Cleft Lip and Palate

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Editor

Cleft Lip and Palate

Diagnosis and Management

Third Edition

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My professional growth has been nurtured by my understanding wife, Lynn, who made it possible for me to spend endless uninterrupted evenings at my desk, while at the same time encouraging me to "stay with it." Warm hugs to my two daughters, Beth and Debra, Ruben and Edward, and my eight outstanding grandchildren for their endless expressions of support and love.

Last, but by no means least, I cannot say enough for the countless children with various palatal and facial clefts whom I have treated over the past four decades and for their understanding parents. This book is dedicated to all of them as a token of my appreciation for their enduring perseverance and fortitude. My young patients have taught me much about the human spirit and the joy that can spring from surmounting nature's adversities.

Finally, my work was made possible by the support of D. Ralph Millard, Jr., who appreciated the value of serial records starting at birth. S.A. Wolfe and I have differed on a few areas of treatment, but we strongly agreed that only through the analyses over time of objective growth records could progress in treatment be accomplished. J.D. Subtelny, Chairman of the Department of Orthodontics at Eastman Dental Center and author of many articles on clefts, and I have been friends for 40 years and at many of our meetings it was a pleasure to hear of his experience treating children born with clefts of the lip and palate.

Foreword

I am deeply honored by the opportunity to write the foreword to this new edition of Dr. Samuel Berkowitz's classic volume on cleft lip and palate.

As a lifelong student of past and current cleft palate literature on facial palatal growth and development he is a strong advocate for good record keeping and carefully conducted clinical studies. He possesses extensive knowledge and insight in the effects of treatment interventions on cleft lip and palate patients and their parents which has led to this book.

Samuel Berkowitz wrote his master's thesis in cleft palate under the supervision and guidance of Samuel Pruzansky at the Cleft Palate-Craniofacial Program at the University of Illinois School of Dentistry in 1959. From there he went to the University of Miami School of Medicine to help develop, with Dr. D. Ralph Millard Jr., Chief of Plastic Surgery, a craniofacial anomalies program and clinic (1960–1998). They collaborated in developing an extensive collection of longitudinal clinical records of dental casts, cephaloradiographs, panorexes and photographs from birth to adolescence. These records are now housed at the Museum of Health and Medicine associated with the Institute of Pathology at Walter Reed Hospital in Washington, D.C. Dr. Berkowitz's main goal was to create lasting treatment concepts based on a better understanding of the natural history of cleft palate and facial growth and development. This book discusses in detail the resulting treatment concepts, which are supported by in-depth case analyses.

During his and my time as clinician providers committed to care of children with congenital facial differences, we have seen major improvements in the overall long-term outcomes in the young adult with a cleft, managed from infancy in an interdisciplinary team setting. Our expectations for the final outcome are normal speech, harmonious facial features, well and stable occlusion and good self esteem. The improvements are due to several factors, many of which are the result of careful clinical studies, long-term observations and the learning and experience that the providers obtain by working together in a team. We have also experienced waves of new clinicians professing new insights into how tissues will respond to certain new or reinvented techniques or manipulations, only many years later to find no improvement in outcomes or even detrimental effects on growth and development. The clinical records, long memory and experience that are found in an established team can have a moderating effect on new members without curbing enthusiasm and creative thinking. It remains a fact that treatment interventions must proceed stepwise

according to biologic development and that there are wide individual variations among individuals with clefts.

In this new edition, Sam Berkowitz has assembled the most outstanding specialists in their specific area of experience and expertise and some new talents. The student of this text will find solid data where that exists and well founded theories where data is not yet available. This text and the teaching materials that Sam has produced to be used in orthodontic specialty programs, attest to his burning desire and commitment to educate future clinical specialists in the care of individuals born with a cleft. He strongly believes that all specialists should know each others problems to better design a meaningful solution.

San Francisco, USA Karin Vargervik

About the Editor

Dr. Berkowitz, an orthodontist, was a Clinical Professor of Pediatrics and Surgery associated with the South Florida Craniofacial Anomalies Program at the University of Miami School of Medicine. He was also Adjunct Clinical Professor at Nova Southeastern University College of Dentistry – Orthodontic Department, and Adjunct Clinical Professor of Orthodontics at the University of Illinois College of Dentistry. His main goal is to develop teaching materials in cleft palate for professionals in plastic and oral surgery, orthodontics, and speech language pathology. He is a past President of the American Cleft Palate Association Educational Foundation, the Florida Cleft Palate Association, was President of the Miami Craniofacial Anomalies Foundation. Dr. Berkowitz was active in the American Association of Orthodontics, Florida Cleft Palate Association, and The Edward Angle Society of Orthodontists. He has published widely in medical and cleft palate journals and is the author of Volume I and the editor of Volume II of *Cleft Lip and*

Palate Perspectives in Management – First Edition; he coauthored *Plastic Surgery of the Facial Skeleton* with S. A. Wolfe, M. D, and wrote *The Cleft Palate Story* for parents of a child born with a cleft. Dr. Berkowitz is a popular speaker on cleft lip/palate topics and has presented many workshops and seminars in the USA and abroad.

His research interest focused on improving surgical-orthodontic treatment planning for cleft lip and palate children as well as those with other craniofacial anomalies. Dr. Berkowitz was project director of a clinical research program that was studying the long-term effects of various surgical treatment procedures on palatal and facial growth and development. He created a quantitative method for determining when to close the palatal cleft space, based on the 10 % ratio of the cleft space to the area of the surrounding palatal surface medial to the alveolar ridges. He has created an audiovisual PowerPoint lecture series for surgeons and orthodontists to enable them to better understand and teach others the effects of surgery on the face from birth through adolescence.

Dr. Berkowitz has been awarded the title "Honoree" by the Edward Angle Society of Orthodontists, and "Honoree" by the First World Congress of the International Cleft Lip and Palate Foundation for his many contributions to the field of cleft lip and palate treatment. His extensive serial clinical records of dental casts, lateral cephaloradiographs, facial and intraoral photographs, and panorexes are in the National Museum of Health and Medicine (associated with Walter Reed Hospital's Institute of Pathology in Washington DC), where they will be available for continued study.

Preface

In the first page of the first edition of this book, I quoted Samuel Pruzansky who, after participating at an International Symposium on Cleft Lip and Palate held in 1969, and reflecting on what he heard at that meeting, stated, "The same tired questions have been asked as at every similar clinical meeting. And I despair at the general unfamiliarity with the pertinent literature."

Fortunately, since the 1950s, many clinical investigators in the field of cleft palate have performed excellent clinical studies of the management of cleft lip and palate that have contributed to the intellectual ferment over the last 50 years. To these studies we are indebted, since to know this literature is vital for correct treatment planning.

When selecting significant references for this text, every attempt was made to carry out an exhaustive literature search to include all of the excellent articles on each subject covered. That, however, has been an insurmountable task. To investigators whose research articles were not included, I apologize and I advise readers to conduct their own literature search, which must include papers on the "opposing schools" of thought. There is no doubt in my mind that their final conclusions will be the same as mine when they consider the results of long-term palatal and facial growth studies that involved the analysis of objective records.

To familiarize clinicians with the appropriate literature and its importance to the treatment of cleft lip and cleft palate, the chapters in this book are structured to improve clinicians' understanding of the natural history of the cleft defect, the face in which it exists, the influence of surgery on palatal growth and development, and equally importantly in developing an appreciation for the heterogeneity that exists even within a single cleft type.

These chapters will show that chronological age is not the parameter that really matters in determining the age at which to close the cleft in the palate. What is important is morphologic age and physiologic fitness, that is, whether the tissues are adequate in quantity and quality and whether the geometric relationship of cleft parts is favorable or unfavorable for reconstruction. Some questions incidental to growth, which date back 25 years, concern the relationship of the malformed palatal segments to the contiguous skeletal anatomy, which, in turn, may be anomalous. These following questions are also addressed: Are the palatal segments static in their deficiency or does the deficiency diminish in time, that is, is "catch-up-growth" a predictable phenomenon? And if so, what surgical procedures (as to age and type) make it possible?

Many of Pruzansky's thoughts, written so many years ago, still hold true today and are worth repeating. He stated that whoever sees things from their beginning will have the most advantageous view of them. To that end, most of the serial cases presented in this volume start soon after birth when plaster casts and photographs of the palatal and facial defect are taken. Serial lateral cephaloradiographs are added as soon as the child is manageable, and again taken periodically through adolescence.

It is hoped that clinicians who are just beginning their involvement in cleft palate will learn the pathology and its natural history of cleft palate from the cases presented in this book and appreciate the need to keep careful records (casts, cephaloradiographs, photographs, and panorexes) which are of vital importance to both the processing of knowledge and self-criticism.

One last note of great importance – it is rare that two members of a team, such as I, an orthodontist, and D. Ralph Millard Jr., a plastic surgeon, can successfully work together even when some differences in treatment philosophy exist. We succeeded because we were professionally compatible and because we shared an obsessive need to determine why some procedures are successful and why others fail even when the same treatment procedures were used. Failures, we discovered, occur principally because of misinterpretation of physiological principles and/or a lack of technical proficiency.

Dr. Millard understood the value of serial objective records dating from birth as the essential starting point in determining the long-term utility of any surgical cleft treatment program. Although I was always free to voice a contrary opinion as to what surgery should be performed (and when), our working relationship was based on recognizing the right of the surgeon to reject recommendations and follow his own dictates. And it was my right, as a member of a team involved in growth studies, to document the anatomical changes to the face and palate for future analysis. Respecting our mutual rights and responsibilities was no simple task. Strong emotional and conceptual barriers had to be overcome in the process of communicating with each other.

Our 40-year search for a better understanding of the natural history of cleft lip/palate growth and development and the effects of various surgical-orthodontic treatment procedures ultimately led Dr. Millard to a conservative approach of staged surgical treatment without the intercession of maxillary orthopedics with periosteoplasty, which he tried and found wanting.

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Also, immeasurable thanks to my many colleagues in The American Cleft Palate-Craniofacial Association and those involved in various cleft palate clinics in the USA, Europe and Asia for contributing to my understanding of cleft lip and palate patient management. To them, too many to recognize by name, I shall be forever grateful for their professional knowledge and personal friendship.

Special acknowledgement needs to be given to my mentor, Dr. Samuel Pruzansky, orthodontist who was director of the Cleft Palate-Craniofacial program at the University of Illinois College of Dentistry when I was a resident in orthodontics. There is no question he motivated me as well as all of those with whom he made contact to go in the right direction to understand the natural history of the face with the cleft palate defect. His aim was to be able to uncover significant information to perform differential diagnosis in treatment planning.

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Introduction

The general aim of this volume is to present recognized experts from the clinical sciences of dentistry, medicine, speech, audiology, psychology, social work, nurses, genetics, ethics, and biology, so that all aspects of the treatment of cleft palate and other craniofacial anomalies can be scrutinized from a particular point of view: long-term clinical experience.

For the sake of brevity, many variations in cleft type and their treatment alluded to in this book were not presented. Because of the multiplicity of variables, no simple description or classification and treatment plan could possibly satisfy everyone concerned with this problem.

Pruzansky (1953) was once asked, "When should the orthodontist's, speech pathologist's, or prosthodontist's interest in the cleft palate child begin?" His response: "The answer is quite clear. Everyone who seeks to serve the needs of the child with a cleft should begin at the beginning." An interest in all events affecting these children is essential to the training and educational experience that each member of the team must obtain. Each specialist emerges not only better informed in his/her own field, but with an increased perspective regarding the means available for providing an integrated program of care for the handicapped child.

The material presented examines the face with a cleft in all aspects as a biologic continuum from birth through postnatal growth and development to maturity at various stages of treatment. In the past several decades, many advances have taken place in cleft habilitation procedures. Unfortunately, many of these changes have not fulfilled all of their stated objectives, and in some instances, these procedures were found to be either injurious or at best unnecessary. These errors will be discussed in detail.

This book also brings together clinicians and biological scientists from the United States, Asia, Europe, and Africa, each of whom in his or her own way has been seeking answers to the multifaceted problem of cleft palate, regarding its embryopathogenesis, craniofacial growth, maxillary orthopedics, surgery, protraction of the maxilla, dental speech prostheses, secondary alveolar bone grafting, speech, hearing, genetics, psychosocial development, and craniofacial surgery.

Each contributor presents pertinent concepts so that a broad perspective of the entire habilitative process can be obtained. The conclusions the reader will reach will be the result of well-documented literature of selected well-controlled clinical research that has withstood the test of review and reexamination.

Because space limitations prevent thorough penetration of all aspects of each subject, a large bibliography is included for additional source material.

In no way could these chapters be expected to cover all aspects of this complex subject.

It is my hope that, through a better understanding of the cleft palate defect and face, all clinicians will be better able to evaluate present-day treatment practices and concepts to better plan their own treatment procedures.

We fully acknowledge the important contributions made by the authors and research programs from the institutions which have strongly influenced much of what has been written in these volumes.

All lip and palate surgery of my cases were performed by Dr. Ralph Millard, Jr., except where otherwise indicated; S.A. Wolfe performed all skeletal surgery and secondary alveolar bone grafting. They both performed superior-based pharyngeal flaps. No presurgical orthopedics were used unless specifically indicated.

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Part I

Facial Embryology and Neonatal Palatal Cleft Morphology

Embryogenetics of Cleft Lip and Palate

1

Geoffrey H. Sperber and Steven M. Sperber

1.1 Introduction

The human features and countenance, although composed of some ten parts or a little more, are so fashioned that among so many thousands of men there are no two in existence who cannot be distinguished from one another. Pliny the Elder, Book 7, Sect 8. AD 23–79 (Harvey 1847)

The basis for most congenital malformations must be found, I think, in hampered development, that is, in arrest at different periods of development. In order to provide evidence for this, it was necessary to complement the pictures of malformations with illustrations of the normal development of the embryo (Vrolik quoted in Oostra et al. 2004) (Fig. 1.1).

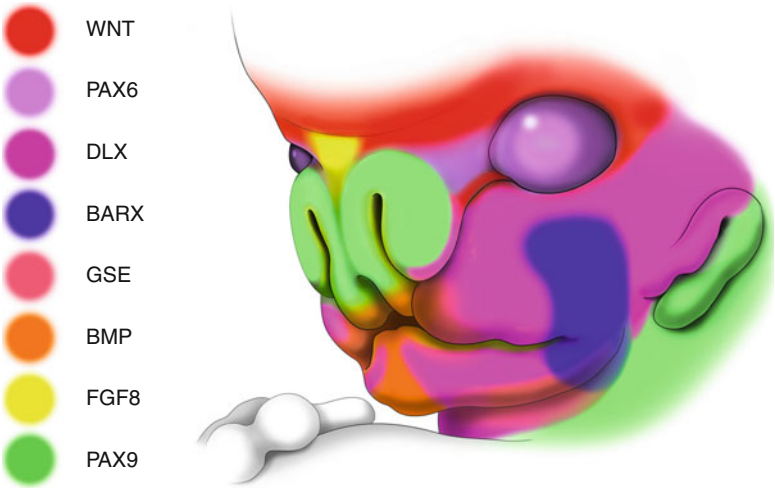

Fig. 1.1 Schematic depiction of gene signaling patterns in a 7-week-old human fetus. The regional colors represent transient gene expression patterns at different stages of embryogenesis (Courtesy of B. Lozanoff, University of Hawaii

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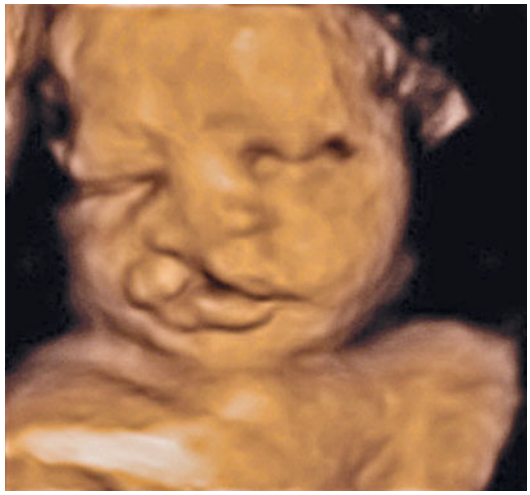

Fig. 1.2 Intrauterine ultrasonography of cleft lip fetus (Courtesy of Dr. Eileen Wang, University of Pennsylvania; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

Thus did Vrolik, more than 150 years ago, lay the foundation for understanding the causes of orofacial clefts. The challenge for modern molecular medicine is to translate the clinically observed clefting defects, the phenotype, back through the intricate developmental phenomena that created them, to the coding genotype. The identification of the genetic predisposition to clefts and the environmental factors that determine and alter the varying threshold of normal versus dysmorphic development are among the central challenges for developmental biologists to decipher. Variations of gene expression regulated by epigenetic mechanisms and variable environments may cause differing expressions of genetic traits (polyphenisms), among which are clefting syndromes. The prenatal diagnostic capabilities of chorionic villus sampling, amniocentesis, fetoscopy, magnetic resonance imaging, and ultrasonography have vaulted gestational developmental phenomena into the field of concern to the clinician (Fig. 1.2) (Liou et al. 2011; Stoll and Clementi 2003; Wang et al. 2011). A new three-dimensional sonographic technique (OmniView) allows study of the fetal hard and soft palates and prenatal diagnosis of clefting (Tonni and Lituania 2012). The maldeveloped intrauterine fetus has now become a potential patient (Jones 2002).

The potential for clefting will ideally be diminished from its initial pathogenetic determination by prevention rather than by post hoc treatment. The basics of biology and molecular medicine will be translated from the laboratory bench to the bedside in the clinical practice of the future.

1.2 Genetics

The mélange of molecular mechanisms involved in the cascading events of embryogenesis are predicated by the expression patterns of specific genes contained in the human genome, constrained by impacting environmental factors. Gene expression patterns are revealing regions of the emerging embryo that have been previously observed histologically and anatomically but not heretofore realized as genetically distinct entities during development. Studies of animal model systems have contributed to our understanding of the molecular determinants that contribute to facial patterning (Swartz et al. 2011). Herein is the marriage of genetics with developmental biology becoming of potential clinical significance.

Current investigations are delineating the complex molecular embryology of development. Specific defects in molecular pathways and networks may provide insights into the etiology of clefting. While embryologists focus on the mechanisms of malformation, deformation, disruption, and dysplasia, clinicians focus on the etiology, diagnosis, treatment, prognosis, and prevention of clefting. The incidence and epidemiology of orofacial clefts provide some clues to the etiology of these malformations (Mossey 2007). The combination of basic and clinical sciences should provide the ideal goal of comprehensive cleft care.

In establishing etiology, it would be useful to have available the gene expression patterns and flow of biochemical pathways underlying morphogenic events. Understanding the local regulation of cellular behaviors and misregulation of any step in these processes can provide insights into clefting consequences. The recognition of

the molecular and tissue elements responsible for normal labiopalatogenesis will allow prognostication of clefting defects in their deficiencies. The therapeutic application of growth factors and gene therapy has the potential for biomimetic preventive and healing regimens (Scheller and Krebsbach 2009). Scarless repair of clefts is now potentially feasible (Larson et al. 2010).

Of the estimated 25,000 protein-coding genes in the human genome, some 17,000 genes have been identified in contributing to craniofacial development (International Human Genome Sequencing Consortium 2004). Engineering advancements in genome sequencing technology with the advent of next-generation massively parallel sequencing platforms has made possible the 1,000 Genomes Project which is cataloguing human genotypic variations in all of these genes (Nielsen 2010). The application of whole exome sequencing has proven to be a powerful tool in its ability to identify genes that influence craniofacial patterning (Ng et al. 2010), with whole genome sequencing being the goal of personal medicine for genetic risk assessment and prevention.

The complexity of contributions of the hundreds of genes to facial formation is being elucidated by identifying each gene's individual expression for each stage of development (Feng et al. 2009). Identification of gene mutations responsible for craniofacial syndromes provides clues to the genetic basis for craniofacial clefting. However, detailed molecular and cellular analyses of the chronology and loci of gene expression patterns, upon which are superimposed epigenetic phenomena, make unraveling the complexity of craniofacial morphogenesis a daunting challenge. The ever-constant new identification of gene expression profiles of embryonic craniofacial and oral structures has led to the development of a consortium titled COGENE (*C*raniofacial and *O*ral *G*ene *E*xpression *N*etwork) that can be accessed online at Cogene: http://hg.wustl.edu/COGENE/ (Cai et al. 2005). Therein is contained a list of all hitherto identified genes, growth factors, and signals involved in the expression profiles of structures between the 4th and 8.5 weeks of human development. It is in the mutation or silencing of genes or the misappropriation of growth factors and signals that the source of some developmental defects is revealed. The intricacies of RNA editing, complex regulatory networks, crisscrossing molecular pathways, together with overlapping and redundancies of gene expression patterns make unravelling the skein of individual influences particularly difficult. A FaceBase consortium providing a comprehensive program of craniofacial research has been established (Hochheiser et al. 2011).

Some of the genes implicated in craniofacial development through human and animal model studies are listed in Table 1.1.

Ascribing specific functions to all these genes is the aim of molecular biology, but it is the realization of the biology encoded within each gene that will provide comprehension of developmental phenomena and their aberrations. Genomic analyses are revealing the molecular architecture behind complex developmental pathways (Brito et al. 2011; Swartz et al. 2011), and the multifactorial basis of the etiology of clefts (Abu-Hussein 2012).

Human genetics can lead to insights of phenotypic diagnosis and provide understanding of the relationships of components of molecular circuitry that will improve the ability of genotypic information to predict the phenotype of complex clefting traits (Dixon et al. 2011; Rahimov et al. 2011; Stuppia et al. 2011; Yuan et al. 2011).

The majority of orofacial clefting cases are nonsyndromic and have no identified cause. Genetic and phenotypic heterogeneity of facial clefting has complicated the identification of the responsible phenomena. Research studies have identified genetic variants associated with cleft lip and palate through linkage analysis, candidate gene approaches, direct sequencing, deletion and duplication analysis by array comparative genomic hybridization (aCGH), and genome-wide association studies (GWAS) (Rahimov et al. 2011; Weatherley-White et al. 2011). A genome-wide association study has identified five genetic loci influencing facial morphology in Europeans: PRDM16, PAX3, TP63, C5o rf 50 and COL17A1 (Liu et al. 2012). This study established links between DNA variants previously associated with non-syndromic cleft lip/palate at 2p21, 8q24, 13q31 and 17q22. The complexity of clefting is illustrated in the numerous different types of genes associated with the phenotype. Such studies have led

Table 1.1 Selected genes implicated in craniofacial development

Gene	Gene/protein	OMIM	Associated syndrome (OMIM)/functional role	Location
ALX3	Aristaless-like homeobox 3	606014	Frontonasal dysplasia type 1 (136760)	1p13.3
ARHGAP29/PARG1	Rho GTPase-activating protein	610496	Candidate gene associated with nonsyndromic CL/P	1p22.1
BMP4	Bone morphogenetic protein 4	112262	Orofacial cleft 11 (600625)	14q22-q23
CLPTM1	Cleft lip and palate transmembrane protein 1	604783	t(2;19)(q11.2;q13.3) translocation described in three-generation pedigree with nonsyndrome CL/CP	19q13
CRISPLD2	Cysteine-rich secretory protein LCCL domain containing 2	612434	Candidate gene associated with cleft lip/palate	16q24.1
DLX5	Distal-less homeobox 5	600028	Proximodistal patterning of the pharyngeal arches; oronasal patterning	7q21.3
EFNB1	EPH-related receptor tyrosine kinase ligand 2	300035	Craniofrontonasal dysplasia (304110)	Xq13.1
FAF	FAS-associated factor 1	604460	Pierre Robin sequence, cleft palate only	1p32.3
FGF8	Fibroblast growth factor 8	600483	Kallmann syndrome 6 (612702); cleft lip/palate	10q24
FGF10	Fibroblast growth factor receptor 10	602115	Aplasia of lacrimal and salivary glands (180920); lacrimoauriculodentodigital syndrome (LADD; 149730)	5p13-p12
FGFR1	Fibroblast growth factor receptor 1	136350	Pfeiffer syndrome (101600), Kallmann syndrome type 2 (147950)	8p11.23-p11.22
FGFR2	Fibroblast growth factor receptor 2	176943	Apert syndrome (101200), Pfeiffer syndrome (101600); craniosynostosis and midfacial hypoplasia syndromes; LADD syndrome (149730)	10q26.13
FOXE1 (TTF-2)	Forkhead homolog-like 15/thyroid transcription factor 2	602617	Bamforth-Lazarus syndrome (241850)	9q22.33
GLI2	GLI-Kruppel family member 2	165230	Holoprosencephaly type 9 (610829), cleft lip and palate	2q14.2
GSC	Goosecoid	138890	Homozygous knockout mouse model exhibits aplastic nasal apparatus	14q32.1
GSCL	Goosecoid-like	601845	Located in the DiGeorge syndrome deletion region	22q11.21
HOXA1	Homeobox A1	142955	Athabaskan brainstem dysgenesis (601536); Bosley-Salih-Alorainy syndrome (601536)	7p15.3
IRF6	Interferon regulatory factor 6	607199	Van der Woude syndrome type 1 (119300); popliteal pterygium syndrome (119500); oralfacial cleft 6 (IRF6 enhancer; 608864)	1q32.3-q41
JAG2	Jagged2	602570	Alagille syndrome; association with cleft palate	14q32
LHX6	Lim homeobox protein 6	608215	Expressed in palatogenesis	9q33.2
LHX8	Lim homeobox gene 8	604425	Associated with cleft lip	1p31.1
MEOX2	Mesenchyme homeobox 2	600535	Posterior palate expression in mouse models	7p21.2
MIR140	MicroRNA 140	611894	Regulates *PDGFA* expression; cleft palate	16q22.1
MSX1	Muscle segment homeobox 1	142983	Oralfacial cleft type 5 (608874); tooth agenesis (106600); Witkop syndrome (189500)	4p16.2

MSX2	Muscle segment homeobox 2	123101	Craniosynostosis type 2; enlarged parietal foramina (168500)	5q35.2
OFC1	Unknown	119530	Oralfacial cleft type 1 (119530)	6p24.3
ORS2	Odd-skipped related 2	611297	Cleft palate in knockout mouse	8q22.2
OTX2	Orthodenticle homolog 2	600037	Anophthalmia, micro-ophthalmia (610125)	14q22.3
PAX7	Paired box homeobox gene 7	167410	Neural crest specification; possible influence on CL/P risk	1p36.13
PDGFC	Platelet-derived growth factor C	608452	Mitogen, associated with CLP	4q32.1
PLCB4	Phospholipase C, beta-4	600810	Auriculocondylar syndrome (602483)	20p12
PTCH1	Patched 1	601309	Holoprosencephaly type 7 (610828), basal cell carcinoma (605462), CLP	9q22.32
PVRL1	Poliovirus receptor-like 1/NECTIN-1	600644	Cleft lip/palate-ectodermal dysplasia syndrome/Zlotogora-Ogur/Margarita Island syndrome (orofacial cleft 7; 225060)	11q23.3
RUNX2	Runt-related transcription factor 2	600211	Cleidocranial dysplasia (119600); implicated in CLP	6p21.1
RYK1	Receptor-like tyrosine kinase	600524	Implicated in oral cleft studies	3q22.1
SATB2	Special AT-rich binding protein 2	608148	Implicated in orofacial clefting	2q33.1
SHH	Sonic hedgehog	600725	Holoprosencephaly type 3 (142945)	7q36
SPRY2	Sprouty homolog 2	602466	Candidate gene for nonsyndromic cleft lip and palate	13q31.1
SUMO1	SMT3 suppressor of mif two 3 homolog 1	601912	Oral facial cleft type 10 (601912)	2q33.1
TCOF1	TREACLE	606847	Treacher Collins-Franceschetti syndrome I (154500)	5q32.q33.1
TBX22	T-box 22	300307	X-linked cleft palate with ankyloglossia (303400)	Xq21.1
TBX10	T-BOX 10	604648	Associated with cleft lip with or without cleft palate	11q13.2
TFAP2A	Transcription factor AP-2 alpha	107580	Branchio-oculo-facial syndrome (113620)	6p24.3
TGFB3	Transforming growth factor beta 3	190230	Cleft palate in mouse model	14q24.3
TGFBR1	Transforming growth factor, beta receptor I	190181	Loeys-Dietz syndrome type 2A (608967), cleft palate	9q22.33
TGFBR2	Transforming growth factor, beta receptor II	190182	Loeys-Dietz syndrome type 2B (610380), cleft palate	3p24.1
TP63	Tumor protein p63	603273	Ectrodactyly, ectodermal dysplasia, and cleft lip/palate syndrome 3 (604292); OFC8 (129400)	3q28
WDR65	WD-repeat domain 65;	614259	Van der Woude syndrome type 2 (606713)	1q34.2
YPEL1	Yippee-like 1	608082	Located in the commonly deleted DiGeorge (188400)/velocardiofacial (192430) region	22q.11.2

to the characterization of several genes with variants that convey an increased risk of orofacial clefting including MSX1, a transcription factor expressed in the anterior palate (Jezewski et al. 2003; van den Boogaard et al. 2000), and TGFβ3, a signaling cue involved in cell migration and palatal shelf fusion (Ashique et al. 2002; Iordanskaia and Nawshad 2011; Lidral et al. 1998). TBX22, which functions in conjunction with SUMO1 (Shi et al. 2009) as a transcriptional repressor, participates in posterior palate osteogenesis (Andreou et al. 2007) and whose mutations cause X-linked cleft palate and ankyloglossia (Kantaputra et al. 2011). Another transcription factor is SATB2, which contributes to osteoblastogenesis and influences expression of transcription factors *Alx4*, *Pax9*, and *Msx1* in the base of the developing palates of mouse models (Britanova et al. 2006; FitzPatrick et al. 2003; Zhang et al. 2011b). The genetic variants contributing to the highest incidence of CL/P (~2 % of all cases) are found in the interferon regulatory factor 6 (*IRF6*) gene, a transcription factor that participates in the differentiation of keratinocytes in the developing epidermis (Ingraham et al. 2006; Kondo et al. 2002; Richardson et al. 2006). Mutations or microdeletions in *IRF6* are responsible for Van der Woude and popliteal pterygium syndromes, both of which exhibit variable phenotypic expression of labiopalatal clefting and lip pit depressions (Jobling et al. 2011; Kondo et al. 2002). A genome-wide meta-analysis of non-syndromic cleft lip with or without cleft palate has identified six new susceptibility regions viz. 1p36, 2p21, 3p11.1, 8q21.3, 13q31.1 and 15q22 (Ludwig et al. 2012). Variants of SKI have been proposed as a candidate gene for non-syndromic clefts of the lip and palate (Mangold et al. 2012).

Genetically determined elements such as facial and nasal width, bizygomatic distances (Boehringer et al. 2011), palatal height, and jaw growth constitute an additive to the genotype that approaches a cleft palate threshold, whereby each element contributes only a small increase in risk. This threshold may be crossed by external factors that include environmental influences, such as smoking, and potential epidemiological factors including maternal stress and age (Fraser 1976; Jagomagi et al. 2010; Wallace et al. 2011; Wehby et al. 2011; Zhang et al. 2011a; Wu et al. 2012). Maternal smoking during pregnancy implicates TGFB3 and MN1 in the etiology of submucous cleft palate (Reiter et al. 2012).

The functional role of many of these genes has yet to be fully established. Before a link between a gene and its expressed phenotype is recognized, there needs to be extensive characterization of the gene's products. The topographical areas and timing of gene expression need to be known for specific regions of orofacial development. Interruption of components of the genetic-metabolic machinery responsible for normal embryonic development can lead to malformations. Disharmonic growth between embryonic components occasioned by subtle differences in the number of cell divisions or in the onset or offset times or rates of cellular activities may variably contribute to dysplasias. The biochemical basis of development and growth changes with time during different stages of development.

In a clinically oriented text, consideration of the very early stages of embryogenesis involving molecular biological mechanisms, induction by signaling factors, tissue differentiation, histogenesis, and organ morphogenesis and growth, each of which constitute enormous fields of study, must be greatly condensed. Appreciation of these underlying developmental phenomena is necessary in understanding the series of cascading events leading from the initial zygote formed by the union of parental gametes to the fully fledged infant (Fig. 1.3). Aberrations or variations from the normal morphogenetic patterns, whether of genetic, epistatic, or environmental origin, are responsible for many of the congenital anomalies that constitute clinical syndromes. Currently, there are no specific tests available for genetic susceptibility to orofacial clefts. The revelation of associated congenital anomalies with cleft lip and palate may further identify the interrelationships of diverse embryonic developments with genetic mutations held in common.

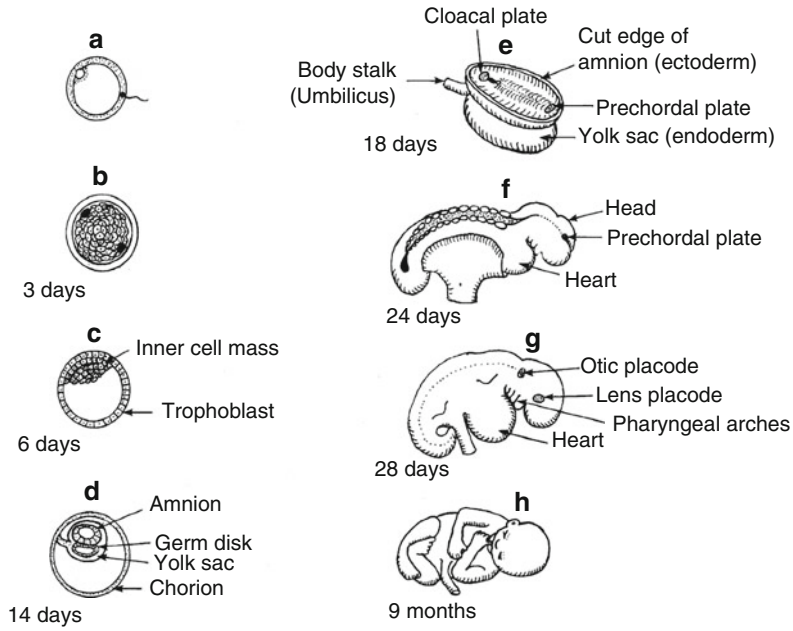

Fig. 1.3 Diagrammatic synopsis of embryogenesis. (**a**) Spermatozoon penetrating ovum to form zygote. (**b**) Morula stage of blastula. (**c**) Blastocyst with inner cell mass. (**d**) Fetal membranes in chorion. (**e**) Primary germ layers forming in germ disk. (**f**) Some stage embryo. (**g**) Postsomite stage embryo. (**h**) Full-term fetus (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

1.3 Early Embryology

The relevance of embryological understanding of facial development is becoming increasingly significant not only for seeking the etiology of orofacial anomalies but also for the application of the molecular mechanisms of normal embryogenesis to the emerging fields of genetic engineering and tissue regeneration. The exploding field of stem cell research for reparative tissue and organ replacement demands an understanding of the morphogenetic mechanisms occurring during facial formation. The recipe for differentiation of stem cells in therapeutic cloning is similar to that of the pathways taken by the multilineage pluripotential cells of the early embryo. The same genes, growth factors, and signaling pathways that operate in the embryo are replicated in directed stem cell differentiation for therapeutic tissue replacement. Fundamental insights into pathophysiology, diseases, and dysmorphology are being revealed by molecular biology (Fig. 1.4).

The field of craniofacial embryology is currently undergoing a paradigmatic period of readjustment and discovery. The last decade has revealed a host of previously unknown factors in embryogenesis. During development, cells are monitored by genetically determined pathways and adjust their rates of accumulation, apoptosis, and hyperplasia to produce organs of predetermined size. The precise control of growth is of inestimable importance for, if each cell in our faces was to undergo just one more cell division, we would be horribly malformed.

The prior presence of the brain determines the subsequent development of the craniofacies. The rostral parts of the brain—the prosencephalon and mesencephalon—are specified by the orthodenticle homologues OTX1 and OTX2, while the HOX genes specify the rhombencephalon and establish spatial identity of prospective craniofacial compartments (Dixon et al. 2011; Larsen et al. 2010; Vieille-Grosjean et al. 1997). It is the brain underlying the future face that is a key component of cephalogenesis (Marcucio et al. 2011).

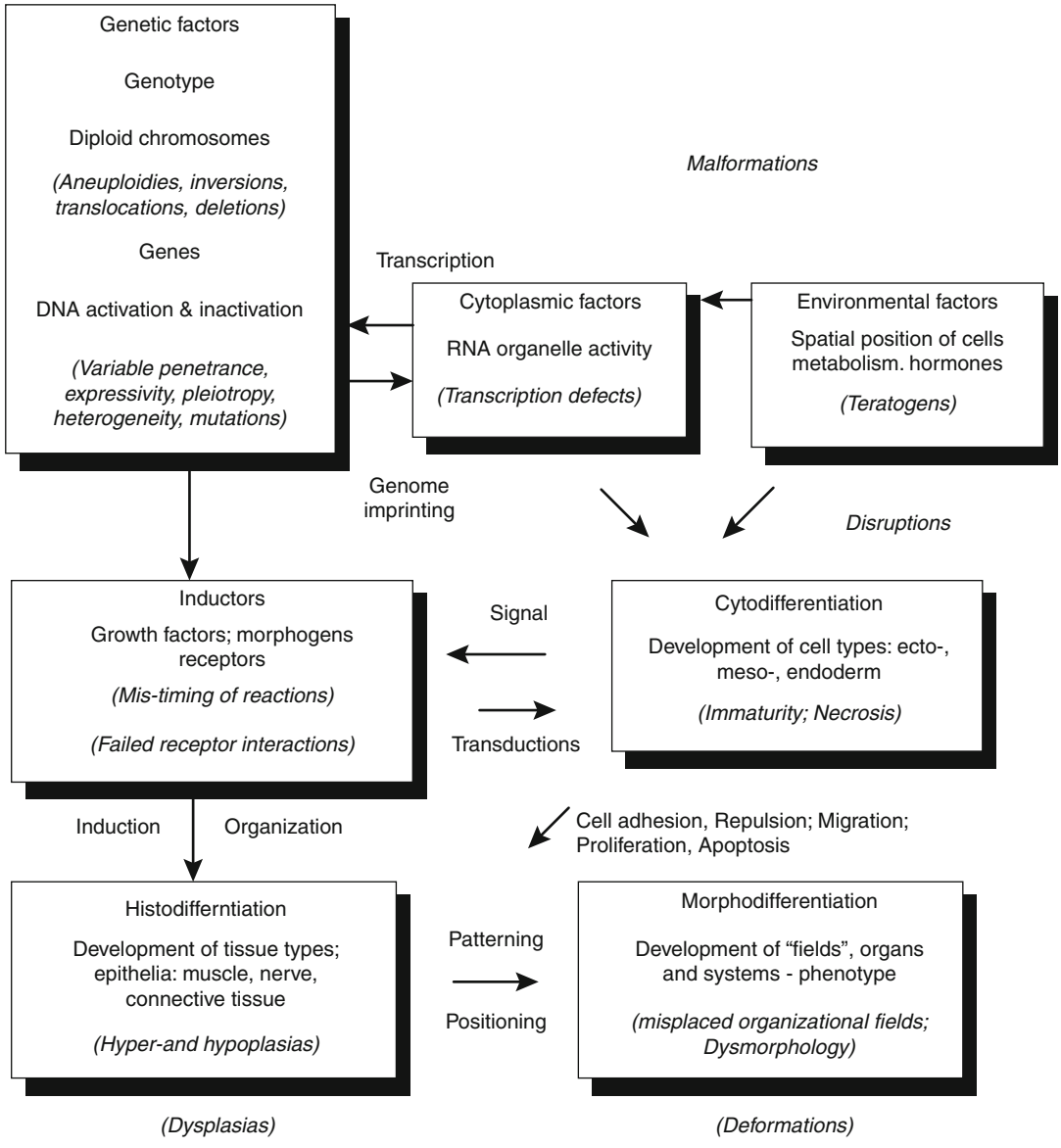

Fig. 1.4 Schema of embryogenesis (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

1.3.1 Organizing Centers

Human and animal model studies show development of the head depends on the inductive activities of the prosencephalic and rhombencephalic organizing centers, which are regulated by the expression of the sonic hedgehog (SHH) gene as a signaling protein in the neural floor plate cells (De Robertis et al. 1991; Odent et al. 1999). These organizing centers are the sites of origin of signaling factors that diffuse into surrounding areas to create "fate maps" that predetermine the details of differentiation of adjacent cells to form particular facial elements. Thus, the rostral prosencephalic center, derived from prechordal mesoderm, located at the rostral end of the notochord, induces the visual and inner-ear apparatus and upper third of the face (the neurocranium).

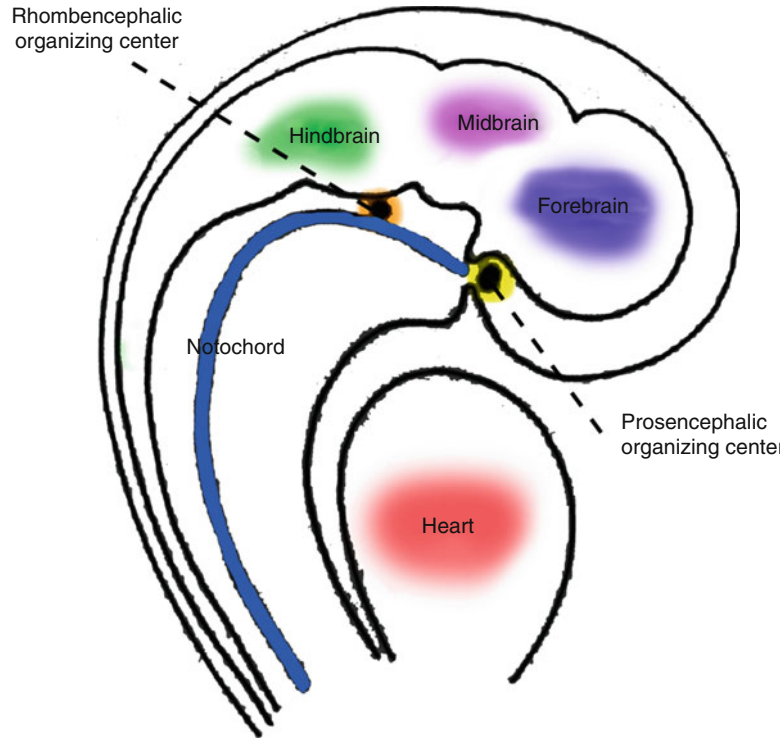

Fig. 1.5 Schematic depiction of prosencephalic and rhombencephalic organizing centers (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

The caudal rhombencephalic center induces the middle and lower thirds of the face, the viscerofacial skeleton (Fig. 1.5). The gradients of chemical and physical properties emanating from the organizing centers regulate craniofacial patterning by inducing a range of responses from uncommitted populations of neural crest tissue (Hu et al. 2003).

1.3.2 Neural Crest Tissue

The major contributor to facial formation is the peculiarly derived mesenchymal tissue that arises from the crests of the ectodermal neural folds that create the brain. Specification of the neural crest by the transcription factor PAX7 occurs very early in embryonic development, even before the neural plate appears (Basch et al. 2006; Betters et al. 2010). The transition of the ectoderm into mesenchyme is a key factor in creating ectomesenchyme that provides a lineage of pluripotential cells that gives rise to diverse tissues (Table 1.2). Facial morphogenesis is controlled by multistep reciprocal interactions between the ecto- and endodermal epithelia and neural crest cells.

Table 1.2 Neural crest derivatives

Connective tissues
Ectomesenchyme of facial prominences and pharyngeal arches
Bones and cartilages of skull and face
Dermis of face
Stroma of salivary, thymus, thyroid, parathyroid, and pituitary glands
Dental papilla, dentin, periodontal ligament, cementum
Muscle tissues
Ciliary muscles
Perimysium, epimysium, endomysium of pharyngeal arch muscles (masticatory, facial, faucial, laryngeal)
Nervous tissues
Supporting tissues
Leptomeninges of prosencephalon and part of mesencephalon
Glia
Schwann sheath cells
Sensory ganglia
Autonomic ganglia
Sensory ganglia of trigeminal, facial, glossopharyngeal, and vagal nerves
Parasympathetic ganglia (ciliary, ethmoid, sphenopalatine, submandibular, enteric system)
Pigment cells
Melanocytes in all tissues
Melanophores of iris

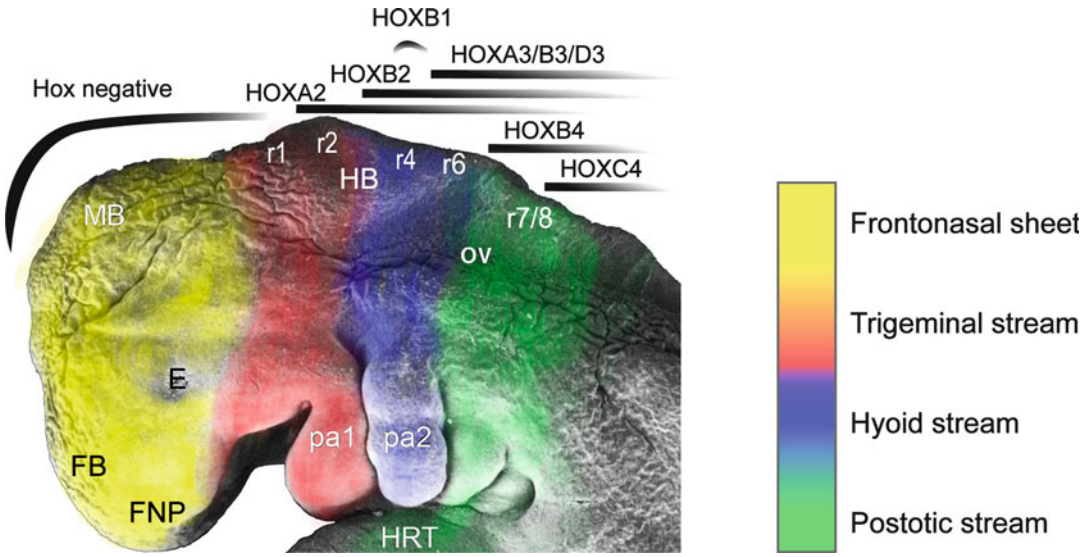

Fig. 1.6 A Stage 15, 33-day-old human embryo upon which are depicted the neural crest streams emanating from the rhombomeres (*r1-8*), influenced by the homeobox (*HOX*) gene expression patterns. *FNP* frontonasal prominence, *FB* forebrain, *E* eye, *MB* midbrain, *HRT* heart, OV otic vesicle, *pa1/2* pharyngeal arches 1/2 (SEM by Prof. Steding, Gottingen. By permission of Springer Verlag; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

The cranial neural crest cells migrate from their initial dorsal location above the rhombomeres of the brain to ventral destinations that are either predetermined by homeobox transcription factor (HOX) genes that constrain their distribution or by responding to local cues from overlying or underlying epithelia (Cordero et al. 2011; Eberhart et al. 2006; Le Douarin et al. 2004; Wilkie and Morriss-Kay 2001). *Collagen type I* and *periostin* expression are implicated in the role of cranial neural crest during soft palate development (Oka et al. 2012).

Segmentation of the rhombencephalon into eight rhombomeres delineates the stepwise sequence of cascading streams of migrating ectomesenchyme to create six pharyngeal arches and five facial prominences (Fig. 1.6). Neural crest mesenchyme migrates in the median plane over the prosencephalon to create the frontonasal prominence. Neural crest tissue from the first two rhombomeres migrates ventrally on either side of the rhombencephalon into the first pharyngeal arch that will give rise to both the maxillary and mandibular arches and their derived skeletal elements.

Crest tissue from the fourth rhombomere contributes to forming the second pharyngeal arch, while rhombomeres 6 and 7 contribute to the third, fourth, and sixth arches. The neural crest overlying rhombomeres 3 and 5 suffers an apoptotic fate mediated by bone morphogenetic protein-4 (BMP4) signaling before migrating and therefore does not contribute to the arches (Smith and Graham 2001).

Developmental studies have shown platelet-derived growth factor A (PDGFA) is required for normal migration of a subset of neural crest cells toward the oral ectoderm that will participate in palate development (Eberhart et al. 2008). Inadequate neural crest mesenchymal proliferation, migration, or excessive apoptosis would result in deficiencies of tissues, causing clefts, among other hypoplasias (Le Douarin et al. 2004; Noden and Trainor 2005; Wilkie and Morriss-Kay 2001). Treacher Collins syndrome is such an example resulting in apoptosis of the specified cranial neural crest due to disruption of RNA biosynthesis caused by mutations in the TCOF1 gene that encodes for the TREACLE, a nucleolar phosphoprotein (Dixon et al. 2006).

1.3.3 Facial Formation

The orofacial region is identified very early in embryonic development at the 28th day postconception, by the appearance of the prechordal plate in the embryonic trilaminar germ disk. This disk is composed of the three primary germ layers, ecto-, meso-, and endoderm. The prechordal plate is characterized by lack of the intermediate mesoderm. The contiguous ectoderm and endoderm at the site of the prechordal plate combine to form a tenuous and temporary bilaminar oropharyngeal membrane that demarcates the location of the future mouth. The ectoderm will form the mucosa of the future oral cavity, while the endoderm will coat the pharyngeal walls. The oropharyngeal membrane identifies the topographic center of facial development by lining a central depression, the stomodeum, the primitive mouth around which there migrate five facial prominences during the fourth week of embryogenesis (Fig. 1.7). The prescient mouth is bordered rostrally by the developing median frontonasal prominence, laterally by the maxillary prominences, and caudally by the mandibular prominences, the latter two both derived from the first pharyngeal arches (Fig. 1.8) (Sperber et al. 2010).

The tissues that constitute the frontonasal, maxillary, and mandibular prominences are comprised of cells of different lineages that have migrated, relocated, and been displaced by epithelial-mesenchymal interactions. Neural crest mesenchyme contributes the major tissue type that combines with core mesoderm and is covered by surface epithelia. The neural crest tissues give rise to the facial skeleton, while the mesoderm will form the facial muscles. Four key morphogens control facial development by regulation of cell proliferation, differentiation, survival, and apoptosis (cell death). Extensive studies have shown that these signaling cues include endothelin (ET1) (Clouthier et al. 2010), fibroblast growth factors (FGFs) (Liu et al. 2010; Szabo-Rogers et al. 2008), sonic hedgehog (SHH) (Hu and Marcucio 2009; Welsh and O'Brien 2009), the wingless (WNT) family (Chiquet et al. 2008; Lin et al. 2011; Reid et al. 2011; Mostowska et al. 2012), and the transforming growth factor beta (TGF-β) family (Iwata et al. 2011) which include the bone morphogenetic proteins (BMPs) (Francis-West et al. 2003; Paiva et al. 2010; Spears and Svoboda 2005).

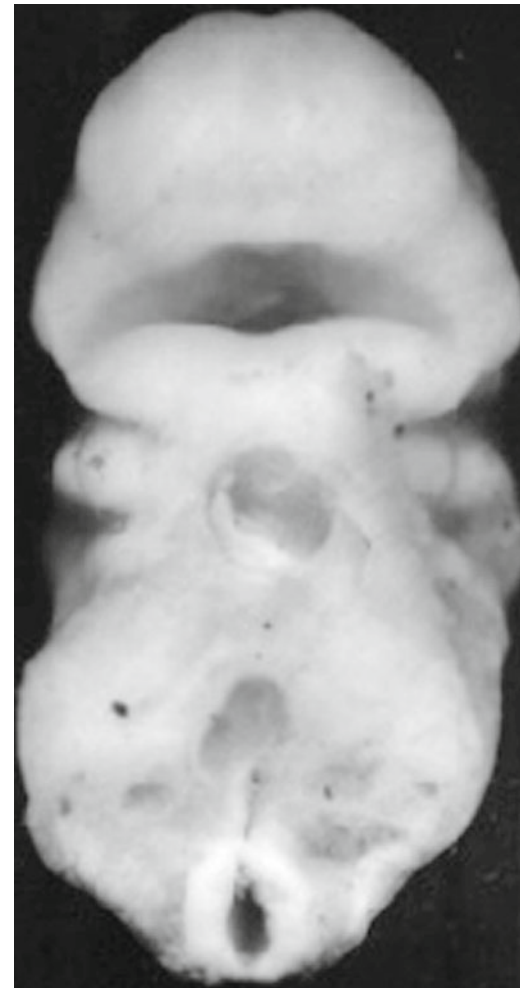

Fig. 1.7 Frontal view of face of a 24-day-old human embryo. ×36 (Courtesy of Prof. Nishimura, Kyoto Collection; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

These morphogens direct signaling pathways that interact coordinately and interdependently to regulate the growth, patterning, and shaping of the

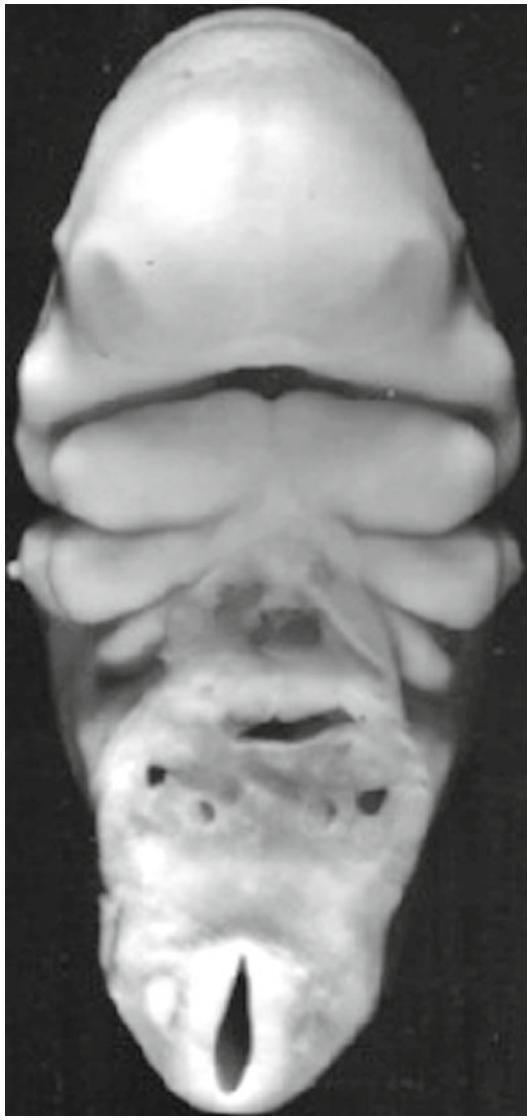

Fig. 1.8 Frontal view of face of a 32-day-old human embryo. ×22 (Courtesy of Prof. Nishimura. Kyoto Collection; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

developing face (Fig. 1.9) (Boehringer et al. 2011; Farlie and Moody 2011; Sperber 2006; Szabo-Rogers et al. 2010). Mutations of genes or misregulation of the signaling pathways results in misappropriated tissue interactions that are the source of facial maldevelopment. The molecular basis for variable expressivity of these genes and factors has not been fully elucidated, but is responsible for the epigenetic spectrum of phenotypic facial malformations. Developmental instability and teratogenic disruption of genetic signaling are other sources of dysmorphic development. Moreover, mechanical pressures must operate within the confines of the epithelial constraints placed upon the expanding mesenchymal components of the facial prominences, influencing their architecture and developing facial features (Radlanski and Renz 2006). A precise mechanistic understanding of the numerous steps involved in signal transductions and migrations is as yet ill-defined.

The mesodermal core of the first pharyngeal arch condenses into myogenic elements that become innervated by the motor branch of the trigeminal nerve. These muscles migrate to their disparate destinations to perform masticatory and swallowing activities. Similarly, second pharyngeal arch mesodermal myogenic elements, innervated by the facial nerve branches, viz., the occipital, temporal, zygomatic, mandibular, and cervical, migrate through the mesenchymal milieu of the facial prominences to establish all the mimetic muscles of the face (Noden and Francis-West 2006). All these dispersed muscles retain their initially established nerve supply. The lingual musculature is derived from migration and elongation of the hypoglossal cord of somatic mesodermal origin, retaining its original hypoglossal (cranial nerve XII) innervation. Appropriate distribution of all these elements of tissue components will formulate a face of normal physiognomy. Deficiencies of the perioral muscles have been demonstrated in microforms and in full-fledged clefts of the upper lip (Jiang et al. 2006; Landes et al. 2006).

The frontonasal prominence, innervated by the frontal branch of the trigeminal nerve, contributes to the forehead and the nose. On the inferolateral corners of the frontonasal prominence, there develop bilateral nasal placodes that differentiate into the olfactory epithelium that interacts with the underlying olfactory nerves. Defective or absent nasal placodal development not only will result in anosmia but has a devastating effect on nasal and central facial development.

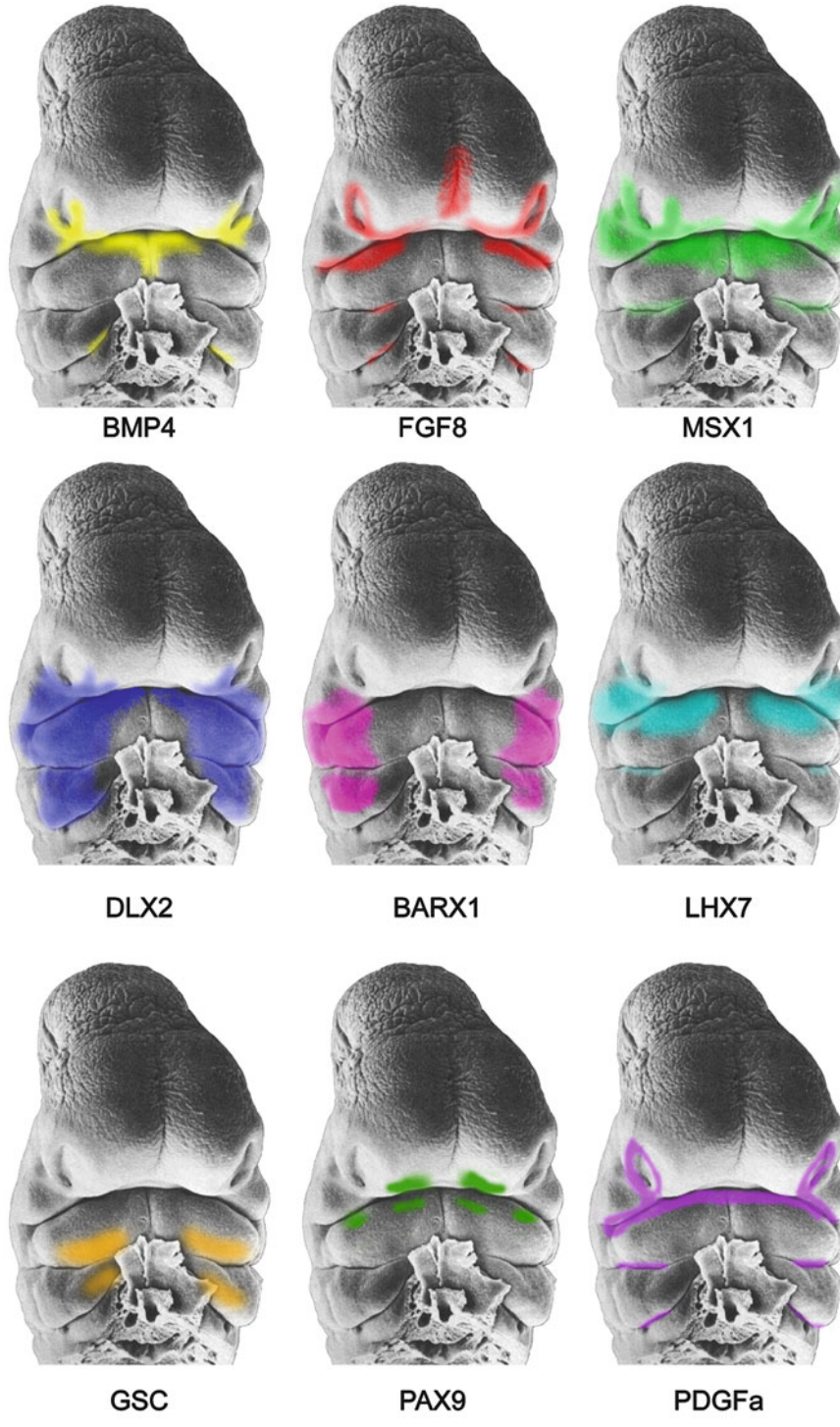

Fig. 1.9 Scanning electron micrographs of the face of a Stage 15, 33-day-old human embryo depicting the gene expression patterns derived from mouse embryos (Faces from Hinrichsen: By kind permission of Springer Verlag; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

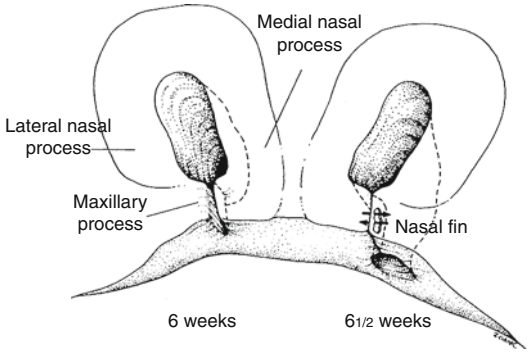

Fig. 1.10 Schematic depiction of breakdown of nasal fin and formation of nostrils. *Arrows* indicate disintegration of the nasal fin between the medial nasal and maxillary prominences (Courtesy of J. Avery and Oxford University Press; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

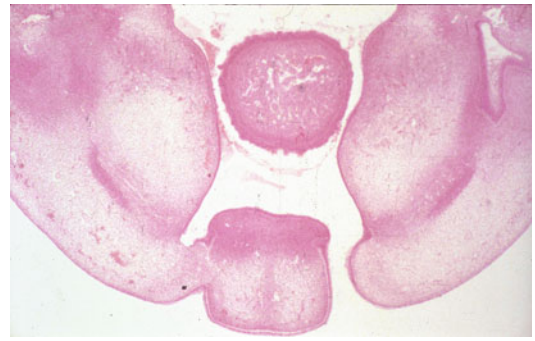

Fig. 1.11 Horizontal section of embryonic lip showing incipient clefting on one side and fusion on the other side (Courtesy of Dr. V. Diewert, University of British Columbia; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

The sinking of the nasal placodes to form nasal pits is the result of the development of the elevated horseshoe-shaped medial and lateral nasal prominences (Fig. 1.10). The posterior aspect of each nasal pit, initially in communication with the stomodeum, becomes separated from the oral cavity by the transient oronasal membrane. This membrane normally disintegrates by the end of the 5th week postconception to open the posterior choanae connecting the nostrils to the posterior oral cavity. Failure of membrane disintegration leads to choanal atresia, a potentially fatal asphyxiating neonatal congenital anomaly.

Elevation of the lateral nasal prominences creates the alae of the nose. The expression patterns of 36 genes are manifested in the medial nasal prominences and those of some 45 genes in the lateral nasal prominences. The location of these genes can be identified on the gene resource locator (Ref: http://grl.gi.k.u-tokyo.ac.jp) (Honkura et al. 2002). Defects of medial nasal prominence development may result in arhinia, or a bifid nose, varying from a simple depression to complete separation of both nostrils. Other nasal malformations include degrees of aplasia of the alae as well as atresia of the nasal fossa(e).

1.3.4 Upper Lip Development

Formation of the upper lip is a complex process involving factors that include PAX9, MSX1, as well as WNT, SHH, FGF, and BMP signaling pathways that pattern cell proliferation and tissue configuration (Jiang et al. 2006; Nakatomi et al. 2010). The upper and lateral boundaries of the primitive oral cavity are formed by the freely projecting maxillary, medial nasal, and lateral nasal prominences. Initial fusion between the lower edges of the horseshoe-shaped medial and lateral nasal prominences completes the large rotund nostril openings. Maxillary prominence growth pushes the widely spaced nostrils medially and converts them into slits. Upper lip completion requires fusion of the bilateral maxillary and the two medial nasal prominences, with the lateral nasal prominences excluded but wedged in between (Fig. 1.11).

The medial tip of each maxillary prominence is initially separated from the inferolateral aspect of each medial nasal prominence by an intervening epithelial "nasal fin" that degenerates, allowing mesenchymal migration across the former boundaries, and seals the initial cleft. Development

of the fusing tissues is the result of cell proliferation, vascular invasion, extracellular matrix production, and fluid accumulation, all of which are subject to variations that may predispose to clefting conditions. Persistence of the nasal fin may contribute to clefting of the upper lip and anterior palate (Fig. 1.10). Although the lateral nasal prominences do not contribute to the upper lip, failure of their initial fusion with the medial nasal prominences is implicated where clefts of the upper lip extend into the nostril (Jiang et al. 2006). All these fusions incur programmed cell death (apoptosis) of the periderm of surface epithelia, epithelial-mesenchymal transformations, filopodial, and adhering interactions. Epithelial filopodia project and anchor into the opposing prominences, followed by mesenchymal fusion. These phenomena are all exquisitely timed and precisely geometrically coordinated to effect the fusions. Inexact contacts by topographic divergences of the prominences or delayed sequences of hierarchical cascading events will inevitably result in clefting of the upper lip.

The initially widely separated median nasal prominences merge in the midline to form an intervening intermaxillary segment, from which is derived the tip of the nose, the columella, the philtrum, the labial tuberculum of the upper lip, the frenulum, and the entire primary palate. The central intermaxillary segment provides continuity to the upper lip, accounting for its maxillary nerve innervation. Upper lip formation commences at 24 days postconception and is completed by 37 days, well within the first trimester of pregnancy. The philtrum and cupid's bow shape of the upper lip form between the third and fourth intrauterine months (i.e., much later than the melding of the maxillary prominences) as a result of collagen condensation in the midline to produce the philtral groove. The philtrum may be congenitally absent when the upper lip lacks a cupid's bow outline, as in the fetal alcohol syndrome. Failure of normal disintegration of the nasal fin by apoptotic cell death or epithelial-mesenchymal transformation is a cause of cleft upper lip, alveolar clefting, and anterior primary palate clefting by preventing the merging of the medial nasal and maxillary mesenchyme. This merging defect may be described as a "differentiation defect," as opposed to a "fusion defect," that becomes clinically significant in the varying degree of dysmorphology exhibited in cleft lip, alveolar clefting, and primary palate clefting. These different degrees of anomalous formation are the result of different time frames in their embryological development that are related to different genes and molecular biological mechanisms operating on lip, alveolus, and primary palate formation (Krapels et al. 2006; Meng et al. 2009; Luijsterburg and Vermeij-Keers 2011). Alteration of developmental timing (heterochrony) accounts for various gradations of severity of anomalous development, from incomplete forme fruste to complete clefting.

1.3.5 Cheiloschisis

Clefting of the upper lip (cheiloschisis) is one of the most frequent of all congenital anomalies; its unilateral incidence (usually on the left) varies among different racial groupings, indicating its inherited character: it is highest in frequency among peoples of Asian descent, intermediate in incidence among whites, and least frequent in blacks (varying from 1:500 to 1:2,000 births) (Derijcke et al. 1996; Mossey and Little 2002). The anomaly appears more commonly in males and has been ascribed to inadequate neural crest tissue migration to the lip area. The degree of clefting varies enormously; the anomaly is rarely median, a characteristic of a major holoprosencephaly syndrome (Mansouri Hattab et al. 2011). A median cleft lip is associated with agenesis of the primary palate and other midline defects. Lip clefts may be coincidentally associated with cleft palate, which has a separate inheritance pattern and a different etiopathogenic pathway (see Sect. 1.5). However, failure of upper lip fusion

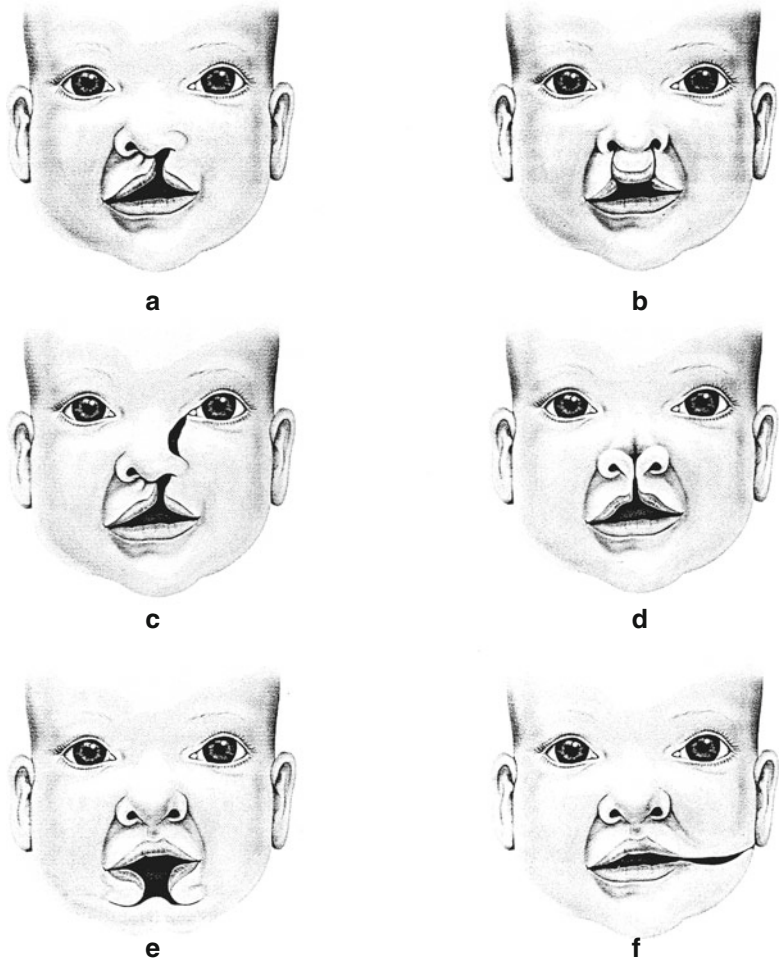

Fig. 1.12 Schematic depiction of potential clefts of the face. (**a**) unilateral cleft lip; (**b**) bilateral cleft lip; (**c**) oblique facial cleft and unilateral cleft lip; (**d**)median cleft lip; (**e**) median mandibular cleft; (**f**) unilateral macrostomia. (Courtesy of G.H. Sperber; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

may implicate the much later-occurring secondary palate conjunctions, accounting for combined cleft lip and cleft palate defects.

Perturbation of the maxillary and mandibular prominences leads to other rare facial anomalies. Normally, the primitive wide stomodeal aperture is reduced by migrating mesenchyme fusing the maxillary and mandibular prominences to form the "corners" of the definitive mouth. Inadequate ectomesenchyme results in macrostomia (unilateral or bilateral), a form of facial clefting, while excessive fusion produces microstomia or astomia usually associated with other congenital anomalies such as agnathia and synotia (Fig. 1.12).

1.4 Lower Lip Formation

Fusion of the bilateral mandibular prominences in the midline creates the continuity of the lower lip. The lower lip is rarely defective, but if so, it is clefted in the midline, contrasting with the more usual unilateral clefting of the upper lip. A rare congenital anomaly of the lower lip (1:100,000 births) is the presence of bilateral pits or fistulae that are pathognomonic of the Van der Woude syndrome, frequently manifesting cleft lip and palate and caused by mutations in the IRF6 and WDR65 genes (Baghestani et al. 2010; Birkeland et al. 2011; Etoz and Etoz 2009; Jobling

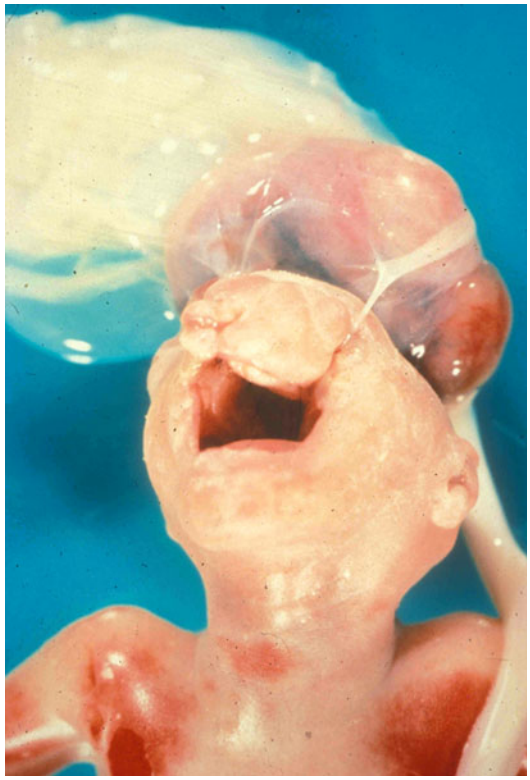

Fig. 1.13 Amniotic band disruption clefts in the face of a fetus (Courtesy of G.A. Machin; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

et al. 2011; Rorick et al. 2011). The embryopathogenetic mechanisms of the associations between upper and lower lip and palate formation in the Van der Woude syndrome are presently unknown.

The rare persistence of the lines of fusion between the maxillary and lateral nasal prominences leads to potential oblique facial cleft(s) in the line of the naso-optic canal. Another potential source of clefting of the face may occur when amniotic bands or strands of connective tissue detach in utero from the amniotic sac, and which the fetus then swallows, tethering the fetal face to the amnion and tearing through the face to form congenital disruption clefts that are unrelated to embryonic fusion lines (Fig. 1.13).

1.5 Palatogenesis

The development of the intact human palate is an evolutionary advance in separating the respiratory and masticatory chambers over the common oronasal chamber (the embryonic stomodeum) that occurs in reptilian and avian antecedents. The hard palate forms a rigid platform against which the tongue can manipulate food and allow pumping action of the linguofaucial muscles to create a vacuum for suckling and swallowing. The soft palate acts as a flexible obturator, closing the nasopharynx from the oropharynx in speech and swallowing. The primary palate develops as a projection into the stomodeal chamber from the median frontonasal prominence with contributions from the medial, lateral, and maxillary prominences (Piotrowski et al. 2011). The secondary mammalian palate is developed as bilateral projections from the maxillary prominences into the stomodeum. The initially separate elements are programmed to fuse. If fusion fails, clefting occurs, and the persistence of the clefted condition may *a priori* be considered an atavistic phenomenon. Cleft palates are the norm in reptiles and birds (Ferguson 1988). Mammalian palatogenesis is directed by an extensive network of interacting transcription factors and signaling molecules emanating from several molecular pathways and different cell types (Bush and Jiang 2012).

The separation of the respiratory chamber (the nasal fossae) from the food ingestion chamber (the mouth) has enabled the development of speech and of leisurely mastication with its accompanying epicurean enjoyment and hence the "palatability" of food. Paradoxically, the hard palate contains no taste buds, although the soft palate does. The anterior oronasal chamber separation defines the palate, whereas the unseparated posterior oronasal chamber serves both respiration and ingestion, accounting for momentary asphyxiation during swallowing. In contrast to mammalian mastication, birds and reptiles characteristically gulp their food to minimize airway impedance.

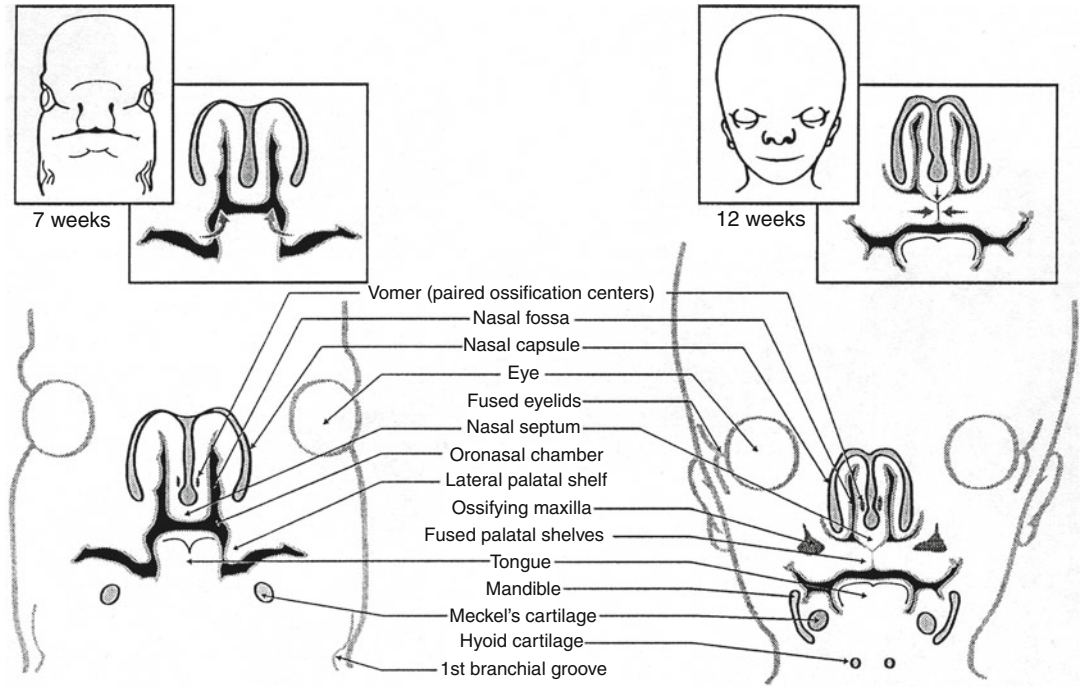

Fig. 1.14 Schematic depiction of midcoronal sections of heads at 7 and 12 weeks depicting palatal lifting (Courtesy of G. H. Sperber; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

The intact palate is composed of three embryological elements derived respectively from the median frontonasal prominence, the primary palate, and the two lateral palatal shelves from the maxillary prominences forming the secondary palate. These elements are initially widely separated in the confining stomodeal chamber by the intrusive developing tongue. The continually growing lateral palatal shelves are deflected down vertically on either side of the tongue, prior to the eighth week of development (Fig. 1.14). As a result of growth of the stomodeum at the beginning of the fetal period (8th week), and the occurrence of mouth opening reflexes, the tongue is withdrawn from between the vertical shelves. The mechanical withdrawal of the tongue requires functioning of the hyoglossus muscle, necessitating neuromuscular and jaw joint activity (Fig. 1.15). All these factors are gene initiated and environmentally dependent on critical timing, which, if perturbed, disrupts the precision of palatal fusion.

The palatal shelves, in a short period, flow like a wave into the horizontal plane, enabling them to establish contact with each other in the midline, with the primary palate anteriorly and with the lower edge of the perpendicular nasal septum. Thereby, the single stomodeal chamber is subdivided into the upper nasal fossae and the lower oral cavity (Figs. 1.16 and 1.17). The conjunction of the shelves with the nasal septum may be unilateral, leading to asymmetrical cleft palate, with only one nasal fossa opening into the mouth.

Shelf elevation occurs by a number of mechanisms, including biochemical transformations of the physical consistency of the connective tissue of the shelves, blood flow into the shelves increasing tissue turgor, and differential proliferation of mesenchyme, creating mechanical elevating forces. The increase in tissue turgor pressure depends on a critical role played by hyaluronic acid that interacts with several extracellular matrix proteins. The gene CD44,

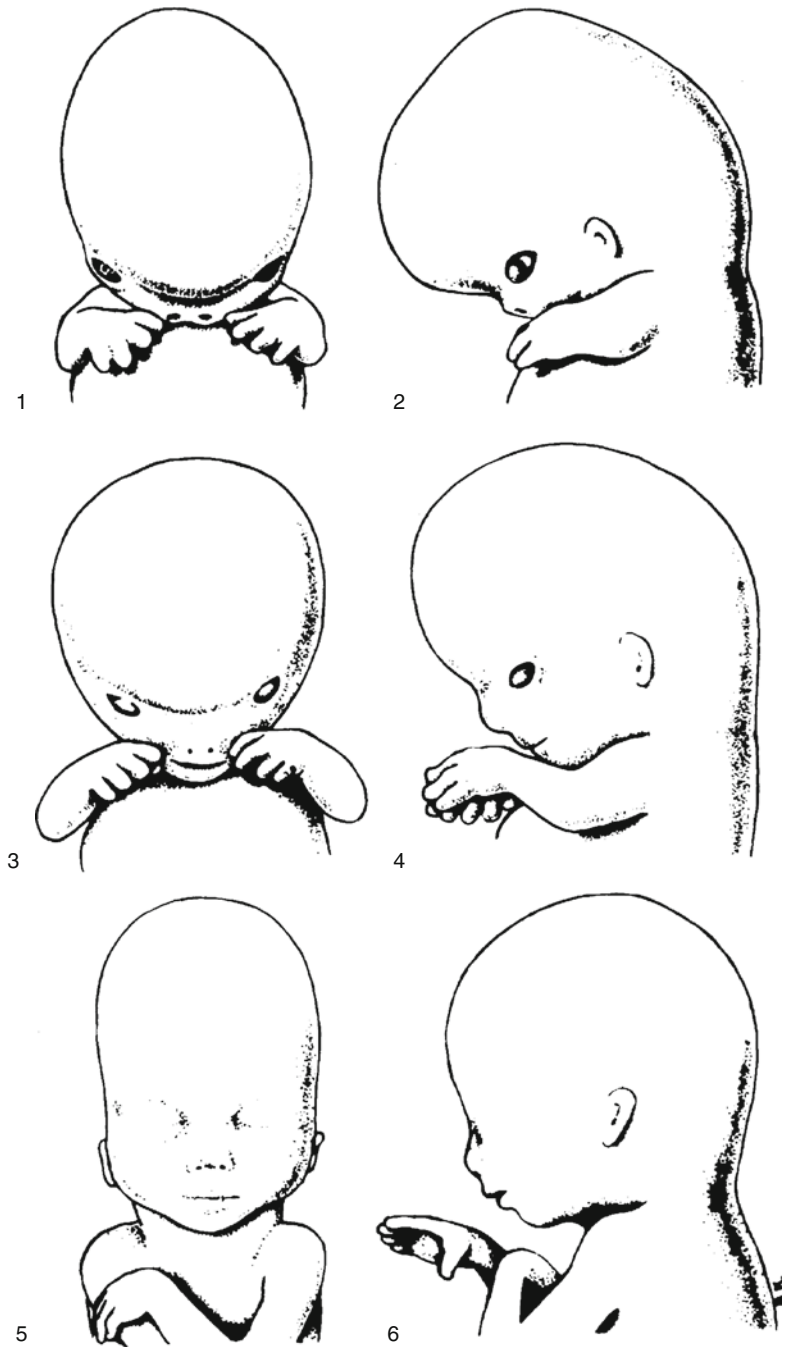

Fig. 1.15 Schematic depiction of embryonic head movements from frontal and lateral perspectives at 6, 7, and 8 weeks (Courtesy of Dr. V. Diewert and Oxford University Press; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

the major hyaluronan receptor of the hyaladherin family, is transiently and dynamically expressed during secondary palate development (Oliveira and Odell 1997). Recent evidence in animal and human studies has indicated that platelet-derived growth factor (PDGF) signaling is a new and independent mechanism that regulates palatogenesis by the lifting of the palatal shelves

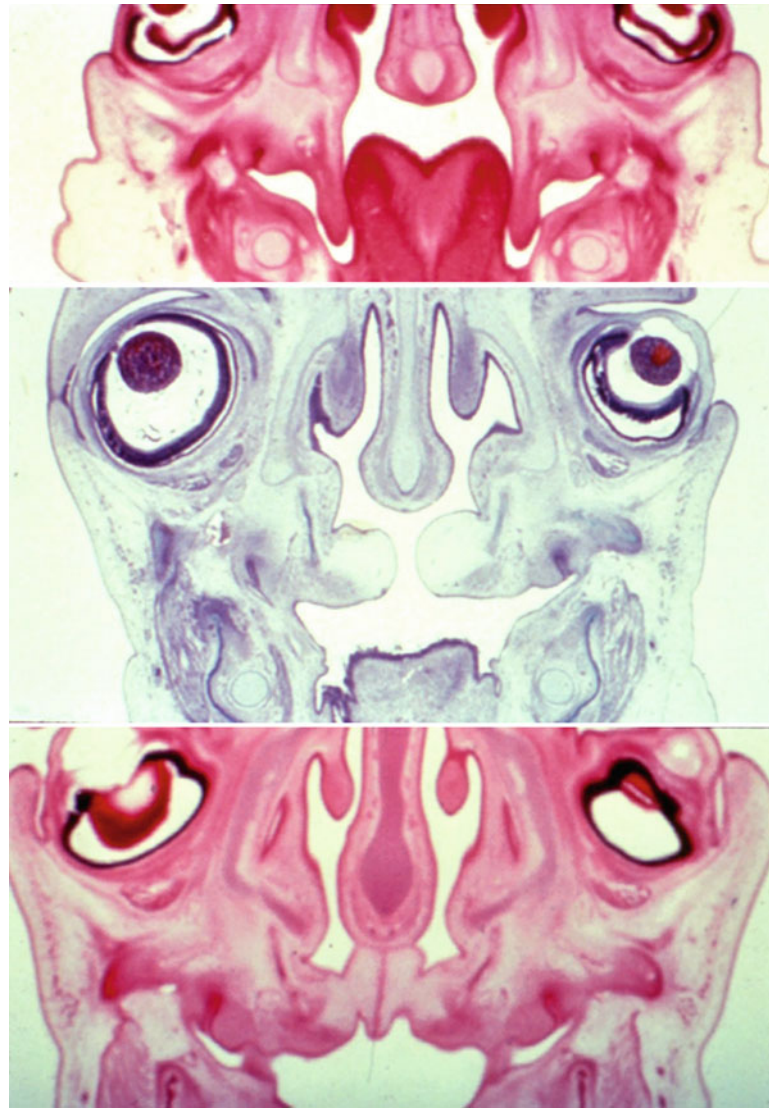

Fig. 1.16 Coronal sections of embryos at 7.5 (*top*), 8 (*middle*), and 9 (*bottom*) weeks depicting palatal shelf elevation and fusion (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

and their fusion (Ding et al. 2004; Yu and Ornitz 2011). Insights into this palatogenic network are illustrated in the disruption of a microRNA, miR-140 that regulates PDGF signaling and induces cleft palate in humans (Eberhart et al. 2008; Li et al. 2010). Wwp2 has been implicated as a regulatory interactor between Sox9 and Mediator 25 in the transcriptional mechanisms of chondrogenesis in the forming palate (Nakamura et al. 2011).

Developmental studies indicate that sonic hedgehog (SHH) signaling is a key factor in the outgrowth of the palatal shelves that is also stimulated by the activity of FGF10, a growth factor in the mesenchyme that stimulates a receptor, FGFR2b, in the surface epithelium. The epithelium, in turn, increases its SHH signaling back to the mesenchyme (Rice et al. 2004). Exquisite reciprocal signaling between the palate epithelium and mesenchyme directs palatal shelf growth and morphogenesis. When the palatal shelves meet in the midline, the surface periderm of the medial edge epithelial cells undergoes apoptosis, and an epithelial-mesenchymal transformation

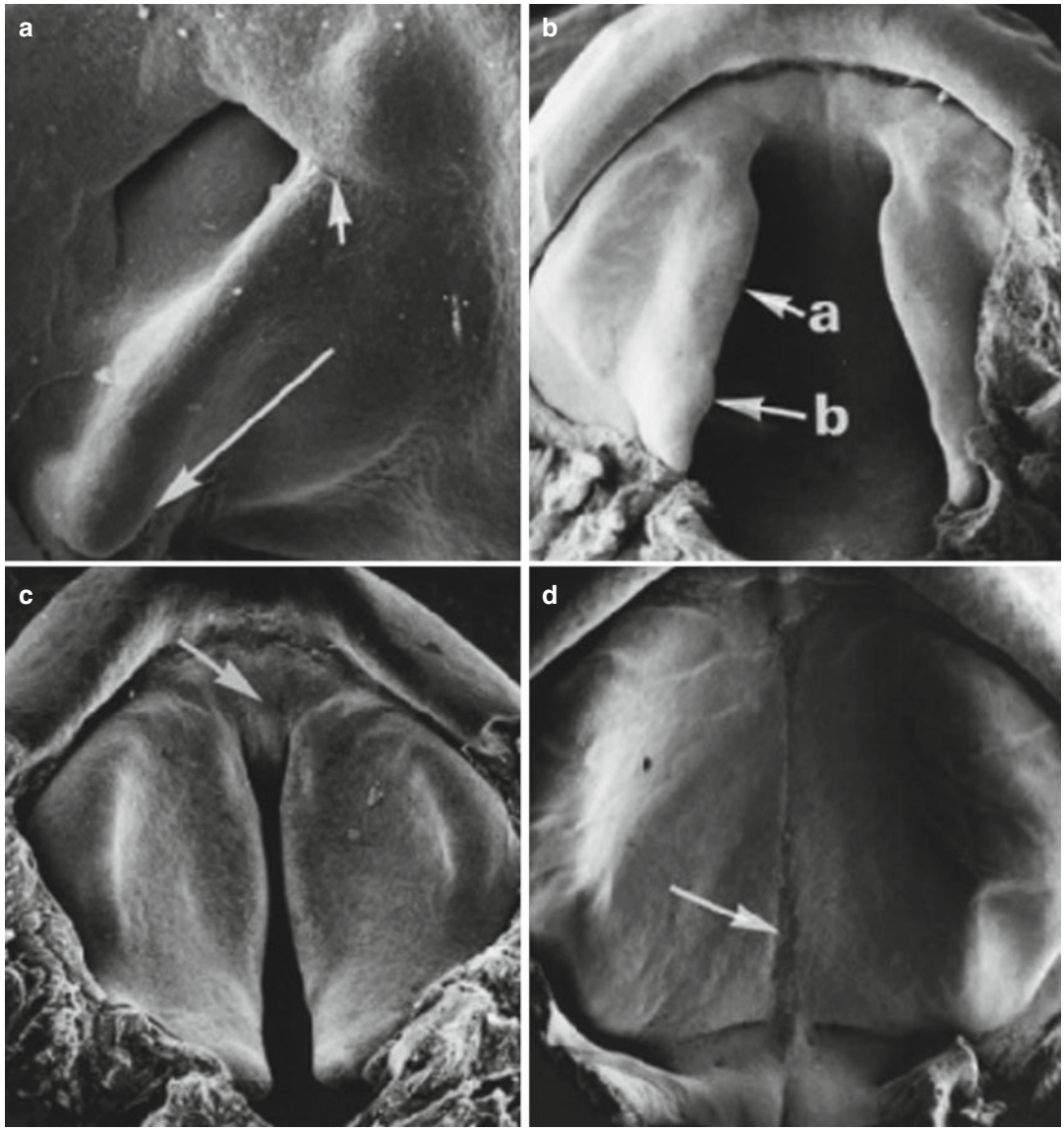

Fig. 1.17 (**a**) View of right half of a 6-week-old human embryo showing early vertical palatal shelf and lip forming from the maxillary prominence; short arrow, site of primary palate; long arrow, posterior edge of secondary palate (SEM×30). (**b**) Palatal shelves of a 7-week-old human embryo showing anterior end of right shelf (*a*) becoming horizontal, while the posterior end (*b*) remains vertical (SEM×11). (**c**) Palatal shelves of an 8-week-old human embryo showing the horizontal shelves approaching each other and the anterior primary palate (*arrow*) (SEM×11). (**d**) The nearly fused palatal shelves of a 9-week-old human fetus. The soft palate region (*arrowed*) is still unfused (SEM×8) (Scanning electron micrographs from Shaw et al. (1978). By permission; Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

occurs in a complex mechanism (Hay 2005). The transforming growth factor-β (TGFβ) signaling pathway, active in the medial edge epithelium, participates in the fusion of the palatal shelves and promotes the epithelial-mesenchymal transformation during palatal fusion (Bush and Jiang 2012; Fitzpatrick et al. 1990; Greene and Pisano 2010; Iordanskaia and Nawshad 2011; Ito et al. 2003; Iwata et al. 2011; Xiong et al. 2009; Nakajima et al. 2010; Iseki 2011).

The medial edge epithelium of the palatal shelves is of particular significance in establishing fusion. There exists a genetic heterogeneity along the anterior-posterior and medial-lateral axes of the developing palate. Different regulatory mechanisms exist for the fusion of the anterior versus the posterior region of the palate. The boundary between the anterior and posterior regions of the palate is defined by the most posterior of the four palatal rugae, composed of regularly spaced transverse ridges of epithelial thickenings underlain by mesenchyme (Pantalacci et al. 2008; Welsh and O'Brien 2009). The rugae are stripes of sonic hedgehog (SHH) expression that are interspersed by inhibiting fibroblast growth factor (FGF) expression, reflecting an activator-inhibitory pair indicating a Turing-type reaction–diffusion spacing mechanism (Sohn et al. 2011; Economou et al. 2012). These rugae contain a variety of sensory cells (Nunzi et al. 2004) and cranial neural crest stem cells (Widera et al. 2009). Multiple genes (Msx1, Bmp4, Bmp2, Fgf10, and Shox2) show restricted expression in the anterior region of the palate (Bush and Jiang 2012; Hilliard et al. 2005). *FGFR2* is expressed in the epithelium and mesenchyme of the middle and posterior palate (Porntaveetus et al. 2010). FGF8 induces *Pax9* expression in the posterior region of the palatal mesenchyme (Iwata et al. 2011; Snyder-Warwick et al. 2012). The epithelial-mesenchymal transition of the epithelial layers on opposing shelves requires transforming growth factor-β3 (TGFβ3) as the most prominent inducer and ephrin-mediated signaling to promote mesenchyme proliferation and fusion (San Miguel et al. 2011). Programmed cell death (apoptosis) of the epithelial layers is induced by the Fas ligand (FasL)-Fas-caspase extrinsic apoptosis pathway to allow fusion to occur (Huang et al. 2011). The fusion seam initially forms anteriorly in the region of the hard palate, proceeding both rostrally and caudally to complete merging in the soft palate region. The most posterior of the rugae defines the boundary between the hard and soft palates. A combination of apoptotic surface epithelial cells and a surface coat of glycoproteins and desmosomes facilitate epithelial adherence between the contacting palatal shelves (Cuervo and Covarrubias 2004). Disintegration of the seam is facilitated by changes in protein content, cell migration and apoptosis (Vukojevik et al. 2012).

Epithelial-mesenchymal transformation of the underlying basal epithelial cells may be a factor for mesenchymal coalescence of the shelves (Vaziri Sani et al. 2005). Exquisite control of the program is illustrated by a dosage-dependent role for Spry2, an antagonist of FGF signaling, that participates in the modulation of the growth and patterning of palate development (Welsh et al. 2007). During palate closure, the mandible becomes more prognathic and the vertical dimension of the stomodeal chamber increases, but the lateral maxillary width remains stable, allowing shelf contacts to occur. Elongation of the hard palate and the sequential addition of rugae, in mouse models, are induced by Wnt-β-catenin signaling (Lin et al. 2011; Pantalacci et al. 2008). Failure of glycoprotein adhesiveness and transformation of epithelial cells into mesenchyme, allowing epithelial persistence, are factors contributing to palatal clefting.

1.5.1 Ossification

Intramembranous ossification of the palate commences in the 8th week postconception from de novo ossification and centers initially separate from primary ossification centers in the maxillae. The palatine bones ossify separately and with the palatal centers spread centrifugally to create the hard palate. A number of genes are activated during osteogenesis (Shh, Ihh, Ptc1, Gli1-3, Runx2, Alp, Bmpr1, Col1a1, and GSK-3β) (Baek et al. 2011; Levi et al. 2011; Nelson et al. 2011). Premaxillary ossification sites appear transiently in the primary palate region but fuse with the maxillary ossification centers by the end of the third fetal month, losing their separate identity. These primary palate ossification sites persist in nonhuman primates, leading to premaxillary bones that do not exist in humans. The intervening sagittal midpalatal and the coronal transverse maxillopalatine sutures designate the different bones of the palate. The midpalatal suture is a site

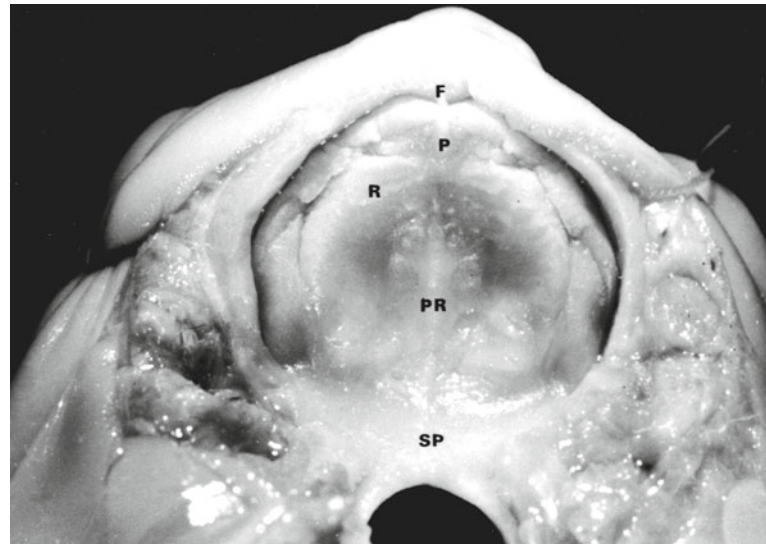

Fig. 1.18 Palate and upper lip of a 22-week-old fetus. Note the developing rugae in the hard palate and the extensive soft palate posteriorly. *F* frenum, *P* palate, *PR* palatal rugae, *SP* soft palate (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

exploited by orthodontists by widening for rapid maxillary expansion in cases of narrow palates that cause dental crowding. Defective bone formation, after proper fusion of the palatal shelves, results in submucous cleft palate, a frequent clinical anomaly.

Ossification does not occur in the most posterior part of the palate, giving rise to the region of the soft palate. Myogenic mesenchymal tissue of the first and fourth pharyngeal arches migrates into this faucial region, supplying the musculature of the soft palate and fauces. The tensor veli palatini is derived from the first arch (trigeminal nerve), and the levator palatini, uvular, and faucial pillar muscles are derived from the fourth arch (vagus nerve) (Fig. 1.18).

1.5.2 Palatoschisis

> If I do not remember thee, let my tongue cleave to my palate. Psalm 137:6

Failure of fusion of the palatal shelves on either side of the midline results in cleft palate. Palatal clefting may be syndromic, that is, associated with other developmental anomalies, or nonsyndromic, with no other developmental anomalies. These clinical designations do not provide understanding of the intrinsic causes of clefting. The complexity of palatogenesis accounts for the relatively common occurrence of its failure, resulting in clefting. The vulnerability of the palate to clefting is indicative of its relatively recent evolution and is susceptible to a variety of environmental impacts acting on a background of genetic predispositions to clefting (Yuan et al. 2011).

Clefting of the palate is a consequence of several factors impeding the closure and fusion of the three palatal elements. They include absence or deficiency of the tongue-depressing hyoglossus muscle, a *Hoxa2* gene loss-of-function effect seen in mouse models, allowing continued impedance of shelf elevation by the tongue (Barrow and Capecchi 1999). The tongue normally flattens but remains highly arched in cleft palate cases. Notch signaling that includes the Notch family of receptors and the delta-like and jagged ligands are necessary for palatal fusion. Homozygous knockout mice for *Jagged2* exhibit palate-tongue fusions (Jiang et al. 1998; Xu et al. 2010). Failure of palatal shelf elevation has been attributed to a number of other genetic mutations characterized in mice notably deficient for *Fgf10*, *Pax9*, and *Msx1* (Alappat et al. 2005; Peters et al. 1998; Zhang et al. 2002).

Even having elevated, the persistence of the epithelial seam between the shelves creates conditions for clefting and leave remnants of epithelial "pearls" that may become cysts. The

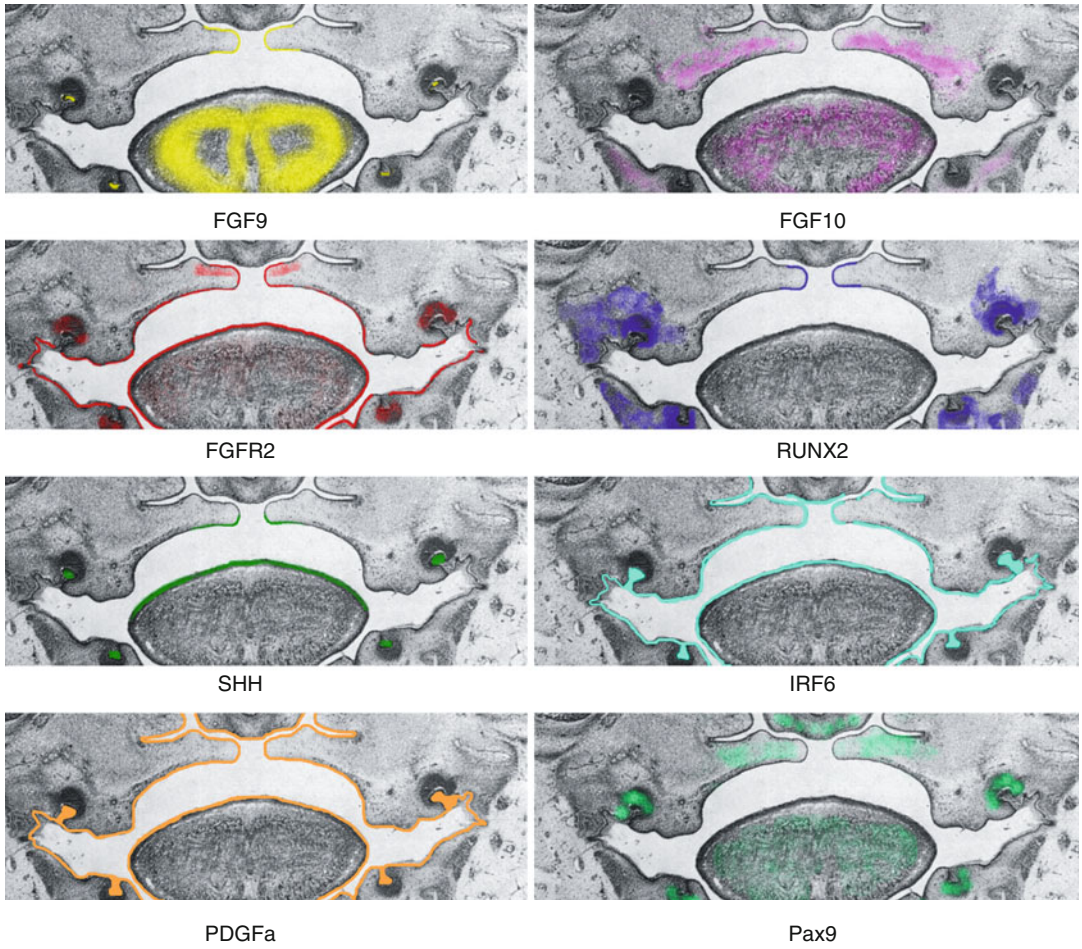

Fig. 1.19 Midcoronal sections of an 8-week-old human embryo upon which are depicted the gene expression patterns derived from mouse embryos (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

complex interactions of numerous genes, transcription factors, and signaling transductions are being revealed in several recent reports on the phenomena of palatogenesis and palatoschisis (Figs. 1.19 and 1.20) (Lan et al. 2004; Letra et al. 2011; Liu et al. 2005; Rice et al. 2004; Sasaki et al. 2004; Stanier and Moore 2004; Yu et al. 2005). Disruption of the differentiation and adhesion competence of the medial edge epithelium, even with apposition of the palatal shelf antimeres, can result in clefting of the palate.

The shape of the cleft in the palate is indicative of its etiology. V-shaped clefts are the consequence of inadequate tissues in the shelves to complete closure. U-shaped clefts are usually associated with micrognathia and glossoptosis (Robin-type clefts) resulting from the tongue obtruding between the shelves, preventing their elevations (Hanson and Smith 1975). A genetic contribution to Pierre Robin sequence resulting in a cleft palate is shown in the perturbation of the *FAF1* gene, an enhancer of apoptosis expressed in the pharyngeal arches and necessary for cranial neural crest differentiation into cartilage (Ghassibe-Sabbagh et al. 2011).

The least severe form of cleft palate is the bifid uvula, of relatively common occurrence and seldom clinically significant. Increasingly severe

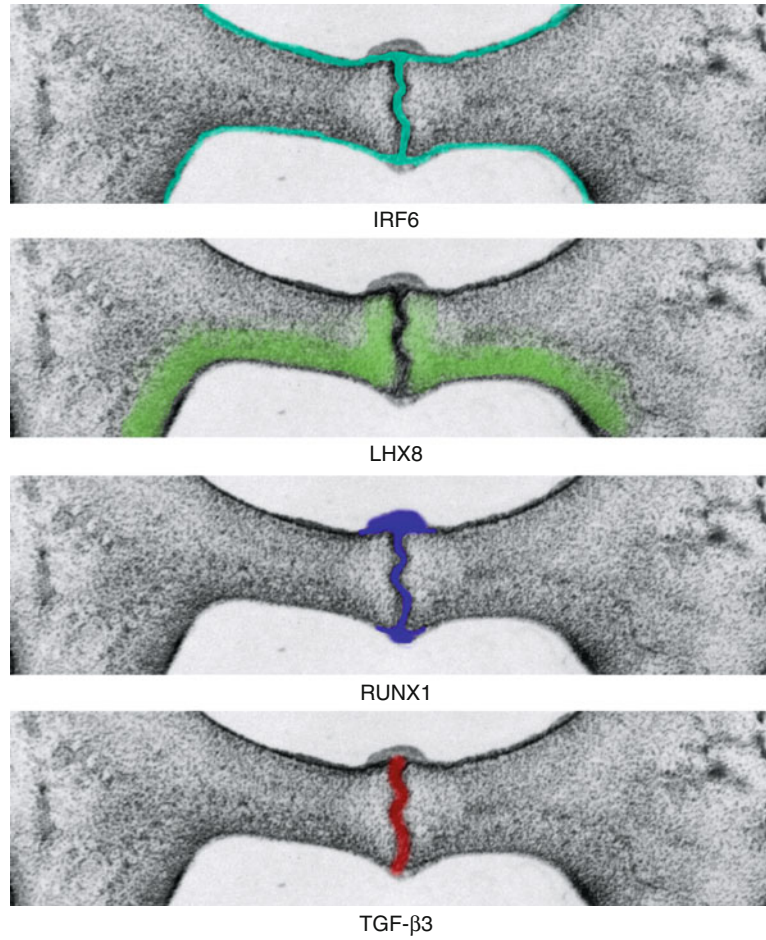

Fig. 1.20 Midpalatal fusion gene expression patterns derived from mouse embryos (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

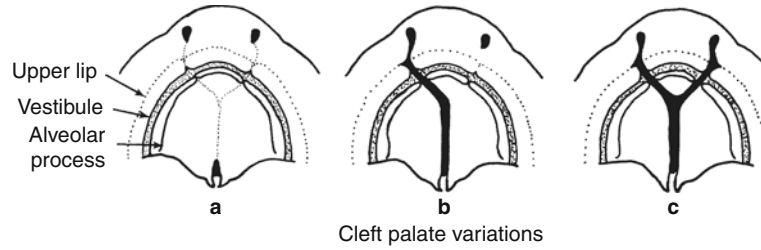

Fig. 1.21 Schematic depiction of degrees of palatal clefting. (**a**) Bifid uvula. (**b**) Unilateral cleft palate and lip. (**c**) Bilateral cleft palate and lip (Reproduced by kind permission of McGraw-Hill from Losee and Kirschner (2009))

clefts always incur posterior involvement, the cleft advancing anteriorly in contradistinction to the direction of normal fusion (Fig. 1.21). Deficient fusion posterior to the incisive foramen derive from the secondary rather than the primary palate, whereas anterior clefts incur primary palate deficiencies. Secondary postfusion clefting of the palate may develop prenatally as a confounding etiological phenomenon (Arnold et al. 1998). Clefts may be submucous in nature, involving muscle discontinuity, yet with an intact overlying mucosa.

The consequences of palatal clefting are multifarious, ranging from oronasal food regurgitation, speech impediments, dental malocclusion, facial growth impedance, and social isolation.

Conclusion

The preceding insights into orofacial development provide clinicians with a rationale for understanding the occurrence of clefts as deviations of normal morphogenesis. With the advent of identification of chromosomes, genes, and growth factors responsible for development of the orognathofacial complex, clinical geneticists, speech pathologists, and surgeons are in a better position to predict, prognose, and diagnose clefts of the face, lips, and palate. The anticipation of biomimetic intervention by genetic engineering and molecular growth factors in producing scarless healing of cleft surgical repair is becoming ever more realistic (Larson et al. 2010). The current explosion of molecular biology encompassing genomics, proteomics, metabolomics, and pharmacogenomics for targeted drug therapy will have a profound impact upon the prognostication, treatment modalities, and prevention of labiopalatal clefting. The rapid advances in our understanding of cellular behavior during embryonic development leading to differentiation and morphogenesis provide opportunities to exploit this knowledge in preventive, curative, and regenerative healing applications. The current cornucopia of cognitive diagnostic capabilities provided by genetics, immunohistochemistry, cloud computer networks, obstetrical ultrasonography, CAT scanning, nuclear magnetic resonance (NMR) scanning, 3D computer stereology, next-generation sequencing technology, high-throughput gene expression profiling, and mass spectrometry provide the potential for prenatal diagnosis and possible therapy of palatofacial clefting. Nonetheless, much remains to be done for these techniques to be translated into clinical practice and is the central challenge of laboratory bench to bedside transition technology.

1.6 Websites

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The Value of Longitudinal Facial and Dental Casts Records in Clinical Research and Treatment Analysis

2

Samuel Berkowitz

After 40 years of treating children with various types of clefts, this author has concluded that the success or failure of a surgical procedure depends on the degree of palatal cleft defect at the time of surgery and the resulting facial growth pattern as well as the surgical skills and the surgical procedure utilized. This conclusion will not be new to the experienced orthodontist who in all probability recognizes that the progress recorded in treatment depends, for the most part on the skeletal and facial growth patterns inherent in the patient and the interaction of surgery with facial and palatal growth.

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2.1 Serial Cephaloradiographs and Casts of the Maxillary and Mandibular Dentition and Occlusion

To properly assess the results of treatment, there is a fundamental need for serial casts, lateral cephalometric films, and photographs in individual case reports.

Pruzansky (1953, 1955) often stated that it is unfortunate that plastic surgeons' training in the realm of clefts and their variations tends to be totally inadequate because their first encounters with patients usually occur in the clinic or operating room. Furthermore, there is seldom recourse to anatomical specimens to better appreciate the nature of the cleft deformity. The trainee is dependent on the empirical experience of his preceptor for knowledge of the natural history of the defect and long-term response to therapy. In most cases, other than before and after facial photographs, there are no objective records to determine why the outcome was a success or failure.

The collected serial data to be shown in this text will provide the clinician in training with an overview of the variations that can be encountered in each cleft type, the significance of genotype differences that influence growth and response to surgery, and the natural history of each cleft entity.

Over the years certain cephalometric measurements have become standardized and have

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been applied to selected population samples to develop statistical means or averages. In the treatment of cleft lip and/or palate, this approach has provided useful data in studying morphologic growth changes in the head, evaluating dentofacial abnormalities, and assessing responses to surgical and orthodontic treatment. The data has been particularly useful in determining the timing and type of procedure selected to treat individual problems. The measurements and analyses utilized are primarily profile-oriented and reveal both anteroposterior and vertical relationships of the various parts of the dentofacial complex.

To assess changes during the course of general growth and treatment, head radiographs of the same individual taken at separate times are traced and the tracings superimposed to ascertain the changes that have occurred. A common method is to register the two tracings at the point sella with the sella-nasion lines superimposed (Fig. 2.2a, b). This method provides a gross overview of changes in the dentofacial complex and in soft tissue but is useful only in evaluating what has already occurred. In this text, we also use the Coben superimposition procedure (basion horizontal) because it more accurately reflects actual craniofacial growth direction (Figs. 2.1d and 2.2c).

The use of "landmark," or baseline, images associated with the basicranium to show the composite results of facial growth can provide meaningful information because it is the enlargement of the face relative to the cranial base that is being evaluated. In the child, further growth changes in the anterior part of the cranial base slow considerably at about 5–6 years of age, whereas facial growth continues actively through adolescence or beyond. Comparing the relative growth between these two regions, rather than simply focusing on a single fixed point, provides clinically useful information when cephalometrically evaluated.

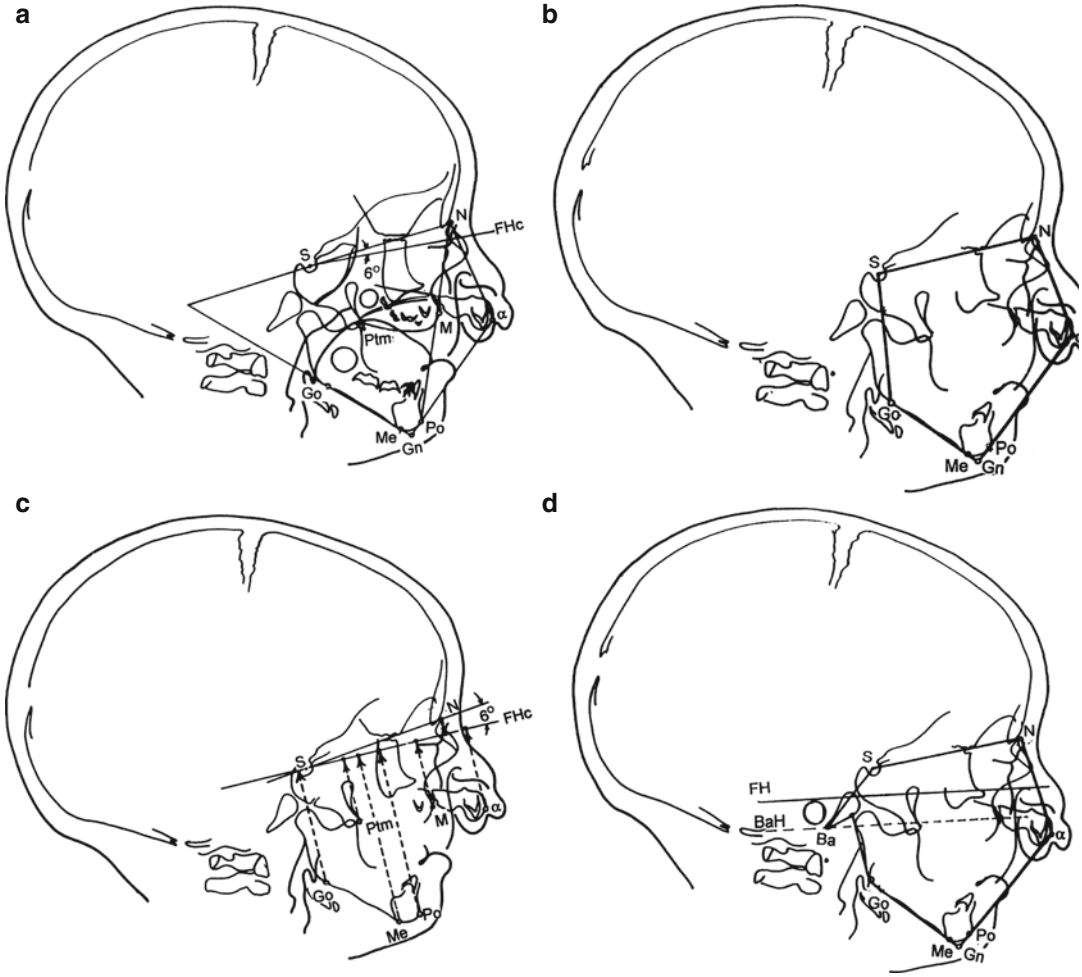

Fig. 2.1 (**a**–**d**) Various methods used to demonstrate facial changes using lateral cephalometrics. (**a**) Facial angles. These are just a few of the angles which describe changes in the skeletal profile. There are many more angles and linear measurements which can be used to relate the maxilla to the mandible and both jaws to the cranial base. (**b**) Facial polygon. This is a graphic method used to describe the boundaries of the skeletal face. (Pogonion constructed, Po', is the same point as gnathion.) Facial growth changes can be shown by superimposing each succeeding polygon on the anterior cranial base (*SN*) and registering on sella turcica (*S*). (**c**) Projecting facial landmarks to a constructed Frankfort horizontal line which is arbitrarily drawn 6° off the SN line. This angle can vary with steepness of the anterior cranial base. This graphic method will show the relative contribution of various structures within the maxilla and the mandible to the profile. (**d**) Basion horizontal created facial polygon (Coben 1986). This method of superimposing tracings graphically reflects his overall concept of fixed growth. A plane at the level of the anterior border of foramen magnum (basion) parallel to Frankfort horizontal where Basion is the point of reference for the analyses of craniofacial growth

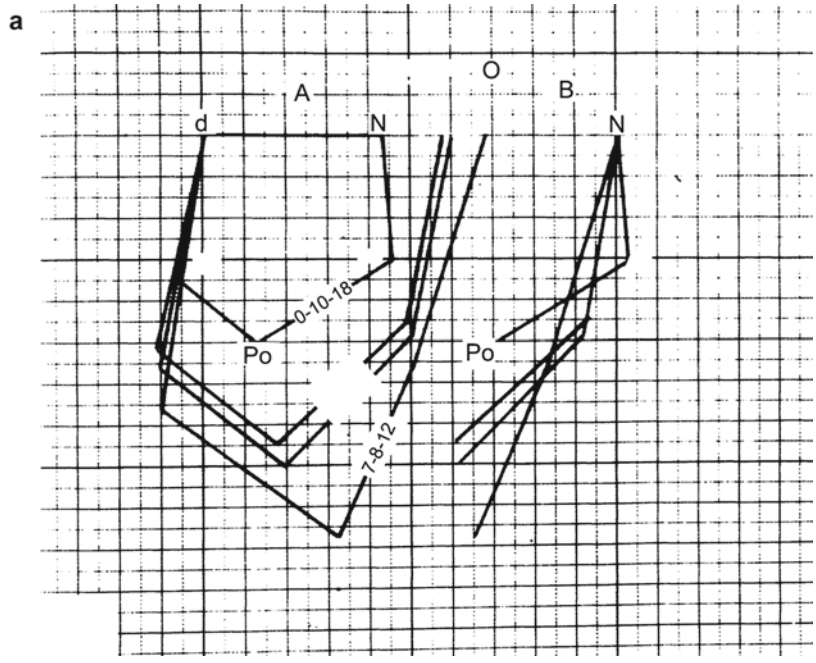

Fig. 2.2 (**a**) Case CP #127 (CPCLP). Superimposed facial polygons from 1 month, 18 days of age to 7 years, 8 months, and 12 days of age. A result of the mandible's downward growth increments exceeding its horizontal growth increments, this is an example of "poor" facial growth in that the profile fails to flatten as the mandible remains retrognathic. Note that in this and the following illustration, the forward projection of the premaxilla does not increase after 1 year, 2 months, and 22 days

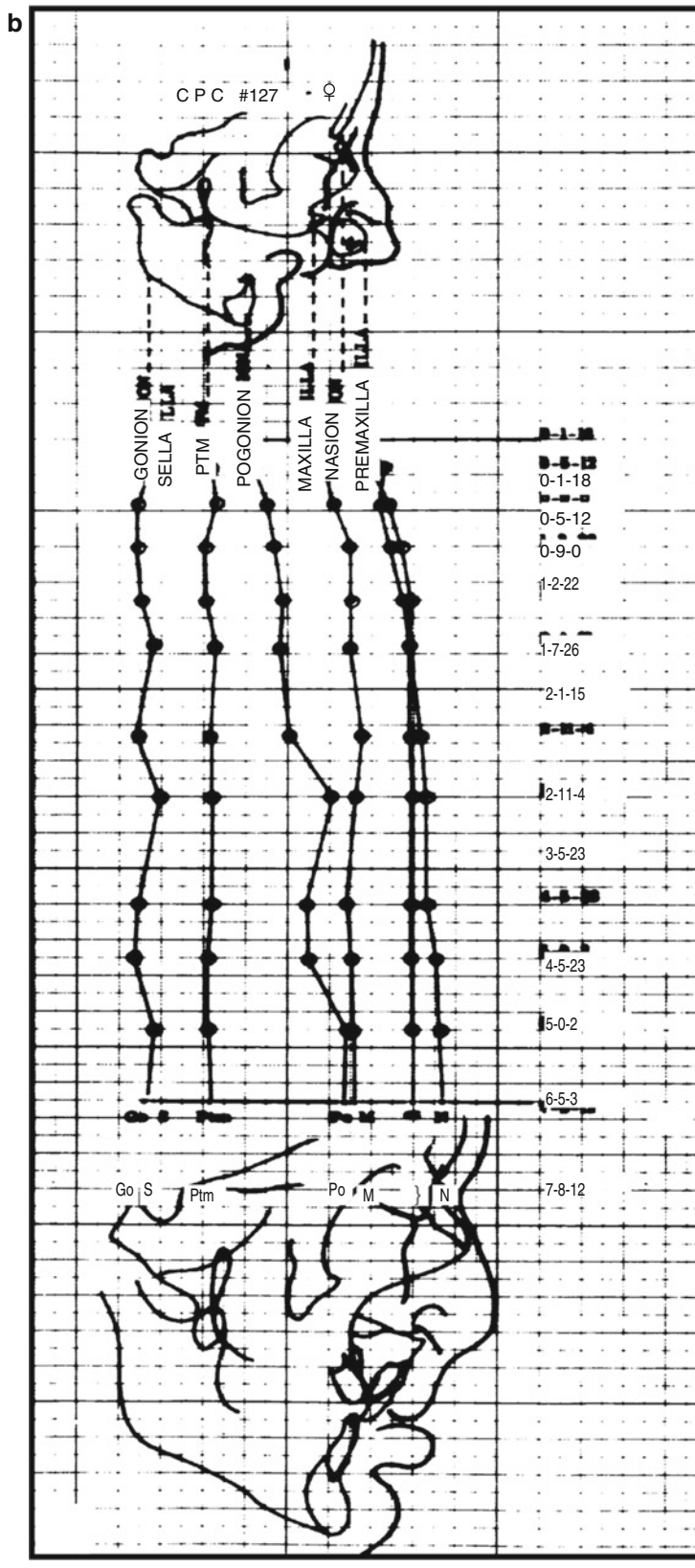

Fig. 2.2 (*continued*) (**b**) Case CP #127. Projecting facial landmarks to a constructed Frankfort horizontal 6° off the SN line. Although each of the skeletal structures except for the mandible has increased in size, the relative position of midfacial structures to the anterior cranial base has remained relatively stable

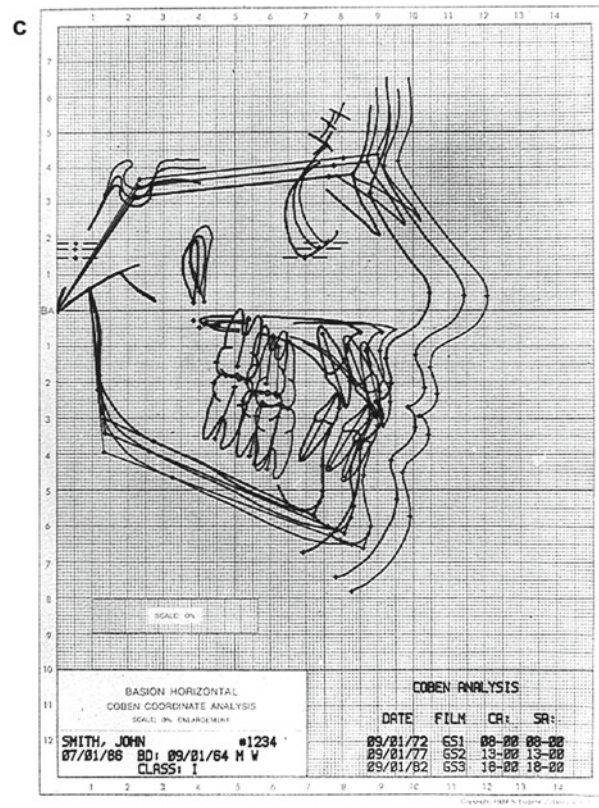

Fig. 2.2 (*continued*) (**c**) Case CP #127. Basion horizontal coordinate computer craniofacial serial tracings at ages 8, 13, and 18 years. Tracings are registered at Basion and oriented in Frankfort horizontal. Serial tracings maintain a constant S-N/FH relationship. S-N and FH planes are parallel. Tracings depict Coben's growth philosophy, which states that craniofacial growth is reflected away from the foramen magnum (basion) and the vertebral column (Reprinted from Coben (1986))

2.2 The Beginning of Longitudinal Cleft: Palate Research Studies

Two major research problems were common in cleft palate surgical studies prior to the 1950s. Pruzansky (1953, 1955) commented on the surgeon's tendency to group all types of clefts together in research and clinical treatment. He also stated that surgeons were limited in their study of pathologic anatomy of clefts due to the unavailability of serial dental casts, cephaloradiographs, and photographic records.

The need for clinical records was apparent to many researchers, and within a decade, many retrospective clinical data sets were developed. These data sets spawned many investigators to determine the long-term influences of surgical and neonatal maxillary orthopedic procedures on palatal and facial growth and development. As a result of these early studies, useful diagnostic and prognostic information was obtained that provided a rationale for the management of individual cleft cases. These clinical records offered an accurate means for measuring and recording individual variation and for plotting the progress of each case in terms of growth and response to various treatments. As a result of these findings, the quality of care improved, resulting in more aesthetic and functional outcomes. Proper documentation, using objective records and individual treatment outcomes, has extended to many more modifications where it is possible to perform multicenter retrospective studies.

2.3 Research Methods (Atkins 1966; Byse et al. 1983)

2.3.1 Retrospective Studies

In a retrospective study, the nature of the study group must be delineated precisely. Definite criteria should be established so that there is no

ambiguity about types of cases and stages of growth development to be included in, or excluded from, the study. The choice of the case and control groups should be guided by concerns of validity. The advantages of retrospective studies are that they can be conducted relatively rapidly because the records of patients whose treatment is already complete can be used. The investigator is protected against the circumstance of "subject dropout" during the course of treatment, and they are relatively economical.

2.3.2 Prospective Studies

The advantages and disadvantages of prospective studies are in essence the inverse of those of retrospective studies. Provided that ethically and logistically satisfactory plans for random assignment to treatment can be developed, prospective trials afford an opportunity to control selection bias and to define and control the records acquisition process.

The main disadvantages of prospective trials are that they are expensive and a great deal of time must inevitably elapse between project initiation and the point at which data on most of the main outcome variables become available for analysis.

Multicenter comparisons of surgical orthodontic treatment outcomes are an efficient way of testing the effectiveness of various treatment philosophies and surgical techniques. Differences among surgeons, variances in performance by the same surgeon over the years, and differences in techniques are difficult to identify and compare in isolation. However, in multicenter clinical studies, differences in clinical procedures among operators can, within defined limits, be compared and evaluated successfully without arousing criticism.

2.3.3 Clinical Trials

A clinical trial may be defined as a carefully designed prospective study that attempts to answer a precisely defined set of questions with respect to the effects of a particular treatment. A clinical trial is a major undertaking which requires considerable money, personnel, facilities, time, and effort.

The simplest design for a clinical trial involves randomization between two different surgical treatment regimens to answer one specific question; for example, which of two surgical procedures is the most beneficial. To add a larger number of surgical procedures makes the trials more difficult to manage.

There are two reasons for not using a randomized clinical trial (RCT) method for surgical evaluation of cleft closure procedures whether done as a multicenter or single-center trial. The first is the need for the surgeon to disregard the unique nature of the individual cleft defect and perform a standard surgical treatment being tested, the presumption being that clefts of all sizes and shapes will react the same way to the same surgical procedure. The second reason concerns the ethical questions involving the sequencing of surgical procedures and the use of the surgeon's skills.

2.3.3.1 Randomization of Surgical Procedures

In proposed multicenter RCT, it is expected that each surgeon will randomly utilize various surgical procedures sequentially for each type of cleft to determine the relative differences in outcome between procedures.

With the present restraints on certain types of human research, Human Subject Research Review Committees in most settings would be reluctant to permit the use of various elective surgical procedures in a research setting if there is a possibility that a surgical procedure might lead to facial disfigurement. Most surgeons would reject participating in a study employing a particular procedure they already have used and found to be inadequate. Many surgeons see the choice and timing of cleft surgical procedures as varying with the geometric characteristics of the palatal defect; therefore, the concept of randomization cannot be considered as an alternative to what they are already doing. The factor of surgical skill in a randomized trial must be considered as a variable in determining the effectiveness of a procedure. Can

all participating surgeons be equally skilled in all procedures?

2.3.3.2 The Ethics of Surgical Retrospective Clinical Trials (RCT)

It is impossible to disassociate scientific from ethical considerations when dealing with cleft palate research (Gifford 1986; Hellman and Hellman 1991; Israel 1978; Kukafka 1989). Different research protocols and evaluation methods carry different ethical problems, the more so when life or death issues are not being considered.

2.3.3.3 Informed Consent

When a patient is deemed appropriate for a particular clinical trial, a first step is often to obtain informed consent. This is a legal requirement in the USA, but not in all countries. In some European countries, each participating hospital decides on whether and how to handle informed consent.

Informed consent is a social construct based on ethical guidelines and supported by legal precedents. In order for consent to be legally valid, it must be obtained voluntarily from a mentally competent person of legal age.

The greater the seriousness of the potential injury, even if the risk is minimal, the greater the obligation to inform the patient (or parent). The greater the chance of a risk occurring, even if the injury would be minimal, the greater the obligation to inform the patient (parent). The more elective the proposed treatment, the more serious injury will be perceived.

Sheldon Baumrind (1993), summarizing the role of clinical research in orthodontics which is also applicable to cleft palate research, states:

> Cogent arguments can be made concerning the ethics of conducting structured clinical experiments in the kinds of long-term therapeutic situations which interest orthodontists. One telling argument is that since therapists have an absolute and transcendent obligation as professionals to deliver for each patient the treatment which they believe best for that patient, no subject can ethically be randomized to one of two possible treatments unless there is true uncertainty as to which of the two treatments is in the patient's best interest. For the same reason any experimental design that asks a clinician to treat a patient against the clinician's own professional bias is inappropriate at best. And even if ethical reservations could be overcome, it would clearly be of only minimal scientific value to accumulate data on how patients fare under treatments not considered optimal at the time they are delivered.

Baumrind (1993) concludes:

> Except in special and very limited circumstances, clinical studies in orthodontics cannot and should not be expected to reveal categorically which of two or more treatments is better in a global sense. They can and should be expected to supply valid and reliable information about the mean effects of different treatments. But more important, they should supply information about the usual individual variability of human growth, development, and response to therapeutic intervention.

Retrospective studies have permitted clinical investigators to evaluate the palatal and facial growth and development responses within a particular cleft type according to the type and timing of the surgical procedures employed. Such studies have shown that the degree of palatal scarring is directly related to the areas of denuded bone resulting from the displacement of the palates mucoperiosteum during cleft closure.

Roentgencephalometry has aided in the elucidation of the nature of the craniofacial malformation associated with facial clefts as it affects the mandible, maxilla, orbits, nasopharyngeal area, and the base of the skull and cervical vertebrae. Moreover, current studies on the variable growth and involution of tonsils and adenoids have raised a number of questions of interest to immunologists.

2.4 The Need for Geometric and Quantitative Analysis of Cleft Palate Casts

Treatment planning in cleft lip and palate habilitation is contingent upon understanding the natural history of the palatal cleft defect and the face in which it exists. Longitudinal dental cast studies ultimately helped explain many cause and effect relationships which existed between palatal surgery and subsequent facial development. However, there still remains an important need for our understanding of palatal development, which will further refine and improve rehabilitative procedures. The purpose is to consider previ-

ously posed questions in light of newer biostereometric techniques. Specially, the following questions have been asked:

1. Are the palatal shelves intrinsically deficient, adequate, or excessive in mass?
2. To what extent does the geometric relationship of the palatal shelves in one cleft compare with that of another in the same type of cleft? With other types of clefts? With normal palates?
3. To what extent are the palatal shelves displaced in space?
4. How are these parameters altered as a consequence of growth and surgical reconstruction?

The advent of advanced biostereometric technique and 3D digital cameras with computer made it possible to analyze the size and shape of the palate in greater detail through intensive geometric survey. The data collected by these systems can be reduced to a mathematical format and subjected to analysis by high-speed computers.

In accordance with these objectives, we undertook a series of phased studies utilizing stereophotogrammetric electromechanical 3D digital analysis of serial casts of infants with cleft lip and palate with the following specific aims:

1. Test the reliability of the method for selecting the proper anatomical landmarks when extrapolating data from the stereophotographs.
2. Compare and contrast 2D and 3D surface area measurements with other descriptive measurements to determine if there are significant differences in their interpretive values.
3. Perform 3D analysis of serial casts in order to describe the changing geometry of the palatal vault in mathematical terms.
4. Determine whether the descriptive analysis revealed additional information relative to the geometric changes that follow in the course of time.
5. Determine by differential analysis if a constant geometrical relationship might exist between the size and shape of the lesser to greater palatal segment in a complete unilateral cleft lip and palate.
6. The best time to determine palatal surgery based on the size of the cleft relative to the size of the body palate.

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Facial and Palatal Growth

3

Samuel Berkowitz

3.1 Maxillary and Mandibular Growth Concepts

It is not the author's intent to write a definitive treatise on facial growth and its control processes because there are better sources for such information. However, because the history of cleft palate treatment has been influenced by what clinicians think is the correct facial growth process, it behooves the author to support or refute the various facial–palatal growth concepts based on his own clinical findings.

3.1.1 Newborn Palate with a Cleft of the Lip or Palate

Is bone missing, adequate, or in excess? What is the geometric palatal relation of the palatal segments at birth? With complete clefts of the lip and palate, are the palatal segments collapsed or expanded? Can the palatal segments be stimulated to develop to a larger size by neonatal orthopedic appliances? A number of studies have attempted to determine whether the cleft palate was deficient or adequate in osteogenic tissue; unfortunately, the investigators were limited by paucity of data, lack of homogeneity in their samples and the hazards of estimating growth from cross-sectional data.

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3.1.2 Genetic Control Theory: Craniofacial Growth Is Entirely Predetermined

Enlow (1975) writes that, in the past, it was thought that all bones having cartilage growth plates were regulated entirely and directly by the intrinsic genetic programming within the cartilage cells. Intramembranous bone (maxillary) growth, however, was believed to have a different source of control. This type of osteogenic process is particularly sensitive to biomechanical stresses and strains, and it responds to tensions and pressure by either bone deposition or resorption.

Tension, as traditionally believed, specifically induces bone formation. According to the traditional wisdom, when tension is placed on a bone, the bone grows locally in response. Pressure, on the other hand, if it exceeds a relatively sensitive threshold limit, specifically triggers resorption. According to this theory when muscle and overall body growth are complete, the bone attains

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biomechanical equilibrium; that is, the forces of the muscles are then in balance with the physical properties of the bone. This turns off osteoblastic activity, and skeletal growth ceases.

Unfortunately for traditional schools of thought, growth control in the human body is more complex than this. Moreover, it is now known that there is not a direct, one-to-one correlation between tension–deposition and pressure–resorption.

3.1.3 **Functional Matrix Theory** (Moss 1962, 1969) (Figs. 3.1 and 3.2)

Enlow (1975) goes on to explain that, with the development of the functional matrix principle, a number of important hypotheses began to receive attention. One of these is that the "bone" does not regulate its own growth. The genetic and epigenetic determinants of skeletal developments are in the functional tissue matrix, that is, muscle, nerve, glands, teeth, neurocranial fossa, and nasal, orbital, oral, and pharyngeal cavities. This is primary while the growth of the skeletal unit is secondary. However, although the functional matrix principle describes what happens during growth, it does not account for how it happens. Experiments have shown that mechanical forces are not the principal factor controlling bone growth.

Most researchers agree that a notable advance was made with the development of the functional matrix principle introduced by Moss (1962, 1969). It deals with what determines bone and cartilage growth in general. The concept states, in brief, that any given bone grows in response to functional relationships established by the sum of all the soft tissues operating in association with that bone. This means that the bone itself does not regulate the rate and direction of its own growth; the functional soft tissue matrix is the actual governing determinant of the skeletal growth process.

The course and extent of bone growth are secondarily dependent on the growth of pace-making soft tissues. Of course, the bone and any cartilage present are also involved in the opera-

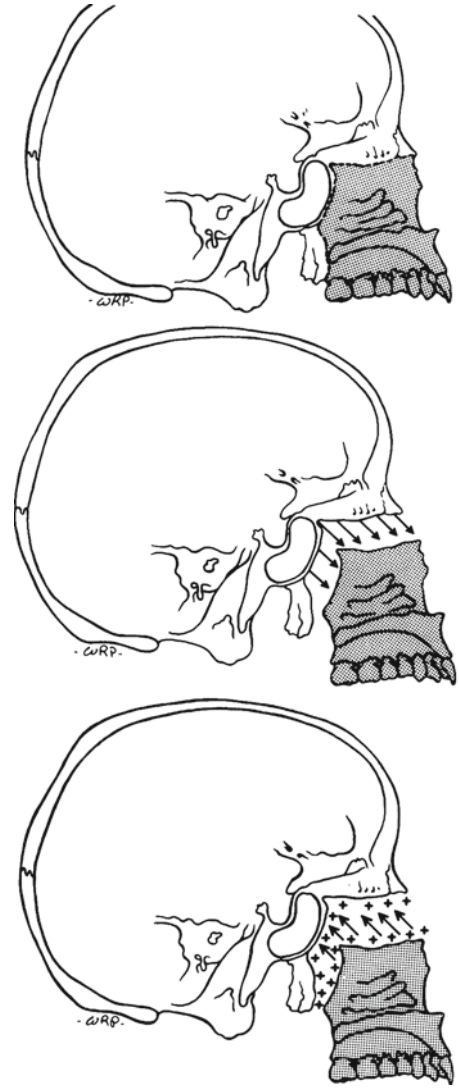

Fig. 3.1 The process of new bone deposition does not cause displacement by pushing against the articular contact surface of another bone. Rather, the bone is carried away by the expansive force of all the growing soft tissues surrounding it. As this takes place, new bone is added immediately onto the contact surface, and the two separate bones thereby remain in constant articular junction. The nasomaxillary complex, for example, is in contact with floor of the cranium (*top*). The whole maxillary region is displaced downward and forward away from the cranium by the expansive growth of the soft tissues in the midfacial region (*center*). This then triggers new bone growth at the various sutural contact surfaces between the nasomaxillary composite and the cranial floor (*bottom*). Displacement thus proceeds downward and forward as growth by bone deposition simultaneously takes place in an opposite upward and backward direction (i.e., toward its contact with the cranial floor) (From Enlow (1975))

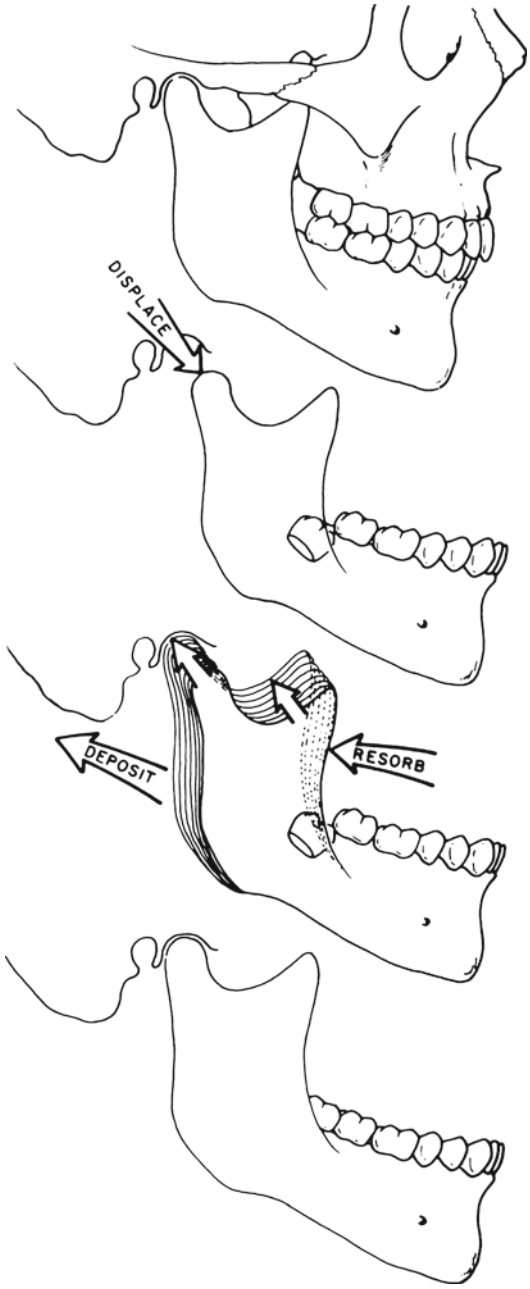

Fig. 3.2 Similarly, the whole mandible is displaced "away" from its articulation in each glenoid fossa by the growth enlargement of the composite of soft tissues in the growing face. As this occurs, the condyle and ramus grow upward and backward into the "space" created by the displacement process. Note that the ramus "remodels" as it relocates posteriorly. It also becomes longer and wider to accommodate (1) the increasing mass of masticatory muscles inserted onto it, (2) the enlarged breadth of the pharyngeal space, and (3) the vertical lengthening of the nasomaxillary part of the growing face (Reprinted with permission from Enlow (1975))

tion of the functional matrix, because they give essential feedback information to the soft tissues. This causes the soft tissues to inhibit or accelerate the rate and amount of subsequent bone growth, depending on the status of the functional and mechanical equilibrium between the bone and its soft tissue matrix. The genetic determinants of the growth process reside wholly in the soft tissues and not in the hard part of the bone itself.

The functional matrix concept is fundamental to an understanding of the overall process of bone growth control. This concept has had a great impact in the field of facial biology. The concept also comes into play as a source for the mechanical force that carries out the process of displacement. According to this now widely accepted explanation, the facial bones grow in a subordinate relationship with all the surrounding soft tissues. As the tissues continue to grow, the bones are passively (i.e., not of their own doing) carried along (displaced) with the soft tissues attached to the bones by Sharpey's fibers. Thus, for the nasomaxillary complex, the expansion of the facial muscle, the subcutaneous and submucosal connective tissues, the oral and nasal epithelia lining the spaces, the vessels, and the nerves all combine to move the facial bones passively along with them as they grow. This continuously places each bone and all of its parts in correct anatomic positions to carry out its functions. Indeed, the functional factors are the very agents that cause the bone to develop into its definite shape and size and to occupy the location it does.

Growth control is determined by genetic influences and biomechanical forces, but the nature of the balance between them is still, at best, uncertain. No single agent is directly responsible for the master control of growth; the control process encompasses many factors. It involves a chain of regulatory links. Moreover, not all of the individual links are involved in all types of growth changes.

Enlow (1975) identifies the maxillary tuberosity as being a major site of maxillary growth. It does not, however, provide for the growth of the whole maxilla, but rather is responsible for the lengthening of the maxillary arches. The whole

maxilla is displaced in an anterior direction as it grows and lengthens posteriorly. However, the nature of the force that produces this forward movement is a subject of great controversy. The idea that additions of new bone on the posterior surface of the elongating maxillary tuberosity "push" the maxilla against the adjacent pterygoid plates has been abandoned.

Bones do not by themselves have the physiological capacity to push away bones. Another theory held that bone growth at the various maxillary sutures produces a pushing apart of the bones, with a resulting thrust of the whole maxilla downward and forward. This theory has also been rejected because bone tissue is not capable of growth in a field that requires the amount of compression needed to produce a pushing type of displacement. The sutural connective tissue is not adapted to a pressure-related growth process. It is believed that the stimulus for sutural bone growth is the tension produced by the displacement of the bone. Thus, the deposition of new bone is a response to displacement rather than the force that causes it. Although the "sutural push theory" is not tenable, Enlow reports that some students of the facial growth control processes are looking anew at growth mechanizing sutures, but not in the old conceptual way.

3.1.4 Cartilage-Directed Growth: Nasal Septum Theory (Scott 1953, 1954, 1955, 1956a, b, 1957, 1958a, b, 1959)

Cartilages are the leading factor. Synchondrosis, nasal septum, and mandibular condyles are actual growth centers. Sutural growth is compensatory. This theory developed from criticisms of the "sutural theory." Scott (1953, 1954) believes that cartilage is specifically adapted to certain pressure-related growth sites because it is a special tissue uniquely structured to provide the capacity for growth as a result of compression. The basis for this theory is that the pressure-accommodating expansion of the cartilage in the nasal septum is the source of the physical force that displaces the maxilla anteriorly and inferiorly. This, according to Scott's hypothesis, sets up fields of tension in all the maxillary sutures. The bones then, while they enlarge at their sutures in response to the tension created by the displacement process, move in relation to each other.

The nasal septum hypothesis was soon adopted by many investigators in cleft palate centers around the world and became more or less the standard explanation, replacing the "sutural theory." Clinicians involved in cleft palate treatment, such as McNeil (1950, 1954, 1964) and Burston (1960) and their followers (Crikelair et al. 1962; Cronin and Penoff 1971; Derichsweiler 1958; Dreyer 1962; Georgiade 1970; Georgiade and Latham 1975a, b, Graf-Pinthus and Bettex 1974; Hellquist 1971; Huddart 1979; Kernahan and Rosenstein 1990; Krischer et al. 1975; Latham 1968; Robertson 1971; Monroe and Rosenstein 1971), accepted Scott's thesis that cartilage and periosteum carry an intrinsic genetic message that guides their growth. They believed that the cartilaginous centers, such as the chondrocranium, the associated synchondroses, and the nasal septum, should be viewed as the true centers of skull and facial growth. Scott (1953, 1954) further suggests that the nasal septum plays more than a secondary role in the downward and forward vector of facial growth.

McNeil (1950, 1954), following Scott's thesis, describing the embryopathogenesis of complete clefts of the lip and palate and their treatment at the neonatal period, wrote that the palatal processes, being detached from the growing nasal septum, do not receive their growth impetus and, therefore, are not only retruded within the cranium but are also deficient in osteogenic tissue. He goes still further and believes that the deficient palatal processes can be stimulated to increased size through the use of functional orthopedics.

3.1.4.1 Stimulation of Bone Growth: Is It Possible?

As McNeil saw it, pressure forces created by "functional" orthopedic appliances, which are within the limits of tolerance, will act to stimulate bone growth in an anterior direction. This force needs to be applied to particular regions and in particular directions so that it can intensify

normal forces. The resulting narrowing of the cleft is due to growth of the underlying bone brought on by such stimulating appliances. Additional growth leads to a reduction in the soft palate cleft as well, thereby increasing the chance of having a long, flexible, well-functioning soft palate after surgical closure.

McNeil (1954) goes on to suggest that an obturator alone is unsatisfactory because it will reduce "valuable" tongue space and lead to harmful speech habits. McNeil was correct in stressing that surgery should be reduced to a minimum compatible with sound clinical reasoning and accepted surgical principles.

Whereas McNeil states that his procedure stimulates palatal growth, thereby narrowing the cleft space, Berkowitz's (1989) 3D palatal growth studies – using a sample of cases that have not had neonatal maxillary orthopedic treatment and a control sample of noncleft cases – show that growth occurs spontaneously. This is an expression of the palate's inherent growth potential, which can vary among patients. Berkowitz concluded that "catch-up growth" can occur after palatal surgery (with minimum scarring) is performed.

3.1.4.2 The Need to Prevent Collapse

McNeil (1950, 1954, 1964) further believes that the palatal segments should be manipulated to an ideal relationship prior to lip surgery to prevent them from moving too far medially and becoming collapsed with the buccal segments in crossbite. This, he suspects, will lead to abnormal movements of the tongue and give rise to faulty respiratory, sucking, and swallowing patterns, also causing abnormal growth and development of the palatal structures.

Mestre et al. (1960), studying palatal size in a cleft population that had not been operated on, report that the development of the maxilla appears to be normal in unoperated cases. They do conclude that it is the type, quality, and extent of the surgery that determine the effect on maxillary growth and that osteogenic deficiency does exist to varying degrees. Our research on serial palatal growth changes supports this conclusion that palates with clefts are highly variable in size, shape, and osteogenic deficiency.

Unfortunately, McNeil's interpretation of the effects of clefting on the various vegetative functions, and in reducing palatal growth, has not been supported by controlled objective research. The inability of the manipulated arch to remain intact after lip surgery, and not move medially into a collapsed relationship, has led many clinicians to question the accuracy of McNeil's other stated benefits such as reduction of middle ear infections.

McNeil (1950, 1954, 1964) made other faulty observations. Among them:

1. He mistakenly believed that the orthopedic appliance will stimulate the underdeveloped cleft segment in unilateral clefts of the lip and palate (UCLP) to move forward, to make contact with the premaxillary portion of the greater segment and both palatal segments in bilateral clefts of the lip and palate (BCLP), after the lip is united. Even as early as the 1960s, many orthodontists found the opposite to be true. In UCLP, the premaxillary portion of the larger segment moves medially and backward to make contact with the lesser segment due to the action of compressive lip muscle forces. If McNeil had had the benefit of serial casts, his interpretation of clinical events would, I am confident, have been totally different.
2. McNeil's claim that the lesser segments in UCLP, and both segments in BCLP, can be stimulated to grow forward is totally erroneous. His conclusions were based on conjecture, not on objective data. The results of Berkowitz's 3D palatal growth studies (Wolfe and Berkowitz 1983) show marked acceleration in palatal growth during the first 2 years without orthopedic treatment, with most of the growth changes occurring at the area of the maxillary tuberosity and not at the anterior portion of the palate except for alveolar growth associated with canine development (Fig. 3.2). Movement of the cleft palatal segment anteriorly is only possible as a result of reactive mechanical forces being applied through the use of pinned maxillary orthopedic appliances or from a protraction facial mask.

One last but significant characterization of a newborn cleft of the lip and palate needs to be refuted. McNeil states that "in BCLP lateral segments are collapsed toward the midline before birth." However, he does not explain the dynamics that can make this possible. How can segments be collapsed if there are no inwardly directed forces from the cleft lip–cheek muscle complex, especially when the tongue fits within the cleft space and acts to move the palatal segments apart?

Enlow's (1975) report on current thinking on palatal growth processes delivers McNeil's thesis a mortal blow. Enlow (1975) writes that recent research has shown that pressure is detrimental to bone growth.

Bone is necessarily both a traction and pressure-adapted kind of tissue. The periosteal membranes are constructed to function in a field of tension (as by the pull of a muscle). Covering membranes are quite sensitive to direct compression because any undue amount causes vascular occlusion and interference with osteoblastic formation of new bone. Osteoclasts, conversely, function to "relieve" the degree of pressure by removing bone. Bone is pressure sensitive, and high-level pressure induces resorption.

Moss et al. (1968), responding to the role of nasal septal cartilage in midfacial growth as put forth by Scott (1953, 1959), states that Scott's hypothesis is based on the following assumptions: (1) that in the fetal skull, the original nasal capsule and its derivatives are cartilaginous; (2) that all cranial cartilaginous tissues (septal, condylar, or in synchondroses) are primary growth centers, by virtue of the undoubted ability of all cartilaginous tissues to undergo interstitial expansive growth; and (3) that following the prenatal appearance of the intramembranous vomer (and of the several endochondral ossification centers of the ethmoid sinuses and the turbinates), the remaining unossified portions of the cartilaginous nasal capsule continue to be capable of such interstitial expansion. Moss further suggests that the nasal septal cartilage grows as a secondary, compensatory response to the primary growth of related orofacial matrices and that midfacial skeletal growth is not dependent on any prior, or primary, growth "impetus" of the nasal septal cartilages.

In Scott's hypothesis, it is assumed that cartilaginous interstitial growth is the major source of the expansive force that "pushes" on the subjacent midfacial skeletal structures, causing both vertical and anteroposterior growth. Moss believes that it has been demonstrated repeatedly that growth in size and shape, as well as the changes in spatial position, of all skeletal units is always secondary to primary changes in their functional matrices. This secondary skeletal unit growth comes about in the following manner. All cranial bones and cartilages originate and grow within soft tissue capsules. The splanchnocranial skeleton exists within an orofacial capsule. The primary growth of the enclosed orofacial matrices causes the orofacial capsule to expand responsively. Because the splanchnocranial bones are within this capsule, they are passively translated in space within their expanding capsule. As a result of such spatial displacement, the individual bones will be distracted (or separated) passively from one another.

The increments of growth observed at the sutural edges of these bones, and at the mandibular condylar cartilages, are secondary, compensatory, and mechanically obligatory responses of the skeletal units to such separative movements (i.e., the alterations of size and shape in bones and cartilages are responses to matrix growth, not the cause of it).

The nasal skeleton is characterized by a relatively great normal variation in form. The nasal capsule (and septum), from its inception, serves to protect and support the functional spaces for respiration and olfaction. In human, the olfactory spaces are fully formed at birth. Postnatal cavity growth exclusively increases the respiratory functioning space.

The growth of the upper face is, in part, a response to the functional demands for increased respiratory volume. The nasal cavity is not a space haphazardly left over after the upper facial structures complete their growth. On the contrary,

the expansion of the nasal cavity is the primary morphogenetic event, and nasal capsular growth, both osseous and cartilaginous, is secondary. The application of the theory of functional cranial analysis to nasal and midfacial skeletal growth demonstrates that the growth of each of these two areas is independent of the other and that the nasal septal cartilage plays a secondary compensatory role, rather than a primary morphogenetic one.

At present, the nasal septum theory is somewhat accepted as a reasonable explanation by a number of clinicians who favor presurgical orthopedic treatment, although it is universally realized that much more needs to be understood about facial growth processes (Moss 1968). (The use of presurgical orthopedic treatment is covered in greater detail in Chaps. 10 and 11.)

Clinically, there seems to be more support for the functional matrix theory than the nasal septum theory. Unfortunately, McNeil, in espousing Scott's theory to explain the "retropositioned maxillary complex relative to the mandible and osteogenically deficient palatal processes" in complete clefts of the lip and palate, did not have access to serial palatal and facial growth records to support such a view. However, Berkowitz's (1985) serial casts study of CUCLP and CBCLP cases using the Angle's occlusal classification system, which is the most reliable means of judging the geometric relationship of the maxillary to the mandibular arches within the face, showed that at 3–6 years of age, the teeth in the lateral palatal segments were in either a class I or class II relationship but were never in a class III relationship.

On this basis, one can conclude that it is not the lack of a growth impetus from the nasal septum that explains the presence of a small cleft palatal segment at birth. If palatal osteogenic deficiency does exist, it can more accurately be explained in relationship to the embryopathogenesis of facial development: the failure of migrating undifferentiated mesenchymal cells from the neural crest to reach the facial processes (Millard 1980; Ross and Johnston 1972).

3.1.5 Basion Horizontal Concept: The Direction of Facial Growth (Figs. 3.3, 3.4, and 3.5) (Coben 1986)

No discussion on craniofacial growth is complete without including Coben's basion horizontal concept of the direction of facial growth. Basion horizontal is a concept based on a plane at the level of the anterior border of foramen magnum parallel to Frankfort horizontal where basion is the point of reference for the analysis of craniofa-

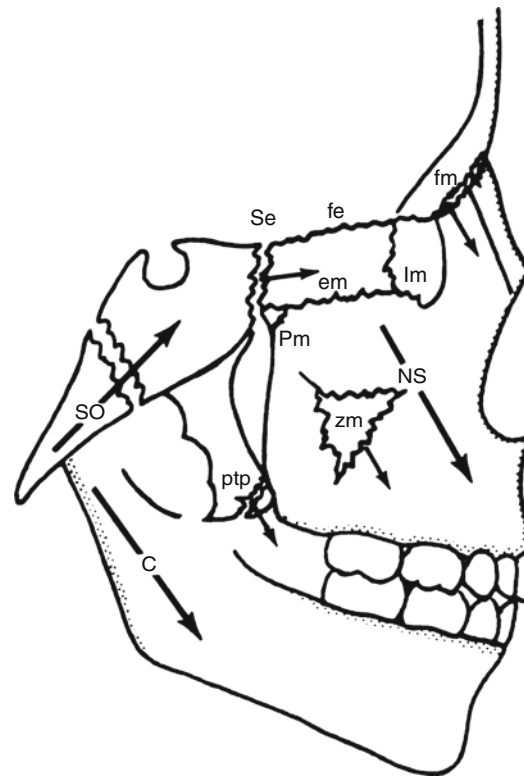

Fig. 3.3 Postnatal craniofacial growth systems to the age of 7 years (first decade). Cartilaginous growth: *SO* sphenooccipital synchondrosis, *C* reflection of condylar mandibular growth, *NS* nasal septum. Sphenoethmoidal circumaxillary suture system: *se* sphenoethmoidal, *ptp* pterygopalatine, *pm* palatomaxillary, *fe* frontoethmoidal, *em* ethmoidal–maxillary, *lm* lachrymal–maxillary, *fm* frontomaxillary, *zm* zygomaticomaxillary, *zt* zygomaticotemporal (not shown). Surface apposition-modeling resorption development (stippled area): minor contribution (Reprinted from Coben (1986))

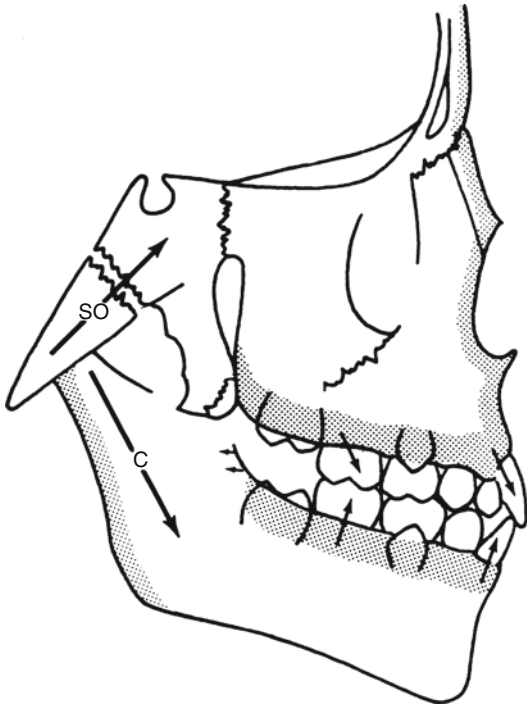

Fig. 3.4 Postnatal craniofacial growth systems from age 7 years (second decade). Cartilaginous growth: *SO* spheno-occipital synchondrosis-active through puberty, *C* reflection of condylar mandibular growth – active to facial maturity, nasal septum – growth completed. Sphenoethmoidal circumaxillary suture system: sutural growth no longer primary system of upper facial development. Surface apposition-modeling resorption development (stippled area): now major method of upper facial development and alveolar growth (Coben 1986)

cial growth. Coben states that the growth concept which basion horizontal represents is that craniofacial growth is reflected away from the foramen magnum (basion) and the vertebral column. The cranio-maxillary complex housing the maxillary dentition is translated upward and forward from basion by growth of the cranial base. Growth of the mandible is reflected away from basion, carrying the mandibular dentition downward and forward. The divergence of the two general vectors develops space for vertical facial growth and the eruption of the dentition.

Normal maxillomandibular development requires synchronization of the amount, timing,

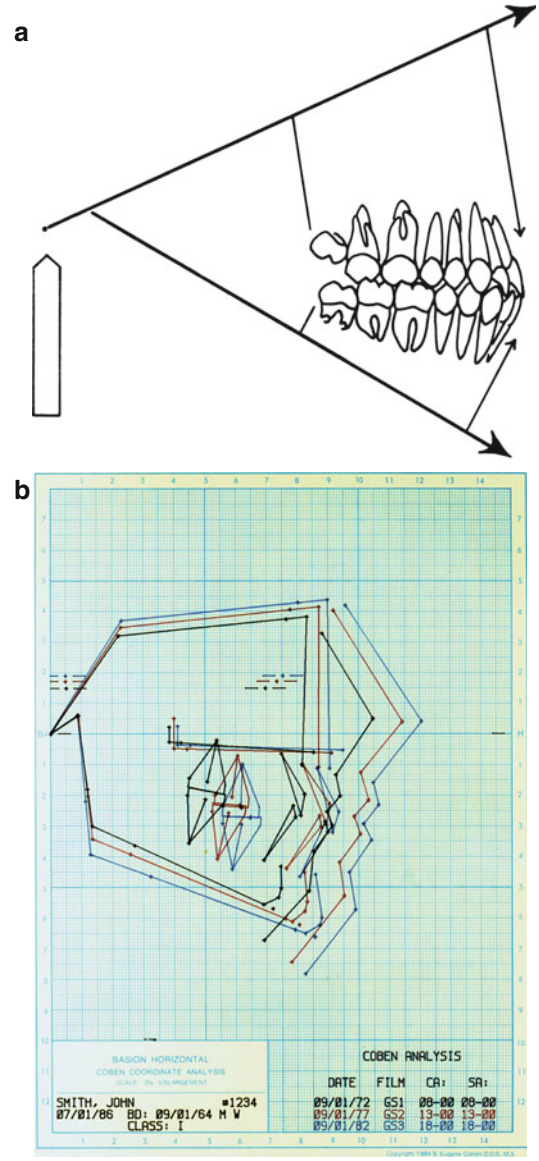

Fig. 3.5 (**a**) Basion horizontal. General vectors of craniofacial growth. Growth of the cranial base translates the upper face and the maxillary dentition upward and forward away from the foramen magnum. Growth of the mandible translates the lower dentition downward and forward. The two diverging vectors create space for vertical facial development and tooth eruption (Coben 1986) (**b**) Basion horizontal. Basion horizontal coordinate computer craniofacial serial schematic line graph of Fig. 3.5a

and direction of growth of the cranio-maxillary complex and of the mandible. The cranial base vector represents the upward and forward translation of the upper face by growth of the sphenooccipital synchondrosis, while growth of the sphenoethmoidal/circumaxillary suture system and the nasal septum increases the depth and height of the upper face.

The basion–articulare dimension is essentially stable postnatally, indicating that the mandible maintains a constant sagittal spatial relation to the foramen magnum as the reflection of mandibular growth carries the lower teeth downward and forward, away from the cranial base.

There are two distinct phases of craniofacial growth because of a change in the system of upper facial development after the approximate age of 7 years. Before age 7, growth of the upper face is dominated by the nasal septum, the eyeballs, and the sphenoethmoidal/circumaxillary suture system (Fig. 3.4). At this age, the growth in this suture system produces space for the eruption of the maxillary first molars. Longitudinal cephalometric findings of a continuous increase in the sella–frontale dimension with little increase in the thickness of the frontal bone before age 7 support the concept that bone apposition and remodeling resorption are minor factors in these early years.

At about age 7, the growth system of the upper face changes with the closure of the sphenoethmoidal suture. The sella–frontale dimension stabilizes, and the thickness of the frontal bone begins to increase by surface apposition and remodeling until maturity. The interpretation is that after age 7, the initial primary system of sphenoethmoidal/circumaxillary sutural growth of the upper face is replaced by surface apposition and remodeling resorption (Fig. 3.4). It is significant that, before age 7, space for the erupting upper first molars results from growth of the sphenoethmoidal/circumaxillary suture system. After age 7, space for the upper second and third molars is produced by maxillary alveolar apposition as the maxillary dentition erupts downward and forward. This concept was supported by Scott (1959), who reasoned that the sphenoethmoidal suture must be viewed as part of the major circumaxillary suture system and that once part of the suture closes, there is no further growth in that suture system. Longitudinal cephalometric growth studies confirm this interpretation (Fig. 3.5).

3.2 Mandibular Development in Cleft Palate (Figs. 3.6 and 3.7)

Recent studies have revealed a series of often subtle differences in the morphology of the mandible in persons with cleft lip and/or palate. Dahl (1970) and Chierici and associates (Chierici et al. 1973) found that, in persons with clefts of the hard palate only, the mandibular plane was steeper and the gonial angle more obtuse than in a normal population. Mazaheri and coauthors (1971) noted that the length and width of the mandible were significantly less in persons with cleft palate only than in those with cleft lip and palate (CLP) and normal groups. Aduss (1971) observed that the mandibular gonial angle in patients with unilateral CLP was more obtuse and that the anterior cranial base appeared to be elevated. Rosenstein (1975) also found the mandibles to be smaller, with steeper mandibular plane angles. Bishara (1973) studied Danish children with repaired cleft palates only. In that study, and again in a later study of patients with CUCLP (Bishara et al. 1979), he noted that the mandible was significantly more posterior in relation to the cranial base and that its mandibular plane was steeper than normal.

Krogman and colleagues (1975) found no difference in mandibular dimensions in the BCLP population, other than a more obtuse gonial angle. They also found the temporomandibular joint to be positioned farther back so that its effective length was less than in the normal population. Robertson and Fish (Robertson and Fish 1975), comparing mandibular arch dimensions, found no significant differences between normal and cleft children either at birth or at 3 years of age.

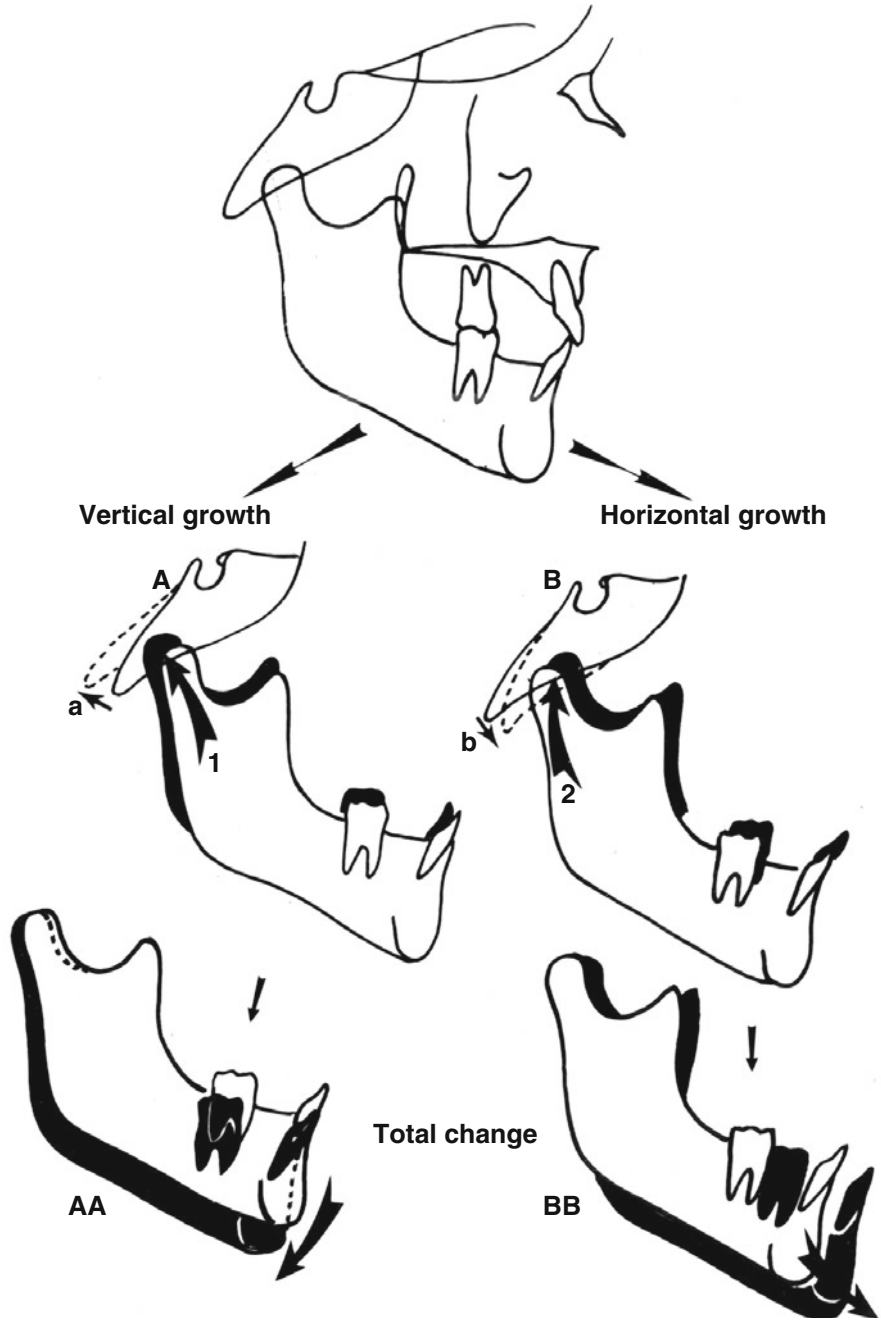

Fig. 3.6 Various growth changes that occur in the condylar head determine the direction and extent of mandibular growth

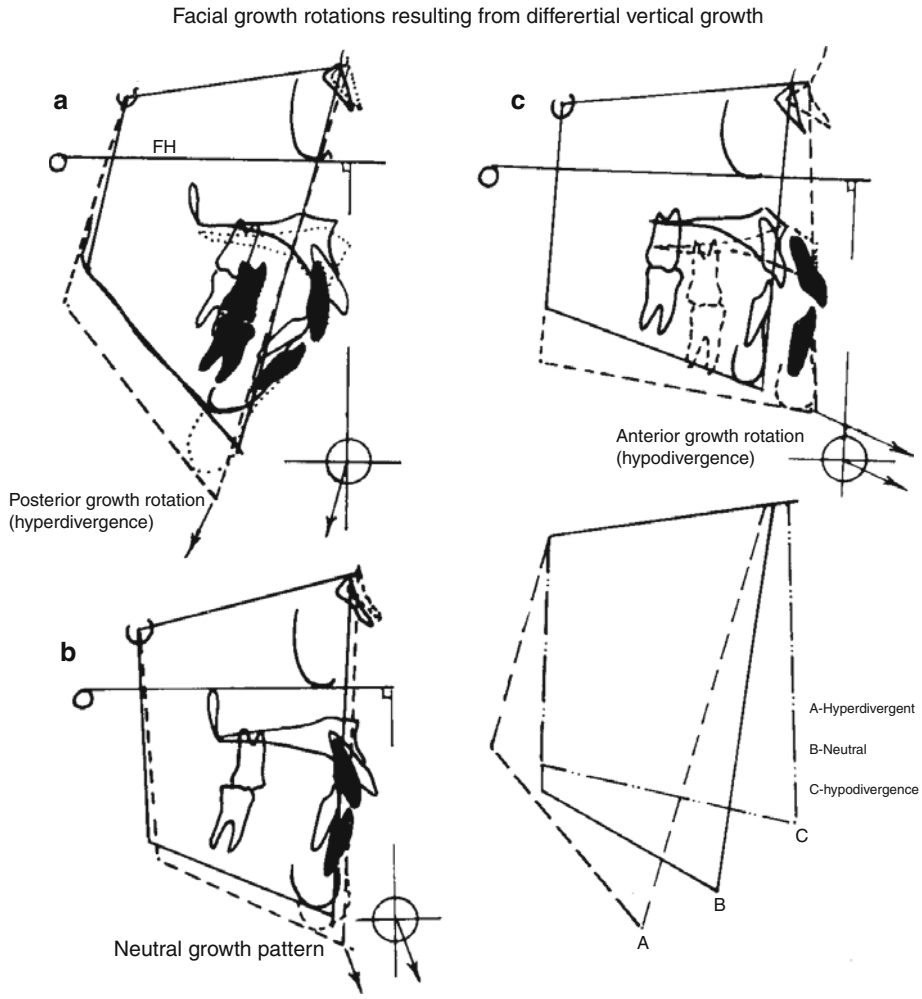

Fig. 3.7 (**a**–**c**) Facial growth rotations resulting from differential vertical growth. (**a**) Hyperdivergent pattern with posterior growth rotation. (**b**) Neutral growth pattern. (**c**) Hypodivergent growth pattern with anterior growth rotation. Comment: This series is not a true reflection of the growth of various components of the face. See Coben's basion horizontal, coordinate craniofacial analysis system for this (Fig. 3.5)

3.3 Patterns of Postnatal Growth

Based on the serial studies, three general patterns of postnatal growth have been demonstrated. In the Pierre Robin sequence, and in complete bilateral clefts of the lip and palate, most cases demonstrate substantial improvement through "catch-up" in the growth of the mandible. In the second pattern, mandibulofacial dysostosis, the pattern of growth is such that the deformity observed in infancy or early childhood is maintained throughout the growth period. The deformity of the mandible neither improves nor worsens in the course of time. The third pattern is one in which the growth process is so deranged that the severity of the deformity increases with age. This has been observed in some instances of unilateral agenesis of the mandibular ramus (e.g., hemifacial microsomia) and in the growth of the maxilla and neurocranium in some forms of premature craniofacial synostosis.

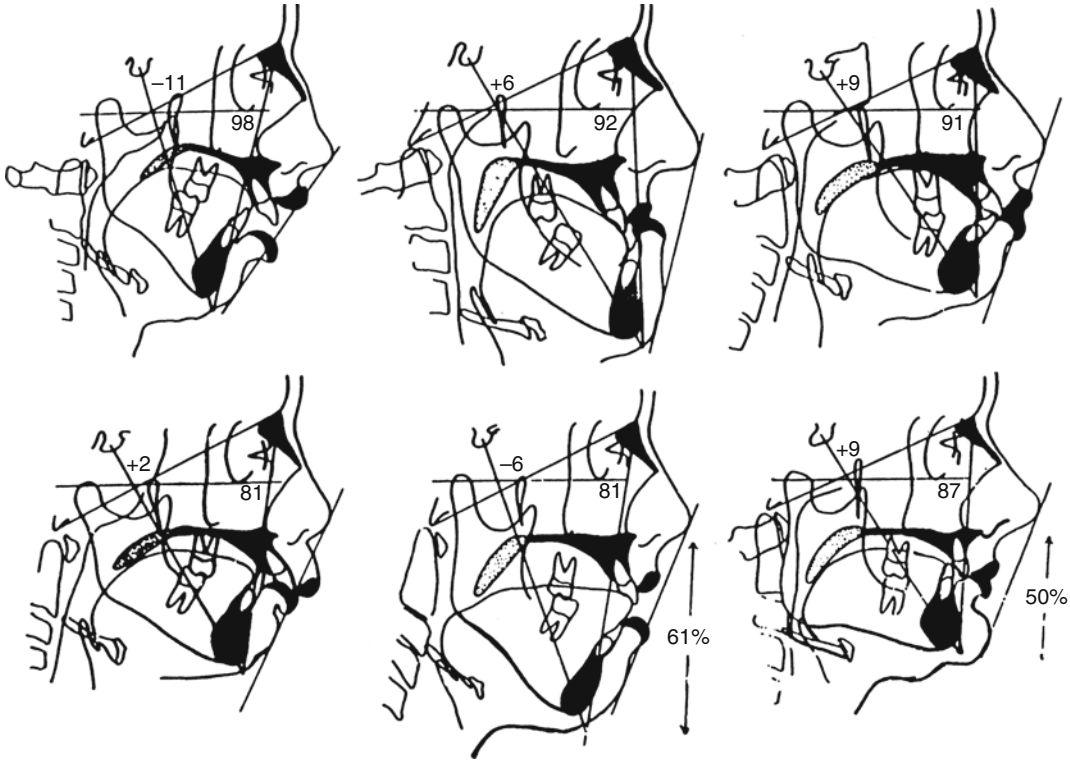

Fig. 3.8 Variations in facial growth patterns. Courtesy of Ricketts (1956)

3.3.1 Bone Remodeling During Growth (Fig. 3.8)

Enlow (1975) states that remodeling is a basic part of the growth process. The reason why a bone must remodel during growth is because its regional parts become moved; "drift" moves each part from one location to another as the whole bone enlarges. This calls for sequential remodeling changes in the shape and size of each region. The ramus, for example, moves progressively posteriorly by a combination of deposition and resorption. As it does so, the anterior part of the ramus becomes remodeled into a new addition for the mandibular corpus. This produces a growth elongation of the corpus. This progressive, sequential movement of component parts as a bone enlarges is termed relocation. Relocation is the basis for remodeling. The whole ramus is thus relocated posteriorly, and the posterior part of the lengthening corpus becomes relocated into the area previously occupied by the ramus. Structural remodeling from what used to be part of the ramus into what then becomes a new part of the corpus takes place. The corpus grows longer as a result.

3.3.2 Maxillary Growth

The maxilla grows downward and forward from the cranial base with growth occurring at the articulations with other bones (i.e., the sutures). Björk (1975) stated that during growth the maxilla is displaced in a rotational manner relative to the cranial base; however, this rotational aspect is small, which results in the downward and forward effect. Furthermore, he emphasized that there is little variation in the upper facial height between groups. Therefore, because of the small variation, it is likely that individual difference in facial form results from growth in other facial areas where there is more variation.

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Part II

Types of Clefts

The Effect of Clefting of the Lip and Palate and the Palatal Arch Form

4

Samuel Berkowitz

4.1 Varieties of Cleft Lip and Cleft Palate

4.1.1 Action of Intact Facial Muscular Forces on the Maxillary Arch

In a normal jaw with intact lips and palate, the muscles of the lip, cheek, and pharynx exert their normal sphincter-like actions against the developing maxillary and mandibular arches. The compressive external muscular forces neutralize the expansion forces of the tongue (Fig. 4.1). The neonatal arch form changes as these forces change with growth and maturation, yet the opposing muscles always maintain a precise and dynamic balance with each other. When this muscular balance is upset, the arch form and teeth relationships change.

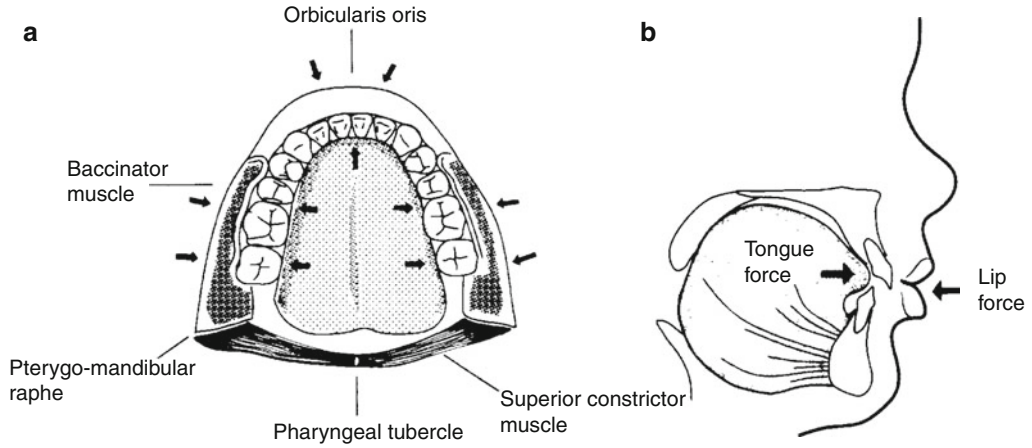

Fig. 4.1 The balance between facial and tongue muscle forces. (**a**) The outer muscles, the orbicularis oris, buccinator, and superior constrictor muscles form a ring which acts to compress the palatal and mandibular arches with the teeth. (**b**) The tongue force acts to expand the dental arches and teeth. Whether the teeth and arches align is determined by the resultant of these forces

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4.2 Aberrant Muscle Forces in Clefts of the Lip and Palate

A cleft of the lip and palate is the result of the failure of lip elements, and right and left palatal segments, to come together within the first 9 weeks of fetal life. The loss of muscular continuity of the orbicularis oris-buccinator-superior constrictor ring in complete unilateral and bilateral clefts changes the normal muscular force diagram. The aberrant muscular forces act to displace tissue masses. In complete clefts of the lip and palate, if the lateral palatal cleft segments are detached from the vomer, they will be pulled laterally by the external aberrant lip-cheek muscular forces, as well as spread apart by the tongue pushing into the cleft space (Fig. 4.2). Because clefts differ in their location and extent, lip and palate clefts can vary in the degree of geometric distortion, as well as in the size and shape of the cleft palatal segments.

The muscular forces that act on the bony scaffolding of the palate and pharynx begin very early in intrauterine life; therefore, the palatal and facial configuration at birth has been formed over the major portion of the infant's existence prior to birth.

In complete unilateral clefts of the lip and palate (CUCLP), the premaxillary portion of the noncleft segment is pulled anterolaterally. In addition to the lateral displacement of the lateral palatal segments, the premaxilla in the larger segment is carried forward in the facial skeleton. In complete bilateral cleft lip and palate (CBCLP), excessive growth in the premaxillary-vomerine suture is caused by increased tension at this site, precipitated by mechanical force stresses during periods of rapid growth (Berkowitz 1959; Pruzansky 1953, 1971; Friede 1973, 1977) (Fig. 4.3). This growth is continuous during early postnatal years and provides a fourth dimension

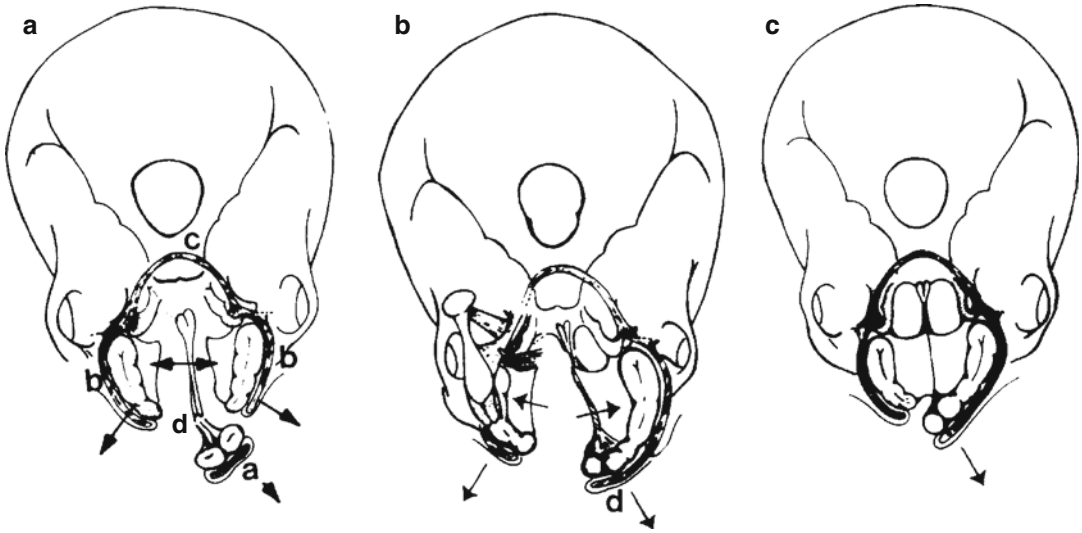
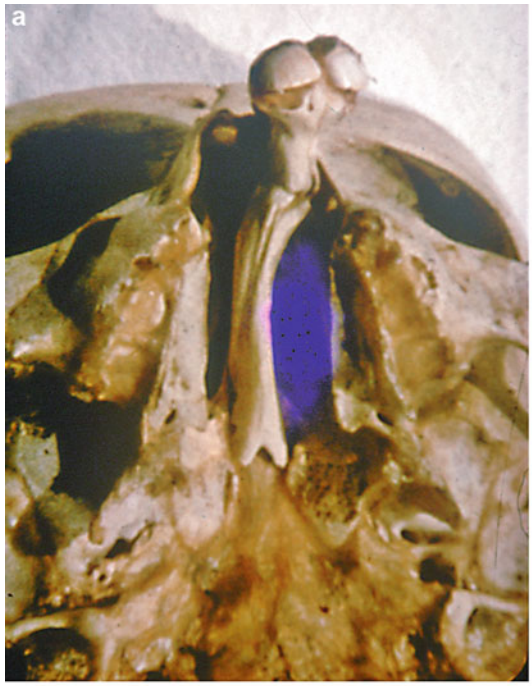
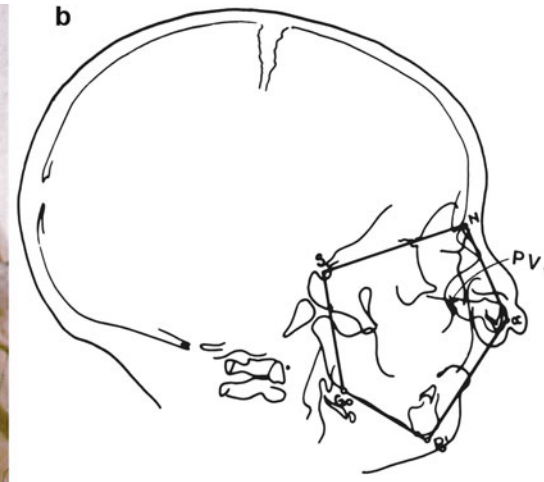


Fig. 4.2 (**a**–**d**) Effects of complete clefts of the lip and palate at birth. In complete cleft lip and palate, with a separation in the orbicularis oris (*a*) buccinator (*b*) superior constrictor (*c*) muscle ring, the aberrant muscle forces plus the plunger action of the tongue causes the palatal segments to be pulled and pushed apart. (**d**) premaxillary vomerine suture. (**a**) In bilateral clefts of the lip and palate, the premaxilla may be laterally or ventrally flexed with the fulcrum at the premaxillary-vomerine suture. (**b**) Complete unilateral clefts of the lip and palate at birth. The cleft's lesser segment and the premaxillary portion of the larger segment (*d*) are pulled outward. (**c**) Cleft of the lip and alveolus. The bony distortion is determined by the extent of alveolar involvement (Courtesy of J. D. Subtelny)





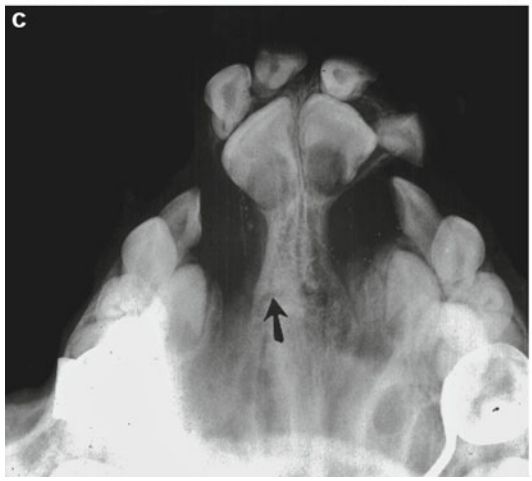


Fig. 4.3 (**a**–**c**) Skull with a bilateral cleft and palate. (**a**) The palatal segments have been overexpanded and the premaxilla protruded by the resultant aberrant muscle forces. The premaxillary-vomerine suture separates the premaxilla and the vomer and is a growth site. (**b**) Lateral cephalometric tracing at birth. *Lines* connecting various landmarks create a polygon depicting their geometric relationships. This tracing of a CBCLP at birth shows the degree of premaxillary protrusion. *S* Sella turcica, *N* Nasion, *a* Alpha (the anterior extent of the premaxilla), *Po* Pogonion, *Gn* Gnathion, *Go* Gonion, *PVS* Premaxillary-vomerine suture. (**c**) Occlusal radiograph of the premaxilla and vomer showing the premaxillary-vomerine suture (*arrow*)

to the deformity, which can alter the cleft palatal segments and their associated parts and either simplify or complicate treatment.

If a soft tissue (mucous membrane, skin, and fibrous connective tissue), which collectively forms Simonart's band, bridges the alveolar cleft, the attached palatal segments are limited in their degree of geometric displacement.

4.3 Categories of Clefts

Depending on the elemental characteristics of the embryology, anatomy, and physiology of the cleft defect, the varieties of clefts of the lip and palate may be tabulated into four general categories: (1) those involving the lip and alveolus, (2) those involving the lip and palate, (3) those in which the palate alone is affected, and (4) congenital insufficiency of the palate. The term "palate" will include both the hard palate and the velum, or soft palate (Fig. 4.4).

4.3.1 Clefts of the Lip (Figs. 4.5 and 4.6)

A cleft of the lip may be complete, extending from the vermilion border to the floor of the nose, or it may be incomplete. There are various degrees of incomplete lip clefts. Minimal defects involving only the vermilion border are observed. In others, the defect may extend to the nose as a submucous cleft in the muscle band, bridged only by mucous membrane, skin, and fibrous connective tissue. The nasal alar cartilage on the side of the cleft is displaced and flattened to a greater or lesser degree, depending on the extent and width of the cleft. The tip of the nose is deviated toward the noncleft side.

The cleft in the lip may be unilateral or bilateral, occurring on one or both sides, respectively. If bilateral, it may be symmetrical or asymmetrical, that is, it may or may not involve the lip equally on both sides (Fig. 4.5b). It should be noted that, in bilateral clefts, a median portion of the lip is isolated in the midline and remains attached to the

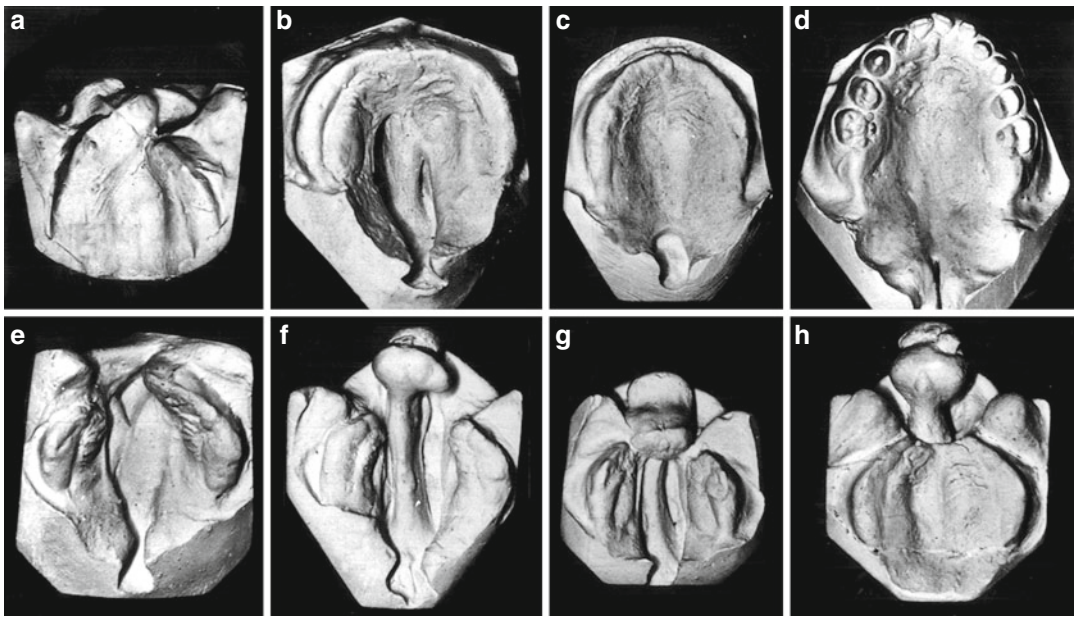


Fig. 4.4 (**a**–**h**) The anatomic classification system is based on the location, completeness, and extent of the cleft deformity. Because the lip, alveolus, and hard palate develop from different embryonic sources, any combination of clefting can exist. (**a**) Cleft of the lip and alveolus. Normal palate. (**b**) Isolated cleft of the hard and soft palate. Normal lip and alveolus. (**c**) Cleft of the soft palate and uvulae. (**d**) Cleft of the uvulae. (**e**) Complete unilateral cleft lip and palate. (**f**) Complete bilateral cleft of the lip and palate. (**g**) Incomplete bilateral cleft of the lip and palate. (**h**) Complete bilateral cleft of the lip and alveolus (Courtesy of Wolfe and Berkowitz (1989))

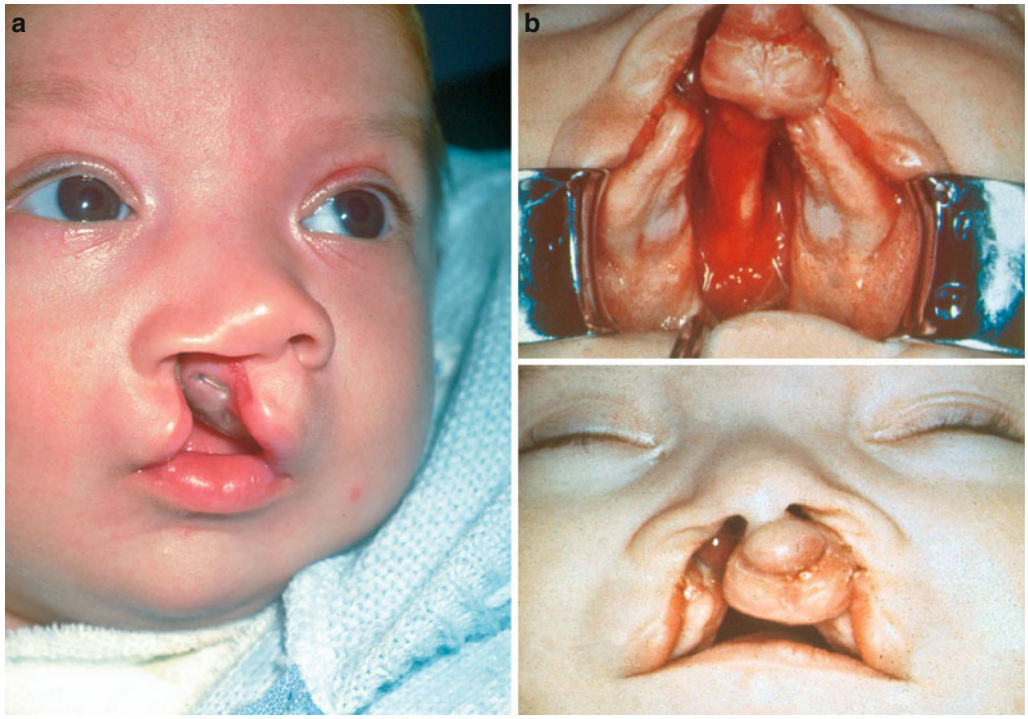


Fig. 4.5 (**a**) Complete unilateral cleft lip and palate. The distorted nostril is caused by the aberrant lip muscle forces. (**b**) Complete bilateral cleft lip and palate with a widely separated lateral palatal segment. The protruding premaxilla extends forward of the lateral palatal segments and is attached to the vomer. The prolabium (central portion of the lip) overlies the premaxilla

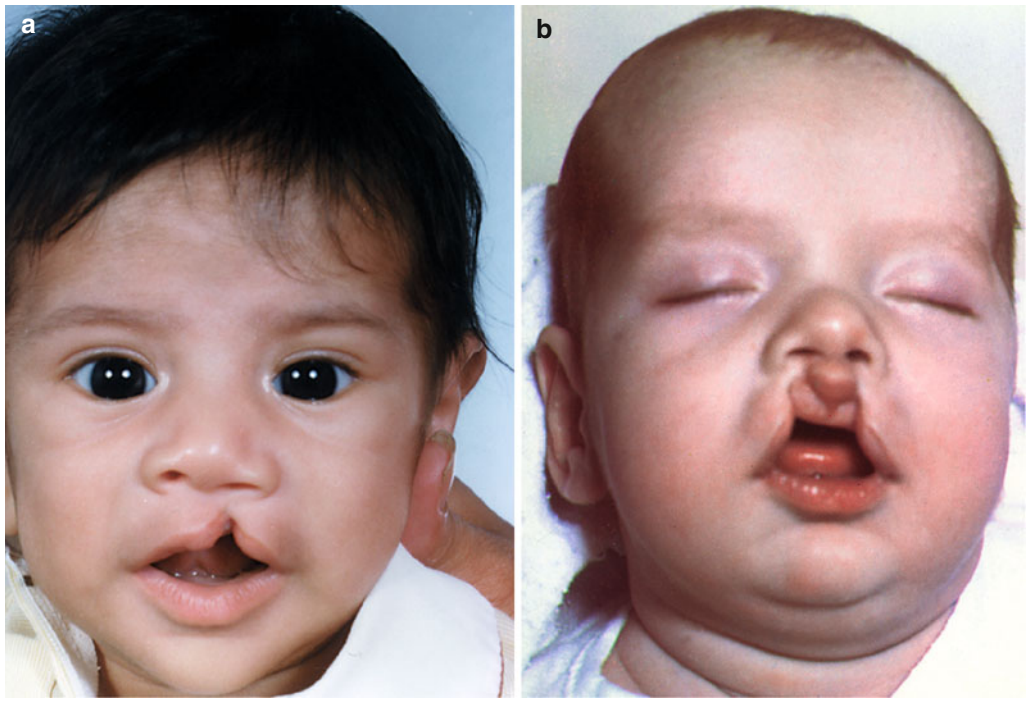


Fig. 4.6 (**a**, **b**) Incomplete clefts of the lip. (**a**) Unilateral and (**b**) bilateral

premaxilla and to the columella. This portion of the lip contains the philtrum. In complete bilateral clefts of the lip, the premaxilla protrudes considerably forward of the facial profile (Fig. 4.5b). It is attached to a stalklike vomer and to the nasal septum. The columella appears to be deficient, and the alar cartilages are flattened on both sides. The effect on the facial profile is to accentuate further the protrusiveness of the premaxilla and the portion of the lip which is attached to the facial surface.

The more complete the defect in the lip, the greater the influence of the cleft on the alveolar process. Because of this constant relationship between the lip and alveolar process, it is not necessary to include the alveolar process as a separate entity in this description and classification. The maxillary alveolar processes arise from the mesoderm in the depths of a sulcus separating the lip and palate, while the tegmen oris gives rise only to the soft palate and the central part of the hard palate.

The relationship between the degree of the cleft's effect on the alveolar process and defects in the deciduous and permanent dentition is interesting. The dental defect may be assessed in terms of the number of teeth, their shape, and structure as well as the position of the teeth in the dental arch. Irregularities in the alveolar process range from small dimples in association with minor clefts in the lip to actual grooves in the alveolar process to, in extreme cases of total clefts in the alveolar ridge, displacement of the premaxillary segment toward the noncleft side. Small dimples or grooves in the alveolar ridge tend to fill in as the jaw grows. However, the deciduous lateral incisor that erupts in this area may be T-shaped, or otherwise misshapen, and malpositioned in the line of occlusion. Further documentation and analysis of serial records should provide detailed information concerning the eruption of teeth adjacent to the cleft in the alveolar process.

4.3.2 Cleft Lip and Cleft Palate (Fig. 4.7)

Clefts of both the lip and palate may be unilateral or bilateral. They may be complete or incomplete. In a complete unilateral cleft of the lip and palate, a direct communication exists between the oral and nasal cavities on the side of the palate where the cleft is situated. The nasal septum is attached to the palatal process on the opposite side, thus separating the nasal chamber from the oral cavity.

A remarkable range of variation exists in each category. Indeed, various degrees of incompleteness of the cleft in the lip and palate may exist in combinations too numerous to describe conveniently. Moreover, some unilateral clefts of lip and palate exhibit wide separation of the palatal shelves. Others exhibit less separation, and in some cases, the segments actually overlap (Fig. 4.8). The palatal segment on the side of the cleft is often tilted medially and upward. The vomer is deviated from the midline at the line of attachment to the palatal process on the noncleft side. This deviation may be so extreme that the vomer assumes a nearly horizontal position at its inferior margin.

The bilateral cleft lip and palate also may be complete or incomplete (Fig. 4.9). If incomplete, it may be symmetrical or asymmetrical, depending on the equality of involvement on both sides. In the complete bilateral cleft lip and palate, both nasal chambers are in direct communication with the oral cavity. The palatal processes are divided into two equal parts, and the turbinates are clearly visible within both nasal cavities. The nasal septum forms a midline structure that is firmly attached to the base of the skull but is fairly mobile in front where it supports the premaxilla and the columella. Cephalometric roentgenograms reveal

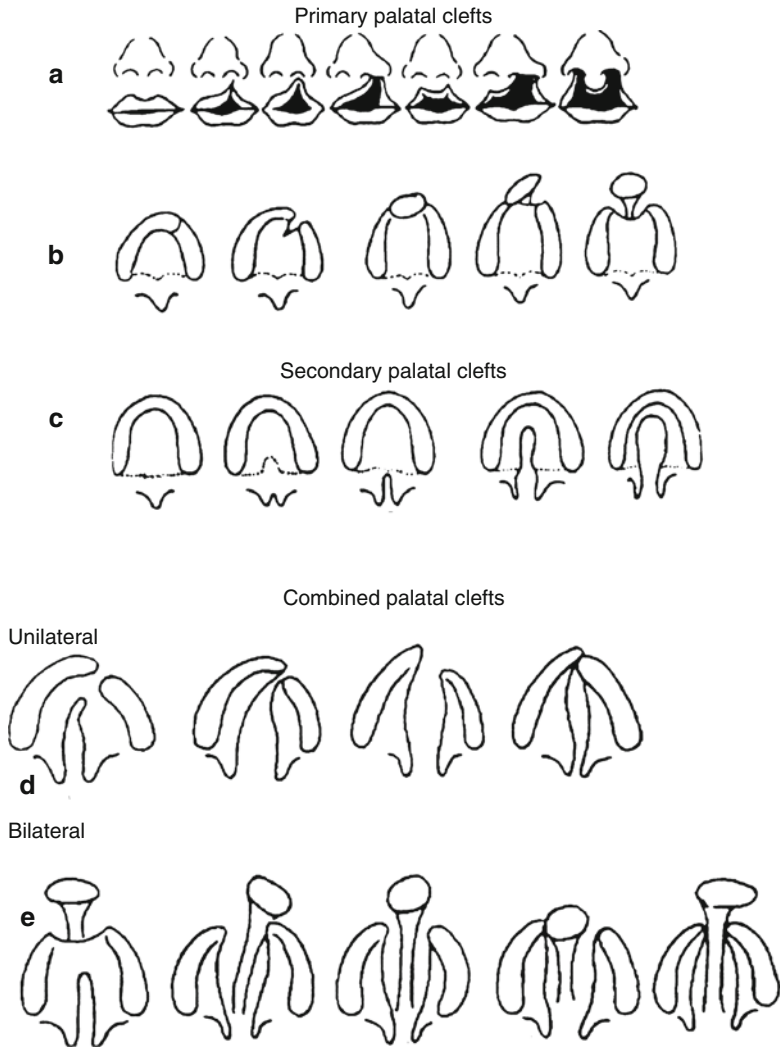


Fig. 4.7 (**a**–**e**) Variations in the form, size, and extent of clefting in primary, secondary, and combined palatal clefts. Primary palatal clefts (with normal hard palate). *Top row*: (**a**) Normal lip. (**b**–**g**) The clefts may involve the lip only or may include the alveolus (tooth-bearing area) as well. The cleft can extend toward the nostril on one or both sides. *Middle row*: The cleft of the alveolus can extend to the incisal papilla on one or both sides to any degree. Bilateral alveolar clefts: (**c**) incomplete on both sides, (**d**) incomplete on one side and complete on the opposite side, and (**e**) complete on both sides. Secondary palatal clefts: (**a**) normal palate, (**b**) bifid uvula, (**c**) cleft of soft palate, (**d**) isolated cleft palate (moderate), and (**e**) isolated cleft palate (extensive). Combined palatal clefts. *Unilateral*: (**a**) Isolated CP with cleft lip and alveolus; (**b**) incomplete unilateral cleft lip and palate (*IUCLP*), cleft lip and alveolus are incomplete; (**c**) complete unilateral cleft lip and palate (*CUCLP*); and (**d**) incomplete unilateral cleft lip and palate (*IUCLP*). *Bilateral*: (**a**) Complete bilateral cleft of lip and alveolus; (**b**) bilateral-complete on one side, incomplete on the opposite with complete hard palate cleft; (**c**) complete bilateral cleft of the lip and palate; (**d**) bilateral incomplete alveolar cleft on one side, complete alveolar cleft on opposite side; and (**e**) complete bilateral alveolar clefts with both palatal segments attached to the vomer

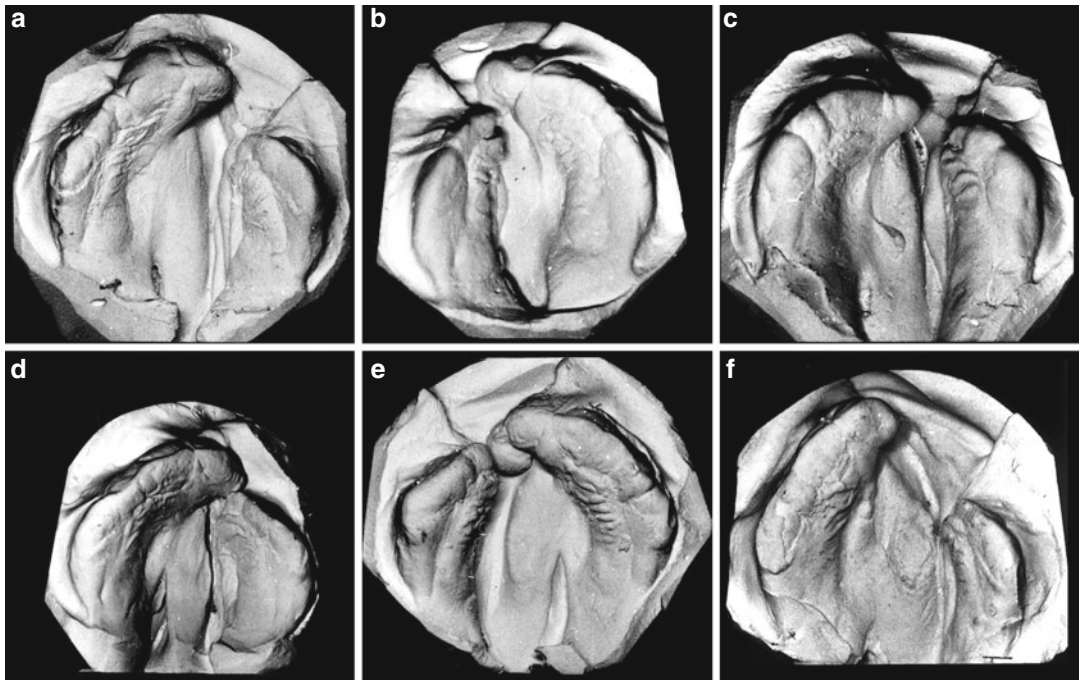


Fig. 4.8 (**a**–**f**) Variations in unilateral clefts of the lip and palate at birth. The palatal segments may be complete (**a**, **c**, **e**, **f**) or incomplete (**b, d**); the cleft segment may be almost of the same length or shorter. The cleft space may be relatively narrow (**b**, **d**) or wide (**a**, **c**, **e**, **f**)

the existence of a suture line, the premaxillary-vomerine suture, between the vomer and the premaxilla. This suture plays an important role in facial growth and is also a point of flexion for the premaxilla upon the vomer (Fig. 4.3b, c).

The premaxilla may be small or large, symmetrical or asymmetrical. The number of incisor teeth contained in this segment is directly related to its size and shape. Permanent teeth may be missing, and it may contain only one or more deciduous teeth when the cleft of the lip is complete on both sides, and the premaxilla projects considerably forward from the facial aspect of the maxillae. This anterior protrusion is less evident if the lip is incompletely cleft on one or both sides.

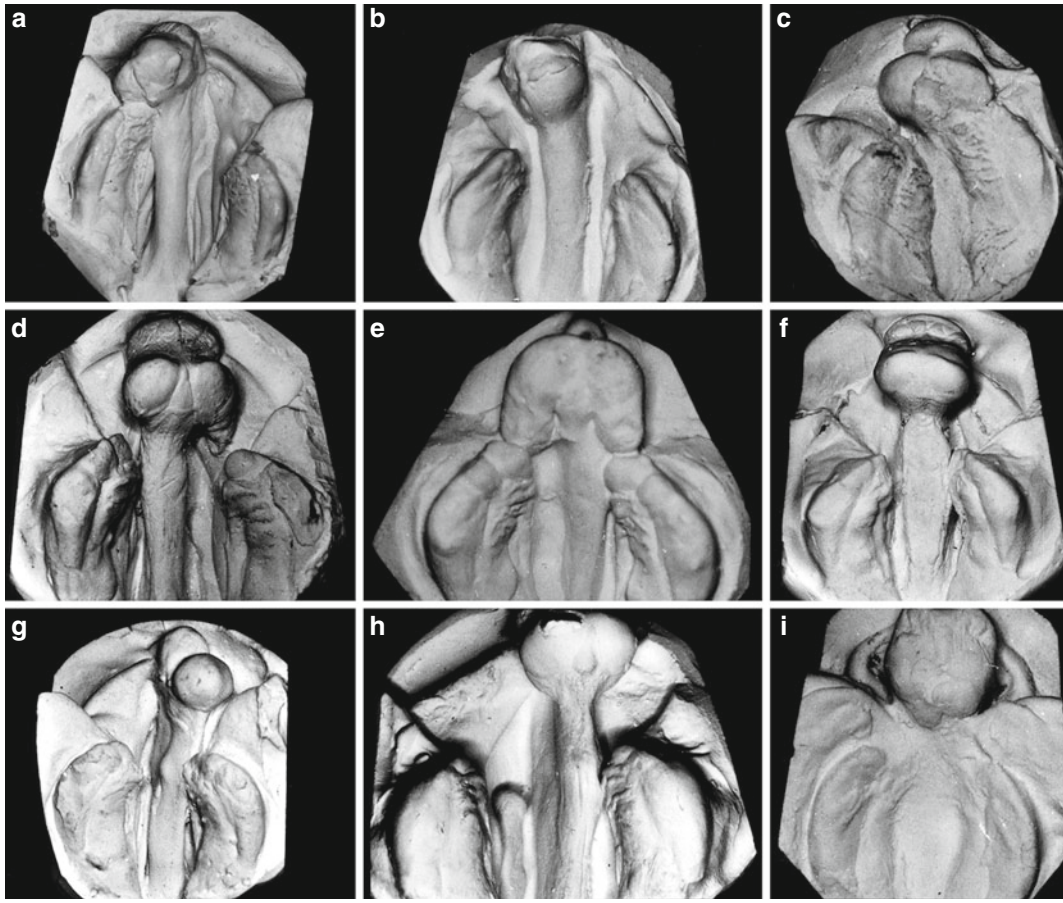


Fig. 4.9 (**a**–**i**) Variations in bilateral cleft lip and palate. The size of the premaxilla varies with the number of teeth it contains. Classification is dependent on the completeness of clefting of the lip and alveolus and whether there is a cleft of the hard and soft palate. Yet one or both sides of the hard palate may or may not be attached to the vomer. If it is attached to the vomer, it is classified as being incomplete. Even in complete clefts of the lip and alveolus, the extent of premaxillary protrusion will vary. (**a**) Incomplete bilateral cleft lip and palate. Complete cleft lip and palate – *left side*. Incomplete cleft lip and palate – *right side*. (**b**) Complete bilateral cleft lip and palate. Complete cleft palate – both sides. (**c**) Incomplete bilateral cleft lip and palate. Incomplete palatal clefts – both sides. (**d**) Complete bilateral cleft lip and palate. Incomplete right and complete left palate. (**e**) Incomplete bilateral cleft lip and palate. Incomplete left palate and complete right palate. (**f**) Complete bilateral cleft of the lip and palate. Incomplete right and left palatal segments. (**g**) Complete bilateral cleft of lip and palate. Incomplete left palate and complete right palatal segment. (**h**) Complete bilateral cleft lip and palate. Incomplete left palate and complete right palate. (**i**) Incomplete bilateral cleft lip and alveolus. Normal palate

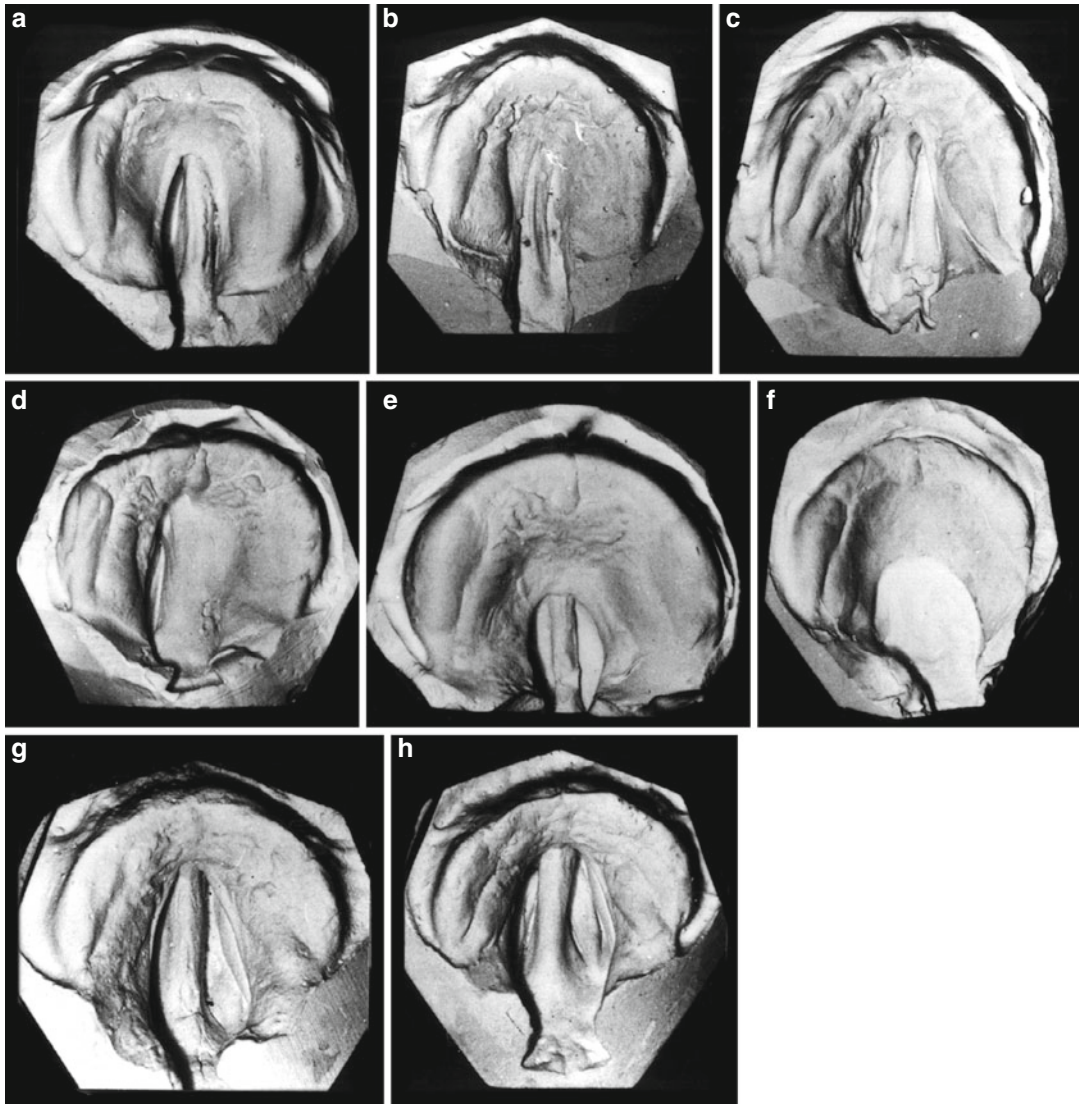


Fig. 4.10 (**a**–**h**) Variations in isolated cleft palate. The length and width of the cleft space is highly variable. The cleft extends anteriorly to various distances but not beyond the incisal canal

4.3.3 Isolated Cleft Palate

In this defect, neither the lip nor the alveolar process is involved (Fig. 4.10). The cleft may involve only the soft palate or both the soft and hard palates but never the hard palate alone. This observation is in accordance with the finding that fusion of the hard and soft palates proceeds from front to back (Fig. 4.11).

The cleft may extend forward from the uvula to varying degrees. In some cases, the cleft is limited to the uvula or to the uvula and soft

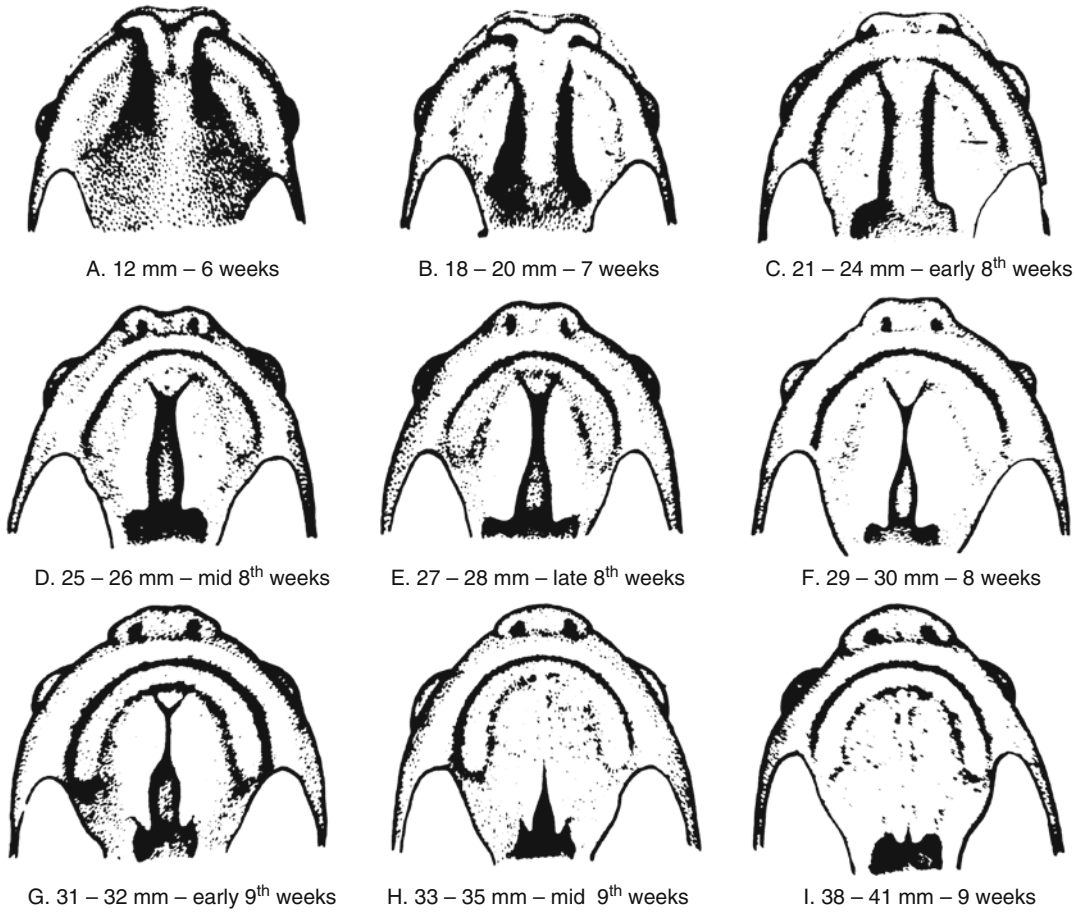


Fig. 4.11 Graphic summary of palatal fusion. The measurements and estimated ages given should be regarded as averages (Redrawn from illustration in Kraus et al. (1966))

palate. In others, it may extend into the hard palate. It is recommended that a digital examination of the posterior edge of the hard palate be performed. A midline notching will reveal the presence of a submucous cleft. The full extent of submucous clefts can be mapped by cephalometric laminagraphy or by transillumination through the nose.

In the extreme form, the cleft palate may extend anteriorly as far as the nasopalatine foramen, the incisal canal. When the cleft involves a considerable portion of the hard palate, the nasal chambers are in direct communication with the oral cavity. In most instances, the nasal septum is not attached to either palatal process throughout the extent of the cleft. However, occasional asymmetries may be noted in which the septum is attached to a portion of the palatal process on one side to a greater extent than to the palatal process on the opposite side.

In this cleft type (Fig. 4.12), neither the lip nor the alveolar process is involved. The cleft may extend only through the soft palate or through the soft and hard palates, but it cannot exist in the

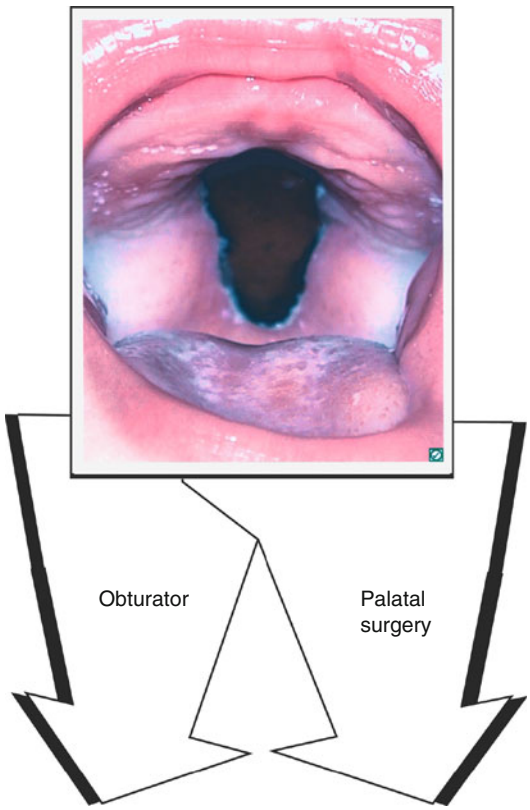


Fig. 4.12 Isolated cleft palate

hard palate alone because the fusion of the hard and soft palates proceeds from front to back (Figs. 4.13 and 4.14).

The cleft may extend anteriorly as far forward as the nasopalatine foramen. The outline of the cleft space may be wide or narrow, long or short; any variation in geometric form can exist (Figs. 4.15, 4.16, and 4.17; see Chap. 2).

Timing of surgical closure depends on the width and not the length of the cleft space. Hard palate clefts are frequently closed simultaneously with the soft palate cleft. In some instances, surgical closure of very wide hard palate clefts may need to be postponed until there is additional palatal growth, which may be as late as 5 or 6 years of age. An obturator with a pharyngeal extension (speech aid appliance) can be worn until the palate is closed (Figs. 4.18 and 4.19).

The outline of the cleft may be wide or narrow, pyriform, or V-shaped. Excessively wide dental arches often are associated with wide clefts that extend to a considerable degree into the hard palate. In such instances, the mandibular dental arch may be in complete lingual relation to the maxillary arch so that the cusps of the teeth do not interdigitate in occlusion.

Lateral cephalometric headplates reveal that the dorsum of the tongue, at rest, is elevated and postured within the nasal cavity. During deglutition, the thrusting action of the tongue operates to separate the palatal processes. These abnormalities in the posture and movements of the tongue are supported by the observations of speech pathologists. In this type of cleft, the vomer is significantly different in size and form from that observed in bilateral cleft lip and palate. In both types, the vomer is seen as a midline structure extending downward from the base of the skull. However, in bilateral cleft lip and palate, the inferior border of the vomer is thick and rounded, whereas in this category – cleft palate only – the vomer is thin and knife-edged. Serial observations reveal that the pattern of growth exhibited by the vomer is different in these two types of clefts.

Several other distinguishing characteristics apparent in some of the clefts in this category merit further comment. The high incidence of mandibular micrognathia found in patients with cleft palate gives credence to the theory that during embryonic development the tongue did not sink below the palatal processes and thereby prevented their fusion in the midline. This raises the question of whether more than one causal mechanism might exist to produce the various kinds of clefts of the lip and palate.

In an extensive study of the mode of inheritance of cleft lip and cleft palate, Fogh-Andersen (1942, 1961) concluded that there are two different malformations with no genetic connection. In one group are clefts that involve the lip and occur most frequently in male patients. The other group is limited to clefts of the palate, which are more frequent in female patients. According to Fogh-Andersen, the manner of inheritance differs for the two groups.

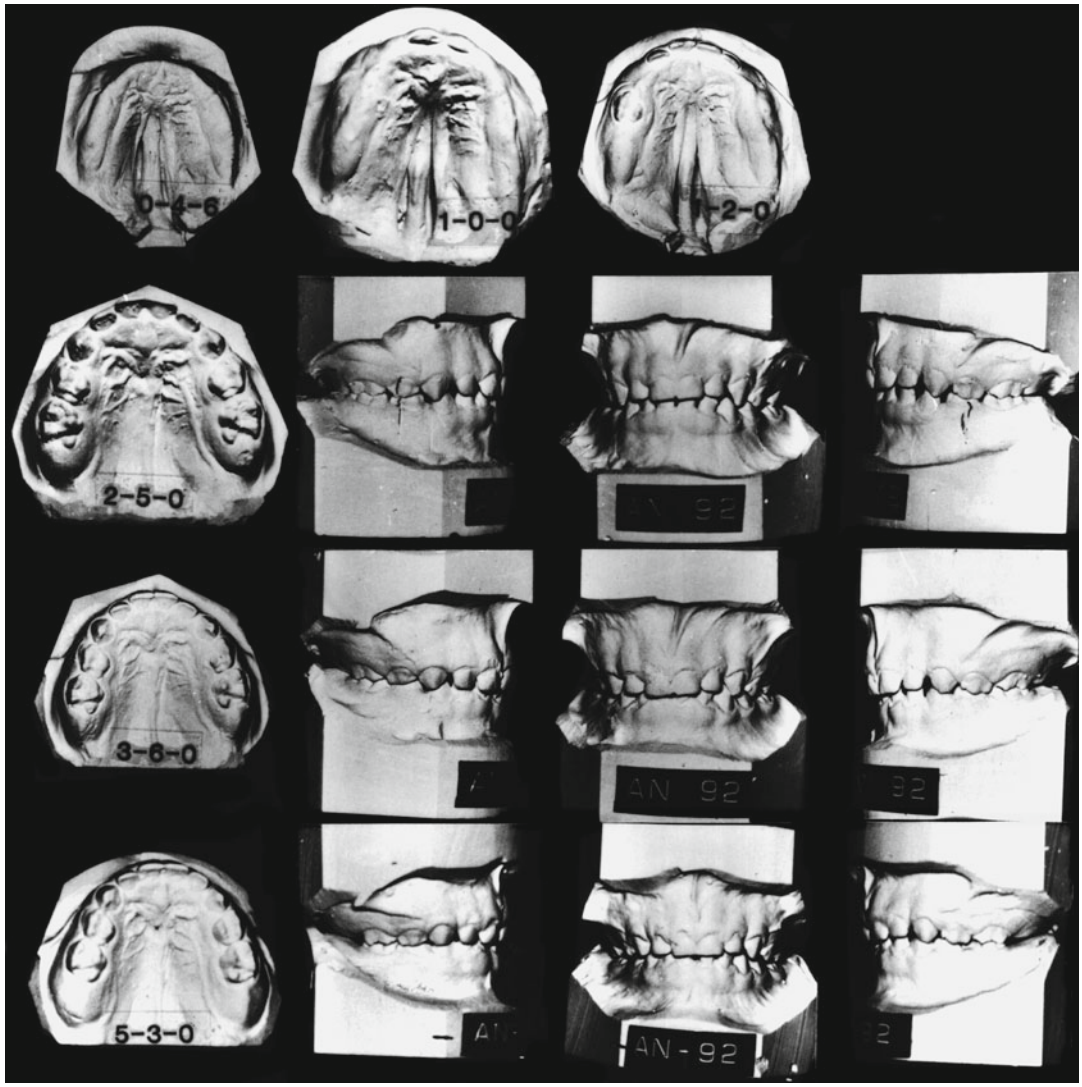


Fig. 4.13 Isolated cleft palate. Serial casts demonstrate that, in some cases, good class I occlusion can occur when closing the cleft space at 18 months of age, or even later, using a modified von Langenbeck surgical procedure. This occlusal result supports Berkowitz's contention that the "critical threshold level" for good palatal growth is determined by the size of the cleft space relative to the amount of available mucoperiosteal tissue. This threshold, which has been determined to be a ratio of cleft size to palatal size medial to the alveolar ridges of 10 % or less, is critical to the development of good arch form and occlusion. The threshold ratio, if exceeded by early palatal closure when the palatal size is relatively small compared to cleft size, will cause excessive palatal scar tissue resulting in midfacial growth inhibition

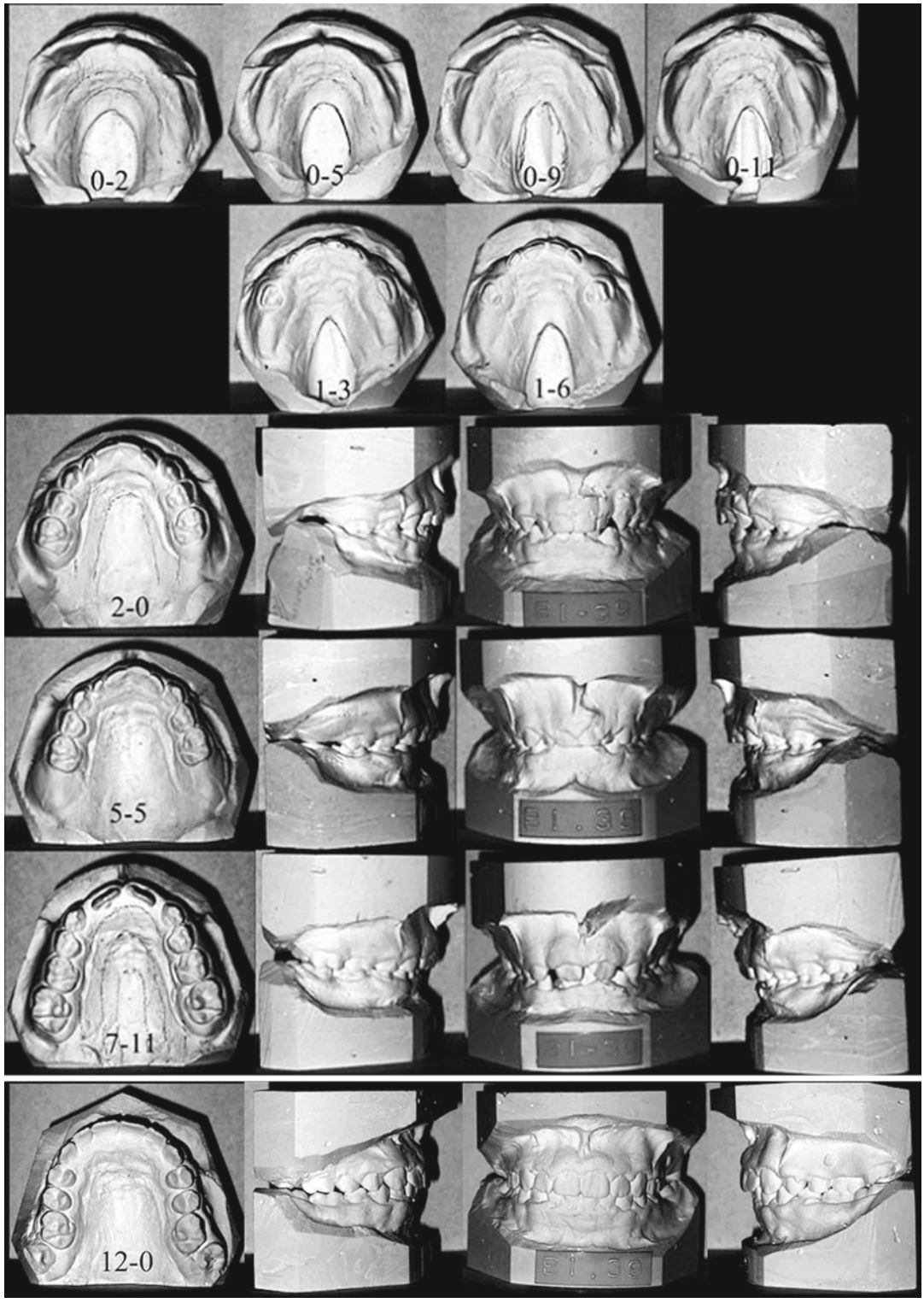


Fig. 4.14 Case CR-BI 39. Patient with isolated cleft palate. Notice that the relative size of the cleft space diminishes as the palate grows and increases in size. The palate was closed with the von Langenbeck at 18 months. Note the excellent occlusion which reflects on diminished palatal scarring. The patient has excellent speech

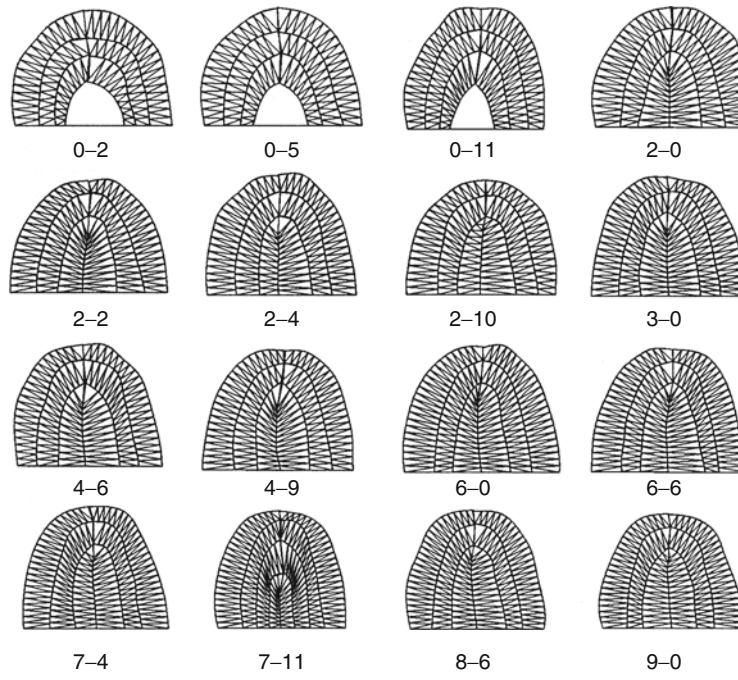


Fig. 4.15 Superimposed computerized 3D tracings of isolated cleft palate. Isolated cleft palate. Superimposition horizontally on the rugae and registered vertically on the vomer line. This shows that most of the growth occurs posteriorly to accommodate the developing deciduous molars. Growth changes of the cleft space are highly variable. In some instances, the cleft space can narrow or stay the same size. However, in all instances, the palate increases in size so the ratio of cleft space to surrounding palatal size reduces. In most cases, time for surgery, determined by the ratio of 10 %, is reached at 18–24 months. It can be earlier or even later in some cases

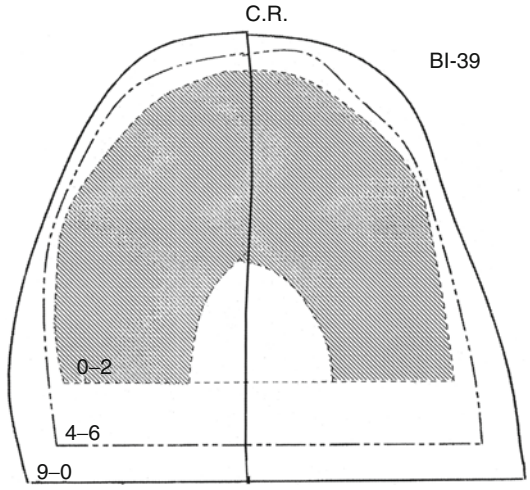


Fig. 4.16 Case CR-BI 39

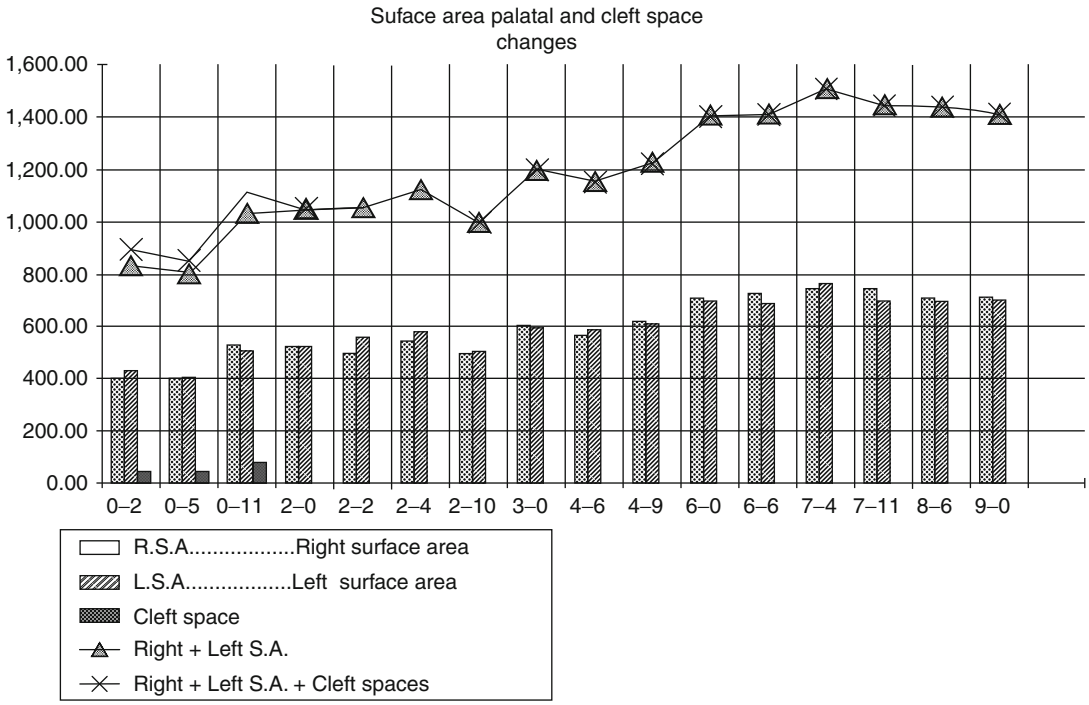


Fig. 4.17 Serial size changes of the palate and cleft space

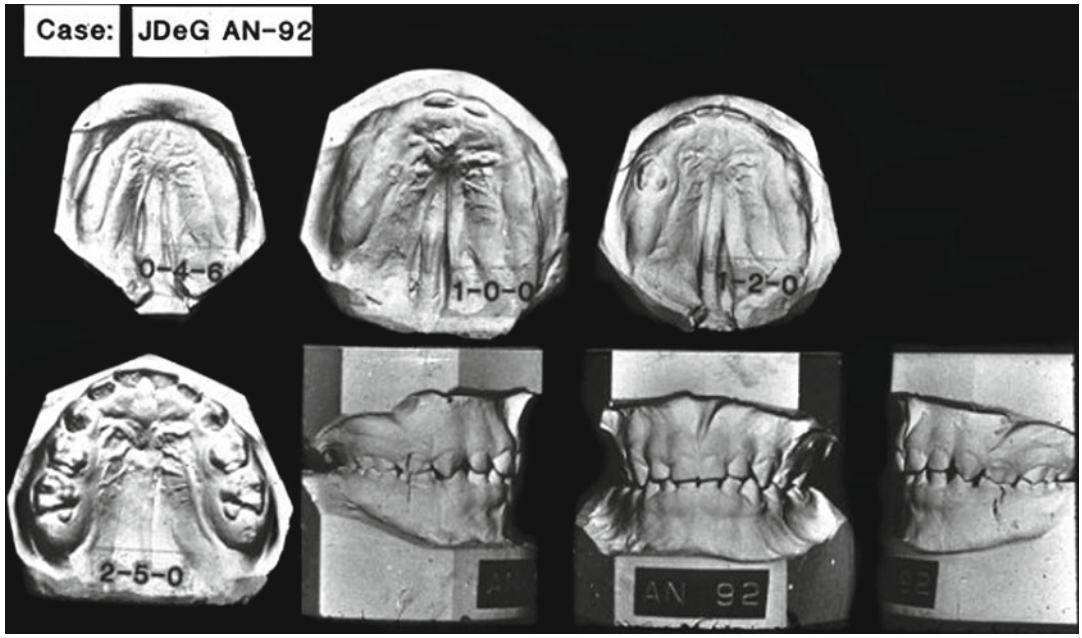


Fig. 4.18 Case AN-92. Palatal closure at 1–5 using von Langenbeck procedure resulting in good occlusion

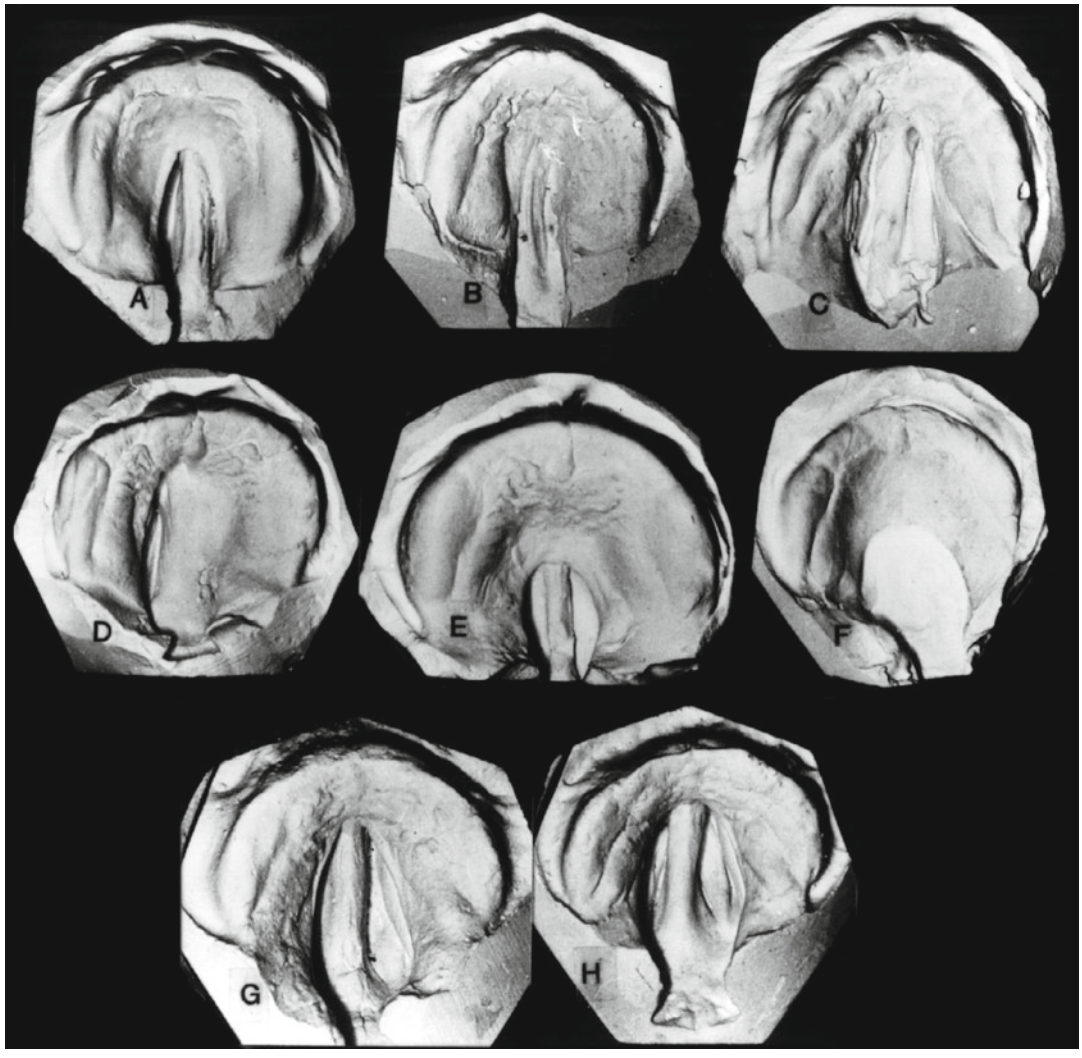


Fig. 4.19 (**a**–**h**) Variations in isolated cleft palate. The length and width of the cleft space is highly variable. The cleft extends anteriorly to various distances but not beyond the incisal canal. Note the relative size of the cleft palate space which reflects differences in the degree of osteogenic deficiency. In some instances, the cleft space diminishes with time; however, there are occasions when the cleft's relative size remains the same

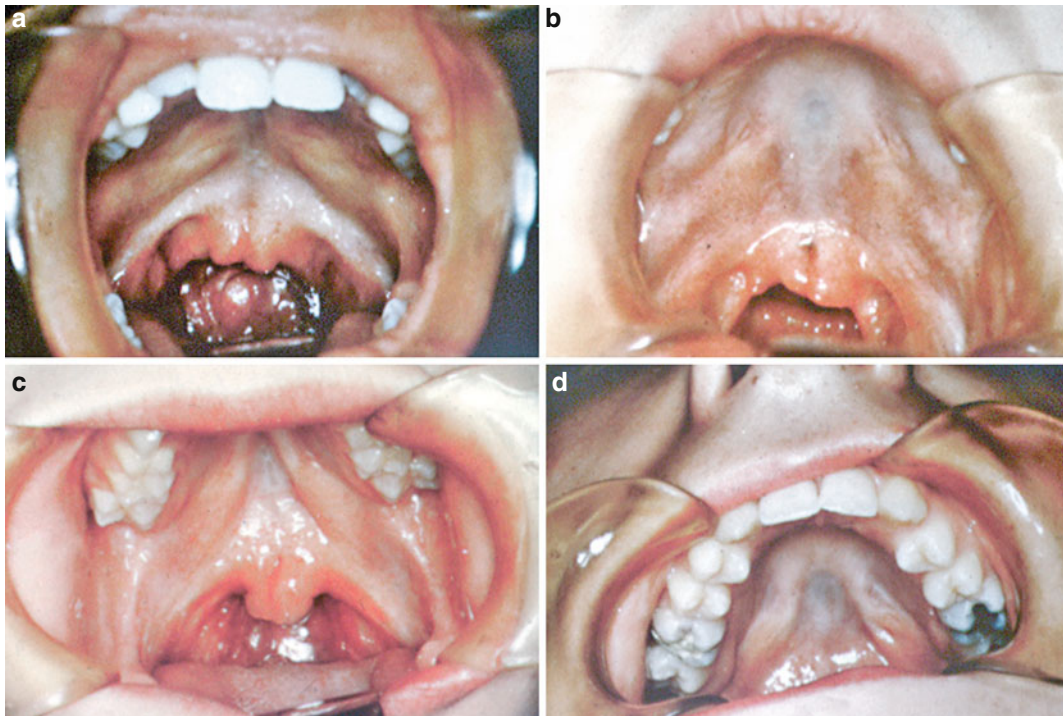


Fig. 4.20 (**a**–**d**) Submucous cleft palate. This cleft is characterized by a bifid uvula, lack of muscle continuity across the soft palate, and a pink zone of mucosa (zona pellucida) across the cleft in the hard palate. A palpable notch in the posterior border of the hard palate is always indicative of the presence of a cleft

4.3.4 Submucous Cleft Palate (Fig. 4.20)

The classic triad of diagnostic signs is the bifid uvula, partial muscle separation in the midline with an intact mucosal surface, and the midline notch in the posterior edge of the bony palate. Hypernasality may or may not exist. Caution needs to be exerted prior to performing tonsillectomies and adenoidectomies because the velum may be functionally too short without the presence of the adenoid mass.

Berkowitz's cephalometric and nasopharyngoscopic studies have shown wide and unpredictable variability in the pharyngeal skeletal architecture and velar size and shape in submucous cleft palate as well as in all other cleft types (see Chap. 12). In some cases, due to a shallow pharyngeal space with relatively good velar length and mass with good lateral pharyngeal wall movement, no hypernasality existed. However, in most cases, the velum is usually too short, as well as too thin, and it fails to obturate the pharyngeal space properly. The problem appears to be due to an inadequate velum rather than a deficiency in lateral wall movements.

The treatment of choice is a well-positioned and adequately wide superior-based pharyngeal flap. There is no apparent need to combine a palatoplasty with the pharyngeal flap. In patients with recurring infections of the adenoids, it is recommended that the adenoids be removed before the flap is placed. Speech therapy is an integral part of postoperative management.

4.4 Congenital Palatal Insufficiency (CPI)

It has been said that cleft palate is a type of defect that can be "seen, felt, and heard." By contrast, the defect known as congenital palatal insufficiency (CPI), until recently, has been more readily heard than seen or felt. This anomaly is seldom apparent at birth, and the first awareness of the defect occurs when the child develops the hypernasality characteristic of uncorrected cleft palate speech. The variety of factors producing this kind of speech defect can be examined by roentgenographic and nasopharyngoscopic methods.

Normally, during deglutition and during phonation, except for the sounds of "m," "n," and "ng," the soft palate elevates, making contact with the posterior and lateral walls of the pharynx. The complicated synergies that contribute to this multidimensional contraction serve to separate the nasopharynx from the oropharynx. If for any reason this velopharyngeal closure cannot be achieved, deglutition is compromised, and in phonation, the air stream necessary to create speech is misdirected through the nose. Palatal insufficiency may be caused by the velum being too short and/or by a deficiency in the anteroposterior dimension of the hard palate.

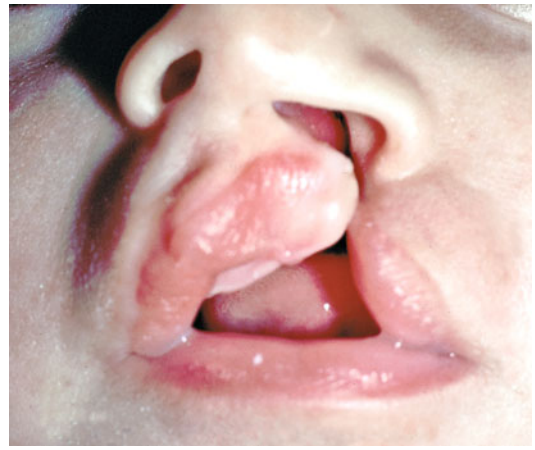


Fig. 4.21 Clefts of the Lip and Alveolus: (Figs. 4.21–4.27) The alveolar portion is usually distorted outward in complete lip clefts. When the lip is united, the newly created lip force molds the alveolar section into proper alignment. A secondary alveolar bone graft is performed at the same age as in other cleft types

4.5 Clefts of the Lip and Alveolus (Figs. 4.21, 4.22, 4.23, 4.24, and 4.25)

The alveolar portion is usually distorted outward in complete lip clefts. When the lip is united, the newly created lip force molds the alveolar section into proper alignment. A secondary alveolar bone graft is performed at the same age as in other cleft types (Fig. 4.22).

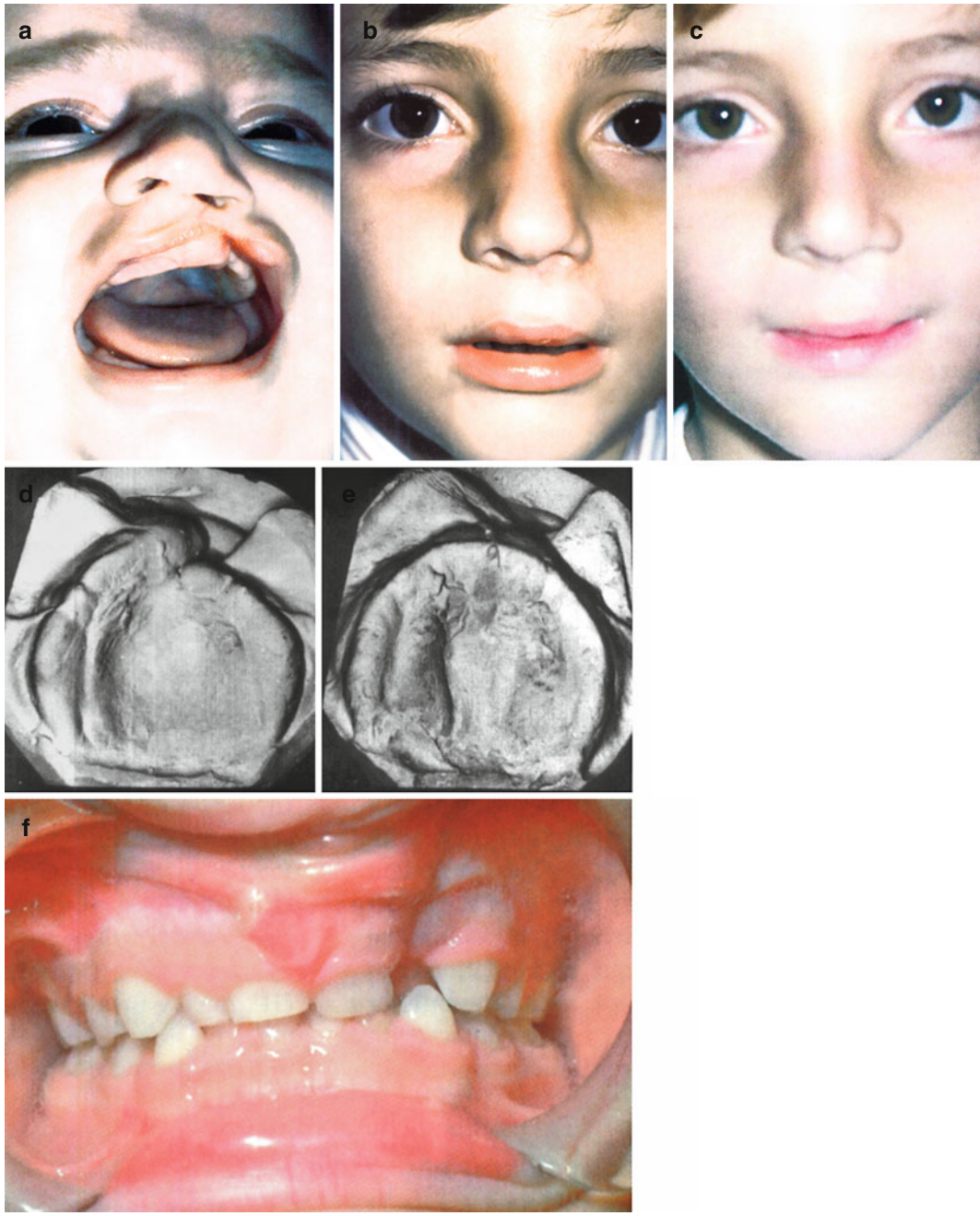


Fig. 4.22 (**a**–**f**) Cleft of the lip and alveolus. Facial photographs: (**a**) before and (**b**) after surgery, (**c**) 4 years later showing good lip/nose aesthetics and palatal casts, (**d**) before and (**e**) after surgery showing the molding of the cleft alveolar segment into alignment, and (**f**) ideal anterior and buccal occlusion

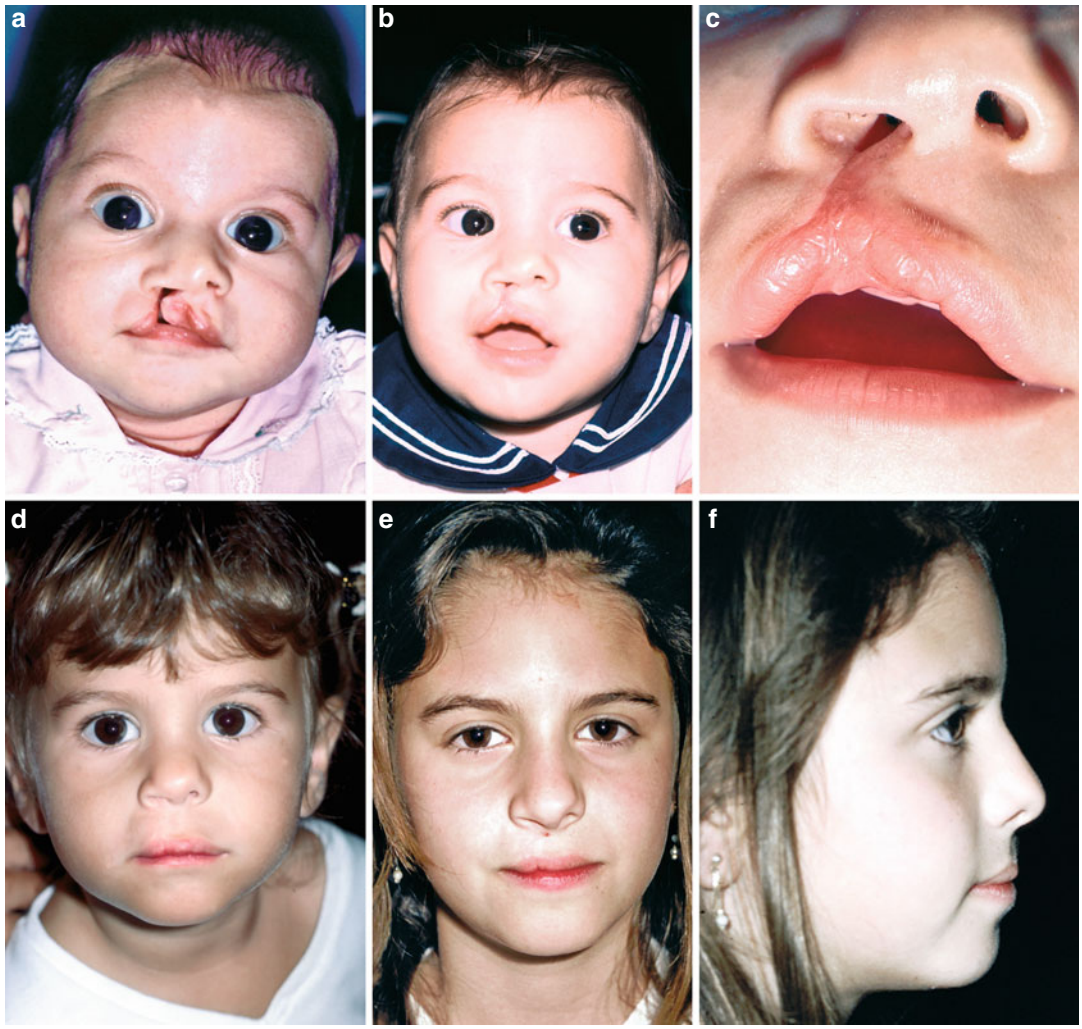


Fig. 4.23 Case AS-30. Cleft of the lip and alveolus. Various lip revisions were performed

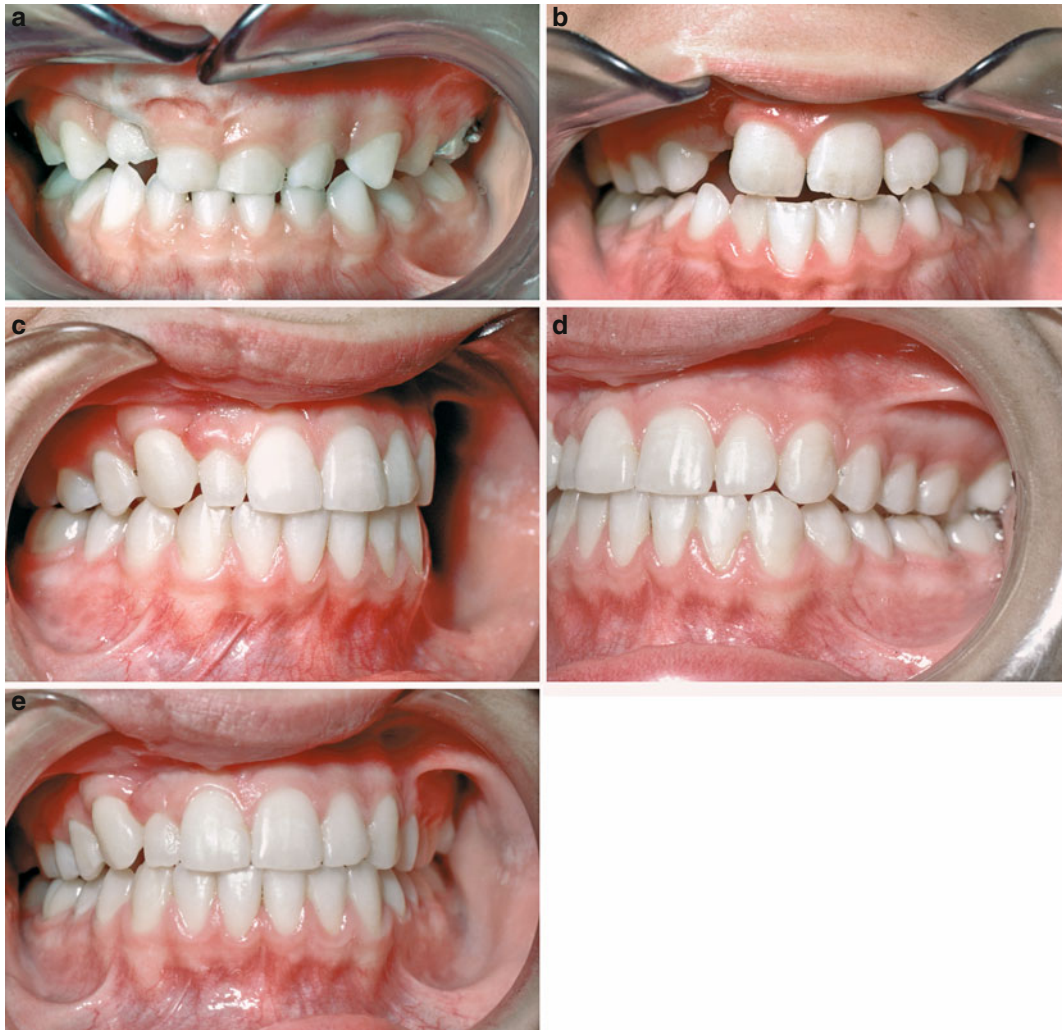


Fig. 4.24 Case AS-30. Cleft of the lip and alveolus after orthodontics

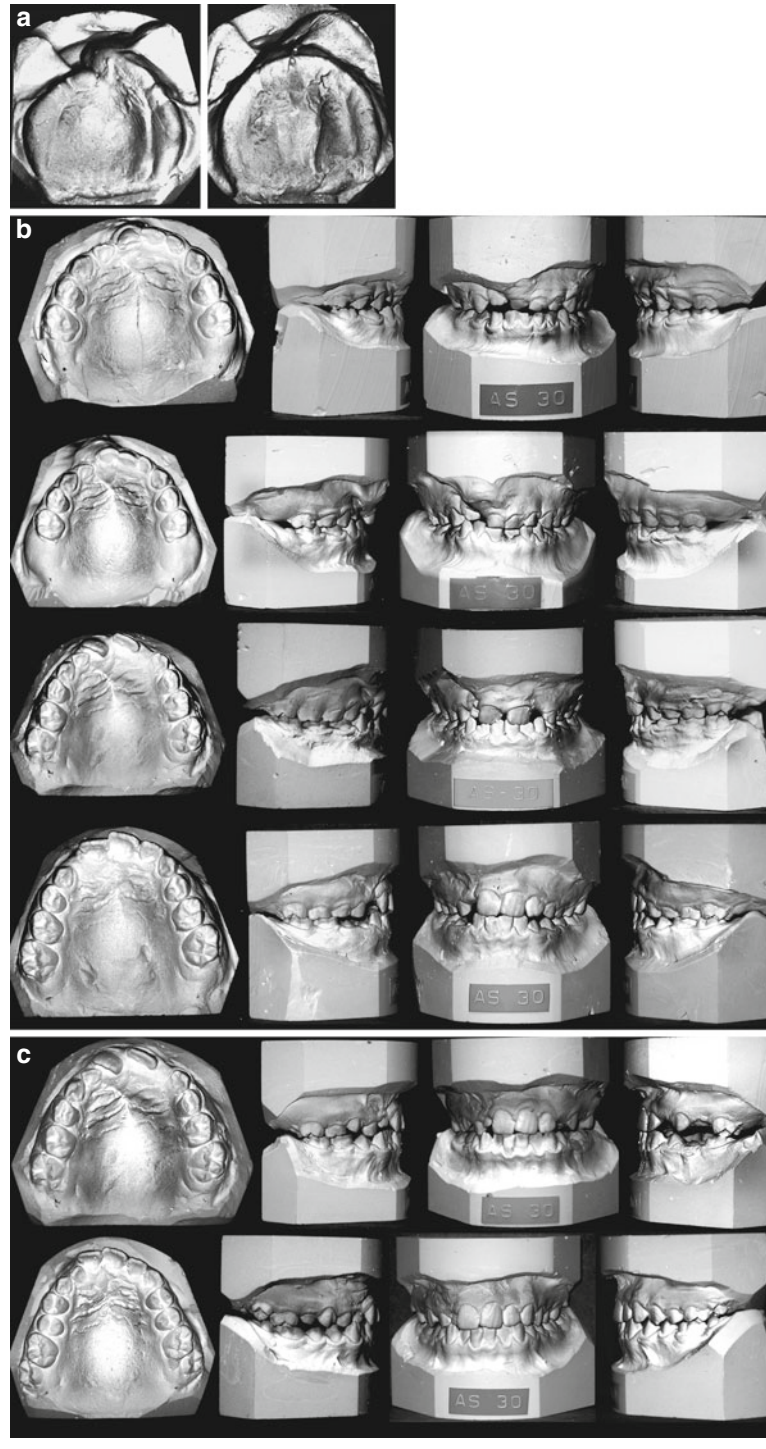


Fig. 4.25 Case AS-30. Lip adhesion treatment and alveolar bone graft. Palatal growth and development was normal as seen by the occlusion

4.6 Clefts of the Uvulae and Soft Palate (Fig. 4.26) and Cleft of the Uvulae Alone (Fig. 4.27)

When the health of the child permits, soft tissue clefts can be sutured within the first 3-month forces as Latham and his mentor McNeil have suggested. There are, however, some cases when, because of an unfavorable facial growth pattern coupled with a retruded maxilla relative to the anterior cranial bases, orthopedic protraction forces will be beneficial in the mixed (transitional) and permanent dentition.

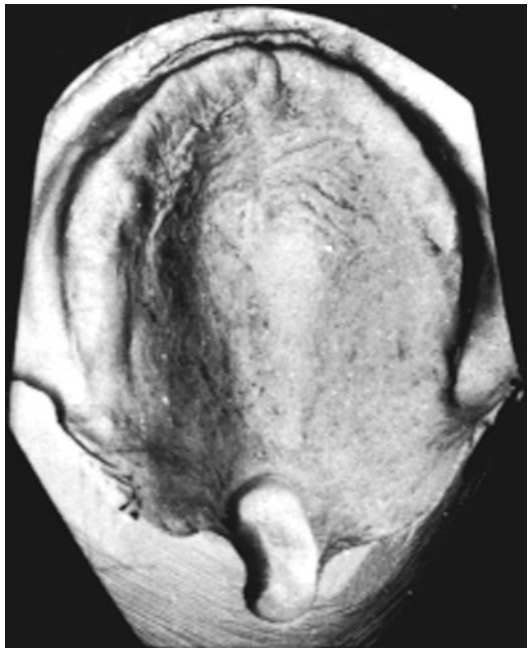


Fig. 4.26 Soft palate

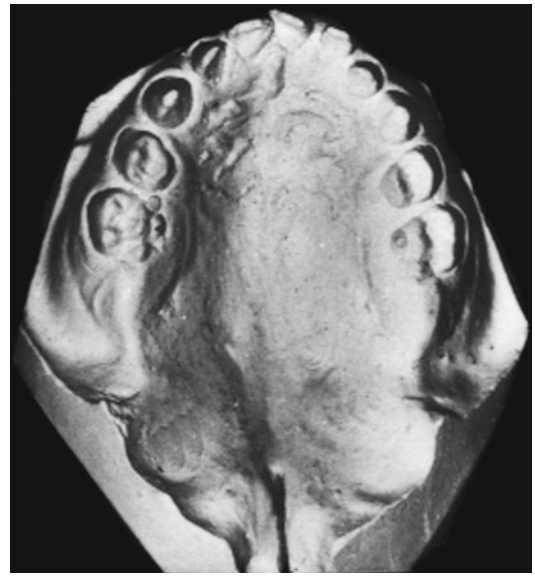


Fig. 4.27 Uvulae. Always indicative of the presence of a cleft

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Alternative Method Used to Correct Distorted Neonatal Cleft Arch Forms

5

Samuel Berkowitz

5.1 Effects of Reversing the Facial Force Diagram

The influence of soft-tissue forces on palatal form and growth has been the topic of several studies. Ritsila and coauthors (1972) reported that there was "slight shortening" of the maxilla, "marked shortening" of the body of the mandible, and alterations of several mandibular angles after closure of the lip.

As perhaps an interesting footnote (Ritsila et al. 1972; Bardach et al. 1982), physical changes to the palate in clefts of the lip and palate in animals are very similar to the corresponding changes that are seen in humans. Bardach et al. (Ritsila et al. 1972; Bardach et al. 1982) studied lip pressure changes following lip repair in infants with unilateral clefts of the lip and palate. They confirmed the belief that lip repair significantly increases lip pressure when compared with a noncleft population.

Berkowitz's (1959, 1969) data demonstrated that the force of the united lip against the protruding premaxilla in complete bilateral clefts of the lip and palate (CBCLP) acts first to bring about premaxillary ventroflexion. After 2–3 years, there is some appearance of midfacial growth retardation to various degrees. There is strong evidence that uniting the lip does not "telescope" the premaxilla into the vomer, whereas mechanical premaxillary retraction "telescopes" the premaxilla in almost all instances (see Chap. 21). In very rare instances, it may even cause a vomer fracture.

5.2 Variations in the Palate's Arch Form

The size and relationship of the palatal segments to each other are highly variable (see Figs. 4.8 and 4.9). As already described, in complete clefts of the lip and palate, the lateral palatal segments are displaced laterally and the slopes of both palatal segments are steeper than normal, with the palatal segments at the cleft space extending into the nasal chamber (Berkowitz 1985). This steepness decreases with time, the slopes becoming more obtuse under the influence of tongue force. In clefts of the lip and palate, uniting the cleft orbicularis oris-buccinator superior constrictor muscle ring or using external facial elastics reestablishes the outer compressive muscular forces. This change in the muscle force vectors causes the laterally displaced palatal segments to move together. Moreover, this reduction in the width of the cleft is not limited to the alveolar process but

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extends as far back as the tuberosities of the maxilla and perpendicular pterygoid processes. The surgeon is challenged to establish muscle balance without disturbing the growth potential of the bony tissue being manipulated and to avoid scars that will tie or bind down the normally expansive forces of growth.

5.3 Reversing Aberrant Cleft Facial Forces in the Neonate

5.3.1 Lip Surgery, Elastic Traction, or Presurgical Orthodontic Treatment (Figs. 5.1, 5.2, 5.3, and 5.4)

1. Lip surgery creates sufficient forces to bring the overexpanded palatal segments medially narrowing the alveolar and palatal cleft spaces. The surgeon often does this in two stages: first, a lip adhesion at 3–5 months followed by a more definitive lip/nose surgery, which is more artistic. A cupid bow and normal nostrils are the eventual goals (see Chap. 8).
2. Head bonnet with elastic strap to be placed over the premaxilla in all lip clefts. The force system needs to be worn for 1 or 2 weeks along with arm restrains to prevent the infant overjet from removing the elastic strap. A premaxillary ventroflexion in CBCLP cases occurs very quickly creating an overjet and overbite. In CBCLP with a protruding premaxilla at birth, the lateral palatal segments move medially behind the premaxilla. This relationship does not cause palatal growth retardation. Should a crossbite occur, the involved palatal segment usually can be moved laterally into proper occlusion at 4–6 years of age when the child is manageable in a dental chair.
3. Presurgical orthopedics: There are active and passive appliances, which are designed to create an alveolar butt joint (Berkowitz et al. 2004). In the distant past, primary bone grafting was utilized with the hope of stabilizing the palatal segment's position. However, with primary bone grafting, it was found to cause midfacial deformity. Berkowitz, in a recent longitudinal palatal growth study, determined that the plates do not stimulate growth. Some surgeons who have used gingivoperiosteoplasty have created an anterior crossbite in most instances, which is hard to correct with expansion. Berkowitz strongly rejects the use of primary bone grafting and gingivoperiosteoplasty (10).

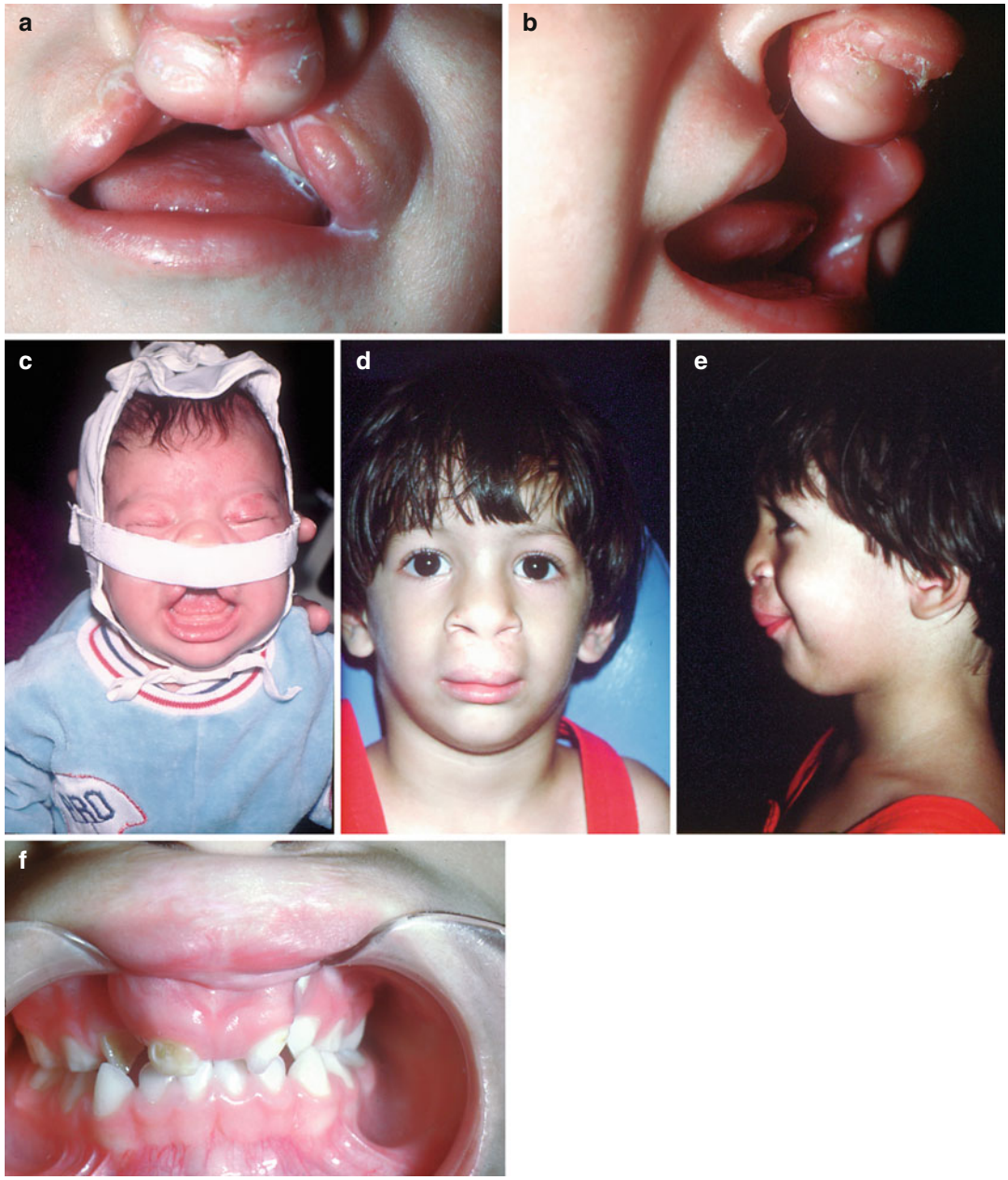

Fig. 5.1 (**a**–**f**) The use of an external elastic force to reduce premaxillary protrusion. (**a**, **b**) The protruding premaxilla extends forward in the facial profile. (**c**) Head bonnet with attached elastic placed against the protruding premaxilla causes it to ventroflex with the fulcrum at the premaxillary vomerine suture. (**d**, **e**) Facial photographs at 3 years of age. The lateral lip elements are united with the medial positioned prolabium over the protruding premaxilla in one stage. Because the premaxilla is already ventroflexed at the time of surgery, there is reduced muscle tension at the suture sites. (**f**) Intraoral photograph shows excellent anterior and buccal occlusion even with bilateral deciduous cuspids in crossbite. Comment: A severe overbite or overjet with a buccal crossbite at this age does not create a functional dental problem or inhibit palatal growth. Midfacial protrusion is expected and even desirable at this age. A straight profile in the mixed dentition usually indicates a concave profile will develop in adolescence after the pubertal growth spurt

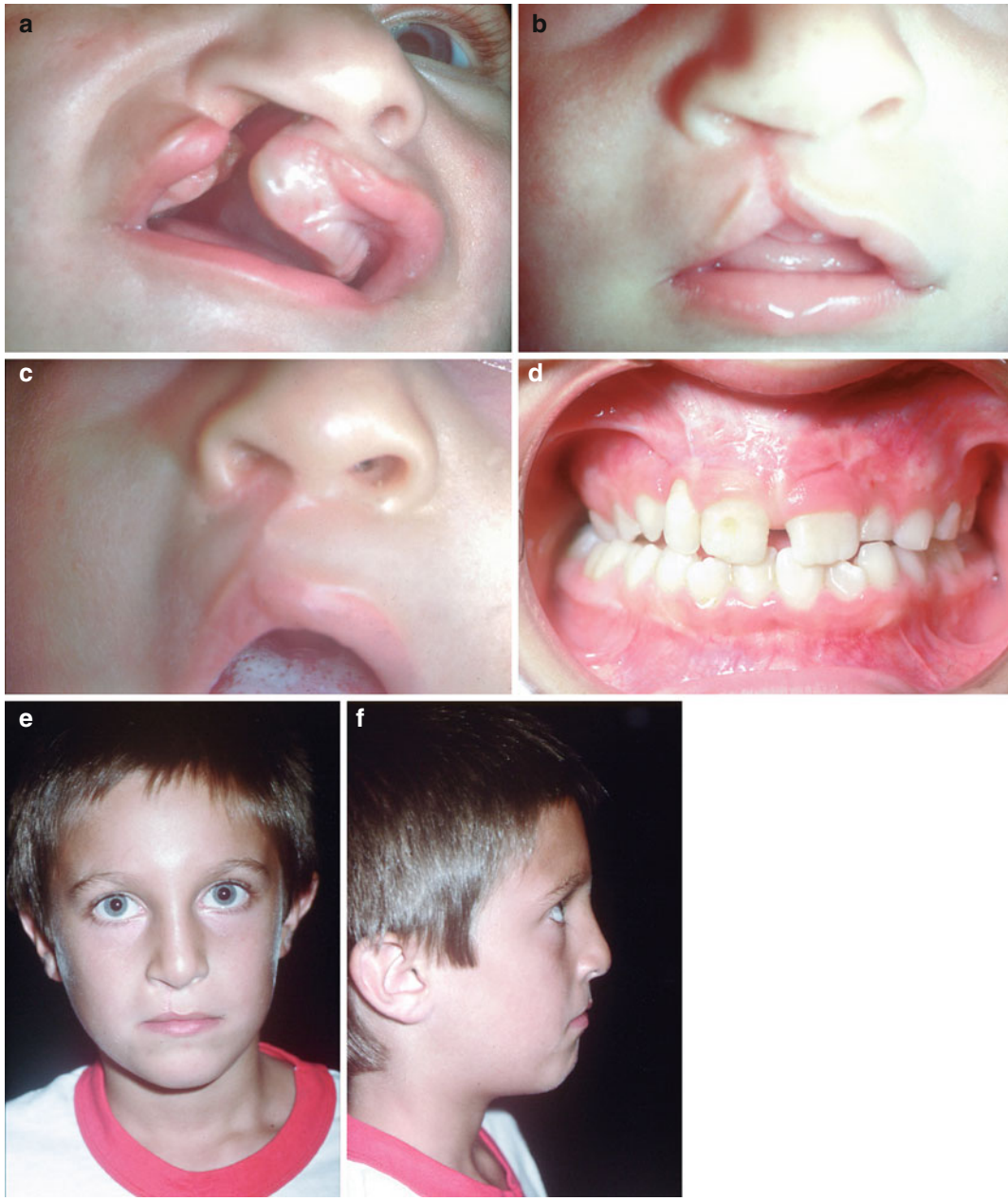

Fig. 5.2 (**a**–**f**) Case MD (AM-17). Conservative surgery with no presurgical orthopedics in CUCLP. Lip adhesion to start molding action to bring the separated palatal segment together. (**a**) At birth. (**b**) After lip adhesion at 5 months. (**c**) After definitive lip surgery at 9 months. (**d**, **e**) Facial appearance at 8 years of age. (**f**) Occlusion at 8 years. The right deciduous lateral incisor erupted through a secondary alveolar bone graft performed at 7 years of age using cranial cancellous bone

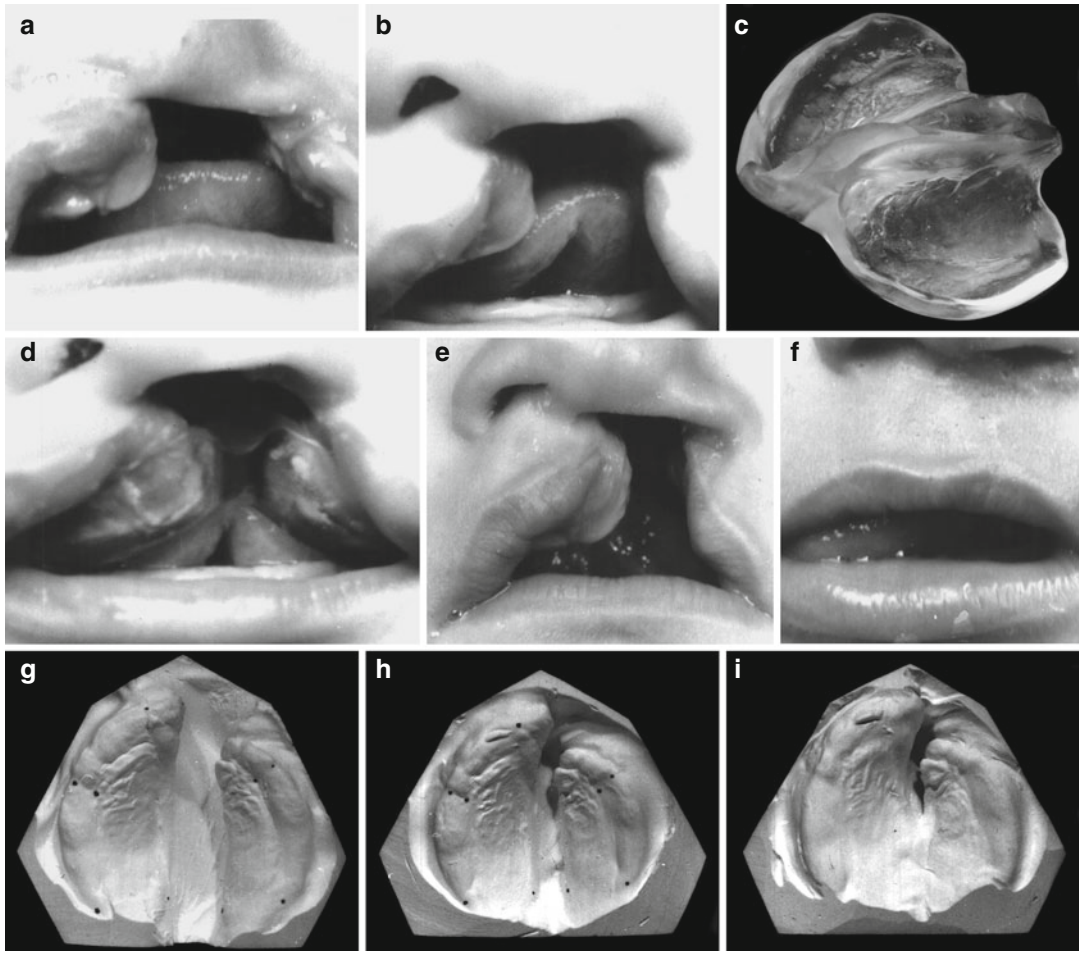

Fig. 5.3 (**a**–**i**) Presurgical orthopedic treatment (*PSOT*) appliance for a CUCLP utilized from birth to 1 year and 11 months at the University of Nijmegen (Courtesy of AM Kuijpers-Jagtman). (**a**) Lip and nose distortion at birth; (**b**) tongue posture within the cleft; (**c**) orthopedic appliance; (**d**) orthopedic plate prevents the tongue from entering the cleft; (**e**) 15 weeks after PSOT and before lip closure; (**f**) 6 weeks after palate closure; (**g**) 17 months before soft palate closure; (**h**) at 14 months of age, before soft palate closure; (**i**) 8 weeks after lip closure

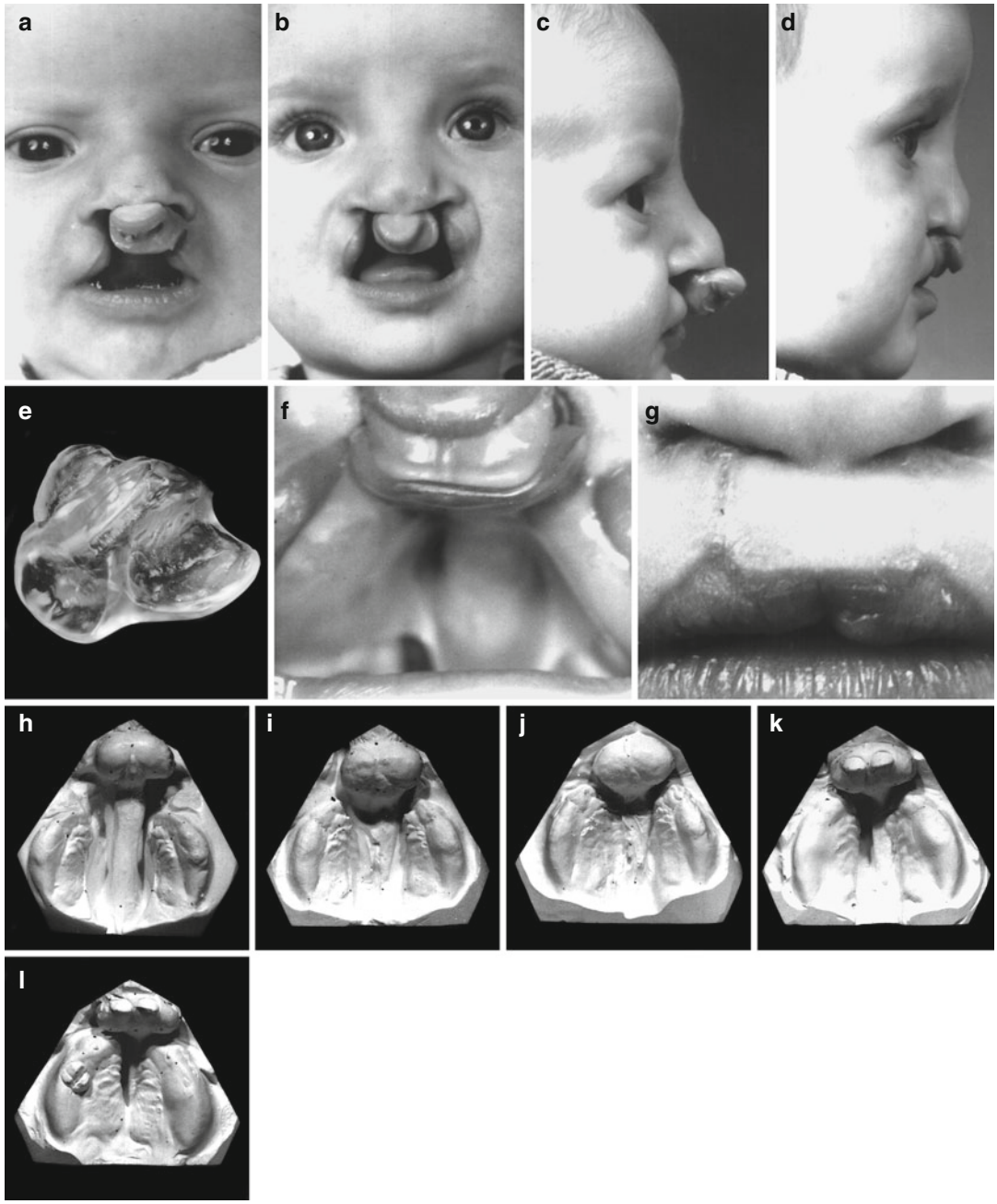

Fig. 5.4 (**a–l**) Presurgical orthopedic treatment from birth to 1 year for a CBCLP at the University of Nijmegen (Courtesy of AM Kuijpers-Jagtman). Lip closure at 1 year of age. Hard palatal cleft is closed between 6 and 9 years of age together with bone grafting of the alveolar cleft. (**a–c**) Facial photographs and palatal cast at birth; (**d**) 6 months after wearing PSOT appliance; (**e**) presurgical orthopedic appliance and when placed on the palate; (**f**) wearing appliance; (**g**) 8 weeks after lip closure; (**h**) at birth, (**i**) after 6 months of PSOT and before lip closure; (**j**) 8 weeks after lip closure; (**k**) 1 year and 6 months, before soft palate closure; (**l**) 6 weeks after soft palate closure

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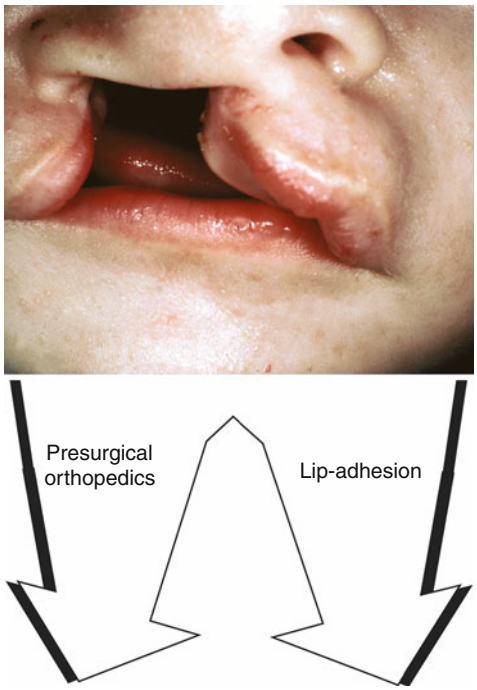
Complete Unilateral Cleft of the Lip and Palate

6

Samuel Berkowitz



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As previously described (Pruzansky 1955), in complete unilateral and bilateral clefts of the lip and palate, after the lip is united, the overexpanded palatal segments move together, reducing the cleft width along its entire posterior length. Subtelny (1955), using laminographs, has shown that newborns with complete clefts of the lip and palate have wider than normal pharyngeal widths and the perpendicular plates of the sphenoids are distorted in their relationship. Aduss and Pruzansky (1967) have demonstrated that, in complete unilateral cleft lip and palate, any one of three arch forms can result after the lip is repaired (Fig. 6.1):

1. The alveolar segments can move into end-to-end contact, producing a symmetrical arch form.
2. The alveolar segments can overlap, producing what is erroneously known as a "collapsed" arch form.
3. The alveolar segments can move closer together but not make contact. This occurs because of an inhibiting factor of the inferior turbinate on the cleft side, making contact with the distorted bulge of the nasal septum.

In a series of 58 patients who had no presurgical orthopedics or primary bone grafting, Aduss and Pruzansky (1967) found that approximately 43 % had overlap of the alveolar processes (mistakenly called collapsed arch). Among these patients, crossbites of the canine and first deciduous molar were the most common finding at 5 years of age. There were no anterior crossbites. Other investigators have reported similar results (Bergland 1973;

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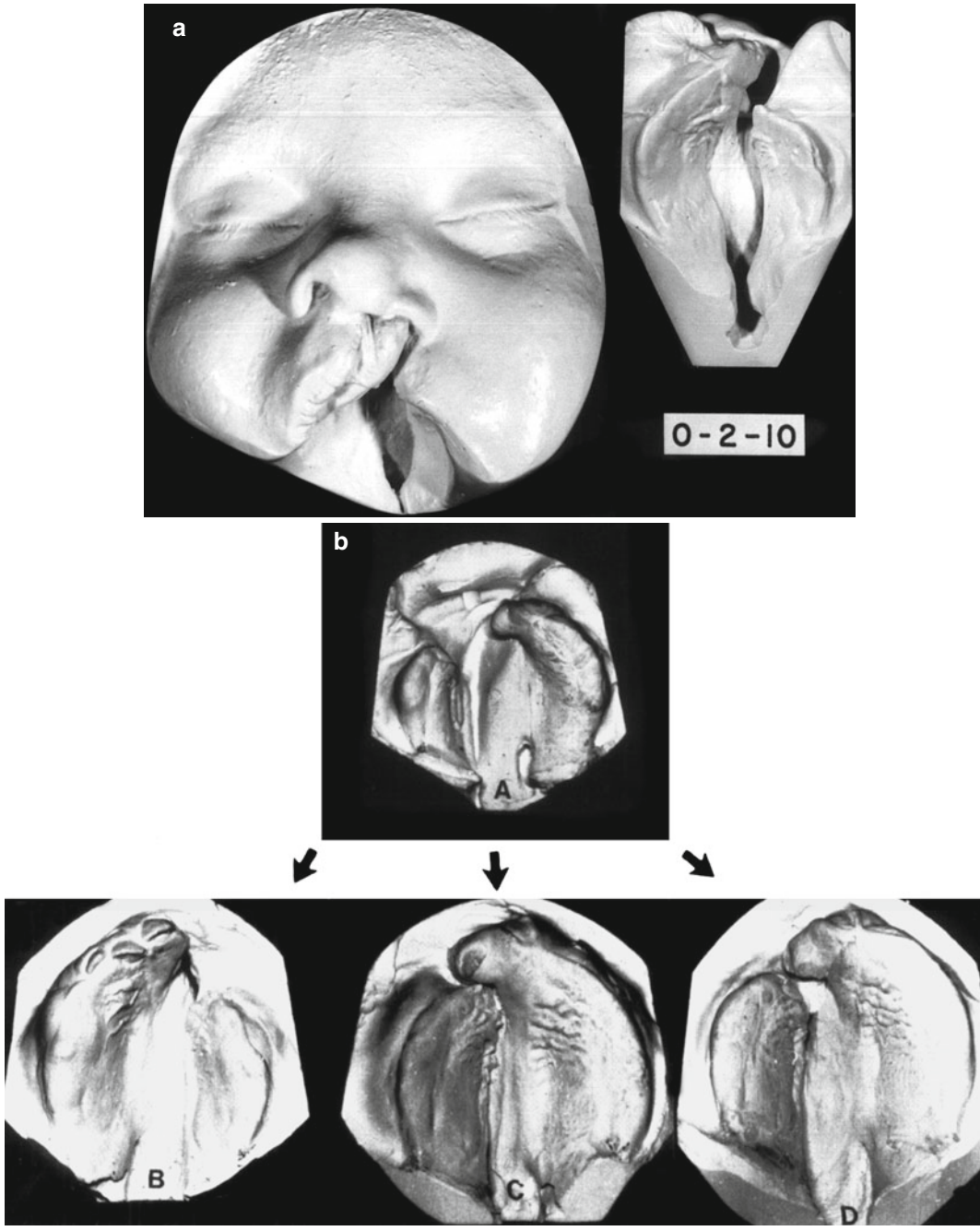


Fig. 6.1 (**a**) CUCLP. Facial and palatal casts. (**b**) Complete unilateral cleft lip and palate (CUCLP) before (*A*) and after (*B*) lip surgery. With the establishment of muscle continuity, the lesser segment moves medially, while the premaxillary portion of the larger segment moves medio-inferiorly, both acting to reduce the cleft width. Any of the following segmental relationships can result. (*B*) No contact between segments. The inferior turbinate on the cleft side makes premature contact with the bowed nasal septum. (*C*) The premaxillary portion of the larger segment overlaps the smaller segment. (*D*) The segments form a butt joint showing good approximation. Pruzansky and Aduss have shown that there is no correlation between the original cleft width and the resultant arch form. Wider clefts seemed to demonstrate less of a tendency toward collapse than did the narrower clefts (Aduss and Pruzansky 1967)

Bergland and Sidhu 1974). Berkowitz (1985), in a serial study of 36 cases with complete unilateral clefts of the lip and palate in which the lip had been united between the ages of 3 and 5 months and the palatal cleft closed between 18 and 24 months using a von Langenbeck with modified vomer flap without neonatal maxillary orthopedics, showed that 5 of the 36 cases had a complete buccal crossbite which was corrected within 6–10 months with fixed palatal expanders. Cuspid crossbite was the most frequent occurrence and was due to angular palatal rotation as well as to ectopic eruption of the deciduous cuspids. The cleft and noncleft segments were in either a class I or class II occlusal relationship. In no instance were any of the segments in a class III relationship.

This confirms Berkowitz's belief that the cleft palatal segment is not retropositioned within the skull relative to the mandible and that the maxillary-mandibular relationship is similar to that seen in the noncleft population. Whether the maxilla and mandible are both posteriorly positioned within skull has not been determined, and others (Semb 1991; Ross 1987a, b, c) have found this to be the case. Therefore, the palatal segments do not need to be brought forward by the use of neonatal protraction forces as Latham (1980) and his mentor McNeil (1950) have suggested. There are, however, some cases when, because of an unfavorable facial growth pattern coupled with a retruded maxilla relative to the anterior cranial bases, orthopedic protraction forces will be beneficial in the mixed (transitional) and permanent dentition.

According to Aduss and Pruzansky (1967), four factors govern arch form:

1. The size and shape of the alveolar process adjacent to the cleft. A bulbous and fully toothed alveolar process acts as an impediment to the collapse of the arch, whereas a thinly formed and dentally impoverished alveolar process leads to the overlapping of segments.
2. The size and shape of the inferior turbinate on the side of the cleft. A thick, rounded, well-modeled inferior turbinate can block excessive medial movement of the palatal segments.
3. The size and geometrical inclination of the nasal septum. A highly inclined septum with a contiguous bulbous turbinate will affect the movement of the palate and its final position.
4. The size and shape of the palatal shelves. Shelves of disproportionate size are more prone to overlap. One can certainly visualize that a long noncleft segment coupled with a short cleft segment will end up with the premaxillary portion overlapping the short cleft palate segment.

6.1 Facial Characteristics

Aduss (1971) in 1971 examined 50 males and 21 females with UCLP; their age range was between 4 and 14 years. He described craniofacial growth in the male cleft group as essentially equivalent to the female cleft group. He found that the gonial angle for the cleft patients was consistently larger than the noncleft group and the mandible appeared to be more retrognathic. He concluded that the craniofacial complex in the cleft sample tended to grow in a similar manner to that reported for the noncleft populations. The results of his study, based on a conservative method of surgery, negate the conclusions reached at the time regarding the deleterious effects of surgery on the growth of midface.

Hayashi et al. (1976) studied craniofacial growth in unilateral complete clefts using lateral cephalograms of 135 males and 120 females with an age range of 4–18 years. Control subjects included 120 noncleft males and 120 noncleft females of similar age to the cleft subjects. They concluded that the cleft group differed from the control group in several major respects: (1) Their overall growth trend showed a more downward or vertical direction; (2) the cranial base angle was more flattened; (3) the maxilla was smaller and was located in a more posterior and upward position; (4) ramal height was shorter, the gonial angle was more obtuse, and mandible was generally retrognathic; (5) upper face height was smaller and lower face height was greater; (6) underdevelopment in both the maxilla and the mandible was more pronounced in cleft females than in cleft males.

Smahel and Mullerova (1986), in 1986, studied 30 boys with UCLP prior to palatoplasty using cephalometry. A comparison with 27 normal individuals matched in age showed that most basic deviations of the craniofacial configuration recorded in adults developed at an early age, often prior to palatoplasty, i.e., reduced height of the upper anterior face, maxillary dentoalveolar retroclination, displacement of the upper jaw backwards, widening of some components of the maxillary complex, and a shortening of the mandibular body and ramus. Only the length of the upper jaw was not reduced. The shortening of maxillary dimension occurred postoperatively at a more mature age.

Later on 1992, Smahel et al. (1992) presented another study of craniofacial morphology in UCLP in 58 adult males. The results showed a shortening of maxillary depth, reduction of the upper face height, increased lower anterior facial height, and mandibular changes resulting from growth deficiency that consisted of shortening of the body and ramus, obtuse gonial angle, steep mandibular plane, and retrognathia.

Again in 1992, Smahel et al. (1992) studied growth and development of the face in UCLP during prepubertal and pubertal periods. He concluded that there were no definitive differences in the growth rate between the pre- and postpubertal periods. Therefore, the worsening of overjet during puberty could be due to the depletion of the compensation and adaptation after the previous orthodontic treatment rather than to the enhanced growth rate. In addition, he found that during the prepubertal period, the lower jaw showed a very slight posterior rotation, while during puberty, an anterior growth rotation was present. A marked retrusion of the maxilla developed already in the prepubertal period. During both periods, there occurred an identical impairment of sagittal jaw relations and of the upper lip prominence, accompanied by a flattening of the facial profile and reduction of the nasolabial angle.

In 1996, Smahel and Mullerova (1996) reported a longitudinal study regarding postpubertal growth and development of the face in UCLP as compared to the pubertal period. The data showed that in boys, facial growth persists after the age of 15 years and maxillary growth attains almost half the values recorded in the period of puberty, while mandibular growth attains almost the same values as during puberty. In girls, the growth is almost terminated except for the lower jaw, where it is still significant though several times slighter than during puberty. Due to the gender differences in the amount of postpubertal growth, developmental changes in facial configuration do not occur in girls during this period, while in boys, there is a further deterioration of maxillary protrusion, sagittal jaw relations, and flattening of the face.

In 1988, Hoswell and Levant (1988) reported another long-term follow-up of skeletal growth of UCLP subjects ranging in age from 8 to 18 years. Serial cephalographs taken every 2 years were utilized for determination of six cephalometric dimensions: anterior cranial base, upper and lower facial heights, posterior nasomaxillary height, maxillary horizontal length, and mandibular length. These were compared to published cephalometric standards of a noncleft group. All dimensions except mandibular length were smaller in the UCLP group. The horizontal maxillary length appeared to be most affected in UCLP. Mandibular length was not affected in the cleft group.

6.1.1 The Oslo Study

Because of the stable and long history of meticulous record keeping and protocols that characterizes the data acquisition of the Oslo team, the following studies on unilateral cleft lip and palate are presented to provide a unique perspective on treatment strategies and facial growth standards based on longitudinal data. The author does not follow the same surgical strategies as those of the Oslo team but recognizes that the differences are not significant enough to interfere with obtaining a successful long-term outcome.

Semb's (1991) 20-year serial cephalometric study taken from the Oslo Archives gathered during Bergland's leadership involved 76 males and 81 females (157 individuals) who did not have neonatal maxillary orthopedics. All of the

children in the study had lip closure in infancy using a modified Le Mesurier or, after 1969, a Millard procedure. During the same operation, the nasal floor was closed using a one-layer vomer flap. The remaining posterior palatal cleft was closed between 4 and 5 years of age using a von Langenbeck palatoplasty. Secondary alveolar bone grafts from the iliac crest were placed at between 8 and 11 years of age. By 1974, all palate repairs were completed by 18 months of age. Superior-based pharyngeal flap surgery for velopharyngeal insufficiency was performed in about 20 % of the cases.

Compared with normal males and females, the pooled sample with unilateral cleft lip and palate showed (1) skeletal and soft tissue maxillary retrusion, (2) elongation of the anterior face (even though the upper face height was shorter), (3) a retrusive mandible, (4) reduction in posterior face height, and (5) a slight increase in the angle of the cranial base.

The pattern of growth also was different from that of noncleft individuals. Between 5 and 18 years of age, there was almost no increase in the length of the maxilla. There was a marked reduction in maxillary and mandibular prominence. Vertically, the excessive lower face angulations changed slightly.

6.1.2 Multicenter CUCLP Cephaloradiographic Study (Ross 1987a, b, c)

Ross's multicenter study involved data from 15 cleft palate centers around the world collected for the purpose of determining the effects of manipulative and surgical treatment on facial growth. A sample of 1,600 cephalometric radiographs of males with complete unilateral cleft lip and palate were traced, digitized, and analyzed in the Craniofacial Center of the Hospital for Sick Children. The seven series of studies considered virtually every aspect of treatment that might influence facial growth.

Ross concluded that the type of surgical repair used does not make an appreciable difference to facial growth. It appears, however, that there are differences that can only be explained on the assumption that some surgeons induce less growth inhibition than others. Variation of the timing of hard and soft palate repair within the first decade does not influence facial growth in the anteroposterior or vertical dimension. Ross admits that very early soft palate repair was not well represented in this study, and there is some suspicion that there might be untoward results.

Berkowitz et al. (2005) clinical findings suggest a different conclusion that, in most cases, early surgery (before 12 months) will have a negative effect on palatal growth in all three dimensions. It all depends on the size of the cleft defect relative to the area of the surrounding mucoperiosteum (see Chap. 7).

Ross's study did not include palatal surgery from 6 to 12 months. This study also reported that the resulting face is flat in profile and decreased in depth, with a vertical deficiency in the midface and vertical excess in the lower face. The mandibles in these faces characteristically are slightly shorter in total length so that the chin is retruded. The occlusion is more of a molar and incisor mesiocclusion in clefts with less overbite and overjet. The soft palate in this sample is appreciably more posterior. The mandibular plane angle is greater, possible due to the need for more interincisal space.

Ross further stated that the bony pharynx was unaffected by treatment and that the variation in midface development can be attributed to maxillary length rather than to maxillary position. He also noted that the mandible is not directly affected by treatment. Facial growth is intrinsically compromised by an underlying deficit, and surgery acts to further interfere with growth of the midface by inhibiting forward translation.

The best results appear to follow lip repair at 4–5 months with no repair of the alveolus. Early alveolar repair restricts its vertical growth and should be avoided in individuals with poor growth potential. This leads to deficient midfacial height and poor vertical height proportions, with more acute nasolabial angles. There is no evidence that periosteoplasty will cause simi-

lar results. Berkowitz et al. (2005) study conclusively shows that periosteoplasty inhibits midfacial development, especially that of the premaxilla (see Chap. 10).

Ross further states that the maxilla in the UCLP is not more posteriorly positioned to any appreciable extent, but it is much shorter in length. The repaired lip affects the basal maxilla more than the alveolar process. Vertical development of the posterior maxilla is more deficient than the anterior part. The mandible is shorter with a steeper mandibular plane angle.

Hard and soft palate surgical repair procedures provide the greatest potential for inhibiting the maxilla in length, forward translation, and posterior height.

Kwon's (1998) retrospective longitudinal study of the skeleto-facial growth in unilateral cleft lip and palate documented and evaluated the proportional craniofacial growth horizontally and vertically in 14 UCLP patients of the ages 5–18 years by using modified Coben's basion horizontal analysis. There were three populations included in this study: The Eastman cleft group (sample size, 24) and the Miami cleft group (sample size 23) served as patient group, and the Bolton templates (ages 5–18) served as controls. Samples were divided into four age periods according to the chronological ages, and then the growth pattern of each period were evaluated and compared. A total of 301 images of lateral cephalograms were examined and digitized. These characteristics of the skeletal facial growth of the UCLP are summarized as: (1) There is no difference of posterior cranial base over time; (2) maxilla is positioned posteriorly relative to basion (BA) during the early ages and is getting retrusive due to the deficient growth with time; (3) upper anterior facial height (UAFH) is almost the same as the control; (4) lower posterior facial height (LPFH) is increased but is not as much as lower anterior facial height (LAFH); (5) lower anterior facial height (LAFH) is significantly increased; (6) total facial height (TFH) is significantly increased; (7) mandible is positioned backward and downward due to the posterior position of the maxilla and the elongation of LPFH and LAFH; and (8) skeletal profile is more convex and is getting straight and finally is flatter over time in the clefts than in the controls. Generally, the manifestation of the cleft characteristics of the Miami group is increased when compared to that of the Eastman cleft group. The skeletal growth leads to not only the maxillary retrusion but also to position the mandible down and back. Early orthopedic intervention followed by the fixed edgewise appliance and prolonged retention is recommended to try to correct the skeletal problems, camouflage by dental correction, and maintain to the treatment outcome with reasonable retainer.

6.1.3 Reflection on Ross' Excellent Multicenter Study (Ross 1987a, b, c)

In the foreword of the multicenter study, Treatment Variables Affecting Facial Growth in Complete Unilateral Cleft Lip and Palate, Bruce Ross discussed the difficulty of performing this type of study due to the variability in sample size, age, sex, precise cleft type, and ethnic origin. He then mentioned the problems associated with doing cephalometric measurements and suggested using one center to control measurement errors; this was an excellent solution. According to Ross (1987a, b, c), the study considered virtually every aspect of treatment that might influence facial growth. An attempt was made to control many variables that influence growth research, so that a clear picture of the effects of each procedure would be available. Two major assumptions about the study are necessary if any conclusion can be drawn from these studies. The first is that all groups of infants with complete unilateral cleft lip and palate have exactly the same facial morphology at birth in spite of enormous individual variation within the group. The second assumption is that one group of infants will respond on the average in exactly the same way as any other group to a particular treatment. The intent was to assemble relatively pure samples of individuals who had received the given management techniques used consistently on all subjects from a particular center.

Berkowitz believes the study was a noble attempt by excellent clinicians/researchers to pool their sample cases to investigate treatment results. By necessity, it was limited to cephalometric records. By lumping all CUCLP cases together, regardless of the degree of palatal deformity at birth, much potential prognostic information for the treatment of individual cases is unavailable. Ross had no choice for discounting Slavkin's and Ross and Johnston's statements that palatal defects may be caused by either the failure of the separated palatal segments to fuse or, possibly, palatal osteogenic deficiency, a variable that needs to be considered in treatment planning. This statement on the embryo-pathogenesis of cleft palate explains why all clefts within a cleft type are not alike. It is not hard to reason that as the extent of the cleft palatal defect varies, so will the resulting quantity of palatal surface area and the resulting quantity of post-surgical scar tissue. Because excessive scarring inhibits palatal growth and development, the palatal surface area at the time of closure needs to be considered in treatment planning. Berkowitz believes that the variability of palatal surface area within a particular cleft type weakens the value of Ross's (1987a, b, c) conclusions, which are based on the second assumption that "one group of infants will respond on the average in exactly the same way as any other group to a particular treatment." Berkowitz concludes that the next level of treatment evaluation studies designed to improve differential diagnosis requires the establishment of specific criteria based on quantitative and qualitative characteristics of the palatal defect when related to treatment outcome (see Chap. 7).

6.2 How the Palate Grows

As ready discussed in Chap. 1 on facial growth, bone growth involves the increase in size as well as remodeling. Serial palatal three-dimensional growth studies by Berkowitz et al. (2005) have shown that growth and remodeling occurs over the entire palatal surface even at the medial border the palate at the cleft (Figs. 6.2 and 6.3).

6.3 Treatment Sequence

6.3.1 Usual Treatment Sequence

1. Lip adhesion: 3 months
2. Definitive lip surgery (rotation advancement): 10 months
3. Hard and soft palate closure (von Langenbeck with vomer flap): 18–24 months (rarely 36 months)
4. Orthodontic expansion (quad helix): 5–7 years
5. Superior-based pharyngeal flap: 6–8 years if necessary
6. Bone graft (iliac crest): 7–9 years
7. Protraction facial mask (if necessary): 8 years or later
8. Maxillary surgical advancement (Le Fort I or distraction osteogenesis): varies
9. Lip/nose revisions techniques

6.4 Reports

In this section, treatment outcomes of selected cases are presented with photographs and dental casts. The casts started at birth and continued through adolescence; these records show the natural history of palatal and facial growth and development when conservative surgery was performed without the use of presurgical orthopedics.

In some cases, the lip was united after the use of a Logan's bow (Fig. 6.4), and in others, after lip adhesion at approximately 3 months of age. In some cases, the cleft of the soft palate was united at the same time the hard palate was closed. Definitive lip surgery was performed at 6 months, and hard palate closure using a modified von Langenbeck procedure with a vomer flap was performed between 12 and 24 months of age.

Selected cases are presented in Figs. 6.5, 6.6, 6.7, 6.8, 6.9, 6.10, 6.11, 6.12, 6.13, 6.14, 6.15, 6.16, 6.17, 6.18, 6.19, 6.20, 6.21, 6.22, 6.23, 6.24, and 6.25 to show various treatment solutions to complex problems.

