Multidisciplinary Management of Pediatric Voice and Swallowing Disorders

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Gratitude with humility does not begin to express what is in my heart for so many people. I give thanks to my co-editors, Matt and Maia, who tirelessly worked this book to completion. I thank the many authors who contributed to this book. I give thanks to my other specialty partners without whom we could not give the care that we do. I give thanks to the patients and their families for allowing me to join their family and help point their children in a direction to achieve their full potential.

I give thanks to Chuck Ford and Diane Bless for taking a chance on me as I started my career. I give thanks to all the cottonoids around the globe who are supportive brothers and sisters, and especially to Mark, Dana, and Mike. I give thanks to Sally Shott, Chuck Myer, and J. Paul Willging. For opening opportunity, I give thanks to Chuck Kashima and Paul Flint. I give thanks to David Hanson and Jack Jiang. To Charles W. Cummings, you started it all for me in otolaryngology as the man I dreamt to try to be. To Robin Cotton, there are no words that can express my gratitude for what you have shared in so many different ways.

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suggested different paths. She has set example for what to try to become. She has always been there for me. I love you and I always will.

I hope this book helps you in your journey, seeking knowledge and honing tools, so that you too may point your children patients in the direction to reach their fullest potential.

–J. Scott McMurray

Thank you to Jack Jiang, for introducing me to the larynx, and Tim McCulloch, for introducing me to the pharynx. Thank you to Scott McMurray, for fostering an interest in pediatric laryngology. Thank you to my daughter, Val, for her laugh, smile, joyful nature, and friendly disposition. Most importantly, thank you to my wife, Marisa, with whom I always have fun and whose support is unwavering, maternal instinct is natural, and patience while I worked on this book was unending; I love you.

–Matthew R. Hoffman

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–Maia N. Braden

Foreword

In over 100 years of collective practice, we have seen many changes in the ways voice and swallow problems are managed, largely due to new knowledge of structure and function, developments in technology, and transmission of information through a myriad of journals, e-records, and the Internet. The explosion of information has resulted in the development of subspecialities and evidence-based practices and the underlying knowledge that it is necessary to have a multidisciplinary team to best treat voice and swallowing disorders, a premise we expounded in our 1991 book, *Phonosurgery: Assessment and Surgical Management of Voice Disorders.*

The editors who conceived the current book have succeeded in creating a richly informative yet very readable comprehensive textbook on assessment and management of pediatric voice and swallowing disorders. All three editors are accomplished authors and clinicians practicing at the University of Wisconsin-Madison. J. Scott McMurray, MD, is the UWSMPH chief of Pediatric Otolaryngology; Maia N. Braden, MS, CCC-SLP, manages pediatric voice and swallowing problems at UWAF Children's Hospital; and Matthew R. Hoffman, MD, PhD, is a remarkably accomplished young senior resident with over 40 publications in major journals. They have selected some of the best and brightest clinicians in their respective fields to contribute chapters. Each chapter stands alone as a contribution to our mutual understanding of how to treat this pediatric population, and the combined chapters are likely to enhance the way clinicians practice.

As former teachers and colleagues of these authors, we are delighted that they articulate the importance of collaboration and communication in clinical practice. Both factors are particularly important in dealing with pediatric patients, where sensitivity, flexibility, and creativity promote optimal assessment and successful outcomes. Insofar as we teach by example, we are particularly gratified because these are principles we have embraced in our practice, teaching, lectures, and writings.

This book describes an effective multidisciplinary approach. We are introduced to clinician specialists skilled in care of pediatric patients with disorders of the aerodigestive tract. Along with the parents, otolaryngologists and speech-language pathologists usually play pivotal roles, often in collaboration with gastroenterologists, pulmonologists, and other specialists. Readers will obtain a wide range of these interacting roles so important in treating the pediatric population with voice and swallow disorders. The chapters articulate the importance of recognizing that children are not miniature adults and

that they need pediatric subspecialists who are cognizant of the disorders, issues, assessments, and treatments unique to the pediatric population.

Assessment of pediatric voice and swallowing problems are presented in this book by clinicians and scientists who participated in developing many currently used techniques. The techniques described cover the gamut of visualization; aerodynamic, acoustic, perceptual, quality-of-life, and manometric assessment procedures; as well as surgical and behavioral management practices. Notably, separate chapters are devoted to the problems typically seen in pediatric populations as well as chapters on issues dealing with singers and gender-affirming voice concerns. Relevant measures and concepts are explained with great clarity, making this book a valuable resource for persons who are beginners in the field as well as for veterans who have been practicing for years. The reader will be struck by an overarching strategic principle the authors embrace in assessment and management: *effective communication* with both patient and parent. Communicating with a young child can be challenging, especially with tasks that might cause discomfort or require active patient participation. This can cause the child – and possibly the parent – to become anxious and stressed. Successful examinations often require the examiner to be flexible by adjusting tasks to the patient's ability. An informed parent will appear less anxious, which can be reassuring to an apprehensive child.

The section on intraoperative evaluation addresses the basics of operating room setup. Topics include essential equipment, solutions to laryngeal exposure problems, and discussion of advantages and limitations of inhalation vs total intravenous anesthesia. Two exciting new technologies are presented that provide alternative approaches to intraoperative assessment and treatment: (1) optical coherence tomography, which is used in diagnosis and depth assessment of vocal fold lesions like cysts, scars, and papilloma, and (2) microendoscopy via minithyrotomy, which is to provide subepithelial access to Reinke's space.

We expect you will find many familiar things in this book to be reaffirming. You will also discover new things to enhance your practice. Hopefully, you will be struck by unexpected things that will stimulate your imagination and enrich your appreciation of what you do. We are so pleased to have written this foreword because the editors and contributing authors not only effectively articulate our vision of best practices, but most importantly, they advance the challenging field of treating pediatric patients with disorders of the aerodigestive tract.

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Preface

Kids are amazing. Taking care of kids is a gift and a joy. The triad of child, parent, and provider creates multiple layers of complexity that never cease to stimulate, and upon whom, a solid foundation can be formed for growth and development. Whereas the goal of adult and geriatric medicine is often to help a patient maintain their current but at times fleeting abilities, pediatric medicine focuses on pointing the patient in the right direction to achieve their own ultimate potential. With the correct assessment and the appropriate intervention, a tremendous future can be unleashed.

Disorders of the aerodigestive tract can impair a child's verbal communication and swallow function which can significantly hinder their personal and social development. As pediatric otolaryngologists and speech-language pathologists, we have the opportunity to help these children reach their full potential. Critical to achieving that aim is a multidisciplinary approach. This begins with collaboration between the otolaryngologist and speech-language pathologist. At our institution, this collaborative relationship was started and epitomized by Drs. Charles Ford and Diane Bless. They have served as leaders in our field and personal mentors to us.

Over the last two decades, our understanding of the numerous ways in which a child's voice or swallow can be altered has dramatically grown. The myriad and often complex interaction of acquired and congenital anomalies requires a detailed assessment with thoughtful attention before an accurate lifelong plan can be developed, discussed, and instituted. We know that not all children presenting to the otolaryngology clinic simply have nodules or reflux (though some do, as described in Chaps. 27 and 28) and that we can offer many of them more than observation and reassurance alone that symptoms will resolve with age.

As our understanding of voice and swallowing disorders has increased, the number of clinicians involved in the care of affected children has also increased. We now routinely work with gastroenterologists, pulmonologists, plastic surgeons, geneticists, physical therapists, and occupational therapists. Delivery of healthcare by this number of providers has been significantly enhanced by the creation of multidisciplinary pediatric aerodigestive clinics. At our institution, we call it the UW PACT or University of Wisconsin Pediatric Aerodigestive Care Team. Families can come from distances to see multiple specialists over the course of a single trip, attending clinics and undergoing operative assessments over a brief time span in order to better understand their child's disorder and receive a unified plan to help.

With this book, we sought to create a practical reference that would emphasize the collaborative relationships among clinicians that are critical to effective clinical care. Accordingly, most chapters are written by a physician and a speech-language pathologist. Furthermore, a straightforward framework for approaching, diagnosing, and managing each disorder is presented, including descriptions of relevant operative interventions. It is our hope that this could serve as a useful resource for not only otolaryngologists and speech-language pathologists but all members of the pediatric aerodigestive team and all other providers caring for children affected by voice and swallowing disorders.

We are very grateful to the authors who contributed to this book, without whose time and expertise, this would not have been possible. We are also grateful to Drs. Ford and Bless who provided valuable advice at the onset of the project and have provided valuable advice and mentorship throughout our careers.

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1 Pediatric Aerodigestive Programs: Role of the Core Team Members, Speech Language Pathology, Pulmonology, Gastroenterology, Otolaryngology, and Parent/Caregiver

J. Scott McMurray, Maia N. Braden, Matthew R. Hoffman, Vivek Balasubramaniam, and Dorota Walkiewicz

Overview

More and more, children who are medically complicated are surviving and flourishing as medicine advances and treatments improve. These medically complicated children pose specific and often difficult challenges as they present with congenital or acquired disorders of multiple organ systems that can impact breathing, swallowing, growth, and verbal communication. No other group of patients epitomize the need for an interdisciplinary team approach with a core group of specialists than patients with aerodigestive disorders. The interest and formation of specialized aerodigestive programs have grown globally as their efficacy, efficiency, and economy have been recognized [1–6]. As more teams developed independently, the need for a consensus has been recognized regarding the types of patients and the typical disorders evaluated, the basic and minimum structure and function of the team, and the quality measures that should be followed. Boesch et al. [7] were the first to use the

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Delphi method to obtain consensus about the structure and function of the aerodigestive program. The future of aerodigestive programs in general and the establishment of the aerodigestive society have been seminally shaped by this work, spawned by a desire to treat these complex patients well.

Based on the consensus developed by Boesch et al. [7], the following definition was developed for an aerodigestive patient. An aerodigestive patient is a child with a combination of multiple and interrelated congenital and/or acquired conditions affecting airway, breathing, feeding, swallowing, or growth that require a coordinated interdisciplinary diagnostic and therapeutic approach to achieve optimal outcomes. This includes (but is not limited to) structural and functional airway and upper gastrointestinal tract disease, lung disease because of congenital or developmental abnormality or injury, swallowing dysfunction, feeding problems, genetic diseases, and neurodevelopmental disability. Common conditions evaluated and treated through aerodigestive programs include structural or physiologic airway disease, congenital or acquired subglottic stenosis, chronic parenchymal lung disease, lung injury from aspiration or infection, gastroesophageal reflux, eosinophilic esophagitis, esophageal dysmotility or stricture, dysphagia, and behavioral feeding problems [8]. Piccone and Boesch [8] polled 50 programs in 31 states in the United States and compiled a list of common presenting conditions based on airway, pulmonary, gastrointestinal, feeding and swallowing, sleep, genetic, and neurologic disorders which are listed in Table 1.1.

There are a significant number of specialists that would be required to cover all of the possible conditions in children with aerodigestive disorders. Through consensus development by Boesch et al. [7], however, the list of essential core members whose input is required for all patients attending an aerodigestive program can be distilled to the following: care coordinator, nursing, speech language pathologist, pulmonologist, gastroenterologist, and otolaryngologist. An aerodigestive program should include these key players at a minimum.

Consensus was also achieved regarding the essential and defining functions and features of an aerodigestive team evaluation [7]. For maximal efficiency and efficacy, the care cycle for an aerodigestive patient would involve the following work flow: consultation request and care coordination, pre-visit intake, team meeting, prescheduling appointments and procedures, shared clinic visit, combined endoscopy with a single anesthetic encounter, wrap-up visit with the family, summary document, and provision of follow-up care if needed.

Table 1.1 Common aerodigestive presenting conditions

Airway	Choanal atresia
	Laryngomalacia
	Glossoptosis
	Vocal fold paralysis
	Laryngotracheoesophageal cleft
	Stenosis: glottic, subglottic, tracheal, transglottis
	Tracheobronchomalacia
	Tracheoesophageal fistula
	Tracheostomy dependence
Pulmonary	Chronic lung disease of prematurity
	Diffuse lung disease
	Asthma
	Bronchiectasis: aspiration, ciliary dyskinesia, immunodeficiency, post-obstructive
	Chronic respiratory failure
Gastroenterology	Gastroesophageal reflux
	Eosinophilic esophagitis
	Esophageal structure
	Failure to thrive
Feeding and swallowing	Swallow incoordination
	Oral aversion
	Behavioral feeding problems
Sleep	Obstructive sleep apnea
	Central sleep apnea
	Hypoventilation
Genetic	Trisomy 21
	CHARGE association
	Pierre Robin sequence
	22q11 deletion
	VATER/VACTERL
	Craniofacial syndromes
	Opitz syndrome
	Cri du chat
Neurologic	Static encephalopathy
	Chiari malformation
	Hypotonia

Adapted from Piccione and Boesch [8], with permission

The typical aerodigestive program will see patients with a mix of medical and surgical needs. The interdisciplinary approach is important to effectively manage and plan the order of events leading to maximization of medical and surgical interventions and outcomes. Piccione et al. [8] also emphasized that there are several consistent structural elements of an aerodigestive program, namely, a (1) interdisciplinary medical and surgical team, (2) care coordination, (3) team meeting, and (4) combined endoscopy.

The team meeting is essential. This allows for distillation and review of historical events and prior tests. This information may be obtained through a telephone-based intake with caregivers and acquisition of previous medical records. This review will help to formulate a patient visit itinerary based on the team review and available best practice guidelines. It will help to ensure that a complete evaluation will be afforded in a short and convenient time without needlessly repeating tests with the associated cost and risk. The telephone contact is also a great opportunity to council the family about expectations. The multidisciplinary visit can be overwhelming with the total number of interactions and the length of the overall day. Families are often thankful despite the long day once they realize the extent and expedience of the evaluation they will receive. The itinerary will include essential laboratory tests, radiographs, and swallow studies leading up to the clinic visit with the core provider team. The team visit confirms historical and physical findings and affirms the need and plan for the endoscopies and adjuvant tests requiring anesthesia. Piccione et al. [8] compiled the common aerodigestive diagnostic tests which are adapted in Table 1.2.

Each of the core specialists will bring their perspective and process for evaluating the chief

Table 1.2 Common aerodigestive diagnostic tests

Diagnostic modality	Strengths	Weaknesses
Chest radiograph	Identification of lower respiratory tract disease	Low sensitivity for bronchiectasis
	Low radiation	Limited ability to differentiate causes of lung disease
Chest CT	Distribution and severity of lung findings of various types	Increased radiation
	Differentiation between airway and parenchymal disease	May require sedation for good imaging
Upper GI series	Evaluation of anatomy: peristalsis, stricture, hernia, gastric outlet obstruction, malrotation	Does not evaluate reflux
Radionucleotide reflux scan	Physiologic conditions	Limited sensitivity
	May document aspiration from reflux	
Radionuclide salivagram	Assess for aspiration of saliva	Poor sensitivity
Radionuclide parotid scan	Assess function of major salivary glands	
FEES (fiber-optic endoscopic evaluation of swallowing)	Evaluate functional anatomy of swallowing	Blind to moment of pharyngeal swallowing and esophageal phase
	Evaluate airway protective reflexes	Not widely available
	Portable	
	No radiation	
VFSS (videofluoroscopic swallowing study)	Evaluates all phases of swallowing	Radiation exposure
	Evaluates for aspiration	Limited anatomic evaluation
Microlaryngoscopy and rigid bronchoscopy	Superior optical resolution	Difficult access to peripheral airways
	Evaluation of the posterior larynx	Limited assessment of airway dynamics
	Access for instrumentation	Requires anesthesia

(continued)

Table 1.2 (continued)

Diagnostic modality	Strengths	Weaknesses
DISE (drug-induced sleep endoscopy)	Assessment of anatomic site of obstruction during sleep	Only an approximation of sleep state
		May miss REM specific obstruction
Flexible bronchoscopy with lavage	Evaluation of static and dynamic airway lesions, nasal-bronchial	Limited evaluation of posterior larynx
	Access to difficult and peripheral airways	Limited optical resolution
	Evaluation of airway inflammation and infections	Access for instrumentation
		Requires anesthesia
Esophagogastroduodenoscopy (EGD)	Evaluation of esophageal mucosal disease: acid and eosinophilic	Requires anesthesia
	Evaluation of esophageal, gastric, and duodenal anatomy	
	Obtain intestinal secretions	
	Evaluation of celiac disease	
Esophageal impedance	Identification and characterization of acid and nonacid reflux	Lack of normative data
	May identify dysmotility	Unclear relationship between impedance indices and extra-esophageal disease
Motility studies	Gold standard for dysmotility	Not widely available
Polysomnography (PSG)	Characterization of sleep disordered breathing and sleep architecture	Expensive and cumbersome
	Titration of respiratory support	Availability issues

Adapted from Piccione and Boesch [8], with permission

complaints and symptoms presented by the patient [8]. Although the group encounter, with all present for the clinic interview and the operative endoscopies, has been found to be the most efficient and efficacious, each provider brings unique and individual expertise. Each of the four core disciplines has overlap but also bring a unique role in the evaluation of these complex patients. The role of each core discipline will be outlined in this chapter.

Role of the Speech Language Pathologist

The multidisciplinary voice, swallow, and aerodigestive team can provide comprehensive, patient-centered and evidence-based care for children and adolescents with a variety of disorders impacting voice, swallow, and upper airway. Multidisciplinary team management of aerodigestive disorders in children has been found to be more cost-effective and has better outcomes than stand-alone care [6]. In voice disorders, the model of speech language pathologist and otolaryngologist working together in evaluation and treatment has been well established since the 1980s and became more common in pediatric voice around a decade later. The strength of these teams lies in both the diverse knowledge and skills of the team members and their ability to work collaboratively to evaluate and treat the patient. The speech language pathologist specializing in these areas provides a focused set of knowledge and skills for these patients. We can provide evaluation of structure, function, and behavior of upper airway as they relate to voice, swallow, and breathing. In many cases we can also provide behavioral therapy to change voice, breathing, and swallow function, provide education, and provide compensatory strategies when needed. We have specialized knowledge of laryngeal structure and

function; the mechanics of voice, breathing, and swallowing; and neurologic controls of voice, swallow, and breathing. We provide valuable contributions with our in-depth understanding of behavior change. On any medical team, but especially with complex children, we do not operate in a vacuum and collaborate with surgical and medical personnel in both evaluation and treatment. According to the American Speech-Language-Hearing Association's scope of practice statement, "SLPs share responsibility with other professionals for creating a collaborative culture. Collaboration requires joint communication and shared decision making among all members of the team, including the individual and family, to accomplish improved service delivery and functional outcomes for the individuals served" [9].

Evaluation of Swallow

Dysphagia is relatively common in children. A rate of 0.9% was found in children aged 3–17 [10], and incidence is higher in certain medically complex populations including those with cerebral palsy and craniofacial syndromes [11–13]. There has been a marked increase in diagnoses of dysphagia in the pediatric hospitalized population, from 0.08% in 1997 to 0.41% in 2012 [14]. While exact reasons for this are not clear, it is often attributed both to increased survival rates of extremely preterm infants and improved diagnosis of swallowing disorders. Often the SLP is the first contact a child with dysphagia has with the multidisciplinary team. Children may be referred directly to us for a swallow evaluation or for treatment of feeding or swallowing disorders, or we may care for the child in the NICU from birth. We have the benefit of being able to spend the time to get a comprehensive history and provide ongoing assessment in therapy sessions. The SLP has several methods of evaluating swallowing, including the clinical swallowing evaluation, flexible endoscopic evaluation of swallowing, and video fluoroscopic swallowing study, as well as less frequently used measures including manometry. These may be used in combination depending on the needs of the patient. According to ASHA, the role of the SLP in evaluation includes participating in determining the appropriateness of instrumental evaluation and follow-up, diagnosing pediatric oral and pharyngeal swallowing disorders, making appropriate referrals to other disciplines, and recommending a safe swallowing and feeding plan [9].

We require the expertise of others when evaluating and planning treatment beyond swallow recommendations for structural and functional deficits impacting swallowing, including (but not limited to) neurologic impairments, cerebral palsy (CP), sensory deficits, tracheoesophageal fistula, laryngeal cleft, acid reflux, esophageal dysmotility, laryngeal mobility impairment, and neurologic disorders.

Treatment of Dysphagia

SLPs on the multidisciplinary team as well as our colleagues working in more general outpatient settings, birth to three, and schools provide feeding and swallowing therapy to habilitate or rehabilitate swallowing and progress feeding skills. Feeding is defined as any aspect of eating or drinking and includes preparing food or liquid for intake, sucking or chewing, and swallowing [15]. Swallowing specifically refers to the complex processes involved in transporting solids, liquids, or saliva from the mouth to the digestive tract while maintaining airway protection [15]. Speech language pathologists are involved in evaluation and treatment of both.

A detailed description of all forms of feeding and swallowing therapy is beyond the scope of this chapter. Approaches to swallowing treatment may include positioning changes, changes in viscosity of bolus, changes in flow rate of bolus, maneuvers, sensory stimulation techniques, oral motor treatments, pacing, and cue-based feeding [15].

Evaluation of Voice

Incidence estimates of pediatric dysphonia are varied, ranging from 1.4% [10] to 26% [16]. Dysphonia rates in children are likely increasing

for some of the same reasons dysphagia rates are increasing, and children are presenting with more complex etiologies of voice disorders, beyond benign lesions. As survival rates of children born extremely preterm, or with complex tracheal or laryngeal anomalies, increase, rates of hoarseness and the complexity of children seen in the voice clinic will increase. For example, 38% of a sample of children born extremely preterm were found to have moderate-severe dysphonia at school age, with only 6% having normal voice [17]. Speech language pathologists often work in collaboration with an ENT in diagnosis and evaluation of voice disorders. According to the ASHA scope of practice, SLPs can perform a comprehensive voice evaluation which includes clinical and instrumental evaluation, assess normal or abnormal vocal function, describe voice quality and function, diagnose a voice disorder, refer to appropriate professionals to provide diagnosis of the underlying cause of the voice disorder (e.g., nodules as a cause of dysphonia), and make referrals to other professionals for other medical, surgical, or behavioral evaluation [9]. We can perform perceptual, acoustic, and aerodynamic evaluation of vocal function. We can also visualize the larynx using rigid or flexible endoscopy with stroboscopy, as well as high-speed digital video imaging of the larynx, and provide skilled interpretation of structure and function based on this. We do not diagnose lesions but can identify and describe the parameters of laryngeal function based on these evaluations and contribute to planning treatment, whether it be behavioral, surgical, or a combination of the two. The voice evaluation is also an important time to assess for stimulability for change based on therapeutic probes.

Treatment of Dysphonia

SLPs in a voice clinic and in other settings plan and deliver skilled treatment to optimize vocal function given the current anatomy, provide pre- and postoperative therapy, and provide therapy to change ingrained vocal functional behaviors. A detailed discussion of the types of voice therapy provided is beyond the scope of this chapter but can be found in other sections of this book and in these and other resources [18–23].

Evaluation and Treatment of Breathing Disorders

Speech language pathologists are also experts in evaluation and management of laryngeal breathing disorders such as paradoxical vocal fold motion disorder, exercise-induced laryngomalacia, and chronic cough [9]. We can behaviorally and endoscopically evaluate laryngeal, pharyngeal, and respiratory function during breathing and provide interventions related to laryngeal sensitivity and control as well as optimizing respiratory coordination [24–28].

Conclusion

The benefits of working as a part of a multidisciplinary team cannot be overstated, for both clinician and patient. We are able to evaluate based on our areas of expertise and then discuss with other team members based on the findings of their specialized evaluations, providing optimal treatment for patients.

Role of the Otolaryngologist

As a specialist of disorders of the upper aerodigestive tract, the otolaryngologist shares the pathway to both the lungs and the gastrointestinal tract. This unique perspective positions them to be able to relate to both the pulmonologist and gastroenterologist. Working in conjunction with the speech language pathologist, the otolaryngologist can help assess the anatomy and physiologic function of the upper aerodigestive tract. Medical treatments of aerodigestive disorders in children are likely made in conjunction with the gastroenterology and pulmonology regarding reflux, inflammation, or infection. Dynamic surgical interventions of the airway may be suggested after functional assessment in collaboration with the speech pathologist. The typical

aerodigestive problems evaluated by the otolaryngologist can be seen in Table 1.1. The role of the otolaryngologist centers primarily on evaluation of airway surgical issues and aspiration [8].

The otolaryngologist should elicit history specific to obstructive sleep apnea, voice and swallowing disorders, recurrent infection, previous surgical history, or instrumentation of the airway. An assessment of possible congenital or genetic disorders is also essential. Growth and weight gain curves are helpful to assess potential feeding or breathing problems.

The otolaryngologist can offer expertise in office and operative endoscopy to evaluate function and anatomy. Identifying sites of abnormal anatomy, obstruction, or function of the upper aerodigestive tract is the prime modality offered. Expertise in nasopharyngoscopy in the awake patient facilitates anatomical and functional evaluation for airway obstruction, voice disorders, and swallowing dysfunction. Expertise with flexible endoscopy with the patient in a state mimicking sleep is also essential for identifying sites of obstruction causing obstructive sleep apnea. Drug-induced sleep endoscopy (DISE) protocols continue to be developed to bring the patient as close to a state of true sleep as possible [29]. Typical dense general anesthesia for airway endoscopy changes muscular tone and can change the site of obstruction that occurs during this type of sleep and can misdirect the clinician during the evaluation. Accurately identifying the true site of obstruction during normal sleep is required to allow for successful surgical management of obstructive sleep apnea.

Plain radiographs of the airway are often helpful during the assessment of the airway and may be ordered by the otolaryngologist. PA and lateral plain radiographs of the upper aerodigestive tract and chest are often helpful. This affords an assessment of the upper airway and trachea. Obstruction from adenoidal hypertrophy (Fig. 1.1), subglottic narrowing, vascular compression, or complete tracheal rings may first be identified or suspected in these films which are easy to obtain. This will help prepare the team for operative endoscopy and prevent unsuspecting catastrophe in cases such as complete tracheal rings.

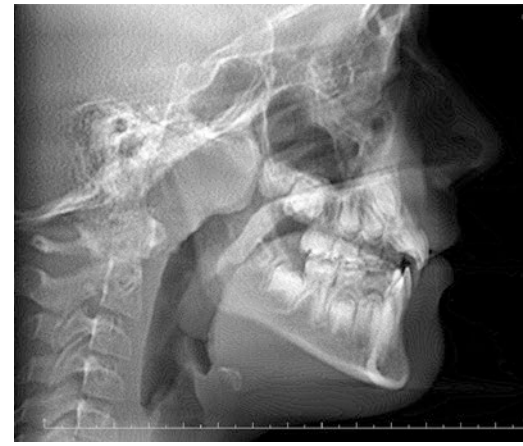

Fig. 1.1 Large adenoidal pad filling the nasopharynx in this patient

Other radiographs and studies such as swallow studies, esophagram, upper GI CT and CT chest angiography, or MRI of the head or chest may also be of interest and are discussed by the entire team to determine utility and need.

Rigid airway endoscopy with its superior optics should also be the forte of the otolaryngologist. It offers superior static visualization, sizing, and intervention. Figure 1.2 is an example of severe laryngomalacia seen during direct laryngoscopy. The otolaryngologist member of the aerodigestive program should be strongly versed and capable with endoscopy of the upper aerodigestive tract.

As the surgical representative of the four core members, the otolaryngologist can also offer surgical or procedural correction for certain disorders. Boesch et al. [7] polled aerodigestive programs regarding the types of procedures requiring proficiency by otolaryngologists in aerodigestive programs for open and endoscopic airway reconstruction. Proficiency should be maintained in the following interventional categories: (1) open or endoscopic procedures that directly increase the diameter of the cartilaginous skeleton of the airway, (2) endoscopic treatment of airway obstruction, (3) surgical procedures to treat aspiration, (4) surgical procedures to improve voice, (5) tracheostomy, and (6) foreign body removal.

One of the most rewarding goals encountered in many aerodigestive patients is the relief

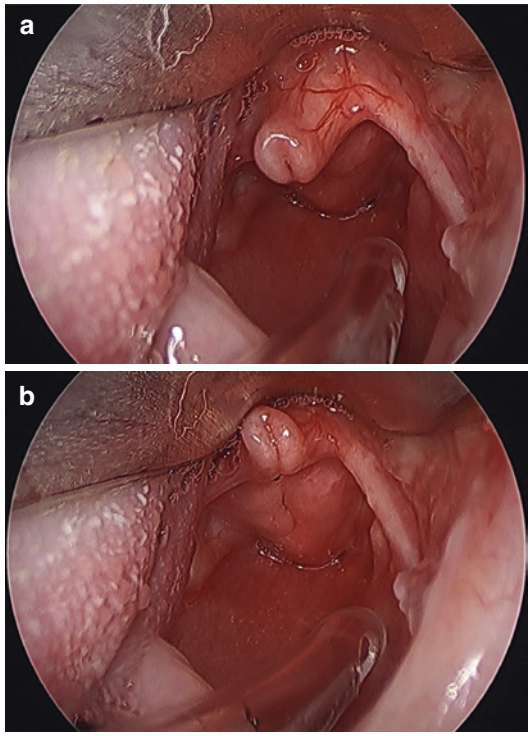

Fig. 1.2 (**a**, **b**) Operative direct laryngoscopy in a 6-week-old child with signs and symptoms of progressive noisy breathing and retractions since birth. Her symptoms were worse during activity of crying and feeding. She has a diagnosis of laryngomalacia. The infantile larynx is curled and obstructed during inspiration with the supraglottic structures

of tracheotomy dependence. Some patients will require airway reconstruction for the tracheotomy to be successfully removed. As stated previously, successful airway reconstruction requires a multidisciplinary approach [30] with sufficient preoperative evaluation of all airway lesions and non-airway diagnoses, appropriate patient selection, appropriate reconstructive technique, staging and timing, and effective patient optimization [8]. An understanding of the surgical approached for airway reconstruction in the context of static and dynamic lesions is essential and must include an appreciation of the interrelatedness with other comorbidities [8].

Working together, the otolaryngologist can bring expertise in diagnostic airway evaluation and surgical airway interventions to maximize patient outcomes through the aerodigestive team.

Role of the Pulmonologist

The role of the pulmonologist in the aerodigestive team is to provide complementary anatomic airway evaluation. Flexible bronchoscopy allows for a better dynamic assessment of the trachea and bronchus. It also allows for a more distal airway assessment and affords a superior bronchoalveolar lavage to help with culture and identifying any inflammatory markers. The pulmonologist is also integral in diagnosing and optimizing respiratory comorbidities prior to airway reconstruction and assisting in postoperative management. Pulmonologists may evaluate for and manage lung injury due to aspiration, active infectious or inflammatory lung disease, impaired airway clearance, interstitial lung disease, asthma, sleep apnea, dynamic airway lesions, and respiratory muscle weakness [8]. The pulmonologist may also make recommendations regarding radiographs, CT chest and CT angiography, as well as pulmonary function tests that may aid in the evaluation.

A list of recommended procedures that a pediatric pulmonologist should be able to provide at an aerodigestive center has been established by Boesch et al. using the Delphi method among a number of aerodigestive programs [7]. This includes bronchoscopy with bronchoalveolar lavage, balloon dilation, sleep state bronchoscopy, biopsy, foreign body removal, and identification of tracheoesophageal fistula.

Role of the Gastroenterologist

The role of the gastroenterologist is to evaluate growth and nutrition and gastrointestinal barriers to safe and adequate feeding and to diagnose and manage esophageal and other gastrointestinal disorders that may present as aerodigestive symptoms (Table 1.1) [8]. They are also essential in managing gastrointestinal disorders that may complicate airway reconstruction. The array of possible diagnoses includes laryngopharyngeal or gastroesophageal reflux, acidic or eosinophilic esophagitis, reflux aspiration, esophageal dysmotility, esophageal stricture, rumination, gastritis, and malabsorption [8].

The gastroenterologist will rely on history and physical findings, upper GI series, swallow studies, EGD (esophagogastroduodenoscopy) with biopsy, pH probe monitoring, impedance manometry, and specific blood work. Procedures which can be performed by the pediatric gastroenterologist on the aerodigestive team include esophagogastroduodenoscopy with biopsy, dilation, cautery, and placement of percutaneous endoscopic gastrostomy (PEG) or gastrojejunostomy (GJ) tubes [7].

Role of the Parent/Caregiver

When working with children, we cannot exclude the role of the parent(s) or caregiver(s) in evaluation and management. DeCivita and Dobkin [31] describe the therapeutic triad that exists among the medical team, the child, and the parent/caregiver. All have shared decision-making, and the impact of the disorder as well as the treatment burden needs to be considered. In interviewing parents of children with dysphonia, Connor and colleagues found that parents reported concerns with social and emotional issues related to their voice, as well as concerns with being understood and fitting in and concerns about comments from others [32]. Parents of children undergoing voice therapy for dysphonia have discussed concerns about social and emotional outcomes, academic and career success for their children, and the impact that their voice has on peer relationships [33]. At the same time, attendance and adherence to voice therapy depend on the parent, as they are usually the ones scheduling and bringing the child to therapy, as well as helping with and monitoring home practice [33]. With infants, very young children, or older children with cognitive or communication impairments, the parent is the only one able to provide a history and description of problems. Parents bear the financial burden, responsibility for transportation, and responsibility for carrying over medical team recommendations at home. They are responsible for making challenging decisions about their child's care, with varying levels of medical knowledge and experience.

Feeding and swallowing are particularly emotionally loaded areas for families. Parents of children born with cleft lip and palate have reported feeling that their ability to feed their baby is linked to their competency as a parent [34]. When feeding modifications are recommended, the parents are primarily responsible for thickening liquids, providing positioning and pacing, and ensuring adequate oral intake. Parents have described feeding difficulties with children as a journey lasting from birth and discussed the impact it has on daily life, from the ability to leave the house to schedule activities, and the need to plan ahead extensively [35]. Parents of children with failure to thrive (FTT) [36] described not feeling heard by medical professionals, conversely feeling nurtured by others; feeling comparisons; being afraid; doing what needed to be done.

When partnering with parents, the medical team needs to take into account their values, their understanding of the medical issues facing their child, and their resources and abilities to cope. In many cases, it is easy for a medical team to see themselves as providing a service for the child, rather than partnering with the child and family, especially when treating conditions that require multiple interventions, whether they are medical, surgical, or behavioral.

Conclusion

The multidisciplinary approach to children with aerodigestive disorders is rewarding. Through this approach, efficient, cost-effective, patient-centered, family-focused, and consistent care can be delivered. The number of new diagnoses and the speed to diagnosis increase. The overall cost to diagnosis decreases, and the efficiency in the OR increases, freeing up more OR time for other unique procedures [6]. These complex and medically fragile patients deserve our best and concerted care to help them reach their maximal potential.

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Operative Evaluation of the Upper Aerodigestive Tract

2

Matthew R. Hoffman and J. Scott McMurray

Overview

Critical components of patient assessment always include history, physical exam, and in-office visualization of the functioning larynx. Instrumented evaluation of swallow function and imaging exams may also be warranted, depending on the clinical scenario. In most children with a significant aerodigestive complaint which is not readily identified on these assessments, the next step is evaluation in the operating room for further physical examination. Operative direct laryngoscopy, bronchoscopy, and esophagoscopy allow for close examination of the upper aerodigestive tract with the opportunity for intervention and remain an invaluable aspect of pediatric patient evaluation. This chapter reviews the indications, equipment, technique, and approach to intraoperative upper aerodigestive tract assessment.

Indications

Direct laryngoscopy and bronchoscopy can be an important aspect of the assessment of any breathing, swallowing, or voicing problem. While awake office flexible laryngoscopy gives a better functional assessment and may be adequate in some cases, direct laryngoscopy and bronchoscopy can provide superior information and allow for direct palpation of the laryngeal structures. In a study of 523 children with history of aspiration, flexible laryngoscopy was able to identify 91 anatomic abnormalities, while direct laryngoscopy and bronchoscopy identified an additional 215 abnormalities [1]. Direct laryngoscopy and bronchoscopy can also be indicated as part of the comprehensive assessment formed by the multidisciplinary aerodigestive team, in conjunction with esophagogastroduodenoscopy and flexible bronchoscopy with bronchoalveolar lavage.

Equipment

Necessary equipment includes devices for direct visualization, magnification and video recording, as well as palpation.

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Laryngoscopes

There are many laryngoscopes available to expose the larynx and place a patient into suspension for detailed operative exam or intervention (Fig. 2.1). Examples include the Philips, Parsons, Lindholm, and Zeitels. The Philips blade attaches to a standard lighted handle and is useful for exposing the larynx as part of a direct laryngoscopy or bronchoscopy. The Philips laryngoscope blade is a straight blade with a short distal curve. The light source is also located distally near the slight curve. The Parsons blade has a port to attach a light cable and is a single-piece unit. It can be connected to a Lewy arm to place the patient into suspension. The Lindholm is often used for operative intervention. It has ports for light and suction and can be connected to a Lewy arm to place the patient into suspension. There is a pediatric version of the Zeitels universal modular glottiscope that can be used to achieve a view of the glottis for phonosurgical procedures.

Suspension Arms

Once the larynx is exposed, patient can be placed into suspension for further examination or operative intervention. The Parsons and Lindholm can be connected to a Lewy arm which is placed on a Mayo stand or Mustard stand for suspension.

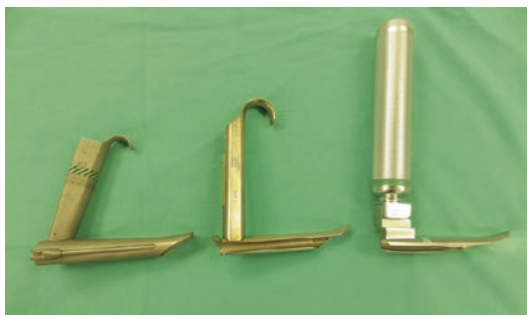

Fig. 2.1 Examples of pediatric laryngoscopes, including the Lindholm (left), Parsons (middle), and Philips (right)

Telescopes and Bronchoscopes

0-, 30-, and 70-degree telescopes should be available for close evaluation of the larynx. Examination can start with the 0-degree telescope to assess the supraglottis, superior glottis, subglottis, trachea, and proximal bronchi. The 30- and 70-degree telescopes can aid in close assessment of the anterior commissure, ventricles, and infraglottic surfaces of the true vocal folds. The telescope should be connected to a light cord for illumination and video camera for recording and projection of the image on a monitor.

Bronchoscopes have four ports: telescope, prism, ventilatory circuit, and suction (Fig. 2.2). The prism directs light through the bronchoscope but is now not typically required as the attachable light source on the telescope provides superior illumination. The prism is still placed to prevent air escape through the port.

Appropriate size of bronchoscope to use based on patient age is presented in Table 2.1.

Microlaryngeal Instruments

Microlaryngeal instruments which can be helpful during general assessment include the vocal cord retractor/posterior glottic spreader and the right-angle probe. The vocal cord retractor can be placed in an inverted fashion to lateralize the false vocal folds while still allowing the surgeon

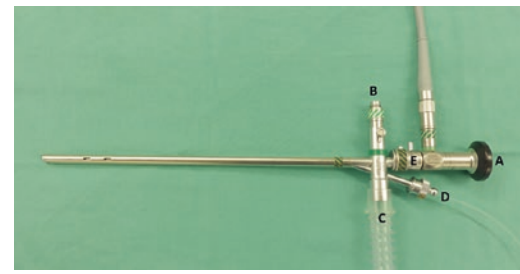

Fig. 2.2 Ventilating bronchoscope with attachments including telescope (**a**), light prism (**b**), ventilatory circuit attachment (**c**), and suction (**d**). A bridge (**e**) connects the telescope to the bronchoscope

Table 2.1 Patient age with corresponding estimated diameter of the cricoid and trachea as well as the corresponding appropriate size bronchoscope to use

	Age	<1 month	1–6 months	6–18 months	18–36 months	3–6 years	6–9 years	9–12 years	>12 years
Cricoid diameter	ID	3.6–4.8	4.8–5.8	5.8–6.5	6.5–7.4	7.4–8.2	8.2–9.0	9.0–10.7	10.7+
Trachea diameter	ID	5	5–6	6–7	7–8	8–9	9–10	10–13	13+
Bronchoscope	Size	2.5	3.0	3.5	3.7–4.0	5.0	5.0–6.0	6.0	6.0
	ID	3.5	4.3	5.0	5.7–6.0	7.1	7.1–7.5	7.5	7.5+
	OD	4.0	5.0	5.7	6.4–6.7	7.8	7.8–8.2	8.2	8.2+

Numbers represent size in millimeters
ID inner diameter, *OD* outer diameter

access to the posterior laryngeal structures and the interarytenoid area. The retractor is then suspended via rubber bands onto the suspension apparatus to provide hands-free exposure. Care must be taken during placement to avoid injury to the true vocal folds. The right-angle probe is helpful in multiple ways. First, it can be used to palpate the interarytenoid space to evaluate for a laryngeal cleft. Second, it can be used to palpate the true vocal folds in a systematic fashion to evaluate for scar, sulcus vocalis, or other glottic abnormality such as a submucosal cyst. During palpation, the probe is placed perpendicular to the vocal fold and passed over its surface in an inferior to superior fashion. This motion is performed over the length of the vocal fold and then repeated on the other side. In this way, subtle changes in vocal fold stiffness can be appreciated that might otherwise be missed on visualization alone. This is especially important when a submucosal cyst is suspected.

Instrument Table Setup

Figure 2.3 demonstrates a typical setup in preparation for direct laryngoscopy and bronchoscopy. Equipment includes a quiver for holding laryngeal suctions, Lewy suspension arm, pediatric Lindholm laryngoscope for use in suspension, Phillips 1 laryngoscope for initial exposure and exam, ventilating bronchoscope, additional rigid telescope, defog pad, topical lidocaine, mouthguard, petri dish for holding pledgets, 0.5″ × 0.5″ pledgets, dry gauze, saline, laryngeal suctions of varying size, uncuffed endotracheal tubes of varying size based on patient's anticipated subglottic diameter, and right-angle probe. Additional equipment which may be needed but is not pictured includes an attachable video camera for the telescope, vocal cord retractor, and angled rigid endoscopes for evaluation of the anterior commissure and infraglottic surfaces of the true vocal folds.

Approach

Preoperative Assessment

Preoperative evaluation should focus on anticipated ease of exposure, development of anesthetic plan in conjunction with the anesthesiologist, and ensuring all necessary equipment is available. Factors associated with difficult laryngeal exposure include restricted head extension, small oral cavity, macroglossia, craniofacial dysmorphism, and reduced thyromental distance [2, 3].

Patient Positioning

The head of the bed is rotated 90° away from the anesthesia circuit. The anesthesia machine is typically to the patient's left. This allows for the laryngeal equipment to be set up to the right and passed to the field from the right. In this way, the otolaryngologist has clear access to

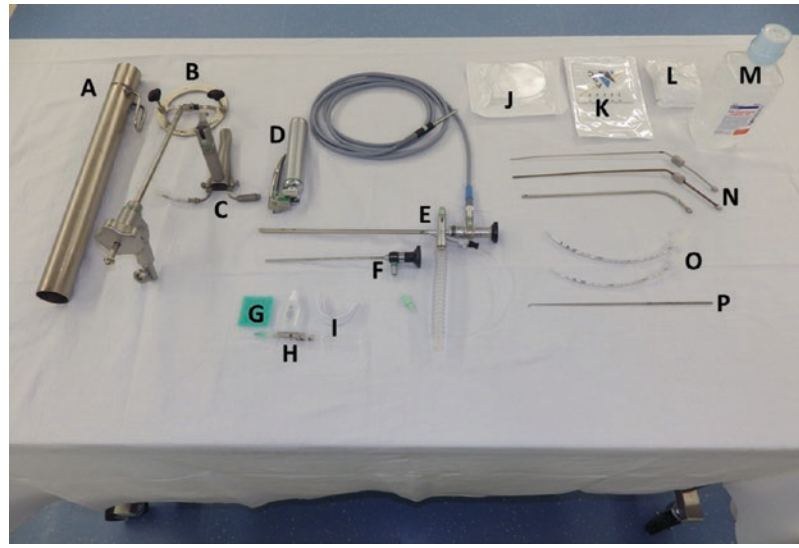

Fig. 2.3 Table setup for direct laryngoscopy and bronchoscopy for purpose of upper airway exam. Equipment shown includes quiver (**a**), suspension arm (**b**), Lindholm laryngoscope (**c**), Phillips laryngoscope (**d**), bronchoscope (**e**), additional telescope (**f**), defog (**g**), topical lidocaine (**h**), mouthguard (**i**), petri dish (**j**), 0.5″ × 0.5″ pledgets (**k**), dry gauze (**l**), saline (**m**), laryngeal suctions (**n**), uncuffed endotracheal tubes (**o**), and right-angle probe (**p**)

the airway, and the anesthesiologist can monitor the patient closely for level of anesthetic and ensure appropriate ventilation. The patient is positioned supine with the scalp vertex at the edge of the bed. Optimal positioning for direct laryngoscopy includes flexion at the neck and extension at the atlanto-occipital joint, the sniffing position. If necessary, the head of the bed can be flexed slightly to aid in flexion at the neck and extension of the head. In some patients (e.g., those with Down syndrome) in whom atlanto-occipital instability is a potential risk, the neck should remain neutral if possible. Atlantoaxial flexion and rotation have been shown to produce the greatest changes in the atlantodens interval (ADI). Preoperative neck films in patients with Down syndrome are controversial, and no definitive recommendations have been made [4]. Trying to maintain a neutral position if possible and early monitoring postoperatively for weakness are recommended. A mouthguard is placed to protect chipping the maxillary dentition, but care must be taken not to apply too much pressure on the dentition to prevent fracture or extraction. If the patient is edentulous, moistened folded gauze is helpful to prevent maxillary gum injury.

Procedure

Once the head of the bed is turned, the otolaryngologist assumes airway management and maintains bag mask ventilation. This can be facilitated by the use of an oral airway. Anesthesia for the endoscopy is achieved by inhalational anesthesia, TIVA (total intravenous anesthesia), or a combination of the two. The anesthetic technique should be discussed prior to the induction and be modified based on physician preference and comfort and the patient's needs. Spontaneous ventilation is preferred if possible, for safety and to allow for a dynamic assessment of the airway as well. Once the patient is in a stable plane of anesthesia and able to tolerate direct laryngoscopy, the oral airway is removed, and the maxillary alveolus is protected. A laryngoscope of the surgeon's choice is used in the right lingual gutter to sweep the tongue to the left and directly expose the larynx. Topical lidocaine is atomized onto the larynx and trachea. Considering the maximal allowed lidocaine dose is important, particularly in infants. For topical plain lidocaine, the maximal allowable dose is 5 mg/kg, and there are 10 mg in each 1 cc of 1% lidocaine (thus, 20 mg in each 1 cc of 2% lidocaine, and so on). After the lidocaine

is applied, the laryngoscope is removed, and bag mask ventilation resumed to give time for the anesthetic to take effect. The larynx is then re-exposed. A telescope with or without a ventilating bronchoscope is then passed transorally to visualize the upper airway. An image is taken to demonstrate the exposure obtained and ease of future intubation (Fig. 2.4). Clear, close-up images of the supraglottis and glottis are obtained. The 30- and 70-degree angled endoscopes can be used to visualize the ventricles and infraglottic surfaces of the true vocal folds. The telescope or bronchoscope is then passed carefully through the glottis, either through the posterior glottis between the vocal processes or aiming at one vocal process and then rotating the telescope/bronchoscope medially. Images are then recorded of the subglottis, mid-trachea, carina, and each proximal bronchus. As the telescope/bronchoscope is withdrawn, attention is paid to movement of the trachea during respiration to evaluate for tracheomalacia, to the posterior tracheal wall to evaluate for tracheoesophageal fistula, and to the anterior tracheal wall to evaluate for vascular compression. Once the telescope/bronchoscope is withdrawn, the subglottis is sized with serial intubations using progressively larger uncuffed endotracheal tubes. Appropriate endotracheal tube size based on age is calculated according to the following formula: (age in years +16)/4. After sizing has been performed, additional examination can be performed as indicated, which can include palpation of the arytenoid cartilages to evaluate for cricoarytenoid joint fixation and palpation of the interarytenoid space to evaluate for laryngeal cleft.

During the procedure, oxygenation and ventilation can be accomplished via several methods. The insufflation technique can be used to allow for oxygenation and delivery of an inhalational

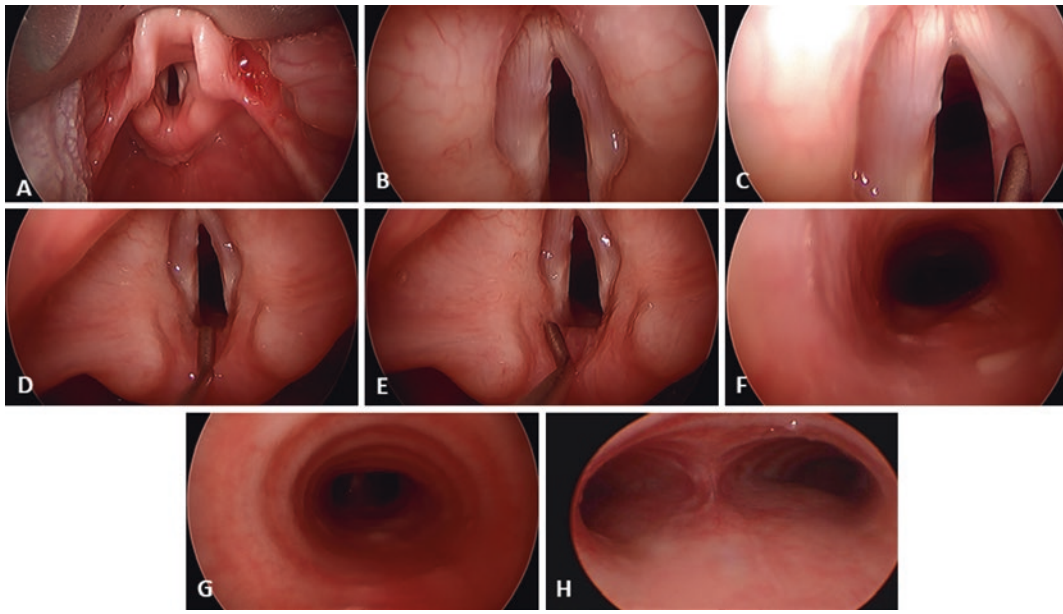

Fig. 2.4 Series of images recorded during standard direct laryngoscopy and bronchoscopy, including intubating view (**a**), close-up of glottis (**b**), use of right-angle probe to visualize infraglottic surface of right true vocal fold (**c**), palpation of interarytenoid region to rule out cleft (**d**), palpation of vocal process to assess cricoarytenoid joint mobility (**e**), and views of subglottis (**f**), mid-trachea (**g**), and carina (**h**)

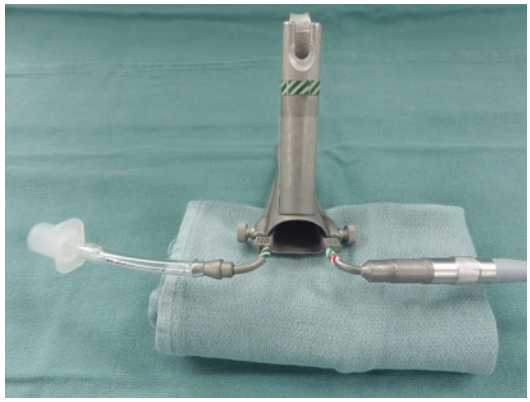

Fig. 2.5 Insufflation technique can be performed with endotracheal tube placed in oral cavity against the oral commissure or, as shown here, with cut endotracheal tube connected to the suction port on a laryngoscope

anesthetic. An endotracheal tube is placed in the oral cavity or, alternatively, a 5.5 endotracheal tube can be attached to the suction port on the laryngoscope (Fig. 2.5). During bronchoscopy, the ventilating port on the bronchoscope can be used (Fig. 2.2). If any desaturations occur, equipment is removed and bag mask ventilation performed.

Considerations for the Difficult Exposure

In the anesthesia literature, difficult exposure is defined as Cormack-Lehane grade III or IV exposure. This equates to visualization of only the arytenoids (grade III) or posterior pharyngeal wall (grade IV). Intubation can be performed without adjuncts (e.g., Eschmann stylet) even with a grade II view (seeing posterior aspect of true vocal folds but not seeing the anterior commissure). For a complete evaluation of the larynx, it is important to see the entire glottis clearly including the anterior commissure. Adjuncts that can aid in exposure include increasing the degree of neck flexion and head extension, utilizing suspension, and applying counter pressure over the cricoid manually or with silk tape attached to the bed. A folded gauze is placed over the cricoid cartilage, and silk tape is wrapped around the entire bed and over the gauze to apply firm, constant

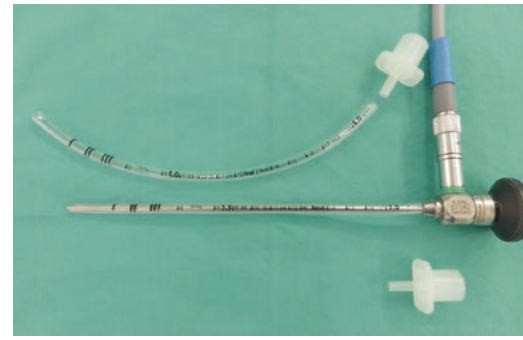

Fig. 2.6 Placing an endotracheal tube over a rigid telescope can aid with intubation in the setting of a difficult exposure

pressure over the larynx and facilitate visualization of the anterior commissure. In cases where the primary goal is intubation or general airway visualization rather than close examination of the glottis, the telescope can be placed at an angle and used to visualize the glottis, subglottis, and trachea indirectly rather than via direct line of sight. In a similar fashion, an endotracheal tube can be placed over the telescope (Fig. 2.6). With the larynx exposed as best as able, the telescope is passed transorally through the glottis and into the trachea. It is then withdrawn as the endotracheal tube is stabilized and then connected to the anesthesia circuit.

Emerging Concepts and Techniques

Two recently proposed methods of intraoperative evaluation may play a future role in the operative evaluation of pediatric dysphonia. Optical coherence tomography (OCT) is a method of performing high-resolution cross-sectional imaging that is analogous to ultrasound, though a light is used instead of sound and thus superior resolution is provided [5]. Light is directed onto a structure of interest (e.g., true vocal fold), and differential backreflection and backscatter result in creation of an image [6]. This approach was recently applied to the operative assessment of pediatric true vocal folds to distinguish among nodules and cysts as well as assess depth of papilloma and sulcus vocalis [7]. Further refinement may offer a

method of intraoperative lesion assessment without the need for vocal fold exploration as well as improved planning prior to lesion removal.

Hoffman et al. recently described the use of microendoscopes to perform endoscopy of Reinke's space via a small neck incision with minithyrotomy [8]. This approach was then used in an animal study to deliver a hyaluronic acid-based hydrogel to porcine larynges with simulated mucosal stripping and mid-true vocal fold biopsy injuries, with consequent improvement in rheologic properties [9]. Microendoscopy of Reinke's space offers an exciting new avenue for examination and treatment of disorders of the lamina propria, including vocal fold scar. This approach does currently require an external neck incision but affords visualization of and potential delivery of therapeutics to Reinke's space without further mucosal violation.

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Anesthetic Considerations

3

Bridget Muldowney

As invasive procedures become part of the workup for pediatric voice and swallowing disorders, an anesthetic to facilitate these procedures is often necessary. Here we will review preoperative assessment, intraoperative management, and postoperative management.

Preoperative Assessment

Preoperative assessment begins with a comprehensive history and physical exam best performed by the physician who has the most continuity with the patient. For all children, but especially for neonates, infants, and toddlers, birth history is important. Prematurity can lead to a number of complications, reactive airway disease being the most pertinent for anesthetic management. Prematurity also has implications for the timing of surgery and the need for postoperative observation. Premature infants are more prone to apnea and bradycardia spells and so require a 24-h observation stay with cardiorespiratory monitoring after a general anesthetic until they are 52–60 weeks post-conceptual age, dependent on institutional practice [1, 2].

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Past medical history should include craniofacial abnormalities commonly associated with difficult airway management. The most common examples include Pierre Robin sequence, craniofacial dysostosis, mandibulofacial dysostosis/Treacher Collins syndrome, and hemifacial microsomia, but many others exist [3].

Family history should include a history of adverse anesthetic reactions. Although rare, malignant hyperthermia (MH) is a life-threatening condition associated with exposure to volatile anesthetics or the depolarizing neuromuscular blocking drug succinylcholine. Any positive family history of MH should be clearly documented and conveyed to the anesthesia team as it would necessitate an alteration of typical anesthetic management. Resources can be found at www.mhaus.org.

One of the most important acute illnesses that is pertinent to anesthetic care of any child but specifically in children presenting for airway procedures is an upper respiratory infection (URI). URIs are ubiquitous in toddlers and young children yet have important implications for timing of the anesthetic and possible complications. Airway surgery puts patients with a URI at a higher risk of adverse events. Multiple studies have shown that children with an active or recent (within 2–4 weeks) URI are more prone to bronchospasm, laryngospasm, breath holding, oxygen desaturation <90%, and overall adverse respiratory events. It would be important to note if a

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patient has the following independent risk factors for these events: history of prematurity, personal or family history of reactive airway disease or eczema, and second-hand smoke exposure [4, 5]. When a patient has an active or recent URI, the decision to proceed versus cancel and reschedule must be a collaborative one between the surgical and anesthesia team. For some children, it can be hard to find a time when they are completely free of URI symptoms to safely proceed with operative evaluation.

Intraoperative Management

The workup for voice and swallowing disorders is now routinely conducted by aerodigestive programs performing a triple endoscopy, also known as the "triple scope": laryngoscopy and rigid bronchoscopy, flexible bronchoscopy with bronchoalveolar lavage, and esophagogastroduodenoscopy. This involves coordination among the otolaryngology team, pediatric pulmonary team, and pediatric gastroenterology team.

Laryngoscopy and rigid bronchoscopy can be one of the most challenging anesthetic cases even for an experienced pediatric anesthesiologist. One must balance maintaining spontaneous respiration while also providing a deep plane of anesthesia to prevent movement, coughing, or laryngospasm. This becomes even more challenging with younger patients, as maintaining an adequate depth of anesthesia often comes with significant hypotension. During the procedure itself, end-tidal carbon dioxide monitoring, a standard American Society of Anesthesiology (ASA) monitor, is not reliable, and the anesthesiologist must use auscultation of breath sounds and visual inspection of chest rise as confirmation of ventilation.

In most young children, anesthesia is induced with a volatile anesthetic provided by face mask. Once an adequate depth of anesthesia is achieved, a peripheral intravenous catheter is placed while maintaining spontaneous respirations. Our institutional practice is to start with rigid bronchoscopy. There are two different anesthetic techniques to facilitate rigid bronchoscopy, and choice is determined by anesthesiologist and/or surgeon preference. There is limited evidence to suggest one technique is superior to another [6].

The first option is a total intravenous anesthetic. This is often accomplished with a propofol infusion with or without additional opiate, most commonly fentanyl bolus or remifentanil infusion. It takes time for the propofol to reach a steady state, but the patient often has residual volatile anesthetic from the mask induction to cover this period. The advantage of this technique is providing a measurable amount of anesthetic as well as limiting volatile anesthetic pollution to the operating room and providers.

The second choice is to continue providing anesthesia with inhaled volatile anesthetic. This can also be augmented with additional opiate analgesics. The advantage of this is simplicity, as no additional infusions need to be started. One may be less likely to induce apnea with this technique as well. The main disadvantage is the difficulty in measuring the amount of volatile anesthetic delivered, as delivery is occurring via a side port of a rigid bronchoscope or via an endotracheal tube positioned in the oral cavity. There is also significant amount of waste gas pollution to the operating room as high flows of oxygen are necessary to carry the volatile anesthetic to the patient. According to the Centers for Disease Control (CDC), exposure to high concentrations of waste anesthetic gases, even for a short time, may cause headache, irritability, fatigue, nausea, drowsiness, difficulties with judgment and coordination, and liver and kidney disease [7].

The anesthesia team will often give a dose of steroid (dexamethasone 0.5–1 mg/kg (up to 10 mg)) to prevent airway swelling. They will also provide the proceduralists with topical lidocaine to apply on the true vocal folds. The total dose is split between the otolaryngology team and the pulmonary team who will want to anesthetize the carina as well.

After completion of rigid bronchoscopy, a classic laryngeal mask airway (LMA) is placed to facilitate flexible bronchoscopy by the pulmonary team. Use of an LMA instead of an endotracheal

tube allows for a larger flexible bronchoscope to be used. During this time, the anesthetic may proceed with IV anesthesia (propofol), volatile anesthetic (sevoflurane), or a combination of the two. One of the biggest challenges is again maintaining a depth of anesthesia to prevent coughing and movement while attempting to maintain spontaneous respirations. After the pulmonologist preforms bronchoalveolar lavage (BAL), the patient may experience transient desaturation due to shunting in the lung segment that was lavaged. The degree of desaturation is often dependent on the amount of fluid instilled and removed by suction.

The patient's anesthetic management can proceed in a number of ways for the final step of the procedure, the upper endoscopy performed by the gastroenterologist. If the patient is stable without desaturation after the BAL, the LMA can be removed, and an IV-based anesthetic can proceed with spontaneous ventilation and oxygen delivery via nasal cannula. This technique prevents further airway manipulation and irritation. Certain patients, particularly younger patients, struggle to maintain unobstructed spontaneous respiration with an endoscope in the esophagus. In this case, there are two options. Some gastroenterologists are willing to work around the LMA already in place, although they often prefer to work around a flexible LMA. If this is not an option, or if the patient's pulmonary status is tenuous after the flexible bronchoscopy, the patient will require endotracheal intubation. This is likely the safest way to proceed, but also the most invasive, and the patient can still struggle with coughing, desaturation, and laryngospasm upon emergence and extubation.

Although this is the typical course, there is currently no national standard for anesthetic management for these procedures, and within a given institution, there is often marked variability between providers. The American Association of Pediatrics (AAP) consensus statement on the structure and function of these programs highlights the benefit of fewer exposures to anesthesia when these services are performed together, yet it makes no mention of anesthetic techniques [8]. It is beneficial for an institution to have an anesthetic guide for management as these patients are often complex and coordination among so many providers on one case can be challenging. At our institution, a clinical guide helped standardize care and management for these procedures.

One final procedure to mention is the drug-induced sleep endoscopy (DISE). This procedure is often requested when there is concern for sleep-disordered breathing or obstructive sleep apnea. The goal for the otolaryngology team is to visualize the upper airway in a state that mimics natural sleep. From an anesthesia standpoint, this can be very challenging. The vast majority of mediations used in anesthesia cause transient respiratory depression and decreased pharyngeal muscle tone. Some otolaryngologists are willing to examine the airway with a small flexible fiberoptic endoscope inserted through the end-tidal sampling line port of the circuit elbow during mask ventilation. Although the anesthesia team will lose end-tidal monitoring during this time, the anesthetic can continue with face mask delivery of volatile anesthetic. Other otolaryngologists prefer to use agents that more closely mimic natural sleep. Our institutional practice limits oral premedications and proceeds with inhalation induction with volatile anesthetic. A dexmedetomidine (selective alpha-2 agonist) bolus is then given over the course of 10 min, while the volatile anesthetic is washed out through spontaneous respiration. We find a small bolus of ketamine (0.5–1 mg/kg) given just before nasal endoscopy helps the patient tolerate the procedure. Both ketamine and dexmedetomidine have the least effects on pharyngeal tone and respiratory drive of the commonly used anesthetic agents. A recently published review on DISE pointed out a lack of agreement for optimal anesthetic management/agents. They described the use of oral premedication, intranasal dexmedetomidine, nitrous oxide, fentanyl, ketamine, and topical anesthesia [9]. Again, standardization of technique within an institution and program will likely improve procedural and diagnostic outcomes.

Postoperative Management

Most patients presenting for triple endoscopy will be candidates for outpatient surgery. During phase 1 recovery, it is important to ensure a patent natural airway and adequate oxygen saturation without supplemental oxygen, especially if the patient underwent BAL. If there is concern for stridor and/or airway edema, racemic epinephrine may be used as a treatment, but the patient will need to stay at least 2 h after administration to ensure they do not have rebound swelling. As none of the endoscopy procedures are particularly painful, postoperative analgesia with acetaminophen is often sufficient. As with all ambulatory surgery patients, discharge readiness should be assessed with a validated tool such as the Pediatric Post Anesthesia Discharge Scoring System (Ped-PADSS). This score measures adequate vital signs, ambulation, nausea and/or vomiting, pain, and surgical bleeding [10]. In rare cases, if oxygen saturation is not adequate, if airway obstruction is present, or patients do not meet the ambulatory discharge criteria above, an inpatient observation may be necessary.

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4 Perioperative Considerations After Pediatric Laryngeal Surgery

Maia N. Braden, Matthew R. Hoffman, and J. Scott McMurray

Overview

Surgery on the larynx is extremely delicate and requires careful pre- and postoperative management to ensure optimal results. While there is much discussion of the "essential ingredients," perioperative management generally falls into the categories of behavioral and medical. Behavioral considerations include how much and how the voice should be used after surgery, and medical management may include steroids and anti-reflux treatment.

When considering surgical intervention on the larynx, pre- and postoperative management are essential in facilitating the best possible outcomes. These recommendations are largely intended to limit irritation and impact forces on healing tissues and promote rapid and effective tissue healing after the surgery. Surgery on the pediatric larynx is not undertaken lightly, and children can have more difficulty than adults in adhering to postoperative recommendations. Especially when surgery is done for reasons of preserving or improving voice quality, consideration of the child's ability and willingness to participate in preoperative therapy and postoperative voice rest and therapy is imperative. While recommendations vary based on individual surgeon and speech pathologist, specific surgery, and individual considerations for the patient, in general, these recommendations encompass preoperative voice therapy and counseling, postoperative voice rest, postoperative voice therapy, and medical management, which may include anti-reflux medication and/or steroids.

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The Role of the Speech-Language Pathologist

In cases where surgical management of the voice or airway disorder is warranted, the speech-language pathologist plays an important role in preoperative counseling and pre- and postoperative voice therapy and voice care. The speech-language pathologist's role typically revolves around preoperative voice therapy, counseling on postoperative voice rest, guiding return to voice use, and rehabilitation of voice after surgery through postoperative voice therapy. As there are no available studies on voice rest or postoperative voice use in children, clinicians must look to the adult literature, the basic science literature, and pediatric wound healing literature for guidance.

Voice Rest

Voice rest is commonly recommended after laryngeal surgery, but there is no clear consensus on the duration, type, or importance of voice rest in postoperative management of voice. The evidence that exists is limited to studies of adults, and a review of the literature revealed no studies of voice rest in children. Recommendations range from no rest at all to multiple weeks of total voice rest, with most otolaryngologists falling somewhere in the middle [1–3]. In a survey, 84/85 respondents recommended some form of voice rest following phonomicrosurgery with up to 14 days of complete voice rest or 35 days of relative voice rest, 7 days the most common recommendation for subepithelial lesions, and 1–4 days recommended for epithelial lesions [1]. Coombs et al. found that most otolaryngologists recommended 1–2 days of postoperative voice rest, but some recommended more than 7 days [3]. Behrman and Sulica found that 51.4% of otolaryngologists preferred complete voice rest, 62.3% preferred relative voice rest, and 15% preferred no voice rest at all [2]. Recommended durations of voice rest ranged from 0 to 14 days of complete voice rest, and 0–21 days of relative voice rest, with the most common duration being 7 days [2]. While otolaryngologists have clear preferences in their recommendations for voice rest, these are often based on experience or expert opinion, but not on the available research.

Much of the standard practice of voice rest is based on a study done in a canine model, in which the vocal fold mucosa was excised bilaterally, and voice rest was simulated by resection of the recurrent laryngeal nerve; based on this study, 2 weeks of voice rest was recommended [4]. A small number of studies have explicitly examined the effects of voice rest in patients after surgery. In a 2015 study, participants with 10 days of voice rest demonstrated longer maximum phonation times than those undergoing 5 days of voice rest, but there were no other significant findings [5]. Conversely, Kaneko et al. found that 3 days of voice rest followed by voice therapy showed better results than 7 days of voice rest in adults undergoing phonomicrosurgery for leukoplakia, carcinoma in situ, Reinke's edema, polyp, or cyst. Specifically, patients demonstrated improved vibratory function as assessed by normalized mucosal wave amplitude (NWMA) on stroboscopy at 6 months postoperatively. Perceptual voice evaluation indicated more perceptually normal voice in the 3-day group than the 7-day group at 1 and 3 months, and acoustic findings and quality of life measures were better in the 3-day than the 7-day group at 1 month after surgery [6]. While optimal duration of voice rest cannot be determined based on these studies alone, it does suggest that earlier controlled phonation with voice therapy may be beneficial in functional outcomes.

The basic science literature on this can help guide clinicians in developing guidelines for voice rest and therapy but is likewise inconclusive on the role of both rest and vocalization on healing. Vocal fold healing and scar have been primarily examined in animal models [7–11]. For an excellent overview of wound healing for the clinician, see Thibeault and Gray or Branski et al. [12, 13] In brief, wound healing consists of three stages: inflammation (days 1–3), proliferation which is made up of angiogenesis and epithelization (days 3–30), and maturation (1 year or more). In the vocal folds, hemostasis is complete in 24 h, and the inflammation stage lasts for

4–7 days, and epithelization is complete in 7 days [12]. Research in pediatric wound healing indicates that it follows the same trajectory as healing in adults but at an accelerated rate; in children, fibroblasts are present in greater numbers, collagen and elastin are produced more rapidly, and granulation tissue forms more quickly [14]. The human infant and pediatric vocal fold differs from the adult in terms of layer structure and distribution of collagen and hyaluronic acid [15, 16]. The infant lamina propria consists of a monolayer, and the layer structure differentiation is not complete until adolescence [16, 17]. Because of these differences, we cannot make assumptions that healing and scar in pediatric vocal folds will be identical to adults.

Vocal fold lamina propria is a form of connective tissue, although unique in composition and features. As such, some parallels can be drawn with orthopedic rehabilitation. The role of rest versus controlled mobilization has been discussed at length in the orthopedic literature, and current research indicates that long-term immobilization has a deleterious effect on healing, including atrophy and alterations in the makeup of connective tissue, while controlled mobilization of the injured area has positive impact on functional outcomes [18–22]. It is not clear if this can be generalized to the vocal folds, as there are differences in the nature of the tissues and the type, frequency, and duration of mobilization required.

The effects of rest and mobilization on tissue healing have not been studied in human vocal folds, but research has been done using animal models and bioreactors. With artificially induced phonation in a rabbit model, Rousseau et al. found increases in gene expression of metallopeptidase (MMP)-1, an enzyme that breaks down collagens I and II, no changes in expression of MMP-9, and no changes in interleukin-1β (involved in inflammation), suggesting that vocal fold vibration may impact gene expression involved in healing.

Using a bioreactor to control mechanical stress on vocal fold fibroblasts, Titze et al. found that genes associated with the extracellular matrix expressed more with mechanical stress than with rest [23]. Kutty and Webb found that vocal fold fibroblasts exposed to vibration showed increased expression of hyaluronic acid synthase 2, fibromodulin, and decorin compared to fibroblasts that were kept still, while collagen and elastin were not significantly affected by vibration, indicating that mobilization plays a role in gene expression and likely influences tissue healing [24].

Adherence to voice rest is difficult in adults and is assumed to be especially difficult in children. Rousseau et al. found that overall compliance with voice rest was 34.5%, although it was higher after surgery than for other reasons [25]. Compliance with voice rest in children has not been studied but, given developmental considerations, is not likely to be better than in adults. As such, during preoperative counseling we discuss strategies for communication when not speaking, which may include use of paper and pen, low-tech strategies such as a picture board with commonly used phrases and requested items, or more high-tech options including text-to-speech apps for phone or tablet for children who can write. The use of a simple text-to-speech app has been shown to result in higher self-reported communicative effectiveness in adults on voice rest after surgery [25]. Activities that are enjoyable but do not require talking can be brainstormed, and parents and children can discuss special privileges they might get during their recovery, such as a special coloring book or the chance to watch a preferred show or movie.

Voice Therapy

Voice therapy is generally accepted by clinicians as helpful in the postoperative period, although again there is little agreement on when to begin, how much to do, and what type of therapy is most helpful. Koufman and Blalock found that in adults, preoperative voice therapy was associated with reduced postoperative dysphonia [26]. There is no evidence that clearly indicates one approach to therapy as favorable to another. In clinical practice, the decision on what to do in therapy often depends greatly on the age of the

child, the type of laryngeal lesion, and the type of surgery. In the case of benign lesions, children will generally have had a course of behavioral voice therapy prior to considering surgical excision. Ideally, the child will have multiple sessions of therapy prior to surgery, to reduce maladaptive compensatory behaviors associated with the vocal pathology, achieve optimal voicing, and train vocal exercises. One session of therapy prior to surgery should focus on discussion of voice rest recommendations, strategies for adherence to voice rest, and training of postoperative voice therapy exercises and target voice. Kaneko et al. used tube phonation (a form of semi-occluded vocal tract exercise) for 6 weeks postoperatively. Resonant voice exercises were used by Verdolini Abbott and colleagues in studying the effect of voice therapy vs voice rest on phonotrauma [27]. They studied participants after an intense vocal loading task assumed to cause phonotrauma and found that certain markers of inflammation in the vocal fold secretions were reduced after resonant voice exercises compared with both spontaneous speech and voice rest [27]. While this cannot be clearly generalized to recovery after surgery, it is encouraging in the role of large-amplitude, low-impact vibrations (as present in resonant voice therapy and semi-occluded vocal tract exercises) in the role of healing. Voice exercises such as cup bubbles, straw phonation, lip trills, resonant humming, and gentle pitch glides are frequently employed, as they are instrumental in coordinating subsystems of voice and facilitating optimal glottal configuration and vibration during voicing. Clinicians may work on resonant voice-based therapy (e.g., Lessac-Madsen resonant voice therapy) or flow phonation to bring the efficiency and coordination into connected speech. As with all voice therapy, clinicians should tailor their approach to both their knowledge of the anatomy and physiology postoperatively and the individual learning style and needs of the patient.

In summary, the available evidence indicates that some degree of voice rest followed by controlled voicing and voice therapy is optimal for functional recovery after surgery. Based on the wound healing literature, our clinical practice is typically to recommend 3 days of complete voice rest, with some voice conservation for 2 weeks, and early implementation of semi-occluded vocal tract exercises and resonant voice beginning 3 days after surgery. We recommend voice therapy continuing for 2–3 months after surgery as there are likely changes continuing to occur in the vocal fold tissue, and patients may need assistance in adapting to these changes and maximizing their vocal gains.

The Otolaryngologist's Perspective

Reflux Treatment

Empiric treatment to prevent laryngopharyngeal reflux during the perioperative period is controversial. It would seem obvious that acid reflux would be detrimental to wound healing as it has been implicated in laryngotracheal injury [28–30]. That said, however, empiric treatment may not come without risk. Neutralization of the stomach may allow for pathogenic bacterial overgrowth which may be detrimental to a positive outcome. Personal experience can testify to gastric pseudomonal overgrowth and subsequent cartilage graft loss after laryngotracheal reconstruction and perioperative empiric proton pump therapy. Prudent use of proton pump inhibitors and prophylactic antibiotics are commonly employed and likely promote healing and graft survival although distinct scientific study is lacking. The exact timing of acid suppression and antibiotic therapy is not known. Time for wound healing and reepithelialization would seem to be the key for treatment length. Four to 12 weeks of acid suppression is typical after airway reconstruction or microlaryngeal surgery.

The implications of the change in the microbiome secondary to acid suppression and antibiotics treatment is beginning to pique interest in clinicians [31–33]. Empiric acid suppression and antibiotic usage may have unintended and seemingly counterintuitive outcomes. If laryngopharyngeal reflux is known or significantly suspected, however, treatment is important. A proton pump inhibitor is indicated when a diagnosis of

laryngopharyngeal reflux is supported. Other treatments such as H2 blockers may be used when the suspicion is lower. As the literature is lacking, personal preference often plays a role in the treatment of patients postoperatively after laryngeal surgery or airway reconstruction. The prevention of caustic exposure from acidic gastric contents must be balanced with the potential disruption and change to the microbiome of the patient and its potential long-term effects.

Perioperative Steroids

Perioperative steroids are often used after airway reconstruction or phonomicrosurgery for the prevention of scar formation and to decrease swelling, assisting in airway management. Oral, intralesional and other parenteral administration of steroids have been utilized for this purpose [34–36]. Intralesional dexamethasone has been used to hydrodissect the epithelium away from the laryngeal lesion with the added benefit of decreasing the risk of scar formation. Care must be taken as there is a risk of causing temporary atrophy of the vocal fold with intralesional injection [37]. Intraoperative intralesional injections with dexamethasone are used during laryngeal phonosurgery. Intravenous dexamethasone is used during the 24–48 h prior to extubation after airway reconstruction. It would be rare to use prolonged oral steroids in the perioperative period.

Voice Rest

As outlined above, there is a sweet spot for voice rest after phonomicrosurgery. The precise length of voice rest and the timing of reinstituting graded phonation for optimal outcome have not been well studied. As stated above, we prescribe 3 days of absolute voice rest followed by 2 weeks of conservative voice use, progressing back to full voicing after this time. We would typically wait until we feel confident that the child can complete this course of voice rest prior to performing phonomicrosurgery. There are no hard rules for when a child will be able to follow the voice rest protocol and must be assessed individually. Typically, children will be older than 4 or 5 years of age, although depending on the urgency of surgery and the temperament of the child, this may be older or younger.

Evolving and Emerging Techniques

As mobile technology has become ubiquitous even for children and adolescents, its role in vocal rehabilitation is growing. Postoperatively, phones and tablets can be used for communication, with simple text-to-speech or picture selection apps for nonverbal communication. Text reminders to rest the voice and to do rehabilitation exercise can be programmed or sent to a smart phone or mobile device. Additionally, vocal dosimeters are a form of mobile technology that can objectively monitor voice rest after surgery and better quantify adherence to recommendations; if this can be combined with already existing mobile technologies like phones and wearables, it could be instrumental in helping patients monitor and adhere to voice rest and rehabilitation.

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5 Imaging Evaluation of the Upper Aerodigestive Tract

Tiffany Zens and Kara Gill

Anatomical Review

Several anatomic differences between children and adults must be considered when interpreting imaging of the upper airway and gastrointestinal tract. The pediatric trachea is shorter than adults. The larynx is more anterior and superior, lying at the level of C3 to C4 [1]. The narrowest portion of the pediatric airway is the cricoid cartilage. The epiglottis is relatively larger than in adults. Additionally, children have larger tonsils and increased soft tissue which can further contribute to airway obstruction [2]. The tongue of a child is short and broad. During suckling, the larynx elevates to allow the child to breathe and feed at the same time. The pediatric esophagus has the same contraction pattern as adults but ends approximately two vertebral bodies higher than in adults [3].

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Indications for Common Imaging Modalities

Plain Films

Upright lateral and anterior radiographs of the neck and chest can be instrumental in determining the etiology of a child presenting with difficulty breathing or swallowing. Patients should be positioned with their head in a neutral position or slightly extended to avoid neck flexion which can exacerbate respiratory symptoms or create pseudothickening artifact of the retropharyngeal tissues [2] (Fig. 5.1). Imaging of the chest can be performed in either the AP or PA projection with best attempt made to obtain the image during inspiration (Fig. 5.2).

The AP view is best for looking at the cervical and upper thoracic airway and adjacent soft tissues. The lateral view is used to evaluate the degree of adenoid and tonsillar hypertrophy, the epiglottis and aryepiglottic folds, as well as the prevertebral soft tissues. Metallic or dense foreign bodies can be seen on both views. It is always important to consider radiation exposure when deciding how many views to obtain. Consider whether one view will answer the clinical question or if two are needed. For example, the adenoids cannot be seen on an AP view, so only the lateral needs to be performed.

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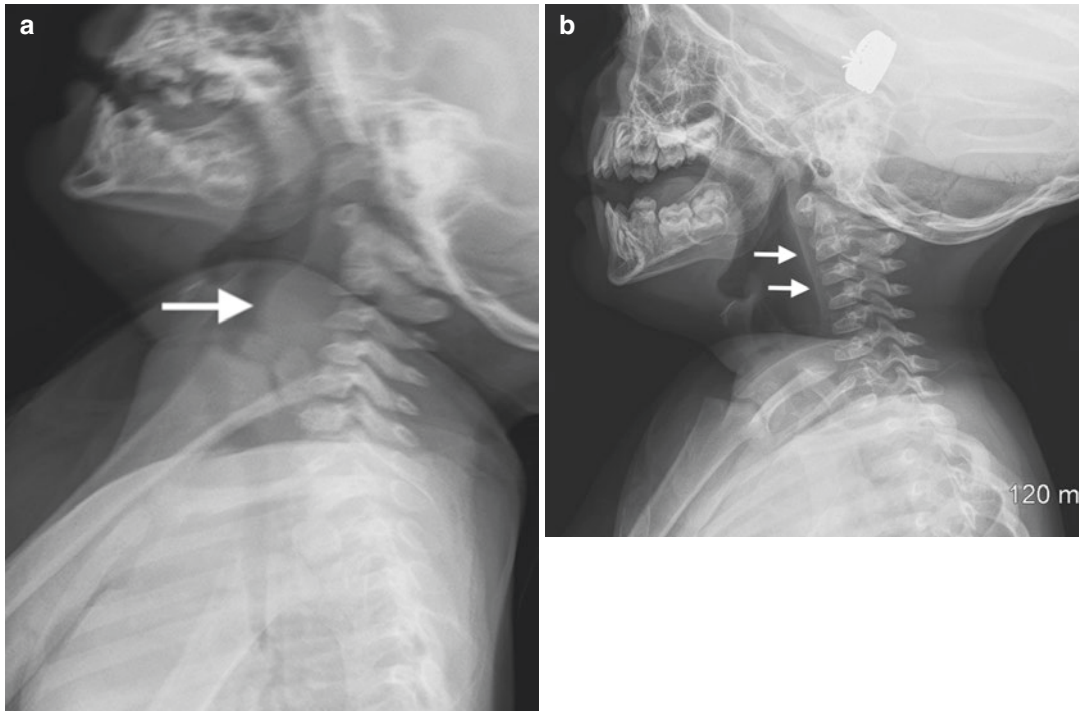

Fig. 5.1 (**a**) Initial lateral image of the neck soft tissues in a 19-month-old male is limited by motion artifact, but shows apparent widening of the prevertebral soft tissues (∗). Within minutes, the image was repeated with better positioning, (**b**) revealing thin, normal prevertebral soft tissues (arrows)

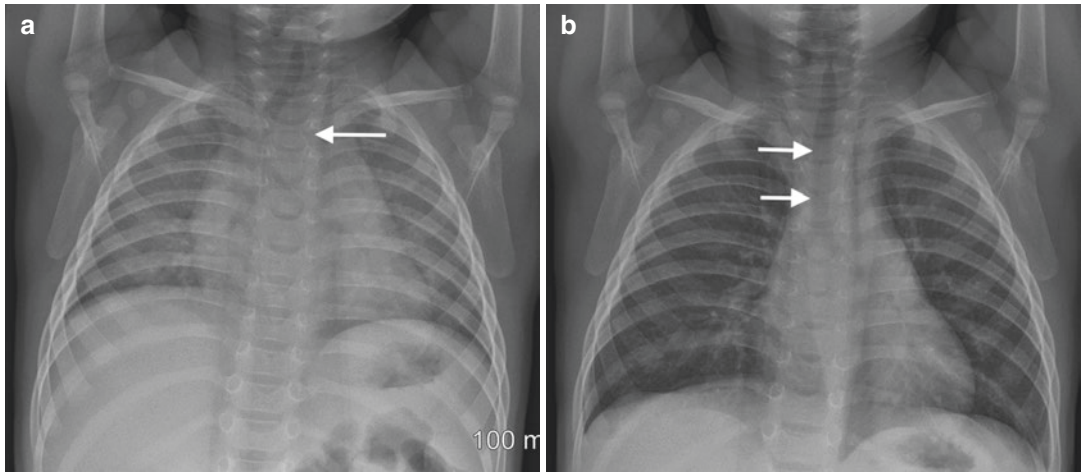

Fig. 5.2 (**a**) Initial AP radiograph of the chest done in a 10-month-old female to evaluate for source of fever shows rightward deviation of the trachea (arrows). The subsequently performed AP radiograph obtained in inspiration (**b**) shows a normal midline trachea. The apparent tracheal deviation on the initial image was related to expiratory technique rather than a mediastinal mass

Modified Barium Swallow Study (MBSS)/Videofluoroscopy

MBSS is performed through the collaborative efforts of a speech pathologist and a radiologist. Images can be obtained in lateral and AP projections. Neonates and young infants are generally imaged in a side-lying position, whereas older infants and children are seated for the exam [4]. Various consistencies of barium (liquid, thickened liquid, semisolid, and solid) can be administered to determine what foods result in aspiration and which are safe for the patient to eat. Beyond determining only the presence or absence of aspiration, the swallow study evaluates where, when, and why the swallow breaks down and looks at alterations in structure and deficits in strength and timing. Positioning, viscosity, and method of delivery can all be altered to determine the safest and most effective way for an infant or child with dysphagia to eat. In older children, it is often best to attempt to cover their favorite foods with barium. For example, mix barium into yogurt or sprinkle barium powder in a peanut butter and jelly sandwich. By allowing the child to choose the foods they eat, tolerance of the exam improves. Additionally, it is important to test foods that parents report the child has problems with at home.

The field of view should be limited to the superior margin of the nasal cavity to avoid radiation to the eyes. The inferior margin of the image should include the upper chest so that tracheoesophageal fistulas and reflux can be seen. The mouth needs to be included in the field of view so that both the oral and pharyngeal phases of swallowing can be evaluated [4]. There is much debate between speech pathologists and radiologists regarding an acceptable frame rate by which to obtain images. There needs to be a balance between trying to decrease radiation exposure in pediatric patients who are more susceptible to the potential deleterious effects of radiation and the need to make a diagnosis and plan treatment. Depending on the institution, frame rates ranging from 3 to 30 frames per second are used. Bonilha et al. described differences in judgments of swallow impairment between 30 frames per second and simulated 15 frames per second in a cohort of only 5 patients [5]. Further study on larger pediatric populations will be required to determine how to optimize exam sensitivity while limiting radiation exposure. Importantly, though a lower frame rate may be adequate to detect the presence of aspiration, a higher frame rate may be needed to determine why it occurred and what interventions could help address it.

Images need to be scrutinized for any of the following abnormalities: nasopharyngeal reflux, penetration (Fig. 5.3a), aspiration (Fig. 5.3b), intraesophageal/gastroesophageal reflux, and retrograde flow of contrast from the esophagus into the airway as with H-type tracheoesophageal fistula. It is also important to comment on adenoid or palatine tonsil hypertrophy (Fig. 5.4) as this can be another cause of noisy breathing or dysphagia. Lastly, as the trachea lies directly anterior to the esophagus, caliber change of the trachea due to tracheomalacia can be observed.

Esophagram

This exam is performed by a radiologist with the patient in the supine versus upright position depending on age. Images of the esophagus need to be obtained in both the AP and lateral dimensions and include the cervical esophagus through the gastroesophageal junction. It is preferable that the patient drink contrast via bottle, straw, or age-appropriate cup. The examination can, however, still be diagnostic by administering contrast via a syringe. If the patient is unable or unwilling to drink via those mechanisms, a feeding tube can be inserted into the proximal esophagus for injection of contrast. If there is a concern for esophageal perforation or anastomotic leak, water-soluble,

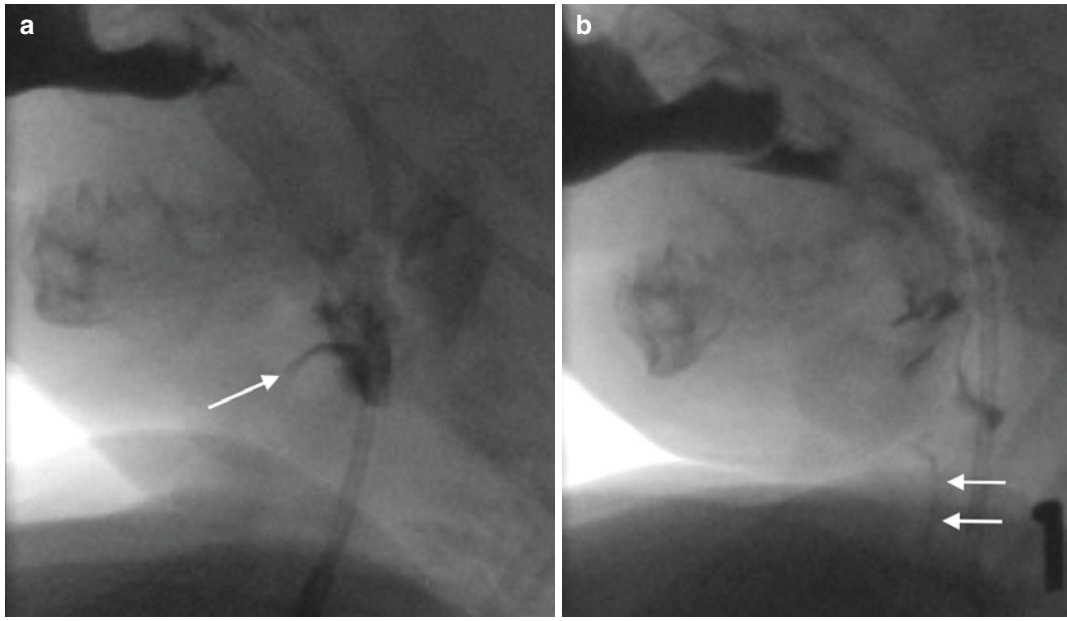

Fig. 5.3 A 3-month-old male with history of laryngomalacia s/p supraglottoplasty 2 months prior who presented with increased gagging and choking with feeds. (**a**) Single image from a videofluoroscopic swallow study performed in a seated, recumbent position shows contrast within the valleculae and piriform sinuses with thin extension into the larynx, consistent with penetration (arrow). (**b**) A second image from the same swallow study shows aspiration: contrast spillage into the trachea with layering along the posterior wall (arrows)

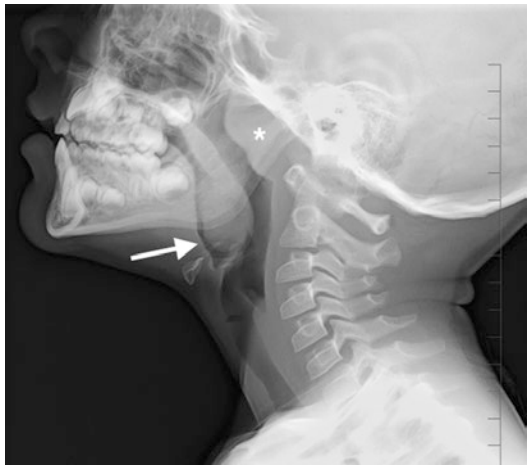

Fig. 5.4 Lateral view of the neck soft tissues in a 6-year-old male demonstrates marked hypertrophy of the palatine tonsils (arrow). The adenoids (*) are also enlarged and cause mild to moderate narrowing of the nasopharynx

low-osmolality contrast should be administered. High-osmolality water-soluble contrast such as gastrografin should be avoided as it can cause chemical pneumonitis when aspirated. All other patients can be given liquid barium. Esophageal caliber and motility should be commented on, as well as abnormal extrinsic mass effect caused by a vascular ring, sling, or mass. There are three normal impressions on the esophagus. First, the aorta should have an impression on the left lateral aspect of the upper thoracic esophagus in the AP view and an anterior impression on the lateral view. Second, an impression by the left main stem bronchus is variably seen sloping downward from right to left on the AP view. Third, on the lateral view, there is a gentle impression on the posterior wall of the esophagus by the left atrium (Fig. 5.5). Lastly, evaluation for gastroesophageal reflux should be performed. For infants and small children who cannot follow directions well, this is done by rolling the patient from side to side. Older children can be asked to cough or perform a Valsalva maneuver. That being said, the esophagram only captures a few moments in time and is not 100% sensitive for reflux.

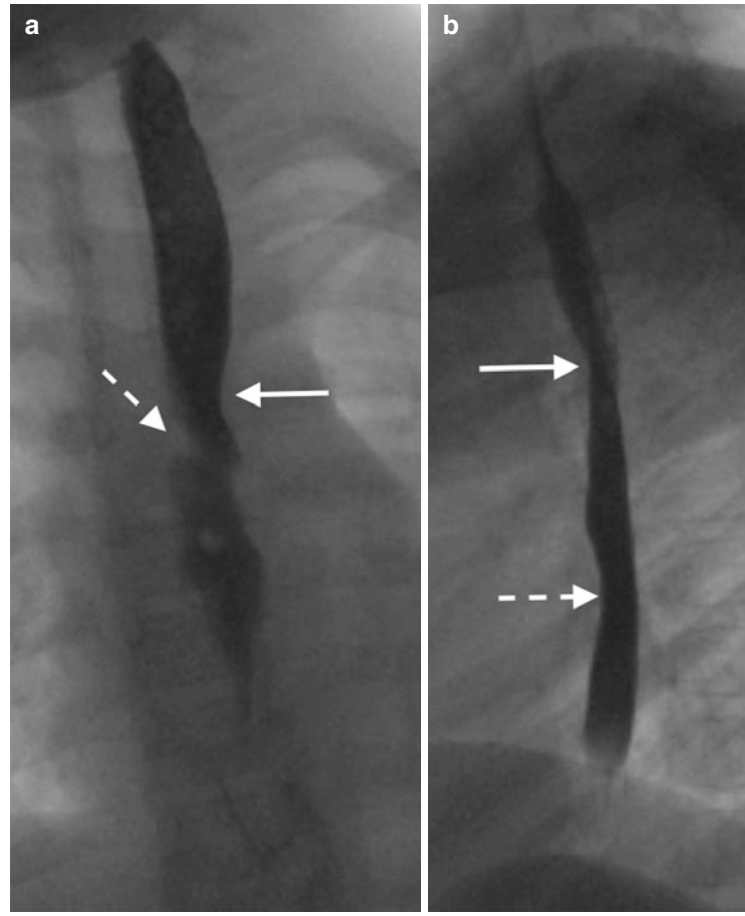

Fig. 5.5 (**a**) AP view obtained during an esophagram shows normal extrinsic compression on the upper left lateral esophagus by the aortic knob (arrow). The relative lucency sloping downward from right to left below this level is due to normal compression of the anterior esophagus by the left main stem bronchus (dashed arrow). (**b**) Lateral view obtained during the same esophagram shows mass effect upon the anterior proximal esophagus by the aortic knob (arrow) and anterior mass effect on the distal third of the esophagus by the left atrium (dashed arrow)

Computed Tomographic (CT) Imaging

When performing CT to evaluate the upper aerodigestive tract in pediatric patients, intravenous contrast is always needed in order to delineate the soft tissues from the vascular structures. Anatomy and pathology will be more clearly identified in case surgery is indicated. When evaluating for a vascular ring or sling, only a single-phase study is indicated following injection of contrast. When the airway is being evaluated for malacia, vascular compression, or congenital abnormality, contrast-enhanced imaging is done with a two-phase scan – one in inspiration and one in expiration. Anesthesia has traditionally been needed to obtain adequate imaging in infants and small children who cannot hold their breath. An endotracheal tube cannot be used as it limits the ability to evaluate the trachea; however, satisfactory inspiratory and expiratory imaging can be done with a laryngeal mask airway.

With newer, wide-detector dynamic CT imaging, continuous low-dose scanning can be performed over the respiratory cycle, providing more physiologic evaluation of the airway during both inspiration and expiration without the need for anesthesia [6]. This allows for evaluation of children with noisy breathing or respiratory distress with a differential diagnosis including tracheobronchomalacia, vascular ring, and innominate artery compression syndrome.

Magnetic Resonance Imaging (MRI)

Newer techniques make it possible to image the large airways with MRI and evaluate for intrinsic large airway disorders such as tracheobronchial

branching anomalies, tracheobronchomalacia, congenital stenosis, vascular rings and slings, intrinsic large airway neoplasms, and infectious disorders [7]; however, this has not yet become universal. Although older children can undergo this procedure with adequate coaching, special equipment including an MRI-compatible spirometer is required for dynamic imaging of the airway. Sedation is required for almost all patients under the age of 6 [7].

Another important application of MRI for evaluation of voice and swallowing disorders is the evaluation for an underlying Arnold-Chiari malformation. Arnold-Chiari malformation refers to the downward herniation of the hindbrain below the level of the foramen magnum [8]. Four subtypes are described, and types I and II are more commonly encountered. Type I is characterized by caudal descent of the cerebellar tonsils; symptoms may not arise until adulthood [9]. Type II involves herniation of the cerebellar vermis, medulla, and fourth ventricle, which may lead to hydrocephalus or myelomeningocele; symptoms are common in infancy [8]. Type III includes occipital encephalocele, and type IV is associated with cerebellar hypoplasia [8]. Importantly, patients with type I and II malformations may present with vocal fold mobility impairment and aspiration [10, 11]. MRI is the imaging modality of choice for diagnosis. Multiplanar T1- and T2-weighted images with a slice thickness less than 3 mm are recommended, covering the entire skull, brain, spine, spinal cord, and cerebrospinal fluid spaces [12]. Diagnostic criteria for type I malformation includes caudal displacement of one cerebellar tonsil >5 mm or both tonsils 3–5 mm below a virtual line connecting the basion with the opisthion of the foramen magnum [12] (Fig. 5.6). Importantly, some children with tonsillar displacement >5 mm may be asymptomatic [13], and others with displacement of only 3–4 mm may be symptomatic [14]; it is important to look not only at the tonsillar position but also whether the tonsils are compressed and the perimedullary cerebrospinal fluid spaces are effaced [12]. If a cause for vocal fold mobility impairment or aspiration cannot be identified based on history and exam, further investigation with MRI can be helpful.

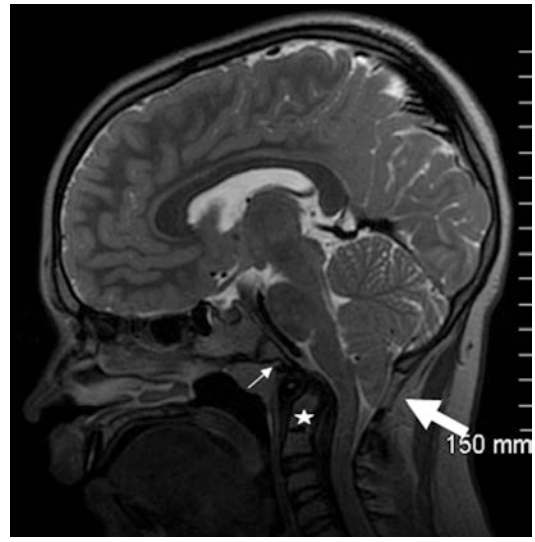

Fig. 5.6 Sagittal T2-weighted magnetic resonance image of the brain in a 13-year-old male with Chiari I malformation showing >7 mm tonsillar ectopia (large arrow) with shortening of the clivus (small arrow) and posterior orientation of the dens (star), resulting in effacement of the cerebrospinal fluid ventral to the brainstem

Acute Disease Processes

Epiglottitis

Clinical Presentation and Etiology

Acute epiglottis is a medical emergency which requires prompt airway evaluation and management. This disease process is caused by infectious agents (*Haemophilus influenzae*, *Staphylococcus aureus*, *β-hemolytic Streptococcus*, *Neisseria*, and *Streptococcus pneumoniae*) or noninfectious agents (angioedema/anaphylaxis, trauma, or ingestion). Epiglottitis results in cellulitis, edema, and inflammation of the epiglottis and surrounding tissue (including the aryepiglottic folds and subglottis) [15, 16]. Children with epiglottitis usually present with acute, rapid-onset stridor, dysphagia, sore throat, fever, and respiratory distress. They typically assume the "tripod position," sitting upright, leaning over with a hyperextended neck, and drooling. Prompt evaluation by otolaryngology and emergent intubation is the recommended initial treatment for acute epiglottitis [16]. Fortunately, vaccines have decreased the overall incidence of epiglottitis.

Radiographic Findings

Acute epiglottitis is often a clinical diagnosis which does not require imaging. That being said, lateral radiograph of the neck will demonstrate classic findings consistent with swelling of the epiglottis called the "thumb sign" (Fig. 5.7). Normally, the epiglottis has defined margins and the aryepiglottic fold is convex inferiorly. In acute epiglottitis, submucosal edema causes a thumb sign and narrowed vallecula [2, 17].

Retropharyngeal Abscess

Clinical Presentation and Etiology

Retropharyngeal abscesses occur in the neck between the posterior pharyngeal wall and prevertebral fascia along a chain of lymph nodes which drain the nasal cavity, adenoids, eustachian tubes, and posterior paranasal sinuses [18]. Abscesses are typically the result of suppuration of lymph nodes from an upper respiratory tract infection, but can also be seen after trauma, instrumentation, or ingestion of foreign bodies or caustic agents. Abscesses are typically polymicrobial and include respiratory anaerobes, *Staphylococcus aureus*, *Haemophilus influenzae*, and *Streptococcus pneumoniae* [2]. The incidence of retropharyngeal abscesses is most common in children under 5 years old [18]. Patients typically present with fever, neck pain, cervical lymphadenopathy, and dysphagia. Retropharyngeal abscesses are treated with supportive care, humidified oxygen, airway management, broad spectrum antibiotics, and possible incision and drainage [18].

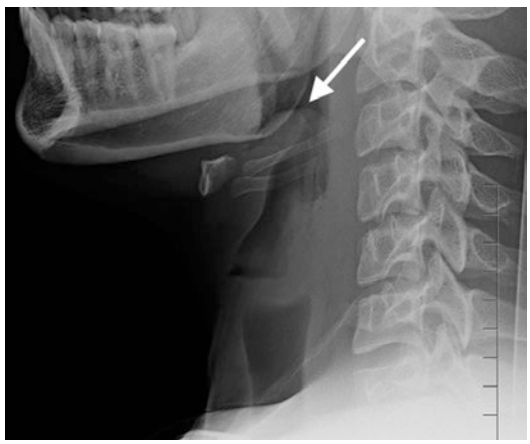

Fig. 5.7 Lateral view of the neck soft tissues in a 17-year-old male with 5-day history of fever, worsening sore throat, and dysphonia demonstrates thickening of the epiglottis consistent with the "thumb sign" (arrow). The aryepiglottic folds are also thickened

Radiographic Findings

Upright and lateral radiographs will demonstrate thickening of the prevertebral soft tissues to >7 mm at C2 or >14 mm at C6 [18] (Fig. 5.8a). Contrast-enhanced CT is generally performed to confirm the diagnosis and assist with surgical planning (Fig. 5.8b). If cross-sectional imaging is obtained for preoperative planning or evaluation of complications, it should be extended to the mediastinum in order to evaluate for secondary mediastinitis [19].

In advanced disease, arterial and venous abnormalities may be seen on ultrasound or contrast-enhanced cross-sectional imaging, including jugular vein thrombosis (Lemierre's syndrome) and pseudoaneurysm [2].

Croup

Clinical Presentation and Etiology

Croup (laryngotracheobronchitis) is a subglottic inflammatory process which affects approximately 3% of children and is caused by the parainfluenza virus in 75% of cases [20]. This infection is most common in children 6 months to 3 years old [21], but can be seen in older school-age children. Children present with respiratory distress and a barking cough. It is typically treated conservatively with oxygen, humidification, steroids, and nebulizers [20]. Croup is generally a clinical diagnosis, with imaging of the neck soft tissues or chest being performed only in those who are not appropriately responding to therapy which raises concern or additional pathology (foreign body, tracheitis, superimposed bacterial pneumonia).

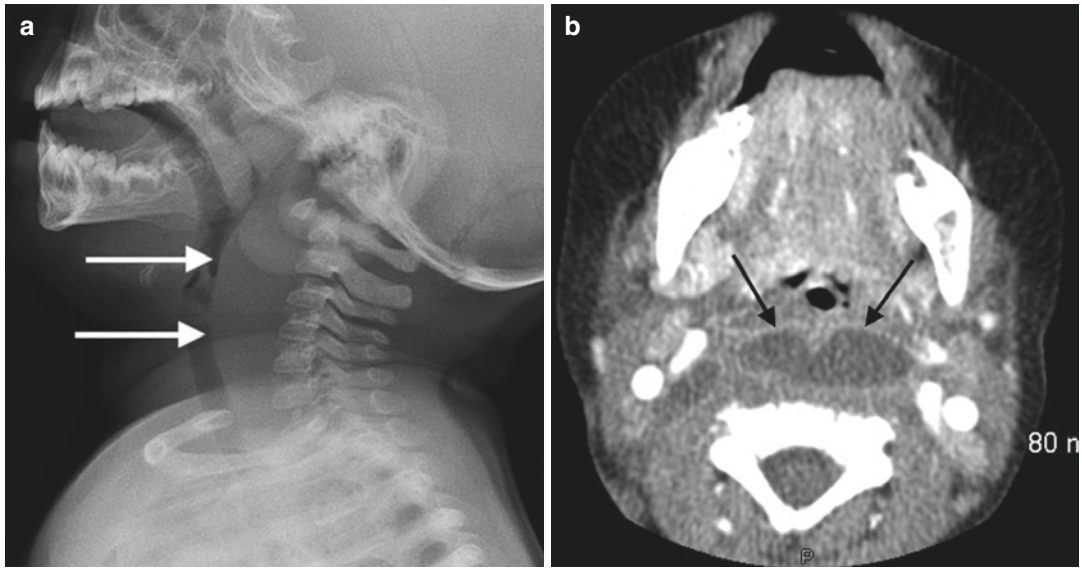

Fig. 5.8 (**a**) Lateral view of the neck soft tissues in a 9-month-old female with 4-day history of fever, cough, respiratory distress, and neck stiffness demonstrates thickening of the prevertebral soft tissues which are greater than the width of the C2 vertebral body (arrows). (**b**) Subsequently performed contrast-enhanced CT of the neck demonstrates a rim-enhancing retropharyngeal fluid collection consistent with abscess (arrows)

Radiographic Findings

Plain film radiographs of the soft tissues of the neck are the diagnostic imaging modality of choice for croup [22]. In croup, the normal, smooth subglottic larynx will have an elevation and loss of lateral convexities, causing a classic steeple sign or inverted V sign on frontal radiographs and subglottic narrowing, hypopharynx overdistension, or increased subglottic density on lateral radiographs [2] (Fig. 5.9). The differential diagnosis for these radiographic findings would include angioedema and tracheitis depending on the clinical history.

Foreign Body

Clinical Presentation and Etiology

Foreign body ingestion is a common occurrence in the pediatric population. The incidence is highest among children 6 months to 3 years old [23]. Ingested items can include toys, magnets, batteries, buttons, or retained food. The most common retained esophageal foreign bodies are coins [23]. Up to 50% of children are asymptomatic after foreign body ingestion, but symptomatic children may present with drooling, gagging, dysphagia, coughing, or hematemesis. The most common area for a retained foreign body is at the level of the upper esophageal sphincter (60–75%). Foreign bodies are also commonly found lodged in the mid-esophagus at the level of the aortic knob and near the lower esophageal sphincter [24, 25]. Some ingested foreign bodies can be allowed to pass through the gastrointestinal tract without incident. Indications for intervention include obstruction at the upper esophageal sphincter, evidence of esophageal or intestinal perforation, intestinal obstruction, and ingestion of potentially dangerous items such as sharp objects, multiple high powered magnets, or batteries [23].

Foreign body aspiration in the pediatric population accounts for approximately 150 deaths annually in the USA [26]. Children are at higher risk for obstruction from aspiration given their narrow airways. The most common aspirated items are food and toys (such as balloons, marbles, and balls) [23]. Bronchoscopy is the gold standard for removal of aspirated foreign bodies [23].

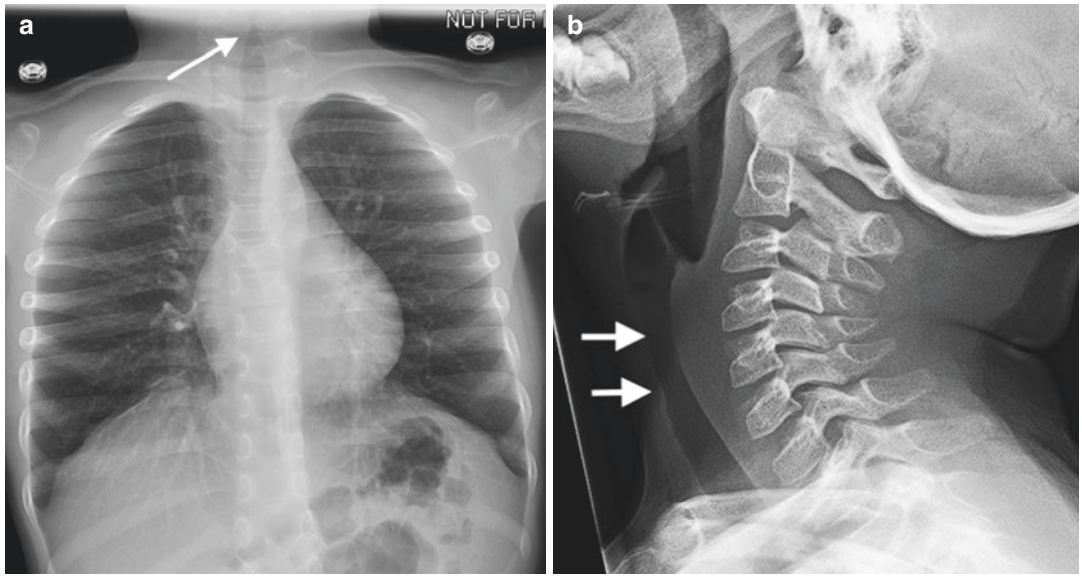

Fig. 5.9 (**a**) AP view of the chest in a 6-year-old male demonstrates narrowing of the subglottic trachea (arrow) resulting in the "steeple sign," consistent with croup. The lungs are clear. (**b**) The lateral view of the neck in the same patient demonstrates narrowing of the cervical trachea (arrows)

Radiographic Findings

Radiographs are the first diagnostic tool used to identify foreign bodies. This often includes both anterior-posterior and lateral projections of the neck, chest, and abdomen. The position of radiopaque objects can be clearly delineated on radiographs. Indirect signs (such as gaseous distention or air/fluid levels in the esophagus) can often be used to identify radiolucent foreign bodies in the gastrointestinal tract [23]. Coins are the most commonly ingested foreign body. If the coin is in the esophagus, it will appear as a circular disc on anteroposterior imaging and on edge as a thick line on lateral imaging (Fig. 5.10). In contrast, coins in the trachea will appear as circular discs on the lateral view and as thick lines on edge in the anteroposterior film [27]. Button batteries will have a double ring appearance when seen en face on radiographs which distinguish them from coins [28] (Fig. 5.11). When magnets are ingested, it is important to correctly identify the number of magnets so they can be monitored as they move through the digestive system. Magnets in adjacent loops of small bowel have the potential to attract the bowel walls together, causing necrosis, perforation, or fistula formation. If a symptomatic child has ingested a foreign body, but the radiographs are negative, CT or fluoroscopy can be used with the help of oral contrast to identify filling defects and the location of the foreign body within the esophagus [28].

When there is a clinical concern for an aspirated foreign body due to respiratory distress or wheezing after a choking/gagging episode, imaging of the chest should be done to evaluate for asymmetric air trapping. An aspirated foreign body, usually radiolucent food products, in the airway acts like a ball valve: air can get in, but the foreign body obstructs the lumen during expiration so that air cannot get out. Imaging protocol will depend on the age and cooperation of the patient. In infants and young children who cannot follow breathing instructions, the AP radiograph is compared to bilateral decubitus views of the chest. Whichever side is down should show volume loss in the absence of bronchial obstruction. If appropriate volume loss is not seen, this suggests obstruction of the bronchus by a radiolucent foreign body (Fig. 5.12).

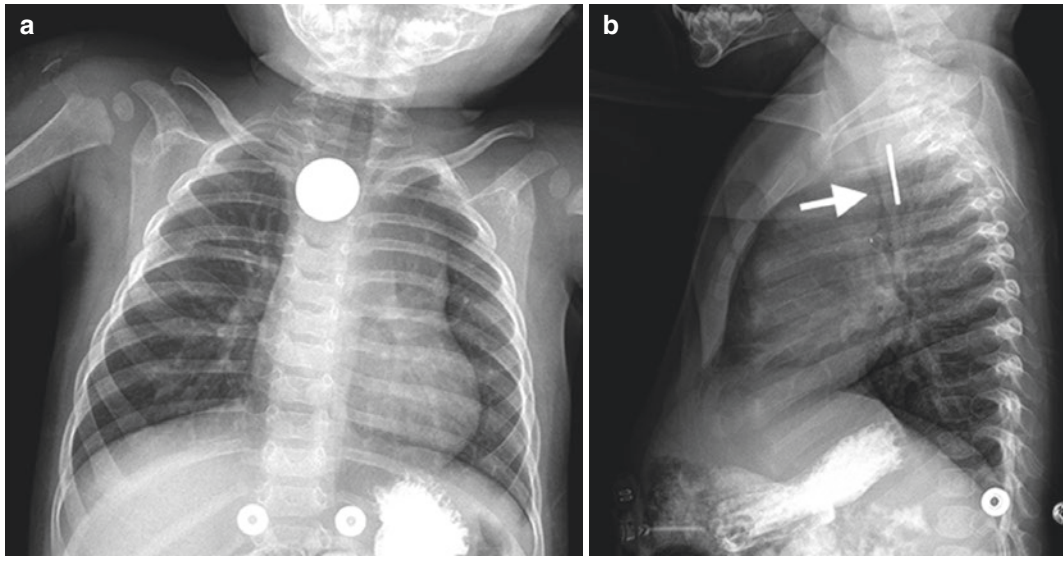

Fig. 5.10 An 8-month-old male with history of esophageal atresia with tracheoesophageal fistula who was found to have a coin in the upper esophagus on a gastrostomy tube check. Subsequently obtained AP view of the chest (**a**) demonstrates a homogeneously dense round metallic foreign body consistent with a coin in the upper esophagus, likely just above the level of the esophageal anastomosis. The lateral view (**b**) shows narrowing of the trachea anterior to the coin (arrow), either due to inflammation related to chronic foreign body or tracheomalacia. This information is important for surgical planning as airway inflammation could impact the patient's ability to maintain their airway while anesthetized for foreign body removal

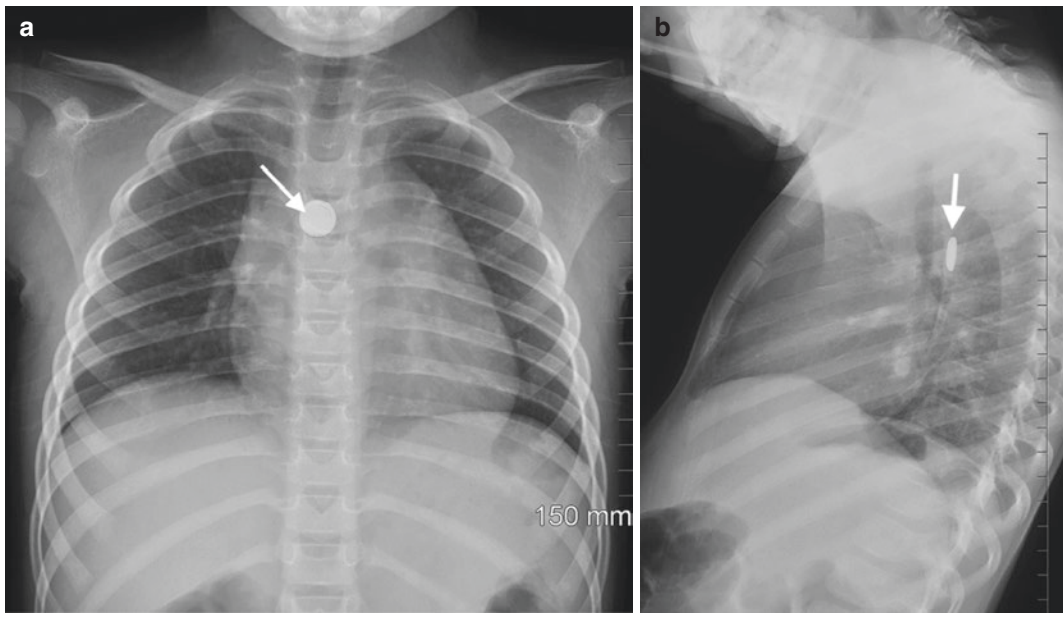

Fig. 5.11 (**a**) A 4-year-old female found to have a button battery in the mid-esophagus at the level of the carina. The AP view of the chest best demonstrates the thin, lucent circle between the anode and cathode plates which differentiates a button battery from a coin (arrow). (**b**) The lateral view demonstrates the beveled edge of a button battery (arrow)

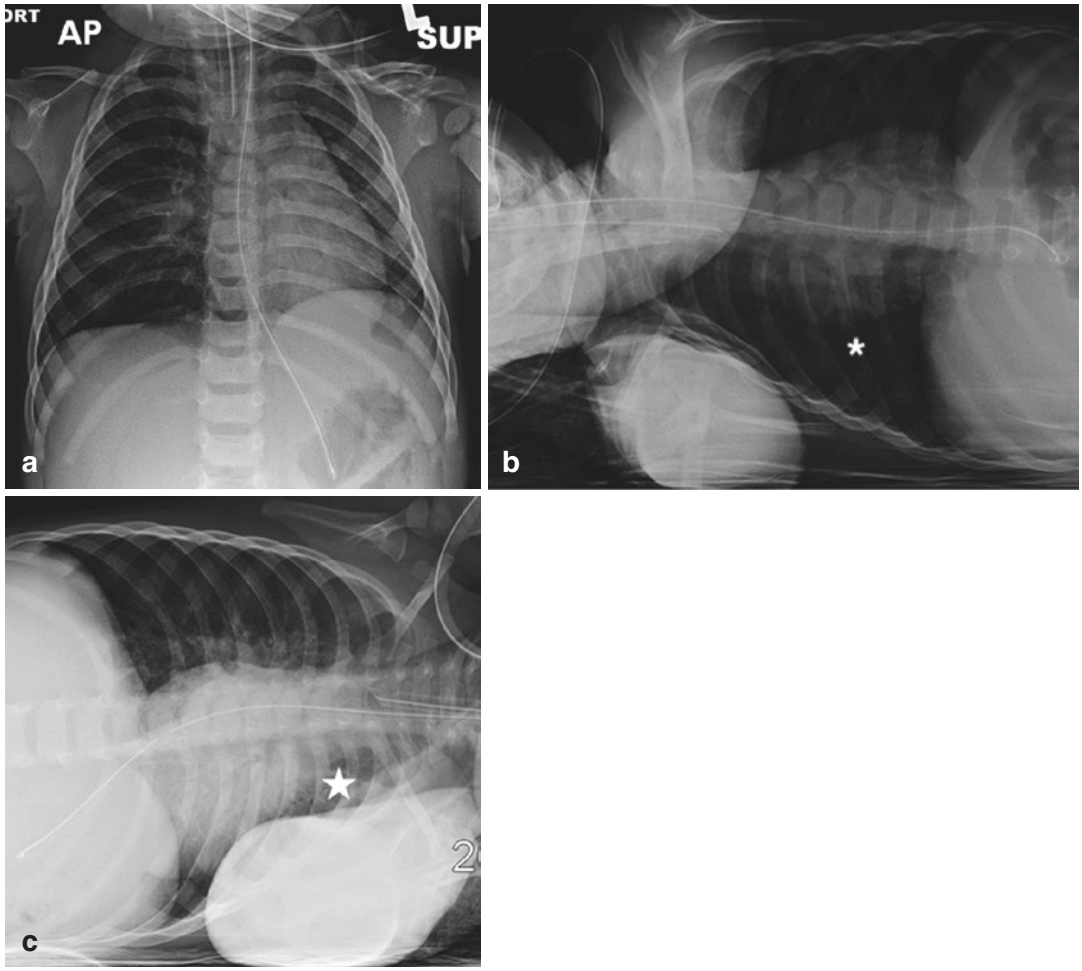

Fig. 5.12 A 9-month-old female admitted to the PICU for acute respiratory failure requiring intubation. (**a**) AP image of the chest demonstrates asymmetric increased lucency of the right hemithorax with leftward mediastinal shift. Given the patient's age and clinical status, bilateral decubitus AP views of the chest were obtained to evaluate for air trapping. In the right lateral decubitus position (**b**), the right lung (∗) remains lucent and does not show appropriate volume loss or development of dependent atelectasis. There is appropriate volume loss in the left lung (star) when the patient was moved into the left lateral decubitus position (**c**). These findings confirm air trapping due to a radiolucent foreign body in the right mainstem bronchus. Multiple peanut fragments were removed during bronchoscopy

Chronic/Congenital Disease Processes

Tracheal Narrowing

Clinical Presentation and Etiology

Tracheomalacia is a dynamic narrowing of the lumen of the trachea during breathing and is caused by a weakness in the tracheal wall. The trachea has C-shaped cartilaginous rings anteriorly and a soft membranous component posteriorly which allows for normal changes in the diameter of the trachea with respiration. In tracheomalacia, this variation is excessive (>50%) and results in obstructive symptoms including cough, recurrent infection, dyspnea, feeding difficulties, or stridor [29]. Intrathoracic tracheomalacia presents with wheezing on expiration, and extrathoracic tracheomalacia presents with stridor on inspiration [30]. Treatment of tracheo-

malacia is based on severity and can include use of positive-pressure ventilation, aortopexy, and tracheostomy placement [31].

Complete tracheal rings are the most common cause of congenital tracheal stenosis but still account for <1% of cases of airway stenosis in children [32]. This anomaly results from a defect during embryogenesis which results in the posterior membranous portion of the tracheal ring being absent [32, 33]. As a result, the tracheal cartilage is circumferential rather than u-shaped, and the lumen becomes narrowed and circular in appearance (Fig. 5.13). Children present with "washing machine" stridor, cyanosis, and respiratory distress. Children with symptomatic or long-segment complete tracheal rings often require surgical repair, while patients with an incidental finding of complete tracheal rings later in life may be treated conservatively with surveillance bronchoscopy [32, 34].

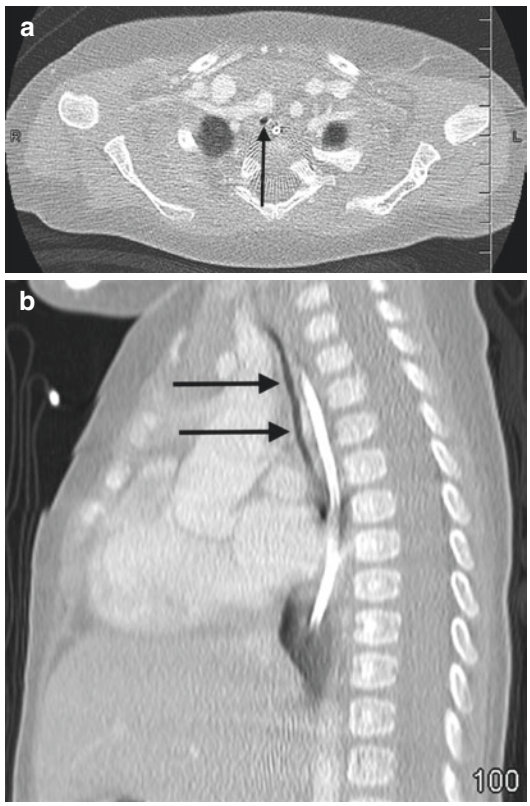

Fig. 5.13 Contrast-enhanced CT of the chest in an 11-month-old male with noisy breathing and abnormal bronchoscopy revealed diffuse narrowing of the trachea which was small in caliber and round, rather than inverse U-shaped (**a**), consistent with complete tracheal rings. Sagittal reformatted image (**b**) shows long segment narrowing of the esophagus

Vascular rings are anomalies of the aortic arch, and vascular slings are anomalies of the pulmonary arterial system which can cause compression of the upper airway and esophagus [35]. These vascular anomalies can be categorized as complete if they circumferentially surround the trachea or esophagus or incomplete if they only partially encase these structures [36]. Vascular rings and slings include a double aortic arch, right aortic arch with aberrant left subclavian artery, innominate artery compression syndrome, left-sided aortic arch with aberrant right subclavian artery, and pulmonary artery sling. A double aortic arch is the most common vascular ring and results from persistence of the right and left embryonic fourth aortic arch [37]. A right aortic arch can be associated with concomitant left subclavian artery, left ligamentum arteriosum, or Kommerell diverticulum [38]. A pulmonary artery sling is less common and occurs when the left pulmonary artery comes off the right pulmonary artery, compressing the distal trachea and right main stem bronchus [38]. A pulmonary sling is a result of failure of development of the left sixth aortic arch [37]. Children with vascular rings or slings often present with symptoms related to compression of the trachea or esophagus including difficulties swallowing solid foods, stridor, noisy breathing, or chronic cough [38]. Symptoms vary based on the degree of compression on the digestive or respiratory tract. Children may also exhibit frequent upper respiratory tract infections, wheezing, decreased exercise tolerance, apnea, or aspiration events. These symptoms usually present within the first year of life [35]. Surgical repair is the treatment of choice for children with symptomatic vascular rings and pulmonary slings, but the surgical procedure and approach varies based on the child's vascular anomaly.

Radiographic Findings

Fluoroscopy or CT can be used to determine decrease in anterior-posterior diameter of the

tracheal lumen during expiration with excellent specificity (96–100%) but poor sensitivity (23–68%) [29] (Fig. 5.14). Bronchoscopy during spontaneous respiration remains the gold standard for the evaluation of suspected tracheomalacia. If focal rather than diffuse tracheal narrowing is seen, innominate artery syndrome (Fig. 5.15) or vascular ring should be considered.

Workup of a child with a vascular ring or sling often starts with a frontal and lateral chest radiograph to identify the location of the aortic arch and narrowing of the trachea (Fig. 5.16a) [35, 39]. In children with vascular rings, a right or double aortic arch may be detected by abnormal right-sided mass effect on the trachea (the normal left aortic arch causes mild mass effect on the left side of the trachea). Additionally, the trachea may be narrowed near the aortic knob, and increased retrotracheal soft tissues can result in bowing of the trachea on the lateral view [39]. Pulmonary hypoinflation, tracheal narrowing, and a horizontal course of the left bronchus can be seen on the chest radiograph with pulmonary sling [35]. Initial imaging evaluation for possible vascular ring or sling often includes an esophagram to evaluate for the classic patterns of abnormal extrinsic mass effect on the esophagus (Fig. 5.16b, c). Posterior indentation of the esophagus on an upper GI can indicate an aberrant left subclavian artery seen with a right aortic arch, while bilateral indentations of the esophagus on an AP view are seen with a double aortic arch [39]. A single anterior indentation of the esophagus indicates the presence of a pulmonary artery sling [39]. Once a vascular ring or sling is suspected, contrast-enhanced CT of the chest is performed for surgical planning. CT allows for high spatial and temporal resolution, the ability to create vascular reconstructions, and cross-sectional imaging of the associated esophagus, trachea, and bronchi [35]. MRA can be used as an alternative to CT for the evaluation of vascular rings and slings (Fig. 5.16d). MRA eliminates radiation exposure and iodinated contrast; however, these imaging sequences are often more time-consuming and expensive, may require pediatric sedation, and can result in poorer spatial resolution [37]. Tracheobronchomalacia can often be seen on CT imaging by utilizing breath holds at full inspiration and end expiration [39].

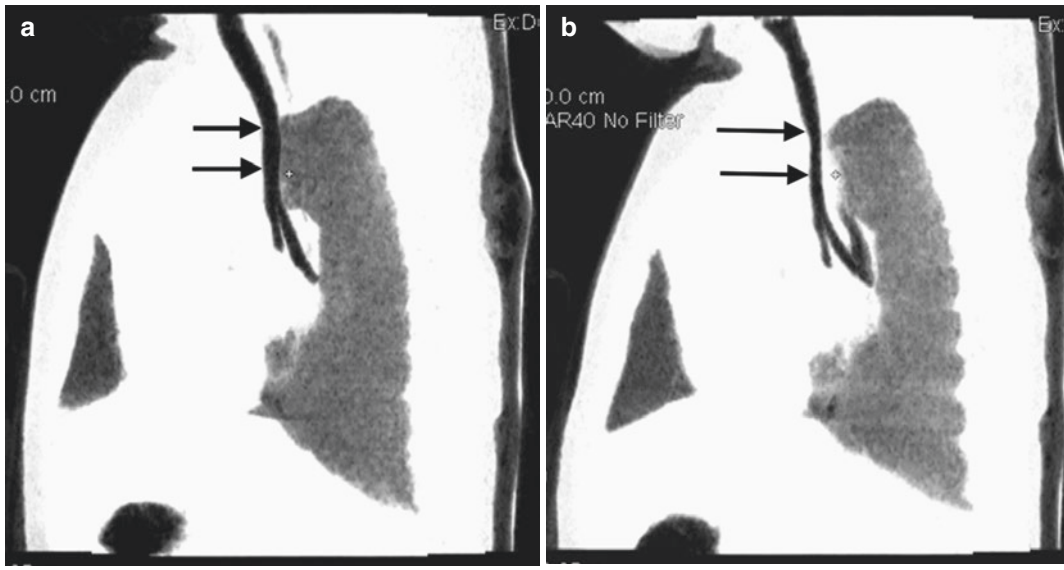

Fig. 5.14 An 11-month-old female with history of right aortic arch with aberrant left subclavian artery status post vascular ring takedown with noisy breathing. Low-dose contrast-enhanced CT scan of the chest was obtained during free breathing over the course of a respiratory cycle to evaluate for possible innominate artery compression syndrome. Maximum intensity projection reformatted images during inspiration (**a**) and expiration (**b**) demonstrate diffuse smooth narrowing of the trachea during expiration rather than focal narrowing, consistent with tracheomalacia

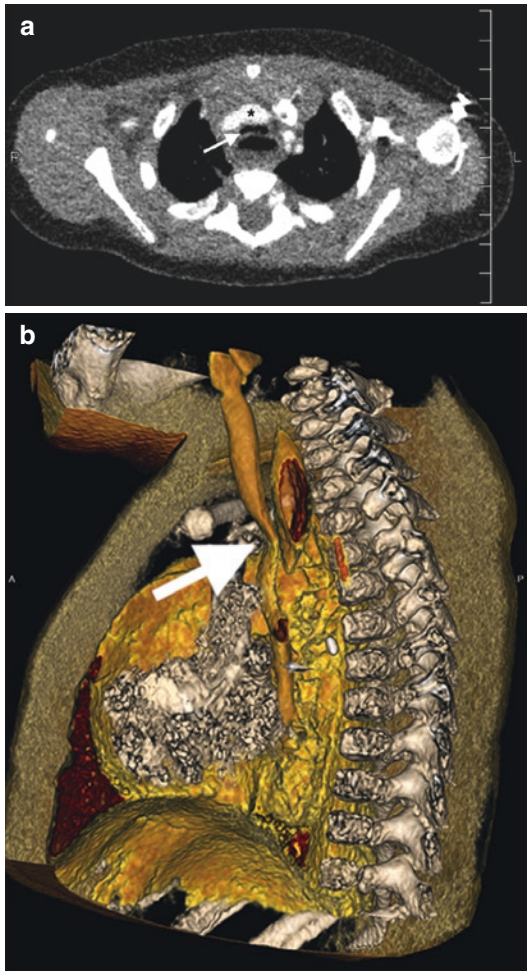

Fig. 5.15 A 4-month-old male with history of esophageal atresia with tracheoesophageal fistula s/p repair with noisy breathing. (**a**) Low-dose axial imaging through the chest performed over the course of a respiratory cycle during free breathing demonstrated focal fixed narrowing of the trachea (arrow) as the innominate artery (∗) crosses anterior to it from left to right. The dilated proximal esophagus related to prior esophageal atresia is seen posterior to the trachea. (**b**) 3D reformatted CT image of the chest in an oblique sagittal projection shows the severe focal narrowing of the trachea at the level of the innominate artery (arrow). The patient subsequently underwent aortopexy with improvement of his symptoms

Additionally, patients are recommended to undergo echocardiogram to assess for evidence of congenital heart disease or cardiac anomalies including patent ductus arteriosus (PDA), tetralogy of Fallot, or ventricular septal defect (VSD) which may occur in up to 12% of children with vascular rings [40]. Finally, children with vascular rings and slings can undergo a bronchoscopy to assess for tracheomalacia and subglottic stenosis [38].

Tracheoesophageal Fistula

Clinical Presentation and Etiology

During the 4th week of gestation, the laryngotracheal tube forms and subsequently divides the trachea and esophagus. Failure to completely bud and separate results in an esophageal atresia or tracheoesophageal fistula (TEF). There are five types of TEF. Esophageal atresia without or without TEF is commonly seen with VACTERL (vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula with esophageal atresia, renal anomalies, and limb anomalies) syndrome [41]. Infants with esophageal atresia with TEF will classically present with large amounts of oral secretions, respiratory distress, dysphagia, chronic respiratory infections, and cyanosis which is worsened with feeds. When suspected, a feeding tube is often passed and typically ends at 10–12 cm at the blind end of the proximal esophageal pouch [42]. Once identified, a TEF is managed operatively via either open thoracotomy or minimally invasive thoracoscopic approach. Postoperative complications after TEF repair include anastomotic stricture, leak, recurrent fistula, or recurrent laryngeal nerve injury [43].

Radiographic Findings

Esophageal atresia with TEF is often identified on prenatal ultrasound as early as 20 weeks' gestation as polyhydramnios with a small or absent stomach. If the diagnosis is in question, fetal MRI can be performed and is highly sensitive [41]. For a select group of children, preoperative imaging for TEF can include bronchoscopy and proximal pouch contrast study [44]. Delayed diagnosis is most common in children with Type E TEF (H type). Given that these fistulae are most commonly found between C7 and T1 [45], the lower cervical and

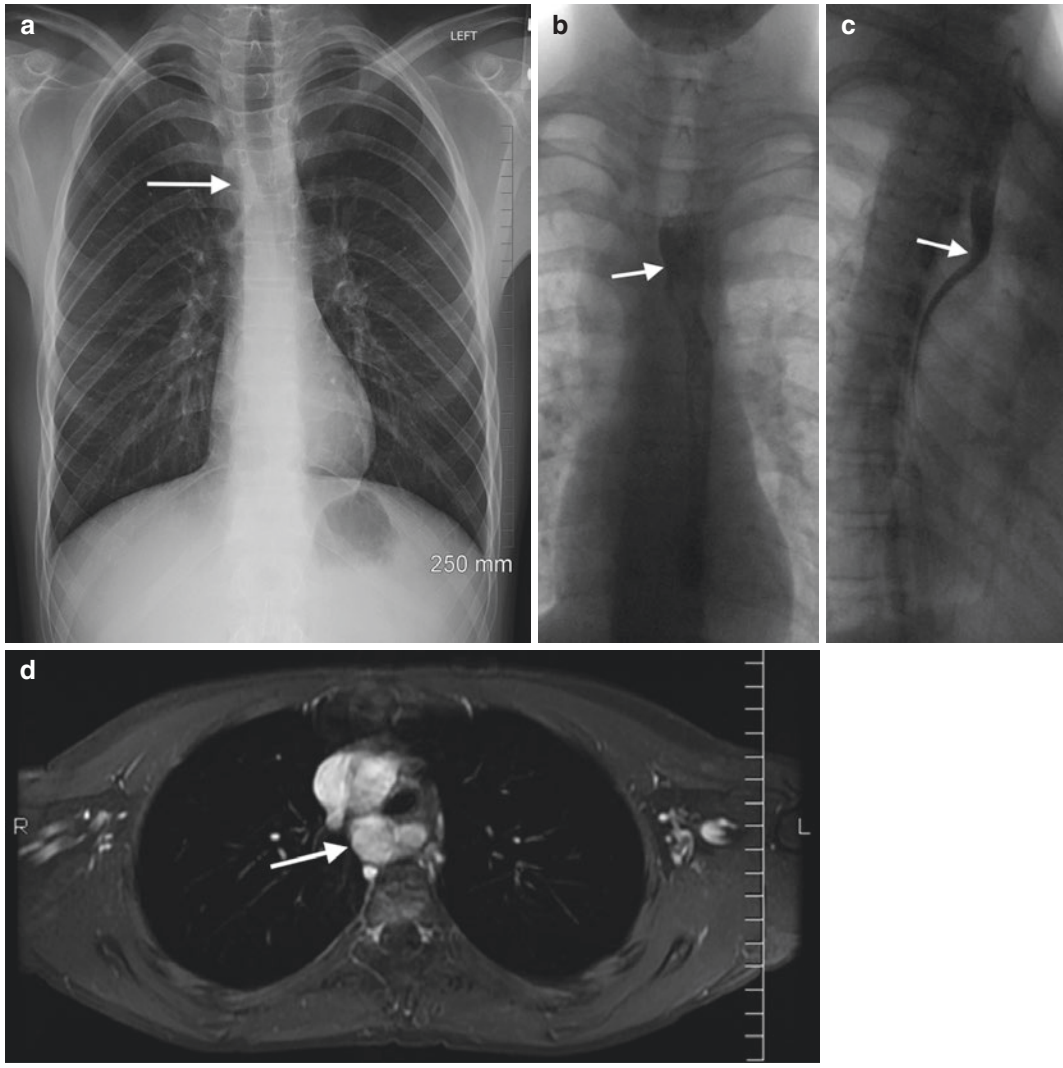

Fig. 5.16 A 14-year-old male with long-standing history of difficulty swallowing meat and pills. During an endoscopy the patient was found to have abnormal compression of his upper esophagus. An AP chest radiograph (**a**) demonstrated abnormal distal right paratracheal soft tissue (arrow) resulting in narrowing of the distal trachea suspicious for a right aortic arch. Subsequently performed esophagram showed (**b**) abnormal extrinsic mass effect along the right side of the upper cervical esophagus (arrow) as seen with a right aortic arch as well as on the posterior trachea (**c**) related to an aberrant left subclavian artery. Subsequently performed MR angiogram of the chest (**d**) confirmed the findings (arrow)

upper esophagus must be closely scrutinized during videofluoroscopic swallow studies and esophagrams. The imaging findings for aspiration from H-type TEF differ from classic oropharyngeal aspiration. With TEF, the contrast flows upward through the fistula into the trachea and then subsequently cephalad toward the glottis (Fig. 5.17). With aspiration, contrast flows downward through the larynx into the trachea. Although bronchoscopy is the most sensitive test, esophagram and/or videofluoroscopic swallow study is generally done as the first-line study in children with suspected TEF as no anesthesia is required.

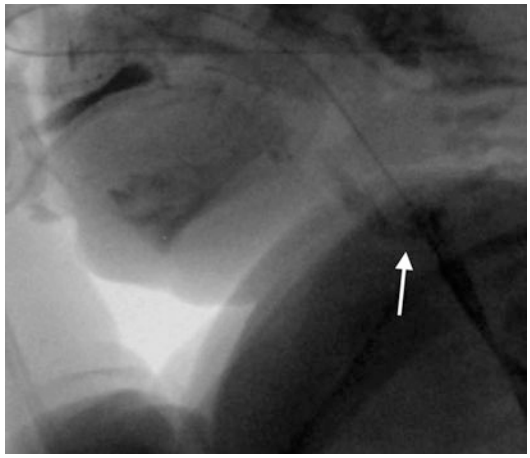

Fig. 5.17 A 6-day-old female with right pulmonary agenesis who had desaturations as well as coughing and gagging with oral feeds. Single lateral image of the neck and upper chest obtained during a videofluoroscopic swallow study demonstrated contrast extending from the esophagus near the thoracic inlet upward into the trachea (arrow) where contrast then flowed superiorly toward the glottis

Conclusion

Children presenting with either acute or chronic pathology of the upper aerodigestive tract often present with either respiratory symptoms of stridor, increased work of breathing, cough, or recurrent pneumonias or gastrointestinal symptoms of dysphagia, drooling, failure to thrive, and vomiting. For those children in acute distress, emergent airway management is imperative. For those with more indolent symptoms, imaging studies are often integral to the diagnosis of the underlying disease process. Typically, plain radiographs of the neck are the initial imaging study done to narrow the differential. These plain films are often followed by either CT and/or MR imaging or dynamic fluoroscopic studies in conjunction with bronchoscopy. Discussing clinical details with the radiologist prior to ordering radiologic studies can optimize the imaging plan in order to limit radiation dose and sedation as much as possible.

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Physiology of Voice Production

6

Matthew R. Hoffman, Maia N. Braden,
and J. Scott McMurray

> Voice is unique and exceptional among physiological phenomena.
>
> –Nobuhiko Isshiki [1]

Overview

Many critical moments in a child's early life relate to voice: the first cry on entering the world, the first laugh, and the first words. As a child grows, voice becomes an important part of identity, reflecting age, emotion, gender, and health [2]. In order to understand and appropriately manage voice disorders, an understanding of normal phonatory physiology is required. This chapter provides an overview of laryngeal anatomy and physiology of voice production.

Laryngeal Anatomy

Framework

Laryngeal structural framework includes the hyoid bone and six cartilages, three of which are paired [3] (Fig. 6.1). The largest cartilage is the thyroid cartilage, which has an angle of 90° in the adult male and 120° in the adult female, accounting for the difference in external prominence.

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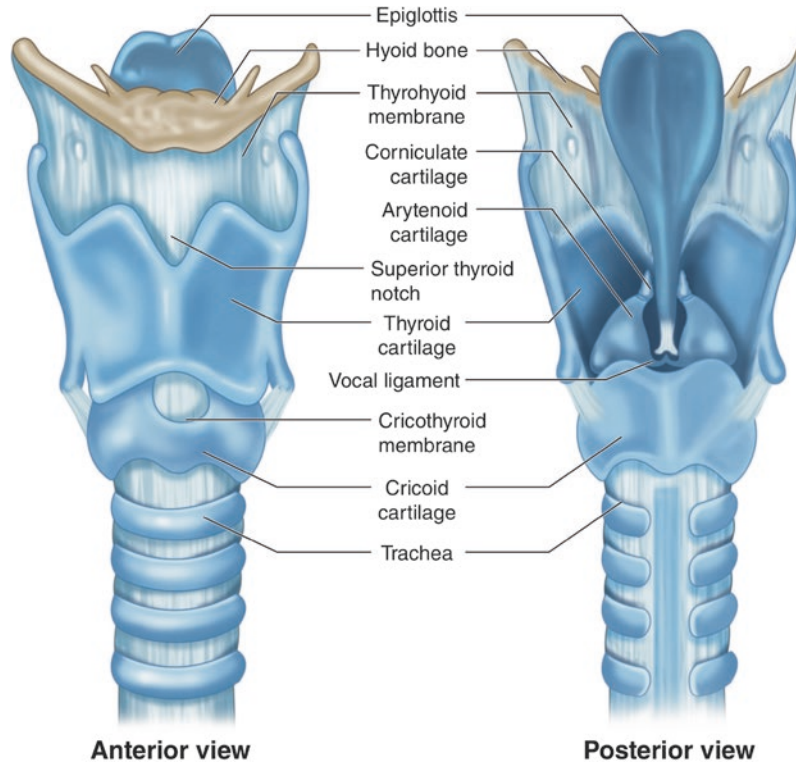

Fig. 6.1 Laryngeal cartilaginous framework

The "thyroid" comes from the Greek word for shield, *thyreos*, and it is this shield which serves as a cartilaginous barrier protecting the endolarynx. In the child, the thyroid cartilage is more pliable and obtusely angled, resulting in a less discrete prominence prior to puberty. The thyroid cartilage is connected to the hyoid bone via the thyrohyoid membrane and to the epiglottis via the thyroepiglottic ligament. The lateral superior cornua of the thyroid cartilage attach to the lateral hyoid bone via the hyothyroid ligaments. The epiglottis aids in airway protection during swallow and can also serve as a resonance chamber during phonation [4]. The cricoid cartilage is the inferior-most cartilage of the larynx and has a longer coronal extent posteriorly. "Cricoid" is derived from the Greek word for ring, *krikos*, as it is indeed the only complete ring structure in the normal airway. It is attached to the trachea via the cricotracheal ligament and has two posteriorly positioned cricoarytenoid facets, which serve as the articulatory surfaces for the paired arytenoid cartilages. The cricoarytenoid joint allows for sliding, rocking, and twisting [5]. The muscular process of the arytenoid cartilage serves as an attachment point for all intrinsic laryngeal muscles except the cricothyroid muscles, and the vocal process serves as the attachment point for the vocal ligament. The corniculate cartilages lie superior to each arytenoid cartilage, and the cuneiform cartilages are positioned anterosuperiorly to the corniculates within the aryepiglottic folds.

Two additional structures provide structural support to the larynx: the quadrangular membrane and conus elasticus. The quadrangular membrane is an elastic structure that extends anteriorly to the lateral epiglottis, posteriorly to the arytenoids, superiorly to the aryepiglottic fold, and inferiorly to the medial wall of the pyriform sinus [6]. The conus elasticus is a fibroelastic structure that originates inferiorly along the cricoid cartilage and extends superiorly to the anterior commissure and vocal processes; it forms the vocal ligament medially and is continuous with the cricothyroid membrane anteriorly [6].

Muscles and Peripheral Innervation

There are intrinsic and extrinsic muscles of the larynx. The intrinsic muscles control vocal fold position and are innervated by the recurrent laryngeal nerve (RLN) or external branch of the superior laryngeal nerve (SLN) (Fig. 6.2). Both the RLN and SLN are branches of the vagus nerve, the tenth cranial nerve. Muscles innervated by the RLN include the thyroarytenoid, lateral cricoarytenoid, posterior cricoarytenoid, and interarytenoid muscles. The interarytenoid muscles receive bilateral innervation, while the others receive unilateral innervation. The cricothyroid muscle is the only muscle innervated by the external branch of the SLN; it controls vocal fold elongation and, thus, helps dictate fundamental frequency. The thyroarytenoid muscle comprises the bulk of the vocal fold and contributes both to vocal fold adduction and control of fundamental frequency. The lateral cricoarytenoid and the oblique and transverse interarytenoid muscles also adduct the vocal folds. The posterior cricoarytenoid muscle is the sole vocal fold abductor, with contraction serving to open the glottic airway during respiration. The posterior cricoarytenoid also plays an important role during voice production, allowing the arytenoid to rock posteriorly, thus increasing tension on the vocal ligament. If posterior cricoarytenoid action is lost, the arytenoid tends to subluxate anteromedially given the lack of a posteriorly directed force to counter arytenoid movement [7].

Extrinsic muscles of the larynx include infrahyoid and suprahyoid muscles and serve to maintain or alter the position of the larynx within the neck. This can include maintaining a stable laryngeal framework so that the intrinsic laryngeal muscles can work effectively in the setting of voice performance [8]. Infrahyoid muscles include the omohyoid, sternohyoid, sternothyroid, and thyrohyoid, with general function serving to lower position of the hyoid bone. Suprahyoid muscles include the stylohyoid, geniohyoid, mylohyoid, and digastric muscles, with contraction serving to raise the hyoid bone.

Laryngeal sensory innervation is from the internal branch of the SLN above the glottis and the RLN at and below the glottis.

Vocal Folds

The true vocal fold consists of five layers (Fig. 6.3). Superficially is a layer of stratified squamous epithelium, able to withstand the stress of vocal fold vibration. Deep to the surface epithelium is the lamina propria, which consists of three layers. The superficial layer is also known as Reinke's space and consists primarily of an extracellular matrix comprised of hyaluronic acid, collagen, elastin, lipids, and carbohydrates [9, 10]; it plays a key role in vibration. The intermediate and deep layers of the lamina propria form the vocal ligament, with the intermediate layer consisting primarily of elastin and the deep layer primarily collagen. At the anterior and posterior ends of the membranous vocal fold are the macula flava, which are formed by thickening of the intermediate lamina propria [11, 12]. This provides additional stiffness which protects the vocal fold from injury during vibration [13].

Hirano described the body-cover theory of the vocal fold, with the cover consisting of the surface epithelium, superficial lamina propria, and intermediate lamina propria, and the body consisting of the deep lamina propria and thyroarytenoid muscle [14].

Superior to the true vocal folds are false vocal folds, also known as the ventricular or vestibular folds. The ventricle lies between the true and false vocal folds. The false vocal folds aid in airway protection during swallow and can also contribute to normal voice production and timbre [15–17]. The false vocal folds also play a prominent role in Tibetan and Tuvan throat singing. Adduction of the false vocal folds is due to contraction of the ventricularis muscle, innervated by the RLN [16].

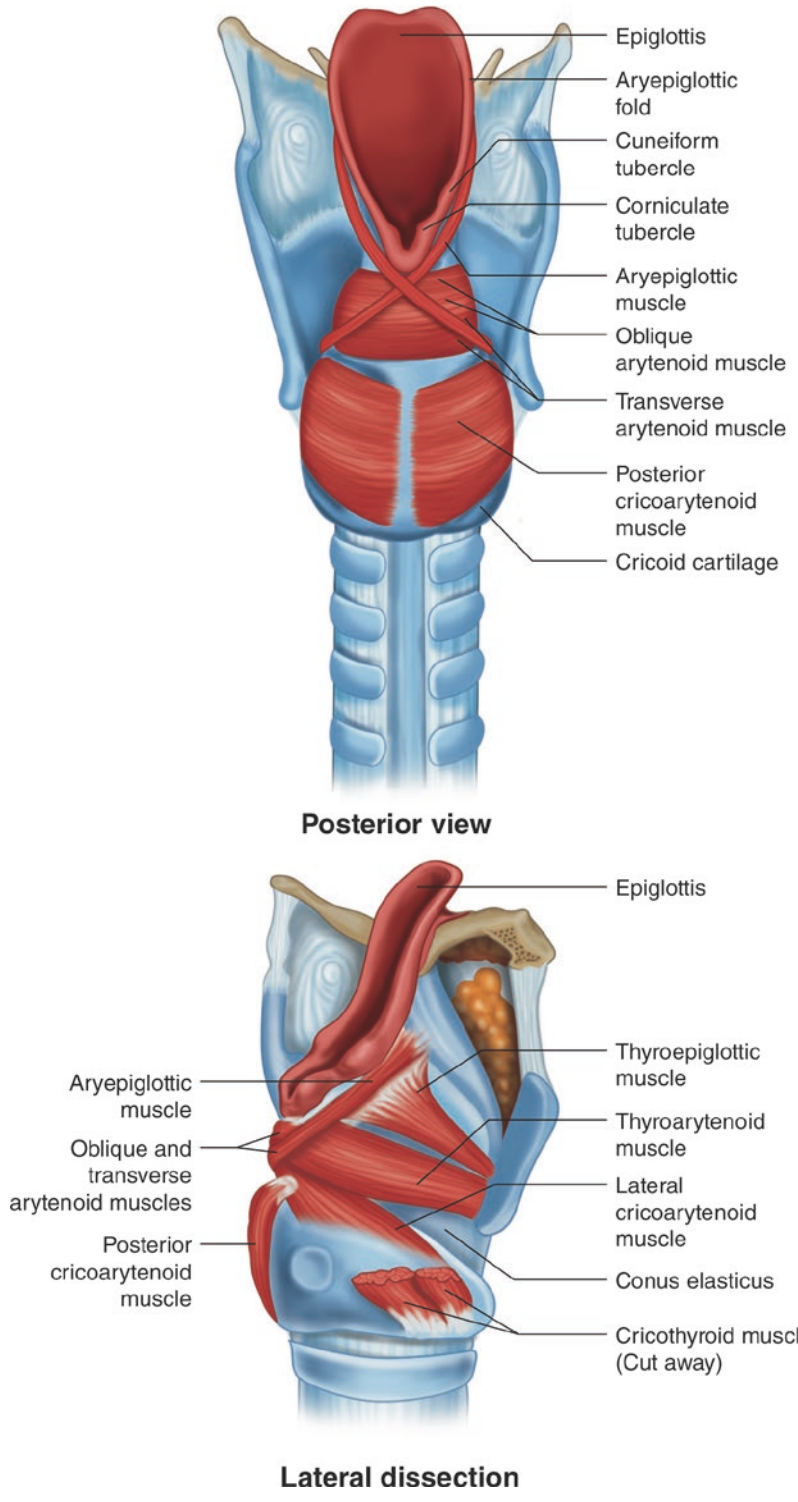

Fig. 6.2 Intrinsic muscles of the larynx

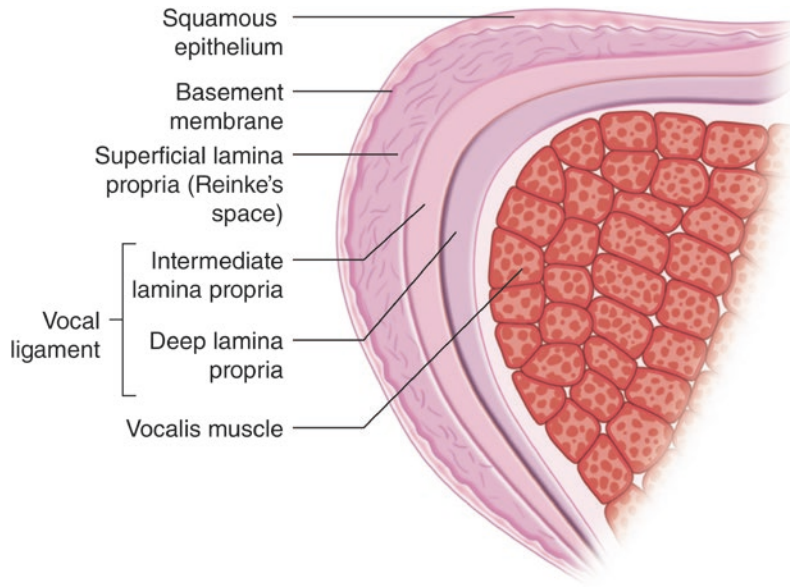

Fig. 6.3 Layered structure of the vocal fold

Blood Supply

Blood supply to the larynx is from the superior and inferior laryngeal arteries. The superior laryngeal artery arises from the superior thyroid artery, which arises from the external carotid artery. The inferior thyroid artery arises from the thyrocervical trunk, which arises from the subclavian artery. Venous drainage is via the superior and inferior laryngeal veins which arise from the thyroid veins.

