Cleft Lip and Palate

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Dina Emam, Aya Elderee,
and Abdelrahman Alsaleh

65.1 Introduction

Cleft lip and cleft palate are openings or splits in the upper lip, palate, or both (Figs. 65.1 and 65.2). Cleft lip and cleft palate result when facial structures that are developing in fetus don't close completely.

Cleft lip and cleft palate are among the most common birth defects. They most commonly occur as isolated birth defects but are also associated with many inherited genetic conditions or syndromes.

In most babies, a series of surgeries can restore normal function and achieve a more normal appearance with minimal scarring.

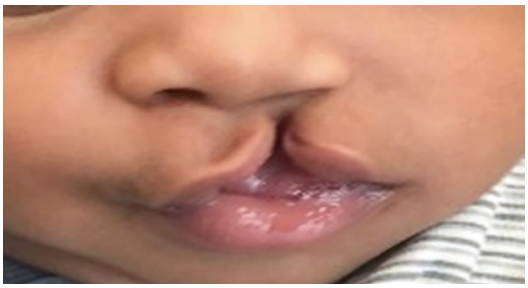

Fig. 65.1 A child with unilateral cleft lip

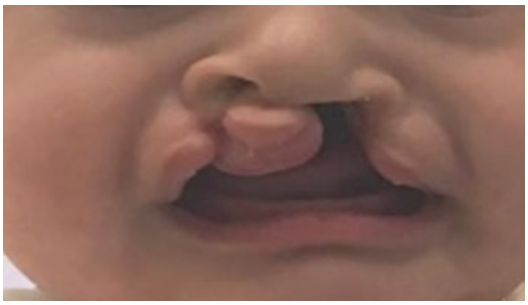

Fig. 65.2 A baby with bilateral cleft lip and palate

65.2 Embryology

- The embryologic development of the face begins at 4 weeks after conception from the neural crest ectomesenchyme that forms five prominences; the frontonasal process, and paired maxillary and mandibular processes surrounding a central depression.
- During the fifth and sixth weeks of embryonic development, bilateral maxillary processes derived from the first brachial arch fuse with the medial nasal process to form the upper lip, alveolus, and the primary palate.
- The formation of the secondary palate begins during the sixth week after conception from the two palatals shelves, which extend from the internal aspect of the maxillary processes.
- During the eighth week, these bilateral maxillary palatal shelves, after ascending to an appropriate position above the tongue, fuse with each other and the primary palate.

D. Emam (✉) · A. Elderee · A. Alsaleh
ENT Department, Hamad Medical Corporation,
Doha, Qatar
e-mail: DEmam@hamad.qa

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– A disruption in the fusion of these embryonic components can occur due to delay in elevation of the palatal shelves from vertical to horizontal, defective shelf fusion, or post-fusion rupture resulting in a cleft of the secondary palate.
– The lateral nasal process forms the alar structures of the nose.
– The lower lip and jaw are formed by the mandibular processes.
– This process of formation of the face is the consequence of a cascade of processes that involve cell proliferation, cell differentiation, cell adhesion, and apoptosis.
– Failure or error in any of these cellular processes that lead to fusion of the medial nasal process with the lateral nasal and maxillary process can cause orofacial clefts.

65.3 Epidemiology (Fig. 65.3)

– Cleft lip and palate are the second most common congenital malformation after the club foot, which is more common.
– Interracial differences exist in the incidence of cleft lip and palate versus cleft palate.
– The mean incidence of cleft lip and palate is 2.1 cases per 1000 live births among Asians, 1 case per 1000 live births among white people, and 0.41 cases per 1000 live births among black people.
– A high incidence of the cleft lip and palate are seen in North American populations of Asian descent, such as Indians of the southwestern United States and the west coast of Canada.

The incidence of isolated cleft palate is constant among the three racial groups at 0.5 cases per 1000 live births.

– The incidence of cleft lip and palate also rises with increased parental age, and older mothers with additional parity have an increased incidence of having children with cleft palate.
– In relatives of children with cleft lip and palate, the incidence of cleft lip and palate is significantly increased. However, the isolated cleft palate anomaly occurs with the same frequency as that in the general population.
– Relatives of children with isolated cleft palate also have a higher risk of this anomaly, without an increased risk of the cleft lip and palate deformity.
– Overall, 5% of patients with cleft lip and palate and isolated cleft palate have identifiable syndromes.
– Associated syndromes are more common among patients with isolated cleft palate than among others.