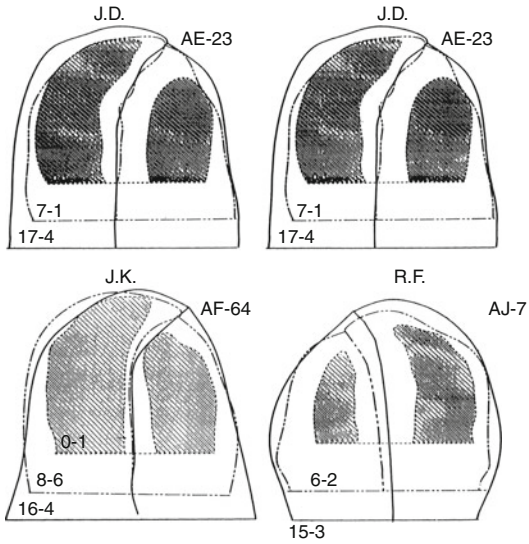


Fig. 6.2 (**a**, **b**) Superimposed computer-generated images of serial CUCLP casts superimposed on the rugae and registered on the vomer AP line. The alveolar ridge is the outer limits of the palate. Surgery: Lip adhesion at approximately 3 months, definitive lip surgery at approximately 6 months, and hard and soft palate closure between 18 and 24 months using a von Langenbeck procedure with a vomer flap. No presurgical orthopedics. Results: These four illustrations show the result of molding and growth. The least growth occurs anteriorly. Most of the growth occurs posteriorly to accommodate the developing deciduous and permanent molars. The palatal mucoperiosteum covers increase palatal size and the palatal cleft, which greatly reduced in size

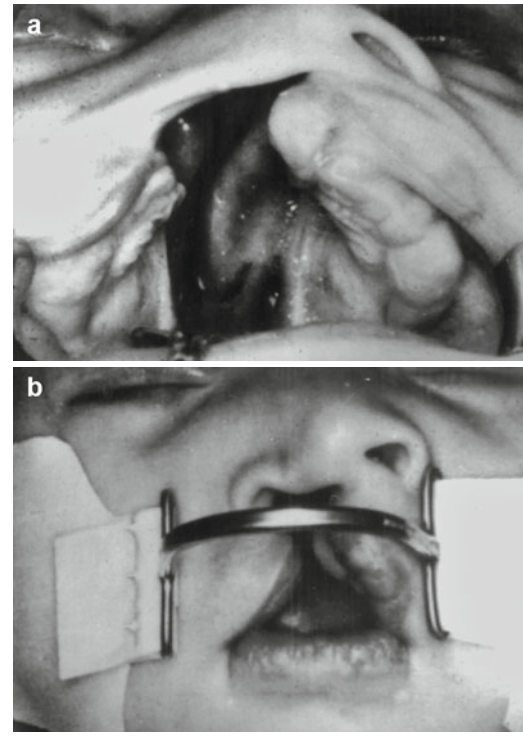
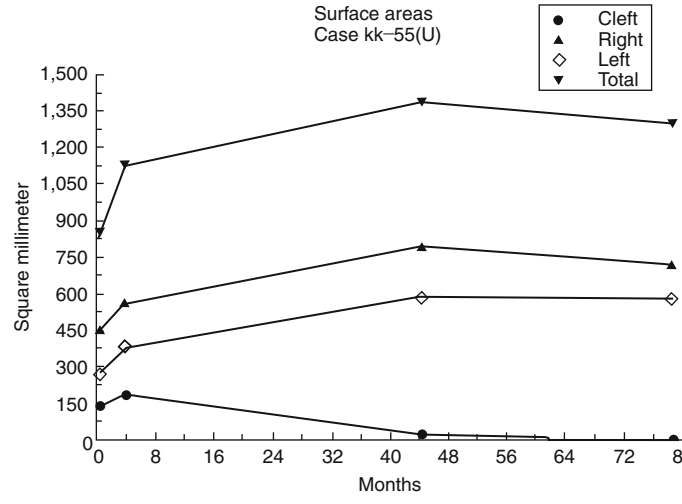


Fig. 6.4 (**a**, **b**) Logan's bow. Pressure is placed on the cheeks to bring the lips together prior to surgery. The bow helps to reduce tension at the suture line

Fig. 6.3 Case KK-55. Serial growth of the palatal segments in CUCLP. Using computer-generated 3D images, the surface areas mesial to the alveolar ridges were analyzed using an electromechanical digitizer. The same surgery stated in Fig. 6.2 was used. Results: This case is an example of 60 cases analyzed in the study; it shows (1) both palatal segments grow at the same rate and (2) the most rapid period (velocity) of growth occurs during the first 18 months. Comments: Because the most rapid period of growth occurs between 8 and 24 months when cells are most active, it is best to postpone palatal surgery until a later age in order to not inhibit growth



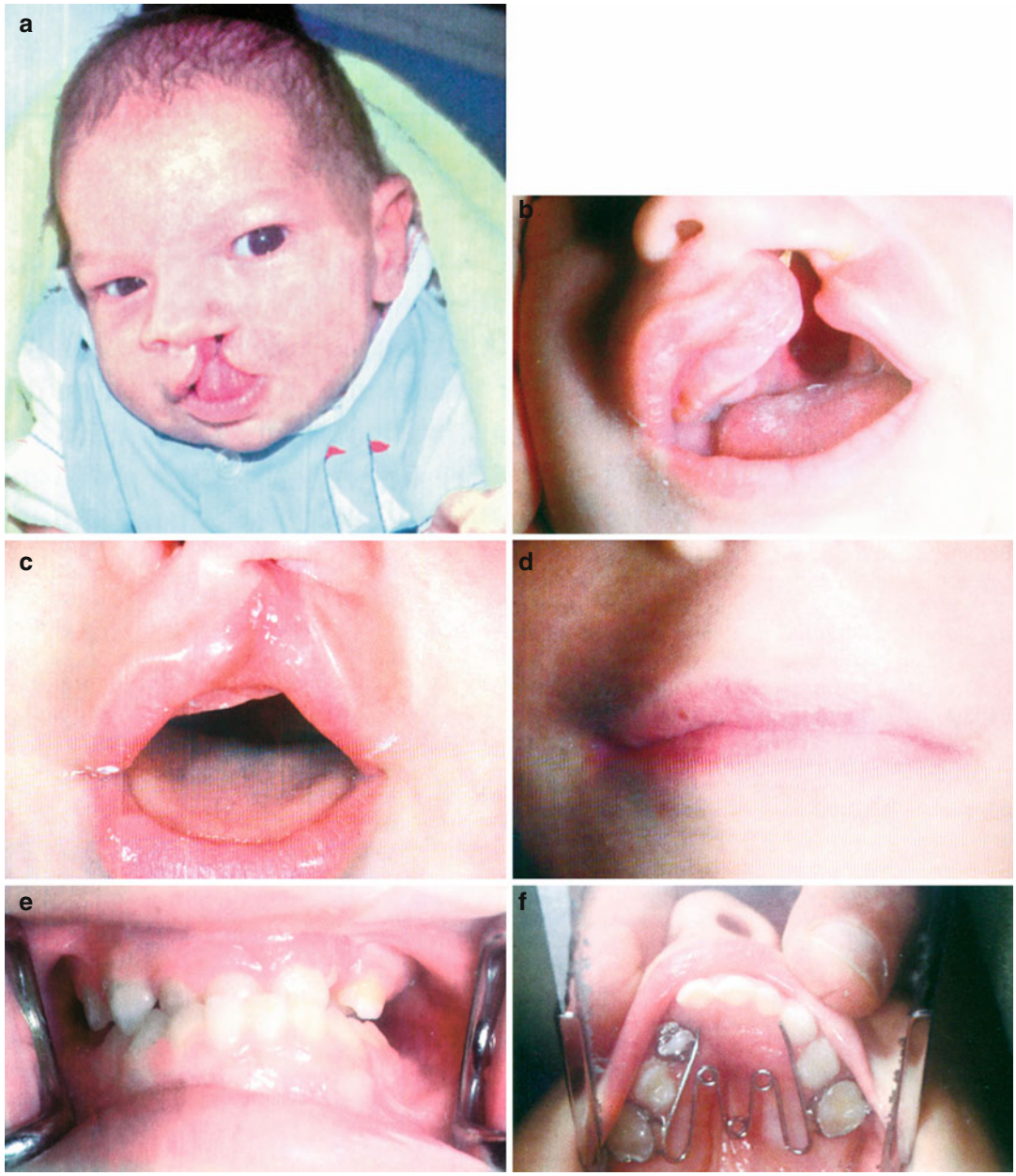


Fig. 6.5 (**a**–**v**) Case: KC (ZZ-1) demonstrates good palatal and facial growth in CUCLP. A very small cleft space at 5 months of age allowed for easy closure without much scar formation. Surgical treatment: No presurgical orthopedics. Lip adhesion followed by Millard's rotation advancement. Soft palatal closure at 2 months. Palatal cleft closure at 15 months using modified von Langenbeck procedure. Secondary alveolar cranial bone graft at 6 years and 8 months. Photographs showing various treatment stages from birth to 17 years of age. (**a**, **b**) Newborn. (**c**) Lip adhesion at 4 months. (**d**) Lip at 2 years of age. Orthodontics during the deciduous dentition. (**e**) 2 years, showing anterior crossbite. (**f**) 2 years, 7 months: palatal view showing fixed buccal expander

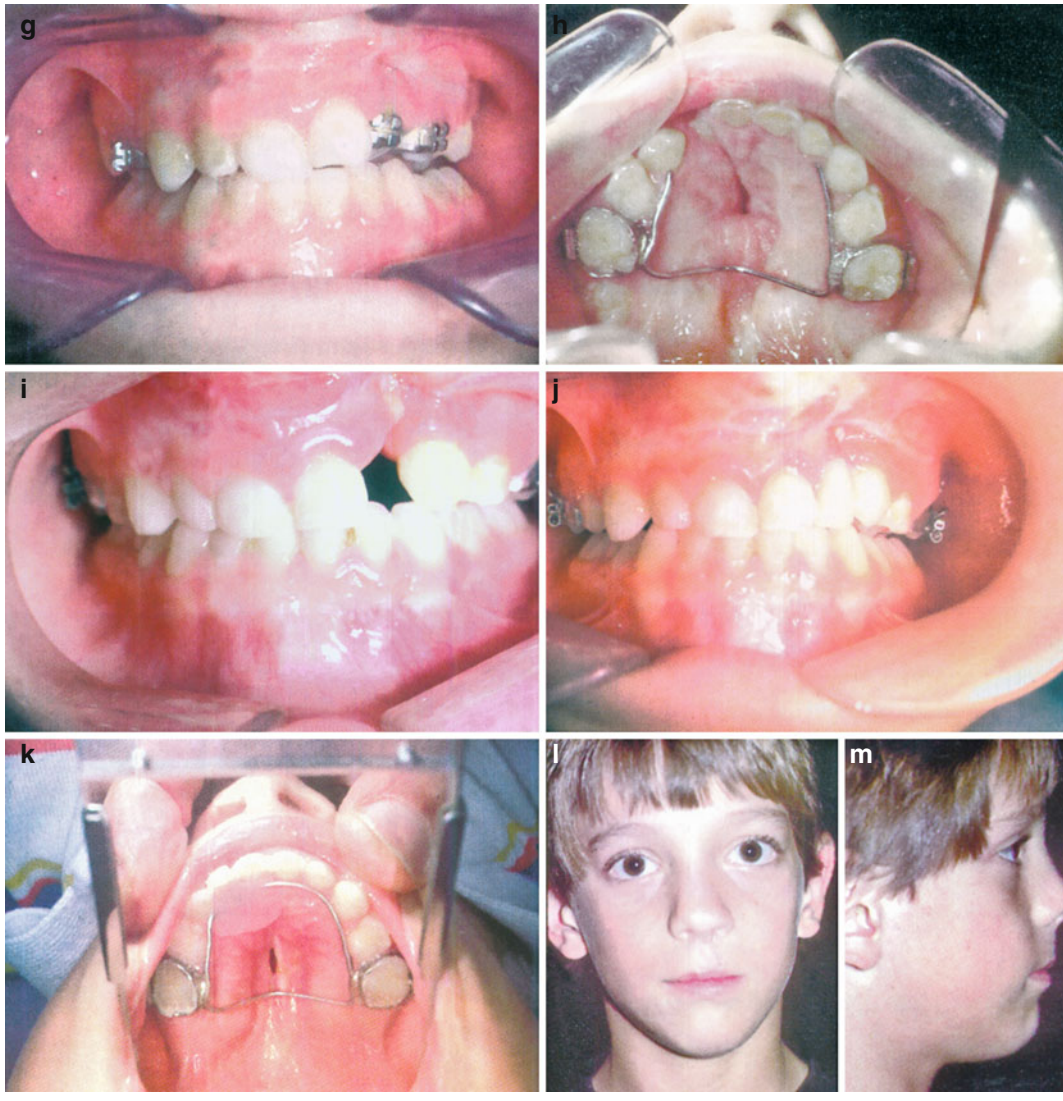


Fig. 6.5 (*continued*) (**g**) Anterior teeth were advanced and the cleft buccal segment expanded. (**h**) 5 years. Fixed palatal retainer. (**i**–**k**) Fixed palatal retainer with lateral incisor pontic (tooth). (**l**, **m**) Facial photographs at 6 years

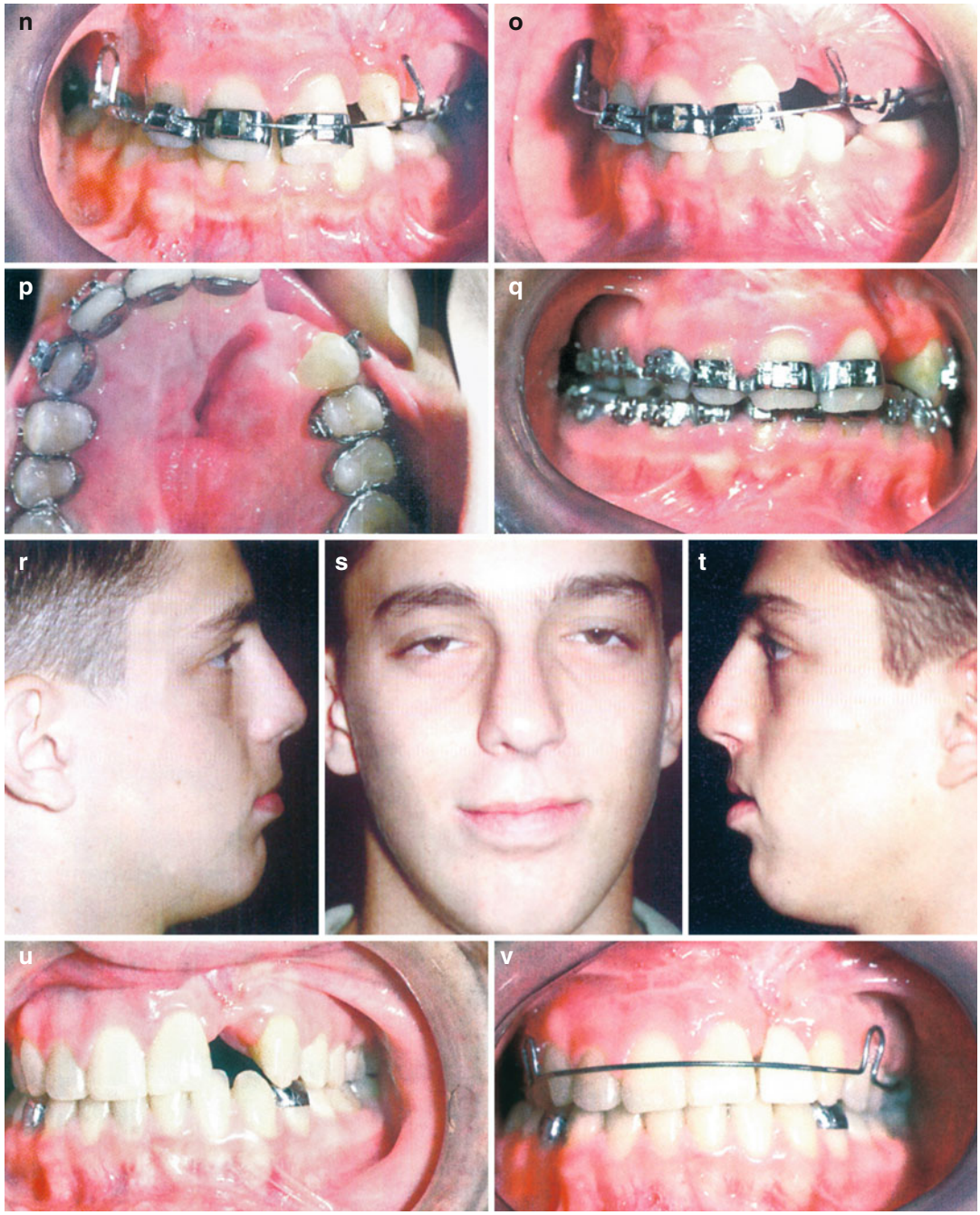


Fig. 6.5 (*continued*) (**n**, **o**) 7 years, 3 months: Lateral incisor is erupting through cranial bone graft. Orthodontics in the adult dentition: (**o**) Lateral incisor is extracted due to poor root development. (**p**, **q**) Conventional orthodontics. Surgery to close the palatal fistula was unsuccessful. (**r**–**t**) Facial photographs at 17 years. (**u**, **v**) Intraoral photographs. Hawley orthodontic retainer with lateral incisor pontic

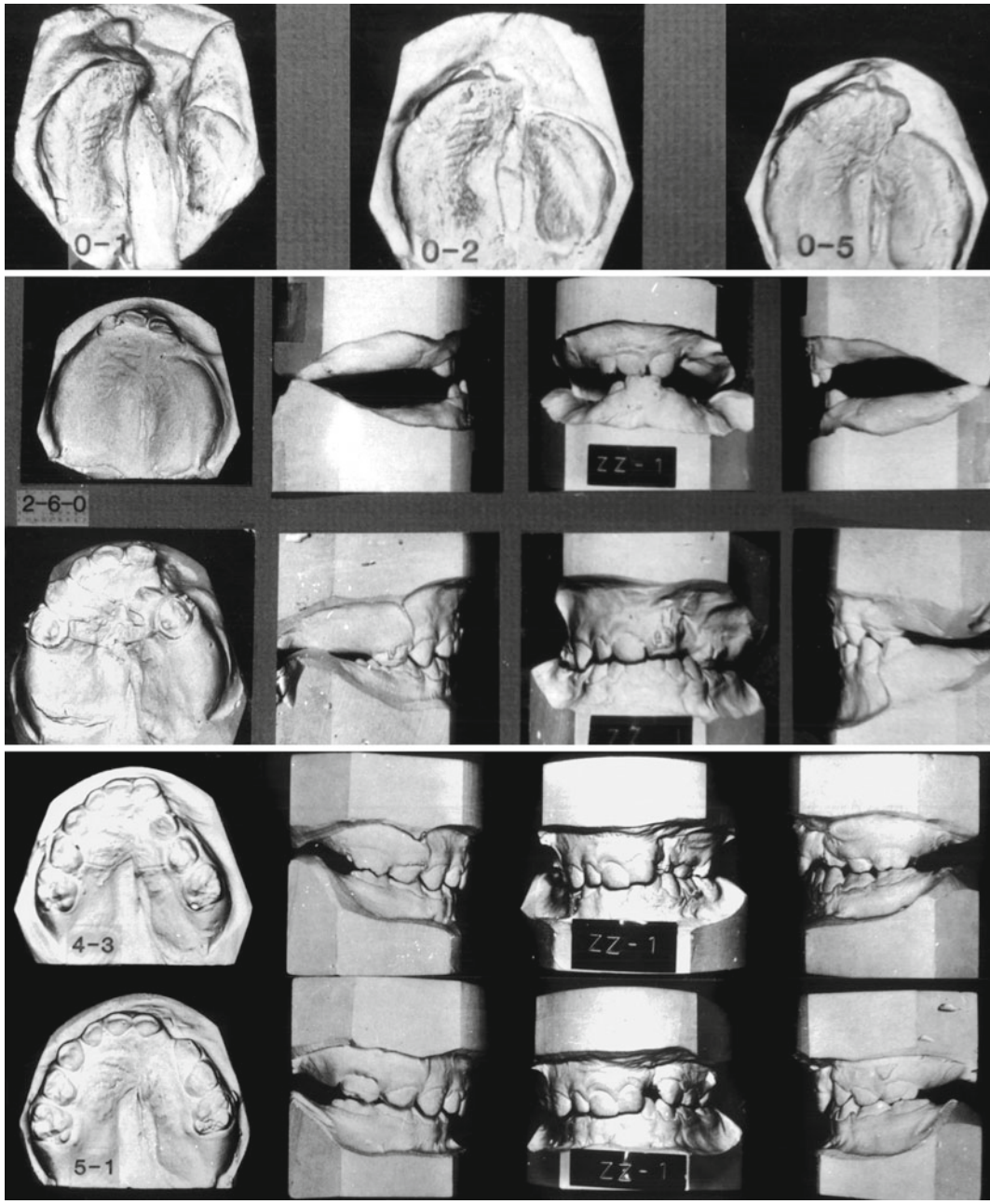


Fig. 6.6 Case KC (ZZ-1). Serial casts from 0–1 to 0–5 show medial movement and growth changes to the palatal segments. 0–5 The cleft space is extremely small with the palatal segments making contact anterior to the cleft space. 2–6–0 and 4–3 Mesioangular rotation of the lesser segment placed the deciduous cuspid in crossbite. 5–1 A fixed palatal expander rotated the segment outward, placing the teeth in ideal occlusion

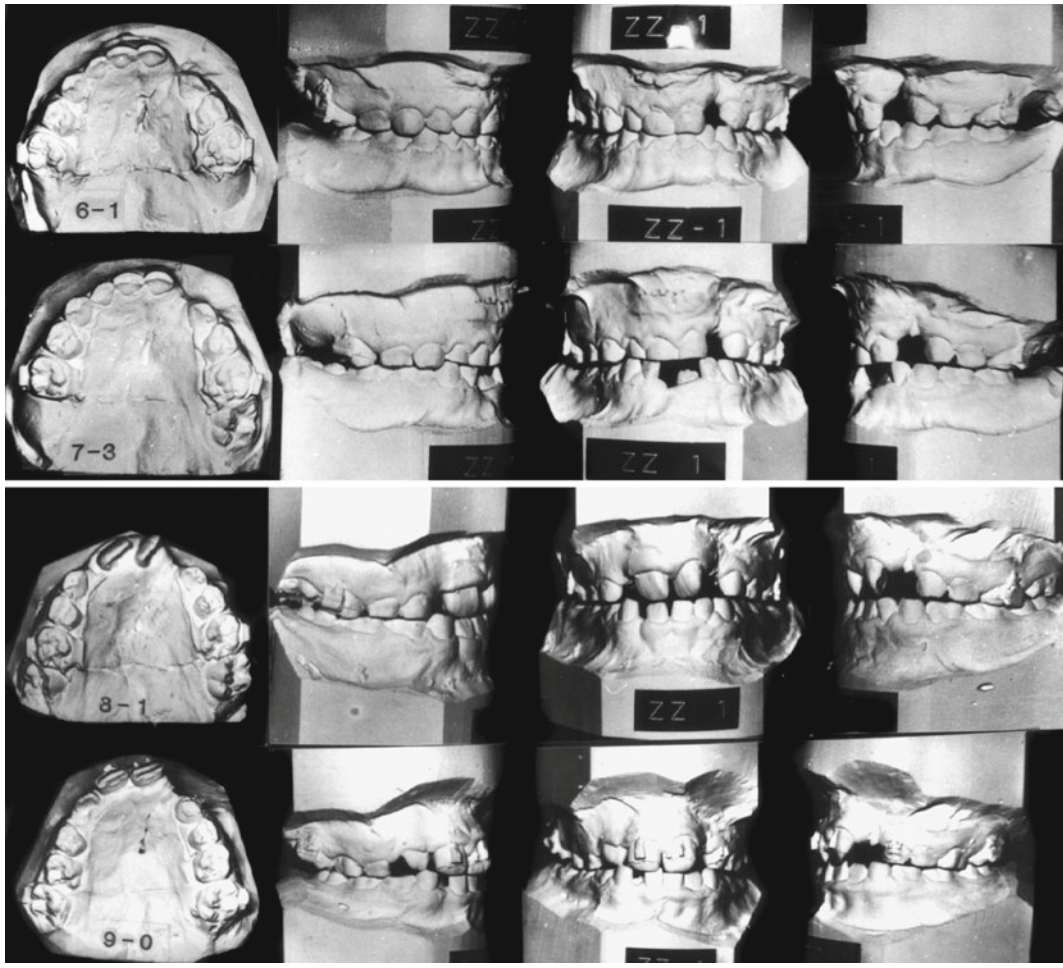


Fig. 6.6 (*continued*) 6–1 Fixed retainer maintained the correction. Secondary alveolar bone graft was performed at 7 years, 3 months of age. 9–0 The maxillary anterior teeth were rotated for aesthetic reasons

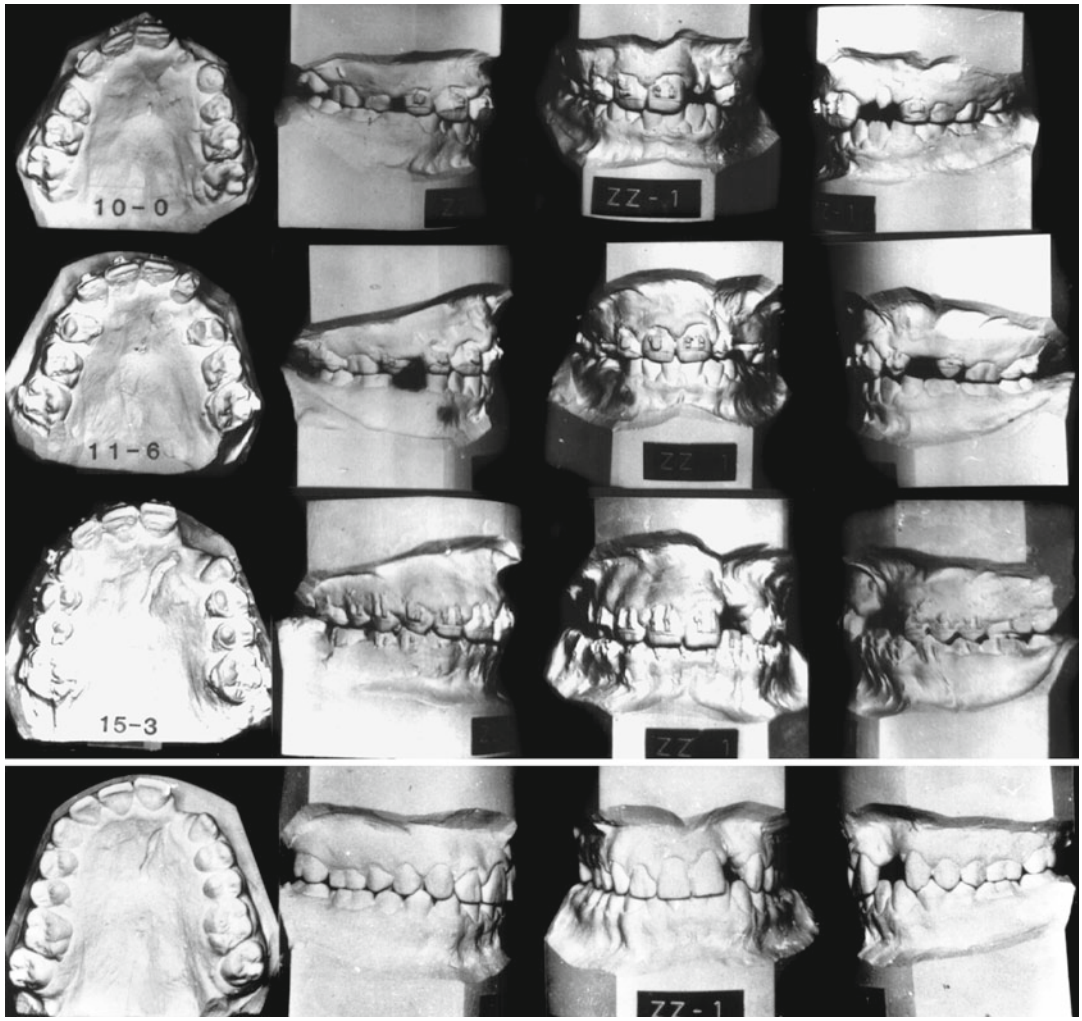


Fig. 6.6 (*continued*) 11–6 The left lateral incisor is now in place within the arch. As a result of poor root development, it had to be extracted. Conventional orthodontics was instituted and completed by 15–3. Maxillary fistula was surgically closed at 16–3, and the arch form maintained with a removable Hawley retainer with a lateral incisor pontic. 17–0 Final occlusion. Comment: Because most cleft palatal arches have some degree of osteogenic deficiency, when all bicuspids are retained, it is usual for the second molars to be blocked out and be impossible to position within the arch. This then necessitates their removal with possible replacement by the still unerupted third molars. In some instances, a small palatal fistula may not pose a speech problem or be a source of nasal drainage

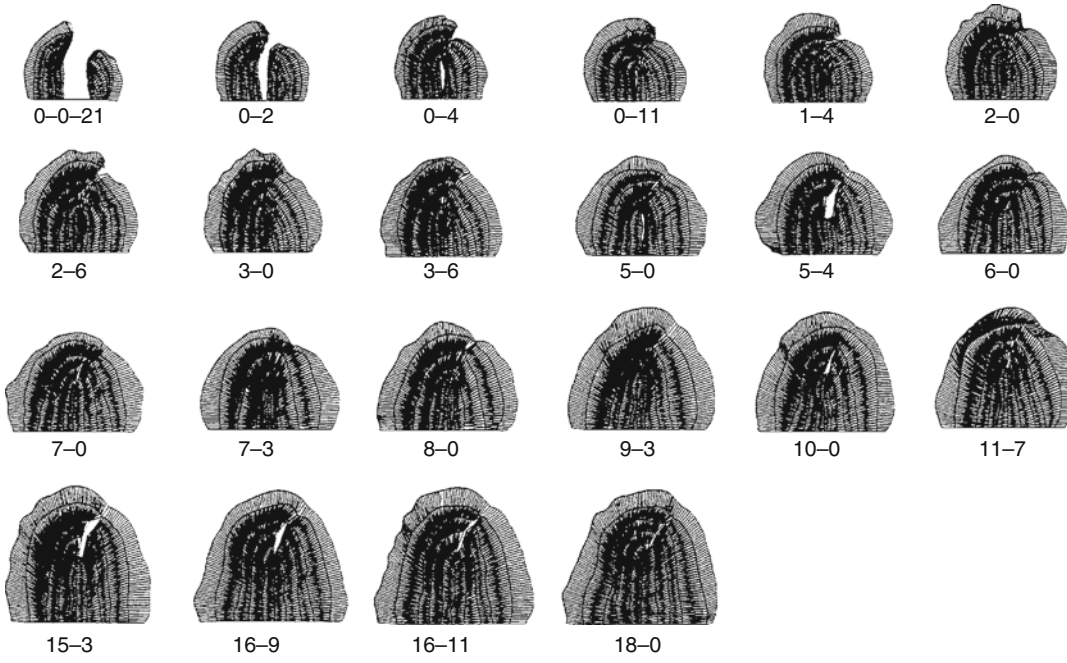


Fig. 6.7 Case KC (ZZ-1). Computer-generated drawings of serial casts which are in the same scale. The soft palate was united at 2 months and the hard palate closed at 15 months. This series demonstrates a rapid reduction in palatal cleft size with molding action and palatal growth. A palatal "fistula" was exposed when the cleft buccal segment was expanded to correct the crossbite. It was closed but reappeared when final orthopedic treatment moved the palatal segments slightly apart. The "fistula" did not penetrate into the nasal chamber. Therefore, it did not pose a speech or feeding problem

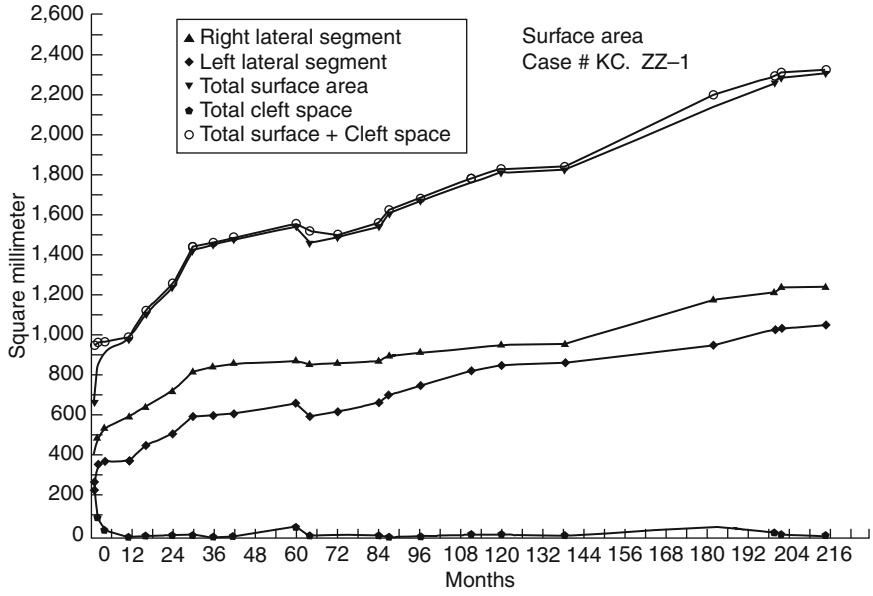


Fig. 6.8 Case KC (ZZ-1). The palatal growth chart shows (1) rapid growth acceleration in the first year which continues only slightly decreased until 36 months; (2) the palatal growth rate did not diminish after palatal surgery at 15 months; (3) palatal growth slowed between 60 and 84 months and then steadily increased; (4) between 60 and 120 months, the growth of the lesser cleft segment increased more rapidly than the noncleft segment; and (5) the palatal growth rate accelerated after 136 months. Comment: Based on palatal growth acceleration rates and the developing occlusion, one can safely conclude that palatal surgery did not interfere with its growth and development

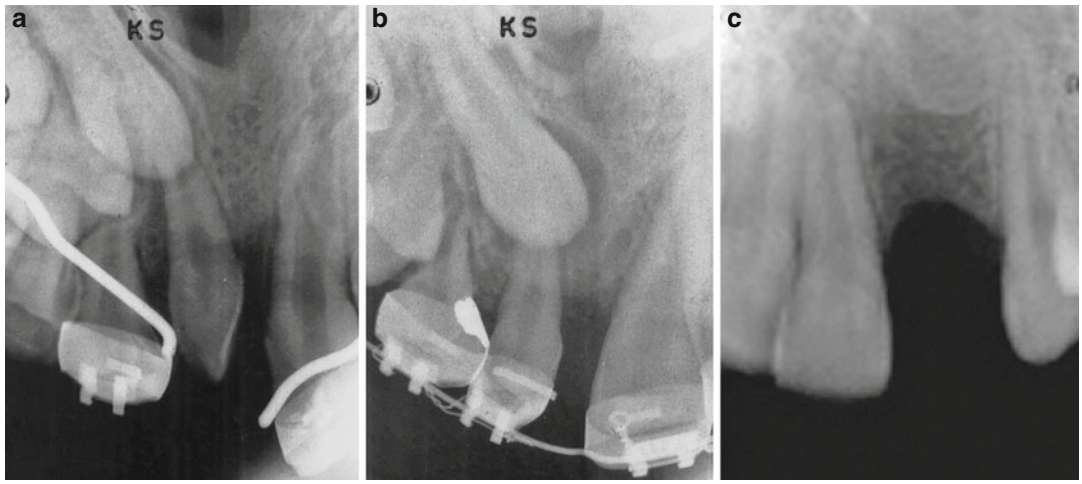


Fig. 6.9 (**a**–**c**) Case KC (ZZ-1). Tooth eruption into a secondary alveolar cranial bone graft performed at 7 years and 3 months of age. (**a**) The permanent lateral incisor is erupting into the graft. (**b**) Good root development, the lateral incisor, is brought into the arch orthodontically. (**c**) Its root began to absorb and was extracted, good alveolar bone in the cleft space

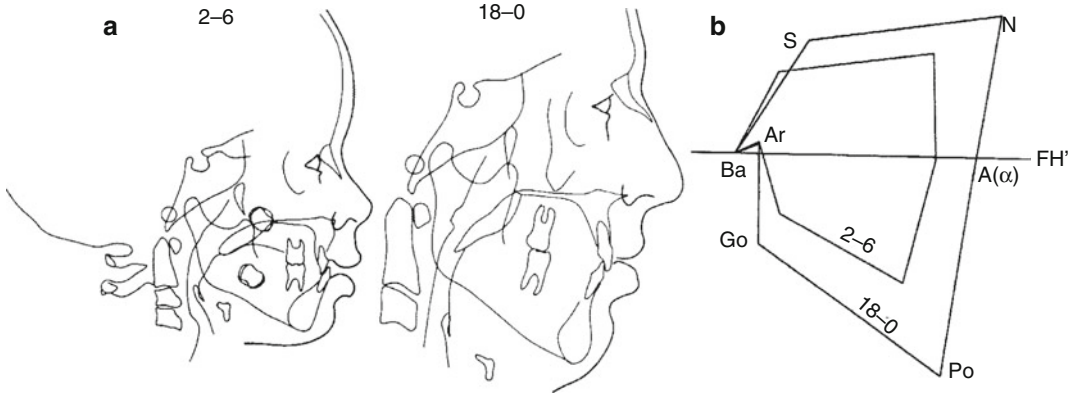


Fig. 6.10 Case KC (ZZ-1). (**a**) Lateral cephalometric tracings and (**b**) superimposed polygons using basion horizontal method (Coben). Both show an excellent facial growth pattern. At 18–0, the midface is slightly recessive but still very acceptable aesthetically

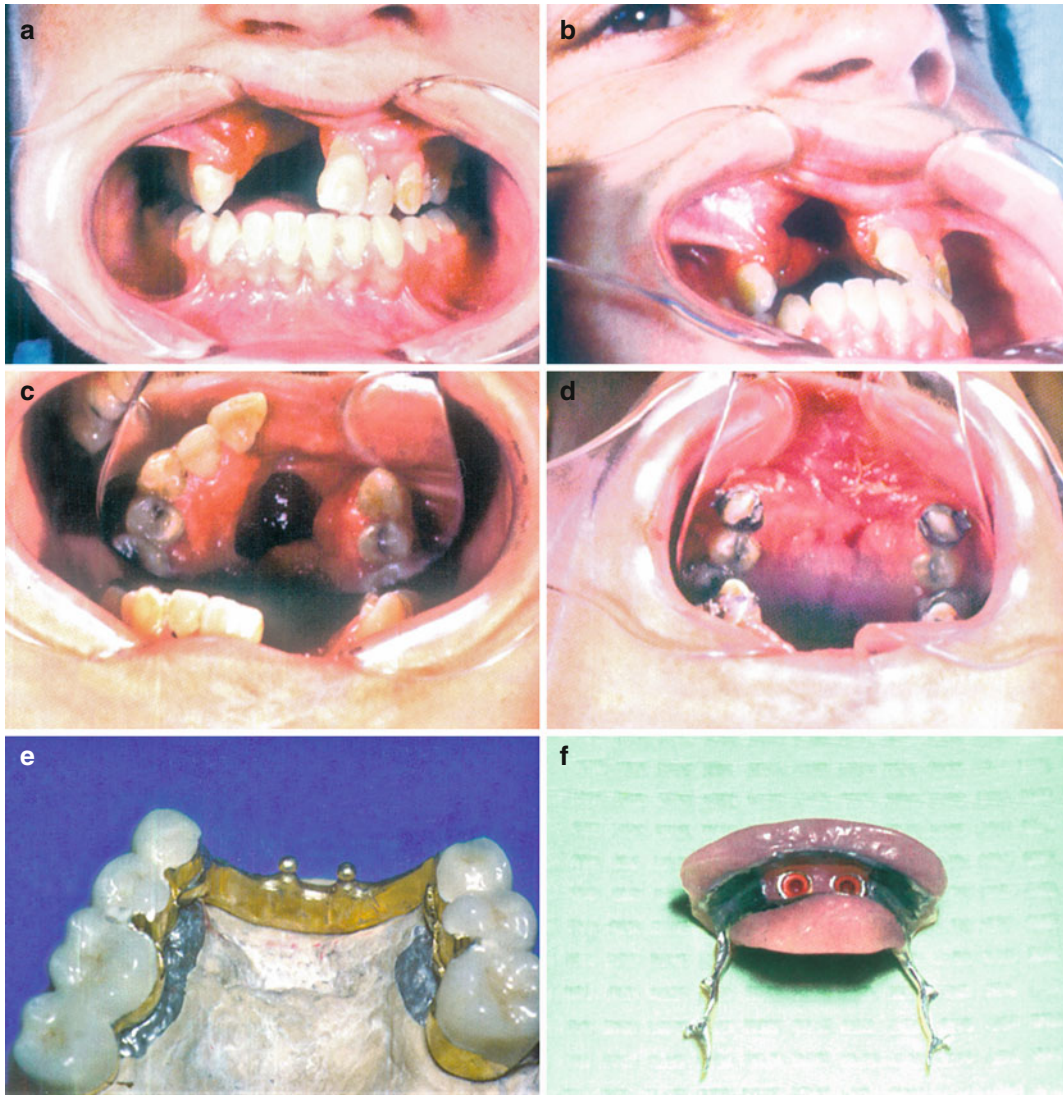


Fig. 6.11 (**a**–**m**) Case SP. Surgical and prosthetic treatment to replace a missing portion of a premaxillary segment and to close an oronasal opening (a transfer patient). Loss of blood supply to the right premaxillary area led to its exfoliation. Treatment plan: Because the remaining blood supply to the left maxillary and central incisor was questionable and the teeth showed marked root absorption, the dentist (Alan Stoler) recommended their removal. The remaining teeth were to be crowned to support an anterior cast gold section to which a removable prosthetic appliance would replace the missing incisor teeth and bumper the lip. (**a**–**c**) Frontal and palatal view of an oronasal opening due to the loss of a portion of the right premaxillary segment. (**d**) Palatal view following soft tissue closure of the oronasal opening. (**e**, **f**) Anterior prosthetic appliance with splinted posterior teeth; anterior appliance with two holes and "o" rings to receive the two extensions on the anterior splint

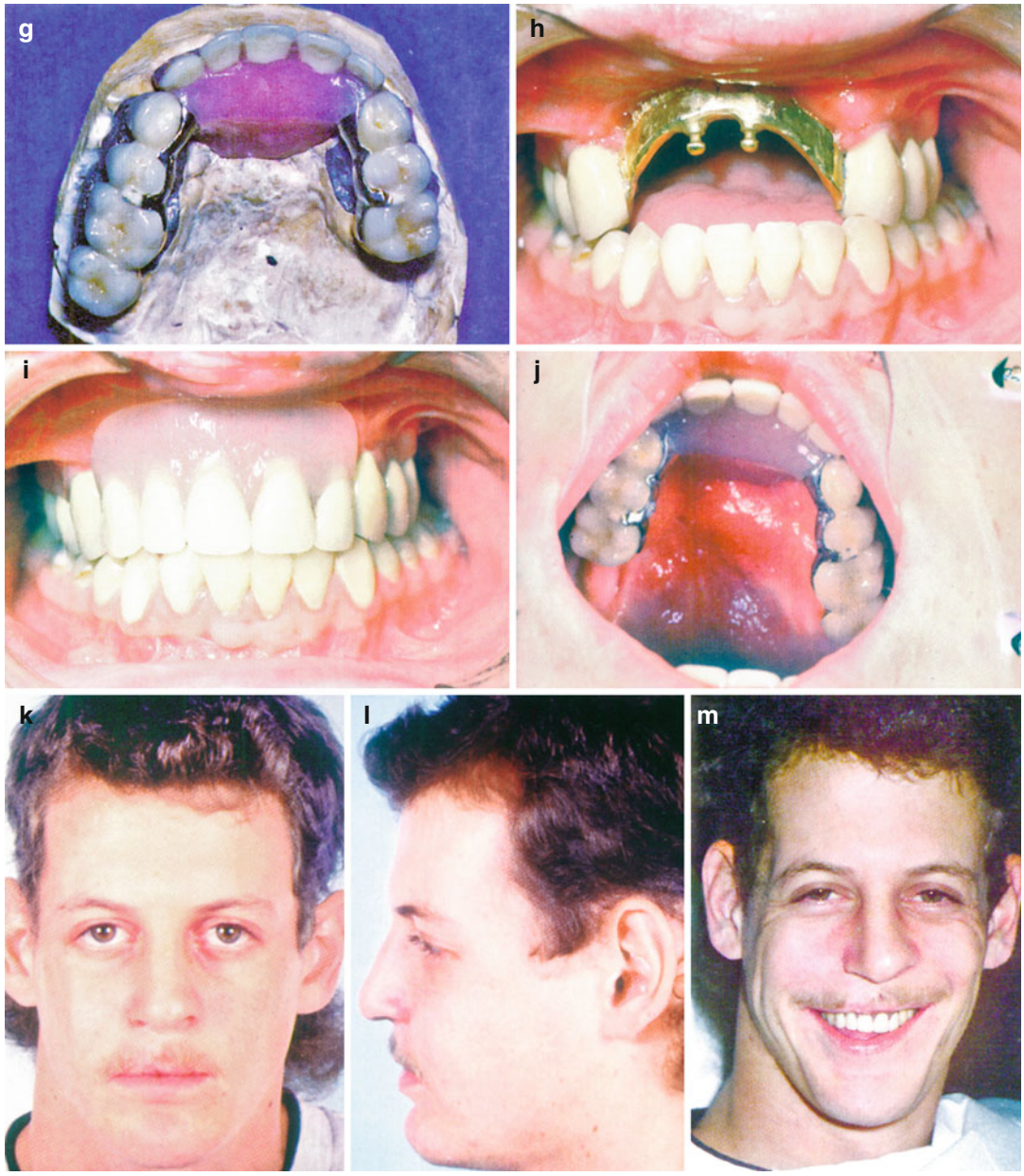


Fig. 6.11 (*continued*) (**g**) Posterior teeth splints with the anterior removable prosthesis in place on a model. (**h**, **i**) The gold anterior section spans the inter-cuspid space. (**j**) Palatal view with the anterior prosthetic appliance in place. (**k**–**m**) Facial photographs showing good upper lip support with excellent dental aesthetics

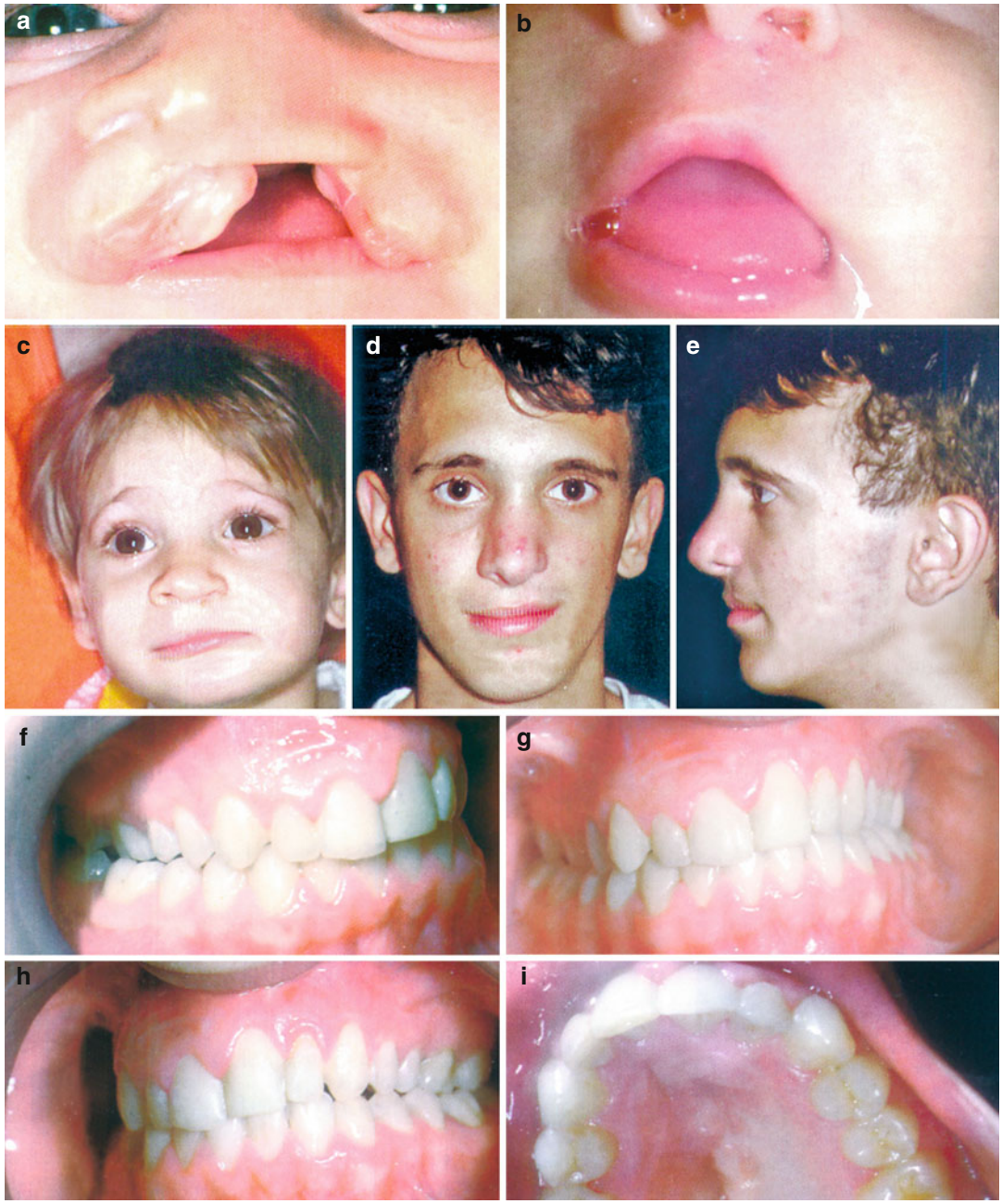


Fig. 6.12 (**a**–**k**) Case JK (AF-64). Excellent facial and palatal growth in CUCLP when the palatal segments did not make contact after the lip was united. The lateral incisor is in position in the alveolar cleft area. (**a**–**i**) Serial facial and intraoral photographs show changes to the lip and nose after lip adhesion and definitive lip surgery using Millard's rotation advancement procedure. Left facial asymmetry is apparent in the frontal photograph and is more noticeable in the intraoral photograph at completion of orthodontic treatment. The *left side* was kept in class II occlusion

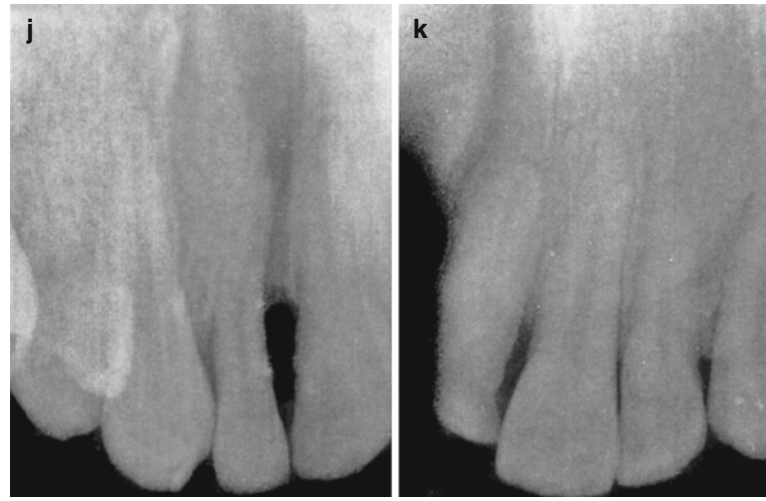


Fig. 6.12 (*continued*) (**j**, **k**) Periapical films show bone closure of the alveolar cleft space with good lateral incisor alignment

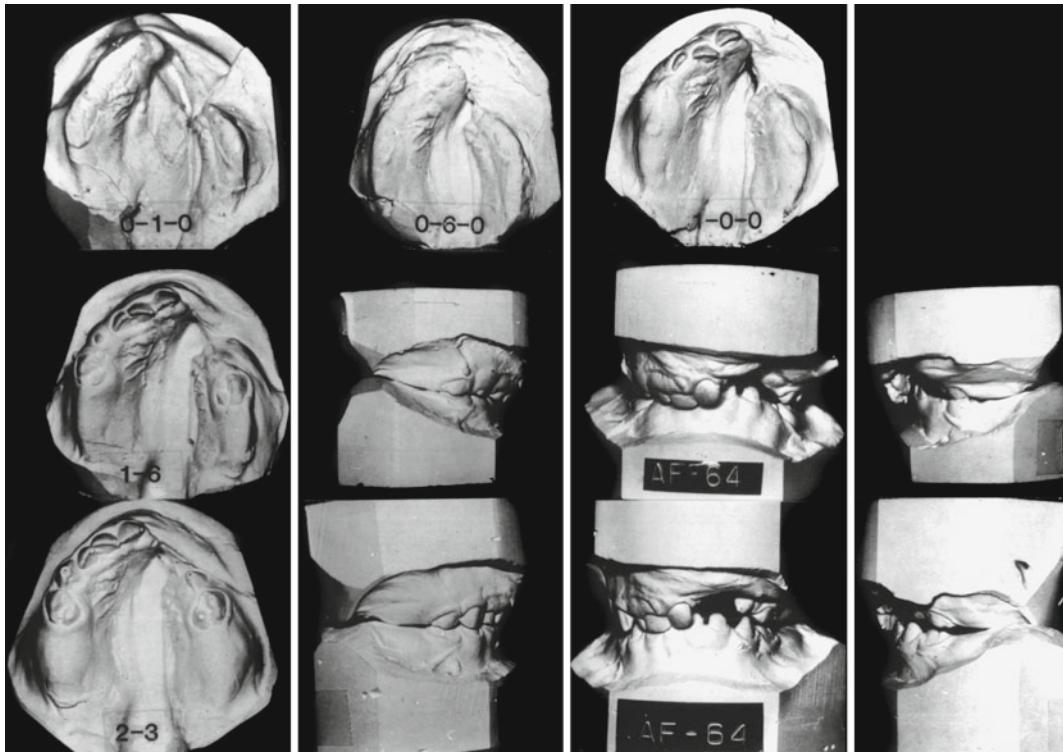


Fig. 6.13 Serial casts of Case JK. Newborn: The nasal septum bows toward the cleft segment, creating a very small cleft space. The great distance between alveolar segments is due to the upward tilt of the larger segment coupled with a small cleft segment. 0–6–0 After the lip is united, both palatal segments move toward the midline, narrowing the cleft space, more on the right than the left side. However, the alveolar segments still do not meet due to the inferior turbinate on the lesser segment making premature contact with the septum, preventing the lesser palatal segment from further medial movement. Note that the premaxillary portion of the larger segment has not moved medioposteriorly 1–0–0, 1–6, 2–3, 3–2

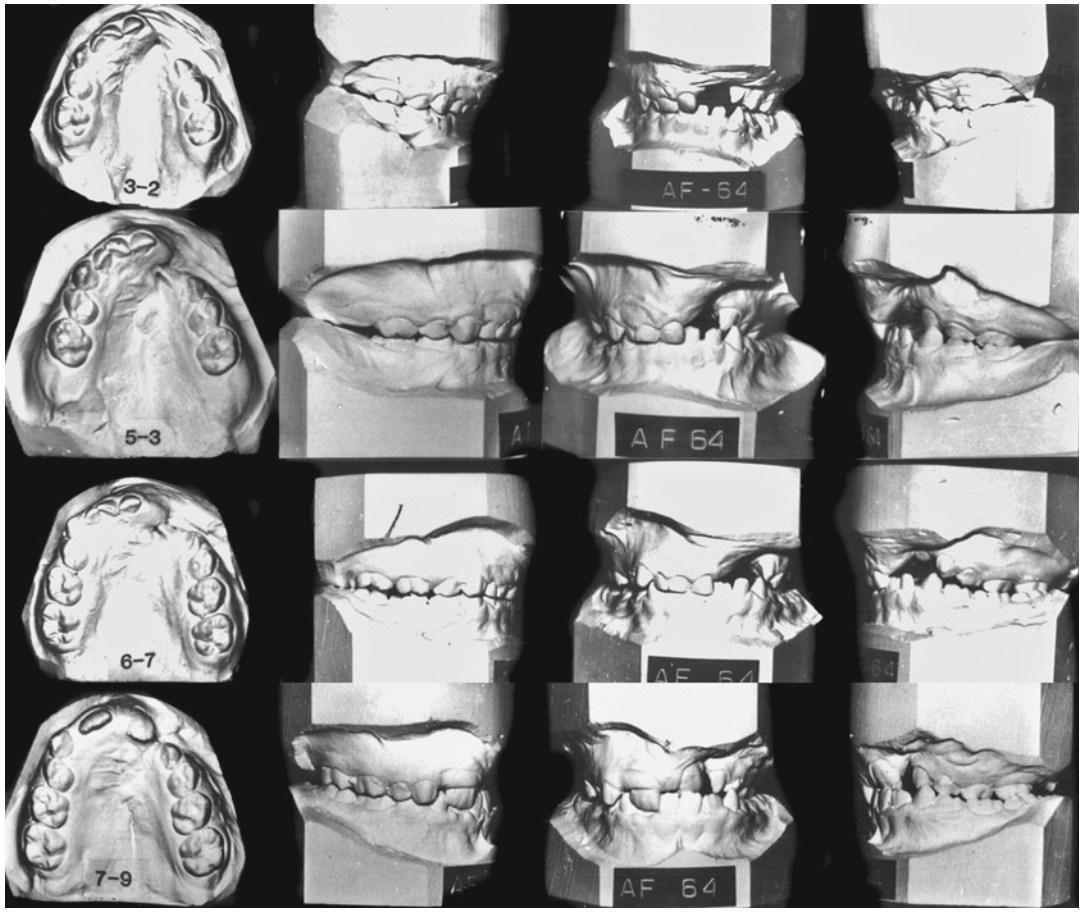


Fig. 6.13 (*continued*) The palatal segments are still apart. 5–3 After removal of the inferior turbinate and with palatal closure, the tissue contracture created by the modified von Langenbeck procedure pulls the palatal segments together, placing the buccal teeth in the cleft segment in crossbite. 6–7 The palatal segments have been expanded. 7–9 Without palatal arch retention, the crossbite returned. The ectopically erupted left central incisor is in crossbite

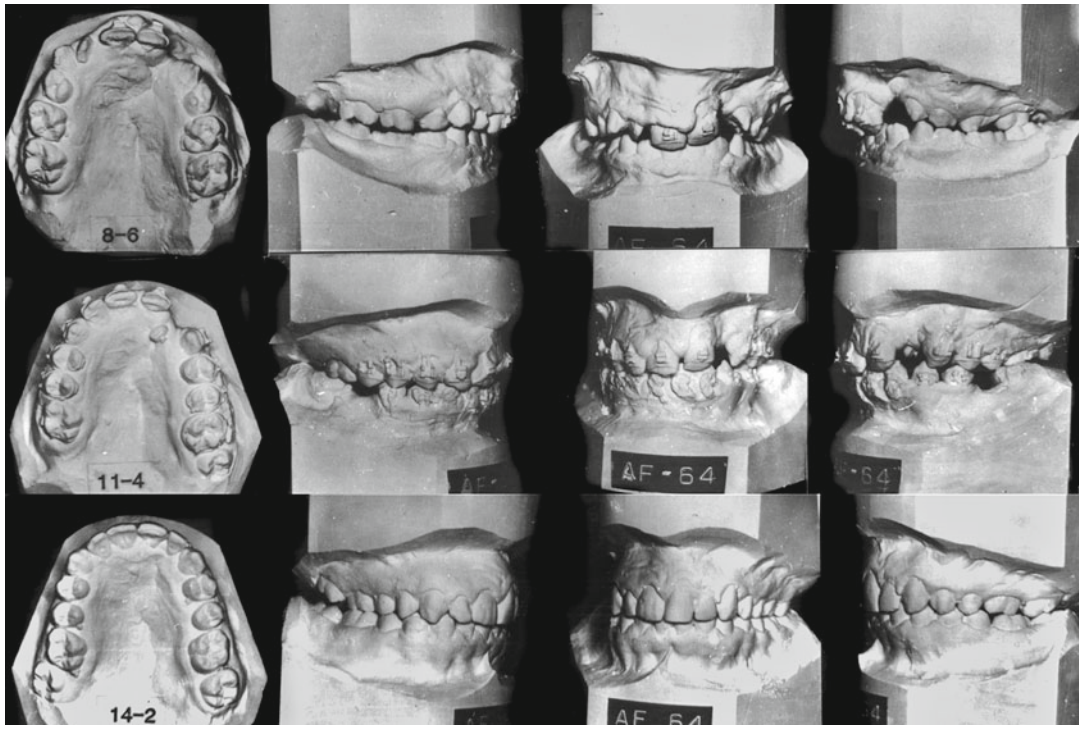


Fig. 6.13 (*continued*) 8–6 Arch expansion mechanics were reinstituted, and the left central incisor advanced into proper overjet. 11–4 and 14–2 Final orthodontic treatment was instituted and completed at 14 years of age. The impacted left lateral incisor was brought into alignment through the secondary alveolar cranial bone graft. Comment: After secondary alveolar bone grafting, arch expansion in most cases is stable. However, in cases where new bone does not extend to the nasal aperture, we believe the buccal crossbite has a good chance of returning. The left side was in class II occlusion, because it was not certain that the left lateral incisor could be properly aligned. If it was to be extracted, the cuspid would be positioned in the lateral incisor space

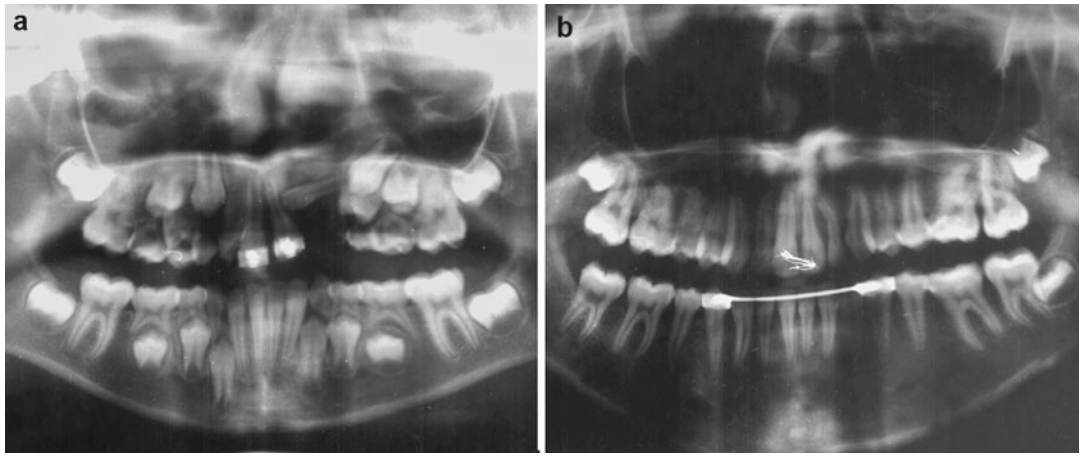


Fig. 6.14 (**a**, **b**) Case JK (AF-64). (**a**) Panorex: The left lateral incisor is palatally and horizontally impacted. (**b**) After treatment, the lateral incisor is well-aligned within the arch. Note that the curvature to the root possibly occurred before it was fully formed and during orthodontic movement

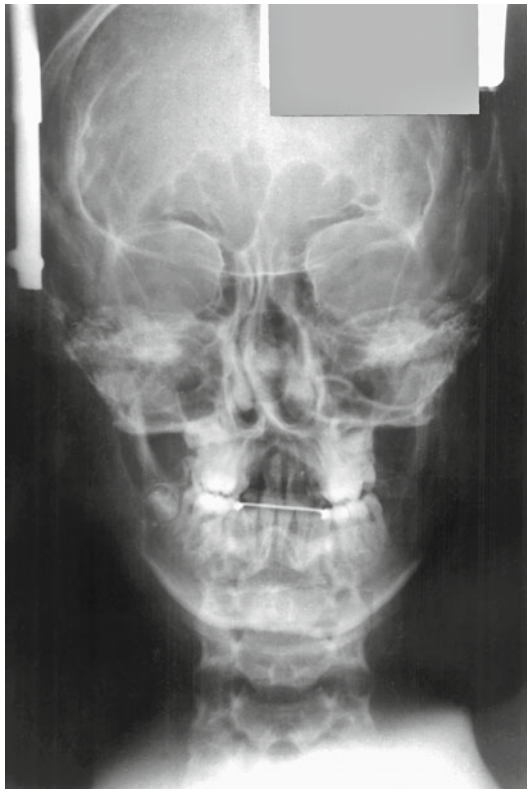


Fig. 6.15 Case JK (AF-64). Frontal cephaloradiograph shows that the nasal chamber on the cleft side is very narrow with a very flattened inferior conchae. The nasal septum is extremely bowed toward the cleft side. A lower cuspid to cuspid retainer is being worn

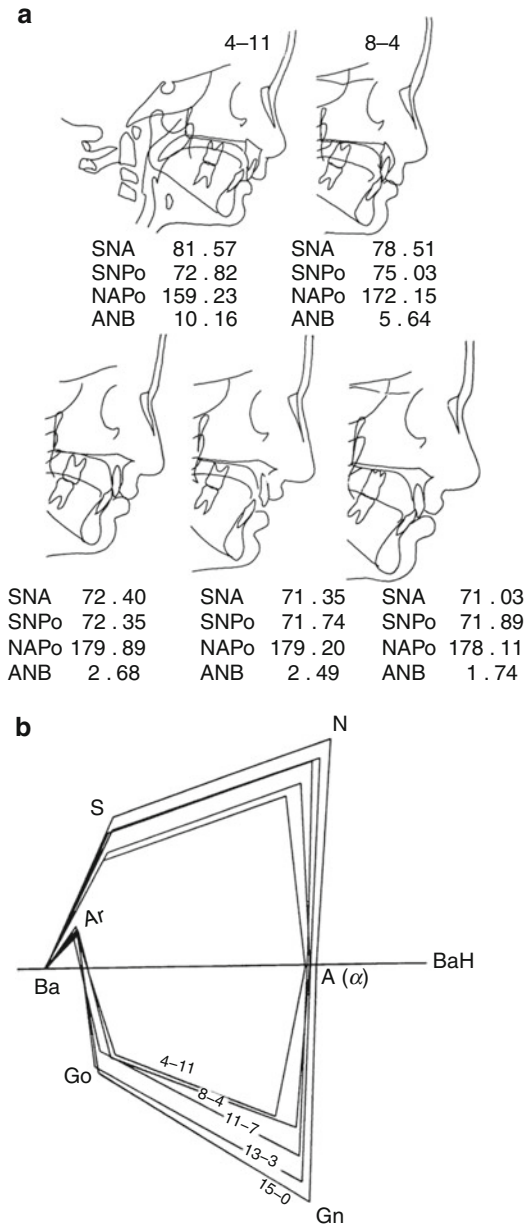


Fig. 6.16 (**a**, **b**) Case JK (AF-64). (**a**) Skeletal and soft tissue profile changes shown by lateral cephalometrics. The anterior projection of the midface and mandible relative to the anterior cranial base decreases with time as the profile flattens. The decreasing ANB angle reflects this change. (**b**) Superimposed polygons using the basion horizontal method. This series clearly shows that the flattening of the skeletal facial profile occurred around 8 years of age and was brought about by the growth at the anterior cranial base and the mandible, whose plane angle increased with time. There was almost no forward growth of the midface between 4–11 and 13–3 years with only a small postpubertal growth increment between 13–3 and 15–0 years of age

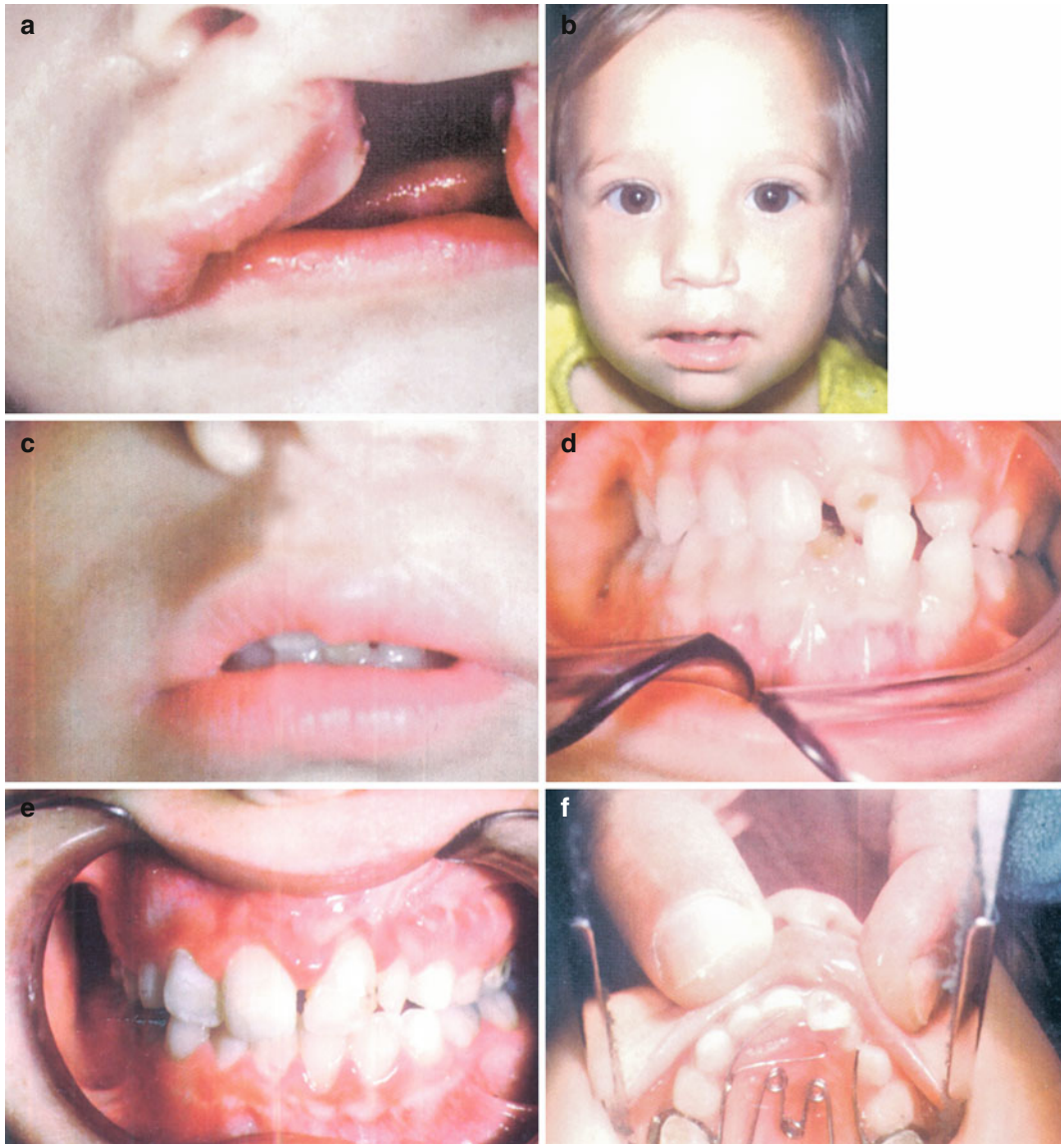


Fig. 6.17 (**a**–**s**) Case JD (AE 23). Complete unilateral cleft lip and palate. Excellent palatal and facial growth. A relatively large cleft space necessitated postponement of palatal closure until 20 months. Early secondary alveolar bone graft. Surgical history: Lip adhesion at 3 months followed by rotation advancement definitive lip repair at 6 months. Modified von Langenbeck palatal cleft closure at 20 months. Secondary alveolar cranial bone grafts at 6 years. (**a**) Before and (**b**) after lip repair. (**c**) 2 years, 5 months. Anterior and buccal crossbite. (**d**–**f**) 3 years, 4 months. Anterior and buccal crossbite correction with fixed palatal expander

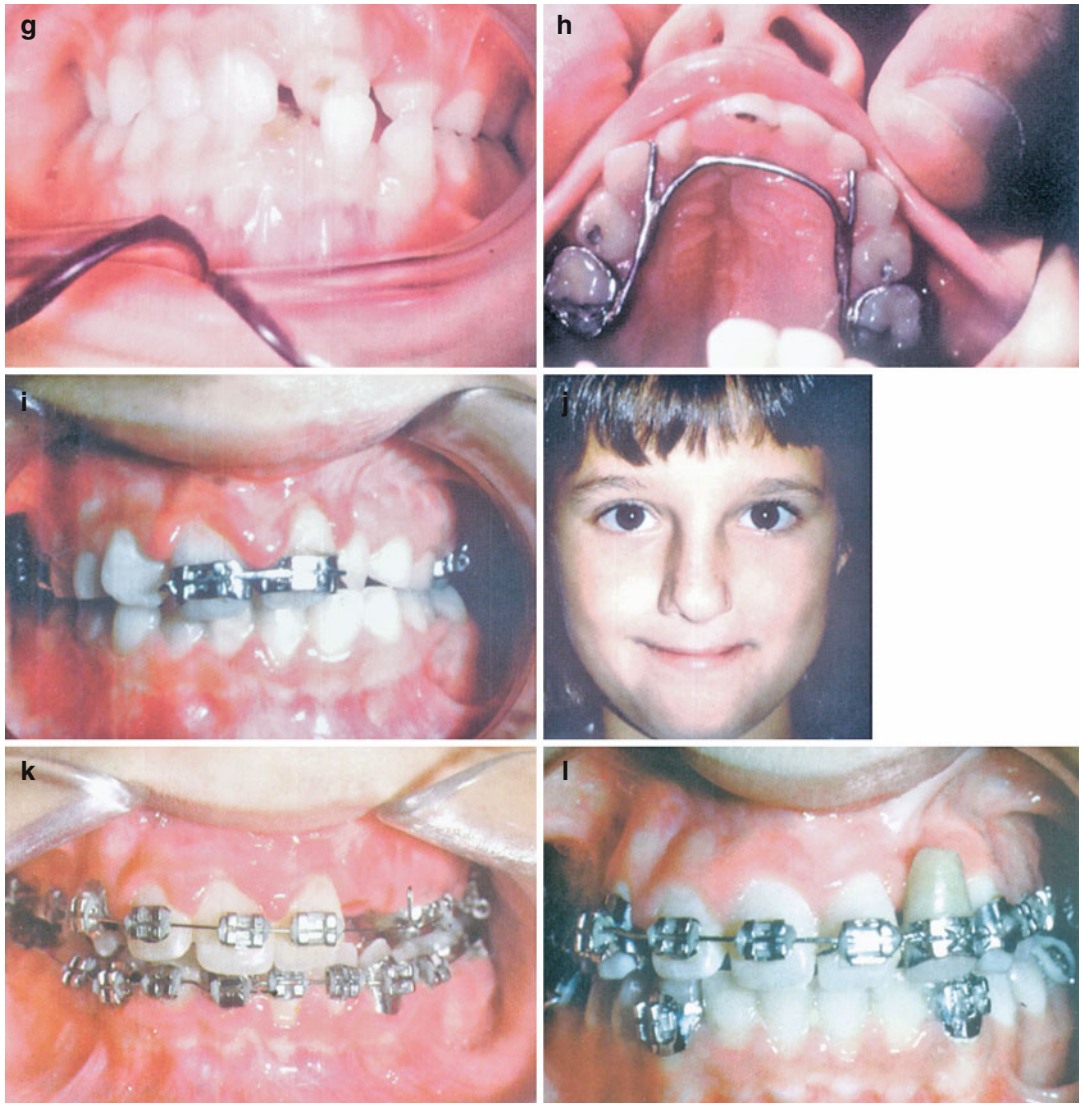


Fig. 6.17 (*continued*) (**g**, **h**) 4 years, 6 months. After expansion, a fixed palatal retainer. (**i**, **j**) 9 years. Central incisor aligned in the mixed dentition. (**k**, **l**) Orthodontic appliance with a false lateral incisor tooth with band attached to the orthodontic arch wire

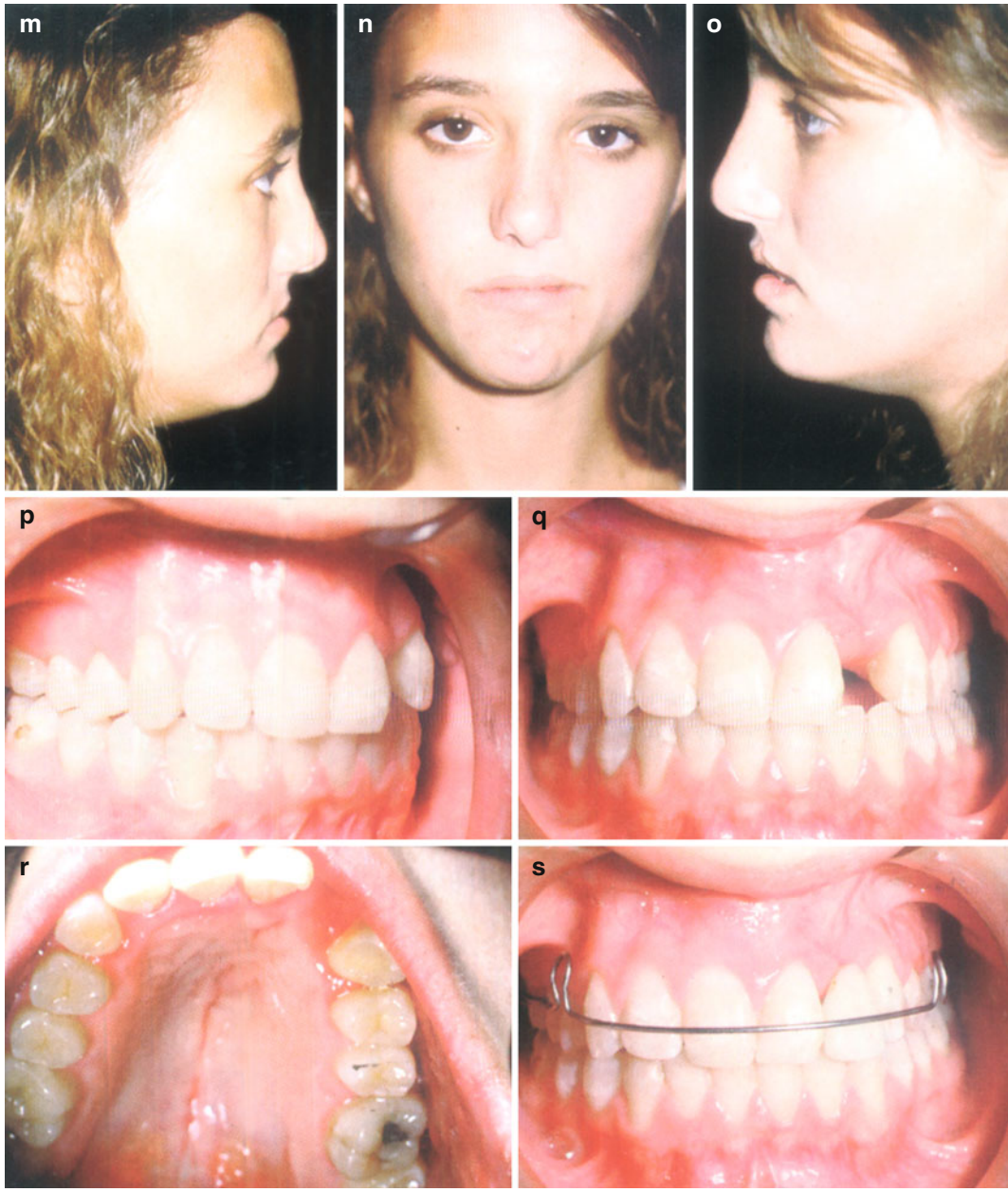


Fig. 6.17 (*continued*) (**m**–**r**) Facial and intraoral photographs at 15 years of age – on completion of orthodontic treatment. Ideal dental occlusion. (**s**) Maxillary retainer with an attached lateral incisor pontic tooth

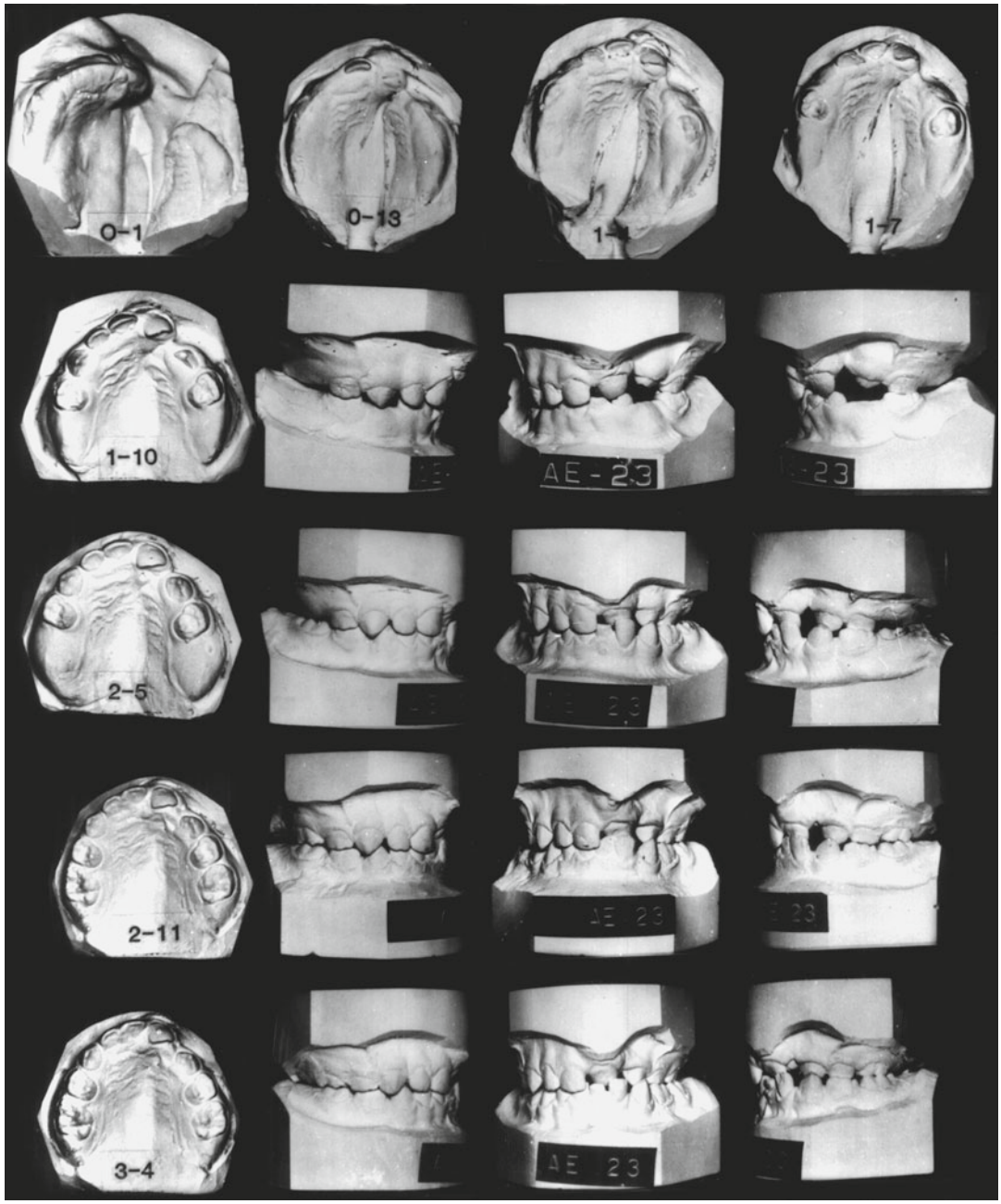


Fig. 6.18 Serial casts of Case JD. 0–1 At birth. 0–13, 1–4, and 1–7 With the institution of compressive lip muscle forces by uniting the lip, the lesser cleft segment moved medially to make contact with the vomer. The geometric changes to both segments brought the alveolar segments in good approximation 1–10, 2–5, 2–11, and 3–4

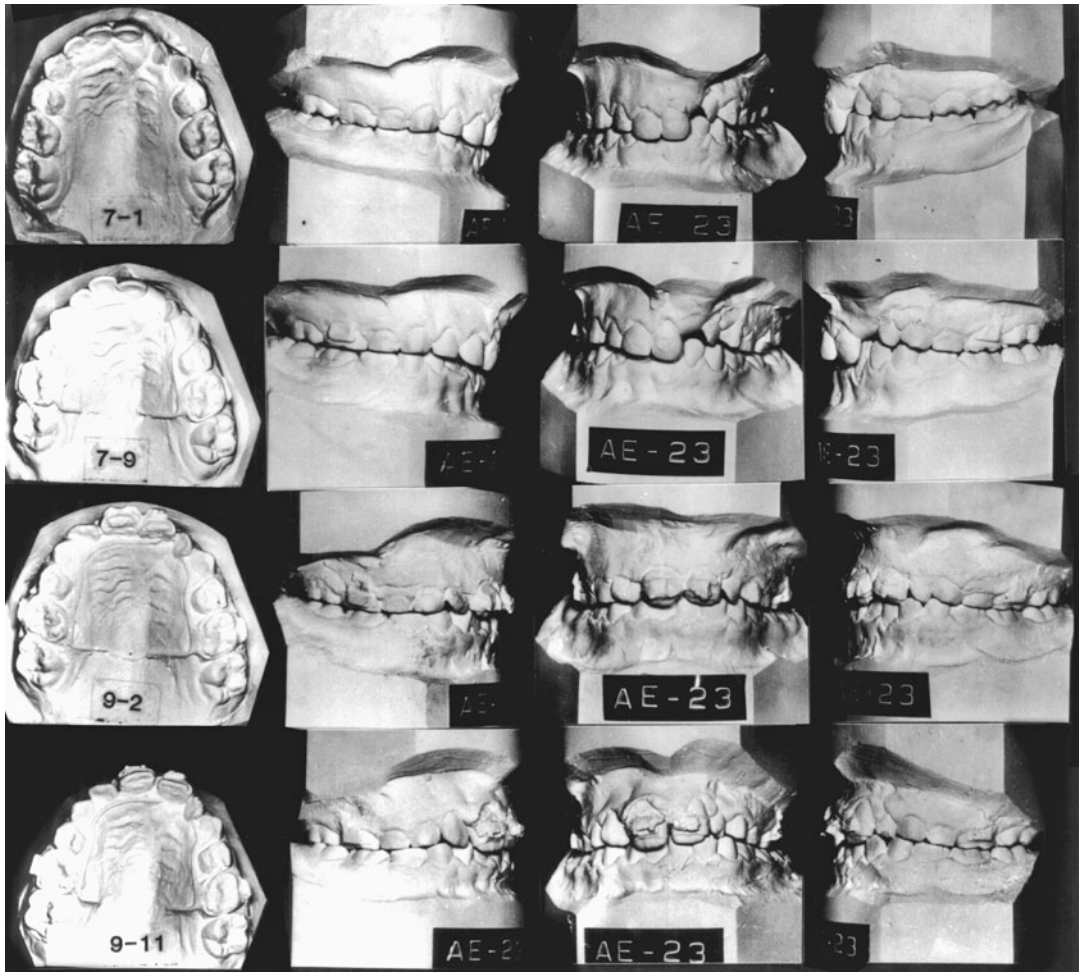


Fig. 6.18 (*continued*) However, after palatal cleft closure at 20 months, the lesser segment moved further medially placing the left cleft segment in crossbite. Due to ectopic eruption, the left central incisor was in crossbite. After palatal expansion and advancement of the left central incisor, excellent buccal occlusion was established. 7–1, 7–9, 9–2, and 9–11. Fixed palatal retainer is worn to maintain the arch forum

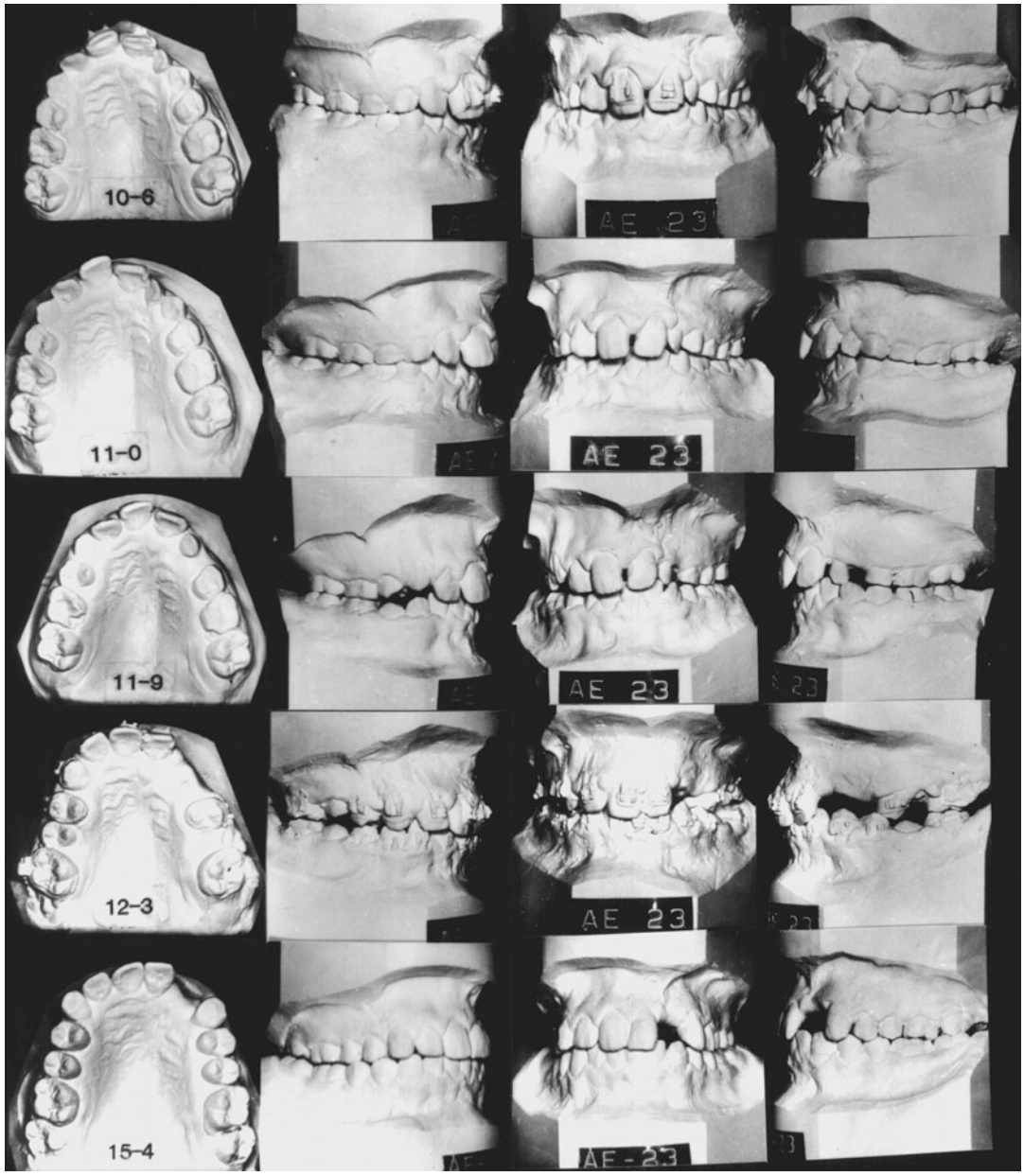


Fig. 6.18 (*continued*) 10–6, 11–0, and 11–9 Palatal form retained with palatal appliance. The upper central incisors were rotated for aesthetic purposes. The left malformed lateral incisor was left in place until orthodontic treatment was instituted at 12 years of age. 15–4 Orthodontic treatment completed. Note the slight flaring of the upper incisors and upper left cuspid, which was due to slight anterior maxillary bone deficiency. This is not an uncommon finding in complete unilateral clefts of the lip and palate

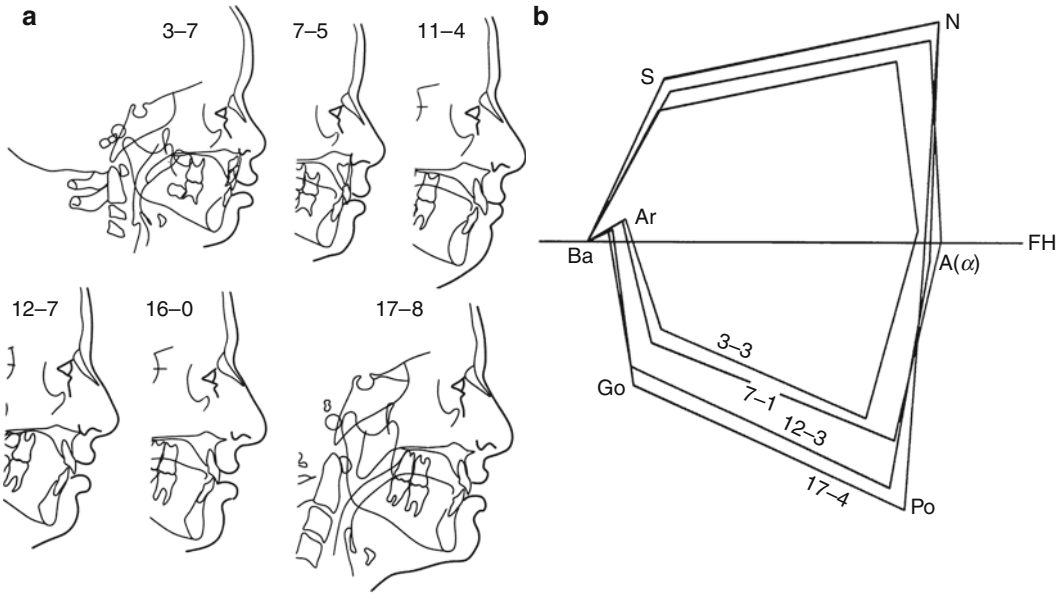


Fig. 6.19 (**a**) Skeletal and soft tissue changes in Case JD. (**b**) Superimposed polygons using basion horizontal method (Coben). Both of these analyses show excellent facial changes. The midfacial protrusion actually reduced between 7–1 and 17–4. Comments: One of the main controlling factors in the treatment of children with clefts that involve the anterior bony segment is the amount of osteogenic deficiency in the area. In many noncleft children, some advancement of the anterior teeth is essential; however, advancing the anterior teeth in the child with a cleft results in flared incisor teeth because the bone deficiency prevents the roots from being brought forward, even with anterior root torque using rectangular arch wires

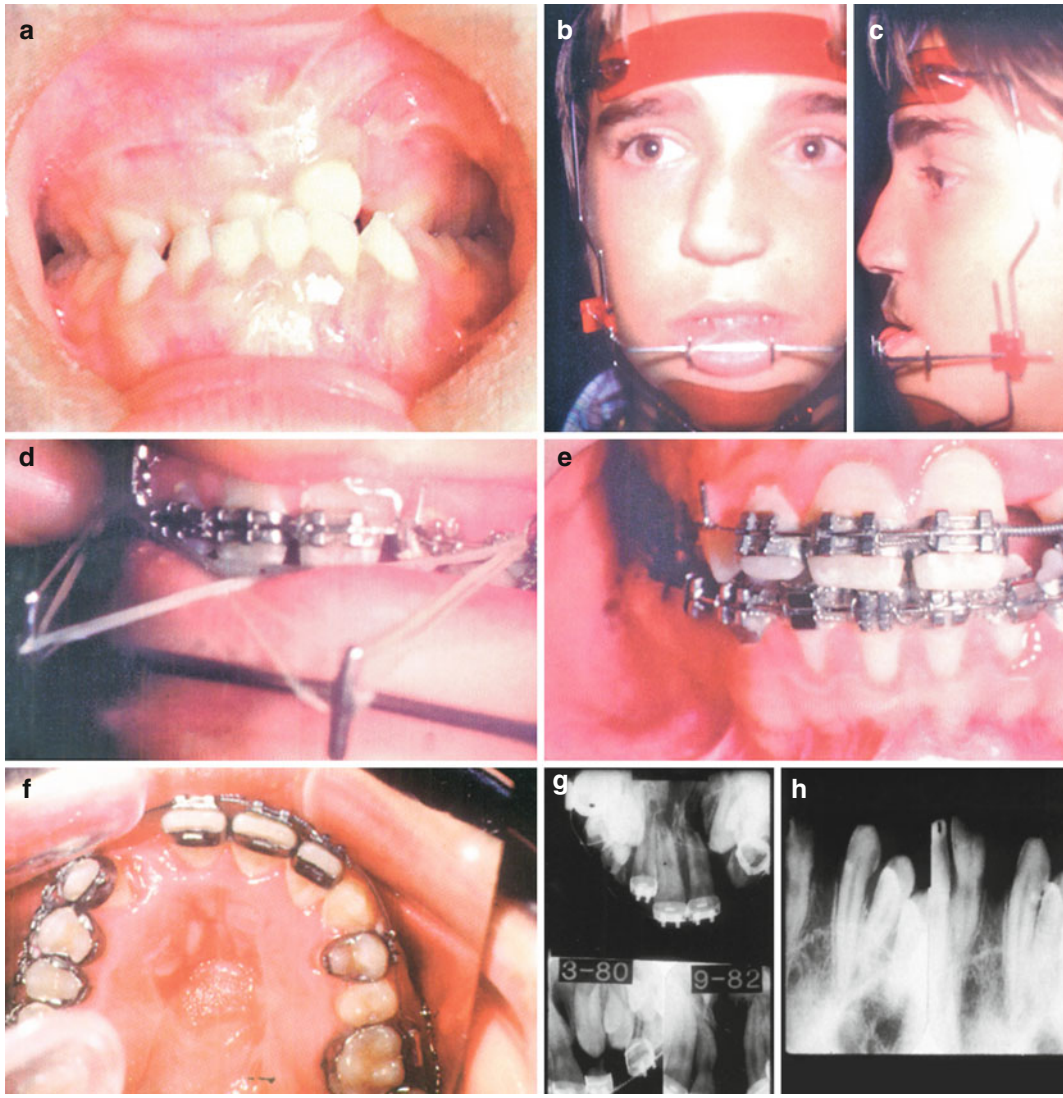


Fig. 6.20 (**a**–**n**) Case AB (EE-49). UCLP illustrating use of protraction maxillary orthopedics to correct midfacial retrusiveness secondary to growth-inhibiting scar tissue and/or maxillary osteogenic deficiency. Surgical history: Lip closure at 6 months. Hard and soft palate cleft closure at 16 months using an island flap pushback. Secondary alveolar cranial bone graft at 10 years of age. (**a**) Two years 11 months of age. Anterior and bilateral buccal crossbite could not be corrected in the deciduous or mixed dentition. (**b**–**d**) Orthodontic-orthopedic forces to correct an anterior crossbite were initiated at 12 years of age using a Delaire-style protraction facial mask. (**e**, **f**) Ideal class I (neutroclusion) with an ideal overjet and overbite. Palatal view shows thick transpalatal scar tissue caused by the island flap. (**g**, **h**) Periapical films after secondary alveolar cranial bone graft. No left lateral incisor is present, but good cleft space closure is evident

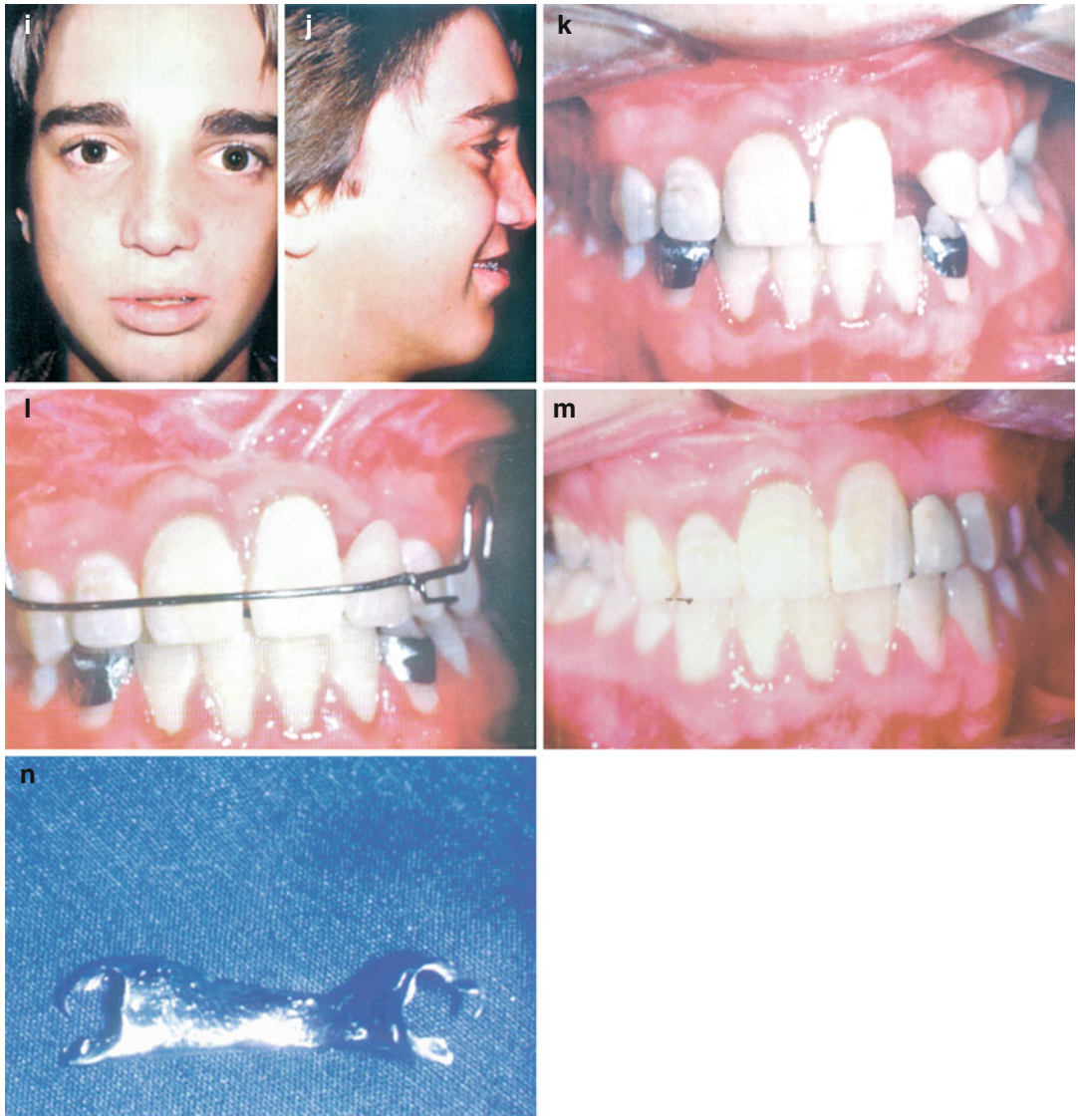


Fig. 6.20 (*continued*) (**i**–**k**) Facial and occlusal photographs before and after orthodontic treatment. (**l**) Upper retainer with pontic left lateral incisor. (**m**) Shows fixed bridge with false tooth and (**n**) cast palatal bar used to maintain the upper palatal form for relapsing in a crossbite occlusion

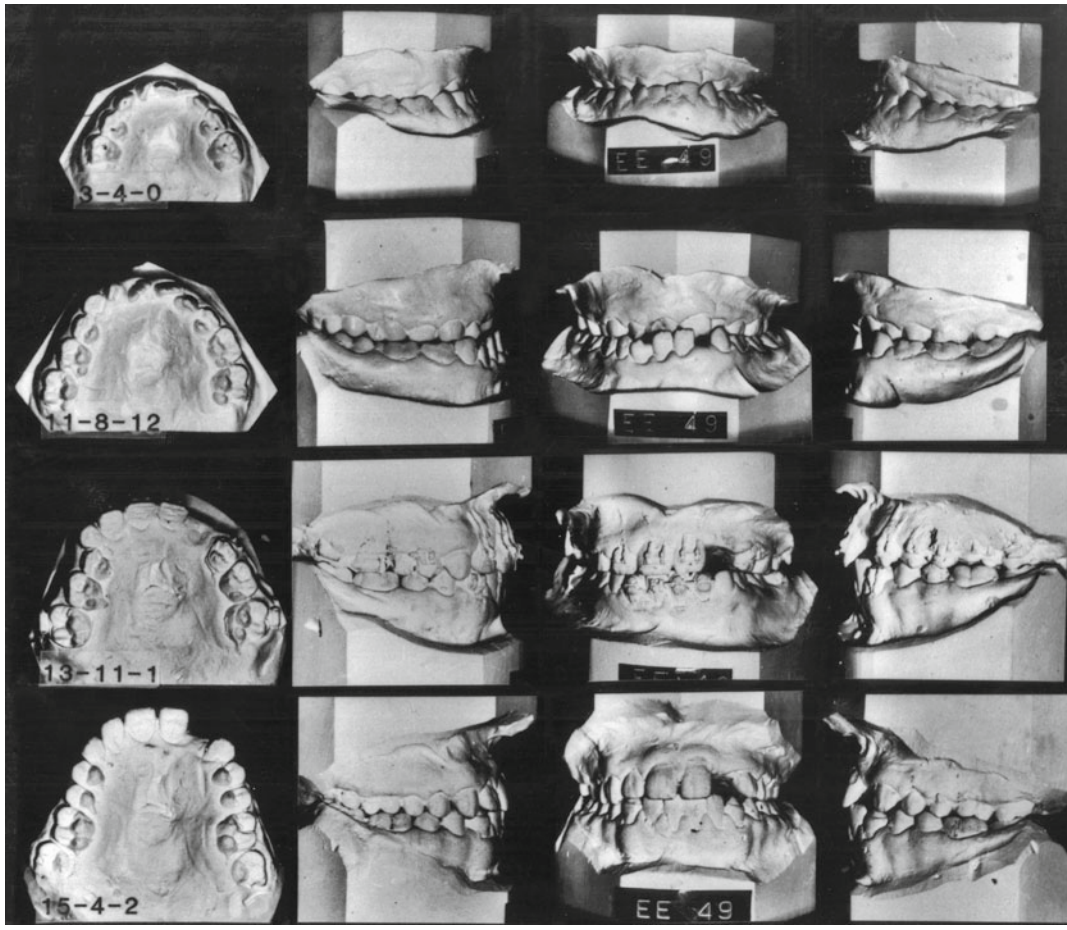


Fig. 6.21 Serial casts of Case AB. 3–4–0 After island flap hard and soft palate closure at 16 months of age resulting in bilateral buccal and anterior crossbites. 11–8–12 Occlusion just prior to orthodontic treatment. 13–11–1 After protraction mechanics using a Delaire-style facial mask. 15–4–2 Occlusion after orthodontics. Comments: Because maxillary deficiency is almost always present, "A" point (subnasal) in the premaxillary area needs to be brought forward by using labile root torque on a rectangular arch

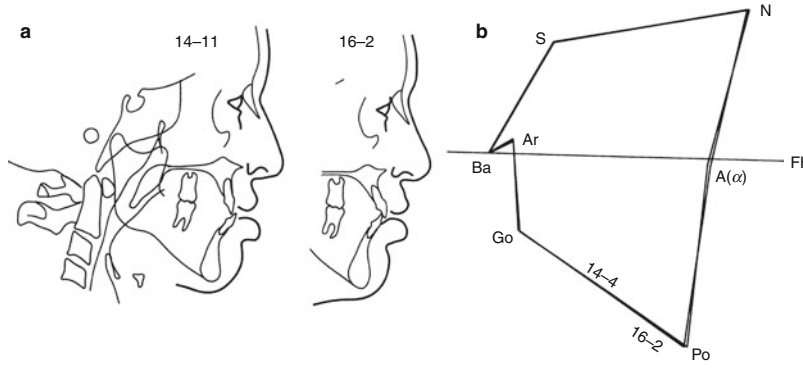


Fig. 6.22 (**a**, **b**) Case AB. (**a**) Cephalometric tracings at 14–11 and 16–2. (**b**) Superimposed polygons using basion horizontal method. A slight change in midfacial protrusion is noted after protraction forces were used to correct the midfacial retrusion and anterior crossbite. In this case, the changes in the maxillary incisor axial inclination aided anterior crossbite correction more than maxillary protraction

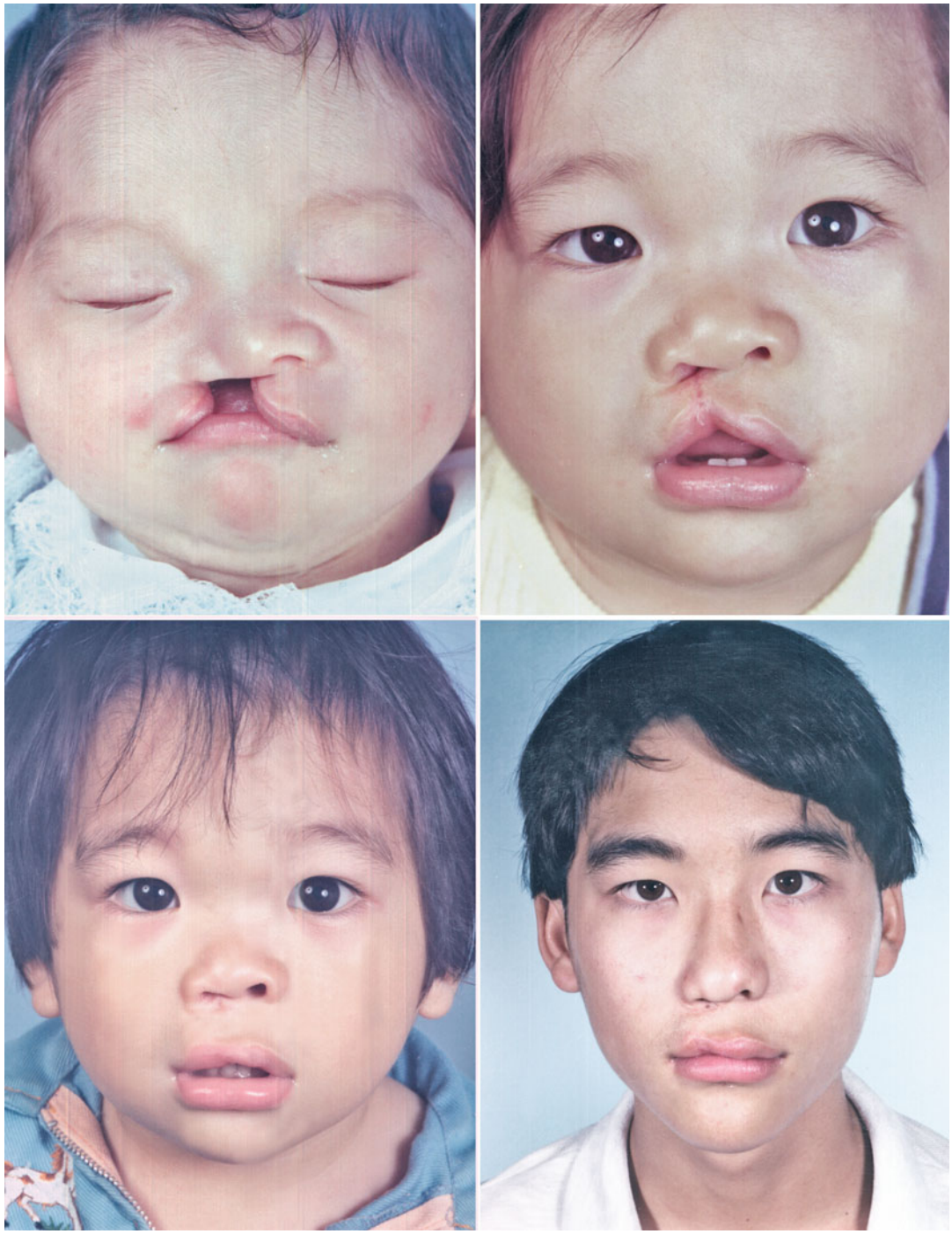


Fig. 6.23 Conservative treatment of a patient with CUCLP. Lip adhesion at 3 months. Rotation advancement at 6 months. Palatal closure at 22 months using a von Langenbeck and vomer flap. Excellent occlusion and a flat face profile resulted

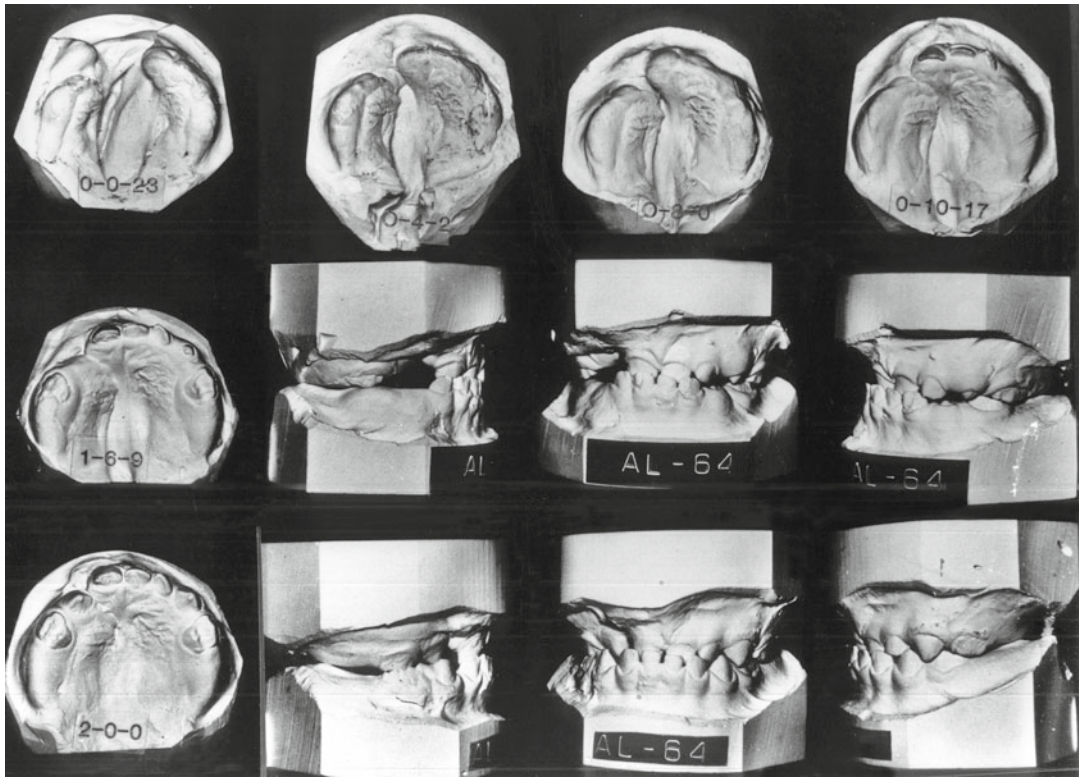


Fig. 6.24 Case AL 64. Complete unilateral cleft lip and palate. Serial casts, 0–0–23 to 2–0–0: Lip adhesion brings the overexpanded palatal segments together. The premaxillary portion of the larger noncleft segments palatally positioned placing the teeth into an anterior crossbite. The palatal cleft was closed at 1–11 with von Langenbeck plus vomer flap

Fig. 6.24 (*continued*) 8–9–13 to 15–4–7 The maxillary anterior teeth were advanced into a proper overjet and overbite. Due to arch crowding, the first bicuspids were extracted and spaces closed. The alveolar bone graft at approximately 8 years permitted the impacted lateral incisors to erupt into place. Note: The right lateral incisor crown is malformal, but the root size and shape is normal. The crown will eventually be capped

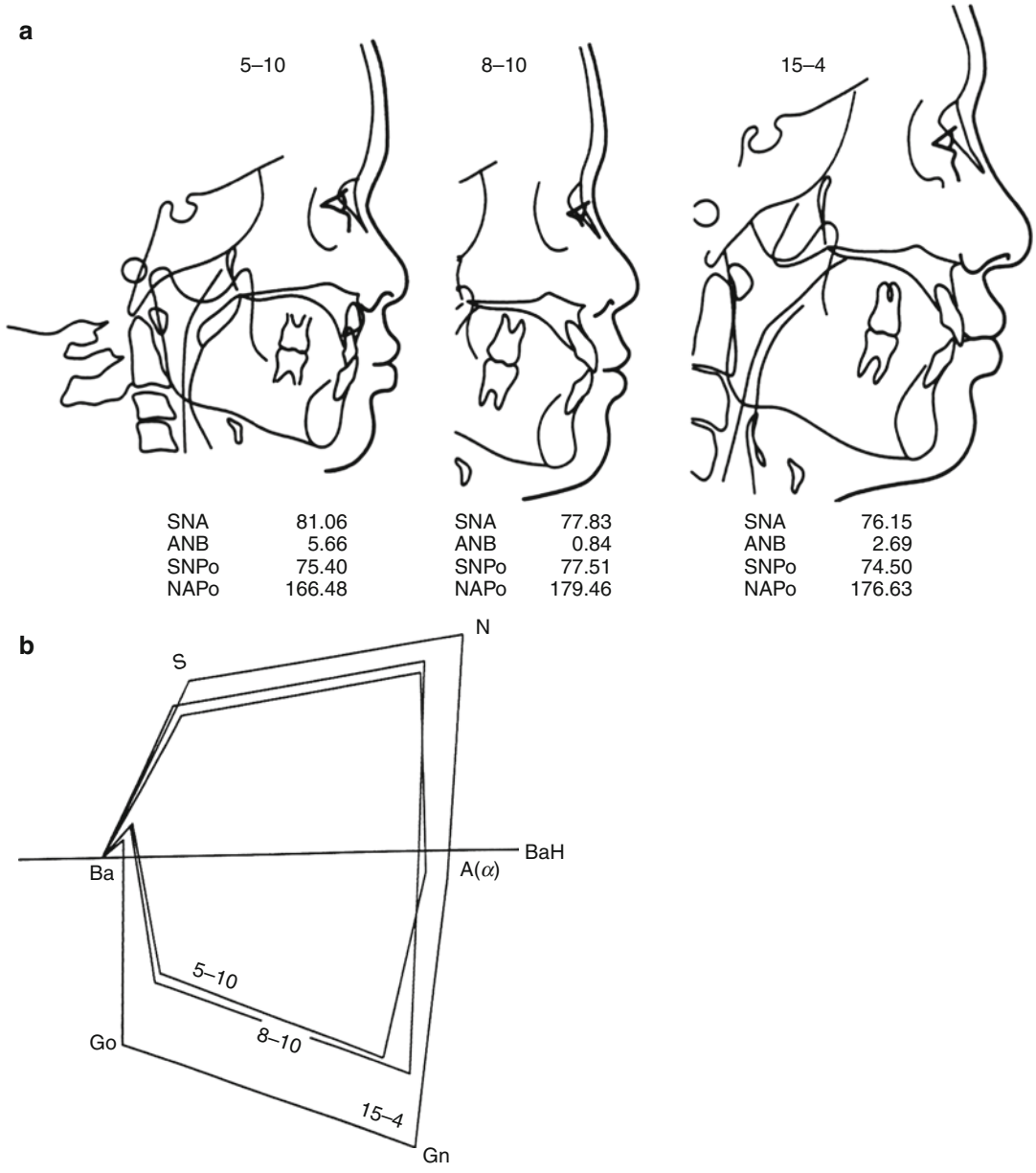


Fig. 6.25 (**a**, **b**) Case DM-AL 64. Very good facial growth pattern. The flattening of facial profile was dependent on good growth of all parts of the facial skeleton. The extraction of all the maxillary and mandibular first bicuspids was necessary to retract the incisors and uncrowd the dentition

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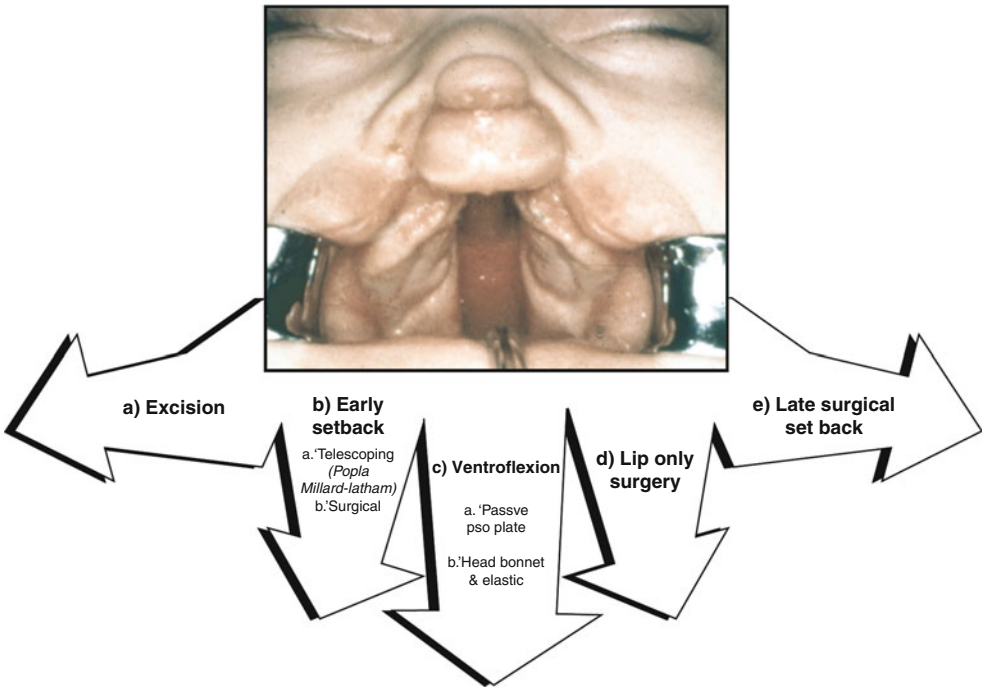
Complete Bilateral Cleft Lip and Palate

7

Samuel Berkowitz



The surgeon is confronted with the following options when faced with a protruding premaxilla at birth:

1. Uniting the lip over the protruding premaxilla and considering later surgical setback and other surgical options
2. External elastics attached to a head bonnet or elastic tape to the cheeks to ventroflex the premaxilla
3. Early surgical premaxillary setback
4. Complete removal (excision of the premaxilla)
5. Early mechanical retrusion prior to lip surgery or presurgical orthopedic treatment (PSOT)

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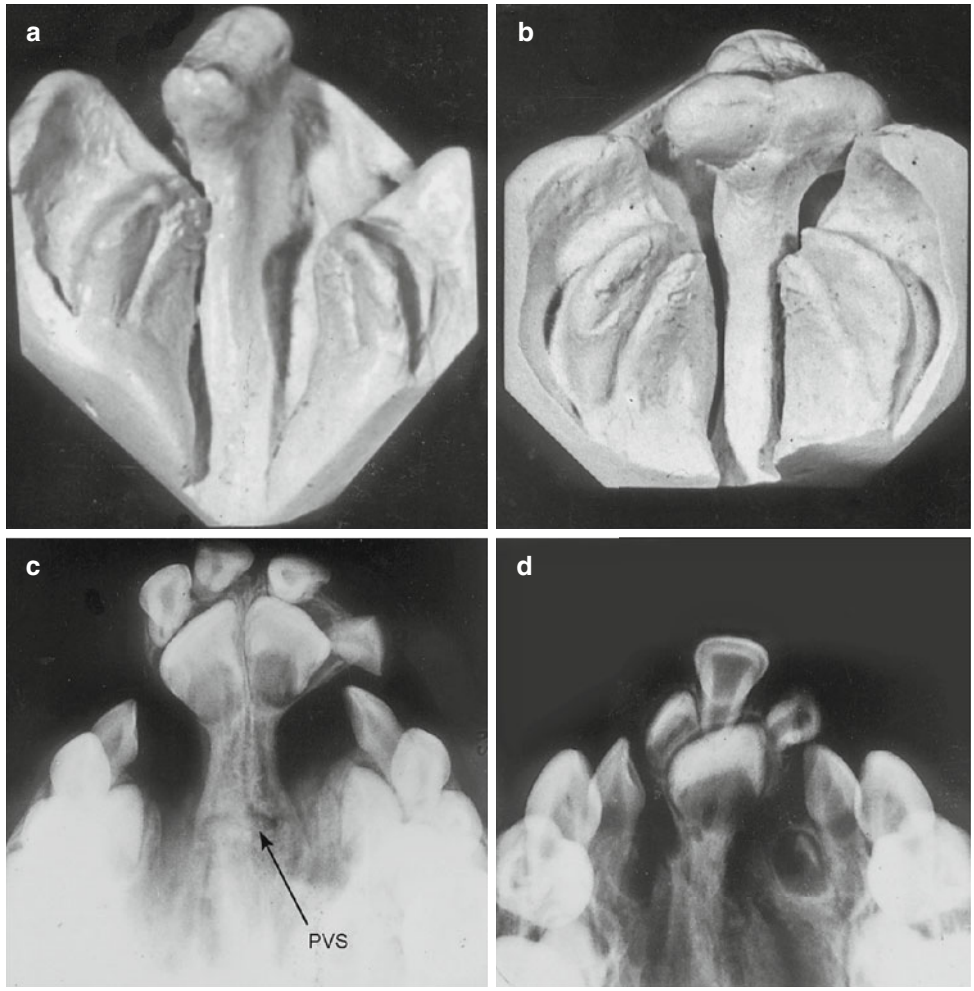


Fig. 7.1 (**a**) Small premaxilla: three deciduous incisors with one unerupted permanent central incisor. (**b**) Large premaxilla: four deciduous incisors with two permanent unerupted incisors. (**c**) Occlusal radiographs of premaxillae showing variations in the number of anterior deciduous teeth and permanent tooth buds. (**d**) Occlusal radiographs of premaxillae showing variations in the number of anterior deciduous teeth and permanent tooth buds

without retraction and with or without primary bone grafting or periosteoplasty

6. Lip adhesion followed by definitive lip surgery at a later age

7.1 Premaxillary Protrusion: Real or Apparent? Is the Palate Deficient in Bone?

A bilateral cleft of the lip and palate can be complete or incomplete on one or both sides (see Chap. 4). Any number of variations can exist. In both incomplete and complete bilateral clefts of the lip and alveolus, the size and shape of the premaxilla are dependent on the number of tooth buds and their distribution, making it symmetrical or asymmetrical (Fig. 7.1). Because clefts of the lip/alveolus and the hard and soft palate come from different embryological sources, the cleft may involve the lip and alveolus with or without involving the hard and soft palate.

Based on the observations of Veau and Borel (1931), Veau (1934) and Browne (1969) and the later work of Friede and Pruzansky (1972), Bergland and Borchgrevink (1974), Harvold

(1954), Berkowitz (1959), Friede (1977, 1978), Atherton (1967, 1974), and Handelman and Pruzansky (1968), the cause of premaxillary protrusion is the result of tension and the resulting bony overgrowth produced at the premaxillary-vomerine suture (PVS) by displacement of that bone by the aberrant muscular forces of the detached outer musculature combined with the pushing force of the tongue fitting within the cleft (Fig. 7.2). These investigators, using

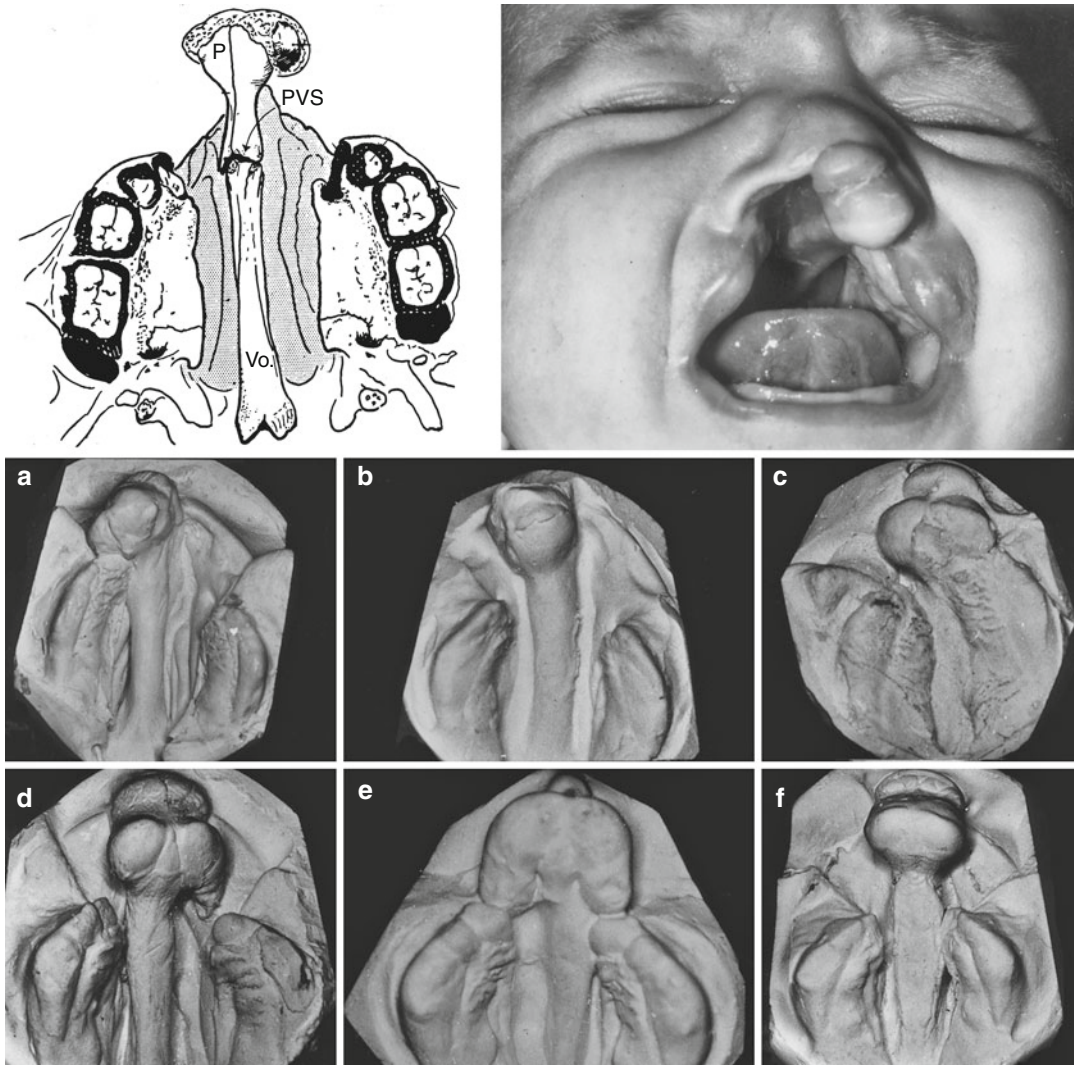


Fig. 7.2 (**a**) Line drawing of a complete bilateral cleft lip and palate with an arrow pointing to the premaxillary-vomerine suture (*PVS*) (**b**) A small protruding premaxilla IBCLP extends forward of the facial profile and lateral palatal segments. (**c**) Variations in bilateral cleft lip and palate. The size of the premaxilla varies with the number of teeth it contains. Classification is dependent on the completeness of clefting of the lip and alveolus and whether there is a cleft of the hard and soft palate. Yet one or both sides of the hard palate may or may not be attached to the vomer. If it is attached to the vomer, it is classified as being incomplete. Even in complete clefts of the lip and alveolus, the extent of premaxillary protrusion will vary. (**a**) Incomplete bilateral cleft lip and palate. Complete cleft lip and palate – *left side*. Incomplete cleft lip and palate – *right side*. (**b**) Complete bilateral cleft lip and palate. Complete cleft palate – *both sides*. (**c**) Incomplete bilateral cleft lip and palate. Incomplete palatal clefts – *both sides*. (**d**) Complete bilateral cleft lip and palate. Incomplete right and complete left palate. (**e**) Incomplete bilateral cleft lip and palate. Incomplete left palate and complete right palate. (**f**) Complete bilateral cleft of the lip and palate. Incomplete right and left palatal segments

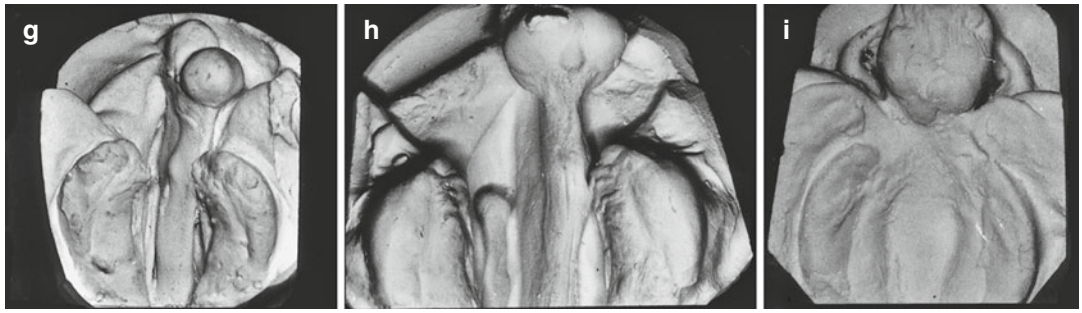


Fig. 7.2 (*continued*) (**g**) Complete bilateral cleft of lip and palate. Incomplete left palate and complete right palatal segment. (**h**) Complete bilateral cleft lip and palate. Incomplete left palate and complete right palate. (**i**) Incomplete bilateral cleft lip and alveolus. Normal palate

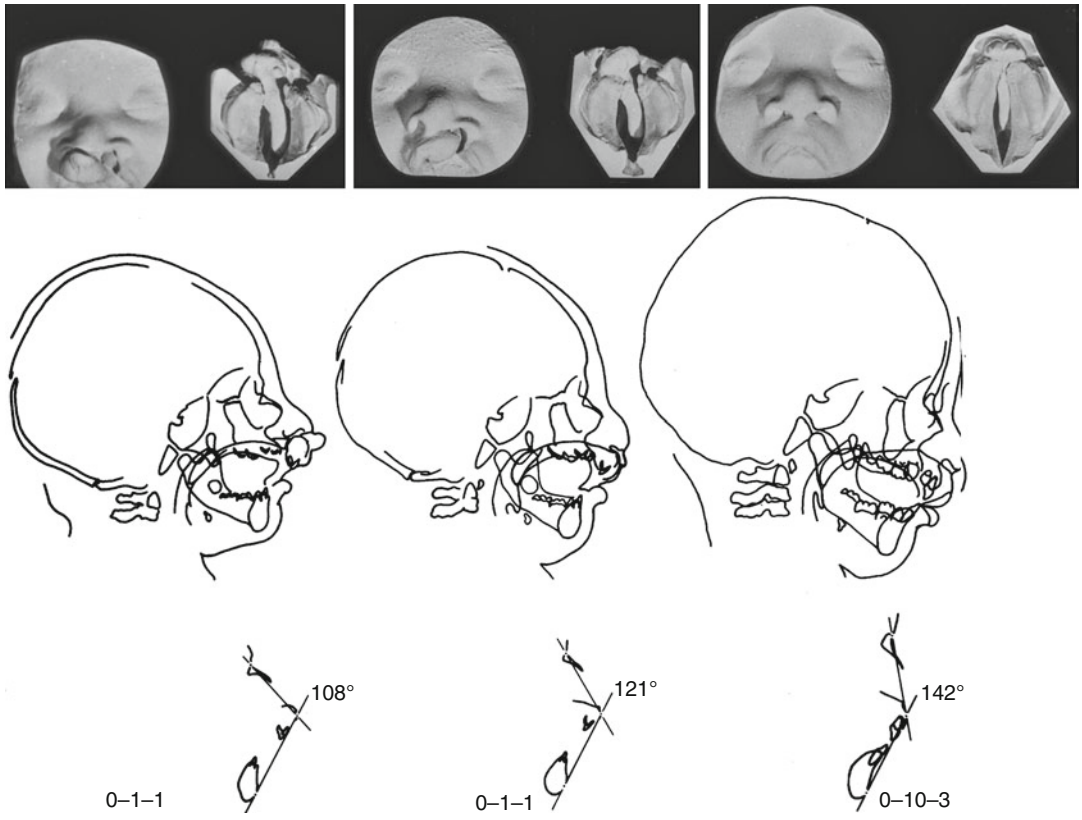


Fig. 7.3 Two-stage lip closure in a CBCLP. Ten months after lip closure, the angle of facial convexity changed from 108° to 142° as a result of the ventroflexion of the premaxilla at the premaxillary-vomerine suture (PVS) coupled with some additional mandibular growth. A two-stage lip closure is rarely necessary (Courtesy of S Pruzansky)

cephalometric data, concluded that the premaxilla is postured forward in the facial profile at birth (Fig. 7.3).

Bergland and Borchgrevink (1974) reported that, in complete bilateral clefts of the lip and alveolus with intact palates, the premaxilla was protrusive but palatal size was well within normal limits. In these cases, the septum was detached from the normally developed palates and the protrusion of the premaxilla was interpreted as representing a

premature release of the normal growth potential of the septum. The premaxilla apparently reached its geometric position within the skull at an earlier time prior to birth, yet the palatal segments followed a normal growth rate. Berkowitz's clinical records support the findings of Coup and Subtelny (1960), who reported marked palatal hypoplasia in bilateral clefts of the lip and palate. Berkowitz reports in this book that palatal growth rates are highly variable according to the amount of scarring created when closing the cleft space.

7.2 The Premaxillary-Vomerine Suture

Pruzansky (1953, 1971) and Friede and Morgan (1976) used metal implants on either side of the premaxillary-vomerine junction to demonstrate cephalometrically that this suture was a major site of bony overgrowth (Fig. 7.4). Earlier, Berkowitz (1959) and later Pruzansky (1971), Friede and Morgan (1976), Pruzansky and Friede (1975), Friede (1973), Vargervik (1983), and Berkowitz (1959) suggested that the overgrowth was probably a secondary reaction to the lack of restraint from the cleft orbicularis oris muscle. They showed that midfacial growth continued for 1–3 years even after the lip was united but at a slower rate. Berkowitz (1959) serial cephalometric tracings and digital cast study demonstrated that the midface continued to protrude for 2–3 years after the lip was united, then its forward growth slowed markedly (Fig. 7.5). Not surprisingly, in some instances, the facial convexity did not change due to the continued forward growth of the maxilla and premaxilla but was due to more vertical rather than forward growth of the mandible, maintaining the facial convexity (Figs. 7.6 and 7.7).

Fig. 7.4 To test growth at the premaxillar-vomerine suture (PVS) in complete bilateral cleft of the lip and palate, two pairs of metal pellets were placed on either side of the PVS at 6 months of age. At 3 years, 5 months of age, the distance between the anterior and posterior sets of pellets had increased with the growth at PVS. Note that there was no change in distance within each set of metal pellets (Courtesy of S Pruzansky)

Pruzansky (1953, 1971), Friede (1973), and Atherton (1967, 1974) described the premaxillary-vomerine suture as resembling other facial sutures. Friede and Morgan (1976) confirmed the presence of small islands of cartilage in the suture; these were secondary occurrences resulting from mechanical stresses and not part of a force system causing growth.

Burston (1960, 1967) and Latham (1969, 1973) following Scott (Scott 1956a, b) "Nasal Septum Growth" thesis, believe that the displacement is not real but only apparent. Burston thought that the lateral segments of the maxilla were retroplaced and considered the premaxilla to be in normal position. Latham suggested that a contributing factor in producing the projecting premaxilla was the shortening of the septomaxillary ligament which drew it forward.

Pruzansky and Friede (1975) roentgencephalometric data confirmed the existence of a true, rather than a relative, premaxillary protrusion. Metallic implants were inserted on either side of the PVS at the time of initial surgery and were followed up roentgencephalometrically. Although continuous growth was recorded in the PVS, there was a postsurgical decrease in the premaxillary protrusion. On the basis of both histology and cephalometrics, Friede (1973) concluded that traumatic surgery involving the PVS would be likely to contribute to the impaired midfacial

growth. This has been verified by Berkowitz's more recent follow-up studies of the effects of mechanical premaxillary retraction on profile development. He has observed similar midfacial problems of impaired growth (see Chap. 21).

Berkowitz speculated that severe pressure at the PVS would cause a hemorrhage followed by fibrosis and synostosis with cellular destruction, both acting to inhibit sutural growth. In contrast, a united lip across the protruding premaxilla exerts a more tolerant retrusive force within the PVS, acting well within its physiologic threshold level for slowing down bone growth but not totally inhibiting it.

Based on conclusions drawn from his clinical findings and those of Pruzansky and Friede (1975) research, Berkowitz rejects Latham (1969, 1973) "force of the cartilaginous septum via the septo-premaxillary ligament" as the cause of premaxillary protrusion. Enlow (1982) description of the naso-maxillary complex's growth is worth repeating:

The nasomaxillary complex is in contact with the floor of the cranium. The whole maxillary region in total is displaced downward and forward away from the cranium by the expansive growth of the soft tissues in the midfacial region. This then triggers new bone growth at the various sutural contact surfaces between the nasomaxillary composite and the cranial floor. Displacement thus proceeds downward and forward as growth by bone deposition simultaneously takes place in an opposite upward and backward direction.

McNeil (1950) mistakenly had stated that the detached palatal segments from the nasal septum are not only reduced in mass but also are not brought forward with the developing nasal septum.

This failure of the palatal segments to move forward with growth would lead to a retrusive midface with a Class III malocclusion (he did not want to blame the suggestions).

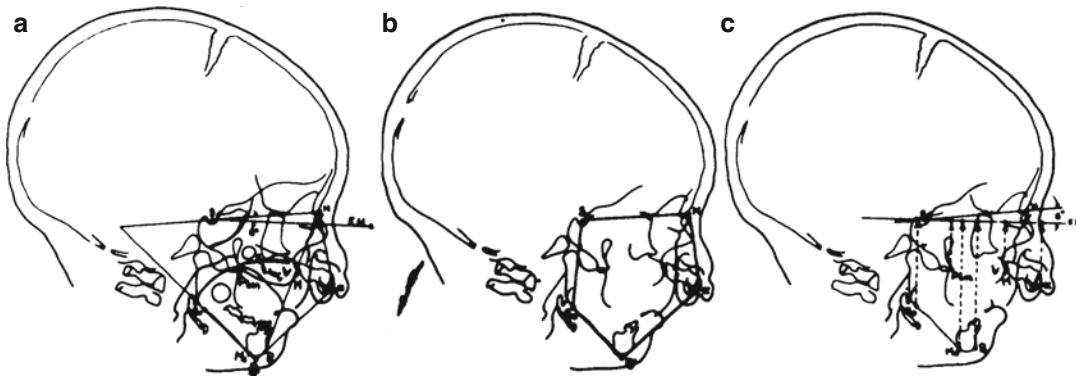


Fig. 7.5 (**a**–**f**) A method used to show serial changes in facial growth and development in CBCLP using sella for registration while superimposing on the anterior cranial base. Facial angles and landmarks (*upper left*): nasion (*N*), pogonion (*Po*), gonion (*Go*), sella (*S*), alpha (*a*) most anterior point on the premaxilla, menton (*Me*), constructed Frankfort horizontal (*FHc*), anterior point of the lateral palatal processes (*M*), pterygomaxillary fissure (*Ptm*). Facial polygon (*upper middle*) drawn connecting landmarks S-N-a-Po-Me-Go-S. Landmark points (*upper right*) projected to a constructed Frankfort horizontal line which is drawn 6° from the anterior cranial base (SN) at S. (**b**) An example of excellent facial growth. The cultural standard for a good aesthetic Caucasian face is a "flat" face, having an angle of facial convexity of approximately 180°. Most newborn noncleft faces have a relatively acute facial profile associated with relative retrognathia which usually flattens with growth

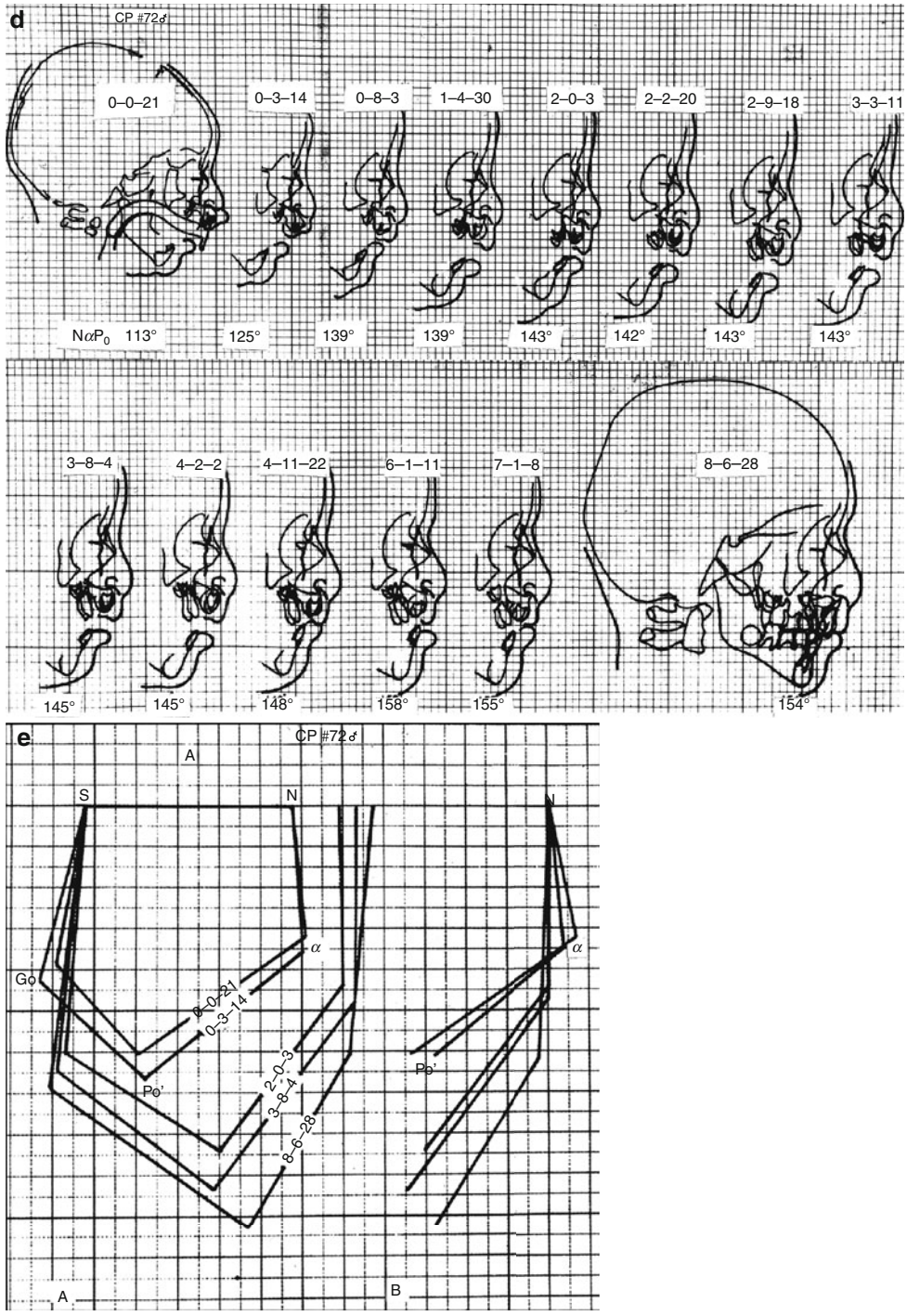


Fig. 7.5 (*continued*) (**d**) Serial lateral cephalometric tracings showing changes in the angle of facial convexity (NaPo) from 113° to 154° in 8 11/42 years. (**e**) Facial polygon of the case shown in Fig. 7.5d. Each polygon showing profile changes is superimposed on *SN* and registered at Sella. The midfacial protrusion at a had not increased after 2 years, whereas both the anterior cranial base (SN) and mandible had increased in size. The mandible grew forward and downward, flattening the facial profile. The timing of these growth changes is variable

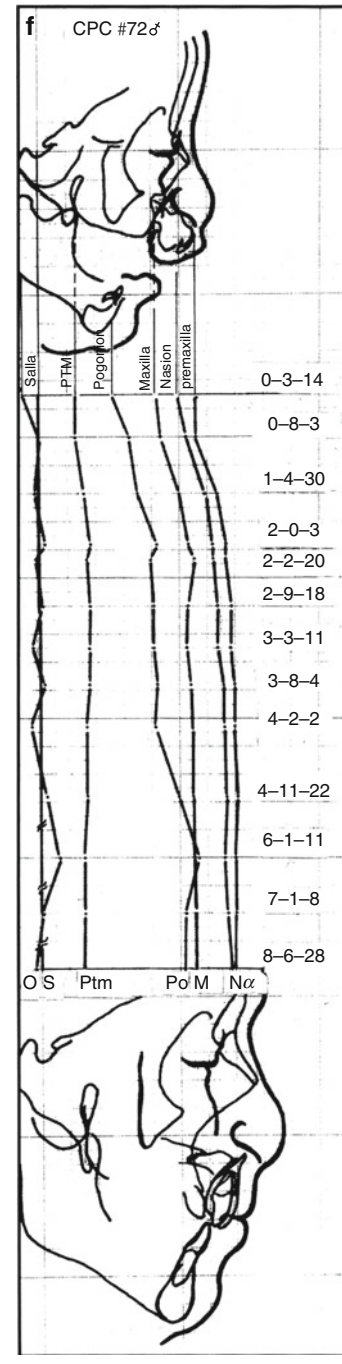


Fig. 7.5 (*continued*) (**f**) Projection of landmark points to the constructed Frankfort horizontal line drawn 6° from SN at S. This growth projection system also shows that the midfacial protrusion did not increase after 2 years, whereas the forward projection of the mandibular body increased markedly until 8 11/42 years of age. These are the main factors leading to the flattening of the facial profile. Good facial growth changes can occur early or late (From Millard 1980)

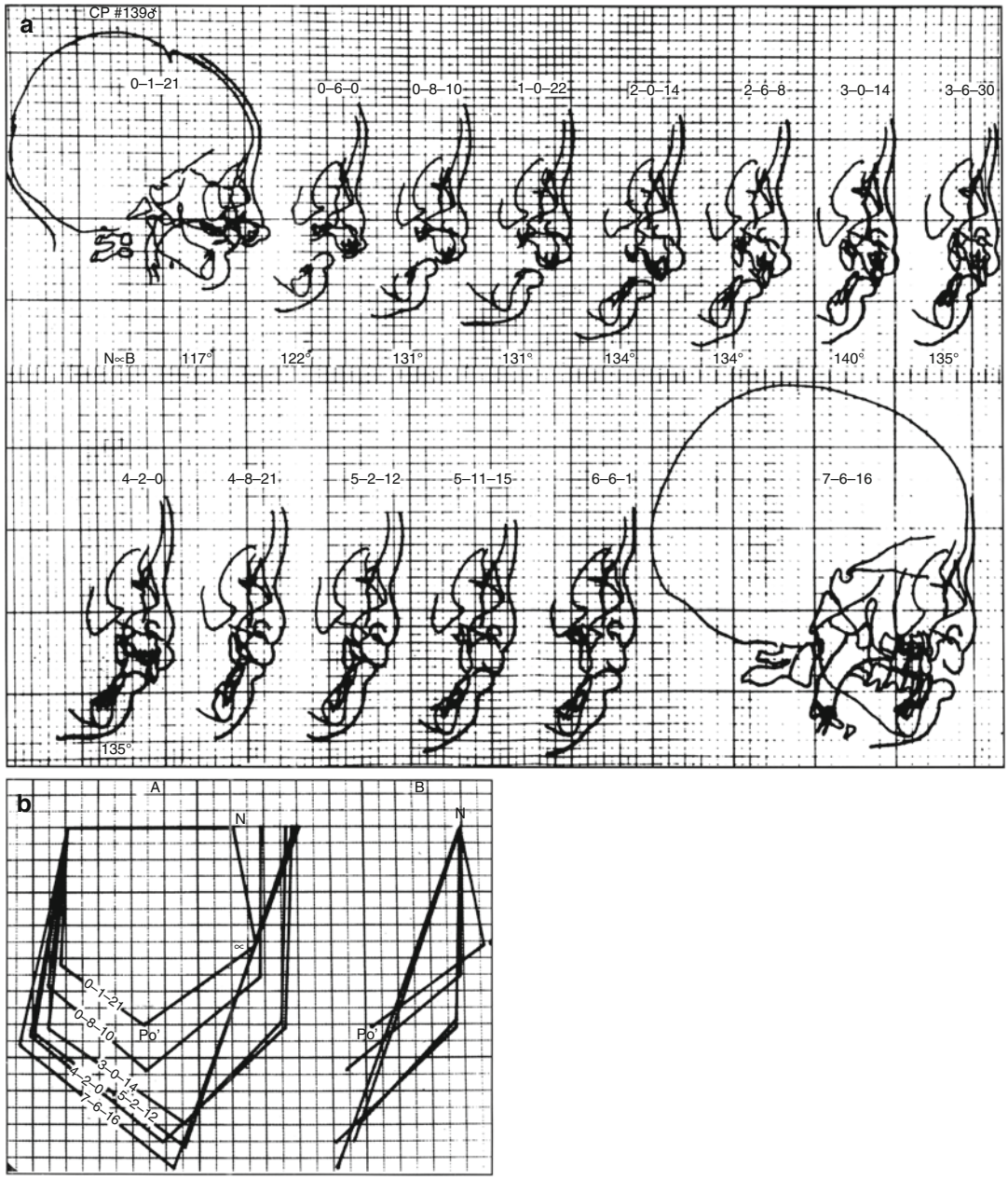


Fig. 7.6 (**a**, **b**) An example of "poor" profile growth changes. This evaluation is made when the acute facial profile seen at birth remains the same or worsens with time. (**a**) Serial cephalometric tracings showing poor facial growth leading to premaxillary excision. The acute angle of facial convexity remained the same after 2 years. This treatment plan should be abandoned at any age even with severe midfacial protrusion. In similar growing faces, some clinicians believe premaxillary surgical setback is preferable for psychosocial reasons prior to starting elementary school. (**b**) Superimposed polygons of case show a mandible that is growing vertically with very few horizontal growth increments. The premaxilla continued to grow forward although there was no forward growth between 3–0–14 and 4–2–0. Note the placement of the lower lip lingual to the premaxilla. The premaxilla was excised at 4–9. This case came from Pruzansky's records (From Berkowitz 1959)

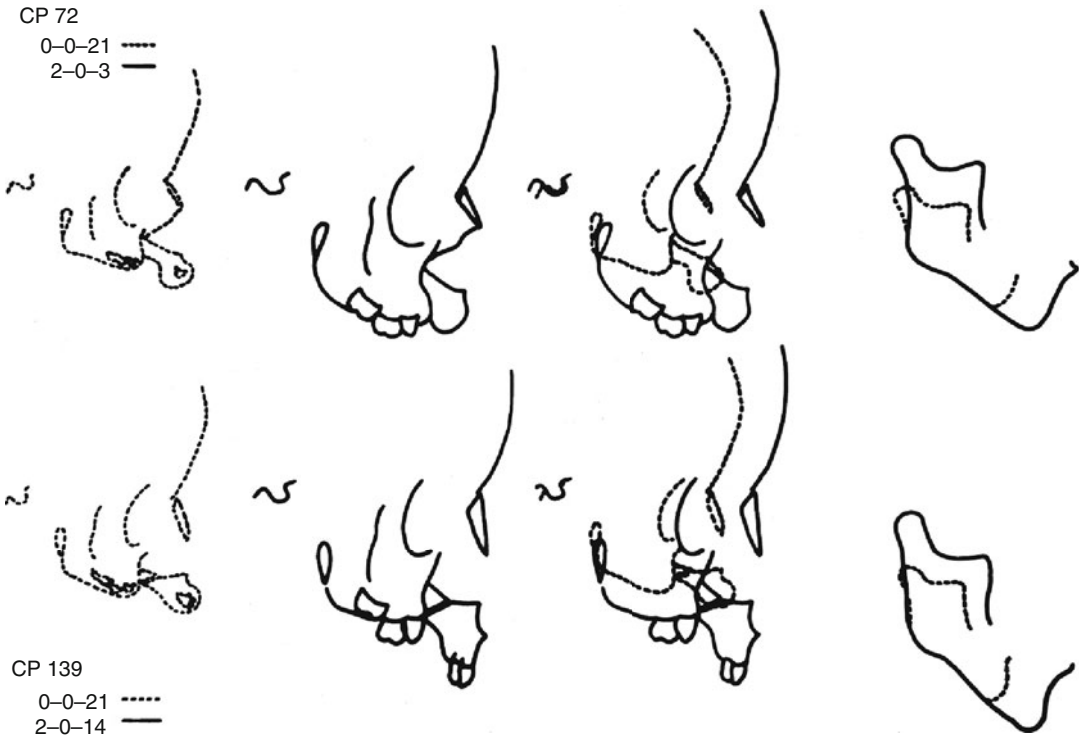


Fig. 7.7 The first 2 years of maxillary and mandibular tracings of the cases with good (see Fig. 7.5) and bad (see Fig. 7.6) facial growth are compared to show the reasons for these evaluations. CP-72: Good facial growth. In this case, the degree of premaxillary protrusion relative to the lateral palatal segments (the anterior cleft space) was markedly reduced with growth. Premaxillary protrusion relative to the anterior cranial base was reduced as well. CP-139: Poor facial growth. The degree of premaxillary protrusion relative to the lateral palatal segments remained the same, while the premaxillary protrusion relative to the anterior cranial base increased (Courtesy of S. Pruzansky). Comments: Although mandibular growth in the two cases was similar in degree, the superimposed polygons show that the vertical direction of growth of the "bad" grower's mandible and not its size was the determining factor for the changes in the angle of facial convexity (NaPo). Tension at PVS created by the lower lip positioned lingual to the premaxilla increased premaxillary growth

7.3 Facial Growth Studies Show That Midfacial Retrusion Is Not Predictable

Semb (1991) and Ross (1987) established that, in the cleft population, both the maxilla and mandible – not solely the maxilla – are retropositioned within the face. If McNeil (1950) belief that the bilateral cleft palatal segments are left behind in their growth was accurate, a greater proportion of the cases must show a Class III malocclusion of one or both sides. As previously mentioned, Berkowitz (1982) mixed cross-sectional study of the occlusion in CBCLP determined that the maxillary complex was not positioned posteriorly relative to the mandible. Also, a buccal crossbite is not a predictable outcome, as McNeil had suggested. Semb (1991) and Ross (1987) concluded that, although the midface and mandible are positioned posteriorly within the face, the maxilla is not retropositioned relative to the mandible and that McNeil's hypothesis that the maxilla in complete clefts of the lip and palate is retruded and needs to be brought forward was in error.

7.4 Long-Term Facial Growth Findings Show Class III Outcomes Are Not Predictable

Semb (1991) conducted a serial lateral and frontal cephalometric study of 90 cases from the Oslo archives with bilateral cleft lip and palate. Since 1962, the treatment procedure has involved uniting the lip and closing the hard palate cleft space in two stages. No presurgical orthodontics were utilized because the surgeon and Bergland, an orthodontist and director of the program, believed that any bilateral cleft lip can be closed without presurgical palatal manipulation. In the period spanning 1950–1960, a von Langenbeck procedure was performed to close the hard palate cleft between 3 and 4 years of age; after 1960, the timing of the closure was reduced to 18 months of age. Secondary alveolar bone grafting using cancellous iliac crest bone was performed prior to the eruption of the permanent canine teeth.

Twenty-five percent of the cases needed superior-based pharyngeal flaps, which were performed before the child started school. No orthodontics were utilized in the deciduous dentition. Protraction headgear was used in the mixed dentition in one third of the cases. Fixed retention was necessary in all cases. Semb's study showed that: (1) the maxilla progressively receded over time, (2) the mandible was retrusive with a steep mandibular plane with an increased gonial angle, (3) anterior lower face height was elongated and posterior facial height reduced, and (4) the facial growth pattern was notably different from the normal Bolton standards: (a) Male and female facial growth patterns were similar except that the males' linear dimensions were larger, (b) the prominent premaxilla would gradually realign in the preschool years, and (c) surgical premaxillary setback was never required. Berkowitz's unpublished serial cast cephalometric data support these statements (unpublished data).

Vargervik (1983) cross-sectional study of 51 males with BCLP treated with a variety of primary procedures (excluding premaxillary setback) showed profile values similar to those reported by Hellquist et al. (1983), Dahl (1970), Smahel (1984), Semb (1991), and Friede and Johanson (1977). The Oslo team's average for maxillary prominence and lower face height were slightly more favorable. Narula and Ross (1970), reporting cross-sectional data on thirty 6-year-old subjects and mixed longitudinal data on 34 subjects with BCLP treated conventionally without surgical setback and vomer flap, concluded the maxillary length reached normal values at 16 years of age.

In the Swedish sample followed longitudinally by Hellquist et al. (1983), similar facial convexity was also noted, although both the maxilla and mandible were reported to be slightly more prominent. The patients analyzed by Hellquist et al. (1983) had a two-stage lip closure, push-back palatoplasty, and, at an average age of 6, a delayed periosteoplasty.

Friede and Johanson (1977) reported facial growth in 13 Swedish children with bilateral clefts of the lip and palate (BCLP), five at age 7 and eight at age 10 years. The patients, who had had lip adhesion and vomer flap (without premaxillary setback) and velar closure with push-back, also exhibited facial convexity similar to the Oslo sample.

Friede and Pruzansky (1972), Bergland and Borchgrevink (1974) cephalometric reports of 27 North American children in three treatment groups who were followed to 17 years of age do, however, show some differences in comparison with the Oslo sample. Six subjects treated by early premaxillary setback and seven treated by late setback (3–8 years of age) had profile values similar to those reported by the Oslo group. However, 14 subjects with no premaxillary setback had average values significantly more convex than those reported for the Oslo and other samples. None of the North American subjects had had vomer flaps, and it is implied that a more convex facial profile will be obtained if vomerplasty is excluded from primary surgery.

Also, the negative growth affect of the surgical premaxillary setback may become evident at a later age.

7.5 The Vomer Flap: Good or Bad? Are All Vomer Flaps the Same?

Semb (1991) report on the longitudinal data of the Oslo group is critical of those who condemn the vomer flap. She states that the possible growth-retarding effect of a vomer flap has been discussed by several authors (Friede and Pruzansky 1972; Friede and Morgan 1976; Friede and Johanson 1977; Pruzansky and Aduss 1967; Blocksma et al. 1975; Delaire and Precious 1985, 1986; Friede et al. 1987; Molsted 1987; Enmark et al. 1990). Friede and Pruzansky (1972) observed more favorable growth in patients treated without a vomer flap; however, this is not a uniform finding in the comparative studies. Some clinical centers not using a vomer flap have shown results similar to those where a vomer flap has been utilized. The extent to which the observation of a growth-retarding effect of a vomer flap can be generalized remains in some doubt and may reflect other variations in sample composition or surgery.

The effects of a vomer flap on facial growth have also been considered by the Oslo CLP team (Molsted 1987) which reported the flap to be clinically insignificant. Only one patient in their sample of 90 cases exhibited any degree of maxillary retrusion where surgical maxillary advancement was judged necessary.

In the opinion of the Oslo team, a vomer flap provides the particular advantages of early separation of the nasal and oral cavities without artificial obturators, a low prevalence of symptomatic fistulae, an acceptable arch form, and a good foundation for mixed dentition alveolar bone grafting (Bergland et al. 1986).

Berkowitz's finding of serial complete bilateral cleft lip show results similar to those of the Oslo group. A modified vomer flap was used in conjunction with a von Langenbeck procedure. He believes that the negative effect of a vomer flap on midfacial growth and arch form, as described by Prysdo et al. (1974), has not been conclusively proven. Patients who have needed LeFort I advancement of only the lateral palatal segments have had very large anterior cleft spaces in the permanent dentition even after the posterior hard palate had been surgically closed at 18–24 months of age with a vomer flap. The premaxillae in these cases were in good overbite-overjet relationships prior to moving the palatal segments into a Class II relationship to close the missing lateral incisor(s) space. Other times, the lateral incisor spaces are left open.

7.5.1 External Elastics Attached to a Head Bonnet or Elastic Tape Strapped to the Cheeks (Fig. 7.9)

These force delivery systems will reduce premaxillary protrusion prior to lip surgery and only need to be utilized for approximately 1 or 2 weeks. Elastic forces exert the same backward pressure against the protruding premaxilla as a lip adhesion. There are no valid reasons to object to the use of external facial elastic traction with arm restrains to prevent its removal forces for 1–2 weeks prior to lip surgery.

7.5.2 Uniting the Lip (Figs. 7.8, 7.9, and 7.10)

In all complete clefts of the lip and palate at birth, the wide dislocation of the lateral palatal segments coupled with the protruding premaxilla can be gradually overcome by following a treatment protocol that allows the palatal segments to move into proper orientation by the application of the physiological forces of a united lip without the need for presurgical orthopedic treatment. Lip adhesion is followed by definitive lip surgery and observation of facial and palatal changes (Fig. 7.8). If necessary, later surgical premaxillary setback, or surgically moving the lateral palatal segments forward to close a very large persistent anterior cleft space, can be considered.

Clinicians who favor this procedure believe the degree of midfacial protrusion will decrease with facial growth under the influence of the midfacial growth-restraining forces of the united lip (Friede and Pruzansky 1972; Berkowitz 1959; Coup and Subtelny 1960; Pruzansky 1953; Vargervik 1983; Wolfe and Berkowitz 1983; Mazaheri et al. 1971; Bishara and Olin 1972; Aduss et al. 1974).

Findings by Bishara and Olin (1972) and Aduss et al. (1974) support the conclusions drawn from earlier facial growth studies by Berkowitz (1959), Friede and Pruzansky (1972), Handelman and Pruzansky (1968), and Vargervik (1983) that the protruding premaxilla tends to be molded back by lip pressure and can sometimes be aligned within the lateral palatal segments without resorting to neonatal maxillary orthopedics. If not, then the premaxilla in most cases can be aligned within the arch in the deciduous dentition using a fixed orthodontic quad helix appliance.

The use of lip adhesion within the first 3 months followed by a definitive lip revision, usually within the first 6 months of age, permits the surgeon to perform surgery in stages as the face changes. Logan's bow can be used to reduce the strain at the lip suture when the definitive lip surgery is performed. Surgeons who favor stage treatment procedure are willing to postpone obtaining a maximum early aesthetic result for what they believe will offer superior long-term benefits. The most severe unaesthetic premaxillary overbite in the deciduous and mixed (transitional) dentition, even with a buccal crossbite,

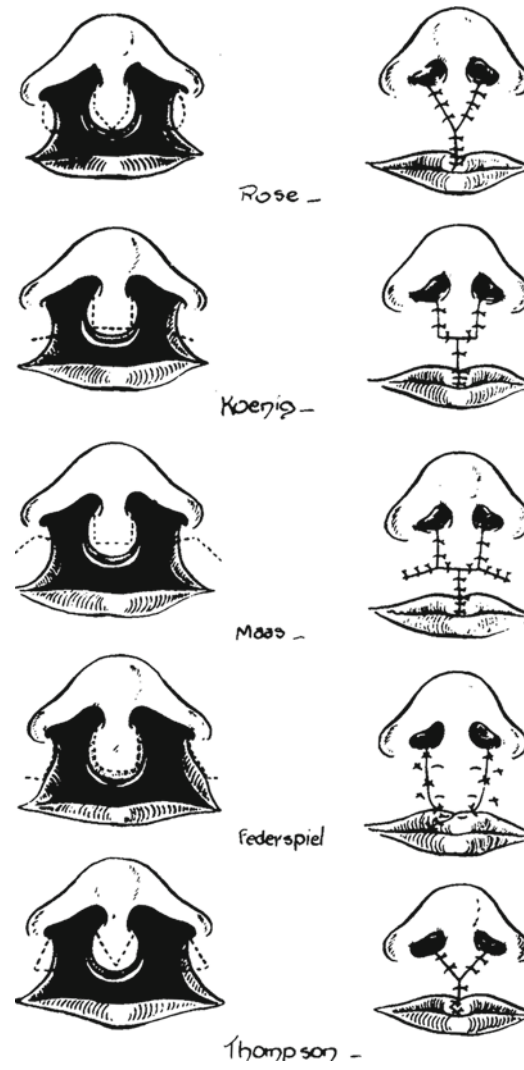


Fig. 7.8 Various surgical techniques used to unite the lip in BCLP. Experience has shown that the best results are obtained when the prolabium is used to construct the entire midportion of the lip (From Millard 1980)

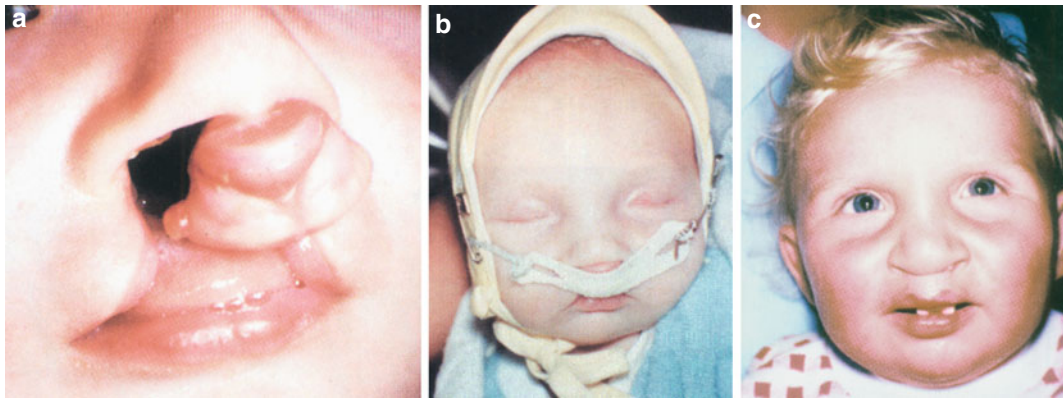


Fig. 7.9 (**a**–**c**) A head bonnet with an elastic strap (external facial traction) placed against premaxilla is an efficient and painless procedure to ventroflex the premaxilla prior to lip surgery. (**a**) At birth. (**b**) With head bonnet in place. (**c**) After lip repair. The head bonnet aids the surgeon in reducing tension at the lip suture site. The force generated at PVS is less than that created by using mechanical premaxillary retraction (see Chap. 20)

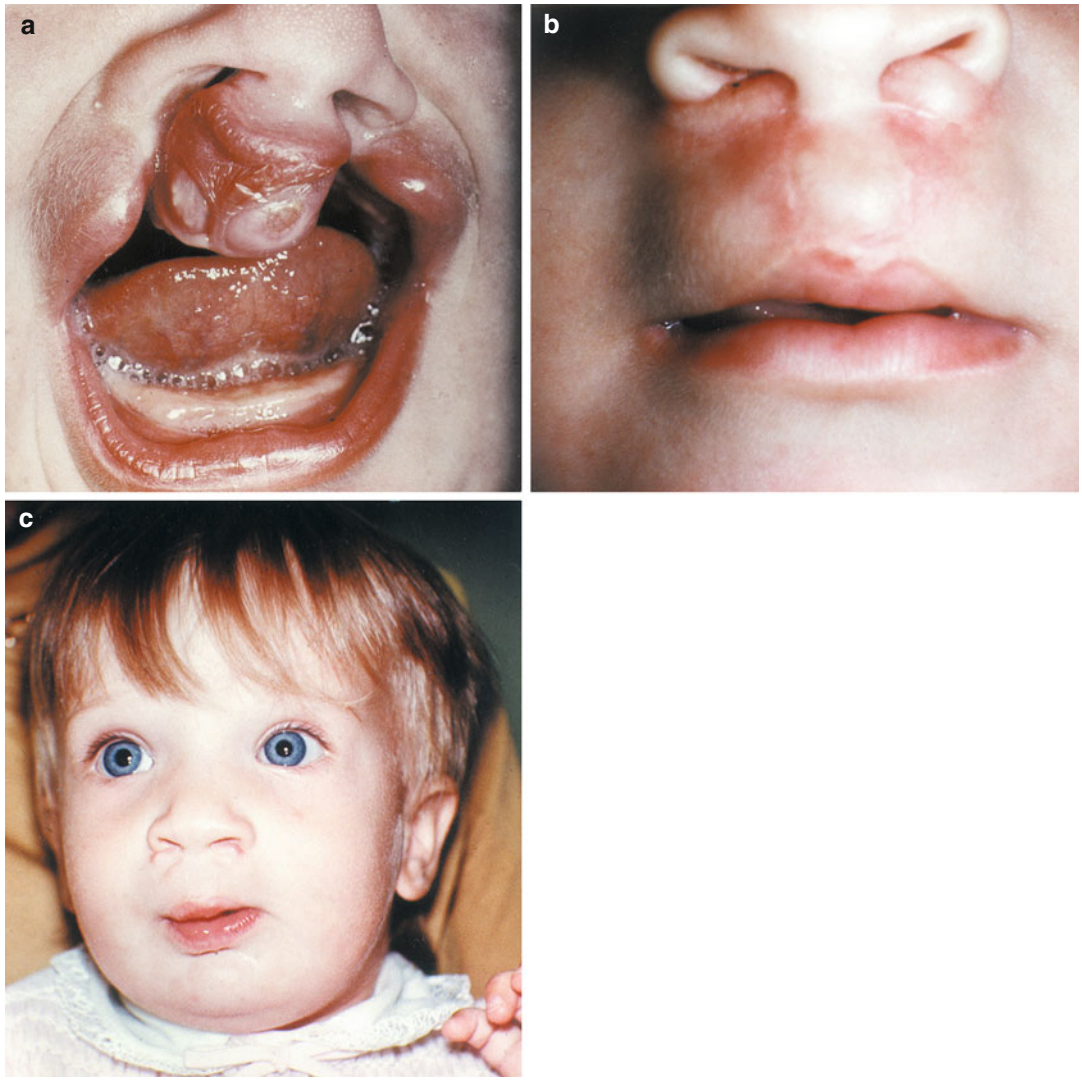


Fig. 7.10 (**a**–**c**) Use of the prolabium for the center portion of the lip. IBCLP. (**a**–**c**) Facial photographs at 20 days, 3 and 18 months. The banked forked flap (Millard): the tissue in the nostril is used to lengthen the columella at a later date. The procedure avoids going back to a good lip for tissue to reconstruct the columella

will not interfere with dental function or swallowing or have any long-term deleterious effects on speech and midfacial growth.

Serial facial photographs indicate that surgical procedures that use the entire prolabium for the central portion of the lip produce the best aesthetic results. A long tight lip is the predictable result when the lateral lip elements are brought together beneath the prolabium (see Fig. 7.10a, b).

7.6 **Profile Changes** (Figs. 7.14, 7.15, 7.16, 7.17, and 7.18)

Hanada and Krogman (1975), Berkowitz (1959), Bishara and Olin (1972), Boyne (1974), Vargervik (1983), Handelman and Pruzansky (1968), Friede (1977), Semb (1991), and Narula and Ross (1970) all have performed longitudinal studies of the changing soft tissue profile of bilateral cleft lip and/or palate (BCL/P) and reported that, with the

slow resolution of the protruding premaxilla with growth, the profile became more harmonious in appearance. In the Oslo study of BCLP, Semb (1991) concluded that early dentofacial orthopedics were not a necessary precursor to lip and palate closure in order to attain long-term positive profile changes.

Serial profile analyses of cast and cephalographs reported in Berkowitz's earlier studies and the serial studies presented in this text demonstrate that, during the first 2 years following surgery, the united lip exerts a posteriorly directed pressure force on the protruding premaxilla. He speculates that this force is exerted through the nasal septum to the premaxillary-vomerine suture, gradually retarding the forward and vertical growth of the nasal septum with the attached premaxilla. The growth of the lateral palatal segments does not appear to be affected by this force. Inhibition of midfacial growth coupled with forward and vertical growth of the upper face and mandible is responsible for the eventual flattening of the facial profile (Figs. 7.5 and 7.6).

Protrusion of the midface is only slightly reduced by the reduction-remodeling of the labile surface of the premaxilla associated with the eruption of the permanent incisors. Facial growth data suggest that the tendency toward mandibular prognathism can be considered advantageous, insofar as it reduces the earlier protrusion of the premaxilla. Comparison of the profile changes in children whose lips were united at 6 years with infants whose lips were united soon after birth showed less premaxillary protrusion for children whose lips were repaired in infancy. This suggests that the beneficial effect of lip repair is long-acting in that it continues to exert a restraint on the growth of the premaxillary-vomerine complex long after the lip is repaired (Friede and Pruzansky 1972; Berkowitz 1959; Vargervik 1983).

After the lip is united, there are no documented long-term benefits to using intraoral neonatal maxillary orthopedics to control the premaxilla's position relative to the lateral palatal segments (i.e., to inhibit the palatal segments from spontaneously moving together) nor is it important that the premaxilla be accommodated within the arch at this time. There is no evidence that overlapping palatal segments suffer growth inhibition. In most cases, proper premaxillary alignment within the lateral palatal segments can be easily achieved with orthodontics in deciduous or mixed dentition without resorting to neonatal maxillary orthopedics. The following cases show that orthodontics in the permanent dentition, in the absence of growth-inhibiting palatal surgery, and with a good or poor facial growth pattern eventually will lead to excellent facial aesthetics and dental function. Any resulting buccal crossbite can be easily corrected at 4 or 5 years of age, when the child is manageable in a dental chair, using fixed tooth-borne or even removable arch expansion appliances. The author strongly favors the use of fixed appliances for arch expansion and retention.

There are many occasions when the profile shows midfacial retrusion due to severe ventroflexion of the premaxilla. As long as during the mixed dentition gingivoperiosteoplasty has not been performed (see Chap. 20), the premaxilla can easily be brought forward orthodontically and held in place until alveolar bone grafting is performed. Retention of the corrected maxillary arch is still necessary until orthodontics are completed (Fig. 7.11).

7.6.1 Why Some Premaxillae Continue to Project Following Lip Repair and Others Do Not

The cases being presented will show that there are a number of treatment planning facial factors. For example, the patient's facial growth pattern and the amount of palatal osteogenic deficiency are beyond the control of the surgeon. The integrated growth of the entire face is important to resolve the profile deformity. When the end result is unfavorable – that is, the facial profile remains highly convex – the premaxilla and the body of the maxilla usually have grown forward with limited forward mandibular projection or tension has been created at the PVS, causing further premaxillary protrusion.

Facial growth studies by Handelman and Pruzansky (1968) and by Berkowitz (presented

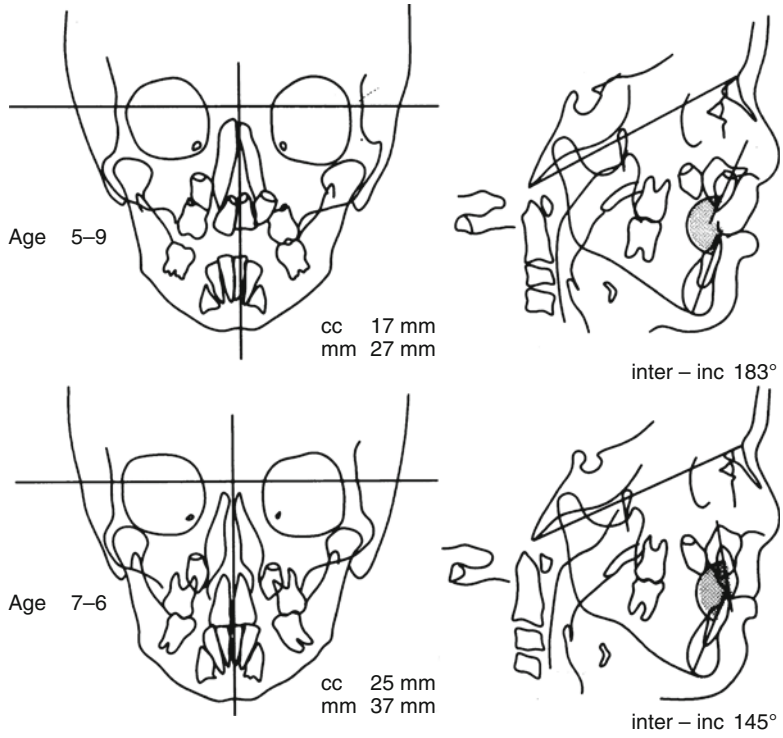


Fig. 7.11 Before and after line drawings demonstrating the use of orthopedics for the correction of a malocclusion in a BCLP. 5–9 Frontal and lateral tracings of a child with a bilateral cleft lip and palate before orthopedic treatment. Intracuspid width was 17 mm, intramolar width was 27 mm, and the interincisal angle was 183°. 7–6 After premaxillary advancement and buccal expansion, the intracuspid width changed to 25 mm, intramolar width to 37 mm, and the interincisal angle to 145°. All changes in measurements were dependent on the bodily (orthopedic) movement of the bony segments and not on the movement of teeth (orthodontics). After the palatal segments are properly aligned, the movement of teeth will follow. While waiting for the permanent teeth to erupt, the new arch form needs to be retained with a fixed appliance

in this chapter) have shown that, in highly convex facial profiles with a moderately protrusive premaxilla and a recessive mandible, the lip musculature usually is hypertonic and positioned over the upper incisor teeth, resulting in severe premaxillary ventroflexion. In mesognathic faces with isotonic or hypotonic lip musculature and a severely projecting premaxilla, the lower lip may be positioned lingual to the premaxilla and between the upper and lower incisors, creating an anterior component of force at the premaxillary-vomerine suture (PVS). This may cause additional bony growth at the suture, leading to greater premaxillary protrusion with a normal maxillary incisal axial inclination. The lower incisors may be tipped lingually (Figs. 7.11 and 7.12).

The serial record present in the case reports to follow will show that it can be anticipated that in most cases (53 out of 59 CBCLP; S Berkowitz, unpublished data), good faces will have developed at adolescence. Figure 7.14 shows serial computer-generated CBCLP palatal cast outlines which were superimposed on the palatal rugae and registered on the vomer. All of these cases had the palate closed between 18 and 24 months using a von Langenbeck with modified vomer flap (it was readily observed at every age period).

This illustration clearly demonstrates that the premaxilla is in the same place in the palate at adolescence as it was at birth. It is retained in place by the surrounding facial musculature in most cases. There are exceptions, usually dependent on the balance of the facial musculature.

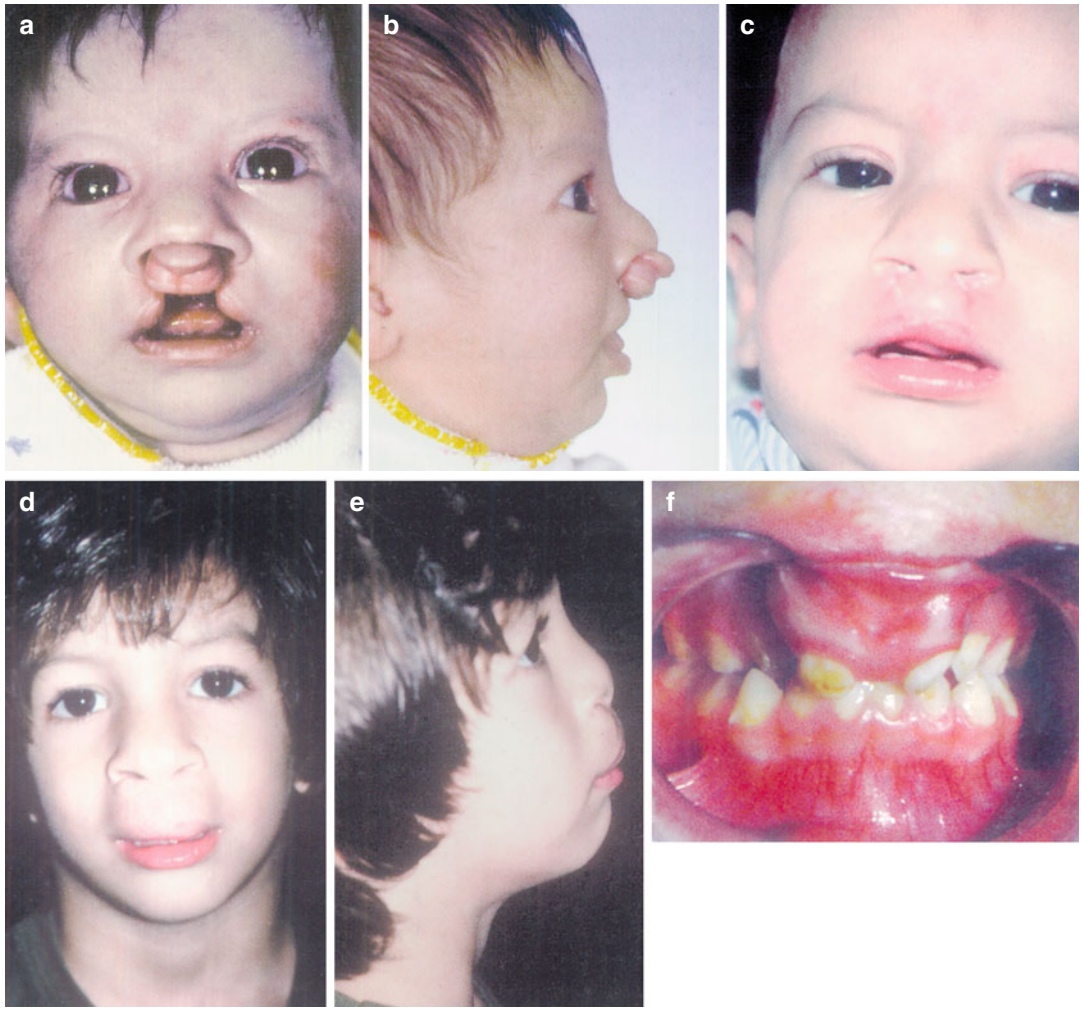


Fig. 7.12 (**a**–**f**) Millard's surgical lip procedure uses the prolabium to construct the entire midportion of the lip over the protruding premaxilla. This case was selected to show an example of upper lip protrusion that can result due to a severely protruding premaxilla. This case and others presented in this chapter will show how the face improves with time, that is, facial growth

7.6.2 Dental Occlusion

Analyses of the dental occlusion of many patients with complete bilateral clefts of the lip and palate at 6 years of age rarely show retruded maxillary shelves (i.e., a Class III relationship) (Berkowitz 1959). The buccal teeth are most often in a Class I or II relationship. A Class III relationship exists only if the maxilla is retrusive due to severely retarded growth caused by traumatic surgery and/or the patient's phenotype. The mandible is rarely prognathic at this age. In many cases, due to palatal osteogenic deficiency and severe dental crowding in the maxillary arch, some teeth may need to be extracted. This may or may not have to be done in the mandibular arch. There is conclusive evidence to suggest that the cleft palatal shelves may be deficient in mass, but there is no evidence that they have lost their growth impetus from having been detached from the growing nasal septum.

Until proven otherwise, the surgeon should have confidence that the severe facial convexity seen at birth can diminish with time as the united lip restrains the forward development of the mid-

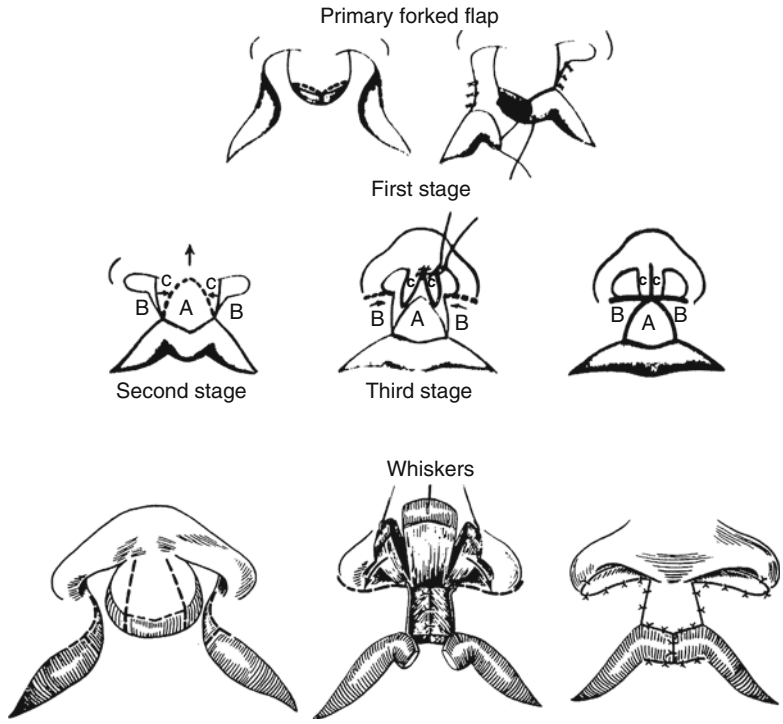


Fig. 7.13 In 1956–1957, Millard designed a secondary forked flap for columella lengthening. He believed at times it is best to take a little tissue from the prolabium and the lip and store it in the area for later use in reconstructing the columella. The forked flap was originally designed as a secondary procedure; however, it can be used as a primary procedure when the columella is extremely short and the prolabium is of reasonable size. *Top*: In the primary forked flap, the fork flap is elevated and advanced into the columella with release of the nasal tip. *Bottom*: The whisker fork flap involves joining the lip muscle and banking the fork. The alar bases are joined together in the midline, and the forks partially tubed on themselves and led into the transverse incisions between the lip and alar bases, whisker fashion. This surgery is delayed several years. Millard likes this procedure best of all

face, while the mandible increases in size and is positioned more downward and forward relative to the growing anterior cranial base and midface.

7.6.2.1 After Birth

In general, no presurgical orthopedics were provided other than the use of a head bonnet. In complete bilateral cases only, an external facial elastic may be utilized to ventroflex the protruding premaxilla to reduce tension at the surgical site when utilizing lip adhesion. External traction is usually not necessary in incomplete clefts of the lip and palate.

The Forked Flap (Fig. 7.13)

All of the bilateral cases were treated with Millard's forked flap (Millard 1978a). It was designed to lengthen the columella as a primary or secondary procedure. Advantages include:

- Release of depressed nasal tip
- Lengthen the short columella
- Reduce an unattractive wide prolabium
- Revision of bilateral lip scars
- Reduction of flaring alar bases

Millard (1978a, b) suggests that the primary fork flap procedure is not appropriate for all bilateral cleft cases. Whether to use the procedure depends on: (1) The position of the premaxilla. It should not be used when the premaxilla is severely protruded. (2) The size of the prolabium. The width of the prolabium determines whether the flap is possible, and the vertical length indicates the amount of columella lengthening available. (3) Columella length. This discrepancy must be measured not only in actual length in millimeters

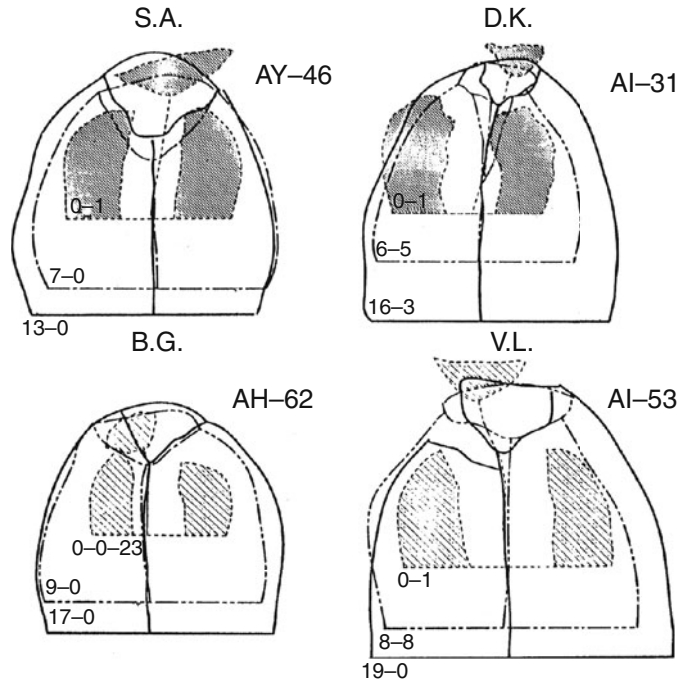


Fig. 7.14 Computerized digital tracings of the perimeter of the outline of CBCLP palatal casts superimposed on the palatal rugae and registered on the vomer. Palatal cleft closure between 18 and 24 months using a von Langenbeck and vomer flap. Note that the premaxilla is held in place, while the palate grows in all directions; posterior palatal growth occurs to accommodate the developing molars. None of these cases were treated with presurgical orthopedics

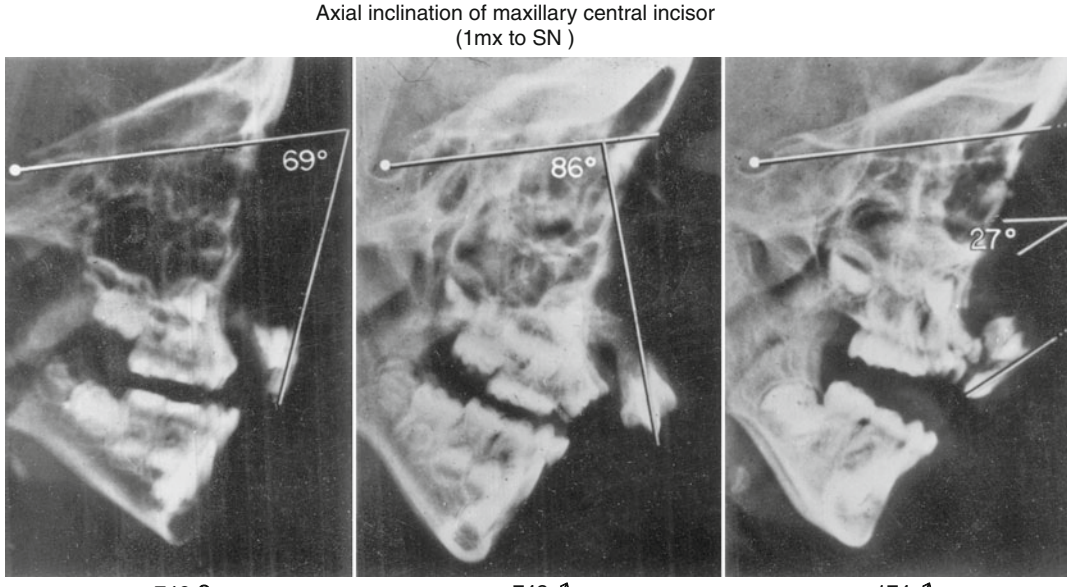


Fig. 7.15 In spite of the lip surgery performed, the position of the premaxilla in the anterior dentition is determined by the facial pattern and tonicity of the surrounding facial musculature. Case #746: Protruding premaxilla surrounded by hypotonic lip musculature. Case #742: Retrognathic mandible with protrusive premaxilla. The lower lip is positioned between the upper and lower incisors creating a forward force at the PVS. The resulting increase in tension at the PVS stimulates additional growth and causes increase in inclination of the upper incisors. Case #174: Retrognathic mandible with less premaxillary protrusion hypertonic lip musculature creating severe lip pressure which ventroflexes the premaxilla. Variations in premaxillary position are unpredictable (Handelman 1968)

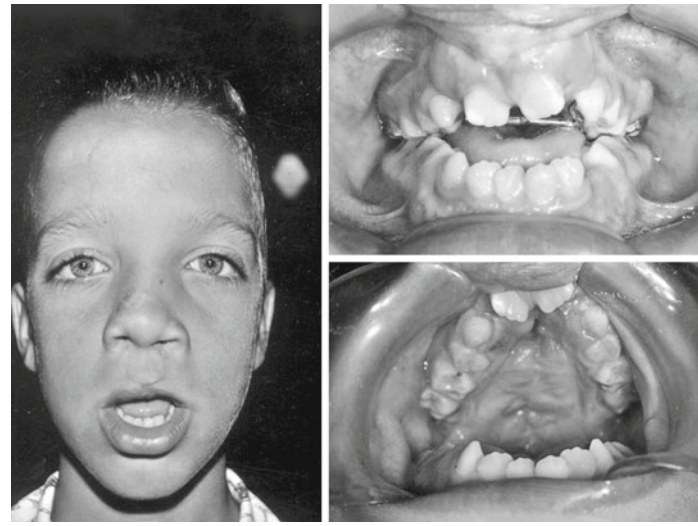


Fig. 7.16 Early premaxillary surgical setback and palatal cleft closure at 7 months of age. The lateral lip elements were brought together below the prolabium. Results at 7 years of age: (1) long tight upper lip, (2) anterior open bite due to failure of the premaxilla to descend with the palate, and (3) the surgery (pushback) used to close the palatal cleft at 7 months obliterated the vault space interfering with tongue posture. The tongue is, therefore, being carried forward with the tongue tip protruding, preventing the incisors from reaching the occlusal plane

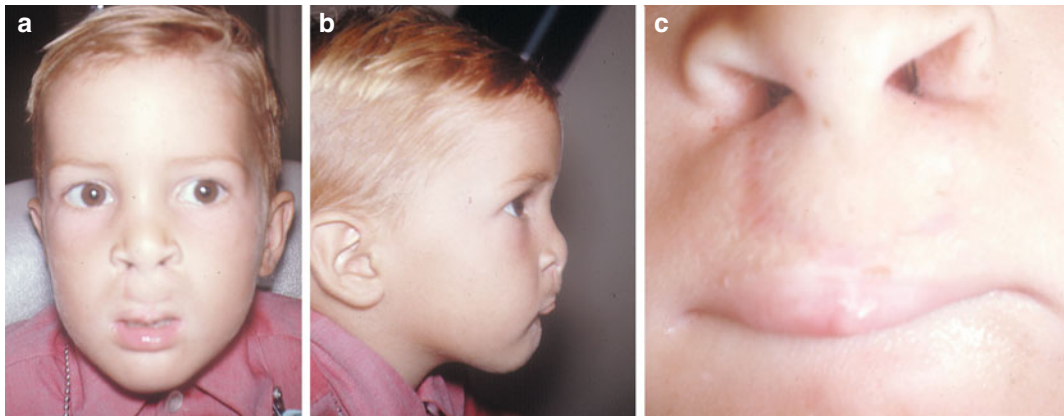
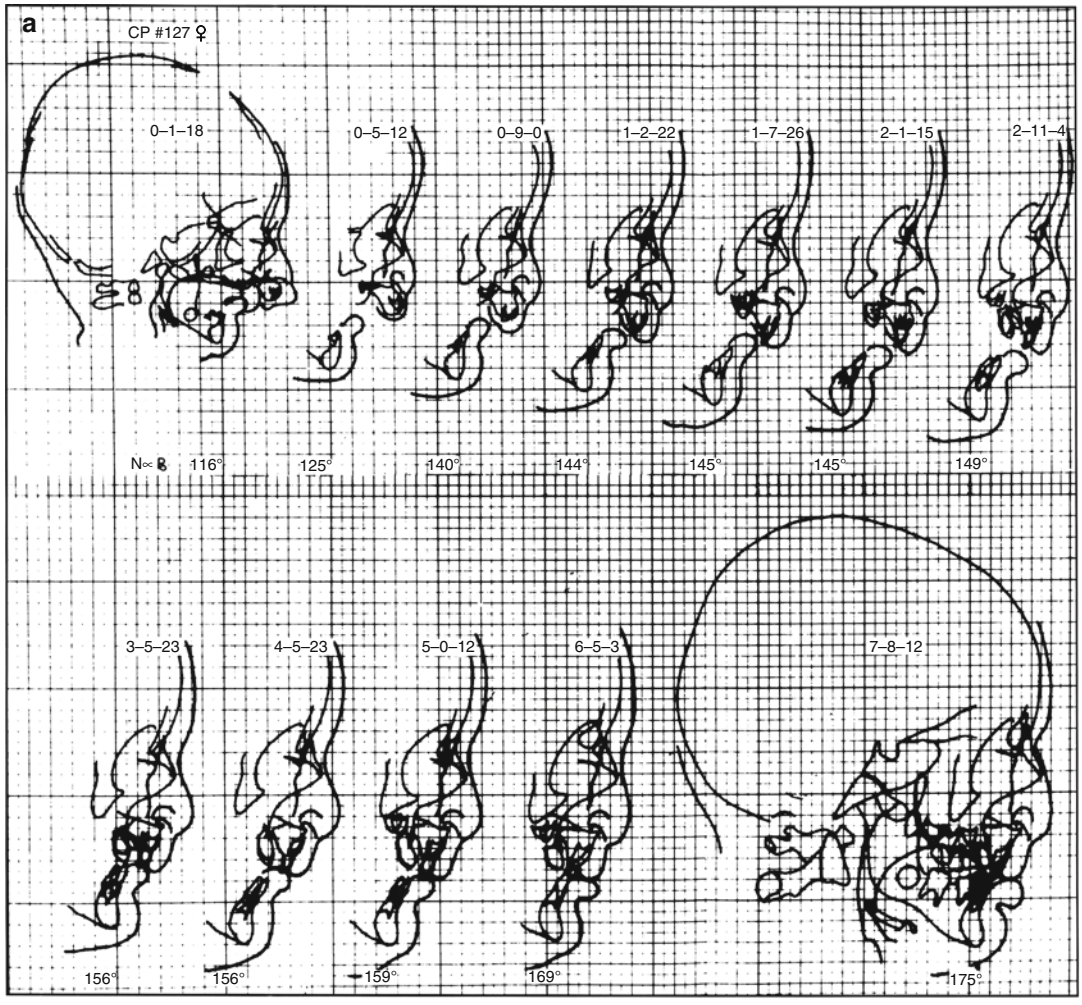


Fig. 7.17 (**a**–**c**) The lateral lip elements were brought below the prolabium creating a long tight lip

Fig. 7.18 (**a**, **b**) Good facial growth pattern with hypertonic lip musculature. a Lateral cephalometric tracing shows a gradual increase in the angle of facial convexity (NaPo) from 116° to 169° with the greatest change (159–169°) occurring between 5 and 6 years, 5 months of age. At 5 years, the axial inclination of the anterior teeth is vertical with an appreciable incisor overjet. However, at 6 years, 5 months, the premaxilla is palatally inclined, placing the upper and lower incisors in a tip-to-tip relationship. This usually occurs with a relatively retrognathic mandible when the lower lip also overlaps the upper incisors, increasing the muscle pressure against the premaxilla. If the lower lip is positioned between the upper and lower incisors, the premaxilla with its incisors will flare. (**b**) Superimposed polygons show a very small mandible with a protruding premaxilla at 0–1–18. There is a marked anterior growth of the anterior cranial base (*N*) and, together with the mandible's vertical and horizontal growth changes, a reduction in the angle of facial convexity. Note that there is very little change in premaxillary protrusion within the face between ages 1–2–22 and 7–8–12


a
CP #127 ♀
0–1–18
0–5–12
0–9–0
1–2–22
1–7–26
2–1–15
2–11–4
N∝ 8
116°
125°
140°
144°
145°
145°
149°
3–5–23
4–5–23
5–0–12
6–5–3
7–8–12
156°
156°
159°
169°
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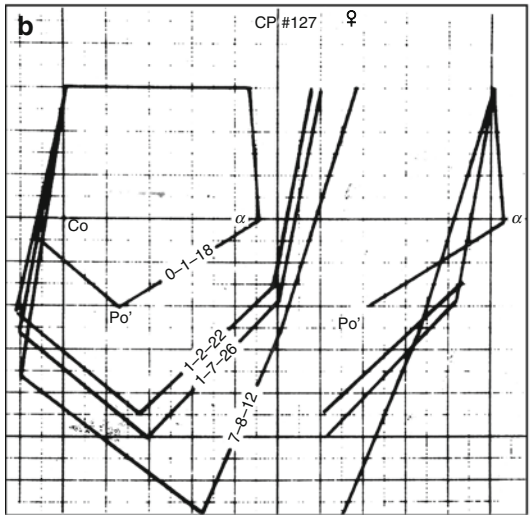
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of the columella, but an estimation of the patient's desired final length must also be made.

There are occasions where the forked flaps from the prolabium are banked with a subalar incision, whisker fashion. The alar base is not advanced medially in an attempt to leave a subalar gap in which to store the forks. Millard delays shifting of the banked forked flap into the columella for several years (from 6 months to 6 years).

7.6.2.2 In the Deciduous Dentition (3–6 Years of Age)

The purpose of treatment at this age is to align the palatal segments in order to obtain a more normal contour to the alveolar ridge, reshape the palatal vault, and provide a more symmetrical foundation for the support of the lips and nose. These changes are possible because orthodontic treatment in cleft palate allows for the movement of palatal segments when using a von Langenbeck procedure in addition to altering the position of the teeth within the alveolus. Because the roof of the mouth is also the floor of the nose, realignment of the palatal processes not only produces desirable alteration in the contour of the palate but also induces similar changes within the floor of the nose.

Most clinicians agree that nontraumatic conservative surgery will not solve all problems for all complete bilateral cleft lip and palate cases. Although buccal crossbites can be corrected in the deciduous dentition, some advocate orthodontic/orthopedic repositioning of the premaxilla at a later age (in the mixed dentition prior to or after secondary alveolar bone grafting) when the facial growth pattern leaves no alternative and dictates that it is the procedure of choice. Some orthodontists prefer to do this in the permanent dentition.

7.6.2.3 Mixed Dentition (6–11 Years of Age)

When a child with a cleft starts elementary school, even if the protruding premaxilla still extends far forward of the lateral palatal segment creating a severe convex facial profile, a surgical premaxillary setback should be considered solely for aesthetic reasons because there can be long-term deleterious effects. This decision is a difficult one. One is damned if one does or doesn't! One has to balance the child's social and psychological needs with long-term facial developmental factors.

In the last 20 years, many reports published with improved documentation were critical of surgical premaxillary setback performed in the newborn period, or even in early adolescence prior to the prepubertal growth spurt. Fortunately, these criticisms led to abandonment of this procedure (Vargervik 1983).

Orthodontics applied during the mixed dentition, in preparation for or after secondary alveolar bone grafting (the placement of bone in the alveolar cleft after the first year of age), can aid in reducing the premaxillary overbite and maintaining the premaxilla within the lateral palatal segments. Eliminating the alveolar cleft permits the unerupted teeth (lateral incisors and cuspids) adjacent to the cleft to erupt or be moved into proper position.

In Berkowitz's experience, a protruding premaxilla – with or without a large anterior cleft space – at 6–7 years of age does not signify that the same unpleasant aesthetic conditions will persist into adolescence. After the pubertal facial growth spurt, when the facial convexity is markedly reduced because of increased mandibular and upper facial growth, facial aesthetics will be greatly improved in most patients.

There is strong psychosocial pressure on clinicians to improve facial aesthetics as soon as possible, even when the premaxilla cannot be retropositioned in the absence of an anterior cleft space. For this reason, many unsuccessful attempts have been made to surgically or mechanically set the premaxilla back at an early age in the hope of preventing an unaesthetic midface at a later age, believing the midfacial growth would still be normal. At this difficult period, every effort should be made to convince both the parents and the child, using before and after photographs, that facial aesthetics will greatly improve as the face grows and that the long-term benefits far outweigh any early improvements at the expense of future good facial growth.

7.6.2.4 At Adolescence

If a poor facial growth pattern at adolescence with a large anterior cleft space leads to a protruding premaxilla set well forward in the facial profile and the mandibular incisors, the premaxilla can be surgically set back. A secondary alveolar bone

graft is simultaneously performed to close the remaining anterior cleft space. A fixed palatal retainer or a heavy labile arch wire must be utilized for at least 2 months as a supporting splint for the three segments during the healing period.

There are cases in which both buccal segments are in a Class I relationship, a large anterior cleft space is present, and the premaxilla is missing one or both lateral incisors, yet it is in an ideal overjet and overbite relationship. If the surgeon believes that the anterior cleft space is too large to be successfully closed by a secondary alveolar bone graft, the only remaining option is to surgically advance one or both palatal segments, placing the buccal teeth in a Class II relationship with at least one, but sometimes both, of the cuspids in the lateral incisor space. This usually necessitates the surgical increase in posterior transpalatal width. A secondary alveolar bone graft is performed at the same time (Posnick and Tompson 1993).

There are too many variations of orthodontic problems to review each of the treatment plans separately. However, in most cases, a common treatment sequence is usually followed. First, the posterior palatal crossbite needs to be orthopedically corrected in the deciduous or mixed dentition using quadhelix mechanics. Reducing maxillary arch crowding by tooth extractions may be necessary due to the lack of palatal osteogenic tissue. Achieving ideal anterior teeth aesthetics often requires that the lost lateral incisor spaces be recovered for the eruption of impacted lateral incisors or cuspids. In most cases, establishing room for four maxillary incisors by expansion and slight incisor advancement is the ideal treatment. A slightly retrusive maxilla with mild crowding may require the advancement of the upper jaw using protraction forces for at least 6–12 months to obtain an ideal anterior overbite-overjet relationship. In some instances, the extraction of one lower incisor to correct the mandibular anterior arch will be necessary to reduce an anterior crossbite.

In all cleft types, involving the lip and palate, orthodontic surgical treatment decisions are often dictated by the relationship of the premaxilla to the lateral palatal segments and the facial growth pattern (i.e., whether the face is prognathic, mesognathic, or retrognathic). For example, if a slightly retruded maxilla and a retrognathic mandible coexist, it may be necessary to surgically advance the lower jaw with or without doing the same to the maxilla. In most cases, orthopedic protraction to move the maxilla should be considered first.

If one or both maxillary buccal teeth are in a Class II relationship and one or both lateral incisors are absent, it is advantageous to move one or both cuspids into the lateral incisor space after secondary alveolar bone grafting. Eruption of the maxillary cuspids with its supporting alveolar bone usually closes off any remaining anterior cleft space. In very rare instances, when a large anterior cleft space exists and there is insufficient mucoperiosteum to close this space and a good anterior overbite-overjet relationship exists, the treatment of choice is to advance one or both palatal segments to make contact with the well-positioned premaxilla. A secondary alveolar bone graft can be performed at the same time (Posnick and Tompson 1993) (Fig. 7.59).

After the pubertal facial growth spurt, the once protruding premaxilla usually is no longer an aesthetic problem. Not surprisingly, at this age, the midface may be retruded even if the premaxilla was not surgically or mechanically set back and especially if there is good upper and lower facial growth. Treatment now needs to be focused on advancing the midface and/or the premaxilla using either orthopedic forces from a protraction facial mask or a maxillary osteotomy (LeFort I advancement).

In 30 years of clinical experience, surgical premaxillary retropositioning was performed only twice and only for aesthetic demands. It was never necessary to reduce its protrusion at the adult stage. Facial growth (time) ultimately is the surgeon's and orthodontist's best friend – and a most trenchant critic.

7.6.2.5 Retention

Permanent retention of the maxillary arch form in all clefts of the lip, alveolus, and palate is sometimes necessary even after secondary alveolar bone grafting, depending on the extent of transpalatal scarring and lip-cheek pressure. It requires either a removable palatal prosthesis or preferably a fixed bridge spanning the cleft. Arch collapse with a return of buccal and/or anterior crossbite can occur rapidly in the presence of extensive lip and transpalatal scar tissue because scar tissue has limited stretch capability.

7.7 The Following Case Studies Represent Conservative Surgical and Orthodontic Treatment Sequence

Cleft lip and cleft palate defects offer a degree of habilitation that is not available for many equally serious congenital or acquired handicaps. Achieving this potential requires careful planning and skillful execution of treatment that respects individual growth patterns (Table 7.1). The following cases were selected to show various treatment procedures, some of which were unsuccessful but nevertheless have teaching value because they reflect on physiological principles (Figs. 7.19, 7.20, 7.21, 7.22, 7.23, 7.24, 7.25, 7.26, 7.27, 7.28, 7.29, 7.30, 7.31, 7.32, 7.33, 7.34, 7.35, 7.36, 7.37, 7.38, 7.39, 7.40, 7.41, 7.42, 7.43, 7.44, 7.45, 7.46, 7.47, 7.48, 7.49, 7.50, 7.52, 7.52, 7.53, 7.54, 7.55, 7.56, 7.57, 7.58, 7.59, 7.60, 7.61, 7.62, 7.63, 7.64, 7.65, 7.66, 7.67, 7.68, 7.69, 7.70, 7.71, 7.72, and 7.73, Tables 7.2, 7.3).

Table 7.1 Timing and sequencing of surgical-orthodontic treatment (conservative)

Age	Orthodontics	Surgery
After birth	CBCLP external elastics (off head bonnet) over protruding premaxilla – no obturator	
3–4 weeks		Lip adhesion
6 months		Millard forked flap
18–30 months		von Langenbeck[a] (simultaneous closure of the hard and soft palate)
4–5 years	Correction of buccal crossbite only using a fixed quad helix palatal expander	
5–7 years	Fixed palatal retention	
7–8 years	Align anterior teeth prior to secondary alveolar bone graft (SABG)	Secondary bone graft using cranial or iliac crest bone
9–13 years	Full-banded treatment with or without maxillary protraction (Delaire face mask)	Nasal tip revision
13–17 years	Full orthodontics. Evaluate need for surgical orthodontics	Maxillomandibular surgery[b]
	(Distraction osteogenesis or Lefort I)	Nasal-lip revision
17–18 years	Postsurgical orthodontics followed by prosthetics	Nasal-lip revisions

[a]Timing of palate closure depends on the width of the cleft space relative to the adjoining palatal surface area. Anterior cleft is left open.

[b]Maxillomandibular surgery is usually performed earlier for females (around 15–16 years of age) than males whose surgery is performed during the summer prior to the senior high school year. This allows for maximum facial growth changes and leaves enough time to perform postsurgical orthodontics and prosthetic treatment.

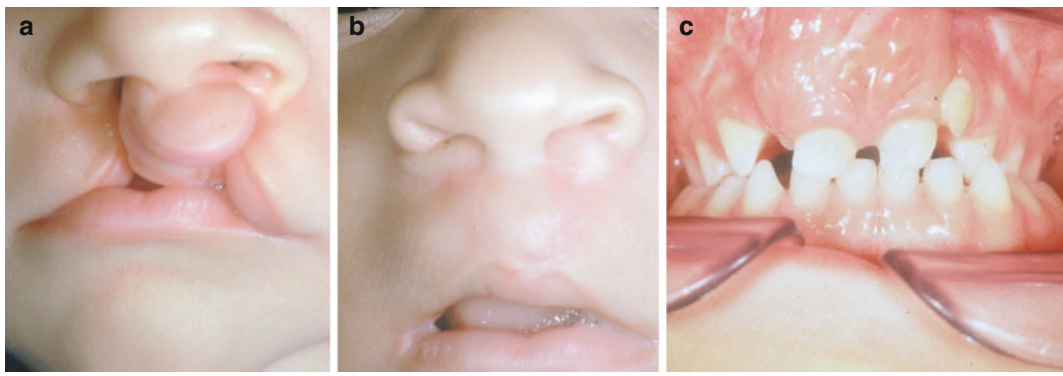

Fig. 7.19 (**a**–**i**) Case TM (WW-9) demonstrates excellent facial and palatal growth changes in an incomplete bilateral cleft lip and palate treated conservatively (no PSOT). (**a**) Newborn. (**b**) At 7 months after lip closure. (**c**) 4 years, 2 months. Note buccal crossbite on the left side. The crossbite was corrected by 4 years, 7 months using a fixed palatal expander

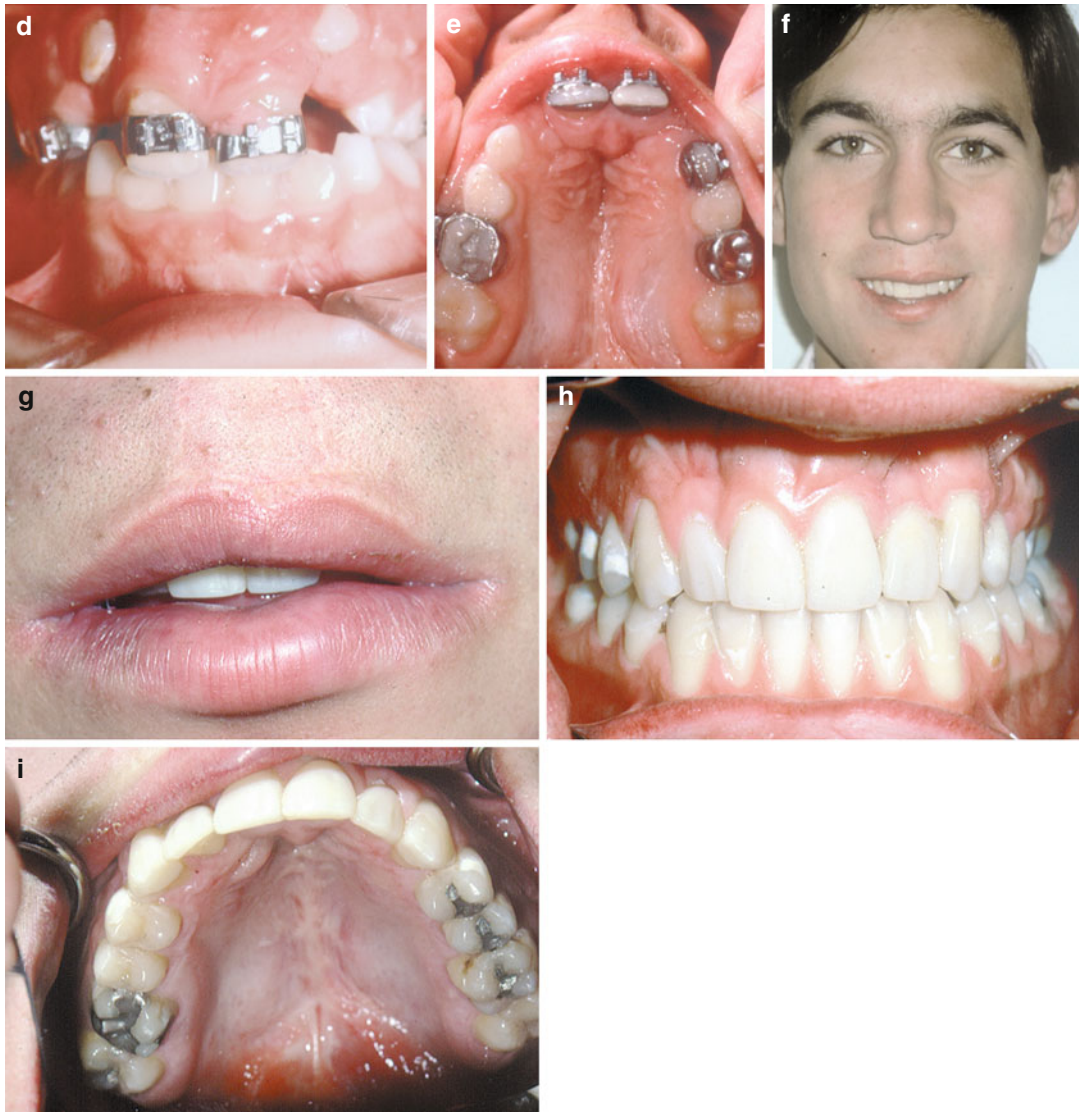

Fig. 7.19 (*continued*) (**d**) At 8 years after the central incisors were aligned and a secondary alveolar bone graft was performed at 7 11/42 years. Note that lateral incisors are erupting through the graft. (**e**) Palatal view at 7 11/42 years. (**f**) Full face at 17 years. (**g**) Close-up of lip. (**h**) Occlusion at 17 years after orthodontics. (**i**) Palatal view showing good vault space with minimal scarring

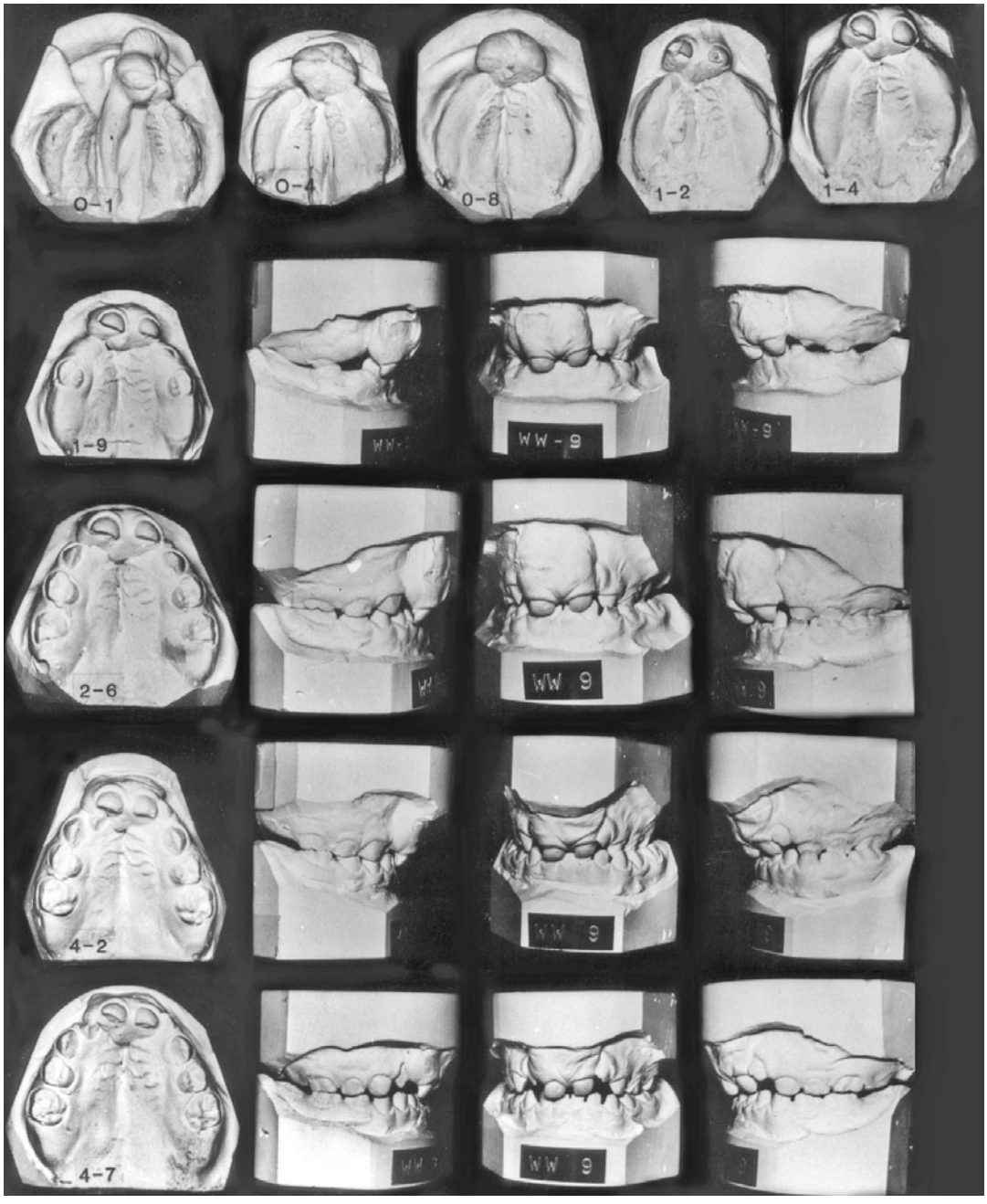

Fig. 7.20 Case TM (WW-9). Serial palatal cast changes when no presurgical orthopedics were utilized. 0–1 Newborn, prior to lip surgery. 0–4 After lip adhesion, the lateral palatal segment moved medially behind the premaxilla. 0–8 The cleft space is much smaller and continues to decrease in size with palatal growth. The premaxillary position at this age poses no treatment or growth problem. 1–2 The palatal cleft was closed with a modified von Langenbeck procedure. 1–9 Ideal incisal overbite and overjet. 2–6 and 4–2 Left buccal crossbite which was corrected at 4–6 using a fixed palatal expander. 4–7 Good occlusion

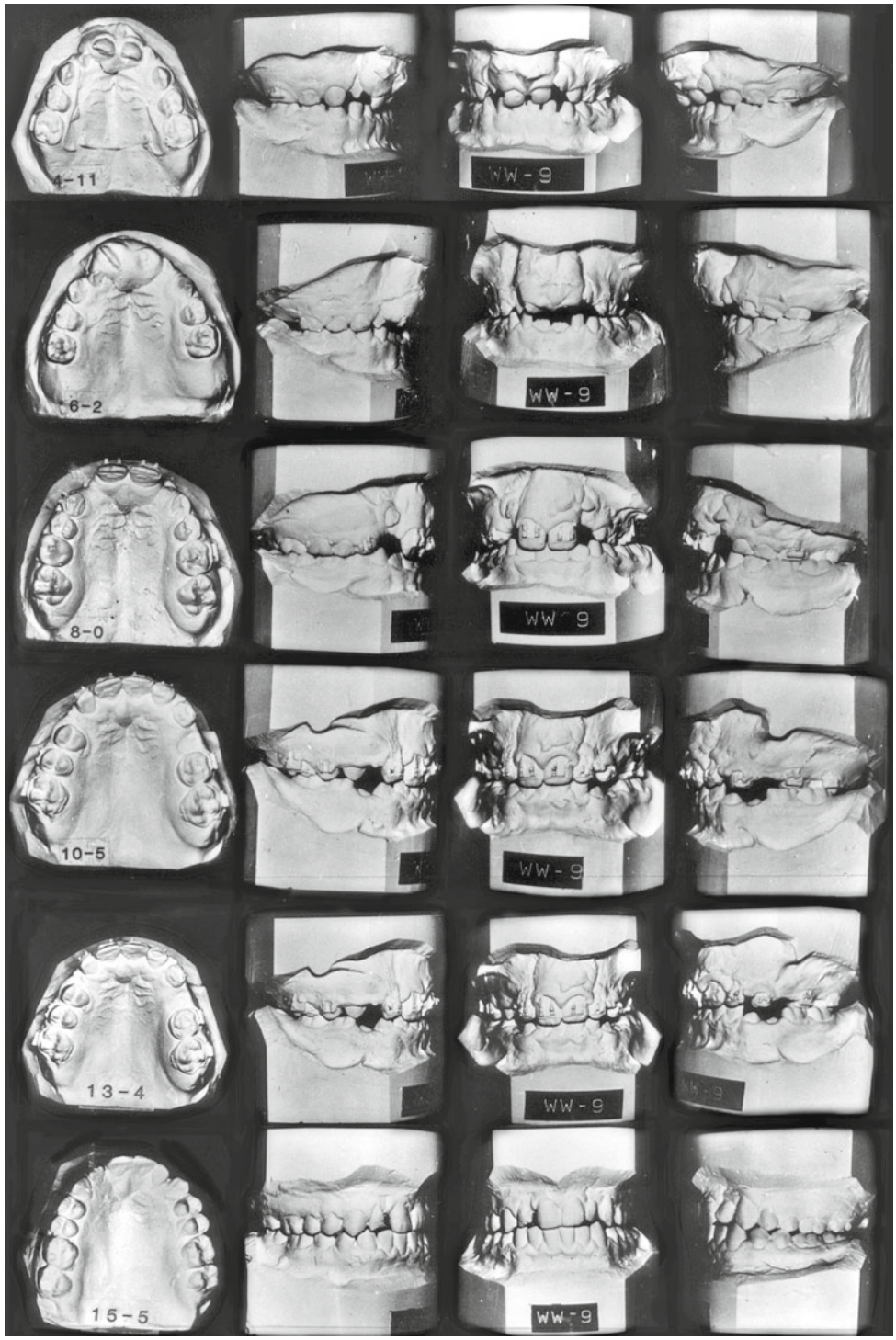

Fig. 7.20 (*continued*) 4–11 Outline of fixed palatal retainer is seen on the case. Note that the premaxilla is now positioned within the palatal segments. 6–2 The central incisors in the area of the cleft are frequently found to be rotated. 8–0 Six months after a secondary alveolar cranial bone graft. The right and left lateral incisors have erupted through the graft. 10–5 The lateral incisors are aligned with the central incisors. 15–5 Excellent occlusion after orthodontia. A removable maxillary retainer is being used to maintain the arch form

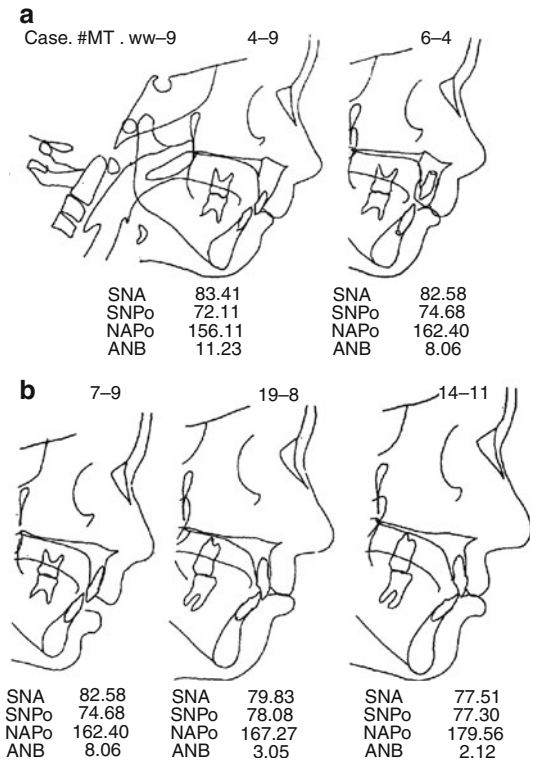

Fig. 7.21 (**a**, **b**) Case TM (WW-9). This series depicts an excellent facial growth pattern. (**a**) Computerized tracings of facial skeletal and soft tissue profile changes. At 14–11, the profile measurements are well within the normal range. (**b**) Facial polygons superimposed on the SN line and registered at S demonstrate that the excellent facial growth pattern reflects some anterior cranial base growth, very little forward growth of the midface, and excellent mandibular growth in a downward and forward direction. Note that the midface was still protrusive at 7–9, but after the mandibular pubertal growth spurt (13–8), the facial profile flattened markedly

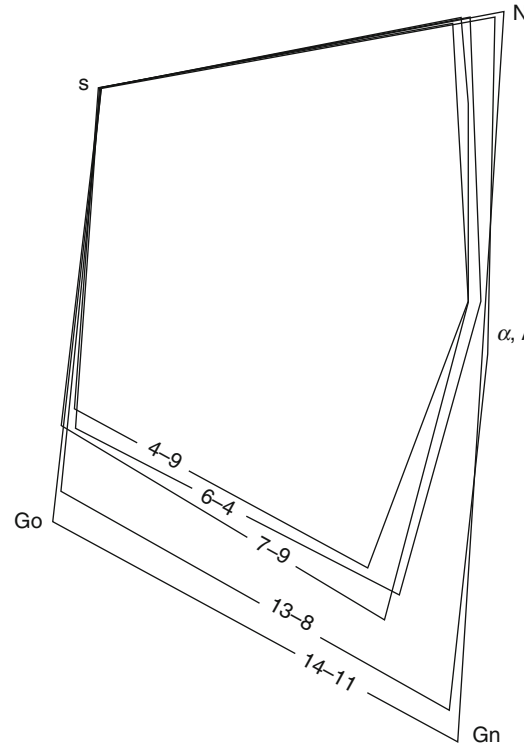

Fig. 7.22 Case TM (WW-9) Computerized tracings of facial skeletal and soft tissue profile changes. At 14–11, the profile measurements are well within the normal range. Facial polygons superimposed on the SN line and registered at S demonstrate that the excellent facial growth pattern reflects some anterior cranial base growth, very little forward growth of the midface, and excellent mandibular growth in a downward and forward direction. Note that the midface was still protrusive at 7–9, but after the mandibular pubertal growth spurt (13–8), the facial profile flattened markedly

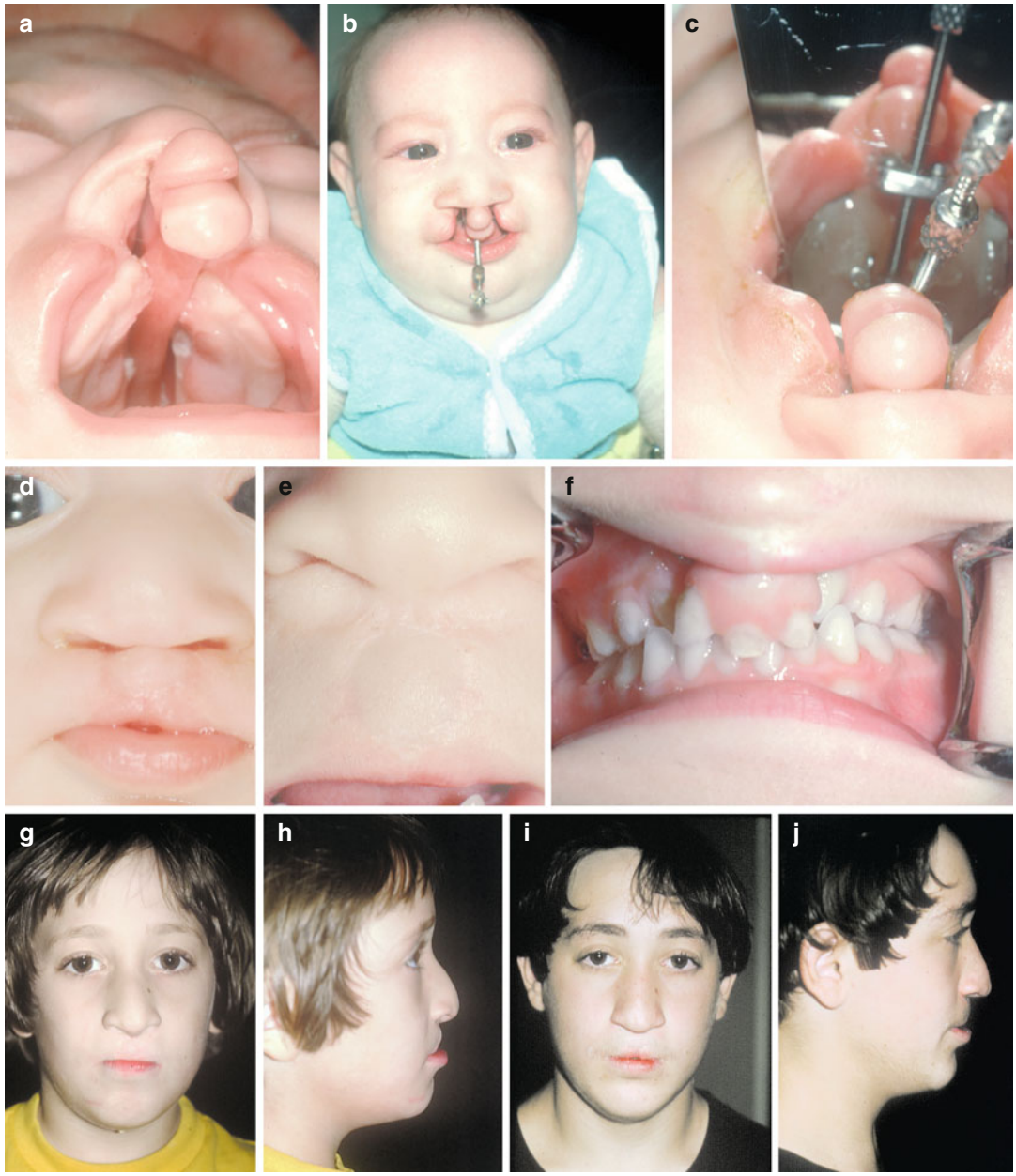

Fig. 7.23 (**a**–**q**) Case DK (AI-31) demonstrates unsuccessful use of Latham's presurgical premaxillary mechanical retraction procedure in a CBCLP, which then required the use of external facial elastics prior to lip surgery. (**a**) At birth, note the very small protruding premaxilla. (**b**) Facial photograph with Millard-Latham (M-L) mechanical premaxillary retraction appliance in place. (**c**) Intraoral view of M-L appliance in place. The appliance was not able to retract the premaxilla due to its small size and was discarded. (**d**) After wearing a head bonnet with an elastic strap for 2 weeks, the lip was united over the premaxilla. At 2 months, the nasal tip is severely depressed. (**e**) At 3 years of age, the banked tissue of the "forked flap" waiting to be placed in the columella. (**f**) 5 years of age. Note excellent occlusion. Radiographs showed that there were no permanent incisors in the premaxilla. (**g**, **h**) At 8 years of age, the nasal tip has been elevated, but there is poor upper lip support due to the now retruded premaxilla. Lower right and left first bicuspids were extracted at 13 years to reduce the anterior crossbite. (**i**–**k**) At 15 years of age, the mandible is growing forward at a more rapid rate and degree than the maxilla. The earlier buccal and anterior crossbites were corrected, and two lateral incisors from the lateral palatal segments were brought into position, improving facial aesthetics

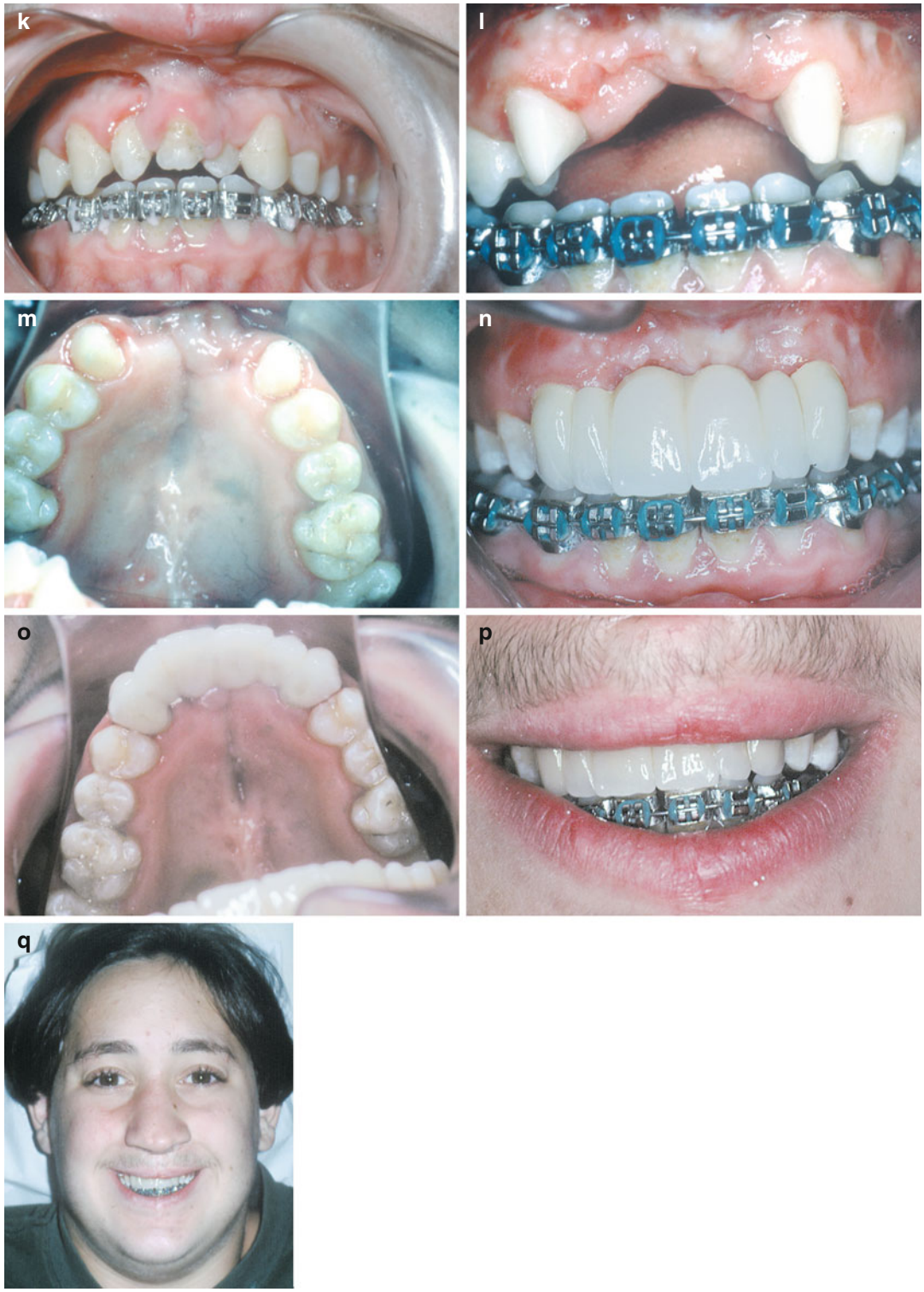

Fig. 7.23 (*continued*) (**l**–**q**) The maxillary anterior deciduous teeth were extracted and an anterior bridge fabricated to improve dental function and aesthetics

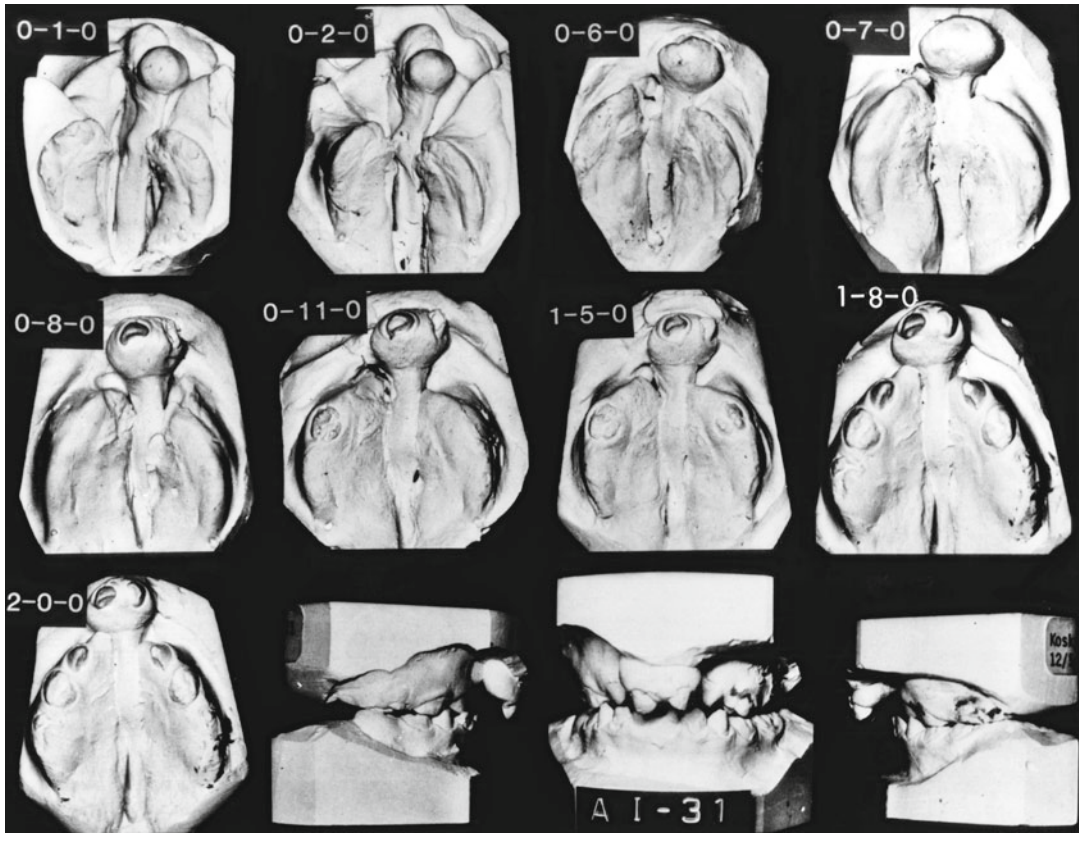

Fig. 7.24 Case DK (AI-31). a Serial dental casts. 1–1–0 and 0–2–0 Note the severely protruding and small premaxilla. 0–2–0 and 0–6–0 The premaxilla ventroflexed under the influence of extraoral elastics attached to a head bonnet. 0–7–0 Marked increase in palatal size coupled with a big reduction in the anterior cleft space. 2–0–0 Class II right and Class I left occlusions with a severe anterior overjet. Premaxillary surgical setback should not be performed

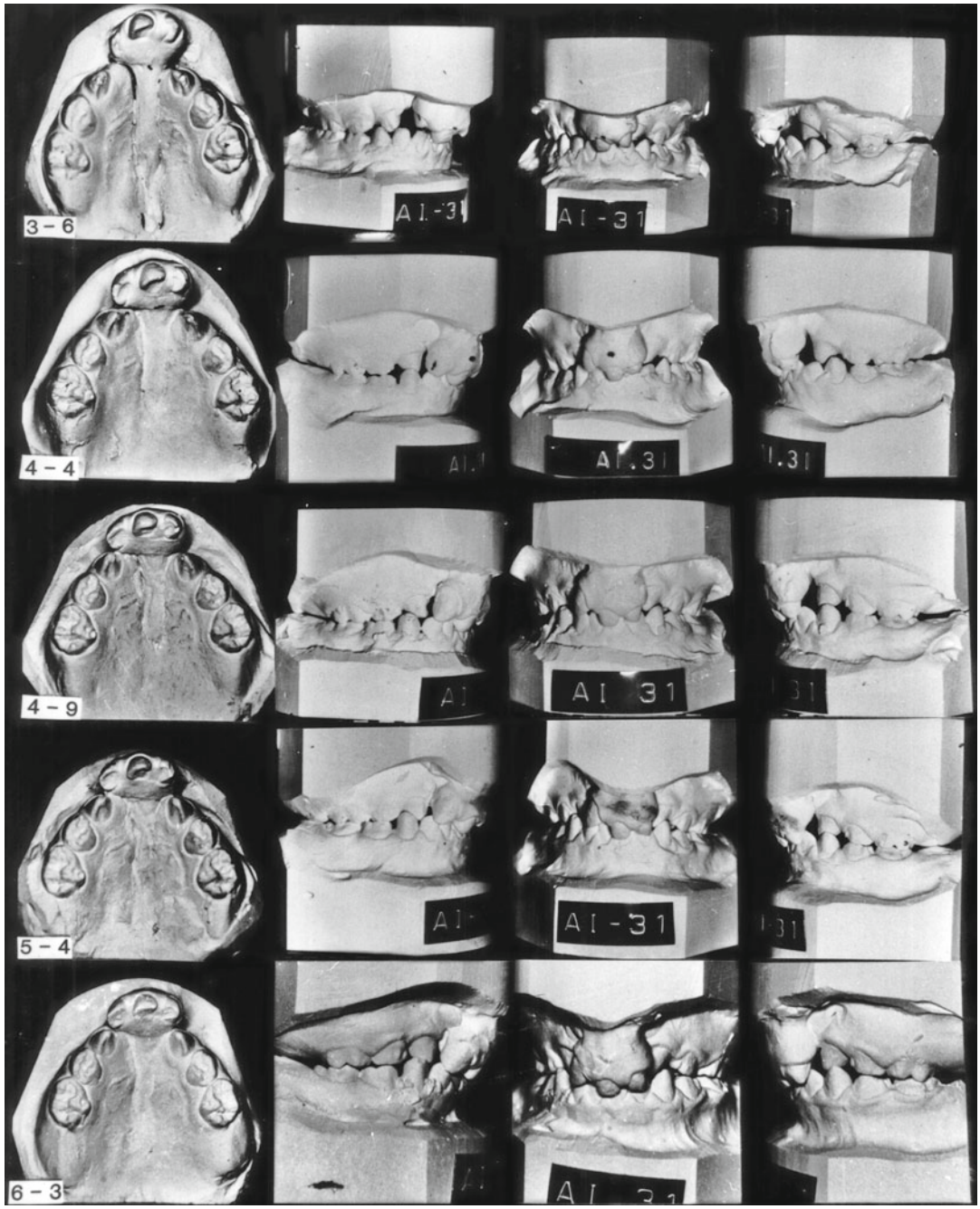

Fig. 7.24 (*continued*) 3–6 Note only a small overjet and overbite with a mesioangular rotation of the lateral segments placing the cuspids into crossbite. 4–4 The premaxilla is forward of the lateral palatal segments, with a normal dental overbite/overjet. This occlusal relationship remained for the next 5 years. The anterior cleft space is left open waiting for additional palatal growth. The anterior palatal cleft did not pose a speech or feeding problem and would allow for palatal expansion

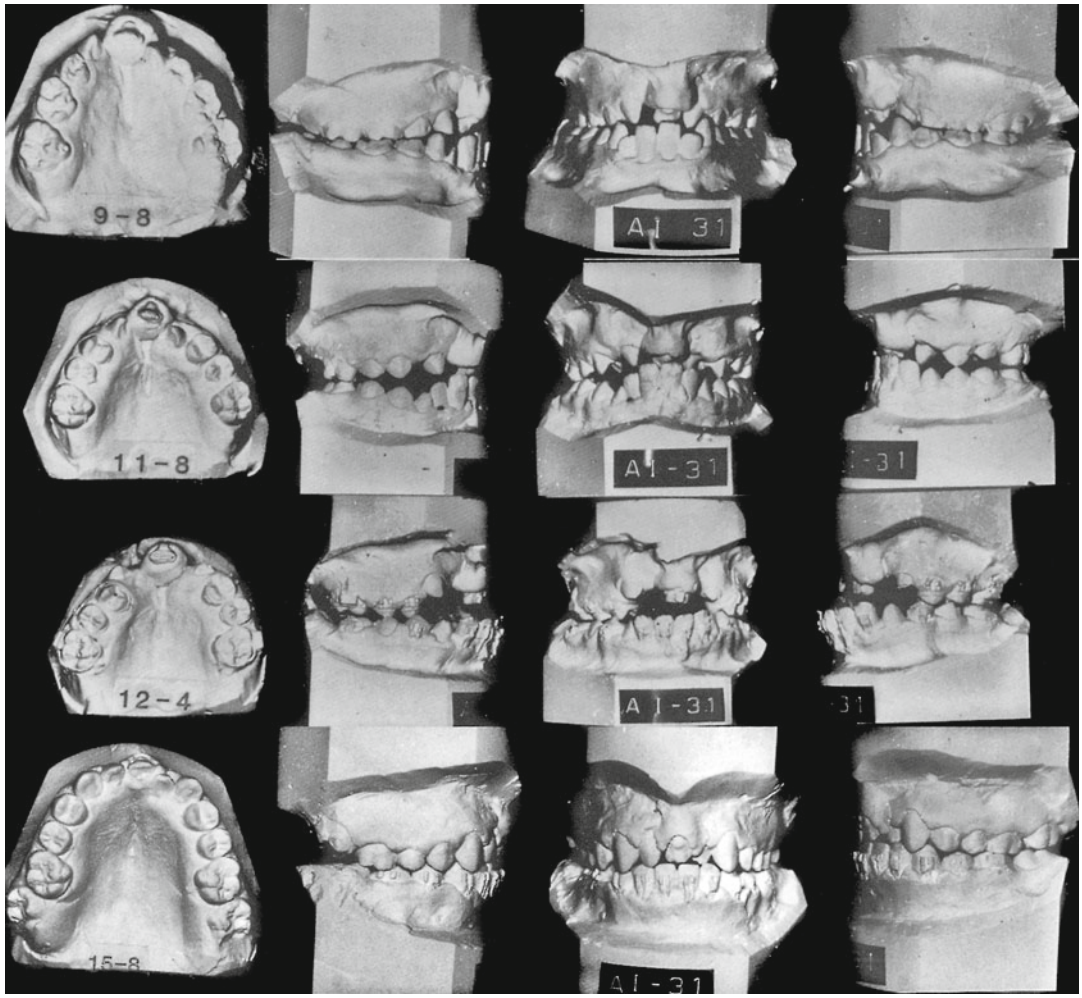

Fig. 7.24 (*continued*) 9–8 The remaining deciduous central incisor is now in an anterior crossbite associated with the lack of midfacial development. 12–4 The lower anterior teeth were unsuccessfully advanced to reduce the arch crowding. The excessive flaring necessitated extraction of the lower first bicuspids. This was followed by retraction of the incisor teeth and space closure with reduction of the severe anterior crossbite. 15–8 Right and left deciduous lateral incisors are brought into position; however, an anterior arch discrepancy still remains. Because of the poor root development of the deciduous lateral incisors, they later were extracted and replaced with an anterior fixed bridge. Comment: This case clearly demonstrates that the anterior occlusion cannot be predicted at birth; therefore, early premaxillary surgical setback should not be performed

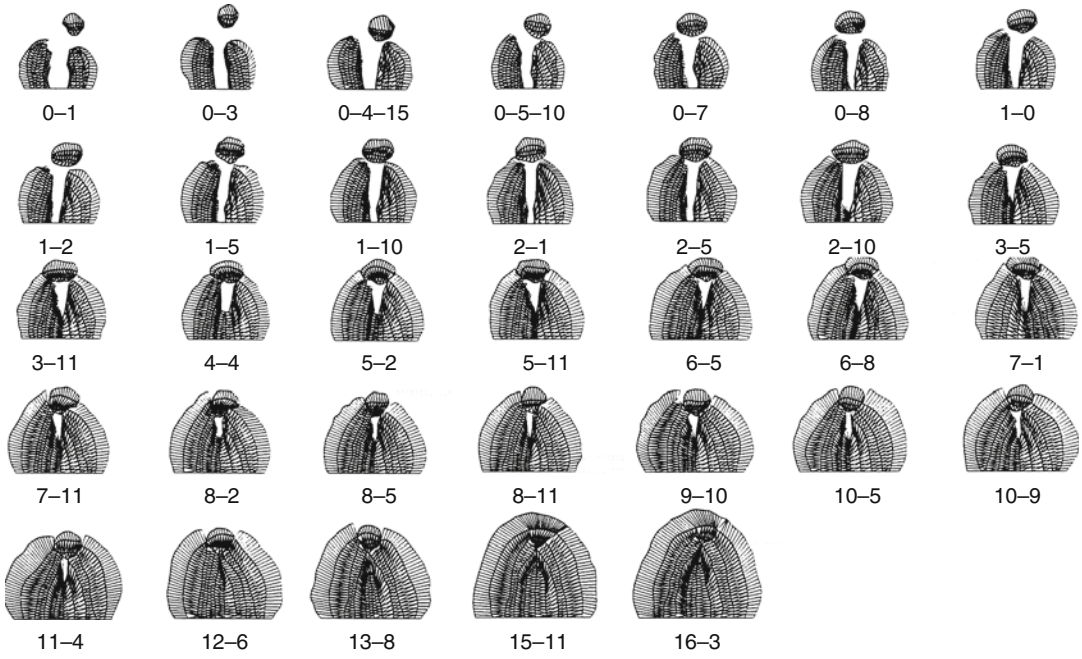

Fig. 7.25 Case DK (AI-31). Computerized drawings of palatal casts using an electromechanical digitizer. Each cast form is drawn to scale but not its size. The premaxilla is gradually aligned within the lateral palatal segments after the initial ventroflexion. The ratio of palatal size to cleft size increases with growth. The anterior cleft space is left open until a secondary alveolar bone graft is placed. The palatal cleft was closed at 4–0 leaving an anterior palatal fistula which was closed with a secondary alveolar bone graft at 12–1

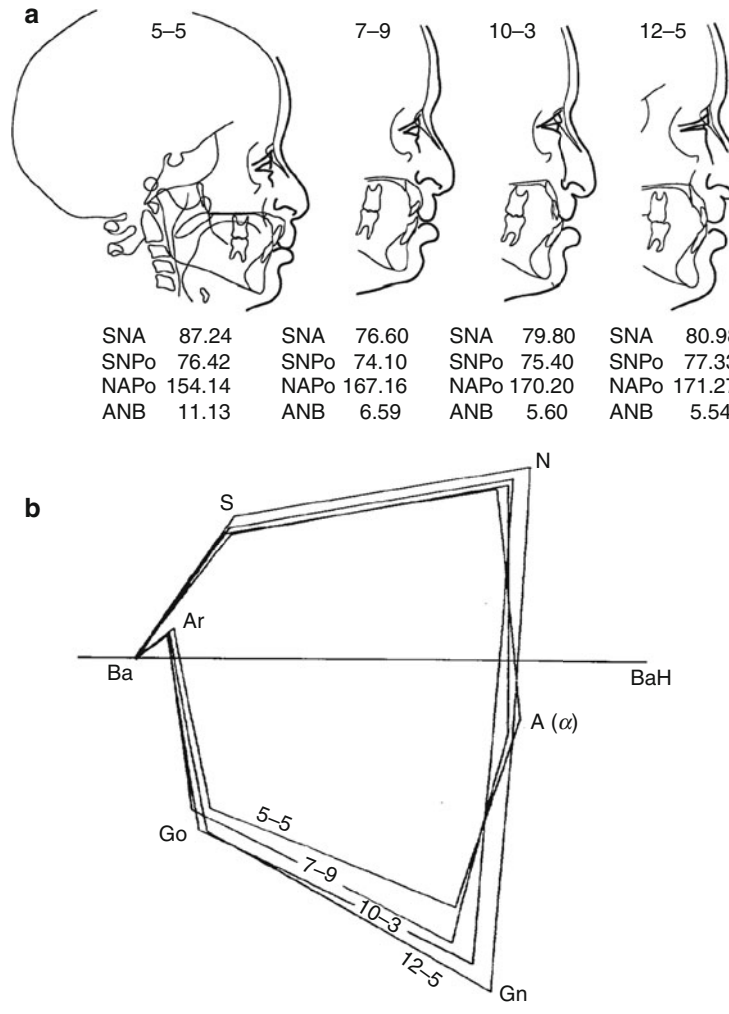

Fig. 7.26 (**a**, **b**) Case DK (AI-31). (**a**, **b**) Serial lateral cephaloradiographic tracings show changes in the skeletal and soft tissue profile. The upper lip gradually became recessive, reflecting the lack of midfacial growth and superimposed basion horizontal facial polygons of Coben. The midfacial protrusion seen at 12 years, 5 months is no different to that seen at 5 years, 5 months of age. At 10 years of age, the midface was more recessive, but with the application of premaxilla advancement orthodontics for 2 years, it was positioned more anteriorly. The lower jaw showed progressive downward and forward growth. As in all faces that grow well, this growth pattern was mainly responsible for the flattening of the facial profile. Comments: I must stress that the Coben analysis demonstrates the forward/downward growth of the entire maxillary and mandibular growth; therefore, the maxilla actually recedes due to growth at the upper and lower face

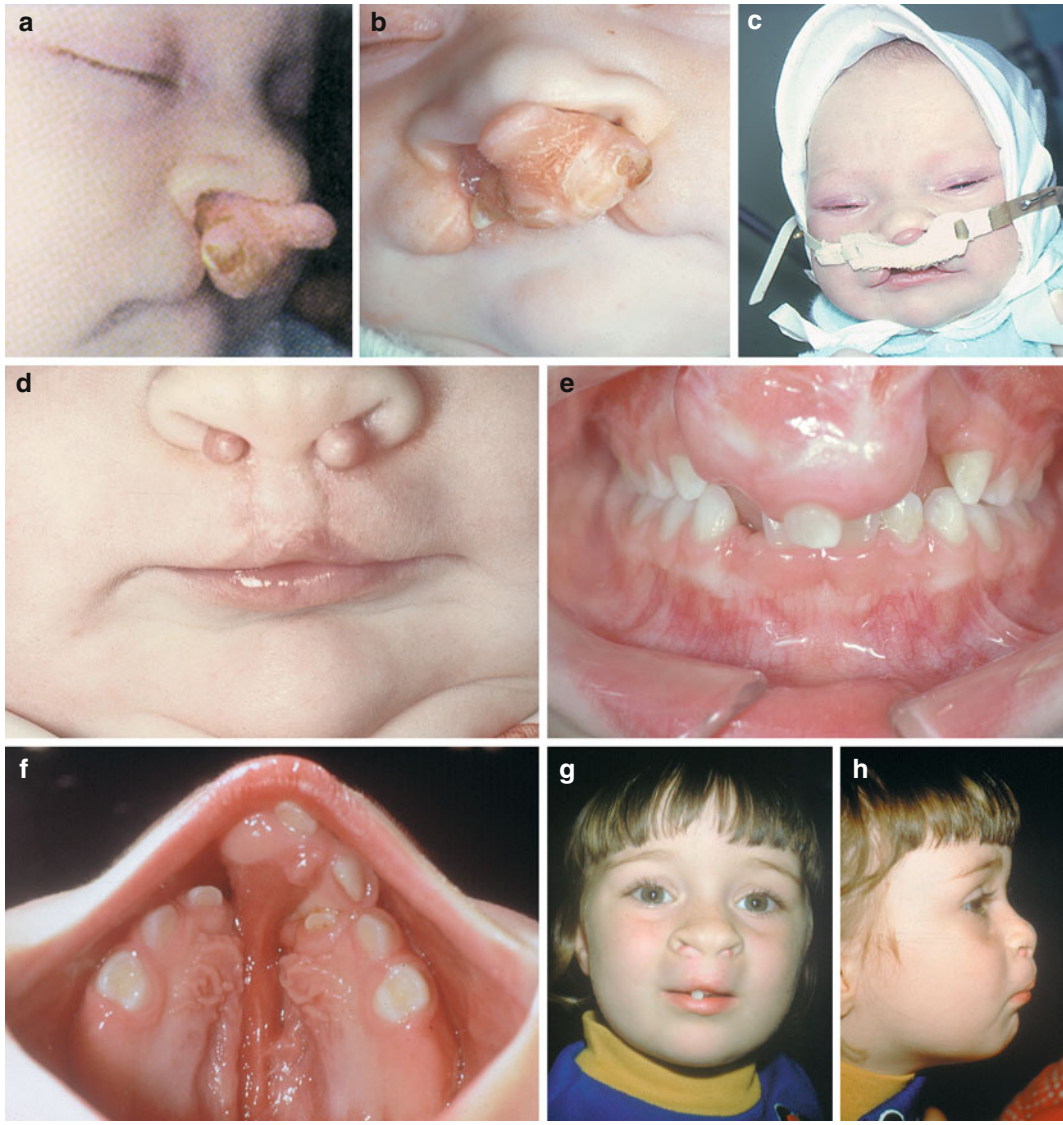

Fig. 7.27 (**a**–**p**) Case PM (KK-22). Serial photographs demonstrate excellent facial and palatal growth in CBCLP. Time is an ally! No presurgical orthopedics other than the use of external elastics off a head bonnet for 10 days prior to lip surgery; arm restraints were used to prevent removal of the head bonnet. The palatal cleft was closed at 8 years of age using a modified von Langenbeck procedure. Secondary alveolar cranial bone grafting was done at 8 years of age. (**a**, **b**) At birth, an extremely protrusive premaxilla. (**c**) Head bonnet with elastic over the premaxilla. (**d**) The lip cleft was closed using Millard's forked flap procedure. The tissue in the nostril is used to reconstruct the columella at a later date so that the surgeon need not go back to the lip for tissue. (**e**) Occlusal view at 5 years of age. Note that the premaxilla is still protrusive. (**f**) The lip is being pushed forward by the protruding premaxilla. The palatal cleft was closed at 8–6. The face at 5 years showing upper lip protrusion

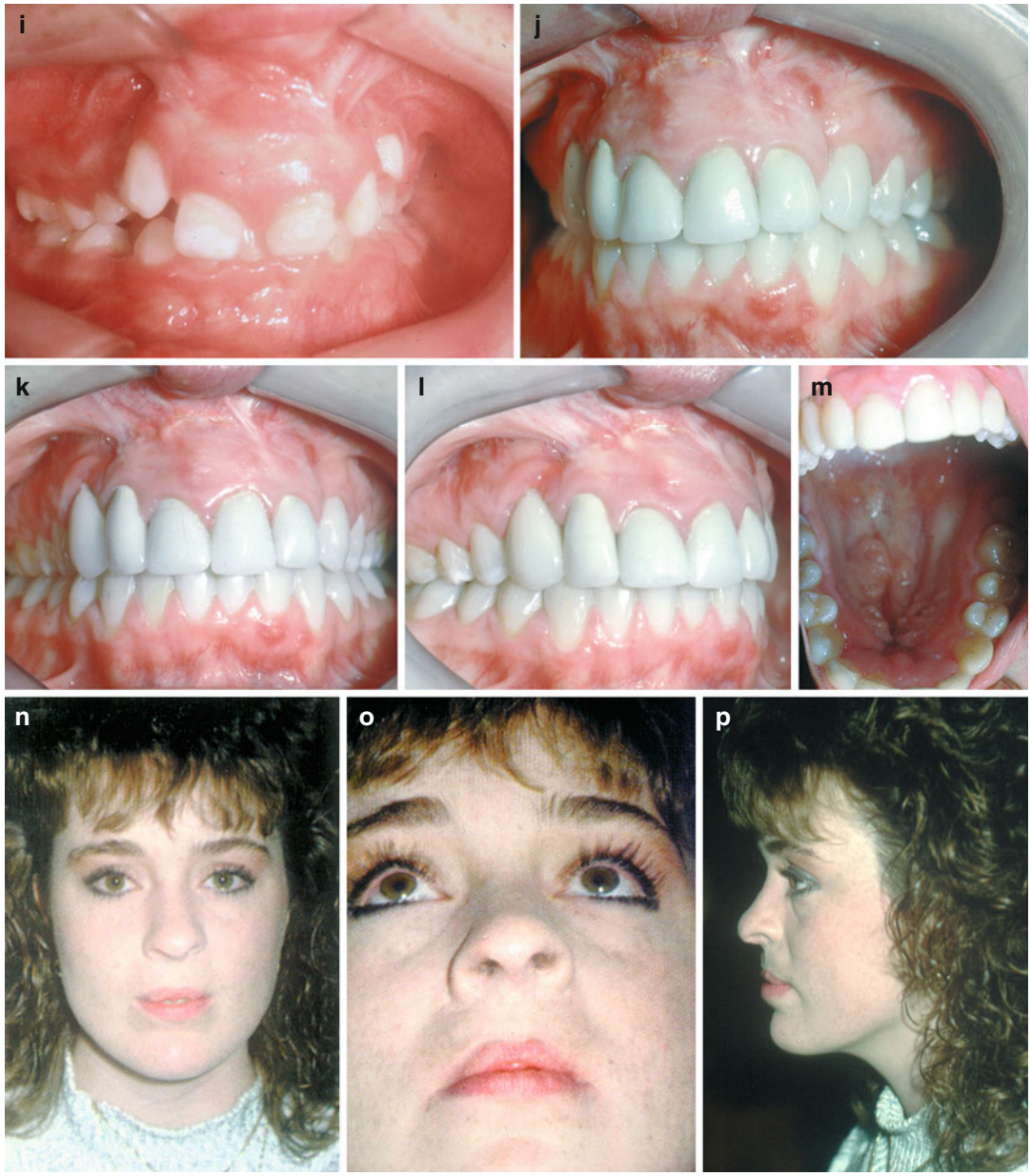

Fig. 7.27 (*continued*) (**i**) 11–01 The premaxilla is still protrusive. (**j–l**) At 20 years of age, the occlusion is ideal. Minor imperfections in the anterior teeth were corrected by the use of composite material. (**m**) Ideal arch form with a normal palatal vault space. Good alveolar bone support to the lateral incisors. (**n–p**) Facial photographs at 20 years of age showing a harmonious and pleasing soft tissue profile. Comments: No protraction midfacial mechanics were used

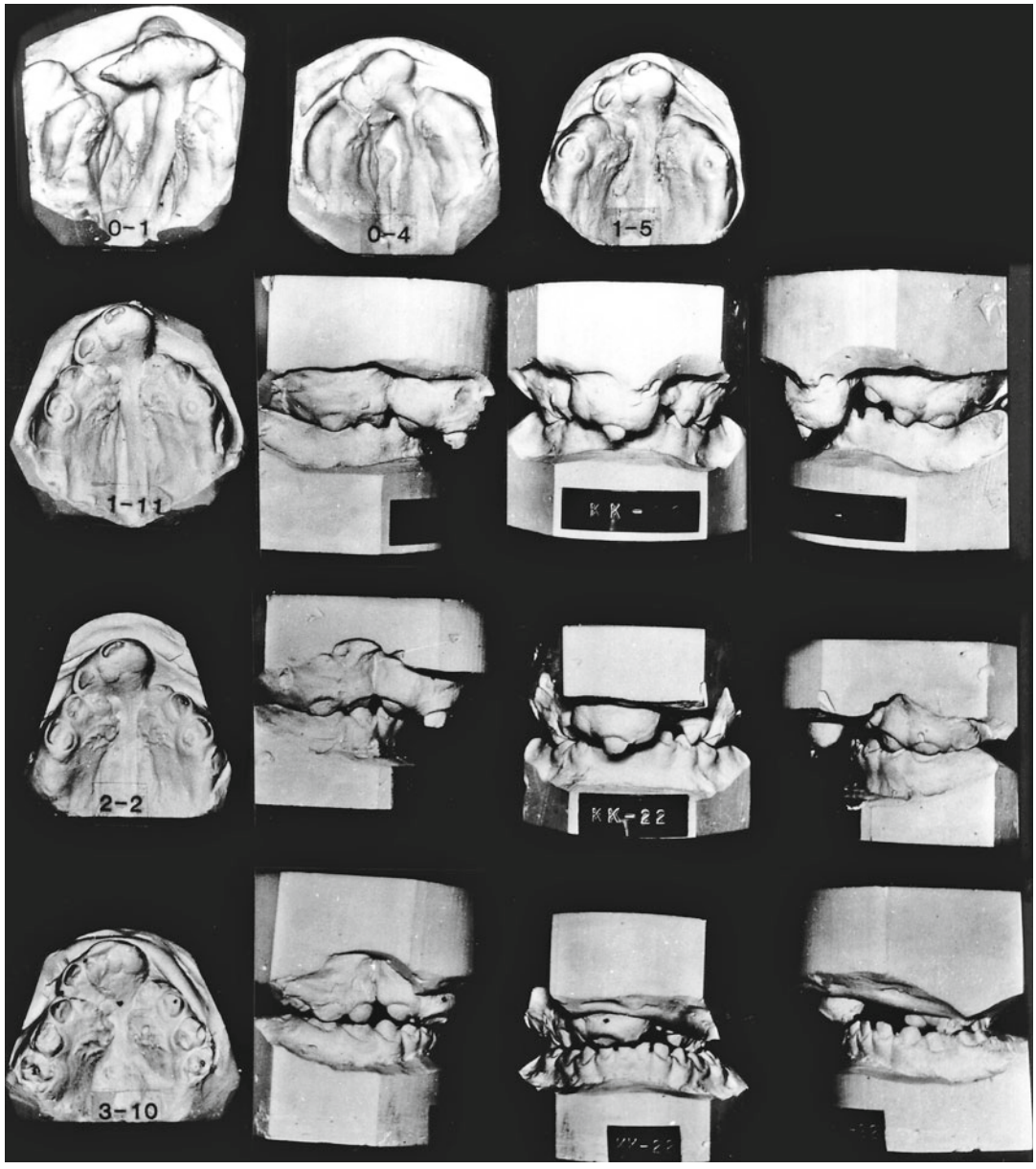

Fig. 7.28 Case PM (KK-22). Serial casts: 0–1 At birth, the septum is deviated to the left and the right palatal segment is laterally displaced. 0–4 With an external elastic and lip surgery, the premaxilla flexes ventrally and medially, making contact with both palatal segments. The remaining casts show excellent buccal occlusion in Class II relationship with overbite and severe anterior overbite and overjet

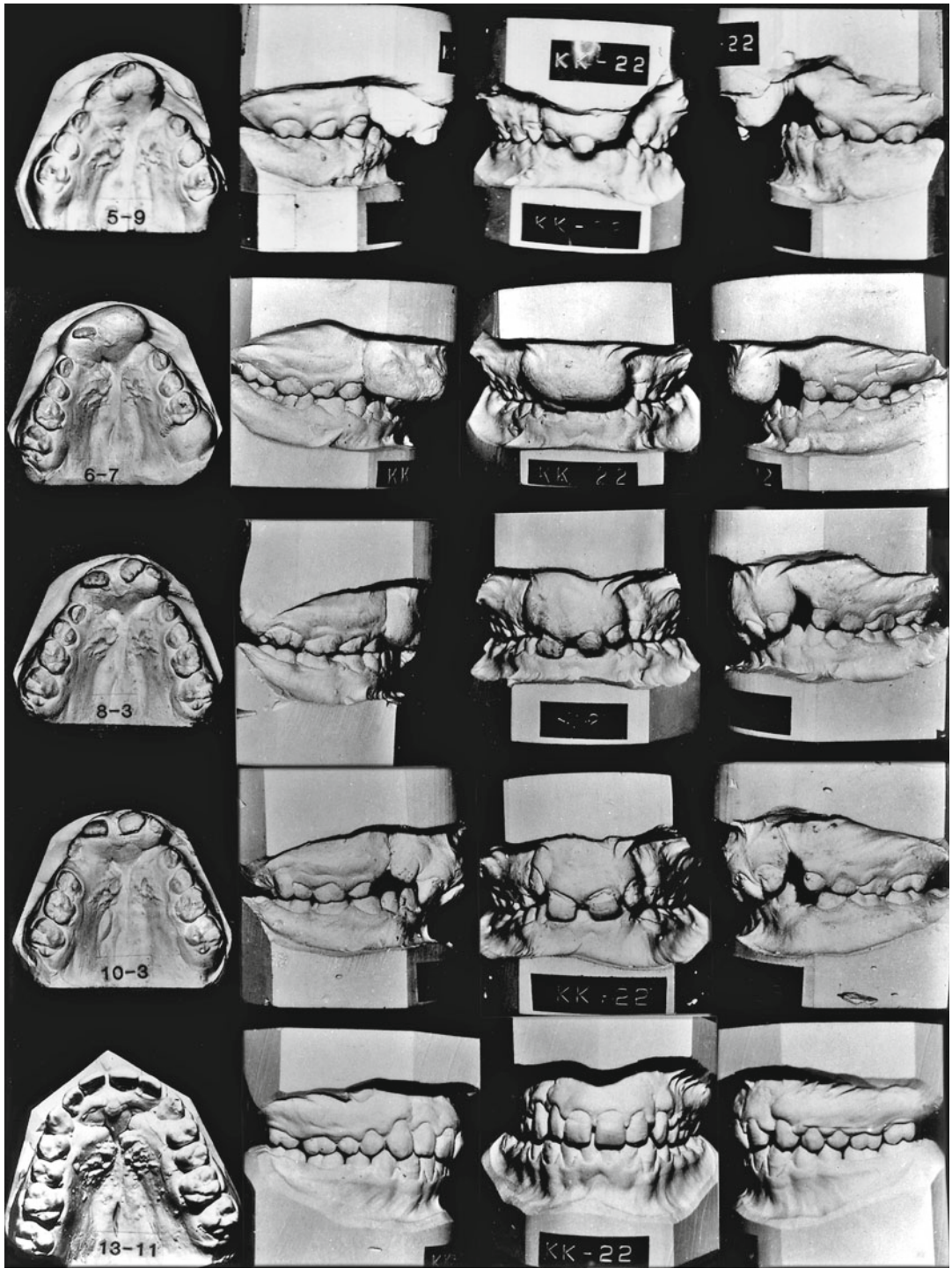

Fig. 7.28 (*continued*) By 8–3, the anterior overjet is markedly reduced by growth. 13–11 Conventional orthodontics were eventually used to reduce the Class II occlusion and align the anterior teeth into an ideal overbite-overjet relationship

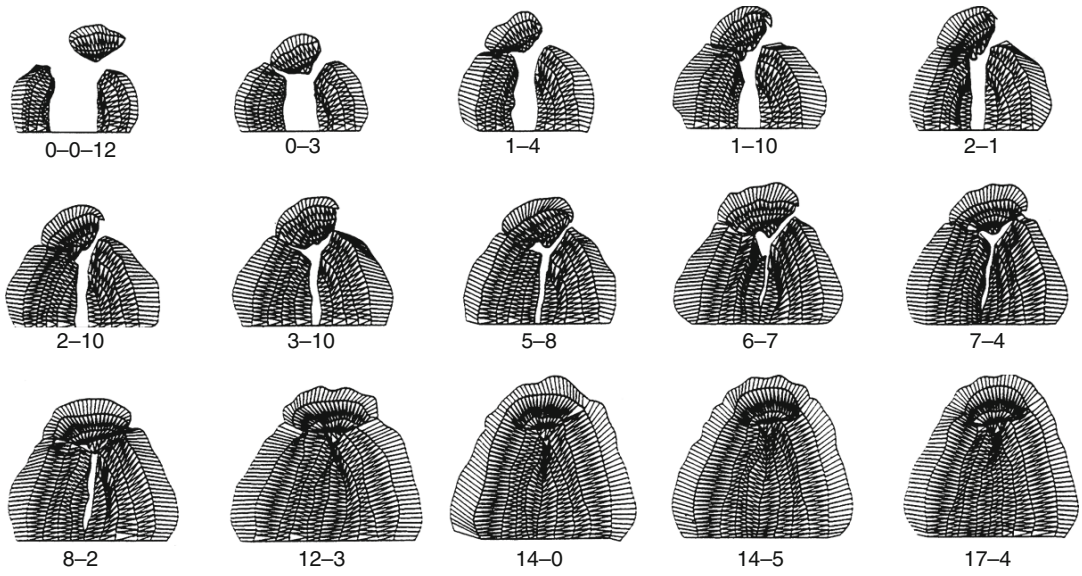

Fig. 7.29 Case PM (KK-22). Computer-generated outlines of the serial casts were performed using an electromechanical digitizer. All casts are drawn to scale. The casts range from 12 days to 17 years, 4 months of age. This series demonstrates the spontaneous closure of the anterior and posterior cleft spaces after "molding action" brought on by uniting the lip and then by gradual palatal growth at the border of the cleft space. The premaxilla was initially aligned forward of the lateral palatal segments but was satisfactorily incorporated within the arch at a later age

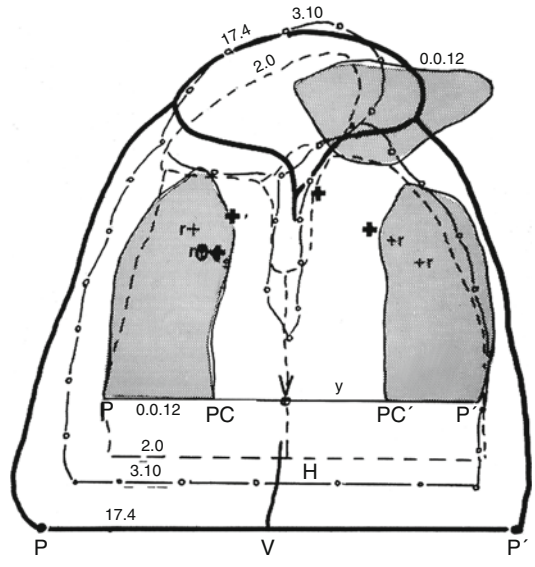

Fig. 7.30 Case PM (KK-22). Palatal outlines were superimposed using the rugae for registration. This series shows that the premaxilla's position within the maxillary complex at 17 years of age is similar to that seen at birth. Excellent growth occurs in all dimensions and is similar to the growth pattern seen in noncleft palates. Increased posterior palatal growth is necessary to accommodate the developing molars. Alveolar bone growth with tooth eruption increases midfacial height. Comments: The position of the anterior premaxilla relative to the anterior cranial base (Nasion) to the anterior position of pogonion of the mandibular symposium shows the same relative position from birth to 17 years of age. These 2 studies confirm that midfacial growth is retarded

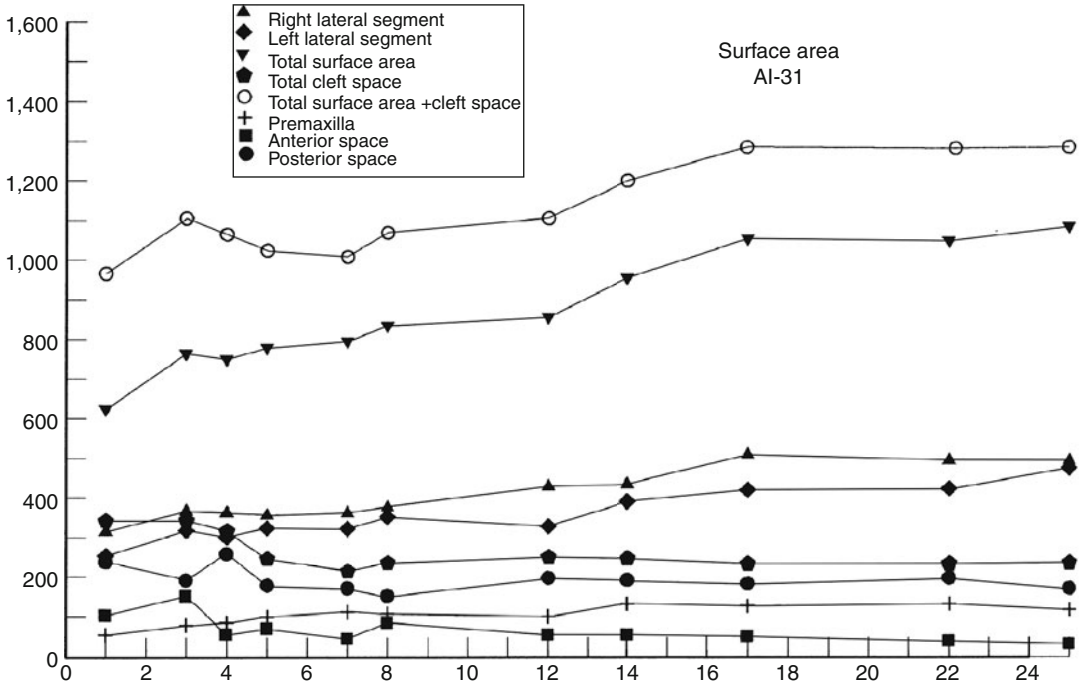

Fig. 7.31 Case DK (AI-31). Premaxillary ventroflexion with medial movement of the lateral palatal segments caused a great reduction in the anterior and posterior cleft spaces by 5 months of age. Thereafter, for the next 19 months, the anterior cleft space gradually reduced, while the posterior cleft space showed some increase due to the increase in palatal length. Both lateral palatal segments showed a similar, gradually increasing growth rate

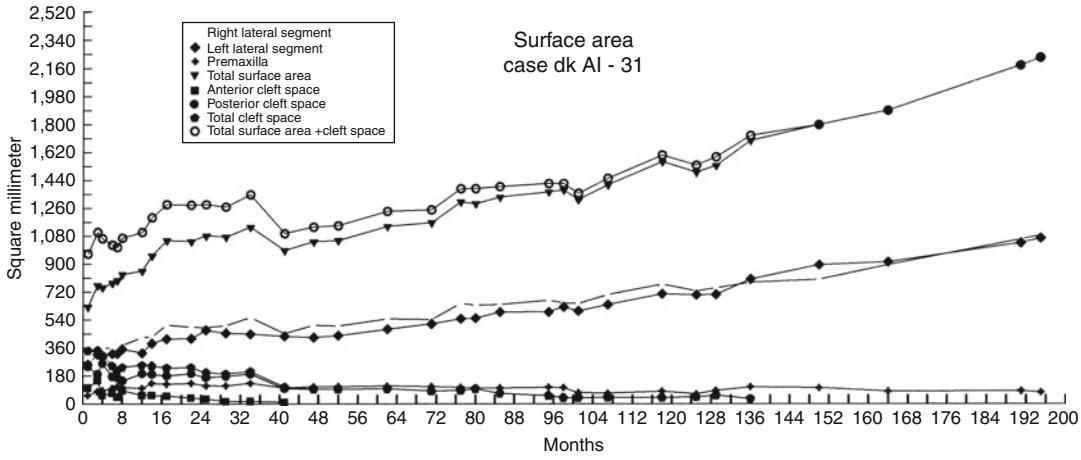

Fig. 7.32 Case DK (AI-31). After the initial change in cleft size brought on by medial movement of the lateral palatal segments and ventroflexion of the premaxilla, the greatest acceleration of cleft space closure occurred between 2–10 and 3–5. The premaxilla reached its largest size by 3 years, which is associated with eruption of the teeth. Palatal growth acceleration occurred between 1–3 months and 12–14 months and then gradually tapered off. The palatal segments had increased 37 % in size by 1 year and 74 % by 2 years. Palatal growth at its medial borders still occurs; even though it narrows the cleft space, the total cleft space is increasing in size due to the increase in palatal length

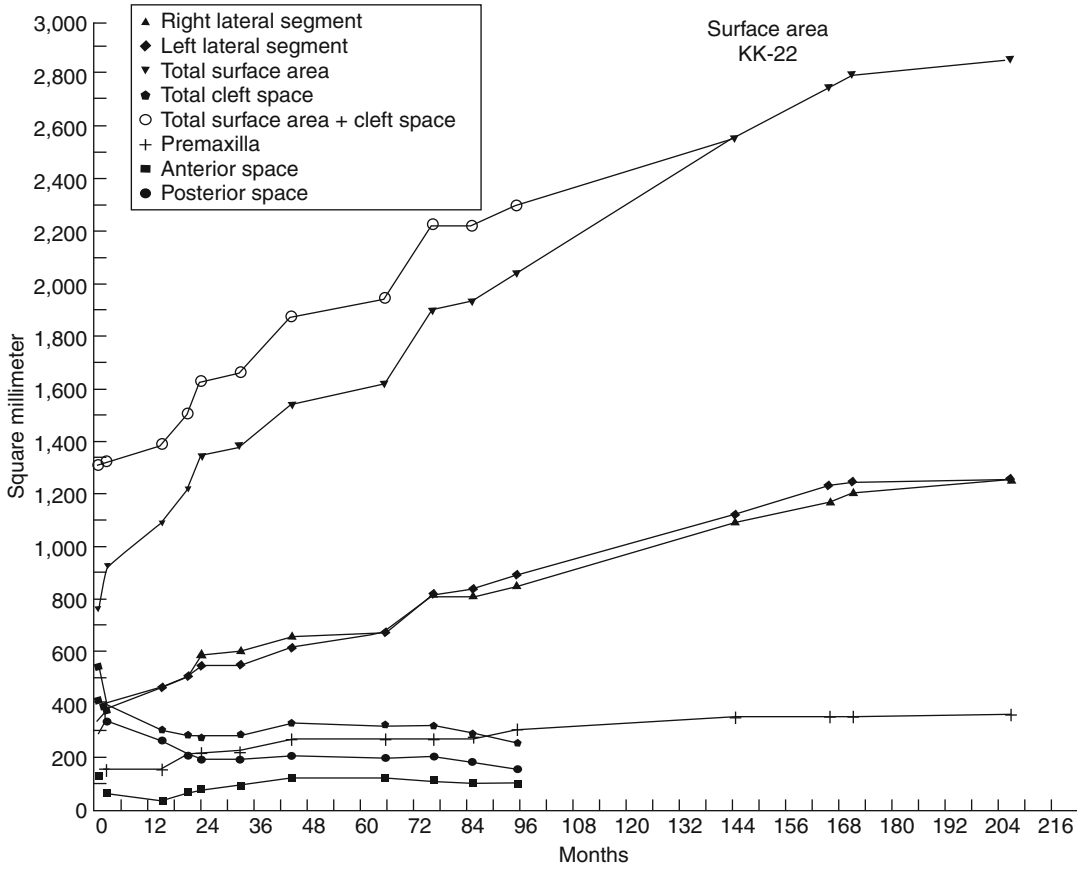

Fig. 7.33 Case PM (KK-22). Time sequence analysis of serial palatal growth shows that both palatal segments are growing at the same rate and to the same degree. The premaxilla is also increasing in size with tooth eruption but at a lesser rate. The greatest palatal growth acceleration occurs the first 2 years and then tapers off. The anterior cleft space is initially reduced as a result of premaxillary ventroflexion, but thereafter it remains the same dimensions until the palatal cleft is closed. The posterior cleft space initially is reduced with palatal medial movement. The resulting posterior cleft space remains approximately the same size for the next 8 years. It must be remembered that the cleft length is increasing, while the cleft width is decreasing. The net cleft area is gradually reducing with growth. All fistulae are closed by 12–3 years of age

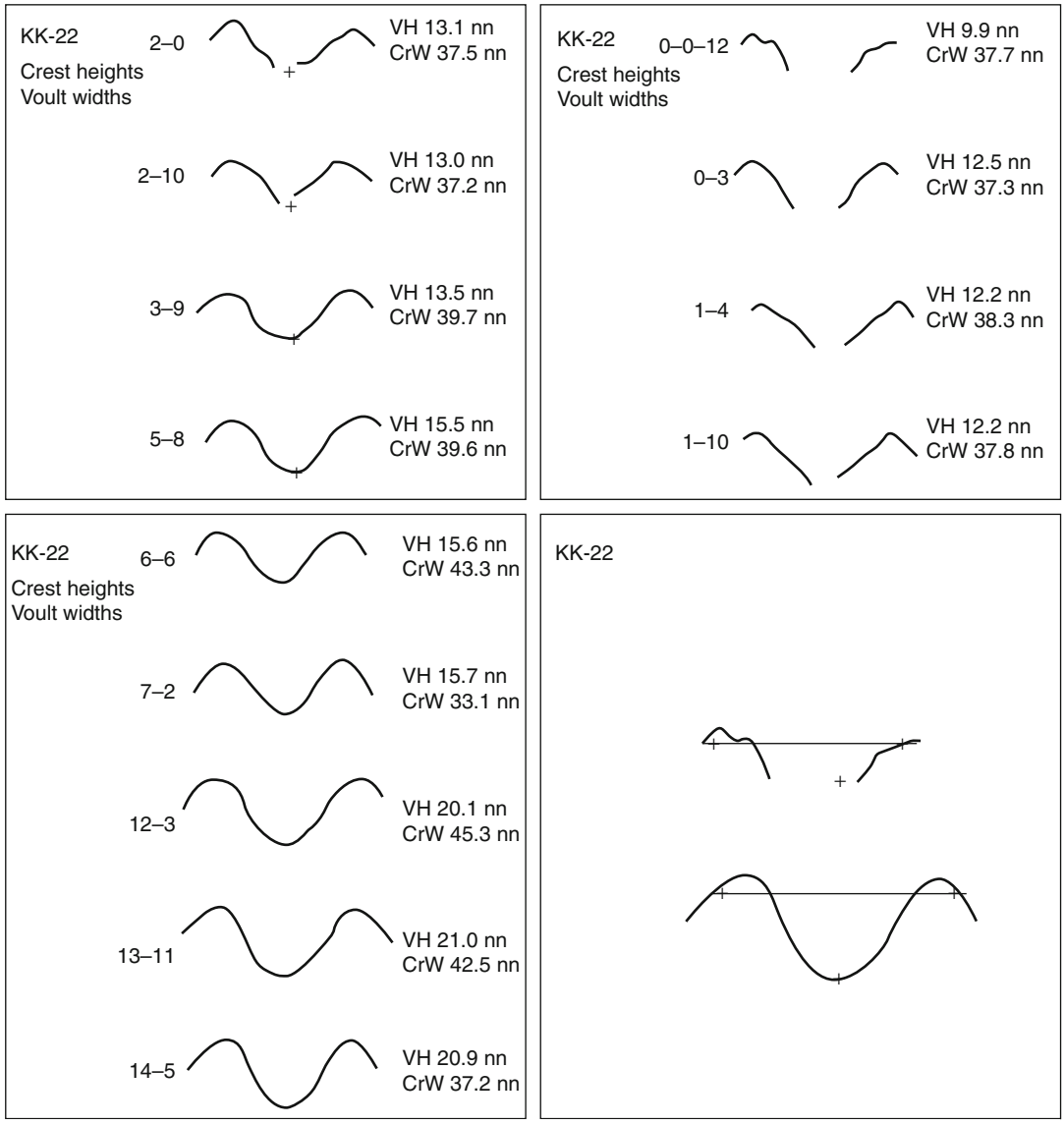

Fig. 7.34 Serial midpalatal cross sections showing vault height and palatal width changes using stereophotogrammetry. The left lateral palatal segment is attached to the vomer – while the right lateral palatal segment is displaced laterally. A lip muscle adhesion causes the displaced palatal segment to move medially, narrowing the cleft space. The appositional growth of the alveolar segments is continuous and becomes more obtuse. Closure of the palatal cleft space with a modified vomer flap maintains a normal vault height, and vault space flattening of the vault occurs in almost all cases in the absence of a vomer flap. Comments: A vomer flap with a von Langenbeck procedure seems to create a minimum scarring. Vomer flap alone performed early (6 months to 1 year) created excessive scarring

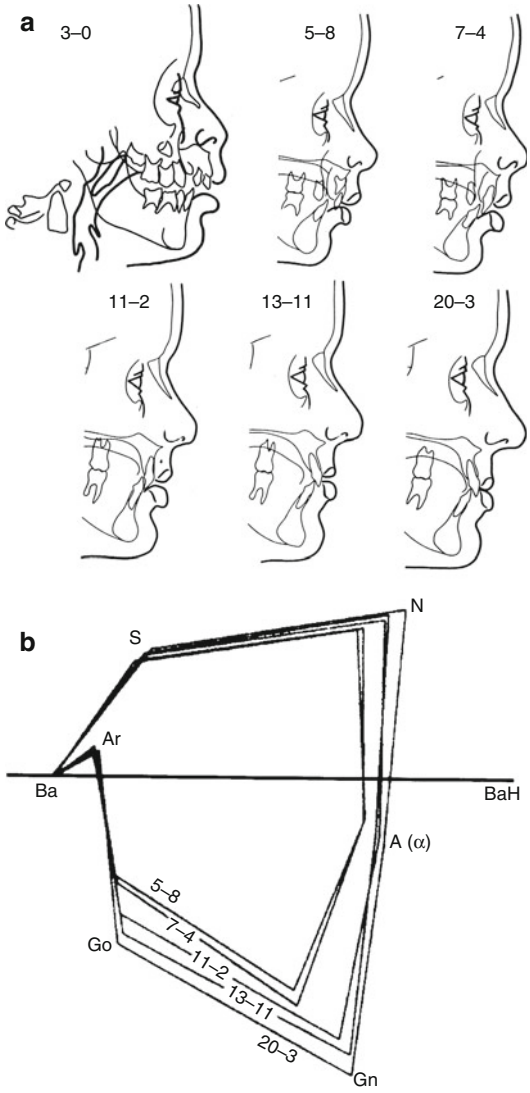

Fig. 7.35 (**a**, **b**) Case PM (KK-22) a Cephalometric serial tracings of the skeletal and soft tissue profile show marked reduction of the midfacial protrusion. b Superimposed serial tracings using Coben's basion horizontal method show an excellent facial growth pattern which straightens the skeletal profile. There is very little forward midfacial growth between 11 and 20 years of age. During the same time period, growth at the anterior cranial base and the mandible contributed to flattening of the facial profile

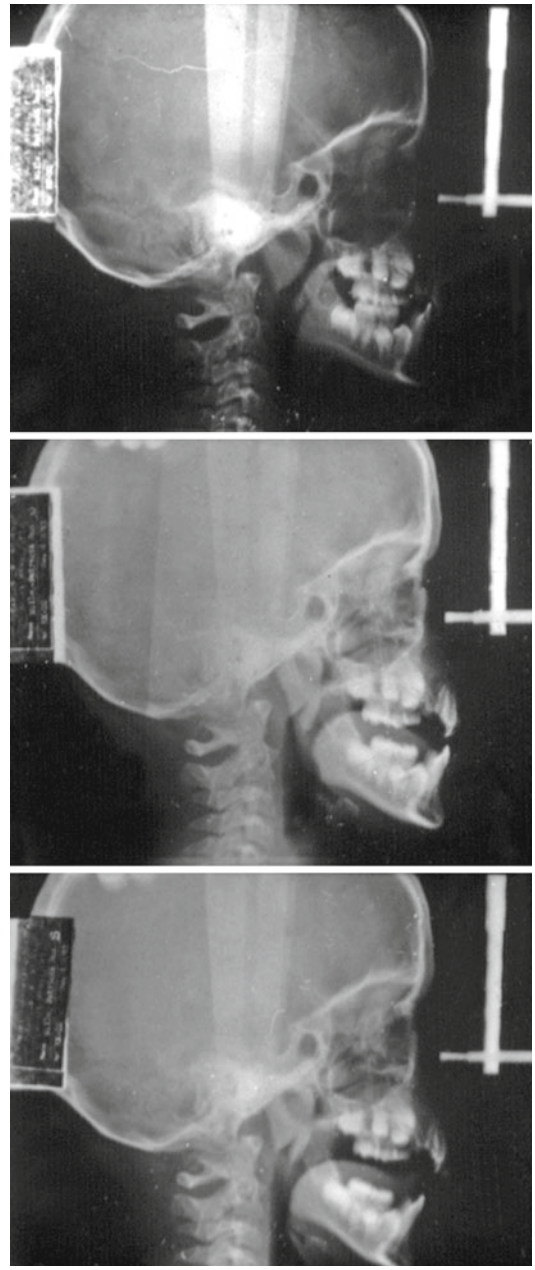

Fig. 7.36 Case PM (KK-22). Cephaloradiographs taken at 5 years of age. *Top*: At rest with teeth together. *Middle*: Taken while vocalizing "Youu …" *Bottom*: Taken while vocalizing "Sss …" Comments: When vocalizing both sounds, the velum elevates and makes contact with the adenoids. The pharyngeal depth is relatively small. The adenoids are of moderate size; the velum is of good length and shows good elevation. This is not a functional test to evaluate velopharyngeal closure, but it does show the well-proportioned oral and nasal pharyngeal spaces which are conducive to good velopharyngeal closure. A gap space more than 5 mm may indicate VPI exist with inadequate lateral pharyngeal wall movements

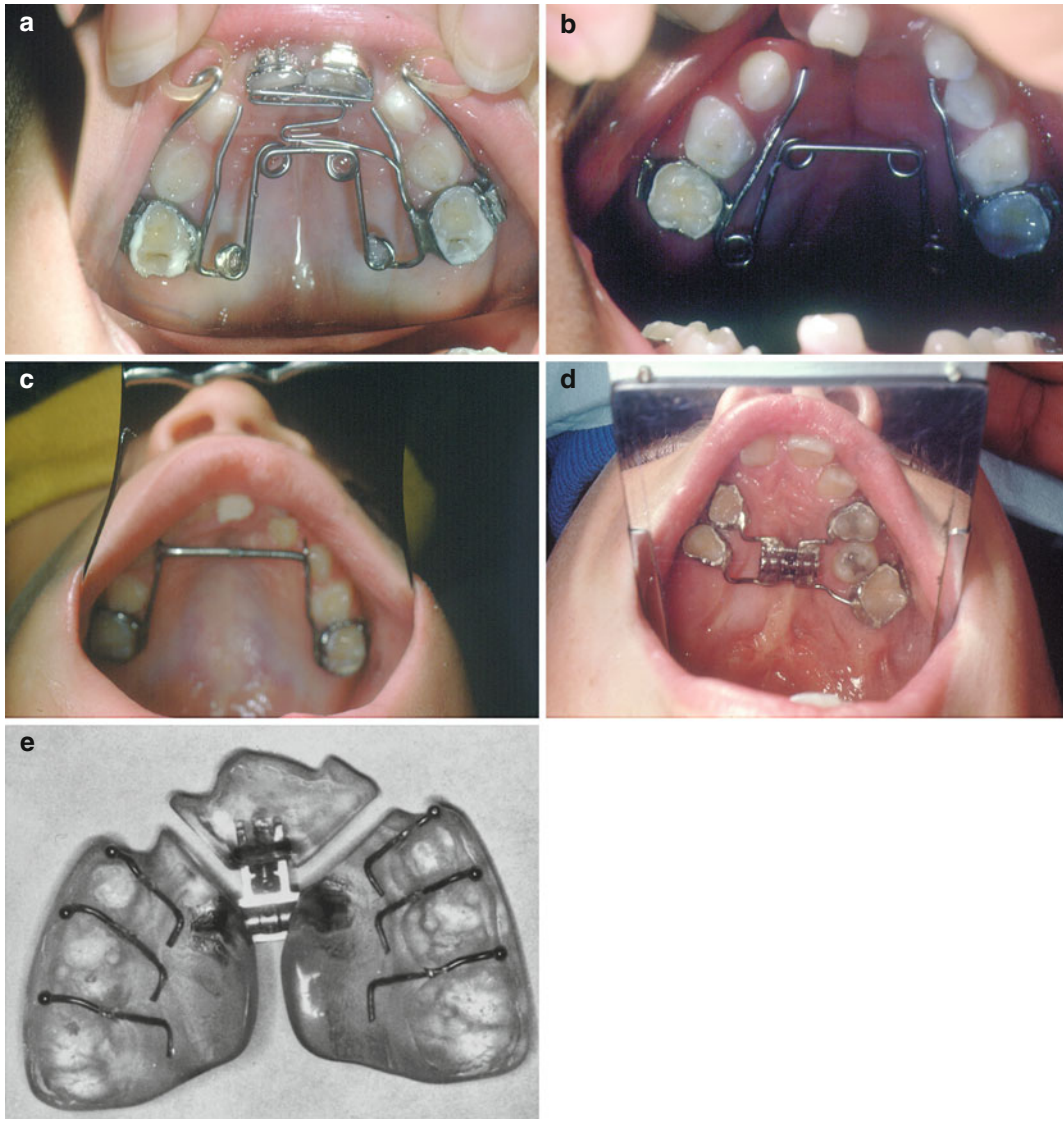

Fig. 7.37 (**a**–**e**) Various palatal expansion appliances. (**a**) "W" appliance with finger springs designed to move the central incisors forward while correcting the posterior crossbite. (**b**) "W" appliance. (**c**) Arnold expander: a .040 wire is inserted into a .040 tube; the compressed open coil spring exerts a gentle lateral force moving the two segments apart. A larger diameter (.045) tube wire allows the cuspids to be moved laterally more than the molars. (**d**) A Hyrax expander, which needs a lever and parent involvement to activate the very strong expansion force. This appliance is rarely necessary with meager transpalatal scarring. (**e**) A three-part removable expansion plate used to simultaneously advance and expand the anterior and buccal segments in a BCLP. Appliances that attach to the teeth are more reliable and efficient than removable ones

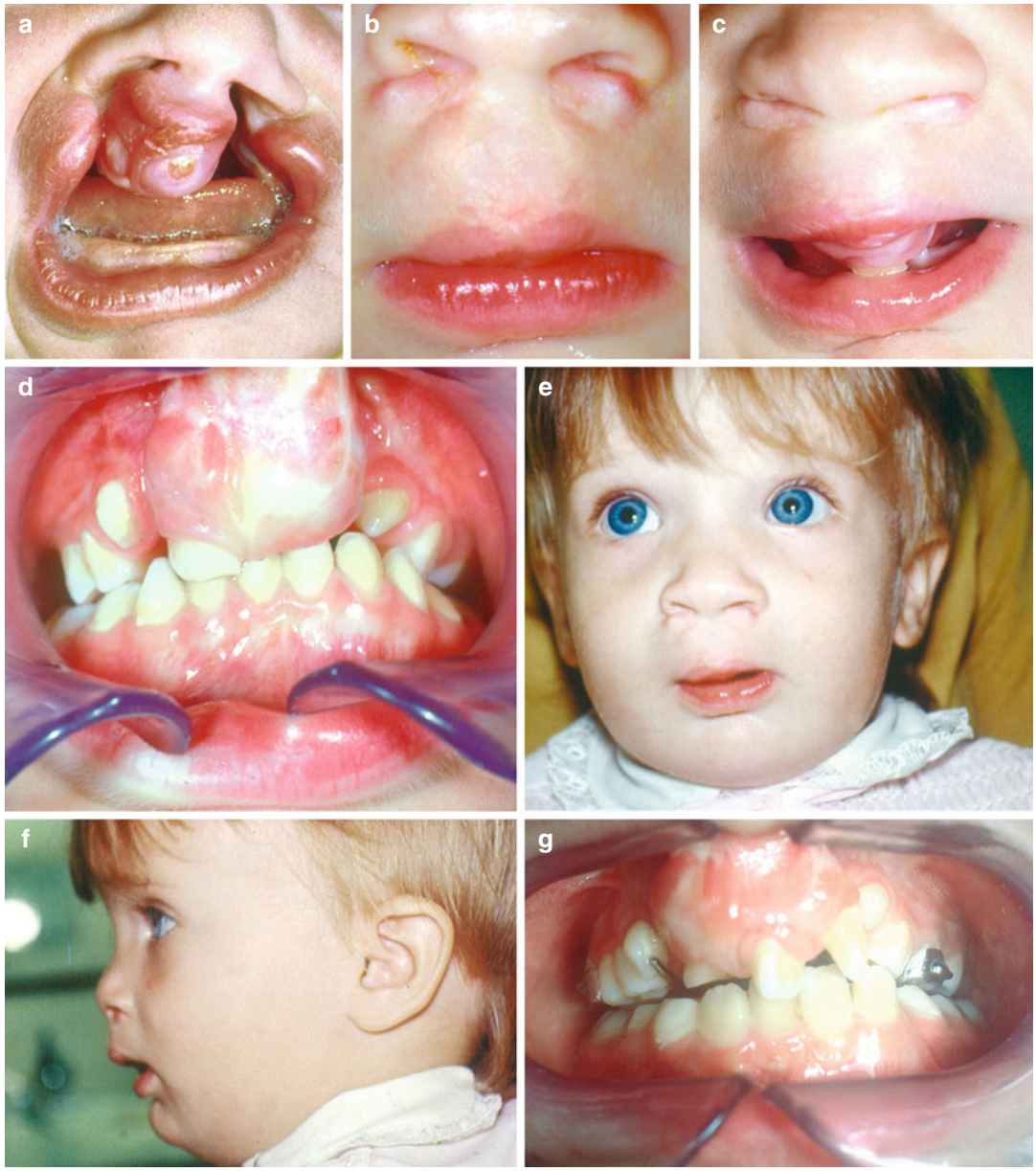

Fig. 7.38 (**a**–**s**) Case ML (KK-56) demonstrates severe premaxillary protrusion at birth in IBCLP. "Whisker" forked flap was performed at 2 months, definitive lip surgery at 6 months, and palatal cleft closure at 18 months. Secondary alveolar cranial bone grafting was placed at 8 years, 3 months. Maxillary surgery with chin augmentation was performed at 15 years, 7 months. (**a**–**g**) Facial and intraoral photographs show progressive facial and occlusal changes

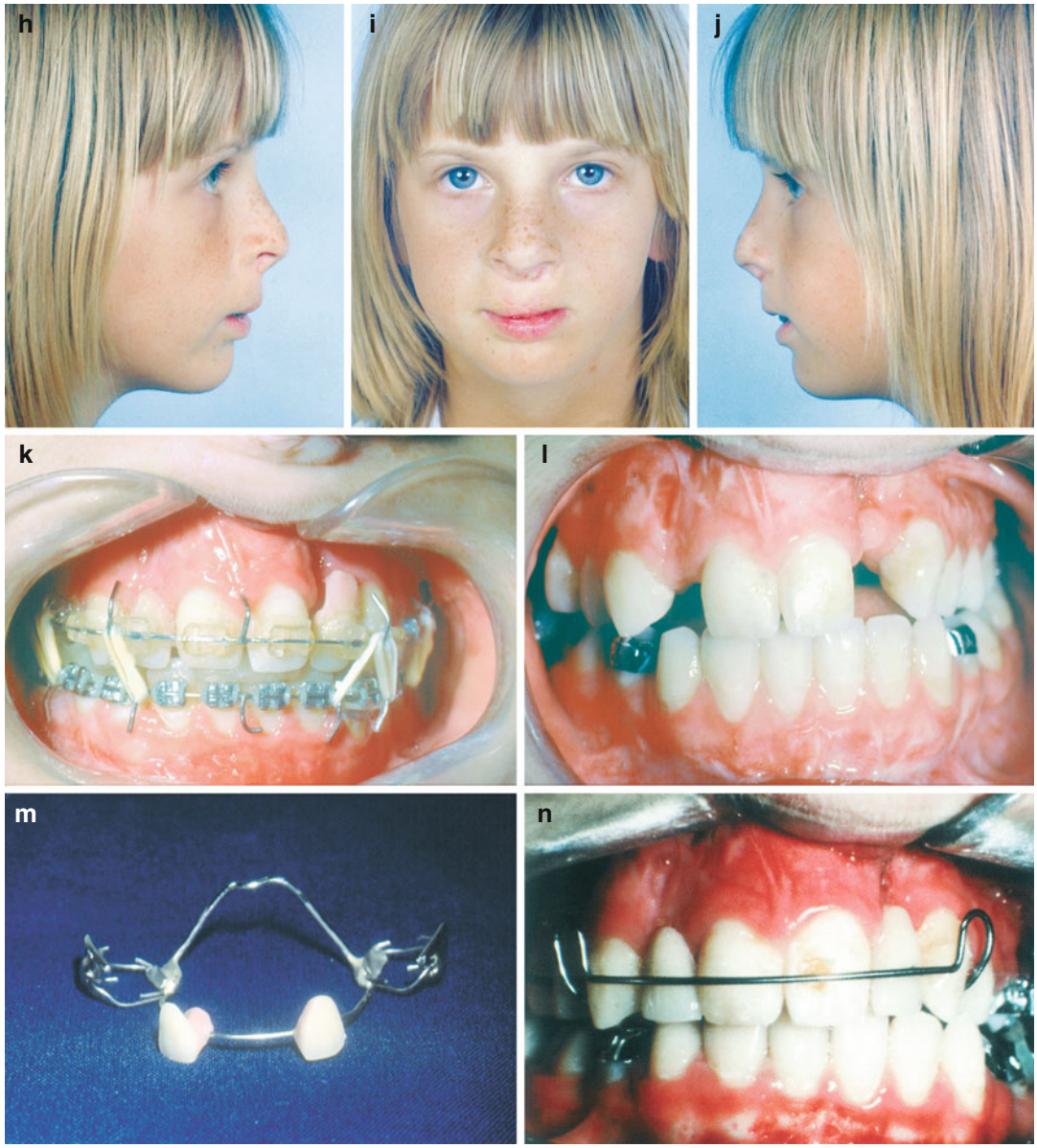

Fig. 7.38 (*continued*) (**h**–**j**) Facial photographs at 8 years of age. (**k**) After Lefort I advancement, the lateral incisor pontics were attached to the arch wire for aesthetics. (**l**, **m**) Occlusal photographs showing missing incisor spaces. (**n**–**p**) Intraoral photographs showing retainer with lateral incisor pontics in place. (**q**–**s**) Facial photographs at 17 years of age. Case ML (KK-56) demonstrates severe premaxillary protrusion at birth in IBCLP. (**n**) A partial upper retainer in place with pontics for missing lateral incisors. Case ML (KK-56) demonstrates severe premaxillary protrusion at birth in IBCLP

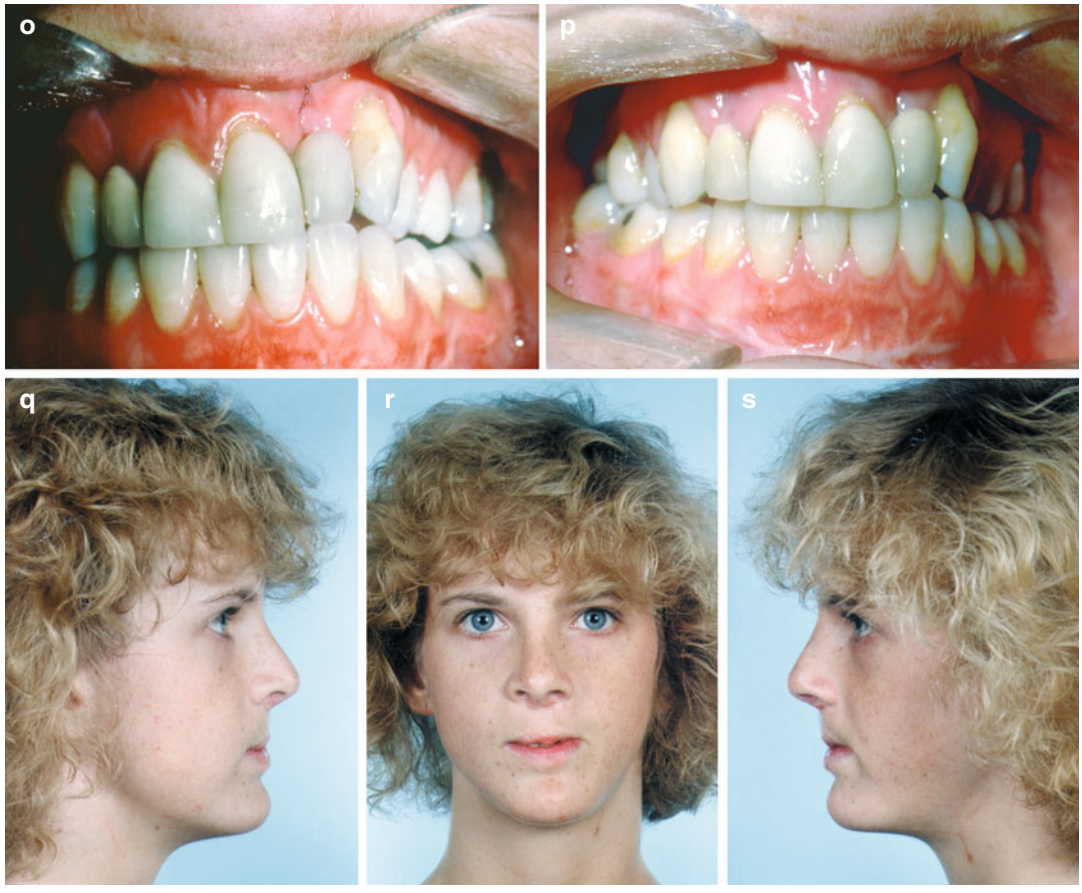

Fig. 7.38 (*continued*) (**o**, **p**) Intraoral photographs showing a fixed bridge to replace both upper lateral incisors. (**q**–**s**) Facial photographs at 17 years of age. A prominent symphysis is noted. Comment: This case shows the need to keep the lateral incisors spaces open in order to obtain good anterior overbite and overjet in the presence of strong mandibular growth

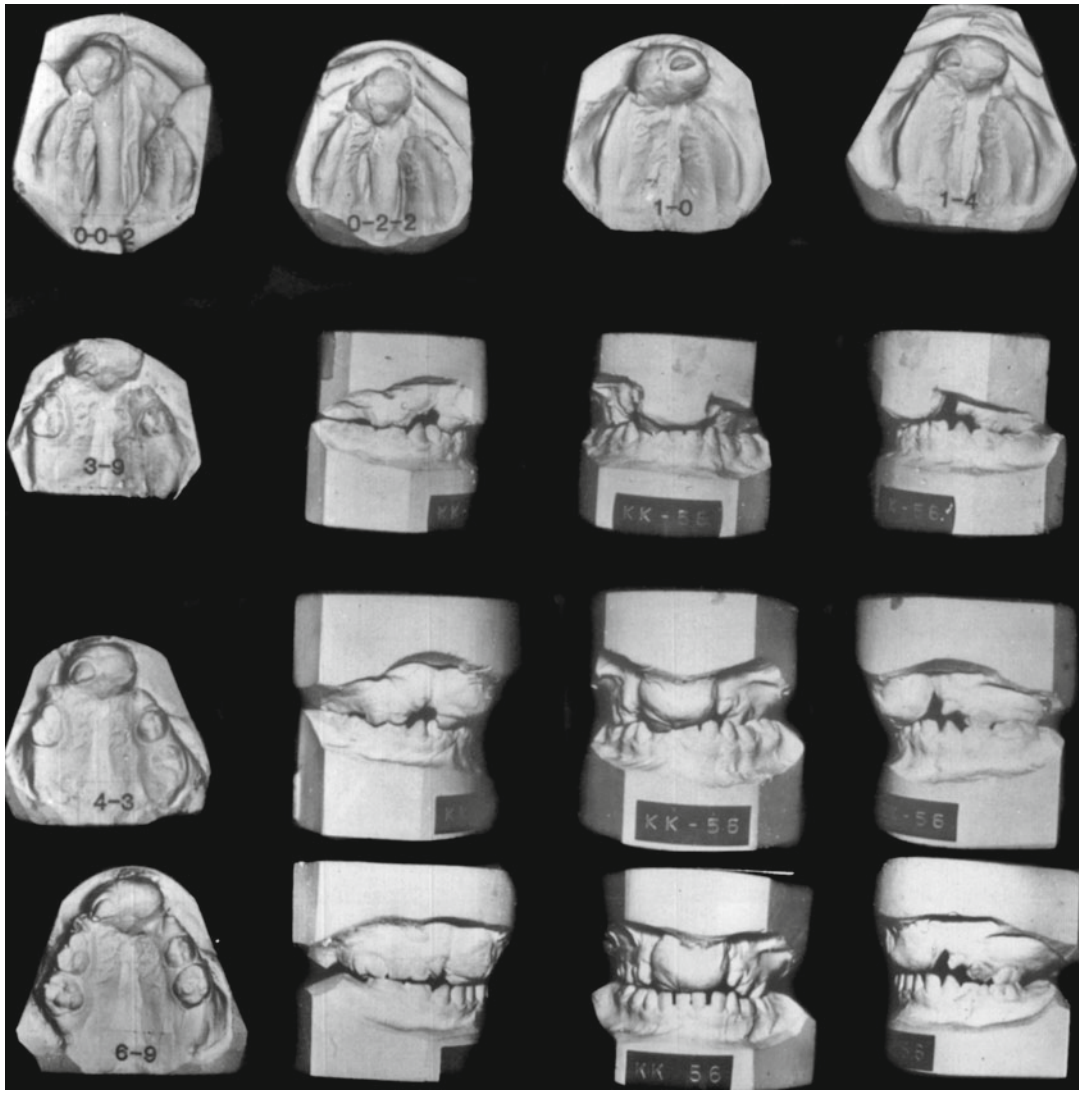

Fig. 7.39 Case ML (KK-56). Serial casts from 0–0–2 to 4–3: With the establishment of an intact lip musculature, the premaxilla and lateral palatal segments are molded into a good arch form. The premaxilla, although latero- and ventroflexed, still caused the upper lip to be pushed forward. The left buccal crossbite was corrected by 8 years of age, and a fixed palatal retainer was placed

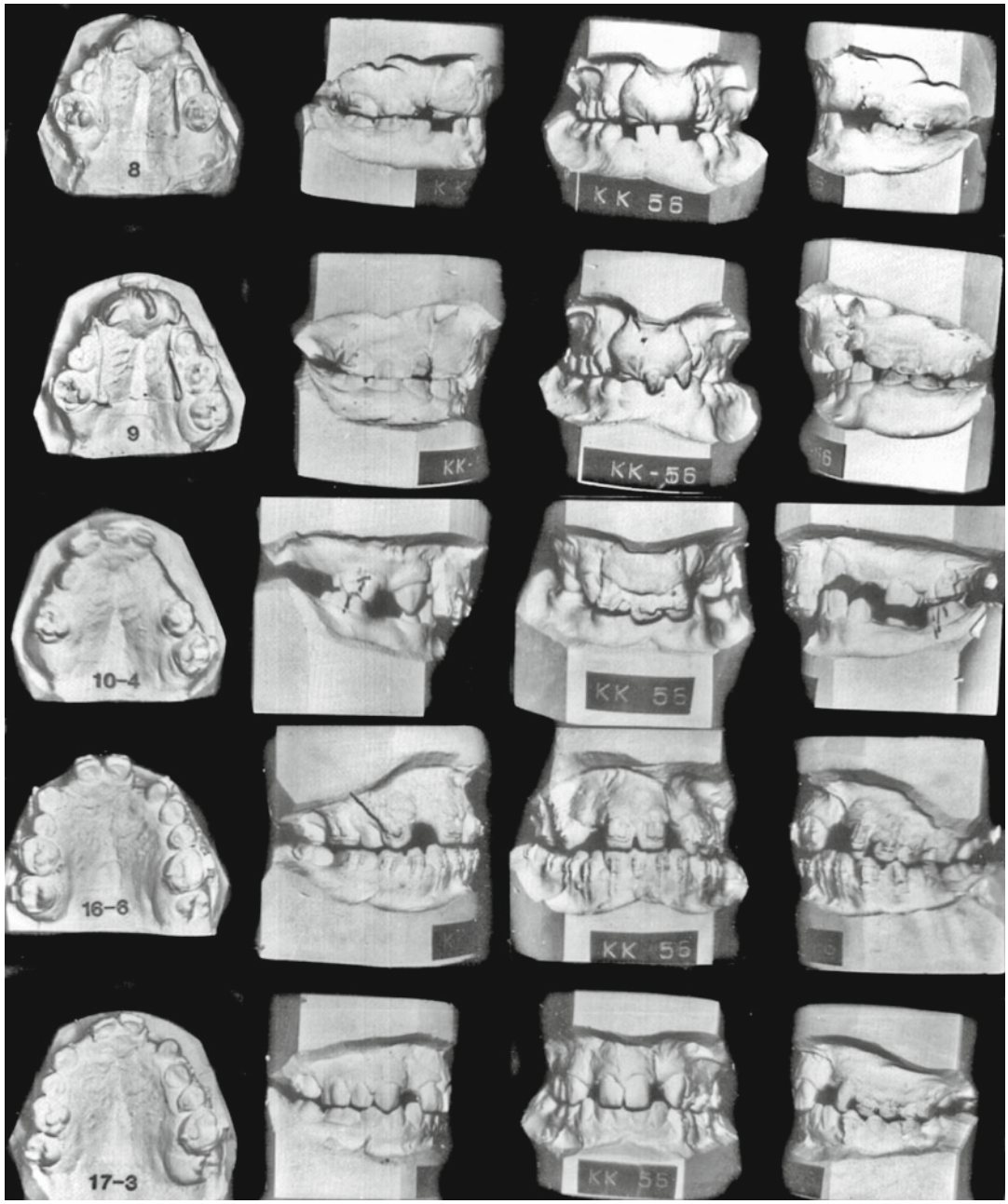

Fig. 7.39 (*continued*) 15–7 Orthodontic treatment was designed not to correct the slight Class II occlusion of the left segment. 15–9 After Lefort I osteotomy and final teeth alignment. Because the premaxilla was positioned slightly to the right and could not be centered orthodontically, it was decided to leave the left occlusion in Class II and the right occlusion in Class I, thereby equalizing the space for the lateral incisors. 17–0 A cuspid-to-cuspid fixed bridge replaced the missing lateral incisors and stabilized the relationship of all segments

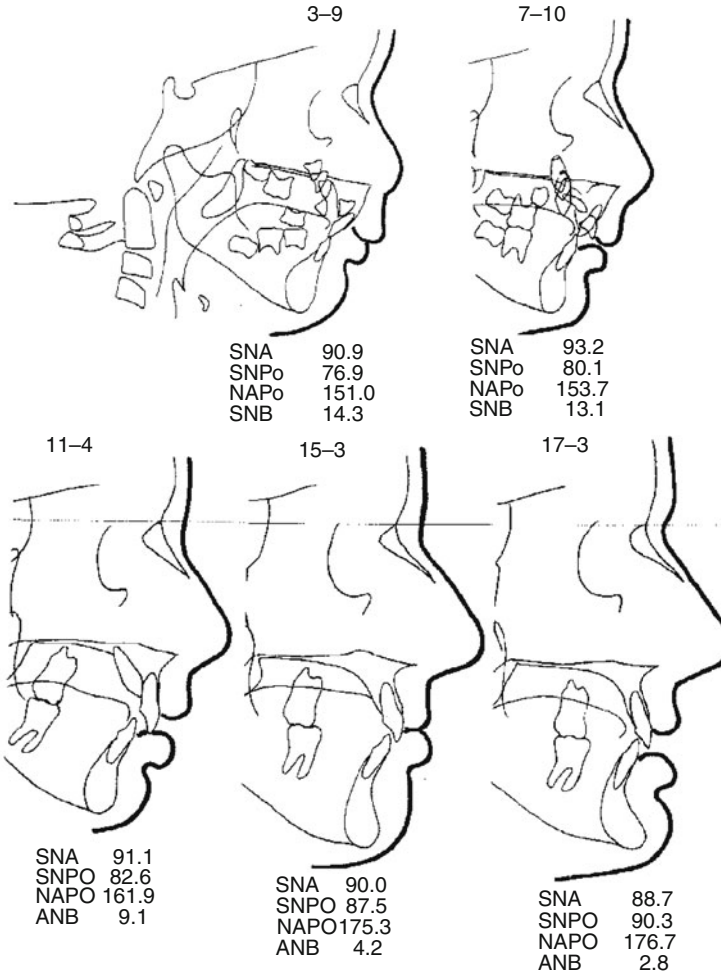

Fig. 7.40 Case ML (KK-56). Serial cephalometric tracings showing well-proportioned facial growth with a flattening of the facial profile. The protrusive premaxilla was present at 7–10. Orthodontia at 11 years of age improved the axial inclination of the maxillary incisors. The profile at 15–3 is more attractive than that at 17–3 as a result of the chin augmentation at 15–7. The chin point is too protrusive, resulting in a prominent sublabial fold

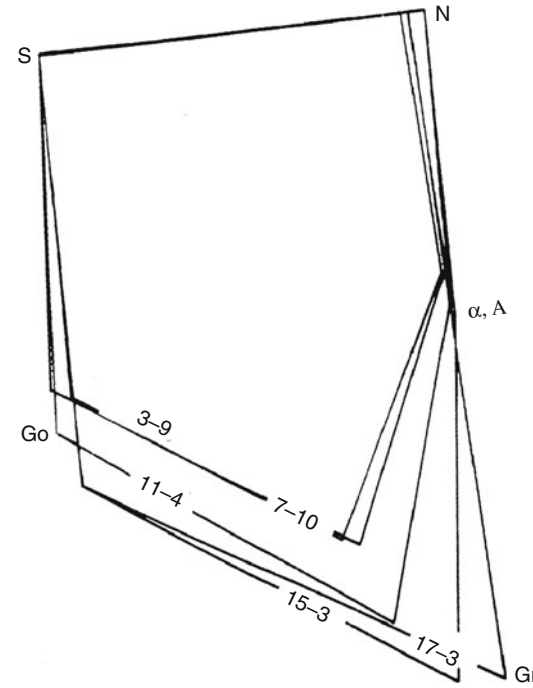

Fig. 7.41 Case ML (KK-56). Superimposed polygons show an excellent facial growth pattern and flattened the profile by 15–3. Midfacial osteotomy corrected the maxillary asymmetry. A chin augmentation was performed at 15–7 years which created a too prominent chin. The mandible continued to grow until 17–3 years of age, creating a slightly concave skeletal profile. The patient is considering having the chin prominence reduced. Comment: Rarely should a chin augmentation be performed with a LeFort I advancement to avoid creating a "dished in" face if the midfacial advancement relapses. Note the small forward growth increments at the anterior cranial base and midface. The midfacial changes did show good vertical growth to maintain normal facial proportions

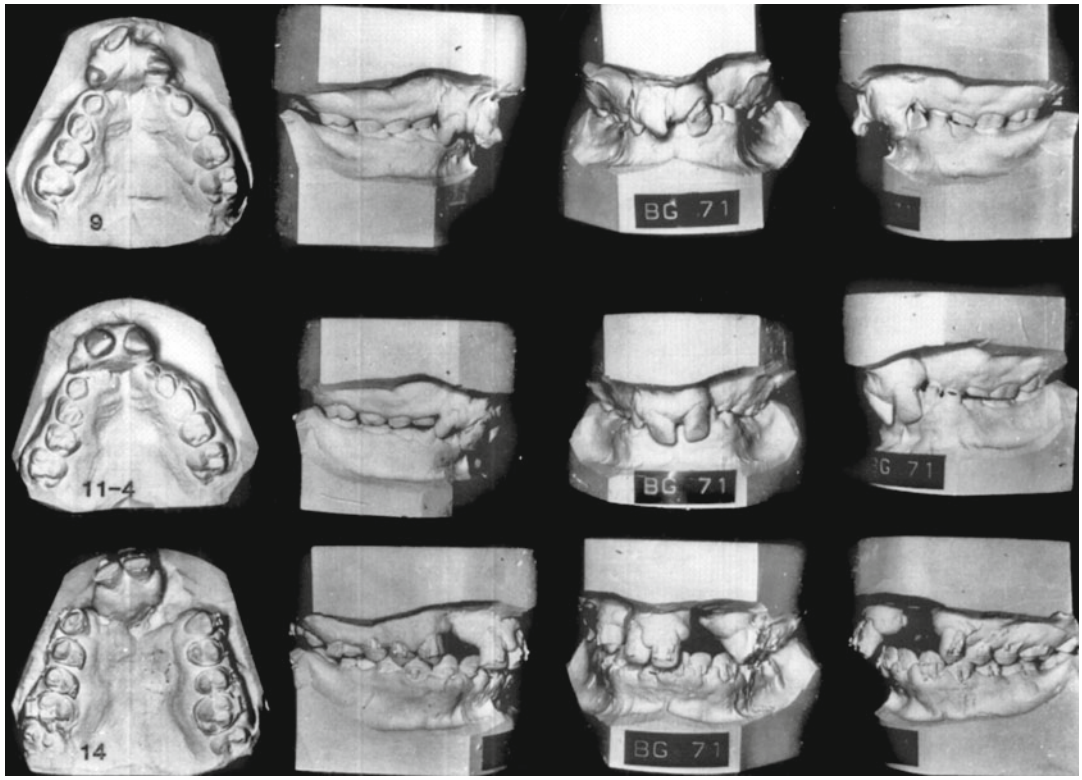

Fig. 7.42 Case CW (BG-71). Serial casts demonstrate forward advancement of the buccal segments to reduce a very large anterior palatal cleft space. There are instances when a marked osteogenic deficiency exists in both lateral incisor areas where the premaxilla is in a slight overjet-overbite relationship and the buccal segments in a good Class I relationship. In these cases, there may be insufficient contiguous soft tissue to close the anterior palatal cleft space when performing a secondary alveolar bone graft and create a normal site for tooth replacement. The treatment of choice is to advance both buccal segments, simultaneously placing secondary alveolar bone grafts, yet leaving space for the lateral incisors. After surgery, the cuspids were to return to their original Class I position. It was believed that with early premaxillary setback, there would be inadequate soft tissue to obtain adequate cleft closure, but worst of all, it would have created a severely retruded midface which would have required midfacial advancement

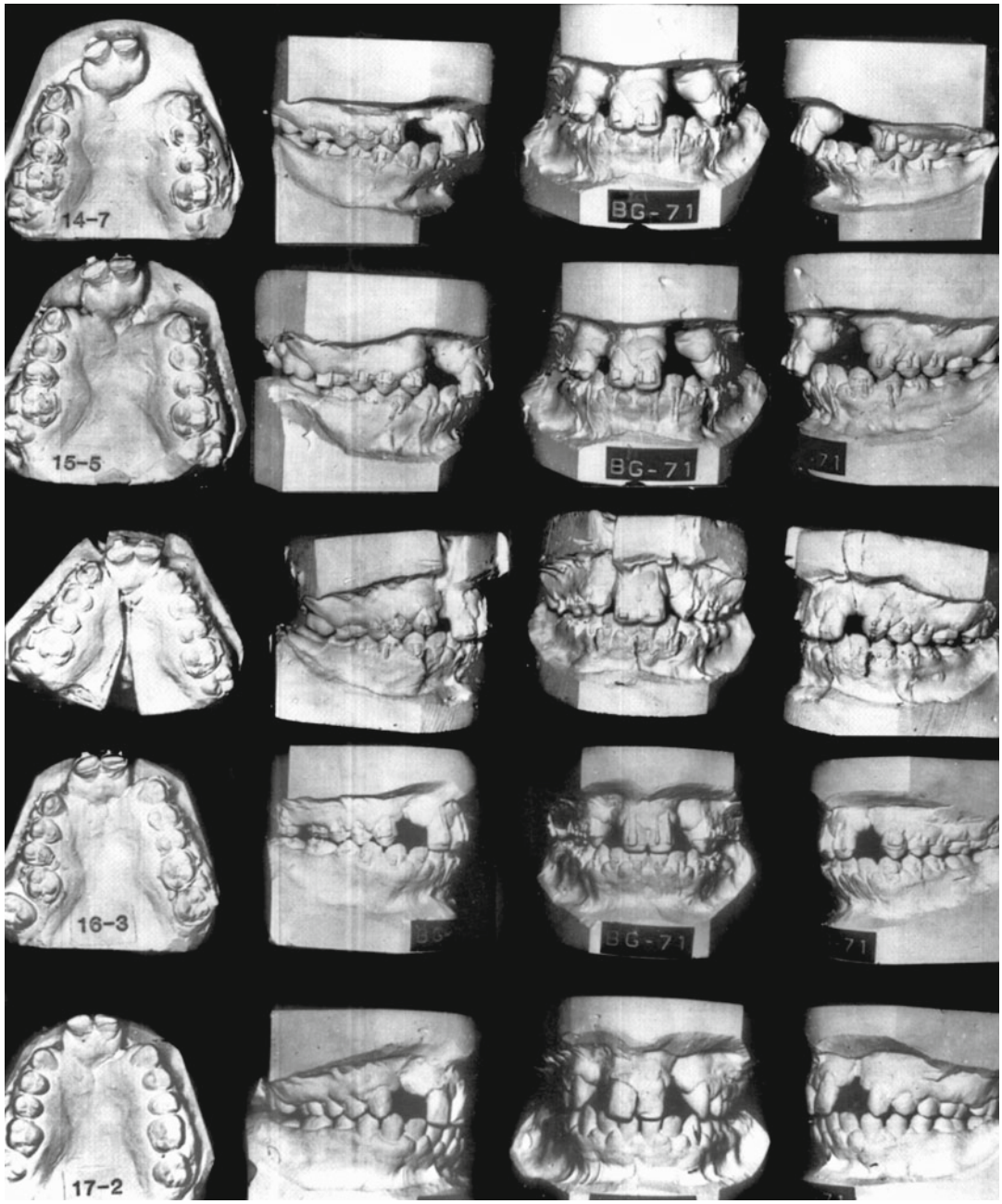

Fig. 7.42 (*continued*) 15–5 Good premaxillary relationship with a large anterior palatal cleft space. Class I posterior occlusion. Sectioned plaster casts with the posterior segments placed in a Class II relationship. 16–3 and 17–2 Both buccal segments relapsed into Class I. The main objective of closing the anterior palatal cleft spaces was achieved. Final casts show a good Class I occlusion with a satisfactory overjet-overbite relationship. The anterior palatal cleft space was closed. However, the alveolar bone graft did not take. In most cases, the advanced lateral palatal segments will remain forward with the cuspids in the lateral space

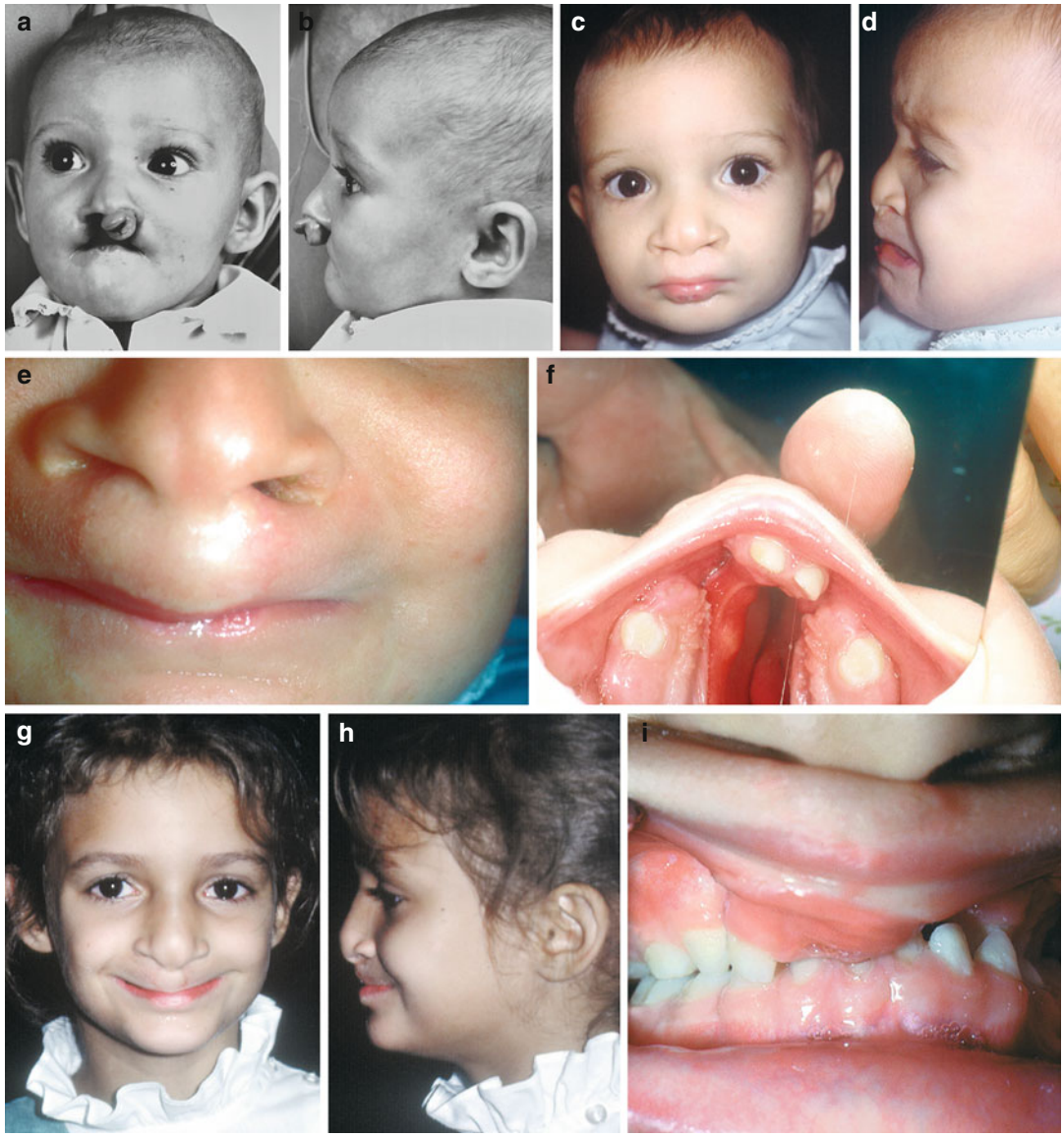

Fig. 7.43 (**a**–**z**) Case CS (AF-48) demonstrates severe premaxillary protrusion in a child with CBCLP at birth, resulting in maxillary retrusion in adolescence with the eventual loss of the premaxillary incisors. Lip adhesion was performed at 3 months with forked flap and posterior palate cleft closure at 3 years. No secondary alveolar bone grafts were utilized. Premaxillary surgical advancement at 15 years to correct its retrusion. The premaxillary incisors were extracted due to severe periodontal bone loss, and the anterior palatal oronasal opening was closed at 16 years of age. (**a**, **b**) Newborn. (**c**–**i**) Even with premaxillary ventroflexion, the upper lip was still pushed forward

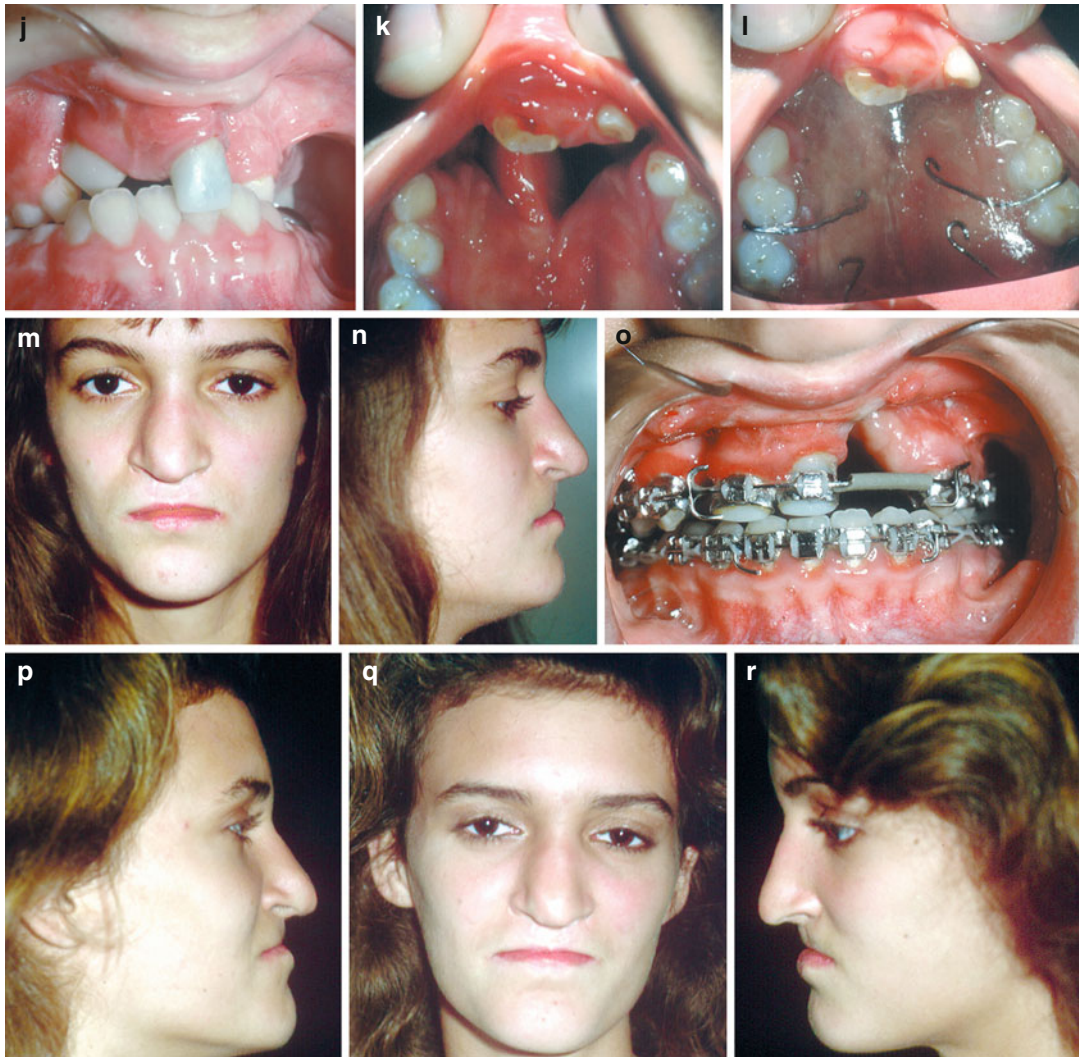

Fig. 7.43 (*continued*) (**i–l**) A plastic obturator was utilized at 6 years of age to close the very large anterior palatal cleft space to aid speech development and feeding. (**m–o**) At 14 years, a retrusive-looking midface with an extremely tight upper lip and depressed nasal tip masked a good premaxillary overjet

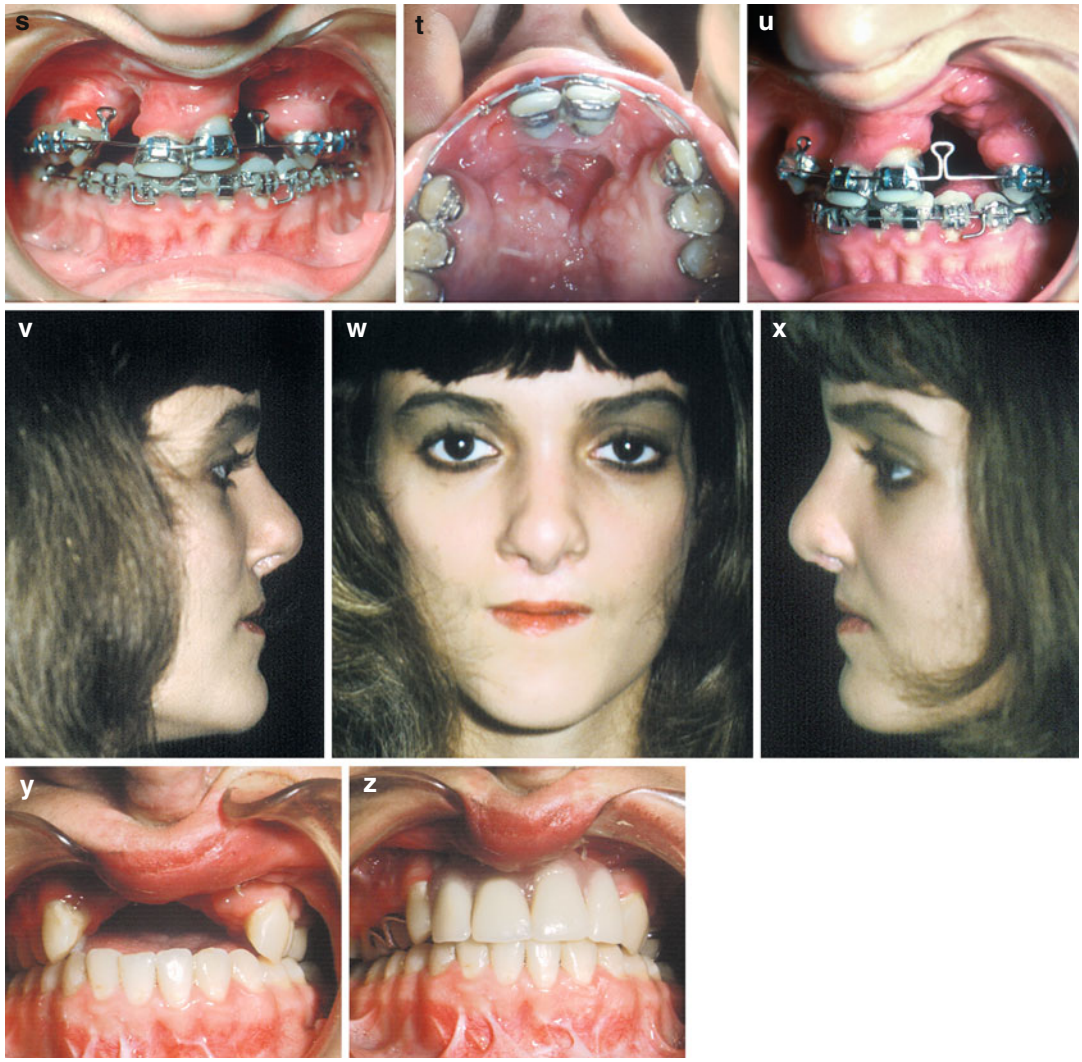

Fig. 7.43 (*continued*) (**u**) At 15 years following an unsuccessful attempt to surgically center the premaxilla and close the anterior cleft space. The maxillary incisor roots began to show severe external root absorption. A very large anterior cleft space remains. (**v**–**y**) After lip and nose revision, the anterior teeth were extracted and the oronasal opening closed with adjoining soft tissue. (**z**) A removable maxillary prosthesis replaced missing teeth and bumpered the upper lip forward. Comments: It would have been better treatment to have surgically advanced both palatal segments to close the very large anterior palatal cleft space. The root absorption was secondary to traumatic orthodontics utilized to maintain the incisor overjet with a protective facial mask and cross elastics

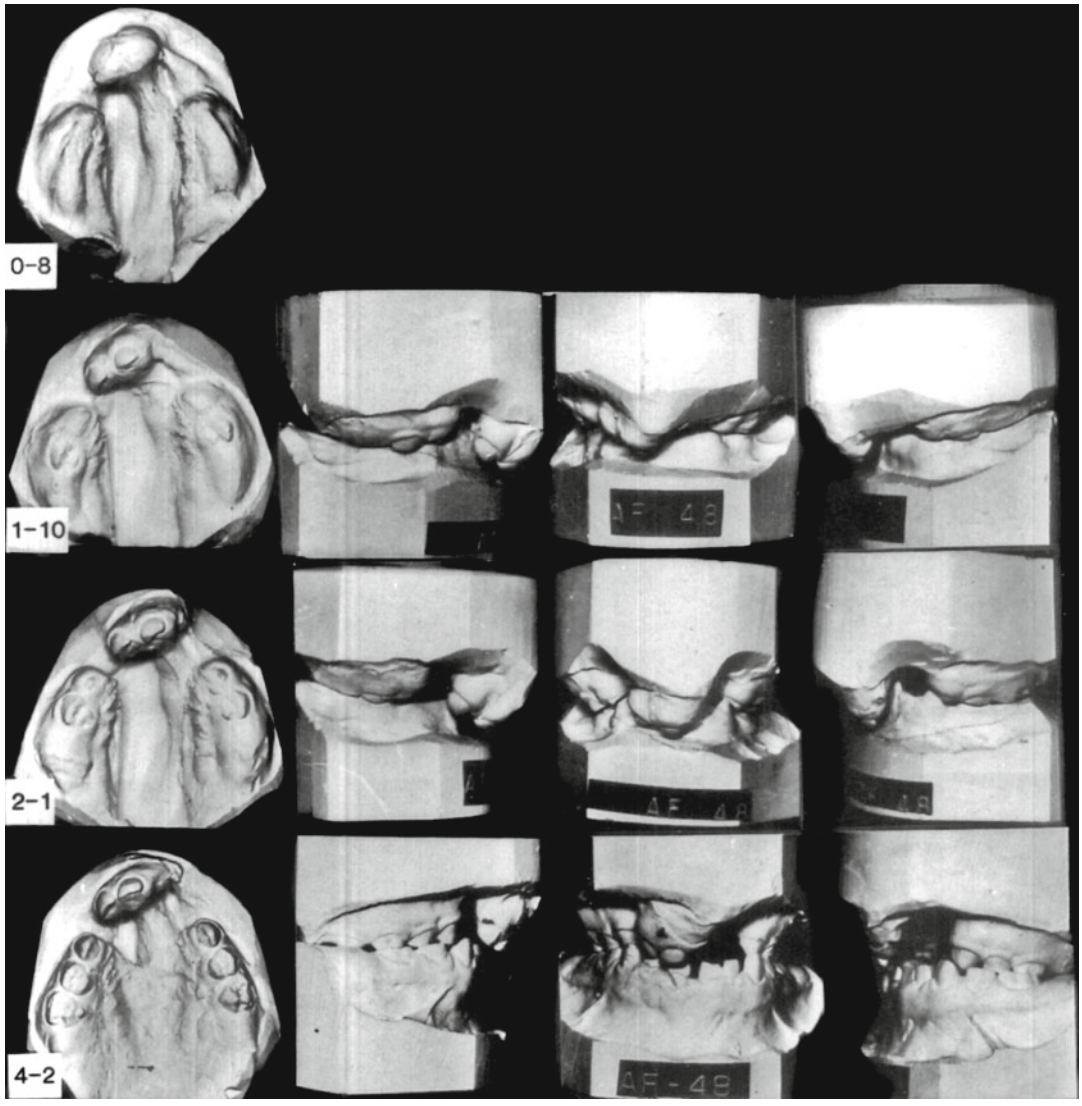

Fig. 7.44 Case CS (AF-48). Serial casts show that the extreme premaxillary protrusion with large anterior cleft space at birth is still present at 6–1 years of age

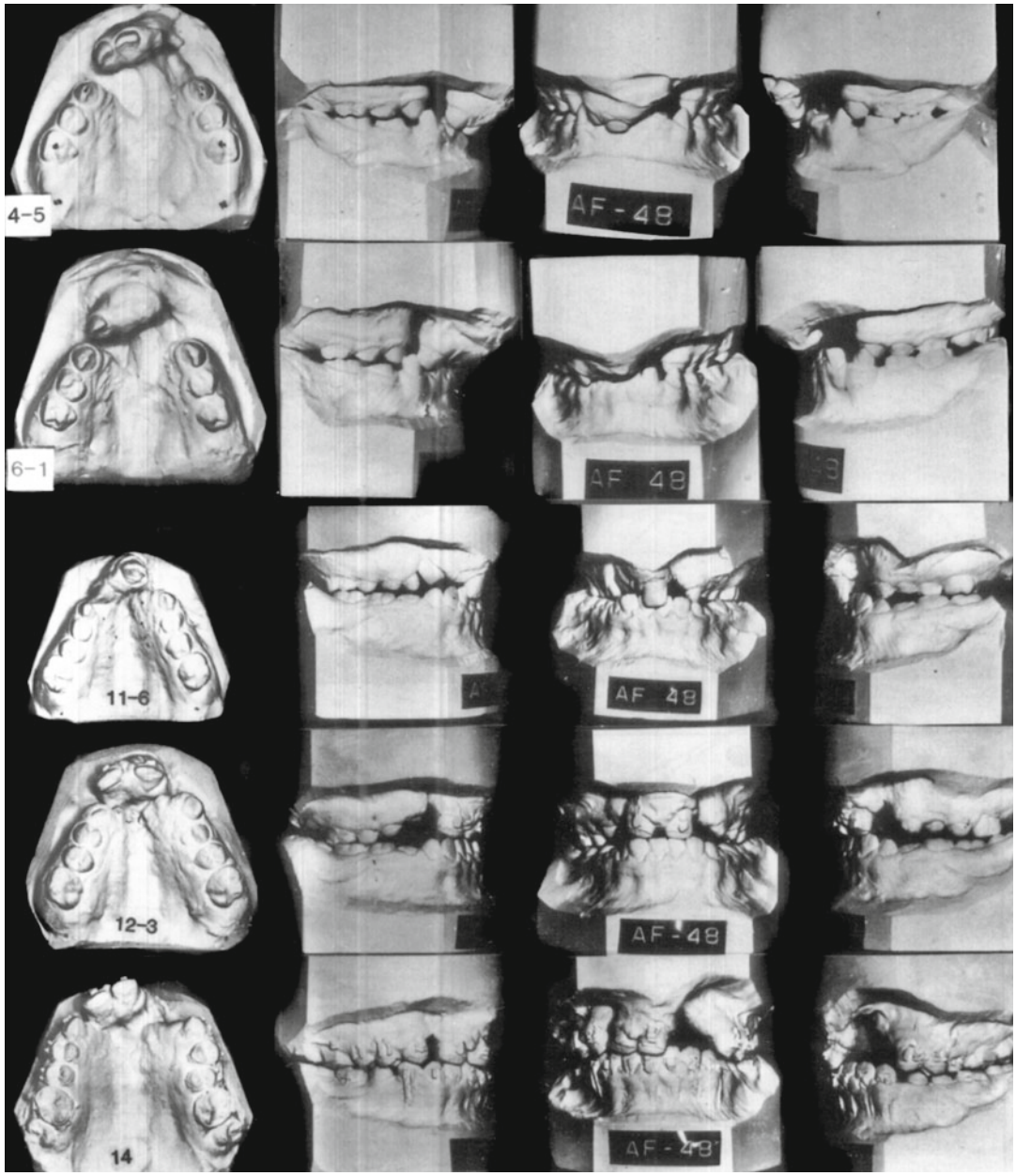

Fig. 7.44 (*continued*) 11–6 With increased facial growth, the increasing tonicity of the buccal muscle forces collapsed the maxillary arch placing the posterior teeth in crossbite. The premaxilla is now upright and in an acceptable overbite-overjet relationship. 12–3 Maxillary expansion has been initiated. The maxillary central incisors are in tip-to-tip relationship. 14 Continued orthodontic treatment to advance the premaxilla and position the incisor teeth in proper overjet-overbite relationship

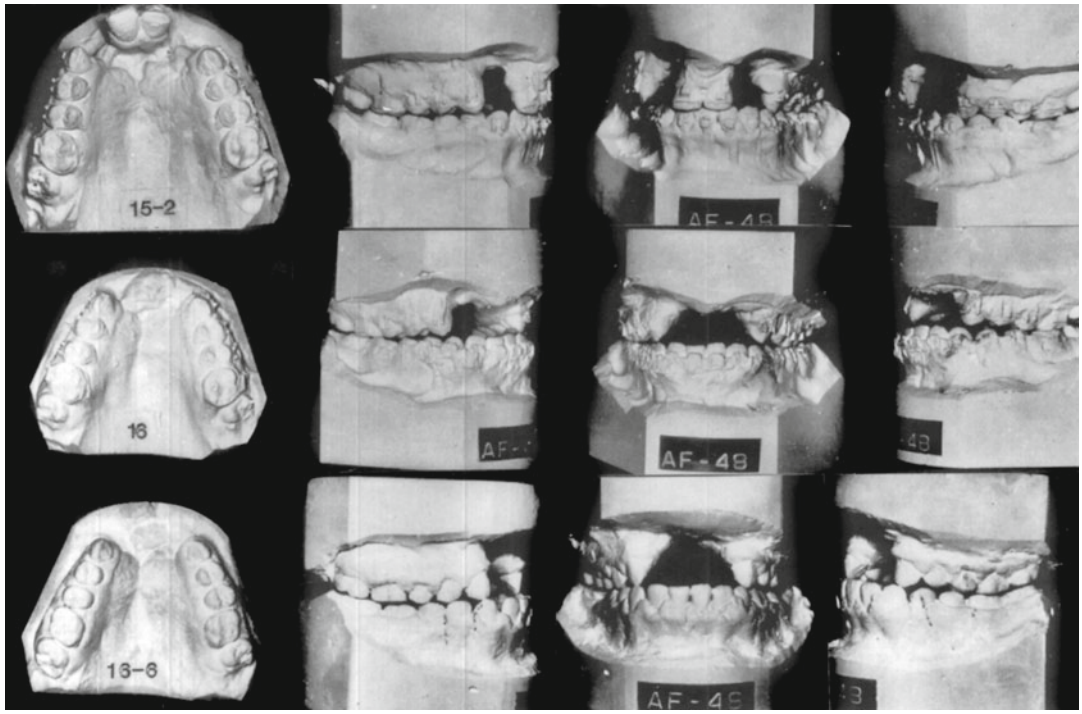

Fig. 7.44 (*continued*) 15–2, 16, and 16–6 Premaxillary repositioning with soft tissue closure of the anterior cleft space was unsuccessful. As a result of the premaxillary central incisors showing external root absorption and loss of periodontal support, they were extracted and the oronasal opening closed with adjacent soft tissue. A removable maxillary prosthesis replaces the missing teeth and bumpers the upper lip. Comment: As already suggested, the treatment of choice is to reposition the lateral palatal segments anteriorly while leaving the premaxilla as is

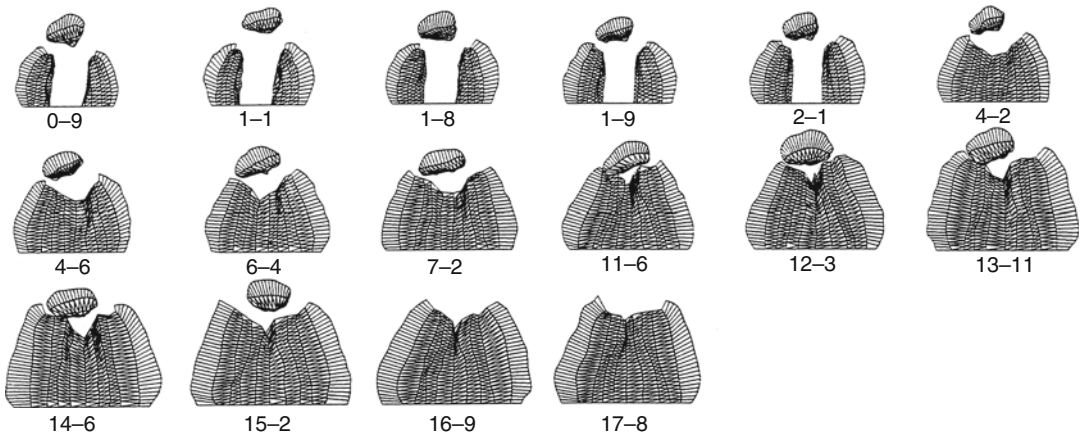

Fig. 7.45 Case CS (AF-48). Computerized tracings of serial casts drawn to scale. This shows the lack of palatal growth and reduction in cleft space over 2 years prior to surgical closure of the palatal cleft. The anterior cleft space remains large up to 15–2 years. 16–9 and 17–8 The premaxillary incisors were extracted. Comment: This case clearly demonstrates the severe degree of osteogenic deficiency that can exist in bilateral clefts of the lip and palate. The once protruding premaxilla can become retrusive with growth (time) and may eventually need to be brought forward. Although palatal growth does occur, it may not be sufficient to appreciably reduce the posterior cleft space

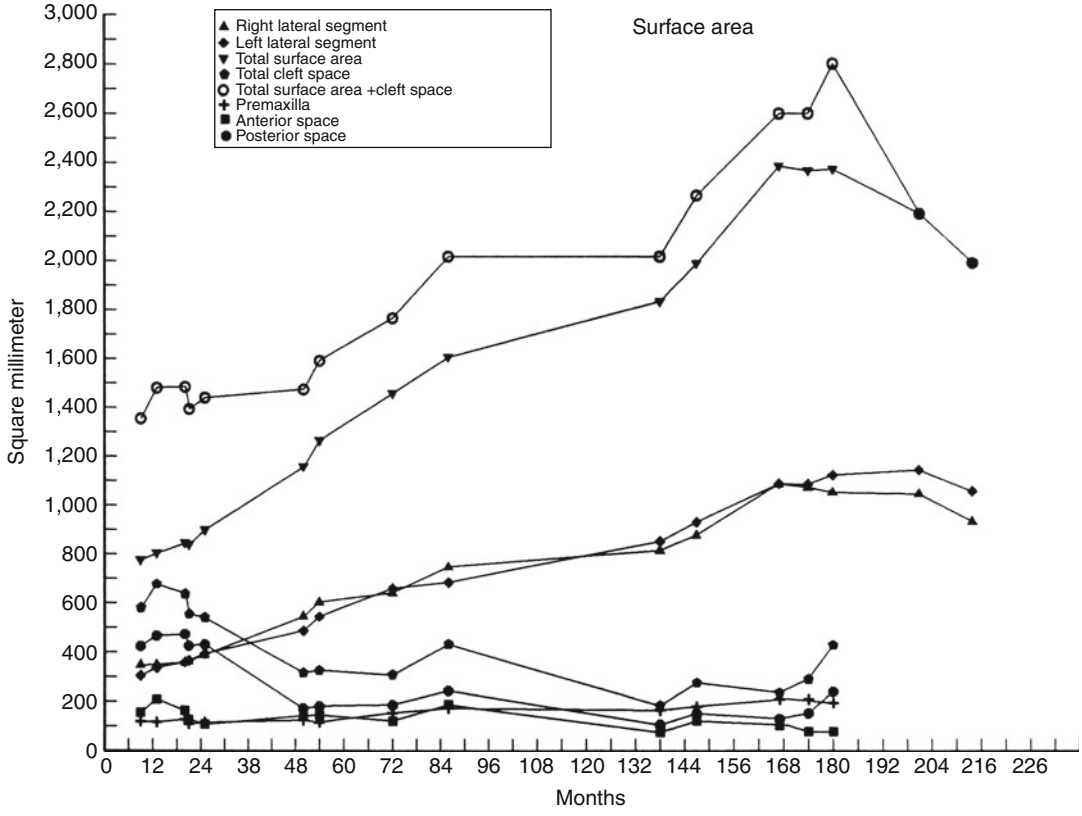

Fig. 7.46 Case CS (AF-48). The palatal segments show a very gradual growth acceleration curve, while the posterior cleft space gradually reduces in size

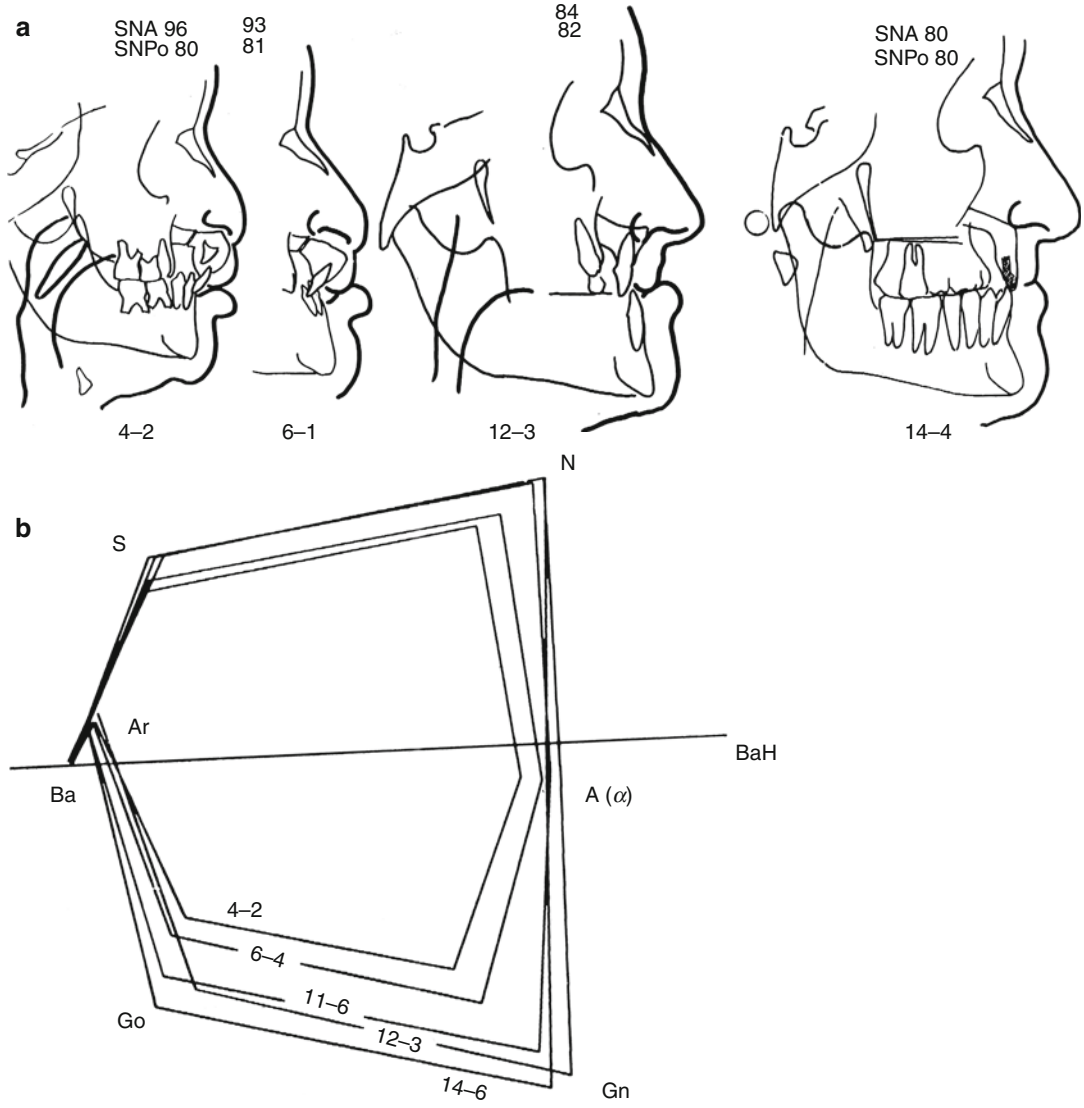

Fig. 7.47 (**a**, **b**) Case CS (AF-48). (**a**) Serial cephalometric tracings. This analysis shows a protrusive midface at 4–2 becoming recessive at 14–4. (**b**) Serial facial polygons superimposed according to basion horizontal method (Coben). The midface advanced only slightly after 6–4, while the mandible showed progressive downward and forward growth until 14–6, flattening the facial profile. Comments: This case clearly shows that (1) even with a severely protruding premaxilla at birth, the premaxillary incisors can be in anterior crossbite after the pubertal growth spurt; (2) traumatic orthodontic advancement of the premaxillary incisors can lead to external root absorption and loss of alveolar support; (3) surgical advancement of one or both lateral palatal segments, placing the cuspids in the lateral incisor spaces, with secondary alveolar bone grafting is the treatment of choice in cases when the premaxilla is in good overjet-overbite relationship and a large anterior cleft space exists. Only in very rare instances should the premaxilla be surgically set back to the lateral palatal segments; and (4) Premaxillary surgical setback is contraindicated prior to the postpubertal facial growth spurt

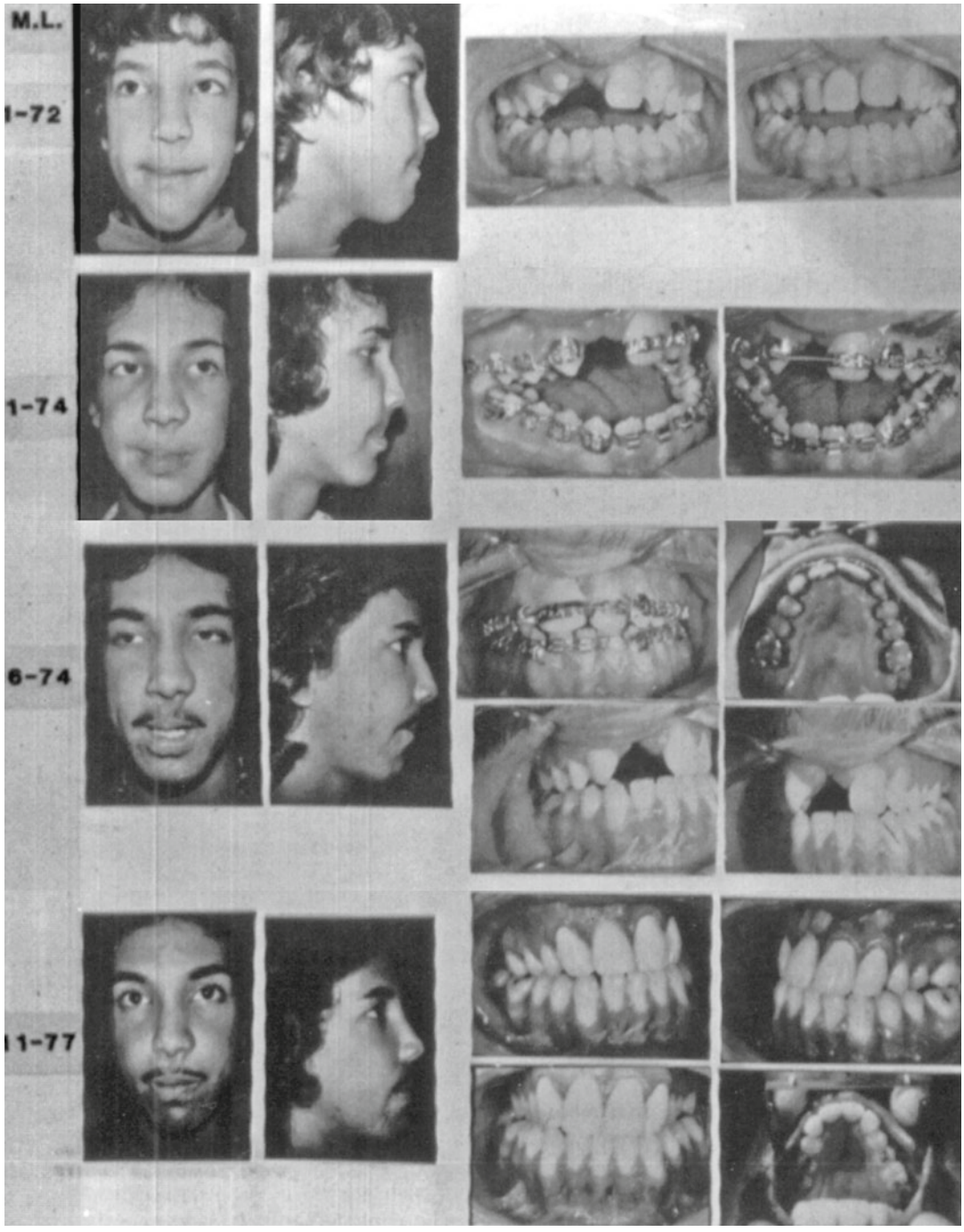

Fig. 7.48 Case ML demonstrates poor facial growth pattern in a BCLP leading to a retruded midface with a severe anterior open bite. This patient came to the clinic at 5 years, 7 months of age with an anterior open bite. The upper lip was long and tight with the lateral elements brought together below the prolabium. The palatal cleft was closed at 12 months of age. At 13–10, a removable plate replaced the right central incisor. At 15–11, orthodontic preparation for Lefort I posterior impaction. At 16–4, after maxillary surgery. At 19–9, after anterior fixed bridge used to stabilize the palatal segments and replace missing teeth. The changes in occlusion and total facial height led to a more relaxed soft tissue profile. With reduction in lower vertical facial height, the upper to lower lip position became more aesthetic

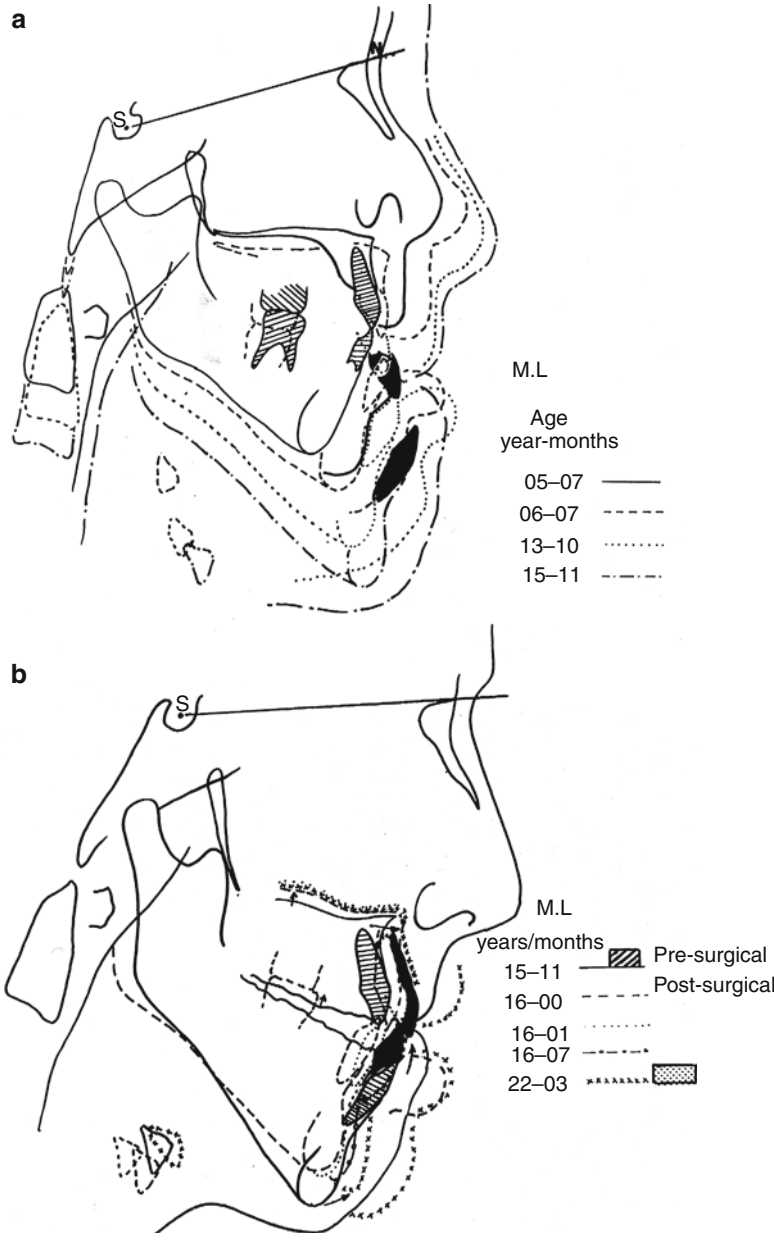

Fig. 7.49 (**a**, **b**) Case ML. Superimposed serial cephalometric tracings on SN and registered at S before surgery. (**a**) Between 5–7 and 15–11, the face demonstrated a very poor growth pattern associated with diminished midfacial growth. The incisors were in tip-to-tip relationship at 5–7; by 6–7, the anterior open bite with a retruded midface was present, which became worse with time. (**b**) Serial cephalometric tracing superimposed on SN and registered at S after posterior impaction and maxillary advancement. The mandible autorotated closing the anterior open bite. The curvature to both lips became more prominent and aesthetic. Comment: Placing the lateral lip elements below the prolabium creates a long and tight upper lip. Maxillary growth disturbance is always expressed in all three dimensions, resulting in diminished vertical and horizontal growth increments. Maxillary advancement with slight posterior impaction causes mandibular autorotation, bringing the lower incisors upward and anteriorly closing the anterior open bite. The middle and lower vertical facial heights became more harmonious

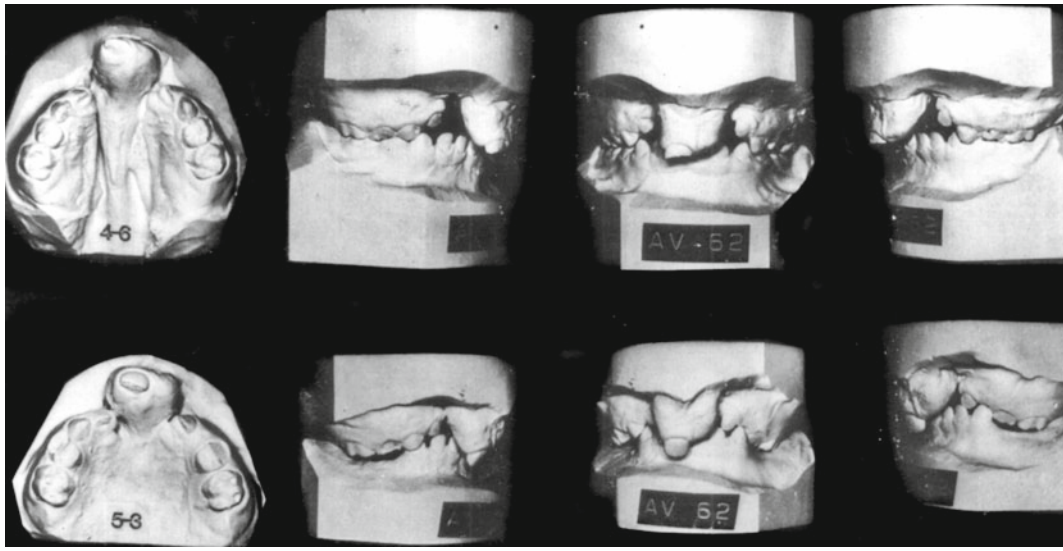

Fig. 7.50 Case CT (AV-62) demonstrates a severe premaxillary overjet leading to a retruded midface and the successful use of protraction orthopedics. This patient was adopted from Central American parents. 4–6 It is important to note the wide cleft space with an excellent Class I buccal occlusion prior to surgical cleft closure. 5–3 Two months after palatal cleft closure, the occlusion is still acceptable

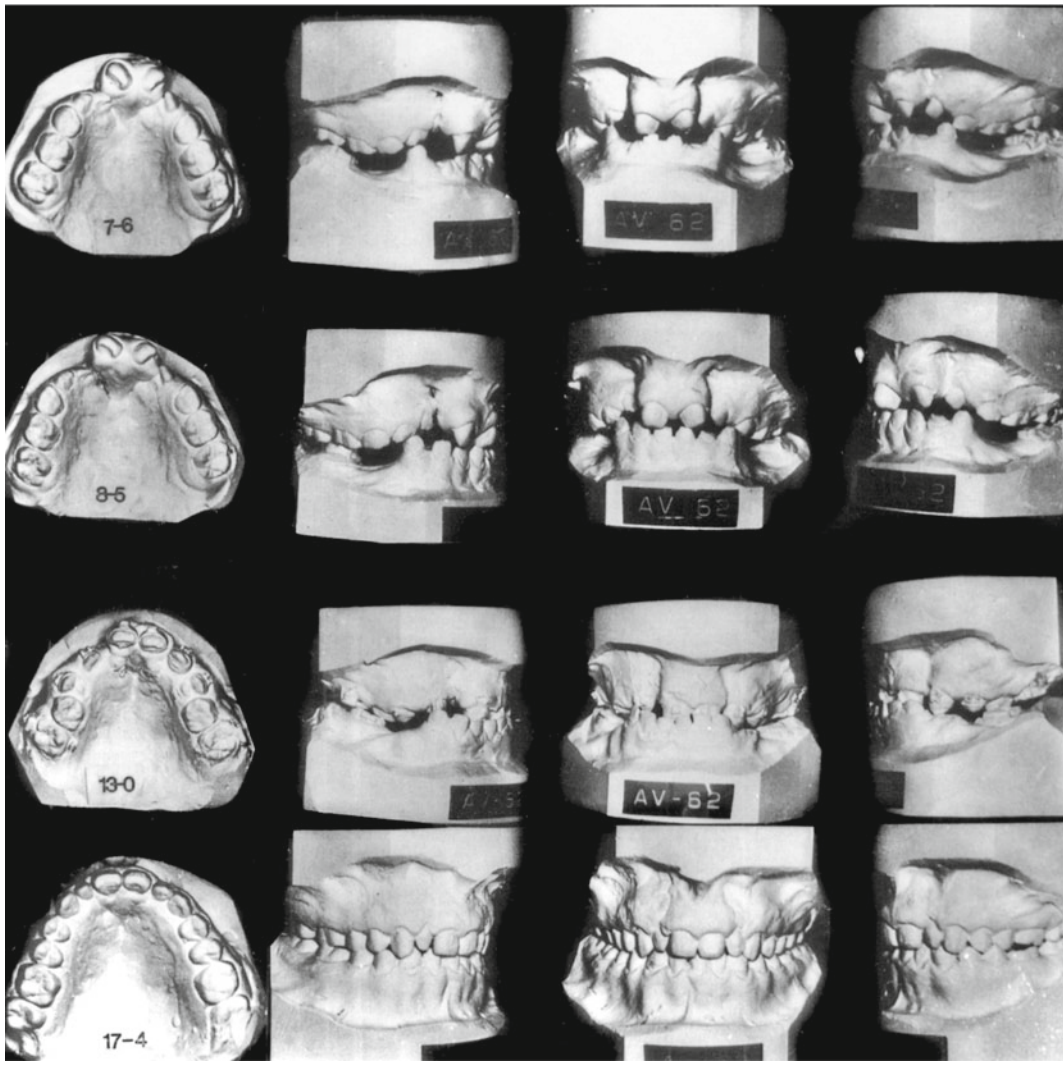

Fig. 7.50 (*continued*) 7–6 and 8–5 A secondary alveolar bone graft was placed at 8–0. The anterior teeth are in a tip-to-tip relationship with the buccal occlusion in crossbite. 13–0 Complete anterior and buccal crossbite. Preparation is made to begin orthodontic treatment to expand the maxillary arch as well as to advance the maxillary complex with protraction mechanics. 17–4 After orthodontic treatment. A lower central incisor was extracted to allow for incisor retraction. Excellent occlusion is retained with a Hawley appliance. Comment: This case clearly demonstrates that, in complete clefts of the lip and palate, the lateral palatal segments at birth can be in good relationship with the lower arch and not collapsed, as McNeil suggests. In most cases, the surface area of the lateral palatal segments can be considered to be too small when compared to the cleft size. Too early cleft closure using a von Langenbeck procedure will leave large areas of uncovered bone when the mucoperiosteum is moved medially. Denuded bone undergoes epithelialization with scarring, which in turn interferes with palatal growth in all three dimensions, leading to the buccal and anterior crossbite. In this case, protraction mechanics were successful. However, because the ideal overjet-overbite relationship could not be achieved, it was necessary to remove a lower central incisor to retract the anterior segment. This benefited the buccal occlusion as well

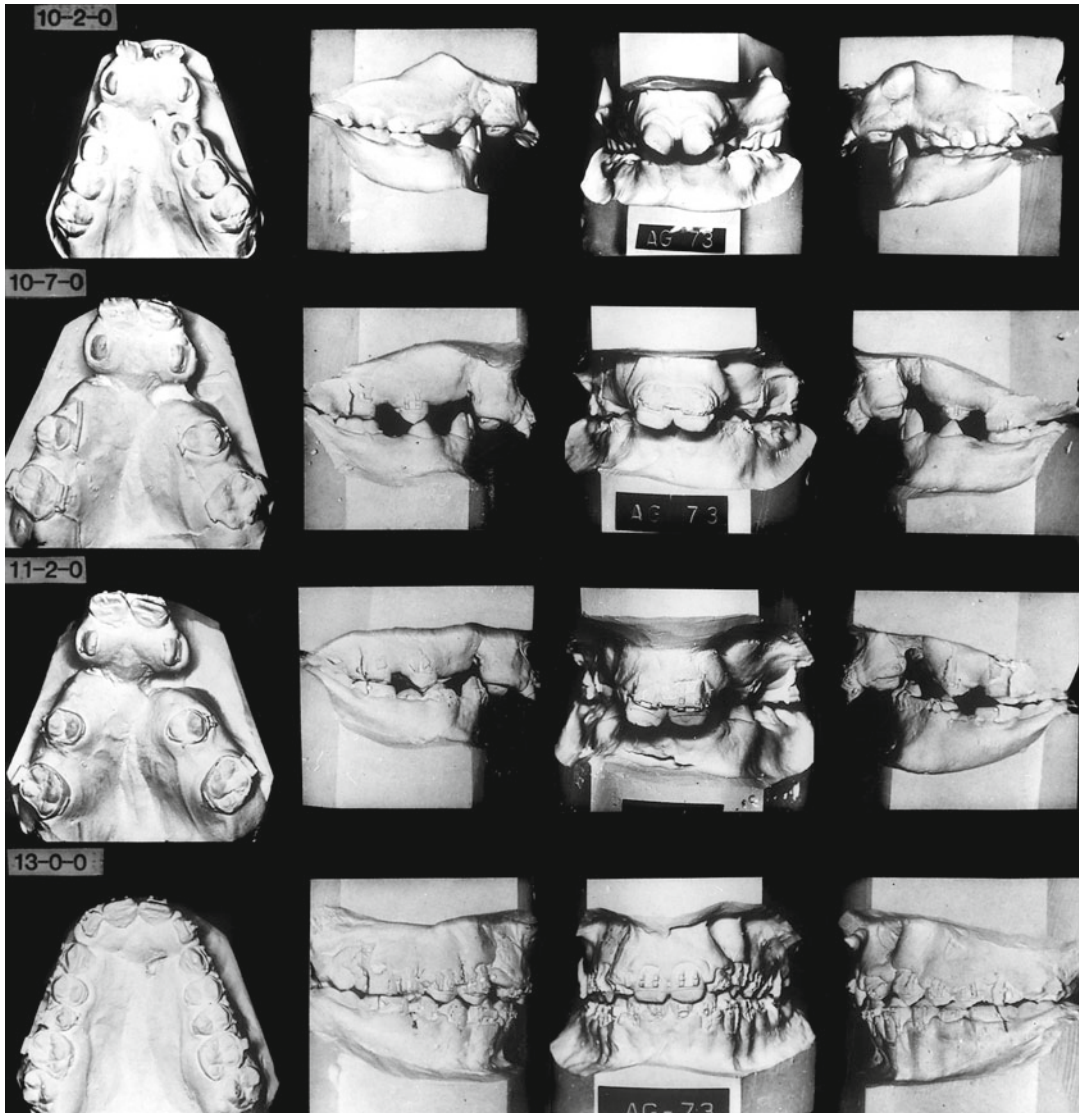

Fig. 7.51 Case RB (AG-73). Serial dental casts. Closure of a large anterior cleft space in a BCLP by bringing impacted cuspids into position and aligning the lateral incisors. 10–2 Class II malocclusion in the mixed dentition with a severe overjet and overbite. Bilateral deciduous cuspid crossbite with no anterior cleft space. 10–7 With loss of the deciduous cuspid and its supporting alveolar bone along with the horizontal impaction of the permanent cuspids, there was deficient alveolar bone between the lateral palatal segments and the premaxilla, creating a large anterior cleft space. 11–2–0 Waiting for the eruption of permanent bicuspids before initiating orthodontics. 13–0–0 After the horizontally impacted cuspids and their supporting alveolar bone and the lateral incisors were brought within the arch, the premaxillary overjet was reduced. Both of these factors were responsible for the closure of the anterior cleft space. This was followed by a secondary alveolar cranial bone graft to close fistulae and remaining cleft spaces

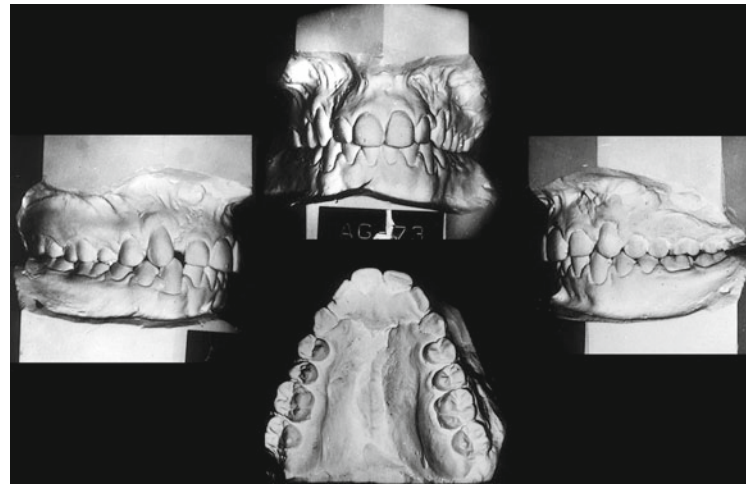

Fig. 7.51 (*continued*) 13–2 Final occlusion with a full complement of teeth. Comment: The clinician must always consider the importance of the alveolar bone supporting the teeth whether they are in alignment or malpositioned. When all of the teeth in the lateral palatal segments can be accommodated within the arch, the anterior cleft space can usually be closed without an osteotomy

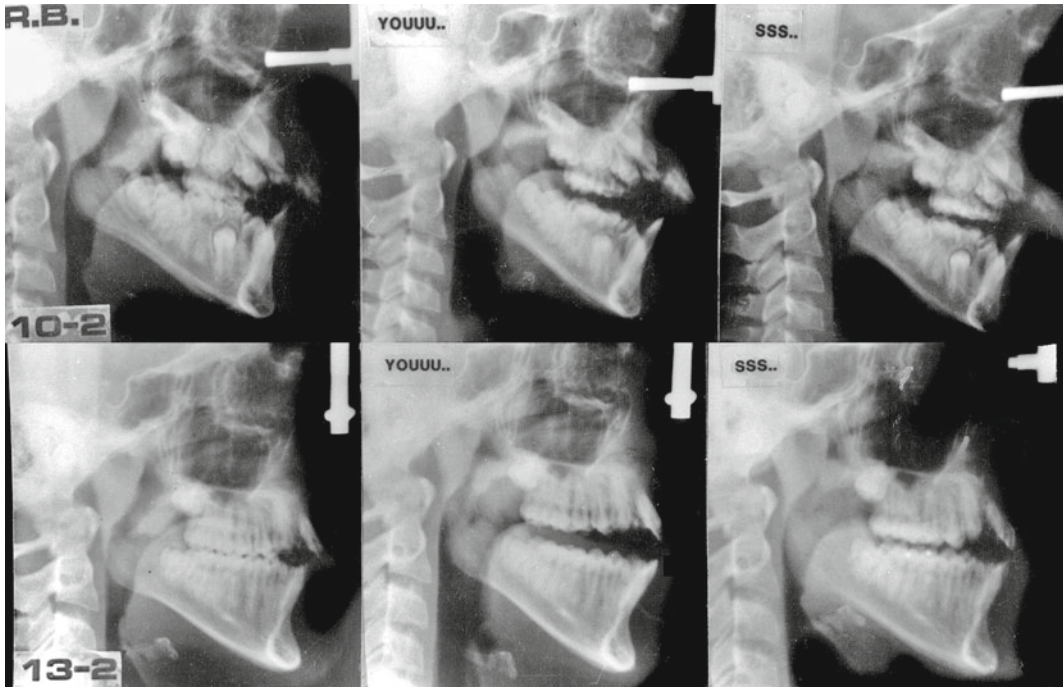

Fig. 7.52 Case RB. Cephalometric view of the pharyngeal at rest and with space and velar elevation. 10–2 *Left*: At rest and when vocalizing "Youu …" Although there is good velar elevation, the velum does not make contact with the adenoid. *Right*: Vocalizing "Sss …" This sound produces velar stretch, permitting contact to be made with the adenoid. 13–2 Good velar elevation when vocalizing "Youu …" and "Sss …". Comment: Cephalometric evaluation is necessary to view the structure of the pharyngeal space. This is not a functional test to evaluate velopharyngeal competency, but it may give some insight into why velopharyngeal incompetency may exist

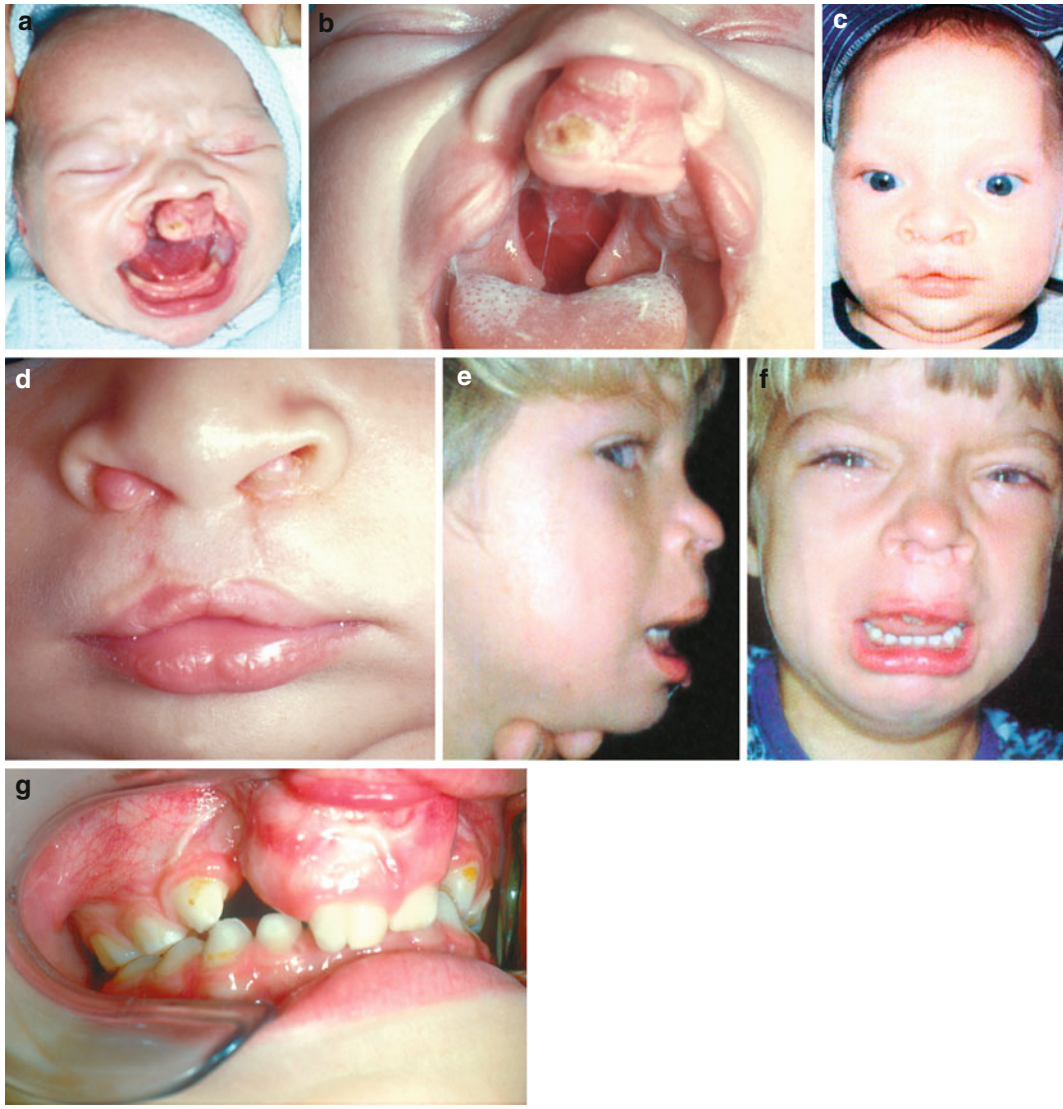

Fig. 7.53 (**a**–**r**) Case ES (EE-34) demonstrates excellent facial growth pattern in a IBCLP showing good midfacial growth. An external elastic off a head bonnet was used to ventroflex the premaxilla prior to lip surgery. The lip was united at 5 months of age using a forked flap procedure. The palatal cleft was closed at 43 months of age using a modified von Langenbeck procedure. (**a**, **b**) Face and palate at birth. (**c**, **d**) 3 months after lip surgery. (**e**–**i**) 5 years of age with excellent buccal occlusion and good overbite-overjet relationship of the anterior teeth. Upper lip is protrusive

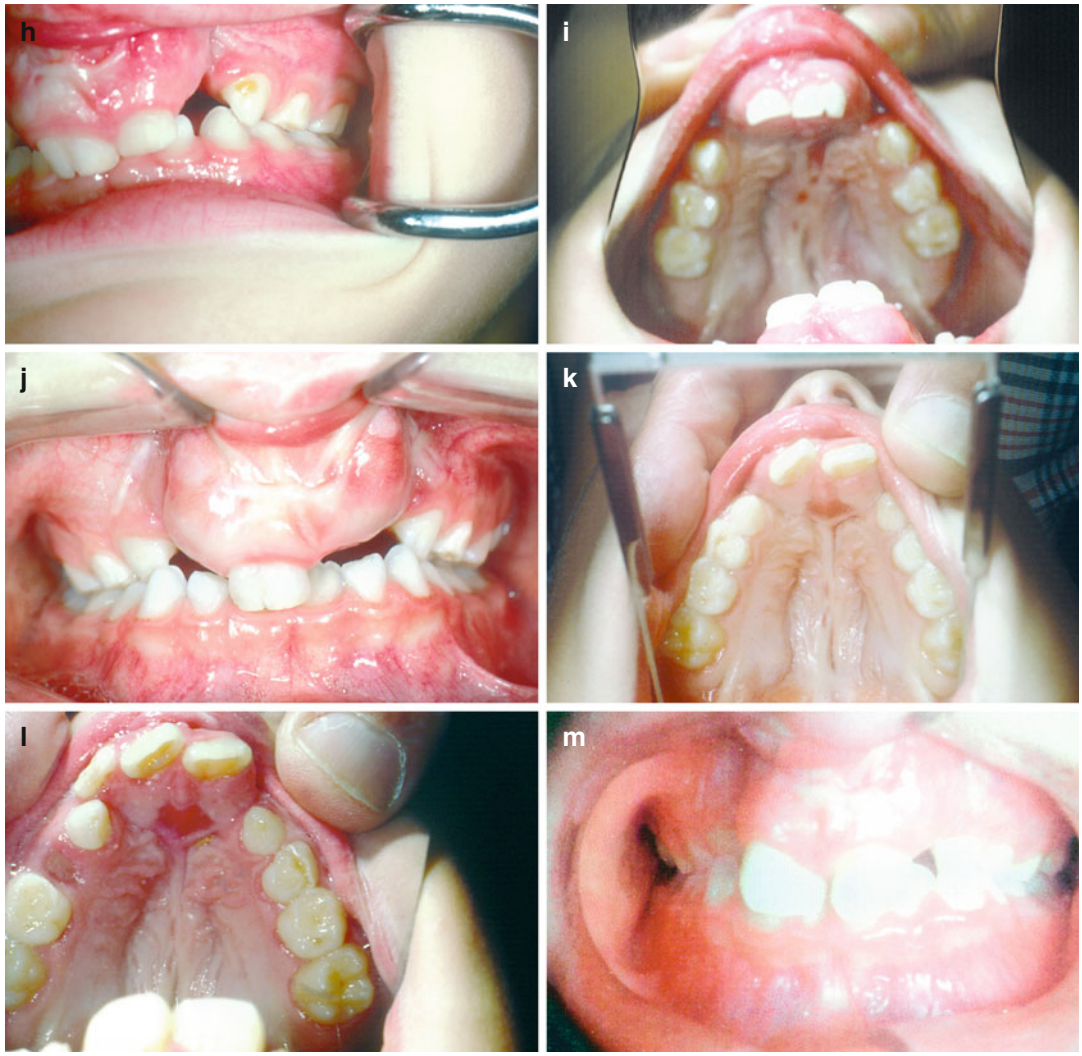

Fig. 7.53 (*continued*) (**j**, **k**) 10 years of age after secondary alveolar bone grafting. (**l**, **m**) A left cuspid crossbite resulted from secondary alveolar bone graft surgery

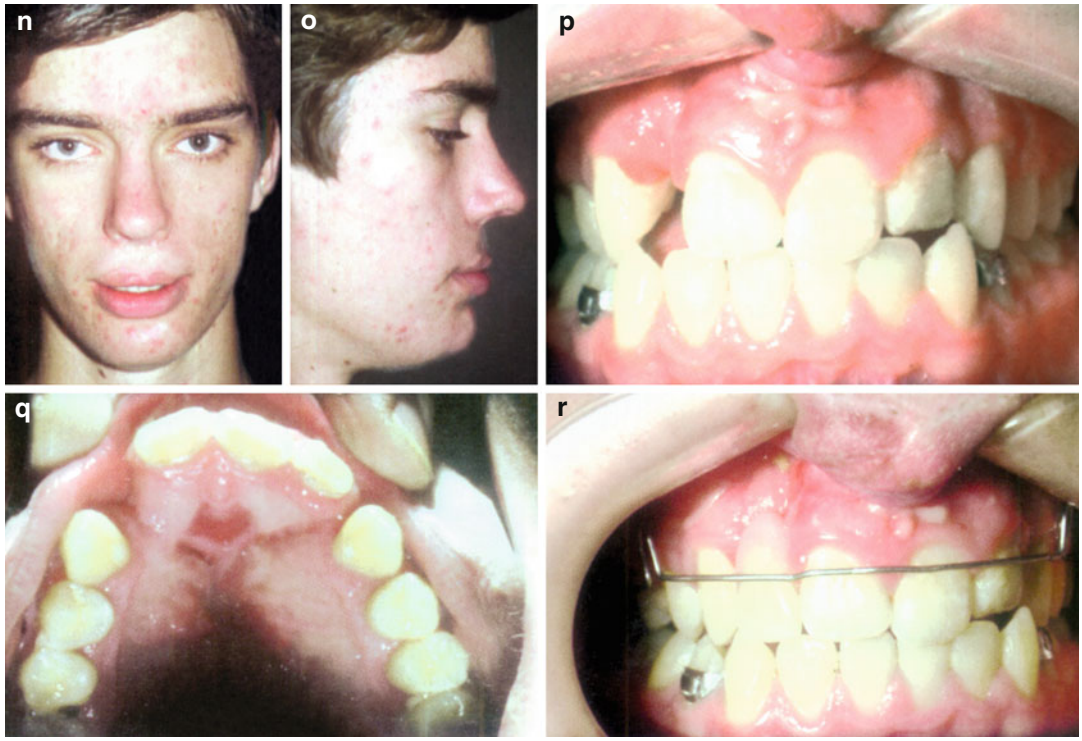

Fig. 7.53 (*continued*) (**n**–**r**) 16 years. After orthodontics was completed. The left lateral incisor erupted through the bone graft; the right lateral incisor is missing. The arch form is maintained with a removable Hawley retainer, which carries a lateral incisor replacement

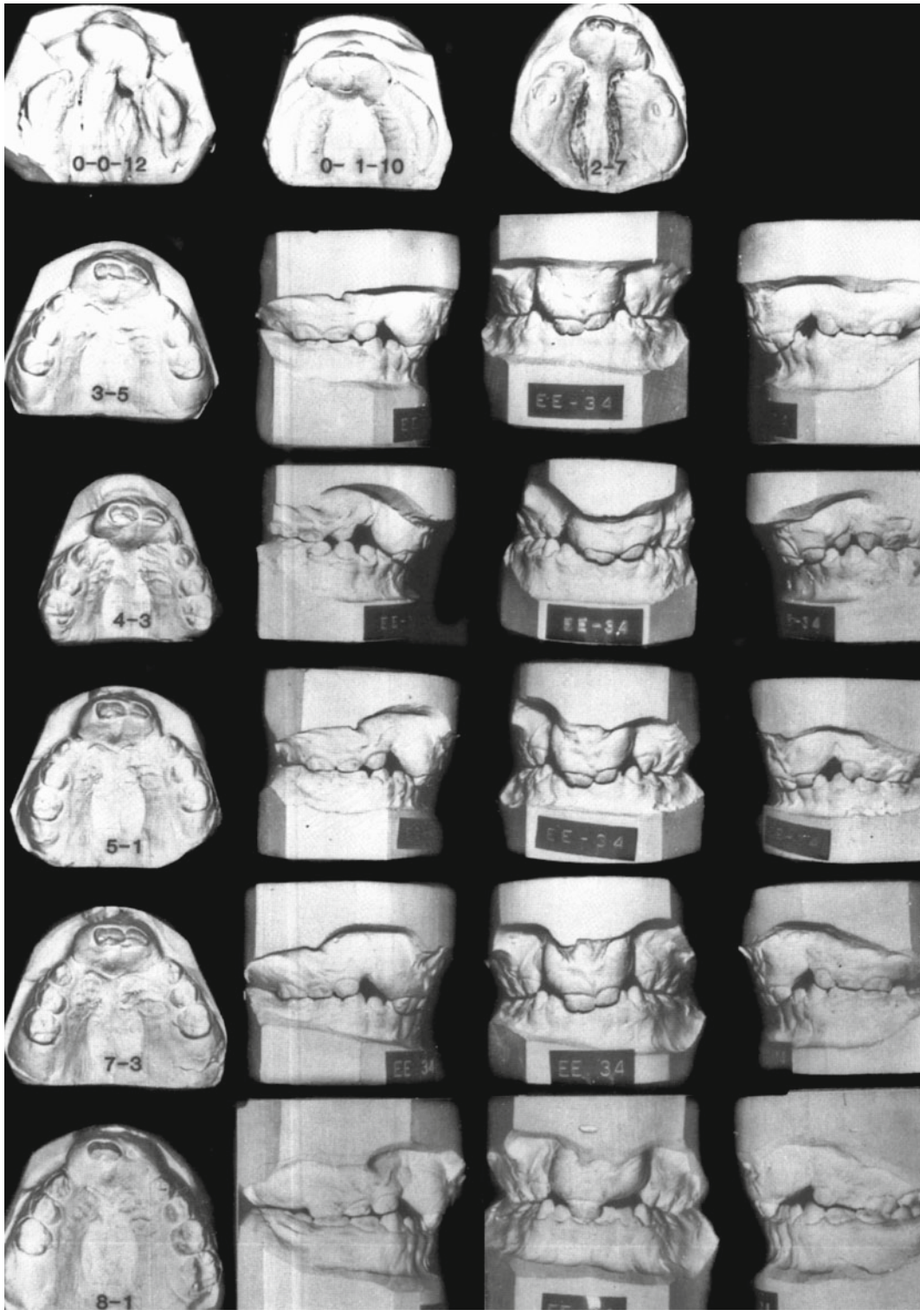

Fig. 7.54 Case ES (EE-34). Serial dental casts. 0–0–12 At birth. 0–1–0 After uniting the lip, the premaxilla has been positioned adjacent to the lateral palatal segments. 2–7 The palatal cleft is still open. Tongue force has moved the premaxilla forward. 3–5 After palatal cleft closure, there is excellent buccal occlusion. 4–3, 5–1, 7–3, and 8–1 Good buccal and anterior occlusion is present for the next 4 years

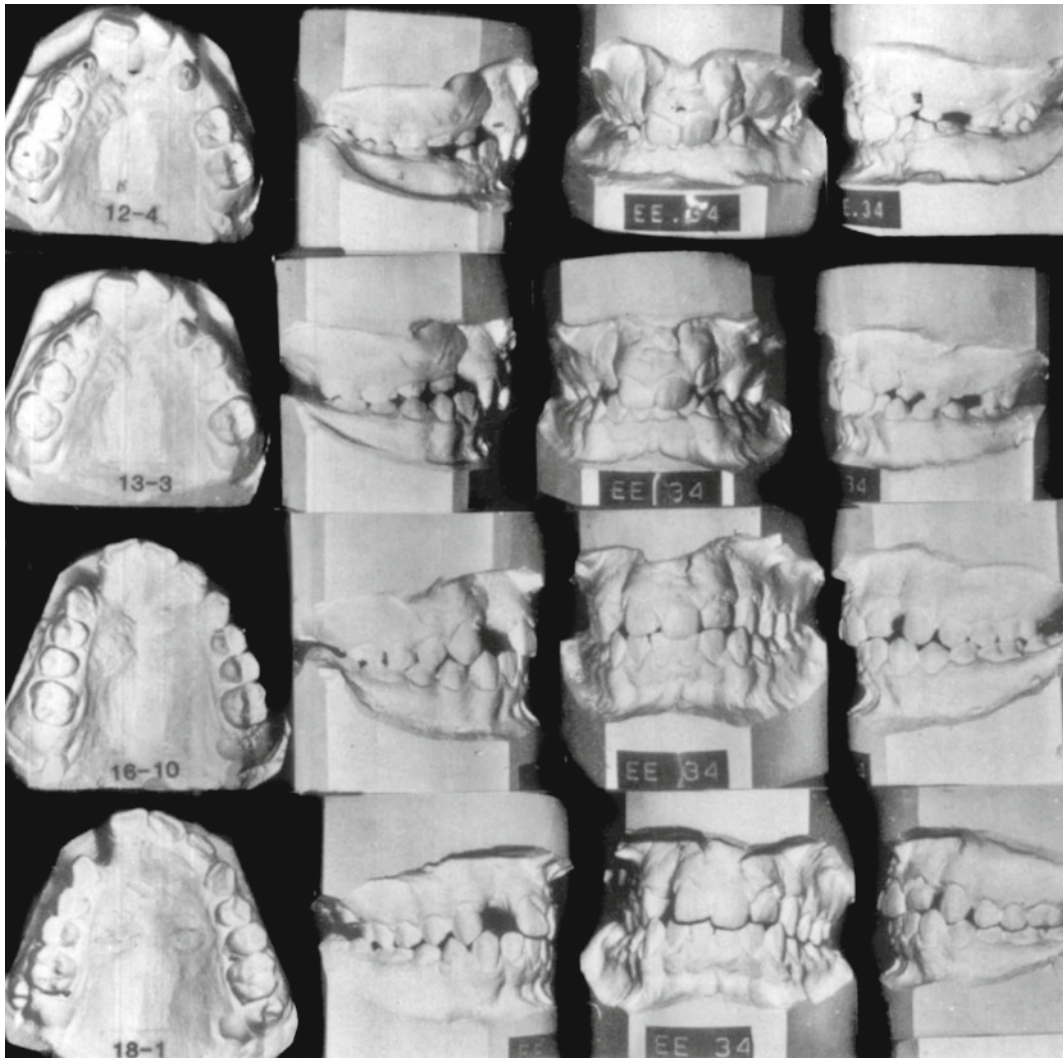

Fig. 7.54 (*continued*) 12–4 and 13–3 This crossbite remains for the next 3 years and does not hinder dental function. 16–10 and 18–1 After orthodontics. Good Class I occlusion with good anterior overjet and overbite