Central Innervation

Nuclei for the motor neurons of the intrinsic muscles of the larynx are in the nucleus ambiguus [18]. Additional nuclei involved in control of voice production include the trigeminal motor nucleus in the pons, facial and hypoglossal nuclei in the medulla, and ventral horn of the spinal cord, with coordination accomplished via the ventrolateral parabrachial area, lateral pontine reticular formation, anterolateral and caudal medullary reticular formation, and nucleus retroambiguus [19]. Voluntary voice control requires input from the cerebral cortex, including Broca's area, the supplementary motor area, and the pre-supplementary motor area [19].

Interestingly, multiple forebrain systems appear to innervate the thyroarytenoid muscles [20]. This includes the anterior cingulate, periaqueductal gray, and ventral respiratory group, which serve as the center for vocalization in primates [19]. The second system consists of the central nucleus of the amygdala, basal ganglia, thalamus, and hypothalamus, which contribute to swallowing, vocalization in response to aversive stimuli, and emotional vocalizations such as laughing or crying [21]. The ventrolateral preoptic area of the hypothalamus provides neural input to the thyroarytenoid during sleep [20].

Changes in Anatomy with Aging

The newborn larynx sits more superiorly in the neck at the level of the C4 vertebra compared to the level of C6–7 in adults. This leads to a shorter vocal tract and altered formant frequencies. The epiglottis has a more tightly curled contour, the vocal folds are about 2.5–8 mm long [22, 23],

and the vocal fold lacks the layered architecture found in adults [24]. There is no well-defined vocal ligament, but this emerges beginning after age 1 and is observable after age 4 [23, 25]. As the child ages, cartilages also increase in size and change shape. The vocal folds develop a layered structure [24, 26]. By age 7, the depth of the superficial lamina propria is approximately that seen in the adult [27]. In terms of distribution of elastin and collagen and orientation of these fibers, the vocal fold layers do not approximate those of an adult until around age 13 [28]. Interestingly, hyaluronic acid (HA) in infant vocal folds has been found to be evenly distributed throughout, in contrast with the distribution in adult vocal folds, in which distribution of HA, elastin, and collagen vary by layer [29]. This has been hypothesized to have a protective effect against phonotraumatic damage from prolonged crying. The ratio of the membranous/cartilaginous glottis also changes with aging, from a ratio of approximately 1.5:1.0 in the newborn to a ratio of 4.0:1.0 in the adult female and 5.5:1.0 in the adult male [30, 31]. The altered ratio with effectively shortened membranous vocal fold may play a role in changing relative impact stresses and the high rate of nodules in children.

Fundamental frequency is 400–600 Hz with crying [32]. This decreases during the first few years of life in both males and females [4]. Further decreases in fundamental frequency occur from age 10 to 18 [33], with a downward shift of 12 semitones in males and 3–4 semitones in females [34]. Other pubertal changes in voice can include a reduction and then expansion in range, more difficulty negotiating register transitions in singing, increased breathiness, and transient difficulties with pitch control [35–37].

Vocal Fold Vibration

During voice production, the larynx acts as an energy transducer, transforming aerodynamic energy from the lungs into acoustic energy heard as voice (Fig. 6.4). How does this process (i.e., vocal fold vibration) occur? The myoelastic-aerodynamic theory of phonation was proposed

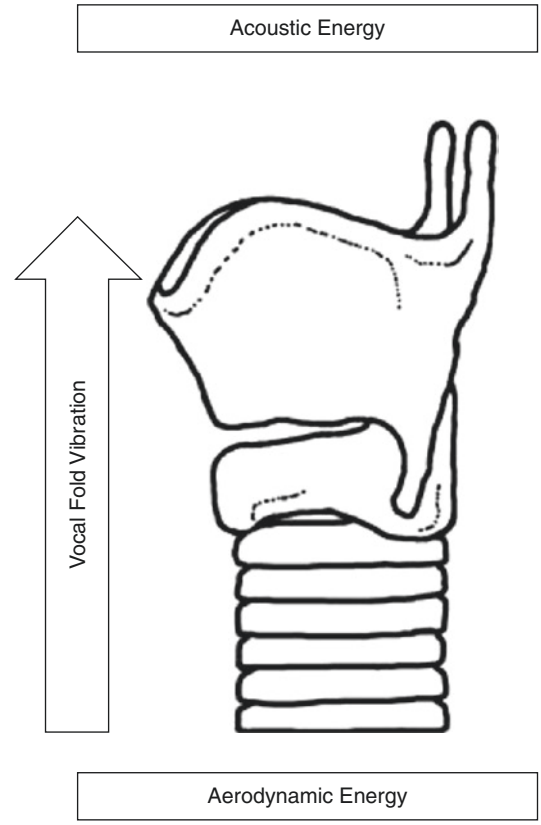

Fig. 6.4 During phonation, the larynx acts as an energy transducer, transforming aerodynamic energy into acoustic energy. Image is courtesy of Dr. Erin Devine

by Johannes Muller in 1839. Air from the lungs passes through the glottis and causes vibration of passive vocal folds, provided vocal fold tension and elasticity are adequate. This theory was modified by van den Berg in 1958 to require that the vocal folds be sufficiently approximated and that the vocal folds are driven into oscillation by Bernoulli's principle [38]. Adequate subglottal pressure can then drive the vocal folds apart, allowing air to escape through the glottis [38]. That adequate subglottal pressure is termed the phonation threshold pressure (PTP), or minimum subglottal pressure required for voice production. PTP is defined according to the following equation developed by Titze [39]:

$$\mathrm{PTP} = \frac{k_t \mathrm{Bcx}_0}{T}$$

where PTP is the phonation threshold pressure, k_t is a transglottal pressure coefficient, B is a damping constant, c is the mucosal wave velocity, x_0 is the prephonatory glottal half-width, and T is the vocal fold thickness. Lateral movement of the vocal folds occurs until elastic forces within the vocal fold and a decrease in intraglottal pressure cause the vocal folds to move medially. Decreased intraglottal pressure occurs due to Bernoulli's conservation of energy principle, which states that the pressure, or potential energy, of an incompressible fluid decreases as particle velocity, or kinetic energy, increases. Notably, the vocal folds have both upper and lower lips, and thus vibration has a vertical phase difference, with initial separation of the lower lips, followed by the upper lips. Closure then proceeds initially at the upper lips and then the lower lips.

Although the above description can explain how vocal fold oscillation is initiated, it does not completely explain how it is sustained. For that, consideration of changes in pressure above the glottis is required. When the vocal folds open, air accelerates through the glottis, and an air column of positive pressure moves superiorly into the vocal tract, which aids further in vocal fold opening. As the vocal folds close, the momentum of the air column continues, creating negative pressure which aids further in vocal fold closing. Vocal tract inertance with time-delayed changes in the supraglottal air column facilitates vocal fold opening and closing and allows for sustained oscillation [4].

Factors Affecting Vocal Fold Vibration

Subglottal Pressure

Subglottal pressure is the driving force of voice production. Two of the primary descriptors of vocal output, fundamental frequency (measured in Hz) and vocal intensity (typically measured as sound pressure level and measured in decibels (dB)), are dictated by subglottal pressure and activity of the intrinsic laryngeal muscles [40]. Increasing subglottal pressure while holding fundamental frequency constant causes an increase in intensity, and increasing subglottal pressure while holding intensity constant causes an increase in fundamental frequency [41]. The amount of change to be expected has been calculated in modeling studies and found to be 0.5–6 Hz increase in frequency per cmH_2O increase in subglottal pressure [42] or 8–9 dB increase in intensity per doubling of the phonation threshold pressure [40, 43]. Children have been found to have higher subglottic pressure than adults even when frequency and sound pressure level are controlled for [44–46].

Vocal Fold Length and Tension

Fundamental frequency is determined by mass and stiffness, which are related to vocal fold length and tension, and controlled by the relative contractions of the cricothyroid and thyroarytenoid muscles. These two muscles are innervated by different nerves (the cricothyroid by the SLN and the thyroarytenoid by the RLN), which allows for precise control of differential contraction and, thus, fundamental frequency. Stiffness is regulated by vocal fold length, proportional to vocal fold tension, and serves as the effective restoring force in vocal fold vibration [4]. Contraction of the cricothyroid muscle causes increased tension in the cover and body and consequent increase in fundamental frequency. Contraction of the thyroarytenoid muscle causes increased tension of the body which, depending on the state of cricothyroid muscle contraction, can either increase or decrease fundamental frequency [47, 48]. Isolated thyroarytenoid contraction causes a decrease in vocal fold length and cover stiffness with an increase in body stiffness, resulting in a decreased fundamental frequency. Combined contraction of the thyroarytenoid and cricothyroid muscles causes an increase in fundamental frequency, with isolated cricothyroid contraction causing the greatest increase in frequency.

Vocal Fold Contour

Vocal fold contour dictates glottic configuration which can be convergent, divergent, or rectangular [4] (Fig. 6.5). During puberty, inferomedial hypertrophy of the thyroarytenoid muscle creates a more rectangular glottis with increased medial

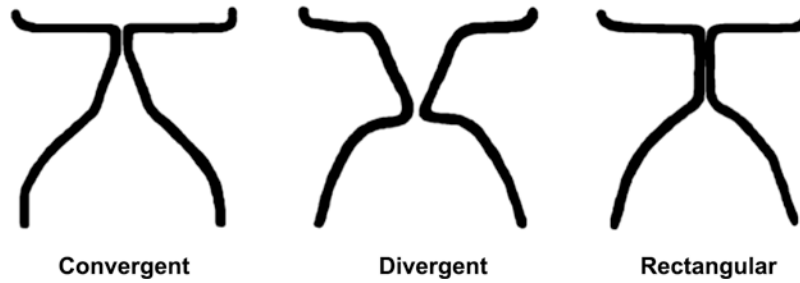

Fig. 6.5 Glottic configurations, including convergent, divergent, and rectangular configurations. Image is courtesy of Dr. Erin Devine

surface thickness, which results in increased mass and thus a lower fundamental frequency [4, 49]. As the medial surface thickness increases, so too does glottal resistance. There is an initial decrease in the phonation threshold pressure, followed by an increase as well as a decrease in glottal airflow [50]. There is also an increase in the closed quotient or portion of the vibratory cycle during which the vocal folds are in contact. This has acoustic consequences, including excitation of higher-order harmonics and a decrease in the difference between the first and second harmonics [50]. A similar change in glottal configuration can be observed as one moves from a falsetto to modal register, where contraction of the thyroarytenoid creates a more rectangular glottis and vibration in the mucosa and ligament changes to vibration in the cover and body [4].

Vocal Fold Adduction

Changes in prephonatory glottal width affect vocal aerodynamics and resultant voice quality, with increases in prephonatory glottal width leading to increased breathiness and decreased vocal efficiency. Increased prephonatory glottal width, as in vocal fold paresis or paralysis, leads to increases in phonation threshold flow and pressure [39, 51]. Flow tends to vary more than pressure with changes in glottal gap [52].

Hydration

Recommendations to improve hydration are a common tenet of vocal hygiene, although the ideal amount and methods of hydration are topics of frequent discussion. Systemic hydration keeps the vocal fold mucosa healthy, and hydration at the vocal fold surface keeps the epithelial surfaces pliable [53]. Dehydration has been shown to increase the phonation threshold pressure [54, 55] and phonation threshold flow in the excised larynx [56]. Phonation threshold pressure did not increase with dehydration in healthy adult singers, although there were increases in perceived vocal effort with desiccation challenge [57]. A systematic review of the literature on hydration and voice found a slight but not statistically significant inverse relationship between hydration interventions and phonation threshold pressure [58]. From a biomechanical perspective, the level of hydration affects vocal fold stiffness and viscosity, with increasing stiffness and viscosity in the setting of dehydration [59], which affects mobility of the vocal fold cover. The importance of maintaining adequate hydration is further emphasized through consideration of the biphasic theory for the viscoelastic behavior of the vocal fold lamina propria (Fig. 6.6) [60]. The lamina propria can be thought of as a porous permeable solid with fluid occupying the pores, and the interaction between the solid and fluid components of the lamina propria dictates the stress relaxation of the lamina propria during vocal fold vibration [60, 61]. Interstitial fluid is a key factor for stress load support, with decreased fluid content increasing the risk for tissue damage during vibration [60].

Symmetry

In the ideal larynx, the vocal folds have vibratory surfaces which are smooth (without mass lesions), slippery (hydrated), straight (without bowing), and symmetric (mass and tension). Notably, slight laryngeal asymmetry is common, particularly for the posterior larynx, cartilaginous framework, or arytenoid position during phonation [62–64]. Significant disparities, though, can

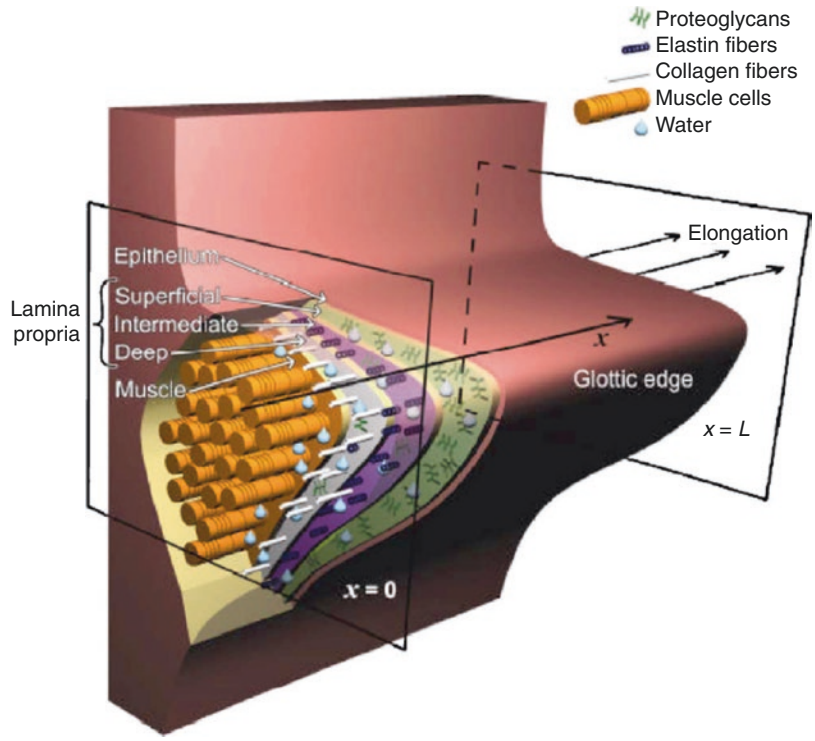

Fig. 6.6 Schematic of the biphasic nature of the vocal fold lamina propria, which includes both solid and liquid portions. Fluid is critical for stress load support during vibration. (From Zhang et al. [60], with permission)

cause significant dysphonia. Critical to maintaining normal voice production is preservation of bilateral neuromuscular activity with activation stimulated by the recurrent and superior laryngeal nerves. In the setting of asymmetric laryngeal adductor muscle activity, as may occur in a unilateral vocal fold paresis, a vibratory phase asymmetry occurs, with the normal vocal fold leading the opening phase of the glottal cycle [65], producing a chasing wave phenomenon [66]. In cases of unilateral superior laryngeal nerve paralysis, ipsilateral cricothyroid muscle contraction is impaired, resulting in a tension asymmetry between the two vocal folds. Phase shift, subharmonics, and diplophonia may occur [67].

Nonlinear Considerations

Nonlinearity is a fundamental characteristic of voice production [68]. This arises from multiple nonlinear aspects of vocal fold vibration, including nonlinear pressure-flow relationships in the glottis and the nonlinear stress-strain curve of vocal fold tissue [69]. A more thorough description of the implications of nonlinearity for clinical voice assessment is provided later in this book. It is important to recognize that chaos, or deterministic nonlinear behavior, has been observed in computer models of vocal fold vibration [70–72], excised larynx experiments [73, 74], and human subjects, including infant cries [75–77]. The importance of this becomes more evident when voice is abnormal, as in the setting of tension or mass asymmetry, or high subglottal pressure input. In such situations, the resulting acoustic signal may contain subharmonics or exhibit aperiodicity potentially heard as roughness [78] or include elements of stochastic noise heard as breathiness [79].

Formant Frequencies

It is easy to recognize the voices of those who are familiar to us. A person's voice provides a signature which is unique to them. This is due not only to their particular laryngeal anatomy and given

fundamental frequency of comfortable phonation but importantly the contributions of the vocal tract as well. The source-filter theory states that the vocal tract acts as a variable filter that shapes sound produced by the vibratory source into what is heard as speech [80]. Resonance within the vocal tract creates formant frequencies or concentrations of acoustic energy at certain harmonic frequencies relative to the fundamental frequency (F_0). Formant frequencies (abbreviated F_1, F_2, F_3, F_4) can be considered from how they are identified, as spectral peaks in the radiated sound spectrum, as well as how they originate, as resonant frequencies within the vocal tract [81]. Importantly, it can sometimes be difficult to identify the formant frequencies on a spectrogram. This is particularly true in children, as the higher fundamental frequency generates widely spaced harmonics that can undersample the spectral shape of the vocal tract transfer function, and noise secondary to glottal turbulence may further interfere with the signal [82]. Interestingly, though differences in fundamental frequency between males and females do not arise until age 10 or 11, differences in formant frequencies can occur as early as 3 or 4 years old [83, 84]. This may be related to prepubescent differences in vocal tract anatomy, including length of the oral cavity [85].

Vocal Registers

Registers are perceptually distinct regions of vocal quality that can be maintained over a range of pitch and loudness [4]. Typical speaking registers include pulse, modal, and falsetto [86]. Pulse register, also known as vocal fry, is characterized by an ability to discern gaps of silence between localized bursts of acoustic energy heard as voice, which can only occur at a fundamental frequency below approximately 70 Hz [87]. Above that frequency, the sound is interpreted as a continuous signal. The degree of thyroarytenoid contraction can control whether one phonates in the falsetto versus modal register. With increased thyroarytenoid contraction, one can transition from the falsetto register, where vibration occurs primarily in the mucosa and vocal ligament with a convergent glottis, to the modal register, where vibration occurs in the cover and the body with a rectangular glottis [4]. With rapid changes in vocal fold length, vocal tract dimensions, and body size, adolescents often report more difficulties with register transitions. While this is not in itself a voice disorder, in our experience the attempts to adjust to these changes can result in vocal difficulties.

Conclusion

Critical to the effective management of children with voice disorders is a strong understanding of how voice is normally produced. In this way, each step in the energy transduction process can be considered when voice is abnormal. Additionally, evaluating vocal fold vibration from a biomechanical perspective can help guide interpretation of videostroboscopic exams and anticipate outcomes of interventions.

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7 Embryonic and Histologic Development of the Vocal Tract

Vidisha Mohad, Hailey Hirsch, and Susan L. Thibeault

Introduction

Voice is a fundamental aspect of effective communication. The larynx seated at the intersection between the digestive and respiratory tracts is responsible for governing three major functions in humans: protection, respiration, and phonation. The primary functions are to regulate airflow during respiration and close the glottis, the space between the vocal folds signaling the entrance to the airway, during swallowing to prevent food and liquid from entering the lungs. The secondary function is phonation. A comprehensive understanding of these functional competencies is essential to the management of the myriad of laryngeal diseases impeding the functionality of this complex organ. This is especially vital in the pediatric population with congenital anomalies. The embryological basis for human laryngeal development and defects remains very poorly understood despite the fundamental role of the larynx and vocal folds in mammalian voice production. Moreover, knowledge of the molecular genetic control of laryngeal development is limited.

Defects in development of the larynx and vocal tract during embryogenesis present with life-threatening respiratory complications and widespread lifelong problems in breathing and voicing [1]. Understanding laryngeal developmental can provide precise analyses of the pathogenesis of aberrations that affect these functional competencies. Understanding the pathogenesis of congenital laryngeal defects is dependent on the improved understanding of the embryonic development of larynx and vocal folds.

Although the anatomy of the larynx and vocal folds is well known, little is known about their embryonic growth patterns. There are few studies describing embryonic development of the larynx and even fewer describing embryonic development of the vocal folds. What research has been done was completed several years ago, and more recent research conflict in terms of their definition of when various structures are visible. The study of in utero development of the human larynx and

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vocal folds dates back to 1820 by Fleischmann [2]. These historical studies of laryngeal development have improved our understanding of the basic stages and mechanisms of mammalian laryngeal development; however, they are limited in elucidating the genetic and molecular mechanisms of embryonic development. With great strides in genetic manipulation along with the high degree of similarity between mouse and human genomes, the mouse has been a powerful model system to study all aspects of laryngeal biology from development to physiology to adult diseases [3]. Further research with murine models to investigate embryonic development and various genetic defects is vital to improve understanding of the morphogenetic processes during laryngeal and vocal fold embryonic development. Thus, a comprehensive understanding of the laryngeal development and function in the vocal apparatus is crucial with significance to both basic science and biomedical research.

Human Embryonic Development

The research on human laryngeal and vocal fold development discusses development in terms of gestational weeks, gestational days, fetal size, and the Carnegie staging system (see Table 7.1 for conversion of gestational weeks, days, and fetal size to Carnegie staging as broken down by Hill, 2007 [4]). The Carnegie staging system characterizes the first 8 weeks of embryonic development based on external changes of the fetus. Prenatal development is divided into three stages, germinal (the first 2 weeks after conception), embryonic (the end of the germinal period until 2 months of development), and fetal (the remainder of in utero development). A summary of the developmental stages and key laryngeal and vocal fold developmental features are presented in Tables 7.2 and 7.3.

Table 7.1 Carnegie stage as it corresponds to gestational weeks, gestational days, and fetal diameter size as outlined by Hill

Carnegie stage	Gestational week	Gestational day (s)	Fetal diameter (mm)
9	3	19–21	1.5–2.5
10	4	22–23	2–3.5
11	4	23–26	2.5–4.5
12	4	26–30	3–5
13	5	28–32	4–6
14	5	31–35	5–7
15	5	35–38	7–9
16	6	37–42	8–11
17	6	42–44	11–14
18	7	44–48	13–17
19	7	48–51	16–18
20	8	51–53	18–22
21	8	53–54	22–24
22	8	54–56	23–28
23	8	56–60	27–31

From Hill [4], with permission

Table 7.2 Comparison of embryonic laryngeal and vocal fold development in utero in murine and humans

Week	Human	Murine	Embryonic day (E)
3	Laryngeal formation begins	Laryngeal formation begins	9.5
5	Lateral walls of the primitive laryngopharynx are almost meeting medially	Initiation of the larynx and VF with apposition of the lateral walls of the primitive laryngopharynx	10.5
5	Primitive laryngopharynx is bilaterally compressed	Fusion of lateral wall of primitive laryngopharynx	11.5
7	Establishment of blood flow to the epithelial lamina begins	Establishment of blood flow to the epithelial lamina; laryngotracheal septum is visible	13.5
8	Cartilages are formed	Epithelial lamina continues to separate; cartilages are visible	15.5
		VF separation is completed	18.5
10	VF are differentiated	VF fully separated and firmly attached to the thyroid and arytenoids	P0/birth

Table 7.3 Summary of fetal developmental key features

Stage of embryonic development	Week of development	Key features in embryonic laryngeal and vocal fold development
Embryonic period	3	First indication of laryngeal development, the median pharyngeal groove appears
	4	Signs of the separation of the esophagus from the trachea are noted
	5	Arytenoid swellings present; superior laryngeal nerve is identified
	6	Recurrent laryngeal nerve is identified
	7	Posterior cricoarytenoid, interarytenoid, cricothyroid, and lateral cricoarytenoid are identified and innervated
	8	Fusion along the posterior cricoid lamina signals the end of chondrification; arytenoids contain muscular and vocal processes; thyroid lamina almost met medially to fuse
	8	Arytenoids develop their muscular and vocal processes
Fetal period	10	True vocal folds and vestibular folds differentiate
	14	Random "flutter-like" movement at the level of the glottis present
	15	Inconsistent tongue thrusting and swallow
	16	Inconsistent tongue cupping
	17	Periodic glottic movement noted through ultrasound
	18	Inconsistent tongue protrusion
	21	Consistent tongue thrusting
	22	Consistent swallow present
	28	Consistent tongue cupping and tongue protrusion
	29	Complex swallow present

The first indication of the larynx appears during week 3 (stage 9) via the median pharyngeal groove [2, 5, 6]. During week 4 (stages 10–12), there are three key changes. The first series occur during stage 10, when the median pharyngeal groove is noted to include the laryngotracheal sulcus with developing pulmonary primordium caudally and identification of the oropharyngeal membrane and second branchial arch is noted [2, 5–10]. The second branchial arch gives rise to the hyoid bone, esophagus, laryngotracheal ridge, trachea, bronchial buds, and thyroid diverticulum by approximately week 5 (stage 15) [8]. Also during week 4 (stage 11), a single layer of the epithelium develops in the laryngotracheal sulcus [11–13], signs of separation of the esophagus from the trachea are noted, and the tracheoesophageal septum is visualized [2, 6, 10, 11]. Lastly (stage 12), motor/sensory fibers of cranial nerves (CNs) XII, V, VII, VIII, IX/X, and XI appear in that order, and the superior ganglia of CN X are identified [11, 14].

By week 5 (stages 13–15), undifferentiated mesenchyme is noted to surround the primitive glottic slit, the lateral walls of the primitive laryngopharynx are almost meeting medially, and tracheoesophageal separation occurs [2, 15–17]. Also of note, arytenoid swellings begin to develop, and the third and fourth branchial arches, which will form future laryngeal cartilages and the ansa cervicalis, are seen with conclusion of development in week 6 (stage 17) (Kallius, 1897, as cited by Zaw-Tun & Burdi) [8, 13, 17]. Week 5 is also significant for the beginning stages of development of the cricoid and arytenoid [2, 11, 16, 18] with the right and left arytenoid swellings aiding in identification of the embryonic laryngeal inlet (stage 14) [6] and full detachment of the trachea (Streeter, 1942 as cited by O'Rahilly & Boyden) [5, 7, 11]. The epithelial lamina of the larynx begins its formation (stage 14) [2, 6, 7] followed by increased cell density of the arytenoid swellings and identification of the wedge-shaped

mesenchyme anteriorly and bilateral to the epithelial lamina that will give rise to the laryngeal musculature and cartilages (stage 15) [9, 11]. The superior laryngeal nerve (SLN) is also identifiable during this stage [2, 19].

During week 6 (stages 16–17), the epiglottal swelling is identifiable [13], the hyoid has begun to develop [2], and the cricoid begins its formation from laryngeal mesodermal anlage (stage 16) [13]. While some report that the arytenoids begin their development during stage 14, Crelin [8] reports them being identifiable during stage 16, and Zaw-Tun et al. [13] report that they are taking on a conical shape. Week 6 is also significant for the epithelial lamina consisting of two closely fused cell layers [20] and identification of the recurrent laryngeal nerve (RLN) [19].

During week 7 (stages 18–19), Lisser [21] noted that the thyroid and epiglottis have begun to develop (stage 18); however, research has also suggested that chondrification has initiated for the hyoid bone and thyroid lamina at this time [2, 6, 16]. The posterior cricoarytenoid (PCA), interarytenoid (IA), cricothyroid (CT), and lateral cricoarytenoid (LCA) muscles are identified [12, 16]. The cartilages are more adult-like in shape [12] with the epiglottis appearing concave (stage 19) [6]. Both the SLN and RLN are innervating their respective muscles with the CT, PCA, and IA being isolated, and the LCA is reliably differentiated from the thyroarytenoid (TA) [12, 16]. Also of note, the epiglottis is concave in shape, and the process of blood flow recanalization begins and is completed during week 8 (stage 23) with the glottis [6, 17].

Week 8 (stages 20–23) is significant for fusion along the posterior cricoid lamina and signals the end of chondrification [14]. The PCA is noted to be in its adult position [16], and the arytenoids have developed their muscular and vocal processes [13]. Toward the end of week 8, the ventricle is formed [8], and all intrinsic muscles are clearly recognizable (stage 22) [12].

By the end of week 8, the last of the structural changes of the larynx occur. The hyoid bone parallels the anterior border of the thyroid cartilage [22, 23], and the two thyroid laminas have almost met medially to fuse [13]. The fetus performs reflexive actions, but no sensory fibers extend to the epithelium in most areas of the pharynx and larynx [10]. An adult pattern of motor innervation is present [10] with the cricopharyngeal muscle identified at the posteroinferior margin of the cricoid [18]. The vocalis muscle begins differentiation with muscle fibers extending toward the vocal ligament [18].

For the remainder of in utero development, few changes to the structure of the larynx occur, but laryngeal function becomes established. During week 8, the vocal folds begin their development and are observed to be a slit-like opening in the larynx [8]. During week 9, the ventral borders of the thyroid lamina begin to meet with fusion completed by week 12 [6, 13]. The true and false vocal folds are differentiated around the laryngeal ventricles between weeks 10 and 12 [13, 19]. During week 11, random "flutter-like" movement of the glottis is noted through ultrasound, becoming more consistent by week 17 [24]. During week 15, the epiglottis has descended to the level of the thyroid [25]. Between weeks 17 and 20, the epiglottis is noted to contain fibroelastic cartilage [6]. During week 21, the epiglottis is in near apposition to the uvula [26]. The free borders of the VF contain one to two cell layers of the nonkeratinized squamous epithelium [27]. The cuneiforms appear during week 26 [28]. The epiglottis takes on its omega shape between weeks 29 and 32 [6]. The membranous portion of the vocal folds contains stratified squamous epithelium and is differentiated from the cartilaginous portion by week 30 and distinct from the cartilaginous portion of the vocal folds by

38 weeks [22] with glottic movement decreasing leading up to birth [20].

Human Embryonic Feeding and Swallowing

Swallowing function is the first motor response to develop and can be seen as early as week 11, coinciding with taste bud development as well as the jaw-opening reflex and beginning signs of tongue activity [29, 30]. During week 15, anti-regurgitation reflex mechanisms appear [31] as well as pre-feeding skill development of inconsistent tongue thrust and non-nutritive suck/swallow [24, 29]. Fetuses have been noted to swallow approximately 18–50 ml/kg of amniotic fluid daily by week 16 [32, 33] as well as demonstrate inconsistent tongue cupping [24]. During week 18, the fetus has inconsistent tongue protrusion with sucking movements identified between weeks 18 and 20 [24, 30]. Myelination of CNs III, IV, VI, VII, IX, and XII occurs with the appearance of jaw-opening/jaw-closing movements, anterior tongue movement, and suckling between weeks 18 and 24 [29]. Through color Doppler imaging, fetuses have been identified to have an uncoordinated swallowing and regurgitation phenomena from weeks 19 to 28 [31]. During week 21, tongue thrusting becomes more consistent with a more consistent swallow developing by week 22, which is also when substantial weight gain of the fetus is appreciated [24, 29]. Fetal facial responses to bitter tastes have been noted between weeks 26 and 28 [34]. Consistent tongue cupping and tongue protrusion have been appreciated during week 28 [24]. Coordination of the swallow with a simultaneous decrease in frequency of regurgitation is noted during week 29 [31]. By weeks 35–38, the fetal nervous system is able to sufficiently carry out integrative functions, such as nipple feeding, with near-term fetuses swallowing amniotic fluid at a rate of 500–1000 ml/day [29, 33].

Human Postnatal Development

At birth, the thyroid cartilage and hyoid bone are attached to one another. Over time, as the larynx descends, the two become separated. At birth, the larynx sits around cervical vertebrae C1–C4 to promote the suck-swallow-breathe phenomena seen in young infants [26, 35, 36]. At 4–6 months of age, infants begin to transition to use of oral respiration coinciding with changes to the neuromuscular control of the larynx and pharynx [26]. At 2 years of age, the larynx is situated between C2 and C5 [26], and by age 5, the larynx sits around C5 [35, 36]. This pattern of descent continues until it reaches its final position at C7 between the ages of 15 and 20 [31, 35]. The cartilages also begin to ossify as one ages with the hyoid beginning at age 2 [36]. Ossification does not begin again until one reaches their 20s when the thyroid and cricoid cartilages begin to ossify [36]. The ossification process is complete by the age of 65, apart from the cuneiform and corniculate cartilages [36].

Changes to the layered structure of the vocal folds occur postnatally. At birth, the vocal folds are a hypercellular monolayer at the level of the lamina propria with single cells randomly distributed and of various shapes with traces of elastin [37]. They are 2.5–3.0 mm in length [38]. By about 2 months of age, signs of cellular differentiation and progression to a bilaminar structure can be seen [37]. There is a hypocellular superficial layer with a deeper layer containing plumper and less spindly shaped cells. By 11 months of age, a three-layered structure can be seen developing based on the various cell

densities [37]. There continues to be a hypocellular layer beneath the epithelial cover, the medial layer that is more hypercellular, and the deepest layer that is just superficial to the vocalis and is also hypocellular [37]. A three-layered structure is clearly defined by the age of 7, with identifiable distinct regions of cell density [37]. The superficial layer continues to remain hypocellular, the medial layer is denser with an increase in cellularity and a region with increased amounts of collagen and elastin, and the deepest layer becomes less cellular [37]. At age 10 years, the vocal folds lengthen from approximately 6–8 mm to about 8.5–12 mm in females and 14.5–18 mm in males. By ages 11 and 12 years, we can see the classic pattern of the lamina propria, containing a hypocellular superficial layer, elastin prominent intermediate layer, and collagen prominent deep layer [37].

The timing of full maturation of the layered structure of the vocal folds coincides with when humans reach puberty. In males, this is perceptually noted as a lower-pitched voice. Research has shown that this is due to the increase in testosterone which thickens the laryngeal cartilages and vocal folds [38]. This is the first of many differences between male and female laryngeal anatomy. In addition to pitch differences, men are noted to have a larger thyroid lamina, an acute thyroid angle (creating the large "Adam's apple"), and larger glottal space [38]. Women on the other hand are noted to have a larger posterior cartilaginous space which has been hypothesized to contribute to how women use their voices culturally giving the perception of a breathy voice [38].

Murine Laryngeal and Vocal Fold Development

Numerous studies have documented the basic steps in the gross anatomic embryonic development of the larynx and vocal folds in humans [13, 17, 37]; however, they have not elucidated the cellular and molecular basis of development. Mice is an established model to study human laryngeal development [39]. According to recent findings in murine laryngeal development, the glottis develops in a region known as the primitive laryngopharynx, which develops from the foregut endoderm. Vocal fold morphogenesis starts at embryonic (E) day 10.5, ten days after the female mouse is impregnated. Throughout murine development, this region undergoes several distinct changes in development involving key morphogenetic events. These developmental events include (a) establishment of the larynx and vocal folds at E10.5 due to apposition of the lateral walls of the laryngopharynx; (b) epithelial lamina fusion (E11.5); (c) epithelial lamina recanalization and separation of the vocal folds; (d) development of laryngeal cartilages and muscles and stratification of the vocal fold epithelium (E13.5–E18.5); and finally (e) maturation of the vocal fold epithelium and lamina propria during postnatal stages [40] (Fig. 7.1). Between E16.5 and E18.5, vocal fold separation is completed. At the same time, the laryngeal cavity lengthens both anteriorly and posteriorly to accommodate vocal fold growth [40]. During postnatal stages, the vocal folds are fully separated and firmly attached to the thyroid cartilage (ventrally) and arytenoid cartilages (dorsally). Epithelial cells lining the vocal folds further stratify from the original two layers at postnatal (P) day 0 to three or four layers in the adult. After birth, the murine larynx further elongates, maturation of vocal fold epithelium occurs, and epithelial cells continue their process of stratification and evolve from a bilayer to a three/four layer in the adult mouse [35] (Fig. 7.2). As in humans, the structures continue to mature; however, the rate at which they do so differs. In humans, this process takes approximately twelve years to fully mature, whereas in wild-type mice, it takes about 6 weeks to fully mature, coinciding with sexual maturity necessary for mating.

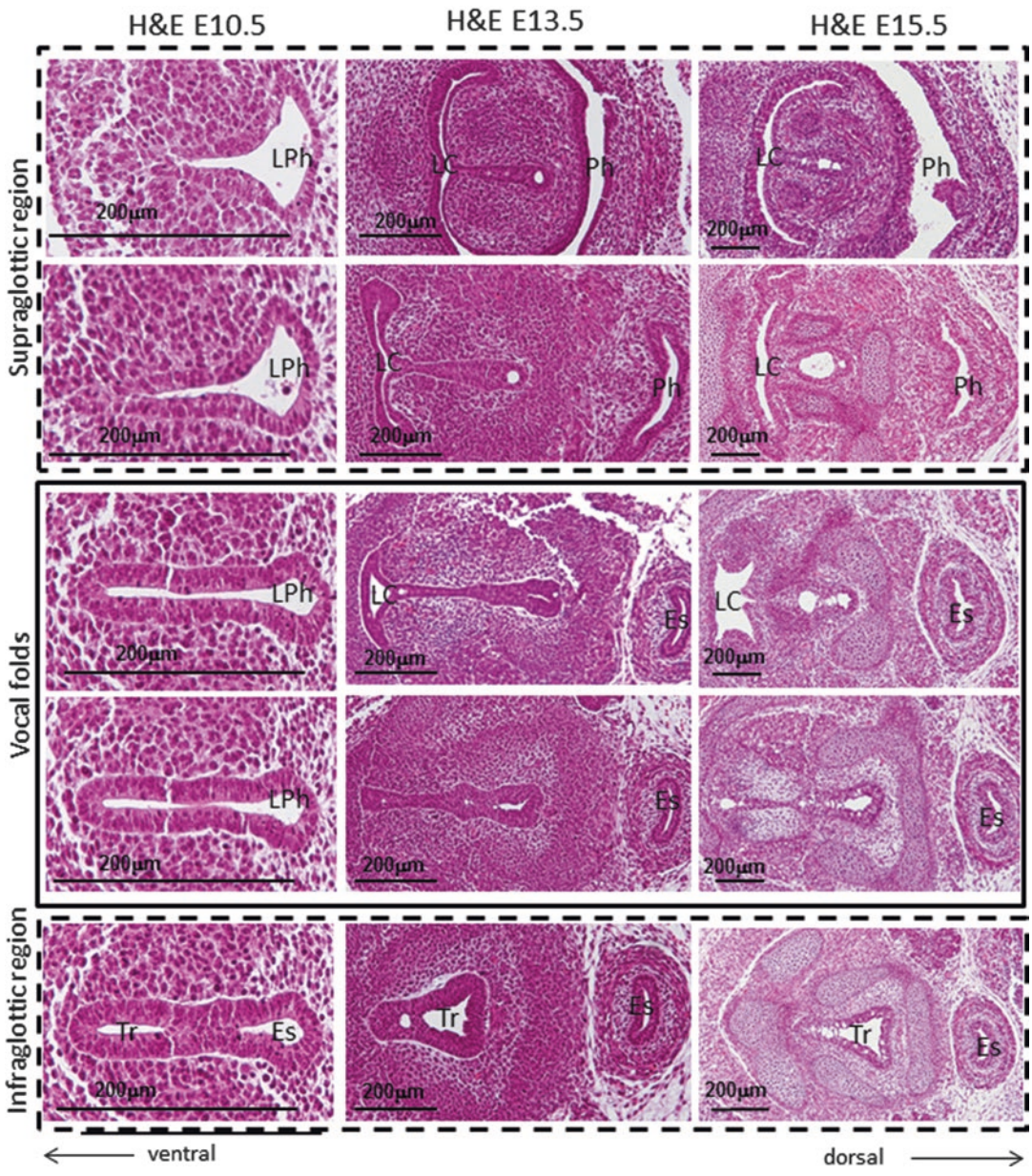

Fig. 7.1 A model for conversion and stratification of VF epithelial cells. (From Lungova et al. [40], with permission)

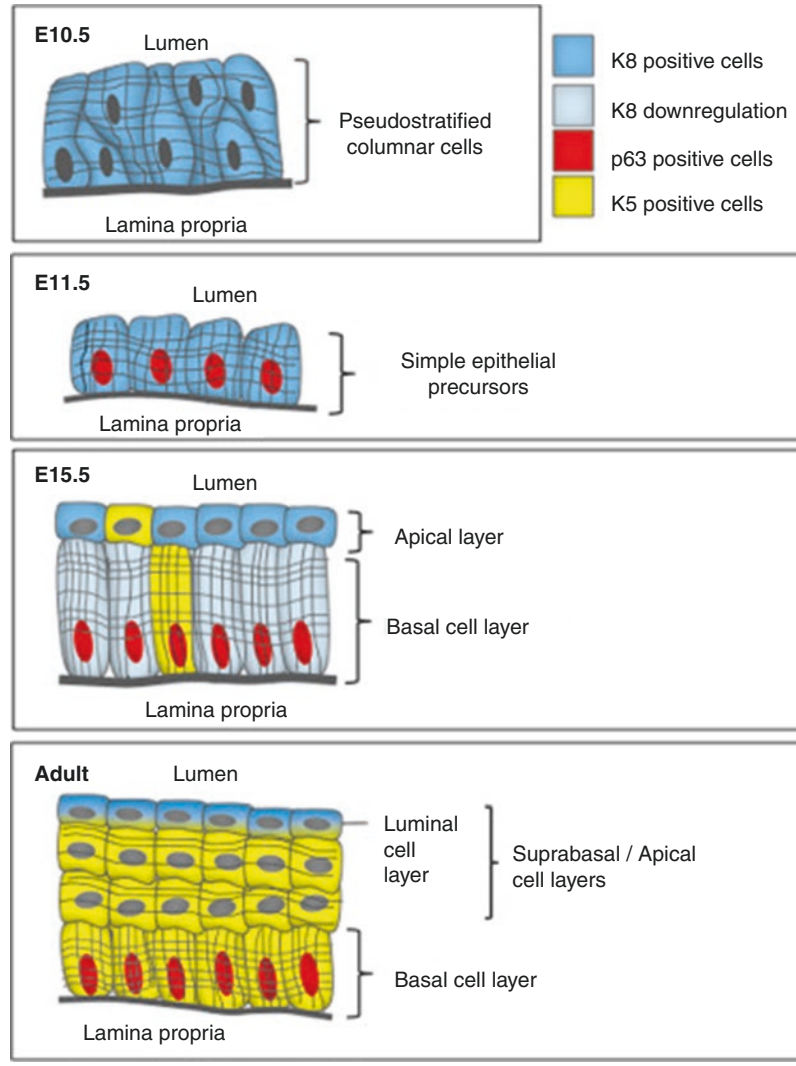

Fig. 7.2 Hematoxylin and eosin staining illustrating morphology of the laryngeal cavity in the supraglottic region, at the level of VF, and in the infraglottic region. (From Lungova et al. [40], with permission)

Conclusion

The field of embryonic development has captivated scientists for decades. Our understanding of laryngeal development has grown dramatically in recent years. This chapter explores embryonic development of the larynx and vocal folds in both human and animal models. Our concept of laryngeal embryonic development and the causes of laryngeal defects is evolving as pathways and genes required for normal development and function are being discovered. As in other organs, this improved understanding will unquestionably have an impact on human health. A more careful consideration of current animal models could further improve our understanding of the molecular processes involved in laryngeal morphogenesis. Thus, the generation of new genetic models that reproducibly create specific laryngeal developmental defects is imperative. Further investigations in this area will reveal the complex process of laryngeal morphogenesis and allow us to develop therapies through guided efforts in this burgeoning field of embryonic development. Eventually, this information may be able to help pediatric patients that suffer from congenital abnormalities.

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Nonlinear Acoustic Analysis of Voice Production

8

Hayley H. Raj, Austin J. Scholp, and Jack J. Jiang

Introduction

Voice production involves motion of the vocal folds, which act as a pair of coupled oscillators [1]. Specifically, the vocal folds and glottal airstream form a mechanical system, where energy from the airstream can be imparted to the vocal fold tissue; with enough energy, the vocal folds will begin to self-oscillate, and columns of air will pass through the glottis, creating phonation [2]. The signal produced by this airflow is then filtered by the vocal tract and heard as voice [3]. Supraglottic structures such as the supraglottic larynx, lips, tongue, palate, pharynx, and nasal cavity then act as resonators to produce the sound that is heard as voice [4]. Small changes in supraglottic structures can affect voice quality. The voice signal produced through vocal fold oscillation is modulated by other sources of internal motion such as respiration, heartbeat, action potentials, and air turbulence, as well as sub- and supraglottal anatomical structures [1].

Evaluating the quality of voice is an important aspect of vocal health assessment as it is a simple and noninvasive method for judging the outcomes of surgical procedures and following someone's progress over the course of therapy. Two main options for analyzing a voice are through perceptual methods and computational signal processing.

Perceptual Analysis Versus Signal Processing

Perceptual Analysis

Perceptual voice analysis is an essential aspect of vocal assessment that involves a trained listener that rates a person's voice based on specific factors such as overall severity, breathiness, and roughness [5]. A major benefit of this type of analysis is that it is cost-effective and noninvasive. However, as everything is decided by the listener's perception of the voice, it is subjective and depends on the listener's experience and the criteria they are using [6, 7]. In order to combat this, the Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) was developed as a way to standardize auditory-perceptual assessment [5, 8]. Another benefit of perceptual assessment methods is that they are able to analyze

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continuous speech along with sustained vowel sounds. Continuous speech is favorable because it includes important features such as rate, dialect, articulation, rapid voice onset and termination, and variations in fundamental frequency—all of which may contribute to perceptions of voice quality [9].

Signal Processing

Computerized signal processing involves the analysis of recorded acoustic signals. Several parameters exist to measure different aspects of the signal such as periodicity, aperiodicity, chaos, and noise. The goal of these methods is to provide an objective acoustic assessment of the voice. Analyzing the acoustic signal with signal processing methods allows researchers to detect trends that cannot be detected perceptually and may provide insight into the mechanism of voice production. Both perturbation and nonlinear dynamic methods are used to quantitatively describe normal and disordered voices, stemming from a variety of laryngeal pathologies [10].

Linear Versus Nonlinear Signals

A key aspect of acoustic signal processing is the linearity of the signal and its parameters. A linear parameter, loosely defined, is one that can have multiple inputs that can be summed together to obtain an output. For example, if you throw a tennis ball forward at 20 miles per hour while riding a bike going 10 miles per hour, you can calculate that the ball's initial velocity in relation to the ground would be 30 miles per hour by adding the two velocities. Thus, in this case, the velocity is a linear parameter. In contrast, the rate of deceleration is a nonlinear parameter, depending on a variety of variables including the air moisture, wind speed, ball texture, and ball velocity. All of these parameters cannot be simply summed together to describe how the ball slows down. Thus, they must be described in a nonlinear relationship [11].

In voice production, normal speech often produces a signal that is nearly periodic; this means that while there are slight deviations, one segment of the signal is approximately the same as other segments that occur on a specified interval. When signals are nearly periodic, cyclic parameters such as amplitude, period, and frequency can be tracked [1]. These are all parameters that can be considered linear. While an acoustic signal itself is nonlinear, when it is nearly periodic, it can be broken down into multiple sinusoidal waves added together allowing linear parameters such as frequency and amplitude to be calculated. This can be visualized in Fig. 8.1.

There are also aspects of voice production that are nonlinear, including the subglottal pressure-airflow relationship, vocal fold collisions, and stress-strain characteristics of laryngeal tissue [12]. This means that under certain conditions, such as high subglottal pressure and asymmetric tension or mass, vocal fold vibration may become aperiodic, producing chaotic acoustic signals [13]. Chaos, in this context, is a phenomenon that describes seemingly random behavior that is both nonlinear and deterministic. This is in contrast to white noise that is stochastic [12] (Fig. 8.1).

Voice Signal Types

To facilitate analysis, voice samples can be further categorized into four signal types. Titze first created a voice classification scheme by defining three types of acoustic vocal signals [1]. This scheme recognizes the process of bifurcation, in which the behavior of a dynamic system changes. Type 1 signals are nearly periodic with few subharmonics. Type 2 signals exhibit bifurcations or subharmonic and modulating frequencies, with a varying fundamental frequency. Type 3 signals are aperiodic [1]. Sprecher et al. [14] went on to modify the voice typing scheme to include a fourth voice type. While type 4 signals are also aperiodic, they are distinguished from type 3 signals by the presence of stochastic noise. This distinction is

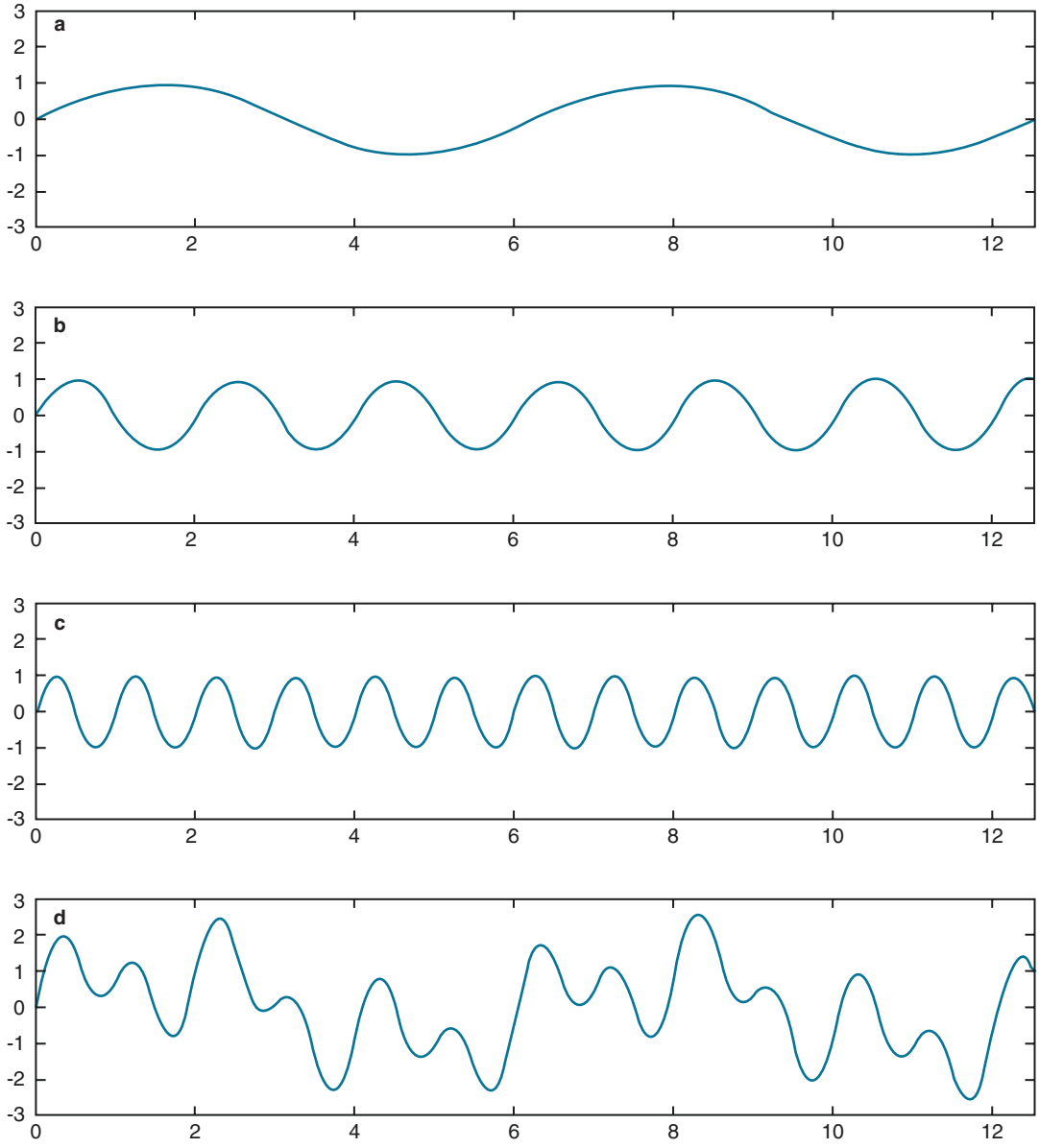

Fig. 8.1 Visual representation of three sine waves of different frequency being added to create a complex waveform

important in acoustic analysis because, while type 3 signals can be described in a finite number of dimensions, type 4 signals are considered high-dimensional. The presence of stochastic noise in a signal is interpreted clinically as breathiness. The four voice types can be qualitatively assessed through spectrogram analysis (Fig. 8.2).

Parameters

GRBAS and CAPE-V

GRBAS is an auditory-perceptual metric that is used to subjectively assess voice based on five characteristics: grade, roughness, breathiness, asthenia, and

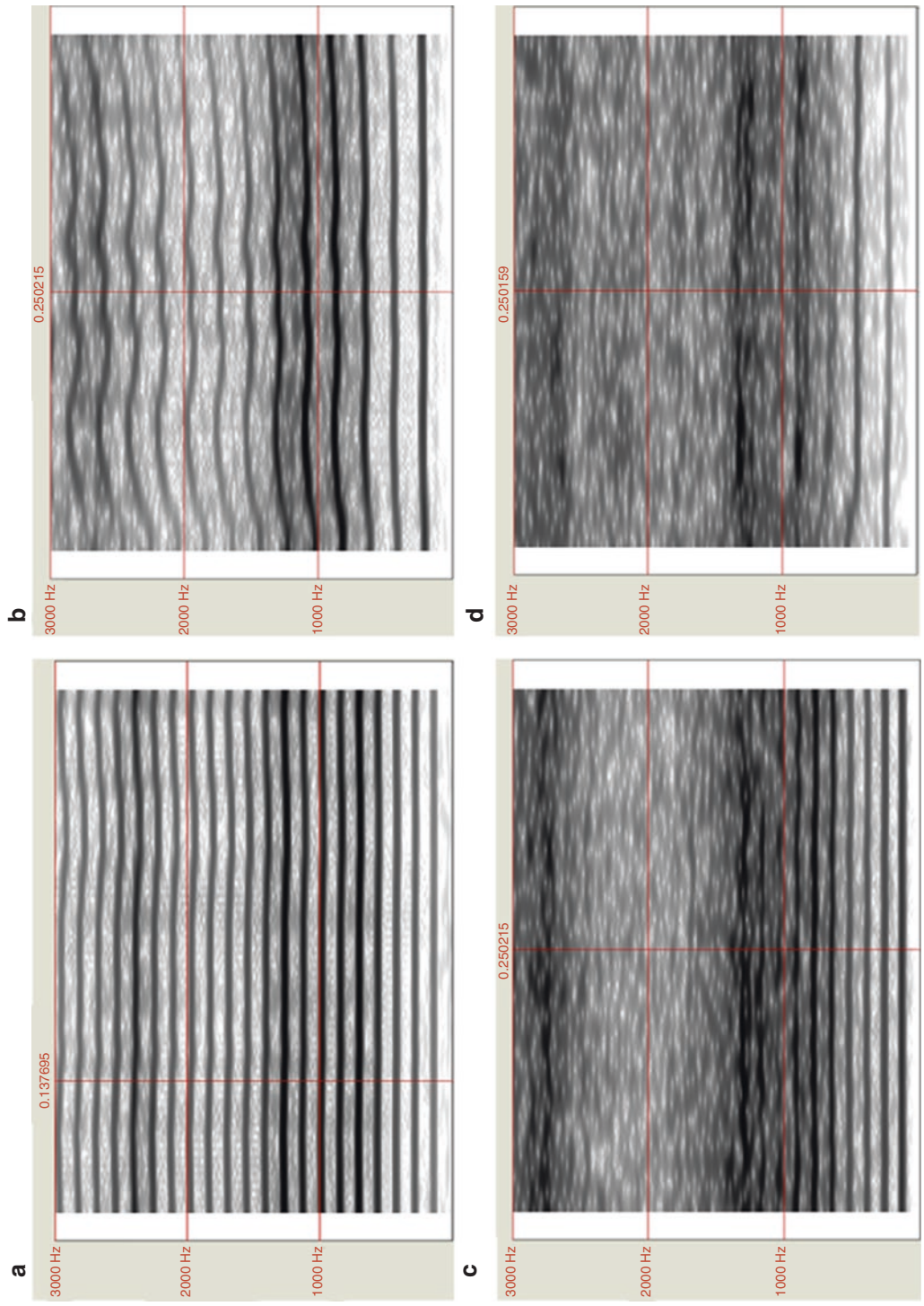

Fig. 8.2 Four spectrograms representative of the four voice types. (**a**) Type 1 voice with minimal subharmonics. (**b**) Type 2 voice with predominately periodic waves with subharmonics. (**c**) Type 3 voice with aperiodic waves and subharmonics. (**d**) Type 4 voice with no discernable periodicity and random noise

strain. Grade assesses hoarseness and abnormality in the voice. Perceived roughness represents the degree of irregular vocal fold vibration. Breathiness assesses glottic air leakage [15, 16]. Asthenia measures perceived weakness in the voice. Strain is an assessment of perceived vocal hyperfunction. The five components of the GRBAS system are rated individually on a four-point scale, where zero corresponds to normal phonation and three corresponds to severely disordered phonation [17].

The Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) is another psychoacoustic metric that allows clinicians to assess voice based on overall severity, roughness, breathiness, strain, pitch, and loudness. This system uses a 100-mm-long visual analog scale, where the far left represents normal vocalization and the far right represents severely disordered voice [8, 18]. Both GRBAS and CAPE-V require a trained rater, typically a speech-language pathologist, to make judgements on a subject's voice. Thus, as mentioned previously, these metrics are subject to the rater's level of experience.

Both GRBAS and CAPE-V have been used with children to successfully distinguish between healthy and disordered voices and evaluate the outcomes of surgical procedures and voice therapy [19, 20, 21, 22, 23, 24]. The reliability of these measures has also been investigated. In a study of 50 children aged 4–20, the CAPE-V metric was used to assess dysphonia after laryngotracheal reconstruction. Seventeen of the samples were then rerated at a later time. Inter-rater reliability was high for perceptions of breathiness, roughness, pitch, and overall severity with intraclass correlation coefficients (ICC) ranging from 67% to 71%. Perceptions of loudness were less reliable (ICC = 57%). Except for strain, intra-rater reliability was strong for all parameters, with the ICC ranging from 63% to 93% [25]. Other studies in adults and children have also found that strain, measured using CAPE-V or GRBAS, is less reliable [26, 27, 28].

Fundamental Frequency

Fundamental frequency (F_o) is the lowest frequency of a periodic or nearly periodic signal. Within a complex periodic waveform, there exists a period (duration of a single cycle of a periodic signal) that is the smallest overall. This is the fundamental period T_o. F_o is then defined as $1/T_o$ [1]. F_o reflects the frequency of vocal fold vibration; qualitatively, changes in fundamental frequency will result in a change of pitch. This is calculated through a Fourier transform, which is a decomposition of the time-domain signal, in this case an acoustic recording, into its frequency components. Figure 8.1 depicts the different waves that can make up a signal. The frequency of each wave can be plotted along with their relative amplitudes. The lowest of these frequencies is taken as F_0.

In a study of 218 healthy children aged 4–17, fundamental frequency was measured from recordings of four CAPE-V sentences. For three of the four sentences, fundamental frequency was found to decrease significantly more rapidly during ages 11–14 for boys, compared to ages 4–11 and 14–17. For girls, fundamental frequency linearly decreased with age. There was no critical age at which frequency dropped more significantly [29].

Jitter and Shimmer

Jitter and shimmer are parameters that track perturbation in the voice. Specifically, jitter is a measure of the change in fundamental frequency from cycle to cycle, and shimmer measures cycle-to-cycle change in amplitude of the signal [1]. Jitter and shimmer have been used extensively in acoustic analysis; however, these methods are less reliable in the analysis of aperiodic voice signals [12, 30, 31, 32].

Signal-to-Noise and Harmonic-to-Noise Ratios

Signal-to-noise ratio (SNR) quantifies how dominant the voice signal is over random noise. Harmonic-to-noise ratio (HNR) is similar to SNR in that it quantifies the dominance of periodic (harmonic) signal elements over noise [33];

however, noise in this case refers to random noise, aperiodic signal elements, and perturbations such as jitter and shimmer [34]. This parameter is helpful for assessing vocal characteristics such as breathiness, which results from turbulent airflow [33]. Both SNR and HNR have been used to successfully analyze healthy and disordered voices in children [35, 33, 36, 37].

Perceptual and signal processing techniques can also be used in combination to provide a comprehensive acoustic analysis of voice. In a study of 39 children aged 7–14 with a diagnosis of bilateral vocal nodules, acoustic analyses were performed to assess progression during voice therapy. Therapy lasted for 8 weeks and consisted of lessons on vocal hygiene and voice abuse reduction, breathing and phonation coordination, and laryngeal massage. Jitter and shimmer improved most significantly post-therapy. HNR was also lower after therapy compared to baseline, but F_o did not change. Perceptual ratings according to the GRBAS system showed that grade, roughness, breathiness, and strain all improved [36].

Correlation Dimension, Lyapunov Exponents, and Kolmogorov Entropy

Nonlinear dynamic methods are useful for analyzing normal and disordered voices, and they provide the advantage of not having to track cycle boundaries or fundamental frequency, which can be difficult for aperiodic voices [38, 39, 40, 41]. Correlation dimension (D_2), Lyapunov exponents, and Kolmogorov entropy are nonlinear dynamic methods that have been used extensively in research [12, 13, 30, 42, 43, 44]. D_2 measures the number of degrees of freedom that are necessary to describe a dynamic system, with more complex systems having a higher D_2 [43]. In the context of voice analysis, D_2 objectively describes the degree of periodicity or aperiodicity and chaos in the voice. When D_2 does not converge to a finite value, this indicates the presence of a high level of random noise [43]. Lyapunov exponents assess a dynamic system by focusing on two trajectories that are initially nearby and measuring their rate of divergence or convergence over time [44]. For voice samples that are nearly periodic, the signal remains stable, and the value of a Lyapunov exponent remains close to zero; however, chaotic signals have positive Lyapunov exponents [30]. Second-order Kolmogorov entropy (K_2) quantifies the rate that information about the system dynamics is lost, with a positive K_2 value indicating chaos in the signal [43]. The calculation of Lyapunov exponents, D_2, and K_2 involves reconstructing the phase space of the corresponding signal, which is used to describe all possible dynamic states of the voice signal over time [30, 45].

Meredith et al. performed nonlinear dynamic acoustic analysis of sustained vowels in 23 dysphonic and 15 healthy children. D_2 was higher among dysphonic children, indicating that the voices of dysphonic children require a higher number of degrees of freedom to be fully quantified and are therefore more aperiodic. Additionally, though jitter was higher among dysphonic children, variability was high for both groups [46].

Linear and nonlinear acoustic analyses were also utilized in a study of 111 healthy female and 101 healthy male children aged 6–12. While jitter and shimmer did not vary significantly with age or sex, fundamental frequency and largest Lyapunov exponent were lower in boys and decreased with age. These findings show that the boys' voices had lower frequency and were more stable than girls' voices [47]. Higher Lyapunov exponents have also been seen in cleft palate patients with hypernasality compared to cleft palate patients without hypernasality [48].

Formants

Resonance created by vocal tract filtering can be described by formants, which are characterized by a center frequency and bandwidth and are influenced by the length and shape of the vocal tract [49]. Similar to fundamental frequency, changes to formant frequency and spacing may provide important information about the voice [49, 50, 51].

In a study of ten 5-year-old children with cerebral palsy, formant measures were used to investigate dysarthria [51]. Analysis of single word recordings showed that children with dysarthria had smaller second formant ranges when uttering words that are known to require larger changes in vocal tract shape [51]. Formants were also used to collect normative acoustic data in Brazilian children aged 4–8. Seven different vowels were uttered by each of the 207 children. Frequencies of the first three formants were generally higher in girls than boys, and formant frequency decreased with age [52].

Emerging Parameters

While existing acoustic parameters have been beneficial to voice analysis in both research and clinical settings, they demonstrate limitations. Specifically, nonlinear methods including correlation dimension (D_2) and Lyapunov exponents are unable to distinguish between low-dimensional, deterministic chaos and high-dimensional, stochastic noise [53]. The implication for voice analysis is that it may be impossible to objectively quantify type 4 voice signals using these methods. This presents a significant limitation, considering that patients with significant breathiness, as in unilateral vocal fold paralysis, may have prominent stochastic noise and thus type 4 characteristics. However, nonlinear dynamic methods have been proposed recently that are better able to handle noise in the voice signal, resulting in fewer computational errors. These methods are spectrum convergence ratio (SCR), nonlinear energy difference ratio (NEDR), and rate of divergence (ROD) [54, 55, 56]. Computation of SCR relies on short-time Fourier transform analysis (STFT), which tracks how signal frequency components change with time. Most importantly, this analysis can be used to detect changes in periodicity. When a signal is more aperiodic or affected by turbulent noise, its spectrum consists of segments that are dissimilar to each other. Because SCR quantifies the convergence of segments, type 1 signals tend to have the highest SCR. A study in adults has shown that SCR can be successfully computed for all four voice types and that average values are significantly different across groups [54].

NEDR is similar to SCR in that it also involves a Fourier transform to decompose the signal into its frequency components. However, NEDR uses an iterative algorithm for calculating spectral energy variation among these frequency components. Briefly, the algorithm uses a nonlinear weighted function to weigh local data points based on their relative position to the data point of interest, before using the weighted function to perform a Fourier transformation. This process is repeated several times to improve the accuracy of the subsequent spectral energy distribution calculation. The output of NEDR characterizes the stability of the voice signal. Periodic signals will exhibit stable energy distributions, while the spectral energy of aperiodic signals will vary over time. NEDR has been found to be lowest in adults with type 1 voices. NEDR has also demonstrated the ability to distinguish among all four signal types [55].

ROD uses a modified algorithm for calculating Lyapunov exponents, which, as discussed previously, are the average exponential rates of divergence or convergence of nearby orbits in phase space. Higher maximum Lyapunov exponents indicate more chaos or instability in the voice signal. A limitation of this parameter is that calculation of Lyapunov exponents requires a known embedding dimension, which cannot be determined for type 4 voices. Rather than calculating Lyapunov exponents directly, ROD calculates the rate of divergence of two nearby points followed in three dimensions only. A pair of points is followed for three sample intervals, before a new pair is chosen. In total, eight fragments are analyzed for each voice sample, and the average value is taken to represent the ROD, which tends to be highest for type 4 voices. ROD has been used to successfully distinguish between all four voice types in a study of adults [56]. The ability to objectively distinguish between voice types 3 and 4 is particularly useful, because it enables the detection of subtler differences that may not be recognized perceptually.

Recently, SCR and ROD were used for the first time to study healthy pediatric voices. Acoustic recordings of 20 adult and 36 pediatric subjects aged 4–17 were taken. Subjects were then grouped according to their voice type using spectrogram analysis and CAPE-V. Mean SCR and ROD were found to be significantly different between the pediatric and adult groups, while jitter and shimmer were not. Using adult reference values for the SCR and ROD boundaries between type 2 and 3 voices, pediatric voices were grouped as primarily periodic or aperiodic. Using the original voice type designation as the true categorization, the adult SCR and ROD reference values were only capable of correctly sorting 36.1% of pediatric subjects. For analysis based on gender, boys in the age groups of 4–7, 8–12, and 13–17 all had similar SCR and ROD values, while girls aged 8–12 and 13–17 had significantly different values of SCR. These findings suggest that future research is needed, particularly for establishing appropriate pediatric reference values for these nonlinear parameters [57]. Potentially, these emerging methods could become clinically useful and be used in conjunction with other parameters to provide a comprehensive acoustic voice assessment.

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Clinical Approach to Acoustic Assessment

9

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Overview

Acoustic assessment is an integral part of the pediatric voice evaluation and requires that the speech-language pathologist (SLP) gathers reliable data that can be compared to a normative sample, thereby allowing for objective measurement of dysphonia. It goes without saying, obtaining a reliable data set with children can be challenging. The clinician must be prepared to deviate from standard protocols as needed, and tailor (customize) the assessment tasks to a given child, with consideration for the child's age, cognitive level, and general level of cooperation. In order to effectively determine appropriate modifications to use during the acoustic assessment, the clinician must have a clear understanding of the principles and foundations of acoustic assessment and analysis. Additionally, assessment tasks should be presented in the order of priority, as attention and cooperation can quickly wane and the clinician needs to be prepared for the child to cease cooperating at any time. Although children can often be coaxed back into finishing an assessment session, some may act silly or stubborn, both of which can result in an affective voice quality. The clinician should note any changes in voice production that may not be related to a voice disorder but rather are attributed to inattention, boredom, or other behavioral factors.

A Note on Age

Children across a wide age range can successfully complete the assessment tasks, and, therefore, there is no strict age recommendation for an acoustic assessment. Rather, to determine if a child may be successful with the assessment, consider the following questions:

- Can the child imitate?
- Is the child willing to interact or engage with the clinician?
- Are the parents willing to help?

If the answer is "yes" to any of the above questions, then we recommend trying to collect acoustic data from that child.

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Prior to Collecting Voice Recording

The child's comfort level can have a significant impact on what acoustic recordings are collected, how the child's voice sounds, and degree of variation from the standard protocol needed. The examination room is unfamiliar and can be an anxiety-provoking place. If a child does not feel comfortable during an assessment, clinicians may hear statements from parents such as "she is much louder at home" or "he is using a 'baby' voice right now because he is nervous." It is important to verify with caregivers that the child's voice during the assessment is consistent with, or different from, their usual manner of speaking.

An essential part of acquiring a usable acoustic sample is making the child as comfortable as possible. Incorporating toys in the session, engaging the parent, and taking the time to allow the child to acclimate to the clinician and the new environment may be beneficial to a successful outcome. When possible, the clinician should take time at the beginning of the assessment to engage the child in natural conversation, before a microphone is placed in front of him or her. The more comfortable the child is, the more likely we are to obtain a recording of the child's natural voice and speech patterns.

Equipment and Calibration

Detailed instructions on equipment specifications and setup can be found in the article constructed by an expert panel entitled "Recommended Protocols for Instrumental Assessment of Voice: American Speech-Language-Hearing Association Expert Panel to Develop a Protocol for Instrumental Assessment of Vocal Function" (ASHA-IVAP) [1]. One key element to successful acoustic assessment with children is a head-mounted microphone. This may be a microphone that is affixed to the head via an adjustable headset or one that goes behind the head and over the ears. Using a head-mounted microphone maintains a consistent distance from the microphone to mouth, thereby improving the signal to noise ratio and reducing the overall impact of environmental noise. To minimize noise in the signal further, the child is encouraged to sit as still as possible. It may be beneficial to ask the parents to assist during the assessment, and, for some children, sitting on their parent's lap may be helpful. When the child tolerates the headset without excessive movement, a calibration value can be acquired prior to completing the tasks, and this allows the clinician to address absolute measures of sound pressure level. See ASHA-IVAP for details on how to perform a calibration [1]. Due to both the added time calibration requires as well as the need for the microphone to stay in a consistent place, calibration can be undertaken with children when there is no concern regarding attentional difficulties or excessive movement. When the calibration is not acquired or the microphone moved during the assessment, absolute measures of sound pressure level cannot be calculated for the assessment.

Some children may not tolerate a head-worn microphone. Whether this is due to sensory difficulties or general compliance, using a microphone attached to a headset is not always feasible. Alternative options include the clinician holding a microphone, placing a microphone on a stand on the table in front of the child, or attaching a microphone to the child's clothes. If an alternative option for microphone placement is used, the acoustic analyses must be interpreted with caution. These alternative methods may result in inconsistency in the signal due to the child's variable distance from the microphone.

The clinician should monitor the gain of the signal throughout the assessment, whether the microphone is mounted on the head or in an alternative placement, as many children present with variable loudness. For example, a child may sporadically speak with a loud voice, lean into the microphone to speak, or intermittently speak forcefully to emphasize a point. These aspects may be natural features of a child's speech; however, they can cause clipping or distortion of the signal. If these elements are intermittent and the gain is appropriate, the clinician can avoid adjusting the gain as long as sections that are clipped are not analyzed. If the signal is consistently clipped, then the gain should be lowered, and that signal should not be used for analysis.

Elicitation of Speech Tasks

It is important for the clinician to consider three points with each task that is undertaken during the assessment: (a) *purpose*, what is the purpose of the task (i.e., what information will it provide about the child's voice); (b) *optimal elicitation method*, what is the standard protocol to collect acoustic data for the task; and (c) *alternative elicitation method*, if the optimal elicitation method wasn't feasible, what was the actual elicitation method performed. Answers to these questions provide valuable guidance for the clinician performing the assessment. Understanding the purpose of the task will help the clinician prioritize which tasks to try and elicit, and in what order. When interpreting the results of the completed tasks, it is essential to note if there was any deviation from the standard administration protocol. It is optimal to use a standardized protocol for elicitation and production, thereby allowing comparison of the assessment results to published normative data (e.g., [2, 3]). When the clinician deviates from the standard acquisition protocol, this should be documented, and consideration must be given to this with the acoustic analysis.

Sustained Vowels

Purpose

The speech tasks for analysis include sustained vowel productions elicited at the child's typical speaking pitch and volume. Measurements that are done include habitual fundamental frequency (f_o) and vocal quality measures such as cepstral peak prominence (CPP). Additional measures of vocal quality including jitter, shimmer, and harmonics to noise ratio may be evaluated as well, although they should be in interpreted with caution.

Optimal Elicitation Method

The suggested protocol for sustained vowel assessment is the production of the /ɑ/ vowel for 3–5 s at a steady pitch and volume [1]. Children may require cues to successfully complete the task. These can consist of prompts to take a substantial (big) breath before speaking or may involve the clinician modeling a sustained vowel. When modeling a task, the clinician should be aware that the child may try to exactly imitate the clinician's voice; i.e., the clinician's f_o may influence the f_o produced by the child [4]. The clinician should try to model as infrequently as possible and, when modeling is done, encourage the child to speak at his or her own pitch. Visual cues are also an effective method of assisting children to produce sustained vowels. This may include the clinician silently counting the seconds of the sustained production out on her fingers or slowly bringing her hands together, with contact signaling the cessation of phonation.

One second of the vowel production that is steady in pitch and loudness should be selected for analysis. Research has shown that f_o measures in children can be consistently measured from vowels using either time-based measures [5, 6] or frequency-based measures [2, 6]. Time-based measures of vocal quality, including jitter, shimmer, and harmonics to noise ratio, were previously considered to be the standard. These have been found to be less consistent [5, 6] and significantly affected by deviations in loudness [7]. There has been a recent shift to focus on frequency-based measures, specifically cepstral peak prominence (CPP; [1]), which provides an overall measure of periodicity. The measure of CPP has been shown to have good reliability in children [6] and is the current recommended protocol for analysis of sustained vowels [1]. Although measures of CPP are more robust than time-based measures, the clinician must still monitor for steady production of the vowel, as CPP values can be impacted by changes such as loudness [8].

Alternative Elicitation Method

There are children who are unable to produce a sustained sound in isolation, whether due to cooperation difficulties or cognitive abilities. Some,

however, prolong a vowel production in the context of familiar words or sound effects elicited during play, especially when toys are involved. For example, a child may produce sustained vowels when asked, *Can you make a sound like a*:

- *Ghost* ("boo")
- *Cow* ("moo")
- *Sheep* ("baa")
- *Owl* ("whoo")
- *Train* ("choo-choo")

These productions may contain different vowels and/or be influenced by coarticulation with surrounding consonants. Although it is not advisable to compare these productions directly with normative values, eliciting sustained vowels using these alternative methods may still provide valuable information on the child's sustained vocalic productions. Clinicians should be aware with alternative elicitation methods when the child's voice demonstrates improved vocal quality as compared to his or everyday voice. This may occur the vowel itself, or the abutting consonant promotes forward focused resonance affecting the vocal quality of the vowel. Additionally, CPP differs based on vowel type [8]; therefore, comparison to normative data should be evaluated with caution.

Connected Speech

Purpose

With connected speech, we want to acquire a sample of speaking that encapsulates the child's everyday speaking voice. Analysis is undertaken to measure habitual speaking f_o and SPL, CPP, and variability of f_o during connected speech.

Optimal Elicitation Method

In adults, the optimal elicitation method for assessment of connected speech is a standard passage [9]. Although there is a standardized passage available for children who can read ("The trip to the zoo"; [10]), we would argue that reading does not provide the optimal representation of a child's everyday speaking voice. Regardless of age, reading even a simple passage out loud is difficult for many children. Reading aloud may a result in a child using a "performance voice," that is, one that is used to present in front of a class rather than a voiced used during every-day play.

Instead of reading, the clinician may obtain a conversational speech sample with vocal characteristics that are representative of the child. This is no small feat and requires building a rapport with the child and finding a topic of interest. If time allows, it is best to engage the child in a casual conversation before the microphone is put in front of him or her. Asking open-ended questions is a great way to search for topics the child may want to talk about. Questions can surround anything the child would like to talk about, such as:

- What did you do this weekend?
- Who are your friends at school?
- What is your favorite class at school?
- Take me through your school day; tell me your schedule and what teacher teaches each class.
- What's your favorite/book/movie/show?

A successful outcome will often require follow-up questions to elicit a more continuous stream of speech. It may be helpful to keep the microphone on the entire time, even when the clinician or parent speak, making it more likely to catch an adequate segment of spontaneous speech from the child to analyze.

For example, below is a common scenario:

Clinician: *What's your favorite movie?*
Child: *Frozen*.
Clinician: *What a great movie! Who is your favorite character?*
Child: *Elsa*.
Clinician: *Can you remind me, what did Elsa do in that movie?*
Child: *She froze everyone, then she ran away, then Anna found her, and made a snowman.*

If the child refuses to talk to the clinician, engaging the parent is a good next step. The clinician can guide the parent that she would like to

hear as much of the child speaking as possible, emphasizing that the ideal sample will have just the child's voice. Recording the entire parent-child interaction increases the chances of catching a longer string of spontaneous speech from the child. Once a speech sample has been selected that the clinician feels is representative of the child, desired measures can be calculated and, if appropriate, compared to normative values. Ideally, the middle portion of the sample would be selected for analysis, providing as stable a portion of speech as possible. In addition to the measures of f_o and CPP discussed in the *sustained vowel* section above, connected speech can also be examined for the variability of f_o and habitual sound pressure level (SPL) values. Clinicians should remember that absolute values of SPL can only be analyzed if a calibration value was collected and the microphone has remained in the same location during the assessment.

Alternative Elicitation Method

If the child is unwilling or unable to produce a spontaneous speech sample, additional tasks may be useful to elicit connected speech. Counting is often a successful method to use for children of all ages. The result is a rote response that children can often produce on their own, without modeling needed. During counting tasks, children may need prompts to slow down, as they may want to show you how fast they can count. Phrase and sentence repetition are also viable options, though the clinician must be very aware of the model they are presenting, similar to what was discussed in the *sustained vowel* section above. When children are presented with a model, they will often copy that model exactly, in this case, using the same f_o, intonation, and stress patterns. If possible, using multiple tasks (e.g., sustaining a vowel embedded in sound effects/words, repeating a sentence, counting, etc.) to measure habitual f_o may provide the most valuable information as there is variable evidence on whether there are systematic differences to the f_o measured from a given task type [4, 11, 12]. If the child is able to remember a short phrase, the clinician can have him or her repeat the phrase a few times. Successive repetition often lends itself to the production of more natural speech parameters, allowing the clinician to have a clearer understanding of the child's habitual f_o. The Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V, [13]) sentences are good stimuli; the sentences are short and allow for examination of voicing in multiple contexts. In addition, as the goals of any assessment with children are both efficiency and accuracy, using the CAPE-V sentences to examine habitual pitch will also provide the opportunity to begin the perceptual evaluation (see Chap. 12).

Pitch and Loudness Ranges

Pitch and loudness ranges are recommended for assessment of adults with voice disorders [1]. However, reliably relating these concepts to children and eliciting adequate responses for analysis are often difficult. If the child is cooperative enough and/or cognitively able to take explicit directions, both minimum and maximum pitch and loudness values can be obtained and subsequently inform the assessment. More informally, pitch and loudness ranges can provide insight into vocal flexibility in children. Some children will initially appear in the clinic with voices that have mono-pitch or mono-loudness; seemingly, this is often attributable to nervousness in an unfamiliar environment. In this case, the clinician may model pitch or loudness glides and have the child imitate them. When wide ranges of pitch or loudness levels are heard, this suggests that the mono-pitch or mono-loudness heard may not be pathological but rather more affective given the current conditions.

Conclusion

With appropriate knowledge, skills, and flexibility, an acoustic assessment can be successfully completed in children of all ages. A key element to this success is creating a comfortable environment and a rapport with the child, thereby encouraging elicitation of the child's natural speaking voice. Deviations from standard assessment pro-

tocols used in adults may be necessary for children, and the clinician must carefully consider the potential impact of these variations. It is essential for the clinician to understand the fundamentals of the assessment tasks and analysis methods, as with this basis she can accurately interpret acoustic assessment results from a variety of elicitation methods.

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Aerodynamic Voice Assessment

10

Hunter Huth, Austin J. Scholp, and Jack J. Jiang

Introduction

During voice production, the larynx acts as an energy transducer that converts aerodynamic energy from the lungs into acoustic energy heard as voice [1]. The efficiency of this energy transduction is dependent on a variety of factors including glottal configuration, hydration, vocal fold tension, as well as the aerodynamic inputs of airflow and pressure [2]. The aerodynamic inputs are analogous to an electrical circuit where airflow, subglottal pressure, and laryngeal resistance correspond to current, voltage, and resistance [3]. Aerodynamic parameters that reflect the energy required to produce voice are helpful indicators of vocal function [4]. Abnormal aerodynamic parameters have been demonstrated in the setting of numerous laryngeal pathologies including vocal nodules [5, 6], polyps [7], and Reinke's edema [8]. Accordingly, aerodynamic measurement remains a critical component of the comprehensive voice assessment.

Parameters

Airflow

Glottal airflow is measured as the volume velocity of air that passes through the larynx during a given period of time. Two parameters describing airflow are phonation threshold flow (PTF) and mean flow rate (MFR). PTF is the minimum airflow required to initiate phonation [9] and is derived according to the following formula:

$$\mathrm{PTF} = L\sqrt{\frac{8x_0^3 Bc}{T\rho}} \tag{10.1}$$

where L is vocal fold length, x_0 is the neutral glottal width, B is the damping coefficient, c is the mucosal wave velocity, T is the vocal fold thickness, and ρ is the density of air. In this model, the damping coefficient correlates to the stiffness of the vocal folds. For example, vocal fold scarring would increase the stiffness, thus increasing the damping coefficient and PTF [10]. A similar phenomenon is observed in the setting of benign mass lesions such as polyps or nodules [7]. Changes in pre-phonatory glottal width, as in the setting of glottic insufficiency, also affect PTF. An excised

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larynx experiment was conducted that showed PTF was more sensitive than phonation threshold pressure (PTP) to changes in glottal width [11].

MFR describes airflow during sustained phonation. Both PTF and MFR have been shown to differentiate between normal and pathological voice productions [10]. MFR is also one of the two parameters used to measure derived laryngeal resistance (R_L), where R_L is equal to subglottal pressure (P_s) divided by MFR.

Pressure

During phonation, subglottal pressure accumulates inferior to the glottis and serves as the driving force for vocal fold vibration. Phonation threshold pressure (PTP) is the minimum subglottal pressure required to produce stable phonation. Similar to PTF, PTP is sensitive to changes in vocal fold thickness and stiffness [12]. However, PTP is less sensitive to changes in glottal width [11]. Through a similar model, it can be described by the equation:

$$\mathrm{PTP} = \frac{k_t B c x_0}{T} \tag{10.2}$$

where k represents a transglottal pressure coefficient, c is the mucosal wave velocity, B is the damping coefficient, x_0 is the pre-phonatory glottal half-width, and T is the vocal fold thickness. PTP is often elevated in disease states [7, 12, 13].

Resistance

In general, flow resistance is defined as the ratio of pressure to flow and is often measured in centimeters of water per liter per minute ($cmH_2O/L/min$). Ideally, this ratio would remain constant regardless of pressure and flow, thus providing an invariant characteristic of the airway [14]. Airflow resistance provides information concerning the glottal size, configuration, and biomechanical properties [15]. Glottal resistance (R_g) is calculated using the pressure across the glottis and the airflow through the glottis [16]. Laryngeal resistance (R_L) is similar but is calculated from subglottal pressure (P_s) and translaryngeal airflow [17]. In a comparison of R_g measurements, Netsell et al. found that females typically have higher resistances than males. As resistance depends on the size of the airway, this was attributed to females having smaller larynges [18]. However, resistance is dependent on other factors including degree of vocal fold adduction, roundness of glottal entry and exit, [19] and the speed of air particles moving through the glottis [20].

Power

Power is a measure of the work done on an object over time and is measured in watts. Electrical power is calculated by multiplying current and voltage. Using our electrical circuit analogy, we can calculate aerodynamic power as the product of airflow and subglottal pressure. The minimal power required to initiate phonation, phonation threshold power (PTW), can also be calculated. Combining the equations for PTF and PTP from above, we obtain the following:

$$\begin{aligned}\mathrm{PTW} &= \mathrm{PTP} \times \mathrm{PTF} \\ &= \frac{k_t B c x_0}{T} \\ &\quad \times L\sqrt{\frac{8x_0^3 Bc}{T\rho}} \\ &= k_t L\sqrt{\frac{8B^3c^3x_0^5}{T^3\rho}}\end{aligned} \tag{10.3}$$

Pathologies that increase the mass or stiffness of the vocal folds or increase the glottal width will increase the power required to start and maintain phonation [21]. In comparison of PTP, PTF, and PTW, PTW had the greatest ability to differentiate patients with mass lesions and vocal fold immobility compared to healthy subjects [13, 22].

Vocal Efficiency

Vocal efficiency is defined as the ratio of acoustic power obtained for a given amount of aerodynamic power. This can be calculated with the following equation:

$$\mathrm{VE} = \frac{\mathscr{P}_{\mathrm{ac}}}{\mathscr{P}_{\mathrm{aero}}} = \frac{4\pi r^2 \times I}{P_{\mathrm{s}} \times U_{\mathrm{g}}} \tag{10.4}$$

where $\mathscr{P}_{\mathrm{ac}}$ is acoustic power and $\mathscr{P}_{\mathrm{aero}}$ is aerodynamic power. Aerodynamic power, as mentioned previously, is the subglottal pressure P_{s} multiplied by the glottal flow U_{g}. Acoustic power is the amount of energy emitted by a source over time and is calculated by multiplying the measured sound intensity I by the surface area of a sphere with radius r. The radius in this case is the distance from the source [7]. During phonation, aerodynamic power is converted into mechanical energy which causes the vocal folds to vibrate. This vibration creates an air column of oscillating pressure perceived as voice. The amount of acoustic power is reduced by turbulence of the air stream as it exits the glottis. There are additional losses through viscous forces and wall vibrations as the air travels through the vocal tract [23]. Vocal pathologies, such as polyps and nodules, can increase the mass of the vocal folds and create pressure leaks in the glottis. This then increases PTP and PTF, thus increasing $\mathscr{P}_{\mathrm{aero}}$ and decreasing vocal efficiency [7]. Pathologies that alter the hydration and stiffness of the vocal folds have similar consequences on efficiency. Studies performed with excised models found that increased longitudinal tension [24], decreased hydration [25], and increased glottal width [26] all significantly reduced vocal efficiency.

Clinical Assessment Methods

The development of methods to accurately assess and quantify these aerodynamic parameters is a current area of research. Measurement of subglottal pressure is of particular interest as it describes the driving force for voice production. Previously utilized methods for measuring subglottal pressures include a transtracheal pressure transducer [17, 27]. While this was accurate, it is invasive and not feasible for routine assessment. Two approaches have been developed for noninvasive indirect subglottal pressure measurement, labial interruption and mechanical interruption.

The first noninvasive method of subglottal pressure assessment was introduced in 1981 by Smitheran and Hixon. Their method was based on the assumption that oral and subglottal pressures reach an equilibrium during the production of a stop-plosive (/pα/) [17]. The validity of this measurement was confirmed through the use of a trans-nasal transducer by Löfqvist et al. [27]. The labial interruption task has been adapted to also measure PTP. To do this, the subject phonates as quietly as possible while producing the stop-plosives [22]. Labial interruption has been shown to be a reliable assessment method for both adults [28] and children [29]; however, it can be difficult for the subject to master, leading to higher intrasubject variability [30].

The second approach is through mechanical interruptions, which was first developed in 1992 by Bard et al. [31]. The principle of mechanical interruption is similar to that for labial interruption; however, control of the interruption is taken away from the subject by replacing the stop plosive with the closure of a mechanical valve, thus theoretically allowing for reduced variability. While there are different variations, typical mechanical interruption uses a tube equipped with a balloon valve that inflates to cut off airflow and phonation (Fig. 10.1). The occlusion causes the pressure in the mouth and tube to equilibrate to the pressure below the glottis. This method was developed further by Jiang et al. to obtain airflow, pressure, efficiency, and resistance measurements in patients with Parkinson's disease [32]. Through a direct comparison of labial and mechanical interruption, it was found that mechanical interruption provided higher measurement precision for laryngeal resistance in adults [28]. A recent comparison of the two approaches found similar measurement reliability for phonation threshold pressure in pediatric subjects [29].

Compared to subglottal pressure, airflow measurement is simpler. An assumption is made that no air loss occurs into the tissues of the vocal tract and thus, airflow exiting the mouth is equal to that passing through the glottis. Devices such as the Rothenberg mask and other flow measurement methods work off the Ohm's law analog mentioned previously. By measuring the pressure

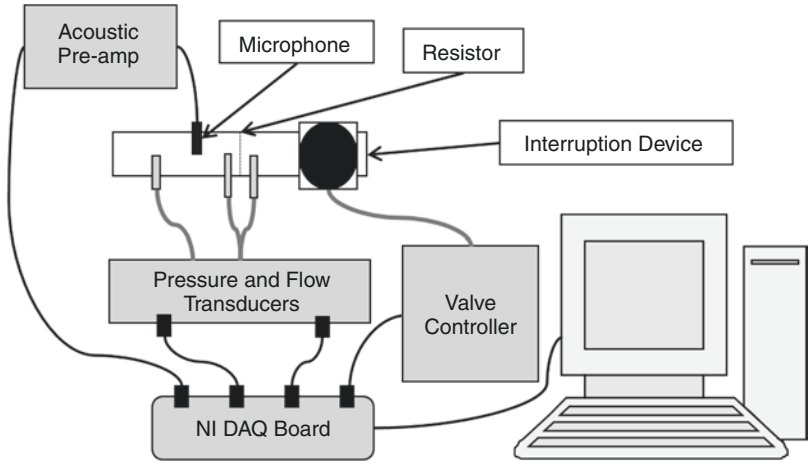

Fig. 10.1 Schematic of the airflow interruption system. The subject produces a sustained vowel into a mask or mouthpiece, and pressure is measured within the device during interruption of airflow by a balloon valve. (From Jiang et al. [37], with permission)

difference across a known resistance, flow can be calculated [33].

Current Clinical Equipment

The Phonatory Aerodynamic System (PAS) is currently used for clinical voice assessment. PAS model 6600 was developed in 2006 by the KayPENTAX Corp. to replace the Aerophone II model 6800 created by Kay Elemetrics Corp. The PAS can simultaneously capture sound intensity, intraoral pressure, airflow rate, and fundamental frequency and has an auxiliary port to allow for the collection of electroglottography. It also includes protocols for common phonatory measurements including vital capacity, air pressure screening, comfortable sustained phonation, vocal efficiency, and running speech analysis as well as normative data for pediatric and adult subjects to assist clinicians with interpreting results [34, 35]. The labial interruption technique is used to assess subglottal pressure with the PAS. In studies that examined the test-retest reliability of the PAS, the parameters of glottal power, efficiency, and resistance had substantial coefficients of variation in both men and women [36].

Mechanical Interruption Methods

The first airflow interruption method developed involved complete occlusion of the vocal tract. During a trial, following the closure of the balloon valve, supraglottal pressure increases until it equilibrates with subglottal pressure. As supraglottal pressure increases, there is a pressure at which phonation ceases. This pressure is subtracted from the final equilibrated pressure to calculate phonation threshold pressure. In other words, when the pressure in the tube reaches a certain value, the pressure difference between subglottal and supraglottal is not great enough to sustain phonation (Fig. 10.2) [37].

Measuring PTP with this method provides an offset PTP, which is the minimum pressure to sustain phonation after it has been initiated. In comparison, labial interruption provides an onset pressure—the pressure required to initiate phonation. Due to a hysteresis affect, PTP offset is lower than PTP onset similar to the differences between

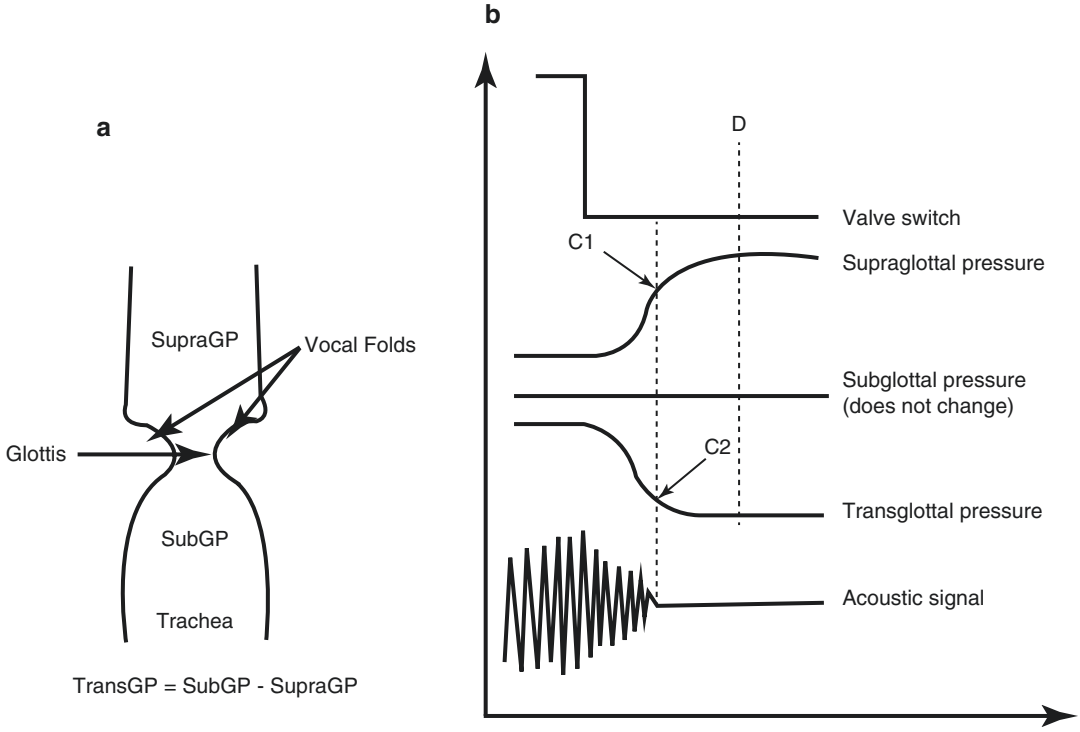

Fig. 10.2 Measurement of phonation threshold pressure using airflow interruption. (**a**) Relationship between the transglottal pressure, subglottal pressure, and supraglottal pressure.(**b**) Supraglottal pressure at moment D (measured by device) after interruption estimates subglottal pressure because the transglottal pressure falls; moment C1, the supraglottal pressure at which phonation ceases, corresponds to moment C2 on transglottal pressure graph; this transglottal pressure is equal to the phonation threshold pressure (PTP); therefore, C2=D – C1. (From Jiang et al. [37], with permission)

kinetic and static friction [38]. Therefore, noting the method of PTP measurement is an important consideration for vocal assessment.

Normally, during an interruption, supraglottal pressure rises and reaches a plateau equal to the patient's subglottal pressure (Fig. 10.2, supraglottal pressure data trace). Some have theorized that the force exerted by a mechanical interruption could potentially trigger a laryngeal adductor reflex (LAR). This could cause a sudden rise in pressure following the interruption that would interfere with accurate P_s and PTP measurement. Hoffman et al. observed experimentally that when using an airflow interrupter, a pressure plateau would occur within the first 150 ms of the balloon valve inflation, but the pressure could sometimes continue to increase after the plateau. The rise in pressure after the plateau may be due to the LAR, which would have an average latency between 150 and 175 ms after interruption. Supraglottal pressure measurements at 150 ms post-closure were determined to be more precise than measurements made via analysis of pressure plateaus [39]. Other methods have been developed that attempt to lessen the effect of the LAR or avoid it completely.

In order to lower the chances of triggering the LAR, the incomplete interruption method was created (Fig. 10.3). As the name suggests, instead of completely occluding subject airflow, the device is only partially occluded during mea-

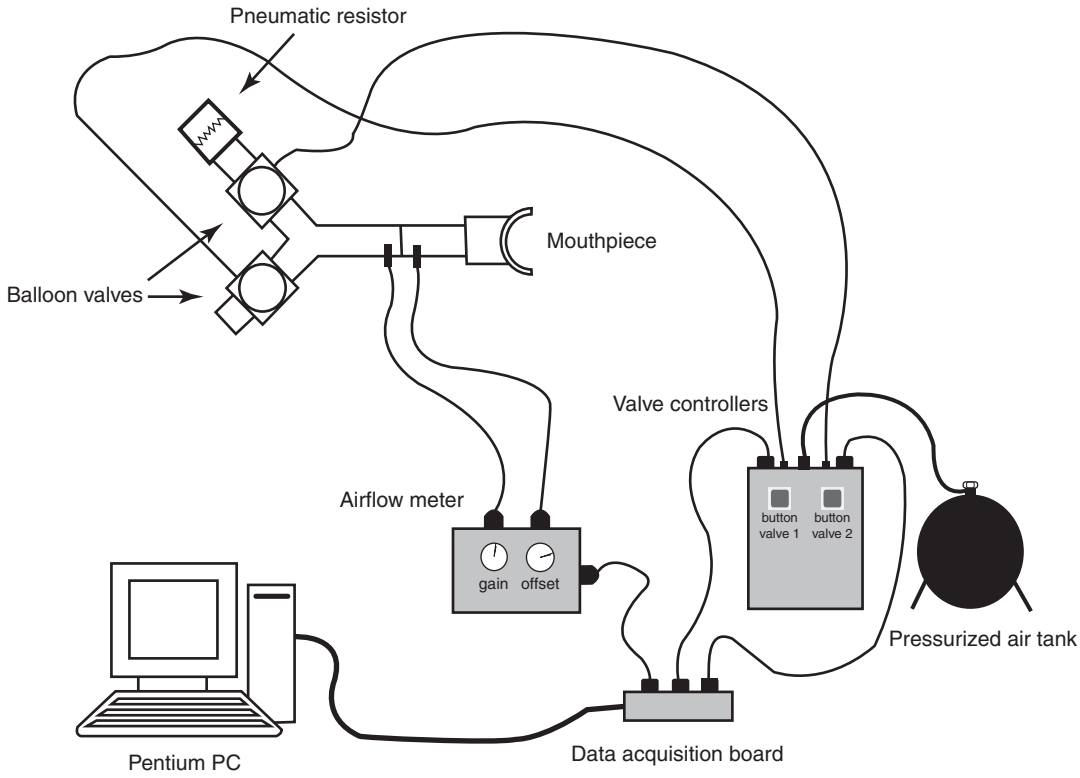

Fig. 10.3 Incomplete airflow interruption setup. (From Jiang et al. [3], with permission)

surement. The experimental construction of this circuit, shown in Fig. 10.4, uses two balloon valves to guide airflow through two pathways of different resistances. A system of equations is constructed using the circuit design:

$$P = Z_g U_1 + Z_1 U_1 \tag{10.5}$$

$$P = Z_g U_2 + Z_2 U_2 \tag{10.6}$$

where P is subglottal pressure, Z_g is glottal resistance, Z_1 and Z_2 are the known resistances of each pathway, and U_1 and U_2 are the flows measured when each pathway is open. This can then be solved for the input pressure P and the glottal resistance Z_g.

$$P = \frac{U_1 U_2 (Z_1 + Z_2)}{U_2 - U_1} \tag{10.7}$$

$$Z_g = \frac{Z_2 U_2 - Z_1 U_1}{U_1 - U_2} \tag{10.8}$$

Human subject trials revealed the incomplete airflow interrupter measured subglottal pressure within similar ranges to previous measurement techniques and was comparable to complete airflow interruption [3].

Airflow redirection, developed by Baggott et al., is another alternative method used to measure subglottal pressure that was designed to bypass the effects of laryngeal reflexes that can lead to inaccuracies in measurements (Fig. 10.5) [40]. The airflow redirector, similar to previously discussed interrupters, uses a fast-closing balloon valve that interrupts subject airflow during sustained vowel production; however, instead of occluding or par-

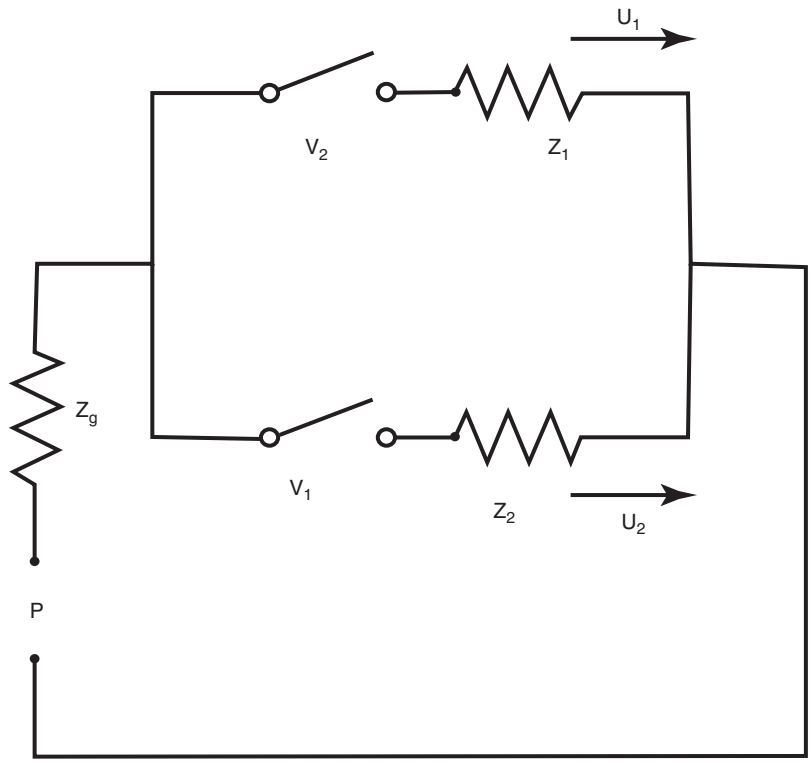

Fig. 10.4 Incomplete interruption circuit. Impedance of each output branch of the mouthpiece (Z_1 and Z_2) is known; airflow (U_1 and U_2) is measured through each output branch; glottal resistance (Z_G) and subglottal pressure (P) are unknown. (From Jiang et al. [3], with permission)

tially occluding the airway, it redirects the airflow through a one-way pneumatic valve into a pressurized tank. When the balloon valve repeatedly closes for short interruptions of ~135 ms, the tank pressure increases until it is equilibrated with subglottal pressure [40]. The airflow redirector has been validated to have a less than 5% error on a tracheostomy patient, but its effectiveness as a diagnostic tool will be a subject of future research [40].

The airflow redirector can also be used to calculate laryngeal resistance. To do this, the air tank of the redirector acts as a capacitor, while the larynx acts as a resistor in an RC circuit. In this system, the time constant (τ) is calculated by multiplying laryngeal resistance (R_L) by the capacitance (C) of the tank.

$$\tau = R_L \times C \tag{10.9}$$

This equation can be solved for resistance to estimate the laryngeal resistance of the subject [15]. Tank capacitance is calculated with the following equation:

$$C = \frac{VV_1}{RT} \tag{10.10}$$

where V is the volume of the tank, V_1 is the mean gas volume, R is the ideal gas constant, and T is temperature. τ is calculated experimentally from the pressure trace at each balloon inflation [15].

In addition to interruption and redirection methods, a device has been developed to allow for airflow to be calculated while allowing for free motion of the mandible and normal articulation. This device (Fig. 10.6) allows airflow to be characterized during conversational speech and singing and provides a natural environment for the subject during airflow measurement collection. The device also operates based on the aerodynamic analog of Ohm's law. By measuring the pressure changes within the helmet and keeping the resistance constant, airflow can be calculated. This device is still in early stages of development [41].

Recently, complete mechanical interruption has been tested in a pediatric population. Both labial and mechanical interruptions were used

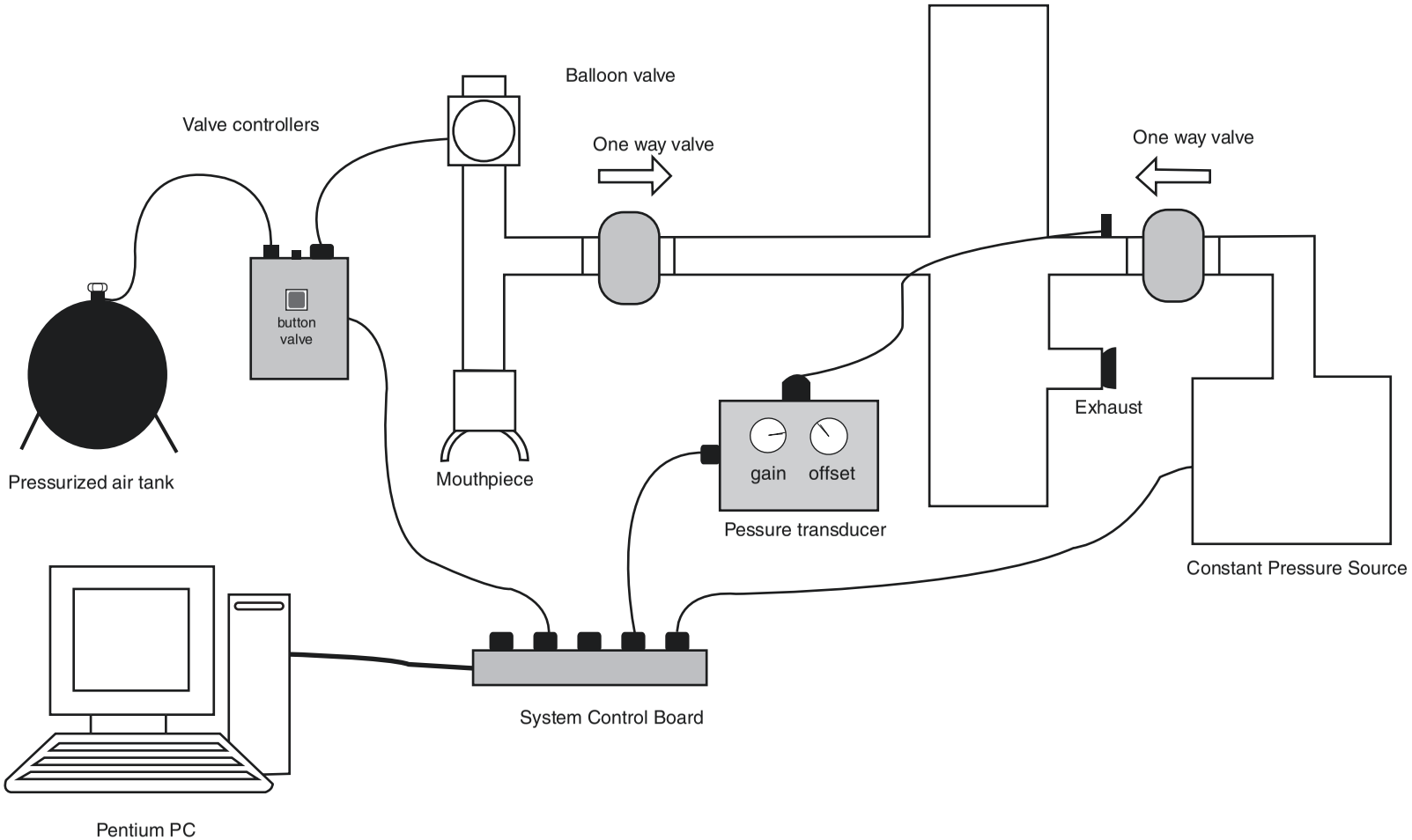

Fig. 10.5 Schematic of the redirection system. The principle is similar to airflow interruption, but airflow is redirected into a larger tank (capacitor) that may improve measurement stability. During the short balloon valve interruptions, supraglottal pressure spikes. This system equilibrates this supraglottal pressure with an air tank attached to the system via a one-way pneumatic valve. When a supraglottal pressure spike no longer changes the pneumatically retained pressure in the air tank, the tank pressure resisting the one-way valve should be equal to the pressure generated by the lungs during constant phonation, subglottal pressure. (From Baggott et al. [40], with permission)

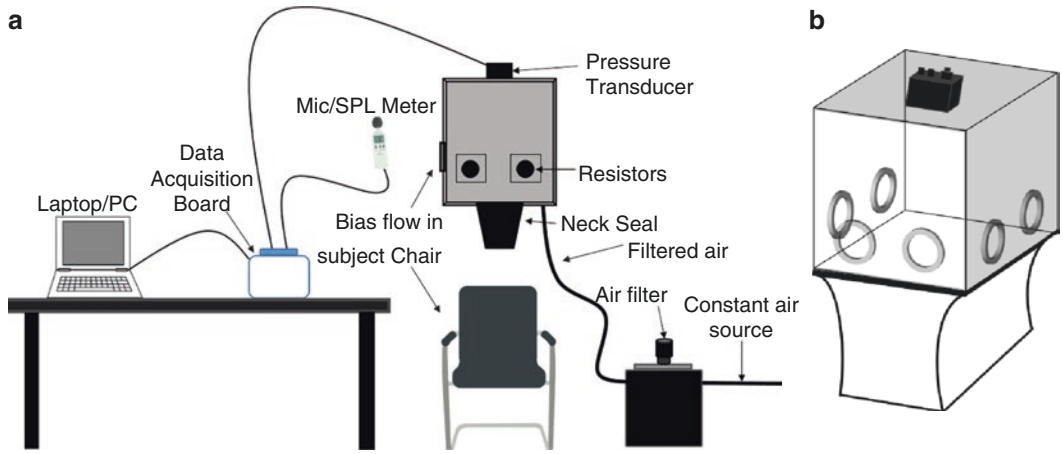

Fig. 10.6 (**a**) Schematic of the singing helmet which allows for continuous airflow measurement with normal articulation. (**b**) Closer view of device. The subject's head fits through the elastic seal on the bottom, and red resistors allow air movement through the device. A biased flow air current is also introduced into the device. (Reproduced from Jiang et al. [41], with the permission of the Acoustical Society of America)

to assess subjects with normal voice as young as 4 years old with acceptable precision. Mechanical and labial interruptions had similar variability for subglottal pressure and phonation threshold pressure, without significant changes in variability related to age [29]. Differences in mean phonation threshold pressure were observed, with labial interruption values being higher than mechanical. This could be related to differences in task as well as the hysteresis effect. Evaluation of children with dysphonia as well as the other assessment devices in the pediatric population will be the focus of future studies.

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Clinical Approach to Aerodynamic Assessment

11

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Overview

Aerodynamic assessment is undertaken to obtain information about vocal function via measurement of subglottal pressure and glottal airflow, i.e., glottal aerodynamics. As is the case with acoustic assessment, aerodynamic assessment can be accomplished with children of a variety of ages [1]. Acoustic and aerodynamic measurements are often conducted concomitantly, and previously detailed suggestions for performing an acoustic assessment are appropriate; please refer to Chap. 9.

Equipment

The primary method of aerodynamic assessment utilizes noninvasive equipment consisting of a pneumotachograph, a device designed to measure airflow. During the assessment, a face mask is placed over the child's nose and mouth, and this directs airflow into the pneumotachograph. Measures of intraoral pressure are simultaneously acquired via an intraoral tube, which rests in the open space in the child's mouth. Finally, acoustic information is collected via a microphone that is placed at a fixed distance away from the mouth. Commercially available systems are typically used to collect all three signals and keep them time-aligned, reducing the burden of analysis on the clinician. Results of the aerodynamic assessment can be compared to normative data in vocally healthy children (e.g., [2–4]). For details on equipment specifications, calibration, and appropriate signal criteria, please refer to the recommended protocols for the instrumental assessment of voice [5].

Familiarization

Allowing time for the child to become familiar with the equipment is a key component of a successful aerodynamic assessment, as the placement of the face mask over the nose and mouth is unfamiliar and may be met with apprehension or even anxiety. This can be especially true among children who have previously undergone surgery, as they may assume similarities between the face masks used to prepare for surgery and that which is used in the current assessment. Children may

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feel more comfortable if they are able to handle the face mask on their own prior to the assessment. The clinician can encourage them to hold it up to their face or their parent's face and take time to become comfortable with the equipment. Many children are put at ease with both the freedom to explore the equipment as well as direct explanations and reassurance that nothing will come out of the face mask.

Task

Aerodynamic measures are collected during a single task consisting of a syllable train of /p/ + vowel productions at a comfortable pitch and loudness. Ideally, the vowel produced allows the tongue to remain in a neutral position such that it does not block the oral cavity and encourages oral airflow and promotes good velopharyngeal closure [6, 7]. For adults, the /i/ vowel is often recommended; however, saying /pi pi pi pi pi/ is silly for many children. Therefore, as vowel choice does not have a significant impact on tasks with similar elicitation methods in adults [8], many clinicians choose an alternate vowel for aerodynamic assessment. In our experience, the /æ/ vowel is a good choice to use for children during this task, and, thus, we will use the exemplar /pæ/ for the remainder of this chapter.

Successful completion of an instrumental aerodynamic assessment involves practice of the task with the child prior to data collection. The clinician should coach the child to produce five to seven consecutive /pæ/ syllables in a single breath. Syllable strings should be produced in an easy manner at a rate of 1.5–2 syllables per second [6]. The /pæ/ syllable strings should be produced with the child's typical pitch and loudness. One method the clinician can use to accomplish consistent productions is to have the child produce a sustained /æ/ sound and then practice closing his or her lips at even intervals to produce the /p/. Alternately, the clinician can use imitation during practice; have the child mimic your production and provide feedback (e.g., give a thumb's up indicator) at moments when the productions are optimal. Visual cues can also assist the child in producing the string of /pæ/ productions at a consistent rate. These may include a visual metronome, quietly tapping or clapping, or silently mouthing the productions along with the child. Many children benefit from the clinician counting on her fingers during the task such that the child knows when it is time to take a breath.

Observations During the Task

When performing aerodynamic assessment in children, clinicians must carefully observe the child as well as the instrumentation during the task. The face mask must remain flush on the child's face the entire time. If necessary, help from the parent or the child can be solicited to ensure that the face mask has a tight seal on the face. If this cannot be accomplished, whether due to a facial abnormality or compliance by the child, aerodynamic estimates will be inaccurate. The clinician will also need to observe for any adverse interaction the child has with the oral catheter tube. The optimal placement is such that the tube sits within the intraoral space and does not impact articulation. However, some children may react to the tube adversely: bunching up their tongue to avoid it, pressing their tongue against it, or keeping their lips open to avoid touching it while they speak. It is important that the clinician encourage the child to close their lips fully around the tube, as a seal at the level of the lips is essential for intraoral pressure estimates. Additionally, bunching up or pressing their tongue against the tube may result in abnormal pressure transducer measures due to tongue position or saliva buildup. Aberrant tongue positions can usually be detected perceptually and often remedied with further instruction.

It is important that the clinician observes the child's natural speech and resonance patterns prior to completing the task in order to identify any speech deviations that would affect the aerodynamic results. First, the clinician should observe whether nasality or nasal emissions are present in the child's speech. This is important because the face mask is covering the child's nose and mouth, and, although stimuli are

selected to optimize oral airflow, nasal airflow will be captured. If nasality or nasal emissions are noted, the clinician can examine the possibility of using a nose clip. If the child tolerates a nose clip, this will prevent air from escaping through the nose during speech production. However, many children will not tolerate wearing a nose clip [1], and, therefore, the clinician must take this into account when interpreting the results of the aerodynamic assessment. Second, the clinician should listen to the child's articulation for production of the voiceless bilabial plosive /p/. If the child displays error production of /p/ with respect to place, manner, or voicing during running speech, the clinician must ensure that the child can accurately produce a voiceless sound with full bilabial occlusion during the assessment tasks. Lastly, the clinician should consider if the child's general respiratory patterns are adequate to support five /pæ/ productions on a single breath. Fewer syllable productions on a single breath may be acceptable, but a minimum of three syllables is needed for analysis, and analysis should only be performed on the middle productions, ignoring the first and last to control for initiation or termination effects [6].

Noninstrumental Measures

When an instrumental aerodynamic assessment is not practical or successful, noninstrumental assessment of maximum phonation durations may provide some information on the efficiency of the laryngeal system. One task used to measure maximum phonation involves asking the child to take a breath and hold out a vowel for as long as she or he can and timing the sustained vowel production. Children often need external cues to achieve phonation durations that accurately reflect their capacity. Cues may be explicit, such as having the child trace a line with his or her finger during phonation, moving a toy down a path, or showing the child a stopwatch. There is some evidence that children will produce longer phonation times if they are given encouragement and/or coaching during the production [9]. Additionally, repetitions may be helpful to elicit the best performance [10], and an average of multiple productions may increase the reliability of the measure [11].

Another informal method of examining maximum phonation durations is the s/z ratio in which the child holds out an /s/ for as long as they can and a /z/ for a long as they can. The clinician should calculate the ratio by dividing the /s/ production time by the /z/ production time. First proposed by Boone [12], this measure provides information on sustaining sounds with and without vocal fold vibration, and the resulting ratio is suggested to yield a measure of laryngeal efficiency. Previous research has shown that vocally healthy children have an s/z ratio of around one [13–15]. However, large within- and between-subject variability across studies suggests that proposed norms should be interpreted with caution [13–17]. While the clinician may use maximum phonation duration measures to provide additional information about the child's aerodynamic function, conflicting findings suggest these values should not be used in isolation for assessment.

Conclusion

Familiarization to both the speech tasks and the equipment used during aerodynamic acquisitions are key elements to successful assessments for children of any age. Clinicians should also be well versed in the common pitfalls that can occur during aerodynamic acquisition, such as blockage of the oral catheter or a leak in the face mask. By understanding the purpose and method of assessment, coupled with observations of the child's natural speech patterns, the clinician can accurately collect and interpret aerodynamic signals.

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Perceptual Evaluation of Voice

12

Maia N. Braden and Sarah D. M. Blakeslee

Introduction

A voice disorder, by definition, exists when the voice of an individual differs from the voices of similar age, gender, geographic location, and cultural group in terms of pitch, quality, or loudness [1]. As such, the clinician's ear is the gold standard for identifying, quantifying, and describing a voice disorder. Clinicians use perceptual evaluation not only in the initial and subsequent evaluations but also as an ongoing assessment of the effectiveness of therapy throughout the therapeutic process. Perceptual ratings are by nature subjective and dependent on the culture, location, age, and gender of the speaker as well as the listener.

There are additional challenges when applying perceptual ratings to children, as children may not be as consistent in their productions or as willing to participate in a task as adults, and listeners may lack a clear sense of what is "normal" in children's voices. Perceptual ratings of voice can be completed based on sustained phonation, repeated sentences, reading, and connected speech. When the stimuli are standardized, this allows for more consistent ratings across speakers, raters, and serial visits. Rating systems in use include descriptors such as "mild, moderate, and severe," equal appearing interval scales, visual analog scales, direct magnitude estimation, or sort and rate systems, to name a few. The rating systems most commonly used in clinical evaluation of voice disorders are the grade, roughness, breathiness, asthenia, and strain (GRBAS) scale [2] and the Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) [3, 4].

Perceptual Features of Children's Voices

When you listen to a child's voice, you can tell that it is a child and not an adult speaking. Why is this? There are perceptual differences between children's and adults' voices. The most obvious is pitch, with mean speaking fundamental frequency declining with age in both boys and girls [5]. However, pitch is not the only factor that makes a child sound like a child. As discussed in Chap. 6, children have shorter vocal folds than adults, resulting in higher pitch [6]. They have incomplete differentiation of the vocal fold

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layered structure, although the impact this has on perceptual characteristics of voice is not well understood [7]. The larynx is also positioned more superiorly, resulting in a shorter resonating chamber and different formant frequencies than adults, which should also result in perceptual differences. Time-based perturbation measures (jitter and shimmer) vary with age and are higher in children than adults [8]. Children use a higher percentage of their vital capacity during speech and have higher tracheal pressures during speech than adults [9] although it is not clear how this translates to auditory-perceptual characteristics in the voice. Lopes et al. [10] found a correlation between shimmer and listener ratings of breathiness, roughness, and overall grade of severity of dysphonia. Additionally, normal glottic configuration in children features a posterior gap, [11] which may be conjectured to produce a breathier voice quality even in children with normal voices.

Limitations of the Perceptual Evaluation

As important as perceptual evaluation is, it has limitations, including variability in the child's voice, inconsistency or unreliability of perceptual ratings of any kind, and differences in definition of aspects of voice and in definitions of severity. Throughout history, hundreds of words have been used to describe voice: "nasal, hoarse, squeaky, creaky, harsh, rough, breathy, airy, rich, resonant," to name only a few [2, 3, 12, 13]. These have been quantified in different ways, which can be more or less precise, "a little rough" or "9/10 in harshness" or "4/5 loudness." Without shared scales, terminology, and understandings of what is normal and how to quantify severity, these terms are as useful as describing a color by saying "that bluish color that is kind of like green and yellow too." As such, perceptual ratings are necessary but troublesome when attempting to make them useful and reliable across clinics, patients, clinicians, and disorders. For perceptual ratings to be reliable across clinicians, institutions, and patients, there are several assumptions that must be accepted. These assumptions apply to all perceptual ratings of speech and are summarized by Kent [14]. First, we must have shared vocabulary and definitions of vocal characteristics such as "hoarseness," "breathiness," "roughness," "strain," and other labels. Second, we have to use the same descriptors and scale values. Third, we need to be able to reliably isolate perceptual features, and fourth, the differences in ratings between judges need to be smaller than the differences needed to quantify severity of disorder or change in status [14]. Unfortunately, these assumptions are not always true. Until relatively recently, there was a lack of consistency in the terminology used to describe disordered voice, although the adoption of the CAPE-V in clinical settings lays out a consistent scale and terminology [15]. Studies have shown that perceptual features of voice are not reliably isolated by clinicians – for example, judgements of pitch have been found to be influenced by roughness [16]. Perceptual evaluation of voice quality can be influenced by articulatory context, visual stimuli, and even information about the medical diagnosis [14, 16–20]. It is not clear if this is more challenging in children than adults. We do not have any clear definitions of what constitutes a significant difference to quantify severity of disorder or to validate a change in status.

Few studies have examined inter- or intra-rater reliability in evaluation of pediatric voices. Kelchner et al. [21] found moderate to strong inter-rater agreement in rating of overall severity, roughness, and breathiness using the CAPE-V, and strong intra-rater reliability, but poor inter-rater reliability in ratings of strain in a population of children status post-laryngotracheal reconstruction [22].

Listener training, use of anchors, and using a rank and sort method of rating voices have been shown to improve inter-rater reliability. Rater training with anchors was found to increase inter-rater reliability in evaluation of dysphonic voices, and that training using synthesized anchors was more effective than natural voices [23]. Listener training can certainly be done in the clinical setting, but anchors and more involved methods of

rank and sort rating are typically more feasible in research.

Clinically, perceptual evaluation of children can be challenging because they may not be as consistent in their productions as adults, they may have difficulty either reading or producing standard stimuli, it may be challenging to get a representative sample of connected speech, and they may simply choose not to do as they are asked. It is ideal to have a set of stimuli that is consistent across patients and across evaluations with the same patient. If this cannot be accomplished, we recommend attempting to get at least a representative sample of conversational speech, or speech in play.

Perceptual Characteristics of Voice

Historically, a wide variety of terms have been used in perceptual analysis of voice. When looking at methods of perceptual evaluation in the literature, several terms tend to be the most frequently used and easiest to define and in most cases can be partially linked with a physical or acoustic correlate [3, 18]. However, descriptors of vocal quality are multidimensional in nature, and it is extremely difficult, if not impossible, to completely isolate individual parameters of voice quality. In spite of these challenges, perceptual evaluation remains a cornerstone of voice evaluation.

Two of the most basic parameters used in perceptual voice analysis are pitch and loudness. *Pitch* refers to the perceived highness or lowness of the voice. It is the perceptual correlate of fundamental frequency as measured in Hertz, which as discussed earlier varies across age and gender. *Loudness*, on the other hand, is the perceptual correlate of sound intensity, measured in decibels, and is typically expressed as a range from too soft to appropriate to too loud. Three other parameters – roughness, breathiness, and strain – make up the other most commonly used terms in perceptual voice analysis. *Roughness* refers to the degree to which the voice is smooth/clear versus gravelly. In general terms, it correlates with the periodicity versus irregularity of vocal fold vibration. *Breathiness* refers to the degree to which excess airflow or "hiss" is detected in a person's speaking voice and roughly correlates with the degree of glottal competence versus incompetence. Is extra air "leaking" during voice production? *Strain* relates to the perception of increased muscle effort or "pushing" associated with voice production. It can be thought of as the perceptual correlate of pressed phonation, or hyperadduction of the vocal folds.

A variety of additional descriptors can also be used to help describe a person's voice. Most commonly, these include asthenia (weakness), glottal fry (low pitch pulsations of voicing or "creaking" – can be perceptually acceptable in certain age/gender/culture groups), tremor (regular oscillations in pitch), and diplophonia (perception of two pitches being produced simultaneously). Other descriptors can also be used, such as presence of pitch breaks or "cracks," aphonic breaks or any periods of aphonia or near-total aphonia, and descriptions regarding vocal register (chest/modal register versus head voice or falsetto). These are not always reported with a measurable rating scale but may also aid the overall description of a person's voice as perceived by the clinician.

As a final note, statements regarding oral/nasal resonance balance are sometimes included in an overall perceptual description of a person's speech, but these are phenomena of the resonance system and not voice, and thus a detailed discussion of this is beyond the scope of this chapter. Briefly, hypernasality refers to excess nasal resonance (relating typically to velopharyngeal incompetence), and hyponasality refers to too little nasal resonance (resulting in a person sounding "stuffy" or congested). Similarly, articulation and language skills are of course separate from voice quality. In a pediatric population, however, even if the primary focus is on voice, these are important parameters to consider as part of an overall evaluation and may warrant further formal testing procedures if they appear to be problematic.

Standard Methods of Perceptual Evaluation of Voice in Children

There have been a wide variety of rating tools used by clinicians and described in the literature to help formalize and standardize the perceptual analysis. The two most commonly used, particularly in the United States, are the GRBAS and the Consensus Auditory-Perceptual Evaluation of Voice, or CAPE-V [2–4]. The GRBAS was first described by Hirano in 1981 [2] and is an acronym for five parameters to be rated: grade, roughness, breathiness, asthenia, and strain. For each of these parameters, the clinician rates the patient's voice on an equal appearing interval scale from 0 to 3, 0 being normal, 1 mild, 2 moderate, and 3 severe. This scale is easy to use and relatively quick but can limit the ability to reflect change over time. For example, if a patient is rated as a 3 or "severe" for any of the parameters, but becomes worse at some point, there is no way to reflect this within the 4-point scale. Additionally, there are no standardized stimuli for administration of this scale, and perceptual ratings can change based on context, length of utterance, vowel, and whether the speaker is sustaining vowels, reading, repeating, or speaking spontaneously [14, 15, 19, 20].

To provide a standardized way of evaluating voices perceptually, work on the Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) began in 2002 with a consensus meeting of speech-language pathologists, speech scientists, experts in psychoacoustics, and experts in perception [3]. After extensive discussion and development work, the CAPE-V instrument was developed. The full form is available for download for clinical use through the American Speech Language Pathology and Hearing Association website [24]. Rather than an ordinal scale, it implements a visual analog scale, with 100 mm lines for each parameter to be rated. Clinicians make a hash mark on the line to indicate their judgment, and a ruler is used to measure in mm where this mark falls from 0 to 100, 0 indicating normal, higher values indicating more severely disordered. The parameters to be rated include overall severity, roughness, breathiness, strain, pitch, and loudness. Several blank lines are also included so that other parameters can be rated if desired (e.g., tremor). The clinician can also indicate with each parameter whether it is consistent or inconsistent and note whether resonance is normal or not. While the results are reported as a number out of 100, there are also general visual guidelines indicating where mild, moderate, and severe fall on the scale.

While the GRBAS has no specific tasks to be completed upon which to base judgments, the CAPE-V has three tasks for the patient to complete – sustained vowels (/ɑ/ and /i/, 3–5 s each), six phonetically distinct sentences, and a sample of spontaneous speech in response to "Tell me about your voice problem." The CAPE-V is more detailed and takes longer to administer and score. At least in part because it uses 100-point scales, it is thought to be more responsive to small changes in voice [25]. Both the CAPE-V and GRBAS have been found to be reliable and valid measures of perceptual voice quality [15, 25], but as noted earlier in this chapter, it is important to consider the factors that affect reliability and validity of any perceptual rating tool and control this with use of standardized procedures, training, and use of anchors when needed.

We have discussed the importance of standardizing procedures and minimizing variability as much as possible to aid reliable perceptual assessment, but when these rating scales are applied to a pediatric population, this can be challenging, and modifications at times need to be made. It can be difficult for young children to sustain vowels for more than a few seconds at a time. Providing models and using child-friendly explanations and visual cueing can be helpful, as can encouraging them with competition. ("See if you can make your voice go all the way to the end of the screen!"). Some of these techniques are described more thoroughly in the acoustic and aerodynamic assessment chapters.

For sentence-level stimuli, ideally an older child could read the same standardized sentences

an adult would produce. If the child is not a fluent reader, however, he or she may need to repeat each sentence after the clinician. While the standardized sentences used with adults are ideal, their linguistic complexity may be too difficult for younger children even when provided with a model to repeat. For these children, we have developed a list of similar more simple sentences that aim to preserve the same phonetic makeup of the original sentences. These include "Harry has a hat," "We were away," "We eat eggs," "Mama made muffins," and "Pet the puppy." These are typically simple enough for even young 3–4-year-old children to repeat successfully. These modified sentences were not developed by the consensus committee and are not an official part of the CAPE-V. As with any standardized instrument, if used in a non-standardized way, this should be noted and taken into account.

Eliciting conversational speech samples can have its own challenges. While we know young children with dysphonia often have more awareness about their voice problem than they are given credit for, they may have difficulty answering a prompt such as "Tell me about your voice problem." We have typically chosen to elicit speech with a more child-friendly prompt such as "Tell me about your favorite vacation" or "Tell me about your favorite movie." Using visually interesting stimuli such as the Cookie Theft Picture [26] or the updated Cookie Theft Picture [27] is another way to help elicit additional speech samples. Sometimes despite a clinician's best efforts, however, a young child may be very reticent to engage in any sort of conversation given the unfamiliar and sometimes anxiety-provoking setting. Sometimes the speech one is able to elicit may not be particularly representative of a child's typical conversational voice, particularly in terms of loudness. In these cases, engaging the help of parents or caregivers in getting the child talking more naturally even a little can provide useful output upon which to base perceptual judgments. When judgments are based on a very limited speech sample, it is necessary to note this in one's documentation.

Emerging and Evolving Practices

Emerging methods and technologies in perceptual voice evaluation are focused on improving inter- and intra-rater reliability, isolating perceptual features, and providing complementary information that may assist in the accuracy or reliability of evaluation. For example, providing a spectrogram of the voice in conjunction with the recording has been demonstrated to increase inter-rater reliability [28]. Synthesized voices have been used to better isolate individual vocal parameters, and better understand both the auditory perceptual characteristics and the acoustic correlates, and to increase reliability in ratings [29, 30]. The use of synthesized voices can provide anchors for varying severity and different parameters of voice and allow clinicians and researchers to isolate the salient characteristics. Currently, acoustic, aerodynamic, and perceptual assessment techniques are complementary, but as voice recognition and analysis techniques continue to develop, we may see more overlap and ability to more objectively quantify what we hear perceptually.

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Health-Related Quality of Life in Pediatric Dysphonia

13

Shannon M. Theis and Nadine P. Connor

Introduction

Childhood dysphonia is a broad condition that can be difficult to investigate and quantify. As clinicians and researchers, we often consider the severity and duration of a child's voice disturbance and then extrapolate the degree of impact that the disturbance may have on the child's ability to communicate effectively. While the field of pediatric dysphonia has advanced by providing objective data via acoustic and aerodynamic measures, it is unlikely that we are able to fully understand the way in which dysphonia affects children's lives without exploring health-related quality of life. Too often in childhood dysphonia, clinical judgments are made regarding the effect of a pediatric voice disorder through a caregiver report rather than asking the child directly. This continues to be problematic because the self-evaluation process is important in establishing therapy goals and measuring subsequent intervention outcomes.

Quality of Life Versus Health-Related Quality of Life

Health-related quality of life has become recognized as an important outcome measure in healthcare research, clinical trials, and quality assurance [1]. Long ago, the World Health Organization (1948) defined health as "a state of complete physical, mental and social well-being, and not merely an absence of disease." The terms quality of life and health-related quality of life are often used interchangeably in the literature, but an important, albeit subtle, distinction must be made between the terms. The term quality of life refers to a broader concept that involves numerous aspects of an individual's life, such as housing options, quality of the environment, and job satisfaction [2, 3]. The term health-related quality of life was introduced to distinguish outcomes that are relevant to health research versus the overall concepts of subjective well-being and life satisfaction associated with the term quality of life [4]. As such, health-related quality of life relates to the measurement of the dimensions of quality of life that are directly relevant to clinical interventions [5, 6]. Although there is no universally agreed-upon definition, health-related quality of life is typically characterized as a

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"multidimensional concept that includes physical, social, emotional, and role functioning" [2].

Types of Instruments

Health-related quality of life instruments can provide different types of information, such as the effectiveness or efficacy of treatment protocols, treatment response among individuals, and patient comparison at different disease stages [7]. Instruments, or questionnaires, can be divided into two categories: generic and disease- or disorder-specific health measurements [8]. Generic instruments focus on overall health measurement and are typically developed for broad clinical use with a variety of diseases and conditions [9, 10]. Disease- or disorder-specific instruments, on the other hand, are developed to measure specific problems within an illness, population, or treatment group [7].

Generic health-related quality of life measures allow for comparisons across disease/disorder groups, interventions, and severity of illness. Generic measures allow for more generalizability when assessing health status; however, they may not be specific or relevant to the disorder being investigated [11]. In contrast, disease-specific instruments are more specific, but then lose generalizability. The lack of generalizability can affect the instrument's ability to be used in a variety of clinical trials. However, disease- or disorder-specific instruments are more responsive to even small changes in health status when assessing treatment outcomes and for use in clinical trials, as well as comparing alternative treatments [12]. Since generic and disease-specific measures of health-related quality of life are both subject to weaknesses and strengths, the type of instrument used should be dependent on the outcomes question being examined. In the case of pediatric dysphonia, a voice-related quality of life measure should be used for assessing intervention and treatment outcomes.

Proxy Report

There are circumstances in health-related quality of life research in which a patient is not able to independently answer questions related to their health status. In these circumstances, a person close to the patient, or a proxy, is used to provide information regarding health-related quality of life. Proxy reports are often used in health-related quality of life instruments for a variety of reasons, such as decreased cognitive functioning, or the severity of illness does not allow for patient report. Proxies can include physician perspective, close family relative, or spouse/partner viewpoint. If a proxy report is used, it should be assessed for any potential bias and is most accurate when the perspective is taken from someone very close to the patient [13]. Physician reports have shown less agreement [14].

Proxies are the most common form of report for pediatric instruments and can consist of parental assessment, teacher viewpoint, or physician perspective. While adult health status measures often use proxies instead of patient report out of necessity, the use of parent proxies as the sole measure of a child's health status is flawed for several reasons. First, parents are often used as proxy informants in pediatric quality of life research because it is assumed that parents know their children best and would be able to accurately report their health status [15]. However, parents are not with their children in every setting, especially as they continue to mature and become more independent, which may make the parent report incomplete [7, 16]. Second, parent perspective can be influenced by their own anxiety regarding their child's illness or disorder and can negatively affect ratings of health-related quality of life [7, 17]. Third, while proxy instruments are frequently used in pediatric health-related quality of life research, proxy reports have been shown to be inconsistent when compared to patient report, particularly on certain domains. In a variety of studies that involved parent proxy reports for child health-related quality

of life, concordance was higher for items that were related to physical and functional domains rather than emotional/social domains [18]. Therefore, parents were able to accurately rate more observable external behaviors, such as difficulty climbing stairs or outward symptoms of their illness, versus more subjective internal behaviors including feelings and emotions [19]. Finally, and most importantly, parent proxy perspective is commonly used in pediatric health-related quality of life instruments due to the questionable validity of children as informants of their own health status because of cognition and developmental issues.

Children as Health Reporters

While the field of health-related quality of life in the adult population has evolved as a respected field of research over the years, pediatric outcomes research has significantly lagged behind [20–22]. A recent development in the field of pediatric health research is the increasingly accepted view that health-related quality of life instruments and questionnaires must be developed for different age groups and the items and domains incorporated should reflect age-appropriate issues [7, 23, 24]. However, this process is complex, labor-intensive, and extremely time-consuming.

The field of pediatric health-related quality of life has developed from theoretical concepts that were initially developed for the adult population, but since children are not just "miniature" versions of adults, there are certain considerations that must be included [25]. Measures of pediatric health-related quality of life need to accommodate developmental changes and domains that are not consistent with those used in adult measures [26]. Furthermore, children's understanding of their health is different from adult's views [12], and a child's concept of health has been shown to change with age [27, 28].

Adult measures typically include domains specific to adulthood, such as economic status, job role performance, and social interactions and relationships [7], which is not typically appropriate for pediatric measures. A specific obstacle that must be addressed are the methodological considerations related to progressive cognitive and emotional development that occurs throughout childhood [25]. The items and domains represented on a pediatric health-related quality of life measure must be appropriate for the different ages and developmental levels of the population that is being investigated [12, 29, 30]. As childhood is a dynamic process, an outcome measure that may be appropriate for one age group or developmental stage may not be appropriate for another [26]. Accordingly, the instrument must be sensitive to the developmental issues of the age group for which it is intended.

To establish sensitivity to developmental stages of childhood, it is imperative that children's perspectives are considered when developing health-related quality of life instruments. Children as young as 7 years of age have been shown to reliably complete health-related quality of life questionnaires [31]. The authors concluded that young children were consistent and accurate in their understanding of both the questions and response options [32]. Therefore, it is critical that child report be included whenever feasible while measuring health-related quality of life, as well developing instruments that represent children's opinions and views regarding their health in a developmental and age-appropriate manner.

Current Approaches for Assessing Voice-Related Quality of Life in Children

Development of adequate methodology for assessing how dysphonia affects the lives of children is critical for providing insight into the depth of disability that a voice disorder may impose on

a child and in assessing how well our treatments address these issues. However, these critical concerns remain largely unanswered in pediatric voice pathology.

Several patient-reported outcome measures (PROMs) for assessment of voice-related quality of life have been developed and are currently used with children (Table 13.1). These measures aim to assess the manner in which a voice disorder affects the lives of children. In general, these instruments were developed for all children with dysphonia across the childhood age range (ages 2–18), although there was some variation in the ages tested in development of these instruments. Some of the instruments have been translated into multiple languages [33–37].

As described in a systematic review on the topic of voice-related quality of life and its measurement [38], instrument development must start by talking with patients. To develop an instrument concerning children's voices, initial input must come from children. As a first step, interview or focus group information must be obtained and incorporated into instrument design and content. Rigorous qualitative research methods, using grounded theory analysis and coding, should be used to discover themes that apply to children who have life experiences with a voice disorder. These interviews or focus groups must be performed by examiners trained in these methodologies. In contrast to trained interviewers and focus group facilitators, healthcare providers are trained to sift, combine, remove irrelevant information, and summarize from a patient's clinical and oral histories to form a diagnosis and treatment plan. While this approach works very well in the clinic, the risk in using these clinical history-taking methods in qualitative research is one of filtering a patient's experience through the lens of the practitioner and not forming a true picture of the information as conveyed by the patient. Thus, it is rarely acceptable to replace a rigorous interview or focus group process with retrospective review of medical histories or chart notes in this initial process of instrument development. It is even worse to start with an instrument developed for a different population and apply it elsewhere.

Table 13.1 Instruments used to assess voice-related quality of life patient-reported outcomes (PROMs) in children

Instrument	Participants	Reporter	Instrument development
Pediatric Voice Handicap Index (pVHI) [39]	Children aged 4–21 years old with laryngeal airway concerns and healthy children	Caregiver (parent)	23 items adapted from an adult instrument, the Voice Handicap Index [40]
Pediatric Voice Outcomes Survey (PVOS) [41, 42]	Children aged 2–18 with tracheostomies or decannulation; 385 children with ORL conditions not limited to voice	Caregiver (parent)	4 items adapted from an adult instrument, Voice Outcomes Survey [43]
Pediatric Voice-Related Quality of Life (pVRQOL) [44]	Children aged 2–18 who visited ORL office with all conditions, not limited to voice	Caregiver (parent)	10 items adapted from an adult instrument, Voice-Related Quality of Life (VRQOL) [45]
Pediatric Voice-Related Quality of Life (pVRQOL) [46]	Children aged 3–15 who visited a pediatric voice clinic	Child and caregiver (parent)	10 items adapted from an adult instrument; children completed a child-adapted version of the pVRQOL
Voice Handicap Index (VHI) [40, 47]	Children aged 6–12 with dysphonia due to vocal fold nodules, edema, paralysis	Child	30 items used directly from an adult instrument; VHI [40]. Some items are not appropriate for children
Pediatric Voice Symptom Questionnaire (PVSQ) [48]	Children aged 9–13 (French)	Child and caregiver (parent)	29 items developed from interview study [49] and further work to assess item comprehension. Separate parent and child versions
Children's Voice Handicap Index (CVHI) and CVHI-P for parent report [50, 51]	Children aged 8–14 (Italian; English translation)	Child and caregiver (parent)	10 items from adult measure, VHI-10; then modified based on child interviews [52]

When themes are discovered in patient interview transcripts through systematic coding, instrument items can be developed that target those themes [39–52]. Appropriate triangulation testing can be used to determine if items are comprehensible to children and written in ways that define those initial themes. The second and third stages of instrument development involve field testing the instrument and psychometric analyses [38]. Unfortunately, most of the PROMs used in the area of voice disorders did not follow these standardly accepted instrument development methodologies [38].

When direct input from children with voice disorders is not part of the development process, the result is a violation in the first step of instrument validation. With the exception of the PVSQ (French) [48], all pediatric voice-related PROMs are derived from adult instruments. That is, development of these instruments began with adult questionnaires that were then revised to reference a child's voice rather than an adult's voice. For example, the pVOS [41, 42], pVHI [39], and pVRQOL [44] were developed by changing item referents from an adult patient's voice (e.g., "your voice") to that of a child (e.g., "your child's voice"). The CVHI [50, 51] used an adult instrument as a starting point, then interviewed children, and revised items. As such, while these PROMs had the goal of assessing the child's perspective, they were unfortunately not developed using methods that allowed realization of this goal.

As shown in Table 13.1, most currently available pediatric voice PROMs use proxy administration. As discussed earlier in this chapter, parent or caregiver proxy assumes that a more accurate report may be obtained from a parent or caregiver than from the child. Other assumptions specific to voice are that children may have a limited awareness of their voice disorders, that parents are knowledgeable about the entirety of their child's perceptions regarding the voice disorder, and that children and adults share a similar framework for the concept of quality of life and the manner in which a voice disorder contributes to quality of life. We know that these things are not true [12, 49, 52]. As such, proxy administration may be biased toward the views of the respondent and can be problematic, especially when items reflect internal states, such as thoughts, emotions, and experiences that occur independently from the parent/caregiver. It is not surprising that in the area of pediatric voice disorders, research has shown that parents and children are often not in congruence regarding the child's voice-related quality of life [49, 52]. Thus, asking children to provide responses about their own voices is paramount. Children as young as 6 years old are aware of their voice disorder and capable of discussing their life experiences regarding their voices [49, 52]. Therefore, the use of parent/caregiver proxy responding for children 6 years old or older is poorly justified. As stated by Branski et al. (2010) [38], "Manipulation of instruments to apply to other populations for proxy application violates the fundamental tenets of instrument development." The need for more instruments that emphasize child reporting is clear.

Currently available pediatric PROM instruments in common use assert that they are reliable and valid [38, 39, 41, 42, 44]. Reliability reflects the degree to which a measure, items within a measure, instrument, or tool provides a stable or repeatable value [53]. For instance, a questionnaire administered twice should have values that are relatively similar to be considered reliable. Additionally, items within a questionnaire that measure similar constructs should be correlated (internal consistency). Validity can be defined as the degree to which an entity, in this case a pediatric voice PROM, accurately reflects an underlying truth. Therefore, a valid pediatric voice PROM would faithfully reflect the viewpoints of children with voice disorders. There are multiple types of validity; a complete list and definitions of different components of validity are beyond the scope of this chapter (see Kimberlin, 2009, for a review) [54]. Validity can be complex to determine [55] and cannot be claimed solely with statistical testing in the absence of appropriate instrument development. In this vein, it has been stated that validity and reliability best refer to *data*, and not to *measures* [55]. An example given by Sechrest [55] is a tape measure – this is a valid and reliable instrument for measuring the length of a piece of lumber only

inasmuch as its use conforms to generally accepted principles of how to use a tape measure. As mentioned previously, validity of an instrument depends upon how it was designed, how it is administered, and whether the population tested is reflective of the target population for use of the instrument, in addition to statistical analyses and psychometrics. Several of the pediatric voice PROMs in current clinical use were validated, using parent proxy, with children who had complex airway disturbances and tracheostomies. While these children may also have had voice disorders, the voice-related quality of life concerns may not be typical of children with more commonly occurring benign vocal fold lesions. Further, voice-related quality of life concerns may have been part of a larger constellation of illness that arguably could have more grossly affected parent perceptions of quality of life. Thus, while statistical tests may reassure us that a pediatric voice-related PROM available for use is valid, and claims of validity are made in published papers, inadequate development protocols, proxy administrations, and other practices may not be consistent with this notion.

The limitations in current approaches for measuring pediatric voice-related quality of life described in this chapter are echoed in the larger context of health-related quality of life assessment in children [26]. In summary, the major limitations include faulty development protocols that do not include qualitative analysis of direct interviews or focus groups with children, use of an adult instrument revised for children, and proxy administration. It must also be acknowledged that children can view the concept of health-related quality of life differently throughout the rapidly changing childhood years. Therefore, thought should be given to the reasonableness of collapsing data from the wide childhood age range into a single version of an instrument [26]. This issue has not been considered in any of the currently available pediatric voice PROMs shown in Table 13.1. Research has shown that children's perceptions of their voice disorders differ across the age range, where younger children are primarily focused on physical variables and adolescents add concerns around emotional factors to the previously established physical concerns [52]. These flaws in instrument development result not only in faulty assumptions regarding pediatric PROMs and inadequate tracking of treatment effects, but do not easily allow study of other interesting aspects of voice-related quality of life in children. With a properly designed and validated pediatric voice PROM instrument, it is possible to study how a voice disorder affects children's function in educational settings and within a family context. In addition, it is possible to examine how developmental changes/maturation affect the manner in which a voice disorder is perceived and how particular dynamic variables, such as emerging independence and social relationships, influence voice outcomes. As pediatric voice PROMs are refined and developed, answers to these important questions about the lives of children will be answered.

Conclusion

Voice disorders are common in the pediatric population [56, 57] and can have lasting, negative effects on communication, social interactions, scholastic performance, and self-esteem (Fig. 13.1) [58, 59]. The science of pediatric voice has not yet provided a set of standardized and meaningful metrics that can define or predict functional outcomes from treatment. Furthermore, we do not have a standard set of agreed-upon measures for assessing severity of a pediatric voice disorder that are acquired the same way across clinics and clinicians; that are reliable, valid, and sensitive to change; and that directly reflect vocal function in natural environments. Valid, reliable, responsive, and child-centered patient-reported outcome measures (PROMs) could fill this void if the content is properly established and meaningful to children with dysphonia. Indeed, improving vocal function outside of the clinic and thus improving the lives of children constitute our main treatment goals. Instruments designed to validly and reliably reflect conceptual models of voice-related quality of life would be very useful for documenting therapeutic

Fig. 13.1 Direct quotes from children with dysphonia obtained during structured interviews. (Quotes from Connor et al. [52], with permission)

change. These instruments could be used to select or influence treatment methods, assess progress through therapy, gauge severity of dysphonia from the child's perspective, and provide a basis for counseling prior to or throughout treatment.

Our view is that health-related quality of life instruments that capture the child's perspective on their voice disorder can, and should, be used in conjunction with objective instrumental measures as additional and important clinical endpoints. Thus, we encourage the further development of valid and reliable child-centered PROMs that follow generally accepted methods of instrument design [38]. We also encourage the use of direct reports from children wherever possible, rather than reliance solely on parent proxy. Until such measures are generally available, we encourage clinicians to perform structured clinical interviews with children that probe emotional, physical, and functional effects of the voice disorder on their daily lives.

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14 Endoscopic Evaluation of the Pediatric Larynx

Rita R. Patel, Stephen D. Romeo, Jessica Van Beek-King, and Maia N. Braden

Overview

Visualization of the larynx is necessary to evaluate structure and function, identify pathology, and plan treatment. There are different methods of evaluating the larynx, and each has benefits and limitations. Flexible endoscopy under halogen light can be performed in the clinic on nearly all children and provides an excellent view of general structure and mobility at the cricoarytenoid joints. Rigid or flexible stroboscopy provides more in-depth evaluation of the vibratory properties of the vocal folds, closure pattern, and any vocal fold lesions. High-speed laryngeal visualization has the advantage of being able to capture vibratory properties of aperiodic or chaotic vibration.

Flexible Laryngoscopy

Flexible laryngoscopy is a key technique in the evaluation of the pediatric larynx. While other techniques exist for evaluating the physiology of the larynx, flexible laryngoscopy is a useful tool for evaluation of anatomical features. There are many strengths unique to flexible laryngoscopy: it is cost-effective, portable, fast, and adaptable to a child of any age. It is often the first instrumented step in the evaluation of the pediatric larynx and can guide further work-up and treatment.

There are alternatives to evaluate the pediatric larynx with indirect mirror exam being one example. Mirror exam has many of the benefits of flexible laryngoscopy, as it is cheap, fast, and portable. However, it cannot be recorded and

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requires a cooperative patient that can be coached through the exam, restricting its use to teenagers. Another option is direct laryngoscopy. Though it does provide an excellent exam and can be performed on children of any age, it requires a general anesthetic in the operating room and does not provide the dynamic information provided by flexible laryngoscopy in the awake patient.

Procedure Details

Flexible endoscopes come in a broad range. Some differences are subtle, such as using an eyepiece versus a separate video tower or a pediatric versus an adult sized endoscope. A more critical distinction, perhaps, is a distal chip endoscope contrasted with a fiber-optic endoscope. One trade-off here is the potential addition of a working channel. The working channel yields a bigger scope, which can be a significant challenge in the pediatric population. Although distal chip endoscopes provide better quality images, they have been found to have similar diagnostic accuracy compared with fiber-optic laryngoscopes [1]. Some studies even suggest that fiber-optic scopes are more accurate [2]. However, with improvements in technology have come smaller diameter distal chip endoscopes, allowing for improved image quality and comfort for smaller children. Fiber-optic and distal chip endoscopes are pictured in Figs. 14.1 and 14.2.

Fig. 14.1 Flexible fiber-optic pediatric endoscope

Fig. 14.2 Flexible distal chip pediatric endoscope

Another consideration in preparation for laryngoscopy is the use of an intranasal anesthetic and/or decongestant. Using a combination spray can be beneficial to examiner and patient: it decreases pain, decreases duration of the exam, and provides a superior view [3]. After using the spray, it is best to wait several minutes prior to the exam to allow maximal benefit. Anesthetics should be used with caution, however, as they can have unwanted consequences depending on the indication for the endoscopic exam. For example, topical anesthetics are known to increase signs of laryngomalacia [4] and may influence and swallow function, although findings on this have been mixed in adults and not extensively studied in children [5–7].