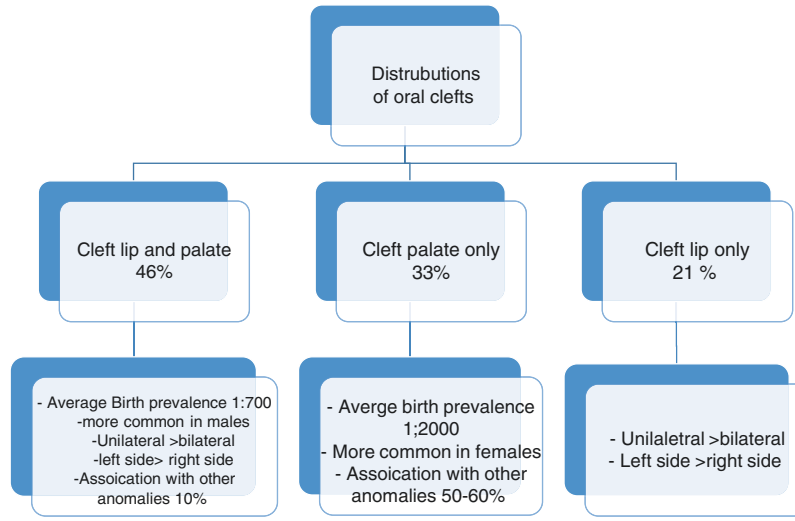

Fig. 65.3 Epidemiology of cleft lip and palate

– Congenital heart diseases are more prevalent in patients with non-syndromic cleft lip and/or palate than in the general population.

65.4 Etiology and Genetics

Non-syndromic CLP is a complex trait with multifactorial etiology resulting from gene–environmental interactions:

- More than ten genes contribute to the development of cleft lip and palate.
- Environmental factors that contribute to the etiology of facial cleft disorders include cigarette smoking; folic acid deficiency during the periconceptional period; and maternal exposure to alcohol and teratogenic medications such as retinoids, corticosteroids, and anticonvulsants (phenytoin and valproic acid).

65.5 Classification of Cleft Lip and Palate

65.5.1 Veau Classification

The Veau classification system divides the cleft lip and palate into four groups, which are as follows and illustrated in Fig. 65.3:

1. Group I—Defects of the soft palate only.
2. Group II—Defects involving the hard palate and soft palate.
3. Group III—Defects involving the soft palate to the alveolus, usually involving the lip.
4. Group IV—Complete bilateral clefts.

65.5.2 International Confederation of Plastic and Reconstructive Surgery Classification

The International Confederation of Plastic and Reconstructive Surgery classification system uses an embryonic framework to divide clefts into 3 groups, with further subdivisions to denote unilateral or bilateral cases:

1. Group I—Defects of the lip or alveolus.
2. Group II—Clefts of the secondary palate (hard palate, soft palate, or both).
3. Group III—Any combination of clefts involving the primary and secondary palates.

65.5.2.1 Submucous Cleft Palate: A Minor Form of Secondary Cleft Palate Defect

Causing a midline diastasis of the levator muscles with the mucosa remaining intact, deficient musculature of the palate causes a blue streak known as zona pellucida, associated with a bifid uvula and loss of posterior nasal spine (notched hard palate).

Risk of velopharyngeal insufficiency is increasing after adenoidectomy.

65.6 Evaluation

A child with a cleft lip and palate present with multiple challenging problems:

- *Airway problems*: Syndromic children, like those with the Robin sequence, for example, present with respiratory distress. Securing the airway is the priority in these patients.
- *Feeding difficulty is the primary problem*, and although these babies have normal sucking and swallowing reflexes, they have difficulty generating enough negative pressure to nurse adequately. As a result, the baby's nutrition must be delivered through bottle-feeding via nipples with large openings to facilitate the delivery of breast milk or formula.
- *Dental and orthodontic problems.*
- *Speech and language difficulties* as hypernasality due to velopharyngeal insufficiency.
- *Hearing impairment* because of Eustachian tube dysfunction leading to recurrent attacks of otitis media secondary to middle ear effusion which will need insertion of auditory tubes to improve the hearing.