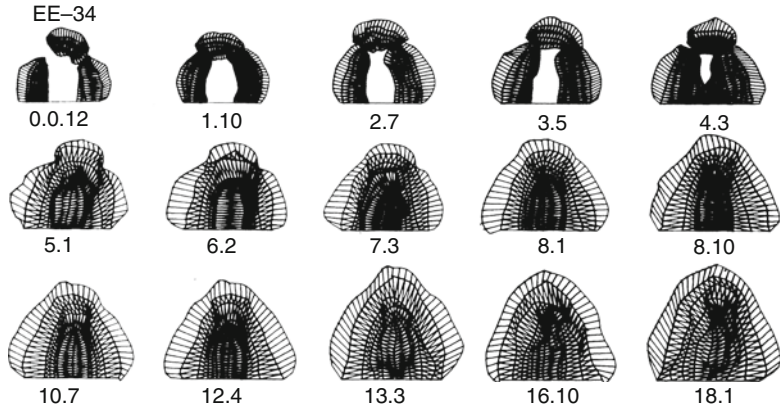

Fig. 7.55 Case ES (EE-34). Computer-generated images of serial palatal casts. All of the casts are in proportion. Palatal growth occurs on all surfaces but mainly transversely and posteriorly to accommodate the unerupted molars. Although the palatal surface is gradually increasing in size, the posterior cleft space remains almost constant in size over the same period of time. When the palatal cleft was closed at 43 months, more mucoperiosteal tissue was available to prevent growth-inhibiting scar tissue from being created

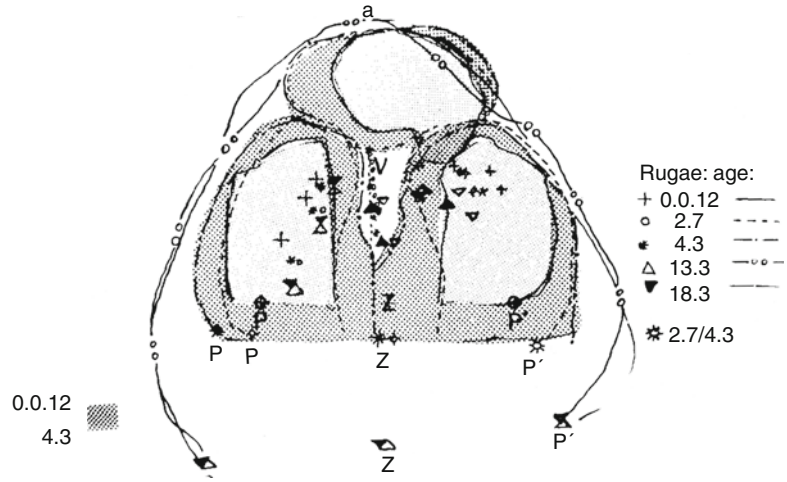

Fig. 7.56 Case ES (EE-34). Computer-generated outlines of palatal casts superimposed on the palatal rugae. This study demonstrates that palatal growth occurs mainly in the posterior and transverse areas with very little premaxillary growth. More significantly, it shows that the premaxilla is relatively stable in its geometric position within the maxillary complex, that is, the face grows up to and around the original position at birth

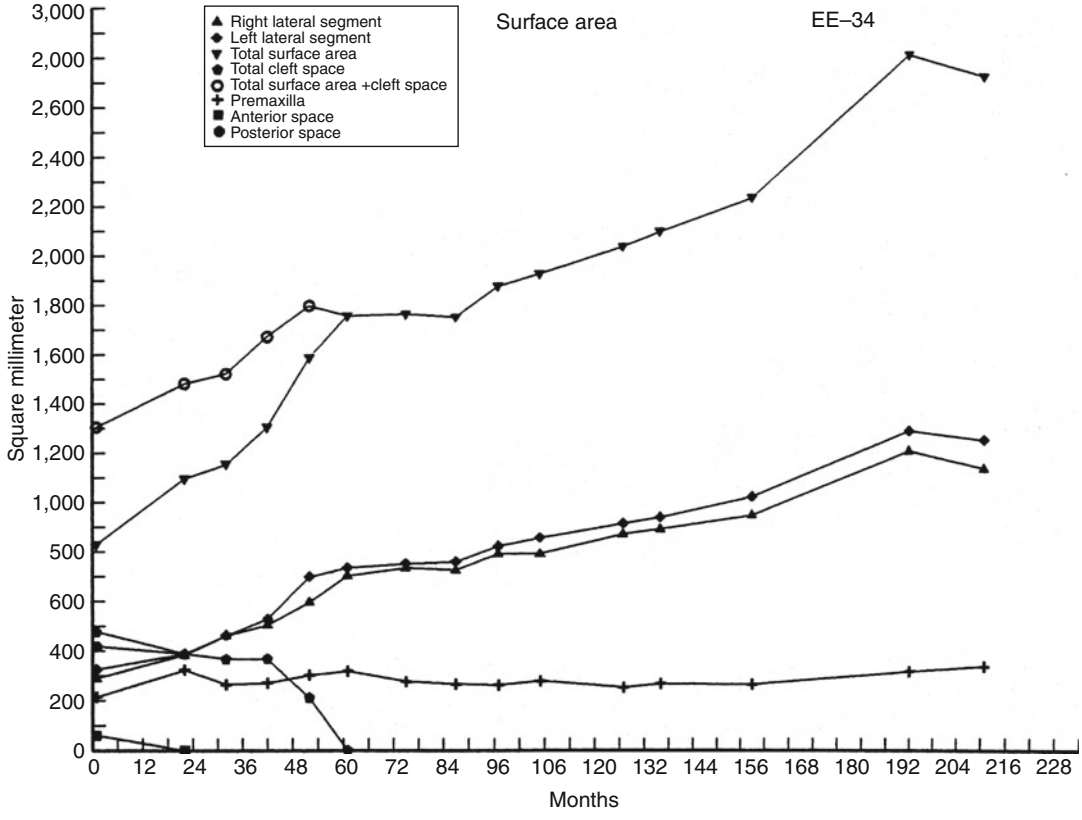

Fig. 7.57 Case EE-34. Very gradual growth acceleration curve, while the posterior cleft space closed very rapidly between 42 and 52 months of age. The premaxilla's growth was negligible

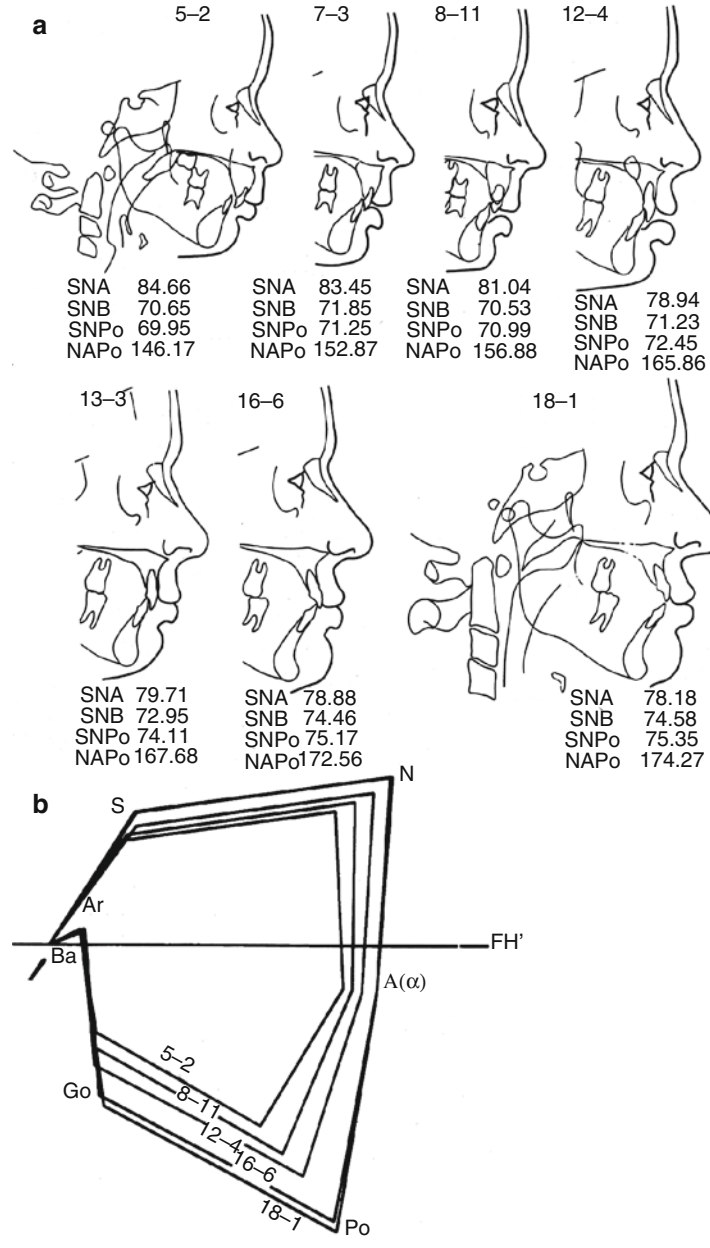

Fig. 7.58 (**a**, **b**) Case ES (EE-34). (**a**) Serial cephalometric tracings show ideal facial measurements evolving. A superior-based pharyngeal flap is outlined in 18–1. (**b**) Serial polygons superimposed according to the basin horizontal method (Coben). This series demonstrates an ideal facial growth pattern and reflects the use of physiological maxillary surgery: (1) The anterior cranial base shows good growth increments. (2) Midfacial growth increments are continuous and only slightly less than those seen at N (Nasion). (3) Mandibular growth shows more forward than downward changes which are conducive to the flattening of the facial profile. Comment: Taken together, all of these growth changes are conducive to the flattening of the facial profile. Such extensive midfacial growth is usually not seen in BCLP cases with such a large cleft space relative to the palate's surface area that have had the palatal cleft closed earlier than 43 months of age. We speculate that this finding strongly suggests that: (1) delaying palatal surgery to 3 years of age may be more conducive to good midfacial growth in some cases than early (before 1 year) palatal closure. However, we believe that it is not necessary to postpone palatal closure to after 5 years of age in all cases. (2) Hypotonic lip pressure has not exerted sufficient pressure through the premaxilla to the premaxillary-vomerine suture to reduce its growth before 16–6. However, between 16–6 and 18–1, midfacial growth has ceased

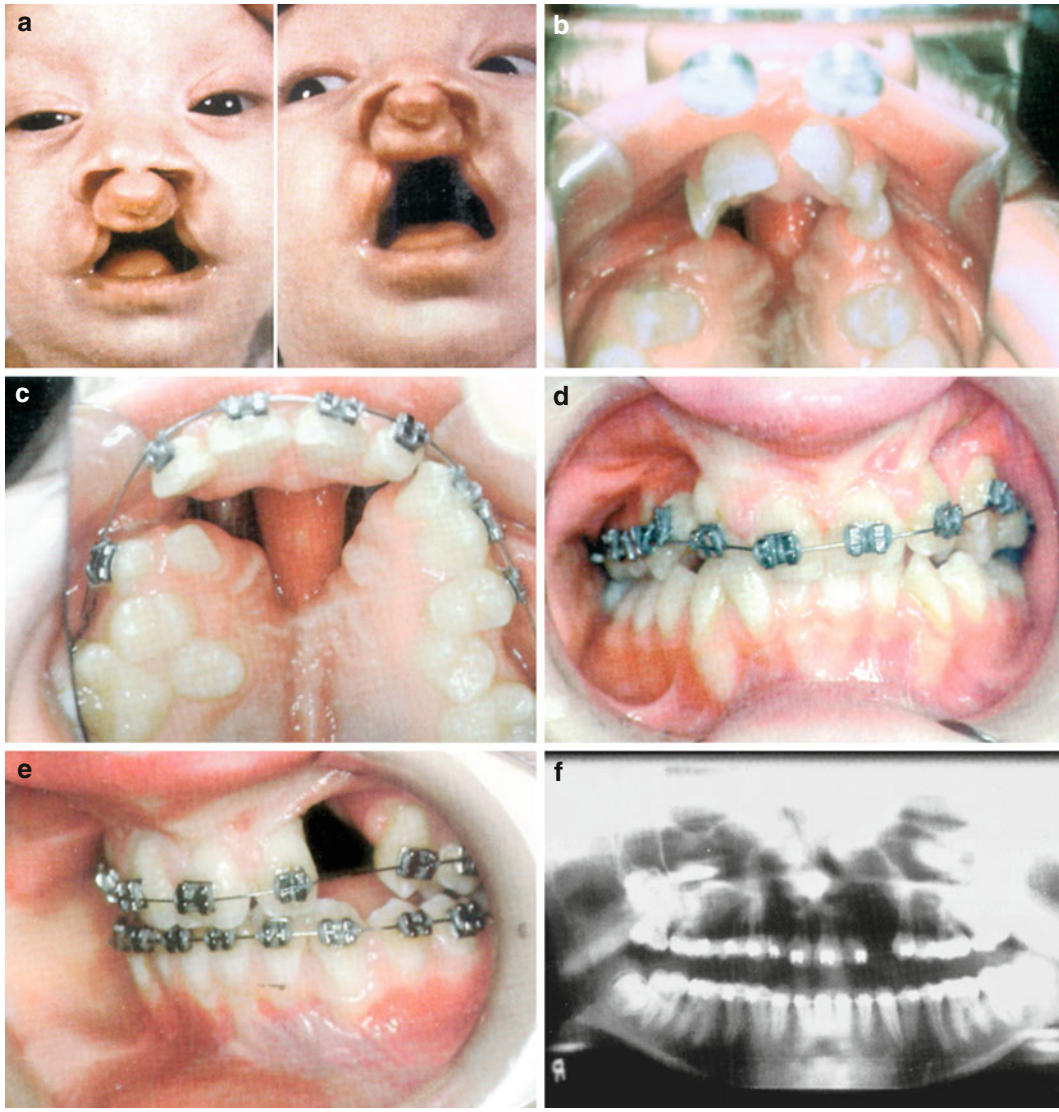

Fig. 7.59 (**a**–**n**) Anterior positioning of the lateral palatal segments in an adolescent with CBCLP to close a large anterior cleft space Semb (1991). A 16-year-old with repaired BCLP underwent combined orthognathic-orthodontic procedure to align the teeth, close the large cleft space while maintaining the existing premaxillary overjet-overbite relationship. (**a**) Initial cleft lip and palate deformity at 8 weeks of age. (**b**) Palatal view at 10. (**c**, **d**) Preoperative palatal and frontal view at 15 years showing large anterior cleft space and dental crowding of the lateral palatal segments. The right second bicuspid is to be extracted. (**e**) Preoperative occlusal view showing large cleft space with posterior positioned lateral palatal segment. (**f**) Preoperative panorex at 17 years of age

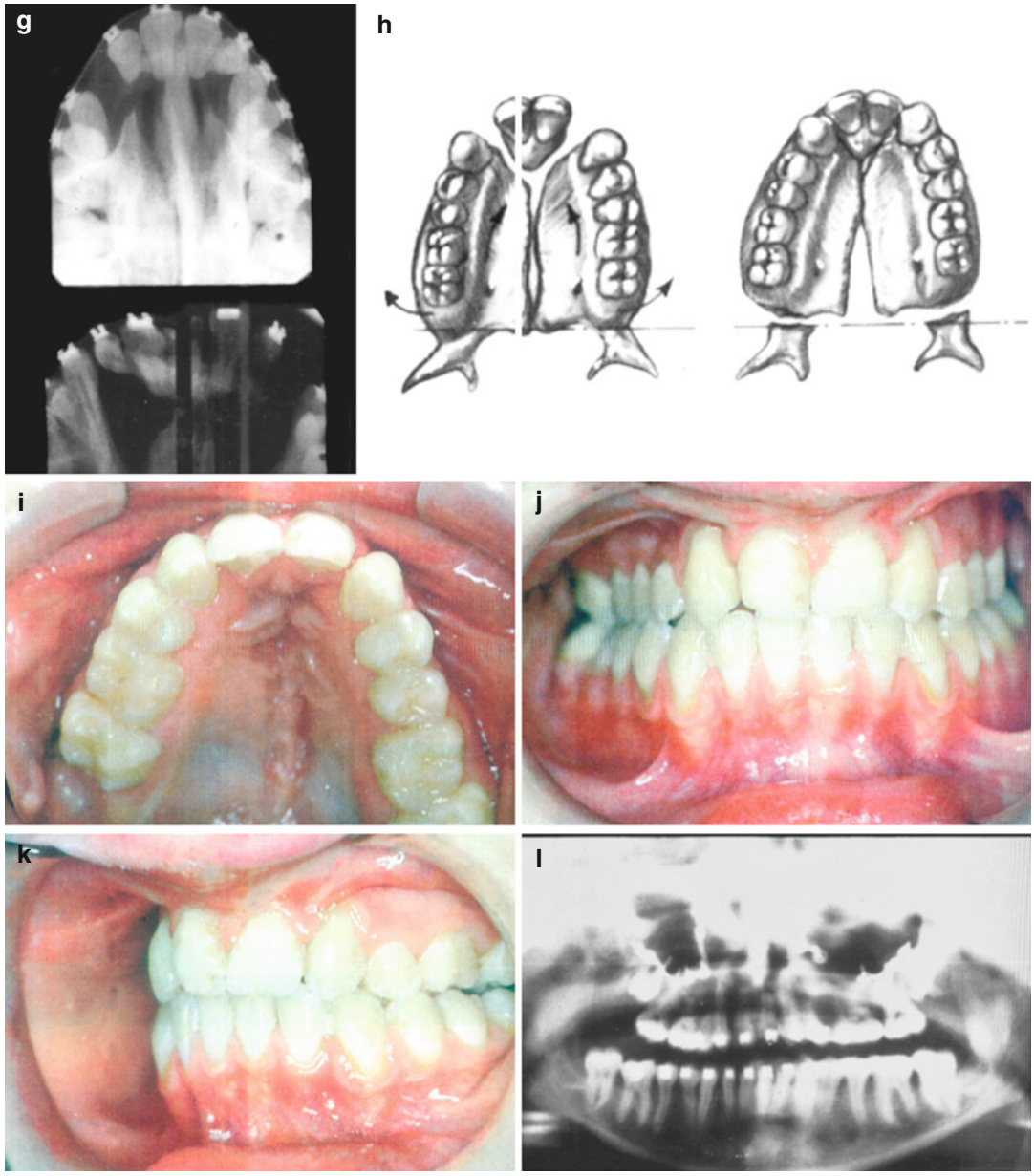

Fig. 7.59 (*continued*) (**g**) Occlusal radiograph. (**h**) Line drawing of the proposed palatal surgery showing the posterior widening and anterior movement of the lateral palatal segments. (**i–k**) Postoperative intraoral photographs. (**l**) Postoperative panorex

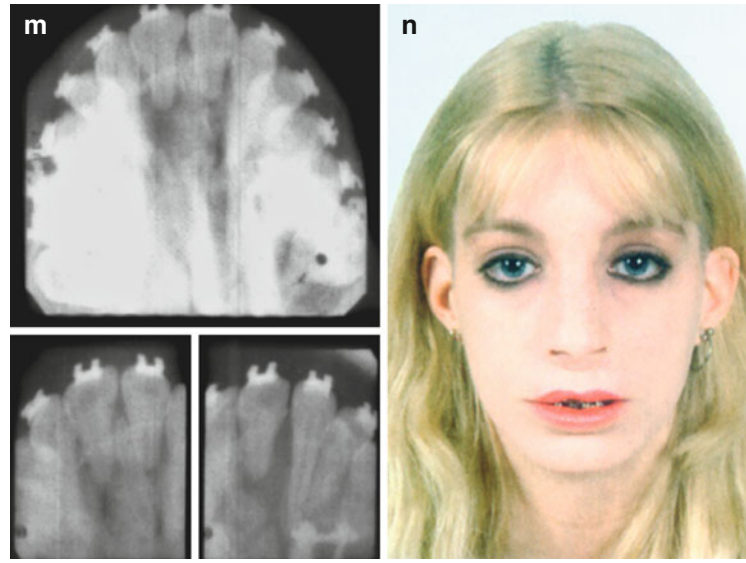

Fig. 7.59 (*continued*) (**m**) Occlusal radiographs. (**n**) Postoperative frontal facial view. Comment: There was limited alveolar bone support to the lateral incisor areas. The premaxilla was mobile. Marked velopharyngeal incompetence with regurgitation of fluid while drinking and intake of air while speaking required a palatal obturator. The surgical plan was to extract the questionable lateral incisors and reposition the maxillary lateral segments to close the lateral incisor and large anterior cleft spaces

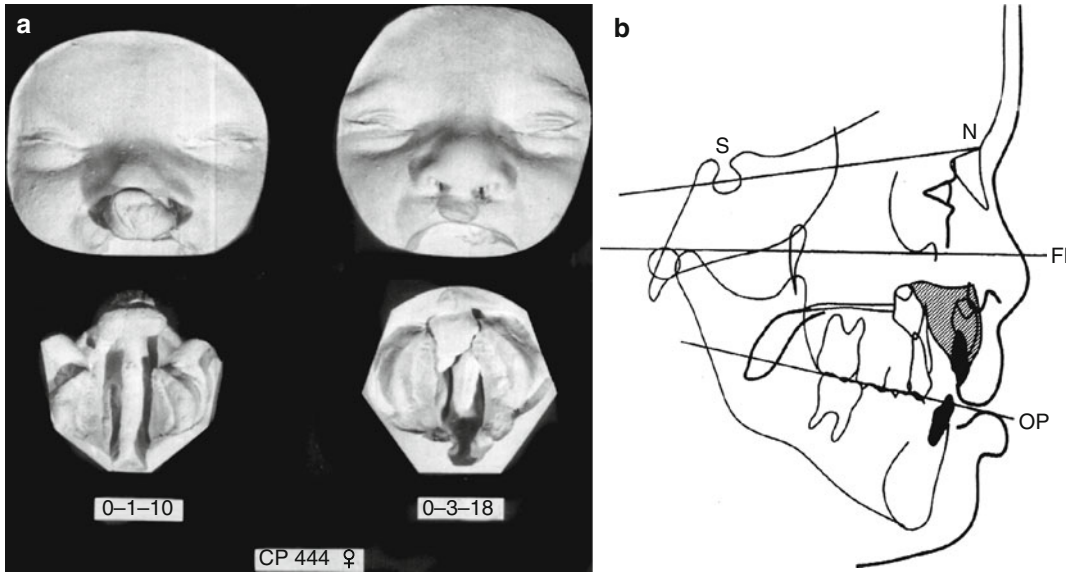

Fig. 7.60 (**a**, **b**) Case CP (444) demonstrates the effects of early premaxillary surgical setback. The purpose of this surgical procedure is to establish ideal arch form prior to lip surgery. The procedure lost favor when surgeons found that it led to severe midfacial growth retardation and an anterior open bite. (**a**) Before and after premaxillary setback. The premaxilla is positioned within the arch. (**b**) 4 years later, the premaxilla, having been detached from the vomer, fails to descend with the palate with growth, creating on anterior open bite. Orthodontics in the permanent dentition will super erupt the maxillary incisors with only slight change in maxillary position (Courtesy of S. Pruzansky)

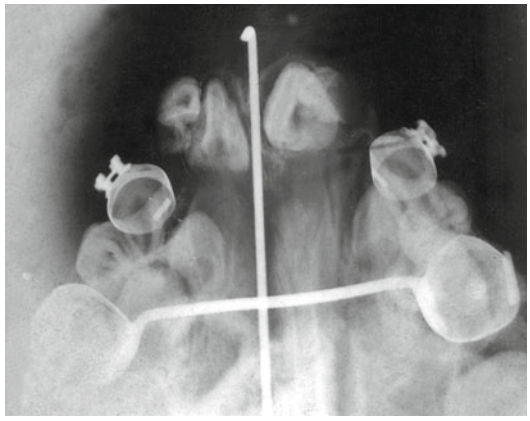

Fig. 7.61 A Kirschner wire that penetrated dental sacs of the central incisors, causing tooth malformation. Due to the spatial relationship of the premaxilla to the vomer, the Kirschner wire frequently fails to enter the vomer as seen here

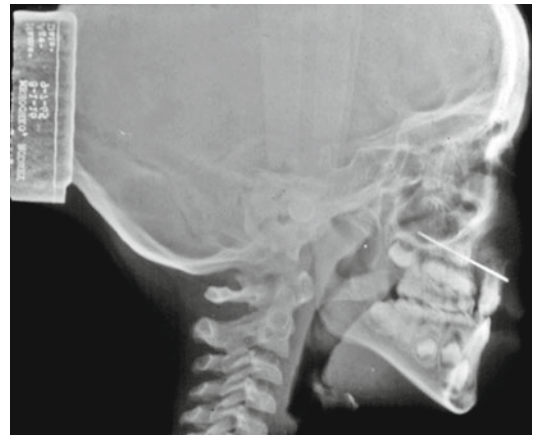

Fig. 7.62 Lateral cephalograph showing a malpositioned Kirschner wire. After the premaxilla is surgically retropositioned, surgeons used to attempt to stabilize it by placing wires of various sizes through the premaxilla into the vomer. The procedure has not been successful and should be abandoned because it can devitalize the teeth in the premaxilla and the wire can migrate to various areas within the skull

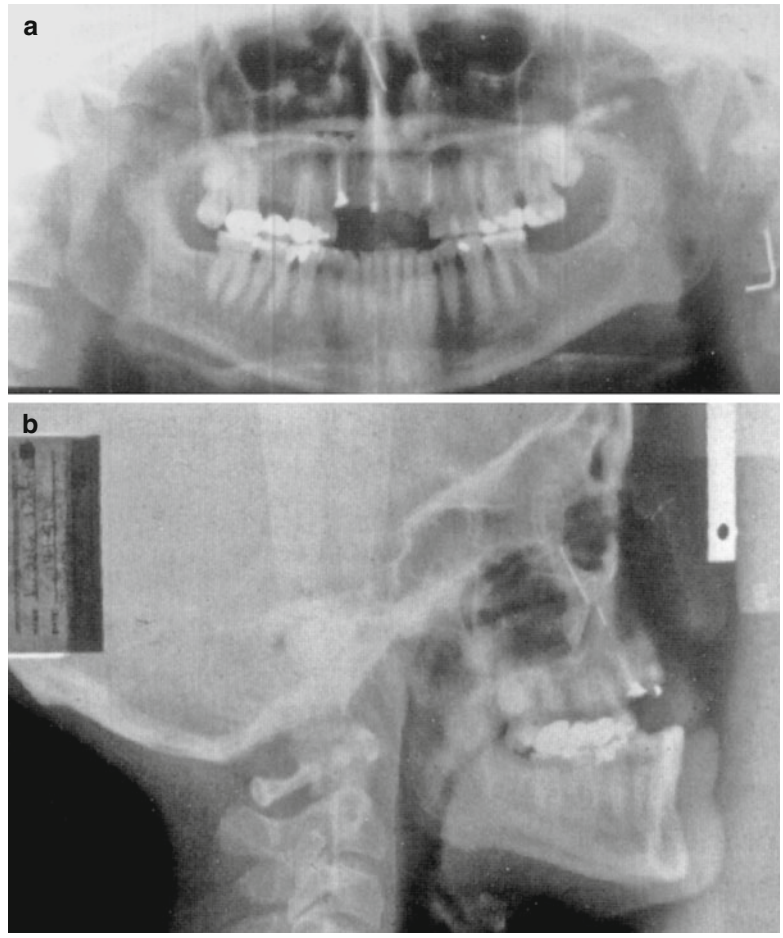

Fig. 7.63 (**a**, **b**) Two views of a Kirschner wire 35 years after insertion. (**a**) Panorex and (**b**) Lateral cephalometric films show the displaced Kirschner wire located high in the vomer

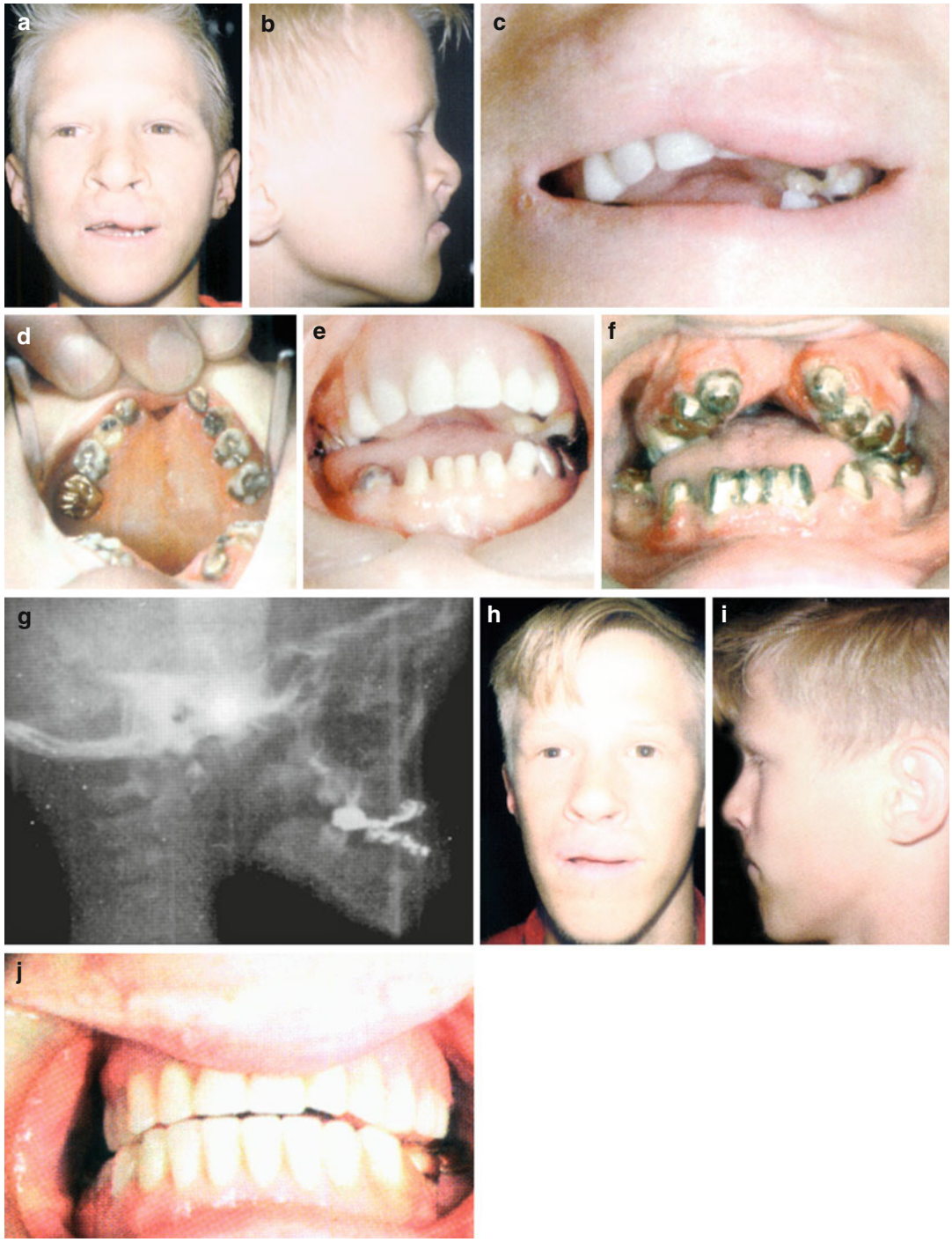

Fig. 7.64 (**a**–**j**) Case LN demonstrates the effects of premaxillary incision. This case was referred to us 25 years ago after the premaxilla had been excised. This is the only case in our files where the premaxilla was removed at birth. The remaining teeth were badly decayed. The treatment plan required overlay dentures to both arches. As a precaution, all the teeth were crowned prior to the construction of the dentures. The upper lip was revised. (**a**–**c**) Facial photographs, at the start of the treatment. (**d**) Maxillary arch with decayed teeth. (**e**) Denture over decayed teeth. (**f**) Upper and lower teeth were crowned. (**g**) Lateral head plate after dental reconstruction. (**h**) Upper and lower over dentures. (**i**, **j**) Facial photographs after lip revision

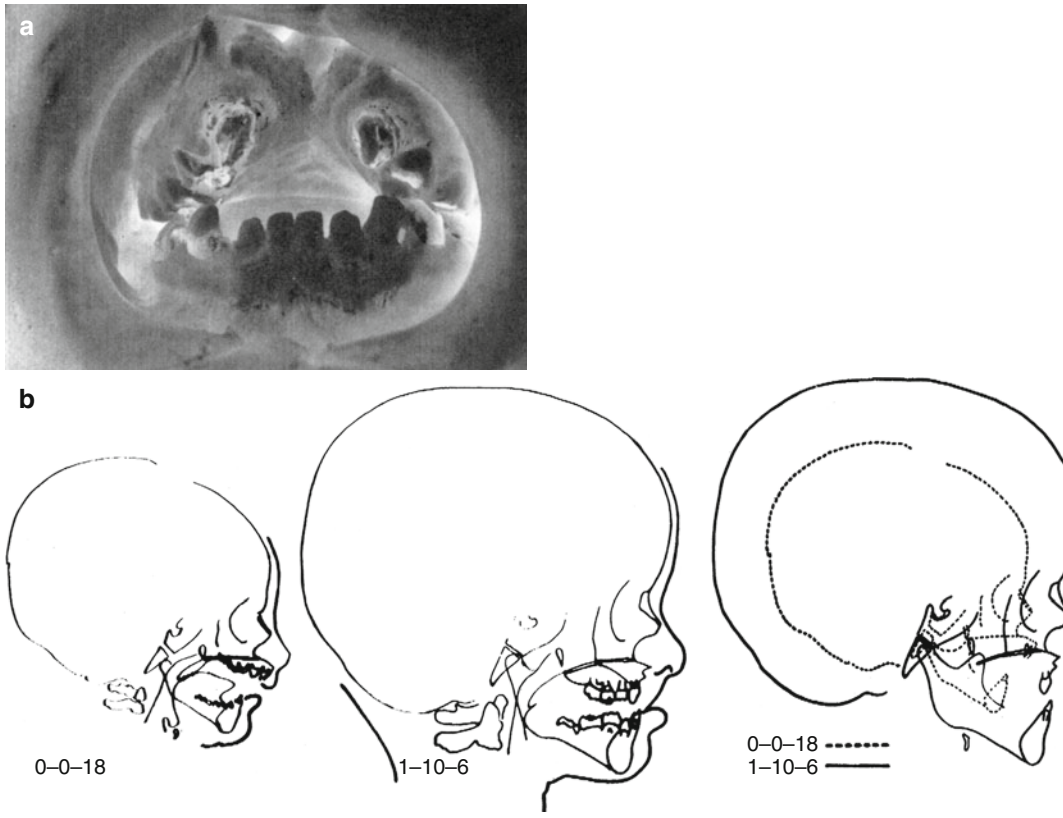

Fig. 7.65 (**a**) Premaxillary excision. Removal of the premaxilla leads to a large oronasal opening with a retruded midface. Even at 1 year, 10 months of age, the upper lip is severely retruded (Courtesy of S Pruzansky). (**b**) Premaxillary excision results in direct communication between the oral and nasal chambers

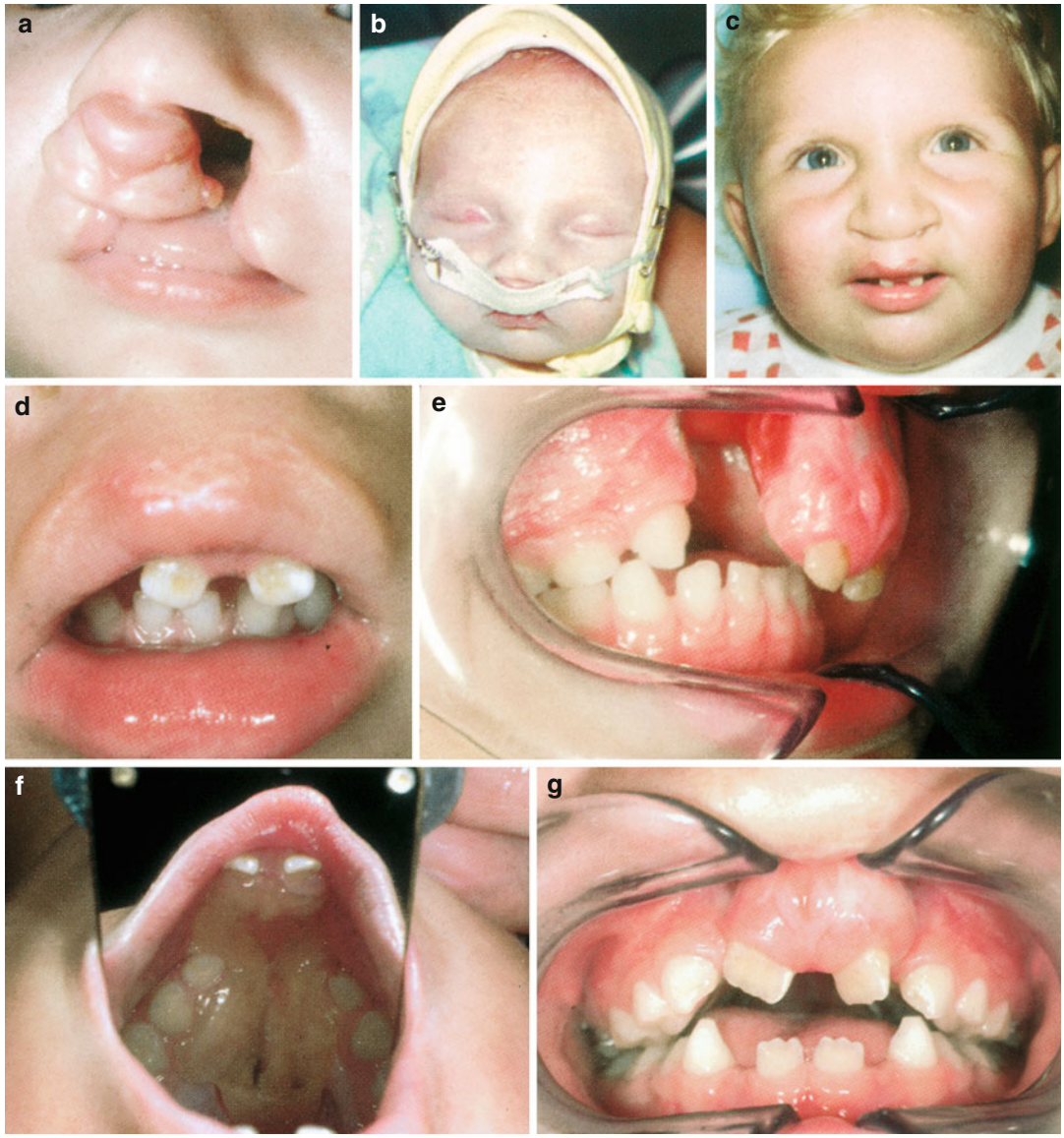

Fig. 7.66 (**a**–**q**) Case CH (II-64). This case demonstrates the need to advance the premaxilla after premaxillary surgical setback. A chin augmentation was necessary to neutralize the effect of excessive upper facial growth. (**a**–**i**) An external elastic off a head bonnet was used to ventroflex the premaxilla prior to lip surgery. As a result of the continuous premaxillary overjet, at 4–9, it was surgically set back. The palatal cleft was closed at 5–6 using a modified von Langenbeck procedure. A secondary alveolar bone graft was placed at 10–5. The patient's family moved to a different state where the orthodontist unsuccessfully attempted to close the lateral incisor space by retracting the central incisors

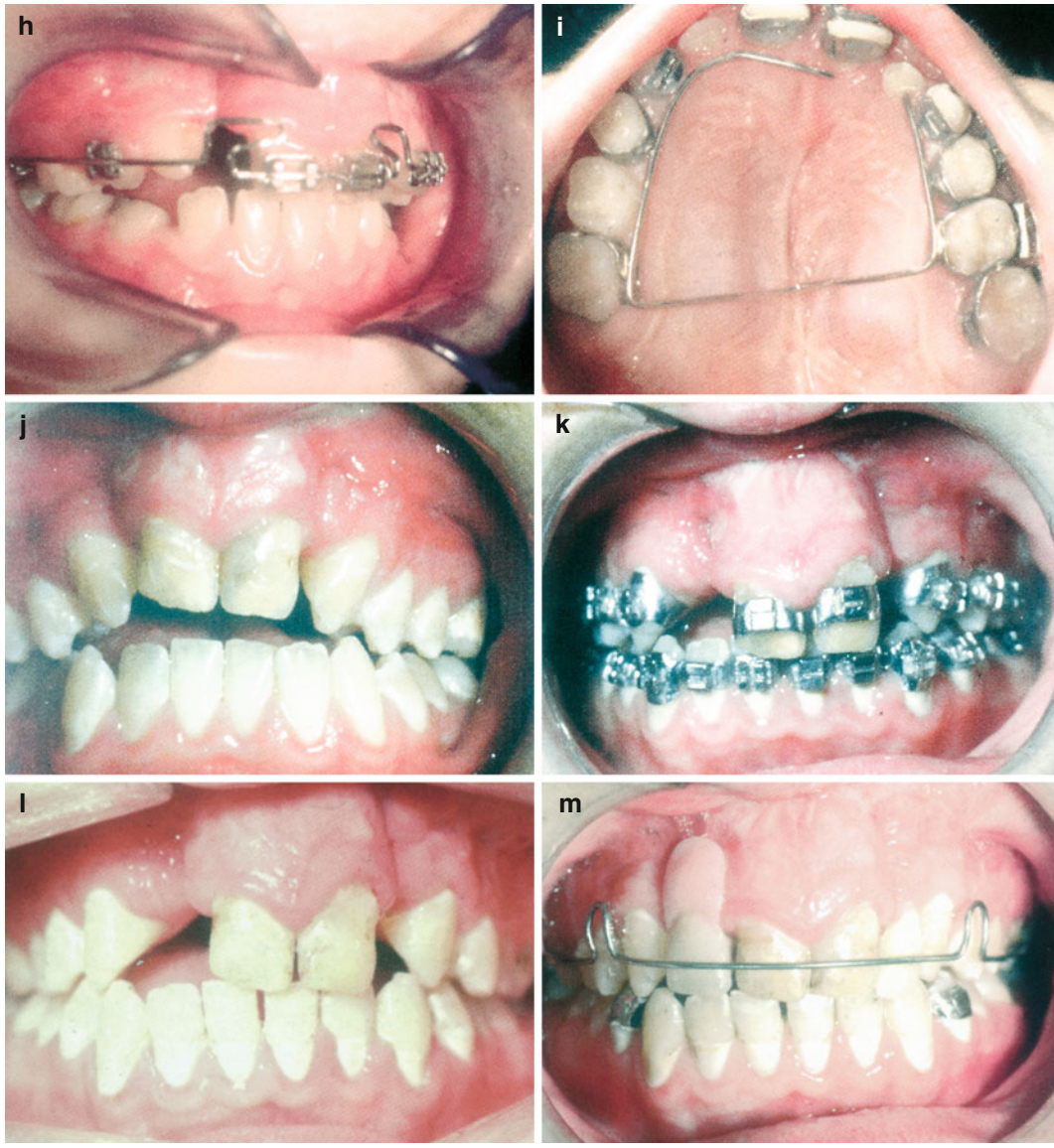

Fig. 7.66 (*continued*) (**j**) An anterior open bite was created. The patient returned to Miami. Treatment plan was changed to: (1) advance the premaxilla and open the lateral incisor space and (2) along with chin augmentation using autogenous bone from the inferior border of the symphysis. (**k**–**m**) After mandibular sagittal split surgery and orthodontic premaxillary advancement to recover lateral incisor spaces and advance the midface. A Hawley retainer with two false teeth were used to maintain the arch form

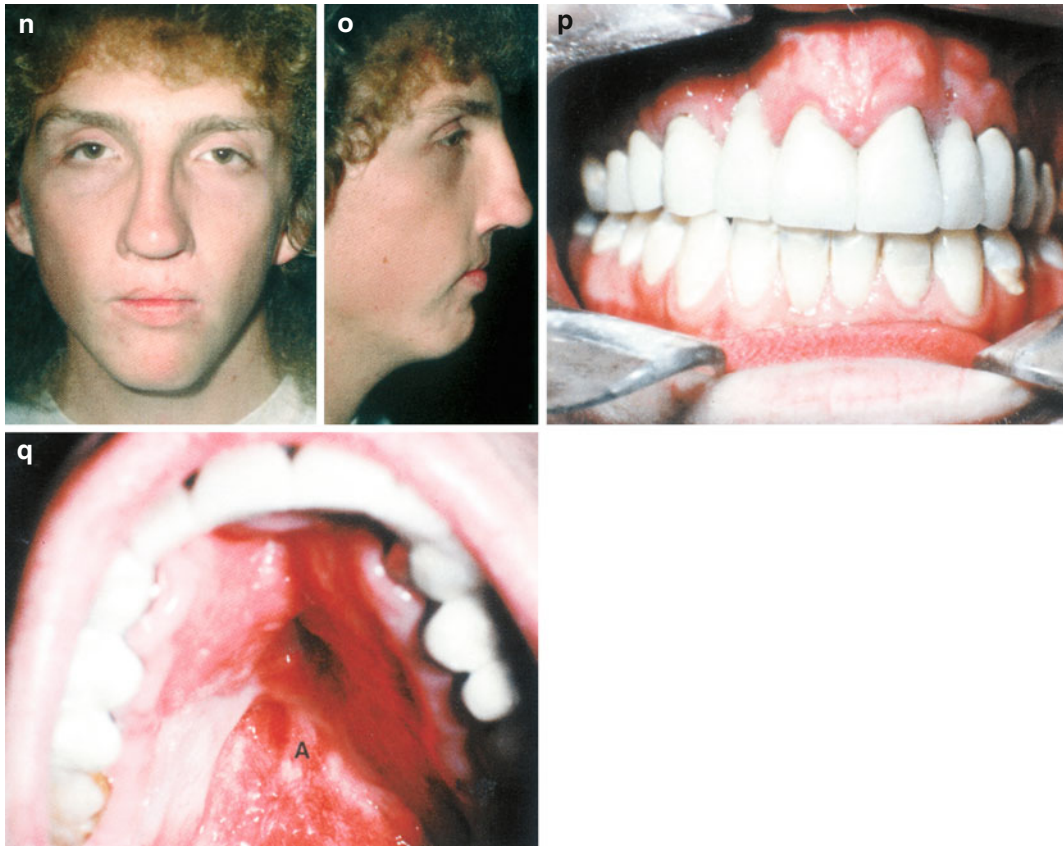

Fig. 7.66 (*continued*) (**n**, **o**) Facial photographs showing a pleasing facial profile. (**p**) A fixed bridge replaced the removable retainer. (**q**) Shows a pharyngeal flap at A

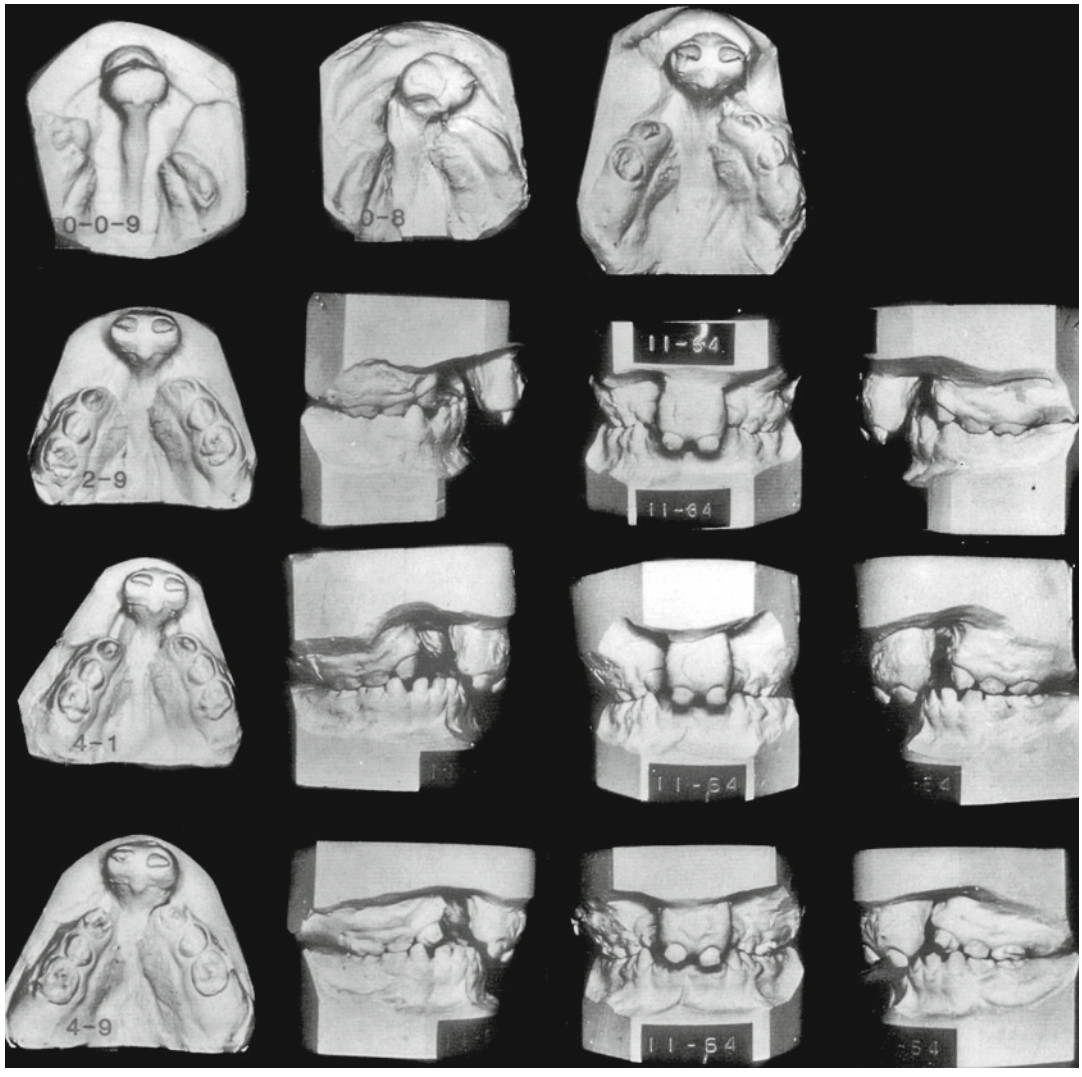

Fig. 7.67 Case CH (II-64). Serial dental casts. 0–0–9 9 days of age. 0–8 Premaxillary ventroflexion after uniting the lip. The palatal segments have moved medially making contact with the vomer. The premaxilla has uprighted, but the anterior cleft space is slow in reducing. 2–9 Severe overbite and overjet. Mesioangular rotation of both palatal segments placed the deciduous cuspids in crossbite. 4–1 and 4–9 The anterior overbite and overjet have reduced spontaneously

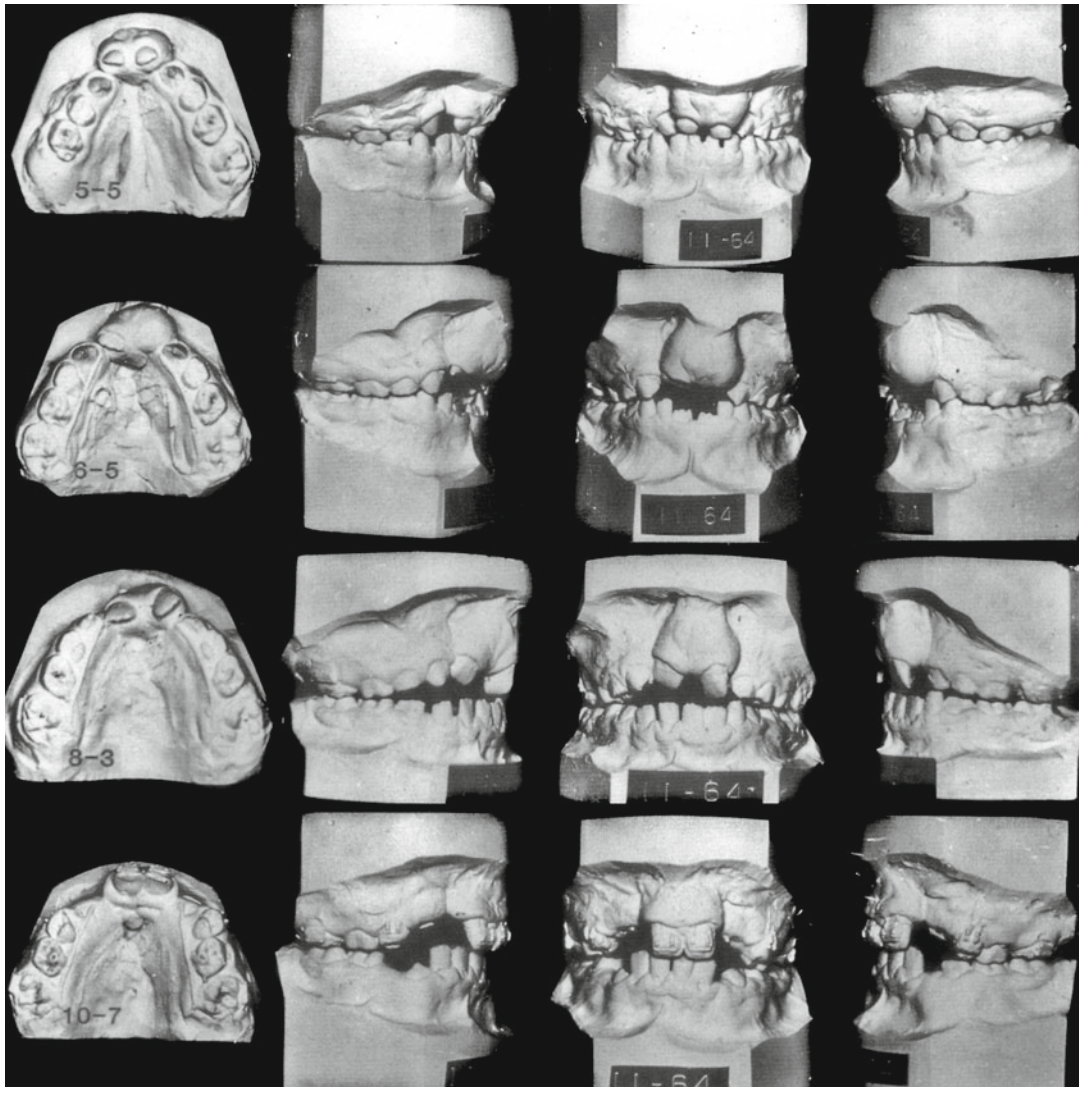

Fig. 7.67 (*continued*) 4–11 The premaxilla was surgically set back. 5–5 The anterior open bite was eliminated. 6–5 The palate was expanded to correct the deciduous cuspid crossbite and to allow the premaxilla to fit within the arch. 8–3, 10–7, and 11–5 Palatal retainer is used to hold the corrected arch form

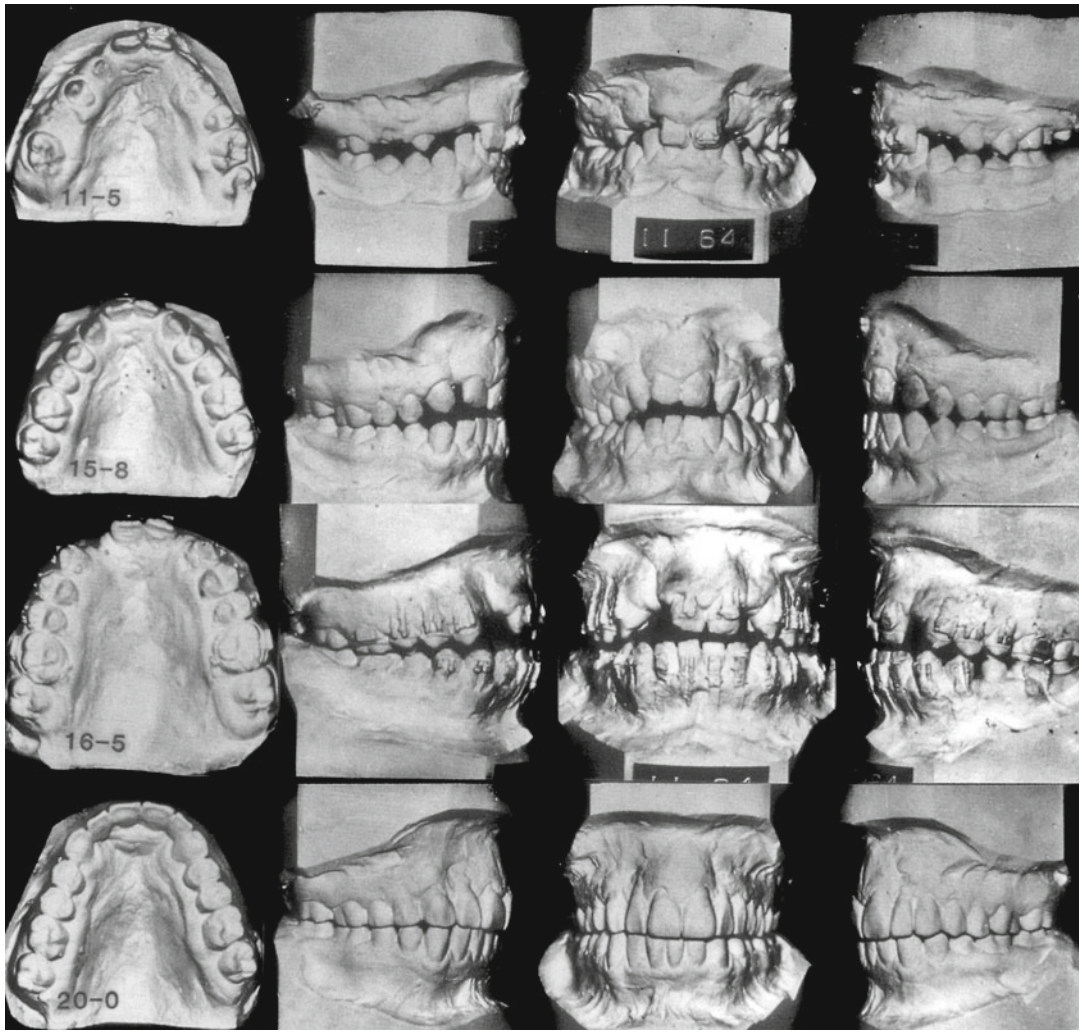

Fig. 7.67 (*continued*) 15–8 An attempt to close the lateral incisor space orthodontically was unsuccessful. 16–5 Premaxilla advanced and lateral incisor spaces opened. 20 Fixed bridge replaces missing teeth and maintains the arch width and form

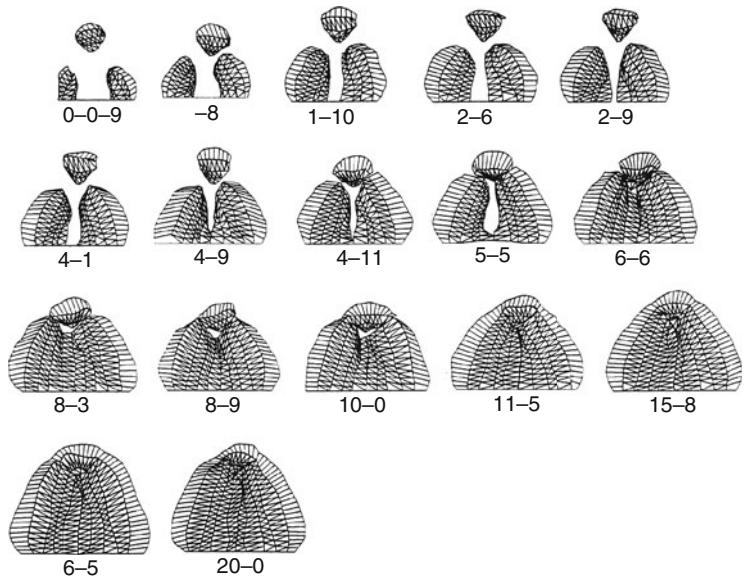

Fig. 7.68 Case CH (II-64). Computer-generated palatal images drawn to scale. This series shows the gradual decrease in cleft space size from birth to 5–5 years. A posterior cleft space remained until 5–6. Alveolar cleft closed at 10–5. Comment: Hopefully, the research project currently under way will determine the best age, based on parameters such as the ratio of the palatal surface area to cleft space size, at which to close the posterior palatal cleft. The various facial-palatal growth studies presented in this chapter show great variation in palatal as well as facial growth patterns. Differential diagnosis to select the "best" time and surgical procedure to close the cleft space should not be based on age alone, but should consider the nature of the cleft defect – its size and the availability of tissue

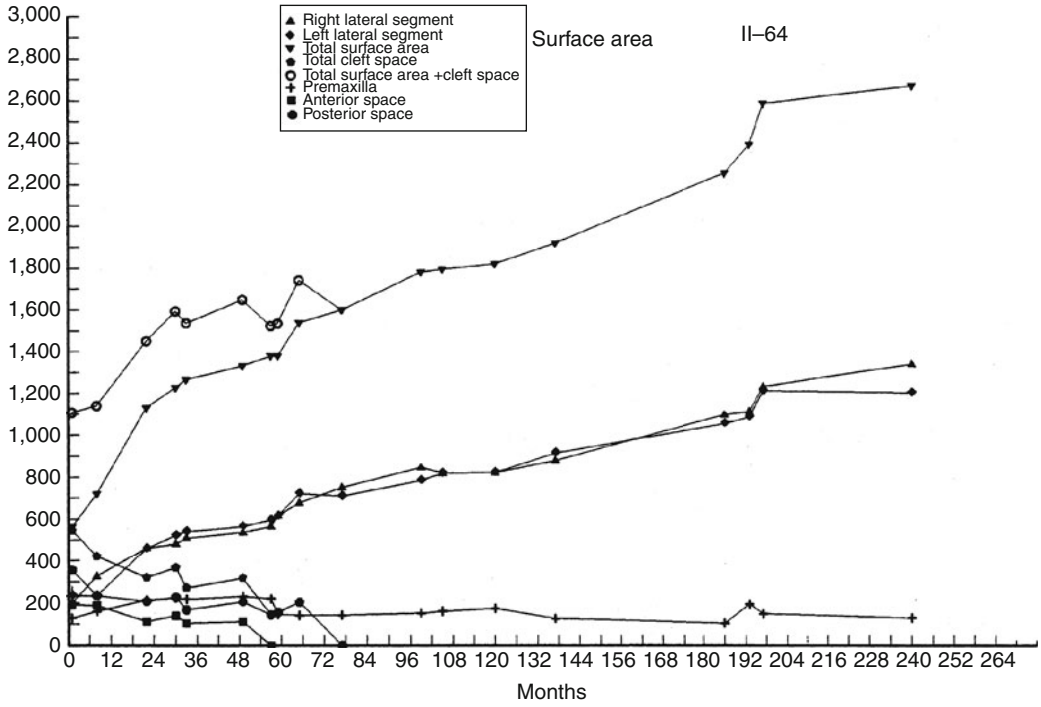

Fig. 7.69 Case CH (II-64)A rapid increase in palatal growth as the posterior cleft space reduces at almost the same rate. The reduction in anterior cleft space proceeds at a lesser rate. Palatal growth rate after surgery is the same as it was before surgery

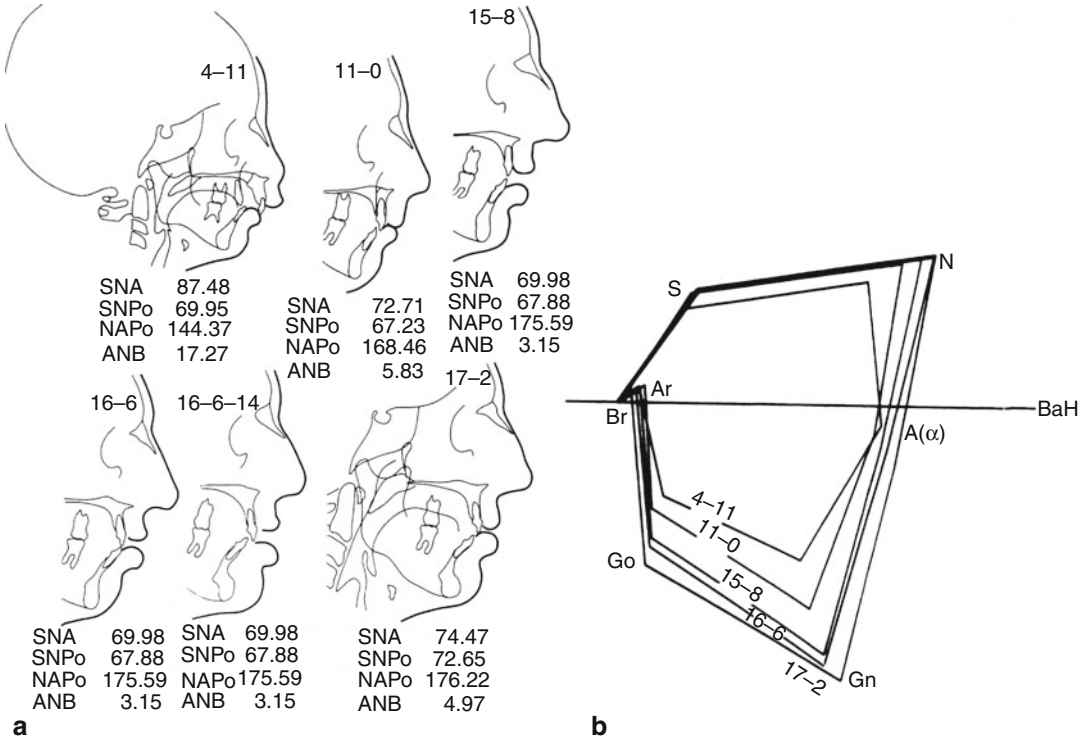

Fig. 7.70 (**a**, **b**) Case CH (IT-64). (**a**) Serial cephalometric tracings showing facial changes with a severely protruding maxilla at 4–11 and (**b**) superimposed polygons using the basion horizontal method (Coben). An excessive amount of growth occurred at the anterior cranial base (at N) and in the mandible mainly in the vertical dimension. With some minor midfacial growth, the skeletal and soft tissue profile became more retrognathic and unaesthetic. Because of the prominent upper face, it was decided to improve facial aesthetics by advancing the premaxilla and to open the lateral incisor space. Notice the resulting larger incremental midfacial change resulting from the movement of A point between 16–6 and 17–2. Chin augmentation increased the mandibular prominence from an SNPo angle of 67.88° to 72.65°. With the lack of continued growth of the anterior cranial base between 16–6 and 17–2, the angle at facial convexity changed slightly from 175.59° to 176.22°. Comments: The clinician should not evaluate the alignment of teeth only within the arch and occlusion without considering the effects of anterior tooth position on the developing facial profile. Retracting the maxillary central incisors along with A point to close the lateral incisor space made the midface appear very deficient and aesthetically unpleasant. Because of the large increment of anterior cranial base growth, it was necessary to advance both the midface and augment the chin. Therefore, when treatment planning, the protruding premaxilla at birth needs to be considered as part of the developing face, and treatment options need to be left open until the facial growth pattern is identified and can be adjusted to its best advantage. It is important to emphasize that a mechanically or surgically retropositioned premaxilla does not advance in the midface with growth as a nonretracted conservatively treated premaxilla will do

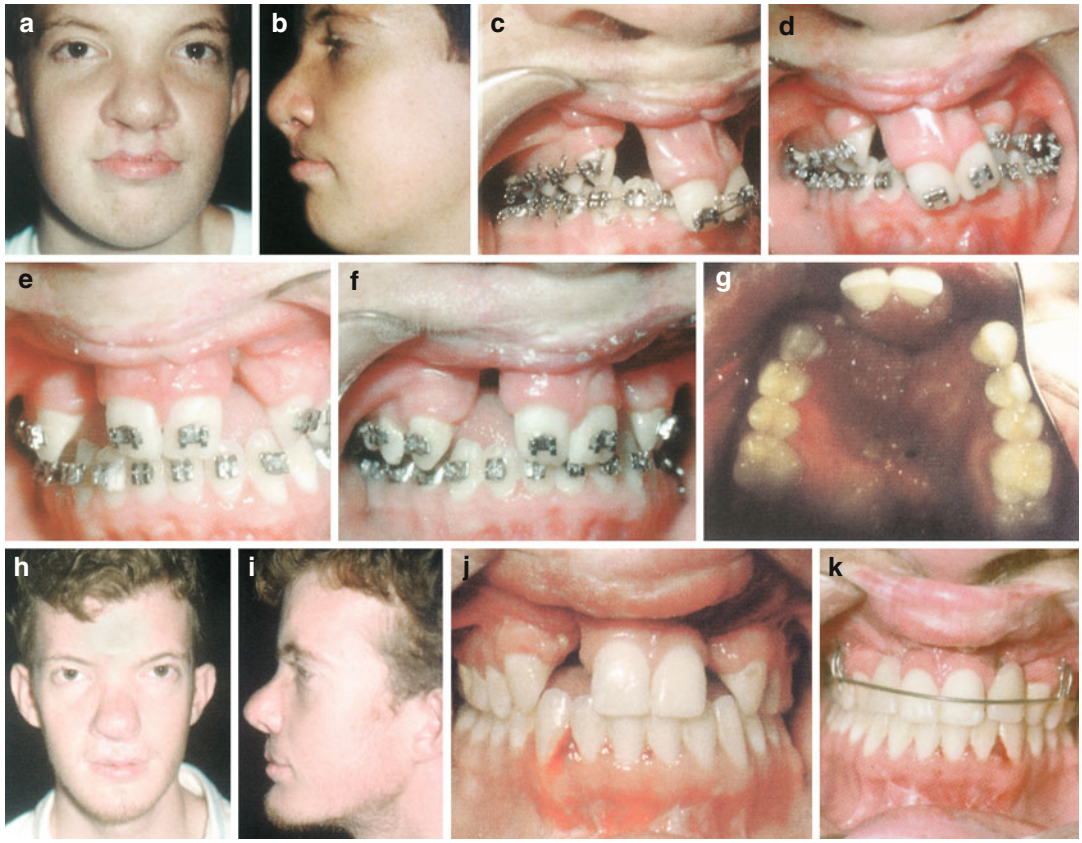

Fig. 7.71 (**a**–**k**) Case AO (BM-82) shows surgical repositioning of the premaxilla in an adult. This patient had been under orthodontic-surgical care elsewhere. The orthodontist was reluctant to orthodontically correct the severe premaxillary overbite and left it to the surgeon. Because of the patient's age, it was decided that surgical premaxillary alignment was the treatment of choice. (**a**–**k**) Before and after surgery. The maxillary central incisors are stabilized with a Hawley retainer with two false teeth in the lateral incisor position

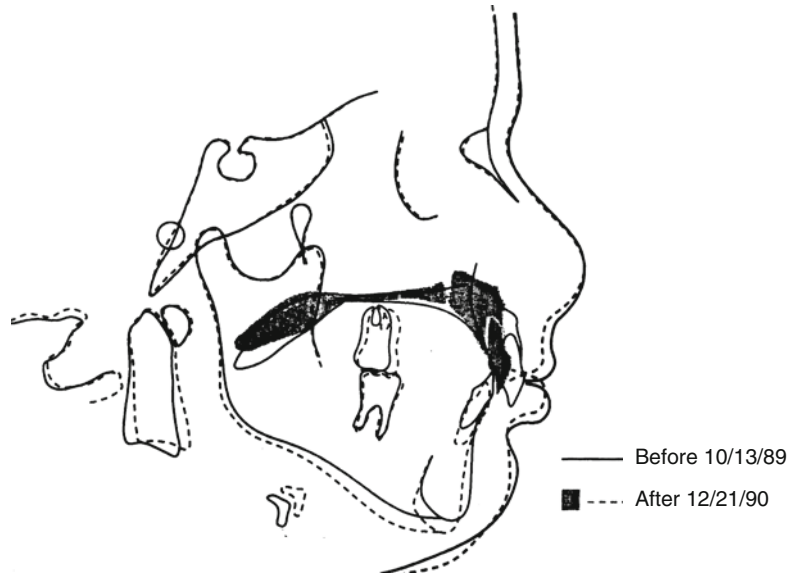

Fig. 7.72 Case AO (BM-82). Lateral cephalometric tracings showing correction of the premaxillary overjet and overbite in an adult. Comment: When a severe premaxillary overbite exists in the mixed and early permanent dentition, in most cases, it can be successfully treated with orthodontics. The surgical cut is made between the premaxilla and the vomer. The need to protect the integrity of the PVS is not crucial at this age. Rigid arch wire stabilizes the corrected premaxillary position during healing

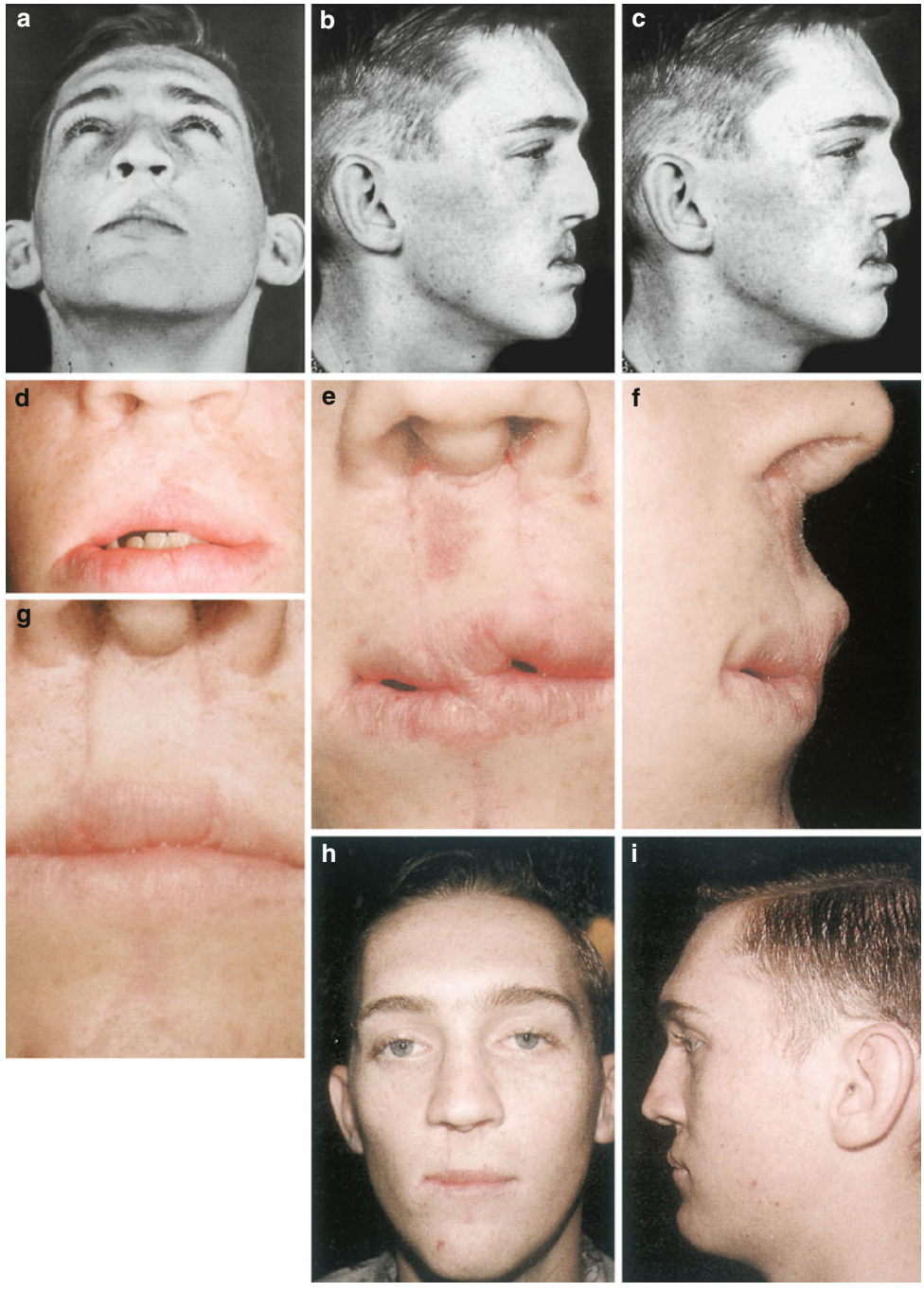

Fig. 7.73 (**a**–**i**) Case TK. Lip-Switch (Abbe Flap). The combination of a short columella and a short or long and tight upper lip requires secondary cleft lip surgery. (Millard 1978b) strongly believes that, had the primary surgery been planned and executed properly, a lip switch flap would most likely never be required. Unfortunately, there are occasions when the destruction of the lip landmarks and midfacial growth retardation require that tissue be brought in from outside and used to remove scars, bring in muscle continuity, create a philtrum and even a bow, correct free border defects, and relieve tension. Millard suggests that the flap form the total central vertical length of the upper lip. It will then resemble the natural philtrum and can appear normal in spite of its scars. (**a**–**c**) Repaired bilateral cleft lip and palate has resulted in a malformed, recessive midface with a scarred upper lip. (**d**–**f**) The flap was taken from the middle of the lower lip and inserted in the total central portion of the upper lip. (**g**–**i**) After lips were separated. The upper lip is symmetrical and shows a more normal looking vermillion. A dental prosthesis bumpers the upper lip

Table 7.2 Surface area of CBCLP. Case DK (AI-31)

	Skeletal area				Cleft space			Total
Age	Premax	RLS	LLS	Tot	Ant	Post	Tot	SA + CS
0–1	55.6	314.1	254.2	623.9	102.9	138.8	341.7	965.6
0–3	79.2	366.6	317.5	763.3	150.5	191.8	342.4	1,105.7
0–4–15	86.1	362.4	300.8	749.3	56.1	259.3	315.4	1,064.7
0–5–10	99.5	356.2	322.5	778.2	70.3	175.8	245.5	1,023.7
0–7	111.7	361.9	321.3	794.9	45.2	168.0	213.2	1,008.1
0–8	106.8	376.0	351.0	833.8	85.6	149.1	234.7	1,068.5
1–0	100.3	428.1	327.2	855.6	55.6	198.8	249.4	1,105.0
1–2	132.8	432.6	389.3	954.7	55.7	189.8	245.5	1,200.2
1–5	127.6	507.3	418.1	1,053.0	51.2	180.8	232.0	1,285.0
1–10	132.6	493.4	421.1	1,047.1	39.4	194.9	234.3	1,281.4
2–1	117.8	492.6	473.0	1,083.4	32.3	168.8	201.1	1,284.5
2–5	116.7	503.0	456.0	1,075.7	16.1	177.3	193.4	1,269.1
2–10	133.6	556.3	449.3	1,139.2	17.8	191.2	209.0	1,348.2
3–5	102.2	451.8	434.3	988.3	11.2	97.7	108.9	1,097.2
3–11	110.7	506.1	427.6	1,044.4		94.4	94.4	1,138.8
4–4	112.1	501.9	438.5	1,052.5		95.4	95.4	1,147.9
5–2	115.3	547.0	481.3	1,143.6		97.2	97.2	1,240.8
5–11	112.2	541.5	514.0	1,167.7		82.5	82.5	1,250.2
6–5	106.9	646.8	547.5	1,301.2		85.7	85.7	1,386.9
6–8	102.2	635.8	552.1	1,290.1		97.8	97.8	1,387.9
7–1	102.1	638.8	591.7	1,332.6		67.8	67.8	1,400.4
7–11	107. 1	667.2	592.4	1,366.7		53.3	53.3	1,420.0
8–2	104.8	650.9	624.4	1,380.1		40.6	40.6	1,420.7
8–5	72.5	647.0	597.4	1,316.9		40.5	40.5	1,357.3
8–11	68.9	704.2	638.8	1,411.9		41.6	41.6	1,453.5
9–10	81.7	770.5	708.4	1,560.6		42.8	42.8	1,603.4
10–5#	64.5	726.4	702.4	1,493.3		44.9	44.9	1,538.2
10–9	86.5	745.0	704.0	1,535.5		55.6	55.6	1,591.1
11–4	109.9	782.8	804.1	1,696.8		32.2	32.2	1,729.0
12–6	103.7	799.8	895.7	1,799.2				1,799.2
13–8	81.1	895.4	914.3	1,890.8				1,890.8
15–11	83.8	1,060.6	1,037.2	2,181.6				2,181.6
16–3	75.4	1,086.4	1,069.3	2,231.1				2,231.1

Note: *Premax* premaxilla, *RLS* right lateral segment, *LLS* left lateral segment, *Tot* total surface area, *Ant* anterior cleft space, *Post* posterior cleft space, *Tot* Ant+Post, *SA + SC* bony surface area + cleft space area, *#* changing teeth

Table 7.3 Surface area of CBCLP. Case PM (KK-22).The palatal surface area increased by 4,096 after 1 year, 4 months and by 7,696 at 2 years, 1 month. By 8 years, 2 months, the palatal surface area had increased two and a half times when the cleft space was closed

	Skeletal area				Cleft space			Total
Age	Premax	RLS	LLS	Tot	Ant	Post	Tot	SA + CS
0–0–12	145.5	335.5	282.7	763.7	127.4	417.0	544.4	1,308.1
0–3	150.2	397.4	377.6	925.2	65.4	331.5	396.9	1,322.1
1–4	154.0	469.5	464.3	1,087.8	36.8	265.5	302.3	1,390.1
1–10	211.8	502.6	506.2	1,220.6	70.4	216.3	286.7	1,507.3
2–1	217.6	589.0	549.5	1,356.1	79.8	195.3	275.1	1,631.2
2–10	220.7	603.9	551.3	1,375.9	95.8	193.2	289.0	1,664.9
3–10	271.6	660.4	616.5	1,548.5	122.6	206.3	328.9	1,877.4
5–8	273.3	673.0	675.9	1,622.2	123.6	201.2	324.8	1,947.0
6–7	273.6	811.0	820.5	1,905.1	115.0	206.5	321.5	2,226.6
7–4	277.3	813.0	839.5	1,929.8	106.7	185.2	291.9	2,221.7
8–2	306.5	844.6	890.8	2,041.9	101.1	155.4	256.5	2,298.4
12–3	346.8	1,087.1	1,116.1	2,550.0				2,550.0
14–0	348.7	1,161.8	1,226.4	2,736.9				2,736.9
14–5	351.1	1,198.8	1,237.0	2,786.9				2,786.9
17–4	353.5	1,241.0	1,246.3	2,840.8				2,840.8

Note: *Premax* premaxilla, *RLS* right lateral segment, *LLS* left lateral segment, *Tot* total surface area, *Ant* anterior cleft space, *Post* posterior cleft space, *Tot* Ant + Post, *SA + SC* bony surface area + cleft space area, *#* changing teeth

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8 Lip Pits: Orthodontic Treatment, Dentition, and Occlusion – Associated Skeletal Structures

Samuel Berkowitz

8.1 Lip Pits

8.1.1 Pits of the Lower Lip in Cleft Lip and/or Palate: Genetic Considerations

Pits of the lower lip such as fistulas of lower lip, paramedian sinuses of lower lip, humps of lower lip, or labial cysts are a very rare congenital malformation, first described by Demarquay in 1845 (Demarquay 1845).

This minimally deforming anomaly is remarkable chiefly for its association with facial clefts. The fact that clefts that occur with lip pits seem to run stronger in families than clefts without lip pits has attracted the attention of professionals dealing with cleft patients (Figs. 8.1, 8.2, 8.3, 8.4, and 8.5).

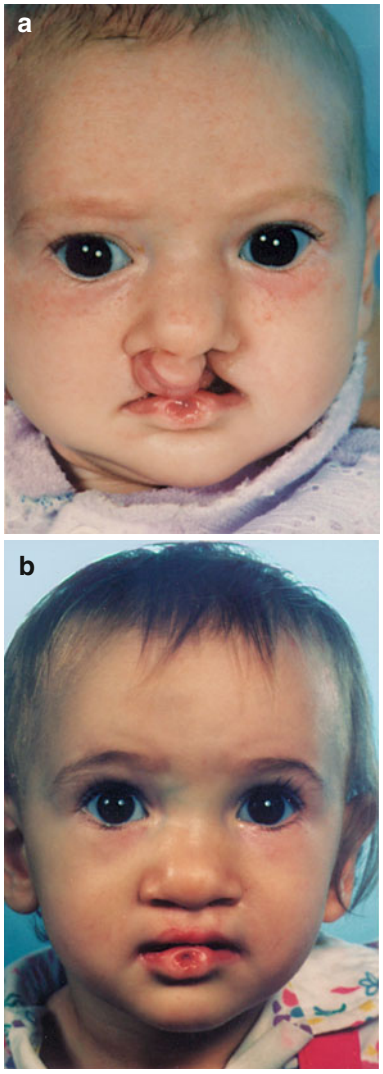

Fig. 8.1 Lower-lip pits in a child with a bilateral cleft lip and palate. (**a**) Before lip surgery. (**b**) After lip surgery at 6 months

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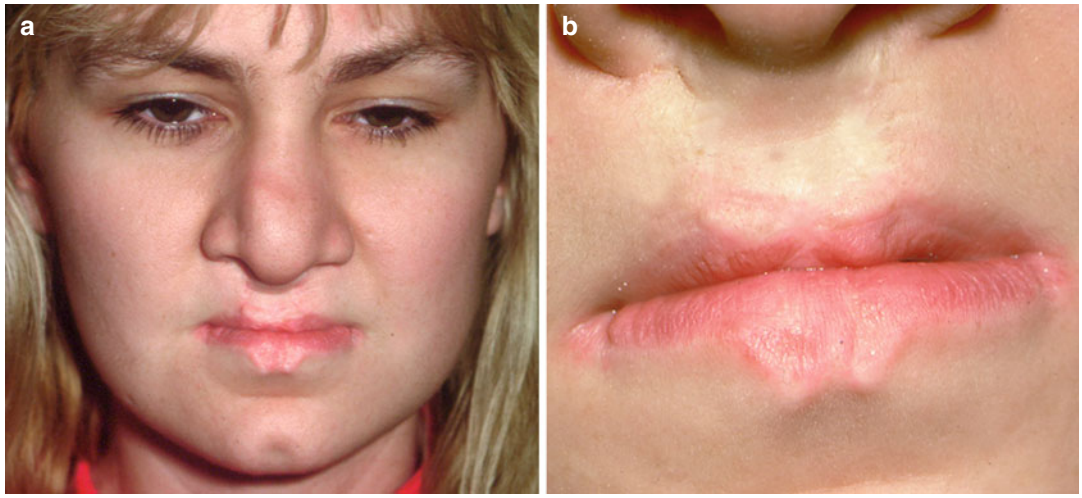


Fig. 8.2 (**a**) Pits in the upper and lower lips in a 20-year-old with a bilateral cleft lip and palate. (**b**) Close-up view

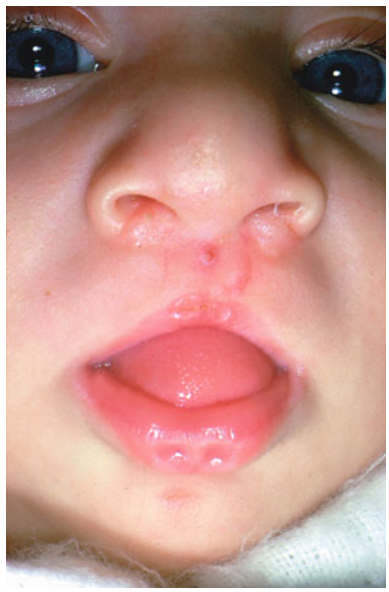


Fig. 8.3 Lip pits in the upper and lower lips in a bilateral cleft lip and palate

8.1.2 Frequency

No survey of lip pits has been carried out among the general population; hence, the frequency of this rare anomaly has been only roughly estimated from its incidence in hospital records.

Assuming that 70 (Gorlin and Pindborg 1964) to 80 % (Van der Woude 1954) of patients with pits of the lower lip have associated cleft lip and/or palate and that the frequency of clefts is 1:650 (one in every 650 births), it can be estimated that the frequency of lip pits among the general population is about 1:75,000–1:100,000 (one in every 75,000–100,000 births).

8.1.3 Morphology

Fistulas of the lower lip usually appear as two pits or humps on the vermilion portion of the lower lip, generally equidistant from the midline. Various kinds of asymmetry may be observed with regard to the midline, or one pit may be positioned more orally. Some pits are mere depressions; others are channels 10–15 mm deep with openings at the top of nipple-like elevations. Some secrete small amounts of viscous saliva but most are asymptomatic.

In exceptional cases, only one pit is present, which may be located either centrally or on one side or the other of the midline of the lower lip. Some cases of single lip pit have occurred in families with members with double lower-lip pits. It can be assumed that a single pit is not a distinct entity but rather an incomplete expression of the trait. On the other hand, the rarely described fistulas of the upper lip (Lannelongue 1879; Radcliff 1940) have not shown any inheritance pattern.

Commissural or angular lip pits – small, usually asymmetrical channels located at the lip

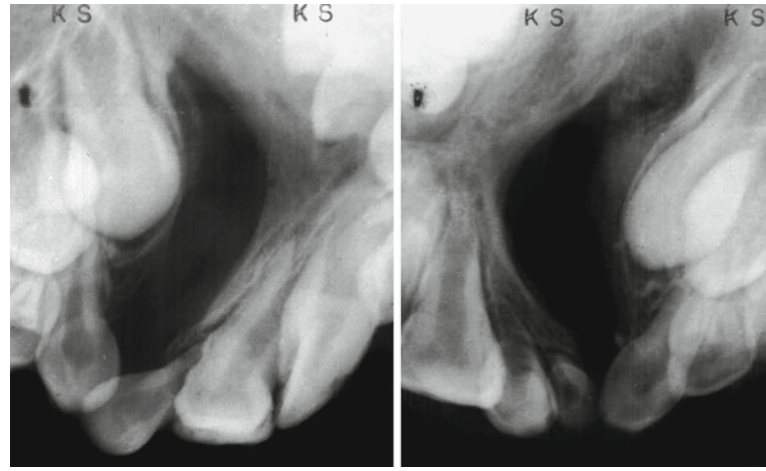


Fig. 8.4 Radiograph of a premaxilla showing two central and one lateral incisors (*left*) and twined lateral incisors and one malformed central incisor (*right*)

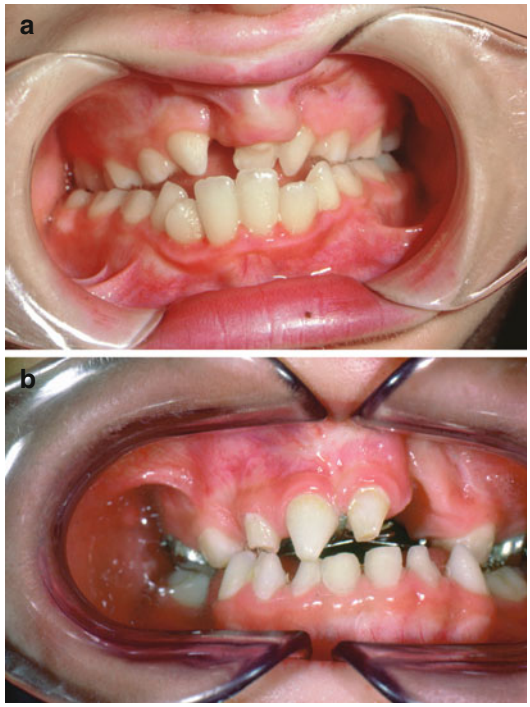


Fig. 8.5 Missing and malformed anterior teeth. (**a**) A very small premaxilla with one deciduous incisor but no permanent tooth buds. (**b**) Malformed deciduous maxillary anterior teeth and missing left deciduous anterior teeth

angles – are also distinct entities with a much higher incidence and different embryology (Gorlin and Pindborg 1964; Lemke 1959; Everett and Wescott 1961; Witkop 1964; Witkop and Barros 1963; Schuermann et al. 1966).

8.1.4 Association with Other Malformations

In addition to their strikingly common association with cleft lip and/or palate, pits of the lower lip have been noted in association with other malformations. Gorlin and Pindborg (1964) have found cases of lip pits along with anomalies of extremities, popliteal pterygia, and anomalies of the genitourinary system. The association with cleft lip and/or palate may well form a distinct new syndrome.

A review of the literature suggests that a variety of other anomalies may be associated with lip pits. Among them: syndactyly of the hands together with cleft lip and palate (Lannelongue 1879; Bernauds 1906); mental retardation and cleft, type not specified (Test and Falls 1947); ankyloglossia and cleft lip and palate (Van der Woude 1954); polythelia (Baxter 1939); symblepharon and cleft lip and palate (Oberst 1910); and ankyloblepharon, adhesion between the maxilla and mandible, and cleft uvula (Neuman and Shulman 1961). In two cases of the orofacial digital syndrome, pits were observed by Gorlin and Psaume (1962).

8.1.5 Inheritance

In most cases of lip pits described during the last 120 years, a marked hereditary pattern was observed. Although all authors have excluded

autosomal recessive inheritance or X-linked inheritance, there is no uniform opinion concerning whether or not the condition is due to a single autosomal dominant pleiotropic gene.

Fogh-Andersen (1942, 1961) was the first to clearly point out that the inheritance of clefts in the families with a history of lip pits is of a different character than in families where no lip pits occur. According to Fogh-Andersen (1942, 1961), the role of genetic factors in families with lip pits is much more pronounced, and both genetically different types of clefts (cleft lip or cleft lip and palate and isolated cleft palates) commonly are found within a single family. Fogh-Andersen also stated that in families in which fistulae of lower lip occur as a dominant hereditary character, there are some cases of cleft lip and cleft palate alone. Possibly, it may be explained as the result of coupling of neighboring genes.

Van der Woude (1954), in a careful study of five pedigrees with clefts and lip pits, found that the combination of pits and clefts is based on a single dominant gene of variable expressivity. She agreed with other authors (Test and Falls 1947) that a mildly affected individual can pass the trait on in a very severe form and a severely affected individual can pass the trait on in mild form. The sex of the individual is not a factor in passing on this anomaly. No sex limitation or preference exists.

Patients with clefts but without lip pits (or their parents) often ask for genetic advice. In most of these cases, only a small risk of cleft lip (less than 10 %) is indicated for the next child. But the counseling situation is significantly changed when associated lip pits are found. In this instance, all clefts in the family are considered part of the syndrome, and risk figures for clefts are remarkably higher.

8.1.6 Evidence of Heterogeneity

It is also clear that the risk of a cleft occurring in a child is significantly higher when the parent has lip pits and a cleft than when the parent has lip pits only. Two alternative explanations for this heterogeneity between families can be considered: (1) The development of clefts in persons carrying a "lip pit" major gene may be influenced by modifying genes at other loci, and (2) in some families, a mutant allele may produce lip pits with only occasional clefts, whereas in other families a different mutant allele (at the same or a different locus) may frequently lead to clefts in addition to lip pits. Thus far, efforts to use the data to support one or the other of these hypotheses have been unsuccessful. Cervenka et al. (1967) reported data from 66 individuals with lip pits in his study and 446 cases with lip pits from the literature with known sex to establishing a 1:1 sex ratio. The frequency of the syndrome was estimated as 1:75,000–1:100,000 in the white population.

Cervenka et al. (1967) further states that family histories can be explained adequately on the basis of autosomal dominant inheritance with variable expressivity of the trait. Penetrance is high, estimated at 80 %. Pits show up more frequently than clefts, and there is a significant association between the types of clefts in parents and their children. Possibly, the development of clefts in this syndrome is influenced by modifying genes or by different mutant alleles with a predilection for the different types of cleft.

8.2 Orthodontic Treatment, Dentition, and Occlusion

8.2.1 Crossbite Correction (Figs. 8.6, 8.7, 8.8, 8.9, 8.10, 8.11, and 8.12)

Bergland and Sidhu (1974) advocated postponing orthodontic treatment until complete eruption of the permanent anterior teeth. Segmental alignment can then be corrected with simultaneous manipulation of the anterior teeth. We believe it is best to start maxillary arch expansion when the deciduous dentition is completely erupted and when the children can be easily managed. A three-phase treatment is followed: (1) Buccal crossbite is corrected at 4–6 years, (2) anterior teeth are aligned at 8–9 years, and (3) final orthodontics is utilized at 11-plus years.

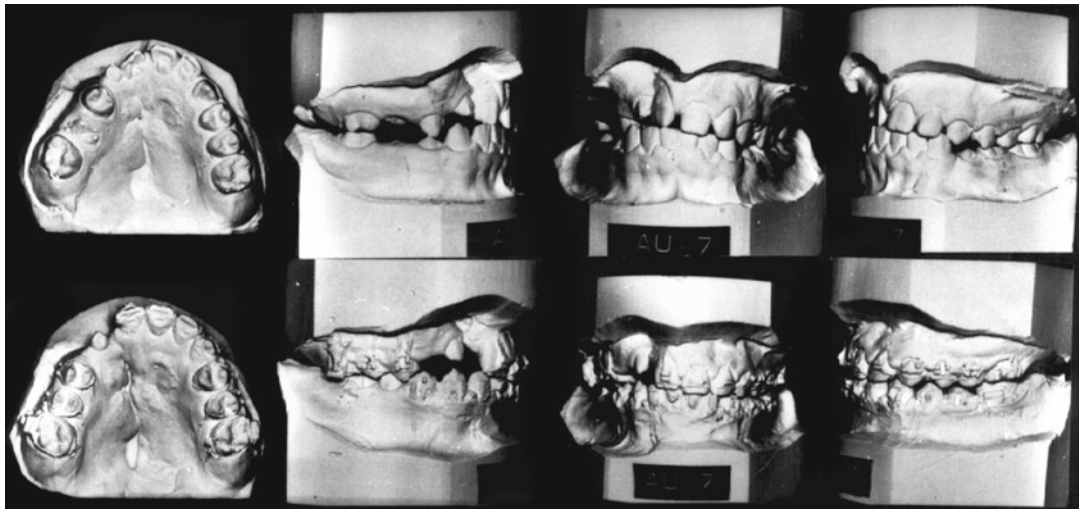


Fig. 8.6 Palatal expansion increases nasal width. At 12 years of age, buccal crossbite correction of the cleft segment was performed by palatal expansion. Crossbite correction is orthopedic in that the cleft bony segment is moved laterally, widening the nasal chamber on that side. *Top*: Right buccal segment is in crossbite. *Bottom*: After expansion: Palatal fistulae are exposed with the separation of the overlapped palatal segments

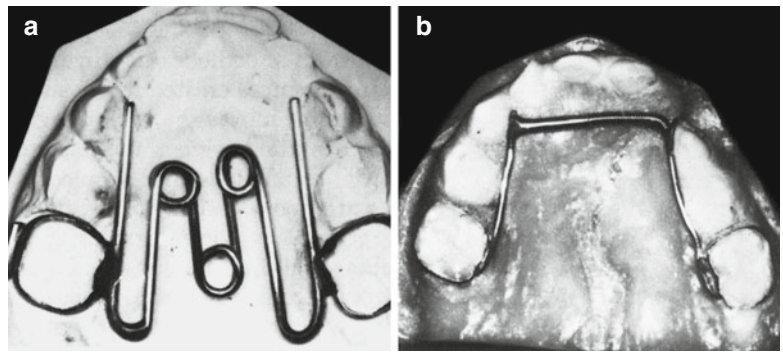


Fig. 8.7 (**a**) Fixed palatal helix expander used in deciduous dentition with or without an anterior finger spring. (**b**) Fixed "Arnold" expander using a compressed open coil spring to create an expansion force

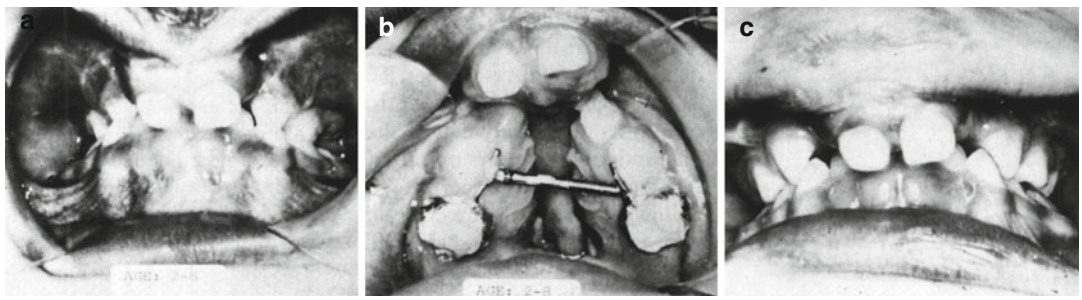


Fig. 8.8 (**a**–**c**) Crossbite correction for bilateral cleft lip. (**a**) Age 2 years 8 months. Note the bilateral crossbite. (**b**) Arnold expander in place. (**c**) Crossbite correction after 3 months brought on by the outward movement of the lateral palatal segments

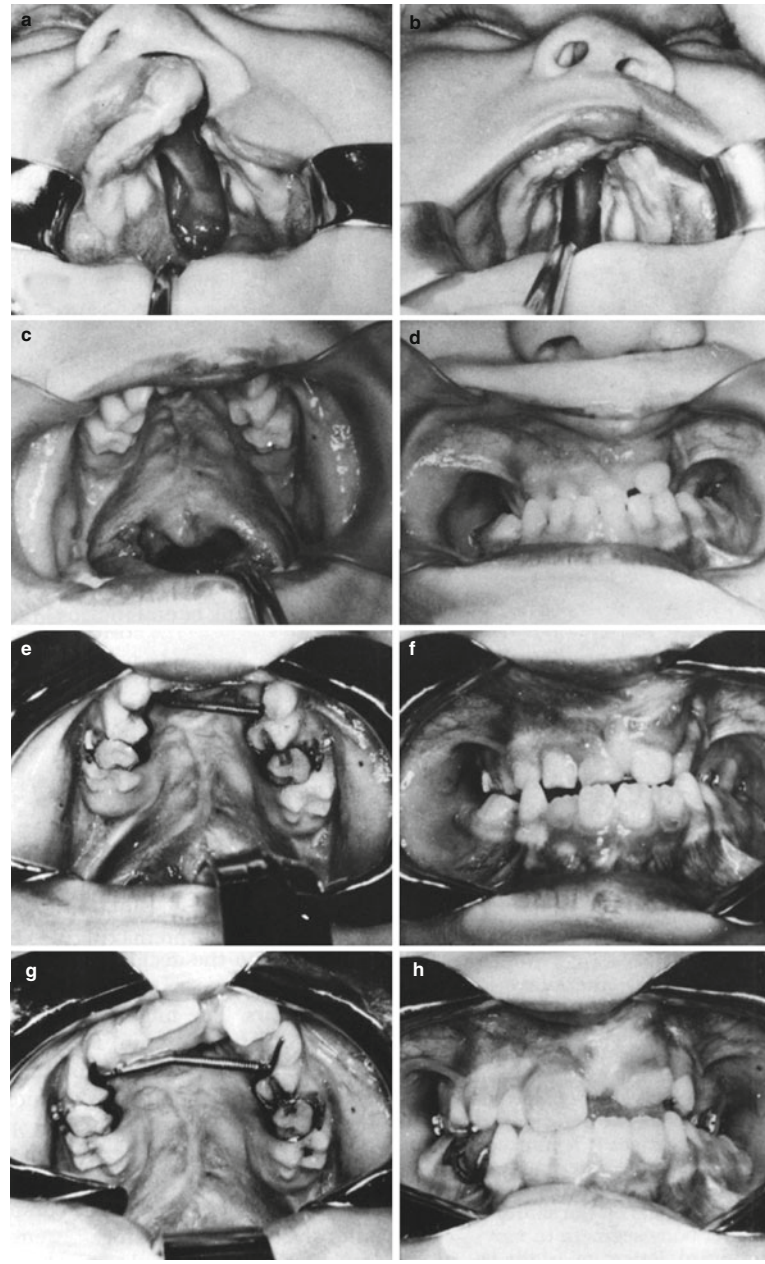


Fig. 8.9 (**a**–**h**) Correction of anterior and posterior crossbite in the deciduous and mixed dentition in a severely scarred palate. (**a**) Complete unilateral cleft lip and palate, unoperated. (**b**) Palatal segments in contact. (**c**) Palatal cleft closed at 6 months of age: von Langenbeck procedure was used to close a very wide cleft space. (**d**) Bilateral buccal and anterior crossbite. (**e**) Arnold expander is in place. (**f**) Crossbite is still present due to excessive scar tissue. (**g**) Expander is fully extended in the early mixed dentition. (**h**) Anterior crossbite was corrected, but the buccal crossbite is still present. Comments: The severely scarred palate prevents the lateral movement of the medially positioned palatal segments. Scars can only be stretched a slight amount, and if that amount is exceeded, the bony segments will not move, and instead the teeth will respond to the orthodontic forces. Should the teeth be tipped outwardly they will have to be permanently retained by a bridge or by teeth splinting

Fig. 8.11 (**a**) Orthodontic correction of an anterior and posterior crossbite in the permanent dentition. Anterior dental crossbite does not necessarily mean that the maxilla is anteroposteriorly deficient in size and requires a LeFort I advancement. In this case, the maxillary dentition was advanced orthodontically with the missing lateral incisor space opened to achieve interarch congruency. The expansion was maintained by complete arch splinting. (**b**) A severely ventroflexed premaxilla was uprighted during the deciduous dentition. In order to maintain the correction after the deciduous anterior teeth are lost, a fixed palatal retainer with an acrylic button is placed on the premaxilla's palatal incline

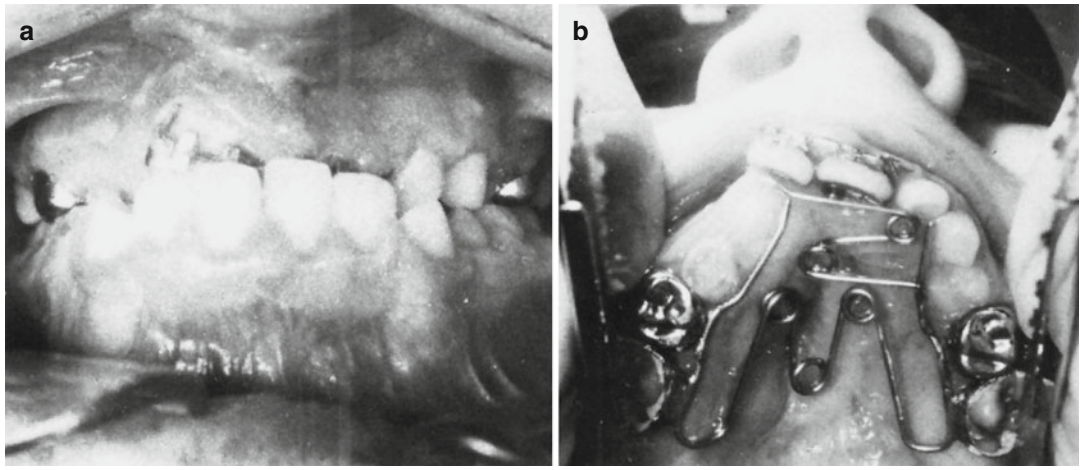
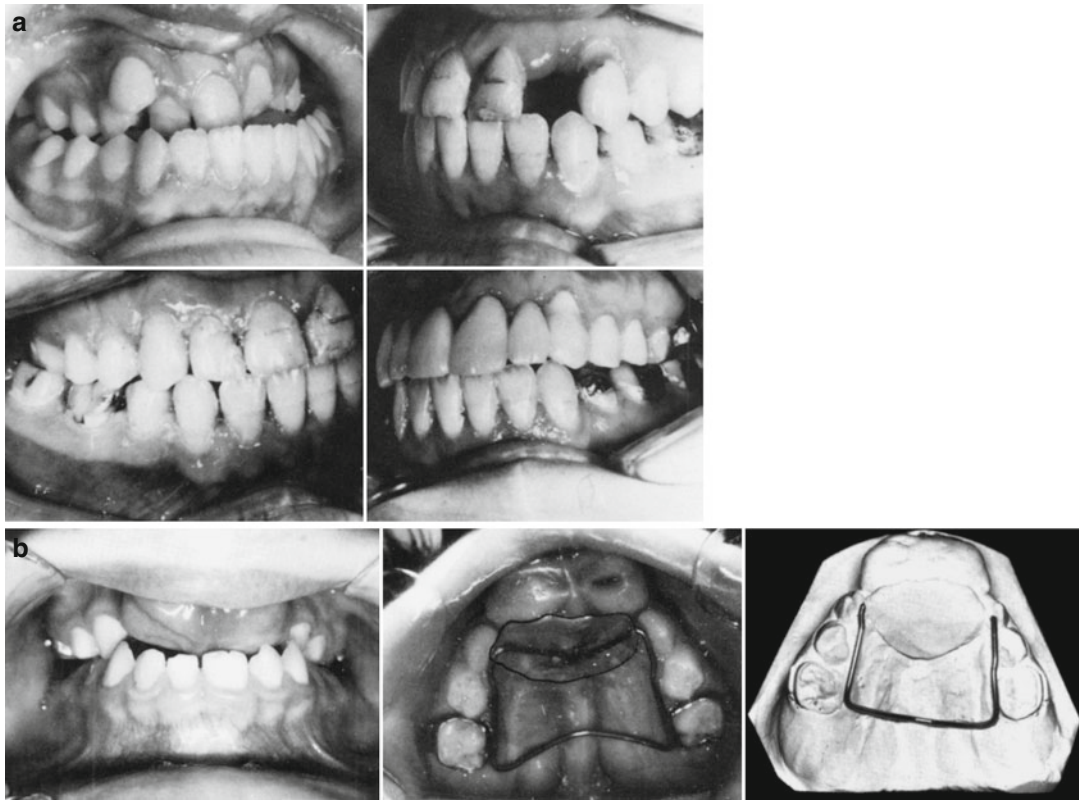


Fig. 8.10 (**a**, **b**) Anterior and buccal crossbite correction the mixed dentition. (**a**) Anterior crossbite was due to palatally displaced deciduous teeth with premaxillary segment and not to growth deficiency. The buccal teeth are in crossbite as a result of the lesser palated segment being palatally displaced. This picture has the maxillary anterior teeth in crossbite; the lower anterior teeth are shown while the upper teeth are hidden. (**b**) A fixed palatal expander with finger springs is used to expand the arch and advance the anterior teeth. In most instances, it is unnecessary to disocclude the anterior teeth to move them forward. Buccal expansion depends on the ability of the lesser bony segment to move outward. Retention of the bony correction is essential



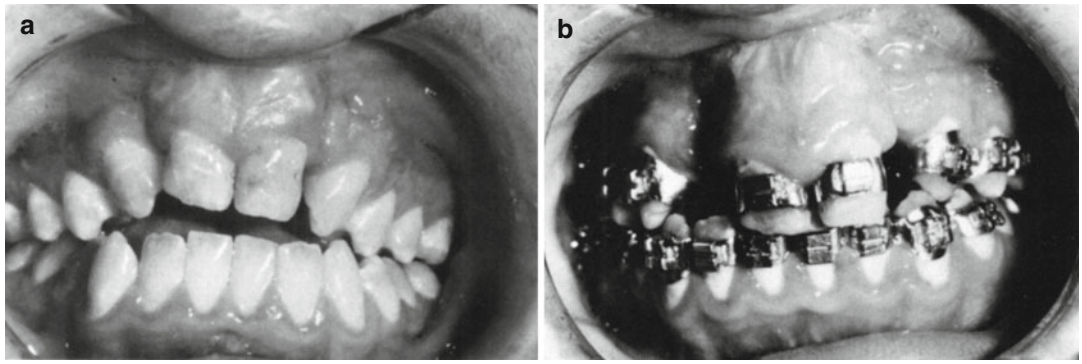


Fig. 8.12 (**a**, **b**) Repaired bilateral cleft lip and palate. An anterior openbite and retruded premaxilla resulting from inadequate orthodontic and surgical planning. (**a**) A slight anterior openbite was present at 8 years of age at which time a secondary alveolar bone graft was performed. The orthodontist attempted to close the lateral incisor spaces by bringing the cuspids mesially, while retracting the central incisors. This created a more severe openbite and an anterior crossbite. (**b**) The orthodontic mechanics were reversed. The lateral incisor spaces were opened, and the central incisors were advanced into an ideal overbite and overjet relationship. One of the bone grafts had to be redone. Comments: When the buccal occlusion is Class I, it is always better to keep the lateral incisor space open in both bilateral and unilateral cleft cases. There are occasions when a mandibular central incisor will need to be extracted in order to avoid flaring the maxillary central incisors and to obtain a proper occlusion. An anterior fixed bridge will stabilize the arch and replace the missing maxillary lateral incisors

Crossbite correction by moving palatal segments laterally in the presence of extensive palatal scarring is difficult and often unsuccessful. Extensive mucoperiosteal undermining, leaving wide denuded palatal bone, was necessary to close the wide palatal cleft at 6 months of age. Uniting the lip moved the palatal segments together (molding action) into good arch approximation. Palatal scar contracture moved the buccal segments farther medially, placing the buccal teeth in crossbite. The anterior dental crossbite does not necessarily reflect diminished anteroposterior maxillary growth but, rather, malposition of the premaxillary teeth of the greater segment.

8.2.1.1 Unilateral Cleft Lip and Palate

Deciduous and Mixed Dentition: When the smaller segment's alveolar process is contained within the premaxillary alveolar segment of the larger segment, a dental crossbite occurs between the maxillary and mandibular teeth. Dental dysplasia (eruption of a tooth out of position) may or may not coexist with segmental dislocation. A simple crossbite of one tooth may be due to its malposition rather than to the palatal segment's collapse. The most frequent crossbite is brought on by the mesioangular rotation of the lesser segment rather than by ectopic tooth eruption. Total buccal crossbite is seen less frequently but is generally present when the palatal tissue is scarred. In either case, the same helix type of palatal expander can be utilized and correction achieved within 2–4 months. Anterior movement of the deciduous central incisors often requires lingual undercuts either by orthodontic bands with lugs or by direct lingual shelf bonding to stabilize the anterior activated palatal finger springs.

A fixed palatal retainer can hold the correction until the second stage of orthodontic therapy is initiated. There is no way to predict the return of a crossbite in the absence of permanent retention. Bone grafting across the alveolar cleft does not guarantee retention of the corrected arch form.

Permanent Dentition: In Class I and Class III cases, it is usually preferable to open the missing lateral incisor space with the expectation of utilizing a fixed bridge to stabilize the corrected arch form and replace the missing tooth. In Class II cases, it may be possible to encourage the cuspid to erupt through the alveolar bone graft in

the lateral incisor space, which eliminates the need for extensive orthodontics and bridgework. The cuspid may need to be splinted to the central incisor in order to maintain the corrected arch form.

8.2.1.2 Bilateral Cleft Lip and Palate

First Stage: Between the ages of 4 and 6 years, the premaxilla is usually ventroflexed and overlaps one or both lateral palatal segments, which may be in partial or complete buccal crossbite. As in unilateral cases, treatment of bilateral cleft lip/palate necessitates moving bony segments into the surrounding muscle ring. If the premaxilla is to be moved forward, bands with lingual lugs are placed on the deciduous central incisors. A fixed helix-type palatal expander with anterior activated finger springs is cemented to the second deciduous molars. The finger springs are positioned under the central incisor-banded lugs for retention. The premaxilla is uprighted prior to correcting the buccal crossbite. The anterior and buccal crossbite correction can be completed within 6 months. As the premaxilla and lateral palatal segments are moved outward, the anterior cleft space is uncovered. A fixed palatal retainer with an acrylic anterior extension to cover the anterior cleft space is placed and kept in position until the alveolar cleft is bone-grafted and all fistulas are surgically closed. Permanent complete palatal retention is necessary, even after alveolar bone grafting. A premaxillary retainer must be placed when the deciduous anterior teeth are lost.

Second Stage: At 7–8 years of age, when the deciduous incisor teeth are replaced by permanent incisors, the anterior crossbite may have to be retreated. The incisors may be rotated and malpositioned. Orthodontic brackets are placed in the upper arch to support a labile arch wire, which will be utilized to reposition the incisor teeth and reduce the premaxillary overbite. A pontic tooth with band is placed on the arch wire to achieve a more pleasing aesthetic result. The premaxilla needs to be properly aligned with the lateral palatal segments prior to alveolar bone grafting; this procedure may be performed between 7 and 9 years of age. A lateral incisor as well as a cuspid may erupt through the area of new bone formation.

Third Stage: Children at 10+ years of age are treated as any other child. The many malocclusion possibilities render it impossible to develop treatment plans for each contingency; instead, basic treatment problems are discussed:

1. Class I malocclusion (with anterior and/or buccal crossbite): The crossbite generally is due to lingual bony displacement. The upper arch should be advanced and expanded. If there is sufficient arch length, the missing lateral incisor area is left open. In arch shortage cases, it may be preferable to move the cuspid adjoining the space into the lateral incisor area rather than extract the first bicuspid on that side. A number of variations of treatment exist according to tooth size and location.
2. Class II malocclusion: The anterior overjet may be corrected by retraction of the premaxillary central incisors in bilateral cleft lip and/or cleft palate cases. If one or both lateral incisors are missing, it may be best to place the adjoining cuspid in that space and extract either the opposite lateral incisor or the first bicuspid.
3. Class III malocclusion: An anterior crossbite does not necessarily signify that a Class III malocclusion exists. In some cases, the anterior teeth can be advanced without excessively increasing their axial inclination. A true depressed midface with poor vertical and anteroposterior development requires midfacial lengthening and advancement. Segmental surgery may have to be utilized to overcome palatal width problems caused by excessive scarring.

8.2.1.3 Use of Orthopedic Forces to Correct Midfacial Recession

Developing anterior crossbites in either mixed or permanent dentition can be corrected using orthopedic protraction forces. These forces must average 800 g/side and pull downward and forward off hooks placed between the lateral incisor and cuspid. The force needs to be applied 12 h/day. With good cooperation, 5–10 mm of midfacial advancement can be accomplished. The preferred facial mask is the Delaire type.

8.2.2 Supernumerary (Extra) Teeth, Missing Teeth, and Aplasia (Malformed Teeth)

This occurs more frequently in children with cleft lip and/or palate than in other figuration of the nasal floor (Figs. 8.5, 8.6, 8.13, and 8.14). Bishara and coworkers (1985) studied untreated adults in India who had clefts of the lip and alveolus only, unilateral cleft lip and palate, and bilateral cleft lip and palate. They observed that the maxilla and cranial base were not different from a matched normal population but that the relation of the maxilla and mandible to the cranial base varied according to cleft type. Moss (1969), Moss et al. (1968), Blaine (1969), Dahl (1970), and Krogman et al. (1975) have stated that the cranial base in cleft palate patients differed in both size and shape from noncleft individuals.

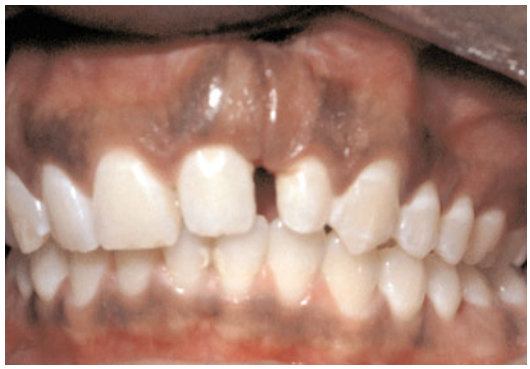


Fig. 8.13 The left central and lateral incisor in the line of an alveolar cleft areas reduced in size

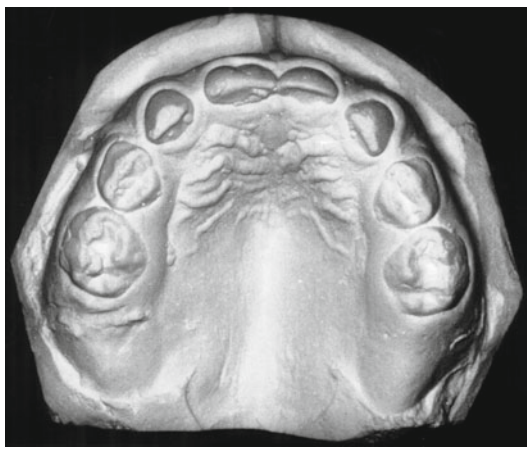


Fig. 8.14 Tooth abnormalities. Coalescence of the right deciduous central and lateral incisors in a right unilateral cleft of the lip and alveolus

The incidence reported in different articles has varied because it is difficult to distinguish between variations rooted in congenital causes and those related to surgery (Bohn 1963). Recently, it has been observed that supernumerary teeth are more common in the deciduous dentition. Moreover, the incidence of supernumerary teeth is greatest in cases of cleft lip only and decreases as the extent of the cleft increases. The relationship is the opposite in cases of aplasia; the incidence of aplasia is lowest for cleft lip only and cleft palate only and increases in proportion to the extent or complexity of the cleft (Brook 1984; Garn et al. 1959, 1960, 1965; Brabant 1967).

In conditions of facial clefting, dental development is, except for the third molars, delayed for all teeth, both maxillary and mandibular (Brook 1984; Garn et al. 1959). Asymmetrical development of tooth pairs, with delayed development on the cleft side, was recorded in approximately half of a group of children with congenital lip and/or palate clefts (Brabant 1967). This supports other observations that eruption is delayed in both dentitions (Garn et al. 1960, 1971, 1977a, b; Ranta 1971, 1972, 1973a, b; Fanning 1961; Dixon 1968; Falkner 1957; Delgado et al. 1975; Demirjian 1986; Fishman 1970; Foster and Lavelle 1971; Galili et al. 1969; Haring 1976; Hatton 1955; Haavikko 1985; Haataja et al. 1972).

Zilberman (1973), from a study on clefts of the lip and alveolar structures, and Mirsa and colleagues (1972), after investigating clefts of the lip and palate, reported that unilateral clefts are more frequent on the left side and are more common in males than in females.

The incidence of dental malocclusion reported in patients with cleft lip and/or palate varied widely in studies by Huddart and Bodenham (1972), Hellquist et al. (1979), Dahl et al. (1981), Norden and associates (1973), Bergland and

Sidhu (1974), Nylen and coworkers (1974), Ranta and colleagues (1974a, b), and Hellquist and Skoog (1976). This may be because the patients had varying types of clefts, and their cases were recorded at different ages. Rehrman and coauthors (1973) found the incidence of malocclusion in the mixed dentition to be twice that in the deciduous dentition.

In cases of cleft palate only, Ranta and colleagues (1974a, b) found only a slight increase in anterior crossbite at the transition from the deciduous to the mixed dentition. A noticeable increase in the incidence of anterior crossbite in the mixed dentition, in cases of complete unilateral clefts of the lip and/or palate, was reported by Bergland and Sidhu (1974). This was irrespective of the arch configuration in the deciduous dentition. They also reported that palatal segments stabilized early after lip repair and that further collapse was the exception. However, contrary to the findings just cited, Nylen and coworkers (1974) found no increase in the frequency of anterior crossbite in their mixed-dentition group.

8.2.3 Caries

Dahl et al. (1989) reported the incidence of caries, gingivitis, and dental abnormalities in preschool children with cleft lip and/or palate in Stockholm, Sweden. Oral health was studied in 49 children 5–6 years old with clefts of the lip and/or palate (CL/P) and 49 healthy children matched for sex and age. The results showed a statistically significant increase in the prevalence and activity of caries among the CL/P children. The average number of decayed and filled tooth surfaces in the cleft group was 7.0 compared with 3.9 in the control group ($p < 0.05$).

The most evident difference between the two groups was found in the number of decayed proximal surfaces. The mean number of decayed proximal surfaces in the CL/P group was 2.5, compared with 0.9 in the control group ($p < 0.001$). No significant differences were found in the prevalence and activity of caries among children with isolated clefts of the lip or palate.

The children with cleft lips/palates also exhibited a significant increase ($p > 0.01$) in the number of gingival units with gingivitis. Other dental abnormalities included increased enamel hypomineralization ($p < 0.05$), supernumerary teeth ($p < 0.001$), unilateral crossbite ($p < 0.001$), and mesial terminal plane ($p < 0.01$). These results clearly show that children with CL/P as a group must be considered to have an increased risk of caries and gum disease and should therefore have the benefit of additional preventive programs (see Chap. 2).

8.3 The Relationship Between the Clefting Process and Contiguous Skeletal Structures

Some studies have indicated that clefting is not an isolated defect but may be a syndrome phenomenon with ramifications in contiguous and often remote structures.

In a study of Danish males, Dahl (1970) suggested that the presence of cleft palate, with or without cleft lip, may have ramifications for distant craniofacial structures and their development. Farkas and Lindsay (1972) identified consistent variations in facial morphology in the cleft population and concluded that the cleft defect was not an isolated condition. They reported that what might otherwise be considered the normal side of the face in cases of unilateral clefts was not completely normal and that the anomaly influenced the development of the face equally on both sides.

8.3.1 The Position of the Cleft Maxilla Within the Cranium and the Mandible

Berkowitz (1985) undertook a mixed cross-sectional study of CUCLP and CBCLP cases to determine whether the maxillary complex relative to the mandible is posteriorly positioned within the face by studying the dental occlusion. None of the cases had presurgical orthopedics,

and the hard palate clefts were closed between 18 and 28 months of age using a modified von Langenbeck procedure with a vomer flap. This study was designed to test McNeil's thesis that the palatal segments, being detached from the nasal septum, are not only reduced in mass but also have not been brought forward with the developing nasal septum. This failure would lead to retrusive midface with a Class III malocclusion.

Berkowitz (1985) found that the occlusal relationships at 6 years of age did show Class I and Class II occlusions, but none of the cases had a Class III occlusion, which would have been present if McNeil's (McNeil 1950, 1954) hypothesis had been valid. Of the 29 bilateral cases, five cases had a crossbite on one side, one case had a complete bilateral crossbite, and six cases had no crossbites at all. It is quite evident that a buccal crossbite is not, as stated by McNeil, a predictable outcome of the presence of a palatal cleft.

Semb's (1991a, b) and Ross's (1987a, b) studies and those already acknowledged elsewhere established that in the cleft population, both the maxilla and mandible are retropositioned within the face (see Chap. 9). However, if McNeil's beliefs were accurate, the bilateral cleft palatal segments would have been left behind in their growth, and a greater proportion of the cases would have shown a Class III malocclusion on one or both sides.

Chierici and associates (1973) and Bishara and Iversen (1974), Bishara et al. (1985) found a relative retrusion of the maxilla and mandible as well as increased steepness of the mandibular plane in various cleft types. Krogman and his colleagues (1975) reported significant differences in the cleft population in the size of the cranial base, its configuration, and direction of growth. They concluded that the clefting process has growth and/or development implications for the contiguous cranial base and facial structures as well as for the maxilla.

Bishara and Iversen (1974) reported that the posterior positioning of the maxilla and mandible relative to the anterior cranial base may result from the cleft's influence on contiguous skeletal structures and that clefting affects maxillary development and facial morphology.

8.3.2 The Cranial Base

Hayashi and colleagues (1976) investigated cranial growth of a large sample of subjects from 4 to 18 years of age with complete unilateral clefts. The investigators found that the cranial base angle was flatter, the maxilla was more retruded, and underdevelopment in both the maxilla and the mandible was more pronounced in girls than in boys. They speculated that upper face height in patients of both genders was less than normal as a result of cleft interference with nasal septal and maxillary suture growth and changes in the configuration of the nasal floor.

8.3.3 Relationship of the Nasal Cavity to Arch Form (Figs. 8.15 and 8.16)

Aduss and Pruzansky (1967) wrote that the anatomic distortions common to all of their patients with clefts included marked deviation of the nasal septum toward the noncleft side; flattening, particularly of the inferior turbinate on the cleft side; and an anterolateral displacement of the noncleft segment, with an outward and lateral rotation of the premaxillary area adjoining the cleft.

These distortions are the result of unbridled septal growth, deviant maxillary growth, and aberrant vectors of muscle pull. Establishing a continuous muscle band across the cleft, by lip repair, can bring the palatal shelves closer together and modify the configuration of the palatal segments, as well as the configuration of the internal nares.

Peyton and Ritchie (1936), measuring the displacement of the soft tissues of the nose in complete unilateral cleft lip and palate, have shown that deviation of the external part of the nose toward the noncleft side extends for the entire length of the nose, with the greatest displacement at the tip. They further demonstrated that growth of the nasal structures is the same in noncleft children and children with complete unilateral cleft and that the early cleft deformity decreases with time. The natural tendency for self-correction of the septal deviation was evident in the continual uprighting and medial movement of the end of the septum observed in all cases.

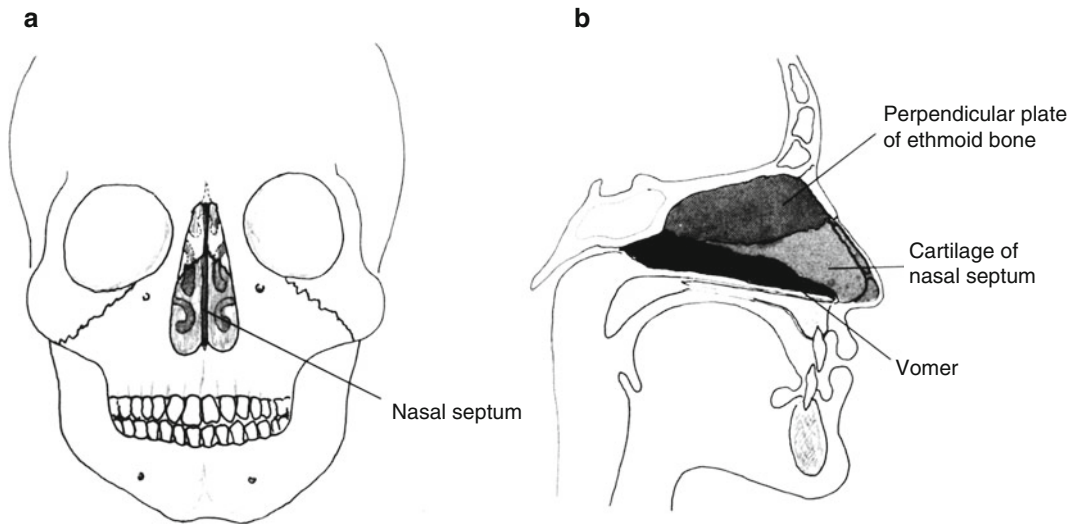


Fig. 8.15 (**a**) Frontal view. The septum is straight dividing the nasal chamber in two equal parts. (**b**) Lateral view. The components which make up the nasal septum: the ethmoid bone, the nasal cartilage, and the vomer

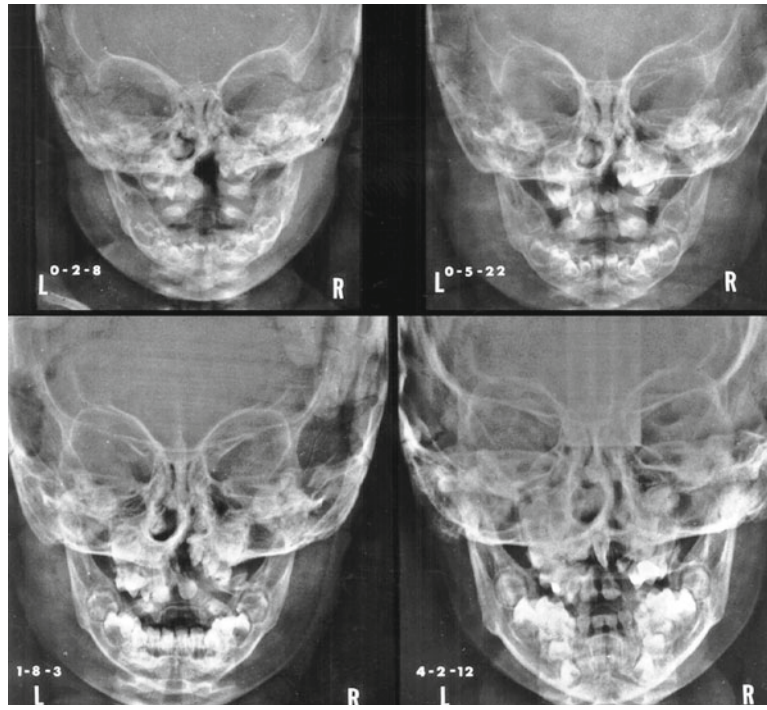


Fig. 8.16 Serial frontal cephalometric radiographs illustrating geometric changes to the nasal chamber in a CUCLP from birth to 4 years of age before and after palatal expansion. *Top*: 0-2-8 At birth a widened nasal chamber is evident. 0-5-22 After lip surgery the nasal chamber has narrowed. *Bottom*: 1-8-3 The inferior turbinate on the cleft side (R) makes contact with the vomer. 4-2-12 After palatal expansion, the nasal width and the septum to inferior turbinate distance has increased. Because the roof of the mouth is also the floor of the nose, any disarrangement in the architecture of the roof of the mouth is reflected in the nasal chamber. Prior to lip repair, the nasal septum is displaced to the noncleft side. After lip repair with the medial movement of the cleft segment, the septum bows toward the nasal chamber on the cleft side. After palate repair, there is continual palatal movement with septal uprighting and decreased septal bowing. The turbinates on the cleft side are flatter, and the buccal teeth on the cleft side may be in crossbite. The nasal chamber on that side is narrowed

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Part III

Facial Growth in Cleft Palate Children

Characteristics of Facial Morphology and Growth in Infants with Clefts

9

Sven Kreiborg, Nuno V. Hermann,
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9.1 Introduction

Congenital clefts of the lip and/or palate can arise in isolation or together with other malformations (syndromes) (Gorlin et al. 2001). This chapter deals solely with "nonsyndromic" clefts. Both individuals with unoperated and operated clefts have a face which differs from those of unaffected individuals. Since the introduction of roentgencephalometry more than 70 years ago (Broadbent 1931), hundreds of cephalometric studies, including both unoperated and operated cleft individuals, have suggested that some deviations are directly caused by the primary anomaly, while others are caused by the surgical interventions and the following dysplastic and compensatory growth of the facial bones (e.g., Graber 1949, 1954; Slaughter and Brodie 1949; Ortiz-Monasterio et al. 1959, 1966; Dahl 1970; Pruzansky 1971; Bishara and Olin 1972; Friede and Pruzansky 1972a, b; Bishara 1973; Friede and Johanson 1974; Bishara et al. 1976, 1985, 1986; Friede and Morgan 1976; Friede 1977, 1978, 1998; Friede et al. 1986; Smahel et al. 1987; Ehmann 1989; Mars and Houston 1990; da Silva Filho et al. 1992b, 1998; Capelozza et al. 1993, 1996; Tomanova and Müllerova 1994; Berkowitz 1995; Dahl and Kreiborg 1995; Semb and Shaw 1996; Sandham and Foong 1997; Friede and Enemark 2001). However, the relative importance of the intrinsic factors, the iatrogenic factors, and the functional or adaptive factors for the facial development is still unclear. There are probably several reasons for this. Firstly, comprehensive knowledge of craniofacial morphogenesis in cleft newborns or infants before surgery, based on large, consecutive, well-controlled samples, is very scarce. This situation is not surprising since, in developed countries, the cleft of the lip is surgically treated within the first couple of months after birth. Thus, the possible period of examining the unoperated state is short, and several methodological problems are involved. Secondly, the cephalometric analyses are most often limited to the lateral projection using simplistic cephalometric analyses, typically based on 15–20 reference points and almost invariably measuring maxillary prognathism as the *S-N-A* angle or similar measurements to the premaxilla, and the use of infant cephalometry has been very limited.

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These authors are of the opinion that incomplete knowledge about the intrinsic factors related to the cleft anomaly has automatically lead to excessive emphasis on the importance of iatrogenic and adaptive factors in facial development of cleft children.

9.2 The Danish Experience

In the middle of the 1970s, we decided to take advantage of the very favorable sampling conditions in Denmark in an effort to contribute to the question of the characteristics of facial growth and development in children born with clefts (Jensen et al. 1988). In Denmark, for more than 65 years, all newborns with facial clefts have been recorded at the Institutes for Speech Disorders in Copenhagen and Århus. Repeated follow-up examinations have shown that the registration of clefts in Denmark is highly reliable and nearly complete. The population is homogeneous and stable, and only very few children are lost to follow-up. Furthermore, all primary cleft surgery is performed in one hospital by one surgeon.

Inspired by Pruzansky and Lis (1958), we constructed a three-projection infant cephalometer, which can obtain truly orthogonal lateral, frontal, and axial cephalograms (Kreiborg et al. 1977). A comprehensive cephalometric analysis system was developed including all craniofacial regions (calvaria, cranial base, orbits, maxilla, mandible, airway, cervical spine, and soft tissue profile) (Kreiborg 1981; Heller et al. 1995; Hermann et al. 2001a), and the method was validated (Hermann et al. 2001a). Furthermore, new methods of visualization of differences in craniofacial morphology and growth between different groups were developed using mean plots (Kreiborg 1981; Hermann et al. 2001a), color-coded vector plots (Hermann et al. 2001a), and color-coded surfaces on a 3D CT-model (Darvann et al. 1999).

During the 6 years from 1976 to 1981, there were 359,027 live births in Denmark. A total of 678 newborns of Northern European ancestry with cleft lip, cleft palate, or both were registered in the period. Twenty-four infants died before 22 months of age, and for practical reasons, material uptake had to be omitted in some patients with isolated cleft palate. Only nonsyndromic clefts were included in the study, but 602 of the 678 children (about 90 %) were examined by us (Jensen et al. 1988) and nearly all at both 2 months of age (before any surgical or orthopedic treatment) and at 22 months of age (before closure of the posterior palate in the children with clefts of the secondary palate). All children were treated by the same surgeon (Dr. Poul Fogh-Andersen), and in the children with cleft of the primary palate, the cleft lip was, in all cases, closed using a Tennison procedure. One-third of the children had isolated cleft lip (CL), about 40 % had combined cleft lip and palate (CLP), and about 27 % had isolated cleft palate (CP). The clefts were subclassified according to the method of Jensen et al. (1988).

In the 602 children included in the study, cephalograms were obtained in the lateral, frontal, and axial projections by three experienced orthodontists (Dr. Birgit Leth Jensen, Dr. Erik Dahl, and Dr. Sven Kreiborg). In addition, impressions were made of the maxilla, and anthropometric registrations (body height, body length, and head circumference) were carried out. The results of the cephalometric analyses have been presented in a number of publications (Dahl et al. 1982, 1989; Kreiborg et al. 1985; Kreiborg and Cohen 1996; Darvann et al. 2001; Hermann et al. 1999a, b, 2000, 2001a, b, 2002, 2003a, b, 2004; Kreiborg and Hermann 2002). So far, we have analyzed infant craniofacial morphology and early craniofacial growth in detail in three dimensions in the following groups: unilateral incomplete cleft lip (UICL), isolated cleft palate (ICP), Robin sequence (RS), unilateral complete cleft lip and palate (UCCLP) (Fig. 9.1a), and bilateral complete cleft lip and palate (BCCLP) (Fig. 9.1b). In the following, we shall summarize our findings, with emphasis on the

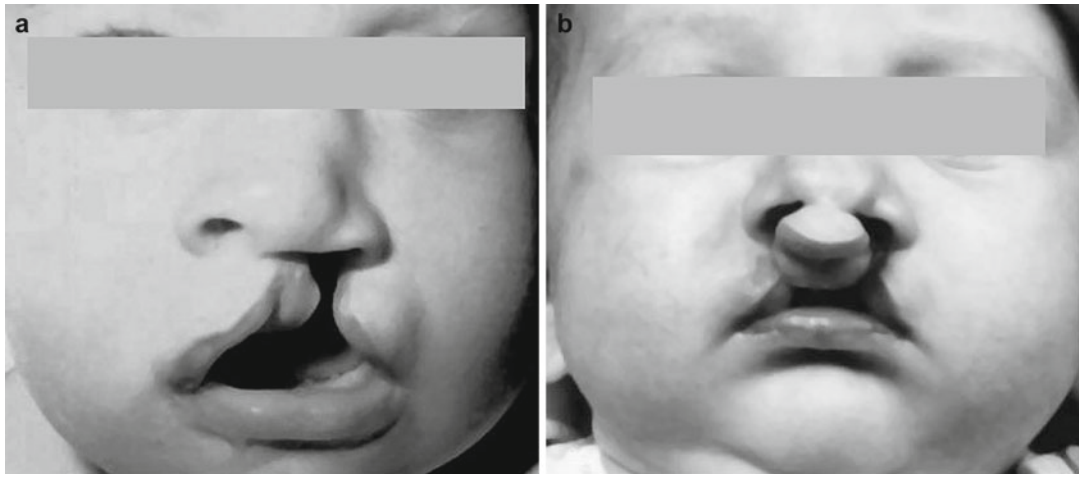

Fig. 9.1 (**a**) The facial morphology in a 2-month-old unoperated infant with unilateral complete cleft lip and palate (UCCLP). (**b**) The facial morphology in a 2-month-old unoperated infant with bilateral complete cleft lip and palate (BCCLP)

unoperated infant to shed light on the intrinsic factors related to the cleft condition (see Fig. 9.2 and Table 9.1), and compare them to data in the literature on unoperated adolescents and adults with clefts.

9.2.1 Cleft Lip (CL)

Isolated CL involves only structures of the embryonic primary palate. The craniofacial morphology in CL subjects has been shown to be fairly normal except for the small region of the cleft including the premaxilla and the incisors. In unoperated bilateral complete CL, the premaxilla may, however, protrude markedly. In unilateral *complete* CL, the protrusion is less pronounced but asymmetric. In subjects with unoperated unilateral *incomplete* cleft lip (UICL), the protrusion of the premaxilla is negligible (Hermann et al. 1999a). The interorbital distance in CL subjects seems to be slightly increased compared to the norm (Cohen 1997). The basal part of the maxilla has a normal prognathism in relation to the anterior cranial base, and the mandible is of normal size, shape, and inclination (Dahl 1970; Hermann et al. 1999a). Following lip surgery, the premaxilla is molded into a normal position, and maxillary prognathism measured to *point A* or *ss* (*subspinale*) is normal (Dahl 1970; Han et al. 1995; Hermann et al. 1999a, b, 2000). In conclusion, subjects with UICL have a very close to normal craniofacial morphology from infancy to adult age, and consequently, we have used our group of infants with UICL as a control group in the study of deviations in craniofacial morphology and growth of infants and young children with ICP, RS, UCCLP, and BCCLP since no actual normative cephalometric data for Danish infants and young children are available.

9.2.2 Cleft Palate (CP)

Isolated cleft palate (ICP) involves only structures of the embryonic secondary palate. In Fig. 9.2a, the mean facial diagrams of the ICP group are superimposed on the mean facial diagram of a group of age-matched infants with UICL (control group). The major deviations in the ICP group were: reduced length and posterior height of the maxilla, maxillary retrognathia, increased width of the maxilla and the nasal cavity, and reduced length of the mandible with mandibular retrognathia. Thus, the ICP group revealed *bimaxillary* retrognathia.

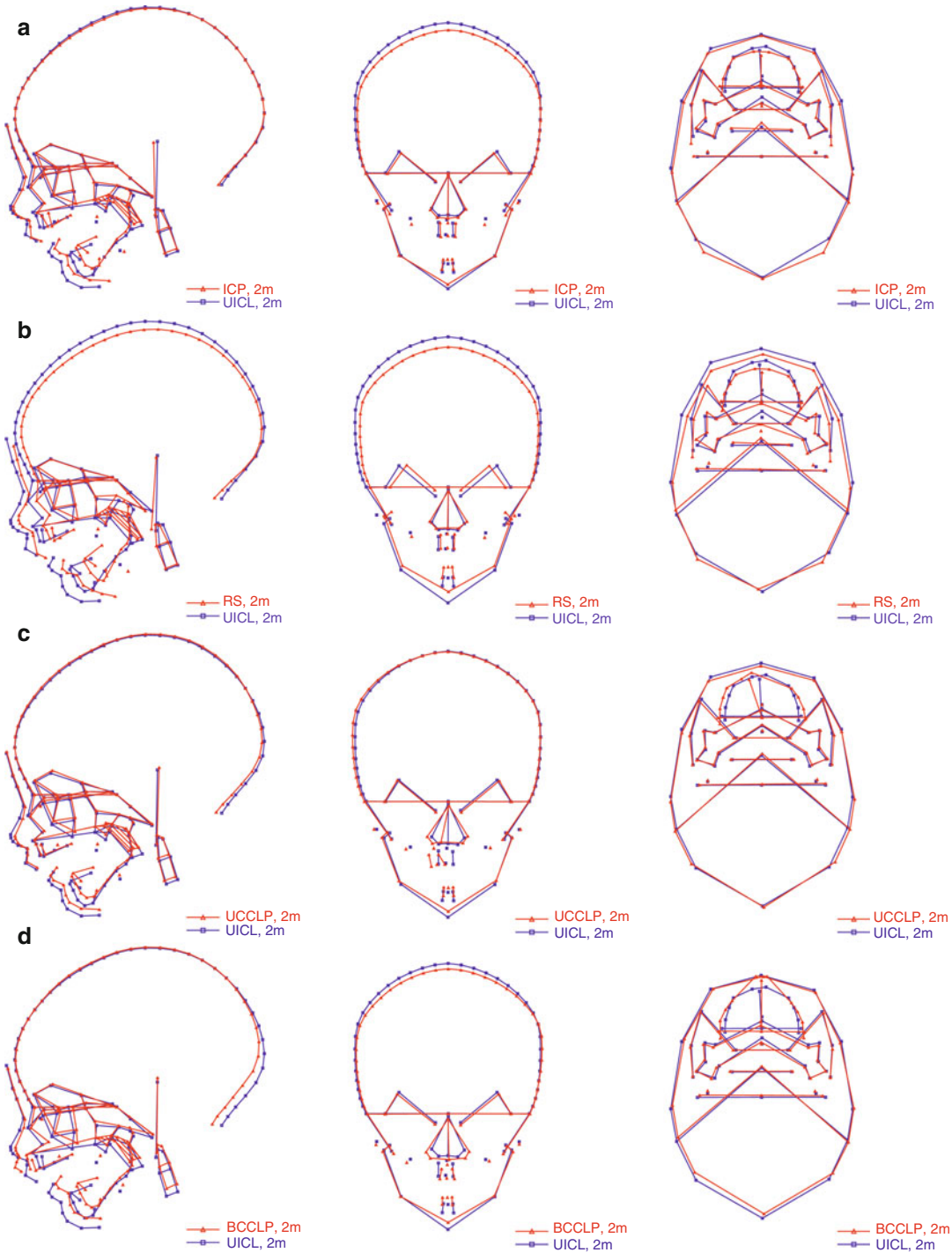

Fig. 9.2 (**a**–**d**) Mean plots in three projections (lateral, frontal, and axial) of the four different cleft groups superimposed on the control group with UICL. The lateral mean plots are aligned on the n-s line and registered at s. The frontal mean plots are aligned on the latero-orbital line and registered at the center point of that line. The axial mean plots are aligned on a line between the two tuber points and registered at the center point of that line. Superimposition of the mean plots for the 2-month-old (**a**) ICP and UICL groups, (**b**) RS and UICL groups, (**c**) UCCLP and UICL groups, and (**d**) BCCLP and UICL groups

Table 9.1 Summary and comparison of the most important findings in the primary anomaly in children with RS, ICP, BCCLP, and UCCLP

Anomaly	RS	ICP	BCCLP	UCCLP
Maxilla				
Decreased length measured to premaxilla (sp-pm)[a]	+[b]	+	–[c]	–
Retrognathia measured to premaxilla (s-n-ss)[d]	+	+	–[e]	–
Decreased posterior length (ci-pm)[f]	+	+	+	+
Retrognathia measured to base of jaw (s-n-ci)[g]	+	+	+	+
Decreased posterior height	+	+	+	+
Increased width	+	+	++	++
Nasal cavity				
Increased width	+	+	++	++
Mandible				
Decreased length	+++	++	++	+
Retrognathia	+++	++	++	+
Pharyngeal airway				
Reduced size	+++	++	+	+

RS Robin sequence, *ICP* isolated cleft palate, *BCCLP* bilateral complete cleft lip and palate, *UCCLP* unilateral complete cleft lip and palate

[a]sp-pm: anterior nasal spine to point pterygomaxillare

[b]The deviation from the norm is shown as + or –, meaning, e.g., that decreased total length of the maxilla was observed in the ICP and RS groups but not in the UCCLP group and that the length of the mandible is decreased in the UCCLP group, very decreased in the ICP and BCCLP groups, and severely decreased in the RS group

[c]The total length was significantly increased

[d]s-n-ss: S-N-A

[e]The prognathism was increased measured to the premaxilla

[f]ci-pm: Point crista infrazygomatica to point pterygomaxillare

[g]s-n-ci: Maxillary prognathism measured to the infrazygomatic crest

The sagittal jaw relationship was, however, normal. In addition, in the ICP group, the upper airway dimensions were reduced. Bimaxillary retrognathia and a short mandible were previously documented in unoperated older children and adults with ICP (Dahl 1970; Bishara 1972).

9.2.3 Robin Sequence (RS)

Robin sequence (RS) is defined as a triad of symptoms: isolated cleft palate, micrognathia, and glossoptosis (Gorlin et al. 2001). RS may be part of several syndromes, e.g., Treacher-Collins syndrome (Kreiborg and Cohen 1996; Cohen 1997). In this chapter, only nonsyndromic cases of RS will be discussed. We consider this group as a subgroup of the ICP group (Hermann et al. 2003a). In Fig. 9.2b, the mean facial diagram of the RS group at 2 months of age is superimposed on the mean facial diagram of the control group. The major deviations in the RS group were decreased length and posterior height of the maxilla, maxillary retrognathia, increased width of the maxilla and nasal cavity, and very short mandible with marked mandibular retrognathia. Thus, the RS group revealed *bimaxillary retrognathia*; the retrognathia was, however, most marked for the mandible, and the sagittal jaw relation was increased. In addition, the RS group had a significantly smaller cranial base angle (*n-s-ba*) resulting in a smaller depth of the bony nasopharynx than the controls, and the upper airway dimensions were markedly reduced. The degree of maxillary retrognathia was similar in the RS and the ICP group. However, the mandibular retrognathia in the RS group was even more marked than in the ICP subjects. It would seem that RS subjects probably represent the extreme part of the ICP population in terms of mandibular retrognathia and upper airway constriction. As mentioned above, we consider the RS group as a special subgroup of the ICP group. Accordingly,

we believe the bimaxillary retrognathia to be intrinsically associated with the cleft of the secondary palate.

9.2.4 Cleft Lip and Palate (CLP)

Combined clefts of the lip, alveolus, and palate involve structures of both the embryonic primary palate and secondary palate. In Fig. 9.2c, the mean craniofacial morphology in 2-month-old unoperated infants with unilateral complete cleft lip and palate (UCCLP) was compared to the control group (Hermann et al. 1999a). The major deviations in the UCCLP group were decreased posterior length and height of the maxilla; retrognathia of the basal part of the maxilla with relative protrusion of the premaxilla; the width of the maxilla and nasal cavity was markedly increased and the premaxilla deviated to the noncleft side; and the mandible was short and retrognathic. Thus, the UCCLP group revealed *bimaxillary retrognathia* combined with a relative protrusion of the premaxilla, which deviated to the noncleft side. In addition, in the UCCLP group, the upper airway dimensions were reduced.

Increased width of the midface and nasal cavity was previously reported in *unoperated* UCCLP infants (Han et al. 1995) and in *unoperated* adults with UCCLP (Motohashi et al. 1994). Relative protrusion and asymmetry of the premaxilla have also been reported in *unoperated* UCCLP children, adolescents, and adults (Ortiz-Monasterio et al. 1959, 1966; Bishara et al. 1976, 1985, 1986; Capelozza et al. 1993). The relative protrusion and deviation are probably due to overgrowth in the premaxillary-vomerine complex (Pruzansky 1971; Friede and Morgan 1976; Friede 1978) and due to the lack of structural integrity of the maxilla on one side. This relative protrusion of the premaxilla explains why we found the measurements *s-n-ans* (*S-N-ANS*) and *s-n-ss* (*S-N-A*) in the infant UCCLP group to be comparable to the values in the control group, despite the fact that the UCCLP group showed significant maxillary retrognathia measured to the basal part of the maxilla.

Dahl et al. (1982) and Hermann et al. (2003a, b) analyzed facial morphology in 2-month-old infants with unoperated bilateral complete cleft lip and palate from our sample. Fig. 9.2d illustrates the mean facial diagram of the BCCLP group superimposed on the mean facial diagram of the control group. The most obvious features in the BCCLP group were protrusion of the premaxilla both in relation to the anterior cranial base and in relation to the basal part of the maxilla; the length of the basal part of the maxilla and posterior maxillary height were decreased; retrognathia of the basal part of the maxilla; markedly increased width of the maxilla and nasal cavity; a short and retrognathic mandible. Thus, the BCCLP group revealed *bimaxillary retrognathia* with a truly protruding premaxilla. In other words, the protruding premaxilla was situated in a totally retrognathic face with a fairly normal sagittal jaw relationship. In addition, the upper airway dimensions were reduced.

The extreme protrusion of the premaxilla is probably the result of marked overgrowth in the premaxillary-vomerine complex secondary to total lack of structural integrity in the region.

For comparison, Mars and Houston (1990) and da Silva Filho et al. (1998) described groups of adult unoperated patients with BCCLP and found extreme protrusion of the premaxilla and a very convex profile measured as the ANB angle. No measurements were performed to describe the position of the body of the maxilla. Da Silva-Filho et al. (1992a, 1998) also found the mandible to be short and retrognathic and discussed whether this finding was related to the primary anomaly or if it was caused by secondary functional adaptations.

The retrognathia of the basal part of the maxilla and the short and retrognathic mandible found in our sample are, in our opinion, variations intrinsically associated with the cleft of the secondary palate as discussed above.

9.3 Discussion and Conclusions

The Danish study of craniofacial morphology in untreated cleft infants is the hitherto most comprehensive and well-controlled since it covers a whole population, which is homogeneous and in

which central registration of clefts has been carried out for more than 65 years, a registration which has been shown to be highly reliable and nearly complete. Furthermore, all cleft infants are surgically treated at one hospital by one surgeon using the same techniques. All infants were examined with state-of-the-art three-projection cephalometry using the hitherto most comprehensive cephalometric analysis covering all craniofacial regions, and the methods were validated. The study included more than 600 children, and even after breakdown into subgroups, the sample sizes were adequate for statistical testing (except maybe for the RS group). Based on these facts, the findings related to the infant craniofacial morphology at 2 months of age, prior to any surgical or orthopedic treatment, must be considered to represent the "true" malformation, primarily caused by intrinsic factors.

In Table 9.1, the most important findings in the primary anomaly in the Danish infants with RS, ICP, BCCLP, and UCCLP are given, revealing a rather clear pattern. The findings support the suggestion of Dahl (1970) and others that facial clefts should be classified based on the embryonic facial development, i.e., into clefts involving the primary palate only (CL), clefts involving the secondary palate only (CP), and clefts involving structures of both the primary and the secondary palate (CLP). The postnatal facial morphology in these groups differs greatly. Infants with cleft of the secondary palate, with or without cleft of the primary palate, shared a number of characteristic morphological traits when compared to the norm: decreased posterior length of the maxilla, maxillary retrognathia, decreased posterior height of the maxilla, increased width of the maxilla and the nasal cavity, decreased length of the mandible, mandibular retrognathia, and reduced size of the pharyngeal airway. As seen from Table 9.1 and Fig. 9.3, the mandibular involvement was most pronounced in the RS group followed by the ICP and BCCLP groups and, finally, the UCCLP group. A similar pattern was observed for the reduced size of the pharyngeal airway.

As for the maxilla, the increased width of the maxilla and the nasal cavity was most pronounced

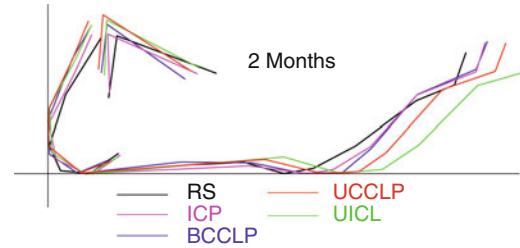

Fig. 9.3 Mean plots of the mandible in the RS, ICP, BCCLP, UCCLP, and UICL groups. Superimposition was made on the mandibular line (ML) registered at pogonion (pg)

in the groups with clefts of both the secondary and the primary palate, i.e., BCCLP and UCCLP. None of these groups showed decreased total length of the maxilla or retrognathia of the maxilla when measured to the premaxilla; the reason for this being a true and relative protrusion of the premaxilla, respectively.

In conclusion, a short and retrognathic mandible was a constant finding in infants with cleft of the secondary palate. The reduction in size of the pharyngeal airway in infants with cleft of the secondary palate was clearly related to the short and retrognathic mandible, being most severe in the RS group, which had the added effect of a reduction in the cranial base angle. But, in principle, all four groups had restricted upper airways as part of the primary anomaly. The increased width of the maxilla and nasal cavity was most pronounced in the groups which also had cleft of the primary palate (UCCLP and BCCLP). The UCCLP group was also characterized by relative protrusion of the premaxilla which was positioned asymmetrically, deviating to the noncleft side, whereas in the BCCLP group, the premaxilla showed true protrusion both in relation to the basal part of the maxilla (the lateral segments) and to the anterior cranial base. On average, the premaxilla was found to be positioned in the midline in this group, although most of the individual cases showed some degree of asymmetry. The protrusion of the premaxilla is suggested to be secondary to the primary anomaly of clefting, allowing for overgrowth in the premaxillary-vomerine complex, due to partial or total lack of anatomical integrity in the region.

It has been the aim of this chapter to summarize our findings about the intrinsic variations in facial morphology associated with the different types of cleft malformations to form a basis for valid estimations of the amount of surgical iatrogenesis, especially to the maxillary development, introduced by different surgical procedures and regimes, including the timing of treatment. In Fig. 9.4, the growth changes of the craniofacial skeleton from 2 to 22 months of age in the UCCLP group have been compared to the UICL group (control group) using color-coded surfaces on a 3D CT-model. In both groups, the cleft lip was surgically closed just after the examination at 2 months of age using a Tennison procedure. In the UCCLP group, the anterior part of the palate was closed with a vomer flap at the same time. The method of producing the illustrations will be given below.

9.3.1 Intuitive Visualization of the Location of Growth Differences

Cephalometric measurements in three projections provided growth vectors at each of the 279 (230 skeletal and 49 soft tissue) anatomical landmarks. The growth vectors, computed as the vector difference between corresponding landmark locations at the ages 2 and 22 months, respectively, after alignment to a common coordinate system (Hermann et al. 2000), have been used to form average growth patterns previously shown in Hermann et al. (1999a, b) (UICL, UCCLP) and Hermann et al. (2004) (UICL, BCCLP). Results of comparisons of growth between the UCCLP and the BCCLP groups, respectively, and the control group (UICL) have been shown as color-coded average growth patterns in Hermann et al. (1999a, b) (UCCLP vs. UICL) and Hermann et al. (2004) (BCCLP vs. UICL). These color-coded growth diagrams disclosed the locations of significantly different growth (1, 5, and 10 % levels) in the study group when compared to a reference group, and the diagrams were shown separately for each of the 3 projections (lateral, frontal, and axial), as well as for the growth magnitude and the two growth directions (x and y in each of the projections, respectively). In order to facilitate the effective comprehension of these diagrams, the locations of significant difference are color-coded onto the surface of a skull reconstructed from a CT scan of a single (noncleft) infant. As an example, Fig. 9.4 shows such color-coded surfaces for the comparison of the UCCLP with the UICL (control group). The color-coded surfaces were created by landmarking the 3D CT scan of the single noncleft infant at locations corresponding to the 230 skeletal cephalometric landmarks and color coding the surface in the vicinity of each landmark by a color corresponding to the significance of the growth difference. The landmark locations are shown in Fig. 9.5. A color table was chosen such that colors signify Student's *t*-test p values smaller than 0.01. Blue colors correspond to locations where the study group exhibits larger growth than the control group, while the opposite is the case at locations colored red. Regions without any significant differences between the two groups remained gray. In the UICL and UCCLP groups, the frontal and axial projection data were mirrored in order to have all clefts on the left side. Accordingly, the cleft is on the patient's left side in Fig. 9.4. The spatial extent of colored surface area in the vicinity of a landmark was governed by the distance to its closest landmark, and a maximum extent (spherically from landmark position) was chosen as 40 mm. Color-coded skulls are shown for differences in growth magnitude, as well as for each of the three growth directions (sagittal, vertical, and transverse). The colors for sagittal growth differences were computed from the x-component of the growth vectors in the lateral cephalometric projection and the y-component of the growth vectors in the axial projection. The colors for vertical growth differences were computed from the y-component of the growth vectors in the lateral projection and the y-component of the growth vectors in the frontal projection. The colors for transverse growth differences were computed from the x-component of the growth vectors in the frontal projection and the x-component of the growth vectors in the axial projection. The method of color coding has previously been described and applied for visualization of the

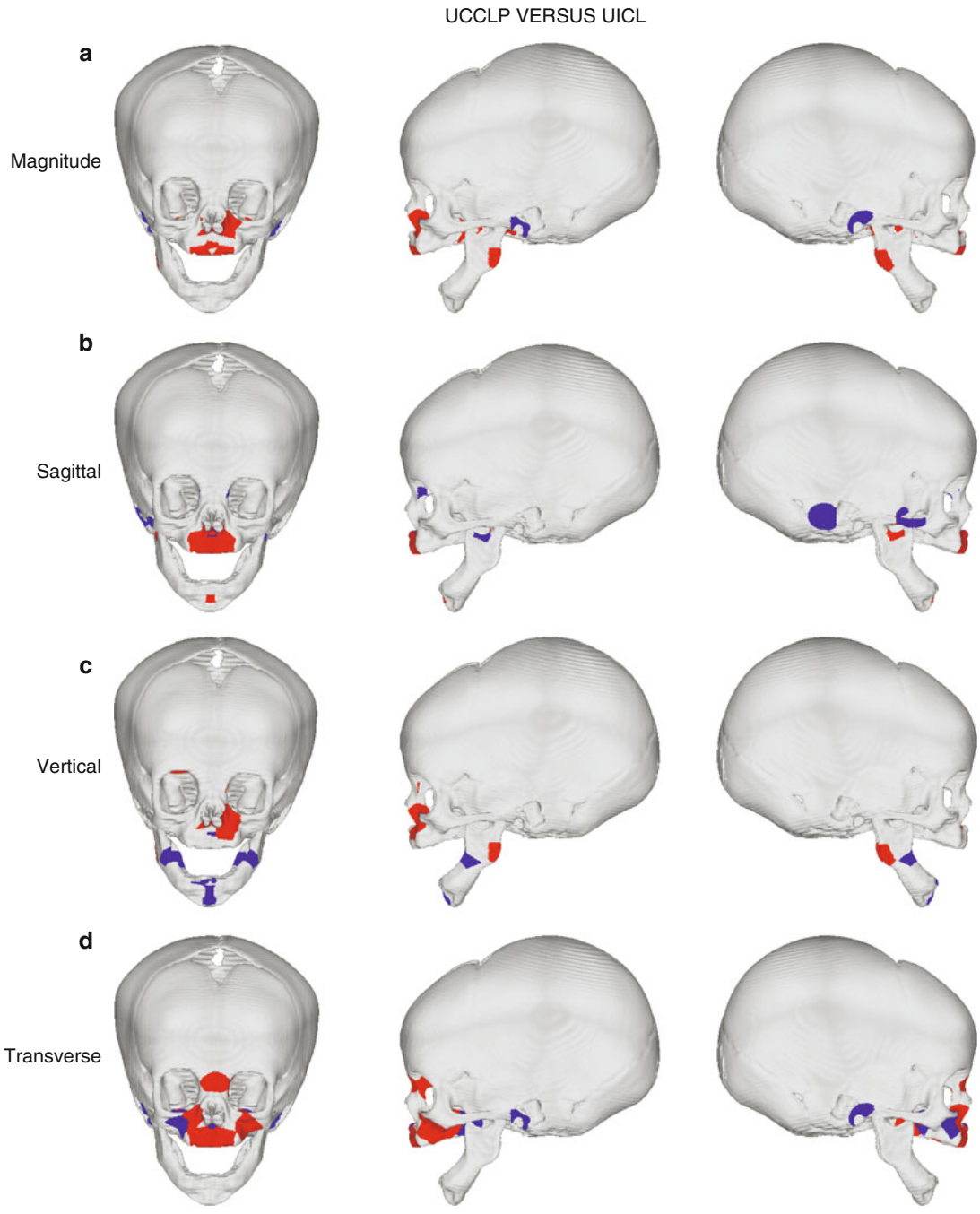

Fig. 9.4 (**a**–**d**) 3D visualization of locations of growth differences. Locations where UCCLP growth differs significantly ($p<0.01$) from UICL growth (2–22 months of age) are colored *red* (UCCLP<UICL) or *blue* (UCCLP>UICL). The surface reconstruction shown is of a noncleft subject of comparable age and is used solely for illustration. Cleft side is on patient's *left* in the figures. Locations of differences in the (**a**) magnitude of growth, (**b**) sagittal, (**c**) vertical, and (**d**) transverse growth components are shown

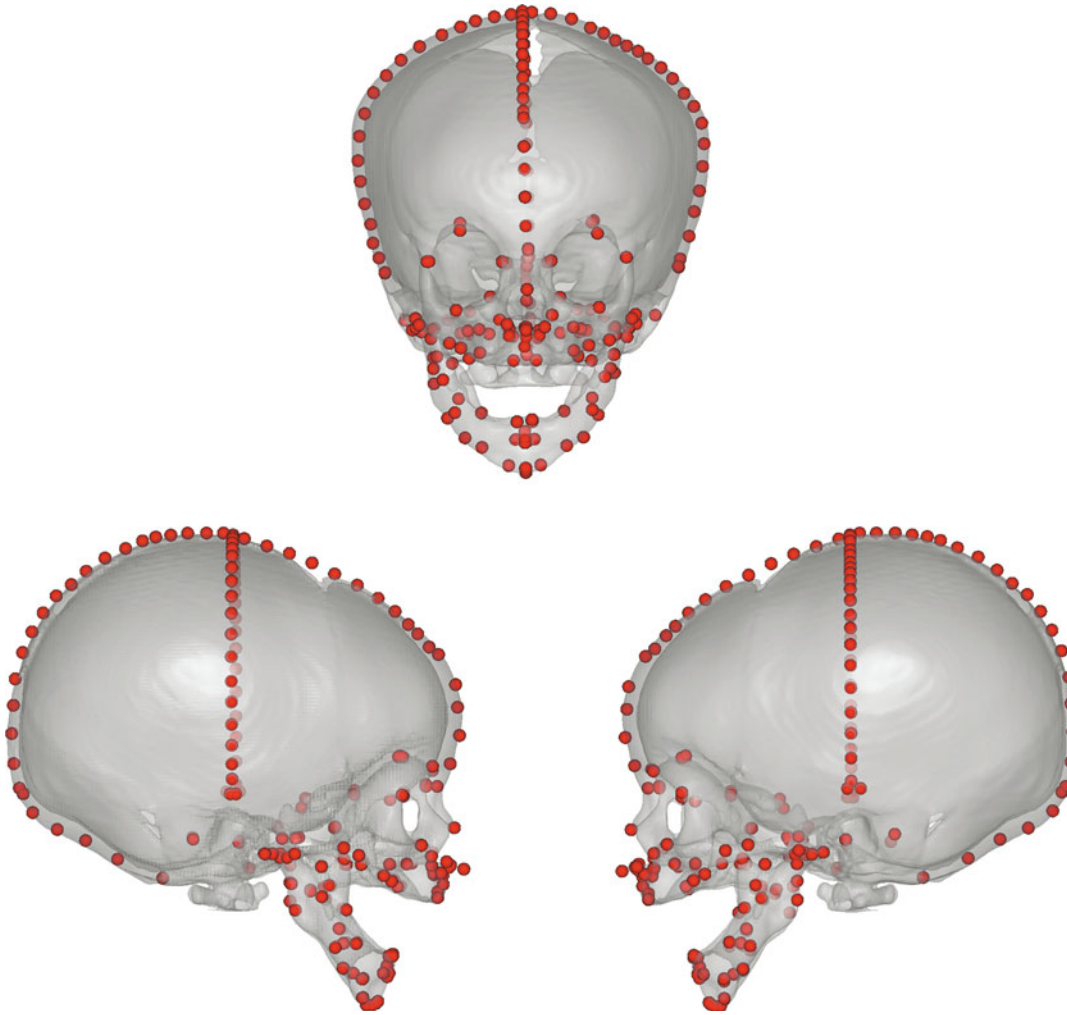

Fig. 9.5 3D landmark locations corresponding to the skeletal landmarks used in the three-projection cephalometric analysis as well as for creating the color-coded surfaces in Fig. 9.4

growth differences between UCCLP and UICL in Darvann et al. (1999).

Secondary to surgical closure of the lip at 2 months of age in the UCCLP group, we found that the premaxilla was molded into place, demasking the intrinsic maxillary retrognathia and leading to a normal sagittal jaw relationship at 22 months of age. Maxillary growth was, besides the premaxillary molding, characterized by smaller vertical growth on the cleft side and reduced transverse development, which could probably be related to the effects of surgery. The amount of mandibular growth was similar in the two groups. However, the direction of growth was slightly more vertical in the UCCLP group. This growth pattern was probably related to the intrinsic pattern of mandibular development. Otherwise, craniofacial growth seemed to be very similar in the two groups.

We found that surgery to the lip and anterior part of the hard palate at 2 months of age in UCCLP subjects seemed to influence the development of the maxillary complex, as observed at 22 months of age, in a number of beneficial ways: the premaxilla was no longer relatively protruding, and it was less asymmetric; the nasal septum deviated less toward the noncleft side; the width of the nasal cavity and the posterior

part of the maxilla became relatively more normal; and the transverse position of the lateral maxillary segment on the noncleft side was closer to normal. The posterior height of the maxilla was, however, still reduced to the same degree; the mandible was still short and retrognathic to the same degree; and bimaxillary retrognathia was still present. The only iatrogenic effect observed was that the lateral maxillary segment on the cleft side had become displaced toward the midsagittal plane anteriorly, resulting in a much too narrow dental arch at the level of the deciduous canine (Hermann et al. 2000). It is noteworthy that several studies of older, *unoperated* UCCLP children and adults find the maxillary prognathism to be within normal limits or even increased when compared to normative data (Ortiz-Monasterio et al. 1959, 1966; Mars and Houston 1990; Capalozzo et al. 1996). All these studies, however, only measure maxillary prognathism to the A-point or to the point ANS, both located in the relatively protruding premaxilla. Ortiz-Monasterio et al. (1959) concluded based on their findings in unoperated adults with UCCLP that: "The embryonic factor responsible for the facial cleft does not interfere with maxillary growth. This evidence leads us to believe that growth defects of the middle third of the face so frequently seen are caused by early or repeated and aggressive surgery." We disagree somewhat with this conclusion. Based on our studies of infants with UCCLP, it would seem that maxillary retrognathia in this group is part of the intrinsic variations associated with the cleft malformation of the secondary palate. In the unoperated infant and the unoperated adult, the maxillary retrognathia is, however, partly masked by relative protrusion of the premaxilla, secondary to overgrowth in the premaxillary-vomerine suture. Surgical closure of the lip at 2 months of age molds the premaxilla back into place, demasking the maxillary retrognathia. Thus in the 22-month-old lip-operated UCCLP group, it is our opinion that the bimaxillary retrognathia illustrates the facial type characteristic of the group rather than an iatrogenic effect of cleft surgery (Hermann et al. 1999b, 2000). Thus, we do not consider the maxillary retrognathia observed at 22 months of age as the result of surgical iatrogenesis; rather, we believe it represents a normalization of the "intrinsic facial type" characteristic of subjects with UCCLP; and at 22 months of age, the face is still harmonious with a normal sagittal jaw relationship. We have, at this point in time, not reexamined the sample at older ages and can, therefore, not comment on facial growth and signs.

In conclusion, we are not arguing that cleft surgery does not lead to disturbed maxillary development during the growth period. But we are suggesting that subjects with cleft of the secondary palate have a special "intrinsic" facial type, primarily characterized by bimaxillary retrognathia and increased maxillary width. We are speculating that this facial type could be a "liability factor" increasing the probability of CP or CLP (Hermann et al. 1999a, b). Finally, we suggest that when outcome of cleft surgery in CLP subjects is evaluated at adolescence or adulthood, comparisons should not be made to normal standards, but rather to the adolescent and adult morphology seen in CP subjects.

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A 25-Year Longitudinal Facial Growth Study of Unilateral Cleft Lip and Palate Subjects from the Sri Lankan Cleft Lip and Palate Project

10

Brijesh Patel and Michael Mars

10.1 Introduction and Background

The Sri Lankan Cleft Lip and Palate Project (SLCLPP) was founded in 1984 and completed data collection in 2009. Surgical visits were undertaken in 1985, 1986 and 1990. More than 500 surgical patients have been followed up longitudinally for 25 years post-operatively, resulting in the creation of a unique multidisciplinary archive. In total, 14 data collection visits were made from 1984 to 2009 (Mars et al. 2008).

Patients presented for surgery at all ages from infancy to adulthood. Many had received no surgery whatsoever; some had received lip surgery but not palatal surgery and others lip and palate surgery by local surgeons. They therefore provided a special opportunity to study the nature and timing of surgical intervention and its outcome on facial growth up to adulthood (nature's experiment).

This chapter is an analysis of the long-term outcome of patients who were over 20 years of age at their last data collection point.

10.1.1 Records Collected for Study

Standardised lateral skull radiographs and dental study models were collected for all subjects as part of this longitudinal study. Impressions were taken in alginate material with the patients sitting upright on a wooden chair.

10.2 Un-operated Unilateral Cleft Lip and Palate

Adults with un-operated clefts of the lip and palate provide the ideal group to study the natural progression of facial growth and to fully assess the inherent growth potential in these patients. Due to the ethical difficulties in withholding treatment for cleft patients, studies of un-operated cleft subjects have been undertaken in the developing world where surgery has not been readily available. As a result, most studies on such patients have lacked a suitably matched local comparison group as most operated patients presented from the developed world (Mars 1993).

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10.2.1 Clinical Features

The most striking feature in the un-operated UCLP patient is the protrusion of the upper labial segment (Fig. 10.1). These subjects present with large overjets, proclined upper incisors, eversion of the major segment, mild contraction of the lesser segment in the anterior region and rarely, buccal crossbites.

10.2.2 Cephalometry

Using the Sri Lankan growth archive, Liao and Mars (2005a) studied the long-term effects of clefts on craniofacial morphology in patients with UCLP. Employing a retrospective case–control study design, they compared 30 un-operated adult UCLP patients with 52 normal (non-cleft) control subjects of the same ethnic background. Cephalometric analysis confirmed the presence of morphological differences between UCLP and non-cleft patients (Fig. 10.2). The adverse effects of clefting were predominantly on the vertical development of the maxilla both anteriorly and posteriorly and to a lesser extent on the anteroposterior development of the basal maxilla. In addition, there were differences in the position and shape of the mandible and the position of the maxillary and mandibular incisors. However, the overall anteroposterior dimensions of the maxilla were not affected by clefting, and these patients did not exhibit maxillary retrusion.

10.2.3 Study Model Analysis

Using the reflex microscope, McCance et al. (1990) studied the maxillary arch form of 41 adults with un-operated complete unilateral cleft lip and palate and compared them to a control group of 100 normal adults (Fig. 10.3).

The teeth in the cleft group were smaller than their equivalents in the control group, the most marked difference being found in the central and lateral incisors. Arch widths of the cleft groups were reduced, more anteriorly (5 mm in the canine region) than posteriorly (1.6 mm in the second molar region), resulting in more V-shaped arches. No differences were found in the arch length or chord lengths between the groups. There was a higher prevalence of crossbites in the cleft group, 19.5 %, compared to none of the controls, and the overjet was greater in the cleft group (mean 8.2 mm) than in the controls (3.7 mm). A higher percentage of missing teeth, most commonly the lateral incisor teeth, was recorded in the cleft group. There was no difference in crowding between the two groups. Although the reductions in tooth size and arch width would suggest a small degree of primary hypoplasia, the differences are small.

The GOSLON yardstick is a robust and reproducible tool for categorising dental arch relationships into five distinct groups of increasing deformity (Mars and Plint 1985, Mars et al. 1987). It was applied to 51 un-operated UCLP cases, and the results showed 98 % of the cases were in groups 1 and 2 (excellent or very good arch relationships) and no cases in groups 4 or 5 (Fig. 10.4). In contrast, only a small proportion of operated patients were in group 1 when UK centres were assessed as part of the CSAG study (Clinical Standards Advisory Group 1998) (Fig. 10.5).

10.2.4 Summary

Studies using cephalometry and study models confirm that there is an intrinsic potential for un-operated UCLP patients to grow relatively normally with minor distortions around the cleft site itself where the dentition is unrestrained because of the disrupted musculature. In addition, the lack of continuity of the arch probably explains the transverse distortions seen in the dental arch. There is a small degree of hypoplasia as discussed above but these are minor and do not account for the gross maxillary retrusion frequently reported in surgically repaired patients.

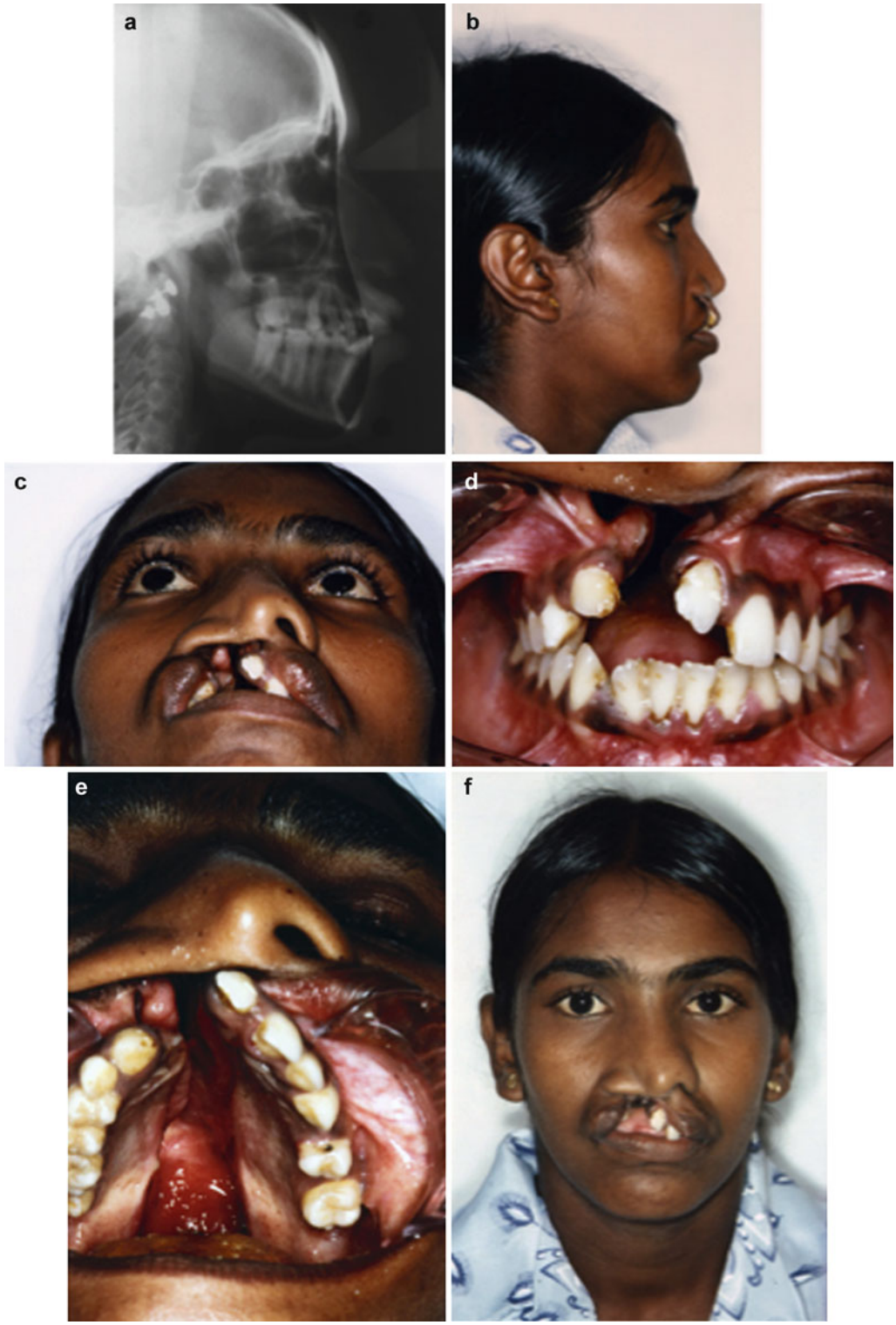
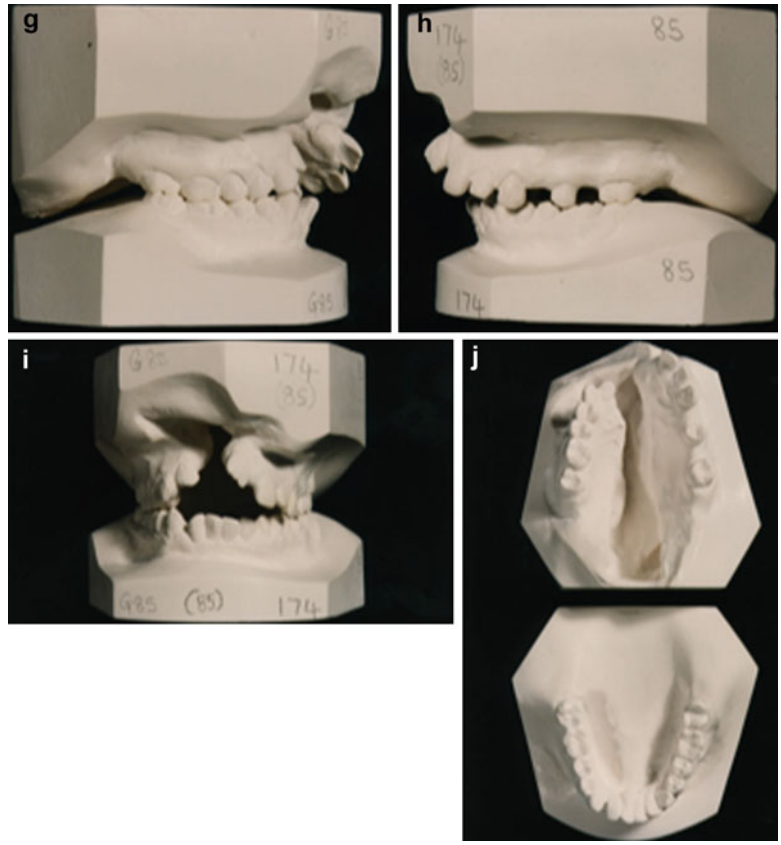


Fig. 10.1 (**a**–**j**) Illustrates a typical example of the facial appearance and dental study models of an un-operated unilateral cleft lip and palate case

Fig. 10.1 (*continued*)



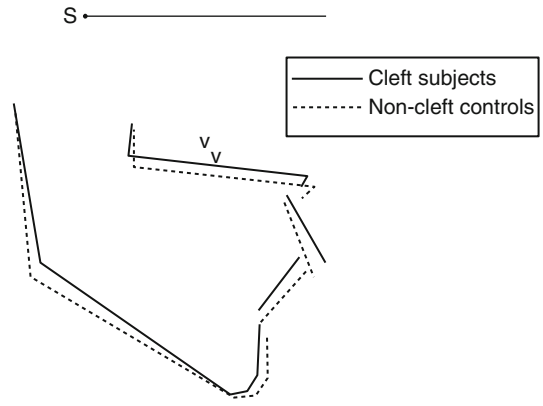


Fig. 10.2 Dental and skeletal effects of lip surgery (Liao and Mars 2005a)

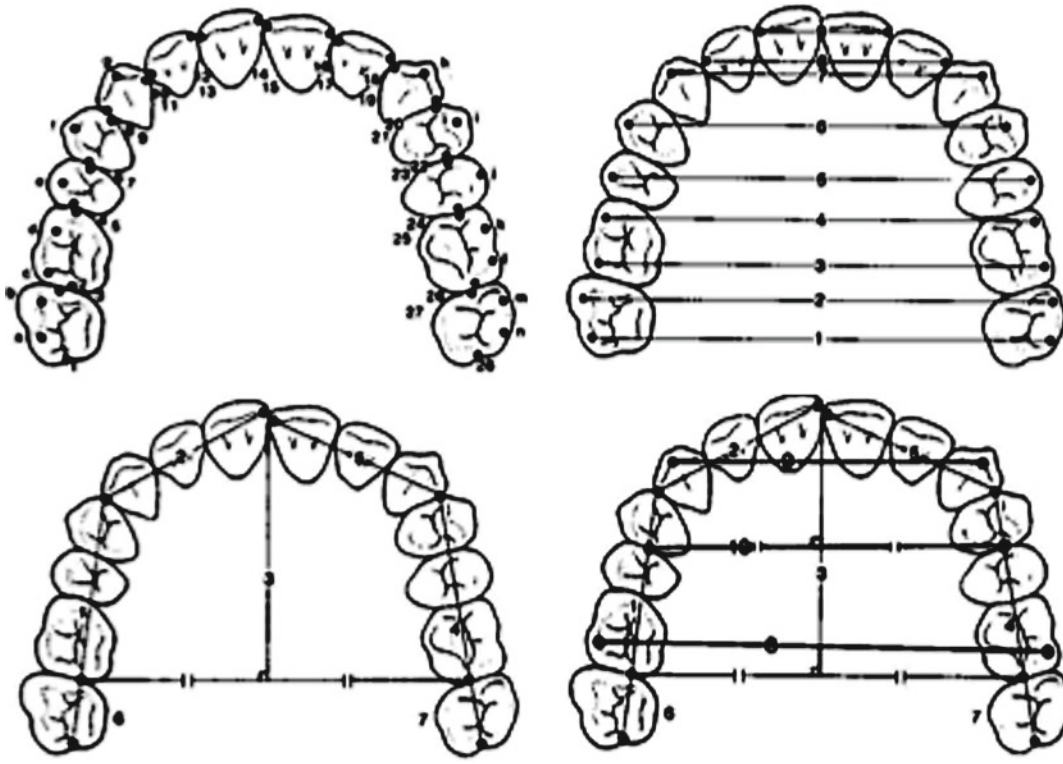


Fig. 10.3 The digitised points, chord lengths and arch widths used in the reflex microscopic analysis of study models

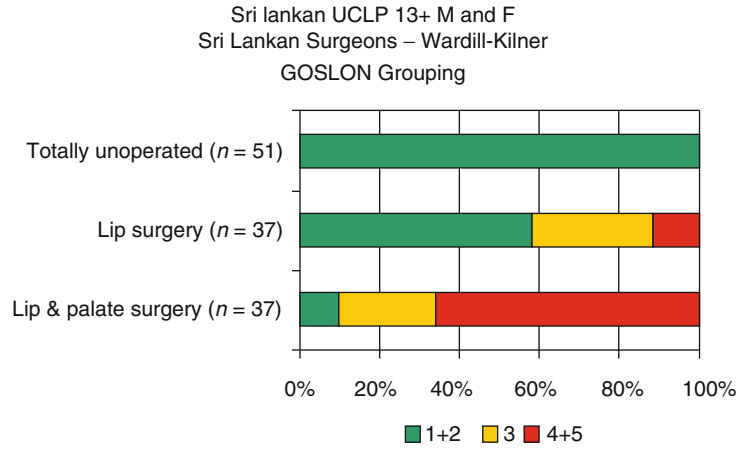


Fig. 10.4 GOSLON grouping of UCLP patients operated by Sri-Lankan surgeons using Wardill-Kilner (Mars and Houston 1990)

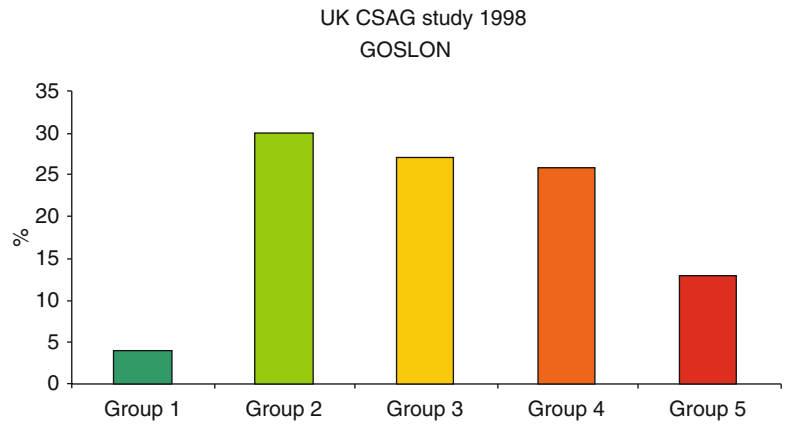


Fig. 10.5 GOSLON for the 1998 CSAG (Clinical Standards Advisory Group) for the whole of the UK

10.3 Effect of Primary Surgery on Facial Growth

It is widely accepted that facial growth and morphology in cleft lip and palate patients is abnormal, with mid-face retrusion common in patients who have had corrective surgery in infancy (Ross 1987; Semb 1991).

Historically, the cause of this mid-face retrusion has been attributed to three possible causes: an intrinsic developmental deficiency, functional distortions affecting growth and iatrogenic factors due to surgical treatment. There has been significant controversy on the extent and relative contributory role of each of these possible causative factors. It was this unresolved conflict of opinions on the aetiology of facial growth distortion in operated cleft lip and palate patients that led to the establishment of the SLCLPP.

A number of seminal papers on facial growth in cleft patients have been published from the Sri Lankan archives.

Mars and Houston (1990) reported the effects of primary lip and palate surgery on craniofacial growth in cleft patients using a cohort of Sri Lankan male patients. The studied patients were divided into three subgroups and compared to a control group of healthy males using lateral cephalometry and study model analysis. The three subgroups analysed included: those who had totally unrepaired cleft lip and palate, those who received lip repair in infancy but not palatal repair and those who had lip and palate repair in infancy. From this preliminary study, it was clearly evident that un-operated cleft patients had the potential to grow normally. Furthermore, in patients who have had a lip repair but no palate repair, the maxilla appears to also grow relatively normally.

In addition to cephalometry, Mars and Houston (1990) applied the GOSLON yardstick to the same cohort of patients aged 13 years old operated by Sri Lankan surgeons. Figure 10.4 shows that all the subjects in the totally un-operated subgroup had excellent dental arch relationships (groups 1 and 2). In the lip only subgroup, almost two thirds of the subject scored in groups 1 and 2, and only a small proportion scored in groups 4 and 5. This contrasts markedly with the lip and palate subgroup where two thirds of the patients were in groups 4 and 5 (poor dental arch relationship) and only a small proportion in groups 1 and 2.

Using preliminary data from the SLCLPP, Mars and Houston (1990) clearly showed the detrimental effects of primary palatal surgery on facial growth.

We now have complete data for 198 UCLP subjects who are greater than 20 years of age. This chapter studies the GOSLON yardstick analysis of all these patients and the cephalometric measures for 154 subjects.

10.3.1 Lip Surgery

Whilst studies on un-operated cleft patients have demonstrated the iatrogenic effects of primary

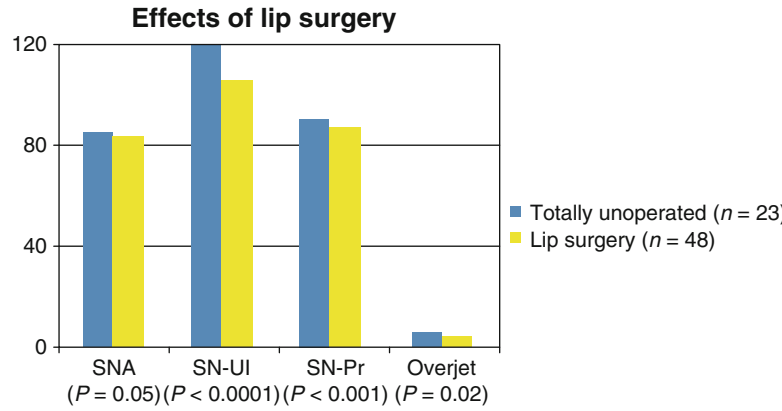


Fig. 10.6 Effects of Lip Surgery (Liao and Mars 2005b)

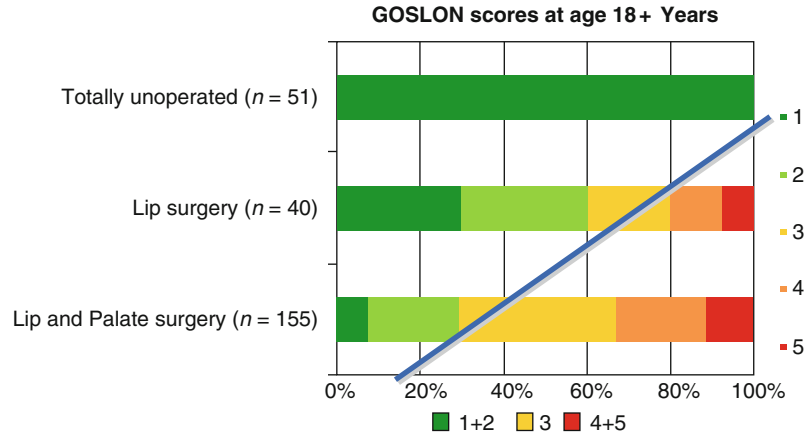


Fig. 10.7 The diagonal line demonstrates the progressively worsening effect from no surgery to lip surgery to lip and palate surgery

cleft surgery on facial growth, there has been controversy as to whether the lip or the palate repair has the most detrimental effect on maxillary growth (Muir 1986; Mars and Houston 1990; Bardach et al. 1984).

10.3.1.1 Cephalometry

Liao and Mars (2005b) further clarified the long-term effects of lip surgery on craniofacial growth. Using lateral cephalograms from the longitudinal growth data obtained from the SLCLPP, they studied 71 adult patients, 23 non-syndromic unoperated UCLP patients and 48 non-sydromic UCLP patients who had undergone lip repair only. This study demonstrated that the major effect of the lip repair was on the anteroposterior and vertical position of the maxillary alveolus and the maxillary incisors. Furthermore, there was a differential influence from the tip of the alveolus and the incisal edge to the base of the alveolus and the incisal apex. This was associated with uprighting of the maxillary incisor and resulted in a decreased overjet and increased overbite in the lip repair only group (Fig. 10.6). The pressure of the lip thus produces secondary bone resorption in the base of the anterior maxillary alveolus.

10.3.1.2 Study Model Analysis

More recently, the authors carried out a review of the SLCLPP archive using the GOSLON yardstick. All patients were operated within the project and are now adults aged over 20 years and have completed facial growth. The subjects were divided into the same three subgroups as reported by Mars and Houston (1990) and had had their surgery at varying ages. Interestingly, the same results were replicated. Figure 10.7 below is almost a replica of Fig. 10.4 above.

Both GOSLON figures (Figs. 10.4 and 10.7) clearly demonstrate that surgery to the lip only has

some effect, but this is mainly dentoalveolar. The most obvious difference is seen in the patients who have undergone both lip and palate repair, clearly suggesting that the palate repair has the most significant influence on future facial growth.

Muthusamy (1998) analysed study models of 26 patients pre and post lip repair using the reflex microscope. These patients had not had any other surgical interventions and were operated on by British surgeons as part of the SLCLPP.

The patients were divided into two subgroups – young being those who had the lip repair prepubertally (15 patients) and mature being those who had it post-pubertally (11 patients).

He found that in both groups, there was a significant reduction in arch width with the greatest reduction in the inter-canine distance. Interestingly, in the younger group, there was a reduction in arch width in all measures (anterior and posterior), whereas in the mature group, there was no significant reduction in the inter-molar and inter-premolar.

After the lip repair, there was a reduction in overjet and an increase in overbite in both groups (Fig. 10.8).

10.3.1.3 Summary

Both the cephalometric and study model studies confirm that that lip repair primarily produces a localised bone-bending effect on the anterior maxillary alveolus (alveolar moulding) and does not have a significant effect on maxillary growth.

10.3.2 Palate Surgery

Gillies and Fry (1921) were the first to suggest that the palate repair has a detrimental effect on facial growth. Interestingly, they were also the pioneers for the policy of delayed hard palate closure. The SLCLPP has been key in demonstrating the relative effects of lip and palate surgery on future facial growth.

10.3.2.1 Cephalometry

Liao and Mars (2005c) looked at the long-term effects of palatal surgery on facial growth. They compared non-syndromic UCLP Sri Lankan adults who had had either a lip repair only (48 patients) or a lip and palate repair (58 patients) using cephalometry. They concluded that palate repair inhibits the forward displacement of the basal maxilla and anteroposterior development of the maxillary alveolus in patients with UCLP. However, they found that the palate repair did not have any detrimental effects on the downward displacement of the basal maxilla or on palatal remodelling in patients with a unilateral cleft lip and palate. Contrary to expectations, the axial inclination of the maxillary incisors is not affected by the palate repair, though is a major effect of the lip repair as discussed above.

10.3.2.2 Study Model Analysis

Both GOSLON charts (Figs. 10.4 and 10.7) above show that the most obvious difference is seen in the patients who have undergone both lip and palate repair, clearly suggesting that the palate repair has the most significant influence on future facial growth.

10.4 Timing of Primary Surgery and Its Effects on Facial Growth

Because patients presented at all ages for primary surgery, it became possible to analyse the effects of such surgery when performed at any age between infancy and adulthood.

10.4.1 Timing of Lip Surgery

In another cephalometric study, Liao and Mars (2006) looked at the timing of lip repair and the relevance of the operating surgeon. Although the sample size was small (23 in the early repair group and 25 in the mature repair group), they noted that early lip repair was found to produce a greater bone remodelling effect in the base of the anterior maxilla. This is possibly related to the relatively greater surgical trauma in a smaller individual or the early onset on tension from the repaired lip or both. In this study, dentofacial morphology was unrelated to the surgeon who performed the lip repair.

Fig. 10.8 An adult patient who had a lip repair but no palate repair demonstrating the localised effects of lip surgery

Fig. 10.9 Effect of timing of hard palate repair on maxillary growth (Liao et al. 2006)

Timing of hard palate repair

Dependant veriable	Regression coefficient (95% CI)	*P* Value
PMP – A point (mm) Maxillary length	0.2 (0.0,0.4)	0.05
SNA (°) Maxillary protrusion	0.4 (0.2,0.7)	<0.001
ANB (°) Relative maxillary/ Mandibular protrusion	0.4 (0.2,0.6)	0.001

The regression coefficient indicates the change in mean (mm or degrees) of the dependant variable at 20 years of age per year increase at hard palate repair.

In summary, patients with earlier lip repairs display more localised bone remodelling than those operated upon later. As the lip repair only has a localised effect, it probably explains why dentofacial morphology is unrelated to the surgeon carrying out the lip repair.

10.4.2 Timing of Palatal Surgery

As the SLCLPP archive has patients operated on at different ages, it has provided an opportunity to undertake longitudinal retrospective studies to assess the question of the timing of hard palate surgery and its effects on facial growth.

10.4.2.1 Cephalometry

Liao et al. (2006) analysed the longitudinal records for 104 patients non-syndromic UCLP patients who had had their hard palate repair by the age of 13 years. A total of 290 lateral cephalograms taken at different ages were analysed using a linear regression model. The timing of hard palate surgery had a significant effect on the length and protrusion of the maxillary alveolus and the anteroposterior jaw relation measured at 20 years. The regression model suggested a maxillary growth improvement of 0.2mm in maxillary length and 0.4° in both SNA and ANB for every year delay in hard palate closure – Figs. 10.9, 10.10, 10.11, and 10.12. They concluded that the timing of hard palate repair significantly affects the growth of the maxilla, earlier palate repair has a more adverse effect on the growth of the maxilla.

Following the most recent data collection visit in 2009, the authors reviewed cephalograms of a further 50 non-syndromic UCLP adult patients, all aged over 20 years of age. Cephalometric analysis to assess the effects of the timing of palatal repair on facial growth concurred with the earlier study (Figs. 10.13, 10.14, and 10.15). Patients who underwent hard palate surgery in infancy showed reduced maxillary length and protrusion when compared to patients who underwent surgery at a later age. Patients who undergo surgery according to accepted protocols in infancy exhibited more class III malocclusions.

10.4.2.2 Study Model Analysis

The authors have recently reviewed all patients treated in Sri Lanka who had records at age greater than 20 years. One hundred and ninety-eight patients were divided up according to the age of palatal repair and the GOSLON yardstick was used to assess the dental arch relationships. The scores clearly show that the earlier the repair, the more the detrimental affect it has on facial growth in the long term (Figs. 10.16 and 10.17).

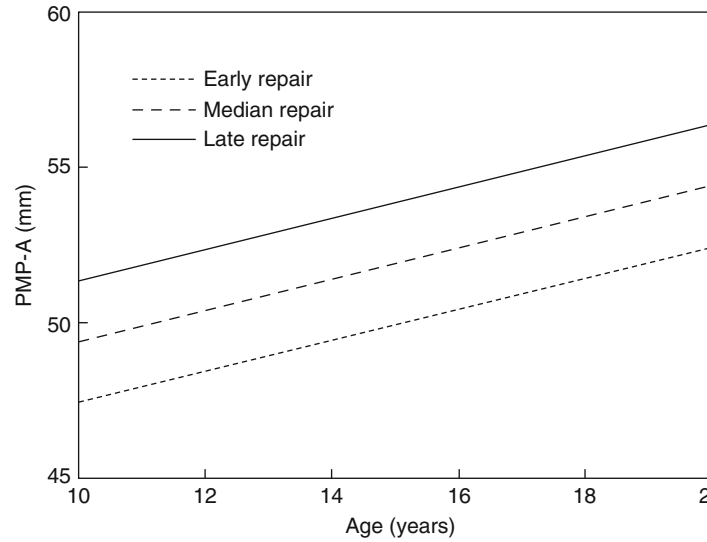


Fig. 10.10 Effect of timing of hard palate repair on palatal length (Liao et al. 2006)

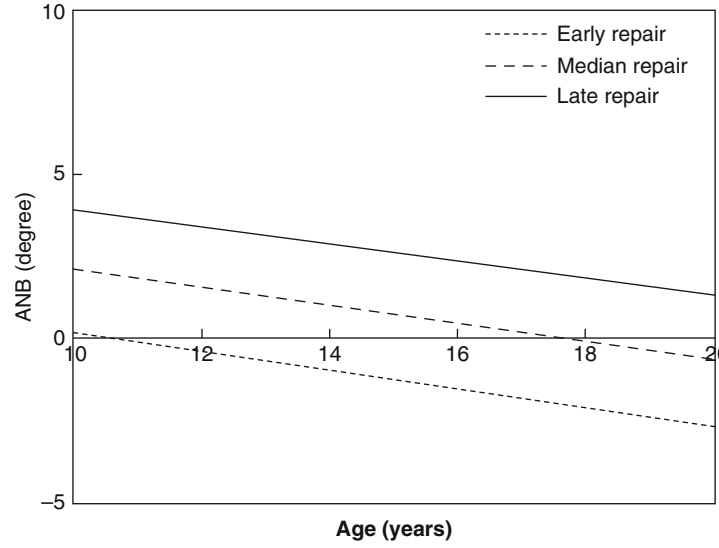


Fig. 10.11 Effect of timing of hard palate repair on maxillary-mandibular relationship (Liao et al. 2006)

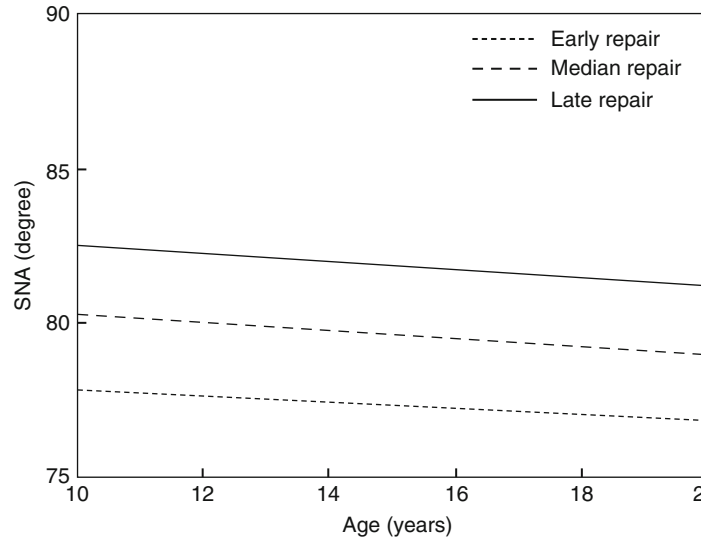


Fig. 10.12 Effect of timing of hard palate repair on maxillary protrusion (Liao et al. 2006)

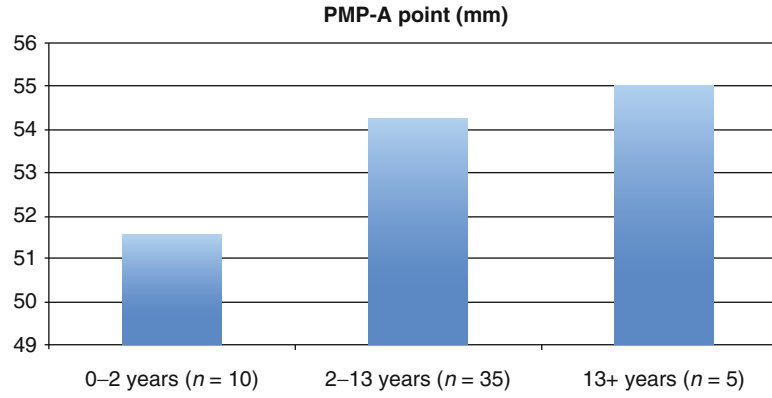


Fig. 10.13 Maxillary length related to age at primary surgery

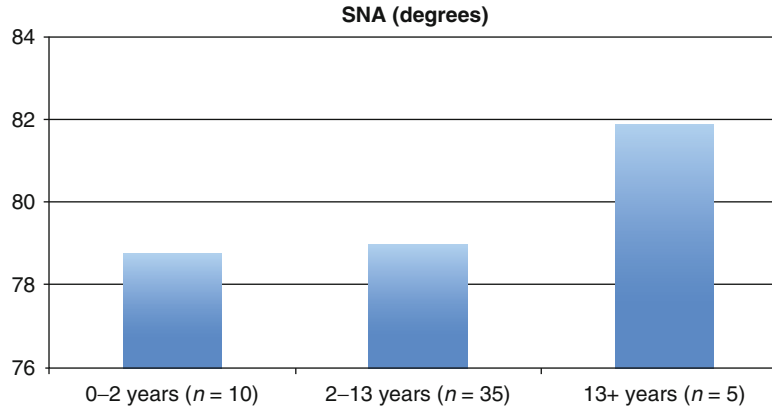


Fig. 10.14 Maxillary protrusion related to age at primary surgery

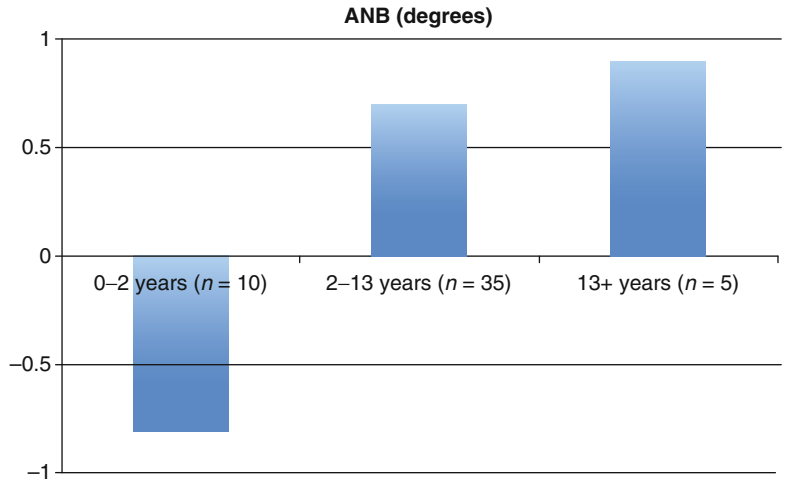


Fig. 10.15 Relative maxillary/mandibular protrusion related to age at primary surgery

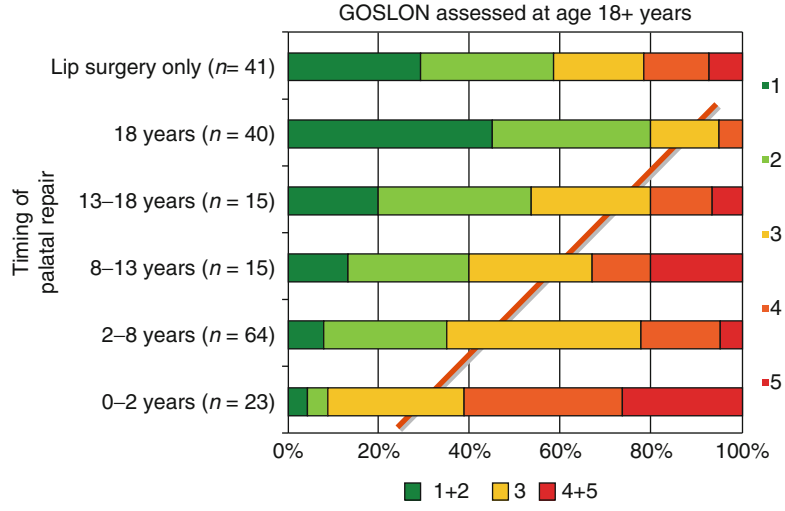


Fig. 10.16 Dental arch relationships and timing of palatal repair. The diagonal line demonstrates the progressively worsening effect with earlier surgery

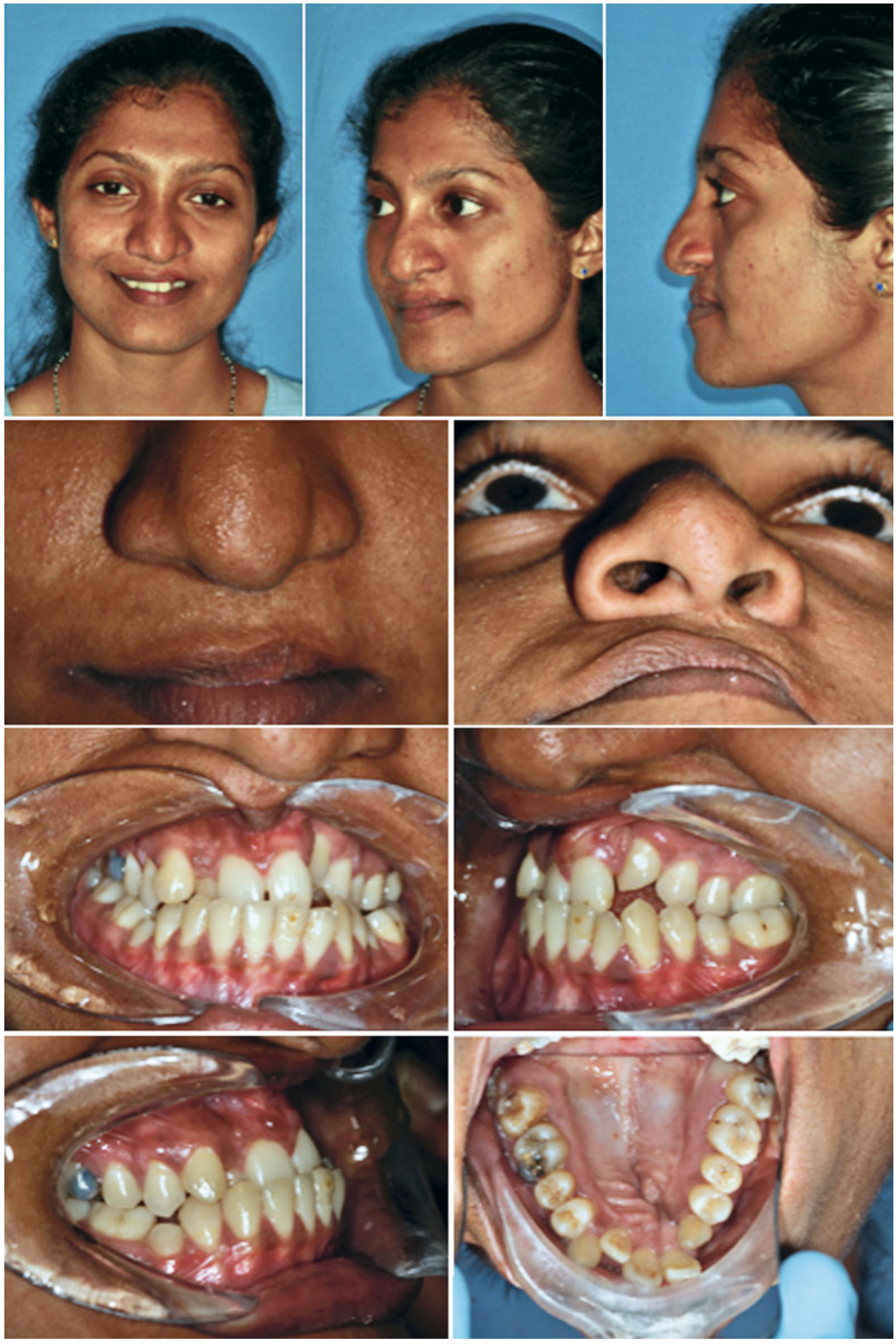


Fig. 10.17 Adjust subject who had lip and palate repair in early infancy demonstrating maxillary retrusion

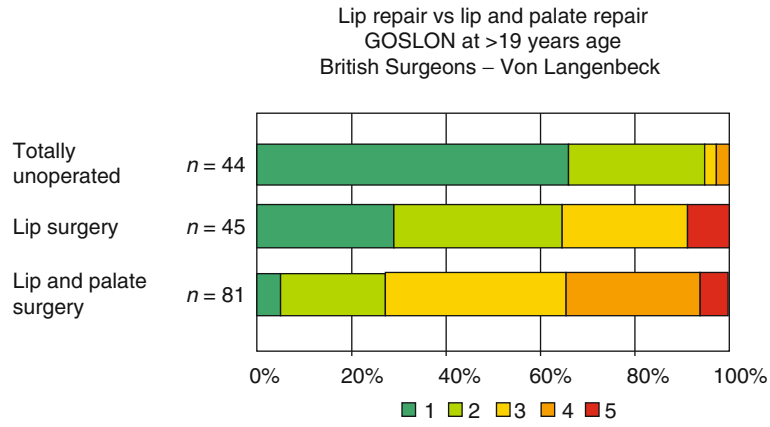


Fig. 10.18 GOSLON scores for adult patients operated by British Surgeons using the Von-Langenbeck procedure (Mars et al. 2005)

10.5 Type of Primary Surgery and Its Effect on Facial Growth

Studies from the SLCLPP data have clearly demonstrated the effects lip and palate surgery on facial growth. The data has been further analysed to assess any correlations on the type of surgery and its effects on long-term facial growth. The results have to be taken in context as subdivision of groups reduces numbers and the power of the study. However, the trends noted can form the basis for future randomised control trials.

10.5.1 Wardill-Kilner Versus Von Langenbeck Repair

There is no agreement on the best surgical approach. Both Pigott et al. (2002) and Johnston et al. (2004) reported increased proportions of poor or very poor GOSLON scores for a Veau-Wardill-Kilner repair group compared to a von Langenbeck group. The SLCLPP archive has patients who had had either the Wardill-Kilner or von Langenbeck procedures. The local Sri Lankan surgeons and some of the earlier visiting British surgeons favoured the Wardill-Kilner procedure. The Scandinavian surgeons and some of the later visiting British surgeons favoured the von Langenbeck procedure. The GOSLON yardstick used to assess outcome clearly shows that patients who undergo the Wardill-Kilner repair (Fig. 10.4) have poorer outcomes when compared to the von Langenbeck group (Fig. 10.18). These results do not take the skill of the surgeon into account, which clearly makes a difference.

10.5.2 Vomerine Flap

The use of a vomer flap has been another area of controversy. It was popularised by the Oslo team who have reported good long-term outcomes with their protocol (Semb 1991). Some groups believe that the use of a vomer flap is actually detrimental to future maxillary growth (Friede and Lilja 1994). Some surgeons within the SLCLPP routinely performed vomer flaps, whilst others did not. It has therefore been possible to tease out the effects of the use of vomerine flaps on dentoalveolar relationships in the long term.

The authors looked at adult UCLP patients who had only had a lip repair (31 patients) who were subdivided into two groups – lip with vomer flap and lip only. The GOSLON yardstick was used to assess outcomes depending on what age the lip surgery was carried out (Fig. 10.19). In both groups, surgery after 2 years of age did not have a marked effect on dentoalveolar relationships. In both groups, patients undergoing surgery in infancy had poorer GOSLON scores with more patients scoring 4 and 5 in the vomer flap group. This suggests that the vomer flap

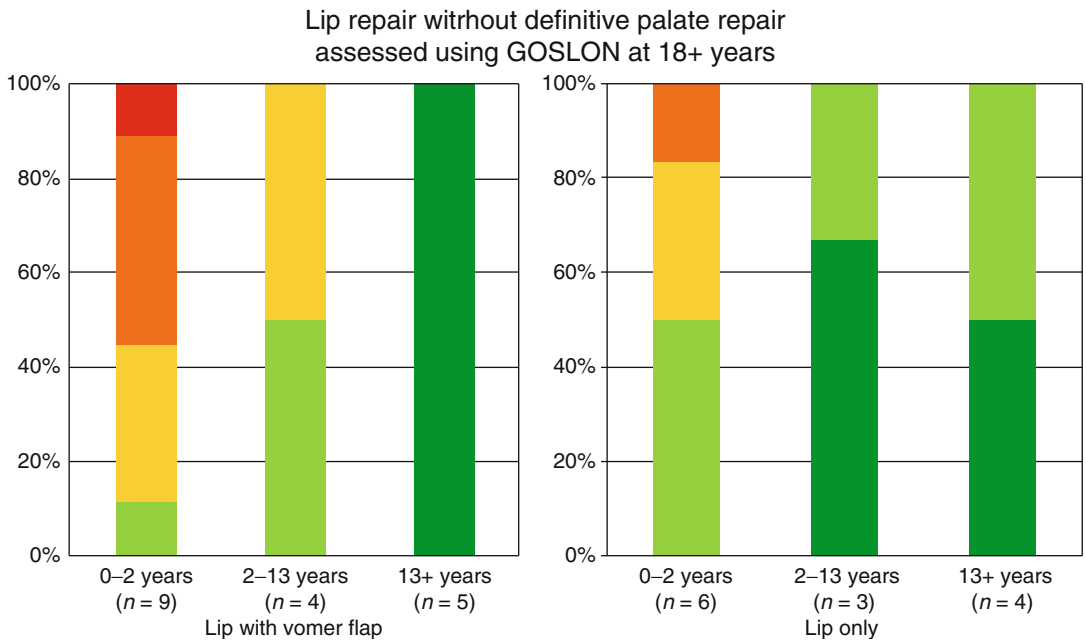


Fig. 10.19 GOSLON at age 18+ years for patients who had lip repair without definitive palate repair

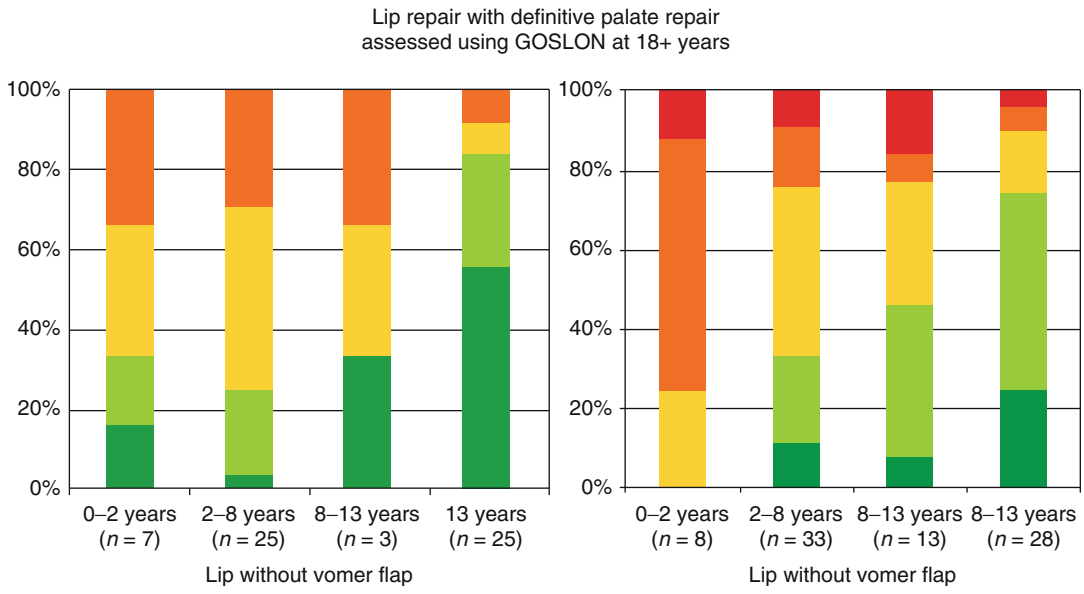


Fig. 10.20 GOSLON at age 18+ years for patients who had lip repair with definitive palate repair

may have some detrimental effect on the growth of the maxilla when assessed without the effects of hard palate repair.

Similarly, the authors analysed adult UCLP patients in the SLCLPP archive who had undergone both lip and palate surgery and subdivided these into two groups: lip with vomer flap (60 patients) and lip without vomer flap (82 patients). All patients underwent a von Langenbeck procedure to repair the palate. The GOSLON yardstick was used to assess outcomes depending on what age the lip surgery was carried out. The figure below (Fig. 10.20) clearly shows that patients who had a vomer

flap repair did better and did not score 5 in any age group. In both groups, patients who underwent repair after the age of 2 did better than the younger patients in the long term. The use of a vomer flap provides 'extra' tissue to repair the hard palate and limits the use of extensive undermining of the soft tissues and relieving incisions. As discussed above, the palate repair has the most detrimental effect on long-term facial growth, and the use of a vomer flap in patients who undergo both lip and palate repair appears to be beneficial.

10.6 Factors Influencing Interpretation of Results from the Sri Lankan Cleft Lip and Palate Project

10.6.1 Malnutrition and Growth and Delayed Onset of Puberty

It should be recognised that the above studies are derived from subjects in the developing world. Although Sri Lanka is a relatively advanced developing country, there is nevertheless significant malnutrition and endemic infections, for example, malaria. The failure of infants with clefts to gain weight adequately has been documented by several authors (Avedian and Ruberg 1980; Ranalli and Mazaheri 1975).

Malnutrition is a well-recognised form of reversible growth hormone resistance, which can be normalised with nutritional supplements. A malnourished mother is likely to give birth to a baby with low birth weight, while children with protein-energy malnutrition do not grow as well as others according to a recent report (Fernando 1990). This kind of malnutrition is an underlying cause of almost one third of the deaths among children under 5 years in Sri Lanka. Malnutrition is still a serious problem in Sri Lanka (Rajapaksha and Siriwardena 2002). Food insecurity is one of the major reasons for malnutrition in that country according to the Department of Census and Statistics. Poor financial and physical access to food is responsible for the malnutrition and food insecurity. Drastic price increases of essential food commodities and stagnating or deteriorating incomes create poor financial access to food. The civil war from 1984 to 2010 in Sri Lanka has exacerbated the provision of essential food and has created financial problems.

A recent survey of 16,000 Sri Lankan children found that only one quarter were properly nourished (Popham 2002). More than one third were suffering from third-degree malnutrition, the level beyond which children exhibit distended stomachs and skinny frames. Supporting evidence from the National Peace Council indicated that only 4,863 children under 5 years out of a random sample of 16,767 were within normal nutritional limits; 6,371 children had third-degree malnutrition, 3,186 with second-degree malnutrition and 2,347 with first-degree malnutrition (National Peace Council of Sri Lanka 1998). According to this report, diseases such as malaria cause malnutrition first, which is still prevalent in Sri Lanka. Secondary causes of malnutrition are by worm infestations and third by a lack of food. Many of the subjects in this study were social outcasts, who dropped out of school. Females in particular were hidden away in their houses, and only one female in the un-operated population married. Children need a good emotional climate to thrive. The mechanism of the effects of emotional deprivation on growth is not well documented but is linked to reduced growth hormone secretion and its associated growth failure.

It has been recognised in the context of growth studies in general and facial growth studies in particular that many patients in the developing world have delayed onset of puberty (Mars 1993). Boys may not attain maturity until after 20 years of age and girls until after 18 years. This has important implications for all studies in developing countries and failure to address this issue can seriously confound the result of research. The large volume of longitudinal data has enabled this problem to be addressed (Liao and Mars 2006).

10.6.2 Speech Implications

Whilst facial growth in the un-operated subject presents without maxillary retrusion – unlike many operated patients – the speech outcomes for the same series of patients demonstrate almost unintelligible speech for the whole sample.

Research on the Sri Lankan Cleft Lip and Palate archive has demonstrated that surgery when delayed beyond eight years of age and even earlier results in permanent irremediable speech disorders (Sell and Grunwell 1990; Sell 1991).

This chapter is careful in not recommending the delay of hard palate repair. Previous studies have consistently demonstrated speech impairment associated with delayed hard palate repair (Bardach et al. 1984; Witzel et al. 1984; Noordhoff et al. 1987; Rohrich et al. 1996, Lohmander-Agerskov 1988).

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Airway Management in Patients with Robin Sequence

11

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Abbreviations

CNMC	Children's National Medical Center
CPT	Current Procedure Terminology
ICD – 9	International Classifications of Diseases Version 9
LOS	Length of hospital stay
NPA	Nasopharyngeal airway
RS	Robin sequence
SPSS	Statistical Program for Social Sciences
TLA	Tongue-lip adhesion

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11.1 Introduction

Robin sequence (RS), the clinical triad of micrognathia (small jaw), glossoptosis (downwardly displaced tongue), and upper airway obstruction, affects approximately 1 in 8,500 births (Bush and Williams 1983; Sadewitz 1992). Cleft palate is also noted in up to 90.4 % of patients (Caouette-Laberge et al. 1994). Infants may present with a wide phenotypic variability, ranging from infrequent episodes of airway obstruction and/or feeding difficulty to severe crises of asphyxia and failure to thrive (Marques et al. 2001). The latter group of patients is at particular risk for hypoxic brain damage, impaired mental development, pulmonary hypertension, aspiration pneumonia, and failure to thrive (Tomaski et al. 1995; Hoffman et al. 1965; Kapp-Simon and Krueckeberg 2000). Despite advances in critical care medicine, mortality is not inconsequential and ranges from 0 to 13.6 % (Sadewitz 1992; Marques et al. 2001; Caouette-Laberge et al. 1994; Cruz et al. 1999; Dykes et al. 1985). Such high morbidity and mortality have been attributed to late diagnosis, delayed airway protection, and multisystem disorder. Furthermore, care of infants with RS can require prolonged hospitalization, with averages ranging from 10 to 60 days and translate into increased costs (Bull et al. 1990; Cruz et al. 1999; Matsas et al. 2004; Wagener et al. 2003).

Several modalities have been proposed to address airway obstruction in infants with RS. Options range from conservative management, namely, prone positioning, nasopharyngeal airway (NPA) placement, and orthopedic devices to

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operative interventions, including subperiosteal release of floor of mouth, tongue-lip adhesion (TLA), tracheostomy, and mandibular distraction osteogenesis (lengthening of the mandible).

The decision on how to manage these infants is often based on the experiences of the provider and the practices at a particular center. Therefore, it is not surprising to see the management of such infants continues to be controversial. Prior studies have attempted to establish evidence-based parameters to help clinicians to devise management plans for these infants. Typically, the management of infants with RS focuses on avoiding tracheostomy either conservatively or surgically, providing adequate respiration and nutrition and preventing long-term sequelae and death. However, by bypassing the obstruction, tracheostomy remains the most definitive treatment for infants with severe respiratory obstruction that is not compatible with life.

Although it is known that tracheostomy in these infants is a long-term commitment with a reported average age of 3.1 years at decannulation (Tomaski et al. 1995; Moyson 1961; Sadewitz 1992), studies on the chronology of decannulation in patients with RS are lacking overall. It is still unclear whether natural "mandibular growth" and time allow for decannulation in patients without further intervention can occur. As part of an overall institutional effort to establish evidence-based guidelines for surgical intervention in patients with RS, we present here a cohort of patients with severe upper airway obstruction treated with tracheostomy to determine length of time for decannulation without further surgical intervention (i.e., "natural" decannulation) and to investigate potential factors associated with successful "natural" decannulation.

11.2 Methods

The records were reviewed for patients who underwent primary airway management at Children's National Medical Center (CNMC) from 1994 to 2010. All study subjects were identified within the accounting departments of *Otolaryngology* and *Plastic and Reconstructive Surgery* using International Classifications of Diseases Version 9 (ICD-9 524.00, 524.06, and 524.10) related to diagnosis of anomaly of jaw size and Current Procedure Terminology (CPT 41510, 20690/20692, and 31600/31603/31605) which include tongue-lip adhesion (TLA), mandibular distraction, and tracheostomy.

Inclusion criteria were patients with documented micrognathia, glossoptosis, and respiratory obstruction who have received a tracheostomy at Children's National Medical Center. The following children were excluded: (a) respiratory difficulties other than upper airway obstruction, (b) patients who were treated at CNMC but with missing or incomplete medical records, and (c) patients of CNMC but who have received definitive airway management at another hospital.

Within the subset of the patients who have received tracheostomy, their demographics, nutritional and respiratory status, laboratory values, and polysomnographic (sleep) studies were reviewed. Perioperative and postoperative complications include tracheitis (inflammation of the trachea), pneumonia, breakdown, stoma infection, hematoma, reoperation, and finally long-term outcomes such as developmental delay, organ systems dysfunction (neurogenic, gastrointestinal, and/or cardiopulmonary), and death were recorded. Finally, the timing of events was investigated. Length of hospital stay (LOS) subdivided into pre-tracheostomy, postoperative hospital stay, and total length of hospital stay was noted. The duration of tracheostomy to "natural" decannulation was also recorded.

Data were analyzed with Statistical Program for Social Sciences (SPSS) version 16.0 software (Chicago, IL) and Microsoft Excel version 2008 software (Redmond, WA). Univariate analysis included chi-squared and Fisher's exact tests for contingency data. Kaplan–Meier curves with log-rank (Mantel–Cox) test were used to estimate the percent of patients on tracheostomy as a function of a time.

11.3 Results

Of 61 infants with RS, 25 infants received a tracheostomy. The other 36 infants were managed with lateral/prone positioning, noninvasive

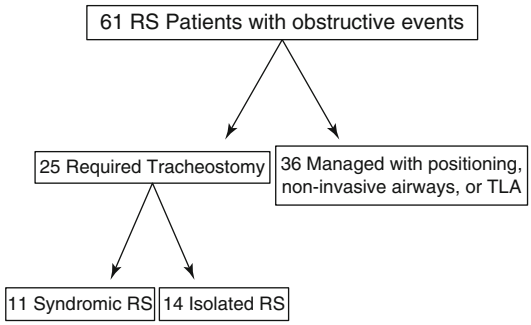

Fig. 11.1 Of 61 patients with obstructive events, 25 required tracheostomy, of which 11 were syndromic (Stickler syndrome (a group of genetic disorders affecting connective tissue), Treacher Collins (congenital craniofacial syndrome), and others) and other 14 patients had isolated Robin sequence

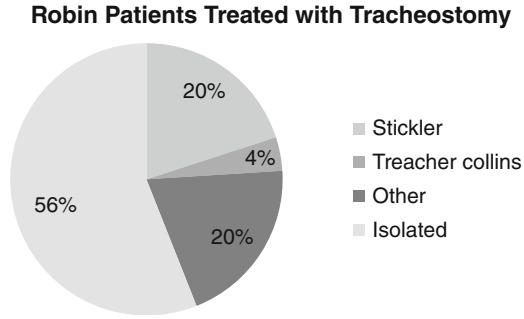

Fig. 11.2 Of 25 patients treated with tracheostomy, 44 % (11 cases) were syndromic, of which 45 % (5 cases) was either Stickler or other syndromes and 10 % (1 case) was Treacher Collins and the other 56 % (14 cases) had isolated RS

oxygen supplement, or TLA in Fig. 11.1. Among the cohort requiring tracheostomy, 14 patients (56 %) were isolated and 11 patients (44 %) were syndromic as shown in Fig. 11.2. Overall, the median time to decannulation was 97 months in Fig. 11.3. The few patients with syndromic RS who were successfully decannulated required a median time >73 months as compared to patients with isolated RS who had a median time to decannulation of 19 months in Fig. 11.4. In total, 13 out of 25 infants (52 %) were successfully decannulated without further surgical intervention; only two patients were syndromic and the remaining 11 patients were isolated.

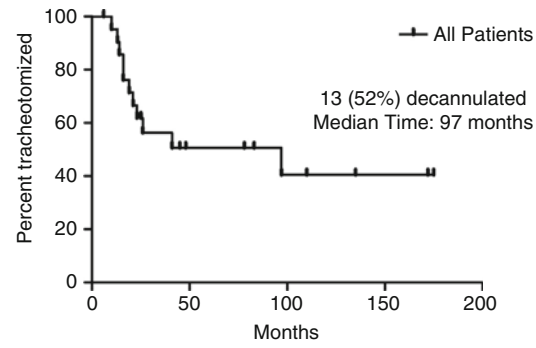

Fig. 11.3 Of 25 infants treated with tracheostomy, 13 infants (52 %) were eventually decannulated with median time to decannulation of 97 months

At mean follow-up of 4 years, the rate of tracheostomy-specific complications (e.g., cannula obstruction or accidental decannulation, delays in speech and language development, tracheomalacia (flaccidity of the tracheal support cartilage)/tracheitis, pneumonia) was 52 % and tracheostomy-specific mortality was 8 %. Patients with syndromic RS stayed in the hospital significantly longer than patients with isolated RS (50 versus 28 days, respectively). There was one death in each group; however, patients with syndromic RS had significantly more events of end-organ dysfunction (neurogenic, gastrointestinal, and/or cardiopulmonary) per patient than patients with isolated RS (2.08 versus 0.69, $p=0.005$).

11.4 Discussion

Based upon the literature, approximately 2/3 of patients with RS can be successfully managed with conservative therapies (Kochel et al. 2011; Gozu et al. 2010; Evans et al. 2006; Horikiri et al. 2010). Prone positioning is often sufficient in less severely affected patients but may result in prolonged hospitalization (Kochel et al. 2011; Sher 1992; Bhat et al. 2006). NPA can be effective in during the early clinical course to avoid emergent tracheostomy, but difficultly maintaining proper position for an extended period of time has limited its use (Chang et al. 2000; Sher 1992; Kochel et al. 2011; Masters et al. 1999; de Buys Roessingh et al. 2007). Lastly, orthopedic devices have been

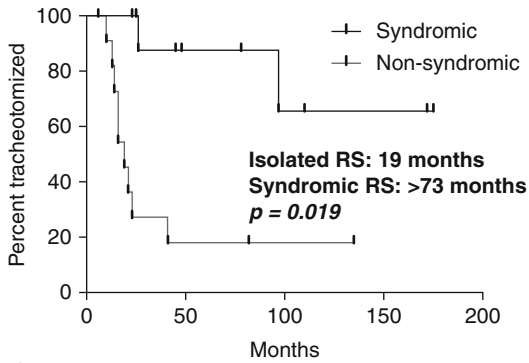

Fig. 11.4 Patients with syndromic RS had a median time to decannulation of >73 months versus patients with isolated RS had a median time to decannulation of 19 months, $p = 0.019$

tried with success in a few small studies but these devices are often expensive (Hotz and Gnoinski 1982; Buchenau et al. 2007; Kochel et al. 2011). More recently, Kochel et al. described the usage of newer orthopedic devices as another noninvasive way to treat upper airway obstruction. Three major types (plate with a posterior wire spur, plate with a posterior acrylic extension, and plate with a pharyngeal tube) of orthopedic devices were used in seven patients based on their mechanisms of the obstruction (Sher's classifications; see Sect. 11.4). All of the seven patients had normal oxygen saturation upon discharge and at the end of study period, five out seven patients tolerated the removal of the orthopedic devices (Kochel et al. 2011).

Surgical modalities such as glossopexy or TLA, mandibular distraction, mandibular extension, and subperiosteal release of floor of mouth muscle have been studied extensively for their effectiveness in avoiding or delaying tracheostomy, improving polysomnographic results, facilitating feeding, and bypassing or correcting the anomalous anatomy. TLA, subperiosteal release of floor of mouth muscle, and mandibular distraction have been described most frequently in the recent literature. TLA temporarily bypasses the obstruction by creating a stable airway with reported success rates between 70 and 100 % (Denny et al. 2004; Bijnen et al. 2009; Kirschner et al. 2003). Dehiscence (breaking down of the incision) still ranges between 0 and 57 % with a mean of 30 % (Sher 1992; Marques et al. 2001; Bookman et al. 2011). Based on a few studies, the need for a secondary invasive airway procedure such as tracheostomy after subperiosteal release ranges between 10 and 100 % (Delorme et al. 1989; Caouette-Laberge et al. 1996; Breugem et al. 2008; Siddique et al. 2000). Mandibular distraction has been shown to improve polysomnographic outcome, to avoid or delay tracheostomy, and to expedite decannulation of a tracheostomy (Scott et al. 2011; McCarthy et al. 1992; Denny et al. 2001; Lin et al. 2006; Schaefer et al. 2004; Schaefer and Gosain 2003). Nevertheless, complications such as infection, device failure, and nonunion occur from 2.5 to 52 % (Caouette-Laberge et al. 1994; McCarthy et al. 2002; Shetye et al. 2009). One large retrospective review of 141 infants with RS who underwent mandibular distraction reported a 52 % overall complication rate with a 5 % major complication rate – a complication that necessitated a secondary invasive therapy (Shetye et al. 2009).

Indications for surgical airway management are also highly debated. While many clinicians rely on a "gestalt" impression of airway obstruction, some have proposed data-driven clinical parameters for airway management. Caouette-Laberge et al. and Cole and colleagues proposed grading systems based on the settings of respiratory obstruction (Caouette-Laberge et al. 1994; Cole et al. 2008). Caouette-Laberge grouped 125 infants with RS into three categories: (1) adequate respiration in prone position and regular bottle-feeding, (2) adequate respiration in prone position and difficulty with feeding, and (3) endotracheal intubation (Caouette-Laberge et al. 1994). Parsons and Smith provided rule of thumb criteria for TLA in infants with RS; those who had progressive weight/strength gain over a 7-day period did not require TLA, while those infants who needed more than 3 days of endotracheal airway support should receive TLA (Parsons and Smith 1982). Freed et al. reported on the use of bedside monitoring and polysomnography to objectively guide airway management for infants with RS (Freed et al. 1988). Criteria for TLA included (1) an average transcutaneous O_2 level below 60 mmHg or transcutaneous CO_2

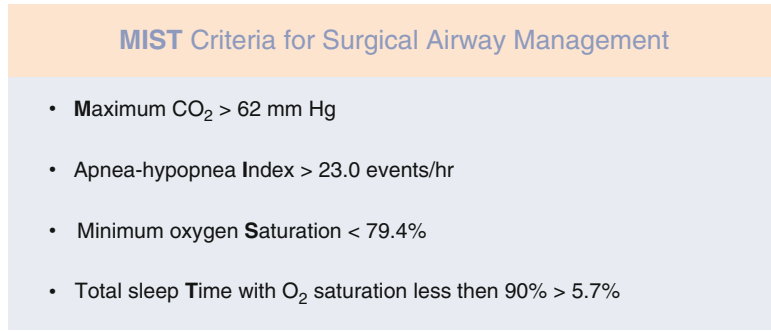

Fig. 11.5 *MIST* criteria for surgical airway management of patients with Robin sequence (*RS*)

level over 50 mmHg for a minimum of 8 h, (2) obstructive episodes on sleep study, and (3) oxygen saturation below 80 %.

More recently, Rogers et al. described the GILLS scoring system where one point was assigned to each of the five variables: *g*astroesophageal reflux, *i*ntubation preoperatively, *l*ate operation, *l*ow birth weight, and *s*yndromic diagnosis. TLA was 100 % successful in infants with a GILLS score of 2 or less but failed in 43 % of infants with a score of 3 or more (Rogers et al. 2011). Sher detailed the use of flexible fiber optic endoscopy to guide the form of surgical treatment in infants with RS failing conservative measures. Based upon nasopharyngoscopy and identification of the site of obstruction, infant airways were classified into four groups. Type I represented airway obstruction solely due to glossoptosis, while types II–IV had additional components of obstruction. Regardless of airway classification, all patients were initially treated with NPA placement for up to 8 weeks. If conservative therapy failed, type I patients received TLA and types II–IV underwent tracheostomy (Sher 1992). Schaefer et al. described an algorithm for approaching respiratory and nutritional dysfunction in infants with RS. Management decisions were based upon the ability to maintain progress on the growth curve, continuous pulse oximetry and bedside polysomnography, and the site of airway obstruction (Schaefer et al. 2004). Finally, data from our own institution (Fig. 11.5) from a retrospective review of airway management in patients with RS from 1994 to 2010 found that four clinical factors (*MIST* criteria) were most associated with surgical airway management: *m*aximum $CO_2 > 62$ mmHg, apnea–hypopnea *i*ndex (index of sleep apnea severity) > 23.0 events/h, minimum O_2 *s*aturation < 79.4 %, and greater than 5.7 % total sleep *t*ime with O_2 saturation less than 90 %. Each of these parameters identified operative intervention with 75–85 % accuracy (Seruya et al. 2011).

Despite the various procedures described for surgical airway management, tracheostomy remains the most definitive treatment for infants with severe respiratory obstruction that is not compatible with life. In many cases, tracheostomy is believed to be a temporary measure until "natural" mandibular growth permits decannulation. The topic of mandibular growth in patients with RS has been heavily debated yet remains poorly defined. Some authors have documented diminished mandibular size and proportions compared age-adjusted norms while others have cited evidence to the contrary (Shen et al. 2010; Hermann et al. 2003; Daskalogiannakis et al. 2001; Figueroa et al. 1991). Rogers et al. found that mandibular length was shorter in all patients with RS irrespective of the type of airway management, and the differences in both mandibular length and sagittal position varied significantly among all the syndromic subtypes (Rogers et al. 2009). Maalouf et al. found that 60 % of patients who received bilateral mandibular distraction maintained proportionate facial symmetry at the median follow-up time of 57 months; mandibular size, however, was not evaluated (Maalouf and Lehman 2011). Finally, Pruzansky and Richmond demonstrated the opportunity for "catch-up" growth of the mandible without the need for invasive procedures aside

from a temporary tracheostomy (Pruzansky and Richmond 2005). Overall, these studies highlight the phenotypic heterogeneity of patients with RS, which may stem from the dissimilar mandibular growth kinetics of isolated versus syndromic patients.

In our retrospective study of 61 infants with RS, 25 patients required tracheostomy (14 cases of isolated RS and 11 cases syndromic RS). This is one of the largest published cohorts of RS children with tracheostomy that have been followed to analyze rates of eventual decannulation due to mandibular growth without other ancillary surgical interventions. The median time to decannulation in our cohort was longer (97 months) than what other smaller series have reported (Tomaski et al. 1995; Demke et al. 2008). Patients with syndromic RS largely contributed to this extended time course, as most of them could not be decannulated by the completion of the study. Outcomes following tracheostomy were significantly poorer in patients with syndromic RS as compared to those with isolated RS: patients with syndromic RS had longer hospital stay and more long-term complications as compared to patients with isolated RS.

Summary and Conclusion

Airway management in patients with RS and severe airway obstruction remains controversial. The benefits of various surgical airway procedures, as well as indications for intervention, are unclear and based upon imperfect data. Our experience with these complex patients has documented some factors associated with the need for surgical intervention, but these criteria await validation in a randomized prospective trial. We have also found that although tracheostomy was intended to be a temporary airway for these patients, the time to natural decannulation was longer than expected. This may be partially explained by the mandible's inability to "catch up" in growth, especially in syndromic patients. We are planning further study into these issues by means of a prospective trial that will incorporate serial lateral cephalograms to document mandibular growth as well as offer mandibular distraction as a surgical modality to patients with severe airway obstruction. Based on our experience, we believe that all potential treatment options should be exhausted before offering tracheostomy to syndromic patients with Robin sequence.

Financial Disclosure Information The authors have no financial or commercial interests to disclose.

Conflict of Interest
None

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Pierre Robin Sequence

12

Samuel Pruzansky and Julius B. Richmond

With some additional photographs by Samuel Berkowitz, DDS, MS

12.1 Growth of Mandible in Infants with Micrognathia (Pruzansky and Richmond 1954)

Since Pierre Robin first described the syndrome of hypoplasia of the mandible, cleft palate, glossoptosis, inspiratory retraction of the sternum, cyanosis, and malnutrition which has come to bear his name (Robin 1929, 1934), numerous reports of cases have appeared in the literature. The fatal termination of some cases is a testimony of the serious nature of the syndrome. Although a variety of mechanical and surgical therapeutic procedures have been suggested (Eley and Farber 1930; Davis and Dunn 1933; Callister 1937; Lleweyllyn and Biggs 1943; Douglas 1946; Nisenson 1948; Longmire and Sandford 1949; May and Chun 1948), a rationale for the proper management of individual patients has not been adequately presented. It is the purpose of this communication to present observations from the serial studies of patients with this disorder, upon which a physiologic approach to management may be based.

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It is generally agreed that the pathophysiologic events in this syndrome are as follows: the receding chin fails to support the tongue in its normal forward relationship and hence fosters the glossoptosis. The retroposed tongue impinges against the posterior wall of the pharynx, obstructing inspiration and impeding feeding. Slight excess in mucus or saliva tends to enhance the pharyngeal obstruction and may precipitate severe cyanotic seizures, resulting in death. Starvation and respiratory infections, or both, may follow as a consequence of the chronic glossoptosis.

Since the major symptoms are definitely related to the micrognathia, it was of special interest to focus our attention upon the growth of the mandible in these infants. In reviewing the published case reports, it appeared that two questions, basic to an understanding of a more appropriate management of these cases, remained unanswered. The first question could be phrased as follows: How soon would mandibular growth be sufficient to accommodate the tongue and hence insure a more adequate airway? This is assuming that the infant could be placed in an adequate metabolic climate. The answer to this question would have an important baring on immediate management. The second question is related to whether or not mandibular growth would be sufficiently sustained to provide an esthetically satisfactory facial profile.

The answer to these questions had to await the development of accurate roentgenographic techniques for measuring the growth of the head in infants, which made it possible to undertake a

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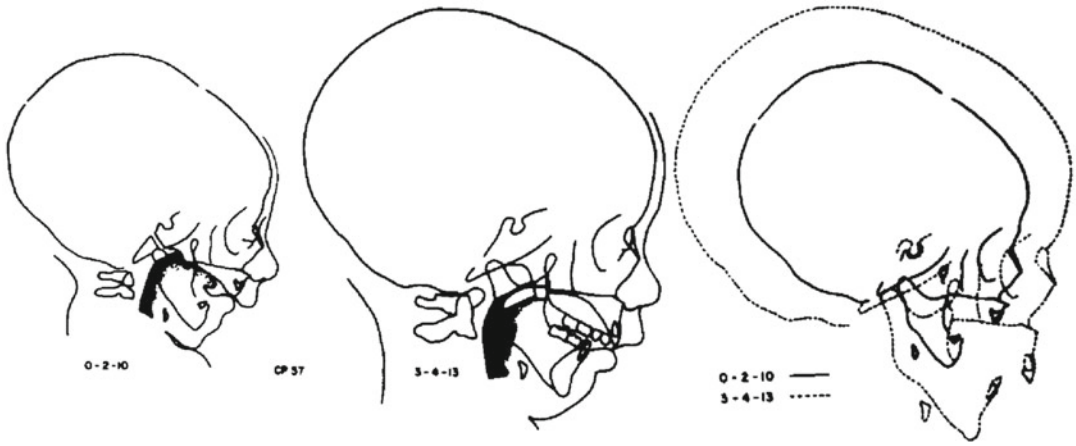

Fig. 12.1 (**a**) Serial cephalometric tracing of a child with Pierre Robin sequence showing a severely micrognathic mandible. At 3–4–13, there is a big increase in the pharyngeal space (*darkened area*). (**b**) Superimposed tracings at 2 months and 10 days (0–2–10) and 3 years, 4 months, and 13 days (3–4–13) shows a rapidly growing micrognathic mandible (Reprinted with permission from Pruzansky (1953))

serial study of the growth of the micrognathic mandible. The results of these investigations have provided useful information as a basis for therapy and prognosis.

During the past 25 years, cephalometric roentgenography has produced valuable information pertaining to the growth of the head in normal (Brodie 1941a, b) and pathologic conditions (Brodie 1941a, b) in man. In addition to the cephalometric roentgenographic measurements, dental impressions of the maxillary arch were obtained at regular intervals. The impressions were cast in dental stone and then subjected to various measurements. The infants were recalled for study every 3 months during the first year of life and twice annually until the age of 5 years. Thereafter, they were observed annually.

From a large series of similar cases now under longitudinal study at the Cleft Palate Center of the University of Illinois (Pruzansky 1953), we have selected three cases for presentation. At birth, each of these infants presents an isolated cleft of the palate and mandibular micrognathia. Despite these similarities, certain important differences existed to vary the management and progress of each infant. The differences to be described in each of these cases represent the major variations which we have encountered in our experience with this syndrome. For the sake of brevity, the case history will be confined to such information as is directly pertinent to the purpose of this chapter.

12.1.1 Case 1

At 2 months of age, D.R.P., a small white dehydrated baby girl, was admitted to the Research and Educational Hospital, with the principal complaint of intermittent pneumonitis for the previous 5 weeks. After a normal full-term spontaneous delivery, it was noted that the baby had an isolated cleft palate and a small mandible. The birth weight was 5 lb 13 oz (2,640 g). Feeding had been difficult because of the tendency of the baby's tongue to fall back into the pharynx. There was no history of a cleft on either side of the family. The mother gave no history of being exposed to a contagious disease or other illness during her first trimester of pregnancy (Fig. 12.1).

Because of the patient's congenital defects, she presented primarily a feeding problem. An attempt was made to design an obturator so that the baby could be bottle-fed. This was met with failure, and gastric gavage was necessary. However, the patient did not gain weight and continued to do poorly. Approximately 1 month after admission, it was considered that a tracheotomy was the only procedure which might

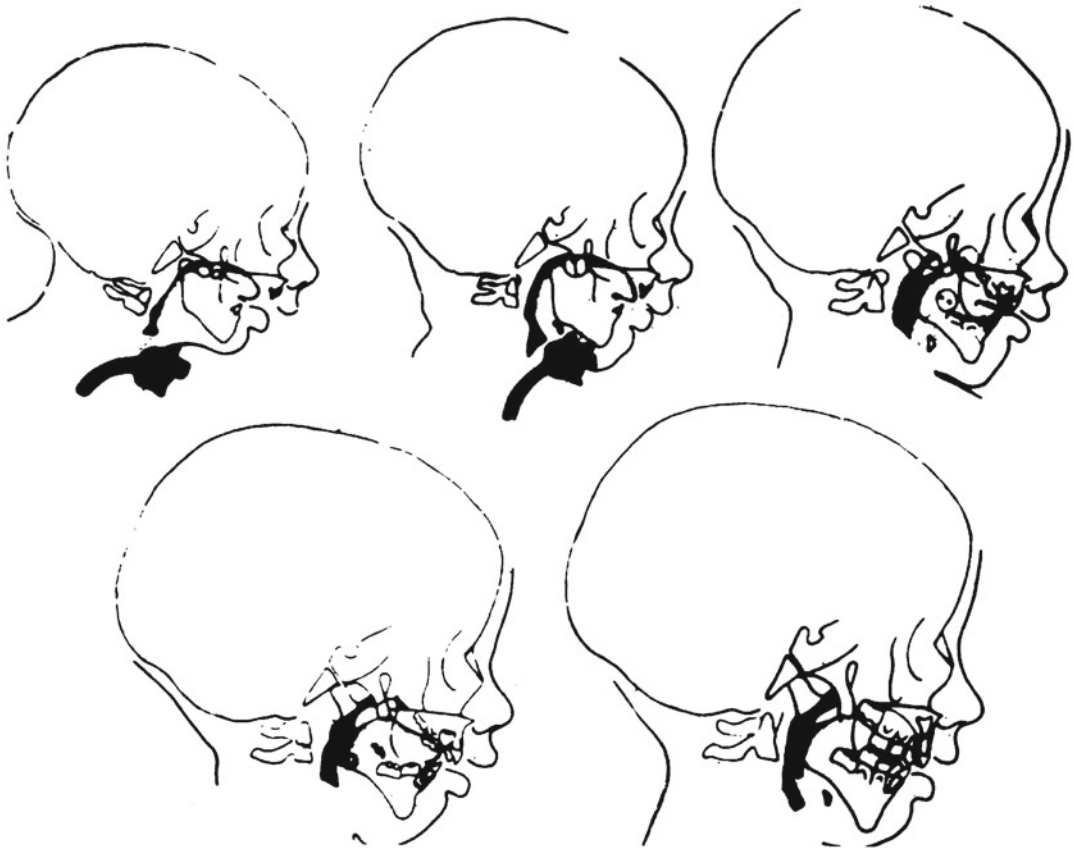

Fig. 12.2 Pierre Robin sequence. A series of tracings of the lateral film from 3 months, 1 day of age (0–3–1) to 3 years, 5 months, 12 days (3–5–12). The pharyngeal airway is filled in (*black*), and the stippled area denotes the border of the tongue. A tracheotomy tube is visible in the first two films. Soon after birth (3 and 4 months of age), the dorsum of the tongue is visible at a level above the palatal plane within the palatal cleft space. Posteriorly, the tongue just above the epiglottis impinged on the airway. At this level, the airway was almost completely occluded. Comparison with the remaining three figures, ages 0–8–10, 1–1–0, and 3–5–12, reveals the configuration of these structures under normal circumstances after closure of the cleft. The airway in its posteroanterior dimensions is fairly wide, and the tongue occupies a more protrusive relationship to the mandible (Reprinted with permission from Pruzansky (1954))

possibly save the baby's life. After the tracheotomy, respiration was greatly facilitated, and soon the patient was removed from the oxygen tent. Her improvement thereafter was constant. The patient was discharged from the hospital 3 months after admission, at 5 months of age.

Growth Studies: Oral examination disclosed a cleft involving the one-half of the hard palate and extending posteriorly throughout the length of the soft palate. Ptosis of the tongue was evident.

On the fifth day following the tracheotomy, at the age of 3 months 1 day, the first lateral film was obtained without sedation. A tracing of this film revealed that the head was in slight dorsiflexion, the position of greatest comfort for the infant. The relative smallness of the mandible and its effect on the facial profile was self-evident. In relation to contiguous anatomical structures, the posture of the tongue was abnormal. It projected through the cleft in the palate into the nasal cavity (Fig. 12.2).

In the lateral head plate, the dorsum of the tongue was visible at a level above the palatal plane. Posteriorly, the tongue impinged on the airway. The posterior surface of the tongue, just above the epiglottis, was in close approximation to the posterior outline of the pharyngeal wall. At this level, the airway was almost completely occluded. The tracheotomy tube was in place. The airway in its posteroanterior dimensions is

fairly wide, and the tongue occupies a more protrusive relationship to the mandible.

The second film in Fig. 12.2, obtained less than 6 weeks later, at the age of 4 months 13 days, revealed considerable enlargement of the airway. At the same time, a considerable increment in the growth of the head and, above all, of the mandible was recorded. Subsequent films, up to the age of 3 years 5 months 12 days, gave proof of the gradual and continued growth of the lower jaw in relation to the total face and the increase in the dimensions of the pharyngeal airway. At 8 months following removal of the tracheotomy tube, the air passage was demonstrated to be quite adequate (Fig. 12.2).

The superimposed tracings, from 3 months 1 day of age to 3 years 5 months 12 days, reveal the progressive growth of the cranial vault, the maxilla, and the middle face and the increase in length and height of the mandible (Fig. 12.1b). Gradual improvement in the facial profile was recorded by the changes in the facial angle and in the angle of convexity. The facial angle is a measure of the degree of protrusion or recession of the chin. In this instance, the facial angle increased from 64°, at 3 months of age, to 70°, at the age of 3 1/2 years, indicating a reduction by 6° in the recessiveness of the chin. While the mandible is still in a retrusive relation to the rest of the face, the potential for further improvement with continued growth still exists.

The changes in the angle of convexity were more interesting. This measurement relates the maxilla to the total facial profile. At 3 months of age, the angle of convexity was 140°, and at 3 1/2 years it measured at 154°. The integrated growth of the several areas of the face was such as to improve the overall configuration of the facial profile. Serial photographs at 2 months, at 13 months, and at 3 years of age further testify to the changes in this child's face. In the last photograph, the patient is posed beside her older sibling.

The changes in the position of the hyoid bone are of particular interest, insofar as they reflect a change in the relative position of the tongue. The tongue is composed of several individual muscles originating from the base of the skull, the mandible, the hyoid bone, and the walls of the pharynx. Changes in the position of any of its bony or fibrous attachments would tend to reflect on the position of the tongue. Conversely, changes in the posture of the tongue would reflect on the spatial relations of the mandible and hyoid bone. Therefore, to study the position of the hyoid bone is, in a sense, to study the position of the tongue. With growth, there occurs a forward and downward migration of the hyoid bone from the base of the skull. The pattern of changes in the posture of the hyoid bone observed in this patient sheds further light on the favorable adjustments consequent to growth. During the first 5 months of our studies, the hyoid bone migrated downward and forward. This resulted in an increase in the angle S-N-H. But, from 8 months onward, this angle became fairly stable and the hyoid bone began to descend principally in a downward direction.

Comment: This case was selected to typify the findings in several similar cases, one of which has been followed to the age of 7 years. Not all cases of Pierre Robin syndrome present such acute histories. When clinical evaluation suggests that there will be no improvement or that possibly death may ensue, tracheotomy should be undertaken without hesitation to prevent further aggravation of the symptoms. Once an adequate respiratory exchange was made possible, improvement in oxygenation and feeding followed. In such instances, we have recorded rapid growth and favorable changes in the facial appearance.

12.1.2 Case 2

J.G, a white girl, was referred to the outpatient clinic of the Cleft Palate Center at the age of 2 months with a diagnosis of cleft palate and mandibular micrognathia. Following an uneventful pregnancy, the delivery was normal and at full term. The birth weight was 6 lb 11 oz (3,030 g). The infant had some difficulty in breathing, but this was relieved by placing her in a prone position. Tube feeding was employed for the first few days after which she was given bottle feedings. At 6 days of age, the infant was discharged from the hospital. There was no family history of cleft palate. The mother suffered no illness during her pregnancy.

Oral examination revealed an unusually small tongue closely attached to the floor of the mouth. In the course of our first examination under sedation, the infant became cyanotic and failed to initiate mandibular movements sufficient to permit the passage of air. This was relieved immediately

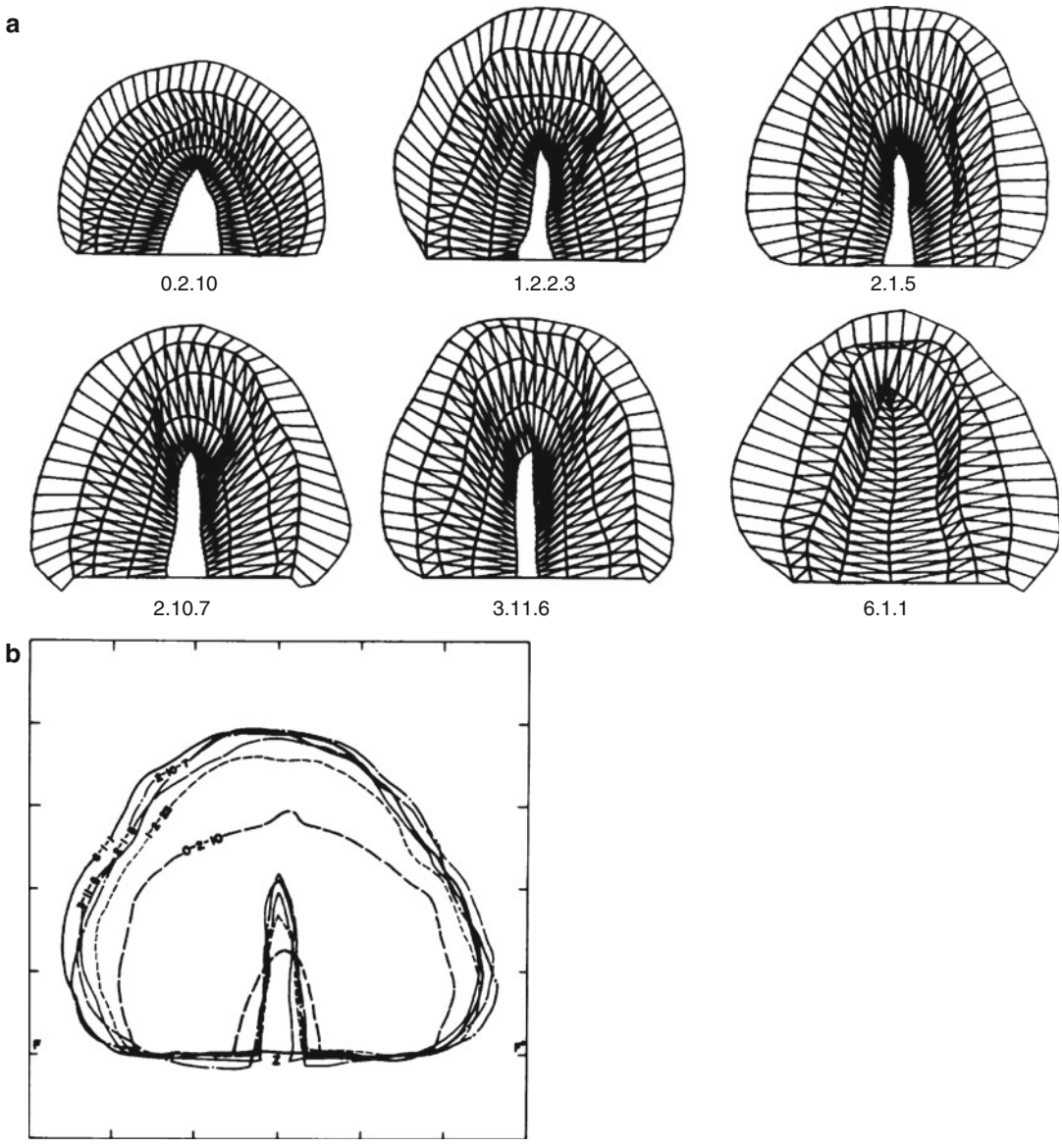

Fig. 12.3 (**a**, **b**) Palatal growth changes in a child with a Pierre Robin sequence. This sequence is characterized by glossoptosis, micrognathia, and isolated cleft palate. In many cases, the cleft palate which is initially wide at birth can spontaneously narrow with palatal growth. (**a**) Computer-generated tracings of the isolated cleft of the hard palate from 2 months and 10 days (0–2–10) to 6 years, 1 month, and 1 day (6–1–1). The palate was closed at 4 years, 2 months. (**b**) Superimposed tracings of each cast (on the baseline created by connecting postgingivale points (the posterior limits of the hard palate) and registered at the bisector of the line) show that the length of the cleft increases with palatal growth and narrows due to spontaneous growth at the medial border of the palatal processes. Obturators which interfere with tongue posturing within a relatively small intraoral space are contraindicated (Berkowitz 1996)

by maintaining forward traction on the tongue and mandible. After about 5 min, the infant recovered control of mandibular movements and respiration normally. Aside from this isolated episode, which occurred under sedation, the parents did not report any similar difficulties. The child has continued to grow and develop at a satisfactory rate.

Growth Studies: The casts disclose symmetrical cleft of the hard and soft palate, extending distally from the region of the nasopalatine foramen. Additional casts obtained at regular intervals revealed that the cleft had narrowed, so that it now presents a narrow V-shaped defect (Figs. 12.3 and 12.4).

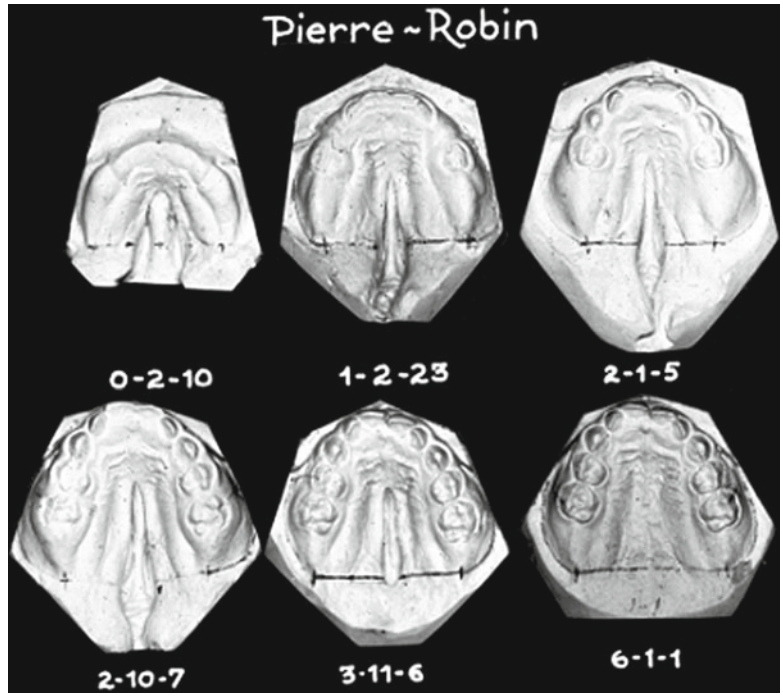

Fig. 12.4 A series of cast of the maxillary arch from 2 months 10 days of age to 6 years 1 month. Note the progressive narrowing in the lateral dimension of the cleft (Reprinted with permission from Pruzansky (1954))

The earliest lateral head palate, at 2 months 10 days of age, displayed a small mandible and small tongue. The latter was positioned high and above the floor of the nose, but relatively remote from the posterior wall of the pharynx. The airway appeared sufficient to sustain respiration without any undue effort on the part of the infant. Progressive growth changes recorded up to the age of 3 years, 4 months, 13 days disclosed mandibular growth and generalized growth in all areas of the face and cranial vault. Mandibular growth was continuous and progressively downward and forward. During the period studied, from 2 to 40 months of age, the facial angle increased from 61.5, becoming more obtuse. The angle of convexity increased from 147° to 155°. Altogether, the changes were in a direction tending to minimize the recessiveness of the chin in relation to the rest of the face.

Comment: Micrognathia by itself is not sufficient to produce glossoptosis and respiratory embarrassment. If the tongue is large or even normal in size, the small recessive mandible will tend to displace the tongue distally and superiorly. It is this displacement that produces the respiratory obstruction both into the hypopharynx and into the posterior choanae. On the other hand, if the tongue is small, there will be no obstruction of the airway even in the presence of a micrognathic mandible. In this instance, the simultaneous occurrence of micrognathia and microglossia averted the respiratory difficulties commonly experienced in such instances.

The tendency to lose reflex control of the muscles of respiration and deglutition under anesthesia or sedation renders such procedures unusually hazardous in these patients because of the limited reserve. It is, therefore, important that such procedures be undertaken with full knowledge and anticipation of possible respiratory obstruction, in order that adequate emergency provisions for the establishment of an airway be available.

12.1.3 Case 3

E.C., a white boy aged 5 weeks, was referred to the outpatient clinic of the Cleft Palate Center for longitudinal growth studies. The delivery had been normal and at full term. His birth weight was 7 lb 8 oz (3,400 g). There was no history of

cleft on either side of the family. No difficulty in breathing was encountered, and the infant was discharged from the hospital on the sixth day. After a brief adjustment period, the infant was readily fed by a combination of a hard nipple and by means of a premature baby bottle nipple.

Some snoring sounds were heard, especially as the infant was placed on its back and the head elevated with slight ventroflexion on the chest. The infant preferred to sleep on either side, and in these positions, the snoring sounds were at a minimum. This baby showed progressive improvement, and at the age of 5 months, he weighed 15 lb 8 oz (7,030 g).

Growth Studies: Two sets of records are available in this case. The first was obtained at the age of 1 month 7 days, and the second at 3 months 25 days of age.

The first cast of maxilla revealed a wide parabolic cleft extending distally from the nasopalatine foramen. The widest portion of the cleft, at the level of the maxillary tuberosities, measured 16 mm. Although the second cast exhibited an increase in the length and width of the palate, there was a decrease of 1.5 mm in the width of the cleft at its widest portion. During the first examination, the tongue was observed to occupy at rest the opening into the nasal chambers provided by the cleft in the palate. This was further confirmed by examining the frontal and lateral views of the head plates. The second series of films indicated that the tongue was now postured in a more inferior position and no longer occupied the nasal cavity to the same extent previously noted. This new position of the tongue could be explained by the downward and forward growth of the mandible that had occurred in the interim.

In the first lateral film, the recessive chin, the distally and superiorly malposed tongue, and relatively restricted airway were clearly observed. Two and one-half months later, considerable growth in the mandible had occurred to improve the facial profile, alter the posture of the tongue, and increase the anteroposterior diameter of the airway. The tongue was no longer in close apposition to the posterior pharyngeal wall, and its superior margin did not extend into the nasal cavity to the degree previously observed. Coincidentally, the mother reported a diminution of the stertorous breathing that had been present.

Superimposition of the tracings of the bony structures revealed the rapid growth characteristic of this early period in life. In 2 1/2 months, that cranial vault and all parts of the face exhibited proportionate increases. Particularly encouraging was the amount and direction of growth displayed by the lower jaw. Mandibular growth was responsible not only for reducing the glossoptosis and increasing the airway but for the improvement in the appearance of this baby's face.

Comment: The problem presented by this baby was unique and different from the two previous cases of the partial obstruction of the airway. Diligent nursing care to determine the most comfortable position for breathing and feeding may be sufficient to tide such cases through their critical period. In some instances, the prone positioning and orthostatic feeding suggested in the literature are most successful. Again, one is impressed by the remarkable potential for prolific growth during this period of life; a potential that is shared by the small mandible. It follows then that every effort must be made to permit the realization of the baby's potential for growth by providing an adequate airway, which, in turn, facilitates feeding. The clinical course to be followed is varied and depends on the severity of the symptoms and principally upon the degree of obstruction of the airway.

12.2 Comment

The representative sampling of cases presented provides an answer to the questions which the study was designed to solve. It is observed from the data presented that the mandible possesses remarkable potentialities for growth in patients with the Pierre Robin syndrome. Thus, all efforts should be directed toward sustaining life in a metabolically favorable climate in order that a more physiologic airway may be established as growth proceeds. With growth, the glossoptosis is minimized and spontaneous resolution of the

respiratory and feeding problems occurs. It is our opinion that tracheotomy should be resorted to promptly if respiratory embarrassment is significant in order to achieve a sufficient airway to provide adequate oxygenation. This is undoubtedly a lifesaving procedure in some patients.

On the basis of our longitudinal growth studies, certain prognostications concerning the future growth of the micrognathic mandible are permissible. In most instances, the increment in mandibular growth, as related to total facial growth, is sufficient to overcome the extreme recessiveness of the chin that is observed at birth. Since mandibular growth continues until late adolescence, it is possible to hope for an esthetically pleasing profile in adulthood. The management of the cleft palate has been in keeping with the established criteria for the treatment of palatal defects.

The lateral cephalometric film served as a valuable diagnostic tool in estimating the degree of obstruction of the airway as a result of the glossoptosis. In our experience, there was a high positive correlation between the degree of obstruction revealed in the x-ray film and the incidence and severity of the respiratory difficulties. When obstruction of the air passage was complete and the tongue was practically in contact with the posterior wall of the pharynx, tracheotomy was recommended as a lifesaving procedure. If the obstruction was incomplete, more conservative measures were employed. Care was taken to ascertain the most comfortable postures for breathing and feeding for the individual case, and the nurse or parent was carefully instructed in the care of the infant. Appropriate nipples were selected to minimize the energy expended by the infant in the feeding process.

In the course of these studies, we were aware of an obvious objection to placing so much reliance on these roentgenograms. Since many of these films were obtained under mild sedation, was it not possible that the posture of the mandible or of the tongue might have been altered by the sedative? Secondly, the film depicted a static view of the airway and represented only two dimensions. Did this view properly reflect the kinetic ability of the infant to manipulate the tongue and jaw hence the consistent correlation between the findings in our films and the clinical state? Moreover, when the films were repeated in the same infant without sedation, similar postures were recorded for the structures under analysis. It was important that the postures of the head in relation to the neck be kept constant. Dorsiflexion or ventroflexion of the head varied to posture of the mandible and tongue and produced changes in the configuration of the airway. To indicate alterations in the posture of the head to the neck, our tracings purposely included at least the first two cervical vertebrae.

We recognize that few institutions possess cephalometric roentgenographic equipment. Therefore, we would like to point out that an ordinary lateral film obtained by carefully positioning the infant can provide useful diagnostic data. To minimize enlargement, a target-object distance of at least 3 ft (90 cm) is recommended. For the sake of definition and to further decrease enlargement, the object film distance should be kept at a minimum. Sjölin (1950) has published interesting films to describe his experiences with a case of micrognathia. Although his films did not permit quantification of the growth changes, they were adequate for diagnostic purposes.

A number of papers in the literature claim to "stimulate" the growth of the mandible by a variety of mechanical devices or surgical procedures. For example, a special nursing bottle was designed to force the infant to protrude his jaw in order to obtain nourishment and, by this protrusion, to stimulate mandibular growth (Eley and Farber 1930). From our data, we would conclude that the nursing care enabled the infant to survive until mandibular growth was sufficient to provide a more adequate airway.

In another report, continuous traction on the mandible was maintained by circumferential wiring around the symphysis. The authors claimed growth-stimulating properties for this procedure (Longmire and Sandford 1949). From the findings in our series, it would seem that mandibular growth probably occurred spontaneously and not because of the stimulus provided by surgical traction.

The important and prime objective in the care of these children is to provide an airway. If possible, this should be accomplished with a minimum

of trauma. Secondly, the infant's total needs should be assessed to provide optimal conditions for somatic growth. As the potential for growth is permitted to express itself, the chin grows downward and forward away from the base of the skull. With this pattern of growth, adequate space for the tongue is provided, the airway enlarges, and there follows a spontaneous resolution of the symptoms. Also, there are progressive improvements in the facial appearance.

There is another dimension to the abnormal posture of the tongue, as observed in these patients, that merits discussion. Not only does the tongue block the pharyngeal processes and hence prevent their fusion. The high incidence of micrognathia in the population of clefts involving only the hard and soft palate lends support to this theory. Mandibular micrognathia is a physiological finding in early intrauterine life. If for some reason the micrognathia persists and fails to carry the tongue down and out of the nasal cavity, a cleft in the palate might result.

In early postnatal life, the tongue acts to keep the cleft palatal processes apart. As the tongue descends with mandibular growth and no longer forcefully intrudes itself into the nasal cavity, the palatal processes tend to approximate in the midline. Fusion of the palatal processes cannot occur, but the narrowing in the clefts is recorded fact.

Summary and Conclusions

The development of the accurate techniques for cephalometric roentgenography of infants has made possible a longitudinal study of the growth of the micrognathic mandible. As a result of these studies, useful diagnostic and prognostic information has been obtained to provide a rationale for the management of individual cases.

The lateral cephalometric roentgenogram is a valuable diagnostic aid in assessing the severity of the glossoptosis and its obstruction of the airway. A definite correlation exists between the degree of constriction of the airway and the severity of the clinical state. On the basis of these findings, it is possible to recommend either conservative management or tracheotomy in extreme situations, or distraction osteogenesis. Three cases, out of a larger series of similar cases, were presented to indicate the spectrum of variations to be encountered.

In all instances, it was found that where an adequate metabolic situation was provided and the infant gained weight, mandibular growth during the first few months was sufficient to provide for a natural resolution of the symptoms attending the glossoptosis.

Longitudinal records have indicated that mandibular growth is proportionally adequate to reduce the retrognathic profile and provide an esthetically harmonious facial appearance.

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Part IV

Audiology/Otology

13 Management of Otopathology and Hearing Loss in Children with Cleft Palate and Craniofacial Anomalies

Amelia F. Drake and Jackson Roush

Children with cleft palate and other craniofacial anomalies are at risk for a variety of otopathologies including otitis media and structural deformities of the outer and middle ear. These conditions are usually associated with conductive hearing impairment; however, when hearing loss occurs as part of a syndrome, there may be sensorineural or mixed hearing loss. This chapter will review the etiology of these conditions and their medical management. We will also examine the assessment of hearing and various technologies available for treatment of hearing loss.

13.1 Etiology and Medical Management of Middle Ear Disease

Because of the shared embryological origins of the ear and other craniofacial structures, patients with cleft palate and craniofacial anomalies are at increased risk for a variety of otopathologies including otitis media, cholesteatoma, and structural abnormalities of the outer and middle ear. Otitis media is, by far, the most prevalent otopathology seen in patients with cleft palate (Paradise et al. 1969; Moller 1975; Hubbard et al. 1985; Rynnel-Dagoo et al. 1992; Sheahan et al. 2003, 2004; Flynn et al. 2009; Zheng et al. 2009); however, a few may require tympanoplasty, tympanomastoidectomy, or ossicular chain reconstruction (Goudy et al. 2006). The high prevalence of otitis media in this population is due primarily to poor Eustachian tube function during swallow (Doyle et al. 1980; Takahashi et al. 1994). In the normal swallow, the velopharyngeal sphincter, which includes the levator veli palatini and tensor veli palatini muscles, closes the nose from the oral cavity to prevent reflux of oral secretions and food into the nasopharynx. The tensor veli palatini, the primary muscle responsible for opening the Eustachian tube, originates from the Eustachian tube and inserts on the palate where it joins muscles from the contralateral side to form a supporting sling for the palate. In patients with cleft palate, the sling is poorly developed and follows a more vertical orientation resulting in greater risk of reflux into the middle ear. The outcome is poor aeration of the middle ear and a high prevalence of otitis media (Bluestone 1971; Fria et al. 1987; Flynn et al. 2009). Although the prevalence decreases with age, some patients continue to experience otitis media as adults (Handzic-Cuk et al. 1996, 2001; Sheahan et al. 2003).

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Diagnosis of acute otitis media (AOM) requires (1) a history of acute onset of signs and symptoms; (2) middle ear effusion based on evidence of a bulging tympanic membrane, limited or absent mobility of the tympanic membrane, observation of an air-fluid level behind the tympanic membrane, or otorrhea; and (3) signs and symptoms of middle ear inflammation with evidence of otalgia (ear pain) or erythema of the tympanic membrane (AAP 2004). Otitis media with effusion (OME) is defined as the presence of fluid in the middle ear without signs or symptoms of AOM. It is more difficult to detect than AOM because in most cases, the only overt symptom is fluctuating hearing loss.

Otoscopy refers to examination of the eardrum or tympanic membrane using an otoscope and is part of any complete physical examination (Stool 2006). Tympanometry, an important adjunct to otoscopy, is a physiologic measure that provides information concerning the mobility of the tympanic membrane and middle ear system as well as middle ear pressure and estimates of equivalent volume, each of which may be associated with various middle ear conditions (Roush and Grose 2006). Figure 13.1 shows a series of tympanograms and their clinical interpretation.

Acute otitis media, when treated medically, is treated with antibiotics to cover for *H. influenzae*, *S. pneumoniae*, or *M. catarrhalis*. For most penicillin-resistant organisms, amoxicillin/clavulanate has proven effective (AAP 2004). When middle ear effusion persists for more than 3 months, as it often does in children with cleft palate, it is considered chronic. Tympanostomy tube placement is a common practice for those children, and improvements in hearing have been noted (Gould 1990) with relatively few complications (Curtin et al. 2009); however, multiple tube insertions have been associated with persistent conductive hearing loss (Goudy et al. 2006). Although it is unclear whether this is due to the placement of the tympanostomy tubes or to middle ear damage from the inflammation associated with otitis media, some clinicians have favored hearing aids over multiple tube placements (Maheshwar et al. 2002).

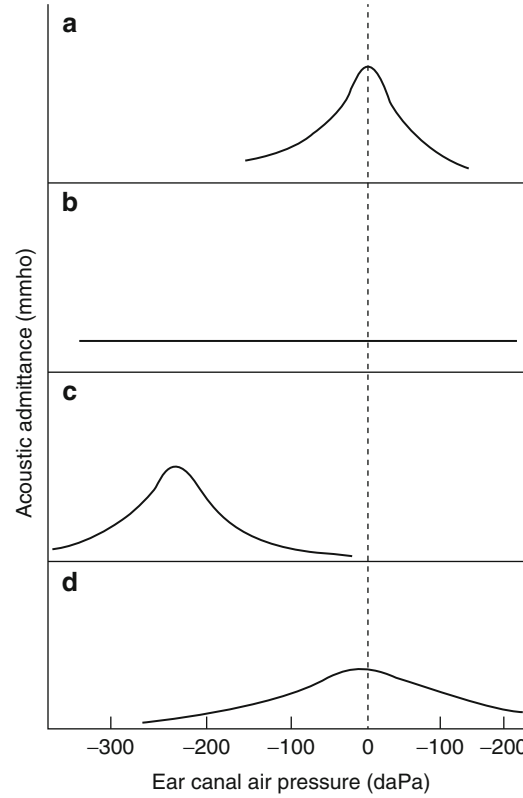

Fig. 13.1 Tympanometric patterns associated with normal middle ear function (**a**), middle ear effusion (**b**), negative middle ear pressure (**c**), and reduced middle ear mobility (**d**) (Reprinted with permission from Roush and Grose (2006), p. 378)

Complications from middle disease, although relatively rare, include perforation or retraction of the tympanic membrane, resulting in cholesteatoma. Cholesteatoma refers to a benign but locally erosive mass of squamous cells that often begins with a tympanic membrane retraction pocket or perforation. It is a potentially serious condition that warrants evaluation by an otolaryngologist and subsequent surgery.

Surgical repair of the palate has been shown to improve Eustachian tube function and decrease the frequency of otitis media and need for tympanostomy tubes (Bluestone 1971); however, it may take several years for Eustachian tube function to fully recover (Smith et al. 1994; Goudy et al. 2006). As with

the non-cleft palate population, the likelihood of hearing loss decreases with age (Gordon et al. 1988); however, hearing loss persists in adulthood for some patients.

13.2 Assessment of Hearing

Newborn hearing screening is now a standard of care throughout the United States (JCIH 2007). Two technologies are used for newborn screening and both employ physiologic methods; that is, they involve physiologic measurement of auditory function obtained without the infant's active participation. The first involves the measurement of otoacoustic emissions (OAEs) which are low-intensity sounds produced by the inner ear in response to acoustic stimuli (tones or clicks) and detected by a sensitive microphone placed in the ear canal. Successful recording of OAEs confirms a healthy inner ear (cochlea) and is consistent with normal or near-normal hearing sensitivity. Since the middle ear is involved in both the conduction of acoustic stimuli to the inner ear and reverse transmission of OAEs to the ear canal, their presence also confirms normal middle ear function. OAEs are "preneural" so their inclusion in the test battery makes it possible to evaluate the auditory system at the level of the inner ear without involving higher auditory centers. However, absent OAEs may be due to a variety of conditions ranging from middle ear dysfunction to profound cochlear hearing loss; other tests are needed to resolve their absence. OAE screening may be performed by a variety of professionals or support personnel, while diagnostic OAE testing is performed by an audiologist as part of a test battery. Another physiologic test used for both hearing screening and diagnosis of hearing loss is the auditory brainstem response (ABR). Like OAEs, ABRs are elicited by acoustic stimuli presented in the ear canal; however, ABRs are neurological responses obtained from surface electrodes attached to the head and provide information regarding the functional integrity of the auditory nerve and brainstem pathway. When used in a diagnostic test battery, audiologists use frequency-specific ABR testing to estimate hearing threshold levels.

Although OAE and ABR tests provide valuable information regarding the auditory system at the preneural, auditory nerve, and brainstem levels, they are not considered true tests of hearing in the perceptual sense. Comprehensive assessment of hearing requires behavioral tests, that is, procedures that involve observation of a listener's response to sound. Methods used for behavioral assessment of hearing vary depending on the child's age and developmental status. When infants reach a developmental age of approximately 6 months, most can be tested using an operant conditioning procedure known as visual reinforcement audiometry. By age 3–4 years, behavioral testing can be accomplished using conditioned play procedures, and by 5 years of age, typically developing children respond by raising their hand or pressing a response button.

To summarize, newborn hearing screening is conducted using physiologic measures: otoacoustic emissions or auditory brainstem responses, alone or in combination. Infants who do not pass the newborn hearing screening are referred to an audiologist for comprehensive assessment using these and other specialized procedures to determine if hearing impairment is present and, if so, to ascertain the type and degree of hearing loss. When infants reach a developmental age of approximately 6 months, they can be tested using behavioral methods. When permanent hearing loss is diagnosed, most children benefit from acoustic amplification (hearing aids). Those with severe-profound hearing loss are likely to benefit from a cochlear implant.

The initial goal of the audiologic assessment, whether conducted using physiologic or behavioral methods, is to obtain a frequency-specific estimate of the child's hearing thresholds for each ear, based on detection levels for air- and bone-conducted test stimuli. Air conduction audiometry involves the presentation of pure tones from an earphone or insert receiver; bone conduction audiometry involves the presentation

of signals through a bone vibrator placed behind the ear on the mastoid process. Testing is usually performed at octave intervals from 250 to 8,000 Hz for air conduction and 250–4,000 Hz for bone conduction. Thresholds are displayed on an audiogram which plots threshold levels in decibels hearing level (dB HL) as a function of frequency, using standard symbols (Fig. 13.2). The degree of hearing loss can be summarized by averaging the pure-tone air conduction thresholds in the mid-frequencies. Terms used to classify hearing levels include normal (0–15 dB HL), borderline normal (16–25 dB HL), mild (26–45 dB HL), moderate (46–75 dB HL), severe (76–0 dB HL), and profound hearing loss (>90+ dB HL). Borderline categories may be described using a combination of terms, such as moderate to severe (Roush and Grose 2006).

When pure-tone air conduction thresholds are abnormally elevated, bone conduction testing is performed to differentiate problems with sound transmission lateral to the inner ear. As shown in Fig. 13.2a, when air and bone conduction thresholds are equally elevated, the loss is described as sensorineural. Conductive hearing loss, illustrated in Fig. 13.2b, is characterized by normal or near-normal bone conduction thresholds with elevated air conduction thresholds. A mixed hearing loss, shown in Fig. 13.2c, is characterized by abnormal responses to both air conduction and bone conduction signals, with air conduction thresholds poorer than bone conduction thresholds.

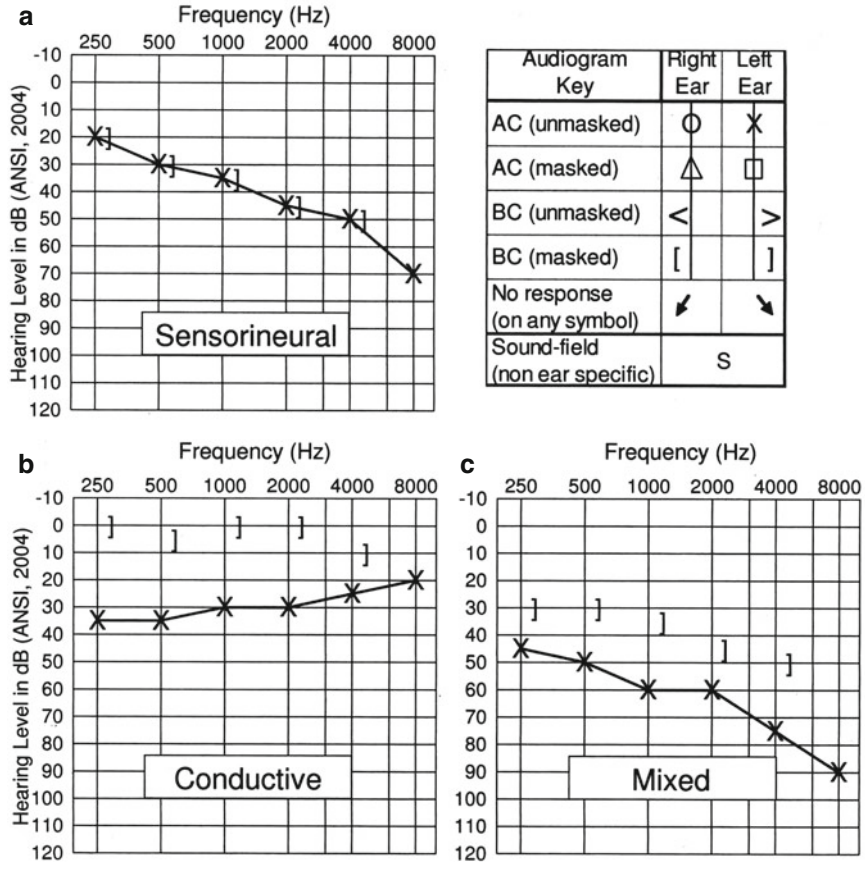

Fig. 13.2 Audiograms illustrating mild to moderate sensorineural hearing loss (**a**), mild conductive hearing loss (**b**), and moderate-to-severe mixed hearing loss (**c**) (Reprinted with permission from Kramer (2008), p. 153)

13.3 Hearing Loss in Children with Cleft Palate and Craniofacial Anomalies

As noted earlier, most hearing losses that occur with cleft palate and craniofacial anomalies are conductive in nature. That is, incoming sounds are attenuated by aural atresia or middle ear disease. When cleft palate or craniofacial anomalies occur as part of a syndrome, they may include cochlear or labyrinthine dysplasias that result in sensorineural or mixed hearing loss. These include Treacher-Collins syndrome, Goldenhar syndrome (oculoauriculovertebral or OAV spectrum), and craniofacial microsomia. CHARGE syndrome may involve abnormalities of the outer, middle, and inner ear, while Stickler syndrome, which is often identified by a Pierre Robin sequence, has been associated with progressive sensorineural hearing loss. Velocardiofacial syndrome (chromosome 22 deletion syndrome) may include sensorineural hearing loss, and when accompanied by cleft palate, Eustachian tube dysfunction, and otitis media, a mixed hearing impairment. Syndromes involving craniosynostosis may include auricular dysplasias, canal atresia, ossicular fixation, and sensorineural hearing loss. Tympanostomy tube placement may be indicated if feasible, and some patients will be candidates for ossicular reconstruction. Jarhsdoerfer et al. (1992) developed criteria to predict the success of atresia surgery based on otologic findings that include the presence of ossicles, an oval window, round window, facial nerve, and external ear. Unfortunately, only about half of patients with aural atresia are candidates for surgical repair, and many of those will require revision surgeries (Jarhsdoerfer et al. 1992).

13.4 Management of Hearing Loss in Children with Cleft Palate and Craniofacial Anomalies

The challenge of determining hearing status for these children often begins at birth. Many infants with cleft palates do not pass the newborn hearing screening because of middle ear effusion and require referral to a pediatric audiologist for follow-up assessment. Although most will not be found to have a permanent, underlying sensorineural or conductive hearing loss, it is important to determine the child's hearing status early as possible. For typically developing infants, the test used to make this determination, diagnostic ABR, can be easily accomplished in natural sleep. But for infants born with cleft palate, the test is often complicated by middle ear effusion. Although ABR testing can be completed using bone-conducted as well as air-conducted stimuli, it is preferable to perform the ABR when the middle ear is free of effusion. Another potential complication is the noisy breathing of some young infants with cleft palate, which may result in artifacts that affect the quality of the ABR when recorded in natural sleep. If an adequate ABR study cannot be performed in natural sleep, it must be completed with sedation or under general anesthetic. Sedated or operating room procedures add additional time and expense but enable the high-quality assessment needed for prompt, accurate diagnosis. The clinician must decide whether a separate procedure for diagnostic ABR is warranted or if the test can be performed at the time of cleft repair or tympanostomy tube placement, being mindful of the importance of avoiding excessive delay between screening and diagnostic ABR evaluation.

When hearing loss is diagnosed, timely and appropriate intervention are essential. In many cases, medical management will resolve a transient middle ear disorder and restore normal hearing. But hearing loss in children with cleft palate and craniofacial anomalies is often chronic, even with appropriate medical management. When conductive hearing loss cannot be resolved through medical intervention, hearing aid use should be considered. For children, hearing aids worn behind the ear are preferred whenever possible because they allow flexibility in programming, and they work well with assistive listening devices used at home and in the classroom. When acoustic amplification is precluded by aural atresia or chronic otorrhea, a bone conduction device is usually indicated. Most bone conduction hearing aids consist of an oscillator fitted to a headband that maintains pressure of the bone vibrator

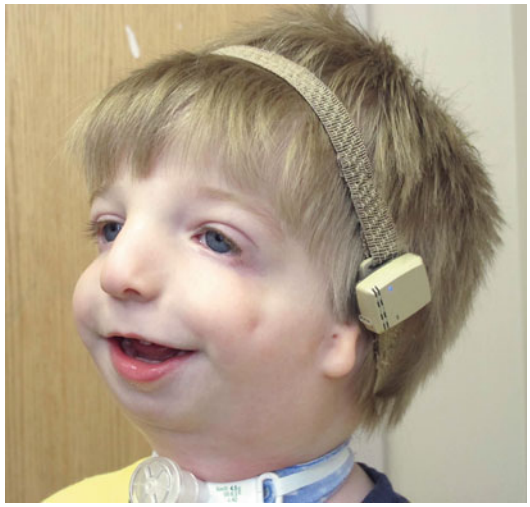

Fig. 13.3 A 3-year-old child with Treacher-Collins syndrome using a Baha softband bone conduction device. A vibrotactile transducer, held in place by an elastic strap, transmits vibrations from incoming sounds directly to the inner ear, bypassing the outer and middle ear

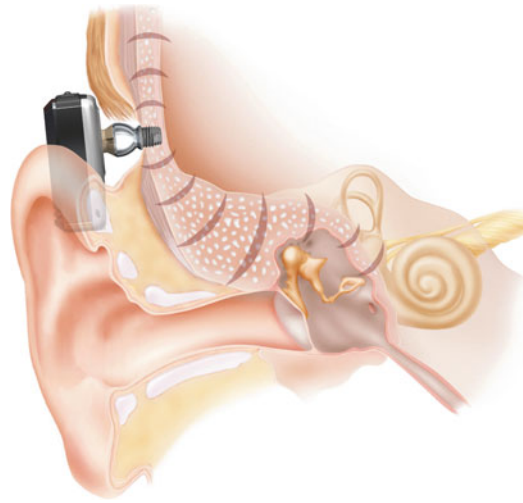

Fig. 13.4 The implanted bone-anchored hearing device consists of an external sound processor attached to a titanium abutment that is surgically implanted into the skull (Courtesy of Cochlear Corporation)

against the scull (Fig. 13.3). Because the inner ear is often unaffected by aural atresia, the bone oscillator is able to bypass the impaired outer and/or middle ear and deliver sound directly to the cochlea. Older children may be candidates for a percutaneous, bone-anchored device that delivers vibrotactile stimulation from a hearing instrument mounted to a titanium abutment surgically implanted in the temporal bone (Fig. 13.4). The surgically implanted, bone-anchored device provides stable retention and efficient transmission of vibrotactile signals, making it usable with mixed hearing impairment.

As noted earlier, although most hearing losses in children with cleft palate or craniofacial anomalies are conductive in nature, it is important to provide comprehensive audiological assessment to rule out a sensorineural or mixed impairment. When sensorineural hearing loss is identified, the audiologist and otolaryngologist will determine the best approach to hearing technology. An ear-level, air conduction hearing aid may be appropriate, or in cases of severe-profound hearing loss, a cochlear implant may be indicated. Cochlear implants bypass the inner ear, providing electrical stimulation from an external sound processer and coil that transmit digital signals across the skin to an internally implanted electrode that provides direct stimulation of the auditory nerve.

13.5 Summary

Hearing is vital for speech, language, cognitive, and social development. Nearly all children with cleft palate and craniofacial anomalies will experience middle ear disease and/or conductive hearing loss, and some are at risk for sensorineural or mixed hearing impairment. Careful monitoring of ear and hearing status by an otolaryngologist and audiologist are essential. With appropriate management, conductive hearing loss can often be avoided or ameliorated. When there is permanent (sensorineural) or chronic conductive impairment, several hearing technologies are available for delivery of sound via air conduction, bone conduction, or electrical stimulation.

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Part V

Effects of Surgery and How It Was Utilized

Palatal Wound Healing: The Effects of Scarring on Growth

14

Johannes W. Von den Hoff, Jaap C. Maltha, and Anne Marie Kuijpers-Jagtman

14.1 Introduction

Cleft palate patients often develop growth disturbances of the midfacial region after primary surgery. Factors such as intrinsic developmental deficiencies and functional distortions are initially involved, but palatal repair seems to be a main factor in these growth disturbances (Ross 1987a, b, c, d, e, f, g). A strong indication for the involvement of iatrogenic factors is the largely undisturbed maxillary growth in untreated patients (Derijcke et al. 1994; Mars and Houston 1990; Ortiz-Monasterio et al. 1966). In treated patients, the healing of surgical wounds originating from palatal repair is probably responsible for the growth disturbances (Kuijpers-Jagtman and Long 2000). This chapter gives an overview of the wound healing process with emphasis on wound contraction and scar formation since they are considered to be key events. Some specific features of the palatal wound healing process are highlighted. Further, the effects of palatal repair on growth of the maxilla and development of the dentition are reviewed, as well as possible means to improve the clinical outcome. This review is based on clinical evaluations, experimental research in animal models, and on in vitro experiments using cell culturing and tissue engineering techniques.

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14.2 Wound Healing

14.2.1 Skin and Oral Mucosa

The general function of wound healing is to restore the integrity and function of the tissue. In tissues like skin and oral mucosa, wound healing involves a partly overlapping sequence of inflammation, tissue formation, and tissue remodeling (Fig. 14.1). During inflammation, hemostasis is restored and bacteria and debris are removed from the wound. Subsequently, the defect is closed by the formation of new tissues and by wound contraction. Finally, tissue remodeling takes place during maturation of the newly formed tissues, which generally leads to the formation of a scar.

In intraoral wounds, the wound healing process is generally faster than in skin and generates less scar tissue. Therefore, intraoral wounds are sometimes considered to be more similar to fetal wounds (Okazaki et al. 2002). This may be related to the presence of lower levels of pro-inflammatory and pro-fibrotic cytokines in mucosal wounds (Szpaderska et al. 2003). The intraoral wound healing process is also influenced by the presence of saliva and large numbers of bacteria (Zelles et al. 1995). Saliva

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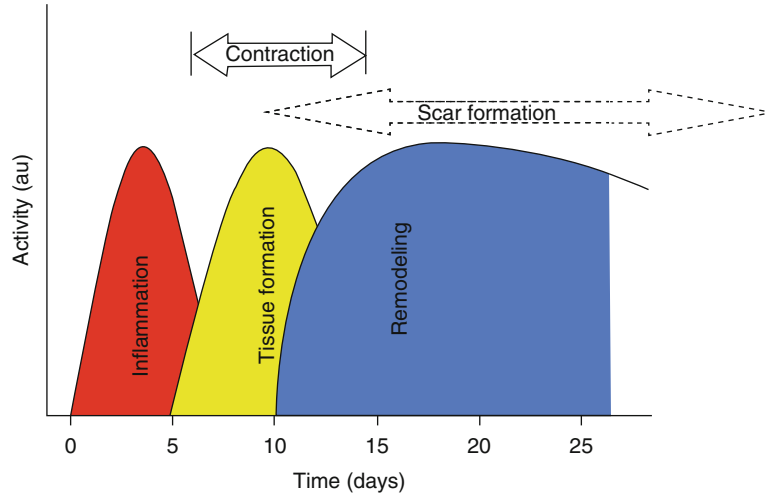

Fig. 14.1 Phases in wound healing. The wound healing process can be divided into three partly overlapping phases. Wound contraction is an early phenomenon, while scar formation is a late phenomenon in wound healing (*arrows*)

contains many growth factors such as epidermal growth factor (EGF). In addition, phenotypic differences between skin and mucosal fibroblasts may be involved (Lepekhin et al. 2002). These considerations, however, mainly apply to buccal mucosa, which has a quite different morphology than the palatal mucosa (Fig. 14.2).

In contrast to buccal mucosa, the palatal mucosa is a mucoperiosteum, which means that mucosa and periosteum are merged and attached to the palatal bone (Squier and Finkelstein 2003). The palatal mucoperiosteum is also much stiffer than buccal mucosa, and it contains less elastin fibers, as was also shown for the gingival mucoperiosteum (Bourke et al. 2000). Furthermore, the epithelium of palatal mucosa is generally thicker than in the buccal areas, and it is keratinized. All this implies that the physiological and mechanical characteristics of this tissue are quite different from buccal mucosa, which might explain differences in the outcome of the wound healing process.

Nevertheless, the general outline of both the palatal and the buccal wound healing process is similar to that of skin, which is described below (Clark 1996). It is important to stress that the phases of wound healing described here are not discrete episodes but they overlap in time (Fig. 14.1). In addition, the progress of healing in the outer wound area is more advanced than in the center, which means that subsequent phases of the wound healing process may be found in adjacent areas. This, of course, concerns only open wounds with a tissue defect in which healing takes place by second intention.

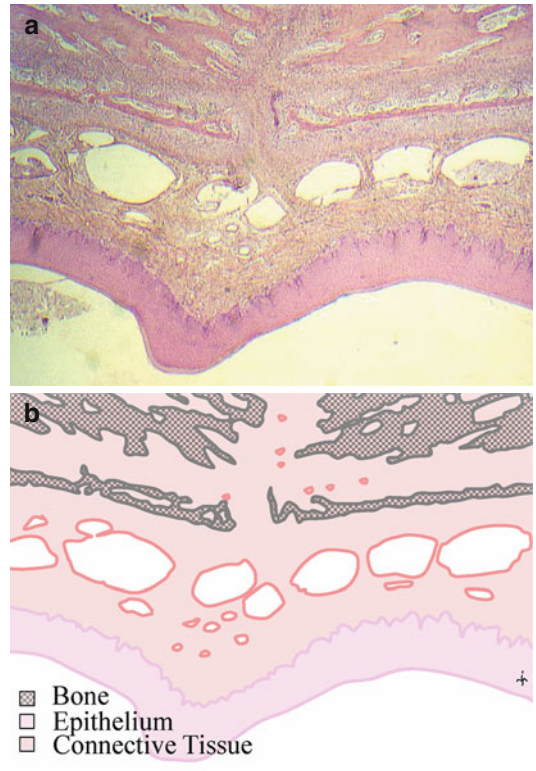

Fig. 14.2 (**a**, **b**) The palatal mucoperiosteum. The mucoperiosteum of the palate contains an epithelium and a submucosal connective tissue and is firmly attached to the palatal bone

14.2.2 Phases in Wound Healing

Tissue injury causes disruption of blood vessels and bleeding. Within seconds, the coagulation cascade starts, leading to the formation of a fibrin-rich blood clot that contains numerous platelets. These platelets are reservoirs of cytokines and growth factors such as transforming growth factors (TGFs) and platelet-derived growth factor (PDGF) that attract inflammatory cells, especially neutrophils and macrophages. The blood clot forms a provisional matrix for the migration of those cells. Proteins such as fibronectin, fibrinogen, and vitronectin allow cell attachment and migration by their interaction with integrins, which are transmembrane cell surface receptors (Yamada and Clark 1996). Neutrophils and macrophages subsequently clear the wound bed of debris and bacteria. In addition, the relative blood volume at the site of injury increases by dilation of the capillary vessels in the surrounding tissue and also by an increase of their permeability, which leads to redness and swelling. This phase is called *the inflammatory phase* and usually subsides several days after wounding (Fig. 14.1).

In the next phase, *the tissue formation phase*, keratinocytes, fibroblasts, and endothelial cells in the wound edge start to proliferate. They migrate into the wound bed and start to form the neoepithelium and the underlying granulation tissue (Clark 1996). This phase already starts a few days after wounding, before the inflammatory phase has come to an end. Keratinocytes seem to be activated by the partial loss of cell–cell contacts at the wound edge and by locally produced growth factors such as epidermal growth factors (EGFs) and fibroblast growth factors (FGFs). Fibroblasts, endothelial cells, and more macrophages migrate into the wound bed. Their migration and other activities are regulated by complex interactions with growth factors and extracellular matrix components within the provisional matrix (Yamada and Clark 1996). Again, integrins play a major role in the interaction between cells and the extracellular matrix. The binding of matrix proteins to integrins is required for attachment and migration, and it also leads to the transmission of additional regulatory signals into the cells (Rojas and Ahmed 1999). Cell migration furthermore requires the activity of matrix metalloproteinases (MMPs), enzymes that pave the way for cells by cleaving extracellular matrix proteins (McPherson 1992).

During migration into the wound, fibroblasts gradually switch to a more synthetic phenotype, a switch that involves the action of TGFβ. The fibroblasts start to produce large quantities of collagen, with collagen type III as the main species, but elastin is not synthesized in the wound. Within this matrix, endothelial cells form numerous capillaries to nourish the newly formed tissue. This process is called neo-angiogenesis and FGFs, vascular endothelial growth factor (VEGF), and many other growth factors are implicated in its regulation as shown by in vitro studies (Clark 1996). As soon as the wound is filled with granulation tissue and the neo-epidermis is formed, the collagen production is reduced, which requires interferon-γ (IFNγ). A negative feedback mechanism based on the accumulated collagen may also contribute to the decrease in collagen synthesis. This event marks the beginning of the third and last phase, *the remodeling phase*.

The remodeling phase starts within 1 week after wounding and will ultimately lead to the formation of scar tissue. The remodeling of the extracellular matrix is mainly carried out by fibroblasts. It involves the degradation of collagen type III by matrix metalloproteinases (Mignatti et al. 1996) and the concurrent deposition of type I collagen by fibroblasts. In the second week after injury, fibroblasts also start to produce proteoglycans. The mechanical properties of the tissue are not only determined by collagens but to a large extent also by these proteoglycans, since they can bind large amounts of water. In addition, many proteoglycans have been shown to regulate cell function, either by direct modulation of cell adhesion and proliferation or indirectly through the binding or release of growth factors (Nakato and Kimata 2002).

At the start of the remodeling phase, part of the fibroblasts within the granulation tissue differentiates into myofibroblasts that possess contractile properties. These specialized cells are

strongly involved in the process of wound contraction. Their differentiation seems to be governed mainly by mechanical tension within the matrix, by TGFβ and by a specific variant of fibronectin, the ED-A fibronectin (Tomasek et al. 2002). Wound contraction causes a rapid reduction of the surface area of the wound and a concomitant rearrangement of the collagen fibers. In the mean time, the neo-epidermis is maturing into a fully differentiated stratified epithelium. After 1–2 weeks, no further contraction takes place because the myofibroblasts have disappeared, probably by apoptosis (Desmouliere et al. 1995). The induction of apoptosis is not completely understood, but several genes are known to govern the process. The expression of these genes is regulated by growth factors as well as by changes in the interaction between the cells and their extracellular matrix. In the next several months, many of the fibroblasts, but also endothelial cells, disappear by apoptosis, which gradually renders the tissue less vascularized and less cell rich. The slow remodeling of the collagen fibers by remaining fibroblasts, which is part of the transition to scar tissue, can go on for a long time.

14.2.3 Contraction and Scarring

Wound contraction and scarring seem to be the two main processes in wound healing responsible for the growth disturbances after cleft palate repair and are therefore reviewed here in more detail. Wound contraction is the reduction of the wound surface area by approximation of the wound edges, and it may account for up to 80–90 % of wound closure (McGrath and Simon 1983). The evolutionary function of this feature obviously is to speed up wound closure and thereby reduces the risk of infection and dehydration. Scar formation might be a negative side effect of this primarily beneficial process.

The cause of wound contraction is not yet exactly known. Two main theories have been described in literature. The first one states that fibroblasts, which migrate from the wound margins into the wound bed, cause traction in the extracellular matrix. This tensional force would be sufficient to contract the wound (Ehrlich and Rajaratnam 1990). This theory does not require specialized cells to explain wound contraction. The second theory assumes that a specialized subtype of fibroblasts, the myofibroblast, is responsible for wound contraction (Desmouliere and Gabbiani 1996; Gabbiani 2003). During wound contraction, fibroblasts, containing intracellular stress fibers are found within the granulation tissue. These stress fibers have been shown to contain alpha-smooth muscle actin (ASMA), a cytoskeletal protein also present in smooth muscle cells. This protein seems to be required for the contraction of myofibroblasts within the granulation tissue. The coordinated contraction of myofibroblasts, attached to the extracellular matrix, causes the reduction of the wound surface.

Nowadays, the two theories have merged into a consensus theory stating that both fibroblasts and myofibroblasts are involved in wound contraction (Tomasek et al. 2002). Initially, migrating fibroblasts in the wound area generate tension within the matrix. The resulting strain within the matrix triggers the differentiation of fibroblasts into myofibroblasts, which also requires the presence of $TGF\beta_1$. The coordinated action of myofibroblasts strongly increases tension within the wound tissue, which subsequently contracts. Thus, in the consensus theory, both fibroblasts and myofibroblasts contribute to wound contraction.

During contraction of the granulation tissue, extensive collagen remodeling takes place in which MMPs play a prominent role (Mignatti et al. 1996). As a consequence of this remodeling, collagen type III is gradually replaced by collagen type I. The new collagen is deposited in an orientation that is guided by the main lines of tension within the extracellular matrix (Huang et al. 1993; Rudolph et al. 1992). The reorientation of collagen fibers and the substitution of type I collagen for type III collagen mark the start of scar tissue formation. If a uniform direction of tension exists within the contracting granulation tissue, the new collagen fibers will also be deposited in a uniform orientation. Consequently, the

resulting tissue will develop the properties of a scar, a process that may slowly progress for several months to years (Rudolph et al. 1992).

During scar tissue formation, the number of endothelial cells and fibroblasts within the developing scar tissue slowly decreases, a process in which apoptosis is involved. The final scar tissue therefore is poorly vascularized and has a low cell density. In addition, elastin fibers, which provide elasticity to normal mucosa and skin, are not resynthesized during wound healing and scar tissue formation. Their absence and the presence of highly oriented collagen fibers make the scar a rigid and stiff tissue. A specific feature of palatal wound healing is the attachment of the scar tissue to the palatal bone (see Sect. 14.4.1). This may cause palatal repair to have considerable effects on maxillary growth.

14.3 Effects of Palatal Repair on Growth

Apart from embryonic distortions and intrinsic growth deficiencies, facial growth in cleft lip and palate patients may be affected by surgical repair, orthodontic treatment, and functional adaptations (Kuijpers-Jagtman and Long 2000; Ross 1987a, b, c, d, e, f, g; Rygh and Tindlund 1982; Semb and Shaw 1996). Since the landmark studies of Graber (1949) and Dahl (1970), numerous descriptive cephalometric studies have been published (for an overview, see Semb and Shaw 1996). It is reasonably well established that cleft surgery, in particular lip and palate repair, can disturb normal growth and development of the maxilla in cleft patients (Berkowitz 1977; Kuijpers-Jagtman and Long 2000). However, the possible growth effects of surgery should be evaluated in relation to the intrinsic abnormalities of craniofacial growth in cleft palate patients. This requires that unoperated cleft lip and palate patients should be studied as well (Capelozza Filho et al. 1996; Derijcke et al. 1994; Lambrecht et al. 2000; Mars and Houston 1990).

Of all surgical procedures that are used in cleft lip and palate patients, palatal surgery has attracted the greatest amount of attention. The reason is that during this procedure, mucoperiosteal flaps are created on the palate to close the cleft, leaving areas of denuded bone. The scar tissue that is formed during healing might be a potential inhibitor of subsequent maxillary growth and dental arch development. Many studies over the past 50 years have focused on the effects of specific techniques of primary palate repair on midfacial growth and development. The effects of palatal closure seem to be mainly confined to the maxillary base and arch (Kuijpers-Jagtman and Long 2000; Semb and Shaw 1998). The maxilla is shown to be narrower, shorter, and displaced posteriorly relative to the cranial base. The dentoalveolar processes are often deflected to the median, resulting in anterior and transverse cross bites. However, since both lip and palatal surgery are generally performed, it is difficult to distinguish between the effects of the two types of surgery.

A problem with the evaluation of the literature on this subject is that the majority of the publications suffer from major methodological drawbacks, which minimizes their value (Ross 1987f; Kuijpers-Jagtman and Long 2000). Hardly, any studies are available that directly compare two types of treatment in a prospective research design. In contrast, there is a vast amount of retrospective studies available that all attempted to relate specific maxillary growth effects to particular surgical procedures. The most comprehensive study of this type is the multicenter study by Ross (1987a, b, c, d, e, f, g). By comparing lateral cephalometric radiographs collected from 15 cleft palate centers from around the world, they concluded that it was difficult to isolate the effects of individual palate repair techniques. However, an inhibition of anterior growth and translation of the maxilla was a common finding. Another problem that is only addressed by few is that not only the surgical technique but in particular the skills of the surgeon are very important for the long-term outcome in terms of growth and development (Kuijpers-Jagtman and Long 2000; Shaw et al. 2000).

In conclusion, strong consensus exists that primary surgery is a major factor in the impairment of dentomaxillary growth. The extent of

growth impairment may be influenced by the specific techniques, the timing and sequence of operations, the use of orthopedic appliances, and possibly the most important of all, the skills of the surgeon. No particular technique has been shown to produce consistently better growth results than any other. Assuming that scar tissue is a primary etiological factor in maxillofacial growth disturbances, most contemporary repair techniques attempt to minimize scarring. Animal experiments are very well suited to determine the exact effects of specific surgical procedures. For a better understanding of the biological mechanisms in wound healing, and for the goal-directed modulation of healing, experimental studies are of major importance. Results of such studies are discussed in the next section.

14.4 Experimental Research

Extensive research in animal models has been performed to evaluate the effects of cleft surgery on growth and development of the maxilla and to study the wound healing process. Animal models are also used to develop new surgical techniques that may reduce the unfavorable effects of surgery. Tissue-engineered constructs are being developed to prevent attachment of scar tissue to the bone or as a substitute for the lacking mucosal tissue. A lot of in vitro research is also aimed at the elucidation of aspects of the oral wound healing process.

14.4.1 Effects of Surgery on Growth

Since suitable animal models for congenital clefts are not available, the effects of reconstructive surgery are evaluated with surgically created clefts in dogs, rabbits, and rats. Two different approaches have been used to evaluate the effects of surgery on midfacial growth and development of the dentition. The first approach is to create a cleft in the soft tissue and the palatal bone by surgical means. This cleft is subsequently closed again as the actual experimental intervention. It is obvious, however, that a surgically created cleft is different from a congenital one. Such a cleft creates a surgical trauma that might act as a confounding factor for the interpretation of the results. Only if the bony cleft is considered to be essential for evoking the disturbances in growth and development does this approach make sense. Bardach and coworkers have used this model since 1975. They performed some of the earliest experiments on the possible negative effect of lip repair (Bardach 1989, 1990; Bardach and Eisbach 1977; Bardach et al. 1979, 1980, 1993). Lip repair in rabbits and beagle dogs with surgically created complete unilateral clefts was found to result in a significant increase in lip pressure and a corresponding maxillary growth deficiency. The authors therefore suggested a causal relationship between the two.

The second approach is based on the assumption that the soft tissue intervention is crucial for the growth disturbances after cleft palate surgery. Already in the late 1960s, Kremenak and coworkers performed mucoperiosteal excisions without affecting the palatal bone in young beagle dogs as a model for the clinical situation after cleft palate repair (Kremenak et al. 1970). This approach led to growth disturbances that were similar to those after surgical cleft closure in children. Kremenak therefore concluded that "in this model, mucoperiosteal denudation of palatal shelf bone adjacent to deciduous molars is the single surgical variable responsible for the maxillary growth disturbances seen" (Kremenak et al. 1970). This is in agreement with later dog studies in which a midpalatal soft tissue cleft was created that was subsequently closed by von Langenbeck technique (e.g., (Wijdeveld et al. 1989, 1991)). Their results not only show the same effects on growth but also that the extent of this effect is related to the age at which surgery is performed. Growth disturbances turned out to be most prominent when surgery is performed before shedding of the deciduous dentition. Furthermore, these studies, as well as recent studies in rats (Kim et al. 2002), show that the deviations in maxillary arch dimensions are not only caused by a decreased sutural growth but also by a palatal tipping of the teeth in the lateral areas. In dogs, this tipping is especially

prominent when surgery is performed at a young age, and it becomes apparent only after shedding of the deciduous dentition.

An explanation for this effect can be found in the healing of the soft tissue wounds and more specifically in wound contraction and scar tissue formation. Wound contraction in dog mucoperiosteum is most prominent in the first week after surgery. In palatal wounds in rats, it has been shown that the number of myofibroblasts increases considerably in that period (Cornelissen et al. 2000b), which also seems to be the case in dogs. Thereafter, the maturing granulation tissue is characterized by a gradual decrease in the number of fibroblasts and inflammatory cells and an increase in number and thickness of collagen type I fibers (Searls et al. 1979). Elastic fibers are not present in the granulation tissue or in the scar tissue at later stages (Wijdeveld et al. 1991).

A specific feature of the healing of open wounds in the mucoperiosteum is the deposition of callus-like cancellous bone on the palate. The granulation tissue adjacent to the palatal bone acquires an osteogenic potential, or osteogenic cells migrate into that area from adjacent periosteal tissues, and new bone is formed (Wijdeveld et al. 1991). This phenomenon is also known from other craniofacial bones and from long bones, where removal or mobilization of the periosteum leads to callus formation. Most of the collagen fibers of the scar are oriented in a transverse direction, but many fibers also show a vertical orientation. These vertical fibers become embedded in the cancellous palatal bone as Sharpey's fibers, generating a strong attachment of the scar tissue to the underlying palatal bone (Wijdeveld et al. 1991) (Fig. 14.3).

The transverse fibers appear to be continuous with the cervical periodontal ligament, thus

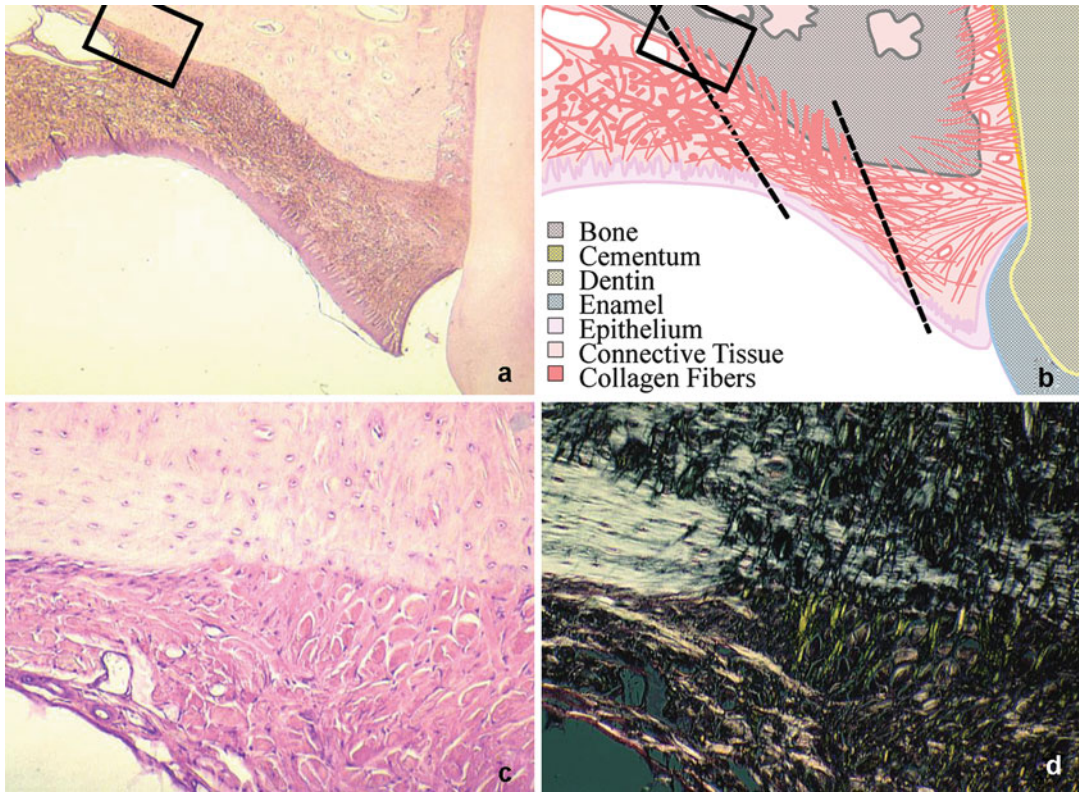

Fig. 14.3 (**a**–**d**) Palatal scar tissue. (**a**) shows the scar tissue adjacent to the teeth. The organization of the tissue is shown schematically in (**b**). (**c**, **d**) Shows an enlargement of the squared section. Note the thick perpendicular collagen fibers (Sharpey's fibers) running into the bone, which are more clear when seen in polarized light (**d**)

forming a mechanical connection between the teeth and the mucoperiosteal scar tissue (Kim et al. 2002; Wijdeveld et al. 1991). At the end of the growth period, the teeth alongside the scar tissue show a palatal tipping which is probably caused by the traction of the scar tissue on the erupting permanent dentition (Wijdeveld et al. 1988, 1989). These findings have led to the hypothesis that the iatrogenic effects of palatal surgery are initially caused by wound contraction, but scar tissue formation and the accompanying attachment of the scar tissue to the palatal bone and the teeth are probably the most important features. This leads to a restriction of maxillary growth and to a palatal tipping of erupting teeth in that region.

14.4.2 Modification of Surgical Techniques

Several researchers have tried to modify cleft palate surgery in order to avoid the appearance of denuded bone areas and the subsequent growth impairment. Perko was the first to use a mobilized mucosal split flap for palatal closure (Perko 1974). The disadvantage of his technique was the high risk of necrosis because the flap was only pedicled at the dorsal side. This technique has been modified by Leenstra et al. to obtain a flap that is pedicled both at the dorsal and the ventral side (Leenstra et al. 1995a, 1996). They used a partially split flap, leaving the lateral bone covered with the osteogenic layer of the mucoperiosteum, without impairment of the major neurovascular bundles (Leenstra et al. 1995a). In dogs, this technique was promising, since it led to less attachment of the mucoperiosteum to the underlying bone, and an improved transversal growth and development of the dentition (Leenstra et al. 1995a, b), and also in a clinical setting, the results were promising (Leenstra et al. 1996).

The denuded bone can also be covered with a biomaterial, which can either be applied as such or supplemented with cultured cells. This type of approach belongs to the field of tissue engineering.

14.4.3 Tissue Engineering

Tissue engineering has been defined in the late 1980s as "the application of principles and methods of engineering and life sciences toward fundamental understanding of structure-function relationships in normal and pathological mammalian tissues and the development of biological substitutes to restore, maintain or improve tissue functions" (Skalak and Fox 1988).

With respect to cleft palate surgery, a variety of approaches have been chosen to improve the outcome of the wound healing process. Firstly, biocompatible membranes have been used to prevent attachment of the scar tissue to the palatal bone or to reduce contraction and scar formation. A second approach is the engineering of mucosal substitutes to replenish the tissue defects. To this end, thin layers of keratinocytes have been cultured that can be used as epithelial grafts. Alternatively, keratinocytes have been cultured on top of a dermal substrate to produce a bilayered or composite graft, which is a substitute for the entire mucosa. The dermal substrate may consist of a collagenous matrix without cells or a matrix with cultured fibroblasts.

14.4.3.1 Biocompatible Membranes

Biocompatible synthetic membranes have been used to inhibit the attachment of scar tissue to the palatal bone by covering the denuded bone areas after surgery. Initially, the principles of guided tissue regeneration were used by inserting membranes in the mucoperiosteal defects (In de Braekt et al. 1992). These membranes were supposed to cover the palatal bone, inhibit osteogenic processes, and thereby prevent the formation of Sharpey's fibers. Bio-resorbable Poly-L-lactic acid membranes and non-resorbable polymer membranes yielded unsatisfactory results. This was caused by an uncontrollable degradation of the lactic acid membranes and exfoliation or incomplete coverage of the bone by the non-resorbable membranes (Leenstra et al. 1998).

Next to synthetic membranes, collagen-based membranes have been used for intraoral surgery. Atelocollagen membranes have been successfully used to improve gingival healing in a rat

model (Minabe et al. 1989). Similar membranes were used later in a model for cleft palate repair in young rabbits (Fujioka and Fujii 1997). In a split-mouth design, the membranes were implanted on the denuded palatal bone at the experimental side, while the control side was left open. The authors report that implantation reduced contraction and allowed more favorable growth of the palatal bone and normal development of the dentition.

To further improve collagen-based membranes, suitable growth factors might be added to promote angiogenesis and regeneration of the submucosa and the overlying epithelium (Nimni 1997; Jansen et al. 2009).

14.4.3.2 Epithelial Sheets

Oral keratinocytes, obtained from a mucosal biopsy, can be cultured to form an epithelial sheet. The keratinocytes are generally expanded using the Rheinwald and Green method or a modification thereof (Rheinwald and Green 1975). Grafts cultured from autologous keratinocytes seem to behave as a permanent epithelial substitute after transplantation (Bodner and Grossman 2003; Tsai et al. 1997). If allogeneic keratinocytes are used, however, the graft behaves as a temporary wound dressing that only accelerates reepithelialization (Sumi et al. 1999). Similar results were obtained after skin grafting with cultured epidermal keratinocytes. Several studies indicate that wound contraction and scar formation may still occur after the application of a cultured epidermal graft (Cooper et al. 1993; Williamson et al. 1995). In addition, the epithelial sheets are very fragile and difficult to handle.

14.4.3.3 Composite Substitutes

An approach that is more widely used nowadays was developed for the grafting of full-thickness burn wounds on the skin. In this approach, the epithelium is cultured on a dermal substrate to produce a composite graft (Pomahac et al. 1998; Liu et al. 2010). The presence of a dermal substrate is supposed to diminish contraction and subsequent scar formation. It can be derived from human dermis or prepared from purified collagen and additional extracellular matrix components. Also, fibroblasts and growth factors may be included within the dermal substrate to improve vascularization and epithelial differentiation. An overview of the production of a composite cultured graft is given in Fig. 14.4.

If dog palatal keratinocytes are cultured on skin-derived substrates, they form an epithelium similar to that in vivo (Ophof et al. 2002) (Fig. 14.5). Such

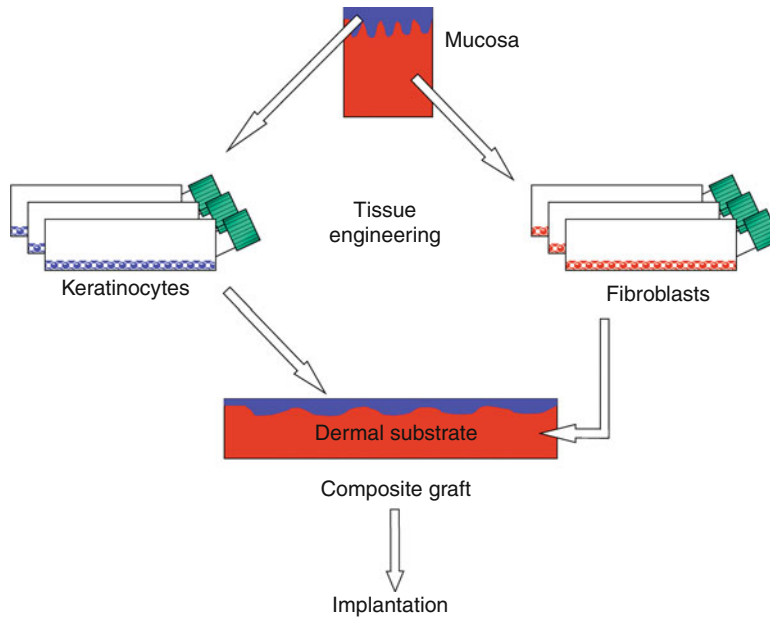

Fig. 14.4 Tissue engineering of palatal mucosa. A mucosal biopsy is taken and divided into epithelium and submucosa. Keratinocytes are cultured from the epithelium, and fibroblasts from the submucosa. The fibroblasts are seeded into a dermal substrate, and the keratinocytes are cultured on top to construct a composite graft for implantation

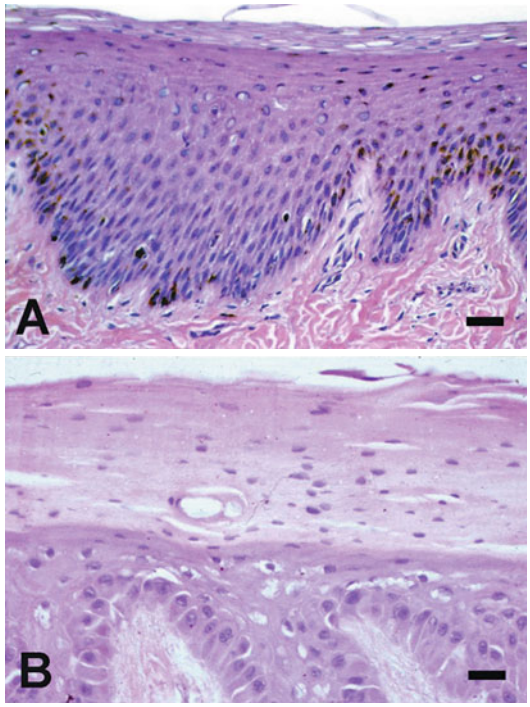

Fig. 14.5 (**a**, **b**) Tissue-engineered mucosa. Dog palatal keratinocytes were cultured on a de-epidermized dermis (DED) for 3 weeks. (**a**) Shows normal dog palatal mucoperiosteum. (**b**) Shows the cultured graft. Note that the cultured graft has a much thicker keratinized layer due to the absence of mechanical abrasion

a composite graft containing human keratinocytes showed a good clinical take rate after intraoral grafting. Compared to a substrate without cells, the composite graft showed enhanced epithelialization and maturation of the submucosa (Izumi et al. 2003). Grafts composed of a collagen substrate and human keratinocytes inside the matrix have been implanted in full-thickness skin wounds in immunodeficient mice (Butler et al. 2002). They showed a reduced wound contraction and a stimulation of epithelial maturation.

Composite grafts for skin and oral mucosa containing both keratinocytes and fibroblasts have also been constructed (El Ghalbzouri et al. 2004; Liu et al. 2008). The fibroblasts within the dermal substrate seem to enhance the differentiation of the overlying epithelium. Preliminary studies have shown that keratinocytes cultured on a collagen gel containing fibroblasts form a well-differentiated epithelium (Igarashi et al. 2003). Transplantation of such a graft into skin wounds in immunodeficient mice reduced wound contraction (Moriyama et al. 2001). Up to now, composite grafts have not been evaluated extensively for intraoral transplantation. However, their suggested capacity to reduce wound contraction and subsequent scarring makes them good candidates for application in cleft palate surgery. The use of tissue engineering techniques may well lead to the development of suitable grafts for the improvement of cleft palate repair.

14.4.4 Mechanisms of Wound Healing (In Vivo Studies)

Recent animal experiments on cleft palate surgery focus mainly on the biological processes during intraoral wound healing and the possibilities of reducing contraction and subsequent scarring. Observational research aims at a more detailed description of the cytokines and growth factors involved in the intraoral wound healing process. Most of these studies have been performed in rats. Several pro-inflammatory factors such as interleukins (ILs) are involved in palatal wound healing. For example, IL-1 seems to be essential for intraoral wound healing but not for dermal wound healing. Its effects are probably mediated by an increase of the antibacterial activity of PMNs and monocytes (Graves et al. 2001). On the other hand, oral wounds contain less IL-6 than skin wounds, while the expression of IL-10 in both types of wounds is similar (Szpaderska et al. 2003).

TGFβs and FGFs are also very important in the early phases of wound healing, as they are involved in the differentiation of myofibroblasts. $TGF\beta_1$ is supposed to upregulate FGF receptor-1 and FGF receptor-2 on palatal fibroblasts and thus increasing the susceptibility of these cells to FGFs. The latter factors are mainly produced by macrophage-like cells and cause an increase in the number of myofibroblasts (Funato et al. 1999; Kanda et al. 2003; Yokozeki et al. 1997). On the other hand, IFNγ inhibits the differentiation of myofibroblasts from rat palatal fibroblasts in vitro (Yokozeki et al. 1997) and in vivo

(Cornelissen et al. 2000a). FGF2 is also involved in the reepithelialization of palatal mucosa since a single topical application already speeds up this process (Oda et al. 2004). After the completion of wound contraction and reepithelialization, myofibroblasts disappear, probably through apoptosis that again is influenced by $TGF\beta_1$ and FGF2 (Funato et al. 1999). The same processes have been described for skin wounds, but they seem to proceed at a slower pace (Nukumi et al. 2004).

In the later phases of palatal wound healing, the number of cells and the amount of collagen are higher than in the normal mucoperiosteum. The collagen type I fibers are densely packed and transversely aligned, indicating a distinct scar tissue (Cornelissen et al. 1999) with a decreased vascular density (Chu et al. 2000). Since IFNs possess antifibrotic and anticontractive properties, they have been used for experimental research on skin wound contraction and the reduction of keloids. IFNα inhibits wound contraction (Nedelec et al. 1998), IFNβ downregulates collagen synthesis of dermal fibroblasts in vitro (Duncan et al. 1995), and IFNγ downregulates collagen synthesis in skin wounds (Granstein et al. 1989).

Strong evidence for the involvement of growth factors in the regulation of wound contraction and scar formation has been obtained from studies on fetal wound healing. Intrauterine wound healing in mammalian fetuses occurs without contraction and scar formation up to a certain gestational age (Longaker and Adzick 1991). In this early phase of gestation, myofibroblasts do not occur during wound healing, and the regenerated tissue is indistinguishable from normal tissue. This scarless healing seems to depend on a specific growth factor profile within the wound and on the typical intrauterine environment. Several researchers have utilized these characteristics of fetal wound healing to perform prenatal cleft palate surgery in animals (Kaban et al. 1993; Sullivan 1989; Zelles et al. 1995). These studies are debatable since they involve only surgically created clefts.

Recently, a model for congenital cleft palate in goats was described in which clefting was induced by the teratogen anabasine (Weinzweig et al. 2002). In affected fetuses, palatoplasty was performed in utero using a modified von Langenbeck technique at 85 days gestation. At 6 months of age, the mucoperiosteum of the palate had healed without scarring, and the function of the soft palate and the architecture of the velum were similar to that of unclefted controls. However, it is highly unlikely that prenatal surgery for cleft palate repair will soon be common practice because of diagnostic and ethical problems (Molsted 1999). The effects of specific growth factors that are possibly involved in scarless fetal healing can be further investigated by in vitro studies.

14.4.5 Mechanisms of Wound Healing (In Vitro Studies)

Several cell culture models are exploited to investigate aspects of fibroblast and keratinocyte biology that are relevant to the oral wound healing process. These studies often aim to elucidate the differences between oral and dermal wound healing or to find pharmacological means to modify the oral wound healing process. Two-dimensional monolayer cultures are suitable to study the expression of certain proteins or the proliferation and migration of cells. However, to study the interactions of cells with their extracellular matrix, a three-dimensional culture model is required. An additional advantage of three-dimensional culture models is that the physiology of the cells is more similar to that in vivo (Mio et al. 1996).

In two-dimensional monolayer cultures, human oral keratinocytes constitutively express higher levels of hepatocyte growth factor and keratinocyte growth factor than skin keratinocytes (Okazaki et al. 2002). In addition, they express higher levels of IL-6 after stimulation with other cytokines (Li et al. 1996). These differences may contribute to the preferential healing of mucosal wounds in comparison with skin wounds. For rat palatal wound fibroblasts, it was shown that interferons can reduce their collagen synthesis, which seems to be favorable in the light of scar reduction (Cornelissen et al. 1999). Oral fibroblasts have also been shown to possess

a lower migration capacity in vitro compared with skin fibroblasts (Lepekhin et al. 2002). Fibroblasts, taken from subsequent phases in rat palatal wound healing, show distinct expression patterns of integrins and cytoskeletal proteins (van Beurden et al. 2003). This indicates that specific subpopulations of fibroblasts may be responsible for wound contraction and scarring.

Fibroblasts cultured in three-dimensional collagen lattices contract this lattice in a time-dependent way (Grinnell 1994). The contraction is caused by attachment of the cells to the collagen and their migration through the lattice. This model therefore represents some aspects of the wound contraction process in vivo. Several investigators have used this model to compare the contraction capacity of oral and dermal fibroblasts (Irwin et al. 1998; Lee and Eun 1999; Stephens et al. 1996, 2001). In general, these studies show that oral fibroblasts have a higher contraction capacity than dermal fibroblasts, similar to that of fetal fibroblasts (Irwin et al. 1998). The contraction of all three types of fibroblasts is inhibited by IL-1b. Fibroblast-populated collagen lattices were also used to investigate the migration of fibroblasts into a wound (al-Khateeb et al. 1997). To this end, an experimental wound was made in the lattice, and the migration of fibroblasts into the wound was measured. Wound repopulation by mucosal fibroblasts was greater than that by age-matched dermal fibroblasts. These in vitro models contribute to the understanding of cellular processes during oral wound healing and the identification of factors with potential therapeutic value. Eventually, this may lead to new strategies to reduce intraoral wound contraction and scarring and subsequent growth disturbances.

14.5 Application of Experimental Results

The available data from clinical as well as experimental research clearly indicate that primary palatal surgery is an important factor in the maxillary growth disturbances in cleft palate patients. Experimental research in animal models shows that wound contraction and subsequent scarring in the surgical wounds are key events. More specifically, the attachment of the scar tissue to the palatal bone and the periodontal fiber system inhibits sutural growth and normal development of the dentition. This can only be partially prevented by modifications of the surgical technique such as the split flap technique, which avoids the denudation of palatal bone. Another development is the use of tissue engineering techniques to construct a substitute for oral mucosa, which can be used to cover the denuded bone areas. There are some indications that this may reduce contraction and subsequent scarring on the palate. Whether this approach will yield better growth parameters after palatal surgery remains to be established. A third approach might be the pharmacological reduction of contraction and scarring after palatal surgery. In vitro studies have shown that some cytokines have the capacity to inhibit specific aspects of scar formation such as the differentiation of myofibroblasts and excessive collagen deposition. Only few of these factors have also shown favorable effects in vivo. However, the efficient delivery of the factor into the wound and the targeting to specific cells are practical problems that have to be solved.

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Facial Growth Related to Surgical Methods

15

Gunvor Semb and William C. Shaw

For the foreseeable future, clefts of the lip and palate will continue to be managed by surgical closure and thus will be accompanied by an inescapable degree of surgical trauma. Surgery will be considered successful when, without undue burden, favourable outcomes have been achieved with respect to nasolabial appearance, speech and hearing and facial growth, thus optimising personal development and social integration.

As far as facial growth is concerned, surgically induced growth impairment has been recognised for more than 60 years (Graber 1949; Slaughter and Brodie 1949). But despite the intervening years, controversy remains about the importance of surgical technique, timing and sequence, ancillary procedures and surgical skill. And there is as much doubt about the mechanisms of facial growth in the absence of clefts, individual variability in response to surgery, the validity of theories that have determined the selection of some techniques over others and the best way to resolve these uncertainties. In this chapter, these issues will be reviewed with respect to complete unilateral and bilateral cleft lip and palate (UCLP and BCLP).

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15.1 The Characteristics of Facial Development in Individuals with Repaired Complete Cleft Lip and Palate

15.1.1 UCLP

A previous review of facial development in individuals with repaired complete cleft lip and palate revealed general consensus on the associated characteristics (Semb and Shaw 1996). Facial form in adult individuals with repaired UCLP and BCLP is similar and, in comparison with the facial form of non-cleft subjects, is characterised by a progressive retrusion of the profile relative to the cranial base involving the nasal bone, the mandible, but especially the maxilla (Fig. 15.1). Both the maxilla and mandible are shorter and retrusive, and the incisors in both jaws are retroclined. There is severe reduction in posterior but only slight reduction in anterior maxillary height. The mandible has an increased gonial angle and a steeper mandibular plane, and there is an increase in lower facial height. The bony nasopharynx is smaller. There is a tendency for facial growth to be more severely affected in males.

The pattern of growth is different from that in non-cleft individuals. Semb (1991a) found only a small increase between 5 and 18 years of age in the length of the maxilla measured to the anterior outline of the alveolar process in a mixed longitudinal study of 257 subjects with complete UCLP. There was a concomitant reduction in maxillary prominence at the dentoalveolar

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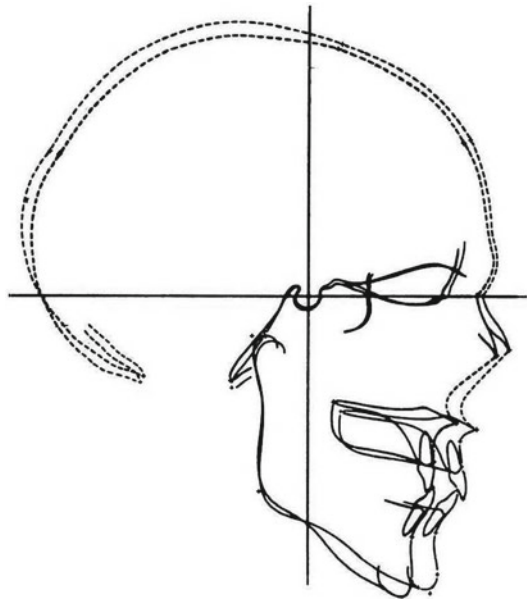

Fig. 15.1 Average facial shape in adult patients with unilateral complete cleft lip and palate compared with a non-cleft sample (Courtesy of Dr. E. Dahl, University of Copenhagen)

level, as seen in Fig. 15.2. The excessive lower face angulation changed little over time in the UCLP sample (approximately 3° greater than the Bolton standards at age 5 years), whereas it was reduced by 5° in the non-cleft sample. The gradual reduction of maxillary prominence over time and increased lower face height has also been described in several long-term studies (Enemark et al. 1990; Paulin and Thilander 1991; Smahel et al. 1993; Friede and Enemark 2001; Brattström et al. 2005; Nollet et al. 2008; Meazzini et al. 2008; Friede et al. 2011).

15.1.2 BCLP

A mixed longitudinal study of 90 patients with complete BCLP from 5 to 18 years (Semb 1991b) found the maxilla in BCLP to be relatively prominent in early childhood (4° more prominent at 5 years), but it steadily receded so that by 7 years, it was similar to the value for non-cleft subjects

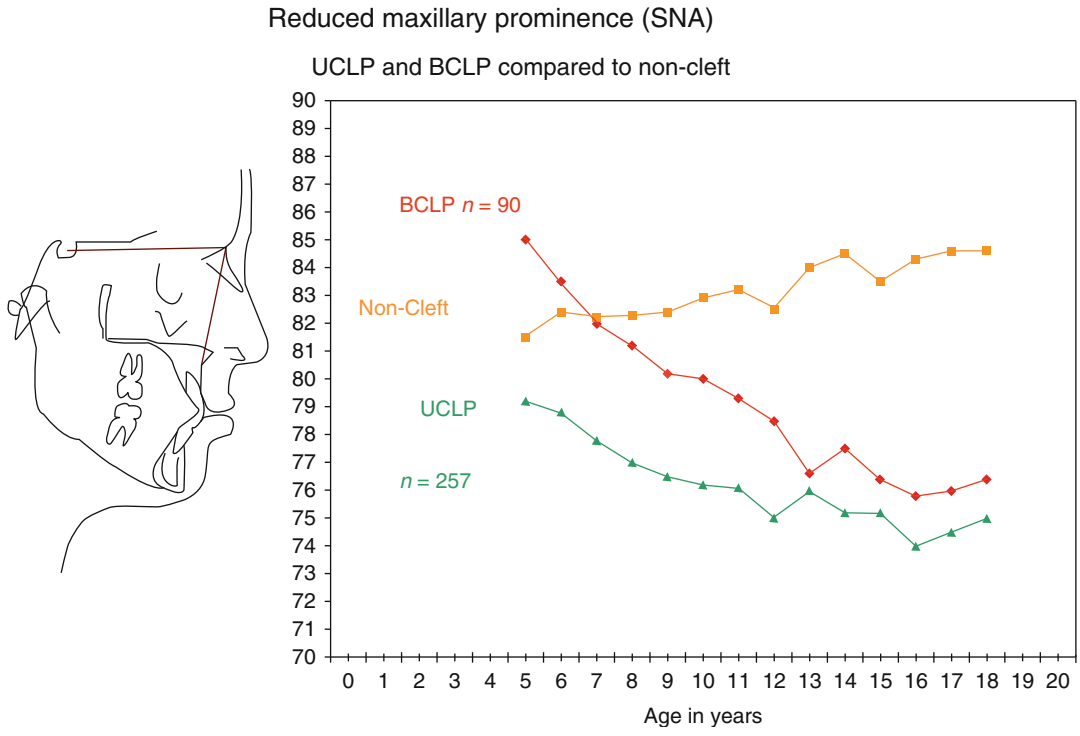

Fig. 15.2 Changes in maxillary prominence (SNA) for 257 patients with complete unilateral cleft lip and palate (UCLP) and 90 patients with complete bilateral cleft lip and palate (BCLP) compared with a non-cleft sample

(Broadbent et al. 1975) and by 18 years, it was 6° less (see Fig. 15.2). Throughout the period of observation, the mandible was less prominent (4° less at 5 years and 6° less at 18 years). Vertically, lower facial angulation remained higher in BCLP (2° at 5 years and 9° at 18 years). Similar findings have been reported (Heidbüchel et al. 1994; Gnoinski and Rutz 2009).

Thus, the growth pattern is different between UCLP and BCLP groups in one obvious respect. In comparison with UCLP patients, subjects with BCLP displayed greater maxillary prominence in early childhood (SNA was 5.3° larger at 5 years), but this difference reduced with time so that by 18 years, the maxilla was only slightly (1.4°) more prominent on average. In other respects, facial growth patterns were similar in both conditions, although the gonial angle was somewhat greater (3°) in BCLP throughout the period of observation.

15.1.3 Late Deterioration

Impairment of maxillary development continues into the late teens and early adulthood while mandibular growth continues (Enemark et al. 1990; Paulin and Thilander 1991; Semb 1991a, b; Friede and Enemark 2001; Brattström et al. 2005; Gnoinski and Rutz 2009; Myklebust et al. 2009; Friede et al. 2011; Semb et al. 2011). Thus, the anteroposterior jaw relationship worsens over time and with it the occlusion.

Since there are few publications of longitudinal follow-up beyond age 20, it is not possible to say when this growth differential ends. Enemark et al. (1990) followed 57 patients with UCLP from birth to 21 years of age. The cephalometric study showed that from 16 to 21 years, the maxillary prominence reduced by 1.1° while the mandibular prominence increased by 1.0°. The same was found in a longitudinal cephalometric study of 60 patients with UCLP from Oslo studied at 16, 18 and 21 years of age (Myklebust et al. 2009). Maxillary prominence (SNA) is reduced by 0.3° from 16 to 18 years and by 0.3° from 18 to 21 years; maxillary length (condylion to A-point) is increased by 0.6 mm from 16 to 18 years and by 0.5 mm from 18 to 21, while the mandibular length is increased by 2.3 mm from 16 to 18 years and with 1.1 mm from 18 to 21 years. A deterioration of the occlusion in the late teens/early adulthood has been reported by Lilja et al. (2006), Marcusson and Paulin (2004), and Semb et al. (2011).

15.2 Intrinsic Facial Differences Not Attributable to Surgery

It would be surprising if the catastrophic events leading to failed union of the facial processes were not associated with dysmorphology beyond the cleft site. Some indication of the extent to which the variations in facial form in UCLP and BCLP described above are intrinsic can be obtained by comparing individuals with unrepaired clefts and normal controls.

On anteroposterior cephalograms, increased width of various facial parts in unoperated infants with UCLP and BCLP has been found (Hermann et al. 2000, 2004). On lateral cephalograms, the maxilla in the UCLP group was smaller, though more protrusive at the alveolar level and at the level of the anterior nasal spine in the male subjects. The mandible was also smaller, the gonial angle was increased, the mandibular plane was steeper and the lower incisors were retroclined in the UCLP group. There was a reduced posterior height and a tendency towards increased anterior lower facial height. These differences in mandibular shape were more marked in females. The nose was less protruded, but as a result of the maxillary protrusion, there was an increase in profile convexity (Capelozza et al. 1993; Mars and Houston 1990). Differences likely to be intrinsic in nature also include abnormalities in the size and form of the cranial base (although the literature is contradictory on this point). One consistent finding is the smaller craniofacial dimensions (including size of the maxilla) found in both unoperated and operated individuals with UCLP and BCLP compared with non-cleft subjects, though this may relate to a smaller overall stature in individuals with clefts (Jensen et al. 1983).

Evidence that the prominence of the cleft maxilla is similar to that of the non-cleft is a key

issue in the debate concerning surgical iatrogenesis. Therefore, it is noteworthy that the comparisons between individuals without cleft and individuals with unoperated cleft have revealed similar anterior projection of the basal maxilla on the greater segment (Capelozza et al. 1993; Mars and Houston 1990), and the same is true preoperatively for infants with complete clefts compared to infants with cleft lip and alveolus (Hermann et al. 2000, 2004).

A variety of reports including twin and fetal studies point to intrinsically reduced nasal airway volume in individuals with clefts, repaired or not. This in turn may contribute the mandibular abnormalities that are observed (Semb and Shaw 1996).

15.2.1 Individuality of Patients

Lastly, it must remembered that children, whether or not they have clefts, tend to resemble their parents and that inheritance, especially with regard to Class II or Class III predisposition, will be an influential background factor in post-surgical facial development. Furthermore, each child entering the operating room will have other inherited traits that may impact on healing and recovery and on a wide range of later development. This individuality is a key factor in explaining the wide variability of outcome demonstrated by any study cohort and the consequent need for adequate sample size in research. As Pruzansky observed, "Within certain defined limits, the success or failure of the surgical procedure depends more on the initial state than on the variables inherent in the manoeuvre. To put it another way, we expect that subtle differences among patients will be more prognostic of the subsequent state than differences between surgeons" Nordin (1957).

15.3 Surgical Iatrogenesis

Surgery has a great impact on maxillary growth that becomes progressively apparent as patients reach maturity with reduced prominence of the maxilla at the basal (anterior nasal spine) and dentoalveolar (A point) level (Liao and Mars 2005a, b). The mandibular growth pattern is unaffected by the surgical repair of complete clefts (da Silva et al. 1992).

It is conceivable that patients with a significant deficiency of tissue are most at risk for post-operative maxillary distortion and restraint. However, the specific cause of growth disturbance remains unclear, but lip closure as well as palate closure has been implicated (Ross and Johnston 1972; Mars and Houston 1990; Normando et al. 1992).

It is possible that the iatrogenic effects of *lip* closure may have been underestimated. Experimental animal studies have reported that increased pressure from the repaired cleft lip is the primary cause of maxillary growth restraint (Bardach et al. 1984a). Lip pressure in infants with UCLP has been measured after lip repair and until 2 years of age and found to be significantly higher than in a non-cleft control group. Furthermore, follow-up of partially operated human subjects with complete UCLP and BCLP where only the lip has been repaired, in comparison to individuals with both lip and palate repair, points to the significant role of lip closure. Indeed, increased lip pressure probably continues to mould the anterior dentoalveolar region and reduce the SNA angle into adulthood.

Two studies have focused on this question on relatively large samples of subjects with UCLP (Mars and Houston 1990; Capelozza et al. 1996). The length of the maxilla (Ar-ANS) was reduced after lip surgery by 6.3 and 4.0 mm, respectively, in the Sri Lankan and Brazilian lip-only samples, an effect that appears to be largely due to moulding of the anterior alveolus (Liao and Mars 2005b). In the samples where lip surgery and subsequent palate surgery were performed, palatal surgery appeared responsible for only a small additional amount of retrusion, 1.0 mm in the Sri Lankan sample and 0.7 mm in the Brazilian sample. The same effect was seen in the prominence of the anterior alveolar process. Dahl (1970) also found a substantial reduction in maxillary prominence in patients with UCLP who had undergone lip surgery only. However, because individuals with complete clefts who have undergone only palatal surgery are not available, this experiment cannot be considered complete. It is by no means certain that the effect of palatal repair alone

would be limited to 1 or 2 mm (i.e. whichever operation is undertaken first could have the major restraining effect).

The transverse dimension of the basal maxilla does not seem to be affected by surgery, but the dental arches are highly affected. Palatal closure often includes incisions alongside the dental arches, and the scars produced may induce an inward deflection of the dentoalveolar processes, resulting in anterior and traverse crossbites (Ross and Johnston 1972; Bergland and Sidhu 1974; Dahl et al. 1981). It is therefore likely that different surgical techniques for closing the palate give rise to malocclusions of different extent without necessarily altering the neighbouring structures.

15.4 Clinical Uncertainty

Considerable ingenuity has been applied in the development and modification of primary surgical protocols for closing UCLP and BCLP. And if one takes the technique, timing and sequence of operations into account, the variations in practice between centres can be extreme. One survey funded by the European Union found that 201 cleft teams employed a total of 194 different protocols in the closure of UCLP (Shaw et al. 2001). The total number of primary operations to close the cleft varied from 1 to 4; a total of 17 possible sequences of operation for closing the cleft were practised; lip closure was performed at all possible points between birth and 12 months; the timing of hard palate closure ranged from birth to 13 years. Almost half the centres used presurgical orthopaedics. In fact, all of the 201 teams would have differed in their protocols had not seven of them previously agreed to participate in a randomised trial.

15.5 Theories That Have Influenced Surgical Management

In general, developments of new or modified surgical protocols have been founded on one or other theoretical premise, and disappointment with current results. Examples of these would include assumptions concerning the underlying mechanisms of maxillary growth, the presence and location of growth centres (to be avoided during surgery) or the role of muscular function in optimising growth potential. However, present understanding of the controlling mechanisms of facial growth is incomplete and derives from observational work and speculation in earlier decades.

However, this field of research no longer seems popular with craniofacial biologists, and understanding of the mechanisms of facial growth has not advanced in recent years. Current research mostly focuses on genetic causation, on gene environment interaction and on Hox genes, growth factors and signalling molecules that influence facial embryogenesis, providing, at this stage, few options for surgical manipulation.

Examples of *theories* that have influenced surgical management in a quest for better maxillary growth include:

- Delaying closure of the hard palate.
- "Later" lip repair may reduce early growth impairment.
- Designing flaps to minimise denuded bone, for example, avoiding pushback techniques and using minimal lateral releasing incisions.
- Avoiding use of the vomerine flap in order to safeguard septal and the vomero-premaxillary suture development and in turn growth.
- Performing extensive facial muscle dissection to optimise function and, in turn, growth.
- Reducing the extent of trauma at any one time by performance of surgical closure in small incremental stages, such as three or four operations rather than one or two.
- Assuming early benefits of repairing the alveolar cleft by bone grafting or other osteogenic technique in infancy would not harm growth.
- Assuming that presurgical orthopaedics/soft tissue moulding would facilitate surgery and improve the long-term result in better or equal growth.
- Assuming that concentrating surgery on high-volume surgeons would achieve better growth.

As we shall see below, however, most of these theories and assumptions have proved unreliable in actual practice, except possibly in the case of

the last. The following appraisal is based upon earlier reviews (Semb and Shaw 1996, 1998), three more recent comprehensive reviews (Liao and Mars 2006; Friede 2007; Yang and Liao 2010) and reports of additional studies that set out to make explicit intra- or intercentre comparisons of different surgical protocols using cephalometry and/or indices of dental arch relationship.

15.6 Clinical Evidence to Support or Refute Theories Concerning Surgery and Growth

15.6.1 Delayed Hard Palate Closure

Delayed hard palate closure is by no means a recent development having been proposed at least 90 years ago (Gillies and Fry 1921). It is still relatively popular; in the last survey, 16.5 % of European centres delayed closure till the child was older (range 3–13 years). But it has not been without critics. In their review of the rationale and supporting evidence for this procedure, Witzel and co-workers (1984) proposed that the beneficial effect on facial growth had not been proved (unless surgery is delayed until after 12 years of age) and deleterious effects on speech had been largely disregarded. Recent findings from a randomised trial confirm this risk (Willadsen 2011). By 3 years of age, the group where hard palate repair was delayed to 3 years had a more restricted phonological system and produced more cleft speech characteristics than the group repaired at 1 year.

Earlier data from single-centre studies indicate that maxillary prominence does not systematically differ between centres practicing early closure of the hard palate with a vomer flap or by different forms of one-stage palatoplasty, including push-back, and those who delay hard palate closure (Semb and Shaw 1996). Only the exceptional delay of hard palate closure in Marburg sample until mean age 13.2 years (range 8–22 years) confirmed a beneficial long-term effect on maxillary growth while assessment of speech development revealed significant speech impairment (Schweckendiek and Doz 1978; Bardach et al. 1984b).

However, earlier reports of very good maxillary growth with delayed closure (till age 8) were recently confirmed by Friede and co-workers (2011) in a cephalometric follow-up of 50 patients to age 19 and an assessment of dental arch relationship at age 19 (Lilja et al. 2006). Liao et al. (2010) also found better maxillary prominence at age 20 in a delayed closure group compared with an early closure group treated in different time periods at the same centre in Taiwan, though delayed closure was abandoned because of poor speech outcome. Noverraz et al. (1993) found no advantage for delayed closure in an intracentre comparison, while Gaggl et al. (2003) compared two groups from the same centre at 18 years, and found maxillary prominence to be worse in the delayed closure group, while another intracentre study, despite use of the original Schweckendiek protocol including obturation and hard palate closure at 7 years, reported very negatively on delayed closure as it led to severe speech problems, deleterious growth and a high rate of retrusion calling for orthognathic surgery (Holland et al. 2007). Negative speech and outcomes for delayed closure were also reported from yet another intracentre study (Rohrich et al. 2000).

*Inter*centre comparisons including at least one delayed hard palate closure group have been reported. Friede and Enemark (2001) compared growth outcome between Gothenburg and Aarhus samples of 30 patients. The first group had soft palate closure at 8 months and hard palate closure at 8.5 years (mean) while the second had anterior palate closure at 3 months (vomer flap) and soft palate closure at 22 months (pushback). Cephalometric values at three intervals from the early to mid-teens for maxillary prominence were better for the delayed closure group, though less so with age.

Swennen et al. (2002), however, found no differences in a comparison between a delayed closure and an early closure centre, nor did Stein et al. (2007) in a similar study. Comparison of delayed with early closure in the same centre revealed no difference in dental arch relationship (Noverraz et al. 1993). This was also the case in a subsequent Eurocleft comparison of records with another centre practising delayed closure (Nollet et al. 2005a). Nor did delayed closure

surpass early closure in a small four-centre Scandinavian comparison (Friede et al. 1991).

Robertson and Jolleys (1974) conducted a randomised trial involving two groups of 20 patients where one group's hard palate closure was delayed until 5 years of age. No benefit for dentofacial growth was found in delaying hard palate closure when the children reached 11 years of age.

One exception to this pattern is the report of a meta-analysis where regression analysis suggested delayed closure to be beneficial for growth (Nollet et al. 2005b). One explanation may be inclusion of a high proportion (65 %) of patients from the UK (where delayed closure is not practised), mainly drawn from a series of studies revealing historical deficiencies of cleft surgical services in general (Bearn et al. 2001).

An alternative approach to the timing question is presented by Berkowitz et al. (2005; see also Chap. 7). This multicentre three-dimensional analysis of study casts found that the velocity of growth decreases over the first 2 years and then plateaus. It was concluded that the optimal time for closure was when the ratio of the posterior cleft space to the total palatal surface area is 10 % or less, generally occurring between 18 and 24 months.

The present overview leads us to concur with the comprehensive reviews of Liao and Mars (2006) and Yang and Liao (2010) who noted that the great heterogeneity in reports precludes reliable conclusions at this time and makes the case that well-designed prospective studies are necessary to resolve this uncertainty; Friede (2007) also drew attention to the forthcoming findings of three randomised trials in the Scandcleft series (Semb 2001, described below).

15.6.2 Later Lip Closure

In truth, not much consideration has been given to the timing of lip closure, but we include it here for consideration as factor that may have been overlooked in interpreting outcomes. Bardach et al. (1984a) drew attention to the high lip pressure that follows repair, and lip surgery on its own is associated with considerable moulding of the anterior maxilla (Liao and Mars 2005a, b) and possibly restraint of the basal maxilla (Capelozza et al. 1996).

Almost one third of European centres perform lip repair at 6 months or later (Shaw et al. 2001). There are no comparative studies of early versus late lip repair that we are aware of, but it is interesting that several diverse protocols that include late lip repair have achieved good growth outcomes. These include "all-in-one repair" undertaken at 9 months (Fudalej et al. 2009), soft palate repair at 3 months followed by lip and hard palate repair at 6 months (Trotman and Ross 1993) and the Zurich protocol with lip repair at 6 months (Perko 1990). Another variation is the Gothenburg protocol where lip adhesion is done at 2 months and definitive repair is deferred to 18 months (Friede and Enemark 2001). Substantial postponement of lip closure of course may raise important sociological issues.