There are several other non-anesthetic considerations that may facilitate a flexible endoscopic exam. These vary by patient age. For a neonate, infant, or toddler, swaddling can help. For a preschool or school-aged child, distracting them during the exam or coaching them through it (if they are amenable to that) may be helpful. Finally, an adolescent should be able to participate more actively in breathing and relaxing techniques. Positioning the patient such that they are sitting up straight, leaning forward, and slightly extending their neck (assuming the sniffing position) is also important.

The steps to performing flexible laryngoscopy are as follows:

1. Administer topical anesthetic and position patient as detailed above.

2. Insert the endoscope along the nasal floor, maintaining a straight endoscope to allow for precise manipulation.
3. Once the posterior nasopharynx is encountered, instruct the patient to breathe through their nose (if they are able to follow instructions) to allow passage into the oropharynx.
4. In the oropharynx, have the patient protrude their tongue to allow for better assessment of the tongue base and valleculae.
5. Advance to the hypopharynx. Instruct the patient to insufflate their cheeks to provide a better examination of the pyriform sinuses.
6. Assess the true and false vocal folds. Have the patient produce a sustained /i/ to evaluate mobility. Spontaneous crying will also suffice for this purpose. Instruct the patient to sniff in to elicit posterior cricoarytenoid muscle contraction and consequent vocal fold abduction.
7. Advance the endoscope to the level of the vocal folds to examine the subglottis.
8. Withdraw the endoscope slowly, evaluating the adenoid pad, torus tubarius, and nasal cavity.

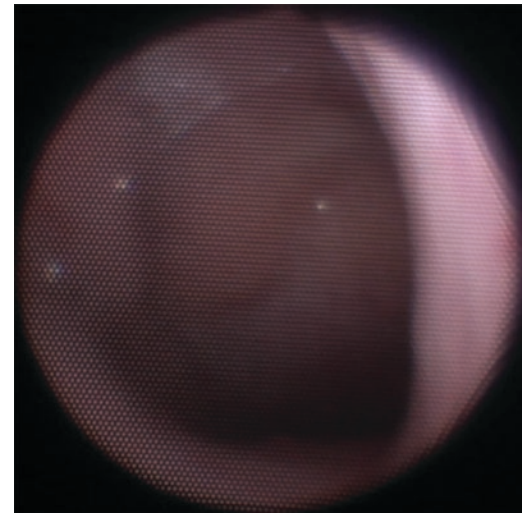


Fig. 14.3 Normal nasopharynx

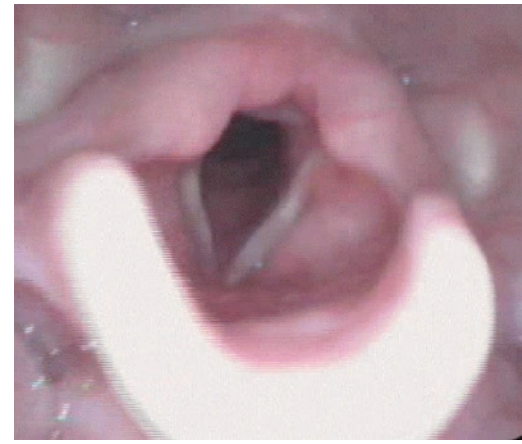


Fig. 14.4 Normal larynx and hypopharynx

Interpretation

More important than the technical ability required to perform flexible laryngoscopy is the interpretation of the exam. Recording the exam is ideal to allow revisiting and comparing across serial exams. The nasal cavity, nasopharynx, oropharynx, hypopharynx, and larynx can all contribute via different mechanisms to alter voice and swallow function.

In the nasal cavity, it is important to assess for mucosal edema as congestion can alter resonance (Fig. 14.3). As such, congestion should be noted, keeping in mind that this may be altered by the use of topical decongestant [8].

Moving posteriorly to the palate, palatal mobility and velopharyngeal competence should be evaluated. Velopharyngeal insufficiency can occur in the setting of various craniofacial syndromes or rarely status post-adenotonsillectomy [9, 10]. The adenoid pad should be examined to determine the amount of obstruction. Adenoid hypertrophy can also have effects on resonance in addition to the negative consequences on eustachian tube function [11].

In the oropharynx and hypopharynx, surface characteristics of the mucosa should be noted (e.g., cobblestoning and erythema) (Fig. 14.4). Posterior pharyngeal wall cobblestoning or lingual tonsillar hypertrophy can be signs of gastroesophageal reflux disease (GERD) [12]. Lingual tonsillar hypertrophy can also contribute to obstructive sleep apnea and is especially common in children with Down syndrome [13, 14]. In the hypopharynx, post-cricoid edema can be a

highly sensitive finding for GERD. Other less sensitive findings include hypopharyngeal cobblestoning and generalized erythema/edema [12]. The pyriform sinuses should be examined for pooling of secretions, penetration of secretions into the supraglottis, and other anatomic abnormalities such as a third branchial cleft sinus tract with an opening at the pyriform sinus.

The supraglottis, glottis, and subglottis should be evaluated from both a functional and anatomic/structural perspective. Using laryngomalacia as an example for evaluation of the supraglottic airway, it is a pathology with both functional (mucosa overlying the arytenoid cartilages prolapsing into the airway) and structural (foreshortened aryepiglottic folds and an omega-shaped epiglottis) components [15]. From a functional perspective, at the level of the glottis, there can be a range of pathologies including incomplete glottic closure, paradoxical vocal fold motion, or vocal fold paralysis. From a structural perspective, benign vocal fold lesions or laryngeal webs/atresia may be present. The subglottis is similar to other parts of the larynx where pathologies such as subglottic hemangiomas or stenosis can contribute to symptoms on the structural side and tracheomalacia can be a factor on the functional side.

Videostroboscopy

While endoscopy under halogen light can evaluate laryngeal structure, mobility, and tissues, and identify the presence or absence of lesions or masses, it lacks the ability to evaluate the vibratory characteristics, pliability of the vocal folds, and closure pattern. The rate of vibration of the vocal folds during phonation is much faster than the human eye can distinguish. Because of this, videostroboscopy allows the evaluator to assess vibratory features through essentially taking advantage of an optical illusion created by stroboscopic light.

Videostroboscopy to evaluate the larynx was well described by Bless, Hirano, and Feder in 1987 [16] and is part of the recommended protocols for instrumental evaluation of the voice set out by the American Speech-Language-Hearing Association (ASHA) expert panel [17]. Videostroboscopy is performed using either a rigid or flexible endoscope (fiber optic or distal chip) attached to a stroboscopic light source and a video recording system [16, 18]. Recommended specifications for equipment are detailed in the recommendations of the ASHA task force [17].

Stroboscopy takes advantage of two phenomena of visual perception: a perception of a flicker-free, uniformly illuminated background (occurring at greater than 50 Hz) and the perception of apparent motion when two objects are displayed in rapid succession [18, 19]. Stroboscopy works by producing a flickering light source at a slightly slower rate than the frequency of vocal fold vibration, so that what is seen is actually a sampling of images across multiple vocal fold vibratory cycles, rather than a single cycle. Due to the mentioned visual perceptual phenomena, the observer's eye perceives this as a continuous motion, allowing them to assess vibratory characteristics of the vocal folds. A minimum of three glottic cycles are needed to make valid perceptual judgements, with each cycle consisting of opening, closing, and closed phases [20]. Rating is not reliable with an aperiodic signal, as the light cannot sync appropriately to provide images that appear to be in immediate succession.

Instrumentation and Procedures

Stroboscopy can be performed with either a flexible or rigid endoscope. When performing rigid endoscopy, the child should be positioned in an upright position, leaning forward from their waist, with their chin up and tongue out. Very young children often have difficulties participating in rigid endoscopy, as it requires them to sit with their mouth open, their tongue out, and sustain phonation in this position. While we have sometimes had success in performing rigid stroboscopy as young as 3 years old, it is more usual for children age 5 or 6 to be able to participate. Flexible visualization requires less assistance from the child but can be more unpleasant for children because, as stated above, the passage through the nose can be slightly uncomfortable.

As with halogen endoscopy, topical anesthetic and decongestant can be applied and often make the procedure more comfortable. For young children sitting on a parent's lap can also be comforting, as well as allowing for the parent to assist with positioning. A laryngeal microphone is positioned on the child's neck so that the stroboscopic light can sync with their fundamental frequency. Flexible endoscopes can be either fiber optic or distal chip, and imaging advances in recent years have allowed for much smaller diameters of distal chip endoscopes. Improved image quality and a smaller diameter combine to improve both patient participation and the ability to interpret stroboscopy.

Parameters and tasks for recommended evaluation are detailed in the recommendations of the ASHA task force on instrumental voice evaluation [17]. Poburka and colleagues created and validated a rating system for both stroboscopic and high-speed video imaging of the larynx, which is included in Fig. 14.5 [21]. The following parameters should be assessed when performing a stroboscopic evaluation in order to fully assess laryngeal function [16, 17, 21, 22].

Parameters which can be assessed with halogen light only:

- Arytenoid mobility – degree of abduction and adduction, symmetry, and speed of movement
- Tissue appearance
- Supraglottic compression – degree of lateral or anteroposterior compression above the level of the vocal folds
- Free edge contour (rated during abduction, each vocal fold rated separately)

Parameters evaluated using stroboscopy:

- Glottal closure (rated during modal pitch) – the degree and configuration of glottic closure during closed phase
- Amplitude (rated during modal pitch, with each fold rated separately) – the magnitude of lateral movement of the vocal folds during vibration
- Mucosal wave (rated during modal pitch with each vocal fold rated separately) – the magnitude of movement of the mucosa during vibration
- Vertical level – the degree to which the vocal folds meet on the same plane (is one higher or lower than the other?)
- Adynamic segments – are there portions of the membranous vocal fold that do not vibrate?
- Phase closure – whether open or closed phase dominates or if it is equal
- Phase symmetry – the degree to which the vocal folds mirror each other during vibration
- Regularity/periodicity – the regularity of vibrations

Evaluation of these parameters is recommended during the following tasks: [17]

1. Rest breathing – three consecutive cycles
2. Laryngeal diadokokinesis (ʔiʔiʔiʔiʔiʔiʔi)
3. /i/ – sniff or /i/ quick inhale
4. Sustained /i/ at modal pitch, at least three stroboscopic cycles
5. Sustained /i/ at low and high pitch, at least three stroboscopic cycles of each
6. Sustained /i/ at varying loudness levels, at least three stroboscopic cycles of each
7. Any additional tasks individualized to the patient's voice complaints

Acquisition of these tasks relies heavily on the patient's willingness to participate, which can be more of a challenge with children than adults. Every attempt should be made to help the child feel comfortable and gain their participation. In pediatric clinics and hospitals, child life specialists can be extremely helpful in making children feel comfortable and relieving some of the potential fear and stress involved.

Interpretation and Evaluation

When an adequate sample can be obtained, stroboscopy has a high level of clinical utility in evaluating the vibratory function of the vocal

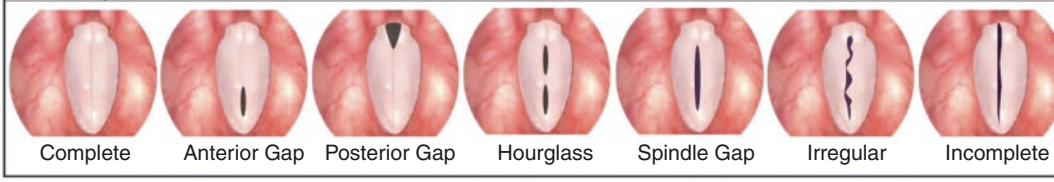
a

Voice-Vibratory Assessment with Laryngeal Imaging (VALI) - StroboScopy
Poburka, B., Patel, R., and Bless, D. 2016

Case #:		Confirm case #:

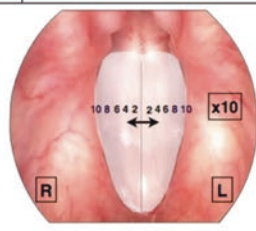
Glottal Closure

Definition:	Appearance of glottis during the most closed portion of the glottal cycle
Rating:	Rate at normal pitch and loudness.



Amplitude

Definition:	Magnitude of lateral movement of the vocal folds.
Rating:	Rate at point of contact.

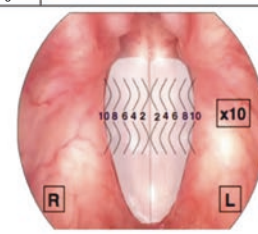


Right: _____ % Left: _____ %

F0: _____ Hz.

Mucosal Wave

Definition:	Magnitude of movement of the muc. membrane.
Rating:	Rate at normal pitch and loudness.

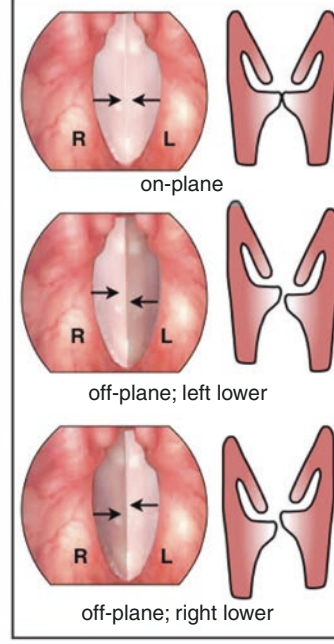


Right: _____ % Left: _____ %

F0: _____ (Hz.)

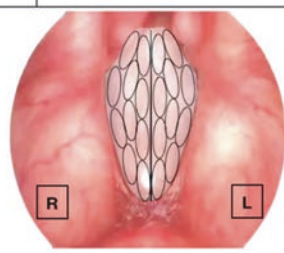
Vertical Level

Definition:	Do VFs meet on plane?
Rating:	Circle one



Non-vibrating Portion

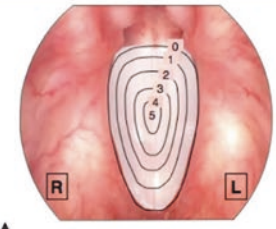
Definition:	Adynamic segments of tissue that appears stiff.
Rating:	Shade in affected area. Full ovals = 10% of TVF.



Right: _____ (ovals = 10%) Left: _____

Supraglottic Activity

Definition:	Constriction of supraglottic structures.
Rating:	Rate anteroposterior & mediolateral planes.

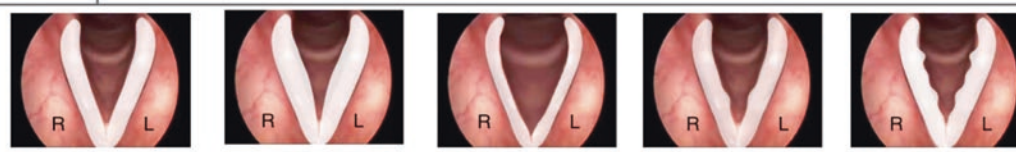


A-P: _____ ML: _____

F0: _____(Hz.)

Free Edge Contour

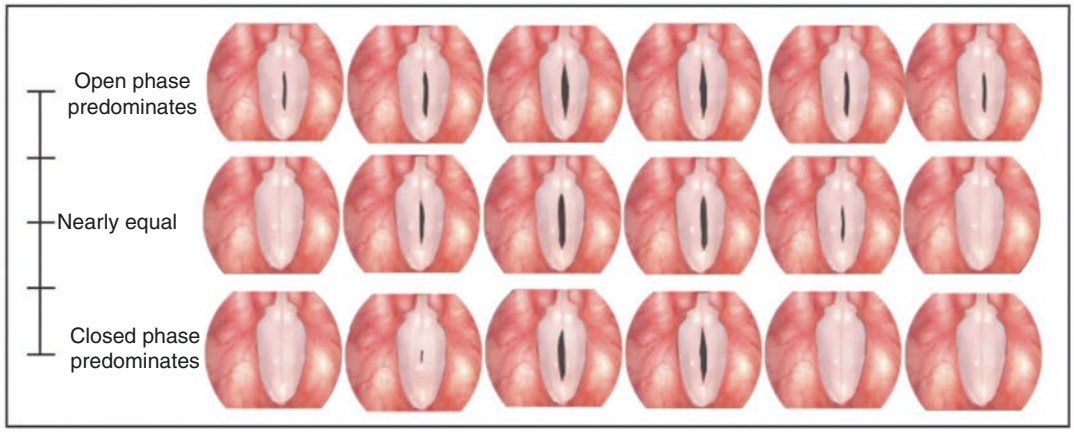
Definition:	Smoothness &/or straightness of free edge.
Rating:	Rate right & left VFs separately during abduction. Write in one rating per vocal fold.



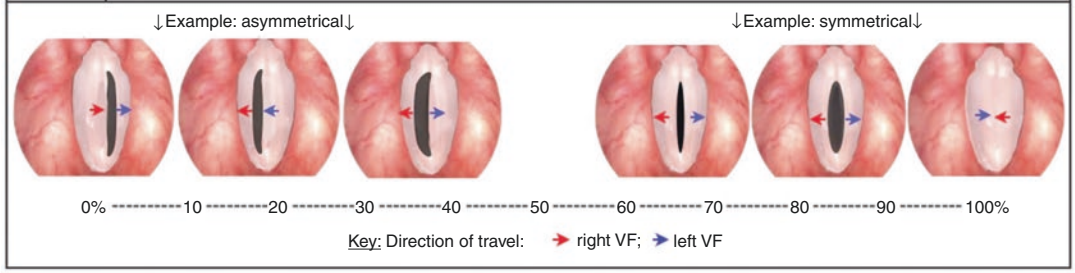
Right: _____	Left: _____

Fig. 14.5 (**a**, **b**) The Voice-Vibratory Assessment with Laryngeal Imaging (VALI) form: Stroboscopy. (**c**–**e**) The Voice-Vibratory Assessment with Laryngeal Imaging (VALI) form: High-speed Videoendoscopy

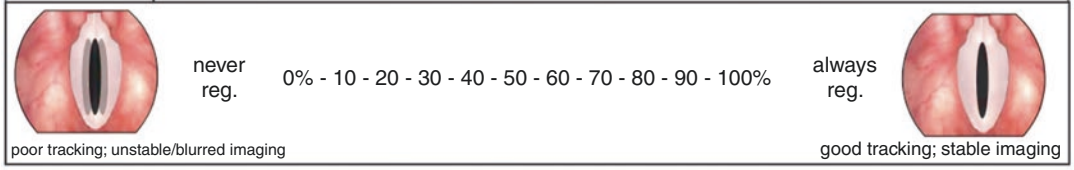
b	Phase Closure
Definition:	The relative durations of appearance of consecutive glottal cycles.
Rating:	Circle a hash mark on the continuum below. Rate at point of contact.



Phase Symmetry	
Definition:	The degree to which the VFs move as mirror-opposite images of each other (180° phase difference).
Rating:	Select the % of exam time that vibration is symmetrical.



Regularity	
Definition:	Consistency of cycles.
Rating:	Circle % of time vibration is regular



Nonvibratory Obserations	
Definition:	Report observations that were not noted elsewhere (e.g., structural differences, erythema, varices, mucous).

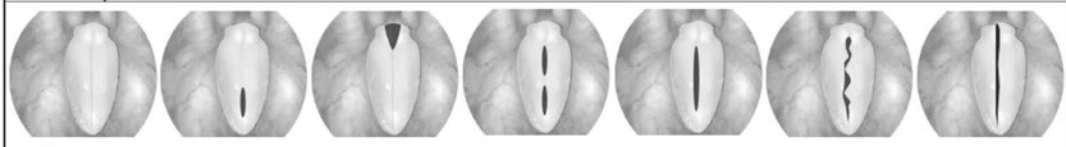
Fig. 14.5 (continued)

c

Case #:		Confirm case #:

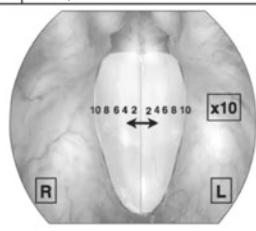
Glottal Closure

Definition:	Appearance of glottis during the most closed portion of the glottal cycle
Rating:	Rate at normal pitch and loudness.



Amplitude

Definition:	Magnitude of lateral movement of the vocal folds.
Rating:	Rate at point of contact.

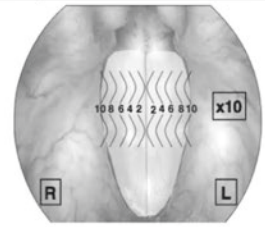


Right: _____ % Left: _____ %

F0: _____ Hz.

Mucosal Wave

Definition:	Magnitude of movement of the muc. membrane.
Rating:	Rate at normal pitch and loudness.

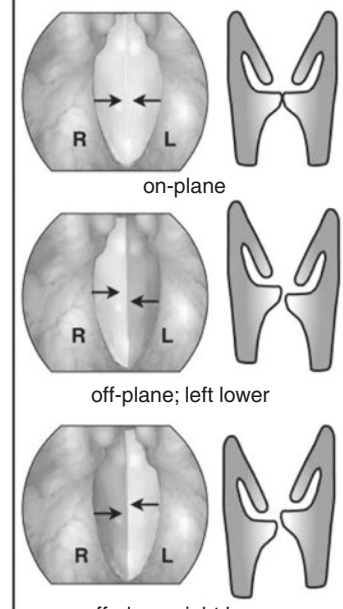


Right: _____ Left: _____

F0: _____ (Hz.)

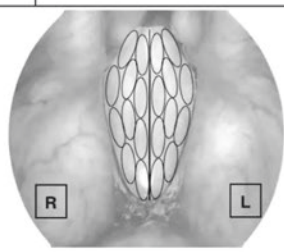
Vertical Level

Definition:	Do VFs meet on plane?
Rating:	Circle one



Non-vibrating Portion

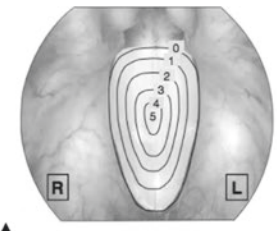
Definition:	Adynamic segments of tissue that appears stiff.
Rating:	Shade in affected area. Full ovals = 10% of TVF.



Right: _____ (ovals = 10%) Left: _____

Supraglottic Activity

Definition:	Constriction of supraglottic structures.
Rating:	Rate anteroposterior & mediolateral planes.

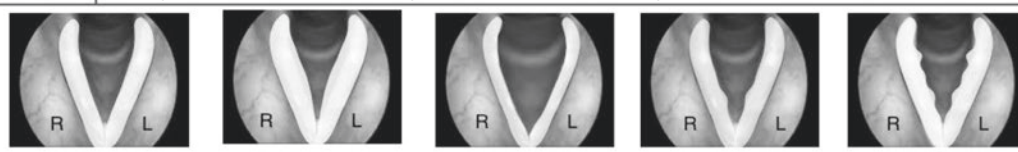


A-P: _____ ML: _____

F0: _____(Hz.)

Free Edge Contour

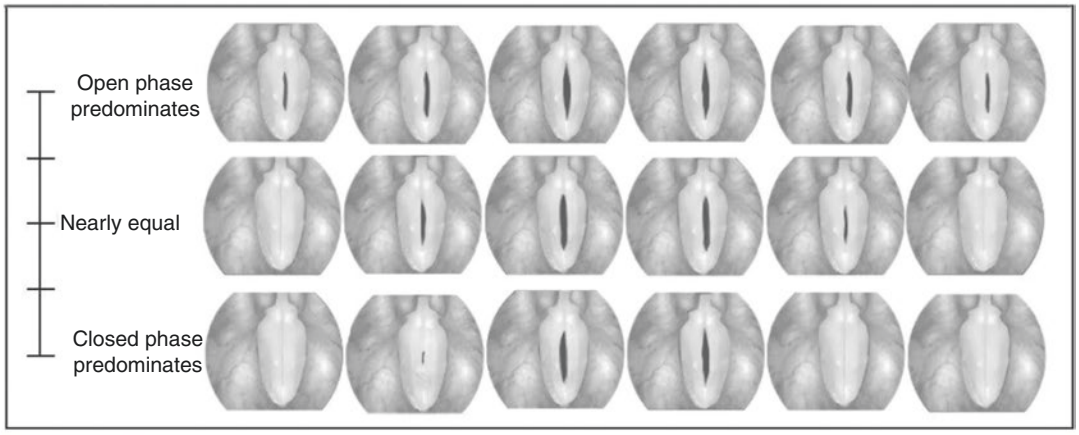
Definition:	Smoothness &/or straightness of free edge.
Rating:	Rate right & left VFs separately during abduction. Write in one rating per vocal fold.



Right: _____ Left: _____

Fig. 14.5 (continued)

d	Phase Closure
Definition:	The relative durations of appearance of consecutive glottal cycles.
Rating:	Circle a hash mark on the continuum below. Rate at point of contact.



Phase Symmetry	
Definition:	The degree to which the VFs move as mirror-opposite images of each other (180° phase difference).
Rating:	Select the % of exam time that vibration is symmetrical.

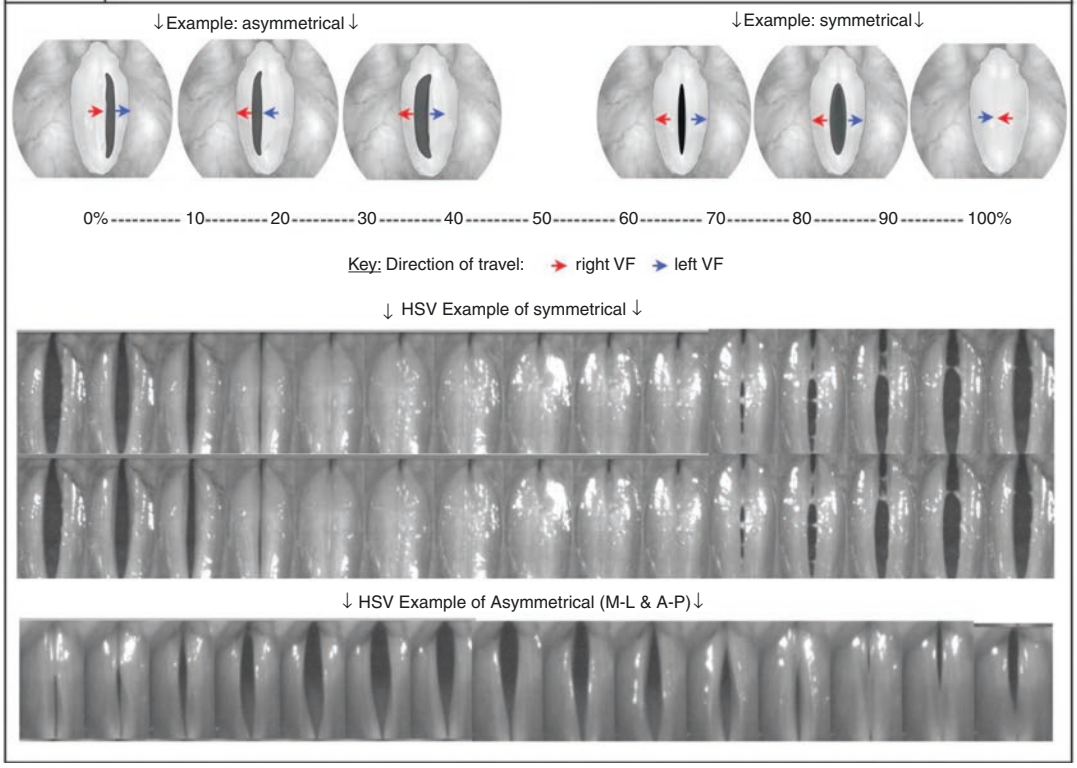
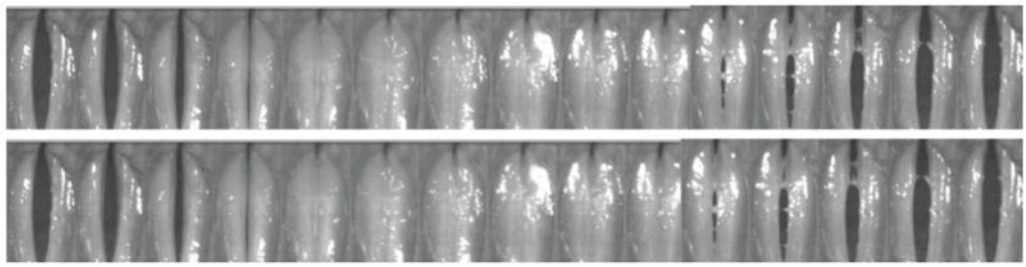


Fig. 14.5 (continued)

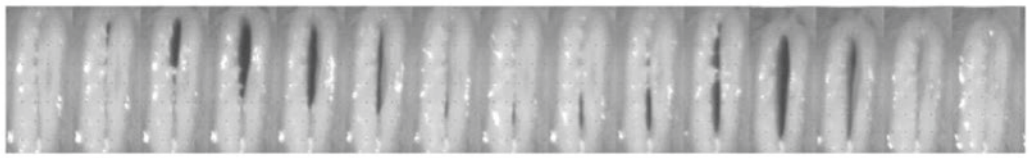
e	Glottal Cycle Periodicity
Definition:	Consistency of cycles.
Rating:	Circle % of time vibration is regular

never reg. 0% —10 — 20 — 30 — 40 — 50 — 60 —70 — 80 — 90 —100% always reg.

↓ Example of regular/periodic ↓

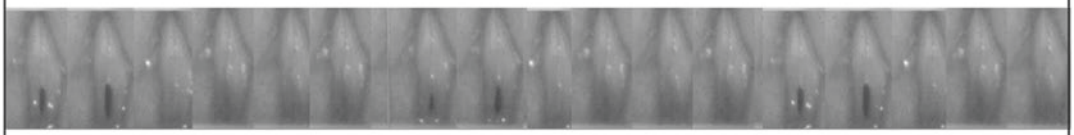


↓ Example of irregular/aperiodic ↓



Other Voice-Vibratory Patterns	
Definition:	Vibratory motion not noted elsewhere (e.g., vocal fry, oscilatory breaks, etc.).
Rating:	Specify what was observed and indicate frequency of occurrence (e.g., occasional, frequent, continuous)

↓ Example of vocal fry ↓



Definition:	Report observations that were not noted elsewhere (e.g., structural differences, erythema, varices, mucous).

Fig. 14.5 (continued)

folds and in differentially diagnosing lesions [23–25]. Successful stroboscopy has been reported on in the literature with children as young as 3 years old [23]. Detailed evaluation may be more challenging in children than adults due to multiple factors, including relative difficulty sustaining a pitch for the required number of cycles, difficulty cooperating, and a smaller larynx. Zacharias and colleagues found that clinicians were able to identify vibratory features in 92% of stroboscopic exams in children but only confidently rate those features in 42% of exams [24]. The researchers

found that raters were more able to rate the features when performed with a rigid endoscope than with a flexible scope and that older children were more able to tolerate the rigid exam than younger children [24]. As stated above, making a child more comfortable with the procedure is important not only for the child's comfort but also in our ability to make adequate observations. As a visual perceptual measure, ratings of videostroboscopy are by nature subjective and subject to the limitations of any perceptual measure. Relatively few studies using stroboscopy as an outcome measure have reported on interrater reliability, and of those that have, many are low [26, 27]. Ratings are dependent on the skill and experience of the rater, as well as their rigor in applying those skills. Efforts have been made over the years to standardize evaluation procedures and ratings in order to be more consistent across raters and clinics, and there are multiple rating forms available for use in evaluating stroboscopic images [16, 21, 26, 28, 29]. The Voice-Vibratory Assessment with Laryngeal Imaging (VALI) form (Fig. 14.5) provides a rating system for both stroboscopy and high-speed digital laryngeal imaging of the larynx [21]. Consistent use of the same methodology across raters, as well as regular practice and training, should improve reliability and clinical accuracy of ratings.

High-Speed Videoendoscopy

Videostroboscopy, the current gold standard in laryngeal imaging, is designed to evaluate periodic vibrations of any nature [16, 22]. In order to obtain reliable and valid visual perceptual judgments of vocal fold vibratory motion from videostroboscopy, a steady-state phonation of at least 2–3 s [20] from which three consecutive glottal cycles [30] can be viewed is required. In the pediatric population, it is often difficult to obtain steady-state phonation of a minimum of 2–3 s with either a rigid or flexible videostroboscopy due to examination factors of ease and cooperation. Other factors such as moderate and severe overall auditory perceptual impairment of voice quality typically also result in short phonations of less than 2 s, resulting in tracking errors on videostroboscopy [31]. The presence of tracking errors renders the exam clinically invalid for documenting the vibratory features of amplitude, mucosal wave, periodicity, glottal closure, etc. [30]. High-speed videoendoscopic systems are able to capture cycle-to-cycle vocal fold vibratory motion for phonations less than 2 s due to the high-temporal resolution of up to 8000 frames per second. In contrast with high-speed videoendoscopy, videostroboscopy is able to provide an averaged vibratory motion at 30 frames per second. The sampling rate of high-speed videoendoscopic systems is fast enough to also capture transient events of oscillatory onset, oscillatory offset, and voice breaks.

Instrumentation and Procedures

Since its first report in 1940 [32], high-speed videoendoscopy systems have undergone substantial modifications making the once impractical research tool now clinically feasible.

High-speed videoendoscopic systems have similar appearance to the videostroboscopy systems but differ substantially in terms of its basic principle and playback capabilities. Like videostroboscopy, simultaneous acoustic and various other signals (e.g., electroglottography, electromyography, etc.) can be captured with high-speed videoendoscopic recordings. However, unlike videostroboscopy, high-speed videoendoscopic recordings do not provide simultaneous playback of the video and audio. Slow video playback rates ranging from 10 to 30 frames per second are required to view and evaluate the high-speed videos captured at high-temporal resolutions of up to 8000 frames per second. Due to the current technological limitations, playback of audio simultaneously with the slow playback of the high-speed videos is not possible. The spatial resolution of high-speed videoendoscopy is generally lower (512×256 pixels) compared to videostroboscopic systems which can range from 720×468 for standard digital videostroboscopic systems to 1920×1080 pixels for high-definition videostroboscopic systems. As is evident high-definition

videostroboscopy is not similar to high-speed videoendoscopy as the former has high spatial resolution but is still lower in terms of the temporal resolution compared to high-speed videoendoscopy. Because high-speed videoendoscopic systems allow for the capture of cycle-to-cycle variations of vibratory motion due to its increased temporal resolution, high-speed videoendoscopy was reported to take less time (2.31 ± 1.92 min) compared to videostroboscopy (2.95 ± 2.41 min) for evaluation of vocal fold vibratory features in adolescents [25]. Common commercially available high-speed videoendoscopy systems are able to record phonations for up to 10 s requiring multiple recordings to capture the range of tasks required to evaluate the vocal fold structure and function. High-speed videoendoscopic systems also require a strong light source of 300 watts; hence care must be taken to turn the light source down between recordings to prevent any heat-related side effects from overheating of the tip of the endoscope. Because high-speed videoendoscopic systems differ in terms of the basic principles compared to videostroboscopy, considerable training is required for its use.

Core tasks and measures similar to those for videostroboscopy can be used for clinical examination with high-speed videoendoscopy. The use of tasks and procedure for videostroboscopy recommended by the American Speech-Language Pathology (ASHA) task force [30] is an ideal place to start as these tasks can also be used for high-speed videoendoscopy. The basic recommended protocol of rest breathing, laryngeal diadochokinetic tasks /i? i? i? i?/, and maximum vocal fold adduction and abduction(/i:/-sniff, /i:/-sniff) can be used for evaluation of vocal fold edges, vocal fold mobility, and the maximum range of vocal fold mobility at the level of the arytenoids [30]. The tasks of sustain phonation of /i:/, sustained /i:/ at varied pitch and loudness levels, and [5] variations in pitch and loudness on sustained /i:/ that elucidate the patients' problem can used to evaluate the vocal fold function features of supraglottic compression, regularity, amplitude, mucosal wave, glottal closure, left/right phase symmetry, vertical level, and glottal closure duration [30]. Often high-speed videoendoscopy is used in conjunction with videostroboscopy clinically rather than in isolation, especially in instances where videostroboscopy results in tracking errors due to short phonation time. Since high-speed videoendoscopy is often used in combination with videostroboscopy, the clinician may choose to limit high-speed videoendoscopy to the evaluation of vibratory function only, thereby reducing the overall time required for the clinical exam.

Evaluation

The vibratory motion obtained from high-speed videoendoscopy can be evaluated both quantitatively and qualitatively. Currently, quantitative tools for evaluating vibratory motion have not attained widespread utility as the custom-developed software systems are not readily available and often too laborious for routine clinical use. Qualitative visual perceptual evaluation of vocal fold structure and function is routinely used in clinic. The Voice-Vibratory Assessment with Laryngeal Imaging (VALI) form for visual perceptual evaluation of vocal fold structure and function can be used for both videostroboscopy and high-speed videoendoscopy (Fig. 14.5) as the VALI rating form was developed a prior for reliable visual perceptual ratings of vocal fold structure and vibratory characteristics for videostroboscopy and high-speed videoendoscopy [21]. The VALI visual perceptual rating form has improved graphics and definition of each parameter to aid the clinician for improved reliability in rating the laryngeal imaging features of interest [21].

The value of high-speed videoendoscopy to the understanding of vocal fold vibrations and voice production is immeasurable. Most of our current knowledge of vocal fold vibrations of normal and disordered voice in adults is derived from classic studies in the early 1960s from high-speed films [33, 34]. The first high-speed study on pediatric vocal fold vibrations was reported in 2011 [35]. Series of studies since 2011 quantify-

Table 14.1 Summary of differences in vibratory characteristics in typically developing children, adult females, and adult males without dysphonia

Vibratory characteristics	Children (5–11 years)	Adult females	Adult males
Glottal closure pattern	Posterior glottal gap (78%)	Posterior glottal gap (75%)	Posterior glottal gap (54%)
Glottal closure duration	Open phase predominant	Closed phase predominant	Closed phase predominant
Vibratory amplitude	Large	Small	Medium
Cycle-to-cycle variability	Large	Small	Medium
Left/right phase symmetry	Greater variability	Small variability	Small variability
Oscillatory onset time	Small	Medium	Large

ing vibratory motion using high-speed videoendoscopy in children have consistently revealed that the vibratory motion in children is complex and not easily predicted from vibratory motion of adults [36–39] (Table 14.1). Typically developing children demonstrate a posterior glottal gap more frequently compared to adult males and females. The posterior glottal gap in children is large extending to the membranous portion of the vocal folds resulting in a diamond-shaped gap [40]. The presence of this diamond-shaped posterior gap (Fig. 14.2) though not a statistically significant finding due to small sample size (boys = 28; girls = 28) could be considered as part of normal development rather than an abnormality on videostroboscopic examination. Typically developing children also had greater cycle-to-cycle variability in both amplitude and time periodicity and left/right phase symmetry during sustained steady-state phonation compared to adult men, suggesting greater aperiodicity of vocal fold vibrations in children [36]. The presence of these aperiodicities/instabilities in vibratory motion should not be confused with the presence of an abnormality but rather part of the normal development of vibratory motion in children. Quantitative measurement of vibratory amplitude revealed that children had large vibratory amplitude compared to the length of the vocal fold, suggesting that the adult normative reference of vibratory amplitude of 50% mediolateral excursion of the vocal fold may not hold true for pediatric vocal fold vibratory amplitude. In the absence of normative findings of vibratory motion in the pediatric population on videostroboscopy, normative findings from high-speed videoendoscopy can serve as a basis for clinical evaluation of vibratory characteristics from videostroboscopy.

High-speed videoendoscopy is the most powerful tool to date to evaluate vocal fold vibratory motion. With future studies, high-speed videoendoscopy will be able to provide further insights into vibratory motion across pitch and loudness variations and will thereby be able to provide detailed functional assessment of various voice disorders leading to timely and improved diagnosis of various vocal conditions in the pediatric population.

Emerging and Evolving Practices

Clinically, laryngeal imaging modalities of videostroboscopy, high-speed videoendoscopy, and videokymography have been primarily limited to providing qualitative or quantitative information about vocal fold vibrations in two dimensions (2D), which are not calibrated in terms of size and distance between the vocal folds and the tip of the endoscope. Vocal fold vibrations are three-dimensional involving not only the lateral and longitudinal dimensions which can be viewed from the superior surface but also the vertical dimension, which is often difficult to visualize from examination of the superior surface. Precise clinical measurements of the vertical dimension have significant potential to improve clinical diagnosis and management of dysphonia. Emerging studies using the latest generation of laser devices coupled with high-speed videoendoscopy have the capability to project a calibrated laser grid of 18 × 18 laser dots [41] and allow in vivo recording of the vertical dimension

in absolute values [42]. The applications of these new laser devices for clinical examination of pediatric vocal fold vibrations have the capability for generating new insights into the clinically relevant diagnostic process and thereby improve evidence-based assessment and management of pediatric voice disorders in the near future.

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Physiology of Normal Swallow

15

Corinne A. Jones

Introduction

Deglutition, commonly referred to as swallowing, is a complex sensorimotor process that involves the coordinated activation of many muscle groups with the goals of moving a food or liquid bolus through the mouth, throat, and esophagus while protecting the airway. Successful swallowing requires 31 paired muscles with sensory and motor information travelling over 6 cranial nerves [1]. This chapter will review typical anatomy and physiology of swallowing in infants and children.

Anatomy of Swallowing

The upper aerodigestive tract is comprised of the nose, oral cavity, pharynx, larynx, and esophagus. Schematics of the infant and adult upper aerodigestive tract with relevant anatomy labeled are in Fig. 15.1 and muscles in Table 15.1.

Oral Cavity

The oral cavity is comprised of the lips, mandible, maxilla, hard palate, soft palate, tongue, cheeks, and floor of mouth muscles. The posterior boundary of the oral cavity is typically delineated at the anterior faucial pillars [2]. Spaces in between the mandible/maxilla and the cheeks are the *lateral sulci*, and the space between the mandible/maxilla and the lips is the *anterior sulci*. The oral cavity is separated from the pharynx by a ring of tissue that includes the circumvallate papillae, lingual tonsils (at the tongue base), and soft palate [3].

Pharynx

The pharynx is divided into three subregions: the nasopharynx, oropharynx, and hypopharynx. The *nasopharynx* connects the nasal cavity with the oropharynx and hypopharynx and serves as a conduit for air and nasal/paranasal sinus secretions. The boundaries of the nasopharynx are the posterior surface of the nasal turbinates (anterior), the skull base (posterior), the adenoids (superior), and the soft palate (inferior). The *oropharynx* follows posteriorly from the oral cavity, bounded by the anterior faucial pillars (anterior), posterior pharyngeal wall (posterior), the soft palate (superior), and the tip of the epiglottis (inferior). In infants, there is no anatomic oropharynx, as the entire tongue resides in the oral cavity, and the soft palate approximates the epiglottis (see Fig. 15.1) [4, 5]. The *hypopharynx* is the most inferior subdivision of the pharynx and

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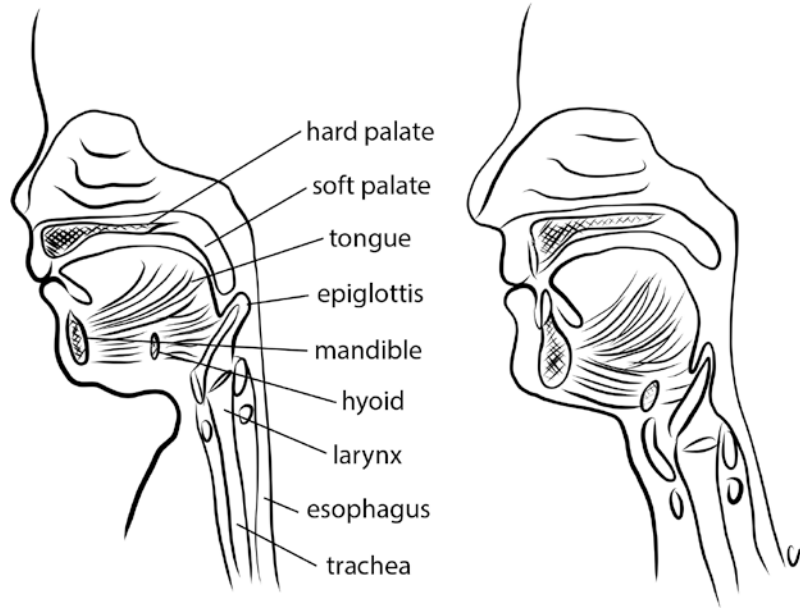

Fig. 15.1 Swallowing-related anatomy from the lateral view in an infant (left) and in an adult (right)

is bounded by the false vocal folds of the larynx (anterior), posterior pharyngeal wall (posterior), the tip of the epiglottis (superior), and the upper esophageal sphincter (inferior). The space between the tongue base and epiglottis is the valleculae, and the spaces posterolateral to the larynx are the pyriform sinuses.

Larynx

See Chap. 7 for a thorough overview of laryngeal structure and function. Structures relevant for swallowing include the true and false vocal folds, the arytenoid cartilages, and the epiglottis (Table 15.1).

Esophagus

The esophagus is a mucous membrane-lined tube that stretches between the upper esophageal sphincter at the junction with the hypopharynx and the lower esophageal sphincter at the junction with the stomach. These sphincters are tonically active to remain closed at rest and sequentially open during swallowing, belching, retching, and vomiting [6]. There are two muscular layers of the esophagus: one with fibers oriented circularly and the other with fibers oriented longitudinally. The rostral third of the esophagus is comprised of striated muscle, with a transition to smooth muscle at the caudal third [7].

Development of Upper Aerodigestive Anatomy

Several aspects of upper aerodigestive anatomy change as an individual develops (Fig. 15.1). The 20 deciduous teeth erupt between 6 and 33 months of age [2, 4, 8]. The sucking pads resorb, resulting in a larger buccal space [4]. As the individual grows, the mandible approximates the size of the maxilla, the tongue takes up a lesser proportion of the oral cavity, the hyoid and larynx descend to a lower position in the neck (~C2–3 to C4–6 for the larynx), and the approximation between the soft palate and epiglottis is lost, resulting in an oropharyngeal space [2, 4, 8–11]. The adenoids grow for the 1st year of life before shrinking around the age of 8, and the angle of the skull base at the nasopharynx becomes more acute [4]. The larynx grows in size and becomes less

Table 15.1 Muscles involved in each phase of swallowing

Phase	Anatomic location	Muscle (motor innervation)
Oral	Lips/cheeks	Orbicularis oris (CN VII)
		Buccinator (CN VII)
		Lip elevators/depressors (CN VII)
	Mandible	Temporalis (CN VII)
		Masseter (CN VII)
		Lateral pterygoid (CN VII)
		Medial pterygoid (CN VII)
	Tongue	Genioglossus (CN XII)
		Hyoglossus (CN XII)
		Styloglossus (CN XII)
		Palatoglossus (CN X)
		Superior longitudinal (CN XII)
		Inferior longitudinal (CN XII)
		Vertical (CN XII)
		Transverse (CN XII)
Pharyngeal	Soft palate	Palatoglossus (CN X)
		Palatopharyngeus (CN X)
		Levator veli palatini (CN X)
		Tensor veli palatini (CN V)
		Musculus uvulae (CN X)
	Pharynx/hyoid/extrinsic larynx	Anterior digastric (CN V)
		Posterior digastric (CN VII)
		Palatopharyngeus (CN X)
		Palatoglossus (CN X)
		Salpingopharyngeus (CN X)
		Stylopharyngeus (CN IX)
		Styloglossus (CN VII)
		Stylohyoid (CN XII)
		Geniohyoid (CN XII)
		Mylohyoid (CN V)
		Thyrohyoid (ansa cervicalis)
		Superior pharyngeal constrictor (CN X)
		Middle pharyngeal constrictor (CN X)
		Inferior pharyngeal constrictor (CN X)
	Intrinsic larynx	Lateral cricoarytenoid (CN X)
		Posterior cricoarytenoid (CN X)
		Transverse arytenoid (CN X)
		Oblique arytenoid (CN X)
		Thyroarytenoid (CN X)
		Thyroepiglottic (CN X)
		Aryepiglottic (CN X)
	Upper esophageal sphincter	Inferior pharyngeal constrictor (X)
		Cricopharyngeus (X)
		Striated esophageal muscles (X)
Esophageal	Esophagus	Striated esophageal muscles (X)
		Smooth esophageal muscles (X)

funnel-shaped, the laryngeal cartilages stiffen, the arytenoid cartilages take up a lesser proportion of the larynx, the angles of the vocal folds approximate parallel to the transverse plane, and the epiglottis stiffens and flattens from an omega shape [4, 10, 11].

Swallowing Physiology

Swallowing can be broken down into four phases: oral preparation, oral transport, pharyngeal, and esophageal [7]. Swallowing is composed of reflexive and voluntary movement patterns, partially controlled by central pattern generators in the brainstem [11, 12]. Swallowing physiology changes as the individual develops, but overall sequencing of biomechanical events remains stable, particularly in the pharyngeal and esophageal phases. An overview of the events that occur in each phase of swallowing is in Table 15.2.

Table 15.2 Events that occur in each phase of swallowing. Swallowing is not a purely sequential process; events within phases overlap in time

Phase	Event
Oral preparation	Food/liquid enters mouth; via sucking in infants
	Cohesive bolus formed and mixed with saliva; mastication if necessary
	Bolus is gathered onto and contained at the center of tongue
	Soft palate contacts posterior oral tongue to prevent bolus spilling into pharynx (except during mastication)
Oral transport	Tongue tip contacts alveolar ridge and strips posteriorly to propel bolus toward pharynx
	Lateral tongue contacts hard palate to prevent bolus spillage into sulci
	Posterior tongue depresses
Pharyngeal	Soft palate raises
	Hyoid moves anterior and superior
	Larynx moves anterior and superior
	Laryngeal vestibule closes
	Epiglottis retroflexes
	Pharyngeal constrictors activate superiorly to inferiorly
	Tongue base retracts to posterior and lateral pharyngeal walls
	Upper esophageal sphincter opens
Esophageal	Peristalsis of esophageal musculature superiorly to inferiorly
	Lower esophageal sphincter opens

Oral Preparation Phase

Anatomy and sensorimotor development largely determine the nature of the oral preparatory phase, and this phase of swallowing changes most with age [6]. However, the goal of this phase remains the same: to prepare the food and liquid bolus for swallowing (Table 15.2). The food or liquid is tasted, mixed with saliva, and prepared into a cohesive bolus of appropriate consistency and size.

In infants, the nipple of the breast or bottle is placed into the mouth, facilitated by rooting and grasping reflexes [11]. Milk is extracted through *sucking*, a coordinated movement pattern of the lower lips, tongue, mandible, and hyoid that occurs in two phases: suction and expression. Suction occurs by negative intraoral pressures generated by expanding the oral cavity [11, 13]. During *suction*, the soft palate closes off the nasopharynx, the lips make a seal on the nipple, and the jaw is lowered to increase the anatomic space of the oral cavity. This atmospheric pressure gradient results in milk flowing into the mouth. *Expression* occurs through the direct compression of the nipple by the tongue, mandible, and maxilla [6, 13], generating a contact pressure gradient that also results in milk flow into the mouth. It is believed that suction is more important than expression for getting milk into the mouth, but the rate of sucking is the major factor for regulating overall intake [6, 14, 15]. As infants mature, sucking matures from a purely reflexive to a more voluntary process, sucking rates increase, and suction and expression actions become rhythmic and alternating [6, 13, 16, 17]. Infants born at term will suck approximately once per second, if in the act of ingestion (*nutritive sucking*), while sucking with no liquid (e.g., with a pacifier) at a rate of two sucks per second (*nonnutritive sucking*) [13].

Preterm infants have underdeveloped sucking patterns. Sucking movements may occur without generating negative intraoral pressures and in shorter bursts [11, 14, 18]. Those with stable cardiac, pulmonary, and gastrointestinal function are generally introduced to nonnutritive sucking at 28–29 weeks and to oral feeding at 32–34 weeks post-menstrual age [19–21]. Sucking patterns continue to develop with post-menstrual age, and do not generally correlate with postnatal age [22]. Nonnutritive sucking during gavage feeding is

important for preterm infants to develop mature sucking patterns and for overall well-being [6, 19, 23]. Success with nonnutritive sucking is also commonly used as an indicator for readiness to transition to bottle feeds in the preterm infant [21].

For the first 3 months, infants will use a similar sucking action for both liquids and solids [5, 6]. Between 4 and 6 months, many of the reflex responses that assist with feeding disappear, and spoon feeding is generally introduced around 5 months [5]. Rhythmic biting develops between 7 and 9 months, as the buccal fat pads resorb and as the tongue protrusion reflex disappears [4, 6, 21]. These initial mastication efforts help to strengthen the muscles of mastication in preparation for transition to solid foods [24]. Rotary mastication efforts tend to emerge around 8–12 months of age [5, 24] with masticatory force increasing when teeth begin to erupt, around 12 months [6]. Efficiency of initial chewing behaviors is low, increasing to approximately 40% of adult level by 6 years and fully mature in the teenage years [6, 24]. Coordination of oral movements for preparation of solids, including prehension, biting, mastication, and tongue and cheek manipulations for gathering of the bolus, emerges with age and overall developmental skill levels [11, 21]. Repeated exposures to a variety of different tastes and textures are crucial for this development [5, 24].

As teeth emerge and the child has mastered many solid food textures, oral preparation mostly resembles that of adults. Once the food has been placed in the mouth, it is ground up with a rotary action of the mandible and tongue [7]; this allows the material to be mixed with saliva. Precise movements in the oral cavity depend on the nature of the material to be swallowed. During this phase, the labial seal is maintained, the pharynx is relaxed, the nasopharyngeal passage is open, and the individual breathes as normal [7, 11]. The soft palate contacts the posterior oral tongue, except during mastication of solids. The final action in this preparatory phase is to place the bolus in the middle of the tongue; the anterior and lateral edges of the tongue press against the maxillary alveolus to prevent the bolus from spilling into any of the oral sulci [7]. During this phase, sensory information is gathered regarding bolus size, taste, and consistency, which is particularly important for motor programming of the pharyngeal phase of swallowing [7].

Oral Transport

The goal of the oral transport phase is to propel the bolus from the oral cavity to the pharynx (Table 15.2). This phase is under voluntary control and consists of an anterior-to-posterior stripping of the tongue against the hard palate. This occurs as the soft palate rises up to the pharyngeal walls and the posterior oral tongue dips to allow for bolus passage from the oral cavity into the pharynx [6, 7, 11]. During sucking, infants will deliver milk directly to the posterior oral cavity, so the need for oral transport is minimal [24]. At around 4–6 months, this tongue stripping action will occur in response to solid and semisolid foods [6, 24].

Pharyngeal Phase

The pharyngeal phase of swallowing has the purpose of propelling the bolus into the esophagus while protecting the airway and consists of multiple necessary events that overlap in time (Table 15.2). As the bolus is propelled past the anterior faucial pillars into the pharynx, activation of mechanoreceptors on the tongue base, epiglottis, and pyriform fossa is responsible for triggering the pharyngeal phase of the swallow [6, 7]. This involuntary, patterned response consists of actions that move structures of the upper aerodigestive tract and that apply propulsive pressures to the bolus.

Multiple valving-type actions occur during the pharyngeal phase of swallowing. Most superiorly, the soft palate raises up against the contracting pharyngeal walls to close off the nasal cavity [6]. The laryngeal vestibule is closed via multiple mechanisms: (1) true vocal fold closure, (2) false vocal fold approximation, (3) epiglottic inversion, and (4) arytenoid to epiglottic base contact [25]. The upper esophageal sphincter is

comprised of the inferior pharyngeal constrictors, cricopharyngeus, and cervical esophageal musculature, which relaxes open prior to arrival of the bolus head and contracts following bolus tail passage into the esophagus [26–28]. Anterior and superior hyolaryngeal excursion facilitates epiglottic inversion, arytenoid approximation to the base of the epiglottis, and upper esophageal sphincter opening [6, 26].

Other actions during the pharyngeal phase of swallowing act to put propulsive forces on the bolus. Following passage of the bolus tail, the tongue base approximates the posterior pharyngeal walls, which in turn constrict in a superior-to-inferior stripping wave [6, 29]. This puts a positive pressure gradient on the tail of the bolus. Hyolaryngeal excursion and upper esophageal sphincter opening create a larger hypopharyngeal space, generating subatmospheric pressure that acts on the head of the bolus, directing it into the esophagus [26, 30]. Gravity also exerts a force on the bolus, but this force can be overcome while swallowing in a supine or side-lying position or even while fully inverted [31, 32].

As the hyolaryngeal complex sits higher in the infant pharynx, hyolaryngeal excursion movements are decreased. These movements increase with age, as the hyoid and larynx become situated more inferiorly in the pharynx [33]. There is not a difference in timing of the pharyngeal stage onset or in pharyngeal duration with typical development, suggesting an adaptation to the increase in pharynx size [34].

Esophageal Phase

The esophageal phase of swallowing consists of bolus passage through the esophagus and into the stomach (Table 15.2). This phase is entirely controlled by the autonomic nervous system [6]. After passing through the upper esophageal sphincter, the bolus passes quickly through the cervical esophagus, which is comprised of skeletal muscle, and then through the smooth muscle-containing thoracic and abdominal esophagus [6]. Primary peristaltic esophageal waves originate with an oropharyngeal swallow, and secondary esophageal waves begin in the body of the esophagus [6]. With development, a greater number of secondary esophageal waves are observed [13]. Esophageal peristalsis can also be influenced by changes in intrathoracic pressure, such as inspiration, expiration, or coughing [6]. The esophageal phase ends with passage of the bolus through the lower esophageal sphincter into the stomach. The lower esophageal sphincter is less competent in infants, which explains the frequent gastroesophageal reflux or "spit-up" events [6].

Respiratory-Swallowing Coordination

A critical component of swallowing at all stages of development is the coordination of respiratory activity and swallowing. During the oral phases, the pharynx is relaxed and the airway is open, allowing for continuous breathing. During the pharyngeal phase of the swallow, the airway closes and respiration ceases [6, 7, 11]. This regulation occurs via coordinated activities of respiratory and swallowing central pattern generators in the brainstem [6].

Infants are obligate nasal breathers and thus must coordinate their sucking and swallowing with respiration, commonly referred to as a *suck-swallow-breathe* cycle [6, 11]. This cycle represents a coordination between the three activities such that the pharyngeal phase of swallowing does not interrupt the respiratory cycle [25]. Mature infants will typically display a 1:1:1 or a 2:2:1 ratio of suck-swallow-breathe [17, 20], with most swallows being followed by expiration [21, 35]. Preterm infants generally show an immature suck-swallow-breathe pattern, with the swallow occurring during different phases of respiration, including mid-inhalation [13, 35, 36]. It is hypothesized that many of the feeding difficulties seen in preterm infants come from discoordination of swallowing and respiration [36]. Nonnutritive sucking and tactile-kinesthetic interventions in the preterm infant can improve this coordination [23]. In adults, most healthy individuals swallow in the expiratory phase or at the inspiration/expiration transition, with expiration following the swallow [37–39].

Conclusions

Swallowing is a complex and dynamic process, involving coordination between multiple sensorimotor systems. Swallowing anatomy, physiology, and motor control develop in the healthy human in order to allow for ingestion of more complex foods with more efficiency. Due to the intricate nature of swallowing, multiple disease processes or injuries can interrupt central or peripheral systems, leading to difficulty swallowing (*dysphagia*), discussed in ensuing chapters.

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Clinical Evaluation of Swallow

16

Jesse D. Hoffmeister

Introduction

The clinical evaluation of swallowing (commonly known as a "bedside swallow assessment") is the evaluation of swallowing function without the use of instrumental measures such as videofluoroscopy, fiber-optic endoscopy, or high-resolution pharyngeal manometry. While clinical swallow assessment cannot be used to describe certain parameters of swallow function (e.g., presence of silent aspiration, relative differences in pressure at varying levels of the pharynx, exact duration of delay in pharyngeal swallow response), the information gained in clinical evaluation is invaluable in helping to understand the basic physiologic causes of dysphagia and in identifying factors that contribute to dysphagia [1]. A clinical swallowing assessment includes thorough review of the medical record and collection of a case history; direct and indirect assessment of oral, laryngeal, and pharyngeal sensorimotor structure and function; observation of bolus trials; and finally modification of bolus properties and modification of method of bolus presentation when necessary. For the purposes of this chapter, we primarily discuss assessment of the skill-based disorder of oropharyngeal dysphagia, as opposed to feeding disorders, which are primarily behavioral in nature [2].

Developmental Changes in Swallowing Anatomy and Physiology

Etiology of dysphagia often differs between pediatric and adult populations. This is not surprising when considering the anatomic and physiologic changes that occur with development. In infants and young children, the oral cavity is small relative to the overall size of the person. The position of the mandible requires that the tongue occupy a larger portion of the oral cavity early in life, and the near absence of a hard palatal arch means that lingual position is less variable in infancy. Subsequently, younger infants use anterior-posterior lingual movements known as "suckling" to elicit a fluid bolus, rather than the superior-inferior lingual movements known as "sucking," which develop with age [3].

In the infant pharynx, the epiglottis rests high (often touching the velum), and the hyolaryngeal complex lies more superior than in the adult (almost immediately below the base of the tongue). This configuration means that young infants are obligate nasal breathers [4]. The early positioning of upper airway anatomy is thought

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to provide some protection against aspiration (although aspiration can still certainly occur); as the larynx descends during development to facilitate vocal communication, the protection against aspiration fades [5].

The neural motor swallow patterns of infancy, often reflexively regulated, are also different from those in the developed system. Some of the relevant reflexes include rooting (head turning toward stimulation with simultaneous mouth opening), phasic bite reflex, suck reflex, tongue protrusion ("tongue thrust"), and gag reflex. Rooting tends to disappear around the fifth month of life, and bite and suck reflexes tend to disappear later in the 1st year of life [3, 6].

In persons with developed swallowing mechanisms, swallowing events are discrete, with identifiable separations between individual bolus swallows. In infants, on the other hand, the anatomic and neurologic differences described above result in a nearly continuous suck-swallow-breathe pattern that occurs at approximately a 1:1:1 ratio requiring exquisite sensorimotor coordination [7]. When coordination of the suck-swallow-breathe sequence breaks down, airway protection is sacrificed, described in further detail below.

Case History

A thorough review of the medical record is essential for both efficiency and diagnostic accuracy in evaluation of swallow function. Table 16.1 describes conditions that are commonly associated with feeding and swallowing disorders [2, 3, 5, 8–13]. Because swallowing requires integration of multiple body systems (i.e., cardiopulmonary, neurologic, gastrointestinal, musculoskeletal), the clinician should take note of the patient's chronic and acute medical conditions, particularly those with a known relationship to dysphagia. Approaching swallowing evaluation from a systems-based perspective during chart review additionally provides the clinician with a framework upon which to begin building hypotheses regarding dysphagia diagnosis, etiology, and treatment options.

Table 16.1 Disorders commonly associated with pediatric dysphagia [2, 3, 5, 8–13]

Neurological disorders
Traumatic brain injury
Microcephaly
Hydrocephalus
Arnold-Chiari malformations
Intraventricular hemorrhage
Cerebral palsy
Guillain-Barré syndrome
Seizures
Spinal muscular atrophy
Respiratory and cardiac disorders
Apnea of the newborn
Respiratory distress syndrome
Bronchopulmonary dysplasia (chronic lung disease)
Infections causing impaired respiration (i.e., respiratory syncytial virus)
Cyanotic and acyanotic heart defects and some associated surgical interventions
Gastrointestinal disorders
Necrotizing enterocolitis
Esophageal dysmotility
Hirschsprung's disease
Gastroschisis
Tracheoesophageal fistula and esophageal atresia
Congenital diaphragmatic hernia
GERD
Eosinophilic esophagitis
Congenital abnormalities
Ankyloglossia (tongue tie)
Cleft lip/palate
Laryngeal cleft
Laryngo-/tracheo-/bronchomalacia
Moebius syndrome
Down syndrome
Cornelia de Lange syndrome
Iatrogenic complications
Tube feeding
Tracheostomy
Respiratory support, including long-duration intubation
Ingestion of foreign bodies and caustic agents
Caustic agents (chemical injury)
Foreign bodies
Prematurity and associated comorbidities

Interview

Caregiver Questionnaires

Several tools that use data obtained from caregiver observations and caregiver report of history relevant to swallowing and feeding assessment

have been published. Heckathorn performed a systematic review identifying such assessments [14]. These assessments are useful not only for collecting information on the patient's swallowing and feeding but for helping the caregiver to organize their thoughts and observations so as to more clearly convey them to the clinician. Despite practical usefulness, inferences taken from these assessments must be made with caution. Currently, only two assessments provide normative data [14]. The Mealtime Behavior Questionnaire is a 33-item list of 5-point ordinal scales divided into subscales of food refusal/avoidance, food manipulation, mealtime aggression/distress, and coughing/choking/vomiting [15]. Total scores of greater than or equal to 100 are considered to be abnormal. *t* and *z* scores can be calculated for each subscale and compared to "nonclinical" samples reported in the original publication. A second tool, the Screening Tool of Feeding Problems, modified for children (STEP-child), provides normal/abnormal cutoffs for subscales of rapid feeding, food refusal, food selectivity, vomiting, and stealing food [16]. While useful for assessment of feeding and consideration of mealtime behaviors that could be related to swallowing dysfunction, this scale does not include items that interrogate swallow function directly.

Caregiver Interview

Interview of caregivers is the primary means of obtaining information about pediatric swallowing and feeding history prior to adolescence. When beginning the case history, it is beneficial to ask open-ended questions in order to get an accurate sense of the problem in question. (For example, "I've heard that Johnny has been having some troubles with (swallowing, weight gain, taking the bottle, choking, etc.). Tell me what you've been noticing") (Table 16.2).

Every aspect of swallowing should be addressed. Determining the *type of bolus* (breast milk, formula, cow's milk, solids, etc.) is important when considering how bolus characteristics impact swallow performance; this can also give the clinician a sense of overall developmental level in late infancy and early toddlerhood. The *methods of bolus delivery* (breast, bottle, open cup, self-feeding versus dependence) and *positioning during feeding* further characterize swallow function and represent primary targets for modification if necessary. *Volume of intake*, *frequency of intake*, and *duration of feeding* are also relevant. While these parameters vary widely both within and between patients, most infants feed every 2–4 h with longer breaks at night, and most feedings last between 10 and 30 min [12]. It can also be beneficial to ask the parent, "Is feeding easy, stressful, or somewhere in between?" This often provides tremendous insight into quality of feeding and can elicit additional caregiver descriptions that may assist in diagnosing the disorder.

Finally, signs of aspiration and swallowing difficulty should be discussed. These include the more obvious signs of coughing, choking, throat clearing, wet/gurgling voice quality, and congestion of breath sounds with feeding [17]. More subtle signs of a swallowing problem in infants and very young children include food refusal and/or signs of distress, such as widely open/"surprised" eyes, splaying of hands/feet, tight clenching of hands/feet, back-arching, head-turning or other attempts to disengage from

Table 16.2 Useful questions for characterizing pediatric swallowing

What does the patient eat and/or drink? (Solids, semisolids, first foods, breast milk, formula, cow's milk, etc.)
How does the patient eat and/or drink? Are they fed or do they self-feed?
What does the patient eat/drink from? (Bottle brand, nipple type and flow rate, exclusively breastfed, sippy cup, finger foods, etc.)
How is the patient positioned during feeding?
How much (volume) does the patient eat per meal/feeding?
How long does it take to finish a feeding?
How often do they feed?
Does the patient let you know it's time to eat, or do you have to remind and/or alert them to feed?
What does the feeding routine look like, step-by-step?
Is feeding stressful for you or for the patient?
Do you notice any coughing or choking? Wet, gurgling breathing sounds? (Other signs of aspiration?) Signs of distress? Refusal? Shutting down/"sleeping" in infants?

the nipple or food source, and even shutting down, which may be described as "falling asleep" by caregivers [12, 18, 19].

Contributing Medical History

After a general understanding of the patient's swallowing has been obtained, the clinician should begin discussing additional aspects of the patient's medical profile that may contribute to swallowing difficulty.

Gastrointestinal symptoms commonly contribute to pediatric swallowing disorders and should be frankly discussed with caregivers. Does the patient have frequent or large-volume spit-ups? Does the patient seem uncomfortable before or during spit-ups (e.g., with back-arching and fussing), or are they "happy spits?" Does the patient have difficulty with constipation? Do there seem to be any triggers for any of these gastrointestinal difficulties? As translational research continues to elucidate mechanisms of neural swallowing control, it is becoming increasingly clear that gastric and esophageal disorders have a direct impact on oropharyngeal swallow function [20].

Respiration and deglutition are intimately related [5, 7]. For infants, respiratory difficulties can interrupt the suck-swallow-breathe cycle and cause some degree of swallowing dysfunction. Asking the caregiver if the patient experiences breathing difficulties at any time, especially during feeding or in certain positions, may be relevant to swallow function. If caregivers endorse breathing difficulty, they should be asked to describe this. Is it noisy or just very fast? If it's noisy, is it on the inhalation, exhalation, or both? Does it change based on position? What about activity (i.e., feeding, crawling, crying, sleeping)? If noisy breathing is endorsed or observed, consultation with otolaryngology and upper airway visualization are recommended.

Voice problems, addressed elsewhere in this text, can point to potential etiologies of swallowing difficulty. A breathy, weak voice may indicate incomplete glottic closure or poor pulmonary drive. Wet, gurgling voice may indicate penetration, aspiration, or pharyngeal residue. Rough voice may indicate presence of a mass lesion in the upper airway. It cannot be emphasized enough that if any of these voice problems are reported by caregivers or observed by the clinician, consultation with otolaryngology and laryngeal visualization are necessary.

Neurologic status has a tremendous influence on swallow function [6]. Neurologic status can be very grossly measured by assessing the patient's level of alertness and arousal. "Calm/alert" and "semi-drowsy but actively engaged" are the typical states of arousal in normal feeding [21]. Because young infants still possess certain reflexive feeding behaviors (sucking/biting reflex, rooting reflex), "passive feeding" can occur. In passive feeding, well-intentioned caregivers may place a nipple in a non-alert infant's mouth, and, with sufficient stimulation, sucking/biting reflexes can be elicited, resulting in bolus transfer. Depending on the position of the infant, the bolus can then passively flow to the posterior oral cavity, significantly increasing risk for pharyngeal swallow dysfunction and reduced airway protection. The clinician should obtain information on the patient's typical state of arousal during feeding and nonfeeding times. Additional insight into neurologic status and its impact on swallowing function can be gained through discussion of which developmental milestones the patient has reached.

For each disordered finding (swallowing, feeding, respiratory, voice), the time of onset, nature of onset (gradual or sudden), progression of the finding (worsening, improving, or stable), transience (constant or intermittent), and current status (most recent episode) should be documented.

Child Interview

If the patient is verbal, they should also participate in providing the case history. Younger children in particular look to their caregivers to determine how to respond to an unfamiliar clinician. If good rapport with the caregiver is established prior to interviewing the child, the child will be much more likely to participate. Optimizing the environment to make the child feel comfortable will also aid in eliciting a meaningful interview. For example, one might set up a child's table and chairs equipped with a variety of

toys in the examination room. In the same vein, initiating conversation with the child by first joining in play and gradually posing questions is often more productive than a face-to-face bombardment of questions. When discussing swallowing with the patient, be sure to use child-appropriate language. For example, when asking the child about reflux, one might pose the question, "Do you ever get mini throw-ups, where you puke a little in your mouth?" rather than "How often do you experience heartburn?"

Feeding and Swallowing Observation

Assessment Tools

In addition to assessment tools completed by caregivers, there are a number of tools designed to be completed by clinicians. These can be used to guide swallowing assessment. Heckathorn's 2015 systematic review identifies four tools that provide normative data related to swallowing and/or feeding [14]. The Oral Motor Assessment Scale is designed for oral motor skills assessment of children with cerebral palsy aged 3–13 years and 11 months [22]. The Schedule for Oral Motor Assessment is completed by a clinician based on feeding observations for ages 8 months to 2 years, separated into discrete categories of puree, solid, semisolid, cracker, bottle, trainer cup, and open cup [23, 24]. Each category contains a norm-referenced cutoff. Other tools with norm-referenced cutoffs include the Pre-Speech Assessment Scale, designed for young children with cerebral palsy or other developmental disabilities aged 0–2 years and 1 month [25], and the Pediatric Assessment Scale for Severe Feeding Problems, designed for infants with severe feeding problems who feed orally and are aged 0–4 months [26].

Physical Assessment

State of Arousal and Position As previously mentioned, state of arousal (calm/focused/alert versus distressed or sleeping) and ability to modify state of arousal will have a significant impact on swallowing function. For infants in particular, the ability to be soothed is important for establishing a well-coordinated suck-swallow-breathe cycle. For example, an infant who is very distressed may not be able to organize feeding behaviors sufficiently to swallow safely and effectively and may not be able to modify the suck-swallow-breathe cycle sufficiently to account for the alterations in respiratory rate associated with crying. Gross body movement patterns can also influence swallowing in infants. For example, a distressed infant with back-arching whose neck is constantly in extension will modify the shape of the aerodigestive tract such that aspiration can more easily occur (i.e., with a "chin up" position). The ideal alternative is a position of comfortable flexion with the nose approximately aligned with the navel. If the infant typically feeds in a distressed state, while in extension or with other suboptimal body positioning, this should be noted, and modification to state or position will likely be a first target for intervention.

Respiratory Status If the patient requires any type of assistance with respiration (e.g., supplemental oxygen), the method of assistance (e.g., via nasal cannula), oxygen percentage, and flow rate should be noted. While flow rate itself does not necessarily contribute to dysphagia [27], it may indicate a degree of medical fragility that could predispose the patient to increased negative consequences of dysphagia and aspiration. For the infant, particular attention should be paid to respiratory rate. As previously described, infants feed in a pattern of suck-swallow-breathe that occurs at approximately a 1:1:1 ratio over the course of about 1 s. If the infant's respiratory rate is faster than 60 breaths per minute, (1 breath cycle per second), the infant will either need to suppress part of the suck-swallow-breathe pattern and subsequently "catch-up breathe," or the caregiver will need to implement external pacing of feeding. These infant-led and/or caregiver-led modifications allow the infant time to swallow the elicited bolus and to "catch up" from a respiratory perspective before additional bolus volume is elicited. If an infant is breathing at a

very fast rate and is neither self-pacing nor being externally paced, coordination breaks down, leading to some form of airway compromise.

Evaluation of the swallowing sensorimotor mechanism should be completed on all patients as a standard part of assessment [21, 28, 29]. This includes assessment of *strength*, *range of motion*, *symmetry of movement*, *coordination of movement*, and *structural integrity* of the mandible, lips, tongue, velum, and cheeks during speech and nonspeech tasks. Pharyngeal and laryngeal structure and function can be indirectly evaluated through testing of gag reflex, volitional cough, and voice quality. Additional reflex testing of phasic bite, tongue protrusion, and rooting should be completed for infants and younger toddlers. Because these reflexes are present in early infancy and are expected to fade over time, their absence (in early infancy) and presence (in later infancy and beyond) should be noted, as they can indicate neurologic dysfunction and have a negative impact on swallowing performance [5, 6, 30]. Modifications to swallowing sensorimotor mechanism assessment will be necessary, particularly for infants, toddlers, and school-aged children. As described earlier, the importance of establishing a good rapport is essential in obtaining information from a clinical assessment. Elicitation of a gag reflex or other testing that may be perceived to be invasive or unpleasant may lead the patient to refuse further participation or refuse bolus trials. This may be especially true for patients who demonstrate oral aversion or other oral hypersensitivity at baseline.

For infants, assessment with a gloved finger can provide a large amount of information about structure and function of the oral mechanism. Beginning with elicitation of the rooting reflex through gentle stroking of the cheek or chin lateral to midline, the clinician can assess suckling reflexes, anterior-posterior lingual range of motion, sensitivity to novel textures, and structural integrity of the palate. Particular attention should be given to palatal/velar palpation, with the clinician making note of any high palatal arching or soft areas that may suggest clefting. Strength of the suck and ability to generate intraoral pressure differentials can be assessed when the finger is withdrawn from the oral cavity. In most infants, intraoral pressure should be strong enough to cause a strong pull of the glove from the finger upon withdrawal. The clinician's smallest finger should be used, with the pad of the finger facing the palate and the nail facing the lingual surface. As above, if the infant is known to have oral hypersensitivity or oral aversion, care must be taken with intraoral palpation. If there is a risk that the infant will refuse all bolus trials following an aversive response to oral palpation, this should be deferred until after oral bolus trials are completed. As the infant continues to develop, tolerance for intraoral palpation will typically decrease. This often will coincide with presence of first teeth.

Bolus Trials

Initial evaluation of swallowing function should be completed through observation of the patient in a way that most closely approximates their typical feeding behaviors. The child should be positioned in a high chair, at a table, or in a caregiver's lap or cradled in the caregiver's arms in the same way that they are positioned at home. Types of food/formula/milk and method of bolus delivery (cup, bottle and nipple type, utensils) should match those of the home environment and/or the environment in which the swallowing behaviors of concern occur. The acute care setting is a potential exception to this more naturalistic method of evaluation, particularly if the typical feeding/swallowing environment and patterns may be considered likely to result in decreased swallow safety in a medically compromised patient.

In toddlers and older children, discrete trials of systematic volumes can be completed. Ideally, the patient will swallow boluses of increasing size (5 ml, 10 ml, 15 ml, etc.) and will sustain an /a/ vowel for approximately 5 s, count to 5, or perform some other automatic speaking task after each bolus trial [3]. The clinician will observe for signs of aspiration including coughing, choking, throat clearing, wetness or "gurgling" of voice quality, or congestion of breath sounds [17, 28]. Timing of these observations is also significant,

in that immediate coughing, choking, or throat clearing may indicate intact airway-protective reflexes in the setting of dysphagia, while weak and/or delayed responses (greater than 20 s after the swallow) may indicate reduced airway protection at the level of the larynx [31]. Following completion of a trial, the oral cavity should be inspected for residue. The location of the residue and response to attempts to remove the residue (automatic second swallow, cued second swallow, tongue swipes, liquid washes, etc.) will help the clinician in understanding the pathology of dysphagia. Additional signs of swallowing difficulty in the oral phase include immature biting or chewing in older children, oral residue, and anterior loss of the bolus from the oral cavity. Signs of difficulty in the pharyngeal phase include delay in swallow reflex or absent swallow reflex when palpating the larynx during the swallow, signs of laryngeal penetration and aspiration described above, and nasopharyngeal reflux.

In infants, discrete trials of systematic volumes are often not possible and are not likely to be ecologically valid. Instead, continuous feeding from the bottle or breast is observed. While overt signs of aspiration tend to be consistent across age groups, signs of swallowing difficulty are often more subtle in infants. Signs of distress can indicate swallowing difficulty [10, 21]. These signs include abrupt changes in facial expression (i.e., wide open "surprised" or tightly shut eyes, furrowed brows), back-arching, fist clenching, finger splay, increase in tone, or "shutting down" [32]. Loss of coordination of the suck-swallow-breathe cycle is a common sign of dysphagia in infants. This loss of coordination can be the result of a breakdown at any stage of the swallow, including preoral, oral, pharyngeal, and esophageal.

An additional sign of swallowing difficulty is the presence of inspiratory stridor that occurs during feeding only. Inspiratory stridor suggests some degree of airway construction during inhalation, whether at glottic, subglottic, or supraglottic levels. Stridor sometimes occurs as a result of the loss of coordination in the suck-swallow-breathe cycle. Specifically, the infant may retain an adducted glottic position (as a part of airway protection during swallowing) during inhalation. This pattern is often observed when bolus size is too large and/or flow rate is too fast for respiratory rate. There are, of course, several potential organic reasons for stridor. Common examples include laryngomalacia, vocal fold paralysis, and glottic web [33]. Stridor that results from organic causes tends to be present to some degree in all breathing situations and is simply louder with increased airflow during activities that require heavier respiration such as crying and feeding. When stridor is present only during feeding and breathing is quiet and comfortable at all other times, it may be due to the loss of coordination of the suck-swallow-breathe cycle. Regardless, when stridor is present, endoscopic airway visualization is warranted.

Modifications

Modification of patient position, bolus characteristics, and method of bolus delivery can help to both improve swallow function and to aid the clinician in describing the nature of the dysphagia. In older children, common interventions include softening solids, increasing textural or flavor stimulation of solids, thickening liquids, reducing bolus size, using different types of cups or straws, and potentially implementing chin tucks, head turns, and other positional modifications [2, 6, 18]. Every modification and intervention should be made with caution and with an explicit goal in mind. In order to effectively introduce modifications, the clinician must have a hypothesis about the reason a given sign of dysphagia is observed. For example, a child with a traumatic brain injury may have delayed or less-robust swallow reflexes. This could result in reduced coordination for airway protection and subsequent overt signs of aspiration with a thin liquid (although, in this particular scenario, aspiration is more likely to be silent and instrumental evaluation would likely be recommended). The clinician could thus reasonably test liquids of increased viscosity (nectar or honey), which could result in slower bolus flow rate and increased sensory input, leading to improved coordination for airway protection [34].

Table 16.3 Swallowing observations, common hypotheses of dysfunction, and subsequent flow rate intervention for hypothesis testing

Observation	Plausible hypothesis	Modification to flow rate
Overt coughing following uncoupling of sucking/swallowing from breathing	Oral cavity fills with a bolus volume that is too large, bolus either spills from anterior oral cavity or to the pharynx and invades the airway before the pharyngeal swallow can be initiated	Reduce flow rate
Anterior loss of bolus		
Collapse of the nipple, infant frustration/fussing, frequent disengagement from the nipple and small volume of intake	Feeding drive, nutritional/hydration requirement, and oral motor strength capacity are mismatched with rate of bolus flow	Increase flow rate

In young infants, modification of flow rate, position, and method of bolus presentation are the first lines of defense, with modification to bolus viscosity avoided when possible [35]. If the infant has difficulty attaining a calm, focused state while in a cradled upright position, for example, reduced organization for latching to the nipple and reduced coordination of the suck-swallow-breathe pattern could result. The clinician could consider swaddling and placement in a side-lying and semi-upright position during feeding, both of which have been found to assist in pain management, emotional regulation, and soothing [36]. Modification of flow rate (faster or slower) is another simple intervention that can help to test hypotheses for why some signs of dysphagia are observed. Table 16.3 describes some observations, hypotheses, and flow rate interventions that are common for infants.

Modification of liquid viscosity can be challenging in infants, and several factors must be taken into account [35, 37]. Studies have shown that use of thickening agents such as xanthan gum in premature infants can increase risk of necrotizing enterocolitis [35]. Additionally, many infants are breastfed or bottle-fed with breast milk, which does not react well to thickening agents. Other food substances are sometimes used to increase bolus viscosity (i.e., infant cereals, purees, etc.); however, the American Academy of Pediatrics recommends that these foods be introduced only later in infancy [38]. Coordination with gastroenterologists and dieticians is advised when it appears that viscosity will need to be modified for infants.

Summary

Clinical swallow evaluations are important tools that help dysphagia specialists to obtain a gross measure of swallow function. Clinical swallow evaluations also allow for hypothesis generation and testing regarding potential causes of and interventions for dysphagia, although additional information will often need to be obtained from instrumental assessments in order to more fully understand an individual's swallow function. For pediatric patients in particular, data obtained during clinical evaluation will streamline diagnosis and treatment, guide decision-making during instrumental assessments, and help to establish the positive rapport that is essential for effective intervention with pediatric patients with dysphagia.

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17 Videofluoroscopic Evaluation of the Swallow in Infants and Children

Bryn K. Olson-Greb

Overview

Dysphagia is defined as a disruption in swallowing that compromises a patient's ability to eat or drink safely and efficiently and/or to adequately meet nutritional and hydrational needs through oral intake [1]. It is important to distinguish dysphagia, which is anatomically and/or physiologically based, from a feeding disorder. Feeding disorders are behavioral and characterized by an unwillingness to consume foods or liquids that is not related to a skill deficit. Dysphagia and feeding disorders can co-occur; however, evaluation for these two disorders is disparate. Videofluoroscopic swallow study (VFSS), is a method of evaluating for dysphagia

Medically complex children who are at risk for oropharyngeal dysphagia, particularly those with neurological compromise, may not demonstrate clinical signs of aspiration such as coughing or choking [1–3]. It is extremely important in these cases that the clinician take the limitations of clinical evaluation into consideration so that, when appropriate, instrumental evaluation can be used to provide an accurate diagnosis and appropriate treatment plan. Instrumental evaluation can help to more objectively characterize the anatomic and physiologic nature of a dysphagia, determine risk for aspiration, guide decisions regarding the optimal setup for swallowing safety, and determine the need for additional intervention. One option for instrumental assessment of pediatric dysphagia is the videofluoroscopic swallow study (VFSS), also referred to as a modified barium swallow study (MBSS). The procedure is typically performed by a speech-language pathologist in conjunction with a radiologist.

The speech-language pathologist's role includes presenting foods and liquids of different consistencies mixed with barium, a radiopaque contrast that allows for visualization of the bolus as it moves through the oropharynx and into the esophagus. Interpretation of the resulting images can identify issues with the oropharyngeal swallow. The speech pathologist is also responsible for testing various modifications to positioning, texture, viscosity, etc. to determine appropriate recommendations for safe and efficient feeding [4]. This chapter provides an introduction to pediatric VFSS, including indications for instrumental swallow evaluation, purposes, advantages and limitations of VFSS, and the role of the speech-language pathologist (Table 17.1).

For patients who are not medically stable enough to bring to the fluoroscopy suite but are determined to be appropriate for instrumental swallowing evaluation, flexible endoscopic

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Table 17.1 Indications and contraindications for videofluoroscopic swallow study (VFSS)

Indications	Contraindications
Need to thoroughly evaluate oropharyngeal swallowing function	Insufficient medical stability to tolerate the evaluation or transport to the fluoroscopy suite
Presence of feeding concerns or poor weight gain for which oropharyngeal dysphagia is a suspected contributing factor	Patient-specific factors that would preclude adequate participation (e.g., cognitive impairment, inadequate alertness, limited willingness to accept oral intake due to severe oral aversion)
Presence of unexplained respiratory complications (e.g., chronic cough, recurrent pneumonia, or frequent or prolonged upper respiratory infections)	Instances in which the evaluation would not change the patient's plan of care [4]
Presence of clinical signs or symptoms of oropharyngeal dysphagia, penetration, or aspiration	
Presence of significant risk factors for oropharyngeal dysphagia, even in the absence of clinical signs or symptoms	
Need to evaluate strategies (e.g., positioning; bottle flow rate; liquid viscosity; need for implementation of an exercise-based dysphagia program) to determine appropriate recommendations for treatment and safe, efficient feeding	
Need to re-evaluate swallowing function in a patient with a history of dysphagia and a suspected change in swallowing function	
Inability to complete flexible endoscopic evaluation of swallowing (FEES) due to poor tolerance or anatomic abnormalities which limit visualization of the relevant laryngopharyngeal structures or preclude passage of a nasendoscope	
Need for additional information in cases where FEES identifies but cannot fully define the characteristics of an oropharyngeal dysphagia	

evaluation of swallowing (FEES), described in detail in the chapter by the same name, may be a viable alternative.

Arvedson and Lefton-Greif proposed four principle factors the clinician should consider when determining if recommendation for VFSS is appropriate [5]:

1. Do patient history, clinical evaluation, or both indicate suspicion for oropharyngeal dysphagia?
2. Do the results of the VFSS have the potential to clarify diagnosis and inform management?
3. Is the child ready and able to participate in the study?
4. Will the findings impact the child's care?

Careful consideration of these factors and employment of sound clinical judgment will ensure that VFSS is appropriately utilized and that nonessential evaluations are avoided. These same questions are applicable to the determination of appropriate timing for re-evaluation. Additional consideration should be given to the level of suspicion for change in swallowing function and whether confirmation of such a change would meaningfully change management. For example, a repeat VFSS may be necessary to determine when it is appropriate to advance the diet of a patient with silent aspiration; however, repeating the study too soon may result in unchanged results and the need for additional studies to facilitate future care management.

Purposes of VFSS

To ensure optimal utilization of VFSS and the information it can provide, it is of the utmost importance that the clinician understand that

VFSS is not simply a tool to determine whether a patient is aspirating. The primary purposes of VFSS include identifying and defining abnormal anatomy and physiology contributing to the patient's symptoms and evaluating potential treatment strategies to enable safe, efficient, and/or adequate oral intake [6].

Structural (congenital or acquired), neurological, respiratory, cardiac, metabolic, and inflammatory disorders can all have an impact on feeding and swallowing in pediatric populations [1, 7, 8]. Aspiration and inefficient swallowing can occur for a variety of reasons including delayed or absent pharyngeal swallow trigger, inadequate tongue base retraction or pharyngeal constriction, inadequate laryngeal vestibular closure, and dysfunction of the UES. Treatment varies depending on etiology; thus determination of the anatomic and physiologic origin of the problem is essential to providing appropriate and effective intervention.

Advantages and Limitations

Advantages and limitations of videofluoroscopic swallow studies (VFSS) are similar for adult and pediatric patients [7, 9]. VFSS allows for dynamic visualization of the oral, pharyngeal, and upper esophageal phases of swallowing, including timing and coordination of the events of swallowing, pharyngeal motility, presence of residue, presence of penetration or aspiration, and functional contributions to unsafe or inefficient swallowing [10, 11] (Table 17.2).

VFSS is noninvasive, which can facilitate cooperation and allow for instrumental assessment of patients who are not able to tolerate a flexible endoscopic evaluation of swallowing (FEES); however, the procedure requires the administration of foods and liquids impregnated with barium, the taste, texture, and appearance of which may be unappealing to some patients and decrease willingness to participate. A sufficient amount of barium must be consumed during the study to allow for adequate evaluation of swallowing function; thus VFSS would not be appropriate for a patient with significant oral aversion or limited experience with oral intake [7, 12]. The need for use of radiopaque barium also precludes the evaluation of breastfeeding via VFSS.

A major disadvantage of VFSS is radiation exposure, both for the child and for the parent or caregiver tasked with feeding the child during the study [7, 12]. Radiation exposure can be managed by limiting the "fluoro-on" time to a maximum of 2–3 min unless a longer study is absolutely necessary [10]. The need for relatively brief "fluoro-on" time is another limitation of the exam, as this "snapshot in time" may not capture deficits that are infrequent or inconsistent.

VFSS must be completed in the fluoroscopy suite, meaning that the patient must be able to tolerate being transported. The equipment in the suite itself may be intimidating for some children, which may result in a nonrepresentative feeding. Patients must be positioned such that the relevant structures can be visualized, which may preclude evaluation in their natural feeding position. VFSS can yield highly useful information about oropharyngeal swallowing function; however, to obtain an optimal study, it is important that clinicians understand the limitations of VFSS such that patient selection and procedure planning may be performed appropriately.

Speech-Language Pathologist Approach

History

Ideally, children with suspected oropharyngeal dysphagia should be seen for a clinical swallowing evaluation prior to undergoing VFSS. During the clinical evaluation, the SLP can gather a thorough medical and swallowing history, complete an oral mechanism exam, and observe a typical feeding. Information about the typical conditions under which the child feeds, optimal positioning for feeding, consistencies the child typically consumes, and consistencies that elicit swallowing complaints can be obtained during this evaluation [9]. The day of VFSS, the evaluating clinician should obtain a swallowing history detailing, at a minimum, the following:

Table 17.2 Comparison of VFSS and FEES

	VFSS/MBSS	FEES
Description	Instrumental swallow evaluation utilizing fluoroscopy and administration of various consistencies of a contrast (barium) to evaluate swallowing function	Instrumental swallow evaluation utilizing a flexible nasendoscope to evaluate swallowing function
Location	Fluoroscopy suite (radiology)	Bedside or clinic
Anatomy	Grayscale images of the oral cavity, pharynx, and cervical esophagus	Full color video of hypopharynx and larynx
View	Sagittal and A/P view of the head and neck	Superior view of pharynx and larynx
Contrast	Barium	Green/blue/white food dye *(may also use barium or naturally green or white food)*
View of swallow	Comprehensive view of all swallowing phases – oral, pharyngeal, cervical, esophageal phases	Able to view pharyngeal phase except for brief (less than 1 s) "white out" during height of swallow. Unable to view oral phase or esophageal phase
Advantages	Can assess all phases of swallow including the moment of the swallow	Can evaluate management of secretions
	Noninvasive and generally well tolerated in pediatric population	Can evaluate swallow with very small volumes
	Radiologic view of structure and function	Can be conducted in most locations including clinic and bedside
	Can modify position, viscosity, feeding equipment, and strategies during the assessment	Can evaluate laryngeal and pharyngeal structures from superior view
	Can detect presence of structural abnormalities such as type I laryngeal cleft, tracheoesophageal fistula (TEF), etc. that are difficult or impossible to visualize on nasendoscopy	Allows for assessment of breastfeeding
		Allows for assessment of child or infant's typical foods
		No time constraints
		No radiation exposure
		Potential assessment of sensory threshold
		Can modify position, viscosity, feeding equipment, and strategies during the assessment
		Easy to position child in parent's lap for comfort and typical feeding
Disadvantages	Radiation exposure and time constraints	Nasendoscope can be uncomfortable and is not tolerated well by all children
	Can be an unfamiliar setting for young children	Does not allow for evaluation of the moment of swallow due to "white out" during the swallow
	Cannot evaluate child's typical foods without addition of barium	Does not allow for evaluation of oral or cervical esophageal phase
	Cannot evaluate breastfeeding	May be difficult to evaluate for aspiration due to anatomic interference (arytenoids or epiglottis obscuring view)
	Brief snapshot of swallow function due to time limitations	
	Does not allow for visualization of saliva/secretion management, as these substances are not radiopaque	

- Duration, onset, and characteristics of swallowing complaints
- Current diet
- Components of a typical feeding, including positioning, use of specific feeding implements (bottles, spoons, cups, etc.), or any other adaptations employed in the child's home environment
- History and timing of any past pneumonias or pulmonary compromise
- Recent concerns with growth or weight gain
- Impact of swallowing impairment on quality of life
- Other contributory factors

A thorough case history can facilitate VFSS planning by clueing the clinician into possible etiologies for the swallowing problem and identifying variables (e.g., bolus consistency, flow rate) that should be assessed during the procedure.

Preparing the Patient and Parent/Caregiver

In preparation for a pediatric VFSS caseload, optimization of the physical setup of the fluoroscopy suite can be beneficial. Try to keep the environment child-friendly and inviting. If available, draw on the expertise of your facility's child life specialists. These professionals have specialized training in infant, child, and adolescent development and well-being and use strategies such as play, developmentally appropriate communication, and psychological preparation to help children and families cope with and minimize adverse effects of healthcare experiences [13, 14].

Adequate preparation of the patient and parent/caregiver is an essential step to ensure a successful evaluation. The purpose and logistics of the procedure should be described to the caregiver in advance so that they may help to prepare the patient. Whenever possible, the parent should accompany the child into the fluoroscopy suite so as to keep the child at ease. Particularly for young children, using parents as feeders may increase cooperation and acceptance of the foreign barium products [10]. Ideally, families should be encouraged to schedule the study at a time of day when the child is alert, rested, and well-regulated. Instruct families to withhold food and liquid for up to a few hours prior to the study, as thirst and hunger may increase the child's willingness to consume the test barium products; however, keep in mind that discomfort from being overly hungry may result in reduced willingness to participate in some children. Caregivers should be instructed to bring feeding implements (e.g., cups, bottles, spoons, containers) that the child typically uses at home and a variety of preferred foods, including consistencies that have presented a problem in the past. For young children, bringing a security item such as a favorite toy or lovey may help the child to feel secure.

Positioning

Positioning of the pediatric patient is an important consideration, as one must balance the need to visualize the structures involved in the oropharyngeal swallow with the need to evaluate the impact of positioning on swallowing function [15]. There are a wide variety of chairs on the market which can facilitate optimal positioning [10, 11]. When possible, start the evaluation with the patient in his or her typical feeding position so as to evaluate what swallowing function looks like under natural feeding conditions. If possible, use a seat (high chair, adaptive chair, etc.) similar to that which the child uses at home. Consider the physical needs of the child, and, when appropriate, reach out to the child's occupational therapist (OT) in advance of the evaluation regarding recommendations for optimal positioning. If the child exhibits poor head, neck, or trunk support, work to position the child such that they are adequately stable to support optimal swallowing. The child's head should be at midline with the spine in neutral alignment, shoulders slightly forward, lower extremities relaxed and slightly flexed, feet flat and well-supported, and hips in natural alignment [16]. Towels may be used to facilitate midline positioning of the head, bring shoulders forward, and align hips [7]. Sidelying or sidelying semi-upright positioning can be considered for children with issues such as poor muscle tone, micrognathia, or laryngomalacia. This can be accomplished by placing the table horizontally or at a slight incline and positioning the patient directly on the table. A wedge may be used to help keep the patient in place. Regardless of the chair or positioning strategy used, it is important that any adaptive seating setup be adequately secured to ensure the safety of the patient.

Children with medical complexity or those considered to be at high risk for aspiration may require additional equipment and monitoring to ensure safety during VFSS. When needed, suctioning equipment should be available, and

personnel or caregivers comfortable administering suctioning should be present in the fluoroscopy suite throughout the study. Children who require O2 supplementation should be hooked up to a portable oxygen tank. Cardiac monitors, respiratory monitors, and pulse oximetry should be available for at-risk patients and personnel versed in reading the output from these monitors and assisting in the event of a status change should be on hand [17].

Procedure

VFSS is typically performed by a speech-language pathologist (SLP) in conjunction with a radiologist or radiology technician who operates the machinery, documents any structural findings, and monitors the overall safety of the study such that it can be terminated if the patient's safety is at risk. In cases where the radiologist is not in the room during the study, he or she should be consulted as necessary to confirm any structural findings. Other specialists, such as otolaryngologists or gastroenterologists, can review the VFSS as needed to provide additional input in cases where issues requiring medical management are identified. During VFSS, the SLP is responsible for determining the order, size, and type of barium-impregnated foods and liquids to be presented, selecting appropriate feeding implements (bottle with specific nipple flow; special utensils, special cups, etc.), interpreting the data obtained from the study, testing feeding modifications (altered bolus flow rate, modified cups or spoons, modified viscosity, changes in posture/positioning, implementation of external pacing, etc.), and making recommendations for diet, feeding modifications, or other therapeutic interventions indicated based on the results of the study.

Patients can be viewed laterally, which enables visualization of the timing and coordination of movement of the oropharyngeal swallowing structures as well as identification of airway invasion [17]. An anterior-posterior (AP) view can provide information regarding the symmetry of velopharyngeal elevation, laryngeal and pharyngeal structures, vocal fold movement, and bolus passage through the pharynx [17], and the function of the upper esophageal sphincter. Previously published texts by Arvedson and Lefton Grief (1998) and Logemann (1998) include detailed explanations of the basic technique for performance of VFSS and continue to serve as excellent references for the dysphagia clinician [6, 17]. Accurate interpretation of VFSS is of critical importance and has been shown to improve with training [18]. Clinicians are strongly encouraged to seek out focused didactic experiences to improve ability to identify the anatomy of interest in pediatric patients as well as the ability to define oropharyngeal swallowing function and disorder based on videofluoroscopic images (Figs. 17.1, 17.2, 17.3, 17.4, 17.5, 17.6, 17.7, 17.8, 17.9, 17.10 and 17.11).

Information that can be obtained from VFSS and used to determine appropriate interventions includes:

- Oral control of the bolus
- Effectiveness of oral transport
- Velopharyngeal function, including coordination and completeness of closure; presence of oronasal backflow (Fig. 17.7)
- Hyolaryngeal elevation

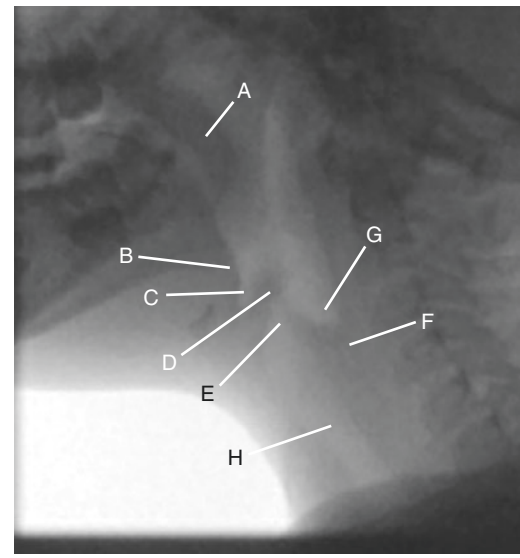

Fig. 17.1 VFSS Image with structures labeled. **A**: soft palate; **B**: base of tongue; **C**: vallecula; **D**: epiglottis; **E**: laryngeal vestibule; **F**: UES (upper esophageal sphincter) **G**: pyriform sinuses; **H**: trachea

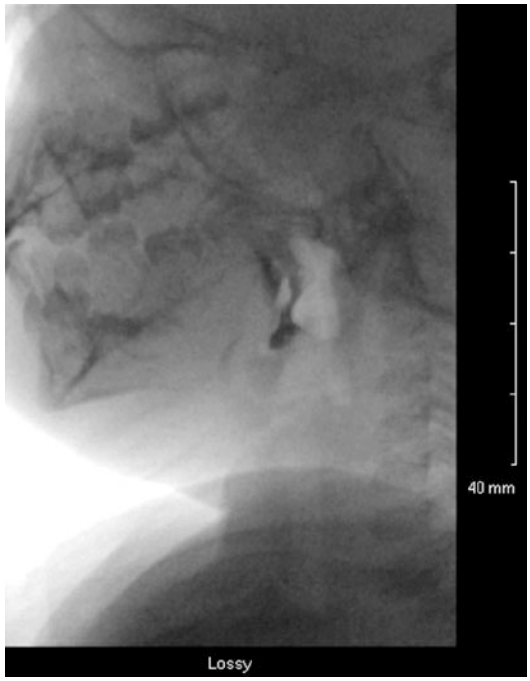

Fig. 17.2 Delay, valleculae

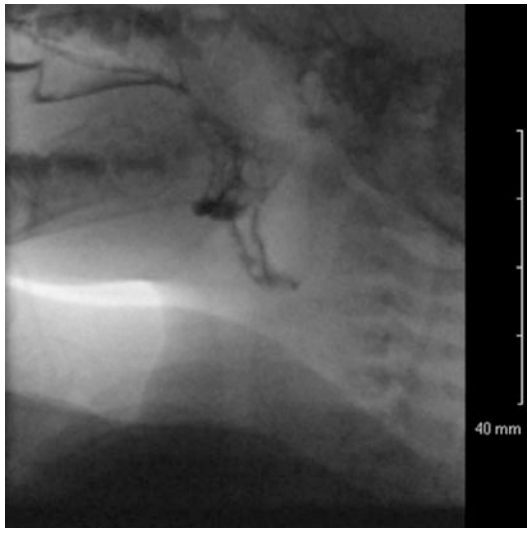

Fig. 17.3 Delay, piriform sinuses

- Airway protection, including timing and completeness of true vocal fold closure, compression of the supraglottic structures, and epiglottic inversion
- Timing of pharyngeal swallow trigger
- Coordination of the events of the pharyngeal swallow

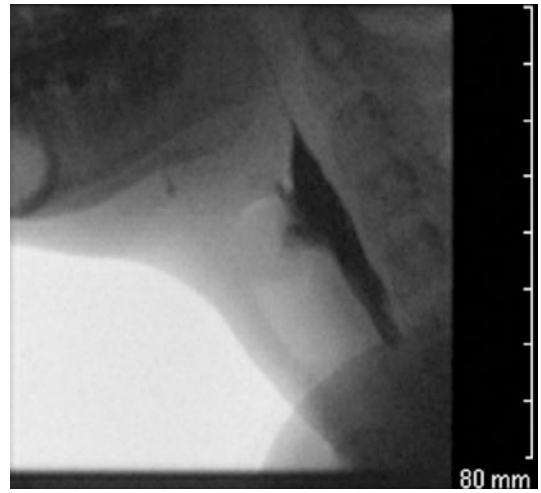

Fig. 17.4 Normal UES distention

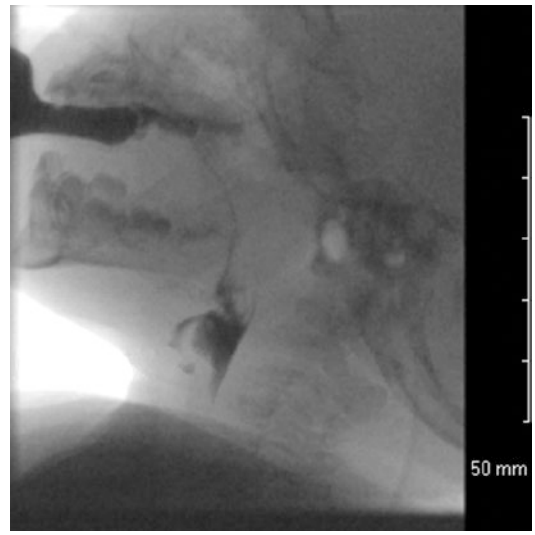

Fig. 17.5 Aspiration during the swallow

- Pooling of bolus material in the vallecula or pyriform sinuses before the swallow (Figs. 17.2 and 17.3)
- Duration of bolus dwell time in the pharynx
- Efficiency of pharyngeal clearance/presence of residual bolus material in the pharynx
- Number of swallows required for pharyngeal clearance
- Presence and timing of airway invasion (penetration or aspiration) (Figs. 17.5, 17.8, 17.9 and 17.11)
- Patient response to airway invasion, when present

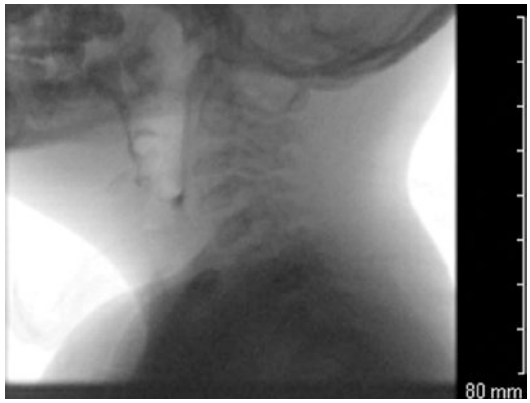

Fig. 17.6 Residue, base of tongue and above the upper esophageal sphincter

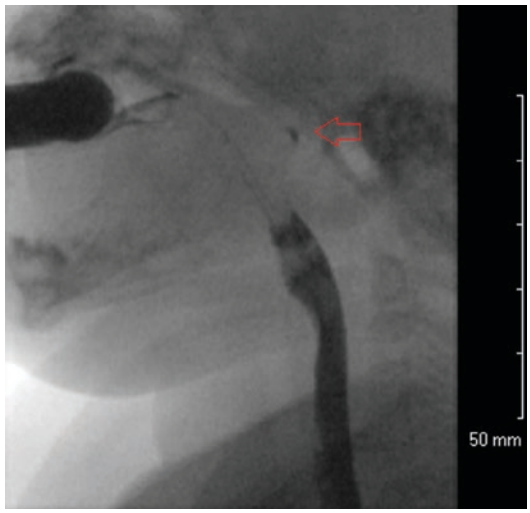

Fig. 17.7 Oronasal backflow

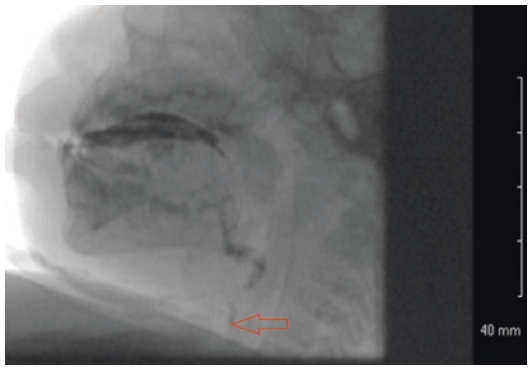

Fig. 17.8 Aspirate in the trachea

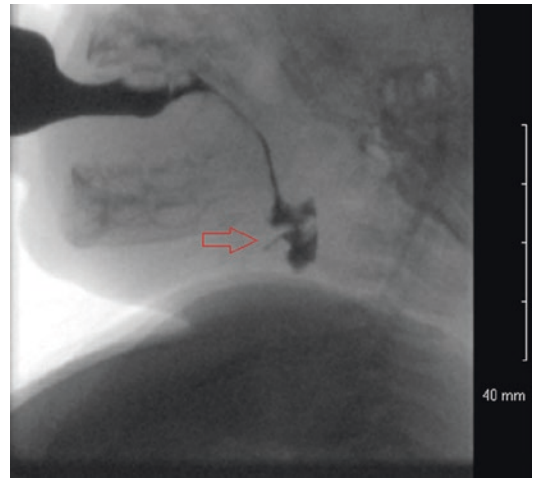

Fig. 17.9 Penetration

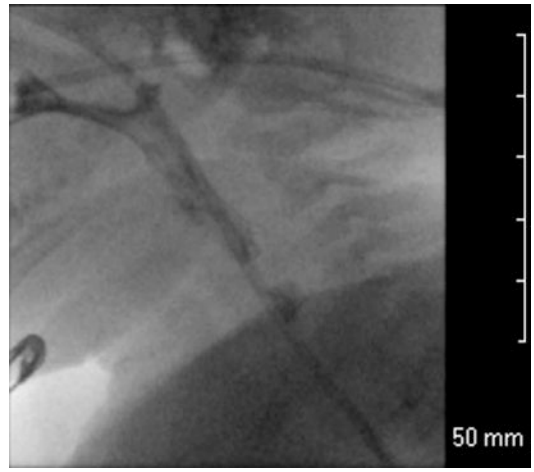

Fig. 17.10 Inadequate UES distention

VFSS should test the full range of developmentally appropriate consistencies. Additionally, it is important that the viscosity of the fluids tested during VFSS matches what is recommended to the patient [1]. Studies have compared the rheological and material properties of the barium liquids used on VFSS to those of "mealtime fluids," meaning those fluids, such as thickened and unthickened infant formulas, that patients typically ingest outside of the context of VFSS. Results indicated inconsistencies in the density, viscosity, and yield stress of mealtime

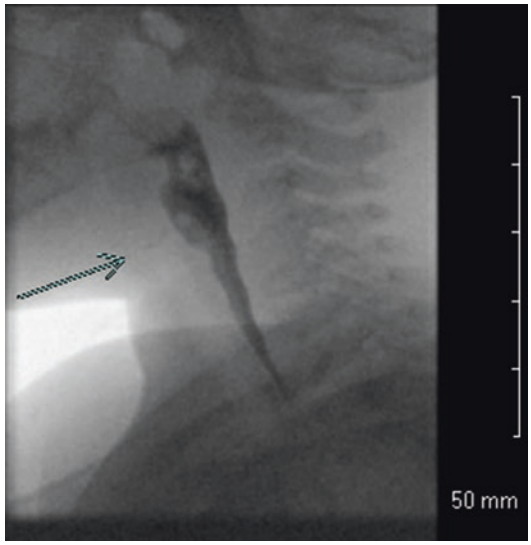

Fig. 17.11 Level 4 penetration

fluids versus barium products, suggesting that swallowing function observed on VFSS may not be consistent with that of a typical feed [19, 20]. This may present problems with respect to recommendations and highlights the need to objectively match recommendations for mealtime fluids with the rheological properties of the fluids used on VFSS. Subjective evaluation of liquid viscosity has been shown to be unreliable [20]; thus clinicians are encouraged to incorporate an objective tool such as the Line Spread Test [21, 22] or the International Dysphagia Diet Standardization Initiative (IDDSI) framework [23–27] into clinical practice. Information about the IDDSI can be found at http://iddsi.org.

Standardization of the VFSS protocol should be considered, with important elements including size and order of bolus presentation, frame rate, and instructions given to the patient [5]. Standardization can facilitate the making and quantification of diagnostic observations, allow for comparison within and between patients, decrease the need for multiple repeat studies, and improve patient outcomes [28, 29]. As of yet, no standardized protocol for pediatric VFSS has been universally agreed upon. In the adult population, the *Modified Barium Swallow Impairment Profile (MBSImP)*™© was developed to standardize the performance, interpretation, and documentation of VFSS in an effort to improve the validity, reliability, and reproducibility of these evaluations [30–32]. The *MBSImP*™© includes clinician training and calibration, a protocol for bolus presentation, specific patient instructions, and a standardized tool for scoring and documenting the various components of the oropharyngeal swallow [30]. It is intended to be used in tandem with the validated *penetration-aspiration scale (PAS)*, which is an eight-point ordinal scale used to rate the depth of airway invasion, when present (Table 17.3) [33]. Work is currently underway by Lefton-Greif and colleagues to develop a similar tool for use with bottle-fed children that will incorporate swallowing components, such as suck-swallow-breathe coordination, that are unique to the infant and pediatric populations [29].

As it currently stands, there are differing opinions in the literature regarding the ideal order of bolus presentation [11]. It is advisable that the clinician consider a standard protocol to use as a framework when performing VFSS; however, it is important to recognize that the primary objective of the assessment is to gather the information necessary to make appropriate recommendations for management of the patient's safe and efficient feeding, which may require altering the protocol. Variable approaches to

Table 17.3 Eight-point penetration-aspiration scale

1. Material does not enter the airway
2. Material enters the airway, remains above the vocal folds, and is ejected from the airway
3. Material enters the airway, remains above the vocal folds, and is not ejected from the airway
4. Material enters the airway, contacts the vocal folds, and is ejected from the airway
5. Material enters the airway, contacts the vocal folds, and is not ejected from the airway
6. Material enters the airway, passes below the vocal folds, and is ejected into the larynx or out of the airway
7. Material enters the airway, passes below the vocal folds, and is not ejected from the trachea despite effort
8. Material enters the airway, passes below the vocal folds, and no effort is made to eject

From Rosenbek et al. [33], with permission

bolus presentation have been suggested in the literature and include starting with the consistency of highest concern [10], that which is thought to be the safest [34], that which is least likely to result in pharyngeal residue [17], or that which is the child's favorite and thus most likely to facilitate cooperation [11]. Lacking clear evidence of one approach over others, the clinician should employ clinical judgment and consider patient-specific factors including patient history, medical fragility, observations made during clinical evaluation, and likelihood that the patient will cooperate long enough to evaluate multiple consistencies when deciding how to proceed.

Presence of Feeding Tubes

The impact of the presence of a nasogastric (NG) tube on pediatric swallowing is not fully understood. Historically, the recommendation has been to remove the tube prior to completion of VFSS to eliminate the risk of the presence of the tube impacting work of breathing, O2 saturation, or velopharyngeal closure [5]. However, studies have suggested that presence of an NG tube does not have a significant impact on oropharyngeal swallowing function or VFSS findings [35, 36]. As such, performing VFSS with NG in place may be appropriate. Risk for aspiration pneumonia when aspiration is present may be higher in patients with an NG tube in place due to the higher likelihood that gram-negative bacteria, which increases pneumonia risk, is present in secretions [37]. This may be an important consideration when making safe swallowing recommendations for this patient population.

For all patients who receive primary nutrition and hydration NG tube or gastrostomy tube, the clinician must balance the need to evaluate swallowing function and determine appropriate recommendations for safe oral intake with the risk that patients with limited experience eating by mouth may be unwilling to take enough contrast during the study to allow for a thorough evaluation. If the patient's medical team is in agreement, patients should be given the opportunity to try therapeutic tastes of the consistencies that will be tested during VFSS for 1–2 weeks in advance of the study [11].

Technical Considerations: Pulse Rate

Though it is unclear whether low levels of radiation exposure associated with medical procedures may result in increased cancer risk [10], pediatric patients are thought to be up to ten times more radiosensitive than adults [38]. Radiation dosage is an important consideration in the performance of VFSS, as fluoroscopy is a major contributor to medical radiation exposure [39].

VFSS, like all medical procedures that utilize ionizing radiation, must comply with the as low as reasonable achievable (ALARA) principle, meaning that radiation must be limited to only that which is necessary to obtain the required diagnostic information [10, 38]. Reducing pulse rate reduces radiation dosage [40]; thus clinicians may encounter radiologist preference for or facility or health system-wide policies dictating the maximum pulse rate that may be used. Studies by Cohen and Bonhila et al., among others, have demonstrated that airway invasion may be missed and ability to accurately judge swallowing impairment may be negatively impacted at pulse rates lower than 30pps [41, 42]. Current evidence supports use of 30pps, also referred to as continuous fluoroscopy [5]. Studies performed below this rate should be interpreted with caution, and clinicians need to work with physicians and medical administration to ensure that an appropriate pulse rate may be used to allow for optimal studies that accurately describe swallowing impairments and allow for appropriate recommendations and interventions.

Feeding Modifications and Recommendations

One of the primary purposes of VFSS is to evaluate feeding strategies to assist in providing appropriate recommendations for safe and effi-

cient oral intake. Interventions vary based on the presence and timing of penetration or aspiration (before, during, or after the swallow), the consistency on which penetration or aspiration occurred, and any observed abnormalities in pharyngeal motility [10]. Several factors have been identified as relevant to aspiration risk. These include tongue strength, hyolaryngeal movement, prolonged stage transition duration, respiratory rate, and the phase of respiration that is interrupted by the swallow [1]. Chronic aspiration can be detrimental to respiratory health [43]; thus when aspiration is observed or oropharyngeal swallowing dysfunction suggestive of high risk for aspiration is noted on VFSS, implementation of appropriate feeding modifications is warranted. Modifications that can be trialed during VFSS vary somewhat depending on patient age and ability to participate. Modifications generally fall into three broad categories [17], including:

- Bolus adaptations (e.g., specialized cups, nipples, or other feeding implements to control bolus size and/or flow rate; liquid viscosity, food consistency, or texture; temperature; flavor (e.g., sour)) or bolus presentation (e.g., placement on one side of the mouth; use of downward pressure on the tongue with bolus placement)
- Posture or positioning modifications (e.g., upright, reclined, or sidelying positioning; chin tuck; head turn; head tilt)
- Behavioral modifications (e.g., volitional cough or swallow maneuvers such as the effortful swallow or Mendelsohn maneuver)

Selection of appropriate modifications varies depending on the nature of the swallowing deficit. Examples include use of a faster flowing nipple to address impaired oral bolus transfer; altered bolus size, texture, or temperature to address delayed pharyngeal swallowing trigger; use of a head turn or sidelying position (head turn toward weak side or positioning on strong side) in the setting of unilateral weakness or paralysis; use of a supraglottic swallow in participatory patients with penetration or aspiration that occurs during the swallow; use of a slower flowing nipple or increased liquid viscosity in nonparticipatory patients demonstrating penetration or aspiration before or during the swallow; or use of an effortful swallow or multiple swallows to reduce pharyngeal residue [17]. This list is far from exhaustive. The skilled clinician must be well-versed in the many options and indications for feeding and swallowing modifications so as to test these appropriately during VFSS and make appropriate recommendations based on observed results [6, 10, 17, 44].

It is important to note that any behavioral intervention is only effective when applied consistently and requires adherence on the part of the child and/or family. Thickening liquids, which results in slower-flowing, more cohesive boli, has the advantage of not relying on the patient to be participatory. As such, this strategy can be used in young children and patients with intellectual impairments that preclude successful implementation of other feeding modifications. Use of commercial thickeners with infants, however, can be problematic and is somewhat controversial.

There are a variety of commercial thickeners on the market, as well as alternative substances that can be added to liquid for the purposes of thickening. Thickening products can include gum- or starch-based commercial thickeners, oatmeal cereal, rice cereal, yogurt, pudding, xanthan gum, guar gum, baby food puree, potato flakes, chia seeds, or blenderized solid foods mixed with liquid. Though a causal relationship has not been clearly established at this time, several studies have suggested an association between use of xanthan gum-based thickeners and development of necrotizing enterocolitis (NEC) in premature infants [45–48]. Use of rice cereal as a thickener has come under scrutiny due to high concentrations of arsenic [49, 50]. Additional concerns with the use of thickened liquids include the impact of thickening agents on hydration and nutritional absorption, caloric content, and risk of additional adverse effects including diarrhea or constipation [51].

Evidence supporting the safety and efficacy of thickeners in the pediatric population is generally lacking, and additional research is needed to determine what level of caution clinicians should employ in recommending use of thickeners for children with dysphagia [51]. Ideally, position changes such as elevated sidelying positioning, which may improve physiologic stability in infants [52, 53], altered flow rate, or use of behavioral strategies such as external pacing should be trialed first, and thickening should be considered only if these options prove inadequately effective. When thickener must be used, the recommended liquid viscosity should be as thin as possible while still ensuring safe swallowing. Patients for whom thickening is recommended should continue to be followed by a speech-language pathologist and appropriate medical team, and the use of thickener should be stopped as soon as it is deemed safe to do so. Patients may benefit from implementation of a systematic weaning protocol with incremental decreases in liquid viscosity to facilitate successful return to thin liquids and reduce the need for serial VFSS [54]. Research looking at the efficacy of such protocols is ongoing.

Young patients may refuse thickened liquids due to the change in texture. Bottle-fed infants may experience difficulty with efficient bolus extraction, resulting in fatigue during feeding. Difficulty with bolus extraction can be managed by selecting a faster flow, y-cut, or crosscut nipple. Bolus management with an appropriate nipple should be tested during VFSS to ensure that safe and efficient oropharyngeal swallowing function is maintained. Clinicians should be aware of research looking at the flow rates of commercially available nipples and recognize that studies have demonstrated variability in flow rate even within like nipples of a given brand [55–57].

Upon completion of VFSS, results and recommendations should be reviewed with the child and caregivers. Ideally, the recorded study should be reviewed and discussed to provide patients and caregivers with a more concrete understanding of findings [17]. Speech pathology recommendations following VFSS can include:

- Feeding or swallowing therapy targeting development of oral feeding skills, oral aversion, monitoring of diet tolerance, or an exercise-based dysphagia program
- Route for nutrition (oral or consideration of alternative means of nutrition if patient is not able to meet nutrition and hydration needs orally)
- Appropriate modifications for safe and efficient swallowing
- Communication with referring physician or PCP regarding consideration of referrals to additional specialties, for example:
  - Consideration of referral to a registered dietitian (RD) in the setting of functional oropharyngeal swallowing but poor growth
  - Consideration of referral to additional specialties, such as otolaryngology, gastroenterology, pulmonology, neurology, or plastic surgery when issues that may require additional medical intervention are identified

When used appropriately by clinicians with adequate training, strong critical thinking skills, and support from an appropriate multidisciplinary team, VFSS can serve as a powerful tool to guide the care and management of children with oropharyngeal dysphagia. Clinicians are strongly encouraged to seek out some of the resources cited at the end of this chapter and to stay current with respect to new areas of research pertaining to VFSS to ensure optimal utilization of this evaluation.

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Pediatric Flexible Endoscopic Evaluation of Swallowing

18

Jennifer L. Maybee, Bridget M. Harrington, and Jeremy D. Prager

Introduction

Flexible endoscopic evaluation of swallowing (FEES) has long been considered one of the gold standards for assessment of swallowing in both pediatric and adult populations [1–3]. In both children and adults, FEES is used to evaluate the structure and function of the upper airway, secretion management, pharyngeal swallowing function, and effectiveness of strategies to improve the safety and efficiency of swallowing. In pediatric FEES, one must consider development, growth, and relative positions of the pharynx and larynx [4–7]. In addition, neurophysiologic maturation including primitive reflex integration, coordination of central pattern generator-mediated feeding/swallowing behaviors, and maturation of respiratory/swallow coordination must be well understood [8].

The purpose of this chapter is to describe the procedures and strategies unique to FEES in the pediatric population. Prioritization of exam goals, roles of team members, and strategies for obtaining an optimal exam are discussed. The parameters of the FEES exam are discussed from a pediatric perspective. Practical considerations for the pediatric FEES clinic and case studies highlighting FEES in various pediatric populations are presented.

Historical Perspective

The use of flexible endoscopes to evaluate swallowing was first described by Susan Langmore in 1988. FEES was first added to the American Speech and Hearing Association (ASHA) scope of practice for speech-language pathologists in 2001 [2]. Technological advancements have occurred including development of endoscopes with smaller diameter, thus improving patient tolerance of the exam. Development of high-definition distal chip endoscopes has greatly improved the quality of images obtained. The use of FEES in the pediatric population was first described by Willging in 1995 [9] and described as a successful assessment of pediatric swallowing function in a population of 568 pediatric patients by Hartnick et al. in 2000 [1]. FEES has been described in the literature as being an effective tool for evaluating the pharyngeal swallow in a wide range of complex pediatric patients, including acute care [10], breastfeeding infants [11], and infants in the neonatal intensive care unit (NICU) [12, 13].

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Fees and Videofluoroscopic Swallow Study in Pediatric Swallowing Assessment

FEES and the videofluoroscopic swallow study (VFSS) are the most commonly used and widely available instrumental assessments of pediatric swallowing function. When selecting an instrumental swallowing exam, the goals of the exam and practicality of the exam are considered. In some cases, both exams may be utilized to thoroughly evaluate a complex swallowing problem, as different and complementary information can be gleaned from FEES and VFSS (Table 18.1).

FEES has been found to be equally or more sensitive than the VFSS in detecting parameters of swallow dysfunction such as penetration, aspiration, and pharyngeal residue [3, 14–16]. Assessment with FEES yields more accurate identification of anatomical markers and assessment of the location of pharyngeal residue [16]. FEES has been found to be *equally effective and valuable* compared to the VFSS for evaluation of swallowing [3] but may not be *interchangeable*, as FEES has been shown to yield higher penetration-aspiration scores and more severe ratings of residue [14, 15]. Guidance regarding diet and health outcomes after the FEES and VFSS have been found to be comparable [16].

Specific patient populations may benefit more from FEES versus VFSS, including breastfeeding infants, children with concerns regarding secretion management, patients who consume very small volumes of food/liquid orally, and children who struggle with efficiency of feeding and are not be able to complete an exam within radiology time constraints. In addition, individuals with known structural and upper airway anomalies may benefit from a FEES as it provides a direct view of upper airway structures and their function during swallowing. FEES is not ideal for individuals without a patent nasal passage, upper airway obstruction, or concerns for poor participation in the exam (Table 18.2).

Table 18.1 FEES vs VFSS

	FEES	VFSS
View	Superior view of the pharynx and larynx	Sagittal view of the head and neck
Anatomy	Full color images of the hypopharynx and larynx	Gray-scale images obtained including structures in the oral cavity, pharynx, and cervical esophagus
Contrast	Green/blue/white food dye *May also use barium*	Barium
Swallow	Able to view pharyngeal phase except for brief (less than 1 s) "white out" during height of swallow. Unable to view oral phase or esophageal phase	Comprehensive view of all swallowing phases – oral, pharyngeal, cervical esophageal phases

Table 18.2 Patient selection for FEES

Best candidates	Less ideal candidates	Not candidates
Breastfed infants Unable to transport from bedside Cranial nerve involvement Pre-and-post upper airway surgery Require assessment of laryngeal/pharyngeal sensation and secretion management Concerns for frequent radiation exposure Require specialized positioning that is not possible in fluoroscopy Limited volume of oral intake (assess readiness for initiation or progression of oral feeding) Require additional time for comprehensive exam that is not possible in fluoroscopy	Toddlers Behavior challenges Oral sensory deficits/oral aversion Infants with laryngomalacia Infants with good coordination of suck-swallow-breathe sequence (frequent "white out") Retrognathia	Nasal obstruction Choanal atresia Pharyngeal stenosis Bleeding disorders Severe sensory or behavioral disorders

Safety, Use of Topical Nasal Anesthetic, and Contrast Material

FEES has been established as a safe exam across the age span in multiple studies [1, 17], including in preterm infants in the NICU [13] and breastfeeding infants [11]. The most commonly reported adverse effects of FEES reported in the literature are epistaxis, vasovagal response, and laryngospasm; however, these are rarely seen. The acuity of patient population must be considered when determining the safety measures that should be in place for FEES. Additionally, careful consideration should be given to whether a FEES should be completed in patients who are acutely ill or medically fragile [18].

The use of topical anesthetic in FEES has been studied in adults, but not specifically in children. In the pediatric population, use of topical anesthetic should be avoided for children with severe neurologic compromise and infants younger than 12 months due to concerns with alteration of laryngeal sensation and potential impact on swallowing function [19, 20]. At this time, there are no published data to guide dosage, concentration, or delivery method of topical anesthetic for children during FEES. When possible, patient comfort during the FEES should be optimized using alternative strategies, including close parent/caregiver participation and support.

Contrast materials that can be considered for use during FEES include green, blue, or white food coloring or liquid barium. There are varying reports in the literature about the utility of contrast material to improve visualization of the pathway of the bolus [21–23]. In breastfeeding infants, contrast material may be swabbed into the infant's oral cavity [11] and/or on the mother's nipple prior to the initiation of breastfeeding; however, contrast may only be visible for the initial few boluses.

Roles of Team Members

The members of a pediatric FEES team and their roles may vary from facility to facility, and the team members present at each evaluation depend on the needs of the patient. A collaborative, integrated, and *interdisciplinary* team is the desired model of care in pediatric FEES. The minimal team present for a pediatric FEES would include an endoscopist and a skilled feeder. The endoscopist may be the otolaryngologist, speech pathologist (pending scope of practice in SLP's region or facility), or in some facilities, the pulmonologist. Advanced practice providers (e.g., physician assistant or nurse practitioner) may also pass the endoscope. The endoscopist adjusts the position of the endoscope throughout the exam in response to vocal and swallowing tasks. Analysis and interpretation of the pharyngeal swallow is typically the role of the speech-language pathologist and may be performed in conjunction with otolaryngologist. Ideally, the team member serving as the skilled feeder is not responsible for interpretation of pharyngeal swallow function. The role of the skilled feeder is to assess the oral phase of swallow, responsively feed the patient, and make adaptations based on exam findings assessed by the speech-language pathologist (i.e., changing bolus size, consistency, pacing, or modality). At our facility, occupational therapists serve as the skilled feeder during FEES. Other team members who may support a pediatric FEES team include a registered dietitian, child life specialist, and lactation consultant (Table 18.3).

Speech-Language Pathologist Approach

The Set Up: Preparing for Pediatric FEES

FEES is a *functional* exam and is most effective when swallowing is visualized in the closest approximation to the patient's typical feeding, including preferred foods and liquids, modalities for eating and drinking, and rate of consumption. Planning and preparation need to occur among team members in collaboration with the patient and caregivers. Organization and environmental supports lay the foundation for a successful clinic visit.

Parent/Patient Preparation

Understanding the purpose of FEES and what will happen during and after the appointment is

Table 18.3 Roles of team members in FEES

FEES team member	Role
Speech-language pathologist	Screening of communication/cognitive status; oral peripheral exam; evaluation of pharyngeal and laryngeal vocal and swallowing function; determination of appropriate adaptations to improve the safety and efficiency of feeding and swallowing function; in some settings, SLP passes and positions the endoscope
Otolaryngologist (ENT)	Evaluation of ENT health/status; passing and positioning of endoscope; evaluation of pharyngeal and laryngeal structure and function; participation in determining the post-exam feeding plan and making appropriate referrals
Occupational therapist (OT)	Evaluation of motor development, sensory processing, oral motor skills, oral sensory processing, and positioning needs relevant to feeding/swallowing function; positions parent and parent/child; assists with feeding patient during the assessment; determination of appropriate adaptations to improve the safety and efficiency of feeding and swallowing function
Nurse	Collection of vitals and medical intake, assists in positioning/supporting patient during the evaluation, including stabilization of the patient's head to prevent excessive movement
Registered dietitian	Evaluation of patient's nutrition status; provision of diet recommendations that optimize nutrition based on swallow study results
Child life specialist	Preparation of patient and family for evaluation; may share social stories or demonstrate the exam on a doll; supports patient and family during the evaluation and may suggest strategies for calming and/or alternate focus (stress balls, watching a video, etc.)
Lactation consultant	Supports the mother-baby pair in the process of breastfeeding; prevents, recognizes, and solves breastfeeding difficulties
Parents/caregivers	Educates staff on their child's current diet and feeding/swallowing challenges; provides insight into home strategies used to decrease child's stress; verbalizes family's goals and priorities and child's medical history
Child	Eat, swallow, vocalize, and participate in the most typical feeding possible

important for parent/caregiver and the child (if developmentally appropriate) prior to FEES. Caregivers are encouraged to bring the child's familiar feeding items (cups, utensils, etc.), items that are calming and soothing (e.g., pacifier, music), and preferred foods and liquids. Team members discuss with parents/caregivers whether they feel the child will be able to tolerate the exam and how the team can support the child's successful participation.

For older children, a practice FEES therapy session with the clinician and/or the child life specialist prior to the exam can be useful to provide exposure to the equipment, setting, and personnel for FEES. Providing a social story can prepare a child for what to expect. Practicing some parts of the exam such as drinking liquids colored with blue food dye at home may be helpful. Establishing a behavioral reinforcement system (e.g., star chart for number of swallows) or providing an incentive can improve a child's motivation to participate. If appropriate, directing a child's attention to his/her exam images may benefit the child's level of engagement as well as provide an opportunity for biofeedback.

Setting Up the Environment

To optimize success and efficiency, it is critical that all food/liquid consistencies, cups, and bottle and feeding utensils are prepared, labeled, and easily accessible before the exam begins. Team members who are present during the exam must bring a calm, attentive, and cooperative attitude. The environment should support patient participation with minimal distraction, calming, and soothing items for the patient. Prior to the exam, infants may benefit from having access to sucrose and non-nutritive sucking, which have been shown to have analgesic properties for neonates, particularly when offered together [24].

Positioning

Supportive positioning with proper pelvic alignment and support of the trunk, neck, and head is critical for optimizing oral motor skills and swallowing in children, particularly those with neuromuscular disorders [25–27]. For infants, swaddling allows for positioning the baby in a supportive posture with hands at midline (accessible to the infant's mouth) and flexion of the elbows, knees, and hips. Swaddling may improve

motor organization, decrease physiologic distress, and increase self-regulation ability in preterm infants [28]. Specialized breastfeeding positions may also be beneficial to support feeding and swallowing [29] (Fig. 18.1). Side-lying is a frequently suggested position postulated to support physiologic stability in both breast and bottle-fed infants, though research findings have been conflicting as to its effectiveness in a limited number of studies [30].

The Examination

Goals

It is critical to establish goals for what the team considers an "adequate" FEES as well as individualized FEES goals for each patient. The child's age, level of development, compliance, and past feeding experience all influence what is able to be observed. A prescribed protocol may or may not be possible. Modifications of vocal and swallowing tasks are often required to optimize efficiency of exam and patient compliance.

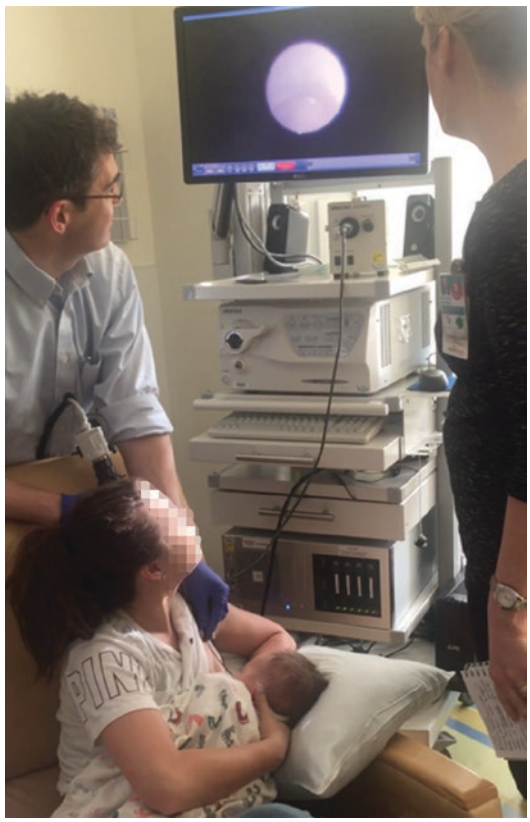

Fig. 18.1 Setup for breastfeeding FEES

Oral Mechanism Exam

Prior to introducing foods or liquids during the endoscopic exam, assessment of facial and oral motor structures and function is critical as part of a comprehensive feeding and swallowing examination [31]. In this assessment, the clinician may evaluate resting posture and tone of oral and facial structures, dental occlusion and dental eruption, and range of motion and coordination of facial and oral motor structures as part of the cranial nerve exam. Non-nutritive oral skills as well as observations of patient's vocal quality, pitch, and loudness are obtained. These observations will inform decision-making regarding which foods or liquids are offered and what modality to use (i.e. open cup, straw) during FEES.

Global and Oral Sensory Processing

In preparation for FEES, the child's global and oral sensory processing function is considered. Exam activities are tailored to achieve a state of arousal supportive of feeding and participation during the exam [32]. Clinicians consider oral sensory processing challenges including hyposensitivity, hypersensitivity, or both when providing modifications during bolus presentation for FEES. Environmental modifications that can be made include reduced lighting, minimizing external distractions, or swaddling to help calm and organize the sensory system and maximize patient participation in the FEES [32].

Oral Motor Observations of Feeding

Oral motor skills may be determined to be age appropriate, impaired, delayed, and/or dysfunctional. Patterns of oral bolus residue, oral bolus

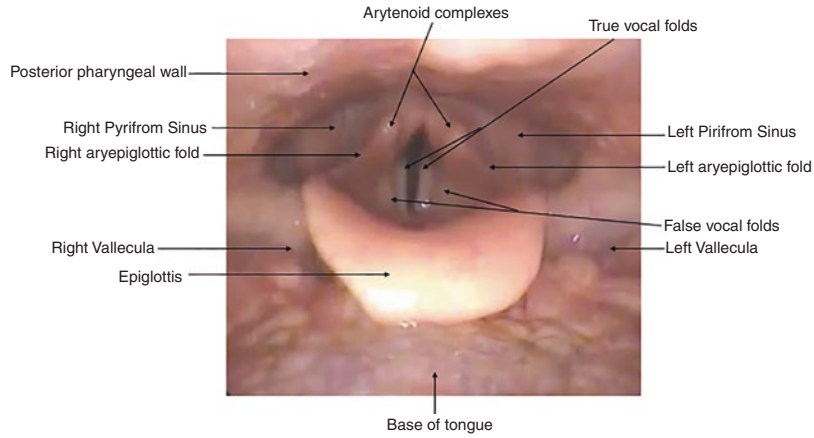

Fig. 18.2 "Home position" with anatomic structures labeled

containment, sensory awareness of the bolus in the oral cavity, and oral manipulation of the bolus directly impact decision-making regarding what adaptations are needed (Fig. 18.2). Online communication between the skilled feeder and the individual viewing the images is critical.

Assessment of Pharyngeal and Laryngeal Structure and Function

Nasal Structures and Nasopharynx

A patent nasal passage is required for FEES. Selection of passing the endoscope through the right or left nare is directly impacted by the presence or absence of nasal obstruction. When a nasogastric tube is in place, the team needs to decide to remove the tube, pass the endoscope around the tube, or pass the endoscope in the contralateral nare. The speech-language pathologist may provide assessment of the velopharyngeal mechanism during speech tasks if there are concerns with resonance and/or nasopharyngeal regurgitation.

Secretion Management

Prior to introducing a bolus, the team evaluates standing secretions in the hypopharynx, looking for the presence/absence, volume, location, color, and viscosity of secretions. The team can assess volitional or spontaneous ability to clear standing secretions. These observations contribute to understanding of the child's global swallow function, determination of risk of aspiration, and inference of pharyngeal swallow function for the patient's current diet [2, 33, 34].

Vocal Tasks

Vocal tasks elicited during FEES will vary pending vocal concerns and patient's developmental level. During vocal tasks, one can infer cranial nerve involvement and appreciate symmetry, strength, range of motion, and coordination of pharyngeal and laryngeal structures.

Pharyngeal and Laryngeal Sensory Testing

Reduced laryngopharyngeal sensory capacity has been shown to be correlated with penetration, aspiration, history of pneumonia, neurological disease, and reflux [35]. Indirect assessment of pharyngeal and laryngeal sensation can be completed given symptoms of deep laryngeal penetration or aspiration without cough, presence of pharyngeal residue that is not perceived or spontaneously cleared by the patient, or minimal response to the presence of the endoscope.

Pharyngeal Swallowing

Direct swallowing tasks are elicited, and the following swallow parameters are assessed (Figs. 18.3, 18.4, 18.5, 18.6, 18.7, and 18.8):

1. *Initiation of pharyngeal swallow*: Anatomical markers are used in determining the location of pharyngeal swallow initiation, including base of

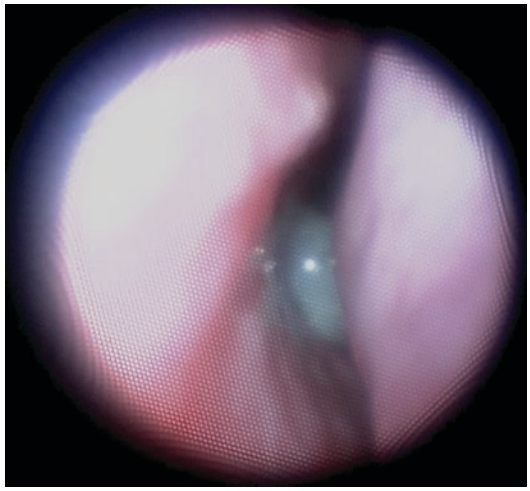

Fig. 18.3 Presence of liquid in nasal passages

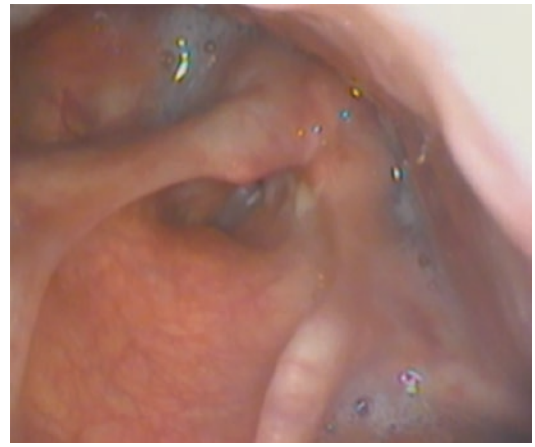

Fig. 18.5 Secretions in valleculae and piriform sinuses

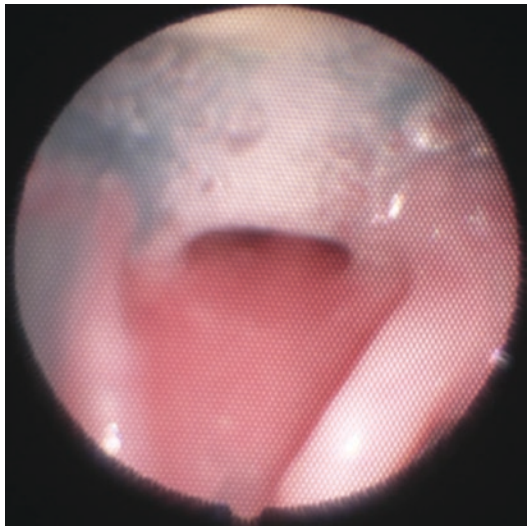

Fig. 18.4 Laryngeal penetration

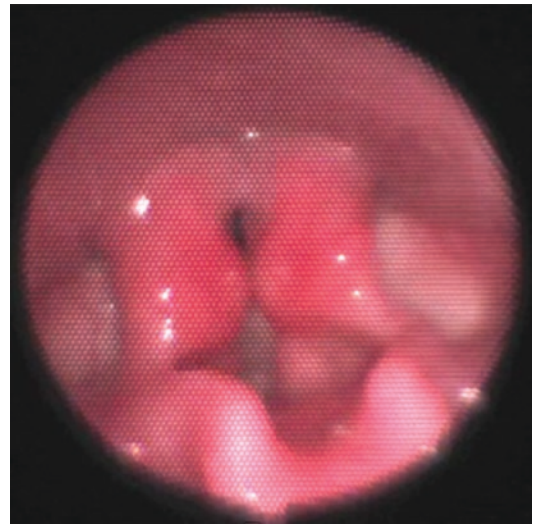

Fig. 18.6 Laryngeal cleft

the tongue, valleculae, and pyriform sinuses (Fig. 18.2). As the bolus enters the pharynx, clinicians visualize where the bolus is held prior to the swallow as well as timeliness of initiation of the swallow. The pattern of bolus flow (lateral channels, right vs left, central) is also directly viewed. Risk to airway protection can be directly assessed by observing the bolus in instances of delay in initiation of the pharyngeal swallow.

2. *Laryngeal penetration*: Presence or absence of laryngeal penetration, when laryngeal penetration occurred (before, during, after swallow), and depth of penetration in the laryngeal vestibule are determined. Challenges in determining the depth of penetration are present due to "white out," especially during FEES with infants with rapid consecutive swallows. Examiners can most reliably assess presence/absence and depth of laryngeal penetration if occurring before or after the swallow.
3. *Aspiration*: Aspiration can be directly viewed. However, "white out" occurs during the height of the swallow when pharyngeal musculature constricts around the endoscope, lasting approx-

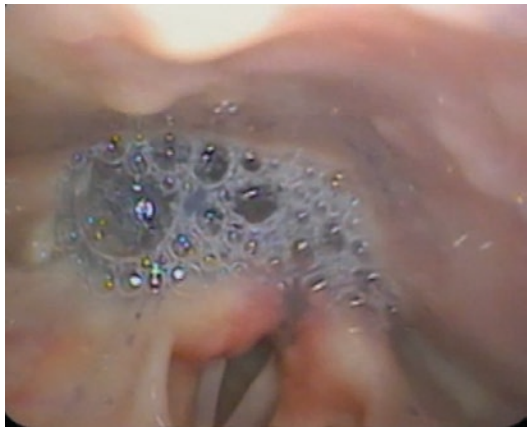

Fig. 18.7 Post swallow residue in piriform sinuses

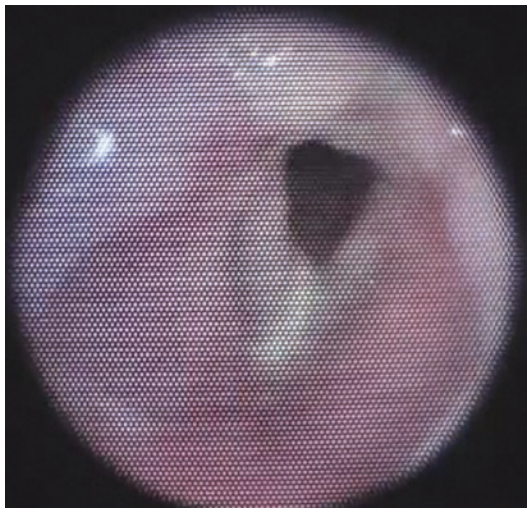

Fig. 18.8 Aspiration

imately less than 1 second. If aspiration occurs before the swallow or after the swallow, images can directly confirm aspiration. Presence or absence of aspiration can also be assessed based on post-swallow residue. Examiners assess the presence/absence, timeliness, and effectiveness of a cough in response to the aspiration event.

4. *Residue*: After the pharyngeal swallow, bolus residue patterns can be assessed. Presence or absence of pharyngeal residue, location of pharyngeal residue using anatomical markers, and the amount of residue are evaluated. In the literature, there are standardized and validated residue severity rating scales designed for use in FEES [36, 37] but are not designed with intended use for pediatric patients.
5. *Upper esophageal sphincter (UES)*: The UES inlet is viewed, but images obtained do not allow complete visualization of the bolus passing through the pharyngeal-esophageal segment. UES function is not able to be directly assessed. If there is concern regarding UES dysfunction, other diagnostic exams (e.g., upper GI, esophagram, VFSS, manometry) may be recommended.
6. *Nasopharyngeal regurgitation*: Instances of nasopharyngeal regurgitation are typically not able to be directly viewed given position of the endoscope during FEES. However, if there are concerns for nasopharyngeal regurgitation, the endoscopist may position the tip of the endoscope in the nasopharynx to determine presence or absence of post-swallow bolus residue.
7. *Compensatory strategies*: Both direct and indirect compensatory strategies may be trialed based on the patient's symptoms. Indirect strategies include modifications to bolus size and bolus flow (change to nipple flow rate, sipper cup with or without valve in place, etc.), changes to the position of the patient, type of positional supports in place, or changes to viscosity of liquids presented [38]. Direct compensatory strategies may be used, but implementation of strategies is impacted by the age and/or developmental level of the pediatric patient. Trial of compensatory swallow strategies or swallow maneuvers is an ideal time to utilize FEES as a biofeedback tool, both to build patient awareness of dysphagia symptoms and also to visualize success of strategies trialed (Tables 18.4 and 18.5).

After the Examination

Creating a Feeding Plan

Given information from FEES regarding swallowing physiology, FEES team members including the family and patient work together to

Table 18.4 FEES worksheet

Nasopharyngeal/laryngeal assessment	
Nasal anatomy	R/L Patent Occluded Other
Velopharyngeal closure (puppy/baby/kitty/sssss)	Complete Incomplete Asymmetric R/L
Secretions	Absent Mild Moderate Severe Cleared Cleared to ____ Did not clear *Color*: Clear Cloudy *Viscosity*: Thin Thick
Tongue base retraction ("Paul is tall"; "Ball-ball-ball")	WNL Reduced Asymmetric R/L
Pharyngeal wall movement (Effortful pitch glide "ee", high pitch "ee-ee-ee")	WNL (pyriforms obscured) Reduced Weak R L
Vocal fold mobility ("ee" – sniff, "ee-ee-ee")	Complete AB/ADuction R/L Partial on R/L Immobile on R/L *Resting position*: lateral paramedian median
Volitional glottic closure (breath hold, Valsalva/bear down, cough, throat clear)	Complete Partial Incomplete Position of VF/supraglottic structures: _____
Laryngeal adductor reflex (touch to L/R arytenoid)	Present Absent R/L
Lesions (tongue side to side, cheek puff)	Present Absent Location: _____ Size: _____ Color: _____ Shape: _____

Pharyngeal swallow assessment	
Swallow initiation	Base of tongue Valleculae Pyriform sinuses not initiated No pooling Prolonged pooling
Laryngeal penetration	Present Absent Mild Moderate Deep Unable to judge depth Before swallow After swallow
Aspiration	Yes No Silent Cough Throat clear Before swallow During swallow After swallow
Residue	Yes No Trace Mild Moderate Severe R/L BOT Valleculae Pyriforms Pharyngeal Wall Laryngeal Vestibule UES Cleared Cleared to _____ amount Did not clear
Upper esophageal sphincter	Closed at rest Open during _____
Nasopharyngeal regurgitation (view during scope exit)	Present Absent

Compensatory strategies	
Strategies	Postural changes Change of bolus size Dry swallow Head turn R/L Head tilt R/L Effortful swallow Chin tuck Supraglottic swallow Super-supraglottic swallow Mendelsohn Change in Modality Other
Result of strategies	Improvement in: _____ No change Worsening

Oral structure and function	
Structures	WNL Atypical: _____
Dentition	Age typical Tooth eruption: _____
Oral/facial tone	WNL Low High
Resting posture	WNL (closed) Open Open with tongue resting forward
Secretion management	WNL Anterior loss: Yes No Mild Significant
Range of motion	WNL Within functional limits Atypical

(continued)

Table 18.4 (continued)

Oral preparatory phase	
Lip seal	WNL Weakness
Jaw excursion	WNL Appropriate for bolus presentation Inadequate Excessive Clonus Weakness
Lingual control	WNL Decreased lateralization Decreased Protrusion Uncoordinated Fasciculations Weakness
Bolus containment and manipulation	WNL Intra-oral scatter Anterior loss Decreased manipulation Bolus hold Prolonged oral prep
Suck/chewing pattern	WNL Immature Disorganized Dysfunctional Dystonic Compressive Nonnutritive Vertical Phasic Munching Rotary Circular Tonic bite Bruxism
Sensory	WNL Hypersensitive Hyposensitive
Feeding skills	WNL Within functional limits Delayed Disorganized Impaired
Modifications	Not needed Needed: Mild Moderate Significant

Table 18.5 Problem-solving in pediatric FEES

Presenting problem	Strategy
Presence of pooled secretions before introduction of bolus	Introduce pacifier or empty spoon to elicit saliva swallow
Endoscopic view occluded by thick secretions or bolus residue	Elicit subsequent swallow or liquid wash
	Direct contact/swipe of endoscope to posterior pharyngeal wall
	Introduce bolus with powdered barium as contrast agent at *end of exam* (in case of barium coating endoscope and occluding view)
Suspicion of trace aspiration	Between swallows, descend tip of endoscope toward laryngeal vestibule for magnified view
	If possible, elicit sustained /ee/ during this task
Prolapse of base of the tongue occluding view of larynx	Elicit mandibular extension (jaw thrust)
	Ensure the head/neck in neutral alignment
Unstable state regulation in infant	Low lights, quiet environment, swaddle, pacifier
Concern for decreased pharyngeal and laryngeal sensation	When residue is present, ask patient if he/she feels any food/liquid in throat
	After completion of vocal and swallowing tasks, endoscopist completes laryngeal adductor reflex test

identify a feeding plan that allows for the least restrictive diet and a safe, pleasurable feeding plan while minimizing aspiration risk and maintaining the patient's overall health. Rarely is this conversation held in isolation of the family and FEES team. In the case of a patient followed by a larger multidisciplinary team (e.g., aerodigestive clinic), decision-making expands to coordination of the feeding plan with a larger group of experts including the otolaryngologist, pulmonologist, gastroenterologist, dietician, nurse, and social worker [39] in addition to a patient's pediatrician.