Table 65.1 Interdisciplinary care of the cleft patient

Time line	Management
Prenatal	Diagnosis and parental counseling
0–6 months	General assessment for associated anomalies. ENT evaluation—breathing, feeding, swallowing, and hearing. Presurgical orthopedics (0–3 months). Primary lip repair (3–4 months)
6 months–2 years	Speech and oral sensory-motor assessment. Ear tubes (as needed). Primary palate repair (9–12 months)
Preschool: 3–5 years	Dental care. Speech assessment and therapy. Assess the need for lip revision
Childhood: 6–12 years	Correction of velopharyngeal dysfunction. Orthodontic treatment-Phase I. Alveolar cleft repair (8–11 years)
Adolescence: 13–18 years	Orthodontic treatment-Phase II. Orthognathic surgery (if needed) 14–16 years (female), 16–18 years (male). Revision Chielo-rhinoplasty. Replacement of missing teeth (as needed)

- *Social and self-esteem challenges* which lead to psychological and behavioral consequences.

65.7 Management of Cleft Lip and Palate

A multidisciplinary approach is required to assist patients and their families with the comprehensive care of these children. Responsibility for their care is shared by a team of pediatricians, plastic surgeons, otolaryngologists, pedodontists, orthodontists, nurses, speech therapists, audiologists, and social workers (Table 65.1).

Cleft lip and palate surgery should aim to achieve the following:

1. Isolation of the nasal cavity from the mouth.
2. Bone continuity throughout the maxillary alveolus to facilitate the eruption of the permanent dentition.
3. A functional velum that will permit normal speech.
4. An esthetic and functional lip and nose.

65.8 Surgical Repair of Cleft Lip and Palate

65.8.1 Unilateral Cleft Lip

Surgical Anatomy

- The most visible anatomic abnormalities of the complete unilateral cleft lip and nose deformity are due to the abnormal position of the orbicularis oris muscle. Orbicularis oris muscle is directed superiorly in the complete cleft or is hypoplastic in incomplete cleft, the floor of the nose communicate with oral cavity and the maxilla is hypoplastic in the cleft side.
- The nasal deformity is proportionate to the severity of clefting. There is slumping of the alar cartilage on the cleft side, leading to an asymmetric nasal tip.
- The alar base is displaced laterally, inferiorly, and posteriorly leading to a widened nasal aperture.
- There is shortening of the medial crus of the alar cartilage.
- The columella and the caudal edge of the septum and anterior nasal spine are deviated to the non-cleft side.

Repair Timing

- Repair of the unilateral cleft lip is usually performed during the first year of life.
- Follow the "rules of 10": hemoglobin more than 10 g, age older than 10 weeks, and weight more than 10 lb.
- A surgical lip adhesion may be preferred as an initial surgical procedure within 6–8 weeks after birth. Lip adhesion helps to align the maxillary alveolar segments and achieve a tension-free definitive lip repair.

Surgical Techniques

- The most popular technique was introduced by Millard (1955) who described the rotation advancement concept.
- In Millard's technique, the medial flap is rotated downward to achieve length, while the lateral flap is advanced. The advantage of this technique is that the suture line lies on the

recreated philtral column, and incision allows easy access for primary rhinoplasty to reposition the nasal septum, lower lateral cartilage, and alar base. The main disadvantage is that the inexperienced surgeon requires good surgical judgment during the operation as it is not based on exact measurements.

- Other Surgical Techniques include:
  The triangular flap technique, described by Tennison and Randall, is based on exact measurements, can be reproduced well, and used more easily in wide clefts of the lip.
  Millard rotation advancement technique.
  Tennison–Randall (single) triangular flap interdigitation.
  Bardach (double) triangular flap interdigitation.
  Bilateral cleft repair (Millard).

65.8.2 Bilateral Cleft Lip

Complete bilateral clefts of the lip are rare, accounting for only 10% of cleft lips, and therefore, the experience in treating these deformities is limited.