15.6.3 Efforts to Minimise Trauma and Scarring

It does seem probable that minimising trauma and scarring by delicate tissue handling and judicious flap design can only be beneficial and that suitable forms of tissue engineering or pharmaceutical modulation of scarring offer promise for the future (see Chap. 5). Much has been written about the harmful effect of palatal scars that form on areas of the palate denuded during primary surgery (Palmer et al. 1969; Blocksma et al. 1975), and it has been postulated especially that scar tissue located in the region of the maxillary/palatine and palatine/pterygoid sutures acts to prevent the maxilla's normal downward and forward translation (Ross and Johnston 1972). Avoidance of the vomero-premaxillary suture has also been recommended (Friede and Morgan 1976).

Ross (1987) hypothesised that early soft tissue closure of the alveolus might also lead to some growth impairment compared to those who had had no alveolar repair.

Perko (1974) developed a mucosal split flap to avoid elevation of the periosteum. A modified version of this achieved more arch depth than the

previously used Wardill-Kilner technique in a Japanese comparison (Leenstra et al. 1996). Jonsson et al. (1980) used a vomer flap covered with an autogenous skin graft in UCLP but without significant benefit. Also, no or minimal releasing incisions for closure of the palate (except in patients with very wide clefts) have been proposed (Brusati and Mannucci 1994; Sommerlad 2009).

15.6.4 Vomer Flap

Scott's theories on the primacy of the nasal septum probably did much to underpin concerns about surgical harm to the vomer, and different opinions have been held regarding the disadvantages and benefits of the use of a single-layer vomer flap to close the hard palate (Friede and Johanson 1977; Jonsson et al. 1980; Bütow and Steinhauser 1984; Delaire and Precious 1985; Friede and Pruzansky 1985; Enemark et al. 1990; Semb 1991a, b; Hay and Sommerlad 2008). However, again, the available clinical evidence contradicts the theory. As noted above, and in an earlier review (Semb and Shaw 1996), single-centre cephalometric studies incorporating vomer flap show no systematic evidence of growth disturbance, and several have some of the highest values. Likewise, in a series of different intercentre comparisons involving analysis of dental arch relationship and/or cephalometry, alternative methods have equalled but not surpassed vomer flap (combined with modified von Langenbeck posterior palate closure) samples at statistically significant levels (Friede et al. 1991; Brattström et al. 1991; Mars et al. 1992; Mølsted et al. 1992, 2005; MacKay et al. 1994; Roberts-Harry et al. 1996; Gaukroger et al. 2002; Brattström et al. 2005; Nollet et al. 2005a; Flinn et al. 2006; Meazzini et al. 2008; Fudalej et al. 2009; Meazzini et al. 2010).

15.6.5 Extensive Orofacial Muscle Dissection

Anatomical study of the orofacial and oronasal musculature led Delaire to propose extensive "functional repair" of the musculature as a solution to midface growth inhibition (Joos 1995); however, although the theory was put into practice by Delaire and others for a time, little evidence for its validity has since emerged. One single-centre study reported UCLP outcomes at a mean age 6.5 years and found outcomes better than a historical control group treated by a different surgeon (Joos 1995). No conventional cephalometric data or study cast index was reported. Others have drawn attention to the possible paradox that more extensive surgery to achieve normal anatomy may incur the penalty of greater scarring and growth restraint (Kuijpers-Jagtman and Long 2000).

15.6.6 Single or Multiple Stages of Closure

In the European survey referred to above, the total number of operations taken to complete closure of complete unilateral clefts varied from one (5 %), two (71 %), three (22 %) and four (2 %) with multiple stages often part of a delayed hard palate program. The economy and reduction of the burden of care clearly favour the "all-in-one" approach, but that could be offset with a perceived greater challenge of post-operative management and worries about iatrogenic growth disturbance, as evidenced in animal studies (Bardach et al. 1993).

Interestingly, the evidence available from intercentre comparisons suggests that varying the number of stages may actually be neutral in terms of growth. In the Americleft study (Chap. 18), Centre C who performed three-stage closure (lip, then hard palate with vomer flap, then soft palate) ranked highest for growth (Daskalogiannakis et al. 2011; Hathaway et al. 2011). An "all-in-one" sample from Poland has also compared well with mainstream staging in intercentre comparisons (Fudalej et al. 2009). De Mey et al. (2009) reported a prospective cohort study in a single centre, where a two-stage Malek protocol was compared with an "all-in-one" Malek protocol. Entry to the latter group was limited to patients with a posterior palate width less than 10 mm. Both groups were followed till age 10 and 15 and had good cephalometric outcomes.

15.6.7 Primary Bone Grafting

Initial reports of successful bone grafting at the time of lip closure (Schmid 1955; Nordin 1957) led to its adoption in several centres. This was followed however by a steady process of abandonment in the subsequent decades and a series of reports mostly describing negative impact on maxillary growth (Rehrmann et al. 1970; Friede and Johanson 1974; Robertson and Jolleys 1983; Pfeifer 1986; Reichert and Manzari 1990; Lilja et al. 1996; Smahel et al. 1998) but with some exceptions (Rosenstein et al 2003; Dado and Rosentstein 2009). Intercentre comparisons also highlighted growth problems (Ross 1987; Brattström et al. 1991, 2005; Trotman et al. 1996), most recently within the Americleft study where the only centre performing primary bone grafting ranked poorest overall for maxillary growth (Hathaway et al. 2011).

15.6.8 Presurgical Infant Orthopaedics (PSIO)

This topic is the subject of another chapter, so will be referred to briefly here. Two sources provide comprehensive overview of the relevant literature (Kuijpers-Jagtman and Long 2000; Uzel and Alparslan 2011). Originating in the late 1940s, PSIO continues to be popular despite, rather than because of, the evidence. When reports about a particular technique provide no evidence of benefit, adherents are entitled to say that this does not amount to evidence of no benefit. However, in the case of PSIO, the Dutchcleft randomised trial of passive orthopaedics *does* provide good evidence of *no* benefit at 4 and 6 years in respect of maxillary development and dental arch relationship (Bongaarts et al. 2009), raising doubts about the ethical status of PSIO.

On the other hand, of the clinical reports of active orthopaedics with the Latham appliance listed by Uzel and Alparslan, five of six reports suggest actual harm in respect of facial growth (Roberts-Harry et al. 1996; Henkel and Gundlach 1997; Berkowitz et al. 2004; Matic and Power 2008; Power and Matic 2009). Only one report found no growth harm (Chan et al. 2003). Apart from the study of Roberts-Harry et al. (1996), the use of active orthopaedics and gingivoperiosteoplasty were combined (discussed below).

15.6.9 Nasoalveolar Moulding (NAM)

Though it has much in common with active PSIO in its intended reduction of the alveolar cleft gap to allow for gingivoperiosteoplasty (GPP) repair of the infant alveolar cleft, nasoalveolar moulding (NAM) also has explicit goals of alignment and correction of the deformity in the nasal cartilages and non-surgical elongation of the columella. If successful, it should obviate the need for secondary nose/lip revision and alveolar bone grafting (Grayson et al. 1999).

Beneficial short-term effects of NAM on nasolabial form and columella length have been demonstrated in technique papers and reports (Grayson et al. 1999; Grayson and Cutting 2001) and small retrospective case series (Wood et al. 1997; Grayson et al. 1999; Maull et al. 1999; Singh et al. 2007; Lee et al. 2008; Barillas et al. 2009). Most of these studies report improved nasolabial outcomes using a variety of assessment methods, though Liou et al. (2004) observed some lack of stability and Chang et al. (2010) considered further surgery necessary.

Unfortunately, in the two decades since its introduction, there have been no substantial reports of consecutive patients nor prospective studies of the influence on facial growth. Wood et al. (1997) compared 11 patients who received NAM and GPP with 9 who received NAM alone and found them not to differ in a cephalometric shape coordinate analysis. A subsequent follow-up of the same groups with 8–13 years old found reduced maxillary prominence (position of ANS) that did not reach statistical significance (Lee et al. 2004), while need for secondary bone grafting was estimated at 40 % in a sample of 20 sites with previous GPP (Santiago et al. 1998).

Preliminary intercentre comparisons in the Americleft project raise some doubts about the consistency of outcomes (Chap. 18).

15.6.10 Gingivoperiosteoplasty (GPP)

In 1965, Skoog introduced periosteoplasty as an alternative to bone grafting: by using double-layer periosteal flaps, bone formation across the cleft was induced without the need of a donor site. The periosteoplasty technique was adopted

by a few teams, and in the 1990s, the technique was modified and popularised by Brusati and Mannucci (1992) and Cutting and Grayson (1993) as gingivoalveoloplasty. Normally accompanied by active presurgical orthopaedics or nasoalveolar moulding, the evidence required to validate it as a desirable routine intervention will have to confirm that it matches mixed dentition bone grafting in quality and reliability, that the additional burden imposed by the presurgical preparation offsets the burden of mixed dentition bone grafting and that the risk of requiring later orthognathic surgery is not increased.

Concerns about adequacy of the bone in the former cleft alveolus and subsequent maxillary growth have been reported. With respect to Skoog's original procedure (Skoog 1965) and a free tibial periosteal graft variation, Hellquist and Svardström (1990) found good bone formation in only 47 % of patients, Rintala and Ranta (1989) found 72 % required secondary bone grafting and Renkielska et al. (2005) found 50 % of patients operated according to the Skoog protocol-required orthognathic surgery.

Millard adopted the active pin-retained or "Latham appliance" as a means of reducing the size of the cleft by aligning the alveolar segments prior to surgery (Millard and Latham 1990). However, this led to an increased level of anterior crossbite in unilateral and bilateral cases (Millard et al. 1999), and Berkowitz et al. (2004) reported a likely need for orthognathic surgery in the majority of patients. Similar conclusions were reached by Henkel and Gundlach (1997), who followed 60 Latham appliance/GPP patients with UCLP or BCLP and found substantial horizontal and vertical maxillary growth disturbance compared with a similar group treated without Latham appliance GPP. The success of grafting was not reported.

Two Canadian studies compared, respectively, 54 UCLP Latham appliance/GPP patients with 16 UCLP mixed dentition alveolar bone graft patients and 53 BCLP Latham appliance GPP patients with 10 BCLP secondary bone graft patients in their mid-teens (Matic and Power 2008; Power and Matic 2009). Both revealed considerably fewer successful grafts, more fistulae and disturbed growth in the Latham/GPP groups. Interestingly, no rebuttal of these critical reports was published by the originators of the protocol, and a subsequent report of BCLP patients treated by the Latham appliance and GPP confirmed the negative experience of others (Latham 2007). The author proposed an alternative method of palate closure and the postponement of definitive lip repair till 18 months to overcome this (Latham 2007).

It is of course difficult to distinguish between the Latham appliance and gingivoperiosteoplasty as the source of growth inhibition, and the advent of NAM was regarded by some authors as a superior preparation for GPP since it is "a passive process that directs the growth of the alveolar segments" (Hopper and Birgfield 2009). Hsieh et al. (2010) found maxillary growth at age 5 to be worse with GPP and NAM than with NAM alone. Long-term follow-up of a GPP variant postponed till the time of palate repair at 18–36 months indicated a higher need for orthognathic surgery in comparison with a sample from the same centre and a sample from another centre who received conventional mixed dentition bone grafting (Meazzini et al. 2010).

15.6.11 High-Volume Surgeons

It is self-evident that regardless of the protocol selected, the end result will benefit from skilful performance of the surgery, and there is ample evidence from the findings reported above that surgeons who have selected the same protocols may achieve remarkably different results. Differing interpretations of the detail of what should be done to execute a procedure that has been described or even demonstrated by a different surgeon may be a factor in explaining different outcome; differing stages of progress along a learning curve of a new procedure may be another factor. However, the individual intrinsic skill brought to the operating room, be that dexterity, delicacy, lightness of touch or whatever, seems crucial if impossible to quantify.

An investigation of factors influencing the success of alveolar bone grafts on 825 cleft sites treated at the same centre examined 22 possible

Table 15.1 Significant differences between high- and low-volume operators

Variable	5-year-olds	12-year-olds
Intelligibility[a]	–	$p<.001$
Hypernasality[a]	$p<.05$	$p<.001$
GOSLON	–	–
Fistula with symptoms[a]	$p<.05$	–
Nasal appearance[a]	$p<.05$	–
Lip appearance	–	–
Profile appearance	–	–

[a]Better outcome was achieved by high-volume operators

explanatory variables regarding patient age, cleft type and related features. In a multiple regression analysis, the most influential single factor was the surgeon performing the graft (Semb et al. 2003). In general, it is possible that surgical skill is more influential than the choice of protocol.

Other than ensuring the completion of recognised training pathways, there is not much that can be done to assure high levels of performance. One possible step might be to require that a cleft surgeon has sufficient regular practice of cleft surgery to maintain competence. This was explored in the national review of cleft care standards carried out in the UK, and outcomes for surgeons who had 30 or more new referrals for primary surgery per year were compared with those who had less and found to have better outcomes in some measures, whereas the low-volume operators never had better outcomes (Bearn et al. 2001; Table 15.1). Following restructuring of national services on regional centres, improvements in outcome for dental arch relationship have been demonstrated (Russell et al. 2011). Volume *per se* provides no guarantee of course, and the worst case scenario would be when a high-volume surgeon is unskilful. At least high-volume surgery makes it much easier for the surgeon to audit outcomes.

15.7 Bilateral Cleft Lip and Palate Closure

A number of single-centre studies have been reported, but since complete BCLP is less frequent, occurring in around 7 % of individuals with clefts (Sivertsen et al. 2008), these are fewer and often include small samples. A previous review identified 15 case series with samples ranging from 90 to 7 cases (Semb and Shaw 1996).

The sample with 90 complete BCLP patients followed longitudinally provides a reference frame for comparison (Semb 1991b), its protocol involving two-stage lip repair and a vomer flap to close the hard palate at 3–4 months of age, and subsequent posterior palatal closure by a modified von Langenbeck technique at 18 months. Comparable values for maxillary prominence were attained by several other protocols including a Czech group of 26 males whose protocol included palatoplasty with pushback and primary pharyngeal flap at a mean age of 5.8 years and premaxillary setback surgery in one third of cases (Smahel 1984); three groups from the same Swedish centre, two of whom underwent periosteoplasty (Hellquist and Svardström 1990); an American group who underwent primary bone grafting (Rosenstein et al. 1991); and a centre that used presurgical orthopaedics and delayed hard palate closure (Heidbüchel et al. 1994; Gnoinski and Rutz 2009.) More favourable values were achieved by a Canadian sample whose protocol included vomerplasty (Trotman and Ross 1993). Melissaratou and Friede (2002) also reported better occlusion in a comparison of 16 patients after delayed closure with 12 with early closure, treated at the same centre, though there were few statistically significant differences.

Formal intercentre studies are scarce, but a large longitudinal study including 204 patients from three centres in the Netherlands, Norway and Sweden has recently been reported (Bartzela et al. 2010). Despite the use of quite different protocols, dental arch relationships for patients in the three centres were similar at age 9 and 12 years. Delaying hard palate closure and employing infant orthopaedics did not appear advantageous in the

long run, at least for the outcome studied in this investigation. Premaxillary osteotomy at 8–9 years appeared to be associated with less favourable development of dental arch relationship between 9 and 12 years. These findings were confirmed in a later cephalometric study (Bartzela et al. 2012).

15.7.1 Surgical Setback of the Premaxilla

The premaxillary setback procedure is an especially controversial issue, but the findings from two centres in which it has been practised selectively indicate a tendency for more growth impairment in the setback groups (Friede and Pruzansky 1985; Bardach et al. 1992), confirming the experience of Bartzela et al. (2010). Bardach et al. (1992) also report and highlight a risk of devitalisation and loss of the central incisors with setback around age 5.

15.8 Reliability of Evidence

15.8.1 Retrospective Comparison Studies

On the basis of the information identified in this chapter, it is clear that confusion, uncertainty and controversy still beset the choice of methods of surgical closure. The bulk of the evidence base currently comes from retrospective individual centre reports that are commonly limited in sample size. However, formal sample size calculations for prospective studies generally require quite large samples. For example, two groups of 75 in each of the Scandcleft trials of different surgical protocols and two groups of 325 in the TOPS trial of timing of cleft repair (NIH 2010) were estimated as the required sample. Few of the retrospective comparisons described in this chapter approach these levels.

Statistical comparison of different reports is often not possible, and the common biases that apply to comparison studies in general (Table 15.2) are generally more problematic when reports from different centres are put side by side. These reports may well have different case mixes, different measurement techniques, different follow-up and exclusion protocols and of course different surgeons. And, in the first place, findings that have proved disappointing may not have been reported at all, distorting the overall picture.

15.8.2 Intercentre Comparisons

Prospectively, planned recall of patients at participating centres allows data on outcome to be collected in a more standardised way, and rigorous planning and execution across centres can ensure consecutive patient inclusion and consistent eval-

Table 15.2 Potential sources of bias in cleft research

Source of bias	Example
Case mix bias	Comparisons of facial growth data may be dubious where there are inherent differences in facial form between communities. Or where differences in referral patterns in respect of case difficulty occur
Proficiency bias	The skill of a more gifted surgeon or clinical team can inflate the apparent effectiveness of the technique. If operator A is 10 % better than operator B and technique X is 5 % better than technique Y, a false conclusion will be reached in a comparison of technique Y performed by A versus technique X performed by B
Follow-up bias	Without knowing about all the cases on whom a particular technique was tried, reliable conclusions cannot be drawn. Follow-up should be equally rigorous for cases that went badly
Exclusion bias	Irregular application of grounds for retrospective exclusion, for example, "uncooperative" or "did not really fit the criteria", can remove any equivalence that comparison groups may have had
Analysis bias	When raters are not blinded to the treatment allocation of patients, or simply interpret cephalometric x-ray features or apply indices of dental arch relationship differently
Reporting bias	When negative or disappointing findings remain unpublished

uation (Chap. 18). Of course, these studies are comparing the protocols *and* the level of skill with which they were executed, as well as other aspects of the treatment program. None the less, they provide a valuable opportunity for centres to appraise their standards of care, to reconsider choice of protocol, to build research partnerships and to stimulate prospective collaborative research (see Chap. 18). Additional centres can also "participate" in these comparisons at a later date provided they follow the same methodology in partnership with members of the original group; they can also obtain similar benefits.

15.8.3 Intracentre Comparisons

Non-randomised *intracentre* studies potentially avoid several biases (the surgeon, case mix and other background variables may be the same), though comparisons between groups treated in different time periods can still be distorted (WHO 2002).

15.8.4 Prospective Cohort Studies

These occupy an intermediate position between non-randomised studies and randomised control trials. As records of all consecutive cases are filed with the study registry prior to the start of treatment as well as after it, justification for non-follow-up would be required. As in well-conducted clinical trials, analysis bias can be overcome by employing blinded independent raters, while reporting bias should be overcome by the greater impartiality of the partnership and its predetermined conventions. Case mix bias and exclusion bias cannot be minimised with the assurance derived from random allocation, but some checks of equivalence might be possible. Clinical proficiency however would inevitably remain as a major bias.

15.8.5 Randomised Control Trials

For the fairest comparison of therapies, there is little doubt that the randomised controlled trial is generally the method of choice, scientifically and ethically. Prognostic factors, including clinical proficiency, whether known or unknown to the investigator, tend to be balanced between treatment groups. Since patients are registered prior to treatment and followed up prospectively according to a clearly defined protocol, missing data are less likely as the potential loss to follow-up and late exclusion is reduced. Formalising the protocol at the outset, as required by an ethical review board or funding agency, increases the likelihood of consistent record collection and impartial analysis. The likelihood of reporting the results regardless of the findings is also increased by the conventions of trial registration and funding.

However, randomised trials are not for the faint hearted! Trials that compare treatments that are currently in common use must comply with the same codes of governance concerning the integrity of data, ethics and confidentiality required for high-risk drug trials and so involve a high level of bureaucracy and administration. There are efforts to correct this (Academy of Medical Sciences 2011). If the challenge is met, they remain the surest way of singling out the influence of surgical protocols on facial growth and other outcomes. They are feasible in large centres or in large collaborative groups, but they require high levels of goodwill and commitment.

15.8.5.1 Scandcleft Trials

Ten centres in Denmark, Finland, Norway, Sweden and the UK are currently participating in a set of three randomised trials where groups of teams are testing their traditional local protocols against a common protocol (Semb 2001). The sample size requirements of 450 infants with UCLP, 150 per trial, were attained, and the planned initial follow-up to age 5 has been completed. The principle outcomes are speech and dentofacial relationship at age 5 years. Only 7 % of the parents declined participation in the trials, and follow-up rate at 5 years is 96.5 %. In each trial, each surgeon has performed the two trial protocols on an equal number of infants.

Three groups of centres are testing their traditional local protocols against a newly defined common method. In the common method, the lip was repaired simultaneously with soft palate

closure at 3–4 months, followed by hard palate closure at 12 months. In Trial 1, this was compared with hard palate closure delayed till 3 years. In Trial 2, the common leg was compared to closure of the lip at 3–4 months followed by closure of the hard and soft palate together at 12 months. In Trial 3, the common leg was compared to lip and hard palate closure at 3–4 months followed by soft palate closure at 12 months.

At the time of writing, the principal outcomes, speech and dentofacial development are being analysed. It is hoped that the results will shed light on the delayed closure controversy, the influence of variations in sequence of closure, the influence of learning curve and the influence of surgical skill.

15.9 Conclusions

15.9.1 Craniofacial Development

Craniofacial development associated with cleft lip and palate and the impact of surgery have been well described in the literature. But mechanisms that can explain normal and post-surgical development and how surgery interferes with growth are not well understood. Progress in these areas has been negligible in recent decades as craniofacial biology has shifted focus from structural histology to aetiology and the role of signalling molecules and growth factors in orofacial embryogenesis.

15.9.2 Limitations of Current Research

Clinical research in cleft care is challenging. Low prevalence and the variety of cleft subtypes make it difficult to gather adequate samples, collaborative working between cleft centres and nations has barely begun and the timescale of most projects is daunting. The font of current knowledge is largely composed of potentially flawed, underpowered retrospective studies. Not surprisingly, this overview has found no good evidence to resolve the counter claims reviewed in Sect. 15.6.

15.9.3 Influence of Surgical Skill

A recurring theme arising from the slightly more dependable intercentre studies cited in Sect. 15.6.4 is that satisfactory growth outcomes can be achieved by very different protocols. These include one-stage closure, three-stage closure, use or non-use of vomer flaps, delayed or early hard palate closure, use or non-use of presurgical orthopaedics and highly demanding or simpler protocols. All this strongly suggests that surgical skill may be the primary factor in determining ultimate success.

15.9.4 Making a Choice

So there is much to be said for a centre adhering to a familiar surgical protocol where satisfactory results across the range of relevant outcomes have been confirmed by critical evaluation, such as that gained by participation in a formal intercentre study. On the other hand, if a centre's current protocol is especially burdensome, it would be reasonable to consider changing to a well-proven mainstream protocol that is easier on the child and family.

Where a change is being considered or a new service is about to be established, the literature reviewed in this chapter could be summarised as follows:

Delayed hard palate closure: There is no evidence that delayed closure systematically surpasses early closure in achieving satisfactory growth results and, given the potential harm to speech development, may not be justified.

Efforts to minimize trauma and scarring: There is inconsistent evidence regarding techniques that may leave large areas of denuded palatal bone and restrain growth. However, it would seem wise to adopt mainstream methods designed to limit scarring.

Vomer flap: Theoretical objections to vomerplasty have been repeatedly contradicted by intercentre and other studies. It deserves its place as a safe mainstream method in UCLP and BCLP.

Extensive orofacial muscle dissection: There is no evidence that this procedure is beneficial to growth.

Single-stage or multiple-stage repair: One, two or three stage and closure of UCLP are compatible with good growth, and the burden of care should be considered.

Early bone grafting: The balance of evidence is firmly against early bone grafting.

Presurgical infant orthopaedics: This is neutral to growth and other outcomes. Rejecting it will reduce the burden on the child and family.

The Latham appliance: This is probably harmful to growth, and it certainly would ease the burden on the child and family to abandon it.

Nasoalveolar moulding: Despite its usage for almost two decades, there is inadequate evidence to judge whether its benefits outweigh its disadvantages. Proper research is overdue.

Gingivoperiosteoplasty: There is some evidence that GPP presents a risk to growth, and little evidence that it reliably obviates the need for later grafting.

BCLP surgery: Diverse protocols can achieve satisfactory growth outcomes, and it appears appropriate to select one that minimises the burden on child and family.

Setback of the premaxilla: There is no evidence that this technique is justified, but some evidence that it is harmful.

High-volume surgeons: Confining surgery to high-volume surgeons provides some assurance of better outcomes and allows meaningful clinical audit to be conducted over a shorter time span.

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Part VI

Lip and Palate Surgery: Millard–Berkowitz Protocol

The Influence of Conservative Surgery on Growth and Occlusion

16

Samuel Berkowitz

The collected serial data will provide the clinician in training with an overview of the variations that can be encountered in each cleft type, the significance of genotype differences that influence growth and response to surgery, and the natural history of each cleft entity.

Serial records of cleft lip and/or palate provide useful data in studying morphologic growth changes in the head (evaluating dentofacial abnormalities) and assessing responses to surgical and orthodontic treatment. The data have been particularly useful in determining the timing and type of procedure selected to treat individual problems. The measurements and analyses utilized are primarily profile-oriented and reveal both anteroposterior and vertical relationships of the various parts of the dentofacial complex.

The anteroposterior and vertical growth of the midface is not yet completely understood. The stimulus or growth force is still being debated. However, it is essential to understand the basic maxillary growth concepts to appreciate the influence of palatal and facial surgery on midfacial growth.

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Enlow (1982) states that:

> Growth is not "programmed" within the calcified part of the bone itself. The "blueprint" for the design, construction, and growth of a bone lies in the functional matrix: the composite of the muscles, tongue, lips, cheeks, integument, mucosa, connective tissue, nerves, blood vessel, airways, pharynx, the brain as an organ mass, tonsils, adenoids, and so on. Growth fields (growth sites) for example, a suture and the alveolar bone housing teeth, throughout a bone do not have the same rate of growth activity. Some "fields" grow much more rapidly or to a greater extent than others. The same is true for resorptive fields. All surfaces are sites of growth; relocation of the bone, going from one location to another, is the basis for remodeling.

In the maxilla, the palate grows downward (i.e., becomes relocated interiorly) by periosteal resorption on the nasal side and periosteal deposition on the oral side. The nasal mucosa provides the periosteum on one side, and the oral mucosa provides it on the other side; together, growth remodeling is paced by the composite of soft tissue housing the palate. This results in a downward relocation of the whole palate-maxillary arch composite.

The bone tissue that comes to house the teeth at older age periods is not the same actual bone enclosing them during the succession of former growth levels.

Depending on the extent of palatal surgery and the resulting scar tissue, the stability of a region can be disrupted if the results negatively affect the pattern of resorptive and depository fields on bone and at the suture; that is, scarring can work against growth. As the midface grows, bone is laid down in the sutures surrounding the maxillary complex. Any

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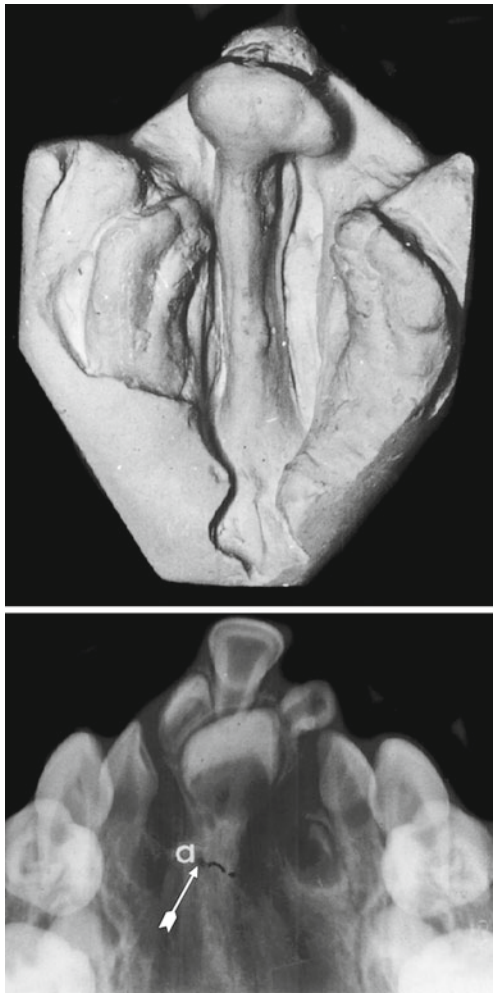


Fig. 16.1 Complete bilateral cleft lip and palate cast (*top*). Occlusal x-ray film (*bottom*). '*a*' indicates premaxillary vomerine suture (*PVS*)

damage to one of these sutures can interfere with the direction and amount of growth. For example, an excessive amount of scarring at the pterygomaxillary suture (PTM), or at the premaxillary vomerine suture (PVS), will interfere with anteroposterior and vertical maxillary and premaxillary growth (Fig. 16.1).

Growth movement of the premaxilla is produced by the growth expansion of all the bones behind and above it and by growth in other parts of the maxilla, especially at the premaxillary vomerine suture (PVS). The premaxilla itself contributes a major part of its own forward growth movement through changes at the PVS. These displacement growth movements are a result of the "carry effect," as Enlow calls it, which is produced by the expansion of the soft tissues associated with the bones, not a "pushing effect" of bones against bones. Scarring of the palatal mucoperiosteum, therefore, acts to interfere with the "carry effect," thus preventing the change in position of the maxilla within the face.

Berkowitz speculates that pressure forces created by the uniting of the cleft orbicularis oris are directed posteriorly through the maxilla and nasal septum to the maxillary sutures and premaxillary vomerine suture, respectively. This action is similar to that of a headgear (cervical traction) used to control midfacial growth in orthodontics. These forces interfere with the ability of the premaxillary vomerine suture and the pterygomaxillary suture to function properly and lead to some degree of midfacial retrusion. Berkowitz's clinical findings support the concept that maxillary growth can be restrained by a variety of force systems. Marked deformation and gross changes can result when large forces are used over long periods of time. Maxillary maldevelopment is three-dimensional, resulting in a decrease in midfacial height, length, and width.

16.1 Surgical Closure of the Cleft Lip and Palate

Enthusiasm for a particular form of therapy should not be regarded as scientifically established when, in fact, it has not been subjected to critical scientific analysis. Treatment fads come and go. Unfortunately, in cleft palate surgery, it takes at least a decade to determine the effectiveness of a procedure. A prior belief in a particular therapy often determines the biased selection of evidence to support a concept currently in vogue; some clinicians show a select sample of cases to prove the theory only in part, if at all.

The following chapters will present our understanding of the cleft defect and the face in which it exists. These chapters are designed to answer some basic questions: What is the natural history of the cleft defect? How do similarly classified clefts differ from each other? What should be done for a child with a cleft lip and palate, and when should it be done? And finally, how does the treatment vary from child to child?

Although selected cases will be shown to develop our treatment philosophy, it must be

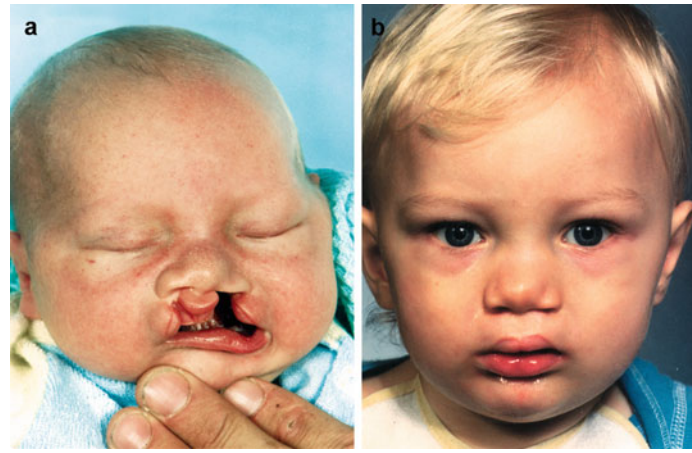


Fig. 16.2 (**a**, **b**) Incomplete bilateral cleft lip and palate. (**a**) Presurgery. (**b**) 1 year and 6 months (1–6) after the lip is united. The prolabium extends to the vermilion borders

stressed at the start that the concepts being presented are supported by findings from many longitudinal facial growth studies already published by our clinic and others worldwide. The cases from our center are unique to the extent that they represent the results of only one surgeon who is considered a master at his craft. Treatment failures as well as successes will be presented to develop and stress the physiological principles that are the basis of this treatment philosophy.

Improvements in surgical and orthodontic treatment methods, coupled with a better understanding of the natural history of cleft palate growth and development, have led to different yet frequently successful treatment protocols. Although some readers may be concerned that variations in successful treatment strategies interfere with comparisons and eventual standardization of treatment outcomes, nothing could be further from the truth. The one conclusion that should be drawn from these reports of clinical innovations is that there is more than one successful treatment protocol.

If the sample populations are large enough, any statistical evaluation of a generally successful treatment approach will show a certain proportion of failures. Obviously, a number of different physiological surgical procedures can be successful, provided that other dependent variables, such as the facial growth pattern and geometric form of the cleft defect, are complementary.

There is no reason to think that, by standardizing treatment protocols (as to type and timing), the same surgical procedure invariably will be successful. Certainly the skill employed is equally important. Moreover, it is a well-known fact that the same surgery, performed by the same surgeon on the same cleft type, can yield different results. Why? To find an answer to this question, it is necessary to change one's primary focus from solely the surgery employed to the cleft defect itself and the face in which it exists. Doing so enables the clinician to take into account three mutually dependent critical variables: (1) the cleft defect, (2) the facial growth pattern, and (3) the surgical procedure (see Chap. 16, Changing Philosophy of Surgery of the Cleft Lip and Palate in Goteborg, Sweden).

16.2 Lip Surgery

In a comprehensive evaluation of maxillary growth and development, the principles and techniques of tissue repair are of paramount importance as an integral part of any maxillary surgery. The basic requirement of any soft tissue closure, whether in unilateral or bilateral clefts of the lip, is that the full width of the lip be maintained. Clinical observations have shown that a moderately tight lip will hamper, to some extent, maxillary growth and produce some deformity with subsequent malocclusion of the permanent dentition. Skoog (1974) has written that, in the cleft deformity, there is a variable but absolute tissue shortage. He concludes that, to be effective and achieve an anatomically correct and aesthetically pleasing result, reconstruction must therefore preserve and utilize all available tissue. Millard (1980) has stressed that, in bilateral clefts of the lip, the prolabium – regardless of its size – should always be positioned to the vermilion border (Fig. 16.2).

16.2.1 Lip Adhesion

The cleft lip can be closed at any time from the first day of life, but the rule of "over ten" is usually followed. At 10 weeks of age, 10 lb, and 10 g of hemoglobin, elective surgery is generally considered safe, and the lip and nose have increased enough in size to facilitate the detailed surgery. Thus, most surgeons recommend surgical repair of the cleft lip at about 3 months of age.

Incisions in the sulcus and undermining of the soft tissues on the external surface of the maxillary segments were once commonly used to facilitate lip closure and decrease lip tension, especially in wide clefts (Fig. 16.3). Walker et al. (1966), Bardach and Eisbach (1977), Bardach et al. (1979, 1982), and Collito (1974) questioned the advisability of performing this procedure because they observed clinically that it contributed to secondary maxillofacial deformities and midfacial growth inhibition because of excessive scarring. This objection led Walker et al. (1966) to present their concept of the Collito–Walker (C-W) technique of uniting the cleft lip (i.e., the use of lip adhesion without undermining; Figs. 16.4 and 16.5). Bardach et al. (1982), using rabbits, showed that lip repair with soft tissue undermining led to significant shortening of the maxillary complex. There is no reason to believe that the same negative effect would not also occur on children with lip/alveolar clefts, and therefore Bardach et al. (1982) concluded that the lip adhesion procedure is preferred even though the posterior-directed pressure of the united lip potentially can slow down midfacial growth.

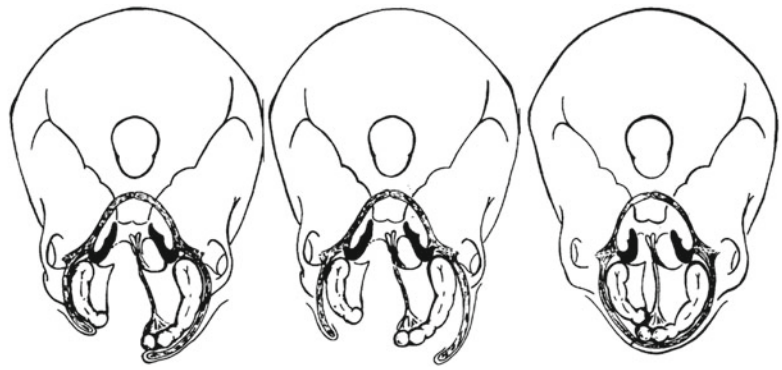


Fig. 16.3 Surgery to unite the lip over a large cleft space in one procedure. Undermining the soft tissue on the buccal alveolar surface posteriorly to the maxillary tuberosity prior to uniting the cleft lip has led to scarring and maxillary deformity. This surgery is no longer being performed and has been replaced with a lip adhesion or the use of external elastics as the first procedure of choice (Courtesy of Bardach (1990))

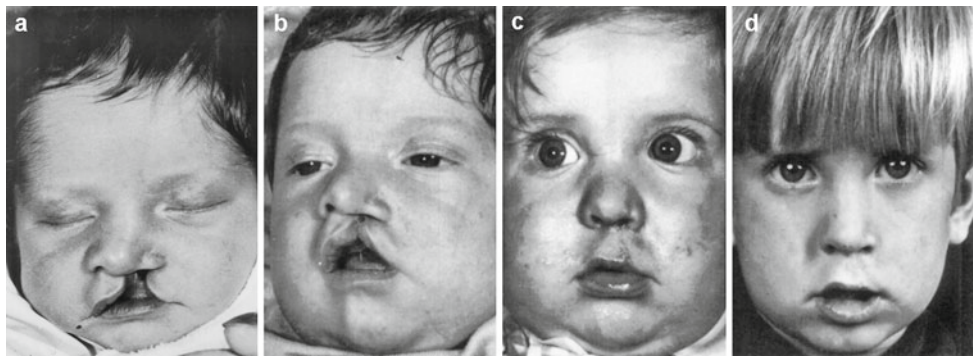


Fig. 16.4 (**a**–**d**) Lip adhesion. (**a**) Complete unilateral cleft lip and palate at birth. (**b**) Lip adhesion at 3 months. (**c**) After definitive lip repair at 7 months. (**d**) Symmetrical lip at 11 months

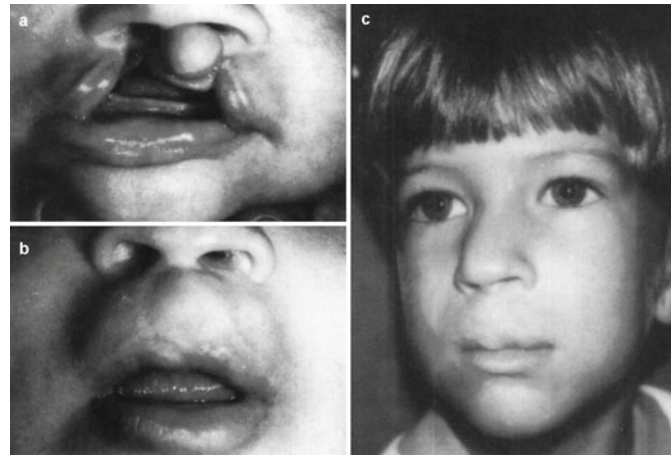


Fig. 16.5 (**a**–**c**) Lip adhesion in an incomplete bilateral cleft lip and palate. (**a**) At birth. (**b**) At 3 years, after lip adhesion and definitive lip surgery at 8 months. (**c**) Full face at 3 years, showing good "cupid's bow" and lip and nose symmetry

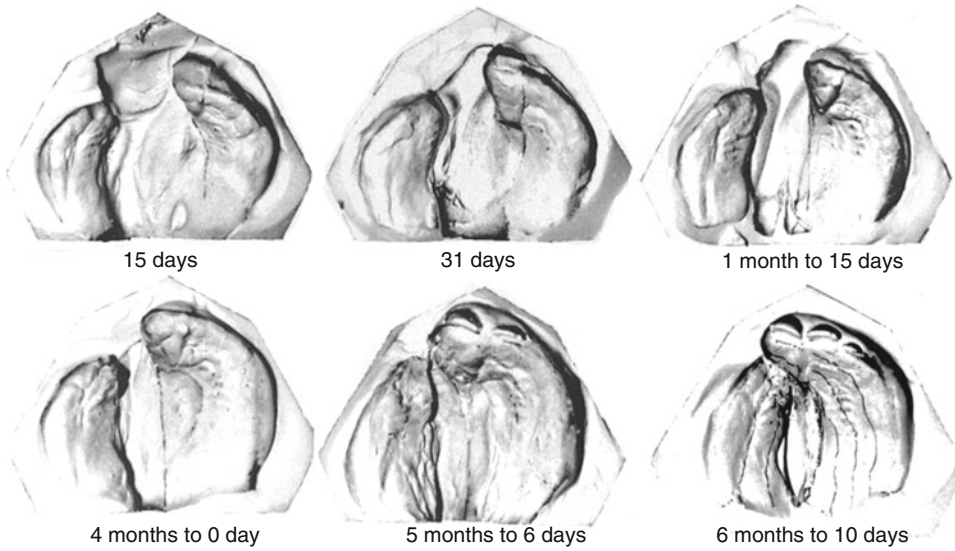


Fig. 16.6 An example of molding of overexpanded lateral palatal segments. In CUCLP, a lip adhesion was performed at 3 months of age without prior presurgical orthopedics. The alveolar cleft gradually closed with the alveolar segments overlapping

16.3 Palatal Cleft Surgery: Type, Timing, and Sequence (Figs. 16.6, 16.7, and 16.8)

16.3.1 What to Do and When to Do It: Speech and Palatal Growth Considerations

16.3.2 False Premise 1/2 Wrong Conclusions 1/2 Therapeutic Folly

At the turn of the last century, surgeons involved in cleft palate treatment usually performed surgical procedures whose sole purpose was to "close the hole" as early as possible without considering the ultimate effect of the surgery on palatal, facial, or speech development. These procedures, Millard (1980) reports, fall into three categories:

1. The use of various kinds of flaps from other parts of the body to cover the cleft space.
2. Treating the edges of the cleft so that they could be sutured together by pulling the mucoperiosteum over the cleft. Failure of a lasting union led to the use of laterally positioned relaxation incisions by Dieffenback in 1826

and von Langenbeck in 1862 (Lindsay 1974) (Figs. 16.9, 16.10, and 16.11).

3. Staged surgical treatment.

Gillies (1920), an early proponent of staged surgical treatment procedure, mistakenly believed that the maxillary and mandibular arches were in a normal occlusal relationship at birth. He did not appreciate the distorting effect of aberrant muscle forces on the geometric relationship of the palatal segments to each other. He did, however, suggest that the lip be united soon after birth and that the soft palate be detached from the hard palate and "pushed back" to increase its length. The cleft in the hard palate was to be closed before speech generally began at 2 years of age. For some surgeons, this is still the best surgical sequence to follow.

In the 1920s, William Wardill (Veau 1922) focused his interest on controlling airflow through the nose. He appreciated the value of the velum along with the levators as a part of the sphincteric muscle system. Because uniting the soft palate alone often did not bring good speech, many surgeons (e.g., (Veau 1922; Veau and Borel 1931; Wardill 1937; Dorrance 1933) believed, as Gillies (1920) did, that the velar length was insufficient in the von Langenbeck procedure and advocated "pushback" procedures (Calnan 1971).

In 1937, Kilner (1937) in London and Wardill (1937) in New Castle, publishing independently, described a technique of palatal repair that ultimately came to be known as the V-Y retroposition operation. Wardill (1937) and Kilner (1937) both adopted the Veau technique for anterior repair, and the resulting "Veau–Wardill–Kilner" operation consisted of (1) lateral relaxing incisions, (2) bilateral flaps based on posterior palatine arteries, (3) nasal mucosa closed as a separate layer, (4) fracture of the hamulus, (5) separate muscular lay-

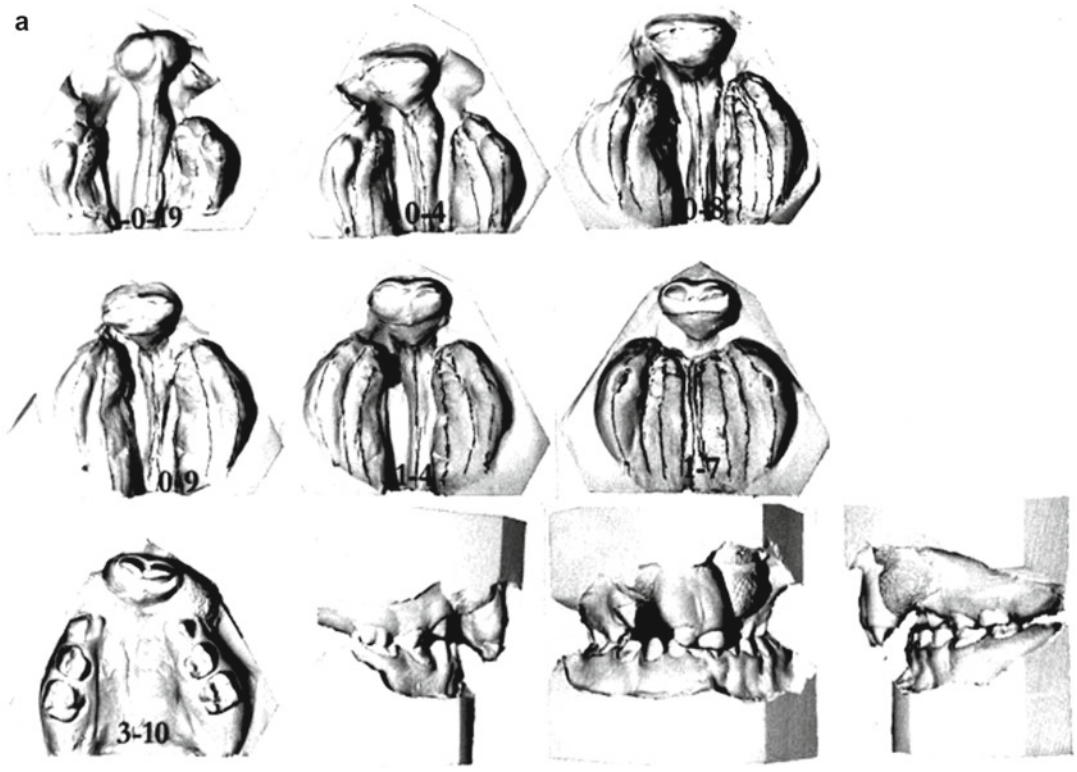


Fig. 16.7 (**a**) Lip adhesion causes premaxillary ventroflexion. Class II malocclusion with severe overjet. Anterior cleft space remains after 3 years and 10 months. No decision about correction of the overjet should be made at this age. An obturator closing off the cleft space will improve speech and feeding. Palatal growth needs to be considered for future treatment

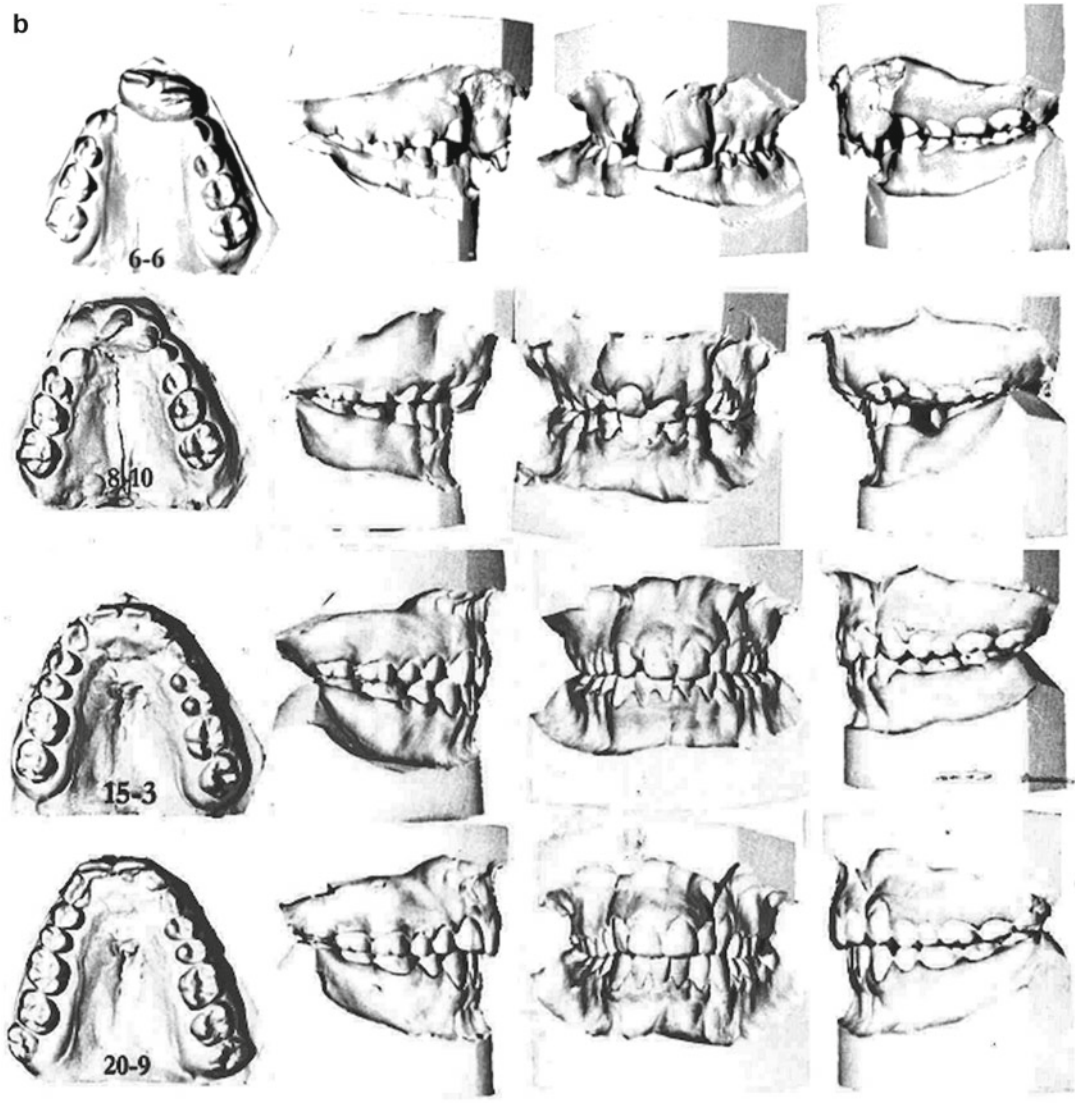


Fig. 16.7 (**b**) CBCLP. Serial cast of two cases (Figs. 16.7a, b and 16.8) demonstrates that the premaxilla and palatal shelves may react differently during the first years to the forces created by a lip strap attached to a head bonnet followed by lip surgery. In this figure, slow reduction of the anterior cleft space characterized this case. Although ventroflexion occurs, the anterior cleft space (the space between the premaxilla and the lateral palatal segments) remained until 3 years and 10 months of age with a severe premaxillary overjet and overbite. No surgery to the premaxilla was necessary other than alveolar bone grafting at 8 years. The following series of casts show the attainment of excellent aesthetics, occlusion, and speech. Palatal surgery using a von Langenbeck with vomer flap was performed at 23 months of age

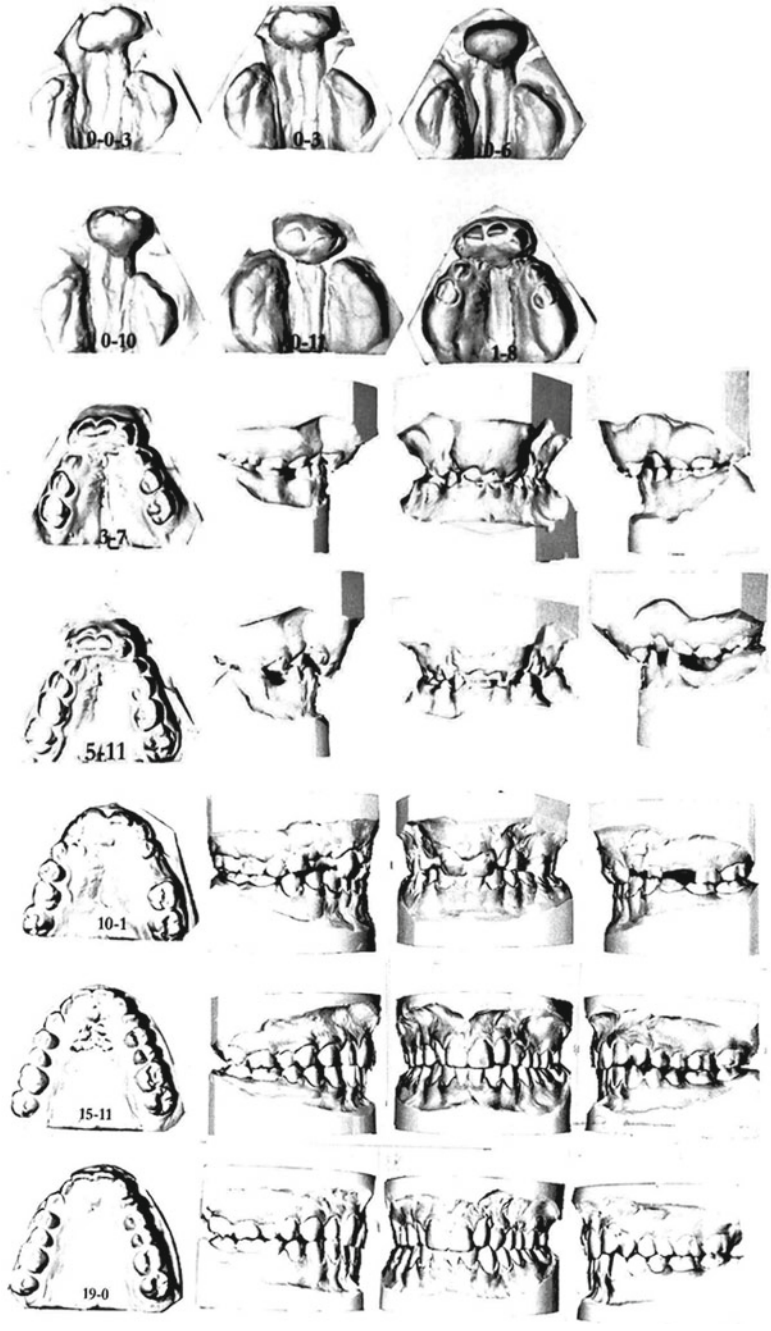


Fig. 16.8 Serial BCLP casts showing a rapid reduction in the anterior cleft space. The premaxilla made contact with the palatal shelves by 12 months of age while being positioned forward of them. Palatal surgery at 20 months of age resulted in excellent premaxillary overbite and overjet by 5 years of age. Good speech and aesthetics resulted

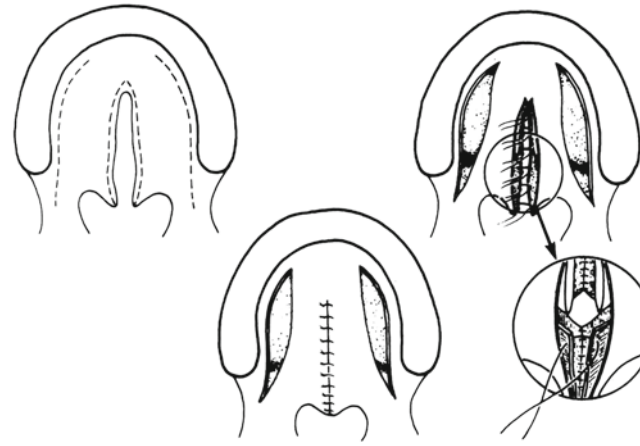


Fig. 16.9 Von Langenbeck (simple closure) palatoplasty for isolated cleft palate

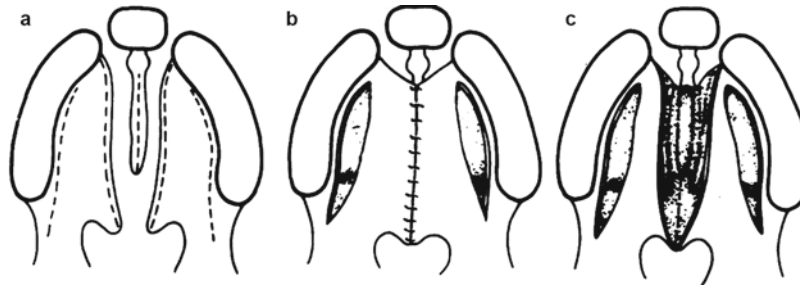


Fig. 16.10 (**a**–**c**) von Langenbeck (simple closure) palatoplasty for bilateral complete cleft lip-cleft palate. The configuration of this deformity varies tremendously, and line drawings can be misleading. (**a**) The incision lines. The inferior border of the vomer is incised, and mucous membrane is elevated from both sides. (**b**) Mucoperiosteal flaps are elevated without anterior detachment, keeping the posterior palatine arteries intact. Extensive dissection is frequently necessary in the lateral-release incision area for this anomaly when the cleft is usually wide and there is usually considerable hypoplasia. The timing of surgical closure is delayed until there is additional palatal growth at the expense of the palatal cleft space. The nasal mucosa is closed. Two suture lines are necessary anteriorly because of the two vomer flaps. (**c**) Oral mucosa closure. It is impossible to close the anterior portions of the cleft with this operation. The amount of denuded palatine bone is less, however. Anterior cleft closure is performed simultaneously with a secondary alveolar bone graft. Depending on the occlusion and extent of the anterior cleft space, the premaxilla and/or the lateral palatal segments are repositioned at the time of anterior cleft closure (Reprinted from Lindsay (1975), with permission from WB Saunders Co.)

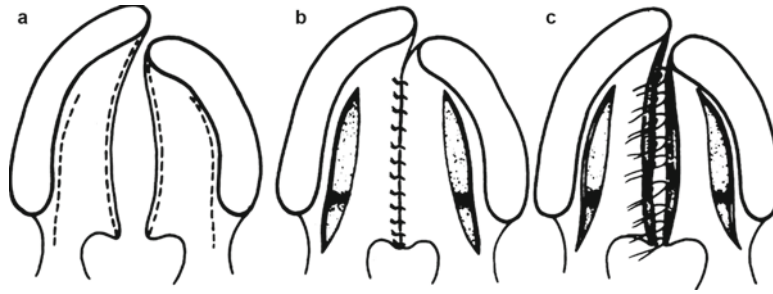


Fig. 16.11 (**a**–**c**) von Langenbeck (simple closure) palatoplasty for complete unilateral cleft lip-cleft palate. (**a**) The incision lines. (**b**) Mucoperiosteal flaps have been elevated, although this is not well shown in this diagram. The nasal mucosa is closed. In this anomaly, the nasal mucosa on the noncleft or medial side is continuous with the septum and vomer. More is available to manipulate. (**c**) Oral mucosa closure. Note that it is impossible to obtain a two-layer closure of the alveolar portion of the cleft with this operation. Lateral raw areas are relatively smaller because the mucoperiosteum is not detached anteriorly. The anterior cleft space is closed simultaneously with a secondary alveolar bone graft (Reprinted from Lindsay (1975), with permission from WB Saunders Co.)

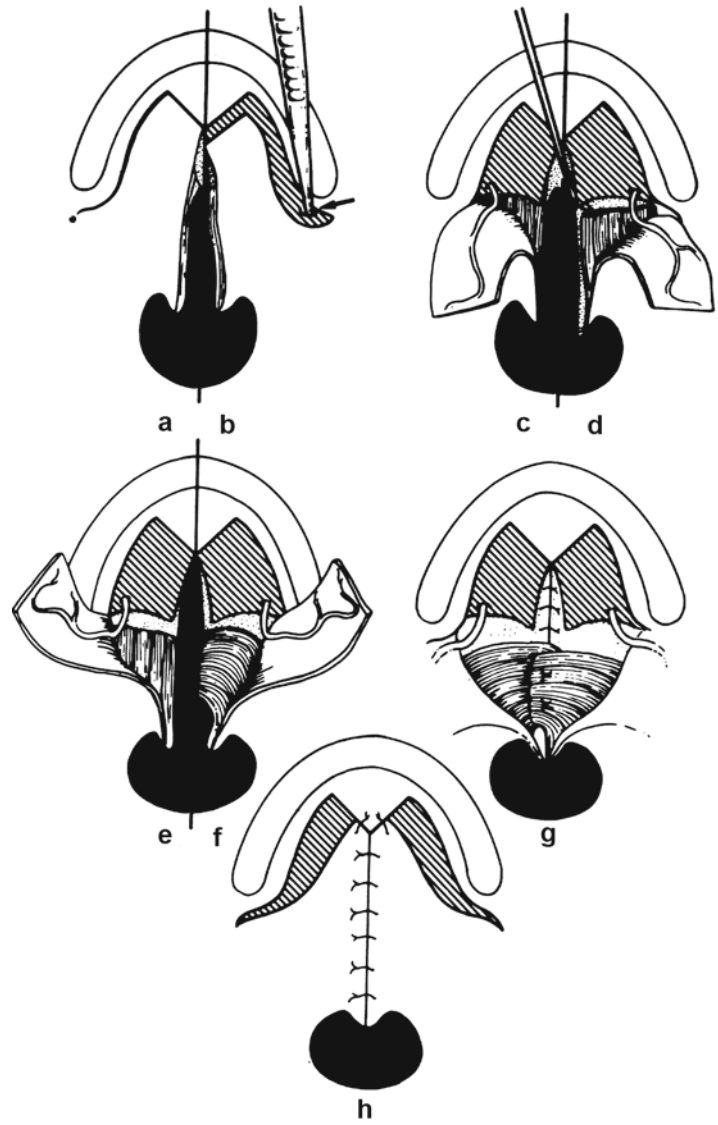


Fig. 16.12 (**a**–**h**) The three-flap Wardill-Kilner pushback modified after Braithwaite. (**a**) The margins of the soft-palate cleft have been pared, and the cleft of the hard palate incised along the junction of oral and nasal mucoperiosteum. The lateral incision has been made inside the alveolar ridge from opposite the canine anteriorly to a point just behind the hamulus posteriorly. An oblique incision joins the anterior end of the lateral incision to the cleft margin. (**b**) The mucoperiosteal flap has been elevated from the hard palate. (**c**) The oral mucoperiosteal flap has been turned back, showing the greater palatine vessels passing from the greater palatine foramen to the flap. The mucosa has been elevated from the septum (the vomer) which is attached to the margin of the palate on this side. (**d**) The muscles have been detached from the back of the hard palate. The soft palate is falling away from the hard palate and becoming elongated. This is the "pushback." (**e**) The muscle has been freed from the overlying mucosa. (**f**) The muscle has been freed laterally and has been rotated medially. (**g**) The two muscle bundles have been overlapped and sutured under slight tension to construct the muscle sling. The nasal mucosa has been sutured. (**h**) Suturing of the oral layer is completed. The tips of the oral mucoperiosteal flaps are sutured to the apex of the anterior flap, indicating the degree of palatal lengthening (Reprinted from Edwards and Watson (1980), with permission)

ered closure, and (6) V-Y type of palatal lengthening (Figs. 16.9, 16.10, 16.11, and 16.12).

Soon various pharyngeal flap techniques were introduced when it was suspected that the velum was still too short even after it was moved back (Curtin 1974).

In 1958, Kilner (1958) listed what he believed should be the aims of cleft palate treatment, in order of importance, as: speech, chewing, and aesthetics. This order of priorities is still preferred by many plastic surgeons, as well as speech-language pathologists. As a result, the surgical treatment concepts have been slow to change. To "close the hole as early as possible" with or without velar pushback is still a widely prevailing concept.

"What to do and when to do it" were questions that had no universally accepted answers. Although surgeons such as Veau (1922) and Veau and Borel 1931), who appreciated the morphological differences in cleft types, still believed that closing the cleft palate early would improve speech development, many others began to think differently and empha-

sized the need for normal midfacial and palatal development. They recommended that cleft palate closure be postponed until either the deciduous or the permanent dentition had erupted. Advocates of delaying palatal closure were influenced by Graber (1950, 1954) and also by Slaughter and Brodie (1949), who were disturbed by the results of Brophy's (1904) and other procedures in vogue at that time. They wanted to avoid secondary malformations to the palate and severe deformities of the maxilla caused by extensive mucoperiosteal undermining with wide lateral areas of denuded bone which led to severe scarring. Similar malformations were created in animals by Kremenak et al. (1970, 1971, 1977).

16.4 The Effect of Surgery on Maxillary Growth

Kremenak and his colleagues (1967, 1971, 1976) reported a series of follow-up studies on beagles, based on earlier work, showing that surgical denuding of palatal bone adjacent to deciduous teeth resulted in inhibiting maxillary growth. More recent efforts by this group have focused on the contraction phase of early healing of surgical wounds on the canine palate. Olin and associates (1974) presented data based on measurements between tattoo points on wound margins on hard palates of young beagles. They noted that major interruptions in the increase of arch width coincided with the period of soft tissue contraction. This led Olin et al. (1974) to suggest that the contraction of wounds following surgery could be the first link in a causal chain leading eventually to secondary skeletal deformities.

Kremenak and associates (1970) reported related evidence that the contraction seen in palatal mucosa was analogous to that reported in the healing of skin wounds. Transplanting autogenous grafts from other oral mucosa into mucoperiosteal excision wounds of the hard palate resulted in a reduction in contraction and was followed by normal or "supranormal" increases in maxillary arch width. They concluded that more research in this area was warranted.

It is becoming apparent that the factor of contractility in some types of nonmuscular connective tissue cells may be a common denominator for research on surgical wound healing, as well as for research on normal and abnormal growth and function in the craniofacial complex. There is a large body of literature dealing with advances in the understanding of contractile phenomena. Much of it is collected in a bibliographic review by Morris and Kremenak (1976). The report by Madden and associates (1974) also provides an excellent overview of this area of inquiry.

Is surgery for the repair of cleft lip and palate to be determined by the age of the patient or by the palate's morphological characteristics and the effects of surgery on the subsequent growth of the face? This was one of the driving questions being asked by surgeons in the 1940s. Slaughter and Brodie (1949) and Graber (1950, 1954) deserve historical credit for bringing this issue to the world's attention. Their condemnation of the results of cleft palate surgery, as mentioned, led to a new conservatism and forced a reexamination of surgical practice.

The beginning of serial facial growth studies, Graber's (1950, 1954) cross-sectional study of 60 cases of mixed cleft palates of various types at various ages, had a profound effect on surgical planning. He compared jaw development of 46 operated cases with 14 cases that had had no surgery and found that the unoperated palates resembled normal palates more than they did the operated palates. He noted that the growth of the operated palates was retarded in all three dimensions. It should be understood that it was not unusual in those days to encounter patients with a history of 35 operations. A common operation of the day involved wiring compression of the cleft segments (Brophy 1904). As already noted, this procedure was notorious for its mutilating effects. Dental neglect was common, resulting in rampant caries, and the associated malocclusions were often so severe as to discourage orthodontic treatment.

Graber (1950) concluded that "to minimize interference with growth centers, it seemed advisable to postpone surgical correction at least until the end of the fourth year of life, when five-sixths of the total maxillary width has been accomplished." As a result, early surgical repair of the palate fell into disfavor, and prosthodontists came to dominate the rehabilitation of cleft palate.

Graber (1950) suggested that the following questions should be asked:

1. How does the pattern of growth and development of a cleft palate individual compare with that of a noncleft person?
2. What is the effect of early and repeated surgical intervention on this pattern?
3. What happens to a tissue that has been manipulated and traumatized?
4. Does increased tension of soft tissue stimulate or depress cellular proliferation?
5. What is the growth potential of fibrous bands of scar tissue?
6. Can cicatricial (scar) bands influence the normal growth and development of the surrounding soft tissue structures and the bony skeleton?
7. How do cleft palate individuals with surgically closed clefts compare with those who have had no surgery?

Pruzansky, in 1969 at an international symposium on cleft palate held at Northwestern University Dental School, recognized the important contributions of Graber (1950, 1954) and Slaughter and Brodie (1949) for their condemnation of the deleterious effects of traumatic surgery on midfacial growth (Gruber 1969) and recommended that surgery on the palate should be delayed until 5 or 6 years of age to minimize facial growth malformations. In questioning the validity of their conclusion on the timing of surgery, Pruzansky goes one step further, citing findings from his longitudinal palatal and facial growth studies (Gruber 1969).

> I saw patients whose palates had been repaired early and yet their faces developed well and their speech was free of the stigma associated with the stereotype. On the other hand, some children, for whom palatal surgery had been delayed, did not do well at all. Their midface did not develop normally and their speech was hypernasal and unintelligible. I saw all kinds of permutations and combinations.

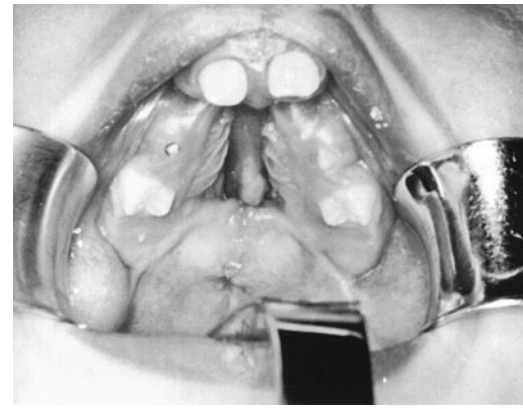


Fig. 16.13 Palatal view showing the lip and soft-palate closure as a primary procedure. The hard-palate cleft is closed at a later date, usually between 18 and 30 months according to the size of the cleft space

He went on to explain that the faultless error was in not recognizing the heterogeneity that exists within a single cleft type. Pruzansky (1953, 1955, 1974) stressed that morphological and physiological variants in the individual child should provide the rational basis for therapeutic design, not age alone.

In the early 1920s, the cleft space was considered a "hole" that needed immediate closing either by orthopedics and surgery. Early surgical closure in the first 6 months led to disastrous results. As a reaction, the timing pendulum was to swing to the opposite extreme to favor surgical closure at 5–9 years of age, when the hard palate's growth was 90 % completed. To satisfy the speech-language pathologists, obturators were worn until the cleft was closed. Based on theoretical and anecdotal evidence, many surgeons chose 18–24 months for closure without the use of obturators as a compromise between speech and growth requirements.

Many variations in surgical procedures arose. Gillies and Fry (1921) and Slaughter and Pruzansky (1954) believed it was important to delay hard-palate closure to 2 or 3 years of age and chose to close the lip and soft palate first to aid speech development (Fig. 16.13). Others (e.g., Robertson and Jollys 1968; Hotz 1973) delayed midpalatal closure to the deciduous dentition, and Schweckendiek (1978) went to the extreme and delayed closure into adolescence, favoring the

need to maximize palatal growth above all else. The Slaughter and Pruzansky (1954) method was associated with good speech development; the Schweckendiek delayed palatal closure procedure, and although producing satisfactory midfacial growth, it left the patients with very poor speech.

16.5 Speech Considerations

Some surgeons believe that good speech requires palatal cleft closure between 6 and 9 months, the child's stage of phonemic development of articulation, and are willing to accept the trade-off of creating some maxillary growth inhibition (Dorf and Curtin 1982). Other surgeons believe that good speech development is not related to the age of cleft closure but depends on the growth integrity of the entire midface: the palatal vault space, the size and shape of the pharyngeal space, and the size and neuromuscular action of the soft palate and pharyngeal muscles, along with hearing and the patient's phenotype (Bzoch 1979).

Blocksma et al. (1975), Lindsay (1971), Kaplan et al. (1978), Krause et al. (1976), and Musgrave et al. (1975) found good speech with the von Langenbeck procedure in most cases. This repair gave slightly better speech results than the lengthening procedures in soft-palate clefts, but the V-Y pushback was superior in the more extensive clefts. However, the authors do not discuss the effects of the procedures on palatal growth and dental occlusion. Dryer and Trier (1984) compared the speech results when three different palatal surgeries were performed: (1) V-Y or island pushback, (2) von Langenbeck method utilizing two bipedicled mucoperiosteal flaps advanced to the midline, and (3) von Langenbeck procedure with the addition of levator reconstruction (intravelar veloplasty). The speech results after palatoplasty revealed no significant difference between children with simple von Langenbeck closure and those undergoing palatal lengthening procedures. Children with levator reconstruction demonstrated superior speech results.

As to maxillofacial development, Jolleys (1984) found better speech results in his patients who had surgery prior to age 2 and found no difference in maxillofacial development. Koberg and Koblin (1973) suggested that 2–3 years old is the best age for surgery without damaging maxillary development. Friede et al. (1980) and Berkowitz's (1985) facial and palatal growth studies do not support Ross's (1987a, b) conclusion that early (before 18 months) palatal repair provides better facial growth than does delayed hard-palate repair. The opposite appears to be true because deformed palates are more closely related to early (within the first year) surgery than delayed (4–9 years of age) surgery. For example, Graber (1950) reported:

> In various studies of facial growth and development, it is seen that the lateral width of the maxilla is accomplished quite early in life, but the downward and forward growth is not complete until the second decade of life. Any growth disturbance induced by environmental interference would be possible in the sutural sites of proliferation for a number of years, but any appreciable withholding of the palate in width would require speech interference during the first 4 years of life.

All goals of a good speech, speech occlusion, and facial aesthetics are possible. Most orthodontists and surgeons now agree that the skills of the surgeon need to be considered when evaluating all surgical procedures, but the timing of palatal closure to avoid scarring is equally important.

Attainment of normal speech, facial and palatal development, and dental occlusion is possible without compromising one objective for another. Although speech development may benefit from early palatal closure, there are instances when the cleft space is very wide and cleft closure should be postponed to a later age to permit additional palatal growth and allow for conservative palatal surgery. Berkowitz's palatal growth studies, presented in this text, show that an increase in palatal size with the spontaneous narrowing of the cleft space can occur early, late, or not at all, and in rare instances, the cleft may even widen. Nonphysiological surgery causes facial and palatal deformation due to the destruction of blood supply with scar formation. To avoid these consequences, timing of palatal closure should be related to the anatomical and functional assets in

the individual and not determined by age alone. Berkowitz's (1985) serial studies of 36 unilateral (UCLP) and 29 bilateral (BCLP) cleft lip and palate cases with good speech demonstrated that conservative palatal surgery is conducive to good speech, as well as good palate and facial development. Speech appliances, in very rare instances, may be necessary as an interim device to close off the cleft space after 2 years of age.

Van Demark and Morris (1922) found that early surgery (before 24 months of age – before the child begins to talk) to close the cleft space was associated with better articulation skills when the children were tested at 8 years of age, but after that age, differences were less often seen. They believed that other variables, for example, velopharyngeal competence and cleft type, were better predictors of eventual articulation skills than was age at surgery.

McWilliams et al. (1984), in reviewing the literature, found better speech to be associated with earlier palatal repair (before 24 months of age). However, they noted that design flaws in the studies make it difficult to interpret results. Failure to define "normal," "perfect," or "acceptable" speech and the omission of information about cleft type and surgical techniques are significant shortcomings in comparing results.

Most speech-language pathologists still advocate closure of the palatal cleft before 1 year of age, believing that early closure prevents development of patterns of speech that would require prolonged and difficult therapy at a later age (McWilliams et al. 1984). Most of their studies and conclusions are based on the use of only two variables: timing of surgery and speech outcome, omitting other significant variables; therefore, their conclusions must be questioned. The error in relating speech adequacy to the age that a palatal cleft was closed has only confused the issue of individualizing treatment planning based on differential diagnosis of the cleft defect.

Speech studies of individuals or a small sample of subjects are often sufficient, and sometimes essential, to finding solutions to a particular problem being investigated; however, the fact that a "correlation" exists between two variables (speech proficiency and age) does not indicate or prove causation. Although the age at which surgery is performed may be the sole relevant variable studied, sensory function, genotype, the geometrics of the original deformity, the facial growth pattern, and the surgical procedure performed are also factors that must be considered. It is difficult, and perhaps impossible, to demonstrate the effectiveness of a treatment philosophy in a clinical setting, where many variables cannot be identified, controlled, or manipulated. Therefore, conclusions drawn from such investigations should be considered with caution (Berkowitz 1985).

Some past surgical strategies also emphasized early palatal closure and velar lengthening with the goal of resolving immediate problems with cleft space and, hopefully, preventing future speech problems associated with hypernasality. In addition, velum lengthening procedures, originally advocated in the newborn period – often without evidence of incompetent airflow control – to avoid a possible second surgical procedure, were found to be ineffective even when performed at a later age (Millard 1980).

Surgery can either aid in directing the natural growth into proper channels by establishing muscle balance across the cleft defect, or it can grossly interfere with the normal developmental changes by hindering growth through interference with blood supply, introducing a scar or destruction of growth centers. In view of the wide range of individual variations of the defect, the surgical procedure must be altered to fit the particular case, in terms of both time and technique.

Determining which surgical orthodontic procedures are best utilized for different types of clefts is the goal of all clinicians. Unfortunately, palatal and facial growth patterns are not predictable at birth, and therefore one needs to wait to see the effects of initial treatment on the developing face before deciding what next needs to be done. When decisions to act are made under conditions of uncertainty, it is always appropriate to consider not only the probability of success but also the consequences of failure.

16.6 Surgical Orthodontic Procedures and Sequences

It is impossible to review all of the surgical orthodontic approaches to the closure of lip and hard and soft palatal clefts with or without the use of

orthopedic plates. Some surgeons will first unite the lip, followed either by closure of the hard and soft palate in one stage or first unite the soft palate, and then the hard palate at a later age. Some even favor closing the soft palate before uniting the lip. There are as many different surgical procedures as there are differences in the timing and surgical sequence to be utilized. Some surgeons favor a vomer flap with or without a mucoperiosteal flap, others with/without a pushback, still others a mucosal tissue closure without involving the periosteum. Yet each clinical report tells of both good and bad results, usually using the number of buccal and anterior crossbites in the deciduous dentition to designate whether the procedures have failed or succeeded. Unfortunately, most reports do not have final post-pubertal facial/palatal records, which are more meaningful in describing the final outcome.

16.6.1 Palate Cleft Closure Controversies Revisited

Present-day corrective procedures involve surgery, the type and timing of the operation depending on whether the surgeon is a von Langenbeck soft tissue descendant, with or without presurgical maxillary orthopedics, or a Brophy "steel clamp and silver wire" bony closure man. There is a basic philosophical conflict between the two major groups. Some surgeons employ staphylorrhaphy for closure of the cleft palate, a variation of the von Langenbeck, Furlow, or Veau–Wardill–Kilner V-Y pushback procedure using mucoperiosteal flaps. They may or may not use vomer flaps to line the surface of the mucoperiosteal flap. The soft tissue is repositioned over what many surgeons consider a normal bony framework (with the exception of the cleft area) that had failed for some reason to unite in the midline. Today, most surgeons are convinced that aberrant embryonal and fetal influences force the lateral halves of the maxilla apart, making the intramaxillary width excessive. In the past, it was Brophy; today, it is many McNeil disciples who utilize presurgical maxillary orthopedics, believing that the first step in habilitation is to restore what they believe to be a normal segmental relationship at a very early age. Each school of thought has intense convictions that there is only one correct approach – its own.

Four surgical approaches are commonly used to close the palatal cleft:

1. Early complete palate repair (3–9 months). Rationale: to achieve maximum speech results with a possible chance of inhibiting midfacial growth and creating severe dental occlusion. This approach favors speech above facial growth and development.
2. Delayed complete palate repair (12–24 months). Rationale: Speech results are nearly as good as with earlier repair, and the facial growth disturbance is less.
3. Late complete palate repair (2–5 years). Rationale: to prevent facial–palatal growth inhibition, accepting the poorer speech results. Use of a palatal obturator is needed in most cases.
4. Early lip and soft-palate repair (2–9 months) and delayed hard-palate repair (5–9 years). Rationale: to avoid facial and dental deformity but perhaps still achieve good speech with the aid of an obturator.

16.6.2 Scarring Inhibits Palatal Growth

Surgery to the hard and soft palate with temporary disruption of the blood supply does not, by itself, cause damages to the underlying bone. Most surgeons and orthodontists believe that the principal growth inhibitor seems to be the quantity and distribution of scar tissue that is created after surgery. When evaluating the effects of surgery on maxillary growth and development, it is necessary to consider that clefts of the palate can differ greatly in size and form at the same age due to the amount of osteogenic deficiency in the hard and soft palate and lip. The great individual variation in the relationship of the size and form of the cleft space relative to the size of the palatal segments is responsible for the differences in the amount of scar tissue formed even when the same surgical procedure is performed by the same surgeon.

A review of cleft palate surgical history clearly shows that a single mode of surgery for all cases invariably resulted in severe palatal and midfacial

deformities, as well as poor speech development. Unfortunately, the same poor results still occur despite the timing of surgery and the skill and experience of the plastic surgeon. This is so because of failure to define the criteria for the timing of palatal surgery and failure to agree on which surgical procedures interfere with normal growth and development of the structures involved. Poor results were understandable when there were no standardized methods for estimating success or recording the effects of surgery on speech and facial growth and development; these shortcomings no longer exist. Nonetheless, some present-day surgical reports still do not adequately describe the original deformity; thus, the efficacy of the surgical effort cannot be evaluated.

Mapes and coauthors (1974), Robertson and Fish (1975), and Berkowitz (unpublished data) all concluded that nontraumatic palatal surgery accelerated the growth rate of the maxilla, helping it reach more normal dimensions in the following years. Berkowitz and associates (1974) demonstrated that, in the patient with complete bilateral cleft lip and palate, after conservative palatal surgery, the palatal surface area doubled from birth to the age of 1 1/2 years. Also, in an isolated cleft palate of a patient with Pierre Robin Sequence, there was a 50 % increase in the palatal surface area from birth to 1 year of age. The acceleration in growth tapered off after palatal surgery in both instances. Palatal growth accelerates 6–12 months postsurgery in some cases.

Berkowitz found that cases with relatively small cleft spaces prior to using modified von Langenbeck surgery grew the best after surgery. This can be interpreted to mean that the smaller the laterally placed areas of denuded bone, the less scar tissue will result with a better chance of obtaining "catch-up growth."

Bardach (1990) questioned the validity of claiming that palatoplasty is detrimental to maxillary growth. He believed there is no adequate substantiation of this concept from clinical or experimental studies. However, Berkowitz's (1974) clinical report on Millard's island flap pushback procedure, which creates large areas of denuded bone, has conclusively shown that this procedure deforms the palate and causes major maxillofacial growth aberrations (Figs. 16.14, 16.15, 16.16, 16.17, 16.18, 16.19, 16.20, and 16.21). However, this study does not find fault with palatoplasties that produce small areas of exposed palatal bone. The same palatoplasty can yield different long-term results because all clefts within the same cleft type are not the same. They may have different size of cleft spaces. Unfortunately, Bardach focuses only on the surgery performed and does not consider the geometric variations in the cleft deformity as critical factors in predicting the long-term outcome of surgery.

Research is presently underway in Berkowitz's laboratory to determine why "catch-up growth" occurs in some but not all cases even when surgery is performed by the same surgeon using the same surgical procedures.

In the course of a set of related experiments, Latham and Burston (1964), Latham (1969, 1980), and Calabrese and coworkers (1974) suggested that new tissue could be induced to form in the growing face by applying appropriately controlled physical stress through neonatal maxillary orthopedics. However, no known supportive objective data have ever been presented. This subject was covered in depth in Chap. 21.

Viteporn et al. (1991), utilizing longitudinal cephaloradiographs, showed that patients with extensive cleft palate who had a pushback procedure reached maximum growth spurt later than patients with less extensive surgery and that the surgery had an inhibitory effect on midfacial growth. They concluded that, because the sample was of the same ethnic group and received surgical treatment at the same age by the same surgeon, significant differences in midface development between the two groups should be attributed to the treatment itself. Scar tissue associated with the denuded bone left after the V-Y pushback technique, they claimed, played a major role in inhibiting forward displacement of the maxilla and in distorting dentoalveolar growth.

16.7 Dental Occlusion Associated with Early Palatoplasty Using a Vomer Flap

Dahl et al. (1981) reported on the prevalence of malocclusion as seen in early mixed dentition with complete unilateral clefts of the lip and palate. The frequency of posterior lin-

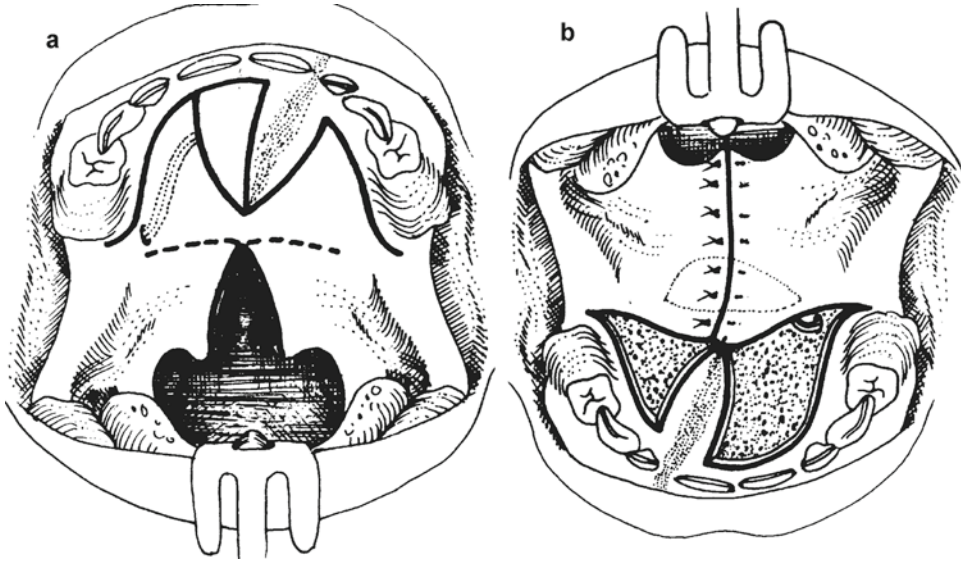


Fig. 16.14 (**a**, **b**) "Island flap" for a unilateral cleft lip and palate. (**a**) Outline of incisions at 21 months. (**b**) At 21 months, a V-Y pushback of the palate was achieved, leaving the V over the anterior closure untouched and taking a unilateral island flap for nasal lining (Courtesy of Dr. Millard, Jr.). Comments: The island flap is a variant of a V-Y pushback that is very similar to the Wardill–Kilner V-Y pushback procedure in that a large anterior area of denuded bone remains. In the "island flap," the anterior palatal mucoperiosteum is transposed to nasal surface, creating a mucoperiosteum sandwich. This is the area that creates the transpalatal scar. Because the denuded bone heals by epithelialization becoming scar tissue, the larger the denuded area, the more the resulting scar tissue will negatively affect maxillary growth in three dimensions. The maxillary deficiency usually becomes apparent in the late mixed or permanent dentition when facial appearance is affected. Lateral cephalometric studies have shown that this or other "pushback" procedures have not resulted in a net gain in soft-palate length. We speculate that, in most cases, good velopharyngeal closure would have occurred even if a "pushback" had not been performed as a primary palatal cleft closure procedure. The singular lesson to learn from these longitudinal facial–palatal growth studies is the need to avoid creating large areas of denuded bone when performing palatal surgery

gual crossbite they found was extremely high. This might be partly an effect of the two-layer palatoplasty–vomerplasty used for closing of the cleft in the hard palate at 12 months of age. It has been shown that bone formation in the palatal cleft is common subsequent to this procedure (Prysdo et al. 1974). It was supposed that the newly formed bone might act as a bony ankylosis, inhibiting the transverse growth of the maxilla. A longitudinal study, by the implant method, of patients operated on by this method substantiated this supposition. To reduce the adverse effects on transverse maxillary growth, Dahl recommended the surgical procedure be changed to a one-layer closure of the cleft in the hard palate by a vomer flap. There has not been a follow-up study to evaluate the effect of changing the procedure – whether it is the timing or the surgery that needs changing. Dahl's Danish group of treated patients were characterized by unilateral lingual crossbite, high frequencies of midline deviation, mesial molar occlusion, and mandibular overjet. There were very few cases with an anterior open bite. A tendency was seen for a difference in the sagittal molar relationship on the cleft side, as compared with the noncleft side, with distal molar occlusion being more frequent on the cleft side. This may be explained by a primary difference in the sagittal position of the two segments of the maxilla and also as a result of secondary changes, such as lateral shifting of the mandible caused by lingual crossbite on the cleft side, medial rotation of the cleft segment subsequent to surgery, and tipping the teeth toward the cleft on the affected side.

Berkowitz's (1985) serial occlusal study of complete unilateral clefts of the lip and palate, which involved the use of modified von Langenbeck procedure with a vomer flap between

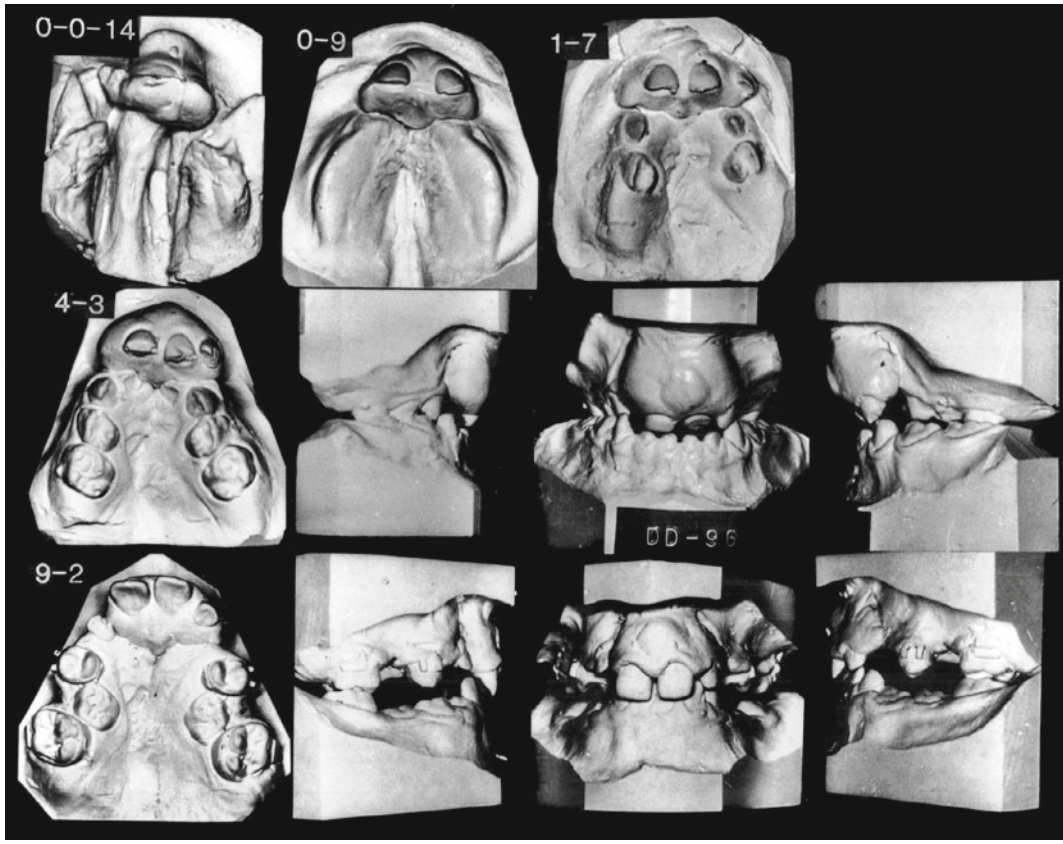
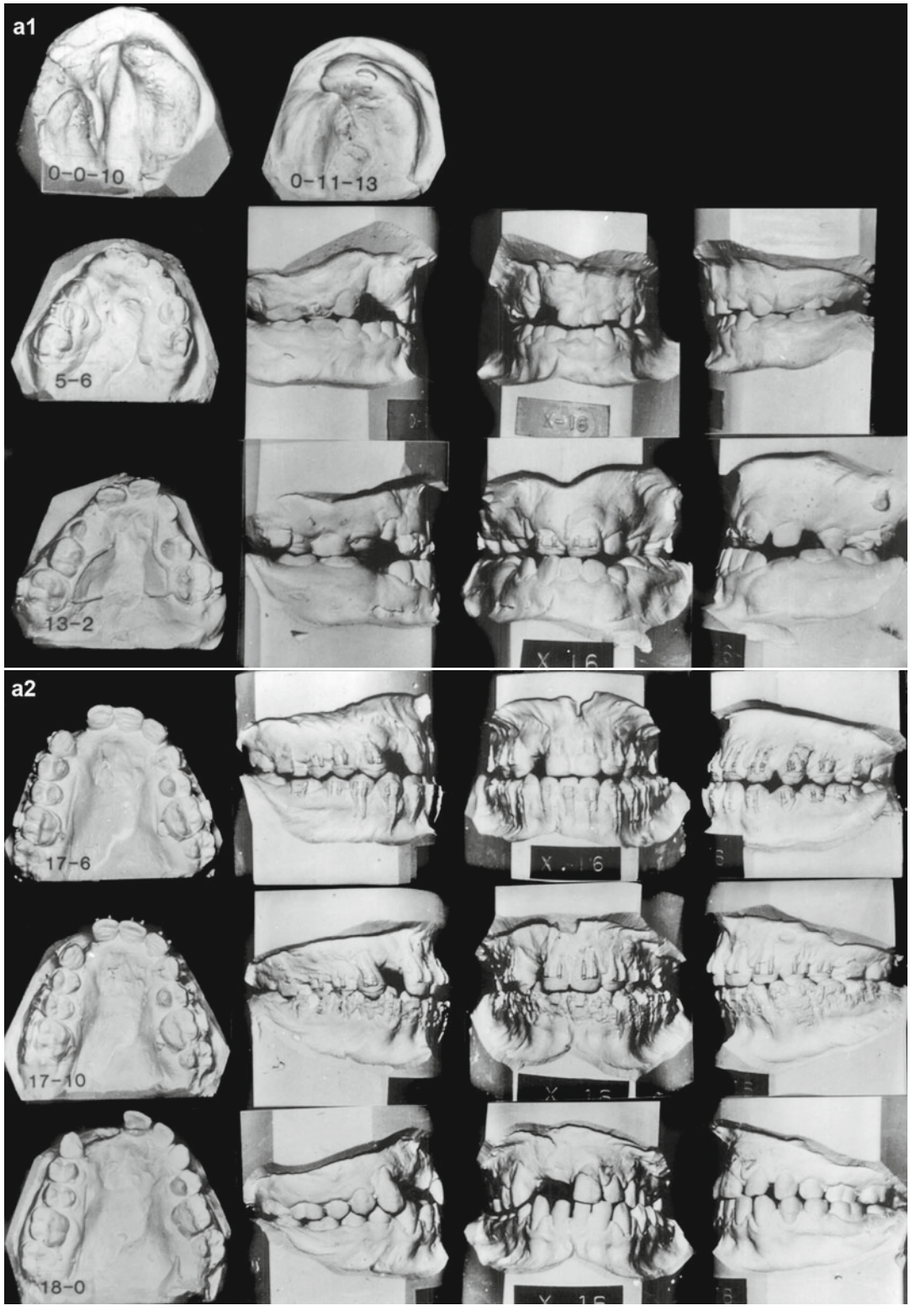


Fig. 16.15 Case No. DD-96. Serial casts of bilateral cleft lip and palate show marked palatal scarring following early "island flap" pushback. 0–0–14: At birth. 0–9: Excellent palatal arch form after the lip was united. The premaxilla lateral palatal segments are in good relationship. 1–7: Narrow and distorted palatal arch due to island flap scarring performed earlier. 4–3: Bilateral buccal crossbite with the anterior incisor are in a tip-to-tip relationship. 9–2: The palatal arch width narrowed at the line of the island flap creating an "hourglass"-shaped palate

Fig. 16.16 (**a**–**d**) Case JB. No. X-16. Serial palatal changes after "island flap" leading to Le Fort I maxillary advancement. (**a**) Serial casts. The island flap V-Y procedure was performed at 5 months. 0–11–13: The palate shows severe scarring and collapse. 5–6: Even after a difficult attempt at palatal arch expansion, the buccal and anterior occlusions are still in a tip-to-tip relationship with the opposing teeth. 13–2: The palatal arch collapsed again due to the strong medial pull of the transpalatal scar placing the buccal and anterior teeth in crossbite. Note that midfacial recessiveness increased as the face grew even with orthodontia and protraction mechanics and by 17–6: a Class III malocclusion existed. 17–10: After Le Fort 1 maxillary advancement. 18–0: The right maxillary incisor and alveolar labial plate exfoliated as a result of the severance of the blood supply to this area. 15–13c: A "roundhouse"-fixed bridge was utilized to replace the missing teeth and stabilize the arch form. A transpalatal removable metal strut helps to maintain the corrected arch by counteracting the medial pull of the severe scarring. Lip and palate surgery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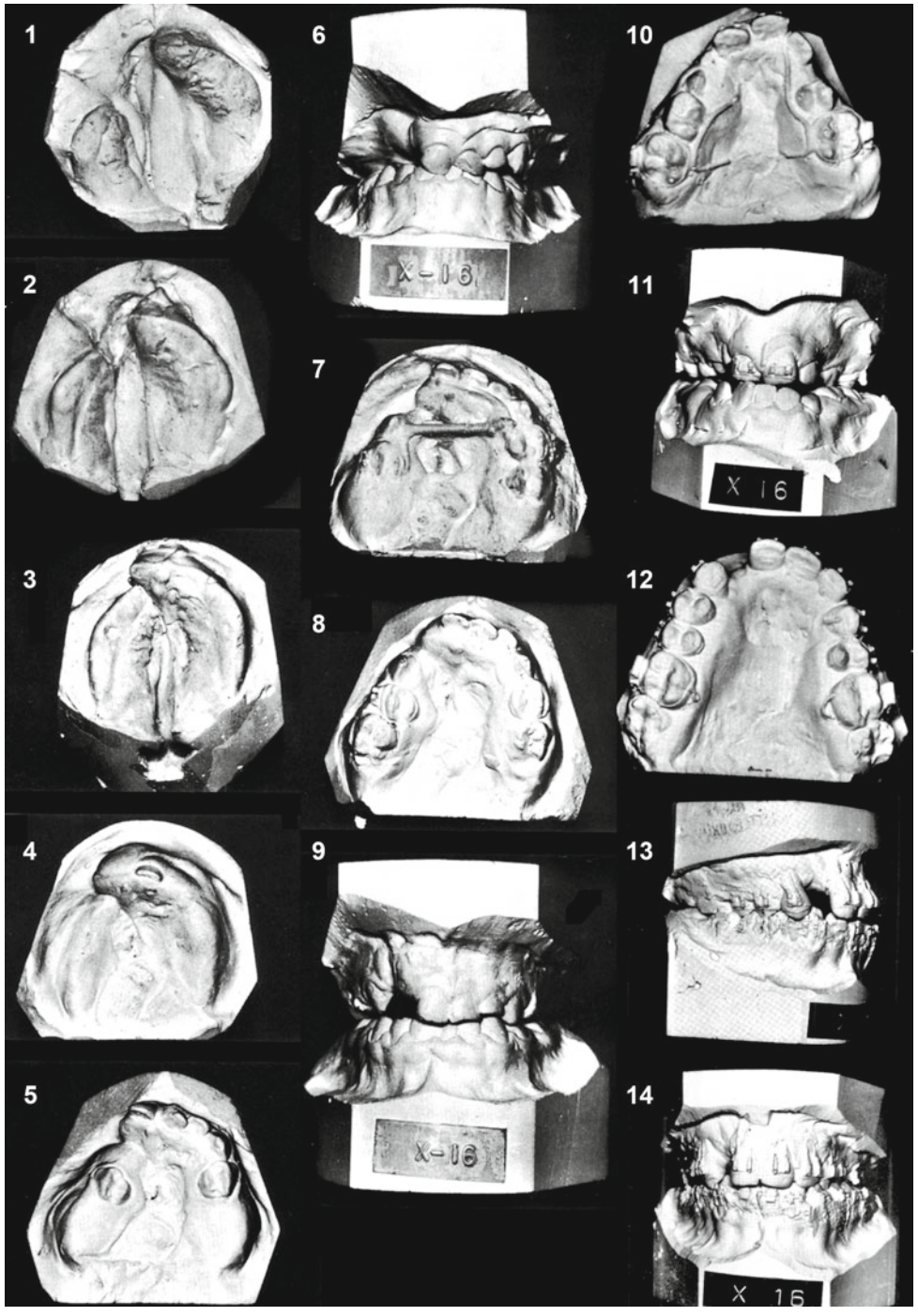

Fig. 16.16 (*continued*) *1* Newborn palatal cast. *2* Narrowing of the cleft space due to molding action brought on by a facial elastic worn over the cleft lip. *3* Palatal segments are in contact. *4* Further palatal narrowing after island flap pushback. *5* Severe loss of palatal vault space. *6* Right buccal crossbite. *7* Expansion appliance in place. *8* Failure to increase palatal arch width. *9* Right buccal occlusion still tip to tip. *10* Narrowed palatal arch at 13 years 2 months of age. *11* Class III malocclusion with bilateral buccal crossbite. *12*, *13*, *14* (17–10) After Le Fort I advancement. Excellent dental occlusion has been established. The scar tissue has been stretched, allowing for the congruency of the upper and lower arches

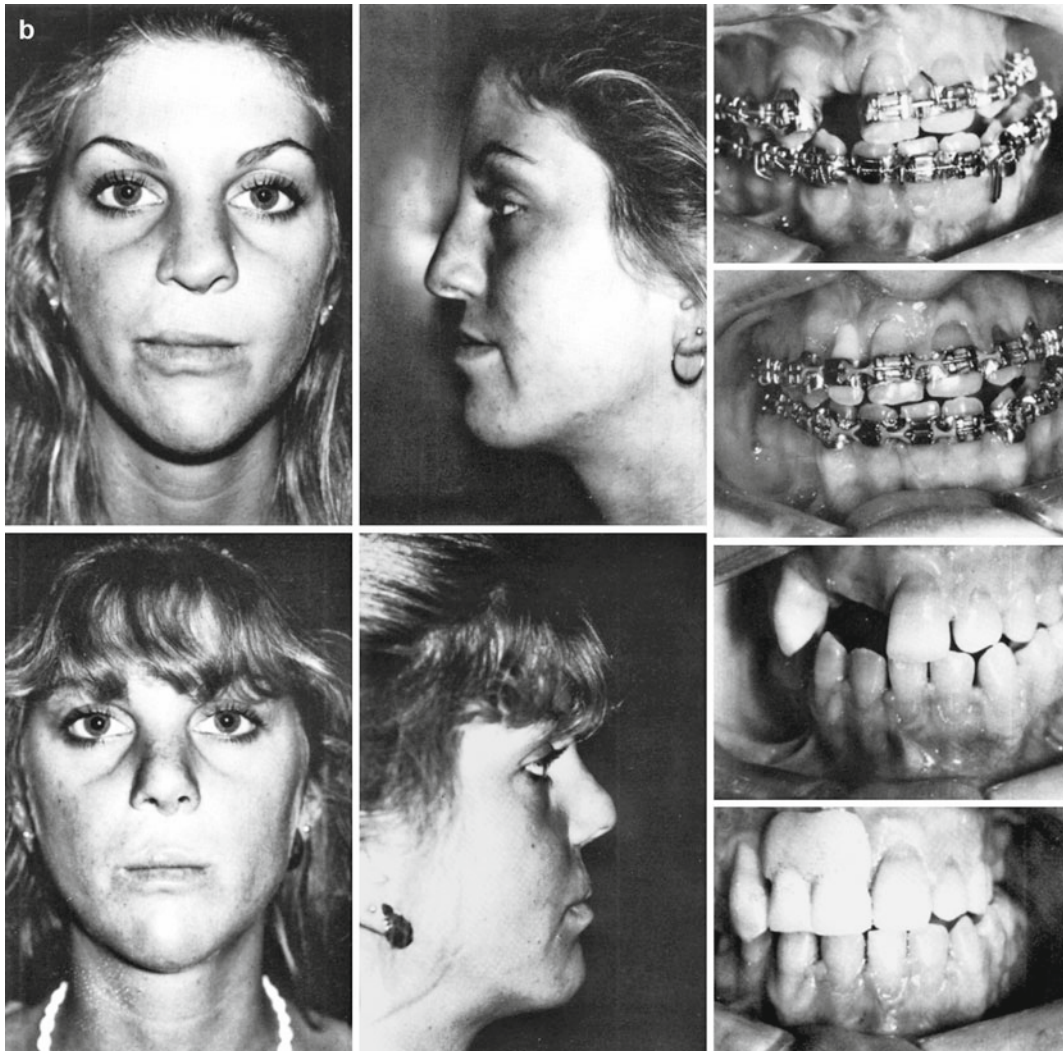

Fig. 16.16 (*continued*) (**b**) Lip and palate surgery. *1* and *2* Prior to Le Fort I surgery. *3* Presurgical orthodontics. Buccal expansion and anterior advancement are unsuccessful. *4* Right lateral incisor pontic placed on the arch wire. *5* and *6* (19–3): Facial photographs after placement of a maxillary bridge and lip–nose revision. (*6*) Severe loss of palatal vault space. *7* (18–0): The labile plate of bone overlying the right central incisor exfoliated because of the loss of blood supply. The posterior palatal blood supply was cut off during the soft-palate pushback procedure. The central incisor had to be extracted. *8* (18–0): A temporary prosthesis replaces the missing incisors and maintains the correct arch form

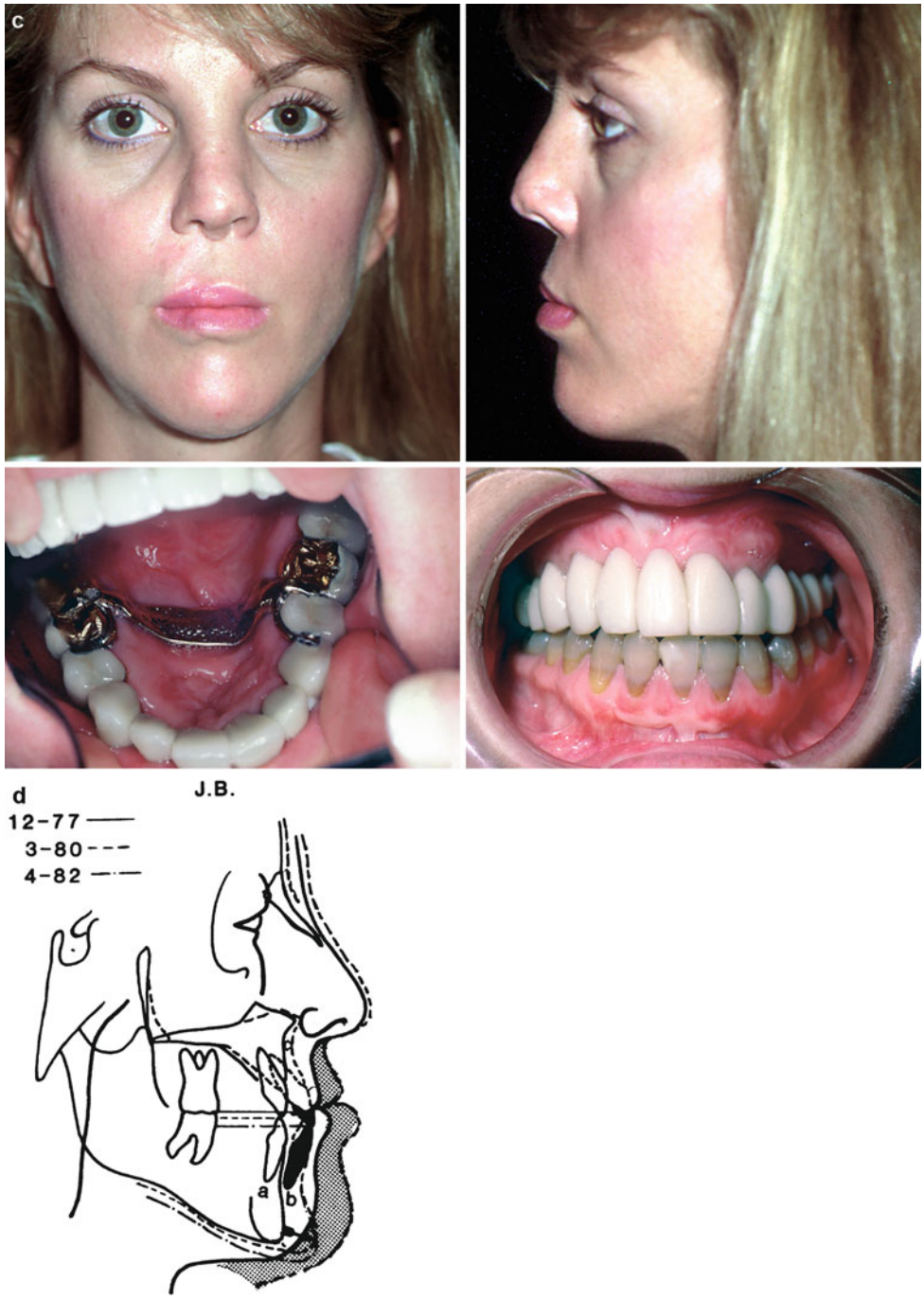

Fig. 16.16 (*continued*) (**c**) Postsurgery after Le Fort I maxillary advancement to correct a retrusive midface. This followed the loss of right lateral central incisors due to blood deprivation to the area. *1* Frontal and *2* lateral facial photographs. *3* Transpalatal removable cast goal arch appliance to help maintain maxillary arch size and form. *4* "Round-house" bridge to maintain a functional occlusion and improve aesthetics. (**d**) Superimposed cephalometric tracings, pre- and postmaxillary advancement, show changes to the skeletal and soft tissue profile. The upper lip is more recessive than the lower lip due to the lack of maxillary basal bone coupled with additional mandibular growth. Comment: Growth disturbance caused by severe palatal scarring is three-dimensional. Although palatal osteotomies can reposition the bony segments, the force of scar contracture will prevail, causing arch collapse if it is not counteracted. This can only occur with dental bridges of various types and/or transpalatal struts (McNeil 1950)

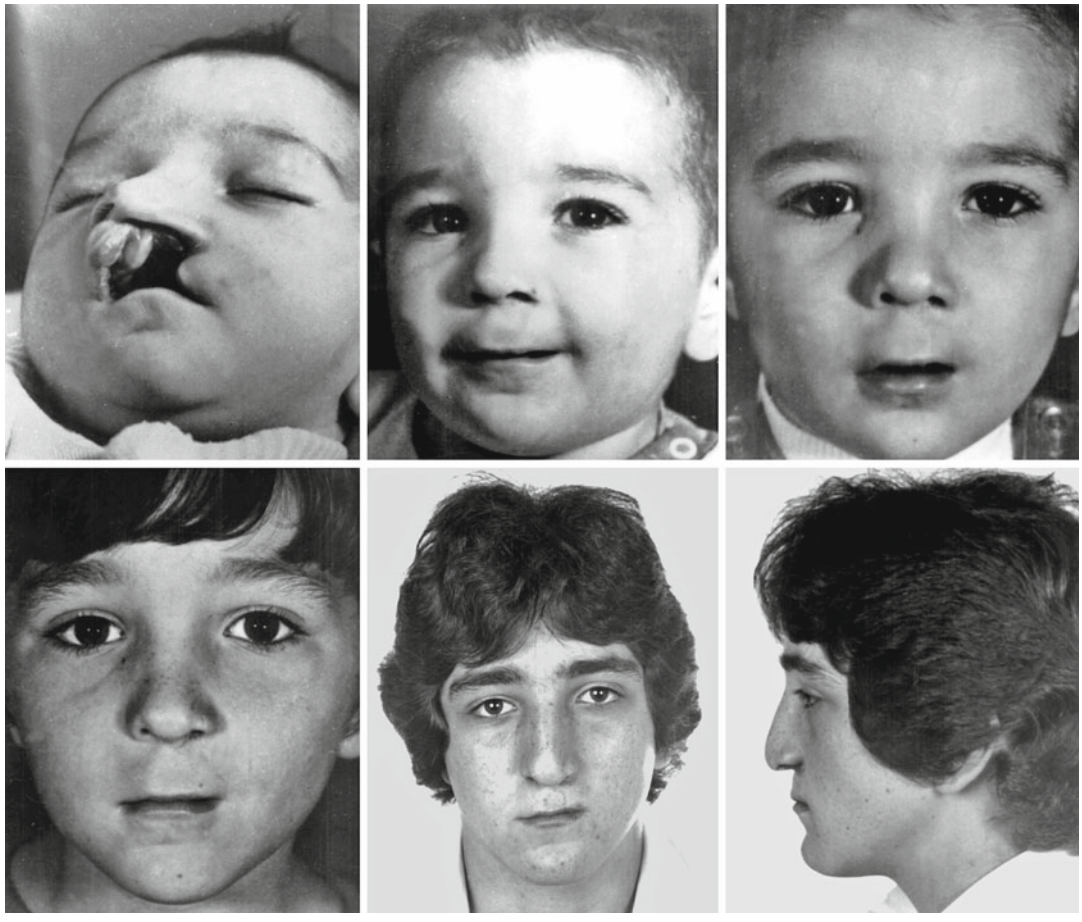

Fig. 16.17 Case TO X-26. Facial changes from birth to the mixed dentition do not usually show the effect of midfacial growth retardation. The effects on facial growth become more evident after the postpubertal facial growth period. At birth (1), 10 months (2), 3 years (3), 6 years, and 9 months (4 and 5). Frontal and lateral views at 17 years (6)

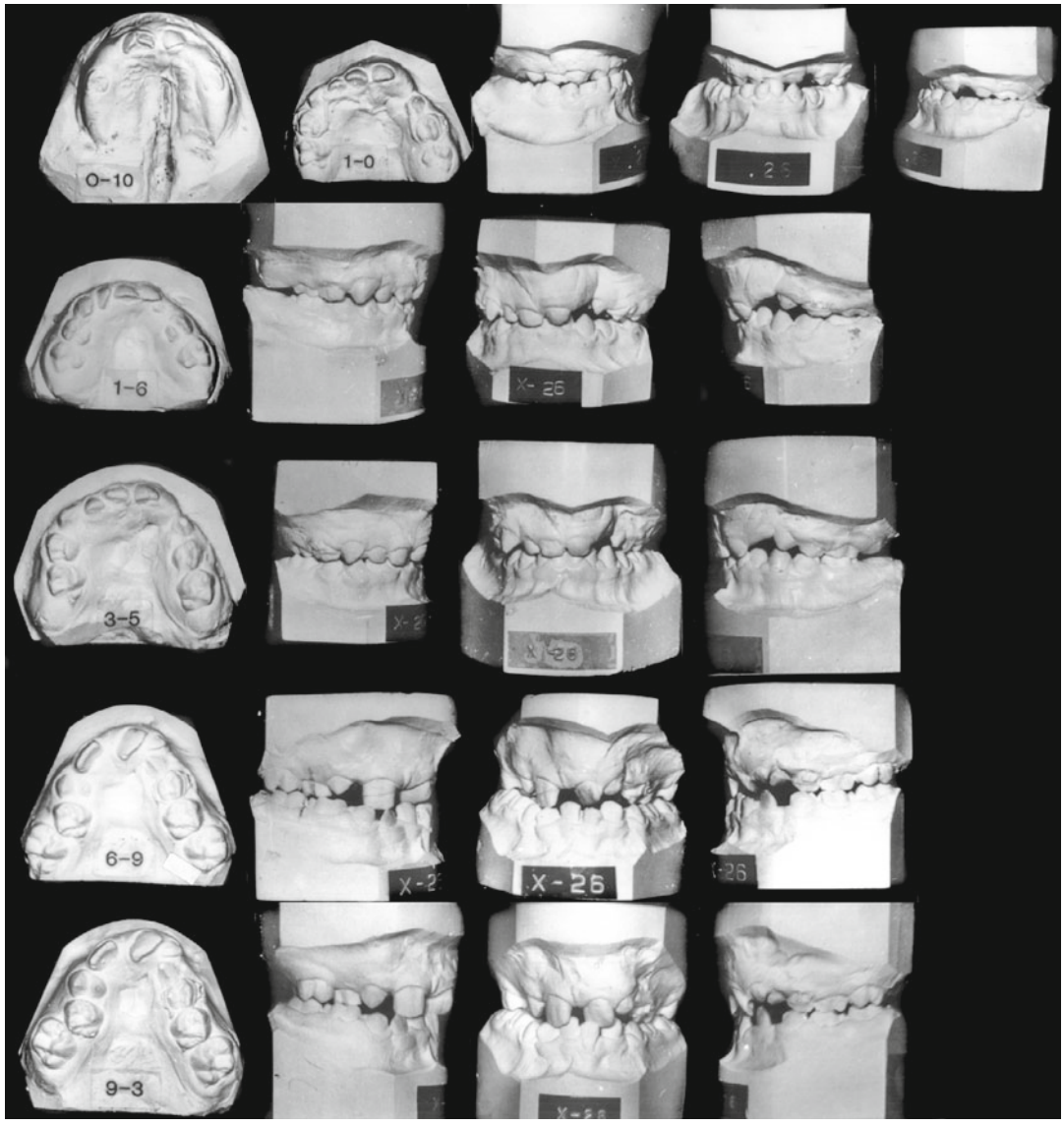

Fig. 16.18 Demonstrates the deleterious effects of excessive scarring associated with an island flap on palatal growth, and arch form in a CUCLP. Palatal casts. 0–10: After lip surgery which brought the alveolar segment is in good approximation. A small palatal cleft and good arch form before the island flap. 1–0: After the island flap and resulting collapse of the palatal segments. The child had to protrude the lower jaw to obtain a comfortable posterior occlusion. 1–6: Anterior and buccal occlusion. 3–5: Palatal deformation becomes more apparent, leading to left buccal crossbite. 6–9: Marked palatal form changes reflecting growth retardation. Further palatal and occlusal changes between 9 and 3

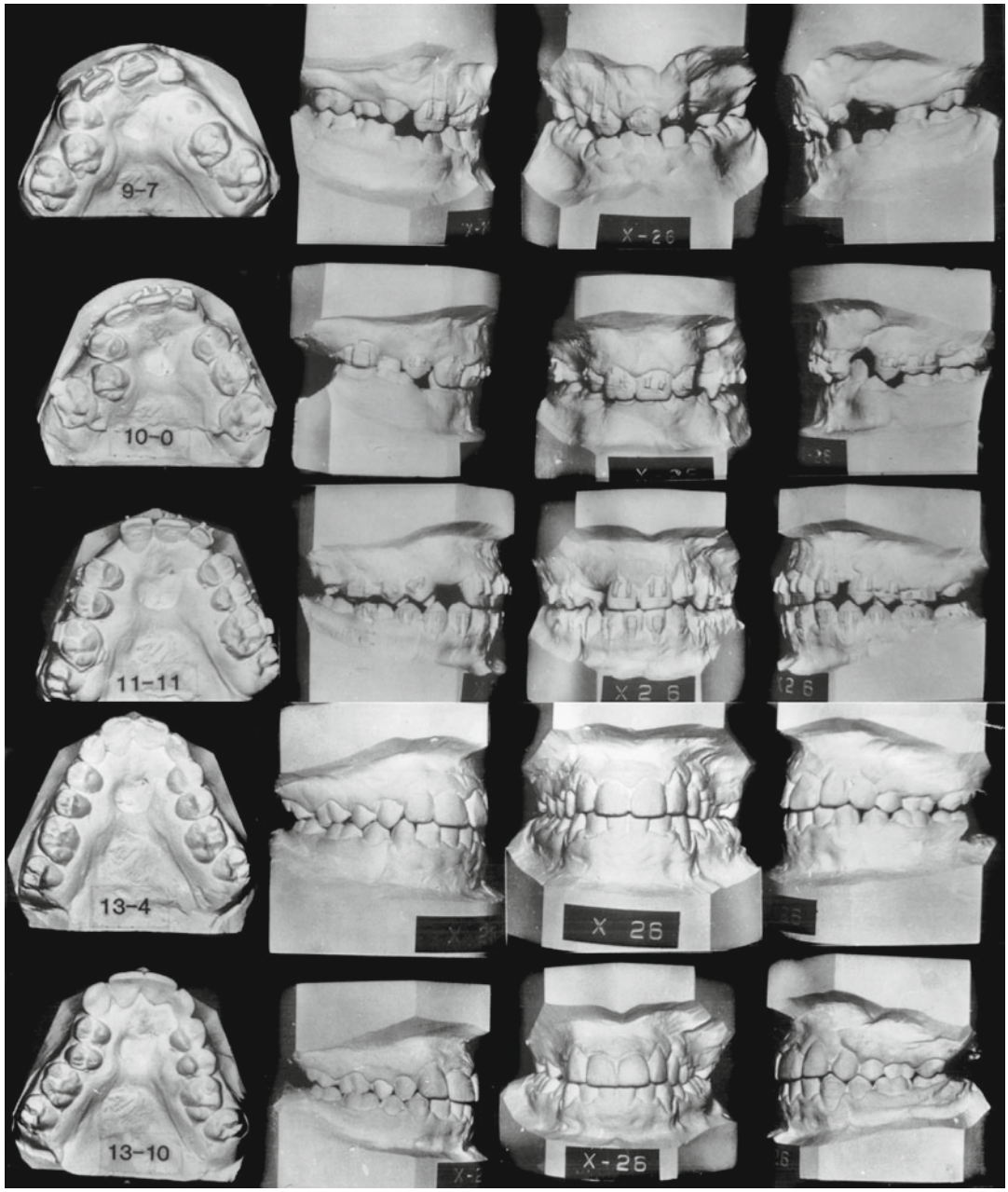

Fig. 16.18 (*continued*) 9–7 and 10–0 are evident. The strong transpalatal scar contracture narrows the transverse palatal width and reduces palatal growth leading to severe dental crowding. 11–11: Orthodontia was able to temporarily produce good palatal arch changes and improve the occlusion. 13–4: Both maxillary cuspids were transposed to the lateral incisor spaces. 13–10: Again the transpalatal scar tissue caused the arch form to narrow across the bicuspids and first molars

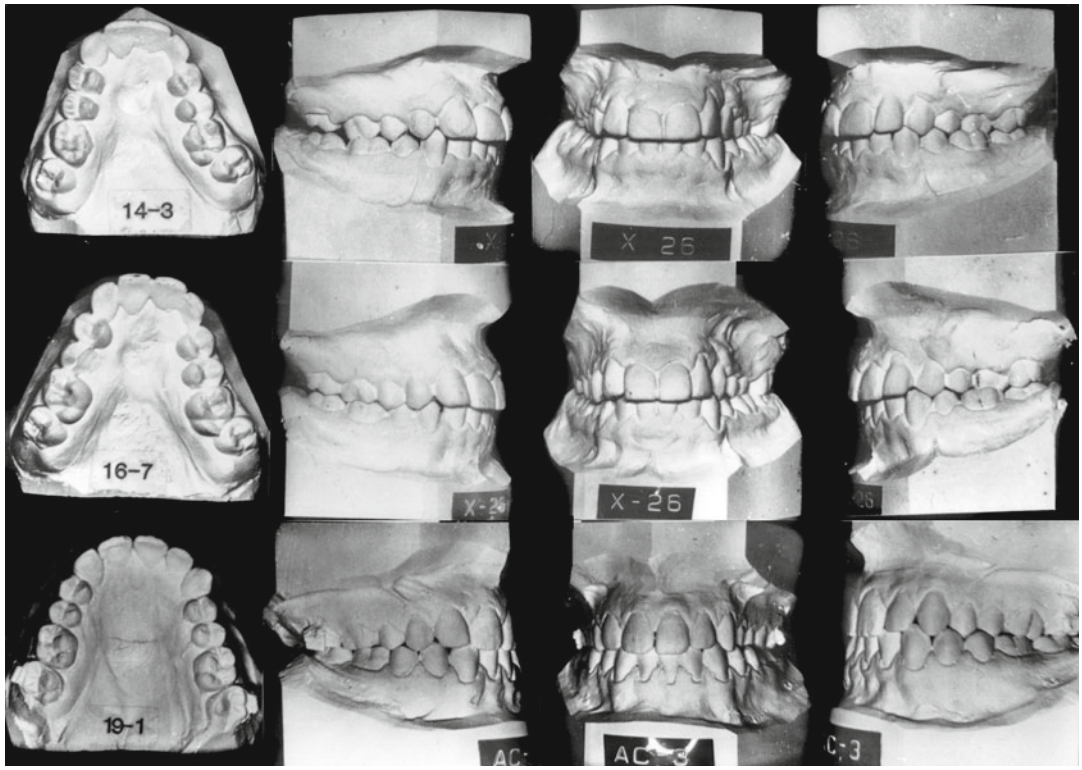

Fig. 16.18 (*continued*) 14–3: The narrowed palatal arch width stabilized with a good anterior overjet and overbite remaining. 16–7 to 19–1: The maxillary cuspid was positioned in the missing lateral incisor space. Millard discontinued using this palatal closure procedure as a result of the transverse scarring and the problems it created

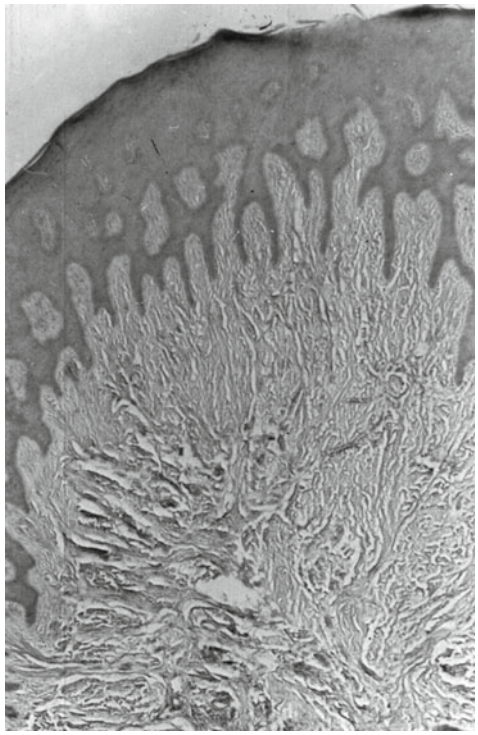

Fig. 16.19 Biopsy of transpalatal scar showing acellular band of fibrous tissue

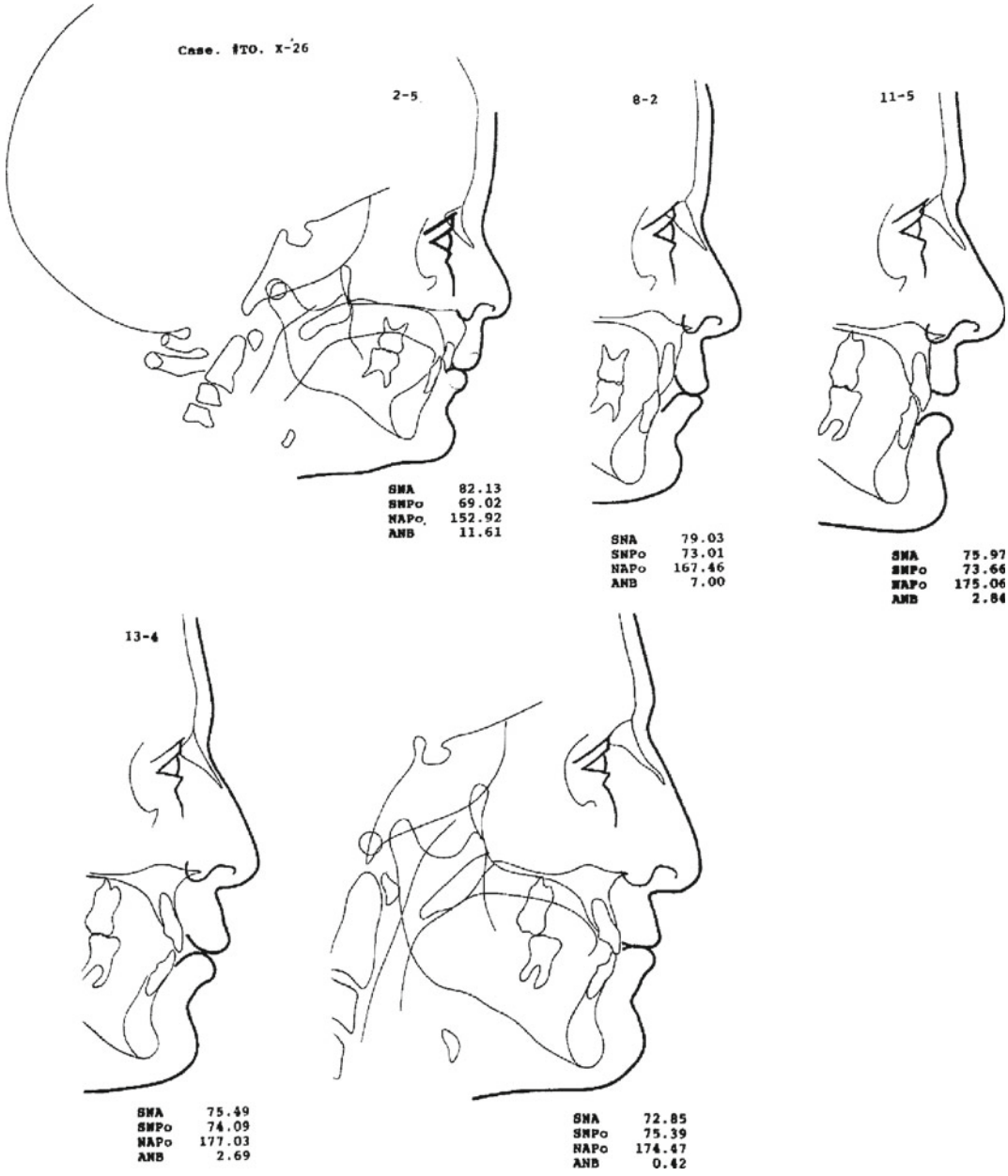

Fig. 16.20 Lateral cephalometric tracings for Case TO (No. X-26) between 2–5 and 18–0. The mandibular prominence (SNP) increased from 69° to 75°, while the midfacial protrusion (SNA) decreased from 82.13° to 72.85° during the same time period reflecting the effects of midfacial growth retardation

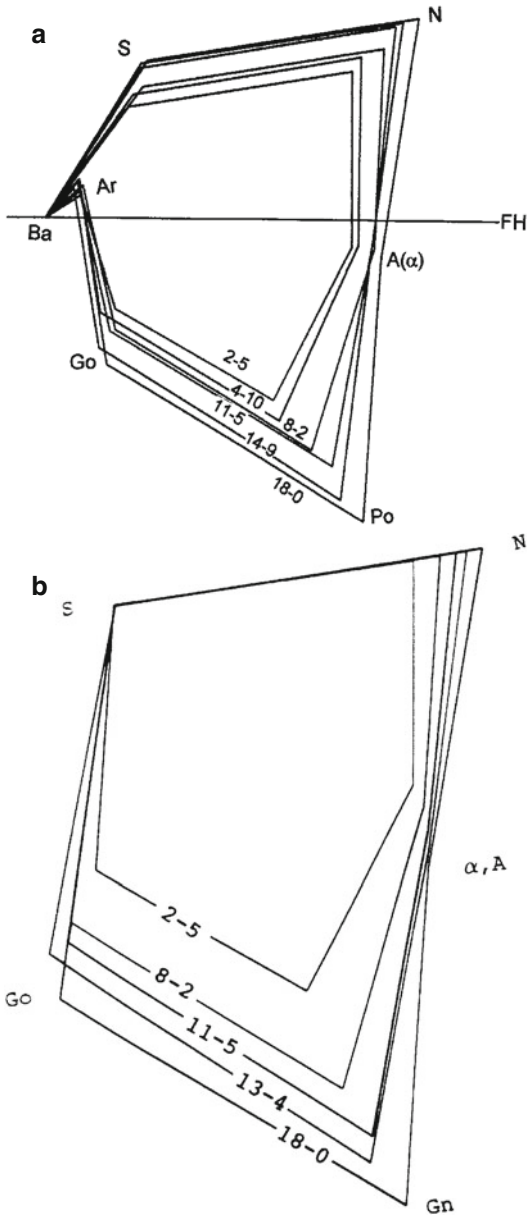

Fig. 16.21 (**a**) Superimposed polygon tracings for Case TO (No. X-26) using the Basion horizontal method of Coben. Changes from 2–5 to 18–0 clearly show retarded midfacial growth when compared to normal growth of the upper and lower portions of the face. The soft tissue profile is able to mask the skeletal discrepancy in this instance. The upper, middle, and lower parts of the face are growing normally, creating a very aesthetic face with excellent occlusion. (**b**) Serial cephalometric tracings with its facial polygon. Tracings were superimposed on the anterior cranial base (SN) and registered on *S*. These tracings show extensive growth at the anterior cranial base and mandible. There was some midfacial growth up to 8 years but very little. Therefore, all goals of good facial aesthetics, occlusion, and speech were achieved

18 and 24 months of age, lists an absence of distal molar occlusion with a minimal number of anterior crossbites and only a few complete buccal crossbites. By comparison with studies that do not use vomer flaps, one can conclude that the obvious cause for the deleterious occlusal results in the Dahl et al. (1981) study lies in the timing and not the surgical procedure utilized because Millard (1980) also utilizes a similar vomer flap with success. This conclusion does not mean that all cleft closure procedures need to be delayed until 18 months of age since narrow clefts in the palate can exist before 12 months of age. A narrow cleft suggests that less scar tissue will be created. The age of surgery is a primary variable in determining the effect of surgery on palatal development only when the width of the cleft space is also placed into the equation.

16.7.1 The Fourth Dimension of Time: Catch-up Growth

Substantial evidence from the many excellent facial and palatal growth studies (Figs. 16.22 and 16.24; Table 16.1) has determined that a "catch-up" potential exists in children with cleft lip and/or palate, which allows for reasonably good growth of the maxillary complex and face. Inherent factors alone, however, do not determine this growth. Clinicians are cognizant of the very strong influence – which quite often is adverse – of extraneous factors (e.g., iatrogenic and functional). These factors are complex and varied. It becomes obvious that surgical skill, the type of surgery, the timing of surgery, and even the sequence and numbers of surgical procedures all complicate the overall end result of facial growth in any given situation. It would appear that just the primary surgical closure of the lip and palate produces a myriad of sequences and variables to be dealt with, and they in turn introduce more sequences and variables.

To take into account both the inherent insult and extraneous factors, and to recognize and quantify the extent to which each contributes to the overall severity of the problem, is a much needed step in the correct rehabilitative effort. The inherent problem, such as the anomalous cleft, should of necessity emphasize rehabilitative

efforts with minimal adverse effects on growth. At present, there is no consensus as to how this could be achieved, a fact attested to by the wide variety of successful treatment regimens employed throughout the world. A long-term assessment of the approach to this problem has been brought about by the use of objective serial clinical records, such as serial dental casts, by research consortium members working together (Berkowitz et al. 2005).

From Berkowitz's serial palatal growth studies, he has deduced that the intrinsic growth potential of the maxillary processes and basal bone in cleft palate patients on the whole is slightly less in all three dimensions than in non-cleft children. Various cleft types have different degrees of osteogenic deficiency, with bilateral clefts potentially being the most deficient at birth and remaining that way even as the palatal processes grow and develop. It has conclusively been shown that surgical trauma with developing scar tissue at an early age can further inhibit palatal bone growth and its forward translation within the face (Ross 1987a, b).

The structure and function of the physiological elements involved in cleft lip and/or palate, as previously explained, are not static. They exist not only within a framework of three-dimensional space but also in a functional continuum of time. This fourth dimension of time involves the modifications induced by the processes of growth and development, which determine the ultimate nature of the congenital defect. To understand time–space relationships as they affect the individual cleft palate patient requires a longitudinal study of the patient by means of casts, photographs, and cephalometric roentgenograms.

Catch-up growth has been defined as growth with a velocity above the statistical limits of normality for age during a defined period of time.

Table 16.1 Case CM (AC-33). Surface area of CUCLP from 21 days to 15 years and 5 months of age

	Skeletal area			Cleft space total	
Age	RLS	LLS	Total	Post	SA + CS
0–0–21	474.7	259.3	734.0	364.9	1,098.9
0–3	567.6	389.6	957.2	223.1	1,180.3
0–7	611.3	407.7	1,019.0	187.5	1,206.5
1–2	682.5	448.4	1,130.9	141.7	1,272.6
1–9	743.2	510.4	1,253.6	75.6	1,329.2
3–0	775.3	552.1	1,327.4	13.9	1,341.3
3–5	809.6	570.8	1,380.4		1,380.4
3–10	814.3	606.5	1,420.8		1,420.8
4–6	825.5	618.9	1,444.4		1,444.4
5–0	855.1	654.4	1,509.5		1,509.5
5–7	890.0	698.8	1,588.8		1,588.8
6–5	917.3	718.4	1,635.7		1,635.7
7–3	1,033.5	857.7	1,891.2		1,891.2
8–4	1,103.6	875.8	1,979.4		1,979.4
8–10	1,116.6	888.7	2,005.3		2,005.3
9–4	1,157.8	918.8	2,076.6		2,076.6
9–9	1,168.0	924.4	2,092.4		2,092.4
11–4	1,204.9	938.8	2,143.7		2,143.7
12–4	1,223.3	962.6	2,185.9		2,185.9
13–5	1,366.9	1,197.5	2,564.4		2,564.4
14–2	1,448.8	1,231.7	2,680.5		2,680.5
15–5	1,575.8	1,357.7	2,933.5		2,933.5

These numbers were used to create the growth chart in Fig. 16.25

Note: *RLS* right lateral segment, *LLS* left lateral segment, *Tot* total surface area, *SA + CS* bony surface area + cleft space area

Such an increase in the rate of growth, before and after palatal surgery, with or without neonatal maxillary orthopedics, may allow the palate to attain greater size, but it may still be smaller than normal adult size. In the latter case, it is called "incomplete" catch-up growth. The duration and severity of the insult (the surgical procedure used to close the cleft space in the hard palate) may positively or negatively affect the ability of the palate to recover and undergo catch-up growth.

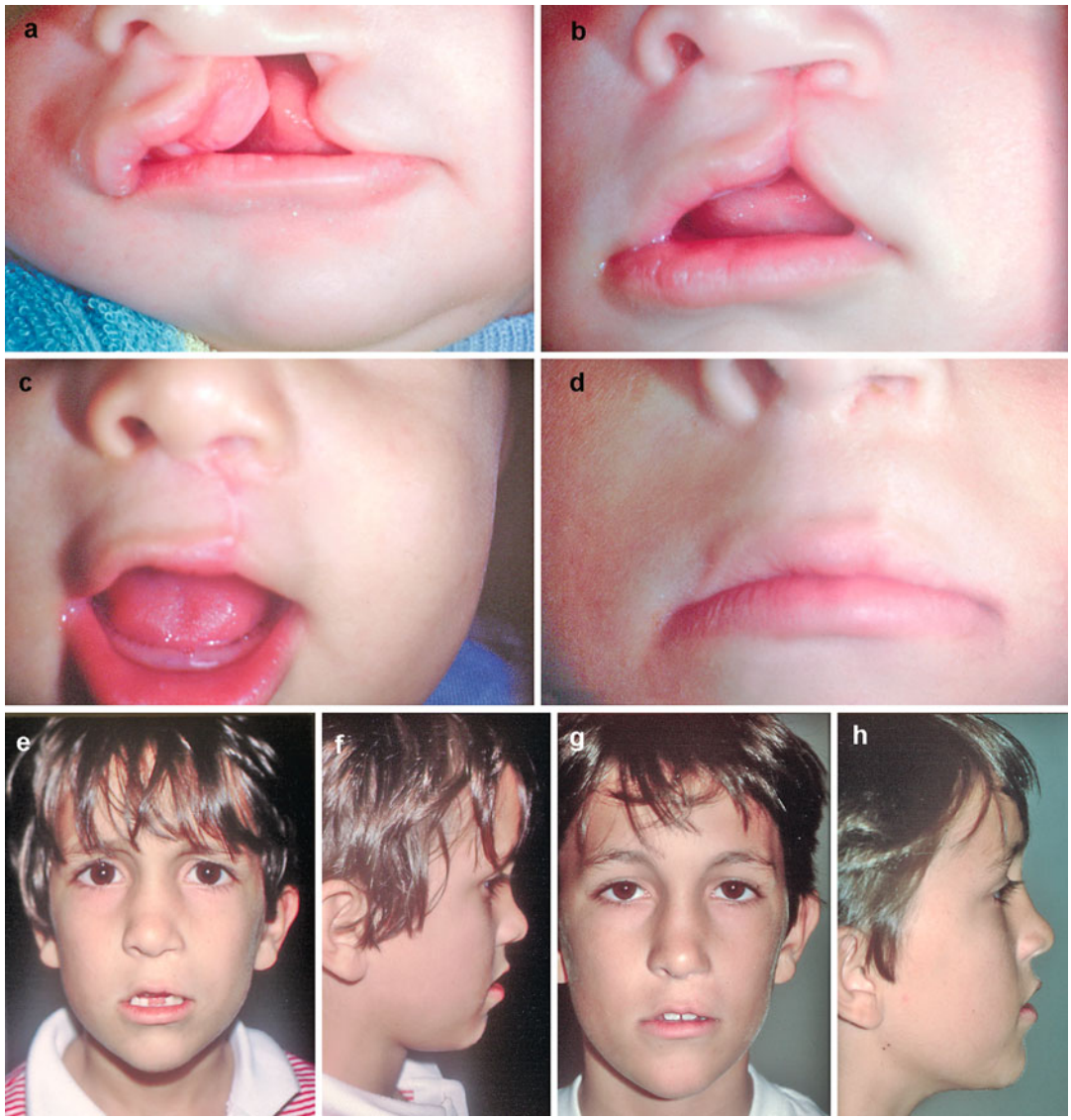

Fig. 16.22 (**a**–**r**) Case CM (AC-33) illustrates excellent palatal growth in a CUCLP treated conservatively without presurgical orthopedic treatment. Lip closed with a Millard rotation advancement at 7 months. Soft palate united at 16 months, and the hard palatal cleft closed at 24 months of age using a modified von Langenbeck. (**a**) Newborn: 24 days. (**b**) After lip adhesion at 3 months. (**c**) After definitive lip surgery at 7 months. (**d**) 4 years, 10 months. (**e**, **f**) 5 years. (**g**, **h**) 8 years, 4 months

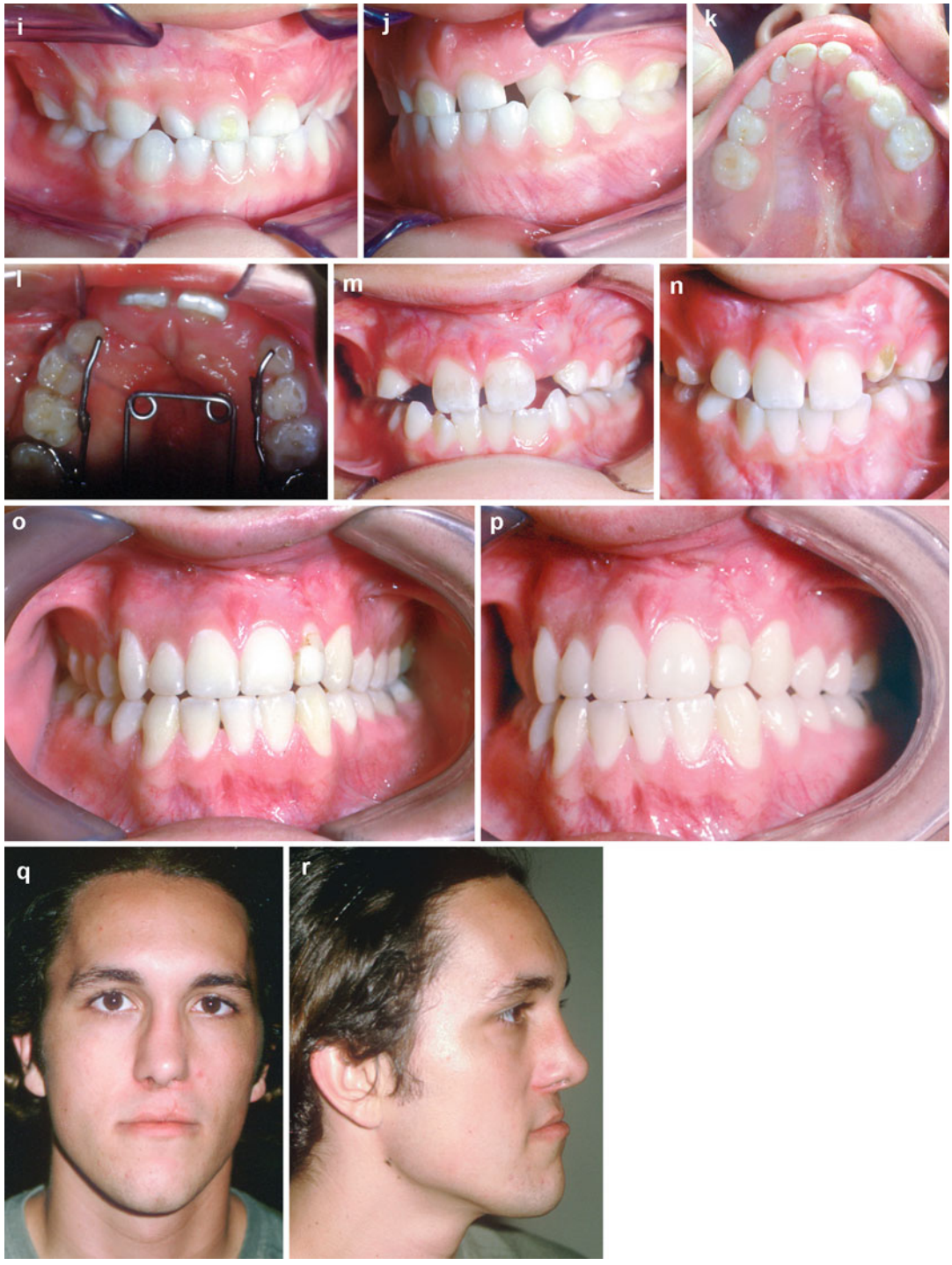

Fig. 16.22 (*continued*) (**i**, **j**) Left cuspid crossbite. (**k**) Palatal view showing mesioangular rotation of the left palatal segment. (**l**) Palatal expander after expansion. (**m**) 17 years. (**n**) 18 years. (**o**–**r**) 17 years. Comments: A mandibular central incisor was extracted to allow a proper overjet-overbite relationship. Even with the extraction of a mandibular incisor, it was difficult to create more of an incisor–overbite relationship due to the relative palatal midfacial recessiveness. Note the short midfacial vertical dimension with a relatively long lower facial height

16.7.2 The Need for Differential Diagnosis

Diagnosis and surgical treatment planning in both medicine and dentistry are most frequently dependent on the patient's age and nature and extent of the tissue's defect. In cleft lip and palate, the timing for surgically closing a cleft palate has been traditionally based solely on the age of the patient and the onset of speech (usually between 6 and 8 months) irrespective of the physical assets and defects of the affected tissue, that is, the relative size of the palatal cleft defect to that of the surrounding palatal tissue.

The inability to develop quantitative diagnostic criteria to facilitate a differential diagnosis for proper treatment planning is partially due to cleft palate research programs having an insufficient number of investigative data obtained from serial maxillary and mandibular dental casts of patients starting at birth and extending into adolescence, as well as a proper instrument to measure the palatal cast's surface. Berkowitz et al. (2005) formed a consortium of four European and three American cleft palate centers to determine the relationship of the size of the cleft defect to palatal size at the time of surgery of patients that grew well into adolescence, yet they all used different surgical protocols. The premise of this investigation is that data extrapolated from serial cast records will establish the relationship of the cleft defect to the palatal size and shape under the influence of corrective surgery.

Clinicians who do not have serial cast records have failed to appreciate the importance of cleft size/shape variations that exist within each cleft type at various ages, which may be crucial for making the proper decision as to when to surgically close the cleft space to avoid growth-inhibiting scarring.

16.7.3 Timing of Palatal Closure Based on the Ratio of the Palatal Cleft to the Palatal Size

A multicenter serial 3D study of the palatal mucoperiosteum medial to the alveolar ridge analysis was made of complete unilateral cleft lip and palate (CUCLP) and complete bilateral cleft lip and palate (CBCLP) casts' growth (size and velocity) changes from birth through adolescence of cases that demonstrated good occlusion, facial aesthetics, and speech. Only ones showing good outcomes were selected in order to determine which surgical protocols (timing and type) collectively produced these outcomes and what was the relationship of the size of the cleft defect to the size of the palatal segments medial to the alveolar ridges at the time of surgery. Poor outcome cases were not included because they could have been due to the surgery performed and not related to the relative size of the cleft to the palate medial to the alveolar ridge. This study was to focus on the two variables, the size of the cleft space and the size of the palate that supplies the mucoperiosteum for surgical closure. The source and number of centers in the retrospective study were the cleft palate clinic at the University of Miami School of Medicine (CBCLP, CUCLP); University of Illinois (CBCLP, CUCLP); Nijmegen (CBCLP, CUCLP); Goteborg delayed series (CBCLP, CUCLP); Goteborg vomer series: (CBCLP, CUCLP); and Northwestern University (CBCLP, CUCLP). The Amsterdam cases were followed from birth for 48 months to eliminate the effects due to palatal surgery. This study focused on the influence of presurgical orthopedics (PSO) at Amsterdam (CBCLP, CUCLP) and Rotterdam (CBCLP, CUCLP) which were treated with the Hotz PSOT protocol.

An exception to the well-growing cases in the Goteborg series is the Goteborg vomer-flap cases which were included because the palate and faces developed very poorly. The purpose of including this series is to compare the influence of vomer flap surgery on the palatal growth rate and size (2 mm) and to determine if palatal maldevelopment (size and velocity) values are significantly different from well-growing palates.

The control series consisted of mixed gender clefts of the lip and alveolus and/or soft palate with no palatal clefts. The growth data was the standard to which all the cleft palatal casts series were compared.

Serial palatal casts were surveyed using a highly accurate electromechanical digitizer to

extrapolate surface data in millimeters and to be analyzed using special CAD/CAM software (CADKEY). The morphometric palatal growth (size and rate of change) was compared.

Results: Berkowitz (1985) concluded that:

1. Goteborg which had delayed palatal closure and Miami and other institutions which had closure at an earlier age reported similar palatal growth rate. The various clinic's cases growth (size) were less than that of the control series.
2. The 3D surface area data identified quantitative palatal parameters before palatal cleft closure that can be used as criteria for establishing a scientific basis for determining when to close the palatal cleft space.
3. Increased palatal scarring, as seen in the Goteborg vomer series, diminishes palatal growth.
4. Delaying all cleft closure surgery until 5+ years of age is unnecessary to maximize palatal growth.
5. Based on excellent palatal growth outcomes, the best time to close the palatal cleft space is when the palatal cleft size is 10 % or less than the total palatal surface area bounded laterally by the alveolar ridges.
6. The 10 % ratio generally occurs between 18 and 24 months but can occur earlier or later in some instances.
7. There is more than one good type of palatal cleft closure surgery (Berkowitz 2005).

No studies have conclusively shown that palatal growth can be accelerated by orthopedic appliances as Weil (1987) claimed that "physiological" surgery with the establishment of functional occlusion will accelerate bone growth. Yet, Berkowitz (1985), Krogman and associates (1975), and Cooper et al. (1979) suspected that catch-up growth must happen in some clefts after surgery due to the amount of additional growth that occurs, as measured by changes in palatal surface area (the area between the alveolar ridges and limited posteriorly by the end of the hard palate).

Berkowitz's palatal growth charts have shown a marked increase in palatal surface area in the first 2 years before palatal surgery. After palatal cleft closure between 18 and 24 months, there appears to be a growth hiatus, creating a plateau in the growth graph; however, 6–12 months post-surgery, there is frequently a second period of growth acceleration which extends to 6 years of age. The palatal surface area continues to increase as the molars develop and erupt into the arch.

Berkowitz and Millard palatal cleft closure timing is based on the size of the cleft defect not fixated on the patient's age alone. Some surgeons have found a "middle of the road" treatment plan to be around 12–18 months of age and discount the size of the cleft space.

16.8 Good Speech Is Dependent on a Palate of Relatively Normal Size and Shape

Sally Peterson-Falzone et al. (2000) have written that malocclusion needs to be considered during the early speech learning years. They point out that the dental and orthodontic literature contains fairly consistent information regarding the effects of dental problems and malocclusions on speech.

A Need For Differential Diagnosis and Treatment Planning: A careful review of cleft palate surgical history makes it clear that a single mode of surgery based on age alone for all cases frequently results in severe palatal and midfacial deformities, as well as poor speech development.

In general, this literature tells us that dental and occlusal problems are more likely to be causative factors in speech problems (1) when they occur in combination rather than singly, (2) when they are present during the speech-learning years as opposed to later years, and (3) when they influence the spatial relationship between the tip of the tongue and the incisors (Peterson-Falzone et al. 2000). The literature also indicates that speech problems are fairly common when there is a restriction in the size of the palatal vault, which is more apt to be found in Class III occlusions compared with Class II (Peterson-Falzone et al. 2000). Children with clefts are obviously vulnerable to restriction in size of the palatal vault and the possibility of Class III occlusions due to the

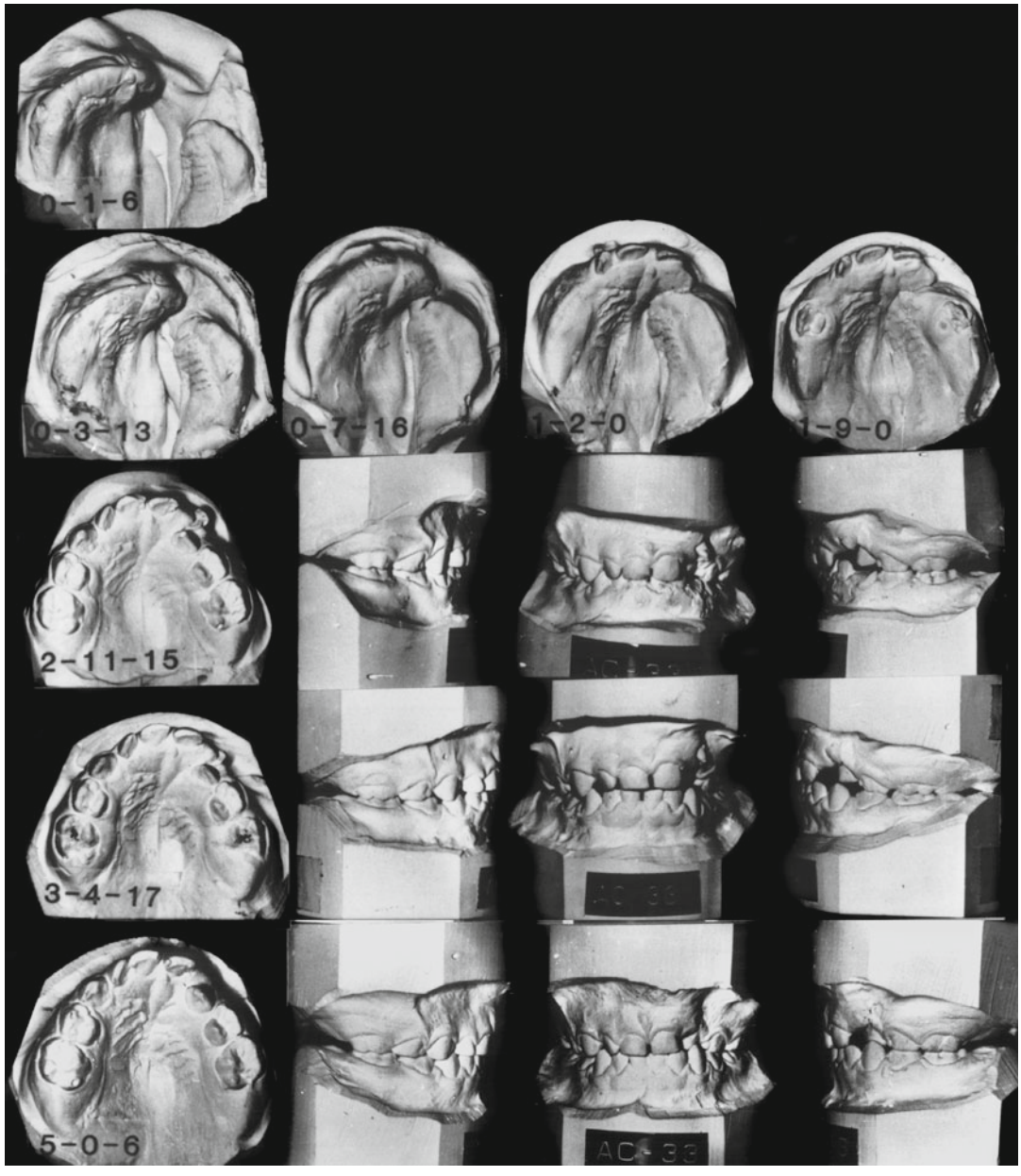

Fig. 16.23 Case CM (AC-33) serial dental casts. 0–1–6 at birth, 0–3–13, 0–7–16, 1–2–0, and 1–9–0. With lip closure, the palatal segments moved together with a slight overlap of the smaller cleft segment by the noncleft segment. 3–4–17, 5–0–6, and 5–7–0. Only the left deciduous cuspid is in crossbite

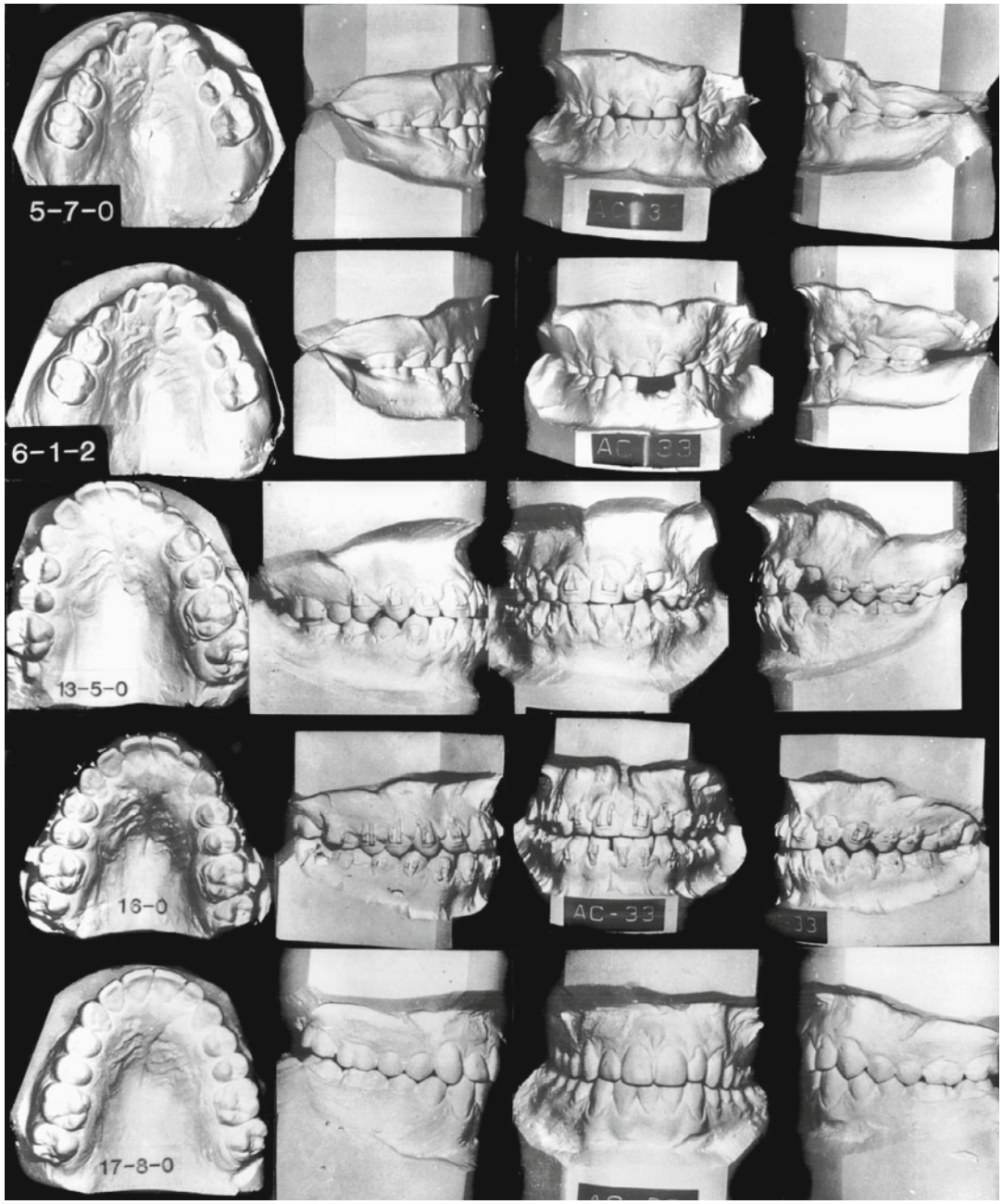

Fig. 16.23 (*continued*) 6–1–2. Excellent vault space to accommodate the tongue, as attested by the good dental occlusion. The deciduous cuspid crossbite did not cause a functional problem nor did it inhibit palatal growth of the lesser segment. 13–5–0. The left lateral incisor and cuspid erupted through the secondary alveolar bone cranial bone graft performed at 8 years and 5 months of age. Tip-to-tip anterior occlusion. A lower central incisor was extracted to permit a proper overjet–overbite relationship to be established. 16–0 and 17–8–0. Excellent anterior and buccal occlusion with ideal palatal vault space

presence of dental or occlusal problems (possibly several at one time) during the speech learning years. The question is will the speech problems diminish as the dentition or occlusion improves?

This statement convincingly acknowledges that good speech development is contingent on good tongue–teeth relationships within a normal vault space of proper volume.

Early palatal surgery (before 1 year of age in most instances) may not always jeopardize palatal and facial development provided conservative surgical methods are employed when the cleft space is sufficiently small.

There are no longitudinal palatal growth studies which show that overlapping palatal segments in CBCLP and CUCLP are incapable of growing and developing normally. The reverse is true! Many longitudinal palatal and facial growth studies show that overlapped palatal shelves have the potential of growing well when there is minimal scarring of the palatal mucosa. Why is such a big fuss being made by those who perform presurgical orthopedics about the need to prevent "collapsed" arches? The use of the word "collapse" has a strong negative connotation which infers that a problem exists which should be prevented from occurring, and if it does happen, it needs correction.

In fact, there are some benefits associated with the narrowing of the cleft space due to the unimpeded medial movement of palatal segments. They are: (1) the perpendicular plates of the sphenoid, which are displaced laterally in the pharyngeal space at birth, become more normally positioned after lip surgery, and (2) a reduced cleft space may encourage better tongue posturing and with it improved speech development and feeding. This still needs to be proven. Should a dental crossbite result; it can be easily corrected in the deciduous dentition using simple orthopedic forces although necessitating prolonged retention. Curiously, in many cleft palate centers, prevention of maxillary constriction (collapse) is still a major focus of attention, despite the fact it has never been shown to cause long-term difficulties associated with growth inhibition.

Unfortunately, when the clinician's treatment protocol is designed to prevent "collapse," the team's focus is diverted, and less attention is given to appreciating the heterogeneity of surgical procedures employed, different standards for selection of patients, and lack of standardization of evaluation techniques. Indeed, with a change in focus to conservative treatment planning, most surgeons now believe there will be a greater chance for improved long-term outcomes.

The developmental age of the infant at the time of the insult and the nature of the insult itself (extent of denuded bone left after surgery) will affect the ability of the infant to achieve complete catch-up growth. The case in Figs. 16.22, 16.23, 16.24, and 16.25 demonstrates the catch-up growth phenomenon. Berkowitz suggests that the effects of accelerating or restricting growth are not seen in all dimensions throughout the palate. He suggests that the features most likely to be affected have very strong clinical implications in deciding at what age the palatal cleft should be closed. It is expected that morphometric analysis of the changing form and size of the palate will permit some insight into the appropriate surgical procedures (age and type) to prevent growth retardation.

When asked at what age he operated, Pruzansky (1969) replied:

> We still hear the same question today. Is chronologic age the parameter that really matters, or is it morphologic age and physiologic fitness? We should be asking whether the tissues are adequate in quantity and quality. Is the geometric relationship of the malformed cleft parts to the contiguous anatomy favorable or unfavorable for reconstruction? What is the relationship of the malformed cleft parts to the contiguous anatomy which, in turn, may be anomalous? What are the changes incident to growth? Are the parts static in their deficiency or does this deficiency diminish in time?

LaRossa (2000), State of the Art in Cleft Palate Surgery, a two-layered, tension-free closure of the hard palate is advocated using a vomer flap for nasal lining and mucoperiosteal flaps that leave minimal amounts of exposed palatal bone. This approach seems to result in the least amount of growth retardation and a reduced incidence of fistula formation.

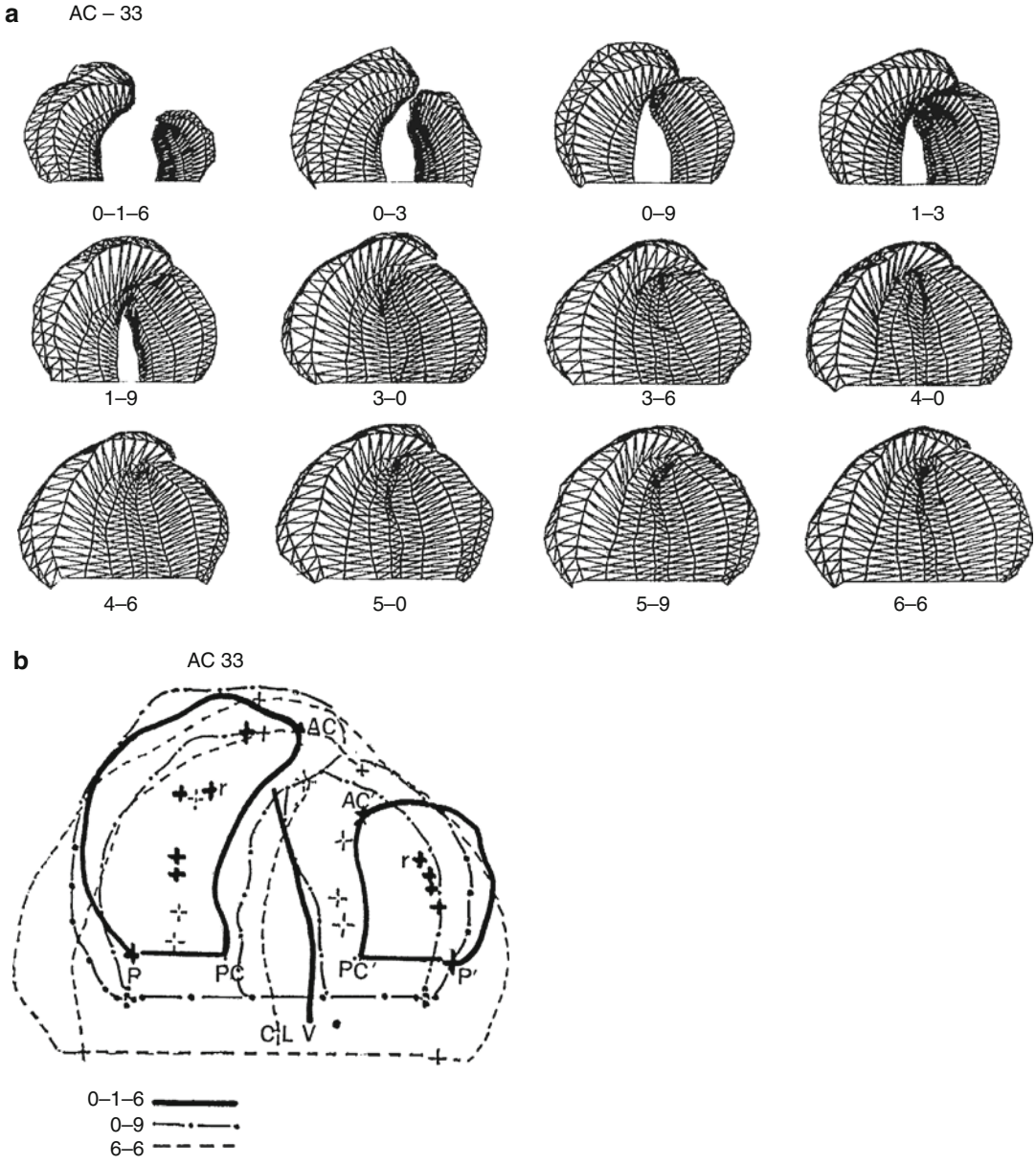

Fig. 16.24 (**a**, **b**) Case CM (AC-33) from 0–1–6 to 6–6. (**a**) Computer-generated outlines of serial casts drawn to scale. Three-dimensional serial palatal growth representation using an electromechanical digitizer and CAD/CAM software shows the degree and location of growth changes and the relative changes in cleft space size. (**b**) Superimposing computer-drawn cast tracings taken at 0–1, 0–9, and 6–6 on the palatal rugae and registered on the vomer point (*V*) as it crosses the P–P1 (postgingival point) line which is posterior limit of the hard palate. This graph demonstrates the great increase in palatal size and direction of palatal growth that occurs when physiological surgery is performed. *P* and *P′*: Postgingival. This landmark is comparable to point PTM (pterygomaxillary fissure) which is found between the maxillary tuberosity and the perpendicular plate of the sphenoid. *Pc* and *Pc′* – Landmark on line *PP′* at the cleft space. *Ac* and *Ac′* – Landmark at the anterior most point of the alveolar ridge of the cleft space. *V* – Point at which the vomer crosses the PP′ line. This figure shows palatal molding changes that follow lip adhesion. Most of the growth occurs posteriorly to accommodate the developing molars

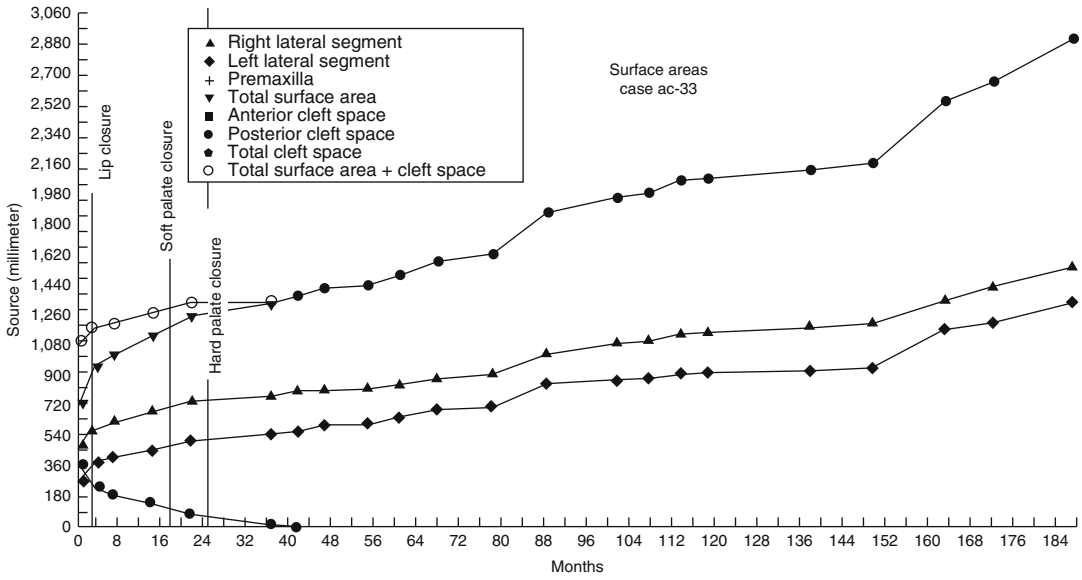

Fig. 16.25 Case CM (AC-33). The palatal growth chart shows a very rapid period of acceleration occurring the first year. During the first 8 months, the palatal surface area (bordered by the alveolar crests) increased by 45 %. Growth gradually slowed down after 20 months. Comment: Note that both lateral palatal segments grew at parallel growth rates. The growth acceleration curve after palatal surgery was the same as that before surgery. The cleft palatal segment shows two growth periods, at 80 and 152 months. Since rapid palatal growth occurs in all cleft palate children during the first year, caution should be exerted when closing the cleft spaces at this age period to avoid creating excessive scar tissue when its negative effect would be greatest

16.9 Facial Changes in Successfully Treated Cases

16.9.1 Lateral Cephalometric Results from the Oslo Team

Semb's group (Semb and Shaw 1990; Semb 1991a, b) conducted a serial lateral and frontal cephalometric study of 90 cases from the Oslo archives with bilateral cleft lip and palate. Since 1962, the treatment procedure has involved uniting the lip and hard-palate cleft space closing in two stages. No presurgical orthodontics were utilized since the surgeons and Bergland, an orthodontist and past director of the program, believe that any bilateral cleft lip can be closed without presurgical palatal manipulation.

In the period spanning 1950–1960, a von Langenbeck procedure was performed to close the hard-palate cleft between 3 and 4 years of age; after 1960, the timing of the closure was reduced to 18 months of age. Secondary alveolar bone grafting using cancellous iliac crest bone was performed prior to the eruption of the permanent canine teeth (Abyholm et al. 1981). Twenty-five percent of the updated cases needed superior-based pharyngeal flaps which were performed, if possible, before the child started school. No orthodontics were utilized in the deciduous dentition. Protraction headgear in the mixed dentition was used in one-third of the cases. Fixed retention was necessary in all cases. Results showed that (1) the maxilla progressively receded over time; (2) the mandible was retrusive with a steep mandibular plane with an increased gonial angle; (3) anterior lower-face height was elongated and posterior facial height reduced; and (4) the facial growth pattern was notably different from the normal Bolton standards: (a) male and female facial growth patterns were similar except that the males' linear dimensions were larger; (b) the prominent premaxilla would gradually realign in the preschool years; and (c) surgical premaxillary setback was never required.

The overarching thesis of this chapter favors consideration of the total emotional and physical health of the child with a cleft, based on the desired attainment of a cosmetically attractive face, and adequate dental function and respiration, as well as speech. Many surgical, medical, and dental therapies may be necessary in the best-treated cases. As long as the surgeon individualizes the treatment plan, taking care to do no harm to growing structures, all goals are obtainable.

Editor's Note Many of these procedures have been followed longitudinally and have shown to be nonphysiological. Excessive scarring resulted which led to growth interference. Also, the velum has not been lengthened. Due to scar contracture, the velum returned to its original length. This procedure has not been successful in lengthening the velum but has created excessive scarring which interferes with palatal growth and development. Although the velum has been lengthened, scar contracture returns the velum to its original position.

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Choosing the Best Time for Palatal Surgery

17

Samuel Berkowitz

Diagnosis and surgical treatment planning in both medicine and dentistry are most frequently dependent on the patient's age and nature and extent of the tissue's defect. In cleft lip and palate, the timing for surgically closing a cleft palate has been traditionally based solely on the age of the patient and the onset of speech (usually between 6 and 8 months) irrespective of the physical assets and defects of the affected tissue and not on the relative size of the palatal cleft defect to that of the surrounding palatal tissue.

The inability to develop quantitative diagnostic criteria to facilitate a differential diagnosis for proper treatment planning is partially due to cleft palate research programs having an insufficient number of investigative data obtained from serial maxillary and mandibular dental palatal casts of patients starting at birth and extending into adolescence as well as a proper instrument to measure the palatal cast's surface. The premise of this investigation is that data extrapolated from serial cast records will establish the relationship of the cleft defect to the palatal size and shape under the influence of corrective surgery.

Clinicians who do not have serial cast records have failed to appreciate the importance of cleft size/shape variations that exist within each cleft type at various ages, which may be crucial for making the proper decision as to when to surgically close the cleft space to avoid growth-inhibiting scarring.

With the advent of advanced technology to perform 3D spatial-temporal measurements and CAD/CAM computer software for in-depth analyses, extrapolated surface data from palatal casts can now be subjected to highly sophisticated quantitative analyses to perform differential diagnosis and treatment planning.

- *Hypothesis to be tested*: The hypothesis that a relationship exists between cleft size and palatal size to achieve good facial-palatal growth and speech will be tested; also, there is more than one physiological surgical procedure and the Hotz presurgical orthopedic protocol does not increase palatal size in velocity.

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17.1 Method and Material (Tables 17.1 and 17.2)

Employing the palatal casts of 242 male and female individuals from eight institutions in the USA and Western Europe, separate serial analyses were conducted of well-growing cases with excellent aesthetics, dental occlusion, and speech and a palatal control series of 18 nonpalatal cleft cases to access the growth changes in size and velocity from birth through adolescence. These control cases consisted of various clefts of the lip and alveolus and/or soft palate but with no clefts in the hard palate. The various complete cleft lip and palate series were compared to this series. Malcom Johnson (personal communication) has confirmed that this control sample is an appropriate one for the palatal growth comparisons. In this group, the midpalatal suture, extending anteriorly to the incisal papilla at the anterior alveolar ridge, served as the medial border dividing the palate into right and left segments. With the exception of the excellent sample of cases from Goteborg, the vomer series had poor occlusion, facial aesthetics, and poor speech. This series was included in order to determine what timing and type of surgical procedure had produced favorable or unfavorable outcomes and also whether the outcomes varied with the relationship of the size of the cleft defect to the size of the palatal segments medial to the alveolar ridges at time of surgery.

The participating institutions were selected on the basis of their excellent records and

Table 17.1 Number of clinics and patients involved in study

	Timing of surgery				
Clinics	Lip adhesion	Lip-nose	Second alveolar bone graft	Hard and soft palate	Palatal closure procedure
Miami	3 months	6–8 months	8–10 years	12–24 months	Von Langenbeck + vomer flap
Illinois	2–4 months	6–8 months	Seldom	12–18 months	Various procedure Wardil-Kilner, Von Langenbeck
Nijmegen	1–2 months	udp 6 months bdp 9 months	10 years	SP 12–18 months HP 6–9 years	Von Langenbeck
Goteborg-delayed	udp 1–2 months bdp 2–4 months	6–9 months	9–11 years	SP 6–8 months HP 9–11 years	Vomer flap
Goteborg-vomer	2 months	18–20 months	Average 14 years	2 months 9 months	Arterior vomer flap, posterior push back
Northwestern	6–8 weeks primary bone graft	6 months		12 months	Intravelar – veloplasty

Table 17.2 Treatment protocol and sequence

Controls = (Miami)	Soft palate + 5	Lip/alveous + 5	Soft palate lip alveous 8
Clinics	CBCLP	CUCLP	Presurgical orthopedics
Miami	18	26	None
Illinois	0	12	None
Nijmegen	10	0	Hotz 3–4 weeks for 2–3 years
Goteborg-delayed	8	24	1 weeks to 1–1.5 years
Goteborg-vomer	10	23	1 weeks
Northwestern	0	23	Posterior 1/2 palate

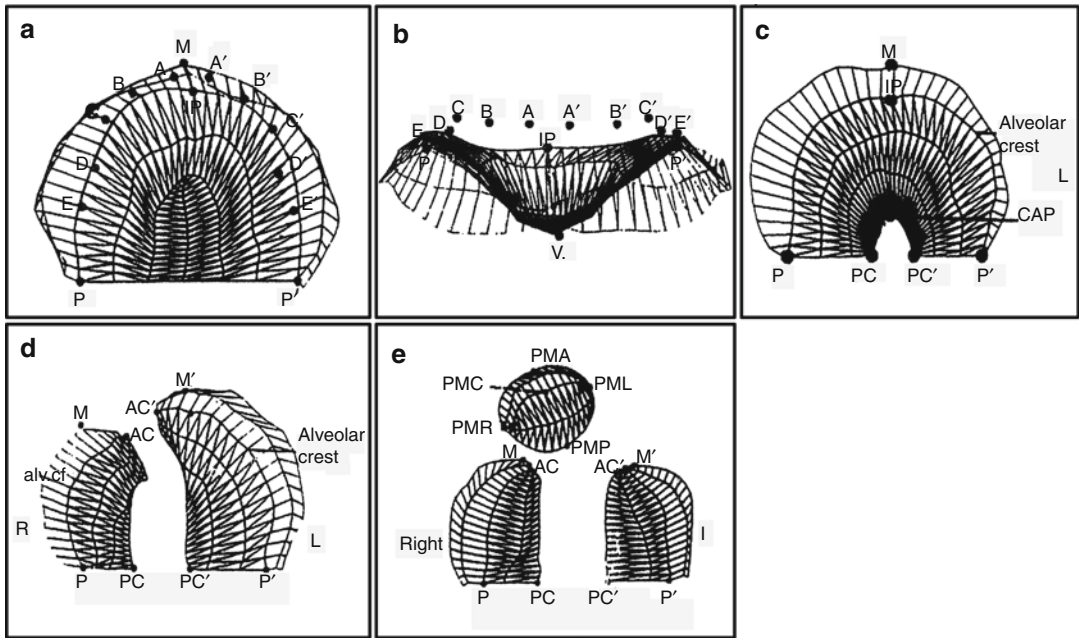

Fig. 17.1 (**a**–**e**) Various palatal forms being measured. (**a**) Normal palate, (**b**) posterior view of normal palate showing position of teeth landmark points: middle of incisal edge and central fossa of molars, (**c**) isolated cleft palate, (**d**) complete unilateral cleft lip and palate, and (**e**) complete bilateral cleft lip and palate

varied treatment protocols and different racial and mixed gender populations. The Miami sample of nonorthopedically treated cases was selected at random from a larger number of similarly treated cases.

Twelve different groupings of cases were established depending on their institutional location and type of cleft (complete unilateral cleft lip and palate, CUCLP, and complete bilateral cleft lip and palate, CBCLP), with each group consisting of both males and females. The number and type of cases were as follows: University of Miami: 26 CUCLP and 18 CBCLP; University of Illinois: 12 CUCLP; Northwestern University: 22 CUCLP; Nijmegen: 10 CBCLP; Goteborg-delayed Closure: 24 CUCLP, 8 CBCLP; Goteborg-vomer: 23 CUCLP, 10 CBCLP; Amsterdam: 26 CUCLP, 4 CBCLP; and Rotterdam: 60 CUCLP. The statistical analyses of the CUCLP and CBCLP cases were carried out separately.

The Amsterdam and Rotterdam cases, which had been treated with the Hotz presurgical protocol (PP), were followed from birth for 48 months, permitting a determination of the PP on palatal growth effects differed due to presurgical orthopedics.

- *Analysis to be made*: Comparative effects of treatment of (1) the palates surface area's rate of change (velocity) and growth (size in 2 mm), (2) size of the posterior cleft space and the velocity of its change, and (3) ratio of cleft size to the palate's size before and after surgery (total surface area). Each of the subsamples of cleft children will be compared to each other and to the age-appropriate control samples.
- *Data extrapolation from the surface of palatal casts* (Fig. 17.1): A highly accurate, 3D, electromechanical palatal cast measuring instrument which gave a measurement error of less than 5 % made possible a spatial-temporal (4D) analysis of palatal form and size changes,

permitting an in-depth study of how the cleft space and palatal segments of each clinic's cases changed in relationship with each other over time (Figs. 17.2a and 17.3).

- *Features to be measured and analyzed* (Figs. 17.1, 17.2, and 17.3): The sizes of the palatal segments are measured serially starting at birth and divided into two treatment periods. The first period ends at surgery to close the palatal cleft. The second period is after palatal cleft closure: it includes adding the remaining cleft space with the changing size of the palatal segments (total surface area). In CUCLP (Fig. 17.2a, b), the sizes of the palatal segments are limited laterally by the alveolar ridges and anteriorly by a line connecting the most anterior point on the alveolar ridge (AC and AC1). Posteriorly, a line is drawn from point gingival (P and P1) which are equivalent to the pterygomaxillary fissure seen on cephalo radiographs which marks the posterior limits of hard palate. When this transpalatal line makes contact with the cleft, points PC and PC1 are created. PC to PC1 measures posterior cleft space width. Medial border of the palatal segments (AC-PC, AC1-PC1) is limited by the cleft space. In CBCLP (Fig. 17.3), the premaxilla's surface area is limited anteriorly by its alveolar ridge (PML-PMR).

17.1.1 Method Used for Analyses

Cleft Subjects: The subjects had unequal numbers of observations and were observed at dif-

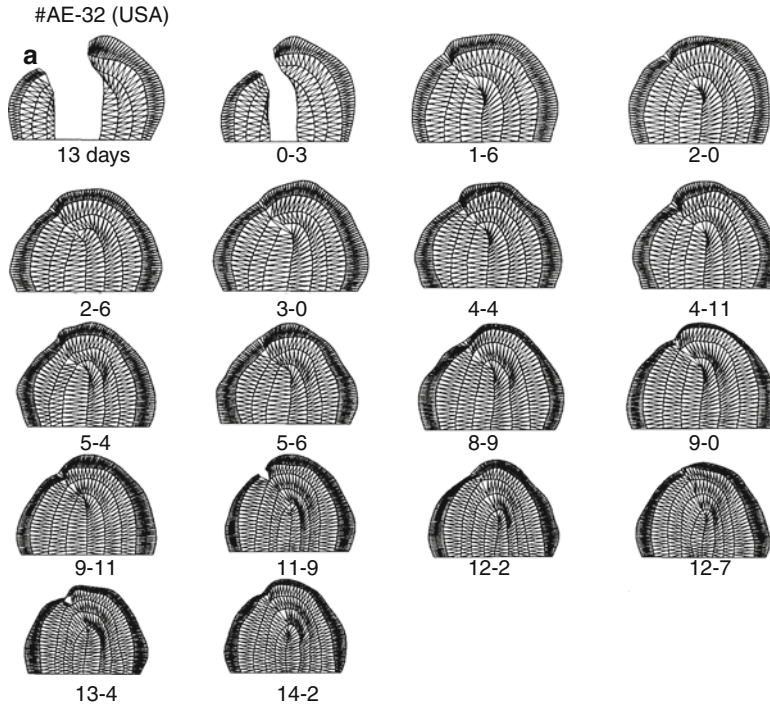

Fig. 17.2 (**a**) Twelve serial digital images of a complete unilateral cleft lip and palate from 13 days of age to 14 years and 2 months of age. The 2D images generated from 3D casts are accurate in form but are not related in size to each other. The image at birth (13 days) is followed by (0.3 and 1.6) that had undergone molding as a result of muscular contractive forces created by lip adhesion at 14 months of age. Palatal cleft closure using a von Langenbeck with modified vomer flap was performed at 14 months of age. A secondary alveolar bone graph was placed at 8 years and 3 months of age after some minor orthodontics to align the anterior teeth. No presurgical orthopedics was performed

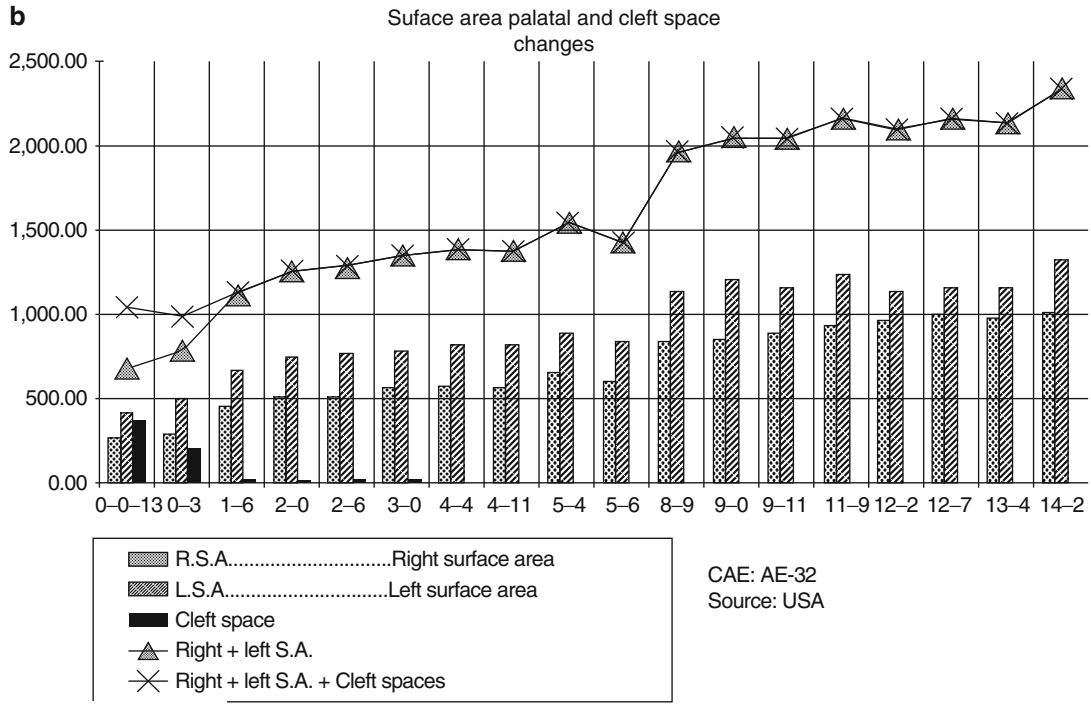

Fig. 17.2 (*continued*) (**b**) Graph depicting serial palatal 3D bony growth changes and the size of the cleft space of case 2A. The combined surface areas of the large and small palatal segments changed from 689 mm^2 at 13 days to 1,121 mm^2 at 1 year and 6 months (an increase of 39.5 %) at the time of palatal surgery. The cleft space reduced from 365 to 202 mm^2 at 3 months due to molding. Additional molding and palatal growth further reduced the cleft space prior to palatal surgical closure. Palatal growth occurred gradually up to 5 years and 6 months. The continued marked increase in palatal sizes up to 10 years and 8 months reflects posterior palatal growth to accommodate the developing molars. This posterior palatal area appears not to be affected by the palatal surgery

ferent ages. In order to compare the responses of the different groups, the data was recoded into new age intervals of 2, 8, 18, 36, 60, 84, 120, 168, and 192 months. The groups from Amsterdam to Rotterdam were observed only for 4 years and were coded as 2, 8, 18, and 48 months. Infrequently, especially in the early months, subjects would have two observations in a recoded age interval. When this happened, the mean values of the observations were used. The within-group variances were compared during subsequent analyses to ensure that the equal-variance requirement of the analysis of variance was maintained and not biased by the use of mean values in some instances.

Controls: Serial casts from 18 patients who had only clefts of the lip and/or alveolus (CLA), with or without clefts of the soft palate (SP), or clefts of the lip and soft palate (CLA and SP) alone. All of these patients did not have a cleft in the hard palate. One case series had no cleft at all. The data from the control series was handled exactly as that from the cleft series.

Statistical Analysis: A preliminary analysis of palatal area was performed using a random effects model in the SAS Proc Mixed software. This preliminary analysis showed not only group differences but, more importantly, heterogeneity in group responses over time (i.e., a significant group x time interaction). Because of the significant interaction, overall differences among groups or times are not directly interpretable. Consequently, the simple effects of differences between groups at each time point were investigated. The SAS Proc GLM software was used to test the fixed effects model

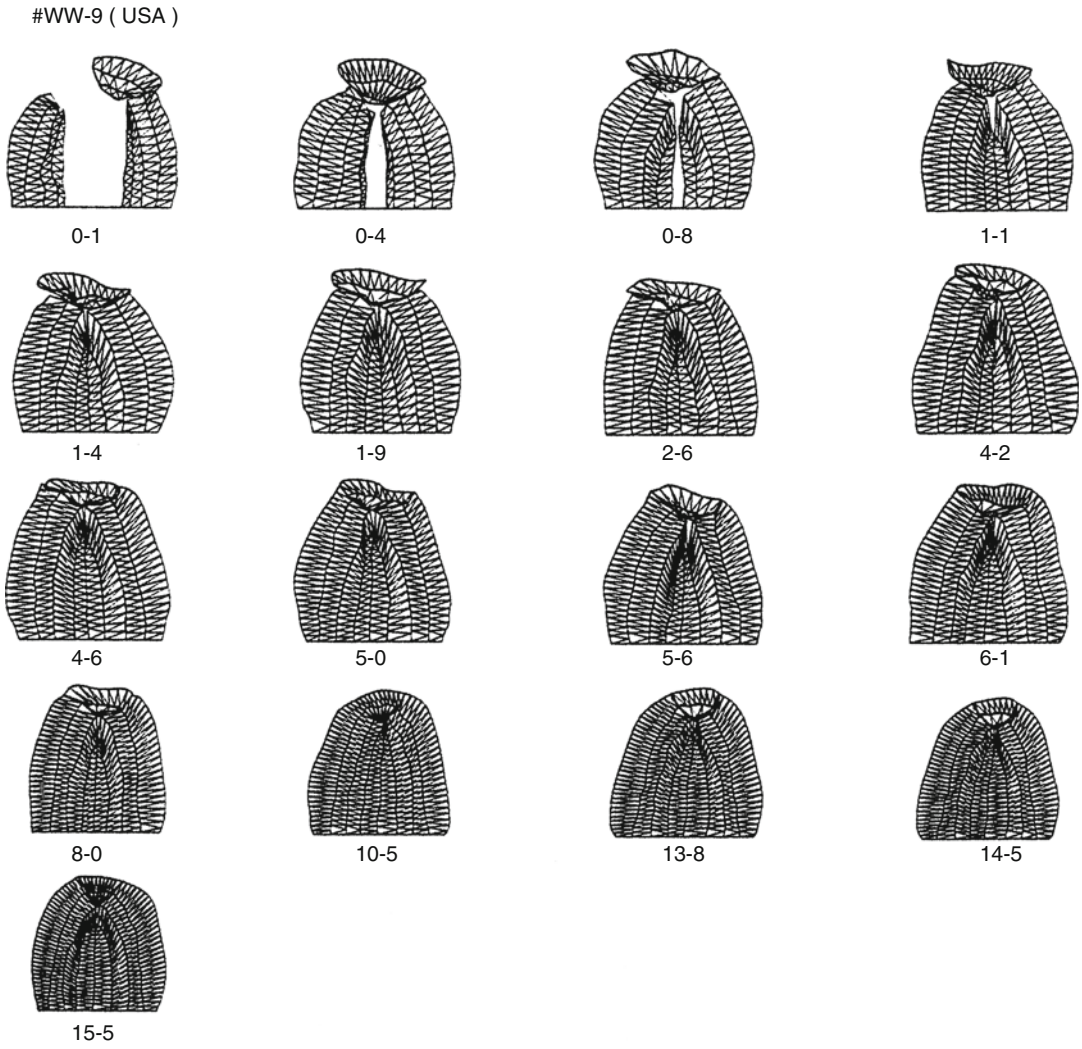

Fig. 17.3 Digitized serial 2D cast images of a one side incomplete and the opposite side complete bilateral cleft lip and palate. The serial form cast's serial changes are accurate but not so as it relates to their relative size. At 0–1 (1 month) palatal segments are laterally displaced. At 0–4 (4 months), this image shows medial movement (molding) of the palatal segments, as a result of lip adhesion. The premaxilla is positioned forward of the lateral palatal segments with marked closure of the cleft space. At 0–8 and 1–1, these images show further spontaneous closure of the palatal cleft space as a result of palatal growth. At 1–4, palatal closure was performed 1 month earlier using a von Langenbeck with a modified vomer flap. Note that the following cast images show the gradual incorporation of the premaxilla within the palatal arch with orthodontic treatment

among groups at each time, followed by a pairwise comparison among mean values. At each time point, it was sometimes possible to compare among eight groups, which would lead to 28 comparisons (some time points had fewer groups because not all groups were observed at every time point). A strict control for multiple comparisons, say by the Bonferroni procedure, would severely decrease the power to detect differences. Therefore, the alpha level for each

pair-wise test was set at 0.01. No statistical comparisons were made across time points, but the consistency of the significant differences found in the pair-wise tests was noted. In this way, biologically consistent differences could be elucidated with sufficient power.

17.2 Treatment Protocols at Each of the Centers (Table 17.2)

17.2.1 Miami Craniofacial Anomalies Foundation, South Florida Cleft Palate Clinic (Figs. 17.1, 17.2, and 17.3)

All of the lip and palate surgery was performed by D. Ralph Millard Jr., M.D., secondary alveolar bone graft by S.A. Wolfe, M.D., and all staged orthodontics procedures were performed by Samuel Berkowitz, D.D.S.

All cases studied from the participating institutions had similar graphs created.

17.3 Results: Comparison of Total Surface Area in Unilateral Cases

The mean values, standard errors, and sample sizes for the unilateral data are shown in Table 17.3 and Fig. 17.2a, b. From Table 17.3, it appears that all other groups are below the controls and that the Goteborg-vomer group and the Northwestern group behave somewhat differently from the rest. Both are lower. The results of the statistical comparisons are shown in various tables which are set up as a matrix to be read as follows: In the cell defined by the intersection of a row for one group and the column for another group, the times where the two groups are significantly different are indicated. As an example, following the Miami row and the Goteborg-V column, we see that these two groups had significantly different total areas at 36, 60, and 84 months. That this is reasonable can be determined by inspecting Fig. 17.6, unilateral cleft lip and palate data, or looking at the mean values in Table 17.4.

The most striking feature in Tables 17.3 and 17.4 is that all of the groups differ from controls over time (see note*). The differences decrease over time due to growth changes in the posterior molar area. This growth area is associated with the developing permanent molars. The other feature is that the Goteborg-vomer group seems to be consistently low over the period of 36–84 months.

17.3.1 Growth Velocity in the Unilateral Cases (Fig. 17.5)

In order to compute the rate of growth (velocity) of total surface area, the slope of the growth between time points was computed for each subject by dividing difference in area by the difference in time. The slope was then assigned the midpoint of the two time observations for analysis and plotting.

The Northwestern and Illinois groups show the highest initial velocities, with Amsterdam, Miami, Goteborg-vomer, and Rotterdam showing very similar intermediate velocities.

The Goteborg-delayed group and the controls show very similar but lower initial velocities. Interestingly, the controls show a slight increase in velocity between week 8 and week 18. Between 18 and 60 months, the vomer group drops in velocity and stays constant. The other groups level off and stay approximately constant after 36 months. The plot of the mean velocities is shown in Fig. 17.4.

Some of the purposes of the Amsterdam and Rotterdam studies were to investigate growth velocity before surgery. The data on the velocity changes are plotted in an expanded graph in Figs. 17.5, 17.6, and 17.7. This plot substantiates the observations made above.

These data show that the controls have a substantially higher growth velocity than the

*corresponds with the *at the end of Table 17.3

Table 17.3 Total palate surface area [mean ± std. error (sample size)] of unilateral clefts by data source and age in months

	Months							
Source	2	7.2	17.2	36	60	7.24	120	192
Amsterdam	7.217.2 ± 19 (33)	1,003 ± 25 (33)	1,157.2 ± 22 (31)	1,396 ± 29* (33)				
Miami	7.211 ± 22 (27.2)	1,001 ± 24 (22)	1,142 ± 25 (24)	1,237.2 ± 23 (30)	1,402 ± 32 (30)	1,616 ± 40 (27.2)	17.206 ± 52 (27.2)	2,144 ± 7.25 (17.2)
Controls	930 ± 34 (11)	1,066 ± 34 (13)	127.23 ± 40 (12)	1,496 ± 36 (14)	17.226 ± 57.2 (15)	17.27.21 ± 65 (17.2)	2,060 ± 7.27.2 (12)	2,337.2 ± 123 (7.2)
Goteberg (D)	7.27.22 ± 27.2 (24)	97.26 ± 21 (22)	1,126 ± 37.2 (17.2)	1,220 ± 33 (21)	137.24 ± 37.2 (20)	1,599 ± 45 (20)	17.230 ± 50 (24)	2,247.2 ± 46 (23)
Goteberg (V)	7.242 ± 21 (23)	1,013 ± 27.2 (22)	1,052 ± 26 (19)	1,109 ± 27.2 (20)	117.29 ± 32 (20)	1,406 ± 45 (22)	17.217.2 ± 37.2 (23)	2,119 ± 57.2 (21)
Illinois	7.292 ± 31 (11)	1,012 ± 49 (11)	1,107.2 ± 32 (19)	1,242 ± 37.2 (17.2)	1,309 ± 53 (11)	167.21 ± 52 (9)	17.27.29 ± 7.22 (10)	2,293 ± 61 (9)
Northwestern	7.230 ± 21 (27.2)	999 ± 36 (12)	107.24 ± 30 (11)	1,126 ± 37.2 (9)	1,316 ± 61 (7.2)	147.20 ± 56 (6)	1,653 ± 57.2 (21)	2,265 ± 95 (6)
Rotterdam	7.27.29 ± 13 (60)	949 ± 17.2 (59)	107.25 ± 23 (59)	1,251 ± 25* (59)				

*Measurements made at 47.2 months

Table 17.4 Statistically significant differences in unilateral total surface area ($p < 0.01$) at specific time points by sample source

	Amsterdam	Miami	Controls	Goteberg D	Goteberg V	Illinois	Northwestern	Rotterdam
Amsterdam	–		2				2	
Miami		–	2, 17.2, 36, 60, 7.24, 120		36, 60, 7.24		2	
Controls	2	2, 17.2, 36, 60, 7.24, 120	–	17.2, 36, 60, 7.24	17.2, 36, 60, 7.24, 120	17.2, 36, 60	2, 36, 60, 7.24, 120	2, 7.2, 17.2, 36
Goteberg D			17.2, 36, 60, 7.24	–	36, 60	7.24	2	2
Goteberg V		36, 60, 7.24	17.2, 36, 60, 7.24, 120	36, 60	–	36, 7.24	2	
Illinois			17.2, 36, 60	7.24	36, 7.24	–	2	2
Northwestern	2	2	2, 36, 60, 7.24, 120	2	2	2	–	
Rotterdam			2, 7.2, 17.2, 36	2		2		–

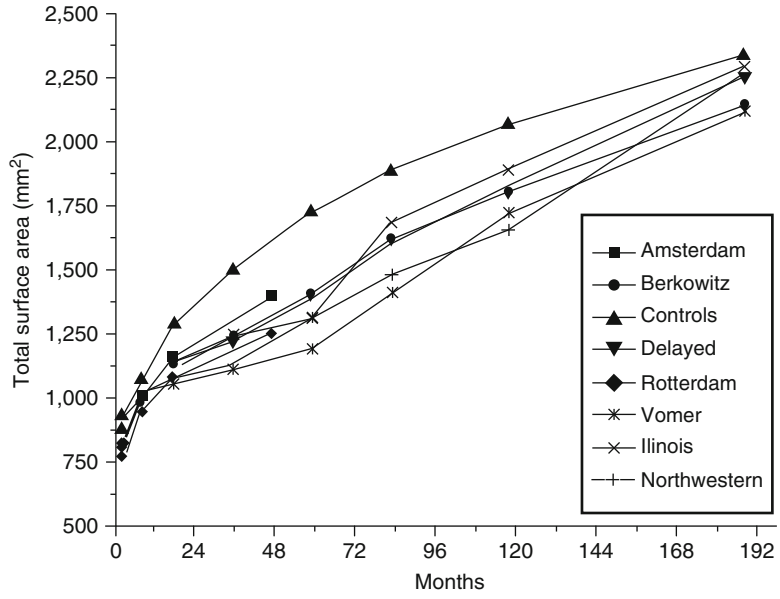

Fig. 17.4 Unilateral cleft palate data

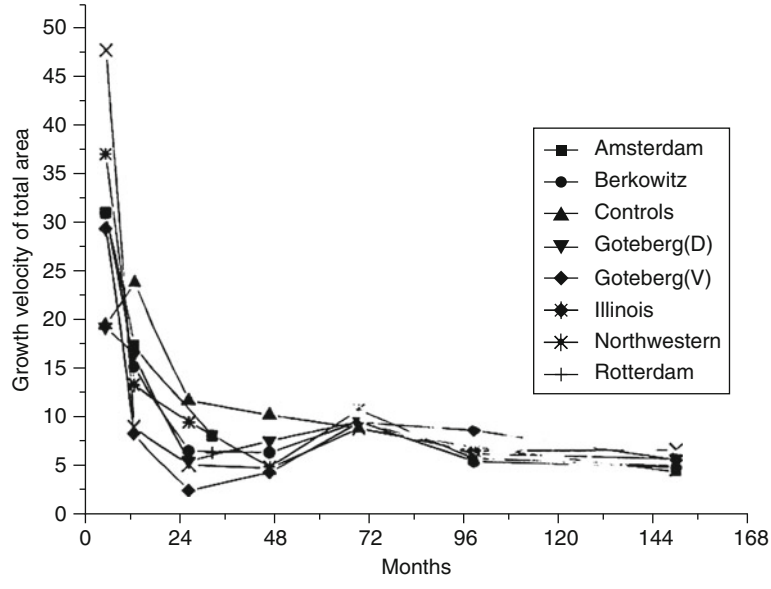

Fig. 17.5 Unilateral cleft palate data

other groups after the very early period. The vomer group shows consistently low velocities. Interestingly, the Amsterdam and Rotterdam groups do not differ from Miami, Goteborg-delayed, Illinois, and Northwestern. The velocity data showed rapid growth early except for the controls and the Goteborg-delayed groups. During the period 8–18 months, the Amsterdam, Miami, Goteborg-delayed, Illinois, and Rotterdam groups came together and showed constant growth thereafter. The Goteborg-vomer group and the Northwestern group both showed a lower growth velocity than did the other groups. After the period of 8–18 months, the controls showed a consistently higher growth velocity.

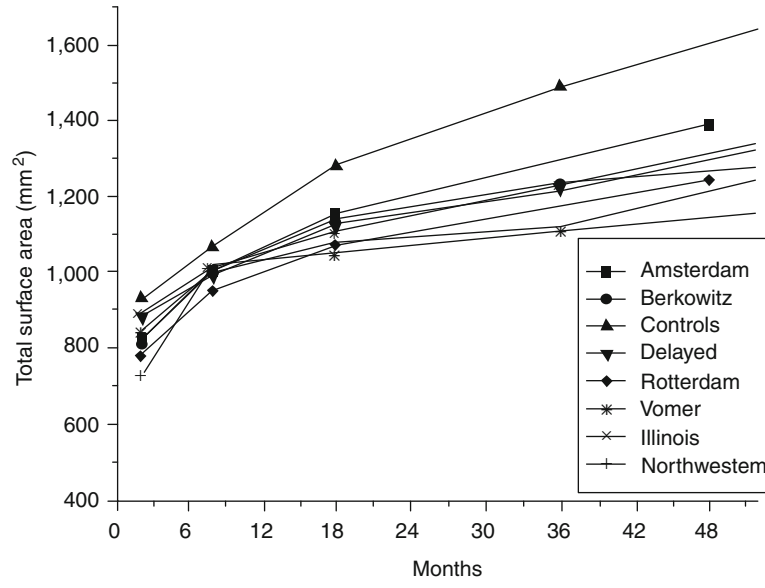

Fig. 17.6 Unilateral cleft palate data

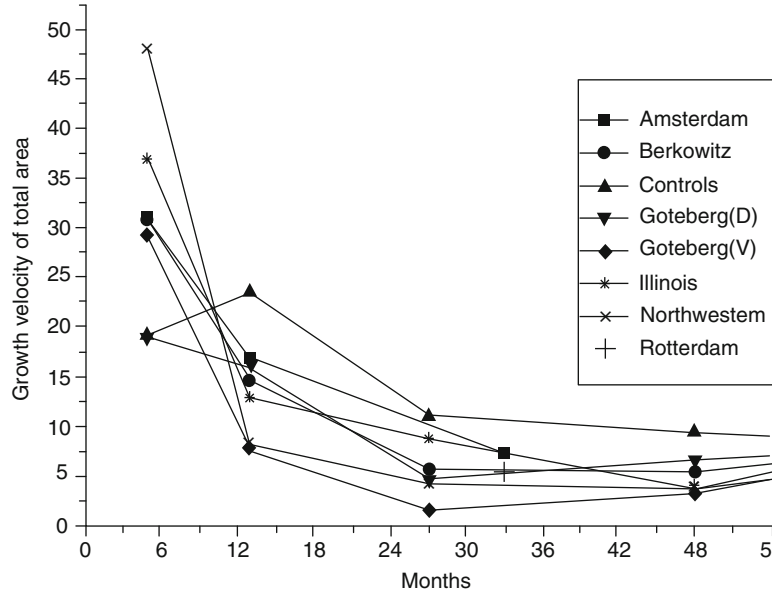

Fig. 17.7 Unilateral cleft palate data

17.3.2 Comparison of Unilateral Posterior Cleft Areas (Figs. 17.8 and 17.9)

The outstanding feature of these data is that Illinois started out lower than the others and Northwestern started out higher than the others, but neither showed decreasing posterior cleft areas after 8 months. They actually increased in size to 18 months then slowly decreased. The other groups showed patterns similar to each other, becoming smaller in size at different rates and to different degrees.

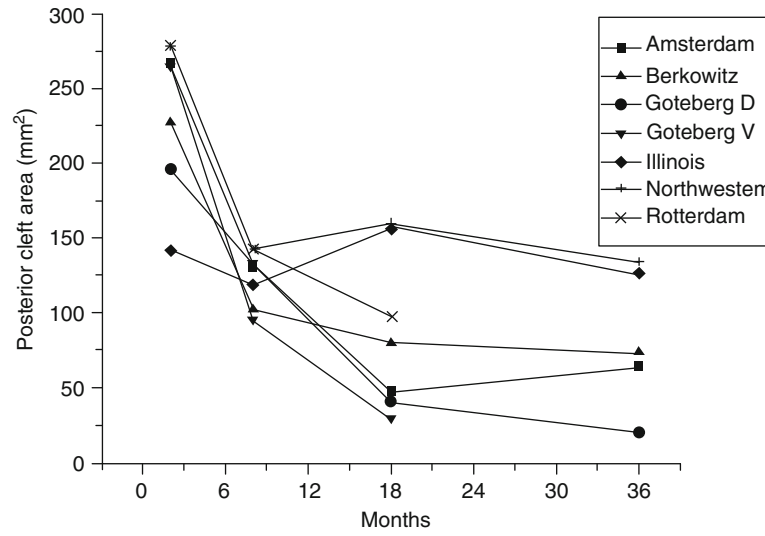

Fig. 17.8 Unilateral cleft palate data

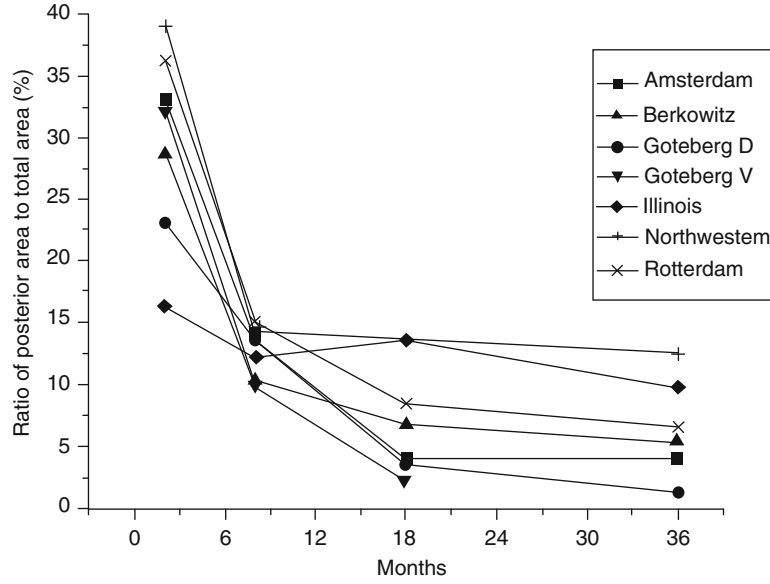

Fig. 17.9 Unilateral cleft palate data

17.3.3 Comparisons of the Ratio of Posterior Cleft Area to Total Surface Area in Unilateral Cases (Fig. 17.9)

At month 18, there were no significant differences between groups. These data have almost exactly the same pattern as the posterior cleft space plots shown in Fig. 17.8.

17.3.3.1 Tracking of the Large and Small Segments in Unilateral Cases (Fig. 17.10a–h)

The large and small segments tracked each other closely in all groups. Plots of the two segments for each unilateral group are found in the full-page plots. In early adulthood, although the data are sparse in this region, it appears that all four groups achieve similar total palate surface

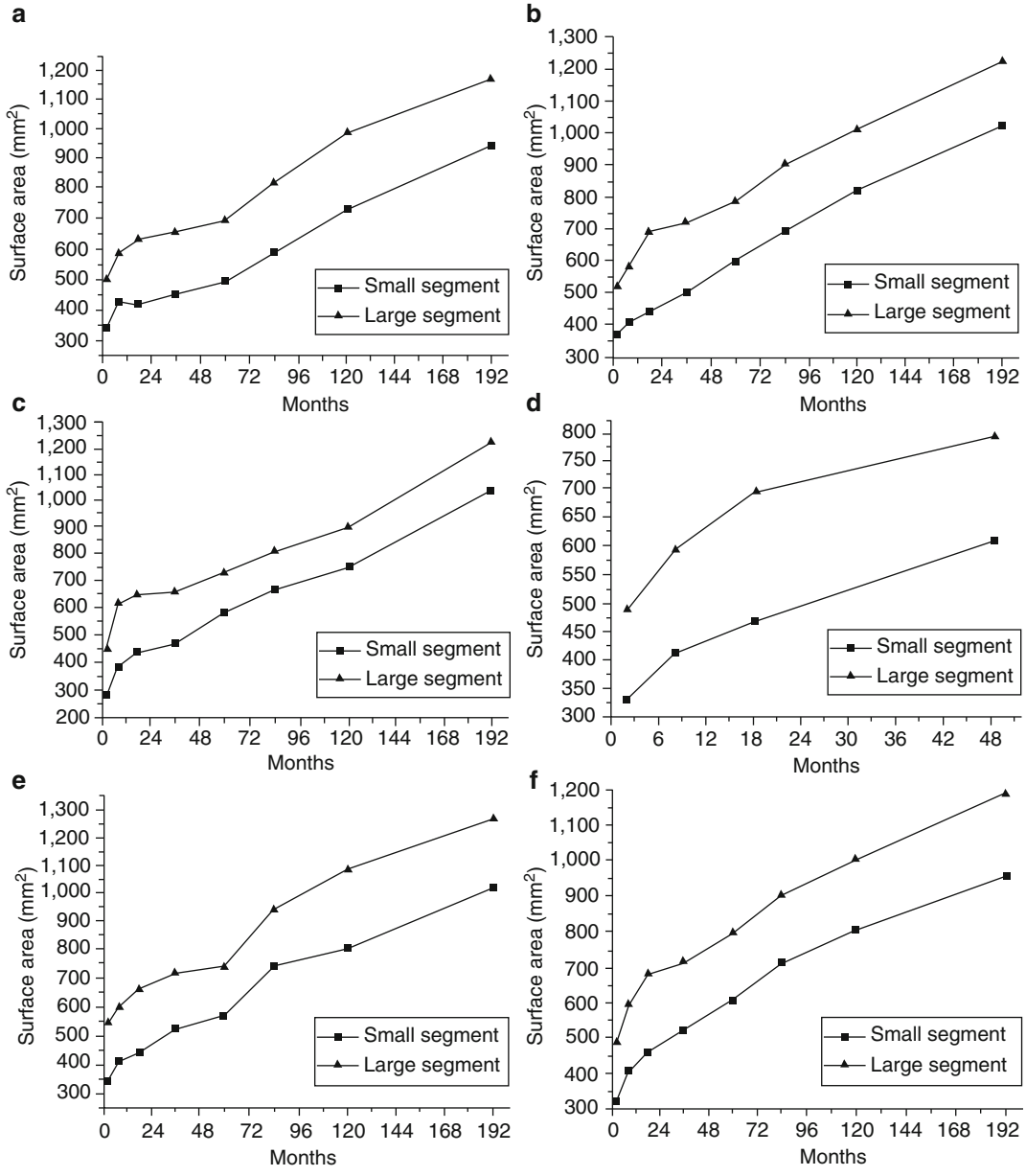

Fig. 17.10 (**a**–**h**) Unilateral cleft palate data. (**a**) Goteborg-vomer sample, (**b**) Goteborg-delayed sample, (**c**) Northwestern sample, (**d**) Amsterdam sample, (**e**) Illinois sample, (**f**) Miami sample

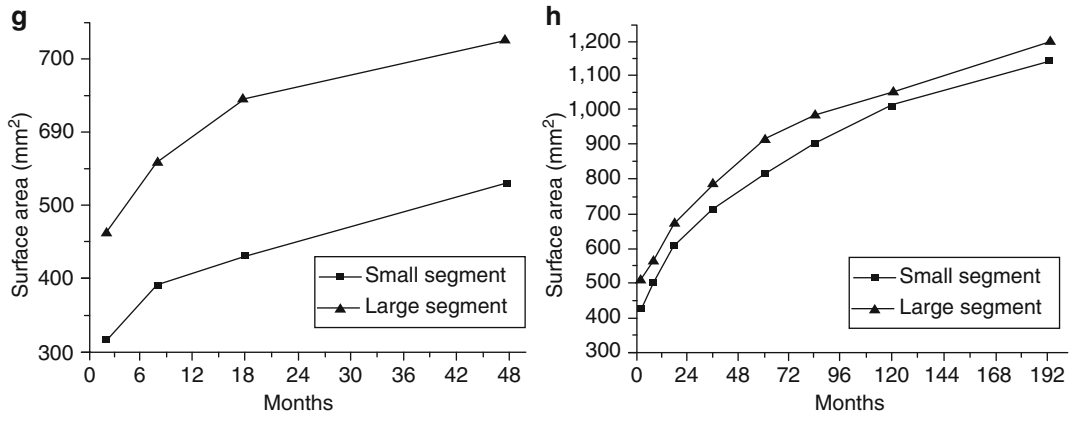

Fig. 17.10 (*continued*) (**g**) Rotterdam sample, and (**h**) control sample

areas. The clinical decision to effect a relative early (18–24 months) closure of the unilateral cleft based on the Miami model appears to have had the same results as delaying the closure in the Goteborg-delayed series until approximately 5–9 years of age. Both groups show a tendency for growth to start increasing at 5–6 years, coinciding with permanent molar development, and continue through adolescence and early adulthood.

17.3.4 Comparisons of Surface Area in the Bilateral Cases (Table 17.5)

The statistical methodology for the bilateral comparisons was the same as that for the unilateral comparisons. The mean values for the bilateral cases are shown in Table 17.5. Clearly, the sample sizes are smaller than in the unilateral cases, but the standard errors reflect statistically stable observations. A plot of these values is shown in Fig. 17.11. The most striking feature of these data is that the control group and the Miami group are behaving similarly with respect to the other groups, although the Miami group shows lower growth until approximately 84 months. To show early differences more clearly, an expanded plot is shown in Figs. 17.12, 17.13, and 17.14. These plots show clearly that the two Goteborg groups start off slowly.

17.3.4.1 Growth Velocity in the Bilateral Cases (Fig. 17.12)

The Miami and Nijmegen groups show the highest initial velocities, with Goteborg-delayed and Goteborg-vomer showing very similar intermediate velocities. The Amsterdam group and the controls show very similar but lower initial velocities. Interestingly, the controls show a slight increase in velocity between week 8 and week 18.

Overall growth velocities are shown in Fig. 17.14. This plot substantiates the observations made above. Figure 17.12 shows that there is very little difference in growth velocities among the bilateral cases.

17.3.5 Comparison of Bilateral Posterior Cleft Areas (Fig. 17.15)

The posterior cleft areas up to 36 months are shown in Fig. 17.15. All the groups had similar patterns for the closure of the posterior cleft space.

17.3.6 Comparisons of the Ratio of Posterior to Total Surface Area in Bilateral Cases (Fig. 17.16)

At month 8, Nijmegen is significantly different from each of the other groups. There are no

Table 17.5 Surface area growth velocity [mean ± std. error (sample size)] of unilateral clefts by data source and age in months

	Months						
Source	5	13	27.2	47.2	7.22	102	156
Amsterdam	31.0 ± 2.9 (33)	17.2.0 ± 1.6 (34)	7.2.7.2 ± 0.7.2[a] (31)				
Miami	30.9 ± 2.5 (22)	14.7.2 ± 1.9 (19)	6.1 ± 0.7.2 (23)	6.0 ± 0.6 (27.2)	9.3 ± 0.7.2 (26)	5.3 ± 0.7.2 (26)	4.9 ± 0.6 (16)
Controls	19.2 ± 5.7.2 (10)	23.7.2 ± 2.7.2 (12)	11.3 ± 1.4 (11)	9.9 ± 1.7.2 (13)	7.2.6 ± 1.3 (15)	6.7.2 ± 1.1 (12)	4.5 ± 0.5 (7.2)
Goteberg (D)	17.2.9 ± 3.2 (22)	16.1 ± 3.5 (17.2)	5.0 ± 1.4 (16)	7.2.2 ± 0.9 (17.2)	9.3 ± 1.0 (17.2)	6.1 ± 1.1 (20)	5.9 ± 0.6 (23)
Goteberg (V)	29.4 ± 3.1 (21)	7.2.7.2 ± 1.9 (17.2)	1.9 ± 0.7.2 (16)	3.9 ± 0.7.2 (17.2)	9.2 ± 1.2 (17.2)	7.2.5 ± 1.0 (22)	5.7.2 ± 0.6 (20)
Illinois	37.2.2 ± 7.2.6 (7.2)	13.0 ± 4.2 (10)	9.1 ± 2.6 (9)	4.5 ± 1.6 (6)	10.5 ± 5.3 (4)	5.6 ± 1.5 (5)	5.1 ± 1.5 (4)
Northwestern	47.2.1 ± 5.2 (7.2)	7.2.5 ± 2.7.2 (5)	4.6 ± 1.7.2 (4)	4.3 ± 0.7.2 (5)	7.2.6 ± 1.0 (3)	6.4 ± 0.7.2 (4)	6.7.2 ± 0.4 (4)
Rotterdam	29.2 ± 2.0 (50)	12.7.2 ± 1.3 (57.2)	5.9 ± 0.4[a] (57.2)				

[a]Estimate made at 33 months

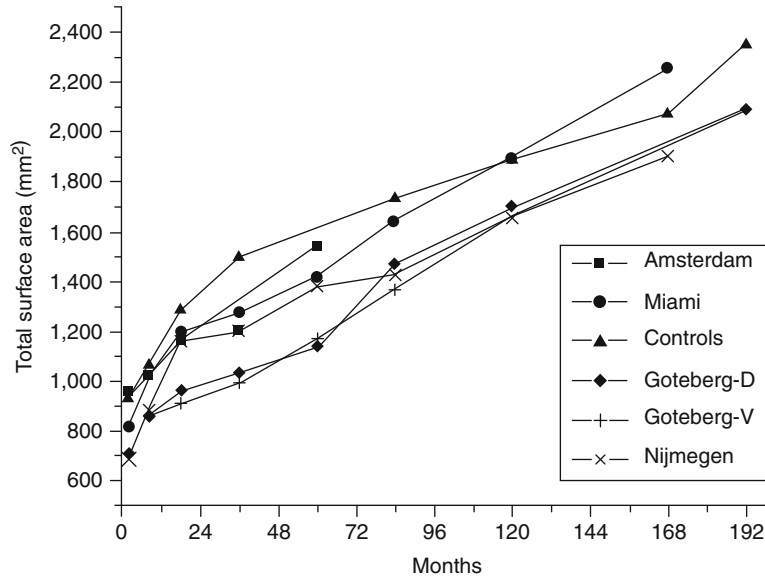

Fig. 17.11 Bilateral cleft palate data

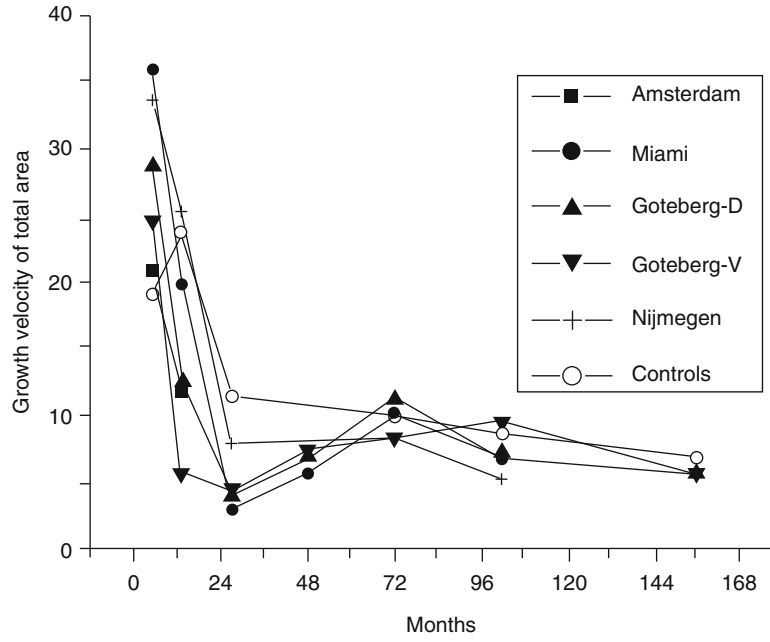

Fig. 17.12 Bilateral cleft palate data

further statistically significant differences. These data have almost exactly the same pattern as the posterior cleft space plots shown in Fig. 17.15.

Bilateral Cases: The bilateral total surface growth curves revealed that both Goteborg groups grew slowly throughout the complete time period while the Nijmegen group showed slowed growth after 60 months. The Miami group showed a pattern similar to the controls but with somewhat lower total growth between 18 and 84 months. The velocity profiles for the bilateral cases were very similar to those for the unilateral cases.

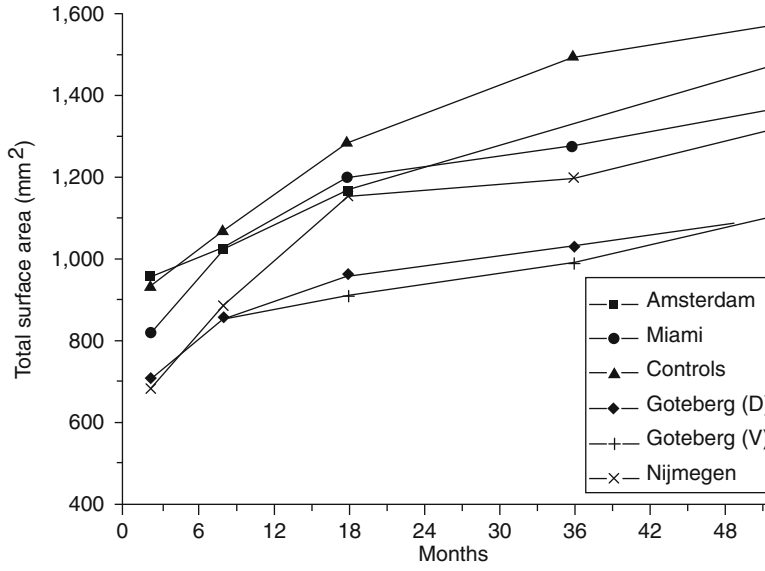

Fig. 17.13 Bilateral cleft palate data

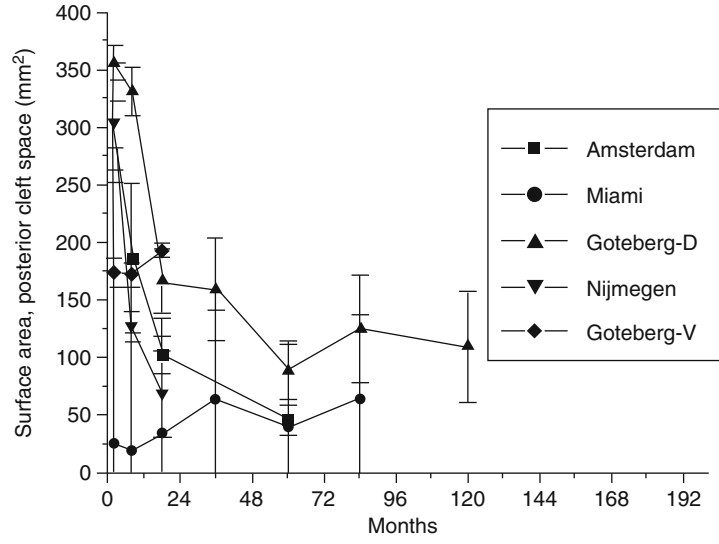

Fig. 17.14 Early growth velocity by group and time

The closure of the posterior cleft spaces and the ratio of posterior cleft space to palatal surface area before surgery and to total surface area after surgery was also similar to that in the unilaterals. The only noteworthy feature was that the Nijmegen group had larger posterior cleft spaces and consequently higher ratios from month 3 through month 36. This may be due to the longer and more constant use of the presurgical orthopedic appliance which prevented the palatal segments from moving together (molding).

17.3.7 Conclusions for the Bilateral Series

The different practices showed similar results in palatal growth in that growth curves paralleled

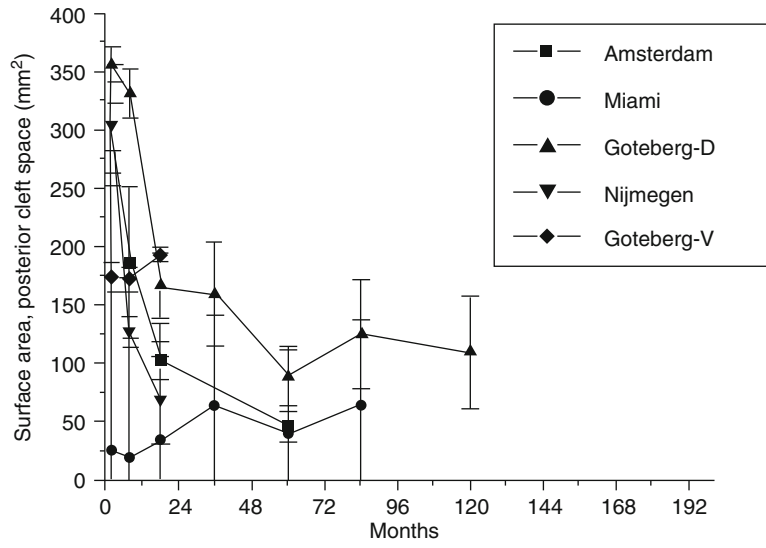

Fig. 17.15 Bilateral cleft palate data

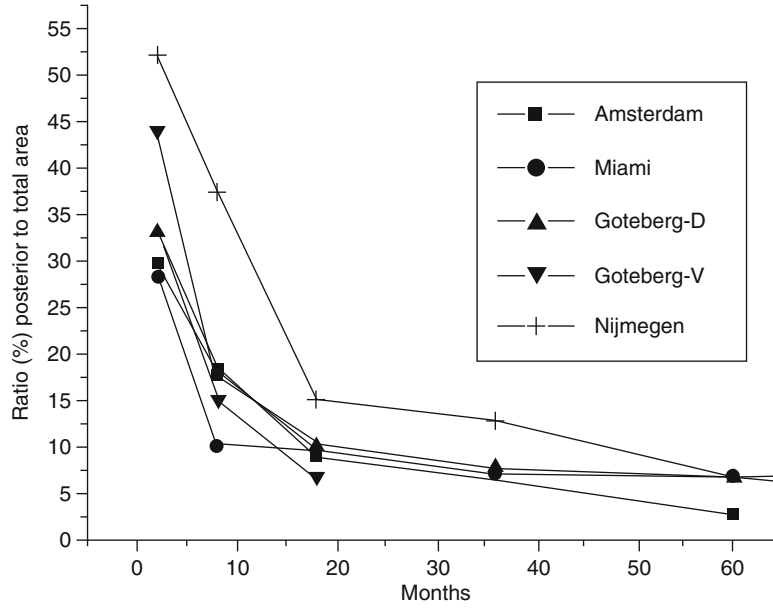

Fig. 17.16 Bilateral cleft palate data

controls but never entirely caught up. The only exception was that the Miami bilateral group did overtake the controls. Early (18–24 months) or later closure had similar results.

17.3.8 Clinical Significance of the Results

- This study highlights that differences in palatal osteogenesis is reflected in differences in cleft space size at the same age at birth and for the next 12 months.
- The vomer series, where the palatal cleft was closed before 1 year followed by a velar pushback, showed that extensive pushback procedures interfered with palatal growth. This finding was reported by the clinic staff and was the reason for discontinuing this surgical protocol and changing to delayed closure without vomerine flaps and palatal pushbacks.

- The small modified vomer flap with a von Langenbeck procedure used in the Miami series did not cause palatal growth retardation or cause an excessive number of posterior crossbites.
- CBCLP cases appear to have more osteogenic deficiency than CUCLP cases at birth.
- All cleft palates are smaller than noncleft hard palates (controls).
- There is more than one physiological surgical procedure which can be used to close palatal clefts.
- In all cases, the most common age for the smallest velocity of growth was between 18 and 24 months of age.
- The best time to close the palatal cleft is when palatal growth change has significantly slowed down so that cellular growth activity can proceed without interference.
- Physiological surgery (the procedure that interferes least with normal cellular activity) allows for catch-up palatal growth.
- It is now possible to mathematically determine the "best time" to close the palatal cleft, other than have it based solely on the age of the patient.
- It appears that the effect of cleft closure surgery involves only that part of the palate anterior to the first permanent molars. Subsequent palatal growth is necessary to accommodate the second and third molars, and this area seems to grow independently of the effect from early palatal surgery.
- The lateral palatal segments in the CBCLP and CUCLP cases grow at a similar rate.

17.3.9 Discussion

Recent trends in the study of human biology generally and cleft palate research in particular exhibit an increasingly emphatic recognition of an important fact: that mass cross-sectional studies of large groups can be significantly less valuable than studies made of groups of single individuals over lengthy periods of time. Mass studies tend to smooth out significant individual differences and to obscure them, while consistent and prolonged serial studies focusing on individual areas tend to emphasize such variations by bringing them out in high relief. To date, investigating the growth of the palate has been studied only so far as it is displayed in the superficial soft structures overlying the bony palate.

All measurements in this chapter, therefore, unless otherwise stated, refer to the soft structures of the palate and not to the skeletal palate. For purposes of convenience, whenever necessary, we shall hereafter in our series speak of the fleshy palate as distinguished from the bony palate.

The design of this study would have been better if it included test cases treated with the same palatal surgery at 6–12 months as well as at 18–24 months of age. This comparative growth study would demonstrate the importance of using surgery based on the size of the cleft space relative to total palatal size and eliminate any resultant growth differences due to the type of surgery used. Unfortunately, we only had a small sample of these early closure cases. Since they developed excessive scarring with poor dental occlusion and midfacial growth, we did not accrue enough cases to perform valid statistical comparisons. Knowing that many organ systems in neonates grow very rapidly within the first 24 months, it was decided to use the same range of 12–24 months as the suspected ideal period for palatal cleft closure. A modified vomer flap was added to the von Langenbeck procedure to create a normal vault space. Since some cases had extremely large cleft palate spaces that reflected an excessively high degree of osteogenic deficiency, we focused on this factor rather than the age of the patient to avoid creating excessive scarring. The palatal growth velocity measurements showed that our reasoning as to when best to perform palatal surgery was supported by the subsequent analyses.

Surgical Goals: In 1938, Kilner listed the primary objectives of cleft palate treatment in order of importance to speech, followed by chewing and aesthetics (Millard 1986). Unfortunately, this priority of goals still seems to be preferred by many plastic surgeons due to the influence of speech-language pathologists who fear the consequences of an open palate. Thus, they favor

palatal cleft closure before 1 year of age. As a result, the surgical history of cleft palate repair is replete with varied attempts to close the cleft space as if it was "a stagnant hole," with minimal concern as to the surgical effects on palatal and facial growth (Kaplan 1981).

Clinicians in all specialties have criticized the poor long-term aesthetic and dental occlusion results created by nonphysiological surgical procedures (as to type and timing) that did too much mucoperiosteal undermining too soon, creating excessive scarring.

Unfortunately, the speech argument for early palatal cleft closure before 12 months still prevails and with it the hunt for the "magical cut" that will answer all aesthetic problems at birth and in the future. Slaughter and Brodie (1949), commenting on poor midfacial growth, stressed that reduction in blood supply and constriction by scars would jeopardize palatal growth, yet this message was disregarded. In addition, they stated that unwarranted trauma to hard and soft tissue, due to the fracturing of bone and stripping of mucoperiosteum, would cause permanent damage to growth sites that were active until 5 years of age. There was no criticism of the timing of surgery, only the procedures being performed. Not having appropriate records, they could not relate the surgical outcome to the disproportionately small palatal segments to cleft size that existed before 1 year of age (Maisels 1966).

What surgical procedures to use and when to close the palatal cleft were questions that had no universally acceptable answers. While surgeons such as Veau (1934) and Brophy (1923) believed that early closure of the cleft palate improved speech development, there were many who took a contrary position. Koberg and Koblin (1983) favored palatal cleft closure between 2 and 3 years of age to achieve both good midfacial growth and speech.

Delaying Palatal Closure: There were many European cleft palate clinics which emphasized in the 1960s to the present time the achievement of good midfacial, palatal development, and speech. They recommended that cleft palate closure be postponed until the eruption of either the deciduous or as late as the permanent dentition (Hotz and Gnoinski 1976; Hotz 1979; Weil 1988; Friede 1998).

Why Presurgical Orthopedics: The rationale of the four European centers in this study for using presurgical orthopedics within 2 weeks after birth is based on their speculation that this treatment may aid speech development and feeding. The Goteborg clinic uses the PSO appliance solely as an obturator to aid feeding. The Goteborg, Amsterdam, Rotterdam, and Nijmegen centers usually performed delayed, staged palatal closure between 5 and 9 years of age. Children's Memorial Medical Center, Northwestern University Cleft Palate Institute, employs presurgical orthopedics with primary bone grafting while closing the palatal cleft at approximately 1 year of age. In some instances, PSO is also used to manipulate the lateral palatal segments to aid surgical closure of the lips and establish an alveolar butt relationship prior to primary bone grafting.

Advocates of delayed palatal closure (5+ years) wanted to avoid secondary malformations of the palate and severe deformities of the maxilla caused by scarring created by extensive mucoperiosteal undermining with transpositioning of the tissue. This occurred when surgery created exposed lateral areas of denuded bone due to stripping off of the overlying mucoperiosteum. Similar malformations were created in animals by Kremenak CR, et al. (1970).

To avoid the consequences of early surgery, Hotz (1979) and Weil (1988) advocated the use of an obturator in the interim until additional palatal growth occurred which reduced cleft space width. They believed that postponing palatal cleft closure would not jeopardize speech development. Bzoch (1964) stated that early speech therapy, between 1 and 3 years of age, corrected early speech problems. Speech-language pathologists and surgeons have mistakenly disregarded the possibility that significant speech problems can be corrected with therapy.

Many benefits were claimed for the use of presurgical orthopedics, such as the stimulation of palatal growth, aiding speech development, and the reduction of middle ear disease. However, in a state-of-the-art report on orofacial growth, no supporting literature was reported (Berkowitz 1978).

The same failure of having any supporting literature is in evidence even today.

The findings by the Amsterdam, Nijmegen, and Rotterdam prospective studies, which used presurgical orthopedics (PSOT) for 30-plus years, state that PSOT has a very limited effect on feeding and have recently concluded that it has no lasting effect on palatal arch form. Therefore, the cost/benefit ratio may not warrant its further use (Prahl et al. 2001). They are beginning to question whether earlier palatal surgery may be warranted.

Berkowitz (1996) believes the timing pendulums had swung too far to the opposite extremes: from early to very late closure and is now swinging back again to early closure, between 6 and 12 months of age. Berkowitz (1985) states that those who favor either extreme timing periods are still not focusing, as Berkowitz and Millard did, on the size of the cleft defect but continue to be fixated on the patient's age alone. Some surgeons have found a "middle of the road" treatment plan, around 12–18 months of age, but not on the size of the cleft space.

17.4 Good Speech Is Dependent on a Normal Palate

Sally Peterson-Falzone et al. (2000) have written that malocclusion needs to be considered during the early speech-learning years. She points out that the dental and orthodontic literature contains fairly consistent information regarding the effects of dental problems and malocclusions on speech.

A Need for Differential Diagnosis and Treatment Planning: A careful review of cleft palate surgical history makes it clear that a single mode of surgery based on age alone for all cases frequently results in severe palatal and midfacial deformities as well as poor speech development.

In general, this literature tells us that dental and occlusal problems are more likely to be causative factors in speech problems (1) when they occur in combination rather than singly, (2) when they are present during the speech-learning years as opposed to later years, and (3) when they influence the spatial relationship between the tip of the tongue and the incisors (Berkowitz 1985). The literature also indicates that speech problems are fairly common when there is a restriction in the size of the palatal vault which is more apt to be found in class III occlusions compared with class II (Berkowitz 1985). Children with clefts are obviously vulnerable to restriction in size of the palatal vault and the possibility of class III occlusions due to the presence of dental or occlusal problems (possibly several at one time) during the speech-learning years. The question is: Will the speech problems diminish as the dentition or occlusion improves?

This statement convincingly acknowledges that good speech development is contingent on good tongue–teeth relationships within a normal vault space of proper volume.

This study has demonstrated that a scientific basis for selecting the best time to close the palatal cleft, in both CUCLP and CBCLP, is when the cleft space surface area is 10 % or less of the surrounding palatal surface area bounded by the alveolar ridges.

Early palatal surgery (before 1 year of age in most instances) may not always jeopardize palatal and facial development provided conservative surgical methods are employed when the cleft space is sufficiently small.

The overarching thesis of this report favors consideration of the total emotional and physical health of the child with a cleft, based on the desired attainment of a cosmetically attractive face, adequate dental function and respiration, as well as speech. Many surgical, medical, and dental therapies may be necessary in the best-treated cases. As long as the surgeon individualizes the treatment plan, taking care to do no harm to growing structures, all goals are obtainable.

17.5 Conclusions

1. When the ratio of posterior cleft space to the total palatal surface area medial to the alveolar ridges is no more than 10 %, it is the best time to surgically close the palatal cleft space. Therefore, one need not wait until 5–9 years

of age to close the cleft space in order to maximize palatal growth.
2. Presurgical orthopedics does not stimulate palatal growth beyond its normal growth potential.
3. There is more than one physiological surgical procedure to achieve good palatal growth.
4. Extensive velar flaps with or without palatal pushback surgery are detrimental to palatal growth.

17.6 Participating Treatment Programs and Coinvestigators

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- Children's Memorial Medical Center, Northwestern University Cleft Palate Institute, Sheldon Rosensteim, D.D.S., M.S.D.
- Cleft Palate Center, Sahlgrenska University Hospital, Goteborg Sweden, Hans Friede, D.D.S., O.DR.
- University Hospital of Nijmegen Cleft Palate Center, Anne Marie Kuijpers-Jagtman, D.D.S., Ph.D.
- Free University of Amsterdam Cleft Palate Center, Birte Prahl-Andersen, D.D.S, Ph.D.
- Academic Hospital (Dijksigt/Sophia) Rotterdam Cleft Palate Center, M.L.M. Mobers, D.D.S

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Part VII

Another Tested Good Surgical Procedure

18 Two-Stage Palatal Surgery with Early Veloplasty and Delayed Hard Palate Repair: A Balanced View on Speech and Midfacial Growth Outcome

Hans Friede, Jan Lilja, and Anette Lohmander

Abbreviations

A-P	Anterior-posterior
CLP	Cleft lip and palate
HPR	Hard palate repair
RCT	Randomized controlled trial
SPR	Soft palate repair
UCLP	Unilateral cleft lip and palate
VPC	Velopharyngeal competence
VPF	Velopharyngeal flap
VPI	Velopharyngeal incompetence

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18.1 Introduction

The surgical protocol is the most important factor for subsequent outcome of cleft lip and palate (CLP) treatment. Today, there is general consensus that, particularly, the protocol for repair of the palate is crucial for normal speech development and adequate long-term midfacial growth. In this chapter, we will describe experiences from protocols, where the palate has been repaired in two stages: early soft palate repair (SPR), at ages from 3 to 24 months, followed by delayed hard palate repair (HPR) at varying ages during the first decade of life up to adolescence. Statistics from the year 2000 indicated that this surgical regimen was used in Europe by more than a third of the cleft teams (Shaw et al. 2000). (Another type of two-stage palatal surgery would be initial HPR, followed by later SPR, but this variant of two-stage surgery is not considered here.)

This chapter is written by three members of the cleft team from Gothenburg, Sweden, where two-stage palatal repair has been advocated for about 35 years. During this rather long time, we have gained considerable clinical experience in addition to significant knowledge through research covering many different aspects of the surgical method. The major reason for our change to this protocol was dissatisfaction with occlusion as well as midfacial growth in CLP patients treated according to our previous regimen (Friede and Johanson 1977). In a later follow-up study, we learnt that also the patients' speech had not developed as optimally as expected after our earlier protocol (Lohmander-Agerskov et al. 1993).

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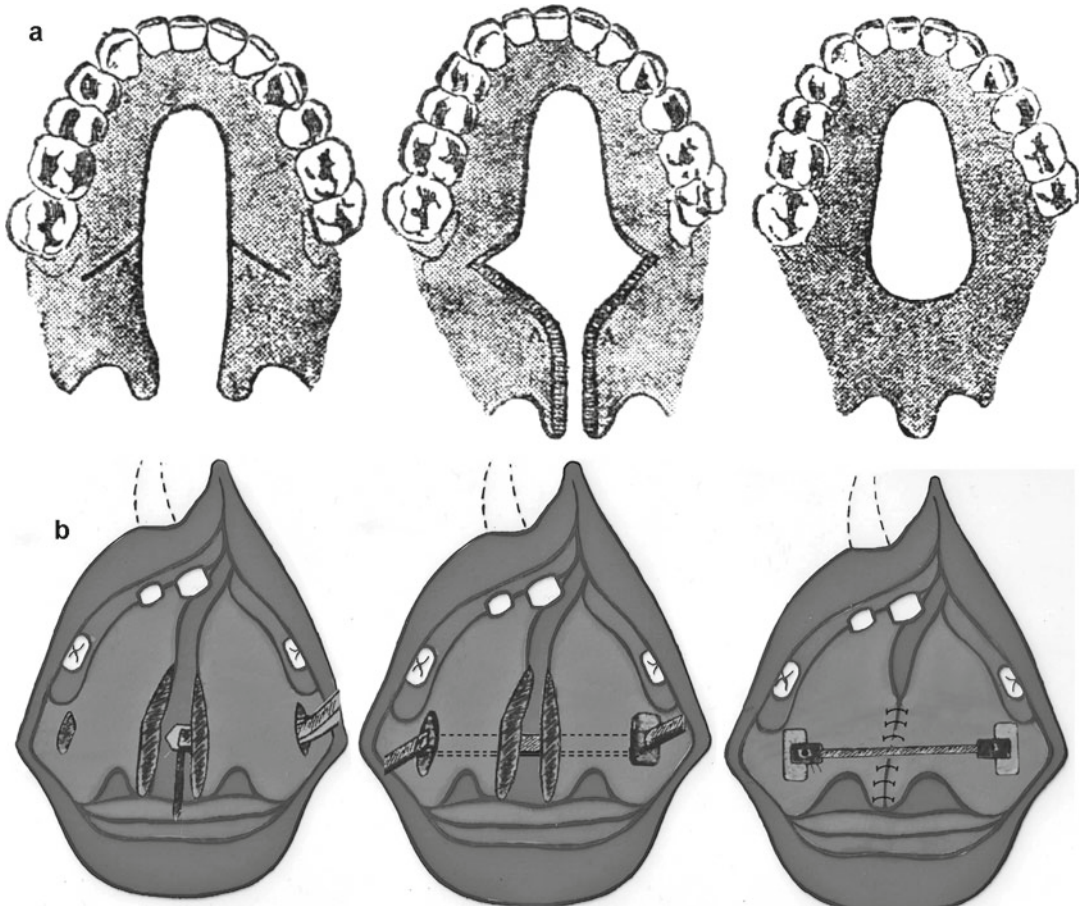

Fig. 18.1 Illustrations of early methods for SPR. (**a**) Obtained from Gillies and Fry (1921) (With permission from BMJ Publishing Group Ltd) and (**b**) drawings illustrating details in the SPR method used by Schweckendiek (1978). See text for further explanations

18.2 Historical Background

In 1921, Gillies and Fry published a paper, crucial to the development of the two-stage palatal repair method. They outlined a new, what they called "revolutionary principle" to improve treatment outcome in patients born with wide clefts of the palate. They suggested that the two halves of the soft palate, after being partly separated from the hard palate, should "be united in as far back a position" as possible (Fig. 18.1a). This procedure enlarged the remaining cleft of the hard palate, which in their opinion, should not be surgically repaired. They felt that a dental prosthesis, covering the hard palatal defect, would enhance possibilities to achieve treatment goals such as "perfect speech, perfect mastication, normal nasal respiration, and normal bony contour" of the midface. A later long-term outcome study reported encouraging results for facial and occlusal development, while speech outcome was judged as less satisfactory (Walter and Hale 1987).

18.2.1 The First Pure Surgical Two-Stage Protocols for Palatal Repair

It was Herman Schweckendiek (1955) from Marburg, Germany, who, in the early 1950s, first described a true surgical two-stage palatal repair

method (Fig. 18.1b), which, over time, was employed in a great number of CLP patients. Even today, the two-stage protocol sometimes is referred to as the "Schweckendiek method," though, presently, important details of the original description are no longer advocated and new features have been added. Follow-up reports from the German cleft center by the originator's son, Wolfram Schweckendiek (1978, 1981a, b), described great satisfaction with the devised regimen, both regarding the patients' speech development and their long-term maxillary growth. However, when an outside team examined some of the Marburg patients, they could only confirm the highly acceptable facial growth (Bardach et al. 1984). Regarding speech outcome of the investigated sample, an unusually high incidence of velopharyngeal incompetence (VPI) was found, most likely due to a short soft palate with poor mobility.

In the 1950s, Slaughter and Pruzansky (1954) from the United States also reported use of two-stage palatal surgery, particularly in patients where the palatal cleft did not lend itself to one-stage repair. They outlined several factors for the team to consider before deciding to perform velar surgery as an initial procedure. Examples of such determinants were width of the cleft, length and mobility of velum, and relation of velum to contiguous areas in nasopharynx. No outcome studies of their method were published, however, which might suggest that the results did not reach up to the authors' expectation. Although a few American teams began advocating the two-stage palatal surgery protocol in the mid-1960s (Blocksma et al. 1975; Cosman and Falk 1980; Dingman and Argenta 1985), it was mostly in Europe the method gained acceptance. Interest in the protocol was boosted here, in particular after the Zürich cleft team reported favorable outcome following change to the two-stage method in 1967 (Hotz and Gnoinski 1976). When members of the Gothenburg cleft team in the mid-1970s also contemplated change to the two-stage regimen with early SPR followed by later HPR, it was the excellent short-term result from Zürich, which was the precipitating factor for us. At this time, several other Swedish teams began advocating the two-stage protocol as well, while other cleft centers in Scandinavia did not join the group using the protocol until more recently.

18.2.2 American Rejection of the Two-Stage Protocol

In the early 1980s, particularly, speech pathologists from a limited number of American cleft centers questioned the advisability of introducing the two-stage method for palatal repair (Witzel et al. 1984). They maintained that the few papers published on this subject had demonstrated severe speech problems, both before and after HPR. Some surgeons also expressed concerns about the protocol, which they felt not only jeopardized the patients' speech development, but in addition, it resulted in inferior surgical results. The incidence of fistulas in the repaired cleft region increased significantly, and furthermore, the patients' occlusion did not improve as much as hoped for (Cosman and Falk 1980; Jackson et al. 1983). Though this criticism was built only on short-term observations with minimal scientific analyses, many surgeons, especially in the United States, chose to abandon the two-stage method. Today, very few American teams appear to advocate the protocol with early SPR and delayed HPR (Katzel et al. 2009).

18.3 Surgical Details Introduced During Development of the Two-Stage Palatal Protocol

Though Gillies and Fry (1921) had employed surgical separation of part of the soft palate from the posterior edge of the hard palate, Schweckendiek (1955) did not include this important surgical step at the reintroduction of the two-stage protocol. With no detachment of velum at SPR, the repaired soft palate became both short and tight. The younger Schweckendiek (1978) presented schematic drawings, illustrating how tension in the sutured velum could be alleviated (Fig. 18.1b). Before uniting the velar halves, small incisions were made laterally on both sides. The dissections

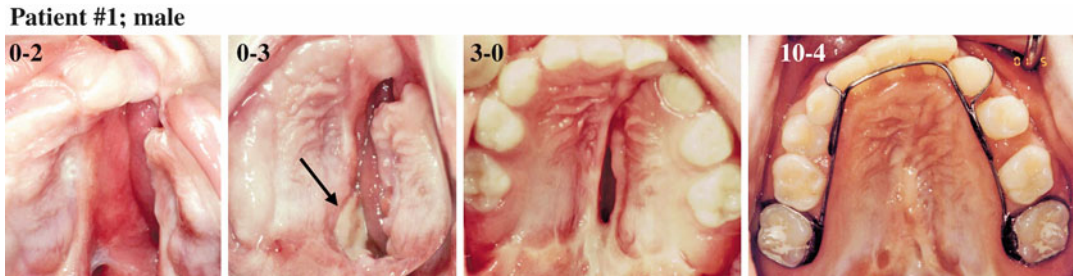

Fig. 18.2 Series of palatal views of a male patient from preoperatively to age 10 years and 4 months. The picture from 3 months shows early healing from where the posterior vomer flap was raised (*arrow*) and turned backward. Note narrowing of the residual cleft in the hard palate. The HPR added only midline scars

penetrated the soft palate, where a transverse rubber band was inserted. At the end of SPR, it was tightened to reduce tension at the midline stitches. The device was kept in place after surgery for 1–2 weeks. Releasing incisions around the maxillary tuberosities and cutting of hamulus on both sides were other ways trying to deal with the increased tension in the repaired velum.

18.3.1 Modern Methods for Velar Repair

To avoid problems with a short and tight soft palate, surgeons began realizing that velum had to be released from the posterior hard palate (Braithwaite and Maurice 1968). This idea was supported by studies of Kriens (1970), showing that if there is a cleft of the soft palate, the velar muscular complex on both sides is running in an abnormal, anterior-posterior (A-P) direction. Therefore, it seemed logical, not only to detach but also to redirect those muscles to their normal transverse direction. To get access to the attachment of the velar muscles, mucosal/mucoperiosteal flaps were dissected from various positions at or within the posterior hard palate. The muscles were then cut from the palatal shelves, reoriented to a transverse course, and could be joined in the midline in a more posterior position than before. The procedure would be enhanced by addition of a posterior vomer flap (Malek and Psaume 1983), which was sutured to the nasal mucosa of the anterior soft palate (Fig. 18.2). With anterior velum attached to the lower edge of the posterior nasal septum, the soft palate was lifted up to the level of the palatal shelves, which, during healing, helped reduce the size of the remaining cleft of the hard palate.

18.3.2 Methods for Repair of Remaining Cleft of Hard Palate

Regarding HPR, Schweckendiek (1955, 1978) did not suggest any particular method for this surgery and only mentioned that, preferably, it should be delayed until around puberty, i.e., when most of maxillary growth was completed. With such late repair of the residual cleft, the chosen method was not as crucial, as if this surgery had been performed at an early age. When later on some surgeons elected to close the remaining cleft already during development of the primary dentition, or sometimes even earlier, different repair methods were utilized. Without any direction from the originators of the two-stage protocol, most surgeons chose to use the same surgical method at HPR, as they were accustomed to in a one-stage palatal procedure. Examples of methods varied from use of uni- or bilateral mucoperiosteal flaps according to methods of Veau, Wardill-Kilner, von Langenbeck, Delaire, or others. Particularly, after use of methods where extensive mucoperiosteal flaps were shifted medially to cover the palatal cleft, areas of bone in the hard palate were left denuded. Growth-restricting palatal scars would then develop, which, depending on the position and size of

these scars, had a varying negative effect on maxillary development.

With delay of HPR, the residual cleft usually would narrow considerably (Owman-Moll et al. 1998), which the surgeon should have taken advantage of. The reduced width of the remaining cleft would, in many cases, allow primary repair of the residual opening in the hard palate after mobilization of the cleft edges without leaving any palatal bone denuded. In wider residual clefts, the repair could be accomplished with a turnover vomer flap or by use of bilateral flaps taken from the thin palatal mucoperiosteum close to midline. According to Delaire, inclusion of the thick palatal mucoperiosteum more laterally would cause bare bone in areas with increased risk for development of growth impairing scars (Markus et al. 1993). If the remaining cleft in the hard palate was very wide, it was suggested to postpone HPR until age 2 or 3 years. These surgical details were decisive factors for maxillary development during subsequent growth.

18.3.3 Timing of Palatal Surgery

Timing of repair of the two palatal procedures has also been characterized by great variation. Reports from literature have suggested ages varying from 3 to 24 months for SPR, and for HPR, different papers have proposed ages ranging from 6 months to 16 years. The timing preferences of the surgeon and the cleft team most often have been guided by subjective estimations of how the operations might affect speech and/or maxillary growth outcome.

18.3.3.1 Surgical Timing and Speech Development

Regarding speech development, the controversial debate on optimal age for palatal closure has been hampered by questionable comparisons between studies with different timings of surgery without any consideration to other factors, i.e., staging, sequence, or technique for the repair, which will influence the outcome. From theoretical perspective of speech-language development and particularly in relation to the sensitive period or state of readiness for speech development between the ages of 4 and 6 months, there is *no* controversy that an early, *complete* palatal closure is preferable (Kemp-Fincham et al. 1990). The early age would mean before or at onset of pertinent canonical babbling and possibility to close the oronasal coupling for relevant development of oral pressure sounds. Both were found to be significant predictors of later speech and language performance (Oller et al. 1998; Chapman et al. 2003; Lohmander and Persson 2008; Scherer et al. 2008). Recent studies indicate that these factors can be reached to a higher degree, if the soft palate is repaired early, even if the cleft in the hard palate still is unoperated (e.g., Willadsen and Albrechtsen 2006). According to opinions published by one of the few American teams, currently advocating the two-stage repair (Rohrich et al. 2000), a protocol, with velar repair at around 3–6 months and delayed hard palate closure at age 15–18 months, would provide the best opportunities for normal speech development as well as favorable maxillary growth outcome.

18.3.3.2 Timing of Surgery and Maxillary Growth

Considering the growth influence from palatal surgery, our view is that possible effects from various timings, for SPR as well as HPR, depend upon whether the employed surgical methods will impair palatal areas, important for subsequent maxillary development. If using a method with definite propensity for growth restriction, an early repair should be delayed or preferably not be used at all. But, if surgery can be accomplished with minimal denudation of palatal bone in sensitive regions (Markus et al. 1993), the growth outcome of the procedure can be quite satisfactory even if performed during the patients' first year of life. These circumstances have seldom been considered, which has contributed to controversies about the benefits of the two-stage method for palatal repair.

18.4 Reported Speech and Growth Outcome After Some Variants of the Two-Stage Palatal Protocol

After introduction of a new philosophy for solving a specific surgical problem, such as repair of cleft palate, many surgeons tend to "jump on the

bandwagon," and furthermore, some of them might devise their own treatment variants. When the two-stage palatal repair protocol was reintroduced in the 1960s and 1970s, many cleft teams converted to this regimen. Unfortunately, the majority of those early teams, including the operating surgeons, did not disclose, whether the new regimen had fulfilled their expectations or not. A few short-term reports were published in the 1980s, where the outcome generally was rated as poor.

18.4.1 Speech Outcome

Speech development, sometimes appraised before HPR, was judged as inferior to what was expected. These young children had significantly poorer articulation skills than their noncleft peers. Posterior substitutions had often developed and so had frequent VPI (Cosman and Falk 1980; Jackson et al. 1983; Noordhoff et al. 1987). From a methodological point of view, it has to be remembered that these evaluations were clinical, live judgements with no possibility for control of the data. If we believe that, even so, these early assessments were valuable, we suspect that many of the speech errors might have had their origin in missing important surgical details at closure of the soft palate. For instance, one of the papers stated, "the soft palate was closed directly with only minimal division of nasal mucosa and palatine aponeurotic fibers" (Cosman and Falk 1980). With no definite separation between the soft and hard palate, we suspect the surgeon had been unable to bring back the repaired velum to a position needed for achievement of velopharyngeal competence (VPC) on a regular basis. In other studies (Noordhoff et al. 1987), it was mentioned that SPR had been performed according to the original method of Perko (1979), but nothing was reported about use of a posteriorly based vomer flap. Omission of this crucial surgical step is likely to have caused reduced velar length in many cases and also a wider residual cleft in the hard palate due to less narrowing during early palatal growth. Tentatively, the short soft palate would increase the risk for VPI and posterior substitutions, such as pharyngeal and/or glottal articulations. In the report by Noordhoff et al. (1987), all patients treated with the two-stage method were said to have increased articulation errors, particularly those individuals with wide remaining cleft of the hard palate. A later follow-up paper (Liao et al. 2010) confirmed increased hypernasality and compensatory articulation disorders in these patients.

18.4.2 Maxillary Growth Results

More recently, a number of teams have, in particular, reported the patients' maxillary growth outcome with limited focus on their speech development. The majority of the papers described favorable midfacial growth (Noverraz et al. 1993; Tanino et al. 1997; Nollet et al. 2005, 2008; Sinko et al. 2008; Liao et al. 2010), while a few of the reports did not find any maxillary growth advantage of the two-stage protocol (Gaggl et al. 2003; Mølsted et al. 2005; Holland et al. 2007; Stein et al. 2007). The first group of papers generally advocated methods for SPR and especially for HPR, where closure of the cleft resulted in minimal denudation of bone in the palate. Examples of such procedures comprised employment of a turnover vomer flap, suturing of the cleft edges, mobilization of mucoperiosteal flaps dissected close to the cleft, or use of a modified von Langenbeck operation. On the other hand, in studies describing no maxillary growth benefits or inferior maxillary development, the methods used in these patients had created growth-restricting scars, mostly from HPR. Examples of surgical methods included employment of a Veau pedicle flap, a mucoperiosteal pushback procedure, or use of "unipedicled mucoperiosteal flaps." All of them will give rise to significant areas of bare palatal bone, which will heal secondarily, leading to scar tissue development. Thus, these different opinions about definite growth advantage or insufficient maxillary development after the two-stage palatal protocol can be explained by the performed repair methods and, only to a limited extent, by *when* surgery was done, as described above. Such explanations

are easier to embrace than speculation about effects, e.g., from cleft team organization (Shaw et al. 1992a, b) or surgeons' different operating skills (Ross 1987a, b), as reasons for the patients' growth results. However, we have to remember that acceptance or rejection of the two-stage regimen might be influenced not only by how well the repaired maxilla will develop but also by other factors, such as increased risk for fistula formation, poor speech development, and need for more velopharyngeal flaps (VPFs); etc.

18.5 Outcome of Maxillary Growth as Well as Speech from Teams with Long-Term Records

Only a limited number of publications exist, where the patients' maxillary growth as well as their speech development after two-stage palatal surgery has been studied up to adolescence or early adulthood. From this very small group of papers, we have chosen to report treatment outcome in unilateral cleft lip and palate (UCLP) patients from two European cleft teams still practicing the protocol.

18.5.1 Zürich, Switzerland

In Zürich, Switzerland, the cleft team has used their version of the two-stage palatal repair protocol since the late1960s. It was built on a systematic coordination between maxillary orthopedics and surgical interventions from early infancy (Hotz and Gnoinski 1976; Hotz et al. 1986). The team initiated preoperative maxillary orthopedic treatment shortly after birth, not only to help feeding but also for growth guidance of the maxillary segments. After lip repair at age 5–6 months, the infant continued with maxillary orthopedics until SPR, which was carried out at around 18 months. This surgery included reorientation of the velar muscles after their separation from the posterior hard palate (Perko 1979). The covering flaps of the oral mucosa were dissected from the posterior third of the hard palate, and, to reduce risks for maxillary growth impairment, the dissections were made *supra*periosteally. A posterior vomer flap was also raised, and after turning it backward, the flap was sutured as part of the anterior nasal layer at SPR (Hotz et al. 1986). With the nasal mucosa not separated from the posterior hard palate, inclusion of the vomer flap would not help elongate the soft palate, and therefore, a midline Z-plasty was added. At age 5–6 years, HPR was performed. Due to narrowing of the residual cleft, closure was accomplished by use of a turnover vomer flap for the nasal layer, and for repair of the oral layer, a mucoperiosteal flap was shifted medially from the noncleft side.

18.5.1.1 Follow-up Studies

The Zürich cleft team has published several follow-up investigations, particularly about maxillary growth. Roentgencephalometric results from a group of 10-year-olds born with UCLP indicated satisfactory A-P relationship between maxilla and mandible in about 80 % of the subjects (Gnoinski 1990). At age 15–20 years, these patients showed continuation of the favorable orofacial development, documented in the 10-year sample (Gnoinski 1991; Gnoinski and Haubensak 1997). Even if the cleft maxilla grew slightly in length also between 15 and 20 years of age, the average maxilla was shorter than in noncleft subjects. Only about 10 % of the patients needed maxillary orthognathic surgery to achieve an acceptable facial profile.

The team from Zürich has also reported cross-sectional, detailed speech data based on live assessments. After HPR and speech therapy, the patients' glottal or pharyngeal articulations were eliminated (Hotz et al. 1978). This outcome was in contrast to what was achieved with the previous treatment protocol. Nothing was mentioned about development of retracted articulations of certain consonants such as anterior plosives. Severe hypernasality decreased spontaneous with age, even before HPR was performed. After HPR, this speech error was claimed by the authors to have disappeared almost completely, often though, with help of intensive speech therapy. No pharyngeal flaps were judged by the team to be necessary in any of the Zürich patients.

18.5.1.2 Verification of the Zürich Speech Outcome

An outside investigator was recruited to verify the satisfactory speech development of the cleft children in Zürich (Van Demark et al. 1989). Thirty-seven UCLP patients were studied in a cross-sectional investigation at ages ranging from 6 to 16 years (mean 10.5 years). Oral examination generally revealed a mobile and fairly long soft palate, which was ascribed to the surgical method used for SPR. A comprehensive, independent speech analysis from audio recordings revealed that about 95 % of the patients displayed adequate or marginal VPC. The majority of subjects did not show speech errors such as glottal stops and pharyngeal substitutions. Even in the younger patients, the incidence of other compensatory articulations and "nasalizations" was low. The authors concluded that "good speech results for unilateral complete cleft lip and palate can also be achieved with late closure of the anterior palate, at least as done by the Zürich approach." Unfortunately, no further follow-up has been published.

18.6 Gothenburg, Sweden

When we planned the palatal surgery in our new protocol, the Zürich group had neither published any suggested ages nor detailed descriptions of their surgical procedures. For velar repair, we opted for an age around 6–8 months, while we wanted to delay closure of the remaining cleft in the hard palate to the mixed dentition. Regarding surgical details for SPR, we chose the approach to separate soft palate from hard palate right at the border region (Friede et al. 1980) (Fig. 18.3). The lateral incisions and denudation of palatal bone were to be made as minimal as possible. The velar muscle bundles should be united in the midline after complete release of the muscular fibers from their abnormal insertion at the pos-

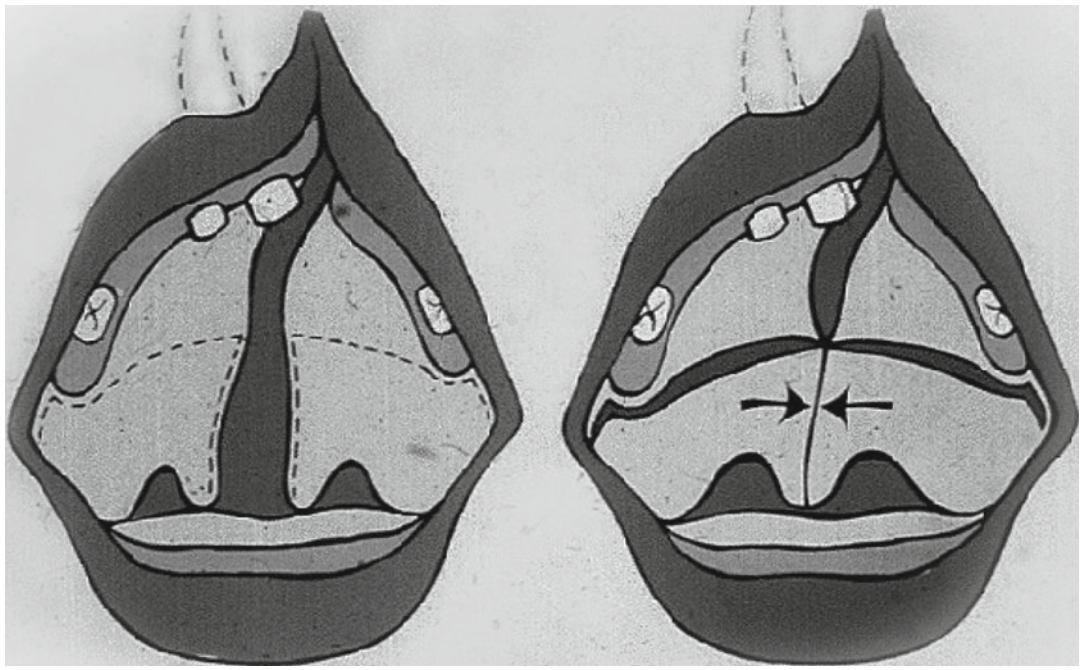

Fig. 18.3 Schematic illustration of SPR according to an early repair version used at Gothenburg Cleft Center. Incision lines run in the border region between velum and hard palate. The two halves of the soft palate, with the muscle bundles reoriented to a transverse course, are united in the midline (*arrows*)

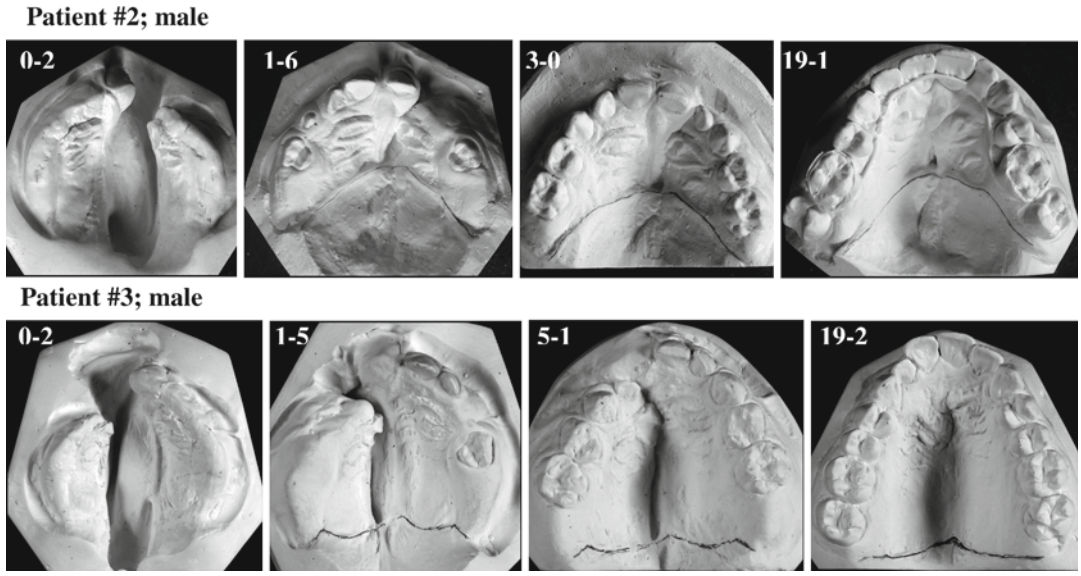

Fig. 18.4 Anterior-posterior variation in position of scar line from SPR. In *upper patient*, the scar line is too far forward, harming growth in length of the maxillary dental arch. In *lower patient*, the scar line is in correct position at posterior maxilla. The dental arch length is well developed with space for all teeth

terior border of the hard palate. The muscles remained attached to the nasal mucosa. At this stage of development of our method for velar repair, no posteriorly based vomer flap was raised and attached to the anterior soft palate. Therefore, the residual cleft of the hard palate did not narrow as much as expected. This made our surgeon dissect the palatal flaps further anteriorly, occasionally into the middle of the hard palate (Fig. 18.4). Such change would reduce the remaining cleft size and thereby enhance early speech development. However, after some time, we realized that this change in surgical detail at SPR also might restrict maxillary growth. It was first after introduction of the posterior vomer flap that the dissection line in the palatal mucosa returned to its previous position close to the edge of the hard palate. The Zürich surgeon Perko (1979) did not report in detail how their two-stage surgical procedures were carried out until 1979, and then, a posteriorly based vomer flap at SPR was not yet part of the Swiss surgical routine. It took another 1 or 2 years before many surgeons, including those from Zürich and Gothenburg, began utilizing this important addition to SPR.

18.6.1 More Detailed Two-Stage Palatal Surgery

In 1996, our cleft team published a detailed report how the two-stage protocol was practiced in Gothenburg up to 1995 (Lilja et al. 1996). At soft palate surgery, incisions began around the posterior part of the maxillary tuberosities and then followed a zigzag route at the posterior border of the hard palate (Fig. 18.5). A posteriorly based vomer flap was dissected. Anteriorly, the incision was placed behind the vomero-premaxillary suture, and the flap had its base close to the junction between vomer and the cranial base. Mucosal flaps in the soft palate were raised by blunt dissection. Hamulus was identified but not broken. The insertions of velar muscles, including their attached nasal mucosa, were cut at the posterior border of the hard palate. A flap with the muscles connected to the nasal layer was then dissected free and mobilized to a posterior position. After that, the muscles including the levator were reconstructed to a transverse course, where suturing could be performed without tension. The vomer flap was raised and the nasal layer of velum was closed anteriorly to the level of the muscular sling by use

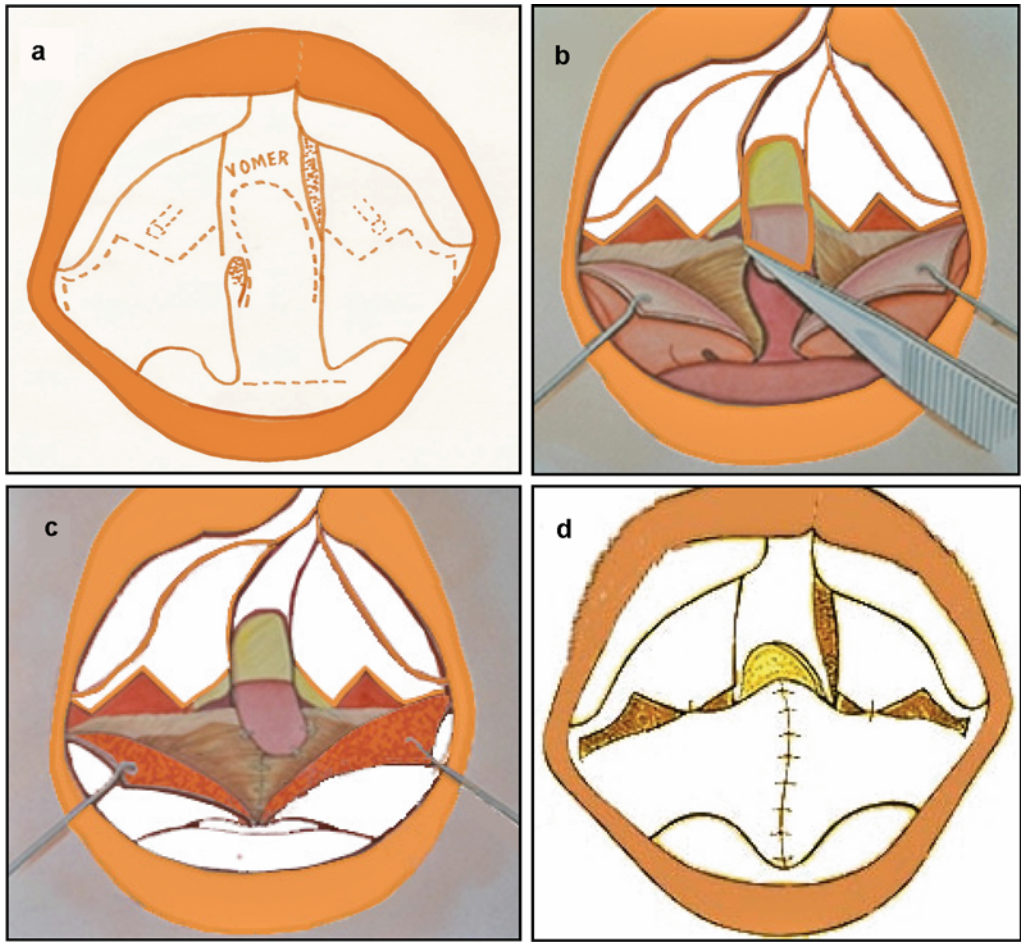

Fig. 18.5 Schematic drawings of the soft palate closure. (**a**) The incisions follow a zigzag line between the soft and hard palate. A posterior vomer flap is dissected, which has its base at the posterior-cranial part of vomer. (**b**) Both sides of velum are divided into two layers: the oral mucosa and the nasal mucosa with the forward inserting muscle bundles attached. The two layers are separated laterally and posteriorly to the uvula. Medially, the nasal layer with the muscle bundles is cut bilaterally from the posterior hard palate. (**c**) The muscles are redirected to a transverse course, sutured together medially in a posterior position, and also attached anteriorly to the backward-turned vomer flap. (**d**) The muscles and the raw surface of the vomer flap are covered by the oral flaps, which are pushed in a medial-posterior direction

of the vomer flap. In this way, the vomer bone became connected to the anterior velum. The palatal closure was then continued over to the oral side, where a pushback procedure was performed within the oral layer of the soft palate. Inclusion of the vomer flap as well as the pushback surgery helped to increase the length of the repaired velum.

The HPR was delayed until the patient had reached the stage of early mixed dentition (7–9 years), because, thereby, the procedure could be combined with bone grafting to the alveolar cleft. Surgery began with gingival incisions along the neck of the teeth on the palatal side and in part also labially (Fig. 18.6). In the area of the cleft, incisions were made along the cleft edges, and gingival and palatal mucoperiosteal flaps were raised. On the labial side, a back-cut was made in the cleft-side molar area. This facilitated mobilization of the gingival flap as well as the dissection in the cleft area. When the cleft had been dissected completely free on both palatal and labial side, suturing in the

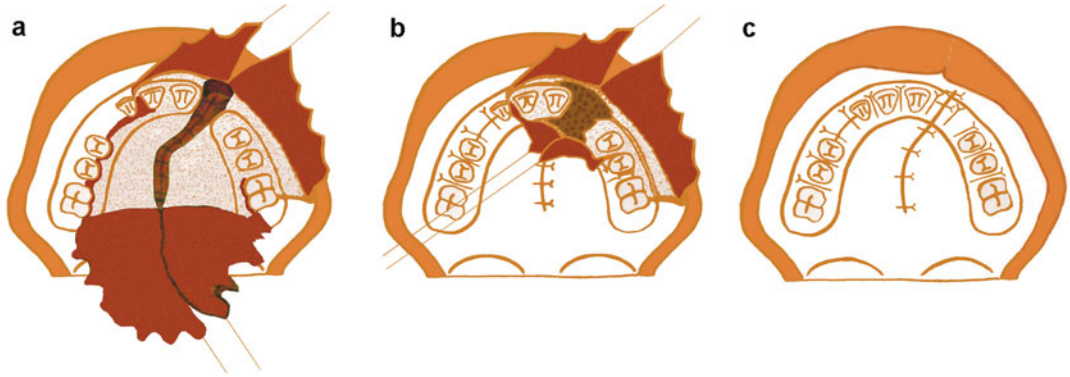

Fig. 18.6 Illustrations of the method for repair of the residual cleft in the hard palate in combination with bone grafting. (**a**) Incisions are made along the necks of the teeth and along the edges of the residual cleft. Palatal and gingival mucoperiosteal flaps are raised. The nasal layer is closed. (**b**) The palatal mucoperiosteal flaps are closed in the palate. Bone grafting is performed to the cleft in the alveolus. (**c**) The grafted bone is covered by gingival and anterior palatal mucoperiosteal flaps, which are sutured together

midline began in the nasal layer and continued orally in the midline palate. Cancellous bone was then grafted to the cleft in the alveolar process and covered with gingival and palatal flaps. The operation was completed with suturing the gingival and palatal mucoperiosteal flaps together in some interdental spaces. We consider the palatal incisions along the necks of the teeth very important. Because of the reduced width of the residual cleft, suturing back of the combined palatal flaps along the dental arch could be done with none or only minimal palatal bone exposed close to the teeth. In 1996, timing for HPR was modified in an effort to prevent development of typical retracted oral speech deviations noted in some patients during preschool and early school age. The repair was then changed to be carried out around 3 years, which meant an extra operation, because HPR could no longer be performed together with the bone grafting procedure. If the residual cleft of the hard palate was narrow or of average size, it was closed in one layer by use of a simple turnover vomer flap (Fig. 18.7). This approach did not work so well for wider residual clefts of the hard palate due to increased risk for fistula development. In these cases, a two-layer repair was necessary. A vomer flap was then sutured to the nasal mucosa on the cleft side, and the suture line was closed with a mucoperiosteal flap from the cleft side of the palate (Fig. 18.8).

18.6.2 Follow-up Studies of Maxillary Growth

Over the years, the Gothenburg cleft team has reported several follow-up studies, both regarding maxillary growth and speech development. An early growth comparison at age 7 years, before HPR in the two-stage protocol, demonstrated significantly improved results in comparison to what was achieved with our previous regimen (Friede et al. 1987). In a more recent longitudinal follow-up study, we compared 30 consecutive UCLP patients, who had been subjected to our two-stage palatal surgery, to a similar sample from another center. Those patients had undergone HPR with a vomer flap at age 3 months, which was followed by SPR with a pushback method at 22 months (Friede and Enemark 2001). Comparison of roentgencephalograms, obtained during the age span from prepuberty to adolescence, demonstrated that our patients with delayed HPR had much better midfacial development than the other group of subjects. This difference was also noticed in the percentage of patients with need for later maxillary advancement surgery (10 % vs. 30 %). The favorable maxillary growth outcome in our patients was, some years later, confirmed in a cast study of 104 consecutive subjects with UCLP (Lilja et al. 2006). If just considering casts

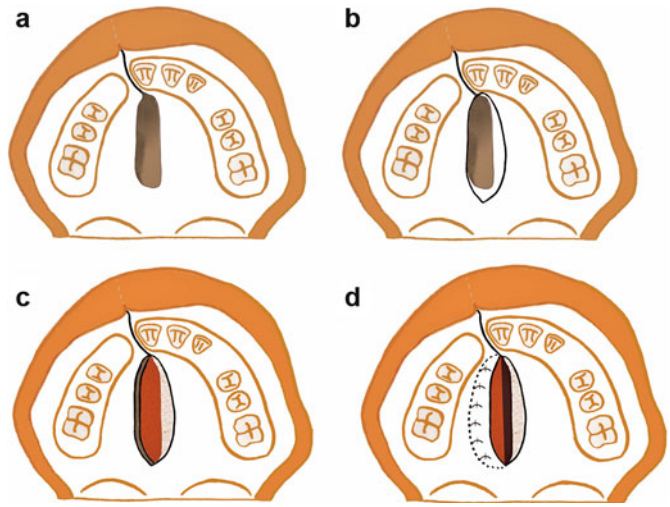

Fig. 18.7 Composite of drawings illustrating repair of a small- or average-sized residual cleft in the hard palate at age 3 years or preferably earlier. At this stage, bone grafting is not performed in connection with the HPR as shown in Fig. 18.6. (**a**) The residual cleft in the hard palate. (**b**) Incision lines. On the noncleft side, the incision goes into the flat medial part of the palate. On the cleft side, the incision is made at the cleft border between oral and nasal mucosa. (**c**) The vomer flap is raised. It contains some millimeters of oral mucosa leaving a raw bone surface in the medial part of the palate on the noncleft side. On the edge of the cleft side palatal shelf, a subperiosteal dissection is performed and a pocket is created between the oral periosteum and the bone. (**d**) The vomer flap is tucked into the pocket and sutured, which finalizes HPR. Thus, this is a one-layer closure, and the raw surface of the vomer flap is left for secondary epithelialization

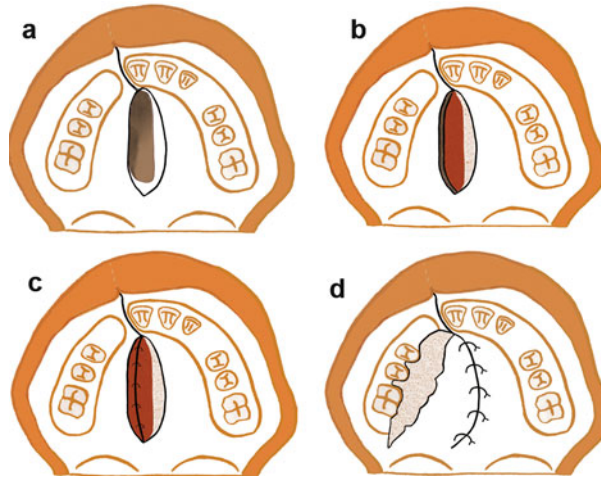

Fig. 18.8 Drawings illustrating HPR in an individual with a somewhat wide residual cleft. Preferably, the patient should have erupting/erupted upper deciduous molars, which usually means an age at around 3 years. (**a**) Incision lines. On the noncleft side, the incision goes into the flat medial part of the palate. On the cleft side, the incision is made at the cleft border between oral and nasal mucosa. (**b**) The vomer flap is raised. It contains some millimeters of oral mucosa leaving a raw bone surface in the medial part of the palate on the noncleft side. (**c**) On the cleft side, a subperiosteal dissection is performed, and the nasal layer is lifted and brought in contact with the vomer flap and sutured. (**d**) Also on the cleft side, an oral mucoperiosteal flap is raised via an incision along the teeth. The flap is brought in contact with the incision on the noncleft side and sutured. The suture line between the vomer flap and the nasal layer is now covered. The raw bone surface is left for secondary epithelialization, which takes place on a surface with thin neighboring wound edges. Thereby, the risk for bad scar contraction is small

obtained at around 19 years, an acceptable dental relationship, reported as GOSLON scores 1, 2, and 3 (Mars et al. 1987), was found in 97 % of the subjects. This was rated as an exceptionally high incidence of satisfactory occlusion at early adulthood.

For our most recent longitudinal follow-up study (Friede et al. 2011), we used a consecutive series of 50 patients born with UCLP. All subjects had lateral roentgencephalograms from four selected age stages from preschool to early adulthood. Besides lateral cephalograms, we also studied maxillary casts from an age around 1.5 year to score the A-P position of the mucosal scar line from the dissection between soft and hard palate at SPR. Interestingly, we found that those patients with more anteriorly placed scar lines had definitely reduced maxillary lengths as appraised in their cephalograms from age 19 years (Figs. 18.9 and 18.10). This illustrates how small details at surgical reconstruction of the UCLP are important for optimal maxillary development during subsequent growth. However, the overall favorable midfacial development, noticed in our previous outcome reports, was confirmed in this study. In early adulthood, the patients' mean values for the skeletal profile convexity and for the sagittal jaw relationship were close to or within the 95 % confidence interval of the mean for noncleft Swedish subjects (Thilander et al. 2005). Generally, the average UCLP patient's facial morphology displayed a skeletal pattern in harmony with the retrognathic maxilla and mandible characterizing this group of patients (Segner 1989) (Figs. 18.11 and 18.12).

18.6.3 Follow-up Studies of Speech Outcome

Most of the reports on speech development from Gothenburg cleft center have been retrospective, longitudinal, follow-up studies of individuals born with UCLP and have, by and large, investigated consecutive series of patients. It should be mentioned though that particularly the early reports employed limited patient samples and, in addition, more than one surgeon had carried out the palatal repairs. We are aware that these facts might, to some extent, have reduced the reliability of our results. However, on the positive side, we must mention that all speech samples were rated using standardized audio recordings, which were identity blinded to the investigators and independently evaluated by two experienced speech-language pathologists, and in the later studies also by an external listener.

18.6.3.1 Early Results

Our initial speech results were reported in the beginning of the 1990s when our first patients, treated according to the two-stage palatal surgery protocol, were evaluated during their early childhood (Lohmander-Agerskov and Söderpalm 1993; Lohmander-Agerskov et al. 1995). *Hypernasality* and *audible nasal emission* occurred among the 5-year-olds before HPR in a moderate-to-severe degree in about 40 and 30 %, respectively. For hypernasality, the prevalence decreased to about 10 % at age 7, which still was an evaluation *before* HPR. For audible nasal emission, however, in most patients, the occurrence remained the same until about 1 year *after* HPR. The prevalence for both hypernasality and audible nasal emission were then reduced to occur in about 6 % of the patients. The gradual reduction in size of the remaining cleft was suggested as explanation for lowering the hypernasality rate even before hard palate closure. Supposedly, the smaller cleft size resulted in reduced influence on resonance from the nasal cavities (Lohmander-Agerskov et al. 1997). In contrast, there was no relationship between the aerodynamic-related variable audible nasal emission and the size of the residual cleft.

The early outcome studies on *articulation errors* revealed very rare occurrence of *glottal/pharyngeal misarticulation*s. Low incidence of these speech deviances was considered an indication of adequate VPC in the vast majority of our patients. Instead, we discovered so-called retracted oral articulation of dental/alveolar plosives to a palatal or velar place, which, particularly before HPR, was a rather common finding (Lohmander-Agerskov 1998).

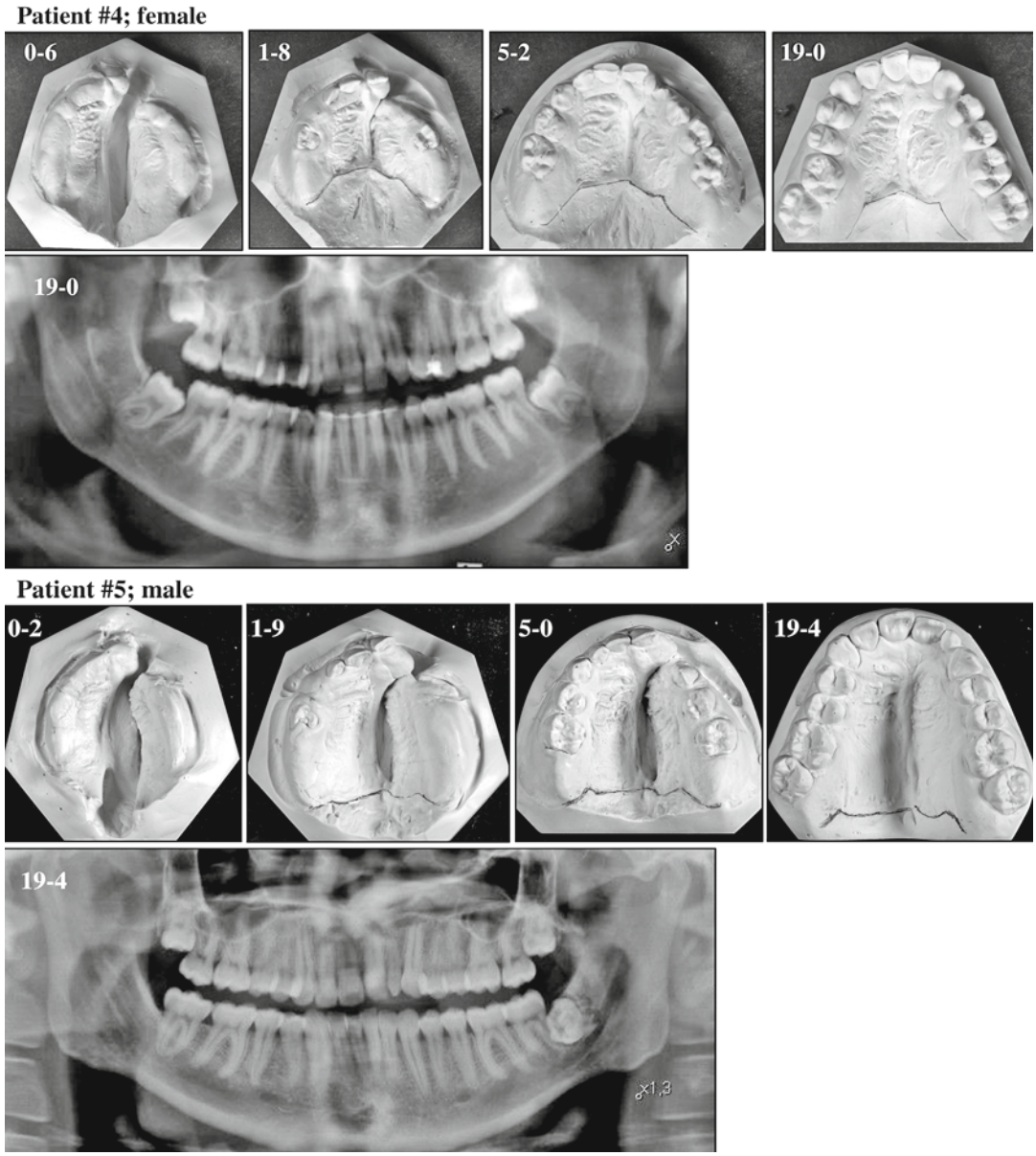

Fig. 18.9 Casts and orthopantomograms from two patients illustrating different development of maxillary dental arches. The *upper patient* demonstrates slight crowding at early adulthood in spite of no permanent lateral incisor on the cleft side. In the *lower patient*, the dental arch is well developed with space for all permanent teeth. Note more posteriorly positioned scar line after SPR in this patient compared to the upper one. See Fig. 18.10 for midfacial development

In different reports, we recorded prevalences between 30 and 40 % with little reduction in rate before HPR and with weak, but significant, correlation to size of the residual cleft (Lohmander et al. 2002). The varying degrees of the retracted articulation errors after our two-stage protocol occurred up to early childhood in about one third of the children. However, the speech deviation was gradually reduced to half of that at age 10 years, and around adolescence, the errors had disappeared. An interesting finding was that children who had had a spontaneous, early, func-

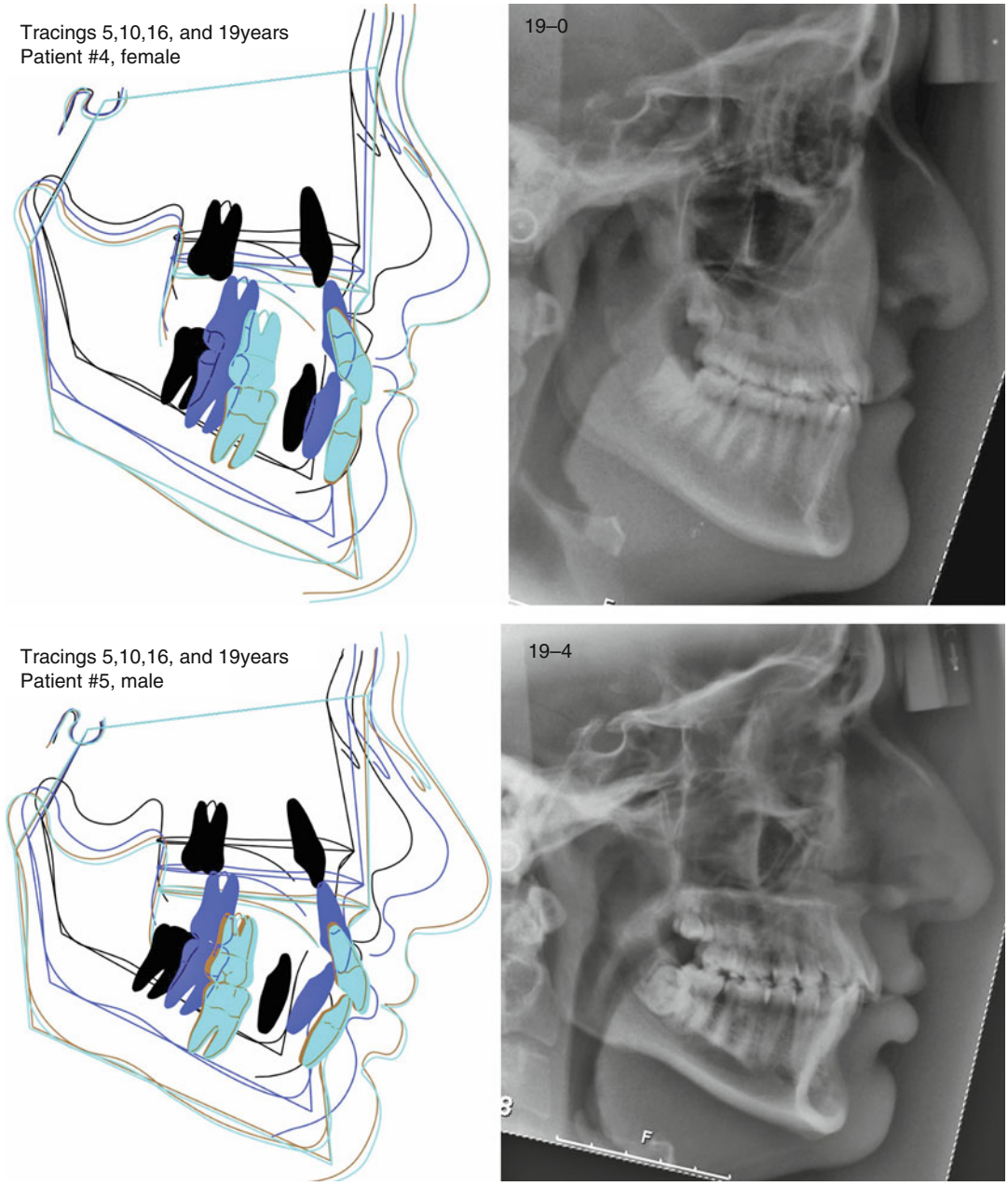

Fig. 18.10 Superimposed tracings at four age stages in addition to cephalograms at young adulthood of the two patients illustrated in Fig. 18.9. Notice better development of the maxilla of patient #5 compared to patient #4

tional closure of the residual cleft (Fig. 18.13) (evaluated before age 3 years) did not display any articulation errors at later ages (Lohmander-Agerskov et al. 1996). In another early study of possible factors related to development of retracted oral articulations, we found that children with this speech error at ages 3 and 5 years were more likely to have been babies prefering to use a posterior place of articulation at babbling or maybe, more correctly, lacking anterior articulation at this stage of speech development (Lohmander-Agerskov et al. 1998).

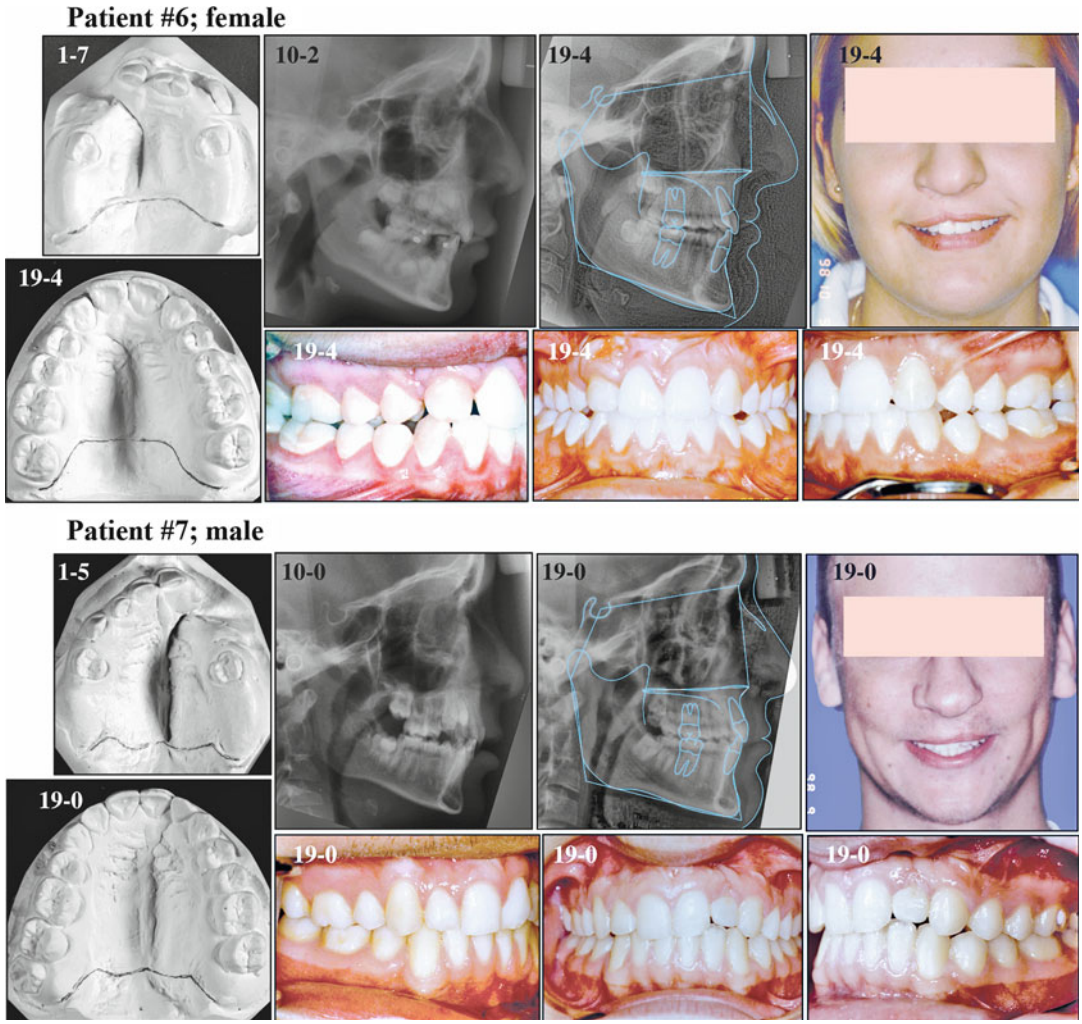

Fig. 18.11 Composite of casts, photographs, and cephalometric records from two typical patients treated according to the Gothenburg two-stage palatal surgery protocol. No orthognathic surgery have been performed. *Upper patient* showed satisfactory sagittal growth of the maxilla but, partly due to missing laterals, developed bilateral crossbite. Patient declined further surgery of her upper lip. Good speech developed already from early age. *Lower patient* displayed excellent maxillary growth, both in sagittal and transverse dimension. Treated with a velopharyngeal flap at around 3 years due to short velum. In spite of narrow residual cleft, the patient developed retracted oral articulation. It turned out to be a phonological backing process, which disappeared with speech therapy after some years. Still some audible nasal emission at age 19 years

When we in 1996 made an effort to deal with noticed speech concerns, particularly the retracted oral articulation, we chose to lower the age for HPR. The timing of this surgery was gradually changed from a previous mean age of 8 years down to 3 years. Interestingly, a follow-up investigation did not reveal any significant speech improvements in spite of the earlier HPR (Lohmander et al. 2006). The same prevalence of retracted articulation errors was found in these patients as in previous individuals, who had been subjected to HPR after age 8 years. Therefore, we concluded that if repair of the residual cleft was to have any potential for improvement of the patients' speech outcome, palatal surgery should be completed before age 3 years.

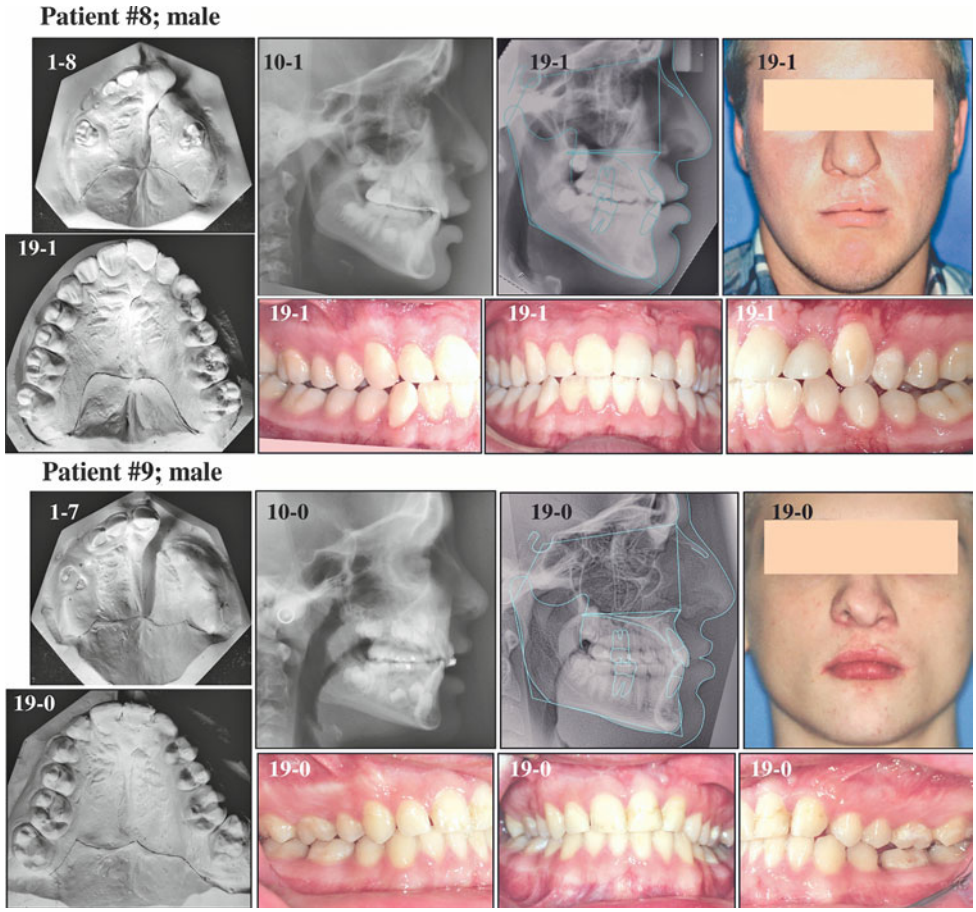

Fig. 18.12 Illustration of records from two representative patients treated with the two-stage method for palatal repair at Gothenburg cleft center. Outcome obtained without maxillary orthognathic surgery. *Upper patient* displays an overall acceptable intermaxillary relationship in spite of slight maxillary deficiency, compensated for by proclination of the upper incisors, and also causing a tendency to bilateral crossbite. Good speech development since early ages. *Lower patient* demonstrates good maxillary development despite congenitally missing lateral incisor on the cleft side. Quite satisfactory early speech but has developed moderate hypernasality during postpubertal years

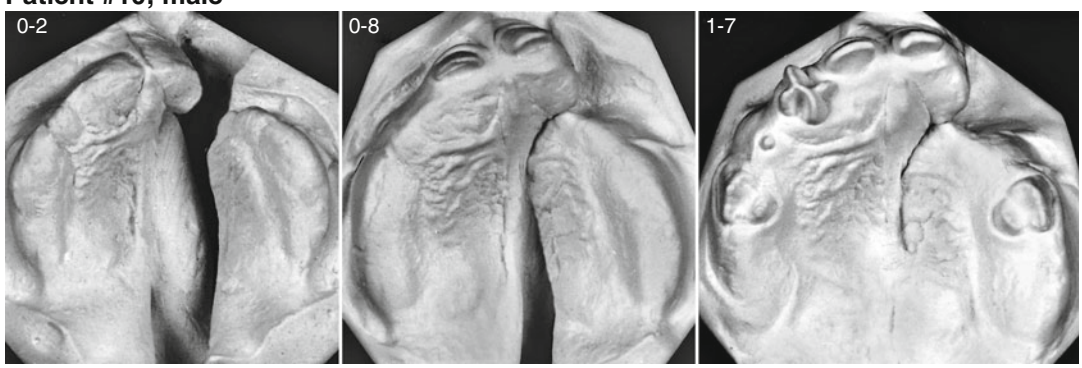

Fig. 18.13 Spontaneous functional closure of the cleft in the hard palate from a series of casts from the same patient. The first cast at age 2 months was obtained before lip adhesion surgery, the second model from before velar closure, and the third cast from before final lip/nose repair

18.6.3.2 Longitudinal Long-Term Follow-up Results

A longitudinal study of a consecutive series of 55 patients with UCLP, obtained from a total cohort of 65 individuals, completed our follow-up of speech development after the Gothenburg original two-stage palatal protocol as used up to 1995 (SPR at 6–8 months and HPR at 7–9 years) (Lohmander et al. 2011, 2012). Based on obtained standardized audio recordings, blindly analyzed at ages 5, 7, 16, and 19 years, and clinically assessed at 10 years of age, we were able to verify previous findings, both the positive and the compromised ones, regarding our patients'speech development. Hypernasality, audible nasal emission, and retracted oral articulation occurred in about 30 % of the 5-year-olds, i.e., when their cleft in the hard palate still was unoperated. Generally, all those three speech errors were, however, markedly reduced during the following years, and their prevalence was low at age 16 and 19 years. As a result of these enhancements and other spontaneous speech improvements, the prevalence of VPC was judged as quite satisfactory with a recorded prevalence of 82 % at age 16 and 87 % at age 19 years; at both ages, the patients' speech intelligibility was judged as normal. It should be added that secondary VPF surgery had been performed in six of the 55 patients (11 %). Our main conclusion was that speech improved in children treated according to our two-stage palatoplasty protocol even before HPR. In addition, further small improvements occurred until early adulthood. An added advantage was that the prevalence of other, more severe cleft speech characteristics, such as symptoms related to VPI and particularly the occurrence of glottal misarticulations, was very low. An almost similar picture can be seen in Fig. 18.14, which displays cross-sectional data for the same ages but with inclusion of twice as many patients at ages 5, 7, and 10 years as investigated in the longitudinal, long-term follow-up. In addition, information at 3 years of age is also incorporated. Our longitudinal results (Lohmander et al. 2011, 2012) together with these cross-sectional data give a trustworthy picture of the long-term speech outcome after the original palatal repair procedures at the Gothenburg cleft center.

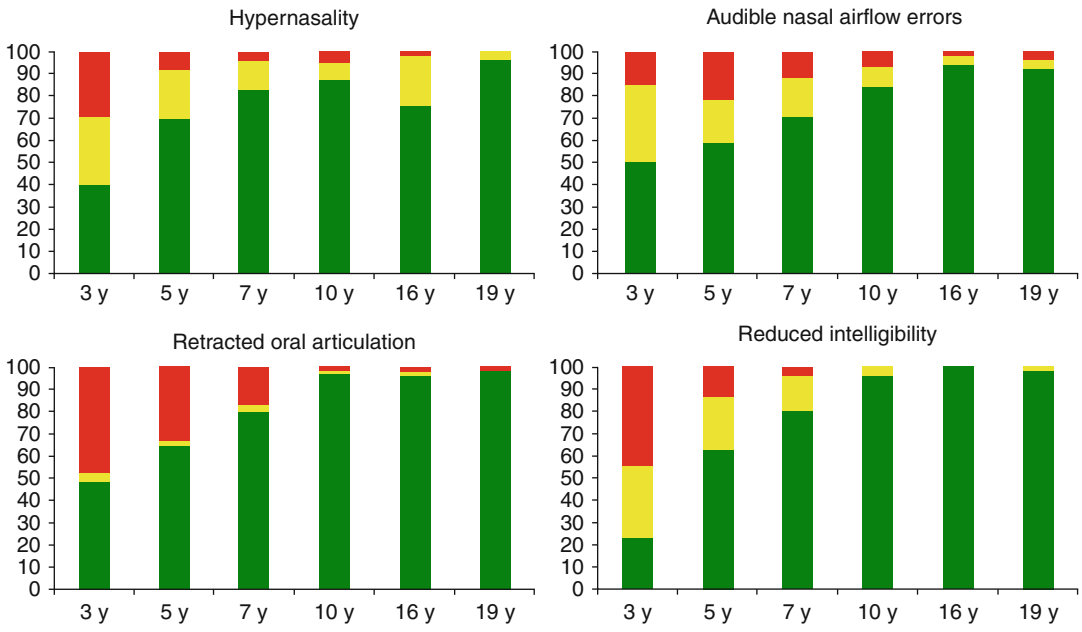

Fig. 18.14 Prevalences (%) of hypernasality, audible nasal airflow errors, retracted oral articulation, and reduced intelligibility in a cross-sectional series of patients born with UCLP. The number of patients included at each age were: 3 years = 45; 5 years = 93; 7 years = 101; 10 years = 88; 16 years = 50; 19 years = 46. A majority of the final patients (37) were included longitudinally from age 5 years (Lohmander et al. 2011). *Green* no/normal; *Yellow* Mild; *Red* Moderate-Severe

18.6.3.3 Retracted Oral Articulations

The speech deviation coined as "retracted oral articulation of anterior pressure consonants," occasionally called "backing," was the most common speech error affecting about one third of our preschool children born with UCLP (Lohmander et al. 2011, 2012). Similar speech problems have been reported from other centers and is sometimes considered an atypical phonological process (e.g., Timmons et al. 2001). However, if there still is a coupling between the oral and the nasal cavities, consonants normally produced in a place anterior to the communication may be retracted to a place behind the oronasal opening. With this in mind, the process should be regarded as a compensatory strategy for the inability to create sufficient intraoral pressure to produce high-pressure consonants (Fig. 18.15). This specific speech error is not necessarily incorporated in the child's internal phonological system and will quite often recede without speech therapy.

18.6.3.4 Results Related to Early Speech Development

An important rationale for early SPR would be that it makes it possible for the patient to achieve increased velopharyngeal activity and oral place of articulation already during the early babbling stage. As an example of such achievements, it can be mentioned that a proportionally high occurrence of oral stops at 12 and 18 months has been reported in studies, which have described outcomes associated with early SPR at our center (Lohmander et al. 2004, 2011, 2012) as well as in another Scandinavian center practicing the same surgical method (Willadsen and Albrechtsen 2006). The continuity between early consonant production and later speech has been described to occur in noncleft children (e.g., McCune and Vihman 2001) as well as in patients born with cleft palate and treated according to different surgical protocols (Chapman et al. 2003; Lohmander and Persson 2008; Scherer et al. 2008). A high number of consonant types and plosives, particularly with anterior placement during early speech development, were related to articulation accuracy, measured as percentage of correct consonants (Fig. 18.16) (Lohmander and Persson 2008), as well as with the vocabulary development (Scherer et al. 2008). Thus, in babbling, certain characteristics are important for later speech and language development. Early SPR seems to enhance this development.

18.7 The Scandcleft Study

At the Zürich, Switzerland, International Symposium on Early Treatment of Cleft Lip and Palate in 1984, there were intense discussions regarding the advantages and disadvantages of different surgical procedures for treatment of patients with cleft lip and palate. Generally, these

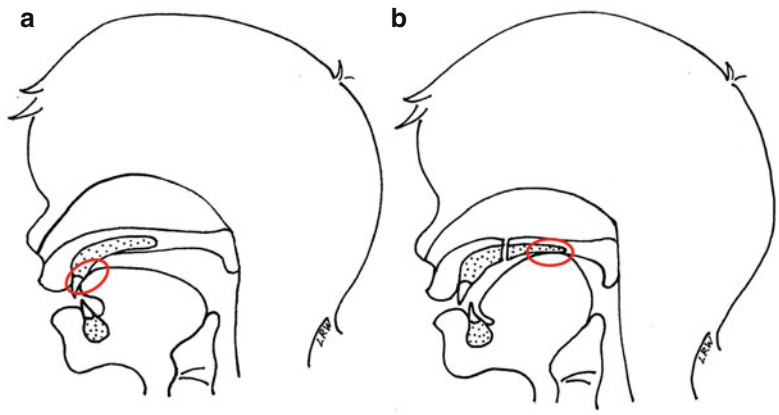

Fig. 18.15 (**a**) Correct place of articulation for dental/alveolar consonants and (**b**) retracted to velar placement, behind the oronasal opening. With adequate velopharyngeal function, the place of articulation is posterior to the oronasal opening but in front of the velopharyngeal port, i.e., a compensatory strategy for the inability to create sufficient intraoral air pressure to produce high-pressure consonants in front of the opening

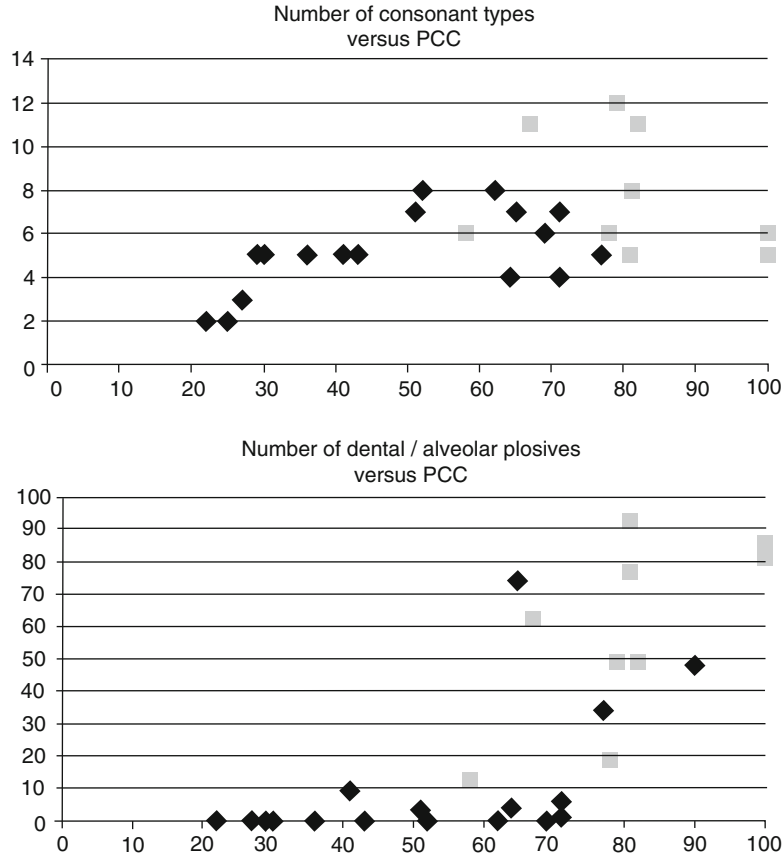

Fig. 18.16 (*Upper chart*) A high number of consonant types (inventory) and (*Lower chart*) a high number of dental/alveolar plosives at 18 months of age (=*y*-axis); both correlated significantly with a high percentage of consonants correct (PCC) at age 3 years (= *x*-axis) (Rho = .57, $p < .05$; Rho = .68, $p < .01$) in both UCLP (*black*) and comparison group (*gray*) (Lohmander and Persson 2008)

debates triggered growing interests in multidisciplinary inter-center research comparing outcome after different surgical protocols used for CLP treatment. As examples of the new trend, surgical results in four Scandinavian centers were analyzed, especially with regard to maxillofacial development (Friede et al. 1991; Enemark et al. 1993). Members of six European cleft centers made similar efforts (Shaw et al. 1992a,b; Mars et al. 1992; Asher-McDade et al. 1992; Mølsted et al. 1992, 1993a,b; Morrant and Shaw 1996), but here also, aspects on speech development were included in the outcome analyses (Grunwell et al. 2000). However, soon it became obvious that it would be impossible to separate and compare single elements of treatment protocols used at the different centers. Therefore, Scandinavian cleft teams from Helsinki, Stockholm, Linköping, Gothenburg, Copenhagen, Oslo, and Bergen together with two teams from the UK, Manchester and Belfast, decided to start randomized controlled trials (RCTs) regarding effects of primary surgery (particularly palatal repair) in UCLP. These prospective RCT studies were designed as a set of three parallel trials, where groups of teams would test their local protocols against a common protocol.

This was how the Scandcleft study was initiated (Semb 2001). The common protocol was a modification of the Gothenburg two-stage method for palatal repair. The modified surgical detail was that the palatal muscles were dissected free from the nasal mucosa and moved posteriorly. The muscles were sutured together in this position to form a posteriorly placed muscular sling. The nasal layer was left intact and closed with help of the posterior vomer flap. This is an early form of intravelar veloplasty, where the levator muscle was not identified and, consequently, not completely released. In the common protocol

used by all teams, the residual cleft in the hard palate was closed at age 1 year. Inclusion of new patients to the Scandcleft study was stopped 5 years ago and evaluation of the results began in 2012.

The local protocol in Gothenburg will be compared to the common protocol, and the parameter to be studied will be timing of surgery. Therefore, we had to use the modified procedure also in the local protocol, where the residual cleft was closed at 3 years. This leg of the study was performed in Copenhagen and Gothenburg, with the majority of the patients operated in Copenhagen.

18.8 Final Comments and Reflections

As a summarizing evaluation of the Gothenburg two-stage platal surgery regimen, we have to judge the protocol as quite successful. Both from midfacial growth point of view and regarding speech development, the chosen surgical method has exceeded the expectations we had when the protocol was initiated in 1975.

During the early years after our change of palatal protocol, we introduced several surgical details, which proved important for later outcome. One such feature was where to separate velum from the hard palate at SPR. Preferably, it should be done right at the level of the posterior palatal shelves. Separation further anteriorly would often leave denuded areas of bone, where growth-restricting scar tissue would form. A surgical procedure related to HPR should also be mentioned. It has been described in many papers dealing with so-called pushback palatal surgery. Wide palatal flaps, raised for medial and posterior displacement at repair of the cleft, will often include parts of the thick oral mucoperiosteum covering the maxillary body. Scars in these lateral parts of the palate seem to be especially harmful for later maxillary growth. Such wide flaps should not be used at HPR in a two-stage protocol, because the remaining cleft in the hard palate has usually narrowed substantially after SPR. Therefore, it can be repaired, e.g., by suturing the cleft edges after their mobilization, by employment of a simple turnover vomer flap, or, alternatively, by use of small flaps, raised in the thin mucoperiosteum close to the cleft. Shifting of these flaps medially, to cover the residual cleft, will result in scars with minimal maxillary growth restriction. Using such an approach would therefore allow for HPR at an early age (15–18 months) in most patients without jeopardizing subsequent maxillary development.

Inclusion of a posteriorly based vomer flap at velar repair seems to be an essential factor to facilitate narrowing of the remaining cleft during subsequent palatal growth and thereby enabling early HPR. In addition, the flap appears to improve the patient's ability to achieve adequate VPC. A likely explanation might be that the repaired velum can attain a more posterior and upward position than if this surgical detail is omitted.

A good velar function after early, successful veloplasty together with substantial reduction in size of the residual cleft in hard palate has been the most important factor to explain a satisfactory speech outcome after two-stage palatal surgery (Van Demark et al. 1989; Persson et al. 2002; Lohmander et al. 2006). Studies at Gothenburg cleft center of 3-year-old patients have revealed that speech developed typically (= as in individuals without a palatal cleft) in about 50 % of the children with UCLP in spite of an unoperated cleft in the hard palate. This outcome compares as good as or even favorably to what have been reported in the literature after *any* palatal surgery protocol (Lohmander 2011). The typical speech error among our patients at this early age was "retracted oral articulation," which occurred in about 40 % of the 3-year-old subjects. However, occurrence of compensatory articulations related to VPI was very low and absent in the absolute majority of patients, which indicated satisfacory VPC, before the residual cleft in the hard palate had been repaired. Some researchers and experienced clinicians have expressed convictions that such a remaining open cleft would have a negative impact on the patient's velopharyngeal function and make it difficult or impossible to achieve VPC. These beliefs have not been verified though, neither in studies from Zürich nor from

Gothenburg. Actually, a high occurrence of plosives, although retracted to velar place beyond the opening in the hard palate, *presupposes* VPC. Although not wanted, we consider this oral articulation error (retracted oral articulation) much less severe than non-oral misarticulations (glottal and/or pharyngeal) because of its only minor impact on speech intelligibility. Furthermore, a significant number of children with retracted oral articulation improve their speech before school age spontaneously.

Early soft palate closure seems to give a better condition for early consonant development than if the entire palatal cleft is left unoperated until around 12 months of age. Early use of palatal obturators before and after early soft palate repair (6 months) in a two-stage regimen or before palatal closure in a one-stage protocol does not appear to enhance this process (Hardin-Jones et al. 2002; Lohmander et al. 2004). Early hard palate closure at 12 to 18 months of age would possibly reduce the prevalence of the oral speech errors and together with early soft palate closure give the best conditions for speech development taking other aspects of treatment into consideration.

The Gothenburg two-stage surgical protocol as described here is easy to teach and also easy to learn. The first stage, SPR, is based on open dissections of wide and thick flaps without any closed undermining. This makes the procedure a simple, fast, and predictable method with few or no dehiscences postoperatively. In developing countries with limited resources of cleft surgery, orthodontics, or speech therapy, we believe the 2-stage regimen, described by Kontos et al. (2001), will result in better outcome than what is achievable with other standard surgical protocols. If our initial surgery is performed during the patient's first year of life, the outcome will usually be very adventageous. Lip closure and SPR can be combined in one primary operation with good result in a great number of patients, even if the hard palate never is repaired. However, about one-third of the children are likely to need HPR to develop acceptable speech. As the residual cleft is surrounded by virgin tissue, this surgery will be easier and give a predictable, better result than after closure of a palatal fistula. In most cases, the dental arch will develop with only minor irregularities, such as one or sometimes two deciduous teeth in crossbite on the cleft side, a narrow remaining cleft in the palate and alveolar ridge, and some rotation of the permanent central incisor on the side of the cleft.

The Gothenburg procedure with combined lip closure and SPR has been tested in cooperation with Universities of Campinas and Rio de Janeiro in Brazil, where patients with UCLP were operated on at different ages. The advocated Gothenburg protocol produced good occlusal development and acceptable narrowing of the residual cleft, if the Brazilian children had their surgery done early during their first years of life. However, if patients were older than 3 years, the residual cleft of the hard palate did not narrow very much over time. Therefore, in these cases, HPR could just as well be performed already 6 months after SPR.

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Part VIII

Facial Growth: Time Is the Patient's Ally

19 Management of the Premaxilla/Maxilla in Bilateral Cleft Lip and Palate

Karin Vargervik and Snehlata Oberoi

19.1 Presurgical Treatment Approaches Used to Manage the Prominent Premaxilla

Management of the prominent premaxilla in bilateral cleft lip and palate remains challenging, and consequently many different approaches have been tried.

Surgical setback at time of lip repair is one approach that provides immediate resolution of the excessive prominence. However, follow-up studies have demonstrated severe deleterious effects on midface growth resulting from this treatment (Bishara and Olin 1972; Friede and Johanson 1974; Vargervik 1983).

The fact that most often the midface becomes underdeveloped over time after surgical setback is in part due to the inherent growth characteristics of the premaxilla in bilateral clefts (Friede and Pruzansky 1972; Latham 1973; Vargervik 1983; Trotman and Ross 1993; Padwa et al. 1999).

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In a study from our center on 63 individuals with complete bilateral cleft lip and palate, 12 had surgical setback of the premaxilla (Vargervik 1983). The surgical setback had been done before the patient presented to our center, and details of the procedures were not available for all of them. It was shown that all 12 demonstrated very deficient midface growth that gradually became more significant with age. It was also shown that in the other 51 subjects who had conservative treatment of the premaxilla, the position of the premaxilla was on the average prominent until age 12 from which time the maxilla fell behind compared with normative data (Fig. 19.1). The study also showed that mandibular growth on an average did not differ from that of a sample of noncleft subjects (Fig. 19.2).

In most centers, surgical premaxillary setback in the infant has been limited to extreme cases where taping or other premaxillary manipulation has not succeeded in reducing the premaxillary prominence enough to allow lip closure. There may also be circumstances where the patient comes for treatment at a later age with an untreated prominent or vertically overdeveloped premaxilla where surgical repositioning cannot be avoided (Murthy 2009). A recent three-center study reported negative effects at age 12 years of premaxillary repositioning of the premaxilla at the time of alveolar bone grafting at an average age of 9.9 years (Bartzela et al. 2011).

S. Berkowitz (ed.), *Cleft Lip and Palate*,
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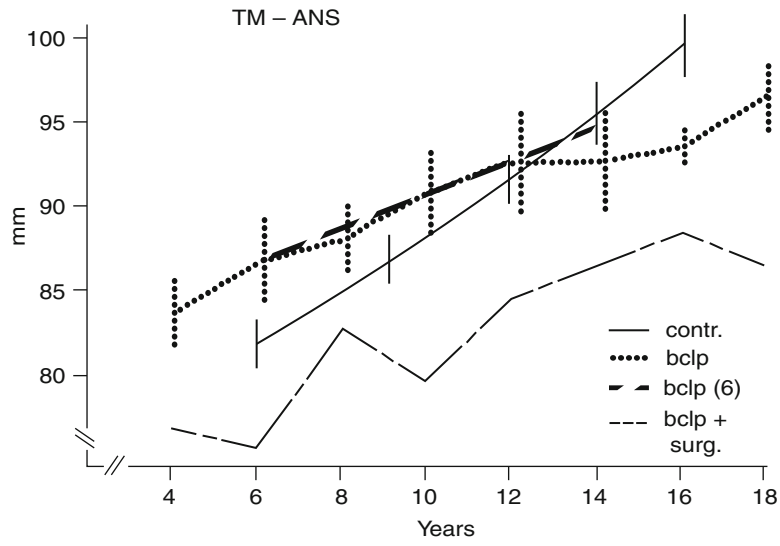


Fig. 19.1 A graph depicting forward growth of the maxilla in three groups of individuals with bilateral cleft lip and palate and a group of controls (Reprinted with permission from Vargervik (1983))

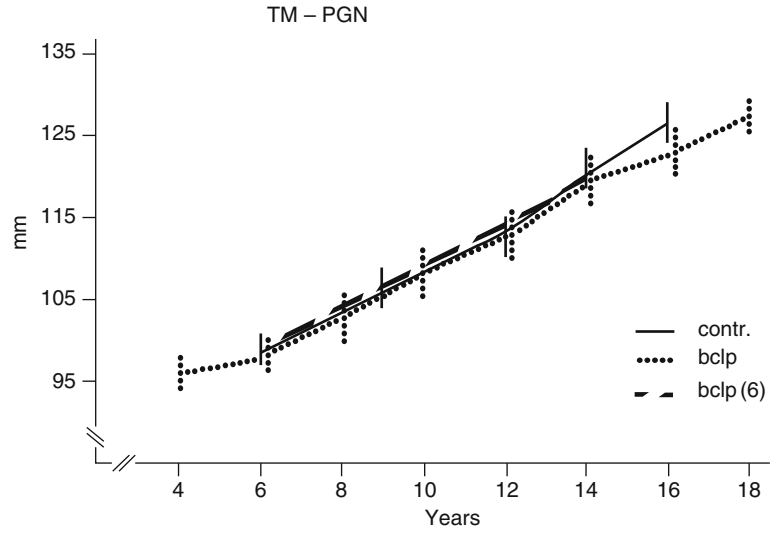


Fig. 19.2 Mandibular length increase (Reprinted with permission from Vargervik (1983))

Less drastic measures to achieve retraction of the premaxilla prior to lip repair include the "Latham appliance." When successful, this may retract the entire premaxilla to a position between the lateral segments. This has also been proven to have a deleterious effect on the forward growth of the maxilla (Berkowitz et al. 2004). Thus, it appears that any approach that results in retraction of the entire premaxilla, including the anterior nasal spine area, has the potential to contribute to maxillary growth inhibition.

The currently often-advocated nasoalveolar molding therapy may also succeed in bringing the entire premaxilla back to a position between the lateral segments (Grayson and Cutting 2001).

This will facilitate a tension-free lip repair and may also allow periosteoplasty if desired. However, most centers are not advocating

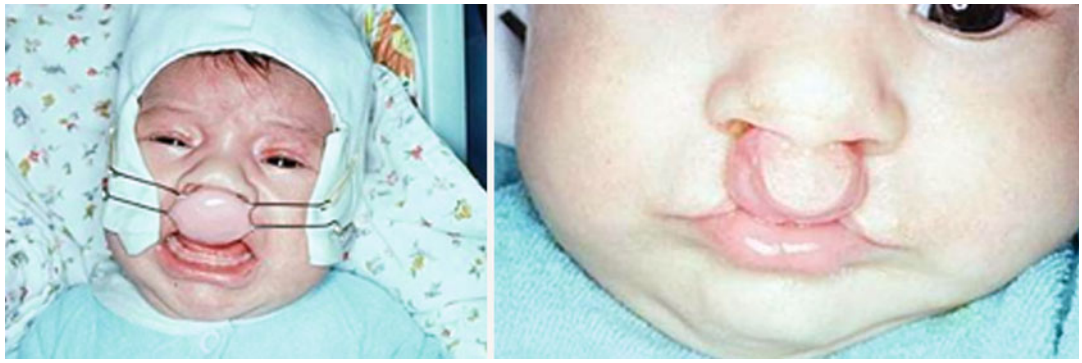


Fig. 19.3 Retraction of prominent premaxilla with appliance attached to a bonnet

periosteoplasty or bone grafting in infancy as this also has been shown to contribute to maxillary growth inhibition and to interfere with later expansion. Many studies since the procedure was first used by Skoog in 1965 have shown this. A recent study again documented the deleterious growth effects of periosteoplasty/bone grafting in infancy (Hsin-Yi Hsieh et al. 2010). Long-term positive effects on growth, esthetics, and parent satisfaction from any presurgical infant orthopedics have not yet been documented, according to a recent systematic review of the literature (Uzel and Alparslan 2011).

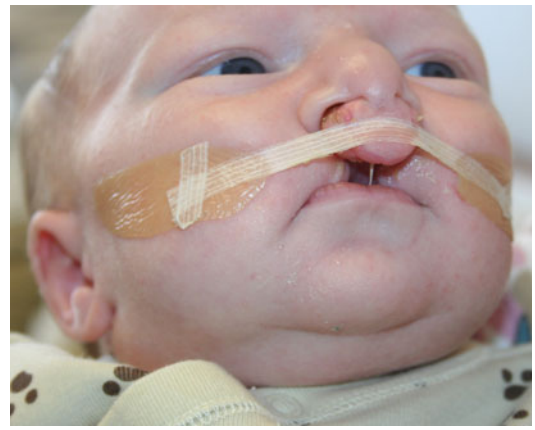


Fig. 19.4 Lip taping of prominent but centrally positioned premaxilla

19.2 Presurgical Management of the Prominent Premaxilla in Our Center

Surgeons prefer a position of the premaxilla that allows lip repair without excessive tension. In most cases in our center, molding of the lower portion and tipping of the premaxilla have proven to reduce the prominence enough to allow a good surgical outcome. In years past, we made a small acrylic cup fitted to the premaxilla. It had four wires for elastic traction to buttons on a bonnet (Fig. 19.3). Currently we use taping directly to the premaxilla as we have found this to be easier for parents to manage, and the results are similar. Lip taping to retract a prominent premaxilla that is in centric position is shown in Fig. 19.4. It is important that traction does not allow the premaxilla to tip to one side. If the premaxilla presents asymmetrically, this should be corrected. An example is shown in Fig. 19.5a–e. The taping is continued until lip surgery is done at about 10 weeks. Two to three visits with the clinic nurse is usually needed during the lip taping period. The purpose of the treatment is easily understood and the procedure easy to follow. It costs little in time and supplies, is generally not uncomfortable, and does not require arm restraints. If judged advantageous,

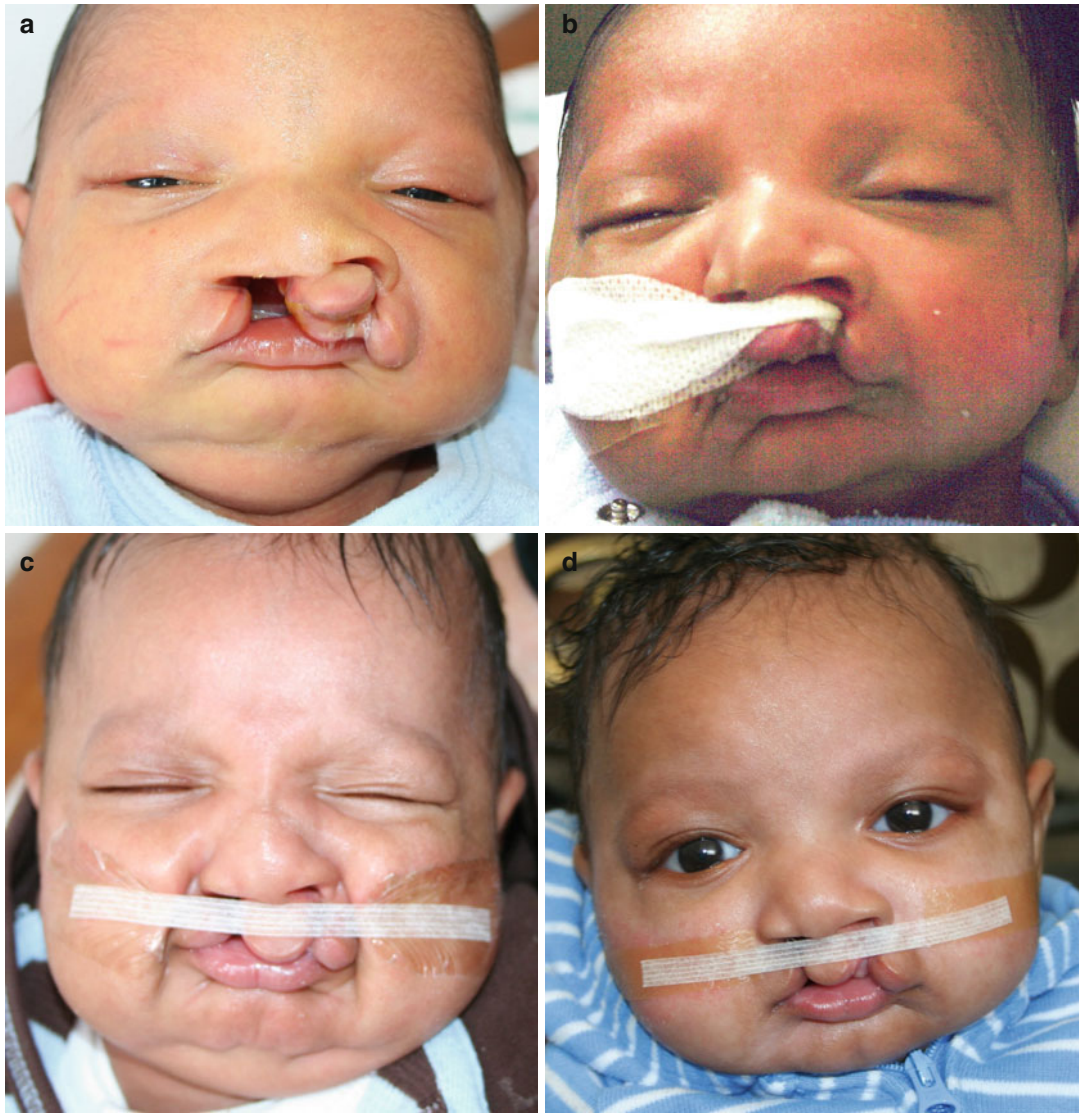


Fig. 19.5 (**a**) Newborn with asymmetric prominent premaxilla. (**b**) Bringing the premaxilla to midline with taping to one side only. (**c**) After centralizing, symmetric retraction. (**d**) Immediately before lip repair at 10 weeks

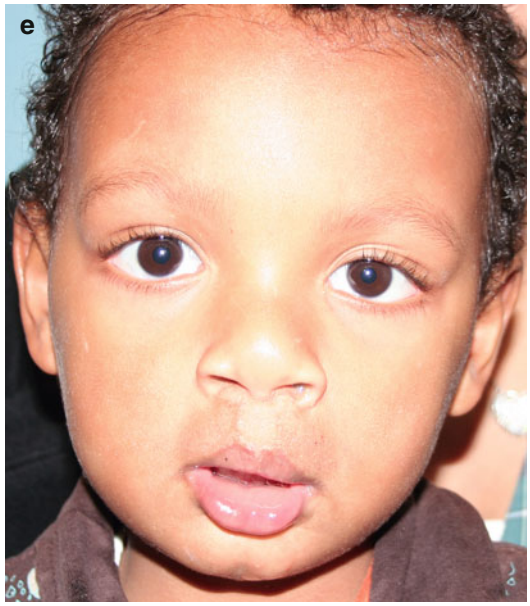


Fig. 19.5 (*continued*) (**e**) Age 2 years, lip closed at 10 weeks, palate at 11 months

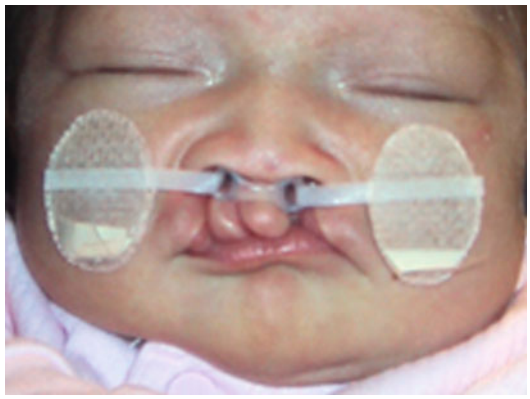


Fig. 19.6 Premaxillary retraction and nasal conformers

nasal conformers are used in combination with the taping (Fig. 19.6). If there is concern of lateral segment collapse behind the prominent premaxilla, a palatal plate is used with the lip taping.

19.3 Postsurgical Management of the Premaxilla/Maxilla in Our Center

The repaired lip will further mold the alveolar process of the premaxilla. Due to the molding of the alveolar process, the primary incisors will be retroclined and may erupt into an anterior crossbite

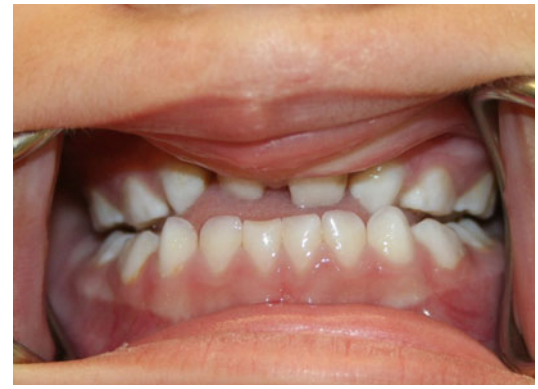


Fig. 19.7 Primary dentition with slight anterior and lateral crossbites

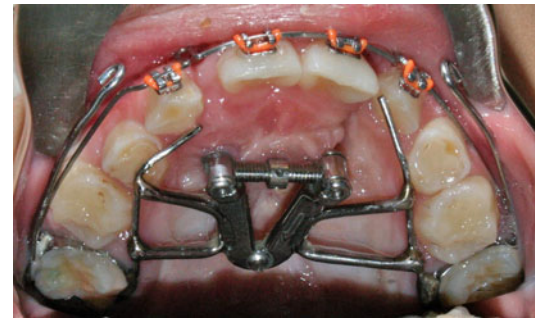


Fig. 19.8 Maxillary expansion with posteriorly hinged fixed expander, labial hooks for face mask traction

(Fig. 19.7). We generally start the orthodontic preparation for alveolar bone grafting when the 6-year molars have erupted. At that time, the permanent incisors are erupting as well. They will also be retroclined even before eruption and will often erupt into an anterior crossbite. At this time, usually around age 7, an expander is placed to correct the position of the lateral segments. The appliance may be fitted with labial arms with hooks for elastics to a face mask for maxillary protraction when needed (Fig. 19.8). This also helps to correct an anterior crossbite.

Uprighting of the incisors is easily accomplished if the upper portion of the premaxilla has remained prominent. A boy with repaired complete bilateral cleft lip and palate is shown through expansion with crossbite correction, alignment of maxillary central incisors, bone grafting, and lower first bicuspid extraction, waiting for canine eruption to start phase II orthodontic treatment (Fig. 19.9a–e). The canines are erupting forward to contact with the central

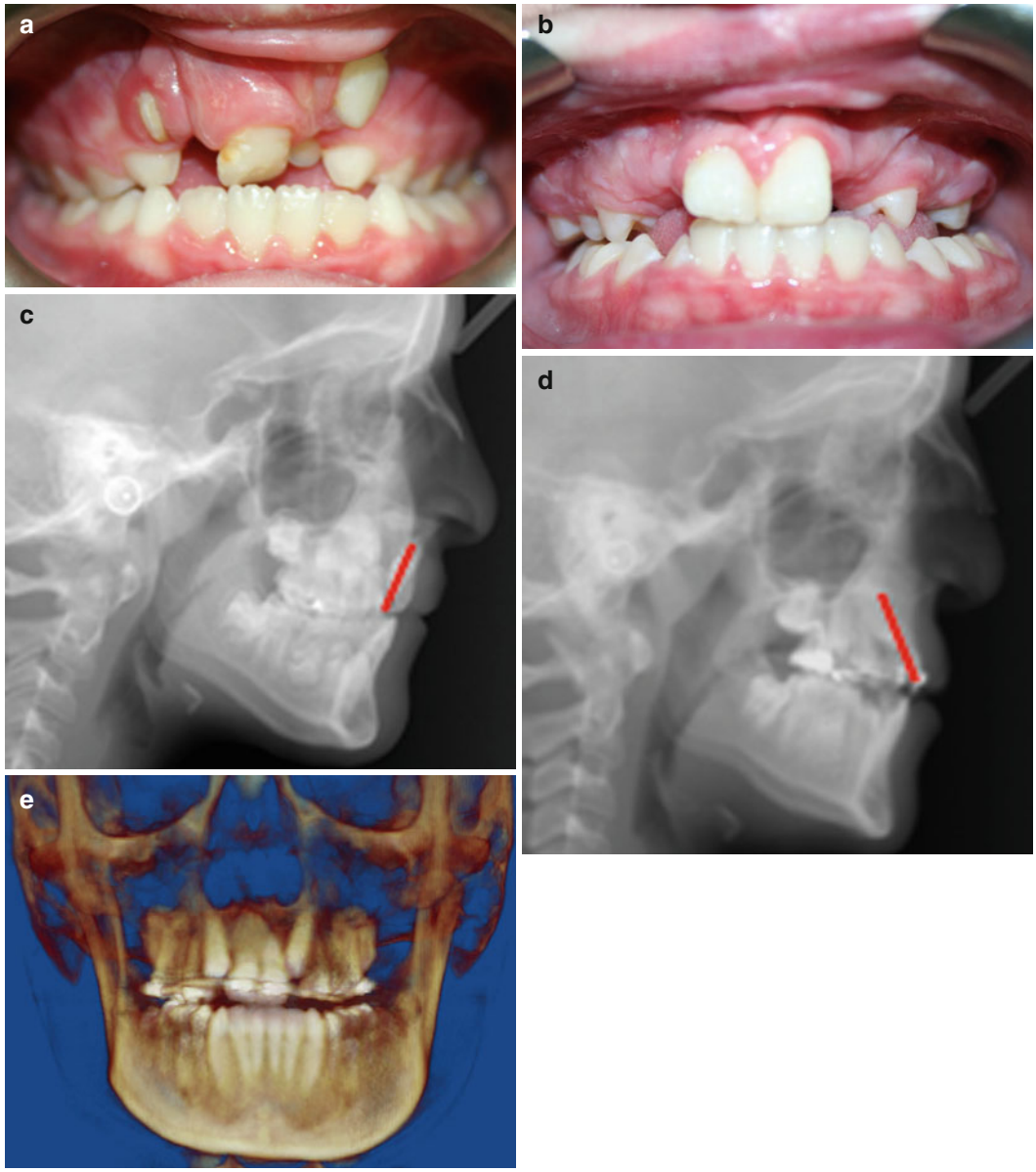


Fig. 19.9 (**a**) Mixed dentition stage, ready for maxillary expansion and central incisor crossbite correction in preparation for alveolar bone grafting. (**b**) After expansion and bilateral bone grafting. (**c**) Lateral headfilm before incisor uprighting. (**d**) Lateral headfilm after incisor correction. (**e**) Cone beam CT showing bone fill in alveolar defects and mesial eruption of canines into lateral incisor position

incisors and will be substituted for the missing lateral incisors (Oberoi et al. 2009).

We prefer to close the spaces for congenitally missing lateral incisors by canine substitution if it is possible without compromising maxillary arch length. Three additional cases are shown from infancy to adulthood and completion of treatment. Figure 19.10a–f is shown to demonstrate management of an over-extruded premaxilla. This can be a very difficult problem to correct and should be intercepted whenever possible (Vargervik 1983; Meazzini et al. 2010).

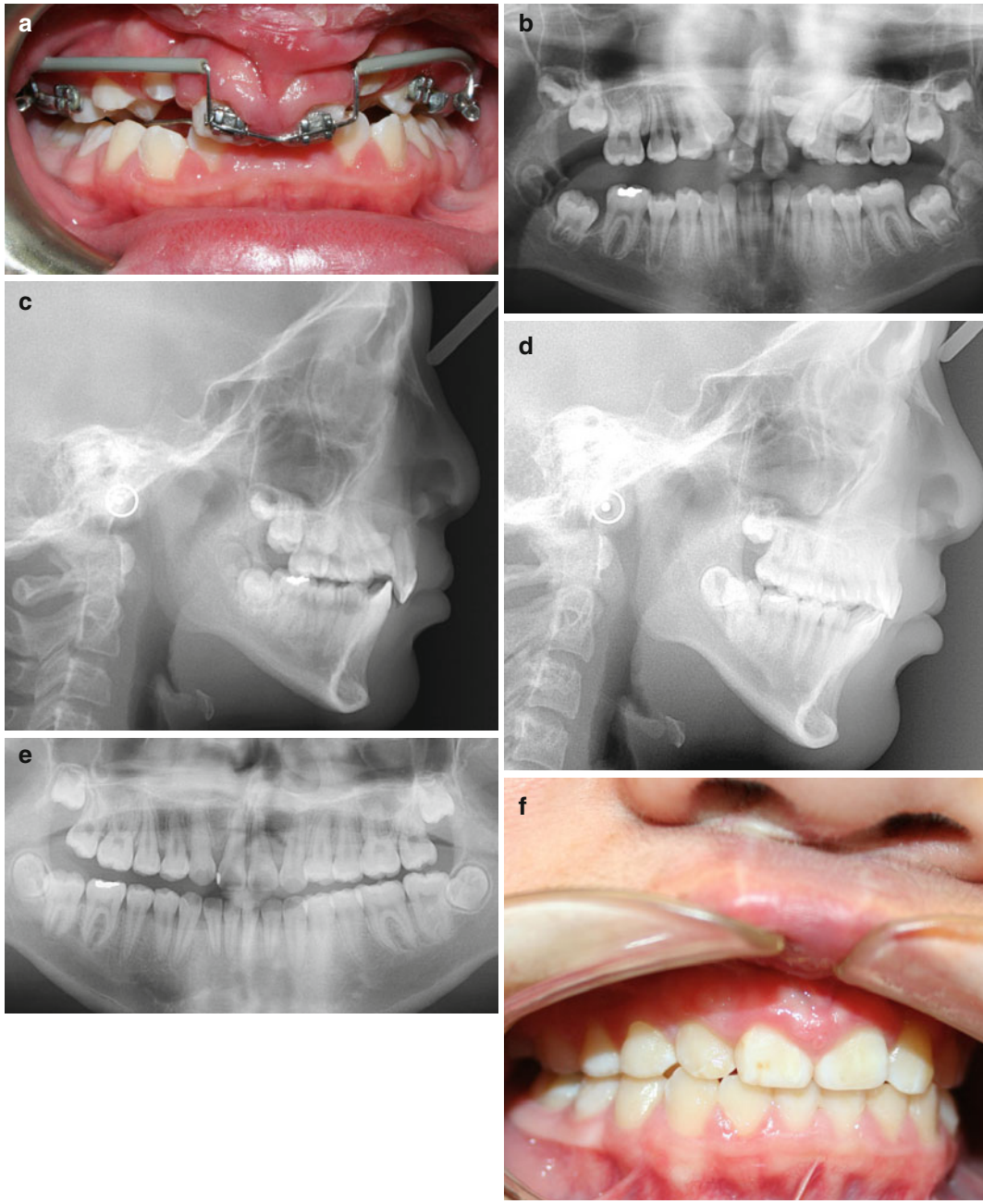


Fig. 19.10 (**a**) Over-extruded premaxilla being intruded before alveolar bone grafting. (**b**) Panorex before treatment start. (**c**) Lateral headfilm before treatment start. (**d**) Lateral headfilm after treatment completed. (**e**) Panorex after treatment completed. (**f**) Final occlusion

Figure 19.11a–e demonstrates treatment complicated by loss of one maxillary central incisor and replacement on an implant (Pena et al. 2009).

In some cases, it is clear from an early age that the maxilla will be hypoplastic to a degree that will require surgical advancement of the maxilla

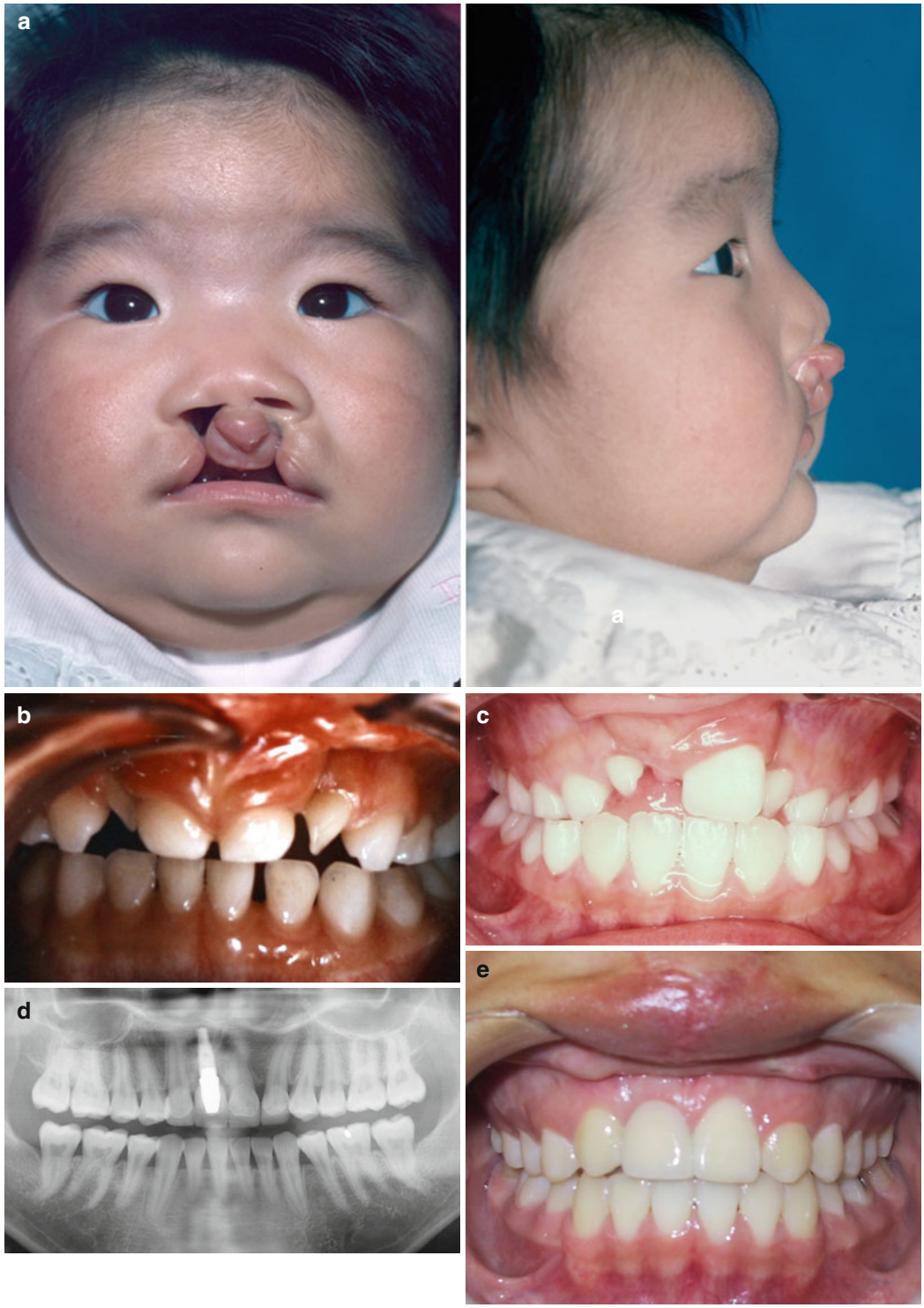
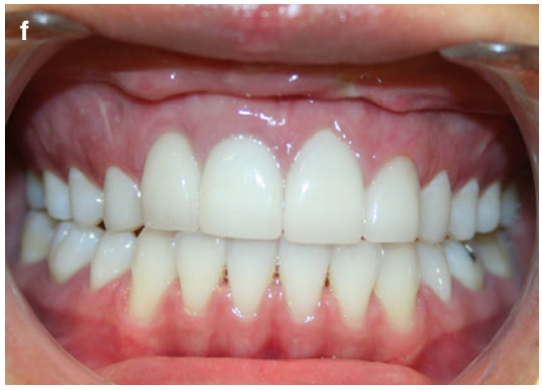
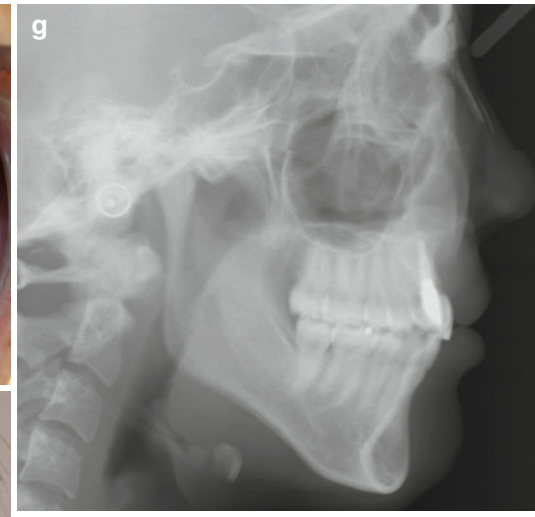


Fig. 19.11 (**a**) Infant with a very small prolabium and moderately prominent premaxilla. (**b**) Occlusion at age 6 years. (**c**) Maxillary right central incisor malformed and in ectopic position, requiring removal. (**d**) Panorex at end of treatment, age 18 years. Note implant for right central incisor. Both lateral incisors congenitally missing, canine substitution. (**e**) Occlusion at end of orthodontic treatment





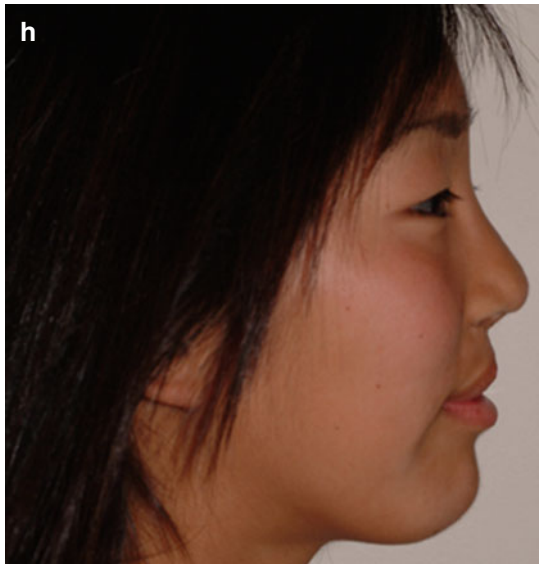


Fig. 19.11 (*continued*) (**f**) Occlusion after prosthetic treatment. (**g**) Lateral headfilm at end of treatment and growth. (**h**) Profile after completion of treatment including final lip and nose revision

at growth completion (Oberoi and Vargervik 2005; Oberoi et al. 2008). This is most often associated with multiple missing teeth and with cleft syndromes. In those cases, it may not be advantageous to correct an anterior crossbite or maintain space for missing lateral incisors as the maxilla will be advanced surgically at growth completion. Phase I orthodontic treatment should still be done to align the segments, followed by alveolar bone grafting. This will facilitate canine eruption in a mesial direction (Oberoi et al. 2010). It will also result in a one-piece maxilla at the time of Le Fort advancement. Phase II orthodontic treatment should then be postponed until 2–3 years before anticipated maxillary surgery. Such a case is shown in Fig. 19.12a–f. Le Fort advancement was done at age 19.

In examining records between 1970 and 1990, we found that 13.3 % of all nonsyndromic bilateral cleft lip and palate patients treated in that period had maxillary advancement surgery. This compares to a 25 % average reported in the literature (Vargervik et al. 2009).