Practical Considerations: FEES "Failures"

Despite excellent patient triage, preparation, and supports offered during FEES, the exam may not yield the desired diagnostic information. Reasons for unsuccessful exams are varied and can include patient refusal, inability to calm with the endoscope in place, obscured view of the swallowing structures, and/or excessive movement. Unsuccessful exams are opportunities for FEES team to learn how to optimize the exam for improved results in the future. Ultimately, it is not always possible to predict when an exam will be unsuccessful. Discussion with the caregiver regarding the possibility that the child may or may not cooperate during FEES is helpful to manage expectations.

Otolaryngologist Approach

Observations of Laryngeal and Pharyngeal Structures

A flexible endoscope is passed through the nasal passage and into the nasopharynx. Note is made of the patency of the nasal passage and the adenoid size, at the least. Once in the nasopharynx, the endoscope is turned inferiorly to observe the oropharynx, hypopharynx, and larynx from a superior perspective. Note is made of palatine and lingual tonsillar appearance and size. The structures of the supraglottis, including the epiglottis and arytenoids, are brought into view. Mobility of the arytenoids and true vocal folds is assessed, as is the quality of phonation and the presence or absence of stridor. Common pathologic observations include the presence of laryngomalacia and/or vocal cord immobility. In addition, the presence of secretions and how they are managed by the patient may be observed as well. Poor management of secretions and a reduction in cough from secretions and the endoscope may imply altered sensation via congenital or acquired conditions (e.g., Down syndrome and stroke, respectively).

Observations of Swallowing

Once the endoscope is positioned posterior to the tip of the uvula, the hypopharynx, larynx, and esophageal inlet are in view. At this point, dry swallowing and voicing tasks may be carried out if desired. Swallowing tasks using various consistencies and amounts are then performed with the assistance of therapy personnel. Observations include the pattern that the food bolus takes to the pyriform sinus (one versus two sides of the hypopharynx), the presence and location of residue, pooling of material in the hypopharynx, penetration, and aspiration. In addition, the elevation of the larynx during the swallow as well as the strength of the swallow to "strip" the food bolus from the walls of the aerodigestive tract and endoscope can be assessed. An inability of swallowing to clean the tip of the endoscope may imply a weak swallow.

Recommended Interventions

For the otolaryngologist, certain findings discovered during FEES may prompt additional testing and intervention. For patients who continue to aspirate over time, central imaging may be recommended in order to rule out a finding that may explain the etiology of dysphagia (e.g., absence of the corpus callosum, Chiari malformation). These findings may occur even in syndromic populations for whom an explanation for dysphagia may already exist.

FEES in Surgical Patients

Given that FEES exams are customizable to the particular patient, this is a most useful tool for swallow evaluation after surgical intervention. While certain ages and neurological status may make FEES challenging, outside of those relative contraindications, FEES is a suitable study for most patients after airway surgery, with some exceptions. If the patient had particular findings on VFSS preoperatively, performing the same study postoperatively may be most appropriate. In addition, if a laryngeal cleft repair were performed, FEES does not assess the esophageal phase of swallowing and may miss a distal repair failure (Fig. 18.6).

Emerging and Evolving Techniques of the Future

Laryngopharyngeal Endoscopic Esthesiometer and Rangefinder (LPEER)

Disruptions to laryngopharyngeal mechanosensitivity are a known, but challenging to measure, contribution to pharyngeal swallowing dysfunction, including in the pediatric population [40]. Previously used technologies to measure the laryngeal sensory function were limited by the challenges with consistent stimulus reliability and the ability to measure the laryngeal adductor

reflex threshold (LART) only. The laryngopharyngeal endoscopic esthesiometer and range-finder has been developed with functionality to better standardize the distance and location at which high-precision air pulses of various intensities can be delivered to determine the LART as well as the cough reflex threshold (CRT) and the gag reflex threshold (GRT) [41]. This technology holds promise for improved understanding of the role of sensory responsiveness (both hypo- and hypersensitivity) not only in pharyngeal dysphagia due to neurogenic and other causes but also potentially in obstructive sleep apnea, chronic cough, and vocal fold dysfunction. Further, assessment of pharyngeal sensory sensitivity in populations who are known to be at risk for dysphagia, such as children with Down syndrome, may improve our understanding of the underlying mechanism of their dysphagia and potential treatments.

Imaging Innovations: Narrow-Band Imaging and High Frame Rate

Narrow-band imaging (NBI) is a filtering technology designed to enhance visualization of the contrast between surface blood vessels and surrounding mucosa during endoscopy. NBI has been described as being useful in FEES as improving visualization of contrast between the bolus and surrounding mucosa and in improving detection of depth of laryngeal penetration, visualization of smaller volumes of aspirated material to the subglottic space, and inter and intra-rater reliability of detection of pathologic findings [42, 43]. While NBI has been shown to improve bolus visualization, use of high frame rate (HFR) videos using high-speed digital imaging (HSDI) has been shown to improve visualization of the rapid motion of pharyngeal structures (i.e., epiglottic tilting, posterior pharyngeal wall movement) prior to the "white out" seen while the bolus passes through the pharynx. As compared to the standard frame rate of 30 frames per second (fps), high-speed recording (4000 fps) improves motion detection [44].

Case Reviews

Case Study 1

Background

Becky is a 14-year-old girl who suffered a traumatic brain injury and spinal cord injury during a motor vehicle accident. Becky was admitted emergently to the hospital, underwent craniotomy, halo placement to stabilize the cervical spine, and placement of a nasogastric (NG) tube. During therapeutic trials of liquids and solids, Becky demonstrated poor oral bolus manipulation, post-swallow oral residue, and wet vocal quality during and after oral tastes/trials. Becky demonstrated right labial and tongue weakness, mildly hypernasal resonance, and breathy vocal quality with decreased vocal intensity. Behaviorally, Becky has a flat affect, short attention span, and working memory deficits. FEES was selected for initial instrumental swallow evaluation due to presence of the halo, ability to assess vocal and swallowing tasks, as well as potential to trial compensatory swallow strategies and/or swallow maneuvers as needed.

Pre-FEES Planning

What is your plan for vocal tasks and swallowing tasks? How will you support positioning changes while the scope is in place, given the presence of a halo? What supports may be used to improve participation in FEES?

Exam

The FEES team explained the exam to Becky and her parents. Becky was supported by a speech-language pathologist with a visual schedule of vocal and swallowing tasks. The occupational therapist provided assistance at the bedside for comfortable and stable upright position with ability for Becky to view images on the monitor. The team then proceeded with placement of the endoscope and initiation of vocal and swallowing tasks.

Online Problem-Solving (Vocal Tasks)

During phonation, the right velum elevated partially resulting in velopharyngeal insufficiency,

the right vocal fold was immobile in the paramedian position, and right pharyngeal wall constriction was poor. Pooling of secretions was noted in the right vallecula and right pyriform sinus (Fig. 18.5). *What do you need to do before introducing food and liquid?*

Online Problem-Solving (Liquid Swallowing Tasks)

Becky was able to clear the secretions with an effortful swallow prior to the introduction of liquids. During the first swallows of thin liquid bolus (consecutive swallows via open cup), you observe deep laryngeal penetration with entrance to the laryngeal vestibule over the right aryepiglottic fold. *What task do you present next?* The team elected to try pacing and bolus size modification in an effort to improve Becky's airway protection during the swallow. Becky was next presented with single swallows of thin liquids via a small diameter straw. In order to determine if she could protect her airway with consecutive swallows of thickened liquids, International dysphagia diet standardisation initiative (IDDSI) [38] level 2 - mildly thick liquids were presented with a regular straw.

Online Problem-Solving (Solid Swallowing Tasks)

After swallowing a single bolus of puree, you notice diffuse pharyngeal residue (right side greater than left side). *How can you determine if Becky is aware of the residue? What compensatory strategies could you trial to clear the bolus residue?* When asked if she had swallowed all of the puree, Becky reported that she had, suggesting decreased sensory awareness of pharyngeal residue. A dry spoon was presented to stimulate a clearing swallow.

Results

Becky demonstrated adequate airway protection with single swallows of IDDSI level 0 - thin liquid via small diameter straw, but consecutive swallows of IDDSI level 0 - thin liquid resulted in aspiration. Becky demonstrated adequate airway protection during consecutive swallows of IDDSI level 2 - mildly thick liquids via regular straw. With purees, Becky required three subsequent swallows to clear pharyngeal residue. When a liquid wash was trialed, Becky presented with an episode of silent (no cough) aspiration of liquid and IDDSI level 4 - extremely thick (puree). IDDSI level 7 - regular solids were not able to be fully cleared, but Becky did not demonstrate airway compromise from regular solid food residue. Of note, her halo is scheduled to be removed in 4 weeks.

Feeding and Swallowing Plan

What is your recommended feeding plan? What tasks would you address in speech and dysphagia therapy? Would you recommend any ENT interventions to address either vocal or swallowing concerns? When would you complete another instrumental swallow examination, and what type of examination do you recommend?

Considerations

The FEES identified Becky's pharyngeal swallow pattern, as well as identified glottic insufficiency and decreased pharyngeal sensation. An interdisciplinary discussion occurred to determine next steps, which included allowing single sips of IDDSI level 0 - thin liquids and small bites of IDDSI level 4 - extremely thick (purees) with "dry" swallows between bites. Medical fragility of the patient influenced oral diet plan. Given unilateral vocal fold immobility, the otolaryngologist offered vocal fold medialization procedure via injection laryngoplasty for better voice outcome and potentially better airway protection during swallowing. A repeat FEES was recommended following removal of halo and injection laryngoplasty.

Case Study 2

Background

Daniel is an 8-year-old with a medical history significant for trisomy 21, history of oxygen requirement at night, recurrent pneumonia, and recent hospitalization with bronchiolitis. Daniel has been consuming a full oral diet including thin liquids and regular solids, and his parents report daily choking with both liquids and solids. His family is concerned about radiation exposure and is requesting a FEES rather than a VFSS.

Pre-FEES Planning

A practice session was scheduled prior to the FEES appointment. During the practice session, he responded well to behavioral reinforcement with a star chart and incentive of a preferred toy as a prize. A social story was made outlining what would happen during FEES appointment.

Exam

Oral mechanism exam was significant for open mouth posture, narrow, high-arched palate, and anterior open bite with occlusal contact present only at the molar surfaces. During observation of feeding, Daniel was noted to have significant oral motor difficulties for solids. The FEES team discussed prioritizing evaluation of liquids during the endoscopic exam, but solid foods were prepared for trial if Daniel is tolerant of FEES.

Online Problem-Solving (Obstructed View)

Upon passage of the endoscope, large tonsils and adenoids were seen, and thick secretions obscured the endoscope. Vallecular space was limited due to large lingual tonsils. Tongue base prolapse was noted, which limited visualization of the laryngeal vestibule. *How would you clear secretions from the tip of the endoscope? What positional changes would you trial to maximize your pharyngeal and laryngeal view during FEES?*

Online Problem-Solving (Participation)

A small bolus of water was provided to clear the endoscope. Daniel's head/jaw was extended slightly forward to improve view of the laryngeal vestibule. Daniel tolerated placement of the endoscope, but began complaining and requesting to take the scope out. *Would you proceed with vocal tasks, swallowing tasks, or both?*

Online Problem-Solving (Vocal and Swallowing Tasks)

Daniel's vocal fold mobility appeared intact bilaterally during spontaneous phonation. Team elected to forgo formal vocal tasks and move directly to observations of swallowing. Multiple episodes of silent aspiration were seen during consecutive swallows of IDDSI level 0 - thin liquid with post-swallow residue in laryngeal vestibule. *What swallowing tasks would you trial next? Would you attempt to clear post-swallow residue or offer additional liquid trials?*

Daniel was able to follow directions to cough to clear post-swallow residue from laryngeal vestibule. He was able to protect his airway with single swallows of IDDSI level 0 - thin liquid. *What swallow task would you trial next? Would you trial a different modality for drinking or change viscosity of liquid?*

Results

Daniel presented with silent aspiration during consecutive swallows of IDDSI level 2 - mildly thick liquids. Daniel demonstrated adequate airway protection during rapid consecutive swallows of IDDSI level 3 - moderately thick liquids. With all liquid consistencies presented, swallow initiation fell at the level of the pyriform sinuses without prolonged pooling before the swallow. IDDSI level 4 - extremely thick (purees) and IDDSI level 6 - soft and bite-sized consistency were swallowed with adequate airway protection and no pharyngeal residue.

Feeding and Swallowing Plan

What is your recommended feeding and swallowing plan given oral motor deficits and pharyngeal dysphagia? How could you support a functional and safe feeding plan for Daniel while maintaining quality of life? Are there other medical examinations or procedures to recommend given findings of oropharyngeal dysphagia, persistent pulmonary symptoms and concerns for upper airway obstruction?

Considerations

Based on the results of his FEES, the swallow team recommended modifying Daniel's diet to thicken liquids to IDDSI level 3 - moderately thick and to provide solid food textures that are a good match for his current oral motor skills (purees, meltable solids, and soft solids), with regular solid foods presented in a finely chopped form only (IDDSI level 5 - minced and moist). Feeding therapy was recommended to assist Daniel and his family with transitioning from IDDSI level 0 - thin liquids to IDDSI level 3 - moderately thick liquids and work toward

improved oral motor skills for consumption of textured solids. Further evaluation with a multidisciplinary aerodigestive team evaluation was recommended considering Daniel's pulmonary health and suspected long-term untreated dysphagia, with plans to evaluate for any structural problems that could be contributing to his dysphagia. The aerodigestive team elected to pursue diagnostic endoscopy. The otolaryngologist elected to consider tonsillectomy and adenoidectomy during this procedure pending results of a sleep study. The family's goal of allowing Daniel to continue taking small volumes of regular water for quality of life will be discussed pending results of the aerodigestive team procedures and assessment.

Case Study 3

Background

Max is a 4-month-old boy with concerns of choking when breastfeeding and bottle feeding. Max was hospitalized for upper respiratory infection and hypoxia and was discharged home with an oxygen requirement. While breastfeeding and bottle feeding, Max demonstrated inspiratory stridor and increased work of breathing, resulting in feeding lasting up to 45 min and mother feeding Max "constantly." At 40 days old, Max underwent a videofluoroscopic swallow study (VFSS). Results revealed incoordination of breathing and swallowing, resulting in mistiming of airway closure and deep laryngeal penetration with thin liquids by a slow flow nipple.

Pre-FEES Planning

What factors do you consider when selecting FEES versus VFSS? What are the goals and priorities for completion of FEES? How will you support Max and his mother during the exam to ensure that he is calm and able to participate in feeding?

Online Problem-Solving (Laryngeal Structures and Participation)

FEES was selected to assess swallow function during breastfeeding and upper airway structures given report of stridor and increased work of breathing. Consideration was also given to reduce radiation exposure. Pre-FEES family counseling was completed to prepare Max's mother for breastfeeding during endoscopy.

Max was swaddled in a flexed position prior to the start of the exam. Upon passing and positioning the nasal endoscope, Max cried continuously for 45 seconds. The otolaryngologist noted severe laryngomalacia demonstrated by significant prolapse of the arytenoids into the laryngeal vestibule and omega-shaped epiglottis. *When do you initiate breastfeeding during the exam? How do you support Max and his mother in achieving a calm state for feeding?*

Online Problem-Solving (Liquid Swallowing Tasks)

Max was calmed using a pacifier dipped in sucrose and was able to latch to the breast. When transferring breastmilk, initiation of pharyngeal swallow occurred consistently at the pyriform sinuses with mistiming of breathing coinciding with instances of intermittent laryngeal penetration. Visualization of the laryngeal vestibule and the vocal folds was challenging due to laryngomalacia.

Results

Between swallows, contrast was noted on the anterior commissure of vocal folds, and trace contrast was present on the ventricular folds. After about 4 minutes of breastfeeding, Max showed signs of fatigue, and the exam was discontinued.

Feeding/Swallowing Plan

What feeding/swallowing plan will you recommend? When should Max repeat an instrumental swallow study? What additional medical interventions are indicated given findings of laryngomalacia, work of breathing during feeding, and pharyngeal dysphagia?

Considerations

Given findings of inadequate airway protection with IDDSI level 0 - thin liquids during FEES and VFSS and patient's history of upper respiratory infections and hypoxemia, the medical team elected to pass a nasogastric (NG) tube. The team

recommended that Max receive the majority of his nutrition via pumped breastmilk delivered by NG tube. In order to preserve Max's skills for breast and bottle feeding, the team elected to allow small volumes of pumped breastmilk via slow flow nipple and/or allowing Max to breastfeed for 5 minutes at the beginning of his NG tube feedings after his mother had emptied her breast via pumping. The otolaryngologist elected to pursue supraglottoplasty to address laryngomalacia. Repeat swallow assessment was recommended 6 weeks following supraglottoplasty. Max's parents requested evaluation via FEES given their goal of resuming breastfeeding.

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Manometric Evaluation of Pediatric Swallow

19

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Introduction

In order to safely swallow, coordinated muscle contraction is needed from 31 pairs of striated muscles and smooth esophageal musculature [1]. These contractions serve to propel the bolus through pressure gradients as well as valve off the nasal cavity and the trachea. Traditional methods of measuring oropharyngeal swallowing function rely on video images through endoscopy or videofluoroscopy (X-ray). These images can provide detailed information about the biomechanics of oral and pharyngeal structures as well as bolus movement and retention. However, these methods require extensive training, and the analysis is largely subjective [2]. Manometry, on the other hand, objectively measures the pressures generated from muscular force and from overall change in shape of the pharynx and esophagus. Pressure sensors embedded in flexible tubing are positioned in the pharynx and esophagus and measure pressure changes over time. The data output is quantitative (numerical), allowing for sensitive calculations to be performed that infer muscle strength, timing, and coordination [3]. While relatively new to the pharynx, manometry has the potential to improve dysphagia diagnosis, management, and outcome measurement [4]. This chapter provides an overview of equipment, data collection, and data analysis of pharyngeal high-resolution manometry with special considerations for the pediatric population.

High-resolution Manometry

Manometry has been used for decades to assess physiologic processes via pressure measurement in the esophagus, stomach, and lower gastrointestinal tract [5]. Since the early 2000s, technology has advanced such that more sensors can be placed in a smaller physical area and computer hardware and software are capable of processing dramatically larger volumes of information. Water-perfused sensors, which required a steady stream of distilled water, have mostly been replaced by solid-state sensors, allowing for a more comfortable placement in the pharynx. Manometric sensors can measure pressure from a single direction (*unidirectional*), can average pressures from multiple axial directions (*circumferential*), or can separately delineate pressures from different axial directions (*3-dimensional*). These technological improvements have allowed

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for a large increase in both spatial and temporal resolution of manometric data, and thus the use of this *high-resolution* manometry has outstripped that of *conventional* manometry [3].

High-resolution manometry has allowed for drastic advancements in understanding of adult swallowing physiology [6–12]. Additionally, all of the advancements that made high-resolution manometry possible improve the feasibility of pharyngeal manometry in children. More pressure sensors that are closely spaced allow for spatial differentiation in the smaller pediatric pharynx. Furthermore, the greater number of sensors allows for an easier and more comfortable catheter placement without the need for the *pull-through technique*, which requires placing the catheter deeper than necessary and then pulling the catheter out until the pressure sensors are situated in the correct location.

Equipment and Procedure

Specifications differ between manufacturers, but all pharyngoesophageal high-resolution manometry setups include (1) a flexible pressure sensing catheter, (2) a data processor, (3) a computer, and (4) a monitor. Pressure catheters typically used in the pediatric population range from 2 to 3.8 mm in diameter and have 20–26 sensors with sensor spacings between 0.5 and 2 cm. Presently, there are no commercially available catheters designed exclusively for the pharynx; all pharyngeal high-resolution manometry is thus performed using esophageal catheters. Depending on child size and equipment specifications, five to ten pressure sensors fall between the velopharynx and upper esophageal sphincter (UES). The system can collect pressures between −20 and 600 mmHg at a sampling rate of 20–50 Hz. Pressures are displayed in real time on the monitor in a spatiotemporal plot (Fig. 19.1).

Pressure measurement can also be combined with impedance measurement on some specialized catheters (Fig. 19.1). In this context, *impedance* refers to resistance to an electrical current. This can be measured by swallowing material that has an ionic charge, such as saline solution. As saline passes between impedance sensors, the electrical charge changes, and thus the data processor reflects that as a change in signal. Therefore, impedance manometry can measure pressures in relation to bolus flow during swallowing without the use of videofluoroscopy [13].

The catheter is placed transnasally into the pharynx and esophagus. Water-based lubricants are often used to assist in passage of the catheter, and a topical anesthetic applied to the anterior nasal passage is sometimes used in older children to reduce discomfort associated with the procedure. Videofluoroscopy or endoscopy can be used to guide catheter placement, but placement is often done blindly, relying on the pressure tracings to identify anatomic landmarks. When accurate catheter placement is confirmed, it can be secured to the nasal tip with adhesive tape, allowing the examiner to move freely. This is an advantage over endoscopy, in which close proximity between the clinician and the child must necessarily be maintained. This ability to move away from the child immediately after catheter insertion can facilitate acclimation to the catheter and subsequently improve the quality of the data obtained. Following placement, a short period of time is allotted for the patient to acclimate to the catheter before swallowing trials are begun. Patients are instructed to fast prior to the examination to reduce the risk of emesis during placement and so the patient is motivated to eat and drink during the procedure.

In both pediatric and adult esophageal high-resolution manometry studies, it is commonplace to follow a standardized protocol [14, 15]. However, no widely accepted protocol exists for pharyngeal high-resolution manometry. Liquids are more frequently used in the literature, with measured boluses ranging from 0.3 to 5 ml for children and bottle-feeding for infants. When possible, bolus trials should be administered methodologically with consistent volumes and consistent methods of delivery. Modifications in position, bolus administration, volume, liquid viscosity, and use of compensatory strategies should be trialed as necessary to answer the clinical question at hand. A benefit of high-resolution manometry over videofluoroscopic or endoscopic

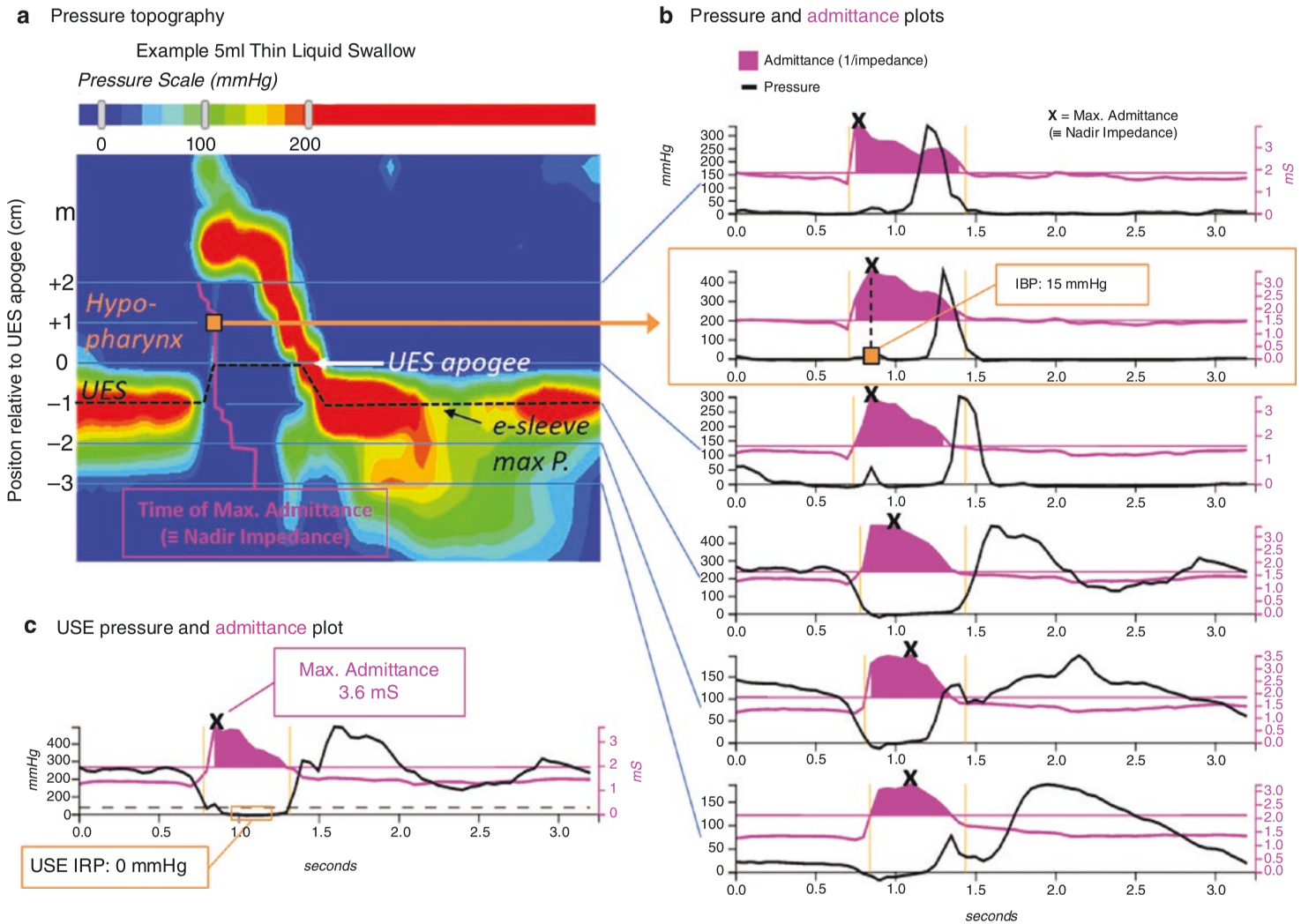

Fig. 19.1 (**a–c**) Pharyngeal high-resolution manometry spatiotemporal output and pressure line plot. (Courtesy of Taher Omari, PhD; Flinders University, Adelaide, South Australia)

swallowing evaluations is that the bolus does not need to be modified (i.e., with barium or food coloring), allowing for a wide variety of foods and liquids that can be assessed during the evaluation. Additionally, once the catheter is in place, the patient may be positioned without much restriction.

There can be multiple indications for performing a pharyngeal high-resolution manometry study in a pediatric patient. The primary indication is when a clinical question cannot be answered through other swallowing evaluations. Specifically, those who aspirate, have unexplained pharyngeal residue, have suck-swallow-breathing discoordination, and have difficulty with solids or those with nasal regurgitation can particularly benefit from this procedure [4]. Pharyngeal swallowing pressures can also be assessed in the setting of esophageal dysphagia [5]. Patients with oral-only dysphagia are not likely to benefit from a transnasal high-resolution manometry study. It is common for the patient to undergo other evaluations of swallowing prior to receiving a manometric study, such as a clinical exam or an endoscopic or videofluoroscopic evaluation of swallowing [5, 16].

Risks of the procedure are often minimal and are similar to those with endoscopy. Practitioners must closely monitor the patient for signs of discomfort, gagging, emesis, epistaxis, laryngospasm, and a vasovagal response [17, 18]. Sedation and use of topical anesthetic should be used with caution, as it can impact participation and may interfere with pressure generation [5, 18]. Most of these risks resolve quickly on their own and can be avoided through use of lubricant, skilled placement, having the child fast prior to the procedure, and managing the child's and family's expectations. One must always assess the risk/benefit ratio when determining the appropriate course of evaluation.

In any assessment of feeding and swallowing in pediatric populations, the patient may refuse to comply. If it appears that the patient will be unable to complete prescribed tasks or if it appears that only a limited number of bolus trials will be completed, attempting to match authentic feeding patterns as closely as possible will provide the most meaningful data for description of the patient's swallow function and for informing clinical decision-making. The clinician should keep in mind, however, that comparison to normative data will be invalidated when modifications to systematic swallow tasks are made. Crying, coughing, gagging, and even excessive movement can impart pressure artifacts in the signal, so care should be taken to make note when these events occur [5].

Data Analysis

Pressure representations are displayed in real time during the manometry examination. This can be used for online, qualitative analysis of the data. Gestalt observations can be made about swallowing pressure amplitude, duration, and coordination, and some systems allow for pausing of the data stream for finer-grained analysis. This display can also be used to educate patients and family about swallowing physiology, can serve as a biofeedback tool, and can be a distractor for the patient.

The power of high-resolution manometry, however, comes in the robust, objective data analysis that occurs after the study is completed. As there are no commercial hardware systems designed for pharyngeal high-resolution manometry, there are no software systems available either. Esophageal manometry software systems often can be modified to extract pressure variables of interest, and one can export raw pressure data for analysis in a third-party software, such as MATLAB (MathWorks, Natick, MA). Research teams have devised pressure and impedance analysis software for adult pharyngeal high-resolution manometry [13, 19], but no such software is currently validated for pediatric manometry.

High-resolution manometry data are first segmented into different regions of interest (Fig. 19.1). The *velopharynx* is a region of swallowing-related pressure that arises from velopharyngeal port closure and some contributions from the oral tongue to propel the bolus [7]. The *tongue base* receives pressure from tongue base retraction and pharyngeal wall contraction,

and the *hypopharynx* is immediately inferior and sits around the laryngeal inlet [11]. Some publications group tongue base and hypopharynx together into the *mesopharynx* region [19]. The *upper esophageal sphincter* (*UES)* is defined by a region of elevated resting pressures that relax during swallowing [20]. As oropharyngeal swallowing is a dynamic process with many moving structures, there is a subset of manometry sensors that register pressures while the UES is at an elevated position [21]. High-resolution manometry analysis benefits from the multi-sensor assessment of pressures along the continuum of the pharynx.

Once the data stream has been segmented into different regions of interest, it can be analyzed according to pressure amplitudes and timing. The measures obtained can help the clinician to describe gestalt strength of the pharynx, relative ability of given segments of the pharynx and UES to generate pressure, and ability of the individual to create pharyngeal pressure differentials necessary for propagation of the bolus from the oropharynx to the esophagus efficiently [22]. See Table 19.1 for a description of pharyngeal and UES pressure parameters reported in pediatric pharyngeal high-resolution manometry. In the pharynx, simple maximum pressures and pressure durations are reported most commonly [23–25]. In the UES, relaxation minimum (*nadir*) pressures and relaxation duration describe the relative ease through which a bolus could pass through the UES [23, 26]. Although most high-resolution manometry systems average pressures circumferentially or measure pressure unilaterally from the posterior aspect, a large proportion of pharyngeal and UES pressures come from anterior and posterior directions, with less of a contribution laterally [10, 11]. In addition to pressure minima/maxima, some research groups use *contractile integral* [25] or area under the pressure curve. Pressure integrals may be more descriptive than pressure maxima, as it is measured throughout the entire pressure wave and is a composite measure of the total pressure generated. Pressure velocity and timing between certain pressure events have also been described [4, 22]; these parameters may be more descriptive of pressure coordination than simple pressure durations. Pressure gradients have been described to relate high propulsive pressure in the pharynx relative to low UES pressure during swallowing [27], but these have yet to be described in the pediatric population.

Table 19.1 Measures calculated from pharyngeal high-resolution manometry data

Measure	Purpose
Pharyngeal peak pressure (PP) (mmHg)	The maximal pressure (over all manometric channels or at a specific pharyngeal region) to which the pharyngeal constrictors contract during deglutition
Pharyngeal contractile integral (mmHg∗sec∗cm)	Product of pharyngeal contractile amplitude, duration, and length; a composite measure of pharyngeal contractile vigor
Pharyngeal propagation velocity (cm/s)	Describes the speed of pharyngeal peristalsis
UES resting pressure (mmHg)	Pressure in the UES during quiet rest
UES nadir pressure (mmHg)	Lowest pressure reached in the UES during relaxation; relevant in considering the ease with which a bolus may pass through the UES
UES peak pressure (mmHg)	Highest pressure reached in the UES following relaxation
UES relaxation duration (s)	Measured at nadir +20% of UES relaxation onset-nadir difference; relevant in considering the ease with which a bolus may pass through the UES
UES relaxation response time (s)	Time needed by the UES to reach its most complete relaxation; speaks to coordination of complex movements during deglutition; relevant in considering the ease with which a bolus may pass through the UES
Time between pharyngeal peak pressure and the UES nadir (s)	A marker for the coordination between pharyngeal contraction and UES function

Parameters and definitions derived from Refs. [13, 22–26]
UES upper esophageal sphincter

Table 19.2 Pressure-flow measurement parameters calculated from pharyngeal high-resolution manometry with impedance data

Measure	Purpose
Pressure at nadir impedance (PNI) (mmHg)	In the pharynx or UES, this is a marker of intra-bolus pressure
UES nadir impedance (ohms)	During UES opening, this is the lowest impedance value, indicating the time point and approximate magnitude of maximal UES opening diameter [28]
Post-swallow impedance ratio	Ratio of post-swallow impedance to impedance during bolus passage, and is elevated when there is a large amount of post-swallow residue [29]
Time from bolus distension of the pharynx to peak pharyngeal pressure (TNIPP) (s)	A measure of bolus flow time through the pharynx, indicating how quickly the bolus passed through the pharynx ahead of the pharyngeal stripping wave
Flow interval (FI) (s)	Duration of impedance signal (bolus presence) in the pharynx
Swallow risk index	Composite score developed to identify swallowing motor function that is associated with risk of penetration, aspiration, and post-swallow residue [9]: $\frac{FI \times PNI}{PP \times (TNIPP + 1)} \times 100$

Parameters and definitions derived from Refs. [4, 13, 24, 25, 27, 33]
UES upper esophageal sphincter

If impedance data were collected, a *pressure-flow analysis* can be completed [13, 26]. This type of analysis adds information about the flow of the bolus in relation to the pressures generated. Impedance data in ohms can be inverted to represent *admittance* in Siemens, acting as a surrogate measure of the bolus. This allows for accurate measurement of intrabolus pressure, which can represent the relative ease through which the bolus passes through the pharynx. A too-low intrabolus pressure may represent weak bolus propulsion, and a too-high intrabolus pressure may represent an obstruction. Impedance measures during UES relaxation can act as a surrogate for actual UES opening width [28] and in the pharynx after the swallow suggest pharyngeal residue [29]. Finally, a swallow risk index has been described as a composite measure that can be used to predict penetration/aspiration and post-swallow residue risk. See Table 19.2 for a description of pressure-flow metrics used in pediatric high-resolution manometry studies.

Ethical considerations preclude the collection of high-resolution manometry data in a large research sample of healthy babies and children. This makes comparison of swallowing pressures in patients with dysphagia difficult. Therefore, most researchers use a comparison group of children who received a high-resolution manometry study as standard of care to evaluate for dysphagia but had swallowing function within typical limits (e.g., [23]). Ranges of mean pressure and pressure-flow values reported by these groups are in Table 19.3.

Table 19.3 Ranges of mean pressure and pressure-flow measures from children without oropharyngeal dysphagia

Value	Ranges reported
Pharyngeal peak pressure	22–148 mmHg
Pharyngeal contractile integral	87 mmHg∗sec∗cm
Pharyngeal propagation velocity	3–20 cm/s
UES resting pressure	30–78 mmHg
UES nadir pressure	3–11 mmHg
UES peak pressure	64–72 mmHg
UES relaxation duration	0.4–0.8 s
UES relaxation response time	0.45–0.48 s
Time between pharyngeal peak pressure and UES nadir	0.13–0.22 s
Pressure at nadir impedance (hypopharynx)	5–76 mmHg
Pressure at nadir impedance (UES)	2–11 mmHg
UES nadir impedance	166–218 ohms
Post-swallow impedance ratio	31–116
Time from bolus distention to peak pharyngeal pressure	0.37–0.45 s
Flow interval	0.38–0.66 s
Swallow risk index	−2 to 8

Values reported in Refs. [4, 22–25]
UES upper esophageal sphincter

Special Considerations for the Pediatric Population

Willing participation is essential for collection of meaningful manometric data in all populations. While high-resolution manometry is not a painful procedure, passage of the catheter can certainly be uncomfortable, and presence of a foreign object extending from the nasal tip to the esophagus can be disconcerting for all ages. Basic knowledge of developmental level, common stressors, and strategies for reducing these stressors at each age helps tremendously in completing a valid study. In addition to the strategies outlined below, it is often useful to consult a child life specialist. Child life specialists are certified individuals who provide evidence-based, developmentally and psychologically appropriate interventions for infants, children, and their families [30].

For school-aged children, stressors often include loss of control, pain (or anticipation of pain), or missing school. Methods to mitigate the negative aspects of the experience and to improve data quality include incorporation of choices, deep breathing, and use of visual distractors including videos on a tablet or other device during the procedure. Medical play with equipment or toy equipment or reading a book about manometry prior to the procedure can also be useful [5]. An additional developmental consideration for this age group is that they are often eager to demonstrate their knowledge and in doing so can better process the experience [31].

Stressors for toddlers can include stranger danger, separation anxiety, pain (or anticipation of pain), or change of routine. Strategies for improving tolerance of the exam include having the child sit with their caregiver or hold the caregiver's hand, working to establish rapport before attempting passage of the catheter, distracting with light-up/interactive toys and bubbles, or singing of familiar songs and medical play with equipment or toy equipment [31].

Primary stressors for infants can include stranger danger, separation anxiety, or overstimulation. When performing uncomfortable procedures with infants, waiting for quiescence is essential; it is not uncommon for the infant to require 15–20 min or more to fully acclimate to the catheter and achieve a state of quiescence [31]. Following insertion of the catheter and confirmation of adequate placement, it can be useful for the clinician to step away and allow the parent/caregiver to comfort the child as they typically do at home. Turning down the lights, reducing noise, limiting other excess stimulation, swaddling, or using a pacifier can also be particularly useful [5]. Alternatively, comfort is often sought through feeding following episodes of distress for infants. If the child is sufficiently calm to feed safely, presentation of a bottle or breast at this time may allow for acquisition of meaningful data. For swallowing data that is more representative of typical mealtime swallows, however, it may be advantageous to begin analyzing swallowing data only after the infant has calmed to a baseline level.

It is common for children to take cues from their parents/caregivers on how to respond to potentially stressful medical situations. As such, thorough discussion of the rationale for and process of manometry with the caregiver is essential. When possible, this should be completed prior to the day of the procedure, allowing both the parent/caregiver and their child to prepare for manometry.

Future Directions

Pharyngeal high-resolution manometry is still in its infancy, especially in the pediatric population. As pharyngeal high-resolution manometry is used more frequently in clinical practice, particularly in clinics specializing in pediatric dysphagia management, establishing normative data across multiple ages and clinical populations of pediatric patients will be essential. This will require larger normative datasets as well as an improved understanding of how different disease processes impact pharyngeal pressure generation. As dysphagia diagnosis and classification becomes more sophisticated, the ability to select appropriate management plans and predict prognosis will improve.

Some measures reported in contemporary literature may prove in the future to be more

meaningful than others. As research progresses, key measures that describe swallowing physiology, classify dysphagia phenotypes, and predict prognosis will arise and be adapted. Nonetheless, current manometric measures add a wealth of information to the clinical picture that are unobtainable with other instrumented evaluations, such as videofluoroscopy or endoscopy.

Conclusions

High-resolution manometry objectively measures swallowing pressures along the pharynx and esophagus. While high-resolution manometry is not a replacement for standard of care evaluations such as videofluoroscopy or endoscopy, it has the potential to significantly increase our knowledge of both normal and disordered swallow function and to guide clinical decision-making both surgical and therapeutic, particularly as samples that include larger numbers of more diverse pediatric populations are obtained.

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20 Quality of Life Assessment in Children with Feeding and Swallowing Disorders

Pamela Dodrill and Hayley Henrikson Estrem

Background

Consuming fluid and food is an unavoidable task and a core activity of daily living. Most children and adults eat at least 3 times during the day – every day. Most babies eat around 8 times per day. Eating is generally an enjoyable activity and usually occurs in a social setting. Families spend more time together in mealtime situations than in any other activity. Children bond with their parents and learn trust over mealtimes, and feeding and nourishing your child is considered a fundamental parenting activity.

Like adults, infants and older children can have feeding and swallowing disorders. Unlike adults, children have rapidly developing body systems, and even short-term problems with feeding or swallowing can interrupt normal development and cause serious long-term sequelae. For a child to reach his or her physical and cognitive growth potential, sufficient energy and nutrients must be consumed. Feeding and swallowing disorders can have a detrimental effect on dietary intake and, hence, growth and development. Further, learning to eat is an important developmental process. Children have to learn increasingly complex oral skills to eat more advanced food textures and need to learn to accept new foods of varying texture, taste, temperature, and color. If early feeding development is interrupted, the child may not develop the skills to eat functionally and/or may not develop an enjoyment of eating.

Pediatric feeding and swallowing disorders can have a profound impact on the child's quality of life, as well as that of their parents and family. Feeding or swallowing difficulties are generally stressful in the moment, for both the child and family. In addition, parents will often be driven by concern for the long-term consequences of their child not eating enough and/or not eating "normally." The high stakes involved underlie much of the stress that follows when pediatric feeding and swallowing disorders are present. Children may have a fear of choking or vomiting. Parents may have that same fear about their child and may also fear that the child's feeding/swallowing disorder is an indicator that the child is not "normal" or will not consume enough to develop "normally."

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Dysphagia and Pediatric Feeding Disorders

Dysphagia

As described in previous chapters, dysphagia is an impairment of swallowing function and may occur at the oral, pharyngeal, or esophageal phases of swallowing. Children with dysphagia generally present with a feeding disorder, but not all children with a feeding disorder have dysphagia.

Pediatric Feeding Disorders

Pediatric feeding disorder (PFD) is defined as impaired oral intake that is not age-appropriate and is associated with medical, nutritional, feeding skill, and/or psychosocial dysfunction [1]. To be fully functional, a child's feeding skills must be safe, age-appropriate, and efficient. Dysfunction in any of these areas constitutes PFD (Table 20.1).

Table 20.1 Indicators of dysfunctional pediatric feeding skills

Unsafe oral feeding may present as:
Choking, aspiration, adverse cardiorespiratory events (e.g., apnea, bradycardia) during oral feeds
Other adverse mealtime events (e.g., gagging, vomiting, fatigue, refusal)
Delayed feeding skills may present as:
A child who is unable to consume age-appropriate liquid and food textures. The child may require food/fluid to be modified from its original form (e.g., blending solids into a puree) or may rely on a natural variant (e.g., a naturally smooth food) that is not age-appropriate
A child who has deficits in use of feeding utensils and devices or self-feeding skills. They may require special feeding equipment, positioning, or feeding strategies
Inefficient oral feeding may present as:
Prolonged mealtime duration (greater than 30 min). These children may require modified food textures or special feeding equipment or strategies
Inadequate oral intake. These children may require nutritional supplementation – orally or via gavage tube

From Goday et al. [1], with permission

PFD can arise in association with dysphagia, aspiration, or a choking event. At other times, there is no apparent physical reason for PFD, although aversive experiences in or around the mouth (e.g., tube feeding, suctioning), undetected pain (e.g., as associated with tonsillitis, pharyngitis, or teething), or sensory disturbances (e.g., oral hypersensitivity) may be involved at some level. Factors within the child, caregiver, and the feeding environment can contribute to and maintain PFD (e.g., increased parent attention when the child gags or fusses). Problem feeding behaviors are generally the resultant dysfunction from having PFD (versus the cause), but are often among the first concerns that caregivers express regarding feeding their child.

In the short term, PFD is often managed through the use of compensations (Figs. 20.1 and 20.2). However, parents often hold onto the hope that the child will develop the skills to able to eat "normally." The healthcare team needs to work with the family to determine which feeding goals are achievable and what would lead to the best health outcomes and quality of life for the child and family.

PFD should not be confused with "eating disorders," such as anorexia, which are associated with body dysmorphia and occur in adolescence and adulthood. PFD occurs when an infant or child is unable or unwilling to eat a range of age-appropriate food (and sometimes any food), as a result of poorly developed feeding skills (e.g., delayed oral motor skills impacting their ability to chew and bite) or a fear of trying new foods, often as a result of hypersensitivity to smell, taste, or texture of foods [2]. PFD typically begins as a delayed transition to solid foods, but can progress to food refusal, taking too long to eat, picky eating according to food type and texture, and choking, gagging, or vomiting when eating (see Table 20.2). If children do not learn the physical skills and cognitive behaviors to eat a wide variety of foods, it will be difficult for them to meet their nutritional requirements through oral diet.

In more severe cases, children with PFD will require full or partial nutritional support via gavage tube feeding as a result of their restrictive

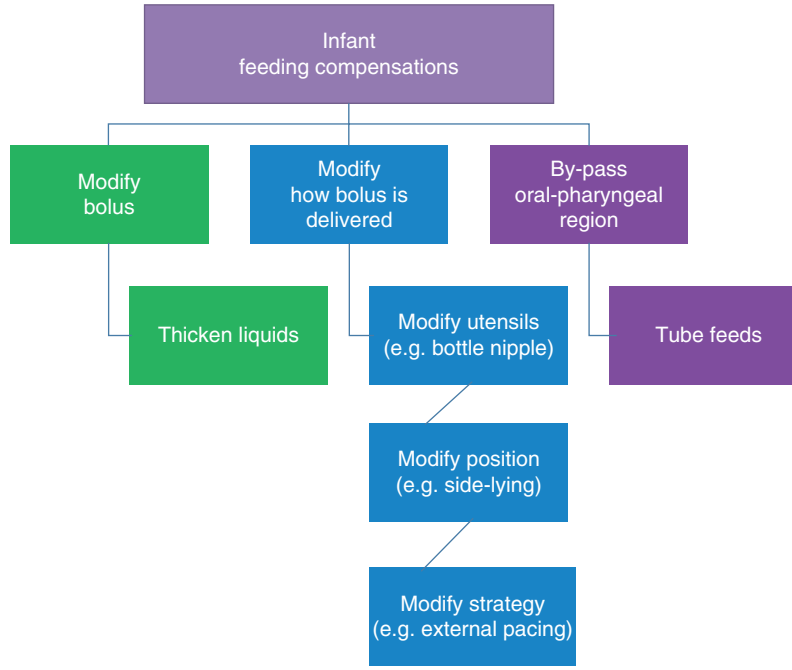

Fig. 20.1 Feeding compensations that may be utilized in infants with pediatric feeding disorders

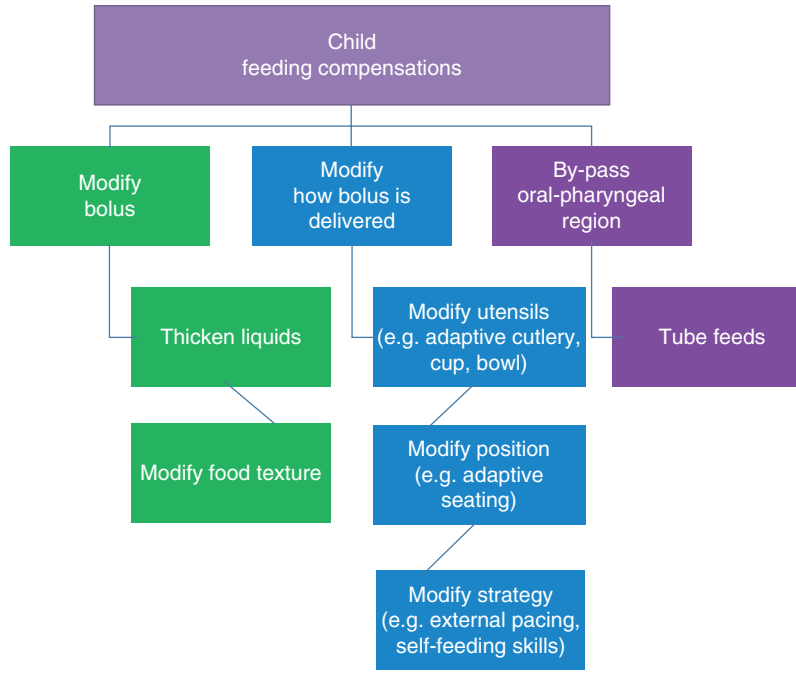

Fig. 20.2 Feeding compensations that may be utilized in children with pediatric feeding disorders

dietary intake. As a consequence, this further restricts the child's opportunities to learn the motor, sensory, and cognitive skills required to eat a variety of healthy fresh foods. Children with *mild* PFD may have a problem in one or more of these key areas, but generally grow sufficiently. Children with *moderate* PFD generally have problems across several of these areas and would not grow sufficiently without nutritional supplementation in the form of oral formula

feeds and/or energy and nutritional supplements. Children with *severe* PFD generally have problems across all of these areas and are unable to meet their fluid/energy/nutritional requirements from an oral diet, thus requiring tube feeding.

PFD can adversely impact a child's quality of life and that of the child's family. Children with PFD often take significantly longer to eat/feed each day, limiting their time to participate in other developmentally appropriate activities (e.g., play) and limiting their parents' time to do the other activities they need to do each day.

Table 20.2 Key indicators of pediatric feeding disorders in children [2]

Restricted oral intake (insufficient intake of energy, nutrients, and/or fluid)
Limited range of food in the diet
Limited range of textures in the diet (often a reliance on "easy-to-eat foods," which are pureed, soft, or dissolvable)
Very low or high weight-for-height
Prolonged mealtime duration (>30 mins at mealtimes, >2 h per day spent trying to feed the child)
Battles/problematic behavior at mealtime
Family stress related to the child's eating patterns

Health-Related Quality of Life (HRQoL)

Health-related quality of life (HRQoL) is defined by the United States Office of Disease Prevention and Health Promotion as "a multi-dimensional concept that includes domains related to physical, mental, emotional, and social functioning. It goes beyond direct measures of population health, life expectancy, and causes of death, and focuses on the impact health status has on quality of life" [3].

There are a number of validated assessment tools for measuring HRQoL in children and their parents or family. Those most relevant to children

Table 20.3 Relevant health-related quality of life (HRQoL) tools for children and families affected by feeding/swallowing disorders

Child HRQoL assessments
PedsQL™ Generic Core scale [4, 5]
Multidimensional: 23 items assess physical, emotional, social, and school functioning
Child with chronic illness self-report and parent-proxy report forms are available
Extensively developed assessments
Generic core scale is not condition specific
PedsQL™ Gastrointestinal Symptoms Module [6] and PedsQL™ EoE Module [7, 8]
These are later versions of the PedsQL designed specifically for children with specific gastrointestinal conditions
These versions come closer to specifically capturing HRQoL for children with feeding disorders, which is often comorbid with gastrointestinal conditions such as eosinophilic esophagitis (EoE)
Parent and family HRQoL assessments
PedsQL™ Family Impact Module [9]
Aimed at identifying impact of health problems on performance of daily activities and relationships:
Child functioning: Physical, emotional, social, school
Parent functioning, family functioning
Not specific to families of children with feeding and swallowing disorders
Feeding/Swallowing Impact Survey (FS-IS) [10]
Specific to parents of children with feeding and swallowing disorders
Parent reported: assesses feeding, worry, daily activities
Feeding Impact Scales (Parent Impact and Family Impact) [11]
Specific to parents and families of children with feeding disorders
Initial item list adapted from Redle's Pediatric Feeding and Swallowing Disorder Family Impact Scale [12]
Item response theory analysis resulted in 13-family impact items and 12-parent impact items
Parent report of impact on self, and parent report of impact on family

with feeding and swallowing disorders are summarized in Table 20.3.

In addition to the tools above, the Functional Oral Intake Scale (FOIS)-Pediatric [13] allows clinicians to describe the degree of functional dietary limitation caused by a patient's swallowing impairment. The original Functional Oral Intake Scale (FOIS) was developed by Crary and colleagues [14] for use in adult patients. An adapted version of this tool was developed for infants and young children [13]. Patients are scored between 1 (minimum) and 6 (maximum) (Table 20.4). This adapted scale has not been formally validated, but has been used in a number of published studies, and our clinical experiences indicate that it adds to the information obtained from the clinical evaluation in infants and young children.

Table 20.4 Functional Oral Intake Scale (FOIS)-Pediatric [13]

1	Nothing by mouth
2	Tube dependent, with minimal attempts at liquids/*foods*
3	Tube dependent, with consistent intake of liquids/*foods*
4	Total oral diet, but requiring special preparation of liquids (thickened liquids) or compensations (e.g., special feeding equipment, feeder uses special strategies)
4.5	*Total oral diet, but requiring special preparation of solids* (e.g., *foods of different texture to peers and/or liquid supplements) or compensations*
5	Total oral diet, without special preparation (i.e., regular thin fluids, *foods of same texture as peers, no additional liquid supplements*), but with compensations
6	Total oral diet, with no restrictions relative to peers

From Dodrill et al. [13], with permission
Italicized items only apply to children over 6 months of age who would be expected to have solids in their diet. Special compensations include special feeding equipment or strategies

International Classification of Functioning, Disability and Health (ICF)

The World Health Organization (WHO) has two main international health classification systems:

- Health conditions (diseases, disorders, and injuries) are classified primarily in the *International Classification of Diseases*, *ICD*, which provides an etiological framework [15].
- Functioning and disability associated with health conditions are classified in the *International Classification of Functioning, Disability and Health*, ICF [16].

These two resources complement each other and are designed to be used together to document health conditions and associated complications.

The ICF model was officially endorsed in 2001 as the international standard to describe and measure health and disability (Fig. 20.3). The *ICF for Children and Youth* (*ICF-CY*) [17] is derived from the ICF and is designed to record

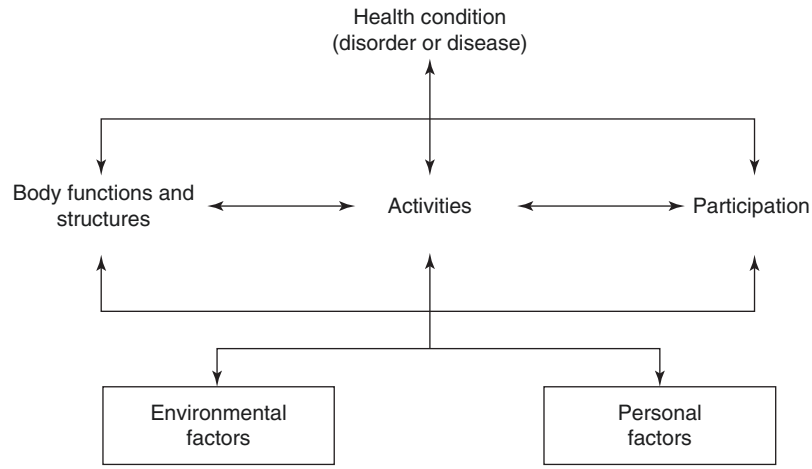

Fig. 20.3 ICF model [15]

Table 20.5 Application of the ICF model to feeding and swallowing in children [13]

Area of ICF model	Relationship to feeding and swallowing
Body structures	Anatomy and physiology of aerodigestive tract
Body functions	Swallowing, sucking, biting, chewing, cognition, motor control, sensory perception
Activity versus disability	Ability to eat a meal, self-feed, drink a bottle, drink from a cup
	Determine, where necessary, whether use of modified food/fluids, special utensils, altered positioning, or special feeding strategies can prevent activity limitations and disability
Participation versus handicap	Participation in family mealtimes and social and educational settings where food/fluid is consumed
	Determine, where necessary, whether social inclusiveness policies and strategies can prevent participation limitations/handicap for children and their families on tube feeds and those who cannot eat developmentally appropriate foods/fluids
Personal and environmental factors	Family's understanding of the child's disorder Family's access to appropriate and hygienic food, fluids, utensils, and seating equipment Where necessary, the family's ability and willingness to prepare modified food/fluids, use special feeding utensils/seating equipment, deliver tube feeds, or apply special feeding strategies Where necessary, the ability and willingness of staff at day care/school to prepare modified food/fluids, use special feeding utensils/seating equipment, deliver tube feeds, or apply special feeding strategies Societal and cultural judgment of families who have a child with feeding disorder Policies to support and include children and families with disability in educational and social settings

health complications manifested in infancy, childhood, and adolescence, as well as relevant environmental factors. Specifically, it is designed to capture health changes associated with their growing competence, societal participation, and independence. The ICF model can be used to map the various areas that can be impacted by a child's feeding or swallowing disorder (Table 20.5).

Child HRQoL

To be fully functional, a child's feeding skills must be safe, age-appropriate, and efficient. Dysfunctional feeding and swallowing disorders may result in a child being offered age-appropriate fluids/foods, but struggling to consume them safely; a child being offered modified fluid/food, or requiring special mealtime positioning, utensils, or feeding strategies; or a child being offered fluid/food via non-oral means.

For some children, not eating by mouth can adversely affect QoL. For others, not having to eat by mouth or not having to eat foods that are beyond their skills can enhance QoL. There are multiple factors which affect QoL in children with feeding and swallowing disorders (Tables 20.6 and 20.7) [10, 18–20].

Therapy Considerations

Nature of the Condition

Depending on the status of the child's underlying medical or developmental condition, the appropriate assessment and treatment plan may be quite different. For some children, the long-term goal is cure of dysphagia or age-appropriate feeding skills. For others, the goal may be to achieve developmentally appropriate feeding skills, knowing they may not achieve age-appropriate skills, or to achieve functional feeding and swallowing skills with the use of modified food or fluids, special feeding equipment, or other compensations. For some children, the best goal may be to try to slow the decline in their feeding or swallowing skills, or to minimize the risk of aspiration or malnutrition by having small oral feeds

Table 20.6 Common factors affecting QoL in children with feeding and swallowing disorders who receive oral feeds

Physical health
Pain and discomfort from eating/drinking
Nausea during meals
Difficulty breathing during meals
Unpalatability of thickened feeds, special diets, or medications
Fatigue from the effort of eating
Fatigue from underlying health issues
Mental health
Reduced enjoyment of eating and mealtime interactions
Frustration at the task of eating and mealtime interactions
Stress manifesting as mealtime behavioral issues – aversion, avoidance (fight or flight responses)
Parental disappointment
Feelings of anger or resentment from parents and other family members
Embarrassment caused by eating differently
Unwanted attention from others
Reduced appetite from stress associated with eating and mealtimes
Exclusion or isolation from family and friends
Participation in activities of daily living
Eating may take up a lot of time and limit time for other activities
Eating can be a hassle when adaptations or compensations are needed
Not eating normally affects participation in mealtimes and special events

Table 20.7 Common factors affecting QoL in children with feeding and swallowing disorders who receive tube feeds

Physical health
Pain or discomfort from presence of the tube
Nasogastric tube: taping, insertion, removal, irritation from presence, increased reflux from presence
Gastrostomy tube: need for surgery, site infections
Nausea if feeds are delivered too quickly
Mental health
Reduced participation in mealtime interactions
Dependence on others to help with tube feeds
Scary medical visits and tests, time spent in hospital
Embarrassment caused by presence of the tube or eating differently
Unwanted attention from others
Exclusion or isolation from family, friends, peers
Daily activities
Loss of daily organization, which is often based around mealtimes
Not eating normally affects participation in mealtimes and special events
Requiring tube feeds may impact access to day care or school
Time and stress for healthcare visits (planned, emergency, cumulative number and duration)
Time in hospital or unnatural environments
Stress from imaging and procedures
Time away from family, friends, home, pets, etc.

and tube top-ups, or to have all feeds via a tube and have a non-oral stimulation program. For many children, feeding goals change at different points in their medical and developmental course. Regular assessment and reassessment is needed to set meaningful, functional goals and to monitor outcomes against those goals. Either the intervention or the goals need to be changed if progress toward the goals is not being achieved.

Nutritional Stability

When an unwell child is nutritionally compromised, the primary focus of nutrition and feeding management is ensuring the child consumes enough energy, nutrition, and hydration to meet basic requirements. At these times, this focus supersedes considerations for promoting an oral diet or a developmentally appropriate diet. This may mean using parenteral feeds or using enteral feeds that are delivered continuously. Some children may require special feeds or supplements that are unpleasant tasting and are better tolerated if given via non-oral means. During this time, the role of the feeding therapist is to assess whether it is possible to introduce activities that can promote normal oral experiences (suckling on a pacifier, chewing on teething toys) and minimize adverse oral experiences while the child is not consuming a typical oral diet.

Medical Stability

In medically complex children, there are times when the greatest focus is on managing acute health complications. Feeding assessment and

intervention may not be appropriate or a priority. In contrast, there are times when care can focus on supporting developmentally appropriate activities, such as feeding. Between these events, there are often times when feeding assessment and intervention can start to be introduced, provided they do not interfere with other essential healthcare activities. Some medically complex children undergo multiple cycles of acute illness and corresponding treatment, and frequent monitoring of feeding skills is required to track progress or regression across these cycles.

Limitations Caused by Medical Treatments and Hospital Environment

In general, it is often hard to replicate normal mealtime experiences in the hospital environment. Fragile infants may not tolerate the handling required for feeding, and older children who are unwell may not tolerate sitting upright for meals. Patients are often confined to their beds and may not have access to normal seating or positioning options for meals or the ability to participate in social mealtimes with others. Hospital food is notorious for being bland and lacking variety and appeal. The sights and sounds of the hospital environment can produce anxiety, and the variety of smells are often unpleasant when eating. In addition, feeding schedules may need to be interrupted if the patient displays nausea, pain, irritability, or fatigue related to his or her illness, medications, or other interventions. Children who need frequent surgeries often have to have their feeding schedules interrupted by the need to fast before, during, and in the time immediately following surgery. Again, the feeding therapist must be considerate of these factors when working with hospitalized children. The feeding therapist also has a role in advocating for developmentally supportive practices, such as the provision of age-appropriate food and feeding equipment, to assist in promoting normal feeding development.

Once the child's medical and nutritional status has stabilized, there is often a sudden push for oral feeding assessment and intervention to be prioritized when a patient is preparing for discharge home. The feeding therapist plays an important role during this time; however, the therapist must resist pressure to clear a child for full oral feeding and discharge if the child or the family is not fully competent in the tasks that will be required for the child to manage full oral feeds at home.

Social Aspects of Eating

Mealtimes are supposed to be social. For infants, more of their awake and interaction time is spent feeding and eating than on any other activity. Much of early parent-child bonding occurs at mealtimes, and children learn early turn-taking and many other communication skills from mealtime interactions. For older children, many important family events are celebrated with meals. In addition, many friendship-building activities are based around sharing meals. Thus, feeding assessment and management needs to consider the effect the child's feeding disorder has on social participation in meals and ultimately the effect on QoL.

Feeding Interactions

The feeding observation should include an assessment of how the caregiver and child work together as a team during feeding. Children who have experienced pain or discomfort with feeds may learn to dislike and avoid feeds and may also show aversion toward the caregiver as part of a classically conditioned response. Unfortunately, caregivers may unintentionally reinforce food refusal behaviors by giving in when the child protests. Long periods of hospitalization and separation can affect the normal bonding process. Children who have been acutely unwell may not know how to interpret the feelings of hunger and fullness and may give mixed or unclear cues to their feeders, which can make the caregivers nervous or apprehensive about feeding the child. Further, parents of children who have been

acutely unwell or who are medically complex are often stressed and fatigued, which can affect their coping mechanisms and their ability to support the child. Many of these children have prolonged mealtimes and need much support and encouragement to feed, which puts a lot of extra responsibility on already stressed caregivers. All of these factors can ultimately affect parent and child QoL.

Parent and Family HRQoL

Parents and families of children with dysphagia and PFD are challenged in unique ways. PFD is a state of dysfunction and is not a classically diagnosed as a disease or condition such as diabetes or congenital heart disease. PFD can be caused by impairments in any of the four key domains of functioning: medical, feeding skills, nutrition, and psychosocial (Fig. 20.4) [1]. In turn, PFD can lead to dysfunction in any of these areas.

Diagnostic criteria for PFD in the psychosocial domain include caregiver distress at mealtimes and the mental health of caregivers and family [1]. Feeding one's child is a core task of parenting. As discussed previously, parents inherently feel pressure to make sure their child receives adequate nutrition to grow and develop.

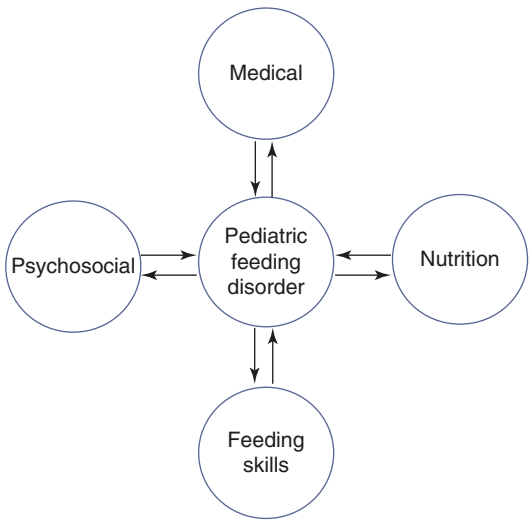

Fig. 20.4 Four domains of pediatric feeding disorders [1]

Table 20.8 Impacts of PFD on parents and family

Impacts on parents
Worry for child's health and development
Feelings of failure
Isolation or being home-bound
Greater than usual amount of time spent preparing food and feeding
Limited ability to work outside of home
Few or no others able to feed child
Impacts on family
Limits on participation in social activities
Restaurants can be difficult or impossible
Disproportionate time focused on one child
Child with PFD may be fed outside of family meal
Parents may disagree on problem and how to act

If a child has PFD, parents often feel overwhelmed and may feel disappointed in themselves. This may be compounded by feeling judged, misunderstood, or stigmatized by others (Table 20.8).

Often the typical go-to resources for guidance on issues with infant and young child feeding are *not* helpful for PFD. Well-intentioned friends and family can provide advice that is unhelpful and sometimes may be frustrating or detrimental. Many health providers are ill-equipped to help with complex issues, and waitlists for specialty providers can be very long. Many parents of children with PFD will report spending months to years struggling before finding appropriate feeding help. This journey is further isolating and stressful, and mealtime interactions can become a negative mealtime cycle (Fig. 20.5) [18, 19].

Feeding therapists generally work directly with children during therapy sessions with the aim of improving the child's skill and/or behavior. It is, however, essential to note that parents are the individuals who will carry out the vast majority of meals, along with being responsible for all other aspects of the child's care. Hence, it is vital that family members are involved in setting meaningful therapy goals and are taught how to implement therapeutic strategies to facilitate generalization to the home environment. There is not yet research on family intervention for children with PFD; however, there is evidence that this is a promising approach. A recent meta-analysis of 37 family research studies with other

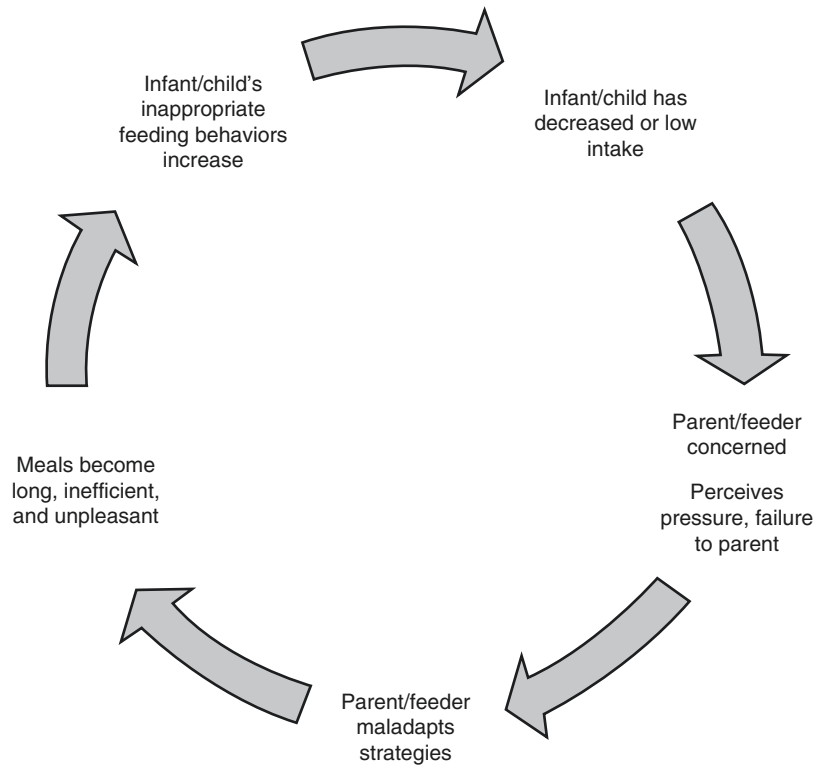

Fig. 20.5 Negative mealtime cycle. (Reprinted with permission from Estrem et al. [19], with permission)

childhood chronic conditions has shown that optimized family function positively impacts child well-being [21]. Additionally, interventions involving family members have been shown to improve both child and family outcomes [22–24].

Generally, the first step in alleviating the impact of health conditions for parents and family and improving their HRQoL is to interrupt the dysfunction in the family system via intervention. For children with PFD, this is often best achieved by a team approach, with input from a mental health provider, in addition to medical, nutrition, and feeding therapy providers [1]. Strategies that all team members can use to support parent and family management of PDF include:

- Partner with parents to personalize a child- and family-centered treatment plan, which can accomplish therapeutic team goals in daily settings.
- Be sure to point out what you see is going well, and celebrate accomplishments.
- Find strengths of the child and of the parents, and reinforce them.
- Facilitate training of other feeders to avoid primary caregiver burnout.
- Know some basics of social program resources the child and family may qualify for in your region.

Support Groups

In addition to the support provided by health professionals, there are a number of feeding and swallowing support groups to assist patients and their families. Some of these are aimed at families of both pediatric and adult patients (National Foundation of Swallowing Disorders – www.nationalswallowingfoundation.com). Others are pediatric specific (Feeding Matters – www.feedingmatters.org; Feeding Tube Awareness – www.

feedingtubeawareness.org). Additional health condition-specific support organizations also exist (e.g., March of Dimes – https://www.marchofdimes.org/).

As discussed, pediatric feeding and swallowing disorders can have a profound impact on the child's quality of life, as well as that of their parents and family. A thoughtful and proactive approach from healthcare providers and other support networks can assist to minimize the functional restrictions caused by the child's underlying feeding and swallowing impairment.

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Additional Material

Meet Lucy.: http://bit.ly/MeetLucyFM. n.d.
Meet Noah.: http://bit.ly/MeetNoahFM. n.d.

Approach to Pediatric Voice Therapy

21

Maia N. Braden

Overview

Voice therapy is frequently used as a primary treatment modality or in conjunction with medical and/or surgical management of voice disorders in children. Advances over the past decade have seen improved delivery of voice therapy to children, and it has been found to be an effective treatment for dysphonia associated with a variety of disorders.

Introduction

Behavioral voice treatment has a growing body of evidence for effectiveness in children. Studies have shown improvements in perceptual and acoustic ratings of voice quality, overall vocal function, vocal stamina, resolution of lesions, and improvements in voice-related quality of life following voice therapy for benign vocal fold lesions and muscle tension dysphonia [1–6]. Overall, available literature suggests that voice therapy, including both indirect and direct voice therapy, can be effective in treating dysphonia in children with nodules, other benign lesions, and muscle tension dysphonia. The exact combination of approaches, duration, and frequency that are most effective is not fully understood, and future research is needed to determine these.

Voice therapy in children, as in adults, focuses on changing the way that the voice is used, to produce a healthy, functional, and, if possible, acoustically pleasing voice. The way that this is achieved is highly individualized. A recent publication by Van Stan and colleagues [7] proposes a taxonomy and structure for describing and categorizing voice therapy approaches. The authors divided intervention into direct and indirect and then characterized direct intervention tools based on overlapping categories. This can serve as a useful way to conceptualize the variety of interventions that can be used. Voice therapy approaches for children should be chosen based on the underlying anatomy and physiology, the patient's current function, their goals and needs, and their learning style and developmental level. Approaches used with adults can very often be adapted to children with some thought about how to make it fun, functional, and understandable at their level. In the past, the focus of voice treatment, especially in children, has tended to focus on reducing overall voice use, and elimination of "vocally abusive" behaviors assumed to be the cause of the voice disorder [8, 9]. Focus on elimination of "vocal abuse" can still be found as a

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primary element of voice therapy in children (as evidenced by the high number of "vocal abuse checklists" available for purchase or download), but overall, the field of speech-language pathology has moved toward more direct, functional, and developmentally focused approaches to voice therapy in children [10–12]. In general, giving children prohibitions rather than solutions is not likely to be effective. The following are examples of commonly used voice therapy approaches, as well as recommendations for adapting these for use with children. These examples should serve as an overview only and do not replace practical training. Clinicians are encouraged to seek out in-person courses and one-on-one mentorship to learn how to deliver these therapy approaches. Clinicians should also draw on their knowledge and understanding of child development to adapt strategies appropriately to the age and developmental level of the child.

Therapy Approaches

Semi-occluded Vocal Tract Exercises

By creating a semi-occlusion at the level of the lips, tongue, or farther forward as with a straw, an optimal glottic configuration is achieved, with the vocal folds barely approximated. The rationale and physiologic underpinnings of these exercises are well described by Titze [13]. Self-sustained vocal fold oscillation occurs with the semi-occlusion, and voicing is produced with maximal output with minimal effort or strain. This can be effective in working with dysphonia related to hyperfunction (muscle tension dysphonia, vocal fold nodules) because it allows children to produce clear, functional voice without excessive impact forces. Conversely, it can also be helpful in achieving better vocal fold vibration in children with hypofunctional voice disorders (vocal fold paresis or paralysis, hyperfunctional underclosure, scar, reduced respiratory support) as it works to coordinate all three subsystems of voice for optimal voice quality. In working with children, semi-occluded vocal tract exercises are often approached through play. Straw phonation, blowing bubbles, and performing lip trills all create a semi-occlusion in the front of the vocal tract, resulting in improved efficiency of vibration at the level of the vocal folds, and more efficient sound with less effort.

These are all easily adapted to play situations. Examples of semi-occluded vocal tract exercises include phonation through a straw, blowing bubbles into water through a straw, humming, lip trills, tongue trills, and a "kazoo buzz" style sustained */u/*. To adapt these to play, one can assign a sound to each vehicle when playing with cars – for example, a fire truck can be pitch glides, a plane can be a hum, a boat can be a lip trill, and a car can be a resonant "voomm" sound. With older children, these same exercises can act as turns in a game – for example, when playing Candy Land, you can do a lip trill for each purple, a hum for each yellow, etc. Many children find a game of "soccer" while moving a ball of paper or a ping-pong ball using straw phonation fun and motivating.

Resonant Voice Therapy

Resonant voice therapy is based on the work of Arthur Lessac [14] and was further developed into Lessac-Madsen Resonant Voice Training [15]. Resonant voice calls for a focus of vibratory sensations in the lips and face, with an absence of strain or effort in the throat. Typically, humming, chanting with nasal sounds, and nasal-loaded words and sentences are used to facilitate this production and generalize it into everyday speech. This is based on the concept that sensation of these vibrations, combined with easy phonation, reflects the optimal configuration of the vocal folds during vibration [16]. The configuration has been described as "minimally adducted, minimally abducted," allowing for efficient, effective phonation without unnecessary effort. Even very young children can identify a sensation of "buzz" or "tickle" in their lips on a hum or /v/ sound, and this can be shaped into words with resonant voice. Training of this approach uses principles of motor learning to enable the child to learn to use their healthier voice all the time. As with semi-occluded vocal tract exercises,

this approach can be effective both with hyperfunctional and hypofunctional voice disorders.

In adapting to children, we often create games with a focus on /m/ or /v/ loaded words, (e.g., *moon*, *mouse*, *mine*, *milk*, *mail*, *very*, *vine*, *vase*, *violin*). These can be as simple as "memory" or "go fish" with articulation cards, self-created Bingo games, or fishing with a magnetic pole. When moving into connected speech, I often use games requiring sentences, such as Guess Who (Hasbro), Headbanz (Spin Master), or 20 questions. Later, depending on the age, connected speech may take the form of imaginative play or a conversation.

Adventures in Voice

Adventures in Voice (AIV) is a resonant voice-based voice therapy program created by Katherine Verdolini Abbott and combines several of the approaches listed above with child-friendly games and activities, as well as a teaching style based on motor learning and child development [17, 18]. AIV is offered through in-person and webinar-based specialty training. This program was recently studied in a randomized prospective clinical trial, comparing AIV with vocal hygiene education only [3]. Both groups showed improvements in quality of life, acoustic, and perceptual measures, with no statistically significant difference in improvement between the groups. There were age-based differences in results, and younger children benefitted more from vocal hygiene, while older children benefitted more from a combined approach. Children recruited later in the protocol benefitted more from the AIV program than hygiene alone, which may indicate that the skill and experience of clinicians plays an important role in therapy success.

Vocal Function Exercises

Vocal function exercises are another form of voice production with a semi-occluded vocal tract. As described by Stemple and colleagues [19, 20], these exercises are designed to rebalance the subsystems of respiration, phonation, and resonance for optimal voice production. Similar to resonant voice therapy and semi-occluded vocal tract exercises, these can be used for both hyperfunctional and hypofunctional voice disorders. These are well described in multiple papers and texts, and typical adaptations are described as well. While not studied in children, vocal function exercises have shown effectiveness in improving voice quality and voice-related quality of life in adults with benign mass lesions [21].

Vocal function exercises as described by Stemple [11, 19] consist of four exercises:

1. Sustained /i/ vowel on musical note F above middle C (for adult females) or F below middle C (for adult males).
2. Stretching: glide from lowest to highest note on the word "knoll" with rounded lips, forward focus, and no voice breaks.
3. Contraction: glide from a comfortable pitch to lowest pitch on "knoll," again with rounded lips, forward focus, and no voice breaks.
4. Sustained pitches: sustain the sound "oll" (as in "knoll") on pitches C, D, E, F, and G above middle C with forward focus, rounded lips, and vibratory sensations in lips. These are produced quietly, but with consistent voicing.

These exercises can be adapted easily to children, although there is frequently a need to change the pitches to suit the child's range and sometimes to simply say "low, medium, and higher" pitches, as very young children can have difficulty with pitch matching. In making this activity more fun or play-based, pitch glides can be done while playing with a toy airplane or involving full body movements of reaching up high and down low. Sustained pitches can be made more fun with moving cars or trains on a track along with the pitch, drawing lines or loops on paper, or simply reinforcing with a turn in a game or play activity.

Flow Phonation

Flow phonation [22, 23] is another form of semi-occluded vocal tract therapy and focuses more on easy flow of air during phonation. I have found it

to be especially useful for children who breath-hold when they speak or have difficulty coordinating phonation with respiration. Cup bubbles (blowing bubbles into a cup), gargling, and prolonged */u/* phonation are the primary components of this therapy approach, with transition into syllables and words. As with resonant voice therapy, different types of games and activities can be used at each level of complexity. Cup bubbles, gargling, and */u/* can be done in coordination with turns in a game, and syllables and words can be part of a game or imaginative play activity.

Holistic or Eclectic Approach

As stated above, therapy should consist of strategies tailored to each individual child's needs and abilities. In our clinical practice, a therapy course might incorporate all of these approaches and more. Based on my experience, therapy can look very different with two different children or two different clinicians, as a skilled voice clinician uses their knowledge of anatomy, physiology, acoustics, learning, and child development as well as observations about the individual child's motivation and learning style to apply evidence-based approaches to each patient.

Emerging and Evolving Practices

With mobile technology becoming ubiquitous, it is no surprise that many of the emerging and evolving practices in voice therapy involve some form of mobile device. Teletherapy, ambulatory monitoring, and mobile app-based practice and therapy are areas of growth in both pediatric and adult therapy. Telehealth is growing in popularity and third-party payor reimbursement, and there is ongoing work on the application of telehealth to voice therapy. While there is still not extensive evidence on telehealth in voice, several preliminary studies indicate that voice therapy delivered through telehealth can be an effective service delivery model [24–27]. Doan and colleagues [28] have reported on a web-based voice therapy portal used for delivery of teletherapy for children with voice disorders, as well as for home practice. This model allows for specialized services to be delivered in more remote areas and also for collaboration with school-based or generalist medical clinicians who may not have experience in voice and voice disorders. Generalization of vocal behaviors outside of the therapy room is a widely recognized challenge in voice therapy. Ambulatory phonation monitoring with small accelerometers placed on the neck has been used in adults to monitor voice use outside of the therapy session and provide feedback and guidance on voice use [29–35]. More recent developments on this device involve an interface with a smart phone, opening up more possibilities for ambulatory monitoring [32]. While this has not yet been studied in children, it is not a large stretch to suppose that it could be helpful in this population as well. Smart phone technology is also being used to support home practice and generalization [36–38], with features ranging from recordings of practice exercises to simulated phone calls to encourage adherence to practicing voice techniques outside of the therapy room. It is likely that all of the technologies will continue to be developed and refined in years to come to support voice therapy in children.

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Benign Mass Lesions

22

Matthew R. Hoffman, Maia N. Braden, and J. Scott McMurray

Overview

Benign mass lesions, particularly vocal fold nodules, represent a common etiology of pediatric dysphonia. Traditionally, a minimalist approach to management has been employed, with observation and counseling that lesions will resolve with aging. With increased knowledge regarding the pathophysiology and natural history of these lesions as well as the psychosocial ramifications of persistent dysphonia, improved assessment and treatment strategies have been developed. Key to effective management of any benign mass lesion is accurate diagnosis, as some (e.g., nodules) are more responsive to behavioral modification and voice therapy than others (e.g., cysts). In the case of a recalcitrant lesion, intraoperative evaluation with vocal fold exploration may be required to make a diagnosis, with definitive treatment potentially being rendered at the same time. This chapter reviews the nomenclature and clinical characteristics of benign true vocal fold mass lesions as well as the treatment of them, including both nonoperative and operative approaches.

Definitions

The most common benign vocal fold mass lesions include nodules, polyps, cysts, and pseudocysts (Fig. 22.1). Nomenclature of benign vocal fold lesions has been a point of contention, with ongoing disparities regarding how lesions are named and described [1]. Rosen et al. conducted clinical consensus conferences to develop a nomenclature paradigm [2]. Definitions from that consensus are provided here and supplemented as indicated.

Nodules are bilateral thickenings of the membranous true vocal folds, often at the junction of the anterior and middle thirds [2, 3]. They can be

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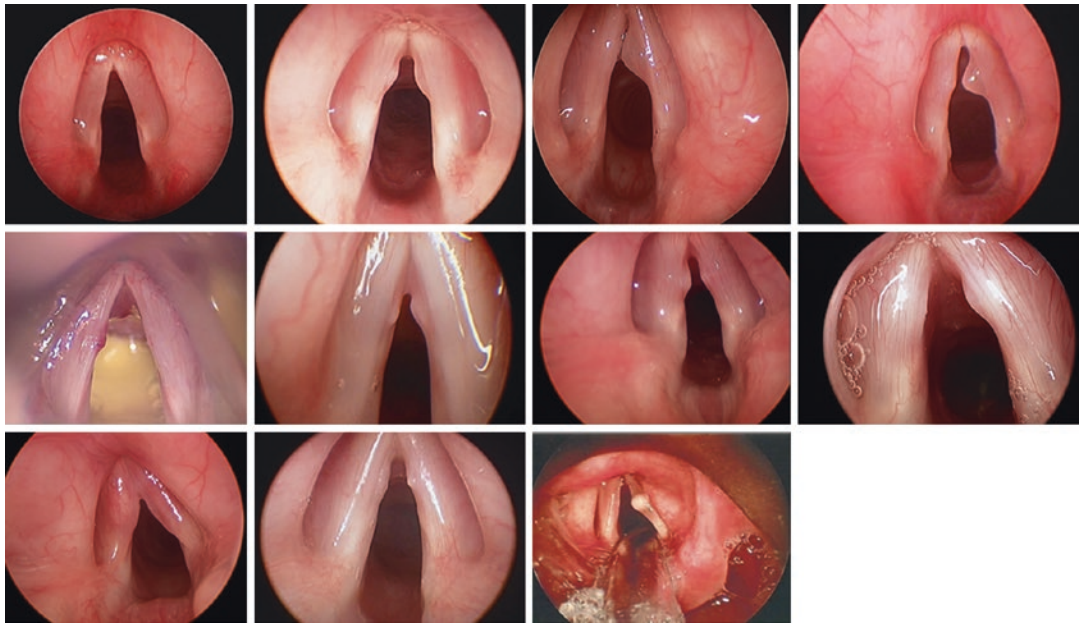

Fig. 22.1 Appearance of benign mass lesions is varied within and across pathology types. Top row: normal; symmetric nodules; right sessile polyp; right sessile polyp with left reactive nodule. Middle row: left sessile hemorrhagic polyp; asymmetric nodules; asymmetric nodules; right pseudocyst. Bottom row: left cyst with right sulcus; right nodule with left reactive fibrosis; right dermoid cyst

symmetric or asymmetric. They are comprised of thick fibronectin deposits in the superficial lamina propria which can be accompanied by basement membrane injury [4]. There is typically minimal impairment of the mucosal wave, and nodules tend to improve with voice therapy [2]. They represent the most common benign lesion in children [5, 6].

Polyps are exophytic masses that are often unilateral but can be bilateral, with a thin overlying epithelium [2]. They can be further classified as pedunculated or sessile according to the nature of attachment to the native vocal fold, as well as hemorrhagic or nonhemorrhagic [7]. The mucosal wave is typically minimally reduced. Surgery is often required for polyps, though voice therapy may be adequate for smaller lesions [8–10]. Polyps are rare in children.

Cysts are subepidermal epithelial-lined sacs within the lamina propria [7]. They may be further divided into those within the subepithelial space versus the vocal ligament [2]. They can be either mucus retention or epidermoid in origin and can be either congenital or acquired. Cysts are typically unilateral but can be bilateral.

Pseudocysts are superficial, subepithelial lesions with a clear, vesicle-like appearance [2]. They are sometimes associated with glottic insufficiency [2, 11]. The lesion is composed of a semisolid fluid or localized edema within Reinke's space [2, 12] deep to thinned epithelium [2]. Importantly, in contrast to cysts, there is no encapsulation.

Epidemiology

Pediatric dysphonia is common. Prevalence estimates vary depending on study, location, methodology, and definitions. A rate of 1% in the United States was reported based on the 2012 National Health Interview Survey [13]. Carding et al. reported a prevalence of 11% based on parental report and 6% based on clinician report in a cohort of 7389 8-year-olds in the United Kingdom [14]. A smaller study in Finland found a rate of 12%, with higher rates for boys (15.8%) compared to girls (7.8%) [15]. A significant portion of children with persistent dysphonia have vocal fold nodules. The estimated prevalence of nodules among children presenting with dysphonia also

varies significantly, from estimates of 5–35% [16, 17] to 38–78% [5]. Data on the prevalence of cysts, pseudocysts, and polyps are less widely reported but are relatively uncommon [18].

Pathophysiology

During vocal fold vibration, the junction of the anterior and middle thirds is exposed to maximal shearing and collision forces, resulting in vascular congestion and edema, with eventual hyalinization of Reinke's space with hyperplasia of the overlying epithelium [7]. Nodules are acellular and composed of thickened epithelium over a dense fibrous stroma with increased type IV collagen and fibronectin [19].

Polyps are caused by impaired circulation followed by thrombosis, exudate, and edema in the lamina propria, with secondary inflammation and atrophy of the epithelium [20]. Hemorrhagic polyps may have a feeding varix [7]. Compared to nodules, they are typically more vascular with less organized collagen, though the distinction can sometimes be challenging [7].

Cysts can be either mucus retention or epidermoid in origin. Mucus retention cysts form secondary to an obstructed mucous gland and expand secondary to retained secretions. Epidermoid cysts can be congenital, developing from cell nests in the subepithelium of the fourth and sixth branchial arches, or acquired, developing from buried mucosa within injured, healing epithelium [7]. Pseudocysts typically develop secondary to phonotrauma in the setting of glottic insufficiency [2].

Presentation

Patients with benign mass lesions will typically present with dysphonia, which can be described as intermittent. If the patient is a singer, there may be inability to maintain phonation in the falsetto register. Voice may be breathy and require additional effort to produce. Vocal fatigue is commonly reported. Dysphagia is uncommon, and the presence should prompt consideration of an alternative or comorbid etiology. In rare cases with large lesions occupying a clinically significant portion of the airway, there may be history of stridor or change in respiration.

We are learning more about the psychosocial impact of dysphonia on children. If questioned, children may report anger, sadness, and frustration, with negative impacts on quality of life [21]. Further, children with dysphonia can be viewed negatively by others [22, 23].

Predisposing factors to vocal fold inflammation should be identified and treated if present. These can include vocal overuse or misuse, secondary or primary smoke exposure, laryngopharyngeal reflux, and allergic rhinitis. A recent study by D'Alatri and colleagues also identified attention-deficit hyperactivity disorder (ADHD) as a potential risk factor [24]. ADHD is one of the most common psychiatric disorders in children [25] and can include loud, impulsive vocalizations as a feature [26], predisposing to phonotraumatic vocal fold lesions.

Speech-Language Pathologist Approach

Benign vocal fold lesions are the most common cause of dysphonia in children, with nodules being the most frequently identified of these. Collaboration between speech pathologist and laryngologist in the identification and treatment of benign lesions is vital. The speech pathologist's (SLP) role in evaluation involves careful evaluation of the child's history, voice use, perceptual and instrumental assessment, and visualization of the larynx. The SLP can assess the impact of both the lesion(s) and the patient's use of adaptive or maladaptive compensation and stimulability for improvement with voice therapy. The SLP's role in treatment is primarily behavioral therapy in the treatment of nonsurgical lesions and perioperative therapy in the treatment of lesions more suited to surgical excision.

History

The SLP should take a detailed history of the child's voice complaints as well as voice use.

They should ask questions to the parent or guardian but also to the child at a developmentally appropriate level. The following components of the history are important in evaluating the dysphonia as well as the child and family's motivation to make any changes:

1. Duration and onset of dysphonia
2. Impact of dysphonia on emotional, social, and academic function
3. Changes in behavior due to voice
4. Typical daily voice use (e.g., does the child talk nonstop, do they scream and throw tantrums, and do they have heavy daily voice needs with sports or theater?)
5. Other health factors that could be contributing to dysphonia, including allergies, asthma, acid reflux, and pulmonary compromise

Quality of Life Measures

These are detailed in Chap. 13 and can be helpful in determining the impact of the voice disorder on the child and their family, as well as their interest in pursuing treatment. Children with benign vocal fold lesions were found to have elevated scores on the Pediatric Voice Handicap Index, and there was no statistically significant difference by lesion type [27].

Perceptual Evaluation

The use of a perceptual instrument such as the consensus auditory perceptual evaluation of voice (CAPE-V) or the grade, roughness, breathiness, asthenia, and strain (GRBAS) scale is necessary to quantify the severity of dysphonia, characterize the dysphonia, and measure change over time. There are no studies indicating that severity of dysphonia differs by lesion type. There is a weak correlation between perceived severity of dysphonia by the clinician and the patient's perception of impact on quality of life [28–30], so these measures should be taken as complementary.

Acoustic and Aerodynamic Evaluation

Detailed descriptions of acoustic and aerodynamic measures and how they are obtained are found in Chaps. 8, 9, 10, and 11. Children with dysphonia due to benign lesions exhibit abnormal values on acoustic parameters such as jitter, shimmer, and noise-to-harmonic ratio [6] compared to peers without dysphonia. Mean values of aerodynamic parameters in children with and without benign mass lesions are lacking, but serial measurements can be used to monitor changes with treatment.

Laryngeal Visualization

The best visualization of vocal fold lesions and their impact on vocal fold vibration and function are obtained using videostroboscopy or high-speed videoendoscopy. 70-degree rigid endoscopes or distal chip flexible endoscopes provide the best images, but it can be difficult for very young children to tolerate and participate in the exam. In these patients, it can be challenging to characterize the stroboscopic parameters, and a combination of history, perceptual, acoustic, and aerodynamic evaluation will be critical to arriving at a correct diagnosis and appropriate treatment plan.

When evaluating stroboscopic video of benign lesions, the SLP does not diagnose the lesion; however, we do describe its presence or absence, appearance, location, and impact on glottic closure and vibration. Careful observation of the primary effects of the lesions on stroboscopic parameters, as well as any compensatory behaviors in reaction to the lesion, is necessary to plan treatment. Closure pattern, pliability or stiffness, and mucosal wave symmetry all influence clinical decisions for how to proceed with therapy.

Treatment

Voice therapy is the gold standard treatment for dysphonia secondary to nodules and either with

or without surgery for cyst and polyp [31]. Voice therapy has well-established effectiveness in treatment of nodules in adults [32–36]. There is a growing body of evidence that voice therapy is also effective in treating nodules in children [37–41]. Studies have included a mix of direct and indirect therapy approaches and have examined different outcome measures including quality of life, perceptual voice quality, acoustic and aerodynamic evaluation of voice, and lesion resolution. As such, it is difficult to compare findings across studies. In general, though, these studies have shown that children who undergo voice therapy achieve improvement or resolution of their dysphonia.

Nodules are frequently cited as being the result of "vocal abuse" or "vocal misuse," terms that are falling out of favor and being replaced by "phonotrauma." Both the quantity and quality of voice use influence development of lesions, but it is not clear why some children who clearly are both heavy and strained voice users do not develop lesions, while some with less exuberant vocal use do. However, while the research is still developing on this, the treatment of dysphonia related to nodules is focused on changing the way that children use their voices, specifically the manner in which the vocal folds contact, the coordination of subsystems of voice, and the use of unnecessary muscle activation. As nodules are assumed to form from excessive repetitive force on the vocal folds, voice therapy focuses on changing the duration, frequency, force, and manner in which the vocal folds contact.

In our practice, children with nodules typically undergo six to eight sessions of voice therapy, once per week. We recommend a combination of approaches tailored to the individual child and their needs. Voice therapy can be characterized as direct or indirect, and a combination of these is often used.

Indirect therapy includes education on vocal health, reduction and replacement of presumed unhealthy voice behaviors, parent education, and implementation of any needed behavioral changes. This is typically addressed at the first therapy session and then briefly discussed in subsequent sessions. Indirect therapy might consist of helping parents tame tantrums, giving children alternatives to yelling, and identifying play noises that may be vocally traumatic. However, simply giving children and parents lists of what not to do is neither practical nor effective. If voice therapy is punitive and children have negative associations with it, they are less likely to be adherent. As we tell toddlers to use their "walking feet" instead of running, we have to give children functional and useful ways of using their voices. Direct therapy teaches more effective and efficient ways of using the voice to achieve a functional sound with lower shearing stresses and impact and without as much perceived effort. In our practice, the majority of time spent in therapy is focused on direct therapy, training healthier voice production. There are a variety of effective ways to address this, but strategies should always be adapted based on knowledge of the anatomy and physiology, impact of the lesion, the child's compensatory behaviors, and the child's motivational factors and developmental level. An overview of voice therapy approaches and ways to adapt to the child's developmental level are provided in section "Overview" of this text. Semi-occluded vocal tract exercises, resonant voice, vocal function exercises, and flow mode phonation, and often a combination of several of these, can be used to address benign vocal fold lesions.

Perioperative Voice Therapy

While voice therapy is the primary treatment modality for most children with nodules, lesions such as cysts and polyps as well as refractory nodules may require surgical excision to achieve optimal voice quality and efficiency. In these cases, we recommend pre- and postoperative voice therapy. Often, preoperative voice therapy is part of the decision-making process in pursuing surgery. If children are able to make the desired changes in voice with therapy alone, surgery is not recommended. However, if they are unable to meet vocal needs despite optimal

adherence to voice therapy, excision may be recommended. When surgery is planned, one preoperative voice therapy session should focus on preparation for voice rest and return to voice use after surgery. The amount of voice rest recommended varies greatly across institutions [42]. A review of wound healing and orthopedic literature indicates that voice rest is preferable to uncontrolled voice use [43], but optimal duration of voice rest is not known. In vitro research has shown that low-amplitude vibrations may have an antiinflammatory effect on healing tissues [44], and Verdolini and colleagues found that resonant voice activities after a vocal loading task resulted in lower biomarkers of inflammation in laryngeal secretions than voice rest or uncontrolled phonation [45]. While it is not possible to generalize this to postoperative conditions, it does suggest that the large amplitude, low impact vibrations associated with resonant voice may play a role in return to vocal health after surgery. Based on the available evidence, but also on the feasibility of voice rest in children, we typically recommend 3 days of complete voice rest followed by gradual return to full voice use over the course of 2 weeks, with use of resonant voice as able, combined with multiple daily practices of semi-occluded vocal tract exercises. Following surgery, the postoperative course of therapy is typically four to eight sessions, once per week, focused on semi-occluded vocal tract exercises, resonant voice, and avoidance of any maladaptive compensatory behaviors that may arise following surgery.

Otolaryngologist Approach

History

Both the patient and parent should be included when eliciting the history, if possible. Critical elements include the nature of dysphonia (e.g., breathy, rough, weak, intermittent, or constant), alleviating and exacerbating factors, and a temporal description. Time of onset and any progression since onset should be noted. Effects of the voice disorder on patient quality of life, functioning in school, home life, and interactions with peers should be evaluated. Presence of any associated breathing or swallowing impairment is questioned. Whether any prior nonsurgical (including observation or voice rest) or surgical therapies have been tried is asked, as well as the results from them.

Potential sources of laryngeal inflammation should be sought, including symptoms of laryngopharyngeal reflux, sleep-disordered breathing, chronic cough, asthma, allergic rhinitis, smoke exposure, and vocal abuse. Extracurricular activities with heavy voice load, including singing, should be noted.

Patients with nodules may report repeated episodes of voice loss, decreased ability to sing softly, breathiness, vocal fatigue, and voice breaks [7]. Patients with polyps can report breathiness, vocal fatigue, diplophonia, and roughness. With cysts, there is often less vocal limitation than may be expected based on lesion size [7]; there can be pitch instability, diplophonia, and compensatory supraglottic hyperfunction. Pseudocysts typically cause minimal impacts on vocal fold vibration, but patients may experience breathiness secondary to impaired glottic closure [46].

Examination

A general head and neck exam is performed. In addition, several aspects are focused on in the patient presenting with dysphonia with concern for underlying benign mass lesion. Any stridor or increased respiratory effort should be noted, though infrequently encountered. The voice should be described, paying attention to presence of breathiness (implying impaired glottic closure), roughness (implying impaired vibration), pressed quality (implying hyperadduction at level of the glottis or supraglottis), projection, and range. The strap muscles are palpated during voice production to evaluate for increased tension, which may indicate primary or secondary muscle tension dysphonia.

Instrumented Assessment

Central to the accurate diagnosis of benign vocal fold mass lesions is careful laryngoscopy and videostroboscopy. In young children, this is typically accomplished with a flexible transnasal endoscope, although children as young as 5 years old may be able to participate in rigid stroboscopy. Image quality is improved with distal chip technology. Older children may tolerate a rigid transoral 70-degree endoscope which will allow for superior image quality. Exams are recorded, which allows for improved visualization of the glottis in the uncooperative patient and provides a baseline for reference during treatment.

Mucosal wave amplitude as assessed by stroboscopy can be helpful in distinguishing polyps from cysts. With a vocal fold cyst, the mucosal wave is often diminished or absent, while it can be preserved in presence of a polyp [47]. Vocal fold pliability is typically preserved with pseudocysts, with resulting minimal effects on vibration [46]. A summary of key laryngoscopic and stroboscopic findings for the four main benign mass lesions is provided in Table 22.1.

Table 22.1 Laryngoscopic and stroboscopic features of benign mass lesions [2, 7, 46, 62]

Lesion	Laryngoscopic features	Stroboscopic features
Nodule	Broad-based, white, opaque prominence at junction of anterior and middle thirds of TVF Typically bilateral, often symmetric Can be unilateral, potentially with contralateral reactive lesion	Hourglass closure Posterior glottic gap Slight decrease in mucosal wave amplitude Vocal fold vibration preserved
Cyst	Broad-based, translucent or yellow fullness at middle third of TVF	Complete closure Decreased or absent mucosal wave on side of cyst Decreased vibration
Pseudocyst	Broad-based, translucent, superficial mass with overlying thin epithelium	May be accompanied by impaired glottic closure Approximately normal mucosal wave Approximately normal vibration
Polyp	Unilateral sessile or pedunculated mass May appear hypervascular (hemorrhagic) with feeding varix	Impaired glottic closure Mucosal wave preserved, with decreased amplitude commensurate with polyp size Phase asymmetry

Differential Diagnosis

Differential diagnosis for benign mass lesions includes nodules, cyst, pseudocyst, polyp, muscle tension dysphonia, vocal fold scar, glottic web, hemangioma, recurrent respiratory papillomatosis, and glottic insufficiency.

Malignant tumors of the larynx in children are very rare but have been reported [48–52]. This includes primary lymphoma, squamous cell carcinoma, choriocarcinoma, and rhabdomyosarcoma. Atypical findings on exam or aggressive clinical course should raise suspicion for these rare entities.

Management

Proper management of benign lesions hinges on making an accurate diagnosis. Unless there is a concern for airway obstruction, initial management can consist of education, vocal hygiene, voice therapy, and management of potential contributors to laryngeal inflammation. This can include treatment of laryngopharyngeal reflux, if present, with twice daily proton pump inhibitors (e.g., omeprazole, 1 mg/kg) and/or histamine antagonists (e.g., ranitidine, 5 mg/kg). Allergic rhinitis can be addressed with nasal saline and topical steroid sprays. Phonotrauma can be addressed with counseling of both child and parent, with emphasis placed on understanding the role of vocal misuse in the pathogenesis of the dysphonia. Hydration should be optimized. Voice therapy with a speech-language pathologist experienced in

the management of pediatric dysphonia should be instituted, with the opportunity to repeat laryngoscopy and visit again with the otolaryngologist if there is inadequate improvement.

For lesions which do not respond to these conservative measures and which continue to cause bothersome symptoms, operative evaluation (Figs. 22.2 and 22.3) with potential phonosurgery can be considered. This is more likely to be required in the case of polyps, cysts, and pseudocysts, which are less likely to respond to conservative measures compared to nodules. If not removed, cysts may rupture which can lead to formation of a sulcus vocalis. However, the presence of recalcitrant lesions which clinically resemble nodules should not be considered a contraindication to surgical exploration with possible intervention.

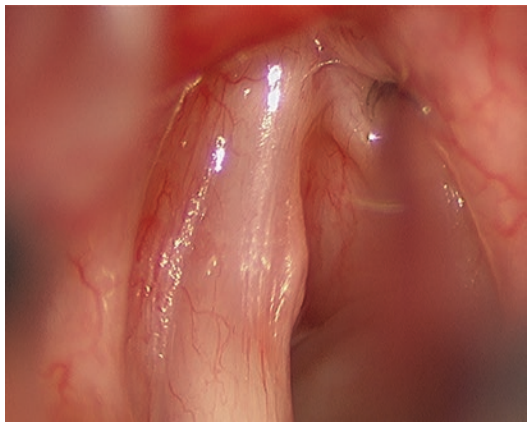

Fig. 22.2 Here, operative evaluation of a left sessile benign lesion also revealed a small microweb. A web can place the effective midmembranous position of the vocal fold where maximal impact stress occurs more posteriorly, resulting in a more posteriorly positioned lesion

Operative Approach: Microflap with Vocal Fold Exploration and Removal of Vocal Fold Benign Mass Lesion

Indications

Surgical evaluation with potential intervention is indicated for benign lesions that are not responsive to nonoperative therapy (voice therapy, proper vocal hygiene, management of contributors to laryngeal inflammation) and affect patient quality of life. Concern for potential airway compromise is also an indication.

Key Aspects of the Consent Process

Risks associated with direct microlaryngoscopy should be discussed, including injury to the lips, gums, tongue, and teeth, as well as the potential

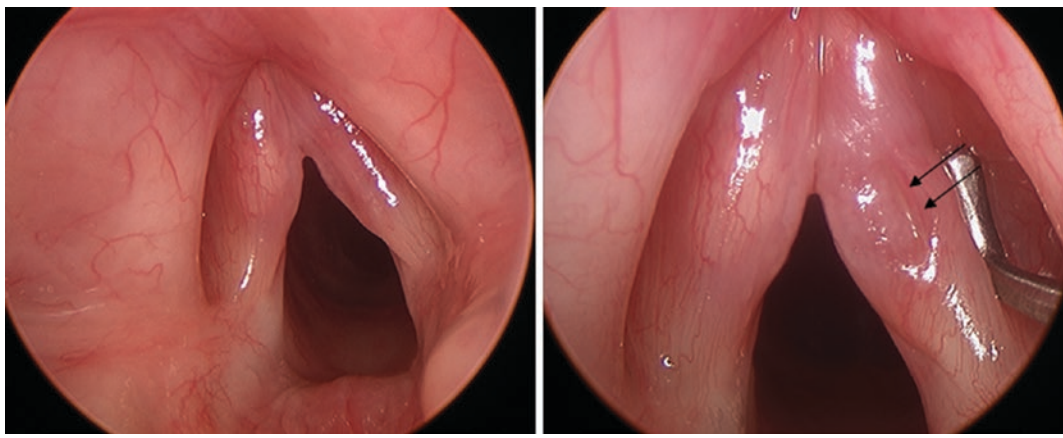

Fig. 22.3 Intraoperative assessment of a presumed lesion should include careful examination of the contralateral true vocal fold. Here, there is a presumed cyst of the left true vocal fold. Palpation of the right true vocal fold demonstrates a sulcus (arrows), which could represent the result of a previously ruptured right cyst

for transient or longer-lasting dysgeusia [53, 54]. Voice or swallow function could worsen or simply fail to improve. Inability to resect an entire lesion, of particular concern in the management of a cyst, may predispose to recurrence. Operating on the airway carries an inherent risk of airway edema which may require overnight observation, temporary placement of an endotracheal tube, or, in rare cases, tracheotomy.

Equipment

A Lindholm laryngoscope is used for exposure. One port is connected to a light cable, and the other can be connected to the end of a 5.5 endotracheal tube to allow for insufflation during the procedure. This is connected to a Lewy arm which is secured on either a Mayo stand or Mustard stand to place the patient into suspension. Rigid 0- and 70-degree telescopes are used for intraoperative exam. An operating microscope is used for the microsurgical portion. A full set of microlaryngeal instruments should be available. This will include a vocal fold retractor to improve visualization, a right-angled probe for intraoperative examination and palpation, sickle knife, left and right scissors, left and right Bouchayer forceps, and microcup forceps. Following dissection, we will typically inject dexamethasone (10 mg/ml) via a Xomed injector into the affected vocal fold due to decrease probability of scar formation.

Steps

1. *Patient positioning.* Depending on patient age and the anesthesiologist's preference, anesthesia can be maintained either by an inhalational anesthetic or by total intravenous anesthesia (TIVA) with spontaneous ventilation and oxygenation via an insufflation technique (typically for patients under age 12) or via orotracheal intubation for older children. The main concern is to provide adequate exposure for the surgical procedure while allowing for the proper plane of anesthesia and ventilation. Working together, the pediatric laryngologist and pediatric anesthesiologist are often very comfortable with insufflation and a nonintubated larynx. This can give maximal exposure and manipulation of the vocal folds during phonosurgery. The patient is positioned supine in the sniffing position, with slight flexion at the neck on the body and extension at the atlantooccipital joint. A mouthguard is used to protect the maxillary dentition. The mouthguard serves more to prevent chips than to prevent fracture or dislodgement of the teeth. Care must be taken not to injury the teeth. For difficult exposures, useful adjuncts include counterpressure to the laryngotracheal complex achieved with silk tape wrapped around the operating table or elevating the head of the bed to increase neck flexion. If silk tape is used, a folded dry 4x4 gauze is placed between the silk tape and patient's skin.
2. *Exposure and suspension.* The tongue and endotracheal tube (if present, as in older children) are pushed to the left, and the Lindholm laryngoscope is advanced into the valleculae. The laryngoscope is lifted to visualize the endolarynx. The Lewy arm is attached and then secured onto the Mayo stand.
3. *Intraoperative examination.* Once patient is in suspension, photodocumentation is obtained with 0- and 70-degree rigid telescopes. A right-angled probe is used to palpate each true vocal fold. The size, consistency, and depth of the lesion are evaluated. The contralateral vocal fold is examined for any reactive change. Each vocal fold is examined to ensure other pathologies, such as sulcus vocalis, are not present. The arytenoids are palpated to ensure normal joint mobility. If the lesion is thought to be a lesion that would be less amenable to surgical excision, the more accurate diagnosis is made, and the procedure may be terminated. Consideration for intralesional steroid injection can be entertained. Using the same injection apparatus for the hydrodissection, dexamethasone (10 mg/ml) can be infiltrated into the lesion.

4. *Hydrodissection with 1% lidocaine with epinephrine.* Local anesthetic is injected into the affected vocal fold in the plane of the superficial lamina propria (just deep to the surface epithelium) to achieve hydrodissection and facilitate elevation of the epithelium off the vocal ligament. Care is taken not to puncture the lesion during this step.
5. *Longitudinal incision with sickle knife.* A sickle knife is used to make a longitudinal incision through the surface epithelium just lateral to the lesion (Fig. 22.4a). The incision should begin posterior to the lesion and extend anterior to it to allow for development of a generous microflap and decrease probability of inadvertent mucosal injury. The distal tip of the knife should be used to tent the surface mucosa to avoid injury to the underlying vocal ligament.
6. *Development of microflap with flap elevator.* An angled spatula is used to elevate the surface epithelium away from the lamina propria and develop the microflap. Start by developing a plane away from any fibrous attachments, and then proceed with dividing the attachments.
7. *Intraoperative vocal fold exploration and determination of lesion type.* The lesion is palpated with the spatula and right-angled probe to determine if it is solid or cystic. In situations where a preoperative diagnosis was uncertain, this can aid in establishing an intraoperative diagnosis.
8. *Develop plane between mass and underlying vocal ligament.* The edge of the flap is retracted gently with a Bouchayer forceps medially (Fig. 22.4b) to apply tension, while a spatula is used to develop a plane between mass and underlying vocal ligament. Care should be taken when handling the flap with the Bouchayer forceps as excess retraction or pressure can cause mucosal injury.
9. *Divide lateral and deep attachments of cyst.* Use the flap elevator to continue dissection lateral and deep to the cyst to free any remaining attachments.
10. *Removal of the mass.*
    (a) *Removal of cyst with preservation of lamina propria.* Dissection is completed around the mass. Care is taken not to violate the cyst wall. If the cyst wall is violated, a microcup forceps can be used to ensure removal of all wall remnants.
    (b) *Removal of other lesions with preservation of lamina propria.* If during exploration of the vocal fold, the lesion is determined not be a cyst but thickening of the basement membrane consistent with a vocal nodule, the thickening of the nodule can be removed to give a smooth, more pliable, vocal fold. Some have been able to remove the nodule matrix manually with cup forceps and scissors. Others have used the smallest laryngeal skimmer with reduced suction and slower speed to plane the nodule

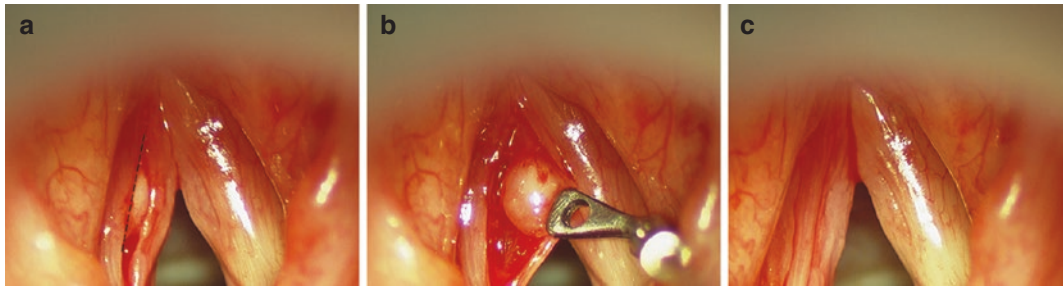

Fig. 22.4 Operative removal of a left vocal fold cyst. (**a**) A longitudinal incision was made along the dotted line with a sickle knife, at the lateral aspect of the lesion. A plane is developed between the mass and underlying vocal ligament. (**b**) The microflap is gently retracted medially with a Bouchayer forcep to expose the mass and facilitate dissection around it. (**c**) After removal of the mass, the flap is draped back. Any excess mucosa is trimmed

matrix off of the raised flap. Care must be taken to avoid amputating the microflap and removing the mucosa. Pseudocysts are removed in a similar manner but are easier to treat as the matrix is not adherent to the basement membrane.

(c) *Removal of polyp with preservation of maximal amount of lamina propria.* The removal of a polyp can be approached in a very similar manner. The polyp may also simply be excised, though, as the redundant mucosa will coapt and not leave a defect. The apex of the polyp may be grasped by atraumatic forceps and placed on tension, medializing the submucosal matrix. Care is taken to assess the amount of mucosa to be excised to allow for complete excision of the lesion while affording approximation and closure of the defect.

11. *Hemostasis.* A cotton ball or ½″ by ½″ pledget soaked in epinephrine (1:1000) is applied for hemostasis.
12. *Replacement of mucosal flap.* The microflap is replaced over the wound (Fig. 22.4c).
13. *Injection of steroid.* Dexamethasone (10 mg/ml) is injected via a Xomed needle into the affected vocal fold to decrease probability of scar formation. A laryngotracheal anesthetic (2% lidocaine) is applied. The airway is suctioned. Patient is then taken out of suspension and returned to the anesthesia team for emergence and extubation (if previously intubated).

Postoperative Management and Follow-Up

Voice rest for 3 days followed by gradual return to full voice use over the course of 2 weeks is recommended for those children who can adhere to it. This can include avoidance of yelling or talking excessively. A 1 week visit with the speech-language pathologist is coordinated to affirm vocal hygiene concepts and initiate postoperative voice therapy, which includes semi-occluded vocal tract exercises and resonant voice therapy. The patient is seen again in 6 weeks by both the speech-language pathologist and otolaryngologist for repeat formal voice assessment and flexible laryngoscopy. Other adjuvant medical therapies are based on the symptoms and suspension of other inflammatory diseases. If reflux is presumed or proven, it should be tightly managed during this initial healing period.

Emerging and Evolving Concepts

Laryngeal Ultrasound

While the standard for office-based laryngeal assessment is flexible transnasal laryngoscopy, the exam is often poorly tolerated in children, and there are patients in whom visualization can be difficult, particularly those with developmental delay. Laryngeal ultrasound has been described for evaluation of subglottic diameter and vocal fold paralysis [55, 56]. Recently, it has also been applied for evaluation of pediatric benign vocal fold lesions [3, 18]. Bisetti et al. used ultrasound to evaluate 16 children, 14 of whom had benign mass lesions [18]. They demonstrated different ultrasonographic appearance for nodules versus polyp versus papilloma. Ongkasuwan et al. used ultrasound to differentiate 23 children with nodules compared to 23 without and reported a sensitivity of 100% with specificity of 87% [3]. Ultrasound has demonstrated potential, and further refinements to image acquisition and analysis may improve ability to distinguish among lesions and characterize lesion size and depth.

Laryngeal Optical Coherence Tomography

Optical coherence tomography (OCT), first presented in 1991 [57], performs high-resolution, cross-sectional imaging of internal microstructure [58]. OCT is analogous to ultrasound, except that light is used instead of sound. A beam of light is directed onto a structure of interest, and the differential backreflection and backscatter

from those tissues result in creation of an image [59]. Images can be obtained in real time. As a light source is used instead of sound, the resolution is superior to ultrasound. This technology has been used in the ophthalmology arena but has recently been applied to the larynx as well [60, 61]. Benboujja used intraoperative OCT with a probe placed in the endolarynx to describe the imaging characteristics of normal pediatric vocal folds as well as those with nodules, cyst, and papilloma [60]. This approach may hold promise for a method of intraoperative lesion assessment which does not require violating the true vocal fold mucosa.

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Laryngopharyngeal Reflux

23

Lauren Sowa, Holly Schmidt, and Mark E. Gerber

Introduction

In the pediatric population, reflux of gastric contents into and above the esophagus impacts different age groups in a variety of ways. It has been estimated that up to 10% of patient visits to an otolaryngologist are for reflux-related issues [1]. Vague symptoms have led to difficulty in research among the involved specialties including gastroenterology, otolaryngology, and speech pathology. This chapter aims to clarify definitions of laryngopharyngeal reflux and its contribution to other aerodigestive pathologies from the perspective of both the otolaryngologist and speech pathologist.

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Definitions

Gastroesophageal reflex (GER) involves reflux of gastric contents up through the lower esophageal sphincter (LES). When this refluxate travels farther into the laryngopharynx, it is termed laryngopharyngeal reflux (LPR), also known as extraesophageal reflux (EER). GER progresses to gastroesophageal reflux disease (GERD) when reflux leads to complications such as esophagitis or esophageal strictures. When symptomatic reflux is less severe, determining GER versus GERD remains controversial [2]. In addition, controversy exists as to whether LPR is truly an extension of GER or a distinct entity given that symptoms, exam findings, and treatments tend to differ [1, 3]. While most older patients with GER will have positional esophagitis, dyspepsia, or heartburn, those with LPR are typically considered "upright" or "daytime refluxers" with symptoms of hoarseness, globus pharyngeus, dysphagia, cough, or sore throat [4]. To be defined as GERD, these symptoms are combined with regurgitation and emesis, which typically interfere with daily function [2]. This will be further broken down into symptoms in infants versus children later in this chapter.

It is difficult to estimate an exact prevalence of LPR, but it is likely that around 20% of infants and children suffer from some kind of reflux disease, similar to adults. In infants, this distinction is particularly difficult because of the universal

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presence of reflux in this age group. From age 6 months to 2 years, there is a decline in the incidence of what is called "physiologic reflux," likely due to maturation of the lower esophageal sphincter with age and the transition from a liquid to a solid diet [2]. A larger proportion of infants with comorbid conditions such as prematurity and neurologic dysfunction suffer from reflux disease.

Pathophysiology and Presentation

The pathophysiology of laryngopharyngeal reflux causing airway symptoms remains under debate. The prevailing theory is that refluxate through a weak upper esophageal sphincter causes damage to the laryngeal mucosa. It has been shown that gastric acid exposure as little as 1 drop per day for 8 days can transform a minor tracheal injury into full-blown subglottic stenosis in animal models [5]. This is thought to be due to the lack of defense mechanisms in airway mucosa and results in findings such as posterior laryngitis and interarytenoid edema, vocal fold nodules, or contact granulomas.

Koufman et al. used canine models to demonstrate the effect of intermittent reflux on laryngeal mucosa with existing damage. This study showed that gastric pepsin, as opposed to hydrochloric acid, was the more injurious agent in reflux contents [6]. Johnston et al. explored the theory that gastric pepsin causes damage to the laryngeal mucosa in both acidic and non-acidic reflux. They determined that pepsin, which is a well-described biomarker of gastric contents, is enzymatically inactive at neutral pH (below 8.0) but is taken up by cells in the larynx and hypopharynx via endocytosis [7]. Once it is intracellular, it is reactivated and can continue to cause damage by activating a pro-inflammatory cytokine cascade [8]. This theory of gastric secretions causing mucosal damage was furthered by Farhath et al. who studied the concentration of pepsin in the tracheal secretions of preterm infants. This group found that infants with a higher concentration of pepsin were at a greater risk for more severe bronchopulmonary dysplasia (BPD) [9]. Other more recent studies, however, have not found an association between the presence of pepsin in respiratory secretions and extraesophageal reflux symptoms and risk of hospitalization [10, 11].

Infants suffering from LPR can present with a conglomeration of symptoms including failure to thrive, feeding intolerance, regurgitation, recurrent croup, stridor, aspiration, and cough. Recognizing the contribution of LPR to these symptoms can be difficult to glean from parental reports, as a certain amount of reflux is considered normal in young infants. Older children are more likely to present with symptoms such as cough, wheezing, hoarseness, globus sensation, recurrent upper respiratory infections, and sore throat [1, 3]. Studies have speculated that recurrent pneumonias are more common in children with a history of LPR, especially when there is also a clinical history of GERD. The thought process behind this association is that chronic reflux bathing the laryngeal mucosa alters sensation. The "laryngeal adductor reflex" is an airway protective mechanism by which the glottis closes and respiration ceases during swallowing in response to chemical or mechanical stimuli to the supraglottic mucosa. If reflux is not recognized and managed, reduced laryngeal adductor reflex responses due to chronic exposure to refluxate make infants and children prone to microaspirations and development of lower airway disease if not recognized [12, 13]. It is also possible for intermittent reflux to create a hyperactive laryngeal adductor reflux sometimes referred to as reflux apnea, which affects approximately 1% of infants. Reflux apnea can occur due to airway closure or laryngospasm in response to acidic or non-acidic stimulation, leading to increased respiratory effort mimicking obstructive sleep apnea [14]. Additionally, patients with swallowing disorders may be further impacted by LPR. Normal mechanisms to clear acid include salivation, swallowing, and peristalsis each of which can be significantly impaired. Dysfunction that is frequently seen in children with cerebral palsy and other types of oral sensorimotor dysfunction can set up a vicious cycle of reflux. It can start with anterior drooling leading to loss

of salivary volume that is further reduced by low frequency of swallowing and made worse still by the presence of abnormal peristalsis [15].

Speech Pathologist Approach

Comparison of Infant and Adult Anatomy for Swallowing

There are significant differences in the oropharyngeal anatomy across newborns, young children, and adults. During the early months of life, there are many changes that occur in the anatomy of the oral, pharyngeal, and laryngeal areas. At birth there is positional stability which provides the close proximity of various structures and the infant's large amount of subcutaneous fat. As the infant matures, greater postural stability is provided. Structures move farther apart and are supported by increased muscle control. These differences predominate until the age of 3–4 months (Table 23.1) [16].

Table 23.1 Comparison of factors that may contribute to LPR in infants, children, and adults

Premature infants	Infants
Weak LES	Weak LES
Positional (supine or side-lying)	Positional (supine or side-lying)
Liquid diet	Liquid diet
Related comorbid conditions (e.g., decreased overall tone, neurologic)	Related comorbid conditions (e.g., decreased overall tone, neurologic)
Constipation	Constipation
Presence of nasogastric tube passing through lower esophageal sphincter	Surgical alternations (e.g., TEF repair)
Immature esophageal motility	
Potential delayed emptying [16]	
Children	**Adults**
Lifestyle	Lifestyle
Comorbid conditions (e.g., Down syndrome, neurological disorders)	Stricture
	Reduced esophageal motility
Constipation	
Surgical alterations	

LES lower esophageal sphincter, *TEF* tracheoesophageal fistula

The oral cavity of an infant is small with the tongue filling nearly the entire oral cavity. The infant's cheeks also have sucking pads which assist with stability in the oral system. Newborns tend to be obligate nose breathers due to the tongue filling the mouth as well as the soft palate and epiglottis touching. Mouth breathing requires more effort to open the mouth and create tone to separate the soft palate and tongue to breathe.

The larynx rides very high in the infant neck and is in much closer proximity to the base of the tongue than in the adult. Because it is directly under the tongue and epiglottis, the larynx is more easily protected when it elevates during swallowing, obviating need for coordinated glottic closure. Elongation of the neck and pharynx during infancy and childhood causes descent of the larynx away from the palate and tongue base, leading to less anatomical protection during the swallow.

Potential Impact of Reflux on Swallowing Function

Sensory impact on swallowing function is a critical component. When normal sensory input is altered, swallowing function may be altered as well. Full-term infants are born with a reflex to eat. This reflex turns volitional between 4 and 12 weeks of life. If there is reflux or aspiration during this time, it is possible that an infant may develop an adverse reaction and begin to refuse oral intake. Regurgitation of acid and other stomach contents into the pharynx, larynx, and oral cavity impacts the normal sensory experience, disrupting the oral and pharyngeal phases of swallowing. The possible impact on the oral phase is oral hypersensitivity, which decreases an infant's interest in eating and impairs bolus preparation. This can be a lasting effect that continues into childhood with repeated insults. The impact on the pharyngeal phase is one from a safety perspective. The pharynx and larynx have a rich supply of chemoreceptors, baroreceptors, and temperature receptors that are highly sensitive to specific kinds of sensory input. When the receptors are overwhelmed due to recurrent/

chronic exposure to gastric contents, responsivity can be dampened, leading to increased risk of aspiration. Silent aspiration is defined as passage of swallowed or regurgitated material into the airway below the level of the true vocal folds without leading to a cough or choking response. Infants are more likely to silently aspirate, leading to a delay in the diagnosis.

Diagnosis

Otolaryngologist Perspective

The most important diagnostic tool for identifying LPR is the clinical history. This can be difficult considering that infants and toddlers cannot verbalize their symptoms. Parents and pediatricians must be astute in their recognition of the aforementioned symptoms that can present differently in infants and children to help determine when it is appropriate to refer to an otolaryngologist for further evaluation and management. Based on the most recent clinical practice guidelines, physicians should also be looking for warning signs that might suggest a pathology aside from GERD as a part of the history. These include weight loss, lethargy, persistent emesis, seizures, chronic diarrhea, or rectal bleeding [2, 17, 18]. These symptoms should prompt further testing to rule out other serious disease processes including gastrointestinal, neurologic, metabolic, toxic, and cardiac disorders.

Fiber-optic laryngoscopic examination can be helpful for both infants and children as an initial diagnostic evaluation since it is brief and minimally invasive. Though cooperation may be a limiting factor, it provides a brief dynamic view of the larynx that can help identify findings consistent with reflux changes or other pathologies such as laryngomalacia or vocal fold nodules [14]. Findings can be subtle, but those most associated with reflux include posterior glottic edema, lingual tonsillar hypertrophy, arytenoid edema, vocal fold edema or nodules, and hypopharyngeal cobblestoning (Figs. 23.1, 23.2, and 23.3) [3, 19]. In adults, the Reflux Finding Score (RFS) can be used to interpret laryngeal findings

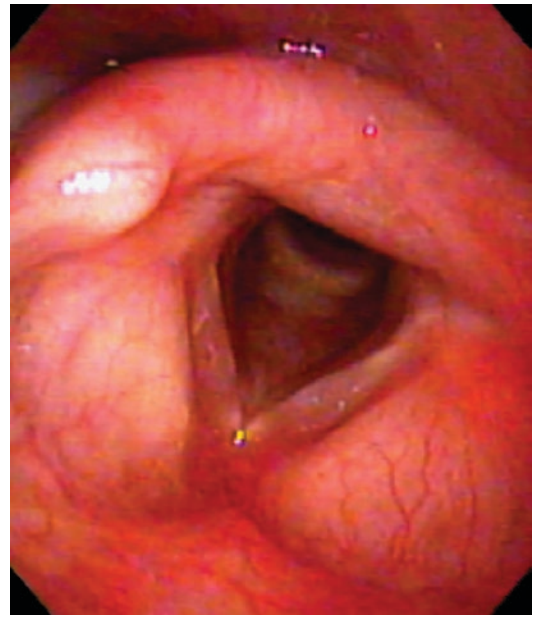

Fig. 23.1 Mild left greater than right true vocal fold with bilateral pseudo-sulcus secondary to infraglottic edema. There is endolaryngeal erythema, most notable at the petiole and anterior commissure. (Courtesy of Dr. J. Scott McMurray)

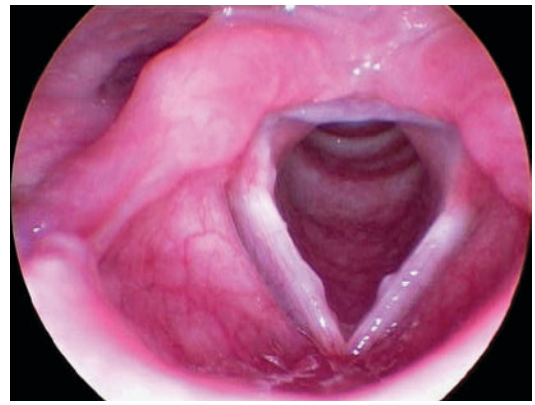

Fig. 23.2 Bilateral vocal fold nodules at the junction of anterior and middle thirds as well as microweb, erythema at the petiole, false vocal folds, and interarytenoid pachydermia. (Courtesy of Dr. J. Scott McMurray)

suggestive of GERD [20, 21]. This was attempted to be extrapolated to children (the Reflux Finding Score for Infants or RFS-I) in a prospective cohort study out of Boston Children's Hospital which determined that laryngeal appearance using the RFS did not correlate with results of

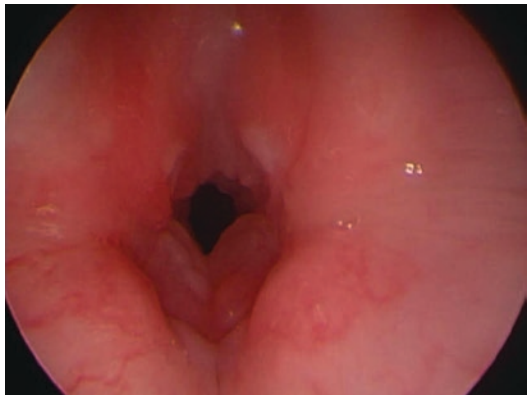

Fig. 23.3 Severe changes related to laryngopharyngeal reflux including diffuse supraglottic and glottic edema, with prominent infraglottic edema as well as extensive posterior glottic and interarytenoid pachydermia and edema. (Courtesy of Dr. J. Scott McMurray)

multichannel pH probe monitoring, suggesting that such scoring systems have a low positive predictive value for detecting reflux in pediatric populations [22]. In response to this study, Ida et al. postulated that these results raise the issue of the "reactive larynx" and non-reflux-related laryngeal inflammation as a barrier to airway surgery [23]. Another group assessed the reliability of the RFS-I in flexible versus rigid laryngoscopy noting no difference between the two diagnostic procedures as well as concern regarding limited reliability of the grading system (only moderate in both cases) [24]. Posterior cricoid biopsy has been shown to have a weak association with reflux symptoms and pH probe findings, but more specific randomized treatment-driven studies are needed to further explore this notion [25]. Two studies attempted to correlate laryngeal findings on direct laryngoscopy with bronchoscopy in an attempt to establish a more consistent grading system. These studies were either retrospective [5] or prospective unrandomized [26]. In both studies, posterior laryngeal edema had the strongest correlation with reflux. Other findings were not consistent enough to establish a definitive relationship [5, 26].

Barium esophagrams, which were used in the past, have been found to have a relatively low sensitivity and specificity for evaluating LPR/GER, with the added disadvantage of radiation exposure [27]. No true gold standard currently exists for the diagnosis of LPR in children. In most cases, four to five events of pharyngeal reflux can be considered normal in infant populations, while one or more episodes in older children and adults are considered abnormal [28]. A common study in pediatric and adult patients to diagnose GERD is 24-h pH probe monitoring, which can involve either a single distal probe or dual probes in the distal and proximal esophagus. In adults, the use of 24-h dual-probe pH monitoring has been helpful in demonstrating pharyngeal reflux at acidic pH (defined as pH <4.0) in the diagnosis of LPR. In children, however, this is difficult to assess because of the common presence of non-acidic reflux into the proximal esophagus and pharynx. Rabinowitz et al. found that distal pH probe monitoring alone gives a high rate of false negatives in pediatric patients with extraesophageal symptoms of GER, suggesting that non-acidic reflux plays an important role in generating these symptoms [29]. Another study reviewed 68 pediatric patients with GER that underwent distal probe pH monitoring and anti-reflux therapy. In patients with extraesophageal symptoms, they found that distal pH studies were not predictive of positive response to anti-reflux therapy and thus should be avoided in this subset of patients [30]. Thus, single distal pH probe monitoring is not considered an adequate study for the diagnosis of LPR.

Dual-probe pH testing involves placing a measuring probe just above the lower esophageal sphincter as well as just below or just above the upper esophageal sphincter in the pharynx. Refluxate at pH <4 is recorded. However, in infants and children, many of these reflux events will occur in physiologic pH range [5–7] and thus not be detected by pH probes alone [31]. Using a pharyngeal probe, Little et al. did show that 76 out of 168 pediatric patients (aged 1 day to 16 years) had pharyngeal reflux as measured by the pharyngeal probe in spite of many having normal levels of esophageal acid exposure as measured by the esophageal probe. They estimated that in the absence of a pharyngeal probe, 46% of participants would have been underdiagnosed [32]. Other studies have found that dual-probe pH

testing was not easily reproducible in children as compared to adults and is therefore not a reliable diagnostic tool on its own [33, 34].

Multiple intraluminal electrical impedance technique (IMP or MII) is a more recent diagnostic tool that is being used in conjunction with pH probe monitoring for the diagnosis of GER. The principle behind this testing is to measure opposition to the flow of current between electrodes along an esophageal probe. This allows movement of refluxate and/or food boluses at multiple heights along the esophagus irrespective of pH [35]. Several studies have shown the superiority of this method as compared to dual pH monitoring alone [36–39]. In a small German study of 50 infants with presumed GER, IMP identified 1866 episodes of retrograde reflux, while pH probe only identified 282 (15%) of these to be acidic with pH <4.0 [36]. The same group looked specifically at respiratory episodes associated with reflux such as apnea, aspiration, and abnormal chest wall movement in the setting of combined IMP and pH monitoring. In 22 patients, 364 episodes of reflux were recorded with IMP, and only 12% of these were found be acidic. Of these 364 episodes, 312 (86%) were associated with breathing abnormalities, 128 of which were associated with an oxygen desaturation of >10%. They also found that longer episodes of reflux greater than 30 s were more likely to be associated with apnea [37]. Both studies demonstrate the importance of impedance monitoring in the evaluation of reflux, particularly in infants when a higher percentage of refluxate will be non-acidic. In older children, combined pH-MII monitoring was shown to be significantly better at detecting cough in the setting of reflux as compared to patient and parental reporting [40].

Despite the clear advantages of pH-MII as compared to pH monitoring alone, there are still widely recognized limitations of impedance monitoring, particularly in light of the fact that there is no control group from which to establish a threshold of what constitutes a "normal" amount of non-acidic reflux. With this in mind, pH-MII testing is an option to consider when trying to correlate troublesome symptoms of acidic and non-acidic reflux in infants and children [2] and prior to airway reconstructive surgery.

In adults, brief trials of anti-reflux medications have been successfully used as a diagnostic tool, with the thought that if symptoms improve with treatment, it is likely that reflux is playing a role. In infants and children, however, this has not been as well studied. The use of proton-pump inhibitors (PPI) for the treatment of asthma and cough has been looked at with no good correlation found. One study explored symptoms of asthma noting that during a trial of a PPI at 4–8 weeks, there were no noted benefits from use of the medication [41]. In infants, a similar study looked at lansoprazole treatment noting no significant difference in extraesophageal symptoms after a 2-week trial [42]. Findings from these studies and concerns about side effects of PPI use have reduced the frequency of diagnostic PPI trials in children.

Speech Pathologist Approach

Workup

Clinical histories can differ among reporters due to varying perceptions of the child's skills and abilities [43], and inconsistencies should be further explored. For example, food refusal may be interpreted as laziness or lack of hunger when, in fact, the child is saying "no" because of the discomfort which might be related to silent aspiration, gastroesophageal reflux, or esophagitis/gastritis [44]. Parents should be asked open-ended questions to describe feeding behaviors.

A speech pathologist's intake includes pregnancy and birth history, general development, and medical background (including prior testing, respiratory issues, and hospitalizations). The feeding history includes a "typical" day in the child's life. Often the 24-h period prior to the appointment is a good example of the child's feeding habits and is easy for the parent to recall. Details surrounding positioning, volume, texture, utensils, and intervals between meals are also collected. If the child is tube-fed, the feeding history also includes looking at feeding volume and any behavioral responses to changes. The timing of onset and duration of feeding problems can help

identify signs and symptoms of gastroesophageal reflux. Questions should include asking about possible pain/discomfort during feeding; crying, gagging, or coughing/choking while eating; refusal to eat/chew/swallow; lack of hunger awareness; and bottle feeding only while falling asleep. If there is recurrent vomiting, the timing and frequency of the emesis and whether or not it is forceful/projectile are important to ask. In addition, questions about bowel movements and/or constipation are helpful. A general history of vocal quality is important for consideration of both vocal fold impairment and irritation. This should include listening to the child's voice, assessing for hoarseness/raspiness, and asking the parent(s) if they are at their baseline in terms of vocal quality.

Clinical swallowing evaluation, videofluoroscopic swallowing study (VFSS), and flexible endoscopic evaluation of swallowing (FEES) can all be useful in evaluating swallowing function in infants and children with reflux. These evaluations are described in detail in previous chapters.

Disease Processes Impacted by LPR

LPR has been linked to several pathologies that predominantly affect the pediatric population, namely, otitis media, sinusitis, laryngitis, laryngospasm, airway stenosis, and lower airway pathology [1]. A significant amount of research has been done to investigate the impact of LPR on disease processes such as laryngomalacia, subglottic stenosis, and chronic cough.

Laryngomalacia is the most common cause of stridor in infants [45]. It is defined by the prolapse of flaccid supraglottic structures inward during inspiration resulting in upper airway obstruction [46]. The presentation can be immediate or delayed for several weeks after birth and often resolves spontaneously by age 12 months. Severe or untreated laryngomalacia can lead to such complications as obstructive sleep apnea, cor pulmonale, failure to thrive, cyanosis, or death. Currently, the treatments for this condition range from conservative lifestyle modifications (e.g., position changes, feeding modifications) to surgical intervention (supraglottoplasty) for refractory cases [45]. In the neonatal and infant population, studies estimate the incidence of reflux associated with laryngomalacia to be between 23% and 80% [47–50]. Matthews et al. aimed to establish this association with a small study of 24 infants diagnosed with laryngomalacia via flexible endoscopy. Each of these infants underwent 24-h pH monitoring with distal esophageal and pharyngeal probes. They found that patients had an average of over 15 episodes of pharyngeal reflux, with 100% of them demonstrating pharyngeal reflux [45]. Though promising, this study was limited by its small sample size and lack of control group. In a larger literature review including 1295 neonates, a correlation between severity of laryngomalacia and prevalence of reflux was present in several studies. When compared to other children with other respiratory disorders, there was no significant difference in reflux between the two groups [46]. Furthermore, six studies included in their evaluation assessed anti-reflux treatment and improvement in symptoms of laryngomalacia, but results were weak and rather inconsistent. In another small study of infants with laryngomalacia, Thompson found that treatment with anti-reflux therapy was associated with a reduction in parent-reported symptoms such as coughing, choking, and regurgitation, particularly in patients diagnosed with mild disease. However, without a control group, it is hard to say if the improvement was due to treatment versus the natural resolution [51].

Laryngeal stenosis can be mild or can present with significant symptoms and require endoscopic and/or open surgical intervention. It can occur due to anatomic abnormalities, instrumentation, or inflammatory processes. Extraesophageal reflux has long been suspected as being a significant contributing factor in many if not most patients with laryngeal stenosis, with the literature citing a prevalence of around 60% [26, 52–56]. In canine models, Little et al. demonstrated that mucosal tracheal lesions bathed with gastric acid developed more rapid and severe stenosis as compared to controls which healed normally [57]. Because of this concern, clinicians have focused on identifying and treating airway reflux prior to surgical treatment of

laryngeal stenosis. In a retrospective review of 25 children with SGS, Halstead examined the perioperative workup and treatment of reflux and their respective outcomes [52]. These children all had subglottic stenosis (SGS) confirmed by flexible laryngoscopy, underwent 24-h pH probe exams, and were treated with proton-pump inhibitors and a promotility agent (cisapride) prior to surgical intervention. Of the 25 children examined, 9 were treated with medication alone, and 16 underwent endoscopic surgery with laser excision of stenosis. Only 1 patient out of 16 failed endoscopic repair and required tracheostomy. Compared to this institution's population prior to the study, the aggressive treatment of reflux decreased the rate of failure of endoscopic surgery from 1 in 5.7 to 1 in 25.

Chronic cough is a diagnosis that continues to puzzle pediatricians, allergists, pulmonologists, and otolaryngologists alike. In the workup of chronic cough (defined as a cough lasting more than 4 weeks), asthma, allergy, and reflux are just a few of the possible etiologies that need to be considered [58]. In the current literature, though, chronic cough has only been formally associated with GER in 3–8% of cohort studies [59]. In an attempt to further characterize this, Chang et al. investigated airway neutrophilia in children with suspected GERD [60]. The study included 150 children undergoing EGD for typical GI complaints, and children over the age of 6 also underwent spirometry. Patients were divided into two groups based on clinical history of cough versus no cough, and both groups were equally likely to have evidence of reflux esophagitis. Furthermore, there was no difference in cellular profile (i.e., number of neutrophils), suggesting that the coexistence of symptoms did not imply causation. With a lack of reliable studies demonstrating a definitive causative relationship between the two, reflux remains more of a diagnosis of exclusion in the workup of chronic cough.

Treatment

Because of the invasive nature and limited normative control data for diagnostic tests, many practitioners utilize empiric treatment with proton-pump inhibitors or histamine (H2) blockers even prior to pursuing testing. This is particularly true of patients that have positive findings on an in-office laryngoscopy such as vocal cord nodules or laryngomalacia [3].

Conservative measures, namely, lifestyle modifications related to feeding frequency, positioning, and consistency, are a reasonable initial treatment strategy (Table 23.2). In a literature review of infants with reflux-related airway symptoms, position changes were found to reduce reflux up to 80%, and thickening of feedings decreased regurgitation by 65% [61, 62]. In older children, conservative measures

Table 23.2 Treatment in the infant/child population includes the following options

Population	Treatment strategy	Purpose/results
Infant	Position modification (left side-lying, prone when awake[a], elevated left side-lying, semi upright, 30-degree elevation of bed)	Gravity assist, improved motility
Infant/child	Frequency/volume of feedings	Lower volume feedings and shorter feedings reduced acidic GER [16]
Infant/child	Thickening in absence of aspiration	Inconsistent conclusions in the literature with necrotizing enterocolitis documented in preterm population [69, 70]
Infant/child	Oral motor therapy	Education, advancing feedings, address oral sensory issues
Infant/child	Occupational therapy/physical therapy	Sensory concerns/positioning strategies, head/neck preference and/or alignment
Child	Voice therapy	If dysphonia is present, address behavioral contribution and train efficient, healthy voice use

[a]Prone positioning in infants is only to be used when they are awake, not sleeping

include dietary education with avoidance of triggers and limitation of nocturnal eating, similar to those for adults.

For patients with symptoms refractory to conservative measures, medical treatment is typically the next step. The recommended first-line therapy is a proton-pump inhibitor (PPI), of which lansoprazole and omeprazole are the only drugs approved by the FDA for use in infants [3]. Duration of treatment is typically 12 weeks, though some say that symptoms of LPR may take up to 6 months to resolve with medical treatment [4]. Histamine (H2) blockers, such as ranitidine, can be used up front as a primary course of therapy or as a second-line additional treatment to supplement a PPI or facilitate weaning from a PPI. Prokinetic agents, such as metoclopramide and cisapride, were historically used in the neonatal population but have since fallen out of favor because of dangerous side effects, including extrapyramidal symptoms [14]. Studies looking at management of LPR in children are very limited. In a review of adult studies of LPR treated with PPI, several double-blinded placebo-controlled trials exist totaling 276 patients. All of these used twice-a-day therapy (high dose), but only one study showed a statistically significant difference in symptoms of LPR as compared to placebo, namely, hoarseness and throat-clearing [63]. Another study of 35 patients treated with a PPI or placebo for 12 weeks and had an additional 4-week follow-up period. Both groups showed improvement based on a validated reflux grading scale and pH probe test, but the group that received the PPI had significant rebound symptoms after cessation of therapy compared to placebo [64]. No such prospective studies currently exist in children. In a retrospective review of infants and toddlers with LPR and dysphagia, Suskind et al. found that, in conjunction with dietary and positional modifications dictated by a speech pathologist, treatment with anti-reflux medications (79%) or a Nissen fundoplication (21%) resulted in a decrease in aspiration on FEES or VFSS from 82% to 14% [12]. It is difficult to ascertain whether this improvement was due treatment or to normal growth and spontaneous resolution of these infants. Given their ages and time course, the authors suggest that treatment may have improved the laryngeal sensation deficits caused by reflux.

Ultimately, the decision to pursue medical treatment for reflux in infants and children must be a thoughtful one, as studies have shown that treatment with a PPI can actually worsen or cause respiratory and gastrointestinal infections in children with underlying respiratory illnesses [41, 65]. Furthermore, the impact of overtreating has public health implications. It has been estimated that the cost of medications and diagnostic testing related to reflux in adults alone is approximately 50 billion dollars per year [66].

In patients with significant pathologic reflux or failure of medical therapy, surgical intervention may be warranted. Nissen fundoplication, which aims to increase the strength of the lower esophageal sphincter, is the procedure of choice [67]. To examine patients with LPR that had undergone Nissen fundoplication, Iqbal et al. performed a retrospective review which determined that 25 out of 40 patients reported a significant quality of life improvement in the 1 year following surgery, which suggests that this is a helpful procedure in appropriate patients for LPR in addition to GERD [68].

The Speech Pathologist Perspective

The speech pathologist's role in reflux treatment most often includes teaching and counseling related to positioning, feedings, and exercises (Table 23.2). When an infant or child presents with a chronic hoarse voice, it may be due to LPR. If there are no improvements to the vocal quality with medical management, further assessments should be considered. LPR could be compounded by overall sensory issues indicating a need for an occupational therapy evaluation. In addition, if the child's voice is not improving, a formal instrumented voice evaluation is indicated in order to evaluate for the cause of the dysphonia and plan treatment. While dysphonia can occur with reflux, it should not be automatically assumed that this is the cause of the voice disorder, overlooking other potential etiologies.

Conclusion

In pediatric populations, the recognition of LPR as an entity that can be pathologic both independently and in conjunction with GERD is essential to accurate diagnosis and thoughtful treatment planning. The age and comorbidities of the patient in question play a significant role in the evaluation and management. It has been well demonstrated that both conservative lifestyle and medical therapies can be helpful in improving some of the symptoms, and in more severe cases, surgical management with fundoplication may be worthwhile. However, prospective, placebo-controlled trials in children are lacking. Above all else, the current literature demonstrates the importance of obtaining a detailed clinical history so as not to underestimate the impact of LPR on aerodigestive function.

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Muscle Tension Dysphonia and Puberphonia

24

Marshall E. Smith and Daniel R. Houtz

Overview

The voice has a significant role in oral communication and is important as an expression of our health, emotion, gender, and age. It forms part of one's individual identity and personality. In children, this organ is developing in physical structure along with the rest of the speech mechanism. At the same time, the neurocognitive, behavioral growth and maturation of the child occurs. The larynx has highly developed neural connections, so it is not surprising that the voice is sensitive to neural input and control. This includes input derived from emotional centers in the brain. The larynx has been labeled "the valve of emotion" [1]. It is highly responsive to emotional state and stress at all ages.

Studies of voice disorders in children have suggested that the majority of dysphonias are due to vocal overuse and misuse. The common manifestation of these is vocal nodules [2, 3]. This disorder may be viewed as "functional" because underlying dysfunction is the cause of tissue trauma that creates the nodules. Dysphonias also arise in children with no identifiable structural or physical pathologic changes to the vocal folds. In this chapter, we define a functional voice disorder as a voice disturbance that occurs in the absence of structural or neurologic laryngeal pathology. In adult voice clinics, these disorders may account for up to 40% of cases [4]. In pediatric patients, functional voice disorders occur less frequently. A series of 427 children referred to a tertiary pediatric voice disorders clinic reported that 7% of cases had a functional etiology [2]. In another recent series, only 4% of 136 children with voice disorder were labeled as functional or neurogenic [3]. In this review, the major manifestations of functional voice disorders in children are discussed. These include muscle tension dysphonia (MTD) and aphonia and puberphonia or mutational falsetto.

MTD has gained common usage as a diagnostic label for functional dysphonia thought to be due to dysregulated or imbalanced laryngeal and paralaryngeal activity [4]. A variety of glottic and supraglottic patterns of laryngeal closure have been described [5, 6]. Their diagnostic utility has come into question because these closure patterns are not unique to MTD, and do not reliably distinguish them from normal speakers, or other voice disorders [7].

The predominant auditory-perceptual feature of MTD is a strained voice quality, disordered pitch (usually pitch elevation), and reduced

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loudness. These features may lead to diagnostic confusion with spasmodic dysphonia [8]. Periods of aphonia may also be present. These may be intermittent or persistent. Another feature that may be present in MTD is that periods of normal voice may occur in between the dysphonic intervals. On physical examination, exquisite tenderness to palpation in the thyrohyoid space and narrowing of the thyrohyoid space are frequently identified.

There have been a variety of explanations offered for MTD, including technical misuse due to excessive vocal demands, altered adaptation following upper respiratory infection, increased laryngeal tone due to local irritative conditions such as gastroesophageal reflux, compensation for underlying glottic insufficiency, and psychological or personality traits that express excess laryngeal tension [4].

The psychological traits of adult MTD patients have been studied in some depth. In the most extensive studies by Roy et al., personality profiles were obtained in large groups of patients with MTD, SD, vocal fold paralysis, vocal nodules, and normal controls [9–11]. MTD subjects scored high on dimensions of introversion, anxiety, depression, and emotionalism. Vocal nodule patients scored similarly on anxiety and emotionalism scales; however, instead of introversion (quiet, unsociable, passive, careful), they demonstrated extroversion (dominant, sociable, active). MTD is described as muscularly inhibited voice production in the context of individuals with personality traits of introversion and neuroticism. In response to certain environmental cues or triggers, elevated laryngeal tension creates incomplete or disordered vocal production in a structurally and neurologically intact larynx [4]. The psychological traits of children with functional voice disorders have not been similarly studied.

Despite the above issues that involve the cause of MTD, successful treatment of MTD through behavioral management has been demonstrated in a number of reports [5, 12, 13]. This focuses on the proximate causes of the dysphonia and rebalancing the laryngeal mechanism to produce normal voice. The most effective technique in our experience is manual circumlaryngeal massage and laryngeal reposturing to lower the larynx [5, 12, 14]. This can yield remarkable improvement, with two-thirds of patients achieving normal voice return from a single treatment session. Successful treatment with behavioral therapy in nearly all patients is expected. Recalcitrant or resistant cases may respond after several sessions of therapy. In a case series of pediatric patients treated for “muscle tension dysphonia” recently published, seven of the eight children had vocal nodules with supraglottic hyperfunction seen on laryngoscopy [15]. One patient had aphonia without lesions. All patients improved with voice therapy. As an adjunct treatment for severe MTD, Dworkin et al. reported the use of topical lidocaine spray to the larynx followed by voice therapy [16]. We have found this to be effective in selected pediatric patients. We also used lidocaine block of the recurrent laryngeal nerve to facilitate phonation in a case of recalcitrant functional aphonia in an adolescent [17]. Sensory or motor perturbation of the laryngeal mechanism may relax excessive laryngeal muscle tension and help the patient gain confidence that they have the capacity to produce normal voice.

The voice of adolescence is characterized by pitch instability. This is true for both males and females but more so in males. In a study of children aged 10–17 without and with vocal complaints, acoustic measures of pitch stability on sustained vowel phonation were not found to statistically distinguish the normal from several disordered voice groups [18]. However, the group diagnosed with puberphonia had the most variability of frequency and amplitude. Puberphonia is a voice disorder of adolescent males. It has also been labeled mutational falsetto, adolescent male transitional dysphonia, incomplete mutation, and persistent falsetto. It can be seen in early adolescence or can persist into late adolescence or adulthood. The voice does not successfully accomplish pitch change during puberty, between 12 and 14 years of age. The voice has been described as weak, thin, breathy, and hoarse in quality [6, 12]. A recent study in a large patient group with puberphonia measured the average speaking F_0 at 241 Hz [19].

It is frequently accompanied by downward pitch breaks into chest register. Coughing sound is also in chest register [6]. The voice of puberphonia may be described as a habituated use of falsetto register accompanied by pitch breaks rather than maintenance of the preadolescent voice. This pattern is commonly seen in MTD, so in our view, puberphonia is considered a variation of MTD seen in adolescent males. The larynx is generally positioned high in the neck, and excessive thyrohyoid tenderness and a narrow thyrohyoid space are found on palpation. Laryngeal lowering maneuvers, including head dorsiflexion, depression of the mandible, hyoid pushback, and laryngeal pulldown, are combined with vocalization [12]. This may create a surprised patient and his mother when his normal deep chest register voice is produced for the first time.

The first-line treatment of puberphonia is behavioral voice therapy [6, 12]. The same techniques of laryngeal lowering and reposturing combined with vocal cues that are used for MTD apply to the treatment of puberphonia. Ideally, this is conducted by a speech pathologist experienced in this approach. These techniques facilitate lowering of the laryngeal to engage the chest register and thyroarytenoid muscle activity to lower the pitch of the voice to the patient's normal male range. A recent study of 45 patients with puberphonia included 16 patients aged 11–15 years and 29 patients aged 16–40 years. All patients were treated successfully with behavioral therapy techniques with maintenance of improvement documented at 6 months [19]. A recent study from our institution documented the successful resolution of puberphonic voice in 12 consecutive patients, with the voice outcome documented by perceptual listener ratings and acoustic measures [20].

For recalcitrant cases of puberphonia, novel approaches have been tried including botulinum toxin injection to relax cricothyroid muscle function [21], pitch-lowering phonosurgical procedures including type III thyroplasty [22], hyoid detachment/laryngeal lowering laryngoplasty [23], and injection medialization laryngoplasty [24, 25]. The reasons for failure of behavioral voice therapy in these cases are unknown. These circumstances should be unusual and are not considered in most cases. The remarkable success of manual reposturing techniques, now documented in several reports, point to this as the first-line approach to treatment of puberphonia [19, 20].