Surgical Anatomy

- The typical anatomical abnormalities that make the bilateral cleft lip deformity so difficult to repair are the absence of muscle in the prolabial segment, resulting in lack of philtral dimple, philtral columns, white roll margin, and the median tubercle; also the central part of the alveolar arch is rotated forward and upward.
- The floor of the nose communicates with the oral cavity bilaterally.
- The nasal columella is short, nasal tip is widened, and cartilaginous nasal septum and nasal spine are deflected forward.

Surgical Techniques

- A number of surgical procedures with many variations for the repair of bilateral cleft lip are well described. Among these are the repairs of Veau, Tennison, Manchester, Millard, and others.
- Bilateral cleft lip repair is much more challenging, and the results are often less satisfactory than those of unilateral cleft lip.

65.8.3 Cleft Palate Repair

Surgical Anatomy

- The muscles of soft palate that help with the function of speech and swallowing include the levator palatini, tensor palatini, palatopharyngeus, palatoglossus, and musculus uvulae.
- The soft palate in a non-cleft individual acts as a muscular valve that can lift superiorly and posteriorly to oppose the pharyngeal wall and achieve velopharyngeal closure during speech.
- In a child with an unrepaired cleft, the soft palate cannot function as a muscular valve leading to velopharyngeal insufficiency.
- The tensor palatini muscle fibers, which control the opening of the Eustachian tube, do not function optimally, often leading to chronic otitis media.

Repair timing: The timing of palate repair to achieve optimal speech with minimal facial growth disturbance has been one of the more debated issues in cleft literature. It is now well accepted, and evidence in the literature shows that speech outcomes are better when soft and hard palate repair is completed before speech development. For most children developing normally, this is around 9–12 months.

Principles and techniques of palate repair:

Surgical repair of the cleft palate falls into two categories. The first is a single-stage repair involving closure with mucoperiosteal flaps. The second involves a multistage approach in which the soft palate is closed initially, followed by delayed closure of the hard palate.

May require orthodontic treatment to move the premaxilla or to realign maxillary segments.

The main principle of cleft palate repair is to detach and retro pose the abnormal insertion of the levator palatini and join the muscles of both halves of the soft palate in the midline at the junction of the middle and posterior third of the soft

palate in order to achieve proper elevation of the soft palate.

In the hard palate, the most important principle is to reflect mucoperiosteal flaps posteriorly based on the greater palatine arteries.

At the time of the primary palatoplasty, the ears should be inspected. If there is evidence of serous otitis, a myringotomy is performed, and fluid aspirated with the insertion of ventilating tubes.

1. The two-flap palatoplasty is a commonly used surgical technique for repair of the complete unilateral and bilateral cleft of the palate.
2. The Veau–Wardwill–Kilner (V–Y pushback) technique: is retro position of the mucoperiosteal flap and soft palate, provides additional palate length, used less often for the repair of cleft of the secondary palate.
3. The double reversing Furlow Z-plasty was introduced by Dr. Leonard Furlow Jr. in 1978. This technique uses two reversed Z-plasties of the oral and nasal mucosa to repair the cleft, it yielded excellent speech results for primary cleft palate repair with minimal and acceptable rates of fistula formation, velopharyngeal insufficiency (VPI), and the need for additional corrective surgery.

Take Home Messages

- Clefts of the lip and palate are common congenital birth anomalies.
- Etiology is complex. Known associations are with antenatal anticonvulsants, steroids, maternal smoking, and alcohol. Some of the genes associated with orofacial clefting have now been identified.
- Cleft palate results from failure of bilateral palatine shelves (from maxillary processes) to fuse at midline with developing nasal septum (from frontonasal process and bilateral medial nasal processes).
- Cleft lip results from failure of fusion of maxillary swelling with the medial nasal process.
- Every child born with a CL/P should be thoroughly assessed by complete physical examination and necessary diagnostic tests to check for associated systemic abnormalities, including congenital heart, renal, or airway anomalies.
- Management of patients with cleft lip and palate requires a multidisciplinary approach delivered by a team that includes an otolaryngologist.

Complications of Palatoplasty

- Early complications include bleeding, airway obstruction, and wound dehiscence.
- Late complications include fistula formation, velopharyngeal insufficiency, and maxillary growth disturbance.

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