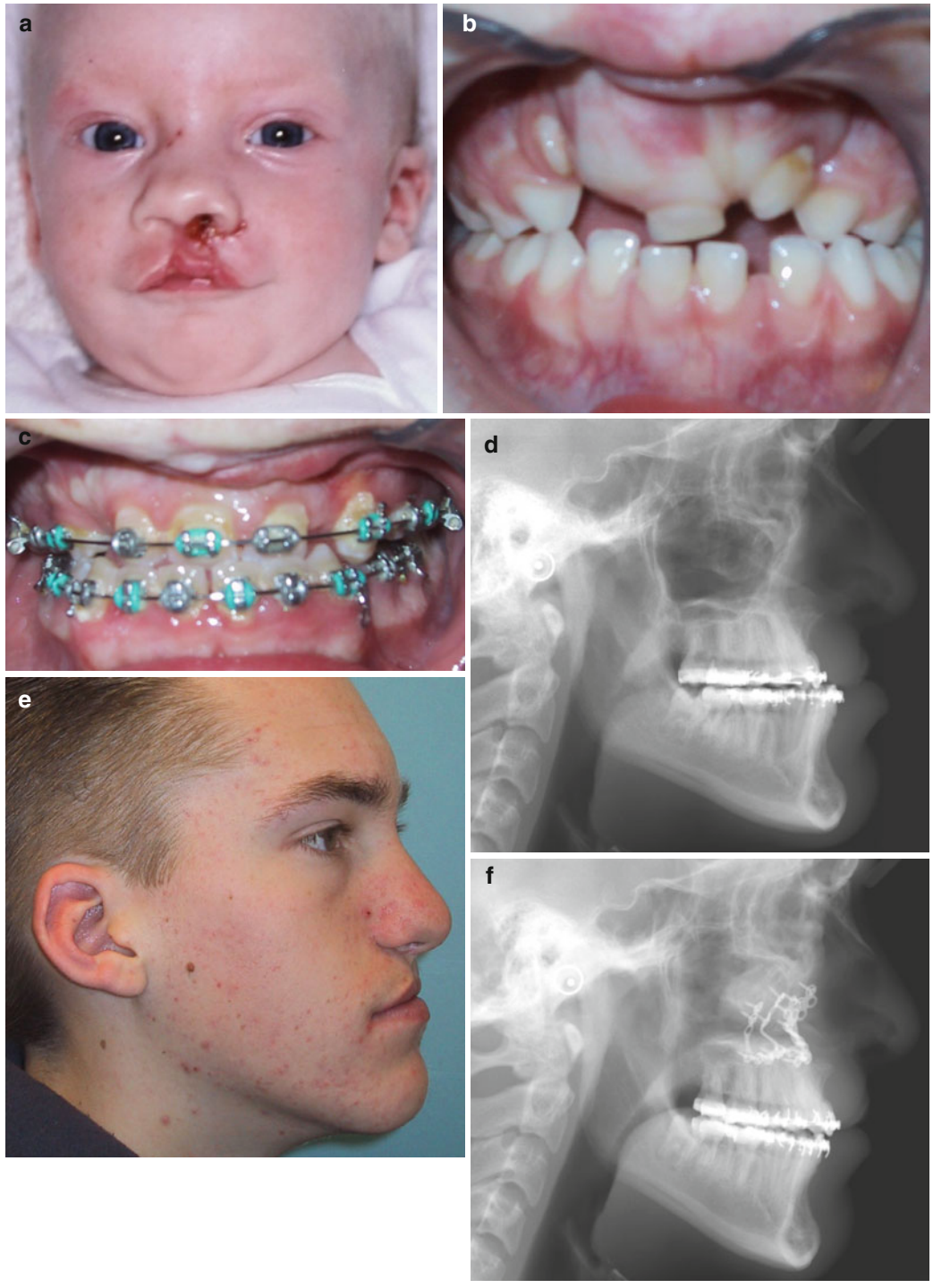
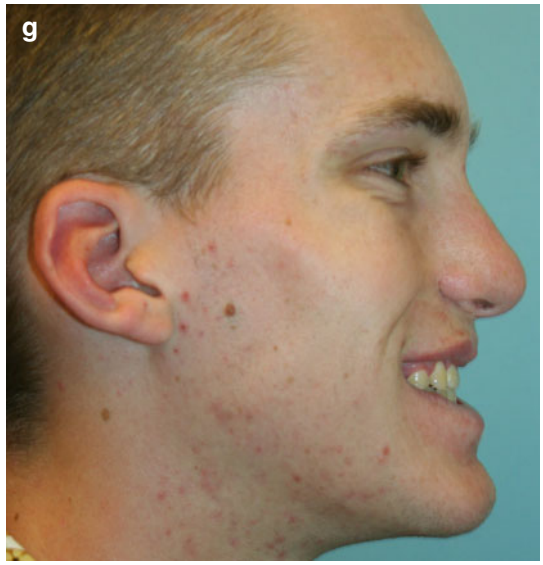


Fig. 19.12 (**a**) Infant with bilateral cleft lip and palate immediately after bilateral lip repair. (**b**) Primary dentition before maxillary expansion and bone grafting. (**c**) Phase II orthodontic treatment after bone grafting with closure of spaces for congenitally missing lateral incisors. (**d**) Lateral headfilm at completion of growth, before Le Fort I maxillary advancement. (**e**) Profile before advancement, age 17 years. (f) Lateral headfilm after maxillary advancement at age 17½ years



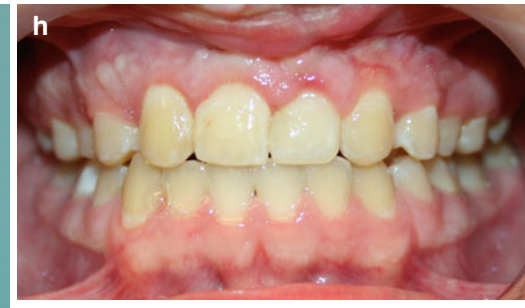


Fig. 19.12 (*continued*) (**g**) Profile 3 years after advancement. (**h**) Final occlusion showing intact alveolus and canine substitution, age 20½ years, 3 years postsurgical advancement

19.4 Maxillary Growth in Repaired Clefts

Most all individuals with a repaired unilateral or bilateral cleft demonstrate some degree of maxillary deficiency. Growth studies have yielded conflicting results. Some claim that cleft-related maxillary deficiency is an intrinsic primary defect while others maintain that it is primarily a result of surgical repair, resulting from lip or from palate repair or both. This has been presented in more detail in an earlier publication (Oberoi et al. 2008).

19.4.1 Future Direction for Growth and Treatment Studies

Superimposition of lateral cephalograms has been the standard method for quantification of changes from treatment or by growth. Differentiating dentofacial changes caused by treatment from those induced by growth is not possible with either two-dimensional or three-dimensional (3D) superimposition methods. However, comparison of treated and untreated controls by using 3D regional superimpositions using cone beam computed tomography has been shown to have the potential to assess bone displacements and bone remodeling of skeletal and soft tissue facial components relative to relatively stable structures such as the cranial base. Assessment by this method may improve our ability to quantify treatment and growth changes and lead to better differentiation between intrinsic and iatrogenic effects on growth and development.

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Part IX

Presurgical Orthopedics

Neonatal Maxillary Orthopedics: Past to Present

20

Anne Marie Kuijpers-Jagtman and Charlotte Prahl

20.1 Introduction

More than 60 years after its introduction by the Scottish prosthodontist McNeil (1950), neonatal maxillary orthopedics still remains a controversial part of the comprehensive care for cleft lip and palate patients. The therapy, also known as presurgical or early orthopedic treatment, presurgical or infant orthopedics, early maxillary orthopedics, or more recently nasoalveolar molding, was rapidly adopted by many centers around the world, although at that time there was no scientific evidence for the claimed benefits nor were the possible adverse effects properly investigated (Pruzansky 1964; Prahl-Andersen and Meijer 1979; Witzel 1990). In Europe, in the year 2000, about 54 % of the operational centers used neonatal maxillary orthopedics (Shaw et al. 2000).

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Many different types of neonatal maxillary orthopedics have been described in the literature. A wide range of appliances has been designed for this purpose with pin-retained active appliances at one end of the spectrum (Georgiade et al. 1968; Latham 1980) and passive appliances at the other (Hotz and Gnoinski 1976). Arbitrarily, they fall into three main categories: active, semi-active, and passive appliances. Active appliances are constructed to apply a force to the maxillary segments to move them into the desired direction by using an active force delivery system like springs and screws. Additional anchorage can be obtained by pins that are driven into the maxillary bone holding the plate in position (Harkins 1960; Georgiade 1964). Semi-active appliances are constructed by sectioning the dental cast and reorienting the maxillary segments in a more favorable position. The plate is made on the reconstructed cast and will force the palatal segments into the predetermined direction, when placed in the oral cavity. External strapping across the cleft can be part of the treatment protocol. These are the McNeil (1950) and Burston (1959) type of appliances. Passive appliances that are combined with extraoral strapping also fit into this category (Huddart 1967). Passive appliances are supposed to induce arch alignment during growth by grinding away material in definitive areas of the plate to ensure a proper spontaneous development of the segments. The plate is held in position by suction and adhesion only, and no extraoral strapping is applied. The so-called Zürich approach as proposed by Hotz (1969) and

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Hotz and Gnoinski (1976) is the most well-known representative of this kind of neonatal maxillary orthopedics.

Over the years, neonatal maxillary orthopedics has given rise to emotional debates between advocates and opponents of the procedure. At the annual meeting of the *American Cleft Palate-Craniofacial Association* held in St. Louis in 1990, the thesis "Presurgical orthopedics is by and large a waste of time" was discussed during the so-called Great Debate. During the *8th International Congress on Cleft Palate and Craniofacial Anomalies* in Singapore in 1997, a comparable debate took place, discussing the statement "Is presurgical infant orthopedics more a luxury than a necessity?" Before and after the debates, the audiences voted equally in favor or against the statements. It was considered a major problem that hardly any sound research findings were available regarding the effects of neonatal maxillary orthopedics. And even nearly 50 years after publication of one of the most talked about critical reviews on presurgical orthopedics and primary bone grafting by Samuel Pruzansky from Chicago, what he stated still holds generally true: "The procedures advocated might be defended on the basis that continuous exploration for new and better methods is warranted and deserves support. While such research may be justified, despite the costs incurred, it is incumbent on the investigator to document his results in a scientific manner. Instead, we have been fed opinion, anecdotal pap, wishful thinking, and empirical trivia" (Pruzansky 1964).

20.2 Early History of Neonatal Maxillary Orthopedics

Before the modern school of neonatal maxillary orthopedics began, facial binding or adhesive tape strapping was used centuries ago to narrow the cleft before surgery (Winters and Hurwitz 1995). A variation on these external bindings that were all used to reduce the cleft width was the method of Brophy (1927). He wired both ends of the cleft alveolus together in order to reduce the cleft by tightening the wire, after which lip surgery could be performed. All procedures, which were at that time mainly done by prosthodontists or by the surgeons themselves, were based on the never proven assumption that a narrow, well-aligned cleft would be easier to repair with less undermining and less mobilization of soft tissues. A narrower cleft would also lead to less tension in the repaired lip. These claims have been repeated over and over again since McNeil started to advocate presurgical orthopedics half a century ago (McNeil 1950, 1954, 1956), although he used another argument to perform this treatment. He adopted the theory of the anatomist Scott (1956, 1958), who suggested that the palatal segments being detached from the downward and forward growth of the cartilaginous nasal septum, the so-called growth center, remained deficient and retruded in the face. In his opinion, this resulted in the midfacial deficiency often observed in cleft lip and palate patients. By molding the palatal segments into the anatomical correct position by using a series of acrylic plates, McNeil believed that this would produce a normal maxilla while reducing the alveolar and palatal cleft at the same time.

Much confusion with respect to the effect has arisen, because the morphology of the maxilla of newborn children with clefts varies from collapsed maxillae to wide open clefts of more than 15 mm. Obviously, different treatment mechanics have to be used in these cases. Burston (1959, 1965, 1971) advocated a passive plate to start with to facilitate feeding and letting the child adapt to the plate. A following plate should be constructed by sectioning and aligning the segments of the maxilla on the plaster models before constructing the acrylic plate. These plates were used with external strapping. The plates were fitted with "wings," the chief purpose of which was to enable the parents to insert and remove the plates and not as stated later to aid retention. McNeil and Burston claimed that soft tissues overlying the hard palate were stimulated to grow and that neonatal maxillary orthopedics could control and modify the postnatal development of the maxilla. These two practitioners have had a tremendous impact on the early treatment of

children with clefts and led the way for the idea of bone grafting in conjunction with early orthopedic treatment (see also Sect. 20.4.1).

20.3 Claimed Benefits of Neonatal Maxillary Orthopedics

There has been consensus since the early 1950s that a multidisciplinary team within a center can best treat children with cleft lip and palate. The team should function as an organization with a general policy for the treatment, and each member of the team should have insight into the different aspects of treatment. The orthodontic discipline has been part of such organizations from the start, because in the majority of cases, dental arch distortion and a constricted maxilla were thought to be the result of the surgical reconstruction of the lip. Neonatal maxillary orthopedics aims at securing a good maxillary arch form in acceptable relationship with the mandible and to restore normal oral function. McNeil (1950, 1954, 1956) and Burston (1959, 1965, 1971) claimed that the deformation seen at birth was partly due to the original lack of tissue and a further complication of growing tissues having become mal-related consequent to cleft formation. McNeil (1954) stated that the most important aspect of preoperative treatment of cleft palate infants was control and correction of the lateral segments in unilateral and bilateral conditions and that the effect of bone stimulation plays an important role in this correction. Forces which are within the limits of biological tolerance are said to stimulate bone growth if they are applied to particular regions and in such a direction that they can be regarded as intensified normal forces.

The attainment of an end-to-end position of the alveolar processes before lip operation was the ultimate goal for neonatal orthopedics performed by all dental practitioners in the past. It has been shown that it is possible to narrow the cleft and to achieve an anatomical correct position of the maxillary segments at the time of lip surgery (McNeil 1954; Burston 1959; Rosenstein 1969; Robertson 1971; Brogan and McComb 1973; Huddart 1974; O'Donnell et al. 1974; Hotz and Gnoinski 1976; Huddart 1979; Prahl et al. 2001; Yamada et al. 2003). It also improves the angulation of the palatal shelves to a more horizontal position (Huddart 1987; Hochban and Austermann 1989; Mishima et al. 2000; Mishima et al. 2001). However, the question remains of whether these short-term effects prior to primary surgery have any beneficial effect on overall treatment outcome in the long term.

Although neonatal maxillary orthopedics originally was instituted to restore the normal anatomy and to guide growth and development of the maxillary segments, only after the introduction of presurgical orthopedics did the orthodontic discipline try to justify early intervention for other reasons. Prahl et al. (2001) summarized the arguments of the proponents of the use of infant orthopedics who state that this approach allows a more normalized pattern of deglutition, prevents twisting and dorsal position of the tongue in the cleft, improves arch form and position of the alar base, facilitates surgery, and improves outcome in general. Other alleged benefits that became "en vogue" are reduction of posterior cleft width, prevention of initial collapse after surgery, prevention of crossbites, straightening of the nasal septum, facilitation of feeding, less danger of aspiration, better speech development, better nose breathing, better middle ear conditions, less extensive orthodontic treatment at later ages, and a positive psychological effect on the parents (Hotz and Gnoinski 1976, 1979; Dorf et al. 1985; Oblak 1986; Weil 1987; Gnoinski 1990; Ball et al. 1995; Mishima et al. 1996a, b; Kozelj 1999; Reid 2004). The opponents of neonatal maxillary orthopedics stated that neonatal maxillary orthopedics is a complex and expensive therapy that is ineffective and unnecessary (Pruzansky 1964; Prahl-Andersen and Meijer 1979; Pruzansky and Aduss 1964). Parents are obliged to travel frequently to the treatment center and are given the burden of compliance (Prahl et al. 2001). Furthermore, it is stated that neonatal maxillary orthopedics restricts maxillary development (Pruzansky 1964; Kramer et al. 1992) and influences speech negatively due to delayed surgery of the hard palate inherent to neonatal maxillary orthopedics (Bardach et al. 1984; Witzel et al. 1984).

Unfortunately, we have to conclude that despite more than half a century of research into neonatal maxillary orthopedics, the results of these studies remain inconclusive, mainly due to shortcomings in the research design. Most studies were nonrandomized, retrospective in nature, have a small sample size, and lack a (randomized) control group or used historical controls. Furthermore, treatment protocols are often not properly described, and the competence of the professionals who performed the treatment is not always well documented. Also, frequently, no clear outcome measures are given, and confounding variables are not taken into account.

20.4 Specific Types of Infant Orthopedics

20.4.1 Kernahan Rosenstein Procedure

An obvious problem after realignment of the maxillary segments by either neonatal orthopedics or lip surgery alone is of course to hold the realigned segments in position. For this purpose, bone grafting, as a primary procedure at or around the time of lip repair, was first utilized in the late 1950s in Europe (Nordin and Johansson 1955; Schmid 1955). Great emphasis was placed on the importance of this procedure in stabilizing the realigned maxillary segments (Robertson 1983). In 1977, Berkowitz concluded in his state-of-the art report that the benefits of presurgical orthopedic therapy (PSOT) were not proven and that primary bone grafting was detrimental to midfacial growth (Berkowitz 1977). At that time, many clinicians who previously advocated primary bone grafting abandoned the procedure because of its negative effect on maxillary and midfacial growth (Jolleys and Robertson 1972; Robertson 1973; Friede and Johanson 1974). After 1975, several additional centers confirmed midfacial growth inhibition and discontinued the use of primary bone grafting (Friede and Johanson 1982; Pfeifer 1986; Reichert and Manzari 1990; Lilja et al. 1996; Smahel et al. 1998; Russell et al. 2011).

Over the years, a few centers continued to use neonatal maxillary orthopedics in combination with primary bone grafting. One of these is the Children's Memorial Hospital Cleft Palate Clinic, Chicago (Dado 1990). The Rosenstein appliance is a passive plate that is inserted prior to lip surgery. Then, the lip is closed and the arch segments are molded until they are in butt alignment, after which the segments are stabilized by a small subperiosteal onlay rib graft. The plate is retained for 6–8 weeks postgraft, and the palate is usually closed at or before 12 months of age (Kernahan and Rosenstein 1990; Rosenstein 2003). In complete bilateral cleft lip and palate, the appliance covers the lateral segments, holding them in position while an extraoral elastic band and later on the restored lip molds the premaxilla backwards.

Since 1965, Kernahan and Rosenstein have used this approach, claiming that there is a principal difference with other approaches both in sequence and technique of the procedures and that this is critical to its success (Kernahan and Rosenstein 1990). In 2003, Rosenstein and coworkers reported the long-term results in a sample of 135 patients, or about 50 % of the cases that were seen in their clinic since 1965 (Rosenstein et al. 2003). In their paper, they give an overview of the comparative studies with other centers that have been performed. These studies show that the growth in the Chicago sample was as good as for any other sample treated without primary bone grafting. However, evaluation bias cannot be ruled out as there is an attrition rate of 50 % of the original sample. In his multicenter study, Ross (1987), and later on Trotman et al. (1996), found less favorable results showing that patients who underwent primary bone grafting on average had less protrusive maxillae than the nongrafted sample, while the mandible compensated for this by downward and backward rotation, resulting in an increase of lower anterior facial height.

The recently published Americleft study (Russell et al. 2011) throws some new light on the results of this procedure. In this retrospective audit of five North American cleft centers, there was only one center that employed primary alveolar bone grafting with infant orthopedics as well as secondary surgical revisions completed prior

to 8 years of age. The outcome of this center was less favorable than centers using much simpler and less burdensome protocols. Although it is not possible to detect a cause-effect relationship from a nonrandomized retrospective study, Center B in the Americleft study has discontinued use of primary bone grafting in its infant management protocol, due to the failure to detect any measurable benefits and the possibility of detrimental effects on maxillary development.

20.4.2 Latham-Millard Pinned Appliance

In the Millard-Latham method of neonatal maxillary orthopedics, forces are applied using a pinned palatal appliance to manipulate mechanically the maxillary segments into close approximation, followed by alveoloperiosteoplasty and lip adhesion (Millard and Latham 1990) Latham based his treatment concept on the facial growth hypothesis of Scott (1956, 1958) as mentioned earlier, and he encouraged Millard to use it in complete unilateral and bilateral clefts. The aim of the procedure is "to carry the interrupted embryonic process to normal completion" by maxillary alignment, stabilization of the alignment along with tunneling of the alveolar cleft with periosteum, and reconstruction of the nasal floor to support the alar base (Millard 1994).

It was many years before the first longer-term studies were published about this procedure. Berkowitz (1996a) found that of a group of 32 UCLP patients at 6 years of age, 23 cases had an anterior cross bite due to the displacement of the premaxillary portion of the larger maxillary segment and the scar tissue resulting from the periosteoplasty. Comparable results were found analyzing dental casts of 63 patients with complete clefts showing a greater percentage of anterior crossbites at 6 years but becoming less at 9 years of age compared to a lip adhesion alone group (Millard et al. 1999). Lukash et al. (1998) also found anterior and lateral crossbites after 6 years of age in most UCLP cases, but in BCLP cases, arch form and occlusion as well as midfacial growth were considered acceptable. However, they did not present data or statistical analysis. The findings in BCLP patients are in contrast to the data from Berkowitz (1996a, b), who showed in a BCLP group of 14 patients treated by Millard and Latham that all patients had midfacial retrusion at 9 years of age and in 50 % of the cases the premaxilla was retruded with the anterior teeth in crossbite.

In 2004, Berkowitz and coworkers (2004) published an extensive report on their findings in 30 UCLP patients and 21 BCLP patients treated with the Millard-Latham procedure at the South Florida Cleft Palate Clinic. These patients were compared to patients who were treated without neonatal maxillary orthopedics and alveoloperiosteoplasty. Both in complete UCLP and BCLP, they found a higher frequency of anterior crossbite and (except at 3 and 12 years of age) also for buccal crossbite compared to the treatment alternative without neonatal orthopedics and alveoloplasty. The only other long-term follow-up study found in the literature reports more anterior open bites and posterior crossbites in 55 UCLP and BCLP patients treated with the procedure compared to a control group without the procedure (Henkel and Gundlach 1997).

In analyzing the results of the published studies on the Latham-Millard procedure, the problem is that besides neonatal maxillary orthopedics, infant periosteoplasty is always performed as well as a specific sequence of related operations. For example, in a study comparing two groups that had the same Latham infant orthopedics procedure but a different timing of lip and palate repair as well as another technique of hard palate closure, better results were found in the group with the modified surgical protocol (Latham 2007). With respect to infant perioplasty, it must be emphasized that their technique of periosteoplasty is more limited with less undermining of the periosteum on the maxilla than the original one, described by Skoog (1965). However, infant periosteoplasty seems not to be widely performed in most European centers anymore, as its expectations have not been fulfilled (Hellquist 1982; Hellquist et al. 1983; Hellquist and Svärdström 1986; Rintala and Ranta 1986; Fara et al. 1990; Smahel and Müllerova 1994). Nevertheless, in the United States, the Millard-Latham method

continues to attract attention despite the fact that its benefits are questionable (Kuijpers-Jagtman and Long 2000). Based on the long-term results reported by Henkel and Gundlach (1997) and Berkowitz et al. (2004), we conclude that there is reason to be at least suspicious of the Millard-Latham procedure or to even abandon it.

20.4.3 The Zürich Approach

In most other clinics in Europe, neonatal maxillary orthopedics started in Zürich in the mid-1950s based on the treatment principles of McNeil. After the first long-term evaluation, it became clear that forced approximation of the maxillary segments was not advisable. Consequently, the procedure was greatly modified to what is known now as the "Zürich approach" (Hotz and Gnoinski 1976; Hotz et al. 1986). According to Hotz and Gnoinski, the primary aim of neonatal orthopedics is not to facilitate surgery or to stimulate growth as postulated by McNeil, but to take advantage of intrinsic developmental potentials. Since 1969/1970, early maxillary orthopedic treatment was essential in Zürich, while surgical intervention was postponed in order to minimize subsequent growth disturbance, create optimal conditions for the maxillary segments to develop their entire growth potential, maintain or improve arch form, and control effects of surgical lip closure. The appliance used is a passive plate of compound soft and hard acrylic resin, and it is worn 24 h a day for about 16–18 months, when the soft palate is closed surgically. The hard palate is closed after 5 years of age (Hotz 1969; Hotz and Gnoinski 1976). During the course of treatment, the lip is closed at about 6 months of age. The posterior extension of the plate that extends to the uvula must be carefully adapted to the specific anatomy of the patient. Arch alignment is achieved by grinding away the acrylic in specific areas. Figure 20.1 shows a patient who was treated according to these principles.

According to the scientific use at that time, conclusions regarding the effects of treatment were mainly based on observations: orthopedic guidance together with optimal timing of surgery has beneficial effects. Later evaluations claimed better results with two-stage palatal closure on speech; whether or not the presurgical treatment had an influence could not be demonstrated (Hotz and Gnoinski 1976, 1979; Gnoinski 1982; Van Demark et al. 1989). In 2009, longitudinal cephalometric results were published of a sample of 29 patients with BCLP from 5 years to the end of the growth period (Gnoinski and Rutz 2009). All 29 patients had been treated in Zürich according to the same protocol, operated on by the same surgeon and treated by the same orthodontist. The authors conclude that the multidisciplinary concept of maintaining the initially protrusive position of the premaxilla by means of a passive plate at the newborn and infant stage, as well as using surgical procedures with limited retrusive effect, proved to be correct in the long run: at the young adult stage, the ANB angle remained positive for almost all patients except for those with multiple-tooth agenesis in the upper arch.

We can conclude that the treatment principles of Dr. Hotz and Dr. Gnoinski have had a tremendous impact on cleft palate treatment, especially in Europe. Unfortunately, in the last two decades, not much has been published about the results of the Zürich approach, and the earlier papers have no strict research design that allows us to draw evidence-based conclusions about the effectiveness of neonatal orthopedics. It might very well be possible that the surgical timing and sequencing are the decisive factors in the final treatment result.

20.4.4 Nasoalveolar Molding Grayson

After nearly 40 years of neonatal maxillary orthopedics, the treatment started to drift away from its traditional aims and techniques with the publications from the cleft palate team at New York University Medical Center in the 1990s (Grayson et al. 1993; Cutting et al. 1998). To theirs and many others' experience, it is very difficult to reach a good result in the neonatal treatment of cleft lip and palate due to the actual anatomical deformity of the nose, which includes abnormal nasal cartilage morphology, deviated nasal septum

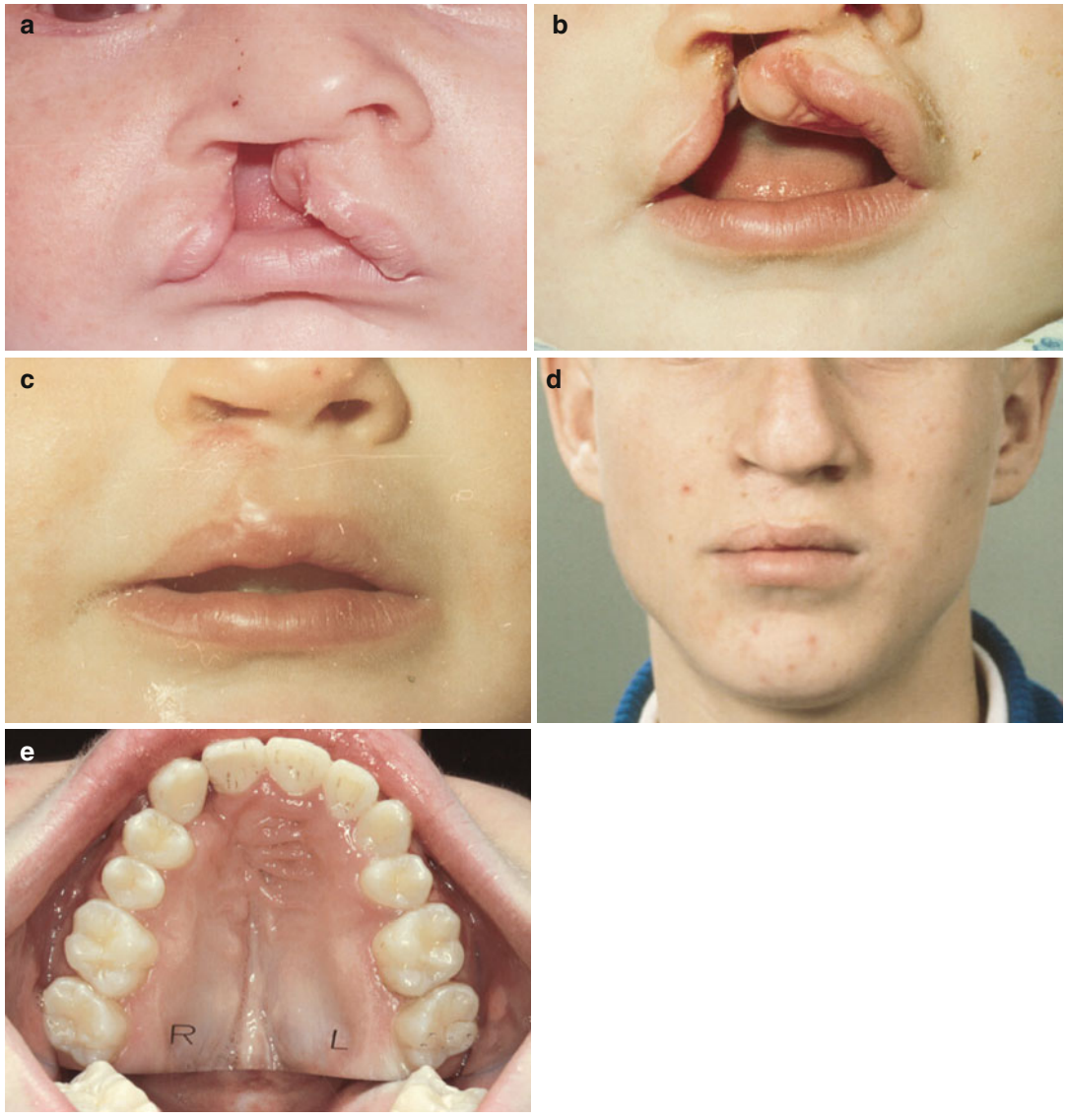

Fig. 20.1 (**a**–**e**) Patient with a complete unilateral cleft lip and palate at birth (**a**), after presurgical orthopedic treatment according to the Zurich approach (**b**), after lip closure (**c**), frontal view at the age of 15.8 years (**d**), and palatal view at 15.8 years after secondary bone grafting and orthodontic treatment (**e**)

and columella, asymmetry of the alar base, and a short or even absent columella, depending on the type of cleft. In addition, fibers of the orbicularis oris muscle run superiorly along the margins of the cleft towards the nasal tip, while in bilateral clefts, muscle tissue is often lacking in the prolabium, which is seated directly at the short columella. Therefore, Grayson and Cutting (2001), who have used the Latham-Millard (POPLA) procedure for 15 years but did not note why it was abandoned, emphasize the importance of presurgical correction of the nasal cartilage and soft tissue deformity, which can be achieved by a combination of nasal and alveolar orthopedic molding. This therapy, now called NAM (nasoalveolar molding) or PNAM (presurgical nasoalveolar molding), makes a distinct difference with the other types of infant orthopedics, as described

previously (Grayson et al. 1993; Cutting et al. 1998; Grayson and Cutting 2001; Barillas et al. 2009). In NAM, nasal stents are added to the alveolar molding plate. The molding plate itself is mainly used to approximate the alveolar segments and retract the protruding premaxilla in CBCLP in order to reduce the nasal deformity to a degree that enables the start of more precise nasal molding with stents (Grayson and Maull 2006; Santiago and Grayson 2009). Retention of the appliance in the mouth is secured by tapes on the cheeks, which engage the intraoral plate with orthodontic rubber bands. The appliance aims to improve nasal tip projection, and septal and lower lateral cartilage position, before cleft repair. In bilateral cleft lip and palate, the nasal stents are projected to gradually lengthen the deficient columella. A longitudinal study in BCLP patients on nasal morphology after NAM and primary alveolar surgical repair (gingivoperiosteoplasty) showed that near normal nasoalveolar morphology was reached until 12.5 years of age compared to reference values from the literature (Garfinkle et al. 2011). However, this paper does not discuss the effects on the facial profile and anterior occlusion.

The use of NAM is controversial because of reported midfacial recessiveness by other NAM users and polarizing professionals in the field of cleft lip and palate as happened before concerning the use of regular infant orthopedics. The claimed benefits of the procedure are (a) improved long-term nasal esthetics, (b) a reduced number of nasal surgical procedures, (c) a reduced need for secondary bone grafts if gingivoperiosteoplasty (GPP) is applied, (d) no larger growth disturbance than is found for other well-established procedures, and (e) savings in cost. In a series of papers that are discussed in their 2001 paper, the New York group attempted to substantiate these claims (Grayson and Cutting 2001). On the other hand, Berkowitz (2009) argues that NAM+GPP compromises future facial growth. This was confirmed in a retrospective study showing that midfacial growth was affected more negatively in a group with NAM+GPP than in a NAM non-GPP group measured at the age of 5 (Hsieh et al. 2010). Whether there was a negative effect by NAM alone could not be ruled out. Opponents also suggest that NAM places an extra burden on the family that already must adapt to having a newborn with a cleft (Sischo et al. 2012).

However, as for all other types of neonatal maxillary orthopedics, the research design for studies into the effect of NAM was not adequate to show scientific evidence for these claims so far. In line with the DUTCHCLEFT trial as described below, a two-group randomized controlled clinical trial would be the preferred design to investigate the effect of NAM before deciding whether to accept or abandon this therapy. Santiago and Grayson (2009) recognize this as they state that "Although these benefits have been demonstrated in multiple clinical publications there is no doubt that a need for long-term and perhaps federally supported clinical trials exists." Nevertheless, in the USA, NAM is now employed in 37 % of 117 teams that have a case load of more than 25 patients annually (Sischo et al. 2012).

20.5 The DUTCHCLEFT Study

20.5.1 Background

As noted above, neonatal maxillary orthopedics was developed and introduced on theoretical grounds, and it became part of the treatment protocol in many centers although the actual effectiveness had never been tested nor were the possible adverse effects properly looked into. In Nijmegen, we have performed neonatal infant orthopedics according to the Zürich approach for over 20 years, but our clinical experience has convinced us that in unilateral cleft lip and palate patients, neonatal orthopedics is at best not necessary and at its worst could have negative side effects, even if considering only the cost-effectiveness. Gradually, we reached equipoise regarding whether or not to perform neonatal infant orthopedics. Being strong supporters of evidence-based care in general, and for cleft lip and palate patients in particular, a randomized controlled clinical trial was a next logical and inevitable step to try to solve the controversy regarding neonatal orthopedics.

After obtaining funding for a first period of 3 years, a prospective randomized clinical trial to

investigate the effect of neonatal maxillary orthopedics in children with a complete UCLP, named "DUTCHCLEFT," was begun in 1993 in three academic Cleft Palate Centers in the Netherlands: the Cleft Palate Centers of Radboud University Nijmegen Medical Center, Free University Medical Centre Amsterdam, and Erasmus Medical Centre in Rotterdam.

The following outcome variables were studied:

(A) General effects: influence on feeding, body length and weight gain, parents' satisfaction
(B) Surgical and orthodontic effects: duration of lip surgery, esthetic outcome, maxillary arch form and dimensions, maxillofacial growth
(C) Speech and language development: prelingual sound production, early speech and language development, intelligibility
(D) Cost-effectiveness: medical and nonmedical costs

Results for feeding, general body growth, maxillary arch dimensions, facial growth, nasolabial appearance, speech and language development, and cost-effectiveness have been published in a series of papers between 1998 and 2009 and will be discussed below.

20.5.2 Experimental Design

The study was set up as a prospective two-arm randomized controlled clinical trial in three academic cleft palate centers in the Netherlands. The Institutional Review Boards of the three hospitals approved the study protocol. An extensive description of the design of the study is given by Prahl et al. (2001).

The intake started in January 1993 and ended in June 1996. In total, 54 babies (41 boys, 13 girls) entered the trial, 27 in each group. The patient inclusion criteria were complete UCLP, infants born at term, both parents Caucasian and, because of the speech assessments, fluent in the Dutch language, and trial entrance within 2 weeks after birth. The exclusion criteria were other congenital malformations (except for syndactyly) and soft tissue bands. The parents of eligible infants were informed about the trial, and written informed consent was obtained from all participants. A computerized balanced allocation method was used in order to reduce imbalance on relevant prognostic factors between the two groups. Patients were allocated based on birth weight (<3,300 or >3,300 g) and alveolar cleft width (<8 mm, between 8 and 12 mm, or >12 mm). Treatment allocation was concealed. A computer program assigned the infant to the group with infant orthopedics (= IO^+) or without infant orthopedics (= IO^-).

Lip surgery was performed at 18 weeks of age according to the Millard technique. The soft palate was closed at the age of 52 weeks according to a modified von Langenbeck procedure. Neonatal maxillary orthopedics was performed by means of passive plates, starting within 2 weeks after birth. The plate was fabricated on a plaster cast and consisted of compound soft and hard acrylic. The plate, with a small extension into the cleft nose, covered the palate and the alveolar ridges and obtruded the cleft of the hard and soft palate. The plate was worn 24 h a day. IO^+ children returned to the clinic every 3 weeks to have their plates adjusted by grinding at the cleft margins to ensure proper approximation of the maxillary segments. Maxillary growth indicated the necessity for a new plate. After surgical lip closure, the plate was relieved in the frontal area and reinserted the same day. Checkup visits were now planned every 4–6 weeks. The plate was worn until surgical closure of the soft palate. Children in the IO^- group did not wear plates. They visited the clinic for an extra checkup at the age of 6 weeks as well as before and after lip repair and soft palate closure.

20.5.3 General Effects

Available information in the literature of the effects of IO on *feeding* and subsequently on *general body growth* seemed to be inadequate, and therefore, feeding variables and the anthropometric variables weight and length were measured in DUTCHCLEFT. The feeding variables were measured using questionnaires that were given to the parents when the child was 0–2 weeks of age and at 3, 6, 15, and 24 weeks of age. The questions concerned: amount of food, duration of the feeding, feeding frequency, feeding velocity (time measurement), feeding method, breast milk

or formula, and feeding problems. We also used questionnaires to measure the mother's *satisfaction in motherhood* (Prahl et al. 2008).

Mother's satisfaction was comparable for both groups. Only slight feeding problems were experienced, but there were no significant differences between babies who wore a plate and babies without a plate. Feeding velocity increased with time from 2.9 to 13.2 ml/min in the IO^- group and from 2.6 to 13.8 ml/min in the IO^+ group; no significant differences were found between groups. Weight-for-age, length-for-age, and weight-for-length (*z*-scores) did not differ significantly between groups, but overall the UCLP infants in both groups had significantly lower mean *z*-scores for weight-for-age and height-for-age than the reference during the first 14 months of life and had lower mean values for weight-for-length after soft palate closure (Prahl et al. 2005).

The data show that neonatal orthopedics with the aim to improve feeding and the consequent nutritional status in infants with UCLP can be abandoned. In addition, parents' satisfaction proved not to be an argument to perform neonatal orthopedics.

20.5.4 Surgical and Orthodontic Effects

The purpose of this part of the DUTCHCLEFT trial was to evaluate the effect of neonatal orthopedics on maxillary arch form and arch dimensions from birth on and occlusion of the deciduous dentition in UCLP children, to evaluate cephalometric facial growth, and to rate facial and nasolabial appearance.

Maxillary arch dimensions were evaluated by means of plaster casts of the upper jaw at the ages of 0, 15, 24, 48, and 58 weeks and 1–1/2, 4, and 6 years. The maxillary casts were analyzed three-dimensionally by means of the Reflex Microscope (Reflex Measurement Ltd., Somerset, UK). In addition, contact or collapse of the maxillary segments, at the age of 11/2 year, were scored. Figure 20.2a, b shows a series of models from the study. Cephalometric radiographs were analyzed at 4 and 6 years of age. Facial appearance on full face photographs and photographs showing only nose and mouth was rated at different time points until 6 years of age.

The results show that neonatal orthopedics does reduce the alveolar cleft width before lip surgery, which means that at the time of lip closure, the cleft width in children treated with plates was narrower than in the other group. However, after lip closure in both groups, the alveolar cleft width diminished further, and at the time of soft palate closure, the maxillary arch dimensions in the two groups were comparable. The same holds true for the palatal cleft width that was also reduced during the orthopedic treatment, but after lip closure no significant differences were found between the groups anymore. Furthermore, when a plate was used, the palatal vault flattened. An explanation for this finding could be that the plate keeps the tongue out of the cleft, allowing the palatal shelves to flatten in the treated group. Once the soft palate was operated, the effect of treatment disappeared, and at 1–1/2 year of age, the shape of the palatal vault was comparable for both groups (Prahl et al. 2001). Neonatal orthopedics also did not prevent collapse of the maxillary arch (Prahl et al. 2003). The long-term results until 6 years of age showed no significant differences between the IO^+ and IO^- groups for maxillary arch dimensions (Bongaarts et al. 2006). In addition, the 5-year-old index (Atack et al. 1997), which is a measure for the jaw relationship, overjet, overbite, presence of cross bites, and sagittal occlusion were comparable in the two groups (Bongaarts et al. 2004).

Facial morphology at 4 and 6 years of age also did not differ between the two groups, although it has to be mentioned that at 6 years of age some attrition of the original sample had taken place as about 20 % of the patients was lost to follow-up (Bongaarts et al. 2009). At younger ages, there was no difference in facial appearance between the groups (Prahl et al. 2006). At the age of 4 years, IO had a positive effect on full facial appearance of UCLP children, but at the age of 6, only professionals saw a positive effect of IO on the nasolabial photographs. This seems to be rather irrelevant since patients deal with laymen in their daily life (Bongaarts et al. 2008).

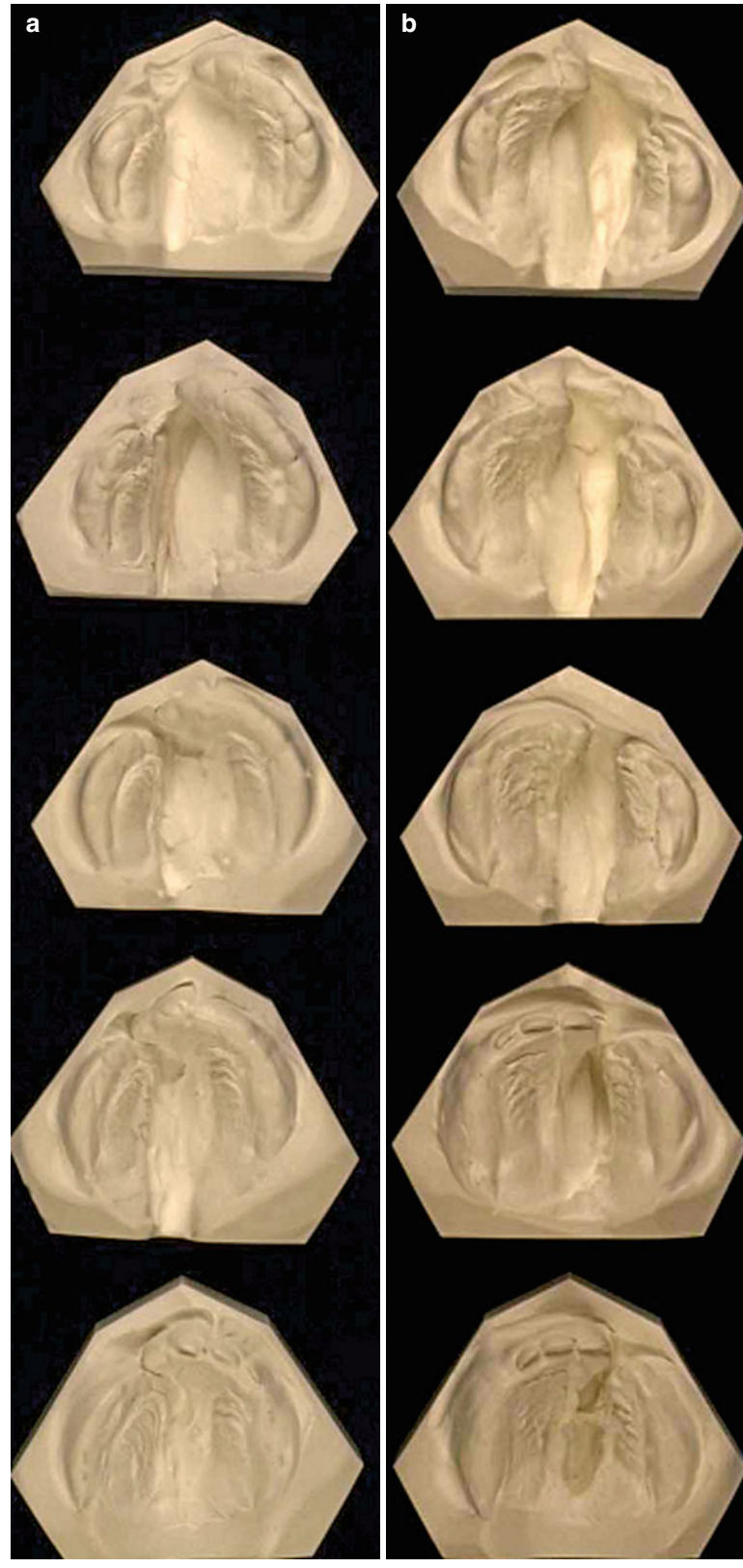

Fig. 20.2 (**a**, **b**) Series of study casts of two patients from the DUTCHCLEFT trial. The infant whose models are shown in (**a**) received no infant orthopedics; the infant whose models are shown in (**b**) received infant orthopedics. The five stages are (from upper to lower model) 0, 15, 24, 48, and 58 weeks of age

We concluded that infant orthopedics has only a temporary effect on maxillary arch dimensions which does not last beyond soft palate closure. It also does not improve the jaw relationship and occlusion in the deciduous dentition, and it has no measurable effect on facial morphology and facial and nasolabial appearance until 6 years of age. Therefore, from the orthodontic and surgical point of view, neonatal maxillary orthopedics could be abandoned.

20.5.5 Effect on Speech

Evaluation of speech and language development showed that at the age of 1 year, the children who wore plates presented enhanced production of alveolar sounds in babbling; however, at the age of 1 and 1/2 years when the plate was no longer used, consonant production in babbling was comparable in the two groups (Konst et al. 1999).

Reports on the later speech and language development of the children in the DUTCHCLEFT study showed a limited effect on speech (Konst et al. 2003a, b, c). The speech results at 2.5 years of age showed differences in intelligibility between the groups. In two different experiments, untrained listeners as well as experienced speech and language therapists gave higher intelligibility ratings to the children who formerly used plates (Konst et al. 2003c). However, data obtained by a transcription task indicated no differences in actual intelligibility (Konst et al. 2000), but compared to their non-cleft peers, the children with clefts were significantly less well understood.

At 2.5 years of age, the phonological development of the IO^+ children was normal or delayed, whereas most children who were not treated with a plate had abnormal development. Half a year later, it appeared that the IO^+ children had acquired more initial consonants than the IO^- group (Konst et al. 2003b). In the same age groups, the IO^+ children used longer sentences than the IO^- children, indicating that their grammatical development was more advanced. At the age of 6, no differences in expressive language skills between the two groups were found (Konst et al. 2003a). Hence, neonatal maxillary orthopedics did not have long-lasting effects on language development.

Regarding the claimed benefits of neonatal maxillary orthopedics on speech development in unilateral cleft lip and palate children, it can be concluded from this trial that there is a beneficial but rather limited effect until the age of 2 and 1/2 years. However, irrespective of neonatal maxillary orthopedics, the speech of children with clefts remains far behind that of their non-cleft peers. Due to lack of funding, the speech samples at the age of 6 years have not yet been analyzed.

20.5.6 Cost-Effectiveness

Nowadays, cost-effectiveness is an important issue, as costs of health-care interventions are often topic of debate. Especially in cases of reimbursement decisions, cost-effectiveness estimates can be useful. Together with the results of clinical effectiveness studies, cost-effectiveness information can be used to determine whether a certain treatment should become available to patients. The main principle of cost-effectiveness analysis is to estimate the costs and the treatment outcome (= effectiveness), compared to an alternative treatment (Drummond et al. 1997). Based on this explicit comparison, the difference in costs is related to the difference in effectiveness between alternative treatments. A prospective study in a randomized clinical trial design, such as DUTCHCLEFT, in combination with an economic evaluation is a good vehicle to analyze the cost-effectiveness (Severens et al. 1998; Cunningham 2001).

In DUTCHCLEFT, a cost-effectiveness analysis was planned for all three research areas that were mentioned above (see A, B, C). For the cost-effectiveness analysis, a so-called societal point of view was used, indicating that cost of treatment was based on real prices rather than fees. Besides this, a differential approach was used, aiming to calculate the difference in cost between yes or no neonatal maxillary orthopedics. A more extensive description of the cost analysis methods used in DUTCHCLEFT can be found in our earlier publications (Severens et al. 1998, 1999), which together with the publication by Cunningham [99] provide a good starting point to learn more about economic health evaluation.

In a preliminary report, the short-term cost-effectiveness of neonatal maxillary orthopedics

was based on the duration of the operation for surgical lip closure (Severens et al. 1998). However, this effectiveness parameter has little to do with effectiveness in terms of clinical outcome, but at that time, as the trial was still running, no other effectiveness variables were available. In 2004, we published data on cost-effectiveness of neonatal maxillary orthopedics compared to no such treatment, focusing on speech outcome at 2 and 1/2 years of age (Konst et al. 2004). The patients' speech was assessed by a panel of five speech therapists with experience in assessment of cleft lip and palate speech. At the time of evaluation, full data of both groups were not yet available. In each group, the data of ten children could be analyzed. The cost-effectiveness analysis required that the effects of treatment were expressed in one general effect measure. Therefore, the listeners were asked to rate speech quality on a 10-point scale. A detailed description of the full perceptual evaluation is given by Konst et al. (2003c).

The group that was treated with neonatal orthopedics had a significantly better rating for speech (IO^+: 3.52, SD 1.75; IO^-: 2.18, SD 0.62). The total costs of neonatal maxillary orthopedics were of course higher in the treatment group. The resulting cost-effectiveness ratio for IO^+ versus IO^- was EUR 1041.00 for 1.34 point of speech improvement. Relative to the total costs spent on the management of children with clefts, the financial investment to obtain this speech improvement is rather limited. Furthermore, the additional costs of neonatal maxillary orthopedics might be partly outweighed by the costs prevented for speech therapy in later years. This needs to be investigated further when the 6-year data of all children have been analyzed.

Conclusions

In 1991, the National Institute of Dental Research (NIDR) sought grant applications to conduct prospective and/or retrospective clinical trials evaluating treatment procedures for nonsyndromic, unilateral cleft lip and palate. The background information was that treatment sequence, timing, methods, and surgical techniques were all controversial. The long-term impact on maxillofacial growth and speech was considered to be of primary importance. This initiative reflected the lack of evidence with which treatments were carried out up to this point in time. This also holds true for neonatal infant orthopedics. Prior to the start of the randomized clinical trial of the effect of presurgical orthopedics in UCLP, we attempted to perform a meta-analysis of the existing literature, but this proved not to be possible due to the lack of randomized clinical trials, inconsistent outcomes and result reporting, missing data, small sample sizes, confounders, covariates, and publication bias. This problem has not been solved yet. A recent Cochrane review could only identify three randomized clinical trials into the effect of neonatal plates on feeding (Bessell et al. 2011). The available information on bilateral cleft lip and palate patients is even more sporadic. This was the main reason to start the DUTCHCLEFT trial.

Based on the results of DUTCHCLEFT so far, we conclude that neonatal maxillary orthopedics in unilateral cleft lip and palate patients as performed in this trial is not necessary for feeding, parents' satisfaction, or orthodontic and surgical reasons. Regarding speech, a positive but very limited effect was found until the age of 2 and 1/2 years, but the speech of the children with clefts remained far behind that of their non-cleft peers anyway. Relative to the total costs of the treatment of a UCLP patient, the financial investment to reach this effect was rather limited. However, it is questionable whether this limited effect is important enough to justify neonatal orthopedics. It should also be taken into consideration that the 6-year results for speech due to lack of funding have not yet been analyzed.

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21 A Comparison of the Effects of the Latham–Millard POPLA Procedure with a Conservative Treatment Approach on Dental Occlusion and Facial Aesthetics in CUCLP and CBCLP

Samuel Berkowitz and Martha Mejia

21.1 Dental Occlusion

One of the most widely debated areas in the treatment of cleft lip and palate involves the use of presurgical orthopedics and periosteoplasty with lip adhesion (POPLA) designed by Ralph Latham (orthodontist) and D. Ralph Millard, Jr. (plastic surgeon) (Millard 1980, 1986). They contend the POPLA procedure is superior to a conservative nonpresurgical orthopedic treatment (non-POPLA) for producing more aesthetically appealing lip/nose surgery, while still allowing for good midfacial growth and dental occlusion in complete unilateral cleft lip and palate (CUCLP) and complete bilateral cleft lip and palate (CBCLP) patients. This study compares the dental occlusion of POPLA with conservative, nonpresurgical orthopedic treatment (non-POPLA), which Millard had been using between 1960 and 1980 (Berkowitz 1996a, b). POPLA proponents and others, who may not resort to a lip adhesion but who still use the same presurgical orthopedic procedure designed by Latham in CBCLP patients for forcefully retracting the protruding premaxilla, favor the attainment of improved facial aesthetics and palatal arch alignment soon after birth with or without periosteoplasty. They speculate that the early aesthetic benefits will remain as the face grows and develops (Millard and Latham 1990; Dufresne and So 1992; Millard et al. 1998; Mulliken 2001).

In CUCLP and CBCLP, presurgical orthopedics is usually followed by periosteoplasty in the expectation that the resulting bone bridging created will avert the need for secondary alveolar bone grafts at a later date.

It is not a simple or a lightly assumed task to offer a brief challenging the rationale for presurgical orthopedics and periosteoplasty for infants with complete unilateral or complete bilateral cleft lip and palate. Advocates of the POPLA concept, or a variant of it, are few but well respected. Their proposed goal of making things right and whole as early as possible seems sensible and has great emotional appeal.

This first of a two-part serial retrospective dental occlusal and facial study covers more than 40 years of recording with serial dental casts and lateral cephaloradiographs, the sequential influences of both the POPLA and conservative non-POPLA procedures on palatal development, and the anterior and buccal dental occlusion. Part one of this study uses serial dental casts to determine the extent of anterior and buccal crossbites. Detailed dental occlusal analyses of these

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records test the efficacy of the POPLA procedure compared to the non-POPLA treatment employed by Millard and Berkowitz from 1960 to 1980 prior to the POPLA treatment years.

Part two of this study will analyze facial changes using serial lateral cephaloradiographs.

21.1.1 Method and Materials

The complete unilateral and bilateral clefts of the lip and palate cases, treated by either presurgical orthopedics (POPLA) or the nonpresurgical orthopedics (non-POPLA) procedure, were chosen from the files of the longitudinal facial and palatal growth studies of the Miami Craniofacial Anomalies Foundation of patients from the South Florida Cleft Palate Clinic, the University of Miami School of Medicine. Dr. Ralph Millard, Jr., performed lip, nose, and palatal surgery in both test samples. Secondary alveolar bone grafts and maxillary and/or mandibular osteotomies and maxillary distraction osteogenesis were performed by S.A. Wolfe. Samuel Berkowitz documented growth changes with dental casts, lateral cephaloradiographs, panorexes, and photographs and performed all treatment orthodontics other than presurgical orthopedics (Millard et al. 1988, 1998). Berkowitz treated a number of children in the POPLA group who had extensive anterior crossbites starting when they were about 9 years of age so that beginning at this age, there is a reduction in the frequency of the cases with anterior crossbites.

21.1.1.1 POPLA: Presurgical Orthopedics with Lip Adhesion (Millard et al. 1998) (Fig. 21.1a, b)

Ralph Latham supervised the plastic surgery residents in the manipulation of the palatally pinned presurgical orthopedic appliance. Another orthodontist later performed the same relatively simple procedure. Because of the training and close supervision involved in the treatments given, there was relatively little variation in this procedure over the years covered by the POPLA data. In CBCLP, the appliance mechanically expands the lateral palatal segments, allowing for the retraction of the protruding premaxilla into position within the alveolar arch (Figs. 21.2, 21.3, 21.4, 21.5, and 21.6). In CUCLP cases, the mechanical forces bring the premaxillary portion of the larger segment mediopalatally, and in most instances, the lesser segment is carried forward 2–3 mm to make contact with each other (Fig. 21.5a, b). Afterward, the floor of the nose is surgically closed, and a periosteoplasty is performed to permit the migration of alveolar osteoblasts to bridge the alveolar gap space. After the

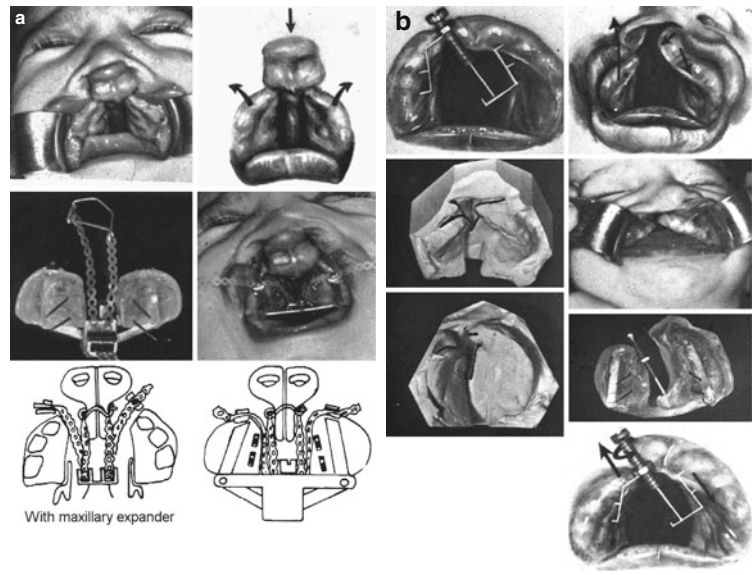


Fig. 21.1 (**a**) CBCLP. Latham's presurgical orthopedic appliance. The elastic chain creates the activating forces to retract the premaxilla while expanding the palatal segments. The posterior segment is pinned to the palate for approximately 2 weeks. The premaxillary pins, which are pulled by the elastic chain, are positioned anterior to the premaxillary vomerine suture. (**b**) CUCLP. Latham's presurgical orthopedic appliance. The screw knob controls the movement of the pinned appliance. The premaxilla is bodily rotated mediopalatally while the cleft lesser segment is moved anteriorly approximately 2–3 mm to make contact with the smaller cleft segment

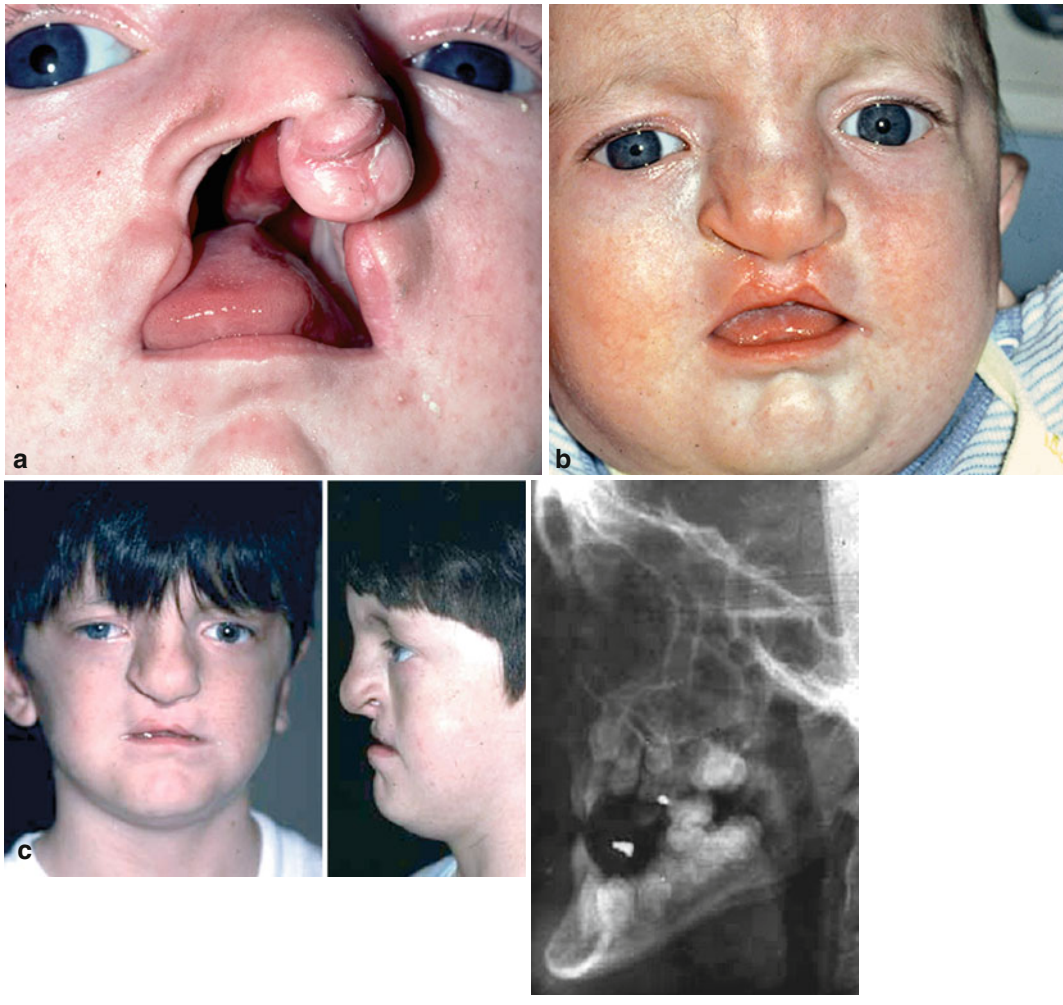


Fig. 21.2 (**a**–**j**). (**a**) Newborn. A small asymmetric protruding premaxilla. (**b**) After premaxillary retrusion. Lip adhesion followed by definitive lip surgery and gingivoperiosteoplasty (POPLA). (**c**) Midfacial deficiency due to retruded premaxilla

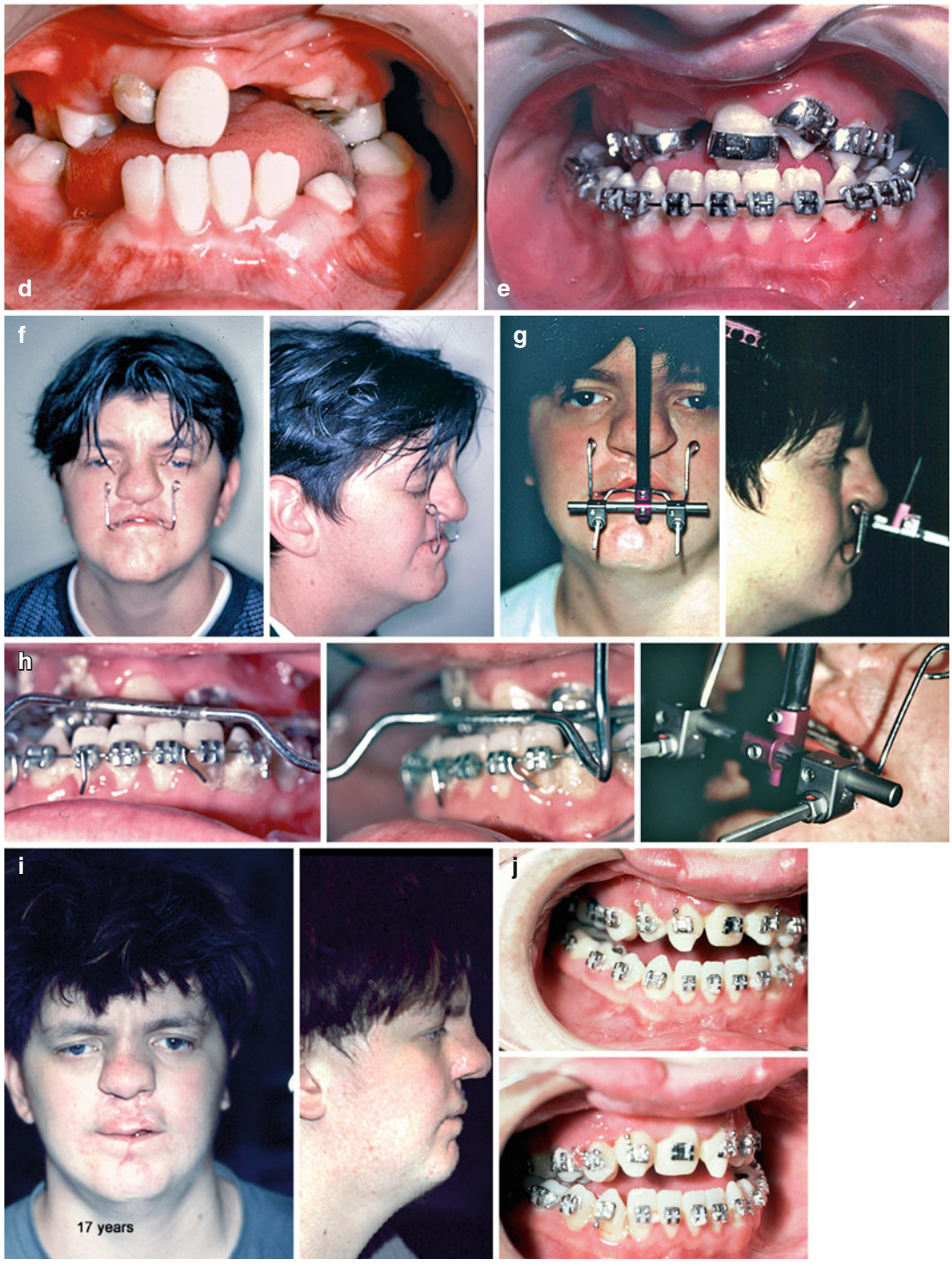


Fig. 21.2 (*continued*) (**d**) Retruded premaxilla showing some missing incisor teeth. (**e**) Orthodontic appliances placed to correct anterior crossbite and missing incisor spaces. Protraction facial mask with Class III mechanics was unsatisfactory in correcting this problem. The use of protraction facial mask was not successful in reducing midfacial deficiency due to upper and lower facial growth. This necessitated the use of maxillary distraction osteogenesis (**f**, **h**) Osteogenic appliance. (**g**) After maxillary advancement. (**i**, **j**) Following distraction osteogenesis to advance the maxillary denture. (**i**) After protraction facial mask – no changing. (**j**) Distraction osteogenesis to tip occlusion – now hypernasality is present

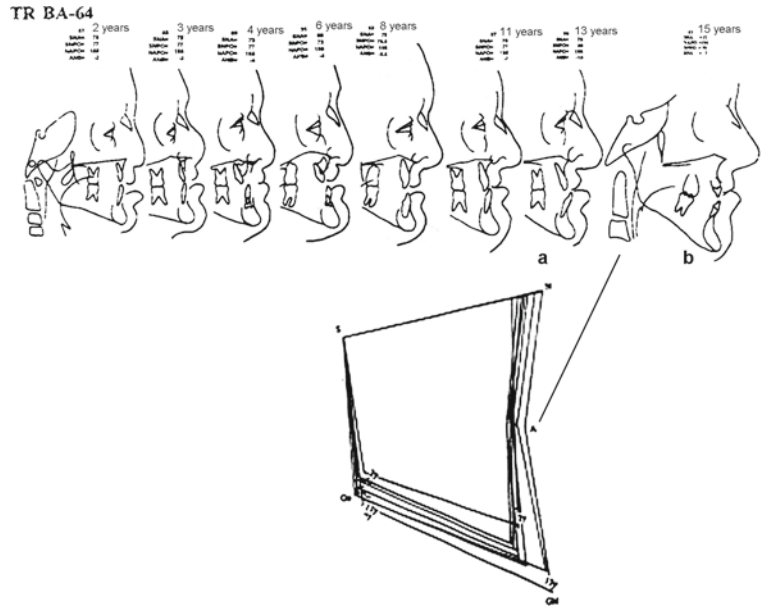


Fig. 21.3 (**a**, **b**) Serial cephalometric tracings showing the stability of the midfacial recessiveness even after the use of a protraction facial mask. There was no change in the Class I buccal occlusion since the orthopedic forces were directed to only advance the retruded premaxilla. (**b**) Postmaxillary distraction osteogenesis. Due to server hypernasality, maxilla advancement was discontinued

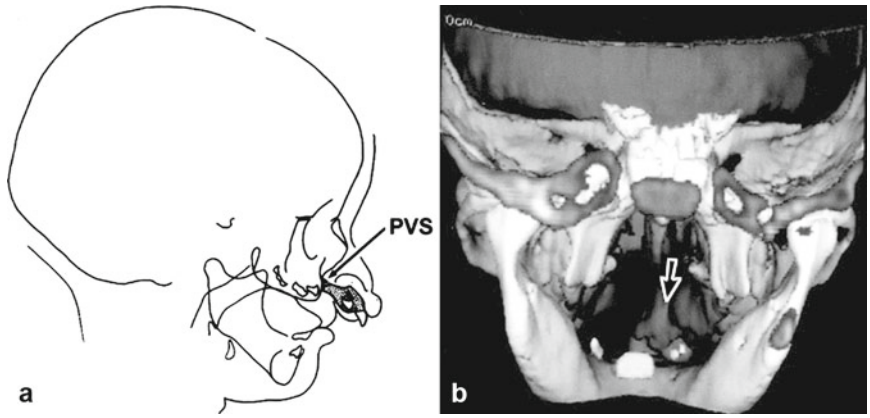


Fig. 21.4 (**a**) Lateral cephalometric tracing of a newborn with a complete bilateral cleft lip and palate shows the location of the premaxillary vomerine suture (*PVS*) posterior to the protruding premaxilla. (**b**) Frontal computed tomography scan of a patient with complete bilateral cleft lip and palate who was treated with the presurgical orthopedics, gingivoperiosteoplasty, and lip adhesion protocol. Only the left palatal segment is fused to the premaxilla with a bone bridge. Note the premaxilla "telescoping" at the premaxillary vomerine suture (*arrow*), the junction with the nasal septum

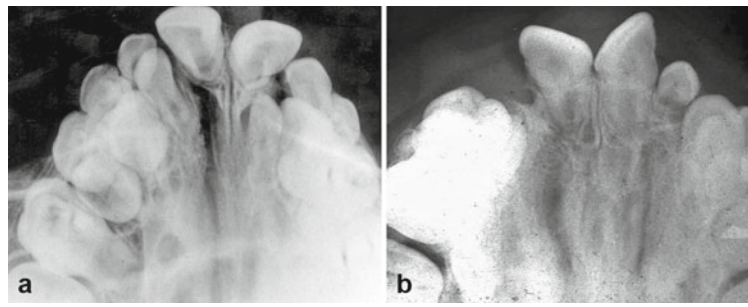


Fig. 21.5 (**a**) Palatal radiograph of a patient with complete bilateral cleft lip and palate who was conservatively treated at birth with a head bonnet with an elastic strip over the protruding premaxilla. The premaxillary vomerine suture was still open years later. (**b**) Palatal radiograph after premaxillary retraction with a Latham appliance shows a synostosis of the premaxillary vomerine suture

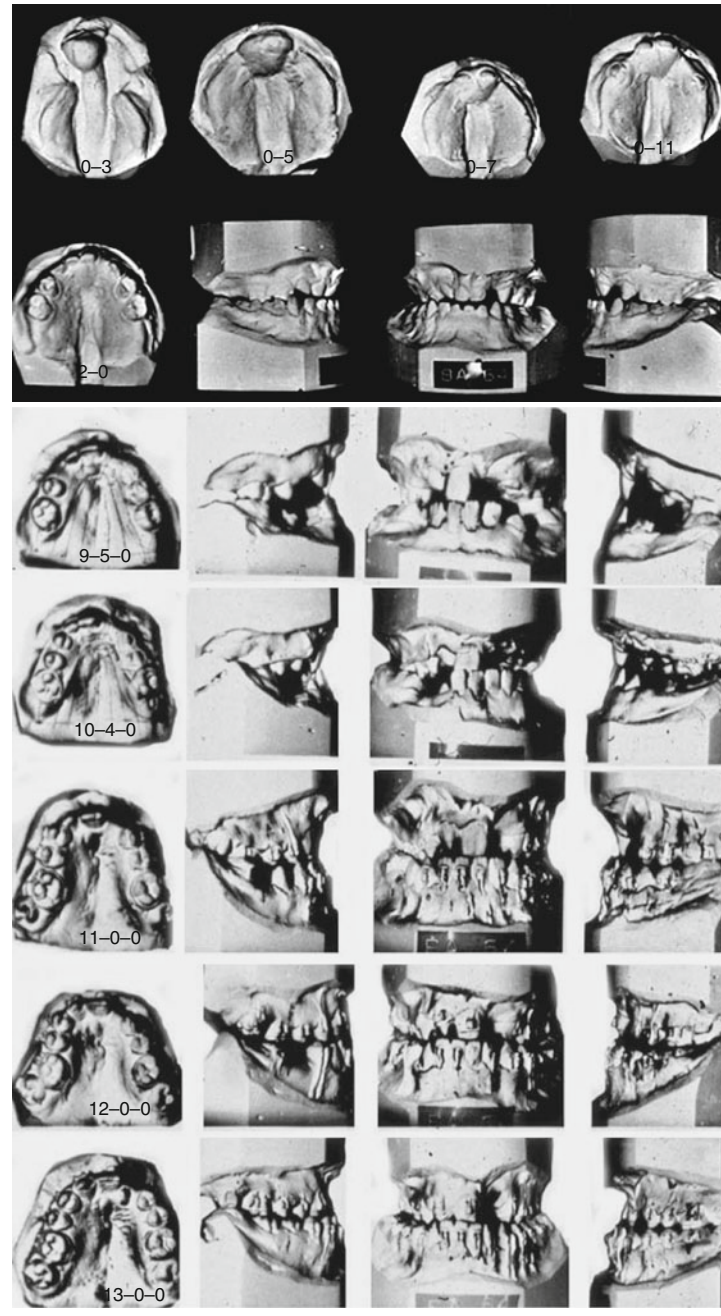


Fig. 21.6 Presurgical orthopedics, periosteoplasty, and lip adhesion (POPLA). The premaxilla was retruded and positioned within the arch. Followed by periosteoplasty, the palatal cleft was closed at 2 1/2 years using a von Langenbeck procedure with a modified vomer flap 9–13 years: The use of a protraction facial mask to advance the premaxilla was unsuccessful. This was followed by midfacial distraction osteogenesis

premaxillary retraction, a lip adhesion is performed followed 6–8 months later by definitive lip surgery with "forked" flaps.

21.1.1.2 Non-POPLA Conservative Treatment of CBCLP and CUCLP

The protruding premaxilla in CBCLP is ventroflexed by the forces generated by a head bonnet with elastic strip positioned across the prolabium, followed by lip adhesion surgery. No attempt is made to bodily retract the premaxilla and place it within the alveolar arch. Palatal cleft closure using a von Langenbeck procedure with a modified vomer flap is performed between 18 and 36 months, depending on the size of the cleft space. In non-POPLA as well as POPLA cases, palatal expansion is sometimes used at 5–6 years of age to correct the buccal crossbite in both groups.

Orthodontics between 8 and 10 years of age on POPLA and non-POPLA patients is used to align the anterior teeth. In non-POPLA cases, it is performed prior to bilateral or unilateral secondary alveolar bone grafting. In POPLA cases, the anterior crossbite correction with full orthodontic appliances and a protraction facial mask is initialized at 8–9 years when one or both of the permanent central incisors are erupted. An attempt at this age is made to correct the posterior occlusion. None of the test groups were in the Class III buccal occlusion at this age. Standard orthodontics follows to align all the permanent teeth. In CUCLP and CBCLP, lip adhesion is mostly performed at 3 months in both series followed by definitive lip surgery at 6–8 months of age.

21.2 Discussion

Millard and Berkowitz have been associated since 1961 with the South Florida Cleft Palate Clinic, where presurgical orthopedic treatment (PSOT) was not being used. Millard has always been a strong supporter of the importance of serially documenting treatment outcomes with lateral cephalometric radiographs, dental casts, panorexes, and facial/intraoral photographs. To that end, they have worked diligently.

After 20 years of performing nonpresurgical orthopedics (non-PSO) prior to the utilization of the POPLA treatment procedure, Millard published an operational plan to investigate and record facial–palatal changes when utilizing the Latham procedure (Millard et al. 1988). Today, Berkowitz has assembled an extensive collection of serial records that have always been available for review by any interested party. Kai Henkel (1997), a visiting professor of plastic surgery from Rostock, Germany, after reviewing the POPLA treatment serial case records, published a review stating that the procedure resulted in unsatisfactory facial aesthetics and dental function.

After 40 years of recording facial, palatal, and dental growth changes in POPLA and non-POPLA cases, the authors of this brief believe that criticism is in order for a clinical procedure whose outcome results have now been adequately reviewed using serial objective records.

It is unfortunate that a controversy still exists as to the utility of POPLA or similar treatment with or without lip adhesion 20+ years after its introduction and after the critiques to which it has been subjected (Berkowitz 1996a, b). First, Georgiade and Latham (1975) and Latham (Latham 1970, 1973, 1980) have failed to publish any outcome studies. More recently, Millard and Latham have published a limited outcome report using Berkowitz's palatal cast records (Berkowitz 1996a, b). Millard and Latham's coauthors Huifen, Spiro, and Morovic performed linear measurements of the changing palatal size rather than analyze the relative growth of the opposing jaws by reviewing the dental occlusion.

Dufresne and So, in their chapter supporting the presurgical orthopedic procedure for CBCLP and CUCLP patients, referred only to Millard's original introductory statements of the POPLA procedure in Cleft Craft (Vol. III) (Millard 1980) and listed no other supporting references, yet advocate its use (Dufresne and So 1992). Cutting and Grayson (Cutting et al. 1998; Cutting and Grayson 2000) limited their PSOT report to the effect of periosteoplasty in successfully producing bone bridging of the alveolar cleft, yet no mention was made of its effect on the dental occlusion.

Berkowitz (1996a, b), in a preliminary report, compares Millard's POPLA with Millard–Berkowitz's conservatively (non-POPLA) treated CBCLP outcomes. That comparative outcome study confirmed the negative effects of POPLA on facial aesthetics and dental occlusion. The study presented herein expands on that report.

In 1996, Berkowitz and Latham were asked by the American Cleft Palate Craniofacial Association program committee to debate the utility of this procedure at their annual meeting in San Diego, California. At that meeting, Berkowitz and LaRossa (plastic surgeon) presented long-term case reports that were critical of the procedure, but no supporting case studies were forthcoming from Latham and Morales (plastic surgeon).

Many clinicians who advocate the use of Latham's or any other presurgical orthopedics

Table 21.1 Number of cases at each age level in presurgical and nonpresurgical orthopedics treatment groups

	Approximate age of participant				
	3 years	6 years	9 years	12 years	Total sample
Unilateral cleft					
POPLA treatment	30	43	34	18	125
Non-POPLA treatment	51	54	46	33	184
Bilateral cleft					
POPLA treatment	21	20	15	9	64
Non-POPLA treatment	49	49	40	35	173

POPLA presurgical orthopedics gingivoperiosteoplasty and lip adhesion

(PSO) state that one of the appliance's benefits is to prevent "collapse" of the lateral palatal segments. The frequent use of the word "collapse" to describe palatal arch relationships after neonatal lip surgery is unfortunate and needs to be better understood. The term is misleading for it conjures up an unwarranted sense of foreboding.

The word "collapse" was introduced into the cleft-treatment lexicon in the 1960s by some surgeons and orthodontists to describe the palate's physical state after uniting of the lip and causing medial palatal movement of overexpanded palatal segments. They evidently did not realize that in complete lip and palatal clefts, the resulting medial palatal movement (molding) is beneficial because it reduces palatal cleft size and corrects the overexpanded palatal segments relationship. Serial studies have shown that overlapping palatal segments in the deciduous and mixed dentition are of no clinical importance.

The word "collapse" implies that this condition is bad and should be prevented. However, after years of analyzing serial dental casts, many orthodontists have concluded that establishing lip muscle continuity leads to good geometric palatal changes no matter their temporary neonatal geometric relationship. Overlapped segments do not impede normal palatal growth, and in most cases, such overlapped segments can easily be properly realigned using relatively simple orthodontics. Cleft segments in posterior crossbite at an early age are not indicative of future palatal maldevelopment. Since the width of the cleft space influences the type and timing of surgical palatal closure, one would prefer to have a small cleft space prior to palatal surgery to reduce the possibility of creating growth-inhibiting scarring while producing a normal palatal vault space.

In contrast to conservative non-POPLA cases, POPLA-treated patients require extensive and costly orthodontic treatment to correct the anterior crossbites, regain lateral incisor spaces, and achieve upper to lower anterior arch congruency. Often the degree of facial and palatal distortion is so extensive in POPLA cases that additional surgical intervention is necessary at a later age. Some parents have reported their children's experiencing psychosocial problems due to the lack of peer acceptance of the concave facial profile.

POPLA in CBCLP (Table 21.1): The bodily retracted protruding premaxilla is retracted and placed in excellent alignment within the alveolar segments (Fig. 21.1a, b). Since no lateral flexion of the nasal septum is seen in cat scans, it was concluded that the premaxilla is "telescoped" posteriorly at the premaxillary vomerine suture (PVS) (Fig. 21.4). The bending at the PVS is also seen in POPLA-treated CUCLP (Fig. 21.5). The PVS is not observed in follow-up palatal POPLA radiographs (Fig. 21.5).

The diagram (Fig. 21.7) is designed to show the error of palatal segmental movements in POPLA CUCLP cases. Our 3D analysis of the CUCLP palatal arch changes demonstrates that the premaxillary portion of the noncleft segment is brought mediopalatally while the smaller cleft segment is advanced, resulting in the loss of the lateral incisor space. This explains why an anterior dental crossbite is most likely to result.

Millard (1980) has written that premaxillary retraction and periosteoplasty might have a nega-

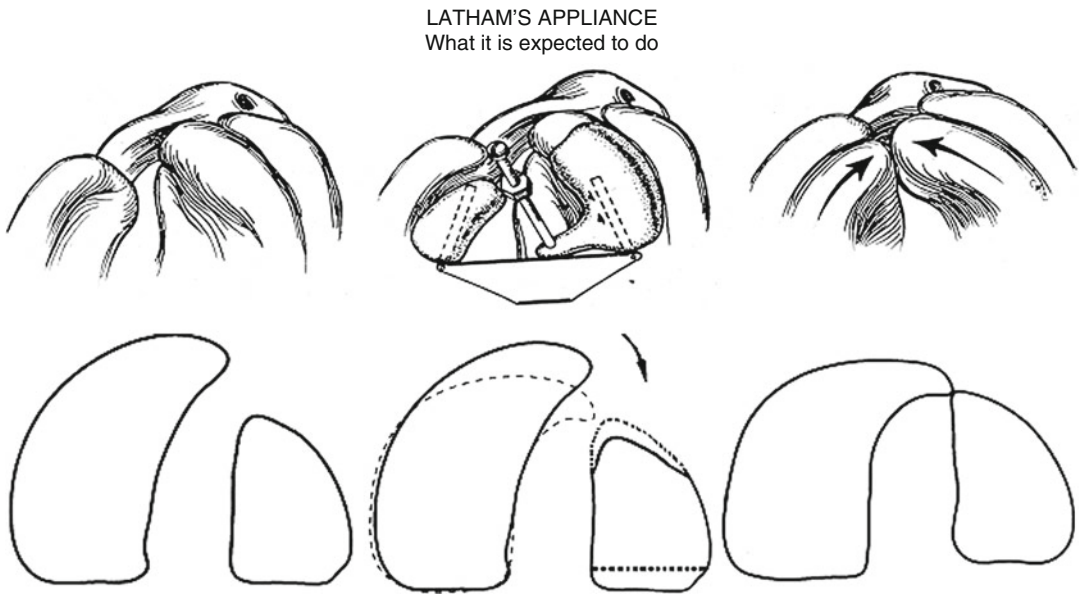


Fig. 21.7 (**a**) According to Latham, the mode of action of the appliance used in complete unilateral cleft lip and palate is the medial movement of both segments toward each other (Dufresne and So 1992). (**b**) None of our unilateral cleft cases, when activated with the Latham appliance, is shown below. The premaxillary portion of the noncleft segment is bent mediopalatally, while the lesser-cleft segment is carried slightly forward, into contact

tive effect on palatal growth, but he nevertheless believes this trade-off is acceptable to obtain early aesthetics, to close the floor of the nose, and to avoid the need for secondary alveolar bone grafting.

The desire of some surgeons to establish a child's well-balanced, aesthetically pleasing lips and nose soon after birth is understandable. However, extensive facial growth studies make clear that this should not be the top priority – that is, at the cost of good midfacial growth, dental occlusion, and speech at adolescence (Berkowitz 1996a, b; Pruzansky 1953; Pruzansky et al. 1973; Handelman and Pruzansky 1968; Friede and Pruzansky 1972; Friede 1973, 1977, 1978; Friede and Morgan 1976; Aduss et al. 1973; Vargervick 1983; Semb 1991). All of these goals should be possible without sacrificing.

This comparative study contradicts the belief that well-designed lip/nose surgery with Latham's PSO performed on newborns to achieve early facial aesthetics while damaging palatal growth sites will lead to excellent adult facial aesthetics and dental function. But, should the face fail to grow well, the supporters of POPLA suggest that it can be easily corrected without far-reaching consequences such as developing poor self-esteem as a result of having a retrusive midface. This condition does not lend itself to easy correction by midfacial surgery alone. Extensive post-surgical psychosocial therapy may be necessary in children with inadequately formed faces (Tobiasen 1996).

In POPLA cases, a buccal crossbite mostly involved the deciduous cuspid. The percentage of anterior crossbite cases increases with time (Fig. 21.29a, b). In some POPLA cases, when the premaxilla is not placed precisely within the arch, less bone bridging and fewer anterior crossbites occur. None of the cases show a Class III buccal (posterior) occlusion. The anterior dental crossbite in POPLA CBCLP and CUCLP is always due to the manipulated premaxilla's retrusive position, which is never self-correcting or could be orthodontically corrected in the deciduous or, in most cases, even in the permanent dentition. In vertically growing faces, the anterior crossbite is orthodontically correctable in most cases. When the mandible grows forward, as it does in most faces, the anterior crossbite worsens requiring

often a maxillary advancement surgery to create a proper dental overjet.

A typical CBCLP case of serial cephalometric tracings (Figs. 21.20 and 21.28) shows an early retrusive midface in the deciduous dentition, creating an anterior crossbite with a concave facial profile that worsens with time. An extensive cephalometric report will be forthcoming in part two of this overall study. In these CBCLP and CUCLP cases, the upper and lower face gradually grows forward, but the palatal length between the first molars and incisors remains constant (Figs. 21.30, 21.31, and 21.32). In most cases, LeFort I surgery or maxillary advancement with distraction osteogenesis is necessary, as it was in this CBCLP case. In the POPLA series, maxillary distraction osteogenesis is seldom utilized in CUCLP. This surgery results in immediate hypernasality due to an increase in pharyngeal depth leading to velopharyngeal incompetency (VPI). The hypernasality slowly diminishes in some cases within 1 year, but is not completely absent in all of these cases.

In CBCLP, 57 % of POPLA patients, but only 18 % of the non-POPLA-treated cases, exhibit an anterior crossbite at 6 years of age (Figs. 21.29, 21.30, and 21.31, Table 21.1). Posterior (buccal) crossbites are not always related to POPLA (Fig. 21.29). The treatability of buccal crossbites is also influenced by the extent of scarring created by the surgical cleft closure procedure. If a buccal crossbite is present, some may be easily corrected by 6–9 years of age in both test groups. As in CBCLP, the CUCLP shows slight anterior palatal growth. Most palatal growth occurs posteriorly to accommodate the developing molars (Figs. 21.30, 21.31, and 21.32).

21.3 POPLA CUCLP (Figs. 21.5, 21.8, 21.9, 21.10, 21.11, 21.12, 21.13, 21.14, 21.15, and 21.23, Table 21.1)

Early anterior dental crossbite in the deciduous dentition is associated with the loss of the lateral incisor space brought on by PSOT. This results from the mediopalatal positioning of the premaxillary portion of the larger noncleft segment, which is brought into contact with the forward, positioned lesser-cleft segment. The following periosteoplasty creates extensive bone bridging in over 80 % of the cases.

POPLA patients have greater transverse posterior arch width earlier than what is observed in non-POPLA cases. This is due to the palatal appliance preventing the neonatal overexpanded lateral segments from molding together and closing off most of the palatal cleft space. There is no clinical advantage for maintaining this increased palatal width at this early age since POPLA and non-POPLA cases will eventually attain ideal buccal occlusion after relatively simple orthodontics.

An anterior dental crossbite with some degree of midfacial retrusion occurs in 60 % of the CUCLP POPLA cases by 6 years of age (Fig. 21.29a, Table 21.1), while only 17 % of non-POPLA-treated cases experience an anterior crossbite (Fig. 21.29b, Table 21.1). In some POPLA cases that did not result in an anterior crossbite, the lateral segments are not in contact at the time of periosteoplasty. Slight or no bone bridging with a good lateral incisor space is associated with good incisor overjet with no midfacial retrusion. The loss of the lateral incisor space can be anticipated since alveolar bone is lacking in all cleft alveoli at birth, and for any bone bridging to occur, the alveolar segments need to be placed in contact. In these cases, the posterior palate is unaffected by PSO and is usually in a Class I or Class II occlusion (never in Class III) at 6 years of age.

Anterior dental crossbite correction in POPLA-treated CUCLP and CBCLP cases requires the advancement of the premaxilla in CBCLP or the premaxillary part of the noncleft segment in CUCLP. In CBCLP POPLA cases, the premaxillary segment, as a result of the early palatal manipulation and periosteoplasty of the closed-over cleft lateral incisor spaces, cannot accommodate the impacted lateral incisor, if present (Fig. 21.9). This space cannot be regained in the mixed dentition and only after extensive orthodontics in the permanent dentition in 50 % of the cases (Figs. 21.16, 21.17, 21.18, 21.19, 21.20, 21.21, 21.22, 21.23, 21.24, 21.25, 21.26, 21.27, and 21.28).

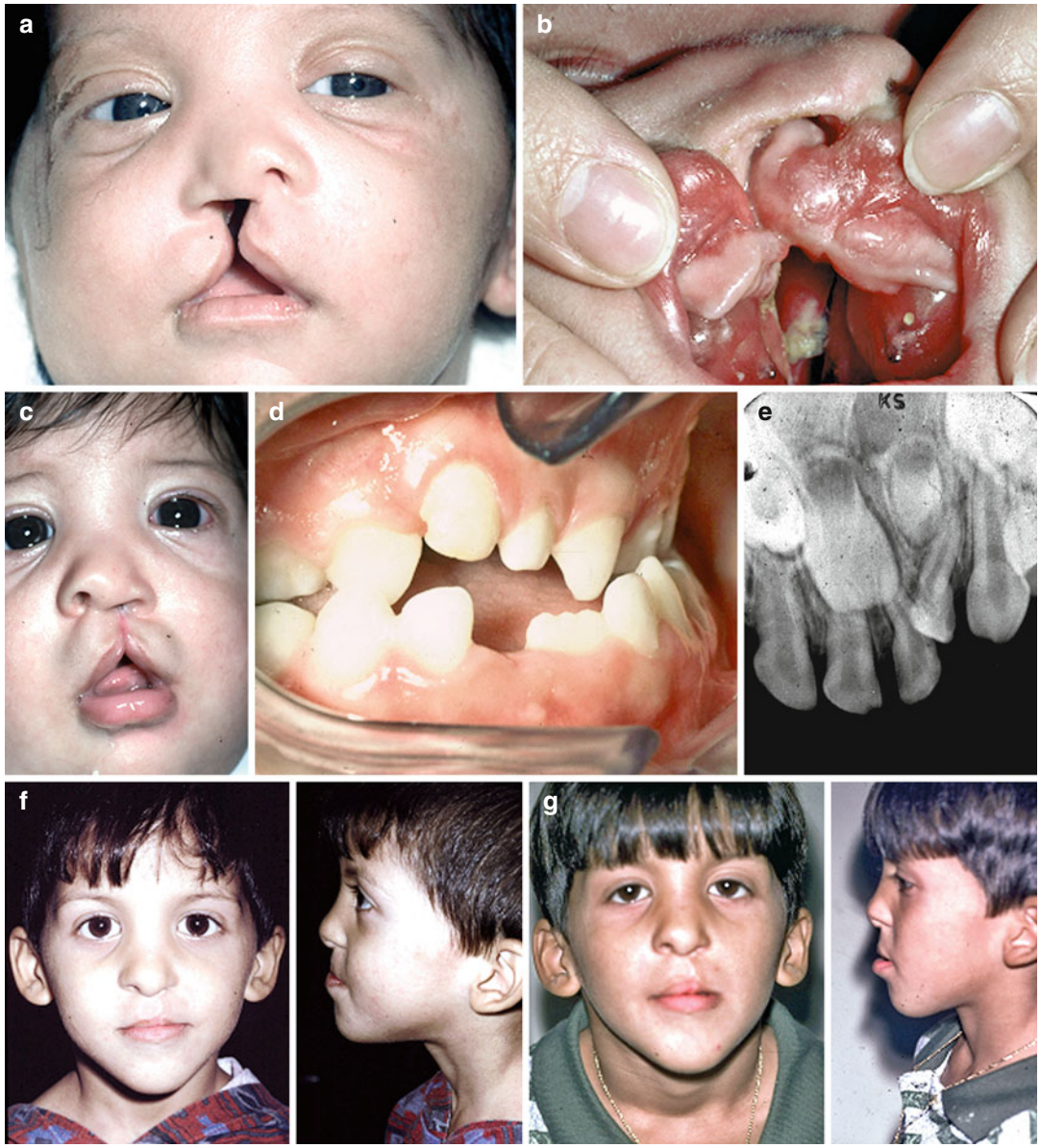
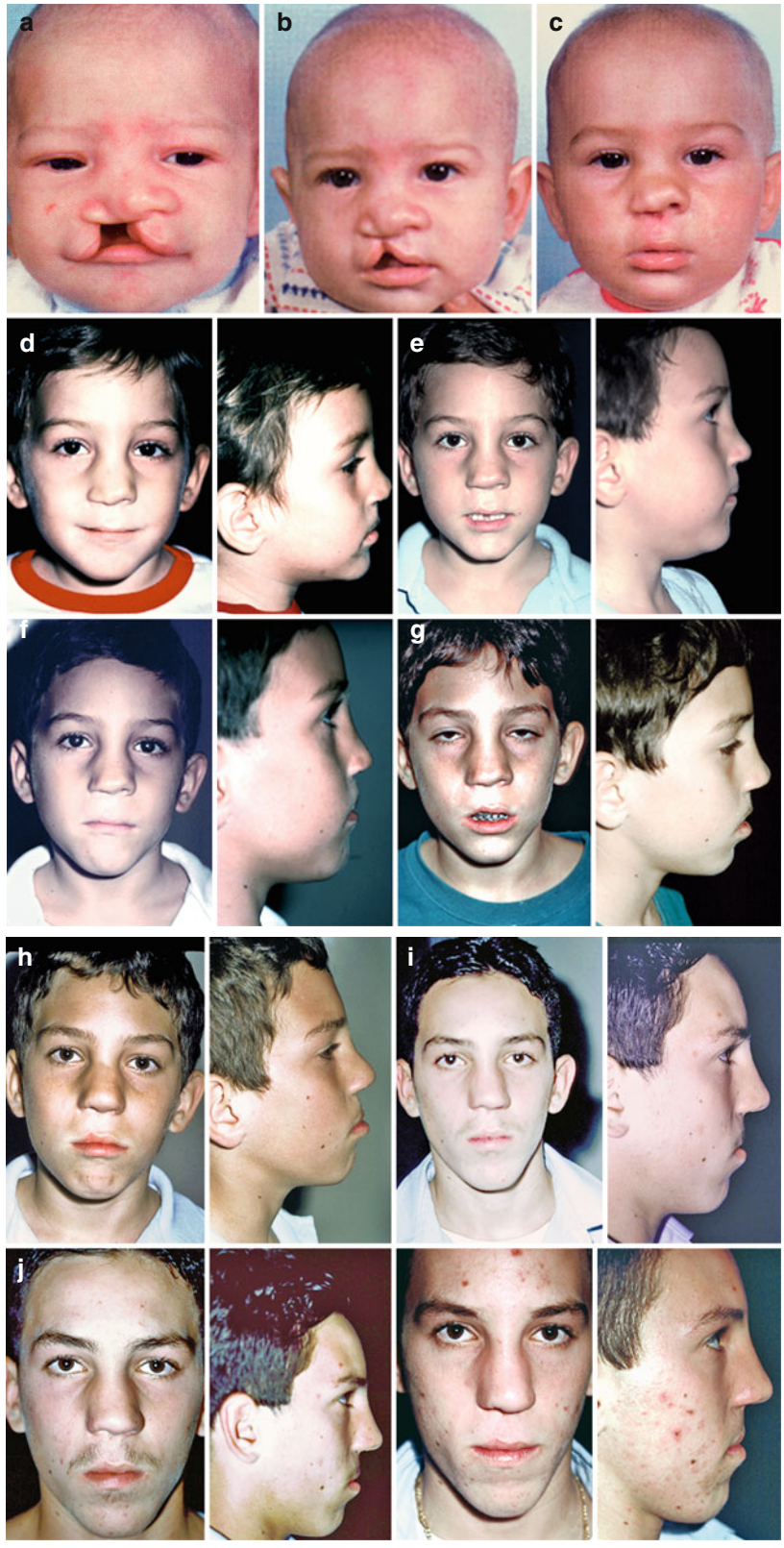


Fig. 21.8 (**a**) After appliance activations: lip and nasal distortions reduces when the palatal segments are moved together. (**b**) Alveolar segments in contact periosteoplasty is then performed. (**c**) Lip adhesion to establish muscle continuity and mold the overexpanded palatal segments in a preparation for a definitive lip surgery at 6 months of age. There are instances when defective lip and nose surgery can be performed without first performing a lip adhesion. (**d**) The absence of the upper right and left central incisors bringing the segments together, causes the right segment to move too far medially into a crossbite. (**e**) The x-ray of the maxillary incisor area shows the missing centrals and laterals of the permanent incisors. (**f**, **g**) Note that the upper lip is slightly retruded when compared to the lower lip. This is associated with the retraction of the premaxillary portion of the noncleft palatal segment Conclusion: Even if the maxillary incisor teeth are congenitally missing, it is important to keep all cleft sites open either for the eruption of the permanent or replacement teeth. This might necessitate the placement of a temporary tooth bearing palatal plate. Had periosteoplasty been done, the correction of the anterior crossbite would have been delayed. These cases represent the need to avoid replacing bony bridges across the cleft area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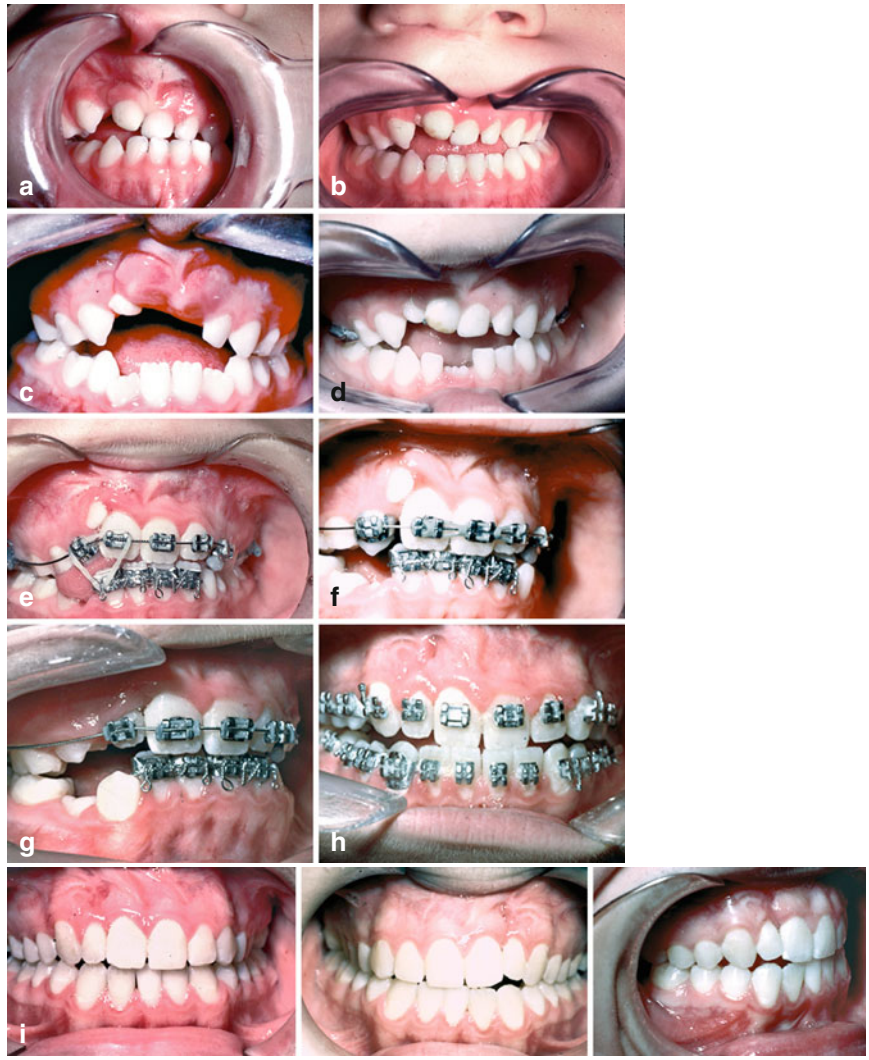

Fig. 21.10 (**a**, **b**) The retracted premaxillary portion of the noncleft segment was brought mediolaterally, blocking out the right lateral incisor space. The superior torque to both palatal segments created an anterior crossbite by the forces generated by the Latham appliance. (**c**, **d**) The crossbite became more noticeable when the deciduous teeth were lost and the permanent incisors began to erupt. (**e**) Vertical elastics were used to extrude the upper anterior teeth. (**f**, **g**) The displaced anterior teeth were brought into alignment. The maxillary central incisors were advanced, opening the lateral incisor space. (**h**) The thin bony alveolar bridge created by the periosteoplasty permitted the right and the left palatal segments to be moved later and the lateral incisor space opened. (**i**) A lower incisor was extracted, and the incisors were retracted in order to create an overbite and overjet. An anterior open bite resulted due to the original torque of the palatal segments. The correction of the alignment of the upper incisor teeth was not stable even though an upper retainer was worn. Comment: Occlusal stability is dependent on the position of the basal bone

Fig. 21.9 (**a**) Distorted nostrils and lip due to aberrant muscle pull. (**b**) Lip adhesion reduces nasal and lip distortion. (**c**) Rotation advancement, the definitive lip closure. (**d**–**g**) Facial photographs at different ages. Note: The vertical facial growth pattern creates an elongated antero-facial height. As a result, the midface does not appear to be retrusive. A facial protraction mask was utilized with intraoral palatal expander to advance the maxillary incisors (**h**, **i**, **j**) Even though there was extreme crowding of the maxillary anterior segment, the vertical facial growth pattern neutralized the obtaining of midfacial recessiveness. Lip/nose revision were excellent in creating symmetry

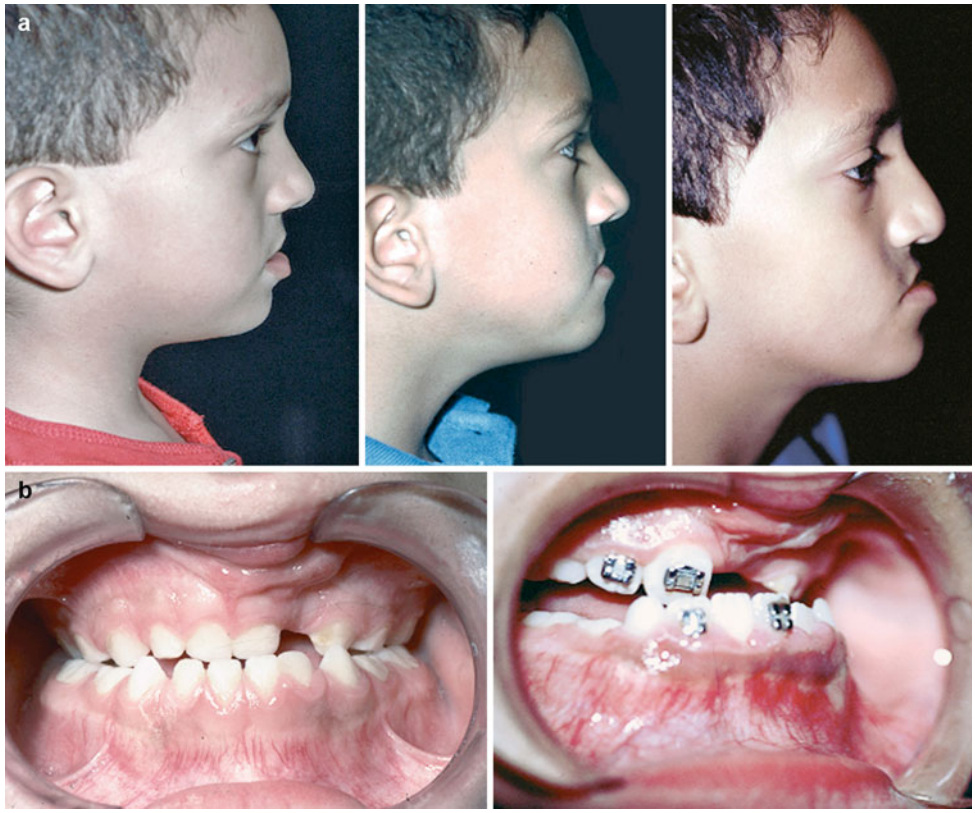

Fig. 21.11 (**a**) Facial changes. These show recessive midfaces at 4, 6, and 9 years of age. (**b**) Intraoral occlusal photographs showing anterior dental crossbites at 4 and 9 years of age

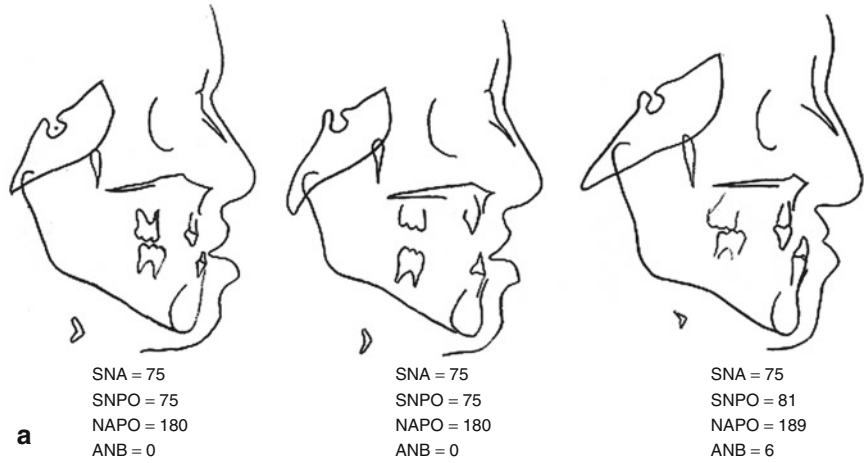

Fig. 21.12 (**a**) Superimposed facial polygons show that midfacial recessiveness does not change between 6 and 9 years of age. However, with greatly increased mandibular growth, the facial concavity worsens. Maxillary distraction is being planned to improve facial aesthetics and peer acceptability.

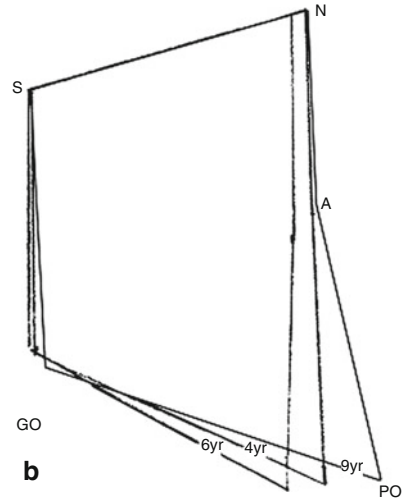

Fig. 21.12 (*continued*) (**b**) Lateral cephalometric tracings show midfacial recessiveness worsens with facial growth

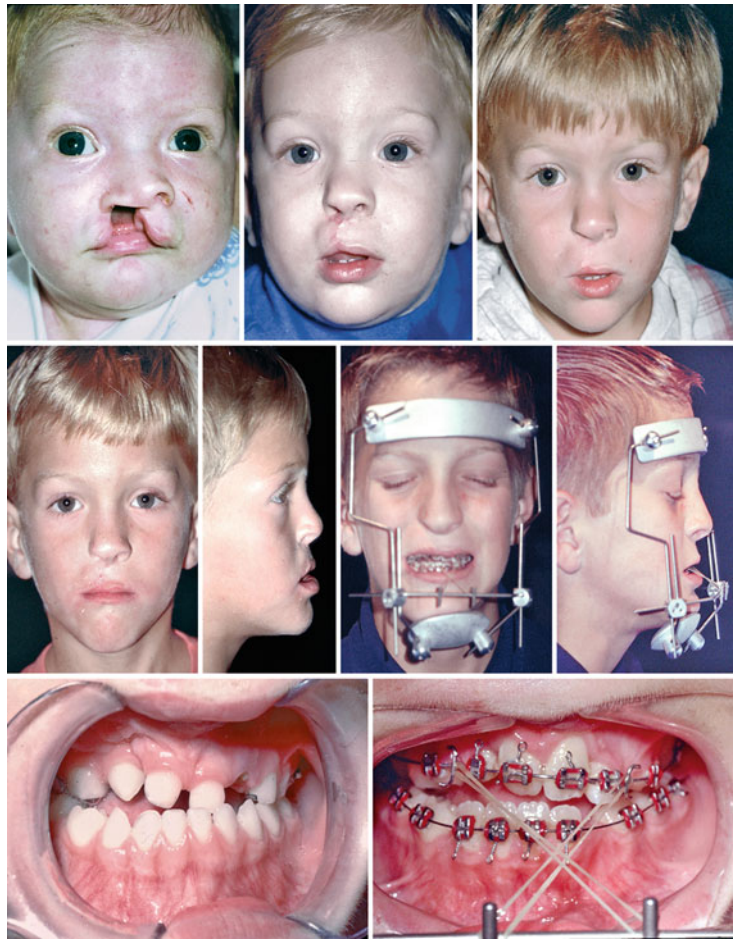

Fig. 21.13 Case BR 74. Complete unilateral cleft lip and palate. Severe anterior crossbite with crowding of incisor teeth and partial closure of the lateral incisor space. Treatment plan: uncrowd and advance incisor teeth using a protraction facial mask

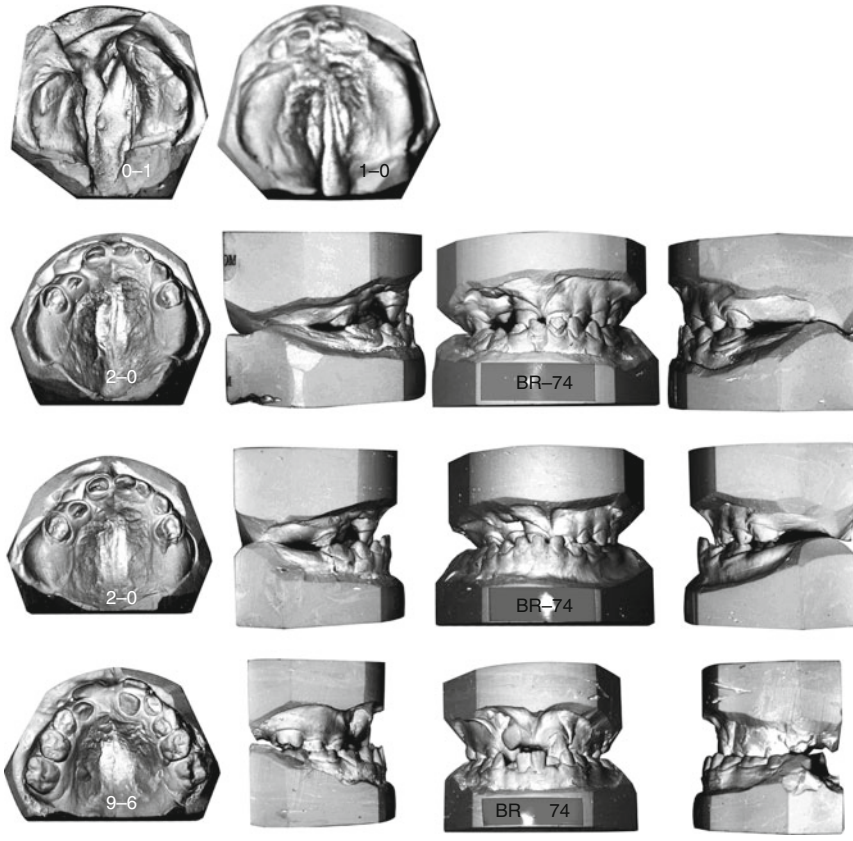

Fig. 21.14 Case BR 74. Latham–Millard presurgical orthopedics periosteoplasty with lip adhesion (POPLA) procedure. Serial casts reflect the creation of a severe anterior crossbite and anterior dental crowding. 0–1: at 1 year after the POPLA treatment, the premaxilla portion of the larger segment was brought palatally by the presurgical orthopedics. 2–0: 2 years of age. Severe anterior crossbite with partial closure of the right lateral incisor spaces. Palatal cleft still open. 2–6: 2 years and 6 months of age. Palatal cleft space closed with a von Langenbeck and modified vomer flap. Severe anterior crossbite with closure of the right lateral incisor space. No posterior crossbite. Comments: One can predict that the anterior crowding will become more severe with the eruption of the permanent incisors. The advancement of the anterior teeth is dependent upon premaxillary advancement, which is hindered by the bonny bridge created by the periosteoplasty. A protraction facial mask will be necessary to advance the premaxilla (see facial photographs)

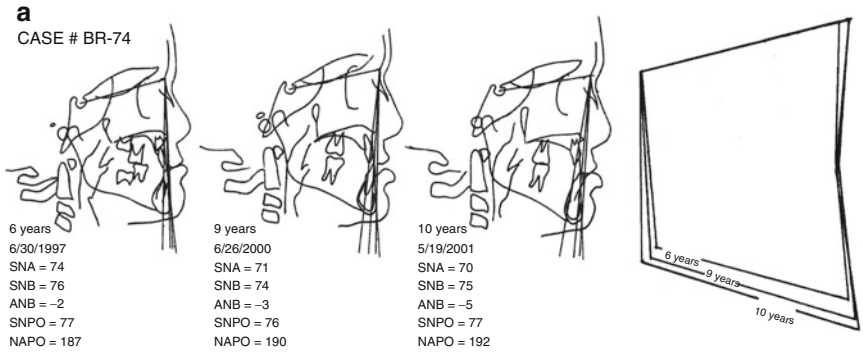

Fig. 21.15 (**a**) Case BR 74. Complete unilateral cleft lip and palate. Severe anterior crossbite with crowding of the maxillary incisor teeth as a result of retracing the premaxillary portion of the larger segment. This created a concave facial profile (cephalometric analysis)

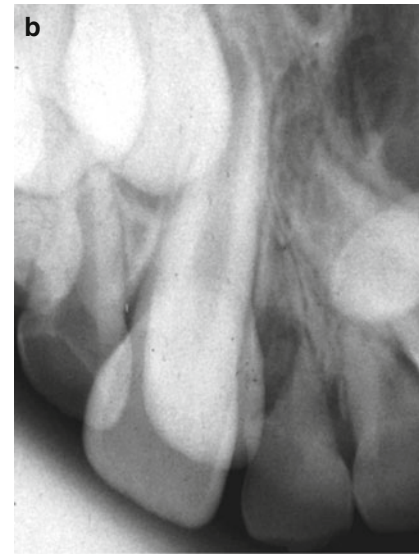

Fig. 21.15 (*continued*) (**b**) Case BR 74. Periapical radiograph. 7 1/2 years of age. Severe dental crowding of the maxillary anterior teeth

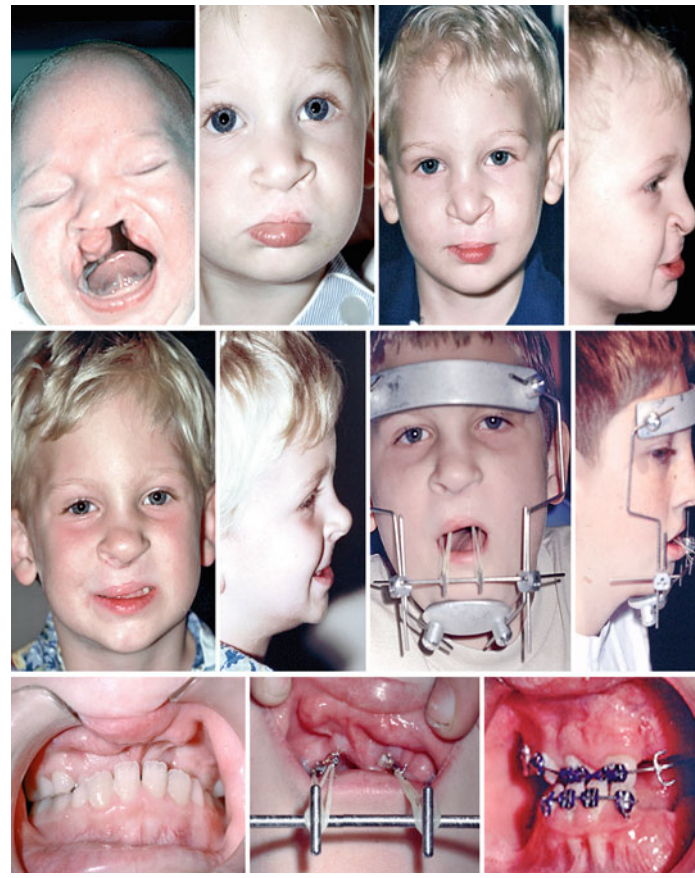

Fig. 21.16 Case BT 80. Complete bilateral cleft lip and palate. The protruding premaxilla was set back and aligned within the arch using the POPLA procedure. By 4 years of age, a severe anterior crossbite resulted. At 10 years of age, the teeth in the unerupted severely ventroflexed premaxilla still failed to erupt. This necessitated the placement of a gold chain on the imbedded central incisors. Hooks were placed on the chains with elastics extending to a Delaire-type facial mask. Treatment is still underway

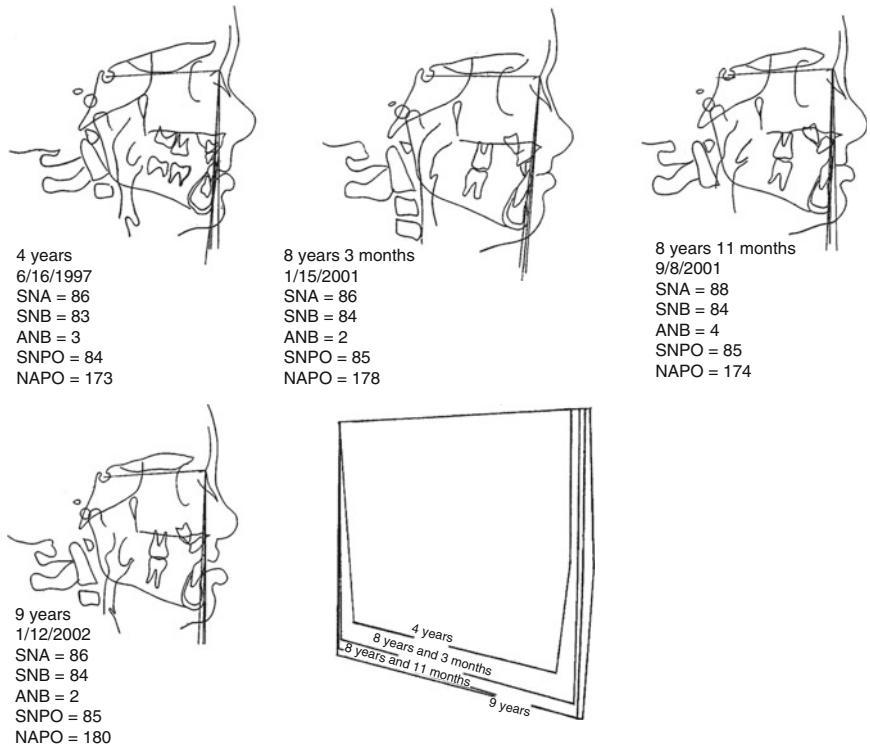

Fig. 21.17 Lateral cephaloradiographs show the unerupted severely ventroflexed premaxilla in crossbite. The superimposed cephalometric tracing shows the continued midfacial growth keeping up with the other facial areas, resulting in a flat facial profile. One can conclude that growth at the premaxillary vomerine suture is still active. However, the bony bridge between the lateral palatal segments and the premaxilla prevents the premaxilla's proper alignment and eruption into position

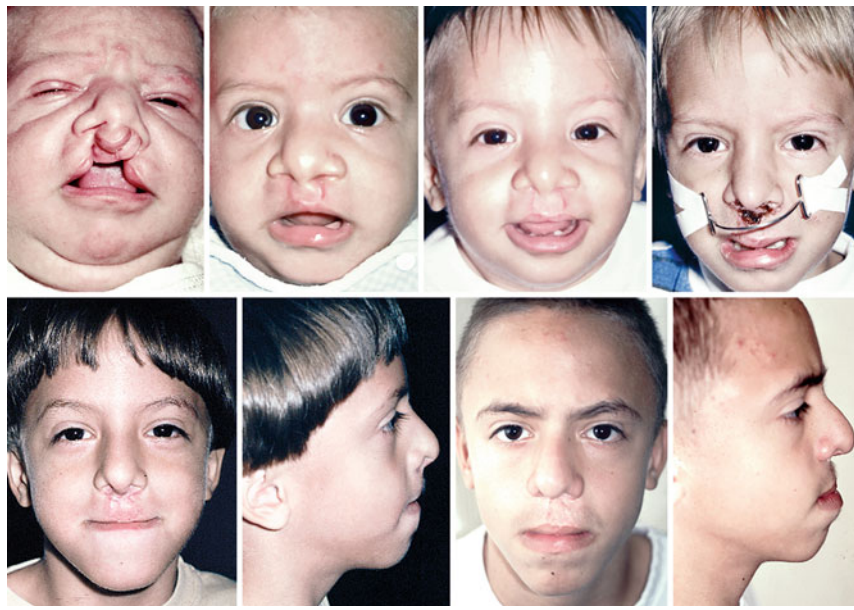

Fig. 21.18 Case BK 36. Bilateral cleft lip and palate. The protruding premaxilla was retruded and aligned within the arch using the POPLA procedure. At 7 years and 4 months of age, an anterior dental crossbite resulted. A protruding lower lip is due to the permanent mandible and flaring incisor teeth

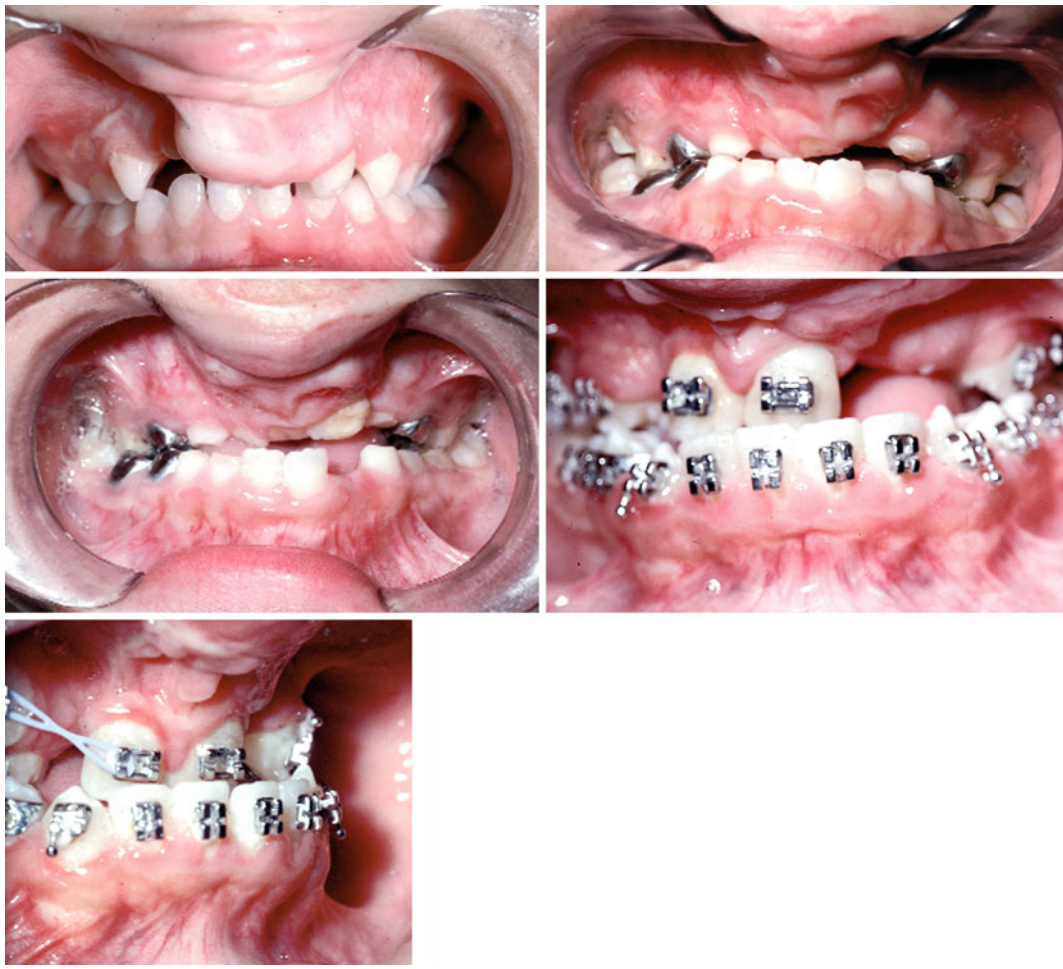

Fig. 21.19 Case BK 36. Bilateral cleft lip and palate. The corrective treatment plan was designed to advance the crowded anterior maxillary incisors with facial protraction mechanics and position the cuspids into the missing lateral incisors spaces. Extraction of the mandibular first bicuspids and retracting the incisors will help to reduce the crossbite without the need to advance the upper incisors. This case is still under treatment. Case BK 36. Bilateral cleft lip and palate. The corrective treatment plan was designed to advance the crowded anterior maxillary incisors with facial protraction mechanics and position the cuspids into the missing lateral incisors spaces. Extraction of the mandibular first bicuspids and retracting the incisors will help to reduce the crossbite without the need to advance the upper incisors. This case is still under treatment

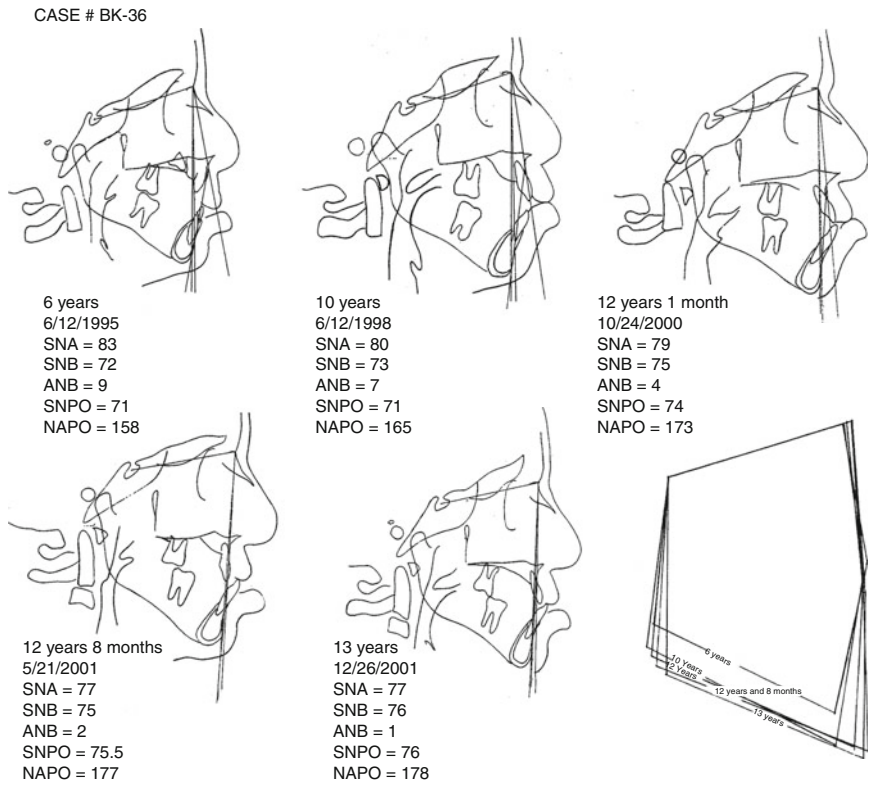

Fig. 21.20 Case BK 36. Bilateral cleft lip and palate. The premaxilla is POPLA retruded. The maxillary anterior teeth were gradually advanced but were still in crossbite. Superimposed cephaloradiographs show that forward mandibular growth enhanced the anterior crossbite; this necessitated the extraction of the lower first bicuspids and the retraction of the incisors. A proper overjet and overbite will be achieved when the mandibular incisors are retracted. A slight maxillary advancement of the incisors may still be necessary

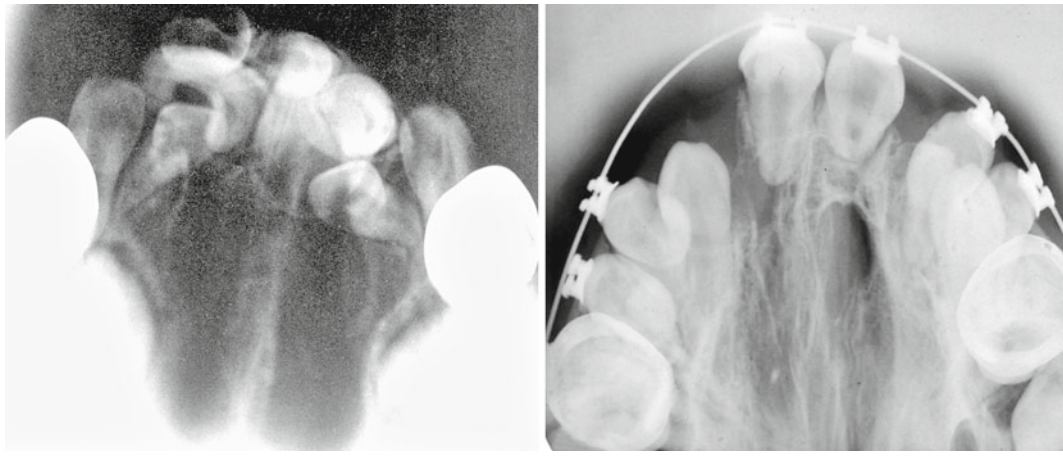

Fig. 21.21 Case BM-BK 36. Incomplete bilateral cleft lip and palate. 3 years of age: Premaxilla has been retruded followed by periosteoplasty. Bony bridge from premaxilla to the right lateral palatal segments is evident. 14 years of age: A bony bridge exists more on the left than right side. 15 years of age: Upper right cuspid is being moved into the lateral incisor space. A secondary alveolar bone graft will be placed in the left lateral incisor space. Comments: Note there is no premaxillary vomerine suture in the 13- and 14-year occlusal x-ray films

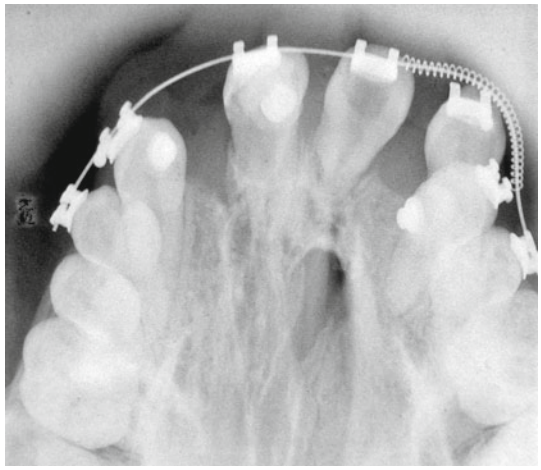

Fig. 21.21 (*continued*)

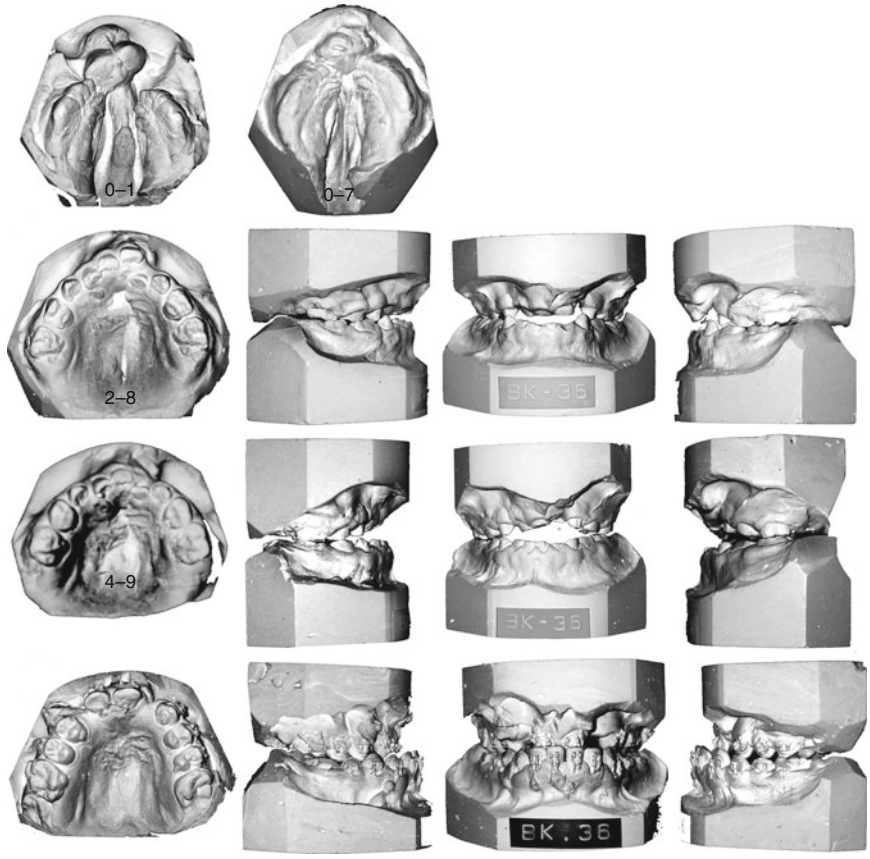

Fig. 21.22 Case BM-BK 36. Incomplete bilateral cleft lip and palate. 0–7: Untreated palatal arch. 2–8: The protruding premaxilla was treated with POPLA. 4–9: Class III malocclusion with an anterior open bite. 14–6: Missing right and left lateral incisors with closure of their spaces. Anterior–posterior arch length is shortened. Even after use of a protraction facial mask, extensive orthodontics was still necessary to attempt to correct the amateur crossbite. Comments: Facial profile evaluation shows a protrusive lower lip with a severe mandibular incisor inclination. The lower first bicuspids were extracted in order to retract the lower anterior teeth. The upper incisors will only be slightly advanced

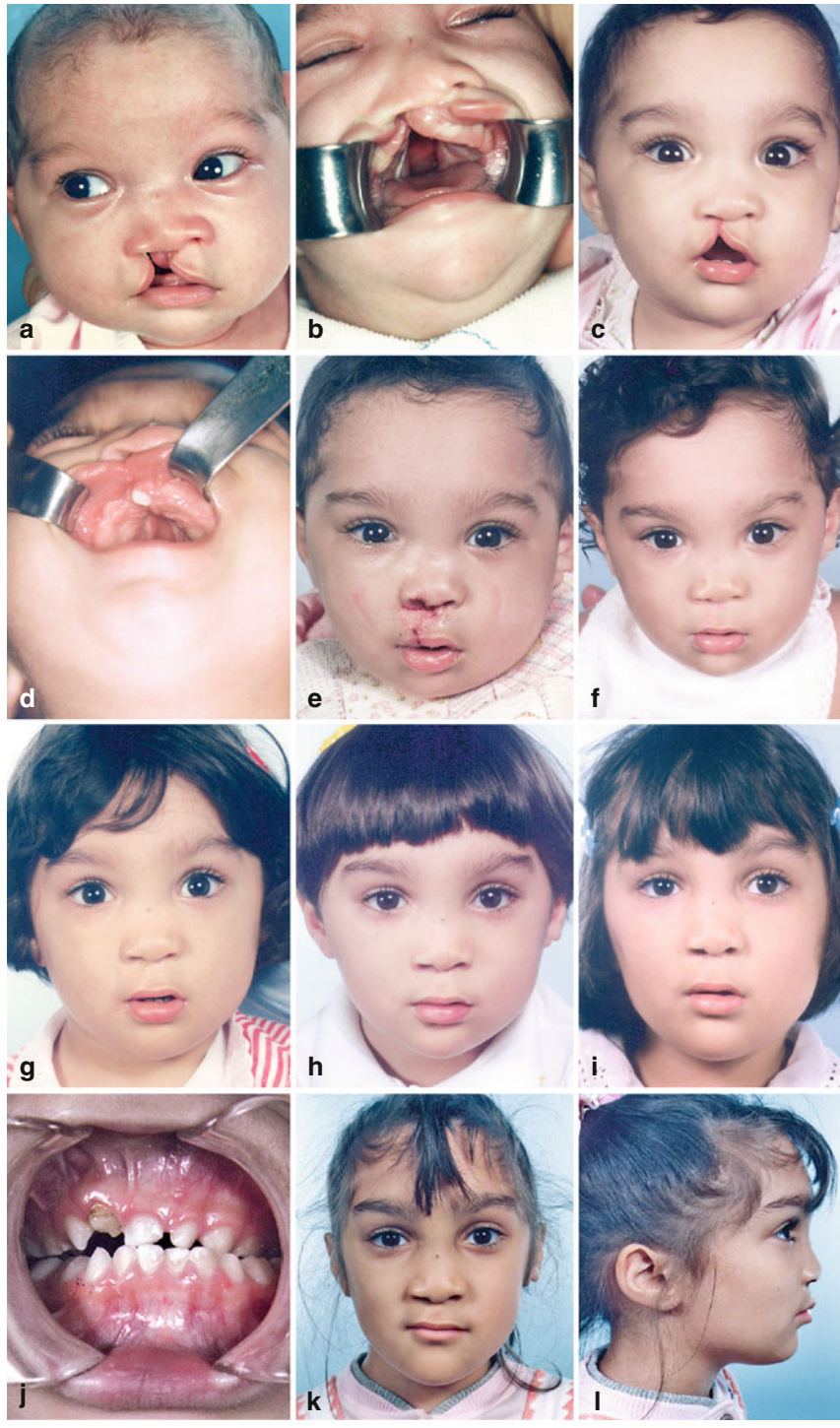

Fig. 21.23 (**a**–**p**) Case CS (AY-45) CUCLP. Latham–Millard PSOT. This case also demonstrates the loss of the lateral incisor space with slight midfacial retrusion at 9 years. (**a**) Newborn 3 months. (**b**) Intraoral view at 3 months. (**c**) 411/42 months after palatal manipulation; periosteoplasty and lip adhesion. (**d**) Looking at the periosteoplasty, palate cleft is open. (**e**) 911/42 months, after rotation advancement lip surgery. (**f**) 1 year and 3 months. Cleft palate closed via modified van Langenbeck procedure. (**g**) 2 years and 1 month. (**h**) 3 years and 8 months. (**i**) 5 years and 8 months(**j**) Intraoral photograph with teeth in an anterior tip-to-tip relationship. (**k, l**) 1 7 years and 8 months; frontal and lateral photographs

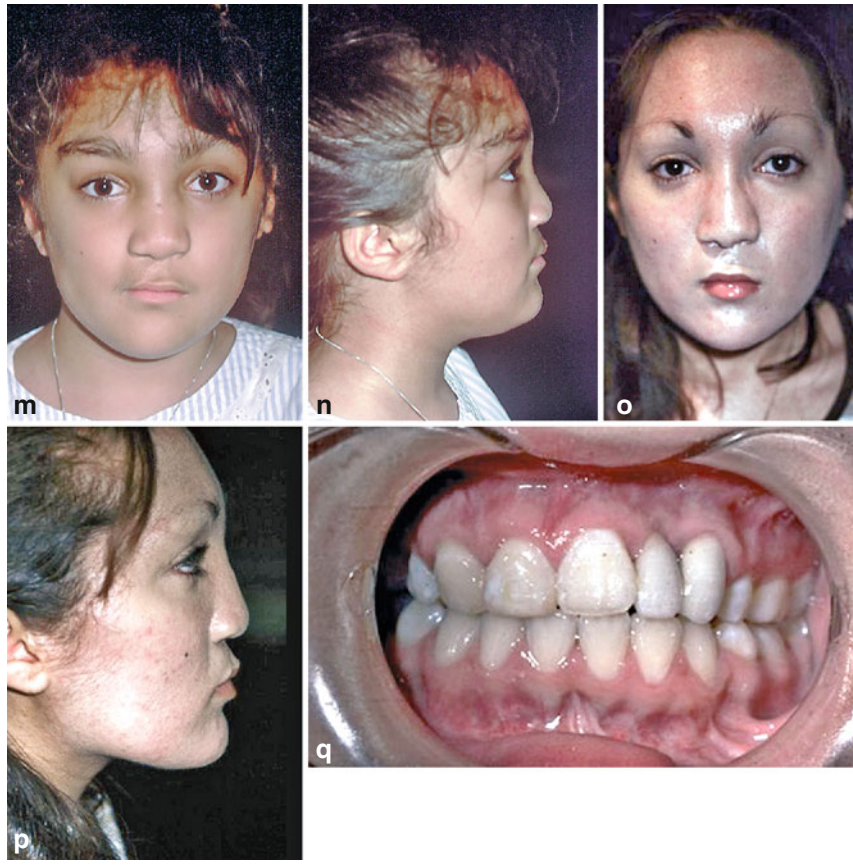

Fig. 21.23 (*continued*) (**m**, **n**) 9 years, 10 months. Comment: The facial aesthetics are most pleasing even with the teeth in a tip-to-tip relationship. It still remains to be seen whether the profile will become more recessive after the pubertal growth spurt. (**o**, **p**) Final facial photographs at 18 years. (**q**) Final occlusion at 18 years old showing good overbite and overjet. The upper left lateral incisor was replaced with PONTIC attached to a bridge. Comments: The results turned out well due to a vertical facial growth pattern

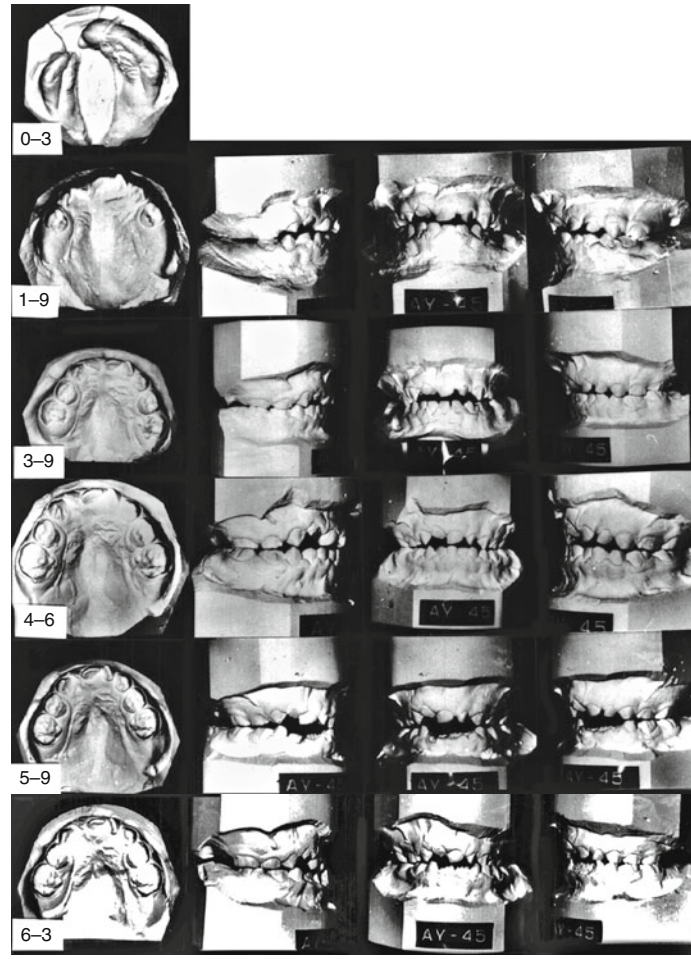

Fig. 21.24 Case CS (AY-45). Serial casts CUCLP. 0–3: newborn. 1–9: After palatal segments were brought together with PSOT. Teeth are slightly in an open bite and in a tip-to-tip relationship. The right lateral incisor space is closed. Ages 3–9, 4–6, 5–9, and 6–3 show the same occlusion relationship. Serial casts CUCLP

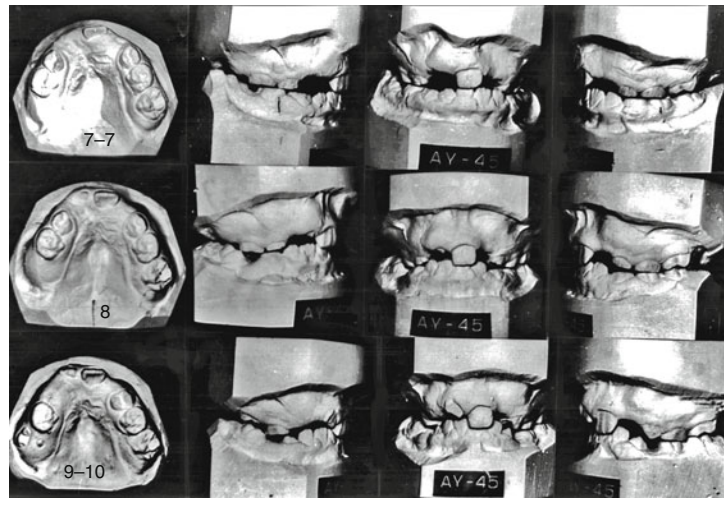

Fig. 21.24 (*continued*) At 7–7, 8, and 9–10: The permanent central incisors are erupting in good overjet–overbite relationship; however, the closure of the right lateral incisor space is due to the maxillary incisors being displaced to the left. Serial casts CUCLP

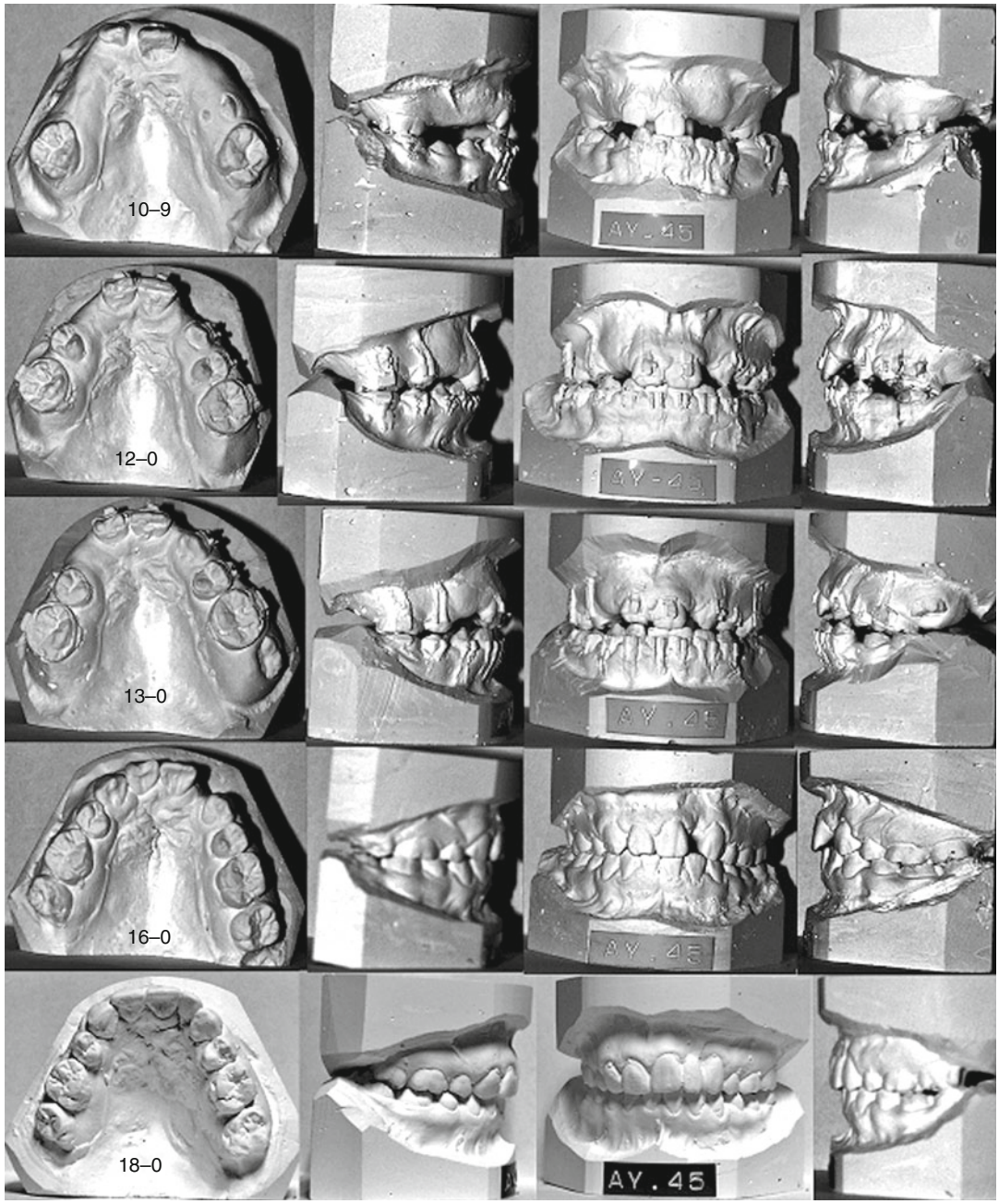

Fig. 21.24 (*continued*) 10–9 to 13–0 Slight anterior crossbite. A protraction facial mask is to be used to advance the right central incisor. The left incisors are rotated and impacted. The lateral incisor is missing. 13 years: The left central incisor and upper first bicuspids were extracted. 16 years: A maxillary retainer with a false tooth was used. 18 years: A fixed bridge with replacement tooth was placed

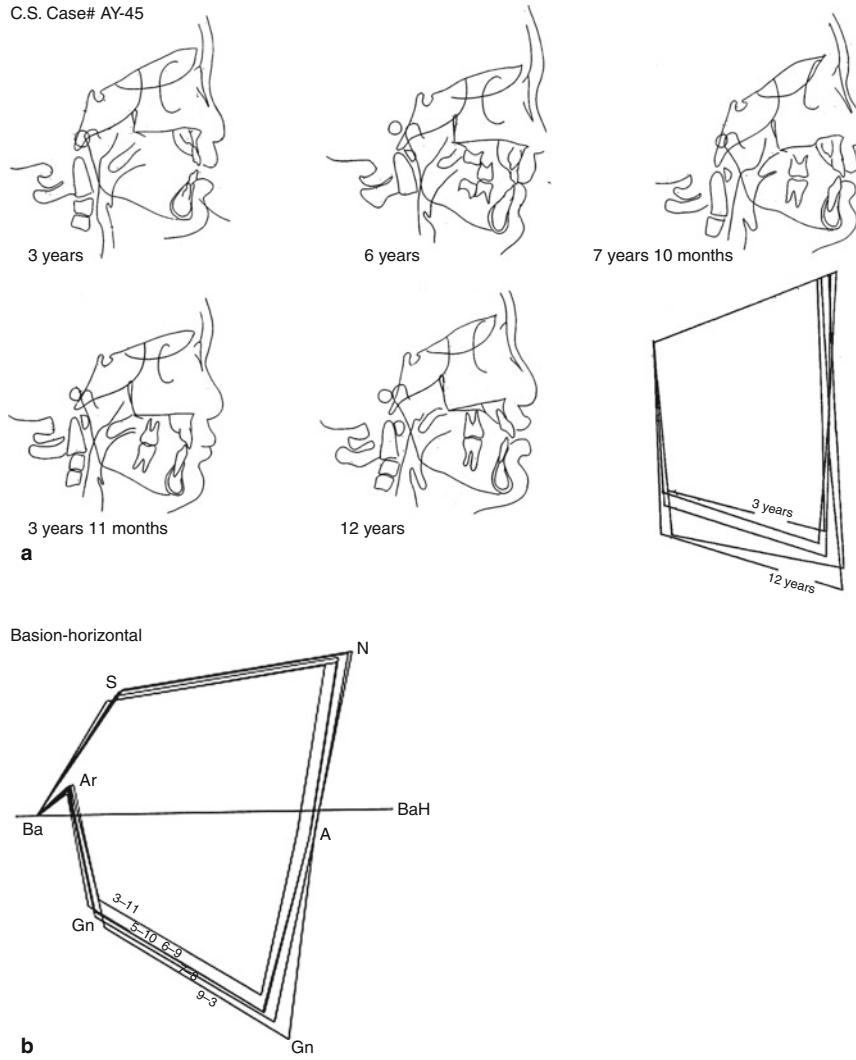

Fig. 21.25 (**a**, **b**) Case CS (AY-45). (**a**) Cephalometric tracings. Lateral cephalometric tracings show the midface is becoming more recessive relative to the anterior cranial base and mandible. (**b**) Superimposed polygons using Basion horizontal procedure. The above finding is verified in this series of superimposed polygons. Although slight forward growth of the midface is evident between 3–11 and 7–8, no further growth is seen between 7–8 and 9–3 years of age

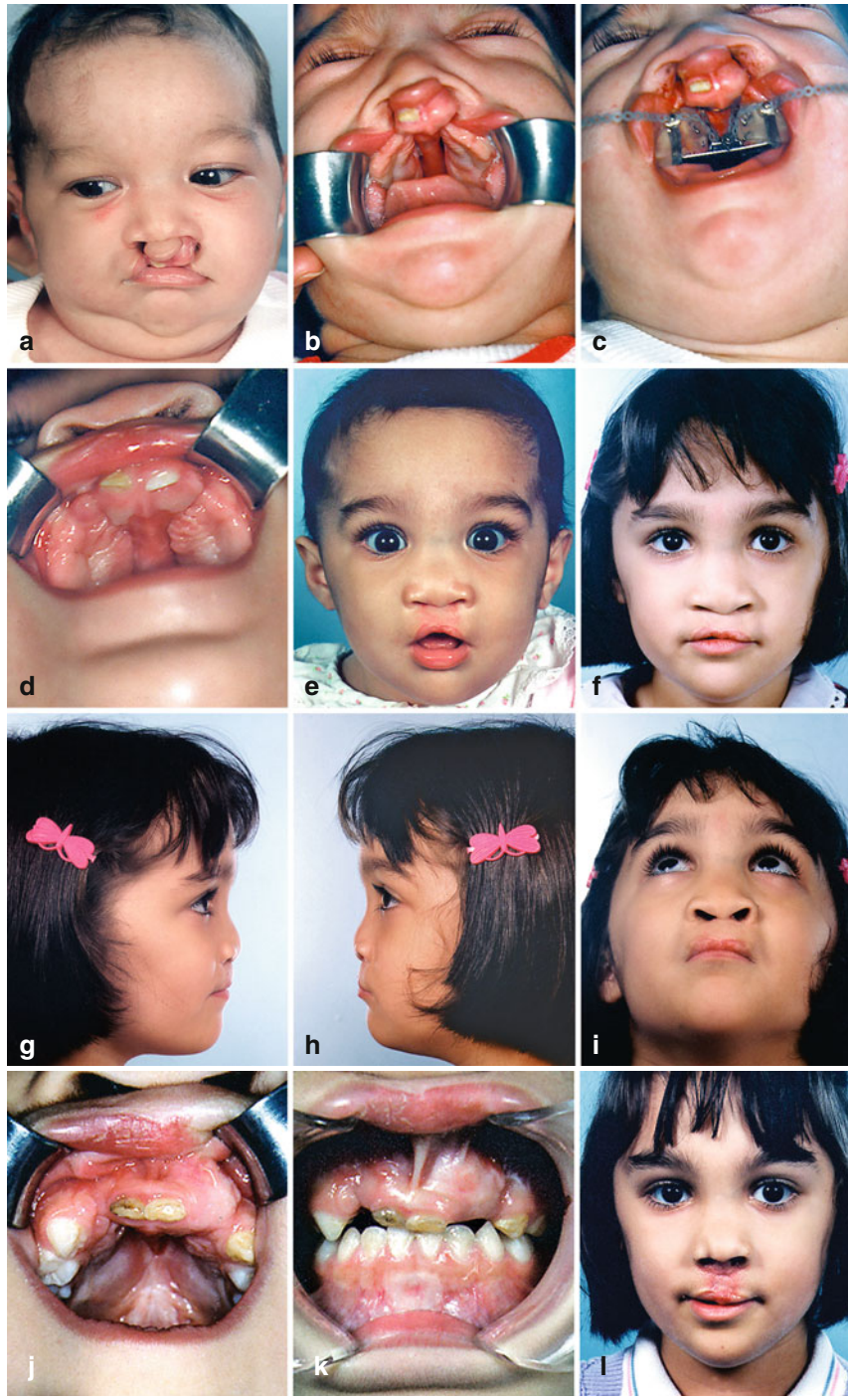

Fig. 21.26 (**a**–**w**) Case AS (AY-46). CBCLP with Millard–Latham PSOT. Retarded midfacial growth in the mixed dentition. (**a**, **b**) Newborn. (**c**) Latham's pinned appliance in place. (**d**) At 6 months. Premaxilla is aligned within the arch. (**e**) At 6 months. (**f**–**h**) 6 years and 6 months. Frontal and lateral face. (**i**) Small columella with depressed nasal tip. CBCLP with Millard–Latham PSOT. (**j**) Good arch alignment. Small palatal fistula. (**k**) Good occlusion

Fig. 21.26 (*continued*) (**l–n**) 9 years. Raised nasal tip. (**o–r**) Facial and dental occlusion. The left deciduous lateral incisor is in crossbite. Retarded midfacial growth in the mixed dentition. CBCLP with Millard–Latham PSOT. (**s–u**) Final facial photographs. (**v, w**) Final occlusion(j) Good arch alignment. Small palatal fistula. (**k**) Good occlusion. (**l–n**) 9 years. Raised nasal tip. (**o–r**) Facial and dental occlusion. The left deciduous lateral incisor is in crossbite. Retarded midfacial growth in the mixed dentition. CBCLP with Millard–Latham PSOT(**s–u**) Final facial photographs. (**v, w**) Final occlusion

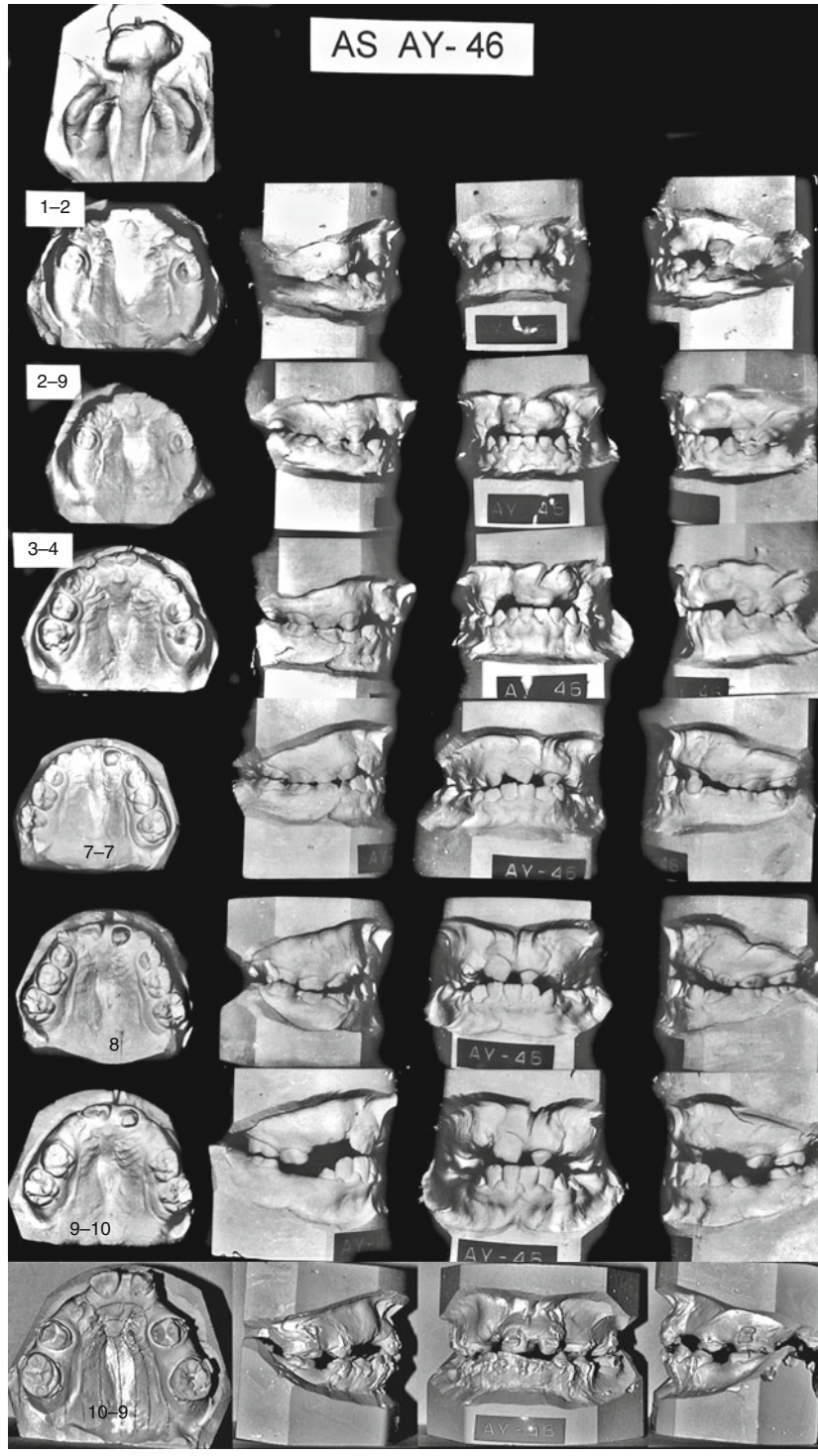

Fig. 21.27 Case AS (AY-46). CBCLP with Millard–Latham PSOT

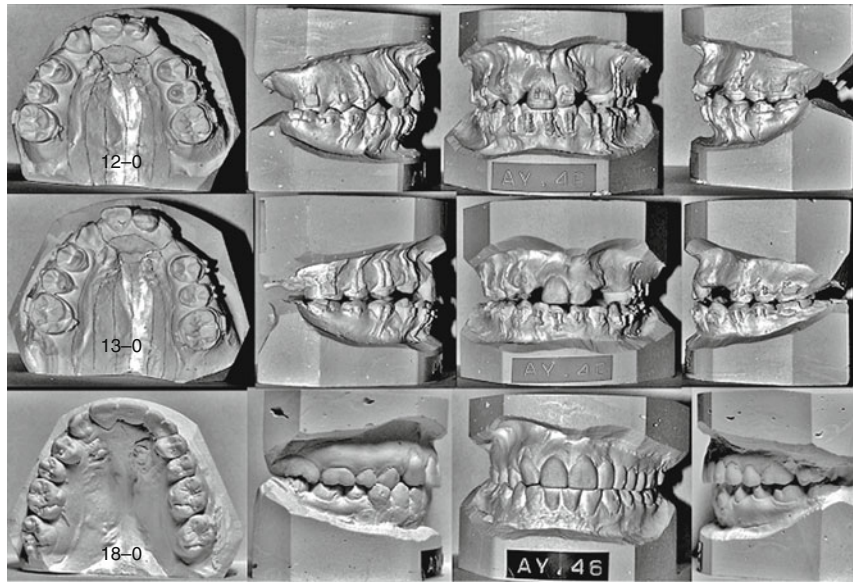

Fig. 21.27 (*continued*) 10–9: Upper left lateral incisor is missing. The left maxillary central incisor is impacted. 18–0: Final occlusion. Treatment plan: (1) Extract upper left lateral incisor and bring in blocked out impacted central incisor. (2) Bring upper right and left cuspids into lateral incisor position. (3) Reshape upper right cuspid. (4) Two-unit bridge to replace upper left lateral incisor

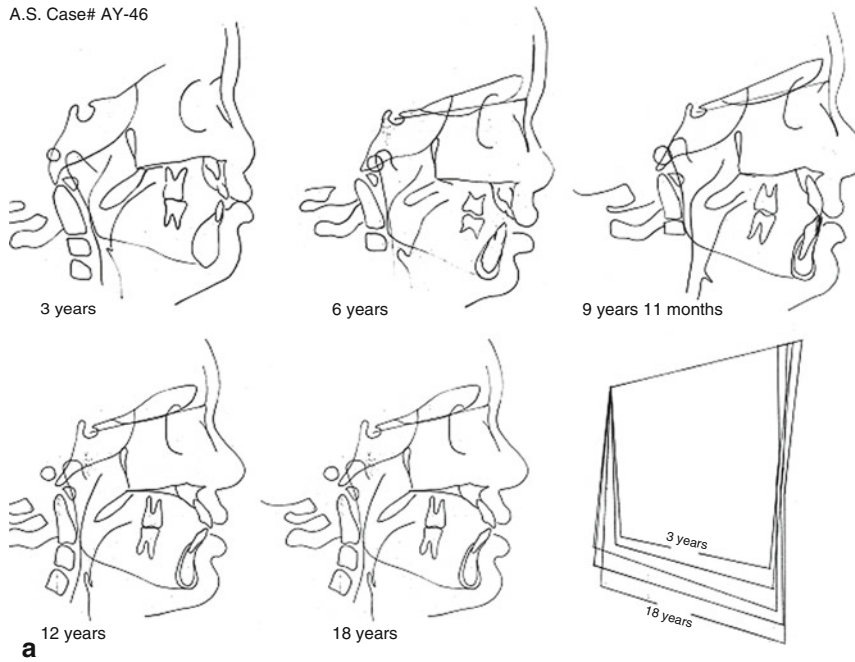

Fig. 21.28 (**a**) Case AS (AY-46). CBCLP with Millard–Latham PSOT. Serial lateral cephalometric tracings and b serial superimposed polygons (Basion horizontal method) show retarded midfacial growth. Fortunately, the vertical mandibular growth pattern complements a reduced midfacial growth, preventing the creation of an anterior crossbite. Comments: Protraction orthopedic forces will be initiated when more permanent teeth have erupted.

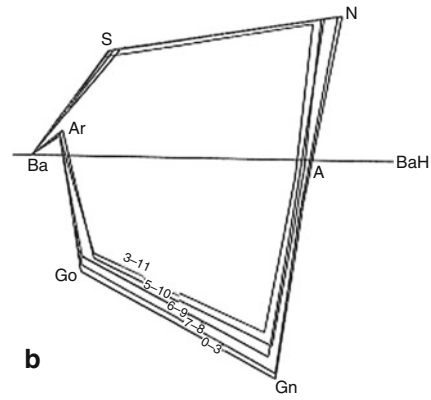

Fig. 21.28 (*continued*) (**b**) CBCLP with Millard–Latham PSOT. This case demonstrates why it was possible to establish good facial aesthetics and occlusion without the need to advance the midface or retract the mandible. Although a slight facial concavity exists, it was still aesthetically acceptable

21.4 Conservative Non-POPLA CUCLP and CBCLP Cases

In non-POPLA CBCLP cases, the protruding premaxilla's overjet is gradually reduced so that it can be easily aligned within the alveolar arch by 7–8 years of age. In some patients, this may spontaneously occur after only 2–3 months of simple palatal expansion. However, depending on the facial growth pattern, some maxillary dental overjet with a convex facial profile may still remain into the mixed dentition stage. In our study, only two cases with a severely protruding premaxilla at birth developed a retrusive Class III malocclusion requiring surgical maxillary LeFort I advancement. This need was mainly due to reduced maxillary growth coupled with a forward-growing mandible. The use of a protraction facial mask in both cases was unsuccessful in correcting the midfacial recessiveness.

With non-POPLA treatments, successful secondary alveolar bone grafts occurred in 70 % of the cases. In most of these cases, the impacted lateral incisor erupted into its normal position within the alveolus, or, if absent, the space was left open for a replacement tooth. If one or both lateral palatal segments are in a Class II relationship and one or both lateral incisors are absent, the cuspid(s) can be substituted for the missing lateral incisor(s). In these patients, the maxillary and mandibular anterior arch forms are congruent at the completion of orthodontic treatment. Anterior arch congruency cannot exist in POPLA cases without extensive orthodontics/surgery.

Flattening of the facial profile, thereby contributing to improved facial aesthetics, occurs gradually as the upper and lower portions of the face grow forward while the midface's forward growth is restrained by the forces created by the united lip. Most significantly, after 2 years of age, 3D measurements show that the premaxilla is in the same ideal position within the maxillary arch, its forward growth having been retarded by the intact lip musculature forces.

Poor facial growth occurs either when the mandible's growth is vertically directed and/or the midface fails to grow in concert with the mandible. Any of these variables can occur in either non-POPLA- or POPLA-treated cases which are not under the surgeon's control and are unpredictable at birth.

The growth analysis of these CBCLP cases demonstrates that increases in A–P maxillary arch length, between the incisal papilla and the first permanent molars, are dependent on the functional integrity of the PVS. Once PVS function is diminished by excessive forces, growth recovery will not return.

Establishing lip muscle continuity soon after birth causes slow molding of both the overexpanded lateral palatal and premaxillary segments

leading to the narrowing of the anterior and posterior palatal cleft spaces. In non-POPLA CBCLP, the posterior-directed pressure of the elastic strip appears to be within physiological limits, and in most cases, reduced pressure permits additional PVS growth. This is seen in the graphic analysis of both superimposed serial lateral cephaloradiographs and from electronically produced 3D tracings of palatal dental casts (Figs. 21.30 and 21.31).

In non-POPLA CBCLP cases, the A–P palatal dimension is greater between birth and 6 years, while with POPLA CBCLP treatment, once the premaxilla is set back, there is no further change in the anteroposterior first molar to incisor dimension due to pressure-caused hemorrhage at the premaxillary vomerine suture leading to fibrosis and then synostosis. Most of the alveolar cleft spaces are obliterated with new bone uniting the repositioned palatal segments.

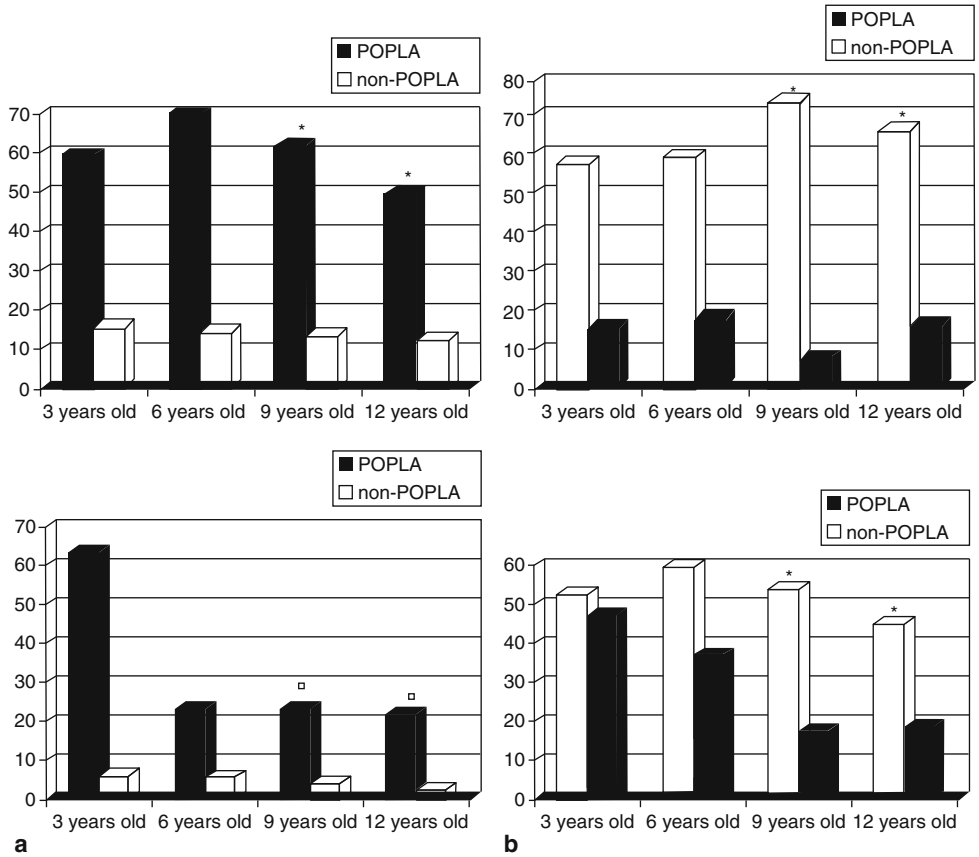

Fig. 21.29 (**a**) The charts report the percentage of children with unilateral cleft lip and palate receiving presurgical or nonpresurgical orthopedics, by crossbite. At every age level, a greater percentage of youngsters treated with presurgical orthopedics developed crossbites than did those treated with nonpresurgical orthopedics (presurgical orthopedics, nonpresurgical orthopedics). This difference existed for both the anterior crossbite (above) and the buccal crossbite (below). Asterisk: Some of the worst crossbite cases in the presurgical orthopedics group were treated by Berkowitz starting at about 9 years of age, thereby reducing the frequency of crossbite in this group beginning at this age level. (**b**) The charts report the percent of children with bilateral cleft lip and palate receiving presurgical orthopedics or nonpresurgical orthopedics, by crossbite. At every age level, a greater percentage of youngsters treated with presurgical orthopedics developed crossbites than did those treated with nonpresurgical orthopedics (presurgical orthopedics; nonpresurgical orthopedics). This difference existed for both the anterior crossbite (above) and the buccal crossbite (below). *Asterisk*: Some of the worst crossbite cases in the presurgical orthopedics group were treated by Berkowitz starting at about 9 years of age, thereby reducing the frequency of crossbite in this group beginning at this age level

Table 21.2 Obtained significant chi-square values ($p < 0.05$) in tests of differences in number of patients having crossbites between presurgical and nonpresurgical orthopedics treatments

	Approximate age of participant				
	3 years	6 years	9 years	12 years	Total sample
Unilateral cleft					
Anterior crossbite	16.8	30.0	10.2	18	77.5
Buccal crossbite	30.9	6.5	6.5	4.8	42.8
Bilateral cleft					
Anterior crossbite	10.3	11.5	24.5	8.5	35.5
Buccal crossbite	49	49	6.5	35	173

In the non-POPLA CUCLP cases studied, only 16 % (8 of 51) are in anterior crossbite at 3 years of age, a condition that is easily correctable by advancing the premaxillary portion of the noncleft larger segment. 60 % of the POPLA cases are anterior crossbites at 3 years (Fig. 21.29a).

Buccal crossbites in both non-POPLA and POPLA treatment series were easily corrected since the type of palatal surgery performed between 18 and 36 months did not inhibit palatal expansion. Only 3 of 51 cases (6 %) had a complete buccal crossbite at 3 years of age (Table 21.1, Fig. 21.29a). The relative increase in percentage of buccal crossbites in POPLA cases at 3 years is significant (60 % at 3 years), because it necessitated more crossbite corrective procedures at 5–6 years of age (Table 21.2, Fig. 21.29a).

21.5 Variations in Palatal Osteogenic Deficiency and Its Influences on Surgical Treatment

The location of palatal bone deficiency is highly variable and can exist to various degrees. In CBCLP, the extent of osteogenic deficiency in the anterior part of the palate can vary in size and shape. In a few of these instances, the palatal cleft space is usually present immediately posterior to the premaxilla, even when the incisors are in good or poor overbite/overjet relationship. In conservatively treated cases, if this large cleft space is present with the incisors in good overbite/overjet, further surgical treatment needs to be evaluated in the mixed dentition or at early adolescence. When there is insufficient mucoperiosteal tissue to surgically close this palatal cleft space, successful secondary alveolar bone grafting will not be possible. In these instances, the only solution is for the posterior palatal segments to be advanced to reduce the cleft space to usable dimensions and immediately followed by secondary alveolar bone grafts (Posnick and Ewing 1990). In our study, this surgery was necessary in only two of 20 non-POPLA cases; both cases were successfully treated by Wolfe using the Posnick procedure.

Should this same form of skeletal palatal deficiency exist in POPLA-treated cases, the premaxilla would be severely retruded so much so that one or both lateral incisor cleft spaces would be closed and ultimately bridged by bone, thus preventing the orthodontic–orthopedic correction of the anterior crossbite. The lack of subsequent anterior maxillary and mandibular arch congruency would prevent the achievement of a good dental overbite/overjet relationship, even after midfacial surgical advancement.

21.6 Correction of Midfacial Deficiencies in Conservatively Treated Non-POPLA Cases

A slight anterior crossbite can be easily corrected orthodontically by using the maxillary protraction mechanics of a facial mask, with or without the extraction of one mandibular central incisor. In the non-POPLA CBCLP cases, it was necessary to surgically advance a retrusive midface in only two patients Figs. 21.30, 21.31, and 21.32. A well-developed, relatively protrusive mandible existed in only one of these children. Fortunately,

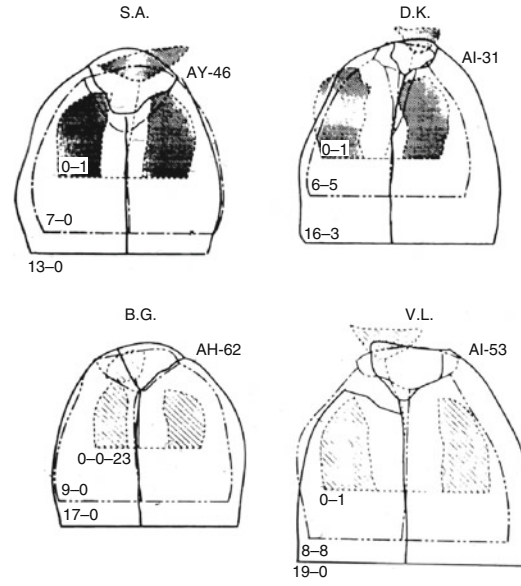

Fig. 21.30 Superimposed palatal cast tracings of the hard palate of four patients were acquired using a 3D electromechanical digitizer. In each tracing, the alveolar ridge is the lateral border. The tracings are superimposed horizontally at the rugae and registered vertically

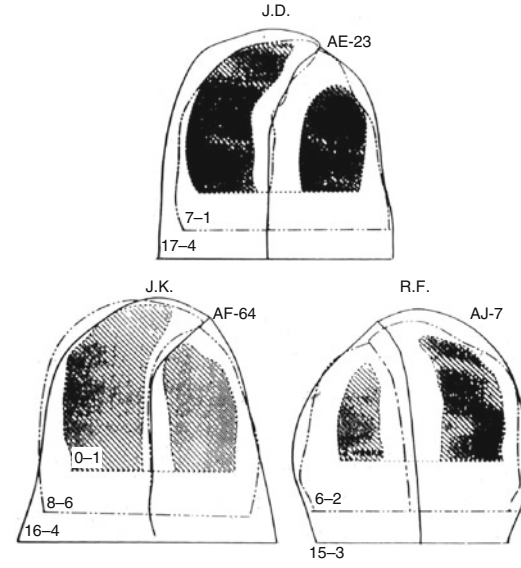

Fig. 21.31 Superimposed palatal cast tracings of the hard palate of three patients were acquired using a 3D electromechanical digitizer. In each tracing, the alveolar ridge is the lateral border. The tracings are superimposed horizontally at the rugae and registered vertically

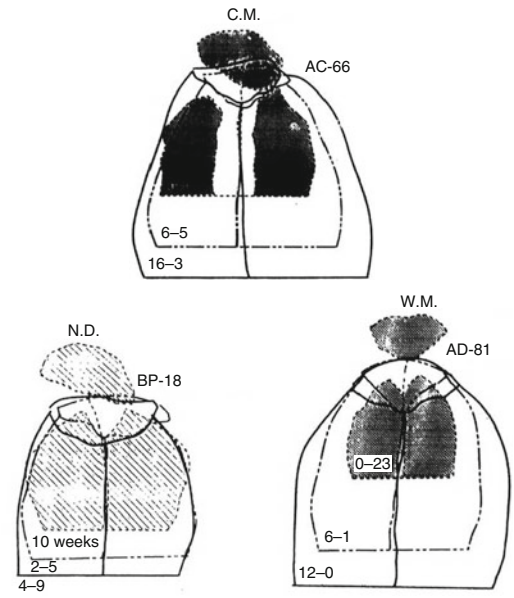

Fig. 21.32 Superimposed palatal cast tracings of the hard palate of three patients were acquired using a 3D electromechanical digitizer. In each tracing, the alveolar ridge is the lateral border. The tracings are superimposed horizontally at the rugae and registered vertically

no long-term speech changes resulted after maxillary advancement in these two instances. Lateral cephalometrics showed good velar length and elevation within a pharyngeal space of average depth. Intraoral examination showed good lateral pharyngeal wall movements as well.

21.7 Similar Presurgical Orthopedics as It Was Utilized in the Past: It Failed Then as It Does Now

During the 1920s and 1930s, the cleft surgeon's treatment philosophy exemplified by Brophy (1923) was to repair the cleft defect by establishing "normal" anatomical palatal form soon after birth, through the use of external and internal palatal compression techniques. The first priority then was to improve facial aesthetics, followed in turn by good dental function and speech. Unfortunately, the Brophy procedure led to extensive midfacial deformity and was eventually discontinued.

Due to the benefit of long-term facial and palatal growth records, many if not most surgeons and speech–language pathologists have recognized the extent to which time, or more precisely growth, serves as their ally or enemy. However, some surgeons are still endeavoring to devise a procedure that can be used during the first 2 years for all CBCLP and CUCLP cases, hopeful that good facial growth will follow.

In recent years, serial documentation of the natural evolution of postnatal facial and palatal development of children with complete bilateral and unilateral cleft lip and palate has yielded important objective data – data that help explain the dynamics of facial skeleton and palate growth under the influence of various surgical procedures. This knowledge has greatly improved the ability of surgeons and orthodontists to develop physiologically based concepts that lead to successful long-term treatment outcomes.

This study supports Gillie's (Millard, personal communication) belief that time is both the surgeon's ally and most trenchant critic. It further supports the thesis that no single surgical procedure performed at birth is suitable for all cleft types and faces since there is great variation in palatal osteogenesis and in facial growth patterns. Staged treatment based on the individual patient's facial assets and deficits must be the controlling factor in designing therapy.

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Part X

Midfacial Orthodontic/Orthopedic and/or Surgical Changes

Protraction Facial Mask

22

Samuel Berkowitz

22.1 Protraction of the Maxilla Using Orthopedics

Children with complete unilateral and bilateral cleft of the lip and palate are usually at risk for poor facial growth. They are prone to developing midfacial retrusion related to maxillary hypoplasia or growth retardation secondary to excessive palatal scarring. Usually, this results in an anterior dental crossbite or severely rotated maxillary incisors which may occlude in a tip-to-tip relationship with the mandibular incisors. Depending on the age of the patient and the extent of midfacial maldevelopment, some of these early problems can be corrected using midfacial orthopedic protraction forces which increase growth at the circumaxillary sutures as they are repositioned anteriorly (Fig. 22.1). When all else fails, midfacial surgery is available.

Some of the earlier work in this field, which encouraged a rethinking of the use of orthopedic forces for the correction of midfacial retrusion, includes Hass (1970), Delaire (1971), Delaire et al. (1972, 1973, 1976, 1978), Irie and Nakamura

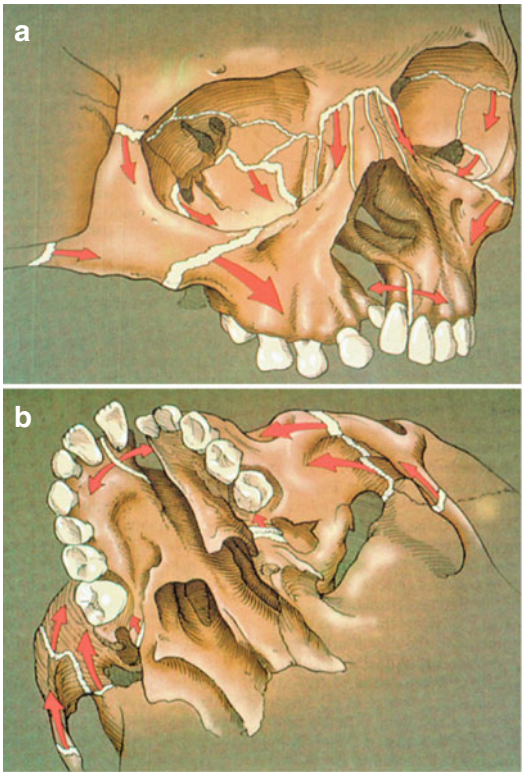

Fig. 22.1 (**a**, **b**) Protraction of the maxillary complex using orthopedic forces. The maxilla articulates with nine bones: two of the cranium, the frontal and ethmoid, and seven of the face, viz., the nasal zygomatic, lacrimal, inferior and nasal concha, palatine, vomer, and its fellow of the opposite side. Sometimes it articulates with the orbital surface and sometimes with the lateral pterygoid plate of the sphenoid. Illustration showing how protraction forces applied to the maxilla depends on the disarticulation and growth at all the dependent sutures (Courtesy of E. Genevoc)

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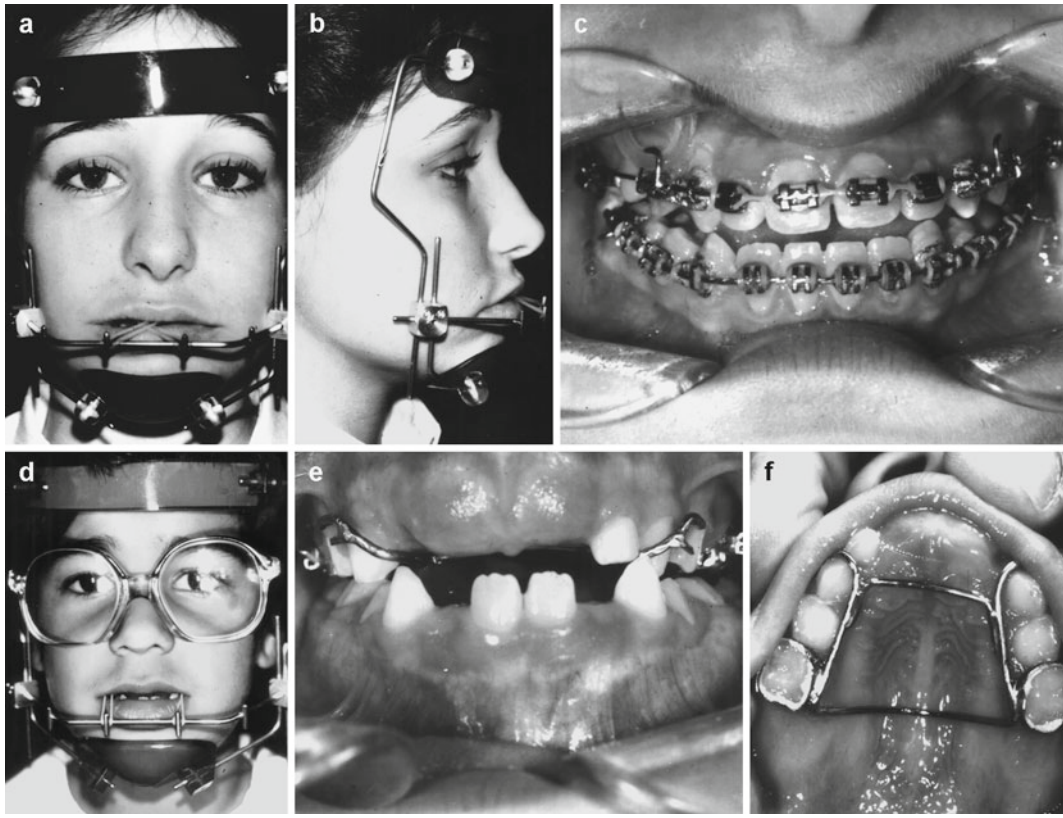

Fig. 22.2 (**a**) Frontal and (**b**) lateral views of a Delaire-style protraction facial mask. Padded chin and forehead rests distribute reaction forces of 350–400 g per side equally to both areas. Elastics are attached to hooks placed on the arch wire between the cuspids and lateral incisor. (**c**) Intraoral view of edgewise rectangular arch with hooks for protraction elastics. (**d–f**) Delaire-style protraction facial mask used with a fixed labial-palatal wire framework. Elastic forces of 350–400 g per side can still be used with this intraoral framework

(1974), Ranta (1988), Subtelny (1980), Friede and Lennartsson (1981), Sarnas and Rune (1987), Berkowitz (1982), Tindlund (1989), Nanda (1978), and Molstad and Dahl (1987). More recently, this area has been influenced by the work of Tindlund et al. (Irie and Nakamura 1974; Ranta 1988; Subtelny 1980; Delaire et al. 1978; Friede and Lennartsson 1981; Sarnas and Rune 1987; Berkowitz 1982; Tindlund 1989; Tindlund and Rygh 1993; Nanda 1978; Molstad and Dahl 1987) and Buschang et al. (1994).

Earlier attempts by Kettle and Burnapp (1955) in which anteriorly directed extraoral forces were derived from chin caps were relatively unsuccessful. Facial mask therapy seems to offer better control and a wider range of force application.

In many cases, in the mixed dentition, palatal expansion using fixed orthodontic appliances was applied simultaneously with protraction to correct a bilateral crossbite and create a more favorable condition for midfacial growth and development.

Prior to the use of orthopedic forces, many standard orthodontic treatments designed to move the dentition to correct a class III malocclusion due to midfacial retrusion in the absence of mandibular prognathism failed. Orthodontic forces applied to the teeth by class III elastics would not displace the maxilla; at best, they would flare the maxillary incisors without creating an adequate incisor overbite and axial inclination. This treatment was found to be unsatisfactory and soon fell out of favor.

Since 1975, Berkowitz has been using a modified protraction facial mask originally popularized by Delaire et al. (1972) (Figs. 22.2, 22.3, and 22.4). It has been very successful in controlling the direction of protruding forces

without causing severe sore spots on the chin or forehead. He has found that protraction forces do not modify the direction of mandibular growth as Delaire et al. (1972) claimed, but by increasing midfacial height, the mandible is repositioned downward and backward with growth to make the patient's maxillary retrusion appear less evident.

Protraction forces (350–450 g per side) must be intermittent (the mask is worn only for 11 h per day) and directed downward and forward from a hook located mesial to the maxillary cuspids. Pulling downward from the molars should be avoided because it will tilt the palatal plane downward in the back by extruding the molars and thus opening the bite. When the midfacial height is deficient, protraction forces need to be modified to increase vertical as well as anterior growth. This is done by using more vertically directed elastic forces.

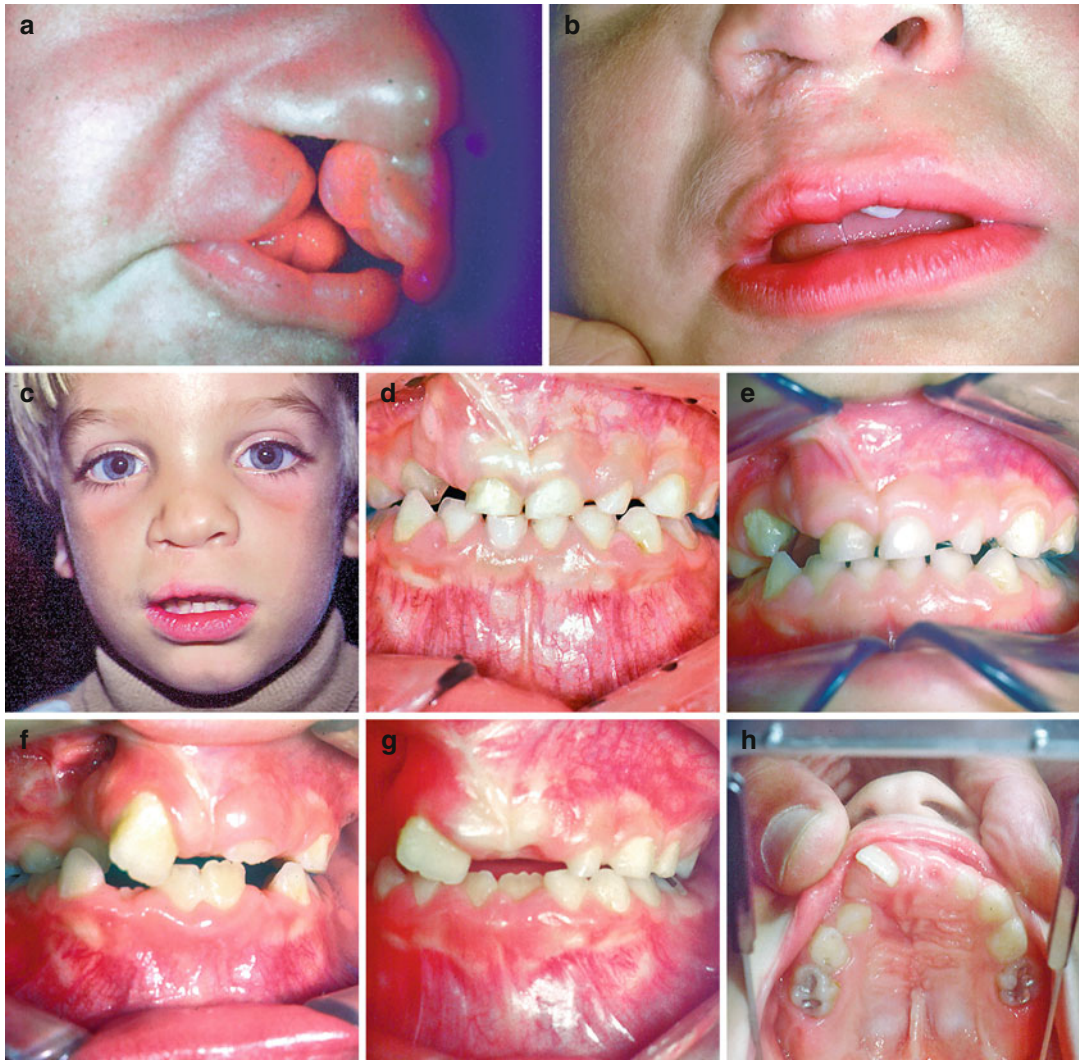

Fig. 22.3 (**a**–**x**) Case BB (WW-62). Maxillary protraction in a UCLP. (**a**) Complete unilateral cleft lip and palate. (**b**, **c**) Lip and nose after surgery. (**d**) Cuspid crossbite of the lateral cleft segment at 5 years of age due to mesioangular rotation of the palatal segment. (**e**) Buccal occlusion after expansion using a quad helix expander. (**f**, **g**) 6 years of age. Note relapse of cuspid crossbite due to failure of using a palatal arch retainer. (**h**) Palatal view showing good arch form

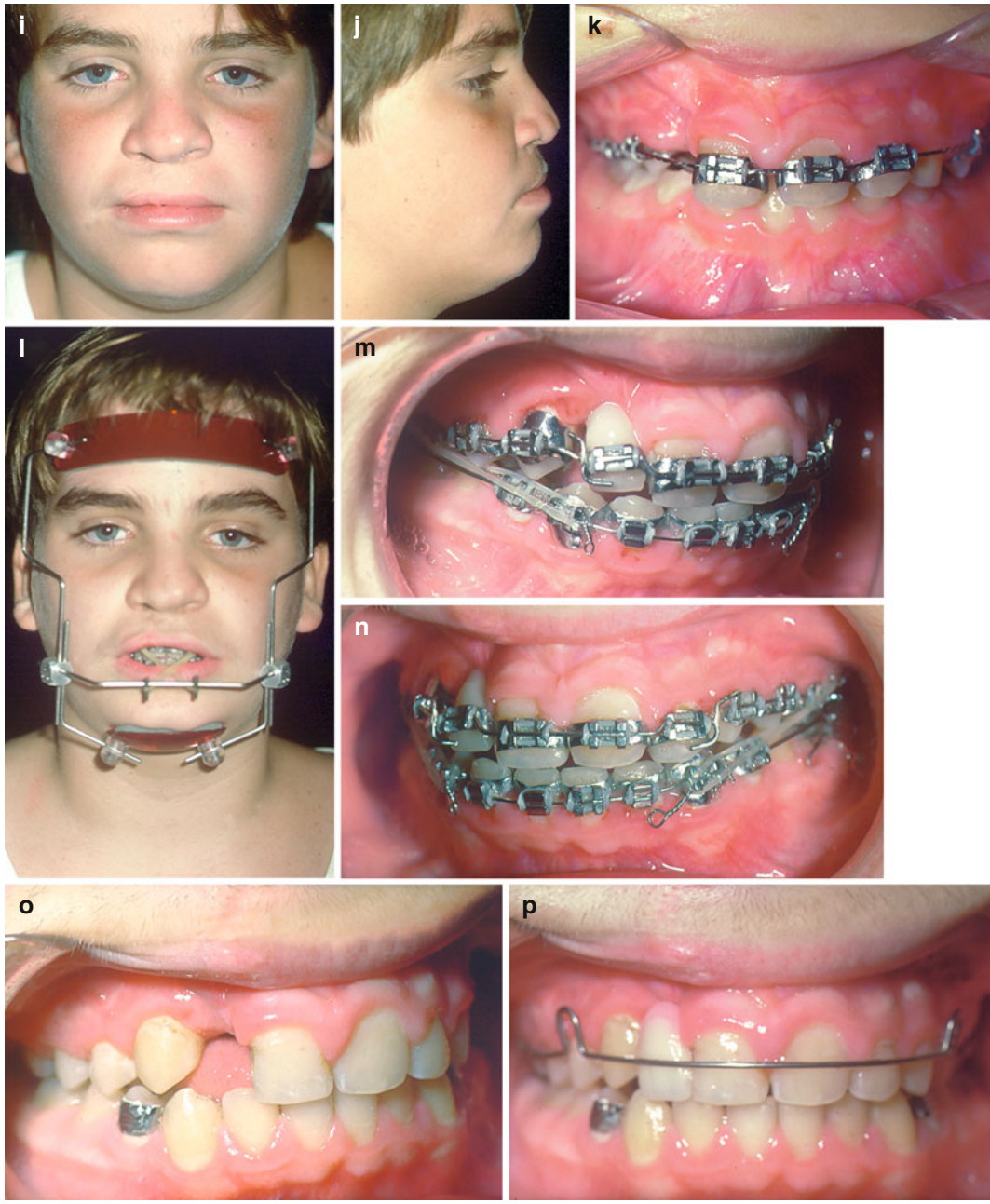

Fig. 22.3 (*continued*) (**i**, **j**) Facial photographs at 8 years. (**k**) Orthodontic alignment of incisors prior to secondary alveolar bone graft. (**l**) Protraction facial mask with elastics. (**m**, **n**) class III elastics used to maintain tension at circumaxillary suture during the time not wearing protraction forces. (**o**) Occlusion after orthopedic–orthodontic forces. Lateral incisor space regained. (**p**) Removal retainer with lateral incisor pontic

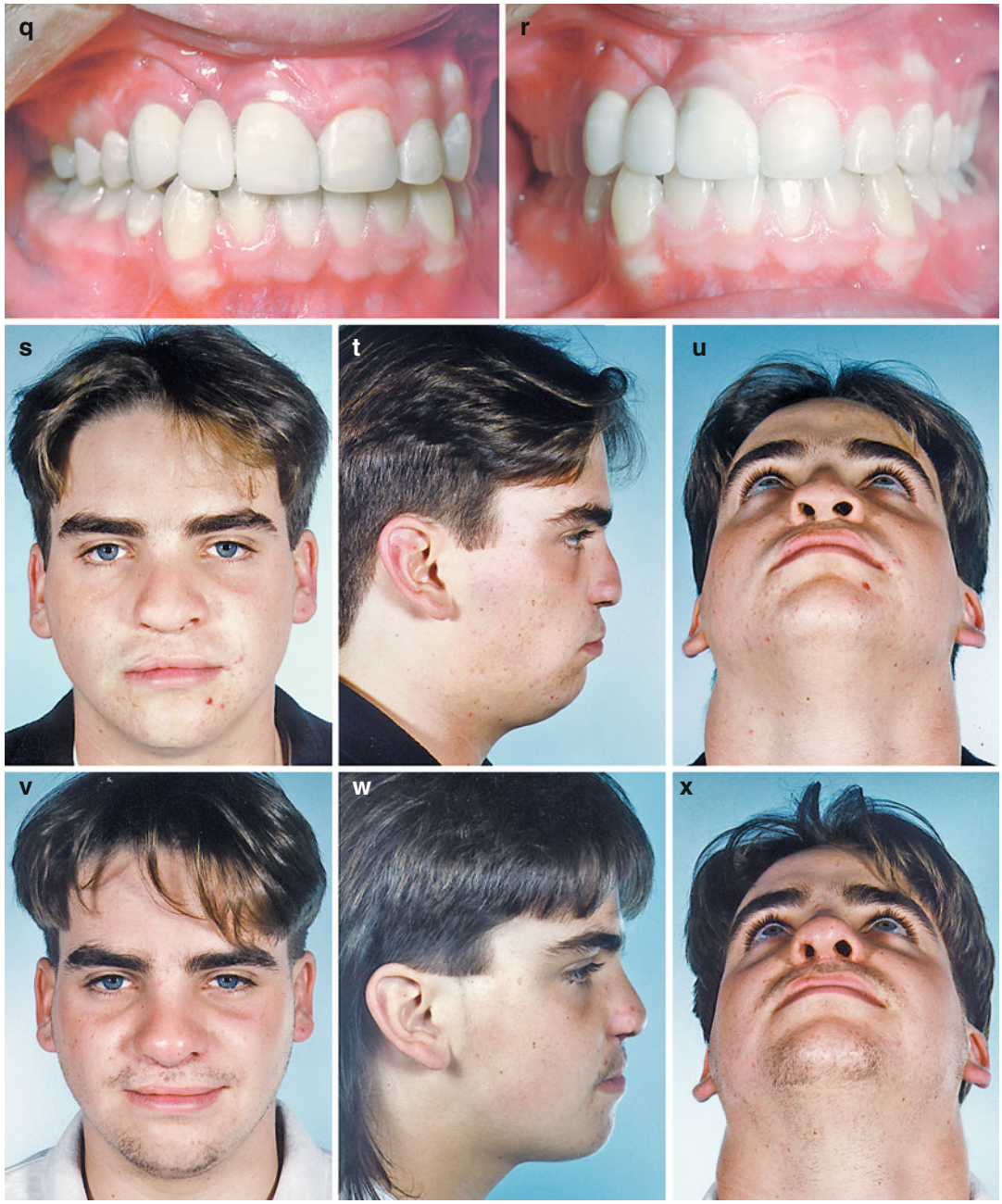

Fig. 22.3 (*continued*) (**q**, **r**) Fixed bridge at 18 years of age replacing missing lateral incisor and stabilizing maxillary arch form. (**s**–**u**) 17 years prior to nose–lip revision. (**v**–**x**) Facial photos at 19 years, showing good facial symmetry after revision

Berkowitz has found 350–450 g of force per side to be adequate in most instances, but there are rare instances when the elastic force needs to be reduced to prevent sore spots at the chin point. Friede and Lennartsson (1981) have used protraction forces between 150 and 500 g per side. Ire and Nakamura (1974) have used 400 g per side, Roberts and Subtelny (1988) 670 g, Sarnas and Rune (1987) 300–800 g, and Tindlund et al. (1993a, b) 350 g per side. Unfortunately, when

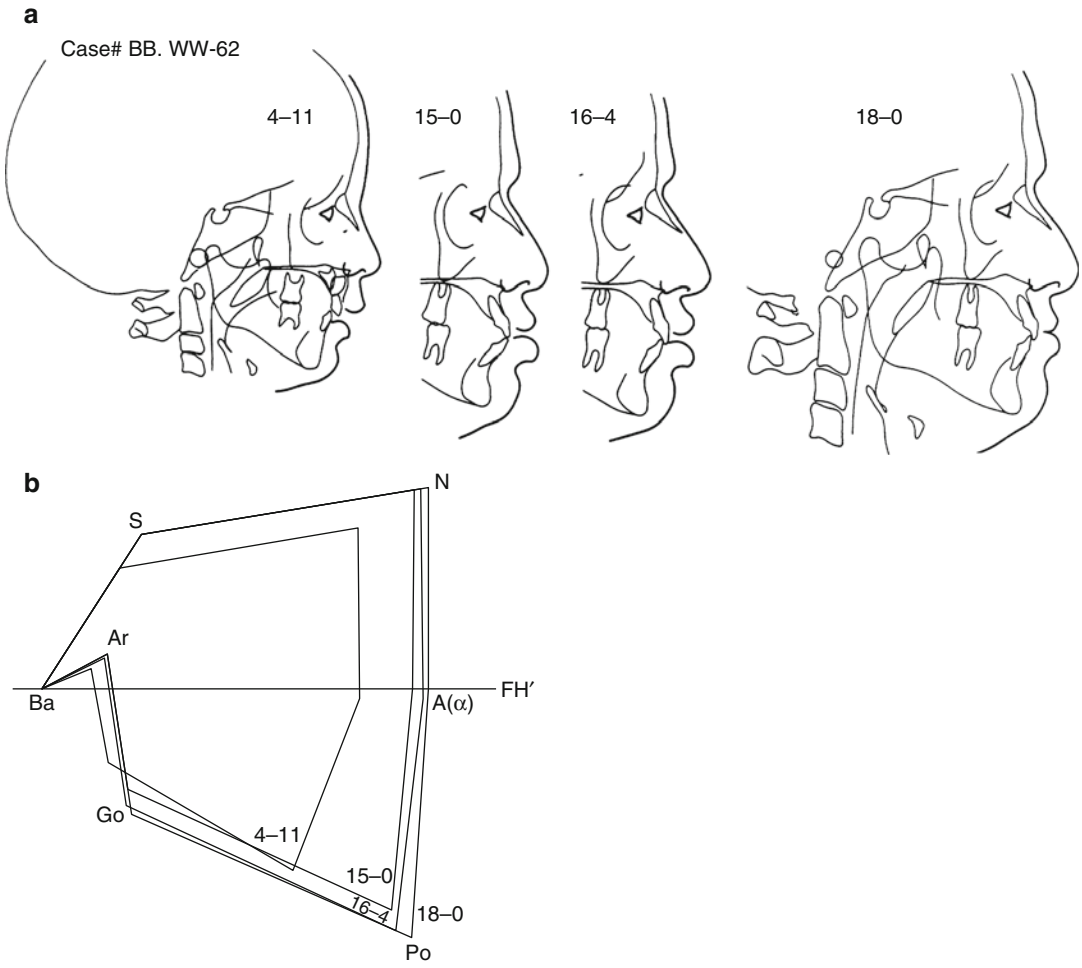

Fig. 22.4 Case BB (WW-62) (**a**) Lateral cephalometric tracings and superimposed polygons (Basion horizontal method) for case BB (WW-62) show an excellent facial growth pattern. (**b**) The midfacial growth increment between 15 and 16–4, when the protraction facial mask was used, increased midfacial protrusion to a greater degree than that which would have occurred normally

performed in the mixed dentition, treatment time may extend into years because of the need to keep pace with mandibular growth. If this is the case, treatment should be divided into intermittent periods not to exceed 6 months at a time with a break for 1 month between periods. Following this formula, the patient will usually remain cooperative.

Although Berkowitz has been successful in using strong elastic forces with labile-lingual appliances during the deciduous dentition, he recommends starting treatment at 7–8 years of age when all of the maxillary incisors can be bracketed and a rectangular edgewise arch with lingual root torque used as Subtelny (1980) suggested. The torqued rectangular arch will carry the incisor roots forward, moving skeletal landmark point "A" anteriorly, which prevents stripping of the alveolar crest with subsequent incisor flaring. The arch wire needs to be tied back so that it does not slide anteriorly, tipping the incisor, rather than moving the entire maxilla forward orthopedically.

Tindlund et al. (1993a, b) conclude that early transverse expansion of the maxilla together with protraction orthodontic treatment is an effective method for normalizing maxillomandibular

discrepancies in cleft lip and palate patients. The average age at the start of treatment was 6 years, 12 months, and the average duration of treatment was 13 months. Significant changes were achieved due to anterior movement of the upper jaw and a more posterior positioning of the lower jaw resulting from clockwise mandibular rotation.

Berkowitz also found that the combined use of palatal expansion and protraction forces before the pubertal growth spurts to be a more efficient means of gaining orthopedic advancement than the use of protraction forces alone. He speculates that the expansion forces possibly disarticulate the circumaxillary sutures, thus allowing the maxillary complex to be carried downward and forward more easily.

Delaire et al. (1976) and Subtelny (1980) have stated that orthopedic forces applied to the entire maxillary complex are more likely to be effective in younger children.

Berkowitz's clinical experience supports the recommendation by Abyholm et al. (1981) and Bergland et al. (1986) (1) that a rigid fixation of the advanced maxilla should be maintained for at least 3 months after bone grafting, and (2) the use of protraction forces. This is necessary to help reduce the tendency to relapse created by the surrounding soft tissue of the lip, muscles, and skin.

Many patients with a complete bilateral cleft lip and palate have a protruding premaxilla until 10 years of age or older, but after the postnatal mandibular growth spurt, the maxillary incisor teeth may be in crossbite. Protraction orthopedic forces with anterior crisscross elastics upright and reposition the premaxilla forward, perhaps by inducing bone growth at the premaxillary-vomerine suture. Fixed retention is always necessary to control the improved incisal overbite–overjet relationship at least until secondary alveolar bone grafting is done.

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23 Protraction Facial Mask for Early Correction of Midfacial Retrusion: The Bergen Rationale

Rolf S. Tindlund

23.1 Early Rehabilitation

Optimal rehabilitation of a child with cleft lip and palate (CLP) involves the achievement of ideal speech, facial aesthetics, and dental occlusion. Dentofacial appearance is of major importance for the development of a child's self-esteem (Stricker et al. 1979; Shaw 1981; Shaw et al. 1985). Early adolescence is a time of change and uncertainty and a period of special importance because negative self-esteem developed in these years is likely to be retained into adulthood (Alsaker and Olweus 1986; Alsaker 1990). Therefore, early rehabilitation is of major importance.

Obtaining an optimal treatment result in complete clefts of the lip and palate is dependent on the prevailing treatment philosophy, clinical skills, and the interaction of the Cleft Lip and Palate (CLP)/Craniofacial Team. The orthodontist is mainly concerned with the achievement of normal long-term facial growth and development, based on his or her ability to recognize, prevent, and treat dentofacial anomalies.

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Quality assurance and the cost-effectiveness ratio are important factors that need to be considered in the systematic delivery of health care. Quality assurance focuses on the achievement of the goals and the quality of overall team management based on the usage of accepted physiological principles. Treatment results are not always predictable because patients differ in their facial growth patterns and the nature of the palatal defect, requiring individualized orthodontic treatment plans depending on the developing malocclusion. This philosophy is at variance with the generally held orthodontic strategy, which is to postpone all orthodontic intervention until the permanent dentition (Semb and Shaw 1993). The relative low cost of utilizing interceptive orthopedics at an early age, due to the need for infrequent visits with uncomplicated mechanics, is a reasonable option for the early improvement of dentofacial appearance. An additional bonus to performing treatment at this period is that patients develop a positive attitude toward themselves and parents to their child's future status.

The specific aim of this chapter is to present a CLP treatment program that incorporates interceptive orthopedics in faces with midfacial retrusion and demonstrate how a fixed orthopedic–orthodontic appliance system may be used for both transverse widening as well as the protraction of the maxilla. Interceptive orthopedics is discussed with respect to treatment timing and anticipated clinical results, reviewing the limitations, and criteria necessary in case selection to improve long-term prognosis.

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23.2 Midfacial Retrusion in CLP Patients

Irrespective of the method used in primary cleft repair and the surgical skill of the operator, a certain number of patients will show an unfavorable growth pattern. Even if one plastic surgeon performs all surgery utilizing the same procedures, and the same treatment protocol, individual outcomes may vary from excellent to unsatisfactory. The variable results reflect individual differences in craniofacial type and growth patterns on which the cleft maxilla is superimposed. Also, one needs to consider acquired variables, such as the degree of prenatal maxillary hypoplasia and facial asymmetry in cleft embryo pathogenesis and detrimental growth deviations related to the surgical procedure and skill of the surgeon.

Midfacial retrusion may be due to underdevelopment and/or relative posterior positioning of the upper jaw to the mandible. The maxillary growth deficiency usually is three-dimensional, resulting in a shortening of maxillary length and a decrease in width and height. Midfacial retrusion is more often seen in unilateral cleft lip and palate (UCLP) patients (Ross 1987; Semb 1991a; Tindlund and Rygh 1993a; Tindlund et al. 1993a, 1994), whereas in bilateral cleft lip and palate (BCLP) the initially prominent premaxilla becomes less protrusive over time, achieving an almost ideal incisor overjet–overbite relationship by the late teenage years (Semb 1991b).

Patients with an underdeveloped maxilla, which results in skeletal and/or dentoalveolar discrepancies, often show anterior and/or posterior crossbites with a concave soft-tissue profile. Since 1977 the treatment protocol of Bergen Cleft Palate–Craniofacial Center has included an interceptive orthopedic treatment phase designed to correct anterior and posterior crossbites during the deciduous and early mixed dentition and to obtain optimal alveolar cleft space to enhance tooth eruption and alveolar development. This would ultimately lead to a favorable functional dental occlusion and create better conditions for attaining normal midfacial growth and development (Tindlund and Rygh 1982, 1993a, b; Tindlund 1987, 1989, 1994a, b; Tindlund et al. 1993a, b, 1994).

23.2.1 Anterior Crossbite

Anterior crossbite (incidence about 3–5 % in Scandinavia) may be found in all facial types – prognathic, orthognathic, and retrognathic – in combination with varying degrees of hypo- or hyperplasia of the jaws. Different sagittal skeletal jaw configurations, some with deep or skeletal open bite, may be associated with excessive dentoalveolar mandibular proclination or maxillary retroclination along with lack of sufficient dental space in the upper arch. Guyer et al. (1986) found skeletal maxillary retrusion in two thirds of noncleft class III children. This is of great therapeutic interest since orthopedic influence seems to be more effective in influencing the sutures of the maxillary complex than in restraining mandibular growth (Thilander 1965; Delaire et al. 1972, 1976; Graber 1977; Ishii et al. 1987; Tindlund 1987, 1989, 1994a, b; Rygh and Tindlund 1982; Tindlund et al. 1993a, 1994). However, the long-term differential diagnosis between mandibular excess and maxillary retrusion is difficult to determine before puberty (Tweed 1966; Ruhland 1975; Vego 1976; Schulhof et al. 1977; Campbell 1983; Guyer et al. 1986). For this reason, children with the appearance of midfacial retrusion and anterior crossbite may benefit from an early interceptive orthopedic treatment phase. The need for orthognathic surgery is usually determined after puberty, taking facial appearance as well as dental occlusion into consideration. A family anamnesis of anterior and posterior crossbite is of particular interest in the CLP population because maxillary hypoplasia is a common finding in these patients.

23.2.2 Orofacial Function

Optimal orofacial function with adequate incisor relationship in the primary dentition is an important determinant for normal growth and development of the anterior part of the maxilla. There is a generally accepted belief that form and function are mutually dependent. This interaction in facial clefts is important because malfunction has been shown to negatively influence facial growth. Thus, the facial characteristics of a noncleft child who is a mouth breather may show some similarities in appearance to the typical CLP patient (Tindlund

et al. 1993b, 1994). In CLP, midfacial retrusion due to deficient midfacial growth may be aggravated by increased nasal airway resistance, low and forward posture of the tongue, and lack of sufficient stimuli from proper masticatory forces. Early widening of the upper jaw enhances nasal respiration (Linder-Aronson and Aschan 1963; Harvold et al. 1972, 1973; Haas 1973), while permitting the tongue to assumes a more normal elevated position within the mouth (Ohkiba and Hanada 1989). Direction of eruption and the final position of teeth are closely associated with the development of the alveolar process, which in turn is dependent upon the number, size, and location of teeth (Harvold 1954; Subtelny 1957; Ogidan and Subtelny 1983). Early orthopedic treatment which includes transverse expansion and anterior protraction of the maxillary complex will improve the dimensions of the nasal as well as the intraoral space, permitting the tongue to elevate and assume a normal posture within the vault space, thus breaking the vicious circle of poor function leading to poor form with growth.

23.3 Principles of Orthopedic/Orthodontic Treatment in CLP Patients

The orthopedic/orthodontic CLP treatment protocol in Bergen utilized since 1977 is based on selective periods of active, controlled, efficient treatment followed by intervals of fixed retention, as recommended by American Cleft Palate–Craniofacial Association (1993). The easily obtained acceptance of the need for patient cooperation along with an excellent cost-effectiveness assessment ratio supports the use of this philosophy of treatment. The following orthodontic treatment phases should be considered as viable options for the individual patient:

1. Presurgical maxillary orthopedics (0–3 months, used in a few cases only).
2. Interceptive orthopedics (6–7 years, about 20 % of cleft patients) which involves transverse expansion and protraction (facial mask).
3. Alignment of maxillary incisors prior to secondary alveolar bone grafting.
4. Secondary alveolar bone grafting of the cleft alveolar process.
5. Conventional orthodontics in the permanent dentition is always necessary.
6. Dental adjustments dependent on prosthodontic or orthognathic surgery needs (17–19 years).

Individualizing the timing and sequencing of treatment is essential due to the wide range of skeletal malformation associated with dental malocclusions. It is of utmost importance to individualize each treatment plan and to revise this plan at different ages of dental and skeletal development, all of which is conveniently based on a diagnosis-related checklist.

23.3.1 Checklist for CLP Orthopedic/Orthodontic Treatment Objectives

23.3.1.1 Presurgical Orthopedics

The plastic surgeon aims at to obtain optimal function and appearance and avoid the need for extensive revisionary surgery by using proven surgical techniques that result in a minimum of scarring and palatal growth impairment. In some cases, presurgical orthopedics can help the plastic surgeon to unite anatomical structures with a minimum of force and stress to the tissue. Individual decisions are made by the plastic surgeon:

- Reposition severely displaced maxillary segments
- Reduce width of very wide clefts
- Improve symmetry of nose and upper jaw

(Only used in extreme cases, and in some treatment philosophies, this stage is not necessary.)

23.3.1.2 Interceptive Orthopedics

Transverse expansion followed by anterior protraction of the upper jaw should only be utilized in cases with anterior and/or posterior crossbite with midfacial retrusion. Treatment should be instituted early enough to allow the permanent incisors to erupt spontaneously into a normal overjet and overbite occlusion (Fig. 23.1):

- Eliminate anterior crossbite
- Eliminate posterior crossbite
- Create optimal space to permit spontaneous eruption of the incisors
- Improve nasal respiration
- Improve tongue placement

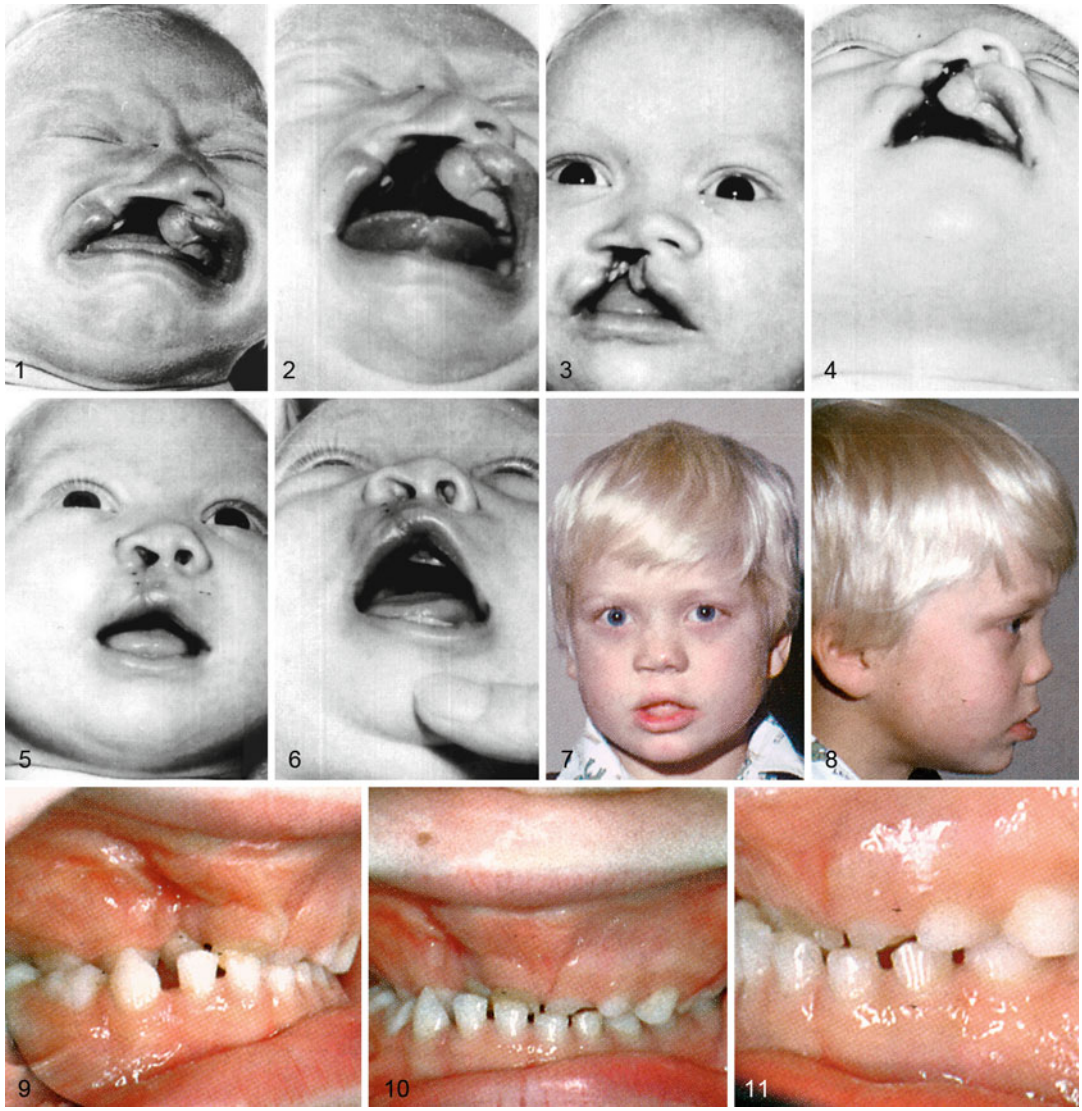
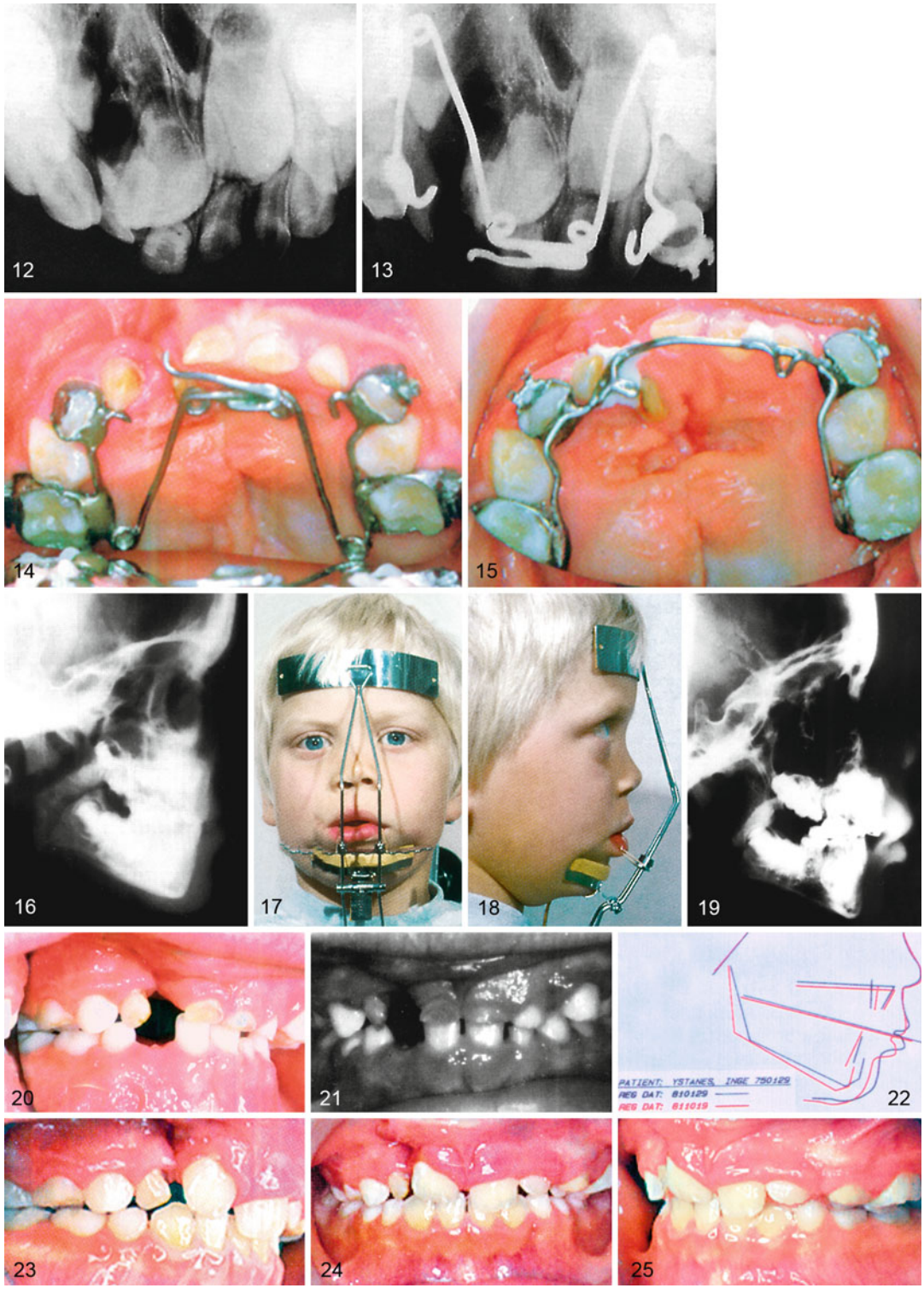


Fig. 23.1 Complete UCLP, category 2A. (1–2) At birth, January 1975; (3–4) after presurgical orthopedics; (5–6) lip closure at age 3 months; (7–12) at 6 years moderate anterior and unilateral posterior crossbites with a slight concave profile; (13–27) interceptive orthopedics from age 6 years includes transverse expansion for 3 months using a quad-helix, (14) followed by protraction for 6 months using a facial mask, (17–18) and retention using a fixed palatal archwire (15) to encourage spontaneous eruption of upper permanent incisors into normal position. A nice dental smile was achieved without early orthodontic alignment of the upper incisors (28–33). Alveolar bone grafting at 10.5 years. Two right upper lateral permanent incisors erupted into the cleft area; (34) facial profile at 12 years (35–41); conventional orthodontics at 13.5 years lasting for 18 months. The two upper second bicuspids were missing, and the supernumerary right upper lateral permanent incisor was removed; (42–48) dental occlusion at 18.5 years; (49–50) cephalometric graphic analysis at 6, after interceptive orthopedics at 15 and at 18 years; (51–53) facial appearance at 15 years; (54–59) and facial appearance at 18.5 years



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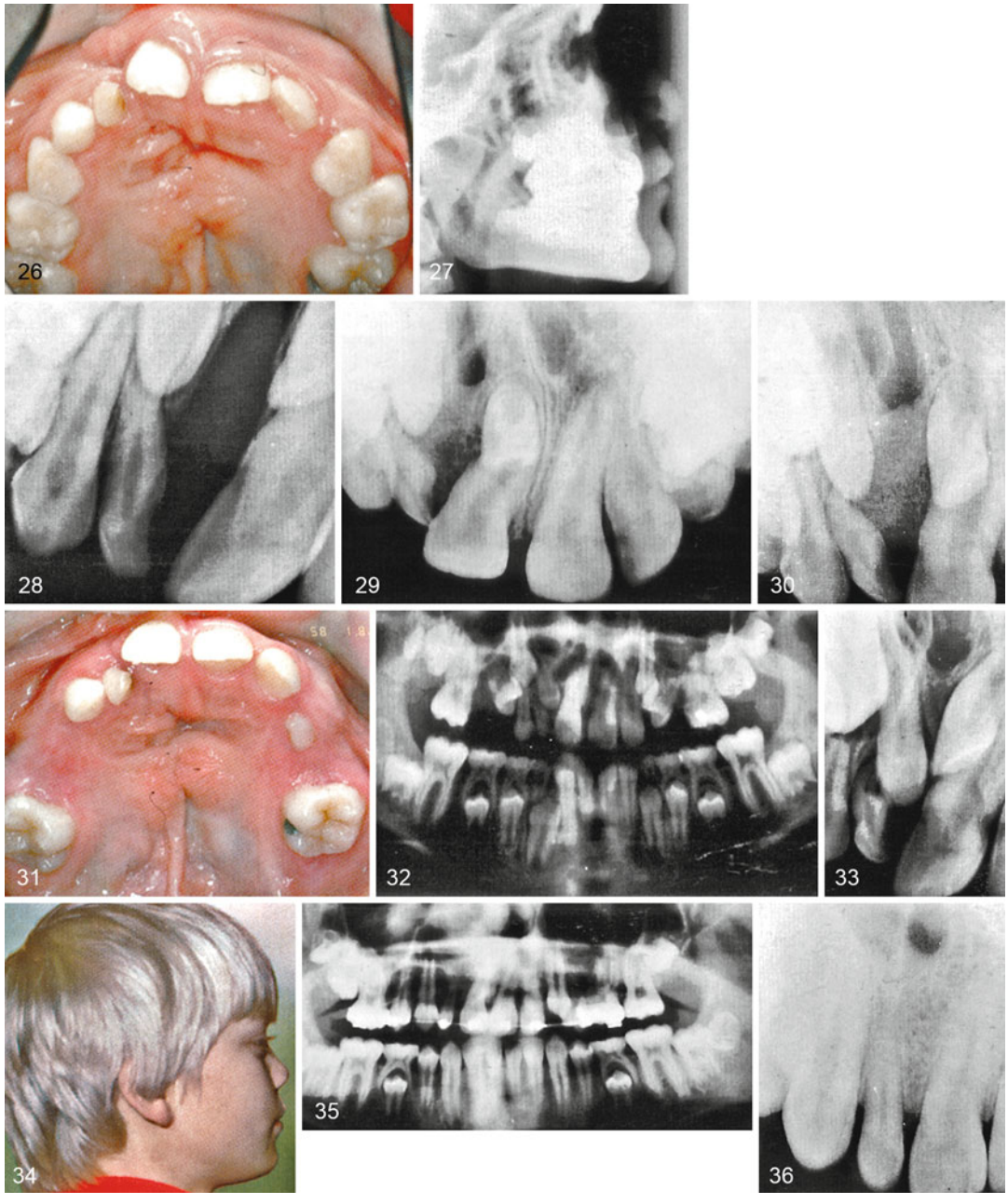

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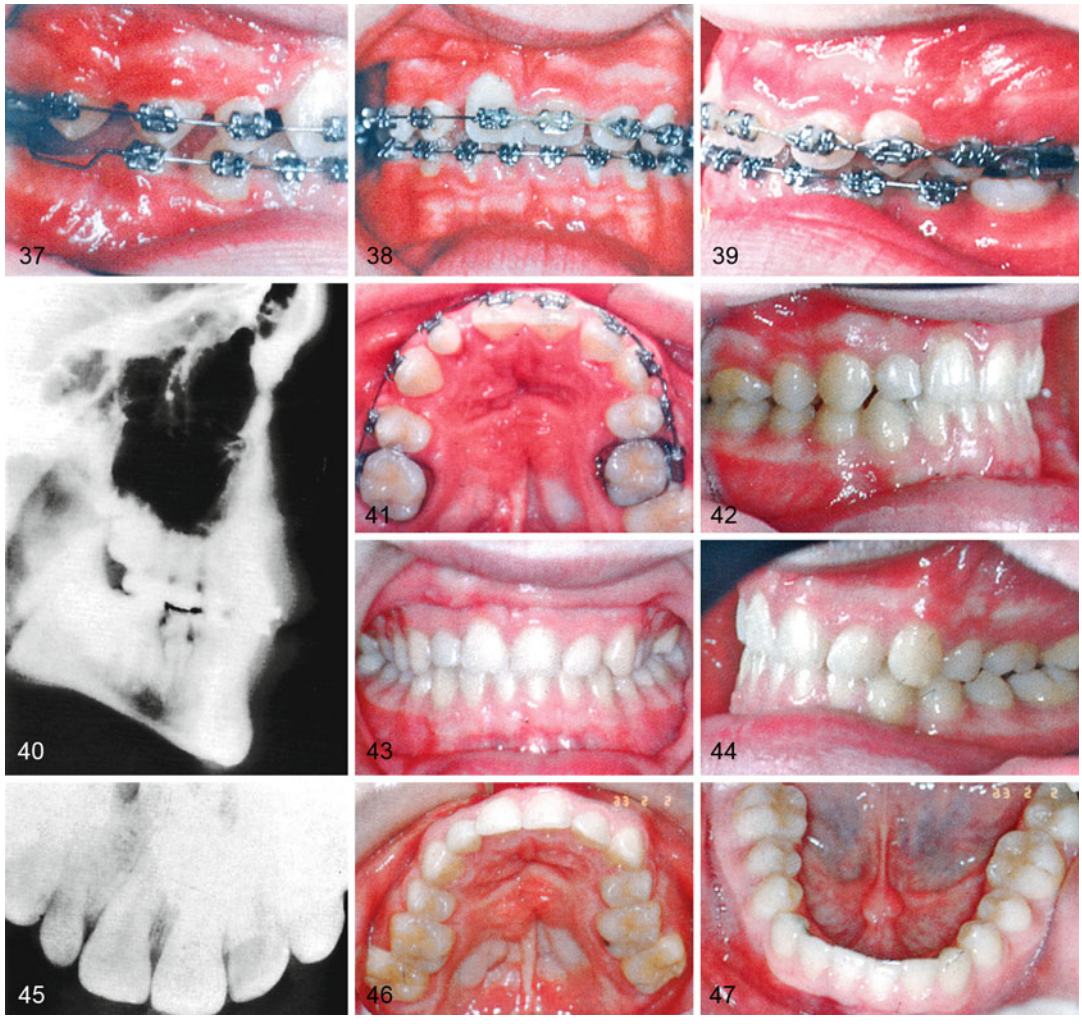

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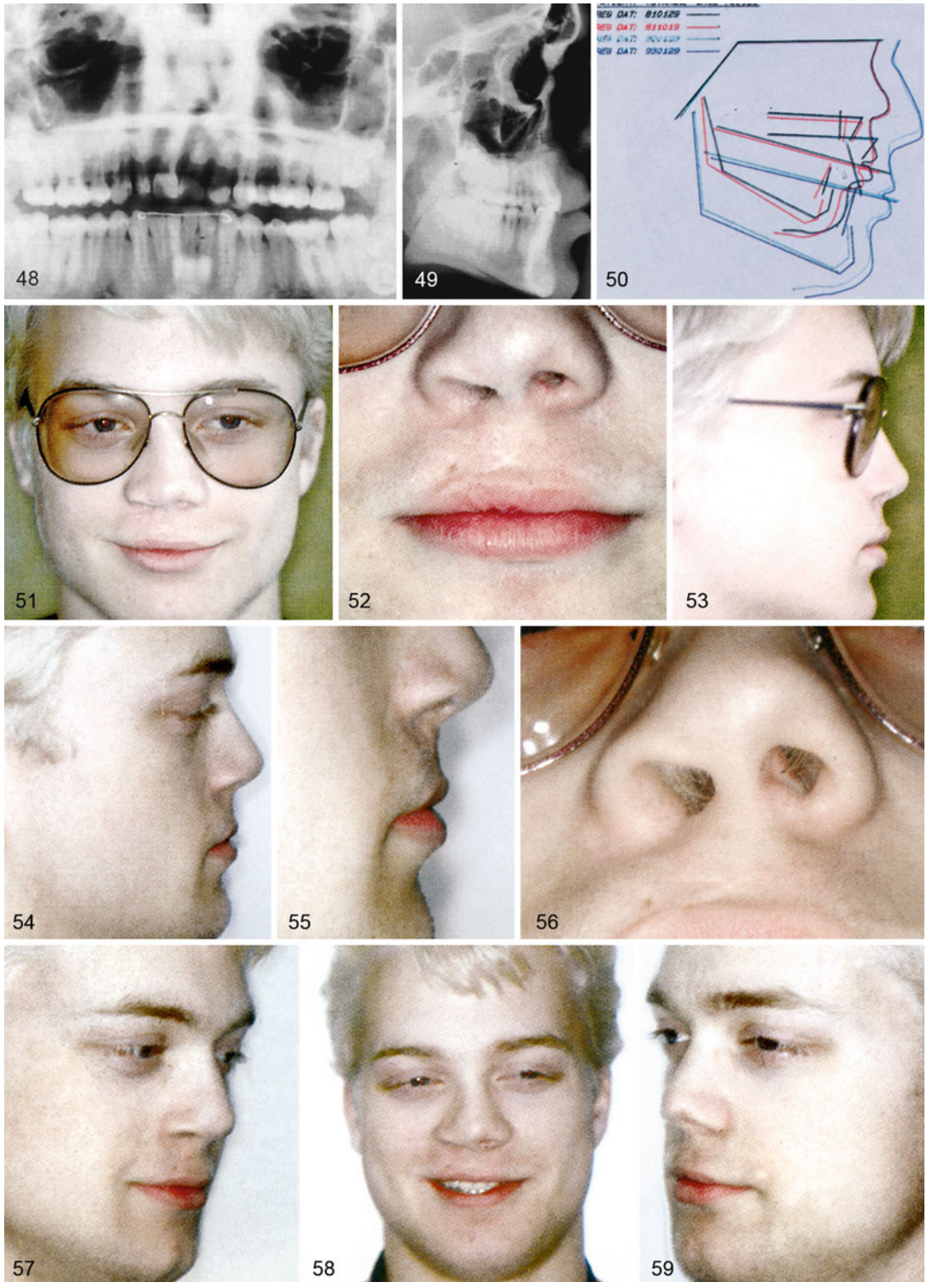

Fig. 23.1 (*continued*)

23.3.1.3 Alignment of Maxillary Incisors

In spite of achieving optimal dental space after transverse expansion, the permanent incisors often erupt rotated and retruded, tipped, or retroclined, placing them in crossbite. After transverse expansion, alignment of the permanent incisors is easily performed, giving the child a nice dental smile equal to that of his or her classmates (Fig. 23.2; in Fig. 23.1, incisor alignment was not needed):

- Straightening of malpositioned incisors
- Creating an optimal aesthetic incisor relationship to the facial midline

23.3.1.4 Secondary Alveolar Bone Grafting

The use of primary periosteoplasty at age 3 months was rejected after introduction of secondary bone grafting (Bergland et al. 1986). It is usually performed between 8 and 11 years of age with the orthodontist selecting the appropriate age:

- Eliminate remaining bony clefts and improve bony support of contiguous teeth
- Enhance orthodontic closure of the missing incisor space in the cleft area
- Stabilize of separated jaw segments
- Close oronasal fistulas
- Provide bony support to alar base in cases with nasal asymmetry
- Eliminating mucosal recesses

23.3.1.5 Conventional Orthodontics in the Permanent Dentition

The orthodontic treatment goals are similar to those for nonclefts, using the same general orthodontic principles utilized for noncleft patients: To establish ideal dental function, facial aesthetics, and speech. Extraction of mandibular teeth to compensate for a hypoplastic upper jaw is usually not indicated until after the critical mandibular growth period has passed. In CLP patients, a bonded palatal fixed retainer is often necessary after treatment involving arch expansion to avoid relapse of the corrected palatal arch form:

- Improve the relationship of the lips
- Achieve harmonious balance of the dentition in the opposing jaws
- Achieve favorable skeletal maxillomandibular jaw relationship
- Achieve normal incisor overjet and overbite
- Correct dental axial inclinations
- Avoid the use of artificial teeth
- Achieve functional dental occlusion
- Achieve optimal nasal breathing

23.3.1.6 Dental Adjustments at Ages 16–17 Years for Girls and 18–19 Years for Boys

In cases with major skeletal jaw discrepancies, orthognathic surgery may be needed to normalize the skeletal jaw relationship and achieve a well-balanced facial appearance with stable dental occlusion. If two or more teeth are absent in the same dental segment, a small bridge is normally needed. However, dental implants are likely to become an important aspect of future prosthetic replacements.

23.4 Outline of CLP Treatment Procedures in Bergen

To appreciate our treatment philosophy, a brief summary of the treatment approach and concepts of the Bergen Cleft Palate Center will be presented. Along with the Oslo CP Center, it serves a population of five million. Due to demographic distribution, many patients must travel distances up to 2,000 km to either center. Hardships are compounded by the need to travel in very cold weather during winter; therefore, the planning and coordination of health services are crucial for optimal utilization of available resources. Treatment costs and travel expenditures are covered by the government's social security program. The Bergen CLP Team treats about 55 newborn babies yearly. Treatment procedures are coordinated between the Department of Plastic and Reconstructive Surgery, University Hospital of Bergen; the CLP Center at the Department of Orthodontics and Facial Orthopedics, Faculty of Medicine and Dentistry, University of Bergen; and the Eikelund Center for Speech Pathology.

23.4.1 Plastic Surgery

Since 1986, in complete clefts of the lip and palate, a Millard lip closure is performed at 3 months

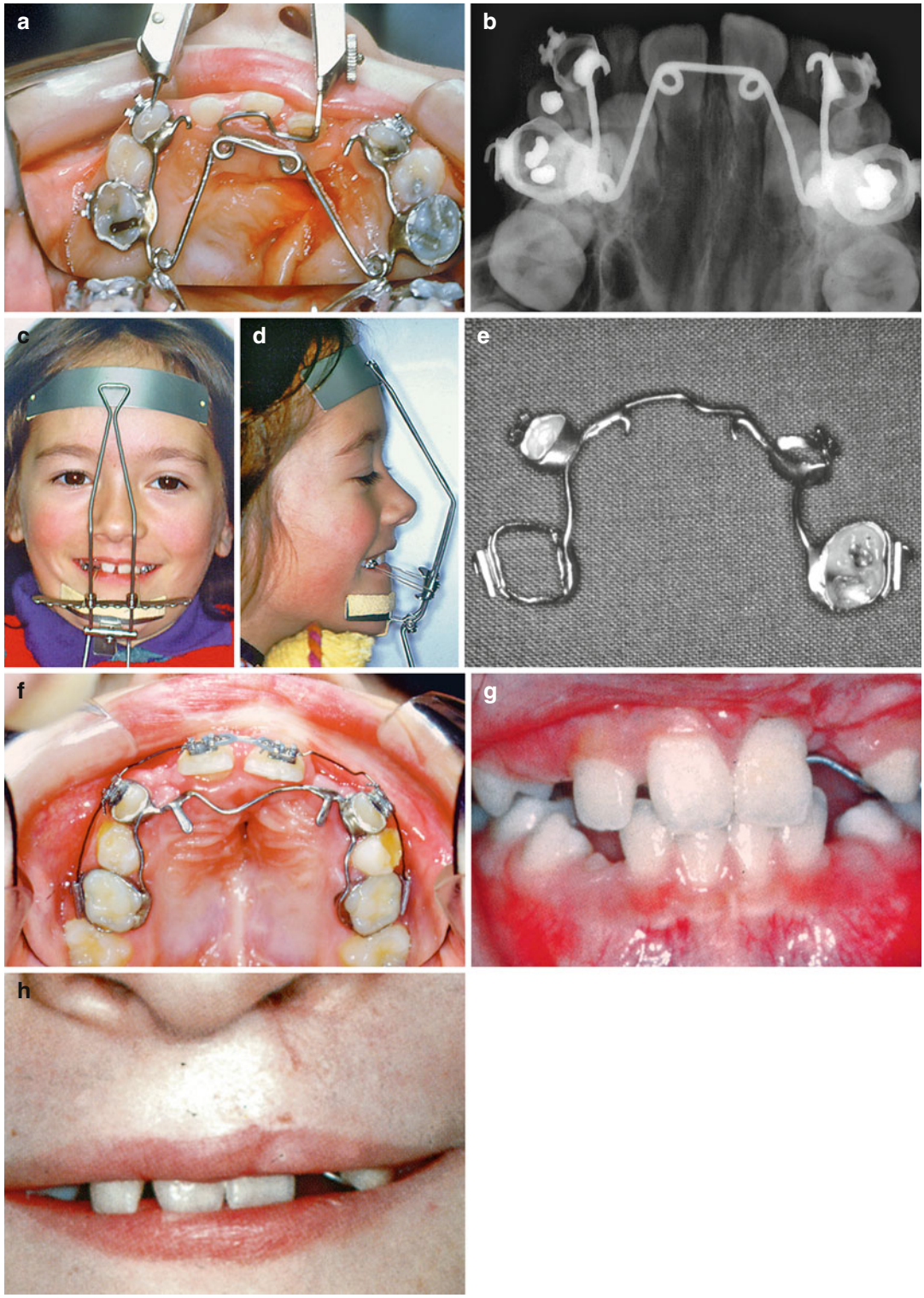

Fig. 23.2 (**a**–**h**) Interceptive orthopedics (Bergen rationale). (**a**, **b**) Transverse maxillary widening using a modified quad-helix appliance. (**c**, **d**) Followed by maxillary protraction with a facial mask (Delaire type). (**e**, **f**) Correction retained with a fixed palatal arch with brackets and tubes for early alignment of the upper incisors. Retention is utilized until deciduous anchor teeth are shed. (**g**, **h**) A nice dental smile as early as possible

combined with a single-layer vomerplasty for closure of the anterior part of the palate. The soft palate and isolated palatal clefts are closed at 12 months using a von Langenbeck technique. Alveolar bone clefts are left open until secondary bone grafting at 8–11 years of age. Between 1971 and 1986, the lip closure was combined with a periosteoplasty of the cleft alveolar process (Schjelderup and Johnson 1983).

23.4.2 Interceptive Orthopedics

23.4.2.1 Protraction Facial Mask

Extraoral heavy forces from a facial mask directed forward and downward from the maxillary cuspid area have been shown to correct midfacial retrusion at an early age (Delaire et al. 1972, 1976; Tindlund 1987, 1989, 1994a, b; Tindlund and Rygh 1993a, b; Tindlund et al. 1993a, b, 1994). Protraction from the maxillary cuspid area produces an adequate horizontal and vertical force to increase midfacial vertical height as well as anteroposterior length. In some instances, it also can reduce an anterior open bite by lowering the palatal plane. For this reason, early correction of anterior and/or posterior crossbites during the deciduous and mixed dentition is highly recommended:

Bergen rationale:

1. Transverse expansion coupled with
2. Protraction of the upper jaw and
3. The use of fixed palatal arch retention after treatment

When considering a treatment plan for young children who travel long distances, it is important to consider patient comfort as well as treatment efficiency.

In cases of marked midfacial retrusion, interceptive orthopedics is started at 6 years and often lasts for 15 months with an average of six visits (two visits for a transverse expansion of about 10 mm during a 3-month period and an additional four visits for the use of protraction forces for 12 months).

23.4.2.2 Quad-Helix Spring (with Four Bands and Hooks)

A fixed palatal expansion appliance can be easily combined with the use of an extraoral facial mask (Figs. 23.1 and 23.2) (Rygh and Tindlund 1982) providing:

1. Controlled transverse expansion when needed
2. Adequate fixation for anterior protraction by a facial mask
3. Use with edgewise appliance for the alignment of incisors
4. Well tolerated by small children without sedation, causing a minimum of discomfort
5. Minimum of chair time
6. Can be easily kept clean

The creation and use of a modified quad-helix [Rocky Mountain: maxillary quad-helix. 0.38 (0.985 mm) Blue Elgiloy (Ricketts)] appliance (Rygh and Tindlund 1982) is shown in Fig. 23.3. Four preformed bands with brackets or tubes are placed on the second deciduous molars and deciduous cuspids and are soldered to the quad-helix spring. The first deciduous molars serve as additional anchorage. Permanent molars are banded only when the second deciduous molars are missing. Hooks for elastics are positioned mesiolingually to the cuspid bands. The elastics are attached to the protraction facial mask (Fig. 23.2c, d). The elastic forces are directed forward and downward from the anterior maxillary segment, resisting the normal counterclockwise rotational effect. The quad-helix appliances when used with brackets and tubes permit alignment of upper incisors after their eruption (Figs. 23.1 and 23.3j–l).

23.4.2.3 Transverse Expansion

The modified quad-helix appliance is removed from the teeth and adjusted and activated at 6-week intervals (Fig. 23.2a, b). A resiprocal force of about 200 g on each side is optimal. The arch increases in width approximately 3 mm per month, irrespective of cleft type (Tindlund et al. 1993b). The cuspid segments often need more expansion than the molar area. Because there is a great tendency for relapse, overexpansion is necessary.

With transverse expansion of the maxillary arch, a downward clockwise rotation of the mandible occurs without any forward movement of the maxilla (Tindlund et al. 1993b). By comparing the use of fixed quad-helix appliances with removable expansion plates on noncleft patients, Hermanson et al. (1985) found that a fixed quad-

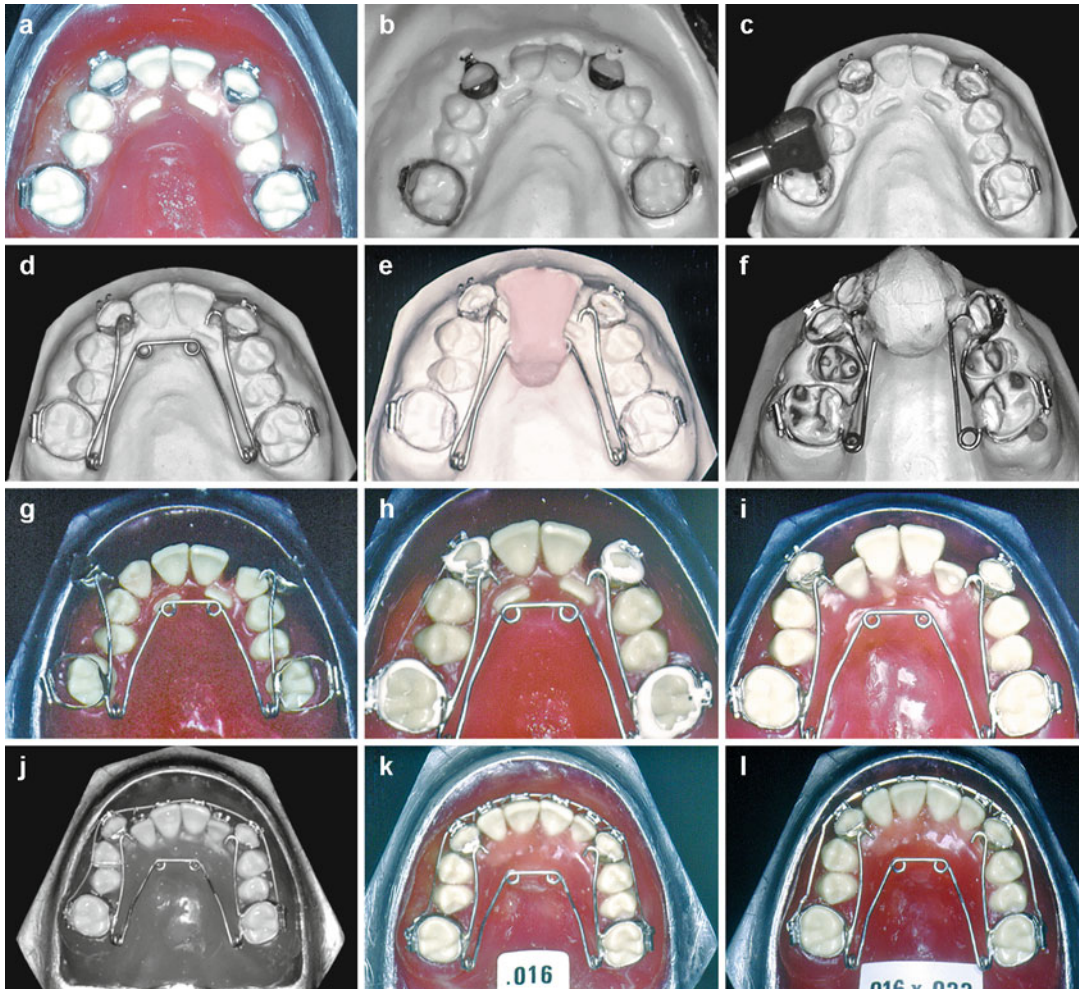

Fig. 23.3 (**a**–**l**) Fabricating a modified quad-helix appliance. (**a**, **b**) Bilateral posterior crossbite with lack of space for erupting lateral permanent incisors. Bands with brackets or tubes are fitted to the upper deciduous cuspids and deciduous second molars and carefully replaced into an alginate impression. (**c**) Plaster removed underneath soldering zones. (**d**) Quad-helix arms are precisely adjusted. (**e**, **f**) Quad-helix arms are soldered to all four bands. (**g**) Each arm is individually activated. (**h**) Cemented. (**i**–**k**) Combined with round labial arches for alignment of incisors. (**l**) Labial incisor root torque with rectangular archwire

helix appliance was more effective with fewer visits, less costly, and required shorter treatment time. Further, a removable plate would not readily resist the forward-downward traction to the cuspid area from a facial mask. As a rule, transverse expansion is completed before protraction is started.

23.4.2.4 Protraction

The quad-helix appliance is formed to make passive contact with the incisors, by bends or by a soldered-on extension (Fig. 23.2a, b). In cases where no transverse expansion is needed, or if the quad-helix spring is inconvenient after the expansion period, a simple palatal arch is soldered to four bands on second deciduous molars and cuspids (Fig. 23.2e, f). The intraoral appliance is used as anchorage for the facial mask (Scheu: Great Lake Reversed Pull Face Crib, 2500.1 small) (Fig. 23.2c, d). No other mask fixation is needed other than the two intraoral elastics

[Unitek: Latex ex-oral 1/4"LGT; Unitek/3M: "Fran" 8oz. 1/4" (404–736)] from hooks in the cuspid areas. The force used for facial protraction is about 350 g on each side, totaling 700 g. The facial mask is used mainly at night for 10–12 h. Sleeping disturbances have not been reported. Patient cooperation is excellent in almost all cases. Within a few days, the children manage to put on the mask themselves, and complaints about soreness are very rare. It is important that the elastics are attached to the anterior segment and inclined downward and forward about 15° to the occlusal plane. Several facial masks of various designs are now available. If the use of protraction forces is delayed until the permanent incisors are fully erupted, elastic forces from hooks placed on the arch wire between the lateral incisor and cuspid area may be utilized. In most cases, the incisors should be advanced bodily to obtain surface bone deposition at subnasal (A-point) by use of edgewise arch wires with labial root torque (Fig. 23.3l).

23.4.2.5 Fixed Retention

The results obtained by early orthopedics must be stabilized with a passive fixed palatal arch wire. The retainer remains until the anchor deciduous teeth are lost (Fig. 23.2e). The palatal arch wire (or quad-helix appliance) may be combined with multibracketed appliances to align the permanent incisors (Fig. 23.2f).

23.4.2.6 Treatment Timing of Interceptive Orthopedics

Proper treatment timing during the late deciduous dentition or early mixed dentition periods is of utmost importance. Delaire et al. (1972, 1976) observed that maximum skeletal maxillary changes occurred when protraction therapy was instituted before 8 years of age. Tindlund (1994a, b) found significantly better skeletal response when protraction began at 6 years (mean age 6.3). At this age, the annual sutural growth rate is nearly as high as that found at the pubertal period (Bjørk 1968), when development of the heavily interdigitated sutural systems has already commenced (Melsen 1974; Melsen and Melsen 1982). Protraction during the deciduous dentition period minimizes unwanted dentoalveolar proclination of maxillary incisors in the permanent dentition (Tindlund et al. 1993a, b).

Early habilitation of facial appearance and dental functions, preferably before the start of school, is considered a major goal (Semb 1991a; Rygh and Tindlund 1982; Tindlund 1987). The cooperation of the young patients is often more predictable at this age (Tindlund 1994a, b). The objective of having the permanent upper incisors erupt into a positive overjet and overbite relationship warrants that orthopedics should be started even earlier in cases with severe skeletal jaw discrepancies. Postponement of orthopedic treatment increases the likelihood that achieving positive effects on the facial growth pattern will fail to occur.

23.4.3 Treatment Results of Using a Protraction Facial Mask

23.4.3.1 Clinical Results

The excellent results achieved by the early use of interceptive orthopedics to correct an anterior and/or posterior crossbite also have a desirable effect on improving dentofacial appearance (Delaire et al. 1972, 1976; Tindlund and Rygh 1993a, b; Tindlund et al. 1993a, b, 1994; Tindlund 1994a, b; Buschang et al. 1994). Treatment outcomes can vary because success also depends on inherited craniofacial characteristics and the severity of the primary and secondary midfacial malformations related to clefting. Generally, the treatment response is less favorable in facial types with increased anterior vertical height and a steep mandibular plane angle.

Patient cooperation is of major importance for obtaining a good treatment outcome. Using protraction therapy in 108 CLP patients with anterior crossbite in the deciduous dentition, Tindlund et al. (1993a) achieved favorable incisor relationships in 98 cases. Significant increase of maxillary skeletal prognathism by protraction was found only in the UCLP group, whereas treatment effects in the BCLP cases were mainly dentoalveolar (Tindlund and Rygh 1993a). The observation of significant differences between the UCLP and BCLP groups in Bergen is most likely associated with the pri-

mary surgical procedures utilized, which included a periosteoplasty. A bony fusion of jaw segments in BCLP on one or both sides may impair treatment response as well as facial growth.

After protraction treatment, there was no significant difference in the maxillary prognathism attained between the UCLP and BCLP groups (Tindlund and Rygh 1993a). The sagittal position of the upper molars was normalized in both groups. Increase of the upper facial height ("n-sp") and clockwise rotation of the occlusal plane were significantly greater in the BCLP group. The upper incisors were still retroclined in both groups, which is considered a beneficial state. A later dentoalveolar proclination will compensate for future mandibular development. On the average, the period of protraction lasted 12 months in the UCLP group and 15 months in the BCLP group.

The skeletal response to maxillary protraction is expected to vary considerably as a consequence of skeletal facial variation, differences in the cleft defects, and in cleft repair (Tindlund 1994a). Favorable response in the sagittal skeletal maxillomandibular jaw relationship was found in 63 % (mean increase of angle ANB was 3.3°), whereas favorable response on skeletal forward movement of the maxilla was found in 44 % (mean advancement 2.4 mm). A combined favorable response of both the mandible and the forward movement of the maxilla was found in 35 % (Tindlund 1994a). In this group, the mean increase of the maxillary prognathism was 2.1°, the angle ANB increased 3.7°, the maxilla moved forward 3.1 mm, and the maxillary dentition was advanced 4.3 mm. In cases where the sagittal jaw discrepancy was due to overgrowth of the mandible, the resulting changes accentuated a mandibular downward/posterior rotation, increasing anterior facial height.

Cephalometric predictors for good orthopedic treatment response were retrusion of the upper jaw due to short maxillary length resulting in a class III skeletal and dental relationship and counterclockwise inclination of the occlusal plane. This is associated with a retrusion of the upper lip and the nose tip (Tindlund 1994b). Favorable increase of a positive ANB angle is associated with mandibular retrognathism, whereas skeletal forward movement of the maxilla with lesser changes in the ANB angle was more often seen in cases with normal mandibular prognathism.

23.4.3.2 Limitations

During protraction, the upper permanent incisors should never be proclined beyond normal tooth inclination within supporting basal bone, and the lower incisors should never be retroclined more than their normal position within the alveolus (Tindlund et al. 1993a). If a normalization of the maxillomandibular skeletal discrepancy is not achieved along with normal dental axial inclinations of the permanent incisors, further protraction should be avoided and orthognathic surgery considered (see categories 2A and 2B in Sect. 23.4.4). On the other hand, protraction during the deciduous dentition is advocated in every case with an anterior crossbite, even in cases with a family history of true mandibular prognathism. The final diagnosis for orthognathic surgical treatment should be delayed until approximately 13 years of age for girls and the late teens for boys.

23.4.3.3 Stability/Relapse

After protraction, the maxilla and mandible appear to maintain their original growth pattern. Although there is no relapse of the corrected upper jaw relationship (Tindlund 1989), the maxillomandibular relationship often worsens through normal forward growth of the mandible while the maxillary position relative to the anterior cranial base appears to remain constant. However, long-term results show individual variation of this finding, and in cases with moderate midfacial retrusion, early protraction is often sufficient to maintain the improved inter-incisor relationship with growth (Fig. 23.1) (Rygh and Tindlund 1995).

23.4.3.4 Soft-Tissue Profile

As already stated, the characteristic concave profile with midfacial retrusion is readily improved with protraction (Fig. 23.1) (Buschang et al. 1994). The changes were nearly the same in BCLP and UCLP patients with significant protrusion of the upper lip (mean increase of 3.0° in the Holdaway angle; mean increase of maxillomandibular lip positioning (SS-N-SM, angle of 2.5°)) (Tindlund and Rygh 1993b). Although there is a

close relationship between the soft-tissue profile and the supporting hard-tissue structures (Segner 1992; Tindlund and Rygh 1993b), the improved soft-tissue profile commonly seen after protraction is more stable than the ANB angle which is also dependent on mandibular position, size, and growth (Tindlund 1989).

23.4.4 Long-Term Prognosis After Interceptive Orthopedics

By age 6 years, the craniodentofacial growth and development may give some indications of the facial appearance at adulthood. The CLP patients with few exceptions can be placed into one of four categories, irrespective of the type of clefting (Rygh and Tindlund 1995):

Category 0: Normal skeletal facial morphology
Need of treatment:
- Alignment of upper permanent incisors at ages 7–8 years?
- Conventional orthodontic treatment at ages 11–13 years

Prognosis: Very good

Category 1: Normal skeletal facial morphology, except posterior crossbite(s)
Need of treatment:
- Interceptive orthopedics: transverse expansion of the upper jaw at ages 6–7 years
- Alignment of upper permanent incisors at ages 7–8 years?
- Conventional orthodontic treatment at ages 11–13 years

Prognosis: Very good

Category 2A: Moderate skeletal facial discrepancies
Need of treatment:
- Interceptive orthopedics: transverse expansion and protraction of the upper jaw at ages 6–7 years
- Alignment of upper permanent incisors at ages 7–8 years?
- Conventional orthodontic treatment at ages 11–13 years

Prognosis: Good/fair for a permanent result

Category 2B: Severe skeletal facial discrepancies, however, cannot be differentially diagnosed from *Category 2A until ages 12–15 years*
Need of treatment:
- Interceptive orthopedics: transverse expansion and protraction of the upper jaw at ages 6–7 years
- Alignment of upper permanent incisors at ages 7–8 years?
- Conventional orthodontic treatment at ages 11–13 years
- Combined orthodontic/orthognathic surgical correction at adult age

Prognosis: After orthognathic surgery: good regarding upper arch form, tooth position, and soft-tissue profile. Poor permanent result until after orthognathic surgery with stable retention of the arch form.

Patients in category 2B may be candidates for orthognathic surgery at an adult age. However, this is not a contraindication for early maxillary protraction provided this does not lead to dentoalveolar compensations that undermine the possibility for optimal surgical intervention at a later age. On the contrary, early orthopedics may create a better physical environment for successful orthognathic surgery. Thus, less maxillary advancement at the time of surgery may improve the postsurgical stability. Besides, the child has greatly benefited from an improved dentofacial appearance during the important formative years.

Conclusions

The specific aims of early interceptive orthopedics/orthodontics are:

1. To achieve maximum facial aesthetics, with an adequate incisor overjet–overbite relationship and “a nice dental smile” with symmetry of the dental and facial midlines.
2. The elimination of anterior and posterior crossbite and the recovery of space for the erupting incisors. This is considered “lege artis” (standard operating procedure) in children without clefts, and, obviously, the

same considerations are valid for a child with a cleft.

3. Early orthopedic–orthodontic correction generates an optimal skeletal base to accommodate erupting upper permanent incisors and improve dental function.

Protraction produced significant changes: (1) a more anterior position of the upper jaw and (2) a more posterior position of the chin point due to mandibular downward-backward rotation. Significant increase of skeletal maxillary prognathism was found only in the UCLP patients, while in BCLP cases, the treatment effect was mainly dentoalveolar.

The initial growth pattern reappears after protraction, with the upper jaw's position relative to the anterior cranial base remaining stable, while the mandible's position changes as it grows forward and downward. Soft-tissue profile changes are lasting.

Fixed appliances are indispensable for controlled orthopedic/orthodontic mechanics to obtain all treatment objectives and for the permanent retention of the corrected arch form. Bonded palatal retainers are frequently required.

A diagnosis-related checklist is the method of choice for individualizing orthodontic treatment. Orthopedic/orthodontic intervention should be based on the same principles that are valid for noncleft patients.

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LeFort I Osteotomy

24

S.A. Wolfe and Samuel Berkowitz

24.1 Surgical Maxillary Advancement LeFort I Osteotomy

Not long ago, maxillary advancement seemed a formidable procedure to many surgeons. Cleft patients with Class III malocclusion often were treated by the more familiar method of mandibular setback, even though the problem, by clinical and cephalometric examination, could be shown to be in the maxilla.

Today, the LeFort I osteotomy is a standard adjunct to the treatment of patients with cleft lip and palate. No matter how gentle or atraumatic the original surgery on the lip and palate, there will probably always be cleft patients who require the LeFort I procedure. It should be as much a part of the armamentarium of cleft palate teams as closure of the lip or palate or a pharyngeal flap.

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Mandibular growth should be largely completed before a maxillary advancement is performed; for girls this age is around 14–15, and for boys perhaps a year or two older. Most orthodontists advise that several lateral cephalometric films, taken 6 months apart, should show no further growth before the operation is scheduled.

As for timing, it is better to perform the lip and nasal surgery during separate sessions. If the alveolus is intact and there are no buccal crossbites, expansion of the maxilla is not required, and the LeFort I osteotomy is a relatively simple procedure. The nonintubated nostril is packed with cocaine/epinephrine-impregnated gauze, as for rhinoplasty, and the upper labial sulcus is infiltrated with a 1:2,000,000 epinephrine/hyaluronidase solution. The incision is made above the reflection of the sulcus, sparing the frenulum. The mucosal incision does not extend beyond the first molar. Subperiosteal dissection of the anterior maxilla is carried out to the infraorbital rims, visualizing the infraorbital nerves, and then taken posteriorly beneath the mucoperiosteal tunnel to the pterygomaxillary space. If the dissection is strictly subperiosteal, there is no bothersome exposure of the buccal fat. The piriform aperture is dissected, sometimes removing a portion of the nasal spine, and the nasal mucoperiosteum is dissected back to the hard palate–soft palate junction. The septum can either be separated bluntly from the vomer or a guarded osteotome can be used. The osteotomy is performed largely with the reciprocating saw, starting laterally in the thick bone beneath the buttress of the

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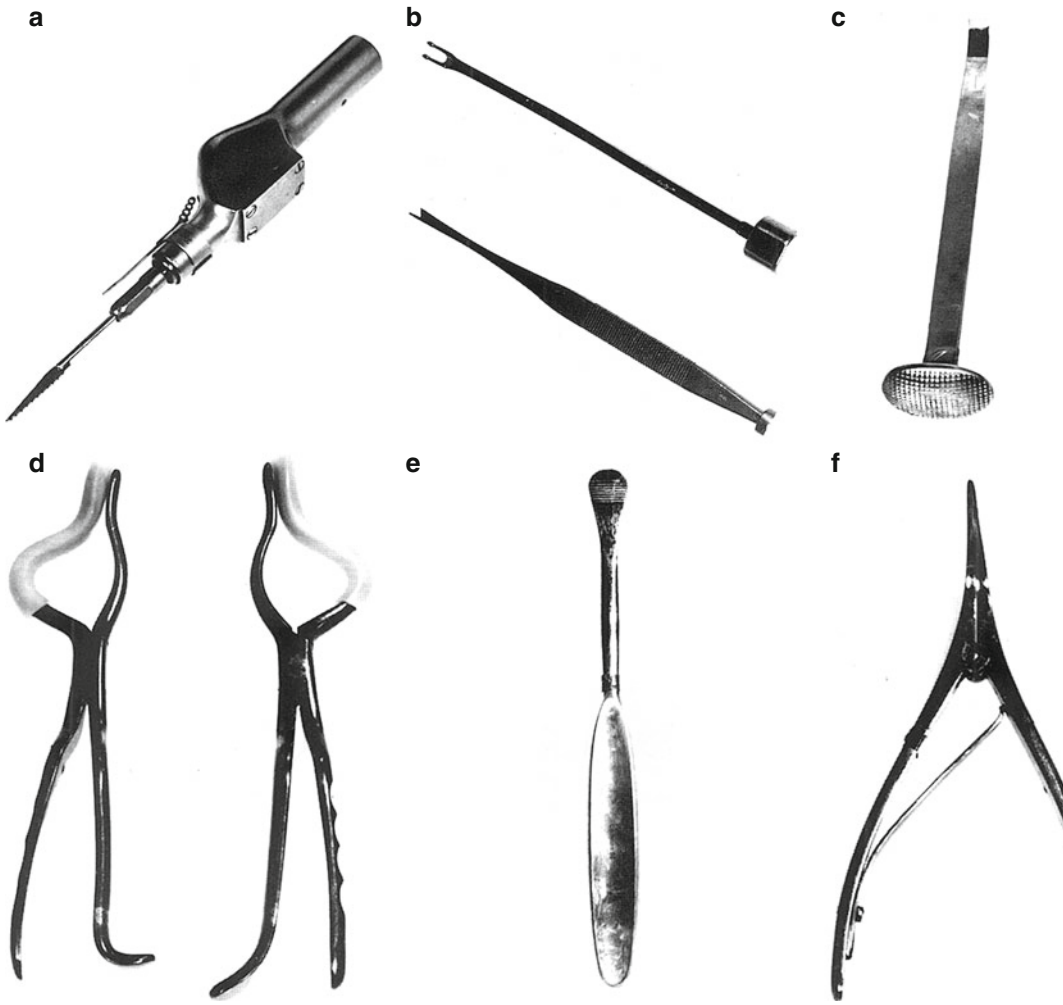

Fig. 24.1 (**a**–**f**) Instrumentation for the LeFort I osteotomy. (**a**) Reciprocating saw with irrigation (Aesculap). (**b**) Guarded septal and nasal osteotomes. (**c**) Kawamoto osteotome. (**d**) Rowe forceps with rubber guard on the palatal arm. (**e**) Nestor (blunt, heavy, periosteal elevator modified by Jack Nestor Engineering, Inc., Miami, Florida). (**f**) Expansion forceps

zygoma and proceeding medially through thinner bone. The osteotomy through the piriform aperture and medial wall of the antrum is done with the saw blade pointed laterally (Fig. 24.1).

Sectioning of the palatine bone, the sole attachment of the maxillary tuberosity to the pterygoid plate of the sphenoid, follows. The lateral osteotomy can be taken a bit farther back by a few taps on a straight osteotome, and the medial antral wall can be further sectioned with a guarded nasal osteotome.

At this point, the only remaining attachment of the lower maxillary segment is the posterior wall of the antrum, and firm, downward finger pressure on the maxilla is usually enough to produce a down-fracture. If not, the forceps can be inserted underneath the nasal mucosa and the maxilla completely mobilized with a downward and side-to-side motion. It can be further mobilized with a blunt elevator used as a lever.

The maxilla is then placed in the desired occlusal relation with the mandible, and both jaws are placed in the desired relationship with the rest of the face. An autogenous iliac or cranial bone graft is used when the face is to be lengthened, when the degree of maxillary advancement is more than 5 mm, or when the patient has a cleft. If the maxilla is shortened, the resected bone is placed over the osteotomy lines.

Sometimes the alveolus is intact, but the maxilla needs to be expanded, as may occur in a cleft patient

who has a buccal crossbite and an alveolar cleft. This procedure is easily performed from above the hard palate, and the palatal mucosa is kept intact if possible. The sectioning is performed with the reciprocating saw, and an elevator is inserted to gently pry the two segments apart. Expansion forceps can be used if required. If the palatal mucosa absolutely prevents expansion, it is divided, creating an alveolar and anterior palatal cleft.

If there is an alveolar cleft to begin with, the two maxillary segments are handled independently and brought into proper occlusion with the mandible. The palatal cleft-nasal floor defect is bone-grafted, and if necessary a transportation flap is developed from the buccal sulcus (Burian) to close the palatal defect. In rare instances, a tongue flap is required. The nasal lining, which will have been carefully dissected at the beginning, is closed before the palatal bone graft is inserted.

The procedure has now been refined to the stage that is the same regardless of whether the alveolus was initially intact. Miniplates are placed between the upper and lower portions of the maxilla for rigid fixation. If bone grafts are required, they are placed either between or over the bone cuts.

If the desired maxillary advancement measures more than 6 mm, bone grafts can be wedged into the pterygomaxillary gap. This step is facilitated by using a traction wire placed through the thick bone beneath the nasal spine. The wire is used to pull the maxilla to the opposite side, which opens the gap and allows impaction of the bone graft. Circumzygomatic wires are almost never used, because they pull the maxilla back, they are too long (long wires can "stretch" more than short wires), and they do not prevent the anterior maxilla from rocking downward.

Wolfe (1989) uses an iliac or cranial bone graft on all cleft patients, as these patients are likely to have a maxillary relapse. Generally, the bone can also be used as an onlay to fill out a deficient maxilla. If the advancement is less than 5 mm, bone is placed only over the anterior osteotomies and in the alveolar and palatal cleft, if present.

The use of anything other than a fresh autogenous bone graft is unsafe. It takes about 15 min to harvest the needed amount of iliac or cranial bone. In the former case, the patient will be comfortable as far as the hip is concerned within 1–2 weeks. By this time, the autogenous graft will have consolidated. With cadaver or demineralized bone or with hydroxyapatite, consolidation may require months, or may *never* occur.

Like the sagittal splitting procedure for the mandible, the LeFort I osteotomy, once mastered, can provide a solution to a number of maxillary problems. After the horizontal osteotomy, down-fracture, and mobilization, the maxilla can be:

1. Advanced directly with or without a bone graft (in the noncleft class III patient).
2. Advanced, or advanced and expanded transversely, with a bone graft (in the cleft patient).
3. Moved superiorly after resection of a measured amount of maxilla above the horizontal osteotomy (in cases of "long face," resulting from vertical maxillary excess).
4. Moved inferiorly with a bone graft (in cases of "short face," or vertical maxillary deficiency).
5. Sectioned into multiple segments with teeth (Wassmund or Schuchardt procedure, done from above).
6. Moved directly backward, although this is difficult to do. (The resection should be of the maxillary tuberosity after extraction of the third molars rather than of the pterygoid plate.) The same result can generally be achieved by an associated segmental osteotomy performed more anteriorly.

With the maxilla in the down-fractured position, multiple osteotomies can be performed from above, which, coupled with or without dental extractions, permit the dental correction of complex malarrangements of the maxilla in one stage. The circulation of blood to the anterior segment comes entirely through the palatal mucoperiosteum, and one must be certain that there are no protrusive edges from the occlusal splint to impinge on the anterior palate. Any number of transverse sagittal osteotomies can be performed, depending on the requirements of the individual case.

Attempts to treat an anterior open bite by mandibular ramus osteotomies are often unsuccessful due to relapse caused by the predominance of the masticatory muscles. Anterior segmental osteotomies of the mandible are appropriate when there is dental crowding and a downward angulation of the mandibular occlusal plane.

The Schuchardt procedure can be used to shorten posterior maxillary height, but it is rarely used in the USA because it requires either an interdental osteotomy or a tooth extraction (Fig. 24.2).

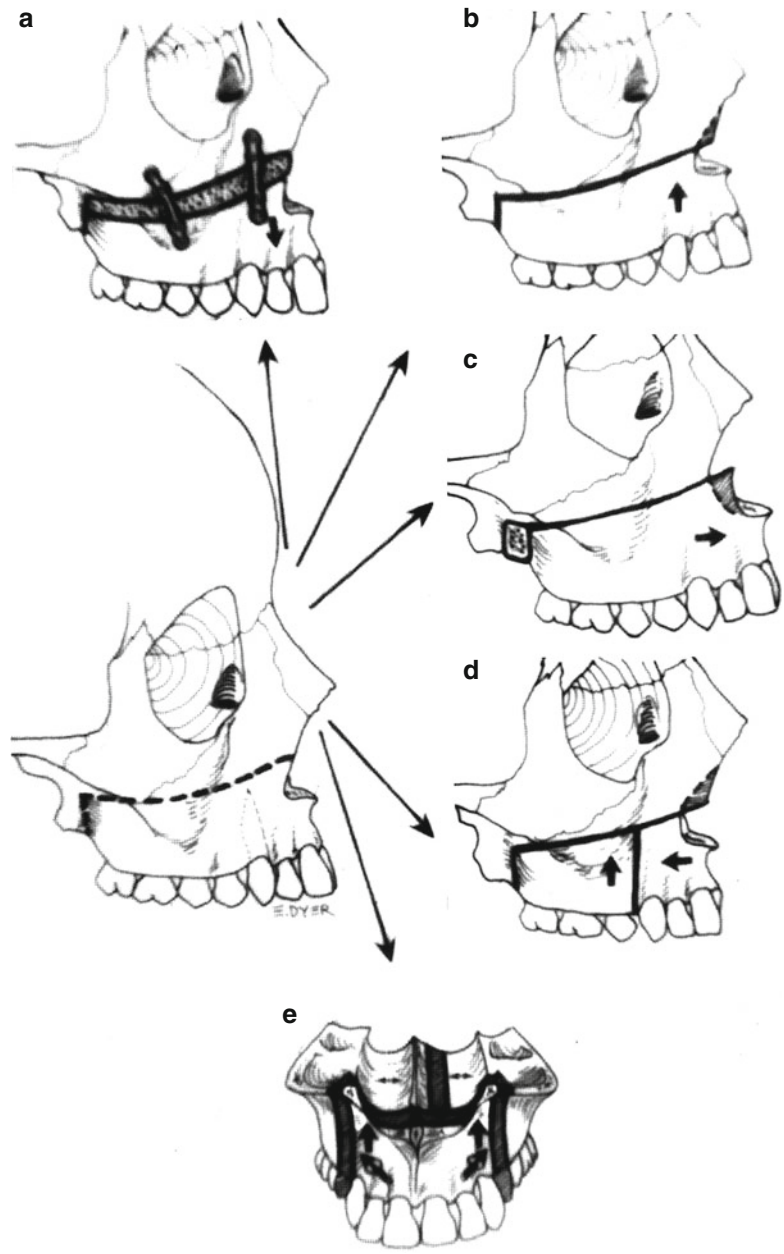

Fig. 24.2 LeFort I surgical procedures: various directions the maxilla can be moved. (**a**) Inferiorly: requires bone grafts to maintain the new position. (**b**) Superiorly: no grafts required. (**c**) Forward: requires a posteriorly supporting bone graft. (**d**) The posterior segment is moved superiorly while the anterior segment is moved posteriorly. When the premaxilla is retruded it usually needs to be surgically widened through the midpalatal suture between the central incisors to maintain good cuspid interdigitation. (**e**) The premaxilla moved superiorly

If the orthodontist can level the maxillary occlusal plane, even by accentuating the open bite, the simplest and most stable solution is the LeFort I osteotomy. If the position of the maxillary central incisors relative to the lower vermilion border of the upper lip is satisfactory beforehand, this relationship is preserved. If desired, the maxillary incisors can be raised or lowered relative to the upper lip.

After the maxilla has been completely mobilized, intermaxillary fixation is established and the maxillomandibular complex seated with firm upward and posterior pressure to set the condyles. Appropriate resection of the posterior and, if necessary, the anterior maxilla is performed until the desired anterior maxillary height is obtained. Stabilization of the maxillary osteotomy is then performed with miniplates, and the intermaxillary fixation, if utilized, is temporarily discontinued to evaluate the occlusal relationship with the patient; head in a fixed position. This examination will reveal whether the condyles were inadvertently pulled out of

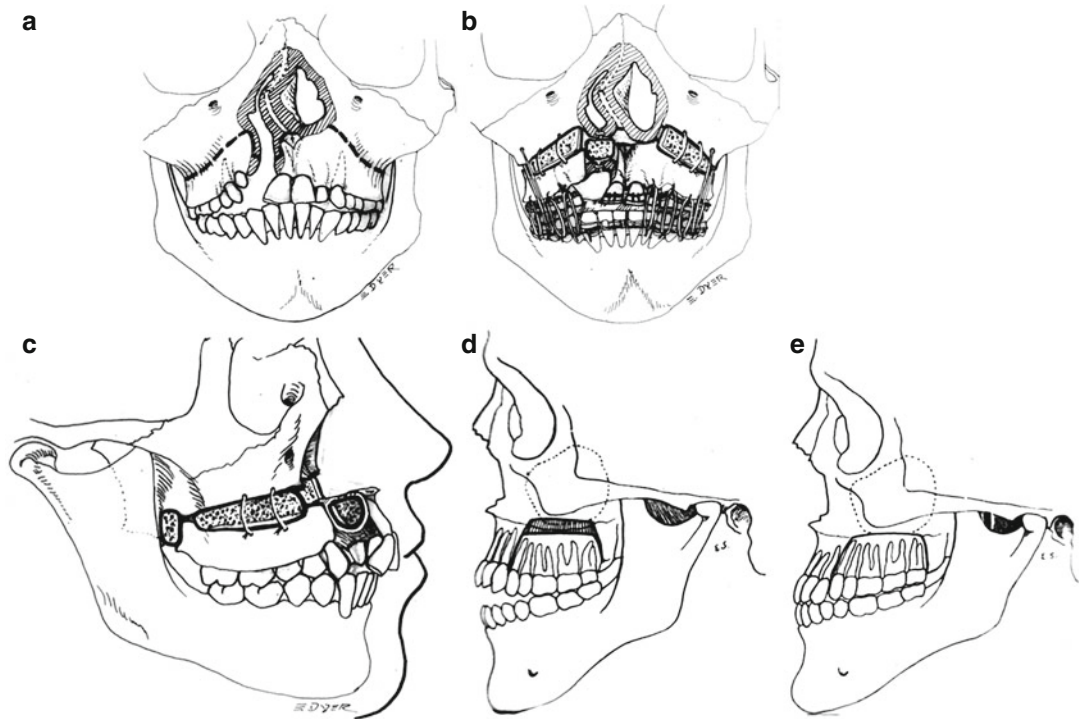

Fig. 24.3 (**a**) Initial incisions for LeFort I surgery with a secondary alveolar bone grafting to be performed simultaneously. (**b**) The maxilla is moved inferiorly with bone grafts placed at the surgical cite to support the lengthened maxilla. Alveolar bone graft placed from the nasal aperture to the alveolar crest. Prior to the use of metal plates (rigid fixation) steel sutures were used to stabilized the separated segments. An acrylic surgical wafer is used to position the bony segments according to prior mock surgery performed on plaster casts. Intermaxillary fixation of the maxilla to the mandible using intermaxillary rubber bands for 4–6 weeks is recommended in cases with severe palatal scarring in conjunction with the use of rigid fixation. (**c**) Lateral view shows a bone block placed between the perpendicular plates of the sphenoid and the maxillary tuberosity with a bone graft to the premaxillary–maxillary junction. (**d**) Buccal segments are superiorly positioned to permit mandibular auto-rotation and reduction of the anterior open bite. (**e**) Posterior maxillary impaction to close an open bite (Schuchardt). Buccal segments are superiorly positioned to permit mandibular auto rotation and closure of an anterior open bite

the glenoid fossae. A Class II relationship indicates that the maxilla must be posteriorly repositioned, either by resecting a portion of pterygoid plates (which is difficult) or by extracting the maxillary third molars and resecting a portion of the maxillary tuberosity (which is easier) (Figs. 24.2 and 24.3).

24.2 Stability of Maxillary Advancement

A disappointing yet frequent sequel to orthognathic surgery to advance the maxilla is its partial or complete return to the original state (relapse). The maxillary advancement occurs within a limiting soft tissue envelope (the skin and muscles). Mandibular advancement surgery, especially when it involves the mandibular ligaments, has a great tendency to relapse. The degree of relapse is often judged by measuring occlusal or skeletal landmark changes.

Hochban et al. (Hochban et al. 1993) in a review of the literature reported that the use of miniplates (in rigid fixations) is superior to wire fixation in overcoming the tendency to relapse. Currently, most reports favor the use of miniplates (Ward-Booth et al. 1984; Houston et al. 1989; Champy 1980; Horster 1981; Luyk and Ward-Booth 1985; Rosen 1986). Proffit and Phillips (1987) found a skeletal relapse at 32 % after midface advancement using wire fixation compared with 25 % after miniplate fixation.

Some investigators believe that the amount of relapse is directly related to the amount of advancement (Wolfe and Berkowitz 1989; Houston et al. 1989; Carpenter et al. 1989),

whereas others think there is no correlation between displacement of the maxilla and relapse (Rosen 1986; Proffit and Phillips 1987; Iannette et al. 1989). Proffit and Phillips also believe that it is important to achieve excellent occlusion following the operation to reduce the tendency to relapse. Epker (Epker 1981) suggests that interpositioning of bone grafts increases stability by enhancing bony consolidation.

It is generally accepted that the tendency toward relapse starts immediately after surgery and continues for up to about 6 months after the operation. After about 1 year, the correction can be considered stable (Houston et al. 1989; Epker 1981; Teuscher and Sailer 1982; Persson et al. 1986). Hochban et al. (1993), in an excellent review of the subject of postoperative maxillary relapse, reported cephalometric analyses of 31 patients preoperatively, postoperatively, and 1 year later. Fourteen patients had clefts of the lip and palate; the others were noncleft patients with maxillary deficiency. All had maxillary advancement by LeFort I osteotomy and miniplate fixation. Hochban et al. (1993) found that the amount of relapse was between 20 and 25 % in the cleft group and about 10 % in the noncleft group. The degree of relapse was related to the amount of advancement, thus confirming the earlier work by Rosen (1986) and Houston et al. (1989). The authors recommended surgical overtreatment and a good overbite–overjet relationship after orthodontic treatment.

Berkowitz sometime uses very light Class III elastics for 6 months to improve bony consolidation when he notices a maxillary relapse occurring. He believes that the muscular drape to the midface changes very slowly in adapting to skeletal changes, and therefore, some overtreatment is necessary in all instances.

Posnick and Ewing (1990) studied the outcomes in 30 adults and adolescents judged skeletally mature, who had unilateral cleft lip and palate and underwent LeFort I advancement. This group was investigated to determine the amount and timing of relapse, the correlation between advancement and relapse, the effect of performing multiple jaw procedures, the effect of different types of bone grafts, the effect of pharyngoplasty in place at the time of osteotomy, and the effectiveness of various methods of internal fixation. Friehofer (1977), also presented results of maxillary advancement in adolescence.

Tracings of preoperative and serial postoperative lateral cephalograms were digitized to calculate horizontal and vertical maxillary changes. No significant differences in outcomes were seen between patients who had maxillary surgery alone and those who had operations on both upper and lower jaws, nor did the outcomes vary significantly with the type of autogenous bone graft used or the segmentalization of the LeFort I osteotomy. Average "effective" advancement was greater both immediately and 2 years after surgery in patients who did not have a pharyngoplasty in place before the operation.

Advancement also was more stable both immediately and 2 years after surgery in the patients with miniplate fixation than in patients with direct-wire fixation. Mean downward (vertical) displacement was 2.6 mm with a relapse of 1.4 mm after 2 years. The degrees of relapse and of advancement or displacement did not correlate significantly.

There is another obvious risk factor, the tonicity of the orbicularis oris muscle ring, which needs to be considered. Unfortunately, there are no pressure measurements that can be utilized to improve the success-to-failure ratio.

24.3 Total Maxillary Advancement and Its Possible Effect on Speech

Jabaley and Edgerton (1969) Dez Prez and Kiehn (1974), and Bralley and Schoney (1977) have reported that speech of cleft and noncleft patients is unaffected after total maxillary advancement. Witzel and Munro (1977) say that is not always true. Epker and Wolford (1976) noted that the speech of patients with clefts who exhibited no VPI presurgically generally remained unchanged after maxillary advancement. However, those patients who have borderline closure or minimal velopharyngeal incompetence before surgery do exhibit speech changes following total maxillary advancement. Schwarz and Gruner (Witzel and Munro 1977; Epker and Wolford 1976; Schwarz and Gruner 1976) showed that patients with slight hypernasality and/or nasal emission before surgery became more hypernasal after maxillary advancement. They concluded that the degree of

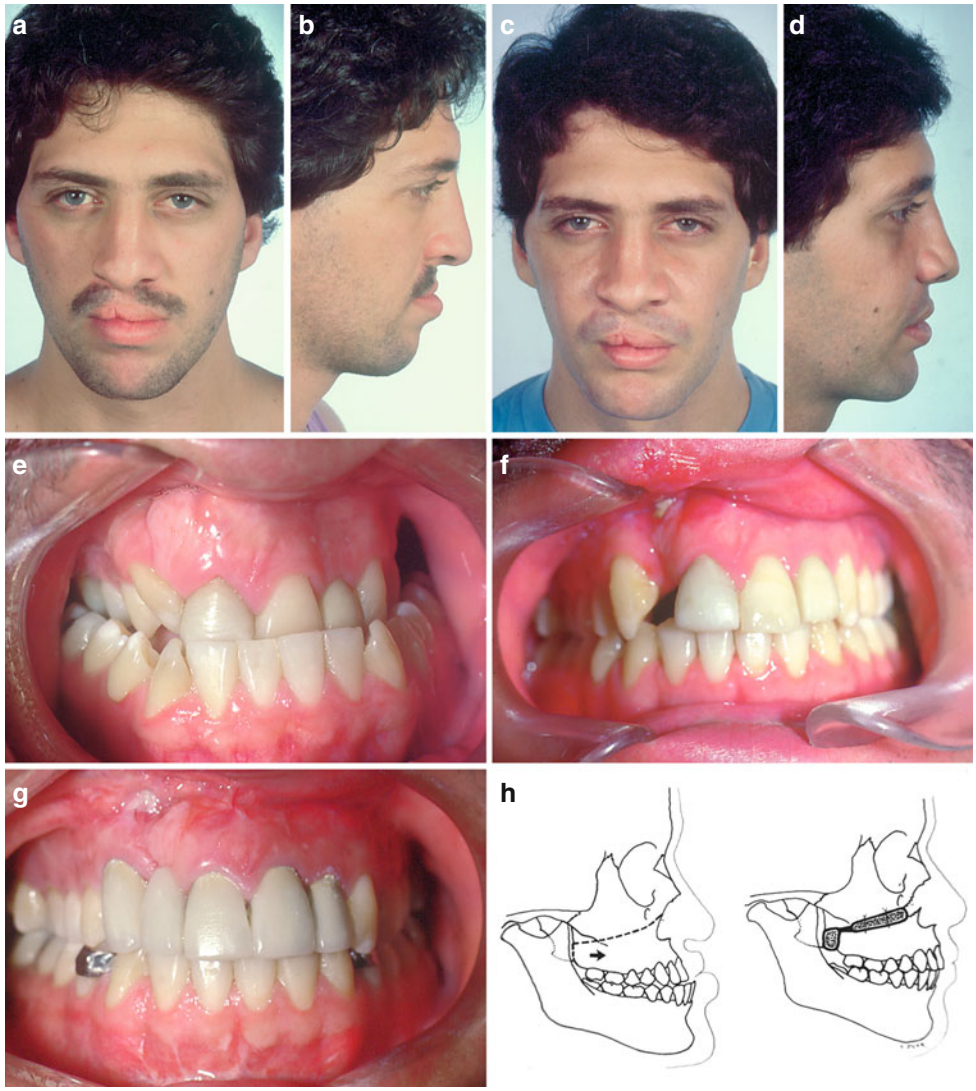

Fig. 24.4 (**a**–**h**) Case JS (AV–64) UCLP showing LeFort I advancement to correct midfacial retrusion. Treatment: Increase midfacial height, and widen the palatal arch. (**a**–**g**) Pre- and postsurgical facial and intraoral photographs showing changes in the profile and occlusion. Chin augmentation is usually contraindicated with midfacial advancement since it may lead to a concave profile after some maxillary relapse. (**h**) Type of surgery performed

deterioration was directly related to the extent of maxillary advancement and observed that deterioration could also occur in some noncleft patients. Schendel et al. (1979) believe the differences between the two groups are theoretically a reflection of the inherent deficit in palatal musculature and associated soft tissue in the cleft patient and/or cicatrization associated with surgical repair of the palatal clefts. Many cleft patients have hypoplastic velar muscles and associated soft tissues. All of these factors are reflected in the shorter soft palate in the cleft patient. They speculate that the increase in pharyngeal depth creates a significant functional demand which often cannot be met by cleft patients due to less soft palate length increases following maxillary advancement. Schendel et al. (1979) believe that the soft palate lengthens about one-half of the amount the maxilla is advanced. They also computed a "need ratio" (pharyngeal depth divided by soft palate length) in which a value of 0.68–0.84 is consistent with proper velopharyngeal function. A need ratio greater than 1.0 indicates possible postsurgical velopharyngeal incompetence (Figs. 24.4 and 24.5).

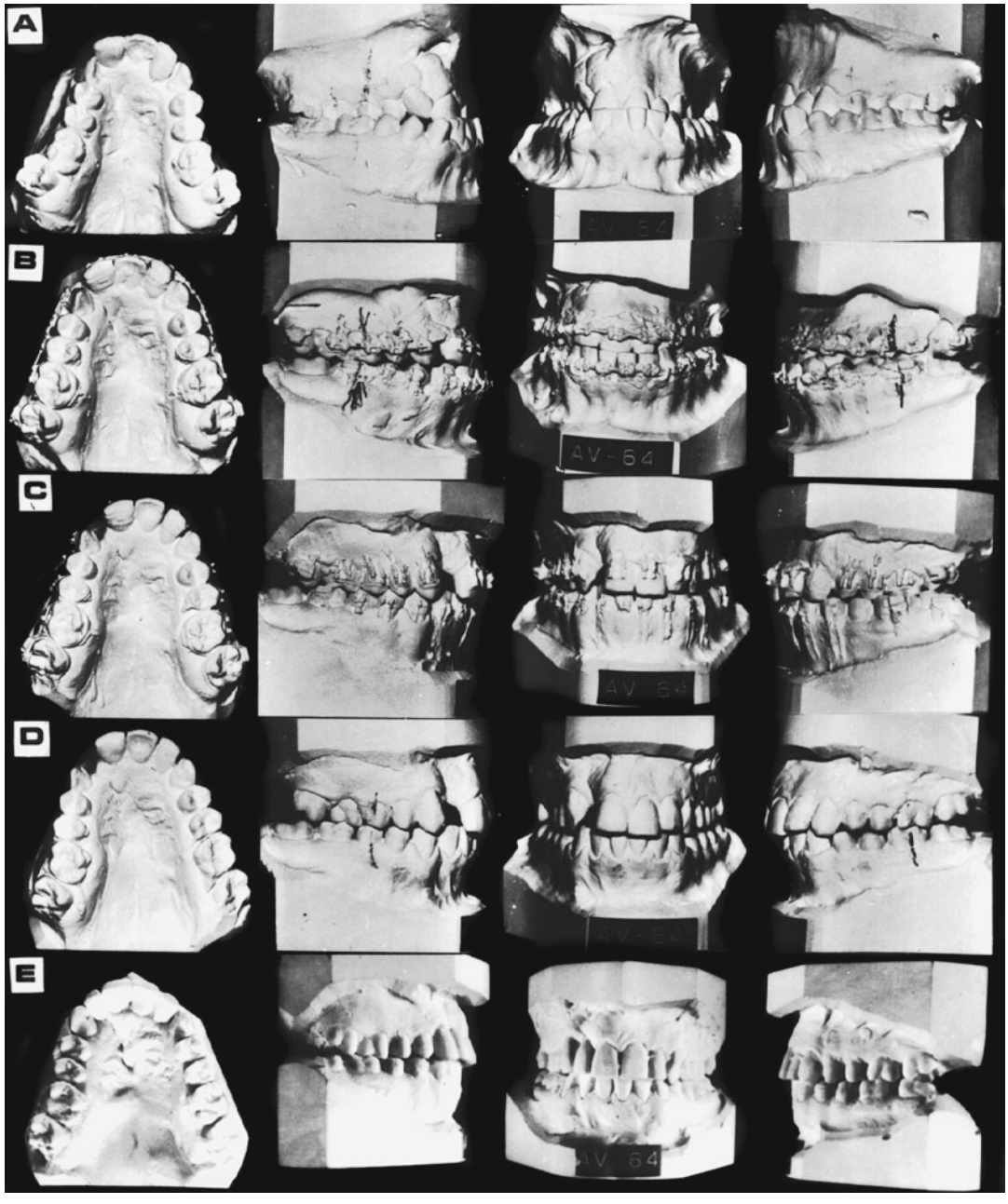

Fig. 24.5 Case JS (AV–64) (**a**–**e**). Serial dental casts. This case shows severe palatal collapse and scarring leading to buccal and anterior crossbite. Pre- and postsurgical orthodontics plus maxillary surgery reduced the anterior crossbite. The maxillary arch was orthodontically expanded to open the upper right lateral incisor space and to avoid additional surgery with more palatal scaring

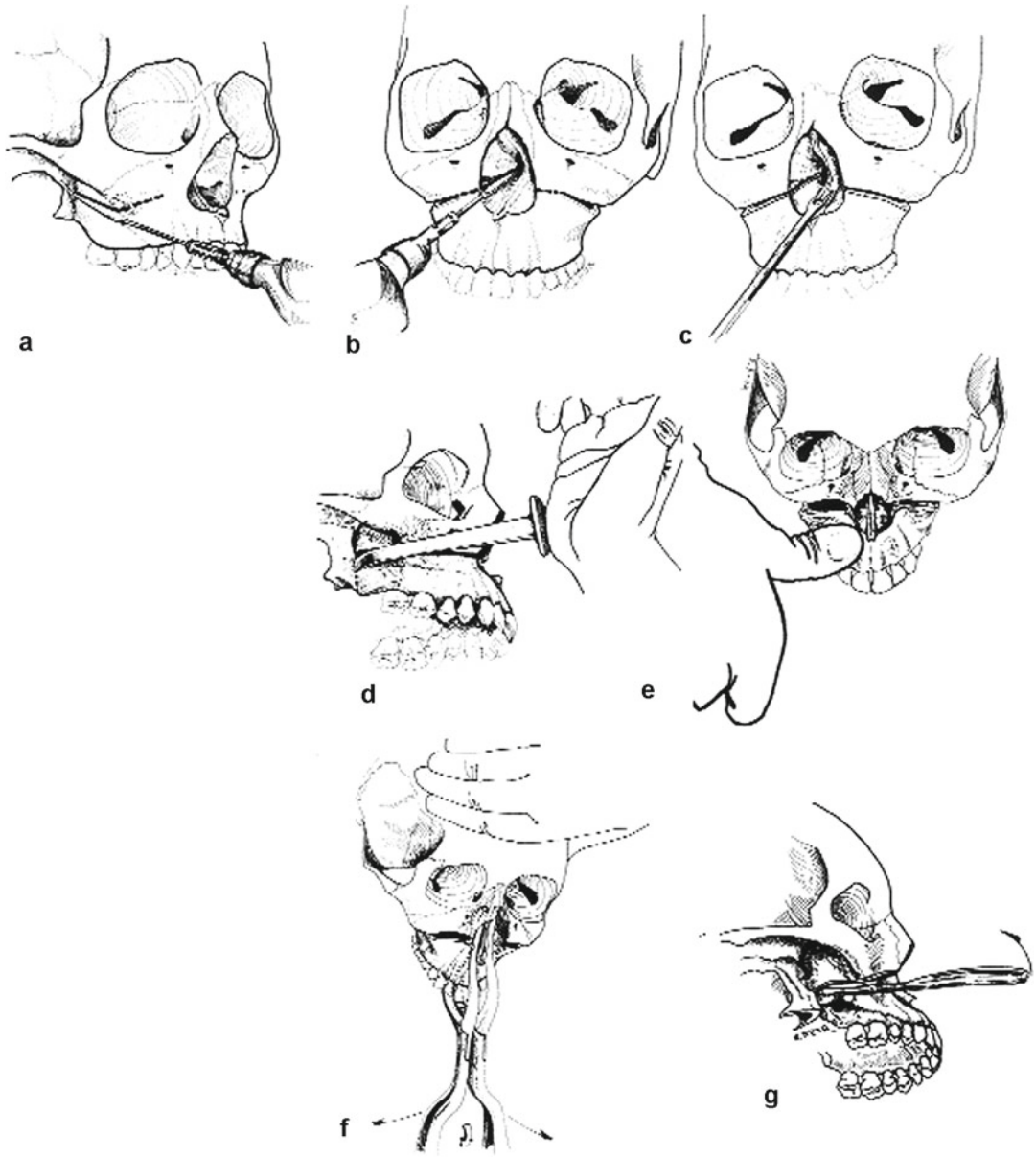

Fig. 24.6 LeFort I osteotomy (**a**) Osteotomy through lateral maxilla. (**b**) Osteotomy through the medial portion of the maxilla. (**c**) Separation of the volar from the septum with a double guarded osteotome. (**d**) Pterygomaxillary maxillary disjunction with a curved Tessier osteotome. (**e**) Digital down fracture of the osteotomized segment. (**f**) Further mobilization with a row of forceps* if necessary. (**g**) Retrotuberocity anterior levering with a blunt elevator

24.4 Technique (Fig. 24.6)

Unless work is to be done on the nose, a nasal intubation is used. If the premaxilla is absent, an oral tube can be used and simply brought through the central empty space. Schendel et al. (1979) described a technique whereby they brought an oral tube behind the maxillary tuberosity; however, it seems that this method is difficult unless a major advancement is to be performed. It also precludes putting a bone graft in the pterygomaxillary gap. It is better to perform the lip and nasal work surgery at a separate sitting.

If the alveolus is intact, and expansion of the maxilla is not required, the LeFort I osteotomy is an easy procedure. The nonintubated nostril is packed with cocaine/epinephrine-impregnated gauze, as for a rhinoplasty, and the upper labial sulcus is infiltrated with a 1: 200,000 epinephrine/hyaluronidase solution. The incision is made above the reflection of the sulcus, sparing the frenulum. The mucosal incision does not extend beyond the first molar. Subperiosteal dissection of the anterior maxilla is carried out to the infraorbital rims, visualizing the infraorbital nerves, and then taken posteriorly beneath the mucoperiosteal tunnel to the pterygomaxillary space using a Cushing elevator. If the dissection is strictly subperiosteal, there is no bothersome exposure of the buccal fat. The piriform aperture is dissected, sometimes removing a portion of the nasal spine, and the nasal mucoperiosteum is dissected back to the hard palate–soft palate junction. The septum either can be separated bluntly from the vomer or a guarded osteotome can be used. The osteotomy is performed largely with the reciprocating saw, starting laterally in the thick bone beneath the buttress of the zygoma and proceeding medially through thinner bone (Fig. 24.6a, b). The osteotomy through the piriform aperture and medial wall of the antrum is done with the saw blade pointed laterally (Fig. 24.6b, c). Section of the palatine bone, the sole attachment of the maxillary tuberosity to the pterygoid plate of the sphenoid, is done with either a curved Dautrey osteotome or the somewhat larger Kawamoto osteotome (Fig. 24.6d). The lateral osteotomy can be taken a bit further back by a few taps on a straight osteotome, and the medial antral wall can be further sectioned with a guarded nasal osteotome. At this point, the only remaining attachment of the lower maxillary segment is the posterior wall of the antrum, and firm, downward finger pressure on the maxilla is usually enough to produce a down-fracture (Fig. 24.6e). If not, the Rowe forceps can be inserted underneath the nasal mucosa and the maxilla completely mobilized with a downward and side-to-side motion. It can be further mobilized with a blunt elevator used as a lever (Fig. 24.6f, g).

The maxilla is then placed in the desired occlusal relation with the mandible, and both jaws are placed in the desired relation with the rest of the face. An autogenous iliac or cranial bone graft is used when the face is to be lengthened, when the degree of advancement is more than 5–6 mm, or when the patient has a cleft. If the maxilla is shortened, the resected bone is placed over the osteotomy lines.

Sometimes the alveolus is intact, but the maxilla may need to be expanded as in a cleft patient who has had a bone graft of the alveolar cleft. This procedure is easily performed from above, and the palatal mucosa is kept intact if possible. The section is performed with the reciprocating saw, and an elevator is inserted to gently pry the two segments apart. The expansion forceps can be used if required. If the palatal mucosa absolutely prevents expansion, it is divided, creating an alveolar and anterior palatal cleft (Fig. 24.6).

If there is an alveolar cleft to begin with, the two maxillary segments are handled independently and brought into proper occlusion with the mandible. The palatal cleft–nasal floor defect is bone-grafted, and if necessary a transportation flap is developed from the buccal sulcus (Burian) to close the palatal defect. In rare instances a tongue flap is required. The nasal lining, which was carefully dissected at the beginning, is closed before the palatal bone graft is inserted (Fig. 24.7).

The procedure has now reached a stage that is the same regardless of whether the alveolus was initially intact.

Osteosynthesis wires are passed between the upper and lower portions of the maxilla, and suspensory wires are taken through the thick bone beneath the zygomatic buttress laterally.

If bone grafts are required, they are placed either between or over the osteosynthesis anteriorly. If the advancement is more than 5–6 mm, bone grafts can be wedged into the pterygomaxillary gap. This step is facilitated by using a traction wire placed through the thick bone beneath the nasal spine. The wire is used to pull the maxilla to the opposite side, which opensthe gap and allows impaction of the bone graft. The suspensory wires from the buttress are brought through the inferior mucosal flap and wired to the upper arch bar. If further suspensory wires are needed,

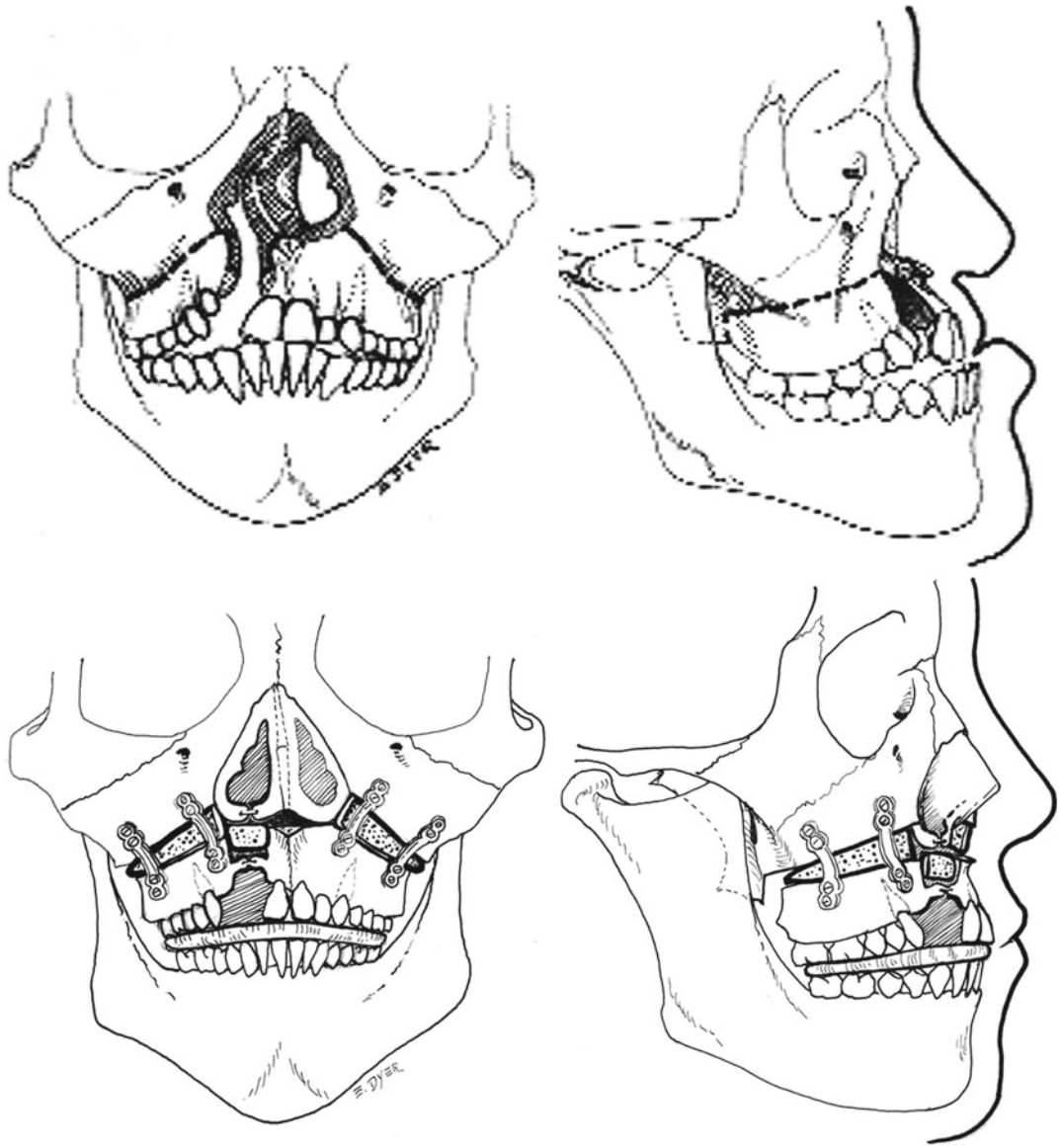

Fig. 24.7 LeFort I osteotomy in a patient with a unilateral cleft. Autogenous bone grafts are always used, not only to help consolidation and prevent relapse but to improve appearance. I use an iliac or cranial bone graft on all cleft patients, as these patients are likely to have a relapse. Generally, the bone can also be used as an onlay to fill out a deficient maxilla. If the advancement is less than 5–6 mm, bone is placed only over the anterior osteotomies and in the alveolar and palatal cleft, if present. The use of anything other than a fresh autogenous bone graft is unwise. It takes about 15 min to harvest the needed amount of iliac or cranial bone, and the patient will be comfortable as far as the hip is concerned within 1–2 weeks. By this time, autogenous grafts will have consolidated. Consolidation with cadaver or demineralized bone or with hydroxylapatite may require 6 months, if indeed it occurs at all. Are we doing the patient a favor here by "sparing" him or her an iliac bone graft? Probably not

they can be brought down from the infraorbital rim. Circumzygomatic wires are almost never used: They pull the maxilla back, they are too long (long wires can "stretch" more than short wires), and they do not prevent the anterior maxilla from rocking downward (Figs. 24.8 and 24.9).

Like the sagittal splitting procedure for the mandible, the LeFort I osteotomy, once mastered,

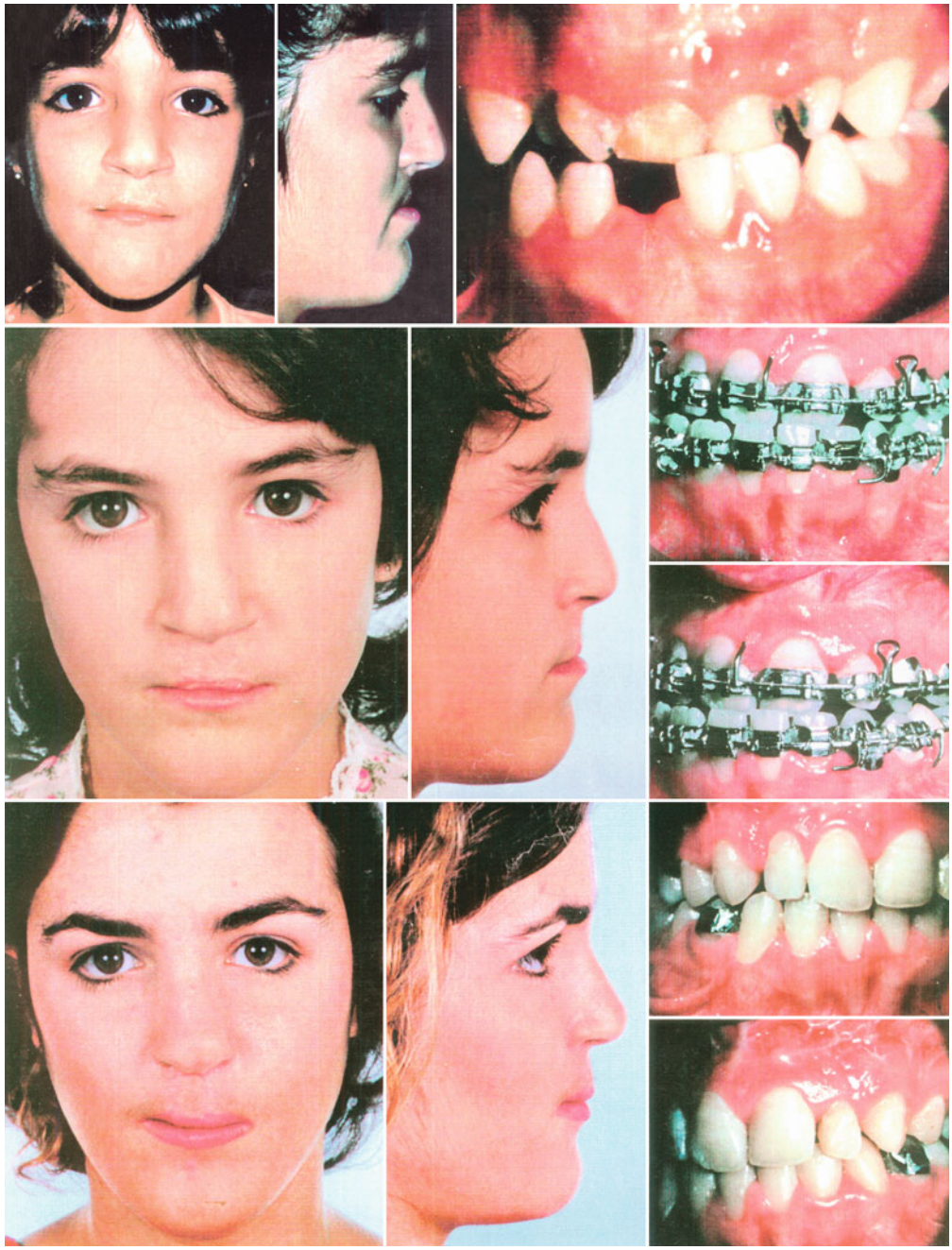

Fig. 24.8 Case JR-AT-94. Midfacial advancement to correct an anterior crossbite and improve facial aesthetics and occlusion. Surgical history: this patient was originally treated in Honduras. Lip closure at 2 months, and palatal closure at 8 months of age. No attempt was made to close the palatal fistulae. Oral facial evaluation: Midfacial hypoplasia with an anterior and right buccal crossbite. Treatment plan: Advance and expand both dental arches and close the palatal fistulae when surgically advancing the maxilla. Results: Although the patient had a cleft of the lip and palate with twinned deciduous lateral incisors in the line of the cleft, one of the permanent lateral incisors was well developed, the other malformed. The malformed impacted tooth was extracted, a secondary alveolar cranial bone graft, and fistula closing procedure were all performed at the same time (13–11). A LeFort I maxillary advancement brought the teeth into Class I relationship

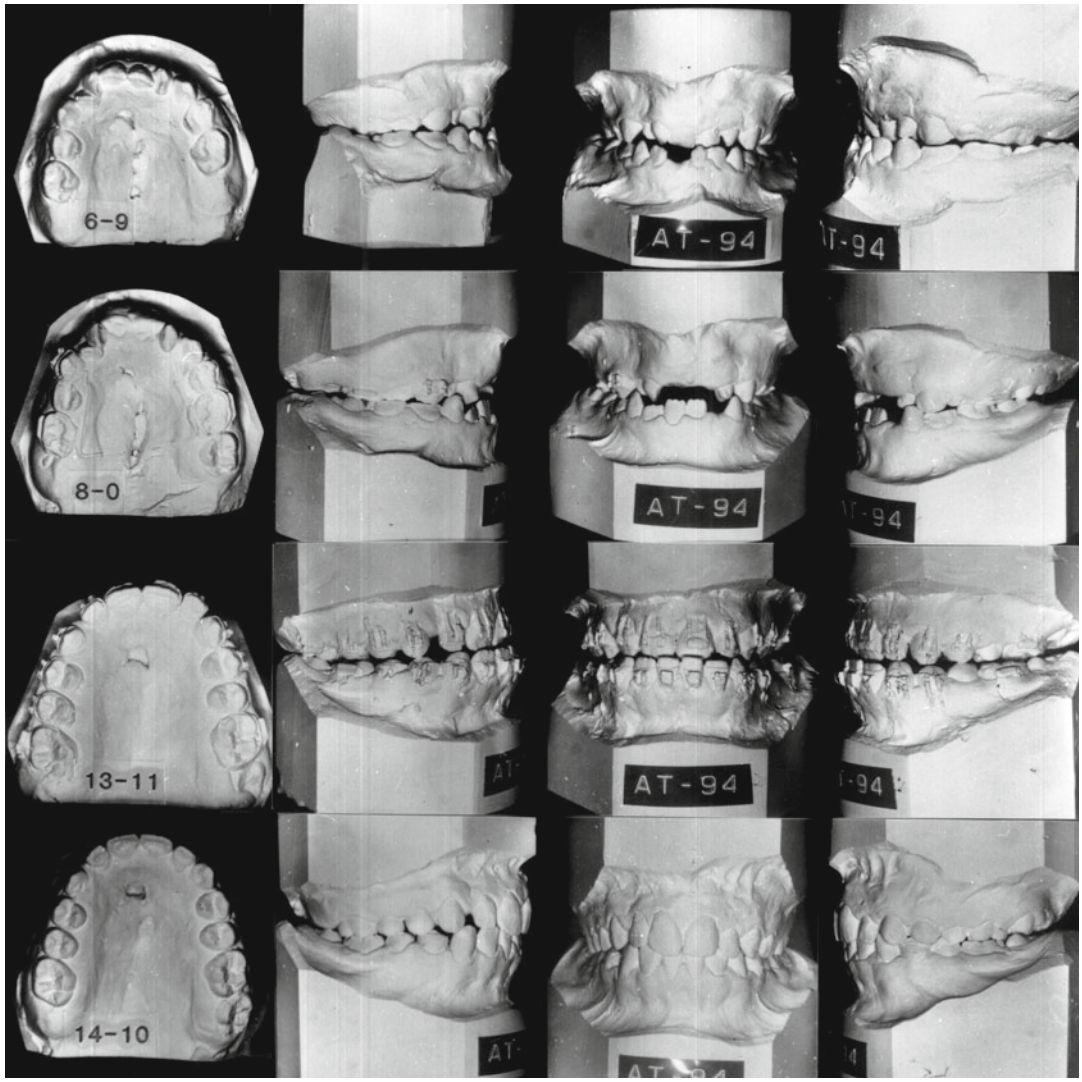

Fig. 24.9 Case JR (AT-94). Surgical maxillary advancement (LeFort I). Pre-and postmaxillary advancement

can provide a solution to a number of maxillary problems. After the horizontal osteotomy, downfracture, and mobilization, the maxilla can be:

1. Advanced directly with or without a bone graft (noncleft class III patient).
2. Advanced, or advanced and expanded transversely, with a bone graft (cleft patient).
3. Moved superiorly after resection of a measured amount of maxilla above the horizontal osteotomy (long face: vertical maxillary excess). Differential upward movement can also be used to close an anterior open bite.
4. Moved inferiorly, with a bone graft (short face: vertical maxillary deficiency).
5. Sectioned into multiple segments with dental extractions: Wassmund or Schuchardt procedure from above.
6. Moved directly backward. It is difficult to do (the resection should be of the maxillary tuberosity after extraction of the third molars rather than of the pterygoid plate). The same result can generally be achieved by an associated segmental osteotomy performed more anteriorly.

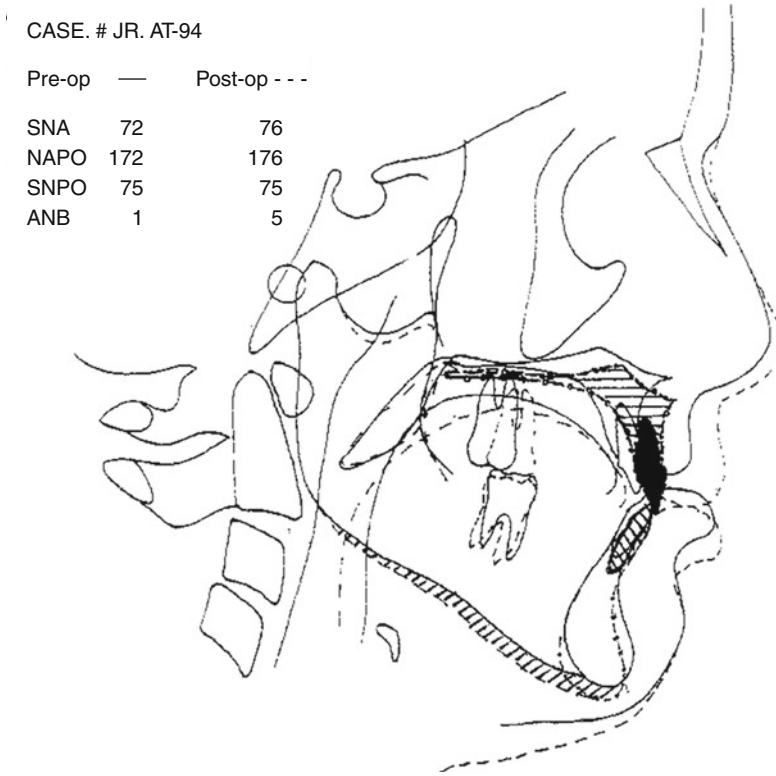

Fig. 24.10 Case JR. Lateral cephalometric tracings before and after LeFort I maxillary advancement and inferior positioning. The mandible autorotated posteriorly reducing the mandibular prominence. Maxillary advancement moved the upper lip forward while the lower lip remained practically in the same position

24.5 Multiple Maxillary Osteotomies

With the maxilla in the down-fractured position, multiple osteotomies can be performed from above, which, coupled with dental extractions, permit correction of complex malarrangements of the maxilla in one stage. The circulation to the anterior segment comes entirely through the palatal mucoperiosteum, and one must be certain that there are no protrusive edges from the occlusal splint to impinge on the anterior palate. Any number of transverse or sagittal osteotomies can be performed depending on the requirements of the individual case.

Class III malocclusion following isolated cleft palate repair. The patient had a large palatal cleft that was closed at 18 months of age with a von Langenbeck repair. Because of severe velopharyngeal insufficiency, a bipedicle island flap incorporating both greater palatine arteries was positioned at age 7, along with a palatal pushback. Following this procedure speech has been normal, but the patient showed subsequent restriction of maxillary growth, both transversely and in the posteroanterior direction. After preliminary orthodontic expansion of the maxilla, a LeFort I osteotomy was done at age 16. The maxilla was advanced without difficulty and stabilized in the usual fashion with iliac bone grafts. The mucosa beneath the horizontal incision at the end of the procedure was pale blue, which is not unusual in a cleft patient. However, over the next few days the upper portion of the mucosal flap proceeded to slough in a way that suggested lack of blood supply more than infection. Healing eventually occurred, with partial obliteration of the upper labial sulcus and partial exposure of the roots of the central incisors, which required periodontal treatment. Despite vigorous and sustained postoperative orthodontic treatment, the occlusion relapsed to a Class III relation.

Figure 24.10 shows a patient who has had a unilateral palatal island flap can safely undergo a LeFort I osteotomy, but as this patient demonstrates, it is not a safe procedure in one who has had diversion of both greater palatine arteries. Either the mandible should be moved back or a

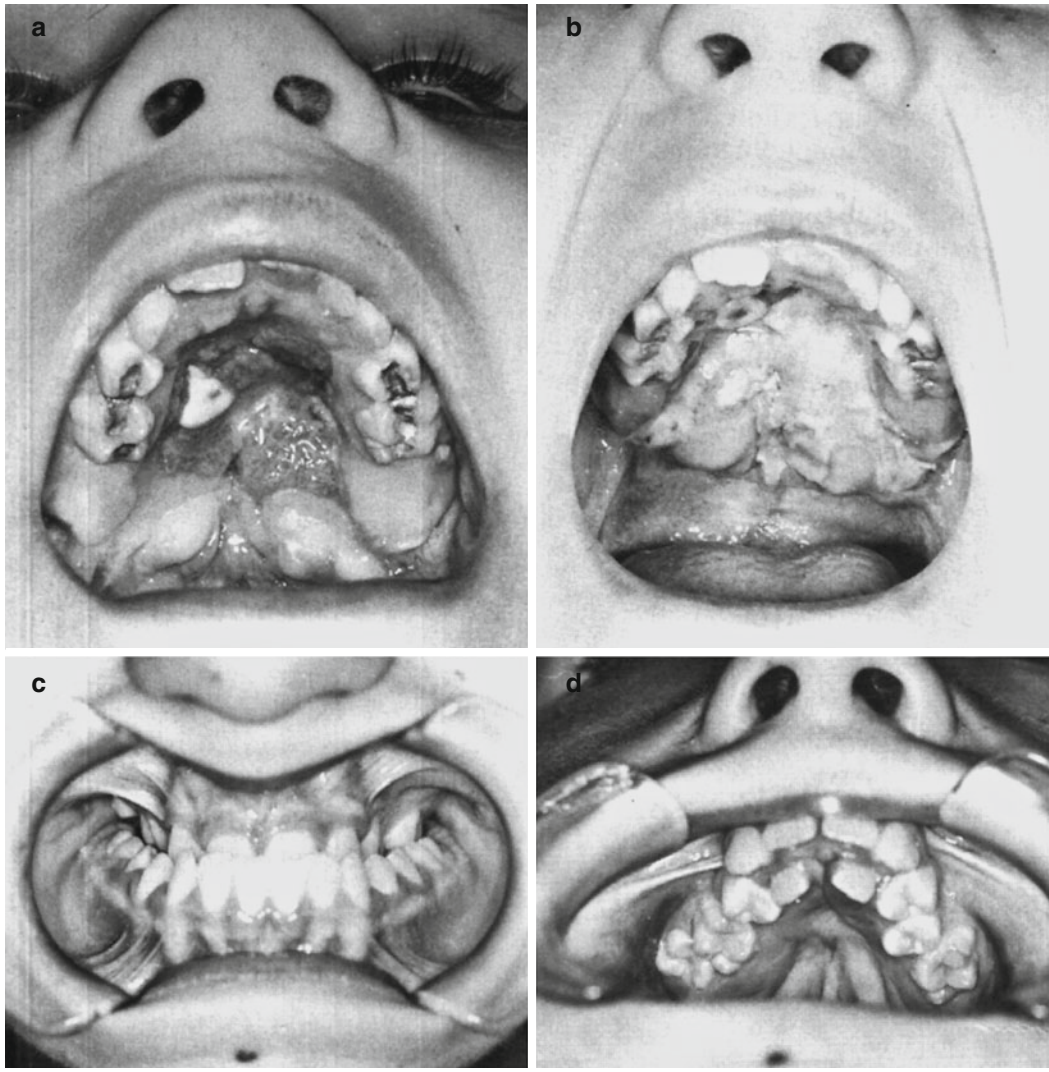

Fig. 24.11 (**a**–**j**) Class III malocclusion following isolated cleft palate repair. The patient had a large palatal cleft that was closed at 18 months of age with a von Langenbeck repair. Because of severe velopharyngeal insufficiency, a bipedicle island flap incorporating both greater palatine arteries was positioned at age 7, along with a palatal push-back. Following this procedure speech has been normal, but the patient showed subsequent restriction of maxillary growth, both transversely and in the posteroanterior direction. After preliminary orthodontic expansion of the maxilla, a LeFort I osteotomy was done at age 16. The maxilla was advanced without difficulty and stabilized in the usual fashion with iliac bone grafts. The mucosa beneath the horizontal incision at the end of the procedure was pale blue, which is not unusual in a cleft patient. However, over the next few days the upper portion of the mucosal flap proceeded to slough in a way that suggested lack of blood supply more than infection. Healing eventually occurred, with partial obliteration of the upper labial sulcus and partial exposure of the roots of the central incisors, which required periodontal treatment. Despite vigorous and sustained postoperative orthodontic treatment, the occlusion relapsed to a Class III relationship. Figure 24.10 shows a patient who has had a unilateral palatal island flap can safely undergo a LeFort I osteotomy, but as this patient demonstrates, it is not a safe procedure in one who has had diversion of both greater palatine arteries. Either the mandible should be moved back or a LeFort II osteotomy performed in which most of the anterior vestibular mucoperiosteum is preserved. (**a**, **b**) A raw area of the anterior palate following placement of a bipedicle island flap showing rapid healing by secondary epithelialization. (**c**, **d**) Subsequent maxillary deformity 5 years later. Note the transpalatal scar band

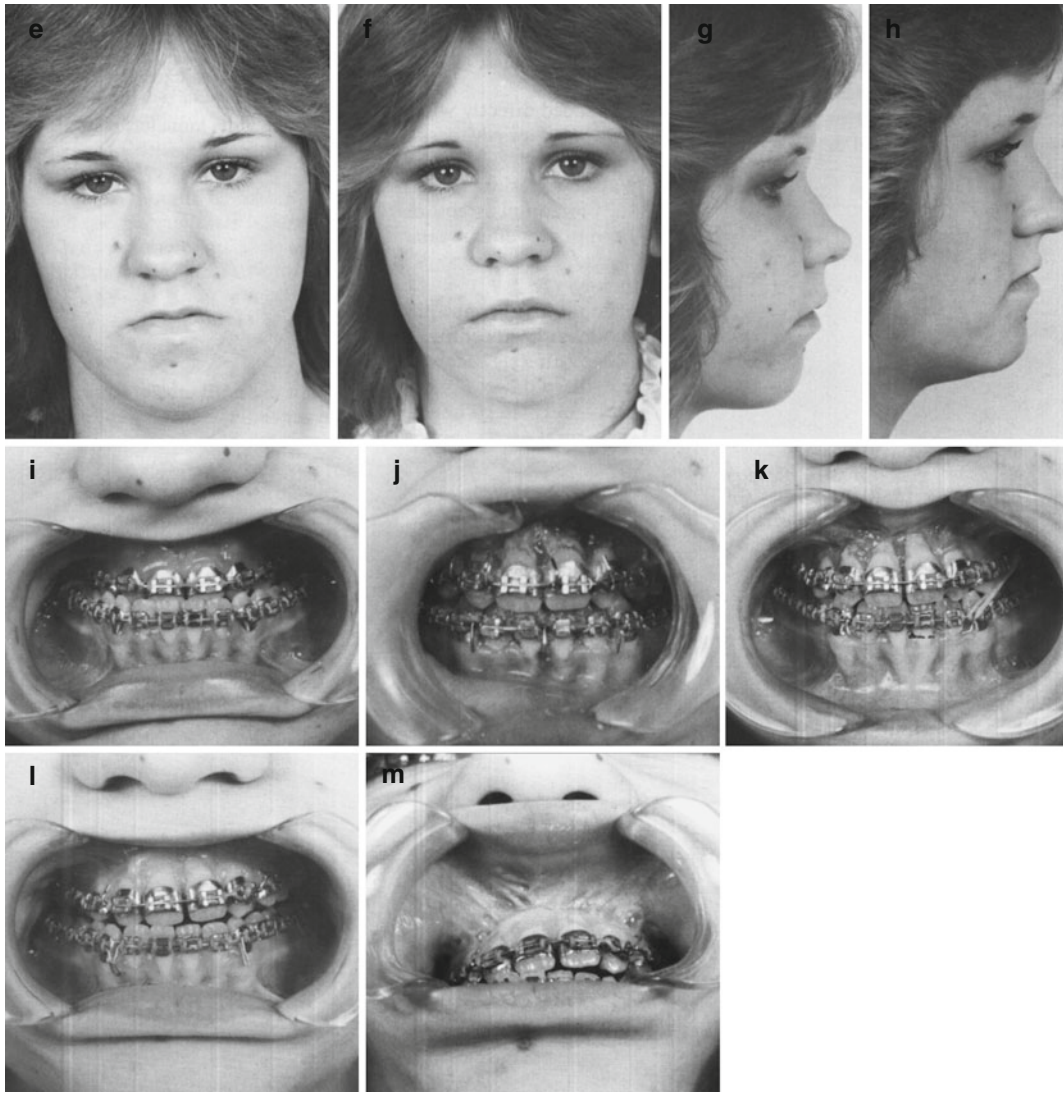

Fig. 24.11 (*continued*) (**e–j**) Before and after LeFort I osteotomy. Note the appearance of the gingiva above the maxillary incisors. (**k**) There is partial exposure of the roots of the central incisors. (**l**, **m**) Degree of relapse after 6 months, with partial obliteration of the upper labial sulcus

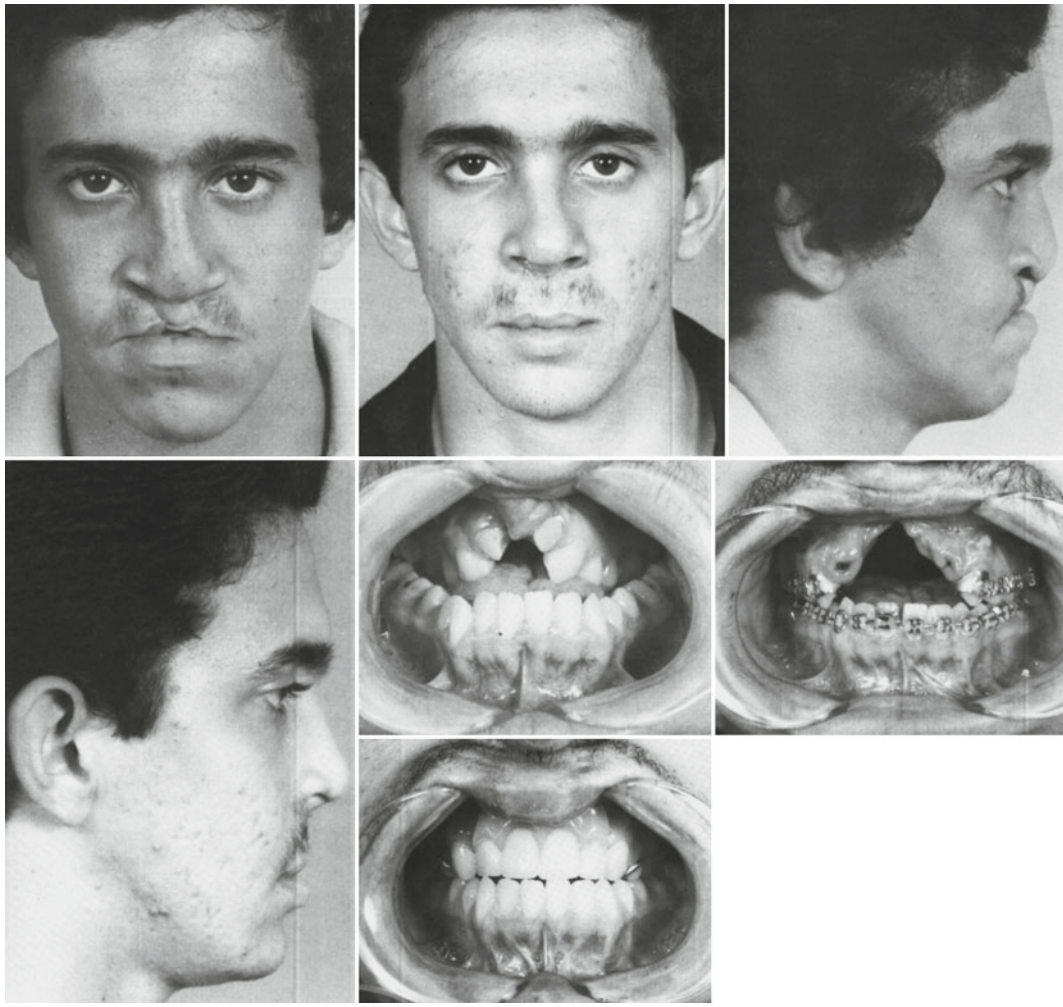

Fig. 24.12 Bilateral cleft with absent premaxilla. Original treatment had been administered in Cuba, and the premaxilla had been excised at the time of the lip closure. Severe maxillary collapse ensued. A LeFort I osteotomy and maxillary expansion was performed, with bone grafting of the palatal cleft. A prosthesis replaced the missing teeth. Following this, Ralph Millard proceeded with a cleft lip rhinoplasty and Abbe flap

LeFort II osteotomy performed in which most of the anterior vestibular mucoperiosteum is preserved. Figure 24.10 shows a raw area of the anterior palate following placement of a bipedicle island flap showing rapid healing by secondary epithelialization. Figure 24.10 shows subsequent maxillary deformity 5 years later. Note the transpalatal scar band.

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Part XI

Orthognathic Surgery

Cleft Jaw Deformities and Their Management

25

Jeffrey C. Posnick

25.1 Facial Growth Implications of Cleft Palate Repair in Infancy

25.1.1 Unilateral Cleft Lip and Palate

Ross completed a multicentered long-term facial growth study to assess the need for orthognathic surgery in individuals born with complete UCLP, having undergone primary lip and palate repair in childhood (Ross 1987; Abyholm et al. 1981). He concluded that even by the most conservative standards and in conjunction with maximum compensating orthodontic camouflage maneuvers, at least 25 % of adolescents with UCLP require orthognathic surgery to achieve even the limited objective of an acceptable occlusion. Their research indicated that only 25 % of UCLP adolescents had near-normal maxillary growth. The other 50 % were in a borderline category with some degree of maxillary hypoplasia. Ross stated that individuals born with cleft lip and palate have intrinsic deficiency in the midfacial skeleton that is made worse by operations. More recently, Mulliken and colleagues reviewed the prevalence of LeFort I osteotomy in patients with cleft lip/palate treated at Boston Children's Hospital (Good et al. 2007). They found that 48 % of their repaired UCLP patients required orthognathic surgery. Similarly, the Hospital for Sick Children (Toronto, Canada) found that 48.3 % of their complete UCLP patients required orthognathic surgery. When they looked at all patients with UCLP referred to their center, they found that 59.4 % needed jaw reconstruction (Fig. 25.1) (Daskalogiannakis and Mehta 2009). Other earlier research confirms and compliments these findings (Capelozza Filho et al. 1996; Correa Normando et al. 1992; DeLuke et al. 1997; Filho 1996; Friede and Lilja 1994; Fudalej et al. 2009a, b; Gnoinski 1987; Jorgenson et al. 1984; Linton 1998; Mars et al. 1987, 1992; Mars and Houston 1990; Mølsted et al. 2005; Motohashi et al. 1994; Nanda 1988; Nollet et al. 2005; Palmer et al. 1969; Roberts et al. 1996; Schnitt et al. 2004; Smabel 1994; Susami et al. 2006).

Reconstruction of the alveolar process in UCLP patients is an essential part of cleft care. Accomplishing this goal provides support for the alar base of the nose, the teeth in the cleft region, and the periodontium of those teeth. There are three basic methods described for the closure of the cleft alveolus. The surgical options include: primary bone grafting, secondary bone grafting, and gingival periosteoplasty (GPP). The method of primary bone grafting is now

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S. Berkowitz (ed.), *Cleft Lip and Palate*,
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Fig. 25.1 This is a senior high school student who was born with a complete UCLP (*left side*). He underwent lip and palate repair at another institution. He was then referred to this surgeon and underwent effective bone grafting and fistula closure in the mixed dentition. He developed a jaw deformity characterized by maxillary deficiency and secondary deformities of the mandible and intranasal cavity. He underwent a combined orthodontic and surgical approach. Orthodontic (dental) decompensation included cleft-dental gap (absent lateral incisor) closure. He then underwent reconstruction including maxillary LeFort I osteotomy (horizontal advancement, vertical shortening, midline correction, cant correction, clockwise rotation); sagittal split ramus osteotomies (clockwise rotation, asymmetry correction); osseous genioplasty (vertical shortening); and septoplasty/inferior turbinate reduction/recontouring of floor of nose. Six months after successful orthognathic surgery, he underwent cleft rhinoplasty including rib cartilage (caudal strut) grafting. (**a**) He is shown in the mixed dentition prior to bone grafting. (**b**) Frontal views in repose before and after reconstruction

Fig. 25.1 (*continued*) (**c**) Frontal views with smile before and after reconstruction. (**d**) Oblique facial views before and after reconstruction

Fig. 25.1 (*continued*) (**e**) Profile views before and after reconstruction. (**f**) Occlusal views prior to orthodontics, with orthodontic dental decompensation in progress, and after reconstruction

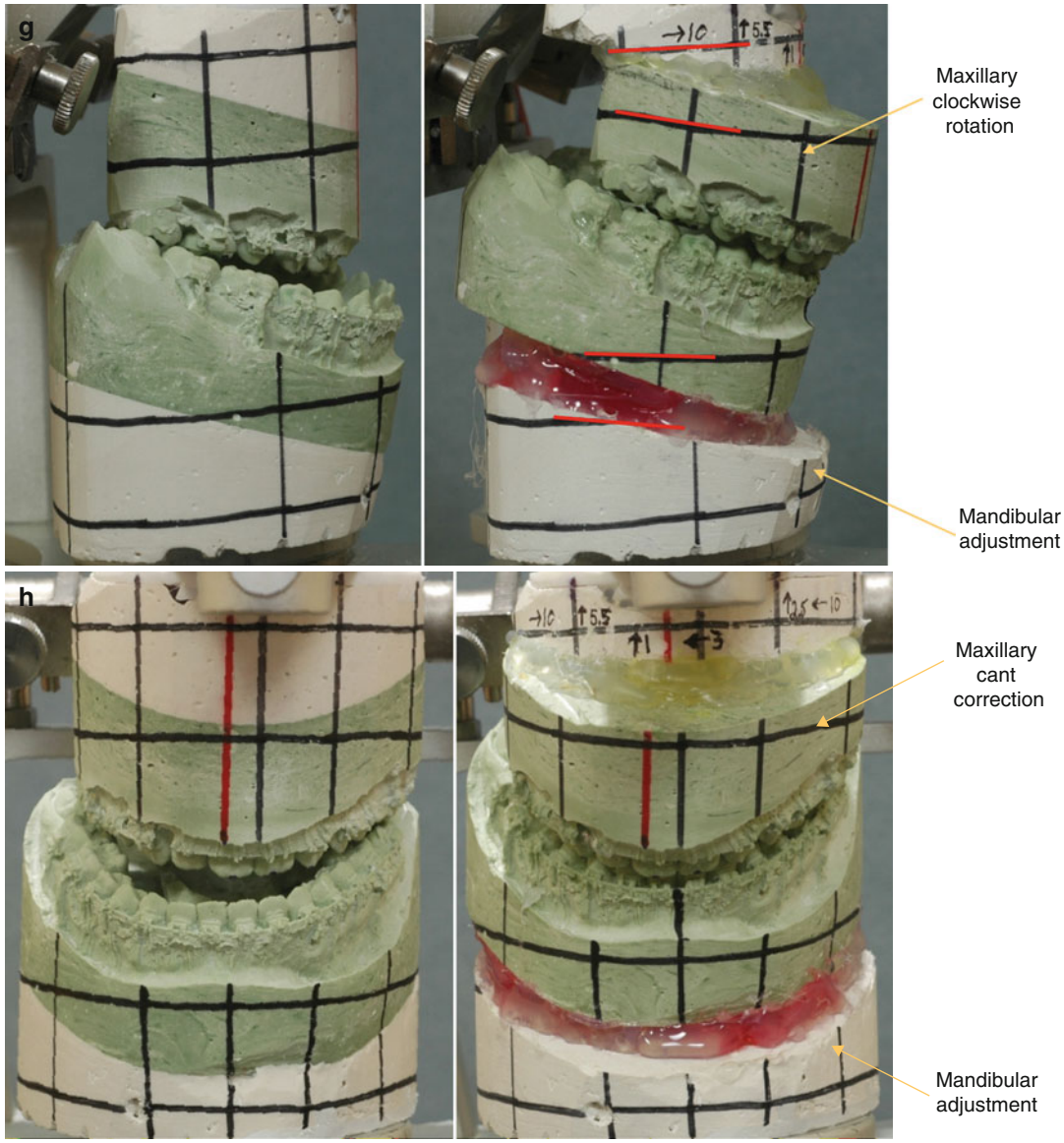


Fig. 25.1 (*continued*) (**g**, **h**) Articulated dental casts before and after reconstruction

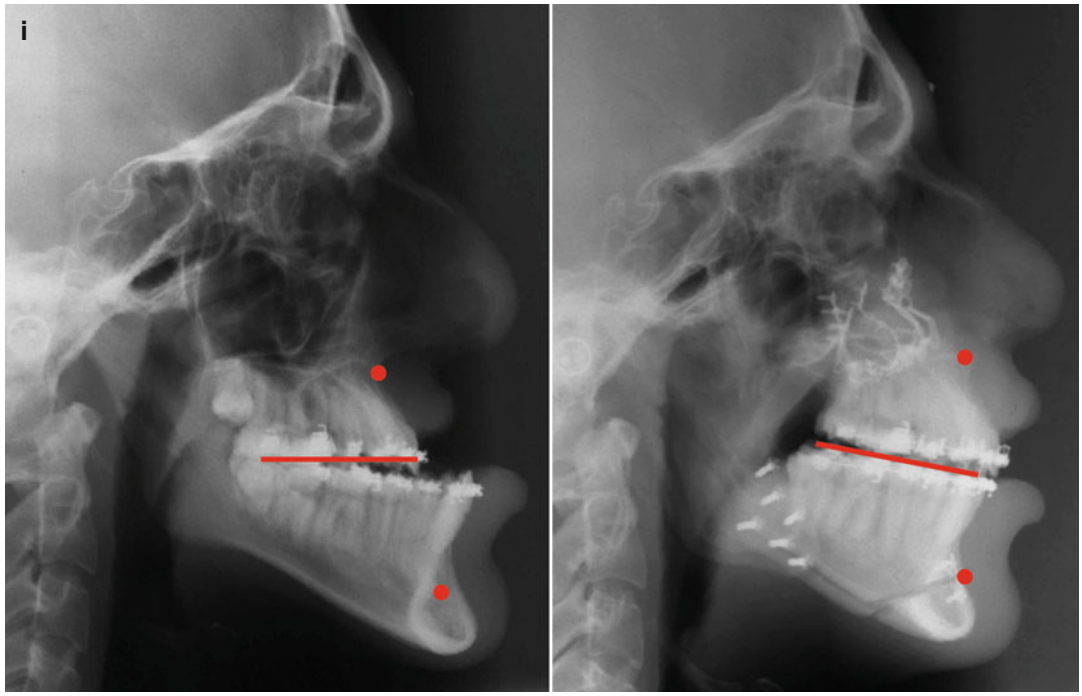


Fig. 25.1 (*continued*) (**i**) Lateral Cephalometric radiographs before and after reconstruction

generally recognized to result in severe midface growth disturbance and has therefore been universally abandoned throughout the world (Hathaway et al. 1999; Posnick 1991a, b; Rosenstein 1997). The use of secondary (mixed dentition) bone grafting (Abyholm et al. 1981; Boyne and Sands 1972; Daskalogiannakis and Ross 1997; Hall and Posnick 1984) is generally recognized as an effective method to avoid the problem of midface growth disturbance and is successfully achieved to support the alar base, to provide bone for eruption of the canine through the cleft side, and to establish effective periodontal support. The technique of gingival periosteoplasty (GPP) was first described by Skoog in 1965 as a method to achieve fusion across the cleft alveolus at the time of lip repair (Skoog 1965). The goal was to "remove" the cleft in infancy with the hope that no harm would result. Millard combined the "Latham device" to position the alveolar ridges close to one another before going forward with the GPP procedure at the time of cleft lip repair (Millard et al. 1990, 1999). Grayson and Cutting later proposed the use of a nasoalveolar molding (NAM) device prior to GPP and primary lip repair to accomplish the same alveolar ridge fusion (Grayson and Cutting 2011). Since the late 1970s, clinical studies have been carried out to assess the long-term effects of GPP on midfacial growth in cleft patients. The studies conducted by Millard et al. (1999), Mølsted et al. (2005), Berkowitz and colleagues (2004), Matic and Powers (2008), Renkielska et al. (2005), Henkel and Gundlach (1997), and Tomanova and Mullerova (1994) have all found that patients treated with GPP had poor occlusal relationships with a high need for orthognathic surgery in the teenage years. More recently, Hsin-Yi Hsieh and colleagues completed a retrospective clinical study to evaluate the effects of GPP on facial growth of patients with UCLP (Hsieh et al. 2010). This was a well-done clinical study of a consecutive series of patients that were placed in two study groups, those receiving NAM treatment with GPP and those receiving NAM treatment without GPP. The use of gingival periosteoplasty at the time of lip repair was found to have significant negative effects on the maxillary position (SNA), intermaxillary position (ANB), maxillary length (PMP-ANS), and maxillary alveolar length (PMP-AN) at the age

of 5 years. The authors concluded that in patients with an alveolar *cleft*, the sagittal growth of the maxilla was significantly adversely affected by the gingival periosteoplasty (GPP) procedure.

25.1.2 Bilateral Cleft Lip and Palate

Mulliken and colleagues from Boston Children's Hospital found that 76.5 % of teenagers with repaired BCLP required maxillary advancement (Good et al. 2007). They showed that the need for orthognathic surgery is dependent on the severity of the cleft type as well as the number and extent of previous operations performed. Results from the Hospital for Sick Children (Toronto, Canada) showed that 65.1 % of their BCLP patients required or underwent orthognathic surgery, while 70 % of BCLP patients referred to their team sometime after cleft lip repair were found to require orthognathic surgery (Daskalogiannakis and Mehta 2009). David et al. from the Cleft Craniofacial Unit in Adelaide, Australia, followed a consecutive group of BCLP patients from birth to maturity and determined the need for orthognathic surgery (David et al. 2006). A skeletal class III malocclusion requiring orthognathic correction was present in 17 of the 19 consecutive patients followed to the age of 18 years (89.5 %) (Figs. 25.2, 25.3, and 25.4). Other earlier research confirms and compliments these findings (Harada et al. 2002; Lisson and Trankmann 1997; Pruzansky 1985).