The negative impact of functional voice disorders in children can be substantial. It may affect their ability to form and maintain social relationships with peers and adults, to communicate in school and home environments, and to enter the world of work. Although they are labeled "functional" because no underlying disease process involving the organs of voice and speech is found, the significance of the problem should not be minimized. The organs of voice and speech are neurally controlled, and this neural control is profoundly influenced by central nervous system controls involving emotional state, personality, and stress response as described above. The impact of voice disorders in children on their social, emotional, and physical function is just beginning to be investigated [26]. Voice-related quality of life instruments that are validated and age-appropriate for children and adolescents are greatly needed. Providers caring for these children need to aggressively advocate for needed services, such as voice and speech therapy provided by experienced clinicians. Documentation by video and audio recordings, patient-based quality of life measures, and references from peer-reviewed publications may all be needed in making appeals to insurance providers to cover speech therapy services for these patients.

Role of the Speech-Language Pathologist

The speech-language pathologist (SLP) has the role of restoring the voice back to the patient's previous normal/baseline (in the case of MTD) or producing and maintaining voice quality and pitch that is expected of patient's age and gender (in the case of puberphonia). In addition to maneuvers/techniques used to interfere with abnormal muscle contraction patterns to stimulate a normal voice, it is our experience that skilled clinicians apply an art of therapy that

is more challenging to quantify and easier to describe. It involves quickly establishing rapport/trust and being able to encourage, coach, and guide the individual to normal voice production. The clinician must also have the knowledge and expectation of the patient's potential for the functional voice disorder to resolve quickly with proper application of the appropriate therapy techniques to coach and guide the patient to normal voicing. This is certainly the case when the patient has failed numerous medical and behavioral treatment approaches. The child and parent may both feel skeptical that his/her dysphonia may be effectively treated with voice therapy, particularly in a single session.

Following auditory-perceptual evaluation, acoustic and/or aerodynamic recording, laryngeal imaging (if not already performed by the otolaryngologist or available for review), and an anterior neck examination with palpation of the perilaryngeal region to access for as described by Aronson [12], the patient undergoes stimulability testing/trial therapy. In our experience, the vast majority of patients with severe MTD/aphonia and puberphonia are restored to normal in a single treatment session as have been reported in functional dysphonia studies [4, 5, 20]. Various therapy techniques may be used in an attempt to stimulate normal voice production. We favor laryngeal reposturing maneuvers as a primary treatment approach. Laryngeal reposturing is combined with sustained voicing. Transient moments of improved or normal voicing is immediately identified and reinforced. After sustained vowels or voiced consonants are produced consistently accurately the improvement in voicing is extended across the speech hierarchy as manual techniques are faded. Negative practice is used to increase awareness of voice production patterns, increase control, and increase likelihood of generalization and maintenance [20]. In negative practice, the patient is encouraged to return and/or simulate the disordered voice during rote and other speech tasks and then quickly alternate back to the normal voicing patterns based on the clinician's verbal cues to "switch." Negative practice in the presence of a parent or other family member and having the patient converse with parent or family member after normal voicing has returned as a generalization activity can be very powerful for the patient's self-efficacy and setting the bar and expectation of the patient maintaining the normal voice and being able to improve his/her voice immediately if disordered voicing patterns return. The patient quickly developing the self-efficacy and ownership of the voice production during the therapy session is key for sustained improvement. To reinforce patient self-efficacy and ownership while simultaneously following the patient's progress, the SLP may ask the patient to telephone him or her later that day or the next day as well as in 1–2 weeks to report progress/maintenance without necessarily scheduling an additional therapy session with reinforcing that the patient is in control and he/she has the tools to improve his/her voice by "switching" or course correcting as he/she performed during negative practice if fluctuations occur.

The patient may have apprehension regarding possible questions by family, friends, and schoolmates following sudden change/normalizing in voice after their voice disturbance had been severe and chronic. In these situations where patients may have difficulties rationalizing how they are going to explain to others what happened to their voice, the patient is encouraged to explain that they saw a speech pathologist who specializes in voice and that the clinician performed laryngeal manipulation like a chiropractor or physical therapist that allowed for normal voicing. An individual with puberphonia may have some difficulty accepting "the new voice." The individual may need to be reassured that it is a "normal" voice that is compatible with his age, gender, and laryngeal size. Although the voice may be much clearer, smoother, stronger, and lower in pitch, the patient may be concerned about drawing more attention to his voice and consequently may be tempted to revert to the disordered voice production pattern. Playing pre- and posttreatment audio recordings for these individuals may be beneficial for the patient to agree that the new voice is more age- and gender-appropriate and will garner less attention than the disordered voice as well as reassuring the patient that after a few days of use, the voice will feel and sound more natural

and comfortable. Positive feedback from parents, family members, and friends is also very beneficial in reducing apprehension and increasing comfort and confidence in the new, normal voice.

Role of the Otolaryngologist

By the time children present to the pediatric otolaryngologist with a suspected functional voice disorder, they have often had the problem for many months. They may have missed a lot of school, had many doctor visits, and had an inordinate number of frustrating and unproductive speech therapy sessions. The frustration level of the family is high. The ideal setting to see these children is in a multidisciplinary voice clinic. However, the problem is so uncommon that these patients often present to physician or physician-extender clinics at a time when the speech pathologist is not present or readily available.

It is important for the pediatric otolaryngologist to identify a functional voice disorder and initiate a referral for voice therapy promptly. Since a fast resolution of the problem is possible with effective therapy, it is important to get the patient seen quickly. The pediatric otolaryngologist's role involves accurately identifying the problem with a medical diagnosis (muscle tension dysphonia ICD-10 code R49.0, puberphonia or mutational falsetto R49.8), explaining it to the patient and family in a way that does not assign blame or guilt, and explaining the necessity of voice therapy. This will usually require an explanation of the necessity of referral to a particular speech-language pathologist who is experienced in functional voice disorder treatment. General pediatric speech-language pathologists usually do not have experience with this patient population. Since children with functional voice disorders are usually older preteens or adolescents, we refer them to speech pathologists at adult voice disorders clinics, although they could also be referred to a speech pathologist who specializes in pediatric voice. Since the child may have already had prior speech therapy with unsuccessful outcomes, the pediatric otolaryngologist must be knowledgeable in why therapy with an experienced clinician is likely to be successful. They should provide encouragement, hope, and optimism for the patient and family to resolve the voice problem. Communication with the primary care provider and school may also be necessary to explain the uncommon nature of the problem and the recommended treatment.

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Vocal Fold Mobility Impairment

25

Karen B. Zur, Kimberly Duffy, and Linda M. Carroll

Vocal fold immobility in children may arise from iatrogenic or idiopathic etiology. The literature is inconsistent as to whether children are more likely to have bilateral or unilateral vocal fold immobility [1–3], but adults are commonly found to have unilateral rather than bilateral vocal fold compromise [4]. Bilateral vocal fold immobility in children has been reported with Charcot-Marie-Tooth disease [5], Arnold-Chiari malformation [6], tracheoesophageal fistula [7, 8], presence of intracranial tumor [9], or complication from vincristine therapy [10].

Unilateral vocal fold immobility in children is more commonly associated with injury to the nerve during neonatal cardiac surgery [2, 11–14], tumor excision, neurological disease, idiopathic, intubation, and birth trauma [1, 2, 15] and is more common with premature or low birth-weight patients [16]. Pourmoghadam et al. [11] reported incidence ranging from 48% to 65% following aortic arch reconstruction surgery and spontaneous resolution of vocal fold immobility in only 74% (Norwood procedure) to 86% (non-Norwood procedure) of patients. Tibbetts et al. [13] found unilateral vocal fold immobility incidence of 68.5% for cardiothoracic surgery. Iatrogenic etiology contrasts with the adult population in whom thyroid surgery [4, 17] and endotracheal intubation [4, 18, 19] are the most common causes of immobility.

Both unilateral and bilateral vocal fold immobility have been found in children following esophageal atresia repair [20] and following tracheoesophageal fistula repair [6, 8]. Genetic etiology is uncommon [21], but it may be seen in cases of bilateral vocal fold immobility. Gestational age does appear to be an influence on incidence of vocal fold immobility [22], but this may be due to increased likelihood for cardiac surgery in the premature patient.

Intubation longer than 3 hours is known to increase the risk for vocal fold paralysis in adults, but there is little formal assessment of maximum length of intubation tolerance in neonates or young children. It is, however, generally accepted

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that prolonged intubation, repeat intubation, or self-extubation in children increases the risk for vocal fold immobility.

Quality of life compromise is greater for children with vocal fold immobility compared to children with dysphonia due to other pathologies [23]. The presence of vocal fold immobility in children affects speech-language development, social development, and sports involvement and can affect academic success [24]. Poor vocal power and lack of voice stability impact on the child's ability to communicate orally, explore language, establish relationships, and participate in social and academic settings [21]. Connor et al. [24] reported that dysphonic children may limit verbal interactions fearing "undue attention" and lack of respiratory skills may limit participation in social and sporting events. Modified diet or the need for a gastrostomy tube [25], which may be needed for the child with vocal fold immobility, further impacts social eating skills and may have downstream effects on nutrition and the ability to thrive [26].

In children with bilateral vocal fold paralysis requiring tracheostomy placement, there is an increased risk of delayed speech and language development. If there is little or no airflow around the tracheostomy, this can lead to aphonia or a decreased ability to achieve voicing. A study comparing tracheostomy-dependent children with and without neurologic impairments found that the tracheostomy alone affects a child's speech and language development [27]. Recently, a cohort study evaluating developmental outcomes of very preterm infants over a 10-year period found that most developmental outcomes, including both receptive and expressive language, were significantly worse in the children with tracheostomies [28]. Specific speech sound deficits in children with tracheostomies include slow acquisition of speech sounds and excessive speech production errors (specifically phonological processes and difficulty with vowel productions) [29, 30].

Patients with unilateral or bilateral vocal fold immobility commonly present with weak cry, weak voice, aphonia [8], stridor [6, 7, 31], dyspnea [8], respiratory distress [32], dysphagia [2], feeding difficulties [33], aspiration [34], and pneumonia [13]. Nichols et al. [22] reported persistent complaints of dysphonia (78%), dysphagia (55%), and respiratory symptoms (39%) for children with vocal fold paralysis.

Among vocal symptoms, access to upper vocal range is compromised, voicing stability is impaired, maximum phonation time is reduced, and vocal power is reduced [35]. Respiratory difficulties in infancy can include stridor, apnea, cyanosis, increased work of breathing, and desaturations [36]. In infants, this can result in poor suck/swallow/breathe coordination while breast or bottle feeding due to obstruction caused by the vocal cord paralysis [37]. The end outcome can be aspiration or decreased oral intake affecting the baby's ability to accept adequate nutrition to grow and gain weight. Babies with airway obstruction may have increased effort for baseline breathing, affecting endurance or reserve to meet the increased demands for feeding tasks [38]. In older children, there is an increased risk for aspiration of liquids due to incomplete vocal fold closure during the swallow.

Laryngeal examination has found left-sided vocal fold immobility to be more common in both children [2, 14] and adults [39]. Endoscopic and stroboscopic features are consistent for children and adults: flaccid membranous vocal fold, possible medial rotation of the arytenoid for the affected side, loss of adduction of the arytenoid during phonation, glottic incompetence, glottal gap, and poor mucosal wave propagation. Because of the quality of life compromise for children with vocal fold immobility for the larynx which will continue to grow and evolve through puberty, management options need to be explored and a treatment plan developed [40].

Immediate management of vocal fold immobility is necessary when the airway is compromised. Daya et al. reported the need for tracheotomy in 57% of children with bilateral vocal fold paralysis in the adducted position [1]. Recently, endoscopic anterior and posterior cricoid split or use of onabotulinum toxin A injection has been advocated for newborns and infants in an effort to avoid tracheostomy [41–44]. Once there is a stable airway, attention is paid to laryngeal function for voice and swallow.

The management plan falls into three options: observe (wait and see), voice/speech therapy, or surgical intervention (injection, reinnervation, or laryngeal framework surgery). When swallow deficits are also present, there is greater urgency for a surgical plan. Parents of younger children may opt for laryngeal reinnervation due to the potential impact of vocal fold immobility on speech and language development. Young children may also be unable to participate in voice and speech therapy to learn compensatory strategies for the vocal fold immobility. Children who are able to compensate well for vocal fold immobility or those who report reduced interference with daily skills are less likely to elect a surgical intervention of injection or reinnervation. The trends toward use of laryngeal reinnervation are now international and have replaced previous attempts of laryngeal framework surgery in the pediatric population.

Recovery from bilateral vocal fold immobility is poor in children, with up to 11 years reported for idiopathic etiology [1]. Recovery of laryngeal function (unilateral or bilateral) without surgery is reported in 3–36% of children with vocal fold paralysis [3, 22, 34]. Laryngeal electromyography suggests poor prognosis of nerve recovery if there is absence of normal-appearing motor unit action potentials by 6 months following patent ductus arteriosus repair (PDA ligation) [45]. Jabbour et al. reported resolution of immobility symptoms in only 28.9% of children [2]. This points to the need to offer both short-term and long-term management for these patients.

Speech-Language Pathologist Approach

Assessment of Function: Voice

Voice is assessed through informal and formal methods [46]. Perception of voice quality is accomplished through the clinician's judgments of overall vocal features during conversation and formal speech and voice tasks and uses either the traditional Grade-Roughness-Breathiness-Asthenia-Strain (GRBAS) [37], Consensus of Auditory-Perceptual Evaluation of Voice (CAPE-V) [47], or Acoustic Voice Quality Index (AVQI) [48]. Parental report of dysphonia severity is accomplished through use of the Pediatric Voice Handicap Index (pVHI) [49], Pediatric Voice-Related Quality of Life (pvRQOL) [50], or the Pediatric Voice Outcome Survey (pVOS) [51].

Instrumental analysis is becoming increasingly available with the emergence of free acoustic analysis programs for computers and smartphones. Objective acoustic assessment includes overall vocal range for fundamental frequency and intensity and vocal stability measures such as jitter, shimmer, noise-to-harmonic, cepstral peak prominence [52], and spectral analysis [53]. Objective aerodynamic assessment includes maximum phonation time, transglottal air flow, and subglottal pressure measures [54]. Except for maximum phonation time, aerodynamic measures remain largely limited in use by clinicians due to the necessary technology to capture aerodynamic signals. At our multidisciplinary voice clinic, typical measures that are obtained include a pVHI for all patients at each visit, GRBAS evaluation, and acoustic measures recorded using VisiPitch.

Respiratory coordination is assessed through observation of breath pacing during speech tasks. Children with vocal fold immobility are recognized to have greater incidence of inspiratory phonation. This may be a strategy to permit long oral communication in the presence of significant loss of air due to vocal fold paralysis but is contraindicated for establishing normal voicing patterns.

Part of an interdisciplinary assessment of voice includes stimulability testing in order to determine candidacy for voice therapy and to develop a treatment plan. In a patient with a vocal fold paralysis, the goal of therapy is to establish the best vocal quality possible in the setting of the paralysis. Therapy can be conducted prior to and/or following surgical intervention. Voice therapy techniques used with adults for vocal fold paralysis can also be used with school-age children provided they have the maturity and cognitive ability to participate in behavioral therapy [55]. During

the evaluation, the speech-language pathologist (SLP) will probe techniques to reduce tension, improve coordination of breathing and phonation, and decrease laryngeal hyperfunction [56]. Laryngeal massage in addition to stretching and relaxation is often used to reduce laryngeal tension and avoid supraglottic phonation [57]. Breath support and behavioral techniques, including use of vocal function exercises [57], resonant voice therapy [58], semi-occluded vocal tract exercises [59], and respiratory coordination for phonation, can also be effective therapeutic strategies in patients with vocal cord paralysis. Kinesio taping has been advocated as an adjunctive treatment for dysphonia in adults with vocal fold immobility, but the techniques have not been applied to children [60].

Assessment of Function: Swallow

Assessment of swallow function includes a clinical evaluation and/or an objective assessment. The first step is often a clinical evaluation conducted by an SLP which includes review of the medical history, a physical examination including assessment of respiration, an oral mechanism exam, and a feeding observation. With infants, there is a focus on pre-feeding behaviors, sucking mechanics, and other structural or functional issues that may affect oral feeding. The goals of a clinical feeding evaluation include developing a safe feeding plan with consideration for signs of airway compromise including inspiratory stridor, stertor, coughing, choking gagging, color changes, and desaturations [61], determining the need for an objective assessment, and making recommendations for other consultations that may be beneficial [62]. The feeding observation looks to determine feeding efficiency, evaluate dysphagia signs including aspiration, and trial of therapeutic strategies and diet modifications. In infants, a feeding evaluation may reveal signs of respiratory distress while feeding including arching, refusal, increased work of breathing, prolonged feeding, and inadequate weight gain [63]. During a feeding observation, the SLP may trial therapeutic strategies including a change in consistency to decrease risk of aspiration [64], position changes such as a side-lying position for infant feeding to improve suck/swallow/breathe coordination [63], a chin down position or head turn with older children [65, 66], pacing, decreased flow or bolus size, and/or other compensatory swallowing strategies.

Following a clinical assessment, an objective assessment of swallowing may be warranted to further assess swallowing function, determine the safety of swallowing by identifying swallow function characteristics and deficits, evaluate the effectiveness of strategies and intervention, establish a safe diet, and develop a rehabilitative plan [67]. Objective assessments should be viewed as dynamic tools with results and recommendations that fall on a continuum rather than a pass/fail test.

Counseling and Education

Once a diagnosis of vocal fold immobility has been made, the voice care team develops a comprehensive and overlapping plan for counseling and education. Due to the complexity of downstream effects of severe vocal fold immobility, the team often expands to include nutritionists, dieticians, and gastrointestinal specialists, as well as continued monitoring by an airway team. Parents of children with vocal fold immobility should be educated on the airway, voice, swallow, and nutritional ramifications of vocal fold immobility. Parents need education on the value of a multidisciplinary team approach to aerodigestive complaints surrounding vocal fold immobility [68]. Once education has begun, counseling should address the stress management skills for the family as well as the patient. Compensatory strategies must be implemented right away to improve the family's ability to provide a safe, effective environment to nurture the child with vocal fold immobility as a short-term and long-term treatment plan is developed.

Patients who choose injection laryngoplasty note an immediate improvement in voice and

swallow function, but the results are not typically long-term. However, this intervention affords an opportunity to "get over the hump" and achieves improved function, enabling implementation of more effective compensatory strategies that may be sufficient for the patient with moderate vocal fold immobility. Patients who elect reinnervation gain the short-term benefit from the concurrent injection laryngoplasty with the long-term benefit of restored intrinsic laryngeal muscle tone. Long-term success rate is high for reinnervation, but there may remain underlying issues for patients with extreme prematurity. Surgical patients are typically seen at 3- to 6-month intervals, with reinnervation patients monitored for 5 years or longer. Patients who live great distances from the phonosurgeon are requested to send regular voice recordings and commentary on their voice and swallow skills, a pVHI form, as well as maintain regular appointments with their local laryngologist.

Otolaryngologist Approach

Assessment of laryngeal function is often adjusted for the young pediatric patient. While school-aged children tolerate examination methods commonly used in the adult population, younger children benefit from a kid-friendly examination routine. Interviewing the parent in the presence of the child and recording voice-only tasks during the initial portion of the exam help reduce fears for the young patient. During the laryngeal exam, the young child should be held on the lap of their parent, and the medical team should be prepared for expected screaming and crying during the flexible endoscopic exam. This is normal. An effective laryngeal exam can be done in children under 2 years of age, and much information can be obtained from even highly resistant patients. It is not uncommon for children to report immediately after the exam "that wasn't so bad!" and then join the post-examination conversations.

Fiberoptic laryngoscopy, with or without stroboscopy, has long been the standard for assessment of vocal fold immobility in children. Use of the smaller flexible pediatric endoscopes is preferable and now available. During the last decade, there has been increasing promise to determine presence of vocal fold paralysis through the use of the less invasive ultrasonography, with Shaath et al. [69] reporting 90% success rate for diagnosing immobility.

Options

If the pediatric patient notes mild difficulty with vocal range (pitch and volume) and has no swallow complaints, vocal fold immobility may be monitored for a period of time before any serious discussion of surgery. This may be the case for a patient who compensates well for the vocal fold paralysis, has a recent injury which may be suggestive of a vocal fold paresis rather than paralysis, has recently undergone thyroidectomy, does not have significant voice disability (typically as measured through administration of the pVHI), or is not of sufficient age or cognitive skill to participate meaningfully in voice therapy.

Voice therapy is the preferred management with mild or moderate vocal fold immobility when some contact between the two vocal folds can be appreciated. Although successful voice therapy can be accomplished in children as young as age 2 years, many clinicians opt for age 3 to begin voice therapy. Children with moderate-severe or severe voice or swallow difficulties are typically offered surgical intervention (injection laryngoplasty or laryngeal reinnervation) with voice/speech therapy during the postoperative period.

Laryngeal Electromyography

Laryngeal electromyography (LEMG) is an invasive yet important tool in prognostication of vocal fold immobility [70]. LEMG is safe and reliable in the pediatric patient and is essential to determine the presence of recurrent laryngeal nerve injury and the best treatment plan [45].

Intraoperative LEMG permits a detailed inspection of the thyroarytenoid, interarytenoid, and posterior cricoarytenoid muscle integrity, including analysis of motor unit action potentials and comparison of right versus left side. This information is essential to determine the appropriate surgical treatment for vocal fold immobility and to ensure no arytenoid fixation is present if reinnervation is under consideration. If the LEMG is normal or if the LEMG shows evidence of paresis rather than paralysis, only injection laryngoplasty is performed.

Operative Approach

Injection laryngoplasty often serves as the first surgical option for both children and adults with vocal fold paresis or paralysis and offers an immediate improvement in glottic closure. Although office-based injection is common for adults, administration of an injectable for younger children is typically limited to the operating room [71]. Typical injection volume is 0.1–0.7 mL, allowing for adequate plumping of the immobile vocal fold. There are a variety of injectable materials available for use in children, but may not be available to all phonosurgeons due to international restrictions. Injectables include carboxymethylcellulose gel (Prolaryn®), micronized alloderm tissue (Cymetra®), polydimethylsiloxane, calcium hydroxylapatite (Prolaryn Plus®), Gelfoam®, and hydrated porcine gelatin powder (Surgifoam®) [71–75]. Multiple injectables have been found to be beneficial in children, but none address the underlying neurological deficit nor provide a long-term solution [35]. Autologous fat may also be used in children, but it is more commonly utilized for the management of adult vocal fold immobility.

The use of recurrent laryngeal reinnervation procedure has been reported by Jackson since the early 1900s, with Crumley and Izdebski bringing this approach to the forefront after the emergence of Isshiki thyroplasty procedures [76, 77]. Laryngeal reinnervation is more successful in children than adults, largely owing to the etiology of the paralysis and the age of the patient. It is now considered a first-line treatment in pediatric vocal fold paralysis [72]. Children are more likely to have laryngeal nerve compromise due to cardiac surgery, whereas adults are more likely to have laryngeal nerve compromise due to thyroid surgery [78]. Therefore, the child patient has a "naive neck" which renders identification of a viable donor nerve and an intact recipient nerve more successful. Many surgeons use a short-term vocal fold injection at the time of reinnervation to help the patient gain immediate benefit from the procedure. This benefit lasts 2–3 months but bridges the time gap from the day of the nerve anastomosis to the time it takes to start seeing the impact of the reinnervation post-procedure. Patients who undergo laryngeal reinnervation are discharged on the day of surgery and return for their first follow-up in 4–6 weeks.

Patients with multiple laryngeal diagnoses (vocal fold paralysis, vocal fold cyst) still benefit from reinnervation but do not gain the same degree of benefit [35]. Parents of young children are often hesitant to pursue an elective surgery due to the emotional trauma they may have experienced from the child's previous surgeries, as well as concerns about a visible scar in the neck. Reinnervation has been successful for children less than 2 years of age and has been shown to be successful in children even when there is over a decade delay from onset of unilateral paralysis to reinnervation surgery [35, 79]. Repetition of laryngeal EMG 2 years or more following reinnervation has shown evidence of successful reinnervation [35]. For those pediatric patients with dysphagia due to vocal fold immobility, Zur and Carroll [80] found normal modified barium swallow study beginning at 6 months post-reinnervation.

Laryngeal framework surgery is more successful in adults than children and is no longer commonly offered for children. The size and placement of the implant is more problematic in children with risk of migration and airway compromise. With the continued growth of the laryngeal structures, both short-term and long-term success is difficult [76, 81].

Operative Approach: Injection

Following confirmation of nerve impairment by laryngoscopy and laryngeal EMG, an injection laryngoplasty is performed with the patient under spontaneous ventilation and in a suspended position. The volume of injectable varies with laryngeal size, severity of the glottic gap, and physical capacity to accommodate the viscosity of the injectate. As with adults, extrusion of the injection material may occur from the needle insertion site. Insertion of the material should be immediately anterior and lateral to the vocal process (Fig. 25.1). The voice improvement is typically noted at about 2 weeks postinjection, with benefits lasting 2–3 months for the gel injections. The longer-term injection with material such as Cymetra can provide a benefit of around 1-year post-procedure. Autologous fat injection durability in children is not well studied. A reduction in pVHI and GRBAS scores is seen following injection laryngoplasty, but the benefit is not long lasting [35].

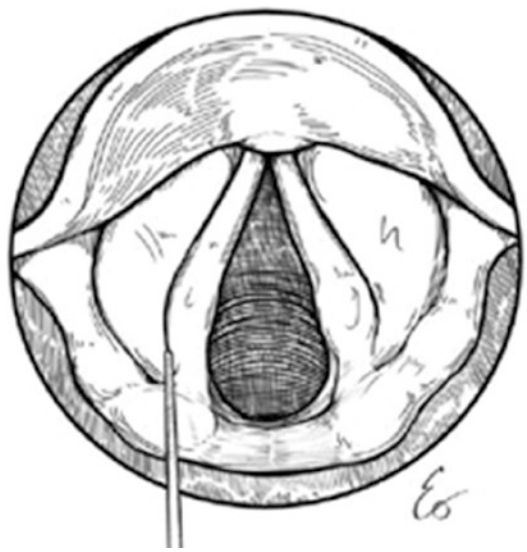


Fig. 25.1 Diagram depicting insertion of an injection needle immediately anterior-lateral to the vocal process. (From Potsic, Cotton, Handler and Zur, *Surgical Pediatric Otolaryngology*. 2nd edition. Thieme, 2016, with permission)

Operative Approach: Recurrent Laryngeal Nerve Reinnervation

Laryngeal reinnervation may be accomplished in children using the traditional open neck approach; however a transaxillary endoscopic robot-assisted laryngeal reinnervation procedure has been reported as well [82]. Experienced phonosurgeons accomplish the open neck operative technique in less than 2 hours. Typically, no drain is required and the patient is discharged on the day of surgery.

Voice and swallow benefits from laryngeal reinnervation emerge after at least 3 months, and there is typically a gradual improvement over the ensuing 18 months. When the voice begins to strengthen, it is typically due to improved glottic closure. When that occurs, patients who had preoperative aspiration and swallowing difficulties would undergo a repeat clinical and/or radiographic assessment of their swallowing function to see if any restrictions on a thin liquid diet can be lifted. A reduction in pVHI averages 15 points more in post-reinnervation patients compared to those patients who chose injection laryngoplasty. Vocal range (measured in semitones) is greater post-reinnervation compared to injection, and the stability of fundamental frequency (measured through jitter) is significantly reduced for reinnervation patients compared to the injection patients. Cartilaginous positioning is improved in the majority of patients, but it is not consistent across cartilaginous features of arytenoid position, arytenoid height, arytenoid rotation, or arytenoid mobility during phonation [35] (Fig. 25.2).

Steps

1. Confirm vocal fold paralysis through laryngeal electromyography and laryngoscopy.
2. Make a curvilinear incision on the affected side, centering incision over the medial belly of the sternocleidomastoid (SCM) muscle.
3. Skeletonize the SCM, and identify the omohyoid as it courses medially.
4. Retract the omohyoid inferiorly and anteriorly (Fig. 25.3).

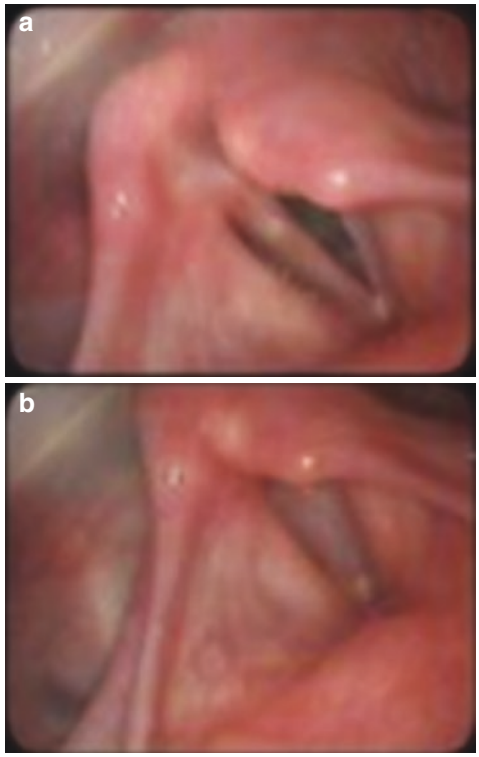
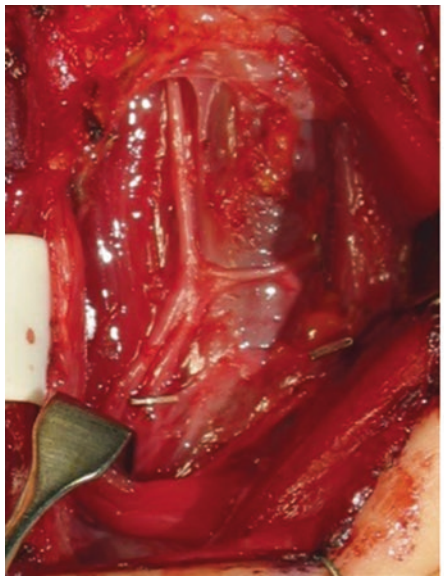


Fig. 25.2 Images before and after a recurrent laryngeal nerve reinnervation. (**a**) Preoperative glottic insufficiency. (**b**) Twenty months post-reinnervation complete glottic closure

5. Dissect the ansa cervicalis along the internal jugular vein. Once the loop of the ansa cervicalis is identified, dissect the more medial branches as possible donor nerves (Fig. 25.3).
6. Dissect inferiorly to the thyroid gland in search of the recurrent laryngeal nerve. Ensure preservation of the parathyroid glands that may be coursing inferiorly.
7. Elevate the strap muscles to form a tunnel to bridge the lateral and medial compartments. Placing a ¼ in. Penrose may be useful to retract the strep medially or laterally depending on the exposure (Fig. 25.4).
8. Stimulate the donor nerve to ensure presence of electrical activity.
9. Section donor nerve and the recurrent laryngeal nerve. Use one or two epineural sutures (9-0 ethilon BV-100-4 monofilament) placed in an end-to-end manner utilizing microscope (Fig. 25.5). Apply Tisseel over the anastomosis.
10. Return strap muscles to their natural position.
11. Irrigate the wound, and close with subcuticular sutures. No drain is needed.



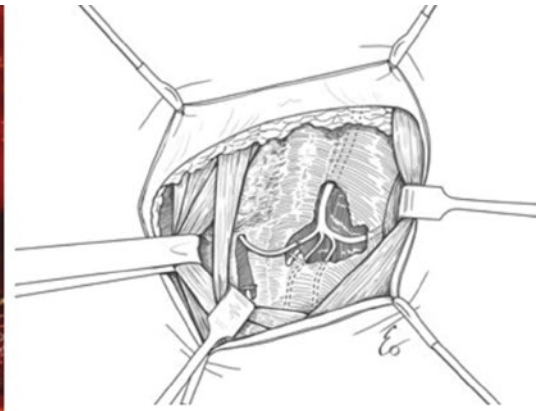


Fig. 25.3 Recurrent laryngeal nerve reinnervation. The omohyoid muscle is retracted medially and the loop of the ansa cervicalis is skeletonized. One of the branches is clipped in preparation for an anastomosis. (From Potsic, Cotton, Handler and Zur, *Surgical Pediatric Otolaryngology*. 2nd edition. Thieme, 2016, with permission)

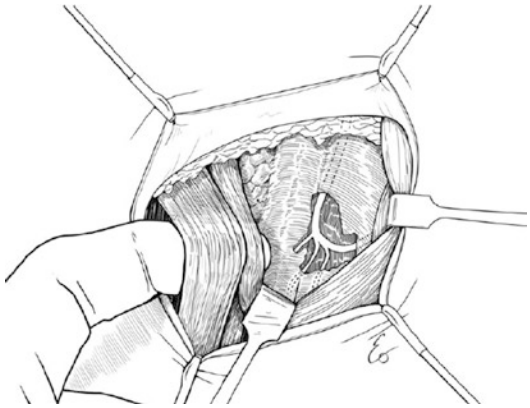


Fig. 25.4 Elevation of the strap musculature to allow a bridge between the donor and recurrent laryngeal nerves with identification of the distal recurrent laryngeal nerve. (From Potsic, Cotton, Handler and Zur, *Surgical Pediatric Otolaryngology*. 2nd edition. Thieme, 2016, with permission)

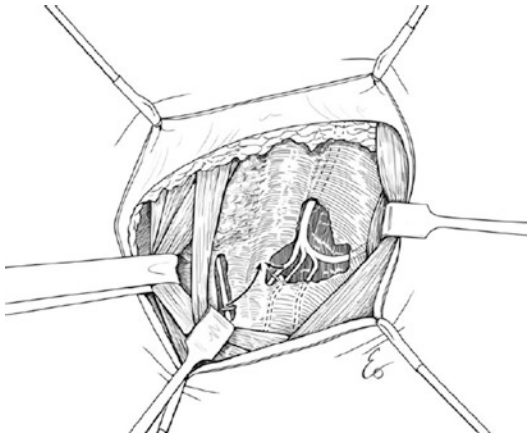


Fig. 25.5 Schematic representation of the surgical anastomosis of the recurrent laryngeal nerve and the ansa cervicalis donor nerve. (From Potsic, Cotton, Handler and Zur, *Surgical Pediatric Otolaryngology*. 2nd edition. Thieme, 2016, with permission)

Summary

The prevalence of dysphonia in the pediatric populations ranges from 6% to 53% [83, 84] with vocal fold paralysis among the top three most common disorders. Whether unilateral or bilateral, patients with vocal fold immobility disorders experience compromise in their quality of life and report poor voice and swallow skills. Due to increased risk for underlying vocal fold paralysis, children with tracheoesophageal fistula or Charcot-Marie-Tooth disease should have laryngeal examination. Because of the high risk for vocal fold immobility following cardiac surgery, these patients should be carefully assessed before and after surgery.

Fiberoptic laryngoscopy remains the standard assessment tool for vocal fold immobility, but there is emerging success of ultrasound imaging [70, 85] to reliably check laryngeal function. The use of ultrasound to determine presence of vocal fold immobility offers great promise to the international community. Confirmation of vocal fold immobility, however, is accomplished with laryngeal EMG, which should be done in all patients prior to laryngeal nerve reinnervation or laryngeal framework surgery.

Team management of the child with vocal fold immobility is essential due to the coexistence of voice, swallow, and respiratory deficits which impact on the child's social and academic opportunities. It is important to create a global treatment plan that encompasses both short-term and long-term solutions for children with vocal fold disorders. With appropriate management, these patients can achieve good voice and swallow function, affording them greater potential throughout childhood and adulthood.

Emerging and Evolving Concepts

Laryngeal reinnervation has been shown to be highly successful in children, whether done through the traditional hands-on open neck approach or robotic-assisted surgery. As Telehealth expands, laryngeal reinnervation may be possible in more remote regions of the world. Management of vocal fold immobility through use of electrical pacing units [86] may also be possible in the future for those pediatric patients who are not candidates for laryngeal reinnervation.

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Laryngomalacia

26

Bethany R. Powers, Bryn K. Olson-Greb, and Jessica Van Beek-King

Overview

Laryngomalacia is a congenital immaturity of the cartilages of the supraglottis that presents with high-pitched inspiratory stridor that worsens with crying, feeding, or lying supine. It is the most common cause of stridor in the newborn and usually presents around 2 weeks of age. However, it may also present anytime within the first 2–4 months of life [1–3].

Laryngomalacia symptoms typically peak at age 6–8 months and resolve by age 12–24 months [3]. About 10% of laryngomalacia cases will require surgical intervention for apnea or failure to thrive secondary to feeding intolerance or utilizing excessive calories for respiration [4].

Classification Systems

There are various classification schemes described in the literature. Nonetheless, a universal classification system is not widely used. In 2006, Goldsmith described a three-type classification system based on laryngeal examination [4]. Type 1 is characterized by a foreshortened aryepiglottic fold. Type 2 is described as excessive soft tissue in the supraglottis, such as overlying the arytenoid cartilages. Finally, type 3 is defined as cases caused by other etiologies such as neuromuscular disorders, with a retropositioned epiglottis.

Another classification system is similarly described as type I, II, and III, but the assigned number corresponds to different structures on laryngoscopic exam [5]. Type I is collapse of supra-arytenoid tissue, type II is foreshortened aryepiglottic folds and an omega-shaped epiglottis, and type III is a retro-positioned epiglottis. Therefore, the numerical description of the classification system should not solely be used to make assumptions about the affected anatomical location.

Most recently, del Do and Camacho published a classification guide where laryngomalacia

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corresponds with the type of supraglottoplasty performed [6]. This simplifies categories to the anatomical site operated on. Type 1 is excessive arytenoid tissue, type 2 is foreshortened aryepiglottic folds, and type 3 is a retro-positioned epiglottis. Therefore, type 1 supraglottoplasty debulks the arytenoid soft tissue, type 2 supraglottoplasty divides foreshortened aryepiglottic folds, and type 3 supraglottoplasty is divided further into three subtypes. These subtypes are 3a, epiglottopexy; 3b, epiglottoplasty; and 3c, epiglottectomy.

Dana Thompson describes symptoms of laryngomalacia with a mild, moderate, and severe classification [3]. Mild is defined as having symptoms of inspiratory stridor and occasional episodes of feeding-associated symptoms such as coughing, choking, and regurgitation. Patients with mild laryngomalacia continue to have a coordinated suck, swallow, and breathe mechanism and have an average oxygen saturation of 98–100% [7]. Moderate laryngomalacia patients have frequent feeding-associated coughing, choking, or regurgitation, and their caregivers report difficulty with feedings. These patients have a lower resting oxygen saturation at 96%. Additionally, 28% of these patients may progress to a severe category and require surgical intervention. Therefore, patients with moderate laryngomalacia should be closely monitored. Severe laryngomalacia presents inspiratory stridor, recurrent apneas, cyanosis, aspiration with recurrent respiratory infections, feeding intolerance, or failure to thrive [3]. Infants with severe laryngomalacia have a lower resting SpO2 of 86% and frequently require surgical intervention to help prevent development of chronic airway resistance sequelae such as pulmonary hypertension or cor pulmonale or to address failure to thrive.

Epidemiology

Laryngomalacia affects 45–75% of all infants presenting with congenital stridor [8]. However, the exact etiology of laryngomalacia remains unknown. Upon initial evaluation of infants with laryngomalacia, 40% will have mild laryngomalacia, 40% will have moderate laryngomalacia, and 20% will have severe laryngomalacia when using Thompson's classification [3].

Medical Comorbidities

Gastroesophageal and Laryngopharyngeal Reflux

Laryngomalacia is associated with concurrent gastroesophageal and laryngopharyngeal reflux in 61–100% of patients [7, 8]. Negative intrathoracic pressure is created secondary to attempted inspiration against a high-resistance system, promoting reflux of gastric contents [3]. Chronic exposure to acid reflux bathes the larynx and supraglottic structures, decreasing sensation in the larynx. This is commonly associated with an impaired swallow [3]. Reflux should be treated in patients with laryngomalacia with either type 2 histamine receptor antagonists (e.g., ranitidine 5 mg/kg twice daily) or proton-pump inhibitor (e.g., omeprazole 1 mg/kg daily).

Secondary Airway Lesions

Secondary airway lesions (SAL) are reported between 7.5% and 65% with the majority of these lesions being subglottic stenosis, tracheomalacia, or bronchomalacia [9–15]. Secondary airway lesions create an additional anatomic level of obstruction, increasing airway resistance further, thus increasing intrathoracic pressure and gastroesophageal reflux. Infants who present with mild or moderate laryngomalacia are 4.8 times more likely to require surgical intervention for their laryngomalacia when there is an associated SAL [3, 13].

Polysomnography

Polysomnography (PSG) was previously studied on laryngomalacia patients, before and after supraglottoplasty. Measures recorded in this study were mixed and obstructive sleep apnea events

as well as desaturation events. The preoperative baseline mixed or obstructive apnea index for the group was 14.75 apnea events per hour compared to 2.2 postoperatively. The number of desaturations per hour was not significantly different [16].

Genetic Conditions

Genetic conditions such as Down syndrome, Pierre Robin sequence, CHARGE, and 22q11.2 microdeletion are all associated with laryngomalacia. Unfortunately, these patients may not show complete resolution of laryngomalacia symptoms with supraglottoplasty owing to the nature of the underlying condition [3]. Still, the senior author's experience is that patients with Down syndrome, in the absence of cardiac or neurological disease, do well with aggressive acid suppression and supraglottoplasty [3].

Pathophysiology

Inspiratory stridor is caused by redundant tissue or poor tone of the supraglottic structures characterized by foreshortened aryepiglottic folds, redundant arytenoid mucosa, or a retropositioned epiglottis. Stridor is created by rapid and turbulent airflow through a narrowed airway due to the aforementioned abnormal structures. This resistance creates increased intrathoracic pressure leading to reflux episodes as described above.

Speech-Language Pathologist Approach

The speech-language pathologist (SLP) serves an important role in the assessment and management of patients with laryngomalacia due to the high prevalence of dysphagia in this population [17, 18]. Duties may include gathering a thorough case history, completing a clinical swallowing evaluation, and performing instrumental swallowing evaluations as needed. The information obtained by the SLP elucidates the functional impact of laryngomalacia and thus may guide decisions regarding medical and/or surgical management.

Case History

The speech-language pathologist should take a thorough case history including the following elements:

1. Onset of breathing symptoms.
2. Factors, such as positioning or activity, which exacerbate or alleviate symptoms.
3. Feeding or swallowing concerns, including signs and symptoms of penetration, aspiration, or feeding distress; history of fever, pneumonia, or other pulmonary compromise; and difficulty with weight gain.
4. Other relevant medical histories that could contribute to breathing concerns include prematurity, complications encountered during gestation or delivery, neurological conditions, GERD/LPR, history of intubation, and other medical comorbidities.

Feeding/Swallowing Evaluation

Dysphagia is a common finding in patients with laryngomalacia. Upper airway obstruction is detrimental to suck, swallow, and breathe coordination and can result in inefficient feeding and/or poor airway protection with resultant penetration or aspiration. Consensus recommendations for laryngomalacia from the International Pediatric Otorhinolaryngology Group (IPOG) include a feeding and swallowing evaluation when coughing, choking, regurgitation, feeding difficulty, poor weight gain, and/or failure to thrive is present [19]. However, silent penetration and aspiration are common in this population [18, 20]. Abnormal swallow studies are seen in patients both with and without clinical signs of dysphagia, and infants with other medical comorbidities are at increased risk for feeding issues [21]. Additionally, the sensitivity of clinical evaluation to detect penetration and aspiration

in this population is poor [18]. As such, an ideal SLP evaluation should consist of the following components:

1. Clinical evaluation with particular emphasis on suck, swallow, and breathe coordination and exploration of strategies to optimize feeding coordination and reduce risk for penetration/aspiration. Strategies that may be beneficial for this population include side-lying or side-lying semi-upright positioning, altered nipple flow rate, and use of external pacing. More thorough explanation of a clinical swallow evaluation may be found in the chapter titled "Clinical evaluation of pediatric swallow."
2. Flexible endoscopic evaluation of swallowing (FEES) is used to both visualize the laryngopharynx to aid in physician diagnosis as well as to assess swallowing function, identify possible penetration/aspiration, and more objectively assess the efficacy of feeding strategies. Please see chapter on Flexible Endoscopic Evaluation of Swallowing for additional information regarding this evaluation.
3. Videofluoroscopic swallow study (VFSS) is used as needed. If the patient is unable to tolerate FEES or inadequate information is obtained with the combination of clinical evaluation and FEES, providers should maintain a low threshold for adding VFSS. Please see VFSS chapter for more information about this evaluation.
4. Repeat of instrumental swallowing evaluation, either FEES or VFSS, may be used to reassess swallowing function as needed (e.g., in patients with a history of silent aspiration who may be ready for diet advancement due to sufficient passage of time and/or recovery from surgical intervention).

Intervention

The speech-language pathologist's role with respect to intervention for patients with laryngomalacia centers around implementation of modifications to allow the infant to feed safely and efficiently. Thorough clinical and/or instrumental evaluation should identify any oropharyngeal swallowing deficits such as impaired suck-swallow-breathe coordination, inefficient feeding, or penetration/aspiration during feeding and subsequently guide the approach to intervention. Breathing difficulty in patients with laryngomalacia tends to be exacerbated by supine positioning [18]; thus, positioning the patient in an upright, side-lying, or side-lying, semi-upright position may be of benefit. External pacing, thickening feedings, and altering bottle flow rate are additional strategies that may be considered depending on the nature of the dysphagia [22, 23]. In cases where feeding and swallowing are so severely impacted as to threaten the patient's ability to maintain adequate nutrition and hydration orally, alternative means of nutrition may be necessary. In such cases, the speech-language pathologist should work closely with the patient's medical team, as feeding difficulty and failure to thrive are among the most common indications for supraglottoplasty [24].

Otolaryngologist Approach

History

Consultation for laryngomalacia may occur in the inpatient or outpatient setting. The referring chief complaint is most commonly "noisy breathing." However, the more precise label describing the respiratory abnormality in laryngomalacia is "inspiratory stridor." The first step with any stridor consult is establishing airway safety. Is the patient stable? Are they currently in any respiratory distress? What was the time of onset of stridor? Once these questions have been answered, a systematic approach to laryngomalacia occurs. A thorough history is obtained with a focus on pertinent information related to the noisy breathing as follows. Does the stridor worsen with change of position, time of day, during agitation or activity, or feeding? Are there any associated episodes of apnea, cyanosis, increased work of breathing such as tracheal tugging or subcostal retractions, or reflux episodes? Furthermore, a weight should be obtained and the growth curve examined for failure to thrive.

Exam

Physical examination begins with evaluation of the patient under resting conditions upon arrival to the clinic. Breathing patterns and any stridor or stertor are noted prior to agitation, which commonly occurs during physical examination in infants. A general head and neck examination should be performed. Breathing should also be evaluated under conditions where stridor and laryngomalacia symptoms are prone to manifest, like feeding or agitation, if possible.

Instrumented Assessment

Laryngomalacia patients are seen either by the physician alone or in conjunction with a speech-language pathologist. A flexible fiberoptic laryngoscopy is performed. If using the team approach, the flexible fiberoptic exam is evaluated together. The hallmark of diagnosing laryngomalacia hinges on dynamic laryngeal examination. Thus, clinic or bedside flexible fiberoptic laryngeal evaluation must be completed prior to making any management decisions. Evaluation should be focused on the three anatomical sites previously described under the classification section above: arytenoids, aryepiglottic folds, and position of the epiglottis. If the examination reveals redundant arytenoid tissue, foreshortened aryepiglottic folds, or a retro-positioned epiglottis and the patient meets requirements for severe clinical laryngomalacia, a supraglottoplasty should be strongly considered (Fig. 26.1).

Differential Diagnosis

Differential diagnosis for stridor in an infant includes infectious etiologies such as laryngotracheobronchitis (croup) or supraglottitis, congenital pharyngeal or laryngeal mass, congenital subglottic stenosis, tracheomalacia, bronchomalacia, retrognathia, airway hemangioma, supraglottic or hypopharyngeal mucous retention cysts, or unilateral or bilateral congenital vocal fold immobility.

Management

If the infant presents with mild or moderate laryngomalacia as previously defined, then conservative management with acid suppression and

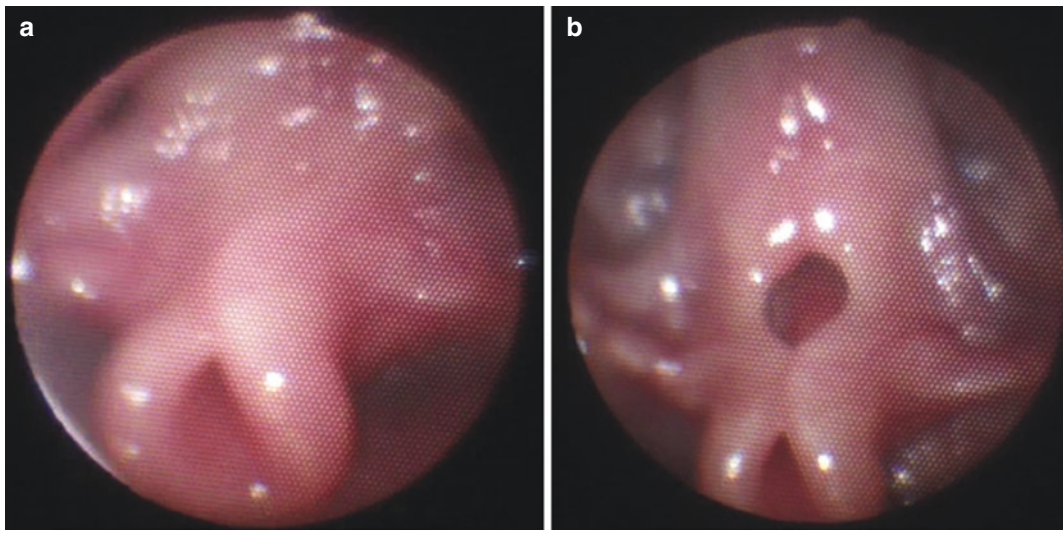

Fig. 26.1 Flexible fiberoptic in office evaluation: severe laryngomalacia with an omega-shaped epiglottis, foreshortened aryepiglottic folds, and redundant arytenoid tissue. The glottis is not visualized during inspiration (**a**) or expiration (**b**)

upright feeding positioning with a raised head of bed position may be trialed. Caregivers and primary care physicians should continue to monitor for signs of apnea, failure to thrive, or frequent coughing or choking with feeds. The patient should be reevaluated by a pediatric otolaryngologist if symptoms worsen.

If symptoms are within the severe category, surgical intervention should be strongly considered. Previously, tracheostomy had been the mainstay of treatment for severe symptoms. Tracheostomy may still be necessary in some cases. However, endoscopic supraglottoplasty has largely become the standard of care [4, 25].

Operative Approach: Supraglottoplasty

Indications

Surgical correction of laryngomalacia with supraglottoplasty is indicated in any patient who has been diagnosed with laryngomalacia at any of the three previously described anatomic subsites of the supraglottis and who has severe symptoms of laryngomalacia.

Key Aspects of the Consent Process

Informed consent should be obtained from caregivers after discussion of all risks, benefits, and alternatives to the procedure. The risks of the surgery may involve damage to surrounding structures such as lips, gums, tongue, and dentition. Furthermore, operating on the airway causes edema in the postoperative period, which may temporarily worsen respiratory symptoms. For this reason, patients are admitted for overnight observation and in some cases may require endotracheal tube placement or very rarely, tracheostomy. Laryngeal stenosis may result from scarring of opposing freshly cut mucosal surfaces. In some cases, this may require reoperation. Swallow function may either transiently or permanently worsen, especially with manipulation of the epiglottis [26].

Equipment

An infant Benjamin Lindholm laryngoscope is used for exposure. One port is connected to a light source and the other port is used for insufflation. The laryngoscope is connected to a Lewy arm and suspended using a Mayo stand. An operating microscope is needed as well as a full set of pediatric microlaryngeal instruments. If the laryngomalacia requires epiglottopexy, then a laryngeal needle driver and knot pusher are necessary. The most commonly used instruments are a microlaryngeal scissors and either a three or five laryngeal suction.

Drug-Induced Sleep Endoscopy

Drug-induced sleep endoscopy (DISE) is performed prior to supraglottoplasty to assess severity, confirm the offending anatomical location(s), and determine the extent of surgical resection necessary. A sleep-like plane of anesthesia is achieved using total intravenous anesthesia in order to maintain spontaneous ventilation and oxygenation. The patient is placed supine, and one spray of oxymetazoline is placed into each nasal cavity for decongestion. A face mask is used to deliver supplemental oxygen per anesthesia and a flexible fiberoptic nasopharyngolaryngoscopy is performed. The flexible fiberoptic scope is placed through the facemask CO_2 port and into the nasal cavity to maintain supplemental oxygen delivery throughout the procedure. The scope is used to examine the following: adenoid hypertrophy, tonsils, tongue base, position of the epiglottis, and if there is evidence of glottic obstruction with spontaneous ventilation, arytenoid mucosa, and if a ball-valve effect is observed with inspiration. Next, the same structures are examined with a jaw lift maneuver to observe if any improvement is visible on endoscopic scope exam. Performing this examination prior to supraglottoplasty helps determine the extent of arytenoid mucosa to trim, aryepiglottic fold to release, and whether to perform an epiglottis manipulation.

Steps

1. *Patient positioning***.** Anesthesia is maintained with total intravenous anesthesia (TIVA) with insufflation, spontaneous ventilation, and oxygenation. With this technique, the supraglottic structures are easily accessible for the surgeon, and the patient is adequately ventilated and oxygenated. The patient is positioned in a sniffing position with the head slightly flexed and the body extended at the atlanto-occipital joint. Moistened gauze is used to protect the gingiva in the edentulous infant or small child. A mouthguard may be used if the size of the mouth is adequate. Care must be taken to not rock the laryngoscope back while gaining exposure as this may place excessive pressure on alveolar ridge or the dentition and dislodge teeth.
2. *Exposure and suspension***.** The mouth is opened using a scissor technique with one finger placed on the posterior maxilla and the thumb placed on the posterior mandible. Next, the tongue is swept off to the left, and the Benjamin Lindholm is inserted into the right lingual gutter and advanced into the valleculae. The laryngoscope is pulled up and forward to expose the supraglottis and glottis and then suspended using the Lewy arm and Mayo stand (Fig. 26.2).

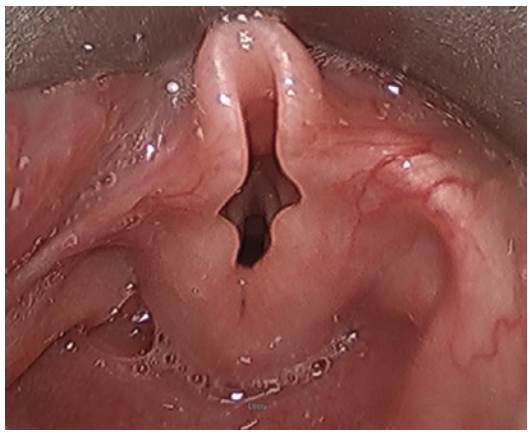

Fig. 26.2 Intraoperative view of laryngomalacia with foreshortened aryepiglottic folds and redundant arytenoid mucosa. Suspended via infant Benjamin Lindholm laryngoscope

3. *Release of aryepiglottic folds***.** We perform supraglottoplasty with cold steel microlaryngeal instruments. The aryepiglottic folds are visualized and are operated on if foreshortened and previously identified to be an issue on dynamic flexible laryngoscopy exam. An internal branch of the superior laryngeal nerve is identified just lateral to the aryepiglottic folds, and care is taken not to injure the nerve. A Bouchayer forcep is used to gently grasp the posterior aspect of the aryepiglottic fold and provide gentle countertraction in the posterior direction (Fig. 26.3a). This places the aryepiglottic fold on gentle tension. Next, microlaryngeal scissors are used to create a 2 mm releasing incision just anterior to the Bouchayer forcep (Fig. 26.3b). After this cut, the aryepiglottic fold and epiglottis pulls anteriorly, toward the vallecula. This is repeated on the other side (Fig. 26.3c).
4. *Trimming of excess arytenoid mucosa***.** The arytenoid mucosa is trimmed if it was previously visualized creating a ball-valve effect in the glottis on dynamic flexible fiberoptic laryngeal exam. A Bouchayer forcep is used to gently grasp the superficial aspect of the mucosa overlying the arytenoid complex (Fig. 26.3d). Care is taken to avoid mucosal injury or manipulation of the interarytenoid space in order to prevent scarring and laryngeal stenosis. Then, right- or left-facing microlaryngeal scissors are used to trim excessive epithelium from the superior surface of the corniculate process (Fig. 26.3e). A deep layer of mucosa should be preserved to prevent exposed cartilage (Fig. 26.3f).
5. *Epiglottopexy*. Epiglottopexy is performed if the epiglottis was retro-positioned and blocking the glottis during dynamic laryngeal exam. This is usually performed in conjunction with release of the aryepiglottic fold to promote further migration of the epiglottis anteriorly. However, it may be performed on its own in the absence of foreshortened aryepiglottic folds. The mucosal surface of the lingual side of the epiglottis and the mucosal surface of the valleculae must be freshened in

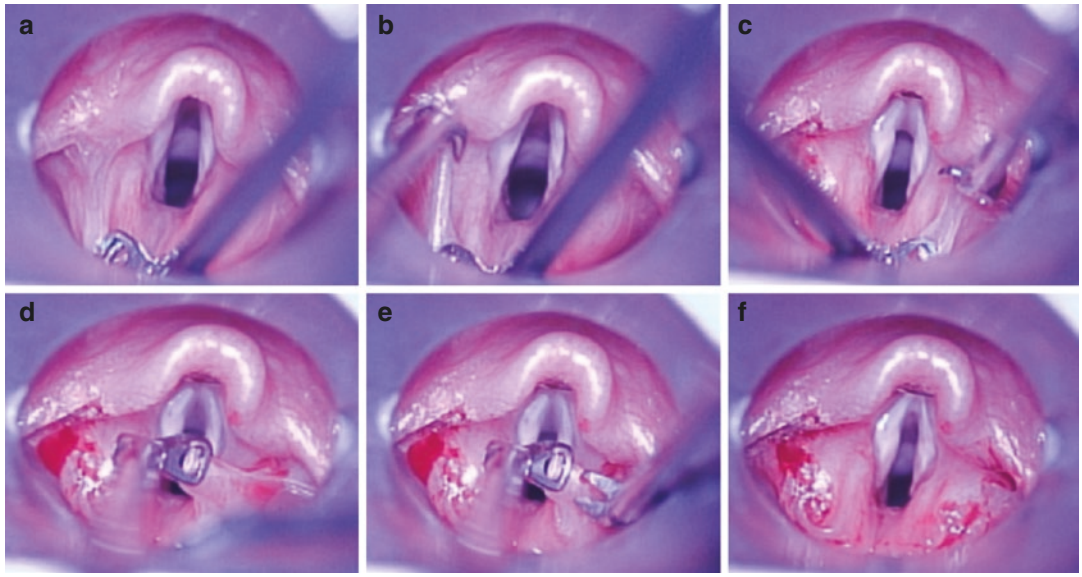

Fig. 26.3 Intraoperative view of supraglottoplasty. (**a**) Left aryepiglottic fold is grasped and retracted posteriorly to place on tension. (**b**) Microscissors are used to divide aryepiglottic fold anteriorly. (**c**) This is repeated on the contralateral side. (**d**) Mucosa overlying the arytenoid complex is retracted superiorly. (**e**) Microscissors are used to excise redundant mucosa. (**f**) View after bilateral aryepiglottic fold release and excision of mucosa overlying bilateral arytenoid complexes

order to promote fusion of the base of the two structures. A CO_2 laser is brought into the field and used to create a raw surface of the aforementioned epithelial surfaces. Laser safety materials including skin and eye protectants with moist towels and moist telfas over the eyes are used on the patient. All staff and observers must wear laser safety eyewear. FiO_2 should be lowered to <30% to reduce fire risk [27, 28]. Care should be taken to not ablate or create a deep cut through the epiglottis. Next, 3–5 vicryl sutures on an appropriately sized, curved needle are placed from the mucosa of the lingual aspect of the epiglottis to the tongue base/vallecula. Stitches are placed using a microlaryngeal needle driver, and knots are tied down using a laryngeal knot pusher. The purpose of the sutures is to create contact between the two newly exposed epithelial surfaces to promote permanent fusion.

6. *Hemostasis*. If necessary, hemostasis is achieved using a cotton ball or 0.5″ by 0.5″ pledget soaked in oxymetazoline. Epinephrine-soaked pledgets may also be used.

Postoperative Management and Follow-Up

Following supraglottoplasty, patients are monitored overnight for development of respiratory distress due to airway edema. Depending on the individual patient, observation may be on the general floor or in the intensive care unit. They are placed on dual acid suppression including a type 2 histamine blocker (e.g., ranitidine 5 mg/kg twice daily) and a proton-pump inhibitor (e.g., omeprazole 1 mg/kg once daily). The patient is continued on this for 3 months postoperatively. If the patient is found to have increased coughing, choking, or gagging with feeds, a swallow study may be necessary. This can be completed at any time, although, early in the postoperative period, swallow may be transiently worse secondary to decreased sensation. Additionally, the patient should adhere to a soft food diet for 1 week if they are old enough to consume solid foods. They are evaluated again at a 1 month follow-up appointment with otolaryngology and, at our institution, in conjunction with the speech-language pathologist.

Emerging Techniques of the Future

Various methods have been used in place of cold microlaryngeal instruments including CO_2 and thulium lasers [29] and the microdebrider [8]. The microdebrider-assisted technique may be used to release the aryepiglottic folds and remove redundant arytenoid mucosa [8, 26]. Likewise, the CO_2 or thulium laser may be used for the same purposes. Authors report that hemostasis is better with the thulium laser in comparison to the CO_2 laser [29, 30]. An additional technique that has been recently used is unilateral coblation of the arytenoid mucosa and suturing of the arytenoid mucosa laterally [31].

When comparing cold steel instrumentation to CO_2 laser, there was no surgical technique associated with increased supraglottoplasty failure [29]. Additionally, there were no differences in median time to feed postoperatively, postoperative pain, or improvement in breathing symptoms [29]. Further studies are warranted to elucidate differences in the new and emerging aforementioned supraglottoplasty techniques. These studies should also aim to break down supraglottoplasty techniques based on their three anatomic subsites.

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Laryngeal Cleft

27

Karthik Balakrishnan and Kari A. Krein

Overview

As has been made clear throughout this textbook, the larynx serves three key functions: voice, breathing, and swallowing. Laryngeal cleft is a congenital anomaly of the larynx that can impair two of these three processes; it may be entirely asymptomatic or may present with significant breathing and feeding problems. In severe cases, laryngeal cleft may cause life-threatening compromise of these functions requiring prompt intervention. This chapter reviews this condition in detail and presents our approach to its work-up and management. The reader should note that some controversy exists about specific aspects of laryngeal cleft diagnosis and management; we have discussed these issues in the body of the chapter.

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Definitions and Classifications

The simplest definition of a laryngeal cleft is the anatomic lack of separation between the airway and swallowing pathway in the posterior midline of the airway. In other words, the larynx and/or trachea are not adequately separated from the esophagus. This failure of separation should be distinguished from tracheoesophageal fistula. In the case of laryngeal cleft, also referred to as laryngotracheoesophageal cleft (LTEC), the abnormal connection between the airway and swallowing pathway begins between the arytenoid cartilages in the supraglottis and extends inferiorly as a continuous cleft to varying points more distal (Fig. 27.1). In contrast, tracheoesophageal fistula involves one or more focal connections between these pathways with normal separation proximal and distal to each fistula. The laryngeal structures are normal with a fistula communication between the esophagus and the trachea through the party wall.

While several classifications have been proposed for laryngeal clefts, the Benjamin-Inglis system [1] is most commonly used (Fig. 27.1). This system divides clefts into four categories of ascending severity based on the distal extent of the cleft. Type 1 clefts are restricted to the interarytenoid space and do not enter the cricoid cartilage; they generally involve the interarytenoid muscle and overlying mucosa in isolation. Anatomically, these can be defined as

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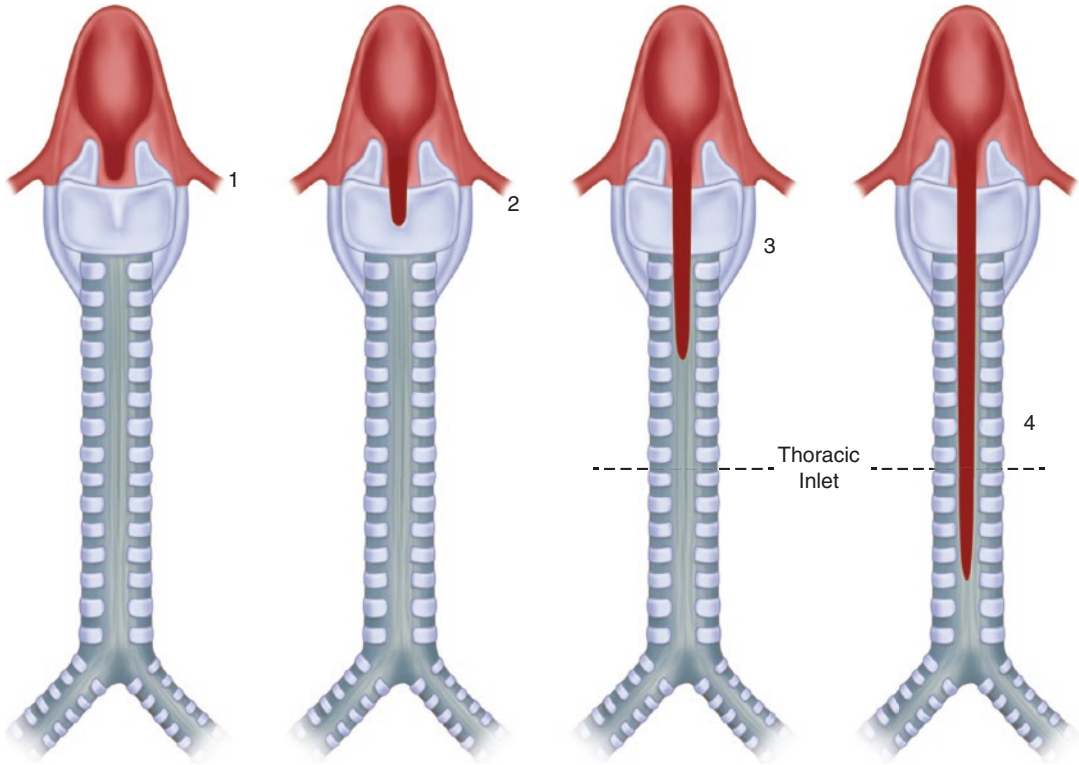

Fig. 27.1 Benjamin-Inglis classification of laryngotracheal clefts. Type 1 is a supraglottic interarytenoid cleft. Type 2 is a partial cricoid cleft. Type 3 extends to cervical trachea. Type 4 extends to thoracic trachea (From Benjamin and Inglis [1], with permission)

"supracricoid, interarytenoid" clefts and extend to or just below the true vocal folds [2]. A related and more controversial condition is the so-called deep interarytenoid notch, also known as a "type 0" laryngeal cleft; this involves an interarytenoid notch that is judged to be deeper than average but that does not extend to the level of the glottis. Diagnosis of this condition is quite subjective. However, it appears that treatment of this condition may improve symptoms of dysphagia [3, 4], suggesting that even a "deep notch" may have functional significance.

Type 2 clefts extend into the posterior cricoid cartilage but do not enter the infracricoid trachea (Fig. 27.2), while type 3 clefts span the posterior cricoid cartilage and enter the cervical trachea (Fig. 27.3). Type 4 clefts, which are the most severe category in the Benjamin-Inglis classification, extend into the thoracic trachea or beyond into a mainstem bronchus.

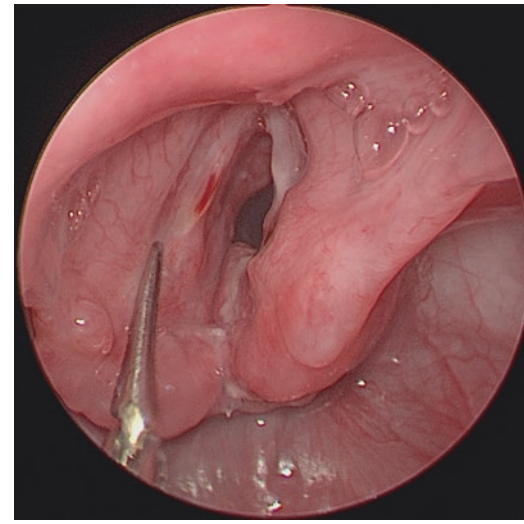

Fig. 27.2 Endoscopic view demonstrating a type 2 laryngeal cleft. The alligator forceps are distracting the left arytenoid cartilage to allow visualization of the cleft

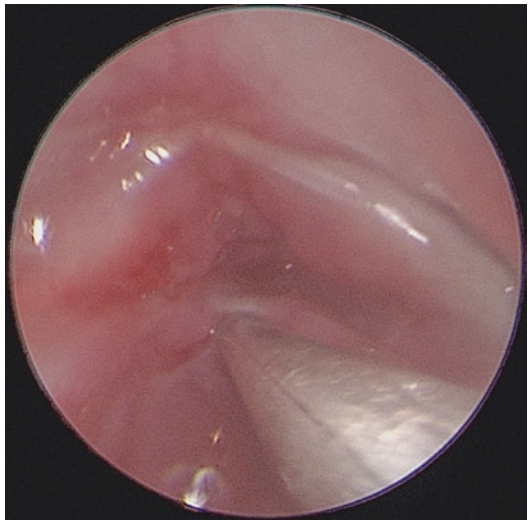

Fig. 27.3 Endoscopic view of an iatrogenic type 3 laryngeal cleft resulting from previous endoscopic posterior cricoid split. The metal probe is in the esophagus

Epidemiology and Associated Conditions

While experience suggests that laryngeal clefts are quite common, their actual incidence is not well-documented in the literature. One available estimate is 1 in 10,000 live births and 1.5% of all congenital laryngeal anomalies [5]. The proportion of syndromic laryngeal clefts is unknown. However, specific syndromes are associated with this anomaly. These include Opitz G/BBB (also known as Opitz-Frias), Pallister-Hall, VATER (vertebrae, anus, trachea, esophagus, renal)/VACTERL (vertebrae, anus, cardiac, tracheoesophageal fistula, renal, limb), and CHARGE (coloboma of the eye, heart defects, atresia of the choanae, retardation of growth and development, ear abnormalities) associations. All of these syndromes also include associations with other airway anomalies, particularly tracheoesophageal fistula [6]. Indeed, some authors suggest that every patient with tracheoesophageal fistula be evaluated for laryngeal cleft [7] regardless of whether the patient has a diagnosed syndrome, and we support this recommendation.

Pathophysiology

The embryogenesis of laryngeal clefts is unknown. In general, these lesions are thought to result from incomplete fusion of the posterior larynx.

Clinical Presentation

Laryngeal clefts vary greatly in clinical presentation, from asymptomatic lesions to those that cause life-threatening compromise of feeding and breathing in the immediate postnatal period. While each Benjamin-Inglis cleft type has not been clearly tied to specific symptoms, experience suggests that symptoms are typically worse in frequency and severity with increasingly distal cleft extent. Type 0 and type 1 clefts may be asymptomatic or may cause symptoms including coughing or choking with feeds, wet voice or wet cough during or after feeds, or recurrent respiratory infections. Our experience suggests that most children with these shallow clefts have normal swallow function unless another factor "tips them over" into aspiration. Examples of such factors include neurologic or neuromuscular disease and poor oral coordination during feeding. This experience is corroborated in the literature by examples of patients not presenting until they acquire other conditions during adulthood [8]. Some children may also have symptoms only when drinking thin liquids, drinking rapidly or with large boluses, or swallowing when they are distracted.

Differential Diagnosis

Work-up and management of laryngeal clefts depend most of all on the clinician's having some index of suspicion that this condition is present. Because the symptoms of laryngeal cleft overlap with multiple other conditions, and because laryngeal cleft may be very difficult to diagnose on in-office flexible laryngoscopy, this condition must be kept consciously in the differential diagnosis of

a patient with the symptoms described in the previous section. Other conditions that may be associated with similar clinical presentation are listed in Box 27.1.

Box 27.1: Differential Diagnosis for Laryngeal Cleft

- Reduced pharyngeal or laryngeal sensation
- Laryngomalacia
- Unilateral or bilateral vocal fold weakness or immobility
- Tracheoesophageal fistula or bronchoesophageal fistula
- Subglottic stenosis
- Nasal or upper airway obstruction (in infants)
- Neurologic causes of dysphagia including Chiari malformation and cranial neuropathies
- Any focal or systemic condition causing dysphagia
- Any cause of recurrent lower respiratory infections, wheezing, or persistent cough, including reflux, persistent bacterial bronchitis, and asthma

Speech-Language Pathologist Approach

Speech-language pathologists (SLPs) have been involved in the evaluation and management of feeding and swallowing disorders in pediatric populations for more than five decades. The prevalence of feeding/swallowing disorders in infants and children has increased as medical and surgical advancements are made including increase in survival of premature births, many resulting in complex medical conditions [9]. It is estimated that in an otherwise healthy pediatric population, the incidence of feeding related disorders is 25–45%, and in those with developmental disabilities, the incidence may be up to 80% [9]. This increased incidence requires more specialized methods of evaluation that are able to assess the anatomy and physiology of the swallowing mechanism that cannot be captured with a clinical swallow evaluation [10]. This includes a small portion of patients that may have dysphagia related to a laryngeal cleft [9].

Clinical Swallow Evaluation

Obtaining a thorough clinical history from parents/caregivers regarding symptoms as well as an examination of the oral mechanism for any evidence of anatomical abnormality or cranial nerve involvement is imperative when evaluating a child for feeding/swallowing issues. If clinical history includes coughing or choking during feeds, difficulty feeding in general, failure to thrive, "wet" or noisy breathing or difficulty breathing during or after feeds, and recurrent pulmonary infections, a laryngeal cleft should be in the differential diagnosis [11, 12]. An oral mechanism examination includes close examination of the face and oropharynx, observation for evidence of respiratory distress, and voice assessment which includes observing for breathiness, hoarseness, etc. that may indicate vocal fold involvement [13].

Instrumental Swallow Evaluation

Videofluoroscopic Swallow Study (VFSS)

In cases where a laryngeal cleft is suspected or included in the differential diagnosis, a pattern of penetration/aspiration that occurs during the swallow may be observed. The patient may or may not be symptomatic to these episodes. The swallow pattern may be well coordinated initially; however, if penetration/aspiration is occurring, the swallow may become increasingly discoordinated as the study continues due to increased difficulty trying to coordinate breathing and swallowing. Typically, there would not be any post-swallow residue or pooling of material in the pharynx after the swallow unless there are other diagnostic considerations.

Overall, according to Johnston et al. [11], a study that shows aspiration in an otherwise healthy child strongly correlates with an anatomic abnormality. They state that 75% of patients who have

a type 1 or type 2 laryngeal cleft will show aspiration on VFSS. However, it is important to note that in patients with intermittent symptoms, the study may appear normal without witnessed events of penetration/aspiration during the examination. Additionally, Miglani et al. [9] showed that patients who have penetration/aspiration and a laryngeal cleft also have a high rate of swallow dysfunction affecting all phases of the swallow. Finally, Johnston et al. [11] remind us that many patients may have multiple swallow studies done in the course of their work-up, and therefore, cumulative radiation exposure should be considered.

In addition to VFSS being helpful in identifying feeding difficulties and penetration/aspiration, it can also be a tool for monitoring patient progress during feeding therapy as well as after procedural interventions [9].

Fiberoptic Endoscopic Evaluation of Swallowing (FEES)

FEES findings in patients with laryngeal cleft are similar to those seen on VFSS. Many children will have a normal-appearing larynx on awake flexible endoscopy. In some cases, the cleft may become visible with abduction of the vocal folds during inspiration, though a high index of suspicion is needed, and the sensitivity and specificity of awake laryngoscopy for this diagnosis remain unknown. In deeper laryngeal clefts, redundancy of the mucosa lining the cleft may lead to visible prolapse of posterior laryngeal soft tissues into the glottis aperture with inspiration, with associated stridor. Some clinicians look for the "ram sign," in which the shape of the redundant mucosa suggests the curling horns of a ram, though this finding is much more visible on rigid airway endoscopy under anesthesia.

During the swallow evaluation portion of FEES, some patients may demonstrate visible laryngeal penetration through the interarytenoid region. This finding is of course very suggestive of a laryngeal cleft. Most patients, however, will either not demonstrate penetration/aspiration or will have the typical "whiteout" during the pharyngeal swallow itself but have visible swallowed material within the larynx, suggesting entry of this material during the "whiteout." Indirect clues to aspiration, such as wet cough, wet voice, or wet breathing, may suggest penetration or aspiration, but they are not specific to the diagnosis of laryngeal cleft. Signs of aspiration may be more evident with thinner textures, more rapid flow of material into the pharynx, or with mixed textures.

Management

Management of swallowing problems in patients with laryngeal cleft who demonstrate penetration or aspiration will vary depending on the severity of symptoms and need for multidisciplinary interventions.

Management options may include surgical intervention for correction of anatomical defect, feeding therapy and/or diet modifications, and medical management. For purposes of this section, feeding therapy and/or diet modifications will be the focus, although it is difficult to completely separate them from each other as it is more likely that a component of all of them will be involved.

Miglani et al. [9] describe conservative management including diet modifications, possibly to include thicker liquids in addition to medical management if laryngeal clefts are small and consequences are considered mild. The specifics of thickening vary between centers and clinicians, with some providers thickening feeds in very young infants and others avoiding this strategy until the patient is at least 1 year of age. More aggressive diet modifications or limitations in addition to medical management and surgical intervention may be required for more severe aspiration events with more significant medical sequelae. Depending on severity, oral intake may not be safe regardless of diet modifications, and other routes for nutrition may need to be considered.

Treatment goals for any child with laryngeal cleft and associated swallowing issues are to achieve the least restrictive diet that allows for continued safe and efficient oral intake. Age limitations may play a role in what consistencies are tested and what consistencies are recommended. Feeding therapy, outside of modifications to diet, may also be indicated to address behavioral feeding issues and skills

(for both caregiver and child) that may reduce risk of aspiration while allowing for continued, efficient oral intake. This may include pacing and positioning strategies. Altering the flow of liquids via a slow flow nipple may also be an alternative for infants who aspirate thin liquids but are too young to thicken feeds.

In summary, the goal of any treatment plan should be to support adequate and safe nutrition; swallowing function can improve simply due to patient growth and maturation over time [11]. Diet advancement or changes in recommendations are determined by the patient's progress as well as repeated instrumental and clinical swallowing evaluations as deemed necessary [13].

Otolaryngologist Approach

History

The medical history should begin at the patient's birth, with questions covering mode of delivery, need for resuscitation, intubation or respiratory support, APGAR scores, and initial feeding strategies and outcomes. While these questions may not relate directly to laryngeal cleft, they help to develop a thorough differential diagnosis for children with feeding and/or breathing problems. The otolaryngologist should then follow the child's development forward to the present, asking about immunization status, neurologic and motor developmental milestones, and feeding development. The clinician should also inquire about recurrent or persistent lower respiratory infections such as pneumonia or bronchitis, including frequency, most recent episode, associated hospitalizations, and need for antibiotic therapy. Specific questions can be tailored to the patient's history based on the differential diagnosis listed earlier.

With regard to feeding, useful questions include current mode of feeding, type of bottle/nipple/cup used, and current textures being taken. The provider should ask about need for non-oral feeding such as nasogastric tube, gastrostomy tube, or parenteral nutrition. Any previous clinical or instrumental swallow evaluations should be reviewed, ideally by examining the actual images, and the provider should elicit any history of prior airway interventions such as intubation, tracheostomy, or other airway operations.

Our practice is also to ask about recurrent croup; clinical experience suggests that subglottic inflammation may be related to aspiration of secretions or refluxed material, which may be exacerbated by a laryngeal cleft.

Physical Examination

The otolaryngologist should perform a thorough head and neck and cranial nerve examination, particularly given the potential association of syndromic diagnoses with laryngeal cleft. In addition, chest auscultation and auscultation at the mouth may reveal subtle stridor or abnormal lower airway sounds.

In-Office Endoscopic Assessment

Flexible nasolaryngoscopy should be a routine part of the examination in patients with suspected laryngeal cleft. It serves two main purposes. First, it is a key part of FEES as described earlier. Second, it allows the otolaryngologist to rule out other diagnostic entities such as laryngomalacia, pooled secretions suggesting impaired sensation, and cranial neuropathies affecting the swallow or vocal fold motion. In some patients, the flexible endoscope may also be passed beyond the vocal folds to assess the subglottis or trachea. Our practice is to limit this to school-age children and older, as long as they are cooperative and can tolerate a weight-based dose of inhaled topical lidocaine to minimize risk of laryngospasm. However, other authors have successfully performed in-office tracheoscopy in younger children [14]. It is important to avoid the administration of any topical anesthetic into the nose or airway prior to FEES to avoid impairing sensation during swallow evaluation.

Flexible in-office endoscopy does not allow adequate visualization of the interarytenoid anatomy to rule laryngeal cleft in or out; indeed, flexible endoscopy by a skilled practitioner only detects this condition 69% of the time when the patient is under general anesthesia [15].

Operative Endoscopic Assessment

The reference standard for diagnosis of laryngeal cleft is rigid airway endoscopy with the patient under general anesthesia. In this setting, the clinician can use a rigid probe or small alligator forceps to gently push the arytenoid cartilages apart and palpate the interarytenoid area. This examination will demonstrate two key findings. First, it shows whether there is a laryngeal cleft and the depth of this cleft relative to the glottis (Fig. 27.2). Palpation and separation of the arytenoids are essential to accurately grade any cleft (Fig. 27.4). Second, it allows the clinician to palpate the midline posterior cricoid plate to determine whether there is any submucous extension of the cleft in or through the cricoid. In both cases, suspension laryngoscopy may be beneficial because it allows the surgeon to elevate the larynx and visualize the esophageal inlet and posterior aspect of the cricoid as well as the endolaryngeal anatomy, permitting a more accurate assessment of the cleft's anatomy.

In the case of recurrent cleft after prior repair or iatrogenic cleft after prior posterior cricoid split or posterior graft laryngotracheoplasty, these steps are even more valuable. The normal anatomy of the posterior larynx may be distorted or the cleft may be off midline, which may affect planning of any repair. Figure 27.3 demonstrates an iatrogenic type 3 laryngeal cleft as a result of an endoscopic posterior cricoid split performed at another institution. Figure 27.5a, b demonstrates an iatrogenic type 2 laryngeal cleft as a result of a failed endoscopic posterior graft at another institution; Fig. 27.6 demonstrates a scar band at the inferior end of a failed type 2 cleft repair at another institution.

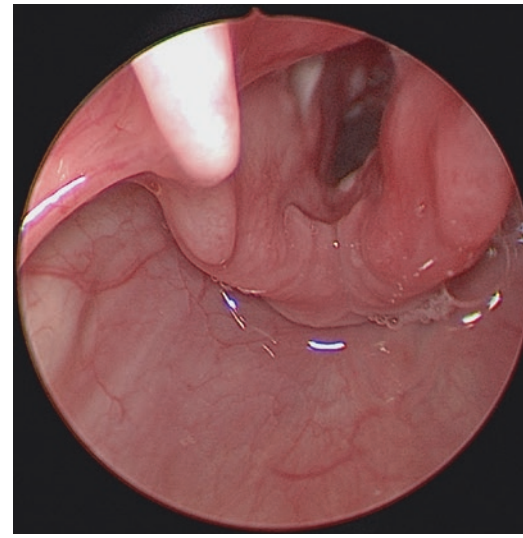

Fig. 27.4 Deceptively normal-appearing interarytenoid area that revealed a type 2 laryngeal cleft upon subsequent palpation. This figure demonstrates the need for palpation and the inadequacy of visual inspection alone in the diagnosis of laryngeal cleft

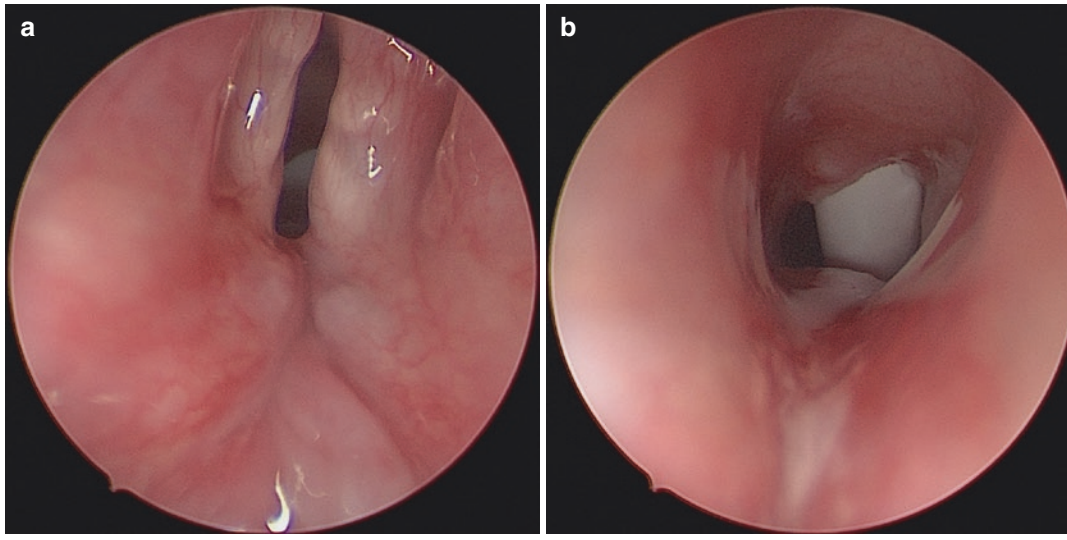

Fig. 27.5 Endoscopic view of type 2 laryngeal cleft at the (**a**) supraglottis and (**b**) subglottis

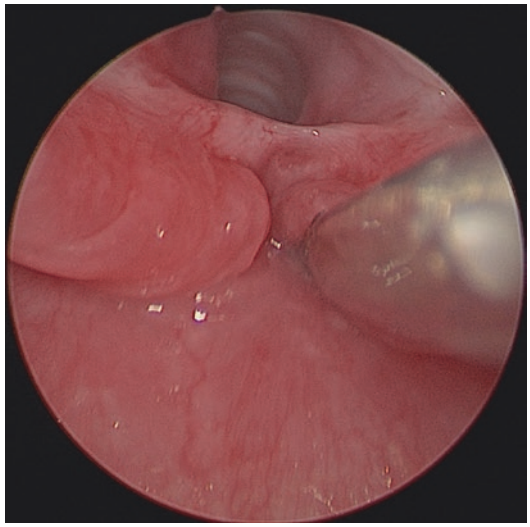

Fig. 27.6 Endoscopic view of type 2 cleft after previous failed repair

Other Studies

Our practice is generally to perform these assessments in conjunction with our pulmonology colleagues to obtain information about the lower airways [15]. Specifically, we are interested in signs of lower airway inflammation and bacterial infection on endoscopy and bronchoalveolar lavage; these suggest either episodic or ongoing contamination of the lower airways that may support a decision to repair any diagnosed laryngeal cleft. We also often send lavage specimens for airway pepsin testing; a positive result suggests aspiration of refluxed material. This finding again would push us to consider repairing a cleft.

Imaging studies may also be coordinated with trips to the operating room. We prefer noncontrast computed tomography (CT) of the chest with inspiratory and expiratory breath holds, done immediately before arrival in the operating room and under the same anesthetic. This study allows localization of any consolidation and may guide location of bronchoalveolar lavage. It also may demonstrate signs of chronic lung inflammation or injury consistent with ongoing aspiration. Finally, comparison of the inspiratory and expiratory views may demonstrate air trapping and other relevant pathology. The drawbacks of radiation exposure and more time under anesthesia must be weighed against the information provided by CT; we typically reserve this study for patients in whom the clinical relevance of any suspected cleft is unclear or in whom other concomitant diagnoses are suspected.

Management

As described earlier, many patients can be successfully managed with dietary modifications. These modifications are typically instituted to buy time while the patient's swallow matures, but they can also be a long-term solution in some children. Ongoing care of patients receiving thickened feeds is best done in collaboration with a dietician to ensure that patients are receiving appropriate calorie and nutrient intake.

If dietary modifications are not an option or are unsuccessful, operative treatment of laryngeal cleft is often useful. A variety of approaches have been described, including endoscopic injection augmentation, endoscopic layered or mass closure suture repair, and open repair. This chapter will describe each of these approaches.

Operative Approach: Endoscopic Injection

Patient Selection

Endoscopic injection of laryngeal cleft is best suited for type 0 and type 1 clefts. Published survey data suggest that more otolaryngologists manage type 0 clefts in the same manner as type 1 clefts [16]. This approach is well-suited to patients with adequate cardiopulmonary reserve to breathe spontaneously for several minutes while under general anesthesia. Patients should have reasonable laryngeal exposure on direct laryngoscopy. Intermittent intubation is possible and reasonable.

Key Aspects of the Consent Process

As with any operation, caregivers should be counseled about the risks of general anesthesia. Specific to this procedure, the surgeon should

mention the risks of direct and suspension laryngoscopy. Depending on the surgeon's preferred injection material, allergic and infectious risks associated with that material should be mentioned. Risks of over-injection including airway obstruction, stridor, and need for temporary intubation or tracheostomy should be mentioned. The surgeon should mention clearly that this is a temporary solution intended to buy time as the child's swallow matures and that further procedures may be necessary. Finally, given the complexity of swallowing, the clinician should make clear the risk of failing to improve swallow or aspiration symptoms [17].

Equipment

Key equipment includes:

- Age appropriate laryngoscope and rigid Hopkins rod telescopes (usually 0 and 70°) with associated light cords and imaging tower.
- Suspension laryngoscope of appropriate size for the patient, with suspension apparatus. Our preference is the Lindholm laryngoscope, though better visualization is achievable in some patients using either the Parsons laryngoscope or Phillips laryngoscope.
- Laryngeal spreaders (optional).
- Operating microscope (optional).
- Injection device, selected by the surgeon depending on the material to be injected.
- Device to spray topical medications onto the larynx.
- 0.5×3 in. cotton pledgets soaked in oxymetazoline.

Steps

1. Patient positioning: The patient is positioned supine with head at the top of the table. The table is turned 90° with the patient's left side toward the anesthesiology provider. No shoulder roll is used. If the patient's head tends to roll to one side or the other, a rolled towel or gel donut can be placed around the head.
2. Laryngeal exposure: If direct laryngoscopy and photodocumentation of the cleft has been done recently, we proceed directly to placement of the Lindholm laryngoscope over a dental guard. If not, standard direct laryngoscopy and tracheoscopy are first performed to assess the overall airway anatomy. The Lindholm laryngoscope is suspended until adequate exposure of the posterior larynx is achieved. The cleft is again palpated and photodocumented.
3. Laryngeal spreaders: If the surgeon wishes to use laryngeal spreaders to better expose the interarytenoid space, we typically use the smallest available spreaders in young children. These are placed through the Lindholm and upside down so that the handles face upward and the distal tips of the spreaders are placed in the laryngeal ventricles (Fig. 27.7). Once the spreader is distended, the handles are looped with the rubber band which is then looped around the suspension apparatus of the laryngoscope to hold the spreaders up out of the way.
4. Visualization: Depending on the surgeon's preference, either a rigid telescope or the operating microscope can be used to visualize the cleft. If the operating microscope was used, we generally set focal length to 400 mm.
5. Injection: Once adequate visualization has been obtained, the injector is brought in through the Lindholm. The needle is placed into the interarytenoid tissues at the distal

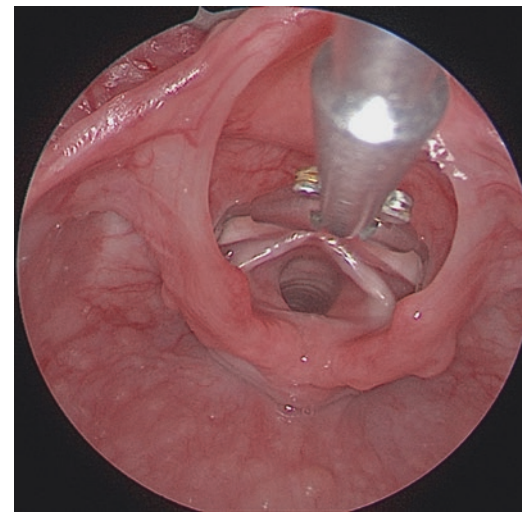

Fig. 27.7 Endoscopic view of type 1 laryngeal cleft with laryngeal spreader in place

apex of the cleft, which is usually in the posterior midline. Under constant visualization, injection is performed until the apex of the cleft is bulked upward toward the arytenoid peaks as much as possible. Care is taken not to create a large endolaryngeal bulge near the posterior glottic commissure, as this may lead to airway obstruction. If additional injection is needed to bulk up the cleft and narrow it further, further injection sites maybe selected along the medial aspect of each arytenoid.

6. Photodocumentation: A final set of images is obtained after injection is complete.
7. Hemostasis: Topical 1:50,000 epinephrine is sprayed onto the larynx to minimize edema and achieve hemostasis at the injection sites. If extensive bleeding is encountered, gentle pressure with oxymetazoline-soaked pledgets will generally stop it.

Operative Approach: Endoscopic Suture Repair

Patient Selection

Endoscopic repair of laryngeal cleft is best suited for type 0 through type 2 clefts, with some shallow type 3 clefts also being amenable depending on the patient's ease of laryngeal exposure. Otherwise, selection criteria are similar to those for endoscopic injection. Patients with concomitant posterior glottic or subglottic stenosis who might benefit from endoscopic posterior graft laryngotracheoplasty can have that procedure done simultaneously via the same approach.

Key Aspects of the Consent Process

These include the same aspects as for endoscopic injection above. Additional risks include airway fire if the laser is used and dehiscence of the repair. Supraglottic stenosis should be mentioned as a risk, but despite extensive experience with endoscopic cleft repair, we have never seen it and are not aware of any colleagues who have seen it after this procedure. Deeper clefts that dehisce at their distal end may result in a tracheoesophageal fistula requiring reintervention. We counsel caregivers that the patient may have transient stridor for 12–24 hours postoperatively and that the swallow may worsen temporarily before it is improved. Our experience suggests that this transient worsening of swallow occurs in 3–5% of patients. No published data reflect this, though we are in the process of formally studying this question.

Equipment

As with endoscopic injection above, additional care equipment includes:

- Operating microscope (required).
- Laryngeal spreaders (required).
- Equipment to demucosalize the medial aspect of the arytenoids and edges of the cleft. This can be done either with laser or with cold microlaryngeal instruments.
- If laser is to be used, then the appropriate laser quit equipment and micromanipulator are needed. Our preference is the Lumenis Acublade CO_2 laser device, set at 10 W continuous with 1.5 mm circle.
- Endoscopic needle driver and knot pusher.
- Appropriate suture. Our preference is 4-0 PDS on an RB-1 taper needle. For a type 1 cleft, three to four sutures will likely be required.
- Microlaryngeal instruments including right and left grasping forceps and right and left curved scissors.
- Laryngeal injection equipment (optional).

Steps

1. Patient positioning: As described for injection above.
2. Laryngeal exposure: As described for injection above. It is critical to obtain an adequate view of the entire larynx including epiglottis, aryepiglottic folds, arytenoids, interarytenoid space, and postcricoid. In some cases, the latter could be achieved by placing a blunt suction behind the cricoid and gently elevating the larynx anteriorly.
3. Laryngeal spreaders: Extremely useful for this procedure. Placed as described for injection above (Fig. 27.7).
4. Visualization: The operating microscope provides hands-free visualization and depth perception, both of which are essential to perform this procedure. Focal length is set to 400 mm.

5. Laser: If laser is to be used, the appropriate generator and micromanipulator should be attached to the microscope and correct function confirmed. The patient should have wet eye pads placed and be draped thoroughly with wet towels to minimize the risk of airway fire and burns. The surgeon should keep in mind that any exposed tissue including teeth can be damaged by laser light. Inspired oxygen fraction should be 30% or less during any use of the laser. The laser should be tested away from the patient to confirm correct function and good alignment with the aiming beam and good function of any shutter function. All personnel in the operating room should have appropriate eye protection depending on the wavelength of laser used.
6. Demucosalization: While we do not inject the mucosa prior to this step, some surgeons prefer to do so to achieve some hydrodissection and hemostasis. Either laser or laryngeal microscissors can be used to remove mucosa from the edges of the cleft. This should include the medial aspects of both arytenoids, and particular attention should be paid to the distal apex of the cleft. Any mucosa left here will prevent healing and may lead to a small fistula. Our preference is to use the laser because it is also hemostatic and allows a very superficial removal of mucosa while preserving as much submucosal tissue as possible for suturing; it is also very fast. Some surgeons have raised concerns about impaired healing from laser damage. We have never seen this to be an issue (Fig. 27.8).
7. Removal of charred tissue and hemostasis: If the laser is used, the charred mucosa is gently wiped away using an oxymetazoline-soaked pledget. The same can be used to achieve hemostasis if cold instruments are used.
8. Suturing: 4-0 PDS on RB-1 taper needle is used, though other surgeons prefer Vicryl. The needle is often overbent slightly to allow easier manipulation through the laryngoscope and in the small pediatric larynx. We prefer a mass closure technique, though other surgeons prefer a layered technique. The mass closure technique has been shown to be equally effective and faster [18]. The distal end of the suture is clamped with a hemostat. The needle is grasped approximately halfway along its length in the endoscopic needle driver and passed through the laryngoscope. The sutures placed such that the knot ends up on the posterior aspect of the larynx. For a right-handed surgeon, the first throw enters just at the free mucosal edge at the posterior aspect of the previously demucosalized area and passes immediately adjacent to the medial aspect of the arytenoid cartilage, exiting just deep to the anterior-free mucosal edge. The return throw reverses this order, entering at the anterior-free mucosal edge, again passing immediately adjacent to the medial aspect of the contralateral arytenoid, and exiting at the posterior free mucosal edge. Care is taken to achieve precise matching of craniocaudal level for the two throws. We typically tie six knots, ensuring that the knots are placed down tight and square. The suture is cut immediately above the knot; our experience has been that longer suture tails lead to irritation of the posterior pharyngeal wall and associated granulation. The first suture is placed immediately at the inferior apex of the cleft, and subsequent sutures are separated by about 1 mm each.

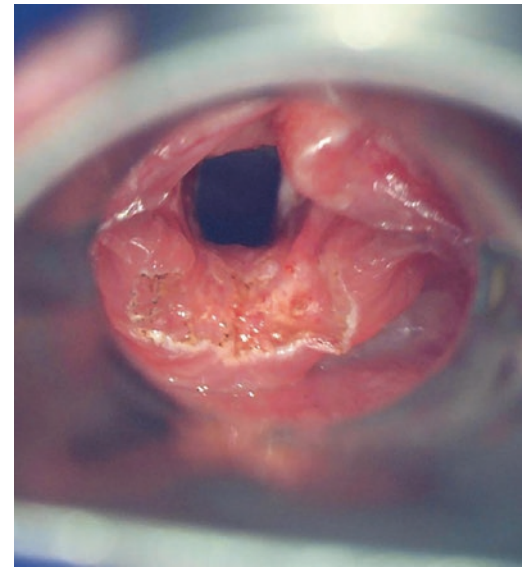

Fig. 27.8 Endoscopic view of type 1 laryngeal cleft after CO_2 laser demucosalization

Care must be taken to tie each suture down toward the previous stitch to avoid any loose areas of the closure that may lead to fistulization. The topmost suture should be passed through the cuneiform/corniculate complex because the mucosa at the arytenoid peak is quite thin and tears easily, leading to dehiscence of this stitch (Figs. 27.9, 27.10a, b, and 27.11).

9. Supraglottoplasty: Once the cleft has been sutured, we invariably find that the arytenoids are tipped slightly forward and the aryepiglottic folds appear tight. We create a small releasing incision in each aryepiglottic fold immediately posterior to the epiglottis, preserving an intact band of mucosa between this area and the cleft repair (Fig. 27.9). We typically inject these lateral

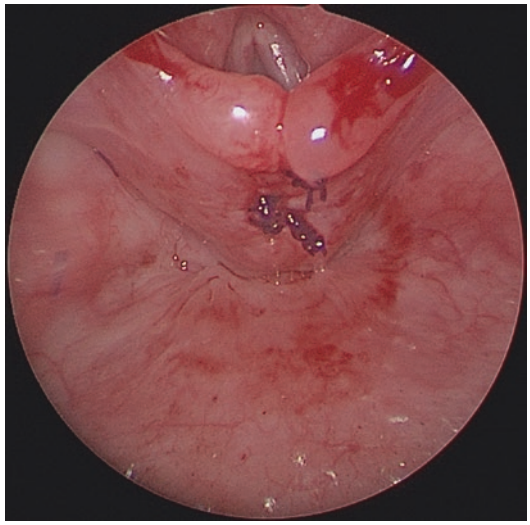

Fig. 27.9 Endoscopic view of repaired type 1 laryngeal cleft

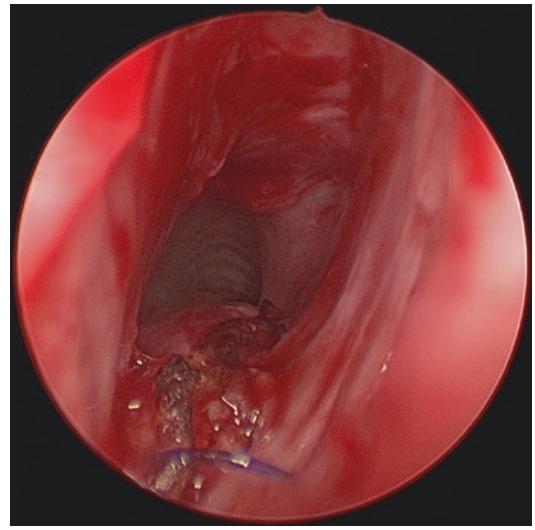

Fig. 27.11 Endoscopic view of repaired type 3 laryngeal cleft

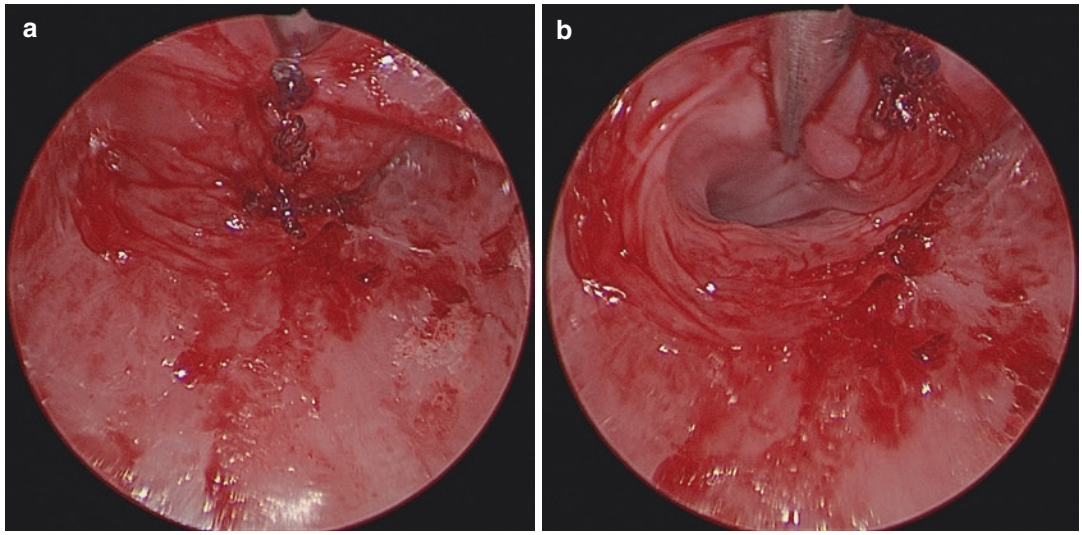

Fig. 27.10 Endoscopic view of repaired type 2 laryngeal cleft (**a**) supraglottis showing four of eight sutures placed, (**b**) esophageal inlet and larynx elevated with suction to show distal sutures

incisions with a small amount of triamcinolone 40 mg/ml to minimize the risk of any scarring or stenosis, taking care not to inject anywhere near the cleft repair and to suction any steroid that may run onto the repair. We have not seen that this is associated with any increased rate of dehiscence of the cleft repair.

10. Photodocumentation: 0 and 70° telescopes are used to photodocument the repair and examine the trachea to ensure that suctioning of any blood or secretions is not required. It is worth examining the endolarynx to confirm that the glottic aperture has not been narrowed by a stitch inadvertently catching and medializing an arytenoid cartilage.
11. Hemostasis: Topical 1:50,000 epinephrine is sprayed onto the larynx to minimize edema and achieve hemostasis at the operative site. If extensive bleeding is encountered, gentle pressure with oxymetazoline-soaked pledgets will generally stop it.

Postoperative Management and Follow-Up After Endoscopic Intervention

Due to the structure of our hospital, we typically admit these patients to the pediatric intensive care unit for monitoring. In some cases we have admitted them to the general pediatric ward, a practice that is supported by at least one study [19]. Patients leave the operating room extubated and breathing spontaneously. We allow the intensive care team to utilize racemic epinephrine, heliox, and noninvasive positive-pressure ventilation as needed, though it is rare that any of these interventions is necessary. If intubation is required postoperatively, which has not occurred at our institution thus far, we plan for intubation by an otolaryngologist, ideally over a rigid telescope to minimize the risk of disrupting the repair.

Patients are observed overnight. The following morning, we request occupational therapy evaluation of swallow function at the bedside in case there has been a worsening of swallow function requiring temporary dietary modification (at other institutions, this could be done by speech-language pathology). Again, in our experience this is about 3–5% of patients. Once a discharge feeding plan is established, patients are discharged home. If no modification is required, we request that patients continue their preoperative diet until follow-up, with the additional request that they avoid hard or crunchy foods for 3 weeks. Follow-up is done around 6 weeks postoperatively with in-office flexible laryngoscopy. Depending on whether FEES or VFSS showed aspiration or penetration preoperatively, that study is repeated at this time as well. None of these follow-up practices are supported by the literature; indeed, there is little published data establishing a best practice follow-up plan, though some consensus survey data and a single patient-based study exist [16, 20]. Our practice is to not repeat VFSS or FEES after the initial postoperative study unless swallowing concerns persist or new concerns develop.

Operative Approach: Open Repair of Laryngotracheal Cleft

Patient Selection

Open repair is typically best suited for patients with type 3 or 4 clefts or significant comorbidities or anatomic restrictions that prevent endoscopic repair. Patients with significant distortion of anatomy due to previous surgery may also benefit from open repair, as do those with multiple previous operations or significant scarring who might need associated posterior graft laryngotracheoplasty or layered repair of their cleft.

Key Aspects of the Consent Process

Caregivers should be counseled that open repair of laryngeal cleft carries risks of infection and bleeding as with any open operation. Open repair often requires complete laryngofissure [18], with associated risks of voice compromise, glottic web, and instability of the laryngeal skeleton. If laryngofissure is required, a stent or keel may also be necessary for some time after surgery, with subsequent operative procedures to remove these devices. Temporary tracheostomy may be

required to allow adequate ventilation and oxygenation during the repair. As with endoscopic repair, distal dehiscence may lead to tracheoesophageal fistula requiring further intervention. Deep type 4 clefts involving the distal half of the trachea may require sternotomy and intraoperative support with cardiopulmonary bypass or extracorporeal membrane oxygenation, and the risks of these interventions should also be discussed carefully. These will typically be done in collaboration with the cardiovascular surgeon, who also should ideally meet the caregivers prior to surgery. Type 4 clefts requiring cricotracheal separation also carry some risk of injury to the recurrent laryngeal nerves with associated vocal fold weakness or immobility.

Equipment

For clefts restricted to the upper half of the trachea and above, setup is essentially the same as open laryngotracheoplasty or other open transcervical airway reconstruction in a child. Deeper clefts requiring a sternotomy or thoracotomy will require appropriate equipment for those approaches.

Steps

The steps of this repair will vary significantly depending on whether a transcervical, transthoracic, or transsternal approach is required, on whether tracheostomy is required, on the depth of the cleft, and on the age and size of the patient. Our general approach for transcervical repairs is to perform partial or complete laryngofissure for clefts through type 3. For type 4 clefts limited to the upper half of the trachea, we prefer to use cricotracheal separation via a neck incision [21]. For transsternal approaches on cardiopulmonary bypass or extracorporeal membrane oxygenation support, we again use cricotracheal separation.

Postoperative Management and Follow-Up

Postoperative management follow-up will depend on the approach and extent of the operation performed.

Emerging Techniques/Concepts

Treatment approaches for type 1–3 laryngeal clefts have evolved over time, from open repair using laryngofissure, to layered endoscopic closure, to the rapid and technically straightforward endoscopic closure technique described here. A few themes have developed in recent years that are likely to affect the care of these patients further:

1. The use of thickened feeds, while inconsistent across institutions, has certainly also had a salutary effect on the care of these patients. The optimal age to start thickening and the optimal degree of thickening remain unknown. Institutions in the United States vary widely in their practice, with the youngest age for thickening ranging from newborn to 1 year. Similarly, definitions of thickened textures, and choice of thickener, have not been standardized. Early efforts to achieve standardization have been pursued at various centers, but we still lack a broader consensus accounting for potential benefits, adverse effects, costs, and uniform terminology.
2. The use of the surgical robot in laryngeal cleft repair was first described in 2007 [22]. Since that time, small series have been described that include type 1 through 3 laryngeal clefts [23]. Data are lacking on whether this technique adds value for the patient in terms of reduced operative time, reduced perioperative morbidity, or improved swallowing outcomes when compared to standard endoscopic techniques, and whether these potential benefits justify any added costs of care associated with use of the robot. While laryngeal cleft and airway surgery in general remain off-label uses of the surgical robot, it seems likely that robot-assisted airway surgery will continue to expand in scope and indications. Formal comparative effective studies will be necessary to evaluate whether this trend is beneficial to patients.
3. Injection laryngoplasty, as discussed earlier, is becoming more popular. Further research is needed to determine which patients are most

likely to benefit from this option, which material is best for injection in terms of effect and duration while minimizing adverse reactions, and how many times injection should be tried before performing a formal suture repair.

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Paradoxical Vocal Fold Motion

28

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Overview

Paradoxical vocal fold motion (PVFM) is a laryngeal disorder characterized by sudden onset of difficulty inhaling that is not typically accompanied by hoarseness or change in voice quality. In the infant or very young child, medical management, diet change, and positioning may be sufficient to manage the symptoms. In the school-age and older child, a combination of behavioral and medical management is used once the diagnosis is confirmed. PVFM is often mistaken for other disorders; therefore, proper differential diagnosis is key to management success.

This chapter reviews the various labels that are used to describe this clinical entity, the differential diagnoses that must be ruled out prior to behavioral intervention, typical medical assessment and management pathways that are pursued, and the basic behavioral intervention approach.

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Definitions

There are many different terms for PVFM as can be seen in Table 28.1. Speech language pathologists and otolaryngologists generally use the term paradoxical vocal fold motion (PVFM) or paradoxical vocal cord motion (PVCM) to describe this condition. Medical specialists in sports medicine, pulmonology, and allergy often prefer the term vocal cord dysfunction (VCD). Given the myriad of laryngeal conditions that are diagnosed and treated by otolaryngologists and speech-language pathologists, use of the term VCD lacks the specificity that PVFM provides.

Table 28.1 Alternative labels for paradoxical vocal fold motion (PVFM)

Munchausen's stridor	Paradoxical vocal fold movement (PVFM)
Functional airway obstruction	Factitious asthma
Laryngeal spasm mimicking bronchial asthma	Paradoxical vocal cord dysfunction presenting as asthma
Stridor caused by vocal cord malfunction associated with emotional factors	Episodic laryngeal dyskinesia
Psychogenic stridor	Functional laryngeal stridor
Episodic paroxysmal laryngospasm	Irritable larynx syndrome (ILS)
Paradoxical vocal fold motion (PVFM)	Paradoxical vocal cord motion (PVCM)
Vocal cord dysfunction (VCD)	Factitious allergic disease

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That being said, it requires acknowledgment that PVFM is a behavioral disorder of the vocal folds that is distinct from neurologic etiologies and airway obstructive conditions that can mimic the same behavior.

Epidemiology

Pediatric PVFM is not common. That being said, accurate diagnosis often takes months and sometimes years due to lack of familiarity with the disorder among medical professionals and misdiagnosis as asthma. The exact incidence and prevalence of PVFM in the general population are unknown, though systematic literature reviews suggest that roughly a third of cases affect the pediatric population. The average age of onset in children is 14 years, but it has been described in infants as young as 1-week-old [1–3].

In terms of demographics, multiple studies report that there is a 2:1–3:1 female-to-male predominance regardless of age. PVFM has also been associated with significant social stressors. In a case series of 22 juvenile patients, Powell et al. [4] found that 55% either participated in competitive organized sports or were heavily involved in other extracurricular activities. In another prospective survey study, Liao et al. [5] found that out of 39 subjects between 12 and 17 years of age, many were regularly involved in competitive activities such as track (30%), swimming (17%), and cheerleading or dancing (15%). Additionally, the majority of study participants were high-achieving students, either making straight-A's (46.2%) or A's and B's (41%).

Pathophysiology

Theoretically, PVFM is often classified along a continuum of disorders that fall under what is called irritable larynx syndrome (ILS; [6]), with chronic throat clearing as the most mild and laryngospasm as the most severe on the continuum of disorders. ILS is believed to develop secondary to irritation to the laryngeal structure that is often multifactorial in nature. Given that all individuals experience some postnasal drip, reflux, and cough, it is only when a threshold of tolerance is passed that laryngeal stridor may occur. The ILS theoretical construct hypothesized that central neuroplastic changes to the laryngeal afferent receptor pathways occur secondary to repeated upper airway exposure or a single overwhelming upper airway exposure to an irritant(s). This hypothesis is supported by the work of Eric Kandel, who received a Nobel Prize for his work in a sea slug model demonstrating the neuroplasticity of the respiratory gill to repeated exposure to noxious agents, with eventual respiratory gill response to agents that the slug did not initially perceive as noxious. Given that the biological role of the larynx is to protect the lower airway, laryngeal adductory behavior in response to repeated or overwhelming exposure to laryngeal irritants is reasonable.

The primary etiology for the development of paradoxical vocal fold motion behavior is described by Mathers-Schmidt [7] as following into one of three primary groups: upper airway sensitivity to irritants (extraesophageal reflux or allergens), neurological, or psychological conditions. For the infant and very young child, it would be unlikely that a psychological condition could trigger the events; therefore the child should be carefully evaluated for probable environmental irritants, allergies, and reflux. Neurological conditions of which PVFM may be a clinical sign or symptom, for example, cerebral palsy or brainstem compression, should be ruled out. The neurological variation of paradoxical vocal fold motion behavior is distinct in its clinical presentation as will be described below.

Presentation

Infants and children with PVFM that is not due to a neurologic etiology will present with discrete, sudden-onset breathing attacks characterized by a primary difficulty with inhalation. In some children difficulty exhaling may accompany the inspiratory stridor. Not all children produce laryngeal stridor, and this symptom is not required to achieve an accurate diagnosis of

PVFM. The predominance of inspiratory difficulty is one distinction of PVFM from asthma. There is not typically any loss of voice during a PVFM attack, making this clinical presentation distinct from laryngeal edema secondary to allergy which can also arise suddenly. Children will not experience oxygen desaturation during a breathing attack, and if the child loses consciousness as the PVFM attack progresses, the child's larynx should relax open and a patent airway will be reestablished.

PVFM attacks can occur during the day with or without activity and can also occur at night. Pediatric PVFM events that occur at night will generally occur around the same time of the night with a sudden wakening with difficulty inhaling. Nocturnal PVFM events are often attributed to extraesophageal reflux events that occur at night. Daytime attacks can occur at any time of the day and may be directly related to mealtimes, exposure to certain allergens, odors, or cigarette smoke.

The neurological version of this behavior is very rare and is distinct from typical PVFM attacks in that these children will experience persistent difficulty with inhalation during their waking hours and will have no inspiratory difficulty during sleeping. The neurologic variant is not characterized by sudden, episodic difficulty with inhalation. The neurologic variant of PVFM has been attributed to four primary etiologies as described by Maschka et al. [8]: brainstem compression, cortical or upper motor neuron injury, nuclear or lower motor neuron injury, and movement disorder. Persistent difficulty with inspiration during waking hours that is secondary to any of the conditions just listed will not be amenable to behavioral intervention, and these children should be referred to the appropriate medical specialist for medical management.

Differential Diagnoses

The differential diagnosis for paradoxical vocal fold motion requires the consideration of many other clinical presentations that are not amenable to behavioral intervention and may warrant medical or surgical management. Paradoxical vocal fold behavior secondary to a neurological etiology is not typically helped by behavioral intervention, and these patients should be referred for medical management [8].

Most of the typical differential diagnoses that require consideration during the assessment process are summarized in Table 28.2. PVFM can also co-occur with many of the conditions listed in Table 28.2, and the authors have clinical experience with patients who have been concurrently diagnosed with three or more of the conditions included in Table 28.2. Clinicians working with upper airway breathing disorders need to take care to acknowledge that a given patient can have laryngeal edema secondary to allergens, panic attack, asthma, and PVFM. Even the young child can tell that these diagnoses result in different onset and experiences of breathing difficulty. It is up to the clinician to help the child untangle the differing symptom profile so that the appropriate intervention is applied. Referrals for PVFM are often made directly from sports medicine, pulmonary, and allergy healthcare providers to speech-language pathologists who specialize in the care of PVFM. SLPs should take care to advocate for inclusion of otolaryngologists in the care of these children.

Speech-Language Pathologist Approach

The role of the SLP in the evaluation and treatment of PVFM differs in many ways from pediatric voice disorders in general. For the infant or child with PVFM, voice is typically not affected; therefore the usual voice acoustic, aerodynamic, and perceptual assessments that are often used for baseline and outcomes evidence are not employed with this population. Laryngeal visualization is paramount to rule out extrathoracic obstruction and other airway conditions for which behavioral intervention is contraindicated. The role of the SLP is sometimes questioned in the treatment of this disorder that does not fit within the well-recognized scope of communication and swallowing disorders. The SLP is the most appropriate professional for treatment delivery in this population given the extensive knowledge of the anatomy

Table 28.2 Pediatric differential diagnoses symptom comparison

Diagnosis	PVFM/VCD	Asthma	Laryngeal edema	Adductor laryngeal breathing dystonia	Exercise induced laryngomalacia	Tracheomalacia	Fixed airway obstruction (stenosis)
Airway noise	Inhalation – *stridor*	Exhalation – *wheezing*	Inspiration or biphasic – *stridor*	Inspiration – *stridor*	Inspiration – *stridor*	Expiration – *stridor*	Biphasic – *stridor*
Origin of noise	Glottis (vocal folds)	Lower airway (bronchi)	Glottis (vocal folds)	Glottis (vocal folds)	Supraglottic (e.g., arytenoids, epiglottis)	Intrathoracic trachea	Any level – glottis, subglottis or trachea
Patient symptoms	Episodic throat tightness accompanied by difficulty inhaling; no loss of voice	Chest tightness with difficulty exhaling; no loss of voice Can wake from sleep	Throat closure and complete loss of voice	Persistent difficulty inhaling during waking hours only; no difficulty during sleep; extreme fatigue Often in context of systemic neurologic disease (e.g., multisystem atrophy)	Throat tightness and inspiratory stridor only during peak exertion; ADLs typically not affected	Shortness of breath and expiratory stridor with exertion Brassy cough even when well	Tightness during inspiration and expiration with or without any voice change; severity may range from affecting ADLs to only when exerting self
Beta-agonist response	No	Yes	No	No	No	No	No

ADL activities of daily living

and physiology of the laryngeal and respiratory systems. Additionally, the SLP is best trained to determine the extent to which a swallowing impairment may be involved and to develop a behavioral treatment plan that both trains and generalizes the breathing recovery method.

The assessment process from the point of view of the SLP relies more heavily on the medical history in general and a highly detailed description of the nature of the problem. The degree to which the SLP and the medical team can obtain a detailed account of the presenting problem will play directly into development of a patient-specific approach for medical and behavioral intervention. Failure to attend to the patient-specific aspects of this disorder will likely result in reduced ability to resolve the symptoms in a timely fashion. Given that this disorder causes the perception of air hunger with associated fear and anxiety for both the patient and caregivers, it is paramount that an accurate diagnosis be made quickly and behavioral/medical intervention be conducted expeditiously.

History

The case history of the infant or child will require a detailed account of birth history, developmental history (gross and fine motor, speech, language, and cognition), breathing and feeding development from birth, and any medical conditions for which the child has been assessed and treated. Given the most probable triggers for PVFM in the pediatric population, it is vital that the child be assessed and treated for all of the following prior to referral to the SLP: extrathoracic obstruction, pulmonary function, allergies, and extraesophageal reflux. It can be frustrating for the SLP when a child is referred for assessment and treatment for PVFM, and the requisite medical assessments have not been completed prior to referral. Failure to do due diligence to rule in or out the likely differential diagnoses (Table 28.2) can delay appropriate diagnoses and treatment for those children for whom behavioral intervention is not appropriate. Case history questions that are useful are included in Table 28.3.

Table 28.3 Pediatric case history questions

Behavioral	Is the primary difficulty breathing in, breathing out, or both?
	Is there a noise when the breathing attack happens?
	Where does the noise come from? The chest or the throat?
	Is there any tightness when the breathing attack happens? Where is the tightness, in the chest or the throat?
	Does anything help the breathing problem?
	Does anything make the breathing attack worse?
	How often do the attacks occur?
	How long does the attack last?
	If the attack resolves, is it likely to come back?
	Is there a voice change or loss of voice when the attacks happen?
	Are there known triggers for the breathing attacks?
Environmental	Does the child live with animals in the house?
	Was the child exposed to any new environments or new construction at the time of onset of the breathing problem?
	Does change in the weather or movement from one environment to another, e.g., going from hot outdoor environment to air conditioned space, trigger the breathing problem?
Medical	Has the child been diagnosed with
	Allergies by an allergist? Any history of laryngeal edema secondary to food or environmental allergies?
	Asthma by a pulmonologist? If asthma medications have been prescribed, is the medication being taken as prescribed?
	Reflux? If reflux medication was prescribed previously, what was the dosing schedule?

Laryngeal Visualization

Visualization of the laryngeal structures during resting breathing and during a PVFM event is diagnostically valuable to determine if the child has isolated PVFM or if it is one aspect of other disorders that may co-occur. Laryngeal visualization may be completed by SLPs depending

on individual practice patterns, regional clinical practice patterns, and state licensure laws. The role of the SLP with regard to laryngeal visualization is different than that of the otolaryngologist who is responsible for diagnosing laryngeal and upper airway pathology. The SLP may use laryngeal imaging for both observation of paradoxical vocal fold motion during the assessment process and for biofeedback during the treatment process.

When imaging the laryngeal behavior of a patient referred for probable PVFM, the imaging is best accomplished with flexible endoscopy without topical anesthetic (to avoid triggering a PVFM event) for the following breathing tasks:

- Tidal breathing – observe the glottal aperture during inhalation and exhalation. A 10–20% lateral adduction of the arytenoid cartilages during expiration is typical for individuals with diagnosed asthma. When observing an infant or toddler, observation of potential supraglottic tissue collapse during resting breathing may help identify laryngomalacia.
- Maximum inhalation – for the preschool, school-age, and teenage child, ask for a deep inhalation through the mouth to observe for any glottal adduction and/or stridor.
- Maximum exhalation – for the preschool, school-age, and teenage child, ask for a deep exhalation through the mouth to observe for any glottal adduction. Moderate arytenoid adduction during a maximal exhalation may be secondary to a diagnosis of asthma.
- Nasal sniff – while the child watches on the monitor, ask them to sniff in deeply through the nose so that they can observe how the vocal folds widely abduct for a more patent airway.
- Train the recovery breathing technique with the endoscope in place. The benefit of the visual biofeedback increases confidence when using recovery breathing technique when a breathing attack seems imminent.

Treatment

The behavioral component for PVFM in children will vary depending on the age and developmental ability of the child as well as other concomitant medical conditions. Treatment can be broken down into three primary developmental levels: infant and toddler, preschool and early school-age, and older school-age and teenage. The specific considerations for each group are as follows:

Infant and Toddler

PVFM in infants and toddlers is not common and will likely fall into the irritant-induced etiology, occurring secondary to reflux or environmental allergies. Because of the very young age and cognitive development of this group of patients, the primary treatment will be medical management of the triggers for the PVFM attacks. SLPs will play a role in counseling caregivers about upright positioning of the child after meals to mitigate reflux of stomach contents into the upper airway and limiting reflux inducing snacks or foods before sleeping or high intensity physical activity. With the very young child, it is important to regularly monitor the responsiveness to treatment and then determine if any medical management for identified triggers can be discontinued once the breathing attacks are completely resolved for a period of time.

Preschool and Early School-Age

The young child who is able to follow directions but is too young to apply the breathing recovery techniques independently will benefit from direct behavioral therapy with a SLP as long as the caregiver is included in the training. For this age group, children can learn the breathing recovery technique but are not cognitively mature enough to recognize when a breathing attack is imminent and apply the technique independently. Caregivers can be trained to help with at-home breathing recovery practice and then guide the child through the technique if a breathing attack appears imminent. Caregivers will also be tasked with making sure that medical management of probable triggers is completed.

Older School-Age and Teenager

For children who are cognitively mature enough to follow through with the breathing recovery technique without adult supervision and can

also identify the pre-attack warning signs that a PVFM attack is imminent, the breathing recovery training can proceed as it would with an adult. The basic breathing recovery training program will be described below. For a detailed description of the breathing recovery technique, the reader is referred to Sandage and Zelazny [9].

Breathing Recovery Technique

The breathing recovery approach has three primary components, the last of which is the actual recovery method for maintaining airway patency during a PVFM attack: training body awareness, lower thoracic breathing, and the recovery breathing technique. Training body awareness as a precursor to the second and third components should not be overlooked. The goal of improved body awareness is to train the child to connect with the physical perceptions of their body and airway so that they can better recognize any and all physiological sensations that precede PVFM attack and serve as warning signs. Recognition of the physiological sensations that precede throat tightness and stridor will help the child learn to avoid the PVFM event altogether. Body awareness can be trained by progressive relaxation techniques or mindfulness strategies that train sustained attention to the body.

Training lower thoracic breathing is intended to help eliminate upper torso and neck tension that can accompany the experience of air hunger that occurs with PVFM attacks. Use of a mirror during breathing instruction to eliminate elevation of the shoulders and clavicular breathing can be helpful. The child should then be instructed to inhale into their rib cage, feeling the sides of their rib cage and their back expanding with every inhalation. This work can also include expansion of the abdomen, but it should be acknowledged that more pulmonary volume can be inspired with expansion of the thoracic cavity than distention of the abdomen.

Once the two primary stages of training are completed, train the recovery breathing technique. This technique requires that the child quickly inhale via a fast nasal sniff or a fast oral "sip" with the lips pursed. The nasal sniff is preferred, but if the child experiences nasal congestion, the oral "sip" can suffice. Clinicians treating individuals with PVFM need to train the rapid, hard sniff of air without clavicular breathing and direction of the inspired air into the lower thoracic space. At the completion of the inhaled deep, rapid sniff, the child should then be taught to exhale as completely as possible using a "sh" or "f" sound. This latter approach will help maintain laryngeal abduction during the expiratory process.

It is highly recommended that the breathing recovery be trained during the initial assessment appointment so that the child can experience relief from the breathing attacks prior the follow-up visit. Schedule a follow therapy appointment within 1 week from the initial evaluation, and ask the caregivers or the older child to keep track of the following data to assess improvement:

- Number of attacks per day
- Length of each attack
- Number of rescue breathing cycles required before resolution of the PVFM attacks
- Rescue inhaler use per day

Improvement in therapy can be gauged by reduction in the length of the attacks, reduction of the overall number of attacks, reduction in the number of rescue breathing cycles required for each attack, reduction/elimination of rescue inhaler, and complete resolution of the PVFM attacks.

Otolaryngologist Approach

History

Otolaryngologists often do not have the privilege of seeing patients during an acute episode of PVFM; thus patient history becomes paramount. Individuals often describe episodic "difficulty breathing," "noisy breathing," or a feeling that their "throat is swelling or closing." Often patients and their families will misuse the term "wheezing," making the differentiation between asthma and PVFM more challenging. Clinicians should redirect the patient and family to avoid

medical terminology and focus on describing the symptoms experienced. Secondary complaints may include cough, tightness in the chest or throat, or difficulty voicing during the episodes. Importantly, patients are asymptomatic between episodes.

Common triggers for PVFM include strong odors, strenuous exercise, and stress. Asthma medications such as inhaled β-agonists or corticosteroids typically do not improve symptoms, and may even aggravate them. Reflux and allergies may also play a role as potential laryngeal irritants. Clinicians should ask about the patient's personal and family history of allergies, atopy, and asthma as well as history of heartburn, belching, or dyspepsia. Previous surgical and intubation history or other medical comorbidities are also important.

A single episode of classic PVFM tends to be short and resolve with removal of the irritant (e.g., rest in the case of exercise-induced) and time. However, episodes can be quite frightening to the patient and caregivers resulting in emergency room visits, intubation, or even tracheotomy if not correctly identified.

Exam

In the otolaryngologist's office, between episodes, patients should not have any noisy breathing. They may however appear anxious and stressed with a tight or hunched shoulder and neck posture. Nasal congestion, boggy turbinates, allergic shiners, and oropharyngeal cobblestoning may be indicative of an allergic component.

Instrumented Assessment

Laryngoscopy

Visualization of the vocal folds via flexible nasolaryngoscopy (FNL) while the patient is symptomatic is the gold standard for PVFM. FNL is also obligatory to exclude the other diagnoses including fixed airway obstruction, exercise-induced laryngomalacia, and vocal fold movement impairment. During FNL the use of topical anesthetics or sedatives may affect vocal fold movement and should be avoided or used with caution. The classical findings described by Morris and Christopher [10] are complete adduction of the vocal folds during inspiration with formation of a small posterior diamond-shaped glottal gap (Fig. 28.1), but PVFM should also be suspected in a patient with more than a 50% closure of the vocal folds on inspiration.

Often, symptoms must be provoked in order to observe the paradoxic vocal fold movement. Previous literature has reported that movement consistent with PVFM can be identified in 100% of symptomatic patients and 55–60% of asymptomatic patients [11, 12]. In the setup

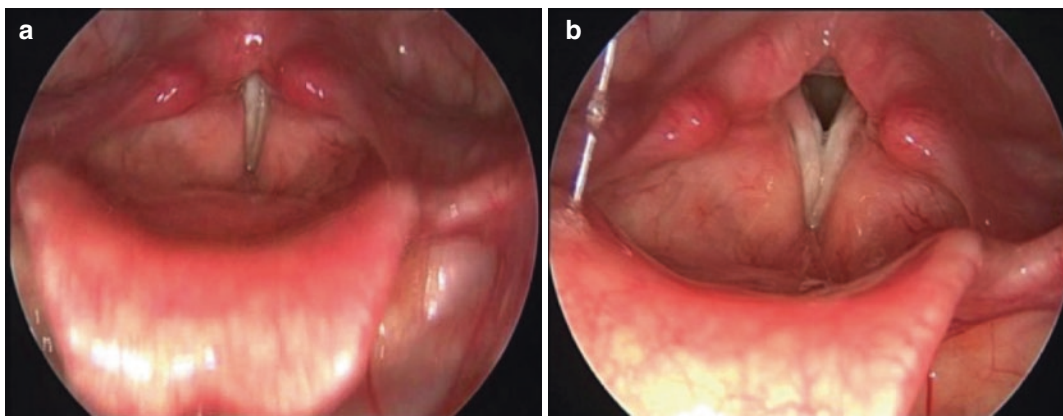

Fig. 28.1 Glottis during an episode of PVFM. (**a**) Adduction and (**b**) abduction with diamond-shaped posterior glottic gap visible

used by Heimdal et al. [13], individuals exercised to exhaustion on a treadmill while attached to a fully equipped ergo-spirometry unit and a flexible nasolaryngoscope linked to a video camera and sound recorder.

If the patient is able to undergo an exercise challenge, combined continuous laryngoscopy and spirometry with exercise is the most accurate test for diagnosis. However, most otolaryngologists do not have access to a full exercise laryngoscopy lab. Thus, clinicians can have patients run or climb stairs, to trigger an episode prior to laryngoscopy.

Pulmonary Function Tests

Pulmonary function testing (PFT's) measures lung volume and rate of air flow. When asymptomatic, patients with PVFM should have normal flow-volume loops; however, during an episode flattening of the inspiratory loop is seen (Fig. 28.2).

Methacholine Challenge

Methacholine is a short-acting cholinergic agonist that is frequently used as a diagnostic aid for asthma. Patients with asthma are more sensitive to the effects of methacholine and other bronchoconstrictors and will demonstrate changes to pulmonary function at lower doses than non-asthmatic individuals. Methacholine challenge testing involves the administration of increasing concentrations of the drug followed by PFTs. Typically, a decrease in the forced expiratory volume in 1 s (FEV1) of greater than 20% from baseline signifies a diagnosis of asthma. A negative methacholine challenge has excellent negative predictive value, and in a patient with chronic dyspnea, chest tightness, wheezing, and/or cough should raise suspicion for alternate diagnoses, including PVFM. However, it should be noted that methacholine is an irritant that has also been shown to induce PVFM symptoms that result in a false-positive result.

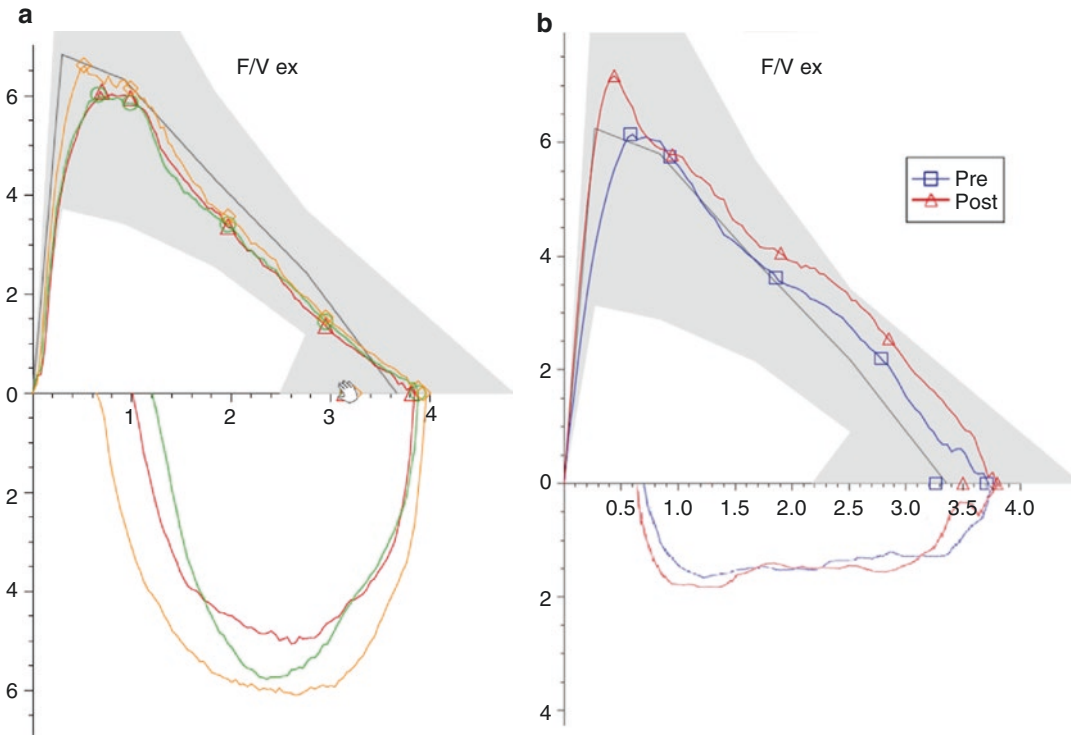

Fig. 28.2 Pulmonary function test flow volume loops. (**a**) Normal. (**b**) Episode of paradoxical vocal fold motion (PVFM). Note the flattening of the inspiratory loop. Blue is pre- and red is post-bronchodilator

Imaging

Imaging has limited utility in the work-up of PVFM; however, a chest x-ray may be performed if pulmonary involvement is suspected, and presence of hyperinflation and peribronchial thickening is suggestive of asthma. Airway fluoroscopy has also been used to rule out other levels of airway obstruction.

Differential Diagnosis

There are many other airway disorders that may mimic PVFM. The differential includes:

- Asthma
- Episodic laryngeal breathing disorders
  - Laryngospasm/spasmodic croup
  - Exercise-induced laryngomalacia/intermittent arytenoid region prolapse
- Dystonia
- Vocal fold movement impairment
- Obstructive airway lesions
- Laryngeal edema

Asthma

Of these conditions, asthma is the most important to distinguish from PVFM. PVFM is often misdiagnosed as asthma, with significant repercussions for patients. A retrospective analysis performed by Traister et al. [14] found that individuals with PVFM who were wrongly diagnosed and treated for asthma had significantly more health care and medication use than those who were not, suggesting that misdiagnosis is a major source of morbidity.

Compared to asthma, PVFM is characterized by inspiratory difficulty rather than expiratory, and this is often reflected in the patient's history and PFT's. A negative methacholine challenge and lack of improvement with the use of bronchodilators also strongly suggest a diagnosis of PVFM over asthma. Diagnostic aids such as the Pittsburgh VCD Index and the Paradoxical Vocal Fold Movement Disorder Screening Questionnaire (PVFMD-SQ) can aid in differentiating between the two disorders but are not validated in the pediatric population [14, 15]. It should be noted that a diagnosis of PVFM does not exclude asthma or vice versa; about half of patients with PVFM also have asthma [16] and may reflect underlying airway irritability.

Laryngospasm/Spasmodic Croup

Patients who experience predominately nighttime symptoms, or symptoms when supine, may be experiencing laryngospasm related to reflux. Reflux may also be a trigger for classic PVFM symptoms. 24 h pH/impedance testing with symptom correlation can clarify the role of reflux. Younger children may have "spasmodic croup" which is a barky cough and stridor which occurs at night without a clear infectious cause (unlike infectious/viral croup). Spasmodic croup may be an expression of irritant (reflux or allergy)-induced laryngospasm.

Exercise-Induced Laryngomalacia

Exercise-induced laryngomalacia is characterized by supraglottic collapse during peak work capacity rather than during submaximal exercise, rest, or recovery [17]. Recovery is usually very rapid, and obstruction is significantly decreased within half a minute to a minute of recovery. Comparatively, exercise-induced PVFM often occurs when the patient is beginning exercise or transitioning, and the timing of the obstruction and recovery is different. The difference between the two conditions is best visualized through continuous exercise laryngoscopy.

Dystonia

Laryngeal adductor breathing dystonias, which may occur with multiple system atrophy or dysautonomia, present with vocal cord adduction that mimics PVFM on laryngoscopy but does not respond well to respiratory retraining. Patients with dystonias often have more continuous symptoms with a worse baseline compared to PVFM. Also, patients may have other neurological findings, such as blepharospasm or involuntary tongue or jaw movements, or have a history of prior brain injury or birth complications.

Vocal Fold Movement Impairment

Bilateral vocal fold paralysis may mimic the laryngeal obstruction seen in PVFM, but symptoms lack a trigger, are present at rest, and worsen with submaximal exercise. Patients with vocal fold movement impairment (VFMI) may have a history of cardiac surgery or other neurologic symptoms. Additionally, compared to in PVFM, where voice quality is minimally affected, in VFMI, the patient may have an asthenic (weak, breathy, or rough) voice.

Obstructive Airway Lesions

An obstructive lesion such as a mass or stenosis in the respiratory tract may cause stridor, but the symptomatology is generally continuous and progressive rather than episodic. Vocal fold lesions such as recurrent laryngeal papillomatosis will typically have significant associated voice change. Laryngotracheal stenosis will present with biphasic stridor rather than pure inspiratory or expiratory stridor.

Laryngeal Edema

Laryngeal edema is a swelling of the laryngeal mucosal tissue in response to an irritant or allergen that can cause throat closure and a dyspneic attack. Patients experiencing laryngeal edema may have greater alteration in or loss of their voice and more signs of allergic response than with PVFM. Visualization via laryngoscopy can help differentiate the conditions, but the priority should be on securing the patient's airway.

Management

Acute

The stridor and respiratory distress seen with an acute episode of PVFM can be quite frightening to the patients, their families, bystanders, and health-care workers. There have been reports of patients who have required intubation or even tracheotomy in this setting. Until the diagnosis is clear, clinicians should adhere to the basics: airway, breathing, and circulation. Of note, most patients with PVFM will not desaturate, despite impressive stridor.

If possible, laryngoscopy during an acute episode is ideal. If PVFM is suspected, coughing or panting may help break an acute episode. Otherwise, the physician can reassure the patient and guide them through metered breathing (as described above).

Therapy

The best treatments for patients with PVFM are respiratory retraining with a qualified speech-language pathologist as described previously. For adolescent athletes, Sullivan et al. [18] reported that 95% of patients were able to control symptoms after working with a speech-language pathologist. Maturo et al. [19] found that respiratory retraining as an initial treatment was effective for about 68% of pediatric patients overall and for 56% of those with any symptoms at rest.

For patients with underlying anxiety or stress and symptoms, psychology or psychiatry can be an essential adjunct [19]. Some children also benefit from psychotherapy, sports psychology, or hypnotherapy. Alternatively, the use of biofeedback, where operant conditioning is applied to gain control of involuntary muscle contractions, is an emerging technique. Warnes and Allen [20] evaluated the use of electromyography biofeedback in a 16-year-old girl. After giving the study participant a visual representation of her muscle tension and training her to relax using the feedback, she was able to reduce overall baseline muscle tension by 60% with a corresponding reduction in episodes of respiratory distress and chest pain.

Pharmacotherapy

Various drugs have been used to try to treat PVFM:

- Benzodiazepines – anxiolytic and sedative effects may function in terminating acute symptoms that are triggered or exacerbated by stress/anxiety.
- Heliox – gaseous mixture of oxygen and helium that reduces airway turbulence and eliminates respiratory noise to provide short-term relief from dyspnea.
- Inhaled lidocaine – may help break the aberrant sensory feedback loop.

- Anticholinergics/Ipratropium bromide – anticholinergic aerosol use prior to activity was shown to prevent exercise-induced symptoms in a retrospective study of 49 patients [21] potentially by inhibiting stimulation of the vagus nerve.
- Proton pump inhibitors (PPIs) or H2 blockers – used to treat reflux symptoms if reflux is felt to be trigger.

Operative Approach

Surgery is very rarely indicated for patients with PVFM, especially in the pediatric population, and should only be considered if multiple other therapies have failed and the patient's quality of life is negatively affected by their symptoms. The main surgical approaches used are injection of botulinum toxin, suture lateralization, and tracheotomy.

Botulinum toxin A prevents release of acetylcholine from the presynaptic nerve terminal, resulting in chemical denervation lasting approximately 3 months. When injected into the thyroarytenoid muscle, it prevents complete adduction of the vocal fold, allowing air passage even during an episode of PVFM. In younger children, injection typically has to be done in the operating room under anesthesia.

Suture lateralization, which prevents vocal fold adduction, can be done endoscopically and is theoretically reversible. However, in the author's experience, the suture often erodes through the vocal fold resulting in a "slow cordotomy" [22].

Tracheotomy is only used as a last resort for patients who have a history of multiple hospital admissions and intubations and experience severe respiratory distress during episodes. Tracheotomy may be indicated in severe dystonias in particular.

Emerging and Evolving Techniques of the Future

New treatments and diagnostic modalities are in development for PVFM, as our understanding of the disorder improves. Currently, there is some focus on the link between dysregulation of the sympathetic and parasympathetic nervous systems and PVFM, as PVFM-like symptoms can be observed in some dysautonomias. Patients with mild cases of dysautonomia with PVFM may have some chronic underlying shortness of breath and core muscle deconditioning and may be benefited by core strengthening exercises. In a case study released in 2017, a 23-year-old female had improvement of symptoms after using a multifactorial approach that included a yoga program with isometric and diaphragmatic breathing exercises [23]. In addition to this, Honey et al. [24] recently described PVFM symptoms secondary to possible compression of a vagus nerve by a looping of the posterior inferior cerebellar artery in three cases. In each case, microvascular decompression relieved all symptoms.

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Vascular Anomalies

29

Austin N. DeHart, Joana M. Mack, P. Spencer Lewis, Shelley E. Crary, and Gresham T. Richter

Overview

Vascular anomalies are physical manifestations of disrupted vascular development in the form of uncontrolled cellular growth or vessel communication and expansion. They may arise as congenital or de novo lesions to cause aesthetic and/or functional problems. The International Society for the Study of Vascular Anomalies (ISSVA) categorizes vascular anomalies between vascular tumors and vascular malformations (VM) [1]. Vascular malformations are further subdivided by blood vessel type and as high-flow lesions, when an arterial component is present, or low-flow lesions which contain capillary, venous, or lymphatic components. These may be simple, comprised of only one vessel type, or combined, with multiple types of vessels involved. Abbreviated ISSVA classification of vascular anomalies is summarized in Fig. 29.1. There are now a large number and wide variety of vascular anomalies known and managed by multidisciplinary teams across the world. This chapter will cover the most common vascular tumors and malformations with a discussion of surgical, medical, and radiological perspectives.

Vascular anomalies can typically be identified by their presenting clinical history and physical examination [2]. Accurate diagnosis is important as the natural history of each lesion differs and affects treatment strategy. Infantile hemangiomas, the most common vascular tumor, are typically absent or very small at birth. They undergo a rapid growth phase during the first year of life followed by progressive involution. Congenital hemangiomas are present at birth, grow commensurate with the patient, and may either involute rapidly (RICH-rapidly involuting congenital hemangioma) or not at all (NICH-non-involuting congenital hemangioma) [3]. Pyogenic granulomas, also called lobular capillary hemangiomas, are acquired vascular lesions of the skin and tend

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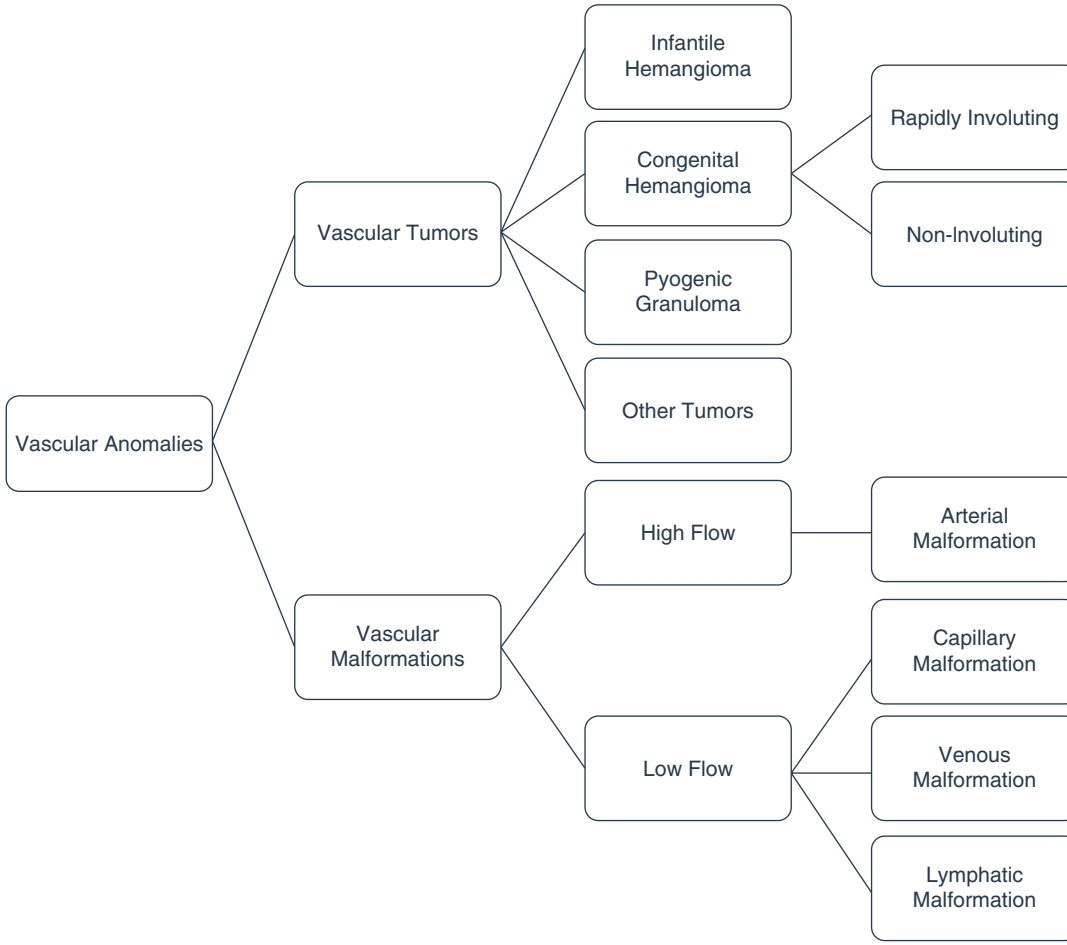

Fig. 29.1 Flowchart demonstrating abbreviated ISSVA classification system for vascular anomalies

to be smaller, pedunculated, and more prone to bleeding [3]. Vascular malformations are present and most often evident at birth but grow slowly over time, often leading to aesthetic and functional issues. Acute growth may occur with trauma, infection, or hormonal influences. There is much variety in the presentation of vascular malformations, from limited "port-wine stain" capillary malformations to complex, infiltrative, multifocal lesions.

Infantile Hemangiomas

Infantile hemangiomas are the most common vascular tumor and occur in ~5% of the population [4]. They express GLUT1, a receptor also found on placental blood vessels [5]. Infantile hemangiomas can be described by their extent and depth of tissue involvement. Focal infantile hemangiomas are smaller, well-defined, and typically solitary. Segmental hemangiomas cover a wide area, are poorly demarcated, and have irregular, geographic shapes. Superficial hemangiomas demonstrate dark red color change in the skin. Deep hemangiomas present as a subcutaneous mass with overlying blue skin discoloration. Compound hemangiomas may have both superficial and deep components.

The natural history of infantile hemangiomas is to rapidly grow over the first year of life and then spontaneously involute, typically resolving by about 7 years of age. Because of this expected resolution, hemangiomas have traditionally been managed with observation alone. The exception

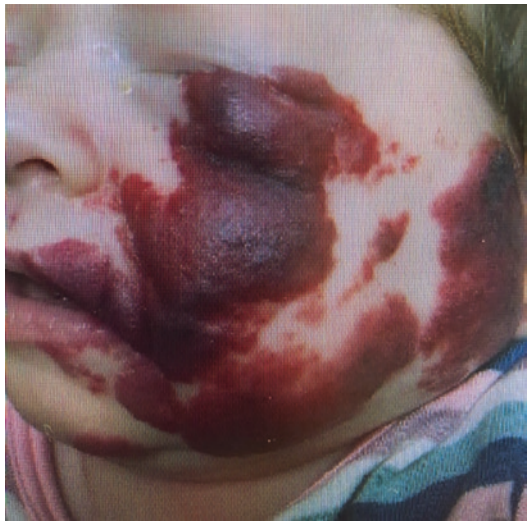

Fig. 29.2 Segmental facial hemangioma in beard distribution

to this is lesions that become symptomatic during the growth phase and develop ulceration, bleeding, vision disturbance, or limit function.

In certain cases, infantile hemangiomas can be associated with syndromes. Infants with multiple focal cutaneous infantile hemangiomas have a risk of hepatic involvement and should undergo hepatic ultrasound [6]. Segmental hemangiomas are associated with PHACES syndrome, the constellation of posterior fossa malformations, hemangiomas, arterial lesions, cardiac abnormalities, eye abnormalities, and sternal cleft. Workup with a head and neck MRI and ophthalmology exam is essential. Segmental facial hemangiomas in a beard distribution (V3) (Fig. 29.2) are associated with concurrent subglottic hemangiomas (50–60% of the time) and should undergo laryngoscopy and bronchoscopy [7].