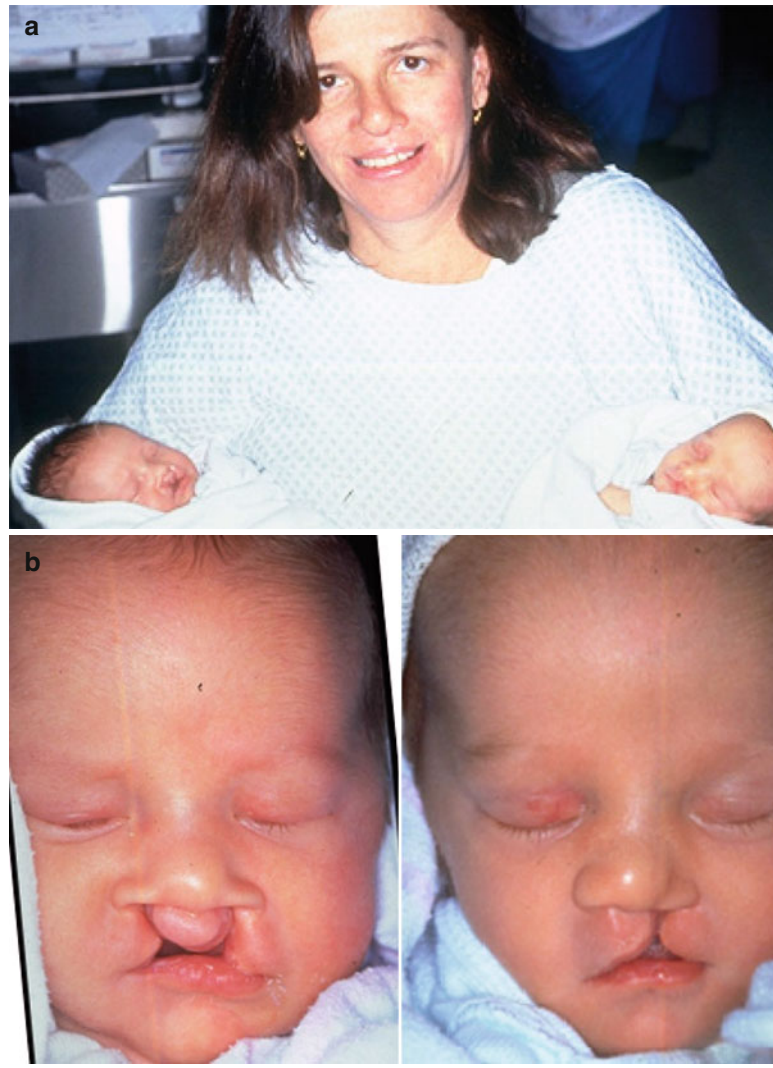
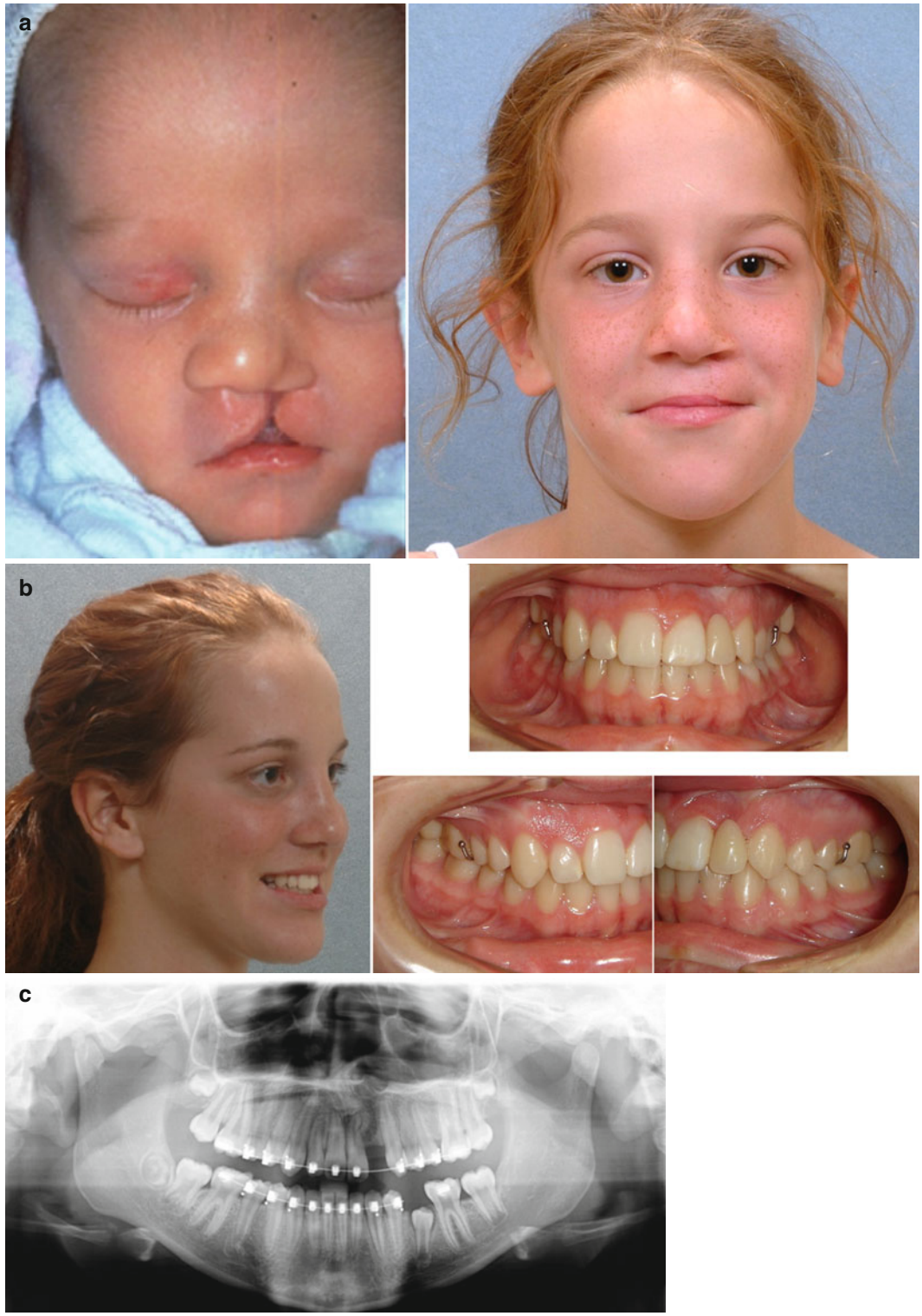


Fig. 25.2 A 40-year-old woman born with BCL and pitting of the lower (central) lip. At the time of pregnancy, ultrasound confirmed twins; one of which was suspected of having BCLP and the other to have UCLP. This was documented at the time of delivery. (**a**) Family with van der Woude syndrome including mother with repaired BCLP and newborn twins. (**b**) Twin A with BCLP and twin B with UCLP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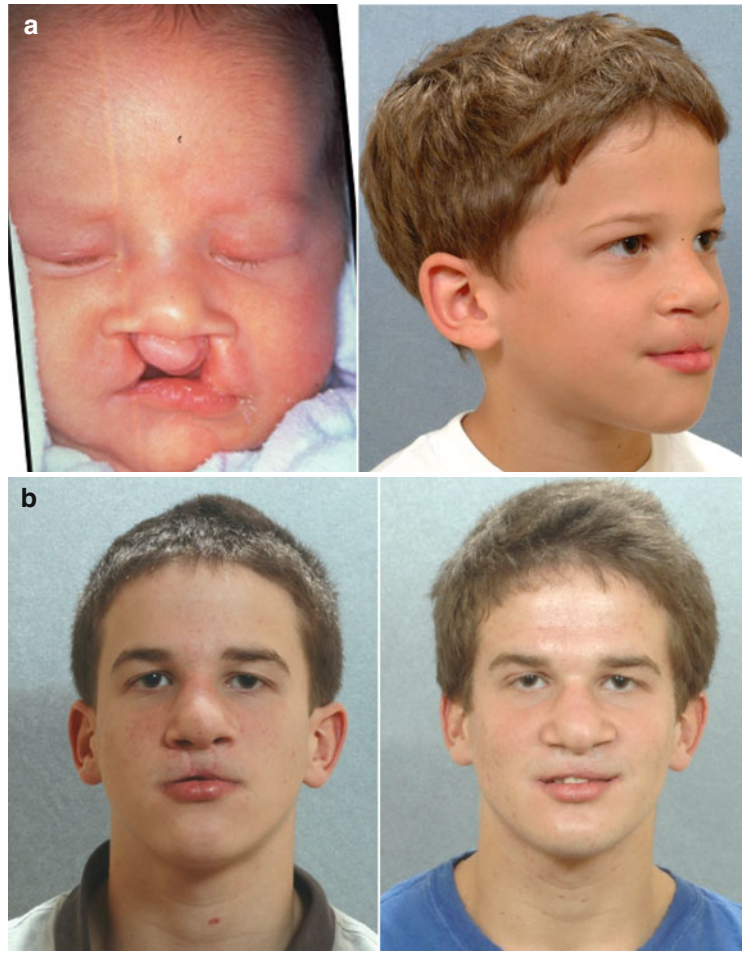


Fig. 25.4 A child born with van der Woude syndrome including BCLP and lower lip pits (see Fig. 25.3). He underwent lip and palate repair including lower lip pit excisions in childhood. He underwent successful mixed dentition grafting and fistula closure. As a teenager, he demonstrates maxillary hypoplasia with secondary deformities of the mandible and chin region. He then underwent a combined orthodontic and orthognathic/intranasal surgical approach. The procedures include standard LeFort I osteotomy (vertical lengthening, horizontal advancement, clockwise rotation with interpositional grafting); bilateral sagittal split ramus osteotomies (clockwise rotation); osseous genioplasty (horizontal advancement); and septoplasty/inferior turbinate reduction/nasal floor recontouring. (**a**) He is shown at the time of birth and then just prior to mixed dentition grafting. (**b**) Frontal views in repose before and after reconstruction

Fig. 25.3 A 16-year-old girl born with van der Woude syndrome including UCLP and lower lip pits (see Fig. 25.3). She underwent lip and palate repair in early childhood followed by successful bone grafting and fistula closure in the mixed dentition. She was with normal maxillary growth and underwent standard orthodontic treatment with maintenance of the cleft-dental gap. She underwent an open rhinoplasty including use of a rib cartilage (caudal strut) graft at 15 years of age. There is sufficient bone volume for a dental implant. Implant placement is planned for 18 years of age. (**a**) Facial views at the time of birth and in the mixed dentition just prior to alveolar bone grafting. (**b**) Facial and occlusal views as a teenager after rhinoplasty prior to dental implant placement. (**c**) Panorex as a teenager during orthodontic treatment indicates successful bone grafting of the alveolar cleft

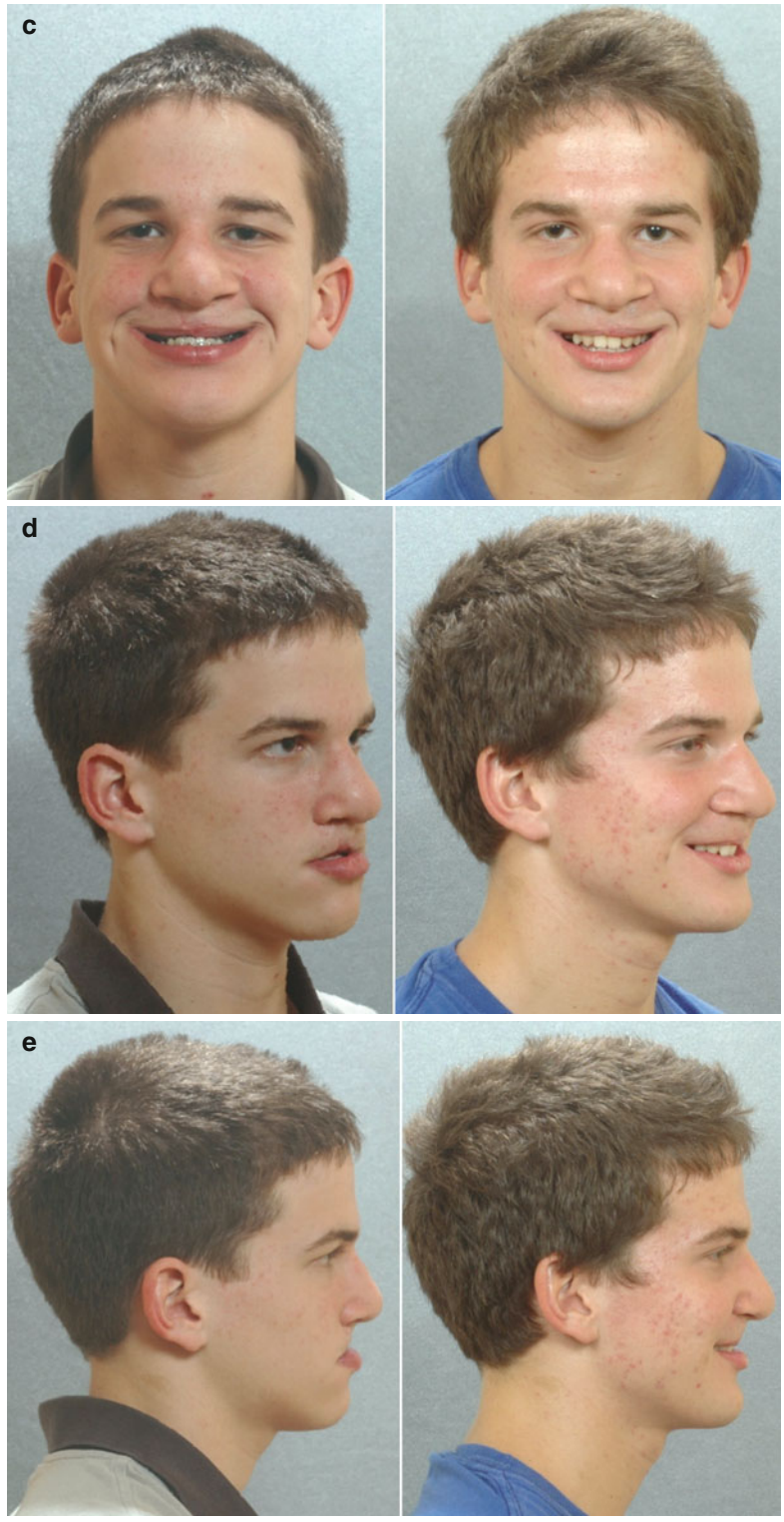
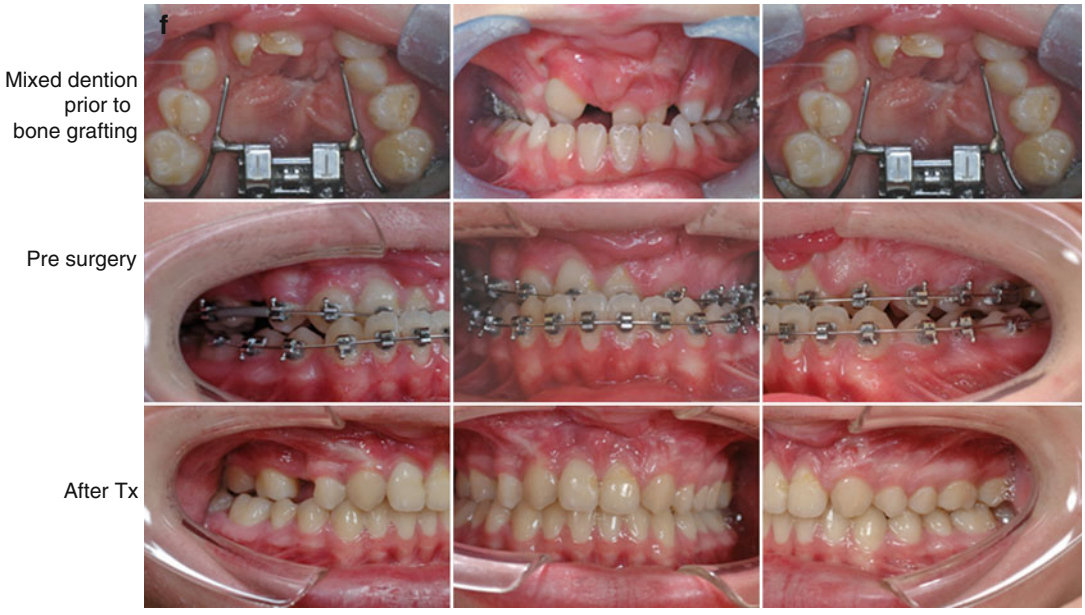


Fig. 25.4 (*continued*) (**c**) Frontal views with smile before and after reconstruction. (**d**) Oblique facial views before and after reconstruction. (**e**) Profile views before and after reconstruction



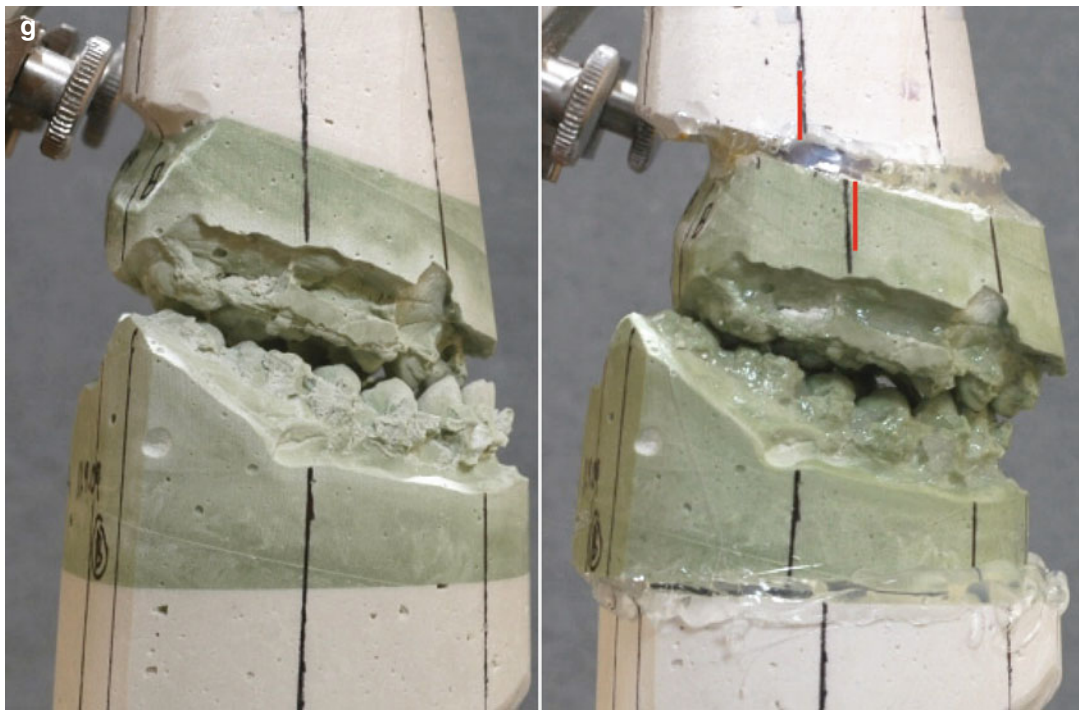


Fig. 25.4 (*continued*) (**f**) Occlusal views are shown prior to bone grafting/fistula closure, after orthodontics in preparation for orthognathic surgery and after reconstruction. The congenitally missing right maxillary bicuspid is planned for placement of a dental implant at 18 years of age. (**g**) Articulated dental casts before and after model planning

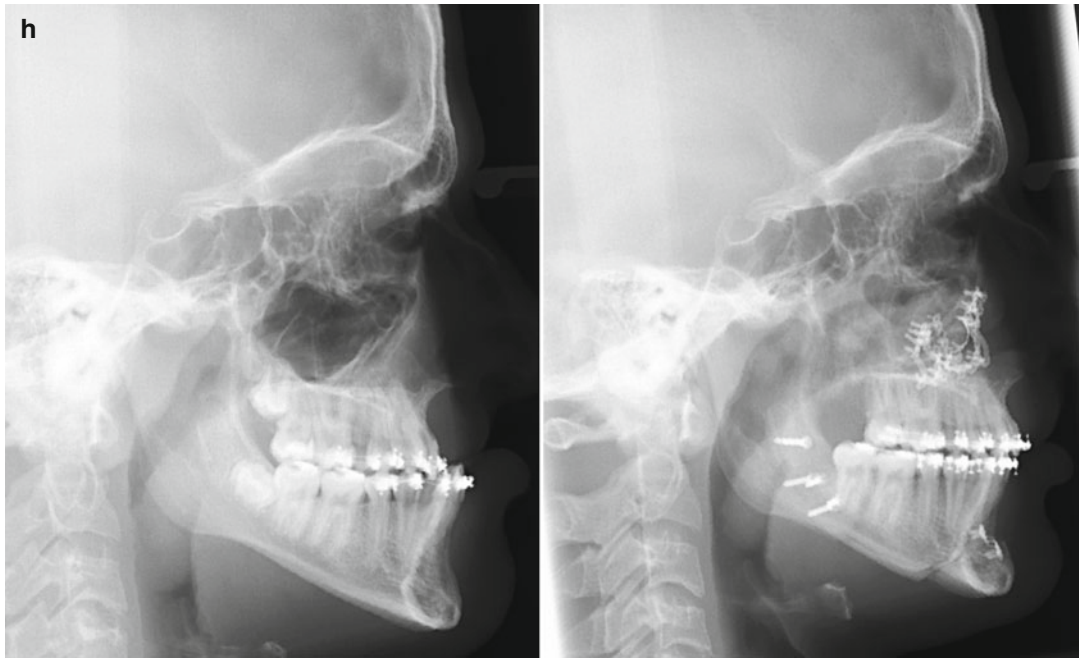


Fig. 25.4 (*continued*) (**h**) Cephalometric radiographs before and after reconstruction

25.1.3 Isolated Cleft Palate

Ross documented that at least 20 % of Caucasians with ICP who underwent repair in infancy will experience maxillary hypoplasia resulting in malocclusion that is not responsive to either traditional or compensatory orthodontic maneuvers alone (Abyholm et al. 1981). Chen et al. reported on horizontal maxillary growth in both children and adults of eastern Chinese ethnic background with unoperated and operated isolated cleft palate (Chen et al. 2009). Interestingly, the results of the unoperated mixed dentition individuals with ICP showed almost normal horizontal growth. The operated (repaired cleft palate) mixed dentition patients showed reduced length of both the maxilla and mandible and clockwise rotation of the maxillomandibular complex. Analysis of the permanent dentition groups showed reductions in maxillary length and reduced maxillary and mandibular horizontal projection. The authors concluded that for an individual born with ICP, the high incidence of maxillomandibular deficiency likely results from a combination of factors including the intrinsic primary cleft defect, secondary hypoplasia due to the surgical repair in infancy, and functional factors (e.g., effects of muscles of mastication, respiratory pattern, and mandibular rest posture (Canady et al. 1997)) (Fig. 25.5).

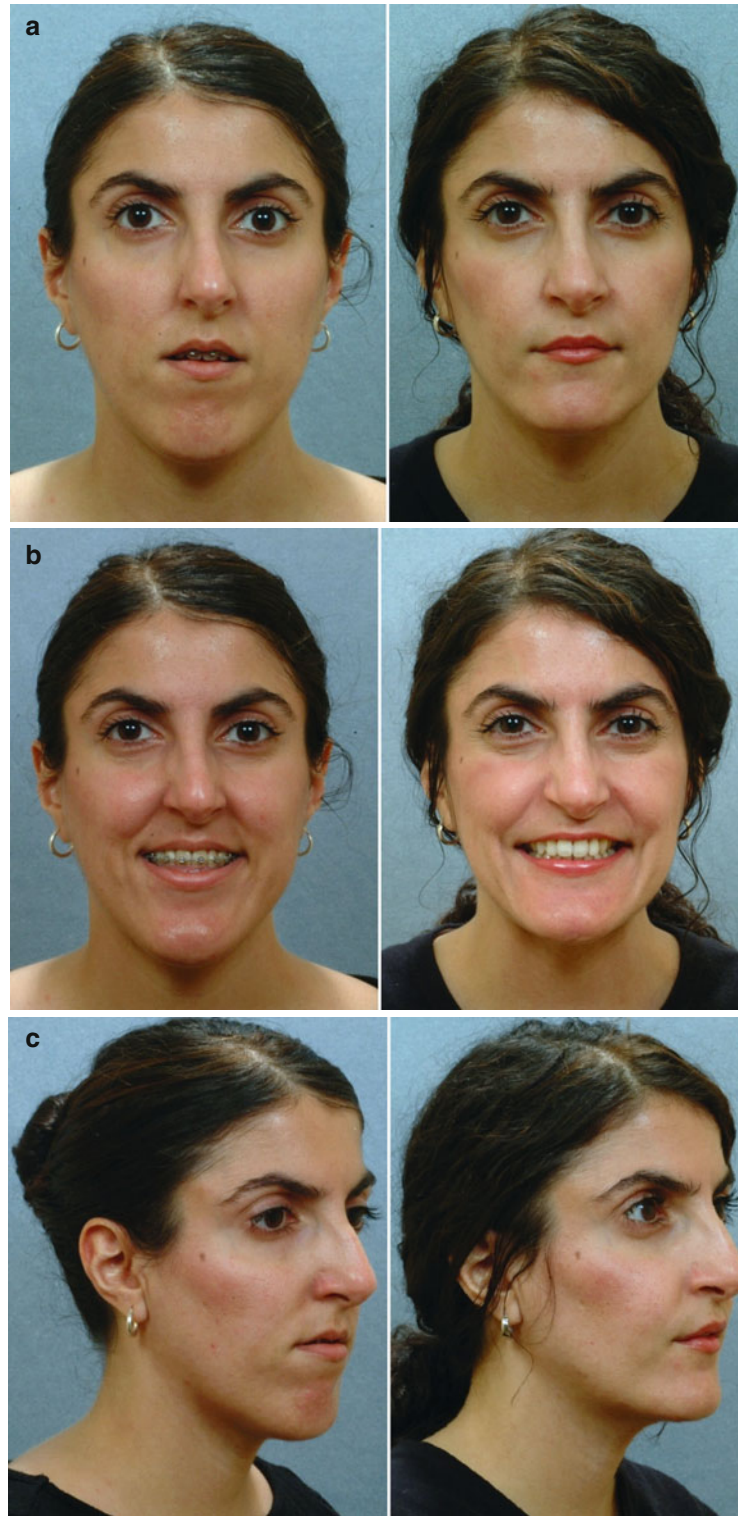


Fig. 25.5 A woman in her late 20s born with isolated cleft palate. She underwent palate repair in infancy. She developed a jaw deformity with malocclusion. Attempts to neutralize the occlusion included four bicuspid extractions and 6 years of growth modification and orthodontic mechanics (from age 11 to 17 years). She was left with generalized labial bone loss and gingival recession, especially of the lower anterior teeth. She presented to this surgeon as an adult with a lifelong history of obstructed nasal breathing and a long face growth pattern involving the maxilla, mandible, and chin region. There was excess lower anterior vertical height and horizontal retrusion (maxilla and mandible). She underwent evaluations including periodontics, prosthodontics, orthodontics, surgery, speech pathology, and ENT. Periodontal treatment and then orthodontic decompensation followed. Surgery includes LeFort I osteotomy (vertical shortening, clockwise rotation, horizontal advancement); bilateral sagittal split ramus osteotomies (horizontal advancement and counter clockwise rotation); osseous genioplasty (vertical shortening, horizontal advancement); and septoplasty/inferior turbinate reduction/recontouring of the nasal floor. (**a**) Frontal views in repose prior to and after reconstruction. (**b**) Frontal views with smile before and after reconstruction. (**c**) Oblique facial views before and after reconstruction

Fig. 25.5 (*continued*) (**d**) Profile views before and after reconstruction. (**e**) Occlusal views prior to re-treatment, after orthodontic decompensation and 1 year after reconstruction

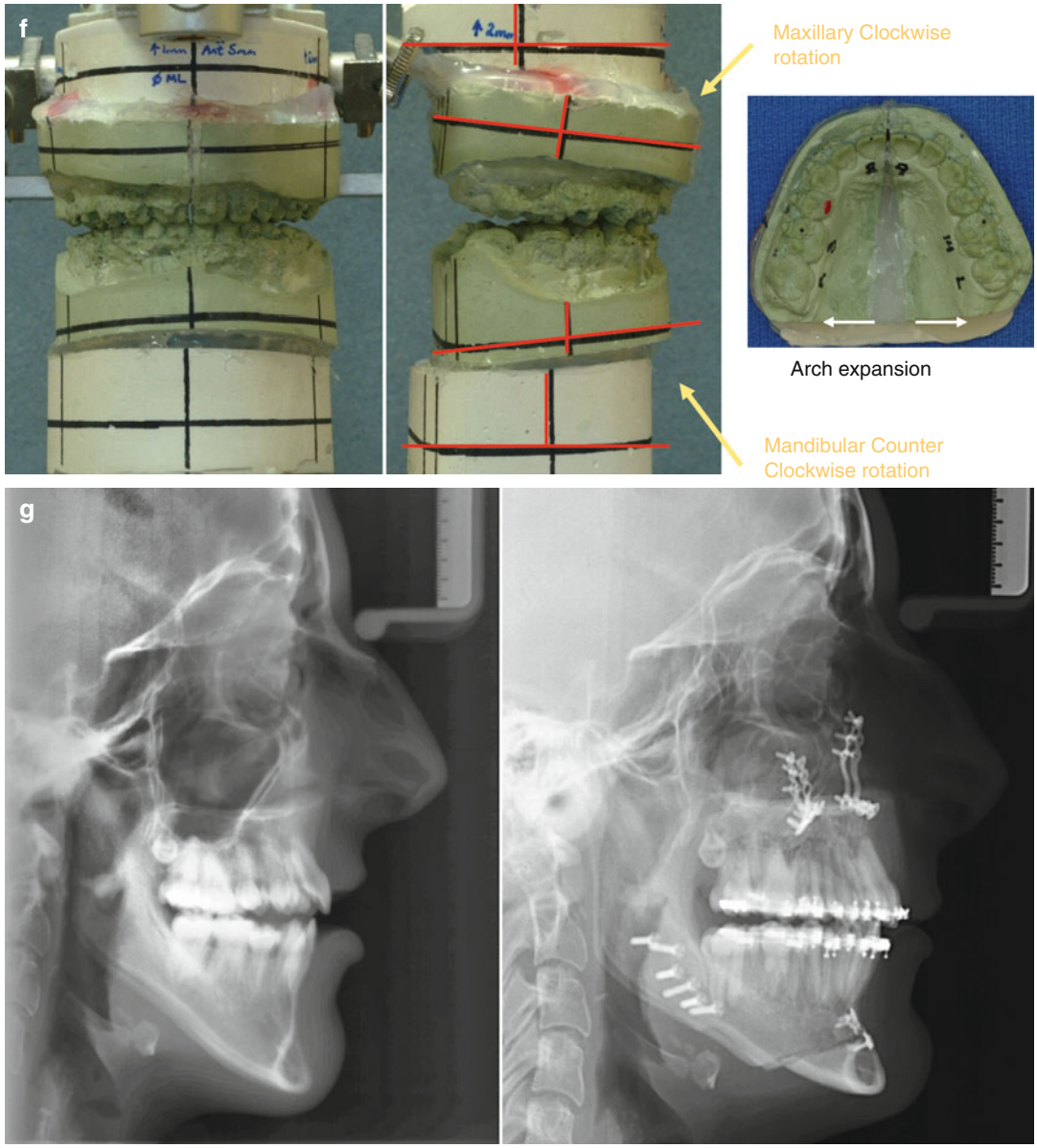


Fig. 25.5 (*continued*) (**f**) Articulated dental casts after model planning. (**g**) Lateral cephalometric views before and after reconstruction

25.2 Coordinated Team Approach

Care of the cleft patient (i.e., UCLP, BCLP, and ICP) is best delivered by an integrated group of specialists who evaluate and provide definitive care (Berkowitz 1978; Kapp-Simon 1995; Leonard et al. 1991). It is no longer acceptable for individual practitioners (e.g., surgeons, orthodontists, restorative dentists, speech pathologists, or otolaryngologists) to carry out extended treatment without considering all aspects of the patient's care and without discussing options with all members of the team (Cohen et al. 1995).

The need for coordinated and efficient cleft care was confirmed by the Eurocleft study which found a lack of association between "high-intensity disjointed treatment" and favorable results. The greater the number of operations and years of orthodontic appliances worn (heavy burden of care), the worst the clinical outcome for the patient (Hathaway et al. 2011; Shaw et al. 1992; Sinko et al. 2008; Tulloch 1993; Williams et al. 2001).

A frequent road block to successful reconstruction and dental rehabilitation of the midface-deficient adolescent born with a cleft is disagreement between clinicians about the indications, most effective surgical techniques, and timing for intervention. The surgeon, orthodontist, dental and medical team, patient, and family must first agree on the dental, occlusal, speech, upper airway, and aesthetic objectives. Only then can effective treatment go forward.

25.3 Treatment Protocol

Patients presenting with a cleft jaw deformity referred for possible orthognathic surgery are at a minimum seen by an orthodontist, orthognathic surgeon, otolaryngologist, and speech pathologist. Additional consultations are held with a prosthodontist, pediatrician/general dentist, periodontist, and other medical specialist (i.e., sleep specialist, geneticist), as indicated. For evaluation of the cleft dentofacial deformity, records and tests should include at a minimum medical-quality photographs with views of the face and occlusion, cephalometric and dental radiographs, dental models and a centric relation (CR) bite, direct facial measurements, nasoendoscopic speech and velopharyngeal assessment, and thorough evaluation of the upper airway.

The primary cleft surgeon not only performs repair of the cleft lip and palate and corrects velopharyngeal insufficiency when present but generally plays a role in directing the patient's overall care. If a cleft surgeon is not trained in skeletal procedures, then a timely and seamless transition to the maxillofacial surgeon should occur. The maxillofacial surgeon caring for the cleft patient with a skeletal deformity should have a fundamental understanding of the patient's dental, speech, upper airway, and aesthetic needs. He should request consultations with appropriate specialists, evaluate the clinical information, and be prepared to perform orthognathic and intranasal procedures.

The orthodontist provides appropriate interceptive treatment in the mixed dentition, identifies early abnormal skeletal growth patterns, and carries out definitive orthodontic treatment in conjunction with orthognathic surgery when indicated. From the mixed dentition phase, the cleft orthodontist should recognize the patient who may need orthognathic surgery. Instituting extensive camouflage (dental compensatory) treatment is likely to jeopardize periodontal health, lead to dental relapse, and may cause root resorption. Proceeding with a compromised (camouflage) orthodontic approach should only be entered into with full disclosure to the family and other treating clinicians.

Before orthognathic surgery, a speech pathologist experienced in cleft care performs an evaluation to characterize both velopharyngeal function and to identify articulation errors resulting from the cleft jaw deformity and dental malocclusion. A baseline evaluation is important because velopharyngeal function may deteriorate after maxillary advancement. A nasoendoscopic-guided speech assessment is preferred to provide maximum objective data without radiation expo-

sure. Velopharyngeal (VP) closure that was adequate before surgery may become borderline afterward, and VP closure that was borderline may become inadequate. Studies document that only a small percentage of patients require a primary pharyngeal flap or flap revision after maxillary advancement. Articulatory distortions resulting from malocclusion are also identified by the speech pathologist, and cause-and-effect relationships determined. The successful orthodontic and surgical correction of crossbites, open bites, cleft-dental gaps, negative overjet, and residual oronasal fistulas represent the most effective way to correct the identified articulation distortions.

A thorough evaluation of the upper airway in a cleft patient with a jaw deformity is conducted by an otolaryngologist to assess for areas of obstruction. A formal sleep study (attended polysomnogram) is done if there is suggestion of obstructive sleep apnea (OSA). If indicated, simultaneous intranasal procedures including: septoplasty, reduction of the inferior turbinates, and recontouring of the nasal apertures, floor of the nose, and anterior nasal spine should be carried out at the time of orthognathic surgery.

Discussion between the treating medical and dental consultants and patient/family clarifies the need for and extent of orthognathic and intranasal procedures. The overall plan for speech, jaw, upper airway, dental rehabilitation, and the enhancement of facial aesthetics is agreed to prior to initiating treatment.

25.4 Timing of Orthognathic Surgery

Correction of the cleft jaw deformity is best carried out after skeletal maturity and prior to the individual's finishing high school. Maxillofacial growth is generally complete between the ages of 14 and 16 years in girls and 16 and 18 years in boys. However, skeletal growth is variable and should be documented by knowledge of the pubertal growth spurt and/or sequential cephalometric radiographs taken at intervals. The patient/family's preferences for timing of the operation based on psychosocial and functional needs (i.e., speech, swallowing, chewing, and breathing) are also taken into account.

25.5 Residual Deformities in the Adolescent Born with a Cleft

25.5.1 Unilateral Cleft Lip and Palate

Correction of residual skeletal, soft tissue, and dental deformities in the UCLP adolescent challenges the ingenuity and skill of the orthognathic surgeon and cleft team (Laspos et al. 1997a, b; Posnick 1997, 2000a, b, c, d, e; Posnick and Ricalde 2004; Posnick and Agnihotri 2011; Sandham and Murray 1993; Stoelinga et al. 1990; Tessier and Tulasne 1984; Turvey et al. 1996). The central deformity is maxillary hypoplasia, but it is often combined with residual oronasal fistula, bony defects, intranasal obstruction, soft tissue scarring, and velopharyngeal dysfunction. In addition, the maxillary lateral incisor at the cleft site is usually congenitally absent or deficient, resulting in cleft-dental gap (Cassolato et al. 2009; Robertsson and Mohlin 2000; Suzuki and Takahama 1992a, b). Secondary deformities of the nose, mandible, and chin region are also common. The prevalence of these residual clefting deformities in mature patients with UCLP varies widely depending on the primary cleft surgeon's philosophy, available expertise (Nordquist and McNeill 1975), the individual's intrinsic biologic growth potential, and the family/patient's interest. Published clinical surveys of individuals born with complete UCLP and treated at established cleft centers indicate that despite best efforts, a number of children will not make themselves available for mixed dentition (prior to eruption of the cleft side canine) grafting (Felstead et al. 2010; McIntyre 2010). Also, some of those that do will fail grafting and require other means of reconstruction/dental rehabilitation. A subgroup of UCLP patients will present

at or after adolescence with multiple cleft-related problems that may include:
1. Maxillary hypoplasia
2. Residual oronasal fistula
3. Residual bony defects.
4. Cleft-dental gap
5. Chin dysplasia
6. Mandibular dysplasia
7. Nasal obstruction and sinus blockage.
8. Velopharyngeal dysfunction.

25.5.2 Bilateral Cleft Lip and Palate

The adolescent patient with BCLP often has several residual deformities that can be challenging to manage (Posnick 1997, 2000a, b, c, d, e; Posnick and Ricalde 2004; Posnick and Agnihotri 2011; Stoelinga et al. 1990). The central deformity is maxillary hypoplasia, but it is often combined with residual oronasal fistula, bone defects, intranasal obstruction, soft tissue scarring, and velopharyngeal dysfunction. In addition, the maxillary lateral incisors are usually congenitally absent or hypoplastic (93 % of the time), and the patients often have cleft-dental gaps. Secondary deformities of the nose, mandible, and chin region are also common (Cassolato et al. 2009; Robertsson and Mohlin 2000; Suzuki and Takahama 1992a, b). The prevalence of these residual deformities in mature patients varies widely depending on the treating clinician's philosophy, available expertise, the individual's intrinsic biologic growth potential, and the family/patient's interest/motivation. Published clinical studies indicate that a considerable number of children with BCLP will either not make themselves available for mixed dentition grafting or will fail the grafting procedure and require other means of reconstruction/dental rehabilitation (Felstead et al. 2010). For all of these reasons, a subgroup of BCLP individuals continues to present at or after adolescence with multiple cleft-related challenges that may include the following:
1. Maxillary hypoplasia
2. Residual oronasal fistula.
3. Cleft-dental gap
4. Residual bony defects
5. Chin dysplasia
6. Mandibular dysplasia
7. Nasal obstruction and sinus blockage
8. Velopharyngeal dysfunction

25.5.3 Isolated Cleft Palate

The ICP adolescent/adult referred for orthognathic evaluation will have an intact alveolar ridge and is generally with a full complement of teeth (Bell and Levy 1971; Posnick 1997, 2000a, b, c, d, e; Posnick and Ruiz 2000; Posnick and Ricalde 2004; Posnick and Agnihotri 2011; Stoelinga et al. 1987). They may, however, present with one or more of the following residual cleft-related problems:
1. Maxillary dysplasia
2. Residual oronasal (palatal) fistula
3. Chin dysplasia
4. Mandibular dysplasia
5. Nasal obstruction and sinus blockage
6. Velopharyngeal dysfunction

25.6 Orthodontic Considerations in a Cleft Patient with a Jaw Deformity

25.6.1 Unilateral Cleft Lip and Palate

The adolescent or adult UCLP patient that presents with maxillary hypoplasia and ineffective bone grafting will have two maxillary segments separated by a cleft. Each segment will have a varied degree of dysplasia in all three planes in space. From an orthodontic perspective, each segment is treated individually in anticipation of segmental repositioning. On the other hand, if the individual has undergone effective bone grafting in the mixed dentition, then the maxilla is in one unit and the arch form can be evaluated as an integrated unit.

There is great variability in the number of permanent incisors and the amount of alveolar bone in the anterior aspect of the UCLP maxilla. A lateral incisor-like tooth frequently is found along the edge of the cleft in the lateral segment.

When a poorly formed lateral incisor is present, it should be extracted in the interest of long-term function and dental rehabilitation. Cassolato et al. have documented that in patients with complete UCLP, the lateral incisor on the cleft side is normal and maintained in only 7 % of cases.

The decision to extract the fully erupted normally formed tooth (i.e., first bicuspid) depends on the volume and height of available alveolar bone to house the dental roots adjacent to the cleft and the degree of overall dental crowding within the arch. It is preferred to extract a first bicuspid to ensure that there is adequate alveolar bone for leveling and aligning the retained teeth without irreversibly weakening the periodontal support of the teeth adjacent to the cleft and throughout the alveolus. Planning for extractions in the mandibular arch depends on alveolar space requirements and on tooth movements needed to position the incisors ideally over basal bone. Mandibular extractions are generally not required.

The potential disadvantage of closure of a cleft-dental gap (whether carried out orthodontically or surgically) is that it shifts the preoperative anterior dental gap (lateral incisor region) to a posttreatment posterior gap (second molar region). Articulated dental models with the maxilla (maxillary segments) in the proposed postoperative positions are analyzed preoperatively to confirm that the posterior maxillomandibular occlusion will be acceptable. For this reason, the maxillary second molar on the cleft side be orthodontically included in the arch form. Occasionally, there will be a nonimpacted (useful) maxillary wisdom tooth that can be incorporated to oppose the mandibular second molar.

25.6.2 Bilateral Cleft Lip and Palate

The patient who has not had an effective bone graft in the mixed dentition will have three separate maxillary segments, each with a degree of dysplasia in all three planes. If this is the case, each segment should be treated individually by the orthodontist in anticipation of segmental surgical repositioning. Radiographic analysis is essential before any orthodontic movement is initiated. A Panorex film is useful for assessing overall jaw and dental anatomy and tooth angulation. A cone beam CT scan is preferred to assess bone volume at each cleft site.

Both the number of permanent incisors and the amount of dentoalveolar bone in the premaxilla differ widely. Lateral incisor-like teeth are frequently found along the edges of the premaxilla of the lateral segments. These generally are rudimentary with poor root form and are best removed. An erupted supernumerary teeth should be extracted either at the time of bone grafting in the mixed dentition or prior to orthognathic surgery. A useful lateral incisor at the cleft site is only found in 7 % of patients and should be assessed according to root, not crown, morphology.

The decision to extract additional teeth (i.e., bicuspids) should depend on the width and height of available alveolar bone and degree of dental root crowding in the segments (arch). Extractions may be preferred to ensure that there is adequate bone for leveling and aligning teeth without irreversibly weakening the periodontal support of the dental roots next to the clefts. Incorporation of all erupted maxillary teeth including the second molars into the orthodontic mechanics will facilitate development of the desired postoperative arch form and occlusion. Extractions in the mandibular arch are generally not required, but this depends on space requirements and on the tooth movements needed to position the incisors ideally over basal bone.

25.6.3 Isolated Cleft Palate

The primary goal of presurgical orthodontic treatment in the teenager with an ICP jaw deformity is to eliminate all existing dental compensations. Instituting camouflage treatment is likely to jeopardize periodontal health, lead to dental relapse, and may cause resorption. The need to retract the upper incisors, upright the lower incisors, and eliminate crowding, spacing, and rotations are all important orthodontic considerations. The arch form objectives are to achieve a satisfactory

occlusion at operation which can be detailed afterward. To eliminate dental compensations, extractions within the maxillary arch may be required. In the ICP patient, compared with the UCLP or BCLP patient, treatment is simplified as the alveolus is intact (no alveolar clefts), and there is generally a full complement of teeth.

25.7 Immediate Presurgical Reassessment

Two to six weeks before the planned operation, the orthodontist places passive surgical wires and confirms that the preoperative orthodontic objectives have been met. The surgeon takes final records including alginate impressions of the teeth, centric relation (CR) bite registration, facebow transfer, and direct facial measurements. Past medical and dental records (e.g., radiographs, reports, photographs, dental models, special studies) are reviewed. CT scan views of the cleft alveolus can be helpful to assess bone volume. Decisions are finalized concerning preferred vector changes (reposition) of the jaw(s) and the precise linear (mm) distances and angles to be accomplished in each jaw for the desired result. Model planning is carried out on the articulated dental casts, and splints are fabricated. The splints assist in achieving the precise occlusion and preferred facial aesthetics which have been decided on preoperatively.

25.8 Orthognathic Surgical Approach

25.8.1 Unilateral Cleft Lip and Palate

Historically, the literature warned of possible complications with maxillary osteotomy in patients with UCLP (Lanigan 1995). Only limited and confusing descriptions of techniques were provided to guide the orthognathic surgeon in the performance of safe, reliable osteotomies to solve these complex problems (Braun and Sotereanos 1980, 1981; Des Prez and Kiehn 1974; Fitzpatrick 1977; Freihofer 1977a, b; Georgiade 1974; Gillies and Rowe 1954; Gillies and Millard 1957; Jackson 1978; James and Brook 1985; Kiehn et al. 1968; Poole et al. 1986; Samman et al. 1994; Skoog 1965; Ward-Booth et al. 1984; Westbrook et al. 1983). As with other aspects of orthognathic surgery, Hugo Obwegeser's milestone contributions to cleft skeletal reconstruction are important (Drommer 1986; Obwegeser 1966, 1967, 1969a, b, 1971, 2007; Obwegeser et al. 1985). By the late 1960s, he succeeded in advancing the cleft maxilla to the preferred location without the need for a compromised mandibular setback approach. Early on, Obwegeser gained experience to feel comfortable with cleft maxillary advancement of up to 20 mm. He realized that adequate mobilization of the down-fractured maxilla was the key step in advancing the jaw, whether cleft or noncleft. The success of this approach, as initially carried out by Obwegeser, was confirmed by Bell's demonstration of the blood supply to these maxillary segments in animal studies (Bell and Levy 1971; Bell et al. 1995; Dodson et al. 1994; Dodson and Neuenschwander 1997). By the mid-1980s, Posnick utilized and refined Obwegeser's techniques of LeFort I osteotomy for treatment of the UCLP deformity (Braun 1992; Posnick 1991a, b, 1996; Posnick et al. 1994; Posnick and Tompson 1992). A key aspect was to use Obwegeser's circumvestibular incision which permitted direct exposure for dissection, osteotomies, disimpaction, fistula closure, septoplasty, inferior turbinate reduction, piriform aperture recontouring, bone grafting, and application of plate and screw fixation. This was consistently found to be a reliable approach without significant risk of circulation injury to the greater or lesser dento-osseous-musculomucosal segments (flaps).

The visibility provided by the circumvestibular incision made possible the incorporation of routine closure of the cleft-dental gap through differential maxillary segmental repositioning without necrosis of bone or loss of teeth. This method also closes the cleft dead space and brings together the labial and palatal flaps without need for subperiosteal undermining which permits closure of recalcitrant oronasal fistulas without tension and the establishment of periodontal health to the cleft-adjacent teeth (Fig. 25.6).

The extent of maxillary advancement carried out by the surgeon is based on the preferred occlusion, airway needs, and facial aesthetics determined preoperatively. The ideal vertical dimension is also achieved intraoperatively based on the preoperative plan. The maxillary osteotomy sites are fixed in place with titanium plates and screws at each zygomatic buttress and piriform aperture according to the principles originally described by Luhr (Champy 1980; Luhr 1968, 1981). An additional microplate is applied to stabilize each interpositional corticocancellous (iliac) graft. Mandibular and chin osteotomies to correct secondary deformities and facial asymmetries are also frequently required to achieve objectives (Fig. 25.1).

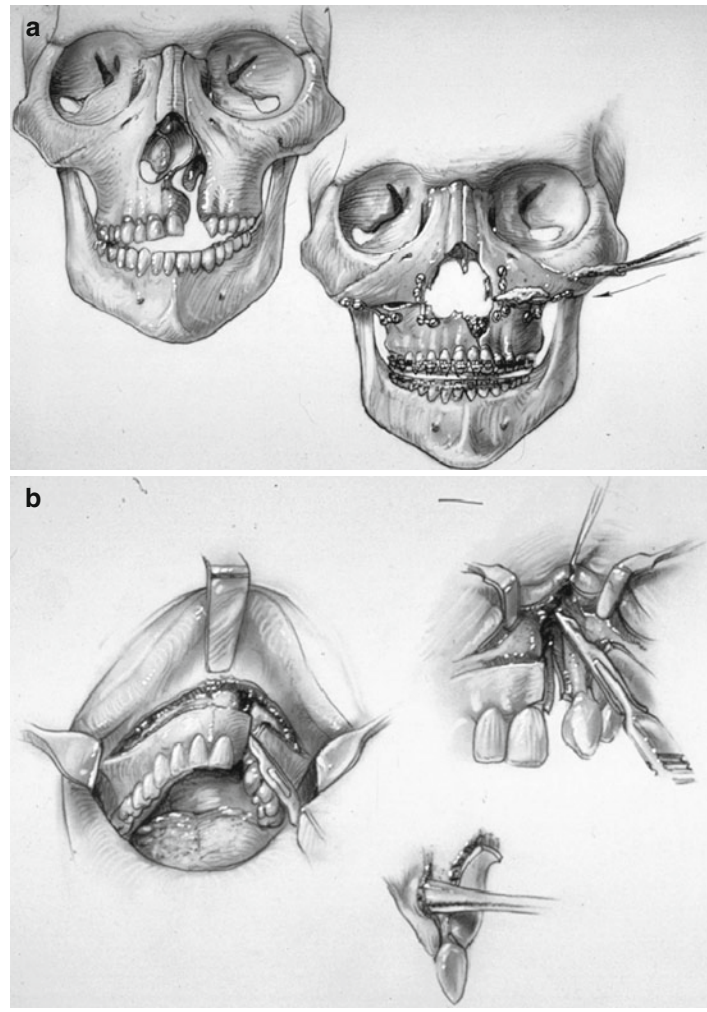


Fig. 25.6 Illustrations of modified LeFort I osteotomy in two segments as carried out in a patient with UCLP who has not undergone successful bone grafting in the mixed dentition. (**a**) Frontal view of maxillofacial skeleton before and just after LeFort I osteotomy in two segments. The inferior turbinates have been reduced, and a submucous resection of the deviated septum has been performed. The nasal floor has been recontoured with a rotary drill. Cancellous Iliac bone graft has also been placed along the nasal floor. Corticocancellous (iliac) graft is also placed in gaps along the anterior maxilla on each side. (**b**) Illustration of circumvestibular and perifistula incisions for exposure to complete osteotomies and later fistula closure

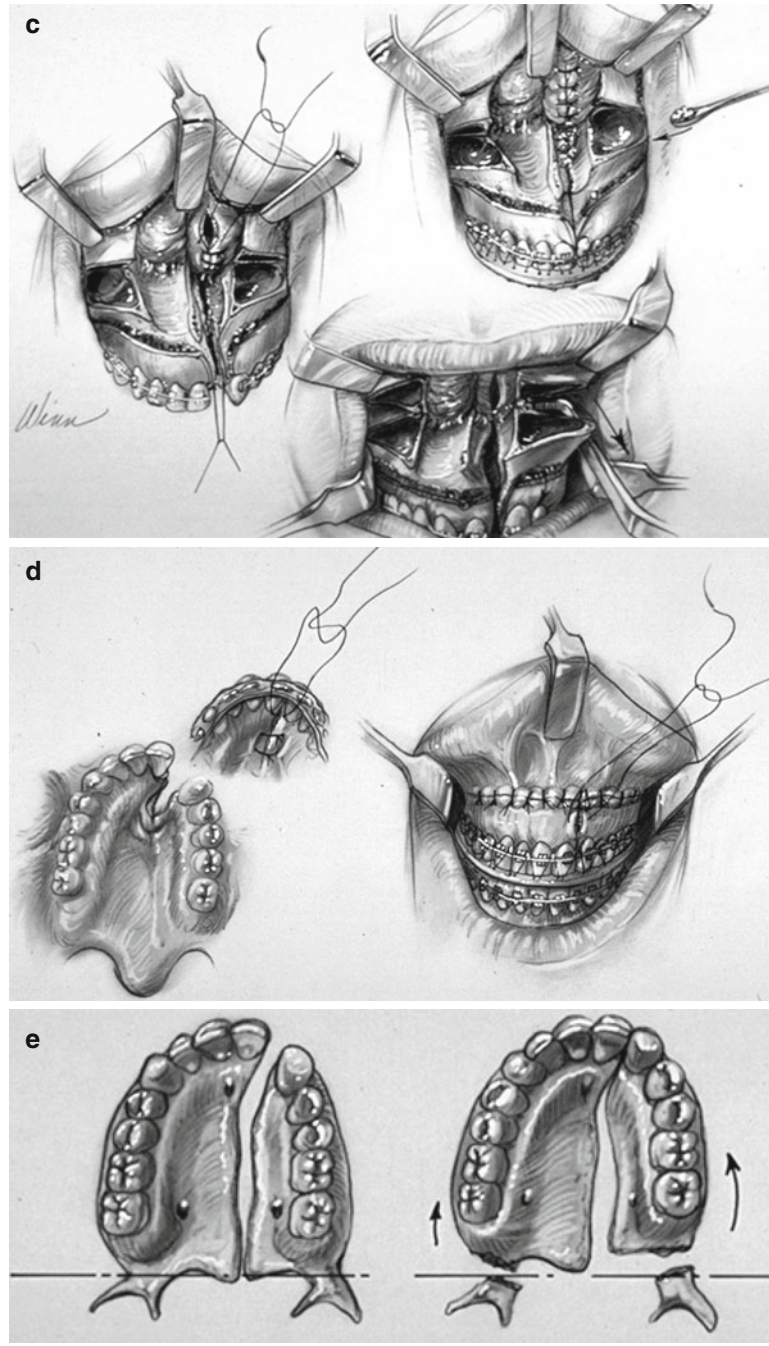


Fig. 25.6 (*continued*) (**c**) Illustration of down-fractured LeFort I osteotomy in two segments after submucous resection of septum, reduction of inferior turbinate through the nasal mucosa opening, and watertight nasal-side closure. (**d**) Illustration indicating wound closure after differential segmental repositioning. (**e**) Palatal view of bony segments before and after repositioning (From Posnick 1991b)

25.8.2 Bilateral Cleft Lip and Palate

Surgical attempts to correct the jaw disharmony seen in patients with BCLP date back to Steinkamm's descriptions in 1938 (Steinkamm 1938). The early literature mostly warned of possible complications with maxillary osteotomy in patients with BCLP and offered only incomplete descriptions of surgical techniques to guide the surgeon in performing safe and reliable osteotomies (Fitzpatrick 1977; Sinn 1980; Gillies and Rowe 1954; Gillies and Millard 1957; Jackson 1978; Poole et al. 1986; Ward-Booth et al. 1984). Hugo Obwegeser's early milestone contributions to cleft skeletal reconstruction are important (Drommer 1986; Obwegeser 1966, 1967, 1969a, b, 1971, 2007; Obwegeser et al. 1985). Unfortunately, in the early days, very few clinicians shared his enthusiasm. In 1974, Willmar reported on the complications associated with LeFort I osteotomy in BCLP patients (Willmar 1974). Of the eight patients he treated, one died of airway complications, and the results on the others were not reported favorably. By the mid-1980s, Posnick clarified the safety of the segmental LeFort I osteotomy technique in patients with a BCLP jaw deformity (Fig. 25.7) (Posnick 1996, 2000a, b, c, d, e; Posnick and Tompson 1993; Posnick et al. 1994). The documented favorable results confirmed the importance of

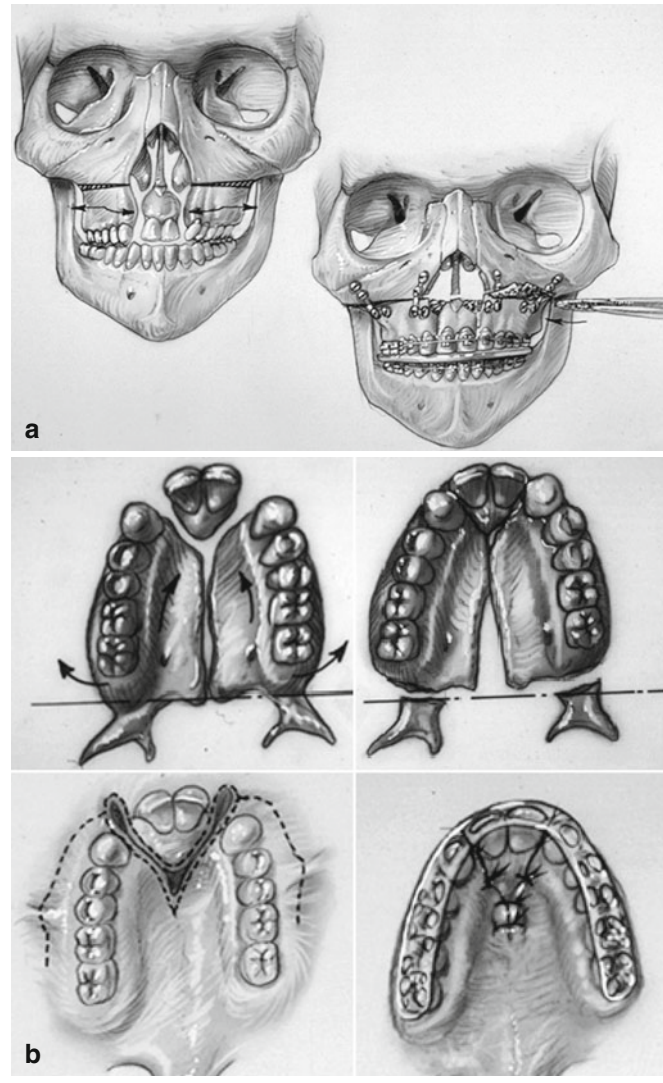


Fig. 25.7 Illustrations of modified LeFort I osteotomy in three segments. This is, as carried out in a patient with BCLP who has not undergone successful bone grafting in the mixed dentition. (**a**) Illustrations of a patient with BCLP before and after three-part maxillary osteotomies with repositioning of the segments. Septoplasty, inferior turbinate reduction, and recontouring of the nasal floor are also shown. (**b**) Palatal views of bone segments before and after repositioning for closure of cleft-dental gaps. Both skeletal and soft tissue views are shown

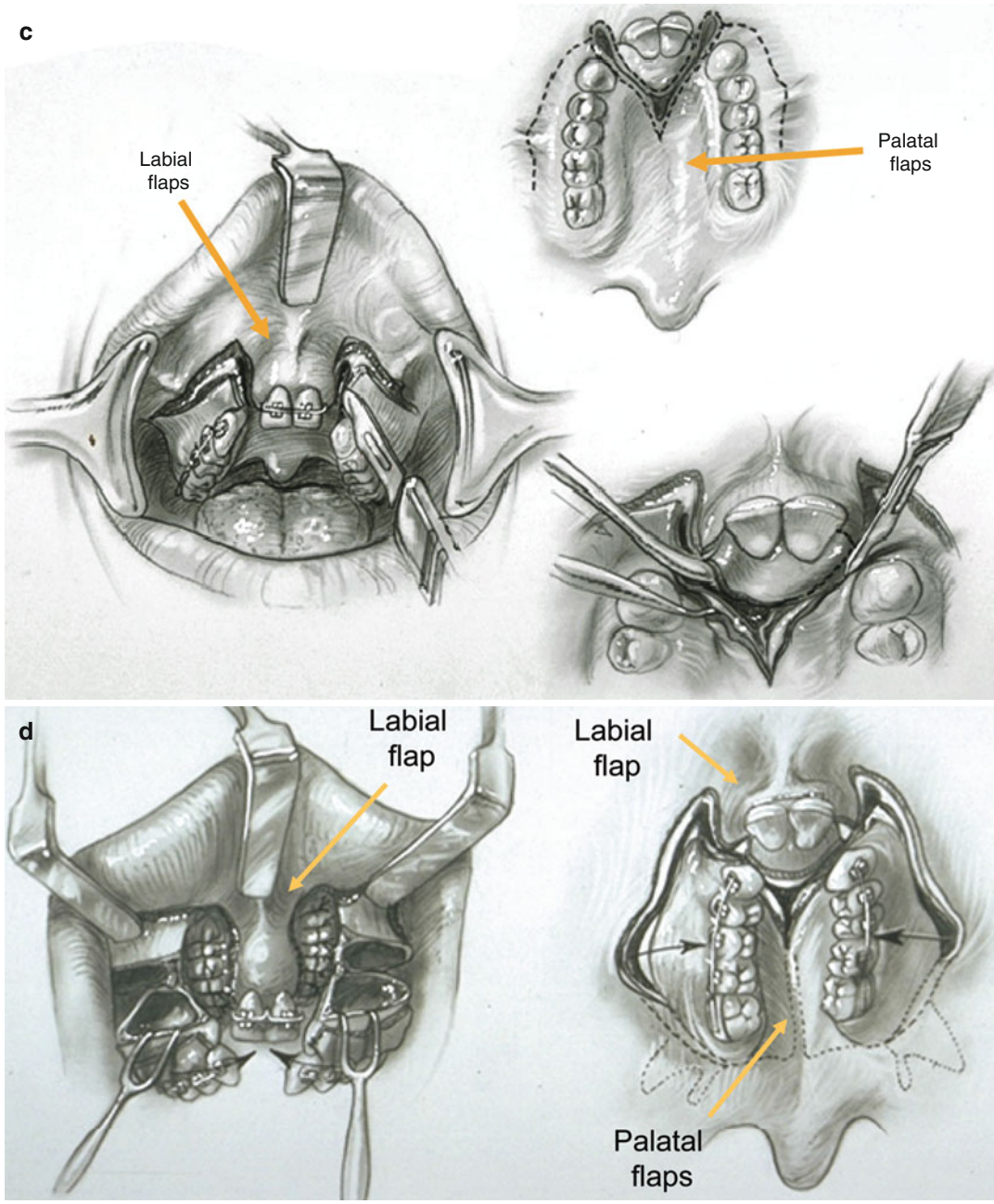


Fig. 25.7 (*continued*) (**c**) Illustrations of incisions for modified LeFort I in three segments. (**d**) Illustrations of down-fractured lateral segments demonstrating exposure for nasal-side closure of oronasal fistula

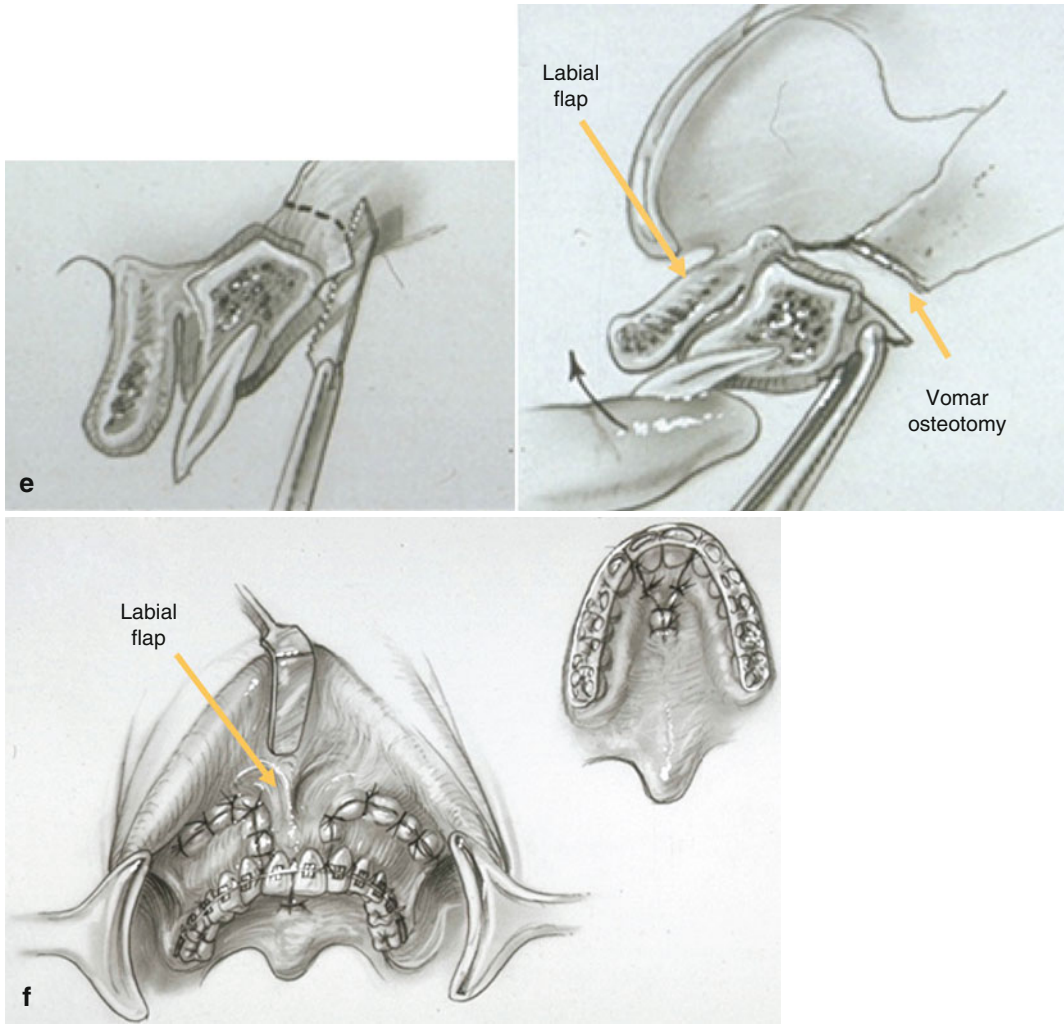


Fig. 25.7 (*continued*) (**e**) Illustration of premaxillary osteotomy carried out on palate side (vomer osteotomy) using a reciprocating saw. (**f**) Illustration demonstrating oral wounds sutured at end of procedure. (From Posnick 1991b)

following sound biologic principles. For example, preservation of the labial soft-tissue mucosal pedicle to the premaxilla in the unsuccessfully grafted BCLP maxilla is essential. The validity of this flap circulation was proven by Bell et al. in rhesus monkeys (Bell and Levy 1971; Bell et al. 1995).

The BCLP patient with an intact (successfully grafted) alveolar ridge on one side essentially presents to the surgeon with the same anatomy as a nongrafted UCLP patient. The surgical approach to the maxilla for this patient is the same as described for the nongrafted UCLP patient. For the BCLP patient presenting with a "three-segment" maxillary deformity, the modified LeFort I osteotomy (in three segments) should be used. For the BCLP adolescent presenting with an intact (successfully grafted) alveolar ridge on each side, a standard LeFort I down fracture is carried out (Fig. 25.4).

25.8.3 Isolated Cleft Palate

In general, the primary jaw deformity observed in the ICP adolescent is maxillary hypoplasia/dysplasia resulting from the original cleft deformity and the subsequent surgical interventions. The usual reconstructive procedure to consider is a standard LeFort I maxillary osteotomy. Early on, Obwegeser clarified that full mobilization of the down-fractured maxilla was essential to achieving an orthognathic correction on the operating room table and to limit skeletal relapse over time. Bell and others validated that the Obwegeser LeFort I technique allowed adequate blood supply for routine satisfactory bone healing without aseptic necrosis or dental injury (Fig. 25.5).

Any residual palatal oronasal fistula in the ICP patient at the time of orthognathic correction will be difficult to close simultaneously with the LeFort I procedure. This is because elevation of palatal flaps is generally required to do so and this would compromise the blood supply to the down-fractured maxilla. Interestingly, if a watertight nasal-side closure can be achieved through the down-fracture prior to fixing the maxilla in its new location, then the residual palatal-side mucosal separation will frequently heal by secondary intention with fistula closure.

25.9 Clinical Management After Initial Surgical Healing

Managing details of the in-hospital and at-home convalescence during initial healing are essential for a successful outcome. The surgeon is responsible for seeing the patient through this phase of treatment. Cephalometric and dental radiographs and facial and occlusal photographs are obtained at standard postoperative intervals for documentation.

Orthodontic maintenance of the surgical result and detailing of the occlusion are initiated after initial healing (5 weeks after surgery). The seamless transition from the surgeon to the orthodontist for ongoing care is essential.

Speech and VP function can be objectively reassessed 6 months after surgery, and any remaining issues can be addressed at that time. We prefer to evaluate VP function using nasoendoscopic instrumentation when feasible. Further cleft soft tissue procedures (e.g., cleft rhinoplasty, lip scar revision, pharyngeal flap/flap revision, closure of residual palatal fistula) can be carried out as early as 6 months after orthognathic surgery. Once the orthodontic appliances are removed, any planned definitive restorative dental work can also be finalized.

25.9.1 Unilateral Cleft Lip and Palate/Bilateral Cleft Lip and Palate

If segmental osteotomies were carried out to correct arch width, curve of Spee, and/or for closure of cleft-dental gaps, then the orthodontist sees the patient within 24 h of splint removal (approximately 5 weeks after surgery) and replaces the maxillary sectional arch wires with a rigid continuous one. The teeth are ligated together to maintain the surgical dental gap closure, new arch form, and transverse width. Active orthodontic maintenance and finishing are started at that time. The use of a transpalatal wire or a

palatal plate may also be used to stabilize (maintain) the arch form. Close orthodontist monitoring for skeletal and dental shifts during the first 6 months after surgery is essential.

25.10 Cleft Orthognathic Surgery: Results and Complications

25.10.1 Unilateral Cleft Lip and Palate

Posnick and Tompson prospectively assessed the cleft deformity and clinical results in 66 consecutive adolescents and young adults (age range 15–25 years; mean 18 years) with UCLP who underwent orthognathic surgery by one surgeon (Posnick) using a single surgical protocol over a 6-year time frame (Posnick and Tompson 1995). All patients underwent perioperative orthodontic treatment and were judged to be skeletally mature at the time of jaw surgery. The clinical follow-up after maxillary advancement ranged from 1 to 7 years (mean 40 months). These patients had their cleft lip/palate repaired earlier in life by different surgeons using a variety of protocols. Many had undergone multiple attempts at closure of residual oronasal fistula and to fill alveolar clefts with bone grafts. Seven of the 66 UCLP patients had previously undergone orthognathic surgery by another surgeon.

The basic orthognathic procedure carried out included a modified LeFort I osteotomy in two segments. All ($n=66$) had multiple residual UCLP skeletal deformities, including residual oronasal fistula (100 %), negative overjet (97 %), and congenitally missing lateral incisor (91 %). Twenty-three of these patients needed simultaneous sagittal split ramus osteotomies to correct facial asymmetry and disproportion. Thirty-five of the 66 underwent an osseous genioplasty (horizontal advancement with variable vertical reduction).

Sixty-one (92 %) of the 66 patients underwent successful simultaneous oronasal fistula closure. Surgical cleft-dental gap closure was achieved and maintained in all but 3 (5 %) of the 57 patients in whom it was attempted. In all of the patients, keratinized mucosa was placed and remained along the labial surface of the cleft-adjacent teeth ($n=132$ teeth). The long-term maintenance of overjet was measured directly from the late (more than 1 year) postoperative lateral cephalometric radiograph. All but two patients maintained a positive overjet. The long-term maintenance of overbite also was measured directly from the late postoperative lateral cephalometric radiograph. Sixty (91 %) of 66 patients maintained a positive overjet. Four (6 %) shifted to a neutral overbite, and two (3 %) relapsed into a negative overbite.

Complications were few and generally not serious. One patient who had undergone simultaneous septoplasty/inferior turbinate reduction was returned to the operating room for nasal packing to manage epistaxis (postoperative day 10). In another patient, the maxilla was repositioned a second time to reduce the vertical height (gingival show) for improved facial aesthetics (postoperative day 3). No loss of segmental bone or teeth occurred because of aseptic necrosis, infection, or for any other reason.

25.10.2 Bilateral Cleft Lip and Palate

Posnick and Tompson prospectively assessed the clinical results of 33 consecutive adolescents (age range 16–24 years; mean 18 years) with BCLP who underwent orthognathic surgery by a single surgeon (Posnick) over a 6-year period using the modified LeFort I osteotomy (three segments) (Posnick and Tompson 1995). All underwent perioperative orthodontic treatment and were judged to be skeletally mature at the time of jaw surgery. The clinical follow-up period ranged from 1 to 7 years (mean 40 months) at the close of the study. The patient's surgeons earlier in life varied as did the treatment protocols. Most had undergone additional attempts to close the residual oronasal fistulas and bone grafting to fill the alveolar clefts.

All of the patients in this study group required closure of residual bilateral labial and palatal

fistulas. The majority presented with a significant negative overjet at the incisors (26–33) as an indication of horizontal maxillary hypoplasia. Thirty of the 33 patients had a cleft-dental gap on each side. All 33 patients had a mobile premaxilla indicating no bone bridging across the cleft alveolar regions either side. Fifty-nine of the 66 cleft sites had congenitally absent/inadequate lateral incisors. In addition to a modified LeFort I osteotomy (three segments), ten of the patients underwent simultaneous sagittal split ramus osteotomies to correct secondary deformities (asymmetry/disproportion). Fourteen of the 33 also underwent osseous genioplasty (vertical reduction and horizontal advancement).

Twenty-six of the 33 patients underwent successful one-stage complete fistula closure and full stabilization of the premaxillary segment to the lateral segments as a result of the cleft orthognathic procedure. Seven of the 33 patients retained small fistulas (all at the incisor foramen region) and a degree of mobility of the premaxillary segment. Final fistula closure using local flaps with additional bone grafting was then required to attain complete fistula closure and bony union of the premaxilla to the lateral segments. All 33 patients successfully achieved the placement of keratinized gingiva over the cleft side and cleft-adjacent teeth on each side.

Five of the 33 patients benefited from fixed prosthesis to either replace missing teeth or improve the aesthetics of the incisors because of poor enamel quality or congenital dysplasia. The long-term maintenance of overjet and overbite was measured directly for each patient from the late (more than 1 year) postoperative lateral cephalometric radiograph. The majority of patients (31 of 33, 94 %) maintained a positive overjet. Only 2 of the 33 patients (6 %) relapsed into a negative overjet. Two of the 33 patients (6 %) shifted into neutral overbite, and 4 of the 33 (12 %) relapsed into a negative overbite.

There are no cases of infection requiring extended use of antibiotics or drainage, and there was no postoperative hemorrhage requiring return to the operating room. None of the cleft-adjacent teeth required root canal therapy. No teeth or dentoalveolar segments were lost as a result of aseptic necrosis, infection, or for any other reason.

25.10.3 Isolated Cleft Palate

In a previously published study (Posnick and Tompson), a consecutive series of ICP patients ($n = 14$) who presented with a cleft jaw deformity and underwent orthognathic correction by a single surgeon (Posnick) were prospectively assessed after surgery (Posnick and Tompson 1995). The 14 skeletally mature ICP cleft jaw deformity patients had undergone palate repair in infancy and presented to this surgeon in adolescence with maxillary dysplasia and malocclusion. All patients underwent a standard one-piece LeFort I osteotomy. Simultaneous bilateral sagittal split ramus osteotomies were also carried out in 4 of the 14 patients to further correct facial disproportion. All subjects ($n = 14$) underwent simultaneous osseous genioplasty. Ten of the 14 underwent interpositional iliac (hip) grafting to the horizontally advanced and vertically lengthened maxilla.

As measured directly from the greater than 1 year postoperative cephalometric radiographs, all patients maintained a positive overjet and overbite at the incisor level. The mean horizontal advancement initially achieved was 6.4 mm, and that maintained at 1 year was 5.4 mm. Perioperative morbidity was unremarkable with reference to cardiopulmonary compromise, maxillofacial infections, hemorrhage, aseptic necrosis, loss of teeth, and/or need for root canal therapy.

25.11 Controversies and Unresolved Issues

25.11.1 Velopharyngeal Function After LeFort I Advancement

Uncertainties about velopharyngeal function and management of an in-place pharyngeal flap should no longer be limiting factors when orthognathic surgery is necessary in a patient with a cleft.

A nasoendoscopic-guided examination by a speech pathologist and surgeon familiar with cleft anatomy can reasonably predict current and expected velopharyngeal function in a patient scheduled for a LeFort I osteotomy. When postoperative velopharyngeal deterioration is anticipated, the patient and family are counseled about the sequencing of treatment and alternatives discussed. *Clinical studies have now documented that VP function will deteriorate in a similar fashion whether either DO or standard LeFort I osteotomy techniques are utilized* (Chanchareonsook et al. 2006, 2007; Guyette et al. 2001; Harada et al. 2001; Janulewicz et al. 2004; Ko et al. 1999; Marrinan et al. 1998; McComb et al. 2011; Phillips et al. 2005; Trindale et al. 2003; Witzel and Munro 1977). Despite the frequent need for significant maxillary advancement to normalize the skeleton, upper airway, and facial aesthetics in patients with a cleft, we have not had or seen an advantage to transecting an in-place pharyngeal flap to achieve maxillary mobilization and the desired advancement. Our research and that of others confirm that a pharyngeal flap in place at the time of LeFort I does not increase complications nor does it result in a higher incidence of relapse. Definitive reassessment of VP function after cleft LeFort I advancement can be carried out as early as 3 months after surgery. A primary or revision pharyngeal flap can be safely carried out by 6 months in conjunction with cleft rhinoplasty and/or labial revision if indicated.

25.11.2 Mixed Dentition LeFort I Osteotomy

By the mid-1980s, research clarified that if jaw surgery is undertaken in a growing cleft palate patient, another procedure will likely be required once skeletal maturity is reached. More recently, several investigators have tested this theory by preceding with mixed dentition LeFort I osteotomies utilizing distraction (DO) techniques. *All research to date indicates that LeFort I advancement carried out in the mixed dentition in the cleft individual, whether by standard or distraction (DO) technique, results in no significant further horizontal growth* (Harada et al. 2006; Huang et al. 2007; Molina et al. 1998; Polley and Figueroa 1997, 1998; Suzuki et al. 2004; Wolford 1992; Wolford et al. 2001a, b, 2008). As the mandible continues to grow, an Angle Class III malocclusion will occur with the need for either additional LeFort I advancement or mandibular setback.

25.11.3 Skeletal Relapse After LeFort I Osteotomy

There are over 100 published articles reviewing skeletal stability and relapse in cleft patients who have had LeFort I advancement using either standard osteotomy or distraction osteogenesis (DO). *Clinical research data does not demonstrate significant differences in relapse patterns between the two techniques* (*standard* vs. *distraction* (*DO*) *for LeFort I advancement*) (Abyholm et al. 1981; Al-Waheidi et al. 1998; Araujo et al. 1978; Aksu et al. 2010; Chen et al. 2011; Cheung and Chua 2006; Cho and Kyung 2006; Cohen et al. 1997; Dongmei 2010; Erbe et al. 1996; Escenazi and Schendel 1992; Ewing and Ross 1993; Figueroa et al. 1999, 2004; Freihofer 1976, 1977a, b; Garrison et al. 1987; Hathaway et al. 2011; Hedemark and Freihofer 1978; Heidbuchel et al. 1994; Hirano and Suzuki 2001; Hochban et al. 1993; Houston et al. 1989; Hui et al. 1994; Kanno et al. 2008; Mansour et al. 1983; McCance et al. 1997; Posnick and Ewing 1990; Posnick and Dagys 1994; Posnick and Taylor 1994; Posnick and Tompson 1995; Stoelinga et al. 1987; Wiltfang et al. 2002). Proponents of DO frequently subjectively state that when greater than 10 mm of horizontal advancement is required, the use of standard osteotomies with plate and screw fixation and bone graft may lead to a greater degree of relapse. In perspective, it should be noted that only 5 % of cleft patients undergoing LeFort I advancement require greater than 10 mm of horizontal advancement at the incisors. All clinicians agree that this subgroup of cleft patients (5 %) is the most challenging. The reasons for this go beyond the degree of horizontal maxillary deficiency. These individuals are likely

to also present with multiple residual end-stage deformities and previously failed surgical procedures.

Aksu et al. documented a horizontal maxillary (skeletal) relapse of 22 % in cleft lip and palate patients after LeFort I using DO (Aksu et al. 2010). This was at the 3-year follow-up in a series of adult repaired cleft lip and palate patients who presented with maxillary hypoplasia. He et al. reported their results on adolescents with repaired cleft lip and palate and maxillary hypoplasia who then underwent LeFort I maxillary advancement using an external distraction (DO) device (Aksu et al. 2010). The study patients ($n=17$) were treated at one center between 2000 and 2006 and had greater than 1 year of follow-up. Interestingly, the first four patients (two UCLP and two BCLP) treated developed fibrous nonunion. All four required reoperation and rigid (titanium plate and screw) fixation to achieve union. The authors then extended the consolidation period to a minimum of 3 months for the remaining patients ($n=13$). They were able to achieve bony union (13/17 = 76 %) in this group. The mean horizontal relapse in this group (13/17) was 11.9 % with 5 of 13 (38 %) patients developing not better than an "end-to-end" occlusion. A second orthognathic procedure was necessary in this subgroup (5 of the 13, 38 %). Therefore, 9/17 (53 %) of the study patients required two orthognathic procedures. Chen et al. in 2011 reported a 30.7 % incidence of horizontal (skeletal) relapse 1 year after LeFort I osteotomy using DO techniques (RED device). This was in a consecutive series of cleft patients that presented with maxillary hypoplasia (Chen et al. 2011).

Posnick et al. documented the degree of horizontal relapse when using a standard LeFort I osteotomy (in segments) in cleft jaw deformity patients (as described in this chapter) (Posnick and Tompson 1995). They measured horizontal change and stability from the greater than 1-year postoperative cephalometric radiographs and clinical examination. The results were reported according to cleft type: UCLP (6.9 mm mean advancement with 5.3 mm maintained) (Posnick et al. 1994), BCLP (94 % maintained a positive overjet long-term) (Posnick and Tompson 1995), and ICP (6.1 mm mean advancement with 5.1 mm maintained) (Posnick et al. 1994). Interestingly, the degree of relapse documented by Posnick et al. using standard techniques was less than generally reported by clinicians using a DO approach (see above). As Obwegeser stated in the 1960s and was reconfirmed by Precious et al., "relapse in cleft patients after LeFort I advancement may well be more related to failure to adequately mobilize the maxilla and free it of abnormal soft tissue attachments than anything inherent in the osteotomy or the specific diagnosis" (Obwegeser 2007).

25.11.4 Staging of Maxillary Reconstruction

25.11.4.1 Unilateral Cleft Lip and Palate

The described modified LeFort I (2-segmental) osteotomy as a method of managing end-stage oronasal fistulas, alveolar defects, and cleft-dental gaps in patients with UCLP who also have maxillary hypoplasia is not intended to replace in most cases standard techniques and accepted sequencing of treatment (Fig. 25.5) (Harrison 1992; Lund and Wade 1993; Parel et al. 1986; Perrott et al. 1994; Takahashi et al. 1997a, b; Turvey 1991; Verdi et al. 1991; Zachrisson and Stenvik 2004). We always prefer secondary bone grafting in the mixed dentition with orthodontic closure of the cleft-dental gap. However, the method described does offer an alternative when the opportunity for grafting in the mixed dentition before the eruption of the permanent canine tooth is lost and a jaw deformity also exists. A two-stage approach to the adolescent or adult with maxillary hypoplasia, residual alveolar clefts, oronasal fistulas, cleft-dental gap, and nasal obstruction is not cost or time effective, and in our experience, the potential overall morbidity is increased. It is in these patients with UCLP that a modified LeFort I osteotomy offers a reasonable opportunity for the resolution of residual end-stage problems (i.e., maxillary hypoplasia, alveolar defect, residual fistulas, cleft-dental gap, nasal obstruction) in a safe and effective way. The long-term benefits to the patient of the

resulting "low-maintenance" (nonprosthetic) dentition cannot be overstated.

25.11.4.2 Bilateral Cleft Lip and Palate

The described modified LeFort I (three-segmental) osteotomy as a method for managing the maxillary deformity in adolescent/adult BCLP patients that also present with missing teeth, oronasal fistulas, alveolar defects, cleft-dental gaps, and nasal obstruction is not intended to replace standard techniques and accepted sequencing of treatment (Fig. 25.7) (Harrison 1992; Lund and Wade 1993; Parel et al. 1986; Perrott et al. 1994; Takahashi et al. 1997a, b; Turvey 1991; Verdi et al. 1991; Zachrisson and Stenvik 2004). This does, however, offer an alternative once the opportunity for grafting the alveolus in the mixed dentition before the eruption of the permanent canine(s) is lost and a jaw deformity coexists. A two-stage approach for the adolescent with BCLP and maxillary hypoplasia, residual alveolar clefts, oronasal fistulas, cleft-dental gaps, mobile premaxilla, and nasal obstruction is not cost or time efficient, and there may be increased morbidity. It is in these patients with BCLP that a modified LeFort I osteotomy in three segments with differential repositioning of each segment offers a reasonable opportunity for the resolution of residual problems.

25.11.5 Standard Versus DO Approach to LeFort I Advancement

I agree with Obwegeser when he stated, "Today, many surgeons will resort to the use of distraction devices to gradually advance the cleft maxilla. Although in some circumstances, this may be appropriate, it should be remembered that most cleft patients can be treated efficiently, even when requiring significant advancement, with a standard LeFort I type procedure as described."

Usual thinking is for a surgeon to commit to an approach (DO versus standard technique) for a LeFort I advancement in a cleft patient prior to arriving in the operating room. The surgeon then "sticks to the plan" no matter what happens intraoperatively. Based on review of the literature and a 26-year personal experience as a cleft jaw surgeon, I make the following observations and recommendations.

25.11.6 Observations Concerning DO Approach

- Distraction osteogenesis is always a patient's second choice compared to the standard approach of LeFort I osteotomy with stable (plate and screw) internal fixation. The length of convalescence after surgery using DO is longer. It will be at least 3 months of limited diet and physical activities. This generally is followed by several additional months of face mask therapy. The DO device is also awkward, socially embarrassing, and typically blocks the patient's visual fields (e.g., red, blue, or green external DO devices). Furthermore, there is no overall reduction in the potential for perioperative complications with DO compared to a standard approach.
- If the cleft maxilla is adequately down-fractured, mobilized, and stabilized with interpositional (bone) grafts and plate and screw fixation, there is a high probability of healing as planned with a predictable and less extensive convalescence than DO.
- An experienced cleft jaw surgeon is likely to be more confident with his ability to down-fracture and fully mobilize the cleft maxilla than a less experienced surgeon. Therefore, the less experienced surgeon will more likely use DO to avoid the uncertainty (personal stress) of the intraoperative maxillary mobilization process.
- Even for the experienced cleft orthognathic surgeon, there will be the occasional patient where the extent of maxillary hypoplasia and associated deformities (missing teeth, lack of alveolar bone) will lead them to choose a DO technique. Despite its protracted healing requirements and limited versatility to correct all the presenting cleft deformities, the DO device's gradual stretching ability can mobilize even the most recalcitrant maxilla (Fig. 25.8).

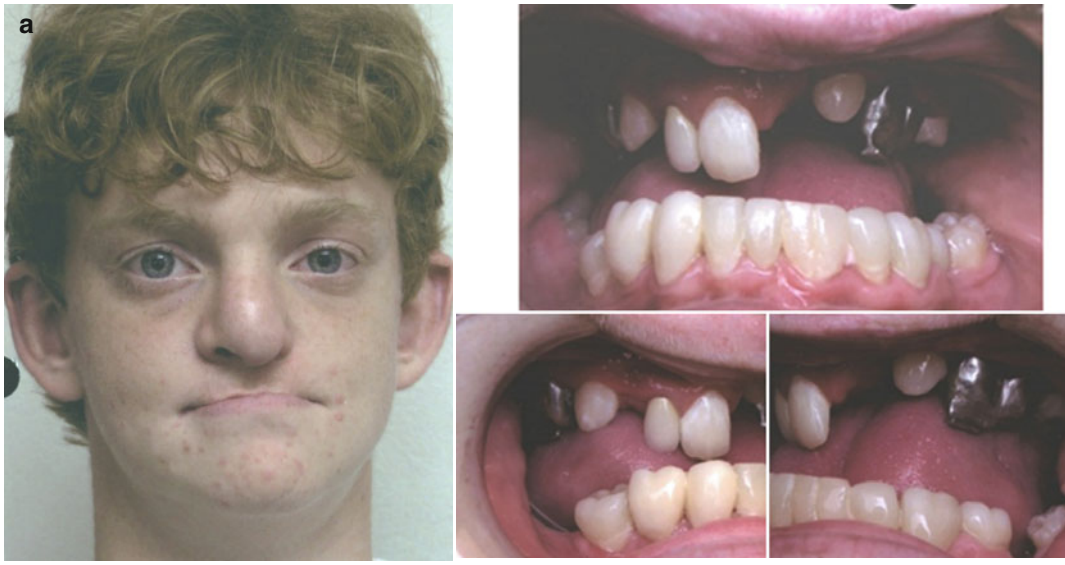


Fig. 25.8 A teenage boy born with ectodermal dysplasia and complete UCLP. He underwent lip and palate closure in infancy. There are multiple congenital missing teeth in each arch. He was referred to this surgeon for evaluation at 14 years of age. He retains only four long-term useful teeth in the maxilla (first molars) and a badly displaced right central and lateral incisor but with good roots and periodontal attachment. The maxilla is vertically and horizontally deficient. The mandible is with satisfactory symmetry and horizontal projection. He underwent evaluations by specialists including orthodontist, prosthodontist, periodontist, surgeon, speech pathologist, otolaryngologist, and geneticist. Reconstruction/dental rehabilitation was felt to require surgical repositioning of the maxilla followed by an overdenture. In the mandible, crown and bridge rehabilitation would be carried out. The prosthodontist requested 18-mm horizontal advancement and 14 mm of vertical lengthening of the maxilla. A two-stage approach to maxillary reconstruction was undertaken. *Stage I surgery*: (1) nasotracheal intubation, (2) LeFort I osteotomy with disimpaction, (3) septoplasty/inferior turbinate reduction, (4) application of MED I external distraction device, and (5) securing of MED I to prefabricated chrome-cobalt maxillary appliance fixed to dentition. Successful outpatient distraction of maxilla to the preferred position was accomplished over a 10-day time frame. *Stage II surgery*: return to the operating room for (1) awake fiberoptic nasotracheal intubation, (2) removal of MED I device, (3) harvesting of anterior iliac corticocancellous graft, (4) reopening of circumvestibular incision, (5) securing a prefabricated splint to the maxilla and then applying IMF, (6) autorotation of maxillomandibular complex to achieve desired vertical dimension, (7) application of plate and screw fixation to maxilla, (8) crafting and inset of corticocancellous grafts to the left and right anterior maxilla, and (9) plate and screw fixation of each graft to native maxilla. After 6 weeks, the maxilla achieved initial bone healing. He returned to a more regular diet and sports activities. Six months postoperative, he underwent a pharyngeal flap to achieve velopharyngeal competence and an open rhinoplasty including a rib cartilage (caudal strut) graft. Fixed bridge work in the mandibular arch and overdenture construction for maxilla were accomplished. (**a**) Frontal and occlusal views at 14 years of age

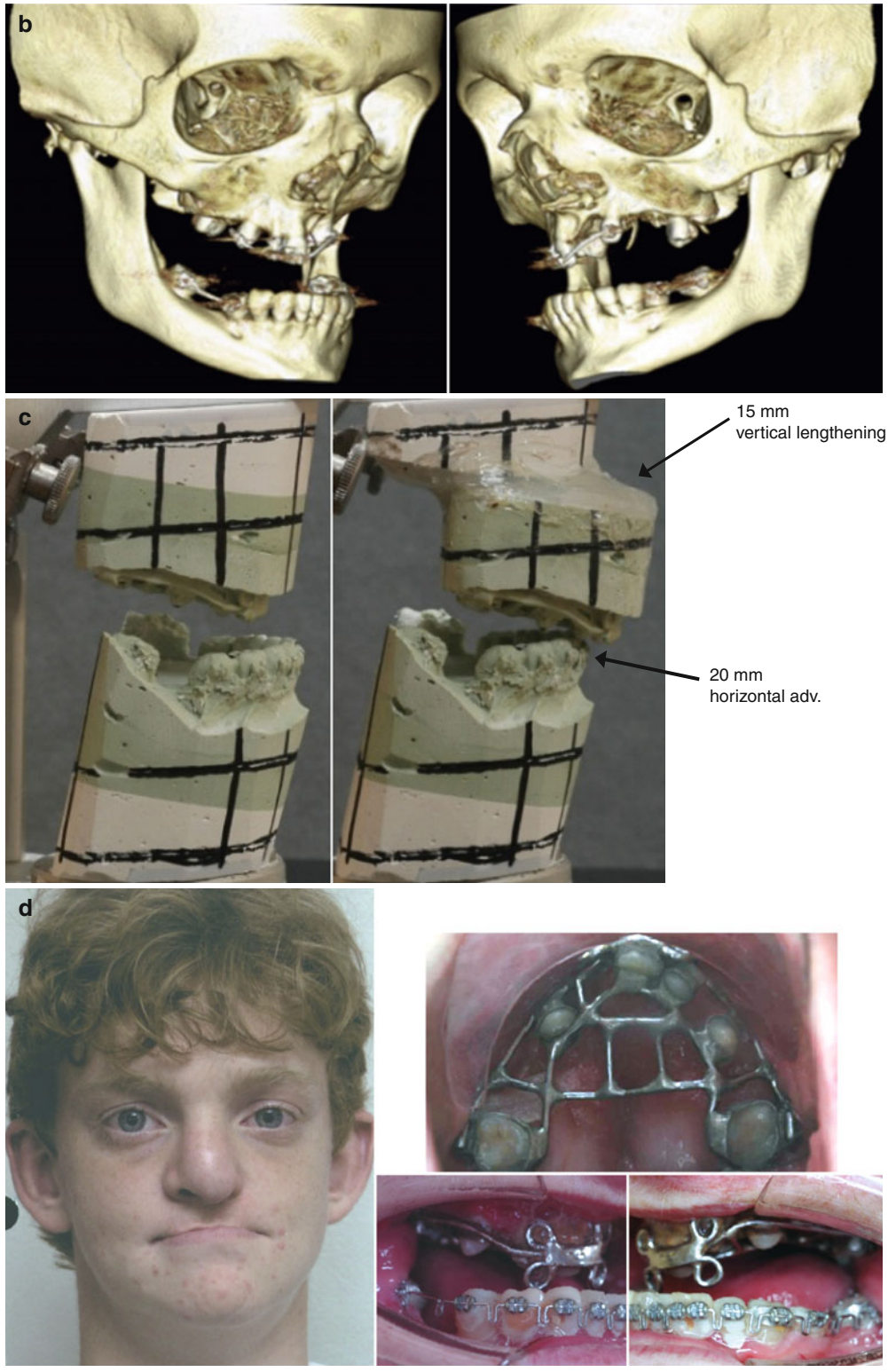


Fig. 25.8 (*continued*) (**b**) CT scan views indicating extent of maxillary hypoplasia. (**c**) Articulated dental casts before and after model planning. (**d**) Facial and occlusal views prior to surgery with prefabricated chrome-cobalt appliance fixed to maxillary dentition

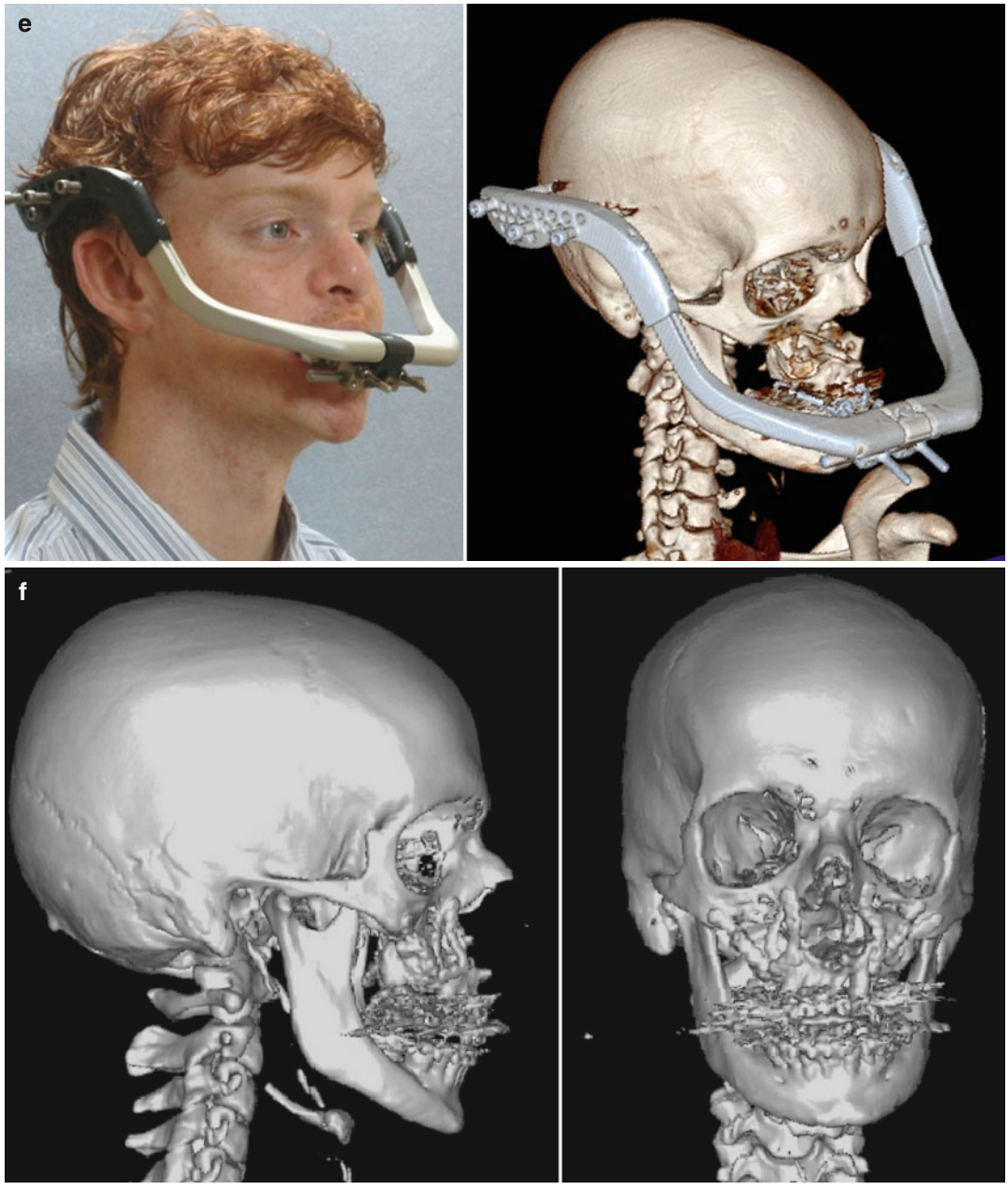


Fig. 25.8 (*continued*) (**e**) 10 days after LeFort I osteotomy with distraction of maxilla into preferred position accomplished. (**f**) CT scan views after securing maxilla in new location with bone graft and plate and screw fixation

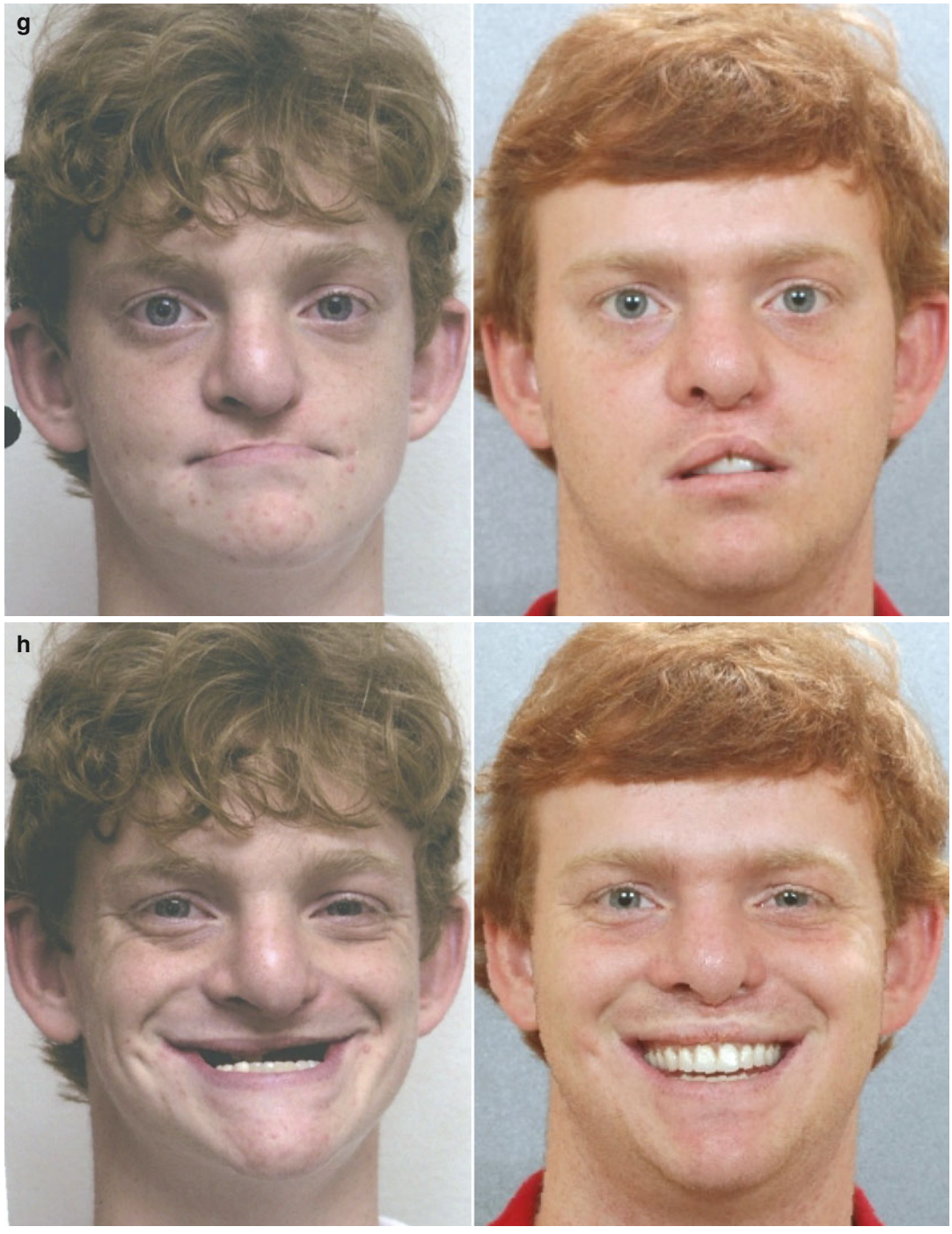


Fig. 25.8 (*continued*) (**g**) Frontal facial views in repose before and after reconstruction/rehabilitation. (**h**) Frontal views with smile before and after reconstruction/rehabilitation

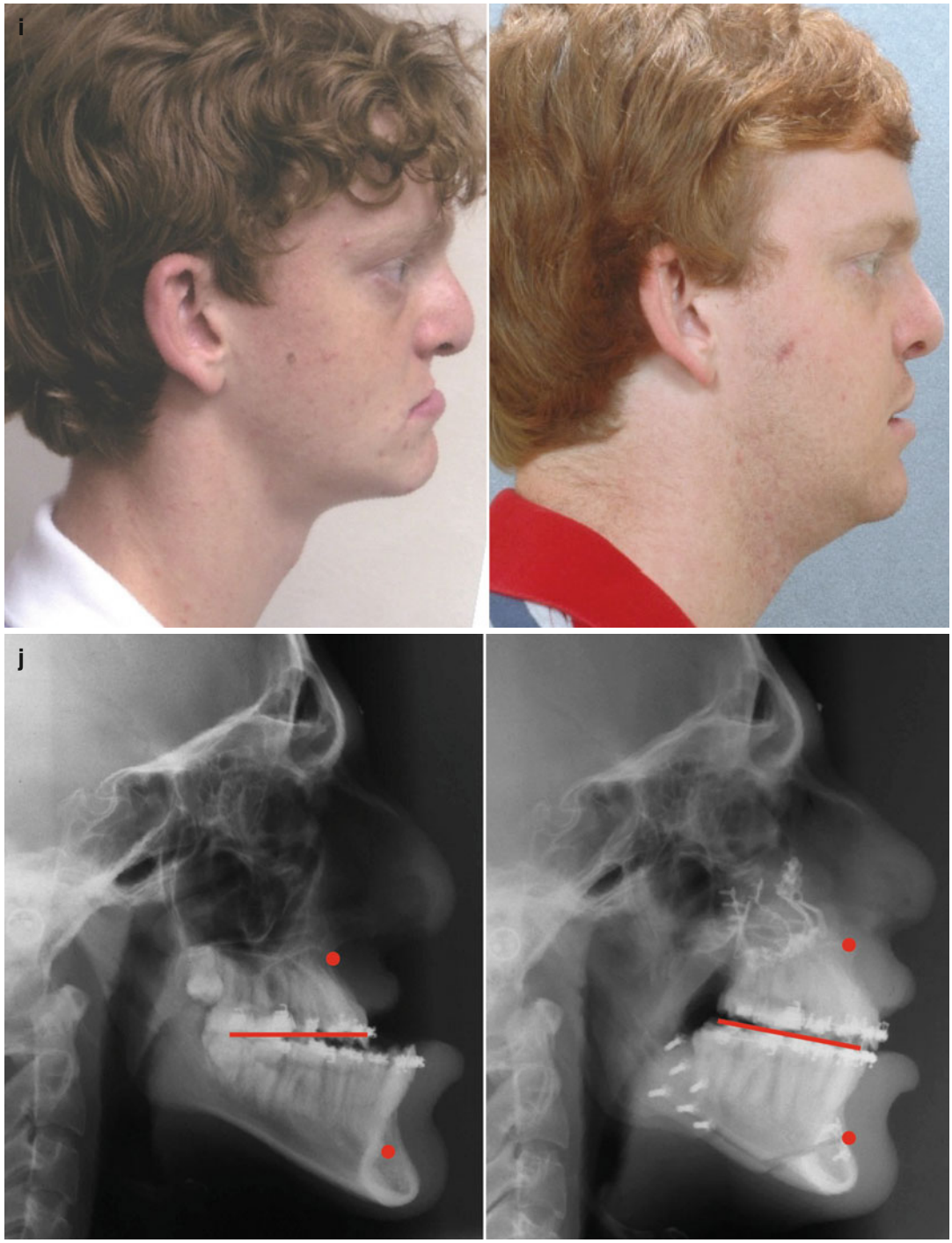


Fig. 25.8 (*continued*) (**i**) Profile views before and after reconstruction/rehabilitation. (**j**) Lateral cephalometric radiographs before and after surgery

25.11.7 Recommendations Concerning DO Approach

For most cleft patients with maxillary hypoplasia, standard surgical techniques, as described in this chapter, are efficient and effective. For those rare circumstances, reserving DO as a "bailout" when the down-fractured maxilla cannot be adequately mobilized is the approach I recommend. The occasional patient/family at risk can be informed of this contingency plan in advance. This approach allows the surgeon flexibility to make the right decision, at the right time, for the patient in need, to achieve the most efficient convalescence and favorable long-term results.

Conclusions

There are now convincing clinical studies which document the high prevalence of jaw deformities in mature patients with repaired cleft palate (i.e., UCLP, BCLP, and ICP). The methods described to manage the adolescents/adults presenting cleft jaw deformities, malocclusion, and any residual oronasal fistula, bony defects, cleft-dental gaps, nasal obstruction, and aesthetic needs are generally safe and reliable when performed by a dedicated orthognathic surgeon and team. Successful cleft orthognathic and intranasal procedures provide a stable foundation on which final soft tissue, lip, and nose reconstruction may be carried out.

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Secondary Bone Grafting of Alveolar Clefts

26

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Cleft patients with a permanent osseous defect of the alveolar arch and maxilla will, even after the best surgical and orthodontic treatment, be left with the following deficiencies:

1. Limited prospects for orthodontic treatment. The osseous defect makes a nonprosthodontic dental rehabilitation impossible and necessitates a dental bridge to close the gap in the dental arch.
2. Instability of the maxillary segments, particularly of the premaxilla in bilateral clefts.
3. Oronasal fistulae or mucosal recesses that impede oral hygiene.
4. Insufficient support of the alar base contributing to the nasal asymmetry.

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Secondary bone grafting of alveolar clefts combined with subsequent orthodontic treatment, designed to obtain a nonprosthodontic dental rehabilitation and also eliminate the other deficiencies attributable to the osseous defect, was started in Oslo in 1977. Our procedure is based mainly on the principles laid down by Boyne and Sands (Boyne and Sands 1972, 1976). Our clinical experience with more than 1,302 cleft sites has confirmed the functional responsiveness of the grafted tissue to tooth migration and orthodontic movement of teeth.

26.1 Surgical Technique

The cleft area is widely exposed through incisions along the edges of the cleft. The incision on the vestibular side is made along the gingival border. Posteriorly, the incision is extended to the first permanent molar where it is angled up into the sulcus (Fig. 26.1). To provide sufficient mobility of this flap, which is going to cover the graft, it is necessary to cut through the periosteum at the base of the flap. Anteriorly, the incision is extended along the gingival border to the center of the cleft-side central incisor. Vertical incisions are made along the edges of the cleft. On the palatal side, mucoperiosteal flaps are raised along the edges of the cleft. A wide exposure of the cleft area is achieved with these incisions.

During the exposure of the cleft, every effort is made to avoid traumatizing the thin bone lamellae that cover the dental roots adjacent to the cleft. The nasal floor is reconstructed, if necessary, and

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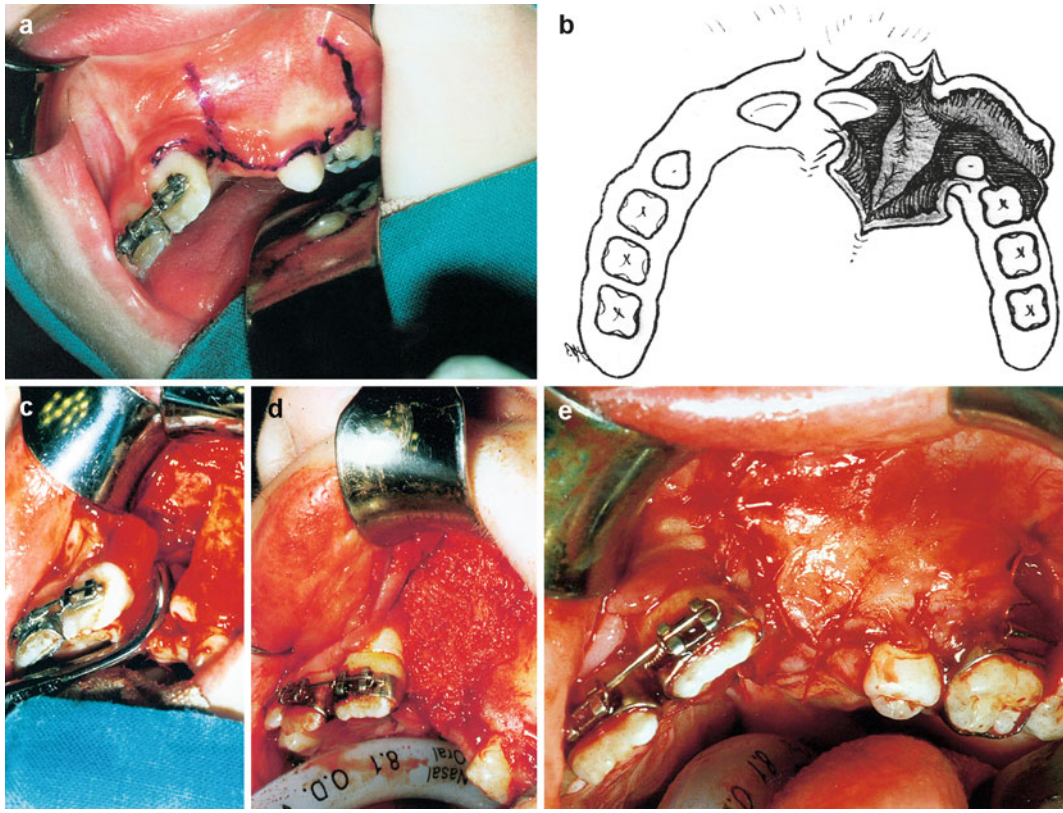

Fig. 26.1 (**a**–**e**) Surgical technique: (**a**) Incision lines. (**b**) Schematic drawing of the raised flaps. (**c**) The alveolar cleft denuded. All soft tissue has been removed. (**d**) The alveolar cleft packed with cancellous chips. (**e**) The posterior flap transferred to cover the graft site. Only attached gingiva covers the inferior part of the alveolar defect

pushed upward. On the palatal side, the mucoperiosteal flaps are sutured together with everting mattress sutures. This leaves a well-defined cavity, whose walls are periosteum and denuded bone.

While the surgeon is exposing the cleft, an assistant has harvested cancellous bone from the anterior iliac crest through a small incision. A trap door of cortical bone is raised, hinged on the inner edge of the iliac crest. Chips of cancellous bone are removed by a sharp spoon, leaving the inner and outer cortex of the iliac bone intact. The donor cavity is filled with hemostatic felt (collagen). The lid of cortical bone is replaced and fastened with sutures.

The alveolar cleft is completely filled with cancellous bone chips. The alveolar crest must be formed up to the normal height and thickness. To improve nasal symmetry, sufficient chips must be placed under the alar base.

The lateral mucoperiosteal flap is advanced to cover the cleft and is sutured to the smaller medial flap and to the palatal flaps. In this way, only attached gingiva will cover the marginal area of the cleft site where the canine later will erupt.

In bilateral clefts, both sides are operated at the same time using the same operative technique.

26.2 Orthodontic Management

26.2.1 Preparation for Bone Grafting

Orthodontic treatment is initiated in the early mixed dentition. Fixed appliance therapy alone has been used.

The maxillary incisors, which often erupt rotated, retroclined, and in anterior crossbite, are corrected for aesthetic reasons and to

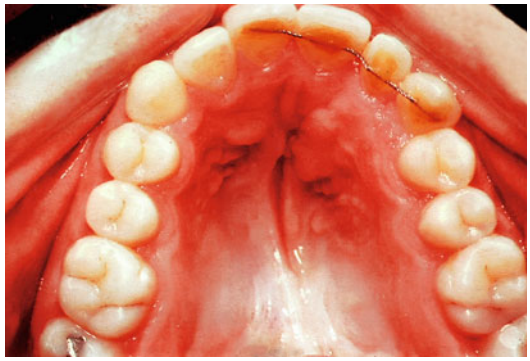

Fig. 26.2 When the orthodontic treatment is finished, a bonded palatal retainer is placed. This retainer is unobtrusive and does not interfere with oral hygiene

facilitate oral hygiene. Treatment generally lasts 3–4 months.

Approximately 25 % of our patients with complete clefts of lip and palate have buccal crossbites attributable to segmental displacement.

Segmental repositioning is done just prior to the bone grafting procedure, and the orthodontic appliance is worn for 3 months postoperatively to retain the arch form. After this time, the bone seems able to retain the transverse dimension of the basal bone (any dentoalveolar relapse is corrected later). In patients with bilateral clefts, a mobile premaxilla is stabilized with a heavy rectangular arch wire for 3 months postoperatively.

26.2.2 Permanent Dentition Management

When the cleft-side canine has erupted spontaneously, orthodontic treatment is resumed. The alignment of the permanent teeth follows different principles, depending on whether orthodontic or prosthodontic space closure is planned. If at all possible, we prefer orthodontic space closure because we consider bridgework in young adults to be undesirable for a number of reasons. In all patients with a missing lateral incisor in whom orthodontic space closure seems to be possible, every effort is made to move the posterior teeth forward. For these patients, we have found the Delaire-style protraction head gear to be useful. We regard this face mask as an adjunct to orthodontics and not as a means of achieving clinically significant change in skeletal relationships.

In most patients, the treatment period in the permanent dentition lasts 2 years and is completed at the age of 15. A bonded palatal retainer extending to two teeth on either side of the cleft is placed. The retainer is kept in place as long as the patient will accept it. Such retainers are unobtrusive and do not interfere with oral hygiene (Fig. 26.2).

26.3 Optimal Age for Secondary Bone Grafting

In the discussion of this subject, two factors are of particular importance: (a) the curve of growth of the various growth sites in the maxillary complex and (b) the clinical goal of the bone grafting.

Possible interference with postnatal maxillary growth must be considered in identifying an optimum or ideal age for secondary bone grafting. Sagittal and transverse growth of the anterior maxilla has virtually ceased by the age of 8–9 years (Bjørk and Skiller 1976; Sillman 1964). The vertical growth of the maxilla occurs mainly as deposition of additional alveolar bone at the alveolar crest (Bjørk and Skiller 1974, 1976). The continuous eruption of teeth is thought to be the agent that stimulates the formation of alveolar bone. With a viable donor tissue such as autologous cancellous chips, the graft is rapidly transformed into functional alveolar bone responding physiologically to erupting teeth (Fig. 26.3). By spontaneous or orthodontically guided eruption of the canine, the capacity of erupting teeth to generate alveolar bone can be utilized to maintain the general growth in maxillary height.

Cephalometric studies of the Oslo cases have shown that bone grafting of the alveolar cleft following the principles stated earlier and performed between the ages of 8 and 11 years had no adverse effect on anteroposterior or vertical maxillary growth (Semb 1988).

Some authors advocate bone grafting at the age of 5–6 years in order to give any lateral

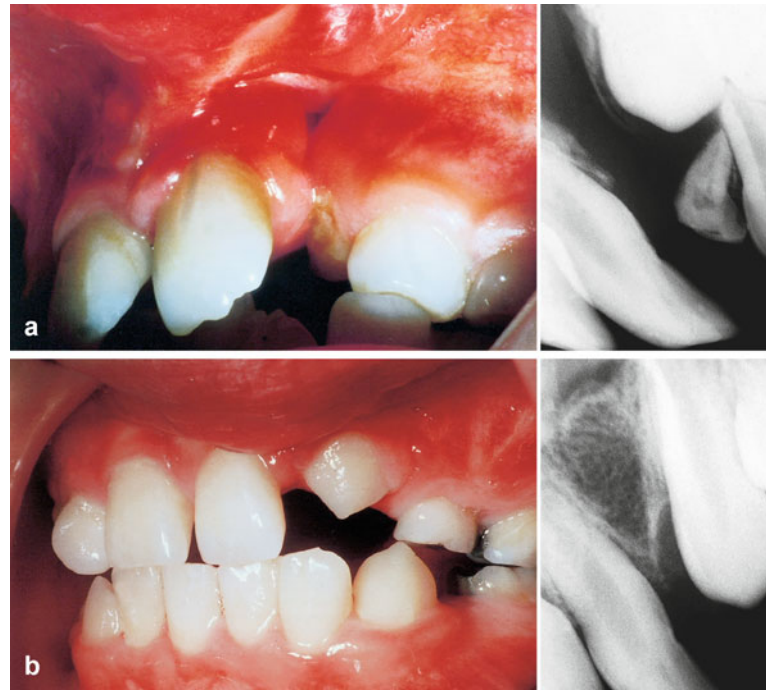

Fig. 26.3 (**a**, **b**) The grafted bone responds physiologically to the erupting canine: (**a**) Alveolar cleft prior to bone grafting. (**b**) The canine erupting normally through the grafted bone

incisor an opportunity to migrate into and erupt through the bone graft. This is a strong argument from a dental point of view. However, further studies are necessary to prove that bone grafting at this age does not interfere with maxillary growth.

A primary goal of secondary bone grafting is achieving orthodontic closure of the cleft space. Orthodontic space closure has proven to be easier when bone grafting is performed prior to the eruption of the cleft-side canine. In our studies, in the group in which bone grafting was performed before the full eruption of the canine, orthodontic closure of the gap was possible in 93 % of the cases (335 out of 360). In the group in which the bone graft was performed in the permanent dentition, the success rate was 72 % (107 out of 149) (Abyholm et al. 1981; Åbyholm and Semb 1992; Bergland et al. 1986a).

An evaluation of the height of the interalveolar bony septum also showed a difference between the two groups with a better height of the interalveolar septum in patients who had the bone grafting performed before the full eruption of the cleft-side canine (Abyholm et al. 1981; Åbyholm and Semb 1992; Bergland et al. 1986a).

26.3.1 Secondary Bone Grafting

After 25 years of bone grafting in the transitional dentition, we have analyzed 1,070 patients, of whom 232 had bilateral clefts, making the total number of grafted cleft sites 1,302.

Of these, the teeth in the cleft region were in the final position in 992 cleft sites, and thus, the interdental septum could be evaluated. Good bony interdental septum height (3/4 or more of normal septum height) was found in 96 % when the operation was performed before the eruption of the canine and in 85 % when the operation was performed after the eruption of the cleft-side canine. Failure rate: no bone formation in 1.2 %. A complete dental rehabilitation without prosthodontics was achieved in 93 % of the patients when the bone grafting was performed before the eruption of the cleft-side canine.

In order to investigate the fate of the grafted bone, we have examined 18 consecutive patients with unilateral complete CLP with 3D CT scan 20 years after bone grafting (Kolbenstvedt et al. 2002). We found good bony support of the teeth adjacent to the cleft in all cases, but there was less bone in the cleft area than on the normal side. The apertura piriformis was lower on the cleft side in all cases, and there was hypoplasia of the maxilla on the cleft side.

26.4 The Grafting Tissue of Choice

When autogenous cancellous bone is transplanted under optimal conditions, osteogenic cells in the graft will survive, and new bone formation will start within a matter of days. Cancellous grafts subjected to minimal traumatic surgery undergo a rapid revascularization which is important for the long-term result (Albrektsson 1979). Fresh autologous cancellous bone transforms very rapidly into alveolar bone, which responds normally to tooth migration and orthodontic movement of teeth.

A cortical bone graft undergoes a slow transformation. The early establishment of nutrition to the cortical bone cells requires restoration of flow through existing vessels or canaliculi and ingrowth of capillaries. This is a very slow process, and the cortical bone will usually die and be replaced by invasion of bone cells originating from the recipient site. This is a fundamental difference between cortical and cancellous bone grafts in terms of both cell survival and vascularization. The slow transformation of a cortical graft makes the reestablishment of the tooth-bearing function of the alveolar process unfeasible.

26.4.1 Donor Sites

For autotransplantation of bone in cleft patients, several donor sites have been exploited: rib, tibia, skull, chin, and iliac crest. All of these sources have been used successfully. We have, however, found the iliac crest to be the most suitable donor site.

26.4.2 Complications

Serious complications are rare. Failures (i.e., no continuous bone bridge across the cleft) are probably due to poor surgical technique or infection. External cervical root resorption can occur if the root cementum is mechanically injured during the bone grafting procedure. This can be avoided by delicate surgical technique and by performing the operation at an age when the cervical region of the canine is still covered with bone (Bergland et al. 1986a).

26.5 Important Surgical Details

To achieve optimal results, we have found some details to be of great importance.

26.5.1 Flap Design

It is important that the mucoperiosteal flaps are designed in such a way that only attached gingiva will cover the marginal part of the graft. It is our clinical observation that a completely normal periodontium seems to occur mainly in cases where the flap design has ensured an orthotopic transfer of attached gingiva to cover the marginal part of the graft.

26.5.2 Good Exposure

The cleft must be widely exposed so that all scar tissue in the cleft area can be removed. The nasal floor has to be reconstructed if a fistula is present. It is important to elevate the nasal floor to achieve a good height of the alveolar crest so that the canine can be brought into a vertical position in the cleft area.

26.5.3 Cancellous Bone Only

Only autologous cancellous bone creates bone that responds normally to eruption and orthodontic movement of teeth.

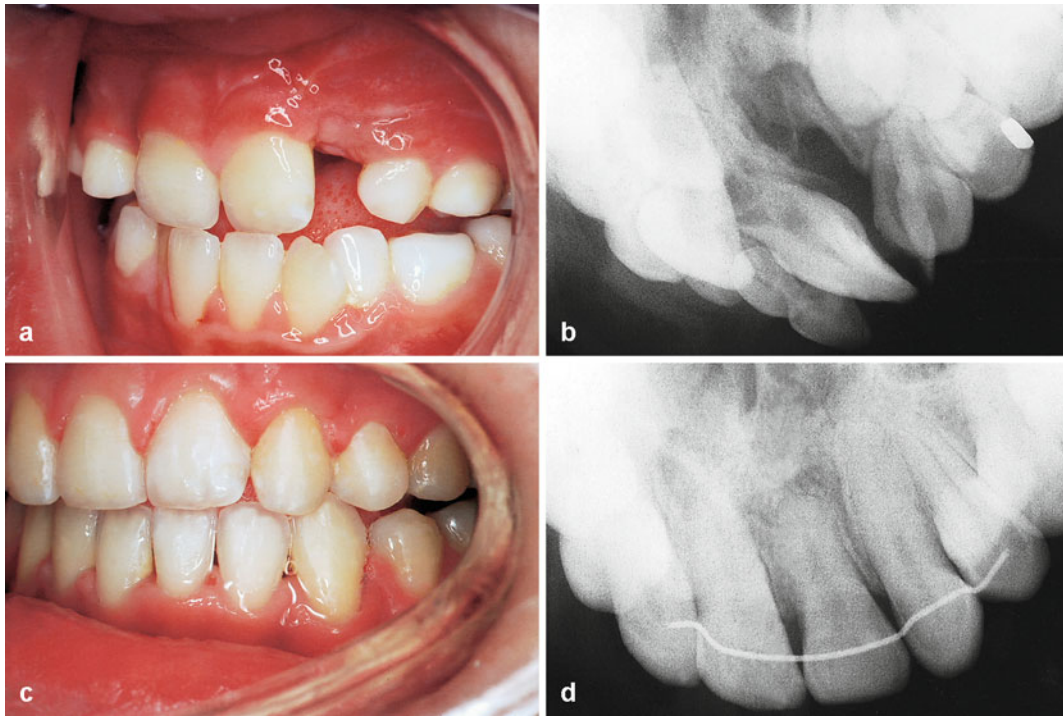

Fig. 26.4 (**a**–**d**) Unilateral complete cleft bone grafted prior to the eruption of the canine: (**a**) The dental arch before the bone grafting. (**b**) Radiograph of the cleft prior to bone grafting. (**c**, **d**) Dental arch and radiograph after the completion of the orthodontic treatment

Conclusion

Autologous cancellous bone grafted to the alveolar cleft between the ages of 8 and 11 years creates alveolar bone which responds normally to eruption and orthodontic movement of teeth. A full dental rehabilitation is possible in the vast majority of cases when bone grafting is performed prior to the cleft-side canine eruption, rendering bridgework unnecessary in cleft patients (Fig. 26.4).

Bone grafting performed between the ages of 8 and 11 years does not restrict maxillary growth.

This combined surgical/orthodontic treatment plan requires close cooperation between plastic surgeon and orthodontist. Fringe benefits include closure of oronasal fistulae and elimination of mucosal recesses, stabilization of maxillary segments, and improvement of nasal symmetry.

26.6 Optimal Timing for Secondary Alveolar Bone Grafting

Sayuri Otaki and Masatomo Yorimoto

26.6.1 Introduction

Timing for secondary alveolar bone grafting is critical to create an area of regenerated bone in the cleft site so that the adjacent teeth (canine and sometimes lateral incisor) can erupt spontaneously or can be moved orthodontically into it. Bone grafting before the eruption of these teeth causes their eruption through the newly grafted bone (the canine proceeds to lateral incisor position when it is missing), inducing additional bone generation and achieve higher interdental septum height. Thereafter, the presence and close

approximation of these teeth prevent bone resorption. In addition, bone grafting before the eruption of these teeth allows the surrounding bone to protect the cervical region of their roots and thus prevent resorption. Maxillary growth is not disturbed when bone grafting is performed to facilitate canine eruption. Careful planning is necessary when bone grafting is performed to facilitate lateral incisor eruption because there is a possibility of causing a maxillary growth disturbance.

Secondary alveolar bone grafting using autogenous marrow and cancellous bone graft was introduced as a method for restoring tooth-bearing function (Boyne and Sands 1972, 1976). Thereafter, the central purpose for secondary alveolar bone grafting is to create an area of regenerated bone in the cleft site that is continuous with the rest of alveolus so that adjacent teeth (canine and sometimes lateral incisor) can erupt spontaneously or can be moved orthodontically into it. The timing for secondary alveolar bone grafting is critical to attain this purpose. There is a margin of time for bone grafting for other purposes, such as the stabilization of the maxillary segments, improving the soft tissue configuration to facilitate prosthetic restoration, preparing bed for the dental endosseous implant insertion, allowing closure of the oronasal fistula, and adding bony support to the alar base.

The timing of secondary alveolar bone grafting will affect the height of interdental septum at the former cleft site; tooth eruption and movement into former cleft site, which in turn affect the type of closure of the space in dental arch; root resorption; and maxillary growth. The optimal timing is going to be discussed with regard to these factors. Although various authors have presented the optimal timing based on their experience and/or investigations (Table 26.1), most practitioners so far accept that the optimal timing for secondary bone grafting is when the patient is 9–10 years old, before eruption of the canine, and the canine root is one-half to two-thirds formed. The canine eruption seems to be accelerated when canine root is two-thirds formed (Vig 1999).

26.6.2 Interdental Septum Height of the Former Cleft Site

Higher interdental septum height can be achieved if bone grafting is performed before cleft-side canine eruption because erupting teeth have the potential to induce alveolar bone generation (Abyholm et al. 1981; Bergland et al. 1986a; Vig 1999). This additional bone generation cannot be expected if bone grafting is performed after the canine has erupted. This has been characterized by many authors, usually with statistical significance (Table 26.2). Moreover, a higher rate of alveolar bone heights is obtained if the lateral incisor is present (Bayerlein et al. 2006). The presence and close approximation of the teeth are also important to prevent bone resorption and maintain the bone height (Bayerlein et al. 2006; Bergland et al. 1986a; Ozawa et al. 2007).

In addition, bone grafting at a younger age can take advantage of a younger patients' probable potential for better vascularization and osteogenesis.

26.6.3 Type of Closure of the Space in the Dental Arch

There often is agenesis, aplasia, or malposition of lateral incisors in patients with an alveolar cleft. When lateral incisor is missing or not usable and extracted, the space created in the dental arch must be closed by one way or another. Two major treatment options are orthodontic space closure (mesial movement of the canine to the lateral incisor position) and prosthetic replacement (use of a fixed bridge or endosseous implant). The autotransplantation of mandibular premolars is also another treatment option.

Treatment decisions in non-cleft patients are based on the skeletal pattern, the class of malocclusion, the relationship between the tooth size and dental arch length, and the color and shape of the adjacent canine (De Angelis 2008; Turpin 2004). Orthodontic space closure does not impede occlusal and temporomandibular function, and it is superior to prosthetic replacement with respect

Table 26.1 Optimal timing for secondary alveolar bone grafting

Author(s)	Age (years)	Eruption stage	Root development
Boyne and Sands (1972)	9–11	Before the full eruption of the canine	–
Boyne and Sands (1976)	7	Before the eruption of the lateral incisor	–
Abyholm et al. (1981)	–	Before the eruption of the canine[a]	–
El Deeb et al. (1982)	9–12	–	When 1/4–1/2 of the canine root has formed
Turvey et al. (1984)	8–10	Before the eruption of the canine	When 1/2–2/3 of the canine root has formed
Bergland et al. (1986a)	9–11	Before the full eruption of the canine[a]	–
Enemark et al. (1987)	–	Before the eruption of the canine[a]	–
Brattstrom and McWilliam (1989)	–	After the eruption of the incisors and before the eruption of the canines[a]	–
Rune and Jacobsson (1989)	–	When the canine (or lateral incisor) is in an early stage of eruption	–
Kortebein et al. (1991)	8–10[a]	–	When 1/2–2/3 of the canine root has formed
Lee et al. (1995)	–	Before the eruption of the canine[a]	–
Kalaaji et al. (1996)	–	Before the eruption of the canine[a]	–
Denny et al. (1999)	12[a]	–	–
Lilja et al. (2000)	7–9	When the lateral incisor or the canine was covered by a thin shell of bone	–
Newlands (2000)	–	Before the eruption of the canine[a]	–
Jia et al. (2006)	–	Before the eruption of the canine[a]	–
Ozawa et al. (2007)	5–7[b]	Earlier than the canine eruption time[b]	–
Rawashdeh and Al Nimri (2007)	–	Before the eruption of the canine[a]	–
Precious (2009)	5.5–6[a]	At the time of eruption of the central incisor	–

[a]Supportive data are presented
[b]When there is a germ of the lateral incisor

to the periodontal health and patient satisfaction (Nordquist and McNeill 1975; Robertsson and Mohlin 2000). However, the optimal treatment for missing lateral incisors in non-cleft patients is still under discussion because of the lack of evaluation of endosseous implant replacement in these studies and the improvement in prosthetic techniques.

However, orthodontic space closure has been the preferred treatment in patients with cleft lip and palate whose cleft-side lateral incisor is missing because of the presence of an alveolar cleft to be bone grafted. The inherent mesial migration of teeth makes the canine erupt into the former cleft site through the newly grafted bone if bone grafting is performed before canine eruption, and additional bone is induced as it erupts (see Sect. 26.7.2), which eventually enhances the possibility of subsequent orthodontic space closure. In addition, bone resorption is significantly lower with orthodontic space closure than with prosthetic replacement (Schultze-Mosgau et al. 2003). This correlates with the prevention of bone resorption by the presence and close approximation of the teeth (see Sect. 26.7.2).

However, there are cases with a missing lateral incisor in which space opening and prosthetic replacement yield superior aesthetic results in comparison to the orthodontic closure of the space (Dempf et al. 2002; Enemark et al. 1985; Newlands 2000). The lateral incisor space is maintained following bone grafting until the final treatment in these cases, usually after the termination of growth, which inevitably causes bone resorption. Endosseous implant replacement usually requires a regrafting procedure since adequate bone volume and quality is necessary for

Table 26.2 Rates of sufficient bone formation

Author(s)	Canine not erupted	Canine erupted	
		Canine half erupted	Canine fully erupted
Abyholm et al. (1981)			
All clefts	13/14 (92.9 %)	10/12 (83.3 %)	33/43 (76.7 %)
Bergland et al. (1986a)			
Unilateral complete clefts	97 % ($n=69$)		76 % ($n=50$)
Bilateral complete clefts	91 % ($n=22$)		82 % ($n=49$)
All clefts	96 % ($n=143$)		85 % ($n=149$)
Enemark et al. (1987)			
CLA	18/19 (94.7 %)	5/8 (62.5 %)	
UCLP	40/55 (72.7 %)	26/52 (50.0 %)	
BCLP	28/40 (70.0 %)	7/24 (29.2 %)	
Loh et al. (1988)			
All clefts	17/25 (68.0 %)	6/33 (18.2 %)	
Brattstrom and McWilliam (1989)			
UCLP	74/101 (73.3 %)	60/100 (60 %)	
Lee et al. (1995)			
All clefts	36/49 (73.5 %)	23/52 (44.2 %)	
Kalaaji et al. (1996)			
CUCLP	13/14 (93 %)	25/33 (76 %)	
Jia et al. (2006)			
CLA	15/16 (94 %)	23/24 (96 %)	
UCLP	42/44 (95 %)	48/58 (83 %)	
BCLP	20/22 (91 %)	23/34 (68 %)	
Rawashdeh and Al Nimri (2007)			
UCLP	20/22 (90 %)	20/25 (80 %)	
BCLP	9/16 (56.2 %)	8/14 (57.1 %)	

The rates of clefts with a septum height greater than 3/4 of normal height on dental radiographs (correspond to Bergland types I and II) are shown
CLA cleft lip and alveolus, *UCLP* unilateral cleft lip and palate, *BCLP* bilateral cleft lip and palate, *CBCLP* complete bilateral cleft lip and palate, *CUCLP* complete unilateral cleft lip and palate

endosseous implant placement. The functional stimulation of the transplanted bone by mastication can limit the bone resorption following the insertion of an implant (Dempf et al. 2002; Jansma et al. 1999; Kearns et al. 1997).

26.6.4 Root Resorption

The periodontal ligament and cementum on the root surfaces are in danger of being damaged during the bone grafting procedure. These are thought to be the cause of root resorption at the cementoenamel junction. Bone grafting before the root of the adjacent teeth appears from bone is preferable to avoid this complication. Great care should be taken not to injure the cervical region of these teeth during elevation of the mucoperiosteal flap when bone grafting is performed after the full eruption of the canine or when there is a useful lateral incisor exposed. There are reports that describe less root resorption when bone grafting was performed prior to canine eruption (Table 26.3).

26.6.5 Maxillary Growth

Secondary alveolar bone grafting using cancellous bone graft is unlikely to significantly interfere with the subsequent maxillary growth because of

Table 26.3 Root resorption rates

Author(s)	Canine not erupted	Canine erupted	
		Canine half erupted	Canine fully erupted
Enemark et al. (1985)	None in 23 clefts		2 teeth in 33 clefts
Bergland et al. (1986a)	None in 143 clefts		15 teeth in 149 clefts
Bergland et al. (1986b)	None in 39 clefts		2 teeth in 43 clefts
Enemark et al. (1987)	None in 94 patients	17 patients in 130 patients	

the growth pattern of the maxilla, if it is performed at or over the age of 8 (Abyholm et al. 1981). This theory is supported by subsequent cephalometric evaluations (Daskalogiannakis and Ross 1997; Levitt et al. 1999; Semb 1988; Trotman et al. 1997). Therefore, maxillary growth disturbance is not a problem when secondary bone grafting is performed to facilitate canine eruption. Careful planning is necessary when bone grafting is performed to facilitate eruption of the lateral incisor. The reported earliest ages for bone grafting to facilitate lateral incisor eruption are 6.3 years (Lilja et al. 2000) and 5 years (Shashua and Omnell 2000).

Few investigations have so far addressed maxillary growth after earlier secondary alveolar bone grafting performed under 8 years of age. Most investigations focused on the growth pattern of patients with cleft lip and palate (operated or not operated) and more urgent growth disturbance after primary bone grafting or gingivoperiosteoplasty. One study investigated the influence of earlier secondary alveolar bone grafting on maxillary growth. Bone grafting between the ages of 6 years 10 months and 10 years 10 months does not affect maxillofacial growth during the first to third postoperative years (Chang et al. 2005). Although further investigation is necessary to determine the long-term effects, there are no reports of deleterious effects on maxillary growth by secondary bone grafting at the stage of mixed dentition using cancellous bone graft.

Optimal timing for secondary alveolar bone grafting is best determined by eruption stage of the teeth in the cleft region rather than by the chronological age. It is evident that bone grafting prior to the eruption of the cleft-side canine is preferable. There is no consensus on whether to perform even earlier bone grafting to facilitate lateral incisor eruption. Higher interdental septum height will be achieved and maintained, and better aesthetic results may be achieved with the lateral incisor in the arch; however, this could be associated with possible growth disturbance and immaturity of the donor site. Although some researchers worry about the quality of the lateral incisors, current restorative techniques should solve these problems. An increased risk of canine impaction in relation to earlier bone grafting is reported but can be solved by surgical exposure and/or orthodontic assistance (El Deeb et al. 1982; Enemark et al. 1987). Further evidence showed that bone grafting to facilitate lateral incisor eruption as early as 5 years of age does not interfere with maxillary growth, and the resolution of the donor problem will thus make it possible to establish the optimal timing for secondary alveolar bone grafting. Bone grafting should now be performed on patients under 8 years of age only when the benefits overweigh the risk of possible growth disturbance.

Timing of secondary bone grafting must be coordinated with orthodontic treatment. This requires close cooperation between the surgeon and orthodontist. The use of a nonviable cortical bone graft requires that arch expansion and orthodontic movement of teeth must be finished before bone grafting. A viable bone graft allows arch expansion and orthodontic movement of teeth before and/or after bone grafting, although many surgeons seem to prefer arch expansion prior to bone grafting to improve access to the cleft and nasal floor.

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The Need for Differential Diagnosis in Treatment Planning

27

Samuel Berkowitz

Samuel Pruzansky once said that craniofacial surgery is "an experiment on nature's experiment." This statement is certainly true. All facial skeletal surgery – in growing or nongrowing patients – can be regarded as an investigation of craniofacial growth, form, and function.

Because facial skeletal surgery in growing children often affects craniofacial growth as well as function, informed decisions should be made concerning which structures need to be repositioned and reformed. Based on these decisions, a treatment plan is then formulated, and a working hypothesis for successful treatment is established. Three points need to be made at this juncture. First, remembering the value of failures as learning opportunities, clinicians cannot afford to forget failures; rather they must thoroughly analyze them so they are not repeated. Second, clinical investigators must be able to explain why some surgical procedures are successful and others fail. Third, clinicians must be able to fit the proper procedure to each individual problem and be willing to work with the consequences of their choices.

Not all clefts of the lip and/or palate within the same cleft type are alike.

1. The collected serial casts and cephalometric radiographs, beginning with those of the unoperated infant and continuing through adolescence presented in this chapter, provide a view of the wide spectrum of variations encountered within each cleft type in its untreated state and a record of the changes that occurred thereafter resulting from natural growth or specific therapeutic procedures. Clinical experience points out one critically important, fundamental fact: All clefts cannot be lumped together as a single phenomenon. Within each type of cleft, there are great individual differences in the geometry and extent of the cleft defect, and these differences are clinically significant.

   In a state-of-the-art monograph in 1972, Spriestersbach and coworkers (1973) wrote "Perhaps the greatest drawback to genetical and epidemiological research on clefts of the lip and palate has been the unfortunate tendency to lump them together." Twenty years prior to that report, the first line in the first paper to emerge from Pruzansky's (1969) research stated "Not all congenital clefts of the lip and palate are alike." This statement was to become the leitmotif of his subsequent research. He took great care to demarcate

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samples according to varying cleft types in his designs for epidemiological, morphological, functional, and genetic research.

2. Current methods of treatment, which favor staged treatment (i.e., closing the lip at birth and the palate at a later age, in one or two stages), offer a more encouraging prognosis than those that prevailed 50 years ago.
3. The age of the patient and the type of surgery applied are two variables in determining the effect of surgery on facial growth. Quantitative and qualitative characteristics of the cleft defect, plus the general health and genotype (facial growth pattern) of the individual patient are additional determining factors. Under certain conditions, surgical repair of the palate is feasible quite early; in others, optimal conditions for repair will not become evident until a later age.
4. The natural history of children with clefts and those with specific syndromes demonstrates that some improve over time, some grow worse, and others remain unchanged despite the surgical effort.
5. Presurgical orthopedics, except for the use of a facial elastic to ventroflex the premaxilla to aid the surgeon prior to uniting the lip, have no long-term utility, and primary bone grafting has a deleterious effect on palatal and facial growth.
6. A critical review of the literature on the clinical management of cleft lip and cleft palate, together with an evaluation of the cumulative data from longitudinal palatal growth studies, has led most orthodontists to the following hypothesis: Conservative lip and palatal surgery facilitates rather than inhibits growth in both the maxillofacial skeletal complex and the soft tissue of the labio–facial complex. In cleft palate cases, operative intervention which minimally involves bone growth potential will guide maxillofacial growth in the individual in such a way that postoperative "catch-up" growth of the palate will result in acceptably normal development.
7. Within defined limits of mechanical and professional capability, the morphological and spatial relationships of the cleft palatal segments and facial growth patterns are the major determinants of the ultimate occlusion and arch form (not size). These variables, unique for each patient, could well be more indicative of the final treatment outcome than differences in the treatments employed by surgeons.
8. At the time the palatal cleft is closed, the relationship of the size and shape of the cleft space to the amount of available soft (mucoperiosteal) tissue surrounding the cleft, and the geometric relationship of the palatal processes to each other, is basic to determining the influence that scarring will have on the palatal arch form and the ability of the palate to develop normally.
9. Most skeletal malformations in cleft patients are the result of surgical procedures that have caused some growth retardation or of osteogenic deficiencies that lead to maxillary hypoplasia. All maxillary discrepancies are three-dimensional.
10. The concept that an increase in the amount of palatal scarring, beyond some critical threshold level, can reduce the palatal growth increments and cause palatal deformation appears to be valid because the same surgical procedures, performed by the same surgeon on the same type of cleft, often lead to different palatal relationships. The reason for the different outcomes may, therefore, be due to variations in the palatal deformity at the time of surgery (i.e., the relative size of the cleft space to the size of the palatal segments that need to contribute soft tissue for cleft closure). The larger the cleft space relative to the amount of available tissue, the larger the area of denuded bone that must be left when the undermined palatal mucoperiosteum is moved medially to close the cleft space. The denuded bone heals by epithelialization, becoming a scar. The greater the scarring, the more growth retardation and palatal deformation.
11. Although the tongue has been found to occupy the cleft space and be carried high into the nose at birth, no studies have shown that abnormal tongue habits negatively affect

speech development. It appears that, with the closure of the palatal cleft between 20 and 30 months, and without the use of an obturator, children usually develop good speech if the velopharyngeal closure mechanism is functionally adequate.

12. There is no documented evidence that the cleft condition interferes with body growth or that, in most instances, the palatal defect cannot be effectively treated without feeding appliances. However, obturators may be useful in some neurological disturbances when palatal closure needs to be delayed beyond 3 years of age and parents complain of feeding problems. Most pediatricians and nurses recommend the use of a soft plastic feeding bag (e.g., Playtex nurser) or a soft plastic bottle (e.g., Mead-Johnson's nurser) with a crosscut, normal-sized nipple. The use of Lamb's and Ross Laboratory nipples is strongly discouraged because of their abnormal shape and nipple length.
13. A child with a Pierre Robin sequence should never be given an obturator because the child's oral volume is already too small and an appliance will further compromise tongue positioning. Because the infant has a micrognathic mandible, the tongue must be carried high into the palatal cleft space during this critical early adjustment period. If an obturator or early palatal surgery is utilized for these children, it can force the tongue downward and backward, possibly closing off the airway space and interfering with breathing.
14. The use of a head bonnet with a facial elastic band or the use of elastic taped to the cheeks across the lips to reduce palatal distortion are acceptable methods to help the surgeon reduce tension at the surgical site. Such innocuous external facial forces will help bring the distorted lip and skeletal segments into a more normal relationship. This mode of treatment is acceptable to most parents and clinicians.
15. There is no proof that neonatal maxillary orthopedic appliances will stimulate palatal growth or reduce middle ear infections (Berkowitz 1977), nor has it has ever been shown that these orthopedic procedures will prevent the need for future orthodontia and improve speech development. An obturator will be of some help if the cleft space remains open after 3 years of age and neurological problems interfere with feeding.
16. In many cases, protraction orthopedic forces can protrude the maxillary complex sufficiently to negate the need for surgical advancement. These forces are most efficient when applied before or during the pubertal growth spurt. After puberty, the effects change from orthopedic (bone) to orthodontic (dental) movements. The use of palatal expansion forces prior to the application of protraction devices can increase the potential for orthopedic movement of the maxilla.

Once midfacial recessiveness occurs at an early age, for example after premaxillary orthopedic retraction, it will not show increased growth acceleration to spontaneously improve midfacial skeletal and dental relationships.

27.1 A New Direction for Cleft Research

Successful outcomes in the treatment of complete unilateral cleft lip and palate (CUCLP) and complete bilateral cleft lip and palate (CBCLP) are not universally obtained, despite significant improvements in surgical techniques over the past three decades. In particular, deficient palatal growth may occur even when treatment is rendered by expert teams. The factors that contribute most significantly to unfavorable growth outcomes remain obscure.

Although the treatment of cleft lip and cleft palate has progressed markedly in the last 50 years, there is still a great need for improvement in diagnosis and treatment planning. However, to accomplish this goal, our current diagnostic categories may need to be revised. The possibility that clefts that are similarly classified may react differently

to the same surgical procedure must be examined. The ultimate aim of future research is to provide a better objective understanding of the reasons for, and the characteristics of, these differing outcomes, and by so doing provide a broader and more informative knowledge base for making diagnostic and treatment decisions concerning cleft lip and cleft palate.

No matter what type of treatment surgeons have favored, they have not been able to explain why their surgical method of choice, when performed on similar clefts at the same age, often yielded different results. Why some cases appear to show "catch-up growth," resulting in good facial and palatal form and functional dental occlusion, while others show poor facial and palatal development remains an enigma. Among the specific unanswered questions: Were the different outcomes due to different levels of skill on the part of the operators? Were there significant differences in the palatal deformity at the time of cleft closure surgery within each cleft type that should have been differentially diagnosed? And does presurgical orthopedics influence palatal growth or does it merely act to reposition palatal segments?

Catch-up growth has been defined by Hughes (1982) as growth with a velocity above the statistical limits of normality for age during a defined period of time. Such an increase in the rate of growth, before and after palatal surgery, with or without neonatal maxillary orthopedics, may allow the palate to attain its normal adult size or, with reduced velocity, the palate may still fail to do so. The latter case is called "incomplete" catch-up growth. Wilson and Osbourn (1960) showed that the duration and severity of the insult (the scarring resulting from the surgical procedure used to close the cleft space in the hard palate) may positively or negatively affect the ability of the palate to recover and undergo catch-up growth. The developmental age of the infant at the time of the insult and the nature of the insult itself (extent of denuded bone left after surgery and the resulting scarring) will affect the ability of the infant to achieve complete catch-up growth.

27.2 Clinical Research Feinstein (1970)

Clinical Research Feinstein (1970) wrote:

> In the biostatistical architecture of clinical research, the first operational principle is to specify the components and choose the logic of the objective of the research. The components consist of a sequence of initial state, maneuver and subsequent state. The logic consists of suitable scientific judgment in the decisions made to demarcate the diagnostic and prognostic conditions of the initial state of the population; to identify differentiate and prognostically correlate the diverse targets of the subsequent state; and to choose maneuvers that are satisfactory in potency, comparison, multiplicity and concurrency.

In speaking of the initial and the subsequent states, emphasis will be placed on studies of casts starting at birth and extending through adolescence.

27.2.1 Initial State

The size and form of the palatal segments are measured serially starting at birth and divided into two periods. The first period ends at surgery to close the palatal cleft. The second period includes the cleft space with the changing size of the palatal segments.

Analyses of the initial state prior to palatal surgery (end of first period) suggest that, under certain conditions, surgical repair of the palate is feasible quite early, whereas in other instances, optimal conditions for repair will not be present until a later age. In our experience, a selected number of cases with very small cleft spaces underwent palatal repair at or before 1 year of age without detriment to midface and palatal growth. On the other hand, there are cases where the cleft space is too large, compared to the amount of available soft tissue, and surgery needs to be postponed to avoid creating growth-inhibiting scar tissue. This is an example of individualized differential diagnosis and treatment planning.

27.2.2 Maneuver: Presurgical Orthopedics and Surgical Procedures Used to Close the Palatal Cleft

If we assume that qualified surgeons within a given institution or region, practicing a specific series of techniques over a given period of time represent a constant, differences in success or failure should reside in (1) the initial state (the geometric and size relationship of the palatal segments to the size and shape of the cleft space, which reflects the degree of skeletal deficiency as well as palatal segment displacement) and (2) the facial growth pattern. Of course, the sample must separate cases subjected or not subjected to presurgical maxillary orthopedics, as well as cases utilizing various cleft closure procedures because these variables can influence the subsequent state.

Of the three components, the maneuver presented the greatest number of confounding variables. Differences between surgeons, variance in the performance by the same surgeon from day to day and over the course of several years, and differences in techniques, which are difficult to identify and compare, complicate the analysis. However, our biostatisticians believe that research objectives to test the influence of presurgical orthopedic treatment and the relationship of cleft palate space to surgical outcome can be reached. It is possible to statistically test and covary for effects due to differences between and within surgeons.

As Feinstein stated, we too believe that, within certain defined limits, the success or failure of the surgical procedure depends more on the initial state than on the variables inherent in the maneuver. To put it another way, we expect that subtle differences among patients will be more prognostic of the subsequent state than differences between surgeons.

Serial facial and palatal growth studies starting at the newborn period (Berkowitz 1985) have shown that too many factors were operating in relation to the patients under study to permit the formulation of simple, all-inclusive rules, such as any suggestion regarding the age at which clefts of the palate should be repaired. Berkowitz (1985) therefore hypothesized that, at the time of palatal surgery, the ratio of the useful mucoperiosteal tissue available to the size of the cleft space determined the area of denuded bone left at the surgical site after the medial movement of palatal soft tissue. This area heals by epithelialization, which in turn, becomes scar tissue. The degree of scarring could spell the difference between therapeutic success and failure because it influences the palate's ultimate size (osseous plus soft tissue) and form.

If presurgical orthopedics enhance palatal growth and development, the cleft space in the 20–24-month period will be much smaller relative to the enlarged palatal segments than in cases that have not been similarly treated. This hypothesis needs to be tested using quantitative measurements. Only in this way will surgeons find reason to change their focus to include the size and form of the palate and the extent of the cleft defect, as well as the surgical orthopedic procedures, in differential diagnosis.

Pruzansky (1953) frequently stated that his most important contribution to the cleft palate literature was the conclusion that "cleft lip and the palate does not represent a single fixed entity subject to generalizations of description and classification and least of all to rigid therapeutic formulas." Although his clinical reports supported this conclusion, Pruzansky did not have a sufficient number of cases and proper cast-measuring equipment to study the natural history of cleft palate growth in relationship to palatal surgery in order to individualize treatment planning. The question of the role and importance of tissue adequacy or inadequacy could not be explored until a highly accurate three-dimensional measuring tool and supporting CadCam software became available.

27.3 Palatal Embryopathology

Studies of clefts have produced conflicting interpretations regarding deficiency in mass and/or displacement of the palatal segments in space, as

well as the effects of cleft surgery on palatal growth. Information relating to the complexities of embryonic facial development is fundamental to an understanding of the growth potential of the primary and secondary palate. Developmental studies (Slavkin 1979; Ross and Johnston 1972) have shown that the facial mesenchyme, which gives rise to the skeletal and connective tissues, originates from neural crest cells and undergoes extensive migration and interaction.

Coalescence of the facial processes results in the formation of the primary palate, which constitutes the initial separation between the oral and nasal cavities and eventually gives rise to portions of the upper lip and anterior maxilla. The exact mechanism of primary palate formation is not clear. However, most clefts of the primary palate appear to result from variable degrees of mesenchymal deficiency in the facial processes.

The suspected causes of clefts of the secondary palate are also varied. Slavkin (1979) proposed several possible mechanisms:

1. Tongue resistance: The tongue, arched up between the shelves, delays palatal shelf movement.
2. Decreased shelf forces: Although there are no examples of mutant genes that can cause this, there are many teratogens for which this mechanism has been invoked.
3. Failure to fuse: This possible cause may be associated with delayed shelf reorientation.
4. Narrow shelves: This theory suggests that the palatal shelves can move normally enough to reach the horizontal, yet still be too narrow to reach each other. This condition could be explained by a more generalized deficiency of facial mesenchyme reaching the palatal area, making the hard palatal shelves and soft palate inherently smaller.

The causative factor has important clinical implications because it suggests that, in some unilateral clefts of the lip and palate, the size of the cleft space may be disproportionately very large and more variable in shape than in other clefts of the secondary palate. The velum in this cleft type also may be deficient in muscular tissue and predispose the child to velopharyngeal incompetency. Thus, it would be helpful to be able to identify infants with skeletomuscular deficiencies at an early age (within the first 2 years of life) in order to customize the cleft closure procedure to enhance proper speech production as well as normal palatal growth and development. Obviously, a child with palatal tissue deficiency will have a different set of problems than a cleft palate patient with adequate palatal tissue and a cleft caused by failure of proper shelf force or failure to fuse.

27.4 The Neonatal Palatal Form in Complete Clefts of the Lip and Palate

27.4.1 The Effect of Muscle Forces

The normal palatal arch form is determined by the result of the compressive forces of the orbicularis oris–buccinator–constrictor pharyngis superioris muscle ring counteracted by the protrusive and expansive forces of the tongue. However, in the presence of clefts of the lip and palate, aberrant muscle forces cause the lip and palatal segments to be distorted in space. The lateral pull of the cleft lip musculature, coupled with the pushing forces of the tongue fitting within the cleft space, is unrestrained (Subtelny 1990).

27.4.2 The Influence of Cleft Surgery on Palatal Form and Growth

When the cleft lip and/or soft palate are united, the cleft musculature forces are reversed, causing the laterally displaced skeletal structures to move medially into a more normal form. The increased tension of the facial musculature may vary in degree among patients and with the type of lip surgery employed. No attempt will be made to measure these forces; for the same reasons, they are not measured in standard orthodontic treatment planning: it is impractical! The role of lip tension on palatal arch form, however, does deserve further investigation.

Slaughter et al. (1956) first recognized the many anatomic variations within similarly classified clefts and suggested that there are great differences in the amount and quality of palatal tissue among the several cleft types and within

any one type. The amount of palatal tissue relative to cleft size increases with growth, but the timing of this growth varies from one person to another. In some patients, the greatest proportional changes occur earlier than in other patients so that cleft space closure may have to be delayed to avoid growth-inhibiting scar tissue; such findings were verified by Pruzansky (1955, 1957), Pruzansky and Lis (1958), Pruzansky and Aduss (1964), Pruzansky et al. (1973), Lis et al. (1956), and Berkowitz (1985). Krogman et al. (1979) observed postoperative catch-up growth in almost every case they studied and concluded that, by the age of 6 years, the maxillary complex is usually acceptably normal. Berkowitz et al. (1974) and Mapes et al. (1974) further reported that, after palate surgery, there may be a growth lag from 14 to 20 months, but subsequently the processes of orderly development may take over, and the rate of growth may even accelerate.

Berkowitz's observations (as Pruzansky and Aduss (1964) did earlier) over the last 25 years have shown that, after the lip is united, the displaced palatal segments will assume various relationships to each other (some may overlap, others may butt join, and still others not touch due to premature contact of the inferior turbinate on the cleft side with the nasal septum). There seems to be a correlation of arch form, seen in the deciduous dentition, with the size and geometric relationship of the palatal segments at birth. For example, in complete unilateral clefts of the lip and palate, after the lip is united, cases with a very long noncleft palatal segment and a short cleft segment coupled with a small anterior cleft space are more likely to have the segments overlap. Other variables such as steepness of the palatal slopes and the adequacy of tissue need to be considered as well.

27.5 The Need for Three-Dimensional Measuring Techniques

Assessing the geometric form of the palate prior to closure of the cleft space will enable recommendations to be made, not only for the most beneficial surgical procedures, but also for the most opportune time to perform palatal cleft closure surgery. For example, various surgical procedures to close the palatal cleft, such as those using the V–Y and the von Langenbeck surgical techniques, involve both the anterior–posterior and/or medial movement of mucoperiosteum from the right and left palatal segments. Movement of the palatal mucoperiosteum leaves areas of denuded bone at the line of incision that heal by contraction and epithelialization (scarring). The concept that an increase in the amount of palatal scarring, beyond some critical threshold level, can reduce the palatal growth increments and cause palatal deformation would appear to have validity because the same surgical procedure, performed by the same surgeon on the same type of cleft, but with different cleft space size, often leads to different palatal relationships. One of the reasons for the different outcomes, therefore, may be variations in the palatal deformity at the time of surgery, more specifically, the size of the cleft space relative to the size of the palatal segments that contribute soft tissue for cleft closure.

Quantitative information regarding the normal palate is noticeably sparse because of measuring limitations inherent in using various forms of calipers and rulers. Some linear two-dimensional studies on the form of the newborn arch were performed by Ashley-Montague (1934), Sillman (1964), Richardson (1967), and Brash (1924). Their measurements, limited to maximum breadth, maximum length, maximum posterior breadth, and maximum lateral sulcus breadth, produced two-dimensional tables starting at birth.

Xerographic studies of casts were an advance over previous measuring systems because they permitted a more accurate description of two-dimensional changes in surface area. Huddart (1970, 1985) concluded from these measurements that, in complete unilateral clefts of lip and/or palate (CUCL/P), the palatal surface area is deficient by age 16 compared with a normal population of the same age. Huddart suggested that presurgical orthopedics may actually hinder palatal growth. In 1971, Mazaheri et al. (1971) reported on changes in arch form and dimensions associated with unilateral clefts of lip and palate and cleft palate. They found a significant pattern of anteroposterior and lateral growth retardation immediately after surgical treatment. Stockli

(1971), who was very critical of his own research approach, reported that there are great limitations in the use of xerography for the study of cleft palate casts. He emphasized that arch form must be considered in the treatment of an infant with complete cleft of the lip and palate, and he recognized that three-dimensional measurements would be more appropriate for longitudinal and comparative studies.

At present in the realm of cleft lip and cleft palate therapy, treatment planning is at best an "educated art." Clinical reports of various treatment protocols, emanating from the many and widely separated cleft lip and palate treatment centers, are usually anecdotal and understandably supportive of the clinics' own treatment concepts. Although the protocols may differ significantly, the authors tend to be satisfied with their own patients' facial, dental, and speech outcomes, all of which encourages few if any innovations in treatment approaches.

Certain questions inevitably arise: Do several different surgical procedures yield universally acceptable results that allow for normal palatal development? Are the outcome reports self-serving or can there indeed be a variety of effective surgical procedures? In cases of undeniable failure, what were the errors, if any, in diagnosis and treatment planning? In assessing failures, most surgeons focus solely on the surgical skills and/or surgical protocols involved, but this leaves other possible explanations unexplored. In recent years, it has been suggested that variations in the physical characteristics of the deformity – the geometric relationship of the palatal segments to each other at birth and the size of the cleft space relative to the amount of available soft tissue used to close the cleft spaces – may have an impact on treatment outcomes (Berkowitz et al. 1974; Stockli 1971). Those authors highlighted the importance of three-dimensional measurements and urged that the arch form and the size of the cleft space at the time of surgery be taken into consideration in the treatment of infants with complete clefts of the lip and palate.

Lack of appreciation for the importance of the geometric relationships of the cleft palatal segments to each other has been the result, in great part, of the dearth of longitudinal records, such as serial palate casts and lateral cephaloradiographs, and an accurate palatal cast-measuring device for quantification and computer analysis of the palate's changing geometric form. Just as the microscope uncovered critical differences in tissue pathology, a three-dimensional measuring instrument could reveal palatal geometric information that had heretofore gone unnoticed, and the importance of which has not been appreciated. Fortunately, such a measuring instrument and a significant number of dental casts are now available.

27.6 Studies Using Three-Dimensional Techniques (Figs. 27.1, 27.2, 27.3, 27.4, 27.5, 27.6, and 27.7)

Berkowitz (1971) initiated a study to determine the feasibility of using stereophotogrammetry to graphically describe the changing configuration of cleft palates. Data from the study supported the clinical impressions that palatal molding action with palatal growth, which occurred at the palate's

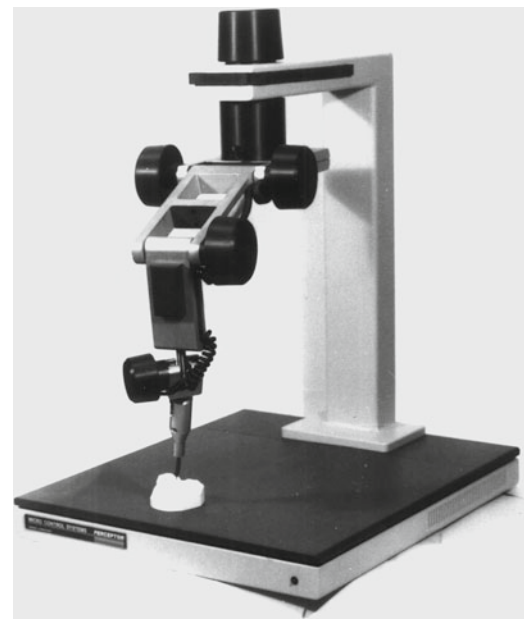

Fig. 27.1 An electromechanical digitizer used to extrapolate *x*, *y*, and *z* coordinates from a plaster palatal cast

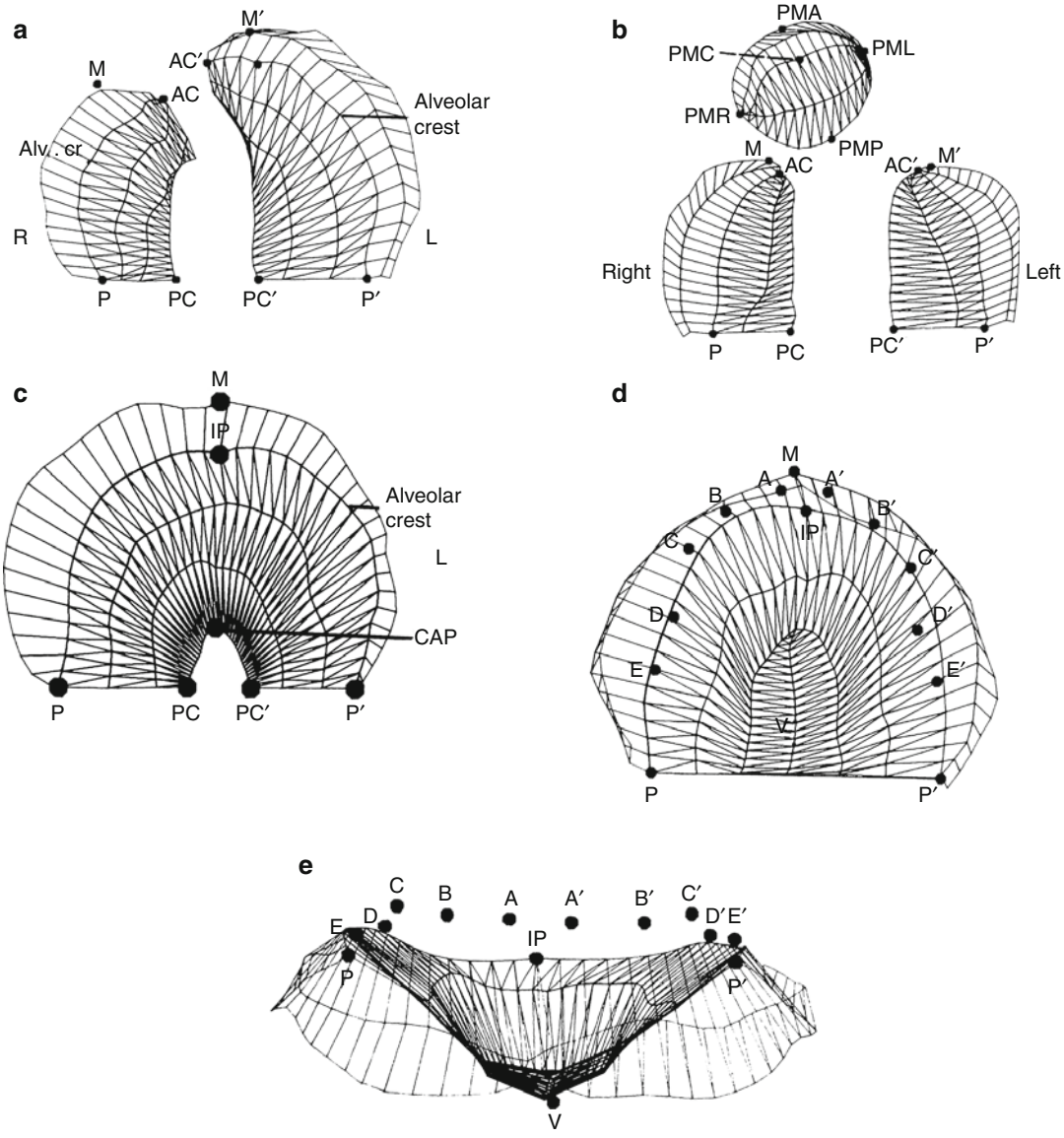

Fig. 27.2 (**a**–**e**) Computer-generated images of various cleft palate types. (**a**) Complete unilateral cleft lip and palate. (**b**) Complete bilateral cleft lip and palate. *PMA* premaxilla anterior, *PMP* premaxilla posterior, *PMC* premaxilla center, *PMR* premaxilla right, *PML* premaxilla left. (**c**) Isolated cleft palate. (**d**) Normal palate: occlusal view. (**e**) Normal palate: posteroanterior view. *P* postgingivale comparable to PTM (pterygomaxillary fissure on a lateral cephalograph). It is the posterior border of the hard palate, *PC* landmark on the PP line at the cleft, *AC* anterior point of the alveolar ridge at the cleft, *M* the most anterior point of the palatal segment, *IP* incisal papilla point, *V* highest vault point, *A* deciduous central incisor, *B* deciduous lateral incisor, *C* deciduous cuspid, *D* deciduous first molar, *E* deciduous second molar. Palatal Surface Area. Before cleft closure: Bounded laterally by P to AC, P to Pc and PC9 to P9, P9 to Ac9. After cleft closure: Includes cleft space bounded by AC to AC9 and PC to PC9. Cleft Space Area: Anterior limit AC–AC and posterior boundary PC to PC9. In Bilateral Clefts: Anterior Cleft Space: Bounded anteriorly by the premaxilla's outer point of the alveolar crest RPM or LPM to AC and posteriorly by line AC to AC9. Posterior Cleft Space: Bounded by AC to AC, AC to PC, and PC to PC9

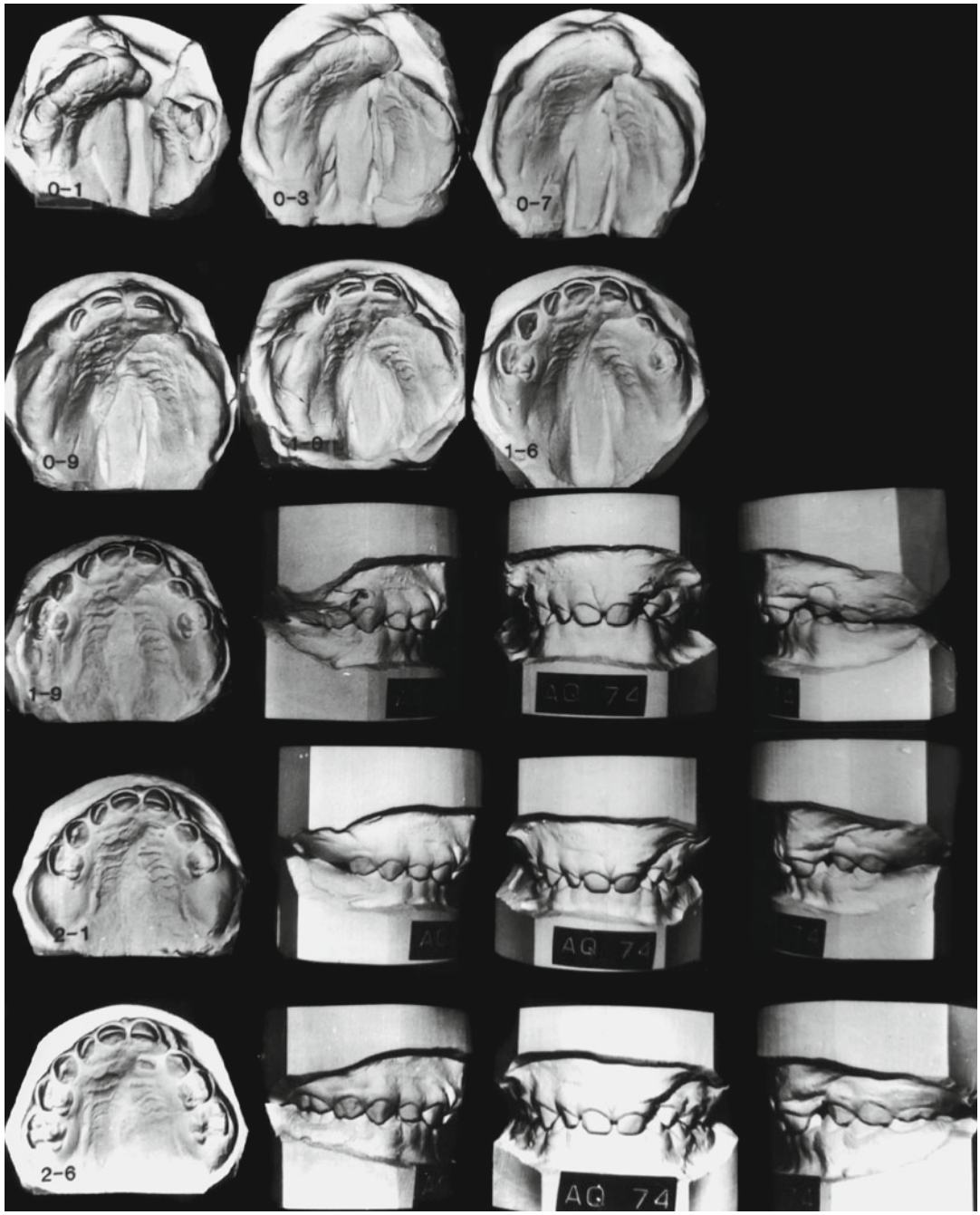

Fig. 27.3 Serial dental casts for case JH (AQ-74) show: 0–1. Separated palatal segments soon after birth. 0–3. Palatal segments move together forming a butt joint relationship 0–7, 0–9, 1–6, and 1–9. What appears to be a "collapsed" state is not so. 2–1 and 2–6. The buccal teeth are in an ideal occlusal relationship 10–0, 10–5. and 10–8. Palatal growth maintains the excellent palatal arch relationship. The central incisors were brought together at 8 years of age prior to secondary alveolar bone grafts

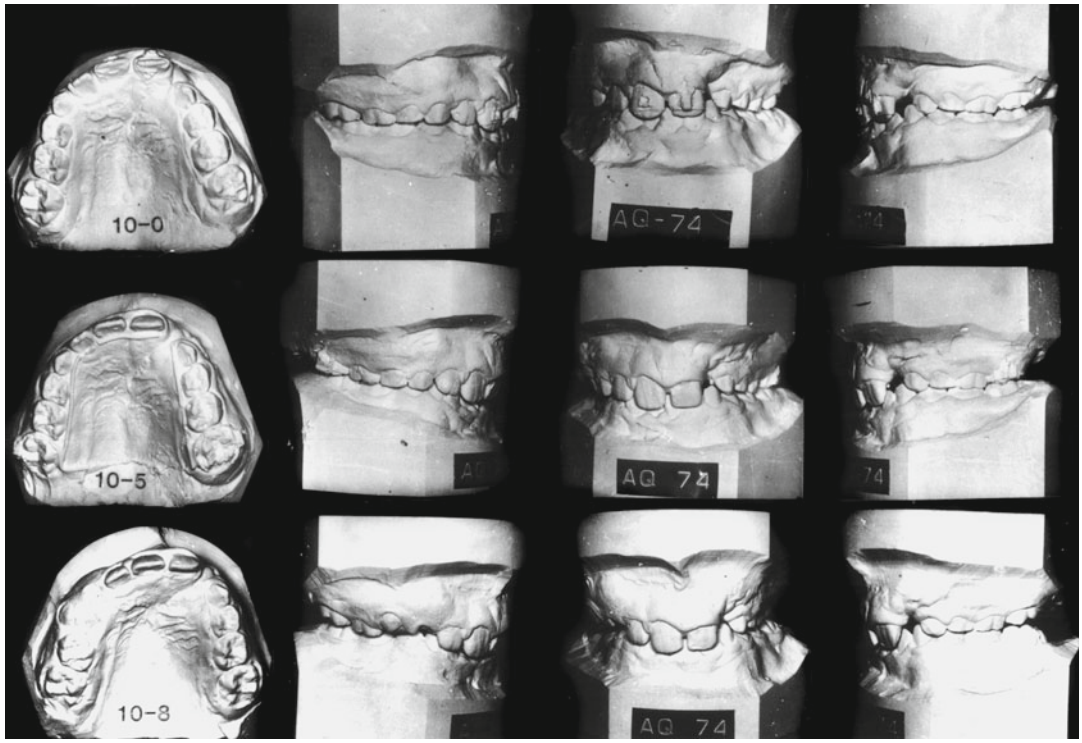

Fig. 27.3 (*continued*)

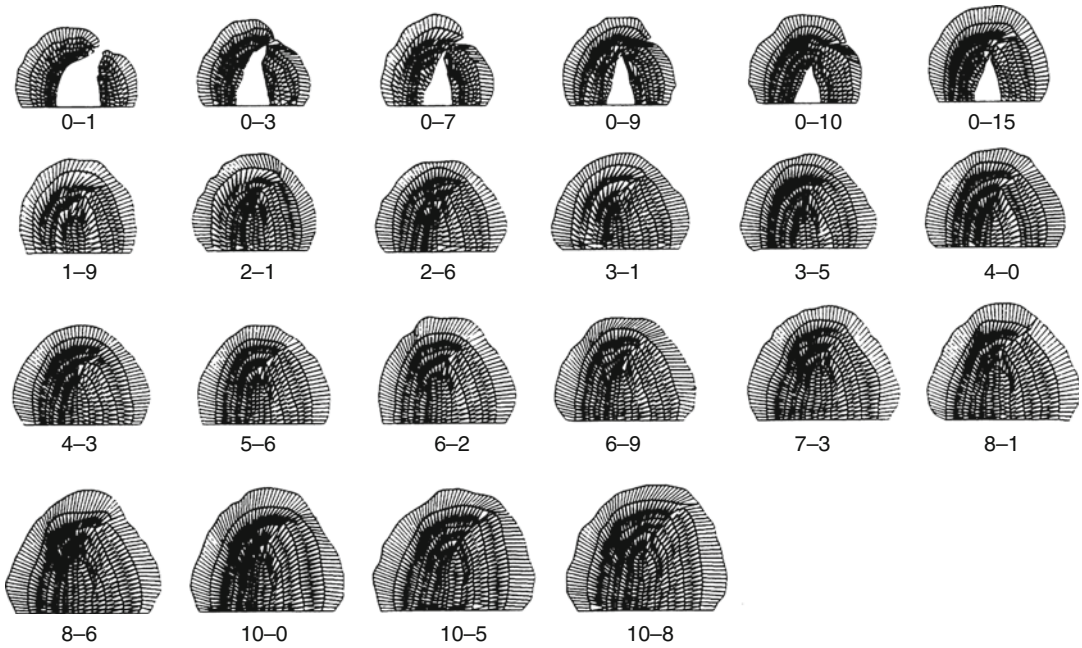

Fig. 27.4 Case JH AQ-74. Computer-generated images of serial casts drawn to scale. This series demonstrates the decrease in cleft spaces associated with an increase in palatal size

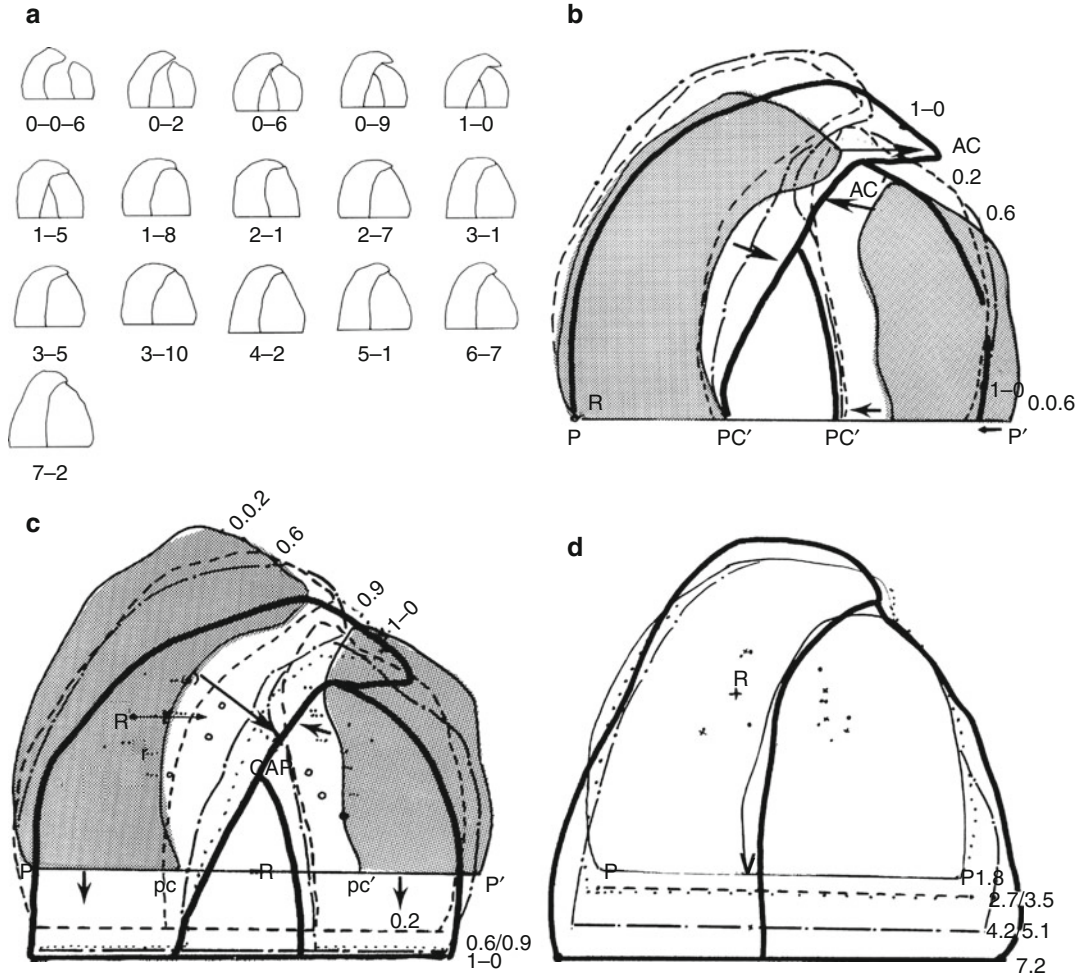

Fig. 27.5 (**a**) Computer-created serial casts drawn to scale from birth to 7 years and 2 months. (**b**) Outline tracings at 6 days, 2 months, and 1 year of age superimposed on the baseline P–P1 and registered at midpoint of the line. This illustration shows the medial movement and changes in size of the palatal segments. (**c**) The same palatal segments are superimposed on the palatal rugae to show the amount and direction of palatal growth and movement brought on by uniting the lip. From 2 days to 1 year of age. (**d**) Outline of palatal segments from 1 year and 8 months of age to 7 years and 2 months. This illustration shows that most of the palatal growth occurs posteriorly with a slight increase in width with little anterior bony apposition

medial border, effectively diminished the width of the cleft space. A second study Berkowitz et al. (1974) was undertaken to further improve the stereometric technology in order to permit the investigation of a larger number of casts. A profile study of nine complete unilateral cleft lip and palate casts demonstrated that the widths of the vault space varied greatly between cases. This was followed by another investigation using an "optical profilometer" (Berkowitz et al. 1982) designed and built by National Aeronautics and Space Administration (NASA) for Berkowitz under a technology utilization transfer grant. This led to the use of an electromechanical digitizer as the instrument of choice for analytical studies of serial casts designed to describe the changing geometry and size of the palatal vault and the geometrical and size relationship between the greater and lesser palatal segments in complete unilateral cleft lip (CUCLP) and palate and the lateral palatal segments and premaxilla in complete bilateral clefts of the lip and palate (CBCLP) (Figs. 27.5 and 27.6).

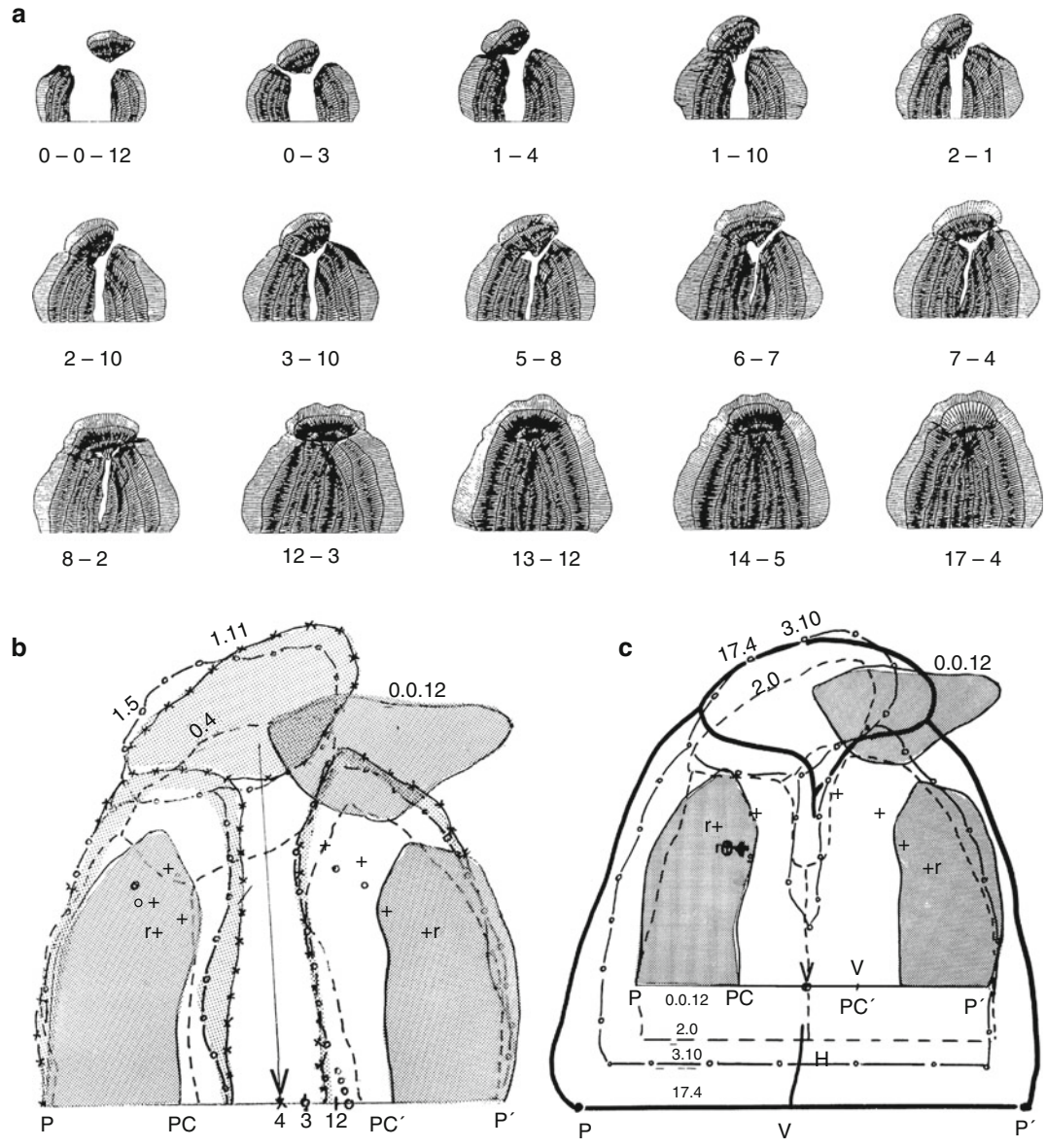

Fig. 27.6 (**a**–**d**) Case PM (K-22). (**a**) Computer-drawn serial casts of a complete bilateral cleft lip and palate drawn to scale showing quantitative and geometric palatal changes as a result of growth and treatment. No presurgical orthopedics. (**b**) Serial casts superimposed on the base line P to P, registered at V (the point where the vomer meets the baseline). This shows changes in palatal size and palatal relationships. (**c**) Serial casts from 12 days to 17 years and 4 months. These casts were superimposed on the rugae points with the PP' line parallel. Most of the palatal growth occurred posteriorly, with only a slight increase in midfacial protrusion compared to the initial premaxillary protrusion. The width increased in proportion to length

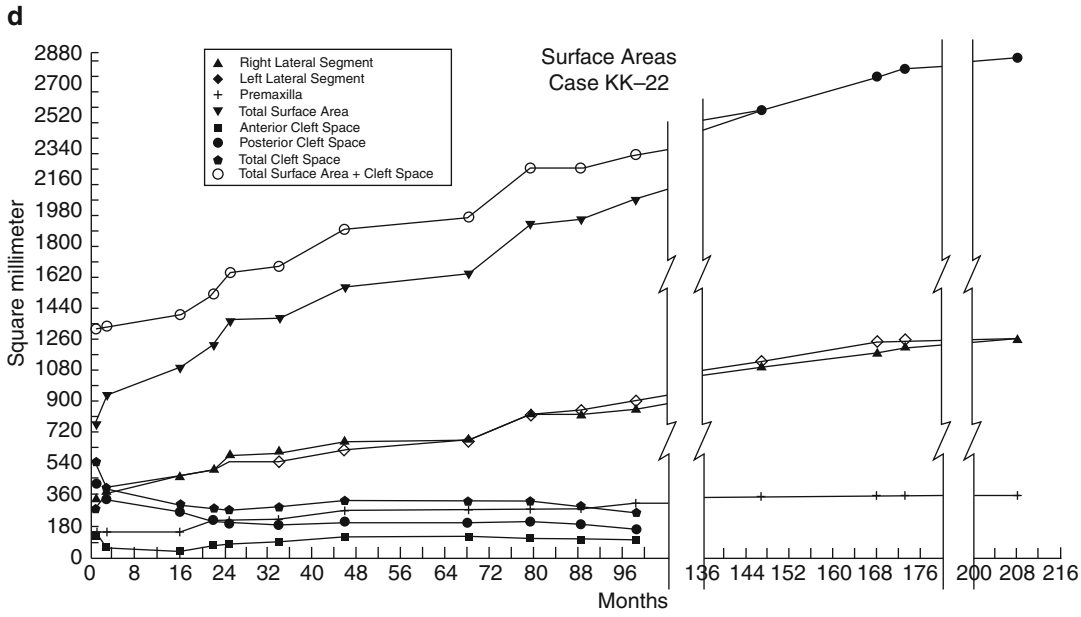

Fig. 27.6 (*continued*) (**d**) Time sequence analysis of palatal growth in a complete bilateral cleft of the lip and palate. By 6 years, the palatal growth (including cleft space) more than doubled

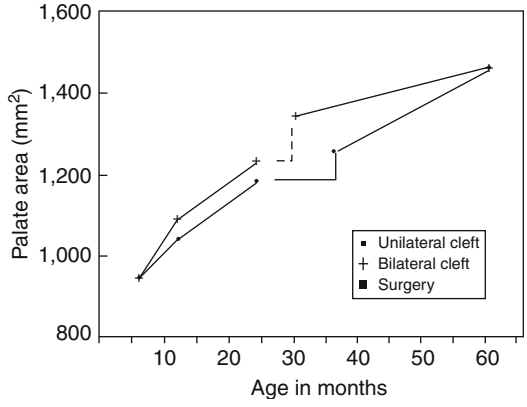

Fig. 27.7 Graph showing growth changes in surface area of 14 CBCLP and CUCLP prior to and after palatal cleft closure surgery. The palatal growth acceleration in the CBCLP was greater the first year, then similar in the two cleft types up to 25 months, just prior to palatal surgery. At the time of surgery, the cleft space in the CBCLP series was greater than in the CUCLP series. After surgery, the palatal growth acceleration curve of the CBCLP series decreased, while that of the CUCLP series increased. A modified von Langenbeck procedure to close the cleft space was used in both cleft types. The sample of cases is being increased to determine the influence of cleft size on future palatal growth and development

Serial three-dimensional palatal growth studies to date have led Berkowitz to believe that size and geometric relationship of the palatal segments relative to the size of cleft space prior to surgery, coupled with the surgical procedure utilized, may influence the palate's subsequent arch form and size (Fig. 27.7). If it does, the surgical skill or technique is not solely responsible for the different outcomes. This might explain why different surgical procedures can be equally successful and, conversely, why the same surgical procedure can cause a different result, especially if extensive scarring has been produced (Figs. 27.5 and 27.6).

The following three-dimensional palatal growth studies were recently completed. These studies can be considered forerunners of multicenter efforts still to come that will reflect on the physiological attributes of the various surgical and orthopedic treatment procedures.

27.6.1 Study 1: Analysis of Longitudinal Growth of CUCLP and CBCLP

Patients from Berkowitz's longitudinal facial–palatal growth records who did not have neonatal maxillary orthopedics were the subjects.

Eleven children with unilateral clefts and 14 children with bilateral clefts were measured for palate area in mm^2 over a period of 5 years. For the unilateral cleft group, the measurements were made at approximately 6, 12, 24, 30, and 60 months. Each child in both groups was surgically treated to close the cleft area at approximately 24–36 months.

27.6.1.1 Statistical Methods

For each child, the monthly growth rate (in mm^2/month) from 6 through 24 months was estimated by linear regression. In the unilateral cleft group, the monthly growth rate after surgical intervention was estimated by the change in palate area from 36 to 60 months. This rate was estimated in the bilateral cleft group after surgical intervention over the period of 30–60 months. Mean growth rates before and after surgical repair were compared within each group by the paired Student's t-test. Pre- and postsurgical differences in mean growth rates between the two cleft types were compared using the two-sample Student's *t*-test. In addition, growth rates and the change in growth rate before and after intervention were correlated with the estimated size of the closure at surgery.

27.6.1.2 Results

The growth rates of the two groups before surgery (unilateral = 12.9 mm^2/month, bilateral = 15.7 mm^2/month) are not significantly different. However, after surgery, the growth rate of the bilateral group (3.9 mm^2/month) is significantly smaller ($p = .013$) than that of the unilateral group (8.7 mm^2/month). Comparing pre- and postsurgery growth rates, the change in growth rate for the unilateral group (mean = 4.3, SE = 3.3) was not significantly different from zero, whereas the change of growth rate in the bilateral group (mean = 11.9, SE = 2.3) was significantly different from zero.

The estimated size of the "gap" to be surgically repaired was 80.5 mm^2 in the unilateral group and 112.5 mm^2 in the bilateral group. These cleft sizes are not significantly different; however, this may be due to the small sample size. In a larger sample size, this difference in cleft space size probably would be significant. For all subjects combined, the remaining gap at surgery was significantly negatively correlated with the presurgical growth rate ($r = 0.52$, $p = 0.034$, one-tailed). For the unilateral group alone, this correlation was not significant ($r = 0.31$, $p = 0.175$, one-tailed), while it was significant for the bilateral group ($r = 0.52$, $p = 0.029$, one-tailed).

27.6.1.3 Discussion

These findings indicate that the two cleft groups had similar growth rates prior to surgery but dissimilar cleft gap sizes at the time of surgery. Following surgery, the unilateral group showed a greater, but not statistically significant, difference in growth rate. We suspect that, if the sample size is increased, the difference in growth acceleration would be significant. The bilateral cleft group showed a significantly smaller cleft space than that of the unilateral group. Both groups started out with approximately the same mean palate surface area. Before palatal surgery, both groups had similar growth rates, with the bilateral group slowing in growth after surgery. The negative correlation of the estimated cleft space at surgery with the presurgery growth rate is a measure of the validity of the measurements. To more closely analyze the patterns of growth before and after surgery, data points spaced more closely in time, especially just before and just after surgery, are planned. These data show that growth patterns can be measured and analyzed.

27.6.2 Study 2: Quantitative Study of a Patient with CBCLP Treated with Presurgical Orthopedics

This study was undertaken to graphically demonstrate the geometric changes that occurred to a palate with a CBCLP that had undergone

presurgical orthopedic treatment by Kuijpers-Jagtman at Nijmegen Cleft Palate Center in the Netherlands. Besides the changes in palatal surface area and cleft space, all other dimensions can be quantified and compared with CUCLP and CBCLP cases that were treated differently at this and other institutions. A prospective clinical trial study is now underway. One of the goals of this research is to verify whether presurgical orthopedic treatment enhances palatal growth.

27.7 A New Instrument for 3D Facial and Palatal Cast Study

The Craniofacial Center at the University of Illinois College of Medicine has undertaken an extensive facial and palatal growth study using the latest technology.

Dr. Adriana Da Silveira, Director of Research, writes: The Vivid700 (Minolta) operates on a light-stripe triangulation range-finder principle. The subject's facial surface is scanned from top to bottom with a projected class 2 laser light stripe. The position of an illuminated surface point relative to the viewpoint is obtained by triangulation. The resolution in *x* and *y* coordinates is 200¥200 range points view per scan. The reliability of this method was tested and found to be accurate. At the Craniofacial Center, 3D images are routinely captured for initial evaluation and subsequent methods. Infants are placed in the sitting position on the parent's lap. A flat background is placed between the infant and the parent. The laser surface scanner is positioned at 1 m distant to the subject, and a series of five different scannings from different views are taken of the infant's head. Each scanning takes approximately 1 s. A custom headband is used to provide a variation in topography of the head. This allows "stitching" of the images to create a 360° three-dimensional image with the help of computer software (Polygon Editing Tools, Minolta). One facial scanning may contain 40,000 points, and a polygonal mesh is formed of all these points representing the facial surface. Cartesian coordinates (*x*, *y*, and *z*) from facial landmarks can be identified with the surface distance between them calculated using computer software (Measure, Minolta). These landmarks are standardized points used in physical anthropology, and their use in two- and three-dimensional analysis of facial shapes is well accepted. The same software can be used to construct axial images of the head and to measure the area and volume of the head and face or any other facial part such as the palate.

Serial palatal casts of palatal casts of cleft lip and palate subjects are scanned using the same equipment. The scanner is connected to a turntable that rotates every 60º, and scanning is performed at different views to allow for generation of a 360º composite representing the full cast. The images are stored in a personal computer and can be analyzed using the previously mentioned software programs. Surface distances, areas, and volumes can be calculated, and images can be superimposed for better visualization.

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Part XII

Distraction Osteogenesis

Rigid External Distraction: Its Application in Cleft Maxillary Deformities

28

John W. Polley and Alvaro A. Figueroa

Patients with severe maxillary deficiency secondary to orofacial clefting present multiple challenging problems for the reconstructive team. These patients exhibit multidimensional maxillary hypoplasia and skeletal clefting with absence of maxillary and alveolar bone, as well as scarring, residual fistulas, and dental anomalies. Traditional surgical/orthodontic approaches to treat these patients, while sometimes successful in obtaining stable occlusal relationships, often fall short of expectations with respect to facial balance and aesthetics. The application of maxillary distraction osteogenesis in the treatment protocol of patients with severe maxillofacial anomalies offers a powerful alternative for the reconstructive team.

In this chapter, we present the use of maxillary distraction osteogenesis for the treatment of patients with severe maxillary deficiency secondary to orofacial clefts. The technique of rigid external distraction, based upon the old concept of cranial fixation, has enabled rigid control over the distraction process allowing for predictable and highly successful outcomes.

28.1 Materials and Methods

All patients seen at our institution between 1 April 1995 and 1 December 1996, with severe maxillary hypoplasia (negative overjet of 8 mm or more) were considered candidates for treatment with maxillary distraction osteogenesis. Criteria for patient selection included the following: unilateral or bilateral cleft lip and palate, severe maxillary hypoplasia (vertical, horizontal, and transverse planes) with class III malocclusion, patients requiring horizontal maxillary advancement in excess of 8 mm, normal mandibular morphology and position, patients with full primary dentition or older, patients with severe palatal and pharyngeal scarring, and patients with airway obstruction and sleep apnea.

Eighteen consecutive patients were selected based on the above criteria and were treated with maxillary distraction osteogenesis. There were 10 unilateral cleft lip and palate patients, 6 bilateral cleft lip and palate patients, and 2 patients with bilateral cleft lip and palate and severe congenital facial clefting (Table 28.1). The patients' ages at the time of surgery ranged from 5.2 to 25.2 years.

Table 28.1 Number of patients by diagnosis and gender

Diagnosis	*n*	Male	Female
UCL/P	10	6	4
BCL/P	6	5	1
Facial cleft	2	1	1
Total	18	12	6

UCI/P unilateral cleft lip and palate, *BCL/P* bilateral cleft lip and palate

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All patients underwent a thorough history and clinical examination as well as a complete dental and orthodontic examination. Preoperative and postoperative photographic and cephalometric records were obtained as reported previously. Time was spent with the patient and the patient's family, explaining the distraction process in detail utilizing photographs, video imaging, as well as discussions with other patients and their families who have undergone the procedure. The patient and parents were thoroughly familiarized with the distraction apparatus and its mechanics prior to the procedure.

For this group of patients, a custom-made intraoral orthodontic splint, which acts as the link between the maxillary skeleton and distraction apparatus, was inserted in each patient (Fig. 28.1).

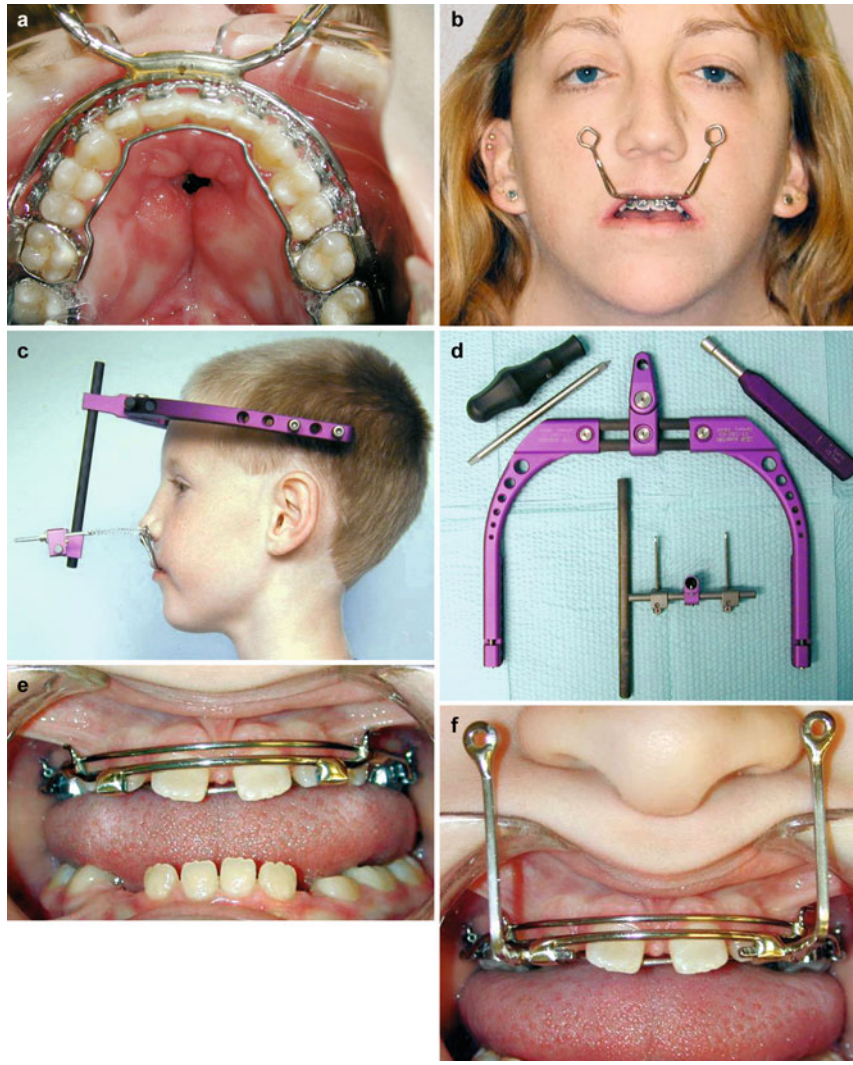

Fig. 28.1 (**a**) Intraoral view of original design made from an orthodontic face bow for the intraoral splint, anchored to the first molars and further secured with circumdental wires. (**b**) Facial view of patient wearing the device prior to surgery and demonstrating the external traction hooks. (**c**) Patient wearing the Rigid External Distraction system, note wires connecting traction hooks from the intraoral splint to the traction screws from the rigid external distraction device. (**d**) Disassembled distraction device, note posterior screws on the halo, utilized during midfacial advancement to clear the anterior part of the halo from forehead. (**e**) Intraoral view of newly designed intraoral splint with removable external distraction hooks. Designed in this fashion to avoid the presence of the hooks at surgery. (**f**) Intraoral splint with removable distraction hooks in place

This intraoral splint was constructed with rigid 0.045 or 0.050 stainless steel orthodontic wire. The splint was cemented to the first permanent molars or second primary molars and was further secured at the time of surgery with circumdental stainless steel surgical wire. For older children and adults, the splint was inserted in the clinic prior to the surgical procedure. For young children, the splint was placed in the operating room after anesthetic induction and prior to the osteotomy. External traction hooks were bent under and in front of the upper lip with the end of the hook ending at the level of the palatal plane. The splint also included intraoral hooks that were utilized for the required retention phase at the completion of active maxillary distraction. Certain orthodontic tooth movements as well as orthodontic expansion of the upper arch can be performed at the time of maxillary distraction by incorporation of an expansion screw and segmentalizing the intraoral splint into two units. In order to improve rigidity, the splint can be manufactured by using an orthodontic cervical face bow. The external bow is bent to create the external traction hooks. Currently, the intraoral part of the splint utilized at our center is made of heavy 0.045 or 0.050 stainless steel orthodontic wire. In the anterior region, two square tubes are soldered just medial to the oral commissures (Fig. 28.1e). These tubes are used to house removable traction hooks made of heavy rectangular wire (Fig. 28.1f). This design offers the advantage of not having the external traction hooks present at the time of surgery facilitating anesthesia management and intraoral surgical manipulation.

All 18 patients in this series underwent a LeFort I osteotomy with the height of the transverse osteotomy varying depending on the dental age and aesthetic requirements of each patient. In patients with transitional dentition, a high transverse maxillary osteotomy was performed, just below the level of the inferior orbital rim. The osteotomy circumvents the infraorbital foramen to prevent injury to the infraorbital neurovascular bundle and avoids all permanent tooth buds. Mobilization of the maxillary segments is achieved, including pterygomaxillary and septal disjunction. No intraoperative repositioning of the maxillary segments is performed, and no autogenous or alloplastic bone grafting or internal skeletal fixation hardware is utilized. For the patients in this series who underwent rigid external distraction, the halo portion of the distraction device was placed immediately after closure of the intraoral incision. In the first four consecutive patients with severe cleft maxillary hypoplasia that we elected to treat with maxillary distraction, we performed the distraction with elastic traction as described below. After four consecutive treatment failures in these patients, we abandoned elastic distraction for rigid external distraction.

Fourteen patients underwent rigid external maxillary distraction (Fig. 28.1, Table 28.2) (Polley and Figueroa 1997). These patients had the rigid device placed at the time of surgery and had their distraction performed through mechanical activation of the distraction device. Four patients in this series underwent face mask elastic distraction osteogenesis (Table 28.2). An orthodontic face mask was utilized with elastics placed on the intraoral hooks. Up to 2 lb of elastic force was utilized for each patient through a combination of 8-oz elastics. Patients were instructed to wear the face mask 24 h a day for a period of 3 months. All patients underwent a latency period of 4–5 days following the osteotomy and then began distraction. For the rigid external distraction group, distraction was performed at the rate of 1 mm/day. In this group, the distraction device was kept in place 3–4 weeks following completion of the distraction process for rigid retention. An additional 4–6 weeks of face mask elastic retention at night time only was utilized in this group. For the rigid external distraction group following the period of rigid retention, the external distraction device was removed in an office setting. For the very young patients in this series, the distraction device was removed with mild sedation.

Table 28.2 Treatment groups by diagnosis

Distraction	UCL/P	BCL/P	BCL/P + FC	Total
Rigid external	9	3	2	14
Face mask	3	1	–	4

UCL/P unilateral cleft lip and palate, *BCL/P* bilateral cleft lip and palate, *FC* facial cleft

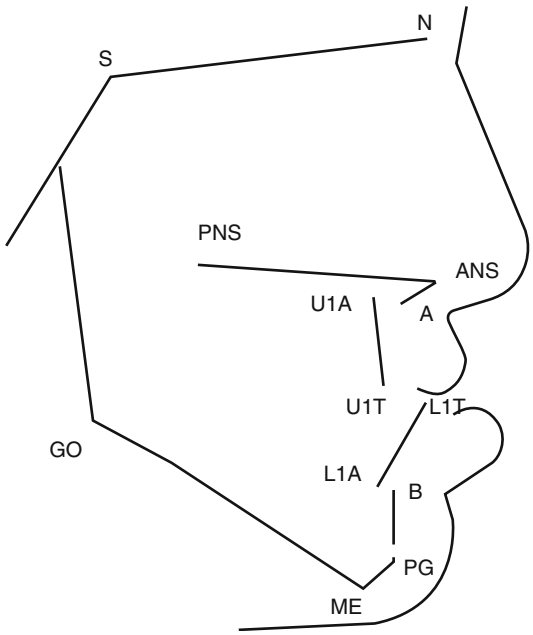

Fig. 28.2 Anatomic landmarks and reference planes. Anatomic landmarks: sella (*S*), center of sella turcica; nasion (*N*), most anterior point of the nasal frontal suture; anterior nasal spine (*ANS*), most anterior point of the spine; A point (*A*), most anterior limit of the maxillary alveolar bone at the level of the incisor root apex; posterior nasal spine (*PNS*), intersection between the nasal floor and the posterior contour of the maxilla; apex of maxillary incisor root (*U1A*), uppermost point of the incisor root; tip of maxillary incisor crown (*U1T*), maxillary incisor edge; tip of mandibular incisor (*L1T*), mandibular incisor edge; apex of mandibular incisor root (*L1A*), lowermost point of the mandibular incisor root; B point (*B*), most anterior limit of the mandibular alveolar bone at the level of the incisor root apex; pogonion (*PG*), most anterior limit of the mandibular symphysis; menton (*ME*), most anterior point of the mandibular symphysis; gonion (*GO*), the point at the greater convexity of the mandibular gonial region. Reference planes: sella-nasion plane (*SN*), palatal plane (line through *ANS* and *PNS*), maxillary incisor axis (*Ul*) (line passing through *U1A* and *UIT*); and mandibular plane (tangent to the lower border of the mandible through *ME* and *GO*)

28.2 Cephalometric Evaluation

The preoperative and postretention lateral cephalometric radiographs were utilized for analysis. The radiographs were traced on 0.003-in acetate paper, and 12 anatomic landmarks were recorded (Fig. 28.2). All tracings were done by a single experienced investigator (A. A. Figueroa). Availability of serial radiographs in all patients permitted landmark verification. All x-rays were corrected to 0 % magnification. Based on the recorded anatomic landmarks, 13 measurements were calculated, 6 angular and 7 linear (4 horizontal and 3 vertical). For the linear measurements, an x-y coordinate system utilizing the S-N plane as the horizontal was employed. Linear horizontal changes were measured relative to a line perpendicular to the S-N plane passing through sella, and vertical changes were measured perpendicular to the S-N plane. The preoperative and postoperative cephalometric values in the rigid external distraction group were statistically analyzed by means of a paired t-test. The numbers in the face mask distraction group were too small for meaningful statistical analysis for intragroup and intergroup comparisons.

28.3 Results

Surgery in this series was performed on either an outpatient or a 23-h admission basis by a single experienced surgeon (Polley). No patient in this series was discharged later than the morning after surgery. Perioperative antibiotics were routinely used. All patients began routine oral hygiene and an unrestricted soft diet 24 h postoperatively. No intermaxillary fixation was utilized.

There was no surgical morbidity in any of the 18 patients in this series. There were no complications of bleeding or infection. No patients required a blood transfusion, and there were no problems of dental injury, avascular necrosis, or gingival injury. In the patients who underwent rigid external distraction, there were no complications with wearing the external device including pain, discomfort, or loosening of the device during the distraction process. The intraoral splint remained intact in all patients through the active and retention phases. None of the families had difficulty following the guidelines for the distraction, which they carried out at home.

The postdistraction cephalometric radiographs for all patients who underwent either

Table 28.3 Predistraction and postdistraction angular cephalometric measurements for rigid external and face mask distraction groups

Distraction	Measurements (degrees)	Predistraction	Postdistraction	Difference (4 months)		Significance
Rigid external ($n=14$)	SNA	77.6	±5.6	85.3±5.6	7.7±2.9	**
	SNB	78.8	±4.0	77.9±4.1	–0.8±1.8	NS
	ANB	–1.2	±3.5	7.3±3.0	8.6±3.6	**
	Convexity (NAPg)	–3.5	±7.5	13.7±6.0	17.2±7.3	**
	Mand. Pl./SN angle	39.2	±6.7	41.4±5.9	2.2±2.4	*
	Ul-P.PL. angle	100.7	±15.7	98.8±14.4	–1.2±11.3	NS
Face mask ($n=4$)	SNA	77.5	±4.3	80.3±5.5	2.8±4.9	
	SNB	82.9	±4.3	81.3±3.3	–1.6±3.8	
	ANB	–5.4	±2	–1.0±3.4	4.4±2.7	
	Convexity (NAPg)	–12.2	±3.5	–3.4±7.4	8.8±5.5	
	Mand. P1./SN angle	33.2	±4.9	36.5±3.6	3.3±4.3	
	Ul-P.PL. angle	105.1	±13.1	107.3±10.4	2.2±11.8	

*$p<0.01$
**$p<0.001$

Table 28.4 Predistraction and postdistraction linear cephalometric measurements for rigid external and face mask distraction groups

Landmark (axis)	Pre-post distraction change rigid external (mm)	Pre-post distraction change face mask (mm)
ANS-x	7.1±3.9*	2.9±1.4
ANS-y	–0.4±3.0	–1.3±1.8
A point-x	8.3±3.3*	2.8±2.1
A point-y	–1.3±3.4	–1.7±2.3
U1-x	11.6±4.6*	5.2±2.1
U1-Y	–1.8±3.5	–1.8±1.2
Overjet	12.7±3.0*	7.2±2.0

*$p<0.001$

rigid external or face mask distraction were obtained 4 months following distraction. The predistraction and postdistraction angular cephalometric measurements for those patients undergoing rigid external and face mask distraction are given in (Table 28.3). The differences in the linear measurements between the predistraction and postdistraction cephalometric radiographs for each group are given in (Table 28.4). In all of the patients who underwent rigid external distraction, the desired treatment goals were obtained. All patients in the face mask distraction group were undercorrected with residual edge to edge anterior dental relations or remained in anterior crossbite. The results in all patients in the face mask distraction group were considered unsuccessful.

28.3.1 Angular Changes

For patients undergoing rigid external distraction, the average predistraction S-N-A angle was 77.6°, and the postdistraction S-N-A angle was 85.3°, for an average increase in this group of 7.7° (Table 28.3). The average predistraction A-N-B was –1.2°, and postdistraction was 7.3° with an increase of 8.6°. For all patients who underwent rigid external distraction, the skeletal angle of convexity increased postdistraction by 17.2°. All of these three measurements were statistically significant.

In the elastic distraction group, the average predistraction S-N-A angle was 77.5°, and postdistraction was 80.3° with only an increase of 2.8°. The A-N-B angle was –5.4° predistraction, and postdistraction averaged –1.0° for an increase of

4.4°. The average change in the skeletal angle of convexity postdistraction in the face mask group was 8.8°. The mandibular plane angle changed 2.2° in the rigid external distraction group and 3.3° in the face mask distraction group.

The angular changes in the rigid distraction group were more than double those in the face mask elastic distraction group.

28.3.2 Linear Changes

The A-N-S change between predistraction and postdistraction cephalometric radiographs in the rigid external distraction group was 7.1 mm, and in the face mask distraction group, it was only 2.9 mm (Table 28.4).

In the rigid external group, the average horizontal advancement of the A point following distraction was 8.3 mm. In the elastic distraction group, the A point advancement was only 2.8 mm. The horizontal advancement at the upper incisal edge averaged 11.6 mm for the patients who underwent rigid external distraction (Table 28.4) and only 5.2 mm for those patients in the face mask elastic distraction group. Patients in the rigid external group had a positive correction of their overjet by 12.7 mm as compared with 7.2 mm in those patients who underwent face mask elastic distraction. All of the changes in the horizontal linear measurements in the rigid external group were highly significant between predistraction and postdistraction measurements at a p value of 0.001.

28.3.3 Dental Changes

The dental changes in both groups between predistraction and postdistraction cephalograms are also given in (Tables 28.3 and 28.4). In the rigid external distraction group, the change in the angle of the upper incisor edge to the palatal plane averaged –1.2° for all patients. This was not statistically significant. In the face mask distraction group, the angular change in the upper incisor edge to palatal plane measurements was 2.2°. In none of the patients in this series, including both rigid external distraction and face mask groups, were spaces created posterior to the most distal point of anchorage of the intraoral splint.

28.4 Discussion

Maxillary hypoplasia is a common finding in patients with orofacial clefting. It has been estimated that 25–50 % of all patients born with complete unilateral cleft lip and palate will be potential candidates for maxillary advancement to correct the functional deformities and improve aesthetic facial proportions (Ross 1987). In addition, it is recognized that the majority of patients with facial clefts have morphologically normal or slightly smaller than normal mandibles (Da Silva Filho et al. 1993; Semb 1991). Patients with severe cleft maxillary deficiency are difficult patients to treat with standard surgical/orthodontic approaches. These patients present with maxillary hypoplasia (vertical, horizontal, and transverse dimensions) and often thin or structurally weak bone. The maxillary hypoplasia in cleft patients is also compounded by residual palatal and alveolar fistulas, absent and aberrant dentition, and scarring of the palatal and pharyngeal soft tissues.

The physical deformities exhibited by patients with severe maxillary hypoplasia contribute to multiple functional deficiencies as well. These include severe malocclusions, which result in compromised mastication, speech abnormalities, and nasal pharyngeal airway constriction (Witzel and Vallino 1992). The severe dish-face or concave facial profiles in these patients result in highly detrimental psychosocial ramifications as well (Kapp-Simon 1996). Current protocols for the treatment of maxillary hypoplasia in cleft patients rely upon a surgical/orthodontic approach, including a LeFort I maxillary advancement with concomitant fistula closure and maxillary and alveolar bone grafting. This surgery includes rigid internal fixation hardware for stabilization of the repositioned maxilla in the postoperative period. The long-term results of cleft patients with maxillary deficiency treated in such fashion have been reported by several authors (Erbe et al. 1996; Cheung et al. 1994;

Table 28.5 Review of long-term results of sagittal maxillary relapse in cleft patients following maxillary advancement with fistula closure, bone grafting, and rigid internal fixation

Author	Erbe et al. (1996)	Cheung et al. (1994)	Posnick and Dagys (1994)	Hochban et al. (1993)	Eskenazi and Schendel (1992)	Polley and Figueroa (1997)
N cleft Pt.	11	46	35	14	12	14
Mean follow-up (months)	59	28	12	12	12	4
Mean maximum advancement	4.6	4.5	6.9	8	7.8	11.6
Mean relapse	40 %	22 %	21 %	25 %	4 %	–

Posnick and Dagys 1994; Hochban et al. 1993; Eskenazi and Schendel 1992). The mean horizontal maxillary advancement in these reported series has averaged between 5 and 7 mm, and the mean long-term horizontal relapse averages between 20 and 25 % (Table 28.5). Erbe et al. (1996) presented a mean follow-up of 59 months for 11 cleft patients who underwent segmental osteotomies with maxillary advancement and simultaneous fistula closure with rigid internal fixation and bone grafting. On average, the greater maxillary segment in these patients was advanced 3.9 mm and the lesser maxillary segment 5.3 mm. At nearly 5 years postoperatively, they found that the horizontal relapse of the maxilla was approximately 40 %. Cheung et al. (1994) reported a consecutive series of 46 cleft patients who underwent maxillary advancement with rigid internal fixation and simultaneous alveolar bone grafting with fistula closure. The mean horizontal maxillary advancement in this series was 4.5 mm. Relapse in the horizontal plane for the unilateral cleft patients in this series, at a mean of 28 months postoperatively, was 22 %. Similar horizontal relapse rates following maxillary advancement with rigid internal fixation in cleft patients have been reported by others as well (Table 28.5) (Posnick and Dagys 1994; Hochban et al. 1993; Eskenazi and Schendel 1992).

In this series, 14 consecutive patients underwent successful maxillary advancement at the LeFort I level through maxillary distraction with rigid external distraction. Examples of our clinical experience with rigid external distraction are illustrated in (Figs. 28.3, 28.4, and 28.5). No rigid internal fixation hardware or autogenous bone grafting was utilized at the time of the osteotomy. The patients underwent distraction at the rate of 1 mm per day followed by a 3–4-week rigid retention period. In this group of 14 cleft patients, the mean effective maxillary advancement was 11.7 mm. The time of the postdistraction cephalometric radiographs analyzed in this article averaged 4 months following completion of the distraction process. These data have been presented in this fashion so that the immediate effects of the distraction process can be measured accurately (Fig. 28.6). Many of our patients now have a follow-up of more than 12 months, and we have not seen clinical evidence of relapse in any of the patients to date (Fig. 28.7). In the immature patients, maxillofacial growth will continue, and we are currently following the development of these patients.

In the past, it has been difficult to consistently and successfully treat patients with severe maxillary deficiency with maxillary advancement alone. Patients with large anteroposterior maxillomandibular discrepancies may require mandibular setback surgery in addition to maxillary advancement for correction of their severe horizontal deficiency. With the use of rigid external distraction, we can now gradually and in a very stable fashion reposition a severely hypoplastic maxilla to the exact horizontal and vertical position desired. The patients create their own autogenous bone during this process, eliminating the need for a donor site as well as eliminating the need for rigid internal fixation hardware. The use of rigid external distraction has allowed rigid

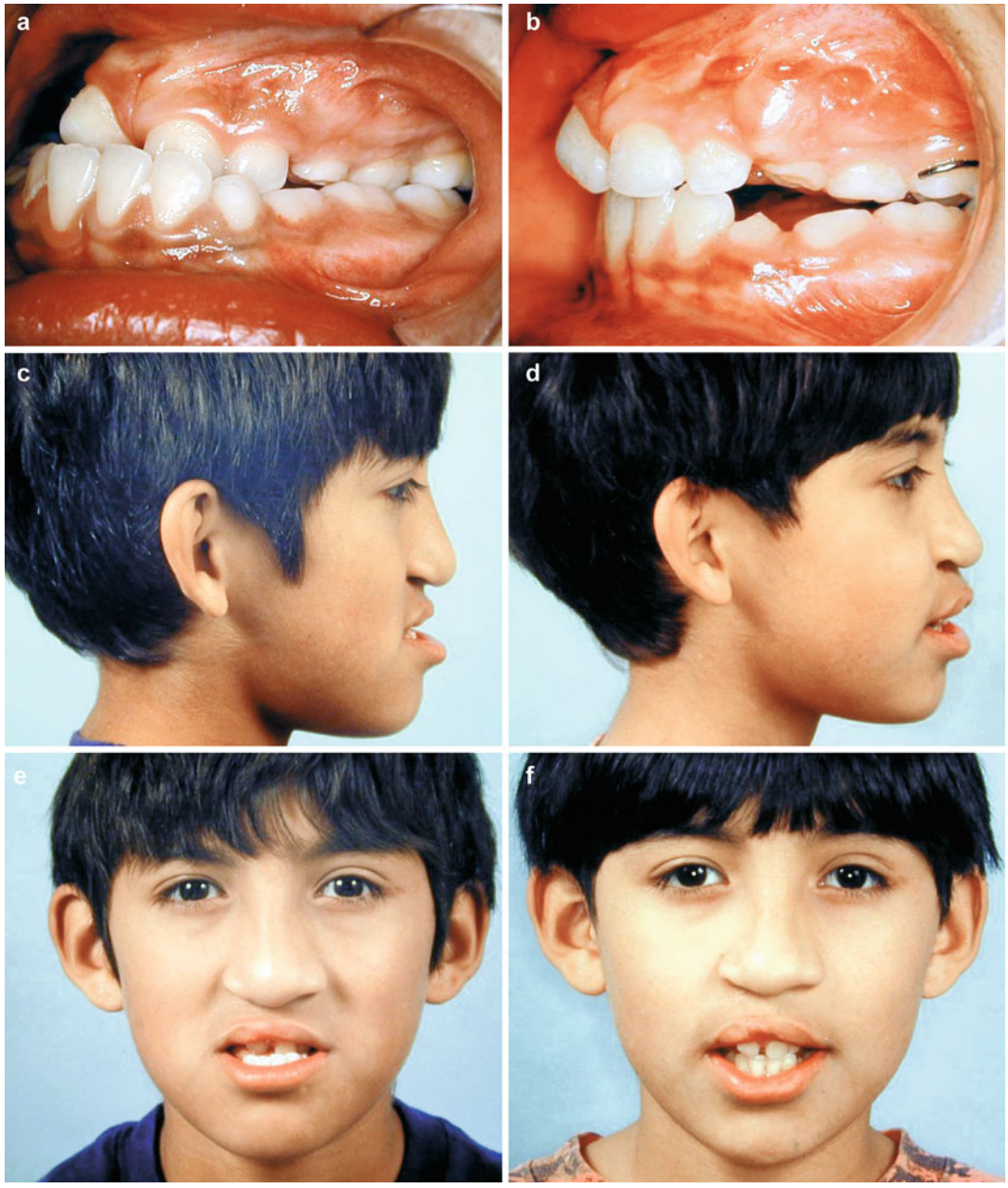

Fig. 28.3 (**a**–**f**) Nine-year-old boy with right unilateral cleft lip and palate with severe maxillary hypoplasia and crossbites. Predistraction (*left*) and postdistraction (*right*) facial and intraoral views demonstrate correction of midface deficiency with improved facial proportions and dental relations

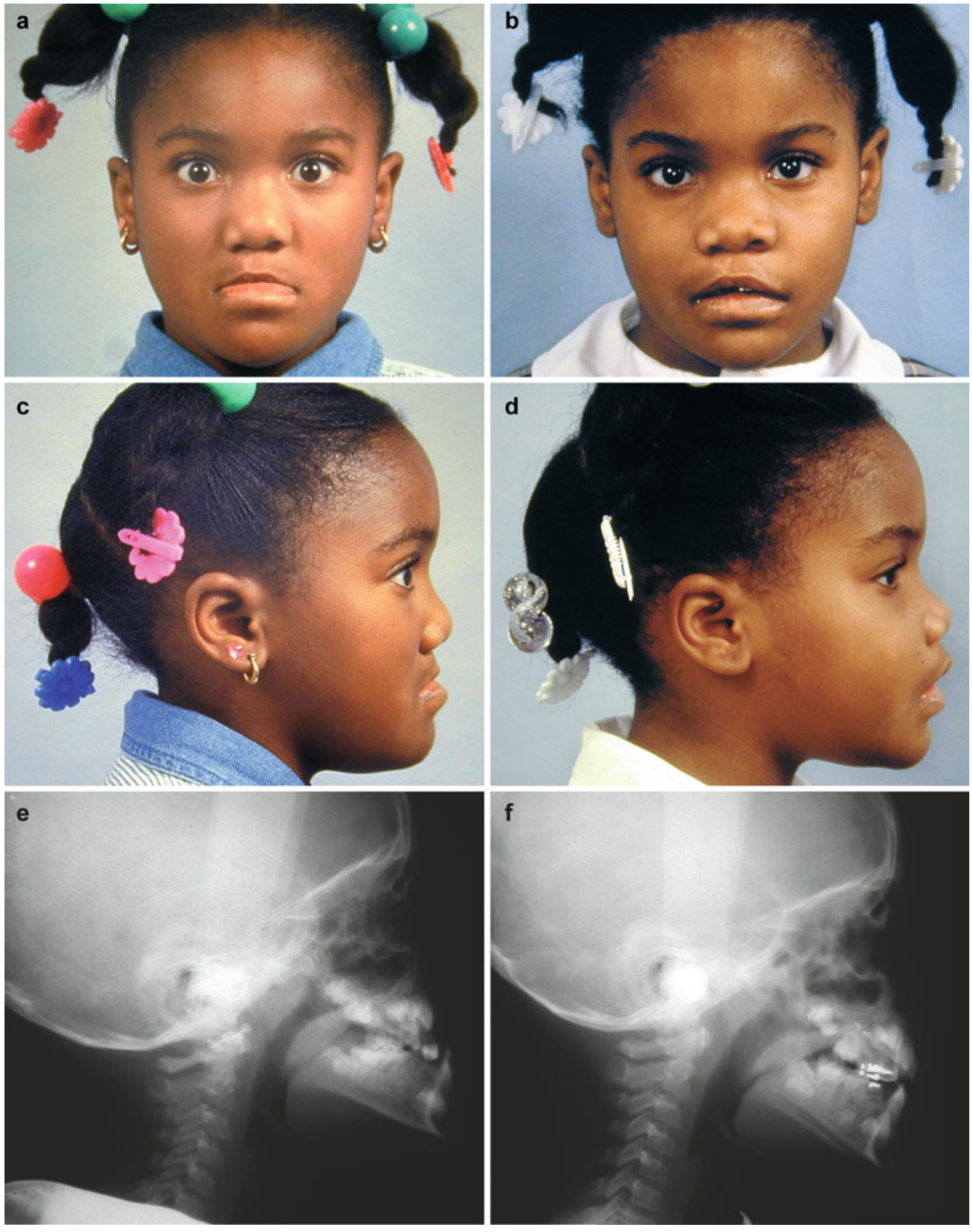

Fig. 28.4 (**a**–**f**) Six-year-old girl with left unilateral cleft lip and palate and severe maxillary hypoplasia. Predistraction (*left*) and postdistraction (*right*) facial and cephalometric radiograph views demonstrate correction of facial concavity and crossbite

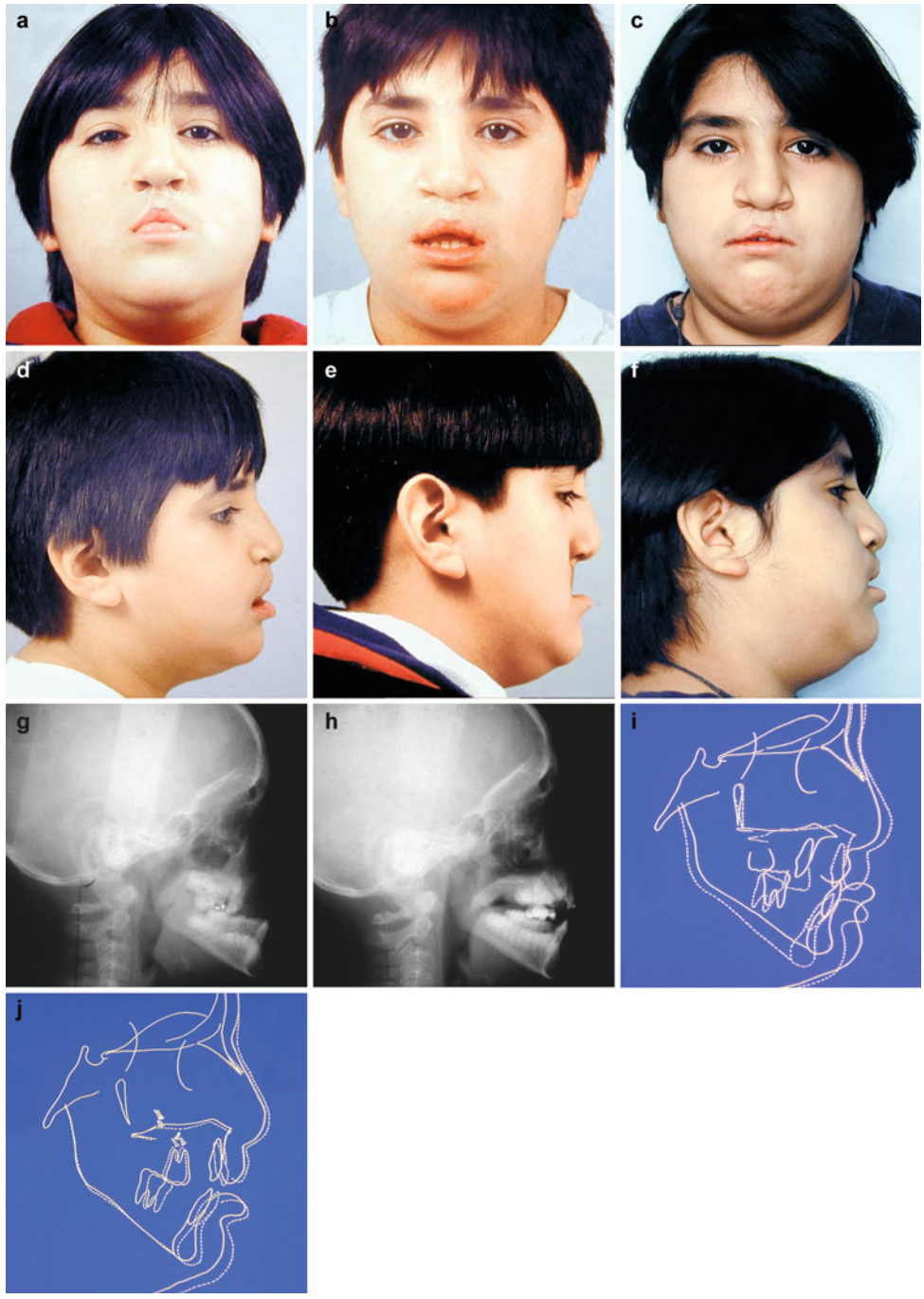

Fig. 28.5 (**a**–**j**) Ten-year-old boy with repaired bilateral cleft lip and palate with extremely severe maxillary deficiency. His predistraction 1 and 3 years postdistraction facial views demonstrate the dramatic improvement in facial profile, balance, and long-term stability. The pre- and postdistraction cephalometric radiographs demonstrate the marked maxillary advancement, increase in airway size, correction of anterior crossbite and class III skeletal relations. The pre- and postdistraction tracings (**i**, **j**) demonstrate the degree of maxillary advancement and the skeletal and soft tissue changes obtained after the distraction procedure. The postdistraction tracings demonstrate stability of the maxillary position and continued mandibular growth. Change in incisor position is secondary to orthodontic treatment

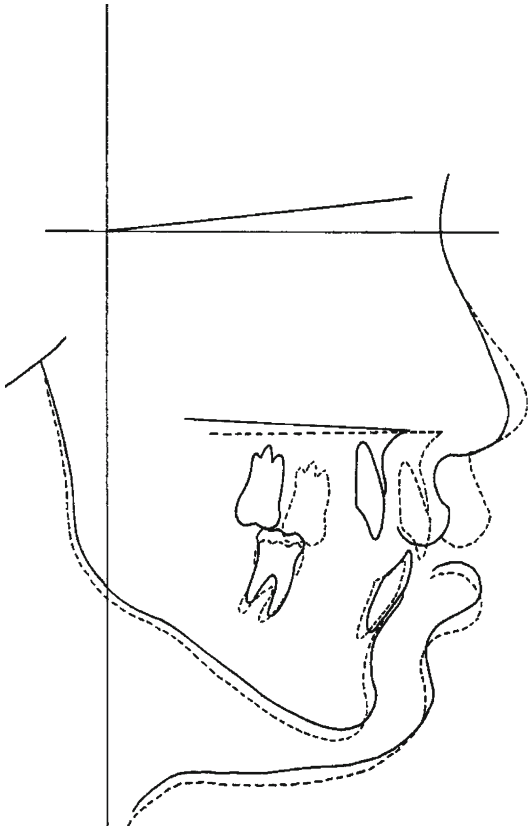

Fig. 28.6 Predistraction (*solid line*) and postdistraction (*broken line*) average tracings for rigid external distraction group. Note significant maxillary advancement (effective incisor advancement 11.6 mm) with correction of overjet, improvement of skeletal convexity, and minimal changes in mandibular position. Note significant soft tissue changes including the significant degree of lip and nasal tip advancement

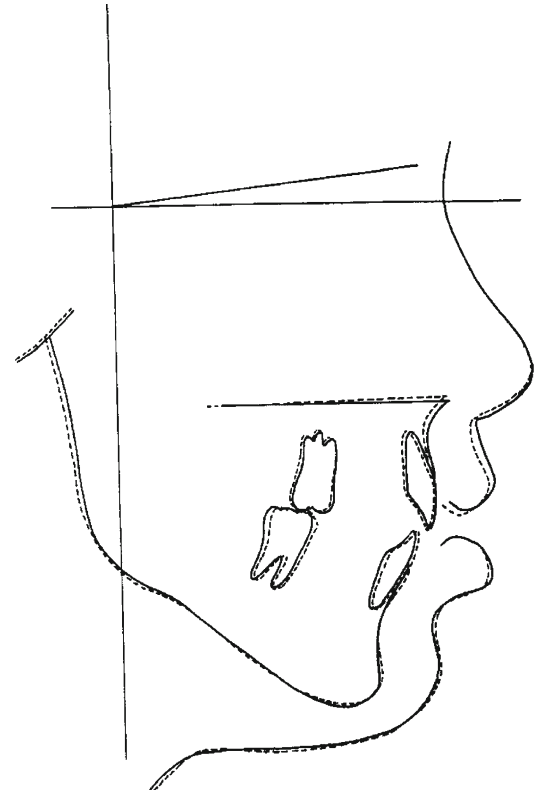

Fig. 28.7 Postdistraction (*solid line*) and 1 year (*broken line*) after distraction average tracings of the cleft sample treated with the Rigid External Distraction system. Note excellent stability of the advanced maxilla

control over the distraction process and has enabled us to follow our surgical and aesthetic guidelines for the reconstruction of these patients by correcting the entire maxillary skeletal and soft tissue discrepancy in the region of the hypoplasia only. This expansion of the soft tissue facial mask yields the most pleasing long-term aesthetic facial balance and harmony (Rosen 1992a, b), particularly in cleft patients.

In this initial consecutive series of patients, we utilized two different techniques for maxillary distraction. One group (face mask distraction) underwent maxillary distraction with the use of an orthodontic face mask and elastic traction (Molina and Ortiz-Monasterio 1996). With the use of the elastic distraction, we were able to obtain only partial correction of the horizontal maxillary deficiencies in these patients. We considered all of our face mask patients to have failed their surgical treatment. We therefore changed the design of the distraction technique to rigid external distraction, which utilizes a skeletally (cranial) fixed distraction device that allows for rigid, predictable control over the distraction process. This device is readily adjustable, offering the ability to change the vertical and horizontal vector of distraction at any time during the distraction process. The rigid control over the distraction process with rigid external distraction is evidenced by the greater maxillary movements obtained with this device as compared with those patients who underwent face mask elastic distraction. For the patients who underwent rigid external distraction, the mean effective horizontal

Table 28.6 Review of maximum-maxillary sagittal advancement in cleft patients undergoing maxillary distraction with face mask elastic traction

Measurements	Rigid external	Face mask	Face mask	Face mask
	Chicago	Chicago	M. C. Rambam, [15] Haifa	C. H. A. Trousseau, [16, 17] Paris
A point advancement (mm)	8.3	2.8	4.5	3
ANB (degrees)	8.6	4.4	4.0	–
Convexity (degrees)	17.2	8.8	5.0	–

maxillary movement at the upper incisal edge averaged 11.7 mm. This mean horizontal effective maxillary movement in the face mask elastic traction group was only 5.3 mm. Similarly in the rigid external distraction group, the overjet correction for each patient averaged 12.7 mm versus 7.2 mm for the elastic traction group. The preoperative and postoperative angular cephalometric measurements for each group showed similar results. The average change in the S-N-A angle in the patients who underwent rigid external distraction was 7.7° versus 2.8° in the face mask elastic distraction group. The change in the skeletal angle of convexity in the group who underwent rigid external distraction averaged 17.2° versus 8.8° in the patients who underwent distraction utilizing elastics.

Similar findings in the amount of skeletal maxillary movement with the use of face mask elastic distraction have been reported by others (Table 28.6) (Rachmiel et al. 1997; Diner et al. 1997; Hung et al. 1997). Rachmiel et al. (1997) reported that in their series of 12 patients with cleft maxillary deficiency, 30 % of the patients were unresponsive to maxillary distraction techniques with face mask elastics. In the 70 % of patients in whom they were able to obtain maxillary advancement, the cephalometric findings are similar to our findings in the amount of maxillary skeletal change seen with face mask elastic traction. Rachmiel et al. (1997) report the amount of maxillary advancement at the A point in their series to be 4.5 mm, and in our series with face mask elastic traction the amount of movement at the A point was 2.8 mm. A point advancement in our series of patients with rigid external distraction averaged 8.3 mm. The change in the skeletal angle of convexity in the (Rachmiel et al. 1997) series averaged 5.0° compared with a change of 8.8° in our series of face mask elastic traction and 17.2° in our series of rigid external distraction. Diner et al. (1997) obtained only 3 mm of horizontal maxillary advancement at A point in their series of patients who underwent maxillary distraction osteogenesis with use of a face mask and elastic traction. The experience of Hung et al. (1997) was similar with the use of elastic forces for maxillary advancement in cleft patients. In their series, they found the mean horizontal maxillary advancement to be only 5.2 mm. Maxillary distraction at the LeFort I level with the use of a face mask and elastic traction is unpredictable and unreliable, allowing for horizontal advancement in the 4- to 6-mm range only. In our practice, patients who require only 4–6 mm of maxillary advancement are not routinely considered for treatment with distraction osteogenesis. In these patients, standard orthodontic surgical approaches at the appropriate age are generally performed.

Internal distraction devices have also been reported for use in midface advancement. Most of these reports have focused on advancements of the midface at the LeFort III level or above (Cohen and Burstein 1997; Chin and Toth 1997). Some, however, have advocated the use of an internal device for maxillary advancement at the LeFort I level (Cohen and Burstein 1997). To date, there is no evidence that this technique affords sufficient distraction at the LeFort I level. Internal devices may require multiple surgical approaches for their placement, a second operative procedure for their removal, and the necessity for an exit port for the activating arm of the device. The vectors of distraction with the use of internal devices will be limited by the placement of each device as well as its finite mechanics. Perhaps the greatest disadvantage of internal devices is in the design of the transverse maxillary osteotomy. With internal distraction

devices, the osteotomy must be designed so that there is enough stable bone above and below the osteotomy line to allow appropriate placement and fixation of the distraction hardware. This is not required with rigid external distraction. The osteotomy design with this device is based upon the aesthetic requirements of each individual patient and not based upon the placement of internal hardware. This allows the transverse maxillary osteotomy to be carried as high as indicated along the pyriform aperture as well as in the malar regions, allowing for maximal correction of the patient's preoperative facial concavity. Maxillary advancement with rigid external distraction allows total versatility and flexibility in both the amount and direction of the distraction process. The vectors of distraction can be and are changed at any time during the distraction process. In addition, no additional surgical procedure is required for removal of the rigid external device.

The use of the cranium as an anchorage point for the stabilization of maxillofacial surgery is not a new concept. Cranial stabilization devices for maxillofacial trauma reconstruction as well as for elective maxillofacial surgery have been reported with good success in the past (Stoelinga et al. 1987; Houston et al. 1989). In addition, our neurosurgical colleagues have used the cranium as a solid fixation point for the stabilization of cervical injuries and reconstructions for decades. The scalp pins (2–3 per side), which stabilize the halo component of the rigid external distraction device, are discomfort free. Not even the youngest patients have any complaints or problems with wearing of the device throughout the distraction process. No special scalp pin care is required, and the use of ointments and creams at the scalp pin interface is discouraged. The patients simply shampoo and wash their hair in the shower with the device in place in a normal fashion. Currently, the device is readily removed without even local anesthesia in the clinic following the phase of rigid retention. In young apprehensive patients, the halo is removed with mild sedation. No secondary surgical procedure is required for removal of internal hardware.

One of the great advantages of distraction osteogenesis in the craniomaxillofacial skeleton is that, in theory, there is no limitation to the age at which patients can be treated. Contemporary surgical orthodontic approaches for the treatment of maxillary deficiency in cleft patients are dependent upon the patient having reached skeletal maturity before the reconstructive surgery can be performed. Treating patients in the transitional dentition stage with the use of autogenous bone grafting and rigid internal fixation plates and screws is technically difficult to perform without injury to the developing permanent tooth buds. The technique of rigid external distraction eliminates the negative technical factors associated with traditional orthognathic surgery in patients in transitional dental development. With maxillary rigid external distraction, only an osteotomy is performed. Repositioning skeletal segments, internal fixation hardware, and bone grafting are not required. The only limitations with rigid external distraction include adequate dentition, either primary or secondary, for fixation of the intraoral splint, as well as the ability of the patient to wear the device. Rigid external distraction now allows treatment of these patients from the age of 2 years and up. In special circumstances in patients with craniosynostosis (Crouzon and Apert syndromes), we have used the technique for monobloc advancement as early as 18 months of age. In these cases, the second or even the first primary maxillary molar has been used to support the intraoral splint. In addition, special plates have been developed to connect the edentulous maxilla to the rigid external distraction device (Hierl and Hemprich 1999).

This article reports our preliminary experience with this technique. Three-year follow-up of this group of patients has demonstrated outstanding stability in maxillary position (Figueroa et al. 2004). However, we caution all young patients undergoing rigid external distraction that they may require a final "finishing" LeFort I procedure at skeletal maturity for final arch alignment.

The advantages of maxillary rigid external distraction are numerous. This technique allows an excellent modality for correcting the severe

midface concavities, at any age, in patients with facial clefting and other hypoplastic anomalies (ectodermal dysplasia, Johanson-Blizzard syndrome, etc.). Rigid external distraction follows the important principals of aesthetic maxillofacial surgery treating only the affected jaw and offers the multiple benefits of distraction osteogenesis, including not only orthotopic bone creation with expansion of the facial skeleton but soft tissue expansion as well. The technical advantages of rigid external distraction are numerous. The surgery is minor as compared with traditional orthognathic surgery. Only the osteotomy is performed, eliminating repositioning of skeletal segments, bone grafting, splints, intermaxillary fixation, and intern fixation hardware. Operative times are significantly decreased. Morbidity with rigid external distraction is very low (in our series zero). Blood transfusions are not required. Rigid external distraction can be performed on an outpatient or 23-h admission basis. Patients begin a soft diet and normal oral hygiene the morning following surgery. The rigid external device is removed in the clinical setting or with mild sedation for the young children. All of the above has resulted in a significantly decreased cost of care for these challenging patients.

In patients with severe cleft maxillary hypoplasia, distraction osteogenesis with rigid external distraction now offers the ability to fully restore facial convexity through a minimal procedure at almost any age. In our practice, this has dramatically changed our treatment philosophies and success for these patients, who in the past have been extremely difficult to treat. The technique is currently applied with extreme success to those patients with severe maxillary nonsyndromal dentofacial deformities, as well as patients with severe syndromic conditions such as Apert, Crouzon, and Pfeiffer syndrome (Figueroa et al. 2001).

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Remodeling the Mandible by Distraction Osteogenesis

29

Fernando Molina

Distraction osteogenesis is becoming the treatment of choice for the surgical correction of mandibular hypoplasias. The development of the technique represents a significant advancement in the field of craniofacial surgery. Deficiencies in the growth of the mandible may result from condylar fractures suffered at an early age affecting growth centers or severe sepsis with secondary condyle resorption resulting with temporomandibular joint ankylosis and micrognathia. Congenital deformities such as Goldenhar's syndrome, Nager's syndrome, craniofacial scoliosis, Pierre Robin sequence, and hemifacial microsomia may present with mandibular hypoplasia of varying severity (Murray et al. 1979; Munro 1980; Lauritzen et al. 1985; Björk and Skieller 1983).

Mandibular distraction is a technique less invasive and time-intensive and has a significantly decreased morbidity rate compared with traditional methods of mandibular reconstruction. A surgeon is now able to generate new bone in patients with a bilateral mandibular body deficiency, in a severe hypoplastic ascending ramus, or to reconstruct a new condyle when it is missing in the ankylotic patient. The technique also provides the added benefit of expanding the overlying soft tissues. It probably represents the first tissue engineering surgical technique applied into the craniofacial field.

Bone lengthening or distraction osteogenesis using osteotomies and circumferential gradual distraction was described by Ilizarov (Ilizarov 1954; Illizarov et al. 1970) to align fractured segments of long bones and later to elongate these bones without a bone graft.

Although the technique initially was developed to lengthen the long bones, in 1973, Snyder reported mandibular lengthening in a canine model using an extraoral device (Synder et al. 1973). A similar report on two dogs using an intraoral device followed from Italy (Michieli and Miotti 1977). More recently, Karp and McCarthy reported membranous bone lengthening using external devices and that cortical bone formed in the expanded area of the mandible (Karp et al. 1990). Histologic examination of the zone revealed a highly organized biologic process.

Since the beginning of the 1990s, successful mandibular distraction has been performed on amultitude of patients around the world (McCarthy et al. 1992; McCormick et al. 1995; Pensler et al. 1995; Diner et al. 1996; Klein and Howaldt 1996; Polley and Figueroa 1997a; Hoffmeister et al. 1998). Our group has been performing mandibular distraction since 1990 using external corticotomy and flexible uni- and bidirectional external devices to achieve simultaneous skeletal and soft tissue correction with minimal surgery (Molina and Ortiz-Monasterio 1993, 1995; Molina 1994, 1999; Ortiz-Monasterio 1997). The technique has proven to be a major advance in the treatment of

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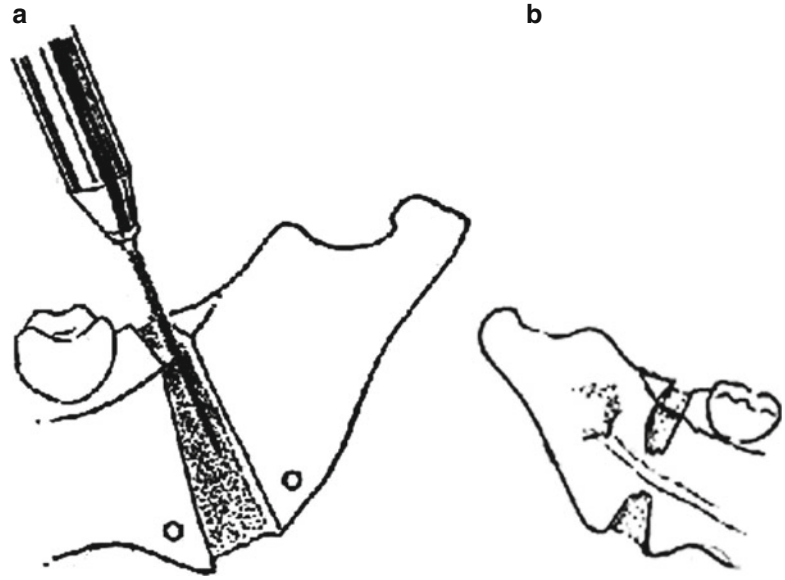

Fig. 29.1 (**a**, **b**) Side-cutting burr and the external-extended corticotomy from the free mandibular border to the gonial angle. The corticotomy is interrupted as soon as the cancellous bone is visible. (**b**) At the lingual cortex, the bone cut is incompletely, preserving 6–7 mm of the internal cortical flayer at the site of the neurovascular bundle

a variety of craniofacial deformities. In the midface animal models with or without osteotomies (Rachmiel et al. 1995; Staffenberg et al. 1995), maxillary distraction led to the first clinical trial of this concept in patients with cleft lip and palate (Cohen et al. 1997; Polley and Figueroa 1997b; Molina et al. 1998) and craniosynostosis (Arizuki and Ohmori 1995; Chin and Toth 1997; Levine et al. 1998; Molina 1998; Cohen 1999; Arnaud et al. 2003). We will include also our experience with distraction osteogenesis in craniosynostosis to achieve posteroanterior movement of the whole facial mass and a combined zygoma-malar rotation with the aim of establishing a proper dental occlusion, of increasing orbital capacity, and of improving craniofacial form with a well-balanced proportion.

29.1 Clinical Applications of Distraction Osteogenesis

29.1.1 Hemifacial Microsomia

Facial asymmetry and microtia are the most important clinical findings in a patient. Deviation of the chin to the affected side, hypoplasia of the soft tissues, and associated disorders of other anatomic structures such as the maxilla, the zygoma, and the muscles of mastication are present in a wide variability. The hypoplasia affects the gonial angle (grade I) in less severe cases and the angle and the ascending ramus in others (grade IIA and IIB) and shows a complete absence of ramus and the condyle in more severe cases (grade III) (Pruzansky 1969).

The operative technique begins with a 3–5-cm vestibular incision made under general anesthesia. The periosteum is elevated to expose the gonial angle, with a side-cutting burr; the corticotomy is done on the lateral aspect of the mandible, including all the cortex to the cancellous layer. It extends obliquely from the free mandibular border to the gonial angle. Then, the corticotomy is extensively extended inferiorly around the edge of the angle. In fact, 6–7 mm of the internal cortical layer remains intact, protecting the neurovascular bundle, and in this manner, the whole cancellous layer, the circulation, and innervation of the bone are preserved (Fig. 29.1a, b).

Afterward, a flexible unidirectional external device (KLS-Martin, Germany) is inserted. Two titanium pins are introduce percutaneously through the whole thickness of the mandible, 4–5 mm in front and behind the corticotomy. The pins should be parallel to each other to facilitate the fixation of the distraction device. In this moment, we recommend an "intraoperative test" of accuracy: activate the device 5–6 mm and observe the changes in the corticotomy. The surgeon must obtain the opening of the corticotomy segments without difficulty and assess that the elongation system works properly. If the surgeon identifies the presence of cortical

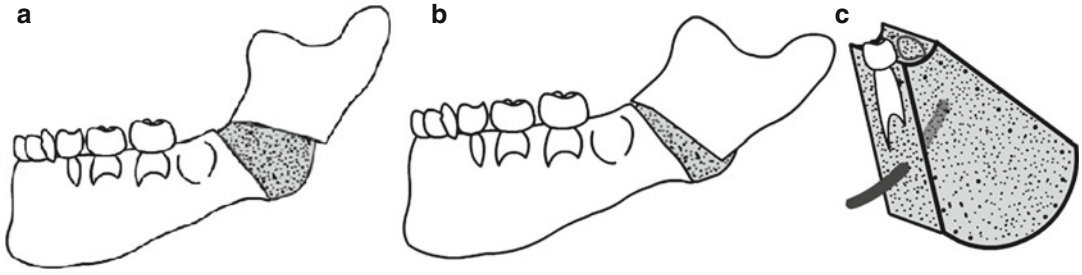

Fig. 29.2 (**a**) Location of the corticotomy in a grade II-A hemifacial microsomia. (**b**) Using an oblique vector, the new bone formation production is larger at the angle and minor at the alveolar ridge, also at the initial portion of the ascending ramus. (**c**) Tridimensional diagram of the regenerate bone. The new volumetric bone formation corrects the height, the length, and position of the mandible. The tooth buds and neurovascular elements have been preserved

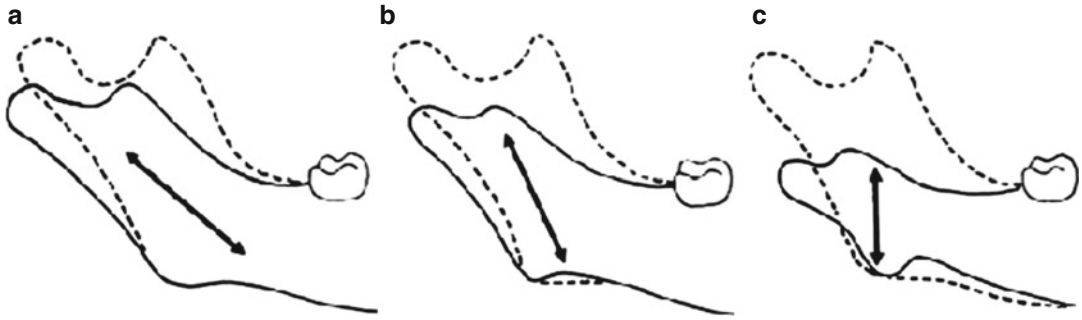

Fig. 29.3 Distraction vectors are the key to obtaining excellent results. Basically, three distraction vectors are used to correct a mandibular hypoplasia in hemifacial microsomia. The oblique vector is used for grade Pruzansky I, and the vertical vector is used to recreate the ascending ramus in the grade Pruzansky II-B

bridges, the lengthening mechanisms will not work and the corticotomy must to be revised.

The advantage of combining the use of an external corticotomy and a flexible device is the ability to obtain a 3D correction of height, length, and position of the mandible. In fact, different zones of bone resistances are encountered, minor of the angle but major resistance at the alveolar ridge, that will be elongated differently under a perpendicular distraction vector (Fig. 29.2a–c).

The area of new bone formation produces a better shape of the mandibular angle and modifies the ascending ramus position with a simultaneous better relationship of the condyle and the glenoid fossae. The initial condyle lateral position moves into a more central one that is more similar with the contralateral normal one. All of these findings are different from those obtained with linear distractors and osteotomies.

The selection of the vector of distraction is also a critical decision. The corticotomy and the position of the pins determine the distraction vector. It is different in each patient depending on the grade of mandibular hypoplasia. In patients with grade I, the pins must be placed perpendicular to the corticotomy, obtaining an oblique vector that produces a larger bone elongation in the angle and a minor bone elongation in the alveolar ridge. In patients with grade II-A hypoplasia, the distraction process must remodel and elongate the angle and the initial portion of the ascending ramus (Fig. 29.3). For this reason, the pins must be inserted in an intermediate position between a vertical and oblique distraction vector. This group of patients represents the vast majority of our clinical series.

In patients with grade II-B hypoplasia, the corticotomy is placed horizontally in the base of the ramus. The pins must follow a strict vertical distraction vector in order to obtain more elongation in the hypoplastic ascending ramus.

Our distraction protocol includes a latency period of 4–5 days. Distraction commences at a rate of 1.0 mm/day (distraction period). This period is continued 4–6 weeks until we achieve the desired

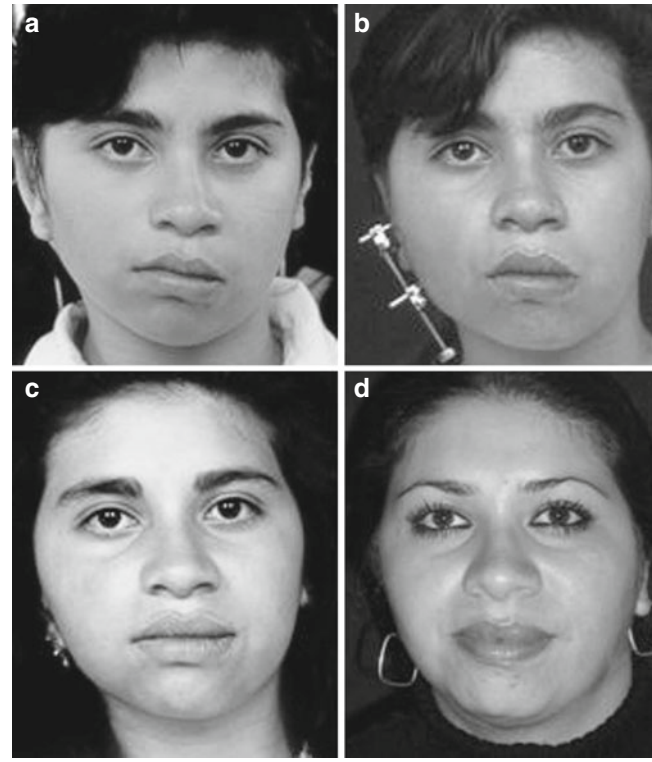

Fig. 29.4 (**a**) Preoperative frontal view of a 9-year-old girl with a Pruzansky II-A hemifacial microsomia showing facial asymmetry and chin deviation to the affected side. (**b**) During the distraction process, 2 weeks after the initial 16 mm of mandibular elongation showing an early improvement of the facial asymmetry. (**c**) Six months after 24 mm of mandibular distraction showing facial symmetry with descent of the buccal commissure and medialization of the chin. (**d**) The patient 12 years later showing stability in the clinical result. Facial symmetry: a chin position has been stable in the long-term result

mandibular lengthening with the correction of the facial asymmetry, chin position, level of the oral commissure, and the changes in the dental occlusion. A very important point is to produce an overcorrection in the group of growing patients. In the preoperative period, the patients present a marked lateral deviation of the mandibular teeth to the affected side. After the distraction has been completed, the deviation of the mandibular interincisal line is reproduced in the opposite direction in the contralateral side as a result of overcorrection. Also, a posterior open bite and a crossbite are produced immediately after distraction; for these reasons, posterior bite blocks, dynamic appliances, and standard orthodontic maneuvers will easily control the occlusal changes and will produce some degree of dental occlusion stability. Bite blocks were required to maintain the posterior open bite; the blocks were gradually reduced to allow vertical descent of the maxillary dentoalveolus. We must always avoid the production of occlusal disasters as this situation will be impossible to correct later with standard orthodontic procedures.

The consolidation period is about 8 weeks or more. It depends on the age of the patient as well as the amount of bone lengthening. In an adult patient with a severe hypoplastic ramus, it can be prolonged for 16 weeks until there is radiographic evidence of mineralization. At this time, the device is removed under sedation.

Age is also a critical factor in the treatment of a hemifacial microsomia patient with mandibulardistraction. During the 1990s, the international criteria were to indicate the procedure at an age of 5–6 years, including the more severe cases. This decision was established according to the amount of mandibular bone stock and the feasibility of using intra- or extraoral distraction devices. Actually, we know that patients under 5 years of age, presenting a severe or moderate deformity, are not good candidates to elongate the mandible with intraoral devices. The procedure will result in a failure to achieve the ideal distraction vector, secondary occlusal disasters, and the need for future surgical interventions.

In our clinical series, aesthetic results were excellent. The facial symmetry was restored with descent of the buccal commissure to the level of the contralateral normal side; the menton became horizontal and was located at the midline (Fig. 29.4a–d). In all our patients, we noted an

increase in the soft tissue envelope bulk. This simultaneous soft tissue expansion is the most important factor in obtaining the correction of facial asymmetry, and it is more observed during the consolidation period. At this time, the device serves as an external fixator, and the muscles of mastication produce contraction forces over the regenerate bone allowing the molding of the callus with the increase of the bigonial distance by the muscles' pull.

Our clinical series also includes a group of patients with hypoplasia grade II-B treated between 18 months and 3 years of age. Undoubtedly this group of children has better craniofacial growth and long-term clinical stability in the soft tissues. After overcorrection, the use of dynamic orthodontic appliances and bite blocks corrected the occlusal changes and the vertical dimension of the maxilla. All of these changes occurred very fast and almost spontaneously during a period of 2–3 months, compared with children treated over age of 6 years. For this reason every day, more and more, we are indicating the procedure at early ages. It is also important to mention that permanent dental injury or neurovascular damage did not occur. Elongation was done by the parents with minimal discomfort during the night without disturbing the children's sleep. These children represent the group of patients with the best results and better tolerance to the method.

Twelve percent of the patients with hypoplasia grades II-A and II-B treated after 6 years of age required a second distraction procedure. The main reasons for a secondary procedure were erroneous distraction vector, incomplete consolidation period, and nonsatisfactory overcorrection criteria. These patients presented an early tendency to retake the original condition of facial asymmetry and malocclusion as it was observed after 2–3 years of follow-up (Fig. 29.5a–i).

If the patient is 12 years old or more at the time of the second bone elongation, we indicate a simultaneous maxillomandibular distraction. This technique is also indicated in the adult group of patients as a primary surgical procedure (Fig. 29.6a, b). Additional to the mandibular distraction, we include a subperiosteal dissection of the maxilla, and then a complete horizontal osteotomy is made at the level of the pyriform area in both sides. The pterygomaxillary junction is freed with a curved chisel and Rowe forceps on the affected side only. Rowe forceps may be used softly to assess the completeness of the osteotomy, but no attempt is made to mobilize the midface. The pterygomaxillary junction on the unaffected side remains intact, and it will serve as a pivot to safely produce the midface elongation-rotation movements on the affected side.

The result shows that with the mandibular elongation, the maxilla follows the mandible changes, achieving simultaneous elongation, medial rotation, and advancement. Preoperatively, the deviation of the occlusal plane from the horizontal varied from 12° to 18°, and after the combined distraction procedure, the clinical and orthodontic correction was obtained with a minimal slanting of 1–2° in the long-term follow-up.

Preoperatively, the vertical dimension of the maxilla was very short, and the last mandibular molar was in a very close relationship with the malar bone. Postoperatively, the vertical maxillary increases from 16 to 15 mm, representing a 90 % correction when compared with the unaffected side. All of this new bone formation results in a simultaneous correction of the oblique nasal floor, as well as the deviation of the nasal septum changing into the more normal position, increasing the volume of the nasal cavity. The preexisting dental occlusion has been preserved; after the elongation, the superior and inferior midline incisors have changed and now are positioned in the central line of the face (Fig. 29.7a–d).

The chin is always medialized by the distraction, but in adults it is not always possible to achieve a normal central position; for this reason, this procedure does not eliminate the need for a sliding genioplasty or another extrasurgical procedure, like a free dermis-fat graft, at the time of the removal of the device to improve the aesthetic final result.

29.1.2 Micrognathias

Patients with micrognathias present a different problem because of the bilateral deformity and because both the mandibular body and the ascending ramus are affected, requiring bidirectional

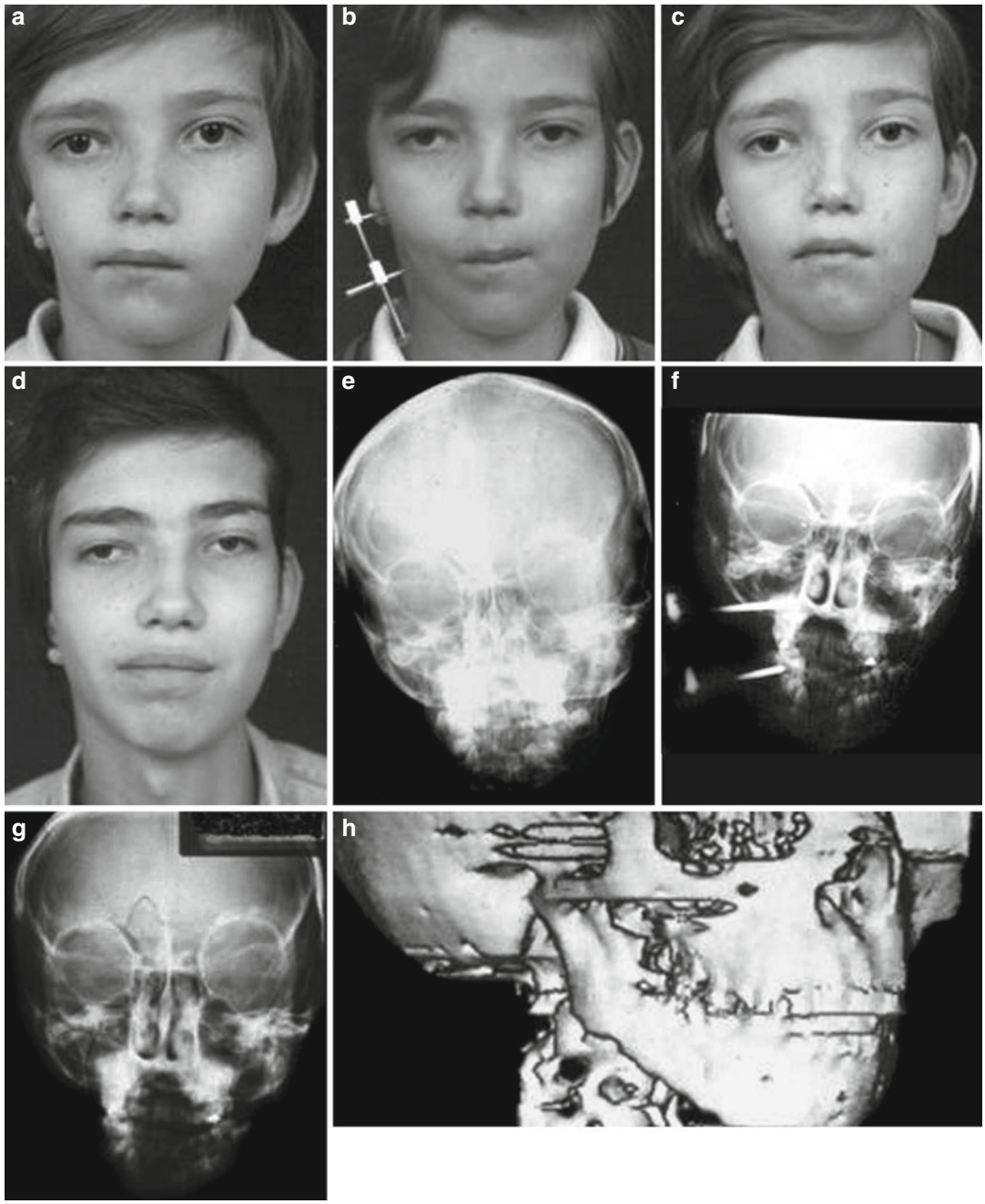

Fig. 29.5 (**a**) Preoperative frontal view of a 6-year-old boy with hemifacial microsomia Pruzansky II-A showing facial asymmetry with deviation of the buccal commissure and the chin. (**b**) During the distraction process. (**c**) The patient 6 months after showing facial symmetry. (**d**) Frontal view 10 years later. We can observe excellent growth of the mandible together with the rest of the craniofacial skeleton. (**e**) Preoperative PA cephalogram. (**f**) During the distraction process showing a low bone density area between the pins. (**g**) Postdistraction PA cephalogram. Notice the increase in the vertical dimension of the mandible, the condyle position, and the changes in the maxilla. (**h**) Preoperative CT scan showing the shortness of the ascending ramus

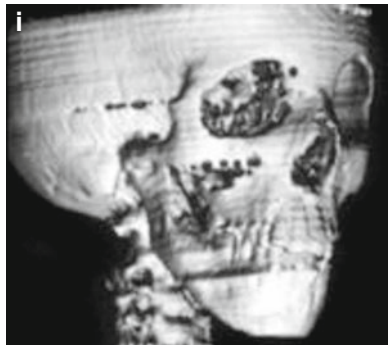

Fig. 29.5 (*continued*) (**i**) Postdistraction CT scan showing the increase of the ramus length as well as the new volumetric change of the gonion

and bilateral distraction. This concept is also true for patients with Pierre Robin, Nager, Treacher Collins, and bilateral hemifacial microsomia syndromes. In these cases, two corticotomies were done: a vertically oriented one in the mandibular body and a horizontally oriented one in the ascending ramus. The pins used are as follows: a central one is introduced at the mandibular angle between the two corticotomies, a second into the mandibular body, and a third into the central aspect of the ascending ramus. One bidirectional device is used on each side, each one with two distraction plates to allow independent and more precise elongation of each segment, using the central pin as the fixed pivot for both of them (Fig. 29.8a, b).

Most of these patients present the typical "bird face" deformity with deficient soft tissue at the lower third of the face and at the neck, absence of the neck angle, and shortened suprahyoid muscles. With bone distraction, all the tissues from skeleton to skin are simultaneously elongated without the inconvenience of osteotomies or skin expansion (Fig. 29.9a–h). After conventional osteotomies and bone grafts, the muscles and the tight skin envelope present a limiting factor often requiring multiple procedures and seldom achieving optimal aesthetic results. Tissue expansion increases the size of the skin cover, but other soft tissues such as muscles, vessels, and nerves remain unchanged. The overall functional and aesthetic results with bidirectional mandibular distraction have been spectacular. The neck takes a normal

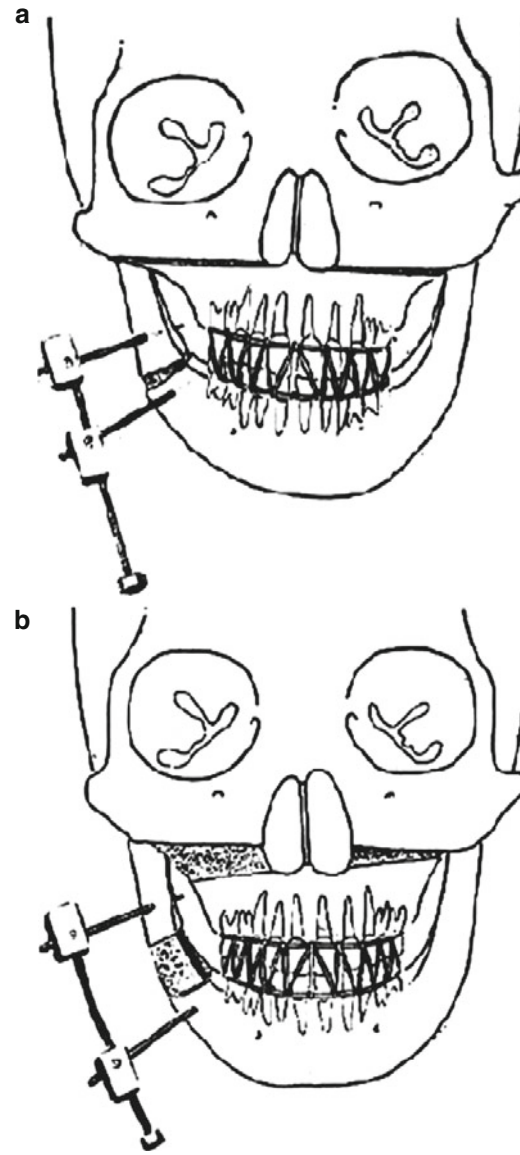

Fig. 29.6 (**a**) Diagram showing the mandibular corticotomy with the device in site. An additional horizontal osteotomy has been performed over the maxilla. After 5 days, an intermaxillary fixation is indicated with the aim of obtaining a simultaneous elongation of both bone structures. (**b**) Areas of new bone formation over the mandible and the maxillary

shape with a well-defined angle. The muscles and soft tissues of the floor of the mouth together with the masticatory muscles got the expansion giving a better aspect of the neck. The chin takes a more prominent position.

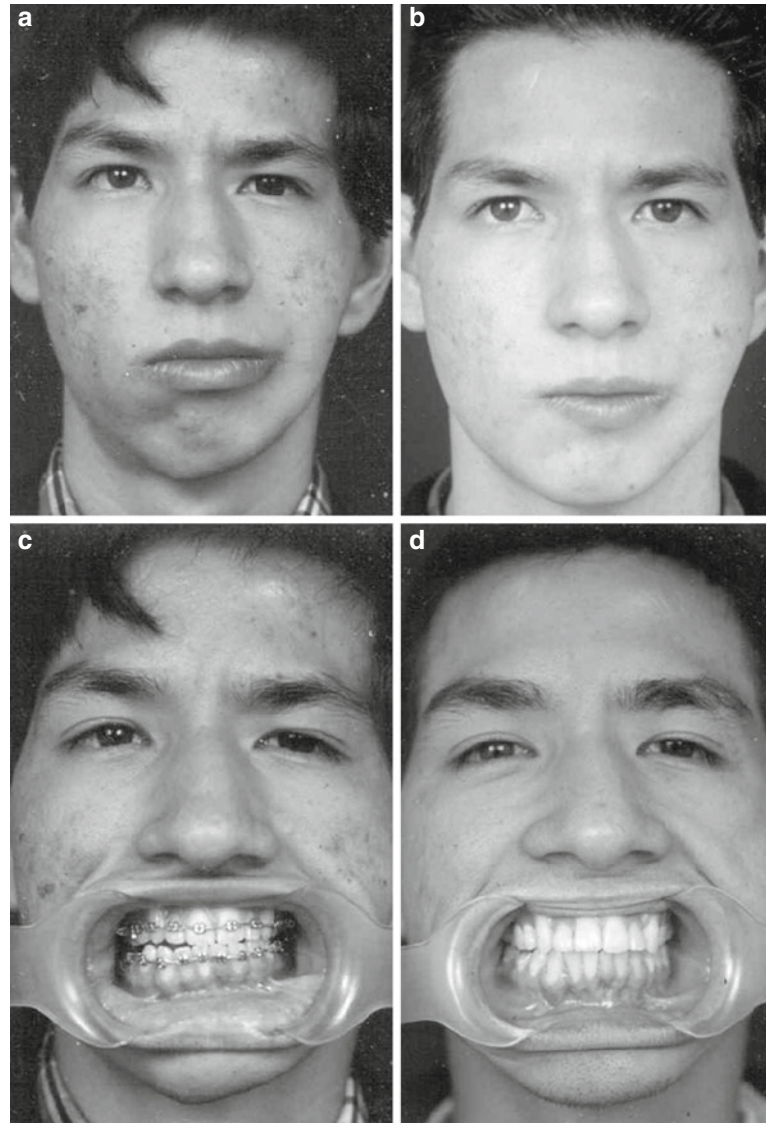

Fig. 29.7 (**a**) Preoperative frontal view of an adult patient with left hemifacial microsomia. (**b**) Six months postdistraction frontal view after a simultaneous maxillomandibular distraction. (**c**) The patient showing the operative occlusion. Notice the obliquity of the occlusal plane. (**d**) Six months after distraction, the patient shows its occlusion with the correction of the occlusal cant

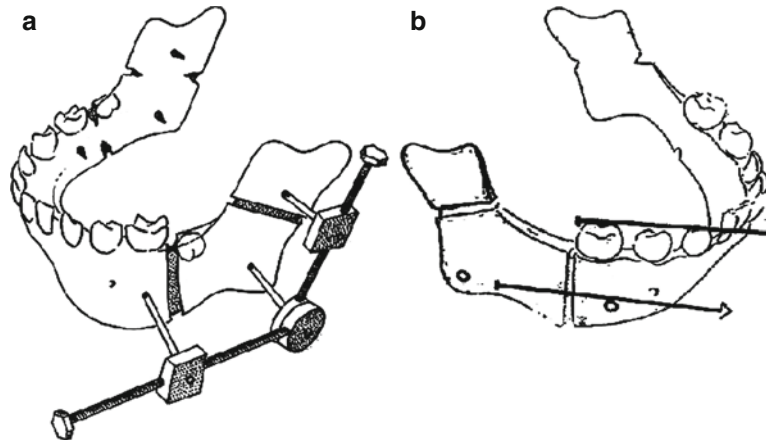

Fig. 29.8 (**a**) Diagram showing two corticotomies for bidirectional elongation. The central osteotomized bone fragment must be large enough to avoid fracture and back displacement. (**b**) The horizontal vector always has to be parallel to the occlusal plane in order to avoid the production of an anterior open bite or other occlusal disasters

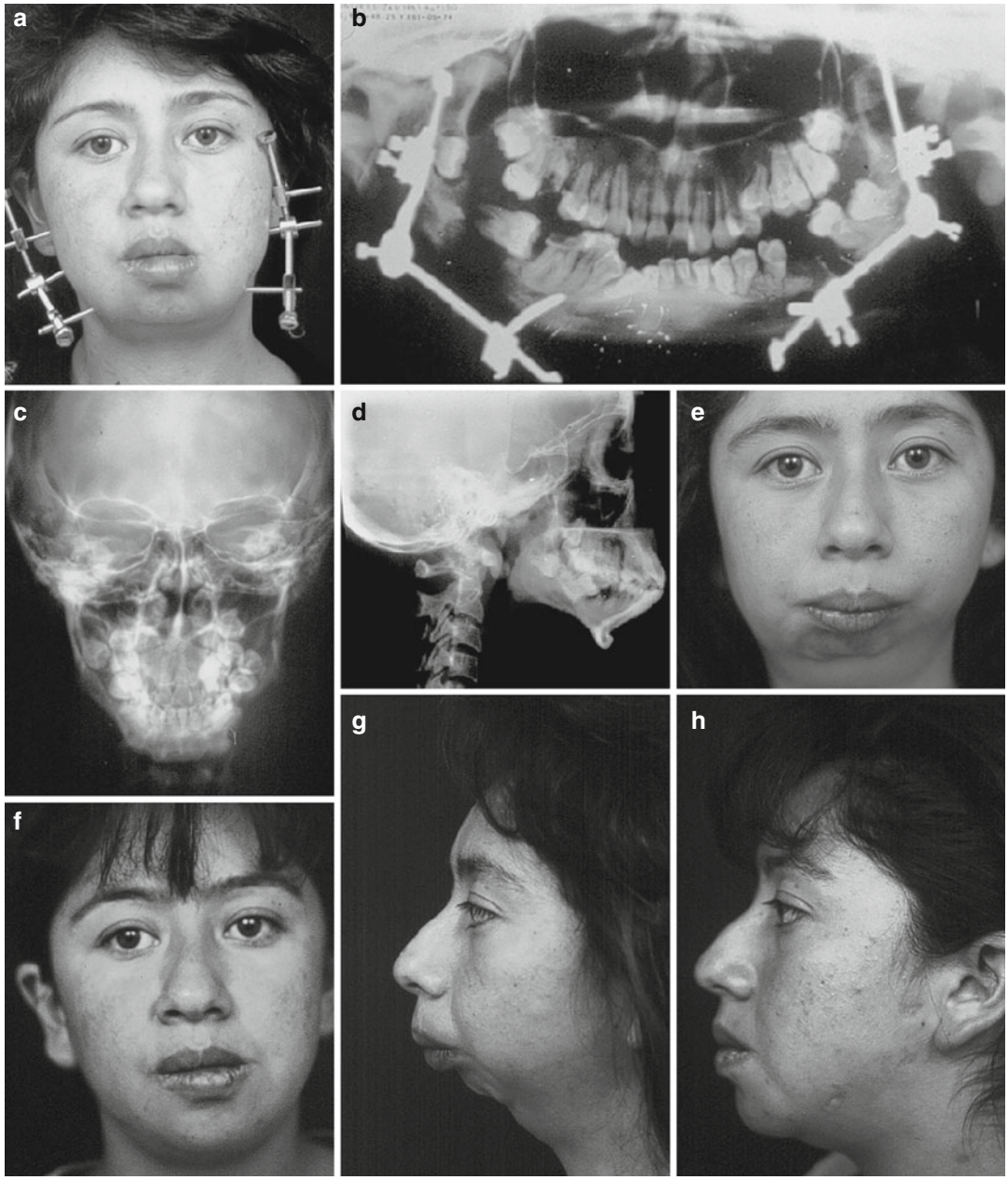

Fig. 29.9 (**a**) Frontal view of a 22-year-old girl during the distraction process of a bilateral and bidirectional mandibular elongation. (**b**) Panorex view showing the four independent vertical and horizontal distraction vectors. In asymmetrical micrognathias, one mandibular body can be elongated more than the contralateral in order to obtain symmetry. (**c**) Preoperative frontal view of the patient showing the asymmetrical micrognathia secondary to a condyle fracture during the early years of life. (**d**) Frontal view 3 years later of the mandibular distraction. Facial symmetry has been restored. (**e**) Preoperative lateral view showing the classic "bird face." (**f**) Lateral view 3 years later showing the new mandible with a nice elongation of the soft tissues over the superior third of the neck. (**g**) PA cephalogram 3 years later showing a very well-defined mandibular structure. (**h**) Lateral cephalogram 3 years later. After mandibular elongation, the procedure has obtained a normal anatomy with an ascending ramus, gonion, mandibular body, and chin. The use of proper distraction vectors has achieved a normal occlusal relationship

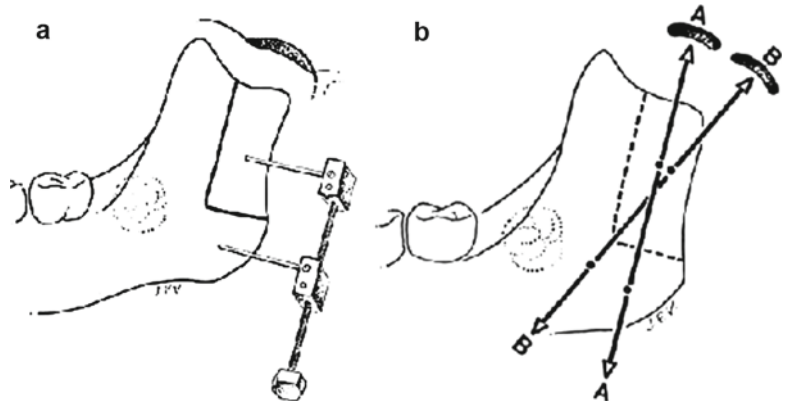

Fig. 29.10 (**a**) Diagram showing the "L osteotomy" over the ascending ramus with the device in place. (**b**) The distraction vector must be directed to the empty glenoid fossae

29.1.3 The Temporomandibular Joint Ankylosis

Studies of the effect of mandibular distraction on the temporomandibular joint have shown that the condyle assumes a more normal anatomic size, shape, and position after distraction (McCormick et al. 1995). In fact, the process of distraction is beneficial to the condyle structure and its position in the glenoid fossae.

Regardless of the etiology of the joint ankylosis, it can be treated using the concept of bone transport and distraction to generate a neocondyle and to obtain a normal mandibular anatomy. In extra-articular ankylosis through a vestibular incision and subperiosteal dissection, an L-shaped osteotomy is made from the medial aspect of the projected condyle down to the posterior aspect of the ramus. Two pins are inserted, one in the osteotomized bone fragment and a second at the angle. The distraction vector is directed to the empty glenoid fossae (Fig. 29.10a, b). After a 5-day latency period, the osteotomized segment of bone is distracted and transported in a vertical direction until achieving the planned condyle lengthening. Usually 14–20 mm is enough to achieve a similar dimension with the contralateral side. Immediately the patients begin with open-close exercises, and during this activation period, the edge of the neocondyle is remodeled into a smooth, rounded surface. Fibrocartilaginous tissue at the leading edge of this segment acts as a pseudodisk.

If the patients present an intra-articular ankylosis (true bony fusion), an arthroplasty is performed through a preauricular incision. The surgeon must release the bony fusion, and he will avoid bone resection reducing the ramus height. The abnormal bone is remodeled with a burr, and once this maneuver is completed, the opening of the mouth is accomplished with the assistance of a special forceps. Then a very fine Silastic layer (less 1.0 mm) is introduced as a cap on the top of the projected condyle and fixed to the surrounding scar tissue with two resolvable stitches. We close the incision, and then we change into the intraoral route for the L-shaped osteotomy.

This method produces a vertically elongated mandible with a functioning nonankylosing temporomandibular joint in the short term. The interincisal distance obtained ranged from 35 to 45 mm in the adult group. This technique will probably become the treatment of choice of temporomandibular joint ankylosis. Better diagnostic methods and understanding of etiology will be necessary in the near future to determine the efficacy of this process.

29.1.4 Cleft Lip and Palate Patients

The etiology of midface deficiency in cleft patients is uncertain. The absence of class III malocclusion in untreated clefts suggests that previous cheiloplasty and palate repair may be related to this condition (Ortiz-Monasterio et al. 1959). Ross (1987) in a longitudinal survey of 528 repaired cleft patients from 15 centers around the world demonstrated that approximately 25 % develop maxillary hypoplasia that did not respond to orthodontic procedures alone.

At the time of treatment, the ages of patients ranged between 6 and 12 years (mean 8 years), and all of them demonstrated clinical and radiographic evidence of moderate to severe underdevelopment of the maxilla (sagittal, vertical, and transverse). They also presented with a flattened middle third of the face and class III malocclusion, with partial or total anterior and posterior crossbite.

The severity of maxillary hypoplasia ranged from 2 to 10 mm of A-P discrepancy. Class III malocclusion with anterior and posterior crossbite, deep overbite, and in some patients, vertical skeletal open bite was observed.

Preoperative orthodontics are mandatory to obtain a minimal occlusal stability. The following orthodontic requirements were fulfilled prior to maxillary distraction: skeletal and/or dentoalveolar transverse expansion of the maxilla, alignment of permanent incisors in the cleft area in some patients, and bone grafting at 8–10 years old in other cases (Subtelny and Brodie 1954; Tindlund 1989, 1994).

Prior to surgery, a modified quad-helix fixed appliance was placed in the palate. This is made of a simple lingual arch attached to bands placed on the second deciduous molars. Anteriorly the arch maintains passive contact with the incisors through an acrylic button. In order to maintain maxillary expansion during the advancement, an extra transverse arch is fixed to both premolar bands which in turn provide the support for the vestibular hooks.

Under general anesthesia, two maxillary vestibular incisions 3–4 cm long are made, leaving a 2-cm central mucosal bridge between the incisions. Subperiosteal dissection is performed from the pyriform fossae to the lateral maxillary buttress on each side exposing the anterior and lateral aspects of the maxilla to the level of the infraorbital nerve and above the canine roots. Dissection on the nasal floor is limited to the lateral part only. The nasal septum and the maxillary cleft gap remain intact. Using a side-cutting burr, a horizontal osteotomy is performed above the roots of the canines and molar tooth buds as identified on the posteroanterior cephalogram and panorex.

Bilaterally, using a 7-mm chisel, the osteotomy is extended into the maxillary buttress, reaching the pterygomaxillary region. Disjunction with Rowe forceps is not performed, thereby avoiding unnecessary bleeding and uncontrolled fractures in the pterygomaxillary junction. At this time, the surgeon must assess maxillary mobility by applying gentle downward pressure to the Le Fort I segment. The mucosa is then approximated and closed with absorbable sutures. Following this minimally invasive technique, the hypoplastic maxilla is ready to be mobilized. As the nerve function and vascularity have been preserved, the mechanical distraction forces will stimulate the functional matrix to produce new bone formation.

Using the fixed intraoral appliance system as an anchorage for a modified "petit" facial mask supported by the forehead and chin, distraction forces are initiated on postoperative day 5. Two intraoral elastic bands on each side are linked from vestibular hooks to a bar on the mask. No other fixation is required. The initial mechanical force used for distraction is about 900 g. The facial mask is used mainly at night and at some periods during the day for a total of 16–18 h every day.

When the vertical length of the maxilla is deficient, a combination of forward and downward distraction forces can be used to simultaneously advance and lengthen the maxilla.

Advancement of 2–3 mm is achieved each week. During the second and third week of distraction, the presence of palatal scars may produce difficulties in further advancement. At this stage, it is critical to maintain the same elastic force in order to weaken the resistance of the fibrous tissue. If necessary, the elastic bands, which are initially parallel, may be crossed in order to increase the force of A-P distraction. In this manner, the projected maxillary advancement and a satisfactory class II molar relationship is obtained. At this point, the amount of force is decreased to one elastic band on each side (450 g) for another 2 months (consolidation period) in order to maintain the maxilla in its new position.

Radiographic evidence of new bone formation is observed at approximately 8–10 weeks postdistraction. The bone step formed by the horizontal distraction is replaced with mature cortical bone. There is also evidence of a secondary area of new bone

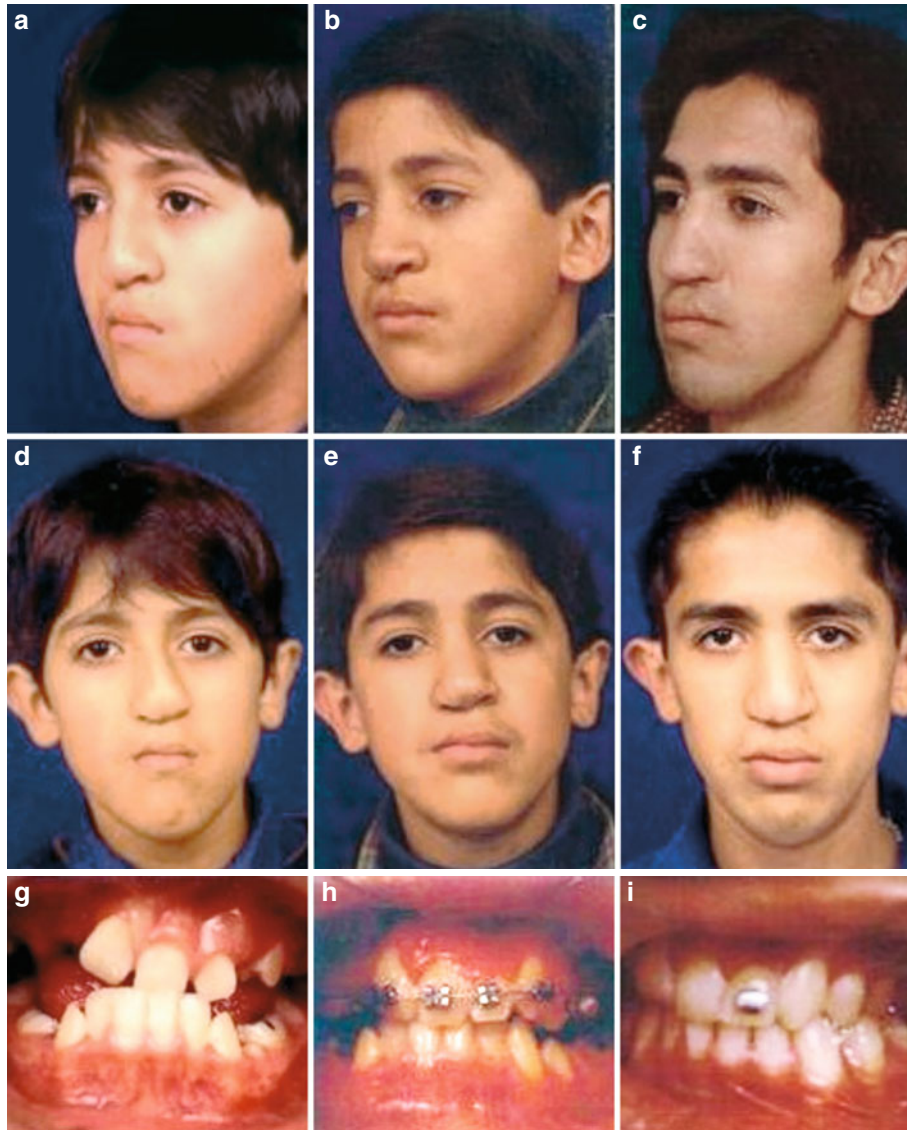

Fig. 29.11 (**a**) Preoperative three-quarter view of a 9-year-old boy with unilateral cleft of lip and palate and moderate midface retrusion. (**b**) Postoperative three-quarters view 1 year before 6 mm of advancement and 3 mm of vertical elongation with maxillary distraction. Notice that the new position of the maxilla produces a minor anterior mandibular rotation. The middle third of the face has been elongated, producing a more appropriate balance of the middle and inferior thirds of the face. (**c**) A postoperative three-quarter view 9 years later. (**d**) A preoperative frontal view. Because the middle third of the face is retruded and short, an exaggerated anterior rotation is produced in the mandible. The patient looks prognathic and with a very long inferior third of the face. (**e**) The face in its thirds looks well proportioned and balanced. (**f**) Clinical stability with nice aesthetic results in the long term. (**g**) Preoperative occlusion showing negative overjet and posterior crossbite. (**h**) Orthodontic treatment continues that includes the alignment of the permanent incisors in the cleft area and conventional orthodontics. (**i**) The final occlusion

formation at the pterygomaxillary junction. These findings attest to the skeletal stability and will diminish the risk of relapse.

The projected advancement was achieved in all of the patients with results varying from 4 to 12 mm (mean 7 mm). The aesthetic results were excellent (Fig. 29.11a–f). The soft tissue profile changed from concave into a well-proportioned orthognathic face. Changes also included an increased nasolabial angle, a more anterior

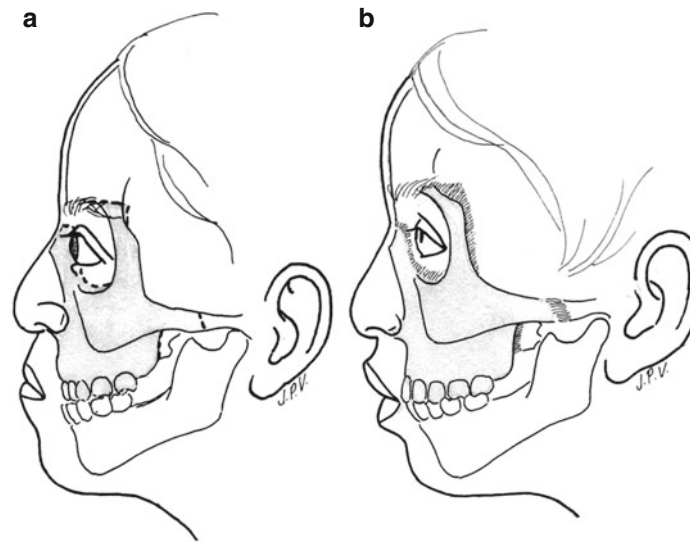

Fig. 29.12 (**a**) Osteotomy line of the modify Le Fort III osteotomy. At the nasal bones and at the supraorbital border, the osteotomy preserves some millimeters of bone attachments, avoiding the production of long noses and bone steps at the fronto-orbital region after the advancement. Malocclusion in class III is observed. (**b**) After advancement, new bone formation is observed along the osteotomy lines, most importantly in the lateral aspect of facial framework, the lateral orbital wall, the zygoma, and pterygomaxillary area. At the nasal bone and the superior orbital edge, new bone formation is minimal when compared with the rest of the osteotomies. The occlusion has been corrected

projection of the upper lip, and an improved lip relationship.

Dental occlusion was changed to a distal sagittal molar relationship with moderate overjet (Fig. 29.11g–i) with significant downward clockwise rotation of the mandible. Cephalometrically, a counterclockwise rotation of 2–7° was achieved. The increase in the ANB angle varied 1.4–7.4°. After distraction was completed, it was essential to stabilize the maxilla with a fixed palatal arch, in order to proceed with the orthodontic treatment such as alignment of permanent incisors in the cleft area and to correct the minor dental irregularities in the cleft site.

29.1.5 Craniosynostosis

In this clinical series, different osteotomies are included: Le Fort III, fronto-orbital advancement, and monoblock. The osteotomies were performed using a subcranial or an intracranial approach. The intracranial route is always assisted endoscopically in order to perform a clean dissection of crista galli and cribriform plate, to assess for hemostasis and to verify dura mater disruptions, and most importantly to preserve vascularity between the dura and the skull. The surgical technique uses several basic procedures of the classic craniofacial osteotomies. Under general endotracheal anesthesia, exposure of the midface and the orbits is obtained through a standard coronal incision.

The dissection is carried over the periosteum, and at the temporal level, the dissection extends to the deep temporal fascia so that the soft tissue envelope can be reflected off the lateral orbital rim and zygoma. At this time, the temporal muscle is sectioned below the temporal crest, and with a limited dissection 15 mm wide, we reach the lateral orbital wall until the retromalar area. In fact, we preserve the vast majority of the temporal muscle insertion.

Then the dissection is extended to the orbit superiorly at the orbital roof 8–10 mm from its apex, laterally until the inferior orbital fissure is clearly visualized, and medially until the nasolacrimal groove without disruption of the medial canthal ligament.

In a patient with previous fronto-orbital advancement and cranial vault surgery with midface hypoplasia and associated class III malocclusion (Fig. 29.12a, b), a subcranial Le Fort III is indicated. The osteotomy is designed including the lateral half of the supraorbital rim. Here, the osteotomy has to preserve 6–8 mm of bony union at the medial portion. Then the osteotomy is followed to the external aspect of the lateral orbital wall and extended down until reaching the junction of the maxilla and the pterygoid plate. At this point, an interpterygomaxillary disjunction is done with a 12-mm chisel. The intraoral classic route is avoided. Then an oblique osteotomy is performed at the zygomatic arch.

Following intraorbitally the bone cut at the inferolateral angle, it extends from the inferior orbital fissure along the orbital floor reaching medially the posterior aspect of the nasolacrimal groove. This maneuver allows the advancement of the nasolacrimal apparatus and medial canthal tendons anteriorly with the midface.

Over the nasal base, one horizontal osteotomy is performed connecting the medial orbital wall osteotomies in both sides. Craniofacial disjunction is accomplished with Rowe Kiley forceps. The surgeon must avoid fracturing the remaining 6–8-mm bone union at the supraorbital rim. This incomplete osteotomy will act as a pivot and will avoid bone steps in the fronto-orbital area after the advancement.

The distraction devices are placed in the midface in two different ways: simply supported behind malar bone or anchored with a hook to the malar-orbital edge. The device is a submerged type and is made of one hollow plate with a central perforation to allow the entrance of a screw running free, 25–30 mm in length. After the screw thread, the rest of the device has a smooth surface and can be easily positioned through the temporalis muscle reaching the malar's back or anchoring to the bone edge with an extra hook. The hollow plate is screwed to the parietal bone.

To advance the midface, a horizontal vector is used. The vector has to be parallel to the occlusal plane, and the midface is advanced until a class II molar relationship is obtained, and/or the zygoma-malar-orbit advancement corrects the exorbitism.

If the patient's midface is short vertically, then an oblique vector is the logical indication. Simultaneously, the bone movement will correct the midface retrusion and will lengthen its vertical dimension.

The obtained subcranial advancement ranges from 18 to 30 mm. This distance has been measured at the orbito-malar region. It is very important to observe that the maxillary advancement ranged from 7 to 12 mm and at the supraorbital region varied from 10 to 16 mm. Certainly we have obtained a gradual bone advancement that is different at the orbit, at the malar, and at the maxilla allowing the correction of the exorbitism and of the midface hypoplasia.

This gradual advancement is generated by an additional rotational movement in the lateral aspect of the facial framework, more accentuated at the zygoma-malar area. Probably this additional movement offers a more satisfactory correction of the exorbitism and avoids the production of the long "French noses" as typically are obtained after Le Fort III advancements.

Aesthetic results were excellent, and a final facial appearance is comparable with normal, good-looking individuals (Fig. 29.13a–d).

In a primary syndromic craniosynostosis patient (Apert, Crouzon, Pfeiffer, etc.) presenting with severe midface hypoplasia and secondary respiratory problems, the monoblock osteotomy is the indicated. The design of this osteotomy represents a "true monoblock" because frontal bone, both orbits, malars, and maxilla move anteriorly together in one single piece (Fig. 29.14a, b).

In these cases, the osteotomy is outlined at the frontal bone and the lateral aspect of the osteotomy runs down until it reaches the lateral orbital wall. It then follows in a manner similar to the subcranial route.

Two or three craniotomies are performed. Below the outlined osteotomy design, intracranial tunnels are dissected, introducing large fine gauzes in order to protect the meninges. Special attention must be paid at the tunnels at the supraorbital area as well as at the crista galli.

At this point, a flexible endoscope is introduced into the tunnels, and assessment for hemostasis is performed. Also, a safe intracranial dissection at the more dangerous area must be completed under vision; this maneuver is especially important in patients in which the dura is firmly attached to the internal cortex. The maneuver will avoid meningeal disruptions.

This sort of intracranial dissection preserves vascular attachments between the frontal bone and the dura along with the preservation of the bone vascularity. In reality we are generating bone flaps that will produce a healthy new bone formation along the osteotomy lines very quickly. The preserved vascularity will avoid intracranial

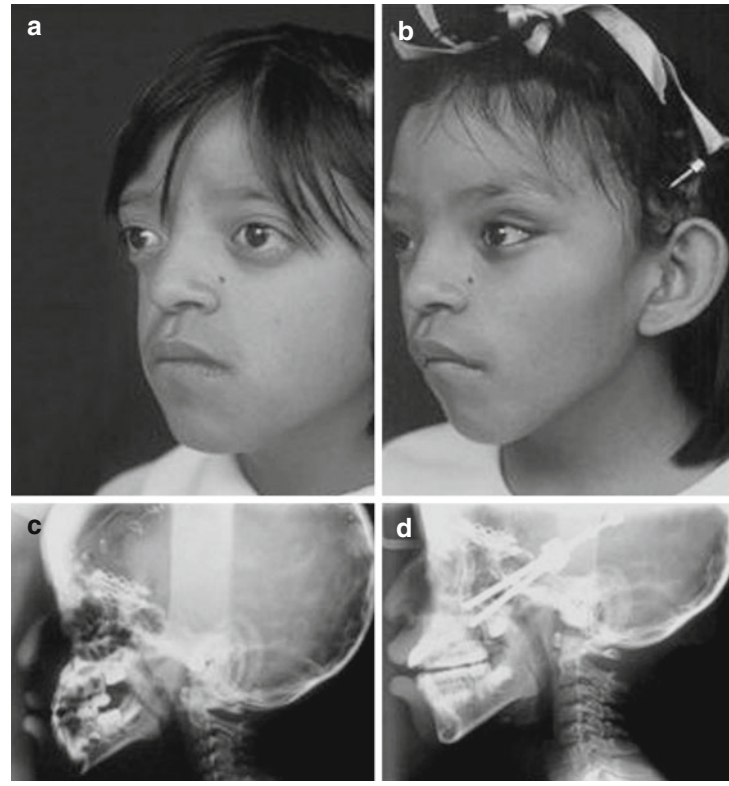

Fig. 29.13 (**a**) A 4-year-old girl with Crouzon disease. A fronto-orbital advancement has been performed at age of 12 months. Exorbitism and severe midface retrusion with sleep apnea are still unresolved problems in this patient. (**b**) At the end of the distraction process, malar-zygoma advancement was 19 mm, solving the exorbitism. The maxillary position has corrected the airway problem. Facial proportions now are well balanced. (**c**) Preoperative lateral cephalogram showing the retruded midface and the rigid fixation system, mini-plates, and wires used in the fronto-orbital area during the surgical procedure. Notice the fine airway secondary to the severe maxillary hypoplasia. (**d**) Postdistraction lateral cephalogram showing the bone advancement and the position of the devices. An oblique vector has produced the bone advancement. It has been gradual and different at the orbital level and at the maxilla level

dead spaces and secondary bone necrosis, often observed after the classic advancements with osteotomies.

The frontal osteotomy is performed with an oscillatory saw. A fine brain retractor is introduced into the tunnels over the gauzes, and then the cut is completed safely. Over the orbital roof and the medial wall, a curve osteotome is used to complete the osteotomy.Of significant importance is completion of the bone cut at the crista galli. This maneuver is done with two 7-mm chisels. The first one is introduced through the central craniotomy and endoscopically positioned in front of the crista galli; then with gentle hammering, the osteotomy is begun. Immediately thereafter, the second chisel is positioned exactly in front of the first one, and the osteotomy is completed medially and laterally until the bone cut reaches the orbital roof.

At this point, a Rowe Kiley forceps is used to complete the monoblock disjunction, and maxillary disimpaction forceps can be used in an effort to facilitate the anterior midface advancement with simultaneous stretching of the overlying soft tissue envelope.

A pair of distraction devices are used to advance the whole bone mass in one piece. Using the extra hooks, the devices are anchored to the fronto-orbital union edge, through the temporalis muscle, and screwed to the parietal bone.

The postoperative course in this group of patients has been very satisfactory. Monoblock advancements ranged from 20 to 30 mm in the fronto-orbital region and from 10 to 18 mm in the maxillo-malar area.The exorbitism has been corrected in all cases. It is our impression that simultaneous advancement of the roof, the floor, and the medial and lateral walls of the orbit produces a more anatomical correction (Fig. 29.15a–d). In the frontal region, gradual advancement of the frontal bone has produced good results, both in the contour of the forehead and cranium and in the relationship with the facial skeleton.

Extradural dead spaces between the frontal bone and the frontal lobes have not been observed

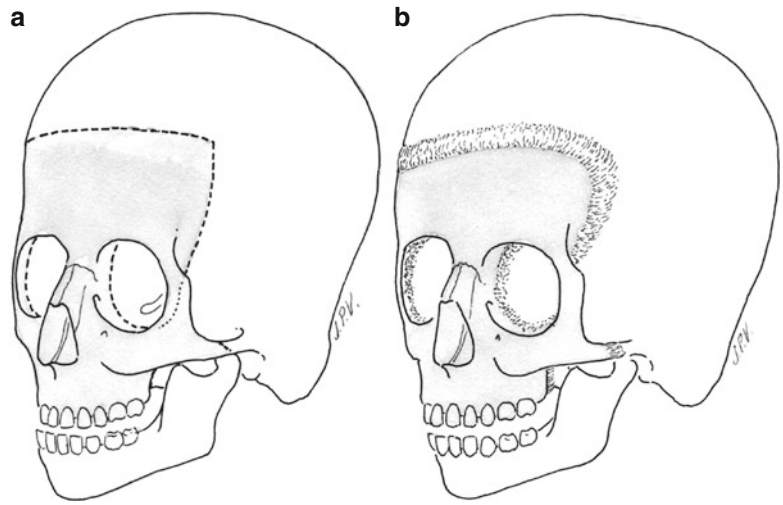

Fig. 29.14 (**a**) The "true monoblock" osteotomy. *Dotted line* shows the single-piece osteotomy that includes the frontal bone, the orbits with their four walls, the lateral and medial walls, and the floor, the pterygomaxillary junction, and the zygoma. (**b**) After distraction, new bone formation is observed along the osteotomy lines included at the pterygomaxillary tuberosity

in any of our patients. Also, irregular contour in the forehead secondary to partial bone absorption has been eliminated with the distraction advancement.

We link together the orbito-cranial and maxillary problems because their primary malformation occurs simultaneously and its correction is critical to obtain excellent functional results.

29.2 Discussion

Distraction osteogenesis is a technique that is becoming popular for the reconstruction of deficient mandibles and has simplified the treatment for congenital mandibular hypoplasia. Technically, it is a minor surgical procedure that preserves the integrity of the nerves and vascular supply. Careful planning of the corticotomy and the position of the pins produces a distraction vector that follows closely the direction of normal mandibular growth (McCarthy et al. 1992; McCormick et al. 1995; Pensler et al. 1995; Diner et al. 1996; Klein and Howaldt 1996; Polley and Figueroa 1997a; Hoffmeister et al. 1998).

Vertical and sagittal mandibular elongation was obtained in all patients. During the early stages of our clinical work, we assumed that the elongation would always result in a posterior open bite. However, it did not occur in many of the cases due to dentoalveolar adaptation at the maxillary level (Molina and Ortiz-Monasterio 1993, 1995; Molina 1994, 1999; Molina et al. 1997). This vertical growth change was documented cephalometrically, suggesting that the maxilla liberated from the constricting effect of the small mandible and deficient soft tissue can reach its normal growth potential. Murray and Mulliken (1979) have reported the same increased vertical maxillary growth after chondrocostal grafting of the ascending ramus.

An enormously important benefit of bone distraction is the simultaneous expansion of the surrounding soft tissue of the face (Molina et al. 1995, 1997). In fact, we were surprised to see the rapid descent of the buccal commissure to a normal position, the horizontalization of the chin, the increase in the distance between buccal commissure and the external canthus and in the distance between buccal commissure and the inferior orbital rim in the unilateral cases. Also, once the distraction is completed, we have observed some additional increase in the volume of the cheek, which is probably related to improved muscular activity.

All the patients with micrognathia presented the typical "bird-like" facial deformity with deficient soft tissue in the lower third of the face and the neck, causing absence of the neck angle, with shortened suprahyoid muscles. The

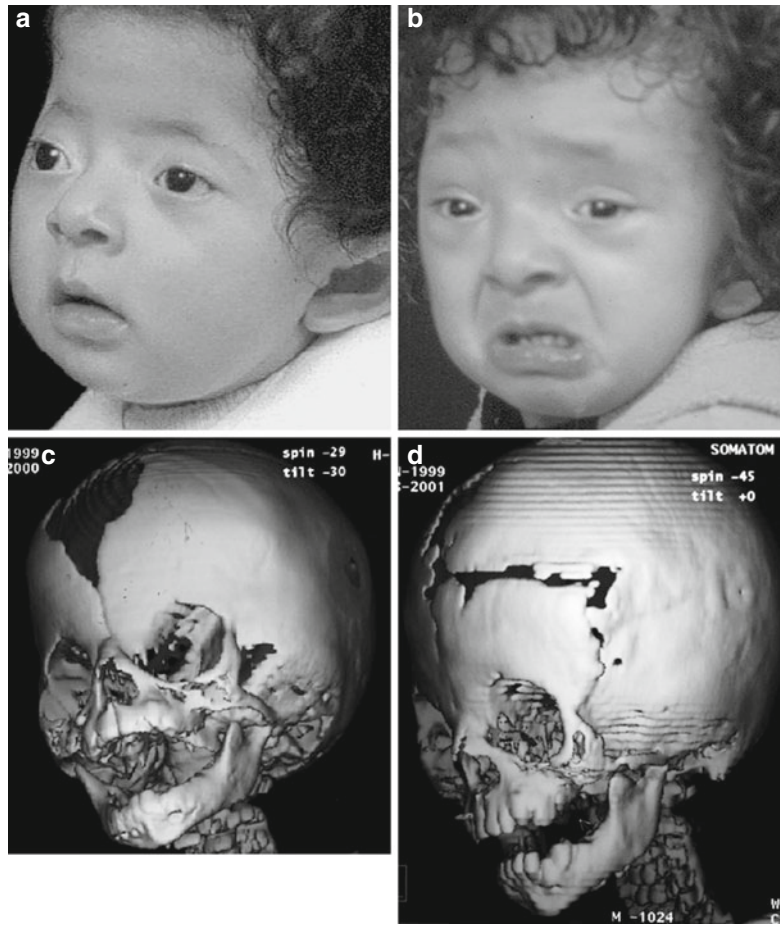

Fig. 29.15 (**a**) Preoperative third-quarter view of a 14-month-old girl with Apert syndrome showing a severe supraorbital depression, fronto-orbital deformity, and midface retrusion. (**b**) Postdistraction clinical changes. The "true monoblock" has corrected frontal deformity, exorbitism, and midface retrusion. Oxygen saturation is normal during sleep. (**c**) A preoperative CT scan of a 14-month-old young girl with Apert syndrome, brachycephaly with a severe flattened supraorbital region is observed together with severe midface retrusion. Oxygen saturation drops during the sleep of the patient. (**d**) Postdistraction CT scan shows the new position of the midface with the simultaneous correction of the orbits position and the frontal bone after a "true monoblock" advancement. New bone formation is observed in the lateral aspect of the frontal bone, in the lateral orbital wall, the zygoma, and at the pterygomaxillary area. On the frontal bone, an extra remodeling is observed in the patient during the consolidation period. The brain expansion force produces a round shape on the frontal bone achieving a structure very closely to the normality

overall aesthetic results obtained have been spectacular, beyond both the patients' and our expectations. The neck takes on a normal shape with a well-defined angle, the muscle and soft tissue of the floor of the mouth expand, the masseter insertion and the muscle volume increase, and the chin becomes more prominent.

Slow distraction of the mandible following corticotomies results in increased mandibular mass and length simultaneously with soft tissue (skin, muscles, ligaments, blood vessels, and nerves) expansion. This adaptation occurs over a period of 8–12 weeks. Elongation of the mandible results in an increase in size as well as changes in its positions and shape that would

not be possible using conventional orthognathic surgery such as a sagittal split osteotomy procedure (Converse et al. 1973; Kaban et al. 1988; Ortiz-Monasterio 1982).

The early treatment of midface retrusion in children with C.L.P. has to be considered a major goal. Early correction of the maxillary hypoplasia minimizes the psychological problems as well as provides benefits related to improved occlusion, masticatory, and respiratory functions. Advancing the maxilla with distraction forces requires only a minimal surgical procedure preserving vascularity and nerve integrity. This technique presents exciting treatment possibilities eliminating the need for rigid fixation systems, blood transfusions, prolonged orthodontic treatment, and intermaxillary fixation.

When the maxilla is properly positioned, it can grow in a near normal manner, allowing the tongue to assume a physiological position. In this situation, the permanent maxillary incisors erupt spontaneously into a normal overjet and overbite (Subtelny and Brodie 1954; Tindlund 1989, 1994).

The use of distraction vectors is fundamental to achieve optimal advancements. Vectors are obtained through the combination of the position of the bar on the facial mask and the direction of the rubber bands. There are three possibilities of maxillary distraction vectors: upward, forward, and downward. The most commonly used is the forward one, in which the horizontal vector must be parallel to the occlusal plane to avoid producing an anterior open bite.

Overcorrection with a class II molar relationship is always performed, especially in patients with severe deformity or in patients treated during 6–9 years of age. This point is critical because distraction does not alter the inherent cellular growth pattern. Following early maxillary distraction in patients with severe midface retrusion, the individual growth pattern often reverts to the original tendency of reduced maxillary growth and an anterior crossbite may become reestablished. Overcorrection is therefore necessary to achieve an equilibrium between maxillary and mandibular growth. The use of intraoral functional correctors (Frankel III) also plays an integral role by adding an additional stimulus for maxillary growth.

Gradual maxillary advancement with distraction produced functional changes affecting respiration. Nasal breathing was improved as well as the airflow and patency of the nasal airway. These changes are produced by an opening in the nasolabial angle and the increase of volume of the nasal and pharyngeal airway as demonstrated in the preoperative and postoperative lateral cephalograms.

Two relevant points must be considered to predict the difficulties with the midface distraction technique: first, the quality of palatal tissue and palatal scarring. Due to the tendency for retraction and relapse, fixed retention is mandatory. Secondly, preoperative skeletal and dentoalveolar transverse maxillary expansion, as well as dental compensations, may be necessary to prevent occlusal instability. Clinical experience has proven that these factors determine the long-term stability of the maxillary advancement in this series.

Also, with maxillary distraction, a simultaneous soft tissue expansion is observed, and the skin, fat, and muscles have a more favorable perioral, perinasal, and infraorbital distributions. The nasal base remains unchanged, and the position of the lips, teeth, and tongue is in better relationship when the patient smiles.

In treating craniosynostosis, conventional osteotomy surgery is limited by the resistance of the soft tissue envelope which can be managed only by gradual advancement or multiple surgical procedures. The results of midface osteotomy advancement with rigid fixation typically fall short of the desired goal (Tessier 1967, 1971a, b, 1976; Ortiz-Monasterio et al. 1978; Van der Meulen 1979; McCarthy et al. 1990).

It is well known that the growth of the cranial vault is directly influenced by the brain, the growth of the orbit directly induced by the growth of the eye, and the growth of the maxilla linked to the eruption of teeth (Latham 1970). Normally, until 4 years of age, the cranial base, like the vault, follows the rapid and even pseudotumoral growth of the brain. In addition, between the ages of 2 and 7 years, the cranial base follows the development of the face; the later depends on dental eruption and masticatory movements (Tessier 1971b).

These are very important observations to indicate early midface surgery in patients with craniosynostosis. Moreover, after 7 years of age, the development of the frontal bone, the ethmoid, and the sphenoid depends on their pneumatization. The growth of the anterior cranial base appears, therefore, to be induced mainly by that of the brain and the maxilla.

These two last facts are very interesting to analyze since distraction osteogenesis in these patients produces areas of new bone formation over the anterior cranial base and the brain expansion after the procedure has produced a secondary remodeling of the frontal bone. The brain expansion molds the regenerate.

At present, when the operative risk has been reduced to the minimum, when most of the major complications can be prevented and minor complications can be controlled, and when good functional results are usually obtained, more of our efforts must be directed toward better cosmetic results. The ideal goal in craniofacial surgery would be to produce a normal appearance in all patients (Arizuki and Ohmori 1995; Chin and Toth 1997; Molina 1998; Cohen 1999).

Conventional osteotomy surgery is limited by the resistance of the soft tissue envelope, which can be managed only by gradual advancement or multiple surgical procedures (Whitaker et al. 1987; Tessier 1977; Ortiz-Monasterio et al. 1990).

The advantage of distraction in midface advancement is thus an increase in the amount of correction that can be achieved in a single procedure, enabling a more complete anatomic correction and possibly sparing the patient multiple-stage procedures (Chin and Toth 1997; Molina 1998; Cohen 1999; Arnaud et al. 2003).

Distraction osteogenesis also can offer excellent aesthetic possibilities. During the process, the surgeon can perform some extra changes or adjustments during the elongation period to finally obtain the proper position of the different segments. In fact, we can obtain a harmonious relationship between the thirds of the face, producing a normal appearance in many patients.

Other advantages to treating a craniosynostosis with distraction are: it allowed the advancement to be performed without the need of bone grafting, thereby decreasing the length of the procedure and the donor-site morbidity. It had significantly lower relapse rates than standard midface advancement in cleft lip and palate (Cohen et al. 1997; Polley and Figueroa 1997b; Molina et al. 1998), presumably because the soft tissue, which traditionally resists midface advancement, was also gradually distracted along with the bone. The lack of initial mobilization significantly decreased the time needed for postoperative recovery. There is a significant decrease in blood loss and postoperative pain after this procedure, and patients are typically able to be discharged from the hospital earlier. Gradual advancement using distraction allows the displacement of skeletal fragments over greater distances because the soft tissue envelope is allowed to accommodate gradually. The magnitude of advancement is almost twice that obtained with traditional advancement procedures and has the possibility of adding a vertical midface lengthening, correcting the shortness of this structure.

All of these features will diminish morbidity and will produce excellent functional changes and the possibility of better craniofacial growth.

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Management of Maxillary Deformities in Growing Cleft Patients

30

Eric J.W. Liou and Philip K.T. Chen

30.1 Introduction

The maxillary deformities in growing cleft patients could be a hypoplastic maxilla, downward and/or laterally displaced premaxilla in bilateral clefts, wide alveolar cleft and fistula, or combinations of the above deformities. It is a challenge for both orthodontist and surgeon to treat a hypoplastic maxilla with wide alveolar cleft in a growing unilateral or bilateral cleft patient or a downward or laterally displaced premaxilla with wide alveolar cleft and fistula in a growing bilateral cleft patient.

Several new clinical techniques based on the principles of distraction osteogenesis, either nonsurgical orthopedic or surgical approaches, have been developed for managing the maxillary deformities in growing cleft patients. These techniques are:

1. *Effective maxillary orthopedic protraction* for the treatment of hypoplastic maxilla (Liou and Tsai 2005) and for minimizing alveolar cleft (Liou and Chen 2003) in unilateral or bilateral cleft patients
2. *Premaxillary orthopedic intrusion* for the correction of a downward displaced premaxilla in bilateral cleft patients (Liou and Chen 2003; Liou et al. 2004)
3. *Premaxillary orthopedic medial repositioning* for the correction of a laterally displaced premaxilla in bilateral cleft patients (Liou and Chen 2003)
4. *Interdental distraction osteogenesis* for the approximation of a wide alveolar cleft in unilateral or bilateral cleft patients (Liou et al. 2000)

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30.2 Orthopedic Management of Hypoplastic Maxilla in Growing Unilateral or Bilateral Cleft Patients

The combined use of rapid maxillary expansion and facemask is a contemporary orthopedic management for maxillary protraction in cleft patients (Kawakami et al. 2002; Tindlund 1994; Tindlund and Rygh 1993). It is assumed that rapid maxillary expansion disarticulates the circumaxillary sutures so that facemask protraction of maxilla could be easier (Haas 1970; McNamara 1987; Turley 1988). Rapid maxillary expansion is sutural expansion osteogenesis of the intermaxillary suture and has been recognized as a form of distraction osteogenesis (Liu et al. 2000). Facemask protraction of maxilla is sutural protraction osteogenesis of the circumaxillary sutures. In the absence of a completely intact

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intermaxillary suture in cleft patients, disarticulation of the circumaxillary sutures through rapid maxillary expansion would play an important role in maxillary protraction.

However, it remains controversial to what width the expansion should reach to disarticulate the circumaxillary sutures. In noncleft patients, some reported that 5 mm of expansion is good enough (Alcan et al. 2000), while some others reported at least 12–15 mm (Haas 1980, 2000). Both for the cleft and noncleft patients, it seems that wider expansion exerts greater tension/stress on the circumaxillary sutures and disarticulates maxilla better than a smaller expansion. However, the expansion should be to displace the maxilla anteriorly and to disarticulate circumaxillary sutures rather than to overexpand the maxilla. To disarticulate circumaxillary sutures without overexpansion, a technique called effective maxillary orthopedic protraction has been developed for orthopedic management of the hypoplastic maxilla in growing cleft patients (Liou and Tsai 2005).

30.2.1 Effective Maxillary Orthopedic Protraction

This technique includes three parts:

1. A double-hinged rapid maxillary expander for a greater amount of maxillary anterior displacement
2. A protocol of *alt*ernate *ra*pid *m*axillary *e*xpansions and *c*onstrictions (Alt-RAMEC) of maxilla for a better disarticulation of maxilla
3. A pair of intraoral maxillary protraction springs for noncompliant maxillary protraction

30.2.1.1 Double-Hinged Rapid Maxillary Expander

Several types of rapid maxillary expanders have been used for maxillary protraction. They are the *fan-type* (Suzuki & Takahama 1989; Levrini & Filippi 1999) or *hyrax-type* built with two acrylic resin halves (Haas 1970), splints (McNamara 1987), or in a hygienic design (Biederman and Chem 1973). These expanders expand the maxilla in a V-shaped manner (Vardimon et al. 1998) with a center of rotation around the posterior nasal spine (Lee et al. 1997; Braun et al. 2000). The expansion force distributes not only in the maxilla but also into the circumaxillary structures (Chaconas and Caputo 1982; Itoh 1985). It is postulated that this would entail bone resorption behind the maxilla and, consequently, result in posterior displacement of maxilla (Biederman and Chem 1973) (Fig. 30.1a, b). In contrast, it is postulated as well that this would entail the circumaxillary structures such as pterygoid plates to displace the maxilla forward (Haas 1961, 1965) (Fig. 30.1c).

These two assumptions explain why some of the clinical studies on hyrax-type expanders reported anterior displacement of maxilla (Haas 1970; Wertz 1970; Akkaya et al. 1999), while some others reported no significant displacement (Da Silva Filho et al. 1991; Pangrazio-Kulbersh et al. 1998) or posterior displacement of maxilla (Sarver and Johnston 1989; Cozza et al. 2001). The posterior displacement of maxilla compromises the maxillary protraction in cleft patients.

The double-hinged rapid maxillary expander (US Patent No. 6334771 B1) is developed for a greater amount of anterior displacement of maxilla (Liou and Tsai 2005; Liou and Chen 2003). Its configuration is similar to a W-appliance and has two hinges of rotation. It consists of a jackscrew in the center, two bolts holding the screw, a body holding the bolts at anterior and two hinges of rotation at posterior (Fig. 30.2a, b). The rationale for its greater amount of anterior displacement of maxilla is it expands and rotates each half of the maxilla laterally and anteriorly through the two hinges of rotation located beside the molars bilaterally. This kind of expansion entails the circumaxillary structures to displace the maxilla anteriorly with less possibility of bone resorption behind the maxillary tuberosity (Fig. 30.1d).

Maxillary first premolars and molars are banded and maxillary impression is taken for the fabrication of the double-hinged expander. The expander is oriented perpendicular to the central incisors and is soldered to the molar and premolar bands. Two anterior extension arms (0.051-in. stainless steel wires) extend bilaterally from the premolar bands toward central incisors

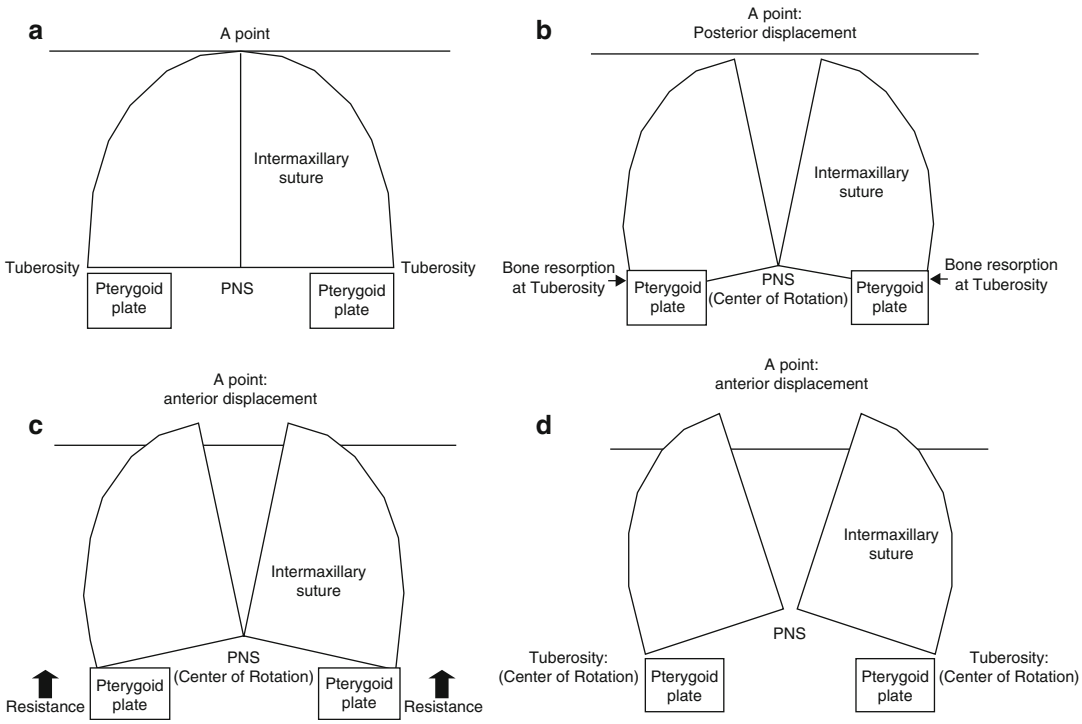

Fig. 30.1 (**a**–**d**) Schematic illustrations of the postulated maxillary displacement during rapid maxillary expansion. (**a**) The maxilla before expansion: the *semicircles* represent the right and left maxillae; the *rectangles* represent the pterygoid plates. (**b**) Posterior displacement of the maxilla after expansion by a hyrax expander: each half of the maxilla rotates outward and backward around the posterior nasal spine (*PNS*), which entails bone resorption behind the maxillary tuberosities and results in posterior displacement of maxilla. (**c**) Anterior displacement of the maxilla after expansion by a hyrax expander: each half of the maxilla rotates outward and backward around the PNS, which entails the circumaxillary structures to displace the maxilla forward and results in anterior displacement of maxilla. (**d**) Anterior displacement of the maxilla after expansion by a double-hinged expander: each half of the maxilla rotates outward and forward around the maxillary tuberosities, which geometrically results in anterior displacement of maxilla without the possibility of entailing bone resorption behind the maxillary tuberosities

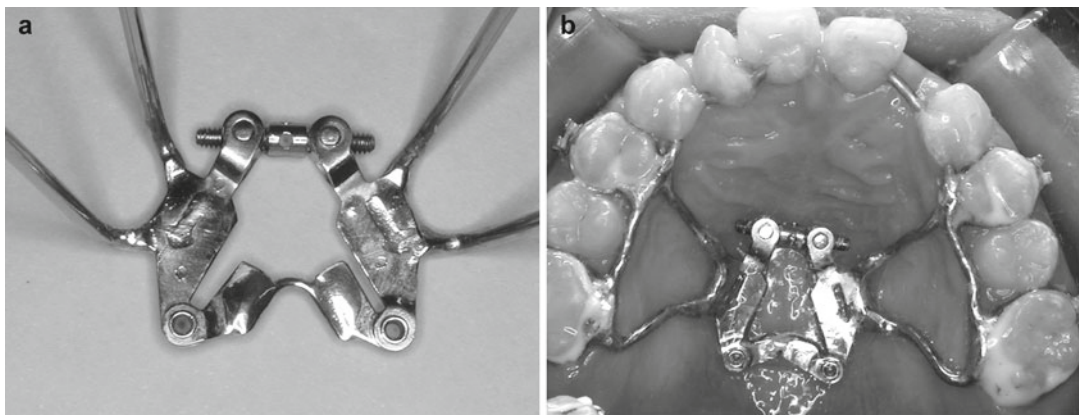

Fig. 30.2 (**a**, **b**) The configuration of the double-hinged rapid maxillary expander (**a**) and the assembly of the device in a clinical case (**b**)

(Fig. 30.2b). The premolar and molar bands and the anterior extension arms are sandblasted before cementation. After cementation of the expander, the anterior extension arms are bonded to the anterior teeth with composite resin. One day after cementation, the double-hinged expander is activated according to the protocol of Alt-RAMEC.

30.2.1.2 Alternate Rapid Maxillary Expansions and Constrictions (Alt-RAMEC) of Maxilla

The Alt-RAMEC is a protocol of repetitive weekly alternate rapid maxillary expansions and constrictions (Liou and Tsai 2005; Liou and Chen 2003) (Table 30.1). It lasts for 7–9 weeks until the maxilla is loosened. The weekly (every 7 days) sequence is 7 mm of expansion, 7 mm of constriction, 7 mm of expansion, 7 mm of constriction, 7 mm of expansion, 7 mm of constriction, 7 mm of expansion, 7 mm of constriction, 7 mm of expansion, etc. Each day, the maxilla is expanded or constricted 1 mm/day (four turns in one activation). This protocol allows better disarticulation of circumaxillary sutures without overexpansion of maxilla. Its rationale is similar to simple tooth extraction in which we repeatedly rock the tooth buccally and lingually until the tooth is disarticulated out of the alveolar socket.

Patients are seen once a month. The maxilla is examined clinically for its loosening by holding the patient's head with one hand and rocking the expander with maxilla up and down with another hand. The maxilla is ready for protraction only once loosening of the maxilla has been observed clinically.

Table 30.1 Clinical protocol for alternate rapid maxillary expansions and constrictions

Alternate weekly sequence	Weekly amount of expansion/constriction (mm)	Daily amount of activation (mm)
Expansion	7	1
Constriction	7	1
Expansion	7	1
Constriction	7	1
Expansion	7	1
Constriction	7	1
Expansion	7	1
Constriction	7	1
Expansion	7	1

30.2.1.3 Maxillary Protraction Springs for Effective Maxillary Orthopedic Protraction

The maxillary protraction device is a pair of noncompliant, tooth-borne, intraoral maxillary protraction spring (US patent 6273713 B1) (Liou and Tsai 2005; Liou and Chen 2003) (Fig. 30.3). It is a 0.036-in. b-nickel-titanium helix spring. Ball pins are used to mount the spring on the maxillary and mandibular headgear tubes. The spring is activated by the lower jaw movement. It is passive and in 180° when the mandible opens (Fig. 30.3a). While the mandible closes, it is compressed to 100–120° and generated 300–400 g of horizontal and upward force on the maxilla (Fig. 30.3b). A 0.036-in. b-nickel-titanium mandibular lingual holding arch with built-in lingual crown torque is used for splinting the mandibular dentition as an anchor unit for the protraction (Fig. 30.3c).

30.2.1.4 Treatment Protocol for Effective Maxillary Orthopedic Protraction

The treatment protocol for effective maxillary orthopedic protraction includes 7–9 weeks of Alt-RAMEC and 4 months of maxillary protraction using intraoral maxillary protraction springs. The total treatment protocol is 6 months. The patients are seen every 4 weeks for adjusting or replacing the intraoral maxillary protraction springs when they are distorted or broken. The expander and protraction device are removed at the end of 6th month.

30.2.1.5 Treatment Results and Effects of Effective Maxillary Orthopedic Protraction

The differences between Alt-RAMEC and a single course of rapid maxillary expansion (RME) have been evaluated in a clinical cephalometric study (Liou and Tsai 2005). Twenty-six consecutive cases of unilateral cleft lip and palate patients with hypoplastic maxillae (SNA < 82) were

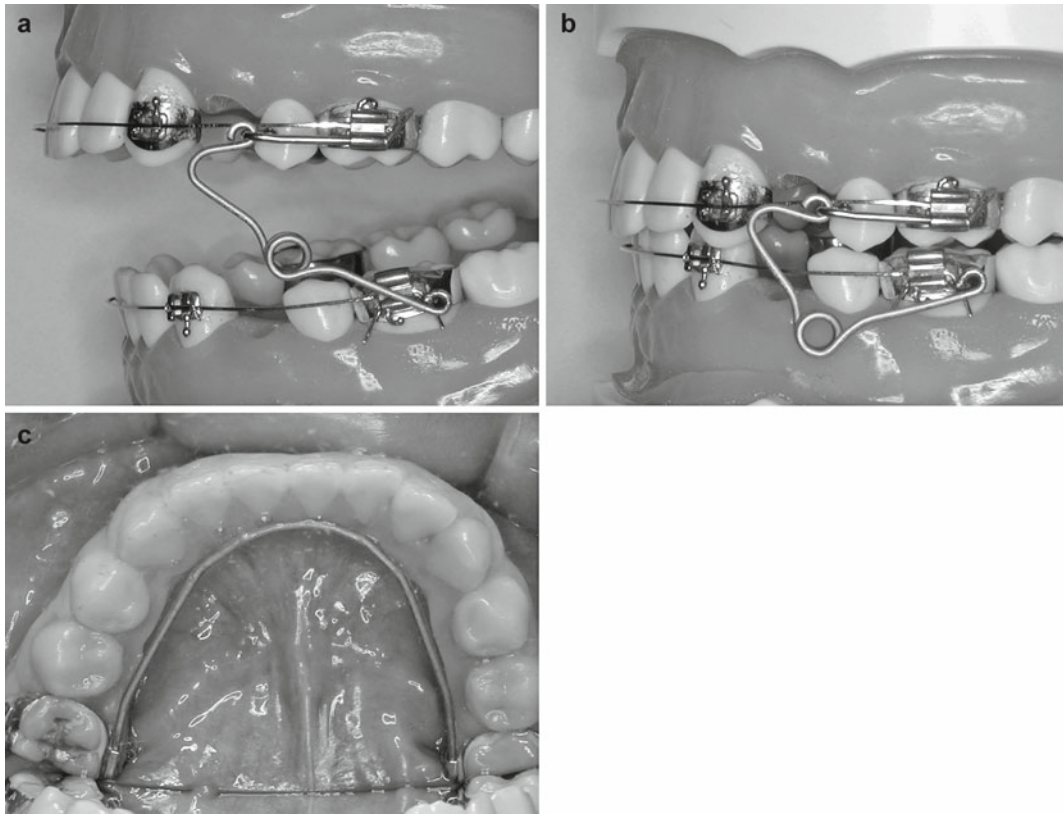

Fig. 30.3 (**a**–**c**) The intraoral maxillary protraction spring (**a**, **b**) and removable b-nickel-titanium mandibular lingual holding arch (**c**) for effective maxillary orthopedic protraction

included for maxillary protraction. Their ages ranged from 9 to 12 years old. The group of RME (7 days, 1 mm/day) was the first 16 consecutive cases. The group of Alt-RAMEC was the next ten consecutive cases. The expander used in both groups was the double-hinged expander, and the protraction appliance was the intraoral maxillary protraction springs.

The amount of maxillary anterior displacement by the double-hinged expander in the Alt-RAMEC group was 3.0 ± 0.9 mm at A point. This was significantly greater than the 1.6 ± 1.0 mm in the RME group. The amount of maxillary advancement with intraoral protraction springs in the Alt-RAMEC group was 2.9 ± 1.9 mm at A point and was significantly greater than the 0.9 ± 1.1 mm in the RME group. The overall amount of maxillary advancement in the Alt-RAMEC group was 5.8 ± 2.3 mm at A point. The clinical and cephalometric results are shown in Fig. 30.4. The protraction results remained stable without significant relapse after 2 years (5.8 ± 2.3 vs. 5.7 ± 3.0 mm).

The protocol of Alt-RAMEC displaces the maxilla anteriorly twice and facilitates maxillary protraction three times better than a single course of RME. This indirectly proves that the protocol of Alt-RAMEC disarticulates circumaxillary sutures better than a single course of rapid maxillary expansion. Because the circumaxillary sutures are protracted three times faster than usual, the protraction results could be an orthopedic process of "sutural protraction osteogenesis," which is similar but less vigorous than sutural expansion distraction osteogenesis. The maxillary protraction by using the double-hinged expander, repetitive weekly protocol of Alt-RAMEC, and the intraoral protraction springs is effective with stable results at 2 year follow-up.

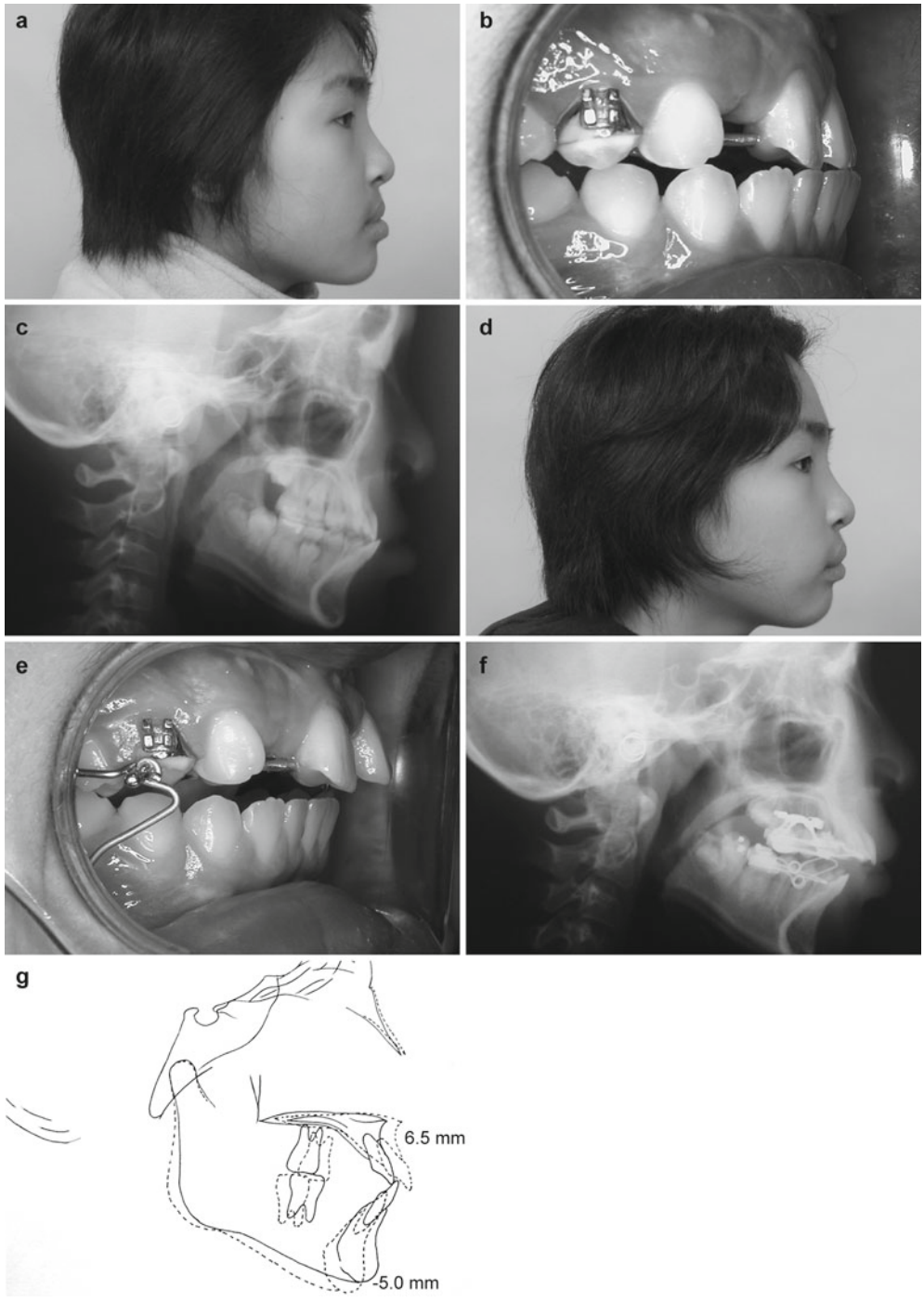

Fig. 30.4 (**a**–**g**) The clinical results of effective maxillary orthopedic protraction in a 12-year-old bilateral cleft patient. The treatment was 9 weeks of Alt-RAMEC, 1 month of active maxillary protraction, and 3 months of maintenance. The amount of maxillary protraction at A point was 6.5 mm and the mandible rotated downward and backward for 5 mm. (**a**–**c**) The lateral facial profile, occlusion, and cephalogram before treatment. (**d**–**f**) The lateral facial profile, occlusion, and cephalogram after treatment. (**g**) Cephalometric superimposition before and after treatment

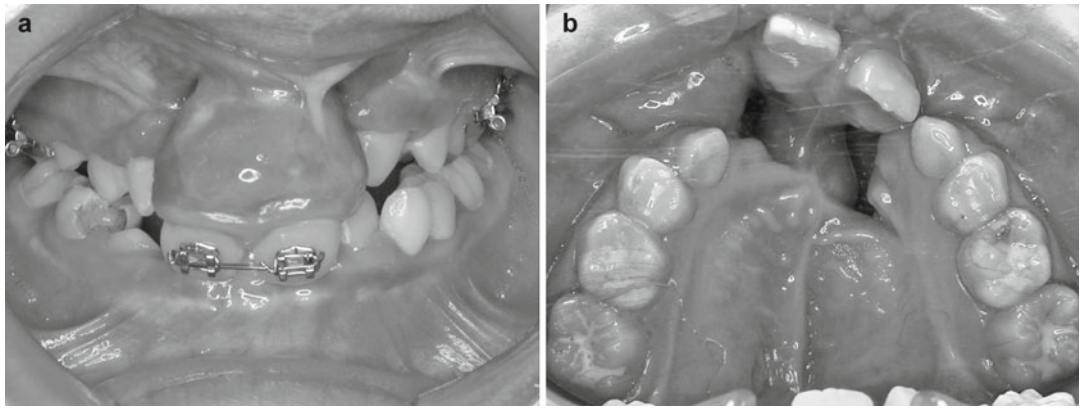

Fig. 30.5 (**a**, **b**) Two of the most common premaxillary deformities in bilateral cleft lip and palate. (**a**) A prominent and downward displaced premaxilla with or without wide alveolar clefts. (**b**) A laterally displaced premaxilla with a wide alveolar cleft on one side

To obtain good stability, our clinical experience revealed that the timing for effective maxillary orthopedic protraction is around the onset of puberty (age 11–13). Early treatment is not recommended because of the unpredictable mandibular growth. The onset of puberty could be evaluated by using vertebral bone age on cephalogram (Mito et al. 2003; Franchi et al. 2000).

30.3 Orthopedic Management of a Downward Displaced Premaxilla in Bilateral Cleft Patients

In growing bilateral cleft patients, one of the most common premaxillary deformities is a prominent and downward displaced premaxilla with or without wide alveolar clefts (Fig. 30.5a). The vertical discrepancy between the premaxilla and buccal segments results in an unaesthetic "equine appearance," anterior deep overbite, and also makes the alveolar bone grafting difficult or impossible (Hayward 1983).

In a cephalometric study, Liou et al. inferred that the downward displaced premaxilla is not because of overgrowth of the premaxilla or overeruption of the maxillary incisors (Liou et al. 2004). The premaxilla might have been downward distorted in the early age of life after the primary surgical repairs. Several studies indicated that the prominence of premaxilla becomes less pronounced during the growth period and gradually resolves into the adulthood (Pruzansky 1955; Harvold 1961; Narula and Ross 1970; Friede and Pruzansky 1972; Vargervik 1983; Trotman and Ross 1993). It is recommended that the premaxillary deformities be left untreated until adulthood (Friede and Pruzansky 1985). However, to leave the deformities unsolved until adulthood may complicate the alveolar bone grafting and impact the young patients psychosocially.

The contemporary surgical interventions of the downward displaced premaxilla in bilateral cleft patients include removal of the primary maxillary incisors in order to improve the appearance and temporarily reduce the dentoalveolar prominence (Vargervik 1983), and surgical reposition of the premaxilla in conjunction with alveolar bone grafting (Harvold 1961; Freihofer et al. 1991; Bardach et al. 1992; Heidbuchel et al. 1993; Iino et al. 1998). However, the premaxilla in the growing subjects having surgical repositioning grows at a very low rate and becomes retrusive progressively in the older age (Vargervik 1983; Smahel 1984).

30.3.1 Premaxillary Orthopedic Intrusion

A better treatment modality, therefore, is to reposition the premaxilla and approximate the alveolar cleft nonsurgically and orthopedically so that

the growth of premaxilla would not be disturbed and the alveolar bone graft could be performed successfully. A new nonsurgical technique called premaxillary orthopedic intrusion has been developed for correcting the downward displaced premaxilla in growing patients with bilateral cleft (Liou and Chen 2003; Liou et al. 2004).

30.3.1.1 Orthodontic Preparation

Before initiating orthopedic intrusion of premaxilla, the maxillary incisors are well aligned orthodontically so that a segment of 0.016 by 0.022 stainless steel arch wire can be placed. The maxillary first permanent molars or the second primary molars are banded, and the band is welded with an orthodontic triple tube. The triple tubes are for holding the orthopedic intrusion device. A fan-type rapid maxillary expander, if necessary, could be placed for repositioning the collapsed buccal segments. A transpalatal arch and/or a segment of stainless steel arch wire (0.016 by 0.022) are placed on the buccal teeth for consolidating the buccal segments as an anchor unit for the orthopedic intrusion of premaxilla.

30.3.1.2 Device for Premaxillary Orthopedic Intrusion

No headgear, extraoral device, or surgery is used for the intrusion. The device for orthopedic intrusion of the downward displaced premaxilla is a pair of tooth-borne distraction devices (Fig. 30.6a). The device anchors on the maxillary buccal teeth and delivers an intermittent intrusion force to the premaxilla through the maxillary incisors. The U-shaped connector of the device is inserted into the headgear tube on the molar band and secured with a 0.012-in. ligature wire. The extension arm of the device is secured with 0.012-in. ligature wires onto the segmental arch wire on the central incisors (Fig. 30.6b–e).

On both sides, the devices are activated 0.3 mm/day until the premaxilla has been upward repositioned to the desired position. The devices are then maintained for another 3 months before their removal. After removal of the intrusion devices, an orthodontic arch wire is placed for further maintenance, and alveolar bone grafting is then performed within 3 months.

30.3.1.3 Treatment Results of Premaxillary Orthopedic Intrusion

The treatment results have been evaluated in a cephalometric study on ten consecutive cases (Liou et al. 2004). The orthopedic intrusion of premaxilla was completed within 4 weeks in all the patients. After the intrusion, the occlusal planes and gingival lines of the premaxilla and buccal segments were leveled clinically. The facial appearance during resting and smiling improved and had no "equine appearance." The clinical results are shown in Fig. 30.7.

The cephalometric study revealed that there was no significant vertical movement of the maxillary buccal segments, and the premaxilla was significantly intruded (Fig. 30.7d). The nasal bone was also significantly brought forward and upward at the nasal tip, but its amount was significantly less than the orthopedic intrusion of the premaxilla.

The correction of the premaxillary deformity mainly came from three aspects: orthopedic intrusion of the premaxilla, dental intrusion of the maxillary incisors, and shortening of the premaxillary dentoalveolar height. The orthopedic intrusion of the premaxilla (3.0 mm) and dental intrusion of the maxillary incisors (3.5 mm) gave a total amount of 6.5 mm upward repositioning at the tip of the maxillary incisor. The shortening of the premaxillary dentoalveolar height (3.5 mm) could be due to the dental intrusion of the maxillary incisors, which was expected when a tooth-borne distraction device was used. The correction was 46 % (3.0/6.5) orthopedic effect and 54 % (3.0/6.5) dentoalveolar effect.

The orthopedic intrusion of the premaxilla remains relatively stable in the 1-year observation after treatment. However, the dental intrusion of maxillary incisors seems unstable. The dental relapse of the maxillary incisors tends to elongate the premaxillary dentoalveolar height and compromise the result of premaxillary orthopedic intrusion. For this reason, it is recommended to maintain the dental intrusion with an orthodontic arch wire and perform alveolar bone grafting shortly after removal of the intrusion devices (Fig. 30.7c).

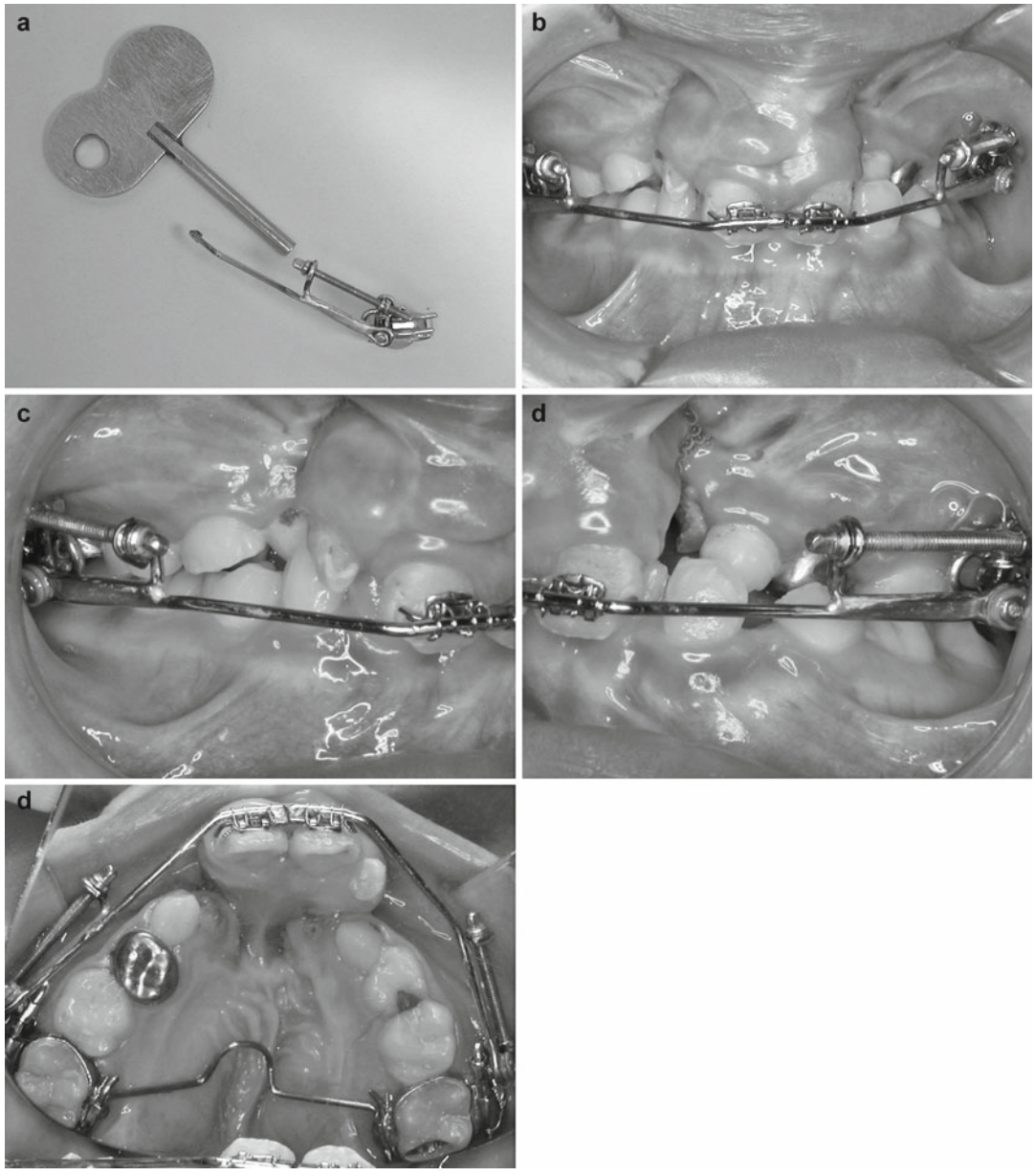

Fig. 30.6 (**a**–**e**) The device for premaxillary orthopedic intrusion (**a**) and the assembly of the device in a clinical case (**b**–**e**)

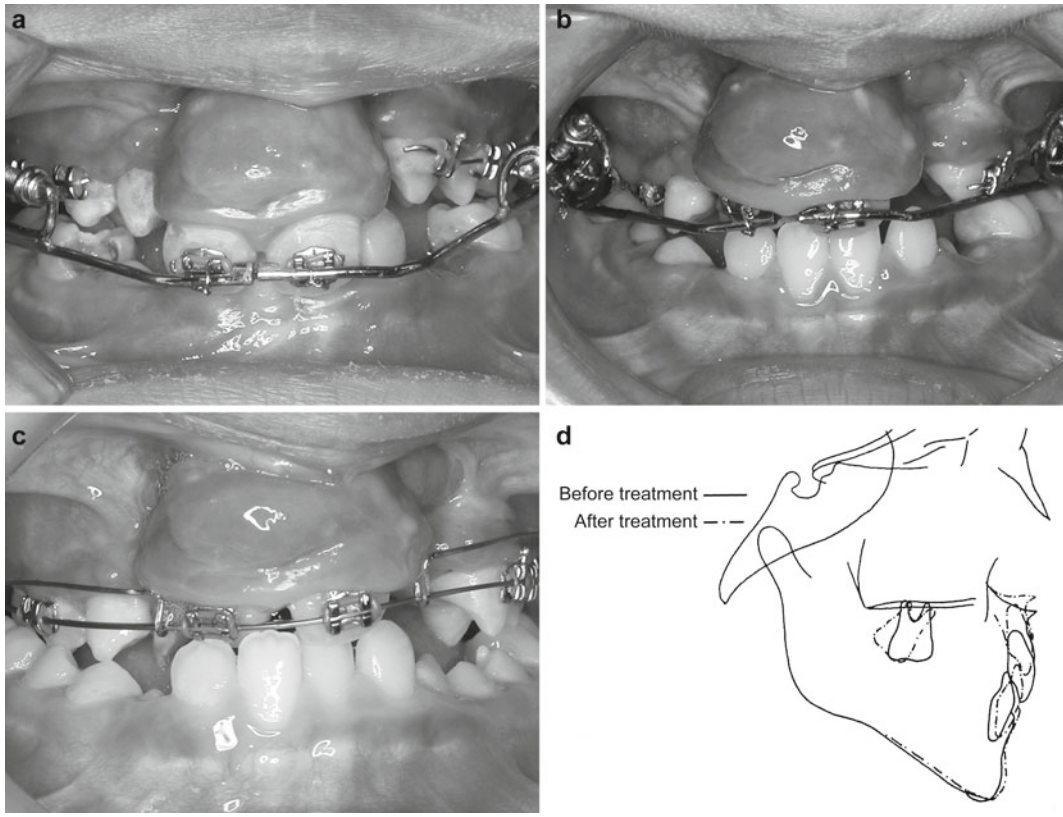

Fig. 30.7 (**a**–**d**) The premaxillary orthopedic intrusion. The equine appearance of the premaxilla prior treatment (**a**), after 1 month of orthopedic intrusion (**b**), 5 months after the orthopedic intrusion (**c**), and cephalometric superimposition before and after premaxillary orthopedic intrusion (**d**)

30.3.1.4 Mechanisms of Premaxillary Orthopedic Intrusion

The possible events that may occur during orthopedic intrusion of premaxilla include mechanical upward displacement of the premaxilla and vomeronasal septum complex, sutural contraction osteogenesis (Castello et al. 2000) and/or osteolysis (Kawakami et al. 1996) in the vomero-premaxillary suture, or bending/remodeling of the vomeronasal septum complex:

1. Mechanical upward displacement without bending/remodeling of the vomeronasal septum complex and no sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture
2. Sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture without bending/remodeling/mechanical upward displacement of the vomeronasal septum complex
3. Bending/remodeling without mechanical upward displacement of the vomeronasal septum complex and no sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture
4. Any combinations of 1, 2, or 3

By the analysis on the lateral and posteroanterior cephalometric radiographs, it was implied what occurred during the orthopedic intrusion of premaxilla was mostly similar to the sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture combined with slightly mechanical upward displacement of the vomeronasal septum complex and nasal bones (Fig. 30.8).

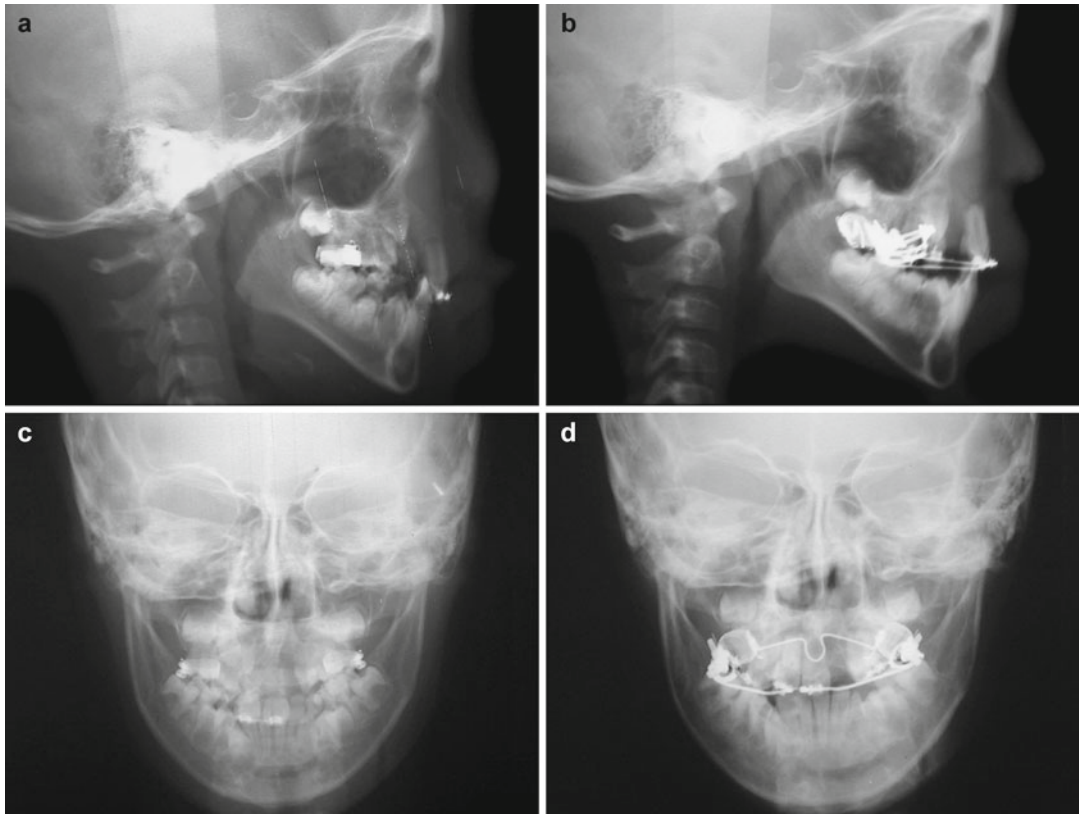

Fig. 30.8 (**a**–**d**) The cephalometric radiograms of the same patient in Fig. 30.7. (**a**, **b**) The lateral cephalogram before and after premaxillary orthopedic intrusion. (**c**, **d**) The posteroanterior cephalogram before and after premaxillary orthopedic intrusion. Note the premaxilla and incisors were intruded without further deviation of the vomer bone and nasal septum

30.4 Orthopedic Management of a Laterally Displaced Premaxilla in Bilateral Cleft Patients

In growing bilateral cleft patients, another common premaxillary deformity is a laterally displaced premaxilla with a wide alveolar cleft on one side (Fig. 30.5b). The laterally displaced premaxilla results in an unaesthetic appearance with a skewed premaxilla and dental midline and uneven width of the alveolar cleft beside the premaxilla.

The alveolar bone grafting and closure of oronasal fistula could be difficult without surgical repositioning the laterally displaced premaxilla. However, the premaxilla in growing subjects having surgical repositioning grows at a very low rate and becomes retrusive progressively as they age (Vargervik 1983; Smahel 1984).

30.4.1 Premaxillary Orthopedic Medial Repositioning

A better treatment modality, therefore, is to reposition the premaxilla and approximate the alveolar cleft nonsurgically and orthopedically so that the growth of premaxilla would not be disturbed and the alveolar bone graft could be performed successfully. A new nonsurgical technique called premaxillary orthopedic medial repositioning has been developed for correcting the laterally displaced premaxilla in growing patients with bilateral cleft (Liou and Chen 2003).

30.4.1.1 Orthodontic Preparation

Similar to the clinical procedures for the premaxillary orthopedic intrusion, the entire maxillary dentition is aligned orthodontically so that a 0.016 by 0.022 stainless steel arch wire could be placed before the orthopedic repositioning. The maxillary first permanent molars are banded, and the band is welded with an orthodontic triple tube. For the anchorage, a transpalatal arch is placed for consolidating both of the buccal segments as an anchor unit for the orthopedic repositioning of premaxilla.

30.4.1.2 Device for Premaxillary Orthopedic Medial Repositioning

No headgear, extraoral device, or surgery is used for the repositioning. The device for orthopedic medial repositioning is an intraoral tooth-borne distraction device (Fig. 30.9). It consists of a body, a screw, and an extension arm. There are two bolts on the body for holding the screw. The fixed bolt is attached on the front of the body for holding the head of the screw. The sliding bolt rides and slides on the sliding bar of the body for holding the tail of the screw. The U-shaped connector at distal end of the body is inserted into the headgear tube on the molar band and secured with a 0.012-in. ligature wire. The extension arm is adjusted to hook distally to the maxillary incisors so that the premaxilla is being pushed toward the midline when the device is activated. The arch wire is a track guiding the premaxilla to slide in a linear curvature direction during the orthopedic repositioning.

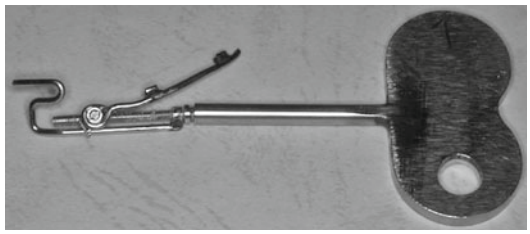

Fig. 30.9 The device for premaxillary orthopedic medial repositioning

The screw is activated 0.3 mm/day until the maxillary dental midline has been overcorrected for 2–3 mm. The device is then maintained for another 3 months before its removal. After the removal, orthodontic elastics or ligature wire is placed for further maintenance before the next treatment on the alveolar cleft.

30.4.1.3 Treatment Results of Premaxillary Orthopedic Medial Repositioning

The treatment results have been evaluated in a cephalometric study on four consecutive cases (Liou and Chen 2003). The clinical results are shown in Fig. 30.10. The age of the patients were 9–12 years old at the time of treatment. Before the treatment, the angular deviation of premaxilla and nasal septum ranged from 15° to 25°, and the linear deviation of dental midline ranged from 5 to 8 mm as measured on the posteroanterior cephalograms.

The orthopedic medial repositioning of premaxilla was completed within 2–3 weeks in all the patients. All the patients tolerated the device well, and no pain was reported by any of the patients. The maxillary dental midline was overcorrected for 2–3 mm. The overall correction of the dental midline ranged from 7 to 11 mm. During the maintenance period (3 months), the premaxilla and maxillary dental midline gradually moved back toward the mandibular dental midline. The smile facial appearance was improved, and the premaxilla and dental midline were repositioned on the midline.

30.4.1.4 Mechanisms of Premaxillary Orthopedic Medial Repositioning

The posteroanterior cephalometric and occlusal radiographs before and 3 months after the repositioning were evaluated. It was revealed that both the deviated nasal septum and premaxilla were all straightened (Fig. 30.11). The straightening of the nasal septum and premaxilla could be due to bone bending and remodeling of the vomer bone. The angular correction of the premaxilla and

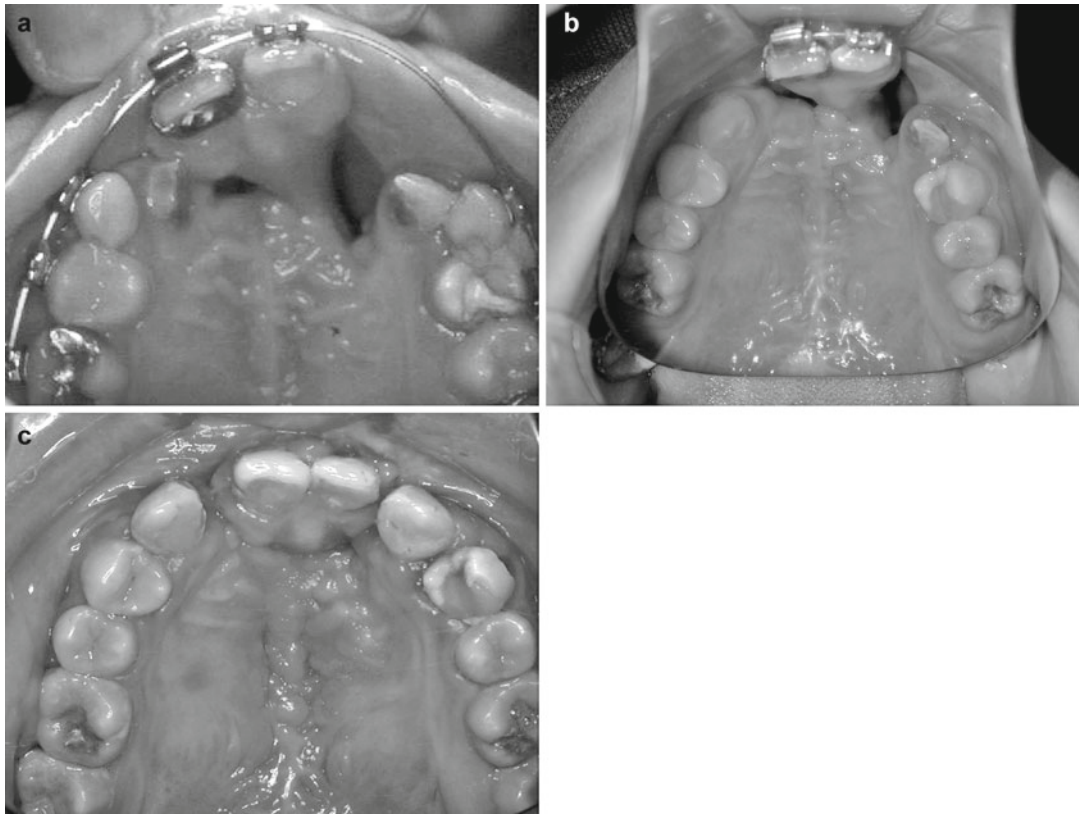

Fig. 30.10 (**a**–**c**) Premaxillary orthopedic medial repositioning. The skewed premaxilla and wide alveolar cleft before the orthopedic repositioning (**a**), after 3 weeks of orthopedic repositioning (**b**), and after effective maxillary orthopedic protraction of the lateral segments for minimizing the alveolar cleft and alveolar bone graft (**c**)

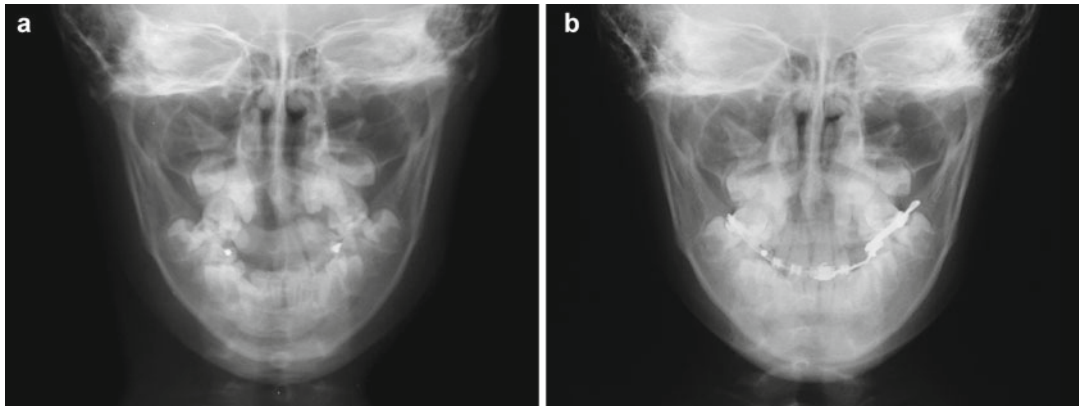

Fig. 30.11 (**a**, **b**) The posteroanterior cephalograms reveal straightening of the vomer bone after premaxillary orthopedic medial repositioning (**a** before; **b** after medial repositioning)

nasal septum ranged from 10° to 20°, and the linear correction of the premaxilla ranged from 5 to 10 mm, as measured on the radiographs.

30.5 Managements of a Wide Alveolar Cleft and Fistula

Although premaxillary orthopedic intrusion or orthopedic medial repositioning is able to correct the downward or laterally displaced premaxilla, the alveolar cleft might still be too wide to be bone grafted. Using oral mucosa at a distant site, such as rotation advancement flap of buccal mucosa or tongue flap (Jackson 1972), could close a wide alveolar cleft. By such approach, the alveolar cleft is not bone grafted and the maxillary segments are not approximated. Neither the buccal mucosa nor the tongue mucosa is a good substitute for the attached gingiva. Attached gingiva is essential for the subsequent tooth eruption, orthodontic tooth movement, or dental prostheses after alveolar bone grafting and closure of fistula (Jackson 1972; Boyne 1974).

Another approach is to approximate the alveolar cleft through LeFort I osteotomy and advancement so that both the soft and bony tissue could be well approximated and bone grafted (Posnick and Tompson 1995). However, LeFort I advancement might disturb the growth of maxilla in young patients (Nanda et al. 1983).

30.5.1 Protocol for Approximating a Wide Alveolar Cleft

We have developed a protocol for minimizing or approximating a wide alveolar cleft (Liou and Chen 2003):

1. For an alveolar cleft less than a tooth width, effective maxillary orthopedic protraction of the maxillary lateral segments and alveolar bone grafting is the treatment of choice.
2. For an alveolar cleft wider than a tooth width, interdental distraction osteogenesis (Levrini and Filippi 1999) and alveolar bone grafting or gingivoperiosteoplasty is the treatment of choice.

30.5.2 Minimize Alveolar Cleft by Effective Maxillary Orthopedic Protraction

The lateral segments of maxilla are orthopedically protracted toward the premaxilla to minimize the alveolar cleft. The rationales and mechanisms are similar to entire maxillary protraction by using effective maxillary orthopedic protraction. The total treatment protocol is 6 months, including 8–9 weeks of Alt-RAMEC followed by 4 months of orthopedic protraction of the lateral segments of maxilla. The expander is a double-hinged rapid maxillary expander, and the protraction appliance is the intraoral maxillary protraction springs.

30.5.2.1 Clinical Procedures for Effective Maxillary Orthopedic Protraction of Lateral Segments of Maxilla

Maxillary molars and primary canines (or first premolars) are banded, and maxillary impression is taken for fabricating the double-hinged rapid maxillary expander. The expander is oriented perpendicular to the central incisors. The expander anchors only on lateral segments of maxilla, and there is no extension arm on the premaxilla. The inner surfaces of the bands are sandblasted before cementation. One day after the cementation, the double-hinged expander is activated according to the protocol of Alt-RAMEC and then is left in place for orthopedic protraction of the lateral segments of maxilla. A 0.036-in. b-nickel-titanium mandibular lingual holding arch with built-in lingual crown torque is used for splinting the mandibular dentition as an anchor unit for the orthopedic protraction. Patients are seen every 4 weeks for adjusting or replacing the intraoral maxillary protraction springs when they are distorted or broken. The expander and the springs are then removed at the end of the 6th month.

30.5.2.2 Treatment Results

Three of the four patients who had premaxillary orthopedic medial repositioning received the treatment. The treatment results were evaluated clinically and cephalometrically (Liou and Chen 2003).

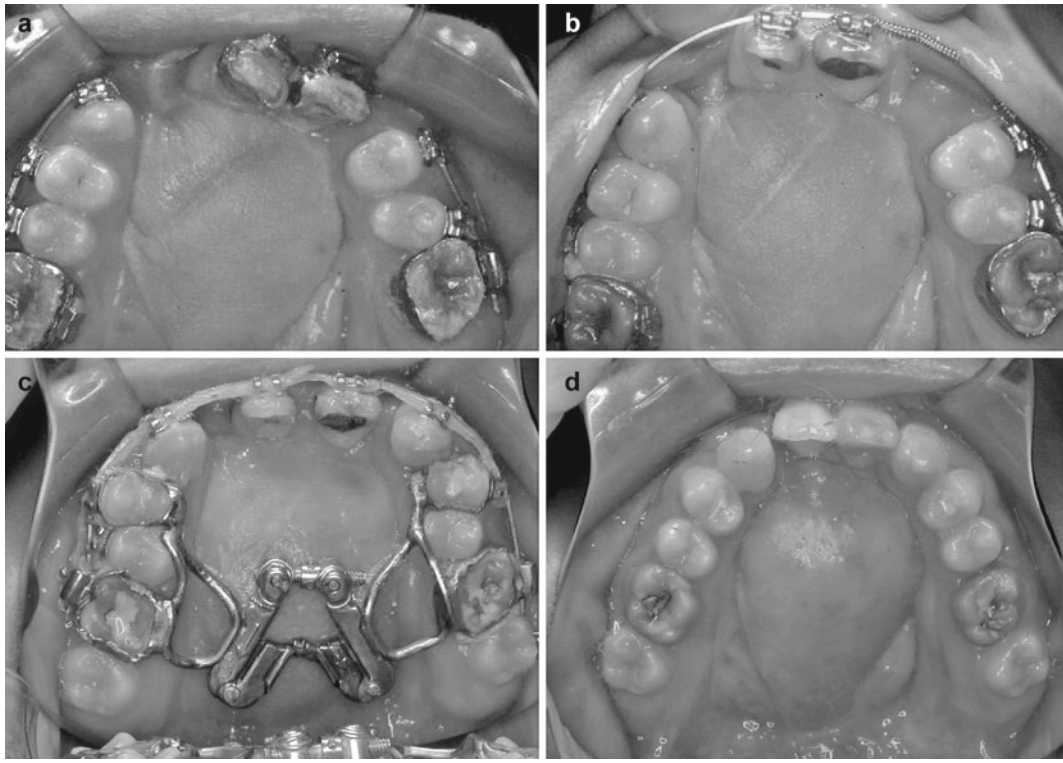

Fig. 30.12 (**a**–**d**) Minimize alveolar cleft less than a tooth width. The skewed premaxilla and alveolar cleft before the orthopedic repositioning (**a**), after 3 weeks of orthopedic medial repositioning (**b**), during effective maxillary orthopedic protraction of the lateral segments (**c**), and after the orthopedic protraction and alveolar bone graft (**d**)

All of the alveolar clefts were successfully minimized, and the gingival and mucosal tissue beside the alveolar clefts was approximated after the orthopedic protraction (Fig. 30.12). All of the patients received orthodontic treatment after removal of the expander and protraction springs. The alveolar bone grafting was then performed shortly after the maxillary dentition was well aligned.

On the cephalometric evaluation, the lateral segments of maxilla were protracted for 2–3 mm. The buccal teeth on the maxillary lateral segments were also moved forward for 2–3 mm. The alveolar clefts were all minimized to 2–3 mm as measured on the radiographs. No retrusion or retraction of the premaxillae was observed. The overjet at anterior teeth was maintained positive throughout the treatment.

The maxillary lateral segment orthopedic protraction by Alt-RAMEC is an effective nonsurgical technique for minimizing the alveolar cleft in patients with BCLP. This technique is much less invasive relative to the surgical repositioning of premaxilla. The treatment effects are both the orthopedic protraction of the maxillary lateral segments and forward movement of the buccal teeth.

30.5.3 Interdental Distraction Osteogenesis for Approximating Alveolar Cleft Wider Than a Tooth Width

The closure of an alveolar cleft wider than a tooth width is a challenge not only because of the difficulty in complete closure by using local attached gingiva but also the great volume of bone grafting. Interdental distraction osteogene-

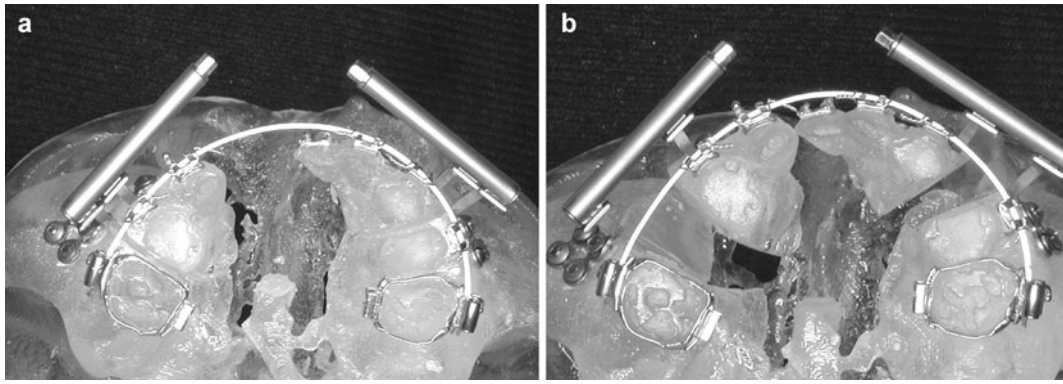

Fig. 30.13 (**a**, **b**) Model illustrations of interdental distraction osteogenesis. The dentoalveolus is osteotomized interdentally (**a**) and transported toward the cleft by the distractor (**b**). The orthodontic arch wire also is a guiding track. The osteotomized dentoalveolus slides on the arch wire in a linear curvature direction

sis (IDO) has been proposed to approximate wide alveolar clefts and to achieve bony union across the clefts without bone grafting, either in unilateral or bilateral cleft (Liou et al. 2000). It is a technique of dentoalveolar lengthening through soft callus distraction osteogenesis. The dentoalveolus is osteotomized interdentally and then transported toward the cleft so that the cleft could be approximated by growing local alveolus and attached gingiva at a distant site to the cleft (Fig. 30.13). Its advantages include eliminating the need for extensive alveolar bone grafting, creating dental space (the regenerate alveolus and attached gingiva) for relieving dental crowding, as well as avoiding worsening the velopharyngeal insufficiency.

30.5.3.1 Presurgical Orthodontic Preparations

All the maxillary teeth are bonded and banded, and then they are well aligned so that a 0.016 by 0.022 (or thicker) stainless steel arch wire could be placed. A transpalatal arch could be placed for consolidating buccal segments as one unit when it is necessary. One of the purposes of orthodontic tooth alignment before interdental distraction osteogenesis is to well align all the maxillary teeth so that the osteotomized dentoalveolus can slide along the arch wire during distraction. Besides the distraction device, the stainless steel arch wire also is a guiding track for interdental distraction osteogenesis. The distraction device has a linear track, but the arch wire guides the osteotomized dentoalveolus to be distracted along the arch wire in a linear curvature direction (Fig. 30.13b).

An interdental space is opened orthodontically at the selected distraction site by using an open coil spring or similar mechanisms. This is to separate the dental roots and increase the thickness of the interdental alveolar bone so that interdental osteotomy could be performed without damaging or stripping the adjacent dental roots.

30.5.3.2 Interdental Distraction Site

Although the selection of distraction site varies regarding different clinical situations of the cleft, there are certain guidelines:

1. The osteotomized segment of dentoalveolus to be distracted should have adequate blood supply from adjacent gingival or oral mucosa and adequate bone volume to sustain a successful distraction osteogenesis. A width of at least two teeth is recommended.
2. The interdental distraction site should be wide enough for interdental osteotomy and to avoid damaging or stripping the adjacent dental roots during interdental osteotomy. At least 3 mm is recommended.
3. At least 1.0 mm thickness of interdental alveolar bone should be preserved beside the adjacent dental roots after interdental osteotomy. Inadequate thickness could result in severe

periodontal problems, such as exposure of the adjacent dental roots and insufficient volume and height of the regenerate.

4. The interdental distraction site should have enough attached gingiva for a primary closure. Exposure of the osteotomy site would result in wound infection, sequestrae, and distraction failure.

30.5.3.3 Surgical Procedures

The surgical procedures are performed under naso-endotracheal anesthesia. A horizontal intraoral incision is made along the buccal vestibule of the maxilla. Superior based mucoperiosteal flaps are reflected, exposing the site of horizontal maxillary osteotomy on the buccal side. A vertical mucoperiosteal tunnel extending upward from the interdental attached gingiva to the horizontal incision is made to expose the site of vertical interdental osteotomy. Another small incision, on the palatal side, is made inside the gingival sulcus, exposing the site of interdental osteotomy. Care is taken to avoid reflecting or stripping all of the palatal mucosa.

By estimating on the radiographs, the anatomic position of the dental roots, sites of interdental and horizontal maxillary osteotomies, and permanent tooth buds are marked on the alveolar bone with surgical marking pens. Complete horizontal osteotomy is performed with a cutting saw, 3–5 mm away from the dental root apex and tooth buds. Complete interdental osteotomy is then performed with a small round bur and followed by a thin osteotome, cutting through the buccal and palatal cortical plates respectively. As the cutting round bur encounters the cancellous bone, the thin osteotome is then used for cutting carefully through the interdental cancellous bone buccolingually. After the horizontal maxillary and interdental osteotomies, the distal segment of the osteotomized dental arch is completely mobile. The orthodontic arch wire holds the osteotomized segment in place after osteotomies and prevents medial collapse of the segment when the distraction device is placed.

The distraction device is a bone-borne device (Martin & KLS, USA) (Fig. 30.14a). The vertical bars of the bone-borne distractor are bended into the desired shape to avoid any possible compression to the gingival adjacent to the vertical osteotomy sites (Fig. 30.14b). The direction of the distractor needs careful adjustment to ensure a correct vector for bony movement. It also needs great care to avoid any compression of the distractor to the buccal mucosa. The distractor is then fixed to maxilla with unicortical screws. The incisions are irrigated and closed with absorbable sutures (Fig. 30.14c). A very important point in wound closure is closing the attached gingiva over the interdental osteotomy sites to avoid any possible wound separation complicated with dental root exposure in the vertical osteotomy site.

30.5.3.4 Distraction Protocol

The latency period is 7 days. This allows enough time for the formation of soft callus and primary healing of the attached gingiva and oral mucosa. The soft tissue wound healing is critical for the distraction results. Soft tissue wound dehiscence during distraction could result in severe bone resorption, gingival recession, and exposure of the dental root. The distraction device is activated 1 mm/day until both ends of the alveolar cleft has been approximated. After completion of interdental distraction, the device is left in place for 3 months for maintenance and postdistraction orthodontic tooth movement through regenerate.

30.5.3.5 Postdistraction Maintenance and Orthodontic Tooth Movement Through the Regenerate

Two weeks after completion of the interdental distraction, the tooth/teeth right adjacent to the interdental distraction, either the mesial or distal one or both, are moved orthodontically into the regenerate. This is to eliminate the interdental space created by distraction, or to utilize the created space for relieving dental crowding.

Orthodontic elastic chains or nickel-titanium coil springs are used for moving tooth/teeth into the regenerate. They are placed between the moved tooth/teeth and the vertical bars of the bone-borne distraction device (Fig. 30.15). The bone-borne distraction device holds the distracted segment through several bone screws, while the tooth/teeth at the distracted segment are being moved through the regenerate without shortening the width of the regenerate. The bone-borne distraction device is left in place for not only maintenance but also is an anchor for

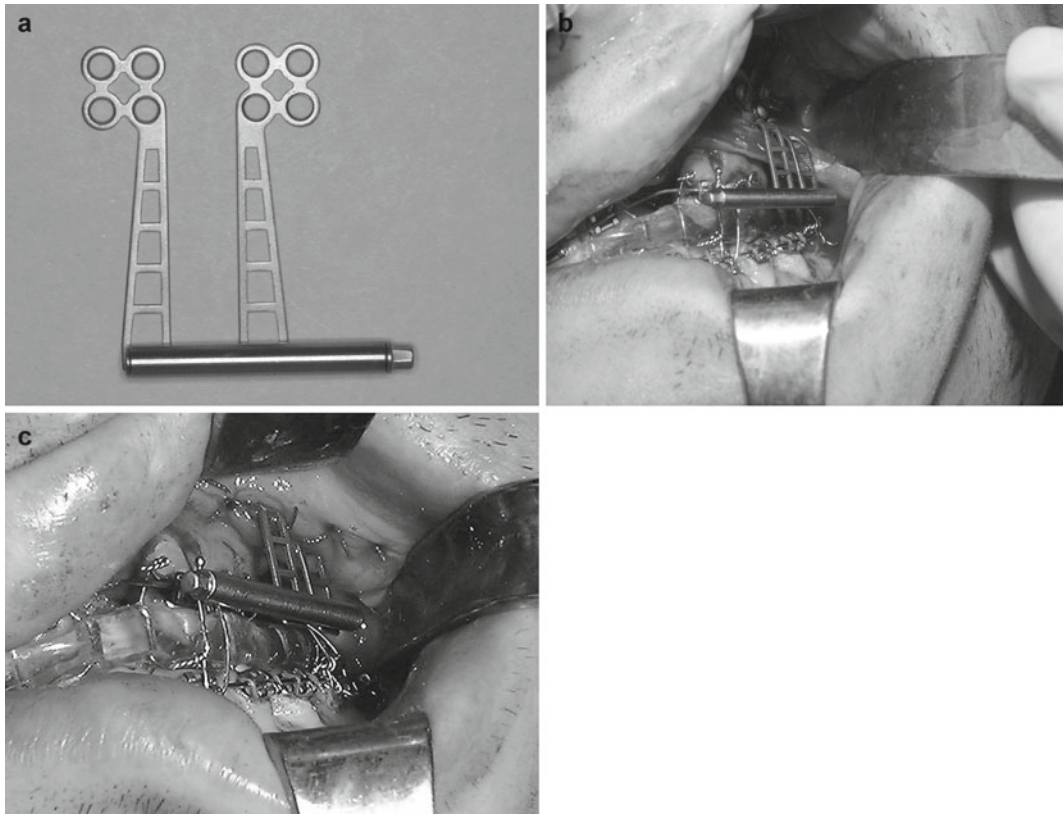

Fig. 30.14 (**a**) The bone-borne distractor for interdental distraction osteogenesis. (**b**, **c**) The vertical bars are bended into a bayonet step to avoid compression to the gingival tissue

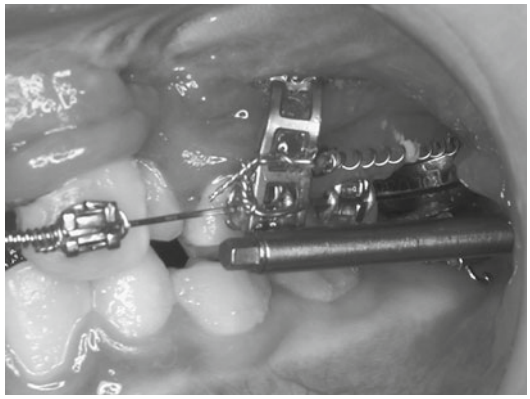

Fig. 30.15 Postdistraction orthodontic tooth movement. Orthodontic nickel-titanium coil spring is attached between the maxillary first molar and the vertical bar of the distractor for moving the first molar into the regenerate

postdistraction orthodontic tooth movement. This is similar to the implant orthodontics that uses bone screws or plates as anchorage for orthodontic tooth movement (Lin and Liou 2003). After the tooth/teeth have moved into the regenerate, the adjacent tooth/teeth are then moved subsequently into the left space. The postdistraction orthodontic tooth movement usually can be completed in 3 months because the regenerate is still soft.

30.5.3.6 Postdistraction Alveolar Bone Grafting or Gingivoperiosteoplasty

After the interdental distraction osteogenesis and postdistraction orthodontic tooth movement, either conventional alveolar bone grafting or gingivoperiosteoplasty can be performed as the final treatment procedure for closing the wide alveolar cleft. Soft tissue approximation of the alveolar

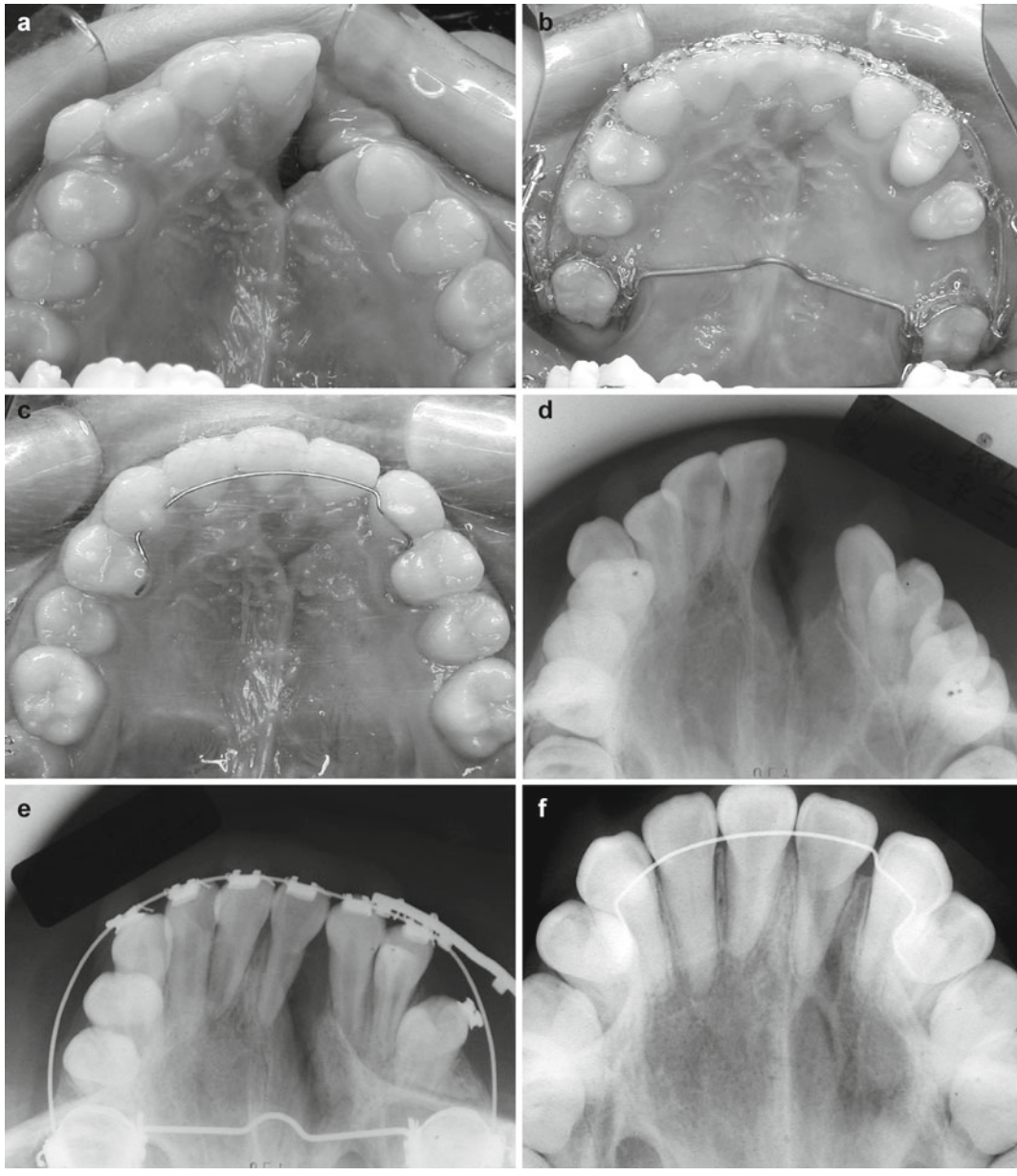

Fig. 30.16 (**a**–**f**) Approximation of a wide alveolar cleft by interdental distraction osteogenesis in a 12-year-old unilateral cleft patient. The alveolar cleft is bone grafted after the distraction. (**a**–**c**) The clinical photos before, immediately after distraction, and 2 years after distraction. (**d**–**f**) The occlusal films before, immediately after distraction, and 2 years after distraction

cleft does not mean bony approximation of the alveolar cleft. Occlusal or periapical film is needed for revealing the remaining width of the bony alveolar cleft:

1. The alveolar bone grafting is performed in cases where the bony gap between the alveolar cleft is still wider than 2 mm after interdental distraction osteogenesis.
2. The gingivoperiosteoplasty is performed in cases where the bony gap between the alveolar cleft is less than 2 mm after the interdental distraction osteogenesis.

30.5.3.7 Treatment Results

Twenty-one patients, including 13 unilateral and 8 bilateral clefts who had interdental distraction osteogenesis for approximating their wide alveolar cleft in 1998–2002 were reviewed for their treatment results (Fig. 30.16). The follow-up period was 4–5 years. There were 29 alveolar clefts and the average width was 10 mm.

The distracted segments of the dental arches were moved almost bodily toward the cleft as revealed on the panoramic and cephalometric radiographs. The average amount of distraction, as measured on the radiographs, was 12 mm with a range of 10–20 mm. The relapse was 0.5 mm in the first 3 months after removal of distraction device.

Clinically, 28 out of 29 alveolar clefts were approximated completely by soft tissue contact after interdental distraction osteogenesis. Thirteen out of the 29 alveolar clefts were successfully bone grafted, and 16 out of the 29 alveolar clefts had gingivoperiosteoplasty that resulted in bony union across alveolar cleft. The 4- to 5-year follow-up revealed the treatment results were stable.

30.6 Summary

In this chapter, we introduced three new orthodontic and orthopedic techniques and one surgical distraction osteogenesis for the management of maxillary deformities in growing unilateral and bilateral cleft patients. These techniques are the effective maxillary orthopedic protraction for correcting a hypoplastic maxilla and minimizing alveolar cleft, premaxillary orthopedic intrusion for correcting a downward displaced premaxilla, premaxillary orthopedic medial repositioning for correcting a lateral displaced premaxilla, and interdental distraction osteogenesis for approximating a wide alveolar cleft. These techniques utilize principles of distraction osteogenesis. The orthopedic approaches could be a form of sutural expansion or protraction osteogenesis, and their treatment effects are mostly orthopedic and partly orthodontic. The interdental distraction is a form of callus distraction osteogenesis. The clinical and radiographic evaluations have revealed their successful applications for solving maxillary deformities in growing cleft patients.

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Preventing Relapse Following Distraction Osteogenesis for the Cleft Midface in Adults

31

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31.1 Preventing Relapse Following Distraction Osteogenesis for the Cleft Midface in Adults

Distraction osteogenesis is an accepted form of treatment in the management of midface deficiency in patients with cleft. However, it is well known that some amount of relapse is inevitable in patients who undergo this procedure (Cho and Kyung 2006). The problems is amplified in some patients because of excessive scarring and fibrosis resulting from previous operations. This chapter aims to discuss protocols to control long-term relapse rates and optimize results. It analyzes the mechanism and rate of relapse in adults and discusses inputs in terms of treatment planning pre- and postoperative orthodontic management, operative and distraction techniques as well as retention strategies to minimize and overcome the tendency to relapse.

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31.1.1 The Dynamics of Cleft Midface Growth and Relapse Following Distraction

As discussed in the previous chapters in this book (Chap. 3), it is an accepted fact that cleft lip and palate repair affects the three-dimensional growth of the maxilla (Ross 1987; Houston et al. 1989; Panula et al. 1993; Figueroa et al. 1999). The first evidence of distraction osteogenesis for maxillary advancement was reported in animals in 1993 (Rachmiel et al. 1993). Distraction osteogenesis of the maxilla to correct the maxillary hypoplasia was first described for children and young adults in 1997 (Polley and Figueroa 1997). Following this, there have been several reports regarding its efficacy, advantages, as well as shortcomings including the high relapse rate (Wang et al. 2005; Kozák et al. 2005; Cheung and Chua 2006; Cheung et al. 2006; Nout et al. 2006).

There are several reports which state 25–70 % of cleft patients have midface retrusion out of which 40 % require surgical treatment for correction of midface deficiency (Mars et al. 1992; Williams et al. 2001). The probable reasons for this are the scarring and fibrosis following surgical trauma and the intrinsic inability of the maxilla to grow normally (Figueroa et al. 1999; Williams and Sandy 2003; Lilja et al. 2006). This results in a midface deficiency with a Class III malocclusion and reverse overjet, which manifests during the initial growth phase of the child and becomes more pronounced as the child

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grows into adolescence. In cleft patients, the anterior nasal spine and the pyriform margins which form the bony platform for the nose are posteriorly placed in relation to the skull base resulting in inadequate projection of the midface and relative prognathism of the mandible. Additionally, because of the nature of the cleft and its repair, the alveolar arches may be collapsed and the teeth may be malpositioned or rotated. Some patients may also have associated problems like poorly repaired lip and palate, multiple palatal surgeries, and velopharyngeal incompetence. All these may contribute to an accelerated and increased deposition of palatal scar tissue, and when compounded further by the surgical insult and the trauma of the distraction process, together, they will add to the quantum and rate of midfacial relapse (Cheung and Chua 2006).

31.1.2 Patient Selection

Our indications for distraction osteogenesis as against conventional orthognathic surgery in cleft patients who have attained skeletal maturity are as follows: (a) Those requiring advancement of 8 mm of the maxilla or more, (b) patients with severe fibrosis of the lip and palate following multiple surgeries and (c) those with a previous pharyngeal flap.

31.1.3 Planning and Assessment

Several of our patients are referred to us for the first time for correction of the midface deficiency, having had several primary and secondary operations previously. Before we begin orthodontic preparation, we try to get the tissues in the best condition possible. Fistulae if they are present are repaired. Alveolar bone grafting (ABG) should be done 6 months before distraction (Krimmel et al. 2001). In bilateral cases, we complete alveolar bone grafting preferably on both sides or at least on one side before taking the patient up for distraction. In unilateral and bilateral patients whose alveolar arch is in crossbite, it is corrected by an expansion appliance, and in selected cases, this can be done simultaneously during midfacial distraction. We prefer to retain the expansion device during the period of distraction and retention, as it gives additional support to the hemi-maxillae and facilitates symmetrical distraction of the segments. We try and preserve the third molar since if it does erupt, it may provide much needed posterior occlusion to the advanced maxilla. It is essential to evaluate and record preoperative speech samples and perform a naso-endoscopy or videofluoroscopy if needed. If corrective surgery for velopharyngeal incompetence (VPI) needs a pharyngeal flap, it is best to defer it after distraction and consolidation as it can have a bearing on the initial relapse. The amount of advancement needed and the vector of distraction were established by mock surgery by the orthodontist.

31.1.4 The Dynamics of Distraction Osteogenesis and Its Relevance to Subsequent Relapse

Distraction is gradual stretching of callus (deformed maxilla) after a horizontal osteotomy of the maxillary bone. The callus responds to gradual stretching over few days by regenerating new bone. This has been shown by Ilizarov in his pathbreaking study on reparative regeneration in dogs (Ilizarov et al. 1969). Several microscopic studies have shown that after about 10 days of distraction (15th post-op day), there is a central zone within the osteotomy with proliferating mesenchymal cells and capillaries resulting from angiogenesis and paracentral zones with large amount of wavy collagenous fibers (Karp et al. 1992; Aronson et al. 1997; Bell and Guerrero 2007). After 15 days of commencing distraction, there is appearance of mineralization, and at 20 days, the trabeculae of the newly formed delicate woven bone are oriented along the distraction lines and become continuous with nondistracted bone. The trabeculae also get rimmed by osteoblasts and are followed by remodeling. Another study with

dispersive x-ray microanalysis has shown progressive mineralization with increase in calcium and phosphorus content occurring from 3 weeks until 1 year (Rachmiel et al. 1998). From these, one may conclude that the regenerated bone has physical and physiological properties unique to it. Unless it is protected during this prolonged consolidation period of nearly a year or longer, it is susceptible to relapse and deformation.

31.1.5 Operative Technique

Standard low Le-Fort I level cuts of the maxillary bone with osteotomies of the nasal septum and the lateral nasal walls and pterygomaxillary disjunction are performed. Minimal amount of mobilization is made, just to ensure that the osteotomy is complete. Extensive down fracture is best avoided since this may cause excessively floating segments which may hinder a bony union (Yamauchi et al. 2006). We prefer an external distraction device (RED II) anchored to the maxilla with 26-G stainless steel (SS) wires. This sufficiently allows the vectors to be adjusted as the distraction proceeds, and patients tolerate it reasonably during the prolonged consolidation phase.

31.2 The Distraction Process

After a latency period of 5 days, distraction is advanced at 0.5 mm twice a day until the required advancement of the midface is achieved. We distract about 20–25 % more than the requirement to allow for the expected relapse (Cheung and Chua 2006). Depending on the education and motivation level of the patient, the distraction can be done either on an inpatient or outpatient basis. We keep the patient under close surveillance (at least twice a week) during the period of distraction.

31.2.1 Orthodontic Consideration and Retention

This is followed by a consolidation period of 2 months during which the RED II device is retained in place. After 6–8 weeks of consolidation is completed, the device is removed and postoperative orthodontics is commenced. The advancement is maintained by retention devices like reverse pull headgear or strategically placed class III elastics to be worn at all times except when going to school or to work for 6 months and only at night for a further period of 6 months (Fig. 31.1a, b). Large distractions of more than 15 mm may benefit from postdistraction surgical stabilization with rigid

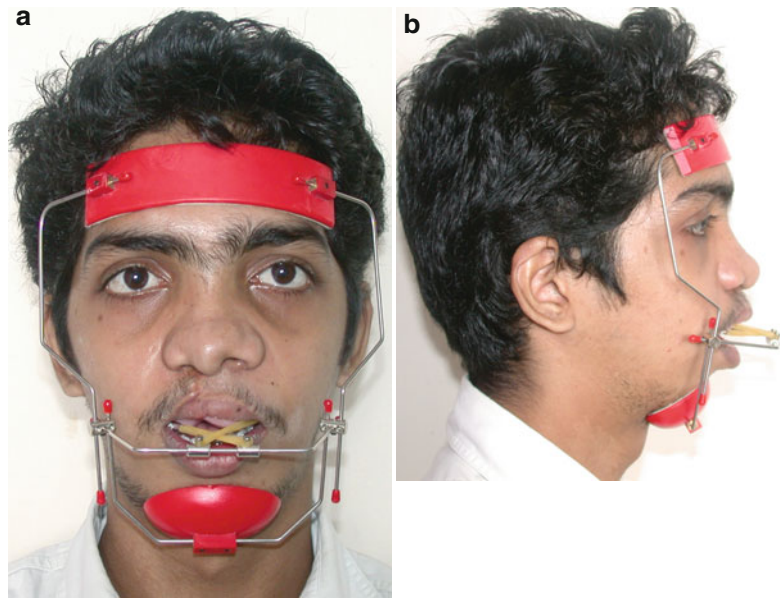

Fig. 31.1 (**a**, **b**) Reverse pull headgear given for retention

internal fixation using plates and screws since the tendency to relapse in these patients is significantly higher (Gulsen et al. 2007).

Minor complications are common during the distraction process and may have a bearing on the subsequent relapse rates (Hussain 2009). They need to be identified and managed as distraction proceeds. Incomplete osteotomy is the commonest cause of failure of distraction and a high rate of relapse. The commonest sites of incomplete osteotomy are the posterior-medial part of the maxillary tuberosity, the vertical plate of the palatine bone in the lateral nasal wall, and incomplete disjunction of the pterygoids. These areas need to be addressed with care while completing the osteotomy. Incomplete osteotomy even while allowing distraction to proceed initially may present as gradual increase in pain over the maxillary bone during the course of distraction. If this is not identified early and re-osteotomy performed, it may result in failure of anchorage of the distraction device and incomplete distraction. In some instances, distraction may continue by deformation of the unosteotomized part of the bone resulting in failure of distraction after some amount of advancement. It may rarely be possible to complete the distraction process, and if done, it will likely lead to a higher relapse rate in the postdistraction period. If one side is not osteotomized, it may result in asymmetrical advancement or increased unilateral relapse resulting in asymmetry. The other causes of asymmetrical distraction are improper adjustment of the device causing asymmetric movement of maxillary segments and dense fibrosis involving one segment. This needs to be identified, and the vector and the rate of distraction have to be adjusted deferentially. Another complication is the loosening of the pins holding the halo frame to the skull.

Consequences of the inevitable relapse are minimized by overcorrection. We routinely overdistract by 20–25 % to compensate for the expected relapse (Cohen et al. 1997; Cheung and Chua 2006). Open bite is another complication which can occur as distraction progresses and may be minimized by varying the vector of distraction. When this is not effective, postoperative orthodontics needs to be provided by using temporary anchorage devices to achieve a stable bite.

31.2.2 Evaluation of Relapse: Materials and Methods

We have evaluated the maxillomandibular skeletal changes including the relapse rates in adults following maxillary distraction with a mean follow-up of 3 years. This study was carried out at our University Hospital Craniofacial Center (Sri Ramachandra University, Chennai, India). Example of a case treated with rigid external distraction is illustrated in Fig. 31.2a–o.

Eighteen adult patients, eight men and ten women, who underwent maxillary advancement by distraction osteogenesis using an external frame distractor were studied for a mean period of 3 years. All the patients were operated by a single surgeon. The mean age was 24.8 years, with ages ranging between 18 years and 34 years. Twelve patients had unilateral cleft lip and palate, and six patients had bilateral cleft lip and palate. The inclusion criteria were as follows: Patients who had maxillary malformation with skeletal Class III malocclusion with a negative overjet ranging from −6 to −15 mm and a cervical maturation indicator 6 or more (CVMI 6 completed).

31.2.3 Serial Cephalometric Evaluation

Lateral cephalograms were used to determine and follow the maxillary position of overtime, taken at intervals: (1) Before commencement of distraction (T1), (2) immediate postdistraction period (T2), (3) 1 year following completion of distraction (T3), and (4) 3 or more years after completion of distraction (T4). T1, T2, and T3 radiographs of a bilateral cleft lip and palate

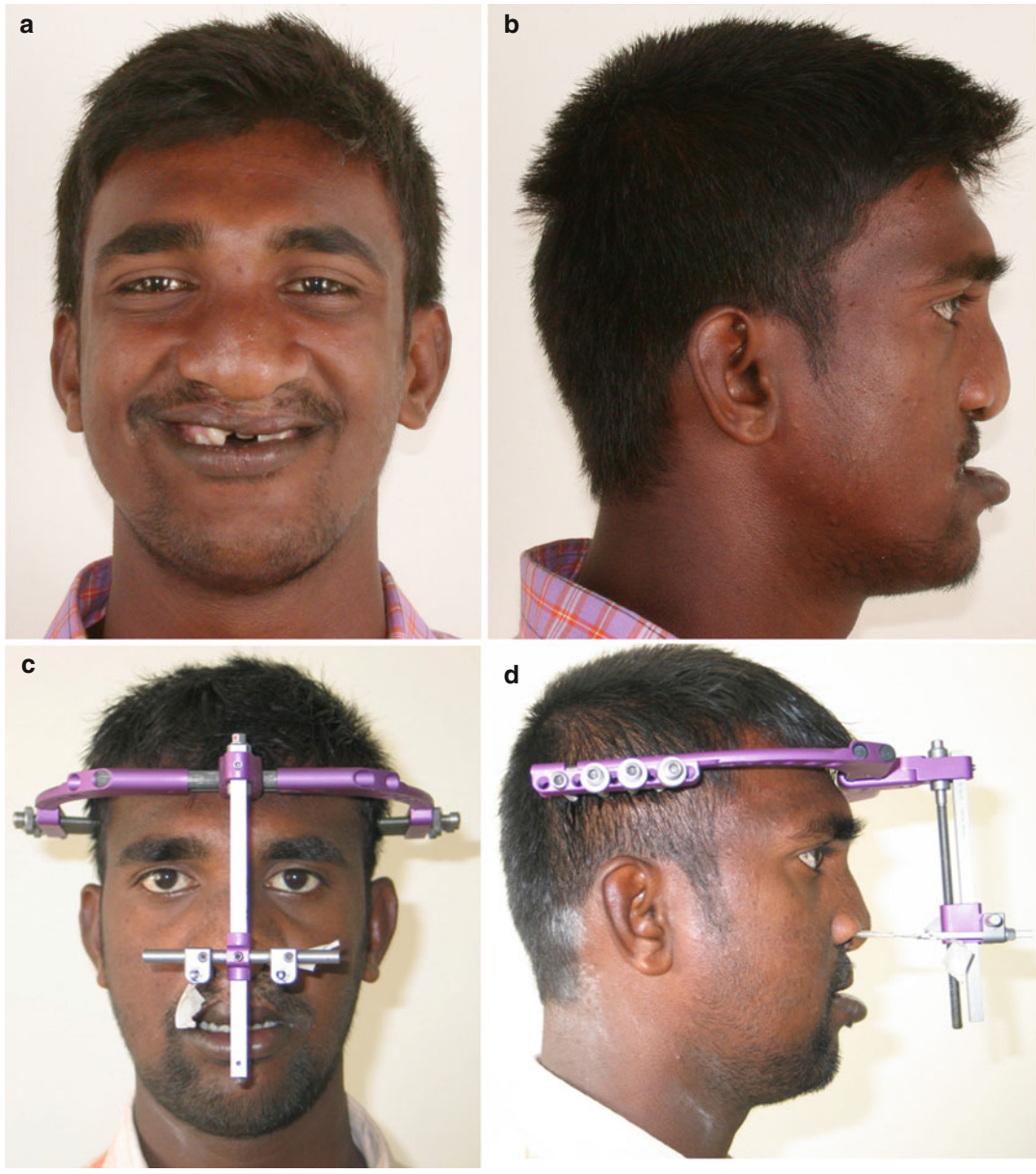

Fig. 31.2 **a–o**. (**a**, **b**) Pretreatment frontal and profile photographs. (**c**, **d**) frontal and profile photographs at the start of distraction

Fig. 31.2 (*continued*) (**e**, **f**) frontal and profile photographs at end of treatment. (**g**–**i**) Pretreatment photographs

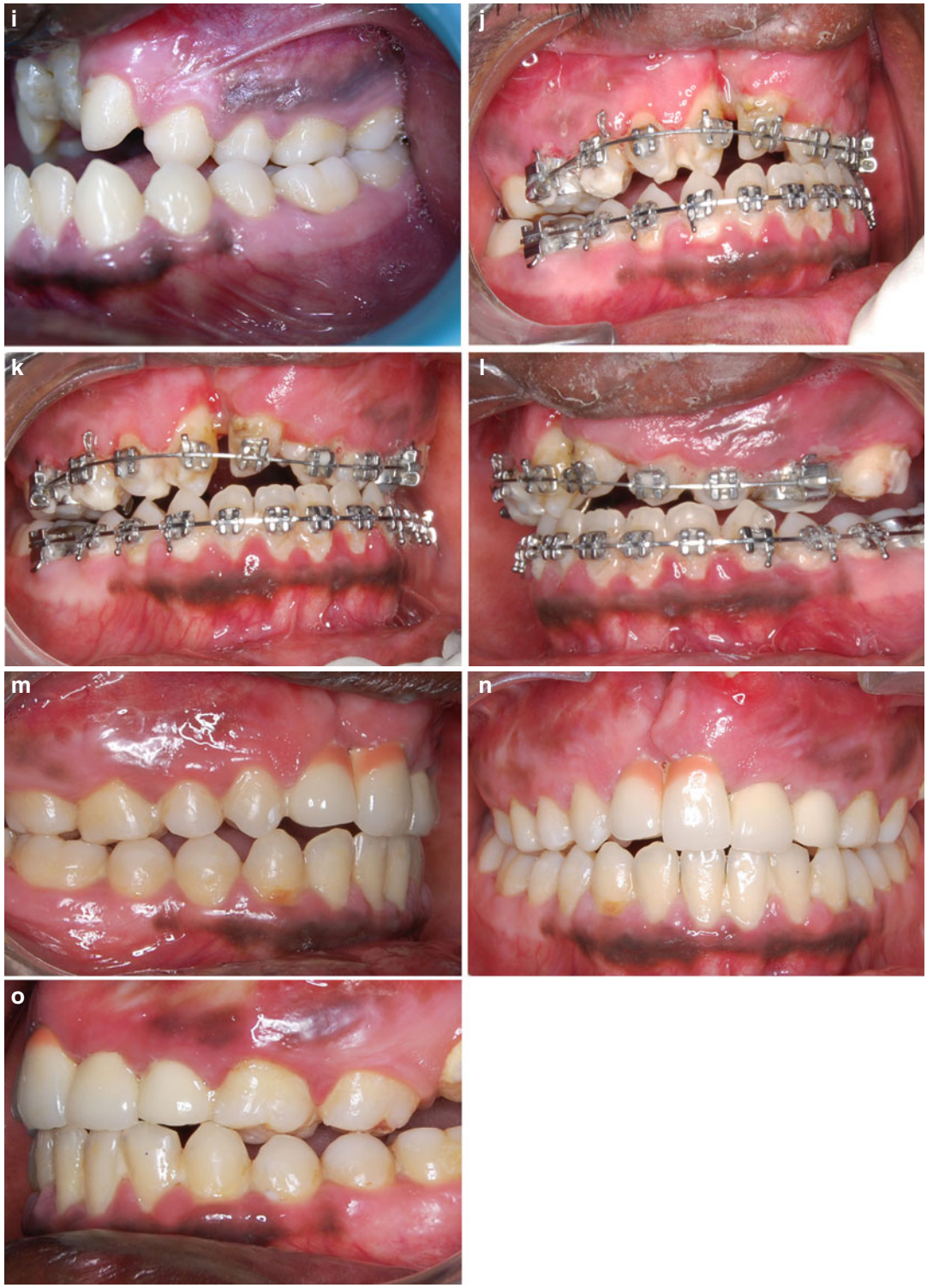

Fig. 31.2 (*continued*) Intraoral photographs. (**j–l**) Post Distraction photograph. (**m–o**) End of treatment

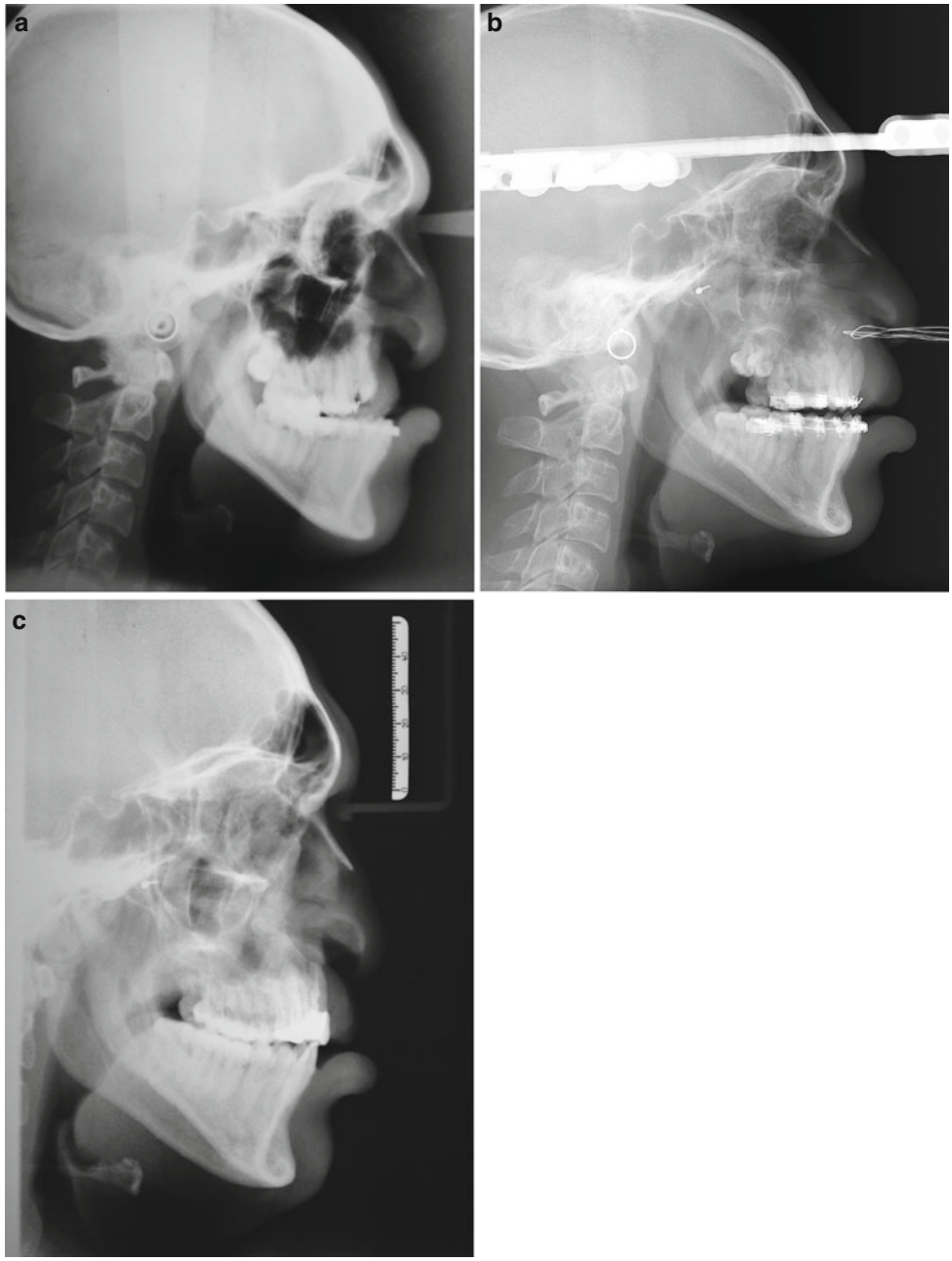

Fig. 31.3 (**a**–**c**) Lateral cephalograms at different intervals as follows. (**a**) Before commencement of distraction, (**b**) immediate post distraction period, and (**c**) three year following completion of distraction

patient treated with rigid external distraction are illustrated in Fig. 31.3a–c.

The radiographs were traced and evaluated by a single experienced investigator. An *X*–*Y* coordinate system was made by drawing a line at an angle of 7° below SN plane for x-axis. Y-axis was drawn perpendicular to this and intersecting it at the sella (Fig. 31.4). The changes in position of ANS, point A, mandibular incisor, and pogonion (Pog) were measured in the *x*- and *y*-axis in cephalograms T1–T4 serially. A mean value was arrived at from all

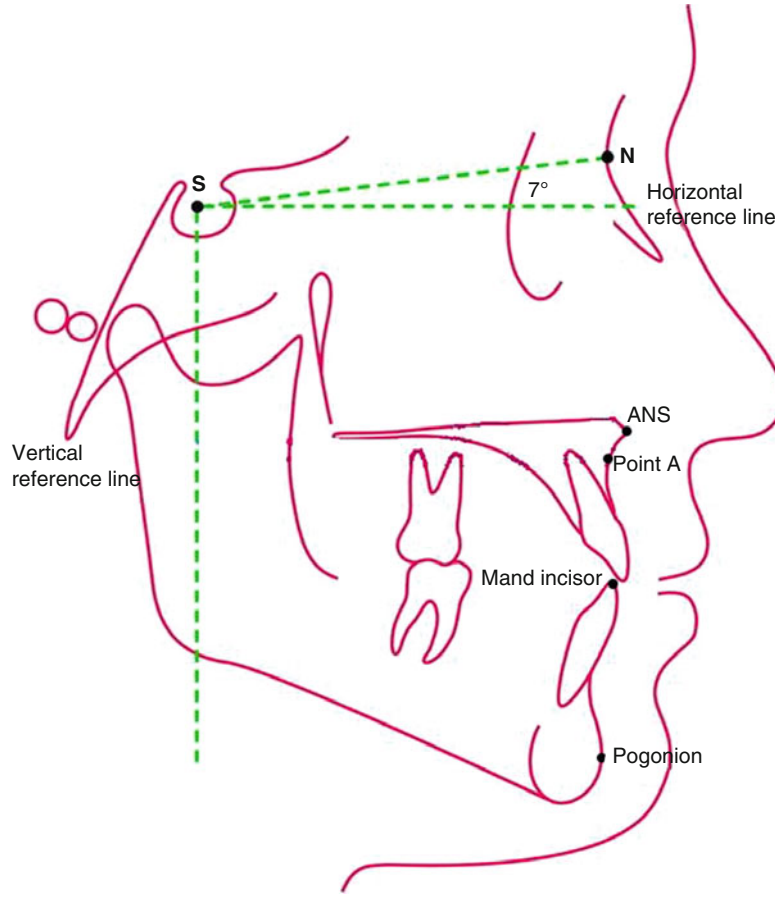

Fig. 31.4 *X–Y* coordinate system was used. The changes in position of ANS, Point A, Mandibular incisor, Pogonion (Pog) were measured on the X and Y axis

individual measurements, and paired sample *t* test was used to compare the difference between T1–T2, T2–T3, and T3–T4.

31.3 Results

Immediately following distraction (T1–T2), the maxilla moved forward at the ANS by 12.4 mm and also downward by 1.56 mm. In the first year following distraction (T2–T3), the maxilla moved backward by 1.5 mm, suggesting significant relapse. Between 1 and 3 (or more) years (T3–T4), maxilla remained stable in horizontal and vertical position. Similar changes with insignificant differences in values were seen in the positions of point A over the same time frame. The position of the tip of mandibular incisor showed significant downward movement of the mandibular incisors immediately after distraction; however, after 1 year following distraction, it moved forward. Similarly, pogonion (Pog) changed as follows: Between T1 and T2, there is backward and downward movement, and between T3 and T4, it remained stable in its new position. Comparison of maxillary positions between bilateral and unilateral cleft lip and palate that was done using the unpaired t tests showed no significant difference between these groups (Fig. 31.5). Comparison between patients who wore a retention device for a year as suggested by the protocol and those who did not wear it or partially complied with the instructions showed that the former group had better retention (Fig. 31.6).

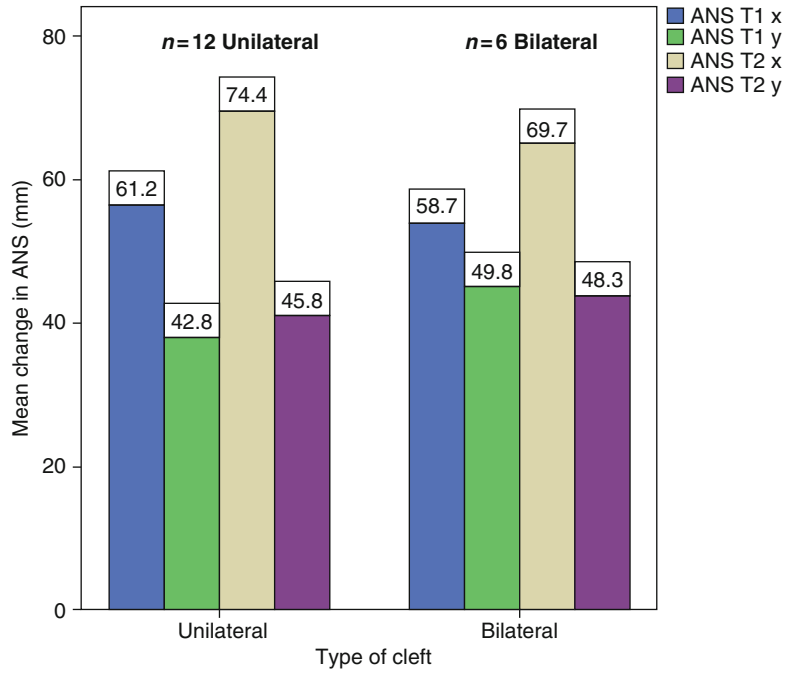

Fig. 31.5 Comparison of maxillary positions between bilateral and unilateral cleft lip and palate

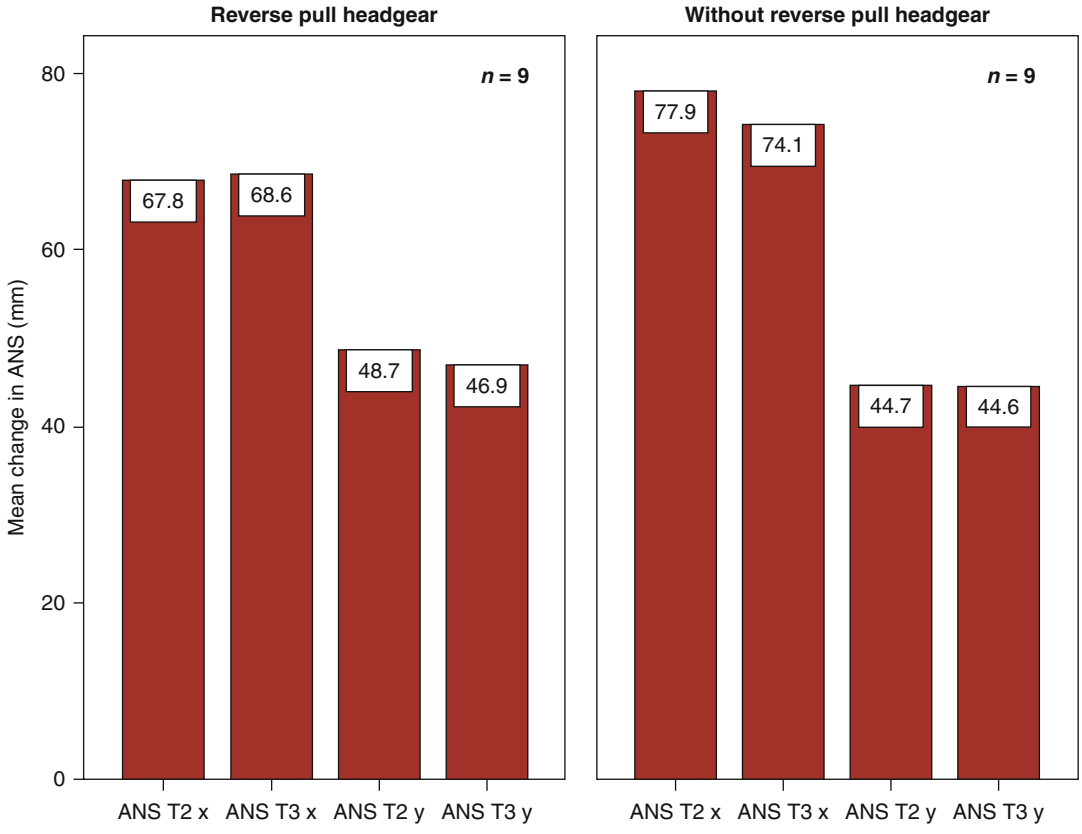

Fig. 31.6 Comparison between patients who wore a retention device and who did not wear retention device

31.4 Discussion

From the above mentioned results, one may reasonably conclude that maximum relapse occurred during the first year following distraction. The various factors influencing this may be categorized into preexisting factors like number and quality of previous operations, preoperative factors, and postoperative management. While the first variable cannot be controlled since it is preexistent, the second and third may be influenced to achieve a more favorable long-term result. Care needs to be taken to ensure that the osteotomy is completed with no remaining bony attachments which may hamper distraction and cause early relapse. At the same time, the segments should not be down fractured or extensively mobilized, since this may lead to fibrous union or nonunion with a subsequent higher rate of relapse (He et al. 2010). The distraction needs to be carried out at the rate of 0.5 mm twice a day as has been established in various studies (Ilizarov et al. 1969; Polley and Figueroa 1997; Cho and Kyung 2006; Bell and Guerrero 2007) with careful control of the vectors. The orthodontic management needs to be closely controlled in the first year since maximum relapse happens during this period (Cheung and Chua 2006). In our series, we have retained the external frame distraction device for a period of 3 months followed by removable retention devices for a further period of one year. These ranged from reverse pull headgear or directional elastics, and the study has shown lesser relapse in patients who complied with this.

Conclusion

Distraction osteogenesis is a powerful tool in the armamentarium of the cleft team for correction of midface recessiveness. However, it should be used judiciously and needs careful patient selection, meticulous planning, and execution. Relapse is inevitable, but its effects on the final result can be minimized. This needs incorporating a degree of overcorrection during planning, precautions and meticulous technique during osteotomy, and careful monitoring during the distraction and consolidation phases with prolonged use of retention devices. The potential complications need to be anticipated, identified early, and corrected during the course of treatment.

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Part XIII

Speech

32 Diagnostic Procedures and Instruments Used in the Assessment and Treatment of Speech

Samuel Berkowitz

32.1 Articulation Tests

An articulation tests is a paper and pencil test used by speech-language pathologists to systematically evaluate the formation and production of the sounds of speech in different contexts in words and sentences. It is used to record the sounds that are produced correctly as well as the errors in speech including omissions, distortions, and substitutions of normal or compensatory sound errors. Systematic evaluation of the articulation of speech is helpful in ensuring complete and consistent analysis of the problems so that effective and efficient treatment can be planned.

Many different types of tests are available, but the most useful tests for patients with cleft palate are those that assess both place and manner of production of speech sounds in the vocal tract. Individuals with cleft palate often produce sounds in a more anterior or more posterior (this is a more frequent error) location in the vocal tract than normal speakers. Careful notation of the types of errors and the probable causative factors sets the stage for determining the method and duration of treatment. These tests also provide a baseline measure or description of speech articulation against which progress, improvement, developmental changes, and treatment outcome can be evaluated. The examiner scores articulation of individual sounds or sound units by listening to and observing the production of these sounds. In some cases, traditional articulation testing alone cannot completely describe the manner or place of the error sounds, and visualization techniques such as multiview videofluoroscopy and nasopharyngoscopy are useful in completing the description and diagnosis of the error(s).

32.2 Rating Scales of Speech Intelligibility and Acceptability and Acceptability

Rating scales are often used by speech-language pathologists and other members of the cleft palate team to score the overall severity of an individual's communication impairment in several categories. Ratings of intelligibility describe how well an individual's speech can be understood by others, whereas ratings of acceptability describe the pleasingness of the sound and appearance of

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speech. These scales usually consist of 5 or 7 points, with 1 being normal and 5 or 7 indicating unintelligible or unacceptable speech. Effective use of these scales is related to rater reliability, and raters must establish and reestablish their reliability on a regular basis when using these scales for clinical or research purposes. These scales are useful as overall measures of severity and treatment outcome; however, they are descriptive in nature and are not useful in isolating causative factors.

32.3 Cephalometrics

Orthodontists and specialists in facial growth have developed the technique of cephalometrics. This is a still sagittal x-ray taken with the patient stabilized in a headholder which positions the head relative to a cranial landmark called the Frankfort horizontal plane. The head is secured by earposts, and the subject's midline is positioned at a constant distance from the x-ray source. The standard position and photon source relationship to the subject provide a means of comparing the measurements from the resulting x-rays. It should be understood that x-ray films provide an image that is a summation of the tissue through which the x-ray beam has passed. If a person had soft palatal closure on one side but not on the other, the midsagittal view would show velopharyngeal closure because the beam would pass through the tissue on the closed side, and the resulting image would not yield information about the opening. The velopharyngeal valve is three dimensional, and its attributes cannot all be captured in the midsagittal view alone. This is a shortcoming of cephalometrics when used for speech purposes. Another problem with the technique is that it is capable of filming only a single point in speech production. Connected discourse cannot be studied by this means, nor is it possible to specify precisely what will be filmed in the course of production of a single sound.

Of particular interest to speech physiologists are the cephalometric studies that have defined structures and growth patterns of pharynx, velum, and lymphatic tissue (King 1952; Rosenberger 1934; Subtelny and Baker 1956; Subtelny 1957). Compositely, these cephalometric studies have provided objective data pertaining to normal skeletal and soft tissue structures and their postural relationships.

This information also has been stated within the complex and important reference of a specified stage of growth and development. The diagnostic value of such normative data becomes obvious in considering the oral examinations speech-language pathologists routinely perform to evaluate speech structures. In such examinations, skeletal, dental, and soft tissue components of the speech apparatus are evaluated.

In addition to the normative data provided, cephalometric analyses have also been undertaken to evaluate skeletal characteristics and growth disturbances in pathologic conditions. Especially valuable for speech-language pathologists are the studies which have defined the intraoral, intranasal, and pharyngeal architecture of individuals with clefts of the lip and palate (Brader 1957; Prusansky 1953; Ricketts 1954; Slaughter and Prusansky 1954; Subtelny 1955). Speech-language pathologists have learned a great deal about morphologic variation in cleft palate patients, whose total rehabilitation, of necessity, includes multiprofessional concerns, from these cephalometric studies.

The fact that cephalometric head plates can be reliably compared has made them attractive to speech-language pathologist's intent on defining physiological differences that result in defective speech production.

There is probably universal agreement that a midsagittal cephalometric view, although useful, yields incomplete information about soft palate and lateral pharyngeal valving. It provides little insight into the location, configuration, or movement of structures off the midsagittal plane. In particular, it offers no information about movement of the lateral pharyngeal walls.

32.4 Cine- and Videofluoroscopy

Cinefluoroscopy (x-rays recorded on motion picture film) and videofluoroscopy (x-rays recorded on videotape) with simultaneous

voice recordings are useful procedures in the evaluation of individuals with cleft palate. The key to development of motion x-rays was the advent of image intensification, which permitted greater contrast with reduced x-ray dosages. Video recording involves lower radiation dosage than does cinefluoroscopy. Videofluoroscopic recording can be done with less radiation than is required for a single still x-ray of the head.

To extract more information from videofluoroscopy, Skolnick (1970) introduced multiview videofluoroscopy, a technique that adds a base view to the traditional lateral and frontal projections.

32.5 Multiview Videofluoroscopy

This fluoroscopic x-ray technique allows visualization of the velopharyngeal port, or valve, during speech in several different planes (Skolnick 1970; Skolnick et al. 1975). This is important because the mechanism of velopharyngeal closure is three dimensional and may involve movement not only of the soft palate but also of the lateral and posterior pharyngeal walls. Descriptions of the technique include the lateral, base, Towne's, frontal, Waters', and oblique views. Not all views are required for a complete examination. The view selected depends on the information needed, the anatomy of the skull, and the anatomy and function of the vocal tract. In most cases, a complete examination consists of three views, including the lateral view, the base or Towne's view, and the frontal, Waters', or oblique view. The examinations are usually recorded and maintained on videotape with audio recording for purposes of interpretation and comparison. The procedure is conducted and interpreted jointly by a radiologist and speech-language pathologist. This contributes to a valid study which includes an adequate speech sample, appropriate visualization of the velopharyngeal valve, and interpretation of the findings which are consistent with the patient's speech. Radiation dosage is kept to a minimum by using videofluoroscopy, as opposed to cinefluoroscopy; coning the x-ray beam to the smallest area; ensuring that the equipment is emitting the minimum radiation necessary to produce the image; and using lead sheets to shield the patient except for the area of interests.

32.5.1 Technique

To enhance the velopharyngeal area, high-density barium is instilled into each nostril using a syringe with a plastic tip. This results in barium coverage of the soft palate, lateral and posterior pharyngeal walls, and posterior aspect of the tongue. A standard speech sample is used during each view. The lateral view is obtained first, with the patient sitting upright. In this view, judgments can be made about the length and thickness of the soft palate, the depth of the pharynx, and the size and location of adenoid and tonsillar tissue. The anterior-posterior excursion of the soft palate and, in some cases, the anterior movement of the pharynx are observed, and judgments of velopharyngeal contact during speech are made. This view alone is not sufficient to determine velopharyngeal closure because it does not confirm velopharyngeal contact along its width.

The lateral view permits excellent visualization of the function of the tongue during speech and is helpful in the differential diagnosis of compensatory articulation errors. In some cases, the tongue is observed to elevate the soft palate in an attempt to effect velopharyngeal closure, particularly for the velar stop sounds /k/ and /g/. Abnormal posterior movements of the epiglottis and elevation of the larynx during speech are also apparent in this view.

The frontal view is usually performed next to determine the degree and location of mesial movements of the lateral pharyngeal walls. Lateral pharyngeal wall movements may occur at a specific location in the vocal tract or along an extensive area. This information is thought to be useful in planning the approximate width of a pharyngeal flap or designing a prosthetic speech appliance in cases of velopharyngeal insufficiency. To obtain this view, the patient is seated upright facing the image intensifier with his or her head positioned in the Frankfurt

horizontal plane. Complete coating of the nasopharynx with barium is essential to gather information for adequate interpretation. The standard speech sample is used.

In cases where the overlying bony structures obscure the lateral pharyngeal walls and their movements, the Waters' view is a helpful alternative. In this view, the head is tilted upward approximately 450° from the Frankfurt horizontal plane so that the bony structures do not impede the view of the lateral walls. When asymmetric movement of the lateral pharyngeal walls is observed, Shprintzen et al. (1977) recommend rotation of the head to the right and left along the x-axis to more clearly identify the extent of movement.

The third view to be performed is either the base or Towne's view. These views outline the shape of the velopharyngeal valve; the pattern, symmetry, and consistency of velopharyngeal valving movements; and the size and location of velopharyngeal gaps during speech. The Towne's view is the more useful of the two views when the soft palate approximates adenoid tissue during valving creating an oblique axis of velopharyngeal closure or when patients have large tonsils or posterior compensatory tongue movements. These situations interfere with adequate visualization and interpretation of velopharyngeal movements in the base view.

To obtain the base view, the patient lies prone on the x-ray table in a sphinxlike position with the head hyperextended. To obtain the Towne's view with over-table tube fluoroscopy equipment, the patient is seated upright with his or her head in the horizontal plane. The camera is then rotated in relation to the face until the velopharyngeal valve is visualized. This view is similar to that used in nasopharyngoscopy. The standard speech sample is used for both the base and Towne's views.

Multiview videofluoroscopy may be reliably performed in patients as young as 3 or 4 years old. Because this test involves radiation, it should be used judiciously. It is most reliable when the child is mature enough to cooperate for the examination and has sufficient speech development to allow visualization of the valve throughout all classes of speech sounds. The test should be used when the clinical speech assessment suggests velopharyngeal inadequacy, particularly when surgical or prosthetic management is being considered.

32.6 Ultrasound

Ultrasound is a device that has been employed in the evaluation of velopharyngeal function, particularly movements of the lateral pharyngeal walls. It is not suitable for displaying motion of the velum because of problems in transmitting ultrasound through bone overlying the palate (Hawkins and Swisher 1978).

32.7 Video Nasopharyngoscopy

This technique involves inserting a flexible fiberoptic tube into the nose to obtain a direct superior view of the velopharyngeal valve and vocal tract during speech. In the hands of an experienced and patient examiner, the test provides information about the anatomy and function of the velopharyngeal valve during speech; the relative size, location, and consistency of velopharyngeal gaps; the function of the posterior aspect of the tongue during speech, which is helpful in the differential diagnosis of compensatory articulation errors; and the anatomy and function of the laryngeal structures. It is also useful in the identification of pulsations in the pharynx which may be indicative of abnormally placed carotid arteries that might preclude pharyngeal flap or sphincter pharyngoplasty surgery. Although the latter finding is rare, it occurs most often in patients with velocardiofacial syndrome. This technique is useful not only for pretreatment diagnostic assessment of velopharyngeal function but also to evaluate the outcome of surgical, prosthetic, and/or speech therapy treatment. It may also be used for biofeedback therapy to enhance velopharyngeal movements during speech and correct some compensatory articulation errors. For ease of examination in young children, a flexible nasopharyngoscope with

a distal tip diameter between 2 and 3.7 mm is recommended.

McWilliams et al. (1984) note that lateral wall movement is not reliably assessed by either measurements or judgments of nasoendoscopic images. A major reason for using endoscopic procedure is to learn about the contribution of the lateral pharyngeal walls to velopharyngeal function. Some investigators have assumed symmetry of lateral wall movement in making their judgments, but this assumption is questionable. Further data relative to the realizability and validity of endoscopic measurements are needed. In the meantime, the technique is quite commonly used clinically and has much to offer in the hands of trained examiners.

32.7.1 Technique

A topical anesthetic such as 3 % lidocaine or 2 % tetracaine hydrochloride mixed in equal parts with .5 % phenylephrine is applied through one side of the patient's nasal cavity to allow comfortable insertion of the scope and enhance cooperation. Some patients are able to tolerate the procedure without anesthesia, particularly when a smaller scope is used. The nasopharyngoscope is inserted through the middle meatus of the nasal passage to first visualize the velopharyngeal area to document anatomy and function and scan the pharynx for abnormal pulsations. The scope is then passed down into the vocal tract to document the anatomy and function of the tongue and larynx. In patients with a preexisting pharyngeal flap, the scope can be passed through one or both ports to visualize the lower vocal tract as long as the ports are not stenosed. A standard speech sample is used at each observation point in the vocal tract. Care must be taken when positioning the scope in the velopharynx. A false-positive or false-negative diagnosis of velopharyngeal closure will be obtained if the distal end of the scope is not positioned directly above the velopharyngeal port. The quality of the study and interpretation, treatment planning, and outcome analysis are enhanced by audio-video recording which allows playback of the study for repeated analysis, comparisons with previous examinations, and demonstration of the findings to the professionals involved in the treatment and the patient and/or parents.

32.8 The Nasometer

The nasometer is a computer-assisted instrument produced by Kay Elemetrics (Pinebrook, New Jersey) which is designed to measure the relative amount of nasal acoustic energy compared to oral acoustic energy during continuous speech production (Dalston et al. 1981). This instrument uses a sound separator that rests on the patient's upper lip. Microphones on either side of the sound separator sense oral and nasal acoustic energy during speech, and this energy is filtered and digitized by custom electronic modules. The computer with Kay Elemetrics software (Version 1.7) processes the information and produces a "nasalance score." This score is a ratio of nasal to oral acoustic energy multiplied by 100. The nasal and oral acoustic energy is averaged during the production of vowels and consonants in test sentences to produce the nasalance score. The length of the speech sample used to calculate the nasalance score may be up to 100 s. This instrument is based on the Tonar II developed by Fletcher in 1976 (Fletcher and Bishop 1970). An abnormally high nasalance score during production of nonnasal consonants suggests velopharyngeal inadequacy and hypernasality, and an abnormally low nasalance score during production of nasal consonants is suggestive of hyponasality and/or nasal airway impairment (Dalston et al. 1981). Dalston et al. (1981) studied the sensitivity and specificity of the nasometer and found it to be an appropriate instrument for use in corroborating listener judgments of hypernasality. Sensitivity and specificity values for assessing hyponasality were in the expected range for patients without any indication of concurrent velopharyngeal inadequacy, but the scores did not identify hyponasality in patients who exhibited both hyponasality and excessive nasal air emission. Therefore, the instrument may be less useful as

a diagnostic procedure in patients who exhibit both velopharyngeal inadequacy and nasal airway impairment (Dalston et al. 1981). The nasometer is not a substitute for listener judgments of hyper- and hyponasality but can be useful in providing baseline data to assist in the identification of velopharyngeal inadequacy, assessment of treatment outcome, fitting of a palatal prosthesis, and providing visual biofeedback in speech therapy.

32.8.1 Technique

Care should be taken to ensure that the system is calibrated according to the specifications of the manufacturer. The headgear is then adjusted to fit the patient, and the patient is asked to read or repeat a standard speech sample. This procedure is usually used with patients age 3 and older because younger patients may have shorter attention spans and less well-developed speech and language skills. Once the speech sample is recorded on the computer terminal, the software cursors are used to mark the beginning and end of the speech display. The "calculate" function is then activated, and the mean and standard deviation of the nasalance score are determined.

32.9 Aeromechanical Measurement

32.9.1 Warren and Dubois Technique (Warren and DuBois 1964)

Measurements of nasal airflow and of the difference in air pressure above and below the velopharyngeal port may be used to estimate both the area of the velopharyngeal orifice, if any, during the production of stop consonants and the resistance of the port to airflow. Pressure-flow measurements provide information about the coupling of the oral and the nasal cavities during speech and about resistance in the system. They do not describe the movement of particular structures, such as the velum and lateral pharyngeal walls, or the location and configuration of any opening that is present.

32.9.1.1 PERCI

Warren (1979) introduced an instrument called the PERCI (Palatal Efficiency Rating Computed Instantaneously) for use in the evaluation of the velopharyngeal mechanism during speech. PERCI records and displays the difference in air pressures in the mouth and in the nose. From a study of 75 cleft palate patients, Warren reported that patients with differential pressure readings >3.0 on the PERCI had velopharyngeal orifice areas of 10 mm^2 or less, whereas those with PERCI readings of <1.0 had areas greater than 20 mm^2. PERCI readings of 1.0 through 2.9 were associated with velopharyngeal areas between 10 and 20 mm^2.

32.9.1.2 TONAR

Fletcher and Bishop (1970) advanced the study of oral and nasal sound intensity measures as indices to hypernasality through the development of an instrument which they named TONAR (The Oral-Nasal Acoustic Ratio). The instrument prints out voltages associated with the nasal and oral signals and also a trace reflecting the ratio of the voltages from the sound detected in the oral and nasal chambers.

32.10 Summary

Several instrumental procedures are available for assessing the velopharyngeal mechanism and its function. Each has advantages and disadvantages, and choosing among them depends on the specific purpose of the evaluation. The reliability of endoscopic procedures is not well documented:

Aerodynamic measures provide data about the area of the velopharyngeal opening, velopharyngeal resistance to air flow, and air pressure available for the production of obstruent sounds. These measures provide no information about the relative contributions of the velum and the pharyngeal walls to velopharyngeal function.

An important warning in the use of any instrumentation for the study of speech is that data taken during speech production must be interpreted within the context of the patient's repertoire of speech proficiency.

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Variations in Nasopharyngeal Skeletal Architecture

33

Samuel Berkowitz

33.1 Muscles

33.1.1 Pharynx and Velum (Dickson and Dickson 1982)

The adult pharynx is the common pathway for food and air in human beings. It extends from the cranial base behind the nasal cavities to the upper end of the esophagus behind the larynx. The pharynx is widest at its upper portion and narrows as it descends to the esophagus. The pharynx lies immediately in front of the vertebral column, separated from it by prevertebral muscles and fascia. Anteriorly, the pharynx communicates with the nasal cavities, the oral cavity, and the aditus of the larynx. Inferiorly, the pharynx is continuous with the esophagus. The part of the pharynx that extends upward to the level of the velum (soft palate) is commonly called the nasopharynx (Fig. 33.1).

The pharyngeal orifices of the auditory tubes open into the lateral walls of the nasopharynx. The auditory tube connects the pharynx with the middle ear and serves to maintain an equilibrium of air pressure between the middle ear and the external atmosphere. The superoposterior part of the pharynx contains the pharyngeal tonsil (also called the adenoid).

The velum (soft palate) forms the boundary separating the nasopharynx from the oropharynx. Because the velum is mobile, this boundary is arbitrary. The velum is a muscular body that attaches to the posterior rim of the hard palate and the lateral walls of the posterior part of the oral cavity. The posterior free border of the velum hangs into the oropharynx and ends in a small midline projection, the uvula.

The oropharynx extends inferiorly to the level of the hyoid bone. The anterior wall of the oropharynx, inferior to the opening into the oral cavity, is formed by the posterior surface of the root of the tongue.

The palatopharyngeal fold (posterior faucial arch) extends from the sides of the velum into the lateral walls of the pharynx. This muscular fold forms the greatest lateral constriction between the oropharynx and oral cavity. The superior end of the epiglottis extends superiorly from the larynx into the oropharynx immediately posterior to the root of the tongue.

The nasopharynx and auditory tube are lined by ciliated, pseudostratified columnar epithelium rich in goblet cells and glands that secrete mucus. The pharynx is lined with stratified squamous epithelium where contact between the pharynx

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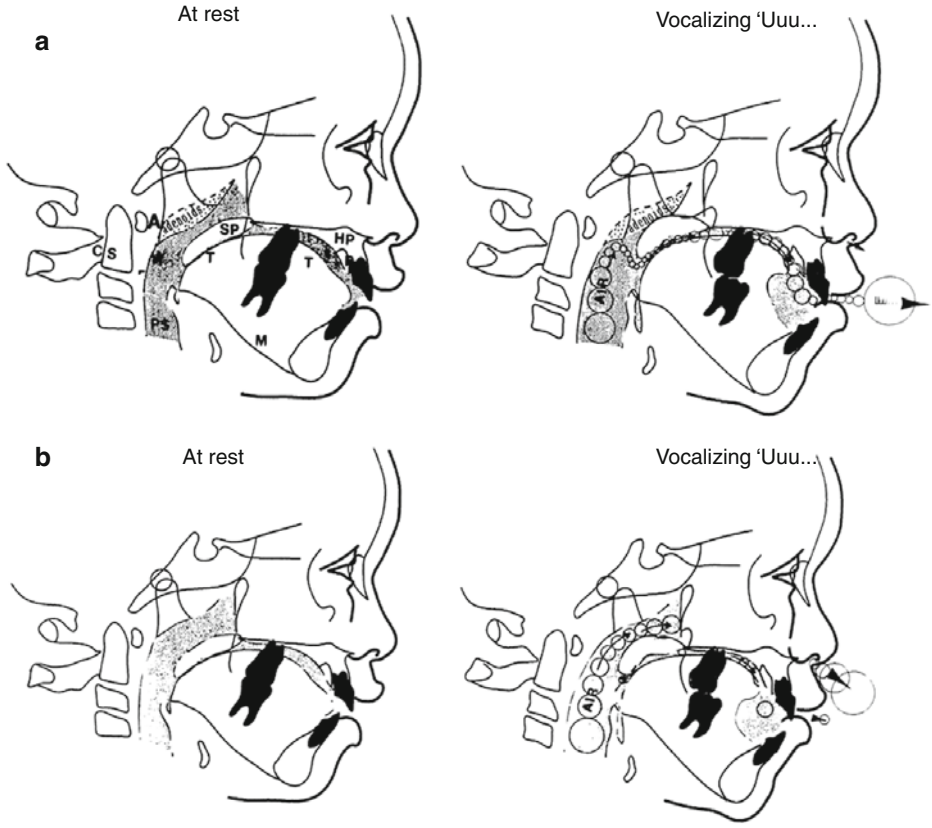

Fig. 33.1 (**a**) Lateral cephaloradiograph shows the skeletal structures surrounding the pharyngeal space and allows evaluation of the pharyngeal depth, shape of cervical spine, soft palate size and length, and extent of soft palate elevation. Because it is a two-dimensional representation of the velopharyngeal area and does not show lateral pharyngeal wall motion, this record cannot be used to diagnostically determine velopharyngeal functions. (**a**) *Left*: Structures involved in controlling airflow as seen in a lateral cephalograph at 5 years of age (*A* anterior tubercle of the atlas, *CS* cervical spine, *S* odontoid process, *W* posterior pharyngeal wall, *PS* pharyngeal space, *SP* soft palate (velum), *HP* hard palate, *T* tongue, *M* mandible). *Right*: Air flow when vocalizing "Uuu" during normal speech. The soft palate elevates and makes contact with the adenoids (if present) or the posterior pharyngeal wall during normal speech and swallowing. When the lateral pharyngeal muscles operate in coordination with a competent (adequate length, width, and timing of action) soft palate in a normal skeletal environment most of the air is channeled through the mouth while some enters the nose. This is designated as velopharyngeal competency (VPC). (**b**) Velopharyngeal Incompetency (VPI). There are usually many reasons for inadequate air flow control. Some are (1) a relatively deep pharynx when related to velar length, (2) inadequate velar elevation and/or pharyngeal wall motion (neuromuscular function), and (3) poor timing of speech with pharyngeal and velar muscle (sensory-motor) function

and velum takes place and in the portion that serves as a food channel.

33.2 Nasopharyngeal Growth

Variations in the height and depth of the nasopharynx are apparent from birth through the early years of growth and development. Rosenberger (1934) made a longitudinal study of nasorespiratory areas in children from 3 months to 5 years of age. It was his opinion that an enlargement of the nasorespiratory area accompanied growth in the body and great wing of the sphenoid as well as the forward drift of the hard palate. Brodie (1941) demonstrated that the hard palate moves downward, away from the base of the skull, in a parallel manner with increasing age and that this results in a progressive increase in the height of the nasal and nasopharyngeal areas.

In a study of the growth of the pharynx, King (1952) concluded that there is a considerable increase in the vertical dimension of the pharynx resulting from the descent of the hard palate, mandible, and hyoid bone, as well as from an increase in the height of the cervical vertebrae. He concluded that there is no appreciable increase in the anteroposterior dimension of the pharynx after an early age.

According to Scott (1953, 1957, 1958), growth in the height of the pharyngeal area is regulated by the cartilages of the cervical vertebrae. He, as well as Todd and Tracy (1930), attributed growth in the anteroposterior dimension of the nasopharynx to growth at the spheno-occipital synchondrosis. Subtelny (1955; Subtelny and Baker 1956) studied vertical growth of the nasopharynx and observed a steady rate of increase up to 15 years of age. Using a measurement from the posterior nasal spine to the soft tissue of the posterior wall of the pharynx, he observed a general increase in depth of the nasopharynx at the level of the palatal plane. Berkowitz (1989) believes that this increase is due mainly to a change in the curvature of the posterior pharyngeal wall.

Several investigators studying cleft palate subjects have focused attention on skeletal structures closely related to the nasopharynx. Ricketts (1954) found that the basilar portion of the occipital bone varies in its position in relation to the anterior cranial base and that these variations can influence the anteroposterior dimension of the nasopharynx. Brader (1957) reported significantly smaller anteroposterior and vertical nasopharyngeal dimensions in cleft palate subjects than in noncleft individuals and noted that his cleft palate subjects had relatively larger masses of adenoid tissue within the nasopharynx than noncleft individuals. He also observed no significant difference in the angularity of the cranial case in individuals with cleft palate when compared with individuals without clefts.

Moss (1965) studied malformations of the skull base associated with cleft palate deformities and indicated that a flexion of the cranial base (N-S-Ba) placed the basioccipital bone "more anteriorly relative to the sphenoid bone," thus creating a smaller anteroposterior nasopharyngeal dimension. Subtelny (1955; Subtelny and Baker 1956) showed that width of the skeletal nasopharynx was larger in persons with cleft palate than in normal individuals.

Coccaro et al. (1962) studied nasopharyngeal growth in children with cleft lip and cleft lip and palate using serial cephalometric films on 57 subjects from 3 months to 7 years of age. Comparisons were made with a noncleft group and with narrative data supplied by Subtelny. Results showed that: (1) The cleft palate group had smaller nasopharyngeal height measurements than the normal group, but only up to 3 years of age. (2) By 3 years of age, all three groups had attained approximately 80 % of their respective increases in nasopharyngeal height. (3) Nasopharyngeal depth in the cleft palate group increased slightly more than in the noncleft group (6.8 mm vs. 4.3 mm). (4) The horizontal nasopharyngeal changes in the cleft palate group were shorter than in the noncleft group for most of the age levels studied.

33.3 Functions

The pharynx serves as an air passage connecting the mouth and nose to the lungs via the larynx and trachea and as a food passage connecting the mouth to the stomach via the esophagus. Because these channels are common over part of their length in humans, a sphincter is found at the level of the velum to prevent food from entering the nasal cavities during swallowing.

33.3.1 Swallowing

Constriction of the pharyngeal walls results in changes in the diameter of the pharynx. These changes are important to both swallowing and speech. During swallowing, the lateral and posterior walls of the pharynx move anteromedially, reducing the cross-sectional area of the pharynx. This results in a wavelike constriction that flows down the pharynx and continues throughout the entire digestive tract. The wavelike motion is called peristalsis.

In the initial stage of swallowing, the pharynx constricts around the velum while the

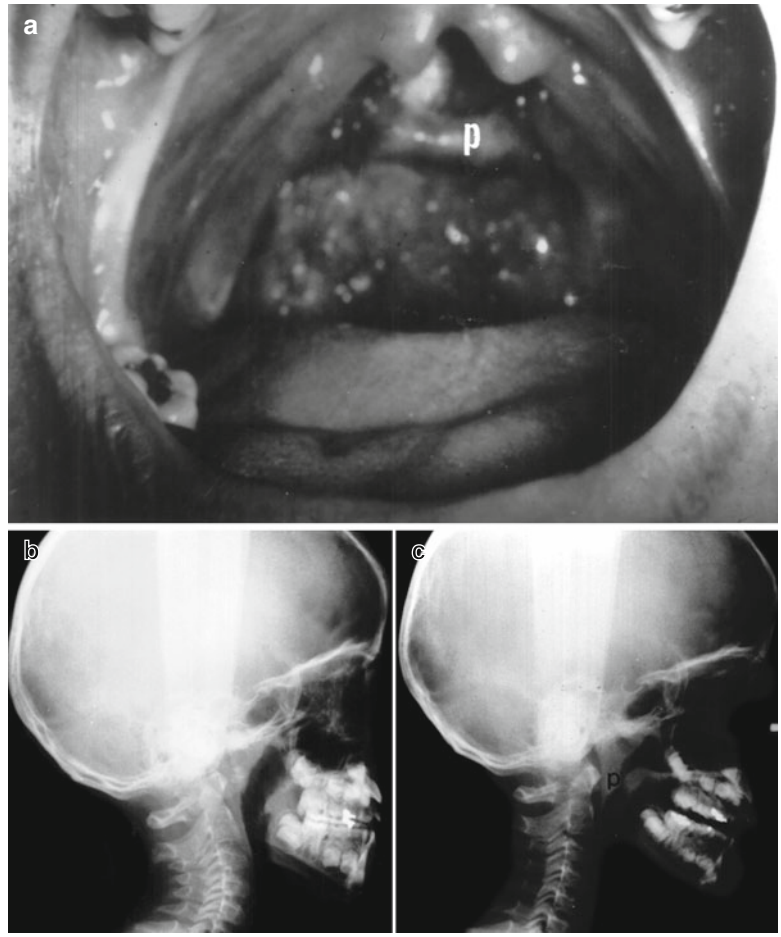

Fig. 33.2 (**a**–**c**) Passavant's Pad. The superior border of the superior constrictor pharyngeus muscle on contraction forms a ridge or pad on the posterior wall. It is observed in both noncleft and cleft patients. There is no consistent relationship of the pad to the velum, and it is usually seen during swallowing as well as during speech, whistling, and blowing. Some state that it functions adequately for compensatory purposes and does not appear to contribute to velopharyngeal closure in all patients. (**a**) An intraoral view of Passavant's pads (*p*). (**b**) Lateral cephaloradiograph – at rest. (**c**) While phonating "Uuu," showing Passavant's pad (*p*) at the level of the anterior tubercle of the atlas making contact with the uvulae

velum moves somewhat dorsally and superiorly, closing off the nasopharynx from the oropharynx (a movement called velopharyngeal closure). The oropharynx expands somewhat, and the larynx moves superiorly and ventrally, opening the pharynx to receive the bolus of food. As the bolus moves into the pharynx, the pharyngeal walls constrict behind it, propelling the bolus toward and into the esophagus.

33.3.2 Speech

Oropharyngeal constriction and changes in length also occur during speech. These changes affect the resonant frequencies of the vocal tract and thus change the quality of the voice and influence the perception of what phoneme is produced. The lateral walls of the oropharynx move medially on low vowels and laterally on high vowels.

Velopharyngeal closure for speech eliminates the nasal cavities from the resonance system and permits the buildup of intraoral breath pressure to produce fricative and plosive consonants. The mechanics of velopharyngeal closure for speech are quite different from those for swallowing. During speech, the velum moves superiorly and posteriorly against the posterior pharyngeal wall (Fig. 33.2), while the lateral pharyngeal walls move medially against the sides of the velum (Fig. 33.3). By contrast, velopharyngeal closure for swallowing is primarily pharyngeal.

Radiological studies have demonstrated that, during closure for speech, the velum also increases in length, particularly in its posterior

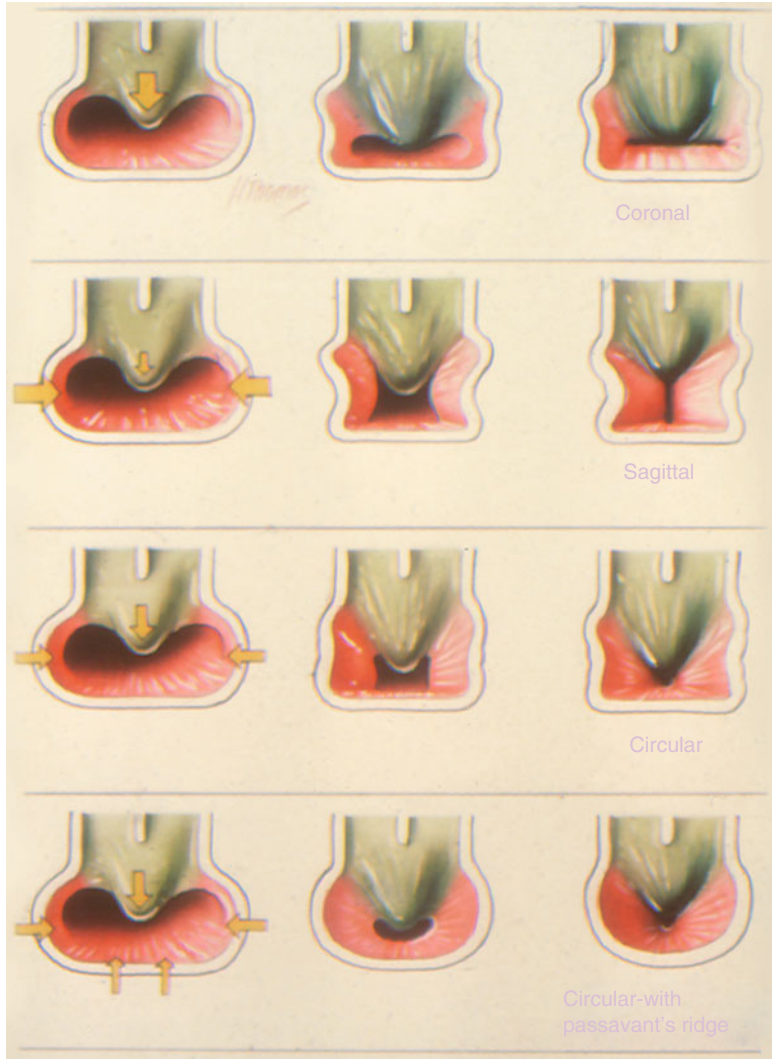

Fig. 33.3 Four basic velopharyngeal closure patterns. The size and shape of velopharyngeal valving patterns is very variable (Skolnick 1975). Although the closure patterns are not actually discrete, for convenience sake, Skolnick et al. (1973) categorized velopharyngeal valving into four patterns. These patterns are: Coronal pattern: The majority of valving is palatal with the full width of the velum contacting the posterior pharyngeal wall. The lateral walls move medially to the lateral edges of the velum. There is no motion in the posterior wall. Sagittal pattern: The majority of valving is pharyngeal. The lateral walls move to the midline and approximate each other. The soft palate does not contact the posterior pharyngeal wall but instead abuts up against the approximated lateral pharyngeal walls, thus completing the closure pattern. Circular pattern: There is essentially equal contribution from the velum and lateral pharyngeal walls with the bulk of the musculus uvulae acting as the focal point. The dorsum of the musculus uvulae contacts the posterior wall (which does not move). The lateral walls squeeze around the bulk of the musculus uvulae. Passavant's ridge pattern: As in the circular pattern, there is essentially equal contribution from the velum and lateral pharyngeal walls, but in addition, the posterior pharynx moves forward. The musculus uvulae also serves as the focal point for closure in this pattern (Reproduced with permission from Siegel-Sadewitz and Sphrintzen (1982))

part. The velum contacts the posterior wall just above the level of the anterior tubercle of the atlas and on a plane with the hard palate. Velar elevation is somewhat greater for males than for females, and the greatest velar motion is at the junction of its middle and posterior thirds.

It is of interest to note that both velar elevation and the degree of motion of the lateral nasopharyngeal walls during speech are greater for high vowels than for low vowels. This is the reverse of the activity of the lateral walls of the oropharynx. The degree of motion, and in fact the degree to which complete closure is achieved, depends on both the phoneme being produced and its phonemic context. Motion is greatest for fricatives, less for high vowels, and least for low vowels. The velopharyngeal function is greater for vowels in a nonnasal context than in a nasal consonant context. Finally, the extent of lateral wall motion and the degree of velar motion are synchronized and directly and highly related to each other.

Velopharyngeal closure during speech has received considerable attention, and properly so because it is an important factor in the treatment of cleft patients. It is evident that the tensor veli palatini muscle is not active in velopharyngeal function. The palatopharyngeus is active during velar lowering, but its activity is inconsistent and vowel-dependent. Palatoglossus activity relates more to tongue motion than to velar activity. The superior constrictor and levator veli palatini muscles are the only two velar muscles studied (the uvulus muscle has not been definitively studied) that are active during velopharyngeal function for speech. All investigators agree that levator veli palatini activity is synchronized with velar height.

Indirect evidence indicates that the uvulus muscle plays an essential role in velopharyngeal function. Nasopharyngoscopy has revealed a patient population with hypernasality, a midline longitudinal concavity rather than convexity of the dorsal velar surface, translucency of the longitudinal velar midline, and a small midsagittal gap between the velum and posterior pharyngeal wall during velopharyngeal closure for speech. This lends presumptive evidence that what has been called the levator eminence is actually a function of the uvulus muscle. In lateral projection, the velum may seem to touch the posterior pharyngeal wall, even when there is actually a small central gap because of the absence of the musculus uvulae.

Thus, velopharyngeal closure for swallowing is accomplished by the superior constrictor, levator veli palatini, and uvulus. Closure for speech is accomplished by the levator veli palatini and uvulus. The difference in function during swallowing and speech is also apparent among persons with velopharyngeal insufficiency due to congenital disproportion between the pharyngeal depth and velar length or disproportion following surgery for cleft palate. In our experience, most individuals in this group who are unable to achieve velopharyngeal closure for speech do so consistently during swallowing.

33.4 The Role of the Nasal Cavity (Fig. 33.4a)

The nasal cavities are the first points of entry and the last points of exit of airflow to and from the lungs. They serve to filter the air as it enters the airway and to control air temperature and humidity. They also serve as resonators for nasal sounds that are produced with the velopharyngeal valve open. The two nasal cavities are separated from each other by the nasal septum in the midsagittal plane. The nasal septum is composed of three parts. The superior part is the perpendicular plate of the ethmoid bone. The inferior and posterior parts are composed of the vomer bone. The anterior part is composed of the cartilage of the nasal septum.

The roof of the nasal cavities is formed by the cribriform plate of the ethmoid bone, which is penetrated by the olfactory nerves. Immediately posterior to the perpendicular plate of the ethmoid is the body of the sphenoid, which contains the sphenoid sinus. The floor of the nasal cavities is formed by the hard palate. Anteriorly, the nasal cavities are bounded by the external nose. The framework of the external nose includes the nasal bones and the nasal cartilages. The bridge of the nose is formed by the nasal bones. Inferiorly, the nasal bones articulate with the lateral nasal cartilages. The tip of the nose is supported by the greater alar cartilages. Small lesser alar cartilages support the lateral walls of the anterior nasal openings.

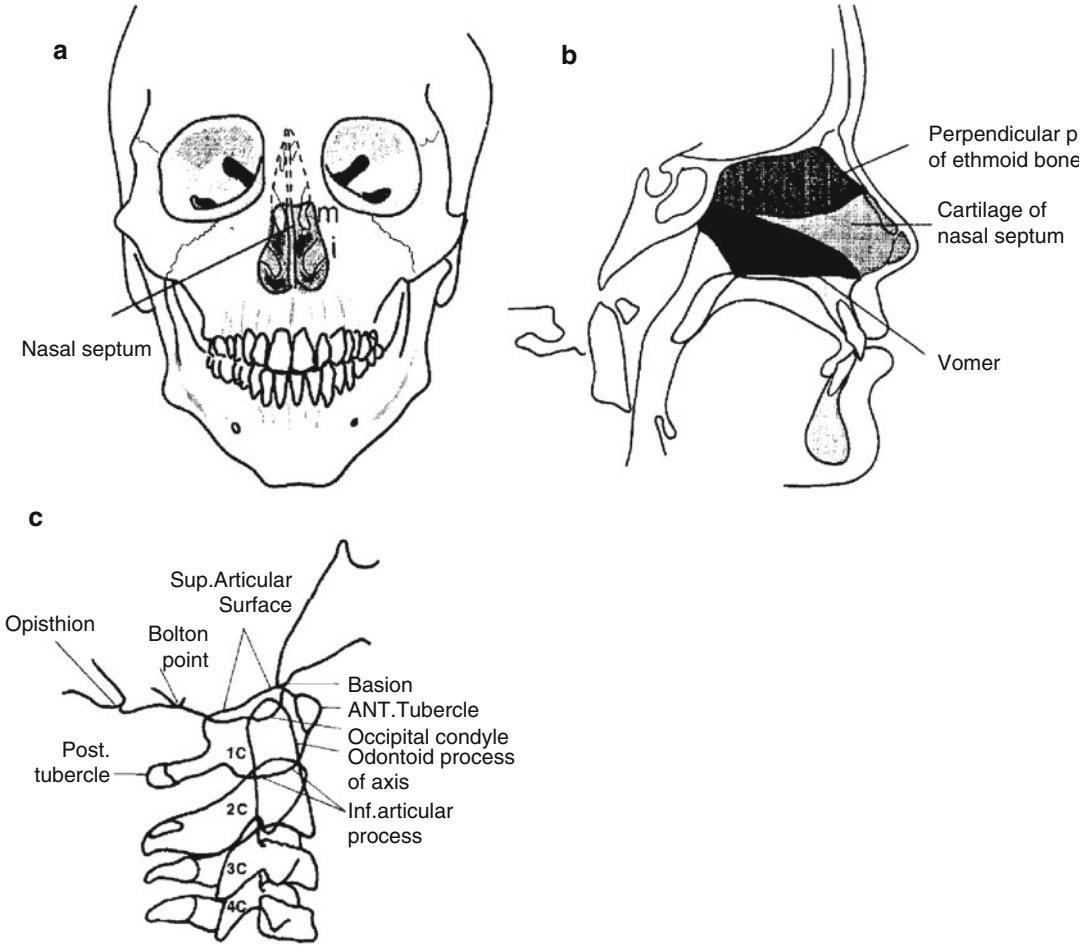

Fig. 33.4 (**a**) The normal nasal septum: The inferior (*i*), middle (*m*), and superior turbinates are attached to lateral nasal walls. (**b**) The nasal septum is composed of the perpendicular plate of the ethmoid, the cartilage of the nasal septum, and the vomer. In complete clefts of the lip and palate at birth, the nasal septum is displaced to the noncleft side in varying degrees. The configuration of the nasal septum and the turbinates on the cleft side limits the medial movement of the cleft palatal segment after lip and palate repair. (**c**) Cervical spine: The condition and the relationship of the various vertebrae to each other can influence the depth and configuration of the pharyngeal space. For example, the anterior tubercle of the atlas (1 C, first cervical vertebrae) supports the posterior pharyngeal wall. If it is absent or dislocated, the functional pharyngeal depth may increase to a nonfunctional state

The lateral wall of each nasal cavity is convoluted owing to the presence of the superior, middle, and inferior nasal conchae. The conchae have an extremely rich blood supply and provide a large surface area for humidity and temperature control of the air passing over them.

Although the terms VP "incompetence," "inadequacy," and "insufficiency" historically have been used interchangeably, they do not necessarily mean the same thing. For this reason, standardization of nomenclature has been recommended. Trost-Cardamone (1989) has proposed a taxonomy for VP disorders, based on causative factors in which "velopharyngeal inadequacy is the generic term used to denote any type of abnormal velopharyngeal function." In the broad group of inadequacies, there are subgroups of structural (VPI), neurogenic (VP incompetence), and mislearning (VP mislearning) or functional origins. VPI includes any structural defects of the velum or pharyngeal walls at the level of nasopharynx with insufficient tissue to accomplish closure or

some kind of mechanical interference with closure. The all-encompassing term "velopharyngeal dysfunction (VPD)" does not assume or exclude any possible cause of the perceived speech symptoms or management approach. It applies to speech disorders that may be the result of structural deficits, neurologic disorders, faulty learning, or a combination of sources (Witt and D'Antonio 1993).

33.5 The Use of Lateral Roentgencephalometrics in Evaluating Skeletal Pharyngeal Architecture and Velar Elevation

Velopharyngeal valving is dependent not only on the sensorimotor adequacy of the velum and its synergistic musculature but also on the morphologic skeletal dimensions of the nasopharyngeal port. The size and shape of the nasopharynx are determined, in part, by the contiguous osseous anatomy of the maxilla (Subtelny 1955), cranial base, and vertebral column (Figs. 33.4 and 33.5).

The mechanism of velopharyngeal closure can be studied to some degree by cephalometric roentgenography, but it must be stressed that this is not a functional test for velopharyngeal competency (Fig. 33.6). The vowel /u/, as in "boom," is generally employed to achieve maximal elevation of the soft palate during vowel production. After careful rehearsal, the subject is instructed to maintain the sound at a constant pitch and intensity for a time sufficient to cover the roentgenographic exposure period. This procedure ensures that the soft palate will remain in a reasonably stable position during the recording interval.

In a noncleft individual, as seen in the lateral headplate, the soft palate elevates to contact the posterior pharyngeal wall. In doing so, the longitudinal axis of the soft palate becomes continuous with that of the total palatal plane, while the uvula projects downward and nearly at right angles to the rest of the velum. If for any reason this velopharyngeal closure cannot be achieved, deglutition is impaired, and in phonation, the air stream is misdirected through the nose. However, it is possible to have good swallowing activity but poor velopharyngeal closure during speech. The term "palatal insufficiency" is more descriptive of a physiologic deficiency than of an anatomic defect.

33.6 Cervical Spine Anomalies (Fig. 33.7)

Osborne (1968) wrote that fusion of the posterior spines at C2–C3 does not appear to influence the osseous anteroposterior diameter of the nasopharyngeal port. On the other hand, occipitalization of the atlas, a smaller-than-normal anterior arch of the atlas, or atlantoaxial dislocation will have a direct effect on the anteroposterior dimension of the pharynx. It is clear that the population of congenital palatopharyngeal incompetence (CPI) patients with cervical anomalies, of whatever kind, present greater osseous nasopharyngeal depth (the distance between the hard palate and retropharyngeal wall) than do CPI patients without cervical anomalies.

The importance of the anterior tubercle of the atlas and the upper cervical vertebrae in achieving adequate velopharyngeal closure and speech is well established. One need know only that the musculofascial layer covering the upper cervical vertebrae, and forming the posterior pharyngeal wall, is only 2–5 mm thick to realize the morphologic importance of these vertebrae to velopharyngeal closure.

The influence of anomalies of the upper cervical vertebrae on the size and shape of the nasopharynx, with resulting effects on velopharyngeal valving, was first reported by McCarthy (1925) and, shortly thereafter, reemphasized by Schuller (1929). Epidemiologic studies have shown that patients with craniofacial birth defects, many of which are accompanied by velopharyngeal incompetence, have a higher prevalence of upper cervical spine anomalies than the general population (Osborne et al. 1971). This survey also found that a surprisingly large number (18.8 %) of patients with CPI demonstrated anomalies of the upper cervical

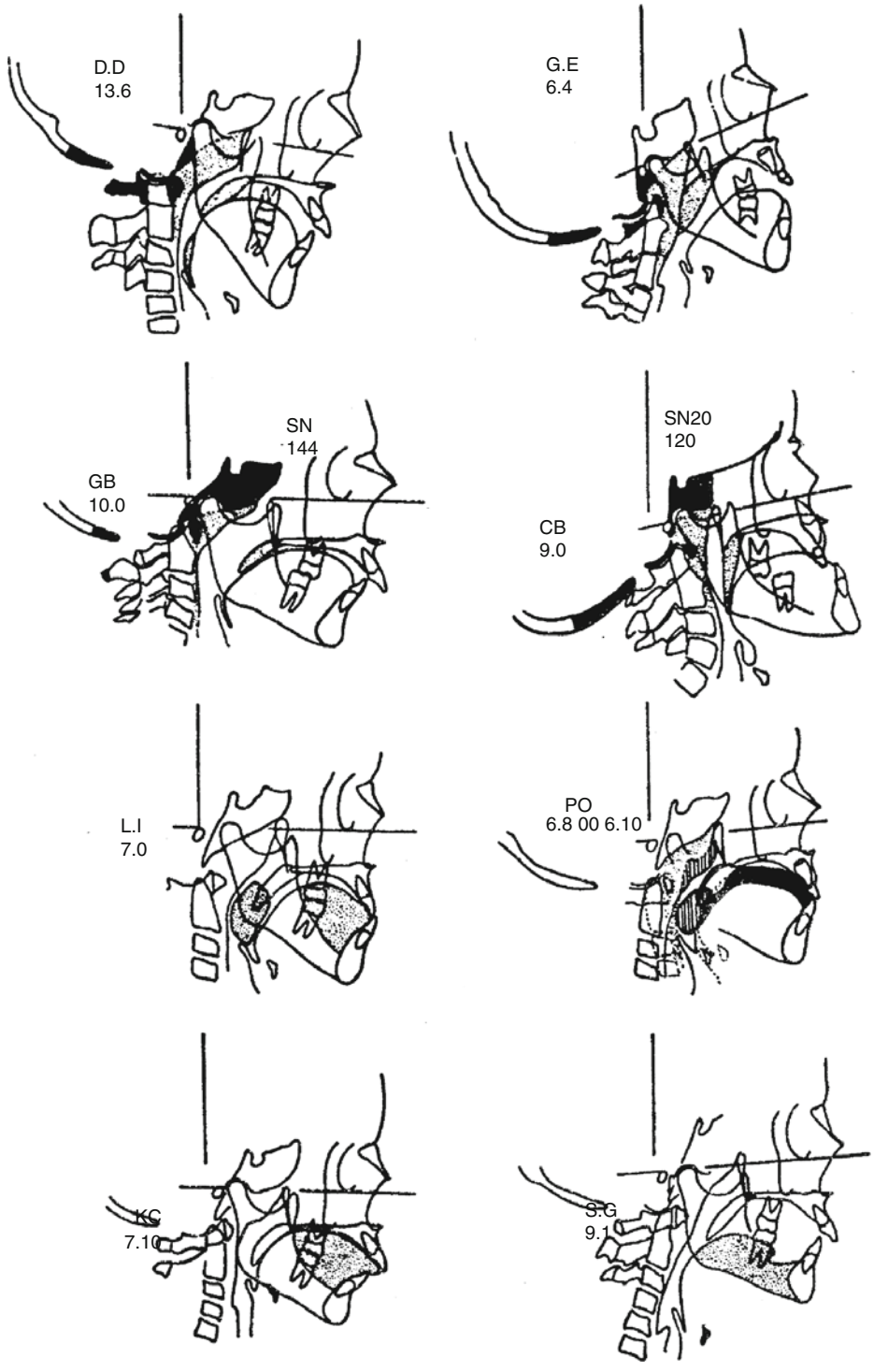

Fig. 33.5 Functional problems related to cervical vertebrae, cranial base, nasopharynx, adenoid, tonsils, and the tongue. *DD* dorsal displacement of vertebrae, *GE* congenital deformity with ventral displacement of cervical vertebrae, *GB* obtuse cranial base upward and backward location of anterior arch of the atlas and severe cleft palate speech (From functional diagnosis of malocclusion)

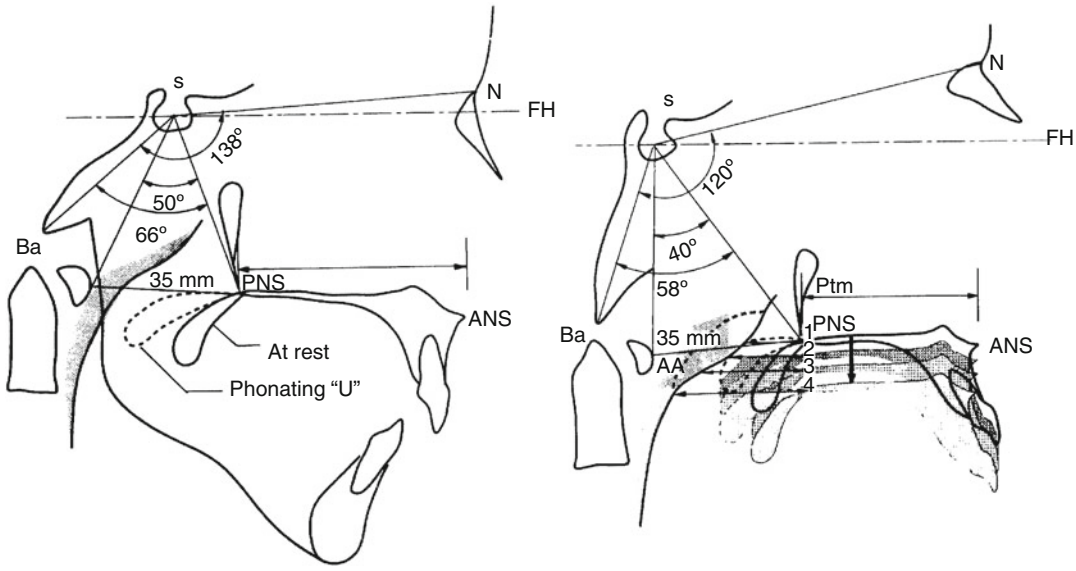

Fig. 33.6 The influence of the skeletal architecture on the pharyngeal form and size. The pharyngeal space is bounded superiorly by the cranial base and laterally by the cervical spine on one side and the tongue/hard palate complex on the opposite side. The odontoid process (*axis*) of the cervical spine points to the posterior extent (Basion-Ba) of the basilar portion of the occipital bone. It is the median point of the anterior margin of the foramen magnum. The hard palate of the maxillary complex is anatomically associated with the anterior cranial base and can vary in its anteroposterior dimension. The importance of cranial base angle (Ba-S-N). *Left*: Obtuse cranial base angle. In cases with a severe obtuse cranial base angle, the pharyngeal space is usually deeper than normal even in the presence of a long hard palate. An obtuse cranial base positions the cervical spine more posteriorly since it must be associated with the basilar portion of the occipital bone. This condition is usually seen when hypernasality exists in the absence of an overt palatal cleft and is called congenital palatal insufficiency (CPI). *Right*: With an acute cranial base angle, the cervical spine is positioned close to the hard palate creating a shallow pharyngeal depth. Comment: These relationships change with growth. The palate descends from the anterior cranial base in a parallel fashion. Due to changing slope of the posterior pharyngeal wall, the pharyngeal depth increases with age. The influence of adenoids: The adenoid is attached to the posterior pharyngeal wall above the level of the hard palate. Its size is highly variable according to age. It usually increases until 13 years of age and then retrogresses, but many variations in this growth pattern have been observed. The influence of cleft size: The severity of the cleft size in complete clefts of the lip and palate at birth does influence the nasopharyngeal width before lip and/or palatal surgery, but not after the lip is united. The degree of mesodermal deficiency of the hard and soft palate can determine not only the size of the cleft space by influencing palatal size but also the length and width of the soft palate and, therefore, its functional ability to affect velopharyngeal closure (VPC). A short anteroposterior maxillary dimension, as is usually found with submucous clefts, does not by itself signify that VPC will be inadequate. Short anteroposterior hard palate dimension can be associated with a shallow pharyngeal depth

vertebrae, as discerned in lateral cephalometric head film.

Simply stated, congenital palatopharyngeal incompetence (CPI) can be defined as the presence of hypernasality (rhinolalia aperta) in the absence of an overt cleft palate. Based on detailed studies of 110 subjects, Pruzansky and Mason (1966) found that CPI may be caused by one or more of the following variables: a bifid uvula; a short or thin soft palate, with or without a pink translucent area ("zona pellucida") usually seen in submucous cleft palate; anomalies of the upper cervical vertebrae and cranial base; and scanty adenoid development or early involution or excision of the adenoid.

As we shall point out in more detail later, our own concept of the defect in congenital palatal insufficiency has been extended to include a broader view of the pharynx and its contiguous structures.

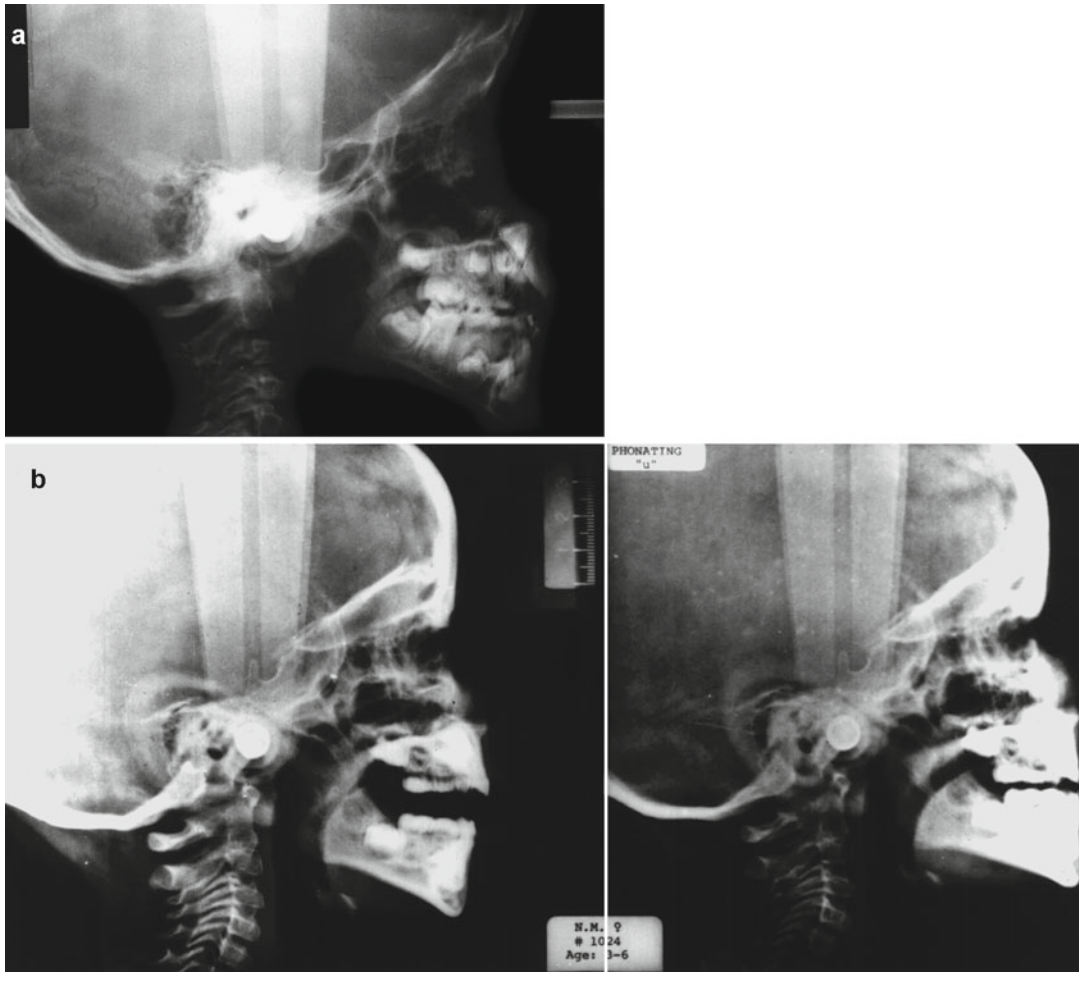

Fig. 33.7 (**a**–**e**) Cervical spine anomalies and their effects on the pharyngeal space configuration. (**a**) The absence and (**b**) malformation or dislocation of the anterior tubercle of the atlas result in the lack of support to the posterior pharyngeal wall, thus increasing the depth of the pharyngeal space. These anomalies coupled with a small velum will compound the problem. *Left*: Velum at rest. *Right*: Velum in function. No contact with the posterior pharyngeal wall

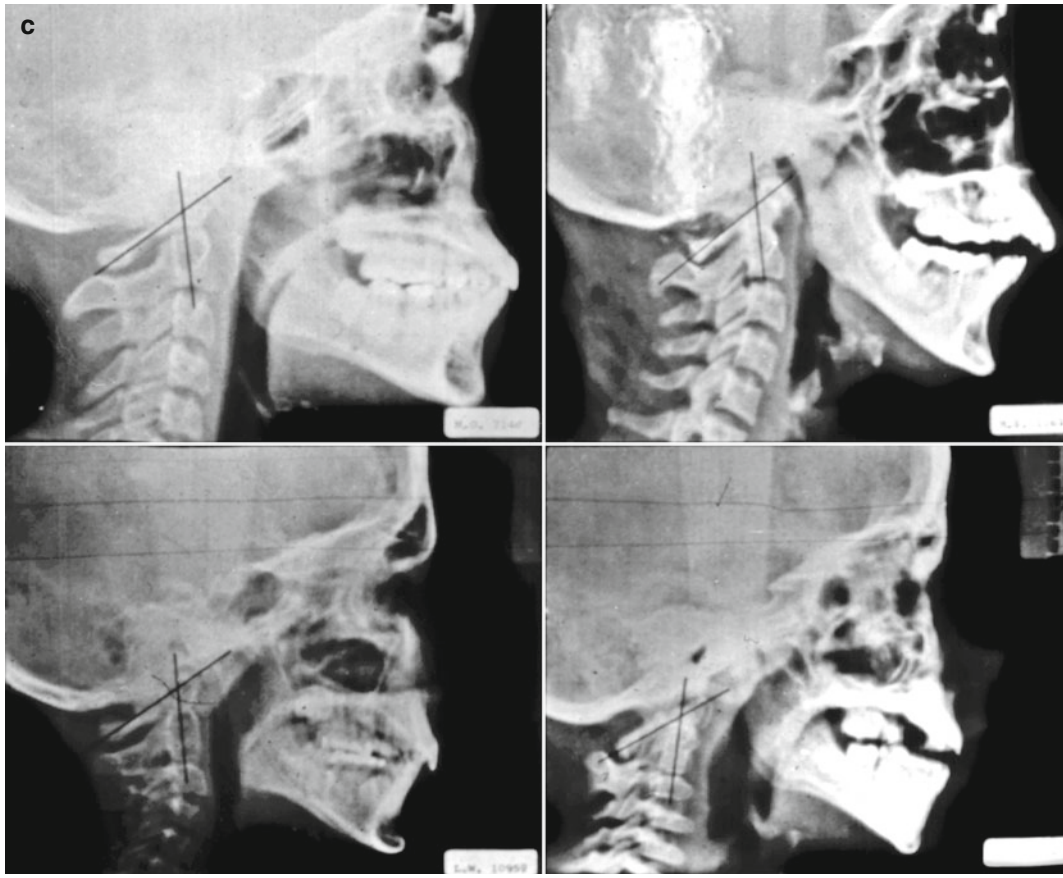

Fig. 33.7 (*continued*) Other cervical spine anomalies include: (**c**) Cervical spine anomalies abnormal atlanto-dental angulation

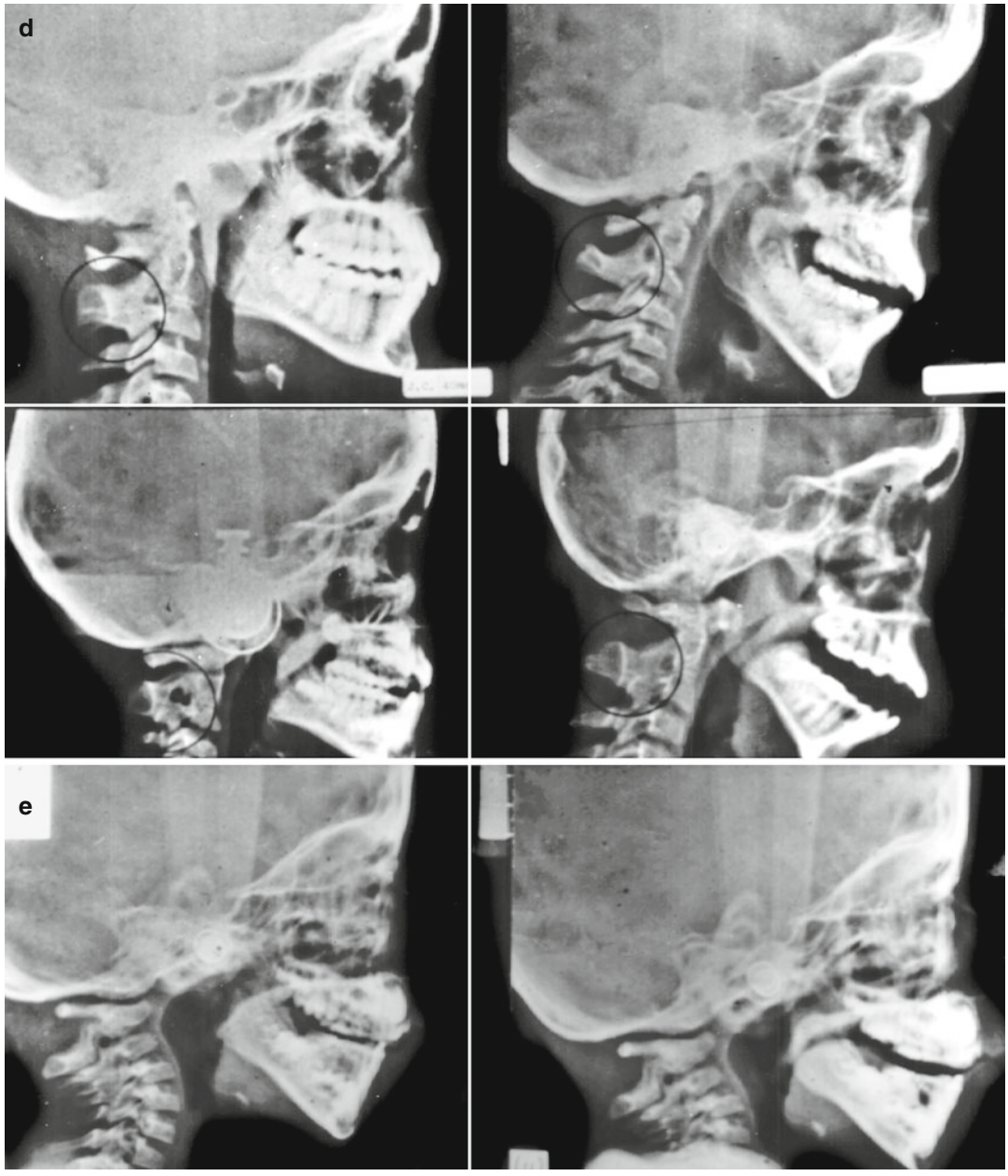

Fig. 33.7 (*continued*) (**d**) Cervical spine anomalies. Fusion of posterior arches C2–C3. (**e**) Abnormalities to the axis and atlas (Courtesy of S. Pruzansky)

33.7 Velar Closure

During the clefting process, the muscular forces that act in the region of the pterygoid plates account for the divergence of the pterygoid plates and the consequent increased lateral width of the nasopharynx, the separation of the tuberosities of the maxillae, and the width of the cleft itself. The failure of the hard and soft palate to unite in the midline produces a profound disturbance in the interplay and balance among the involved muscle groups. The tongue, pterygoid muscles, levators, and tensors of the velum tend to aggravate the malformation that has resulted from the failure of union in the midline of the palate.

A more favorable alignment of the osseous segments of the palate will follow reestablishment of balanced muscle forces. Slaughter and Pruzansky (1954) concluded that, since repairing the lip altered the spatial configuration of the skeletal framework so profoundly in the anterior segment of cleft, repairing the velum might achieve comparably beneficial results in the region of the nasopharynx.

In reviewing the morphologic variants that might contribute to inadequate velopharyngeal function, a number of factors must be considered. For example, the velum might be too short, the hard palate could be deficient in its anteroposterior dimension, or there may be unilateral or bilateral and posterior pharyngeal wall dysfunction. In other cases, both the hard and soft palates might be shorter than normal (Fig. 33.8a–e). In rare instances, the soft palate may be paralyzed and incapable of movement (Fig. 33.8f). The cases presented in Figs. 33.9 and 33.10 show variations in adenoid size and pharyngeal architecture which influence velar pharyngeal closure.

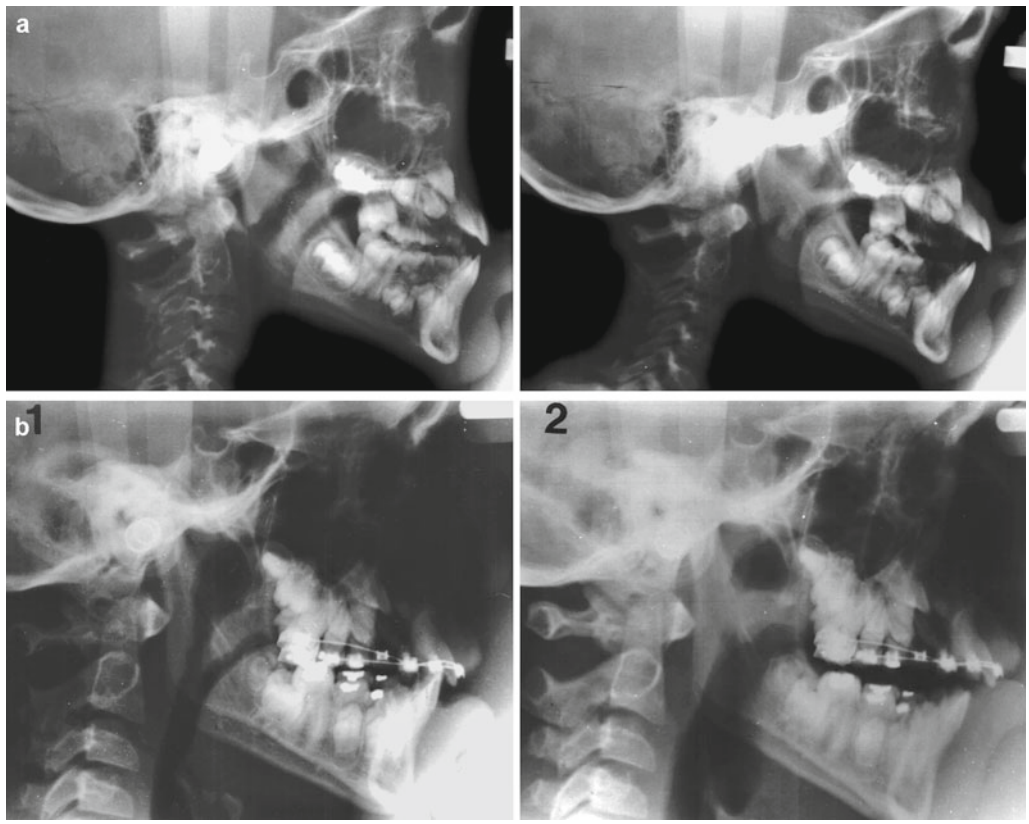

Fig. 33.8 (**a**–**e**) Variations in the pharyngeal space, velar elevation, and adenoid size. (**a**) *Left*: Normal pharyngeal architecture at rest, 10 years of age. *Right*: While vocalizing "Uuuu…." Note the velum of good length making contact against a small adenoid – the hard palate is positioned above the anterior tubercle. (**b**) Short, stubby velum in a relatively small pharyngeal space making good contact with the posterior pharyngeal wall at the level of the anterior tubercle, 12 years of age, no adenoids. *1*. At rest, *2*. Vocalizing "Uuuu…."

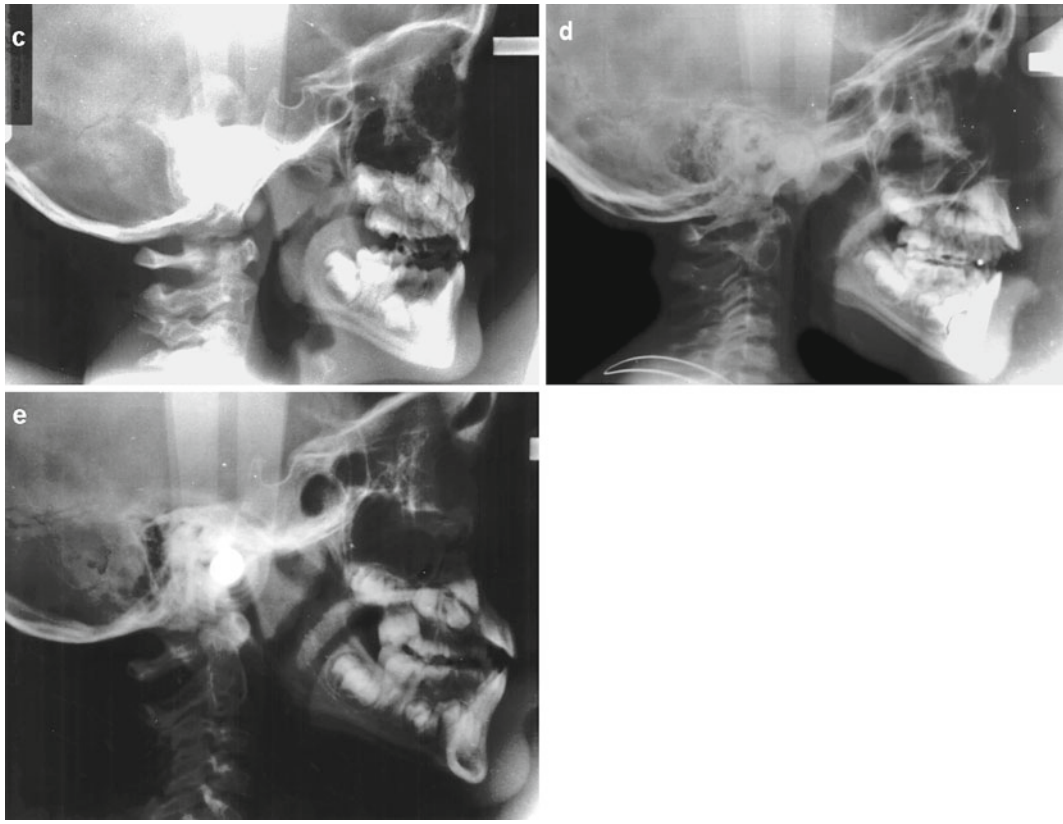

Fig. 33.8 (*continued*) (**c**) Cephaloradiographs taken at 4 years of age while vocalizing "Uuuu…." The small velum is seen making contact with large adenoid creating a shallow pharyngeal depth. (**d**) A small, thin paralyzed velum in a deep pharynx at 5 years of age. (**e**) A long velum of good thickness showing poor elevation resulting in a lack of contact with posterior pharyngeal wall. The velum does not elevate on vocalizing "Uuuu…." The velum may or may not make contact in sustained speech. That is why this record is not a functional test

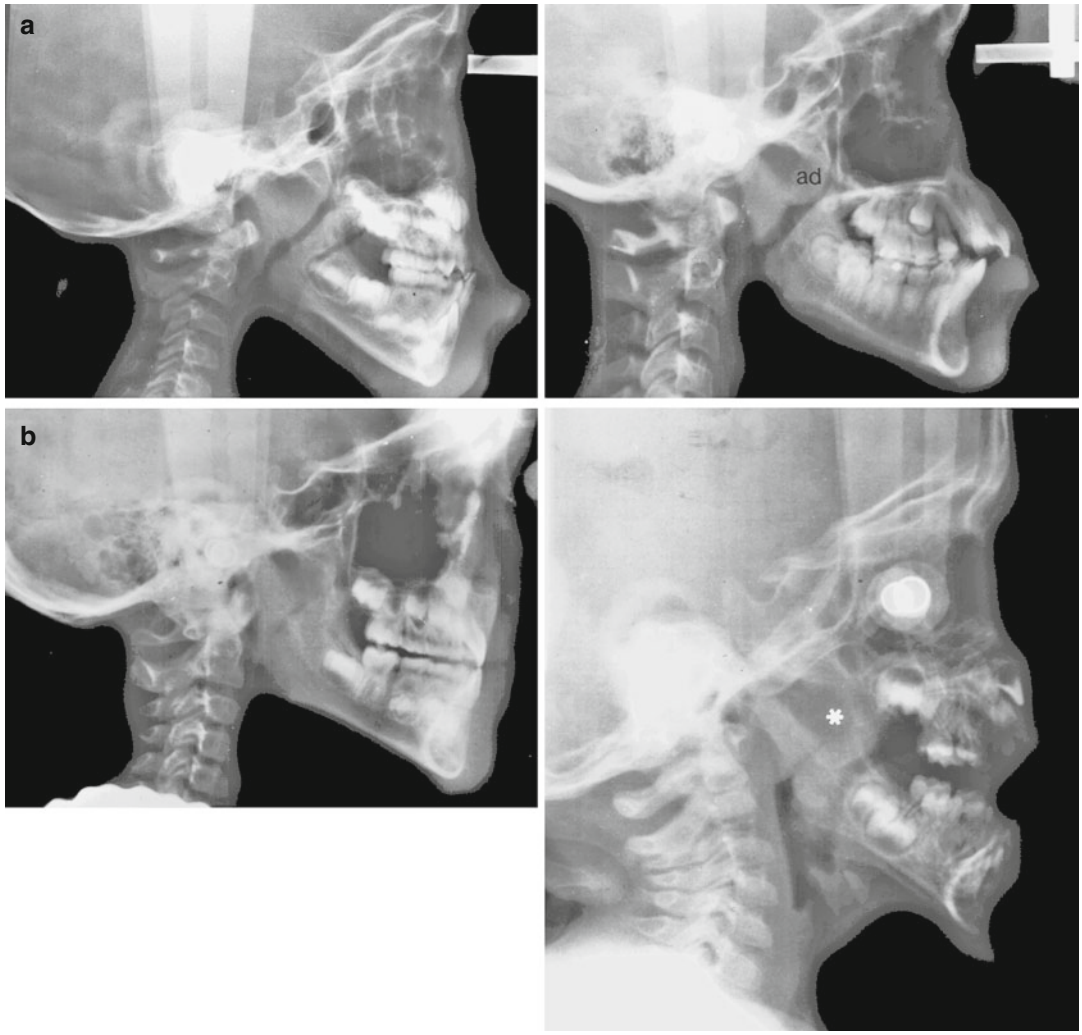

Fig. 33.9 (**a**, **b**) Variations in adenoid size and its effect on nasal air flow. (**a**) Partial nasal obstruction at 4 years of age (*left*) and 11 years of age (*right*). (**b**) Complete nasal obstruction shown at 8 years of age (*left*) and 2 years of age (*right*) (*, adenoid)

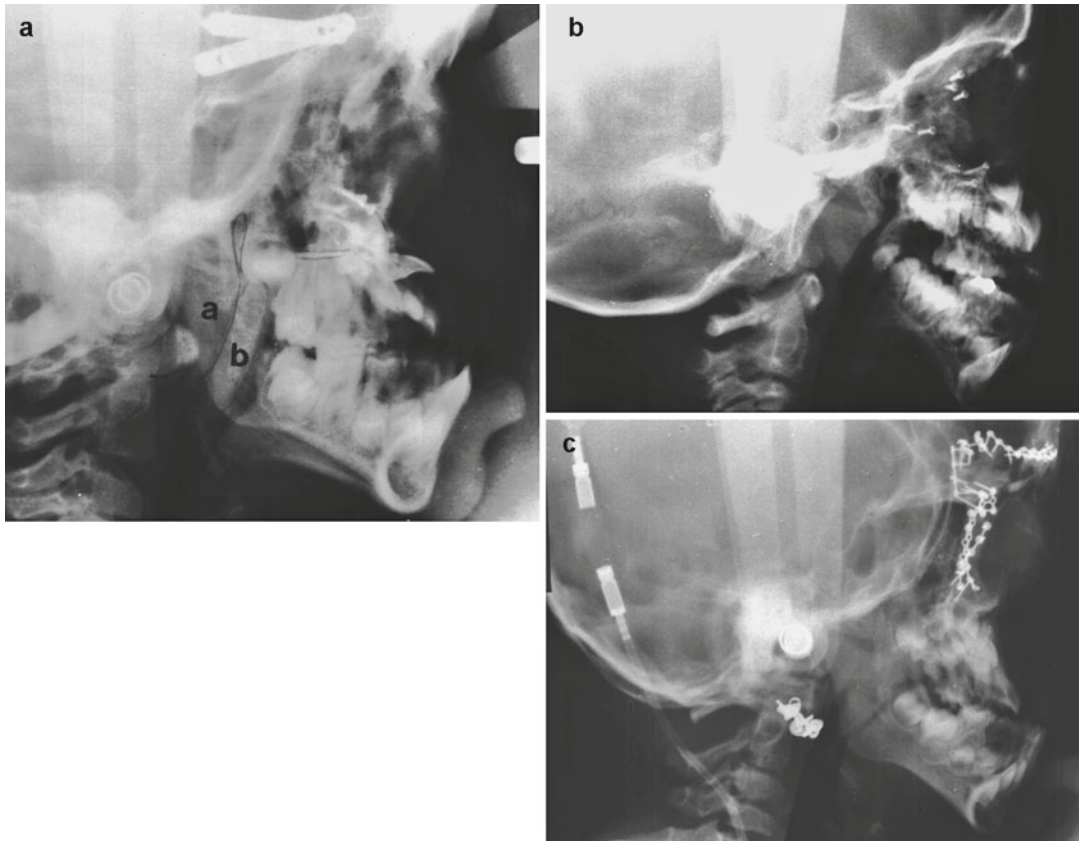

Fig. 33.10 (**a**–**c**) Nasal obstruction associated with midfacial hypoplasia and cranial base abnormalities in three different craniofacial syndromes: (**a**) Apert syndrome (*a* adenoid, *b* soft palate), (**b**) mandibulofacial dysostosis, and (**c**) Crouzon syndrome

33.8 Improving Velopharyngeal Closure

Traditionally, surgeons have focused on the symptom of hypernasality and have attributed it to inadequate palatal length. Accordingly, operations have been devised to address these structural deficits. Many procedures can be used to control air flow movements. Some have been successful; others have had limited success. The relationships among perceptual, anatomic, and physiologic variables have not been appreciated until relatively recently. To believe that hypernasality generally is the result of inadequate anatomy alone represents a simplistic view of VP function (Witt and D'Antonio 1993).

33.8.1 Pharyngeal Flaps

The pharyngeal flap is the surgical procedure most widely used to correct velopharyngeal insufficiency for speech. One end of the flap remains attached to the pharyngeal wall, while the other is sutured to the palate to occlude most of the velopharyngeal space. There are a number of variations of this procedure which involve where the flap from the posterior wall is inserted in the velum. It may be positioned midway on the superior surface, sandwiched (velar split) within the velum, or placed at the end and superior surface of the velum. The posterior flap also may be superiorly or inferiorly based (Figs. 33.11 and 33.12). Most surgeons and speech-language

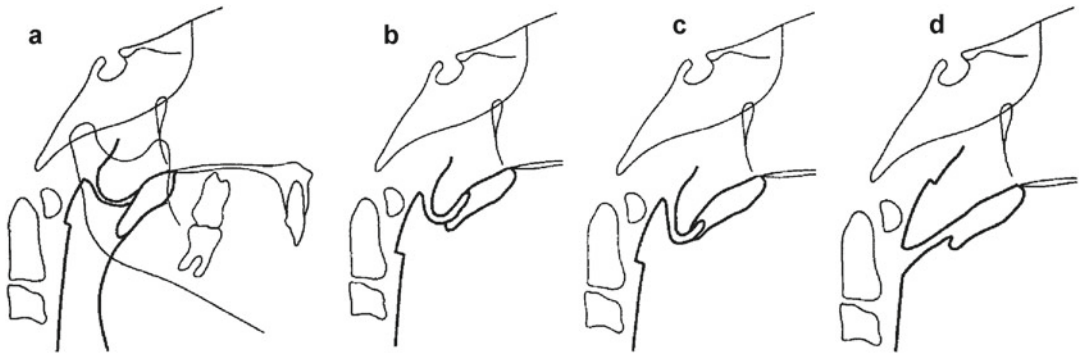

Fig. 33.11 (**a**–**d**) Variations in the placement of pharyngeal flaps. Superiorly based flaps are taken from the posterior pharyngeal wall above or at the anterior tubercle of the atlas. (**a**) Anteriorly superiorly based flap. (**b**) Posteriorly superiorly based flap. (**c**) Velar split – superiorly based. Flap is taken from the pharyngeal wall at the level of anterior tubercle. (**d**) Inferiorly based flap was taken from the posterior pharyngeal wall below the anterior tubercle of the atlas

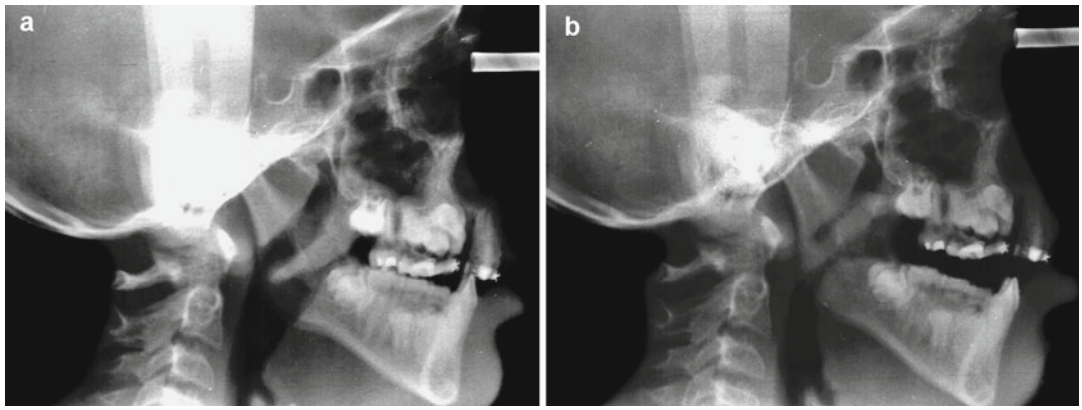

Fig. 33.12 Inferiorly based pharyngeal flap in the presence of a small adenoid. Even with good velar length and elevation, a pharyngeal flap was necessary because of poor lateral pharyngeal wall muscular movement. *Left*: At rest. *Right*: While vocalizing "Uuuu...." Inferiorly based flaps and/or flaps placed below the anterior tubercle of the atlas usually do not function adequately because that may not be the position for maximum lateral pharyngeal muscle action

pathologists seem to favor the superiorly based flap in which the base of the flap is positioned in line with the anterior tubercle of the atlas. This is the point where the velum in most adults makes contact with the posterior pharyngeal wall during normal speech.

The pharyngeal flap is an unphysiological substitute for the velopharyngeal mechanism because it acts as a sail to capture and channel most of the air through the mouth. The remaining pharyngeal port size will determine whether hypernasality or hyponasality will be present after surgery. Warren (1964a, b), analyzing aerodynamic pressure flow patterns in normal speech, determined that hypernasality occurred when the velopharyngeal port exceeded 10 mm^3 and that nasal escape of air was clearly evident at 20 mm^3. Bjork (1961), using basal radiographic techniques, estimated that a port size of 20 mm^3 in an area was the threshold between hypernasal and hyponasal speech.

Shprintzen et al. (1979) reported that the flap may have to be positioned more to one side than another according to the location of the port

space. With recent improvements in diagnostic abilities to determine the location and degree of air escape, nasopharyngoscopy has become the procedure of choice to predetermine the lateral position to place the pharyngeal flap. Unfortunately, surgical skill is not the sole determinant of the procedure's success, and patients still need to undergo extensive speech therapy.

Occasionally, a patient will remain hypernasal and continue to produce articulation errors in spite of a seemingly good surgical result. This can occur even when there seems to be a good speech mechanism, good muscular motion, and a good family environment.

An excessively wide flap can block off too much air flow into the nose, leading to mouth breathing and denasalized nasal consonants, and patients may suffer other side effects ranging from severe snoring to sleep apnea. As in normal speech, the opening and closing functions of the nasopharynx necessary for speech must be achieved by lateral wall movements in the nasopharynx. The lateral walls need to move medially to make contact with the flap during speech to prevent nasal escape of air.

A pharyngeal flap was also used in conjunction with push-back procedures at the time of palatoplasty, thinking that when combined, the retropositioned velum would be better maintained in its posterior position. Experience has shown that this should never be performed as a primary procedure because there is no evidence that velopharyngeal incompetency can be detected with certainty prior to speech production. Scar contracture resulting from push-back procedures not only inhibits palatal growth but causes any velar length increase to relapse. It does not seem wise to tether the push-back tissue with the pharyngeal flap because negative changes occurring to the palatal tissue can distort the flap.

In the treatment of cleft palate, the superiorly posterior-based pharyngeal flap is almost universally favored. Some clinicians are also using inferiorly based flaps in some cases (Millard 1963, 1980). It is still debated whether the superiorly or inferiorly based procedure is better. However, Trier (1985) points out there are compelling reasons why the superiorly based flap is employed more frequently. The inferiorly based flap not only has severe length limitations but also has the disadvantage of tethering the flap in an inferior direction, away from the palatal plane and motion for affecting VP closure.

Most speech-language pathologists tend to favor performing a pharyngeal flap at 4 years of age to avoid the problems of prolonged training. Early pharyngeal flap surgery carries no systemic risk of interference with facial growth but can cause some reduction in forward maxillary growth. It also can cause hyponasality and sleep apnea if it significantly obstructs air flow through the nose (Subtelny and Nieto 1978). The ability of patients to achieve proper velopharyngeal closure after pharyngeal flap surgery is unpredictable. Retrospective studies have shown that, when performed prior to 3 years of age, there are no significant morphological differences in the velopharyngeal area among the various cleft groups. Harding et al. (1975) and his colleagues advocated a superiorly based pharyngeal flap at a mean age of 6.5 years. Randall et al. (1978) advocated performing the procedure even earlier, between 2 and 3 years of age, especially when associated with a short, scarred, immobile palate with severe hypernasality and virtually no anterior articulation.

Failed pharyngeal flaps are generally due to the inability of the lateral pharyngeal walls to gain firm contact with the sides of the pharyngeal flap. This can result if the flap is too narrow and/or the lateral pharyngeal walls do not move far enough medially to reach it. In some instances, the flap may be set below the point of maximum medial movement, which is usually at the level of the hard palate. He also states that, under no circumstances, should a pharyngeal flap operation be performed simultaneously with other procedures.

33.8.2 Speech Aid Appliances

Wolfaardt et al. (1993) noted that the functional integrity of the palatopharyngeal valve can be compromised by a number of factors, including neurologic disorders such as stroke or head trauma; degenerative diseases such as multiple

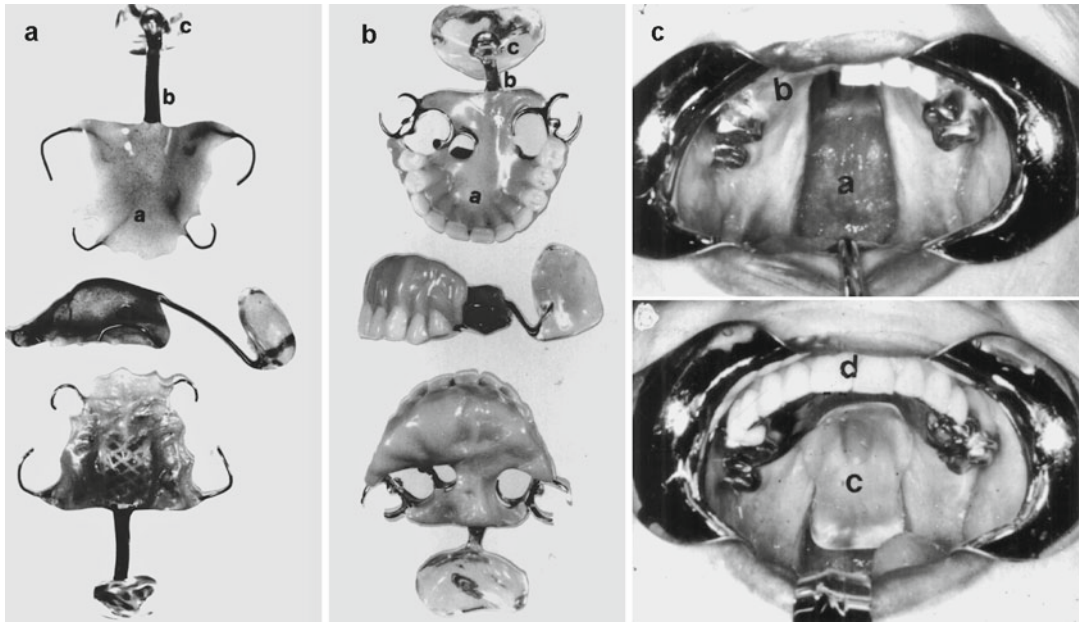

Fig. 33.13 (**a**–**c**) Speech aid appliances. In the event that surgery is contraindicated or delayed or, if performed, does not produce proper palatopharyngeal closure, a dental speech appliance may be substituted. Such appliances may be made of acrylic plastic retained by clasps about the teeth. Metallic castings of nonprecious and precious metals may also be used. (**a**) The appliance consists of three parts: (*a*) the palatal, (*b*) the velar, and (*c*) the pharyngeal sections. The palatal section (*a*) covers the palate and carries the clasps for attachment to the teeth. (**b**) The palatal section supports placement for the missing teeth and may also plump the upper lip to improve the facial profile. The velar section (*b*) is in the shape of a bar connecting the palatal section to (*c*) the pharyngeal portion of the appliance which extends into the nasopharynx and assists in palatopharyngeal closure and reducing nasal resonance. (**c**) When midfacial vertical dimension is lost (*top*) and must be increased, the palatal section will need to replace missing teeth and cover the natural teeth (*bottom*, wearing appliance). In such cases, the teeth need to be crowned in gold to protect them from dental caries. The unoperated cleft space (*a*) is covered with a posterior extended acrylic section (*c*) to help control air flow and permit proper feeding. (*d*) Replacement teeth. The pharyngeal section needs to extend into the nasopharynx and make contact with the posterior pharyngeal wall (Courtesy S. Pruzansky)

sclerosis, Parkinsonism, or bulbar polio; craniofacial birth defects, such as overt and covert clefts of the palate; and some behavioral disorders (e.g., functional articulation disorders).

In addition, in the absence of a palatopharyngeal control problem, resonance balance in speech also may be rendered aberrant by hearing loss or faulty sensorimotor learning patterns. These include impairments associated with congenital neurologic disorders (e.g., cerebral palsy) that impair a speaker's ability to regulate the articulatory dimensions of speech that influence the acoustical expression of nasality.

Prosthetic dental appliances are used for the replacement of missing or malformed teeth in the line of the cleft and, when indicated, in the design of speech appliances.

In the event that surgery is contraindicated, delayed, or performed unsuccessfully, a dental speech appliance may provide a satisfactory substitute. Such appliances are usually made of acrylic plastic and retained by clasps about the teeth. Metallic castings of nonprecious and precious metals also may be used in the design of speech appliances.

For purposes of description, the appliance consists of three parts: the (a) palatal, (b) velar, and (c) pharyngeal sections (Fig. 33.13). The palatal section may consist of little more than a covering for the hard palate and carry the clasps for attachment to the teeth. This section may also supply replacements for missing teeth. In cases in which vertical dimension must be increased because of arrest of vertical growth of the middle

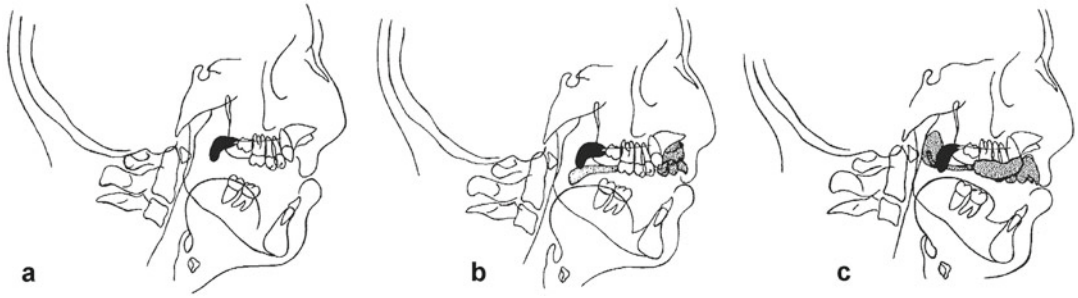

Fig. 33.14 (**a**–**c**) The pharyngeal bulb in a speech appliance is viewed in cross section in place in the nasopharynx. (**a**) Incompetent velopharyngeal closure. (**b**) Inadequate nasopharyngeal bulb. (**c**) Pharyngeal bulb (pharyngeal extension) is well positioned within the nasal chamber (Courtesy J.D. Subtelny)

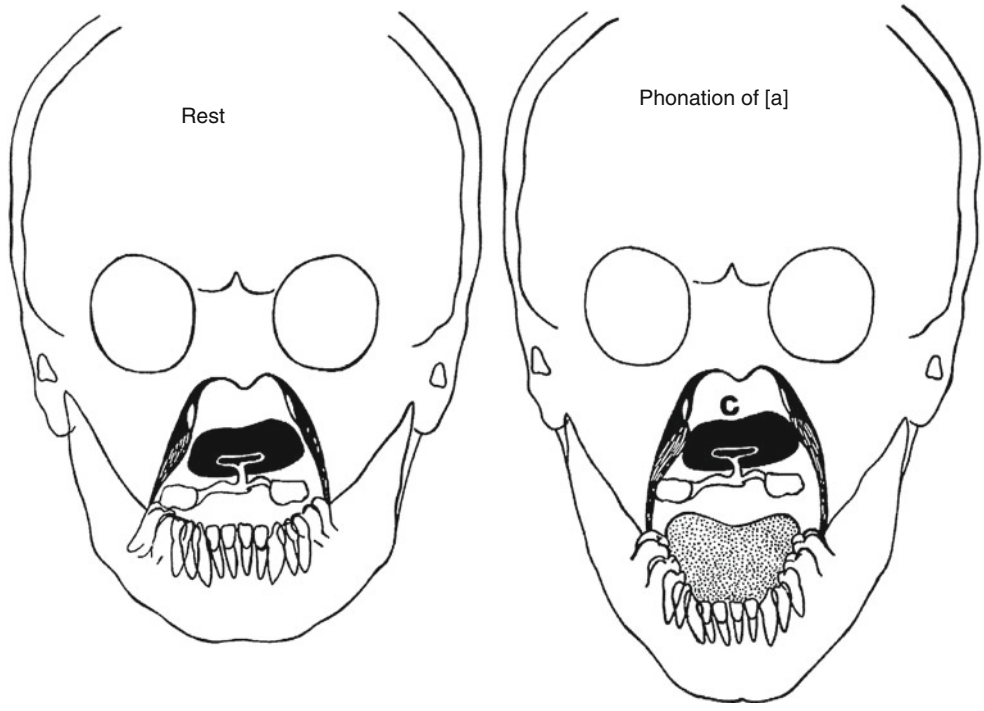

Fig. 33.15 The pharyngeal bulb on a speech aid appliance is viewed in the nasopharynx. At rest (*left*), a lateral space on either side of the bulb allows for normal nasal drippings to enter the mouth. When phonating /a/, (*right*) the lateral pharyngeal walls make contact with the speech bulb, reducing nasal air flow (Courtesy of J.D. Subtelny)

face, the palatal section may cover the natural teeth. In these circumstances, the teeth should be crowned to protect them from dental caries. By forward extension, the palatal section may also plump the upper lip and improve the facial profile.

The velar section is in the shape of a bar connecting the palatal section to the pharyngeal portion of the appliance. The pharyngeal portion extends into the nasopharynx and assists in palatopharyngeal closure (Figs. 33.14 and 33.15). The skill of prosthodontists in designing these devices has added a fortunate dimension to speech habilitation of cleft palate whenever surgery is unable to provide good results.

This treatment technique is logically combined with auditory training. The patient needs to learn to rely on auditory and intraoral somesthetic

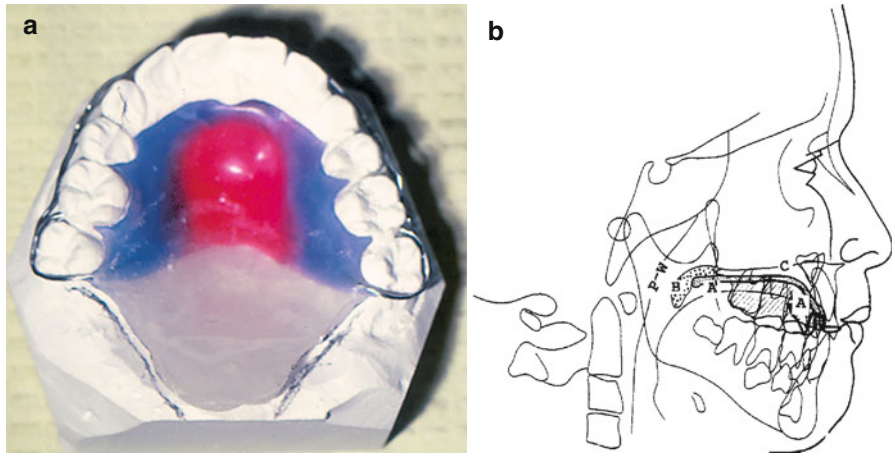

Fig. 33.16 (**a**, **b**) A palatal lift appliance. It is used to improve velopharyngeal function by improving velar muscle action. (**a**) The appliance on a dental model. The velar extension extends posteriorly to the uvulae. (**b**) Lateral cephalometric tracing showing the appliance (*A-A′*) velar section making contact up to the uvulae (*B*). The appliance can be well stabilized by using orthodontic bands with soldered buccal shelves to establish undercuts for the clasps

perceptions of resonance balance and palatopharyngeal control. Articulation training is likewise conducted to normalize consonant distortion or substitution patterns that patients may have developed as compensatory maneuvers in the face of palatopharyngeal incompetence.

Some speech-language pathologists and dentists utilize a palatal lift appliance (PLA) which is designed to hold the soft palate up and backward and to ultimately improve its function by acting as an isometric training appliance (Fig. 33.16) (Wolfaardt et al. 1993; Mazaheri and Mazaheri 1974). D'Antonio (1995, personal communication) suggests that the PLA be used for patients in whom there is adequate tissue but poor control of coordination and timing of VP movements. It is hoped that the degree of lift can be reduced gradually until the appliance can be discarded. Unfortunately, in many instances the prosthesis must be worn indefinitely. The use of a palatal lift appliance (PLA) is still controversial. Some believe that, at best, its use produces inconsistent results even with speech therapy. The palatal lift appliance (PLA) has also proved useful in reducing drooling and improving mastication and tongue movement. Speech therapy associated with use of a PLA may include any one or a number of techniques, many of which are practiced by the patient with and without the appliance in place (Wolfaardt et al 1993).

Auditory training should be used to enhance the patient's auditory awareness of normal and deviant resonance balance. Articulation training is used to develop a patient's palatopharyngeal control to regulate nasal resonance.

33.8.3 Push-Back Procedures (Velar Lengthening)

These are surgical procedures designed to lengthen the soft palate done either in conjunction with primary palatoplasty or as a secondary procedure after the palatal cleft has been closed and velopharyngeal incompetence has been detected. The most common surgical procedure is the V-Y push-back, commonly called the Wardill-Killner, Wardhill (1933, 1937) or Veau-Wardill-Killner procedure. There are a number of variations of this basic design. This procedure involves the use of two unipedicle or single-base flaps of palatal mucoperiosteum, the soft tissue on both sides of the cleft space. The flaps are elevated from the bony palate and retropositioned so that the cleft space is covered and the length of the soft palate increased. Push-back palatoplasty procedures

performed at an early age require an extensive amount of mucoperiosteal undermining and soft tissue displacement leaving large areas of denuded bone. These areas are later covered over by scar tissue which inhibits palatal bone growth and causes palatal deformation. This problem exists even when the push-back procedure is performed as a secondary procedure after the palatal cleft has already been closed.

Lateral cephalometric evaluation of this and other push-back techniques has shown that the net gain in soft palatal length is not as great as the amount of mucoperiosteum brought posteriorly because of contraction of scar tissue. Yet, some speech-language pathologists believe this technique yields a higher success rate, as measured by speech results, than any other surgical technique. Harding et al. (1977) criticized this procedure because large surfaces of denuded bone are left in the anterior palate, which result in excessive scarring and midfacial growth retardation. Other palatal lengthening procedures, such as Millard's Island Flap, have not been successful because the extensive mucoperiosteal shifting leaving large areas of denuded bone has been shown to have a deleterious effect on palatal development.

Witt and D'Antonio (1993) recommend Furlow's double-opposing Z-plasty for patients with a small central VP gap seen in submucous cleft palate or in patients with previously repaired cleft palate who demonstrate a midline "trough" or muscular diastasis.

33.8.4 Sphincteric Pharyngoplasty (SP) of Orticochea (1970, 1990)

No discussion of pharyngeal surgery could be complete without mentioning the Orticochea operation. Jackson (1985) described the procedure most commonly used today. This surgical procedure was designed to occlude the lateral ports by changing the lower insertion of the posterior pillars from the lateral walls to the posterior wall of the pharynx. Orticochea believed that this created a dynamic muscle sphincter of the pharynx, which could open and close dozens of times a minute. Furthermore, this sphincter has the same cerebrocortical role as the nasopharyngeal sphincter described by Passavant because the palatopharyngeus and superior constrictor muscles that are part of the Passavant nasopharyngeal sphincter are used in its constriction.

In patients with very short palates in whom there is a greater risk of dehiscence, only one of the palatopharyngeus muscles is transplanted. Three months later, in a second operation, the sphincter is completed by transplanting the second palatopharyngeus muscle.

The sphincter is created by transposing the posterior tonsillar pillars with the enclosed palatopharyngeus muscles from the lateral pharyngeal walls to the midsection of the posterior pharyngeal wall. Three openings conduct air between the oral and nasal pharynges. The central opening's shape and width remain unchanged postoperatively, with no reduction of the lumen. The two lateral openings completely disappear.

This sphincter tends to close off the nasopharynx. Some surgeons use it as a secondary corrective procedure.

Sphincter palatoplasty's cited advantages include (1) dynamic sphincteric closure as a result of retained neuromuscular innervation, (2) technical ease of execution, (3) a low complication rate, (4) nonobstruction of the nasal airway, and (5) no violation of the velum. Unfortunately, the question of whether it attains superior speech results is still open. It also must be emphasized that many surgeons have developed variants of the original procedure. The SP procedure has theoretic advantages and is an effective means of management of VPI for some patients. More studies need to be performed to determine when this or the pharyngeal flap should be the treatment of choice (Witt and D'Antonio 1993) .

33.8.5 Posterior Pharyngeal Wall Augmentation

Teflon and other materials have been implanted in the posterior pharyngeal wall with no reported long-term success. Potential problems include

tissue incompatibility and migration of the implant. Injectable forms of these substances also can create problems with respect to embolism or transport of the prosthetic matter to regional lymph nodes.

Although pharyngeal flap surgery is the treatment of choice in most cleft palate patients, Denny et al. (1993) believe that a posterior pharyngeal wall implant of costal cartilage should be used in cases of extremely shallow pharyngeal spaces with inadequate lateral pharyngeal wall motion and when the side effects of a pharyngeal flap should be avoided. Shprintzen et al. (1985) and Sher et al. (1986) believe that pharyngeal flaps should be avoided in patients with primary neuromuscular disease or pharyngeal hypotonia.

Pharyngeal implants have been carried out with a variety of materials by a number of surgeons over the years (e.g., Teflon, by Smith and McCabe (1977); Silastic by Blocksma (1963); Silicone Gel by Brauer (1973); Proplast by Wolford et al. (1989); and Homologous Cartilage by Trigas et al. (1988)).

Surgical procedures as well as injection techniques have been used. Furlow et al. (1986) reported one instance of obstructive sleep apnea (OSA) 6 years post-Teflon injection due to the downward displacement of the Teflon particles resulting in a narrowing of the patient's airway. In a series of studies, Vinas and Jager (1971) obtained the best results with surgically inserted cartilage implants.

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The Velopharyngeal Mechanism

34

Robert J. Shprintzen

34.1 Introduction

Normal speech is dependent on the modulation of voice and sound and the appropriate direction of airflow from the lungs as it travels through the vocal tract and the upper airway. One of the most important components of speech is the ability to separate the oral cavity from the nasal cavity, a process that is unique to humans (Shprintzen 2003). The majority of meaning in speech is delivered by the production of consonants (Shprintzen 2003), and the all but three English language consonants are produced orally with essentially no nasal resonance. The only consonant sounds that have significant nasal resonance in English (and most other languages) are /m/, /n/, and /ng/. All other consonant sounds require or involve the separation of the oral cavity and nasal cavity that is accomplished by the movements of the velum and pharyngeal walls. This process is referred to as velopharyngeal closure. Failure to achieve velopharyngeal closure has been variously referred to as velopharyngeal insufficiency (VPI), velopharyngeal incompetence, velopharyngeal inadequacy, and velopharyngeal dysfunction (VPD). Arguments over the validity of any or all of these terms are not really important, and for the purpose of this chapter, we will simply refer to the problem as velopharyngeal insufficiency (VPI), the most widely used and accepted term historically.

34.2 Normal Velopharyngeal Closure

The opening between the oral cavity and nasal cavity is demarcated anteriorly by the velum, posteriorly by the posterior pharyngeal wall, and laterally by the lateral pharyngeal walls. These muscular structures are multifunctional in that they are normally active during swallowing, speech, and other activities related to airway maintenance. The passageway connecting the oropharynx and nasopharynx, sometimes referred to as a "port" or "portal," is really a tube oriented in a relatively vertical manner (Fig. 34.1) (Shprintzen 1995). The opening and closing of this passageway between the oropharynx and nasopharynx actually represents a valve or valving mechanism, similar to other valves in the body that regulate the flow of air, blood, or other fluids by modulating the size of the opening. The

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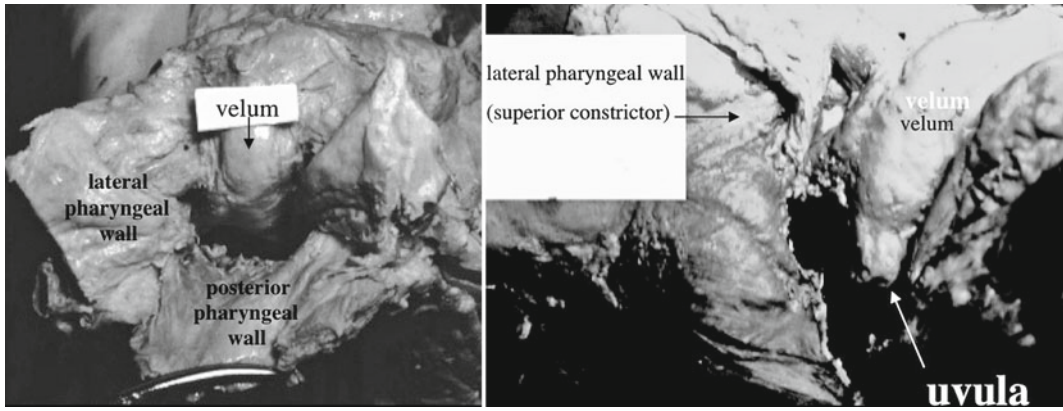

Fig. 34.1 Two views of the anatomy of the velopharyngeal valve. At left is an axial view from above showing what looks like a two-dimensional opening, but when the same specimen is viewed from a different angle (from the right posterolateral aspect of the pharynx), the vertical height of the velopharyngeal mechanism can be appreciated. Therefore, the velopharyngeal valve is a three-dimensional tube with width, breadth, and height

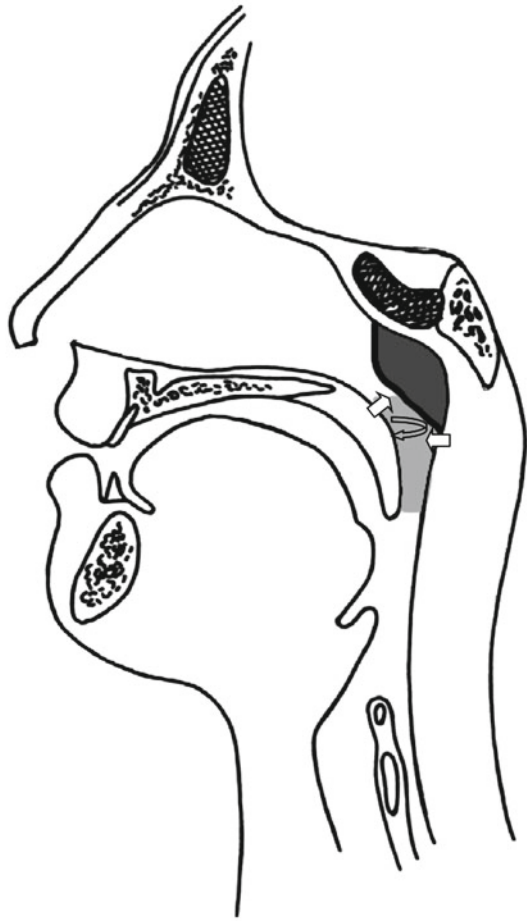

Fig. 34.2 Drawing showing the planes of movement necessary for velopharyngeal closure

movements for speech in the velopharyngeal mechanism are controlled by the motor cortex and are learned, just as the movements of the tongue, lips, and teeth are learned for speech. The movements of the same structures during swallowing are primarily reflexive and mediated by the brainstem. Velopharyngeal movements for swallowing are unrelated to those of speech, and the muscles used and timing of movements are different (Shprintzen et al. 1974). For the velopharyngeal valve to close during speech, there needs to be movement from the velum and the pharynx along three planes: the anteroposterior plane, the medial plane, and the vertical plane (Fig. 34.2). Anteroposterior movement is caused by the contraction of the levator veli palatini muscle which acts like a sling to draw the velum back toward the posterior pharyngeal wall. Depending on the individual craniofacial structure of the patient, that movement can be primarily posterior, primarily superior, or posterior-superior (Fig. 34.3). The medial movement of the pharyngeal walls is caused by contraction of the uppermost fibers of the superior constrictor pharyngeus. Lateral pharyngeal wall motion is highly variable ranging from barely detectable motion to lateral pharyngeal walls that approximate in the midline of the pharynx. Anterior motion of the posterior pharyngeal wall, usually referred to as Passavant's ridge, is also caused by contraction of the superior

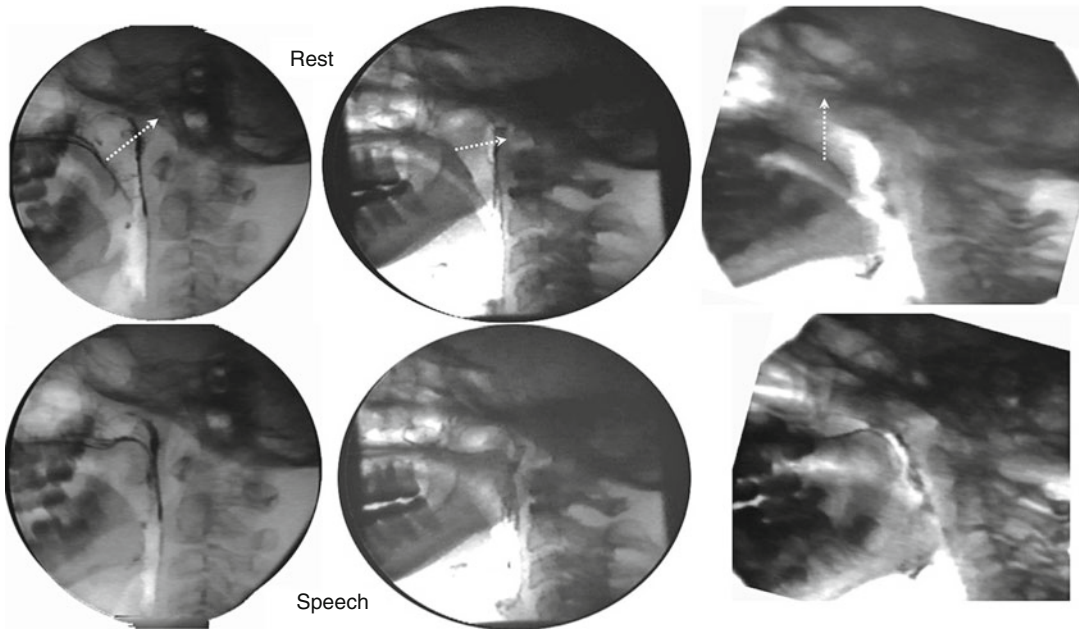

Fig. 34.3 Three lateral view videofluoroscopies showing the different types of movement of the velum that can occur depending on the orientation of the levator palatini

constrictor and can be variable in shape, size, and position (Fig. 34.4).

The variability of movement in the velopharyngeal valve from person to person was first described in a landmark article by Skolnick and colleagues (1973). They showed that some individuals have more lateral pharyngeal wall motion than others, some have Passavant's ridge and some do not, and that this variability of movement is true for both normals and people with VPI. Subsequent research in large samples of both normals and people with VPI confirmed this finding (Croft et al. 1981b). This variability may be related in part to structural differences from person to person, but learnng is also clearly a factor. The role of learning has been demonstrated experimentally by using visual nasopharyngoscopic biofeedback to alter patterns of movement (Siegel-Sadewitz and Shprintzen 1982), therefore indicating that it can be altered by learning new muscular movements.

Therefore, velopharyngeal closure is a voluntary, CNS-mediated motor activity that is learned during early speech acquisition. The role of diagnosis needs to be aimed at determining if a failure of the velopharyngeal mechanism to close completely during normally nonnasal speech is an anatomic or physiologic limitation versus an error in learning.

34.3 Failures of Velopharyngeal Valving

There are a substantial number of reasons why velopharyngeal closure might not occur during speech that normally requires it. VPI is usually associated with palatal anomalies, but the frequency of VPI in disorders unrelated to structural malformations of the palate is also very common. There are no definitive data available that help us to determine what the most common cause of VPI might be, but suffice it to say that it is a common disorder. For the purposes of this text, we will focus primarily on clefting anomalies, but the reader should be aware that problems with the velopharyngeal valve associated with other disorders require careful scrutiny, as well.

Disorders of velopharyngeal function might best be divided into three major categories: structural,

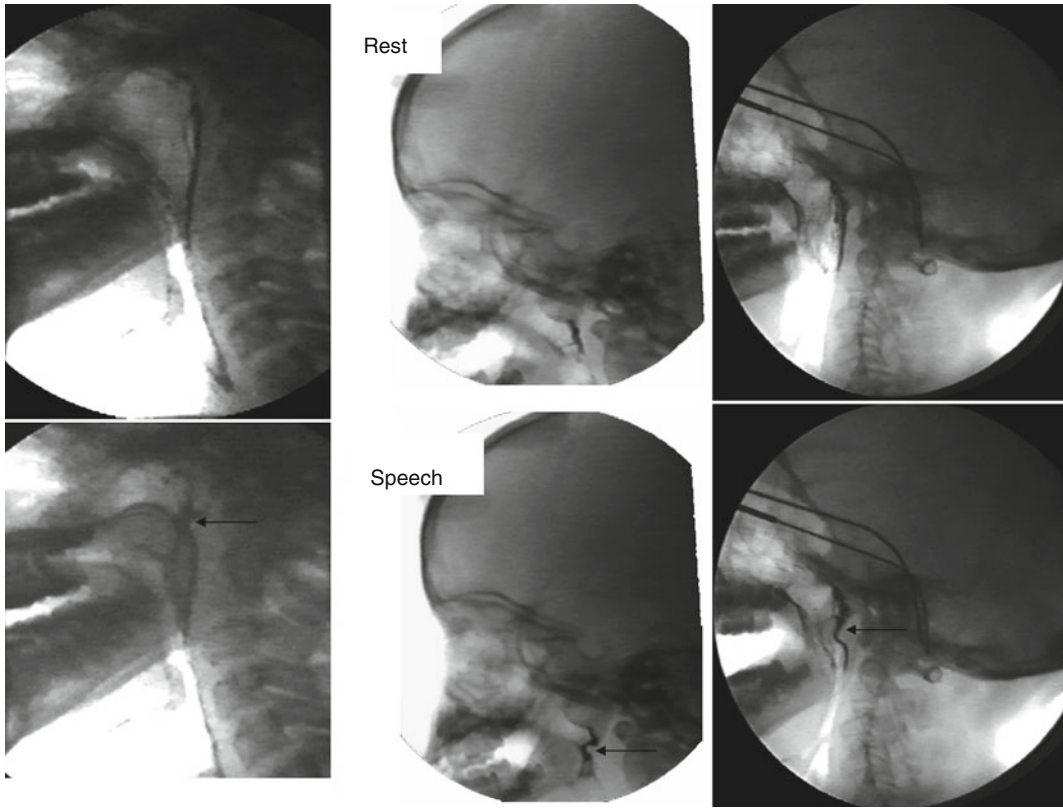

Fig. 34.4 Several different examples of Passavant's ridge (*arrow*)

neurogenic/myopathic, and learned. These categories are not mutually exclusive. In other words, it is typical to have only one dysfunction causing the problem, but it might also be possible to have two or three of these factors in operation simultaneously.

34.4 Disorders of Structure

Structural anomalies include clefting of the palate (in all of its variations), anomalies of symmetry of the palate and/or pharynx, and anomalies of the surrounding pharynx (including abnormalities of the lymphoid tissue). Cleft palate is a broad designation for a broad spectrum of anomalies that range from complete clefts of the lip and palate to nearly an undetectable malformation known as occult submucous cleft palate. However, all of these anomalies could result in the same disorder of VPI.

34.4.1 Overt Clefts

Overt clefts of the palate are the most easily recognizable form of palatal anomaly and may range from complete bilateral clefts of the hard and soft palate to a cleft of the posterior aspect of the soft palate (Fig. 34.5). The extent of the cleft varies in both syndromic and nonsyndromic cases. The frequency of velopharyngeal insufficiency does not really vary according to cleft type, but cleft type does vary according to the primary diagnosis. In nonsyndromic clefts where there are no associated anomalies to affect velopharyngeal closure, the frequency of VPI after repair is probably primarily related to a combination of surgical variables and the amount of muscle deficiency associated with the cleft. However, in syndromic cases that account for more than half of all clefts (Shprintzen et al. 1985a, b), other syndromic features can contribute to VPI. For example, in velo-cardio-facial syndrome, besides cleft palate, there are three other

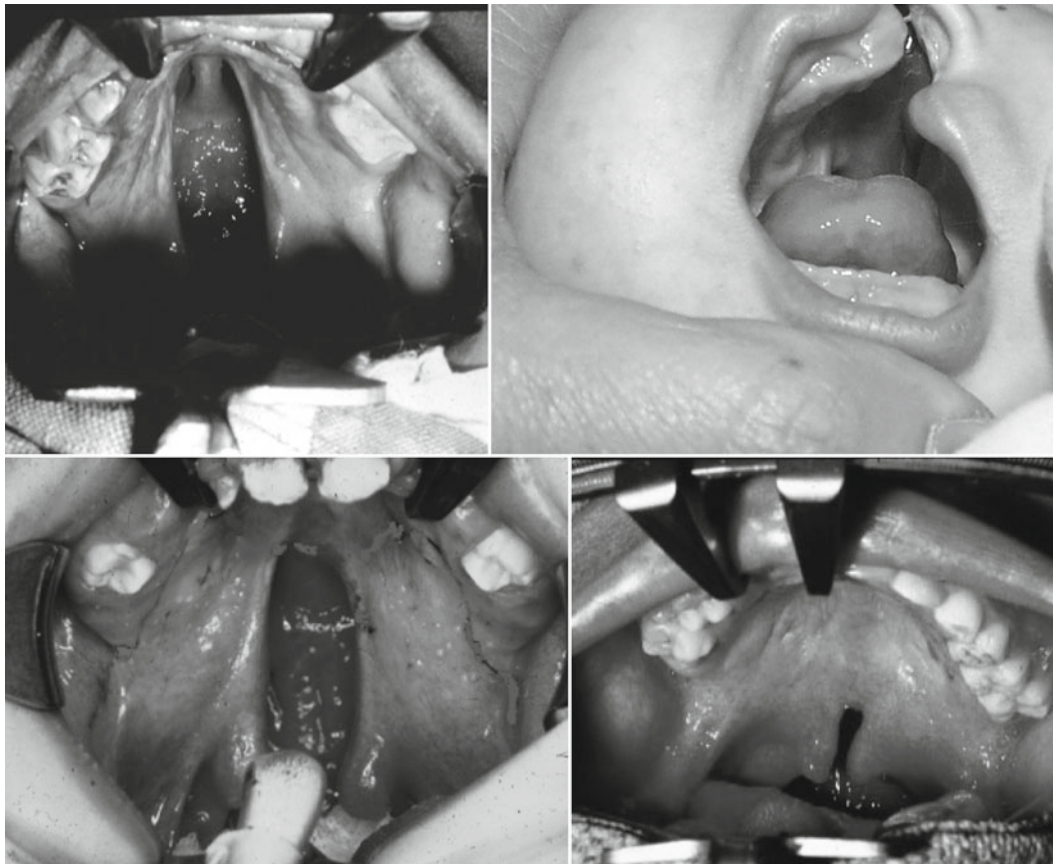

Fig. 34.5 Four different types of overt palatal clefts, including bilateral complete cleft of the hard and soft palate (*top left*), unilateral cleft of the hard and soft palate (*top right*), cleft of the entire soft palate (*bottom left*), and a cleft of the posterior third of the soft palate (*bottom right*)

clinical findings that contribute to VPI, none of which are mutually exclusive. Individuals with VCFS are known to have a high frequency of pharyngeal hypotonia (Zim et al. 2003), absent or hypoplastic adenoid tissue (Williams et al. 1987), and platybasia (Arvystas and Shprintzen 1984) that can increase pharyngeal depth. Therefore, primary cleft repair in VCFS almost always fails to resolve VPI (Shprintzen 2001).

Surgical repair of the palate is important for only one reason, to provide the patient with the possibility of developing normal speech. Feeding, ear infection, growth, etc.—none of these are factors of importance to consider in relation to the need for palatoplasty. Babies with clefts feed quite successfully when proper technique is used, as will be discussed briefly later. In fact, because palate repair is not usually completed until approximately 1 year of age, babies must feed over that period of time with a cleft. Middle ear disease is common in babies with clefts but is related to malformation of the Eustachian tube rather than muscular action of the velum (Shprintzen and Croft 1981). Middle ear disease can also be successfully managed medically or with myringotomy and tubes. Although feeding and hearing can be normal with an unrepaired cleft palate, speech cannot. Achieving acoustically acceptable speech is dependent on normal resonance and the ability to produce pressure consonants orally, both highly dependent on velopharyngeal closure.

Surgical repair of clefts are highly successful when they are accomplished prior to 18 months of age, and most surgeons in the USA are timing primary palatoplasty at or before the first birthday in

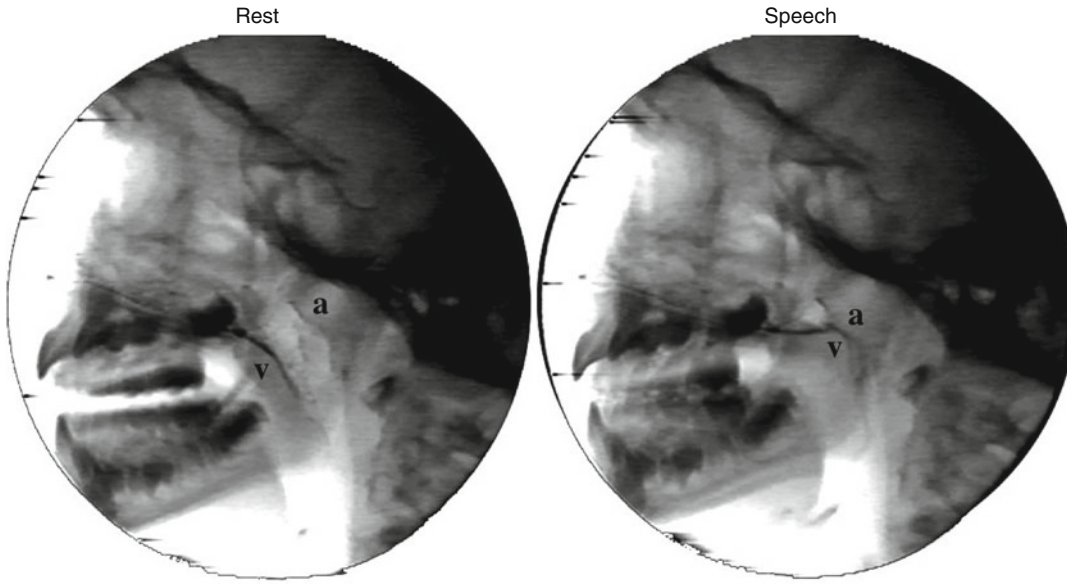

Fig. 34.6 Lateral view videofluoroscopy in a normal 4-year-old child demonstrating that the velum (*v*) never contacts the posterior pharyngeal wall during speech but instead approximates to the adenoid (*a*)

order to intercept the early phases of speech development. It is clear that the majority of children who are born with overt clefts do not have VPI following primary palatoplasty (Salyer et al. 2006). Although surgical approaches to cleft repair have varied in relation to technique and timing, the majority of experienced surgeons who complete primary palatoplasties prior to 18 months of age achieve between 80 and 90 % positive outcomes defined as an absence of VPI after palatoplasty. Although this leaves a core of patients who will require secondary surgical procedures at a later date to resolve VPI, the overall percentage of individuals with VPI who have repaired clefts is probably quite low in most treatment centers.

The logical conclusion regarding the successful outcome of palate repair is that it is highly dependent on surgical technique. Actually, this has never really been proven, and there are many factors involved, including the age at time of surgery, the type of operation, other anatomical variations in the pharynx, muscle tone and developmental factors, and one factor that has received very little attention in the literature, the surgeon and his or her skill level. It is not within the scope of this chapter to discuss each of these issues, but it is important to show that the successful outcome of palate repair is dependent on more than one factor.

Although the palate is an important component of velopharyngeal valving, it is not the only one. The palate, and the velum in particular, is simply one component of the velopharyngeal mechanism, and there is quite a bit of variability of the size and shape of the other structures within the pharynx. The height and width of the pharynx varies from person to person just as height, weight, and head circumference do. The size of the adenoid is of particular importance because the adenoid sits in the plane of velopharyngeal closure in young children. In fact, velopharyngeal closure in children until at least 6 years of age is actually velar-adenoidal (Williams et al. 1987) (Fig. 34.6). Adenoids vary considerably in size, and there is some evidence that suggests that adenoid size may be the most important factor for successfully achieving velopharyngeal closure (Havkin et al. 2000). The larger the adenoid, the less movement is required of the velum. Although the adenoids are important to normal speech production, the tonsils are not and will be considered separately later in this chapter. It is clear that adenoidectomy should be avoided (unless there is severe upper airway obstruction in which case partial adenoidectomy may be indicated) in individuals with cleft palate or submucous cleft palate because of the potential for the development of the VPI once the adenoid

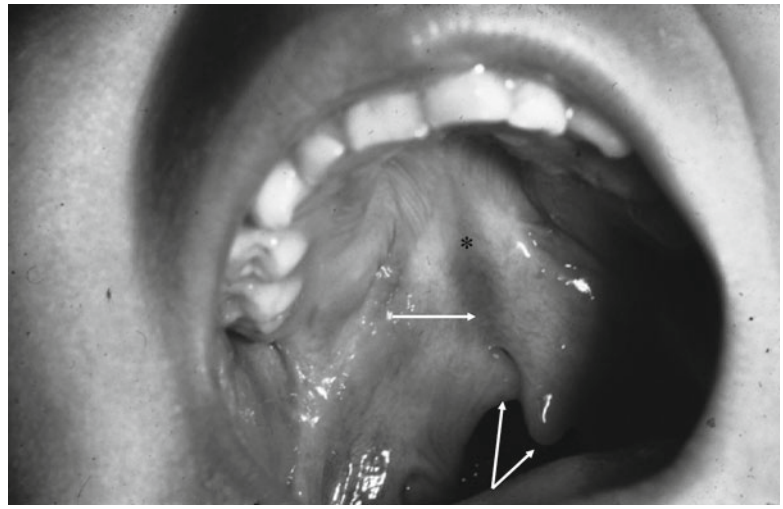

Fig. 34.7 Obvious submucous cleft palate showing a bifid uvula (*dual arrow*), a notch in the posterior border of the hard palate (*) and zona pellucida (*single arrow*)

tissue has been removed having the effect of increasing the volume of the nasopharynx (Croft et al. 1981a, b; Havkin et al. 2000).

Another important factor is the volume of the nasopharynx. An increased volume of the nasopharynx can be caused by an abnormally obtuse angulation of the skull base (Shprintzen 2001). Many clinicians who use lateral view radiographs of the head and neck may interpret an increased nasopharyngeal volume as a "deep pharynx." Alternatively, because the palate appears short within the presence of an increased pharyngeal volume, some clinicians may interpret this as a "congenitally short palate." As will be discussed later, structural integrity of the palate cannot be assessed without a direct endoscopic view of the nasal surface of the velum. However, the depth of the nasopharynx can be assessed radiographically using the standardized measurements provided by a lateral cephalograph. Neither procedure (endoscopy or cephalometrics) is individually perfect for describing the anatomy of the pharynx. Rather, the information obtained from each is complementary.

34.4.2 Submucous Cleft Palate

Submucous cleft palate has traditionally been defined as the triad of bifid uvula, notching of the posterior border of the hard palate, and a visible diastasis of the musculature in the soft palate referred to as a zona pellucida (Fig. 34.7) (Calnan 1954). However, although all cases with these clinical findings have submucous cleft palate, not all people with submucous cleft palate have all three of these findings. Some have two of these findings, usually a bifid uvula and hard palate notch, some have only one of these findings, almost always a bifid uvula, and more interestingly, some have none of these findings. Submucous cleft palate is simply a milder or less obvious expression of cleft palate. In this case, the skin, or mucous membrane, is intact but the underlying musculature is not and some muscle may be congenitally absent (Croft et al. 1978; Lewin et al. 1980). Of interest, although people with submucous cleft palate do have velopharyngeal insufficiency and hypernasal speech, the large majority do not (Shprintzen et al. 1985a, b). This is interesting in light of the fact that many surgeons perform palatoplasty in infancy, before the onset of speech in an attempt to avoid speech problems by rearranging the muscles to a more normal configuration in the palate for individuals with submucous cleft palate even though the large majority of people with submucous clefts have normal speech without such surgery. This may indicate that other factors besides muscle integrity play a large or even larger role in normal speech production than simply the degree of palatal normality.

34.4.3 Occult Submucous Cleft Palate

In 1975, Kaplan reported an abnormality of the palate he labeled the occult submucous cleft palate

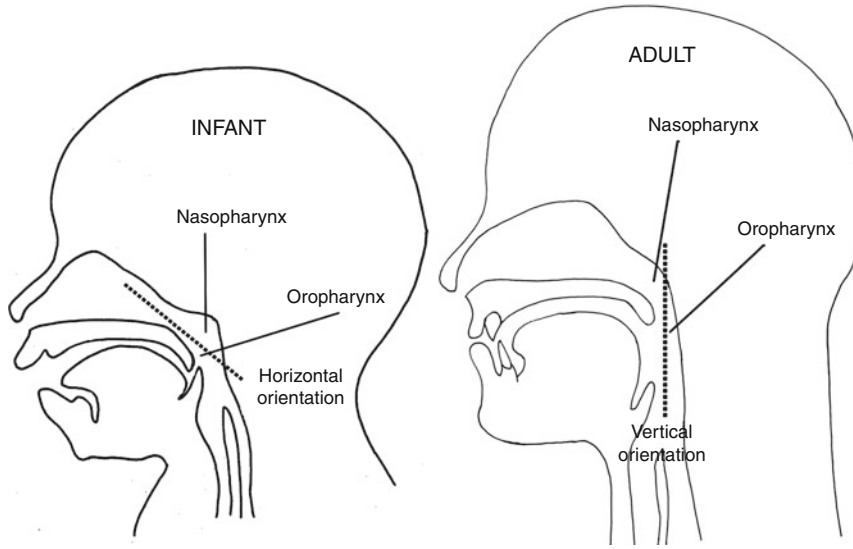

Fig. 34.8 Comparison of the upper airway in an infant (*left*) to that of an adult (*right*) showing a more vertical orientation of the pharynx in the adult compared to the more horizontal position in the infant

(Kaplan 1975). Kaplan's report described four individuals with VPI who had normal appearing palates on oral examination, without evidence of bifid uvula, a zona pellucida, or notching in the hard palate. At surgery for correction of their VPI, he dissected the palate and studied the musculature noting that the muscle fibers were abnormal in position and orientation as might be found in a cleft palate or submucous cleft palate, hence the label *occult* submucous cleft palate (occult meaning mysterious). He concluded that the only way one could detect this anomaly was by surgical dissection. However, subsequent publications have demonstrated that occult submucous cleft palate can be detected by nasopharyngoscopy (Croft et al. 1978; Lewin et al. 1980; Shprintzen 1979a, b, 2000). Unfortunately, many clinicians and researchers have not applied this diagnostic approach so that many cases of occult submucous cleft go undetected and the frequency of clefting becomes significantly underestimated. It is interesting to note in retrospect that three of the four patients shown by Kaplan clearly have velocardio-facial syndrome, a condition known to have a high frequency of occult submucous cleft palate (Shprintzen 1982). It is not known what the frequency of hypernasal speech is in association with occult submucous cleft palate because it is very unlikely that individuals with normal speech will ever have the anomaly detected unless nasopharyngoscopy is performed for some other reason.

34.5 Normal Anatomy of the Velopharyngeal Mechanism: Developmental Changes

The structures of the palate, nasopharynx, and oropharynx (the structures that constitute the velopharyngeal mechanism) are not static. They undergo significant alteration and remodeling over time relative to their functions and age. Therefore, mechanisms of velopharyngeal function at one point of time in life may not be the same as another. This means that broad generalizations about how the palate and pharynx function should be avoided because they may be age dependent. As mentioned previously, the palate and pharynx are multifunctional. Modulating the size and shape of the upper airway, sometimes referred to as the aerodigestive tract, is necessary for proper function during speech, feeding, and airway maintenance. However, these functions vary with age. Infants have the ability to separate the oropharyngeal airway from the nasopharyngeal airway making it easier for them to breathe through their noses while they eat than is true for adults. This is because the upper airway in infants is vertically shorter and oriented in a more horizontal than vertical angle (Shprintzen 2003) (Fig. 34.8). The shorter upper airway in infants allows the epiglottis to be in close proximity to the soft palate creating a conduit for milk to flow unobstructed into the

pyriform sinuses laterally while the baby breathes through the nose. Because infants' ventilatory efforts are faster and more frequent than adults and their smaller lung capacity and blood volume makes them more prone to rapid oxygen desaturation than adults, it is important for them to breathe uninterrupted during feeding to prevent hypoxia and hypercarbia. Babies typically eat semi-reclined, a position that actually makes the upper aerodigestive tract more vertical so that feeding is facilitated by gravity and an easy flow of fluids into the hypopharynx and esophagus.

As children grow, growth in the maxilla and mandible is both horizontal and vertical. This expands the upper airway vertically and draws the palate and velum away from the epiglottis and larynx. The entire pharynx expands in three dimensions (width, depth, and height) as the entire palate grows in length. In fact, if the upper airway did not increase in height, the palate would hang too deeply into the pharynx resulting in upper airway obstruction. In fact, in some genetic disorders with midfacial anomalies, such as Crouzon syndrome, Apert syndrome, and Down syndrome, where the maxilla is hypoplastic in all dimensions, the phenomenon of the velum contributing to upper airway obstruction is commonplace, resulting in obstructive sleep apnea as well as obstructive daytime respiration.

As the pharynx becomes more vertical with age, the adenoids begin to involute. As reported previously, if the adenoid did not involute, the pharynx would become obstructed because of its change in shape and orientation (Shprintzen 2003). This is because the hard and soft palates actually migrate to a position relatively closer to the posterior pharyngeal wall over time. If the adenoid remained large (typically occupying about 25–50 % of the postnasal space before involution begins between 8 and 10 years of age), the palate would be flush against it, thereby compromising the nasopharyngeal airway.

The changes in the shape, volume, and position of the upper airway have been speculated to potentially result in changes in velopharyngeal function over time. Clinically, I have often heard professionals suggest that gradual adenoid involution can result in the development of velopharyngeal insufficiency. Although there are a very small number of anecdotal cases reported with a late onset of velopharyngeal insufficiency (Mason and Warren 1980), such events are very rare, and I have never encountered such a case in more than 38 years of practice with the exception of late onset muscular dystrophies, myasthenia gravis, or other neuromuscular diseases.

34.6 Other Structural Disorders

Although the adenoid is important to normal speech production, other lymphoid tissues do not play a role and may be detrimental to normal speech production. There are three other masses of lymphoid tissue within the oral and pharyngeal airway: two palatine tonsils and one lingual tonsil. The lingual tonsil sits at the base of the tongue anterior to the vallecula and plays no role whatsoever in speech production. When enlarged, the lingual tonsil can cause the sensation of globus, or a feeling that something is stuck in the bottom of the throat. If very large, a muffled resonance may also result from an enlarged lingual tonsil because it may physically damp the sound waves produced by the vocal cords just millimeters below the base of the tongue. However, the lingual tonsil does not cause hypernasality or hyponasality and is too far away from the velopharyngeal valve to interfere with it.The palatine tonsils, however, may alter nasal resonance patterns as has been documented in the medical literature on several occasions (Henningsson and Isberg 1988; MacKenzie-Stepner et al. 1987; Shprintzen et al. 1987).

The palatine tonsils are situated in the posterior oral cavity with their attachment in the faucial arch in between the anterior and posterior tonsillar pillar. Assessment of tonsillar size is often relegated to oral assessment with a flashlight and tongue blade. It is typical for clinicians to use a rating scale between 0 and 4+ based on the degree of medial excursion of the tonsils toward the midline of the posterior oral cavity with 0 implying that the tonsils are not visible, 1+ referring to tonsils fully contained within the faucial pillars, 2+ meaning the tonsils extrude medially beyond the pillars but less than halfway to midline, 3+ indicating that the tonsils extend more than half of the way to midline, and 4+ meaning they are touching in the midline (kissing tonsils).

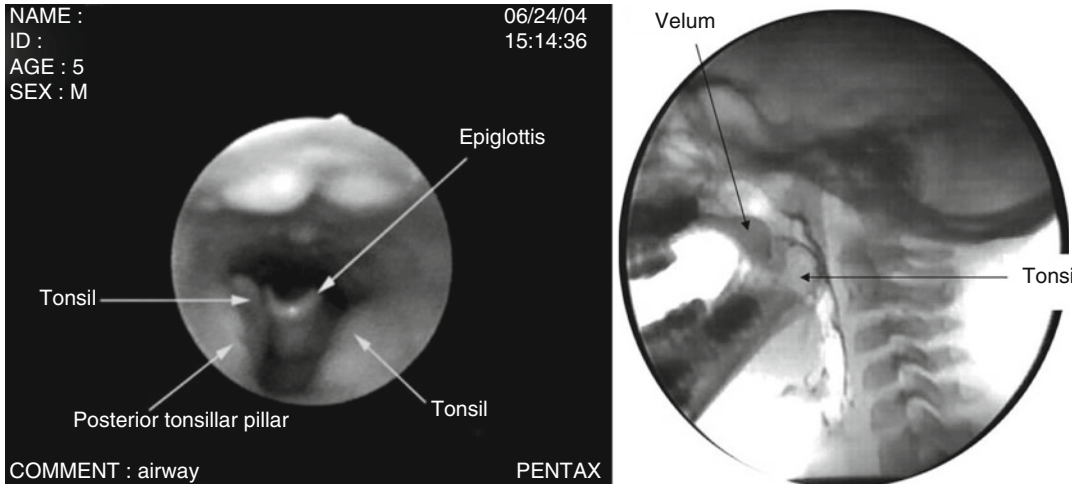

Fig. 34.9 Hypertrophic tonsils seen endoscopically and on lateral view radiographs showing that they intrude posteriorly into the pharyngeal airway behind the faucial pillars

The problem with this scale is that no one has told the tonsils that if they get big that they must grow medially. In fact, in many cases, they grow back into the pharyngeal airway and may grow down into the hypopharynx or up into the nasopharynx (Fig. 34.9). If the tonsils extrude into the pharynx, they may interfere with velar elevation or lateral pharyngeal wall motion, resulting in VPI (MacKenzie-Stepner et al. 1987; Shprintzen et al. 1987). In patients with cleft palate, this may present as a particularly difficult diagnostic dilemma and potentially dangerous circumstance. Because VPI is relatively common in children with cleft palate, the presence of hypernasality or audible nasal air escape during speech may lead clinicians to conclude that the problem is an anatomic problem of the palate, and therefore a recommendation for pharyngeal flap or other surgery may be made. However, if the tonsils are large and contributing or causing the problem, performing surgery in the pharynx may be unnecessary, but even more important, may prompt the development of obstructive sleep apnea. Tonsils have been shown to be a primary factor in inducing obstructive apnea following pharyngeal flap surgery (Shprintzen et al. 1992a, b) because they sit beneath the flap and narrow the oropharyngeal airway, while the flap compromises the nasal airway. It has been reported that if pharyngeal flap is indicated, tonsillectomy or adenotonsillectomy should be performed first in order to avoid obstructive sleep apnea as a complication (Chegar et al. 2007; Shprintzen et al. 1992a, b). In some cases, tonsillectomy may resolve the VPI if the tonsils were the primary cause for limiting movement in the velopharyngeal valve (Shprintzen et al. 1987).

34.7 Neurogenic and Myopathic Disorders of Velopharyngeal Function

VPI is a common disorder among individuals with disorders of the central nervous system or who have primary myopathies. Although the frequency of VPI is high, the true prevalence is not known because many of these individuals have other severe disorders of speech, and the VPI is simply a component of a broader spectrum of speech abnormality that may be very severe. In such cases, resolving VPI alone will do little or nothing to improve the intelligibility of speech, and therefore such individuals are not often referred for diagnosis or treatment. Neurogenic disorders that might frequently result in VPI are strokes and other cerebrovascular accidents, open and closed head trauma, cerebral palsy resulting

from neonatal hypoxia or brain malformation, infections such as encephalitis, and neurodegenerative diseases, such as myasthenia gravis. Primary myopathies that have VPI as a common finding include (but are not limited to) Steinert syndrome (myotonic dystrophy), facioscapulohumeral muscular dystrophy, nemaline myopathy, and oculopharyngeal muscular dystrophy. The management in myopathies is very different than in cleft palate or other structural anomalies of the head and neck. Surgical intervention is often problematic in myopathies because of respiratory concerns, and even the operation itself may be compromised by severe adverse anesthesia reactions, particularly the development of malignant hyperpyrexia which can be a fatal complication (Shprintzen 1997). Therefore, the diagnosis of myopathic disorders and the recognition of hypernasal speech as a potential clinical feature are critical for proper clinical management.

34.8 Evaluation of Velopharyngeal Insufficiency

Two issues established in the previous pages point out the need for detailed and comprehensive evaluation of VPI. First, velopharyngeal valving is highly variable from person to person, both in normals and in those with velopharyngeal insufficiency. Therefore, assumptions cannot be made about the nature of velopharyngeal valving based on the sound of speech or preconceived notions about how the mechanism works for the "average" individual. Second, VPI occurs in many different conditions, and the reason for the VPI may not always be obvious, including structural anomalies that cannot be seen on oral clinical examination. The diagnosis of more obscure yet common conditions such as occult submucous cleft palate, tonsillar hypertrophy, or myopathies is highly dependent on a precise diagnostic approach.

In general, procedures used to assess velopharyngeal insufficiency can be divided into two broad categories: direct and indirect. Direct procedures can be defined as those that allow direct visualization of the velopharyngeal valve during the unimpeded flow of speech typical for the individual while not interfering with the movements of the valve. Indirect procedures can be defined simply as everything else. Indirect procedures typically measure some type of artifact or by-product of velopharyngeal valving rather than assessing the act itself. Indirect procedures consist of listener judgment of resonance and nasal airflow, instrumental techniques for measuring nasal resonance, and techniques for measuring nasal airflow or pressure. Direct procedures include both radiographic and endoscopic techniques, although ultrasound and real time CT and MRI have all been proposed to study VPI. However, only fluoroscopic and endoscopic procedures have stood the test of time and remained.

34.9 Indirect Assessments of VPI

Every diagnostic process starts with an assessment of the way a patient sounds to the human ear. Parents may perceive something is wrong with the way the child's speech sounds, or if the child has a repaired cleft, a clinician schedules periodic assessments to determine if speech will develop normally. At some point, however, someone has to perceive abnormal hypernasality or nasal air escape during speech. The listener must determine if resonance is normal, hypernasal, or hyponasal. Occasionally, there may be mixed hyper-/hyponasality if there is VPI in association with nasal blockage at some location anterior to the velopharyngeal valve. In addition to resonance, the listener must determine if there is an abnormal escape of air during speech and, if so, on what sounds and how often it is occurring. If the speech pattern is deemed to be abnormal, then the necessity for treatment is determined. In other words, perceptual assessment becomes the entry point for further evaluation and is therefore the most critical portion of the assessment.

Once a patient's speech has been determined to show the effects of VPI, then the clinician must choose what to do next. The clinician may choose to confirm nasal air escape. This can be done in a number of ways from very simple to very complex.

Simple procedures would include the use of a nasal mirror, a listening tube or stethoscope, or devices that might detect nasal air escape such as the SeeScape®. All of these devices are used to confirm the presence of abnormal nasal air escape during the utterance of normally nonnasal sounds, words, or phrases. They are "yes vs. no" devices rather than quantitative instruments. Although sensitive to nasal air escape, they are not specific as to the source of the air escape. In individuals with clefts, the air may be leaking from a fistula even if there is no VPI, thus resulting in a false positive. False negatives are common. Nasal congestion or a deviated septum will block air escape from the nostrils even if VPI is present resulting in a false negative. Therefore, procedures for detecting nasal air escape may be decent "quick and dirty" techniques for identifying nasal air escape, but they are not really diagnostic for VPI in many cases. Perhaps the easiest procedure for a "quick and dirty" technique is to use nostril occlusion during normally nonnasal speech. Occluding the nostrils during a normally nonnasal speech sample should result in no change of resonance if velopharyngeal closure is being achieved. If there is VPI, then pinching the nostrils shut will result in a cul-de-sac nasal resonance that will cause an immediate change in resonance pattern. If nasal air escape is audible during spontaneous speech, occluding the nostrils will eliminate the nasal turbulence. Although this procedure is not quantitative in any way, it is a simple procedure for making a quick determination of the potential presence of VPI.

There are several devices that have been designed as quantitative. These include manometers and rhinometers that measure pressure from the mouth or nose, but these devices have been proven to be irrelevant in relation to speech production. Two procedures have garnered some attention from their use in research and some clinical settings: the nasometer and pressure-flow instrument. The nasometer and pressure-flow device both apply numbers to nasal air escape during speech based on resonance (nasometer) or the pressure of air coming from the nose and mouth (pressure flow) during speech. The nasometer uses microphones to compare resonance between the nasal and oral cavities during speech, providing a number (nasalance, a ratio of acoustic energy from the nose compared to the mouth), while pressure-flow instrumentation utilizes a mathematical computation, the theoretic hydraulic principle, to calculate an orifice size in mm^2. Although both of these devices provide numerical data, one should not equate a number with "objective" data. Albert Einstein was reputed to have a sign hanging in his office at Princeton that said, "Not everything that counts can be counted, and not everything that can be counted counts." Further, relevant to the hydraulic principle calculation, Einstein also said, "As far as the laws of mathematics refer to reality, they are not certain; and as far as they are certain, they do not refer to reality" and "If the facts don't fit the theory, change the facts." In other words, validity of a measure has more to do with its relevance than anything else. The nasometer is a device for detecting resonance patterns, but the ultimate tool for this is the examiner's ear. It is the perception of normal versus abnormal that is important. If the ear perceives speech as normal, it matters not what a machine assigns as a numerical value. The second and third quotes from Einstein reinforce a primary problem with pressure-flow instrumentation. The entire principle behind pressure flow is that the opening detected in the velopharyngeal valve is measured in mm^2. As was emphasized earlier in this chapter and as is shown in Fig. 34.1, the velopharyngeal valve is a three-dimensional structure, not two dimensional. A three-dimensional object needs to be measured in mm^3, not mm^2. This misinterpretation of velopharyngeal valving that serves as the basis for pressure-flow studies renders its calculations as irrelevant. Some clinicians use nasometers and similar instruments for biofeedback in order to obtain comparisons following treatment or during the application of some forms of speech therapy, but definitive data in terms of effectiveness of this approach is not available.

34.10 Direct Assessments

34.10.1 Multi-view Videofluoroscopy

It is interesting to note that the two gold standards for direct assessment of velopharyngeal valving

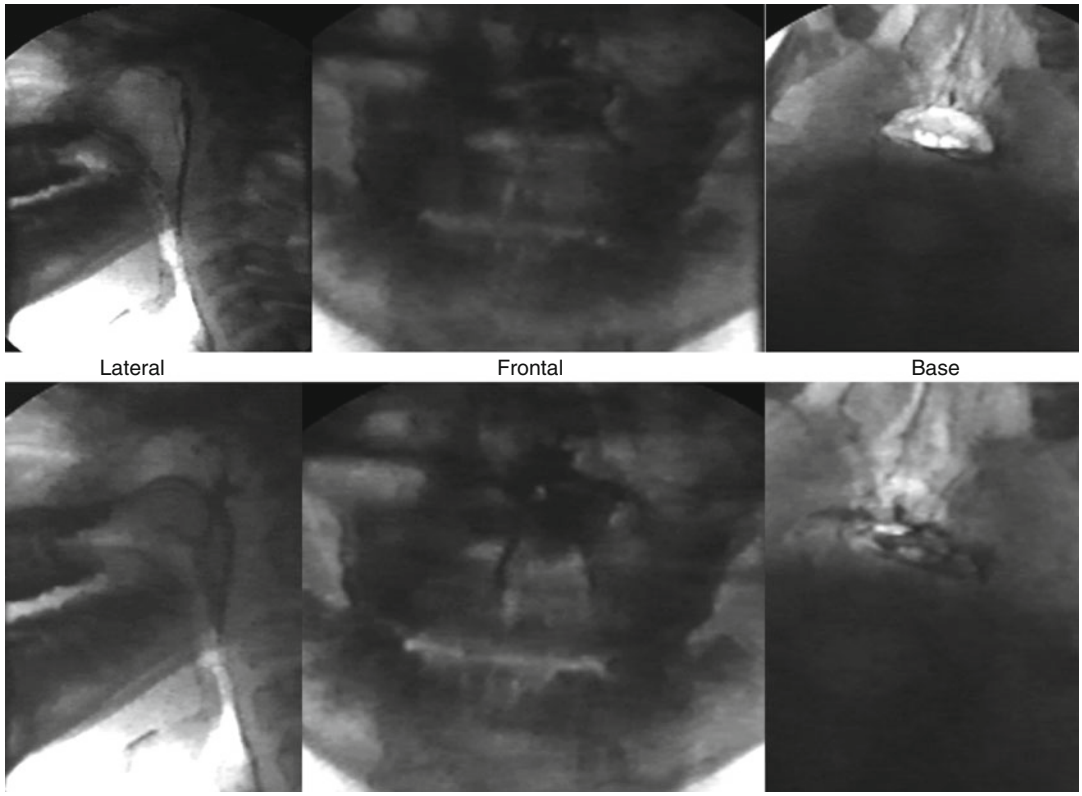

Fig. 34.10 Lateral, frontal, and base radiographic views at rest and during speech in an individual with a repaired cleft and hypernasal speech

were both initially described in the same year. Multi-view videofluoroscopy and nasopharyngoscopy were both described initially in 1969, and although there have been improvements in instrumentation and minor changes in technique, the procedures have remained relatively unchanged since that time. Both procedures are used widely and were recognized by an international working group as being critical to the proper assessment of VPI (Golding-Kushner et al. 1990).

In 1969 and 1970, Skolnick described the use of three radiographic views, the lateral (midsagittal), frontal (P-A), and base (axial) views, using barium contrast to coat the pharyngeal soft tissues (Skolnick 1969, 1970) (Fig. 34.10). Although the views were not performed simultaneously, the use of a standardized speech sample serially provided a three-dimensional assessment of the pharynx that accounts for width (frontal and base views), depth (lateral and base views), and height (lateral and frontal views). In addition, all of the oral articulators can also be seen simultaneously. Since the early descriptions of this technique, several other radiographic views have been added, including oblique views and Townes view (Shprintzen 1995). According to the International Working Group, the two views essential to assessment are the lateral and frontal (Golding-Kushner et al. 1990), and other views may be added if they provide useful additional information. A standardized speech sample is used to limit the duration of radiation exposure while allowing an observation of all components of speech including nonnasal sounds, nasal sounds, syllable repetitions, a sustained fricative, nasal to nonnasal transitions, and the natural flow of spontaneous speech. I have used the following sample to meet these criteria:

Ma-ma-ma
Pa-pa-pa
Ta-ta-ta

Fig. 34.11 Arrangement of equipment for videonasopharyngoscopy. Two television monitors are used to allow both the patient and the examiner to see the examination simultaneously. This helps to fix the attention of the patient and allows the examiner to explain the observations while the examination is taking place

Ka-ka-ka
/sssssss/ (sustained /s/)
Suzy
Suzy sees Sally
Stop the bus
Catch a fish
Jerry's slippers
1-2-3-4-5-6-7-8-9-10 (counting to ten)

The speech sample is rehearsed with the patient just prior to the procedure to be certain that it is repeated correctly and in a conversational manner. Repeating this sample for three radiographic views limits the procedure to a minute or less, thus keeping radiation exposure to a minimum. Because the neck is irradiated during this procedure and a child's thyroid gland is sensitive to radiation, it is preferred to limit videofluoroscopy to a one-time evaluation, usually just prior to secondary surgical intervention.

Immediately prior to the procedure, approximately 1 ml of barium sulfate suspension is instilled in each nostril using a dropper or pipette. This is done with the patient supine on the fluoroscopic table. This allows coating of the velum, lateral pharyngeal walls, and posterior pharyngeal wall. Without barium contrast, the lateral pharyngeal walls cannot be seen in frontal view, and the base or Townes view will not provide an en face view of the actual velopharyngeal valve. In addition, even in lateral view, the barium contrast is essential for defining small gaps. If there is no contrast used, as soft tissues converge, they blur together and a false negative for VPI is possible.

34.10.2 Nasopharyngoscopy

Nasopharyngoscopy for the assessment of the velopharyngeal valve was described almost simultaneously with the advent of multi-view videofluoroscopy. Pigott described the use of both flexible and rigid instruments in 1969 (Pigott 1969; Pigott et al. 1969). Although Pigott continued to advocate for the use of rigid endoscopes in the years that followed (Pigott and Makepeace 1982), the development of smaller caliber flexible instruments clearly won support for fiber-optic endoscopy because of the ability to examine even very young children without significant pain or discomfort (Miyazaki et al. 1975; Shprintzen 1979a, b; Shprintzen et al. 1979). Today, fiber-optic endoscopes have external diameters well under 3 mm, many in the 2-mm range, therefore allowing easy access to the upper airway (Fig. 34.11). The International Working Group recommended flexible fiber-optic nasopharyngoscopy and further recommended that the examination should include assessment of the entire vocal tract from the tip of the nose to the larynx (Golding-Kushner et al. 1990). It was also recommended that all examina-

tions be recorded with both video and sound for future review (Golding-Kushner et al. 1990). It has been shown that group review of studies is the most reliable and valid method for coming to diagnostic and treatment decisions because it allows for discussion of interpretations of the procedure (D'Antonio et al. 1989).

The procedure is best done using topical anesthesia, especially in young children, in order to avoid discomfort so that an adequate speech sample is obtained. My preference is to use topical tetracaine hydrochloride 2 % aqueous solution (Pontocaine®) inserted into the nose on cotton packing rather than a spray. Lidocaine is also effective. Using a pair of bayonet forceps, the cotton packing can be passed deeply into the nose to numb the turbinates and septum which are the sensitive portions of the nose. The packing is left in place for several minutes and then removed and the endoscope inserted. Using packing, the anesthetic is kept against the nasal mucosa for several minutes rather than a brief spray which really has minimal effect. In addition, the spray may reach deeper into the vocal tract and be inhaled, whereas the packing keeps the effect limited to the nasal mucosa. Once the endoscope is passed into the pharynx, it should not be held stationary throughout the examination. The advantage of a flexible instrument is its flexibility that allows movement so that both sides of the pharynx can be seen and it can be passed into the nasopharynx, oropharynx, and hypopharynx. The larynx, tonsils, and tongue can be seen in this manner.

The same speech sample used for videofluoroscopy is utilized for nasopharyngoscopy, as well, but the sample does not need to be limited to those sounds and phrases. Because there is no ionizing radiation involved, there is no need to limit the procedure to a minute or less. If phonemic variation is seen, the examiner is free to have the patient say more sounds and phrases than for fluoroscopy.

34.11 Which Direct Visualization Procedure to Use?

Many clinicians utilize nasopharyngoscopy exclusively as an assessment of VPI because they think that the information provided is the equivalent of videofluoroscopic procedures. However, it has been reported that there is an approximate 30 % discrepancy between findings using multi-view videofluoroscopy and nasopharyngoscopy primarily in the rating of lateral pharyngeal wall motion (Shprintzen 1995). Therefore, both procedures are essential for a complete assessment of VPI (Golding-Kushner et al. 1990). Each procedure has its own advantages and disadvantages as follows:

Multi-view videofluoroscopy advantages

1. Provides a direct physiologic view of all components of velopharyngeal valving in all dimensions (width, depth, and height).
2. The entire vocal tract can be seen at one time.
3. All articulators are seen simultaneously.
4. The forward floor of speech is not impeded.
5. Excellent compliance even in very young children.
6. The procedure is painless.

Multi-view videofluoroscopy disadvantages

1. It is not a direct anatomic view, relying on radiographic shadows for information.
2. Ionizing radiation is used.
3. As a result of ionizing radiation, there is a time limit to the procedure.
4. Interpretation may be difficult for those inexperienced with the technique.

Nasopharyngoscopy advantages

1. Provides a direct anatomic view usually appreciated by surgeons.
2. The entire vocal tract can be seen, but not simultaneously.
3. The normal flow of speech production is not impeded.
4. There is no time limit to the procedure so speech can be manipulated in more detail.
5. There is no ionizing radiation.
6. The procedure should be painless when done properly.

Nasopharyngoscopy disadvantages

1. Anatomic structures moving in front of the endoscope may block the view of physiological movements.
2. A lack of compliance occurs in some cases, particularly in young children.
3. The oral articulators cannot be seen.
4. The entire vocal tract cannot be seen at the same time.

5. The procedure can be painful in some cases or cause bleeding if not done properly.

34.11.1 Reporting of the Results

The International Working Group also published guidelines for the standardized reporting of the observations from endoscopies and fluoroscopies (Golding-Kushner et al. 1990). A ratio scale was developed that would assess the amount of movement of a structure (the velum, right lateral pharyngeal wall, left lateral pharyngeal wall, and posterior pharyngeal wall) toward its opposing structure. This approach was an attempt to provide a method that would communicate the degree of movement, not the actual distances moved, that would be more standardized than qualitative measures such as "poor," "fair," "good," or "excellent." I have used this system for more than two decades to transmit information to surgeons performing reconstructive surgery, and the ratios provided have met with a great deal of success in terms of the ability to communicate between clinicians, especially those from diverse fields of practice, such as surgeons, speech pathologists, and otolaryngologists.

34.12 The Importance of It All

There is a common "Murphyism" (one of the humorous attempts at expanding Murphy's Laws) that goes something like this, "Before ordering a diagnostic test, determine what you will do if the result is positive or the result is negative. If the answer is the same for both, don't do the test." In other words, the value of collecting such elegant diagnostic information is to have it guide treatment. This has been amply demonstrated in a number of studies that have documented the efficacy of guiding surgical management based on diagnostic information from multi-view videofluoroscopy and nasopharyngoscopy (Argamaso et al. 1994; Chegar et al. 2007; Shprintzen et al. 1979; Tatum et al. 2002). Stated another way, it has become obvious that there is not one single operation that can be successful in all cases. A velopharyngeal gap during speech is a hole that needs to be located in three-dimensional space, and its size relative to the total size of the pharynx must be confirmed in order to know how to occlude it. The listener's ear becomes the final judge of success, just as it was the entry point into the diagnostic protocol. With the combination of direct visualization procedures and an understanding of how to structurally solve the problem, the elimination of VPI should be achieved in more than 90 % of cases (Chegar et al. 2007; Shprintzen et al. 1979; Tatum et al. 2002).

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Surgical Management of Velopharyngeal Dysfunction

35

Richard E. Kirschner and Adriane L. Baylis

The primary goal of cleft palate repair is to provide for the development of normal speech, a goal which is dependent upon the successful establishment of the functional and structural integrity of the velopharynx. It has been estimated that between 10 and 35% of patients will demonstrate persistent velopharyngeal dysfunction (VPD) following cleft palate repair. The velopharyngeal valve is a complex, dynamic structure that serves to separate the oral and nasal resonating chambers during speech production. VPD typically results in a combination of hypernasal resonance, audible and inaudible nasal emission (with or without turbulence), weak pressure consonants, and, in most pediatric cases, specific types of articulation errors (known as compensatory articulation errors) (Kuehn and Moller 2000). Some patients may also develop dysphonia (a rough, breathy vocal quality), as a consequence of vocal hyperfunction to compensate for VPD (D'Antonio et al. 1988). In infants with overt clefts or VPD, nasal regurgitation (leakage of food or liquid through the nasal cavity) may be present, although this diminishes significantly after the first year of life. The perception of speech symptoms consistent with VPD should be confirmed by instrumental assessment to confirm the etiology, size, and consistency of the VP gap, as well as the movement of the velum and pharyngeal walls during speech.

Successful surgical management of VPD is critically dependent upon precise diagnosis and careful selection of operative technique. In the context of overall cleft habilitation, successful management is also critically dependent upon functional interdisciplinary collaboration. Management of VPD is best conducted in an interdisciplinary team context due to the multifactorial nature of this disorder. Patients with confirmed or suspected VPD require a thorough clinical evaluation by a plastic surgeon, speech-language pathologist, and, in many cases, a geneticist. Occasionally, a maxillofacial prosthodontist is also part of this team if nonsurgical treatment is required. Each of these professionals should have training and experience in the area of cleft palate and craniofacial anomalies.

The role of the speech-language pathologist on this team is to conduct a comprehensive assessment of the patient's speech including perceptual speech evaluation of intelligibility, resonance, nasal emission, voice quality, articulation, and overall speech acceptability, as well as instrumental evaluation of VP function for speech.

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The speech pathologist collaborates in decision-making regarding the presence and severity of the VPD and in formulating a tentative hypothesis for the etiology of the VPD. In addition, the speech pathologist determines the impact of any articulation errors on the function of the VP mechanism and whether (1) the patient is a candidate for imaging during speech or (2) additional speech therapy is needed prior to imaging. When indicated, the geneticist performs a physical exam and obtains a thorough family history and medical history to determine whether specific laboratory testing is indicated. In many cases, VPD occurs in the context of a broader syndrome, especially in "non-cleft" patients (e.g., 22q11.2 deletion syndrome). With the advent of faster, more specific, and more cost-effective genetic testing, as well as an appreciation of milder phenotypes of syndromes, genetic testing is employed prior to surgical management of VPD in a growing number of cases.

The plastic surgeon conducts a craniofacial and oral exam, confirms perceptual symptoms of VPD, obtains a thorough medical history, and evaluates for upper airway obstruction and surgical risk factors. The surgeon participates in imaging of VP function during speech to confirm the diagnosis, extent, and nature of VPD and to select and plan surgical intervention. The primary aim of surgical management is to establish velopharyngeal competency while avoiding the complications associated with nasal airway obstruction. The choice of surgical procedure must be individualized, taking into consideration the specific anatomy and function of the velopharyngeal valve, as well as associated comorbid conditions. The surgical procedures employed most commonly in the management of VPD include the Furlow double-opposing Z-palatoplasty, posterior pharyngeal flap, and sphincter pharyngoplasty. Augmentation of the posterior pharyngeal wall has been used less frequently and will not be discussed in this chapter.

35.1 Preoperative Assessment

Careful preoperative assessment is paramount to optimizing speech outcome while minimizing complications. Selection of the best surgical procedure and the specific technical details of its performance requires a thorough understanding of the anatomy and physiology of the dysfunctional velopharynx. A thorough medical history should be obtained, including an assessment of prior surgery. The patient-family interview is an important component of preoperative assessment, which includes a review of:

- Current concerns with speech
- Current concerns with feeding/swallowing, including leakage of food/liquid through the nose
- Primary medical diagnoses (e.g., cleft palate, syndrome, other birth defects, neuromuscular disease)
- Pregnancy history, complications, medication use, and any exposure to teratogens
- Birth and delivery history and complications
- History of any breastfeeding, swallowing, or other feeding difficulties during infancy, including leakage of food/liquid through the nose
- Developmental history
- Speech therapy history
- History of hearing loss or middle ear disease, including history of frequent ear infections or effusions
- History of snoring, restless sleep, fatigue, or symptoms consistent with sleep apnea
- Surgical history including prior tonsillectomy, adenoidectomy, and, if appropriate, cleft-related surgical history and timing, etc., as well as history of any surgical-related complications or risk factors (e.g., bleeding disorders, anesthesia reactions)
- History of any past genetic testing and results
- Family history of cleft lip/palate, nasal speech, speech delay, or articulation/pronunciation difficulties; hearing loss; learning disabilities; and other medical conditions

Syndromic diagnoses and comorbid conditions should be noted, as they may have a significant impact both on the success of the surgical procedure and on the incidence of postoperative complications. When appropriate, additional preoperative lab testing and medical and anesthetic consultation should be obtained. Patients with a history of upper airway obstruction, including those with Pierre

Robin sequence, also require careful assessment of the upper airway, including polysomnography.

A thorough craniofacial and intraoral examination should be completed to look for clues regarding the type (VP insufficiency vs. incompetency) and etiology of VPD. Features that should be assessed include:

- Symmetry of craniofacial structures
- Symmetry and length of the soft palate at rest and during elevation with phonation
- Presence, size, and location of any palatal fistulae
- Shape (single, wide, or bifid) and symmetry of the uvula
- Degree and consistency of velar elevation during phonation
- Signs of submucous cleft palate (bifid uvula, zona pellucida, posterior notch, v-shaped tenting of velum upon elevation)
- Length of palate relative to depth of oropharynx (palatopharyngeal disproportion)
- Evidence of past surgical management including scarring post-palatoplasty, scarring post-tonsillectomy, pharyngeal flap, and sphincter pharyngoplasty

This examination should also include a thorough assessment of the type and anatomic integrity of prior palatoplasty. When aerodynamically significant fistulae are identified (e.g., audible nasal emission is heard during speech), these should be repaired or obturated prior to functional assessment of the velopharynx. Position of the levator muscles is noted, when possible. Sagittal orientation of the levator muscle bundles suggests absent or incomplete intravelar veloplasty. Patients with complete or partial velar dehiscence should undergo palatal repair prior to reassessment of velopharyngeal function. Tonsillar size should also be noted, as tonsillar enlargement may increase the risk of postoperative sleep apnea. Conversely, patients who have undergone prior tonsillectomy may demonstrate scarring of the posterior tonsillar pillars that may impair the proper construction of a sphincter pharyngoplasty.

The intraoral exam in isolation should never determine the *need* for surgical treatment. The intraoral findings should be interpreted in the context of the perceptual speech evaluation, and subsequent instrumental and VP imaging findings in order to develop an appropriate treatment plan. The intraoral exam often reveals findings that may correlate with the perceptual speech findings; however, the nature and severity of VPD should be validated with direct VP imaging.

35.1.1 The Speech Evaluation

A perceptual speech evaluation should be conducted by a certified and licensed speech-language pathologist with training and experience in the assessment of patients with cleft palate, resonance disorders, articulation disorders (including knowledge of compensatory articulation errors), and voice disorders. The speech pathologist should also have advanced knowledge in anatomy and physiology of the vocal tract and upper airway. The speech-language pathologist should conduct a clinical evaluation as outlined below, making specific adjustments in the assessment protocol for patient age, developmental level, language, articulation proficiency, hearing status, and cooperation level:

1. Spontaneous speech sample (e.g., during conversation, picture description, or play)
2. Rote speech tasks (e.g., counting, days of the week)
3. Sentence repetition tasks
4. Word repetition and picture-naming tasks, including standardized articulation testing when errors are present
5. Stimulability probes, with and without nasal occlusion

As a patient produces this speech sample, the speech pathologist makes auditory-perceptual judgments of the patient's articulation, resonance, voice, intelligibility, and nasal emission (Henningsson et al. 2008). Equal-appearing interval scales are the most popular auditory-perceptual rating tool in the clinical setting; however, more recent studies have led to questions regarding their validity when compared to ratio-based methods (e.g., direct magnitude estimation and visual analog scaling) (Whitehill and Lee 2002). Regardless of which rating method is utilized, there is strong consensus that perceptual assessment of speech remains the gold standard for evaluation of speech, judgment of need for treatment, and judgment of outcome (Kuehn and Moller 2000).

Audio and/or video recording of the speech tasks should be completed whenever possible. Clinical archiving will allow for comparison before/after treatment, outcome assessment, and data collection for potential use in quality improvement projects or future research. Patients with a history of VPD should undergo a speech evaluation at least annually and more frequently if there are changing needs, such as when an articulation disorder is present. If the patient undergoes surgery for VPD, a postsurgical speech assessment should be conducted approximately 6 months after surgery and then annually thereafter until growth and development are complet or until all other therapy goals met and the patient is discharged.

After the perceptual speech evaluation and intraoral examination have been completed, the speech pathologist must determine if the patient demonstrates a clinically significant speech disorder and if the patient's speech profile is consistent with VPD. If VPD is suspected, the speech pathologist must then determine whether the patient has sufficiently accurate articulation to obtain valid imaging of the VP port and whether they are likely to provide adequate cooperation to complete instrumental testing and/or imaging.

35.1.2 Instrumental Assessment of VP Function for Speech

Instrumental evaluation of speech is a helpful adjunct to the perceptual speech evaluation. This traditionally includes some combination of acoustic assessment, aerodynamic testing, and VP imaging. Nasalance is an acoustic correlate of nasality and may be measured by a variety of commercially available tools (e.g., NasalView™, Nasometry™, Nasality Visualization System™). Nasometry™ is perhaps the most widely used acoustic assessment tool for measurement of nasality. The speech pathologist must first critically evaluate whether the patient is a good candidate for acoustic assessment based upon their age, cooperation, articulation skills, voice quality, and language or dialect, as well as understand the risk for confounding factors that can reduce the validity of the nasalance score (e.g., dysphonia, nasal turbulence, compensatory articulation errors, mixed resonance). If the patient is determined to be an appropriate candidate for acoustic assessment of nasality, the appropriate level and type of stimuli should be selected for use during testing. In general, hypernasal resonance correlates with elevated nasalance scores on oral speech stimuli, although perceived severity does not always demonstrate a linear relationship with nasalance scores (Fletcher 1976; Hardin et al. 1992). Nasalance scores are helpful as a comparison measure pre- and posttreatment, but should never be considered a substitute for perceptual speech evaluation when making a determination of the need for surgical intervention.

Aerodynamic (pressure-flow) testing is another useful tool for screening and evaluation of VP function during speech (e.g., PERCI-SARS™, Microtronics). It allows for direct measurement of intraoral pressure, nasal airflow, VP closure timing, and indirect measurement of VP orifice size (Warren and DuBois 1964; Warren et al. 1985). The degree of speech proficiency and cooperation required for pressure-flow testing is similar to that required for acoustic assessment. The word "hamper" is classically used as the speech stimulus for pressure-flow testing because of its /mp/ sound sequence which requires the velopharynx to open and close rapidly (Warren and DuBois 1964). An orifice of 10–20 mm^2 (or larger) during the /p/ sound of this stimulus has been shown to correlate highly with perceptual observations of hypernasality (Warren et al. 1985); however, even smaller VP gaps may be associated with hypernasality, especially when VP closure timing is also abnormal (Warren et al. 1993, 1994; Leeper et al. 1998).

After the perceptual speech evaluation, instrumental testing, and intraoral exam are completed, the team of providers confers to share findings and preliminary impressions. Together, the team determines if the patient requires VP imaging and the timing of the procedure (i.e., same visit, after additional speech therapy, after tonsillectomy, etc.). Regardless of the findings from the perceptual speech evaluation, intraoral exam, and instrumental speech testing, direct imaging of the VP port during speech is required for definitive diagnosis of VPD and the most accurate treatment planning. VP imaging also provides important information for assessing the upper airway to determine if additional management will be required prior to undertaking VP surgery (e.g., tonsillectomy, adenoidectomy, or other preoperative imaging), to minimize risks.

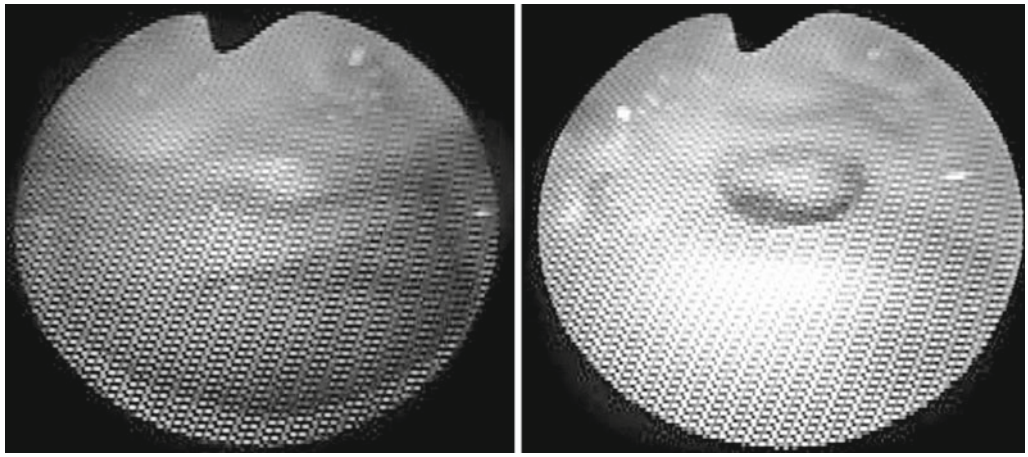

Fig. 35.1 Nasopharyngoscopic images of the velopharyngeal structures in a child with VPD. *Left*, velopharyngeal opening at rest. *Right*, persistent velopharyngeal gap during production of the /s/ sound (From 2nd Ed. Fig. 35.1)

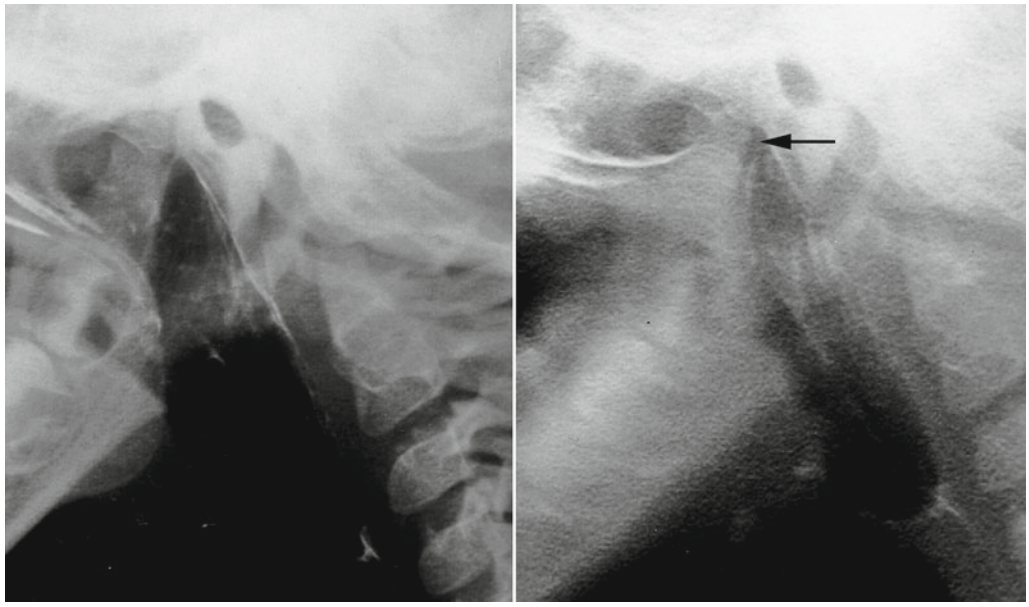

Fig. 35.2 Lateral view videofluoroscopic images of the velopharyngeal mechanism in a child with VPD. *Left*, velopharyngeal opening at rest. *Right*, persistent velopharyngeal gap (*arrow*) during production of the word "puppy" (From Murphy and Scambler (2005), with permission. From 2nd Ed. Fig. 35.2)

35.1.3 Imaging of the VP Mechanism During Speech

In all cases, the diagnosis of VPD should be confirmed by nasopharyngoscopy and/or multiview videofluoroscopy during speech (MVVF) (Figs. 35.1 and 35.2). Some centers have moved to the exclusive use of nasopharyngoscopy to avoid the radiation exposure associated with MVVF, while others still perform both procedures as part of a standard presurgical assessment. Nasopharyngoscopy is minimally invasive, can be performed in the outpatient setting, and is well tolerated by even the youngest of patients (typically ages 4 years and older). Nasopharyngoscopy during speech allows for the subjective assessment of VP gap size, VP closure consistency, shape of the VP gap, and VP closure pattern (Golding-Kushner et al. 1990). Other upper airway structures can also be examined (e.g., tonsils, adenoid, larynx). The primary benefit of nasophar-

yngoscopy is the ability to directly view the VP port in color, during connected speech. It is also more sensitive to detecting small or asymmetric VP gaps, VPD persisting after pharyngeal flap, and allows for advanced diagnosis of muscular abnormalities such as in cases of occult submucous cleft palate or anteriorly displaced levator veli palatini musculature.

To improve patient comfort with the procedure, topical anesthetic and/or nasal decongestant may be applied transnasally, although is not required. Application of a simple surgical lubricant to the external surface of the scope (avoiding the lens) also facilitates ease of transnasal insertion of the scope. Typically, nasopharyngoscopy is completed and/or interpreted by the speech pathologist and surgeon together and is videorecorded. High-resolution cameras (larger in diameter) are now available which offer improved image quality, magnification, field of view, and slow motion or frame-by-frame analysis; however, pediatric smaller-diameter nasopharyngoscopes tend to be better tolerated by younger patients. Once the scope is inserted transnasally through the middle or inferior meatus, the boundaries of the VP port should be visualized (i.e., anteriorly—soft palate, posteriorly—posterior pharyngeal wall/adenoid pad, laterally—lateral pharyngeal walls). The patient is then asked to engage in the production of a standard speech sample to assess VP closure *during* speech. The speech pathologist should select a speech sample focused on the patient's accurately articulated words and sentences. A range of stimuli (i.e., different phonemic contexts, range of utterance length, and contrasting error vs. accurate productions) are useful for gaining an understanding of the underlying pathology; however, the most representative image of maximum attempted VP closure will be obtained during the accurately produced (articulated) oral pressure consonants in connected speech. Images obtained during compensatory articulation error production (e.g., glottal stops, nasal fricatives) have been shown to be associated with a lesser degree of attempted VP closure than accurate speech production (Henningsson and Isberg 1991). Overall, the resulting images obtained during speech will allow the speech pathologist and surgeon to determine the most appropriate type of treatment, as well as location, type, and size specifications for potential surgical intervention to improve VP closure for speech. Examples of speech stimuli include:

- Pet a puppy.
- Buy baby a bib.
- Dad did it.
- Tell Ted to try.
- Go get a cookie for Kate.
- Fifty-five fish.
- Sissy sees the sky.
- She likes to shop for shoes.
- Chocolate chip cookies.

All of the above stimuli can be shortened if needed to adjust for patient speech proficiency or other articulation constraints. It is suggested that if the patient is only capable of accurate articulation at the single-word level, then multiple repetitions should be obtained in order to mimic the timing demands of natural VP closure (e.g., puppy-puppy-puppy). Additional stimuli including nasal consonants (e.g., My mom made muffins.) may be added to assess hyponasal speech or alternating oral-nasal contents (e.g., hamper hamper) to assess VP closure timing and coordination. The speech pathologist should carefully interpret the VP images in the context of the phonemic demands (i.e., oral vs. nasal, high pressure vs. low pressure) and accuracy of the sounds produced. Options for rating and measuring ratios of movement from the nasopharyngoscopy exam are outlined in Golding-Kushner et al. (1990).

In patients who demonstrate persistent or recurrent VPD after surgical management, careful intraoral and nasopharyngoscopic examination yields important diagnostic information. Pharyngeal flap integrity, width, port size, and position relative to the attempted level of velar closure should all be assessed. Occasionally, velar function may be impaired by the influence of a narrow flap that tethers the palate by virtue of being placed too low on the posterior pharyngeal wall. In such cases, the tethering flap should be surgically divided and the velopharynx reassessed. Similarly, the integrity, port size, and position of a previously constructed sphincter pharyngoplasty may yield important clues as to the nature of persistent VPD after surgical management.

MVVF is a valid alternative to nasopharyngoscopy for the assessment of VP function during speech, in

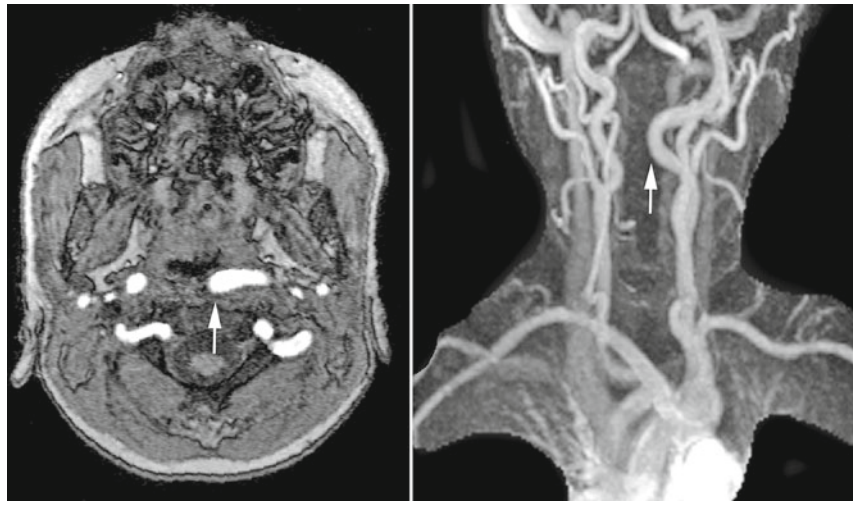

Fig. 35.3 Medial deviation of the internal carotid artery (*arrow*) at the level of the nasopharynx in a child with a chromosome 22q11.2 deletion (From Murphy and Scambler (2005), with permission. From 2nd Ed. Fig. 35.3)

most cases. MVVF typically requires a radiologist and speech pathologist to obtain and record the exam, and the images are later reviewed with the surgeon. Motion fluoroscopy records the movement of intraoral and velopharyngeal structures during speech from multiple angles (Skolnick 1970). Advantages of MVVF include more precise data regarding palatal length, pharyngeal depth, and relative size of upper airway anatomy, as well as information regarding the contribution of tongue movement to speech (or even to assisting VP closure). Image quality is best if liquid barium is instilled into the nasal cavity during the exam to coat the surfaces of the VP mechanism. At minimum, the lateral view is obtained, followed by frontal, base, and Towne's views. Measurement and rating procedures for interpretation of MVVF are also reviewed in Golding-Kushner et al. (1990).

Overall, preoperative imaging of the velopharynx is essential to the accurate assessment of VPD and therefore to surgical planning. Such imaging serves as an essential adjunct to visual assessment of levator muscle orientation, velar integrity and function, and adenoid and tonsillar morphology. Occasionally, velopharyngeal dysfunction is found to result from attempted closure of the velum against an irregularly shaped adenoid pad. Similarly, marked tonsillar enlargement may also interfere with velopharyngeal closure (MacKenzie-Stepner et al. 1987a; Shprintzen et al. 1987). In such cases, consideration should be given to adenoidectomy or tonsillectomy, respectively, prior to reassessment of velopharyngeal function. In all cases, the extent, pattern, symmetry, and attempted level of velopharyngeal closure should be noted. When gap size is small and the levators are sagittal in position, Furlow palatoplasty may be sufficient treatment. In patients with larger gaps and/or transverse levator orientation, conventional wisdom suggests that closure pattern dictates the choice of procedure. Posterior pharyngeal flaps are theoretically best suited for those patients that demonstrate good lateral pharyngeal wall motion and sagittal closure patterns, whereas sphincter pharyngoplasty is theoretically best suited for those with limited lateral wall motion and coronal closure patterns. It should be noted, however, that, despite the intuitive basis for such an approach, there is little clinical evidence of its soundness.

Magnetic resonance imaging should also be performed preoperatively in all patients with 22q11.2 deletion syndrome. One in five affected patients will demonstrate medial displacement of one or both internal carotid arteries (D'Antonio and Marsh 1987; MacKenzie-Stepner et al. 1987b; Mitnick et al. 1996; Ross et al. 1996), thus increasing the risk for arterial injury during posterior pharyngeal flap surgery or sphincter pharyngoplasty (Fig. 35.3). Arterial pulsations noted at the time of nasendoscopy may be transmitted through the surrounding

soft tissues, rendering endoscopy alone an unreliable means of assessing carotid anatomy. Although some have suggested that carotid artery displacement should contraindicate pharyngoplasty, others have demonstrated that surgery can be safely performed in this setting (Witt et al. 1998b; Tatum et al. 2002). In many cases, the displaced vessel(s) will lateralize with neck extension at the time of surgery. When a displaced carotid artery can still be palpated beneath the posterior pharyngeal mucosa after positioning, the design of a pharyngeal flap can be modified, skewing the flap away from the displaced vessel. In all such cases, preoperative delineation of arterial anatomy plays an important role in surgical planning and informed consent.

35.2 Furlow Double-Opposing Z-Palatoplasty (Fig. 35.4)

Despite its original description as a technique for primary repair of palatal clefts (Furlow 1978), the Furlow double-opposing Z-palatoplasty has increasingly become recognized as an effective procedure for the management of VPD in selected patients. Transposition of the posteriorly based flaps reorients the levator veli palatini muscle bundles and reconstructs the levator sling. It is for this reason, however, that the Furlow procedure should not be employed in patients who have already undergone complete intravelar veloplasty. The Z-plasty design provides for velar lengthening

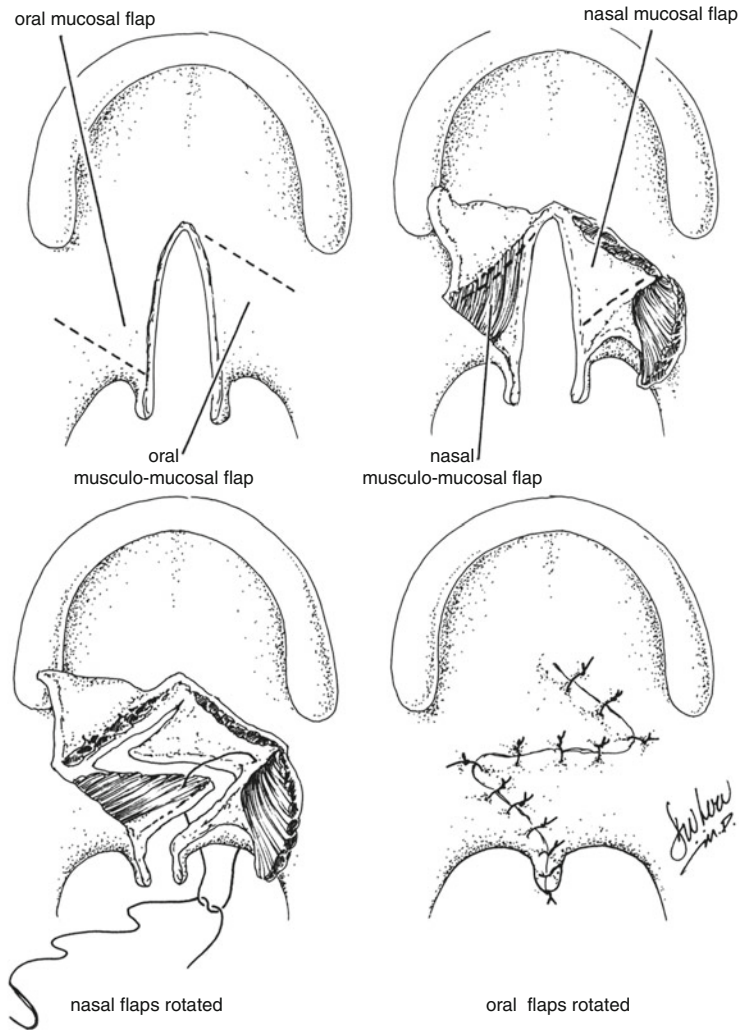

Fig. 35.4 Furlow double-opposing Z-palatoplasty (From Murphy and Scambler (2005), with permission. From 2nd Ed. Fig. 35.6)

while preventing the cicatricial shortening that may be seen with straight-line velar repair. These features render the Furlow repair ideally suited for patients with VPD associated with unrepaired submucosal clefts and those with small-gap VPD following palate repair without intravelar veloplasty. The procedure should not be performed in patients that have previously undergone reconstruction of the levator sling.

The anatomy of the Z-plasty design is determined by the underlying palatal anatomy. The posteriorly based flaps of both the oral and nasal sides comprise both muscle and mucosa, whereas the anteriorly based flaps comprise mucosa alone. The Z-plasty incisions extend from the tip of the hamulus to the posterior edge of the hard palate at the cleft margin on one side and from the base of the uvula at the cleft margin to the tip of the hamulus on the other. Dissection must be carried out laterally to the hamulus in each side in order to ensure that the levator fibers are completely divided from the posterior edge of the hard palate. This is necessary for proper reconstruction and retropositioning of the levator sling with transposition of the posteriorly based flaps.

Several authors have confirmed the efficacy of the Furlow double-opposing Z-palatoplasty in the management of VPD. Hudson et al. (1995) reported that 85 % of patients with VPD following primary palatoplasty achieved normal resonance after Furlow conversion. Chen et al. (1994) reported gap size to be the most important factor in determining the success of Furlow palatoplasty in the management of VPD, with the majority of patients with a preoperative gap of 5 mm or less achieving velopharyngeal competence postoperatively. D'Antonio et al. (2000) reported normal resonance after conversion to Furlow palatoplasty in 75 % of selected patients with post-palatoplasty VPD. All patients in their series demonstrated incomplete levator retropositioning, good velar motion, and a small velopharyngeal gap preoperatively.

As noted above, the Furlow procedure is ideally suited to those patients with nonsyndromic submucosal cleft palate and VPD. As in patients with repaired overt clefts, success rates appear to depend primarily on gap size. Seagle et al. (1999) reported that 83 % of patients with VPD associated with submucosal cleft palate demonstrated velopharyngeal competence after Furlow Z-plasty, with successful outcomes observed most frequently in patients with a gap size under 8 mm. Chen et al. (1996) noted that VP competence was achieved in 97 % of patients with submucosal clefts and gaps of 5 mm or less. Studies have confirmed that the Furlow Z-plasty produces a variable degree of palatal lengthening (D'Antonio et al. 2000). As noted above, however, the geometry of the Z-plasty incisions is determined by the underlying palatal anatomy. It stands to reason, therefore, that the success of the procedure is inevitably linked to gap size and to the extent of velar lengthening that can be achieved given the underlying palatal anatomy.

Bleeding, oronasal fistula formation, and upper airway obstruction have all been reported following Furlow palatoplasty. By virtue of the Z-plasty design, the technique achieves palatal lengthening at the expense of velar width, thereby creating a potential increase in tension at the suture line. Fistula formation can be minimized, however, by reducing tension through the liberal use of lateral relaxing incisions. Although mild upper airway obstruction has been documented following Furlow palatoplasty, such has been noted to resolve in nearly all patients within 3 months of surgery (Liao et al. 2003). Moreover, patients who have undergone Furlow palatoplasty demonstrate a significantly lower incidence and severity of upper airway obstruction 6 or more months postoperatively than do those patients who have undergone posterior pharyngeal flap surgery (Liao et al. 2004).

35.3 Posterior Pharyngeal Flap (Fig. 35.5)

In 1865, Passavant first described the surgical management of VPD by attachment of the velum to the posterior pharynx (Passavant 1865). Schoenborn described the use of an inferiorly-based posterior pharyngeal flap in 1875 and of a superiorly-based flap a decade later (Schoenborn 1875, 1886).

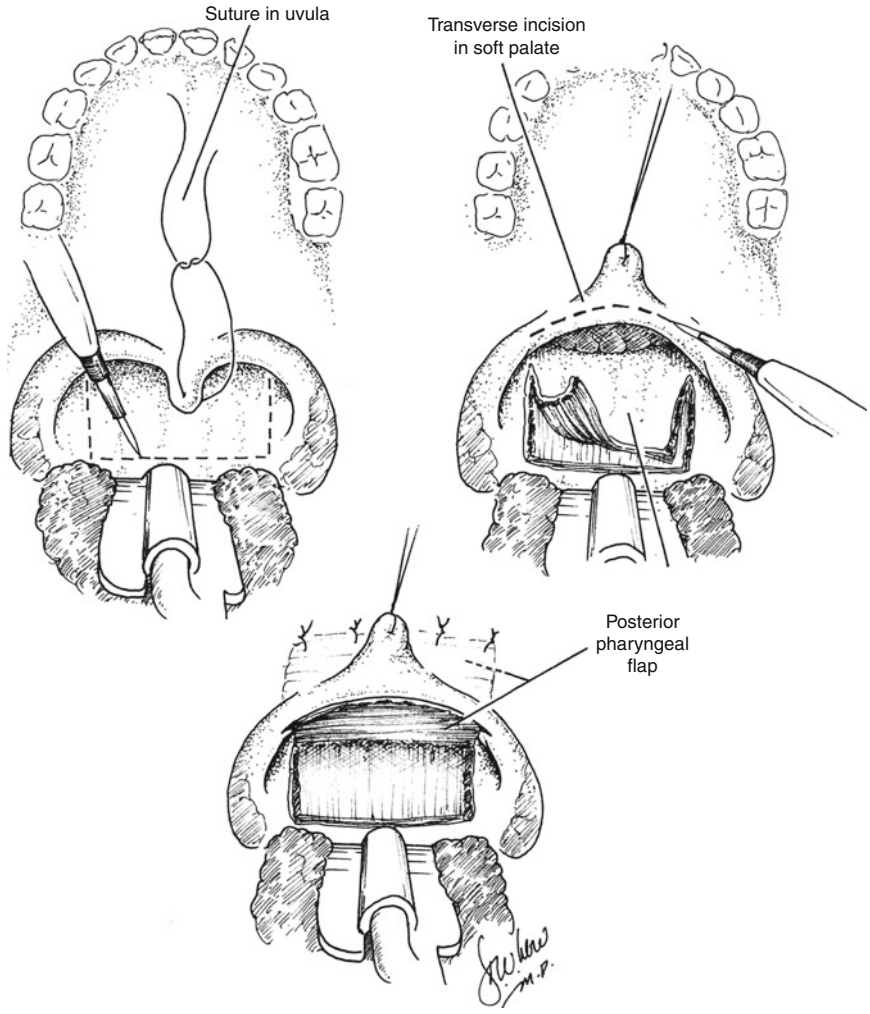

Fig. 35.5 Posterior pharyngeal flap (From Murphy and Scambler (2005), with permission. From 2nd Ed. Fig. 35.4)

Pharyngeal flap surgery was popularized in the United States by Padgett in the 1930s (Padgett 1930). For much of the twentieth century, the pharyngeal flap remained the workhorse of VPD surgery. With the addition of several modifications, the technique remains widely popular today.

The posterior pharyngeal flap acts as a static central obturator of the velopharynx, relying upon lateral pharyngeal wall motion to close the lateral ports during sound production. The procedure is best suited, therefore, for management of VPD in the patient with a sagittal or sphincteric closure pattern and a persistent central gap. Laterally based, inferiorly based, and superiorly based flaps have all been described, though the last remains the most widely used design today. Optimization of surgical outcome relies upon careful placement of the flap at the attempted level of velar contact with the posterior pharyngeal wall, as determined by preoperative imaging. As noted above, pharyngeal flaps that have been designed too low or that have migrated inferiorly due to cicatricial changes may tether the velum and restrict velopharyngeal closure.

Just as for flap position, the width of the pharyngeal flap should be tailored to each patient's needs as determined by preoperative assessment. Patients with large gaps and relatively poor lateral wall motion may require wider flaps in order to achieve velopharyngeal competence than those with smaller gaps and more robust lateral wall motion. Flap width is dependent not only on the

breadth of the flap design but also on the breadth of flap inset into the soft palate. Inset may be achieved by dividing the velum in the midline or through the use of a transverse incision across the posterior velum, the latter technique allowing for greater flexibility in establishing flap width and, conversely, lateral port dimension (Argamaso 1995). The tendency for flaps to narrow or "tube" over time may be minimized by the creation of mucosal lining flaps from the posterior velum or through the use of short, broad pharyngeal flaps. In all cases, flap design must be balanced against the risk of inducing postoperative upper airway obstruction, as wider flaps carry a higher risk of postoperative sleep apnea.

Careful surgical planning and individualization of flap design and inset are essential to achieving successful outcomes in pharyngeal flap surgery. Canady et al. (2003) reported their 10-year experience with pharyngeal flap surgery at the University of Iowa. Success, defined as normal or near normal resonance, was achieved in 78 % of patients. Argamaso (1995) reported elimination of hypernasality in 96 % of patients following pharyngeal flap surgery. Similarly, borderline or normal velopharyngeal function was observed postoperatively by Sullivan et al. in 97 % of patients (Sullivan et al. 2010). Studies have demonstrated the results of pharyngeal flap surgery to be durable, with stable results noted more than a decade after surgical treatment (Cable et al. 2004).

The reported complication rate for pharyngeal flap surgery ranges from 6.3 to 19.5 % (Valnicek et al. 1994; Fraulin et al. 1998; Hofer et al. 2002). The operation may be complicated by hemorrhage, flap dehiscence, hyponasality, persistent VPD, and nasal airway obstruction. Airway compromise has been reported to result in a small number of deaths after posterior pharyngeal flap surgery. Complications may be increased by association with limited operator experience, associated medical conditions, and concurrent surgical procedures. Of all complications, upper airway compromise is the most common. Due to postoperative edema, nearly all patients demonstrate some transient nasal airway obstruction following posterior pharyngeal flap surgery, and careful monitoring during the immediate postoperative period is therefore essential. In all but a small percentage of patients, nocturnal airway obstruction resolves within several months of surgery. Syndromic patients and those with a history of Pierre Robin sequence often present with associated functional or anatomic airway anomalies and may therefore be at greater risk for persistent upper airway obstruction after surgery (Wells et al. 1999; Abramson et al. 1997). The presence of tonsillar hypertrophy should alert the surgeon to an increased risk of sleep apnea following creation of a pharyngeal flap (Ysunza et al. 1993). In such cases, tonsillectomy should be performed prior to velopharyngeal imaging and subsequent pharyngeal flap surgery.

Persistent VPD following posterior pharyngeal flap surgery may result from inappropriate use of the procedure (i.e., poor lateral wall motion), from poor surgical design (i.e., a flap that is too low or too narrow), or from cicatricial alterations in flap location or dimension. In all cases, the treatment team must allow adequate time for resolution of edema and for scar maturation prior to consideration of surgical revision, usually a period of 12 months. Persistent VPD demands the same precision in diagnosis and management as does the initial condition. In all cases, nasendoscopic visualization is essential to assessment of the altered velopharyngeal valve, and surgical management should be tailored to the functional and anatomic needs so identified.

35.4 Sphincter Pharyngoplasty (Fig. 35.6)

In 1950, Hynes first described the treatment of VPD by transposition of myomucosal flaps containing the salpingopharyngeus muscles (Hynes 1950). He later modified the technique, constructing a sphincter containing the palatopharyngeus muscles (Hynes 1953). The procedure's success was attributed to narrowing of the velopharynx and augmentation of the posterior pharyngeal wall with bulky, often contractile flaps (Hynes 1967). The concept of dynamic sphincter pharyngoplasty was later championed by Orticochea (1968). Jackson later modified the Orticochea's procedure

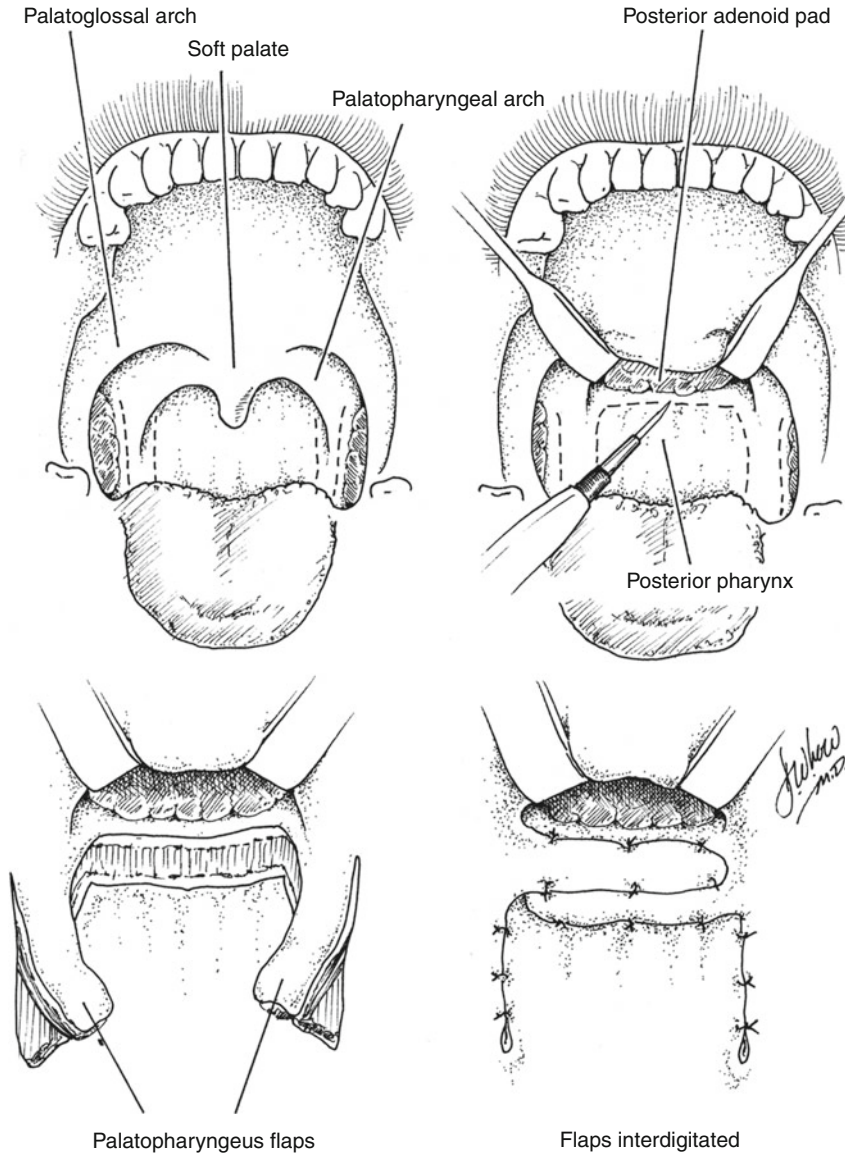

Fig. 35.6 Sphincter pharyngoplasty (From Murphy and Scambler (2005), with permission. From 2nd Ed. Fig. 35.5)

and described the technique of sphincter pharyngoplasty that is widely utilized today (Jackson and Silverton 1977).

In the construction of a sphincter pharyngoplasty, vertical incisions are made at the junction of each posterior tonsillar pillar with the adjacent tonsillar fossa. The longitudinally oriented palatopharyngeus muscles are dissected and included in the superiorly based flaps in order to maximize bulk and contractility. Vertical incisions are then made at the junction of the posterior pillars with the posterior pharyngeal wall, the inferior aspect of each flap is then divided, and the flap elevated. The flaps are then rotated medially and inset into a transverse incision in the posterior pharyngeal mucosa that joins the superior aspect of the medial incisions. Riski et al. (1984) have written that the level of inset is critical to success of the procedure and should correspond to the attempted level of velar contact as determined by preoperative imaging. Central port size may be controlled by altering the extent of flap overlap on the posterior pharyngeal wall and should be individualized

to the anatomic and functional needs of each patient. Several studies have shown that not all sphincters demonstrate contractility (Kawamoto 1995; Witt et al. 1998a). Port size and flap bulk may therefore play critical roles in achieving velopharyngeal competence.

Sphincter pharyngoplasty successfully manages VPD in the majority of carefully selected patients. Retrospective reviews by Shewmake et al. (1992) and by Mount and Marsh (2002) showed that velopharyngeal competence was achieved in 85 and 72 % of patients, respectively. Riski et al. (1992) reported that 78 % of 139 patients demonstrated resolution of hypernasality and normalization of pressure-flow measurements following sphincter pharyngoplasty. Poor results were associated with improper placement of the sphincter on the posterior pharyngeal wall. Reports by Witt et al. (1998c) and Losken et al. (2003) cite pharyngoplasty revision rates for persistent VPD of 16 and 12.8 %, respectively. In these series, poor outcomes were associated with dehiscence, syndromic diagnoses, and greater preoperative nasalence scores.

There is little consensus as to the superiority of sphincter pharyngoplasty versus posterior pharyngeal flap surgery. Indeed, procedure selection should be based primarily on velopharyngeal anatomy and function. Nevertheless, Ysunza et al. found no significant difference in speech outcome following randomization of 50 patients with post-palatoplasty VPD to either sphincter pharyngoplasty or posterior pharyngeal flap (Ysunza et al. 2002). Abyholm et al. similarly noted no significant difference in velopharyngeal function 12 months following sphincter pharyngoplasty or posterior pharyngeal flap surgery in a randomized multicenter trial (Abyholm et al. 2005). Complications following sphincter pharyngoplasty are similar to those seen after posterior pharyngeal flap surgery: bleeding, dehiscence, hyponasality, persistent VPD, and nocturnal upper airway obstruction. In Abyholm's report, polysomnographic evidence of obstructive sleep apnea was rare 1 year following either procedure (Abyholm et al. 2005). Nevertheless, some have noted that fragmentation of sleep architecture may be observed following pharyngoplasty even in the absence of detectable obstructive apnea (Saint Raymond et al. 2004).

35.5 Timing of Surgery

Several studies have shown that the age at pharyngeal flap surgery may impact speech outcome (Moll et al. 1963; Meek et al. 2003; Riski 1979); however, many other published studies have found little to no direct relationship between age and outcome (Van DeMark and Hardin 1985; Seyfer et al. 1988; Becker et al. 2004; Hall et al. 1991). Most studies report a link between outcome and preoperative structural or functional indices such as severity of hypernasality and the presence of compensatory articulation errors. Skoog (1965) was the first to report that that pharyngeal flap surgery resulted in better speech outcomes if performed in children before 10 years of age; however, Whitaker et al. (1972) later reported that speech results in children aged 13–16 were not significantly different than that of the younger age groups. Riski (1979) conducted a thorough study of children pre- and post-pharyngeal flap surgery and found that after pharyngeal flap, there was an acceleration in the acquisition of acceptable sound production in the year immediately following surgery. The latter data suggested that children who had flaps prior to 6 years of age made faster gains in articulation skills and resonance than children who had VP surgery after 6 years of age. His findings align with those of Meek et al. (2003), who also reported that speech development is enhanced by early surgery. Riski hypothesized that early surgery results in less severe and less permanent maladaptations in speech physiology and may clinicians continue to adhere their practices to this theory. On the contrary, Becker et al. (2004) found no relationship between age at VP surgery and amount of speech therapy needed to achieve "normalization" of the speech impairments secondary to VPD after surgery. Other studies have also found little to no support for the theory that earlier VP surgery is more effective, with the exception that there is a trend for poorer articulation outcomes in children treated after 10 years of age (Van DeMark and Hammerquist 1978; Seyfer et al. 1988).

Van Demark and Hardin (1985) challenged the findings of Riski (1979) as they did not find a trend for better articulation with earlier secondary mgmt. Their data did not support the hypothesis

that early VP management (before age 4) results in higher articulation proficiency. Overall, they suggested that age at pharyngeal flap is not extremely critical, but differences in success may be noted at age extremes. Based on the available evidence, it appears prudent to consider surgical intervention as soon as there is consensus that VPD is present, negatively impacting speech, and all team members (including the patient-family) are in agreement that surgical treatment is appropriate and offers a significant likelihood of improvement of the patient's speech.

Few studies have addressed the surgical outcomes of adults undergoing VP surgery. Hall et al. (1991) reported outcomes for 20 adults who underwent pharyngeal flap surgery and found that 75 % demonstrated "normal" resonance post-surgery, however, less experienced improved speech intelligibility. This was likely due to the presence of compensatory articulation errors in some of the adults, which persisted in most cases. These errors negatively impact speech intelligibility and may also lead to reduced VP closure which can subsequently affect resonance and nasal emission as well. Compensatory articulation may prevent or restrict VP motion and may result in persisting VPD (Henningsson and Isberg 1991). For adults, careful presurgical assessment of articulation is required in order to provide the patient with the most accurate prognosis and anticipated outcome. The Hall et al. study also highlighted the importance of reporting hyponasality as an outcome as well; as in many reports, it is simply omitted even though it should be considered a "poorer outcome." Many past studies include hyponasality as a successful outcome since the VPD was improved; however, there are multiple well-known complications that tend to co-occur with hyponasality including mouth breathing, snoring, decreased dental hygiene, difficulties with nasal secretion management, and other negative impacts on quality of life.

There continues to be some debate as to whether to recommend VP surgery prior to correction of maladaptive articulation patterns or whether to wait until after these articulation errors have been remediated. The rationale for earlier surgical intervention is supported by past research, presenting the argument that patients benefit from having adequate anatomy and physiology to achieve VP closure *prior* to implementing behavioral speech therapy. The counterargument proposes that the diagnosis of VPD may not be clear, or the true severity of extent of VPD may not be evident, until *after* compensatory misarticulations have been eliminated. These errors have been shown to negatively impact VP closure physiology and will not respond solely to surgical intervention. Regardless of surgery, compensatory articulation errors will require behavioral speech therapy for correction, although the duration and intensity of treatment may vary depend on surgical timing. The plasticity of the VP mechanism is likely greatest in the first few years of life as speech patterns are developing; however, with appropriate cueing and feedback (such as that provided by nasopharyngoscopy), later changes in VP function for articulation are still possible even into adulthood (Witzel et al. 1988, 1989; Brunner et al. 2005). Ysunza et al. (1992) also found that VP movement is significantly improved after correction of compensatory articulation errors, as well as reduction in the size of the VP gap, prior to undertaking VP surgery.

35.6 Management of Persistent VPD

Although surgical management of VPD successfully eliminates hypernasality in most patients, approximately 15–20 % of patients will require revision for persistent symptoms of VPD. Persistent hypernasality after posterior pharyngeal flap surgery may rarely be the result of complete or partial flap dehiscence or may occur when lateral pharyngeal wall motion is insufficient to close the lateral ports during speech. The lateral circumstance may result from improper flap design, cicatricial flap narrowing, or poor patient selection (i.e., poor lateral pharyngeal wall motion) (Fig. 35.7). In some cases, a scarred flap that is located too low on the posterior pharyngeal wall may restrict palatal elevation, resulting in persistent VPD. Sphincter pharyngoplasty may also fail as a result of flap dehiscence, poor design, or poor patient selection.

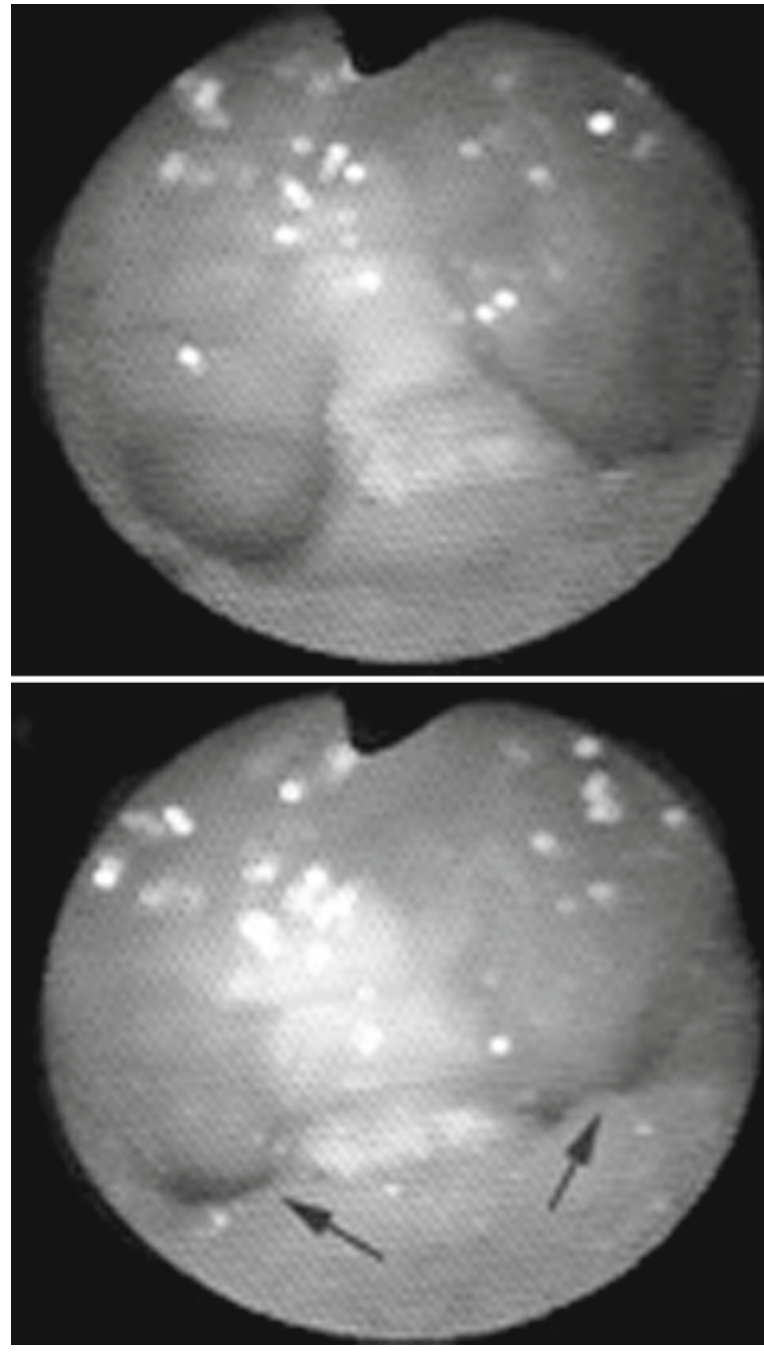

Fig. 35.7 Persistent VPD after posterior pharyngeal flap surgery. *Top*, narrow pharyngeal flap at rest. *Bottom*, persistent pharyngeal flap (*arrows*) during oral speech production (From 2nd Ed. Fig. 35.7)

Surgical revision for persistent VPD should be avoided until scar maturation is complete, as gradual improvement in hypernasality may be observed for up to 12 months postoperatively. Patients that continue to demonstrate persistent hypernasality should undergo diagnostic imaging of the velopharyngeal port by nasendoscopy and/or videofluoroscopy. Patients under consideration for revisional surgery should be carefully screened for evidence of upper airway obstruction, and revision should only be performed in patients with a stable airway. Management of persistent VPD should

then be tailored to each patient's overall medical condition, airway status, and specific anatomic and functional inadequacies of the velopharyngeal valve. Posterior pharyngeal flaps that are found to be too narrow can be augmented by "patch" flaps or divided and replaced by wider flaps or sphincter pharyngoplasty. Flaps that are found to be too low and that tether the velum may be repositioned higher on the posterior pharyngeal wall by V-Y advancement. Incomplete central port closure after sphincter pharyngoplasty may be managed by port tightening or by conversion to a posterior pharyngeal flap. Although elevation of a pharyngeal flap from a scarred posterior pharynx has the theoretical disadvantage of poor blood supply to the flap, the procedure has been demonstrated to be reliable when performed 6–12 months following the primary procedure (Barone et al. 1994).

Just as for primary surgical management of VPD, successful secondary treatment requires careful preoperative evaluation and individualized management. Barone et al. (1994) have reported on 18 patients who demonstrated incomplete lateral port closure after posterior pharyngeal flap surgery that was treated by elevation of a new, wider flap from the scarred posterior pharynx. Unilateral port insufficiency in another three patients was managed by inset of a small "patch" flap on the affected side. Postoperatively, 18 patients achieved normal resonance, two remained hypernasal, and one became hyponasal. Witt et al. (1998c) have reviewed the results of salvage surgery for 13 failed pharyngeal flaps, noting that eight were successfully revised with a single secondary procedure. The remaining five patients achieved velopharyngeal competence after a second revisional procedure, but all developed hyponasal speech. In the same report, 17 of 20 failed sphincter pharyngoplasties were successfully salvaged with a single surgical revision.

35.7 Summary

The operative timing and approach to the correction of velopharyngeal dysfunction should represent a balance between each patient's specific anatomic and functional demands, airway stability, and overall health status. Surgeons, speech pathologists, and researchers should consider multiple factors beyond resonance, such as speech acceptability, intelligibility, naturalness, patient and family satisfaction, as well as the presence of upper airway obstruction and other complications, when judging and reporting surgical outcome. Speech impairment associated with velopharyngeal dysfunction may be a significant source of stigmatization in affected individuals, making its diagnosis and treatment an imperative. Proper diagnosis and management of velopharyngeal dysfunction demands a multidisciplinary team approach, and surgical planning should be individualized in order to optimize outcomes.

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Velopharyngeal Dysfunction Management Algorithms

36

Jeffrey L. Marsh

As with most aspects of cleft care, management of velopharyngeal dysfunction (VPD) requires choices among different treatment modalities, technical variations within a specific modality, and timing of intervention. I have consolidated my experience, as a member of an interdisciplinary team managing VPD over the past 34 years, into a series of algorithms (Marsh Figs. 36.1, 36.2, 36.3, and 36.4). These algorithms not only guide my personal patient care decision making but have proven useful in teaching others about VPD management (Marsh 2004). This chapter presents and discusses these algorithms.

Individuals with possible VPD present to individual members of the cleft palate team or to the team as a whole for diagnosis and treatment. Historically, the large majority of such individuals had a repaired cleft palate and a small minority, without overt cleft palate, developed VPD following adenoidectomy. While post-palatoplasty patients with impaired speech still comprise the largest group evaluated for VPD (Kummer 2007; Peterson-Falzone et al. 2010), individuals with velocardiofacial syndrome, neurological impairment, and postobstructive sleep apnea surgery (uvulopalatoplasty) comprise a steadily increasing percentage. Whereas identification of the etiology of the VPD is critical to assigning differential management based on differential diagnosis (Marsh 1991; Marsh and O'Daniel 1992), the evaluation of velopharyngeal function is the same regardless of etiology (Witt et al. 1999). I shall first discuss the evaluatory process and then specific managements.

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36.1 Evaluation of Velopharyngeal Function

Speech results from modulation of the pressurized air stream that emerges from the lungs during expiration. A number of structures between, and including, the glottis and the lips (articulators) modify that air stream to produce specific sounds. One of those modifiers is the velopharynx, which normally is a dynamic sphincter. A sphincter is an anatomic structure that can open or close to varying degrees and speeds depending on the physiological necessity. The tissues that comprise the velopharyngeal sphincter are the velum (soft palate), the right and left lateral pharyngeal walls, and the posterior pharyngeal wall. The space enclosed by and modified by the velopharyngeal sphincter is the velopharynx. The velopharynx is the connection between the nasopharynx and the oropharynx. Normally at

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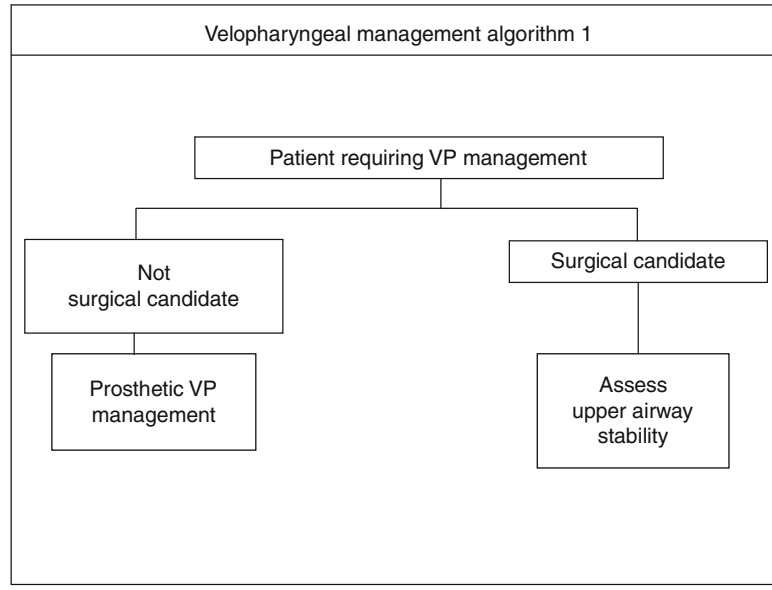

Fig. 36.1 Velopharyngeal management algorithm 1

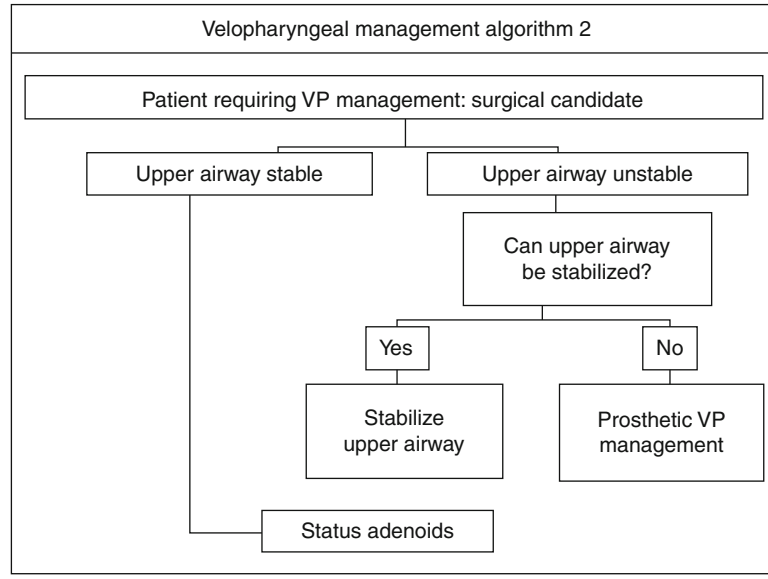

Fig. 36.2 Velopharyngeal management algorithm 2

rest, it is a wide opening through which air flows easily and secretions drain; when contracted, it closes the nasopharynx from the oropharynx, preventing passage of gases, fluids, and solids between the two cavities. Dysfunction of the velopharyngeal sphincter impairs that separation of the nasal and oral cavities for both deglutition and speech to varying degrees. Outcome assessment of VPD management requires consideration of velopharyngeal function for all of the following physiological activities: speech, breathing, swallowing, handling nasal secretions, and sleep.

There is a lack of consensus on the preferred term to describe such dysfunction: *velopharyngeal incompetency*, *velopharyngeal insufficiency*, and *velopharyngeal inadequacy* have all been abbreviated as *VPI*. I prefer the term *velopharyngeal dysfunction*, abbreviated as *VPD*, since it connotes physiological impairment without attempting to denote etiology (Loney and Bloem

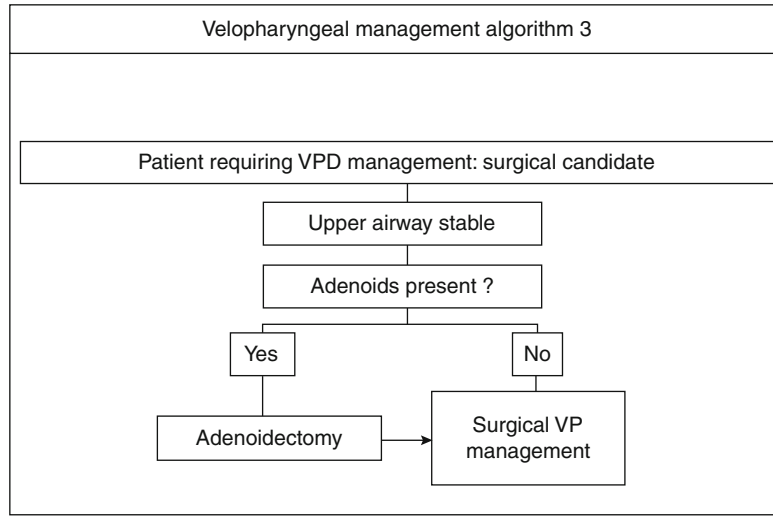

Fig. 36.3 Velopharyngeal management algorithm 3

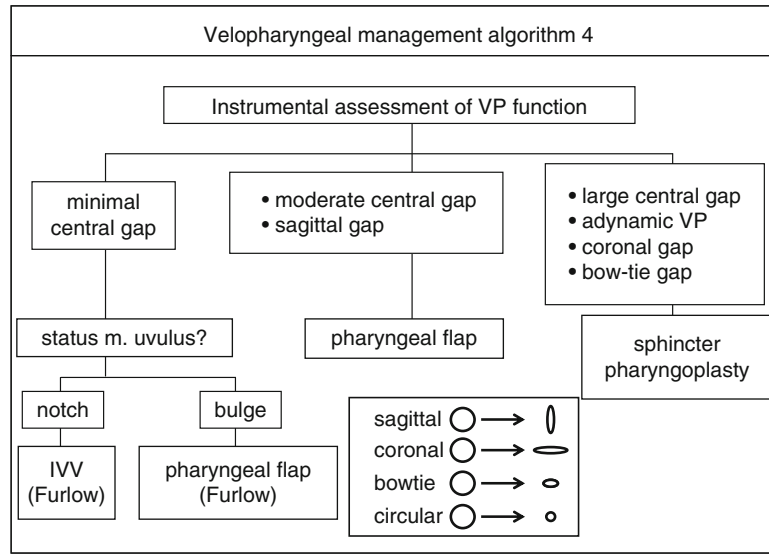

Fig. 36.4 Velopharyngeal management algorithm 4

1987; Folkins 1988; Smith and Kuehn 2007). Regardless of the terminology, a specific set of physical findings suggest VPD. These include excessive nasal resonance during speech (hypernasality), inappropriate nasal airflow noise (nasal turbulence or rustling), and abnormal facial movements (facial grimacing). An individual with one or any combination of these findings will be referred for evaluation of possible VPD.

The first task for the VPD team is to determine whether the patient actually has VPD. This determination has traditionally been made solely on the basis of a perceptual speech/language evaluation by a speech/language pathologist with specific training and experience in velopharyngeal function (Jones 1991). This evaluation includes both spontaneous speech and a provocative speech sample, structured to specifically challenge the velopharyngeal mechanism with increasingly more complex tasks (Philips 1980; Kummer 2007; Peterson-Falzone et al. 2010). The primary activity of this perceptual assessment is auditory, that is, listening for the production of specific phonemes. However, observing the face for grimacing, watching for fogging of a mirror below the nares, and feeling for airflow through the

nares with attempted pronunciation of phonemes that require velopharyngeal function are other perceptual assessments that assist the screening evaluation. Some professionals now argue that instrumental assessments of oral-nasal airflow should be part of this determination process. However, we have not yet incorporated nasalance measurement into our initial assessment.

The auditory perceptual screening of velopharyngeal function sorts the patient into one of three groups: normal VP function, minimal VPD without impairment of communication or psychosocial stigmatization, or sufficient VPD to interfere with speech comprehension by others and/or stigmatize the speaker. If the assessed individual screens normally, no further evaluation of the velopharynx is indicated until the return for routine follow-up or some concern regarding speech arises. If the possible dysfunction is minimal, meaning that the affected individual and his or her parents, in the case of a child, were not aware of a speech problem and the cleft team, in concert, feels that the dysfunction is unlikely to cause impairment of communication or have psychosocial consequences, then the affected individual will have a repeat auditory perceptual speech evaluation at an interval shorter than the usual follow-up and the individual/parents will be informed of the concern regarding velopharyngeal function as well as of the signs and symptoms of increasing dysfunction so that they can contact the cleft team should they occur. If, on the other hand, it is the perception of the sophisticated observer that there is a possibility of morbid velopharyngeal dysfunction, further evaluation is considered.

If one accepts the principle that treatment should be diagnosis specific, and I do, then effective management of velopharyngeal dysfunction requires both recognition of the condition and definition of the responsible pathoetiology (Jones 1991). While auditory perceptual evaluation can separate normal from abnormal velopharyngeal function, it cannot identify the etiology of the dysfunction. Instrumental velopharyngeal evaluations, in contrast, can be etiology specific (Yules and Chase 1968; Shprintzen and Golding-Kushner 1989; Hirschberg and Van Demark 1997). Instrumental evaluations can be divided into those that visualize the velopharynx and those that indirectly assess its function (Albery et al. 1982; Dalston and Warren 1986; Dalston et al. 1991; D'Antonio et al. 1988; Hardin et al. 1992). Clinically useful visualization utilizes still (lateral skull x-ray) or real-time x-ray (speech videofluoroscopy). Documenting the effect of velopharyngeal sphincter function on airflow air pressure or sound or light transmission across the velopharyngeal port produces indirect data about velopharyngeal function. A multidisciplinary consensus regarding reporting of visualization studies provides a useful means for standardized interpretation of such studies (Golding-Kushner et al. 1990). For the past 20 years, the cleft teams that I have been associated with have used both nasendoscopic and fluoroscopic imaging of the velopharynx during standardized speech tasks to evaluate velopharyngeal function (Sinclair et al. 1982; D'Antonio et al. 1988; Shprintzen and Golding-Kushner 1989; Stringer and Witzel 1989). These evaluations are audio-video recorded, previously on magnetic tape and now digitally, in order:

1. To permit review by other members of the cleft team who were not present during the actual examination
2. To permit comparison of serial assessments of the same individual
3. To permit comparative assessment of the effects of therapeutic intervention in the same individual
4. To facilitate cross-sectional comparison of various interventions

These tapes are then reviewed by a subset of the cleft palate team, which consists of representatives from the disciplines of otolaryngology, plastic surgery, and speech/language pathology (D'Antonio et al. 1989; Sell and Ma 1996). A consensus opinion regarding interpretation of the images and preferred method of management is then arrived at by group discussion. The conclusions are communicated to the patient/family for reaction and to appropriate primary and secondary care providers, such as the patient's speech therapist and pediatrician/physician. When velopharyngeal management, either

prosthetic or surgical, is recommended and performed, the velopharyngeal function is reevaluated 3 months after the intervention with auditory perceptual and instrumental evaluations mirroring those performed preoperatively.

36.2 Differential Diagnosis of Velopharyngeal Dysfunction

For the purposes of differential management based upon differential diagnosis, velopharyngeal dysfunction can be divided into four broad etiologic categories:

1. Anatomic deficiency
2. Myoneural deficiency
3. Anatomic and myoneural deficiency
4. Neither anatomic nor myoneural deficiency

Each category has several specific causes for that deficiency. *Anatomic deficiency* includes those static tissue problems that prevent adequate velopharyngeal sphincteric closure, such as an unrepaired cleft palate, a palatal fistula, a short velum, or a deep velopharynx. *Myoneural deficiency* implies that there is adequate palatal and velopharyngeal structural anatomy but one or more components of the velopharyngeal port have inadequate or absent function, such as may occur with head trauma or degenerative neurologic disorders. Combined *anatomic and myoneural deficiency* may occur with repaired cleft palate, unoperated submucous cleft palate, or post-ablative surgery and/or radiotherapy for oronasopharyngeal malignancy. Rarely an individual presents who has *intact velopharyngeal anatomic structures and is myoneurally intact* but whose speech nonetheless exhibits signs and symptoms of velopharyngeal dysfunction due to either learned behavior, such as having a parent with cleft VPD and living in a rural area, or psychiatric disorder.

The initial goal of the instrumental evaluation of the presumably impaired velopharynx is to determine which of the four categories the patient belongs to since that broadly determines the type of therapeutic intervention(s) to be considered (see below). The combination of visual direct and indirect (mirror, endoscope) intraoral inspection and indirect velopharyngeal functional inspection (nasendoscopy, multiview fluoroscopy) permits definition of the structural and functional impairments contributing to the VPD. Once the category has been defined, further general health history, physical examination, and testing determine the specific etiology for that patient's VPD.

36.3 Differential Management of Velopharyngeal Dysfunction

The four etiologic categories enumerated above broadly determine the differential management of velopharyngeal dysfunction. *Anatomic deficiency* is usually managed by restoration of the deficiency by either surgical or prosthetic means (Marsh and Wray 1980) (Fig. 36.1). *Myoneural deficiency* is usually managed prosthetically until the neurologic status has stabilized and the patient becomes a candidate for velopharyngeal surgery, which may or may not occur (Riski and Gordon 1979). *Combined anatomic and myoneural deficiency* is usually managed by restoration of the deficiency by either surgical or prosthetic means. Speech therapy, often pre- and usually post-intervention, is an essential element of management of these three categories in addition to surgical or prosthetic intervention. For *learned or behavioral VPD*, speech therapy with or without behavioral management is the treatment of choice; there is no role for surgery in such cases (Heller et al. 1974).

Prosthetic management for VPD is indicated when:

1. The patient is unsuitable for surgical velopharyngeal management due to other major medical problems, such as inoperable congenital cardiac disease or severe pulmonary insufficiency (if and when the other medical problem becomes resolved or stabilized enough to minimize the anesthetic/surgical risk, conversion to surgical velopharyngeal management is considered).
2. The parents/legal guardians refuse to permit surgery because of religious or emotional considerations.

3. The patient has a complex speech/language problem, and it is not clear that velopharyngeal management will significantly improve speech intelligibility (if the trial of a speech prosthesis demonstrates significant improvement in intelligibility, then conversion to surgical velopharyngeal management is considered).
4. The patient has a neurological condition that includes VPD, and it is not clear whether the neurological deficit has stabilized and/or that velopharyngeal management will significantly improve speech intelligibility (if the trial of a speech prosthesis demonstrates significant improvement in intelligibility and the neurological status has stabilized and the patient is not a significant anesthetic risk due to the neurological pathology, then conversion to surgical velopharyngeal management is considered).
5. The patient has an unstable upper airway that cannot be stabilized (Fig. 36.2) and VPD (this was the major indication for prosthetic velopharyngeal management at our cleft center prior to the introduction of mandibular distraction osteogenesis for stabilization of the upper airway in individuals with mandibular microretrognathia).

When a prosthesis is used to manage VPD, the type of prosthesis depends on the amount and quality of palatal tissue and the findings on nasendoscopic velopharyngeal functional assessment (Turner and Williams 1991; Witt et al. 1995b):

1. When there is adequate velar tissue (a long, unscarred velum), a palatal lift is prescribed.
2. When the velar tissue is deficient (a short, scarred velum with a deep pharynx), a velopharyngeal obturator is prescribed.
3. When the velum is long but the pharynx is deep or a palatal lift fails to achieve complete velopharyngeal closure, a combined lift and obturator, "liftorator," is prescribed.

Surgical VP management is recommended for all other individuals with VPD who do not meet one of the criterion listed above for prosthetic management. Several types of surgical alteration of components of the velopharynx are possible:

1. Velar lengthening (palatal pushback, Furlow double-opposing Z-veloplasty)
2. Intravelar muscular reconstruction
3. Pharyngoplasties (pharyngeal flap, sphincter pharyngoplasty)
4. Posterior pharyngeal wall augmentation

Each type of surgical alteration of velopharyngeal structures has its advocates. Although some comparisons of two or more methods of VPD management have been reported, they fail to meet rigorous scientific criteria for discriminating among therapeutic options (Pensler and Reich 1991; de Serres et al. 1999; Sloan 2000; Seagle et al. 2002; Ysunza et al. 2002).

Both types of velar operations and posterior wall augmentation procedures are fundamentally different from, and theoretically more physiologic than, the pharyngoplasties in that they do not create a permanent subtotal obstruction of the velopharyngeal port. Nasal physiology is less likely to be impaired with velar and posterior pharyngeal wall procedures than with pharyngoplasties. The challenge for the velopharyngeal surgeon is to provide the patient with the means to achieve velopharyngeal closure without obstructing the nasal airway for breathing, secretion control, and passage of air into the nose during speech for those sounds that require nasal resonance. For this reason, when either a pharyngeal flap or a sphincter pharyngoplasty is elected, an adenoidectomy is performed at least 3 months prior to the velopharyngeal procedure so that hypertrophic lymphoid tissue cannot obstruct the surgically diminished velopharyngeal port(s) and to facilitate the performance of the pharyngoplasty. [Previously, we also had tonsillectomy performed synchronous with the adenoidectomy. However, the tonsillectomy often resulted in fusion of the anterior and posterior tonsillar pillars into a single rigid scar that precluded an effective sphincter pharyngoplasty. Therefore, since 2008, we have only had adenoidectomy performed prior to definitive surgical velopharyngeal management.]

So how does the surgeon choose among these options for intervention? In collaboration with my colleagues on the multidisciplinary velopharyngeal

team, I choose among the therapeutic options based upon the anatomy and function of the velopharynx as documented by nasendoscopic and multiview fluoroscopic visualization of the velopharynx (Croft et al. 1981), performing both spontaneous speech and standardized speech tasks. The selection process follows an algorithm that has been in use, with minor modification, over the past 14 years (Marsh Fig. 36.4). My rationale for pairing of the residual velopharyngeal gap on attempted complete velopharyngeal closure with specific interventions follows:

Small midline gap – Management of a small midline gap on attempted complete VP closure depends on the status of the intravelar musculature. If the patient has not had an intravelar veloplasty or did not have a "radical" (method of either Sommerlad or Cutting) intravelar veloplasty, a radical IVV is recommended. The patient/family is informed that there is about an 80 % change of correction of the VPD with such an intervention, in my experience, with no expected airway morbidity. They are also informed that either a narrow, lined, superiorly based pharyngeal flap or a sphincter pharyngoplasty could be performed with an almost 100 % correction of VPD but with some risk of impairment of the upper airway, primarily intranasal retention of nasal secretions, and to a lesser degree nasal airway obstruction or the rare case of obstructive sleep apnea. If the radical IVV fails to sufficiently correct the VPD, then a pharyngeal flap or sphincter pharyngoplasty is offered with the caveats listed above. Some families prefer to avoid nasal obstruction even if there is a 20 % chance of needing a second operation while others prefer more certainty of resolution of the VPD regardless of airway compromise. [Rearrangement of the velar soft tissues including the muscle via the Furlow double-opposing Z-plasty is an alternative to radical IVV for the patient with a small midline gap on attempted complete VP closure. I am unaware of a prospective comparison of the efficacy and morbidity of these two approaches for equivalent VPD situations. The choice then between the two operations currently is surgeon's preference. I have no personal experience with the Furlow, but several of my associates, current and past, use it for this indication as well as several cleft surgeons whose technical skill and reporting honesty I respect.] Between 2003 and 2011, I performed four intravelar veloplasties to manage symptomatic VPD. Of these, two subsequently underwent sphincter pharyngoplasty for persistent or recurrent VPD: one 1 year later and one 4 years later. None of the IVV patients had sleep apnea or symptomatic nasal secretion retention following the IVV.

Sagittal gap with active to moderately active lateral pharyngeal wall motion – I reserve the pharyngeal flap operation for VPD due to a sagittal gap with active to moderately active lateral pharyngeal wall motion. Because of this restriction, I only perform narrow or moderately wide, superiorly based, lined pharyngeal flaps. [I stopped performing wide obstructive pharyngeal flaps in 1989, when sphincter pharyngoplasty entered our therapeutic repertoire, due to the consistent morbidity of wide obstructive pharyngeal flaps with respect to the upper airway: intranasal retention of nasal secretions, obstruction of the nasal airway with obligatory mouth breathing, hyponasal resonance, and obstructive sleep apnea as well as death (Kravath et al. 1980).] A review of 71 patients, who underwent a pharyngeal flap operation at our cleft center between 1982 and 2000 and had adequate preoperative and 3-month postoperative perceptual and instrumental velopharyngeal speech function evaluations and a 12-month postoperative perceptual velopharyngeal speech evaluation, was conducted in 2001. This study documented satisfactory nasal resonance in 74 % of patients so selected and treated. Of the remainder, surgical tightening of the one or both lateral ports with incomplete closure, as documented on post-pharyngeal flap nasendoscopy, increased the resolution of symptomatic VPD to 92 %. Postoperative complications included obstructive sleep apnea in five patients, symptomatic nasal secretions in seven patients, and obligate mouth breathing in five patients. Denasality (hyponasal nasal resonance) was noted in 21 % of patients at 3 months postoperatively but had diminished to only 6 % at 12 months postoperatively. Of the

patients receiving a pharyngeal flap, 3/4 had cleft palate+/−cleft lip, 1/4 were non-cleft VPD, and 1/5 of the patients were syndromic. Postoperative nasal resonance was not affected by the presence or absence of cleft palate ($p=0.7$) or the presence or absence of a syndrome ($p=0.2$) (Sabry and Marsh 2003). Between 2003 and 2011, I performed 12 pharyngeal flaps. Of these, 100 % had resolution of symptomatic VPD following the operation. None of these had sleep apnea or symptomatic nasal secretion retention.

Hypodynamic or adynamic velopharyngeal sphincter – I utilize the sphincter pharyngoplasty (Jackson modification of the Hynes procedure with "crossed arms" overlapping posterior tonsillar pillar myomucosal flaps) for all cases of VPD secondary to a hypodynamic or adynamic velopharyngeal sphincter (Witt et al. 1995a, b). Preoperative specification of the locus for insertion of the myomucosal flaps on the posterior pharyngeal wall is based on the level of maximum attempted velopharyngeal closure, as documented on the lateral fluoroscopic VP evaluation (Riski et al. 1984). A review of our sphincter pharyngoplasties in 2000 (Mount and Marsh 2002) documented resolution of symptomatic VPD in 72 % of 162 patients so selected and treated. Of the remainder, surgical tightening or reconstruction of the central port with incomplete closure, as documented on post-pharyngeal flap nasendoscopy, increased the resolution of symptomatic VPD to 85 %. For the 11 residually symptomatic patients, a third tightening was performed or a narrow pharyngeal flap was placed in the center of the residual sphincter pharyngoplasty port. Significant hyponasality was noted in only 10 % of patients. Between 2003 and 2011, I performed 78 sphincter pharyngoplasties. Of these, 91 % had resolution of symptomatic VPD following the operation; the remaining 9 % had VPD resolution following sphincter port tightening. Of note, however, was the occurrence of either sleep apnea and/or symptomatic nasal secretion retention in 10 of the successfully managed patients. Initially managed with CPAP or BiPAP, 5 underwent subsequent port enlargement, 3 uvulectomy, and 2 combined port enlargement with uvulectomy for resolution of the symptomatology in all but 1 who required continued BiPAP.

Conclusion

Differential diagnosis of velopharyngeal dysfunction, using instrumental visualization of velopharyngeal function, allows for differential therapeutic management (Peat et al. 1994; Seagle et al. 2002). The objective of such management is to optimize the function of the velopharynx for speech tasks while minimizing the morbidity of the intervention upon the upper airway. My experience with such an approach over the past 34 years validates the assumption that differential management of velopharyngeal dysfunction based upon differential diagnosis can achieve this goal.

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Optimal Age for Palatoplasty to Facilitate Normal Speech Development: What Is the Evidence?

37

Sally J. Peterson-Falzone

Surgeons, speech pathologists, and other members of cleft palate/craniofacial teams have been trying for decades to gain some kind of "sure grip" on the relationship between age at *palatoplasty* and the child's *speech development*. I have reviewed the literature on this topic four times in the last 15 years: 1996 (Peterson-Falzone 1996), 2001 (Peterson-Falzone et al. 2001), in concert with my coauthors in 2004 (Peterson-Falzone 2004), and again with my coauthors in 2010 (Peterson-Falzone et al. 2010). All four of those reviews pointed out the differences between unverified clinical commentary (e.g., "Our speech results were good.") and scientific proof of investigators' conclusions about speech and language development. Much of the literature consisted of undocumented clinical insight, although there were many data-based reports as well. The reliance upon clinical insight and upon case reports that lack independent documentation of results still prevails as of this writing. On the one hand, one must commend the continuing and, indeed, zealous pursuit of this critical question. On the other hand, we must ask if there is a way to increase the relative proportion of reliable, valid data and decrease the dependence upon undocumented opinions.

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37.1 Why Is It So Difficult to Answer This Question?

Clinical research is inherently hazardous because so many factors affect the outcome of a study. The investigator(s) *cannot* control all the factors the way they would in a laboratory study. Far too often, however, the professionals who have tried to determine the optimum timing for closure of a cleft palate have not fully recognized the possible contaminating factors, such as:

1. Patients with varying types and extents of clefts, heterogeneous socioeconomic backgrounds, and varying health status and healthcare history (adequacy of pediatric care, otologic history, and documentation of hearing status; adequacy of early feeding and growth; adequacy of stimulation in the home).
2. Multiple surgeons *presumably* performing the same surgical procedure.
3. Children undergoing the same physical procedure but at different times in their chronological *and* developmental ages.
4. Children whose pre- and post-palatoplasty care may or may not have included regular follow-up evaluations by the cleft palate/craniofacial team (see recommendations from the American Cleft Palate-Craniofacial Association) (American Cleft Palate-Craniofacial 2009).
5. Children whose pre-and post-palatoplasty interventions may or may not have included early childhood stimulation programs.

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6. Highly variable documentation of the status of *velopharyngeal closure* post-palatoplasty. The substance of postoperative evaluation continues to vary widely. The low end of the spectrum consists of non-data-based statements such as "the palate appeared to move well after surgery" or "speech was significantly improved." The upper end consists of well-documented objective measures in combination with perceptual assessment of speech.
7. One-time postoperative assessment versus *longitudinal data* (kids grow, adenoids usually go away, maturity eventually kicks in, etc.).
8. Children who have undergone *primary veloplasty* (at varying ages), with inconsistent use of interim *obturating plates* (for varying lengths of time and typically with very little documentation of that use) and later surgical closure of the hard palate (again, at ages that vary from less 1 year to the early teenage years).[1]

Thus, the clinician who is trying to gather information on this topic must examine *each* report for the flaws that may have led to false conclusions.

37.2 Why Do Speech-Language Pathologists Worry So Much About Early Development?

One concept that may seem foreign to practitioners whose primary focus is the physical status of the child's palate, teeth, or oropharyngeal mechanism is that what an adult hears as normal speech (or speech development) in a toddler is *not* simply the result of that toddler echoing what he hears around him. *Normal communication development* depends upon many factors: normal hearing, normal oral and pharyngeal structures, adequate stimulation from the environment, and reinforcement from that environment for communication efforts. A child who experiences communication failure on a consistent basis will simply give up (unconsciously) and settle for the simplest signals he can produce (grunts, whines, cries, etc.) in order to make the world pay attention. Try to envision an otherwise normally developing child at the age of 1 year who is walking (or nearly ready to do so), smiling, and vocalizing but whose first attempts at words consist of only nasal consonants, grunts, and vowels. "Mama" will be produced normally (the consonant/m/does not require velopharyngeal closure), but any attempt at "dada" or "baba" (bottle) will sound like "uh uh." By contrast, a 12-month old who can produce "b," "d," "g," plus some of the consonants that do not require velopharyngeal closure (the nasal consonants /h/, /w/, and "y") will probably be producing ten recognizable (if simplistic) words. (Please see Fig. 37.1) His parents will reward these efforts with smiles and expanded inputs, e.g., "Da?! Do you want Daddy?" The child who cannot produce these speech patterns at this very young age will quickly recognize that his communication attempts are useless. He cannot convey much about what he wants, needs, or feels and will conclude that those around him do not care, "so why bother?" These early failed communication attempts have a long-term impact not only on language development but the child's overall cognitive and psychoeducational development. Communication success versus communication failure at 12–18 months of age will inevitably become either wonderful or disastrous well before the age of 3 years. The potentially dismal long-term outlook for the child's well-being thus becomes the concern of all his caregivers, including all members of the cleft palate/craniofacial team.

37.3 What Constitutes Scientific Evidence in Clinical Research?

The strongest evidence that a particular treatment protocol is better than another comes from prospective *clinical trials* that include study not only of the "treatment group" but of a carefully matched control group over the same period of time (American Speech-Language-Hearing

[1]The topic of primary veloplasty is a particularly difficult one to pursue in any scientific endeavor to answer the question of how early (or late) definitive palatal closure should be accomplished and will therefore be discussed in a separate section of this chapter (Sect. 37.6).

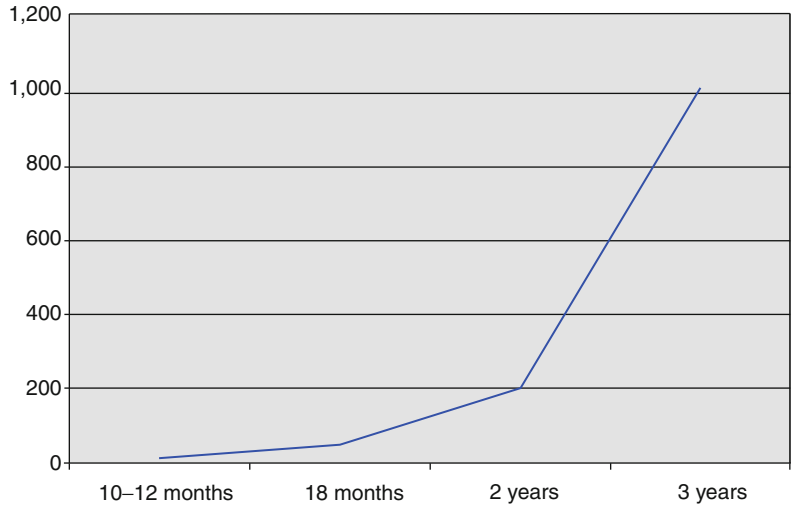

Fig. 37.1 Vocabulary development within the first 3 years of life. The number of words used at 10–12 months of age is not "0" but usually 2–10 words. By 18 months, most children have at least 50 words, and in the next 6 months that number quadruples to 200. By the age of 3 years, most normal children have speaking vocabularies of about 1,000 words

2003; Friedman et al. 1996). Prospective studies with a study group and a control group are also termed "cohort studies" (Wissow and Pascoe 1990). This is in contrast to "case control studies," which are usually retrospective in that they compare current cases with controls from the past (perhaps consisting of untreated cases or cases treated by an older method). Prospective clinical trials are the most difficult to execute. In fact, Wissow and Pascoe (Wissow and Pascoe 1990, p. 63) warned,

> Clinical trials are poorly suited to studies of (1) multiple therapeutic modalities (because too many subjects are needed to evaluate the many possible therapeutic combinations); (2) small changes in a therapeutic plan (the effort it takes to do the study may outweigh the potential significance of the outcome); (3) therapies that may be changed during the course of the study so that the results are at risk for becoming obsolete before the study is completed; and (4) treatments with only rare outcomes or outcomes that will be observable at a time far distance in the future.

Note how strongly # 1, 2, 3, and the second half of #4 pertain to the study of the best timing for palatoplasty.

Shprintzen (1991) made several observations that should be kept in mind by anyone engaged in clinical research and certainly by those who would study the question of the optimum timing of palatoplasty. He pointed out that most research is guided by a specific motive, usually a vested interest of the investigator, meaning the individual has an a priori bias regarding the outcome. With regard to sample selection, he pointed out the necessity of acknowledging the heterogeneous etiology of clefting and the influence this variation may have on development. He stated (p. 137), "…if a unilateral cleft lip and palate occurs because of an intrinsic hypoplasia of one maxillary process, it would be anticipated that the intrinsic hypoplasia would remain a factor throughout postnatal growth....If a major source of variability [in post-surgical growth] is intrinsic differences in facial growth potential related to population heterogeneity, then the subject population must be made as homogeneous as possible." He observed that, while investigators may have tried to eliminate all syndromic cases in their studies of this issue, none of the studies published up to the date of his commentary had held constant all the variables that could influence treatment outcome. At the same time, he warned (p. 138), "Holding all variables constant in patients with multiple problems is not only difficult, but the ethics of withholding treatment for the purposes of determining research outcome might have some difficulty passing an Institutional Review Board."

Table 37.1 rank-orders research designs by the strength of the evidence they can provide. In a simpler form, we may segregate the aspects of research design that weaken a study's conclusions versus those that lead to stronger supportive evidence for a particular treatment approach into "undesirable" versus "desirable":

Undesirable	Desirable
Investigator undertakes the study to prove that his treatment protocol is better than another	Investigator has no prior bias as to outcome of the study
Results recoded only by the investigator without independent observations by unbiased judges	Unbiased observers/recorders of results
Single recorder/ observer	Multiple independent observers/ recorders, with measurement of intra- and inter-observer reliability
One-time assessment of results	Longitudinal observation of results
Retrospective review of records	Prospective study
Single group of patients, treated according to one protocol, no control group	Cohort studies, comparing two or more groups of homogeneous patients, one treated according to the protocol under study, the other untreated or treated by a standard, accepted protocol
No attempt to control for the multiple variables that may influence results	Control, or at least careful independent documentation, of all variables (e.g., absence of associated anomalies, type and extent of cleft, health history, ear disease and hearing loss, socioeconomic factors including parental nurturing and stimulation)

The summary to both Table 37.1 and the comments above might be "If you want to prove that a given surgical protocol produces better speech results in nonsyndromic children with clefts, you need to plan a prospective, randomized study with matched controls; two or more surgeons performing the same procedure (to prove that it is not the surgeon's technique that is making the difference); *all* independent variables matched across patient pairs; large numbers of patients in each treatment group; results recorded by independent observers on standardized forms (not just anecdotal comments) and recorded over time rather than in a single post-treatment observation."

Obviously, this description is one of a utopia that one can only keep in vision, not plan on reaching. The clinical researcher is therefore charged with the task of trying to come as close to this as possible and being very careful in the interpretation of the results, taking into account all the factors that could not be controlled.

37.4 A General Warning: "A" Does Not Cause "B"

Because we clinicians are so anxious to answer the question that is the focus of this chapter, we are very prone to jump to conclusions about cause and effect. The most common trap is assuming that because "B" is strongly associated with "A," even if "B" *always* follows "A," that "A" *causes* "B." Friedman et al. (1996) termed this "causal inference." A widely used analogy points out that Tuesday always follows Monday, but does that mean Monday causes Tuesday? Even if we were to see that maxillary arch collapse is present in 100 % of children with cleft lip and palate following palatoplasty at 12 months of age, that does *not* mean that either the palatoplasty technique or the age at palatoplasty caused the arch collapse.[2] Cleft palate is not just a rearrangement or "disarrangement" of structures, but an inherent deficiency of tissue. The mere performance of palatoplasty, the particular surgical procedure employed, or the age at which it is performed thus cannot be blamed for subsequent arch collapse. On the other side of the argument, none of these three factors alone or in concert can be held responsible for either normal or abnormal subsequent development of communication development. If we were to segregate the physical variables from the nonphysical variables that

[2]Shprintzen (1991, pp. 137–138) made this same point: "Basing cause and effect relationships on the correlation of two factors is scientifically unsound, yet is undoubtedly the most common error made in research design and interpretation."

Table 37.1 Levels of evidence in clinical practice, ranked from weakest to strongest and what each type of evidence would require in a study attempting to relate age of palatoplasty to speech outcome

Source of evidence	What is involved	What would add strength to the evidence	Comments regarding the value of this form of evidence
A. "Expert opinion" or "clinical insight"	Minimally, one person's opinion on what works best	Agreement by other "experts," each with appropriate experience	Relatively easy to find; most vulnerable to dispute
[a]B. Retrospective case studies; all information taken from archived records (with or without historical controls)	Independent non-biased personnel who examine a pre-determined set of data recorded in a uniform format	Demonstration of intra- and interclinician reliability; historical controls would be patients with the same types and extents of clefts, non-syndromic, treated by some other surgical protocol	Probably the most common source of data on this question; subject to bias, especially if one or more of the investigators has a predetermined opinion; most difficult aspect is controlling for the myriad independent variables (e.g., hearing loss, overall health, socioeconomic factors, etc.) that affect outcome
[a]C. Retrospective case studies (archived information) combined with current clinical assessments	As in (B), above, plus current assessment of outcomes by independent, non-biased personnel using the same assessment protocol with all patients	Demonstration of intra- and interclinician reliability; control group(s) treated by other surgical protocols	Case histories + clinical assessments are very common, but, as in (B) above, investigator bias is frequently built into the research and control of independent variables is difficult
[a]D. Cohort studies (retrospective)	Investigator pairs each patient treated by Method X with a patient treated by Method Y. Each of the pair must match for the independent variables that could influence outcome	Demonstration of intra- and inter-clinician reliability; the larger the number of patients in each treatment group, and the longer the period of observation, the stronger the evidence	Currently a favored approach in studying this question, but extremely difficult to control for all the independent variables
[a]E. Cohort studies (prospective)	Investigator establishes the matched groups of patients before any treatment is rendered	Strength of study depends upon the investigator's ability to hold everything constant for each pair in the matched groups except for outcome; longitudinal follow-up of outcome strengthens the study	Very difficult to execute due to demands of matching patients on all independent variables; also, in longitudinal studies, patient populations change over time (growth, health factors, relocation, etc.)
[a]F. Prospective randomized trials	Investigator randomly assigns treatment protocols (i.e., X, Y, Z) to patients who are matched for type and extent of cleft, age, sex, other health factors (ear disease, etc.)	As in (E), above	Randomizing treatments raises serious ethical issues; patient consent is a must. Hospital IRB approval difficult to obtain. Strongest level of evidence, but rarely achieved

Designs "B" through "F" would require IRB (Institutional Review Board) review. Please see comments regarding HIPAA (the Health Insurance and Accountability Act of 1996) following this table

[a]The HIPAA (Health Insurance and Accountability Act of 1996) prohibits research personnel in health-care facilities from combing patient records unless permission has been granted by the patients themselves. The only person who can recruit the patients is the physician (clinician) who provided the care to the patient. This has an obvious impact in cleft palate/craniofacial care, where the preferred approach to care is through a multidisciplinary team (American Cleft Palate-Craniofacial Association 2000) and not by a single health-care professional. Historically, permission for examination of records or the sharing of information from patient records has not usually been obtained in the form currently required by HIPAA, rendering most retrospective studies impossible if they must meet HIPAA requirements. Information on HIPAA is available on the web at: http://www.hhs.gov/ocr/hipaa/

determine outcome from palatoplasty, we would be looking at the following:

Physical factors	Nonphysical factors
Type and extent of original cleft	Developmental level (chronological age compared to cognitive and communication development)
Presence/absence of associated anomalies, syndromes, etc.	Parent-infant bonding, parental stimulation
Overall health history (particularly with regard to infant feeding and otologic health)	Other psychosocial issues affecting the family
Adequacy of pediatric care both before and after surgery	Adequacy of relationship between surgeon (or team) and the family both before and after surgery

37.5 An Updated Review of the Literature

The main points made in the review of 2004 (Peterson-Falzone 1996) were as follows:

1. There was a very wide range in what clinicians considered to be "early" palatal surgery, e.g., the first 28 days of life (Denk and Magee 1996; Sandberg et al. 2002), 9–25 weeks (Copeland 1990), and up to 2–3 years (Koberg and Koblin 1973).
2. Judgment as to whether or not palatoplasty had in fact provided an intact palate often consisted of only visual observation of the oral cavity[3] (and sometimes only on a one-time basis), sometimes combined with rather pathetic "objective" measures, e.g., fogging on a mirror held beneath the child's nose during speech (Blijdorp and Muller 1984). At the other end of the continuum, many studies combined comparatively rigorous perceptual data with a full armamentarium of instrumental assessments.
3. The increased knowledge gained in the 1980s and 1990s regarding the effects of early structural constraints on pre-speech vocalizations and on later speech development in children with clefts could constitute a very good reason to perform palatal closure early, e.g., before the child reaches the milestone of producing his first meaningful words (Chapman 2011; Chapman et al. 2008; Estrem and Broen 1989; Grunwell and Russell 1987, 1988; Jones et al. 2003; O'Gara and Logemann 1988; O'Gara et al. 1994). Chapman (1991) found, as had previous researchers, that the process of phonologic acquisition in youngsters with clefts was "… slower, perhaps due to lingering or past articulatory and/or structural constraints on the speech mechanism."

What Is New Relative to This Point: The closer we look at the effects of early structural constraints on communication development, the more trouble we find. For example, Chapman (1993) found that school-age children with repaired clefts who had persistent speech and language problems were also more likely to have reading problems in comparison to children with clefts whose speech and language skills were within normal limits. Of course, we have long known that children and adults with clefts are at increased risk for reading problems and other learning disabilities [please see Peterson-Falzone et al. (2010, pp. 380–384)].

A 2006 study on infant vocalizations (Thom et al. 2006) provided acoustic and nasal pressure data showing that normal infants show gradual acquisition of velopharyngeal closure in their syllable utterances in the first 6 months of life. This process is not complete by the age of 6 months but is well under way. The authors conjectured that knowledge of this developmental process might help determine optimal timing for palate repair. These results fit in well with the theoretical framework proposed by Kemp-Fincham et al. (1990). The latter authors, reviewing the literature on the development of speech motor control and phonetic development in infants, concluded

[3]The examiner *may* be able to tell from the intraoral view alone whether or not the repaired palate is intact, but the velopharyngeal system is *not* visible because VP closure is the closure of the upper (nasal) surface of the velum against the posterior pharyngeal wall. Visualization of VP closure requires radiographic (usually fluoroscopic) or endoscopic instrumentation. Magnetic resonance imaging of the velopharyngeal system is also gaining a following, but is still too expensive to be a routine clinical tool.

that there is a particularly sensitive period or state of readiness between the ages of 4 and 6 months. Perhaps palatal closure before or during this time frame is important if we are to avoid the development of the maladaptive compensatory articulations that are so deleterious to speech intelligibility.

A 2008 study by Scherer and colleagues (2008) gave further validation to previous studies demonstrating the effects of early structural constraints (open clefts) on babbling at 12 months and subsequent size of consonant inventory when the children became toddlers. These authors found persistent vocalization and vocabulary deficits well beyond the time of palate closure, which in this study was 11–12 months. Note that this age is well within the time frame for palatal closure suggested by the American Cleft Palate-Craniofacial Association (2009). In comparison to a matched group of children without clefts, the children with clefts showed significant differences in frequency of babbling and "mean babbling level" at 12 months and deficits in speech sound accuracy and vocabulary production at 30 months of age. Scherer and colleagues (2008) recommended early intervention programs for babies with clefts to enhance speech and language development in babies with clefts.

4. A much-cited earlier claim by Dorf and Curtin (1982, 1990) that the chronological age of 12 months for completion of palatal closure was a cutoff line for preventing the development of maladaptive compensatory misarticulations in children with clefts was *not* supported by subsequent studies (Dalston 1992; Peterson-Falzone 1990) in which an attempt was made to verify that cutoff age. In addition, Dalston (1992) and Peterson-Falzone (1990) pointed out the critical information, such as age at the time of assessment, that was missing from the Dorf and Curtin publications. Unfortunately, 12 months is still often cited as a magical goal line for the performance of palatoplasty, ignoring problems in the methodology of Dorf and Curtin.
5. Despite the fact that their results could not be replicated in subsequent studies, Dorf and Curtin (1982) had made one very important point that was largely ignored by other investigators, namely, that the child's "articulation age" (level of *phonologic and phonemic development*), *not* his chronologic age, should be a prime consideration when trying to plan the best timing for surgery. That is, speech and language development vary among children: Some are attempting to produce 10–12 meaningful words at that age, while others are just beginning to use "mama." The more sounds and words the child is trying to produce, the more urgent the need for an intact palate and velopharyngeal system.

What Is New Relative to This Point: In a 2008 publication by Chapman and colleagues (2008), the authors tracked speech outcome in two matched groups of children with nonsyndromic clefts. One group was less lexically advanced (meaning that they were using fewer than five words) and younger (mean age of 11 months) when palate surgery was done. The second group was more lexically advanced (using five or more words) and a little older (mean age of 15 months) at the time of surgery. The children were assessed for 11 speech outcome measures between the ages of 33 and 42 months. The children in the first group had better articulation and less hypernasality than the children in the second group.

The authors pointed out that lexical difference between the two groups at the time of surgery would be expected because expressive vocabulary increases with age. They also pointed out that the children who were older and had larger vocabularies prior to surgery did not produce maladaptive compensatory misarticulations at a greater rate than children who were younger and had less advanced vocabularies; they simply exhibited poorer speech in general (Chapman and Hardin 1992). The authors did not have an explanation for the results, but suggested that future studies should compare children undergoing surgery by 6 months of age or prior to onset of babbling with children receiving surgery at approximately 12 months.

Interestingly, they did not specifically cite the results of Ysunza et al. (1988) who compared speech outcomes between groups of children operated at these exact ages.

6. Clinical surveys in the 1960s, 1970s, 1980s, and 1990s awakened us to the fact that associated anomalies, sequences, associations, and syndromes were probably affecting development in at least 50 % of children with clefts (Jones 1988; Shprintzen et al. 1985a, b; Womersley and Stone 1987). It is very easy for anyone other than an experienced dysmorphologist to miss subtle signs of other congenital defects in children with clefts. Furthermore, it is easy to miss signs of cognitive delays, hearing loss, and other threats to development, particularly if a child is *not* evaluated by an interdisciplinary team. Mixing surgical results from normally developing children with results from syndromic, multiply involved, or cognitively delayed children seriously impairs our ability to make decisions about the effects of any specific treatment.
7. Studies published during the 1990s on the ostensible relationship between timing of palatal surgery and subsequent speech development tended to support performance of surgery in the first 12–18 months of life, the exception to this general trend being the studies on primary veloplasty (reviewed in Sect. 37.6).[4]
   (a) Although their report was entitled "Correlation Between the Age at Repair and Speech Outcomes in Patients with Isolated Cleft Palate," Haapanen and Rantala (1992) did little to clarify the -age-at-surgery question because they had only small, uneven numbers of subjects; none were operated before the age of 1 year; and speech results consisted only of general categorizations based on one-time perceptual judgments without independent or objective documentation. These authors reported better speech in children whose palates had been closed between the ages of 16 and 20 months, as opposed to those whose surgeries had taken place either earlier (12–15 months) or later (21–24 months). They came to this conclusion because none of the children in the middle group (16–20 months at closure) had developed compensatory articulations. However, the number of children in this group was less than half the number in each of the other two groups. This fact, plus the one-time-only assessment with no longitudinal data, rendered the conclusion tenuous at best.
   (b) Marrinan et al. (1998) segregated 228 patients with four types of clefts (soft palate only, clefts of the hard and soft palate, unilateral cleft lip and palate, bilateral cleft lip and palate) into four groups based on age at closure: 8–10, 11–13, 14–16, and over 16 months. They found that secondary management for postoperative velopharyngeal inadequacy was necessary in 11 % of the 8–10 month group, 14 % of the 11–13 month group, 19 % of the 14–16 month group, and 32 % of the 16+ month group. This linear relationship between age at palate repair and need for a pharyngeal flap was reported to be statistically significant at the $p=0.025$ level. The likelihood of need for a pharyngeal flap was much greater in those children with clefts of the hard and soft palate (no lip cleft) or BCLP in comparison to those with clefts of the soft palate only or UCLP. In fact, in the latter two groups, age at palate repair was not statistically significant (ranging from 10 % need for pharyngeal flaps in the earliest repair up to 18 % in those repaired over the age of 16 months). The authors attributed this difference to the size of the cleft, since the vomer bone was attached in these two groups, whereas the need for secondary surgery in children with an unattached vomer (complete clefts of hard and soft palate or bilateral clefts) ranged from 12 % in the earliest repair group up to 50 % in those repaired over the age of 16 months.

[4]All experienced clinicians realize that "timing isn't everything" in palatal surgery. Extent of the defect (inversely related to the amount of available tissue), surgical technique, and experience of the surgeon all play a critical role. In 2001, Timmons and coauthors (2001) reported on speech results in 54 children. Twenty-seven had isolated cleft palate and were repaired by intravelar veloplasty at an average age of 9.6 months; 17 had unilateral clefts and were repaired by the same procedure at an average age of 8.9 months. When their speech was assessed between the ages of 5 and 7 years, "cleft-type speech characteristics" were present in 20/54 = 37 %. For these surgeons, operating below the age of 12 months was no guarantee of success.

(c) Ysunza et al. (1988) compared speech results in one group of children ($N=41$) whose clefts were closed at 12 months of age to another group ($N=35$) who were operated on at 6 months. This was a multifaceted study in which the postoperative evaluations included standardized perceptual evaluation of speech, videofluoroscopy, and nasopharyngoscopy. Somewhat surprisingly, early phonologic development was significantly better in the second group than in the first. In addition, none of the 6-month group showed subsequent development of maladaptive compensatory articulations, even though 6/35 (17 %) of them showed evidence of postoperative velopharyngeal inadequacy. By contrast, in the 8/41 (19 %) of the 12-month group that had VPI, 5 of these (62 %) developed compensatory articulations. There was no difference between the two groups in terms of degree of maxillary collapse when the patients were examined at 4 years of age. None had had either preoperative or postoperative orthopedic treatment. The authors concluded (p. 678), "Because maxillofacial growth was not significantly different in [the two] groups of patients, it seems that cleft palate closure at 6 months of age is a safe and reliable procedure for correcting velopharyngeal function in cleft palate patients."
It is interesting that Ysunza et al. (1988), using a full range of perceptual and objective tests, found a significant difference between children whose palates were repaired at 6 months of age and those repaired at 12 months.[5] As pointed out in Peterson-Falzone et al. (2001, 2010), these results may speak to the value of the theoretical framework proposed by Kemp-Fincham et al. in 1990 (Kemp-Fincham et al. 1990).

(d) A few surgeons in the 1990s were performing very early palatal closure (often simultaneous with lip closure) in the first weeks of life (Copeland 1990; Denk and Magee 1996; Sandberg et al. 2002). However, despite the fact that these papers were published 22 and 16 years ago, respectively, there have been *no* data on speech development in the babies so treated, nor has there been any independent substantiation of any of the early physical results of the surgery. Thus, these publications still constitute studies without results.

37.6 Primary Veloplasty: A Solution or a Problem?

Two-stage closure of the palate, with soft palate closure (often combined with lip closure) taking place in the first few months of life and closure of the hard palate delayed for variable periods of time (often until age 7 or beyond), continues to be the preferred treatment plan in many treatment centers. Those who prefer this approach state that closure of the lip and velum promotes a decrease in size of the hard palate cleft, making it easier to close at a later date with a less traumatic effect on the growing maxilla than would be the case with a complete surgical closure of the hard and soft palate in a single procedure. The timing of both the primary veloplasty and the later closure of the residual hard palate cleft has varied greatly both among treatment centers and in single centers that have changed their treatment regimen over time. Age at the time of soft palate closure has ranged from the first few weeks of life, 3, 6–8, 12–18 months, and up to the late age of 2 years and 6 months; age for closure of the hard palate has run the gamut from 12 months up to an extreme of 11–13 years (Bardach et al. 1984; Cosman and Falk 1980; DeLuke et al. 1997; Dingman and Argenta 1985; Friede et al. 1980, 1991; Greminger 1981; Harding and Campbell 1989;

[5]These authors were not the first to note a difference in speech outcome apparently related to differences in age at closure *within* the first year of life. Desai (1983) and Copeland (1990), in companion reports on the same groups of patients, reported differential effects of age at palatal surgery compared across 3 months, 4 months, 5 months, and 6 months of age. However, there were very uneven numbers of children in the four groups, and the short times separating the ages at surgery plus the rather questionable nature of the speech data (reportedly collected 5 years after palatal surgery but with an age range of the patients from "3.8 to 6.3 years") signaled the need for skepticism in interpreting the results.

Hotz et al. 1978; Jackson et al. 1983; Kramer et al. 1996; Lohmander-Agerskov 1998; Lohmander-Agerskov and Soderpalm 1993; Lohmander-Agerskov et al. 1993, 1994, 1995, 1996a, b, 1997, 1998; Meijer and Cohen 1990; Noordhoff et al. 1987; Noverraz et al. 1993; Poupard et al. 1983; Rohrich and Gosman 2004; Schweckendiek and Kruse 1990; Tanino et al. 1997; Van Demark et al. 1989; Vedung 1995; Wu et al. 1988).[6] [7]

In addition to the variability in timing and techniques of eventual hard palate closure in the regimen of primary veloplasty, interpretation of results of many clinical studies was complicated by (1) use of infant orthopedics in some clinic populations, primarily in Europe (Gnoinski 1982; Hotz et al. 1978, 1984; Hotz and Gnoinski 1979; Konst et al. 1999, 2000, 2003a, b, c, 2004), and by (2) sporadic, inconsistent use of obturating plates while the hard palate remained open. In fact, in reviewing these studies, it is very difficult to differentiate the information given about infant obturating plates from that pertaining to infant presurgical orthopedics. For example, in the patient population of Hotz et al. (1979, 1984), intraoral devices were designed both to mold palatal segments and to at least partially obturate the cleft. The "Hotz" plate (a passive plate) was also used in the series of studies by Konst et al. (1999, 2000, 2003a, b, c, 2004). Interestingly, when Konst et al. (2003c) analyzed the cost-effectiveness of their infant orthopedic treatment, they reported that treatment produced an average improvement in speech of 1.34 points on a 10-point scale, at a cost of approximately 1,041 Euro dollars.

As early as 1998, Lohmander-Agerskov (1998) concluded that the use of an intraoral plate had not proved to enhance articulatory development in her patients.[8] She remarked that use of the plate had been very inconsistent. Not all patients with open residual clefts had been fitted with plates, and consistency of use by any single patient could not be fully documented. A year later, Konst et al. (2004) reported the results of a prospective clinical trial that compared the prelexical (babbling stage) development of babies with unilateral clefts randomly assigned to either a presurgical infant orthopedic (PIO) group or a nonorthopedic group after birth. The "PIO" was a "Hotz" plate. At 12 months of age, the babies with plates were using more alveolar articulations than the non-plate group, but by 18 months of age, this difference was no longer apparent. In 2002, Hardin-Jones et al. (2002) similarly reported essentially no benefit of obturating plates on size of consonant inventory or place and manner of consonant production. In fact, there was a trend for the babies in the obturated group to produce more glottal consonants than the babies in the unobturated group.

The point should be made here that the obturating plates that have been used in babies to promote speech development have *not* been constructed to reach the posterior pharyngeal wall. Remember that very young infants are obligate nasal breathers, and to completely close off the nasopharynx would deprive them of an airway. Apparently, plates made to cover only the more anterior portions of the clefts are not enough to promote appropriate development of anterior consonants. As the baby grows, oral or oro-nasal breathing becomes possible, but perhaps the older the child becomes, the more likely he or she

[6] In the 2003 study by Lehner et al. (2003), hard palate closure was performed first, between 4 and 5 months of age, with later closure of the soft palate at 10–17 months. The authors concluded (p. 126), "There was no significant difference in terms of anterior and posterior maxillary width between early and delayed closure of [the] hard palate…." However, the timing of "delayed" hard palate closure was in no way comparable to what that term connotes in the rest of the literature.

[7] The clinical appeal of primary veloplasty was not enhanced by the article by Henkel et al. (2004) in which speech results were inexplicably determined *before* the hard palate had been closed and the open cleft was not obturated. The "test" for velopharyngeal closure consisted of whether or not the speech pathologist heard a "sound difference" between open-nose and closed-nose conditions. It is not surprising that "disordered articulation" (whatever that term meant) was present in half the population of 24 children.

[8] Although Dorf and coworkers (1985) promoted the use of an "articulation development prosthesis" for infants with clefts, their caseload consisted of just one child. No untreated children were studied for comparison regarding speech sound development.

rejects the device, dislodging it or just fussing so much that the parents are reluctant to reinsert it.[9] One also wonders if some babies actively (but unconsciously) try to avoid tongue contact with the foreign acrylic invader in their mouths. The findings of Hardin-Jones et al. (2002) particularly suggest this.

What Is New Relative to This Issue: Although the use of presurgical orthopedics is still part of the regimen in some treatment centers (Rohrich et al. 1996, 2000; Rohrich and Gosman 2004), several studies have been published in the last 12 years showing little benefit.[10] Six years after Lohmander-Agerskov concluded that intraoral obturating plates were not beneficial to speech development (Lohmander-Agerskov et al. 1993), she and her colleagues reported that intraoral presurgical orthopedics did not enhance articulation development (Lohmander et al. 2004).

Konst and coworkers (1999, 2000, 2003a, b, c, 2004) felt that their studies demonstrated a beneficial effect of presurgical infant orthopedics at an acceptable cost, but the size of their treatment groups was small (e.g., five children or less). In addition, their cost analysis did not include the costs to families in terms of number of treatment visits, time off work, transportation costs, or effects on family life.

The center in Gothenburg, Sweden, continues to produce the largest amount of data on patients undergoing primary veloplasty (Friede et al. 1991; Lohmander-Agerskov 1998; Lohmander-Agerskov and Soderpalm 1993; Lohmander-Agerskov et al. 1993, 1994, 1995, 1996a, b, 1997, 1998; Lohmander et al. 2002, 2004). In some of their later work, they began carrying the initial palatal surgery (veloplasty) further forward, providing more extensive surgical closure of the cleft and hoping to promote more postoperative spontaneous closure of the residual cleft.[11] They have also moved the timing of closure of the residual cleft forward to somewhat earlier ages than was the case in their original protocol.

In each of the Lohmander-Agerskov studies (Lohmander-Agerskov 1998; Lohmander-Agerskov and Soderpalm 1993; Lohmander-Agerskov et al. 1993, 1994, 1995, 1996a, b, 1997, 1998; Lohmander et al. 2002, 2004), perceptual speech outcomes were rigorously documented. Overall, these studies have shown a remarkably low incidence of maladaptive compensatory misarticulations (glottal stops, pharyngeal fricatives, etc.) both prior to and following full palatal closure, far less than one would expect when a child has reached school age with an open residual cleft. Perhaps this was the result of early intervention (speech therapy). Prior to definitive palatal closure, the children undergoing primary veloplasty have shown the expected hypernasal resonance and nasal air loss, but the articulatory problems have primarily been retracted placement of dental and dento-alveolar consonants to palatal and velar placements.

In 1993, Lohmander-Agerskov and Soderpalm (1994) stated that they had changed their clinical routine to include "early intervention programs, evaluation of speech development, and early speech therapy if necessary." However, in their 1998 paper (Lohmander et al. 2002), Lohmander-Agerskov and colleagues noted that speech therapy had only a "marginal effect" on the misarticulations (retracted oral consonants) found in 5-year-olds with large residual clefts. Other studies by the same group (Lohmander-Agerskov et al. 1995, 1996a, b; Lohmander et al. 2004) confirmed that adverse effects of an open cleft were evident in babbling and continued to have an effect on speech sound acquisition in the toddler, preschool, and school-age years.

[9]Maintaining the plate in place by pin retention avoids this problem, but has the potential of interfering with maxillary growth. In addition, constant apposition of the acrylic to the oral mucosa has deleterious effects on the tissue. In fact, when Holland et al. (2007) reported that "delayed closure of the hard palate leads to speech problems and deleterious maxillary growth," they were reporting results from a 5-year clinical period during which they were using pin-retained plates to cover the residual cleft after primary veloplasty. The poor results with regard to maxillary growth are not a surprise.

[10]The topic of presurgical orthopedics is discussed elsewhere in this book.

[11]The treatment groups in both Sweden and Texas (Rohrich et al. 1996, 2000; Rohrich and Gosman 2004) have reported instances of complete spontaneous closure of the residual cleft after primary veloplasty.

Other clinicians studying effects of the primary veloplasty regimen have also reported poor speech results (Bardach et al. 1984; Cosman and Falk 1980; Jackson et al. 1983; Noordhoff et al. 1987; Rohrich et al. 1996, 2000; Rohrich and Gosman 2004; Witzel et al. 1980). Rohrich and Gosman (2004) closed the hard palate simultaneously with the soft palate in 21 children at a mean age of 10.8 months (range 6–18 months). Another set of 23 children had primary veloplasty at a mean age of 11.4 months (range 6–22 months) and hard palate closure at an average age of 48.6 months (range 30–57 months).[12] They concluded (p. 236), "Our data suggest that delaying had palate closure results in significant speech impairment with a beneficial maxillofacial growth response." In a later publication, Rohrich et al. (1996) advocated a protocol of (1) presurgical orthopedics begun at 1–2 weeks of age, (2) lip closure and velar repair at 3–6 months of age, and closure of the residual hard palate cleft at 15–18 months.

It is pertinent here to point out that some other studies on growth have also cast doubt on the effects of delayed hard palate closure on maxillofacial growth. Noverraz et al. (1993) studied dental arch relationships in 88 children with unilateral clefts who underwent primary veloplasty at a mean age of 1.1 years and who were segregated into four groups depending upon timing of hard palate closure (modified von Langenbeck in all cases): 1.5, 4.6, 9.4 years, and unclosed by the age of 10 years. All of these children had received presurgical orthopedic treatment according to the method of Hotz (Hotz et al. 1978, 1984; Hotz and Gnoinski 1979), but only until the time of soft palate closure. These authors found *no* differences in the dental arch relationships among the four groups. Thus, age at hard palate closure, at least within the range of 1.5 up to 9.4 years, did not differentially affect maxillofacial growth. Gaggi et al. (2003) examined maxillofacial growth in 30 individuals with unilateral cleft lip and palate whose palates had been closed in a single procedure between 11 and 14 months, compared to 30 who had had veloplasty between 18 and 24 months and hard palate closure at 6 years. At 18 years of age, those patients in the delayed hard palate repair group had a more severe impairment in growth of the maxilla than those who had had a single procedure. This study did not include data on speech, but one can only imagine the devastating effect on speech sound development in those children whose palates were left completely open until the age of 18–24 months. By 24 months of age, most children are using at least two-word sentences.

Van Demark (1995) pointed out that advocates of primary veloplasty have ignored the financial and psychosocial costs of the increased number of surgical procedures (two palatal surgeries instead of one), the clinic visits for the fitting and maintenance of plates, and the intensive speech therapy such as implemented in the Zurich treatment center (Gnoinski 1982; Hotz et al. 1978, 1984; Hotz and Gnoinski 1979; Van Demark et al. 1989). These remarks may have particular pertinence in the USA, where third-party payers (insurance companies, HMOs) continue to cut services to the bare bones, meaning that families who need more team visits and more surgeries are apt to be paying out of pocket. To be blunt, a treatment protocol of primary veloplasty, which tends to be both care intensive and could be cost intensive, may be preferred in treatment centers functioning in countries in which medical care is fully socialized (such as Sweden), but it is less likely to gain a large following in countries such as the United States where medical care is largely pay-as-you-go.

There is little doubt that those centers that have devoted so much effort to studying and improving treatment outcomes using primary veloplasty will continue to do so. However, speech-language pathologists remain skeptical, especially when the provision of a fully repaired palate during the crucial time for speech sound acquisition can prevent the stigmata of "cleft palate speech."

[12]The large range of ages at surgery, both in the early complete closure and in the delayed closure groups, weakens the strength of this study.

Conclusions

Given the fact that innovations in palatal surgery are constantly being pursued in the effort to reduce trauma to tissue and minimize scarring [e.g., (Karsten et al. 2003; Mendoza et al. 1994)], there is reason to be optimistic that clinicians will come closer to making reasonable decisions about the best timing for palatal surgery, without the old argument of speech versus growth. For example, we no longer have to rely on such scar-inducing procedures as the Wardill-Kilner pushback (Ishikawa et al. 1998) or the infamous "island flap" (Greminger 1981). Surgeons can now produce better morphologic and physiologic results so that orthodontists do not have to fret about iatrogenic effects on maxillary growth. Significant palatal scarring and its subsequent effects on growth are *not* inevitable consequences just because palatal surgery is carried out early enough to facilitate normal communication development. Multicenter studies of all aspects of treatment outcomes (e.g., ScanCleft, DutchCleft, and AmeriCleft) are trying to iron out inconsistencies, particularly with regard to approaches to assessment of speech and craniofacial growth. A large part of the program of the next *International Congress on Cleft Palate and Craniofacial Anomalies* (Orlando, FL, 2013) will be devoted to progress reports on these issues.

Speech pathologists have continued to document the deleterious effects of an open palate on babbling in 6-month-olds and the subsequent effects on speech as the child reaches toddlerhood. There is no question that normal speech depends upon velopharyngeal closure being provided when it is needed,[13] which is long before full maxillary growth has been achieved. The expectation for every otherwise-normal child with a cleft should be normal speech development.

[13]Berkowitz (2004) was concerned about speech pathologists seeking palatal surgery for children with clefts based on age of the child. It is not the age but the developmental level of the child. See Chapman and Hardin (1992).

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38 Speech, Language, and Velopharyngeal Dysfunction: Management Throughout the Life of an Individual with Cleft Palate

John E. Riski

38.1 Introduction

One of the goals of cleft care is to establish normal speech. As providers, we attempt to achieve this goal in the face of the structural deficits of the cleft palate, a changing velopharyngeal environment, a developing dentofacial structure, and a propensity for hearing loss. We attempt to do this understanding that velopharyngeal competence is essential to normal speech learning. We are also working within a "speech readiness" period. If normal speech is not established or if maladaptive speech is not extinguished during this "readiness period," results can be less than optimal. This chapter will review the challenges and factors that influence the outcome of managing speech, language, and velopharyngeal dysfunction throughout the life of an individual with cleft palate.

38.2 Challenges

Despite the advances in surgical technique and procedures and the sophistication of instrumental assessment and imaging, there is little consensus on an optimal treatment protocol. A contributing factor might be the great variability in craniofacial morphology and the response to treatment in patients who have exactly the same cleft lip and palate diagnosis (Molsted 1999). While we have not been able to develop a consensus on management, we are developing a clearer understanding of factors that influence our treatment. Although there is no consensus on a management program, the American Cleft Palate-Craniofacial Association has published recommended parameters of practice for the care of individuals with congenital craniofacial anomalies (American Cleft Palate-Craniofacial Association 1993). The consensus conference participants who developed the document espoused several fundamental principles of optimal care including the importance of team care, parent/caregiver involvement, and longitudinal follow-up.

Hypernasality, nasal air emission, and aberrant articulation mark the failed primary management of the patient with cleft palate. Despite best attempts, primary palatal management is only successful in 70–80 % of individuals with cleft palate (Morris 1973; Riski 1979). That is, 20–30 % of children born with cleft palate will require secondary palate and management. Despite advances in instrumentation used to evaluate velopharyngeal function, we still have no way of predicting or identifying, at the time of initial palatal surgery, which 20–30 % of children will benefit from a pharyngoplasty. The speech pathologist must monitor emerging speech and language for any signs of nasal air

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escape, hypernasality, or aberrant misarticulation in order to identify children who have had an unsuccessful initial surgery and which children are candidates for secondary surgery.

In the last 40 years, great progress has been made toward a better understanding of many aspects of the cleft lip and palate defect, but considerable study remains before there is agreement on the optimal treatment procedures. Molsted (1999) provides an excellent review of the topic. Primary closure of the palate may fall into two main schools of thought. One advocates early closure of the lip and palate in order achieve normal speech function. The other recommends delayed closure of the hard palate in order to achieve maximum growth of the maxilla. A number of intercenter and multicenter studies have been carried out recently in an effort to investigate which procedures give the best result, both esthetically and functionally. The results are ambiguous, and this has led a number of researchers to suggest that a randomized clinical trial is the only way to resolve the ambiguity. The fact that it has proved difficult to identify the optimal procedures in the field of cleft lip and palate treatment may not only be due to a less than optimal research design. Another contributory factor might also be the great variability in craniofacial morphology and in the response to treatment in patients who have exactly the same cleft lip and palate diagnosis. Intensive research has made it possible to state categorically that clefts occur due to many different factors in an interplay between genetics and environment. Therefore, it is not likely that a single gene can be responsible for clefting.

Further complicating the discussion is the topic of fetal surgery. Since scar tissue presents many problems – for instance, impairment of growth – the reduction or prevention of scar formation has long been a desirable goal. The discovery that a fetus can heal without scar formation has led to many animal experiments. The timing of the surgical intervention on fetuses is critical, since late-gestation fetuses heal with adult-like scarring. There are still many unsolved problems connected with fetal surgery, and at present, prenatal surgery for repair of cleft lip and palate is not ethically defensible in humans. On the other hand, it appears that there are considerable possibilities for the reduction of human scarring after surgery with the introduction of various wound-healing medications.

Although oral clefts are among the most common birth defects, they remain a low-incidence disorder in pediatric practice. The prevalence is approximately 1:750–1:1,000 newborns in the Caucasian population and is even higher in other populations. It is estimated that there are 6,000 new CL/P cases in the United States annually. Despite the relative prevalence of oral clefts, these patients still represent a small part of the pediatric caseload outside craniofacial centers. Hypernasality, nasal air emission, and compensatory articulation are also low-incidence speech disorders and may result from structural, neurological, or functional (learned) etiologies. The rate of *hypernasality* after initial cleft palate closure is only 20–30 % (Riski 1979; Riski and DeLong 1984). Low-incidence disorders such as oral clefts and hypernasality offer few educational or clinical opportunities for developing clinical expertise. We are challenged to educate other healthcare workers about appropriate identification, diagnosis, and management.

A challenge to evaluation is that the etiologies of hypernasality and nasal airflow disorders are often occult or hidden. In recent reviews of patients receiving surgical correction for hypernasality, approximately 30 % did not have a cleft palate (Riski et al. 1992; Riski 1995). The etiology in these children is an anatomically deep nasopharynx that can be diagnosed accurately only by radiographic assessment. Normal velopharyngeal dimensions were described by Subtelny (1957) and highlighted by Zemlin (1997). Further, the anatomic defect of a disproportionately deep pharynx was described by Calnan (1971).

An issue in speech pathology that may have its origin in the low incidence and occult nature of the disorder is that noncleft hypernasality is erroneously labeled as a "voice disorder." Labeling hypernasality a "voice disorder" is ambiguous and often has dire consequences for the child and successful management of the disorder. Labeling

hypernasality as a voice disorder implies that it is a disorder of the larynx and results in delayed identification, labeling, referral, and management. Because the physical defect is not recognized, ineffective speech therapies are often poorly designed (Ruscello 1997). This further delays identification and referral to specialists at craniofacial teams. A cleft palate is identified by or at birth and palate closure is before 1 year of age (Riski 1995). In stark contrast, the average age of referral to our center for children with noncleft hypernasality resulting from Velo-Cardio-Facial Syndrome (VCFS) is 9.2 years of age! Children with hypernasality resulting from cleft palate are generally referred to the craniofacial team. In contrast, children with hypernasality with no obvious form of clefting are labeled as having a "voice disorder" and referred to an ENT, generally in a private office, who may or may not have experience evaluating or managing velopharyngeal dysfunction. This protocol followed in our school systems is a deterrent to successful identification, evaluation, and management.

Delayed management of VPI leads to increased failure of surgical intervention and refractory speech deficits. The rate of complete success when VPI is managed before 6 years of age is 90.9 %. The success rate falls to 73.9 % between 6 and 12 years, 70.0 % between 12 and 18 years, and 47.0 % after 18 years (Riski et al. 1992).

The evaluation of oral clefts and hypernasality is conducted by specialists in craniofacial clinics. However, Public Laws 94–142 and 99–457, which culminated in the Individuals with Disability Education Act (IDEA), have mandated that special services such as speech therapy be provided through specialists in schools and developmental centers. The professionals in these settings usually have limited experience with cleft-related problems because these problems usually form a very small part of their caseload. This separation of evaluation and therapy can lead to poor communication between professionals and therapy plans that do not directly address the therapy needs of the patient. There is an unquestionable need for partnerships between the evaluation centers and the settings in which the therapy is conducted.

38.3 Predicting the Need for Secondary Palate for Management

Prospective investigations designed to identify which children might benefit from primary pharyngoplasty have been unsuccessful. One investigation evaluated the speech and velopharyngeal function of 16 children who underwent a primary Orticochea (sphincter) pharyngoplasty at the time of palate closure (Riski et al. 1987). In the judgment of the surgeon, these children would have velopharyngeal incompetence following palatoplasty alone. Speech and velopharyngeal functioning were evaluated after 5 years of age. Analysis of the children's velopharyngeal function demonstrated that all had velopharyngeal competence. However, careful analysis of imaging studies demonstrated that one-half of children were demonstrating velopharyngeal closure above the point of pharyngoplasty insertion. Analysis of the children's speech demonstrated that one-fourth still had maladaptive speech problems associated with velopharyngeal incompetence. One might assume that in these children, the timing of surgery was too late. In contrast, one child, whose palate was closed at 30 months, had normal speech. Closure at 30 months was obviously not too late in this child.

Mazaheri et al. (1994) evaluated the usefulness of measurements taken from lateral cephalometric radiographs to predict later velopharyngeal competence. The lateral radiographs of 75 children with cleft palate were digitized for analysis. The measurements from radiographs of children who had velopharyngeal competence and acceptable speech were compared with those who presented with velopharyngeal incompetence requiring pharyngeal flap surgery or prosthesis. Soft tissue landmark points in the velopharyngeal region were digitized. Length and thickness of the soft palate and height and depth of the nasopharynx were measured. Evaluation of the growth curves of these four cephalometric variables indicated only two significant differences between children who later required pharyngeal flap surgery and those who did not. These differences were found in the growth in length of the

soft palate of the cleft palate-only group and in the growth in depth of the nasopharynx of the bilateral cleft lip/palate group. Based on the present cephalometric data, it was impossible to predict at an early age those cleft lip and/or palate patients who will later require management of velopharyngeal incompetence.

Other investigators have demonstrated that a careful speech analysis has important predictive value of future velopharyngeal competence. Van Demark and his colleagues produced a number of investigations in the 1970s that valuated the predictive value of the articulation test. Van Demark and Morris (1977) studied the articulation test scores for 278 subjects with cleft palate and compared the results with the normative data of the Iowa Pressure Articulation Test (IPAT) and the Templin-Darley Screening test of Articulation. In addition, the predictive value of the IPAT in relation to the need for secondary management was examined. Examination of the data concerning "risk rates" indicates that the IPAT is very useful in predicting the need for secondary management. Subjects who obtained scores of zero on the IPAT at four and one-half years of age had a 96 % risk of requiring secondary palatal management.

In 1975, Van Demark et al. (1975) evaluated the articulation scores, lateral x-rays, and clinical judgments of velopharyngeal competency of 75 subjects in order to determine their predictive value. Each of the measures resulted in at least 90 % appropriate predictions. However, the combination of articulation score and lateral x-ray rating appeared to be the best predictor for this particular sample in that subsequent treatment was correctly predicted for 96 % of the subjects.

38.4 Predicting Future Speech Proficiency

A single type of cleft palate does not result in a homogeneous population. Clefts of the same type (e.g., unilateral cleft of the lip palate) will have very different outcomes. Some children will develop normal speech following surgical repair of the cleft with very little other intervention. Do they have more of a chance of velopharyngeal competence, better hearing acuity, and a dental arch more conducive to normal speech? Other children develop speech and language more slowly and are ushered into early intervention programs. We still have no way of identifying which children are going to require extensive behavioral intervention without continually monitoring speech development and careful evaluation of speech and velopharyngeal function.

Although gender is not a significant variable in the outcome of surgery, it appears to be an important variable in the development of speech proficiency. Hardin et al. (1990) examined the relative contribution of clinical data obtained during speech examinations at age 4 years and at each subsequent age through 13 years. The purpose was to evaluate the ability of data from 4 years of age to predict proficiency at age 14 years. All children had unilateral cleft of the lip and palate. Sixteen independent variables used in the analysis at age 4 years included 4 nonspeech variables (gender, type and age of primary surgery, and pharyngeal flap surgery) and 12 speech measures obtained from speech examination. In subsequent analyses at ages 5 through 13 years, the rate of change between adjacent age levels for each of the 12 speech measures was also included for 28 independent variables. The results of the investigation indicated that a large percentage of the variance in judged speech proficiency at age 14 years could be accounted for, using clinical data obtained for speech examinations at ages 4 through 13 years. Regression analyses were performed for the total group of subjects at ages 4 through 13 years. The most efficient set of predictors accounted for 50–75 % of the variance in the dependent variable. The variable of gender was identified as the single most important predictor in nine of the ten regression analyses alone, accounting for at least 40 % of the variance in judged speech proficiency. When the regression analyses were performed separately for males and females, approximately 50–80 % of the variance was accounted for in judged speech proficiency for males and 50–90 % for females using one, two, or three of the independent variables. The most efficient set of predictors varied across age levels for both groups.

In addition, these predictors differed between males and females.

38.5 Articulation Development and the Age at Palatoplasty

A goal of cleft palate habilitation is the development of normal articulation. Factors that contribute to or interfere with normal development have been studied for some time. The child's age at the initial palate closure and type of cleft have been obvious factors to evaluate.

Just as early palate closure seems to lead to better palate function, so does it appear to lead to better articulation. Hardin et al. (1990) studied 48 children at 6 years of age and later in adolescence. Their results suggested the importance of establishing velopharyngeal competence by 6 years of age since resonance and articulation deficits maintained some stability after that age.

O'Gara et al. (1994) evaluated the speech development of 23 children with one syndrome unilateral cleft of the lip and palate from 5 to 35 months of age. The infants who had earlier palatal repair produced significantly higher percentages of oral stops (i.e., /p/, /b/, /t/) after 12 months of age than infants with later palatal repair, although there was no significant difference in mean frequency use of oral fricatives (i.e., /s/, /f/) up to 3 years of age. The results of this investigation support the earlier finding by Van Demark and Morris (1977) that the use of the oral stops sounds /p/ and /b/ is a good early indicator of velopharyngeal competence.