Arteriovenous Malformations

Arterial venous malformations (AVM) present as masses that are characteristically warm and pulsatile, with a palpable thrill, and often associated with a red cutaneous vascular stain and telangiectasias. They are very rare and may be misdiagnosed as hemangiomas because both are high-flow lesions. AVMs lack the early vertical growth and involution seen in hemangiomas. AVMs can develop local symptoms and deformity due to progressive growth and infiltration as well as ulceration and bleeding, which may become life-threatening. Triggers for acute growth are treatment interventions, hormonal fluctuations, and trauma. They are classified as either focal, with discrete borders and one to two feeding vessels, or diffuse, with no discrete boundaries and multiple arterial feeders [8]. Diffuse malformations are challenging to treat due to their infiltrative nature and have a high recurrence rate. Arterial imaging with CTA or arteriogram can better characterize the extent of the lesion and locate the feeding vessels.

Capillary Malformations

Capillary malformations, often called "port-wine stains," are superficial red-stained lesions made of thin-walled, small-caliber vessels (Fig. 29.3). When left untreated, many will thicken, become darker, and develop tissue hypertrophy, so early treatment is indicated. They can occur spontaneously or in association with syndromes. A somatic mutation in the GNAQ gene has been linked to development [9]. Sturge-Weber syndrome is a sporadic disorder defined by a facial capillary malformation associated with abnormal vessel development in the eyes and brain which can lead to glaucoma, seizures, and strokes [10]. Capillary malformations found on the extremities can be associated with bone or soft tissue hypertrophy. When occurring with varicose veins or venous malformations, it is known as Klippel-Trenaunay syndrome [11].

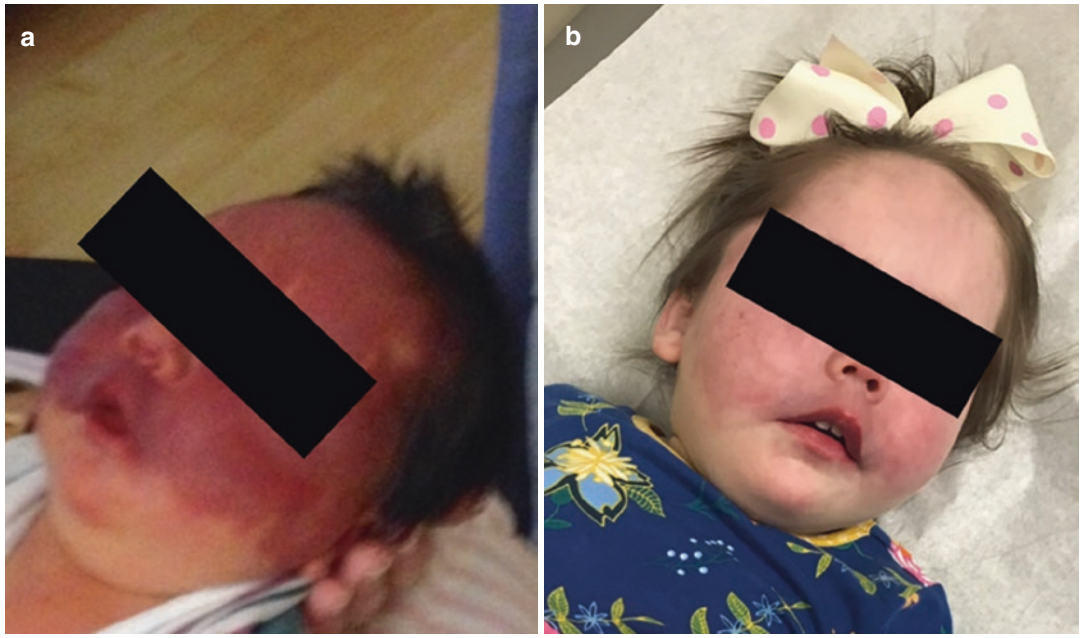

Fig. 29.3 Capillary malformation of facial skin before (**a**) and after (**b**) laser treatment

Venous Malformations

Venous malformations (VM) are made of abnormal venous channels with thin walls with abnormal smooth muscles. When present, they form a sponge-like network of venous channels with poor drainage. While they are benign and uncommon, VM will continue to expand over time with vascular dilatation and infiltration of normal tissue, so treatment is merited that targets the malformation and spares local tissue. They present as blue, compressible, soft masses and can develop clots or calcifications (phleboliths) within the malformation, which can be painful (Fig. 29.4). They often involve the skin and aerodigestive tract. Ultrasound with color Doppler and MRI can help confirm the diagnosis and evaluate lesion extent.

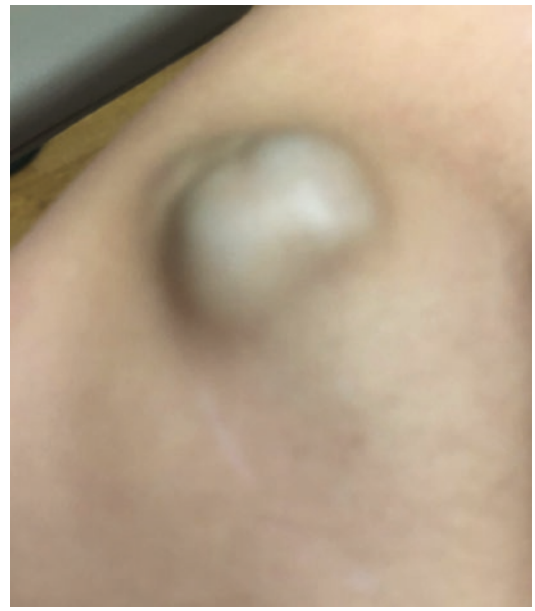

Fig. 29.4 Venous malformation of the arm

Lymphatic Malformations

Lymphatic malformations are comprised of dilated, abnormally formed lymphatic channels and sacs (Fig. 29.5). They are classified as microcystic (<2 cm) or macrocystic (>2 cm) based on size and can also be mixed. While present at birth, they are often undetected until they enlarge, often after an acute infection or trauma

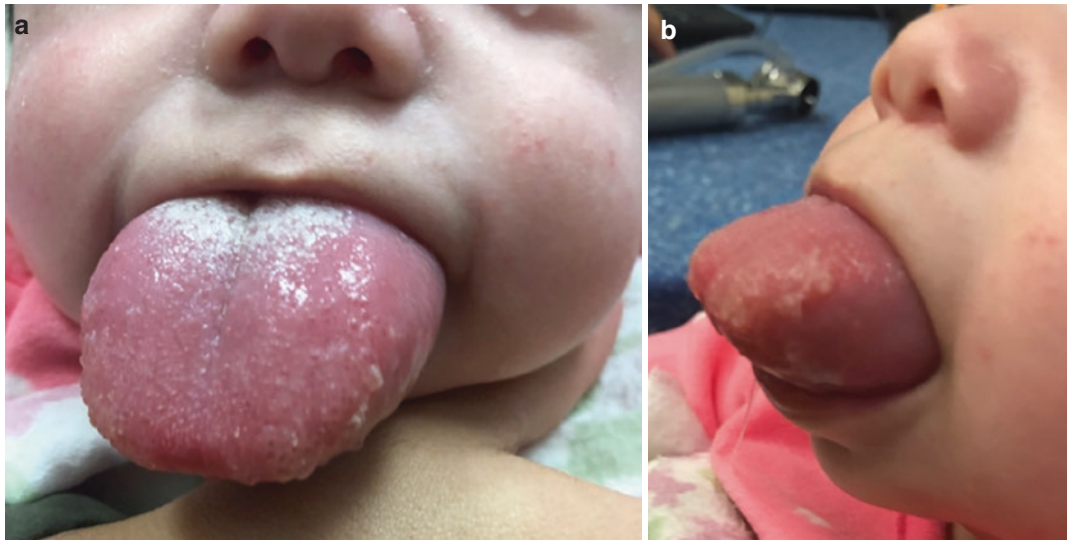

Fig. 29.5 (**a**, **b**) Microcystic lymphatic malformation of the tongue

or with puberty due to hormonal changes. They are soft, compressible, water-filled neck masses, sometimes with superficial vesicles. Symptoms are generally related to pain, swelling, mass effect, and infiltration based on their location. Ultrasound will demonstrate a low-flow lesion, while a contrast-enhanced MRI can help determine the extent and depth of disease.

Otolaryngologist Approach

Management of vascular anomalies in the head and neck is challenging due to the high density of critical structures and complex anatomy. Vascular anomalies can significantly impact appearance and function, causing deformity and disability. A crucial tenet in the management of vascular anomalies is to ensure that the treatment is no worse than the disease.

History

A thorough history is necessary to assess the lesion and correctly diagnose it. Particular importance should be placed on birth history, growth of the lesion, and any associated symptoms. Stridor, feeding or swallowing problems, bleeding, ulceration, and chronologic history should be elicited.

Exam

A complete head and neck exam should be performed to determine the superficial or deep extent of the vascular anomaly. Changes to oral mucosa coloration may indicate their presence. Flexible fiber-optic laryngoscopy in the clinic is often necessary to visualize the upper aerodigestive tract and evaluate laryngopharyngeal disease involvement.

Management of Infantile Hemangioma

Of particular importance in the head and neck are the functions of speech, swallowing, airway patency, and facial cosmesis. Infantile hemangiomas that bleed, cause functional limitation, or affect vision should be treated and are labeled as problematic (Fig. 29.6). Since the discovery of successful treatment of infantile hemangioma in 2009 with oral beta-blockade (propranolol), this has been the medication of choice for large or symptomatic infantile hemangiomas. However,

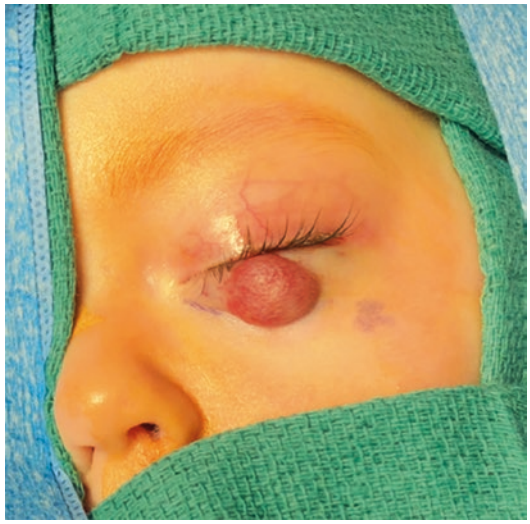

Fig. 29.6 Eyelid hemangioma affecting visual fields

multimodal management with cutaneous flash pump dye laser, intralesional steroid therapy (triamcinolone), and propranolol is often required in larger or more complex disease to achieve complete resolution. Typically, an EKG is obtained prior to initiation of propranolol therapy. In a 2013 consensus conference, guidelines were released that patients older than 8 weeks are safe to start therapy on an outpatient basis, while younger infants should be admitted for observation [12]. Propranolol is generally safe with low rates of hypoglycemia, bronchospasm, or cardiac events. Topical beta-blocker use in the form of timolol 0.5% has been an alternative treatment for superficial, localized, small infantile hemangiomas and has a 91% resolution rate [13]. With topical therapy, treatment during the proliferative phase typically gives the best outcome.

Special considerations apply to the treatment of subglottic hemangiomas. These typically present in an infant with new-onset, progressive stridor during the proliferative phase and are diagnosed with airway endoscopy in the operating room. Systemic propranolol is the mainstay of treatment and has success rates reported at 90% [14]. Of utmost consideration in these patients is airway patency, and endotracheal intubation may be necessary as a temporizing measure until the hemangioma responds to therapy. Intralesional steroid injection may also provide some benefit. If either medical modality fails, open resection with laryngotracheoplasty is appropriate.

Parotid hemangiomas have similar response rates of 90% with propranolol but may require prolonged treatment [15]. If started early in the proliferative phase, propranolol may be an adequate therapy alone. If disease persists superficially in the skin, this can be treated with flash pump dye laser therapy (585–595 nm), but surgical intervention may be necessary to remove residual soft tissue bulk through a parotidectomy approach with facial nerve identification and preservation.

Other hemangiomas with a significant vertical growth pattern may also leave undesirable fibrofatty residuum, which can be treated with elliptical surgical excision (Fig. 29.7). This is particularly common in scalp hemangiomas which develop alopecia during the involution phase, so these are typically excised [16].

Management of Vascular Malformations

Multiple treatment options exist for vascular malformations including surgery, laser photothermolysis, sclerotherapy, and systemic targeted drugs. While small, focal vascular malformations may be successfully treated with a single modality, deep or infiltrative lesions ben-

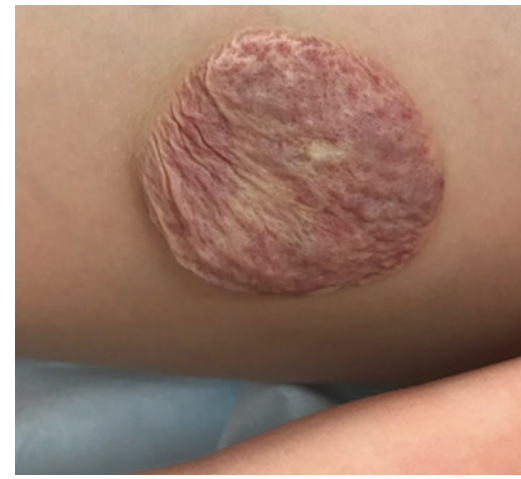

Fig. 29.7 Fibrofatty residuum after hemangioma involution

efit from a multidisciplinary, staged, multimodal treatment plan to achieve optimal outcomes.

Lasers

Laser therapy is a valuable treatment option for the superficial, pigmented component of vascular anomalies due to selective photothermolysis. Laser therapy treats specific vessel sizes and chromophores, typically oxygenated or deoxygenated hemoglobin, to selectively absorb energy which injures tissue in a targeted fashion. This allows for treatment of the anomaly without damage to surrounding normal structures. Superficial, slow-flow lesions respond better to laser therapy due to higher concentration of thermal injury [17]. This makes lasers a particularly appealing modality for superficial hemangiomas, capillary malformations, and mucosal or skin venous and arteriovenous malformations. Laser treatments are not beneficial for purely lymphatic malformations. The risks of laser treatment include local pain, blistering, ulceration, scarring, and pigmentation changes.

Multiple lasers are used in the treatment of vascular anomalies depending on the depth and vessel size of the lesion. Pulsed dye laser (PDL), with wavelength of 585 nm, targets superficial, small-diameter vessels. This is used commonly for the treatment of cutaneous lesions, especially capillary malformations [17]. Multiple treatments are typically needed. When PDL was used in infancy on capillary malformations of the skin, 75% lightening was achieved after four treatments [18]. The Nd:YAG laser has a wavelength of 1064 nm and targets larger vessels, typically in venous malformations. This laser can be delivered through a fiber, which lends itself to the treatment of upper aerodigestive tract lesions during operative laryngoscopy or to interstitial treatment of a lesion. The Gentle YAG laser uses the same wavelength but comes with a coolant spray to treat darkly pigmented skin and penetrates up to 8 mm for deep venous lesions [17]. The carbon dioxide (CO_2) laser has been used for mucosal vesicles of microcystic lymphatic malformations with success but requires repeated treatments. It has also been used for infantile hemangiomas of the subglottis but is associated with high rates of scarring and airway stenosis, so alternative treatments are now preferred [19].

Surgical Resection

Surgical resection remains a valuable tool for the treatment of vascular malformations (Figs. 29.8 and 29.9). For focal, small lesions, excellent cure

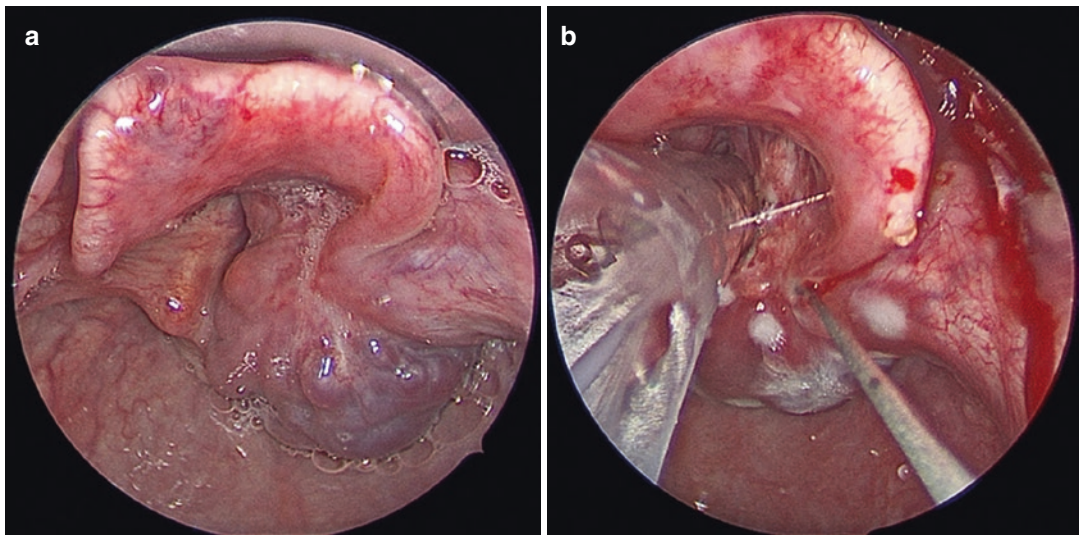

Fig. 29.8 Venous malformation of the airway, before (**a**) and intraoperative image during (**b**) laser and steroid injection treatment

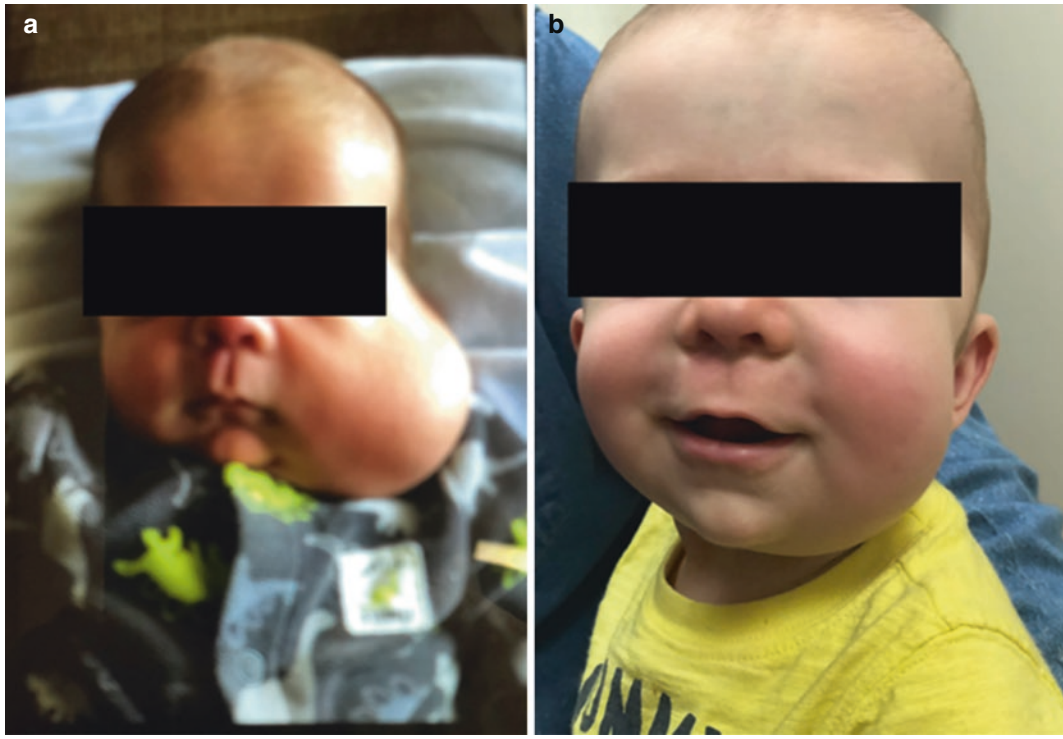

Fig. 29.9 Cervicofacial macrocystic lymphatic malformation, before (**a**) and after (**b**) surgical resection

rates can be achieved with wide local excision. Larger, diffuse lesions present difficulty due to tissue infiltration and high recurrence rates. However, macrocystic malformations are amenable to surgical resection with a high rate of cure as they expand around soft tissue rather than within.

For more vascular lesions, preoperative sclerotherapy/embolization can induce thrombosis in the malformation and helps reduce blood loss. In particular, NCBA glue can be used in either venous malformations or arteriovenous malformations to define the lesion and provide borders for resection. Resection in previously treated areas is prone to distortion of anatomy and tissue planes, which makes the identification of critical structures more difficult. Ideally, surgical resection is performed in conjunction with preoperative embolization to targeted areas in a staged fashion, treating one area at a time at 3-month intervals. Complete resection of large lesions often leads to unacceptable functional and aesthetic results and so is often not feasible. Maintaining a global perspective and selectively targeting symptoms are key to a good outcome.

Hematologist-Oncologist Approach

History

Vascular anomalies are comprised of malformed arteries, veins, capillaries, lymphatics, or a combination of two or more of those vessels. They are further divided into vascular tumors and vascular malformations (VM) [20]. Large VM can be debilitating and lead to significant complications. As with any complex disease, a detailed history, specifically focused on the evolution of the vascular malformation, is crucial. Eliciting the following information is important: time of onset, growth over time, initial color and change of color, and exacerbating symptoms (i.e., illnesses, onset of puberty, pregnancy, menopause). Patients may

even express a feeling of warmth, throbbing, and compressibility or an increase in size with change in extremity position.

Because a simple or complex VM can be associated with other syndromes (e.g., PHACE {posterior fossa, hemangioma, arterial, cardiac, eye abnormalities} syndrome or LUMBAR {lower body hemangioma, urogenital anomalies, myelopathy, bone deformities, anorectal/arterial malformations, renal anomalies} syndrome), it is prudent to do a full review of systems. A patient's past medical history may also elucidate associated syndromes (history of renal anomalies, eye anomalies, etc.). Family history is needed due to the hereditary nature of certain vascular malformations.

VM can further be delineated into low-flow and high-flow entities. High-flow malformations include arterial malformations, arteriovenous fistulas, and arteriovenous malformations. Low-flow VM include capillary, lymphatic, and venous malformations. There are several complications seen in patients with low-flow VM which include a disturbance in the hemostatic system. Therefore, it is important to elicit a history of episodic or persistent pain associated with "knots" within their malformation indicating thrombosis. Previous surgical history with an emphasis on complications with bleeding requiring blood products or thromboembolism is important as well.

Exam

Examination from head to toe is needed since vascular anomalies can be focal or diffuse and superficial or deep. The color of the malformation is dependent on the type of vessel that is involved. Superficial lesions that appear a reddish-maroon color may be capillary malformations. Bluish lesions may be deeper and signify a venous malformation.

Valsalva maneuvers may enlarge venous malformations due to the weakened vessel walls. Change in position of the involved lesion (lowering the extremity or placing a child in the supine position) below the level of the heart can enlarge the vessel due to filling and decreased venous return. Limb length and size discrepancies should be measured. A genitourinary exam can reveal pelvic and rectal involvement. Some lesions may have no color and present as a protruding mass. Warmth, pulsations, and/or an audible bruit will favor a high-flow lesion and can help differentiate from a low-flow VM. Evaluating the skin for ulceration, leakage of fluid, swelling, or hard well-circumscribed masses (indicating thrombi) is also important.

Diagnostic Evaluation

Localized intravascular coagulopathy (LIC) occurs mainly in low-flow VM but can be devastating if it progresses to disseminated intravascular coagulopathy (DIC). Obtaining a complete blood count (specifically looking at hemoglobin/hematocrit and platelet count), D-dimer, and fibrinogen at baseline and prior to any surgical/invasive procedures is critical. If there are abnormalities in the values, further management is needed.

Other coagulopathies such as Kasabach-Merritt phenomenon (KMP) can be seen in kaposiform hemangioendothelioma (KHE) and tufted angioma (TA), which are locally aggressive vascular tumors. KMP is a consumptive coagulopathy classically involving severe thrombocytopenia and hypofibrinogenemia due to platelet activation within the rapidly growing tumor. This can be life-threatening; thus, early recognition by evaluating a complete blood count, prothrombin time (PT), activated partial thromboplastin time (aPTT), and fibrinogen is essential [21].

Medical Management

Due to the complexity and uniqueness of each individual VM, proper diagnosis is essential. Current treatment strategies require a multidisciplinary approach. There is a multitude of treatments available, but those that will be discussed in this section require the expertise of a hematologist-oncologist.

Sclerotherapy, a process in which intralesional injection causes vessel injury, has been shown to be effective [22]. Bleomycin, an antitumor antibiotic, leads to an inflammatory and fibrotic effect on endothelial cells when injected into an affected vessel [23]. Although there is no gold standard sclerosant, our institutional experience with bleomycin has shown a less painful inflammatory response.

With the increasing use of bleomycin in VM, there needs to be close monitoring for cumulative doses and long-term side effects. Bleomycin has been used in malignancies such as Hodgkin lymphoma (HL) and germ cell tumors and is associated with several side effects, the most concerning of which is pulmonary dysfunction [24, 25]. Our institution has a specific protocol in place and includes obtaining chemotherapy informed consent for every patient receiving bleomycin. Consent is obtained by an oncology physician or a treating member of the vascular anomaly team. The treatment and side effects are discussed thoroughly. The oncology faculty is required to provide a second signature prior to a patient receiving bleomycin. Four to 15 units of bleomycin is used in patients in one setting, depending on the size of the lesion being treated and age of patient. Prior to the first dose, a baseline physical exam is warranted with focus on the pulmonary history and exam. If the patient is >7 years of age, pulmonary function tests (PFTs) are obtained with specific focus on the diffusing capacity of the lung for carbon monoxide (DLCO). We no longer monitor chest x-rays since evidence of pulmonary fibrosis would be a late finding. Oxygen saturation is obtained immediately prior to each procedure to be certain there is no hypoxia. It is important to avoid high concentrations of oxygen during any general anesthetic. A complete blood count is obtained 2 weeks following each bleomycin treatment to monitor for cytopenia as seen in patients with malignancies. PFTs are repeated after the patient has received 60 units/m^2 to evaluate for pulmonary dysfunction. The maximum cumulative dose of bleomycin sclerotherapy is 100 units/m^2 at our institution.

It is difficult to make a direct comparison for systemic absorption between intralesional and intravenous bleomycin, but the intralesional bleomycin systemic distribution is likely lower than intravenous doses. Adults who received intravenous bleomycin showed evidence of pulmonary fibrosis in about 10% of adult patients after being exposed to a cumulative dose of 400 international units (~200 units/m^2 in an average size adult) [24, 25]. Four hundred units is the lifetime dose maximum recommended for oncologic processes. Thirty-four asymptomatic childhood HL survivors received a maximum of 60 units/m^2 of intravenous bleomycin with 40% showing evidence of restrictive or obstructive lung disease at a median follow-up of about 2 years [26].

Another drug that requires the expertise of a hematologist-oncologist is sirolimus (rapamycin). Sirolimus, a mammalian target of rapamycin (mTOR) inhibitor, has been shown to be effective and safe in patients with complex vascular anomalies [27]. Several VM genetic mutations have since been identified along the PI3K pathway. The mTOR serine/threonine protein kinase is involved in the PI3K pathway and therefore is a fruitful target in preventing cellular growth and survival [28, 29].

Sirolimus is an immunosuppressive medication that has historically been used in solid organ transplantation to prevent graft rejection. Therefore, a lot of the information of side effects has been reported in the transplantation literature. No live vaccinations should be given during treatment. Because of its immunosuppressive properties, patients are susceptible to opportunistic infections, specifically *Pneumocystis jirovecii* pneumonia (formerly *Pneumocystis carinii*). Prophylaxis with trimethoprim/sulfamethoxazole is the recommended first drug of choice. Patients need to be evaluated for significant illnesses and/or fever of 101 °F or more. Blood work should be considered including a complete blood count and blood culture.

Dosing of sirolimus can be started at once to twice a day (1.6 mg/m^2/day), and trough levels must be obtained after 2 weeks from the start and then every 1–3 months while on the medication. Sirolimus dosing is titrated for response

and trough level between 10 and 15 ng/mL [30]. Sirolimus should be held for serious infections such as pneumonia, bacteremia, mononucleosis, etc. Likely, but usually reversible, side effects include high blood pressure, loss of appetite, swelling, mouth sores, increase in cholesterol and/or triglycerides, anemia, and skin rashes. Less likely but severe side effects include severe infections, low white blood cell count, poor wound healing especially after surgery, hypotension, fluid accumulation around organs (heart, lungs, kidneys), kidney failure, cancers (lymphoma, skin), and infertility [29].

Use of topical sirolimus has little to no systemic absorption; therefore no level monitoring is needed. It has been used in port-wine stains (PWS) and has shown some efficacy with the use in conjunction with laser therapies [31].

As discussed earlier, slow-flow VM have a unique complication of LIC due to the slow blood flow through the abnormal and sometimes ectatic vessels. LIC occurs due to stagnant blood flow which can lead to alteration in the coagulation system causing bleeding and/or localized thrombosis [32]. This can further lead to disseminated intravascular coagulopathy (DIC) which can be life-threatening and result in severe bleeding. A significantly elevated D-dimer with or without hypofibrinogenemia, in addition to a history of pain and "knots," and evidence of phleboliths on imaging can confirm LIC and may warrant anticoagulation, especially periprocedurally. These low-flow lesions can be treated with compression garments and antithrombotic therapy with aspirin or low-molecular-weight heparin (LMWH) to treat pain [33]. Direct oral anticoagulants can also be utilized for treatment or prevention of thrombosis in adults but are not currently FDA approved in children [34].

Emerging Treatment

The phosphoinositide 3-kinase (PI3K) pathway is a signaling network that involves the cell cycle and is frequently altered in human tumors [35]. Through advancements in understanding the molecular pathways that are involved in vascular anomalies, several disorders have been associated with mutations in the PI3K pathway. The pathway involves several proteins including PI3K/AKT/mTOR for which development of targeted inhibitors is promising [35, 36]. Several genetic mutations in complex, disfiguring, and debilitating VM have been discovered. The following mutations have been associated with the following syndromes: AKT1 in Proteus syndrome, PIK3CA in Klippel-Trenaunay syndrome (KTS), CLOVES (congenital, lipomatous, overgrowth, VM, epidermal nevi, spinal/skeletal) syndrome, and FAVA (fibro-adipose vascular anomaly) [37]. Recently, Canaud et al. evaluated 19 patients with PROS after using BYL719, an inhibitor of PIK3CA inhibitor, which showed improvement in disease symptoms in all patients [38]. We have entered the era of molecular classification, and novel targeted therapies are quickly being developed which will advance the treatment of VM.

Interventional Radiologist Approach

In addition to surgical and medical treatments for vascular anomalies, there exists a wide range of percutaneous treatments which are performed under radiologic guidance. Simpler lesions may be amenable to treatment with sclerotherapy under the sole care of an interventional radiologist; however, more often the interventional radiology treatment is complimentary with treatments by other specialties. There is no universally agreed best treatment for vascular anomalies with each center and practitioner using slightly different sclerosants, preparations, and methods, but many of the general principles are used by many interventional radiologists.

Venous Malformations

Venous malformations range from isolated large varicosities to more diffuse and infiltrative disease. The location and characteristics of the venous malformation will tend to favor treatment with either surgical resection or sclerotherapy.

Venous malformations with well-defined margins which are reasonably isolated from critical structures are effectively treated with surgical resection, often requiring only one hospitalization. Glue embolization of the venous malformation can be extremely beneficial to the surgeon, both by making the malformation firmer and more easily resectable and also by limiting intraoperative blood loss [39].

Many venous malformations are not candidates for surgical resection, often due to infiltration of important structures such as muscle and bone which would result in morbidity if resected. These lesions are frequently treated with sclerotherapy. Ethanol, sodium tetradecyl sulfate, doxycycline, and bleomycin are some of the more commonly used sclerosants. There is no consensus as to which sclerosant is best; however, ethanol is often considered the most potent sclerosant and ideal for lesions where adjacent injury is better tolerated (e.g., intramuscular) but should be used with caution near the skin and critical structures such as nerves. Bleomycin has shown promise to treat venous malformations with lower rates of inflammation and injury to adjacent structures and may be used for sensitive areas such as orbits, airways, and near nerves.

Additional technical considerations for treatment of venous malformations are the characteristics of the drainage of the malformation. Some venous malformations have no significant central drainage (sequestered) and can be treated by removing the venous blood and simply replacing it with sclerosant. For lesions with central drainage, the centrally draining veins must be blocked in order to allow the sclerosant to be effective. Techniques range from coil or glue embolization of the draining veins to simple pressure from a tourniquet.

Lymphatic Malformations

Lymphatic malformations broadly fall into two groups, macrocystic and microcystic. If the lymphatic cyst is large enough to puncture directly, near complete remission can often be accomplished with a few sessions of sclerotherapy. Percutaneous needle puncture with drainage of intraluminal contents is performed using a 1:1 replacement with sclerosant, frequently doxycycline, which is allowed to dwell for 30–60 min. Although doxycycline has been reported to cause tooth discoloration in pediatric patients when used as a systemic antibiotic, this side effect has not been reported for sclerotherapy of macrocystic lymphatic malformations, likely in part due to removal of the sclerosant at the end of the case.

Microcystic lymphatic malformations often come as a cluster of tens if not hundreds of tiny cysts, and treating each tiny cyst with intraluminal sclerosant is not possible. These lesions often benefit from adjacent, interstitial administration of sclerosant. Many sclerosants have been used, although bleomycin is one of the most common and effective for this indication.

Arteriovenous Malformations

Most interventionalists consider arteriovenous malformations the most difficult to treat. The high-flow nature of these lesions makes them difficult to treat intraluminally due to the rapid washout of sclerosant. In addition, partial treatment often results in increased angiogenesis which can worsen the disease over time. If possible, complete destruction of the malformation should be attempted. Unfortunately, the nature and location of many malformations make them incurable, and in these cases selective, incomplete treatment may be the only option.

There has been a great deal of literature detailing innumerable methods of treating arteriovenous malformations, and an entire book could be written detailing the different methods. For brevity, some of the more commonly used methods will be described here. However, to prevent vascular collateralization and recruitment, management of AVM should be staged and performed at periodic intervals (3–4 months). Although cure may not be possible in diffuse AVM, control can be maintained with gradually increasing treatment intervals.

If the AVM is amenable to surgical resection, preoperative glue embolization can aid the

surgeon and minimize blood loss. The goal of glue embolization is both to inject as much glue into the nidus of the malformation as possible and also to embolize the feeding vessels and venous outflow tracts. Single large feeding vessels may be occluded intraoperatively; however, many malformations have numerous small feeders arising from different arteries. Focal AVM (one to two feeding arteries), though, can often be cured with a single intervention with embolization followed by surgical resection.

For nonsurgical lesions, intraluminal ethanol injection is often considered the best treatment. Small aliquots of ethanol can cause rapid protein denaturation and endothelial cell injury resulting in a marked inflammatory response. The end result is fibrosis of the malformation. The injection site is selected to maximize the amount of alcohol that will flow into the nidus of the malformation, and 1 mL/kg of ethanol up to 50 mL is often injected during each treatment. Depending on the exact anatomy of the lesion, it may be best approached with arterial access, venous access, or direct puncture. These are long, difficult cases and almost always require numerous treatments to achieve the desired result.

Intraprocedural and Postprocedural Care

With the exception of bleomycin, all forms of sclerosis cause moderate to intense inflammatory responses and result in pain and swelling to the site. Steroids are often given both during the procedure and for 5–7 days afterward to reduce discomfort. Hemolysis is common and can result in hemoglobinuria. For this reason, patients are hydrated during the procedure, and if there is evidence of hemoglobinuria (often cola-colored urine), fluids are switched to D5W with 75 mEq sodium bicarbonate per liter to cause alkalization of the urine. Pain control medications can range from NSAIDs to narcotics. Skin discoloration seen during the procedure often precedes blistering and rarely ulceration. Patients should be given antibiotic ointments and followed closely.

Summary

Vascular anomalies represent a complex spectrum of diseases that benefit from a multidisciplinary management team. Routine questioning for symptoms of voice, swallow, and breathing symptoms to suggest airway involvement is recommended. Consideration of flexible laryngoscopy and, at times, direct laryngoscopy with bronchoscopy in the operating room may be warranted depending on the clinical context. A variety of treatment options including laser, surgical resection, sclerotherapy, and systemic medications should be part of the vascular anomaly team armamentarium. Optimal outcomes can be achieved using treatment that targets abnormal tissue and preserves the form and function of normal structures through a multimodal, staged approach.

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Vocal Fold Scar

30

Maria E. Powell and Bernard Rousseau

Introduction

Chronic vocal fold scar is the consequence of a well-orchestrated wound healing process gone awry. There are multiple causes of scarring in the pediatric population. Patients with a history of prolonged intubation are at risk for trauma to the vocal folds, which can precipitate scarring. Vocal fold scarring is the primary cause of dysphonia following laryngeal surgery [1]. Specifically, chronic scar may present as a secondary sequela following surgical intervention for juvenile-onset recurrent respiratory papillomatosis (JORRP), vocal fold webbing, tracheal stenosis, or other upper airway reconstruction procedures. Over the past two decades, a substantial body of research in wound healing and regenerative medicine has led to significant advances in the characterization of the molecular and structural changes in chronic scar. As ethical considerations preclude widespread investigation of vocal fold wound healing and scarring trajectories in the vulnerable pediatric population, much of our understanding of wound healing arises from animal models and adult human studies. The normal wound healing cascade in the pediatric population is similar to the adult trajectory described below; however, wound healing among neonates and children is much more efficient than adults. As a result, normal wound healing with limited medical intervention is the currently accepted clinical expectation in the pediatric population [2], though specific applicability to vocal fold wound healing is not understood. Despite the decreased incidence of vocal fold scarring in children, when chronic scarring does occur, it represents a clinically significant challenge.

Wound Healing

Scarring is a natural aspect of the body's repair process in response to trauma. Insults to soft tissue disrupt the native microarchitecture and subsequently alter tissue function. The initial insult disrupts the surrounding vasculature, causing blood to spill into the wound bed. As platelets come into contact with connective tissue within the extracellular matrix (ECM), they initiate a "cascade of healing" which progresses through four distinct yet overlapping phases: (1) homeostasis, (2) inflammation, (3) proliferation, and (4) maturation. During the *homeostasis* phase, platelets release growth factors and cytokines (principally transforming growth factor beta, TGF-β)

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to form a fibrin-rich mesh to effectively stop the bleeding and create a scaffold for cell migration.

Once homeostasis is established, *inflammatory* cells infiltrate the tissue to sterilize the wound bed. Within 24 h of injury, neutrophils begin the cleaning process by attacking bacteria and debriding damaged tissue through phagocytosis. Within 48 h of injury, macrophages infiltrate the tissue to continue phagocytosis and further secrete factors to attract immune cells responsible for tissue repair [3, 4]. Fibronectin is a glycoprotein of soft tissue that also plays a key role in chemotaxis and cell signaling and helps to initiate the *proliferation* stage of wound healing.

Fibroblasts are the key players in the deposition of new ECM; however, fibronectin mediates fibroblast migration into the wound bed and adhesion to ECM components [5]. Fibroblasts produce granulation tissue made up of collagen III, elastin, and fibronectin to repair the matrix, and glycosaminoglycans and proteoglycans to occupy space within the matrix, increasing vocal fold volume. Fibroblasts also support endothelial cells in the revascularization of damaged vessels via angiogenesis [6]. Myofibroblasts are contractile fibroblasts which play the vital role of pulling the wound bed margins toward the center of the wound by translocating collagen fibers into a more organized matrix [7, 8]. This contraction of the wound bed reduces the size of the scar and facilitates tissue continuity. As the wound contracts, reepithelialization occurs restoring the tissue's protective barrier within 5 days post-injury [9]. Fibroblast response to injury is greater in children, with more rapid deposition of collagen and elastin. As a result, granulation tissue forms more quickly, and wound closure occurs at much faster rates in the pediatric population in comparison to adults [10–12].

Whereas the first three stages of wound healing occur in quick succession over the course of hours to days, the final *maturation* stage typically begins around 21 days post-injury and may continue for weeks to years. During this period, the tissue is remodeled to approximate the pre-injury microarchitecture. The inflammatory and immune cells active in the initial phases of wound healing are subjected to apoptosis, or programmed cell death, as they are no longer needed. Although overall collagen production has been shown to stabilize around 21 days post-injury, a dynamic process of degradation, synthesis, and remodeling may continue over the next year [13]. Collagen III that was initially produced is remodeled to collagen I, which is much thinner, more organized, and more abundant in healthy tissue than collagen III. In time, these mature collagen I fibers cross-link, creating a basket-weave formation along tension lines to reduce mass and increase tensile strength. Although the scar becomes thinner and more flexible, the remodeled tissue never fully recovers to premorbid function. Dermal wound healing studies suggest that scarred tissue only ever achieves approximately 80% of its pre-injury tensile strength [4].

Pathologic Vocal Fold Scar

Pathological scarring occurs when some aspect of the healing cascade is disrupted or continues unchecked [14]. Fibrosis associated with excessive scarring develops as a result of deviations during the proliferation and maturation stages of wound healing. An overabundance of fibroblast infiltration during the proliferation stage results in the excessive deposition of ECM connective tissue, particularly collagen III, which may contribute to fibrosis. However, in chronic scar, the overall abundance of collagen appears less influential on tissue function than the inadequate remodeling of collagen III into collagen I during the maturation stage [15]. Chronic vocal fold scarring has been characterized in multiple animal models by an increase in the density of disorganized collagen type I and III fibers throughout the lamina propria [15–19]. During the normal maturation phase of wound healing, collagen III is remodeled into a well-organized matrix of collagen I, which is critical as collagen III is structurally much thicker and less organized than its collagen I phenotype.

Similarly, the persistent fragmentation and disorganization of elastin fibers, particularly in the subepithelial region, have been shown to influence tissue function more substantially than what would be expected due to simply an overabundance of the protein [20]. Characterization of elastin following tissue insult shows fragmented and disorganized fibers with increasing density in the deeper layers of the lamina propria [19, 21].

Hyaluronic acid (HA) is a non-sulfated glycosaminoglycan which functions as a gel-like buffer

to maintain separation between collagen, elastin, fibronectin, and other fibrous protein structures within the ECM [22–26]. It is hypothesized to play an important role in the vocal fold's viscoelastic tissue properties, as it functions to absorb shock from vibratory impact [27]. Studies investigating the presentation of hyaluronic acid post-injury report contradictory findings [15–17, 19, 25]. During the wound healing process, the amount of HA increases within the first week following injury [25]. However, in the chronic scar, HA levels vary, with some studies showing no differences in HA levels between normal vocal folds and chronic scar [15–17, 19], while other studies show a significant reduction in HA in the ECM [16, 19]. Such variability may be due to differences in experimental hyaluronan preparation, investigative time points post-injury, and animal model utilized [26, 28].

Fibronectin is increased in the superficial lamina propria in both the immature scar (2 months post-injury) and the mature scar (6 months post-injury), highlighting the long-term role of fibronectin in scar formation and maturation [29, 30]. Immunohistochemistry of canine vocal folds at 6 months post-injury revealed that fibronectin is co-deposited with collagen (Fig. 30.1) [30].

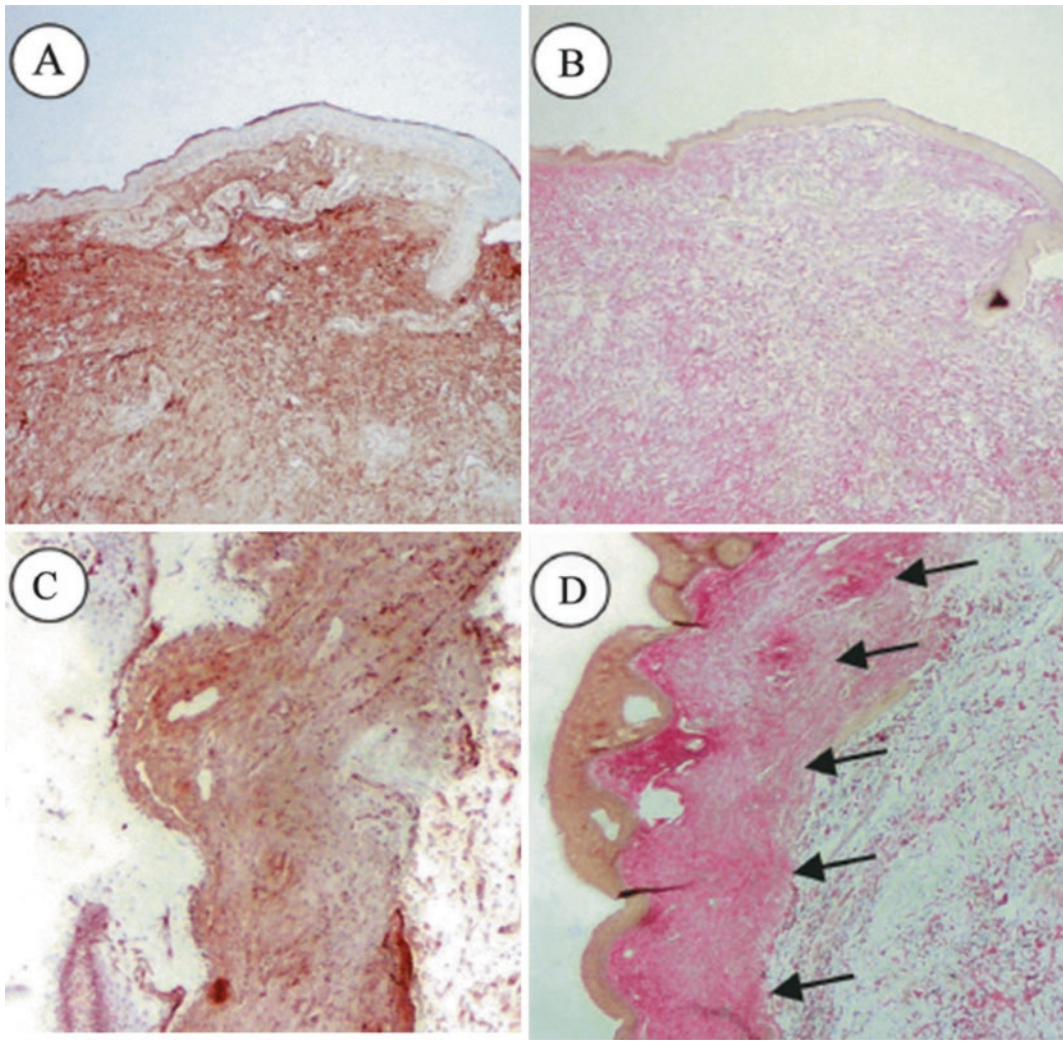

Fig. 30.1 (**a**) Expression of fibronectin (original magnification x4) and (**b**) Elastica van Gieson stain (original magnification ×4) in immature scar (2 months post-injury). Collagen (stained red) is sparse and is not co-deposited with fibronectin. (**c**) Expression of fibronectin (original magnification ×10) and (**d**) Elastica van Gieson stain (original magnification ×10) in mature scar (6 months post-injury). Collagen deposition is observed in the matrix of fibronectin as indicated by arrows. (From Hirano et al. [30], with permission)

Earlier research indicated that while fibronectin and collagen interacted as part of the wound healing process, they did not bind to each other [29]; however, more recent findings confirm that fibronectin is capable of binding to intact, unwound collagen fibers at physiological temperatures [31]. It is therefore possible that these interstitial proteins are not only co-deposited but also bound, contributing to the increased stiffness of the ECM in the pathologic scar.

Sulcus Vocalis

Sulcus vocalis is often grouped with vocal fold scar due to similarities in patient symptoms, functional deficits, and treatment options. Whereas vocal fold scar is characterized by increased deposition of abnormal, unorganized proteins within the ECM, sulcus vocalis is characterized by a loss of ECM. Sulcus vocalis is characterized by a distinct groove, oriented anterior-posterior along the vocal fold free edge. These benign lesions can present unilaterally or bilaterally. Bouchayer and Cornut describe the presentation under laryngoscopy as a "whitish furrow running parallel to the free edge of the fold and producing an aspect of ovular [or spindle-shaped] glottis" [32]. Sulci can result from a congenital malformation and are often believed to occur secondary to a ruptured epidermoid cyst. Acquired sulci may occur following trauma to the vocal folds which significantly damages the lamina propria and creates tethering of the epithelium to the vocal ligament or thyrovocalis muscle [33]. Alternatively, acquired sulci have also been linked to degradation of the maculae flavae. The maculae flavae is responsible for producing fibroblasts, which are critical for ongoing maintenance and repair of the ECM [34]. Ford et al. described three types of sulci, and clinical presentation can range from normal to severely perturbed voice qualities [35].

Type I: Physiological Sulcus

The sulcus affects only the epithelium and the SLP. Gross anatomical inspection may reveal a slight divot in the mucosa as well as a spindle-shaped glottal configuration during adduction. This minor deviation in the mucosa is believed to be congenital and is not always considered pathological.

Type II: Sulcus Vergeture

This pathological sulcus is characterized by an anterior-posterior groove between the upper and lower margins of the vocal fold. In this case, the SLP is significantly involved or even absent, with the epithelium tethered directly to the vocal ligament. Increased collagen deposits surround this focal lesion, creating a stiff, non-vibrating band along the medial edge of the vocal fold. This band affects vocal fold closure (resulting in a spindle-shaped glottal configuration) and mucosal wave propagation.

Type III: Sulcus Vocalis

The third sulcus type is the most severe, involving the full depth of the vocal fold. The epithelium folds into the LP with the deepest portion of the invagination tethered directly to the vocalis muscle. Collagen fibers are present in the LP surrounding the sulcus. The lumen of the pocket formed is lined with stratified epithelial cells that become more keratinized as they approach the vocalis muscle. Some hypothesize that these deep sulci are actually ruptured or open epidermoid cysts [32]. These sulci are often challenging to appreciate upon indirect endoscopic evaluation, but palpation of the region during direct laryngoscopy will reveal the groove. The complete absence of the SLP and vocal ligament as well as the increase in collagen deposits surrounding the

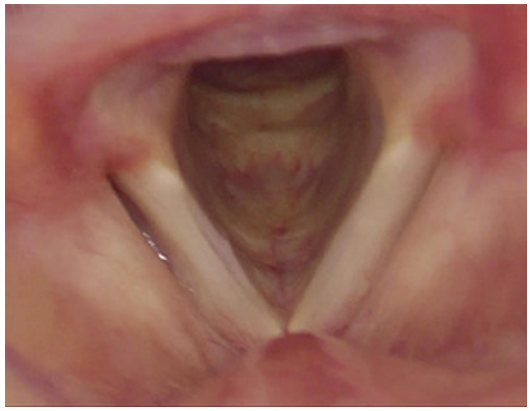

Fig. 30.2 Right vocal fold sulcus vocalis observed during videostroboscopic examination. (Courtesy of Vanderbilt University Medical Center electronic medical records)

sulcus can be appreciated functionally using stroboscopy, as the non-vibrating segment around the sulcus has a significantly reduced or absent mucosal wave (Fig. 30.2).

Clinical Evaluation

Case History

Children and adolescents who present with chronic vocal fold scarring frequently have extensive and complex medical histories. Individuals at particular risk for developing vocal fold scar include those with a history of JORRP [36, 37], prolonged intubation and/or upper airway reconstruction [38, 39], laryngeal web [40, 41], mass lesions involving the vocal ligament [35], and—while rare—laryngeal cancer [42–45]. When obtaining a case history, clinicians should assess the onset, duration, and current presentation of vocal complaints; daily voice use and vocal behaviors; and quality of life and academic impact of the vocal complaint. In addition to these standard questions, it is also important to obtain a thorough medical history. Many of these patients may present with active concomitant disease processes at the time of evaluation that may take precedence over treatment of poor vocal function. Therefore, as part of the case history, it is essential to evaluate not only the patient's current medical status but also the patient's and caregivers' readiness and ability to pursue voice treatment.

Perceptual, Acoustic, and Aerodynamic Assessments

The Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) [46, 47] and dysphonia grade, roughness, breathiness, asthenia, and strain (GRBAS) [48] scales are perceptual assessments conducted by a certified speech language pathologist that should be included in a comprehensive evaluation. Voice quality for patients with vocal fold sulcus or scarring (informed by findings from adult studies) is typically characterized by harsh voice, diplophonia, and abnormal pitch or volume; however, it has also been reported that patients with vocal fold sulcus may instead present with breathy voice due to incomplete glottal closure [35]. Patients with either disorder may complain of increased effort or vocal fatigue. Acoustic and aerodynamic data for patients with vocal fold scarring in the pediatric population are limited; however, based upon our understanding of the structural changes associated with vocal fold scar or sulcus, elevated perturbation measures of jitter and shimmer, and decreased harmonic to noise ratio would be expected. Fundamental frequency may be abnormal for age and gender due to changes in tissue mass, and airflow measures would be expected to be abnormal due to increased subglottal pressure and potentially incomplete glottal closure.

Laryngeal Imaging

Otolaryngologists visualize the gross anatomy and function of the larynx during a laryngoscopy using a flexible endoscope. The goal is to

evaluate structural anomalies including irregular vocal fold edge (Fig. 30.3) and identify concomitant factors such as laryngopharyngeal reflux or chronic inflammation related to allergies or other disease processes which may contribute to, or exacerbate, vocal fold scarring. Laryngologists or speech pathologists will also conduct a videostroboscopic or high-speed videoendoscopic assessment of vocal fold vibration using either a rigid endoscope or distal-chip flexible endoscope. Although videostroboscopy is the current gold standard for functional assessment, challenging cases may benefit from high-speed videoendoscopy for improved temporal resolution of vocal fold vibration [49]. During the laryngeal imaging assessment, clinicians evaluate vibratory function for amplitude, mucosal wave, and glottal closure patterns. Deviations in these characteristics may range from mild to severe. The hallmark feature of scarring is a non-vibratory or adynamic segment visualized as reduced, asymmetric, or absent amplitude and mucosal wave. Visualization of the scarred region may be intermittent and brief (particularly if located along the inferior medial aspect of the vocal fold) and therefore challenging to appreciate [50]. Additional attention should be given to characterize any negative compensatory strategies that have been developed to manage disordered vibratory function.

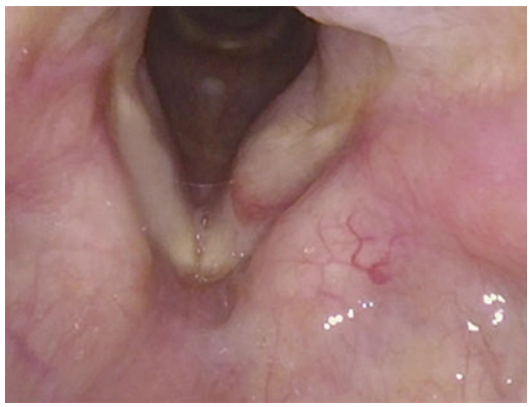

Fig. 30.3 Left vocal fold scarring observed during videostroboscopic examination. (Courtesy of Vanderbilt University Medical Center electronic medical records)

Clinical Management

Minimizing Risk

Since vocal fold scarring is a common sequela following intervention for other, more serious disorders of the larynx and upper airway, the surgical team must often balance the competing goals of maintaining or restoring the airway (intubation and/or airway reconstruction) and optimizing voice outcomes [38]. With this challenge in mind, one of the most effective approaches for reducing the effect of chronic vocal fold scarring is to take a conservative approach when treating other structural disorders which may ultimately result in scarring. Many of the recommendations for conservative treatment are borrowed from the adult population; however, they serve as valuable guidance for treating the pediatric population as well.

JORRP is the second most common cause of hoarseness among children and adolescents [37]. The nature of the JORRP disease process necessitates multiple surgical interventions to remove lesions from the vocal folds to maintain an adequate airway, and the management of JORRP depends on the degree of airway involvement. Recommended treatment options include surgical removal using laser, cold instrumentation, or microdebrider as well as potential use of adjuvant pharmacological therapies. To minimize the risk of vocal fold scarring, only diseased tissue should be removed, and the underlying lamina propria should be preserved as much as possible [51].

Prolonged intubation in medically fragile neonates and children also represents a significant risk factor for developing chronic vocal fold scar and/or laryngeal stenosis. Scarring may be caused by acute trauma to the vocal folds and posterior larynx during intubation. Long-term placement of an endotracheal tube may also cause irritation to the posterior larynx, as pressure from the wall of the tube against the posterior larynx can obstruct blood flow, delaying wound healing [39, 52]. Selection of the appropriate endotracheal tube materials and tube size is critical to reducing the risk of developing

chronic scarring [53]. It is recommended that the tube with the minimum outer diameter necessary to maintain adequate ventilation should be used [39, 52, 53]. Benjamin further describes the composition for an ideal endotracheal tube for prolonged intubation. Specifically, it should be made of nontoxic, synthetic materials which are smooth enough to prevent irritation and have thermoplastic properties which allow it to conform to the tissue contours at body temperature. Additionally, both the tube walls and cuff should allow for a wide pressure distribution to minimize focal contact pressures [39]. As scar formation and possible posterior laryngeal stenosis may develop days to weeks following extubation, one could consider performing serial flexible laryngoscopies if injury was seen on prior exams or there was concern for injury based on post-extubation dysphonia or airway restriction symptoms [38, 39].

Laryngeal webbing may also present as a secondary sequela to prolonged intubation or surgical intervention, particularly when the mucosa is disrupted bilaterally, allowing web formation. Congenital laryngeal webbing has also been reported but is considered to be rare [40]. Clinical presentation ranges from asymptomatic to severe dysphonia with respiratory compromise. In mild cases, surgical intervention may not be warranted, as further disruption of the mucosa may increase the risk of additional scarring [54].

Voice Therapy

Despite implementation of adequate precautions, laryngeal scarring may be unavoidable. Voice therapy is an appropriate initial, noninvasive intervention for treatment of vocal fold scar. While voice therapy may not be completely effective in recovering premorbid vocal function, early intervention by a speech language pathologist is critical for developing optimal voice behaviors and preventing the development of negative compensatory strategies [38, 55, 56]. Therapeutic techniques should focus on improving tissue pliability while eliminating laryngeal tension or supraglottic compression. Attention to deficits in the respiratory and resonance systems may further improve overall system function. The voice therapy trajectory is highly dependent on the severity of the scar, the resources and ability of the family to support therapy, and patient buy-in and compliance. Clinical reports suggest that as the child matures, awareness of their voice disorder and subsequent readiness for change increase. Thus, some patients may pursue therapy years after the initial scar development, and clinical reports indicate that therapy is still beneficial despite scar maturation [55]. Patients who do not see acceptable improvement with voice therapy alone may receive benefit from surgical intervention.

Medical Intervention

There is little consensus on the optimal medical or surgical intervention for vocal fold scarring. Temporary therapeutic options for adults include collagen [57] or fat injectables [58–61] used primarily to increase tissue volume and improve glottal closure. Given the dynamic nature of laryngeal development in the pediatric population, temporary interventions such as injectables may be a first line of approach to mitigate symptoms. More invasive treatment approaches include microcauterization, repeated dilation of the glottis (particularly for webbing), and microflap elevation to release adhesions [38, 62]. Depending on the severity of the fibrosis, introducing a series of vertical incisions across the length of the sulcus or scar may improve voice quality. This technique, developed by Pontes and Behlau, releases the contracted tissue to improve pliability [39, 63]. Despite the seemingly traumatic nature of this treatment, positive outcomes have been reported [63]. Regardless of the type of intervention, care must be taken to prevent additional scarring due to surgical technique; if the mucosa is exposed bilaterally, placement of a laryngeal keel or other temporary prosthesis is recommended to separate the divided tissues and prevent additional scarring or web formation [54].

Emerging Topics

Tissue regeneration for the treatment of vocal fold scarring is an emerging science. An ideal treatment for chronic vocal fold scar should promote an environment that facilitates remodeling of the scarred microstructure to pre-injury lamina propria organization and composition. This normalized microstructure would address the altered viscoelastic state of the vocal folds and in so doing improve vibratory outcomes. Over the past decade, several new therapeutic options incorporating tissue engineering with cells, biomaterial, growth factors, or a combination thereof have emerged [20, 64–68], with the intent to provide an optimal environment to upregulate hyaluronic acid and maximize the deposition, synthesis, and appropriate organization of collagen and elastin in the ECM. Results are encouraging; however, many of these treatments are in early stages of development and are being investigated using in vitro and in vivo animal models. Further, the applicability of many of these interventions has not been investigated in the pediatric population to date. However, research in tissue regeneration to restore native lamina propria tissue properties shows early promise and may result in novel and more effective treatment options for children and adolescents with intractable dysphonia due to vocal fold scar.

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Recurrent Respiratory Papillomatosis

31

Brandyn Dunn, Kevin Huoh, and Hema Desai

Overview

Recurrent respiratory papillomatosis (RRP) is the most common neoplasm of the larynx in children. Although it is considered histologically benign, RRP has the potential for devastating outcomes and is often difficult to manage, requiring multiple medical and surgical approaches. RRP occurs in both children as well as adults; however, it tends to take a more aggressive clinical course in children with potentially fatal outcomes due to its propensity to recur and spread throughout the aerodigestive tract.

Epidemiology

RRP is a rare chronic disease of viral etiology caused by human papillomavirus (HPV), most commonly types 6 and 11, and is characterized by the proliferation of benign squamous papillomas within the aerodigestive tract. RRP is the most common benign neoplasm of the larynx among children and the second most frequent cause of childhood hoarseness [1]. The true incidence and prevalence of RRP are uncertain. It is estimated that between 80 and 1500 new cases of childhood-onset RRP occur in the United States each year [2, 3]. Several large epidemiologic studies, including from Campisi et al. as well as a Danish survey, estimated a national incidence of 0.24 cases per 100,000 to 3.62 cases per 100,000 children, respectively [4, 5]. Additionally, the National Registry of children with RRP, which is comprised of 22 pediatric otolaryngology clinical practice sites, found that children with RRP undergo a mean of 19.7 procedures or an average of 4.4 procedures per year [2, 6]. Ultimately, this is equivalent to more than 10,000 surgical procedures annually for children with RRP in the United States.

Pathophysiology

HPV belongs to the *Papovaviridae* family. It is a small, icosahedral (20-sided), capsid virus without an envelope with double-stranded circular DNA. In the 1990s, HPV was confirmed as the causative agent of RRP with the most identifiable airway subtypes being HPV6 and HPV11. Children infected with HPV11 appear to be at higher risk of obstructive airway disease and have a greater likelihood of needing a

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tracheostomy [7–12]. Two other major subtypes of HPV that are associated with mucosal lesions are HPV16 and HPV18. These subtypes are associated with an increased risk of malignancy in the genital and aerodigestive tracts [13].

Although the complete pathogenesis of HPV-induced lesions is not understood, HPV is thought to infect stem cells within the basal layer of mucosa [14, 15]. After infecting the stem cells, the viral DNA can either be actively expressed or remain latent with no clinical or histologic changes. The induction of cellular proliferation is unclear; however, similar to other viruses, the integrated viral DNA reactivates the host replication genes resulting in the production of viral proteins. Ultimately, the development of RRP is thought to be due to the virus's ability to inactivate certain cellular tumor-suppressor proteins [16, 17] along with the activation of epidermal growth factor (EGF) receptor pathway [18]. It is likely that the host immune system plays a significant role in the pathogenesis of HPV-induced lesions [19].

Presentation

Papillomas usually appear on the vocal folds (Fig. 31.1), with hoarseness being the initial presenting symptom. Vocal changes are often dismissed or attributed to other causes such as nodules in young children, resulting in missed or late diagnosis. Stridor is the second symptom to develop. Other symptoms such as chronic cough, recurrent pneumonia, failure to thrive, dyspnea, dysphagia, or respiratory distress are less common. In some cases, though, dysphonia which has been dismissed as probable nodules without visualization of the larynx may progress to cause significant glottic and/or supraglottic airway obstruction (Fig. 31.2). Occasionally, a tracheotomy may be necessary [20].

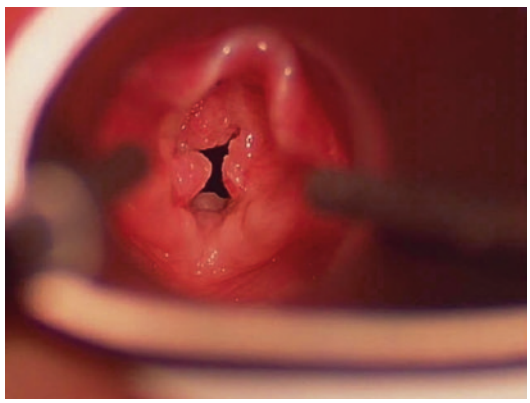

Fig. 31.1 Intraoperative view of endolaryngeal papilloma after exposure with the Lindholm laryngoscope

Speech-Language Pathologist Approach

Collaboration between the speech-language pathologist (SLP) and the pediatric otolaryngologist is vital. While the mainstay treatment for RRP is surgical, the role of the SLP is integral in managing RRP to providing comprehensive care resulting in optimal voice outcomes. The SLP is involved in the ongoing care of children with RRP providing information, support, and help in the avoidance of inappropriate compensatory behaviors and to preserve or restore the voice postoperatively [21, 22].

Evaluation

History

If a child presents with a voice concern in the outpatient setting, a complete history detailing birth, labor, maternal complications during pregnancy/birth, and other concomitant medical diagnoses should be taken. Surgical history to identify procedures that may cause injury to the recurrent laryngeal nerve and previous intubations is also important. Finally, vocal history, looking at time of onset, precipitating causes, chronology, exacerbating or alleviating factors, and severity should be obtained. Additional symptoms such as stridor and dysphagia should also be taken into consideration [23]. In the case of papilloma, some red flags in the history may be hoarseness since birth and stridor or breathing difficulties. When a patient with known papilloma presents for a voice evaluation, knowledge about the

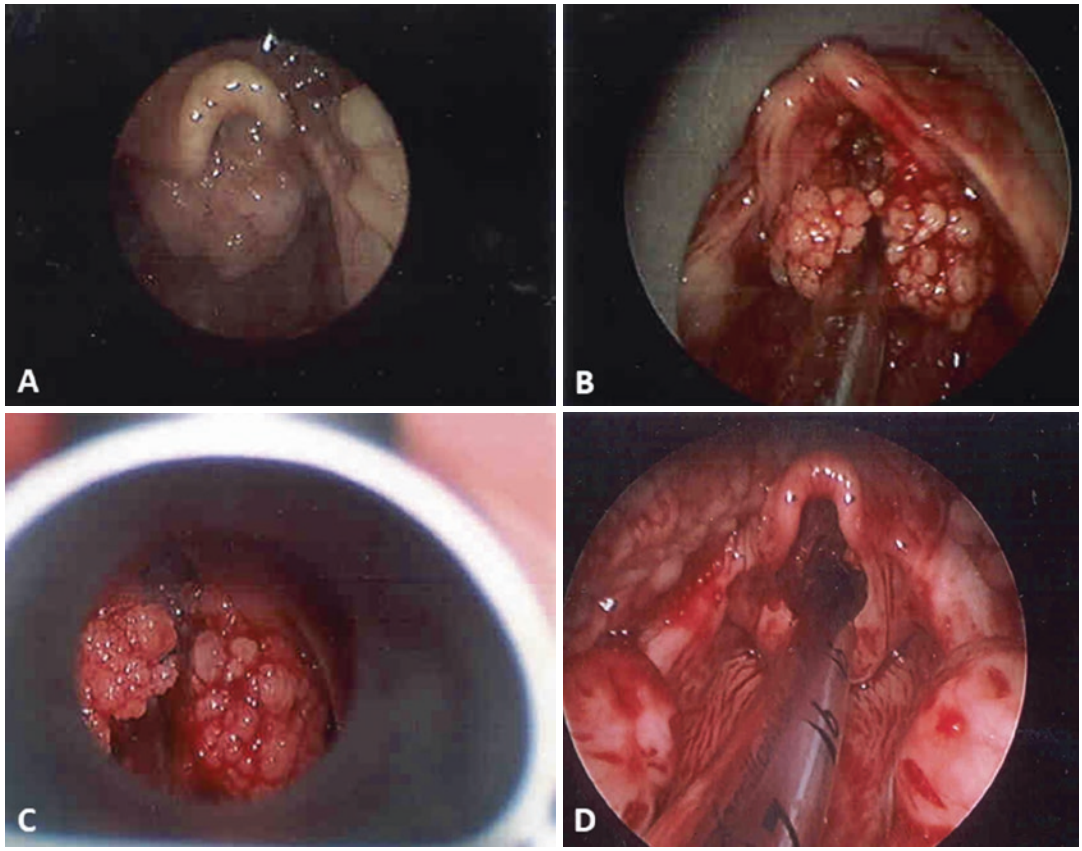

Fig. 31.2 Intraoperative photos of patient who presented with airway distress and was found to have severe supraglottic papilloma. (**a**) Initial view obtained at direct laryngoscopy after child was anesthetized with inhalational anesthetic. (**b**) An endotracheal tube has been passed through the glottis. (**c**) The laryngoscope can be repositioned to view more posteriorly positioned disease. (**d**) The supraglottic disease has been removed with the microdebrider, and the glottis is now better visualized

number, frequency, and type of past surgeries is important, as well as their typical disease course. Current and planned voice use and vocal needs are important as well.

Subjective Evaluation

Quality of Life Tools

Quality of life (QOL) questionnaires such as the Pediatric Voice-Related Quality of Life (PVRQOL) [24] and the Pediatric Voice Handicap Index (pVHI) [25] can provide important information about the impact of the voice disorder. These are both parent/caregiver proxy instruments and may not fully capture the child's perspective on their voice disorder. Adult quality of life instruments may be difficult for children to complete as they are not written at age level. Lindman et al. found discrepancies in adult VRQOL self-rating scale scores of four young children with RRP compared to subjective (grade, roughness, breathiness, asthenia, strain) and objective (acoustic analysis) measures, indicating potential decreased reliability of questionnaires for children with RRP [26].

Perceptual Evaluation

Perceptual evaluation is an important part of the full voice evaluation. The GRBAS [27] scale to assess grade, roughness, breathiness, asthenia, and strain is useful to qualify vocal quality, as is

the Consensus Auditory Perceptual Evaluation of Voice (CAPE-V) [28]. For in-depth discussion of perceptual evaluation of children's voices, see Chap. 12 (Perceptual Evaluation of Voice). Children with RRP have been found to have more hoarse, breathy, and rough vocal quality than age-matched controls [29].

Objective Assessment

Acoustic Analysis and Aerodynamic Evaluation

Acoustic evaluation provides information regarding fundamental frequency, relative average perturbation, jitter, shimmer, noise to harmonics ratio (NHR), frequency range, and spectral/ cepstral measures such as cepstral peak prominence, while aerodynamic evaluation measures maximum phonation time (MPT), mean airflow rate (MFR), and subglottic pressure to assess efficiency of respiratory and phonatory patterns and velopharyngeal and glottic insufficiency [21]. Children with RRP have been found to have significant differences in fundamental frequency, jitter %, MFR, NHR, and MPT compared to age-matched norms [20]. As with other populations who may have severe dysphonia, some acoustic measures may not be reliable or valid if a periodic signal cannot be obtained.

Laryngeal Visualization

Videostroboscopy can be utilized to assess glottal closure, mucosal wave, and symmetry of vocal fold movement. The possibility of RRP is a strong argument in favor of laryngeal visualization for anyone presenting with dysphonia, and not assuming that all hoarseness comes from nodules. While laryngeal visualization while awake can be challenging in very young children, videostroboscopy or high-speed videoendoscopy provides valuable information regarding laryngeal function and the impact of the disease or surgeries on vocal fold closure and vibration and should be attempted. Asymmetric vocal fold mobility and mild delays in closing and opening time have been documented in children with RRP after surgeries [20].

Treatment

Voice therapy to improve hoarseness, resonance, and respiratory patterns and decrease hyperfunctional behavior patterns has been shown to be effective in individuals with benign vocal fold lesions [29, 30]. Although RRP does not have the same pathophysiology as vocal cord nodules and polyps, similar voice parameters are negatively affected, and in some cases this allows for a similar therapeutic approach. A systematic review by Desjardin et al. identified indirect and direct treatment approaches that lead to significant improvement in at least one outcome measure (self-assessment, perceptual judgment, acoustic analysis) [31]. While Jani et al. revealed that a combination of direct and indirect voice therapy would be the best intervention for functional dysphonia rather than no therapy, Speyer et al. found that direct voice therapy resulted in improved outcomes compared to indirect voice therapy [32, 33]. A review of the literature on pediatric voice therapy, as well as a discussion of therapy approaches, is included in Chap. 21 (Voice Therapy). Studies of voice therapy in children have not specifically included patients with RRP, likely due to the variable and fluctuating nature of the disease process, especially in children.

Goals for voice therapy should be tailored to the individual's laryngeal structure and function, as well as their vocal needs. The goals for voice therapy in children with RRP may include the following [22]:

- Education and communication strategies regarding postoperative voice rest
- Information regarding vocal hygiene
- Elimination of hard glottal attack
- Reduction of phonotraumatic behaviors
- Improvement of resonance and tone focus
- Improvement of coordination of respiratory, phonatory, and resonatory subsystems of voice
- Improvements of respiration patterns during speech
- Elimination of maladaptive compensatory behaviors

- Training and support in compensatory strategies to adapt to changing structure and function of larynx

Indirect therapy provides parent and child education about current anatomy and physiology of voice, individualized instruction on internal and external vocal hygiene, recognizing vocal danger zones to monitor speaking in loud environments, and emotionally charged situations due to potential use of loud phonation without conscious technique. Recommendations for amplification to reduce vocal effort and loudness required when speaking in noisy environments can also be made to parents. Using stretches or relaxation to decrease compensatory muscle tension in the head and neck and upper torso can also be beneficial if this is present [34].

Direct voice therapy for children with RRP focuses on decreasing hoarseness from two separate potential causes: an increase in mass of vocal folds from papillomas, interfering with appropriate closure and entrained vibration, and reduced pliability and incomplete closure due to scarring from repeated procedures. As a result, children often compensate by speaking louder (with more force), causing increased effort and strain or compensatory extrinsic muscle activation. Decreasing these hyperfunctional behavior patterns is also addressed in voice therapy to decrease tension in the head and neck and respiratory musculature, as well as optimize vibration of the vocal folds, respiratory support, and use of forward resonance.

Non-conversational techniques promote vocal fold tissue healing from behavioral or surgical trauma, efficient vocal fold vibratory patterns, and increased anterior sensory awareness of voice production. The use of semi-occluded tract postures (straw phonation, vocal function exercises, basic training gestures, tongue trills) and resonant voice therapy has been found to be effective to improve perceptual voice quality and overall efficiency of production [35, 36]. Conversational techniques including Lessac-Madsen Resonant Voice Therapy (LMVRT), Casper-Stone Confidential Voice Therapy (CSCVT), Stretch and Flow Phonation, and Accent Method can be helpful in bridging the gap to connected speech [34]. Specific to the treatment of dyspnea, inspiratory muscle strength training was found, in a single subject, to reduce the perception of dyspnea [37].

In providing voice therapy for children with RRP, a particular challenge is the recurrent nature of the disease and the resultant fluctuating voice quality depending on extent of disease and effects of recent or remote surgeries. Given the sometimes rapid progression, we may be dealing with different laryngeal structure and function at each therapy visit. For example, a child with bulky disease may not be able to achieve entrained vibration for phonation and may instead need to focus on compensatory strategies, including amplification, to communicate effectively. Conversely, a child who is several weeks post-surgery may need to focus on improving respiratory/phonatory coordination and forward resonance to achieve their best voice. Another child may have extensive scarring from repeated procedures and may benefit from work on forward resonance and balanced respiratory support to optimize the vibration they can get. Clinicians need to consistently reassess function as well as stimulability for change in each session. While challenging, this should not be given up on, as voice therapy can be extremely helpful in improving a child's communicative function and quality of life.

Otolaryngologist Approach

History

A thorough history involving both the parent and the patient should be obtained. Persistent or progressive stridor and dysphonia, with the possible development of respiratory distress, are the most consistent signs and symptoms of RRP in children. Focused questions regarding the time of onset of symptoms, possible airway trauma including a history of previous intubations, and characteristics and quality of the cry or voice changes are important. In most pediatrics series, the time from onset of symptoms to diagnosis of RRP is approximately 1 year [2, 38], although the

duration of symptoms before diagnosis varies. The vocal fold is usually the first and predominant site of papillomatous lesions resulting in hoarseness being the principal presenting symptom [39]. Stridor is often the second clinical symptom to develop, initially inspiratory, then becoming biphasic. Less common symptoms include chronic cough, recurrent pneumonia, failure to thrive, dyspnea, dysphagia, or acute respiratory distress. Not uncommonly, a diagnosis of asthma, croup, allergies, vocal nodules, or bronchitis is entertained before a definitive diagnosis is made.

The natural history of RRP is highly variable and unpredictable. The disease may undergo spontaneous remission, persist in a stable state requiring only periodic surgical treatment, or may be aggressive, requiring surgical treatment every few days to weeks and consideration of adjuvant medical therapy.

Exam

Children who present with a history consistent with RRP must undergo a comprehensive physical examination. First, clinical signs of respiratory distress including rapid respiratory rate, degree of distress, signs of fatigue, nasal flaring, and use of accessory muscles must be quickly assessed. If the child appears to be in severe respiratory distress, additional examination should be delayed, and the patient should be transferred immediately to either the operating room (OR), emergency department, or pediatric intensive care unit (PICU) depending on the level of severity. In the stable, well-oxygenated child, additional examination can be carried out. Care should be taken to listen closely to the child's voice and/or cry as well as begin to characterize the child's stridor and its relationship to the respiratory cycle. Using a stethoscope to auscultate over the nose, mouth, neck, and chest can localize the location of upper airway obstruction. A child with RRP would not be expected to demonstrate much change in the stridor with position change, in contrast to infants with laryngomalacia, a vascular ring, or a mediastinal mass.

Instrumented Assessment

The diagnosis of RRP is best made with flexible fiberoptic nasopharyngoscopy. Systematic inspection of the pharynx, hypopharynx, and larynx including the subglottis provides critical information necessary for the diagnosis and can assist with preoperative planning. Evaluating airway lumen size and vocal fold mobility helps to determine the urgency of operative intervention. Although dynamic evaluation can be performed when children are spontaneously breathing, endoscopy in the OR under anesthesia is warranted in any child suspected to have RRP who cannot be fully examined in the outpatient setting [40].

Staging systems are helpful in tracking disease progression in individuals and communicating with other professionals. Derkay et al. created a numeric scoring system for evaluating clinical and anatomic disease severity [41, 42]. Clinical symptoms of dysphonia, stridor, intervention urgency, and respiratory distress are scored. Anatomic subsites of the larynx (lingual epiglottis, laryngeal epiglottis, anterior commissure, aryepiglottic folds, false vocal folds, ventricle, true vocal folds, arytenoids, and posterior commissure) are graded on a scale of 0–3, where 0 is none, 1 is surface lesion, 2 is raised lesion, and 3 is bulky lesion. A final numeric score is calculated to determine the extent of disease at each assessment (Figs. 31.1 and 31.2). Airway endoscopy is crucial to determine the full extent of disease and monitor treatment response.

Differential Diagnosis

As stridor and dysphonia are the most common presenting symptoms, differential diagnosis for RRP is broad including benign laryngeal or tracheal tumors, malignant laryngeal or tracheal tumors, foreign body aspiration, gastroesophageal reflux disease, laryngitis, subglottic stenosis, tracheomalacia, vocal fold dysfunction, and vocal fold paralysis.

Management

At present, there is no cure for RRP, and no single treatment has consistently been shown to be effective in eradicating RRP. The current standard of care is surgical intervention with a goal of complete removal of papilloma with preservation of normal structures. In patients with disease burden in sensitive locations, including the anterior or posterior commissure, or highly aggressive disease, the overall goal is removal of sufficient disease to clear the airway while preserving normal structures and avoiding complications of glottic or subglottic stenosis and webbing.

When RRP presents with severe respiratory distress caused by papilloma obstructing the airway, tracheostomy may need to be performed. It has been suggested that tracheostomy may activate or contribute to the spread of disease lower in the respiratory tract [43]. As a result, most otolaryngologists agree that a tracheostomy is a procedure to be avoided unless absolutely necessary, and, when tracheostomy is unavoidable, decannulation should be considered as soon as the disease has been managed effectively.

The carbon dioxide (CO_2) laser is now preferred by some over previously used cold instrumentation for removal of RRP involving the upper airway [44]. The use of the CO_2 laser with an operating microscope has gained popularity due to its excellent precision with minimal bleeding. Multiple procedures performed over time are recommended to avoid tracheostomy and allow for optimal phonation with preservation of normal laryngeal anatomy.

In addition to the CO_2 laser, the potassium titanyl phosphate (KTP) laser can also be used [45]. The 532-nm wavelength selectively targets hemoglobin and coagulates the vascular supply to the papilloma. This allows for spontaneous involution of the lesions postoperatively and decreases the probability of scar or web formation that may occur with complete lesion removal [45]. A recent study demonstrated that HPV DNA was not present on the laser fiber after the procedure, decreasing some concern for potential transfer of disease from patient to surgeon [46].

While there are many advantages to the laser in patients with RRP, the drawbacks relate primarily to safety. Although uncommon, the laser beam may reflect off nearby metal resulting in potential injury to the surgeon or areas of the patient that are not protected by a wet towel. Additionally, the laser smoke or "plume" has been found to contain active viral DNA, a potential source of infection [47–49]. The most dreaded safety concern is that the laser beam generates heat that, if the beam inadvertently strikes the endotracheal tube in the oxygen-rich environment, could lead to an explosion or fire in the airway. Employing standard laser safety precautions can help mitigate these potential risks. First, all team members in the room should wear eye protection, and the patient's face and shoulders should be covered with wet towels. Clear closed loop communication should be performed between the surgeon and assistant who is operating the laser, specifying "laser on" and "laser on standby" when warranted. Oxygen should be kept at 30%, and a laser-safe endotracheal tube should be used.

An alternative technology which can be incorporated in the therapeutic regimen for pediatric RRP is the microdebrider. Initially adapted from the sinus microdebrider, the laryngeal microdebrider is effective in removing bulky, exophytic papillomatosis disease with reduced postoperative pain scores, improved voice quality, shorter procedure time, and decreased procedure cost compared to the CO_2 laser [50]. Although uncommon, significant complications including major vocal fold scar, airway compromise, severe hemorrhage, and unintentional tissue loss have occurred [51].

Operative Approach: Microdebrider Excision of Recurrent Respiratory Papilloma

Indications

In pediatric RRP, the normal airway lumen is inherently small and thus maintaining adequate

airway patency is of the utmost importance. For this reason, parent education regarding early signs of airway obstruction as well as compliance with regular clinical and endoscopic monitoring is crucial. A key management principle in RRP is to focus attention and effort on preventing the need for a tracheostomy. Indications for surgical intervention include any voice, swallowing, or airway symptoms.

Key Aspects of the Consent Process

Risks associated with microdirect laryngoscopy should be discussed, including injury to the lips, gums, tongue, and teeth, as well as potential for transient or longer-lasting dysgeusia. Voice or swallow function could worsen or simply fail to improve. It may not be possible or safe to remove all disease, which may predispose to earlier recurrence. Operating on the airway carries an inherent risk of airway edema which may require overnight observation, temporary placement of an endotracheal tube, or, in rare cases, tracheotomy. Particularly relevant for papilloma is the risk of inducing a web, which is avoided by not treating the medial surface of both vocal folds at the same time.

Equipment

An operating laryngoscope with its associated suspension system is utilized for optimal exposure. Both rigid 0- and 70-degree Hopkins rod telescopes are used for intraoperative exam and photodocumentation. An operating microscope is used for the microsurgical portion. A full set of microlaryngeal instruments should be available. An ENT Microdebrider with laryngeal blade attachment is needed.

Steps

1. *Patient positioning.* The patient is placed supine in the sniffing position.
2. *Exposure and suspension.* A Benjamin-Lindholm laryngoscope is advanced into the valleculae and lifted to visualize the endolarynx (Fig. 31.1). The Lewy suspension arm is attached and secured onto the Mayo stand.
3. *Intraoperative examination.* Once the patient is in suspension, inspection (visual and by palpation) is performed, and photodocumentation is obtained. This is done with both high-power microlaryngoscopy along with Hopkins rod telescopes (0- and 70-degree rigid telescopes). The telescope should be advanced beyond the larynx to evaluate for tracheal and bronchial disease (Fig. 31.3).
4. *Microdebrider removal of RRP.* The smallest and most conservative microdebrider blade should be placed on the microdebrider handle, especially at the start of the case. Generally, a straight or angled Skimmer® Laryngeal Blade for the Straightshot M4 Microdebrider is used. This is especially true for the subglottis, glottis, and posterior glottis. The safest method for RRP removal is to hold the microdebrider "blade or port" 1–2 mm over the RRP disease and allow the suction from the instrument to draw the RRP tissue away from the deeper aspects of the laryngeal tissue and be removed by the internal blades of the microdebrider. It is often helpful to "pin" the vocal fold in a stationary position with a blunt probe or suction (Fig. 31.4) to prevent the deeper tissues (e.g.,

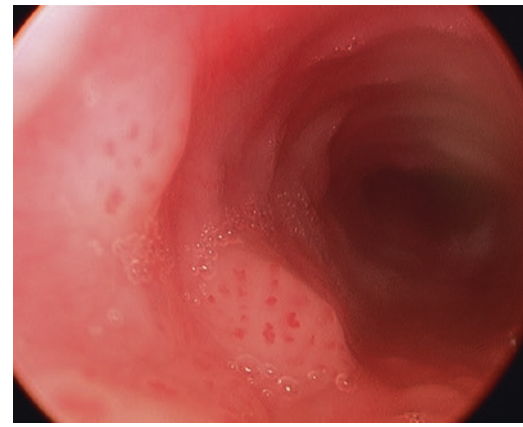

Fig. 31.3 Careful tracheobronchoscopy at the time of operative intervention is essential to evaluate for more distal disease, as shown here with papilloma affecting the left lateral and posterolateral tracheal walls

lamina propria) from being suctioned into the microdebrider chamber. It is especially helpful to begin microdebriding from inferior to superior to minimize difficulties with visualization from bleeding raw mucosal edges.

5. *Surgical approach in setting of bilateral true vocal fold disease.* In patients with papilloma on the bilateral true vocal folds, a careful intraoperative exam is required to develop an appropriate operative plan (Fig. 31.5). It is critical to avoid treatment of the anterior medial surfaces of the bilateral true vocal folds, as that will create apposing raw surfaces and place the patient at risk for a glottic web formation. The papilloma should be carefully examined to identify its underlying base. If there is bilateral disease on the medial surfaces at the anterior commissure, one side should be treated with the first procedure and the second side treated approximately 6 weeks later.
6. *Hemostasis.* Apply epinephrine-soaked or oxymetazoline-soaked pledgets to the surgical site to obtain hemostasis after removal of the RRP.
7. *Pathology Specimen.* To obtain RRP tissue for pathologic examination, a suction trap can be placed "inline" with the microdebrider suction and at the end of the procedure sent for pathologic examination. For the initial surgery for a patient with newly diagnosed RRP, a cup forceps can be used to remove tissue for histologic analysis and viral subtyping.

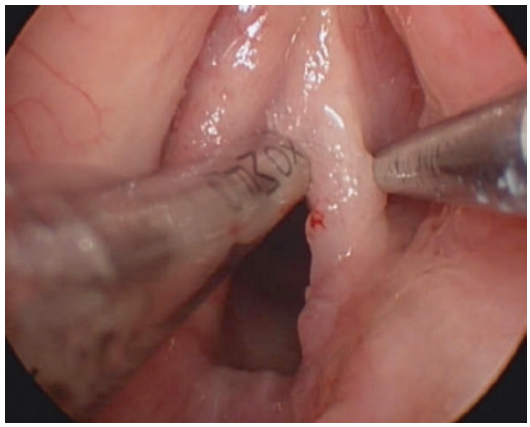

Fig. 31.4 The suction or a blunt right-angle probe can be used to manipulate the vocal fold to allow for better access to infraglottic and medial surface disease. Importantly, the debrider should be used to remove only the exophytic disease and not violate the underlying lamina propria

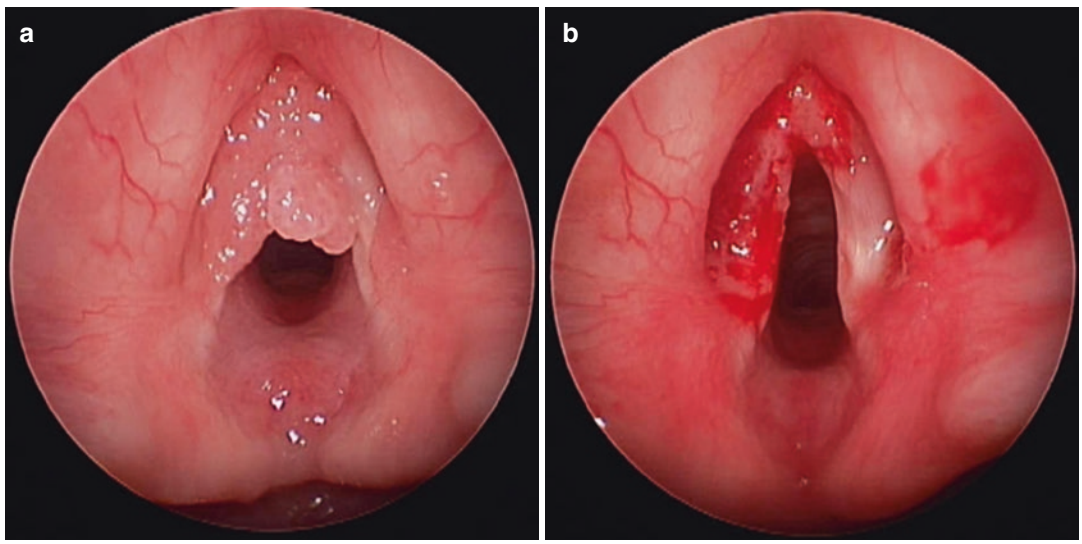

Fig. 31.5 (**a**) Bulky papilloma is seen emanating from the anterior commissure. Careful examination revealed that majority arose from the left true vocal fold. (**b**) The left-sided disease has been removed, and disease at the right side of the anterior commissure has been left in place, to be removed at a second stage

Operative Approach: KTP Laser Ablation of Recurrent Respiratory Papilloma

The indications and key aspects of the consent process are similar to those for the microdebrider. Use of the KTP laser may be particularly helpful if the patient has predominantly surface lesions rather than bulky disease. Additional risks associated with the laser include airway fire and inadvertent damage to surrounding structures. Risks of scar and web formation should be discussed.

Equipment

A KTP laser is used with settings of 35 W, 15 ms per pulse, and 2 pulses per second. A 0.4 mm fiber is prepared by passing it through disposable suction tubing and then a size 7 laryngeal suction. After passing the fiber through the suction, the tip is stripped with the 0.0125″ stripper. The fiber tip is extended approximately 1 cm beyond the suction tip for visualization during the procedure. The spot is tested to ensure the fiber was stripped cleanly. If the spot had blurred edges or aberrant extensions, the fiber should be cut and re-stripped.

Laser safety precautions are ensured, including moistened eye pads, moistened towels/surgical drapes, a laser-safe endotracheal tube, and eye projection for operating room personnel. An additional laryngeal suction or suction port on the laryngoscope is used to suction the laser plume.

Steps

1. *Positioning.* Patient positioning is the same as for the microdebrider approach. The eyes, face, and exposed neck and chest are protected with moist towels.
2. *Exposure.* The larynx is exposed with the Lindholm laryngoscope seated in the valleculae, and the patient is placed into suspension.
3. *Laser safety precautions are ensured.* FiO_2 is lowered to 30%. The patient's face is protected. Laser safe goggles are worn by all people in the room. Clear communication is used between surgeon and assistant at the laser to state when the laser is on and off. Saline-soaked pledgets are placed in the subglottis and against the contralateral vocal fold.
4. *Equipment.* The KTP fiber in the seven laryngeal suction is passed through the laryngoscope and positioned just superior to the lesion of interest. Laser energy is applied to ablate the papilloma at settings of 35 W, 15 ms per pulse, and two pulses per second. Laser plume is removed with a second suction.
5. *Laser treatment of papilloma.* Care is taken to avoid delivery of laser energy to healthy adjacent epithelium or underlying lamina propria. Laser energy is applied in a systematic posterior to anterior, medial to lateral fashion. In the setting of exophytic lesions, superficial portions can be suctioned away after initial ablation to allow for treatment of deeper disease. In the case of sessile surface lesions, lesions can be treated until blanching occurs, which indicates coagulation of the feeding vasculature. It is preferable to leave a thin layer of blanched papilloma remaining rather than remove all visible disease, as any blanched lesions will spontaneously involute over time. If disease is present on the medial edges of both vocal folds, only one should be treated at a time, and the second can be treated with a separate procedure about 6 weeks later.
6. *Laryngotracheal anesthetic.* At the conclusion of the procedure, a laryngotracheal anesthetic is applied.

Postoperative Management and Follow-Up

Intraoperative or immediate postoperative intravenous steroids are often used to reduce postoperative edema. Depending on the extent of the disease process, most cases are considered an outpatient surgery or admitted to the hospital for one night to monitor for any signs or symptoms of airway obstruction. Although somewhat controversial, most clinicians encourage voice rest or

limited voice use for 3–5 days followed by gradual return to full voice for those patients who can adhere to it. Postoperative laryngopharyngeal reflux (LPR) treatment including both proton pump inhibitors and behavioral modifications is often recommended to reduce additional inflammation during the immediate postoperative healing process. Patients will generally follow up outpatient with the otolaryngologist in 4–6 weeks for clinical evaluation including flexible laryngoscopy. Regular clinic visits along with outpatient flexible laryngoscopy are crucial to monitor disease progression and safely plan for future surgical interventions.

Emerging and Evolving Techniques of the Future

Adjuvant Treatment Modalities

Although surgical management remains the primary treatment modality for RRP, some form of adjuvant therapy may be needed in up to 20% of cases [44]. The most widely accepted indications for adjuvant therapy are a need for more than three or four surgical procedures per year, rapid regrowth of papilloma with airway compromise, and distal multisite spread of disease [2]. Current adjuvant therapies range from immunomodulation, disruption of HPV replication, control of inflammation, and prevention of angiogenesis.

Interferon (IFN) therapy was one of the first systemic adjuvant treatments used to manage RRP [52]. Despite positive results regarding the efficacy of IFN, it is rarely used due to the emergence of intralesional adjuvants such as cidofovir and bevacizumab, which have fewer local and systemic side effects.

The antiviral cidofovir is a cytosine nucleotide analog that blocks replication of DNA viruses by inhibiting viral DNA polymerase [53]. The mechanism of action against HPV is not well understood; however, it is hypothesized that it acts by augmenting the immune system or induces apoptosis [54]. Intralesional administration of cidofovir has been fairly well tolerated, with limited systemic toxicity; however, there are a few reported cases of dysplasia after intralesional administration [55–59]. In 2013, the RRP Task Force released an 18 statement consensus regarding intralesional cidofovir for RRP in adults and children. In general, cidofovir use is recommended in patients requiring surgical debulking at least every 2–3 months with routine intraoperative biopsies performed [60].

More recently, bevacizumab has gained popularity in adjuvant treatment of RRP. Bevacizumab (Avastin) is a recombinant monoclonal humanized antibody that blocks angiogenesis by inhibiting human vascular endothelial growth factor A (VEGF-A) [53]. Several studies have been performed since the FDA approval of bevacizumab in 2004 revealing improved disease control along with increase time between procedures. Specifically, Rogers et al. and Sidell et al. showed that the combination of potassium titanyl phosphate (KTP) laser and intralesional bevacizumab yielded improved surgical interval along with decrease disease burden [61, 62]. In a recent case series by Best et al., systemic bevacizumab showed significant lengthening of surgical interval for patients with advanced, treatment-resistant papillomatosis having tracheal and pulmonary spread with a low complication profile [63].

HPV Vaccination

One of the newer developments in the management and prevention of RRP is the HPV vaccination. Gardasil, the quadrivalent HPV vaccine, is directed against both low-risk HPV type 6 and 11 and high-risk HPV type 16 and 18. Since HPV6 and HPV11 are the predominant etiologic factors for RRP, Gardasil has been primarily used to manage and prevent RRP [50]. While there are currently no multicenter randomized controlled trials to fully assess the efficacy of HPV vaccination, there are several small studies and case reports with promising results showing increased time between surgical interventions along with decrease in number of surgical interventions required annually [64–67]. Additionally, given the modern trend to vaccinate pre-adolescent females and males, many

researchers predict a reduction in the incidence of secondary laryngeal infections to newborns via vertical transmission [68].

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Glottic Web

32

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Overview

Laryngeal webs may be congenital or, more often, acquired. Broadly, a glottic web refers to a band-like membrane of variable thickness and composition that occurs between the vocal folds and may have extension into the subglottis. Both upper airway obstruction and phonation must be considered in the diagnosis and management of glottic webs. Understanding the etiology, composition, and thickness of the web is critical to proper management. This chapter describes the definitions, classification, and clinical characteristics of glottic webs as well as their treatment, including both nonoperative and operative approaches. While we have focused on congenital glottic webs in this chapter, important distinctions and considerations for acquired glottic webs are also included.

Definitions

A glottic web is a membrane between the vocal folds with a length of 1 mm or greater and with any depth [1]. Seymour Cohen developed a classification of laryngeal webs based on the percentage of glottic involvement and the presence or absence of subglottic extension (Table 32.1) [2].

The first description of a congenital laryngeal web was by Fleischmann in 1892 with the autopsy of an infant. Congenital laryngeal webs may be glottic (75%), subglottic (12%), or supraglottic (12%). An example of a congenital glottic web is depicted in Fig. 32.1. Congenital glottic webs are diagnosed in patients without previous intubation or airway trauma and are often associated with chromosome 22q11.2 deletion [3].

Acquired glottic webs are essentially synechiae between the vocal folds and occur most frequently in the anterior glottis. These webs develop following a traumatic insult to the bilateral true vocal folds which puts two raw mucosal surfaces in apposition, such as laser treatment of glottic lesions, laryngeal trauma, or infection. An acquired, or iatrogenic, glottic web in a young child following intubation is depicted in Fig. 32.2.

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Table 32.1 Classification of glottic webs

Classification	Percentage of glottis	Characteristics	Breathing	Voice	Treatment
Type I	<35%	TVF easily seen in web	Clear, unobstructed	Slight hoarseness	Often not required; dilation or excision via various techniques may be effective
Type II	35–50%	Anteriorly thick, minimal subglottic involvement; TVF usually visible within the web	Symptomatic with infection, exertion, inflammation	Deep, husky, weak	May be required via serial procedures; endoscopic keel placement and mucosal flap have been described
Type III	50–75%	Thick, anteriorly solid (may thin posteriorly) with subglottic extension or stenosis	Moderately severe obstruction	Weak, whisper	Placement of a keel, mucosal flap, and LTR are possible options
Type IV	75–90%	Densely thick anteriorly and posteriorly, no visible true vocal folds, with subglottic extension and stenosis	Severe obstruction	Aphonic; posttreatment voice results often poor	Urgent/emergent tracheostomy; strong consideration of external approach: laryngofissure with keel or LTR

From Cohen [2], with permission

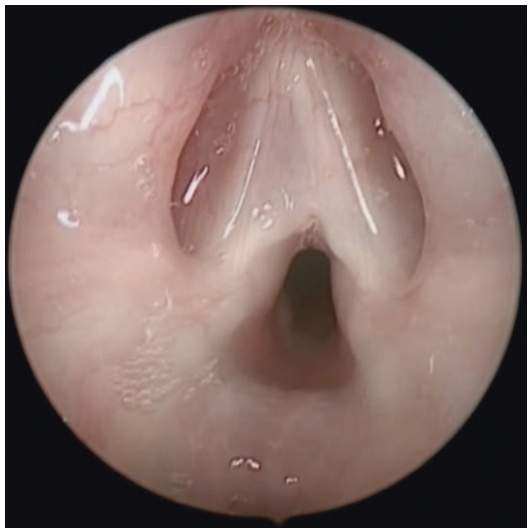

Fig. 32.1 Congenital glottic web

Note: Intentional anterior glottic web formation for the purposes of changing vocal pitch is outside the scope of this chapter and will not be reviewed. The reader is referred to literature describing this procedure for the purpose of voice feminization.

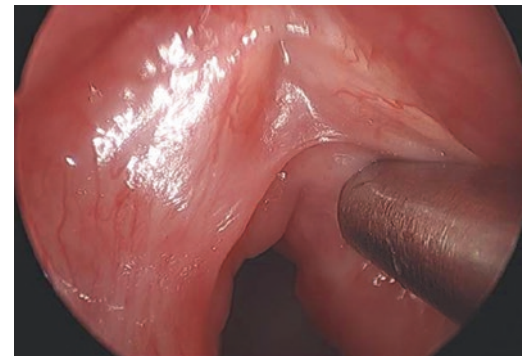

Fig. 32.2 Acquired glottic web following intubation

Epidemiology

Congenital glottic webs are rare and thought to comprise 5% of congenital laryngeal anomalies [2]. Patients with congenital anterior glottic webs also often have a diagnosis of 22q11.2 deletion syndrome, commonly known as velocardiofacial syndrome or DiGeorge syndrome [3]. In the largest published series, 11 (65%) of 17 patients with anterior glottic webs tested positive for the

chromosome 22q11.2 deletion and demonstrated clinical manifestations of velocardiofacial syndrome [3]. As a result, patients who are diagnosed with an anterior glottic web should be counseled regarding this association and given the opportunity to pursue genetic analysis.

Acquired glottic webs are more common than congenital glottic webs and tend to occur in the older child and adult populations. Rarely, a small, asymptomatic congenital web will be incidentally found at the time of intubation in an adult patient. Because thin congenital webs may be obliterated at the time of intubation during infancy, the true incidence of small anterior webs is unknown. In contrast, thicker webs with subglottic extension are more commonly symptomatic and do not resolve following intubation alone.

Pathophysiology

Congenital glottic webs may be conceptualized on a spectrum of embryologic recanalization failure, the most extreme being complete laryngeal atresia. Figure 32.3 depicts near-complete laryngeal atresia. While mild webs include gossamer-thin anterior bands that occur without significant posterior or inferior extension, intermediate webs are thicker in nature and extend into the subglottis.

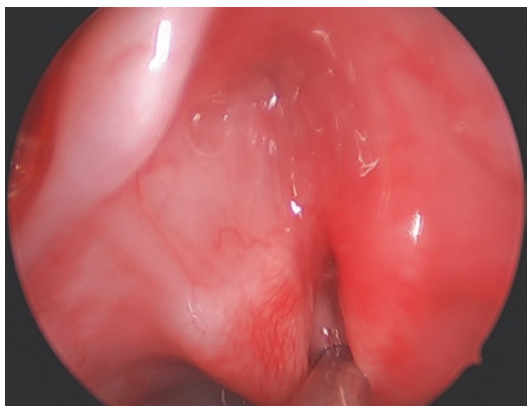

Fig. 32.3 Type IV glottic web demonstrating near-complete laryngeal atresia. Rigid laryngeal suction is larger than any discernable glottic opening

These webs, which are often considered partial laryngeal atresia, have a mucosal extension or "subglottic sail" that extends to the inferior border of the cricoid ring and is often composed, in part, of cartilage. This distinction has important implications when considering treatment options and when providing counseling regarding postoperative expectations [2, 4–6].

Speech-Language Pathologist Approach

Infants with congenital glottic webs frequently present with dysphonia, even in the absence of breathing difficulties. The speech-language pathologist (SLP) may detect a high-pitched cry, weak or absent cry, or intermittent breathy dysphonia during the feeding evaluation of an infant or during a language assessment for a young toddler. A referral to otolaryngology is warranted in these situations.

To date, there are no published data demonstrating perceptual ratings or acoustic measurements in cohorts of children with different types of glottic webs. In 1985, Cohen et al. found that patients who presented with a thin glottic web had more improvement in voice after surgical intervention; however, this study did not include perceptual measures or acoustic data as outcome measures. Aphonia was also noted in patients whose web involvement ranged from 50% to 90% of the glottis [2].

Unfortunately, there is also a paucity of data in the literature pertaining to postsurgical voice outcomes for patients with congenital or acquired glottic webs. One small study of 11 patients by de Trey and colleagues indicated voice improvement in 10/11 patients after surgery; however, no standardized voice measures or questionnaires were used for this young population under the age of 3 years [7].

History

The SLP should take a detailed history of the child's voice concerns as well as voice use. In

addition, the SLP will also take into account a thorough airway history, including previous intubation events, history of intubation difficulty, as well as history of tracheostomy. Understanding the patient's voice history and the chronic versus intermittent nature of the dysphonia is essential. Given the occurrence of glottic webs in the population of children with 22q11.2 deletion, the SLP should elicit any syndromes or genetic evaluations in the medical history.

Quality of Life Measures

For children with less severe glottic webs, it is essential for the SLP to understand the impact of the voice disorder on the child and the family. Some families may not wish to pursue surgical intervention for less severe dysphonia. For children with congenital glottic webs, their dysphonia may be considered part of "who they are" by family members. Conversations regarding the risks and benefits of surgery to improve pitch or volume should occur as a team approach among the family, SLP, and otolaryngologist.

Laryngeal Visualization

Flexible laryngoscopy is often sufficient to identify the presence of a glottic web in most children. In the newborn, small, less-symptomatic webs may be difficult to identify, and close outpatient follow-up is thus recommended in the first weeks of life. Laryngostroboscopy may be used frequently in children over 8 years of age and in some children as young as 4 years of age who are able to participate. Stroboscopic measurements such as mucosal wave and closure pattern can be noted outside of the affected area of the glottic web in Type I and Type II classifications. Laryngeal hyperfunction can be identified on laryngostroboscopy and serve to direct therapy prior to possible surgical intervention for those children who are candidates for behavioral intervention, based on developmental ability and age.

One case study in the literature describes a 5-year-old female who presented with complaints of a high-pitched voice and chronic dysphonia with previously undiagnosed anterior Type II glottic web. She demonstrated laryngeal hyperfunction and lack of mucosal wave within the web on laryngostroboscopy [8]. Postsurgical perceptual and acoustic measurements indicated improvement in pitch range and normalization of her perceptual habitual speaking pitch [8].

In addition to laryngoscopy to identify possible glottic web, patients who present with hypernasal resonance secondary to 22q11.2 deletion syndrome may also undergo nasopharyngoscopy to evaluate palatal movement, depending on their ability to produce connected speech during that procedure.

Treatment

Patients who present with Type I or Type II glottic web and laryngeal hyperfunction may benefit from a trial of voice therapy using one or more of the voice therapy approaches outlined in this text. When considering surgical intervention, voice therapy to "unload" the hyperfunction may help the child understand their compensatory voice patterns and achieve improved voice outcomes after surgery.

Perioperative Voice Therapy

While there is little documented regarding pediatric postoperative voice outcomes for patients with congenital glottic web, children who present with laryngeal hyperfunction secondary to Type I or Type II glottic web will benefit from continued therapy postoperatively to unload the hyperfunction. Given the high percentage of patients who present with voice symptoms at birth and shortly thereafter, the majority of children who require surgical intervention will undergo a procedure at a young age. These children should return for intermittent postoperative voice evaluations based on their age and ability to participate in assessment.

In our experience, children who present with more severe glottic webs and extensive subglottic obstruction will often continue to use supraglottic or mixed glottic and supraglottic phonation postoperatively as their most effective means of communication. Laryngoscopy and voice assessment in conjunction with otolaryngology will determine if the patient is able to produce pure glottic phonation. In these cases, stimulability during assessment will guide therapy recommendations.

Otolaryngologist Approach

History

Congenital glottic webs often present with abnormal cry, biphasic stridor, and respiratory distress in infancy or early childhood. It is estimated that 75% of patients with symptomatic congenital glottic webs will present at birth and nearly all by 1 year of age. The most common presenting symptom is an abnormal cry, which may range from high pitched to deep or even absent. As the infant grows and glottic airflow increases, respiratory manifestations such as stridor and retractions may become apparent. The degree of respiratory distress is grossly correlative to the degree of glottic and subglottic obstruction.

Acquired glottic webs present similarly, but the course is variable in progression and symptoms occur following some airway insult. Both congenital and acquired webs can present with dysphonia, dyspnea, and recurrent respiratory infections secondary to impaired airway clearance. At times, glottic webs may present only after a second concomitant laryngeal insult such as infection or laryngopharyngeal reflux. Webs may also be discovered at the time of endotracheal intubation, where an unanticipated glottic web can lead to difficult airway management.

In the non-emergent setting, obtaining a history should include the perception of the child and parents. Evaluation in a multidisciplinary Aerodigestive Center is optimal for concerted evaluation and management preoperatively and to optimize reconstructive outcomes postoperatively.

Exam

A general head and neck exam is performed. In addition, several aspects of an extended physical exam are included for the patient presenting with congenital anterior glottic web as these may be associated with chromosome 22q11.1 deletion syndromes: velopharyngeal insufficiency, cleft palate, otitis media, dysphagia, cardiovascular malformations, behavioral and developmental disabilities, and renal anomalies. As described by Miyamoto et al., anterior laryngeal webs may be the only indication of velocardiofacial syndrome, and it is reasonable to refer all children with anterior glottic webs for genetic evaluation [3].

Cardiopulmonary exam is essential including auscultation and evaluation for extremity clubbing (in patients presenting outside of infancy). If the child is able, pulmonary function tests may also be conducted to characterize the baseline degree of extrathoracic airway obstruction. There may be a flattening of both the inspiratory and expiratory portions of the flow-volume loop if the glottic web impairs respiratory flow.

Occasionally, glottic webs are diagnosed at the time of difficult intubation as an emergent consult to the otolaryngologist. In these instances, it is reasonable to maintain spontaneous ventilation while performing microdirect laryngoscopy and bronchoscopy. The otolaryngologist should have rigid and fiber-optic intubation equipment and smaller-than-anticipated endotracheal tubes on hand. Consideration of a laryngeal mask airway as a rescue device can also be considered, depending on the nature of the glottic web. Lastly, tracheostomy may be necessary to secure the airway if the glottic and subglottic airway will not permit passage of an endotracheal tube.

Instrumented Assessment

In the office setting, videolaryngostroboscopy, perceptual measures, acoustic measures, aerodynamic measures, and quality of life questionnaires, such as the Pediatric Voice Handicap Index, should be completed. Videostroboscopic

examination should be carried out for voice and anatomic evaluation. Rigid laryngoscopy may be completed with a 70-degree laryngoscope if patient age and cooperation allow. The standard stroboscopic protocol, including vocal tasks such as habitual pitch production, pitch glide, and respiratory tasks, is completed and recorded. Essentially, a glottic web will shorten the vibratory margin of the true vocal folds which can lead to higher pitch. Laryngeal hyperfunction and posterior glottic flaring may also be evident [8]. The mucosal wave and amplitude may be difficult to delineate, especially in thick glottic webs. Because many children will not cooperate with rigid videostroboscopy, endoscopic flexible stroboscopy is an acceptable alternative. During flexible transnasal laryngoscopy (with or without stroboscopy), attention is also paid to any evidence of velopharyngeal insufficiency (VPI) via nasopharyngoscopy. VPI has been reported to be concurrent with glottic web in patients with 22q11.2 deletion syndromes. VPI may result in hypernasal resonance despite adequate treatment of the glottic web.

Operative endoscopy with microdirect laryngoscopy and bronchoscopy is essential to diagnose and fully characterize the glottic web and is ideally done with both zero-degree and angled rigid telescopes as a staging procedure prior to deciding upon further treatment. This affords evaluation of web thickness, involvement of the conus elasticus and subglottis, and Cohen classification grade. A right-angled probe is used to evaluate for concurrent laryngeal cleft and can also be used to palpate behind the anterior aspect of the web to assess the cartilaginous component within the subglottic lumen. The arytenoids are palpated to ensure normal joint mobility, with the caveat that advanced glottic webs may limit arytenoid movement. Nonetheless, normal posterior glottis has been proposed as a favorable factor for endoscopic management of anterior glottic webs [6]. A comprehensive airway evaluation is essential so as to identify concurrent pathologies such as subglottic stenosis, tracheoesophageal fistula, tracheomalacia, and laryngeal cleft. Airway sizing should also be performed in this setting, and information regarding recommended airway management should be given to the parents and documented in the medical record. Photodocumentation is strongly encouraged at each operative evaluation and intervention. Of course, if the child is experiencing respiratory failure or severe distress, securing the airway takes precedent over staging the glottic web which can be conducted at a later time.

Differential Diagnosis

The differential diagnosis for glottic webs most commonly includes subglottic stenosis in those patients presenting with stridor or respiratory insufficiency. Subglottic stenosis may also be present as part of the inferior extent of thick anterior glottic webs. More severe forms of laryngeal atresia are also included in the differential diagnosis if suggested by the clinical presentation. Patients with cri-du-chat syndrome may have similar vocal presentations to children with an anterior glottic web.

Management

Proper management of glottic webs rests on making an accurate diagnosis and fully characterizing the lesion. An estimated 60% of congenital glottis webs require operative intervention. From an operative standpoint, both open and endoscopic management techniques have been described. Historically, the gold standard for operative management included an anterior thyrotomy and tracheostomy. However, in 1991, Lichtenberger developed an endo-extralaryngeal needle holder which afforded the placement of a silicone keel into the anterior glottis after lysis of the web while obviating the need for tracheostomy [9]. Currently, the decision for open versus endoscopic techniques rests largely on the characteristics of the web and surgeon preference. The extent of reconstruction is largely determined by the subglottic characteristic of the web. Current literature suggests that the web thickness, specifically with subglottic extent, may be the most important

consideration in choosing operative technique. Grossly, thinner webs need less invasive surgical technique. Specifically, thin glottic webs may respond well to simple endoscopic lysis with or without adjunctive dilation or microsurgical mucosal flap procedures [10, 11], whereas uniformly thick webs with subglottic extension will often require more advanced techniques such as laryngotracheal reconstruction with stenting or laryngofissure with keel placement (Fig. 32.4) [12, 13]. Such webs are most often seen in Cohen Types III (Fig. 32.5) and IV (Fig. 32.6); however, glottic extension is not always correlative with subglottic extension. In those webs with a thick subglottic "sail," the laryngeal surface of the anterior cricoid cartilage is the extent of web incision, and therefore, an anterior or anterior/posterior graft is often needed to address the respiratory symptoms and decrease relapse. Treatment of large glottic webs with significant subglottic extension by laryngotracheal reconstruction in the neonatal period is controversial. While some authors advocate early intervention, others maintain that definitive surgical correction should be undertaken at a later age [5, 14–16]. Considerations including patient comorbidities, swallowing dysfunction, and pulmonary status must always be considered. For that reason, it is the opinion of the authors that patient-specific characteristics are the most important consideration when determining the age of repair.

Whereas Type I and Type II anterior glottic webs do not often require surgical intervention

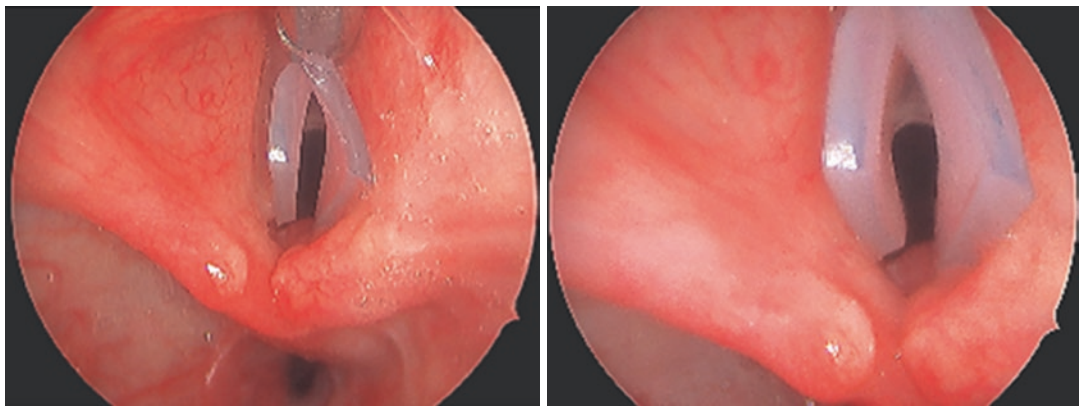

Fig. 32.4 Intraoperative images with keel in position

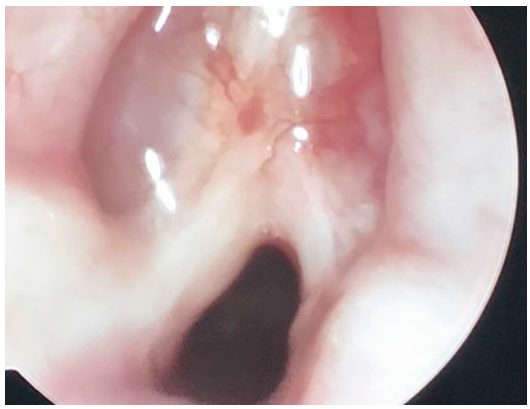

Fig. 32.5 Type III glottic web

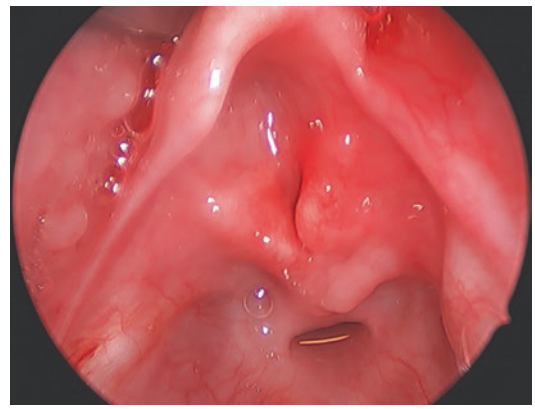

Fig. 32.6 Type IV glottic web

from a respiratory standpoint, there have been reports of improved vocal quality and improvement in quality of life after surgical endoscopic division [8].

Regardless of the operative technique, there is a risk of recurrence if inflammation continues to promote readhesion. Intraoperatively, several pharmacologic adjuncts have been described to decrease early adhesions and inflammation: Kenalog-40 injection, topical mitomycin C, fibrin glue, systemic corticosteroid administration, and coverage of keels and endotracheal tubes with topical Tobradex ointment have been described. Although variable success is met with each adjunctive measure described, it should be noted that many gossamer-thin webs without subglottic extension require only endoscopic lysis (without adjuvant therapy).

Operative Approach

Indications

Surgical intervention is required in patients presenting with respiratory distress or failure. Patients with recurrent respiratory infections or other pulmonary compromise are also counseled toward surgical management. Concern for potential airway compromise, such as known difficult intubation or intermittent symptoms with URIs, is also an indication to consider definitive treatment. For patients without respiratory compromise and no pulmonary sequelae, vocal quality and quality of life influence the decision to intervene. A careful risk-benefit-alternative discussion is essential in elective cases and should include the child's perspective when possible. Children and parents should demonstrate a commitment to improvement by participating in preoperative voice therapy; this will also help lay the foundation for improved postoperative voice outcomes.

Key Aspects of the Consent Process

Risks associated with direct microlaryngoscopy and bronchoscopy should be discussed, including injury to the oral cavity, oropharynx, larynx, and trachea. Patients and parents should be counseled on realistic postsurgical voice outcomes as an element of dysphonia, especially in Type III and Type IV lesions, often persists. Baseline swallow study is also requested prior to surgical intervention. If surgical intervention is elective, such as the desire to improve vocal quality in a Type I or Type II glottic web, the importance of shared decision-making cannot be overstated [8].

Steps

Patients with extensive anterior glottic webs and subglottic extension are often surgically treated in a double-stage fashion. Due to respiratory distress at birth, tracheostomy placement is frequently performed in infancy, with subsequent repair of the laryngeal web occurring months, or on occasion, years later. Under most circumstances, an open approach to the otherwise-unoperated larynx yields the most satisfactory outcomes. Lysis of the web in the midline can be performed first through an endoscopic approach, through the standard open approach, or through a hybrid approach. In the hybrid approach, the incision is created by the operating surgeon at the time of laryngofissure while observing on a monitor, with an assistant providing telescopic visualization transorally. After division of the vocal folds, an anterior laryngofissure allows for elevation of the web with the mucosa of the endolarynx, which can then be retracted through the laryngofissure, and the newly created anterior commissure can be suspended in a more natural location at the anterior thyroid alar cartilage. The laryngofissure is then closed over the anterior commissure, and the cricoid can be expanded using a standard anterior costal cartilage graft. Prior to closure, a suprastomal stent with a keel is used to stent the endolarynx, maintaining a sharp anterior commissure following web repair. Sizing the keel is paramount, and thick keels or keels left in place for too long may cause irritation of the posterior glottis with consequent granulation tissue. The keel and stent are removed after several weeks, with postoperative endoscopic adjunctive treatment (e.g., dilation, removal of

granulation tissue) performed on an as-needed basis [6, 9, 12–14].

Postoperative Management and Follow-Up

After operative management, serial examination in the operating room is recommended for early diagnosis and intervention of reformation. Depending on the operative technique used, this evaluation may be undertaken every 4–6 weeks for the first several months to 1 year postoperatively. A mainstay of proper subsequent interventions is the prevention of opposing raw mucosal surfaces. At times, treatment of one side in isolation followed by contralateral treatment at 2–4 weeks may be required. Changes in breathing or voice, either subjectively or clinically, should prompt expedited evaluation in the operating room.

Postoperatively, control of laryngopharyngeal reflux by both behavioral and pharmacologic (if needed) methods is essential to surgical success. Furthermore, voice therapy is considered an important adjunct to postoperative success by educating patients on techniques that may decrease trauma, enhance healing, and maximize preoperative therapy gains. Because some degree of postoperative dysphonia is anticipated, voice therapy is essential in helping patients achieve the best voice outcome. The timing of postoperative voice therapy depends on the specific operation chosen and whether or not a stent or keel remains in place.

Emerging and Evolving Techniques of the Future

Virtual bronchoscopy and reformatting of multislice imaging (CT or MRI) can demonstrate a static view of glottic webs. However, the degree of dynamic tracheobronchomalacia is less readily appreciated [17]. Furthermore, the true thickness of a web and the relationship to the cricoid as well as the status of the posterior glottis and evaluation of laryngeal cleft can only be determined with operative endoscopic laryngoscopy and bronchoscopy.

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Syndromes and Congenital Anomalies

33

Gregory Rice, Maia N. Braden,
and J. Scott McMurray

Overview

Children may present to voice, swallow, and aerodigestive clinics with known syndrome diagnoses, or in some cases speech-language pathologists and otolaryngologists may be the first medical professionals to note a constellation of symptoms that lead to genetic testing and diagnosis. In either situation, it is important to have a basic understanding of some of the more common syndromes and congenital anomalies associated with laryngeal and airway abnormalities. This is not a comprehensive list, but is designed to give clinicians an overview of some of the syndromes they are most likely to see in clinic.

Common Syndromes and Associations

22q11.2 Deletion Syndrome

22q11.2 deletion syndrome has a variety of names, given to the constellation of traits before the deletion was identified. These include DiGeorge syndrome, velo-cardio-facial syndrome, conotruncal facial syndrome, and a subset of Opitz G/BBB syndrome (Fig. 33.1). However, since the availability of accurate genetic testing, it is typically referred to by the name of the deletion [1]. It is the most common microdeletion syndrome, estimated to occur in 1 in 4000 live births [2]. It is typically caused by a 1.5 Mb submicroscopic deletion resulting in haploinsufficiency of the critical gene TBX1. Patients are usually diagnosed with congenital anomalies at birth; however, diagnosis is delayed into adult in some patients due to extreme

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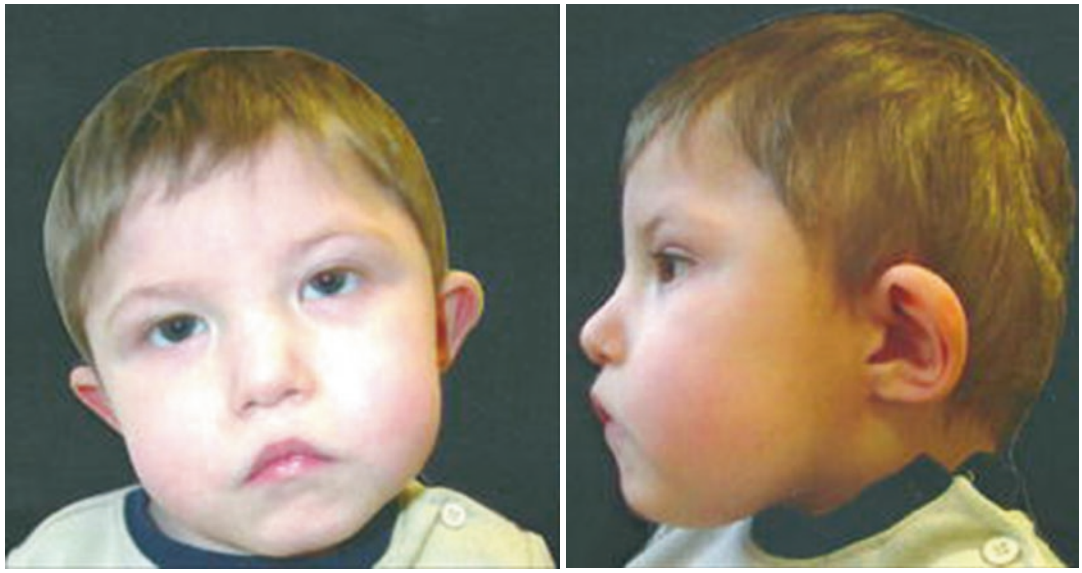

Fig. 33.1 Child with 22q11.2 deletion syndrome. (From Habel et al. [4], with permission)

phenotypical variability. The deletion is inherited in an autosomal dominant fashion in approximately 7% cases but typically occurs de novo due to nonallelic homologous recombination. Affected individuals have a 50% chance of passing the abnormality onto their own offspring.

The condition has extreme phenotypic variability, but the classical clinical features include congenital heart disease (including tetralogy of Fallot) [3], Pierre Robin cleft palate secondary to micrognathia, thymic aplasia with immune deficiency, and hypocalcemia due to hypoparathyroidism. Other cardiac problems include pulmonary atresia with ventricular septal defect, truncus arteriosus, interrupted aortic arch, isolated anomalies of the aortic arch, and ventricular septal defect. Most individuals have some degree of velopharyngeal insufficiency, even in the absence of a cleft palate. Submucous cleft palate is also common. Other features include speech and cognitive delays, hearing loss, short stature, psychiatric disorders, and an increased propensity to autoimmune disorders. Congenital anomalies of the cervical spine and kidneys are also common.

At diagnosis affected individuals should have imaging for occult structural anomalies including echocardiogram and renal ultrasound. Cervical spine x-rays should be performed after age 4. Patients should be referred to immunology at diagnosis and regularly screened for hypothyroidism, hearing loss, hypocalcemia, and thrombocytopenia. Patients should also be watched for cognitive and developmental delays. Guidelines for care of children with 22q11.2 deletion syndrome, including screenings and evaluations at recommended ages, are available [1, 4].

Feeding and Swallowing

Children with 22q11.2 deletion syndrome are more vulnerable to feeding and swallowing problems than the general population [5]. This can result from a variety of causes and is often multifactorial. Children have been found to have difficulty with coordination of suck, swallow, and breathe, slow feeding, and gastrointestinal problems including vomiting, reflux, and constipation [6]. Those children with velopharyngeal dysfunction (with or without cleft palate) can have feeding problems related to the inability to generate enough negative pressure to suck from a breast or standard bottle, leading to prolonged feedings, poor weight gain, and failure to thrive. Hypotonicity can lead to difficulty with the oropharyngeal swallow. Vocal fold paralysis following cardiac surgery can impact airway protection.

Voice, Laryngeal, and Velopharyngeal Abnormalities

Velopharyngeal and laryngeal abnormalities occur frequently in this population. Ebert and colleagues [7] found that 18% of patients studied had a laryngeal abnormality, and those reported include laryngeal web, subglottic stenosis, vocal fold paralysis (often secondary to cardiac surgery), and laryngomalacia (Fig. 33.2) [7, 8]. Voice evaluation revealed that 65% of children had perceptual voice quality within normal limits [7]. Individuals with 22q11.2 deletion syndrome have more severe speech difficulties than most children with cleft palate and even in the absence of an overt or submucous cleft may have velopharyngeal insufficiency. The severity of hypernasality has been attributed to both reduced velar thickness and platybasia [9, 10].

It is extremely important to be aware that children with 22q11.2 deletion syndrome can have a medialized internal carotid, and knowledge of the location of the carotid is needed before any surgery to the head or neck. Additionally, adenoidectomy in children with this syndrome can unmask velopharyngeal dysfunction and should be undertaken carefully.

While detailed discussion of this is outside of the scope of this text, it should be noted that children with 22q11.2 deletion syndrome are at high risk for speech, language, and learning problems. They are also at risk for psychiatric problems [11].

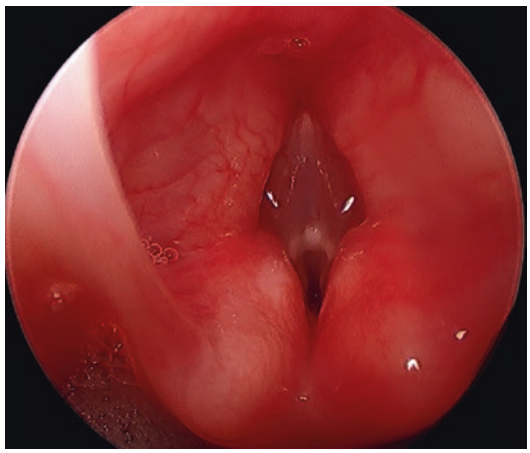

Fig. 33.2 Severe laryngeal web found in a child with 22q11.2 deletion syndrome. This child required tracheotomy shortly after birth and will require laryngeal-tracheal reconstruction prior to decannulation as the web also involves the cricoid

Treacher Collins Syndrome

Treacher Collins syndrome is due to inactivating mutations in the gene TCOF1. The condition often occurs de novo but can be inherited from an affected parent in a dominant fashion. The primary congenital anomaly is hypoplasia of zygomatic arches resulting in malar hypoplasia with downslanting palpebral fissures. Ear malformations, preauricular tags, and lower eyelid coloboma are common (Fig. 33.3) [12]. Patients have normal cognitive abilities, and congenital anomalies outside the head neck region are rare. Miller syndrome is an autosomal recessive disorder due to mutations in the gene DHODH. Miller syndrome is similar to Treacher Collins syndrome in that malar hypoplasia is common; however, patients with Miller syndrome also have anomalies of limbs, including radial and/or ulnar hypoplasia.

Feeding and Swallowing

While feeding and swallowing problems are frequently reported in the Treacher Collins population, these are frequently related to micro-/retrognathia and difficulty breathing, resulting in difficulty coordinating suck/swallow/breathe and potential aspiration, poor feeding efficiency, and poor weight gain. It is not clear that there are any primary oropharyngeal swallow deficits, but rather these result from anatomic differences [13]. Side-lying positioning during feeding and external pacing can be helpful in improving feeding.

VACTERL Association

VACTERL association is a constellation of congenital anomalies that often occur together, the genetic basis is unknown, and the condition is generally not inherited. The clinical features that often occur include vertebral anomalies (V), anal atresia (A), cardiac malformations (C), tracheo-

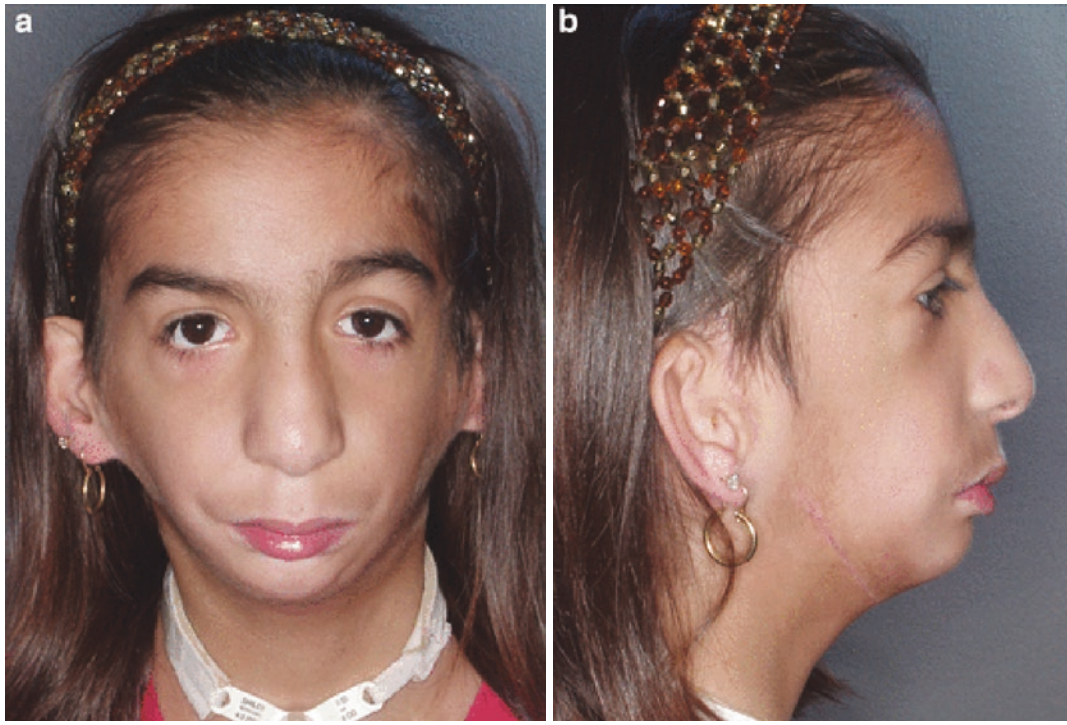

Fig. 33.3 8-year-old child with Treacher Collins syndrome. (From Teichgraeber et al. [12], with permission)

esophageal atresia with fistula (TE), renal anomalies (R), and limb malformations including radial atresia (L). Some patients have speech and/or feeding delays, but cognitive development is usually normal. The condition is a diagnosis of exclusion in that overlapping genetic conditions (such as Fanconi anemia, CHARGE syndrome, and 22q11.2 deletion syndrome) should be ruled out before making a diagnosis of VACTERL. At diagnosis investigations for occult congenital anomalies should occur including radiographs of the spine, echocardiogram, and renal ultrasound. An image of a typical limb malformation with thumb hypoplasia is seen in Fig. 33.4.

Feeding and Swallowing

Feeding and swallowing problems in children with VACTERL association are highly variable and depend on the constellation and severity of clinical features. Tracheoesophageal fistula can lead to airway invasion prior to repair and frequently poor esophageal motility after repair. Cardiac malformations can put children at risk for vocal fold paralysis.

CHARGE Syndrome

CHARGE syndrome is due to point mutations in the gene CDH7. It can be inherited in an autosomal dominant fashion from an affected parent, but must cases occur de novo. The common clinical features include ocular coloboma (C), congenital heart disease (H), choanal atresia (A), growth retardation/renal malformations (R), genital malformations (G), and characteristic ear malformations (E) (Fig. 33.5) [14]. Esophageal atresia, immune deficiency, Mondini malformation, and facial nerve palsy are also common features. Intellectual disability can occur.

Feeding and Swallowing

Feeding and swallowing difficulties are frequently reported in children with CHARGE

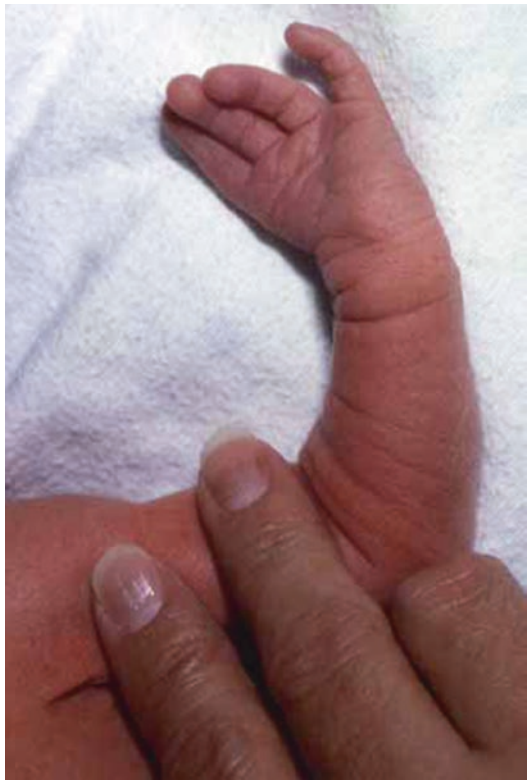

Fig. 33.4 Typical limb malformation with thumb hypoplasia seen in VACTERL association. (Courtesy of National Human Genome Research Institute https://elementsofmorphology.nih.gov/index.cgi?tid=901e03be3fdf0a9e)

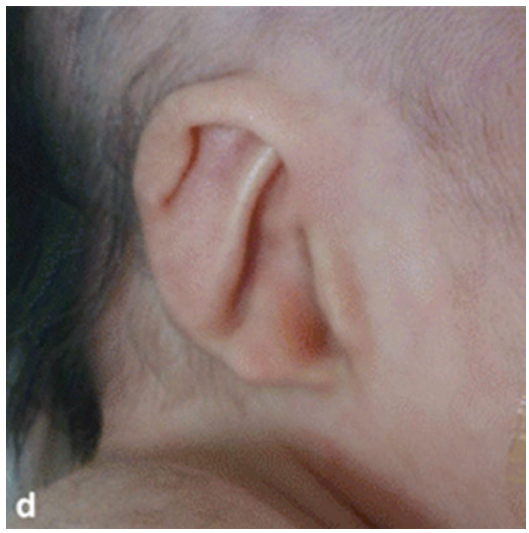

Fig. 33.5 Characteristic appearance of the ear in CHARGE syndrome. (From Chang et al. [14], with permission)

syndrome, with up to 90% having some form of feeding or swallowing problems. While often attributed to the structural abnormalities (tracheoesophageal fistula, cleft palate, choanal atresia), the role of cranial nerve deficits should not be underestimated. Deficits in cranial nerves VII, VIII, IX, and X all occur in children with CHARGE association and can impact feeding and swallowing [15]. Coughing, choking, aspiration, and poor feeding are all reported. Eighty percent reported a history of aspiration, 89% reported a history of reflux, and 92% had some form of feeding tube during their lifetime, with 72% requiring a G-tube [13]. FEES and VFSS exams have shown premature spillage, penetration, aspiration, and pooling [16]. Lack of interest in food due to poor olfaction, packing and stuffing of the oral cavity due to reduced sensation, gastroesophageal reflux, dysmotility, and oral aversion are all reported [17].

Voice and Laryngeal Abnormalities

While the prevalence of voice disorders is not specifically described in the literature, laryngeal and airway abnormalities could contribute to voice problems, and voice should be screened in children with CHARGE syndrome, with evaluation if a voice abnormality is appreciated.

Opitz G/BBB Syndrome

Opitz G/BBB syndrome is an X-linked disorder caused by mutations in the MID1. Common features include hypertelorism, cleft lip and palate, congenital heart and renal anomalies, hypospadias, and imperforate anus. Laryngeal tracheal malformations can occur. Developmental delays can be seen in some affected boys.

Feeding and Swallowing

Cleft palate, heart defects, and laryngeal-tracheal cleft are all common in children with Opitz G/BBB syndrome and can contribute in various ways to feeding and swallowing problems (Figs. 33.6 and 33.7) [18]. As with other children with cleft palate, they cannot generate negative pressure to suck from a breast or stan-

dard bottle and require a cleft specialty bottle. If a laryngeal cleft is present, this can result in aspiration prior to repair, and evaluation of the larynx should be completed in children with signs/symptoms of aspiration, confirmed aspiration on a swallow evaluation, or respiratory problems concerning for aspiration. Depending on the type and severity of cardiac anomalies, children with this syndrome may have challenges with strength and stamina for feeding, and following a cardiac surgery, vocal fold paralysis or paresis should be considered as a possibility if there is a change in voice, breathing, or feeding.

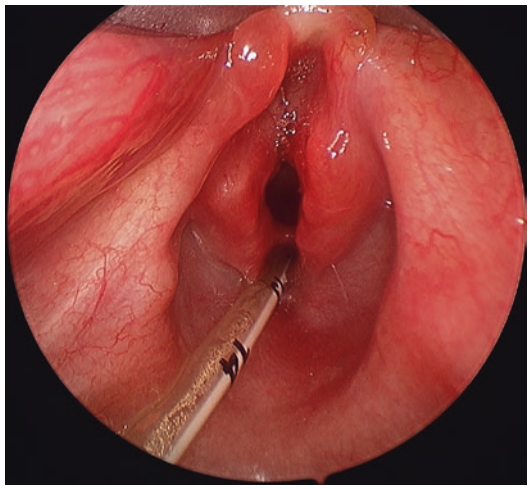

Fig. 33.6 Laryngeal cleft found in a child with Opitz G/BBB syndrome. This child required a tracheotomy and repair of the cleft. The feeding tube is seen entering the cervical esophagus, and the entrance to the trachea can be seen above this. The cleft extends through the cricoid and into the cervical trachea

Fig. 33.7 Two children with Opitz G/BBB syndrome. Notice the wildly spaced eyes (telecanthus). (From Aparicio-Rodriguez et al. [18], with permission)

Kabuki Syndrome

Kabuki syndrome is a multiple congenital anomaly syndrome due to point mutations in one of two genes, KMT2D which is autosomal dominant and KDMA which is X-linked. Common features include characteristic facial features with high arched eyebrows, large ears, long palpebral fissures with eversion of the later aspect of the lower lid, and flat nasal tip (Fig. 33.8) [19].

Other common features include cleft palate, hearing loss, congenital heart and renal disease, short stature, and an increased propensity to autoimmune and infectious diseases. Most patients have mild to moderate developmental delays.

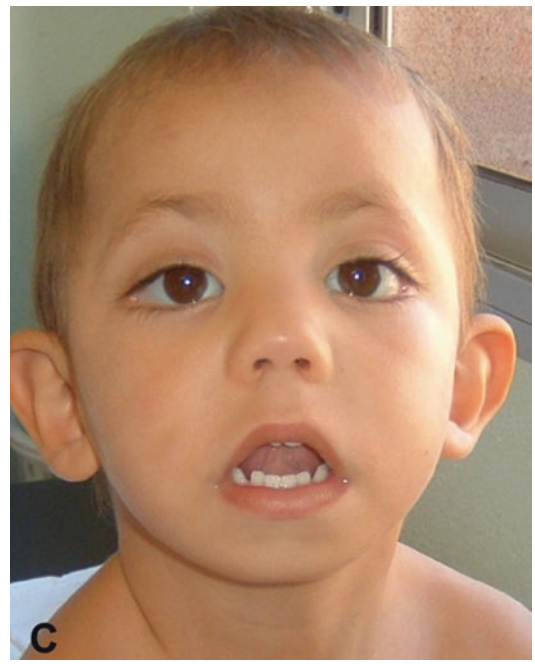

Fig. 33.8 Child with Kabuki syndrome. (From Cusco et al [19], with permission)

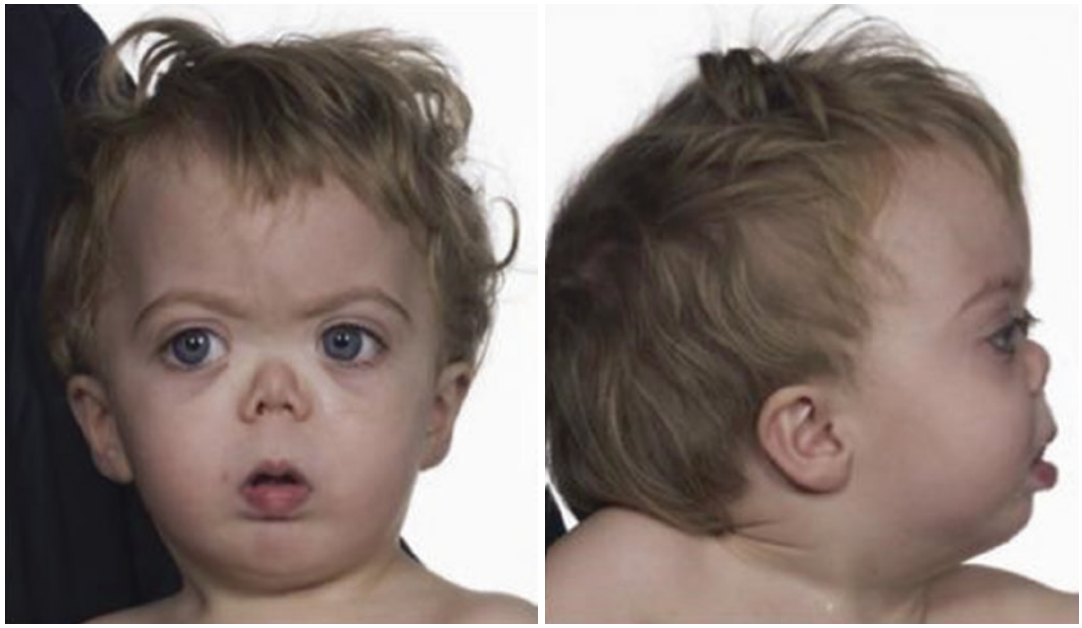

Fig. 33.9 Child with Stickler syndrome. (From Lauritsen et al. [20], with permission)

Feeding and Swallowing

Feeding and swallowing problems are reported in children with Kabuki syndrome. These are most often related to cleft palate or high arched palate and can often be treated with the introduction of a specialty bottle, but feeding and swallowing concerns should be fully evaluated.

Stickler Syndrome

Stickler syndrome is an autosomal dominant disorder due to point mutations in one of four collagen genes (COL2A1, COL9A1, COL9A2, COL11A1). Common features include typical facial features with malar flattening, sensorineural hearing loss, high myopia, cleft palate, and a mild spondyloepiphyseal dysplasia with prematurity osteoarthritis (Fig. 33.9) [20]. Most patients have normal stature and development.

Feeding and Swallowing

Children with Stickler syndrome may have feeding and swallowing difficulties related to cleft palate or micrognathia. The cleft palate-related problems are generally able to be addressed with introduction of a specialty bottle. If micrognathia is severe enough to negatively impact breathing during feeding, surgical intervention may be needed. In less severe cases, side-lying positioning and external pacing may be sufficient.

Summary

Children with these syndromes frequently have more than one feature contributing to voice, swallow, and airway concerns. It is important to know the features of syndromes commonly associated with speech pathology and otolaryngologic issues in order to provide the best evaluation and treatment.

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Esophageal Dysmotility

34

Matthew R. Hoffman, Maia N. Braden, and J. Scott McMurray

Overview

The normal esophageal swallow is a complex process requiring coordinated autonomic innervation, complex contractions of striated and smooth muscle, and appropriately timed relaxation at the lower esophageal sphincter (LES) to deliver a bolus to the stomach [1, 2]. Impairment of any aspect in that process can result in esophageal dysmotility, a group of disorders characterized by abnormal peristalsis of the esophageal body or impaired relaxation at the LES. Esophageal dysmotility can occur both primarily or secondarily as a feature in other disorders, and its effects can be debilitating. Appropriate management of esophageal dysmotility hinges on accurate assessment, best accomplished by a multidisciplinary team with careful history and utilization of both functional imaging as well as high-resolution manometry (HRM). Treatment ranges from conservative approaches such as dietary modification to surgical intervention including Heller myotomy. An understanding of esophageal dysmotility is important for those caring for children with dysphagia.

Normal Esophageal Motor Function

Esophageal functions include transporting a bolus from the pharynx to stomach and preventing reflux of contents from the stomach. The esophagus is comprised of four layers: mucosa, including the stratified squamous epithelium, lamina propria, and muscularis mucosa; submucosa, which includes connective tissue, vasculature, lymphatics, and the submucosal Meissner plexus; muscularis externa, including an inner circular muscle layer, the myenteric Auerbach plexus, and an outer longitudinal muscle layer; and the adventitia, a fascial layer [2]. The

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esophagus can be divided into three functional regions: the upper esophageal sphincter (UES), body, and lower esophageal sphincter (LES). During a normal swallow, the cricopharyngeus of the UES relaxes, allowing bolus transit from the pharynx to the esophagus. Primary peristalsis propels a bolus inferiorly and secondary peristalsis clears any residue [3]. Relaxation at the LES allows for delivery of the bolus to the stomach.

Critical to normal function is the underlying autonomic innervation, which develops during the first trimester of gestation [4]. The esophagus receives both parasympathetic innervation from the nucleus ambiguus and dorsal motor nucleus of the vagus as well as sympathetic innervation from the cervical and thoracic sympathetic chain which regulate secretions, blood flow, and muscle activity [2]. The myenteric Auerbach and submucosal Meissner plexuses also contribute to esophageal muscle activity control. The myenteric plexus includes both excitatory neurons which release acetylcholine, causing smooth muscle contraction, and inhibitory neurons which release nitric oxide, causing smooth muscle relaxation [5].

Contractile segments in the esophagus have been identified in infants at 27 weeks gestation [6]. During a swallow, inhibitory neurons in the caudal dorsal motor nucleus of the vagus cause inhibition throughout the esophagus via release of nitric oxide that lasts longer in the distal than proximal esophagus. Excitatory neurons in the rostral dorsal motor nucleus, via release of acetylcholine, then cause contraction in the proximal followed by the distal esophagus that propels the bolus to the LES [2]. The LES receives both excitatory and inhibitory input from the vagus nerve [7] and is characterized by a state of tonic muscle contraction [8]. After primary peristalsis is initiated, excitatory input ceases, and release of nitric oxide causes LES relaxation, allowing for bolus passage into the stomach [9].

Chicago Classification

The Chicago Classification of esophageal motility disorders uses a hierarchical approach to classify patients with nonobstructive dysphagia or esophageal chest pain according to standardized esophageal high-resolution manometry (HRM) metrics [10]. Diagnostic categories include disorders of esophagogastric junction (EGJ) outflow, major disorders of peristalsis, or minor disorders of peristalsis characterized by impaired bolus transit [10] (Table 34.1).

Implementation of the Chicago Classification requires assessment with HRM. HRM uses a high number of sensors (often 36) spaced 1 cm apart to capture pressure-related events during

Table 34.1 Version 3 of the Chicago Classification of esophageal dysmotility disorders

Disorder	Criteria
Achalasia and esophagogastric junction outflow obstruction	
Type I achalasia (classic)	Elevated median IRP (>15 mmHg)
	100% failed peristalsis
	DCI <100 mmHg
Type II achalasia (with esophageal compression)	Elevated median IRP (>15 mmHg)
	100% failed peristalsis
	Panesophageal pressurization on ≥20% of swallows
Type III achalasia (spastic achalasia)	Elevated median IRP (>15 mmHg)
	No normal peristalsis
	Premature spastic contractions with DCI >450 mmHg∗s∗cm on ≥20% of swallows
Esophagogastric junction outflow obstruction	Elevated median IRP (>15 mmHg)
	Peristalsis present
Major disorders of peristalsis	
Absent contractility	Normal median IRP
	100% failed peristalsis
Distal esophageal spasm	Normal median IRP
	≥20% premature contractions with DCI >450 mmHg∗s∗cm
Hypercontractile (jackhammer) esophagus	DCI >8000 mmHg∗s∗cm on at least two swallows
Minor disorders of peristalsis	
Ineffective esophageal motility (IEM)	≥50% ineffective (failed or weak) swallows
Fragmented peristalsis	≥50% fragmented contractions with DCI >450 mmHg∗s∗cm

Adapted from Kahrilas et al. [10]
IRP integrated relaxation pressure, *DCI* distal contractile integral

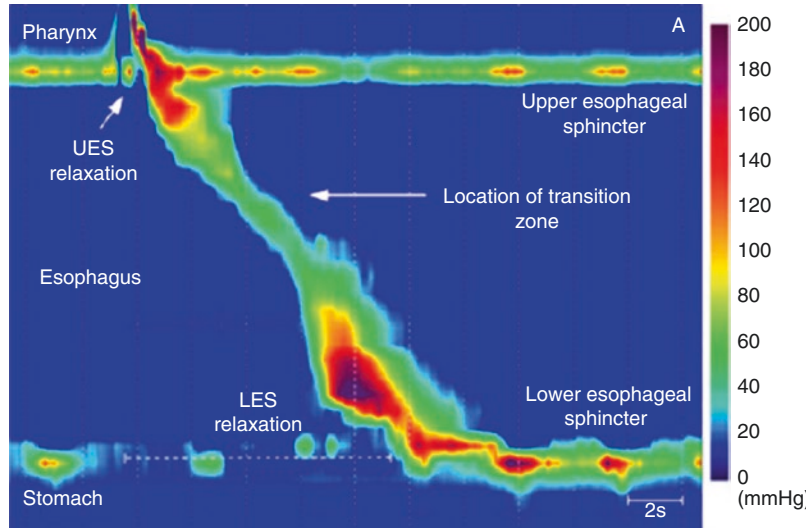

Fig. 34.1 Sample normal high-resolution manometry (HRM) spatiotemporal plot. Time is on the x-axis, sensor position is on the y-axis (with upper esophageal sphincter at top of image and lower esophageal sphincter at bottom), and pressure represented by color. (From Kessing et al. [13], with permission)

swallow with high temporal and spatial fidelity [11, 12]. This is in contrast to traditional manometry which employed three to five widely spaced sensors, which may not capture all salient data regarding bolus propulsion. Data from HRM are presented as a three-dimensional spatiotemporal plot, with time on the x-axis, sensor location on the y-axis, and pressure represented by color (Fig. 34.1) [13]. Various parameters of interest can then be extracted, including local pressure maxima and minima, duration of pressure above or below baseline, and total pressure generated in a region of interest. Importantly, data are available along the length of the entire esophagus, and thus abnormalities can be characterized more completely (Fig. 34.2) [14].

Several HRM metrics are used in the Chicago Classification. These include integrated relaxation pressure (IRP), measured in mmHg, or the mean of the 4 s of maximal deglutitive relaxation in the 10 s window beginning at upper esophageal sphincter relaxation and distal contractile integral (DCI), measured in mmHg∗s∗cm, or the amplitude∗duration∗length of the distal esophageal contraction exceeding 20 mmHg from the transition zone to the proximal margin of the LES [10]. IRP serves to identify EGJ outflow obstruction, with elevated values in the presence of obstruction. DCI is a surrogate for distal esophageal contractile vigor, with hypercontractile disorders having elevated values and hypocontractile disorders having lower values.

There are several limitations relevant to application to children with dysphagia. Most importantly, there are no large normative datasets for esophageal HRM in children [15], and cutoffs for relevant manometric variables described in the Chicago Classification have not been thoroughly studied in the pediatric population [16]. Additionally, esophageal manometry, though low risk, is still an invasive test that requires cooperation by the patient. Effects of catheter size on data collection should also be investigated.

Presentation

Presenting symptoms of esophageal dysmotility in children include dysphagia, pyrosis, chest discomfort, regurgitation, nausea, vomiting, chronic cough, and a change in feeding habits. More severe symptoms include malnutrition, weight loss, and recurrent pneumonia [17]. After starting solid food, children may also present with esophageal food impaction, which is characterized by acute onset of dysphagia, pain, and vomiting [18].

Associated Conditions

Esophageal dysmotility can occur primarily or secondarily in association with other disorders. Disorders commonly featuring esophageal dysmotility include eosinophilic esophagitis (EoE),

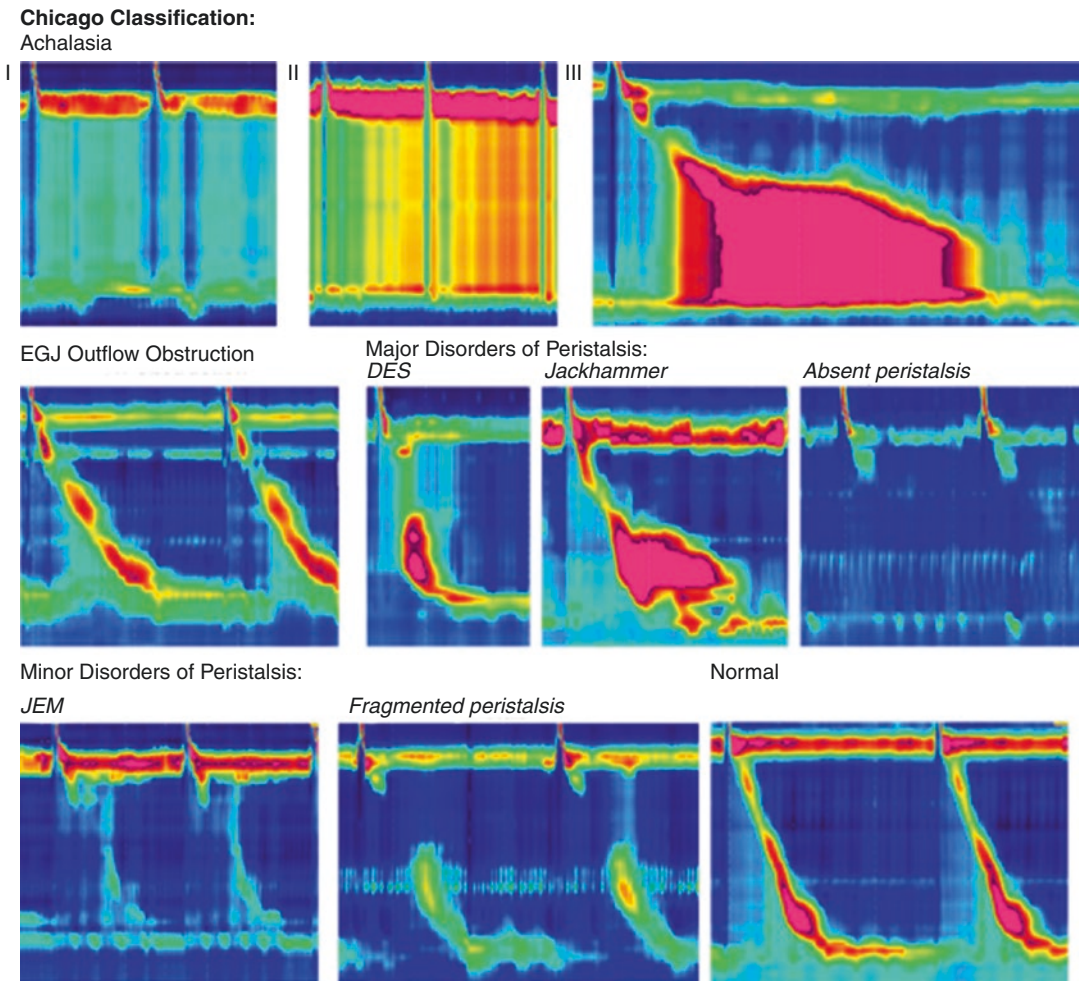

Fig. 34.2 Sample high-resolution manometry (HRM) spatiotemporal plots showing typical appearance of different pathologies within the Chicago Classification. (From Rohof et al. [63], with permission)

esophageal atresia (EA) with or without tracheoesophageal fistula, neurologic impairment, and gastroesophageal reflux disease (GERD). Less common disorders associated with esophageal dysmotility include scleroderma and megacystis-microcolon-intestinal hypoperistalsis syndrome.

Eosinophilic Esophagitis

Eosinophilic esophagitis (EoE) is an immune-mediated antigen-driven disease featuring eosinophilic inflammation of the esophagus [19, 20]. The incidence in children is approximately 10 in 10,000 [19]. Children typically present with pyrosis, regurgitation, emesis, dysphagia, and food impaction [21]. Endoscopic findings include linear furrows, mucosal rings, strictures, and white plaques [22, 23]. Multiple motility abnormalities have been described in patients with eosinophilic esophagitis (EoE), including achalasia, delayed transit (Fig. 34.3), diffuse esophageal spasm, nutcracker esophagus, and tertiary contractions [24]. The pathophysiology of dysmotility in the setting of EoE is not well delineated. Potential mechanisms include eosinophilia causing the release of pro-fibrotic products including TGH-B, IL-13, IL-8, and vascular endothelial growth factor (VEGF) that cause tissue remodeling or eosinophils secreting cytotoxic products such as eosinophil peroxidase which may destroy esophageal intramural neurons [25–27].

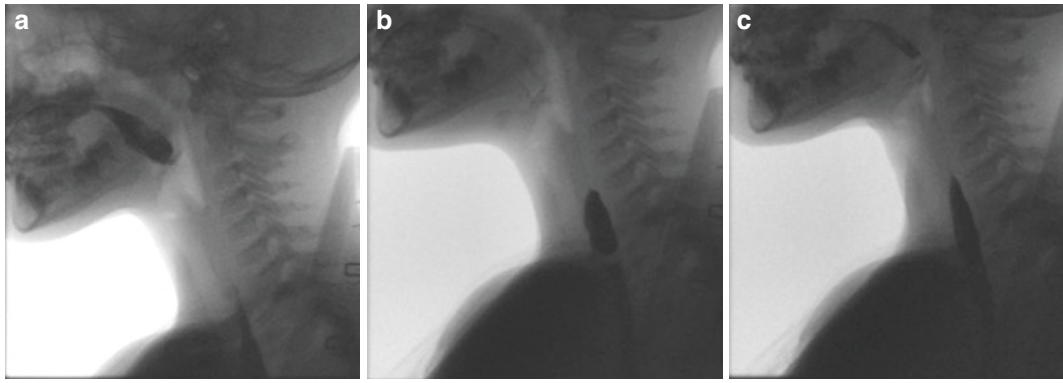

Fig. 34.3 Videofluoroscopy and esophagram showing poor progression of a food bolus. This patient had symptoms of an atypical nonproductive cough and globus. (**a**–**c**) These images showed movement of the food bolus into the proximal esophagus where there was stasis and slow transit. The food bolus stayed in this position for 20 s. Her multidisciplinary endoscopy esophageal biopsies confirmed eosinophilic esophagitis, and her symptoms responded to treatment

Esophageal Atresia

Esophageal atresia with or without tracheoesophageal fistula is the most common congenital esophageal anomaly, with incidence ranging from 1 in 2500–4500 live births [28]. Survival has improved over the last several decades due to improvements in intensive care, anesthesia, nutritional support, respiratory support, and surgical techniques, to the point where mortality is primarily associated with those patients who have additional life-threatening comorbid anomalies [28]. Esophageal dysmotility is an important problem and the most common long-term issue for patients with esophageal atresia [29, 30]. Potential mechanisms underlying dysmotility include developmental neuronal defects, surgical trauma during repair of the atresia, and esophagitis [31]. These changes contribute to abnormalities including aperistalsis, isolated distal contractions, and pan-esophageal pressurization [32]. The dysmotility predisposes to gastroesophageal reflux disease, with consequent exposure of the esophageal mucosa to acid and corresponding inflammatory changes [33]. Thus, long-term follow-up is warranted with treatment targeted at decreasing acid exposure and inflammation as well as monitoring for complications of chronic acid exposure such as Barrett's esophagus [34].

Neurologic Impairment

Children with neurologic impairment often experience feeding problems, in part related to esophageal dysmotility with gastroesophageal reflux disease [35–37]. These children are also more likely to exhibit persistent issues following fundoplication compared to children without comorbid neurologic impairment, potentially secondary to ongoing prolonged LES relaxation or esophageal body spasticity [35, 38, 39].

Gastroesophageal Reflux Disease

There is considerable overlap in symptoms of gastroesophageal reflux disease (GERD) and esophageal dysmotility, with patients with each disorder commonly reporting dysphagia, chest discomfort, regurgitation, and pyrosis. Whether one disorder preceded the other temporally can be debated, but one can certainly perpetuate the other. Patients with esophageal dysmotility may have defective acid clearance, with the persistence of acid within the esophagus causing esophagitis which then further impairs esophageal motor function [31]. For those patients with symptoms of GERD who do not benefit from antacid therapy, additional evaluation with endoscopy and manometry is warranted to evaluate for

alternative or comorbid diagnoses, including dysmotility and eosinophilic esophagitis [40].

Scleroderma and Systemic Sclerosis

Juvenile systemic scleroderma is a rare disorder, occurring in approximately three per one million children [41]. Esophageal dysmotility can occur in these children [42, 43]. Presenting symptoms may include dysphagia, regurgitation, and pyrosis [44], and patients may exhibit low-amplitude peristaltic contractions, tertiary contractions, and low LES resting pressure, with poor esophageal bolus clearance [45]. Use of steroids to treat the underlying disorder can help improve esophageal symptoms [45].

Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome (MMIHS)

MMIHS, or Berdon's syndrome, is a rare smooth muscle myopathy resulting in an enlarged bladder, microcolon, and small intestine hypoperistalsis [46]. It was first described in 1976 [47], and less than 300 cases have been reported [48]. Both autosomal dominant inheritance and de novo mutations have been described [49]. A recent series of six patients with the disease identified normal LES resting tone and relaxation but absent esophageal peristalsis in all patients [46]. Though this is a rare disorder, esophageal dysmotility is reasonable to consider in any child presenting with dysphagia in the setting of an underlying myopathy.

Speech-Language Pathologist Approach

Esophageal motility problems are extremely challenging when working with pediatric dysphagia. While it can be tempting to remain focused on the oropharyngeal swallow, events below the upper esophageal sphincter are also an essential component of effective bolus passage. Children with esophageal dysmotility are at increased risk for aspiration, food refusal, and poor weight gain. The American Speech-Language-Hearing Association (ASHA) states that speech-language pathologists should have knowledge and skills regarding the interrelationships of the oral, pharyngeal, and esophageal stages of swallowing, and "If esophageal screening is completed, describe any suspected anatomic and/or physiologic abnormalities of the esophagus which might impact the pharyngeal swallow, deferring to radiology for diagnostic statements" [50]. Thus, while we do not diagnose esophageal disorders, we are responsible for knowledge of typical and atypical esophageal structure and function and making appropriate referrals and recommendations for further evaluation.

Additionally, as we are treating increasingly complex infants and children, we are more likely to encounter esophageal dysmotility as sequelae of tracheoesophageal fistula, prematurity, neurologic conditions, and inflammatory conditions. Familiarity with the presentation and treatment of esophageal dysmotility is increasingly important. Speech-language pathologists should be aware of symptoms associated with dysmotility, as well as signs on clinical and instrumental evaluation, and know when to recommend further evaluation.

Presentation

Symptoms of esophageal dysmotility in infants may include spitting up, vomiting, slow feeding, food refusal, fussiness, poor weight gain, and even failure to thrive. In toddlers and older children, it may manifest in vomiting, food refusal, slow eating, and regurgitation. Verbal children may report food sticking or pain or discomfort in their chest [17, 51]. Coughing after eating or when lying down can be a symptom as well. When these symptoms are reported, an esophageal issue should be considered. These patients may be referred for a videofluoroscopic swallow study. It should be noted that even when an esophagram is a part of the evaluation, the esophagram has poor sensitivity in identifying esophageal motility disorders, in comparison with manometry [52].

Videofluoroscopy

The videofluoroscopic swallow study is intended to evaluate the oropharyngeal swallow and the cervical esophagus, and as such is not the ideal test for evaluating esophageal dysmotility. Ideally, a barium esophagram will have been planned as part of the evaluation based on history and presentation, but in some cases, it is not, or the child is unwilling or unable to take adequate volumes to complete the esophagram. When esophageal dysfunction is suspected, an esophageal screening should be done.

Signs on videofluoroscopic swallow study can include the following: stasis at the UES or proximal esophagus, retrograde motion of the bolus, and even aspiration of contrast that did not pass through the esophagus. On esophageal screening or barium esophagram, tertiary esophageal contractions, limited or inconsistent passage of the bolus through the esophagus, retrograde movement, stacking of the food in the esophagus, a "nutcracker" appearance of the esophagus, or a "bird beak" appearance of the lower esophageal sphincter may be seen [53, 54].

Treatment

While speech-language pathologists cannot directly treat esophageal dysmotility, we can educate patients and parents on the disorder and offer compensatory strategies. Compensatory strategies that may help with optimizing safe, efficient, and comfortable oral intake include the following:

- Positional changes: upright feeding; remain upright after meals
- Texture changes: thinning or mechanically altering solids to allow for easier passage and decreased stasis
- Behavioral changes: recommending a liquid wash for solids if safe; recommending smaller, more frequent meals
- Nonnutritive suck: especially when oral feeding is not a viable option, promoting nonnutritive sucking, maintaining interest in oral stimulation, and pre-feeding skills

When esophageal dysmotility is suspected, it is the role of the SLP to bring this concern to other team members for further medical evaluation and treatment. Reevaluation with therapy as appropriate is indicated after medical or surgical intervention.

Otolaryngologist Approach

As stated above, there is considerable overlap in the symptoms of GERD and esophageal dysmotility, with both potentially featuring dysphagia, chest discomfort, regurgitation, and pyrosis. Some patients may be referred to otolaryngology for dysphagia, with a presumptive diagnosis of GERD that has not responded adequately to antacid therapy. The question of whether or not surgical therapy targeted at improving reflux, as with a Nissen fundoplication, may then be posed as a next step in management. If the underlying disorder is unrecognized esophageal dysmotility rather than undermanaged reflux, though, a Nissen would only worsen patient symptoms. In scenarios where a disorder has not responded as anticipated to treatment, it is important to consider whether the initial diagnosis was accurate and complete. Esophageal dysmotility will ultimately primarily be managed by the pediatric gastroenterologist and general surgeon, but the otolaryngologist can play a role in organizing initial instrumented assessment, directing patients to those specialists, and evaluating for other treatable comorbid conditions.

History

A thorough history should be obtained, with an emphasis on swallowing function. Presence of regurgitation, vomiting, prolonged mealtimes, food refusal, and retrosternal pain should be assessed. Growth curves should be checked to evaluate for poor weight gain or failure to thrive. Comorbid voice and respiratory issues should be queried. Given the overlap in symptoms as well as the association with eosinophilic esophagitis, a history of food allergies, asthma, atopic dermatitis, and food impactions should be sought. Past

medical history should also be reviewed for esophageal atresia, neurologic disease, myopathy, connective tissue disease, and any aerodigestive surgical interventions. Prior treatments including trials of antacid therapy or diet modification should be noted.

Instrumented Assessment

If esophageal dysmotility is considered, esophagram and esophageal HRM with impedance should be obtained. This may be done in concert with the pediatric gastroenterologist, depending on the institutional practice. An esophagram may demonstrate the classic "bird's beak" appearance in the setting of achalasia, though a normal study does not rule out early disease, and manometry is still warranted for complete evaluation [55]. Manometry is a commonly performed procedure in pediatrics and provides helpful, quantitative information on the pressures underlying bolus transit. A recent consensus statement from the American Neurogastroenterology and Motility Society (ANMS) and North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) reviewed indications for performing esophageal manometry in children [56]. Notably, the indications are wide and include diagnosis of primary and secondary esophageal motor disorders, assessment of dysphagia, choking with feeding, vomiting, chest pain, and regurgitation, evaluation of obstruction after fundoplication, assessment of esophageal stasis in the setting of pneumonia or aspiration, and evaluation of esophageal function prior to any surgical intervention [56]. This is particularly relevant if one is considering proceeding with a fundoplication for symptoms of persistent reflux, as manometry would help evaluate for underlying dysmotility which may be worsened by fundoplication.

Management

If there is clinical concern for dysmotility or finding of dysmotility on manometry, evaluation by pediatric gastroenterology is warranted. Depending on the clinical scenario, esophagogastroduodenoscopy may be warranted, such as to evaluate for eosinophilic esophagitis, stricture, or changes related to chronic acid exposure, or to rule out other source of obstruction. In the setting of achalasia, referral to pediatric general surgery is also warranted. There is significant heterogeneity in management of pediatric achalasia across institutions, with common interventions including laparoscopic Heller myotomy, pneumatic dilation, serial botulinum toxin injection, and peroral endoscopic myotomy (POEM) [55, 57]. Evaluation by both gastroenterology and surgery is helpful for determining the most appropriate option for each patient.

Emerging and Evolving Techniques

Disorder Classification

Though the Chicago Classification provides a logical framework to approaching the complex problem of esophageal dysmotility, it was developed in adults. Further, it employs quantitative analyses that have not been validated in children [56]. Singendonk et al. demonstrated that patient's age and esophageal length had an effect on the measures of integrated relaxation pressure and distal contractile integrity included in the Chicago Classification algorithm [58]. As esophageal HRM is applied more routinely to children and larger datasets are created, a pediatric version could be developed.

Innovative Therapies for Dysmotility in Esophageal Atresia

Esophageal dysmotility is a lifelong problem for patients with history of esophageal atresia, in part related to dysfunction of the esophageal enteric nervous system. Recently, decreased expression of GDNF, a neurotrophic factor for the neural crest cells which give rise to the enteric nervous system, has been demonstrated in patients with

esophageal atresia [59, 60]. Therapeutic manipulation of the normal foregut developmental signaling pathways may provide an avenue for treatment of dysmotility. Additionally, enteric neural stem cells (ENSCs) have been identified in both human fetal and postnatal tissue [61, 62]. Theoretically, ENSCs could be harvested from one region of the gastrointestinal tract and reimplanted into the esophagus to restore neural input in patients with ongoing neuropathic dysfunction leading to dysmotility [59].

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Cricopharyngeal Achalasia

35

Tony Kille and Laurie Matzdorf

Introduction

The four phases of swallowing (i.e., oral preparatory, oral transit, pharyngeal, and esophageal) involve chewing a bolus into an appropriate size and consistency, moving the organized food bolus into the pharynx where it is moved sequentially by the tongue base and pharyngeal constrictors past the relaxed upper esophageal sphincter into the esophagus, all while protecting the airway from aspiration. Swallowing is an intricate function that relies on precise coordination of these voluntary and involuntary processes. Cricopharyngeal achalasia is a disorder wherein the upper esophageal sphincter – which is tonically contracted in its baseline state – fails to relax appropriately, preventing normal transit of the bolus from the pharynx to the esophagus [1]. While this condition is not infrequently encountered in adults, it is an uncommon cause of dysphagia in the pediatric population which was only first described in children in the late 1960s [2].

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Anatomy and Physiology

The upper esophageal sphincter (UES) is a functionally musculocartilaginous structure. Anteriorly, it is composed of the posterior aspect of the cricoid cartilage. Laterally and posteriorly, it is comprised primarily of the cricopharyngeus muscle, but with contributions from the proximal esophageal muscle fibers, and the inferior pharyngeal constrictor. The cricopharyngeus muscle inserts onto the dorsolateral aspect of the cricoid cartilage on each side and forms a raphe posteriorly. Working together, these structures generate a high-pressure zone that can be identified on videofluoroscopy and manometry [3].

The cricopharyngeus is innervated by the recurrent laryngeal nerve and pharyngeal plexus bilaterally [4]. The lower motor neuron cell bodies for these nerves reside in the ipsilateral nucleus ambiguus, with significant dendritic arborization into the adjacent reticular formation. These cell bodies have both excitatory and inhibitory synaptic contacts, suggesting a basis for the variety of afferent inputs (predominantly carried to the brain by the glossopharyngeal nerve) that influence reflex control of the UES [5].

The muscle is predominantly made up of slow-twitch (type 1 oxidative) fibers but also contains fast-twitch (type 2 glycolytic) fibers. This combination of slow- and fast-twitch fibers allows the cricopharyngeus to maintain a basally contracted state, yet relax briskly when necessary during

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swallowing, eructation, and emesis [5, 6]. The UES is opened by a combination of cricopharyngeal muscle relaxation as well as by anterior and superior excursion of the larynx due to the pull of the suprahyoid musculature. Further, the muscle of the UES has a high degree of elasticity, allowing the sphincter to be passively stretched by the laryngeal excursion and the passive pressure of the food bolus itself [5].

Pathophysiology

The exact etiology of cricopharyngeal achalasia in the pediatric population remains elusive and may likely be multifactorial. Neurologic conditions – such as cerebral palsy, Arnold-Chiari malformation, and syringobulbia – are often associated with cricopharyngeal achalasia. Muscular abnormalities (e.g., dermatomyositis and muscular dystrophy) as well as neuromuscular conditions (e.g., myasthenia gravis) have also been correlated to cricopharyngeal achalasia [7]. Neuromuscular immaturity is a likely contributing factor in pediatric cricopharyngeal achalasia. In general, the UES is discernible at 32 weeks gestational age, and it should be functioning in a coordinated fashion in full-term newborns [8]. Thus, prematurity is also a risk factor for cricopharyngeal dysfunction and achalasia.

Cricopharyngeal achalasia can result from failure of relaxation, incomplete relaxation, or abnormal timing of relaxation of the UES. Specifically, high-resolution esophageal manometry performed in pediatric patients with cricopharyngeal achalasia reveals elevated basal UES pressures, elevated pressures during swallowing, and premature contraction of the UES which decreases the time available for bolus transit across the sphincter into the esophagus [8–11].

Presentation

Children with cricopharyngeal achalasia present with dysphagia characterized by choking episodes, prolonged feeding times, pooling of secretions, excess salivation, and nasal regurgitation. These feeding difficulties can often lead to recurrent respiratory infections related to aspiration as well as failure to thrive [12]. The differential diagnosis for pediatric patients with these symptoms would include severe gastroesophageal reflux, an abnormal structural communication between the airway and esophagus (such as laryngeal cleft or tracheoesophageal fistula), palatal abnormalities (such as submucous cleft palate), esophageal webs or stenosis, esophageal dysmotility, and extrinsic compression on the esophagus (e.g., from a vascular ring) [13]. As compared to other causes of pediatric dysphagia, cricopharyngeal achalasia is quite uncommon. Thus, a high index of suspicion for this entity is needed in order to avoid a delay in diagnosis. Given that the etiology of pediatric cricopharyngeal achalasia is often idiopathic, the natural history of this disorder is difficult to predict. Indeed, spontaneous resolution has been reported, particularly in infants [14].

Speech-Language Pathologist Approach

Assessment of children with feeding difficulties begins with obtaining a thorough history, including the child's general medical history as well as a focal feeding/swallowing history. Important aspects of the general medical history should include gestational age and birth history (including any perinatal complications), weight gain and progress on the growth chart, noisy or effortful breathing, recurrent or chronic respiratory infections (especially if hospitalization was necessary), as well as appropriate advancement through the developmental milestones – particularly motor and speech milestones.

Diagnosis of cricopharyngeal achalasia is usually made by identification of a posterior cricopharyngeal "bar" seen on standard videofluoroscopic swallow evaluation (modified barium swallow study) (Fig. 35.1). The cricopharyngeal bar suggests failure of the cricopharyngeus muscle to relax during swallowing. Additional abnormalities seen on videofluoroscopic swallow

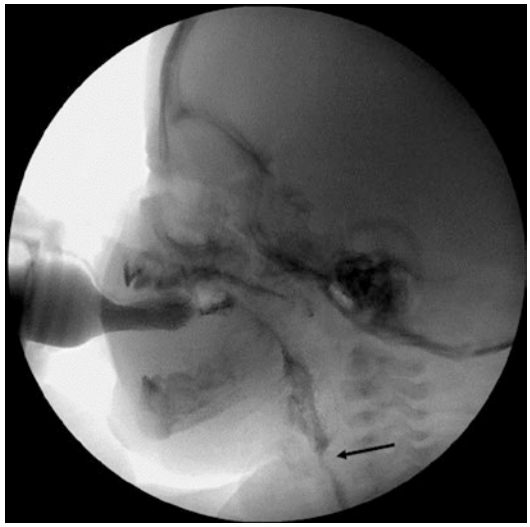

Fig. 35.1 Still image from videofluoroscopic exam showing impression due to cricopharyngeal bar (black arrow) which narrows the pharyngeal outflow

studies in patients with cricopharyngeal achalasia might include residue and pooling in the hypopharynx, penetration and/or aspiration, and occasionally nasal regurgitation.

As stated earlier, high-resolution manometry can be used to evaluate cricopharyngeal resting pressure and relaxation during the swallow. When the cricopharyngeus fails to adequately relax on swallow evaluation, clinicians should be aware of different etiologies that can contribute to this, and understanding the etiology is vital to planning treatment and making appropriate referrals. As described earlier, it may be neurologic or inflammatory. However, opening of the UES is also dependent on hyolaryngeal elevation and pharyngeal propulsion, and care should be taken not to ignore any oropharyngeal swallow abnormalities that could result in failure of the cricopharyngeus to relax.

While direct treatment of the cricopharyngeus may require medical or surgical intervention, speech-language pathologists can often provide compensatory strategies to aid in feeding, including texture modifications, bolus presentation, and positioning. If there is an oropharyngeal component such as reduced tongue base propulsion or reduced pharyngeal constriction, swallow therapy may play a role. Evaluation and treatment after medical/surgical management should be done to assess changes in swallow function and provide therapy or modifications as needed.

Otolaryngologist Approach

As with all patients presenting with dysphagia, a thorough otolaryngologic examination is critical. This would include assessment of general appearance for any syndromic features, nasal patency, and all oral structures. Breathing should be examined/auscultated before, during, and after feeding. Flexible fiber-optic nasopharyngoscopy should also be included to assess pharyngeal and laryngeal anatomy. Though the physical exam of patients with cricopharyngeal achalasia is generally normal, there may be clues present including pooling of secretions in the hypopharynx.

While modified barium swallow study is considered the "gold standard" for diagnosis of cricopharyngeal achalasia, there are a few other studies that may be beneficial in the overall work-up of these patients. These include (1) upper GI endoscopy, (2) pH probe studies, and (3) esophageal manometry.

Upper endoscopy is likely to reveal a tight upper esophageal sphincter but otherwise no other findings specific to cricopharyngeal achalasia. Endoscopy is also useful to rule out any obstructive lesions, as well as to evaluate for reflux and/or eosinophilic esophagitis – factors that could potentiate UES hyperactivity and hypertonicity [9]. Similarly, pH probe studies can help assess for reflux.

Esophageal manometry can be utilized to better define the swallow dysfunction as it relates to the UES. Manometry would be expected to demonstrate high baseline UES pressures with limited relaxation of the UES during swallowing. Manometry – particularly high-resolution manometry – can also assess for abnormal timing and dyscoordination of pharyngeal contraction in relation to UES relaxation, which would result in premature closure of the UES and decreased time available for bolus transit.

Finally, if cricopharyngeal achalasia is diagnosed, further evaluation for the etiology of this issue is recommended. Specifically, a thorough neurologic evaluation should be undertaken, with strong consideration of head MRI to assess for posterior fossa abnormalities such as Chiari malformation.

Options for management of cricopharyngeal achalasia range include simple observation, dilation, Botox injection, and surgical (open or endoscopic) myotomy. In all cases, treatment to prevent reflux should be included, as exposure of the UES to refluxate is thought to aggravate muscle spasm and hyperactivity.

Nitrates and calcium channel blockers – such as nifedipine – have shown promise in the treatment of lower esophageal sphincter disorders; however, these do not have clear and consistent effects at the UES and, therefore, are not considered viable treatment options for cricopharyngeal achalasia.

Though difficult to predict, there are reports of spontaneous resolution of cricopharyngeal achalasia, particularly in neonates and infants. Thus, treatment options that are temporary (such as dilation or Botox injection) may be reasonable in the hopes of being able to avoid the risks involved with myotomy.

In some cases, observation alone – with provision of an alternative feeding modality such as a gastrostomy tube – might be considered appropriate for treatment of cricopharyngeal achalasia when there are significant additional neurologic and functional deficits that would otherwise preclude development of normal feeding and swallowing. In cases where the child is otherwise normally developing, however, observation without specific treatment is not ideal. Cricopharyngeal achalasia typically results in severe swallow impairment that is not likely to improve with a change in feeding technique or food consistency. And while the natural history of this condition is unpredictable (and spontaneous resolution has been described), placement of a nasogastric or gastric feeding tube to merely temporize during a period of observation has the additional disadvantage of inhibiting development of normal feeding behaviors.

Operative Approaches

Indications

Indications for procedural intervention include ongoing dysphagia (characterized by choking episodes, prolonged feeding times, pooling of secretions, excess salivation, and nasal regurgitation) with UES dysfunction identified as a significant contributing factor. There are a variety of procedural options available for treatment. The decision about which intervention to employ depends on several factors, including experience of the operative team, availability of appropriate instrumentation, the patient's gestational age, and the duration of anesthesia the patient can tolerate. For premature infants, where there may be some chance of spontaneous resolution of cricopharyngeal achalasia, a temporary or less invasive treatment modality (such as Botox injection or dilatation) may be warranted.

The success of transcervical myotomy is generally better than Botox injection and comparable to dilation and endoscopic cricopharyngeal myotomy. Endoscopic myotomy, however, has the advantages of shorter operative time, shorter length of hospital stay, and lower rate of complications [15]. The choice of operative technique for cricopharyngeal myotomy is dependent upon surgeon experience and availability of appropriate instrumentation. Patients with difficult endoscopic exposure (e.g., limited neck mobility or trismus) may be better candidates for the transcervical approach.

Key Aspects of the Consent Process

If direct laryngoscopy is performed, associated risks including injury to the lips, gums, tongue, or teeth as well as dysgeusia should be discussed. If botulinum toxin is injected, there is potential for diffusion of toxin to adjacent structures with potential for worsening of dysphagia or airway obstruction related to bilateral vocal paresis. Potential complications of transcervical cricopharyngeal myotomy include hematoma, wound infection, recurrent laryngeal nerve injury,

pharyngeal or esophageal perforation, salivary leak, pharyngocutaneous fistula formation, and mediastinitis. Potential complications of endoscopic cricopharyngeal myotomy include mediastinitis, bleeding, supraglottic edema, and dental injury related to laryngoscopy.

Dilatation

Cricopharyngeal dilation stretches the UES muscle fibers, providing easier transit of food into the esophagus. Experience with this technique is better described in the adult literature, though there are reports of success in pediatric patients [10, 16].

Steps

1. *Patient positioning*. General anesthesia is induced, and an orotracheal tube is placed, taped off to the left corner of the mouth. Place patient supine with neck extended. The maxillary alveolus is protected with a tooth guard.
2. *Exposure.* A pediatric Parson laryngoscope is placed in a post-cricoid position to elevate the larynx and expose the UES. Any laryngoscope of adequate length to reach and elevate the post-cricoid mucosa can be used.
3. *Dilation.* Bougie dilator or a dilating balloon is positioned within the UES to affect the cricopharyngeus muscle. Application of constant low pressure is performed for 15–60 s.
4. *Additional points.* Though general anesthesia is needed, dilation is generally low risk with only mild mucosal tears described in various case series [17]. The duration of effect is variable, with repeated dilatations often needed to maintain symptomatic improvement [16–18].

Botox Injection

Botulinum toxin (Botox) is a polypeptide that inhibits presynaptic release of acetylcholine at the neuromuscular junction, resulting in flaccid paralysis. Focal injection of Botox into the upper esophageal sphincter for the treatment of cricopharyngeal achalasia was first described in adults in 1994 [19] and in pediatric patients in 2005 [20]. Botox can be injected into the cricopharyngeal muscle either endoscopically or percutaneously, with or without EMG guidance for each. Endoscopic injection has the advantage of relatively easy identification of the cricopharyngeus as well as accurate localization of the injection needle – both under direct visualization, thus making EMG guidance superfluous.

Steps

1. *Patient positioning.* General anesthesia is induced, and an orotracheal tube is placed and taped off to the left. Patient is positioned supine with neck extended.
2. *Exposure.* A pediatric Parsons laryngoscope is placed in a post-cricoid position, and the larynx is elevated to visualize the cricopharyngeus.
3. *Injection.* Botox is injected into the posterior and lateral aspects of the UES using a laryngeal injector needle (Fig. 35.2) [21]. Injection into the anterior portion of the UES complex should be avoided to prevent diffusion into the posterior cricoarytenoid muscles – the sole abductors of the vocal folds – that could result in bilateral vocal paralysis and possible airway obstruction.
4. *Dosage.* The dosage of Botox is debatable, but the amounts used safely and successfully in various reports range from ~1.5 to 7 units/kg [12, 22]. Higher doses of Botox may have a

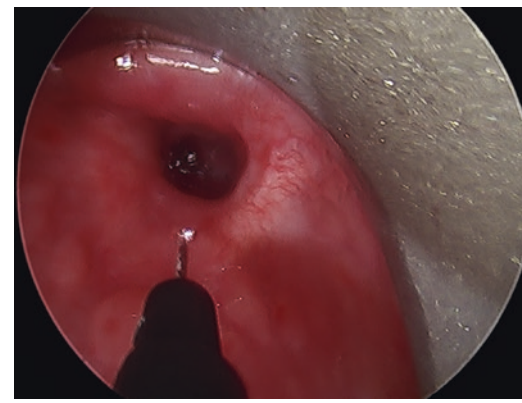

Fig. 35.2 Endoscopic injection needle is aimed toward the posterolateral aspect of the cricopharyngeus muscle

faster onset (approximately 2.5 days versus 5 days), but the duration of action (~3–4 months) is not dose-dependent [21, 23] and is determined by other metabolic and biochemical factors.

5. *Additional points.* Since the effect of Botox is not permanent, repeat injections are generally needed to maintain effect. Some clinicians utilize Botox as a diagnostic tool, with consideration of a more permanent treatment option (myotomy) if Botox proves helpful [12].

Transcervical Cricopharyngeal Myotomy

Transcervical cricopharyngeal myotomy was first described in the 1950s and continues to be considered a highly effective treatment of cricopharyngeal achalasia. Experience with this technique is more extensive in the adult literature as it pertains to treatment of Zenker's diverticula, but has traditionally been considered the standard of care in the treatment of pediatric cricopharyngeal achalasia as well.

Steps

1. *Positioning*. General anesthesia is induced and an orotracheal tube is placed. A shoulder roll is also placed. Patient is positioned supine with neck extended and rotated.
2. *Incision.* A transverse skin incision is made at the level of the cricoid cartilage. Incision is carried down through platysma.
3. *Identify sternocleidomastoid (SCM).* The SCM and carotid sheath are identified and retracted laterally to expose the larynx, trachea, and UES.
4. *Identify cricopharyngeus.* The laryngotracheal complex is rotated to allow view of the thickened transverse cricopharyngeal muscle fibers attaching to the cricoid cartilage at the esophageal entrance.
5. *Divide cricopharyngeus fibers.* Cricopharyngeus muscle fibers are divided longitudinally at the midline posteriorly, incorporating the entire length of the functional upper esophageal sphincter. The myotomy is taken down to where submucosal tissues are seen bulging out. Dissection should remain extramucosal. Care is taken to avoid injury to the recurrent laryngeal nerve in the tracheoesophageal groove [7, 24, 25].
6. *Place a Penrose or rubber band drain.* A drain is placed. The wound is closed in layers. Patient is observed for one night postoperatively, and the drain is removed the following day.

Endoscopic Cricopharyngeal Myotomy

Cricopharyngeal myotomy can be performed endoscopically as well. This has been described more recently (1990s) and – given improvements in endoscopic equipment and instrumentation as well as improved surgeon technical experience and expertise – may be supplanting the transcervical myotomy as the technique of choice for management of cricopharyngeal achalasia.

Steps

1. *Positioning*. General anesthesia is induced and an orotracheal tube is placed, taped off to the left. Patient is positioned supine with neck extended. The maxillary alveolus is protected with a tooth guard.
2. *Exposure*. A Parson laryngoscope is placed with the distal end in the post-cricoid area to expose the upper esophageal sphincter. The laryngoscope is then suspended, lifting the larynx forward and placing the UES under tension. This allows the UES to be easily palpated and for the muscle fibers to more easily splay apart after endoscopic division.
3. *Divide cricopharyngeal muscle fibers*. Carbon dioxide laser is used at a setting of 7 W continuous to transect the cricopharyngeus in the posterior midline down to the underlying buccopharyngeal fascia, a contiguous gray-white sheet of connective tissue that appears distinctly different than the overlying muscle (Fig. 35.3). Violation of the buccopharyngeal fascia should be avoided, as it would allow communication with the

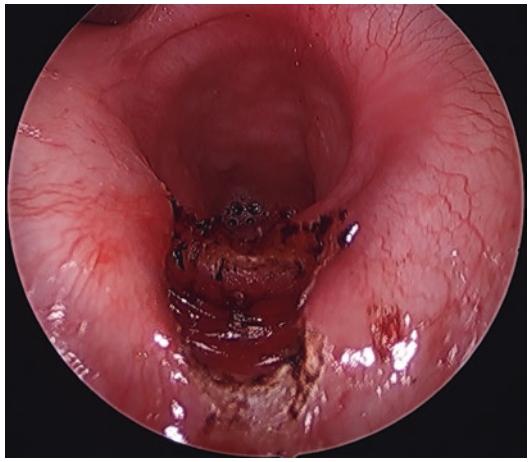

Fig. 35.3 Endoscopic view of cricopharyngeal bar after division with the CO2 laser. Exposure is obtained with a Parson laryngoscope. Fibers are divided down the middle with preservation of the underlying buccopharyngeal fascia

underlying retropharyngeal space which extends down into the mediastinum.

4. *Additional points*. The CO_2 laser is ideal for endoscopic cricopharyngeal myotomy, as it is precise and can provide sufficient hemostasis without significant unintended spread of thermal damage [9, 26].

Postoperative Care

Postoperative management after cricopharyngeal myotomy (either transcervical or endoscopic) – in terms of swallow studies, initiation and advancement of oral feeding, and length of stay – is varied among surgeons, and various protocols have not been adequately compared to determine the best practice. Perioperative antibiotics are generally recommended, as is an initial swallow study to determine safety of starting oral feeds. The pace of oral feeding advancement depends upon postoperative recovery as well as the status of preoperative feeding skills. In those patients with long-standing dysphagia – particularly infants – acquisition of appropriate feeding skills and coordination may take time, even after resolution of obstruction at the level of the UES. Swallow improvements related to cricopharyngeal myotomy should be durable over time, though recurrence of achalasia could theoretically occur if scar forms in the myotomy site. Thus, close monitoring by surgeons and speech pathologists is recommended over time.

Emerging Concepts

Swallow is a pressure-driven process. In the setting of cricopharyngeal achalasia, pharyngeal outflow is obstructed, and intrabolus pressures increase, thus increasing residue and risk of aspiration [27]. High-resolution manometry (HRM) uses a large number of sensors spaced 1 cm apart to capture pressure-related events along the length of the pharynx with high temporal and spatial fidelity [28]. It was recently applied to children with signs of oropharyngeal dysfunction and demonstrated increased pharyngeal intrabolus pressure in those patients [27]. HRM offers a quantitative, objective method of assessing pharyngeal function and may help inform assessment of cricopharyngeal dysfunction and decisions on appropriate therapeutic management.

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36 Congenital Tracheal Anomalies: Complete Tracheal Rings, Tracheomalacia, and Vascular Compression

Lyndy J. Wilcox, Claire Miller, and Michael J. Rutter

Overview

The normal trachea consists of approximately 15–20 cartilaginous rings situated in a "horseshoe" or "C-shape" as well as a posterior membranous portion in a 4–5:1 ratio (Fig. 36.1) [1]. Alterations in this ratio are seen in two of the most common congenital tracheal anomalies, tracheal stenosis and tracheomalacia. Congenital disorders of the trachea, while rare, can cause a wide range of symptoms with varying severity. The diversity in presentation mandates a high degree of clinical suspicion to identify these disorders. Management has evolved significantly, such that there is potential to address both morbidity and mortality in this patient population. Concomitant congenital anomalies may be present and can complicate management. A multidisciplinary approach to care is necessary to ensure the best airway, feeding, and overall outcomes for these complex patients. This chapter will focus on the clinical presentation, diagnosis, and management of congenital tracheal stenosis, most commonly caused by complete tracheal rings. Additionally, other causes of tracheal stenosis and disorders involving tracheal collapse, including both intrinsic and extrinsic causes, will be reviewed.

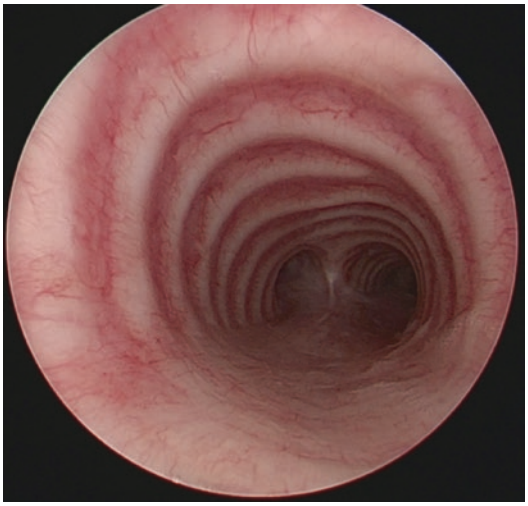

Fig. 36.1 Normal trachea with a 4–5:1 cartilage/membranous trachea ratio

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Definitions

Congenital Tracheal Stenosis

Complete tracheal rings are the most common cause of congenital tracheal stenosis. In this anomaly, there is an "O-shaped" trachea (or a portion of it) with absence of the usual posterior membranous aspect (Fig. 36.2a, b) [2]. Complete tracheal rings can take on several patterns including (1) a "stovepipe" airway or "generalized hypoplasia" with a long segment of complete rings of similar diameter; (2) a "funnel-shaped" trachea with narrowing distally; (3) short-segment stenosis; and (4) complete rings associated with a tracheal or pig bronchus (or other anomalous branching pattern) [3]. A *sleeve trachea* is a tracheal anomaly in which the trachea consists of a sheetlike cartilage formation. The sleeve trachea may extend from the cricoid proximally into the bronchi distally. This does not uniformly result in stenosis; however, the posterior cartilaginous trachea can overlap creating a "figure-9" trachea (Fig. 36.3) that is stenotic and requires intervention [4]. *Tracheal webs* are a short-segment, circumferential form of tracheal stenosis that typically spare the underlying cartilaginous framework [2]. *Tracheal agenesis* is a rare embryologic anomaly resulting in a partial or complete absence of the tracheal airway. Floyd and colleagues described three anatomic variants, all with an absent proximal trachea and a distal airway remnant (i.e., distal trachea, carina, or bronchi) with or without esophageal fistula [5].

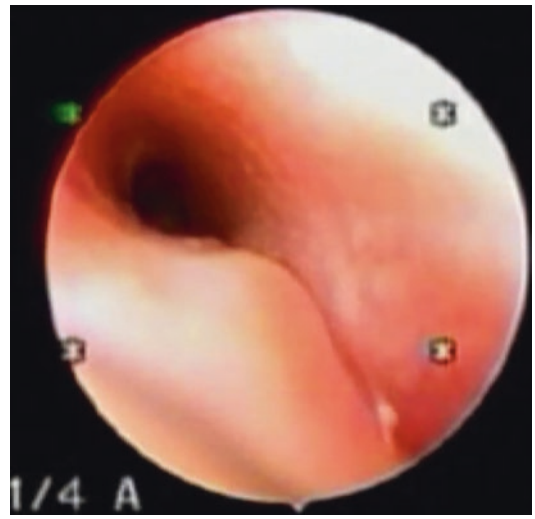

Fig. 36.3 "Figure-9 trachea" seen in tracheal sleeves with overlap of the posterior cartilaginous portions

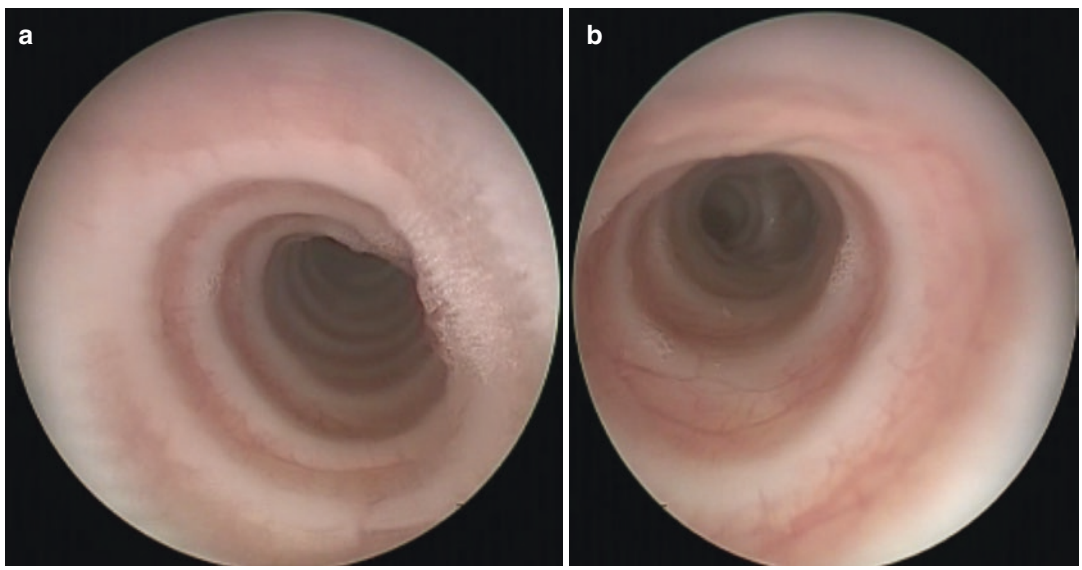

Fig. 36.2 Proximal (**a**) and distal (**b**) views of complete tracheal rings with an "O-shaped" appearance of the cartilage

Tracheomalacia

Tracheomalacia is defined as abnormal softness or collapsibility of the tracheal airway due to inadequate support (Fig. 36.4a). This may be a *primary* structural issue (i.e., *intrinsic*) due to abnormal cartilage framework or atrophy of the longitudinal elastic fibers of the pars membranacea [6]. In the most severe intrinsic form, absent tracheal rings, there can be complete collapse of a short segment (2–3 rings) of the airway that acts more like a true stenosis (Fig. 36.4b) [7]. Tracheomalacia may also be a prominent finding after repair of tracheoesophageal fistulas (Fig. 36.4c) or laryngotracheal clefts. Alternatively, tracheomalacia can occur in a *secondary* or *acquired* fashion. This can be an intrinsic structural issue secondary to prolonged positive pressure ventilation or prior tracheostomy, or it may be due to external compression (i.e., *extrinsic*) from vascular rings, esophageal anomalies, skeletal anomalies, or neck/mediastinal masses. Intrinsic tracheomalacia can also be seen secondary to other conditions. For example, prolonged extrinsic compression can result in degeneration and weakening of the cartilage that can persist even after the resolution of the inciting compression.

A comprehensive discussion of each type of vascular compression is beyond the scope of this chapter; however, several articles and book chapters available provide excellent summaries of this information [8–13]. Briefly, a *double aortic arch*, the most common vascular ring, is seen when the ascending aorta bifurcates to surround the trachea and esophagus and then rejoins to form the descending aorta. A *left-sided aortic arch with aberrant right subclavian artery* and *right-sided aortic arch with aberrant left subclavian artery* occur when there is abnormal involution of the right or left fourth arches, respectively, such that the aberrant subclavian artery takes a retroesophageal course to perfuse the respective side. In a *pulmonary artery sling*, the left pulmonary artery arises from the right pulmonary artery, travels over the right bronchus, and then passes through the tracheoesophageal groove. Finally, an *aberrant innominate artery* is seen when the innominate artery takes off from a more distal, leftward position along the aortic arch.

Figure 36.5 provides a framework to classify and approach congenital tracheal anomalies. Note that although tracheal stenosis and tracheomalacia or vascular compression may coexist, the distinction is important due to the different pathophysiology and management approaches.

Epidemiology

Congenital Tracheal Stenosis

Compared with subglottic stenosis, tracheal stenosis is relatively rare in the pediatric population and is more commonly congenital in origin as opposed to acquired [4]. Congenital tracheal stenosis has been estimated to occur in only 1 in

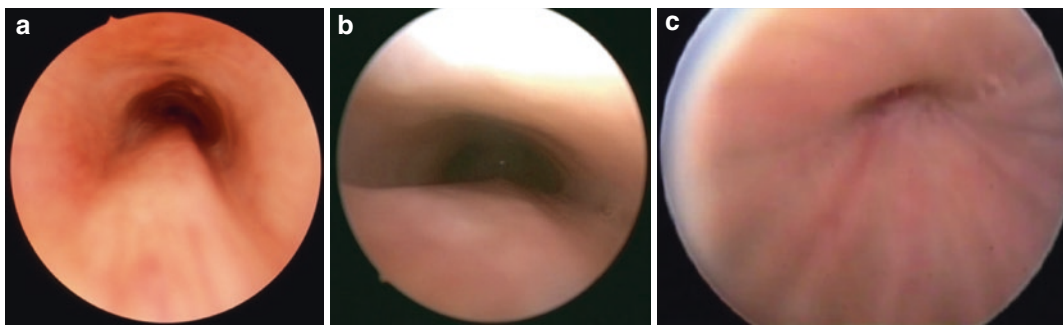

Fig. 36.4 (**a**) Intrinsic posterior tracheomalacia with a "bowed" appearance to the cartilage and a cartilage/membranous trachea ratio of <4–5:1. (**b**) Severe tracheomalacia seen with absent cartilage rings causing a functional tracheal stenosis. (**c**) Tracheomalacia in the setting of a tracheoesophageal fistula

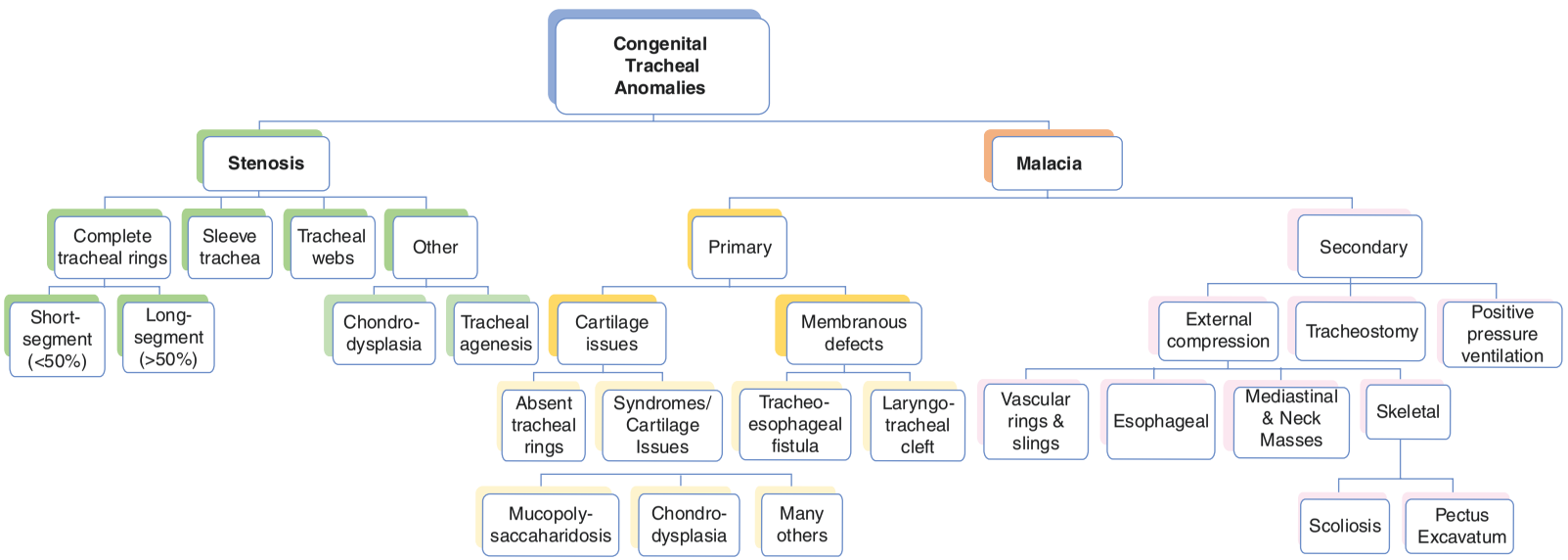

Fig. 36.5 Framework to classify and discuss tracheal anomalies, including stenosis and malacia

64,500 births [14] and accounts for only 0.1–0.3% of all laryngotracheal stenosis cases [2]. The most common form of congenital tracheal stenosis is complete tracheal rings; however, these are still extremely rare and account for <1% of all airway stenoses [2]. Complete tracheal rings are associated with other congenital malformations in 60–75% of cases [15–17]. These include cardiovascular anomalies (most commonly a pulmonary artery sling), tracheal bronchus, lung hypoplasia or agenesis, Down syndrome, and variations of the VATER/VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, limb abnormalities) spectrum.

Other causes of congenital tracheal stenosis, including laryngotracheal webs, tracheal sleeves, and trachea agenesis, are even more rare. Tracheal sleeves are universally associated with craniosynostosis syndromes, such as Pfeiffer, Crouzon, or Apert [18]. Tracheal agenesis has a prevalence of less than 1:50,000 births and occurs twice as commonly in males. This is typically associated with premature birth (52%), polyhydramnios, and other congenital malformations (90%) [19]. The incidence of tracheal webs has been reported to be 1/10,000 births [20]. Reports in the literature are sparse, however, and some authors question whether more frequent webs occur and are incidentally treated via intubation [4]. Tracheal agenesis has been reported to occur in 1 in 50,000 to 1 in 100,000 live births [21]. This anomaly is almost universally fatal; however, the presence of a tracheoesophageal fistula or distal trachea may allow for temporary ventilation via esophageal intubation or tracheostomy, respectively, and potential future reconstruction [5, 22, 23].

Tracheomalacia

Primary tracheomalacia is the most common congenital anomaly of the pediatric trachea occurring in approximately 1/2100 children [24]. Congenital tracheomalacia has been associated with other airway anomalies, including laryngomalacia and bronchomalacia [25–27]. Primary tracheomalacia is also seen in the setting of mucopolysaccharidoses [28], connective tissue disorders, and chromosomal abnormalities and may present with more diffuse tracheal involvement [15, 29, 30]. As mentioned previously, congenital tracheomalacia also occurs in patients with tracheoesophageal fistula [31] or large (type 3 or 4) laryngotracheal clefts [25–27]. The prevalence of severe tracheomalacia in patients with esophageal atresia has been reported to range from 11% to 33% [29]. Absent tracheal rings are an extreme form of primary tracheomalacia and are exceedingly rare. Children with this anomaly are typically otherwise normal. Associations with left vocal fold paralysis and esophageal atresia have been reported [4].

Secondary tracheomalacia is not in itself uncommon; however, it is asymptomatic or minimally symptomatic in the majority of cases. As depicted in Fig. 36.5, tracheomalacia can occur secondary to a variety of conditions. Children treated with prolonged mechanical ventilation are one of the most commonly affected patient populations [24, 32]. The prevalence of tracheomalacia in infants with bronchopulmonary dysplasia has been estimated to be at least 16% [33]. However, the true incidence is difficult to determine, as only symptomatic children are typically assessed endoscopically [34]. Patients who have undergone tracheostomy are another population commonly found to have secondary tracheomalacia. Malacia has been reported to occur in at least 10% of patients after tracheostomy due to trauma to the tracheal rings with resultant increased compliance of the suprastomal airway, stoma, site of the tracheostomy cuff, and/or distal end of the tracheostomy tube [15, 34].

Despite vascular rings accounting for <1% of all congenital cardiac defects, they are the most common congenital anomaly resulting in secondary airway compression [10, 35, 36]. The double aortic arch is the most common form of symptomatic vascular ring in the pediatric population, accounting for ~50–60% of vascular rings [8, 9, 11, 12]. Other vascular rings and slings causing airway compression are the left-sided aortic arch with aberrant right subclavian artery, right-sided aortic arch with an aberrant left subclavian artery, pulmonary artery sling, anomalous innominate

artery, and cervical aortic arch [8–11]. Innominate artery compression occurs in up to 30% of children [37], but is only symptomatic enough to undergo surgery in 13.7% [38]. Of note, at least 50% of children with a pulmonary artery sling will also have long-segment congenital tracheal stenosis in the form of complete tracheal rings [39]. Additionally, vascular airway compression has been showed to occur in approximately 1–2% of children with congenital heart disease, even in the absence of vascular rings [10].

Pathophysiology

The embryologic development of the aerodigestive tract, particularly in relation to the surrounding vascular structures, is a complex process. An understanding of these mechanisms and the ways they can go wrong, however, can help explain many of the tracheal anomalies encountered by clinicians. In general, developmental lesions occurring by week 4 of gestation result in more severe manifestations of disease (i.e., tracheal agenesis), while those occurring after week 8 may have less drastic effects (i.e., complete tracheal rings) [40].

The laryngotracheal groove appears in the proximal foregut in the third week of development. The trachea and esophagus develop as a single tube from the ventral and dorsal anterior foregut endoderm, respectively, at the fourth week, while the lung buds expand caudally. Complete separation of the trachea and esophagus occurs by the sixth week of gestation as proliferating ridges within the lumen of the foregut unite to form the tracheoesophageal septum [41]. Notably, there is some debate on how this process occurs, but the "septation" model is currently the most widely accepted [42, 43]. The tracheal cartilage, connective tissue, and smooth muscle arise from mesenchyme from the fourth and sixth pharyngeal arches and surround the tracheal tube in weeks 8–10, forming the tracheal rings, the tracheal walls, and the trachealis [41]. The laryngotracheal lumen is occluded by an overproliferation of endoderm that eventually recanalizes by week 10 to create the normal glottic opening. The branching pattern of the lower airway is complete by 16 weeks. With initial development complete, the second half of gestation is characterized by airway maturation and remodeling [44]. During this time, the cartilage strengthens, demonstrating a fivefold decrease in airway compliance [45].

This basic framework provides a primitive schema for how many tracheal anomalies occur. For example, tracheal agenesis results from an early defect in development at the fourth week. Tracheoesophageal fistula form when there is failure of complete separation of the foregut into the tracheal and esophageal lumens. Laryngotracheal clefts arise when there is incomplete fusion of the tracheoesophageal septum. Complete tracheal rings result from a defect in embryogenesis after the eighth week of gestation causing a complete cartilaginous ring with absence of the usual posterior membranous portion of the trachea. Primary tracheomalacia can, in a general sense, be explained by a failure of appropriate development of the tracheal cartilage. While findings may vary based on the etiology of the malacia, the trachea generally appears more "U-shaped" or even "bowed" as compared with normal (Fig. 36.4a). There is also an alteration of the typical 4–5:1 cartilage/membranous trachea ratio. Secondary tracheomalacia, as previously discussed, can result from external compression, as a result of tracheostomy, or following positive pressure ventilation.

While development of individual cardiovascular anomalies with resultant airway compression is beyond the scope of this chapter, the normal embryologic development of the vasculature provides a basis for understanding these complex anomalies. The five paired pharyngeal arches (numbered 1, 2, 3, 4, and 6) each have an associated primitive aortic arch that connect the paired dorsal and ventral aorta. The fifth pharyngeal arch is rudimentary in humans and regresses quickly without any contribution to the ultimate arterial system [1, 46]. Appropriate involution, regression, and persistence of the remaining five primitive arches are required to ultimately have normal anatomy. Typically, the first and second arches regress on leaving the maxillary and stapedial

arteries, respectively. The third and fourth arches appear as the first and second regress. The third arch persists as the common carotid and proximal internal carotid arteries, while the fourth arch develops into the proximal subclavian artery on the right side and aortic arch on the left side. Finally, the sixth arch forms and results in the bilateral pulmonary arteries from the ventral portion and the left-sided ductus arteriosus from the left dorsal portion. The right dorsal aorta involutes while the left dorsal aorta becomes the distal aortic arch and descending thoracic aorta. The dorsal intersegmental arteries bilaterally become portions of the subclavian arteries. A double aortic arch is created when both fourth arches persist [12, 46, 47].

Presentation

Clinically, patients with tracheal stenosis and tracheomalacia may have overlapping symptoms. It is important, however, to distinguish between the two, as the management is very different [4, 15].

Congenital Tracheal Stenosis

Children with complete tracheal rings most commonly present in infancy with worsening respiratory function characterized by biphasic "washing-machine" stridor, retractions, apnea, cyanosis, and "dying spells." Symptoms are typically worse when the child is agitated or feeding. Failure to thrive may also be present. Decompensation is commonly seen around 4 months of age as the child "outgrows" the airway [48]. Respiratory infections may also exacerbate symptoms causing significant respiratory distress requiring intubation [39]. If the tracheal stenosis is undiagnosed, intubation may be described as very difficult, requiring the endotracheal tube to be "screwed in." A traumatic intubation may further escalate the already tenuous airway into a critical airway, sometimes requiring extracorporeal membrane oxygenation (ECMO) [48]. The severity of the presentation will largely depend on the degree of airway stenosis and the comorbid conditions. While it is the exception, there is a subset of patients that present either incidentally (e.g., when being intubated for another reason) or with mild symptoms (e.g., dyspnea on exertion). Some may even present as young adults with asthma-like symptoms [49]. While tracheal sleeves are not typically stenotic, the posterior tracheal cartilage can sometimes overlap causing airway narrowing. In these cases, symptoms would be similar to other forms of congenital tracheal stenosis; however, tracheal sleeves are universally seen in children with craniosynostosis syndromes [18].

Tracheal agenesis will present with severe respiratory distress at birth and no audible cry, despite obvious effort. Attempts at intubation will be unsuccessful; however, mask ventilation or inadvertent esophageal intubation may temporarily improve symptoms in the setting of a tracheo- or bronchoesophageal fistula. Prenatally, a congenital high airway obstruction syndrome (CHAOS) may develop if there is no tracheoesophageal fistula [50].

Tracheomalacia

The majority of children with tracheomalacia are asymptomatic. When symptomatic, however, these children may not present until weeks to months after birth with insidious onset and worsening of symptoms. There may be significant variation in the severity of presenting symptoms based on both the degree of malacia and if there are associated cardiovascular anomalies. Because of these factors, a high index of suspicion may be required to identify patients with tracheomalacia. Common symptoms include shortness of breath, a "brassy" cough, dyspnea on exertion, noisy breathing (expiratory stridor), and, when more severe, apneic and cyanotic spells (sometimes termed "dying spells"). Poor airway clearance is common; thus, these children are more prone to recurrent respiratory infections [34, 51]. In the intubated patient, tracheomalacia can be a cause of failure to extubate or apparent life-threatening events (ALTEs) despite having a secure airway in place. Additionally, several conditions are often comorbid with tracheomalacia, including cardiovascular abnormalities (20–58% of patients) [52, 53], bronchopulmonary dysplasia (up to 52% of patients) [27, 33, 34], gastroesophageal reflux

(50–75% of patients) [15, 27, 54], developmental delay (26%) [52], and/or neurologic impairment [6, 27, 52]. Concomitant airway anomalies, including laryngomalacia, bronchomalacia, vocal cord paralysis, laryngotracheal clefts, tracheoesophageal fistula, and subglottic stenosis, may also be present [6, 24–27].

Similarly, children with vascular compression are often asymptomatic or mildly symptomatic. Symptoms, when present, are typically related to airway compression – including noisy breathing (biphasic stridor) and a "seal-bark" cough. If the compression is more severe, apnea, respiratory distress, cyanosis, and ALTEs or "dying spells" may also be present. Dysphagia is not typically a prominent symptom until the child is old enough to take solid foods. Concomitant cardiovascular anomalies may be present in up to 12% of patients [55]. In general, children with a double aortic arch or pulmonary artery sling tend to have more severe symptoms and present in infancy to the first months of life. Approximately 50% of children with pulmonary artery slings will also have complete tracheal rings contributing to a more severe presentation [56, 57].

Speech-Language Pathologist Approach

Children with congenital or acquired conditions that compress the esophagus and/or trachea are at increased risk for feeding and swallowing issues, depending on the severity and location of the underlying condition [58]. Tracheomalacia, in conjunction with extrinsic defects or anomalies such as vascular rings or congenital or intrinsic tracheal abnormalities, may be associated with respiratory compromise during oral feeding [59]. The extra-respiratory effort expended during oral feeding may exacerbate breathing problems and compromise airway protection during the swallow. Dysphagia associated with an aberrant right subclavian artery (dysphagia lusorum) is characterized by increased intraesophageal pressure and a functional partial obstruction with swallowing [60]. Unrepaired complete tracheal rings result in the potential for respiratory distress, which can be exacerbated by the increased respiratory effort required during feeding, threatening airway integrity with swallowing. The role of the speech-language pathologist in the evaluation of associated dysphagia includes a careful review of the medical history and underlying condition, a clinical dysphagia evaluation, and often an instrumental assessment of swallowing function to rule out swallowing dysfunction and/or airway compromise associated with swallowing. Management of feeding and swallowing issues can be determined following the clinical and instrumental assessments.

History

There is a wide range of patient presentations, from those who have severe respiratory distress and require urgent intervention immediately after birth to those children who present with stridor, dyspnea, cough, wheezing, dysphagia, and recurrent respiratory tract infections during early childhood [10, 15, 61]. Review of the medical history and presenting symptoms is completed prior to the clinical oral motor/feeding assessment. The review includes the following components: prenatal and birth history, confirmed or suspected medical diagnoses, respiratory history, current feeding status (enteral, oral), and social history, including parent/caretaker perception of the feeding difficulty and any barriers to accessing dysphagia treatment. The SLP should be knowledgeable about the underlying condition and the medical plan (including potential or past surgical/medical interventions), confirm physiologic stability prior to the feeding assessment, and collaborate with the medical team during the dysphagia assessment and management process.

Clinical Evaluation

The clinical dysphagia assessment serves as an opportunity to directly assess oral structures and function, to confirm physiologic stability during feeding, and to document clinical signs and symptoms of possible swallowing dysfunction.

Nonnutritive assessment of sensorimotor function, ability to integrate sensory input, and the strength and range of oral motor movements is completed prior to the nutritive assessment. Direct observation of a feeding by a familiar feeder when possible is recommended for assessment of a typical feeding.

During the clinical assessment, careful monitoring for clinical signs and symptoms of swallowing dysfunction is essential. Coughing, choking or gagging, noisy wet respirations, and physiologic signs of respiratory compromise such as bradycardia, apnea, increased respiratory rate, or decreased oxygen saturation levels may signal airway protection issues associated with oral feeding. The clinical presentations of symptoms that may be correlated with the type of condition are documented during the clinical assessment [62]. For example, airway symptoms such as stridor, wheezing, and cough that worsen with the respiratory effort of feeding may be associated with an underlying tracheal compression. Reduction of ventilation may lead to declining oxygenation and to periods of apnea and bradycardia during oral feeding attempts. Respiratory and heart rate changes during feeding outside of the normal baseline should be noted and communicated immediately to the medical team.

Compression of the esophagus may be manifested by discomfort during feeding attempts, overt coughing, choking, refusal or vomiting of solid textures, and a preference for intake of liquids, possibly secondary to partial esophageal occlusion. The signs and symptoms of feeding difficulty vary according to the severity of the condition and the age of the child. For example, problems with esophageal clearance of solids secondary to esophageal compression may only become apparent when the child matures to the point that solids are introduced.

A significant and known limitation of the clinical oral motor/feeding assessment is the ability to accurately identify pharyngeal swallowing dysfunction and/or airway protection problems associated with feeding and oral intake [63]. Threats to airway protection such as aspiration may be silent in nature and not detectable during the clinical examination [64]. Instrumental assessments of swallowing are therefore beneficial in providing an objective analysis of swallowing function and informing the goals for dysphagia management.

Instrumental Assessment of Swallowing Function

The examinations that are used most often for objective assessment of swallowing function are the videofluoroscopic swallowing study (VFSS) and fiber-optic endoscopic evaluation of swallowing (FEES). The VFSS provides a comprehensive, dynamic assessment of the oral, oropharyngeal, hypopharyngeal, and cervical esophageal phases of the swallow and helps to identify the type and location of swallowing impairment. For example, diagnosis of an aberrant subclavian artery and the degree of external compression can be made by the VFSS. Compensatory therapeutic strategies such as the use of liquids to clear any persistent residual in the esophagus can be introduced during the examination to directly visualize the effect.

Feeding supersedes the normal ventilator chemoreceptor control mechanism in young infants, and a repetitive swallow pattern without pause intervals may be identified during the VFSS [65]. Feeding under fluoroscopy provides an opportunity to introduce pacing intervals to alter feeding rhythm, increase ventilation time, and determine the effect on maintenance of airway protection. Additional strategies may include slowing the rate of liquid flow to decrease the frequency of swallowing-related apnea, thereby increasing the potential for physiologic stability during feeding.

Fiber-optic endoscopic evaluation of swallowing (FEES) is advantageous for defining airway protection integrity and safety of swallowing in infants or children who have never fed orally or who have suspected secretion management issues. Laryngeal structures and function can be clearly viewed. The integrity of laryngopharyngeal sensation can be assessed, which provides important predictive information about the child's ability to achieve and sustain airway protection during swallowing [66].

The endoscopic view provides an opportunity to assess the child's ability to achieve glottic closure and to maintain airway protection during oral feeding. Difficulties with respiratory compromise during feeding and consequential penetration or aspiration can be readily detected during the FEES examination prior to swallow onset. Airway compromise secondary to inadequate respiratory pauses and ventilatory needs can be visualized with the abrupt opening of the airway during feeding. In such circumstances, responses to compensatory strategies to improve the coordination of respiration and swallowing can be determined. As with VFSS, imposed respiratory pauses or pacing intervals to facilitate appropriate respiratory pauses and adequate ventilation can be introduced. The effect of pacing can be assessed by close inspection of the glottic and subglottic areas during the respiratory pause cycles to detect any evidence of aspirated material. It should be noted that the FEES examination is limited to visualization of events before and after the swallow. There is a loss of view during the swallow secondary to the upward excursion of the larynx, contractile force of the hypopharyngeal musculature, and subsequent light deflection from the scope. In addition, the view is frequently obscured during rapid, sequential swallowing sequences, such as during bottle-feeding.

Treatment

Once the interpretation of the instrumental examination is completed, recommendations for dysphagia treatment are formulated, if appropriate. Each set of recommendations is dependent on the individual patient and the particular set of medical circumstances. As above, the patient's response to compensatory swallowing strategies during the assessment and/or instrumental examination guides recommendations for dysphagia treatment.

Direct dysphagia treatment approaches refer to rehabilitative maneuvers or specific exercises to change the physiology of the swallow and are usually most appropriate for adults and older children who can follow directions. Indirect dysphagia treatment strategies refer to compensatory techniques to eliminate symptoms of dysphagia and improve the safety and efficiency of the feeding/swallowing process. The majority dysphagia treatment strategies in children with a history of tracheal and/or esophageal compression are compensatory and indirect in nature. Modifying position to facilitate increased respiratory support during feeding (side-lying positioning), modifying flow rate (nipple flow rate, altering liquid viscosity) to facilitate organization of airway protection during swallowing, and altering liquid and solid boluses during oral intake to facilitate esophageal clearance are the mainstays of dysphagia treatment. Feeding and swallowing issues may persist even after surgical intervention of the underlying condition; continued follow-up by the SLP to implement compensatory strategies to assist with oral feeding safety and efficiency may be necessary.

Otolaryngologist Approach

History

The history is typically obtained from parental or consulting physician report as this patient population often presents within the first days to months of life. Symptoms may include noisy breathing, increased work of breathing with retractions, respiratory distress, apneic or cyanotic episodes, and recurrent respiratory infections. The time course and evolution of symptoms, description of any noisy breathing, and any alleviating or aggravating factors should be elicited. The otolaryngologist should inquire as to feeding difficulties, reflux symptoms, and if weight gain has been appropriate. Any prior airway surgeries and history of intubation should be discussed. Furthermore, associated syndromes or congenital anomalies may provide clues as to the diagnosis. In older children, it is important to assess for exercise intolerance or dyspnea on exertion.

Of note, patients with congenital tracheal stenosis may present in an emergent fashion and

available history may be minimal. In these cases, information regarding noisy breathing, the presence or absence of a cry, history of polyhydramnios, known syndromes or cardiac anomalies, and prior attempts at obtaining or evaluating the airway should be elicited.

Exam

From the otolaryngologist's perspective, the examination should begin with determining the degree of respiratory distress and if urgent intervention is required. This includes assessment of stridor, retractions, work of breathing, cyanosis, and apnea. Evaluation for other anomalies (i.e., craniofacial anomalies, chest wall deformities, limb abnormalities), listening to the quality of the cry, and assessment of the vital signs and growth chart should be performed.

Differential Diagnosis

The differential diagnosis for a child presenting with noisy breathing or respiratory distress from a congenital tracheal anomaly is outlined in Fig. 36.5. Several other tracheal pathologies that should be considered are acquired stenosis, a tracheal bronchus, and tracheal injury. Acquired tracheal stenosis can occur following intubation or tracheostomy. Congenital or acquired subglottic stenosis may also present very similarly. A tracheal bronchus occurs in 0.1–5% of patients and can be associated with congenital tracheal stenosis, Down syndrome, tracheoesophageal fistula, and rib abnormalities. While it is usually an asymptomatic, incidental finding, in some children, it can be a source of recurrent pneumonia, stridor, and cough [67–69].

Additional levels of airway obstruction should be considered and may be found concomitantly. In addition to affecting the trachea, malacia can also affect the pharynx, larynx, and bronchi. In children with generalized hypotonia, these may all occur to some degree. Choanal atresia and pyriform aperture stenosis can cause significant respiratory distress, particularly in the neonatal period when the child is an obligate nasal breather. Adenotonsillar hypertrophy is typically not seen in the first year of life but is a frequent contributor to upper airway obstruction in older children. Also at the pharyngeal level, micrognathia, glossoptosis, and macroglossia can result in severe airway obstruction. These are more commonly seen in syndromic children or those with Pierre Robin sequence.

Instrumental Assessment

Endoscopic Assessment

Flexible fiber-optic laryngoscopy may be performed in the office setting to evaluate for supraglottic and glottic anomalies, such as laryngomalacia and vocal fold immobility. Information regarding the subglottic and tracheal airway, however, will be limited with this exam.

Evaluation of tracheal pathologies is best performed with a combined flexible and rigid bronchoscopy in the operating room. Flexible bronchoscopy is advantageous for assessing the degree and locations of malacia, as well as response to positive pressure and normal bronchial branching patterns. In cases of tracheomalacia, rigid bronchoscopy may stent the airway open and underestimate the degree of malacia. On the other hand, rigid bronchoscopy provides the most accurate assessment of the length and degree of airway stenosis and best evaluation of laryngotracheal clefts. Formal sizing of the stenotic airway can be performed using endotracheal tubes or modified endotracheal tubes [70]. This also allows airway growth to be monitored over time in patients who are being managed conservatively. Importantly, care must be taken not to traumatize the mucosa in cases of severe stenosis as even mild swelling can precipitate a critical situation. Rigid and flexible bronchoscopy, as well as esophagoscopy, provides useful information regarding tracheoesophageal fistulas and vascular compression. Table 36.1 delineates the areas of airway compression associated with specific vascular anomalies.

Table 36.1 Site of vascular compression on diagnostic studies

Congenital vascular anomaly	Bronchoscopy	Esophagoscopy/barium swallow
Double aortic arch	Anterior and bilateral	Posterior and bilateral
Right arch anomalies	Right anterolateral	Right posterolateral
Left arch anomalies	Left anterolateral	Left posterolateral
Anomalous innominate artery	Anterior	N/A
Pulmonary artery sling	Posterior	Anterior
Aberrant right subclavian artery	N/A	Posterior

Imaging Modalities

Plain films of the airway may suggest areas of narrowing; however, these are not definitive and further evaluation is required. Children with congenital tracheal stenosis or concern for vascular compression should undergo a high-resolution contrast-enhanced computed tomography (CT) scan of the chest with three-dimensional reconstruction, as well as an echocardiogram to evaluate for coexisting cardiac anomalies. Magnetic resonance angiography can also be employed. While imaging primarily is used to evaluate the intrathoracic vasculature, it can also aid in surgical planning for the airway. CT imaging may underestimate the degree and length of airway narrowing, however, and should be used adjunctive to endoscopic evaluation [14, 71].

Evaluation of Swallowing

These studies will be further discussed in the Speech-Language Pathology approach portion of the chapter. Briefly, a fiber-optic endoscopic evaluation of swallowing (FEES) and videofluoroscopic swallowing studies (VFSS) are often employed to help assess the safety for feeding. They can be used individually or combined to provide complementary information. Additionally, a barium swallow may suggest the presence of vascular compression, but further evaluation with endoscopy and a CTA would be necessary.

Management

A collaborative team approach is recommended in these medically complex children. This often involves some combination of pediatric otolaryngologists, pulmonologists, gastroenterologists, cardiothoracic surgeons, general surgeons, intensivists, neonatologists, and speech-language pathologists.

Congenital Tracheal Stenosis

The minimally symptomatic subset of patients with complete tracheal rings may be managed conservatively with serial microlaryngoscopy and bronchoscopy (MLB) to ensure continued tracheal growth and an adequate airway. While previous reports have shown approximately 17% of patients with complete tracheal rings can be managed conservatively, the majority of children will require surgical repair [49, 70].

The method of surgical repair for complete tracheal rings has evolved over the past several decades. Primary resection with reanastomosis [3], augmentation tracheoplasty (with costal cartilage [72–74], pericardial patch [75–77], tracheal autograft [78], or cadaveric homograft [79, 80]), tracheal allografting [81, 82], and laser division with balloon dilation [83] have all been described and successfully employed. However, since it was first described by Tsang [84] and popularized by Grillo [85, 86], slide tracheoplasty has emerged as the largely undisputed method of choice for repairing complete tracheal rings as it avoids the problems of excessive tracheal shortening with anastomotic tension, circumferential scarring, loss of mucociliary clearance, graft prolapse, and granulation around grafts [87]. Additionally, the versatility of slide tracheoplasty allowed it to be adapted for use in repairing tracheoesophageal fistulas [88] and the residual pouches [89], long-segment acquired tracheal stenosis (and even short-segment stenoses) [90], sleeve trachea [4], deficient tracheal or bronchial rings [7], and salvage airway rescue [16, 91]. Historically, the prognosis was quite poor with a mortality rate

for tracheal stenosis approaching 80%. Prognosis now depends more on the child's comorbidities than the stenosis itself [4].

Tracheal webs typically involve only a short segment of the trachea and can generally be divided endoscopically. Occasionally, a short-segment resection or slide tracheoplasty may be required [2, 40]. Tracheal agenesis is typically an unrecoverable condition. If a tracheoesophageal fistula is present, esophageal intubation may allow for temporary ventilation, but this is not usually sustainable. If diagnosed prenatally, an ex utero intrapartum treatment (EXIT) procedure may allow for low tracheostomy if a distal trachea is present or being placed on ECMO [2, 15, 40].

Tracheomalacia

Many children with tracheomalacia will not require intervention and will outgrow the symptoms by 1–2 years of age as the tracheal cartilage becomes more rigid [29, 34, 92]. Intervention should be dictated by the child's symptoms – not by how the airway looks. In children who have significant symptoms secondary to the malacia (e.g., frequent respiratory infections, chronic cough, exercise intolerance, ALTEs, respiratory failure, failure to thrive, or bronchiectasis), available therapies include medications, positive pressure, and surgery.

Medical therapy is aimed at increasing the smooth muscle tone in the trachealis, thus decreasing tracheal compliance [93, 94]. Bethanechol is a cholinergic agonist that, when dosed at 0.1 mg/kg/dose three to four times per day, can increase trachealis tone with a low risk of inducing bronchospasm [95]. Given this pathophysiology, one can imagine how bronchodilators, such as albuterol, could have deleterious effects on the smooth muscle tone and worsen tracheomalacia. Additionally, gastroesophageal reflux has been shown to contribute worsening laryngotracheal symptoms. Since up to 75% of children with tracheomalacia have reflux, aggressive reflux management may improve symptoms [54, 96].

Positive airway pressure acts to stent the airway open throughout the respiratory cycle. This can be administered in a noninvasive fashion (e.g., facial or nasal mask) or via an invasive mechanism with an endotracheal tube or tracheostomy [97, 98]. Additionally, the endotracheal or tracheostomy tube itself may act as somewhat of an airway stent. As such, a longer, flexible tracheostomy tube is often preferable in these patients. The otolaryngologist may be asked to perform a tracheostomy in cases where the child has intractable tracheomalacia, tracheomalacia not amenable to a lesion-specific surgical intervention, or when bronchomalacia is also present. This remains one of the most common interventions for these patients [4].

Finally, for children with certain airway lesions and severe symptoms or those unresponsive to the above therapies, other surgical intervention may be necessary. In cases of primary tracheomalacia related to absent tracheal rings [7], residual tracheal pouches after repair of tracheoesophageal fistula [89], or secondary focal collapse from external compression or tracheostomy-related issues, slide tracheoplasty can be employed [16]. Additionally, tracheopexy with a variety of approaches and techniques has also been described [99–102]. When there are other sources of external compression, the inciting source should be addressed. For example, skeletal anomalies can be addressed by surgery for scoliosis or pectus excavatum and neck or mediastinal masses may be excised. Briefly, if there is external vascular compression contributing to malacia, this is typically repaired by cardiothoracic and/or pediatric surgery, depending on the anomaly and need. Aortopexy with thymectomy and anterior suspension is the mainstay for innominate artery compression [29, 103–106], but innominate artery reimplantation has also been described [107, 108]. In cases of other vascular rings and slings, division and/or relocation of the vessels is employed as indicated [13, 109]. It should be noted that some degree of tracheal deformity often persists even after repair of these anomalies [110, 111]. Rarely and in certain situations, both internal and external tracheal stents have been used for management of tracheomalacia [112–115]. These approaches, however, should be used with caution, in short-term situations, and after other therapies have

failed due to risk of malacia recurrence, granulation, mucosal erosion, stent migration, and mortality, particularly in the pediatric population [29, 34, 116, 117].

Operative Approach

This section will focus on the approach for repair of complete tracheal rings. While this does sometimes involve concurrent repair of congenital cardiac anomalies and vascular rings/slings, details of those interventions are beyond the scope of this chapter.

Indications

Surgery is indicated in the presence of significant airway obstruction, respiratory failure, worsening respiratory status, recurrent cyanotic or apneic spells, or dyspnea on exertion. Additionally, an airway that does not accommodate a 2.5 endotracheal tube, even in the absence of significant symptoms, should be considered tenuous and in need of surgical repair. In general, the younger a child is at initial presentation, the more likely surgical repair will be required [49]. Slide tracheoplasty may also be applied to patients with acquired tracheal stenosis, tracheoesophageal fistulas or pouches, tracheal sleeves, absent tracheal rings, or salvage cases [16].

Key Aspects of the Consent Process

Risks associated with cardiac bypass and sternotomy should be discussed by the cardiothoracic surgeon. Regarding slide tracheoplasty, there are both intraoperative and postoperative risks that should be discussed. Intraoperatively, there is always risk for loss of airway prior to being placed on bypass, particularly of concern in children with very stenotic complete tracheal rings. During dissection, there is risk to the great vessels, trachea, bronchi, and recurrent laryngeal nerve.

Equipment

This procedure is most commonly performed in conjunction with pediatric cardiothoracic surgery via a sternotomy approach on cardiac bypass. Thus, a pediatric cardiac anesthesiologist and an ECMO team are required. In the senior author's experience, it is also advantageous to have a pediatric pulmonologist who is adept in airway bronchoscopy available to assist throughout the case.

Steps

1. *Patient positioning and preparation*

   The patient is brought to the operating room and placed in the supine position on the operating table. Depending on the patient's current airway status and intravenous access, the patient typically undergoes a rigid and/or flexible bronchoscopy at the beginning of the case. If the patient is not already intubated, microlaryngosopy and bronchoscopy with a Phillips 1 blade and rigid Hopkins rod 0-degree telescope should be performed. A 1.9 mm (scope 27,017) or 2.7 mm (scope 10,018) Hopkins rod should be available based on the anticipated diameter of the complete tracheal rings. The telescope or endotracheal tubes of various sizes (with or without modification, Fig. 36.6) can be used to estimate the diameter of the complete tracheal rings [70]. Sizing is of less import, however, if the need for repair is imminent and care should be taken not to traumatize the mucosa, particularly in a patient with very small-diameter rings. After endoscopy is complete, an endotracheal tube is placed with the tip sitting above the level of the complete tracheal rings. Ideally, at least a 3.5 endotracheal tube is placed to allow for therapeutic bronchoscopy with a 2.8 mm flexible bronchoscope. Nasotracheal intubation is also beneficial both intraoperatively (i.e., easier access, less kinking, and less migration) and if the patient will not be

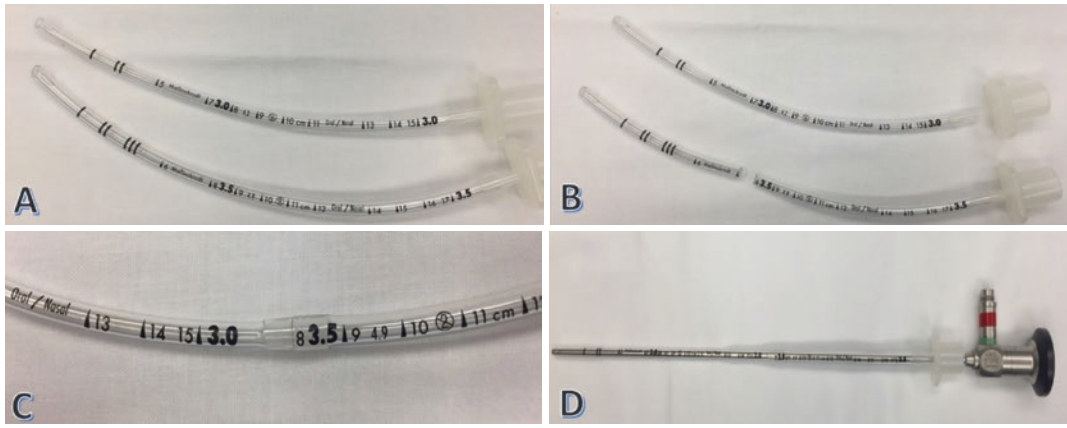

Fig. 36.6 Creation of a modified endotracheal tube (ETT) to allow for sizing of the distal airway. (**a**) Standard 3.0 and 3.5 uncuffed ETTs are used in this case. A half-size larger tube will fit over a half-size smaller tube. (**b**) The larger of the tubes is cut at the mid-to-distal one third. The connector should be removed from the smaller size tube. (**c**) The proximal end of the smaller-diameter tube is then inserted within the distal end of the larger-diameter (cut) tube. The larger-diameter tube adapter should remain in place. (**d**) This unit is then fed over appropriate length and size telescope for sizing. In this example, the airway would then be sized with a 3.0 ETT. (From Wilcox et al. [70], with permission)

extubated at the conclusion of the case. Once the airway is secure, appropriate anesthetic lines and any additional monitoring can be performed.

2. *Exposure*

   At this point, the cardiothoracic surgeon will place the patient on bypass and perform the sternotomy. Adequate exposure of the trachea is then ensured jointly by the cardiothoracic and otolaryngology surgeons. Adequate tracheal mobility is important, but care must also be taken to preserve both the tracheal blood supply and the recurrent laryngeal nerves laterally.

3. *Identification of the complete tracheal rings*

   With the trachea adequately exposed and mobilized, the flexible bronchoscopist is then asked to perform a bronchoscopy via the endotracheal tube. A 30-gauge needle is then placed at the expected start and end points of the complete tracheal rings. These positions are confirmed endoscopically as complete tracheal rings are surprisingly difficult to identify via external examination of the trachea alone. It is crucial that the full length of the complete rings be addressed and that the approximate midpoint of the complete rings can be determined.

4. *Entering the trachea* (Fig. 36.7a)

   The trachea is then entered using a Beaver blade to make a transverse incision just proximal to the approximate midpoint of the complete tracheal rings. The cut should be slightly beveled from superior to inferior.

5. *Beveling the divided tracheal edges*

   The edges of the trachea are then further beveled using scissors.

   – On the proximal aspect of the divided trachea, the edges are beveled from anterior to posterior.
   – On the distal aspect of the divided trachea, the edges are beveled from posterior to anterior.

6. *Dividing the complete tracheal rings* (Fig. 36.7b)

   The complete extent of the length of the complete tracheal rings is then divided in the following fashion:

   – On the *proximal* aspect of the divided trachea, a midline incision is carried from

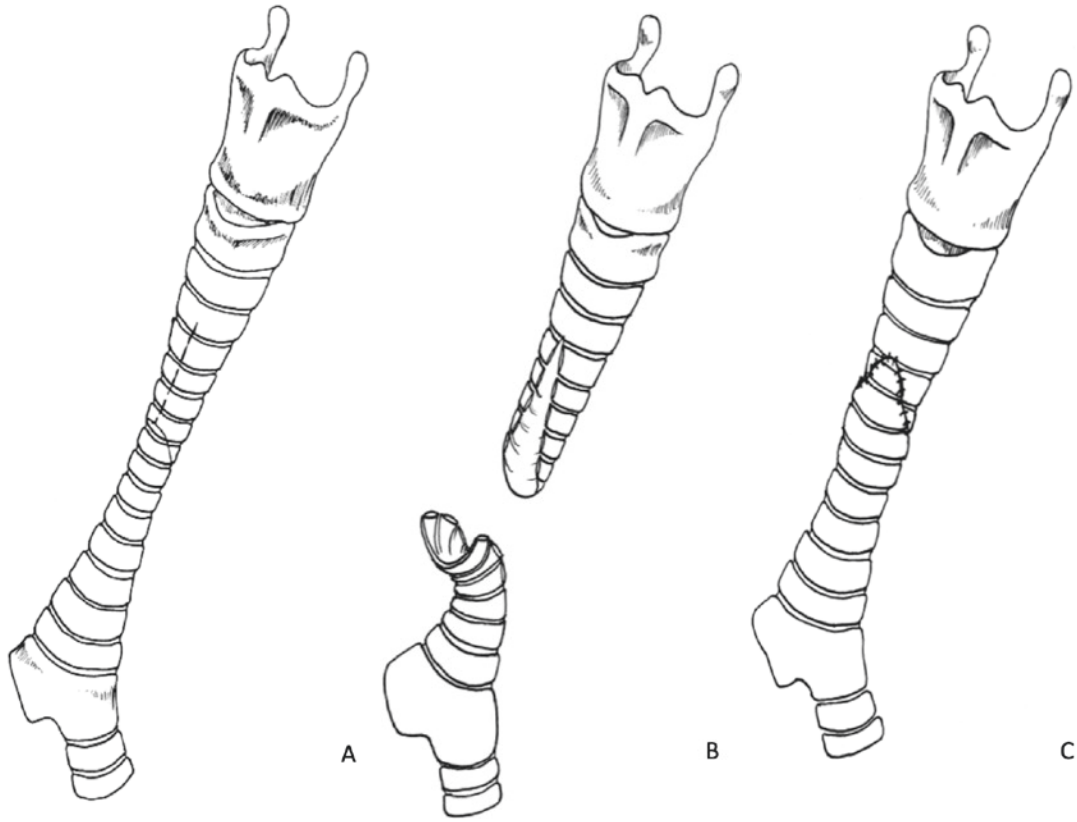

Fig. 36.7 Slide tracheoplasty technique. (**a**) Initial transverse incision just proximal to the approximate midpoint of the complete tracheal rings. (**b**) Dividing the length of the complete tracheal rings along the anterior aspect of the proximal trachea and posterior aspect of the distal trachea. (**c**) The trachea is then slid on itself and reanastomosed in a running fashion using a double-armed 6–0 PDS suture on a BV-1 needle. (From DeMarcantonio et al. [125], with permission)

the divided edge inferiorly up to the beginning of the complete rings superiorly along the *anterior* tracheal wall using scissors.

– On the *distal* portion of the trachea, a midline incision is made using scissors on the *posterior* tracheal wall inferiorly through the extent of the complete rings.

Of note, the orientation of the division can be altered, the cuts can be extended into a main stem bronchus, or the cuts can be extended into the anterior cricoid as needed to address patient-specific anatomic concerns.

7. *Confirm that all complete rings have been divided*

It is crucial that the surgeon confirm that all complete rings have been divided. This should be confirmed by direct intraluminal visualization and palpation. Extending the cuts into normal trachea is prudent.

8. *Reanastomosis* (Fig. 36.7c)

The trachea is then slid on itself, allowing it to become shorter and wider. Placing bilateral retraction sutures on the distal trachea may help reduce tension during suturing. Reanastomosis is then performed using a double-armed 6-0 PDS suture on a BV-1 needle. Suturing begins at the posterior apex and extends laterally and anteriorly until cartilage is reached. Shod forceps are used to tag the first end as suturing is then carried in the opposite direction along the posterior

wall and laterally until cartilage is reached with the second needle. The anastomosis is completed anteriorly after using blunt right-angle nerve hooks to tighten any loose loops in the suture. Care should be taken to evert the edges as the suture is tightened down to help prevent a figure-8 tracheal deformity. The suture is then tied down on the anterior aspect of the trachea.

9. *Closing*

   The wound bed is irrigated and hemostasis is assured. A leak test is performed up to 30 cm water. A fibrin sealant is then placed over the suture line. The cardiothoracic team then proceeds with the closure. At the same time, the pulmonologist typically repeats a flexible bronchoscopy to suction the endotracheal tube and lungs prior to decannulating the patient off bypass. During the remainder of the procedure, the anesthesiologist is asked to place several saline drops down the endotracheal tube every 15 min.

10. *Final bronchoscopy*

    The Pulmonary team performs a final bronchoscopy after the patient is taken off bypass. At this point, the endotracheal tube position is confirmed if the patient is to remain intubated.

Postoperative Management and Follow-Up

The patient is typically extubated within 48 h, if not on the table in the operating room. Positive pressure is typically avoided if possible or kept at less than 30 cm water. After extubation, high humidity is provided via a constant face mask and nebulizers to prevent thick secretions from trapping at the anastomotic site. A repeat microlaryngoscopy and bronchoscopy is typically performed in the operating room 1 week and 2 weeks postoperatively. As long as healing seems appropriate, additional follow-up endoscopies are then typically spaced out by doubling the time between examinations (i.e., 2 weeks, 4 weeks, 8 weeks, etc.). In the author's experience, if the repair looks good and the child is doing well at 3 months postoperatively, they tend to continue to do well in the long term [118].

Emerging and Evolving Techniques of the Future

The slide tracheoplasty has been the major recent development for congenital tracheal stenosis. In the future, more options for airway replacement and augmentation with potential for patient-specific customization, including 3D printing and tissue engineering, as well as tracheal transplantation, may be available [119–122].

Regarding malacia, more procedures are being performed using less invasive methods. Airway stenting, both internal and external, is continuing to evolve. Biodegradable stents, as well as drug-eluting stents, are being developed and trialed [123, 124]. Future studies will hopefully provide a more mainstream method of stenting that avoids many of the current issues with stents.

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Eosinophilic Esophagitis

37

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Overview

Eosinophilic esophagitis (EoE) is a chronic, immune-mediated disorder of the esophagus that commonly presents with dysphagia in the pediatric population and is characterized histologically by eosinophil-rich inflammation. Traditionally, these patients have been managed by providers in allergy/immunology, gastroenterology, and otolaryngology. With increased knowledge regarding disease presentation and progression, improved assessment and treatment strategies have been developed. When feeding dysfunction and airway conditions present, a multidisciplinary approach to management with speech-language pathology and otolaryngology has been employed. This chapter reviews the clinicopathology of eosinophilic esophagitis as well as its evolving multidisciplinary treatment.

Epidemiology

The first cases of suspected eosinophilic esophagitis were reported in the 1960s–1970s, but EoE was first described as a distinct clinical entity in the 1990s [1]. EoE is considered a rare disease, yet the prevalence (or recognition) of EoE in the pediatric population has risen significantly in the past few decades. Average annual percentage increases in prevalence have been reported from 12% to 56% [2, 3]. Population-based prevalence estimates vary depending on study, location, and definitions. A study in the United Kingdom reported a prevalence of 0.2 cases per 100,000 children [4]. A rate of 8.9 cases per 100,000 children was reported in Australia [5]. Noel et al. reported a prevalence of 43 cases per 100,000 children in Ohio [2]. A recent report by Robson

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et al. found a prevalence of 118 cases per 100,000 children in Utah [6].

Pathophysiology

Eosinophilic esophagitis is characterized by predominate eosinophilic infiltration of the esophageal mucosa. Studies have demonstrated that this inflammatory reaction involves numerous cell types including T helper (Th) 2 cells, basophils, mast cells, and invariant natural killer T (iNKT) cells and cytokines such as eotaxin-3, interleukins (IL-4, IL-5, IL-13), and thymic stromal lymphopoietin (TSLP) [7–11]. After activation by antigen-presenting cells (APCs) and iNKT cells, Th2 helper cells secrete IL-4, IL-5, and IL-13. IL-5 stimulates the growth, differentiation, and survival of eosinophils. IL-4 and IL-13 secreted by Th2 cells promote the release of eotaxin-3 and TSLP by epithelial cells and increase eosinophilic migration [11]. Eotaxin-3 is a strong chemotactic agent for eosinophils. TSLP primes Th2 responses through the activation of APCs and prevents apoptosis of eosinophils by direct activation of the TSLP receptor present on eosinophils [11].

Eosinophils synthesize and release many proteins such as major basic protein, eosinophil peroxidase, eosinophil cationic protein, eosinophil-derived neurotoxin, and transforming growth factor beta (TGF-β). Esophageal biopsies from patients with EoE typically show signs of eosinophil degranulation, suggesting that direct damage to the esophageal epithelium by granule proteins may contribute to disease pathogenesis [12, 13]. TGF-β secreted by eosinophils can contribute to esophageal fibrosis by inducing myofibroblast activation and proliferation and esophageal dysmotility by inducing smooth muscle hyperplasia [14].

There is a higher risk of EoE in monozygotic twins (41%) and in families with affected patients which suggests a genetic predisposition for EoE [15, 16]. Single candidate gene studies have identified single nucleotide polymorphisms in the genes encoding for eotaxin-3 (*CCL26*) and filaggrin (*FLG*) as being associated with EoE [17, 18]. Genomic association studies have identified EoE genetic risk loci such as the 2p23 locus encoding the *CAPN14* gene, 5q22 locus encoding the *TSLP* and *WDR36* genes, 11q13 locus encoding the *EMSY* and *LRRC32* genes, and 16p13 locus encoding *CLEC16A*, *DEX1*, and *CIITI* [19–22]. EoE has often been reported in patients with Mendelian-inherited connective-tissue disorders such as Marfan syndrome, Ehlers-Danlos syndrome, and Loeys-Dietz syndrome, which are associated with increased TGF-β signaling [23]. The risk of EoE was found to be increased eightfold in patients with these connective-tissue disorders.

Presentation

EoE has been reported to be present from infancy into adulthood. Children under the age of 18 years comprise almost a quarter of the patients with EoE [24]. EoE presents more commonly in males (range 66.2–83%) and has a Caucasian predominance [2, 5, 6, 24–26]. Children with EoE typically present with symptoms of esophageal dysfunction. However, the clinical presentation can vary by age. Younger children are more likely to present with liquid dysphagia, feeding difficulties, failure to thrive, and nonspecific gastrointestinal issues such as reflux, vomiting, and abdominal pain, whereas more specific conditions such as solid food dysphagia and food impaction can be the presenting symptoms in preteens and adolescents [2, 25–27]. Patients can also develop chest pain and early satiety.

Endoscopic findings of esophageal furrowing (Fig. 37.1) and patchy infiltrates are common in the early stages of EoE. Fibrostenotic changes such as concentric rings, fixed structures, and luminal narrowing are frequent late endoscopic findings in EoE patients with uncontrolled inflammation. Dysphagia that is non-responsive to therapy is suggestive of esophageal fibrostenosis which should be investigated with videofluoroscopy and esophagram and may respond to endoscopic dilation.

EoE in children is often associated with atopic disease. Approximately 33–67% present with asthma, 30–90% have allergic rhinitis, and

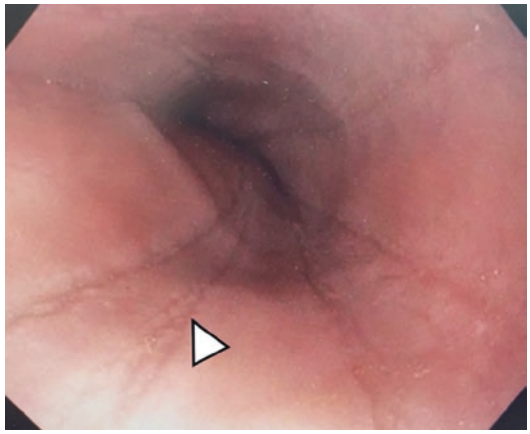

Fig. 37.1 Esophageal linear furrowing (white arrowhead)

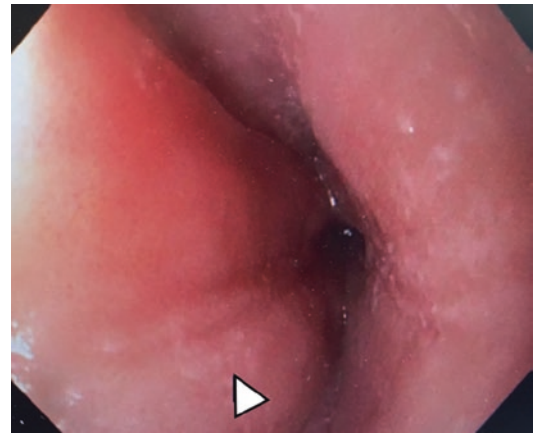

Fig. 37.2 White plaques in esophagus (white arrowhead)

20–60% have atopic dermatitis [25, 26, 28, 29]. Almost a quarter of the pediatric EoE population presents with food allergen sensitization [28, 29]. Multiple studies suggest that atopy and specifically food allergy is associated with more severe symptoms at presentation and resistance to steroid treatment in a subset of EoE patients [30]. Thus, recognition and treatment of atopic disease in children with EoE is a key component of their multidisciplinary care.

Gastroenterologist Approach

Eosinophilic esophagitis (EoE) is an increasingly common disease affecting children and adults alike. It is characterized by progressive eosinophilic infiltration and fibrosis of the esophagus. These changes lead to impaired esophageal function and swallowing difficulties.

Clinical Presentation

Esophageal phase dysphagia is a common presentation of EoE. This is typically seen in school-age children and teenagers who present to the emergency department with esophageal food impaction. Many of these children will describe prior episodes of food feeling "stuck" in their esophagus and engaging in maneuvers to dislodge the bolus, such as drinking water. Vomiting is another common symptom, particularly in infants and young children. Other symptoms include abdominal pain and feeding refusal [31]. There are no characteristic physical or laboratory findings of EoE. Diagnosis rests on endoscopic and histological findings.

Diagnosis

Endoscopic findings suggestive of EoE include edema, linear furrowing (Fig. 37.1), white plaques (Fig. 37.2), trachealization, and paper esophagus [32]. Esophageal stenosis can be found in advanced cases. However, the esophagus can appear normal in some patients (Fig. 37.3), especially in infants and young children. Mucosal biopsies will show eosinophilic infiltration, eosinophilic microabscesses, basal cell hyperplasia, and varying degrees of fibrosis involving the lamina propria. Eosinophilic infiltration of the esophagus is the cardinal feature, and most authorities consider greater than 15 eosinophils per high-power field consistent with EoE [32–34]. The predictive power of this finding is greatest when present in biopsies proximal to the distal esophagus. For this reason, obtaining proximal and distal esophageal biopsies is recommended when evaluating children for EoE (Fig. 37.4). The differential diagnosis includes gastroesophageal

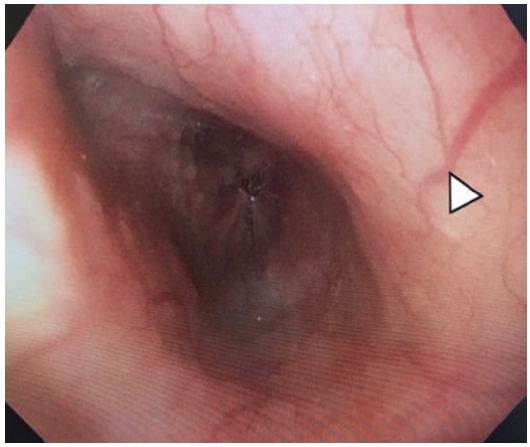

Fig. 37.3 Normal esophageal mucosa with apparent blood vessels (white arrowhead)

reflux disease, inflammatory bowel disease, and eosinophilic gastroenteritis. The latter two should be suspected when the eosinophilic infiltration extends beyond the esophagus.

Treatment

The treatment of confirmed EoE usually involves collaborative care from gastroenterology, allergy, otolaryngology, and nutrition. The treatment goals include providing relief from current symptoms and preventing progression to esophageal stricture. The treatment strategy can be divided into two broad categories. The first is elimination of the offending antigen if possible.

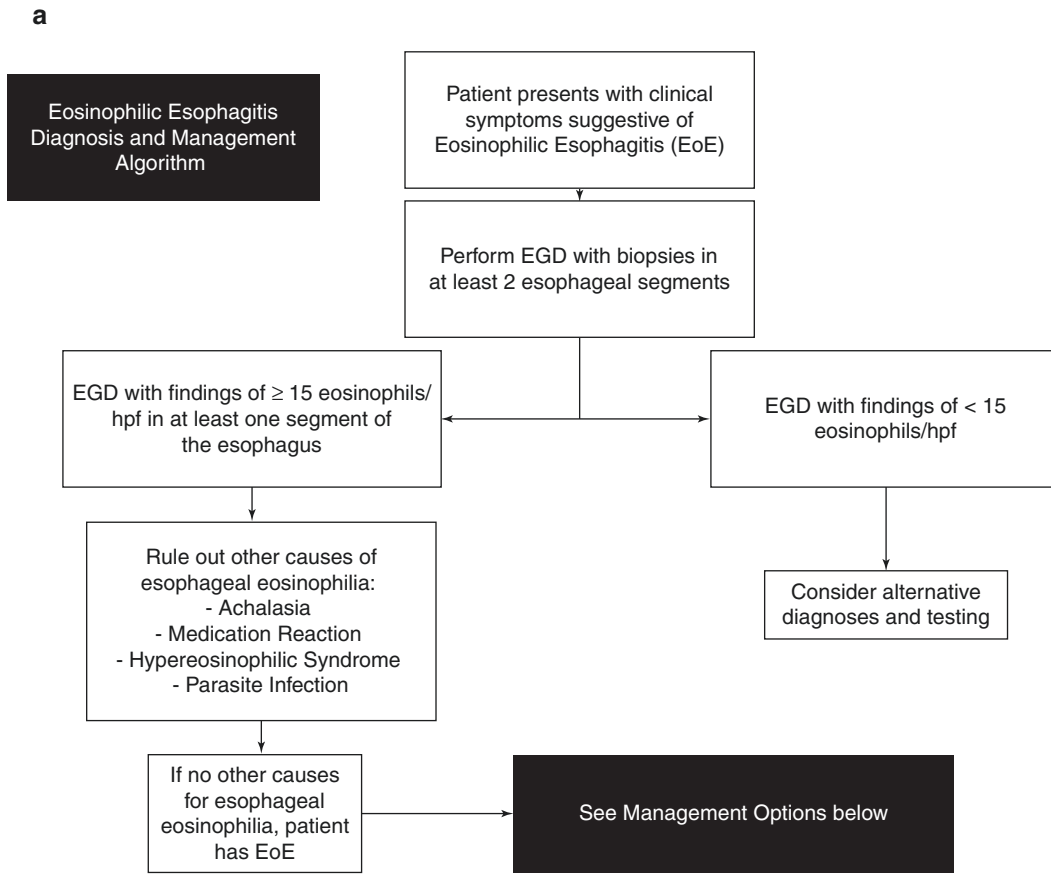

Fig. 37.4 Clinical algorithm for the diagnosis (**a**) and management (**b**) of eosinophilic esophagitis

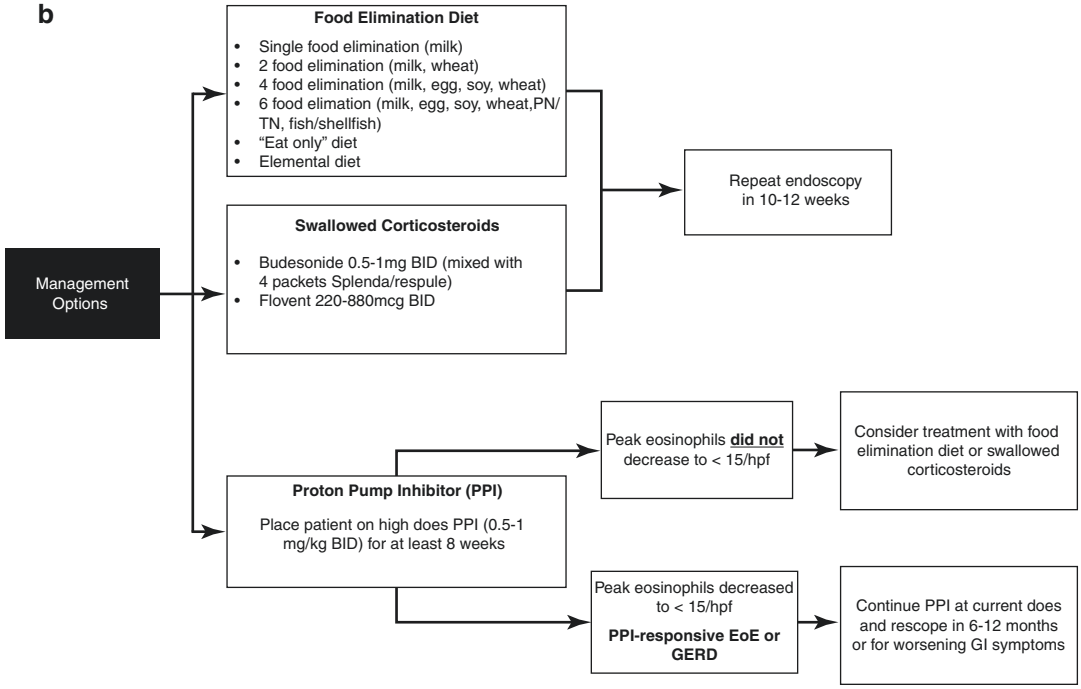

Fig. 37.4 (continued)

This strategy is not possible if the antigen cannot be identified. It may also not be possible if it is part and parcel of the patient's environment or if the patient has numerous inciting antigens. The expertise of an allergist is key to this strategy. Food antigens are most amenable to this strategy, and empiric elimination of the common foods associated with EoE is often carried out (Fig. 37.4). This can be done one at a time. A more common practice is to remove four or six of the most likely foods all at once and then reintroduce them one at a time after histological follow-up. Sometimes an elemental diet may be required. Empiric food elimination is challenging and requires the involvement of an expert dietitian. Drug therapy is the second approach. Topical steroid therapy in the form of a slurry or swallowed from a metered dose inhaler is the most widely used drug treatment. Drug therapy is useful when an antigen cannot be found or removed or where compliance with an elimination diet is inadequate [35].

Prognosis

The main long-term complication of uncontrolled EoE is esophageal stricture formation. Compliance with an effective diet or drug regimen should prevent this complication. Unfortunately, compliance can be problematic in part due to the difficulty of adhering to an elimination diet or cumbersome treatments. This can be compounded by the lack of symptoms to motivate the patient despite ongoing destructive esophageal inflammation. At present, histological follow-up is the only practical way to assure treatment efficacy, but parents may be reluctant to subject their children to the endoscopy required to obtain the biopsies for histology.

Otolaryngologist Approach

Eosinophilic esophagitis (EoE) is a complex inflammatory disorder of the esophagus that is an increasingly important diagnosis for children with

otolaryngologic conditions. Although the presenting symptoms are typically gastrointestinal in nature, aerodigestive complaints are common in children and can greatly impact their quality of life. Otolaryngologists will encounter pediatric EoE patients as they present for initial evaluation of dysphagia or airway symptoms, now commonly in multidisciplinary aerodigestive centers. Thus, the otolaryngologist plays a key role in the diagnosis and management of EoE.

Clinical Manifestations

The presenting complaint for EoE in the otolaryngology setting is similar to gastroenterology with one or more symptoms of dysphagia. In young children, this includes laryngopharyngeal reflux, regurgitation, vomiting, food avoidance, liquid dysphagia, failure to thrive, and nonspecific gastrointestinal complaints. Atopic dermatitis (atopy) in a child with uncontrolled laryngopharyngeal reflux may be evidence of EoE related to cow's milk protein allergy [36]. The adolescent will present with difficulty swallowing and discomfort with solid food. However, dysphagia with solid food and food impaction is usually a late finding of EoE and a direct consequence of uncontrolled eosinophilic inflammation leading to esophageal fibrostenosis, a result of chronic secretion of cytotoxic agents that damage the esophageal epithelium and the release of remodeling factors such as transforming growth factor beta [12–14].

Nasal symptoms and rhinosinusitis are reported in approximately one-quarter of children with EoE [37, 38]. Symptoms include nasal congestion, obstruction, rhinorrhea, postnasal drip, facial pressure and pain, cough, pharyngitis, fever, and headache. There are several common symptoms shared between EoE and rhinosinusitis including chronic cough, hoarseness, and dysphagia [39]. Eosinophilic inflammation is a major hallmark of both rhinosinusitis and EoE. Tissue response to eosinophil degranulation is likely the pathophysiologic connection between the two diseases [39]. A possible mechanism for the relationship between EoE and rhinosinusitis is chronic allergen exposure through the nose and mouth. Chronic exposure in the nose results in rhinosinusitis. When the allergens are swallowed and enter the esophagus, eosinophil activation results in mucosal damage and EoE. In patients with recalcitrant gastrointestinal manifestations of rhinosinusitis (vomiting, regurgitation, nocturnal cough), EoE is a likely culprit.

Children with EoE may present with laryngeal symptoms of cough, hoarseness, globus sensation, and sleep-disordered breathing. Patients with EoE-associated laryngeal disease share similar characteristics to those with gastrointestinal symptoms, including a history of atopy and allergic rhinitis [39]. The initial indication may suggest laryngopharyngeal reflux as the cause of these symptoms. However, several reports describe patients with laryngeal disease refractory to acid reflux therapy who were eventually diagnosed with EoE [40–42].

EoE has also been implicated in the etiology of laryngeal diseases such as recurrent croup and subglottic stenosis. Recurrent croup is characterized by the abrupt onset of inspiratory stridor (most commonly at night) accompanied by a barky cough, hoarseness, and dyspnea [39]. Many of the triggers of recurrent croup are associated in the etiology of EoE, such as aeroallergens and asthma [37, 39, 41, 42]. Additionally, some patients with EoE and recurrent croup have been found to have concurrent subglottic stenosis (SGS). SGS is caused by chronic inflammatory changes in the subglottic region that lead to airway fibrostenosis. Patients with SGS present with symptoms of stridor, cough, croup, and airway obstruction. Severe stenosis requires surgical correction. Several groups have reported on the association between EoE and SGS [40, 41, 43]. An increased failure rate after airway reconstruction surgery has been observed in patients with EoE [44, 45]. Thus, in many institutions, an evaluation for EoE and treatment, if necessary, has become the standard of care for airway surgeons before attempting laryngotracheal reconstruction [39]. This includes esophagoscopy with proximal and distal esophageal biopsies when conducting preoperative airway surveillance (microlaryngoscopy and bronchoscopy).

The pathophysiology of laryngeal symptoms and diseases in EoE patients is poorly understood.

The larynx is located in close proximity to the oronasal cavity and esophagus, especially in young children. Airway inflammation may be mediated by esophageal eosinophilic degranulation and Th2 cytokine response leading to laryngeal inflammation and edema [39]. Diffuse laryngeal edema can lead to hoarseness and the other previously described laryngeal symptoms. Additionally, it is plausible that eosinophilic proteins such as eosinophilic cationic protein and major basic protein can exert a cytotoxic effect on airway mucosa leading to the obstructive airway symptoms seen in patients with EoE and laryngeal disease.

Management

Similar to gastroenterology, early and accurate diagnosis is key to the management of EoE. This is often in the setting of an aerodigestive work-up for dysphagia and airway stenosis. Awake flexible fiberoptic laryngoscopy is used to examine possible disease manifestation in the upper airway in the early evaluation of patients with suspected EoE.

Close collaboration with gastroenterology, allergy/immunology, nutrition, and speech is paramount to effective treatment and is often culminated in a dedicated EoE multidisciplinary clinic. Feeding modifications, dietary restrictions, proton pump inhibitor, and topical steroid therapy (inhaled or slurry) are employed. Treatment strategies are tested for effectiveness with esophageal biopsies and histologic evidence of improvement. Laryngotracheal reconstruction will be delayed until EoE is safely controlled. Solid food dysphagia and esophageal fibrostenosis will be addressed with endoscopic dilation (often repeated), but treatment of the underlying disease is still required.

Allergist Approach

As with many complex disorders, a multidisciplinary approach across otolaryngologists, gastroenterologists, speech-language pathologists, and allergists is important for patients with eosinophilic esophagitis (EoE). Multiple studies have shown the importance of allergens in triggering disease activity, and there are many similarities between the pathogenesis of EoE and other allergic disorders. As a result, the allergist is uniquely positioned to assist in the management of this disease.

Role of Allergies in EoE

EoE is considered an atopic disease similar to asthma and allergic rhinitis as all are driven by a T helper (Th) 2 cell response leading to production of cytokines including IL-4, IL-5, and IL-13 [9, 46]. These cytokines activate and recruit eosinophils, as well as mast cells and basophils, to the esophagus leading to inflammation of tissues. As a result, patients develop clinical symptoms of gastrointestinal dysfunction and are at long-term risk for developing esophageal fibrosis [14, 47]. For many, this allergic inflammation is driven by exposure to antigenic food proteins which can be lessened by removal from the diet [2, 48]. There is some evidence that aeroallergens may activate EoE in a similar manner. As a result, identification of these triggers is an important aspect of EoE management.

Role of Allergy Testing in EoE

There are three different types of testing that can be performed to confirm a clinical suspicion of allergy: skin prick testing, serum-specific IgE testing, and atopy patch testing. Skin prick testing (SPT) involves the placement of an allergen extract onto the skin which is then "pricked" with a device to introduce the allergen into the epidermis or upper dermis where it can interact with mast cells in the skin. If a patient has IgE antibodies to the allergen, a reaction will be elicited, and a wheal with surrounding erythema will develop at the test site. Serum-specific IgE testing is an ELISA-based test utilizing the Phadia ImmunoCAP system (Thermo-Fisher/Phadia, Kalamazoo, MI) to detect the presence of IgE antibodies in the serum. In IgE-mediated food

allergy, skin prick testing and/or serum-specific IgE testing are useful tests to confirm a clinical history of food allergy. Positive and negative predictive values are generally high, and clinical decisions can be made based upon the findings. Another form of testing called atopy patch testing (APT) can be utilized to identify non-IgE-mediated triggers in allergic conditions such as eczema and allergic contact dermatitis. In this form of testing, fresh foods or chemicals are placed into an aluminum Finn chamber which is then applied directly to the patient's back. The test is left on for 48 h after which it is removed and the area examined and again at 72 h. If the patient has the presence of erythema and/or vesicles at the site of the test, the reaction is considered positive.

Although it is well established that foods are a common trigger for EoE, the role of allergy testing to identify the specific food trigger is less clear. There have been multiple studies evaluating the effectiveness of different forms of allergy testing for guiding the management of EoE. Overall, the results have been somewhat disappointing. In a study by Rodriguez-Sanchez et al., 73.1% of adults with EoE could obtain disease remission by utilizing an elimination diet based upon serum-specific IgE results compared to only 53% in those who empirically eliminated the top six most common triggers [49]. Unfortunately, other studies have not found allergy testing to be as reliable. In a study by Gonsalves et al., 50 adults underwent food elimination diets for treatment of their EoE. Skin testing was able to predict triggering foods in only 13% of subjects [50]. Spergel et al., in their study of pediatric EoE, found the negative predictive value for skin prick testing and atopy patch testing to be 92% (milk, the most common trigger, was only 44%); however, the positive predictive value was only 44%. If these tests were utilized to guide dietary management, patients had histologic remission only 53% of the time [51]. In a study utilizing APT to direct treatment of EoE in adults, nearly 50% had positive testing results, but only 16% had histologic remission based upon elimination of the food(s) that caused the positive test result. The authors estimated that the sensitivity of APT for EoE was only 5.9% with a specificity of 92% [52]. In a meta-analysis of dietary interventions for EoE, allergy testing directed elimination diets led to remission in only 45.5% of subjects, which was less efficacious than empiric elimination or elemental diets [53]. Given these results, the ability of currently available forms of allergy testing is limited to accurately predict EoE food triggers.

Role of Dietary Modification in Treatment of EOE

Although allergy testing remains limited in its ability to identify specific food triggers, there is little doubt that food elimination diets are an effective form of treatment for EoE. Elimination of the suspected culprit food(s) frequently reduces esophageal eosinophilia and improves clinical symptoms. One of the seminal studies demonstrating the effectiveness of food elimination diets was performed by Kelly et al. who demonstrated that use of an elemental formula for 6 weeks could lead to significant reduction in clinical symptoms as well as number of esophageal eosinophils [48]. Subsequent studies have shown that elemental formula diets can induce remission in more than 95% of EoE patients [54, 55]. Two other approaches have also been utilized: allergen-directed elimination and empiric elimination. Allergen-directed elimination diets involve the removal of food(s) based upon clinical symptoms seen following ingestion and/or positive results determined through allergy testing. As described above, remission utilizing this form of therapy can be achieved in 24–65% of patients [49–53]. Certain foods appear to be more common triggers including cow's milk, wheat, hen's egg, and soy, which have led to the use of empiric elimination diets (Fig. 37.4). Rather than removing foods based upon history of clinical reaction or allergy testing results, food triggers are removed individually or in groups. These diets are generally more effective than allergen-directed elimination and are better tolerated by patients than elemental diets. Single food elimination of cow's milk can be effective in up to 65% of cases, and elimination of the six most common food

triggers (6-FED) (cow's milk, hen's egg, wheat, soy, peanut/tree nuts, and fish/shellfish) can be effective in up to 81% of patients [56, 57]. Other combinations such as two food (milk and wheat) and four food (cow's milk, hen's egg, wheat, and soy) elimination have also been utilized [58, 59]. In general, if single or limited group elimination diets are ineffective, patients are asked to remove additional foods until remission is obtained. Once remission is achieved, suspected trigger(s) may be kept out of the diet long-term or reintroduced one at a time with periodic endoscopic and histological evaluation. If patients experience worsening of their clinical symptoms and/or worsening endoscopic and histologic findings on subsequent endoscopy, the food is then re-removed.

Aeroallergens in EoE

Although food is the most common trigger for EoE, aeroallergens have also been implicated. The literature remains sparse, but several case reports and studies from single centers have shown that aeroallergens can trigger worsening clinical symptoms and eosinophilic inflammation of the esophagus [60, 61]. This is unsurprising given that many EoE patients have other atopic diseases, many of which can be triggered by aeroallergens. In sensitized subjects, the mechanism is believed to be due to the inhalation of pollen into the upper airway which is then swallowed, activating Th2 cytokines. There may also be cross-reactivity between food and aeroallergens that share similar protein structures leading to increased recognition of aeroallergens by tissue eosinophils and mast cells [62]. In such patients, symptoms usually worsen during the peak pollen season and improve when pollen counts subside [63–65]. Some authors have suggested that EoE is diagnosed more frequently during certain times of the year, although a meta-analysis by Lucendo et al. found no significant variations in seasonal diagnosis [66]. These findings may indicate that EoE patients living in certain regions may be more affected than others. It is also uncertain whether aeroallergens actually trigger disease development or serve as an exacerbating factor only. Management of those patients affected by aeroallergens is also uncertain. Typical seasonal allergy treatments such as antihistamines do not seem to reduce clinical symptoms or eosinophilic inflammation. Swallowed corticosteroids do abate both clinical symptoms and inflammation and have been utilized as treatment during pollen seasons. Allergen immunotherapy, which is an effective long-term treatment for seasonal allergic rhinitis, has an unclear role in EoE. There has been limited work to evaluate its utility in treating EoE, although at least one case report has suggested efficacy [67].

Summary

The allergist plays an important role in the multidisciplinary approach to managing EoE. Given the association with other atopic disorders, the allergist is uniquely positioned to accurately assess food and environmental triggers as well as other confounders such as allergic rhinitis that may impact a patient's disease. Although the role of current allergy testing is limited, there is important information that can be gleaned that could guide management. For those patients that elect to start a food elimination diet, the allergist, in combination with a dietitian, can ensure successful implementation. Close collaboration with other specialists, including otolaryngology, gastroenterology, and speech-language pathology, should be consistent to optimize the changes for obtaining disease remission.

Speech-Language Pathologist Approach

First recognized as its own disease entity in 1993, eosinophilic esophagitis (EoE) has been increasing in prevalence in the pediatric population over the past two decades. Since reported symptoms for infants and children often vary, an EoE diagnosis can be delayed for years [27]. Before a diagnosis of EoE is made, the speech-language pathologist (SLP) may be the first specialist referral for these children with feeding difficulties.

History

Children with reported feeding issues should initially undergo a thorough clinical feeding evaluation under the supervision of an experienced SLP with knowledge of pediatric feeding and swallowing dysfunction. This referral may come from the child's primary care physician or after the child has had an initial visit with a specialist such as an otolaryngologist, gastroenterologist, or allergist.

Children referred for a feeding evaluation may present with any of the following conditions: failure to thrive, vomiting, reflux symptoms, choking with liquids, lengthy feeding times, and/or gagging or choking with early solids [25, 27]. Toddlers and older children referred for a feeding evaluation are often described by caregivers as "picky eaters," who will not accept solids that are not pureed. These children also typically prefer liquids to solids [68].

During the feeding evaluation, the SLP obtains detailed information from the caregiver about the child's past and present feeding history, including any other concomitant medical issues that could be negatively impacting the child's feeding progression. These would include upper airway anomalies, reflux, and possible food allergies. Information about the feeding environment at home and the child's behaviors during mealtimes is also pertinent. During the feeding evaluation, the child should be observed by the SLP with a typical feeding.

Oral Sensory and Oral Motor Assessment

The child's oral motor and oral sensory systems are assessed during the clinical evaluation. Bottle feeders are evaluated for safety and efficiency during feeding. If the child is having frequent emesis and/or is not gaining adequate weight, referral to a gastroenterologist may be warranted if the infant's bottle-feeding skills are judged to be adequate and there are no overt signs/symptoms of aspiration.

For older infants and toddlers, parents may report that a child gags with initial offerings of more textured solids. While this type of concern is common for this age group, more thorough oral sensory and oral motor assessment is warranted. A child may have delayed or impaired oral motor skills and is not able to safely manage a solid, thus requiring more manipulation prior to swallowing and prompting a gag response. Oral motor deficits, such as ankyloglossia, may contribute to the child's issues with oral preparation of the bolus. Reflux-related changes or upper airway obstruction may contribute to the child's sensory gag response. The SLP should observe the child's sensory responses to varying tastes, temperatures, and textures of solids presented. Though an unknown diagnosis of EoE can be present at this time, oral motor and oral sensory system delays or deficits independent of EoE should be carefully examined as contributing factors to the child's feeding difficulties [69].

Treatment

In cases of children with feeding disorders who have a mild reflux history or resolved upper airway obstruction, caregivers may be able to use learned therapeutic feeding techniques at home to help with a child's feeding progression. However, children with diagnosed eosinophilic gastrointestinal disorders will require more intensive interventions. Though reflux may occur concurrently with EoE, a child's symptoms may not significantly improve with acid-suppression therapy alone [32]. Using an amino acid-based formula or a six-food elimination, diet has been shown to be particularly effective in resolving symptoms in patients with suspected food allergies [48, 57].

Direct feeding therapy, in conjunction with medical therapy, is fundamental in helping children with both oral motor and oral sensory dysfunction and aiding in the acceptance of food reintroduction in cases of pediatric EoE [68]. A child has likely developed negative responses to foods which do not simply disappear with reduction or absence of eosinophils. In addition, children may be fearful or anxious about mealtime, and mealtime dynamics can become complicated and stressful for both the child and the family

members, affecting quality of life for the entire family [27].

Feeding therapy sessions may focus first on the child's oral hypersensitivity and slowly progress with acceptable tastes and textures of foods. Oral motor skills may then be addressed once a child's hypersensitivity improves. Focus on the overall mealtime environment is also discussed with the caregiver(s), as consistency and routine can play key roles in the success of the child's feeding progress.

Conclusion

Children with feeding difficulties require thorough evaluation by a team of specialists to determine possible sources of feeding dysfunction. In cases of eosinophilic esophagitis, even with clinical improvement in a child's symptoms, ongoing adverse responses to age-appropriate foods are common. Medical intervention, concurrent with treatment by a feeding specialist, a registered dietician, and a psychologist, is critical [68].

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Tracheoesophageal Fistula

38

Scott M. Rickert

Overview

Tracheoesophageal fistula (TE fistula or TEF) is an abnormal communication between the trachea and esophagus. While tracheoesophageal fistula can be secondary to malignancy, trauma, infection, or surgery, the vast majority are congenital in nature. It is the most common congenital abnormality of the trachea and found in 1 in 3000 live births [1]. This chapter reviews the variants of tracheoesophageal fistula, the clinical presentation of these variants, management and treatment strategies, as well as operative planning for repair.

Definitions

Tracheoesophageal fistula (TE fistula or TEF), an abnormal communication between the trachea and esophagus, has five different variations (although only four are in fact a TEF) (Fig. 38.1) [2, 3]:

1. The TE fistula is a connection between a fully formed trachea and a distal esophagus. There is an upper esophageal blind pouch. Typically, this connection occurs at the level of the distal trachea, just above the carina. This is the most common presentation and represents approximately 87% of the TE fistulas (Gross Type C, Vogt 3B).
2. The TE fistula is at one junction with a fully formed trachea and a fully formed esophagus (Gross Type E or H type). This occurs approximately 4% of the time.
3. The TE fistula is a connection of the upper esophagus with a completely unconnected distal esophagus. This occurs approximately 1% of the time (Gross Type B, Vogt 3A).
4. The TE fistula is in fact a combination of two distinct areas, an upper esophageal connection with the trachea and a slightly more distal lower esophageal connection with the trachea. There is an area of esophageal atresia between these two unconnected parts of the esophagus which connect only through the two TE fistula sites. This occurs 1% of the time (Gross Type D, Vogt 3C).
5. There is also a "TEF" variant which is not actually a TE fistula as there is no connection between the trachea and esophagus. This is a normal trachea with a mid-esophageal atresia, allowing an upper esophageal blind pouch and a lower esophageal connection to the stomach and subsequent GI tract. This occurs 8% of the time (Gross Type A, Vogt Type 2).

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Fig. 38.1 Types of esophageal atresia/tracheoesophageal fistula based on the Gross classification. (Reprinted with permission from Singh et al. [3])

Pathophysiology

For the congenital variant of TE fistula, an incomplete fusion of the tracheoesophageal fold allows for a defect in the tracheoesophageal septum [4]. This defect allows an open communication between the trachea and esophagus. Embryologically, the trachea and the esophagus both develop around 4–6 weeks from the caudal foregut. The longitudinal tracheoesophageal fold then divides the foregut into a ventral laryngotracheal tube and a dorsal esophagus. The tracheoesophageal fold fusion occurs around the 4th–6th week of embryonic development. The location of the incomplete fusion is where the TE fistula forms, hence the variability in presentation of TE fistulas.

There are several syndromes that are predisposed to incomplete fusion of the tracheoesophageal ridges and form TE fistulas. These include trisomy 13, 18, and 21 as well as VACTERL syndrome (Vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) [1, 5, 6]. Other congenital anomalies are noted to have an association with TE fistulas or esophageal atresia. These include children with muscular/skeletal anomalies, kidney or genitourinary issues, gastrointestinal tract issues (imperforate anus, congenital diaphragmatic hernia), or heart deficits (tetralogy of Fallot, ventricular septal defect) [7, 8]. Also notable is that some patients with TE fistulas will also present with laryngeal cleft and may have further dysphagia or aspiration secondary to the cleft [9].

For the acquired variant of TE fistula, presentation can be equally variable as the cause can have a significant impaction on presentation. Typical causes for an acquired TE fistula include malignancy, trauma to the chest and neck, prolonged intubation in conjunction with a nasogastric/orogastric tube in place, infection (tuberculosis is most common), or ruptured diverticula [10–12]. Location is dependent on the area of disease or trauma.

Presentation

Patients typically present at birth or near birth with symptoms, including respiratory distress. The most common TE fistula, one in which the upper esophagus is a blind pouch, fills with food and saliva at first. Once full, liquids cannot pass to the stomach as it is a blind pouch and is regurgitated into the pharynx and aspirated into the airway, causing coughing, choking, and respiratory distress. This may lead to significant desaturations and result in a "blue baby" in moments of distress. Other cases may be more subtle in initial presentation but cause significant respiratory distress when the oral contents are aspirated.

Physical exam and imaging can be particularly helpful in diagnosis of a TE fistula. Passage of a small nasogastric tube into the esophagus in conjunction with an x-ray can determine the distal end of the esophagus which is particularly helpful in cases of proximal esophageal atresia/blind pouch or TE fistula. General x-rays of the chest and abdomen can be helpful in determining air column patterns in patients with concern for esophageal atresia. TE fistula can be further evaluated with cross-sectional imaging, including

magnetic resonance imaging (MRI) or computed tomography (CT) [13], which will help clarify the location of the esophageal atresia and TE fistula and the nature of the fistula. CT has disadvantages of radiation exposure, and thus consideration of MRI instead is warranted depending on the clinical scenario. In general, Gastrografin swallow studies should not be performed on patients with suspected TE fistulas as it may predispose patients to chest infections or allergic reactions. While MRI or CT with thin cuts remains the mainstay of anatomic imaging and surgical planning, a diagnostic bronchoscopy and endoscopy helps to localize the TE fistula most accurately. There are some TE fistulas which are challenging to assess within the mucosal folds of the trachea/bronchus or esophagus. In these cases, during bronchoscopy and endoscopy, methylene blue dye can be placed in the esophagus via endoscopy, and the trachea can be visualized via bronchoscopy to see if there is any extravasation of dye within the airway. This helps to confirm the communication as well as identify its location.

Otolaryngologist Approach

History

Typical presentation of a TE fistula occurs shortly after birth, as children with TE fistula present with labored breathing, choking, and coughing. The pregnancy history should be elicited as polyhydramnios is a risk factor for TE fistula. Family history as well as other genetic syndromes such as trisomy 13, 18, and 21 or VACTERL may be picked up prenatally as well, and those patients should be monitored closely for possible TE fistula.

If there is any concern for a possible TE fistula in utero, a prenatal ultrasound and a prenatal MRI can be performed to better understand the area of concern.

Once born, the baby should be carefully assessed for signs of labored breathing, choking, and coughing. If they are unable to swallow their secretions, a nasogastric tube should be carefully placed to see if there is patency of the esophagus.

Exam

A general assessment of the patient and a head and neck exam are performed. From the general assessment, the patient should be observed to see if there are any signs of choking or coughing with their secretions that may represent aspiration or esophageal atresia. A full round abdomen may be a clinical sign of TE fistula as well. In addition, an endoscopic exam can be useful to assess vocal fold function (before any surgical intervention) as well as any other anatomic abnormalities within the upper airway (e.g., laryngeal cleft).

Imaging

Imaging of the chest and upper airway is particularly important in localizing a TE fistula. Prenatal ultrasound and MRI can be useful in those cases with high suspicion prenatally. Chest and abdomen x-ray are useful as first-line imaging studies, particularly when a nasogastric tube is placed to evaluate for esophageal patency versus atresia. Thin-cut CT or MRI can be used to better delineate the area of concern prior to any surgical intervention. If the diagnosis is in question and the patient is taking orally, videofluoroscopy with water-soluble contrast may help visualize a fistulous tract (Fig. 38.2).

Instrumented Assessment

Incomplete passage of a nasogastric tube can be helpful to determine esophageal patency. Definitive assessment with a preoperative diagnostic bronchoscopy and endoscopy is a mainstay of treatment. Methylene blue can be used to aid the assessment when direct visualization is challenging on endoscopy [14].

Management

Management of a TE fistula requires preoperative management, surgical management, and proper postoperative surveillance. Proper preoperative

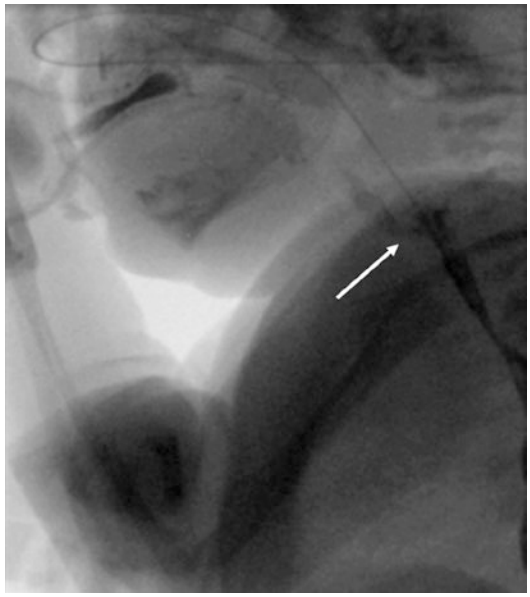

Fig. 38.2 Videofluoroscopic image demonstrating fistulous tract from the esophagus to the trachea. (Courtesy of J. Scott McMurray, MD)

assessment with imaging and bronchoscopy/endoscopy is essential to understand the nature and type of TE fistula as well as to assess for any other concomitant lesions such as laryngeal cleft. Preoperative management of the patient's general health is paramount as they are more prone to recurrent pneumonias secondary to aspiration events as well as poor nutrition secondary to the nature of the TE fistula. Once their nutritional and respiratory status are stabilized, successful surgical repair can be properly performed.

Operative Approach: Endoscopic and Open Repair of Tracheoesophageal Fistula

Indications

Surgical intervention is indicated for patients with TE fistula as the disorder adversely affects patient quality of life with concern for recurrent pneumonias, inability to tolerate secretions, and poor nutrition.

Endoscopic repair is indicated for smaller or more localized TE fistula and can be done via localized biological glue, laser/cauterization, or a combination of the two. Open repair is reserved for larger TE fistulas, TE fistulas which are difficult to expose endoscopically, or previously failed endoscopic repair.

Key Aspects of the Consent Process

Risks associated with bronchoscopy, endoscopy, and TE fistula repair should be discussed, including injury to the lips, gums, tongue, and teeth, as well as potential for transient or longer-lasting dysgeusia from the bronchoscopy/endoscopy. Furthermore, TE fistula repair can result in damage to the recurrent laryngeal nerves, causing vocal fold paresis or paralysis. This may lead to a poor voice, poor coordination in swallowing, and aspiration. Operative repair of the esophagus may lead to strictures at the area of repair, potentially causing prolonged dysphagia and need for further interventions on the stricture (dilation, additional open repair). Operative repair on the trachea may lead to stenosis or narrowing within the airway and predispose them to airway compromise, further recurrent pneumonias, and need for future surgical interventions on the airway (dilation, additional open repair). Poor healing may result in recurrence of the fistula and subsequent infections, including pneumonia or mediastinitis.

Equipment

A flexible bronchoscope and a gastrointestinal endoscope are used with their individualized endoscopic towers to visualize the areas of concern. If methylene blue is needed, no more than 1 cc can be placed through the biopsy port of the endoscope or bronchoscope during the procedure.

For definitive endoscopic repair, a rigid 3.5 bronchoscope can be used to evaluate the trachea, while an appropriately sized gastrointestinal endoscope is used to visualize the esophagus.

For definitive open repair, a rigid 3.5 bronchoscope can be used to provide endoscopic

visualization while the open repair is being performed. A pediatric open head and neck set has all the basic equipment needed for open repair.

Endoscopic Repair Steps

1. *Patient positioning*. The patient is placed supine, and a bite block is placed to prevent damage to the lips and teeth as well as damage to the endoscopic equipment. Anesthesia can be maintained by an inhalational anesthetic through a nasal approach or by total intravenous anesthesia (TIVA) with spontaneous ventilation and oxygenation via a nasal insufflation technique or via orotracheal intubation if necessary.
2. *Endoscopic exposure.* Rigid and flexible bronchoscopes and esophagoscopes can be used to identify and assess the fistula (Fig. 38.3). Once the fistula is exposed, a small flexible suction (4 or 5 Fr) can be placed in the area to see the extent of the fistula.
3. *Cauterization/application of biological glue to TE fistula.* Once it is fully identified, a Bugbee cautery can be placed through the biopsy port of the bronchoscope and passed within the fistula. Using a low-powered setting of 2–4, it can be intermittently activated to attempt to seal the TE fistula endoscopically with cautery (Fig. 38.4a–d). It is important to stay within the fistula only and not use the cautery within the lumen of the esophagus or trachea to minimize the incidence of synechia or stenosis. Once the Bugbee cautery is used, it is carefully withdrawn into the bronchoscope. Once this is complete, Prolaryn or other augmentative injectable can be injected just lateral to the fistula to encourage coaptation of the cauterized edges (Fig. 38.4f). Additionally, a small amount of biological glue (e.g., Tisseel) can be placed through a small suction tubing in a similar fashion through the bronchoscope into the fistula. If the decision is made to perform the closure with biological glue solely, then this can be done through the bronchoscope as the first step without performing any cautery.

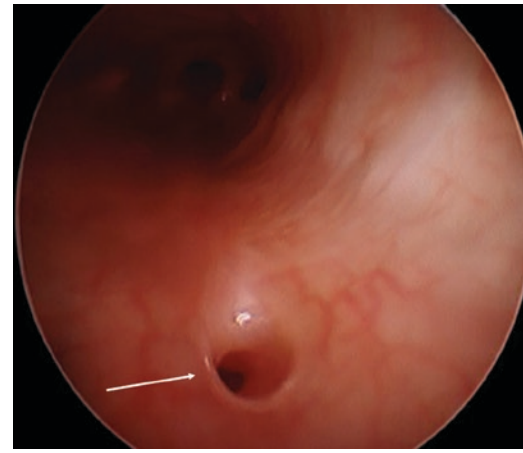

Fig. 38.3 Intraoperative image obtained during rigid bronchoscopy demonstrating opening of fistulous tract into the trachea (white arrow). (Courtesy of J. Scott McMurray, MD)

4. *Optional alternative technique: endoscopic clip placement.* Application of an endoscopic clip can be used in select cases to physically close the fistula [15, 16]. This is usually attempted through the esophageal exposure and placed along the edge of the esophageal outpouching to pinch the TE fistula closed. Possible dislodgement of the clip may lead to the need for further intervention or other attempts at repair.
5. *Optional alternative technique: endoscopic stenting.* Although this is primarily a technique used in adults, endoscopic placement of stents can be used to seal off a fistula effectively when the option is viable [17]. Stents are typically made of either membrane-covered metal alloy or silicone, and preference and use are very individualized. Stent design can be based on the endoscopic exam and imaging. 3D-printed stents based on the 3D image reconstruction of the airway are a newer design technique but provide a very individualized approach to tailoring the stents appropriately.
   (a) *Esophageal stenting.* Esophageal stenting alone is a viable option and is a good option for those without concomitant airway stenosis. Once the fistula is adequately assessed, the stent length and

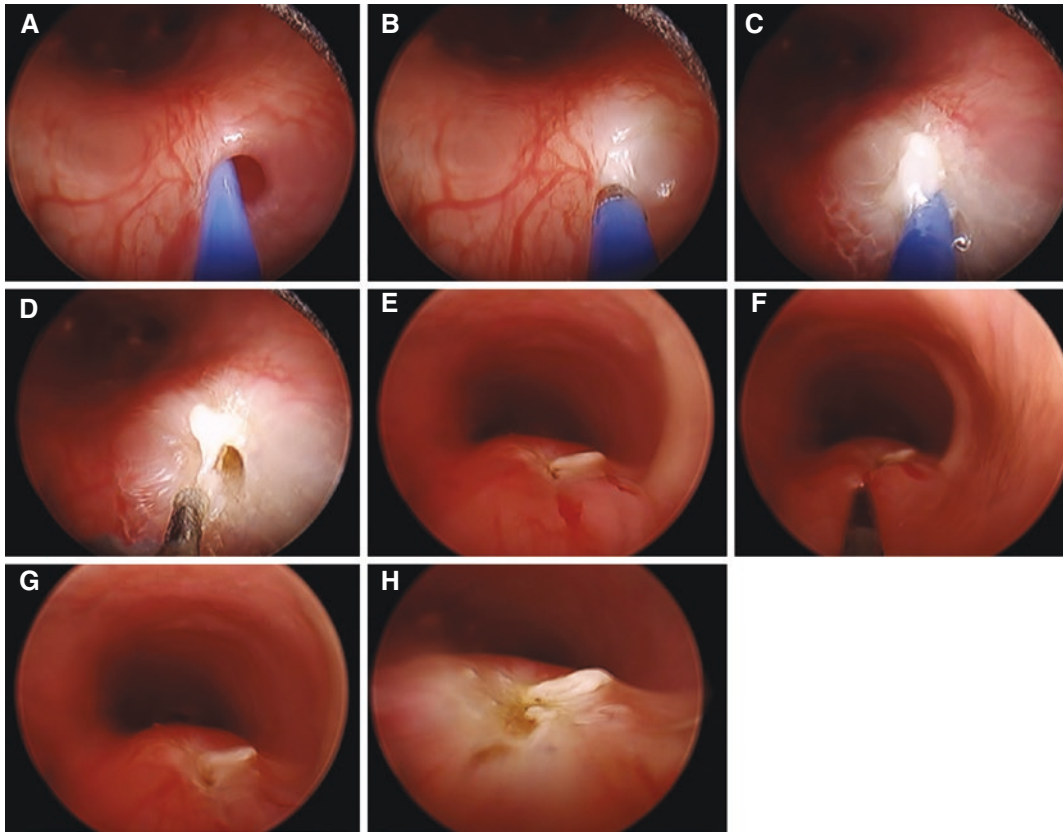

Fig. 38.4 Series of intraoperative images demonstrating endoscopic repair of a tracheoesophageal fistula using the Bugbee cautery method. (**a**–**d**) Bugbee cautery is inserted into the tract and used to cauterize the fistula. Care is taken not to violate the adjacent mucosa. (**e**) Visualization of tracheal opening into the tract after cautery has been performed. (**f**) Prolaryn is injected adjacent to the tract opening to encourage coaptation of the cauterized edges. (**g, h**) Tracheal opening of the fistula at the end of procedure. (Courtesy of J. Scott McMurray, MD)

diameter can be determined to cover the entire length of the fistula. The stent is placed endoscopically using basic endoscopic principles. When placed, it should cover the fistula site and press firmly against the esophageal wall. There is a risk of displacement of the stent which would then lead to the need for further intervention and repair [18, 19].

(b) *Airway stenting.* Airway stenting alone is a viable option in difficult cases when an esophageal stent cannot be placed. While rare in children, the selection and design of the stent are similar to the esophageal stent. The stent needs to fully cover the airway side of the TE fistula while having sufficient diameter to press firmly against the wall. Airway stents are typically longer in length than the fistula itself and are carefully designed to prevent displacement as true airway compromise can occur from stent displacement. Stenosis at the site of the airway stent can also occur, and possible need for localized endoscopic intervention may arise if stenosis causes airway compromise or recurrent pulmonary infections.

(c) *Esophageal and airway stenting.* A combination of airway and esophageal stenting can also be performed [20]. Dual stenting is particularly helpful in prevention of stent migration or displacement as they keep each other in place from their expansion in the airway/esophagus. If

dual stents are placed, it is important to place the airway stent first to prevent possible airway stenosis secondary to the esophageal stent. After the airway stent is confirmed to be in the proper position, the esophageal stent can be placed.

Open Repair Steps

1. *Patient positioning.* Once the TE fistula is identified on initial assessment, the patient is placed supine, and anesthesia can be maintained via orotracheal intubation. The patient is prepped and draped for a planned open repair and neck incision. The location and length of the incision depend on the location of the TE fistula and planned technique of repair.
2. *Incision options.* Depending on the location of the TE fistula, there are a variety of incision sites that could be appropriate. Cervical collar incision is the mainstay approach although it may be modified based on the location of the fistula. Upper sternotomy may be utilized in conjunction with a cervical incision if needed for exposure. For lower TE fistulas, a median sternotomy or thoracotomy may be performed.
3. *Surgical approach.* After a cervical collar incision is made (with or without a sternal split), subplatysmal flaps are raised and the strap muscles are lateralized. Typically, the thyroid isthmus is divided and the thyroid is lateralized. The pretracheal fascia is dissected and the trachea is exposed. The lateral aspects of the trachea are exposed with careful attention to the locations of the recurrent laryngeal nerve in the tracheoesophageal groove. By staying directly on the trachea, the nerve is less likely to have any stretch injury.
4. *Operative management of the trachea.* The trachea is exposed only around the area of the fistula to avoid devascularization. The level of the dissection and fistula is confirmed by endoscopic exam. Dissection is continued along the fistula in the plane between the trachea and esophagus, and the abnormal trachea is resected as either a formal tracheal resection (remove entire rings) or a localized resection with reconstruction (similar to a laryngotracheal reconstruction). In a formal tracheal resection, the trachea is typically reconstructed by end-to-end anastomosis. In a more localized resection, the reconstruction follows the basic principles of a laryngotracheal reconstruction with possible rib grafting for further cartilaginous support of the resected area and reconstruction.
5. *Operative management of the esophagus.* Once the TE fistula is exposed and the tracheal defect is addressed, the esophageal defect is addressed by full dissection in the area and closed in at least two separate layers. A nasogastric tube is placed to ensure the suture line does not narrow the esophagus and limit the risk for postoperative strictures.
6. *Closure with vascularized tissue.* Once both areas of concern at the site of the fistula are addressed, pedicled vascularized tissue is locally mobilized (typically from the strap muscles) and placed between the esophageal and tracheal repair to ensure better healing and prevention of localized complications such as a tracheo-innominate fistula. A laryngeal release may be needed to prevent excessive tension on the tracheal or esophageal anastomosis. A flat drain is placed in the subplatysmal plane to prevent hematoma. A chin suture may also be used to help prevent excessive tension on the repairs.
7. *Complications.* As this is complex surgery involving the esophagus and trachea, there are many possible complications. Poor division of the TE fistula may result in reformation of the fistula and subsequent pneumonia and mediastinitis. Localized scarring in the airway may cause tracheal stenosis and airway compromise with potential need for further repair. Localized scarring in the esophagus may cause dysphagia and poor esophageal motility due to esophageal strictures. Aggressive dissection around the trachea may cause damage to the recurrent laryngeal nerves. More than half of repaired patients are diagnosed with reflux and require anti-reflux treatment.