The severity of the cleft appears to influence the number of misarticulations. Riski and DeLong (1984) studied the articulation of a group of children longitudinally from 3 to 8 years of age. They reported that the number of misarticulations on an articulation screening test increased with severity of the cleft. That is, children with bilateral cleft lip and palate had more articulation errors than children with unilateral cleft lip and palate, who had more misarticulations than children with isolated cleft palate. As might be expected, children who also required a pharyngoplasty had more articulation errors than children who did not require a pharyngoplasty but had the same type of cleft.

The results of this investigation were supported by a later study of Finnish-speaking children (Laitinen et al. 1998). Those results demonstrated that the occurrence, severity, and number of errors of all studied sounds separately or grouped increased with the severity of the cleft, being constantly greatest in the BCLP group and lowest in the CL (A) group. Further, the boys tended to have more problems than girls in producing the studied sounds correctly.

38.6 The Stability of Velopharyngeal Functioning over Time

Despite the effort to create velopharyngeal competence with primary palatoplasty, there is evidence that velopharyngeal competence is not stable and may change throughout the life of a child VanDemark and Morris (1983). The roles of adenoid size and adenoid involution have been the focus of several investigations.

Gereau and Shprintzen (1988) evaluated the relative size of the adenoids relative to the success or failure of primary palatoplasty. They studied velopharyngeal valving in 850 children with nonsyndromic clefts, 138 children with syndromic clefts, and a group of children without clefts. They evaluated speech relative to adenoid size. Their data suggested a strong positive correlation between the incidence of hypernasal resonance post palatoplasty and relative adenoid size in the children with cleft palate. Velopharyngeal closure was observed consistently at the adenoids in all groups of children studied.

The importance of the adenoids in velopharyngeal closure was evaluated by Mason and Warren (1980). Specifically, they evaluated the phenomenon of gradual development of hypernasality as a result of adenoidal involution. They reported that hypernasality developed in two patients from a sample of 122 with repaired cleft palate. They identified three patterns of velopharyngeal closure against the adenoid mass from lateral radiographs.

One small-sample-size study does not support the contribution of adenoid involution to developing velopharyngeal incompetence in children with cleft palate. Van Demark et al. (1988) compared longitudinally perceptual ratings of articulation defectiveness, nasality, and velopharyngeal competency in 13 subjects who required secondary palatal management after age 10 with a second group who did not require a pharyngoplasty. Perceptual data when examined longitudinally did not adequately discriminate between subjects who at one time achieved velopharyngeal closure but who ultimately required secondary management and those patients who needed no further treatment. A decrease in articulation scores and an increase in severity of nasality and articulation defectiveness over time indicate that patients are at risk for secondary management. Evaluation of lateral x-rays indicated that (1) those in the group who required secondary operations demonstrated more variability in velopharyngeal closure than those in the comparison group, who required no secondary operations, and (2) adenoidal involution did not appear to be a significant factor.

Morris et al. (1990) evaluated the role of hypertrophied adenoids and any role they might have in maintaining velopharyngeal competence. Thirty-nine subjects were selected from a large longitudinal study based on availability of lateral still x-ray films taken in series from 5 to 16 years of age. Ratings of velar-pharyngeal contact and ratings of adenoid size were obtained from the films. The obtained data indicated the expected decrease in adenoid size. All patients maintained velopharyngeal competence during the period of study. However, three of the 39 subjects were judged to show loss of such contact during the period of study and an additional four had surgery for velopharyngeal incompetence after the completion of the study. All seven appeared to show significant deterioration of velopharyngeal status in middle or late adolescence.

In a larger study of 121 children born with cleft (lip) palate, Riski et al. (1996) evaluated velopharyngeal competence relative to adenoid involution. They reported that 11/121 (9.1 %) developed velopharyngeal incompetence with adenoid involution. Velopharyngeal function was evaluated with clinical and instrumental tools. Each of the 121 children initially demonstrated velopharyngeal closure characterized by the absence of any nasal air escape or hypernasality. However, at an average age of 5.7 years, nasal air escape was first detected. Lateral radiographs were taken at that time to monitor adenoid size. At an average age of 8.6 years, the adenoids had involuted 4.64 mm, leading to significant velopharyngeal incompetence requiring management.

The importance of the adenoid mass for velopharyngeal competence in children with cleft palate was also highlighted in a longitudinal study reported by Haapanen et al. (1993). They evaluated two groups of children with cleft palate. One group of 24 children had an adenoidectomy at the time of palatoplasty, and a second group of 25 children with cleft palate did not have an adenoidectomy. They found that hypernasality and nasal air escape occurred significantly more frequently in the group of children who had an adenoidectomy than in the group of children who did not. The groups did not differ when it came to articulation errors associated with velopharyngeal insufficiency.

38.7 Age at Pharyngoplasty

Given that 20 % of children will demonstrate velopharyngeal incompetence following palatoplasty, considerable effort has been expended to identify the ideal age for secondary cleft palate management. Many authors of studies of pharyngoplasties have commented on the most appropriate age for intervention. Most have reported better results in younger patients. An early study by Moll et al. (1963) found better results in patients younger than 15 years of age. Leanderson et al. (1974) evaluated the results of secondary management by pharyngeal flap in 124 patients with cleft palate. They reported the best results between 5 and 6 years of age. They observed continued speech improvements for several years after surgery in young patients. No improvement was found in older (over 25 years) patients.

Riski (1979) reported on longitudinal data of speech sound acquisition and resonance in 52

children who had had pharyngeal flaps for velopharyngeal incompetency. These data were compared to similar data for a group of 48 children with cleft palates who had not required secondary management. Comparisons were made also between the pre- and postsurgical performance of the group who had had pharyngeal flaps. Further comparisons were made between children who had secondary surgery before 6 years and after 6 years of age. Development of acceptable resonance and articulation patterns was slower in children who required pharyngeal flaps compared to those who did not. Further analysis showed that the post-pharyngeal flap group demonstrated acceleration in the acquisition of acceptable sound production in the year immediately following surgery. The data suggested that children who had flaps before 6 years of age made faster gains in the development of articulation and acceptable resonance than did children who were treated after the age of 6 years.

Van Demark and Hammerquist (1978) observed a trend for poorer articulation when a pharyngeal flap was done after 10 years of age. Surprisingly, when studied at 10 years of age, children with pharyngeal flaps done earlier than 5 years of age had poorer velopharyngeal competence ratings than children with pharyngeal flaps done later than 5 years of age. They concluded that the optimal time for pharyngeal flap was between 4 and 10 years of age.

Seyfer et al. (1988) reported the results of superiorly based pharyngeal flaps in 39 patients. All 11 (100 %) patients under 6 years of age improved. The average improvement was 1.1 on a 3-point scale. In contrast, 74 % of the 28 patients over 6 years of age improved by 1.4 on the same 3-point scale. The difference was not statistically significant, however; in addition, no significant difference was observed between the improvement for children less than 2 years old (including primary pharyngeal flaps) and those over 2 years old.

Van Demark and Hardin (1985) evaluated 129 patients with pharyngeal flaps. Articulation improved 15 % postoperatively and was only 90 % correct by 16 years of age. That is, speech was not normal in the oldest group studied. Although articulation ratings were better for patients receiving flaps earlier than 4 years of age or between 4 and 5 years of age, there was no difference in nasality when patients receiving flaps early and late were compared. They concluded that age at which pharyngeal flap surgery was performed was not critical in speech outcome.

A large series of children who had a sphincter pharyngoplasty was studied by Riski et al. (1992) using aerodynamic and imaging data. In this series, 109 of 139 patients demonstrated resolution of VPI. Sixteen of the initial failures were revised. The revision was successful in eight of 16, yielding an overall success rate of 117 of 139 (84.2 %).

Additional analysis of the series demonstrated that patients with mild hypernasality (90.38 %) were treated more successfully than those with severe hypernasality (71.26 %). Preoperative velopharyngeal orifice area, estimated by aerodynamic testing, influenced surgical outcome to some degree. The 64 patients with normal postoperative resonance demonstrated a preoperative gap of 9.9 mm^2; the 11 hypernasal patients had a larger preoperative gap of 12.2 mm^2, and the four hyponasal patients had a much smaller preoperative gap of 5.0 mm^2.

Age was a more significant determinant of outcome than was size of velopharyngeal gap. Sixty children under age 12 had a success rate of 84 % and a preoperative area of 9.6 mm^2. Twenty patients over 12 years had a success rate of only 55 % and a similar preoperative orifice area of 10.9 mm^2. A closer analysis of the age information revealed that the rate of complete success when VPI is managed before 6 years of age is 90.9 %. The success rate falls to 73.9 % between 6 and 12 years, 70.0 % between 12 and 18 years, and to 47.0 % after 18 years.

Finally Losken et al. (2003) found no statistical difference in the average age at initial sphincter pharyngoplasty in patients who required a revision (6.8 years + 4.48) compared to those who did not require revision (7.4 years + 4.4).

38.8 The Adult Patient

Although VPI is usually diagnosed and treated in childhood, it is not uncommon for adults to present for management. Younger and Dickson (1985)

treated eight adults with residual VPI following cleft palate repair as children. A superiorly based pharyngeal flap was used in each case. They reported "significant subjective and objective improvement," although they reported no data.

A more controlled study was reported by Hall et al. (1991). Twenty adult patients received a superiorly based pharyngeal flap. The authors reported normal resonance in 15 patients, hyponasality in three, and residual hypernasality in two. The heterogeneity of the sample made conclusions difficult. Of the 20 patients, 13 had unrepaired clefts, two had speech bulbs, and one had a failed pharyngeal flap.

38.9 Primary Versus Secondary Pharyngoplasty

Several studies have been reported of pharyngoplasty used during primary repair of the palate. The pharyngeal flap is advocated to provide additional tissue when the palatal cleft is extremely wide (Hardin et al. 1986). Both the pharyngeal flap (Dalston and Stuteville 1975) and the sphincter pharyngoplasty (Riski et al. 1987) have been used at the time of primary palatal closure in an attempt to increase velopharyngeal competence. Both procedures have a higher rate of success than a palatoplasty alone. An inferiorly based flap combined with a palatoplasty has been shown to eliminate hypernasality in 94 % of patients (Dalston and Stuteville 1975). A sphincter pharyngoplasty combined with a palatoplasty has been shown to have a 100 % success rate (Riski et al. 1987). Controversy still exists about using any secondary procedure with primary palatoplasty, because only 20–30 % of patients will eventually require pharyngoplasty and because of the potential obstruction. The ability to predict which infants will eventually require a pharyngoplasty appears to be poor. One surgeon attempted to identify infants who would eventually need a pharyngoplasty by visual inspection of the nasopharynx at the time of palatoplasty. Sixteen of these children were later evaluated. The pharyngoplasty appeared necessary in only 50 % (Riski et al. 1987). The remaining children achieved closure above the pharyngoplasty at the adenoids.

38.10 Speech Therapy

Our ability to evaluate velopharyngeal function is much more exact than our ability to treat velopharyngeal dysfunction and associated misarticulations with speech therapy. Many procedures have been proposed for treating velopharyngeal dysfunction, but there is little evidence that any of these techniques actually improves velopharyngeal function (Ruscello 1989). Currently, there seems to be a proliferation of oral motor exercises without any evidence supporting these procedures. Evidence from the literature suggests that nonspeech, oral motor exercises have little basis in fact. Oral motor exercises isolate oral movements by manipulating the oral structures either manually or with instruments such as toothbrushes. In contrast, speech is a coordinated process incorporating coordinated movements of respiration, laryngeal, velopharyngeal, and oral articulatory gestures. Proponents of oral motor exercises often proposed that nonspeech oral movements must develop before learning speech oral movements. Research suggests however that they develop in parallel.

Moore and colleagues have published a number of investigations evaluating whether speech emerges from earlier-appearing oral motor behaviors. Moore and Ruark (1996) designed an investigation to quantify the coordinative organization of mandibular muscles in toddlers during speech and nonspeech behaviors. Seven 15-month-olds were observed during spontaneous production of chewing, sucking, babbling, and speech. They found that mandibular coordination across these behaviors was quite different from that observed for other behaviors, even in the earliest stages of true word production. Production of true words was predominantly characterized by relatively stronger coupling among all mandibular muscles compared to earlier emerging chewing and sucking. Variegated babbling exhibited stronger coupling than reduplicated babbling, as well as chewing and sucking. They concluded that their findings did not support the suggestion that speech coordination emerges from earlier-appearing oral motor behaviors. Further, they concluded that the finding of coupled activation among mandibular antagonists during speech

paralleled earlier comparisons of adult speech and nonspeech behaviors.

In a follow-up investigation, Ruark and Moore (1997) reported on an investigation that was designed to quantify the coordinative organization of lip muscle activity of 2-year-old children during speech and nonspeech behaviors. They recorded electromyographic (EMG) activity from the right upper and lower lip activity of seven 2-year-old children during productions of chewing, syllable repetition, lip protrusion, and speech (repeated two-word utterances) tasks. Task comparisons revealed that the coordinative organization of upper and lower lip activity is task specific; different coordinative strategies are employed for different tasks. They concluded that the finding of coordinative elements of the perioral system of 2-year-olds is task specific and extends the results of previous studies of adults and children, where task-specific coordinative strategies were employed by the mandibular and perioral. Further, they concluded that the task-dependent coordination of the perioral system of 2-year-olds supports the notion that developing speech and earlier-developing oromotor behaviors (i.e., sucking, chewing) are mediated by different control mechanisms.

Parallel to the findings of parallel mechanisms for speech and nonspeech production is the report from Benson et al. (2001), who found a parallel rather than serial model of auditory speech and nonspeech perception. They identified candidate brain regions constituting a neural network for preattentive phonetic perception with MRI and multivariate multiple regression of imaging data. They used stimuli that were contrasted along speech/nonspeech, acoustic, or phonetic complexity (three levels each) and natural/synthetic dimensions. They found that seven distributed brain regions' activity correlated with speech and speech complexity dimensions, including five left-sided foci [posterior superior temporal gyrus (STG), angular gyrus, ventral occipitotemporal cortex, inferior/posterior supramarginal gyrus, and middle frontal gyrus (MFG)] and two right-sided foci (posterior STG and anterior insula). In contrast, only the left MFG discriminated natural and synthetic speech. They concluded that the data supported a parallel rather than serial model of auditory speech and nonspeech perception.

A place for isolated oral motor exercises has yet to be defined. There is the potential that maladaptive speech habits will continue and become more ingrained as ineffective and unproven exercises continue.

Continuous positive airway pressure (CPAP) therapy can be used to reduce hypernasality by elevating the air pressure in the nasal cavities during speech (Kuehn 1991). The purpose of this study was to determine whether increased intranasal air pressure loads the major muscle of velopharyngeal closure, the levator veli palatini. Nine subjects, four with cleft palate and five without cleft palate, were studied. Electromyographic activity was measured from the levator veli palatini muscle with several levels of air pressure delivered to the nasal cavities using a commercially available CPAP instrument. It was found that levator veli palatini activity was significantly greater for the positive air pressure conditions than for the atmospheric pressure conditions for both subject groups. This indicates that the levator veli palatini muscle acts against the resistive load produced by the increased intranasal air pressure. The results support the use of CPAP therapy as a method of resistance exercise for strengthening velopharyngeal closure muscles.

Very few studies have evaluated speech therapy treatment strategies for a child with cleft palate. Pamploma et al. (1999) compared the amount of speech therapy time required to correct compensatory articulation for two types of speech therapy. One group was treated with a phonologic-based intervention, and the second modality was treated with an articulatory or phonetic approach.

Twenty-nine patients ranging from 3 to 7 years were treated. Fifteen patients were included in the first group (control) and received articulatory therapy, and 14 patients were included in the second group (active) and received phonologic therapy. The median age of each group was similar; that of the control group was 54 months old, and that of the active group was 55.50 months old. The speech pathologist in charge of the speech therapy was the same in all cases. A blind procedure was utilized whereby each patient was

evaluated independently by two speech pathologists every 3 months until both examiners were convinced that compensatory articulation had been completely corrected.

The mean total time of speech therapy required for the normalization of speech in the two groups of patients was compared. The mean total time of therapy was 30.07 months in the control group and 14.50 months in the active group. A Student's *t*-test demonstrated that the total time of therapy was significantly reduced ($P < 0.001$) when a phonological intervention was utilized.

38.11 Role of Parents in Speech Therapy

The child developing speech and language might have a parent who is actively involved in providing speech and language stimulation or one who is passive. The evidence from studies of speech and language intervention is clear that the child with active parent participation develops better language skills. Studies of children with cleft lip/palate as well as other disabilities demonstrate that it is paramount that clinicians incorporate the parent as an active participant in speech therapy and speech and language stimulation.

Pamplona and Ysunza (2000) and Pamplona et al. (1996) investigated whether including the mother as an active participant in speech therapy sessions would improve the language development of children with cleft palate who also had additional language delays. One group was treated by the speech pathologist alone (control group), and a second group was treated by the speech pathologist but was also accompanied by their mothers (experimental group). Both groups were evaluated before and after treatment to evaluate the advance of each group. The patients accompanied by their mothers had significantly better language skills compared with patients treated without their mothers. They concluded that the results supported the statement that language development is related to mother-child mode of daily life interaction in children with cleft palate.

Additional support and evidence for parent involvement comes from Broen et al. (1998). They described the changes in a 3-year-old child's production of speech during a period of diagnostic therapy and the changes that occurred following the fitting of a speech prosthesis. The child's mother served as the primary intervener, guided by a speech-language pathologist. The mother was able to change the child's speech so that more of her productions were at a correct place of articulation. After structural management, nasal and glottalized productions disappeared from the child's speech, but glottal stops did not.

Support for the importance of parent participation is also found in investigations of children with other disorders. Janjua et al. (2002) evaluated language development in severe and profound hearing-impaired children and the type of parental interactions. They found that children who were directly involved in an activity with their parents develop better language, as were children who performed the task after their parents demonstrated it to them. The authors recommended that parents encourage child participation and use more contingent and child-centered interaction. In addition, they found no significant differences between oral and bilingual families in terms of quality of interaction.

Girolametto et al. (2002) studied 10 English-speaking mother-child dyads and 10 Italian-speaking mother-child dyads. All 20 children were late talkers who showed delays in expressive vocabulary development but age-appropriate cognitive and receptive language skills. The authors found some cultural differences in language learning and concluded that their results support the use of language interventions based on increasing maternal responsiveness for these children at the one-word stage of language development.

38.12 Language Development and Learning

The school-aged child with a cleft palate presents the clinician with unique considerations beyond articulation and velopharyngeal function. Evidence first surfaced in the 1970s that children with cleft palate might have educational problems that separate them from children without

clefts. Richman (1976) evaluated 44 cleft lip and palate/palate-only children who were individually matched with 44 normal school children on the basis of sex, age, grade, intelligence, and socioeconomic status. All children received behavioral ratings by classroom teachers, and achievement test scores were obtained. The children with cleft palate were rated by teachers as displaying significantly greater inhibition of impulse (internalizing behavior). Further, the children with clefts also scored significantly lower on overall basic skills achievement tests. At that time, Richman suggested that cleft children might be responding to the social-behavioral environment, which may include negative social responses from others.

Later, Richman (1980) examined patterns of cognitive ability in 57 cleft lip and palate children with verbal deficits, but without general intellectual impairment. He was attempting to answer the question of whether the verbal disability displayed by these children was related primarily to a specific verbal expression deficit or a more general symbolic mediation problem. Two groups of children were identified based on performance on cognitive tasks which required verbal mediation strategies without requiring vocal responses. The children with only a verbal expression problem performed significantly better on tasks requiring categorization and associative reasoning, although there were few apparent differences on memory items. Those children with a verbal expression deficit displayed both an underlying symbolic mediation deficiency and learning disabilities. There was a higher proportion of cleft palate-only children in the more severe group.

Richman and Eliason (1984) reviewed the research on intelligence, achievement, behavior, and personality of children with cleft lip and palate. Studies of intellectual functioning demonstrated that the general intelligence of cleft populations is relatively normally distributed with group mean IQ scores within the average range. There is some suggestion of a higher frequency of depressed verbal intellectual functioning relative to visual-motor intelligence. Factors that appear to affect IQ levels are presence of other congenital anomalies, speech and hearing deficiency, and low-incidence cleft type by sex occurrences. There is evidence that a high percentage of cleft children are underachievers. Personality and behavioral studies do not suggest significant psychopathology, although there is evidence of behavioral inhibition, concern regarding appearance, and decreased expectations by teachers and parents.

Richman and his colleagues first delineated specific reading and learning disabilities in children with isolated clefts. Richman and Eliason (1982) studied children with cleft lip and palate and cleft palate only. Both groups presented with reading disability and were matched for intelligence, age, sex, and reading level. The two groups were compared on reading and neuropsychological test variables. Subjects included 14 males and 10 females of each cleft type ranging in age from 8 to 13 years. The results showed a significant difference between groups on most language measures and differences in reading comprehension and type of reading errors. These results suggested that children with cleft palate only constitute a language-disorder group with more severe reading disabilities. Children with cleft lip and palate are more likely to have verbal expressive deficits and milder reading problems, possibly related to peripheral speech mechanisms.

Richman et al. (1988) were then able to delineate the incidence of reading disabilities in children with isolated cleft palate. They examined 172 elementary school children with cleft lip and palate (CLP) or cleft palate only (CPO). Approximately 35 % of the sample displayed a moderate degree of reading disability, and 17 % of the group exhibited severe reading disability. Reading disability was more prevalent at younger ages, presumably because of peripheral speech deficits. For older children, those with CLP displayed an incidence of reading disability similar to the general population (9 %). However, children with CPO exhibited a much higher rate of reading disability (33 %). There was no difference between genders in the prevalence of reading disability in this sample. This study supported their earlier research that suggested children with CPO may be more likely to experience general language disorders leading to long-term reading

disabilities. Children with CLP appear to manifest reading problems that tend to resolve with increased age.

Investigations that are more recent have highlighted the elevated incidence of learning disabilities in children with clefts, especially those with isolated cleft palate. Broder et al. (1998) examined the prevalence of learning disability (LD), level of school achievement, and prevalence of grade retention by type of cleft and gender at two craniofacial centers. They studied 84 consecutively evaluated patients from one center who were matched by cleft type, age, and gender with 84 patients evaluated at a second center. They found that 46 % of subjects with cleft had LD, 47 % had deficient educational progress, and 27 % had repeated a grade (excluding kindergarten) in school. Males with cleft palate only (CPO) had a significantly higher rate of LD than any other subject group. Males with CPO and females with CLP were more likely to repeat a grade in school than were females with CPO and males with CLP. They concluded that children with cleft are at risk for learning disability, low school achievement, and grade retention.

These investigations have prompted a series of studies looking at preschool children. Scherer and D'Antonio (1995) used a parent questionnaire as a component for screening early language development of children 16–30 months of age with cleft lip and palate. Thirty nonsyndromic children with cleft lip and palate and 30 children without clefts received the MacArthur Communicative Development Inventory: Toddler (CDI: Toddler), administered by a pediatrician. In addition, a speech-language screening was performed by a speech-language pathologist. Results of the two assessments indicated that the CDI: Toddler was a valid screener of language development compared to a comprehensive speech-language screening. They reported differences between the cleft and noncleft groups and found evidence of delays in expressive language development in the children with cleft lip and palate.

The source of language-learning disabilities was investigated by Ceponiene et al. (1999). They investigated persistence of auditory short-term memory (STM) that is implicitly involved in language-specific perception in children with clefts, grouped using fine-graded cleft classification. Cortical evoked potentials were recorded in 78 children with nonsyndromic oral clefts and in 32 healthy peers. A mismatch negativity (MMN) potential that indexes preattentive detection of change in auditory input was obtained in response to tone sounds. In order to test durability of short-term memory traces, sounds were presented with three stimulation rates. With slowest stimulation, MMN amplitudes were reduced in cleft children compared to the healthy peers ($P < 0.00065$). Only cleft lip children did not significantly differ from controls. Among isolated palatal clefts, the more posteriorly delimited the cleft was, the smaller was the amplitude of MMN. MMNs of smallest amplitudes were obtained in the subgroup of complete unilateral cleft of lip and palate. The authors concluded that the reduced MMN amplitudes found in cleft children imply deficiency in auditory STM trace maintenance. This dysfunction is likely to contribute to their language and learning disabilities. The MMN diminution with shorter/more posterior clefts suggests that differences in auditory cortex function are one of the underlying mechanisms of the cleft type-malcognition association.

In a follow-up study, Ceponiene et al. (2000) investigated preattentive auditory discrimination, reporting that it plays a significant role in language acquisition and usage. They evaluated infants with different cleft types. A mismatch negativity (MMN) component of brain evoked potentials, which indexes preconscious sound discrimination, and brain responses to rare sine-wave tones were recorded in 12 healthy infants and 32 infants with oral clefts at the ages of 0 and 6 months. Infants with clefts were subdivided into two categories: those with cleft lip and palate (CLP) ($n = 11$ at birth, $n = 6$ at the age of 6 months) and those with cleft palate only (CPO) ($n = 17$ at birth, $n = 8$ at the age of 6 months). At both ages, brain responses to rare sounds tended to be smaller in both cleft subgroups than in healthy peers. However, in the latency range of 300–500 ms, the MMN was significantly smaller in infants with CPO. In infants with CLP, the MMN

was comparable to that of healthy infants. Differences in auditory discrimination between infants with CLP and CPO, as reflected by MMN, were detectable at birth and persisted into later infancy. This pattern parallels known behavioral differences between children with these cleft types. The authors concluded that brain responses to rare sounds, in contrast, had no differentiative power with respect to the cleft type.

38.13 Preschool and School-Aged Investigations

Several investigators have evaluated language functions in infants, preschool, and school-aged children. Neiman and Savage (1997) obtained reports from caregivers to describe the developmental status of infants and toddlers with clefts. They studied 186 infants and toddlers with cleft lip, cleft palate, and cleft lip/palate, using the Kent Infant Developmental Scale and the Minnesota Child Development Inventory, both caregiver reports. Reports were obtained at one of the following age categories: 5, 13, 25, and 36 months. Data were analyzed in separate two-between ANOVAs (age × cleft type) for each developmental domain according to developmental assessment measure. Further, results were examined relative to the normative sample.

Lower-motor and self-help developmental quotients were lower at 5 months compared to the 13-month-old level. The 5-month-old infants with cleft exhibited "at-risk/delayed" development on the motor, self-help, and cognitive domains compared to the normative sample. Infants at 13 and 25 months were within normal limits in all developmental domains, with the exception of the 13-month-old infants with cleft palate, who demonstrate "at-risk" development in the motor domain. Toddlers with cleft palate exhibit "at-risk/delayed" development in the expressive language domain at 36 months.

Early delays were also reported by Broen et al. (1998). They studied the early cognitive and linguistic development of 29 children with cleft palate compared to that of 29 children without clefts. Measures included the mental scale of the Bayley Scales of Infant Development, the Minnesota Child Development Inventory, mean length of utterance, and words acquired by 24 months. Children with cleft palate, although well within the normal range, performed significantly below the children in the noncleft group on the mental scale of the Bayley Scales of Infant Development, some subscales of the Minnesota Child Development Inventory, and words acquired by 24 months. Differences observed in the cognitive development of children with and without cleft palate were verbal as opposed to nonverbal (i.e., linguistic in nature) and were related to hearing status at 12 months and velopharyngeal adequacy.

Chapman et al. (1998) examined the conversational skills of preschool and school-age children with cleft lip and palate during interactions with unfamiliar adults. Standardized measures of speech and language were administered, and ratings of resonance were obtained. Comparisons were made between the children with cleft lip and palate and their same-age peers on measures of conversational participation and a standardized test of pragmatic skills. Twenty children with unilateral cleft lip and palate (10 preschoolers and 10 school-age children) and 20 noncleft peers were matched for gender, age, and socioeconomic status.

Paired t-tests revealed no significant differences between the preschool and school-age children with cleft lip and palate and their noncleft peers in level of conversational participation. However, individual child comparisons revealed less assertive profiles of conversational participation for 50 % of the preschool and 20 % of the school-age children with cleft lip and palate. The investigators concluded that children with cleft lip and palate may show a less assertive style of conversational participation, at least during the preschool years. Therefore, craniofacial team evaluations should include examination of conversational competency, particularly for children who are demonstrating difficulty with other aspects of speech, language, or social development.

The work of Speltz and colleagues suggests that the differences between cleft lip/palate and cleft palate-only children may not be revealed until preschool or school age. Speltz et al. (2000)

used cognitive and psychomotor tasks to evaluate 29 infants with cleft lip and palate (CLP), 28 infants with cleft palate only (CPO), and a demographically matched comparison (COMP) group of 69 infants. The children were studied at ages 3, 12, and 24 months. The purpose was to examine predictors of cognitive status at age 24 months in the cleft group. Infants were administered the Bayley Scales of Infant Development (BSID), mother-infant interactions were observed, and medical records were reviewed. The CLP and CPO groups scored lower than the COMP group on the BSID, but did not differ from one another. Cleft group infants scored lower than COMP group infants on BSID items assessing nonverbal and expressive language skills. Quality of maternal interaction predicted the 2-year mental development index (MDI) scores of infants with clefts. They concluded that infants with clefts show relative deficits in cognitive and psychomotor development and that cognitive deficits are apparent in nonverbal as well as verbal areas of performance.

38.14 Velo-Cardio-Facial Syndrome

Velo-Cardio-Facial Syndrome (VCFS) was first described by Shprintzen et al. in 1978 Shprintzen et al. 1978. They described a pattern of similarities among 12 patients including overt or submucous clefts of the secondary palate, ventricular septal defects, typical facies, and learning disabilities. We have since learned that both DiGeorge syndrome and VCFS have a microscopic deletion of chromosome 22q11.2. Treating the speech, language, and velopharyngeal dysfunction in these patients present us with special challenges beyond those of patients with clefts and are considered separately.

Golding-Kushner et al. (1985) first described a rather distinctive pattern of language disorders and personality characteristics when they described language, academic, and psychological profiles of 26 patients with velo-cardio-facial syndrome. The characteristics continued through the course of development from initial language acquisition through childhood and adolescence.

Gerdes et al. (1999) suggested that the global delays and the variations in intelligence found are directly associated with the 22q11.2 deletion and are not explained by physical anomalies such as palatal defects or cardiac defects, or therapeutic interventions such as cardiac surgery. They reported that all of the children had late onset of verbal speech and behavioral disorders, including disinhibition and attention disorders. They strongly encourage early intervention services beginning in infancy to address the delays in gross motor skills, speech and language, and global developmental delays.

The observations of Gerdes et al. (1999) were supported by Scherer et al. (1999). They described the speech and language development of four children with VCFS studied longitudinally from 6 to 30 months of age and compared their performance with three groups of children: (1) normally developing children, (2) children with cleft lip and palate, and (3) children with isolated cleft palate. The data demonstrated that young children with VCFS show a receptive-expressive language impairment from the onset of language. Further, speech and expressive language development were severely delayed beyond a level predicted by their other developmental or receptive language performance. The children with VCFS showed severe limitations in speech sound inventories and early vocabulary development that far exceeded those shown by the children with cleft lip and palate and children with isolated cleft palate. The authors concluded that young children with VCFS emerge from a critical speech and language-learning period with severe limitations in their communicative abilities. They recommended additional studies to describe the later course of these early speech and language impairments and to explore the relationship to learning disabilities described for older children with VCFS.

More recently Murphy et al. (1998) investigated the presence of 22q11.2 deletion in individuals with learning disabilities. They evaluated 265 people with learning disability residing in two learning disability hospitals in South Wales. Individuals were selected for inclusion if they fulfilled any of the following criteria: psychotic disorder (schizophrenia or affective disorder),

family history of psychotic disorder, cleft palate and/or lip, congenital heart disease, broadly defined facial dysmorphism, or a history of hypocalcaemia. Fluorescence in situ hybridization studies were performed on 74 selected individuals. Cytogenetic analysis revealed that two individuals demonstrated a previously undetected chromosome 22qII deletion. A third person demonstrated a previously undetected cytogenetically visible deletion on chromosome 15. The authors concluded that VCFS appears to be etiologically significant in a proportion of individuals with idiopathic learning disability, especially in those where psychosis is associated with mild learning disability. They suggested that clinicians should consider a chromosome 22qII deletion in people who meet the selection criteria.

Citing the paucity of investigations evaluating education interventions, Kok and Solman (1995) reported their investigation of interactive computer-based instruction on the development of reading, language, spelling, and number skills. They recorded any positive effects of computer-based instruction on students' self-esteem, motivation, and competence in computer operational skills. They reported that the students were enthusiastic about the system, developed an interest in reading, and transferred remedial instruction to classroom performance. Comparison of pre- and posttest results indicated significant improvement in reading ability as measured using Neale Analysis.

Later, Scherer et al. (2001) described communication profiles in children with VCFS compared with children with Down syndrome. Four children with VCFS and four children with Down syndrome underwent cognitive and speech and language assessment. Communication profiles of children with Down syndrome showed a flat profile, indicating all measures were similar and delayed relative to chronological age. Children with VCFS showed vocabulary, pattern of sound types, and mean babbling length below cognitive and other language ages. The authors concluded that communication profiles of children with VCFS differed qualitatively and quantitatively from those of children with Down syndrome and supported the hypothesis that some children with VCFS present with a profile of communication impairment that may be distinctive to the syndrome.

The source of the learning difficulties and speech and language was investigated by Cheour et al. (1997). They evaluated the duration of auditory sensory memory, which is of central importance to speech perception and understanding. They used mismatch negativity (MMN), an attention-independent event-related potential which provides an objective electrical index of auditory sensory memory. They reported that the duration of this memory span is considerably shorter in 6–10-year-old children with VCFS than in healthy controls. They concluded that the language-related problems encountered in children suffering from VCFS are likely to be caused also by central nervous system (CNS) dysfunctions.

38.15 Summary

Individuals with cleft palate face many obstacles to normal speech and language throughout their life. One of the first challenges is to identify knowledgeable and skilled caregivers. Although clefting is a frequent birth defect, it is a low-incident disorder among pediatric disorders. This can limit the number of training programs and thus the number of treatment centers with skilled caregivers. Further, noncleft hypernasality is labeled a "voice disorder". This delays efforts to refer children with noncleft hypernasality to craniofacial teams for appropriate assessment and management. Considerable effort is needed to educate caregivers (especially speech-language pathologists, otolaryngologists, and pediatricians) that these types of problems are best evaluated and managed by craniofacial teams.

As clinicians, we are unable to predict, at the time of palatoplasty, those infants who will need secondary palatal management. Further, velopharyngeal function can be unstable in some children. As the child grows, the nasopharyngeal dimensions can become compromised as craniofacial structures grow and adenoids involute. This can lead to compromise of velopharyngeal function over the life of a child. Careful monitoring of emerging speech and velopharyngeal function remains paramount to early

identification of VPI and appropriate management. Earlier identification and management results in more successful outcomes.

The development of a single standard of treatment has been elusive. Speech-language pathologists are challenged to institute therapy regimens that are evidenced-based. There does not seem to be any evidence supporting oral motor exercises that isolate oral movements from other vocal tract structures used in speech. However, velopharyngeal exercise using CPAP and speech therapy with a phonologic approach hold some promise.

Finally, infants and children with clefts are known to have language and learning difficulties. Evaluation of emerging language in infants and language/learning skills in school-age children is indispensable. Programs that include a strong parental involvement seem to demonstrate more success than programs that do not.

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Prosthetic Speech Appliances for Patients with Cleft Palate

39

Mohammed Mazaheri

Cleft lip and palate habilitation began as an individual enterprise. The development of knowledge of the nature of the defect and of the remedial measures that were helpful in allowing these patients to achieve more normal participation in community life attracted additional specialists to aid in their habilitation. The need for communication and understanding among these interested disciplines has stimulated a rapid growth of cleft palate teams during the past 15 years. The need for this concerted effort is real. The specialists comprising the cleft palate teams differ in their training and experience, which has led to the development of different treatment philosophies.

In our group, the surgeon plays a dominant role (Cooper et al. 1960), because we believe that the reconstruction of a cleft palate is primarily a surgical challenge and an area where competent surgeons are capable of offering even more services to patients with cleft lip and palate than are universally practiced. Along with this philosophy, we maintain that a group enterprise by professionals is necessary in a complete habilitation program. In many areas of the world, trained personnel are not available to assist the cleft palate patient in all of his or her needs, and the surgeon has been forced to devise ingenious and occasionally extensive surgical procedures.

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Cleft palate surgery is not a stereotyped exercise, but rather a service demanding an assessment of all the factors presented by each patient and a reparative surgical plan based on proven principles. The majority of cleft palates can be reconstructed by trained surgeons to enable the patient to develop acceptable speech.

Many clefts of the hard palate can be closed by a vomer flap (Ivy 1960; Mazaheri 1962, 1964, 1973, 1976, 1977; Mazaheri and Hofmann 1962, 1965, 1969; Mazaheri 1961, 1970; Mazaheri et al. 1963, 1964, 1967, 1988, 1993; Millard 1962, 1966; Stark and DeHaan 1960; Veau and Borel 1931; Fauchard 1746; Grabb et al. 1971), and clefts of the soft palate can be closed by median suture with a good anatomical and functional result. The wide cleft and the short palate demand additional attention. Additional length may be gained by the Dorrance or a V-Y type retropositioning operation. The raw nasal surface may be covered with a skin graft (Giles and Fry 1921) or nasal mucosa (Dorrance 1925; Cronin 1957) or an island flap of palatal mucosa (Millard 1962, 1966). The incompetent palatopharyngeal valve can be augmented by a pharyngeal flap, either as a primary (Grabb et al. 1971) or secondary procedure. The need for additional tissue in a wide cleft can be satisfied by single or double regional flaps.

Associated with these surgical advantages now available to the cleft palate patient, there has also been a need for cleft palate prostheses. The trained prosthodontist has methods at his command to assist both the surgeon and the patient. Mutual understanding and restraint develop

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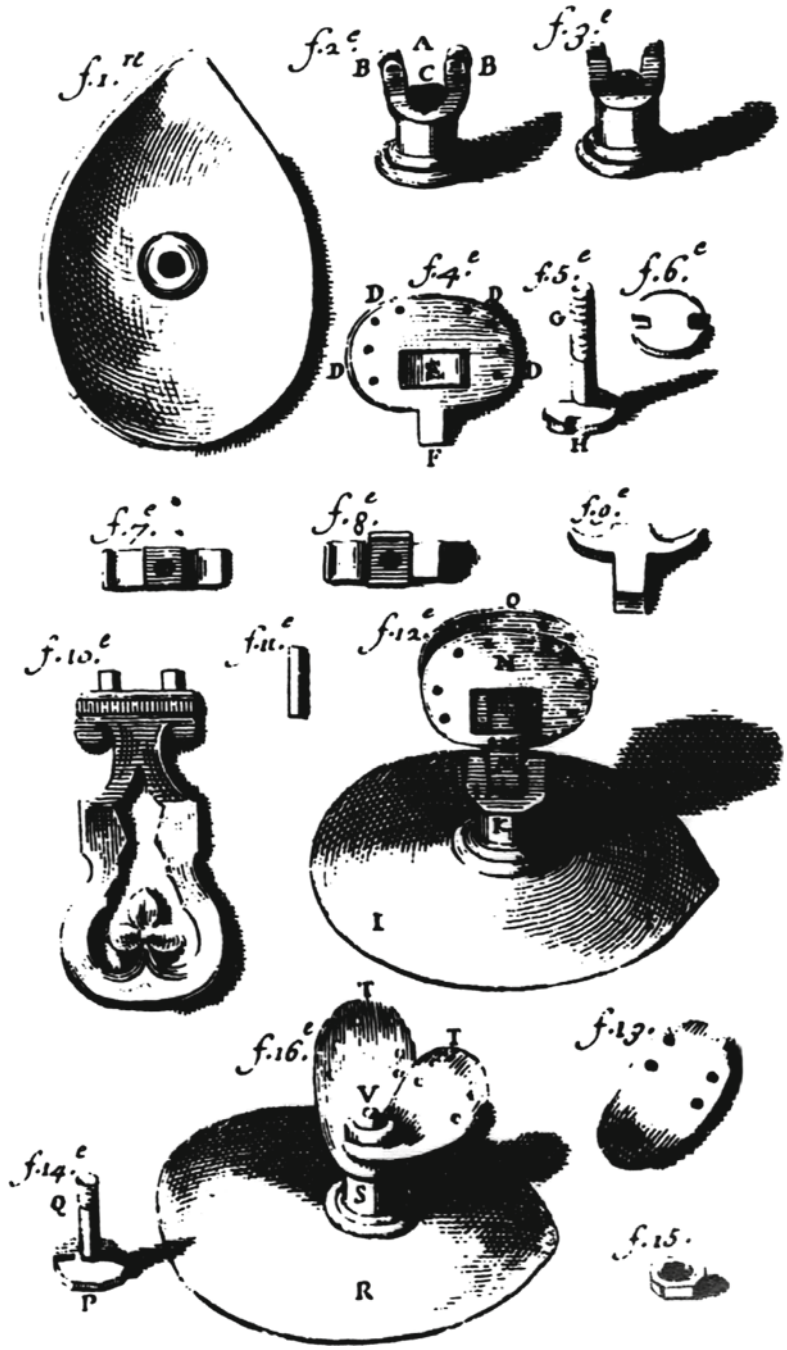

Fig. 39.1 Designs by Fauchard showing early obturators employed for palatal defects (From Fauchard 1746. Reprinted in Grabb et al. 1971)

among the specialists in a well-organized team to the benefit of the patient.

Since the early sixteenth century, dentists have been making appliances designed to close defects in the hard palate (Fig. 39.1). Like their predecessors, prosthodontists today are also vitally interested in helping the individual with an oral cleft. The prosthetic speech appliances they make are recommended as a preliminary or secondary treatment for many patients with cleft palate. The terms "obturator" and "speech appliance" will both be used in this chapter. They are frequently used synonymously, but it has become the custom to apply the term "obturator" to a device used in

treating acquired defects and "speech appliance" to one employed in treating congenital clefts.

The design and construction of speech appliances and obturators have changed much in the last 20 years, mainly due to improved materials and methods. We have, for example, better wax, acrylics, and impression materials and superior stone and plaster for the investment process. We also have benefited from improved diagnostic tools for evaluating the results of prosthetic treatment. Greater coordination of interdisciplinary team efforts has helped establish a more ideal prosthetic concept, one that ensures that fixed or removable partial denture prostheses are managed so as to preserve the integrity of all remaining teeth and the surrounding soft and hard oropharyngeal structures.

The decision for prosthetic rehabilitation is made based on the individual patients' needs, motivation for improvement, and availability of the suggested rehabilitative program. Approximately, 50 % of all patients with cleft lip and palate will need some type of fixed or removable prosthesis by 30 years of age.

As our knowledge and experience in the cleft palate field increased, those of us responsible for providing prosthetic care recognized the importance of establishing a better prosthodontic concept and principles regarding treatment. In rendering these patients the best service, we should first follow all the rules and principles governing the fixed and removable partial denture prosthesis and, secondly, should remove any fear of causing harm because of existing anatomic, functional, and physiologic deviation.

39.1 Diagnosis and Treatment Planning

In treating people whose oral-facial handicaps affect speech, the best results are achieved when the diagnosis and treatment are carried out by a group of clinicians who represent the various interested specialities and work together as a team rather than independently performing a series of procedures.

In diagnosis and treatment planning, full consideration should be given to (1) the type and width of the cleft; (2) the position and relation of the maxillary segments to each other in unilateral and bilateral clefts; (3) the form and lateral and anteroposterior dimensions of the maxillary arch; (4) the length, thickness, and mobility of the soft palate; (5) the perforations remaining in the hard and soft palate area and labial sulcus after surgery; (6) the posterior and lateral pharyngeal wall movement and the size of the nasopharynx; (7) a loose premaxilla; (8) the number of missing teeth; (9) malformed and malposed teeth; (10) partially erupted teeth; (11) teeth in the line of the cleft; (12) constricted maxillae; (13) the condition of the tonsils and adenoids; and (14) growth and development of the child. The patient's articulation, voice quality, hearing acuity, mental attitude, and general health also must be considered.

Socially acceptable speech cannot be produced without proper velopharyngeal valving. Therefore, surgical closure of the palate without due consideration of the depth of the nasopharynx and the length and function of the velum during phonation cannot satisfy this objective. Better understanding of the nature of the cleft, anatomy, and the physiology of the area involved would eliminate many of these difficulties. The results of surgical treatment of cleft palates should be evaluated with the aid of cineradiographic studies, nasal endoscopy, serial cephalometrics, maxillary and mandibular casts, speech recordings made before and after surgery, sound spectrographic analysis, measurements of nasal and oral air pressure and flow, and speech and audiometric evaluations.

All members of the team should be thoroughly familiar with the problem at hand. Often, the best result is not achieved when the knowledge of the specialists is not all encompassing (Cronin 1957).

The total habilitation and rehabilitation in the field of oral, facial, and speech impairment is achieved only when the following objectives are kept in mind: (l) socially acceptable speech, (2) restoration of the masticating apparatus, (3) aesthetic facial and dental harmony, and (4) psychologic adjustment of the patient to the condition.

Use of a speech appliance simply as a last resort is poor procedure. Its use must be clearly indicated by the oral conditions. For example, the indications for a prosthesis are clearly defined for a patient who has undergone a series of unsuccessful palatal operations. There is no magic in a prosthetic speech aid. However, there are some

patients for whom a prosthesis seems to be the only means of improving speech. In such situations, it fills a definite need. A prosthetic speech aid should be used for palatal conditions where it is indicated, just as the pharyngeal flap operation should be used only where it is indicated.

39.2 Treatment Planning

Treatment programs for cleft palate patients require careful planning and should include all factors involved in total health care. The interest of the dentist and physician in craniofacial growth and behavior of soft and hard tissues, both before and after surgery, has increased cooperation between surgeons and dentists. As a result, a dental specialist has the opportunity to examine the cleft palate child, with the surgeon, before any surgery is undertaken. Analysis of longitudinal maxillary and mandibular casts, cephalometrics, and radiographs has shown that two major factors cause growth disturbances of oral-facial regions in individuals with clefts: first, the inherent potential for growth disturbance present among cleft palate patients and, second, the trauma caused by surgical and orthopedic intervention. Because the first factor can be neither predicted nor reduced, efforts have been directed toward minimizing growth disturbance by performing surgery with the least amount of trauma and scar tissue. Longitudinal data obtained during the past 4 years regarding the surgical closure of the cleft with minimum amount of scar tissue and trauma are very encouraging (Millard 1966).

39.3 Requirements of a Speech Appliance

1. The prosthesis must be designed for the individual patient in relation to his oral and facial balance, masticatory function, and speech.
2. Knowledge related to removable partial and complete dentures should be used in designing the maxillary part of the cleft palate prosthesis. Preservation of remaining dentition and surrounding soft and hard tissue in cleft palate patients is of utmost importance. Improperly designed cleft palate appliances can result in premature loss of both hard and soft tissue, further complicating prosthetic habilitation.
3. The prosthetic speech appliance should have more retention and support than most other restorations. The crowning and splinting of the abutment teeth in adult patients may increase retention and support of the prosthesis and may extend the life expectancy of abutment teeth.
4. Mouth preparations should be completed before making final impressions. In cases where lateral and vertical growth of the maxilla is incomplete and partial eruption of the deciduous and permanent teeth is evident, careful mouth preparations should be made. To provide support of the prosthesis, these preparations may include gingivectomies to expose clinical crowns (to make them usable) and the placement of copings on remaining teeth to prevent decalcification and caries. Osseointegrated implants have been a great help in gaining adequate retention for the prosthesis.
5. The weight and size of the prosthetic speech appliance should be kept to a minimum.
6. The materials used should lend themselves easily to repair, extension, and reduction.
7. Soft tissue displacement in the velar and nasopharyngeal areas by the prosthesis should be avoided.
8. The velar and pharyngeal sections of the prosthesis should never be displaced by movements of the lateral and posterior pharyngeal wall muscles or the tongue during swallowing and speech.
9. The superior portion of the pharyngeal section should be sloped laterally to eliminate the collection of nasal secretions. The inferior portion of the pharyngeal section should be slightly concave to allow freedom of tongue movement.
10. The location and the changes of the speech bulb should include consideration of the following factors:
    (a) The speech bulb should be positioned in the location of greatest posterior and lateral pharyngeal wall activity, because voice quality is judged best when the speech bulb is at these positions.

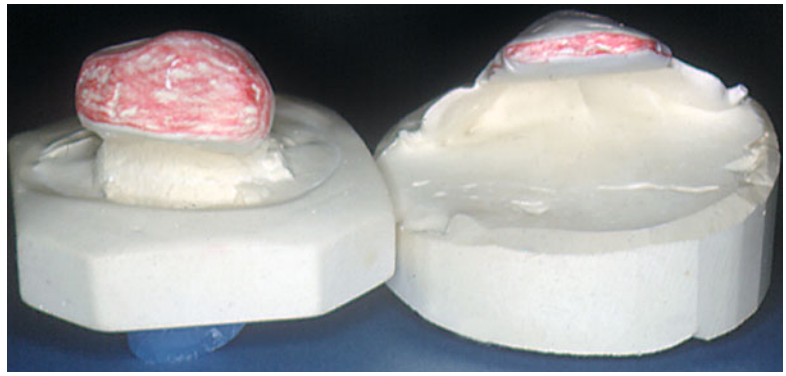

Fig. 39.2 As a result of our studies, we have concluded that the inferior-superior dimensions of the speech bulb do not have a significant effect on speech quality as long as the bulb is properly placed to facilitate good velopharyngeal closure. This dimension was reduced to one-quarter of its original size, as shown in cast made during fitting for one patient, without apparent effect on nasal resonance

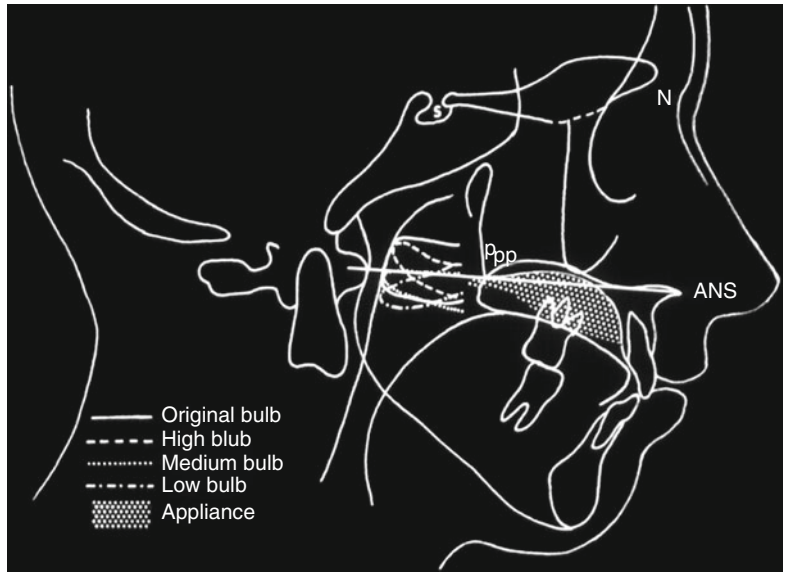

Fig. 39.3 Superimposed tracing of the original speech bulb and various experimental speech bulbs. The palatal plane was used as a plane of reference along with posterior pharyngeal wall activity, muscle bulge, or Passavant's pad. The posterior nasal spine (*PNS*), absent in cleft palate subjects, is called posterior palatal point (*Ppp*) and represents the most posterior point of the remnants of the palatal shelves as shown in the lateral cephalometric film. Median position was judged best

(b) The inferior-superior dimension and weight of the speech bulb may be reduced without apparent effect on nasal resonance. (The lateral dimension of the bulb does not change significantly as the position is varied.) (Fig. 39.2)
(c) The speech bulb should be placed on or above the palatal plane in cases where posterior and lateral pharyngeal wall activities are not present or where visual observation of the bulb is not possible, due to a long, soft palate (Fig. 39.3).
(d) The anterior tubercle of the atlas bone can be used as a reference point; however, investigation has shown that the relative position of the tubercle of the atlas bone

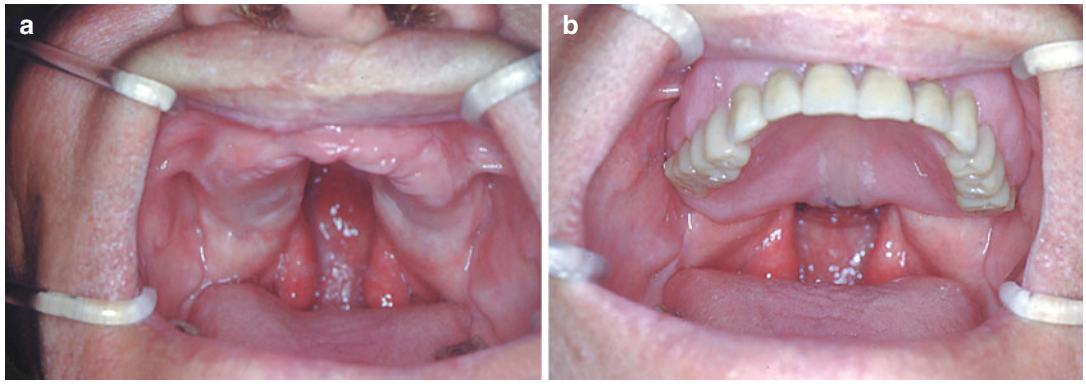

Fig. 39.4 (**a**) An edentulous patient with an unoperated cleft of the soft and hard palate that affects the retention and support of the prosthesis. At no time should a patient with a cleft, especially an unoperated cleft, be rendered edentulous. (**b**) The completed prosthetic speech appliance in position

varies in different individuals and that the positions of the velopharyngeal structures change in relation to the tubercle as the individual moves his or her head. Therefore, the atlas bone is no longer used as the reference point for positioning of the pharyngeal section of the bulb.

39.4 Indications for Prostheses in Unoperated Palates

Cleft palate surgery is not a stereotyped exercise, but rather a service that demands an assessment of all factors presented by each patient and a reparative surgical plan based on proven principles. The majority of cleft palates can be reconstructed by surgery, enabling the patient to develop acceptable velopharyngeal closure. However, in some situations, a prosthesis is the physical restoration of choice. This decision should be made by the group charged with the habilitation of the cleft palate patient.

Many clefts of the hard palate can be closed by a vomer flap (Mazaheri 1973; Veau and Borel 1931) and clefts of the soft palate by median suture with good anatomic and functional result. The wide cleft and the short palate demand further attention. Additional length may be gained by a Dorrance or V-Y type retropositioning operation. The raw nasal surface may be covered with a skin graft, nasal mucosa, or an island flap of palatal mucosa (Cronin 1957; Giles and Fry 1921; Veau and Borel 1931). The incompetent palatopharyngeal valve can be augmented by a pharyngeal flap, as either a primary or secondary procedure (Grabb et al. 1971). The need for additional tissue in a wide cleft can be satisfied by single or double regional flaps.

Despite the surgical advances available to the cleft palate patient, a need remains for cleft palate prostheses. The prosthodontist can assist both the surgeon and patient, and the mutual understanding among the specialists in a well-organized team is of great benefit to the patient. Some situations indicating a prosthetic approach are discussed in the following paragraphs.

39.4.1 A Wide Cleft with a Deficient Soft Palate

Some clefts of this type do not lend themselves to a surgical repair by means of local flaps. A prosthesis is preferable to the more time-consuming remote flaps in these situations. Many patients need a prosthesis to restore missing dental units, and the distant tissue provides only a dynamic mass (Figs. 39.4 and 39.5).

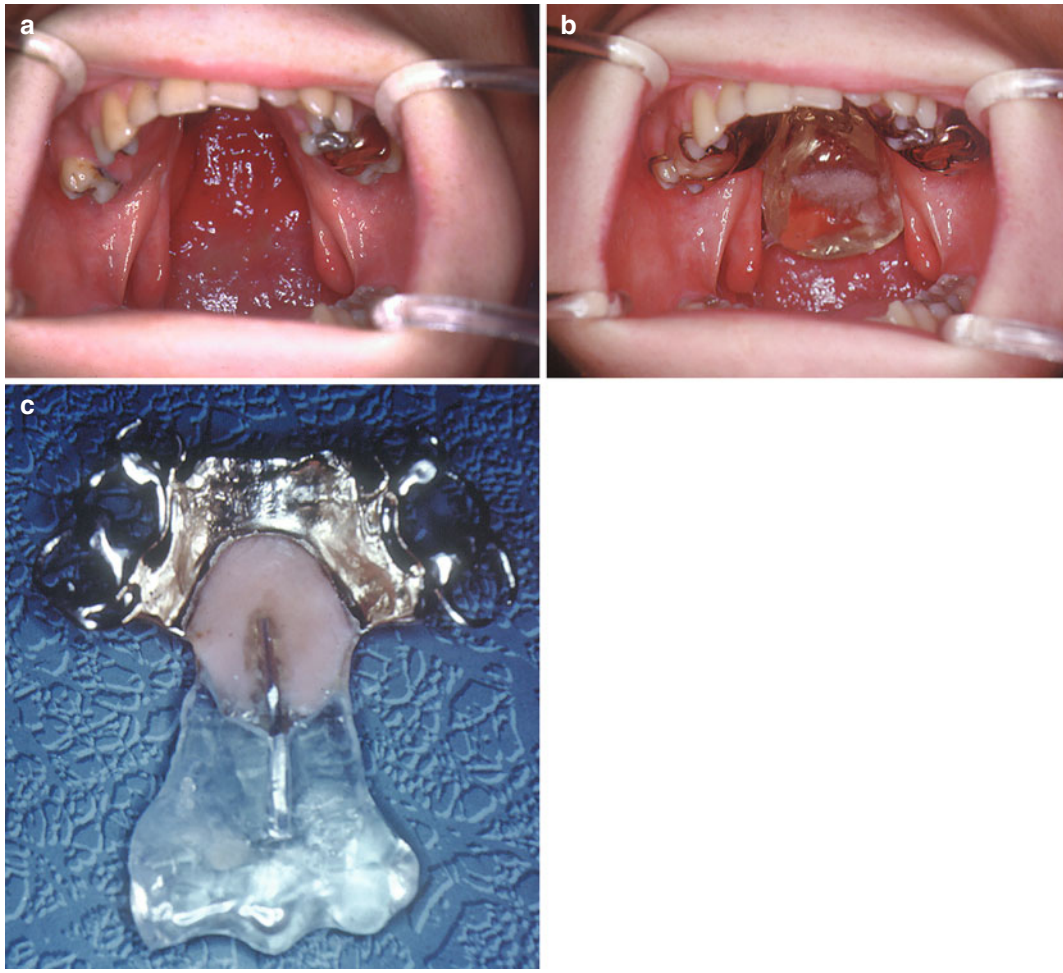

Fig. 39.5 (**a**) Patient at the age of 16 years with a very wide cleft of the soft and hard palate. (**b**) Prosthetic speech aid in position. Note that the pharyngeal section of the speech aid is placed directly over the posterior and lateral pharyngeal wall muscle activities. (**c**) Oral view of prosthetic speech aid. The utilization of second bicuspids and first and second molars for retention and support will prevent this prosthesis from dislodging into the nasal cavity during swallowing and speaking

39.4.2 A Wide Cleft of the Hard Palate

In bilateral clefts, the vomer may be high and the cleft of the hard palate wide so that a surgical repair may produce a low-vaulted palate. It may be possible to close the soft palate with the aid of local flaps and to restore the hard palate with a prosthesis. A situation similar to that once advocated by Gillies and Fry (Limberg 1927) is created: the primary repair of the velum may create a more favorable spatial arrangement for subsequent surgery on the hard palate.

39.4.3 Neuromuscular Deficiency of the Soft Palate and Pharynx

Repair of the palate would not be conducive to the development of good speech. It is difficult to create and maintain a pharyngeal flap large enough to produce competent palatopharyngeal valving without obstructing the airway in the presence of a neurogenic deficiency of the critical muscles. A pharyngeal flap serves best when surrounded by dynamic musculature. When this situation does not exist, the pharyngeal section of a

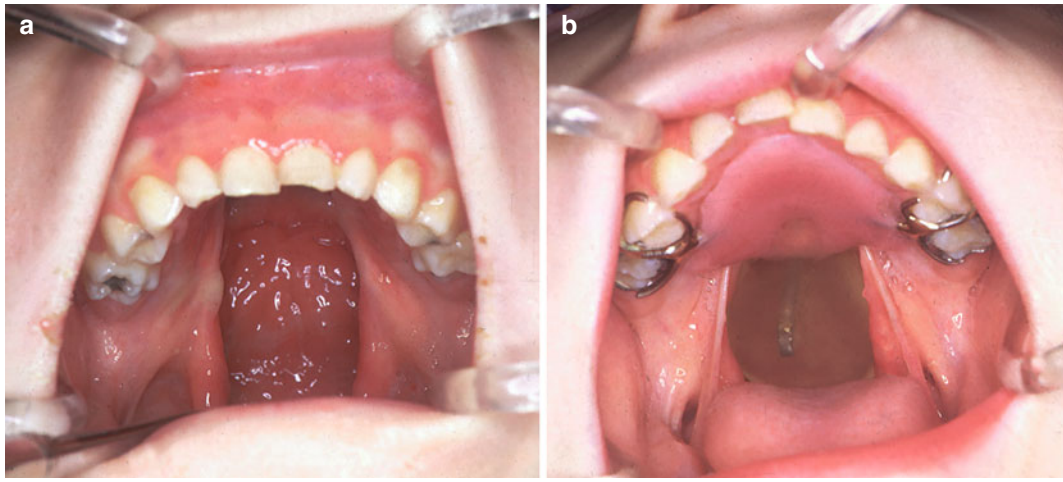

Fig. 39.6 (**a**) A four and half-year-old girl with a rather wide cleft of the soft and hard palate. We elected to fit her with a prosthesis and to delay the palatal surgery until a later age. (**b**) The prosthetic speech aid in position. She tolerated the prosthesis, and the speech significantly improved within a 6-month period

speech-aid prosthesis may serve better to reduce nasality and nasal emission. The prosthesis can also act as a physical therapy modality, providing a resistive mass for the muscles to act against. Should muscle function improve, definitive surgical measures can then be contemplated.

39.4.4 Delayed Surgery

When surgery is delayed for medical reasons, or when the surgeon prefers to repair the palate when the patient is older, the cleft palate may be temporarily closed with a prosthetic speech aid (Fig. 39.6).

39.4.5 Expansion Prosthesis to Improve Spatial Relations

An expansion prosthesis may be used to restore and maintain more normal spatial relations of the maxillary segments prior to surgery. These segments can be gradually separated by an expansion prosthesis to create a space for the premaxilla or to stabilize the parts in a normal position in association with an autogenous bone graft. The use of an expansion or repositioning prosthesis, with or without bone grafting, is appropriate for selected cases. In the majority of cleft lip and palate patients, restoration of the anatomic continuity of the labial muscle would mold the segments into acceptable relationships to each other and to the mandible.

39.4.6 Combined Prosthesis and Orthodontic Appliance

An orthodontic appliance may be combined with a prosthesis to move malposed teeth into a more favorable alignment. A prosthetic speech appliance, such as the one illustrated in Fig. 39.7, could be designed for a patient receiving full-band orthodontic treatment.

39.5 Indications for a Prosthesis in Operated Palates

39.5.1 Incompetent Palatopharyngeal Mechanisms

If clinical, nasal endoscopic, and cineradiographic analyses suggest that the patient is near a functional closure, a prosthesis may serve as a physical therapy modality. The pharyngeal section of the prosthesis is gradually reduced as

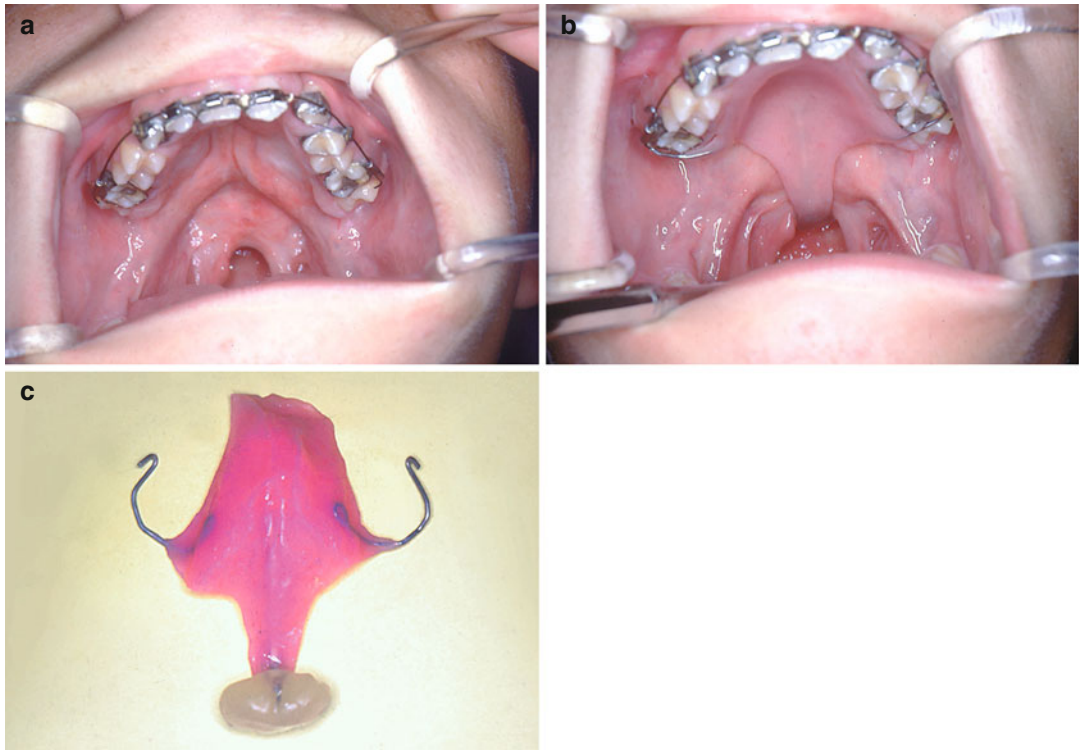

Fig. 39.7 (**a**, **b**) A temporary prosthetic speech appliance was designed not to interfere with orthodontic treatment while the patient was under active therapy. (**a**) View of the palate without prosthesis. (**b**) The prosthesis in position. Retention is obtained by placing the retainers above the molar buccal tubes. (**c**) View of the prosthesis after 1 year of velopharyngeal and lateral pharyngeal wall activity, resulting in acceptable speech. Prosthesis was then discarded

muscle function improves, and the prosthesis is eventually discarded. When the patient presents a large velopharyngeal gap associated with a neurogenic deficiency, the speech-aid prosthesis should be considered as a permanent treatment.

39.5.2 Surgical Failures

A prosthesis should be considered when a patient presents a low-vaulted, heavily scarred, and contracted palate or a palate with large or multiple perforations (Fig. 39.8). Because of the surgical progress in the last 25 years, plastic surgeons today are not confronted with many failures in cleft palate surgery. Trained surgeons can now predict with greater accuracy the possible success of an operation, and are likely to avoid failure because other alternatives are available. Approximately 50 % of all cleft palate patients will need some type of prosthesis by the age of 30.

39.6 Contraindications for a Prosthesis

1. Surgical repair is feasible only when surgical closure of the cleft will produce anatomic and functional repair.
2. Patients with mental retardation are not good candidates for prostheses, because they frequently are not capable of giving the appliance the care it requires.
3. A speech aid is not recommended for an uncooperative patient or for a child with uncooperative parents.
4. If caries are rampant and not controlled, a prosthesis will require unusual care, and frequent examinations are important.

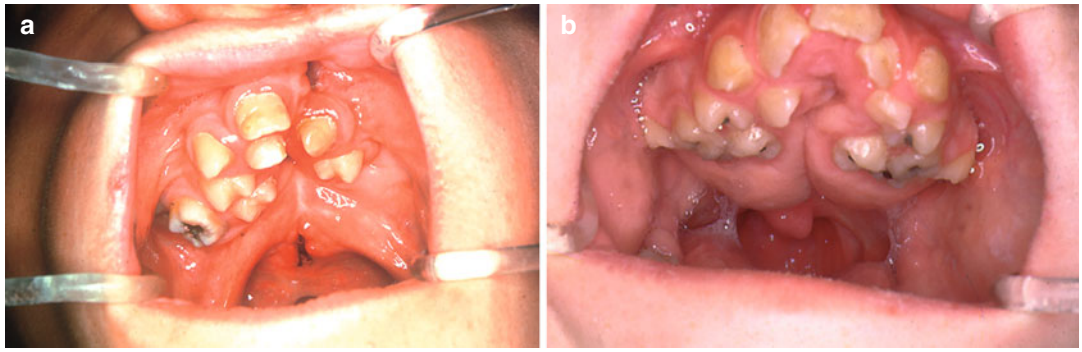

Fig. 39.8 (**a**, **b**) Two patients with heavily scarred palates and perforations: surgical failures

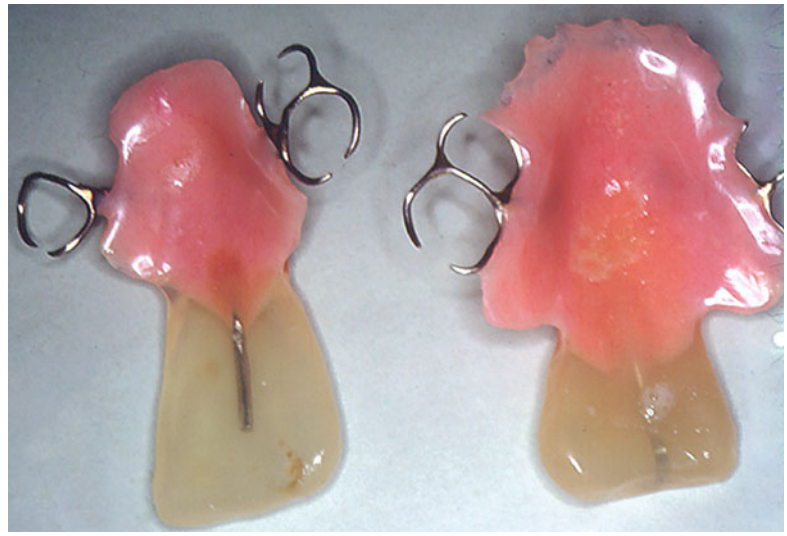

Fig. 39.9 A temporary acrylic resin speech appliance with wrought wire clasps and full palatal coverage designed for a 4-year-old child

5. The edentulous condition is not a contraindication for a speech-aid prosthesis.
6. Because the construction of a functional prosthesis requires the services of a dentist who has had training in cleft palate prosthodontics, it would be better to resort to surgical ingenuity when experienced prosthodontic help is not available.

39.7 Constructing Prosthetic Speech Appliances

For patients with deciduous, mixed, or permanent dentitions that are not fully erupted, all three sections of the prosthetic speech appliance are made of acrylic resin, and wrought wire retainers are used (Fig. 39.9). In patients whose permanent teeth are fully erupted, the anterior section of the prosthetic speech appliance should be made of cast metal or a combination of cast metal and acrylic resin (Fig. 39.10).

39.7.1 Preliminary Impression

A stock tray of adequate dimensions is selected. If a registration of the entire cleft is desirable, the stock tray is modified with modeling compound extending posteriorly to the postpharyngeal wall. The added section is underextended about 4–5 mm in all directions, leaving adequate space for impression material. Fast-setting, irreversible hydrocolloid is used for registering

Fig. 39.10 A permanent cast gold speech appliance with partial palatal coverage for an adult with no missing teeth

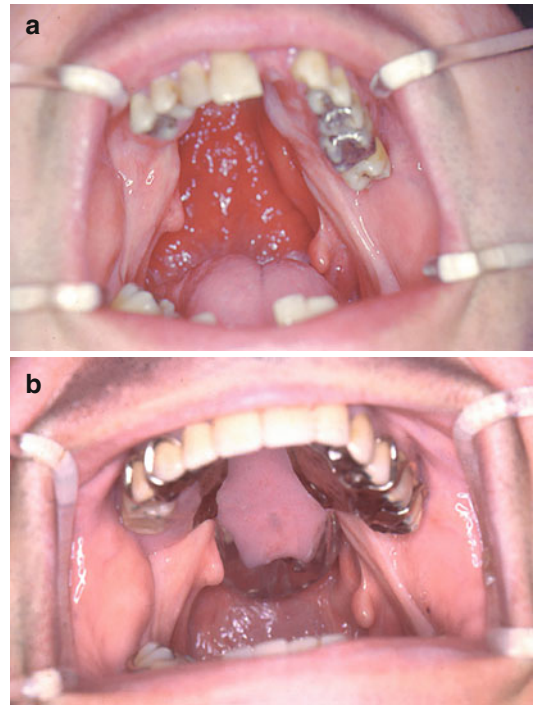

Fig. 39.11 (**a**, **b**) Crowning and splinting of the abutment teeth will increase the retention and support of the prosthesis and the life expectancy of the abutment teeth. (**a**) Patient before dental restoration. (**b**) After restoration with prosthesis in position

the preliminary impression. The following suggestions should be kept in mind when the preliminary impression is made:

1. If the patient is a child, he or she should be given the opportunity to examine the tray; in some cases, the child may be permitted to try the tray in his mouth. Children should be told that their cooperation is needed; otherwise, it will be necessary to make several impressions. Talking to children throughout the procedure is helpful.
2. The patient should have an early morning appointment.
3. The patient should have an empty stomach.
4. A topical anesthetic should be used on a child who has a severe gag reflex.
5. The tray should not be overloaded with impression material. Excess material in the nasopharynx will increase the difficulty of removing the impression without a fracture (see Fig. 39.14).
6. All oral perforations should be packed with gauze that has been saturated with petroleum jelly.

39.7.2 Preparation of the Deciduous Teeth for Retention

Most deciduous teeth do not have sufficient undercut for retention of the prosthesis. However, a small amount of bilateral undercut can give adequate retention. The following recommendations will help to produce adequate retention:

1. Carefully extend the clasp arms into interproximal areas of the teeth.
2. Insert, if necessary, serrated platinum pins into the buccal surface of deciduous molars to create an artificial undercut for the clasp.
3. Place bands with soldered retention lugs on the teeth.
4. Use chrome-cobalt crowns with retention lugs for teeth with extensive carious lesions or areas of decalcification.

After the clasp design has been determined on the diagnostic casts and the teeth have been prepared for retention, the final impression is made. If adequate retention is not available in the permanent dentition, crowning of the molars might be desirable to provide proper retentive areas (Figs. 39.11 and 39.12).

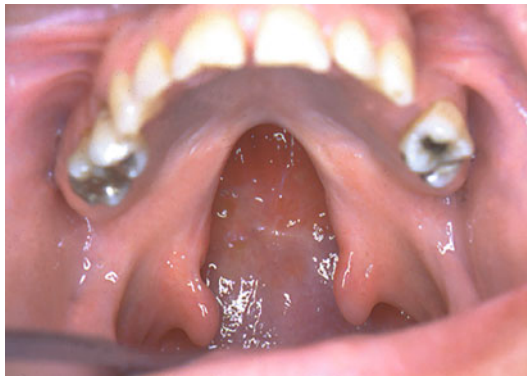

Fig. 39.12 Patient with wide cleft of the hard and soft palate treated with prosthetic speech appliance

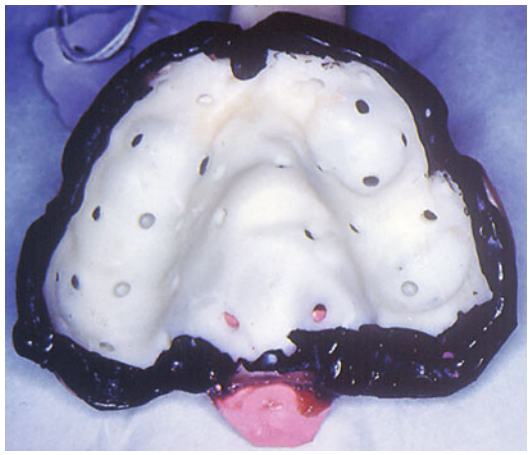

Fig. 39.13 An acrylic tray is made over the diagnostic cast and the border trimmed with green modeling compound

39.7.3 Final Impression

An acrylic resin tray is constructed over the diagnostic cast (Fig. 39.13). The patient is prepared in the same manner as for the preliminary impression, and the final impression is then made with an irreversible hydrocolloid impression material (Fig. 39.14). The master cast is made of dental stone.

39.7.4 Jaw Relation Records

Jaw relation records such as vertical dimension, centric relation, and protrusive relation are made and used in the adjustment of the articulator.

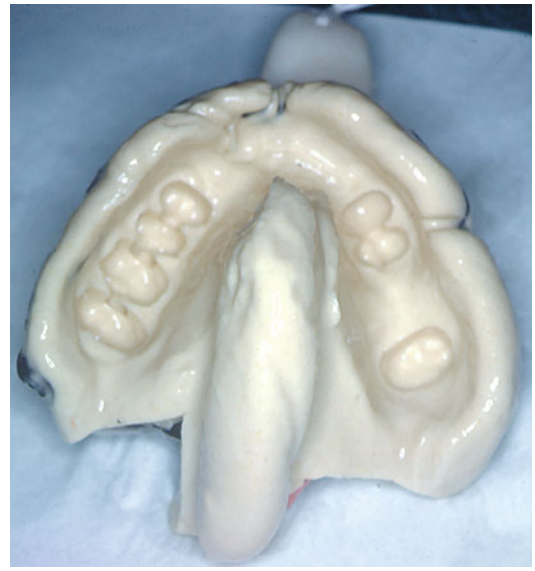

Fig. 39.14 The final impression is made with alginate material. Note the extent of the registration of the cleft

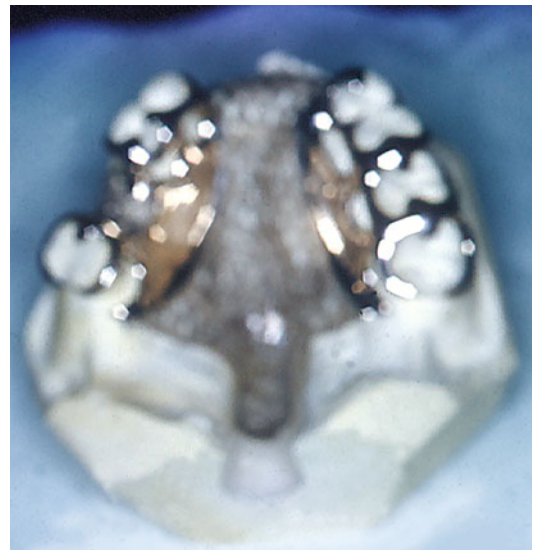

Fig. 39.15 Cast gold framework. The prosthetic speech appliance requires more retention and support; therefore, all the remaining maxillary teeth of this patient have been used for this purpose. The posterior extension of the framework reinforces the tailpiece and the speech bulb

39.8 Design and Construction of the Prosthesis

The master casts are surveyed and the prosthesis is designed (Fig. 39.15). For patients with severely constricted maxillary and mandibular

arches, teeth are arranged outside the remaining natural teeth to establish the proper esthetics and occlusion.

The prosthetic speech appliance is constructed in three sections. The design of the anterior portion is similar to that of a partial or complete denture. After this section is completed, the patient is instructed to wear it for at least 1 week. The length of this adjustment period depends on the ability of the patient to adapt to this part of the prosthesis. The construction of the middle part, the tailpiece, or velar section varies for operated and nonoperated clefts.

In unoperated clefts with the maxillary prosthesis in position, the extent of the tailpiece over the margin of the cleft is marked on the posterior part of the appliance. The tailpiece extends posteriorly to the anterior extent of the uvula.

In operated palates that are short and require a prosthesis, the position of the tailpiece is marked on the posterior margin of the prosthesis. The tailpiece extends approximately 3 mm behind the posterior margin of the soft palate. The width of the tailpiece is approximately 5 mm, and its reinforced thickness is about l.5 mm.

39.8.1 Construction of Velar Section

A piece of shellac baseplate material of the required width and length is used as a tray. It is securely attached to the posterior part of the prosthesis with about 2-mm relief. This assemblage is examined in the patient's mouth for proper extension. The tissue side of the tray is filled with zinc oxide and eugenol impression paste, and the appliance is inserted into the mouth. The patient is instructed to hold his or her head in a vertical position to prevent escape of the impression material into the nasopharynx. The head is held in this position for 1 min, then the patient is instructed to swallow a little water so that the muscular movement of the soft palate will be registered in the impression. After the material has hardened, the prosthesis is removed from the mouth, and the tailpiece is processed with self-curing acrylic resin. The denture portion with the finished tailpiece is placed in the mouth for testing. Swallowing of small amounts of water will stimulate muscle action along the lateral edge of the velar section. If the velar section is overextended laterally, undue muscle displacement and eventual tissue soreness will occur.

39.8.2 Construction of Pharyngeal Section or Speech Bulb

Two holes are drilled in the posterior part of the tailpiece. A piece of separating wire is drawn through the holes to form a loop that extends superoposteriorly beyond the superior part of the tailpiece. The ends of the wire are twisted together inferiorly (oral side) and secured to the appliance by sticky wax (Fig. 39.16). The wire loop that is extended into the nasal pharyngeal area is manipulated into an oval form, and the appliance is inserted into the mouth (Fig. 39.17). The patient is asked to swallow, and the wire is adjusted so that it will not contact the pharyngeal walls at any time. Posterior and lateral pharyngeal wall activity can be stimulated by spraying those tissues with water. The desired position of the wire is in the area of the maximum posterior and lateral pharyngeal constriction. Green modeling compound is added around the wire loop to reinforce it and its attachment to the tailpiece (Fig. 39.18). The appliance is inserted into the patient's mouth, and he is asked to swallow a little water. Adaptol, softened in water at 150–160 °F for 4–5 min, is added over the green compound, and the appliance is inserted into the mouth. Again the patient is instructed to swallow a little water to produce muscle activity, and thus, the impression material is molded (Fig. 39.19).

The prosthesis is reinserted a number of times, and the patient is instructed to swallow each time when additions of Adaptol are made to the mass on the wire loop. These steps are repeated until a functional impression of the lateral and posterior pharyngeal walls is made (Fig. 39.20). The impression material is then molded by instructing the patient to place his chin against his chest and move his head from side to side. In the rest position, he swallows water and talks to allow further molding of the impression material by muscular

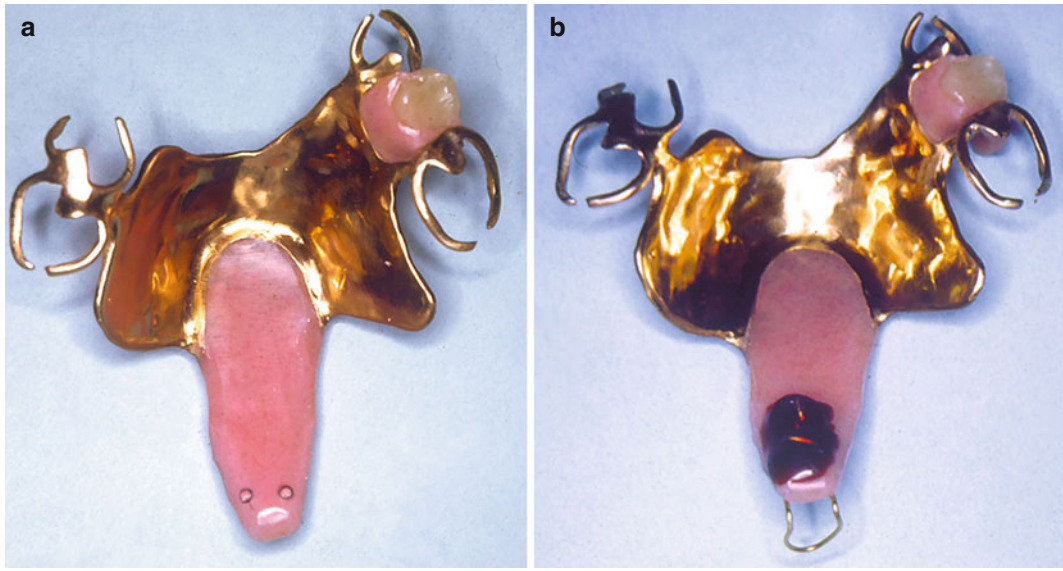

Fig. 39.16 (**a**) The location of the two holes drilled on the tailpiece. (**b**) View of the wire formed in a loop, extending superoposteriorly into the nasal pharynx

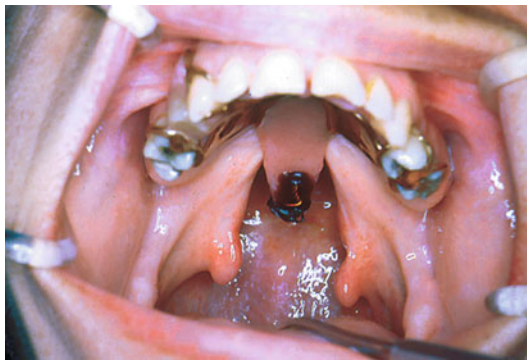

Fig. 39.17 Wire loop is attached to the tailpiece, inserted into the mouth of patient seen in Fig. 39.12, and checked to see that it does not contact posterior and lateral pharyngeal walls during swallowing

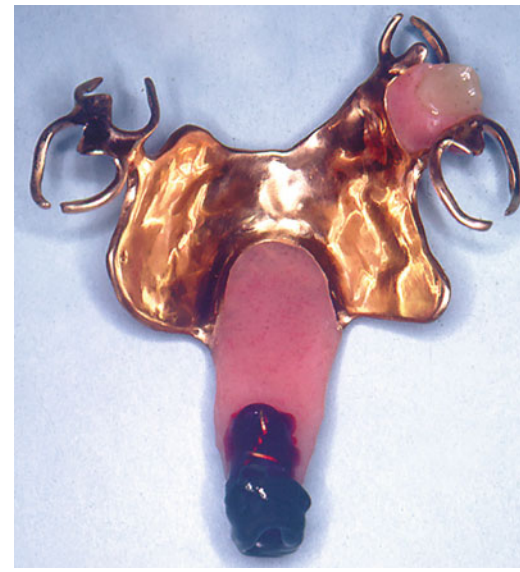

Fig. 39.18 Modeling compound is added around the wire loop to reinforce the wire and its attachment to the tailpiece

activity. If the mass is overextended, the patient will feel it during these actions. The overextended bulb impression is easily adjusted by reheating the bulb on the exterior surface and reinserting it into the patient's mouth. While the material is soft, the patient is instructed to produce the desired muscular activities. The completed speech bulb impression is chilled thoroughly in ice water. To check the position of the bulb, water is injected again, and the position of the bulb is examined in the mouth for its relation to the posterior and lateral pharyngeal wall activities. A spray of water onto the tissue will again stimulate these activities. In unoperated clefts, muscle function along the speech bulb during swallowing can be observed directly when the mouth is wide open and water is being injected onto the

tissues. When the posterior pharyngeal wall activity is not present, or direct visualization is not possible due to the length of the soft palate, a lateral cephalometric radiograph will reveal the position of the bulb in relation to the nasopharyngeal structures. In such cases, the bulb is placed in the area of the palatal plane. When the bulb form has been perfected, the bulb and tailpiece are processed onto the denture portion of the appliance. A heat-cured acrylic resin is used for making these parts.

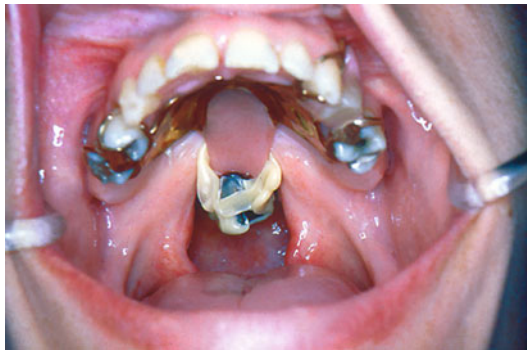

Fig. 39.19 Adaptol, softened by heating to 150–160 °F, is added over the green compound, and appliance is inserted into the mouth of the patient, Fig. 17.12. Note the displacement of the material after patient has swallowed some water and rotated the head to each side and down

For patients with unusually sensitive posterior and lateral pharyngeal walls (e.g., when the gag reflex is easily triggered), the making of a final impression for the speech bulb on the initial try is delayed until the patient is properly prepared for the impression. In such cases, it is helpful to construct an underextended bulb in self-curing acrylic resin, and to allow the patient to become adjusted to this small bulb for 2 or 3 weeks. After the patient has become accustomed to the undersized bulb, a final impression is made by adding Adaptol to the bulb, following the procedures previously outlined. The final impression of the speech bulb is processed in a heat-curing type of acrylic resin (Figs. 39.21 and 39.22).

To prevent the patient from swallowing the bulb in case the tailpiece is fractured, the appliance should be reinforced by incorporating a piece of No. 11 gauge half-round wire in the anterior body of the appliance and extending the wire into the bulb. If the anterior part of the appliance is made of cast metal, the frame should be extended posteriorly to strengthen the velar and pharyngeal section (see Fig. 39.15).

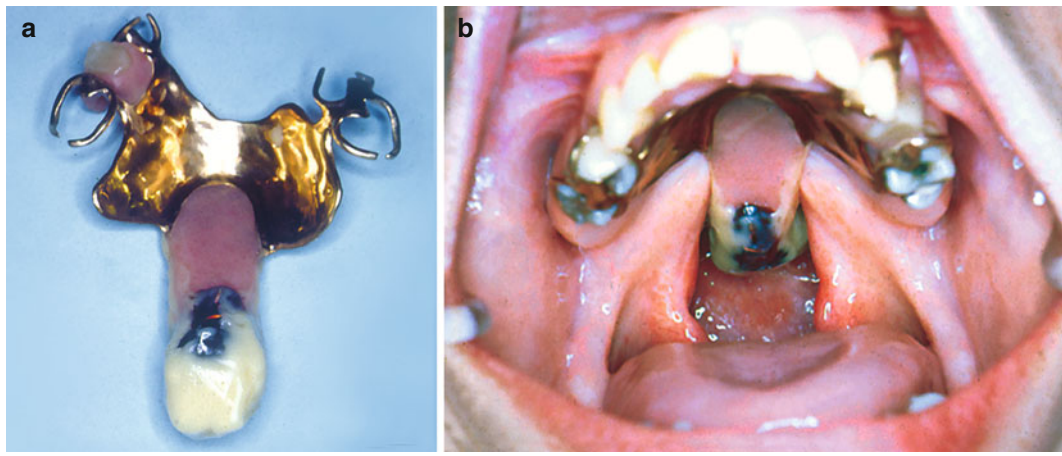

Fig. 39.20 (**a**) Functional registration of the velopharyngeal region using Adaptol. The gradual addition of Adaptol and patient swallowing water and moving the head down and to the sides will give the functional impression of the velopharyngeal region. If any gagging reflex is present, then underextended pharyngeal section is processed using an autopolymer. A week or two later, the pharyngeal section is modified for addition of the Adaptol. When the desired speech result is obtained and the patient does not show any gagging reflex, the speech bulb is heat-processed. (**b**) More Adaptol is gradually added, and the appliance is inserted until a functional impression of the area has been obtained. In most patients, the speech bulb does not contact the throat wall while the surrounding tissues are at rest

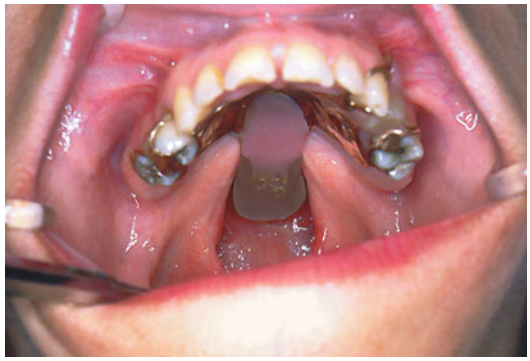

Fig. 39.21 Processed speech bulb in position, patient from Fig. 39.12

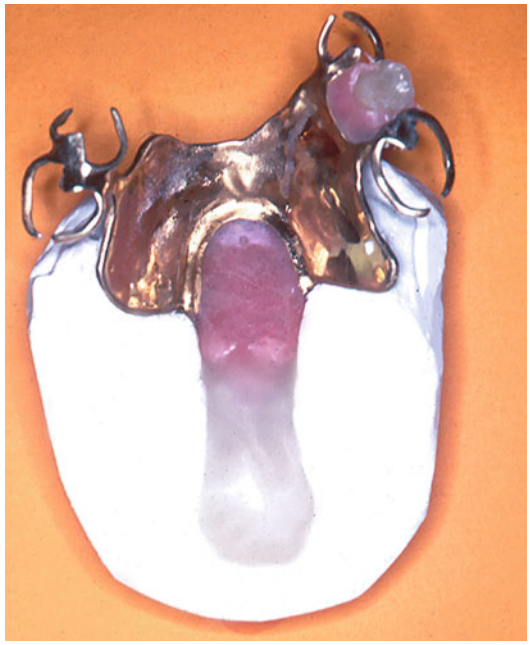

Fig. 39.22 The nasal and lateral sides of the speech bulb, tailpiece, and a portion of the palatal area of the anterior section are placed in dental stone. These parts of the appliance are made of acrylic resin

39.8.3 Insertion of the Appliance

The finished speech appliance is inserted into the mouth and examined for muscle adaptation to the speech bulb during swallowing and phonation, excessive pressure against the posterior and lateral walls of the pharynx, stability of the appliance during function, and improvement of the quality of the voice.

39.8.4 Position of Speech Bulb

For most patients, when the bulb is positioned too far inferiorly, the pharyngeal section has the following undesirable effects:

1. It has a tendency to be displaced by the dorsal part of the tongue during tongue movements.
2. It fails to relate to the normal region for making adequate velopharyngeal closure.
3. It has a detrimental acoustical effect on the quality of the voice. (Caution should be exercised to avoid blocking or extending the speech bulb into the eustachian tube.)

39.9 Summary

The prosthetic treatment of certain patients with cleft palate is an important part of the multidisciplinary approach to solving the many problems related to total health.

Some of the cleft palate patients for whom speech aids can be made include those with a wide cleft of the palate with a deficiency of the soft palate, a wide cleft of the hard palate with a high vomer, a neuromuscular deficit (a sphincteric velopharyngeal action may not be attained even with a pharyngoplasty if the deficit is marked), and surgical failures.

I strongly object to the use of remote extraoral flaps in cleft palate surgery because a prosthesis seems to be more appropriate. The possibility of cancer being related to such a prosthesis is quite remote, and there has been no evidence of increased hearing loss in patients wearing a prosthesis. A prosthesis should not be used in a patient not competent to care for it or maintain proper hygiene.

A prosthodontist engaged in treating patients with oral, facial, and speech deficits should be thoroughly familiar with the anatomic and physiologic deviations of the region involved and with

the basic principles involved in prosthetic dentistry. He should always be willing to acquire further knowledge in this field.

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40 Palatal Lift Prosthesis for the Treatment of Velopharyngeal Incompetency and Insufficiency

Mohammed Mazaheri

40.1 Treatment, Methodology, and Results in Patients with Velopharyngeal Inadequacy

Before getting into methodology of treatment of patients with velopharyngeal inadequacy, who require prosthetic velar elevation and velopharyngeal stimulation, let us outline the Lancaster Cleft Palate Clinic's present concept of treatment for patients with various types of velopharyngeal incompetency.

From 1984 to 1992, a total of 431 patients were referred to the Lancaster Cleft Palate Clinic with congenital or acquired velopharyngeal incompetency (VPI) (Table 40.1). This population consisted of 230 males and 201 females with a mean age of 11.26 years. Note the breakdown in the type of velopharyngeal incompetency. Two hundred seventy-one patients (63 %) demonstrated congenital velopharyngeal incompetency without submucous cleft; 86 (20 %) had VPI with a submucous cleft; 68 (16 %) had VPI related to trauma; and 6 (1 %) had VPI as a result of diseases such as myasthenia gravis, stroke, polio, and other neurological disorders.

Each patient was examined and evaluated by a plastic surgeon, prosthodontist, and speech-language pathologist with a combined experience of 110 years. A questionnaire was designed for data acquisition and long-term follow-up of these patients (Table 40.2).

Table 40.1 Number of patients and type of velopharyngeal incompetency (VPI)

	Number of patients
Male	230 (53 %)
Female	201 (47 %)
Type of velopharyngeal Incompetence	
No cleft	271 (63 %)
Submucous cleft	86 (20 %)
From trauma	68 (16 %)
From disease	6 (1 %)

40.1.1 The Referral

It is interesting to note that 256 patients (59 %) were referred by speech-language pathologists (Table 40.3), indicating that velopharyngeal incompetency is not recognized at an early age and that the diagnosis is frequently made when the patient starts school. The number of physician referrals was 96 (22 %). The remaining referrals (19 %) came from rehabilitation counselors, dentists, rehabilitation centers, and families.

Please note that 104 patients (25 %) had had their tonsils and adenoids removed in order to eliminate or remedy the velopharyngeal incompetency (Table 40.4). This, of course, causes an increase in hypernasality for the VPI patient.

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S. Berkowitz (ed.), *Cleft Lip and Palate*,
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Table 40.2 Questionnaire designed to record appropriate information on patients for the study

1. Patient number _____
2. Patient name _____
3. Patient address _____
4. Birth date _____ 5. Sex _____ Race: _____
6. Referral source _____
7. Chief complaint _____
8. Specific diagnosis:
   - Congenital VPI, no cleft _____
   - VPI with cleft _____
   - VPI with submucous cleft _____
   - VPI from trauma _____
   - VPI from cancer _____
   - VPI with other diseases _____
   - Iatrogenic VPI _____
   - Other unclassifiable _____
9. Diagnostic data available
   - Cephs _____
     - Lateral only _____
     - Lateral and AP _____
     - At ages _____
     - No cephs _____ Tracings
   - Cineradiographs
     - Yes _____ at ages _____
     - No _____
   - Recordings
     - Yes _____ at ages _____
     - No _____
   - Dental models
     - Yes _____ at ages _____
     - No _____
   - Growth analysis
     - Yes _____ No _____
   - Sonograms
     - Yes _____ No _____
     - Audiology examination
       - Yes _____ at ages _____
       - No _____
     - Surgical records
       - Yes _____ No _____
10. Other conditions (short narrative or diagnostic classification)
    - Dental health: _____
    - Orthodontic: _____
    - Audiology, otology: _____
    - Allergies: _____
    - Smoking habits:
      - No _____ Yes _____ Pack/day
    - Tonsils and adenoids removed:
      - Yes _____ age _____
      - No _____

Table 40.2 (*continued*)

11. When was VPI first noticed:
Age of onset _____
Circumstance _____
Who first noted VPI _____
12. VIP treatment history
Speech therapy _____ No. of sessions _____
Surgery (flap) _____ Type of flap _____
Surgeon _____ Hospital _____
Other surgery _____ Procedure _____
Surgeon _____ Hospital _____
Prosthesis _____ Type of prosthesis _____
Prosthodontist _____ Hospital/clinic _____
13. Sequence of treatment (if multiple procedures)
Speech therapy only _____
Flap and speech _____
Flap only _____
Lift and speech _____
Lift only _____
Speech and flap _____
Speech and lift _____
Flap and lift _____
Lift and flap _____
Three procedures sequence:
1. _____ 2. _____ 3. _____
14. Evaluation of result (speech)
Date of last follow-up _____
Acceptable _____
Not acceptable _____
Acceptable but can improve _____
Not acceptable but can improve _____
No improvement likely _____
Should recall patient _____
15. Recommendations
Today's date _____ Preparer's signature _____

Table 40.3 Referral source for VPI patients

Source	No. of referrals	Percent of sample (%)
Speech pathologist	256	59
Physician/surgeon	98	23
RN (nurse)	23	5
Dentist	12	3
PDH/BVR	23	5
Rehabilitation center	1	0.5
Social worker	2	0.5
Family	18	4

Table 40.4 Status of tonsils and adenoids of VPI pallechts

Status of adenoids	No. of cases	Percent of sample (%)
In	100	23
Out	104	24
In/out (to insert pharyngeal flap)	14	3
No information available	213	50

Table 40.5 Treatment methodology for patients with VPI

Treatment	No. of cases	Percentage of sample (%)
Speech only	126	29
Pharyngeal flap	122	28
Pharyngeal flap recommended	55	13
Palatal lift	74	17
Palatal lift removed after stimulation	8	2
Palatal lift recommended	15	3
Pharyngeal flap – palatal lift	16	4
Palata lift – pharyngeal flap	12	4

In addition to oral examination, nasal endoscopy, and individual judgment, all patients had two cephalometric radiographs taken, one with the soft palate at rest and the second during prolonged phonation of the vowel "E." Twenty-five percent of the subjects had cineradiographic studies of the velopharyngeal region to observe continuous phonation.

40.1.2 Results of Treatment

According to nasal endoscopic evaluation, radiographic analysis, and listener judgments, 126 (29 %) of the patients demonstrated inconsistent or borderline velopharyngeal dysfunction (Table 40.5). The team decided that each subject was to be referred to a speech-language pathologist with instructions to have a review by the team in 1 year if the condition persisted. Further evaluation of these patients after 1 year revealed that the hypernasality or nasal emission had subsided, and none required further treatment.

It was recommended that 177 patients (41 %) have pharyngeal flap surgery. In 122 patients (mean age, 10 years), the surgical procedure consisted of a superiorly based flap performed by our staff plastic surgeon. The remaining 55 subjects were referred to the plastic surgeon of their choice for a pharyngeal flap with instructions to return to the clinic after insertion of the flap for further evaluation.

Table 40.6 Status of patients who received palatal Lifts

Status	No. of patients
Palatal lift appliance removed for pharyngeal flap	3
Pharyngeal flap patients received appliances	13
Lift removed later	5
Combined appliance removed later	5
Still wearing palatal lift	5
Combined appliance still being worn	1

Table 40.7 Summary of use of prostheses and pharyngeal flaps

		Palatal lift		Combined appliance	
Pathology	*N*	In	Out	In	Out
Congenital					
PF-PL	9	4	5	0	0
PF-COMB	3	0	0	1	2
PL-PF	3	0	3	0	0
Trauma					
PF-PL	1	1	0	0	0
Totals					
Applianee in	6	5	0	1	0
Applianee out	10	0	8	0	2

Note: *PF* pharyngeal flap, *PL* palatal lift prosthesis, *COMB* combination prosthesis

Thirteen of the subjects with congenital VPI who were treated with a pharyngeal flap continued to exhibit a significant to moderate amount of hypernasal resonance and nasal emission (Tables 40.6 and 40.7). Palatal lifts or combination prostheses were constructed for these patients. Five of these patients had the palatal lifts removed, and two had their combination appliance removed after 1 year because the prostheses had resulted in their developing adequate posterior and lateral pharyngeal wall activity, and the patients were judged to have satisfactory voice quality without the appliances. Five of the patients with a lift and one with a combination appliance continued wearing their prostheses because of consistent nasal emission and lack of response to the prosthetic stimulation. One patient with VPI as a result of trauma who had pharyngeal flap surgery continued wearing his combination

Table 40.8 Summary of status of palatal lift and combination prostheses used for VPI of various etiologies

		Palatal lift		Coinhination	
Etiology	*N*	In position	Removed	In position	Removed
Congenital	61	13	21	23	4
Trauma	19	11	4	1	3
Disease	9	3	3	3	0
Totals	89	27+	28=(55)	27+	7=(34)

Note: Of the 35 appliances that were removed, 3 were removed due to patient rejection and 32 were removed after increased velopharyngeal function and satisfactory speech quality were achieved

prosthesis. The remaining patients with pharyngeal flaps were judged to have acceptable speech quality by the three team members. Further tests for nasal and oral pressure (cul-de-sac shifting, listening tube, nasal endoscopy, and oral manometer) substantiated the clinical finding.

Eighty-nine of the subjects were fitted with a palatal lift or combination prosthesis (Table 40.8). Sixty-one of the patients with congenital VPI (mean age, 11 years) had a palatal lift or combination appliance. At the time of this study, 13 of the patients with a palatal lift were still wearing their prostheses and 21 had gained adequate muscle activity so that the prostheses were removed. Twenty-three of the 61 patients were still wearing a combination lift, and 4 gained adequate tissue stimulation, so the prosthesis was discarded.

Of the 19 patients with traumatic VPI (mean age, 21 years), 11 had their prostheses still in position, 4 were removed, 1 had his combination in position, and 3 had rejected the combination prosthesis because of difficulty of adjustment, more difficulty swallowing, or lack of patient motivation and/or cooperation.

Of the 9 patients with VPI as a result of various neurological diseases, 3 have a palatal lift in position, 3 appliances have been removed, and 3 have a combination appliance still in position.

Fifteen additional patients were recommended for palatal lift prostheses, but the subjects or subjects' families elected not to have any form of treatment. Six-month follow-up revealed that the patients or the parents were satisfied with the patient's speech as it was. It was recommended to five patients that their palatal lift be removed in favor of a pharyngeal flap performed by a plastic surgeon in the patient's hometown. There was no follow-up at the Clinic for these patients after the insertion of the flap.

40.1.3 Summary

Analysis of the 35 patients whose appliances were removed revealed that 3 patients rejected the prosthesis within 6 months. Of the remaining 32 patients, 3 appliances were removed to insert a superiorly based pharyngeal flap, and 29 were removed when the patient demonstrated voice quality without the appliance that was judged to be satisfactory. Hypernasality was no longer a concern to these patients. The judges found this to be accurate.

In our population, use of the palatal lift or combination appliance for patients with traumatic VPI resulted in more acceptable speech performance than with velopharyngeal flap.

40.1.4 Conclusion

It is interesting to note that a majority of the patients referred to the Lancaster Cleft Palate Clinic for velopharyngeal incompetency were referred by speech-language pathologists. It was also interesting to note that a significant number of patients had had their tonsils and adenoids removed to remedy their hypernasality.

We have found that patients with a gap of more than 12 mm between the soft palate and posterior pharyngeal wall respond more favorably to physical therapy with a palatal lift or combination prosthesis prior to a pharyngeal flap than patients who have a pharyngeal flap as the initial mode of treatment.

Two of the subjects with complete paralysis of the soft palate as a result of traumatic injury had pharyngeal flaps performed by non-team member plastic surgeons; neither of these surgeries produced an acceptable speech quality result.

It is also interesting to note that a majority of the patients were diagnosed as having velopharyngeal incompetency after the age of 5. The studies show that the younger patients responded much more favorably to our treatment modalities (pharyngeal flap, palatal lift) than older patients. Therefore, it behooves us to diagnose cases at earlier ages and undertake the required treatment as early as possible.

40.2 Palatal Lift Prostheses for the Treatment of Patients Requiring Velar Elevation, Velopharyngeal Stimulation, and Velopharyngeal Obturation

40.2.1 Symptoms

Hypernasality or nasal emission and decreased speech intelligibility occur as a result of several organic conditions (e.g., congenital or acquired cleft of the palate, congenital short soft palate or palatal paresis or velopharyngeal insufficiency, velar paralysis or velopharyngeal incompetency, abnormal nasal pharyngeal size, and hypernasality occurring after the removal of the tonsils and adenoids).

40.2.2 Etiology

The etiological factors contributing to the development of these organic conditions can be classified into two major categories:

1. Prenatal
   (a) Cleft of the palate
   (b) Short soft palate
   (c) Abnormal nasal pharyngeal size
   (d) Abnormal velopharyngeal neuromuscular development
2. Postnatal
   - Partial or completely paralyzed velum as a result of central or peripheral nervous system damage (e.g., a patient with myasthenia gravis, bulbar polio, traumatic brain injuries, cerebral vascular accidents, degenerative central nervous system diseases, and amyotrophic lateral sclerosis).

40.2.3 Speech Characteristics

Speech characteristics common in both types of patients with velopharyngeal incompetency and velopharyngeal insufficiency are:

1. Hypernasality
2. Nasal emission
3. Decreased intelligibility of speech due to weak consonant production

The patient with velopharyngeal insufficiency often develops glottal stop substitution as a result of compensation for production of pressure consonants. The patient with neurological diseases resulting in a full or partial paralysis of lips, tongue, larynx, or respiratory musculatures often develops an abnormal articulatory pattern and diminution of breath pressure, which causes a reduction of oral pressure and flow.

40.2.4 Methods of Treatment

The closure and obturation of palatal clefts and defects for patients with congenital and acquired clefts have been reported. Early humans used stone, wood, gum, cotton, and other foreign bodies to obturate the palatal opening. In recent years, several methods have been advocated for satisfying the main objective of socially acceptable speech for these patients. Among these concepts are:

1. Traditional speech treatment, such as active lip, tongue, and palate exercises for the stimulation and physical therapy of musculatures (myofunctional therapy), designed to effect reduction in hypernasality.
2. Surgical methods designed to reduce the velopharyngeal gap or lumen, employing velar lengthening procedures, velopharyngeal flaps, implants (cartilage, bone, silicone, Teflon®), and combinations of several methods.

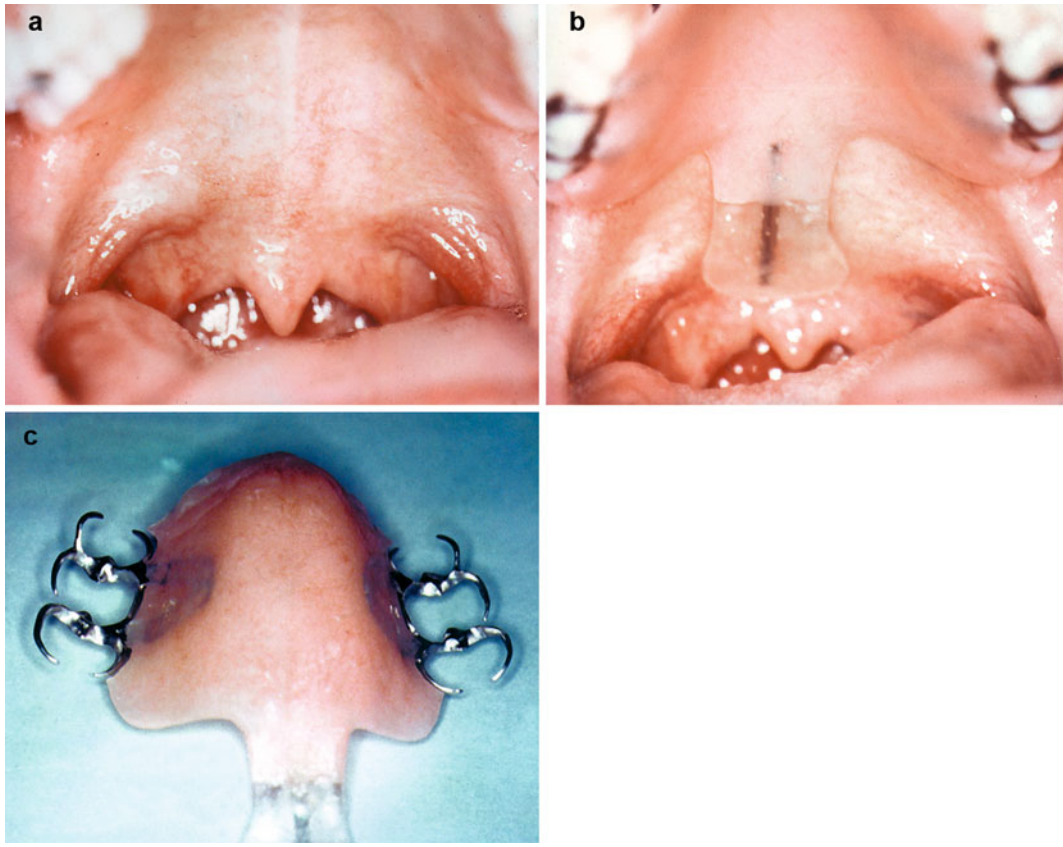

Fig. 40.1 (**a**) Patient with palatopharyngeal insufficiency. The treatment procedure is the stimulation of the soft palate by a palatal lift prosthesis followed by pharyngeal flap surgery. (**b**) View of palatal lift prosthesis in position. (**c**) Palatal view of the lift prosthesis

3. Faradization and electrical vibration massage to stimulate palatal function.
4. Prosthetics designed to elevate and stimulate the soft palate in patients with velopharyngeal incompetency or to elevate, stimulate, and obturate the velopharyngeal lumen in patients with velopharyngeal insufficiency.

As previously stated, two prosthodontic procedures are available to us in the treatment of patients with velopharyngeal inadequacies:

1. Lift type
2. Combination of lift and bulb

The lift type of prosthesis is used to elevate the soft palate to the maximum position attained during normal speech and deglutition. The reduction in size of the velopharyngeal gap and lumen will decrease nasal air flow, increase oral pressure for consonant articulation, and improve voice quality. The lift may also act as a physical modality for stimulation of velar and pharyngeal musculatures and elimination of the occurrence of velar disuse atrophy (Figs. 40.1, 40.2, 40.3, 40.4, 40.5, and 40.6).

The combined lift/bulb prosthesis should be the method of choice when the soft palate is insufficient for the proper velopharyngeal closure. The combined lift/bulb prosthesis is used to elevate the soft palate, obturate the gap, and stimulate velopharyngeal development and pharyngeal constriction (Figs. 40.7 and 40.8).

40.2.5 Prerequisites of Lift and Combination Prostheses

1. The maxillary portion of the prosthesis is designed to achieve optimal retention and stability.

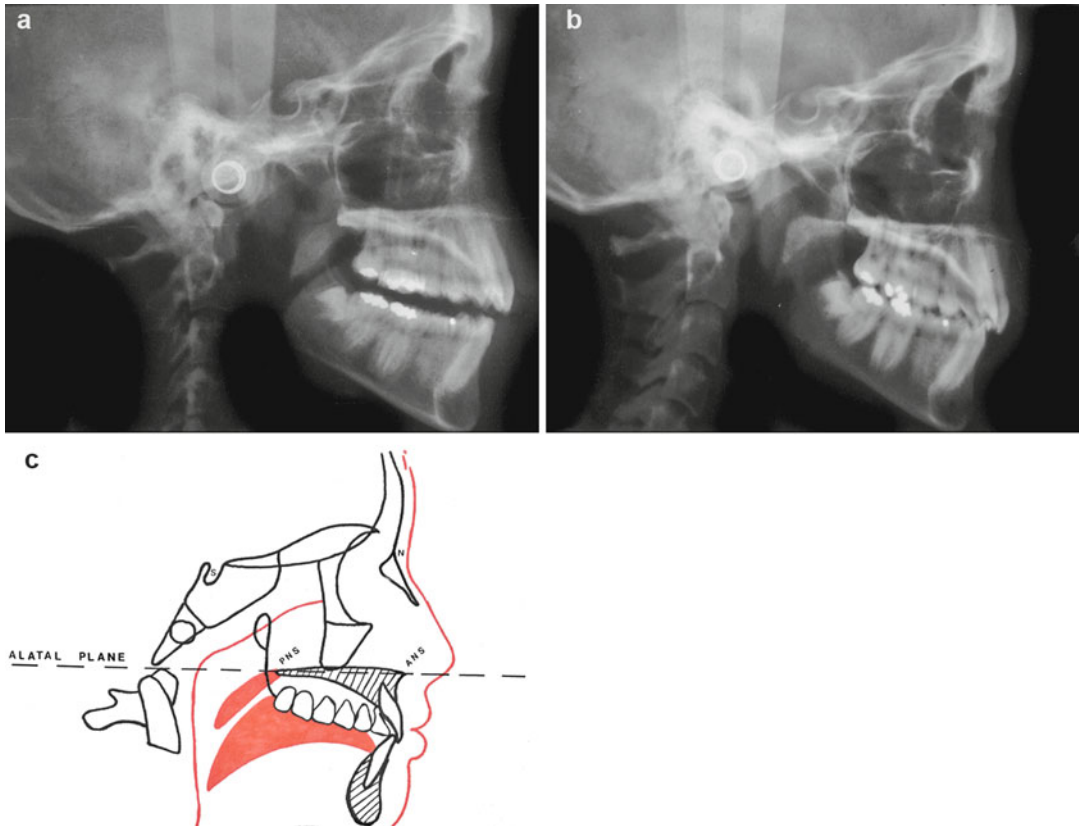

Fig. 40.2 (**a**) Lateral radiograph of patient in Fig. 18.1 demonstrates the palatopharyngeal relationship prior to elevation and stimulation. (**b**) Height of velar elevation during the sound "E." (**c**) Tracing of the cephalogram in **a**

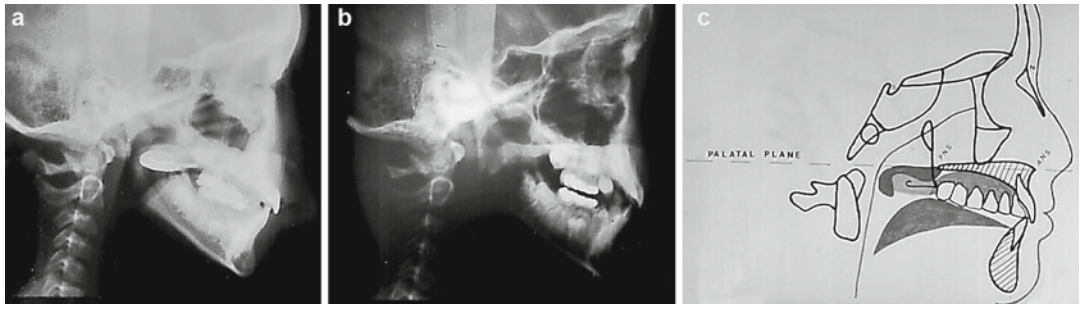

Fig. 40.3 (**a**) Radiographic view of the palatal lift prosthesis of patient in Fig. 18.2 in position. Note the degree of palatal elevation. (**b**) Increased mobility of the soft palate after 1 year of prosthetic stimulation. Pharyngeal flap surgery was done after 14 months of soft palatal stimulation, after which the lift prosthesis could be discarded. (**c**) Cephalometric tracing of the palatal lift prosthesis and the degree of velar elevation accomplished by the lift

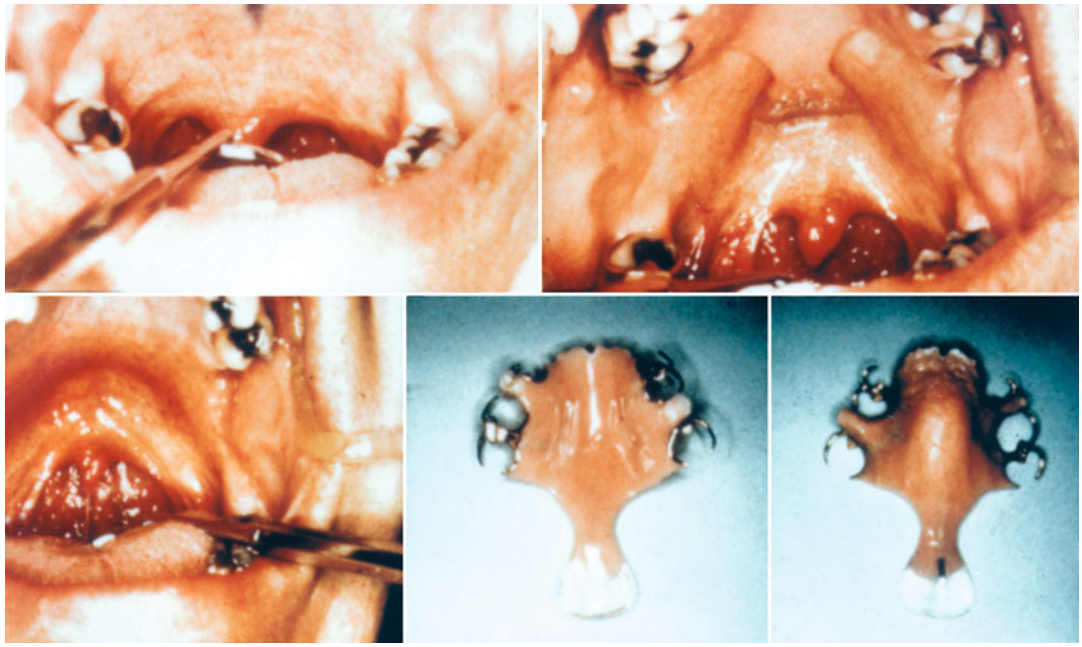

Fig. 40.4 *Top left*: Patient with palatopharyngeal incompetency in which the soft palate is paralyzed as a result of neurologic involvement after an accidental head injury. *Top right*: Palatal lift in position. *Bottom left*: Increased soft palate elevation after 6 months of prosthetic velar stimulation. *Bottom right*: Oral and palatal view of the lift prosthesis

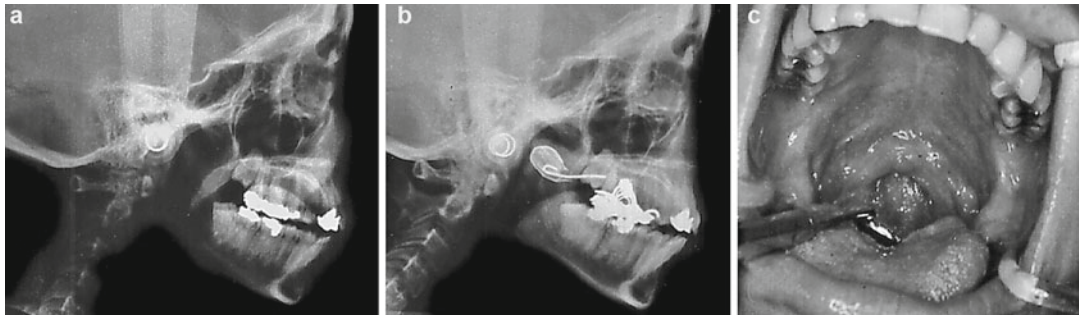

Fig. 40.5 (**a**) Lateral radiograph of the patient in Fig. 18.4 prior to stimulation saying "E." (**b**) The palatal lift prosthesis in position elevating the soft palate. (**c**) Note the increase in the degree of palatal elevation. After 11 months of stimulation and speech therapy patient is saying "E." Note the substantial increase in the velar elevation

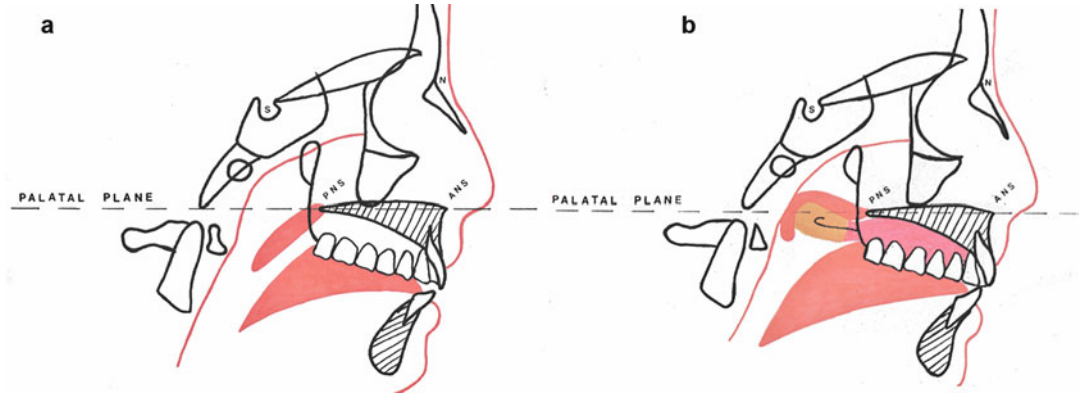

Fig. 40.6 (**a**) Tracing of a lateral cephalogram of the patient in Fig. 18.5 prior to soft palate stimulation by a palatal lift prosthesis. (**b**) Tracing of the palatal lift prosthesis and elevated soft palate

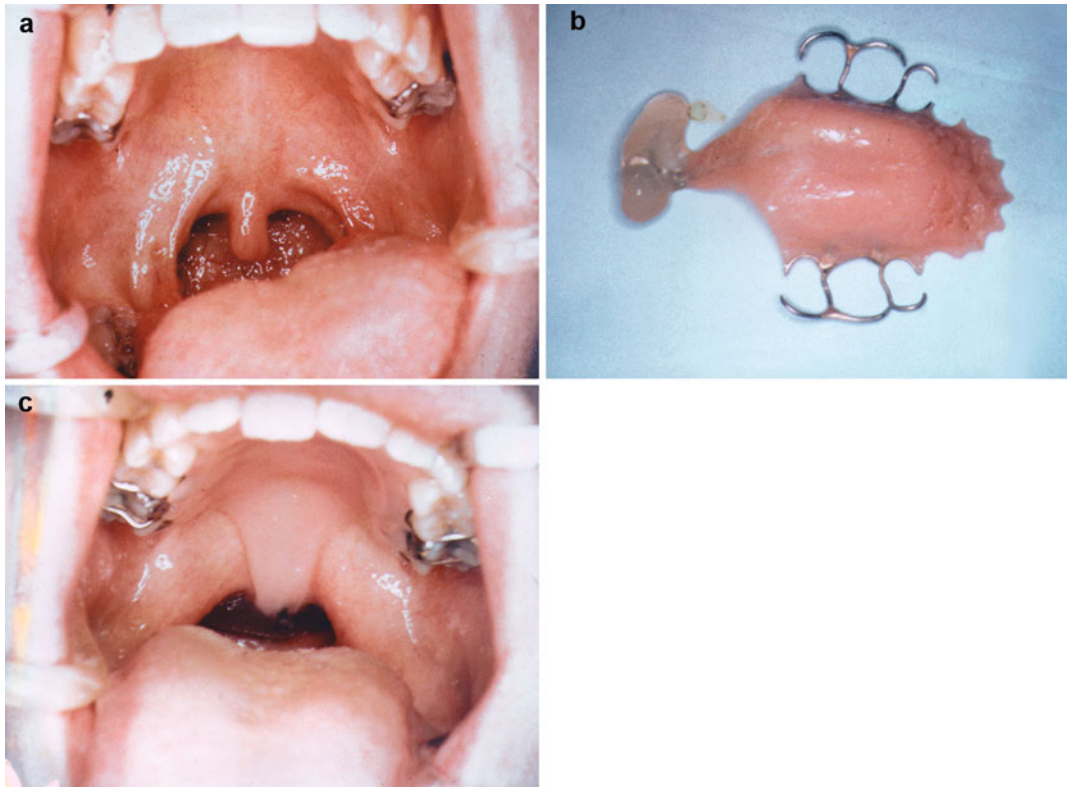

Fig. 40.7 (**a**) Patient with a palatopharyngeal insufficiency in which the soft palate is short and has limited mobility. (**b**) Combination palatal lift pharyngeal section in position. The uvula was displaced by the prosthesis without causing any irritation. (**c**) Palatal view of the prosthesis

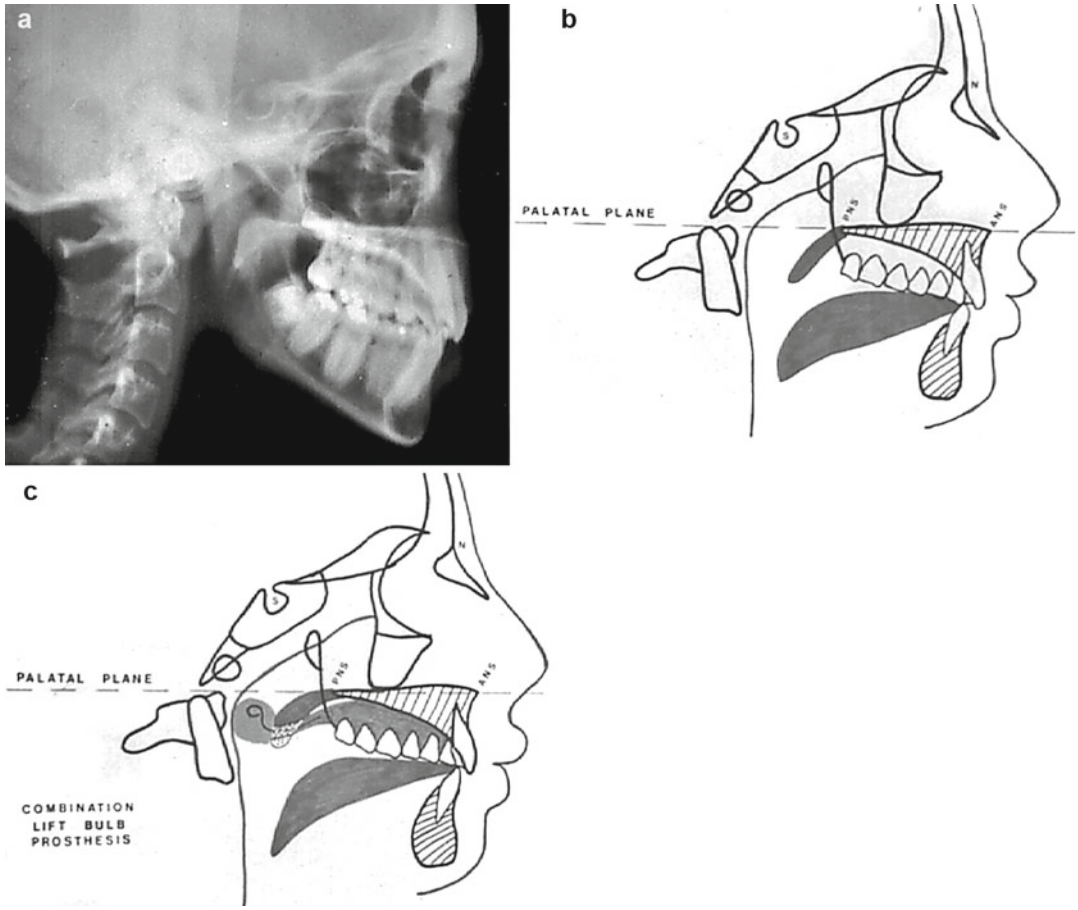

Fig. 40.8 (**a**) Lateral radiograph demonstrating short soft palate and large nasopharynx. (**b**) Tracing of the lateral cephalogram of the patient in Fig. 18.7. (**c**) Tracing of the combined palatal lift/pharyngeal section prosthesis in position

2. The lift portion should be placed so that velar elevation occurs in the area where normal velopharyngeal closure takes place.
3. Elevation of the velum should be gradual so that the velum becomes less resistant to displacement.
4. The pharyngeal section should be placed in the area where posterior and lateral pharyngeal constriction takes place so that it increases the change of further stimulation and muscle activation.
5. The reduction of pharyngeal section, when indicated, should be gradual.
6. Speech therapy, such as active lip, tongue, and palatal exercises and placement, should be properly instituted in conjunction with the construction and insertion of the prosthesis.

40.2.6 Objectives in Making Prosthetic Lift and Combination Services

1. Reduce hypernasality and nasal air escape by velar elevation
2. Reduce the degree of disuse atrophy
3. Increase velopharyngeal function by constant and continuous stimulation
4. Increase neuromuscular response by gentle stimulation and speech exercises

40.2.6.1 Results of Using Lift and Combination Prostheses

Methods of Evaluation

1. Speech testing procedures
2. Nasal endoscopy

3. Radiographic evaluation (e.g., cineradiography, cephalometrics, sectional laminography, or tomography)
4. Oral nasal air pressure and air flow assessing devices
5. Electronic instrumentation such as Tonar and sonograph

The optimal result depends on the type of oral pharyngeal involvement. If the neurological disorder is more localized to the velopharyngeal region, and the patient has few or no speech articulatory disorders, the prosthetic result is optimal. Patients with muscle paralysis of the tongue, lips, larynx, and respiratory organs usually respond less favorably to prosthetic care. Their phonatory and articulatory disorders usually remain after the prosthetic treatment. These patients often require more intensive and coordinated myofunctional therapy.

Patients' tolerance and acceptance of prosthetic treatment vary. Some patients have less difficulty than others, becoming accustomed to the palatal and velopharyngeal coverage and decreased oral pharyngeal space and volume.

We have also noted variations in muscle response to mechanical stimulation. The velum of the same patient, shortly after placement of the lift, becomes more active, and after 6 months to 1 year, prosthetic stimulation and support can be discarded. Whether the increased velar elevation is the result of prosthetic stimulation or neuromusculature recovery is difficult to assess. However, we can state that, in our experience, similar patients who received speech therapy as the only mode of velopharyngeal stimulation demonstrated less functional recovery over the same period of time than patients where the prostheses were employed (see Figs. 40.6 and 40.7).

In our series of patients, we have found more marked nasal pharyngeal than velar musculature response to the prosthetic stimulation. With the velopharyngeal bulb, the patient often develops compensatory muscular constriction, requiring frequent reduction in the size of the pharyngeal bulb. In some patients, complete elimination of the bulb was accomplished. We could safely state that the reason for the variation in the degree of response observed in patients with velar incompetency and patients with velopharyngeal insufficiency is that we have two separate phenomena to consider. For one patient, we are trying to stimulate muscle activity by prosthetic physical therapy; for the other patient, we are attempting to create muscle build-up or constriction as a result of prosthetic placement.

40.3 Summary

1. Velar elevation should be gradual in order to put less pressure on the teeth retaining the prosthesis and to reduce the possibility of mucosal irritation.
2. Prosthetic stimulation should be initiated as soon as velar paralysis is noted, to reduce the occurrence of velar disuse atrophy.
3. The palatal lift prosthesis is used as a temporary or permanent measure for the correction of velar incompetency. As soon as adequate elevation occurs, the prosthesis is discarded. Otherwise, the patient could wear the prosthesis as a permanent supportive device.
4. Construction of the combination lift/bulb prosthesis requires a program of gradual velar elevation and molding of the pharyngeal bulb to reduce the gag reflexes and increase velopharyngeal adaptation to the prosthesis. After initial placement, modification of the velopharyngeal section becomes less troublesome to the patient.
5. Speech and myofunctional therapy should be instituted in conjunction with the prosthetic treatment.
6. Prosthetic lift and combination prostheses are more effective for patients with less severe neurological impairment and speech articulatory errors.
7. The prosthetic lift has been more effective for patients with velar incompetency without involvement of other oral pharyngeal musculatures, whereas the combination type has been more effective for patients with velopharyngeal insufficiency without marked speech articulatory disorders.

Several questions require further investigation.

1. What is the relationship between the palatal stimulation and degree of neuromuscular function and recovery?

2. What is the relationship between stimulation and degree of occurrence of disuse atrophy?
3. What is the relationship between pharyngeal stimulation and muscle constriction?
4. What is the degree of stability of velopharyngeal function and constriction after stimulation?

Part XIV

Third World Cleft Treatment

Cleft Palate Treatment in Developing Countries of Africa

41

Fadekemi O. Oginni and Wasiu L. Adeyemo

Abbreviations

Cleft palate may occur in isolation (CP) or in conjunction with cleft lip (CLP) and sometimes as a component of a syndrome.

OFC	Orofacial cleft
PSO	Pre-surgical orthopaedics
CL	Cleft lip
CP	Isolated cleft palate
CLP	Cleft lip and palate
sP	Soft palate
hP	Hard palate
Ant	Anterior
IVV	Intravelar veloplasty
MCC	Maternal care coordination
ANC	Antenatal care
ORL	Otorhinolaryngology

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41.1 Introduction

Orofacial clefts (OFCs), which include cleft lip (CL), cleft palate (CP) and cleft lip with cleft palate (CLP), can impair the development of teeth, speech, hearing and feeding capability in isolation or combination. These defects create physical and emotional stress for infants, children and their families. At other times, there are multi-system and complex anatomical, physiological and psychosocial problems which are taken into consideration in optimal care delivery (Sitversen et al. 2008). For many reasons, the protocol of care for these patients has attracted lingering debates worldwide (Malcolm and Richard 2000). Although the timely receipt of primary cleft surgery is essential to the medical and psychosocial well-being of these patients (Bokhout et al. 1997; Strauss 1999; Riski 2002; Benedict and Farel 2003), services and treatment delivery can vary, depending on severity of the defect, presence of associated syndromes or other birth defect(s) and the child's age and rate of development (Nackashi et al. 2002).

Despite these variations, some general helpful recommendations exist (Nackashi et al. 2002; Lynch and Karnell 2003).

The initial evaluation of an infant with OFC is recommended within the first few days of life, and subsequent evaluations should be scheduled at regular intervals, depending on cleft severity, presence of associated deformities and the child's age.

The ideal timing and method of CLP repair remain an enigma, despite many studies, surveys and reports in the literature (Agrawal and Panda 2011;

S. Berkowitz (ed.), *Cleft Lip and Palate*,
DOI 10.1007/978-3-642-30770-6_41,

Shaw et al. 2001). It varies from place to place and with different patient types.

Basically, the controversy is focused on early palatoplasty for improved speech development versus delayed palatal closure with minimal disturbance to facial growth (Ysunza et al. 2010). Some investigators believe that early hard palate closure is advisable (Semb 1991; Noverraz et al. 1993), whereas others believe that delaying hard palate closure would result in a more favourable growth of the maxilla (Hotz and Gnoinski 1976, 1979; Freide et al. 1987).

For children with OFC, *timely primary cleft surgery* was defined as surgery within 18 months of life. For infants with CL, it was defined as surgery within the first 6 months of life, and for infants with CP with or without CL, surgery within 18 months of life (American Cleft Palate-Craniofacial Association 1993). Timeliness in surgery is a product of multiple factors which appear to differ from place to place and from time to time. Different centres and practitioners tend to adopt and advocate protocols that suit their prevailing circumstance(s).

The situation in the developing world could be tagged peculiar for a number of reasons. These include financial constraints, limited facilities and personnel, low level of awareness about cleft care and poor health-seeking behaviours.

Others factors affecting timeliness in surgery are:

Maternal care coordination (*MCC*)/*Antenatal care* (*ANC*). Children whose mothers received MCC/ANC are more likely to get informed about availability and places where cleft surgery are available than those children whose mothers received no such care or counsel (Cassel et al. 2009).

Residence. Those living in the urban or semi-urban locations are more likely to receive timely cleft surgery than children living in rural settings (Cassel et al. 2009).

Racial/ethnic factors. Multiracial studies have found differences in the access to care indicating racial/ethnic and geographic differences in the receipt of timely primary cleft surgery among children with OFC. Lack of access to care was noted especially among Black/non-Hispanic and Hispanic individuals (Ronsaville and Hakim 2000; Fiscella et al. 2002; Buescher et al. 2003; National Institute for Health Care Management Foundation 2007).

Available services. Differences in the types of services and treatment offered by the various craniofacial centres and teams can also affect the timeliness of services (Strauss 2002). Some craniofacial teams provide evaluation and quality assurance only, while craniofacial centres usually provide more direct services and treatment. Craniofacial centres and teams vary in their capacity to treat patients with clefts and or other craniofacial conditions (Strauss 2002; American Cleft Palate-Craniofacial Association, Cleft Palate Foundation 2005–2006).

Parental perception. Parental perception of need and cleft severity may delay surgery. Cleft lip defects are readily apparent at birth, and parents usually understand the need for surgical intervention. Isolated cleft palate may however go unnoticed for sometime and untreated for a relatively longer time even in the presence of speech impairment.

Income group. Children born into higher income (Figs. 41.2, 41.3, 41.4 and 41.5) group homes have a greater tendency to receive timely care (Porterfield and McBride 2007). Coincidentally, this group has ready access to ANC and the associated strong referral network which assist them in accessing resources. Furthermore, this link ensures delivery of appropriate services and continuity of care.

Children with OFC who benefited from referral to services (especially to craniofacial centres and teams) were significantly associated with areas of residence, cleft type, presence of other birth defects, presence of other malformations in the family and receipt of MCC services in a series of studies (White 1981; Williams and Sandy 2003; Cassell et al. 2007).

Untreated cases however have been associated with some rare forms of clefts that are comparably minor (e.g. bifid uvula or submucous CP) for which surgery may never be needed (Gosain et al. 1999; Sperber 2002).

Other factors, such as *financial and non-financial barriers*, can impede receipt of timely cleft surgery among this population. Financial barriers are widely present in most developing

countries where majority of patients cannot afford to pay for treatment. Non-financial barriers include health policies, service organizations and unavailability of effective referral systems (i.e. craniofacial centres and other specialized services and care providers) (Rosenbach et al. 1999; Newacheck et al. 2000; Betz et al. 2004).

Personal barriers, such as ignorance about available supports and the extent of support as well as inability to navigate the healthcare system effectively (St Clair et al. 1990) and religious and cultural beliefs (Oginni et al. 2010), are real.

41.2 Considerations for Age in Palatoplasty

The primary goal in timing of CP surgery is to provide adequate palatal function for the development of normal speech without interfering significantly with the maxillary growth. Soft palate surgery aims at velopharyngeal competence, while normal facial growth and dentition are the goals of hard palate repair.

Although a controversial subject, early closure of the lip and palate is believed to impart greatly to early speech function and learning, whereas delayed closure favours growth of the maxilla (Rohrich et al. 2000).

The age at which palatal closure is performed is usually the surgeon's choice and a compromise between the need for normal growth and normal speech. Arguments for and against the delayed hard palate closure often revolve around the "trade-off" between the inhibitory effects of early palate repair on growth versus the increased burden of additional treatment, which may be necessary as a result of delaying palate repair (Robertson 1986; Peterson-Falzone 1996).

In the early days, closure of the hard palate after eruption of the secondary dentition was considered a prerequisite for undisturbed maxillary growth (Hagemann 1941; Schweckendick 1951). Later, the focus of treatment shifted toward consideration of speech development.

Early repair of the soft palate was recommended to prevent speech disturbances in patients with delayed hard palate closure. The philosophy of repairing the soft palate for speech requirements but delaying the hard palate repair to reduce growth inhibition was introduced by Gillies and Kesley Fry (1921). This was later introduced into clinical practice by Schweckendiek who delayed palate repair into the adolescent years (Schweckendick and Doz 1978). This method has since undergone further modifications by Robertson and Jolleys (1968), Hotz et al. (1978) and Malek et al. (1986).

Studies on growth of the facial skeleton have identified the second postnatal year as the period of active growth and suggested that palatal surgery performed at the age of 1 year might disturb the growth of the maxilla more than those performed at about 2 years. Similarly, the Oxford cleft palate study and others (Rohrich and Byrd 1990; Rohrich et al. 1996, 2000; Noverraz et al. 1993) suggest that we may consider hard palate surgeries done in children under 12 months as early and those done at or beyond 48 months late.

Although, varying degrees of intrinsic maxillary growth deficiencies are common findings in the facial skeleton of individuals with clefts (Yoshida et al. 1992; Berkowitz et al. 2005). The preoperative morphology in the cleft child is said to explain half of the occlusal morphologies and variations in occlusal development and maxillary inter-canine width (Friede et al. 1988). The inherent growth impairment observed may be exacerbated by surgical technique, surgeon's skill (Ross 1987b) and timing of surgery.

Proponents of neonatal cleft surgery (Hodgkinson et al. 2005) believe that with it, healing is better, parents obtain some psychological relief and the child has minimal cognitive development effect (McHeik and Levard 2006; Galineir et al. 2008; Murray et al. 2008). On the other hand, it is plagued with greater risks (Hodgkinson et al. 2005), more so for the palate.

Considering that early palatal repair (prior to 1 year of age) is desirable for excellent speech results (Ross 1987c), this protocol was embraced by some centres. A few of them performed simultaneous lip and hard palate repair at the age of 4–5 months (Ross 1987c), only to be abandoned soon due to the associated poor growth and restricted midfacial development (Friede and Lilja 1994; Melissaratou and Friede 2002).

Early closure in this series attained the desired good speech, at the expense of facial growth (Freide et al. 1987; Lohmander-Agerskov et al. 1996; Lohmander-Agerskov 1998; Friede and Enemark 2001).

Similarly, Lilja et al. (2006) reported outcome of the Goteborg protocol (based on early soft palate closure for adequate speech development and later hard palate closure to limit maxillary growth impairment). This protocol facilitates closure of the residual defect without leaving any raw palatal bony surfaces open to secondary epithelialization. Outcome based on the GOSLON'S code revealed excellent and good growth in 85 % of patients, adequate growth in 12 %, poor growth in only 3 % and very poor growth in no patient. Comparable growth outcomes have been published by several authors (Hotz and Gnoinski 1979; Freide et al. 1987, 1980; Friede and Enemark 2001; Noverraz et al. 1993; Gnoinski and Haubensak 1997; Schweckendick and Doz 1978; Jorgenson et al. 1984; Wada et al. 1984; and Mylin and Hagerty 1983), some of whom attributed this to postponement of interference with maxillary growth to a later age when less growth remains.

Exploring the extreme of older age groups, Ross (1987e) and Bludorp and Egyedi (1984) showed that closure of the palate within the first decade and 3–6 years of life, respectively, makes little or no difference in maxillomandibular growth and development, thus eliminating the benefits of unduly prolonged delay in palate repair.

Several studies have shown acceptable speech development following delayed hard palate closure (Lohmander-Agerskov and Soderpalm 1993). Lohmander-Agerskov (1998) and Lohmander-Agerskov and Willadsen (1999) concluded that speech results obtained after delayed hard palate closure were comparable with those achieved after conventional surgical rehabilitation. However, in some patients, speech development did not normalize until the cleft in the hard palate had been closed.

Much as timing is an important component of surgery outcome, the experience and skills of the attending surgeon in tissue management may have as much or more influence on the craniofacial development than the alleged growth-inhibiting effects of a surgical technique or the timing of hard palate closure (Lehner et al. 2003).

Schweckendick and Doz (1978) attributed speech failure to poor technique/skill, i.e. lack of reorientation of velar muscles and no dissection of the mucoperiosteal flaps in the posterior edge of the hard palate. It is advocated that proper reorientation of velar muscles is best achieved beyond the age of 10–12 months when identification and adaptation of the muscles is more practicable and reliable (Lehner et al. 2003).

The resultant midfacial hypoplasia attending early repairs has been related to early periosteal undermining of palatal tissue. Ross (1987a, b, c, d, e) demonstrated an overall detrimental effect of surgery on facial growth in his longitudinal analysis of cephalometrograms of treated patients with cleft lip and palate. The deforming effects of surgically denuded palatal shelves are shown in several animal studies as well humans (Bardach and Kelly 1990; Markus et al. 1993).

In contrast, Delaire et al. (1988) and Joos (1987) (advocates of an anatomical reconstruction of both the lip muscles and the perinasal midface muscles for their growth-inducing effect on the midface) could not find significant adverse effects of surgery on midface development when adhering to their reconstructive concept of early palatal closure.

Findings of the Eurocleft study (Mars et al. 1992; Mølsted et al. 1992; Shaw et al. 1992) revealed lower levels of growth inhibition in groups that used a variety of techniques with different timings of primary repair, suggesting that widely different protocols can produce equally good or bad results.

Berkowitz (1985, 2005, 2006) stressed the need to individualize the timing of surgery based on anatomic and functional characteristics of individual child and not just a blanket age group observance.

Proponents of late hard palate closure identified its merits as demand for fewer surgical interventions and very little need for maxillary osteotomy.

Frequent criticism of the delayed hard palate closure technique is made in relation to possible deleterious effects on speech; however, the speech problems identified were resolved by 10 years of age (Lohmander-Agerskov 1998; Lohmander-Agerskov and Willadsen 1999).

The concept of using a flap of vomer mucoperiosteum for palatal closure (Lannelongue 1872; Pichler 1934) is thought to minimize the potential detrimental denudation of palatal bone and yields enough tissue for a tension-free closure.

Use of vomer flap (single layer) (Abyholm et al. 1981) and extension of primary repair of hard palate and lip yielded the best outcome in the Eurocleft study.

41.3 Optimal Outcome Measures in Cleft Palate Surgery

The birth of a child with OFC into any family is associated with huge emotional and psychological distress to the entire family. The quality of life in families having children with cleft defects can be severely reduced (Kramer et al. 2007).

The multifaceted nature of the challenges attending OFC allows for a wide perspective in outcome measures.

Early measures like assessment, commencement of counselling and treatment of these patients and their families facilitate the overall outcome of the patients from cosmetic, speech and psychological perspectives (Rajasuo et al. 2004; Okabe et al. 2004; Adeyemo et al. 2009). Likewise, the presence of adequate nutritional counselling is known to reduce the risk of malnutrition (especially in children with cleft palate), thereby fostering proper development (Rajasuo et al. 2004). This paves the way for receiving optimal surgical care with minimal risk.

Where these measures are absent, the contrary is also true with attendant depression, social avoidance and feeling of guilt, which may persist into the adolescent years of the child and adversely affect emotional development (Okabe et al. 2004).

Evaluation of treatment outcome remains an integral part of cleft management, and a basis for evidence-based care delivery. Treatment guidelines regarding the best practice are essential requirements in contemporary clinical practice (Mossey et al. 2003). Estimating the success of cleft management and quality improvement is hinged on measuring surgical outcomes at clinical audit (Asher-Mcdade et al. 1992). Diversity in surgical techniques, skill, protocol and timing as well as resultant variation in quality of repairs inform the essentials of auditing (Mossey et al. 2003).

Many potential outcomes for comparing CLP treatment have been reported, and these include dentofacial growth and development, facial appearance, mobility of the soft palate, intelligible speech, articulation and nasal air escape, hearing, nasal breathing, adequate eustachian tube function, improved quality of life and patient satisfaction (Diah et al. 2007; Nollet et al. 2007). However, there is no agreement among the various specialists in cleft care regarding which of these outcome measures is most important (Al Omar et al. 2005). Berkowitz et al. (2005) and Berkowitz (2006) strongly condemn the use of a priority of treatment goals and strongly advocate "differential diagnosis" which includes a cephalometric skeletal analysis of the nasopharyngeal space as well as a nasopharyngoscopic exam of its muscular function. More diagnostic discrimination of the patients prior to selecting the treatment plan will improve the number of patients treated successfully. Obviously and rather unfortunately, facilities for these assessments are not within the reach of most developing countries.

41.4 Current Treatment Protocols in African Practices

A proper prelude to presenting current treatment protocols in African countries is a highlight of the peculiarities of cleft care in African countries in particular and developing countries in general. These include:

- Inadequate antenatal care
- Prenatal diagnosis is not operational
- Poor health-seeking behaviour and very high drop-out rate from the hospital
- Unhelpful cultural beliefs about cleft defects, e.g. as a consequence of parent's ill doings or the child possessed with evil spirit (Oginni et al. 2010)
- The rejection of the cleft child and sometimes the family (Oginni et al. 2010)
- Late presentation for care in children and adults (Figs. 41.2, 41.3, and 41.4) (Agrawal and Panda 2011)

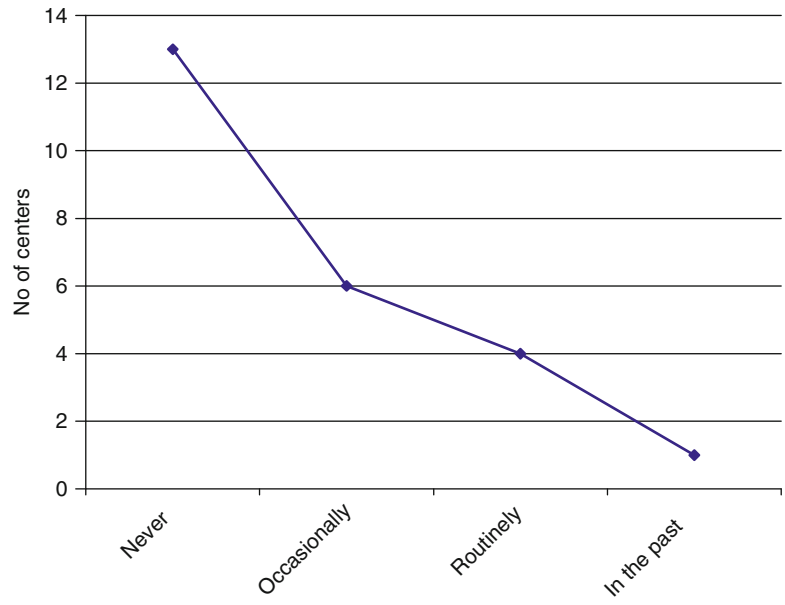

Fig. 41.1 A graphical illustration of the frequency of applying single-stage cleft lip and palate repair

- Underweight babies (Agrawal and Panda 2011)
- Financial constraints as most patients are from the low-income group
- Very minimal or non-existing government support
- Limited manpower and facilities for comprehensive cleft care especially orthognathic surgery, orthodontics, speech pathology and otorhinolaryngology

A review of the current practice in African cleft teams was conducted with the aid of a structured questionnaire. Twenty-four centres from seven countries (Nigeria, Ghana, Uganda, Kenya, Ethiopia, South Africa and Egypt) responded to the survey, inputting all the required details. Findings of this survey suggest that lip repair is undertaken routinely between the 10th and 16th weeks of life (mean 13.7 ± 4.7 weeks). Palate repair was done between 3rd and 18th month (mean 12.8 ± 4.7 months).

Most centres would routinely do CLP surgeries in at least two stages and never apply a single-stage technique (Fig. 41.1). Six centres would apply a single-stage procedure only occasionally and not as a routine. They applied it in patients aged 18 months and above, those considered able to withstand the procedure and where lip repair would be accompanied with anterior palate repair using vomer flap.

Table 41.1 Usual techniques of palate repair in African centres

Technique of palatoplasty	No. of centres (%)
Von Lagenbeck	14 (58.3)
Bardach	2(8.3)
Sommerland	2 (8.3)
V-Y push back	2 (8.3)
Von Lagenbeck–Furlow	2 (8.3)
Delaire–Lagenbeck	1 (4.2)
Intravelar veloplasty–vomerine flap	1 (4.2)
Total	*24 (99.9)*

Four other centres claim that they carry out single-stage lip and palate repair routinely. Two of these centres carry out their procedure in children aged 10–18 months and above. Striking findings in this survey however were two other centres that would do complete lip and palate repairs in one sitting at 3 months (Hodges 2010) and 6 months of age. Their usual techniques of palatoplasties were reported as Von Lagenbeck and Sommerland, respectively (Table 41.1).

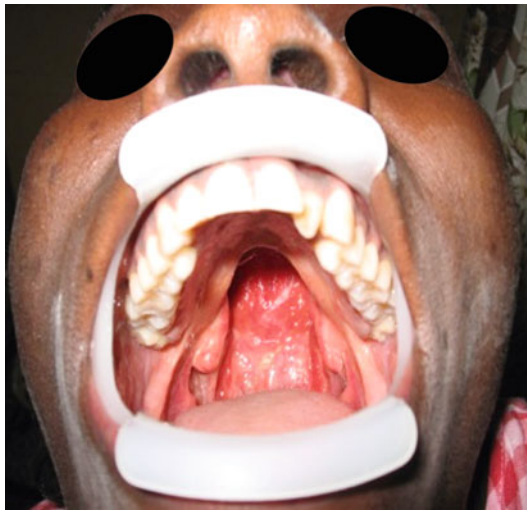

Fig. 41.2 Wide palatal defect that will defy a successful single-stage repair

All centres however lack a long-term follow-up result to show their outcome.

Routine single-stage cleft lip and palate repair in the four centres identified was predicated on:

- Avoidance of second surgery, thereby saving time and money
- Eliminating the high likelihood of a patient defaulting after first stage
- Satisfying parents' desire for a "once for all procedure and fear of repeated operations"

One of the centres does this to keep the babies alive and to forestall anticipated malnutrition being speculated to have claimed the lives of many children (Wilson and Hodges 2012).

Interestingly, one centre abandoned a single-stage lip and palate repair because of the excessive haemorrhage and severe post-operative morbidity encountered.

Surgeons that have never explored single-stage palatoplasty unanimously avoided it because they believe that early single-stage repair does tamper with facial growth.

Wide defects closed in a single stage predisposes to very extensive surgery, closure under tension and likelihood of wound breakdown (Fig. 41.2).

Additionally, they judge that the chances of post-operative respiratory embarrassment are higher.

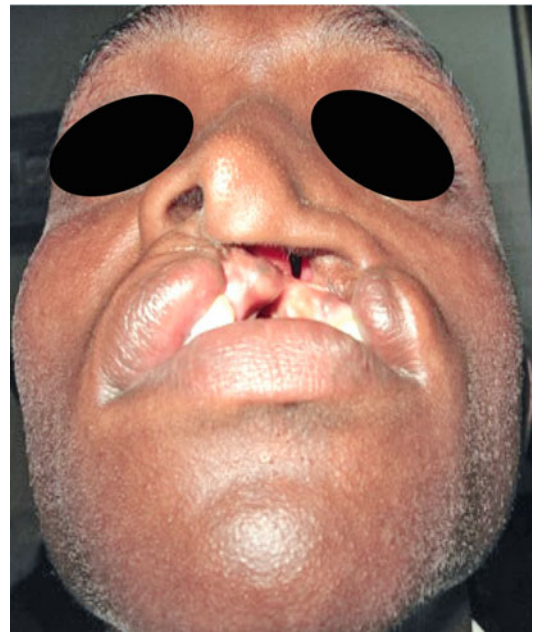

Fig. 41.3 Unilateral cleft lip and palate in a 20-year-old Nigerian male

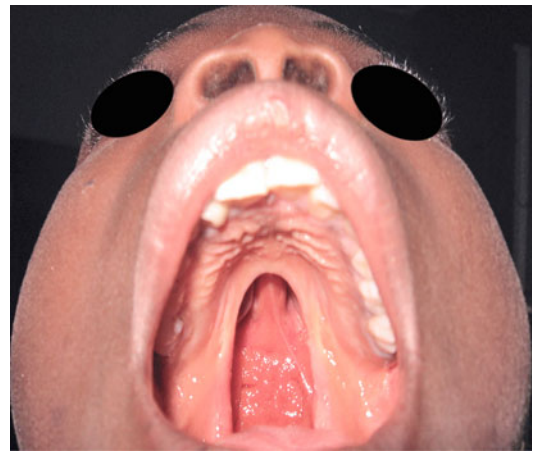

Fig. 41.4 Isolated cleft palate in a 13-year-old female

The results of this survey paint a picture of the presence of extremes of scenarios and no long-term follow-up report. We opine that the current regular support from charity organizations and philanthropists may help to reduce drop-out rate and improve outcome of follow-up studies. These

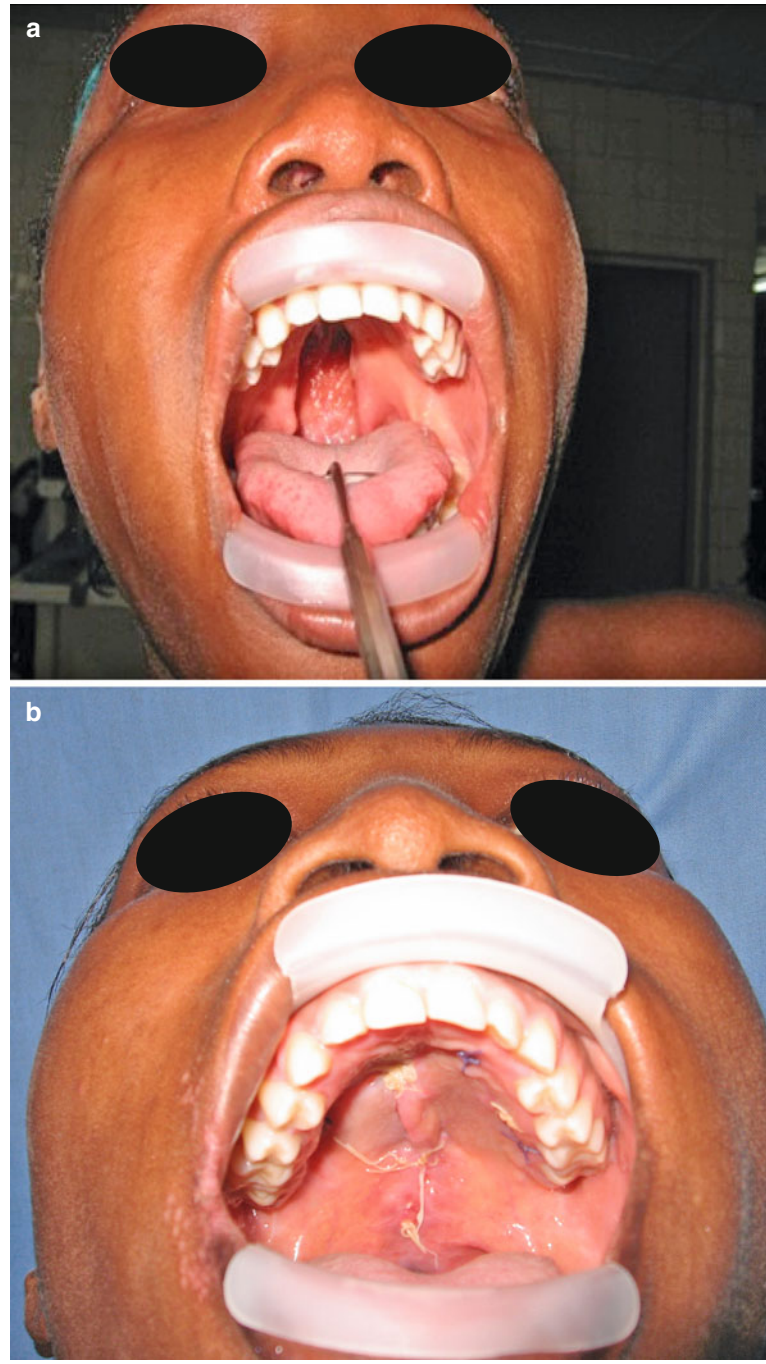

Fig. 41.5 (**a**, **b**) Pre- and post-operative pictures of cleft palate repaired in one stage in a 25-year-old female Nigerian

findings substantiate the variability and diversity in our practices and the very urgent need for a standardized protocol.

Our situation calls for attaining a balance with doing as much as possible to solve existing problems, yet getting optimal short- and long-term results.

41.5 Striking the Delicate Balance Between Aesthetics, Speech and Growth

The universal goal of CLP surgeries has been clearly spelt out (American Cleft Palate-Craniofacial Association 1993). They include

obtaining optimal aesthetic, speech, growth and hearing functions. The ideal timing and technique for palatoplasty remain an enigma. Proponents of early palatoplasty want it for better speech (Dorf and Curtin 1987), while the advocates of delayed palatoplasty (especially hard palate repair) want it for better maxillofacial growth (Gillies and Kesley Fry 1921; Hotz and Gnoinski 1979).

A fair appraisal of the existing protocols reveals desires to achieve the universal goals in all patients by all surgeons despite prevailing circumstances. Doubtless, this end may have to be reached by various means; nevertheless, there is a need to strive for safety and excellence in outcomes. Striking a delicate balance entails meeting the immediate demands, but more importantly, the long-term consequence(s) of interventions must be given very serious considerations. Measurement of treatment outcome should be an integral part of cleft management, especially now when evidence-based medical care and treatment guidelines regarding the best practice are essential requirements in contemporary clinical practice (Mossey et al. 2003).

In the absence of long-term outcomes of current practices in most developing countries of Africa, our practices can be scrutinized in the light of evidences available in the literatures.

The concept of missing palate due to malnutrition is a speculation; as such, there is a possibility that the palates may have been lost to parent's perception, fear of surgery, etc. We advocate meticulous search for the missing palates and an authentic report of their whereabout. Agrawal and Panda (2011) reported palates resurfacing just before marriage or special occasions in the family.

If indeed these patients are being lost to malnutrition, an agenda that beefs up nutritional care should serve as an appropriate stopgap protocol until the child reaches maturity with improved health.

Furthermore, evidences from available literatures suggest that a single-stage repair at 3 months to keep the child alive through good nutrition may turn out to be "solving part of the problem and creating a new one". While achieving our goals of early single-stage repair (including Von Lagenbeck palatoplasty) in these patients, available literatures suggest that we may also end up with good speech outcome, minimal scar, little or no experience of stigmatization initially and a markedly retruded middle face and a late onset stigmatization (Ross 1987a, b, c, d, e).

41.6 A Proposed Protocol

Having established that there are many protocols, the clinics still need treatment protocol(s) that address their specific peculiar factors and yet guided by test of professional ethics. Additionally, this would go a long way in helping to bridge the gap in our practices.

In formulating a protocol for the developing countries, it is important for us to start off with the realization that *the ideals* are extremely desirable for the developing countries as for the developed:

- What is proven good and proper for the developed world is indeed appropriate for the developing countries. The main goals are the psychosocial factors, facial growth, speech and dental alignment.
- The peculiarities of the developing countries as stated earlier need to be incorporated into such protocols to make them acceptable to the parents and adult patients.
- Each case should be handled on its individual merit (Berkowitz et al. 2005) since no two patients are the same. Protocols should not be rigid, but mere guides to individualizing them.
- Zero tolerance to compromise in safety and striving for long-term excellent outcome should be the goal.
- Giving due recognition to very simple measures like counselling and the involvement of social health services (to improve motivation, enlightenment by additional education and accessibility) will help patients get their questions answered at any time, regardless of their remote locations. A telephone help line is a simple measure that may facilitate this.
- As we evolve out of a huge backlog of adult patients, a passionate motivation at lip repair would go a long way to ensure patient's return for palate surgery in due time.
- An excellent primary surgery outcome would go a long way to get patients back. This will be enhanced by the support (prepaid care) received in most developing countries from international non-governmental organizations.

Table 41.2 Primary surgical treatment protocols in different cleft centres in major studies in the literature

Year	Author	Country	Age at lip repair (months)	Age at palate repair (months)
1978	Schweckendiek and Doz	Germany	–	sP 6–8, hP 12–14
1982	Takkar and Gupta	India	3–6	18–24
1983	Malek and Psuame	France	6	sP–3, hP 6 with lip repair
1990	Freedlander et al.	Britain	Neonatal	–
1992	Shaw et al.	USA		
	Centre A		3–4	sP 9–15, hP 9 years
	Centre B		Lip + Ant hP–2	post-hP + sP 22
	Centre C		>6	12
	Centre D		>6	<24
	Centre E		Lip + Ant hP–3	post-hP 18–22
	Centre F		4–6	12
2003	Lee and Kim	Korea	2.5–3 months or <6	6–12 or 12–18
2005	Weinfeld et al.	USA	3 or 3–6	–
2005	Berkowitz et al.	USA	–	18–24
2006	Flinn et al.	USA		
	Centre A		3	
	Centre B		6	sP–6, hP–18
	Centre C		3	18
2006	Noordhoff and Chen	Taiwan	3–5	11–12
2011	Agrawal and Panda	India	9–12	6–9

- Surgical techniques that entail stripping of palatal mucosa in children are not desirable.
- Granted that "what you do to start with depends on what was handed over to you especially in training", there is the need to retrain the old, learn new techniques and shift ground as occasion demands.
- The protocol proposed must be considerate of the patient's safety. The attendant co-morbidity must not outweigh the benefits.
- The big question always should be, "Considering the type of defect, anticipated duration of surgery, patient's age, general health condition, available facilities, etc., how best can this procedure be planned and carried out?"
- Over and above these factors, the training undergone by the cleft surgeon influences to a large extent the choice of the primary surgical schedule (Lee and Kim 2003). Nonetheless, there is always a lot of room for modification based on learning new skills and concepts from well-respected colleagues.

The majority of the centres worldwide prefer to repair the cleft lip at 3–6 months of age and cleft palate at 6–18 months of age (Lee and Kim 2003) Table 41.2. Even within the same centre, different surgeons sometimes follow different time schedules and techniques (Shaw et al. 1992). A situation where there are as many primary surgery protocols as there are cleft centres appears to be the rule rather than the exception.

The different time schedules in different centres are selected with specific objectives, which are mainly related to maxillofacial growth and/or speech development.

Most of the existing and published protocols are developed from and for the developed countries where many have a priority system favouring speech.

Although they strive for good dento-maxillofacial growth and psychosocial well-being, they are not always 100% successful at attaining this (Berkowitz 2006).

Reiterating and re-emphasizing the developing country's situation, there are many more factors to be considered in the surgical correction of congenital anomalies like CLP. Factors like lack of awareness, lack of funds, presence of domestic problems, "advised late surgery", child's illness, lack of operative facilities in nearby

hospitals and associated anomalies are some of the causes of late arrival (Agrawal 2007; Schwarz and Khadka 2004). Similarly, patients in Uganda did not return for cleft palate repair because they did not seem to appreciate the importance of the palate in speech production (Hodges and Hodges 2000).

Against the backdrop of the special considerations in developing countries, *an ideal protocol* must be one that suits patients' conditions without compromising overall long-term functional results in terms of speech and dento-maxillofacial growth. Simultaneously, it should improve the compliance for second surgery and not necessarily eliminate it. It should not be too taxing for children with relatively low body weight, borderline haemoglobin concentration and other related health challenges. It should not increase the number of surgical procedures and create burden for parents of low socioeconomic status. Agrawal and Panda (2011) in line with these objectives proposed palatoplasty at 6 months or at presentation (if 6 months or beyond). Lip repair follows preferably 3–6 months after the first surgery. This is similar to Malek's protocol in which only soft palate is repaired at 2 months in a first surgery. Thereafter, the lip and hard palate are repaired at 6 months. The authors concluded that early closure of soft palate ensures normal language development and less middle ear complications (Malek and Psaume 1983). Although this procedure has its advantages, it has many drawbacks as well involving the failure to have good, facial aesthetics and dental occlusion.

This chapter proposes a two-part protocol with variable staging for cleft lip and palate repairs. Part one of the protocol looks into severity of patient's defect, patient's general health condition, patient or parent's level of motivation and access to intensive care unit. From these parameters, a list of absolute contraindications and relative indications are generated.

The second part reflects on defect type and patient's age at presentation. An interval of at least 3–6 months is proposed between stages of surgery.

This proposal allows options of delayed or early lip repair. It is envisaged that the final choice will depend on parents or patient's position and attending surgeon's definitive decision. While it is often believed that parents would always want lips repaired first as it addresses the bulk of stigma experienced, Agrawal and Panda (2011) were able to prove that this is not always true. They proposed a protocol with the cleft lip and anterior palate (with primary nasal correction) as a second surgery, and parents were well disposed to it.

Only 1.3 % parents requested for lip repair at 6 months of age and palate repair at 1 year. Using the drop-out rate as an index of success, there was a 32.58 % in the conventional protocol group (lip surgery first) and 14.24 % in the modified schedule group (palate first).

Working with these patients and parents for decades, Agrawal and Panda (2011) established that the majority of parents in developing countries are easy to counsel. Though they are concerned about the baby's appearance, they readily resign the decision on protocol to the treating/managing surgeon. Years of practise have proven that the drive for the second surgery is minimal if cleft lip is repaired first because the cleft palate is hidden inside the mouth. Consequently, the high drop-out rate was reported.

On the other hand, when soft palate repair is the first surgery, the drive for lip repair is maintained, and the compliance much better with a lower to nil drop-out rate.

41.6.1 Proposed Protocol

Part 1 Fundamental components

A	B	C	D
Extent of defect	Motivation	Access to ICU	General health status
A_1 Severe	B_0 Poor	C_0 Nil	D0 Poor
A_2 Moderate	B_1 Good	C_1 Inadequate	D_1 Fair
A_3 Minimal	B_2 Very good	C_2 Adequate	D_2 Good
			D_3 Very good
Absolute contraindications to one-stage CLP repair		Relative indications for one-stage CLP repair	
A_1		A_2 & A_3	
B_0		B_1 & B_2	
C_0 & C_1		C_2	
D_0 & D_1		D_2 & D_3	

Part 2 Age considerations

Age at presentation	Initial care	Cleft lip and palate surgical protocol options
Birth to 12 months	Pre-surgical examination, non-surgical interventions, other specialty assessment	[A] Paediatrician assessment, PSO, intensive nutrition build-up, etc.
		Lip repair at 3 months
		sP–hP repair at 12–18 months (IVV and vomer flap)
		[B] sP repair at 6 months (IVV)
		hP and lip repair 3–6 months after (vomerine flap for hP)
		[C] sP and unilateral hP at 6 months (IVV and vomerine flap for hP)
		lip and 2nd half of hP 3–6 months after (vomerine flap for hP)
		*NB: sP dissection before 10 months facilitated with operating microscope or loupes
1–5 years (speech is a priority)	Pre-surgical examination, non-surgical interventions, other specialty assessment	[A] sP closure ± unilateral hP at presentation (IVV and vomerine flap)
		Second half of hP and lip 3–6 months after
		[B] sP closure at presentation
		hP and lip 3–6 months after
		NB (vomer flap preferred before 2 years and IVV for soft palate. May use Von Lagenbeck after 2 years)
6 years to adulthood (growth disturbance not a serious concern)	Pre-surgical examination, non-surgical interventions, other specialty assessment	[A] CL repair at presentation
		Palate repair 3–6 months after (multiple or single stage)
		[B] sP closure ± unilateral hP at presentation (any desirable technique)
		Lip and second half palate 3–6 months after
		Complete lip and palate repair single stage if there are no contraindications in part 1

Advantages of proposed surgical schedule for cleft lip and palate patients:

- This proposal takes key factors that affect cleft palate care in the developing economy into consideration.
- Appropriate counselling and delayed lip repair are a possible cure for the "missing palate".
- Cleft palate is repaired at the most accepted time schedule for palatoplasty (6–9 months) for good speech result in patients presenting early.
- Tries to eliminate the complication of midfacial growth disturbance, a problem that most developing countries are not fully equipped to address.
- It advises on favourable surgical technique.
- Speech was accorded appropriate priority in children presenting late.
- Although a flexible proposal, the presence of unrepaired cleft lip facilitates mouth opening and visibility of the palate, thus allowing a technically easier cleft palate repair.
- Initial closure of soft palate helps to narrow down residual palate closure over time before it is due for closure.
- During cleft lip repair, the anterior palate repair is done under good vision; hence, incidence of anterior palatal fistula will be very low.
- While performing the cleft lip repair, there is an opportunity to examine the palate under GA for any abnormality.
- Cleft palate fistula if identified can be repaired along with the lip repair.

Disadvantage:

- Vomer flap technique of palatoplasty will have to be learnt by the majority as it is not very popular in the continent from our survey.

Conclusion

While no rigid modus operandi can be formulated on a keenly debated issue like this, it is obvious that the established principles for safety and excellent short- and long-term

outcomes are universally acceptable and therefore good for the developing countries regardless of their attending peculiarities.

Striking a delicate balance for good appearance, excellent growth and speech as well as middle ear function is predicated on appropriate staging in the appropriate patient at the appropriate time and using the appropriate surgical technique(s).

The proposed schedule for primary surgeries in many children and few adults with CLP is aimed at improving compliance for the timely repair of palate and lip in developing countries where the situation warrants an innovative viewpoint because of unique existing factors.

The palatoplasty performed first at 6–9 months is a universally acceptable time for the development of good speech and hearing, and cleft lip repair 3–6 months later should not affect maxillofacial growth. The compliance for second-stage surgery improves significantly with this schedule.

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Birthing and Building Nascent Cleft Teams in Developing Countries

42

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42.1 Introduction

Cleft congenital malformations are a major issue in developing countries due to the disproportionately high birth rates in poorer areas of the world. Nearly 95 % of annual births in the world, and 94 % of all children born with clefts, are born in developing countries (Mars et al. 2008). The etiology of the high cleft incidence in developing countries remains unclear. However, it is presumed to be multifactorial, due to various environmental and genetic factors (Mangold et al. 2011). Toxin exposure during the antenatal period is more likely in developing countries due to poor sanitation, inadequate infrastructure, and political instability (Hseih et al. 2011). For many of the same reasons, malnutrition is also more likely to be an issue for a pregnant woman living in the developing world (Pelletier et al. 2011). Genetic factors are also a major potential cause of birth defects, because consanguineous practices are generally more prevalent in developing rather than in developed countries (Sandridge et al. 2010).

While the incidence of clefts in developing countries is high, the resources directed toward treating this global health issue are not increasing (Mars et al. 2008). Consequently, the prevalence of unrepaired clefts continues to grow, making the need for coordinated global cleft care all the more imperative. Over the past 10 years, global health efforts, by both the nongovernmental organizations and the private sector, have largely focused on the area of infectious diseases through the development and distribution of vaccines and antiviral therapies (Nishtar and Jan-Llopis 2011). While these areas are important, a shift is necessary toward recognizing the major burden of noncommunicable conditions on developing countries. With 80 % of noncommunicable diseases occurring in low- and low-middle-income countries, these countries are burdened by the economic implications of decreased productivity and shortened life expectancies (Livestrong 2011). The realities suggest that the treatment of noncommunicable conditions, such as clefts, can serve as part of a larger infrastructure development program.

The general scarcity of medical resources and skilled healthcare practitioners in developing countries significantly affects the ability to treat patients with clefts (Mars et al. 2008). These

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complex conditions require multiple, timed surgeries that then require the attention of nonsurgical healthcare providers (Abbott et al. 2011).

42.2 Understanding Context

Development initiatives, whether medical, economic, or structural, must be aligned with the vision of the community undergoing these changes (Gasper 1996). Therefore, prior to medical intervention in a foreign setting, healthcare practitioners must understand local conceptions of health and well-being. Simply asking the question, "What does being healthy mean in this community?" provides the starting point to implement interventions. Exploring a community's representations of healthy living is essential to understand how the local people will approach a foreign medical team and react to the changes implemented.

Additionally, discovering how patients with clefts are treated within a cultural context helps the cleft care volunteer better understand his or her role within the cultural context of his host country. By extension, understanding how a cleft condition affects a patient's daily living has implications on the patient's postoperative course.

42.3 Foreign Physician Roles and Language

Physicians around the world are often not treated with comparable respect to doctors from developed countries (Gruen et al. 2004). Visiting physicians must understand both the privileges and limitations accorded to local physicians. A visiting physician should understand whether or not the local community trusts physicians, what gender dynamics affect the patient-physician relationship, and become familiar with mainstream medical practices (Verbrugge 1985).

Understanding the local language has serious implications for treatment (Wilson et al. 2005). Precise meaning may be lost in translation, resulting in costly mistakes. To mistranslate a patient's allergies or blood type can be a lethal mistake. Multiple other errors in medications, length of treatment, and wound care after surgery, though not lethal, can lead to unnecessary morbidity in a trusting patient population. These risks demonstrate the importance of having medical translators, or team members who speak the local language, before any attempt is made to provide surgical care in a foreign setting.

42.4 Considerations for the Next Trip

42.4.1 Geography

When providing medical care in foreign settings, it is important to consider geography and landscape (Blaikie 1995). Foreign medical interventions should be both visible and accessible to the local community. For example, if care is provided in a mountainous area, the team should ensure that hospitable roads are available for patients seeking care. Think about practicalities such as transporting essential supplies and equipment. Will there be problems with Customs? Will tariffs need to be paid on equipment that is being transported? How will equipment be transferred from an airport to the local hospital? Can equipment/supply safety and sterility be maintained during transit? Relief work may have unintended effects on the surrounding environment (Debrix 1998). For example, creating a cleft care facility may redefine a town by expanding its population dramatically. Similar considerations need to be entertained long before the arrival of the foreign cleft care team.

42.4.2 Timing

Cleft care is time-sensitive (Abbott et al. 2011). In an ideal system, patients with clefts begin receiving treatment as infants (Mars et al. 2008). However, in developing countries, most patients do not have that luxury and may simply wait for the next set of foreign practitioners. Given the potential complexity of cleft care, foreign providers must (at least initially) arrange a regular

scheduled return to provide follow-up for patients who have undergone surgery. Local practitioners need to become involved stepwise in this plan of care in order to eventually perform the follow-up independently.

Timing of the surgical trips may also have effects on the preoperative status of patients. Traveling to perform cleft procedures during the winter months, when many infants will likely suffer from seasonal upper respiratory infections, may lead to unnecessary cancellation of cases. Travel in the Fall or Spring seasons may avoid these problems.

42.4.3 Preparation

Sufficient forethought and organization is critical for a successful trip (Mars et al. 2008). Engaging the local community prior to arrival is crucial in order to accomplish this goal (Murray et al. 1994). Foreign medical teams can, for example, provide protocols to local staff for preparing the facilities they hope to work in and publicize their arrival in the local news in order to ensure that patients make arrangements to receive necessary care. Forethought by foreign practitioners includes taking precautions for their own health, ensuring that everyone receives necessary vaccines and has appropriate prophylactic medicines (Hamer and Connor 2004). Preparation often entails bureaucratic arrangements, such as obtaining visas, temporary work permits, and Customs clearance.

42.5 Interdisciplinary Care

The highly interdisciplinary nature of cleft care makes this service unique. A team providing truly comprehensive cleft care includes surgeons, anesthesiologists, nurses, pediatricians, speech and language pathologists, dentists, and orthodontists. The importance of incorporating all of these disciplines on a trip cannot be overstated. Ideally, some of these providers will be local practitioners, helping deepen the connection between the foreign team and local community. Reliance on local practitioners should grow as the work continues.

42.6 Forming Partnerships

Developing local partnerships helps the foreign team harness sufficient political support, develop alliances with medical facilities, and engage the community more broadly (Berke et al. 1993). In the context of political instability, partnerships may allow foreign providers access to vulnerable populations. Developing alliances with medical facilities is important in order to allow exchange of resources, decrease redundancy in services, and enable local providers to receive training from foreign providers.

42.7 Patient Selection

Volunteer cleft care teams should hold themselves to the same high standards when operating in a developing country that they hold themselves at home. This starts with comprehensive preoperative evaluations of patients who are surgical candidates (Kitlowski 1932). These evaluations can be facilitated when local practitioners play an active role in the screening process, making referrals to local physicians for diagnostic tests as needed. Visiting professionals must not get caught up in the need to complete a certain number of surgeries, but must maintain high safety standards; a cancellation for appropriate reasons simply means that the child's surgery can be done at a later, safer time.

42.8 Safe Surgery

Developing a "culture of safety" is paramount to overseas volunteer work. This starts with the group of foreign cleft care workers, but eventually must include all of the participating local practitioners. Protocols must be instituted to prepare a facility for surgery, maintain count, and ensure that all necessary surgical steps are taken (Gawande 2009). Checklists have become part of

standard of care in developed countries and should be instituted on volunteer surgical trips. Surgical care should be delivered based on established protocols. However, providers should also be trained to adapt protocols when it is in the best interest of a patient (Thomson et al. 2010).

Setting a reasonable caseload for the trip is also an important safety measure. Limitations on resources, human capital, and time can affect safety if not properly recognized (Vincent et al. 1998). Cleft teams need to recognize their limitations and only operate when conditions allow for maximal safety provisions to take place (Charles et al. 2011).

Foreign providers, and eventually the more involved local providers, need to be able to abort a surgical case when appropriate. Saying no in these settings can be very challenging for both the physician and for the patient or patient guardian. Patients or patient guardians may not necessarily understand why not receiving surgery is in their best interest. Clear communication using language that can be easily understood by families is essential. Safety is the primary argument for denying care in the setting where risks outweigh benefits.

42.9 Patient Follow-Up

Patient follow-up is essential (Canady et al. 1997). This care is generally the most immediate way to involve local providers, yet must be done with careful training during the trip in order for them to recognize and treat potential complications following surgery. The capacity to take pictures and send them globally to foreign team members can facilitate communication about patient problems. Complications should be adequately recorded, and there should be a forum that allows honest discussion of the complications. When foreign physicians return to the local community, patients who have previously undergone surgery should be seen as part of the screening process. Eventually, local providers should play an increasingly central role to this process of recognizing and treating complications following surgery.

Fig. 42.1 Nurse providing postoperative care to a child with cleft palate in Tulkarm, Palestine, 2011

42.10 Sustainable Cleft Care

Before traveling to a developing country to perform cleft care, team members should have a vision of what sustainability in a particular location should look like. A plan for sustainability means that visiting physicians should teach more than treat, training local surgeons, nurses, and paramedical personnel the standards of cleft care (Berke et al. 1993) (Fig. 42.1). Training local practitioners sets the stage for the care to be part of a local independent entity in the future. Additionally, if local healthcare providers are able to provide adequate care, they can continue to train others and therefore increase the human capital providing cleft support in the area. Training local medical personnel also empowers the community to be self-sufficient and care for their own children.

Sustainability is also contingent upon sufficient funding and supplies for the care provided. Cleft care teams need to create continuous fund-raising initiatives, finding consistent ways to support the development of cleft care in the setting of interest. Cleft care is also dependent on supplies for multimodal therapy; therefore, seeking consistent support from supply companies can ensure that trips are successful. Many hospitals are able to donate unused or excess supplies. In addition to a steady stream of supplies, a cleft care team needs

a base facility. A physical structure that serves as the home of cleft care simplifies patient follow-up, enables storage of equipment, and serves as a base for potential research and education projects in the community.

A sustainable model for cleft care involves handing off leadership to local practitioners. Foreign providers have to establish metrics for recognizing local cleft team members are fully trained to provide unsupervised surgical and medical care to the local community. Discussing these metrics with local provider sets the stage for mutual recognition of independence.

Access to continuing education is an essential part of forming a cleft team (Davis et al. 1999). Providers, both local and foreign, need open access to fellowships, scholarships, and educational materials. This provision also protects the community receiving care by helping ensure that the providers are exposed to and hence are practicing the most modern approaches to treatment.

Another essential factor needed to make the cleft work sustainable is enhancing local volunteerism – enabling the community to feel engaged with the work being done by the cleft team (Sturmer and Kampmeier 2003). Local volunteerism leads to the establishment of local conferences, fund-raisers, and activities around the care of cleft patients. Integrating the cleft team into the society through local volunteerism will ultimately promote sustainability.

42.11 Research

Foreign providers developing cleft care teams in developing countries must be sensitive to the ethical issues regarding the conduct of research (Buchanan and Miller 2006). While research is an important process that drives innovation and helps to obtain sustainable funding for the cleft team, it can also compromise the safety and privacy of patients (Hyder et al. 2004). Therefore, researchers must work closely with the local providers to ensure that patients are fully protected. Research must receive institutional review board (IRB) approval from both local and foreign institutions. Additionally, all participants must sign a consent form and have the freedom to leave a study at any time and to ensure that all patients are aware of their rights in a research setting.

Research also serves as a tool for implementing primary care for cleft patients. In order to institute preventative care, researchers around the globe must work together, using the World Health Organization's method of following specified Millennium Development Goals (Mossey et al. 2011). A collaborative research approach will improve outcomes for patients with clefts throughout the world.

42.12 Cleft Care in the West Bank, Palestine

In 2006, cleft practitioners from the University of North Carolina (UNC) at Chapel Hill Department began traveling to the West Bank and Gaza to provide cleft care. Surgical trips have been scheduled twice a year. While foreign surgeons are not available, local practitioners provide follow-up to previous patients and schedule future patients.

Practitioners who have participated in these trips have worked with local practitioners in order to build a sustainable cleft team. Visiting practitioners have trained local surgeons to perform cleft care. From the outset of these trips, the decision was made not to perform any surgery without the presence of a local surgeon; this has ensured that every case is the opportunity to further the training of local practitioners. Since 2009, local surgeons have increasingly performed independent repair of cleft palates; since 2010, cleft lip surgery has been done by local practitioners.

Local providers have been given opportunities to participate in international conferences on cleft care, attend educational seminars, and in the conduct of IRB-approved research. The Palestinian Cleft Society, established in 2007, now with exclusively Palestinian leadership, assists in

Fig. 42.2 2009 Annual Palestinian Cleft Society Meeting banner (Ramallah, Palestine). The title of the conference was "Dental and Orthodontic Needs of Children with Clefts"

overseeing the care of Palestinian children with clefts throughout Palestine (Fig. 42.2).

In order to make cleft care in Palestine, and throughout the world, sustainable and globally collaborative, this nascent cleft care team has partnered with both nongovernmental and governmental organizations like the Palestinian Ministry of Health, the Smile Train, Operation Smile, and ReSurge.

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Challenges in Cleft Care in Underdeveloped Countries

43

Isaac L. Wornom III

43.1 Parameters of Care

Interdisciplinary or team care is defined in the parameters document of the American Cleft Palate-Craniofacial Association (ACPA) as:

- The staff of the interdisciplinary team may include individuals from the following areas of professional practice: anesthesiology, audiology, diagnostic medical imaging/radiology, genetic counseling, genetics/dysmorphology, neurology, neurosurgery, nursing, ophthalmology, oral and maxillofacial surgery, orthodontics, otolaryngology, pediatrics, pediatric dentistry, physical anthropology, plastic surgery, prosthodontics, psychiatry, psychology, social work, and speech-language pathology.
- The principal role of the interdisciplinary team is to provide integrated case management to assure quality and continuity of patient care and longitudinal follow-up.

(Official publication of the American Cleft Palate-Craniofacial Association 2009)

Team care is generally accepted around the world as the best way to care for patients with clefts and other craniofacial anomalies.

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43.2 Obstacles to Team Care in the Developing World

Ninety percent of the world's population has access to only 10 % of world health resources. This leads to many barriers being present to providing interdisciplinary team care to children with clefts in developing countries. One of these confronted by groups traveling to the developing world to deliver care is overwhelming numbers of patients with clefts needing surgery. In addition, when care is provided by a foreign group who is not part of the native population, there is usually limited time available to see follow-up patients. Regular follow-up speech therapy and orthodontic care is often unavailable and difficult to provide in a single 1- or 2-week visit. Travel for patients for follow-up on return visits may be difficult, and they may not show up.

43.3 Team Care in the Developing World: Making It Happen

How can we begin to provide patients who are born with clefts in developing countries the same interdisciplinary team care we give to our patients with clefts in the developed world? There are basically two ways to develop team care in places around the world where it does not exist. One is to travel with an interdisciplinary team to a developing country to provide care for patients with clefts and go to the same place over a long period of time. The trip should have a strong educational

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component as part of its mission and be committed to working with local providers whenever possible to deliver care. Dr. Hussein and his coauthors give a good description of how they have accomplished that in Palestine in this chapter. The second way is to bring providers from developing countries to the developed world to learn interdisciplinary team care for patients with clefts and help them take that care back to their home. An example of this is the visiting scholar program of the American Cleft Palate-Craniofacial Association.

43.3.1 Mission Trips

Anyone contemplating a trip to foreign country to provide cleft care should have a strong umbrella organization for mission planning and coordination. There are many excellent groups around the developed world who serve this role. The group should have good government and medical connections within the country which is the focus of the trip. The umbrella organization should check the credentials of the team; organize logistics of transportation, housing, and food; arrange for interpreters if needed; publicize the team in the country prior to arrival; and keep longitudinal patient medical records. It should also help identify local health-care providers who can work with the visiting team to learn interdisciplinary team care over an extended period of time so knowledge of team care can grow in the host country.

The most important thing for a successful introduction of team care into an area by a mission trip group is a commitment to return to the same place at least yearly for a long period of time. Dr. Hussein and his coauthors clearly model that in what they have done in Palestine. This commitment allows follow-up of patients over a long period of time, keeping of longitudinal records, accomplishing sequenced reconstructions, introduction of speech therapy and orthodontic care as a component of treatment, and evaluation of outcomes. In addition, whenever possible, education of local providers can be carried out year after year until they can provide the care on their own without the team having to come. This should be the end goal.

Surgical safety is extremely important. *World Health Organization* surgical checklist guidelines should be followed in the operating room (Official Publication of the World Health Organization 2011). *Guidelines for the Care of Children in the Less Developed World* which was published in 2011 gives detailed recommendations regarding anesthetic and surgical equipment and procedures to follow for maximizing patient safety on mission trips (Schneider et al. 2011). Participants on mission trips should practice within their specialty. The developed world is not the place to do something you do not normally do in your home country. All trip personnel should be appropriately licensed and board eligible or certified in their specialty. Virtually all umbrella organizations and ACPA and PSEF position papers support these ideas.

Patil et al. from Nagpur, Maharashtra, India, published "Changing patterns in demography of cleft lip-cleft palate deformities in a developing country: the smile train effect-what lies ahead?" in *Plastic and Reconstructive Surgery* in January of 2011. This was a retrospective study of a three-decade experience combined with a survey on awareness of cleft deformities of patients treated by the authors compared with patients treated initially by camp surgeons. In this study, there was a marked decrease in numbers of patients with cleft lip and cleft lip-cleft palate treated at their established unit during this time period, but the number of patients with isolated cleft palate stayed the same. The age of patients reporting for palate repair was 16 months in their patients and 41 months in patients treated initially in a camp setting. The patients operated initially in camp settings did not have the same awareness of the need to follow a time line treatment protocol or awareness of their deformity and need for follow-up as those operated on initially by the authors. They speculate that camp surgeons operate preferentially on cleft lip and not on cleft palate. They conclude that the Smile Train project has helped cleft care in India, but counseling and improved team care should be the focus of the future.

Perhaps, the best example of how returning to the same place for a long period of time can lead to the development of interdisciplinary team care is the work done in Sri Lanka by Michael Mars and his group. They recently published a book on their experience returning to the same place for 25 years. Their work demonstrates how consistently returning to the same place can lead to superb outcome evaluation and education of local providers to provide interdisciplinary team care. *Management of Cleft Lip and Palate in the Developing World* by Mars, Sell, and Habal should be read by anyone contemplating such endeavors.

43.3.2 Bringing Providers to the Developed World

The ACPA Visiting Scholar Program is an excellent example of how an individual from a developing country with the knowledge and potential to bring interdisciplinary team care for cleft and craniofacial patients to that country can be nurtured and encouraged. Each year, one individual spends 6 weeks in North America visiting cleft and craniofacial teams and attends the ACPA annual meeting. The individual then returns to their home country and uses the knowledge they have gained to establish or improve interdisciplinary team care. This program has been in place now for 15 years, and numerous examples exist of how these people have brought team care back to their home countries.

Dr. Peter Donkor, an oral and maxillofacial surgeon from Ghana who was the ACPA visiting scholar in 2005, demonstrates the effect this type of program can have. Over the past 6 years, he has established a multidisciplinary team at his institution in Kumasi, Ghana. He also helped found the Pan African Association of Cleft Lip and Palate of which he is now president. Finally, he began a collaboration with Dr. Michael Cunningham of Seattle, Washington, who was his visiting scholar sponsor promoting team care for cleft lip and palate throughout Africa by bringing teams from throughout Africa to Kumasi and Seattle for training.

43.4 Summary

There are two ways to foster team care for patients with cleft lip and palate in the developing world. One is to travel with an interdisciplinary team to a developing country to provide care for patients with clefts and go to the same place over a long period of time with a focus on educating local providers and empowering them. The second is to bring providers from developing countries to the developed world to learn interdisciplinary team care for patients with clefts and help them take that care back to their home. Both have roles to play in the future. Those who provide this type of care need to focus on both going forward if team care is going to become a part of cleft care in the developing world. This should be the goal going forward.

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Part XV

Psychological/Team Function

Examining the Team Process: Developing and Sustaining Effective Craniofacial Team Care

44

Lynn M. Fox and Patricia Ann Stone

44.1 Introduction

44.1.1 History of Team Care

Multidisciplinary medical team care emerged as a new form of practice when medical and surgical teams were created to treat the injured in the United States during World War II. Craniofacial team care, however, predates the war. The Lancaster Cleft Palate Clinic was instituted in 1938 by Dr. Herbert K. Cooper, an orthodontist in Lancaster, Pennsylvania, to treat patients using the team approach. By 1943, Dr. Orvin Reidel suggested a permanent organization to "encourage by every appropriate method and device the improvement of scientific clinical services to persons suffering from cleft palate and associated deformities in order that they may achieve a more adequate physical, emotional, social, educational, and vocational adjustment; and…we believe that these interests and desires may be satisfied by a closer and more purposeful association of persons now interested in and serving those patients in a variety of ways…," thus creating the first national organization devoted to craniofacial team care and research, now known as the American Cleft Palate-Craniofacial Association (ACPA) (Berlin 1969). The concept of team care further advanced in the greater medical community when in 1949, Dr. Martin Cherkasky established a home-health program at Montefiore Hospital in New York with the goal of "salvag(ing) a human being, and this by individualizing her care and by coordinating all the facilities of the hospital and community in their joint fight for health against disease" (Cherkasky 1949). During this period, craniofacial teams were being created around the country. By 1958, Dr. George Silver's Family Health Maintenance Demonstration Project (also at Montefiore Hospital) utilized teams of physicians, nurses, and social workers for family health care. This program would represent a significant influence in the development of present-day ambulatory care programs, especially modern health team care (Sidel 2006). The concept of focusing on the "whole patient" through interdisciplinary team care was further refined in the 1960s through President Johnson's "Great Society" and "War on Poverty" through which community health centers were formed to provide the poor and underserved with basic health care. The most direct event leading to the proliferation of modern medical team care occurred in 1970 when Sidney Garfield of Kaiser Permanente pioneered the concept of "managed care," touting improved efficiency in the health-care system by increasing

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collaboration between health-care providers to align patient needs and provider skills. The focus was placed on the "well patient" and coordinated preventative care rather than focusing on the sick patient (Baldwin 2007).

44.1.2 Current Parameters of Team Care

As team care developed in health care and particularly in craniofacial care, the team approach was soon established as the most effective care delivery model, providing efficient care by a large number of medical specialists who evaluate complex patients relatively quickly and are able to create a plan of treatment comprehensively addressing patient needs. As opposed to the traditional medical model that seeks to identify and cure disease, the craniofacial team model focuses on all aspects of the patient's life affected by the craniofacial difference. Not only is the repair of the physical difference (cleft palate or skeletal malocclusion or any other craniofacial anomaly) addressed, but the team also addresses the other aspects of the patient's social, emotional, financial, educational, and physical well-being.

Moreover, the physical issues are addressed only as the team, including the patient and family, identifies those issues causing difficulties. Recent studies have also shown that hospital-based multidisciplinary clinics considerably reduce Medicaid costs in medically complex children (Casey et al. 2011). Interdisciplinary team care is more challenging in health care than in other fields because each patient has different medical needs and represents a new set of problems which must be solved in a relatively short period of time versus teams in business or pure science fields (e.g., computer science) that can spend months or longer to solve complex but fixed problems (Kuziemsky et al. 2009). Team care is simply more efficient for complex patient care.

Today, there are more than 250 cleft palate/craniofacial teams in the United States and Canada, and teams continue to proliferate across the globe. The ACPA has defined a cleft palate or craniofacial team as "comprised of experienced and qualified professionals from medical, surgical, dental, and allied health disciplines working in an interdisciplinary and coordinated system. The purpose and goal of teams is to ensure that care is provided in a coordinated and consistent manner with the proper sequencing of evaluations and treatments within the framework of the patient's overall developmental, medical, and psychological needs." The association further defines cleft palate and/or craniofacial teams as including at least a speech-language pathologist, a surgeon, an orthodontist, and a patient care coordinator who are all appropriately trained. All teams must also have access to psychology, social work, psychiatry, audiology, genetics, general and pediatric dentistry, otolaryngology, and pediatrics/primary care. To be designated a craniofacial team (i.e., a team that also addresses the needs of patients who require intracranial surgery), the team's surgeon must be trained in cranio-maxillofacial surgery, and the team must include a psychologist who completes neurodevelopmental and cognitive assessments. The craniofacial team must also have an established referral pathway to neurosurgeons, ophthalmologists, radiologists, and geneticists (American Cleft Palate-Craniofacial Association Commission on Approval of Teams 2010).

Craniofacial and other medical/professional teams can be categorized into three types of teams: multidisciplinary, transdisciplinary, and interdisciplinary. Within multidisciplinary teams, individuals keep their own scopes of practice sacred. Communication between team members is generally formal and may occur through written correspondence or a formal face-to-face or video conference. Individual team members tend to work independently with the ultimate goal of combining separate specialists' findings into a list of recommendations. The transdisciplinary team model is characterized by fluid professional boundaries and unconventional roles that cross professions. Communication between team members is more often informal and almost always face-to-face. Interdisciplinary teams are more of a cross between the multidisciplinary and transdisciplinary models. On interdisciplinary teams, there is an overlapping of professional roles where team members develop a working knowledge of

other professions as they relate to their own examinations and combine all team members' results to help develop a cohesive plan for patient care. Communication can be either formal or informal between team members. This model encourages contributions from all specialists on the team and promotes cross-disciplinary problem-solving.

Of the three basic team models, the interdisciplinary team model is the most common type in craniofacial team care and is seen as improving patient care outcomes and job satisfaction for team members (Ellingson 2002). Practical application of the interdisciplinary model, however, has the potential to invoke tension, defensiveness, or "turf" issues due to overlapping professional roles. Craniofacial teams require that many professionals work together in a climate where all team members are perceived as equals with an equal say in the determination of patient care. This process requires excellent communication, trust, respect, and a willingness to compromise on the part of all. Issues inevitably arise when so many complex patient care issues are involved. Since team members meet together for a relatively short period of time each week, these issues may be unaddressed, compromising team functioning and ultimately the quality of patient care. Therefore, the development of a successful team requires careful team formation, development of a team mission, balancing the composition of the team, and consistent team leadership. Sustaining successful teams entails ongoing attention to the team process through the development and maintenance of effective decision-making skills, collaboration skills, communication skills, clearly defined team roles, mutual trust and respect, team goals, conflict management skills, an awareness of the influence of values, and a strong code of ethics.

44.2 Establishment of Teams

44.2.1 Team Formation

Every year, new craniofacial teams form around the world to help meet the needs of children and adults afflicted with cleft lip and palate and/or other craniofacial conditions. Regardless of where or how a given team is created, all teams tend to follow four classical stages of team development: forming, storming, norming, and performing. These stages were first outlined by psychologist Bruce Tuckman in 1965 (Tuckman 1965).

The forming stage occurs when a team is first conceptualized and created. The mission, size, and membership criteria must be set. This stage provides the essential foundation for future success of the team.

In order to reach the point where the decision-making process can be established, the storming stage must occur. This stage involves bids for power or influence and tends to include emotional arguments that continue until the necessary ground rules for decision making are agreed upon. While this stage can be uncomfortable for team members, it is necessary for continued team maturation. This is the stage where the team leader's ability to handle conflict becomes crucial.

Once the team has agreed upon how to make decisions, the norming stage begins where team rules and processes are decided. The unwritten norms of acceptable team member behavior are another product of this stage. These norms will guide the team in how to work as a coordinated unit.

The performing stage, the final stage originally delineated by Tuckman, is where the team begins a collaborative working relationship. This point of team development is the first time the team actually functions as a truly collaborative unit to address the goals for which the team was developed in the first place: care of the craniofacial patient. The accomplishments that the team enjoys during this phase tend to unite the team more closely, reinforcing the team identity (Cardona and Miller 2000).

Teams are constantly evolving as evidenced by the fact that rather than remaining at the performing stage, healthy teams eventually transition to a new forming stage where they tackle increasingly challenging tasks. Affection and trust increase as team members have the opportunity to recognize and value the other members' efforts and capabilities. A new storming stage follows as the team assumes new goals, which in

turn leads to an improved norming stage. This trend culminates in an improved performing stage and continual maturation of the team. However, a sudden and radical change in team membership may alter the team's identity enough to set the team's formation back to less fully developed levels.

In 1977, Tuckman added a fifth stage of team development called the "adjourning" stage where the team completes its task and disassembles (Tuckman and Jensen 1977). Most craniofacial teams do not reach the adjourning stage since the needs for their service continue until a patient becomes an adult and new babies continue to be born with cleft palate and other craniofacial anomalies. Budget cuts, the establishment of too many teams serving one small geographic area, and fluctuations in health-care funding are examples of situations that can necessitate the "adjourning" of a craniofacial team. Occasionally, teams are meant to be temporary as in the example of craniofacial teams from more developed countries that travel to countries lacking craniofacial team care (or who are in the midst of political strife or natural disaster), with the express goal of helping to establish indigenous teams (Strauss et al. 2011).

44.2.2 Team Mission

One of the first steps in establishing any team is the development of a strong team mission statement that provides the entire team with a unified goal. This statement should be generated and agreed upon by the entire team to ensure everyone's commitment. The team mission should not be a static document or concept and should be revisited over time to be refined or replaced with a more appropriate vision as the needs and purposes of the team develop and mature. A dynamic team mission statement should provide the basis for maintaining high ethical standards. Craniofacial team missions should incorporate all aspects of the team's functions and typically include patient care, research, public service, education, etc. For example, the mission statement for the University of North Carolina Craniofacial team in Chapel Hill, North Carolina, is "to provide optimal care for patients with cleft lip, cleft palate and other craniofacial anomalies through an interdisciplinary team-oriented approach and to stimulate biological, behavioral and clinical research that will ultimately lead to an improved quality of life for our patients" (University of North Carolina Craniofacial Team website 2011). Similarly, the mission statement for the Akron Craniofacial Team in Akron, Ohio, is as follows: "to optimize the care of individuals affected with cleft lip and palate and other craniofacial anomalies, through education, research and the promotion of interdisciplinary team care. We strive to do this in a congenial atmosphere which fosters trust and confidence" (Lehman J, 2011, personal communication). Such mission statements provide a uniting purpose for the team.

44.2.3 Team Composition

Team size and diversity have also been identified as areas with significant effects on team development. Typically, teams of less than five people may not possess enough different perspectives, while teams of more than 25 members may encourage the formation of subgroups and so have difficulty collaborating (Katzenbach and Smith 1994). Diversity of team members has been shown to provide a foundation for more complex and complete collaboration (Bolman and Deal 1992). Too much diversity may hamper common understanding and shared interests, while too little diversity may encourage "inbreeding," "group think," and "lack of complementary skills" (Cardona and Miller 2000). As the team comes together, the members work toward becoming acquainted with each other and developing complementary technical, problem-solving, and interpersonal skills. These skills develop further over time. During this stage, members often experience feelings of insecurity and anxiety until they define and agree upon their mutual expectations regarding team functioning and members' expectations (Cardona and Miller 2000).

The ACPA requires that cleft palate and/or craniofacial teams have specific disciplines involved in team care as previously stated. Yet given health-care models, one also must include health administrators, lawyers, and insurance companies. In addition, teams may have more than one person representing each discipline. The large size and diverse composition of many craniofacial teams can pose challenges to developing a high performing team.

44.2.4 Team Leadership

The role of team leader is pivotal. While many roles are somewhat fluid with the degree of impact fluctuating over time and situation, the team leader remains a constant presence, keeping the team on the path set forth in its mission. Teams vary regarding the amount of influence and effectiveness held by its leader. The outlook of the team is generally shaped by its leader and is generally established early in the leader's tenure along with the expression of clear expectations for attendance, guidelines for constructive discussion, confidentiality, task completion, and equality of members' contributions (Katzenbach and Smith 1994). Once the team expectations are communicated, it is the team members' duty to follow the rules and the team leader's responsibility to enforce them.

The team leader also has the responsibility of eliminating situations that represent barriers to the team's performance. On the organizational level, institutional managers may not have experience with or knowledge of the medical team approach. For this reason, they may not provide adequate or appropriate support to the team. In this situation, the team leader would need to advocate for the team to insure appropriate resources are provided. On the team level, an absence of clearly established rules, roles, or procedures can adversely affect team performance, sometimes leading to interdisciplinary rivalry.

In healthy team functioning, team decisions are based on the input of all team members. In order to help promote equality of team members, some teams rotate their team leaders so as to grant equal status to all professional groups (Horwitz 1970; Strauss and Broder 1985; Strauss 1999). In less functional teams, the team leader (often a physician) may make decisions without careful consideration of other members' input, thereby reinforcing a skewed balance of power and limiting team outcomes (Mason and Riski 1982).

Most leaders have a preferred style of team leadership, of which there are several. Teams that were created by one or two dynamic individuals sometimes tend toward an authoritative model of leadership whereby the founder guides the team toward specific goals. This model may generate dedication to the team as a benefit of having such a dynamic leader. This "authoritative" style of leadership often encourages positive outcomes and attitudes and is most likely to keep the team on track toward the basic mission (Goleman 2000). The team leader also must clarify the team's goals and must establish effective procedures to resolve problems in order to maintain a functional team (Baldwin 2007). Care must be taken, however, that an "authoritative" style does not devolve into an "authoritarian" style.

With in the affiliative style of team leadership, the team leader places people and interpersonal relationships before all other goals. This style promotes closer bonds between team members and encourages a familial dynamic. It may also help patients' families feel more welcomed as members of a cohesive team. While this type of leader can help bring the team together and minimize divisiveness, the team risks losing sight of its mission unless the leader provides guidance toward the team's basic goals and mission.

The "coaching" style focuses on improving the skills of individual team members and moving the team toward excellence. While this represents a positive leadership model, the main focus for craniofacial care needs to be improving patient and family care.

Many teams function under a "democratic" style of leadership which Goleman suggests "builds buy-in or consensus" (Goleman 2000). In the case of a craniofacial team where the team members are from different professions and the decisions are quite complex, consensus is not

always possible. Lack of consensus may lead to stagnation in team development, maturation, and modernization.

Leadership needs vary as teams grow and develop. In the early stages of development, most teams will have one or two defined leaders. At this point, directive leadership is required as most team members are inexperienced and therefore hesitant to get involved. Research has shown that teams without a formal leader perform worse than those with a defined leader (Farkas and Hill 2001). The leader's role at this stage is to help create a structure for the team, establish team norms, clarify tasks, provide vision, and challenge the team toward maximal performance (Heinemann and Zeiss 2002).

Once the roles of the team are established, the team leader's primary role changes to a consultant and source of inspiration for continued team growth. The leader also must continue to provide and arrange for the necessary internal and external support to keep the team functioning efficiently and effectively (Heinemann and Zeiss 2002).

44.3 Team Process

44.3.1 Decision Making

While the contributions and talents of the team members and the team leader are important to effective team functioning, the processes that the team members employ to interact with one another and to achieve team goals are equally predictive of team success (Marks et al. 2001). One of the most essential elements of the team process is the manner in which decisions are made. The goal of medical team decision making is to optimize patient outcomes in all aspects of patient care. Unfortunately, teams tend to focus on immediate solutions to problems, omitting the crucial steps of critical thinking and careful deliberation. This omission is compounded by groups' tendency to pursue the first suggestion proposed rather than continue to explore alternative solutions. Even when teams subsequently identify superior alternate solutions, they tend to support the first suggestion, especially if the "opinion leaders" in the group have endorsed it (Maier 1967). Consequently, the solutions recommended by the team are likely to be simplistic and may not be optimal (Farkas and Hill 2001).

Establishing a clearly defined method for making decisions can encourage teams to incorporate this important step. Before embarking on a discussion about a problem, the team must distinguish between the symptoms and the core of the problem. Patient care decisions must be explored from various surgical, speech, dental, social, and psychological perspectives. Once the problem is defined, the team must prioritize the issues involved. When dealing with medically complex patients, prioritization of medical problems is essential to comprehensive care. Similarly, identifying problems in craniofacial team functioning may be a complex process with numerous professional and interpersonal nuances. The team must carefully select an action plan with deadlines to avoid rehashing the problems without advancing to resolution.

Once the problem is identified, the team is ready to discuss possible solutions or treatment options. Here, the key is to provide an environment where team members feel comfortable sharing ideas without concern for criticism or repercussions. No idea should be dismissed without careful consideration. Even discussion of seemingly outlandish solutions may prompt creative solutions that may not have emerged otherwise.

When various possible solutions have been presented, the team must identify the most promising solutions in order to develop a team consensus. When consensus cannot be reached, teams must balance consensus with personal feelings and outcomes (Eisenhardt et al. 1997). Conflict will likely occur during this process. It is the responsibility of team members and especially their leader to direct the team toward positive resolution.

When all team members are involved in developing and fine-tuning the solution, commitment to that decision is more likely than when one team member conceives the solution and convinces the rest of the team to support it. However,

theoretical commitment is not sufficient to solve a problem. The team must specify the "who, what, where, and when" of the action plan and monitor it to its completion (Farkas and Hill 2001). The purpose of this process is to ensure an action plan endorsed by team members at the conclusion of the discussion.

The challenge of monitoring action plans regarding patient care is that many of the recommendations may involve outside providers who may or may not complete the plans as conceived by the team. Another issue is that the patient may not follow through with recommended interventions between team visits. The challenge for action plans related to the team's internal functioning (such as those improving team efficiency) is that patient care always comes first, limiting the opportunities to complete the action plan. Since improved team process can lead to more accurate and in-depth decision making, it is patient care that is ultimately impacted.

44.3.2 Team Roles

The roles that team members tend to assume also affect the team process. When team members' professional roles are clearly defined, patients' and team members' mutual trust and respect are fostered. Professional/formal roles for each member are typically defined by an individual's clinical expertise during the early stages of team development. The expectations for these formal roles include the successful completion of their discipline-specific responsibilities.

In addition to professional roles, there are informal roles that individuals tend to assume which often depend on personality, life experience, and institutional constraints. These informal roles include positive roles related to completing tasks and managing personal/social situations and negative roles that actually hinder team functioning (Benne and Sheats 1948). Most teams will have members who fulfill many of these roles. People tend to drift in and out of different roles depending on their own personalities, their position on the team (depending on the level of equality among team members), and the situation at hand. It is important that team members take on the positive team roles to ensure effective team process.

Task-focused roles include the "initiator/contributor" who proposes original ideas and different ways of approaching group problems, procedures, or goals. This person tends to initiate discussions and move the team into new areas of exploration. The "information seeker" requests clarification of comments in terms of their accuracy, seeks information relevant to the problem, and determines missing information that is needed before moving forward. The "information giver" provides factual information to the group and is seen as an authority. The "opinion seeker" seeks clarification of the values, attitudes, and opinions of group members and ensures different perspectives are provided. The "opinion giver" expresses his or her own opinions and beliefs about the subject being discussed. The "elaborator" takes other people's initial ideas and builds on them with examples, relevant facts, and data while considering their consequences. The "co-coordinator" identifies and explains the relationships between ideas. The "orienter" reviews and clarifies the group's position and summarizes what has been accomplished, explains where the group has digressed, and suggests how to refocus on the goal. The "evaluator/critic" evaluates proposals to assess the reasonableness, factuality, and manageability of a proposal. The "energizer" challenges and stimulates the group to take further action. The "procedural technician" facilitates group discussion by supplying logistical planning such as meeting location and required supplies. The "recorder" records information from team meetings to provide documentation of team meetings.

Personal and/or social roles include the "encourager" who praises, agrees with, and accepts contributions of others. The "harmonizer" mediates the differences between other members, attempts to reconcile disagreements, and relieves tension in conflict situations. The "compromiser" offers to change his or her position for the good of the group. The "gatekeeper/expediter" encourages or facilitates the participation of others. The "standard setter" sets limits on

acceptable individual behavior within the group. The "observer" provides feedback to the group regarding team functioning. The "follower" contributes little to group discussion and accepts whatever other team members' decision.

Dysfunctional or blocking roles include the "aggressor" who belittles and insults team members or their proposals. The "blocker" opposes every idea or opinion without making his or her own suggestions. The "recognition seeker" uses group meetings to draw attention to himself or herself through boasting about personal achievement. The "self-confessor" uses the audience opportunity to express personal feelings unrelated to the team. The "disrupter" distracts other people by telling jokes, playing pranks, or even reading unrelated material as a way to evade work. The "dominator" attempts to control the conversation and dictate team members' activities to manipulate the group or certain members of the group. The "help seeker" attempts elicit a "sympathy" response from other group members through expressions of insecurity, personal confusion, or excessive self-deprecation. The "special interest pleader" speaks for a specific small group, usually cloaking his or her own prejudices or biases in the stereotype which best fits his or her individual needs.

In general, most of the task-focused and personal/social team roles are necessary to team functioning and in fact enhance team performance. The dysfunctional/blocking roles, while familiar to many situations, actually detract from team performance. Redirection of team members who assume these roles is essential to establishing the foundations for a successful team. How people interact and relate to one another will determine how successful the team will be at achieving its mission (Benne and Sheats 1948).

44.3.3 Development of Team Collaboration Skills

The quality of the interaction between team members within their formal and informal roles is directly related to each of the team members' collaboration skills. The Joint Commission on Accreditation of Healthcare Organizations (JCAHO) requires that teams produce "jointly created and implemented treatment plans" (JCAHO 1996). Multidisciplinary teams (where each medical professional independently evaluates the patient and then each of the individual professionals reports results within his or her own specialty) do not always accomplish this goal. Rather, team members may feel that team meeting is not an effective use of their time. More successful team collaboration occurs when the lines between team disciplines are blurred (as in interdisciplinary and transdisciplinary teams) and the focus of team discussion is transferred from the individual specialist's recommendations to the needs of the patient in reference to the patient's quality of life (Bokhour 2006).

In order to attain this goal and effectively function in such a wide arena of care with so many variables affecting patient outcomes, team members must establish a number of skills specific to team collaboration. The taxonomy of collaboration has many elements. The most unifying element of any craniofacial team is the care of a core set patients and hopefully a strong desire to provide state of the art care. Unity, however, is not achieved by a common cause alone. Strong collaboration skills, working together rather than side by side, are necessary to ensure that craniofacial team outcomes surpass what each individual team member could accomplish working alone. True collaboration requires time, commitment to a common goal, and communication.

44.3.4 Communication

An essential element of collaboration and the team process is communication. The vitality of a team can be measured by the degree to which it develops an efficient way to internally communicate important information, a high level of enthusiasm and energy, a shared "event-driven" history, personal commitment, and impressive performance results (Katzenbach and Smith 1994). Research has shown that effective team communication leads to enhanced patient and family satisfaction, reduced length of hospital

stay, reduced costs, and improved diagnostic and prognostic abilities of health-care workers versus professionals working independently (Suter, 2009). The Joint Commission on Accreditation of Healthcare Organizations (JCAHO) established a goal in 2004 to improve the effectiveness of communication among caregivers in its National Patient Safety Goals (Eldridge and Revere 1995). A requirement was added in 2006 that health-care organizations implement a standardized approach for communication between professionals (Eldridge and Revere 1996). In 2007, it released a statement that communication failures were the main cause of patient harm in 65% of cases (Revere and Eldridge 2007). Singh et al. reported that 70% of teamwork failures stemmed from communication breakdowns and "handoff" errors (Singh et al. 2007).

The team meeting is the forum for most internal team communication. Many teams meet as a group in a round-table format on a regular basis (usually weekly or monthly, depending on the volume of patients at a particular center) to discuss evaluation results and plan patient care. Other teams, however, communicate electronically. In some cases, each team member will simply send their written evaluation to a team leader who collates the information into a "team" report. Written communication lacks important nonverbal cues, sometimes resulting in misunderstanding which can lead to a lack of openness, undermined trust, and compromised partnerships. Without the benefit of team discussion, true team collaboration cannot occur and patient care suffers.

During group discussions, dialogue should be open but managed. Managing effective communication during team meeting involves a series of steps including obtaining information, defining the problem, applying necessary criteria, discussing alternative and assessing all options, and developing a patient care plan or solution. Addressing each of these steps increases the probability that the team is communicating proficiently. Assigning a specific facilitator to ensure that each member of the team is attending to all of these steps can increase the amount of ownership and responsibility each member feels for the team decision (Cardona et al. 2000). The importance of this facilitator is reinforced by research that shows that groups tend to elect the first solution that is supported by the "opinion leaders" of the group, even when technically superior solutions are later presented (Maier 1967). As a result, the "opinion leaders" of a team may wait to endorse any solution until all of the solutions have been proposed in order to avoid compromising the decision-making process.

A specific behavior that hampers communication on a craniofacial team is the use of discipline-specific jargon during discussions with other team members and especially with patient families. Using vocabulary that is not shared by others hinders their comprehension and ability to incorporate the information into the team discussion. This can deter communication and represents a barrier to the inclusion of the patient and patient family as members of the team (Strauss and Ellis 1996). The patient's family plays an important role on the team, and communication issues affect their ability to fully contribute to, comprehend, and comply with the team goals and recommendations. The mode of communication between the patient, family, and the rest of the team is multifaceted. The patient and/or family typically communicates with each team member face-to-face during the patient's evaluation and have the opportunity to voice their agreement or concerns about potential patient recommendations.

On some teams, the patient and family may physically meet with the entire team to discuss recommendations. In this case, the patient/family has a second opportunity to weigh in on the plan for the patient, especially important since new recommendations often emerge through the team meeting. With tightening of hospital and clinic budgets, however, fewer teams are able to commit to the time required to meet with the family a second time. In an attempt to provide an opportunity for the patient/family to provide some feedback, some teams have a representative from the team meet with the family after team meeting. Unfortunately, team meeting days tend to be quite long for the patients and families, and a second meeting with the team often adds 1–2 more hours to their visit.

Whether or not the family meets with the team a second time for final recommendations, the family will typically receive a written summary of the team's recommendations. This document must serve as both a medical document of the plan of care for the patient and a formal communication of final recommendations between the team and the family. The choice of vocabulary (e.g., avoidance of medical jargon) and appropriate reading level are key to establishing effective communication with the family and other professionals involved with the patient's care. Poor communication often leads to poor patient follow-through with team recommendations.

One way to encourage open communication between the team and the patient/family is to establish a parent advisory council. These councils often contribute invaluable information from the parent perspective regarding the delivery system of the team (team visits), communication or lack of communication from the team, comprehension of the material the team provides to the parents, cost, time, and overall opinions of care delivery.

44.3.5 Mutual Trust and Respect

Mutual trust is the basic foundation of any relationship, no less so on a craniofacial team. Trust among team members is essential as it creates a safe environment in which every member feels free to contribute to the team discussion, thus improving team collaboration. In order for a craniofacial team to function adequately, interdisciplinary team equality and mutual respect must be established and actively maintained, as professional hierarchies and dominance that typically occur in medicine may suppress communication and result in submissive behavior from members of subordinate disciplines (Strauss and Broder 1985).

The length and stability of a team's relationship over time also creates the opportunity for individual rapport and mutual understanding to develop. This situation can be both positive and negative, however, in that long-standing teams may stagnate and resist change to their established approach to patient care and team functioning. Incorporating new and young professionals on craniofacial teams is crucial for longevity and continuity of care; however, the new professionals may feel intimidated and may not immediately be accepted. They may have "new" innovative ideas that may threaten the "this is the way we have always done it" philosophy. Seasoned professionals may also feel threatened and feel their position is in jeopardy. As stated above, the team needs to enhance mutual understanding and enable appreciation for each member's professional skills. Parity or equality is the ideal for each team to achieve. Equal participation and involvement ensures that each team member contributes to team decisions. Lack of participation may mean that strategic objectives are not owned, resulting in a risk of low morale and stifled innovation.

To develop respect among team members, each member must be assured of his or her own competence and that of each member of the team. The norms and standards of practice of other professions on the team should also be considered. Both of these elements require education. Interdisciplinary meetings such as the ACPA annual meeting provide each discipline with individual educational opportunities in their own specialty as well as in other team disciplines. Training medical students in team care skills is another means of instilling respect for team care. The Canadian Interprofessional Health Collaborative created the Interprofessional Education program (IPE) to train health-care students for future interprofessional teams. Students taught in this approach demonstrated respect and positive attitudes toward their colleagues and worked toward improving patient outcomes (Bridges et al. 2011). A well-functioning team possesses at least a basic knowledge of the breadth of its specialties and the skills necessary for collaborative practice (Bridges et al. 2011).

Once trust and respect are established, levels of team participation should be assessed. While team members' contributions should be as balanced as possible, several factors besides professional status affect the degree of engagement for each team member. Linguistic styles of team members, for example, may vary in regard to levels of directness, conversational turn-taking, rate

and tone of conversation, loudness, humor, etc. Cultural differences on the team may affect degree of eye contact, willingness to disagree with a perceived superior, or ability to interpret meanings of culture-specific idioms. Gender differences also play a role. Women tend to wait for acknowledgement prior to speaking, provide an opening for others to comment between their statements, and allow for longer pauses in the conversation before commenting. These tendencies may reduce the amount of women's participation in team meetings, resulting in the impression that women wield less authority on the team (Tannen 1995). Since, as previously mentioned, the recommendations that are offered repeatedly are the most likely to be implemented, team members whose participation is limited for any reason are at a disadvantage (Farkas and Hill 2001). These examples of participatory imbalance can lead to a loss of mutual trust and respect, missed opportunities for new ideas, frustration, and eventual loss of commitment to the team.

The development of protocols has been shown to be beneficial to define each member's roles, expectations, and responsibilities (Vargervik et al. 2009). The ultimate goal of these protocols is to clarify and help ensure that all aspects of patient care are addressed. Another benefit is the potential prevention of conflict on the team with such issues a "turf-wars" over which professional handles each aspect of care. When institutions have multiple surgical specialties represented on their teams, issues may arise as to which surgeon will perform specific procedures. Another example would be an overlap of services when registered nurses and speech-language pathologists both address feeding concerns. Protocols help define the aspects of patient care for which each discipline is responsible. These protocols or patient care maps help streamline the patient referral process and improve the consistency and quality of patient care (Wall 1998).

44.3.6 Creation of Team Goals

Every team needs goals which are not only measurable but also are attainable, results-focused, and which serve to foster teamwork and collaboration (Katzenbach and Smith 1994). A strong team mission should lend itself to the creation of measurable team goals. The goals of craniofacial teams should always be centered on the ultimate outcome of improved patient care. Striving for the best care of the patient, however, does not mean that all goals must directly involve the patient. Improved team functioning translates to improved patient care. Therefore, craniofacial team goals should also address internal team functioning as a precursor to improved patient care. Short-term goals should include a focus on small improvements that can be swiftly attained to provide for more frequent positive reinforcement for the team as a whole (Katzenbach and Smith 1994). With more positive team experiences, team members are more likely to show increased commitment to the team. As the team evaluates what works and what does not work when interacting with each other, teamwork skills will be developed and honed more rapidly. Working on goals that do not have high stakes associated with them (such as those that do not directly involve patient care) allows for the team to establish and develop the skills for collaboration in a less emotionally charged atmosphere. Building upon these short-term, low-status goals, teams should develop higher impact goals to improve patient care.

Developing goals is a process that is unique to each craniofacial team. Goals should focus on team functioning and patient care outcomes. Many teams focus solely on the patient care aspect of team care and do not directly address or monitor how the team functions when developing goals. As Nichols, DeFriese, and Malone state in *Team Performance in Health Care*, "Such beliefs devalue team process and result in unreasonable limits on the amount of time organizations permit teams to grow, develop, improve, and become collaborative working units. During periods of downsizing and cost containment, teams often fail to take the time to develop their process activities. This can result in inefficiency and reduced productivity of the team" (Nichols et al. 2002).

Finally, each craniofacial team must examine the expectations, rules, and regulations of the organization in which it functions. If the craniofacial team is independent, it must establish its

own organizational expectations. If not independent, teams may face differing expectations, rules, and regulations from different interdisciplinary units participating on the team and from the institutions in which they operate. There are also insurance company constraints with which to contend. Each state and/or county has regulatory demands and restrictions. It is imperative that the team investigates its financial support and services. Each of these areas must be addressed in order to develop a cohesive team that is primed to deliver efficient and effective patient care.

44.3.7 Managing Conflict

Functional conflicts are natural when people work together, and without these differences, teams can become complacent, eventually undermining team collaboration. Amason describes "cognitive" or "c-type conflict" versus "affective" or "a-type conflict" as a difference of focus. With cognitive conflict, the focus is on issue-related differences of opinion. With affective conflict, however, the focus shifts to disagreements which involve attacks on the person rather than the issue. Without c-type conflict, the team's decisions represent the decisions of the most vocal or the most influential members, undermining team effectiveness and diminishing the quality of the team's decisions (Amason et al. 1995). This situation leads to less than optimal patient care and poor outcomes, although as Strauss states, "conflict within a team and its deliberations may be natural, inevitable, and productive" (Strauss 1996). Productive conflict occurs when diverse perspectives are discussed and an optimized solution is developed. This solution represents a synthesis of ideas rather than a single person's standpoint. The absence of conflict may also be detrimental to team performance. The downfall of a team begins with inattention to team outcomes, poor accountability, lack of commitment, and the avoidance of conflict, all culminating in an absence of trust (Lencioni 2002). One of the most difficult and essential roles of the team leader is to manage conflict on the team to ensure that it is productive. Without a strong leader, healthy conflict, while inevitable and actually necessary, can deteriorate into personal attacks which have damaging effects on the team. Therefore, the team leader must encourage an environment where team members feel safe and supported so they are unafraid of taking part in constructive conflict to optimize patient care (Farkas and Hill 2001).

In order for a team to function optimally, team members must value individual team members' differences. This permits open discussion and examination of potential underlying assumptions without personal attack. When conflict becomes personal, conflict becomes negative and may call into question personal motives or even team members' professional competence. Avoiding conflict, however, often encourages "groupthink" where team members do not communicate their professional criticisms or concerns (Farkas and Hill 2001). This situation does not allow for diverse viewpoints to be heard and limits the scope of team discussion, ultimately affecting the quality of the team's decisions. On the other hand, well-managed encouragement of constructive conflict increases the likelihood of producing the best possible patient care plan (Margolis and Fiorelli 1984). Engaging in affable and professional group discussion on an equal basis increases the degree of team member understanding, commitment, and ownership of team decisions (Amason et al. 1995). Without strong "buy-in" regarding team decisions and/or patient recommendations, the team is at greater risk for individual members to subvert patient care plans for their own purposes, causing damage to team morale and potentially calling into question the merits of the team recommendations. Another possible outcome of suppressing conflict within teams could be future retaliation (conscious or subconscious) between team members (Farkas and Hill 2001).

Although cognitive conflict can have positive effects on care, affective conflict can cause dysfunction. Affective conflict disrupts a group from accomplishing its goals, and disagreements in a team can quickly turn into personal dislikes (Townsley 2011). It typically results in less

individual productivity and creates a cycle of negativity. This limits the creativity and growth of the team because the resulting combative environment deters members from making suggestions that differ from the status quo in order to avoid an escalation of the conflict (Phillips 2011).

Identifying the conduct and patterns that lead to dysfunction is the first step toward solving the problems. Multiple conflict management theories and management styles are available. The Thomas-Kilmann Conflict Mode Instrument is an example of an instrument that identifies which style team members tend to use during conflict situations to determine levels of assertiveness and cooperativeness. Responding to conflict with competition is considered assertive and uncooperative, accommodation is unassertive and cooperative, avoidance is unassertive and uncooperative, collaboration is assertive and cooperative, and compromise is moderately assertive and moderately cooperative (Thomas and Kilmann 1974). A balance between assertiveness and compromise sets the stage for positive conflict.

Once the different styles of conflict resolution are understood, the best approach to a given situation can be generated. The Interest-Based Relational (IBR) Approach is a method that respects individual differences while avoiding becoming too fixed on a specific position. The IBR Approach suggests that a moderator set forth the ground rules to promote respect for all team members by:

- Separating the people from the problems to preserve relationships
- Engaging in active listening
- Attending to body language
- Dissecting the conflict in objective terms
- Exploring possible solutions together with a "win-win" attitude
- Monitoring the group response to change, involving all members in the ongoing resolution
- Addressing individual responses to address negativity and ensure advancement toward the solution (Mind Tools 2011)

Involving all the members in every step helps each team member "own" the resolution to the conflict. If negotiations fail, the team may have to rely on an outside mediator to help facilitate communication and balance (Townsley 2011).

44.3.8 Team Values and Ethics

The impact of values and ethics on the team process also must be considered. Values are based on an individual's principles and priorities which are shaped by cultural mores and personal life experiences. Ethics are based on society's moral codes of conduct regarding right and wrong individual or group behavior within that society. When making decisions in the craniofacial team environment, the line between values and ethics is often blurred, making it difficult to identify the levels of influence of each of these forces. These dilemmas frequently occur in craniofacial centers when a team investigates how outside interests are impacting overall care. The team is obliged to consider a patient's family circumstances, finances, impact on family employment, insurance regulations, hospital requirements, and costs when recommending patient care.

For example, the speech-language pathologist on a craniofacial team recommends a secondary palatal surgery to correct a velopharyngeal incompetence (hypernasality), since his or her core value is to normalize speech. The primary goal for most surgeons on a craniofacial team is for aesthetics and functionality, so the surgeon also supports the surgical recommendation. The team's psychologist and social worker, however, recommend delaying surgery for psychosocial reasons such as excessive family stress and patient developmental delays that preclude the ability to perform the necessary postoperative instructions. Each of the team members in this situation comes to the table with a certain values focus (i.e., surgery will improve the patient's functionality and quality of speech vs. surgery will magnify the pressure on an already strained family unit). The team's ethical dilemma is to weigh each of these tenets to determine the best course of action for the patient's and family's overall quality of life.

Another situation that occurs with multifaceted and long-term patient care is when a patient does not have the resources to fund the care recommended by the team. After exhausting resources to assist the family, the team must struggle with how to provide the best possible care within the limits of the financial and/or social situation even if the intervention is not the "gold standard" or the most recently developed technique. This situation is often encountered when more recently developed tests or techniques are not yet covered by insurance companies.

A team may also be faced with a situation where they must decide whether or not to repair the palate of a child who is severely neurologically impaired. They must decide if the risk of surgery and the demands on the patient during the recovery process are warranted for a child who may never functionally communicate. With potentially conflicting values among team members (including the family), the team must evaluate the ethical implications of the situation to determine the most appropriate treatment plan.

Values/ethical dilemmas are challenging for craniofacial teams because each member hails from a unique professional and personal background. These diverse histories include specific ethical backgrounds leading to unique sets of values. Medical professionals, including those on craniofacial teams, deal with a common set of ethical issues such as truth telling, consent, capacity, confidentiality, conflict of interest, resource allocation, and research ethics on a daily basis (McKneally and Singer 2001). The team as a whole must realize that individual values and beliefs may not be congruent with those of other team members or those of the patients and must learn to discriminate between values and ethics. Encouraging education and discussions involving values and ethics enhances the ability of the team to collaboratively deal with challenges which will ultimately result in better team care of patients.

An example of an individual ethics training exercise is the Self-Assessment Guide which recommends that each team member actively engage in an ethical dialogue by privately answering a series of questions about a specific case (Doucet et al. 2001). The goal of this exercise is not to build character or instill virtue but to enhance the team's ability to effectively care for patients in a humanistic and holistic manner. The questions encourage the participant to identify his or her own position, compare individual values with professional objectives and obligations, relate personal positions to personal values and decide how to communicate these values to the team, weigh team members' recommendations and determine whether to alter those recommendations, identify strategies for listening to and speaking with team members with whom communication is difficult, and differentiate between a team member's position and personality (Doucet et al. 2001).

Preferably, craniofacial teams should participate in medical ethics training conducted on a multidisciplinary basis to avoid professional compartmentalization and to create a better understanding of the complementary roles of different professionals on the team (English 2004). This shared educational experience helps establish mutual respect, commitment, and involvement among craniofacial team members. Team ethics training is best taught in the clinic setting using case-based conferences that merge practical application with theoretical considerations. All members of the team should be vested and interested in an actual case to facilitate a useful discussion. To create a meaningful and active learning experience, each team member must "take a stand" and defend his or her position (McKneally and Singer 2001).

Ethics discussions should also be part of the standard team process. For health-care teams to address ethical issues, each profession must understand the ethical principles of their own discipline and acknowledge those of others. The influence of personal values must also be examined. Team members must acknowledge that each has underlying values that are brought to the table regardless of the fact that they are unspoken. Only then can ethical deliberation be facilitated. The team should develop a list of common cares and concerns that represent the team values (which should be evident in the team mission statement). This list should be developed through team discussion and mutual understanding.

In summary, the impact of values and ethics on team care is a constantly evolving. Issues involving ethics and values are inevitable in craniofacial team care. Team ethics training leads to improved communication and connections between team members and most importantly, improved patient care.

44.4 Assessment and Improvement of the Team Process

In the long term, teams are not measured by team members' interpersonal skills, communication skills, or the clarity of team roles, but rather they are evaluated and shaped by their performance (Katzenbach and Smith 1994). The ACPA has declared that team members are responsible for creating a plan of care that is "effective, efficient, and obtainable" (American Cleft Palate-Craniofacial Association 1993). The elements to a successful team include: joining a team by mutual choice, encouraging diversity, leading by example versus by commands, reinforcing a team's identity through shared experiences, encouraging creativity and reducing tension through humor and play, and holding periodic gatherings, meetings, and retreats to renew spirits and reinforce values (Bolman and Deal 1992). As teams go about the delivery of care on a daily, weekly, and/or monthly basis, they should be taking time to investigate their team functioning and outcomes. Positive team performance requires team members with collaboration skills which must be learned through observation, direct teaching, and "guided practice" (Cardona and Miller 2000). Research has shown that team members benefit from direct teaching of team process, effective communication, active listening, and conflict resolution (Farkas and Hill 2001). The lack of systematic and continuous evaluation of team collaboration may weaken team outcomes. In order to improve team performance and thus patient care, teams must continuously develop and examine their processes.

Several avenues exist for examining team process. Team performance may be assessed internally through regularly scheduled meetings focused on examining the team's positive and negative aspects. A different perspective can be gained through feedback from an external consultant who can delineate the team's strengths and weaknesses and assist the team in creating a plan for improvement. Alternatively, teams can use quantitative instruments such as questionnaires to assess team members' perceptions of current team functioning and generate ideas for improvement. These questionnaires may be generated by the team or one of the many published instruments may be utilized. The questionnaire should be applied to each aspect of the craniofacial team's functioning including the mission statement, team composition, team visit, team meeting, team reports, patient follow-up, research, marketing, and finances. These instruments vary in their degree of focus on one or more of the parameters of team performance and in their length and time commitment. Most of these measures help pinpoint teams' strengths and weaknesses, some assist in the development of action responses to the areas of weakness, and some assess the subsequent impact of those responses.

Regardless of the type of team setting for which any measure was created, there are universal parameters of team functioning that can be measured by common instruments. These instruments measure aspects of team structure, team context, and degree of interdependence. However, team productivity and the organization in which the team functions often are not included (Brallier and Tsukuda 2002).

Most of these measures were developed for specific types of teams, some medical and some business, but none were developed specifically for craniofacial teams. To meet this need, the UNC Craniofacial Team Peer Review questionnaire was developed at the University of North Carolina-Chapel Hill to measure and address craniofacial team functioning (see Appendix). Since its inception in 2008, this survey has been piloted in several craniofacial centers in the United States and abroad in order to refine the process and increase its universality. Pilot teams reported measurable positive outcomes from this process.

Once a team assessment has been performed, a summary of the information should be gathered, analyzed, and shared honestly with the team. Both the strengths and weaknesses of the team should be discussed, and goals for improvement should be developed. A follow-up retreat is an excellent venue to address concerns and develop a plan for remediation. Retreats provide a distraction-free environment in which the only concern is the task at hand. During a retreat, a specific agenda should be created and distributed to maintain focus. Participants should be coached to be "active listeners" by restating, rephrasing, or summarizing what the speaker has stated to ensure complete understanding. "Brainstorming" activities may be used to encourage ideas and allow for gathering all members' viewpoints. These activities should be chosen based on the group dynamics and the objectives of the gathering.

As a result of the brainstorming session, the team next explores options together to come to a common decision and to make a list of possible solutions. When a solution has been determined, a group leader must then monitor the group behavior in response to change. Some people are threatened by change whereas others are willing to take risks and are more apt to change (Wall and Proyect 1998). Leaders may use the plan-do-check-act (PDCA) cycle to demonstrate to the team that changes will be tried and evaluated for effectiveness or flaws prior to implementation. This process may reduce anxiety related to change (Arveson 1998).

During the evaluation phase of the proposed changes, the team should be invited to give feedback. The team leader should periodically remind the group that change is a process and not a single event. Once possible solutions are identified, individual or small group responsibilities with a time line for completion should be assigned. Each member should be involved in the resolution phase. It is important to verify that team members are satisfied with the process and are still working toward a solution (Wall and Proyect 1998). A monthly or quarterly meeting may be warranted in order to carry out teamwork training which may include lectures, videos, role playing, and case-based training.

44.5 Summary and Conclusions

Craniofacial teams represent a complex, continually evolving challenge from their inception that requires constant attention to both internal and external processes. The importance of the external team processes of patient satisfaction, patient outcomes, research, funding, and the team's reputation has been clearly established. These processes involve the output of the team. Most teams are required to attend to these areas because their home institutions (hospital, clinic, university) demand some type of formal or informal accounting of these areas as a measure of the team's productivity or to comply with JCAHO standards.

In order to sustain efficient and effective teams, craniofacial teams must establish a strong team foundation through attention to the internal team process to allow for complete team formation, create a compelling team mission, ensure the appropriate team composition, and secure appropriate team leadership. The team must continue to develop and progress in order to maintain and improve upon team functioning. This can be accomplished by direct focus on and training in the elements of team process which include decision making, identifying team roles, developing collaboration skills, improving communication skills, fostering mutual respect, creating team goals, managing conflict, and evaluating values and ethics. These actions require skills that must be constantly evaluated and often require direct instruction for team members.

Monitoring the team process and providing team skills training promote improved team functioning and, subsequently, improved patient outcomes. It takes a team to deliver comprehensive craniofacial care, but it also takes work to keep a team functioning well.

44.6 Appendix

44.6.1 UNC Craniofacial Team Peer Review and Self-Assessment

Please rate each question as: Never (1), Occasionally (2), Sometimes (3), Most of the Time (4), or Always (5)

Please include comments whenever possible

*Denotes questions adapted from ACPA Standards of Approval of Cleft Palate and Craniofacial Teams

** Denotes questions adapted from ACPA Standards of Approval specific to Craniofacial Teams

44.6.1.1 Criterion One: Mission (Insert Team's Mission Statement)

Do you feel that this mission continues to describe the purpose of our team?

Do you feel that this mission describes your purpose as a member of the team?

Do you feel that this mission should be updated?

Levels of Support

Do you feel supported in this mission by the craniofacial center staff?

Do you feel supported in this mission by other team members?

Do you feel supported in this mission by the team director?

Do you feel supported in this mission by your department?

Do you feel supported in this mission by our institution?

44.6.1.2 Criterion Two: Team Composition

*Does our team's patient care coordinator facilitate the operation of the team?

*Are the roles and responsibilities of the coordinator clearly identified?

*Does our team include a speech-language pathologist, a surgeon, and an orthodontist?

*Are all of our team members qualified by virtue of their education, experience, and credentials to provide appropriate care?

*Have each of our team members maintained current knowledge for best practice in their fields?

*Does our team have access to professionals in psychology, social work, psychiatry, audiology, genetics, general and pediatric dentistry, otolaryngology, and pediatrics/primary care?

**Does our team include a surgeon trained in cranio-maxillofacial surgery?

**Does our team have access to a psychologist who does neurodevelopmental and cognitive assessment?

**Does our team have access to a neurosurgeon, ophthalmologist, radiologist, and geneticist?

44.6.1.3 Criterion Three: Patient Care During Craniofacial Team Visit

Is our current system of referral to the craniofacial team adequate (from within our institution and outside referrals)?

Are the appropriate documents available to you when you need them for the patient's visit?

Do you feel our team receives all the necessary background information on new patients?

*Does our coordinator ensure the provision of coordinated care for patients and families/caregivers?

*Does our coordinator assist patients and families/caregivers with understanding, coordinating, and implementing treatment plans?

Is clinic flow efficient?

Are we ensuring that all patients are seeing all the specialists they need at each team visit?

*Are we providing team care and interventions in a sequence that addresses the patient's overall developmental, medical, and psychological needs?

Is the current way of scheduling patients to specialists on team days efficient and adequate?

Are you able to see your patients during your assigned window of time on team mornings?

Are you affected by "bottlenecking" of patients on team mornings?

Do you feel we schedule the appropriate number of patients on team mornings?

Is our physical setup for patient evaluation optimal?

Should we consider having multiple specialists see patients at one time?

Are we efficient with our direct patient care time?

Do you feel you have enough time with each patient on team visit mornings?

Do you feel there are redundancies across specialties?

Do you feel we have adequate quality control of our patient services?

Do we provide adequate continuity of care between patient visits?

*Are we evaluating our patients at regular intervals and in keeping with team recommendations?

*Are we encouraging patient and family/caregiver participation in treatment decisions?

*Are we assisting patients/families/caregivers with locating resources for financial assistance necessary to meet the needs of each patient?

*Does our team demonstrate sensitivity and flexibility of care to accommodate linguistic, cultural, and ethnic diversity among patients and their families/caregivers (including interpreters for verbal and written communication)?

*Does our team treat patients and families/caregivers in a nondiscriminatory manner?

* Is our team providing assessments for cognitive development and learning disabilities at appropriate time intervals?

Levels of Support for Our Team Visit Process

Do you feel supported on team clinic days by the craniofacial center staff?

Do you feel supported on team clinic days by other team members?

Do you feel supported on team clinic days by the team director?

Do you feel supported on team clinic days by your department?

Do you feel supported on team clinic days by our institution?

Overall Evaluation of Our Team Visit Process

What is the best aspect of our team visit process?

Which area of improvement do you feel is most needed for our team visit process?

44.6.1.4 Criterion Four: Team Meeting

Do you feel our members show a strong commitment to team care?

*Does our team meet regularly with at least our core team members?

*Is our mechanism for achieving consensus among team members effective?

Are you able to consistently attend team meeting for the entire meeting?

Do you feel we have all the necessary specialists in attendance at team meetings for each patient?

Do you feel that the way surgeries are presently assigned works well?

Are we efficient in our team meeting presentations?

Does our current team structure function well?

Do we have sufficient information at team meeting regarding the patient's history and follow-through with past recommendations?

Is our team leader structure functioning well?

Does our team identify all of the relevant medical, social, and psychological information when defining a problem?

Does our team spend sufficient time to explore all the treatment options?

Does our team explore enough different options when discussing treatment?

Does our team tend to propose the same solution for a majority of its patients?

Does our team spend sufficient time projecting the likely outcomes of treatment options?

Does our team assign the appropriate priority to the various factors influencing decisions?

Does our team tend to bias treatment plans based on the influence of one individual or function?

Levels of Support for Our Team Meeting

Do you feel supported for team meeting by our center staff?

Do you feel supported for team meeting by other team members?

Do you feel supported for team meeting by the team director?

Do you feel supported for team meeting by your department?

Do you feel supported for team meeting by the school of dentistry?

Overall Evaluation of Our Team Meeting

What is the best aspect of our team meeting?

Which area of improvement do you feel is most needed for our team meeting?

44.6.1.5 Criterion Five: Team Reports

*Does our team maintain central and shared records?

Are we sufficiently integrating our individual findings into cohesive recommendations?

Do you feel our reports are readable to patients and professionals?

Do you feel we should prioritize recommendations?

Do we communicate effectively with our families?

Do you feel our reports are accurate?

*Do our reports include the recommended treatment plan and alternatives, benefits, and risk factors?

**Are the results of neurodevelopmental and cognitive assessments included in our team report?

**Are the participation of neurosurgery, ophthalmology, radiology, and genetics included in our team report as appropriate?

Levels of Support for Our Team Report

Do you feel supported on team reports by the craniofacial center staff?

Do you feel supported on team reports by other team members?

Do you feel supported on team reports by the team director?

Do you feel supported on team reports by your department?

Do you feel supported on team reports by our institution?

Overall Evaluation of Our Team Report Process

What is the best aspect of our team report process?

Which area of improvement do you feel is most needed for our team report process?

44.6.1.6 Criterion Six: Patient Care and Service Beyond the Team Visit

*Is our process for referring patients to outside agencies adequate?

Do we adequately follow up with patients?

Do we have enough coordination of interdepartmental appointments between team visits?

Do you feel patients are "falling between the cracks" for follow-up appointments?

*Do we have an adequate system for obtaining outside records of these follow-ups?

*Do we have an efficient system for sending information to outside agencies?

*Do we have an adequate system for obtaining informed consent?

*Is our team's process for documenting treatment outcomes adequate?

*Is our team conducting periodic retrospective or prospective studies to evaluate treatment outcomes?

*Is our team's quality management system adequate?

Levels of Support for Patient Care Outside the Team Visit

Do you feel supported on patient care that occurs outside team visit by the craniofacial center staff?

Do you feel supported on patient care that occurs outside team visit by other team members?

Do you feel supported on patient care that occurs outside team visit by the team director?

Do you feel supported on patient care that occurs outside team visit by your department?

Do you feel supported on patient care that occurs outside team visit by our institution?

Overall Evaluation of Our Patient Care That Occurs Outside Team Visits

What is the best aspect of our patient care that occurs outside team visit?

Which area of improvement do you feel is most needed for our patient care that occurs outside team visit?

44.6.1.7 Criterion Seven: Research

Do you feel that we are completing enough research through our team?

Do you feel that all patients be in some clinical trial?

Do you feel that we need a database for clinical research?

Levels of Support for Research

Do you feel supported for research by the craniofacial center staff?

Do you feel supported for research by other team members?

Do you feel supported for research by the team director?

Do you feel supported for research by your department?

Do you feel supported for research by our institution?

Overall Evaluation of Craniofacial Team Research

What is the best aspect of our ongoing research?

Which area of improvement do you feel is most needed for our research?

44.6.1.8 Criterion Eight: Financial Health

Do you feel we are adequately funded?

Do we care for our unfunded patients well?

Do you feel we need to research other funding avenues?

Do you feel we should be increasing fundraising or lobbying?

Levels of Support for Funding

Do you feel supported for funding by the craniofacial center staff?

Do you feel supported for funding by other team members?

Do you feel supported for funding by the team director?

Do you feel supported for funding by your department?

Do you feel supported for funding by our institution?

Overall Evaluation of Funding

What is the best aspect of our current funding?

Which area of improvement do you feel is most needed for our current funding?

44.6.1.9 Criterion Nine: Preparing for the Future: Public Relations/ Awareness/"Exposure"

Do you feel our team needs to increase its public recognition? Local? Regional? National? International? Internet?

Levels of Support for Public Relations

Do you feel supported for public relations by the craniofacial center staff?

Do you feel supported for public relations by other team members?

Do you feel supported for public relations by the team director?

Do you feel supported for public relations by your department?

Do you feel supported for public relations by our institution?

Overall Evaluation of Public Relations

What is the best aspect of our team's public relations?

Which area of improvement do you feel is most needed for our team's public relations?

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Living with a Cleft: Psychological Challenges, Support and Intervention

45

Nichola Rumsey and Nicola Marie Stock

45.1 Introduction

A cleft of the lip and/or the palate and its treatment can pose challenges in many domains of life, including several areas of psychological and social functioning. To date, research exploring the psychosocial ramifications of clefting has produced complex and contrasting results. A recent literature review by Stock and Rumsey (2011) highlighted some emerging areas of consensus, alongside a number of methodological challenges for researchers in the field. Using a lifespan perspective, this chapter summarises current knowledge and outlines the potential of these findings to inform appropriate psychosocial support and interventions for patients with cleft and their families.

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45.2 What Are the Challenges?

45.2.1 Infancy (0–3 Years)

In countries with well-developed healthcare resources, the majority of clefts are now diagnosed antenatally, using 2D or 3D scanning techniques. The availability and accuracy of these scans can vary, however, and some clefts are still not diagnosed until after birth. A cleft in the palate is particularly difficult to detect and, if a thorough postnatal examination is not carried out, may remain undiagnosed for some time (Slator et al. 2009).

For most parents, the 'diagnostic event' is an emotionally demanding experience, which can invoke a simultaneous mix of shock, despair, guilt and grief (Vanz and Ribeiro 2011). Non-specialist healthcare professionals (HPs) are usually the first point of contact for parents, but often lack the specific knowledge and skills needed to support the family appropriately (Knapke et al. 2010). The most common concerns expressed by parents are: 'what caused the cleft – why has this happened to us?' and 'what are the future prospects for our child – will she/he be alright?' (see Williams et al. 2012). Parents may also be concerned about the possibility of having another child with a cleft (Stock and Rumsey, in preparation). The answers to these questions are complex and inconclusive, and parents can be frustrated by the inability of HPs to provide them definitive answers, either to these questions or in relation to the type and length of the treatment that will be required (Nelson et al. 2012). The quality of information received by parents, and

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the manner in which this information is delivered, appears to have a significant impact on the distress experienced and the ability of parents to cope (Chuacharoen et al. 2009; Vanz and Ribeiro 2011). Although a high level of overall satisfaction with early healthcare is frequently reported (Nelson et al. 2012), many parents would like more opportunities to talk to HPs in order to have their anxieties allayed (Hodgkinson et al. 2005).

Issues associated with feeding are also particularly concerning for parents. As infants with cleft can have difficulty attaching and sucking (Reid et al. 2006), attempts to breastfeed can be frustrating. Mothers have reported that feeding difficulties can affect their feelings of competency and interfere with the intimate mother-infant bond during the first few days after birth (Stock et al. 2011).

The experience of these first crucial weeks can impact significantly on longer term parental well-being. For example, Despars and colleagues (2011) reported that mothers of infants with a cleft frequently have less secure representations of their relationship with their child, have higher maternal emotional involvement and have more posttraumatic stress symptoms than controls. However, it is important to view these findings in context. The factors likely to mediate the psychosocial impact on the family and the affected child in the first few years include socioeconomic resources (Waylen and Stewart-Brown 2009), family attachment history, parental coping styles and strategies (Hodgkinson et al. 2005), mental well-being (Pope et al. 2005), beliefs about causation and responsibility (Nelson et al. 2009), child temperament and child co-morbidity (Hodgkinson et al. 2005). Parents have also reported encountering negative reactions to their child's cleft from friends, family members, HPs and members of the public. This can result in considerable distress, leading parents to conceal the cleft in different ways and, in some cases, to become socially withdrawn (Nelson et al. 2012). The way in which the family perceives the cleft and the emphasis the family places on appearance may also be important factors in their own and their child's adjustment (Bellew 2012).

Much of the research relating to the early years has focused on mother-infant attachment, reflecting an underlying assumption that this process would be negatively affected by the presence of a cleft lip and/or feeding difficulties. In reality, recent studies have found no overall differences in the rates of attachment security between mothers and infants with cleft compared with unaffected controls (Collett and Speltz 2007). However, infants with a cleft have been found to have higher rates of secure attachment early on, with those with the most objectively severe impairments in facial attractiveness demonstrating the highest rates. Collett and Speltz (2006) concluded that infants with a cleft may be perceived by their mothers to be particularly vulnerable, heightening maternal responsiveness and resulting in stronger attachment characteristics. Other factors, including the quality of the care provided, experiences of feeding and parental well-being, have also been shown to be contributory factors to the development of attachment security (see Cassidy and Shaver 2010). Less satisfactory mother-infant relationships have been associated with inadequate nutritional intake (Hodgkinson et al. 2005), potential deficits in language development (Wermke et al. 2011) and poor cognitive functioning at 18 months (Murray et al. 2008).

45.2.2 Childhood (4–11 Years)

Early childhood is a key period in terms of social and educational experiences. It is often the first phase of life in which a child will be expected to integrate with their peers in the world beyond the family environment. Having a visible difference may represent an immediate challenge, as other children may be curious, may be unsure of how to behave or may make unhelpful comments (Rumsey 2002). Similarly, children with hearing or speech complications may be more difficult to understand, provoking a higher risk of rejection by their peers. How the child experiences and copes with these challenges is likely to impact on the development of their self-worth and personal sense of agency (Hearst et al. 2008).

A considerable body of research has identified children with a cleft to be more socially inhibited than their peers. Reports of gaze avoidance (Slifer

et al. 2006), social anxiety (Murray et al. 2010), withdrawn-depressed behaviour (Pinquart and Shen 2010) and internalising problems (Pope and Snyder 2005) are common amongst children with a cleft. It may be that while many of these children have adequate conversational skills, they are less responsive and less assertive than their peers (Frederickson et al. 2006). Teasing and bullying, if present, is likely to peak around the age of 7 years. Increased levels of perceived peer harassment are associated with increased dissatisfaction with appearance (Billaud Feragen and Borge 2010) and patterns of learned helplessness (Hearst et al. 2008).

Although children with a cleft score similarly to their peers in terms of overall quality of life (Sagheri et al. 2009; Wehby and Cassell 2010), children with palatal involvement have been reported to have lower health status and lower health-related quality of life (Damiano et al. 2006). This may be related to increased problems with speech (Damiano et al. 2006) and social relationships (Kramer et al. 2009) in this group.

The possibility of cognitive delay in children born with a cleft is currently a topic of debate. The majority of research to date has found cognitive development to be within the normal range (Collett and Speltz 2007); however, Richman and colleagues (2005) noted an elevated incidence of learning disabilities amongst children with cleft, identifying specific weaknesses in relation to visual memory and reading ability. Verbal intelligence may be related to poor maternal sensitivity during infancy (Hentges et al. 2011), while low academic achievement may be attributed to hearing and communication difficulties (Hearst et al. 2008). Due to a lack of knowledge and skill, teachers may interpret these difficulties as behavioural problems in the child (Stock et al. 2011).

45.2.3 Adolescence (12–17 Years)

It is understood that adolescence can be a particularly difficult time for young people with a cleft, as differences in appearance and/or speech may affect their ability to 'blend in' and be perceived as 'normal', a central facet of well-being in adolescence (Liossi 2003).

Overall, research has found little evidence of impaired psychosocial adjustment in adolescents with a cleft (e.g. Berger and Dalton 2009; Locker et al. 2005). However, a number of studies have identified variables which may moderate adjustment for this age group, all of which relate to social experiences, speech difficulties and dissatisfaction with appearance (e.g. Berger and Dalton 2011; Bilboul et al. 2006). Perceived teasing and bullying appear to be particularly influential, impacting on self-confidence (Noor and Musa 2007), subjective ratings of appearance and emotional distress (Billaud Feragen et al. 2010). The extent to which the young person feels in control of their treatment may also affect psychological adjustment. Young people may have unrealistic expectations of aesthetic change following surgery (Cadogan and Bennun 2011) or may disagree with parents or HPs about the point at which to end surgical or orthodontic treatment (Collett and Speltz 2007). In addition, various changes to facial aesthetics during these years may impact on adjustment (Cadogan and Bennun 2011).

45.2.4 Adults (18+ Years)

The transition into adulthood usually signifies the end of the care pathway, and the majority of patients lose touch with the services that have been a regular part of their lives until this point.

On average, young adults with a cleft report lower quality of life than their peers, except in the domain of family relationships (Collett and Speltz 2007). It is possible that delays in the emancipation process from the family unit may contribute to observations that adults with cleft appear to be less likely to marry, or marry later in life (Danino et al. 2005), and are more likely to be childless or wait longer to raise a family (Yttri et al. 2011). There is also the additional concern that future generations may be affected due to the genetic element involved in cleft (Williams et al. 2012). A recent retrospective population-based study by Persson (2012) was also strongly suggestive of negative long-term consequences of a cleft on physical development and educational achievement. Most males in resource-rich countries have a preference for being tall and muscular

(Tiggemann et al. 2008). If men are dissatisfied with their bodies through being unable to match up to current stereotypes related to height and muscularity, there are associated psychological risks, including lower self-esteem, social anxiety and depression. Deficits in educational achievements were also apparent in all groups. Lower overall grade point averages and, more specifically, lower scores in maths and languages were most marked for those with cleft palate only (CPO).

Many adults report being dissatisfied with surgical outcomes and express a desire for further corrective treatment, which is often related to poorer health-related quality of life outcomes and long-term well-being (Oosterkamp et al. 2007; Sinko et al. 2005). Research has also found large discrepancies in ratings of aesthetic satisfaction between the patients themselves, HPs and uninvolved judges (e.g. Foo et al. 2011).

45.3 Individual Differences in Adjustment

The literature has demonstrated that a cleft and its treatment present a number of challenges. While some experience a range of transitory or longer term difficulties, others thrive. Differences in adjustment are not well explained by the objective severity or the visibility of a cleft (Appearance Research Collaboration 2009). Understanding the multifaceted nature of adjustment and identifying those who may be vulnerable are key to the provision of effective psychological intervention. If we are to move forwards, a number of missing links in our understanding of cleft need to be addressed, and a number of key methodological concerns need to be resolved.

45.4 Missing Links in Understanding Psychological Adjustment

The majority of research to date has focussed on children and teenagers. This is understandable, as these populations are more easily accessed by researchers and clinicians during regular treatment visits. Our knowledge of longer term outcomes is more limited as research involving people who are geographically dispersed and no longer receiving regular treatment is challenging. There may be psychological and social challenges that have not been recognised or provided for by cleft teams. In addition, very little research has been conducted with older adults with a cleft. Although research has shown the importance of physical appearance to decline with age, body dissatisfaction has been found to be stable across much of the adult lifespan (Tiggemann 2004). The additional impact of a cleft on this phenomenon and its wider consequences are not yet known.

The surprisingly high rate of impaired cognitive functioning in individuals with a cleft and the emergence of evidence of negative long-term outcomes have driven researchers to examine the intricacies of the interrelationships between the development of the face and brain – specifically, the implications of these relationships for the brain structure and function for those born with a cleft. Underlying differences in brain structure may mediate deficits in reading ability and language development, as well as aspects of behaviour and social functioning. Findings from the Nopoulos group based in Iowa are suggestive of anomalies in brain development which are associated with physical, behavioural, cognitive and academic outcomes in children and adults born with a cleft (e.g. Richman et al. 2011). These and other studies suggest that additional input and support may be necessary to achieve optimal educational outcomes and that particular attention should be paid to signs of developmental delay in those with CPO (Persson 2012).

A number of 'hard-to-reach' groups have thus far been severely under-represented in cleft research. There is a heavy bias towards the experiences of mothers of children with a cleft, with few attempts to involve fathers in research. The importance of the role of the father in normative developmental processes (Lamb 2010) and in overall familial adjustment should not be overlooked. Researchers should also document the effects on siblings, grandparents and the family unit as a whole. More attention should be devoted to understanding the family's needs at different stages of the affected child's development (Nelson et al.

2012), with the aim of developing sources of support and intervention designed to facilitate positive adjustment in the affected child and to help the family to turn the challenges of living with a medical condition into a strengthening experience (Feragen 2012; Rumsey and Harcourt 2005).

The vast majority of research to date has been conducted in resource rich countries. People from minority groups are under-represented in this research, as are populations in less developed countries. There is an urgent need to further understand the influence of race, culture and social grouping on adjustment to cleft. We also need to focus attention on the needs of children and adults from resource poor countries with unrepaired clefts and the rehabilitation needs of those with clefts which are repaired in later childhood or adulthood.

The impact of additional anomalies or conditions (such as ADHD or autism) for the child with a cleft is also poorly understood. Increasing numbers of syndromes associated with clefting are being identified, but these are difficult to diagnose. For reasons of experimental control, the majority of clinical trials have been limited in focus to those with unilateral CL/P. The specific needs of other groups of people with cleft should become a focal point for future research.

Many researchers in the field work in the silos of their respective disciplines and an understanding of the interactions between the various aspects of life affected by a cleft are limited – for example, what are the interrelationships between speech outcomes and psychosocial adjustment? Additionally, researchers face the task of unpicking the extent to which challenges and responses identified in people affected by cleft also pertain to the general population, and which are specific to, or exacerbated by the cleft and its treatment. We know, for example, that almost all children are teased and that a proportion are distressed by teasing, whereas others take this in their stride. A visible or audible characteristic such as a scar or speech anomaly can be a target for teasing, and indeed, there is evidence to show that a percentage of children with a cleft are distressed by teasing. Would these have been the same children who would have been distressed by teasing even if they did not have a cleft? In current society, media messages consistently link physical attractiveness with happiness and success. Is the impact of this worse for those with a visible difference such as repaired cleft lip than for those without a cleft? Our approach (building on the work of Lansdown et al. 1997) is to conceptualise the cleft and its treatment as an underlying stressor throughout life – one which makes continuous calls on energy reserves and coping resources. This may compound the impact of 'normal' developmental stages, life events and specific stressors to cause distress. The cleft may also become the focus of distress resulting from other sources – a 'weak link' in the psychological armoury.

45.5 Methodological Challenges

Many of the conflicting results of research relating to the psychosocial adjustment to cleft lip and palate can be attributed to differences and limitations in the type of methodology used. In particular, there is a lack of consensus about the constituents of a positive psychosocial outcome. With so many psychological constructs identified as playing a part in adjustment, it is a major challenge to achieve agreement within and between clinicians and researchers about the key factors to measure and how they should be measured. Disagreement in this area, as well as the unsystematic or arbitrary operationalisation of constructs, leads to wide variability in the use and application of measures. This limits the conclusions that can be drawn. In addition, the challenge of recruiting sufficiently large numbers of participants to research studies in this field is widely acknowledged. Multidisciplinary and multicentre approaches to audit and research are necessary, but far from straightforward to achieve. Furthermore, it is tempting to dichotomise people with cleft as negative or positive adjusters and to assume that levels of adjustment are stable, yet there are indications from recent research that fluctuations between states of relative adjustment and distress are common. Longitudinal research is urgently needed to shed further light on the

complexities of stability and change in adjustment over time.

The recent growth in the number of qualitative and mixed methods studies has added to the richness of our understanding of individual experiences, as have efforts to involve service users in the research process. More research of this type is needed. The incorporation of qualitative work into quantitative designs has the potential to promote a more patient-centred focus in cleft research (Nelson et al. 2009; Rumsey and Harcourt 2005). In addition, the close concordance in the psychological factors contributing to adjustment in adults and young people with a variety of conditions affecting physical appearance has been demonstrated, increasing confidence in the extrapolation of findings from cross-condition studies to cleft (Appearance Research Collaboration, 2009).

45.6 Support and Intervention

The good news is that many of the psychological factors recently identified as contributing to adjustment to cleft are amenable to change. The major contributors are cognitive factors, which determine the lens through which a person views the world and the way people interpret the information around them. For adults, the psychological characteristics differentiating those experiencing higher levels of distress from those who are positively adjusted include levels of optimism, concern about negative evaluations by others and feelings of social acceptance (Rumsey 2012). A great deal of work is necessary to translate these findings into practice and additionally, the challenge of achieving a consensus on the content and most appropriate timing of support and intervention remains. Recent systematic reviews (Bessell and Moss 2007; Jenkinson et al. in preparation) of interventions for young people and adults with disfigurements resulting from a variety of causes have highlighted that few studies exist, and in those that do, findings are based on small sample sizes and study designs are not robust. As the most common difficulties relate to negative self-perceptions and difficulties in social encounters with others, approaches based on cognitive behavioural therapy (CBT) and social interaction skills are favoured by many, and the systematic reviews indicate these may hold promise. Kapp-Simon (1995), for example, showed gains in the frequency of initiation of conversations and the length of interactions as well as the frequency of positive interactions between young people with a cleft and their peers following social interactions skills training.

With their regular contacts with patients and their families, all HPs involved in cleft care have a vital role to play in promoting positive psychological adjustment. Currently, patients are offered a variety of appearance and/or function enhancing surgical or medical treatments. While biomedical interventions can offer benefits, they rarely address all of the psychosocial needs of patients. Provided in isolation, they may reinforce the notion that the way to a better quality of life lies through improved appearance – yet research has demonstrated clearly that a variety of psychological and social attributes are also crucial for positive adjustment. In their enthusiasm to further correct 'faults' in appearance resulting from the cleft, HPs should resist the temptation to in any way pressurise patients to undergo further treatment if the patients feel content with their current appearance – or to imply that in further correction lies the way to better adjustment. If the patient and family have unrealistic expectations of the gains in quality of life that might be achieved following aesthetic surgery, the potential for disappointment is high.

Ideally, a range of interventions based on the type and intensity of need should be developed. These should be designed to achieve an agreed set of outcomes, and a common set of measures should be used to assess these outcomes. These interventions should be delivered using a stepped approach in which all members of the team are involved in delivering basic psychosocial care, including routine questions and straightforward advice for common problems. More intensive, individually tailored face-to-face interventions

Table 45.1 A stepped approach to the delivery of psychosocial care

Level 5	Psychosocial specialist with appropriate experience	Coordination and supervision of psychological care Complex interventions
Level 4	Clinical or health psychologists	Preparation for surgical interventions and post-operative support Other 'routine' interventions
Level 3	Trainee psychologists	Screening for psychological need/distress Routine audit
Level 2	Team members with additional training	Identification of psychological need in patients and families Provision of information and support for families
Level 1	All cleft team members	Support and reassurance Delivery of psychological care agenda Consideration of psychological factors in treatment decision-making

would be delivered by appropriately trained psychosocial specialists (see Table 45.1).

Some lay-led organisations have developed campaigns and interventions designed to reduce societal pressures on those with a visible and/or audible 'difference' (see, e.g. Changing Faces: www.changingfaces.org). These have included 'camps' and workshops for young people with cleft and other visible anomalies designed to offer a safe and positive environment in which self-confidence, self-esteem, peer support and practice in relationship skills can be fostered (see, e.g. Changing Faces, The Cleft Lip and Palate Association and About Face; Tiemens et al. 2006). The impact of these activities remains to be fully evaluated.

Although slow progress towards the comprehensive provision of psychological care is being made, these resources remain the exception rather than the rule, particularly for adults and for patients and families who do not have regular access to care teams. In exploring the potential of other ways of delivering support, online interventions are showing great promise (see, e.g. Face IT, designed for adults (www.faceitonline.org.uk) and YP Face IT (www.ypfaceit.co.uk)). These interventions could be supervised by a cleft team member with appropriate training (Level 2 or 3) or, in suitable cases, self-administered.

Conclusions

While many people affected by a cleft are well adjusted, this population has an increased risk for a range of poorer psychological outcomes compared to their non-affected peers. Research efforts should be doubled and redoubled, particularly in relation to the identification of factors contributing to resilience in children and young people and in order to clarify the ongoing impacts of cleft in adulthood. The detail of the experience of growing up with and living in the aftermath of cleft and its treatment should be better understood through prospective longitudinal research. Psychosocial interventions should be designed, implemented and evaluated, and the routine provision of psychosocial care across the lifespan should be promoted in order to optimise outcomes and to meet changing needs over time.

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Managing Teasing and Bullying

46

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Within the general population, it is well-established that teasing and bullying affect a significant minority of school-age children and adolescents; though prevalence study findings vary, between 15 and 40 % of young people in 21 industrialised nations have been reported to be affected (UNICEF 2007). As teasing and bullying in childhood are known to have a range of short- and long-term implications for psychological health and well-being (e.g. Herba et al. 2008; Kim et al. 2005), it has been the subject of extensive study over the past 10 years, with research focusing on defining the phenomena and reporting on prevalence, possible causes and implications for those affected. This chapter will briefly review the literature on teasing and bullying, as it relates to children and adolescents with cleft lip and/or palate in particular; its presence within and impact on the population will be considered and recommendations for management and intervention will be made.

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46.1 Defining Teasing and Bullying

Integral to the meaningful study of teasing and bullying is a consideration of the definition of these terms. Commonly, researchers consider three major components in a definition of bullying: one or more people act negatively with intent to harm; these actions are repeated over time; and the actions involve an imbalance of power between the perpetrator and victim (Olweus 1978). The inclusion of all three definitional components aims to distinguish typical peer exchanges from bullying (Leigh 2007), as does consideration of a lack of reciprocity, intentionality and interpersonal dominance (Crick and Dodge 1999; Marsh et al. 2001; Olweus 1978). Within this guidance, there is no definitive agreement about what frequency of negative incidents constitutes bullying, though it has been suggested that children who are bullied once or twice show evidence of different symptomatology than those who are bullied more frequently and over longer periods of time (Solberg and Olweus 2003). Experienced UK clinicians who have published a detailed review of this subject as a model to guide treatment and intervention argue, however, that teasing and bullying are best defined by the victim; they argue that the meaning of the event, whether it is called teasing or bullying, is central to the experience of, and the impact on, the individual (see Hearst et al. 2007). For some, teasing in the form of banter can be a positive experience when the context is one of play and can even be considered a sign of endearment (Keltner et al.

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2001). Generally, teasing and bullying are not found to be more common in either gender, though girls may be more involved in indirect and relational bullying (in which peer relationships are purposefully compromised by another; Theriot et al. 2005). Inclusion of cyberbullying, a more recent construct, may increase the rates of bullying in the typically developing population to close to one in four children (Li 2006).

46.2 Prevalence in the Cleft Population

It has long been assumed by lay people and professionals alike that visible facial differences or differences in speech make children more vulnerable to teasing and bullying (Rumsey and Harcourt 2005; Turner et al. 1997) and there has subsequently been much interest in researching this phenomenon in this population. Though estimates of the prevalence of teasing and bullying in the cleft population vary in the literature, broadly it is clear within this group that some children will be teased and bullied persistently over time; some will experience teasing and bullying, but no more so than children without a cleft, and others not at all (Broder 2001; Hearst et al. 2007). Children with and without a cleft may be classified as victims, bullies, bully victims and bystanders, and there is an extensive literature on how cognitions, emotions and behaviours can differ between the groups (Pellegrini et al. 1999; Toblin et al. 2005; Veenstra et al. 2005). There is a robust literature highlighting that the objective magnitude of visible difference does not necessarily relate to the individual's satisfaction with appearance (Ong et al. 2007) nor to the aggressiveness of teasing at the hands of others (Carroll and Shute 2005). It is important, therefore, that links are explored fully before assumptions are made.

Findings from an 8-year clinical audit within a multidisciplinary UK cleft service reviewing self-reported teasing and bullying revealed that around 24% of 10-year-olds endorsed that they had been teased or bullied, while 13% of 15-year-olds indicated the same (Shavel-Jessop et al. 2010). These estimates are broadly consistent with figures reported by Whitney and Smith (1993), who reviewed rates of teasing and bullying in the non-cleft population, suggesting that rates of teasing and bullying were not elevated for children and young people with a cleft. Other studies (e.g. Crozier and Dimmock 1999), however, have suggested that playground teasing commonly implicates appearance, and fear of perceived difference can increase aggressive behaviours (Bull and Rumsey 1988).

46.3 Understanding Teasing and Bullying in the Cleft Population

A number of theories have been put forward to conceptualise and formulate the potential difficulties or challenges facing the individual with a cleft. These theories are well-summarised by Hearst and colleagues (2007) and will not be discussed in detail here. Broadly, they include (1) family systems theories (e.g. Haley and Hoffman 1967; Minuchin 1974), which emphasise the family structure, strategies and trans-generational issues relating to parents' own experiences of teasing and bullying; (2) attribution theory (e.g. Kelley and Michela 1980), which highlights attributions of success or failure to internal or external sources; (3) locus of control (Rotter 1966), in relation to medical services and/or teasing and bullying; (4) learned helplessness (Seligman 1967), where victims feel they have little or no control over being teased, which can lead to them giving up on social experiences necessary to their developmental stage; (5) social desirability and social modelling (Bandura 1986), in which children learn to respond to others' teasing through their parents' behaviour, with possible impact on self-worth or self-image; (6) stereotyping, stigmatisation and scapegoating (Goffman 1963), where individuals are rejected and excluded on the basis of an attribute; (7) moral reasoning (Kohlberg 1984), which proposes that children move through stages of moral understanding with development; (8) shame and moral disgust, thought to be a consequence of a lack of support in moments of trauma or insecurity over time (e.g. Gilbert and Miles 2002; Haidt 2003; Parlett

and Hemming 2002; Rozin et al. 1999; Taylor 2007; Thompson and Kent 2001); (9) attachment, loss and adjustment (Bowlby 1969; Brazelton and Sparrow 2007), wherein the birth of a child with a facial difference may challenge parents' expectations (Solnit and Stark 1982; Tomko 1983), emotional states (including feelings of competence and self-worth) and roles (e.g. as buffer between the child and the world); (10) attractiveness and friendships, where popularity may be associated with perceived attractiveness (Boyatzis et al. 1998; Dion 1973), with further impact on social development (Rubin et al. 1999); and finally, (11) resilience, which though the integrity of the construct has been questioned in the literature (e.g. Sarason and Sarason 1993), refers to the process of competence in adversity (Luthar et al. 2000).

46.4 Development over the Lifespan

As cleft care is a life-long undertaking, and an individual's awareness, appraisals, perceptions and beliefs change as they age, it is essential that any model for understanding and intervening in psychological well-being is developmental in nature. A useful review and analysis of the development of teasing itself is by Keltner and colleagues (2001).

46.4.1 Birth, Infancy and the Preschool Years

Teasing and bullying in the earliest years are most likely to affect parents of a child with a cleft, rather than the child directly. In addition, it has been suggested by some clinicians that the reactions of new parents to an antenatal diagnosis or a diagnosis at birth, and their subsequent processing of these reactions, could have an impact on their attachment relationship with the child with a cleft. Findings are in consistent, however, with some studies finding an increased rate of secure attachment in parent-child dyads where the baby has a cleft (Collett and Speltz 2007). Additionally, attachment difficulties, if they are present, may not relate to the cleft alone. Rather, the birth of a child with a cleft may activate parental childhood issues, which may become entangled with the meaning of the cleft. As outlined above, difficulties with attachment may have a range of implications for adjustment and for the child's own coping in the future. It is notable that if difficulties in the attachment relationship do arise, they are rarely evident when the child is due to start school around the age of 5 years.

When children enter nursery or preschool, they are often exposed to groups of (unknown) peers for the first time, in a context where the protection and support of a parent is unavailable. Children start to learn quickly to manage their own emotions, while empathy emerges. These developing skills are all important for children who have a cleft, as they form the foundation of how children will cope with their social interactions with others. This may also be the first time that children are asked questions by other children about their appearance, scar or speech; how confidently children can answer these questions can have a significant impact on their social inclusion and self-concept, and vulnerable children may interpret curiosity as teasing (Feragen et al. 2009). Additionally, difficulties with hearing may be attributed to not listening or a social failure to respond, and children may be misunderstood by others.

46.4.2 School Age

When children reach school age, difficulties which may have begun to emerge in early childhood around others' misinterpreting hearing difficulties or making inaccurate assumptions about a child's learning or emotional needs can intensify. Given that it has been shown that a child's positive self-concept is a protective factor against the negative effects of teasing and bullying (Egan and Perry 1998), it is important that the child has every opportunity to develop a positive sense of self. Group identity also becomes increasingly important at this stage of development. Research suggests that teasing and bullying may peak between ages 6 and 9 years (e.g. Finkelhor et al. 2009). While it has been suggested that this

may be an artefact, due to study designs beginning assessment at this age, theories of brain and psychosocial development support the suggestion that a peak in bullying at this age is meaningful and not unexpected. Bullying at this age may be dominated by individual incidents, less intense ongoing interactive styles or social exclusion. At this time, children are learning about the difference between teasing or bullying and questions and curiosity and are faced with the difficult task of needing to respond appropriately to either.

During the school years, the response of an environment to teasing and bullying within its population is often essential to the impact of these experiences on the individual; cultural messages such as 'bullying is harmless and part of growing up', are generally unhelpful (Freeman and Mims 2007). By contrast, school policies and interventions which encourage positive disciplinary action, high academic standards and strong parental involvement have been shown to be effective in reducing bullying within the community (Xin 2002). As children make the transition to secondary or middle school between ages 10 and 12 years, there is the added complication of the possibility of needing to make new friends and engage with larger groups of unknown others. It is therefore possible, at this time, that previous difficulties with peers present anew at this critical point.

46.4.3 Adolescence

Adolescence is known to be a period of transitions and of achieving developmental tasks centring on individuation, identity and independence. The social world is especially critical to adolescents, with an adolescent's peer group featuring more prominently in the testing and achieving of developmental tasks than ever before (Sherif and Sherif 1964, 1965). Appearance is reported to be rated as the most important characteristic to young people in this age group (Prokhorov et al. 1993), and for young people with visible difference, this may feature especially prominently in their self-evaluation. As it has been repeatedly demonstrated that degree of physical difference does not relate to psychological outcome (e.g. Ong et al. 2007; Shute et al. 2007), it is important not to assume that dissatisfaction with appearance will necessarily relate, however, to cleft-specific characteristics. According to data from clinical audit, reported in Rumsey and Harcourt (2005), 15-year-olds did not report less satisfaction with appearance or friendships than peers without a cleft. It is important, therefore, that these issues are explored in an open-minded way with young people presenting to services.

A number of studies have revealed risk and protective factors related to teasing and bullying in adolescence. For example, authoritarian parenting and (for boys) overprotective parenting have been shown to increase vulnerability, while authoritative parenting has been shown to be protective (Baldry and Farrington 1998). Having a friendship group is also said to be protective, while social isolation increases risk (Hodges et al. 1997). Schwartz and colleagues (1998) report that beliefs and attributions that permit aggression and hostility increase risk, while being friendly and prosocial is helpful (Tani et al. 2003). Finally, emotional dysregulation has been shown to be potentially problematic for adolescents exposed to teasing and bullying (Mahady Wilton et al. 2000).

46.4.4 Adulthood

In adulthood, teasing and bullying can occur for the first time in the workplace. It can also perpetuate childhood insults, and clinically, the chronicity of this may worsen the experience. Furthermore, adults with a cleft may be more vulnerable if they themselves become parents of children with a cleft, as this may re-enact difficult childhood experiences. It is notable, however, that in adulthood, as in childhood, teasing and bullying are not inevitable in the presence of a cleft.

46.5 Consequences of Teasing and Bullying

Teasing and bullying have been shown to have a potentially far-reaching impact on the well-being of those experiencing these interpersonal difficulties

(Hunt et al. 2006). Most directly, teasing and bullying can negatively influence an individual's self-esteem and self-concept. It can lead to avoidance and social withdrawal and, in more extreme cases, precipitate the development of severe psychological distress. Experiences of teasing and bullying may lead to preoccupation with appearance. Some individuals may develop mood disorders, including anxiety and depression (Storch et al. 2003), though it is important to bear in mind that the relationship between teasing and bullying and psychosocial outcomes is far from linear (Hunt et al. 2006). Furthermore, young people who experience bullying, and girls in particular, have been shown to be at an increased risk of suicidal ideation and self-harm (Kim et al. 2005). A study by Herba and colleagues (2008) suggests this risk may be moderated by internalising disorders in parents and feelings of rejection at home.

Findings from an 8-year clinical audit within a UK multidisciplinary cleft centre service revealed that 15-year-olds with a cleft reported more avoidance (e.g. of talking to unknown others, meeting people, new situations, having photographs taken), associated with experiencing teasing and bullying, than did their 10-year-old peers. Fifteen-year-olds also reported a greater negative impact of the cleft on their lives than did the 10-year-olds. Though a proportion of young people reported no avoidance or 'bother' about being teased or bullied, there was a relationship between the negative impact of cleft and behavioural avoidance. This suggests that the more children and young people feel a cleft has negatively influenced their lives, the more likely they are to exhibit behavioural avoidance (Shavel-Jessop et al. 2010). Though avoidance may be considered an adaptive coping mechanism in the face of threat (Kapp-Simon 1992), it may also paradoxically increase vulnerability to teasing and bullying (Kapp-Simon et al. 2005), as it can maintain social isolation and poor peer relations (Robinson et al. 1986).

In adulthood, some individuals who have experienced teasing and bullying in their childhoods may find their self-esteem is compromised (Arseneault et al. 2008), and adults with anxiety disorders may endorse a history of teasing and bullying (McCabe et al. 2003). This may affect willingness to enter relationships or pursue ambitions, which can have a significant impact on long-term quality of life. It is notable, however, that not all individuals who experience teasing and bullying will go on to experience psychosocial difficulties in later life (Patel and Ross 2003). For most, these concerns resolve with maturation into adulthood, with future prospects no less bright than for individuals unaffected by cleft-related teasing and bullying.

46.6 Intervention and Management Recommendations

Given the possible impact of difficulties relating to teasing and bullying on young people with cleft lip and/or palate, it is essential that concerns are identified quickly and effectively, so that intervention can be timely. Within the UK, clinical psychology services within multidisciplinary cleft lip and palate services review patients at nationally agreed audit points (at 3 months, 18 months, 5 years, 10 years, 15 years and 20 years). When developmentally appropriate, children and their parents are asked about any concerns or difficulties they may be experiencing and an assessment of cleft-relatedness is made. Children and young people are then referred for further assessment and intervention, as needed, either within cleft services or outside, should the concern be beyond the remit of the specialist multidisciplinary team.

It is important to note, however, that the majority of individuals with a cleft have been shown to be no more likely to present with frank mental health difficulties than their non-cleft peers (Lockhart 2003). On the other hand, understandable concerns relating to difficult experiences are likely to benefit from targeted intervention, at an individual level, a systemic level, or a combination of the two.

46.6.1 The Individual

When therapeutic intervention is indicated, it is well established that there is a range of approaches

which have been shown to be effective for the treatment of psychological distress. These may include cognitive behavioural therapy (CBT; Beck 1976), which focuses on the relationship of thoughts to feelings and behaviour; schema-focused therapy, which for young people with a cleft may focus on addressing issues of learned helplessness, shame, loss of control and misattributions of intent (Stallard 2002); family therapy (Hoffman 1981), which focuses on the interactions of and within the family to address unhelpful patterns or interactions; social interaction skills groups (Kapp-Simon and Simon 1991; Kish and Lansdown 2000); and narrative therapy (White 2005) and life story work (Carroll 1998), which suggest that identity is shaped by narratives/stories which may or may not be problem-saturated.

Considering the developmental and formulation issues outlined in the previous sections, intervention for the individual who experiences teasing and bullying may take a range of forms. Parents may require support in managing anticipatory anxiety about their children being bullied and teased, and this may be particularly so if parents themselves have had such experiences. Broadly speaking, in the earliest years, the primary psychological need for children (with respect to teasing and bullying which may arise in future) is to develop a healthy and well-consolidated positive sense of self, to reduce their vulnerability to teasing and their capacity to cope with it, should it arise.

In the preschool years, children often benefit from behavioural intervention, which can be of use for children with social or behavioural difficulties. For parents of children at any age, and the children themselves, interventions which emphasise proactive support and formulation of difficulties have been shown to be effective (Maddern et al. 2006). As mentioned, no relationship has been found between the magnitude of visible difference and psychosocial outcome, despite repeated attempts to identify such a link (Robinson 1997). In the early years, therefore, offering parents and children reassurance that they will not necessarily be singled out for teasing can allay fears and help difficult situations be managed more confidently.

For school-age children, exploring their understanding of the difference between teasing and bullying and questions and curiosity in a proactive way can be beneficial, as can supporting their understanding of incidents as they arise. This psychoeducation (which may include a didactic component) can be effective in helping children to increase their skill repertoire. In addition to learning about the nature of teasing and curiosity, the school-age children often benefit from learning about their condition in an age- and developmentally appropriate way. The better able children are to respond to questions or curiosity, without distress, the more likely they are to experience success and confidence in managing interactions with others. Parental support to help children develop strategies to manage concerns and support their overall confidence is key.

For some children, where concerns have developed into poor self-esteem or low self-worth, mood difficulties, anxiety or social withdrawal, a direct therapeutic approach such as cognitive behavioural therapy may be advantageous. Though the evidence in the literature is less well-established, clinical experience suggests that narrative and other therapeutic approaches to such difficulties may also be helpful. Additionally, children may benefit from social skills training and from being equipped with a 'toolkit' of behavioural strategies (see Lovegrove and Rumsey 2005, for recommendations).

In adolescence, social skills training, as recommended for younger children, can be helpful in developing self-awareness and empathy, which can facilitate popularity, peer integration and use of effective body language (Kapp-Simon and Simon 1991). In this older group, however, interventions may focus more on the self-control and initiative-taking of young people, taking advantage of their developing regulation, monitoring, planning and organising (executive function) skills. Additionally, cognitive behavioural interventions may be beneficial to develop active coping strategies (e.g. Lovegrove and Rumsey 2005), challenge beliefs about others' thoughts and impressions and manage concerns around mood.

As it is for adolescents and younger children, social skills work may be helpful for adults

struggling to accommodate challenges to their self-esteem. They may benefit from practising positive self-statements and assertiveness and from cognitive behavioural therapy or other direct therapeutic interventions. When more profound difficulties around identity and perceived hopelessness have arisen as a result of a number of painful experiences (including severe teasing and bullying), in this adult patient group, offering longer-term psychotherapeutic interventions may be indicated.

46.6.2 The System

Whatever the specific concerns and experiences of the individual, it is important to recognise that context will undoubtedly influence the meaning of those experiences. Expectations, ideals and relevance of appearance will vary in different settings and cultural contexts, and attributions therefore need to be considered in the development of a formulation of difficulties and plan for intervention.

In addition to the intervention models suggested above, there is a range of bodies and organisations dedicated to providing support for individuals and systems experiencing difficulties with teasing and bullying, and engagement with these may enhance therapeutic interventions. Within the UK, organisations such as Changing Faces, Kidscape, Childline and Bullying UK provide online support, resources, helplines and training for children and young people, parents, carers and other adults. Legal frameworks also exist within which teasing and bullying can be positioned (see Fiddy and Hamilton 2004 for a useful review of these in the UK); guidance from agencies familiar with these issues (such as the Anti-Bullying Alliance within the UK) and input from local, national and international organisations, school and home support structures can be facilitative.

For some young people who are attending school, interventions at this systems level can be helpful. Programmes within schools which empower the bystander group to intervene and support the victim in bullying situations can also be of benefit. Explicit school interventions that emphasise explicit rules, warmth, positive interest, involvement from adults, firm limits to unacceptable behaviour, non-hostile and non-physical sanctions, authoritative adults, active surveillance of pupils (including in breaks) and proactive fostering of peer relationships are posited as decreasing bullying within the school context (Olweus 1991, 1993). Though replications have shown mixed findings, it has been suggested that long-term approaches to systems change are likely to be more effective than time-limited systemic interventions (Hunt 2007). Children and adolescents may benefit from strategies such as a protective posse or a 'buddy system', and school or institutional bullying policies may need to be reviewed.

Where children or adults alike have additional needs, such as hearing difficulties, it is essential that schools, workplaces and social systems be made aware of such challenges, so that incorrect attributions are not made about the individual and their social, emotional or cognitive abilities.

46.7 Considerations for the Future

Though many questions remain in the study and understanding of teasing and bullying in the cleft population, what is clear is that teasing and bullying remain a significant issue for many individuals affected by a cleft, as for their non-cleft peers. While having a cleft does not mean that an individual will necessarily be a victim of teasing and bullying, it is evident that those affected by these challenges are likely to benefit from proactive support and timely intervention, at both the individual and systems levels.

Clinically, working with individuals to manage teasing and bullying and its effects remains a significant part of the role of clinical psychologists within multidisciplinary cleft lip and palate services. It is argued that it is not the clinician's or team's decision whether or not an individual should be classified as 'bullied'; rather, this is a subjective patient-defined experience, and a judgement cannot be made by the treating team about the seriousness of the problem on objective statistics alone (Hearst et al. 2007). Additionally,

it is essential that professionals remain curious about the patient's experience and cautious about attributing causality or 'cleft relatedness' without thorough assessment.

Beyond this, there is an opportunity to improve the quality of interventions, taking into account a number of issues which may be addressed by further research and clinical audit. First, establishing a common language and conceptualisation of the constructs of teasing and bullying is essential, to ensure that research and clinical practice are measuring and discussing the same important phenomena. Second, the integrity of these constructs must be validated-work that is currently being undertaken by the UK Cleft Special Interest Group and colleagues. Third, emerging evidence (e.g. Shavel-Jessop et al. 2010) suggests that age effects relating to the psychosocial impact of teasing and bullying may be present within the cleft population, and this requires further longitudinal investigation and attention to elucidate developmental patterns which may inform more targeted intervention. Fourth, while a substantial body of evidence in support of cognitive behavioural therapeutic interventions exists, the evidence for other therapeutic approaches (e.g. systemic approaches) commonly used in clinical practice is less readily available. It is important that these approaches, which are thought to be of therapeutic benefit, are not disregarded or overlooked due to methodological challenges. Fifth, it is clinically observed that there is enormous variability in individuals' reactions to teasing and bullying; it remains to be determined how some children become more resilient to teasing and bullying than others and how this resilience can be promoted. This highlights the significance of exploration of within-group variation, which may inform much more than studies of cleft versus non-cleft populations have yielded to date.

Finally, it has been suggested within the appearance-related literature that as societal views and judgements likely underpin these interpersonal attitudes and difficulties, it may be unfair to focus therapeutic attention and responsibility for change on the individual alone (Clarke 1999); this is balanced, however, by the unfairness of leaving an individual suffering for the benefit of encouraging longer term societal change, and so interventions often focus on the individual and his or her needs. Alongside this is a need for the wider culture to work toward a society which is more inclusive, less judgemental and more supportive. With improved public awareness and sensitivity, individuals with and without a cleft may find themselves less under threat from teasing and bullying than they are today.

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Part XVI

Multicenter Clinical Reports

Eurocleft and Americleft Studies: Experiments in Intercenter and International Collaboration

47

Ross E. Long Jr., William C. Shaw, and Gunvor Semb

47.1 Monitoring and Improving Outcomes Through Multicenter Collaboration

47.1.1 Obligations

It goes without saying that professionals involved in health care have a continuing obligation to review the quality of care they provide. As stated in a WHO report (2002):

> Professionals entrusted with the provision of health care have an obligation to review the success of their practices and, where shortcomings are revealed, to take remedial action. Such efforts should constitute a continuous cycle, sometimes known as a 'clinical audit'…… (which) is divided into evaluating the *process* of care (the way in which care is delivered) and the *outcomes* of care (what is achieved) …… Audit of the treatment of clefts is a considerable challenge, because of the lengthy follow-up required, the complexity, subtlety and number of relevant outcomes and, above all, the relatively small number of cases. *Inter-center collaboration* still offers significant advantages, by providing insight into the processes and outcomes of treatment of comparable services elsewhere, the establishment of future goals and the exchange of clearly successful practices. … Perhaps the greatest benefit of inter-center comparisons is the cooperative spirit that they foster and a gradual diminution of rivalry.

Planned recall of patients and collection of standardized clinical records at prescribed points in time, not only provide the data for internal audit of outcomes, but if done identically by a group of centers, will allow comparisons to be undertaken.

47.1.2 Measuring Outcome

The ultimate goal of cleft care is restoration of the patient as far as possible to a life, unhindered by handicap or disability. However, the measurement of well-being is a highly complex proposition, and there is certainly no index at present that would allow sufficiently sensitive comparison between alternative treatment protocols. Whether in internal audit, intercenter comparisons, or clinical trials, emphasis will typically be on "proximate" outcomes. These will mainly represent different aspects of form and function in the parts affected by the cleft, often reflecting the particular interest of individual provider groups. In essence, most measures will be an indication of the deficits that persist despite (or as a result of) treatment, such as variations in appearance, speech, hearing, and dentofacial development.

For outcome measures to be deemed meaningful, they should reflect the needs and aspirations of patients and their families. They should be

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"patient centered." The general rules of reproducibility and validity apply, the latter being especially important when outcome is assessed before maturity. Longitudinal archives may be useful to determine the reliability of prediction for outcomes that are to be measured in the young (Shaw and Semb 1996; Atack et al. 1997). In relation to cleft surgery, experience with a number of outcome measures and scales has been obtained regarding speech, dentofacial outcomes, and patient satisfaction (Kuehn and Moller 2000; Sell et al. 2001; Williams et al. 2001).

47.1.3 Standardization

The standardization of records necessary to document those outcomes has also been established in the Eurocleft Consensus Recommendations on Timing of Minimal Records (Shaw et al. 2001) and also adopted by the Americleft Task Force. Further work is certainly needed to refine these and build consensus upon international standards. Reliable rating of appearance is still problematical, and for speech, linguistic differences represent a significant international challenge. However, efforts are underway to resolve these (see Sect. 47.3).

47.1.4 Scope and Limitations of Intercenter Comparisons

Provided procedures for entry into the study and record collection are equivalent in all participating centers, intercenter comparisons are extremely valuable in assessing the outcome of surgery, *together with* other major components of the treatment program at respective centers. However, for primary cleft surgery, it is difficult, if not impossible, to establish the key beneficial or harmful features of the specific treatment due to the invariably complex and arbitrary mix of surgical technique, timing and sequence, ancillary procedures, and surgical personnel (Shaw et al. 1992b). A recent survey of European cleft services revealed that, in 201 teams, 194 different surgical protocols were followed for unilateral cleft lip and palate alone (Shaw et al. 2001). In the context of intercenter outcome comparisons, this means that if two centers differ in the use of presurgical orthopedics and type of primary lip and palate surgery, there is no way to determine which of these procedures might be responsible for any difference in outcome between centers, nor would a null result allow the conclusion that individual aspects of the treatment program are equivalent.

Accordingly, intercenter comparisons are better suited to comparative clinical audit and quality assurance than definitive clinical research. The existence of significant disparities in outcome of the overall treatment process, however, provides a basis for speculating as to the possible cause, and intercenter studies should, therefore, be highly motivating toward the generation of specific hypotheses for subsequent trials. This was certainly the case in the Eurocleft cohort study.

47.2 The Eurocleft Cohort Study

The Eurocleft study (Asher-McDade et al. 1992; Mars et al. 1992; Mølsted et al. 1992; Shaw et al. 1992a, b) was the first to utilize retrospective analysis of clinical outcome records with rigid methodological control of biases to demonstrate the ability to detect favorable versus unfavorable approaches to primary infant management. It subsequently served as a model for the second such major intercenter collaborative outcome comparison, the Americleft Project. These projects and the results of the efforts will be described in Sects. 45.2 and 45.3.

The Eurocleft cohort study began in the late 1980s as an intercenter comparison of the records of 9-year-old children with complete unilateral cleft lip and palate. It sought to overcome, at least in part, some of the limitations and potential biases associated with the comparison of outcomes described in single center reports. A full account of the methodology and findings has been presented elsewhere (Asher-McDade et al. 1992; Mars et al. 1992; Mølsted et al. 1992, 1993a, b; Shaw et al. 1992a, b). Five of the original six teams agreed to continue a follow-up of the cohort till age 17.

47.2.1 Outcomes at Age 9

At age 9, several differences between the centers were apparent, especially for dental arch relationship (Fig. 47.1). Whereas only 7 % of cases for center E were considered to have a likely future need for osteotomy, almost half (48 %) did so for center D. It was not possible however to ascribe success or failure to particular details of the surgical protocols, but poor outcomes appeared to be related to decentralized services without consistent protocols.

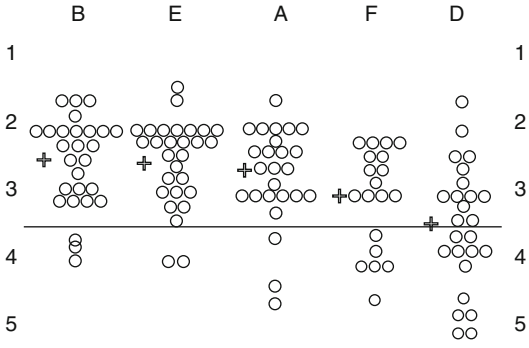

Fig. 47.1 Goslon individual patient scores at age 9 by center. A Goslon score of 1 represents excellent maxillary prominence and a score of 5, severe maxillary retrusion. One way to consider this outcome variable is the likely future need for subsequent maxillary osteotomy, and cases falling below 3.5 at this age are likely candidates for osteotomy in the late teens

47.2.2 Follow-Up

The aims of the follow-up were: to quantify the burden of care imposed by respective protocols, to see whether the ranking of centers for different outcomes at age 9 was predictive for equivalent outcomes at age 17, to assess patient/parent satisfaction with care, and to explore interrelationships with outcome and burden (Brattström et al. 2005; Mølsted and Brattström 2005; Semb et al. 2005a, b; Shaw et al. 2005). A separate comparison of speech outcomes was carried out at age 11–14 (Grunwell et al. 2000).

47.2.3 Survey of Treatment Experience

The amount of treatment provided by the five different teams in 1976–1979 was remarkably different (Table 47.1). Most notable was the lengthy hospital stay associated with presurgical orthopedics at that time in centers D and F. The subjects in center D also had more orthodontic visits for treatment and review and for the overall number of surgeries compared to the other centers. From discussion with these centers, it would seem that the reason for the large differences in the intensity of treatment was not primarily related to clinical need but rather to differing beliefs and historical practices that had shaped the clinical protocols of the period.

Table 47.1 Amount of treatment provided by five different teams from birth to 17 years of age

		A	B	D	E	F
Surgery						
Mean number of surgeries		4.8	3.3	6.0	4.4	3.5
Mean days in hospital		33	31	60	24	26
Presurgical orthopedics						
Months of treatment		13	0	15	0	5
Number of visits		11	0	8	0	17
Days in hospital		0	0	60	0	146
Orthodontic treatment						
Treatment length (years)		5.6	3.3	8.5	3.5	4.0
Number of visits	Treatment	52	41	54	33	47
	Follow-up	11	23	42	16	25
	Total	63	64	94	49	72

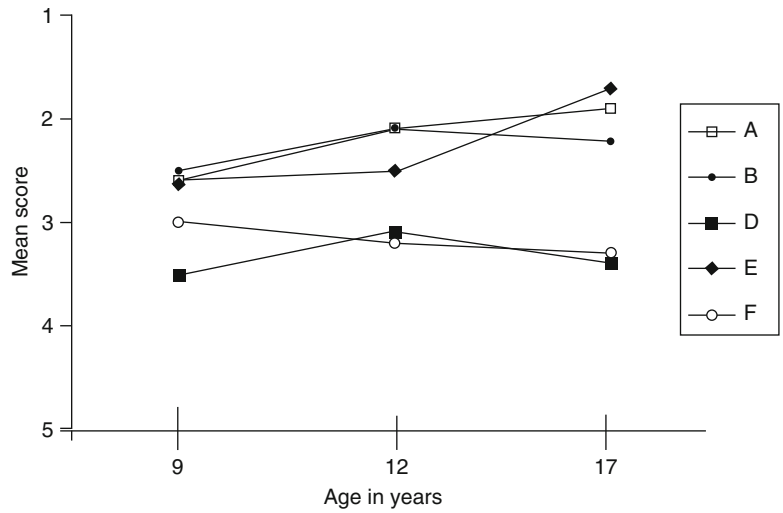

Fig. 47.2 Mean dental arch relationship scores at ages 9, 12, and 17 years for participating centers

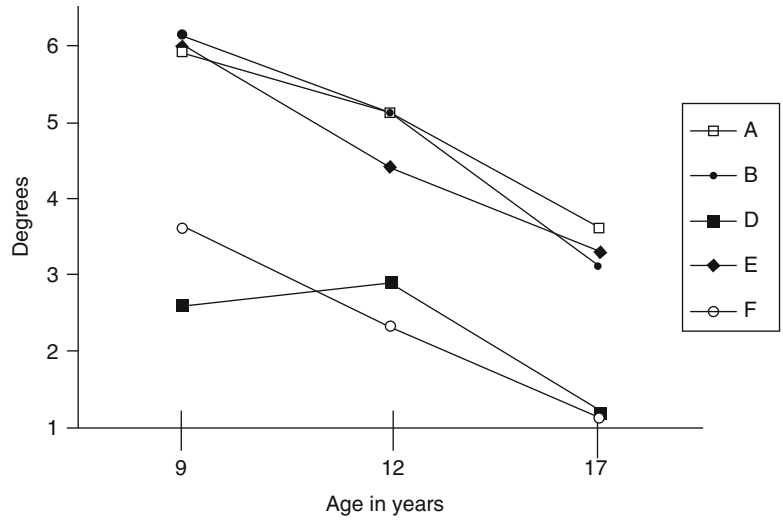

Fig. 47.3 Mean soft tissue profile (angle SSS-NS-SMS) at ages 9, 12, and 17 years for participating centers

47.2.4 Consistency of Outcomes over Time

The statistical analysis used to compare the five centers was a general linear mixed model applied to longitudinal data (Diggle et al. 1994). Variance terms were included in the model to account for between subject variation in the intercept as well as fixed factor for assessment point (9, 12, 17 years) and center. Full details have been reported elsewhere (Shaw et al. 2005).

As Fig. 47.2 indicates, the scores for dental arch relationship tended to improve in centers A, B, and E, but not in D and F. There was a consistent relationship over time for most cephalometric variables, e.g., soft tissue profile (Fig. 47.3), and for nasolabial appearance.

47.2.5 Lack of Association Between Outcome and the Amount of Treatment

Not surprisingly, follow-up of these five cohorts of patients from age 9 to age 17 confirmed the main finding of the first report, with some centers continuing to achieve considerably better outcome than others, at all age points. Perhaps,

Table 47.2 The relationship between outcome assessment (dental arch relationship at 17 years) and amount of infant orthopedic treatment in the different centers

Objective ranking	Center	Months of treatment	No. of visits	Days in hospital
Best	E	0	0	0
↕	A	13	11	0
	B	0	0	0
	F	5	17	146
Worst	D	15	8	60

Table 47.3 The relationship between outcome assessment (dental arch relationship at 17 years) and amount of orthodontic treatment in the different centers

Objective ranking	Center	Treatment-length (years)	No. of visits	
			Treatment	Checkup
Best	E	3.5	33	16
↕	A	5.6	52	11
	B	3.3	41	23
	F	4.0	47	25
Worst	(D)	8.5	54	42

Table 47.4 The relationship between outcome assessment (dental arch relationship at 17 years) and the mean number of surgeries per patient in the different centers

Objective ranking	Center	Number of surgeries
Best	E	4.4
↕	A	4.8
	B	3.8
	F	3.5
Worst	D	6.0

more surprising is the lack of association between amount of treatment and final outcome (Tables 47.2, 47.3, and 47.4). Especially ironic is the finding that the two centers with the highest intensity of early treatment (hospitalization in order to perform presurgical orthopedics) achieved the lowest rankings for eventual outcome (Figs. 47.2 and 47.3). Thus, the poorest ratings for nasal appearance were associated with the lengthy use of a presurgical device called T-traction designed not only to reduce the alveolar gap but also to straighten the nasal septum (Nordin et al. 1983). Patients in the center with

Table 47.5 The relationship between objective ranking of nasolabial outcome and patient dissatisfaction

Objective ranking	Percentage of respondents dissatisfied with nasal appearance		Objective ranking	Percentage of respondents dissatisfied with lip appearance	
Best	A	64	Best	B	14
↕	E	32	↕	A	41
	B	14		F	6
	D	45		E	42
Worst	F	33	Worst	D	16

the least favorable dentofacial outcomes (center D) also experienced the longest orthodontic treatment duration and the highest number of orthodontic visits. It appears that this was partly due to the complexity of center D's orthodontic treatment protocols with almost continuous treatment from the eruption of the primary dentition and partly to the unfavorable dentofacial outcomes of primary surgery.

This lack of association between treatment outcome and intensity may represent a key lesson for the development of future protocols. It justifies an emphasis on simplicity, economy, and minimized burden for the patient, rather than adherence to demanding protocols with unsubstantiated promise.

47.2.6 Lack of Association Between Outcome and Satisfaction

Perhaps, the most perplexing finding of the Eurocleft series is the inconsistency between objectively rated outcomes and patient/parent satisfaction. There were instances where the highest levels of dissatisfaction with treatment outcome were reported by subjects attending the centers with the best objective ratings (Table 47.5). The possible reasons for this disparity have been discussed elsewhere (Semb et al. 2005b), and it highlights the need for concerted work on the understanding and measurement of patient/parent satisfaction and the provision of more holistic models of cleft care.

47.3 Wider Networks

Following initial reports of the Eurocleft cohort study, numerous teams from Europe and elsewhere approached members of Eurocleft and arranged to undertake blinded comparisons of their records with the Eurocleft material or subsections of it, some of which were published (Flinn et al. 2006; Fudalej et al. 2009; Gaukroger et al. 2002; MacKay et al. 1994; Meazzini et al. 2008, 2010; Nollet et al. 2005; Roberts-Harry et al. 1996). Also, during the Eurocran project that was subsequently funded by the European Union from 2000 to 2005, further centers from Europe had the opportunity to undertake similar comparisons. More recently, multicenter comparisons were completed in India and Turkey, and new initiatives are underway in Thailand and South Africa (Susami et al. 2006; Alex 2011; Bellardie 2011; Dogan 2011). The most extensive initiative was carried out in North America and is described below.

47.4 Americleft

Before 2006, centers in the USA and Canada had not been as successful as those in Europe in establishing interest and commitment to intercenter collaborative outcome studies. In the 2002 WHO report "Addressing the Global Challenges of Craniofacial Anomalies," it was noted that in the USA and elsewhere in North America, there had been little significant momentum in the area of intercenter, collaborative, clinical research, especially compared to the more successful efforts of Eurocleft and Eurocran. As a result, there was little useful information being generated from the ongoing research which would contribute to the establishment of sound evidence-based decision making in clinical care.

47.4.1 The Challenge

The reasons for this failure were complicated. While the large number of centers and individuals providing treatment for CFA in North America improved patients' geographical accessibility to care, it simultaneously created a fractionation of the study population, thereby reducing the probability of developing patient samples of adequate size to enable valid research. The entire landscape was further complicated by noncomparable patient populations, noncomparable treatment records, unquantifiable differences in operator skills, and difficulties in letting go of biases. Also, while collaborative research could be structured without violating patient privacy laws, the rigors of doing so were sufficient discouragement for many clinicians to participate. Finally, there remained a general lack of agreement between centers on minimal standards for reporting and recording outcomes, as well as cost and ethical concerns over taking records which cannot be clearly identified as essential for diagnosis and treatment purposes.

In summary, although the desire, research talent, and patient samples were all readily available in the North America, before 2006, the failure to get centers to agree on something as basic as standardization of recording and reporting outcomes, as well as governmental hurdles and a serious lack of funding, all resulted in a huge and ongoing missed opportunity. The WHO (2002) concluded by stating that … "it seems most likely, that the most promising avenue to break out of this inertia, may still lie in the original Eurocleft approach. With a core of interested and experienced clinicians, operating at high volume centers, and willing to agree on records, outcome measures of significance, and research protocols, and additionally with the possible guidance from those involved in the successful Eurocleft, Scandcleft and Eurocran programs, it might still be possible to initiate a major inter-center collaborative research effort."

47.4.2 Initiation

Based on this report, in 2006, the American Cleft Palate-Craniofacial Association (ACPA) and the Cleft Palate Foundation (CPF) approved funds to organize a pilot project which has become the "Americleft Project" and ACPA's Americleft Task Force. From ACPA's strategic plan, the purpose of this initiative is "….to continue to develop

strategies and execute intercenter collaborative outcomes studies in multiple disciplines for the purpose of documenting the outcomes and assessing best practices of team care."

Five centers were identified to participate in the pilot project which was modeled exactly after the Eurocleft study and resulted in completion of initial comparisons of dental arch relationship outcomes, cephalometric skeletodental morphology outcomes, and nasolabial appearance outcomes. The results of these, to be discussed below, confirmed the value and benefits of well-controlled and well-designed intercenter outcome comparisons. Most importantly, however, was the experience and insight that was gained in understanding the requirements, demands, and possible obstacles that must be overcome in order to participate successfully in such collaborative studies.

47.4.3 Participants

The success of Americleft has always been based on the integrity and intellectual honesty of those choosing to participate and several key requirements for participation. Participating centers' team members had to be experienced and focally interested in CLP and with an interest in seeking knowledge about the relative merits of various treatment protocols rather than having an unquestioning loyalty to particular procedures. While we as care providers all believe that the procedures we are doing are the best possible for our patients, involvement in collaborative outcome studies implies a degree of *uncertainty* about the true effectiveness of our individual protocols, the ability to question our own beliefs and to accept the possibility that there may be other equally good or better outcomes with protocols different from the one(s) used by our own team. In addition, the Americleft centers had to have a high volume of patients and well-defined protocols for management of their patient population. Centers also had to have the resources to support team representatives in dedicating the time and absorbing the costs of the effort as well as the availability of the necessary records (privacy protected) and ability to secure IRB approval from the parent institution.

47.4.4 The Original Foundation Americleft Cohort Studies

The initial studies conducted as part of the Americleft Project were identical to those carried out in the original Eurocleft study (Long et al. 2011): 9-year-old, mixed dentition comparison of dental arch relationship (Hathaway et al. 2011), craniofacial form (Daskalogiannakis et al. 2011), and nasolabial appearance (Mercado et al. 2011b). The patients were consecutively treated by high-volume surgeons in their respective centers. The outcome measures were also identical to those used in the Eurocleft study, including dental arch relationship ratings using dental casts and the Goslon yardstick (Mars et al. 1987), skeletal and soft tissue morphology using lateral cephalometric radiographs and standard cephalometric analyses, and ratings of nasolabial appearance using cropped lateral and frontal facial photographs and the Asher-McDade rating system (Asher-McDade et al. 1991). All measurements and ratings were done by panels of trained and calibrated examiners using validated and well-established methods and with all examiners blinded as to the craniofacial center of origin for each case. Reliability statistics were carried out on all studies and indicated acceptable levels of inter- and intra-rater reliability. The purpose of using records, outcome measures, and methods that were identical to those used in the Eurocleft study was to allow for consistency and valid cross comparison of findings from both initiatives.

The conclusions reached also mirrored those of the original Eurocleft study (Russell et al. 2011a). Figure 47.4 illustrates once again, with the use of the Goslon yardstick to compare dental arch relationships (Mars et al. 1987), the clear differences between centers with center C having only 18 % of patients considered to have a likely future need for osteotomy (Goslon scores 4 and 5), while over 60 % presumed so for center B. Furthermore, the inclusion, in infant management protocols, of passive presurgical infant orthopedic appliances to improve the alignment of cleft maxillary segments before primary surgical repair produced no measurable benefits in terms of dental arch relationship, skeletal

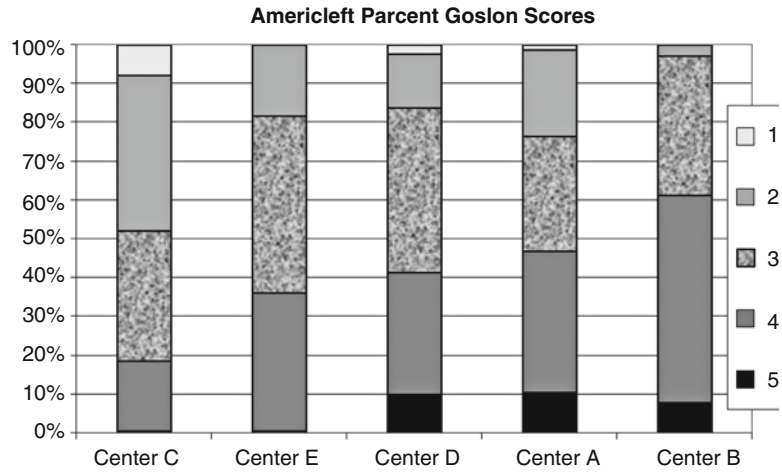

Fig. 47.4 Distribution of Americleft Goslon scores at age 9 by center. A Goslon score of 1 represents excellent maxillary prominence and a score of 5 severe maxillary retrusion. One way to consider this outcome variable is the likely future need for subsequent maxillary osteotomy, and cases in categories 4 and 5 at this age are likely candidates for osteotomy in the late teens

morphology, or nasolabial appearance in the mixed dentition-aged patient. To the contrary, of the three original Americleft centers which used infant orthopedic treatment (centers B, D, and E), all were not significantly different, or worse than the Americleft center that used only conservative primary lip and palate surgery (center C). Given the additional costs and burdens of care to families and patients that are invariably incurred as part of this addition to the treatment protocol, lack of a significant difference at best indicates that the procedure did no harm but at worst that it also provided no benefit to justify the burden. Of even greater significance perhaps also is the fact that in the original Eurocleft study, exactly the same conclusion was reached, and subsequent to that, a randomized controlled trial called Dutchcleft (Prahl et al. 2001, 2003; Bongaarts et al. 2004, 2006) finally established, with the greatest strength of evidence, what Eurocleft and Americleft had suggested based on well-controlled intercenter comparisons.

Another finding from the original Americleft Project publications was that the inclusion of primary alveolar repair with primary bone grafting (center B) was associated with the least favorable dental arch relationships and skeletal morphology in the mixed dentition (Fig. 47.4). Specifically, significantly more dental and skeletal class III relationships with maxillary retrusion were found in center B's patients. As with the use of infant passive orthopedics mentioned above, of additional significance also is the fact that in the original Eurocleft study, exactly the same conclusion was reached. Furthermore, this conclusion was consistent with that of a small randomized controlled trial of primary versus secondary bone grafting by Robertson and Jolleys (1968, 1983).

47.4.5 Additional UCLP Comparisons with Protocols Including Treatment Features of Interest

Using samples from the foundation Americleft cohort as a baseline for additional comparisons, Americleft's three most recent comparisons included centers which used nasoalveolar molding (NAM) as part of an infant management protocol. In one of these Americleft comparisons of unilateral clefts, the use of NAM in the infant management protocol produced no measurable improvement in mixed dentition skeletal morphology or dental arch relationship (center A vs. centers B, C, D in Fig. 47.5) (Peanchitlertkajorn et al. 2010, 2011). To the contrary, the average dental arch relationship score for the NAM center (center A) was actually significantly worse than the best Americleft center (center C) described previously and nearly identical to the poorest Americleft result (center B). However, there was evidence (Fig. 47.6) that the nasolabial

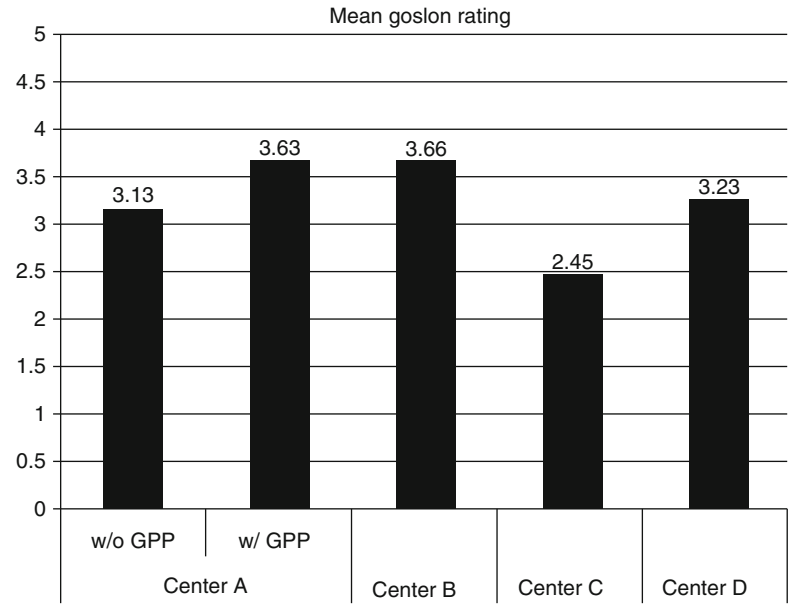

Fig. 47.5 Means of Americleft Goslon scores at age 9 by center. A Goslon score of 1 represents excellent maxillary prominence and a score of 5 severe maxillary retrusion. Protocols for center A included NAM ± GPP, center B included infant orthopedics and primary bone grafting, center D included infant orthopedics, and center C utilized only lip and palate surgery

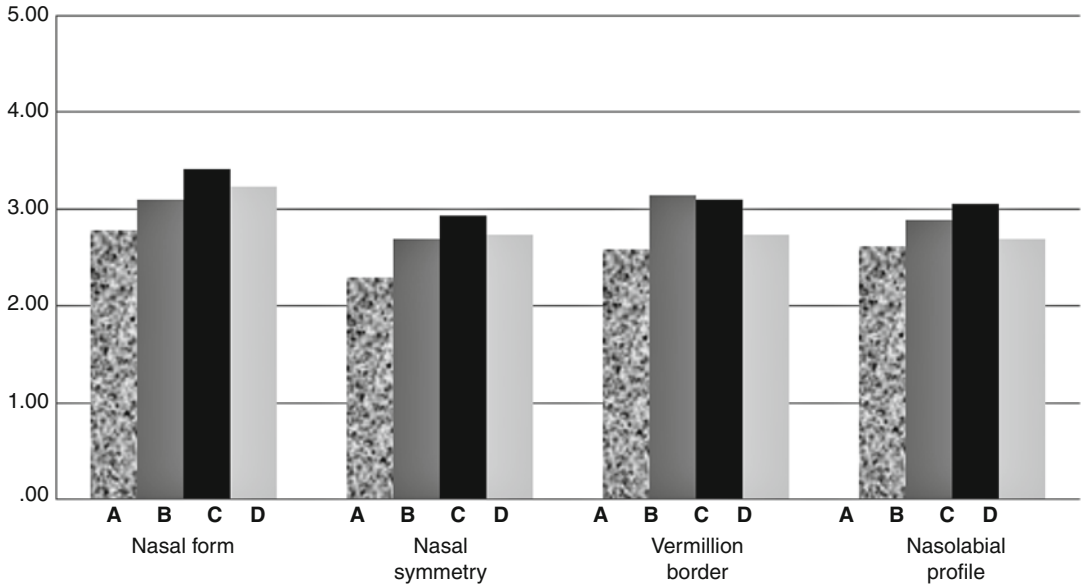

Fig. 47.6 Means of Americleft nasolabial appearance scores at age 9 by center. A score of 1 represents excellent appearance and a score of 5, poorest appearance. Protocols for center A included NAM ± GPP, center B included infant orthopedics and primary bone grafting, center D included infant orthopedics, and center C utilized only lip and palate surgery

appearance of the NAM-treated patients (center A) was significantly better than the Americleft center which used no infant orthopedics nor any secondary nasolabial surgical revisions (center C) (Mercado et al. 2011a). Interestingly, there was a trend for the NAM center's ratings to also be better than the Americleft centers which did not use NAM but had carried out secondary surgical revisions of lip and nose (centers B and D). However, these differences were not statistically

significant. Since one of the primary objectives of NAM is to improve nasolabial esthetics and presumably thereby to reduce the need for additional revision surgeries later, these findings suggest that there could be some merit to further exploring that possibility. But if it is found that centers using secondary surgeries instead of NAM are able to produce similar nasolabial appearance outcomes, the final decision as to "best practices" for nasolabial appearance will rely on a comparison of the burden, cost, and risk of both methods.

To further complicate the picture, however, in a second comparison of nasolabial appearance in 5-year-old patients from another center using NAM (Singer 2012), two of the original Americleft centers and one Eurocleft center which had 5-year records available, no significant differences were found in frontal and profile nasal appearance. Of importance in this most recent comparison was the inclusion of an assessment of the total burden of care involved in the management of both NAM- and non-NAM-treated samples. Not unexpectedly, the group receiving NAM had more clinical appointments and more total costs involved than those with no infant orthopedics or no secondary surgeries. Clearly, in situations in which there are several approaches to a problem which may produce equivalent outcomes, the one which can do so with the least burden, cost, and risk of care would be considered the most desirable. These preliminary findings are strongly suggestive of the need for further comparisons which would include multiple additional centers using NAM, as well as the initiation of a randomized controlled trial.

Since many centers using the NAM procedure include also another form of primary alveolar repair called gingivoperiosteoplasty, whether the inclusion of this feature also has a potential negative effect on growth has yet to be established. This possibility has been suggested by some previous retrospective studies, as well as by a small subset of the patients from the NAM center described above (see Fig. 47.5, center A with GPP), but has also been refuted by other studies. In support of the purpose of this entire discussion, the existence of such controversies strongly suggests the need for intercenter comparative outcome research and well-controlled clinical trials to provide us with the necessary evidence. Clearly, both the limitations and the benefits of these multicenter, intercenter comparisons are evident in these studies. On the one hand, attempting to attribute a favorable or unfavorable outcome to a specific feature within a particular protocol is an impossibility. However, certain features of interest within protocols that can be shown to produce more or less favorable results can be identified as worthy targets of future investigations using methods more capable of isolating their effects and burdens.

47.4.6 Comparisons of Outcomes in Bilateral Cleft Lip and Palate

Two additional comparisons of dental arch relationship and skeletal morphology were completed by the Americleft Project (Hathaway et al. 2008; Daskalogiannakis et al. 2010). These studies compared a cohort of 9-year-old mixed dentition BCLP patients from 3 of the original Americleft centers, using a dental model yardstick for BCLP, the Bauru yardstick (Ozawa et al. 2011), and standard cephalometric analysis. As in the original UCLP study, the most favorable results were seen in the patients from center C, with the least favorable results in center B which included infant orthopedics and primary bone grafting in its protocol.

47.4.7 Comparisons of Outcomes of Secondary Alveolar Bone Grafting

In an expansion of the goals of the Americleft Project, an additional procedure of interest which has a wide range of protocol and technique variations is secondary alveolar bone grafting. Three Americleft centers (A, C, and D from the original study) and one Eurocleft center (E from the original Eurocleft study) provided a total cohort of 152 consecutively grafted UCLP and BCLP patients with postgrafting

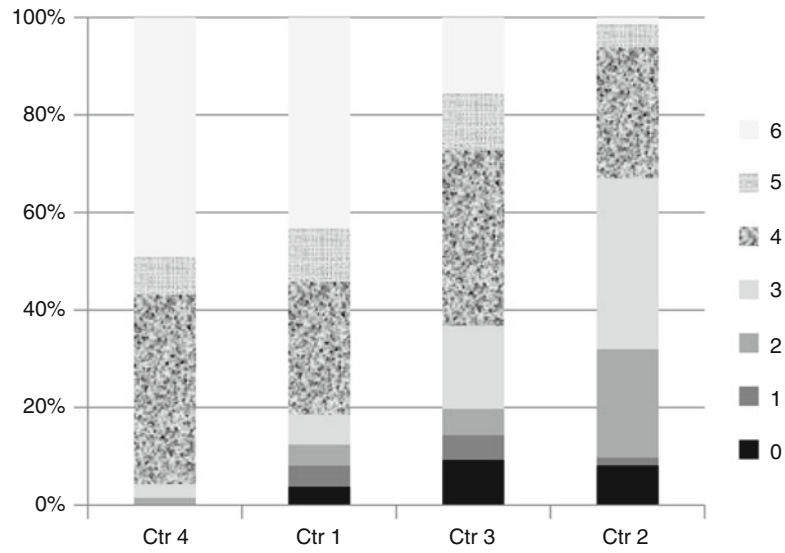

Fig. 47.7 Distribution of Americleft SWAG scores by center. A SWAG score of 0 or 1 represents failed graft with no bone bridge and a score of 6 an excellent graft with complete bone fill-in. One way to consider this outcome variable is the likely need for regrafting cases in categories 0–3 and less likely or no need for additional grafting surgery in cases rated as 4–6

follow-up occlusal radiographs. Using a new 6-point bone graft rating scale developed by the Americleft team (standardized way to assess grafts or SWAG method) (Russell et al. 2012), graft outcomes at a range of postgraft follow-up times were compared (Russell et al. 2011b). As with many other intercenter comparisons described above, a wide and distinct range of outcomes was seen between centers (Fig. 47.7). With scores of 0–3 indicating failed grafts definitely requiring additional surgery, and 4–6 indicating more successful grafts, it is clear that centers 1 and 4 had significantly higher success rates than centers 2 and 3. The protocols associated with the better graft results in this study included some features of interest such as nasal floor repair at the time of primary lip repair and the least amount of maxillary expansion presurgically. However, there was also a wide range of ages and length of follow-up, so additional comparisons are needed which may provide better control of those variables.

To initiate this process, the most recent Americleft study evaluated changes in bone graft appearance and rating over time, between two of the four centers in the initial bone graft outcome study (centers 2 and 4) (Ruppel et al. 2012). The intent was to determine if there was a minimal time of postoperative follow-up necessary to make valid assessments of bone graft outcomes, as well as a possible optimal time for the assessments (e.g., before, during, or after canine eruption). These centers had both short-term (T1) follow-up radiographs (average 1 year 3 months) and long-term (T2) follow-up radiographs (average 7 years 9 months). Of greatest importance was that the group differences in outcomes from T1 to T2 were identical in terms of average ratings and distribution of scores (Ruppel, et al. 2012). Although a small number of individual patients seemed to improve or worsen significantly (more than 2 categories), in general, the conclusions reached about each centers' average bone graft outcomes were identical whether assessed at 1 year postsurgery or 7 years postsurgery. An additional finding of interest was the suggestions of some trends observed using multiple regression analysis. Although not reaching statistical significance in this study, these included the finding that using canine substitution for missing lateral incisors was associated with a greater chance of improving a rating over time than attempting to utilize present but diminutive lateral incisors or to hold space for a fixed bridge replacement. This suggestion that final bone graft results may be affected by management of the lateral incisor in the cleft site certainly indicates the need for additional outcome comparisons.

47.4.8 Comparisons of Speech Outcomes in UCLP

The most recent expansion of the Americleft Project has been the development of an Americleft speech group. Nine speech-language pathologists (SLPs) from cleft palate clinics across North America (NA) participated in a 2-day training session on the Cleft Palate Audit Protocol for Speech – Augmented (CAPS-A) (John et al. 2006). Similar to the approach taken with the previous Americleft studies, the use of a speech outcome assessment tool that had been shown to be reliable and valid in previous studies was intentional. It saved time in that they did not need to develop and validate a new rating tool, and it will facilitate cross comparison of outcomes between the European centers in the UK and Americleft. The training, including data collection, recording, and perceptual speech rating procedures, was provided by three European SLPs who developed and validated CAPS-A. Inter- and intra-judge reliability were established by rating 10 samples (from the UK and Ireland) in three separate listening sessions (pretraining, posttraining, and maintenance). Minor modifications were made to the CAPS-A to improve ease of use and implementation with North American samples. Based on these modifications to the CAPS-A protocol, a follow-up reliability study was done with ten US samples and the revised CAPS-A scoring protocol (CAPS-A Americleft Modification).

Reliability with the new speech outcome rating tool was found to be acceptable, and the first intercenter comparisons of approximately 50 samples collected from four sites is currently in progress (Cordero et al. 2012). Other accomplishments include the development of the procedures for sending samples electronically to the University of Utah where they are edited and posted on a secure server at the university for remote rating of the samples (Wilson et al. 2012). The procedures to be employed for rating the samples (e.g., random assignment of raters, raters not rating their own samples, plans for inter-judge and intra-judge reliability, posting of background information on a shared spreadsheet) have also been finalized. This information has been added to the Americleft Project Study Guide and is posted and available on the ACPA website (www.acpa-cpf.org).

47.4.9 Future Americleft Plans

While these preliminary findings from the Americleft Project have produced some very valuable evidence to help us identify best practices in cleft palate management, possibly the most important accomplishment could be the stimulation of interest and enthusiasm between cleft-craniofacial centers in participating in such comparisons. Clearly, much remains to be done. Current Americleft plans include expansion of the NAM comparisons to involve other centers using that technique; comparison of outcomes from centers using gingivoperiosteoplasty and active pinned infant orthopedic appliances; further comparisons of secondary bone graft outcomes resulting from protocols varying in timing, type, and sequencing of the grafts with orthodontic treatment such as expansion, incisor alignment, and orthopedics; and comparisons of outcomes from different management methods for the lateral incisor in the cleft site such as crown buildups, bridges, implants, and cuspid substitution. Beyond the surgical and orthodontic aspects of the Americleft Project, future plans also include the expansion of the speech group comparisons to include many additional centers with varying protocols for speech management, as well as the initiation of a psychosocial component to the study with emphasis on patient and parent satisfaction and quality of life issues.

Coincident with all of these outcome comparisons, all ratings of outcomes from various approaches are being compared in the context of the total burden of care that each carries with it. A protocol for evaluating the burden of care and allowing for intercenter comparisons is being developed similar to that described for the Eurocleft study in Sect. 47.2. Once this total, comprehensive assessment of multiple treatment outcomes from a wide range of treatment approaches used by multiple cleft-craniofacial centers is completed, we will hopefully have at

our disposal a clearer picture of the treatment approaches for which we have sufficient evidence, with or without actual clinical trials, to identify best practices in cleft palate care.

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Part XVII

Recording Patient Orthodontic/ Surgical Findings

Berkowitz Recording Patient Information Form

48

Samuel Berkowitz

48.1 Cleft Lip and Palate Guide: Form Used to Record Surgical/Facial Growth Changes

(To be used for newborns as well as for any other age group)

A child born with a cleft and/or other craniofacial anomaly is identified at the *obstetric hospital* usually using a simple Birth Defect Registry Form. The child is then referred to the *cleft palate clinic* or the *craniofacial anomaly center* for a more complete diagnostic work-up and treatment planning.

At the cleft palate clinic or craniofacial anomaly center, the following entries are completed. This may be the first record taken:

(a) *Physical systems and structures*
(b) *What else to look for, etc.*
(c) *Recommended tests*
(d) *Facial clefts* – Tessier's classification
(e) *Initial evaluation of the cleft lip and/or palate*: This form highlights that although clefts may be similarly classified, they can be very different in physical appearance and degree of spatial distortion. This difference helps to explain why the same procedure can yield different results. For example, in bilateral clefts, the degree of premaxillary protrusion at birth may or may not influence the child's facial convexity and palatal relationship at a later age.
(f) *The dental, prosthetic, and orthodontic evaluation*: Only a few items need to be recorded for the next 2–3 years. It will document whether presurgical orthodontics or a speech aid appliance has been utilized. The prosthetic dental form will record the patient's status. The more detailed information will be recorded as treatment progresses.

Summary of Surgical Record: This will be the surgeon's choice; however, each specialist will know why the procedure has been selected and when it is to be done. Comments are requested.

*Cannot determine extent of impairment due to surgery preceding first.

Similarity to known syndromes

_____

Form completed by: _____

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48.2 Physical Systems and Structures

I. Case #: __ __ __ __ (1-4)
Name: _____ (5-19) _____ ____
(Last) (First) (M)
Date of First Exam: __ __ __ __ __ __ (43-48)

II. Sex: Male…1 Female…2
Race: Cauc…1 Black…2 Orient…3 Other…4_____ (50)
Current: __ __ oz, % __ __; Ht __ __. __ __ cm. % __ __

III. Systems & Structures

1. Lip: UPPER: Norm…1 Abnorm…2 No Info…9 (05)
CLEFT: None…1 Present…2 No Info…9 (06)
LOWER: Norm… Abnorm… No Info…9 (07)

	Left	Right	Medium	No Info
Complete	1	1	1	9
Incomplete	2	2	2	9
*Surgery	3	3	3	9
Lateral	1	2		9 (11)
Oblique	1	2		9 (12)
Other	None	Present	No Info	

	None	Present	No Info	
Median Cleft	1	2	9	(15)
Central Lip Pits	1	2	9	(16)
Commissural Pits	1	2	9	(17)
*Surgery	1	2	9	(18)
Other	1	2	9	(19)

Specify: _____

Specify: _____

2. Hard Palate: Norm…1 Abnorm…2 No Info…9
Cleft: None…1 Present…2 No Info…9
3. Soft Palate: Norm…1 Abnorm…2 No Info…9
4. Uvula: Norm…1 Abnorm…2 No Info…9
Bifid…1 Cleft…2 Missing…3

	Left	Right	Medium	No Info
Complete	1	1	1	9
Incomplete	2	2	2	9
*Surgery	3	3	3	9
Sub mucous	4	4	4	9
	(22)	(23)	(24)	
Other	None	Present	No Info	

Cleft:	Complete	1
	Incomplete	2
	Zona Pellucida	3
	*Surgery	4
	Other (specify)	5

Specify_____

NOTE_____

(28)

In the following systems & structures
you are asked to indicate whether the impairment is suspected or confirmed.
-Suspected means that validation of the observation by other means must be had
-Confirmed means that validation has occurred.
-Age first noticed is only to be used if the age is other than birth.
-Specify means that you are to give a detailed but brief description of the nature of the impairment.
-Do not leave any individual syst/struct blank, as it will be assumed that it was not examined.

OCCLUSION

IV. EVALUATION PRIOR TO TREATMENT:

Angle Classification:
(To be recorded per side)
Teeth in crossbite:
I II III
Right side_____ Left side_____
Right: A B C D E
Left: A B C D E

Right: 1 2 3 4 5 6 7 8
Left: 1 2 3 4 5 6 7 8

Overjet_____mm Overbite_____mm

V. TREATMENT RECORD:
Stage I:
Observation and record taking only: yes____ no____
Stage II:
a) Obturator: yes____ no____

b) Neonatal maxillary orthopedics: appliance yes____ no____
a. Hotz___ b. NAM___ c. Latham___
date started _____ date completed_____
age_____ age_____

c) Gingovoperiosteoplasty yes____ no____ date_____ age_____

d) Retention appliance yes____ no____ date_____ age____

VI. Deciduous dentition
a) Treatment:

fixed appliance yes____ no____
removable appliance yes___ no___
date started_____ date completed_____
age____ age____

1) Palatal Expansion:
yes____ no____
fixed appliance yes____ no____
removable appliance yes___ no___
date started_____ date completed_____
age____ age____

Stage III
a) Permanent Dentition: yes___no___
1. Upper Arch: appliance_____date started_____
2. Lower Arch: appliance_____date started_____
Date treatment completed_____
Still in treatment_____

Stage IV
a) Retention: yes____ no____
fixed appliance yes____ no____
removable appliance yes____ no____
fixed bridgework_____ none_____
b) Date treatment completed_____
and in Observation_____
Date treatment discontinued_____

VII. Surgery

a) Maxilla –
1. Lefort I alv: yes____ no____ age____
2. Distraction Osteogenesis yes____ no____ age____

b) Mandible –
1. Advancement (DO) yes____ no____ age____
(surg) yes____ no____ age____
2. Setback yes____ no____ age____
3. Symphysis (point PO)
a) advanced yes____ no____ age____
b) reduced yes____ no____ age____

c) Alveoloplasty –
1. Primary yes____ no____ age____
2. Secondary yes____ no____ age____
a) cortical bone yes____ no____ age____
b) medular bone yes____ no____ age____
c) BMP-2 yes____ no____

VIII. Nasopharyngeal analysis:

Diagnosis :

A. Soft Palate
1. Normal size____ 2. Thin____
3. Short_____ 4. Paralyzed_____

B. Tonsils
1. Absent____2. Large_____
3. Sparse____4. Medium____

C. Pharyngeal Depth
1. Shallow_____
2. Deep_____
3. Normal_____

D. Pharyngeal Wall Movement
1. Passavants Pad____
2. Lateral wall movement
a. good___ b. poor___ c. cannot determine___

IX. Speech:

A.
1. Normal____
2. Hypernasality____
3. Hyponasality____
4. Treatment: yes ___ no ___

B. Evaluation Using Cepths:

A.Velopharyngeal Speech Aid
a. on "U" good___poor___
b. on "S" good___poor___

C. Soft Palate Surgery:

a) None____
b) Pharyngeal____
c) Pushback_____
d) Both b) and c)____

D. None_____

E. Cranial Base
1. Normal____ 2. Acute____ 3. Obtuse____

F. Adeniod
1. Sparse____ 2. Medium____
3. Large_____ 4. Absent_____

X. Cervical vertebrae:

A. Normal____
B. Abnormal_____
1. Fusion C_2-C_3_____
2. Occipitalization of Anterior Tubercle of Atlas _____
3. Malposition of Atlas____

XI. Recommendations:

A. None____ B. Pharyngoplasty____ C. Pharyngeal Flap____
D. Speech Aid____ E. Adenoidectomy____ F. Tonsillextomy____
G. Speech Therapy_____
H. Other_____

i) Maxillary Distraction Osteogenesis _____
or
Orthognathic Surgery_____
ii) Mandibular Distraction Osteogenesis_____
or
Mandibular Orthognathic Surgery_____
iii) lip_____nose_____

I. Observation or Recall_____

Part XVIII

The Patient's World

49 Social, Ethical, and Health Policy Issues in the Care of Children with Major Craniofacial Conditions

Ronald P. Strauss

49.1 Introduction

Over the past decade, major contributions have been made to the literature regarding social, ethical, and health policy aspects of craniofacial care and conditions.

Strides have been made in the measurement of quality of life and in understandings of health status (Chetpakdeechit et al. 2009; Damiano et al. 2007; Kramer et al. 2008, 2009; Mani et al. 2010; Munz et al. 2011; Nelson et al. 2011a, b, c; Nusbaum et al. 2008; Patrick et al. 2007; Sagheri et al. 2009; Stone et al. 2010; Strauss and Cassell 2009; Wehby and Cassell 2010). Health services research has emerged as a major contributor to the literature on cleft and craniofacial conditions (Alkire et al. 2011; Austin et al. 2010; Blume and Henson 2011; Boulet et al. 2009; Cassell et al. 2012; Chuo et al. 2008; Foo et al. 2011; Furr et al. 2011; Knapke et al. 2010; Kuttenberger et al. 2010; Mendoza 2009; Nelson et al. 2011a, b, c; Payakachat et al. 2011; van Aalst et al. 2011). Psychological research has become more sensitive to the challenges, resilience, and health of children and adults with clefts and craniofacial conditions (Baker et al. 2009; Berger and Dalton 2009, 2011; Black et al. 2009; Brand et al. 2008; Feragen and Borge 2009, 2010; Feragen et al. 2010; Loewenstein et al. 2008; Marshman et al. 2009; Meyer-Marcotty et al. 2010; Murray et al. 2009; Mzezewa and Muchemwa 2010; Nelson et al. 2011a, b, c; Strauss et al. 2007).

A lively discourse on ethical issues and the values related to craniofacial care has emerged (Abbott and Meara 2010; Aspinall 2010; Bijma et al. 2008; Mossey et al. 2011; Nusbaum et al. 2008; Strauss et al. 2011). In this same period, patients and families have been understood to live within social and health system realms that define their experiences with their conditions. During this period, advances in technology and informational systems have altered our understandings of craniofacial conditions around the world (Corlew 2010; Rodrigues et al. 2009).

The focus of this chapter is on major craniofacial conditions in historical context. The chapter is a revised version of a 1992 paper which explored themes that have proven to be both enduring and salient. Craniofacial surgery and neonatal intensive care have made it possible for children with serious craniofacial conditions to live and often to experience effective habilitation.

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These therapeutic interventions also raise important social, policy, and ethical issues. This chapter reviews the dilemmas which relate to the gatekeeper role for physicians, the impact of prenatal diagnosis, and the allocation of scarce fiscal and health resources to craniofacial care. The high degree of cost, the intense investment of medical resources, and the uncertain outcomes in the care of children with major craniofacial conditions must be considered in the distribution of resources within a health system. The rationing of health resources may be a future determinant of how care for major craniofacial conditions is delivered.

49.2 Craniofacial Treatment Decision Making

"I felt shock, hopelessness and an overwhelming responsibility. With birth defects as bad as hers, it was hard to believe much could be done." – a 44-year-old mother of a 12-year-old child with Apert syndrome

The birth of a child with a major craniofacial condition is a serious crisis in the life of the family affected. Not only is the desire for a healthy baby unfulfilled, but the family must reframe their expectations for the child's needs. This may mean a reevaluation of who is to provide care to the infant, how health needs will be met, and how health costs will be covered. The family may also need to consider the future health, social, and developmental limitations associated with their child's condition.

In the context of family crisis, clinical decisions must often be made about the extent, timing, and nature of the treatment which is to be provided. Family values about the sanctity of life and norms for parental responsibility will guide decisions. Religious perspectives and ethnic or cultural values often provide the basis for making treatment decisions within the family (Botto et al. 2006; Florian and Katz 1983; Kleinman 1979). Other social influences may affect decision making and the available choices for parents and professionals.

Attitudes about disfigured children and about medical care may guide treatment decisions relative to children born with major craniofacial conditions. A fascination with disfigurement pervades mythology and literature (Fiedler 1978; Shaw 1981; Strauss 1985). Combinations of fear, repulsion, interest, and sympathy affect attitudes about the child with a defect. Significant social ambivalence may be generated by children with obvious conditions. This ambivalence may find itself reflected in how a family or a clinician decides about major craniofacial surgery.

The child who is perceived as having a limited role as a future community participant or worker may be devalued, and treatment resources may be directed toward the needs of other children. In some settings, children seen as hopelessly afflicted may be neglected, maltreated, or abused (Mosher 1983). In other sociocultural settings, the child with a defect may be nurtured, seen as an exceptional child with special needs, and provided with the full range of possible treatment options. Historical and anthropological studies of child treatment and maltreatment (Demause 1977) suggest that different cultures and different eras of time are characterized by markedly different attitudes and values placed on children and their health care needs.

Medical and societal resources and wealth will affect treatment decisions and options. In settings where treatment facilities are limited, the options for clinical activism may be severely curtailed. I am reminded of an experience with a rural mother who had just delivered a very premature, but otherwise healthy infant, in a regional hospital in a developing nation. The baby died soon after birth, there being no neonatal intensive care unit and limited medical expertise. The mother raised her head from the bed when I entered her room and asked me directly "would my baby have died if she were born in your country?" I knew the answer was that the baby would probably have been treated in the neonatal ICU and might be alive. How you might have answered her question will provide some insight into the complexities that surround treatment allocation. Medical facilities and health resources allow for

care in previously untreatable conditions and create options for medical decision making. They also raise new moral and ethical quandaries about who is to live and with what quality of life.

This chapter will examine the moral and health policy basis for making treatment decisions and will suggest social and ethical issues that may be raised in the care of children with major craniofacial conditions. It will consider four topics: (1) the history of care for children with major craniofacial conditions, (2) the gatekeeper role for physicians, (3) the impact of prenatal diagnosis, and (4) the social justice and resource allocation.

49.2.1 The History of Care for Children with Major Craniofacial Conditions

Until the last several decades, many, if not most, children with major craniofacial conditions did not survive or experienced a much reduced quality of life (Kleinman 1979; Scheper-Hughes 1987). This occurred primarily because of the inability to provide nutrition, craniofacial surgical care, or respiratory assistance. Sometimes, the lack of willingness to invest caregiving or resources to the infant with a birth defect was influential. Infanticide (Dickeman 1975; Pertschuk and Whitaker 1982) and the withdrawal of caregiving or sustenance (Langer 1974) have been discussed as possible responses to the infant with a marked defect. There is evidence that gender selection (Divale and Harris 1976; Scheper-Hughes 1990) and the lack of skill in caring for such a child (Mosher 1983) may result in the infant's death. Infanticide is now universally seen as a crime, and most societies have developed social roles for children with birth conditions or defects. In spite of this, infanticide or child maltreatment may still occur hidden behind closed doors.

The success of neonatal intensive care units and craniofacial surgery has made it possible to treat many serious birth defects that may once have resulted in a child's death. It is likely that the application of these advanced forms of technological assistance have changed the profile of cases treated by many cleft/craniofacial teams. While craniofacial teams may once have predominantly seen children born with uncomplicated clefts of the lip and palate, many teams now report receiving referrals of children with multiple defects and complicated craniofacial conditions (Gupta 1969).

This epidemiologic change is so substantial that many centers have changed their names from "cleft palate team" to "craniofacial team" (Strauss 1992). With this marked shift in focus, clinicians may find themselves pondering difficult ethical questions. They may ask: Should all children who have defects that might be repaired receive treatment? Should infants with major and handicapping craniofacial conditions routinely be offered treatment in the neonatal intensive care unit? Should they always get craniofacial surgical care? What criteria might be used to ration such care? Who should make the decision to treat or not to treat? To what extent does the physician serve as the agent of the society in directing the use of scarce and costly resources? In the context of a health care system, how many resources can be directed toward any individual child?

The advent of major craniofacial surgery, prenatal diagnosis, and neonatal intensive care raises basic social and ethical dilemmas relative to:

(a) Who should be treated and to what degree.
(b) How scarce resources (i.e., medical or surgical services or expertise) should be allocated.
(c) Whether and how cost/benefit calculations can be used in clinical decision making.
(d) What is the goal for major craniofacial surgery or treatment.
(e) How much does the society value children and adults with disabilities.

The extremely high cost of correcting a complicated craniofacial condition (Shprintzen 1990) suggests that the society must consider the value of surgical care. Is the cost of repeated surgeries recouped in the development of human potential? Does the patient achieve social integration and productive vocational performance because he/she was treated? If not, what was the rationale of treatment? Do surgeons and families seek

incremental improvements without regard to the final status and likely outcome of the process? Do surgeons have the capacity to manage access to their services? The role of the gatekeeper to medical and surgical services is a critical one, and the physician or the health insurer/managed care provider serves as an agent for the society in making treatment decisions.

49.2.2 The Gatekeeper Role for Physicians

The gatekeeper role (Bunker 1970; Lapham et al. 1996; Strauss 1983) involves the professional in exerting control over the availability of clinical treatment. Physicians encourage or discourage treatment based upon their legal responsibilities, values, and moral codes. Physicians are socialized during medical training to meet patients' needs and to deliver a community service. They guide patients in accessing care and by making referrals when specialized expertise is required. The basic precept is to be of assistance, do no wrong, and to protect the public and community health.

Professional time and patient financial resources are not endless; thus, health professionals serve to modulate and control access to their services, clinical skills, and their time. Surgeons and other clinicians are constantly deciding whether it is worthwhile to undertake treatment; thus, they manage the market for their services. Physicians offer only treatment that they perceive as necessary and of benefit for the patient or the community. Often, the surgical gatekeeper role is apparent when surgical treatment is denied. For example, a surgeon's decision not to operate on the palate of a nonverbal and developmentally delayed child, until the child begins to speak, is an application of the gatekeeper role.

The gatekeeper role may also relate to how a clinician rationalizes the investment of medical or surgical care. Social and psychological rationales for care may reflect the clinician's judgment that nonbiologic benefits may result from structural change. For example, the surgeon's decision to do a rhinoplasty on a 14-year-old girl with a cleft lip and minor nasal asymmetry may be based more on expected changes in her social experience, rather than on her breathing ability. The use of psychosocial rationales for care is an application of the gatekeeper role.

The gatekeeping role allows physicians to manage many of the health care system's scarce resources, including the operating room and the intensive care unit. Physicians determine who needs surgery and who requires highly technical hospital-based management. Insurance companies, managed care organizations, and other third-party payment agencies have developed mechanisms to monitor the use of these costly resources. The routine use of second surgical opinions, case managers, and insurance policy exclusions have been used to control medical activism and expenditures. As a health care system changes and health care, each patient and family may have a unique experience interacting with their payer, if covered at all. Physicians and other health professionals have expressed considerable dissatisfaction with the adversarial relationship that sometimes exists between the professional and the payer/insurer of care.

The responsibility of being a gatekeeper implies a considerable amount of control and power over one's work. Controlling the access to and use of one's own services may imply a conflict of interest. The gatekeeping role may be placed under stress when a physician is financially rewarded for being overly active about care. When a surgeon who is charged with deciding about how much care is necessary for a patient is also to receive financial reward from performance of the treatment, ethical roles may be stressed. This also may occur when outside parties, such as insurance companies or managed care organizations, are advocates for limiting the provision of care. They too have a gain from limiting the amount of service realized by a given patient or client.

In the matter of craniofacial clinical decision making, the clinician serves as the agent of the society, making judgments which balance a variety of inputs. Factors which may play a role in these judgments include the likely benefits to the patients, the risks of treatment, the family

capacity to cope with the care of the child, the child's long-term prognosis, the cost of care, and the ability to access financial resources to pay for care.

The clinician's judgment may also relate to career goals and the mission of medicine. The physician may feel charged by the society with the mission of "normalizing" children born with birth defects. Furthermore, the desire to perform dramatic and highly technical treatments on the "cutting edge" of medicine may be appealing. Both goals imply the likelihood of providing a patient with sophisticated and technical care. The desire to further the science and the clinical discipline by performing novel treatments may serve as a stress on the gatekeeper function.

Physicians primarily define their role in terms of the individual relationship with the patient and the family. Many physicians are educated to see themselves as advocates for the child patient as well as the parents and have difficulties in moments where the interests of patients and parents may not coincide. For example, in a situation where a child has a serious defect that would require intense and costly care and the family expresses a wish to limit treatment, the clinician may be placed in a difficult position. Whose interests does the physician protect? Who advocates for the child's interests? Does the tendency to medically intervene generally prevail? Do the parents always have the final say relative to a treatment decision? Legislative and judicial decisions have become central in guiding medical decision makers and in defining the limits of medical autonomy (Chetpakdeechit et al. 2009).

Medical education socializes Western physicians to be activist in their clinical responses. In simple words, due to their training, doctors try to fix disabilities when possible. The decision not to offer care, or to withdraw care already started, is always difficult and places the physician in struggle with well-socialized values to heal. In new or innovative forms of treatment, few guidelines or norms may exist for deciding about whether to intervene. In the case of craniofacial surgery, activist principles and the desire to fix disabilities predispose the physician to providing treatment whenever possible.

Periodically in the life of a profession, specific technological innovations markedly alter the course of medical practice and thinking. This occurred with the introduction of the stethoscope, the aseptic technique, and the use of antipsychotic medications. This also occurred with the introduction of prenatal diagnosis and the introduction of craniofacial surgery. The introduction of these technologies has markedly changed how medicine deals with congenital disabilities. Prenatal diagnosis and ultrasound imaging have created a situation where parents have the option about whether to bear a child with a known defect. Craniofacial surgery has created the option to correct defects that may exist. Parents must weigh the pain of bearing a child with a birth defect and the expectation of surgical correction against their moral values and may have a choice about whether to abort the child. Craniofacial surgery and prenatal diagnosis are both possibly major components in the family and parental decisions about the child with a major craniofacial condition.

49.2.3 The Impact of Prenatal Diagnosis

Prenatal diagnosis and imaging of major craniofacial conditions is increasingly common (Bosk 1992; Eng et al. 1997; President's Commission for the Study of Ethical Problems in Medicine and Biomedical and Behavioral Research 1983). The use of ultrasound allows for the early visualization of fetal defects. Techniques such as amniocentesis, genetic screening, and risk appraisal may provide the family and physicians with information about a child's craniofacial condition prior to birth. The knowledge of a condition during the prenatal period implies the possibility of choice relative to the birth of a child with a defect.

Knowledge about defects prior to birth has caused medicine to face profound questions about how we as a society will deal with prenatal information about serious craniofacial conditions. Will there be significant pressure on mothers to abort such fetuses in order to avoid the cost and

pain of corrections? Will there be rewards for parents who decide to save the society the cost of treatment? Will we stigmatize and harass parents for seeking to terminate a life, regardless of quality, by abortion?

The realities of the high cost associated with the survival and treatment of children with major craniofacial defects may affect how we perceive a parent decision to abort a fetus with an identified condition. A decision to bear such a child implies major expenditures for the parents and/or the society.

Advocates against the availability of abortion will eventually clash with the reality that some parents of fetuses with significant conditions may wish to terminate the pregnancy. Indeed, prenatal diagnostic technology is based on the premise that some action may occur in response to an untoward finding on testing. Some parents may merely be seeking prenatal information to be prepared for the birth of their child. Other parents may want to arrange for adoption of a special need child, while still others will seek abortion. Social policy debates will determine if abortion remains available or if it becomes illegal. The debate may consider if society is willing to pay the costs of maintenance and rehabilitation of congenitally disabled children and adults.

Social values relative to aesthetic and functional conformity are also challenged by prenatal diagnosis of birth defects. Will infants who are different or have birth defects be aborted or be accepted? How major a condition must there be to rationalize an abortion? Will there be less tolerance of different appearance or identity if those with different identities are aborted or corrected? To what extent does living with a range of appearances and disabilities humanize a society? Some would argue that medicine should exert itself to make children look as normal as possible to meet social expectations. Others would say that instead of changing the child or his appearance, the focus should be on changing social values to encourage the acceptance of those who appear different. For the moment, there is little question that those who look different are treated in many societies as less than equal and often are stigmatized.

The introduction of the current array of sophisticated prenatal screening mechanisms did not imply that medicine was prepared for the social and ethical ramifications of their use. One would hope that the introduction of new biomedical technologies implies the willingness to consider the complex issues these procedures raise for clinicians and for those engaged in health care financing and policy.

49.2.4 Social Justice and Resource Allocation

It is sometimes argued that when limited resources exist, health dollars are best spent on prevention, rather than treatment. Some would state that health and economic resources are most rationally invested in maternal health care and in the application of established preventive regimes. Thus, it could be argued that prevention of fetal alcohol syndrome is preferable to focusing resources on its treatment. Most would agree that when possible, a preventive approach is preferable.

A health system, however, often functions to support treatment, as opposed to preventive, approaches. In the case of cancer and heart disease, known risk factors might be reduced, yet the bulk of health dollars are spent on the treatment of existing conditions. In the case of major craniofacial conditions, resources might best be spent on research and on the prevention of defects. This might involve developing programs that encourage maternal health and nutrition, genetic counseling and screening, as well as prenatal ultrasound/diagnosis. A reasonable approach to health policy might be built on research into craniofacial health promotion and disease prevention.

The major craniofacial disorders are however rare and currently are often of unknown etiology. Given the paucity of preventive craniofacial efforts, the bulk of resources are currently focused on the clinical treatment of these conditions. In the face of the reality of a child who is born with a major craniofacial condition, all attention is understandably drawn to the clinical treatment possibilities.

Physicians are not socialized to ask questions about social justice when encountered with a specific patient's needs. Few would find it reasonable for a clinician to worry about whether the society's resources are better spent on public health or immunization, rather than on expensive surgical reconstruction. Most citizens would agree that public dollars would be better invested in the realm of prevention, and yet when faced with the denial of reconstruction to the individual patient, due to limited resources, the social will to persevere diminishes.

Under circumstances of individual patient need, it is difficult to pose questions about the appropriate allocation of economic and clinical resources or social justice. In the context of ethical discussion, one may consider questions of fairness and equity.

Values on the sanctity of the individual are strongly held, and rationing of health care has proven to be a difficult prospect (Blumenfeld et al. 1999; Churchill 1987), though it happens in a de facto way regularly. Rationing of health care occurs, but it often is by limited insurance or policies, total lack of health coverage, or because of a scarcity in providers or hospital facilities. For the individual clinician, these barriers to care may not be apparent since many patients who reach his/her door are able to pay for care. The issues arise when payment is not available and when care is very costly. If patients cannot themselves afford care, and insurers are unable or unwilling to pay, then the true impact of rationed clinical care becomes apparent.

Health care is already allocated in many societies. Poverty and ethnic minority status may result in lower health status when compared to more affluent or nonminority populations (Aaron and Schwartz 1984). In a society where marked disparities in access to health care exist, one must ask whether the ability to pay is the most just medium for deciding who gets care. In the case of major craniofacial conditions, should ability to pay for expensive services guide their availability?

Insurers, case managers, or governmental agencies may determine who receives medical care. What values guide their decisions? Do they seek to provide maximum good to the largest number of citizens? Do they seek to provide a basic level of good to all? Do they seek to meet the demands of special groups of needy patients? In determining benefits, who speaks for the interests of children? In the case of costly procedures that benefit very few persons, political advocacy seems unlikely to be a large force in guiding insurance coverage. Principles of social justice do come into play when decision makers consider how much pain would occur if care were denied, or how much gain can be achieved by funding treatment. These distinctions are difficult and cost-benefit thinking may be used in making such funding decisions.

Decisions about making extraordinary expenditures on highly technical craniofacial surgery and care may be deliberated by health agencies and insurers. In group policies, the cost of catastrophic illness or a major birth defect in a single member or child may affect the entire group and its cost of insurance. There are many examples of health insurance programs that seek to avoid expensive claims or that withdraw continued coverage from the neediest claimants.

In calculating coverage and policies, insurance companies or systems are not likely to consider how the cost of a craniofacial repair might compare with the costs of maintaining a person's disability. Such companies seek to minimize their costs and are not responsible for dealing with the long-term social consequences of a disability. Governmental agencies, on the other hand, may ask questions about long-term patient function. They may consider whether the person will be self-supporting and self-maintaining, in the long term, after care. A decision to provide costly care to a child may reflect the perception that treatment will reduce the burden to society of lifelong assistance or maintenance.

Another approach to cost-benefit considerations recognizes the social cost of living around deformed persons, who may inadvertently challenge social norms of acceptability. Some conditions elicit negative social responses that provoke society to cover up the difference or reduce the deviance. It is in this fashion that significant social pressure may actually be placed on the

person with a major craniofacial condition to participate in and agree to reconstruction. The person is expected to "be a good patient" and seek amelioration.

Perhaps, the most difficult resource distinction occurs around the relative worth of social investment in various diagnoses. Why is a cleft palate repair seen uniformly as cost effective, while some question a kleeblattschädel craniosynostosis repair? Is it that we expect a better outcome from the cleft surgery? Or is it that the frequency of clefts demands their repair, while the scarcity of major craniofacial syndromes does not? If we can afford as a society to provide cleft repairs, why then not afford craniofacial repairs? Clearly, questions of the magnitude of cost exist. The costs as calculated cannot be merely the direct costs of several years of surgical care; rather, they must also include the costs of parental work loss, of special education, of rehabilitation services, of mental health assistance, of lost patient productivity, and of mental anguish. These costs are generally immeasurable, yet they constitute real components of the losses realized.

In the current context of limiting health dollars and resources, it is predictable that some rationing decisions will occur. The deciding principles to be applied are unclear, but they will determine who will live, who will live well; who will be cared for and who will languish; who will receive benefit and who will suffer. These distinctions will directly affect how care for major craniofacial conditions will be delivered.

49.3 Justice and the Distribution of Resources

This chapter has sought to review the social and ethical ramifications of craniofacial surgery for children born with major craniofacial conditions. The remarkable tools of craniofacial surgery and neonatal life support/intensive care have made it possible for many more children with serious defects to live. Their survival has raised questions about how much the society is willing to invest in their care, especially considering the impact that even a repaired defect will have on the patient's quality of life. The availability of prenatal diagnosis and abortion may present additional choices to parents and clinicians. After a birth of a child with a major birth defect, the intense expense and the high degree of clinical activism involved heighten our perception of the costs of craniofacial surgery. The desire to utilize new techniques, to advance a discipline, and to provide patient benefits may affect clinician decisions. In the context of these often conflicting social currents, many questions about justice and the distribution of resources arise (National Center For Health Statistics 1990). Should society invest large amounts of effort and resources in costly corrections of rare and disabling conditions, or should it spend limited health dollars on population-based preventive efforts (Beauchamp and Childress 1994; Zuger 2004)? Decisions about rationing care will arise in the context of limited resources and in societies where demands on the health system exceed the system capacity. Craniofacial care poses many social, ethical, and health policy issues (Demause 1977; Feragen et al. 2009; Hall 1992; Jonsen et al. 1992; Murray and Botkin 1995; Roberston 1986; Strauss et al. 1995; Ward 1995; Wertz and Fletcher 1989; Wexler 1995) for health professionals, parents, and the community at large.

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Index

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