Postoperative Management and Follow-Up

Close follow-up and surveillance bronchoscopy and endoscopy are necessary to ensure good healing. Follow-up swallow studies help with evaluation of swallowing, and working closely with the pulmonary team ensures that the lung exam is closely followed for any signs of recurrent pneumonia or aspiration. Flexible videostroboscopy can be used in patients with suspected vocal fold immobility secondary to aggressive dissection around the trachea and possible recurrent laryngeal nerve injury.

Emerging and Evolving Techniques of the Future

Endoscopic Approaches to Tracheoesophageal Fistula

Endoscopic approaches for tracheoesophageal are desirable, eliminating the need for an external neck incision and also limiting morbidity associated with extensive dissection and reconstruction. However, it can be difficult to identify the fistula endoscopically and achieve adequate exposure for repair. A variety of approaches have been described, as discussed in this chapter, though large comparative series are lacking, in part secondary to the rarity of the disorder [19]. Determining how to select the appropriate procedure for each patient will be the focus of ongoing clinical investigation.

Endoscopic Stenting

Although this is an evolving technology, the use of endoscopic stenting has been a helpful adjunct to traditional methods for addressing TE fistulae in adults. Further investigation is required to determine which pediatric patients may be appropriate candidates for this approach.

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39 Post-intubation Glottic Insufficiency

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Introduction

Posterior glottic insufficiency (PGI), also termed posterior glottic diastasis, is a triangular or keyhole aperture between the vocal processes and the posterior laryngeal wall [1, 2]. The pathological finding is the result of inadequate arytenoid closure and apposition, often arising from one or more of three factors: loss of normal interarytenoid mucosal soft tissue, pressure necrosis of the arytenoid, and medial cricoarytenoid joint injury [2–4]. These injuries can be a result of long-term indwelling endotracheal tubes [5]. Because the corniculate regions and the upper arytenoids tilt forward to compensate for PGI, patient voices often sound worse than would be expected based on laryngoscopy alone. This anatomical compensation impairs complete visualization of PGI, and thus the pathology has been historically underdiagnosed [2]. Posterior glottic diastasis is often seen after laryngotracheal reconstruction where in a posterior graft can cause separation of the cricarytenoid joints. Posterior glottic stenosis is another sequelae of prolonged intubation, however, it is outside the scope of this chapter.

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Factors Contributing to Laryngeal Intubation Trauma

There are several identified factors that contribute to laryngeal intubation trauma. The severity of intubation injury is most strongly influenced by two factors: the duration of intubation and the endotracheal tube's physical characteristics. The following predisposing factors must be considered in the assessment of not only potential PGI but also other forms of laryngeal trauma.

Duration of Intubation

A longer duration of intubation increases the likelihood of damage [6]. Although there are little data, the time of intubation before risk of irreversible damage is likely longer in infants than in adults. With skilled neonatal ICU care, intubation can extend for several weeks with a low incidence of problems [7].

In one study, six of seven patients with PGI had multiple or prolonged intubations. Two of the seven patients were intubated for surgical procedures [1]. Because laryngeal injuries and

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ulcerations can develop as early as 3 hours after intubation, it is important to consider PGI in the differential even if the patient was only intubated for surgical procedures [8].

Characteristics of the Endotracheal Tube

The external diameter, shape, firmness, and composition of the endotracheal tube all shape the pathogenesis of laryngeal trauma. In practice, tube size should be chosen for each individual patient. For men, the endotracheal tube external diameter should be approximately 8 mm, whereas the diameter should be no larger than 6 mm for women. Standard practice indicates that infants and children up to 8 years of age should use a low-pressure cuff and a tube with a diameter that allows an air leak in the subglottic space with approximately 20 cm of water ventilation pressure under ideal circumstances [7].

Difficult or Complicated Intubations

Difficult or complicated intubations include those performed in an emergency, conducted by an unskilled person, following repeated intubations, associated with improper use of an introducer, or involving abnormal laryngeal anatomy. Specifically, the tube pressure and friction aggravate existing injuries and cause new trauma. Such movements include not only those conducted by healthcare workers but also coughing, swallowing, transmitted ventilator movements, and movements during suctioning [9, 10].

Bacterial Infection

Bacterial colonization increases during intubation, which can be further complicated by the formation of a biofilm; 4 days is a critical period [11]. In patients with a tracheotomy, bacterial infections occur quickly, especially in immunodeficient patients or those relatively immunosuppressed due to stress. When a tracheotomy is performed in the setting of prior laryngeal intubation trauma, contamination of the tracheotomy stoma can prolong laryngeal healing and influence scar formation [12].

Gastroesophageal Reflux

Many patients who have sustained posterior glottic injury from intubation also have underlying gastroesophageal reflux [13]. Spillover and aspiration of acid into the larynx and subglottic region aggravate existing injuries and increase the risk of infection, granulation tissue, and ulceration [14]. Experiments on the canine larynx found that intermittent reflux can severely damage the larynx if there is prior mucosal injury, even if the pH of the refluxate is 4.0 [13]. Furthermore, reflux is more likely with a nasogastric tube in place, which irritates the lower esophagus and can cause pressure necrosis and ulceration in the post-cricoid region [15].

Acute/Chronic Diseased States

Conditions that cause poor tissue perfusion are all associated with more severe changes. These include, but are not limited to, malnutrition, diabetes mellitus, dehydration, hypoxemia, anemia, hypotensive episodes, and liver, heart, or kidney failure. Decreased capillary perfusion leads to an increased risk of necrosis and ulceration, thereby increasing the risk for not only PGI but also other forms of laryngeal damage [7].

Pathogenesis of Pressure-Induced Injury

Endotracheal tubes lie in the posterior larynx. Intubation therefore results in pressure on the following structures: the mucous membrane and mucoperichondrium covering the medial surfaces of the arytenoid cartilages and their vocal pro-

cesses, the cricoarytenoid joints and cricoid cartilage, and the posterior glottic and interarytenoid region. When pressure from the endotracheal tube exceeds capillary pressure, the microcirculation in the mucosa and mucoperichondrium becomes interrupted. The resultant pathology is broken into a sequence of fundamental lesions: edema, ulceration, granulation tissue, ulcerated troughs, and chronic healed furrows. Other miscellaneous gross injuries without a diagnostic category may occur, particularly in difficult or complicated intubations. Uncontrolled, the fundamental lesions lead to chronic injuries, such as PGI. Identifying the sequelae of pressure-induced injury provides an accurate prognosis, and a more confident decision can be made regarding whether to continue intubation or to move to tracheostomy [7].

Edema

Edema may manifest as protrusion of the edematous ventricular mucosa within minutes to hours. Swelling of the supraglottis and glottis is typically nonspecific and resolves quickly post-extubation and does not indicate future morbidity [7]. Protrusion of the edematous ventricular mucosa should not be confused with prolapse of the ventricles, which is a separate pathological finding involving varying degrees of metaplasia, reserve cell hyperplasia, edema, inflammation, glandular atrophy, and fatty infiltration [16].

Ulceration

Superficial ulceration can begin within hours of intubation. It involves the mucosa and typically heals without scarring post-extubation. Continued pressure from the endotracheal tube, however, results in deep ulceration into the perichondrium or cartilage, which carries the implication of delayed healing. Characteristics of post-extubation recovery include healing by second intention and fibrosis, vascular damage, hyperpermeability, migration of inflammatory cells, and possible future formation of scarring or squamous metaplasia [7, 15, 17]. As the ulceration further exposes cartilage, frank cartilage necrosis may occur, resulting in weakening, disintegration, and deformation of the cartilaginous framework. Subsequently, chronic phase lymphocytes and macrophages may accumulate at the necrotic area. Fibroblasts activate and scar tissue begins to form [7]. Deep ulceration involving the cricoarytenoid joints likely leads to airway or voice defects [15].

Granulation Tissue

Granulation tissue, which may arise within 48 hours of intubation, is attempted healing at sites of irritation and ulceration (Fig. 39.1) [15]. Granulation tissue at the vocal process is often the first pathology to be identified, but whether superficial ulceration preceded its development is difficult to determine. With removal of the endotracheal tube, flaps of granulation tissue can be seen, almost encircling the anterior surface of the tube on each side. This also forms under the posterolateral aspect of the endotracheal tube, but because they are compressed into

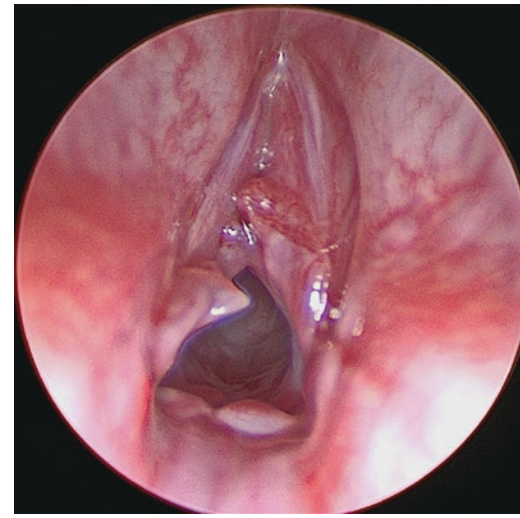

Fig. 39.1 Glottic granulation tissue

the posterior glottis, they can only be visualized post-extubation.

Because the flaps move with inspiration and expiration, they may cause inspiratory obstruction. Removal of the flaps may be beneficial, at least from one side, thereby possibly improving the airway and the prospect of extubation. Removal from both sides is not recommended, as the two raw surfaces may form an interarytenoid synechia, adhering to each other across the midline and joining the vocal folds. Others have suggested removal at the time of tracheostomy, should extubation have failed [18]. Removal of the flaps is usually unnecessary, however, because most cases completely resolve after extubation. Should the granulation tissue not heal properly, scarring (covered by mucous membrane) or a reddish-yellow intubation granuloma may form on the medial edge of the vocal process of the arytenoid cartilage [7].

Ulcerated Trough and Chronic Healed Furrows

The originally superficial ulcerations may become deeper and wider. These ulcerations may then expose the cartilaginous medial surface of the arytenoid and cricoid cartilages and sometimes the cricoarytenoid joints. Such deep ulcerations are referred to as ulcerated troughs, which are visible only after removal of the endotracheal tube [7]. At the margins of the ulcerated trough, granulation tissue proliferates. Post-extubation, the healing and fibrosis that take place weeks to months later eventually replace the trough with a chronic healed furrow (Fig. 39.2). Both the ulcerated trough and the chronic healed furrow suggest dysfunction of the cricoarytenoid joints, thereby causing chronic voice problems (including PGI) [7].

Miscellaneous Injuries

These include, but are not limited to, damage to muscles [19], perforation of the airway, and laceration of the true or false vocal folds. These injuries are more likely to result from difficult intubations and may be caused by either the endotracheal tube or the inducer. Perforation of the airway may lead to spreading surgical emphysema and infection into the soft tissues of the neck or mediastinum. Acute lacerations typically heal but may leave a small scar permanently. Without complete healing, a granuloma may form [15].

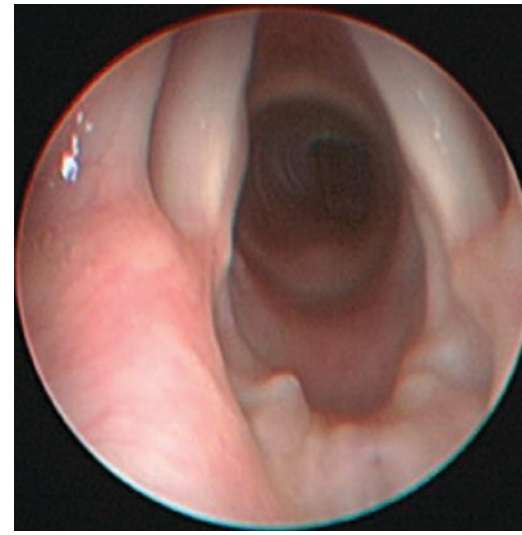

Fig. 39.2 Ulcerated trough and subsequent chronic healed furrow in the interarytenoid space

Speech-Language Pathologist Approach

Posterior glottic insufficiency (PGI) results in a posterior glottic gap that results in dysphonia. The dysphonia is characterized by breathiness due to air escape, asthenia due to poor glottic closure, and roughness due to aperiodic vibrations. The management of PGI is important for a child's educational and psychosocial development, as well as physical and emotional health [20]. While current surgical intervention is being investigated to manage PGI, voice therapy can also be beneficial. Voice therapy techniques specifically to

treat PGI have not been thoroughly explored. Of equal importance is the detailed preoperative and postoperative assessment conducted by a speech and language pathologist to provide invaluable diagnostic information and possible treatment options [21].

Treatment

Beginning treatment with a child with PGI may first involve optimizing voicing technique. It has been our experience that over time of using a chronic weak, breathy, rough voice, the child may develop poor voicing techniques as they are accustomed to having a voice that does not meet their daily functions (projection, quality, etc.). This may include reduced breath support, an increase in pitch to aid in vocal fold closure, rapid rate of speech to produce as many words on one breath as possible, and poor overall effort. The opposite may also occur where the child becomes hyperfunctional to compensate for a dysphonic voice. Increasing breath support, implementing elements of Conversational Training Therapy (CTT), and promoting vocal hygiene to reduce laryngeal irritants can optimize voice production in children with PGI [22]. Specifically within CTT, the use of "clear speech" facilitates a reduction in speaking rate, an increase in amplitude and prosody ranges and phrase and speech sound lengths, and more precise vowel productions [22]. To accomplish these common voice therapy goals, an increase in breath support is necessary and indirectly gained.

Additionally, use of vocal fold adduction exercises, resonant voice therapy, inhalation phonation, and vocal function exercises have shown to assist in maximizing vocal fold closure. Adduction exercises must be closely monitored to prevent development of hyperfunction. Optimizing vocal fold contact through resonant voice therapy and increasing frontal tone focus increase resonant projection [23]. Inhalation phonation can be useful when aphonia, abnormal methods of vocal fold phonation (i.e., ventricular phonation), or hyperfunction is present [24]. This maneuver approximates true vocal fold adduction, activates true vocal fold vibration, relaxes the ventricles, and stretches the vocal folds [24]. Vocal function exercises are a systematic procedure designed to strengthen and coordinate laryngeal musculature, increase vocal fold adduction, and improve balance to the vocal mechanism [25].

Summary

Evidence-based treatment options specifically for PGI are scarce. More research is necessary for this particular patient population. Stimulability testing for the particular patient is vital to developing the most appropriate treatment plan. Overall, the above techniques are beneficial in optimizing vocal production and encouraging vocal fold adduction.

Otolaryngologist Approach

Traditionally, PGI was treated by injection laryngoplasty to improve vocal fold approximation; the treatment was ultimately found to be effective mainly for anterior gaps, as opposed to the posterior gaps seen in PGI [26, 27]. Aryepiglottic fold flaps and endoscopic posterior cricoid reduction laryngoplasty (EPCRL) are newer techniques that have been shown to produce better outcomes [2]. In one retrospective study, six of seven patients (11 months–20 years) presenting to pediatric otolaryngologists/laryngologists for PGI had minimal or no improvement with injection laryngoplasty. In comparison, three of three patients (15–20 years) who had undergone EPCRL had significant improvement in voice function without any resulting dyspnea or stridor [1]. EPCRL, however, involves removal of a segment of the posterior cricoid cartilage, thereby narrowing the airway. Patients that are at risk of airway obstruction should be reevaluated when they are older, or other options should be considered [1].

Another surgical option is grafting of buccal mucosa into the damaged interarytenoid space.

Although the operation has exhibited strong outcomes for adults, results have not yet been reported for children. Regardless of the extent of injury, it is recommended that the graft encompass the entire posterior glottis. This is partially because the graft is thicker than the normal posterior glottic mucosa. Even for unilateral injuries, the resulting graft provides better outcomes, including greater posterior glottic obstruction, better airflow through the vocal folds, and a stronger voice [28].

Potential Chronic Complications

In addition to posterior glottic insufficiency, the otolaryngologist should be aware of other potential chronic complications which may arise and can impact patient voice and airway patency.

Complete Obstruction

Obliteration of the glottic or subglottic lumen may occur in advanced cases of intubation trauma. This can be caused by poorly judged repeated attempts to dilate the lumen or from excessive laser surgery that worsened existing injury from prolonged intubation [15].

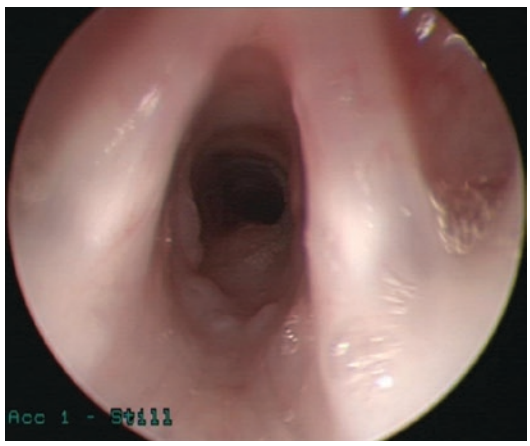

Fig. 39.3 Retention cysts in the posterior glottis

Ductal Retention Cysts

Ductal retention cysts are accumulations of mucus in obstructed and subsequently dilated ducts of submucosal mucous glands, as opposed to distension of the glands themselves (Fig. 39.3). They are commonly seen in infants after days or weeks of intubation and usually coexist with other complications of intubation trauma [29]. Ductal retention cysts often do not require treatment, especially if they are small. Larger cysts may be associated with airway obstruction. In such cases, laser-based removal is recommended. Recurrence is rare [15].

Dislocation, Fixation, or Subluxation of Arytenoid Cartilage

Trauma to the arytenoid cartilages is more common in difficult or complicated intubations. The left arytenoid cartilage is at higher risk for damage because the endotracheal tube is usually angled through the right side of the mouth and directed down to the left during intubation [15]. The patient may experience hoarseness, throat discomfort, difficulty swallowing, painful swallowing, cough, and difficulty breathing. Indirect laryngoscopy shows displacement, swelling, asymmetry, and reduced or absent movement of the affected side.

A fixed or dislocated arytenoid causing serious airway obstruction should be evaluated for arytenoidectomy, whether endoscopic or open via a laryngofissure. Though presentation is often delayed, treatment by microlaryngoscopy and attempted closed reduction to relocate the arytenoid cartilage is more likely successful when conducted earlier [7].

Interarytenoid Adhesion

Interarytenoid adhesion, or interarytenoid synechia, or type 1 posterior glottic stenosis, occurs when flaps of granulation tissue fuse together (Fig. 39.4). Patients experience stridor and difficulty breathing, whereas the voice typically

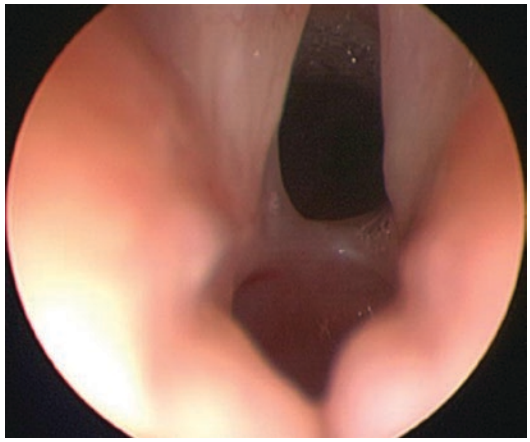

Fig. 39.4 Interarytenoid adhesion

remains functional. If identified within the first few weeks, interarytenoid adhesion can easily be divided. If left untreated, the fibrous scar becomes more difficult to separate, and scissors or a laser must be used. Results at any time of treatment tend to be uncomplicated, and recurrence is unlikely [30].

Cricoarytenoid Joint Immobility

Cricoarytenoid joint mobility must be assessed with direct laryngoscopy under general anesthesia. Loss of mobility may be obscured by the presence of interarytenoid adhesion, posterior glottic stenosis, or healed furrows. Some individuals may consider endoscopic arytenoidectomy or vocal fold medialization for treatment [30].

Assessment of Laryngeal Injury Associated with Prolonged Intubation

Proper assessment of intubation-related laryngeal injury is best performed using a rigid telescope during direct laryngoscopy under general anesthesia. The presence of an endotracheal tube prevents complete visualization of the posterior glottis and thus should be temporarily removed to facilitate evaluation [31].

Preparation for Extubation and Consideration of Tracheotomy

No "safe" duration of intubation has been established, but Benjamin et al. claim that 5–7 days after intubation is a reasonable time to consider changing to tracheotomy for adults [7]. When extubation was unsuccessful in the ICU or will be attempted in the setting of moderate laryngeal edema, granulation tissue, or ulceration, an endotracheal tube of smaller diameter can be placed for 24–48 hours. In considering this intervention, it is imperative that ulcerated troughs (deep ulcerations) indicating perichondritis are absent. If the tube is removed at this stage and the trachea remains undisturbed by further intubation, the changes can be expected to resolve without any treatment under ideal conditions. As such, chronic complications such as PGI can be avoided. Although the use of steroids is controversial and not proven to be beneficial, they are used regularly by many ICU physicians and laryngologists [15].

Compared to the firmer cricoid cartilage of adults and older children, the immature cricoid cartilage ring is more compliant with the pressure of an endotracheal tube. Therefore, long-term endotracheal intubation is fairly well-tolerated for children under 3 months of age, and risk for long-term complications is lower [32].

For older children, tracheotomy should be considered if any of the following occur: ulcerated troughs through the perichondrium into the cartilage of the arytenoid, the cartilage of the cricoid, or the cricoarytenoid joint, or diffuse and concentric ulceration in the subglottic region. In such cases, chronic intubation injuries may arise [32].

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Dysphonia After Laryngotracheal Reconstruction

40

Mathieu Bergeron, Lyndy J. Wilcox, and Alessandro de Alarcon

Overview

The main objective of airway reconstruction is to establish a patent laryngeal airway without the need of a tracheostomy tube as well as being able to support respiration, airway protection, and voicing [1]. To achieve these goals, several surgical techniques may be used to either expand the airway diameter, remove the stenosed segment, or slide the airway. The most common airway expansion procedure is laryngotracheoplasty (LTP) with anterior and/or posterior costal cartilage grafts, while the most common resection procedure is cricotracheal resection (CTR). Slide tracheoplasty improves the airway diameter by sliding one segment of the airway onto the other, effectively doubling the size of the slid section. Each of these procedures has its advantages and disadvantages regarding surgical correction of the stenotic airway. Similarly, these techniques, as well as the initial airway injury, impact voice outcome to different degrees.

Childhood dysphonia is associated with social withdrawal and depression and has negative influences on emotional, educational, and occupational outcomes. Moreover, it has been reported that teachers have a negative bias toward adolescents who have a voice disorder [2]. Ultimately, voice disorders may lead to psychosocial problems that will affect patients over their lifetime, specifically their career choices and their long-term quality of life [3]. Once a patent airway has been secured, efforts should be made to improve patient's voice and overall quality of life as it can have significant effects on their well-being.

Patients with airway disorders represent a unique subset of patients. The majority of them have undergone numerous hospitalizations, were premature, and are typically involved in multiple complex medical and surgical interventions. When caring for pediatric airway patients, consideration of additional interventions should be individualized, which requires a thorough and complete workup. Voice evaluation provides further crucial, detailed functional information that can specifically direct the management of ongoing airway problems and voice problems.

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Definitions

Supraglottic Phonation

Supraglottic phonation implies the patient is voicing by means of supraglottic rather than glottic vibration [4]. The source of vibration from the supraglottic tissues may vary across patients and may include any of the following structures or combination of structures: the ventricular folds, the aryepiglottic folds with or without the petiole, and the interarytenoid mucosa. The sound generated from these structures results in a distinct, perceptually low-pitched, rough voice quality. Patterns of supraglottic compression are typically documented as either lateral-medial (medial movement of the ventricular folds during true vocal fold phonation) or anterior-posterior (anterior movement of the arytenoid cartilages toward the petiole) [4] (Fig. 40.1).

Posterior Glottic Diastasis

Posterior glottic diastasis is suspected in patients with a history of airway expansion or prolonged intubation who present with breathy dysphonia. Rigid endoscopy is performed to confirm the presence of a broad posterior cricoid plate and interarytenoid space contributing to a persistent posterior keyhole aperture (Fig. 40.2).

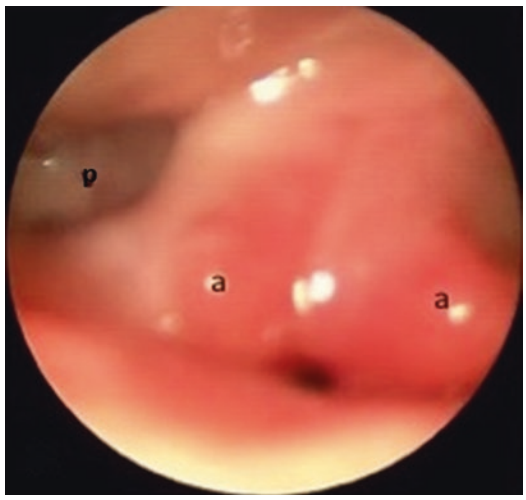

Fig. 40.1 Supraglottic collapse as noted during a microlaryngoscopy (a arytenoids, p pyriform sinus)

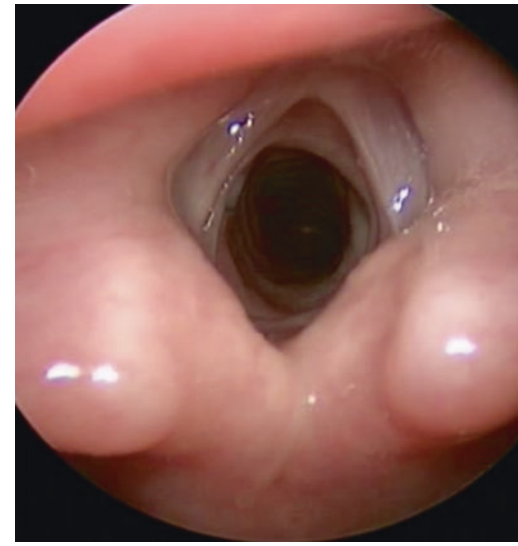

Fig. 40.2 Posterior glottic diastasis seen on microlaryngoscopy

Laryngotracheoplasty (LTP)

LTP is a surgical procedure to expand the airway diameter by placing an anterior and/or posterior costal cartilage graft (ACCG, APCCG, PCCG), most often costal cartilage or thyroid ala cartilage. This procedure can be performed as a double-stage surgery (placement of a tracheostomy) or as a single-stage surgery (removal or non-placement of a tracheostomy). Multiple factors may impact voice outcomes, such as prelaryngeal muscle dissection during surgery, baseline subglottic stenosis severity, and laryngeal nerve injury.

Cricotracheal Resection (CTR)

CTR involves the excision of the anterolateral cricoid plate and anastomosis of the distal tracheal ring to the proximal thyroid ala with suture lines placed in the posterior cricoid mucosa to reapproximate the trachealis to the more proximal cricoid plate [5]. This procedure should be

reserved for experienced surgeons as results are highly surgeon dependent. Furthermore, this procedure may significantly alter the voice. In particular, CTR decreases the fundamental frequency of connected speech and vowel phonation and changes the acoustic signal type [6].

Slide Tracheoplasty

Slide tracheoplasty is a surgery to increase tracheal diameter. The trachea is opened anteriorly and posteriorly. It is then slid up onto itself and reconnected to make the trachea shorter, but wider. This procedure is classically performed for long-segment congenital tracheal stenosis. Limited data exist regarding specific voice outcomes after such procedures, but vocal fold paralysis has been reported in approximately 5% of patients [7].

Epidemiology

Voice disturbance following airway reconstruction is not uncommon; more than half of children who undergo airway surgery are reported to have postoperative dysphonia, which is often described as severe [3]. Parents and patients concerns about normalization of the airway and removal of the tracheostomy often overshadow initial concerns about voice outcome and dysphonia. Concerns about voice become more relevant during middle school and high school. Typically, children in these age groups are moving from one environment to another and therefore encountering novel situations where their abnormal voice becomes more relevant. Consequently, in our experience, children in these age groups are more likely to seek care related to their dysphonic voice. Additional benefits of early voice evaluation include counseling about potential voice problems post reconstruction. Risk factors for poor voice outcomes after an airway reconstruction are numerus and include [8, 9]:

- Complete laryngofissure
- Cricotracheal resection
- Posterior grafting
- Higher grade of subglottic stenosis (SGS)
- Revision airway surgery
- History of multiple airway reconstructions

Patients with low-grade stenosis (grade 1–2), single-stage procedure, and fewer comorbidities are less likely to have post-reconstruction dysphonia [9, 10].

Pathophysiology

Post-reconstruction dysphonia is multifactorial and occurs in more than half of patients. Postsurgical causes of dysphonia include, but are not limited to:

- Abnormal vocal fold mobility
- Persistent subglottic stenosis
- Anterior commissure blunting
- Posterior glottic diastasis
- Prolapsed petiole
- Vertical asymmetry of vocal folds
- Vocal fold scaring
- Supraglottic compression

Dysphonia after airway reconstruction may depend on the baseline airway problem and the type of surgery performed. Glottic incompetence is often a problem, either secondary to vocal fold immobilization due to laryngeal nerve dysfunction, cricoarytenoid fixation, or glottic diastasis with a posterior graft. With vocal fold immobility and cricoarytenoid fixation, patients often compensate with supraglottic phonation, using their ventricular folds to produce voice [11]. Such compensatory compression patterns and alternate sources of vibration used by these children often result in moderate to severe dysphonia. These patients also typically complain of breathiness, strain, and fatigue. Additionally, patients may have difficulty modulating or creating sound due to excessive scarring and/or injury to the prelaryngeal muscles during dissection for their airway reconstruction [5]. A complete laryngofissure violates the anterior commissure and depending on postoperative healing or long-term changes

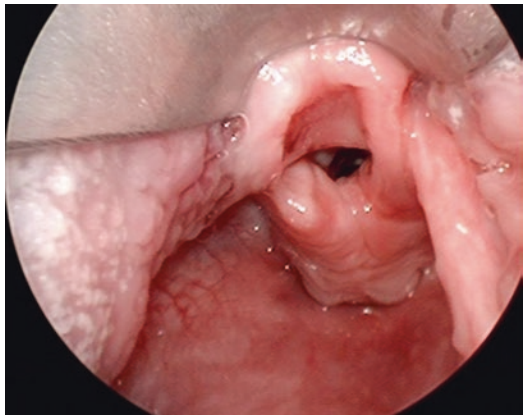

Fig. 40.3 Petiole prolapse as noted during a rigid microlaryngoscopy

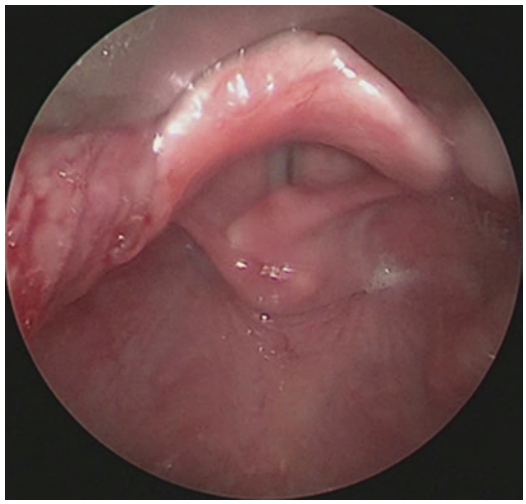

Fig. 40.4 Right arytenoid prolapsing over the vocal folds

during pubertal growth may result in off-level vocal folds, anterior commissure blunting, or petiole prolapse (Fig. 40.3) [12, 13].

As previously mentioned, CTR will frequently lead to a more severe degree of dysphonia. This is partly explained by either the removal of the cricothyroid muscle and obliteration of the cricothyroid membrane [5]. Moreover, there is a possibility of arytenoid prolapse (Fig. 40.4) due to destabilization of the cricoarytenoid joint or vocal fold paralysis secondary to recurrent laryngeal nerve injury during surgery.

Presentation

Most patients will report some degree of dysphonia after airway reconstruction. Typically, voice outcomes after airway reconstruction include roughness, breathiness, supraglottic phonation, or inappropriate pitch [1, 14]. These children often present when they are becoming more social or making decisions about future careers. In younger children (ages 5–10 years), issues may arise as children begin participation in sports and other social activities. As children reach middle school age, they may have more issues related to being in multiple different classroom settings with different peer groups and teachers who are not familiar with their voice. Concerns include peers noticing this dysphonia as well as difficulty being heard in a noisy environment, embarrassment about their voices, and reduced willingness to participate in class [15]. In adolescents, the voice is important for social interactions, defining their identity, and determining their future occupation. Teenagers report embarrassment and fear of peer responses to their voice, as well as frustration and social isolation [15].

Otolaryngologist and Speech-Language Pathologist Approach

Patients with dysphonia after airway reconstruction are typically complex, and there are a variety of parameters that must be evaluated. As such, the evaluation is best performed in a multidisciplinary fashion with both an otolaryngologist and speech-language pathologist well-versed in voice pathology. Multidisciplinary voice evaluation frequently influences the course of treatment for patients with airway reconstruction and dysphonia [14, 16].

History

The history for these patients is largely obtained from the parents, but the child's perspective is also very important when discussing the management

of voice concerns. Many children will have had voice issues throughout their life related to prior intubations, scarring in the airway, and prior surgical interventions. At the time of presentation for voice evaluation, it is necessary to determine the child's airway history including intubation history, history or presence of a tracheostomy, and prior surgical interventions. Furthermore, attention should also focus on current medications and relevant comorbidities such as pulmonary or swallowing disorders. Details of the surgical interventions performed, such as if anterior or posterior grafts were placed or if a laryngofissure was performed, are useful in helping predict potential causes of the dysphonia. Reviewing operative notes and operative videos, if available, may also be helpful to determine potential causes of the dysphonia.

As with any history, the clinicians should determine the nature and course of the dysphonia, alleviating and aggravating factors, and if prior interventions (e.g., voice therapy, injections, surgery) have been attempted. If they have had prior interventions, the specifics of those and their outcomes should be elicited. The family's perception of the voice quality (e.g., weak, breathy, rough, raspy, harsh, deep) and how it is affecting the child's quality of life in all environments (e.g., home, school, social, work) is important. Families will sometimes report that the child has "two voices" – typically a weaker one (presumably the glottic voice) and a stronger but deeper one (presumably a supraglottic voice). They may describe this as a "duck" voice or "superhero" voice. For a younger preverbal patient, a formal preoperative voice evaluation may be challenging; however, the breathing pattern, presence of stridor, and babbling can be documented.

Indices such as the pediatric Voice Handicap Index (pVHI) [17] or Voice-Related Quality of Life (VRQL) [18] can assist in elucidating the perceived impact of dysphonia. Similar factors that may cause laryngeal irritation and exacerbate voice problems in other patients, such as laryngopharyngeal reflux, allergic rhinitis, chronic cough, asthma, sleep-disordered breathing, smoke exposure, and vocal misuse/overuse, must be evaluated, as well.

Additionally, operating on the larynx can significantly and disparately impact voice, airway, and swallowing. As such, the current airway status (e.g., tracheostomy, decannulated), presence of stridor or dyspnea on exertion, and time of last airway evaluation should be noted. Lastly, patients who undergo open airway reconstruction are also likely to experience some degree of postoperative dysphagia symptoms and delayed return to oral intake. Cough, choking events, and aspiration pneumonias should be documented. The patient's current feeding status and swallowing safety should also be assessed.

Examination

The examination is typically completed in conjunction with the speech-language pathologist. Combined evaluation has been proven to be beneficial for decision-making regarding voice management and potential surgical interventions [14]. A general head and neck examination should be completed. Special attention should be paid to the intelligibility, voice quality, effort for voice production, and voice range. Specific perceptual instruments, such as the grade, roughness, breathiness, asthenia, and strain (GRBAS) [19] scale or Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) [20], are used to document the quality and severity of the dysphonia. These scales assist in establishing a baseline and monitoring progress over time. Interestingly, a prior study showed only weak-to-fair correlation between the parent-reported pVHI and expert ratings of voice quality using the CAPE-V [21]. The presence of diplophonia and if two discrete voices can be elicited should be documented.

The otolaryngologist should pay attention to the presence of stridor or respiratory distress; however, most children presenting for voice evaluation after airway reconstruction have typically overcome this challenge. Finally, patients who have undergone airway reconstruction may have

other factors that can affect their voice outcome or ability to participate in therapy, such as syndromes, other congenital abnormalities, or developmental delay that should be noted.

Instrumental Assessment

Endoscopic Evaluation

Laryngoscopy and videostroboscopy should be performed. Again, the speech-language pathologist and otolaryngologist serve complimentary roles. Depending on the child's age, ability to cooperate, and anatomy, flexible transnasal and/or rigid transoral 70-degree stroboscopy may be completed. Whereas one or the other is often adequate for evaluation of common laryngeal lesions, both transnasal and transoral exams may be necessary to fully evaluate the anatomy and function in these post-airway reconstruction patients. The use of a distal chip telescope will improve image quality, and recording the examination is useful for reference. Careful attention should be paid to attempting to determine the sound generator for phonation (e.g., glottic or supraglottic or both), presence of a posterior glottic gap, if the vocal folds are level, the degree of scarring, the mobility of the vocal folds and arytenoids, and the degree of effort/strain with vocalization. Of note, examination of the glottis during phonation is often difficult due to the degree of supraglottic collapse and/or squeeze seen in these patients as well as postsurgical anatomical variation. Parameters of vibratory patterns should also be evaluated via stroboscopic exam.

Rigid endoscopy in the operating room alone is not adequate for evaluation of vocal pathology. However, the mobility of the arytenoids, presence of posterior glottic scarring or diastasis, and other structural anomalies can be assessed and may add important information to the clinical picture.

Acoustic and Aerodynamic Evaluation

Post-airway reconstruction patients should undergo acoustic and aerodynamic evaluation. This is typically completed by the speech-language pathologist, and detailed descriptions of these exams can be found in other chapters. Briefly, acoustic analysis provides information regarding the fundamental frequency; jitter, shimmer, and noise-to-harmonic ratio; and frequency range and may also include spectral/cepstral measures such as cepstral peak prominence. Aerodynamic measures provide information regarding glottal efficiency by determining the average airflow rate and estimated subglottic pressures. Not all patients will be able to produce a type I (periodic) signal and a measurement of fundamental frequency. Common features seen in post-airway reconstruction patients are a lower pitch and reduced pitch range, breathiness, and a reduced maximum phonation time [14]. These assessments help provide a baseline and can be used to measure response to voice or surgical therapies over time. A prior study suggests that the majority of children are able to complete the acoustic and aerodynamic assessments with a significant proportion of post-airway reconstruction patients having severe dysphonia [22]. Recording enough voicing segments may be challenging and sometimes impossible; protocols should be tailored to the patient's capability.

Other Modalities

Dynamic voice computed tomography (CT) has been described but is not yet widely available. This CT is performed with the patient holding a sustained /i/. The CT does not require contrast and is a relatively low dose of radiation (same as a general head CT). The main benefit of the voice CT is that it allows for evaluation of the glottis during phonation. As previously mentioned, this is often difficult to assess endoscopically due to the supraglottic structures. This exam is particularly useful for evaluating the degree of glottic gap during phonation (Fig. 40.5), but cannot evaluate the mucosal wave.

High-speed videography is another tool that is still largely used in a research setting, but is also used in some clinical settings. This exam can provide extremely detailed information regarding the mucosal wave and sound generator with

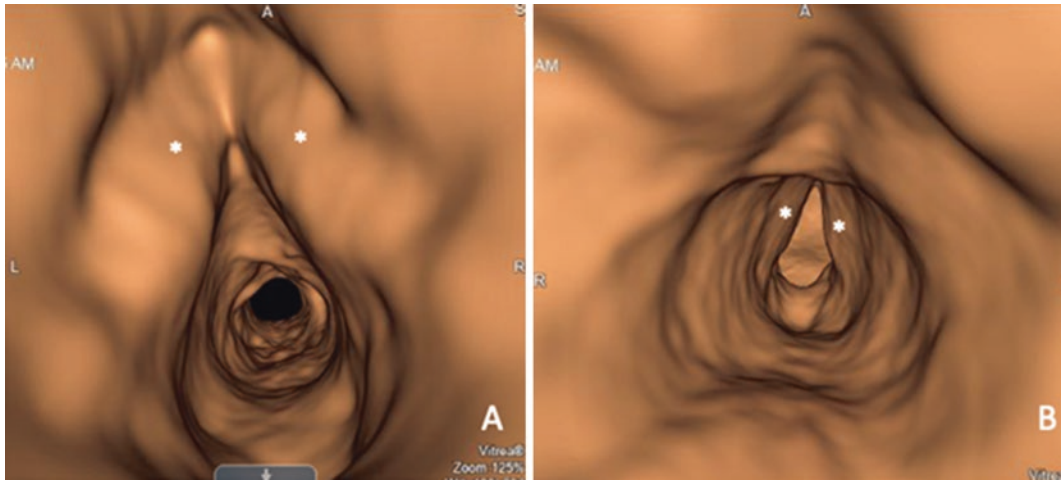

Fig. 40.5 View of the glottic gap from above (**a**) and below (**b**) with the 3D reconstruction of the dynamic voice CT scan. ∗ = vocal fold

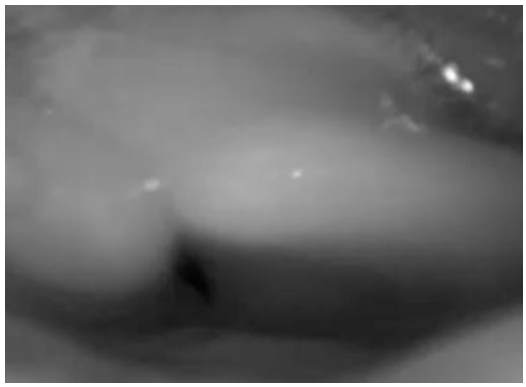

Fig. 40.6 High-speed videolaryngoscopy showing supraglottic phonation

higher reliability when compared with traditional videostroboscopy (Fig. 40.6) [4]. Limitations of this exam, however, include the availability of the equipment and the massive amount of storage space required for the data obtained for even very short examinations.

Ultrasound for evaluation of the supraglottic and glottic structures has also been described by some pediatric voice specialists. While the presence of cartilage grafts may alter visibility to a degree, the larynx does not typically calcify until around 40 years of age making this a viable tool in children.

Differential Diagnosis

The differential diagnosis for a child presenting with dysphonia after airway reconstruction is broad. While more typical laryngeal pathology (e.g., nodules, cysts, polyps, papilloma) may be present, the post-airway reconstruction vocal pathology is typically more complex. The dysphonia may relate to scarring in the subglottis, glottis, or supraglottis, arytenoid prolapse or fixation, vocal fold atrophy, vocal fold scar, or vocal fold vertical asymmetry preventing an adequate mucosal wave, anterior commissure blunting, posterior glottic diastasis, and the compensatory use of supraglottic structures for phonation. Furthermore, patients with history of airway reconstruction often have several comorbidities, such as pulmonary and neurologic disorders that can also impact voice quality. Often, a combination of these pathologies can be identified.

Management

Understanding the family and patient's motivations for voice evaluation and how it is impacting the child's day-to-day life is important in helping the family determine goals of therapy and/or

surgical interventions. Not every patient will have the same needs or goals for their voice. Additionally, as opposed to the airway procedures these children underwent in their youth, voice interventions are more about quality of life; thus, a discussion with both the patient and family about their goals and expectations is crucial. For example, a young girl using a supraglottic voice may desire a more feminine-sounding voice. In that case, transitioning to a glottic voice may be more appropriate even if it is slightly weaker or breathier. On the other hand, if the patient is a young male whose goal is a louder voice, finding ways to help him use the supraglottic voice may be more appropriate. Additionally, some patients may not be bothered by their voice at initial evaluation. It is important to discuss with the families that voice interventions are not a "now-or-never" option. The child and family committing to voice therapy and perceiving a need for change will improve adherence to exercises. If surgery is to be undertaken, children of adequate age and maturity should be included in the discussion as voice is such an integral part of a person's identity.

Management of these patients typically involves voice therapy alone, before surgery, and/or after surgery. Voice therapy can help the child access and use the supraglottic and/or glottic voice more easily when appropriate. Prior studies have shown that some children can achieve periodic vibration when phonating with non-glottic structures suggesting that in appropriate children, therapy can help achieve a better adapted and more acceptable supraglottic voice [14].

Initial surgical intervention should be tailored to the anatomic considerations of the patient. This often includes injection laryngoplasty, which can help improve a glottic or a supraglottic voice. Injection in the typical location, however, may be more difficult due to scarring, and, in some cases, an intracordal injection is required. Other procedures have been described specifically for posterior glottic diastasis including laryngofissure with partial posterior cricoid reduction, endoscopic pharyngoepiglottic-aryepiglottic fold advancement-rotation flap with interarytenoid interposition, interarytenoid submucosal implant augmentation [23], buccal flap augmentation [24], and endoscopic posterior cricoid reduction [25]. The following section will focus specifically on the latter intervention for posterior glottic diastasis. As awareness of post-airway reconstruction dysphonia increases, surgeons should certainly think more critically when performing airway procedures. Some strategies to help minimize the impact on voice include avoiding a complete laryngofissure when possible, meticulous reapproximation of the vocal folds in the setting of complete laryngofissure, and creating appropriately-sized (not oversized) posterior grafts when they are indicated.

Operative Approach

This section will focus on a surgical approach to posterior glottic diastasis: endoscopic posterior cricoid reduction.

Indications

Endoscopic posterior cricoid reduction is a surgical procedure to address posterior glottic diastasis. This may be caused by prior intubation and/or prior airway reconstruction, particularly when posterior grafts are placed. Patients with posterior glottic diastasis, as demonstrated on endoscopy and/or dynamic voice CT, who cannot obtain an adequate voice with voice therapy, whose voice is impacting their quality of life, and who do not have concerns for airway compromise, are candidates for this procedure. The ability to obtain adequate exposure endoscopically is also a consideration.

Key Aspects of the Consent Process

The procedure includes suspension microlaryngoscopy, use of a CO_2 laser, and reduction in the size of the airway. Risks associated with each of those should be discussed. For suspension microlaryngoscopy, the risk of injury to the lips, teeth, and gums should be noted. Additionally, there is

potential for dysgeusia or hyperextension injury to the neck. With use of a CO_2 laser, risk of eye injury, burns to head and neck structures, and the risk of airway fire should be acknowledged. With the posterior cricoid split, there is a risk of tracheoesophageal fistula and potential need to open the neck for repair should it occur. Finally, the family and patient should understand the balance of voice, airway, and swallowing. It should be explicitly stated that making in reducing the glottic inlet to help improve the voice, there is an inherent decrease of the airway diameter. While care is taken to minimize the risk, there may be airway compromise and need for future airway interventions, including intubation, tracheostomy, and/or revision airway reconstruction. Additionally, the voice may fail to improve despite the surgical intervention, and other therapies may still be required (including voice therapy and/or additional surgical interventions).

Equipment

Traditional suspension laryngoscopy equipment should be available. Additionally, a CO_2 laser, laser technician, and laser safety equipment should be available. While the procedure can be performed with cold instrumentation, in the senior author's experience, it is advantageous to have the laser for this case.

Steps (Fig. 40.7)

1. *Patient Positioning and Preparation*

   Discussion with the anesthesia team regarding preference for spontaneous ventilation and low oxygen levels while the laser is in use should be performed prior to the procedure. The patient is brought to the operating room and placed in the supine position on the operating table. Perioperative antibiotic prophylaxis (e.g., cephalexin) is given at induction. The patient should undergo initial rigid microlaryngoscopy and bronchoscopy (MLB) with a dental guard, an appropriately sized Phillips blade, and a rigid Hopkins rod 0-degree telescope. Laryngotracheal anesthesia should be employed. Sizing of the airway should be performed pre- and postoperatively with endotracheal tubes. Photodocumentation should be employed throughout the case.
2. *Exposure*

   After the initial MLB has been performed, the patient should be placed in suspension with the largest Lindholm laryngoscope the patient can accommodate. If the exposure is not adequate with this, taping of the anterior neck to provide constant cricoid pressure or a different laryngoscope (such as a Zeitels universal modular glottiscope placed in the laryngeal vestibule) may be employed. Furthermore, a shoulder roll may give some additional degree of exposure.
3. *Injection*

   Once adequate exposure is obtained, the posterior cricoid can be palpated. An orotracheal injector is then used to inject 1% lidocaine with 1:100,000 epinephrine into the posterior cricoid plate to assist with hemostasis.
4. *Laser Precautions*

   While allowing time for the local anesthetic to work, standard laser precautions should be instituted. The patient's eyes should be taped with silk tape and wet eye pads and the skin covered with wet towels. All room staff should have adequate eye protection. The windows to the room should be covered and signs placed on each entrance warning that the laser is in use. The microscope should be appropriately prepared for use with the laser, and a smoke evacuator should be turned on. The accuracy of the laser beam must be ensured *off the field*. Communication should be instituted with the anesthesia team regarding safe oxygen levels. A basin of water or saline should be available on the scrub table in case of fire.
5. *Cricoid Reduction*

   Once all necessary laser precautions have been instituted, the operating microscope is brought into the field. The posterior split and reduction are then performed using the CO_2 laser (SurgiTouch+ set at 16 W, 2 mm depth, and approximately 1.6 mm circle shape). The surgeon should have a predetermined width

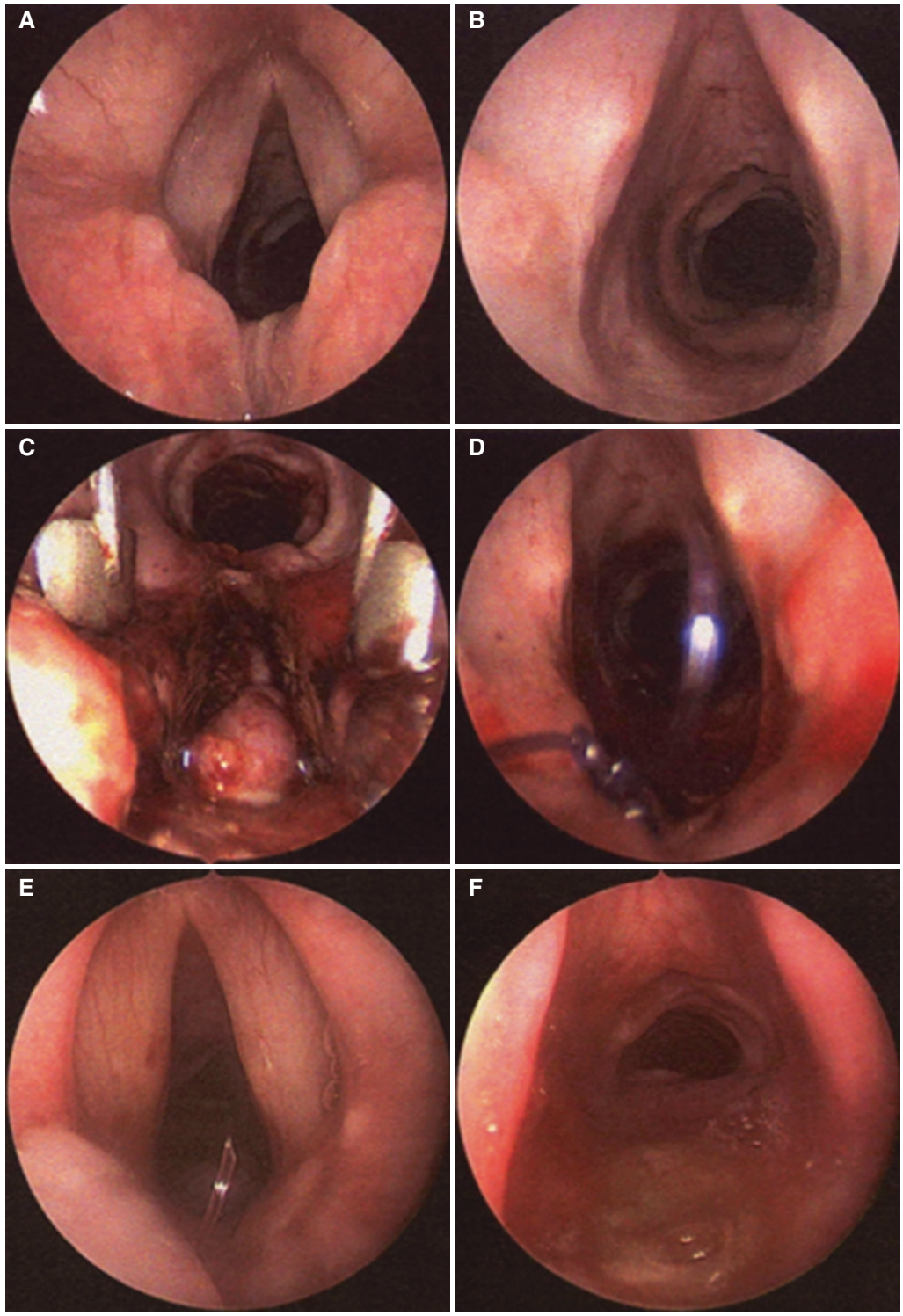

Fig. 40.7 Endoscopic cricoid reduction. (**a**) Preoperative view of the posterior glottic diastasis (**b**) Closer look at the preoperative glottic diastasis. (**c**) CO2 laser cricoid split with vocal folds spreader in place demonstrating the split cricoid and the preserved posterior wall. (**d**) Endoscopic sutures of the posterior cricoid plate. (**e**) Postoperative view of the glottis at 1 week. (**f**) Postoperative view of the subglottic area at 1 week

of reduction planned and size the laser shape accordingly. Care should be taken to stay in a straight line in the midline when performing the split and to not take a wider segment than planned. This can be completed with a D-knife or Blitzer knife if the CO_2 laser is not available. The surgeon should confirm that the split extends the full length of the cricoid by using a vocal fold spreader to distract the cricoid laterally. The posterior common party wall should be respected throughout the dissection and carefully inspected afterward to ensure a tracheoesophageal fistula is not created. The microscope and the zero-degree telescope can be used for visualization throughout this process, as needed. Afrin-soaked pledgets may then be used to attain hemostasis.

6. *Refinement of the Cricoid Split*

   The edges of the split should be refined as needed using the CO_2 laser (with a straight line instead of a circle) to allow for excellent midline approximation. Once the laser is no longer required, it may be helpful to communicate to the anesthesia team that the oxygen level can be increased. At this point, attention is turned to suture repair of the cricoid.

7. *Cricoid Closure*

   Using an endoscopic needle driver and 4-0 PDS suture on an RB-1 needle, the cricoid is reapproximated in the midline with simple interrupted sutures. A distal suture is placed followed by a proximal suture. Two sutures are typically adequate for closure. A post-procedure photograph should be taken, and the patient can then be taken out of suspension.

8. *Final Bronchoscopy*

   The larynx should again be exposed with the Phillips blade and repeat sizing of the airway performed using endotracheal tubes.

9. *Final Tips*
   - Intermittent intubation may be employed throughout the case.
   - Excellent communication with the anesthesia team is helpful.
   - Size the airway before and after the procedure.
   - Make sure the midline split is both midline and straight. Right-handed surgeons will tend to veer to the right and left-handed surgeons veer to the left.
   - Zeitels universal modular glottiscope often provides excellent exposure for more difficult cases.

Postoperative Management and Follow-Up

The patient should be admitted for overnight observation with airway monitoring to either an "airway stepdown" unit or the intensive care unit. Acetaminophen and ibuprofen can be employed for pain control along with oxycodone as needed for severe pain. The senior author allows the use of ketorolac on postoperative day 1, if needed. The patient should be on antibiotic prophylaxis (amoxicillin-clavulanate) and a proton pump inhibitor for 7 days and 1 month postoperatively, respectively. The patient can be orally fed after the procedure. Ideally, the patient should not receive corticosteroids that may inhibit adequate healing. A repeat microlaryngoscopy and bronchoscopy is performed one week postoperatively to ensure adequate healing of the cricoid.

Emerging and Evolving Techniques of the Future

Innovative technologies exist to optimize the evaluation of these complex patients prior surgery.

- As already mentioned, high-speed videoendoscopy improves the ability to rate tissue vibratory characteristics when compared with videolaryngoscopy in children with supraglottic phonation. This information may allow better understanding of the underlying mechanisms of voice production in these individuals, leading to improved therapeutic and surgical recommendations [4].
- Predicting the impact of the surgery on airway dynamics may decrease morbidity and

improve overall quality of care for these complex patients. Cine magnetic resonance imaging (MRI) combined with computational fluid dynamics (CFD) has been reported to model the airflow through the dynamic airway in complex airway cases. CFD modeling might reveal the specific portions of pressure and energy losses in both inhalation and exhalation, allowing targeted interventions for these specific locations.

- Also, the use of dynamic voice CT may provide complementary information to the videostroboscopy [26, 27]. For patients with complex airway history, the pattern of laryngeal closure could be detected more frequently compared to the standard endoscopic examination. Moreover, the location of gap closure and the vertical closure pattern of the glottis may have a better yield with the dynamic voice CT scan. Dynamic voice CT shows promise as an additional tool for evaluation of patients with a history of complex airway procedures by providing complementary information that might alter surgical decision-making.

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Non-cleft Velopharyngeal Insufficiency

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Katherine M. McConville and Catharine B. Garland

Overview

Non-cleft velopharyngeal dysfunction is an eclectic concept. It involves the intersection of multiple etiologies of disorders (neuromuscular, functional, or postsurgical) that impact the normal function of the aerodigestive tract for voicing, speaking, swallowing, and other oral pressure generation tasks (like blowing up balloon and playing a brass or woodwind instrument).

Specifically defining the affected population is challenging, as even patients with similar etiologies may present in a widely different manner. Milder cases may go undetected or ignored due to burden of care, medical complexity, or poor awareness of the signs and symptoms of velopharyngeal dysfunction. A thorough and flexible assessment of the disorder and its impact on swallowing, resonance, speech intelligibility, and overall patient quality of life is particularly important in order to tailor a treatment protocol.

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Definitions

Velopharyngeal dysfunction (VPD) is a broad term for any condition wherein the normal, consistent, and complete closure of the velopharyngeal port is not achieved for the production of oral sounds. More specifically, velopharyngeal *insufficiency* (VPI) denotes a structural or anatomic cause for velopharyngeal dysfunction, such as stenosis or loss of tissue. Velopharyngeal *incompetence* (also abbreviated VPI) describes neurophysiological impairment of muscles aiding in palatal closure, such as stroke or traumatic brain injury. Both velopharyngeal insufficiency and velopharyngeal incompetence are sometimes accounted for using the term velopharyngeal inadequacy (confusingly, also abbreviated VPI). While velopharyngeal inadequacy is often an accurate descriptor of many presentations, it should be noted that it does not fully account for any behavioral cause or contribution to a resonance issue in the way that the broader term velopharyngeal dysfunction (VPD) does. The symptoms of velopharyngeal dysfunction include hypernasality, nasal emissions, difficulty producing high pressure oral consonants, and nasal regurgitation.

Hypernasality is an increase in the amount of sound energy resonating within the nasal cavity during voiced sounds. It is typically attributed to velopharyngeal dysfunction, although it has been documented that some amount of nasalance can

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be accounted for by sound transfer through the palatal bones and tissues [1]. Nasal emissions are the escape of air through the VP port and nose during the production of nonnasal speech sounds. This escape of air is most readily heard on high pressure consonants like /s/ and can occur in speech with or without hypernasality. Lastly, nasal regurgitation is the retrograde flow of food or liquid into the nasal cavity during oral intake. While this is not directly a speech or resonance issue, it can be a symptom of incomplete closure of the velopharyngeal (VP) port and may be a meaningful symptom of complaint for patients with velopharyngeal dysfunction.

Epidemiology

With the large number of potential causes or contributors to velopharyngeal dysfunction, prevalence data on non-cleft velopharyngeal dysfunction is not reported. The presence of VPI following neurologic injury or surgical or radiation treatment is only reported when the VPI is severe enough to cause significant speech intelligibility or swallowing issues and is thus likely underreported in these populations.

Causes

Any condition or event that alters the orientation of the velopharyngeal port or its structures has the potential to cause velopharyngeal insufficiency. Treatment of oral and pharyngeal cancers is the most common non-cleft cause of VPD in adults but much less common in children. Surgical treatment may involve resection of the structures that separate the oral and nasal cavities. In addition to loss of these tissues and structures, stiffened or scarred tissue from surgical manipulation or radiation can render structures in the VP port less mobile, also contributing to velopharyngeal insufficiency. Notably, since radiation changes can occur slowly over time, patients may not note speech or resonance changes immediately, and the emergence of resonance issues may be more insidious. In children, congenital masses of the oral or nasal cavity requiring early surgical intervention may have similar effects.

Platybasia refers to a flattening of the cranial base angle, where the points of the angle are formed by the anterior border of the foramen magnum, the center of the hypophyseal fossa, and the frontonasal suture. When this angle is more obtuse (sometimes due to craniofacial anomalies like 22q11.2 deletion), the pharyngeal spaces are increased which may cause velopharyngeal insufficiency [2]. Platybasia may alternatively render VPI more likely when combined with other structural changes, such as adenoidectomy or uvulectomy. Similarly, surgeries to advance the midface (maxillary advancement) for cosmetic or functional reasons alter the spatial orientation of the palatal structures and the posterior pharyngeal wall, potentially impacting VP closure.

Velar dysplasia refers to abnormal development or growth of the velum. This can be caused by a craniofacial anomaly such as hemifacial microsomia. In the case of hemifacial microsomia, growth of the face is restricted on one side. This can both render a portion of the velum small or insufficient for closure and can also present with limited development of the nerves for innervation of the muscular velum, impacting velopharyngeal competence [3].

An irregular adenoid surface has the potential to prevent the velum from sealing smoothly against the portions of the posterior pharynx which can interfere with complete closure. More frequently, adenoidectomy can result in temporary or permanent resonance issues. This is thought to occur in particular if a patient is accustomed to closing the velum against the adenoid, but typically should resolve within 6 weeks of surgery [3]. If scarring occurs along the posterior pharynx from adenoidectomy or if velopharyngeal closure was limited to begin with, a change in the spatial orientation of these structures could potentially cause longer-term issues. Tonsillectomy does not appear to bear the same risks as the involved structures are lower in the pharynx. However, anecdotally, very large tonsils can extend into the velopharynx shunting the port open.

Neurophysiologic impairment impacting the muscular control of the structures of the velopharyngeal port and their closure or coordination for speech most often include stroke or traumatic brain injury. Dystonias of the palate have also been reported. Those causing velopharyngeal incompetence are marked by the involuntary depression of the palate during speech [4]. This may be mistaken for spasmodic dysphonia due to the presence of some overlapping symptoms such as breathy voice breaks or increased frequency of sound disturbance in connected speech versus singing. Medical and procedural management are described in the limited literature on a case-by-case basis. Although available case series describes speech therapy as not beneficial, there may be benefit in exploring compensatory strategies for speech and communication. The author recalls a case of negative dystonia that ultimately benefitted most from a palatal prosthesis. However, strategies such as whispering with an amplifier did bolster intelligibility until more effective treatment was rendered.

Behavioral velopharyngeal dysfunction in patients without any history of velopharyngeal inadequacy as a precipitating factor is thought to be rare. Despite this, patients with purely functional VPD do present clinically. The risk factors for this are not well understood but have been noted to include tonsillectomy, with the notion that postoperative pain may lead to avoidance of use of some of the pharyngeal structures [5]. Other presentations may appear to be psychogenic. However, this should never be the first assumption and can only be considered after a thorough process of elimination with a comprehensive assessment.

Stress VPI is a presentation noted in instrumentalists playing brass or woodwind instruments. It has been described as gap or incomplete closure of the VP port during higher pressure activity but can occur with low pressure production. In some cases, air leak just prior to playing an instrument may fuel complaints [6]. This is sometimes reported in persons who have been engaging in protracted practice, instrumentalists who have recently undergone adenoid surgery, and sometimes those complaining of embouchure changes (possibly due to compensation). Stress VPI is only documented in instrumentalists because of their unique use and pressure demand of the VP port. Feasibly, it could present as velopharyngeal insufficiency or incompetence, and so it necessitates careful assessment with symptom provocation while the patient plays his or her instrument.

Presentation

The key features of non-cleft velopharyngeal dysfunction are hypernasality and/or nasal regurgitation during intake of liquid. Severe maladaptive articulatory patterns, sometimes associated with cleft-related VPI, are feasible in severe non-cleft-related cases, but anecdotally are not observed to the same extent. Presumably this is because many cases of non-cleft VPD have established phonological patterns prior to the onset of velopharyngeal dysfunction.

Speech-Language Pathologist Approach

Non-cleft velopharyngeal dysfunction can negatively impact speech and communication, comfortable and effective deglutition, or other activities occupying the aerodigestive tract. Since the causes and presentations of these types of disorders vary so widely, speech pathologists may collaborate with a number of specialists to best assess and treat patients who may be affected.

Treatment teams may include speech pathologists, otolaryngologists, plastic surgeons, dentists, and prosthodontists. The SLP is valuable to the treatment team in assessing a patient's overall communication or swallow function. They help determine the level of impairment presented by velopharyngeal dysfunction as a consequence of, or in comparison to, other impairments present (dysarthria, postsurgical defect, aphasia, voice disorder, other contributor to dysphagia, etc.). In evaluating VPD, a speech pathologist is tasked not only with assessing the severity and characteristics of the presentation but also determining

if treatment would resolve the VPD. When therapy cannot eliminate the VPD, the speech pathologist must confer with the team and the patient to communicate about whether intervening in other aspects of speech, voice, swallow function, etc. could improve function in some capacity. In this manner, the patient and care team weigh the risks and benefits of the available management options. Even when speech therapy is not indicated as a primary or adjunct form of treatment, the speech pathologist's assessment provides essential baseline and outcomes data for the treatment team as a whole to better judge and quantify a patient's response to any intervention.

Assessment

A thorough case history should collect details regarding any changes in functional status and enquire about any pre-existing conditions that could impact the patient's baseline function or predispose them to resonance changes (such as prior history of adenoidectomy, sleep apnea surgery, or stroke). Additionally, it is exceedingly important to identify the patient's perspective on any changes to speech and swallow function and to determine what the most troublesome aspect is. Is it, for example, not being understood, difficulty projecting, having speech that is remarkable or distorted to others, or difficulty with nasal regurgitation? It is helpful to know if the patient is chronically bothered by these issues or if it is only in specific settings (eating in front of others, meeting new people, speaking at school) that the symptoms are concerning as this may also shape the course of treatment. In children, a prior history of difficulty breast feeding or drinking from a standard bottle may be telling.

Ideally assessment of velopharyngeal function should include both perceptual and objective measures that span not only function of the VP port but overall speech or communicative function as well. Articulation testing should be fairly broad, sampling all speech sounds not only to account for errors or distortions related to the VPD but any other errors (such as developmental errors, reduced labial contact for both oral and nasal labial sounds due to oral surgery, weakness from stroke, or prominent dentition) that could interact with VPD or otherwise contribute to the overall impression of a speech-related issue.

Regarding detection of VPD, speech tasks to identify hypernasality would consist of vowels and voiced speech sounds. but should also sample some voiceless pressure sounds like /p/, /t/, /s/, and /ʃ/ that help to identify nasal emissions. Listeners often struggle to distinguish hypernasality from hyponasality. This is a difficult task and can be further complicated when resonance is mixed. It may be helpful to compare a spoken passage without nasal sounds against a passage replete with them. If the nasal sentences sound "better" than those without nasal sounds in them, one may suspect some hypernasality. Conversely, if the nasal sentences sound "off" or congested, hyponasality may be the contributing factor.

High pressure sounds should be sampled in multiple contexts including syllables, words, and connected speech (see Appendix 1 for suggested stimuli). More structured tasks render analysis of hypernasality or nasal air emission more easily, but some issues may be masked in shorter contexts or worsen due to the need to sustain effort and/or attention for connected speech. When selecting syllables and words for assessment, it is important to remember that hypernasality is more readily detected in high vowels like /i/.

Objective measurements made without VP port visualization, but through measurements of the acoustic or aerodynamic features of nasal air and sound energy are known as "indirect measures." A nasometer typically uses a computer interface with hardware to measure the ratio of oral versus nasal sound energy produced with speech. From this, a "nasalance" measure is obtained. Nasalance offers an objective benchmark in assessing a primarily perceptual phenomenon but does not necessarily directly reflect hypernasality. Nasalance varies with a number of factors including vowel selection and nasal consonant coarticulation. Global normative data for nasalance has not been established due to the various factors that can affect the readings [7]. For these reasons, nasalance measurements should never be the sole criteria used to separate

normal voice and speech from VPD, but nasometry can provide helpful data in assessing resonance and documenting changes in nasalance with treatment.

Pressure flow measurement instruments using pneumotachographs to measure nasal air escape during speech tasks are described in the literature as beneficial in distinguishing normal VP function from abnormal function [8]. These instruments are less common clinically than nasometry equipment. This is unfortunate, as quantification of air pressure and flow escaping through the velopharyngeal port during speech closely represents the degree of incomplete velopharyngeal closure [9]. While hypernasality is primarily an acoustic-perceptual phenomenon, weak pressure consonants and nasal air emission associated with VPD are *aerodynamic* in nature.

When objective measures are not available, a clinician may consider recording data from lower-tech assessment methods. This could include counting the number of nasal emissions observed while extending a straw from the patient's nare to the clinician's ear while reading a set of high pressure sound sentence stimuli aloud.

Methods of visualizing the velopharyngeal port are mostly described and generated from the cleft palate literature and are an essential part of evaluating VPD. Fluoroscopic and endoscopic imaging are the predominant methods for examining elevation of the palate and closure of the velopharyngeal port [10]. Some research literature also describes use of MRI to view the length of relevant structures and muscles including the levator veli palatini, depth and width of the pharynx, and various calculations of the length and range of motion of the velum [11], but this has not been used widely in clinic settings as both the cost and complexity of sampling speech during MRI may be limiting factors. Cephalometric x-ray imaging was used more frequently in the past to gauge VP closure, but images are limited to those obtained in production of isolated, sustained sounds and thus are not favorable for diagnostics [12].

Multiview videofluoroscopy during production of speech stimuli can be used to view velopharyngeal port dynamics for real-time speech-based tasks. This involves collection of lateral views for observing dynamic palatal movements and the degree of closure along this dimension as well as a frontal view to examine palatal symmetry, lateral pharyngeal wall movement, and vertical level of palatal elevation [12]. Nasendoscopy affords many of the same visualization benefits as videofluoroscopy but is thought to be more helpful in detecting small gaps in palatal closure that can often be the causes of non-cleft VPD [10]. Since endoscopy bears no radiation-related risks, speech samples collected with endoscopy can also be lengthier if the patient tolerates it [12], offering further benefit in terms of stimulability probing or treatment with biofeedback.

Treatment

Since the etiologies and presentations of non-cleft velopharyngeal dysfunction vary, formal directives regarding treatment are lacking. The clinical speech-language pathologist must heavily synthesize data and observations collected from assessment to select a treatment approach when appropriate.

In the cleft literature, which is more robust, there continues to be a debate surrounding the clinical indicators that implicate therapy versus additional surgery as the treatment of choice. It is generally accepted that behavioral therapy can be pursued when hypernasality or nasal emissions are specific to certain speech sounds as this indicates a learned hypernasality [10]. While behavioral or functional hypernasality can occur in patients without a history of cleft, these cases are relatively rare, when acquired after speech development. As such, the determination of a patient's appropriateness for speech therapy for non-cleft causes of velopharyngeal dysfunction is influenced by a broader set of factors. This includes the cause and presentation of hypernasality and/or nasal emissions, a global awareness of the patient's overall speech and communicative function and what aspects of these things can be altered, medical history and course, and overall

candidacy and desire for surgery versus prosthetic management.

The clinician treating this population is tasked with determining whether the patient's hypernasality and/or nasal emissions can be eliminated or masked and, if so, whether this will have a significant impact on the patient's intelligibility or communication. In cases where hypernasality is present but cannot be addressed behaviorally, the speech pathologist may divert to an assessment-based care model wherein speech and resonance or swallow outcomes are monitored and documented following interventions from other members of the team. This ongoing monitoring may be helpful to identify residual treatment targets for other specialists. For example, the SLP can aid the prosthodontist in identifying regions to target for VP closure or articulatory contact.

In some cases, alternative speech or communication targets may be selected to aid communication when hypernasality cannot be addressed by the medical/surgical team. When elimination of hypernasality may not be an effective or ideal treatment target, compensatory strategies may help improve overall intelligibility or reduce perception of nasalance. Treatment targets may include softer articulatory gestures to limit the perceived turbulence of nasal emissions or increased oral opening during speech. If a patient has more severe communication or intelligibility issues that are not addressed with speech-based compensatory strategies, augmentative and alternative communication (AAC) may be considered.

For velopharyngeal dysfunction with a presumed behavioral cause or component (mislearning, functional contribution, etc.), attention to redirection of airflow may be helpful. This can be achieved using a finger or cotton ball to monitor airflow, having a patient monitor for nasal air emissions with a straw, or even during nasendoscopy to provide biofeedback.

For VPD that is related to weakness or very mild insufficiency that could potentially be overcome by increased recruitment of pharyngeal constrictors, the use of continuous positive airway pressure (CPAP) has also been suggested as a means of treatment. The rationale for this is that the provision of resistance during speech tasks could promote strengthening of the muscles for velopharyngeal port closure [13]. This has been explored in small samples of patients with a history of persistent hypernasality following cleft repair and some with hypernasality associated with traumatic brain injury [14]. While the rationale for this treatment is interesting, efficacy data is lacking. There is a need for additional research on the effect of CPAP-assisted speech therapy on VP port closure and resonance outcomes and refined guidelines on factors that may predict improvement in resonance with this treatment approach.

Velopharyngeal dysfunction alone is not thought to prevent a functional oropharyngeal swallow [15], yet symptoms of nasal regurgitation may be a meaningful complaint for patients and can detract from the pleasure of oral intake. It is important to verify that that nasal backflow is attributable to limited VP contact or closure, rather than outflow obstruction such as UES dysfunction [16]. When complaints of nasal regurgitation persist, addressing this behaviorally may include postural adjustments, bolus modification, or strengthening other musculature to better compensate for loss of driving pressures from the nasopharynx.

Oral motor exercises often are discussed by clinical care providers as a means of treating VPD but are not supported in the research literature and are currently discouraged by most expert opinion. It should be noted that while there is a paucity of treatment outcomes data for many speech resonance therapy approaches, oral motor exercises such as pushing, pulling, and blowing lack strong theoretical basis as it is not felt that these nonspeech exercises generalize meaningfully to speech-based tasks [17], nor do they align with the principles of exercise physiology that would potentially increase strength [18].

Surgeon Approach

History

The patient is typically referred to the surgeon by the speech and language pathologist if it is felt

that speech therapy techniques alone may not be sufficient for correcting symptomatic VPD. In many instances, children have had a trial of speech therapy to improve concurrent articulation disorders, but hypernasality and nasal air emissions remain. Nonetheless, obtaining a history including prior therapies is important. Surgical history, particularly any prior nasal, palatal, tongue, or oropharyngeal surgeries, is obtained. Tonsillectomy and/or adenoidectomy may frequently be a precipitating factor in the development of non-cleft VPD. A history of recurrent ear infections and myringotomy tubes may lead to suspicion for Eustachian tube dysfunction caused by occult or overt submucous cleft palate. Medical history should include any cardiac, renal, or spine abnormalities and workup of any genetic abnormalities. VPD may be the presenting complaint leading to a subsequent diagnosis of a genetic condition such as 22q11.2 deletion syndrome in 12–37% of patients [19, 20]. Birth and development history should be recorded including history of prematurity and any motor, speech, or cognitive delay. Family history of any speech disorders or other congenital abnormalities should be considered as all of these factors may help to determine the etiology of the VPD, whether structural or functional.

Exam

Complete craniofacial assessment should be undertaken including assessment of the cranial shape and facial features. In children with 22q11.2 deletion, for example, characteristic facial features include malar flatness, hooded eyelids, a broad nasal bridge and tip, and low-set ears with overfolding of the helix [21]. Intraoral examination should include an assessment of dentition and occlusion, presence of tonsillar hypertrophy, and structure of the alveolus and hard palate. A hard palate notch, zona pellucida, or bifid uvula may indicate a submucous cleft palate. The motion of the soft palate may be evaluated for symmetry of movement, strength and quickness of movement, and pattern of elevation. Occult submucous cleft palate may be identified either with phonation or gag when a vaulted V-shaped pattern of elevation is identified instead of the expected curved pattern of elevation in the posterior palate (Fig. 41.1). Neurologic disorder or weakness may also be suspected if there is relatively little movement with phonation or gag.

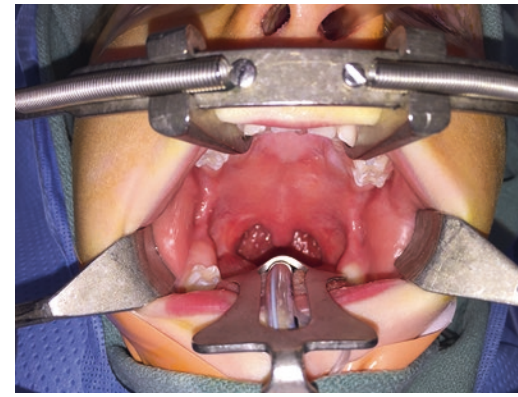

Fig. 41.1 Vaulting V-shaped pattern within the soft palate connotes anterior insertion of the palatal musculature on the hard palate, such as that seen in submucous cleft palate. In some patients this can be seen at rest, as shown here, while in others it is clearly visible with palatal movement in the clinic setting during either phonation or gag

Instrumented Assessment

Nasendoscopy or videofluoroscopy can be extremely helpful in defining the closure pattern and anatomy of the VP port. Often this assessment is critical in determining the appropriateness of surgical therapy. These studies help to delineate whether there is an anatomical abnormality with an associated surgical target vs. a neuromuscular deficiency. Correspondingly, they help the surgeon decide whether a more static or dynamic approach is likely to be successful for the individual patient or whether referral for prosthodontic therapy might be most beneficial. At other times, or in patients who may be difficult to examine in clinic, oral exam under anesthesia may help delineate the specific anatomy. In the OR, medialized carotid arteries may be visible, submucous cleft palate may be detected, or other abnormalities of the palate or velopharyngeal port may be clarified.

Management

Options for surgical treatment of VPD are wide-ranging and target different aspects of the velopharyngeal mechanism. If the velum is deemed to be short, one approach is to lengthen the palate to improve its ability to reach the depths of the posterior oropharynx during speech. Other approaches such as sphincter pharyngoplasty and posterior pharyngeal flap decrease the diameter of the VP port, thereby aiding in closure of the port with speech. Finally, prosthodontic treatment with a speech bulb is another approach for statically addressing VP inadequacy.

Operative Approach

Indications

The indications for surgery for VPD include those patients who have failed correction of their VPI with speech therapy or for whom speech therapy is felt likely to be inadequate. They must be making attempts at verbal communication and be motivated to participate in therapy after surgery. The patient must be deemed medically safe for surgery under general anesthesia on the airway. In children with complex medical problems or history of cardiac anomalies, clearance by their cardiologist or primary care physician is necessary prior to surgery.

Key Aspects of the Consent Process

Risks associated with surgery for VPD must be discussed, including bleeding, infection, and risks of general anesthesia or airway complications. Risks of palatal fistula, or dehiscence of a pharyngeal flap or sphincter pharyngoplasty, are specific to each procedure. Furthermore, all patients and their parents must understand the possibility for incomplete correction of their speech dysfunction and possible need for additional surgical procedures if the primary procedure is insufficient or inadequate. Finally, all VPD procedures that narrow the VP port may increase the risk of developing obstructive sleep apnea (OSA). Development of OSA after surgery may require additional treatments or even revision of the surgery to improve the breathing concerns.

After surgery, most surgeons employ specific dietary restrictions such as a liquid or soft diet for several weeks postoperatively to allow for healing to take place and prevent the risk of dehiscence. Parents must also understand the importance of initiating or returning to speech therapy after the healing time to address any mislearning or articulation concerns and optimize resonance in these patients. In some patients, speech and resonance may continue to improve with speech therapy over the next several years [22, 23].

Equipment

A Dingman mouth gag is used for exposure of the palate and oropharynx. The Hurd elevator can be used to palpate the palate, evaluate the palatal anatomy, and evaluate the adenoid pad along the posterior pharyngeal wall. Good-quality and well-positioned overhead lighting may be used, or a headlight and loupe magnification are helpful to optimize intraoral visualization. Anesthetic solution with epinephrine is typically infiltrated into the tissues prior to incision to help with hemostasis. Standard palatal surgical instruments may be used including long toothed and smooth forceps, long needle drivers, tenotomy dissecting scissors, periosteal elevators, and needle-tip bovie electrocautery. #11, #12, or #15 scalpel blades may be used per surgeon preference. For sphincter pharyngoplasty and posterior pharyngeal flap, often a 12F red rubber catheter is helpful for lifting the uvula and posterior soft palate out of the palatal plane to achieve proper positioning of the flaps. 4-0 Vicryl or chromic sutures are typically used, per the surgeon preference.

Approach 1: Palatal Lengthening

Palatal lengthening is often the first choice for correction of VPI when there is active sphincter movement and a relatively small gap of 5 mm or less. Approaches to lengthen the palate vary. Options include a hard palate pushback (Fig. 41.2) or lengthening of the soft palate with the addition of buccal myomucosal flaps at the junction of the hard and soft palate (Fig. 41.3) [24, 25]. When an overt or occult submucous cleft palate is present, palatal lengthening with a Furlow palatoplasty is often the procedure of choice. This procedure has the advantage of lengthening the palate, narrowing the velopharyngeal sphincter, and creating a functional muscular sling containing the levator veli palatini muscles. Furlow palatoplasty is the author's preferred approach and is described in more detail here.

Furlow palatoplasty includes the creation of two oral and two nasal flaps for a double opposing Z-plasty on each mucosal surface. The anteriorly based flaps on each surface are mucosa-only flaps, and the posteriorly based flaps are myomucosal flaps containing the levator veli palatini muscle, as well as the palatoglossus and palatopharyngeus muscles. Bilateral velar relaxing

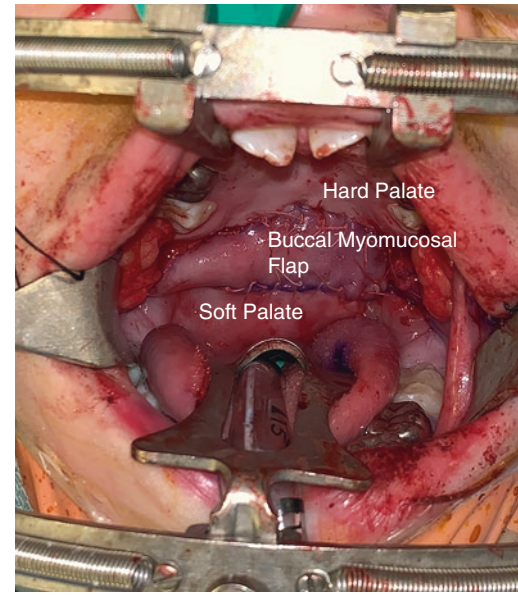

Fig. 41.3 Lengthening of the palate with buccal myomucosal flaps

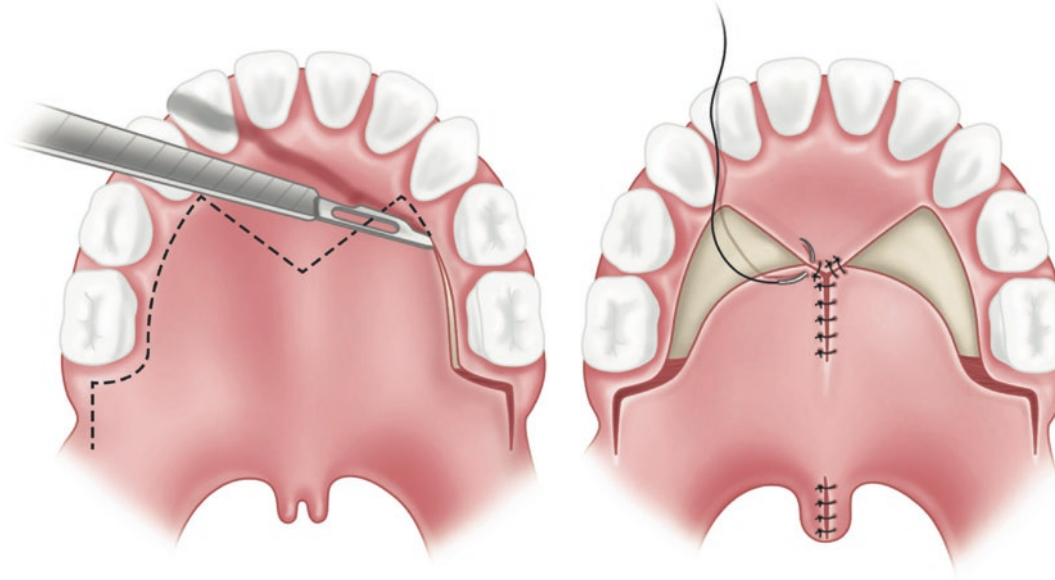

Fig. 41.2 V-Y palate pushback operation for palate lengthening

incisions may be used if necessary to allow for complete release and retropositioning of the flaps. In the non-cleft palate, these incisions are less commonly needed. These relaxing incisions are drawn in the crease formed between the junction of the vertically oriented cheek side walls and the horizontal velar shelves. They may extend from the hard palate around the maxillary tuberosity toward the retromolar trigone of the mandible as needed.

The Z-plasty is then marked at four key points: the junction of the hard and soft palate, the base of the uvula, and the hamulus on each side. In Fig. 41.4, a left-sided posteriorly based oral myomucosal flap and a right-sided anteriorly based oral mucosal flap of between 60 and 90° are marked. The incisions are made through the oral mucosa. The left-sided posteriorly based oral myomucosal flap is then elevated off of the nasal lining. Any abnormal muscle attachments are freed from the posterior edge of the hard palate and tensor veli palatini aponeurosis anteriorly and from the superior pharyngeal constrictor laterally. Releasing these attachments allows the flap to be rotated posteriorly and medially without tension. The right-sided anteriorly based oral mucosal only flap is elevated off of the underlying palatal muscle, which is left attached to the nasal lining (Fig. 41.5a).

After the oral flaps are elevated, the nasal flaps are marked and divided. Here the Z-plasty is designed in the opposite configuration, with a left-sided anteriorly based nasal mucosal flap and a right-sided posteriorly based nasal myomucosal flap. Any muscle on the right side is again freed from its abnormal attachments to the posterior edge of the hard palate, tensor aponeurosis, and superior constrictors. The nasal lining is divided and this flap carries the muscle posteriorly. The left-sided nasal mucosa lining flap is incised, and the right-sided flap can then be inset posteriorly and medially to begin the nasal lining repair of the Z-plasty (Fig. 41.5b). After complete closure of the nasal lining, the oral lining is repaired, mobilizing the left-sided myomucosal flap posteriorly to overlap the muscle of the soft palate to create a functional levator veli palatini muscular sling (Fig. 41.4b, c). Note the change in direction of the flaps after closure with lengthening of the palate. After the palate is well healed, it has increased length and normalized muscular position to reach the posterior pharyngeal wall.

Approach 2: Sphincter Pharyngoplasty

Sphincter pharyngoplasty functions to reduce the transverse diameter of the VP port by medializing the lateral walls. This leaves a smaller central VP port and also augments the posterior wall of the pharynx at the point of velar contact. On nasendoscopy or videofluoroscopy, evidence of poor lateral wall movement with coronal closure pat-

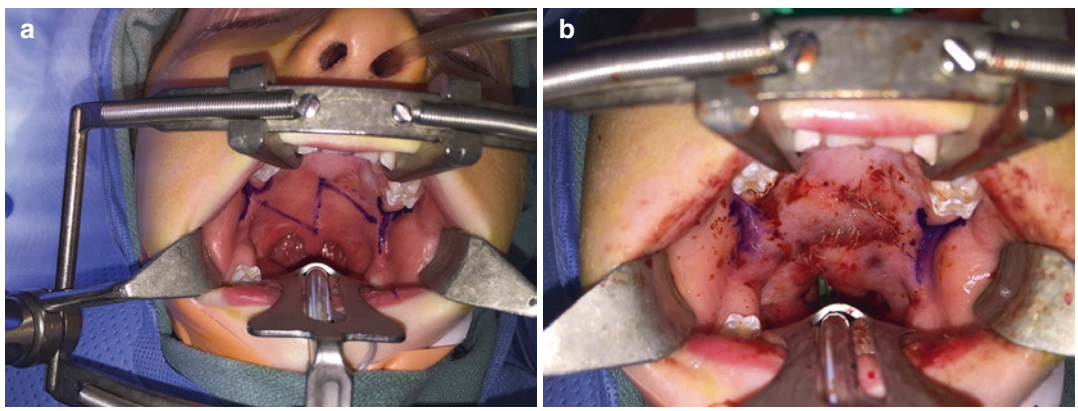

Fig. 41.4 Furlow palatoplasty. (**a**) Markings for oral Z-plasty. (**b**) Completion of repair demonstrating lengthening of the palate

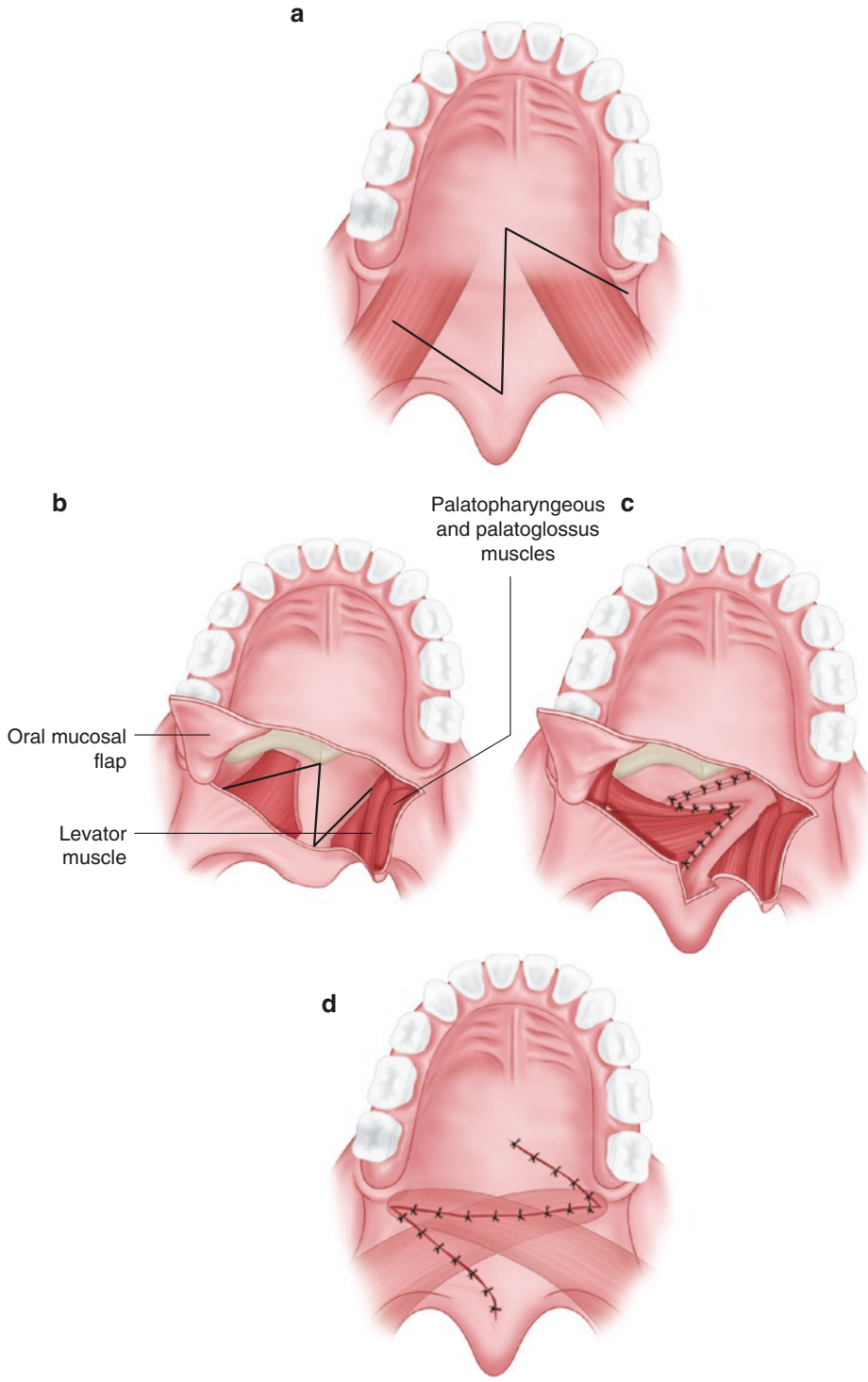

Fig. 41.5 Furlow palatoplasty. (**a**) Z-plasty incision on the oral mucosa. In a submucous cleft palate the muscle may be abnormally oriented longitudinally and insert on the posterior edge of the hard palate. (**b**) Elevation of the oral flaps, with the muscle included in the left-sided posteriorly based flap. Note the nasal Z-plasty is designed in the opposite configuration with the muscle being included in the right-sided posteriorly based flap. (**c**) Nasal Z-plasty after repair with new transverse orientation of the right-sided palatal musculature. (**d**) Oral Z-plasty after repair with overlapping transverse orientation of the muscle on both sides

tern is considered a good indication for sphincter pharyngoplasty. Sphincter pharyngoplasty requires a mobile velum and normal levator orientation to achieve closure in the smaller VP port. Often this procedure may be performed in combination with Furlow palatoplasty to optimize palatal function either in a staged or simultaneous operation [26].

Superiorly based myomucosal flaps are created from the palatopharyngeus muscles of the posterior tonsillar pillar (Fig. 41.6). A key element of this procedure is anchoring these flaps at the appropriate height along the posterior wall of the oropharynx to optimize velar contact. Often this point is slightly above the tubercle of the first cervical vertebra (C1) which can be identified by palpation. After marking these flaps and the transverse incision, the tissues are infiltrated with epinephrine containing solution. The posterior tonsillar pillar flaps are incised and mobilized superiorly, incorporating the palatopharyngeus muscle with its overlying mucosa. A transverse incision is made in the mucosa of the posterior pharyngeal wall. Care is taken to avoid complete transection of the posterior muscle to decrease the risk of inferior migration of the flaps. The myomucosal flaps on both sides are then rotated 90° and anchored to the posterior pharyngeal wall mucosa. The muscle flaps are typically overlapped in a Z pattern to create optimal tightness with a sphincter effect. Finally, the lateral pharyngeal wall donor sites are closed. Postoperatively, if the patient has either persistent VPI or hyponasality and OSA, the limbs of the sphincter may be taken down and either loosened or tighten to adjust the size of the VP port.

Approach 3: Pharyngeal Flap

A pharyngeal flap is commonly performed in patients with a large central gap and/or poor velar mobility or hypotonia. In general, many feel this surgery is optimal in the presence of good lateral wall movement. Some surgeons suggest that a coronal closure pattern is better treated with sphincter pharyngoplasty; however others recommend a wide pharyngeal flap in patients with large gap coronal closure VPI, such as patients with 22q11.2 deletion [27]. This procedure creates a static bridge of tissue extending from the posterior wall of the oropharynx to the velum. A pharyngeal flap therefore creates permanent passive obturation centrally with two lateral ports for nasal airflow and dynamic closure.

The pharyngeal flap may be superiorly or inferiorly based; however superiorly based pharyngeal flaps are most common (Fig. 41.7). The flap is marked with the base at the tubercle of C1 or optimal point of VP contact. The width of the flap may vary depending on lateral pharyngeal wall movement, with wider flaps used when this movement is poor or in the setting of hypotonia. Often the width is between 1/3 and 2/3 of the total width of the pharynx. The flap length is also relative to need, but is typically around 3 cm in length. This flap is infiltrated with epinephrine-containing solution prior to being incised. The incision travels through the palatopharyngeus and superior constrictor muscles, leaving the prevertebral fascia intact. This myomucosal flap is then mobilized completely to its superior extent to prevent any tension on the inset. The donor site muscle and mucosa are closed in a single layer.

The soft palate is split down the middle to allow for inset of the posterior pharyngeal flap. A key element is insetting the flap at the level of the palatal plane to minimize inferior displacement and recurrence of VPI. There are several variations on flap inset. The Hogan modification involves the creation of nasal palatal flaps based along the free edge of the palate to help line the muscular surface of the pharyngeal flap. The flap is inset with multiple sutures to position the flap anteriorly as well as laterally to set the size of the lateral ports. The oral and nasal palatal flaps are then closed over the raw surface of the flap on the lingual side.

Risks of this surgery include hyponasality and obstructive sleep apnea. While OSA may occur in up to 38% of patients in the early postoperative period [28], this resolves by 6 months in most patients to a rate of <10% [28–30]. The lateral

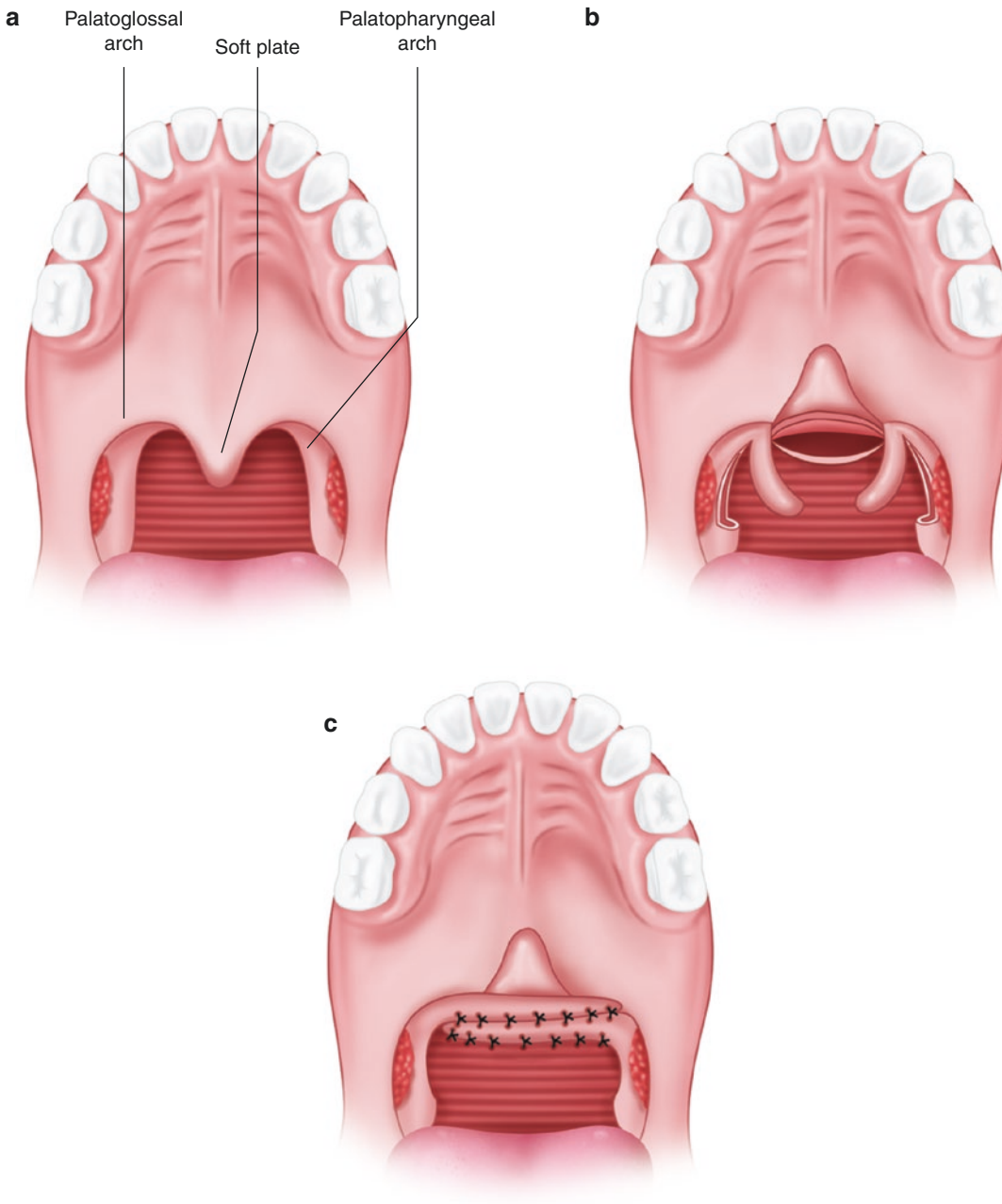

Fig. 41.6 (**a**–**c**) Sphincter pharyngoplasty. (**a**) The mucosa and muscle from the palatopharyngeal arch (posterior tonsillar pillar) is elevated from inferiorly to superiorly. (**b**) The uvula is retracted out of the way to expose the posterior wall of the pharynx. A transverse incision is made in the mucosa. (**c**) The flaps are inset in an overlapping fashion to the back wall of the pharynx to create the sphincter

ports may become scarred or stenotic, increasing the likelihood of OSA or respiratory difficulties. Alternatively, the pharyngeal flap may become tubularized with contractile scarring, leading to narrowing of the flap and suboptimal obturation with persistent VPI. Finally, the flap may dehisce and fall either early in the healing process or late in response to trauma to the area.

Approach 4: Augmentation Pharyngoplasty

Augmentation pharyngoplasty is another option for treatment of minimal VPI or in patients in whom pharyngeal flap or sphincter pharyngoplasty may be contraindicated due to comorbidities. Most commonly, this approach is felt to benefit patients with a small central gap of less than 3–4 mm or with touch closure of the VP port on nasopharyngoscopy or videofluoroscopy. Some authors have suggested this may be appropriate for those children with VPI after adenoidectomy. Multiple different substances have been described for use in augmentation pharyngoplasty including silicone, cartilage, collagen, fat, fascia, acellular dermal matrix (ADM), and calcium hydroxyapatite (CAHA) [31, 32]. Depending on the substance used, risks may include infection, extrusion, absorption, or migration of the substance. Fat and CAHA appear to be most widely studied for this purpose [31–36].

In terms of the location of injection, various sites have been described. In the posterior pharyngeal wall, the substance is ideally placed into the retropharyngeal space just deep to the superior constrictor muscle and superficial to the pharyngobasilar fascia. Fat is typically over-injected by 30–50% to offset anticipated fat resorption. With any substance, multiple injections may be necessary to achieve adequate impact on VP closure. Other authors advocate targeting fat grafting to the uvula to allow for improved closure when touch closure is present [33]. Some studies have evaluated injection in the posterior pharyngeal wall, soft palate, and pharyngeal arches [34].

Augmentation pharyngoplasty demonstrates safety and success in the literature [31, 32], but the indications remain limited. All studies to date contain relatively small numbers of patients, are retrospective in nature, and have limited follow-up. The necessary volume needed for injection varies from a mean of 5–6 ml of fat in some studies to 11–13 ml in others. Injection sites vary between centers, and the technique lacks consensus on indications. Outcomes may vary depending on the indication, with some suggesting fat grafting may be better as a secondary procedure for VPI following another

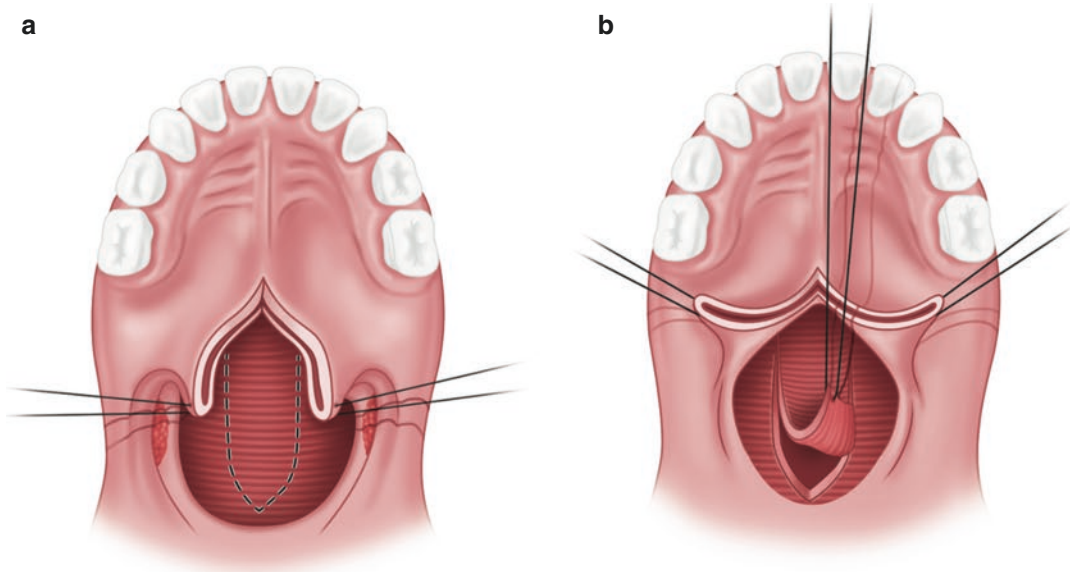

Fig. 41.7 (**a**–**e**) Pharyngeal flap. (**a**) The soft palate is split and a superiorly based flap is designed on the posterior wall of the pharynx. (**b**) The pharyngeal flap is elevated from inferiorly to superiorly and (**c**) inset into the soft palate nasal lining. (**d**) The pharyngeal wall donor site may be closed and the uvula and oral side of the palate are closed over the pharyngeal flap (**e**)

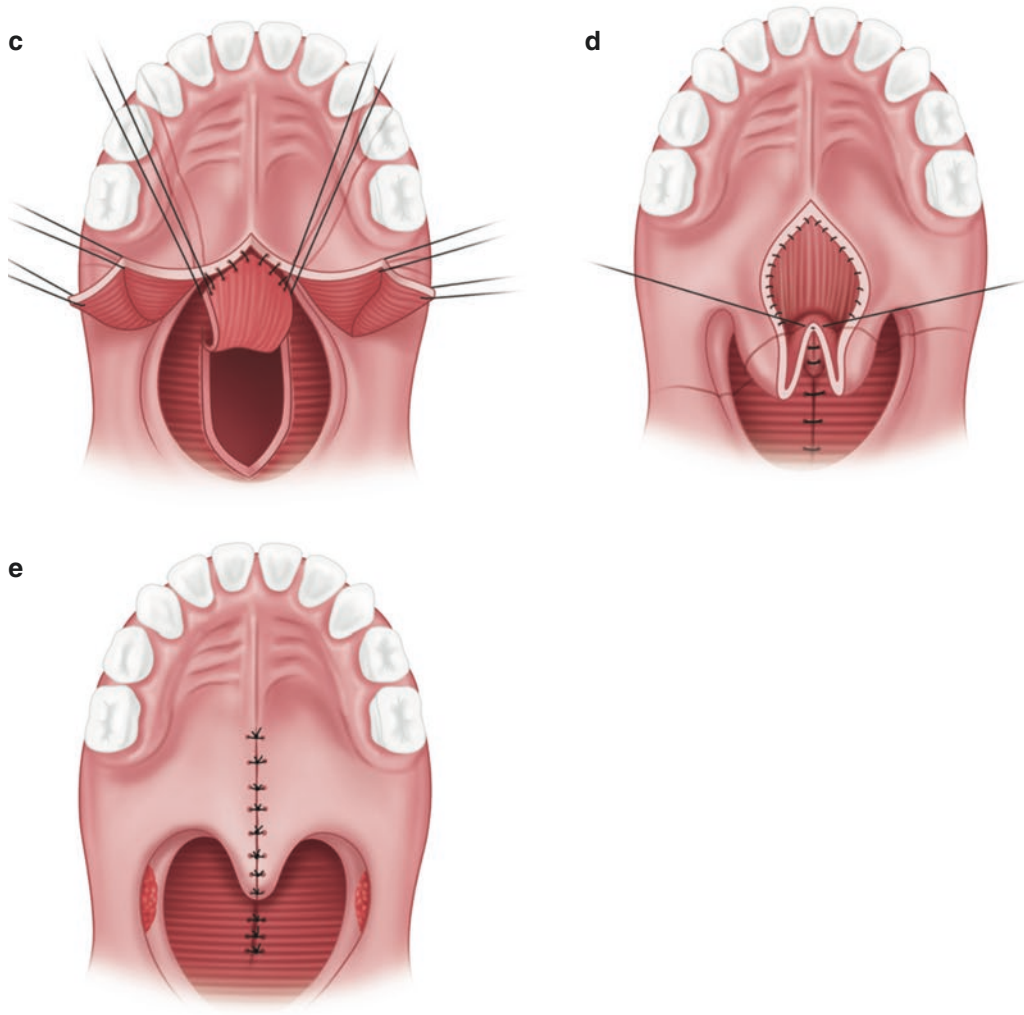

Fig. 41.7 (continued)

primary treatment [37]. Long-term data on the use of this approach in children is lacking at present. Fat hypertrophy may increase the risk of OSA as children age and gain weight. In one reported case of fat hypertrophy, the patient subsequently required two debulking procedures [38]. Finally, in children with 22q11 deletion, the surgeon must be aware of the abnormal course of the carotid arteries in the pharyngeal space to ensure intra-arterial injection is avoided as a potentially devastating complication. An unpublished report of MCA infarct due to presumed fat embolism has been reported, and autologous fat grafting has other known risks in the head and neck including blindness and cerebrovascular accident [31]. Avoidance of injection into the lateral pharyngeal walls has been recommended to minimize the risk of intravascular fat injection.

Comparison of Surgical Approaches

In general, the literature on the use of these different surgical approaches for the treatment of VPI is diverse, without consistent conclusions, and lacks high-level evidence to support one procedure over any other in terms of indications or speech outcomes [39]. In general, Furlow palato-

plasty is thought to be less likely to lead to nasopharyngeal obstruction or OSA. No consensus exists regarding the increased risk of OSA between sphincter pharyngoplasty or pharyngeal flap. The majority of studies on VPI in children are on patients with cleft palate, and this population may fundamentally differ from patients with non-cleft VPI as discussed in this chapter. Nasopharyngoscopy or videofluoroscopy to evaluate closure pattern has been traditionally used to help determine the appropriate surgical approach. Some authors have recently called this into question, suggesting that coronal closure patterns traditionally thought to be best treated with sphincter pharyngoplasty may actually be better treated with pharyngeal flap [27].

Postoperative Management and Follow-Up

After any of these surgical approaches for VPI, the patient is typically admitted to the hospital for overnight monitoring with continuous pulse oximetry given the risk of swelling and airway obstruction. Antibiotics to cover for oral flora may be used for a short duration, and many surgeons will keep patients on a liquid and soft food diet restriction while the palate is healing for 2–6 weeks. Patients will typically return to speech therapy approximately 6–12 weeks after surgery to begin working on their compensatory articulation errors. Speech therapy is essential for the patient to learn how to effectively use the new anatomy. Many centers perform a formal speech assessment between 3 and 12 months postoperatively to assess progress with VPI following surgery.

Finally, any postoperative protocol must include screening for and testing for obstructive sleep apnea (OSA) in the long term. When OSA is identified in the early postoperative period, it may be related to tissue swelling, and observation may be adequate. When persistent, it may be treated with CPAP. If severe, or persistent despite other optimal medical management and CPAP management, surgery to decrease the amount of obstruction may be necessary. For pharyngeal flap, takedown of the flap has been shown to often have the benefit of preserved speech function, possibly due to the bulk of tissue being maintained with the velum [40].

Prosthodontist Approach

Nonsurgical treatment of VPI may include a palatal lift or speech bulb. This is most commonly used in patients in whom surgery may be contraindicated, on a trial basis, or per patient preference. These devices are often most helpful in cases of velopharyngeal incompetence in which neuromuscular function is limited. When the palate is hypomobile, poorly coordinated, or paralyzed, the surgical treatments may be less effective. These devices are made for an individual patient by a maxillofacial prosthodontist and anchor to the maxillary teeth. Similar to a retainer, these devices have a posterior extension that statically elevates the soft palate upward to narrow the velopharyngeal space. When the tissue is inadequate, the prosthesis may extend beyond the limit of the soft palate to optimize velopharyngeal closure.

22q11.2 Deletion Syndrome

Children with 22q11 deletion syndrome make up a unique subset of patients with non-cleft velopharyngeal insufficiency. Approximately 70% of children with 22q11.2 deletion have speech delay or velopharyngeal insufficiency. Children with 22q11.2 deletion may have an overt cleft palate, overt or occult submucous cleft palate, or no cleft at all. They commonly have a short or atonic velum and a deep cavum leading to anatomic discrepancy creating incomplete VP closure [41]. This subset of children can present with a range of speech and language concerns including expressive language delay, cognitive delay, and variable speech and articulation patterns, making their management more challenging for both the speech and language pathologist and the surgeon.

Of particular concern to the surgeon, operating on these children is medialization of the internal carotid arteries in patients with 22q11.2

deletion. In one study, the carotid arteries were found to be medialized in 25% of children undergoing imaging [42]. The surgeon should evaluate for pulsations along the posterior pharyngeal wall during nasendoscopy and also at the time of surgical intervention. Some surgeons routinely perform preoperative imaging (either CT angiogram or MR angiogram) in this patient population to determine the course of the carotid vessels. Others argue that the injury rate remains very low and imaging is not cost effective [43].

Surgical treatment for VPI in children with 22q11.2 has demonstrated a lower success rate in multiple studies when compared to children without this diagnosis [44–47]. Pharyngeal flap is reported to have between 85% and 100% success rate [44–46] vs. 78% success rate in sphincter pharyngoplasty [47]. The effectiveness of Furlow palatoplasty in achieving normal resonance in children with 22q11.2 and a submucous cleft palate is reported between 45% and 74% [23, 46]. In general, the revision rate is higher in children with 22q11.2 deletion than nonsyndromic children, between 3% and 22% depending on the study. Furlow palatoplasty is associated with a higher risk of secondary surgery, but careful patient selection leads to ultimately equivalent rates of achieving normal resonance [23]. Given the lower risk of obstructive sleep apnea with Furlow palatoplasty, we suggest that Furlow palatoplasty may be selectively employed in children with 22q11.2 deletion and VPI, a kinetic submucous cleft palate, and a relatively smaller defect with at least 70% closure on imaging. Those children with larger gaps are recommended to undergo pharyngeal flap surgery [23].

After surgical treatment, children with 22q11.2 deletion syndrome consistently require a much longer time after surgery in speech therapy to achieve normal resonance [23, 48, 49]. Compared to non-syndromic children, who require a median of 8 months to achieve normal resonance, children with 22q11.2 deletion take a median of nearly 3 years to achieve normal resonance after surgery [23]. This highlights the importance of ongoing intensive speech therapy for longer periods in this patient population and the need for patience on the parts of the surgeon, therapist, patient, and family in dealing with this unique population of children with non-cleft velopharyngeal insufficiency.

Appendix 1

Suggested stimuli for non-cleft assessment of resonance and velopharyngeal function.

To test for hypernasality

and nasal emission, consider a sample of syllable repetitions using CV combinations with high pressure sounds to test for nasal emissions and a combination of high and low vowels to assess hypernasality:

Pa pa pa…pi pi pi

Ta ta ta…ti ti ti
Ka ka ka…ki ki ki
Sa sa sa…si si si
Sha sha sha…shi shi shi

Longer passages with high pressure sounds and no nasal sounds:

The Zoo Passage (Fletcher, 1972)
Look at this book with us.
It's a story about a zoo. That is where
Bears go. Today it is very cold out of
doors. But we see a cloud over head
that's a pretty white fluffy shape.

Additionally/alternatively:

- Counting from 60 to 70 will produce many repetitions of high pressure sounds with few nasal sounds

Sentences with nasal sounds (Mckay-Kummer SNAP test, 1994)

Mama made some lemon jam.
Ten men came in when Jane rang.
Dan's gang changed my mind.
Ben can't plan on a lengthy rain.
Amanda came from Bounding Maine

Additionally/alternatively:

- Counting from 90 to 99

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The Young Aspiring Singer

42

Debra Jean Phyland and Neil A. Vallance

Overview

Singing is a basic human behaviour, and children are well known to imitate and play with pitch and resonance and to produce song from an early age. The value of singing has been regularly described and is strongly linked to communication, education, play, musicality, cultural cohesion, emotional wellbeing, and various other aspects of children's development [1–4]. As a child matures, this can burgeon into a desire or expectation to participate in group singing activities and also to perform in front of others as a soloist or chorister which introduces an array of potential psychological sequelae and vocal demands (Fig. 42.1). For some children, singing may be related to a musical production and simultaneously involve movement or dance, and, for others, the performance may be associated with singing with a band incorporating the use of microphones and sound systems. The performance context and music genre will have bearing on the nature and physicality of the child's voice production as well as their performance confidence, exposure to judgment, artistry, and self-esteem. An appreciation of these aspects and the possible short-term and developmental implications for the young aspiring singer is important when assessing and managing the child's vocal health and overall wellbeing.

Fig. 42.1 Parents influence their child's performance in many ways. (Original illustration by Joey Phyland)

Young Singers in the Professional Entertainment Industry

With the global success of television talent shows such as *The Voice, Idol,* and *Got Talent* and the explosion of social media in recent times, the performing child as 'star' has become increasingly

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J. S. McMurray et al. (eds.), *Multidisciplinary Management of Pediatric Voice and Swallowing Disorders*, https://doi.org/10.1007/978-3-030-26191-7_42

more celebrated [5]. Similarly, in the music theatre genre, many of the shows introduced this century such as *Billy Elliot*, *Matilda,* and *School of Rock* feature children as central to the plot and can even involve a larger number of children than adults in the cast. Along with evergreen shows such as *Oliver! and Annie*, these professional productions now abound worldwide, and audiences embrace the child as a key performer in the story-telling. Perhaps as a direct reflection of the increased prevalence of children performing in these various contexts, we have also witnessed a boom of commercial enterprises such as talent training and performing arts schools focusing on both the child and adolescent performer.

Although it is beyond the scope of this chapter to muse on the potential benefits and downsides of children working in the entertainment industry, it is important to recognise there can be psychosocial, logistic, and developmental implications. Workplace regulations and child employment laws vary enormously across and within countries [6, 7], but performer children may work long shift hours, be highly scheduled, become dependent on external validation, suffer fatigue, experience performance anxiety, and also miss out on childhood and educational experiences for production seasons that can last for months and even years. Of course, on the other hand, children can thrive in this environment, develop a stronger sense of self-esteem and social inclusion, and be well rewarded financially and psychologically for their hard work and talents. When clinically assessing and managing these children's voices, it is prudent to consider these performance-related factors and their potential impact on the vocal apparatus.

Other Singing Contexts

Young choirs, glee clubs, and student musical productions also abound in schools and in local community as part of our contemporary culture. For some children, singing is a compulsory school activity, whereas others may seek involvement and performance opportunities in school or external contexts such as cathedral and worship choirs, community theatre, busking, concerts, cultural events, and the like. They may sing with specific or mixed age groups and genders, with several different groups in school and with outside organisations, and with repertoire that may or may not be age-appropriate or easily achievable in terms of their vocal range, pitch, loudness, and style. In our experience, young singers may try to emulate adult voice characteristics or be placed or maintain singing within a vocal range or voice category that does not reflect their potentially changing tessitura (e.g. a transitioning boy soprano continuing to sing treble when undergoing mutation). The child singing in the school concert or musical production may be regularly instructed to sing louder or required to 'binge' sing during technical week, without adequate preparation for vocal fitness. For some, the vocal load can be extreme, and the vocal techniques adopted to achieve the vocal demands may be maladaptive and potentially injurious to vocal health. For other children, their natural singing capabilities and competencies are extended by positive vocal experiences along with increasing maturity – it is these children that perhaps achieve greater singing success for reasons other than simply 'survival of the fittest'.

Influence of Singing on Vocal Development and Health

Relative to the fully developed adult voice, there is surprisingly little known about the effects of singing on the developing larynx. Although several studies have demonstrated the positive effects of regular singing and individual voice training on vocal efficiency and health among children [8–11], whether this translates to a reduction in risk of voice disorders has also not been previously well investigated. Of the few studies available, most relate to children engaged in choral singing. Rather than espousing the positive benefits of choral singing, several authors suggest child choristers experience more symptoms of voice problems than non-singers or soloists, particularly if undertrained, due to factors associated with choral singing such as increased

vocal load, reduced auditory feedback, and possible competitive effects causing vocal strain or misuse [10–13]. Whether these are only short-term or have a cumulative negative effect is not established. More recently, however, Clarós et al. [14] reported on 1544 children (half of whom were singers) and found the opposite – children singing in a choir were significantly less likely to be diagnosed with voice problems [14]. Whether this is also the case with soloists and young singers within other genres such as musicals and other contexts requires further investigation.

The manner or style of singing may also influence the nature and amount of impact sustained by vocal fold tissue and effects on neuromuscular function and should be an important consideration in the estimation of the vocal load demands among singers. Along with the duration of voicing, singing activity can vary enormously in terms of loudness and pitch parameters and also in other vibratory characteristics such as glottic closure durations and patterns, phonatory onsets, and aerodynamic aspects which all contribute to their vocal load [15–18]. In the young performer, the potential long-term effects are not known of the current tendency in popular music for children to sing in a predominantly modal register up to high frequencies or to regularly use a 'belt-like' vocal posture. Such laryngeal postures typically involve high vocal intensities and subglottic pressures, increased glottic closure times, high vocal effort, and hence heavy vocal load [15]. Since the vocal folds of children are structurally smaller, they are arguably experiencing greater vocal tissue load than the adult singing equivalents when singing the same repertoire. Whether these vocal choices may influence the short-term and long-term health of the not yet fully developed instrument has not been established. It is also not clear whether there are age-related differences in the ability of the vocal fold lamina propria and muscles to withstand or recover from heavy vocal load [19–21]. For a more comprehensive explanation of vocal fold structure and morphology in children and the potential impact of heavy vocal load, the reader is directed to Chaps. 8 and 9.

There are inherent difficulties in establishing whether singing in childhood poses a risk to vocal health since there are so many potential confounders. By nature, children who sing are more likely to be gregarious [22], and therefore it is difficult to ascertain whether their personality and potentially associated heavy speaking voice load are influencing overall vocal health. Another issue is that paediatric singers may be overrepresented in treatment-seeking clinical settings compared to non-singing children due to differences in the importance attached to their voice and with their vocal needs. As previously mentioned, however, there is an overall need for an increased understanding of the potential physiologic and functional impact of sustained vocal load on children's vocal fold development. The perceived trend towards more young people aspiring to be performers also provides a wealth of opportunities for increasing our understanding of the impact of performance (and training) load on the developing larynx and potential for injury and the development of pathology.

Influences of Vocal Health on the Singing Experience

Good vocal health will obviously contribute to optimal singing competence and capability and therefore successful singing experiences. There are also other potential benefits of this, in that children who sing are more likely to have a positive self-concept and sense of being socially included [23]. Conversely, it would seem logical that children with compromised vocal function or voice disorders may be less likely to engage in singing or to enjoy the singing experience which may lead to negative consequences such as frustration, lower self-esteem, and reduced social participation. Both these arguments provide a strong rationale for maximising vocal health in children in order to provide the option of singing participation and optimal performance.

Influences on Singing Voice Health

Differences in vocal function and vocal health of children according to age, gender, voice use

patterns, training, and various medical factors have been well described elsewhere in this textbook, but it is of interest to further explore these variables in relation to their singing voice relevance.

Age and Gender

The age of a child will influence the expected pitch and loudness ranges, voice quality, and resonance characteristics of the singing voice as these dynamics are mostly dictated by the size, shape, and nature of the vocal tract structure and the development of increasing neuromuscular control. Physical changes occur throughout the respiratory, laryngeal, and resonatory systems so there are differences across developmental stage in power, source, and filter aspects of vocal function. Specifically, with advancing age, there are increases in breathing capacity, changes in vocal fold structure, increases in neck length and width and relative descent of the larynx, and subsequent enlargement of the vocal tract and resonatory system. There is also growth of the paranasal sinuses and nasal turbinates with atrophy of the tonsils and the adenoids, thereby creating more resonance space. When singing through childhood, it is therefore important to a child's optimal vocal health that the repertoire and vocal demands are commensurate with the developmental stage and capabilities of the child in terms of respiration, voice, and resonance [4, 24–28].

Prior to puberty, the singing voices of boys and girls are similar in terms of their pitch range and vocal qualities, in keeping with the vocal tract structures being relatively the same shape and size [24]. Differences between the genders across childhood in singing activity have been suggested, however, with boys being less likely than girls to participate in singing activities, especially at school [28]. This may no longer be the case perhaps due to social and cultural shifts, and indeed gender representation in the childhood entertainment industry would now seem relatively even.

The onset of adolescent voice change particularly for males can be dramatic and rapid or a gradual and relatively undetected process. Neither age nor the onset of puberty seems to be the best indicator of the advent of voice changes [4], but the mean average onset is suggested to occur between 10 and 12 years [29, 30], and the peak of pubertal voice changes around 12–14 years of age in both females and males [29–38]. The Cooksey six-stage classification of pubescent voice change is based on singing range and tessitura and can be useful in tracking singing alterations across puberty [12, 29–38]. Some male singers can pass through all these musical stages of adolescent voice change in 12 months, but it is also possible for this process to be much slower and to last several years. Singing dynamics will be affected by both the hormonal and psychological influences of puberty, and it is sometimes difficult to tease out the relative contribution of each to the pubescent singer's vocal profile. It is also worth noting that, in our experience, it is not only the voice quality and pitch that change but often there are subtle resonance changes too that yield a richer timbre and signal adolescence is nigh.

The prediction as to when a boy's voice is likely to start breaking is therefore not an exact science, but some sense of the timing of this is desired when casting young male singers in music theatre productions. Some seasons of the shows featuring prepubescent male roles, such as the roles of Bruce in *Matilda,* Billy and Michael in *Billy Elliot*, and Oliver and Artful Dodger in *Oliver!*, may span over 2 years from audition to end of the contract. Producers and creatives invest much in these performers and understandably wish to avoid cast changes but, in addition to changes in physical attributes (such as height and weight), declining ability to achieve the vocal demands of the role due to puberty invariably leads to compromised performances or a cessation of the pubescent child playing these roles. In our experience, young male performers are often acutely aware of their vulnerability and can become highly anxious about any voice issues in case they are symptomatic of mutation. We have also noticed, although typically in a less obvious way, that the prepubescent female singer can similarly alter for some weeks or months around the time of the menarche, becoming lower in modal pitch and variable in quality, more prone to vocal fatigue and fluctuating vocal fold oedema.

There is some conjecture within the scientific literature that singers may differ from non-singing children in their ability to control and extend some aspects of their singing dynamics, such as vocal quality, pitch, and loudness ranges, and thereby exceed age-related physical constraints [4, 12, 30, 31]. This is attributed to singing training, singing experience, and perhaps inherently superior singing competency or talent [9, 23]. Although purely anecdotal, it is our experience that, although children's vocal folds can be very resilient and can demonstrate improved efficient voicing for singing, repeated overextension beyond their comfortable physical limits in singing can be problematic for a child's short-term vocal health. Whether this is potentially injurious to long-term vocal health has not been ascertained, but common sense suggests that clinicians and vocal pedagogues should be strong proponents of children singing efficiently within their current comfortable range and adapting repertoire accordingly.

The Team Approach

The concept of team management within all aspects of performance medicine has been increasingly favoured [39] and is particularly cogent for young performers. Our experience working together over the past 26 years in a Voice Clinic with a strong singer focus has reinforced the value of a team approach, with each member offering specific and complementary expertise to optimise patient care. An expert understanding of singers' needs, performance, and vocal demands and of vocal pedagogy is required within the team composition, in addition to the usual clinical skillset described in Chap. 2.

In our experience, there can be additional complexities and frequently emotionally-laden stakes associated with the vocal care of the child performer that distinguish them from non-singing children. Managing both the singer and parental concerns can be challenging, and there is often an imperative to meet performance expectations and requirements above other priorities, including medical recommendations. Parents may be unintentionally providing an array of alternative remedies that are confounding recovery or adding medical complexity with multiple visits to various specialists. In addition, the children themselves may be reluctant participants in the treatment-seeking process and highly anxious about the potential ramifications of vocal care advice. For example, a recommendation for a reduction in vocal load may be the difference between performing or not. Similarly, the suggestion to sing in the alto rather than treble group in the choir may jeopardise the potential for any involvement, for solo opportunities or reduce singing satisfaction.

We have also seen countless young singers who are concerned they may have vocal nodules (colloquially coined 'nodes') and can even seem somewhat comparatively relieved when they are diagnosed with another pathology. In popular culture, vocal nodules have become associated with catastrophic performance consequences, as best described in the clip *Chloe tells The Bellas that she has nodes* from the much celebrated movie *Pitch Perfect*. When asked to explain her diagnosed 'nodes', Chloe responds, 'They sit on your windpipe and crush your dreams' (http://pitchperfectmovie.com). This concords with our experience that a diagnosis of nodules is commonly perceived as a dire outcome for young singers, despite the findings that children's voices can fluctuate significantly and also that well-established nodules among paediatric singers of both genders and in post-pubescent males are extremely rare. Such potential preconceptions and the extra layers of importance attached to vocal health for this population highlight the necessity for a considered and sensitive team approach to the clinical assessment and management of the young singer. There is also a frequent need for the team to de-catastrophize the clinical findings for both the child and parent/s and to be cognisant of the recent vocal load of the young singer, in order to account for acute rather than chronic issues, and not 'over-call' pathology. The overall message must be one of vocal survival and success, and indeed, in our experience, with appropriately targeted care, there is almost never a need to stop a child from singing, unless recovering from sickness or surgery, and from pursuing their performance goals.

Speech Pathologist Approach

Assessment

A comprehensive voice assessment is routinely indicated for all children. For the singer, the case history information will also revolve around the singing voice activities, vocal needs, singing voice symptoms, singing training, and performance expectations (see Table 42.1). In particular, it is essential to ascertain the child's current vocal load for both speaking and singing across a week. It is useful to hear the child's perspectives on these aspects, as well as the parents', to gauge congruence in the level of concern, motivation, singing goals, and the description of the voice problem and how it relates to singing activity. In our setting, the singing teacher is often the person who initiates the referral, due to concern about a child's voice, which the child or their parents may not have previously noticed, so it is highly useful to have the singing teacher's report of their impressions too. Problems with the singing voice may not be audible or obvious in connected speech tasks and there are also singing-related aspects that need to be evaluated, so it is useful to

Table 42.1 Special considerations in the assessment of the singing child

Singer factors	Examples of areas to consider in assessment and management
Age and gender	Relevance to vocal fold development and layered structure
	Pre- or pubescent
	Relevance to timing of vocal training commencement
Personality	Extroverted (gregarious)
	Confidence and self-esteem
	Anxiety
Singing activity	Singing performance (and rehearsals)
	1. Performances, rehearsals, practice, general singing at home
	2. Solo or ensemble
	3. Repertoire-pitch, loudness, and quality ranges (tessitura)
	4. Genre-singing style/s required, certain characters with certain voicing requirements
	5. Duration of each session and of singing time within these
	6. Number of sessions per week
	7. Context (e.g. with a band, on stage, in studio, church)
	8. Additional simultaneous activities (e.g. dancing, acting, playing a musical instrument, etc.)
Singing confidence	Own understanding of vocal range and limitations
	Whether educated on changing voice during adolescence
	Mismatch between singing confidence and set singing task
Demands of performance	Performance expectations and stakes
	Singing within competitive environments
	Impact of performing on schooling and lifestyle effects
	Parental expectations
	Key personnel influencing nature of singing activity
Vocal load	1. Singing load
	2. Speaking voice use patterns – context, amount, type, environments
	3. Overall vocal load intensity, spacing, and recovery
Singing training	1. Individual or group
	2. How many teachers involved in past (and now!)
	3. Frequency and duration of individual lessons
	4. Teacher approach, method, and/or favoured genre (classical, music theatre, contemporary, rock, jazz, choral, etc.)
Individual	Talent, competency and capability
	Singing awareness and knowledge
	Aspirations and drive

hear the child sing on specific vocal tasks (such as interval glides, vocal runs, and sustained notes) and also in song. Context, however, is everything – the demonstration the child gives in the clinical setting may not reflect their usual manner of voice production at home practising or in performance!

Perceptual evaluation of the singing voice is assessed similarly to the speaking mode and includes the features of breathing for singing (such as mode, adequacy, timing, duration, and presence of audible inhalations), voice quality (including breathiness, strain, roughness, vibrato), loudness and pitch (ranges and control), and resonance. It is helpful also to listen to the voicing onsets and offsets, transitions across pitches (obvious register shifts or discrepancy in quality between low notes and high), length of breath phrases, and variations or inconsistences in voice quality, in order to get an overall impression of the vocal efficiency achieved in singing voice production. Whether the voice quality and resonance characteristics produced in singing matches the speaking voice production and whether the voice sounds 'authentic' or appropriate for the child's age and capabilities are also worth noting. Finally, a general impression of confidence, degree of effort or of the vocal ease with which the child sings, can be yielded in the perceptual evaluation.

In our experience, the most common technical issues or symptoms encountered in young singers related to singing voice function are breathy or pressed singing voice quality, reduced vocal range (pitch and loudness), overly hard or overly aspirated glottal attacks on notes with vowel onsets, an obvious discrepancy between a rich and loud (chest/modal register) voice on the lower pitched notes, and a soft and breathy quality in the higher registers, audible inhalations, and reduced subglottic control of loudness and pitch in favour of laryngeal hyperfunction. Whether these features are symptomatic of habitual or recent inefficient voice production for singing, vocal fatigue, or signs of laryngeal pathology may require some teasing out. If the overall picture is one of a relatively recent deterioration in the voice, the voice problem is most commonly related to a heavy vocal load and usually resolvable with a reduction in load. If it is perceived to coincide with hormonal changes, teaching children about vocal physiology and anatomical changes during puberty may assist in their understanding of their own vocal abilities and limitations. For both scenarios, it is crucial to normalise rather than pathologise the voice concerns and reassure that the singer will be able to continue singing well once the short-term issues are addressed.

Management

Voice therapy for the singing child is not different in essence compared to that for the non-singer except that the functional focus may be more on the singing voice, although it is preferable for the child to understand they have one voice, and so the same principles apply to both modes. Specific exercises and approaches that have proven to us to be the most favoured and effective are promotion of simultaneous breath with phonation onsets (rather than hard attacks or breathy onsets), use of semi-occlusive vocal tract exercises (especially straw 'voice-bubbling'), subglottic control of loudness and pitch (akin to or a variation of the Accent Method), and establishment of best resonance and quality ('sweet spots') on sustained notes and interval 'slides and glides' [40–43].

An important consideration throughout the assessment and treatment process is that these singers are child, not adult, performers (although a child may often think he or she is meant to sing as an adult). Our primary tenets are that children should sound like children and aim to be the best singers they can be within their current physical and functional vocal capacity and we recommend these principles are considered foremost when working with young performers. When experiencing voice difficulties, it is recommended that the therapy be focused on extending and building upon what the child can do well to enhance their vocal efficiency, flexibility, and endurance. Whether it is within the scope of the voice-oriented speech pathologist to also treat the artistry is arguable as it is a fine line between treating and teaching voice. We strongly recommend

recognition of our professional competencies and limitations in this space and of the specialised skills and experience of our singing teacher colleagues [44]. As many singing children may not be receiving individual or appropriate singing training, it may be necessary to facilitate transfer of the skills gleaned by voice therapy into the singing performance context. If so, this must be undertaken with care, transparency, and endorsement so that all those involved with the child's singing activities can work together as a team to avoid confusion and conflict for the child and to optimise artistic success.

With the young singer, depending of course on the underlying reason and motivation for seeking help and the nature of the voice problem, therapy is usually, and should be, fast-moving and rewarding for both the child and clinician. The child is more likely than their non-singing cohort to have a good understanding and mastery of their vocal instrument and be more confident changing laryngeal postures, imitating sounds and participating in vocal play or singing. The child must be able to achieve improved voice production independently and to perceive quick progress in the singing voice output and ease of production in order to maintain motivation and continue with therapy. Avoid prolonged and frequent therapy intervention, being overly scientific, too methodical, or measurement-oriented with young singers, or you run the risk of killing the joy of singing for them. Singing provides a form of artistic expression, at the very least, and our primary role is to help them flourish as vocalists and get back to it – not to become scientists.

Otolaryngologist Role

The late 1970s saw significant advances in the diagnosis and management of voice disorders and the development of laryngology and voice as a subspecialty in otolaryngology. This is especially so in adult medicine but not so clear in the paediatric population. Specialist paediatric otolaryngologists are small in number, and of these, very few could claim specialist skills in the larynx and voice. This has potentially caused the development of paediatric laryngology and voice science to lag significantly behind the gains made in these areas for adults.

Most investigative methodologies are adult-oriented and not easily translatable to a child, especially an anxious or uncooperative one. It is important for the otolaryngologist to therefore maintain a very open mind and be flexible in assessing, examining, and investigating the child with a voice disorder. In addition, the laryngologist must act primarily as physician more than surgeon, taking a holistic approach and focusing on the medical needs, in addition to the vocal issues at hand. The usual mainstays of history, examination, and investigation should be adhered to. Proper treatment relies on accurate diagnosis. Accurate diagnosis and treatment should be very much considered a team event and in particular with the otolaryngologist and speech pathologist clinicians working with the family, singing teacher, and other parties as found necessary from time to time.

History

Parents will clearly play a significant role in the history-taking process, and it is not unusual for a singing teacher to be present as well. It is important to include general health issues as well as voice issues. Large tonsils and adenoids, for example, can impart a significant hyponasal resonance pattern and also reduce loudness in voice production, so many young singers will become hyperfunctional in their laryngeal effort to compensate. Inflammatory tonsil disease whilst being generally disabling can also create an infective environment leading to an inflammatory laryngeal disorder. Asthma and steroid-based preventative sprays are also important factors to consider in terms of their potential impact on respiratory and vocal health, as are allergy and allergic rhinitis, upper respiratory obstruction, and sleep-disordered breathing. It is also useful to determine if the child was known to experience significant reflux as an infant. If so, consider that this may not have resolved and still be present but induce symptoms to which the child has become accustomed.

Parents of performers are often understandably highly concerned about their child's vocal health, sometimes being more anxious than the child. In trying to help their singing child's vocal recovery, they can be overly zealous in providing various remedies, and it is important to explore the nature of these well-intentioned interventions during the consultation. It is also important to recognise that performance anxiety among child singers is common and to appreciate the potential for this to be playing a part in the symptom presentation and description.

Examination

As described elsewhere in this textbook, a general head and neck examination is essential. Of particular importance for young singers is examination of the oropharynx to assess tonsils and the nose to exclude obstruction and allergy.

Visualisation of the vocal folds is usually possible in the child performer with patience and a gentle approach. Most will respond favourably to rigid videolaryngoscopy/stroboscopy. Many are shy of transnasal flexible endoscopy, but it is certainly worthwhile attempting if rigid endoscopy fails. Performing children are often curious about where their voice comes from and a gentle approach usually leads to successful visualisation of the larynx.

Specialist paediatric flexible and rigid scopes are readily available. The videostroboscopy should always be recorded and even if unable to achieve prolonged phonation sufficient to determine vibratory characteristics. Often an appropriate freeze frame will clearly identify pathology in an otherwise difficult examination.

Finally, it is important to stress that it is fruitless and potentially damaging to persist or be forceful with a reluctant child when trying to visualise the larynx. It is worth remembering that another time may yield a different result. It is also important to consider the clinical picture to determine whether there are any medical indications suggesting it is imperative to visualise the larynx. However, in almost all cases of young singers seen at our clinic, if no view was obtained, we have trialled voice therapy first rather than persist with endoscopy attempts or rush to a microlaryngoscopy.

In our assessments of young singers seeking treatment for a recent-onset voice disorder, we most commonly find bilateral oedema which may or may not be symmetrical. In these acute cases, we have rarely found cysts which may give weight to the argument that in children cysts may be mostly congenital [45, 46]. It is worth noting, however, that there should be high suspicion of a cyst and contra-coup lesion mimicking nodules if the singer has always sounded hoarse, there is little variability in vocal quality across time, they do not respond well to expert therapy, and/or the lesions are obviously asymmetric in presentation.

By virtue of the nature of our clinic and personnel, we tend to see many child singers particularly from the music theatre industry who develop sudden voice difficulties rather than longer-term or chronic voice disorders. It is of perhaps of interest to note that, unlike their adult co-singers, we have never seen a child with a vocal fold haemorrhage and indeed speculate that this may be rare due to the undeveloped layered vocal fold structure [44] and associated differences in vasculature.

Investigation

It is rarely necessary to perform any special investigations. Occasionally allergy testing can be fruitful in older children. Respiratory specialists are better placed to assess asthma and other lung issues. Sinus radiology in children is to be avoided unless one is considering sinus surgery in older children.

Management

Commonly the most difficult management plan for parents to accept is one of inaction or an absence of prescribed or alternative medications, but sometimes simply time, rest, and reduction of vocal load are the best recommendations. In management of the child singer, it is important to

firstly advocate for the child's health rather than the performance or singing activity they are eager to undertake. Over the past 26 years, we do not recall any vocal situation for a performing child that necessitated administration of oral steroids for a performance-related 'emergency'. In many cases, understudies or alternates are available in lieu, and this is usually preferable to the possible side effects of steroids and the psychological and artistic risks associated with a child performing suboptimally. Conversely, children recover quickly from illness, and it is easy to be overcautious, so over the years, we have become increasingly more inclined to maintain voicing through singing activity with careful and expert guidance and support.

Operative Approach

The aim of phonomicrosurgery (in the absence of dangerous disease) is to restore voice that has been unfavourably altered by pathology when other less invasive remedies have failed and when the patient has requested a remedy. It is therefore driven by voice rather than the appearance of the vocal fold and is definitely not driven by the laryngologist's desire to remove a lesion. A variety of circumstances will play a role in this decision-making.

If the lesion is acute, then time and expert voice therapy may help resolve it. If congenital, it may well not require anything, and if chronic it may require management if problematic.

The impact on the voice and how this may affect the patient's aspirations is significant. Many singers, for example, work with imperfect vocal folds, and often their 'vocal signature' results from the imperfection.

It is preferable to operate on a larger larynx than a smaller one, and deferring surgery for a time may be reasonable. For this reason, there is always a delicate balance between a 'wait-and-see' approach versus removal of pathology to increase vocal 'choice' at an earlier age, when the vocal folds may have better healing properties [20, 25]. Surgery may also prevent the potential ill effects of prolonged abnormal voicing, secondary to pathology, on the development of the layered structure of the vocal folds and the entrenchment of dysfunctional laryngeal postures.

It is unusual to surgically remove nodules from child performers, and indeed from children at all, particularly as they are most commonly soft and pliable rather than well established. Nodules in child singers usually respond to appropriate voice therapy, judicious singing training, reduction of vocal load, and behaviour modification and disappear with adolescence as the vocal folds lengthen, especially in males [47]. If absolutely necessary and all agree it is in the child's best interests for both their short- and long-term vocal health, for example, to remove polyps or cysts, then surgery should follow the standard phono-microsurgical and perioperative principles that have been very well established. When such surgery is indicated, excellent results can be achieved, and the child can enjoy significant vocal improvement giving them greater singing opportunities and choice.

Conclusion

We must emphasise that we have found children's vocal folds seem to be highly robust and resilient. Almost all use-related voice concerns can be averted expediently especially for the singing child who invariably has a greater interest or investment in vocal recovery and a unique and relatively higher insight into their individual vocal capabilities, than non-singers. High levels of vocal fitness can be achieved and seemingly extended with expert guidance, appropriate singing training, and positive singing experiences. With optimal efficiency and balance among the power, source, and filter aspects of voice, appropriate vocal repertoire and singing conditions, the young singer can happily 'load' the voice, singing healthily with endurance levels that may exceed our expectations in view of their size and stature.

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Gender-Affirming Voice

43

Sarah L. Penzell

Overview

The field of transgender healthcare is rapidly expanding and evolving in many parts of the world. Medical practitioners and other providers working in transgender and gender-diverse services attempt to keep pace with the needs of those communities. For speech-language pathologists (SLPs), our role is critical in helping individuals align their voices and communication with their gender identity. The increasing demand for voice and communication services obligates our profession to increase the population of SLPs that are comfortable, competent, and prepared to provide services in this burgeoning practice area. This chapter on gender-affirming voice serves as an introduction to a variety of concepts for transgender and gender-expansive voice and communication services and how they may apply to pediatric/adolescent patients.

Terminology and Concepts

A brief list of some important terminology and concepts that will be used throughout the chapter is provided here. The scope of this chapter does not allow for a comprehensive review of gender terminology, and speech-language pathologists who plan to work in the area of transgender and gender-expansive voice and communication will need to pursue further education including training in cultural competence as well as in clinical methods.

Gender Terminology

The booklet, website, and video series published in Australia entitled, *TRANS 101: Gender Diversity Crash Course*, explains *gender* or *gender identity* as "…part of a person's internal sense of self. It can be female, male, neither, a combination of the two, or exist completely outside of that! A person's relationship with their gender can also change over time as well" [1].

The words **transgender** or **trans** are broad terms, generally used for individuals whose gender identity or gender expression is different from the sex which they were assigned at birth, whereas **cisgender** or **cis** refers to people who identify as having the same gender identity as the sex they were assigned at birth [2].

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Gender expansive and **gender diverse** are terms that help us to describe more broadly a wide range of gender identities and expression. These differ from terms that were often used in the past, which were limited to a binary concept of gender.

Non-binary is defined by *TRANS 101: Gender Diversity Crash Course* as "an umbrella term people use to describe gender that doesn't fit squarely into male or female. This can include people who feel their gender is a mix of both, changes often, is something totally separate, or have no strong sense of a gender at all" [1].

Gender dysphoria is explained in Section III of the World Professional Association for Transgender Health (WPATH) Standards of Care, 7th Version, as such:

> Gender dysphoria refers to discomfort or distress that is caused by a discrepancy between a person's gender identity and that person's sex assigned at birth (and the associated gender role and/or primary and secondary sex characteristics) [3, 4]. Only some gender-nonconforming people experience gender dysphoria at some point in their lives. [3–5]

The Gender Identity & Expression Map by Antonia Clifford describes the terms sometimes written as **MtF** and **FtM** as, **"**MTF (M2F) Male to Female: A person who was assigned a male sex at birth, identifies as female and/or has taken hormonal or surgical steps to transition to Female. FTM (F2M) Female to Male: A person who was assigned a female sex at birth, identifies as male and/or has taken hormonal or surgical steps to transition to Male" [6].

While these terms are often seen in the medical literature, and are therefore important to recognize, when discussing voice and communication services with clients, the terms **transfeminine** and **transmasculine** are often used. Transfeminine and transmasculine do not carry the potentially problematic implication that a person has not always been their affirmed gender.

The IMPACT LGBT Health and Development Program at the Northwestern University Institute for Sexual and Gender Minority Health and Wellbeing has published Antonia Clifford's interactive learner-friendly *Gender Identity & Expression Map*, which is a highly engaging online resource for learning about gender identity and expression (Fig. 43.1) [6].

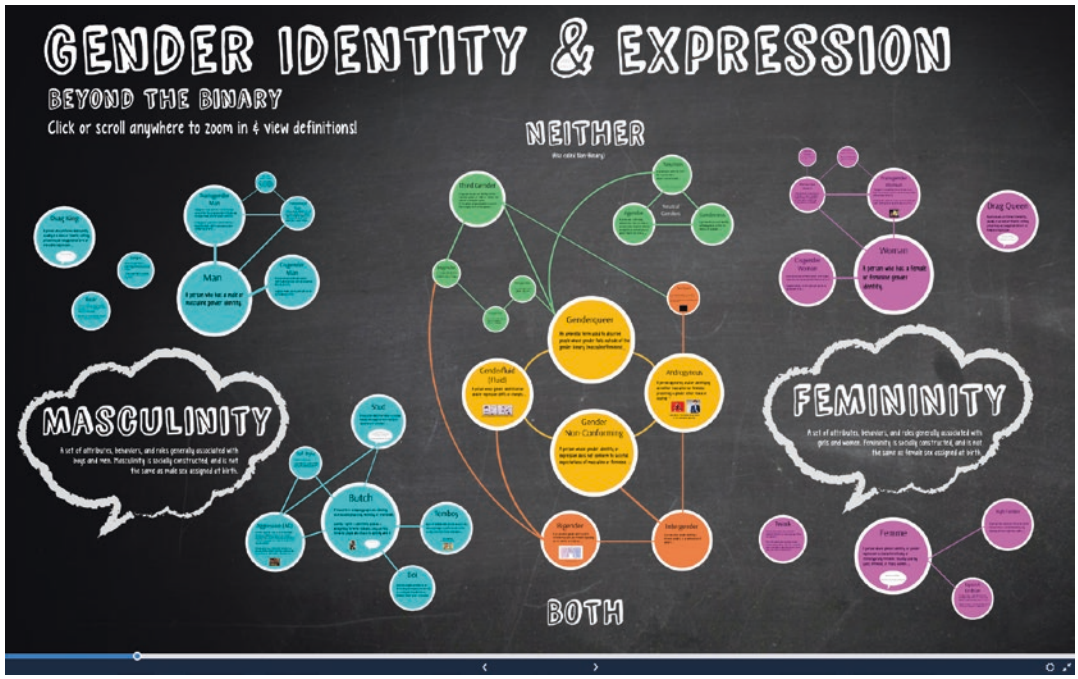

Fig. 43.1 Gender Identity & Expression Map. Text within the image can be seen more clearly within the interactive website page. (Courtesy of Antonia Clifford: https://prezi.com/yvqu4hrcexig/gender-identity-expression-map/)

Transitioning

TRANS 101: Gender Diversity Crash Course explains that "Transitioning is when someone takes steps to socially or physically feel more aligned with their gender identity" [1]. Steps a person may take when transitioning could include telling others about being transgender or gender diverse (coming out); using a different name or different pronouns; or changing use of gendered spaces (e.g., bathrooms, locker rooms). If a person chooses to physically or medically transition, it might involve altering appearance or seeking medical support or interventions, which can involve surgery or taking hormones [1]. Other steps could include a legal transition, a process in which a person might change their gender marker and/or name on legal documents. Important for our provision of services and for our cultural competency is an understanding that the steps involved in transition vary from person to person. A person may transition on a variety of timelines, including all at once, gradually, or not at all [1]. For some transgender and gender-expansive people, voice and communication modification can be an essential part of transition, to align communication with their affirmed gender.

Speech-Language Pathology Services

The World Professional Association for Transgender Health (WPATH)

Medical providers and others working in the area of transgender and gender-expansive services often look to the World Professional Association for Transgender Health (WPATH), which is one of the most highly recognized organizations for creating policy and providing essential communication and training for transgender service providers worldwide. They issued the "Standards of Care for the Health of Transsexual, Transgender, and Gender-Nonconforming People, Version 7" in 2011 [5], which is available free to the public at www.wpath.org. Of particular relevance to pediatrics and to voice and communication services are the sections entitled "Voice and Communication Therapy" and "Assessment and Treatment of Children and Adolescents with Gender Dysphoria" [5].

In the section entitled "Voice and Communication Therapy," the authors introduce the need for transgender and gender-expansive services:

> Communication, both verbal and nonverbal, is an important aspect of human behavior and gender expression. Transsexual, transgender, and gender nonconforming people might seek the assistance of a voice and communication specialist to develop vocal characteristics (e.g., pitch, intonation, resonance, speech rate, phrasing patterns) and nonverbal communication patterns (e.g., gestures, posture/movement, facial expressions) that facilitate comfort with their gender identity. Voice and communication therapy may help to alleviate gender dysphoria and be a positive and motivating step towards achieving one's goals for gender role expression. [5]

While we generally consider pediatrics to include people under the age of 18, a discussion of pediatric voice and communication services is primarily limited to a discussion of adolescents who have begun pubertal changes. Prior to the onset of puberty, our speech-language pathology services will likely be limited to consultation, education, and monitoring, as voice changes will not have occurred.

When discussing physical interventions used for adolescents, much consideration is given to whether the intervention is fully reversible, partially reversible, or irreversible [5]. The WPATH Standards of Care states that "A staged process is recommended to keep options open through the first two stages. Moving from one stage to another should not occur until there has been adequate time for adolescents and their parents to assimilate fully the effects of earlier interventions." [5] Some physical interventions can impact voice, including use of agents to suppress puberty and hormone therapy to masculinize or feminize the body. A positive outcome of puberty suppression when undertaken for transfeminine individuals is that some of the physical changes that pubertal testosterone causes to voice will not occur [7–9]. Considerations for hormone therapy include that taking the masculinizing hormone testosterone will deepen

the voice and the physical changes are not considered reversible [5, 10]. However, feminizing hormone therapy for transfeminine clients is not expected to raise pitch [9, 11, 12]. Risks of interventions must be carefully weighed with the risks of not providing an intervention [5]. If intervention for adolescent or preadolescent voice and communication is undertaken, the status of pubertal changes and any hormone interventions should be carefully considered [9]. WPATH reports that "Increasing numbers of adolescents have already started living as their desired gender role upon entering high school" [5, 13] and providing comprehensive care and support to these adolescents is crucial.

In some areas of the USA, multidisciplinary medical and support teams have been established to provide comprehensive and coordinated services to transgender and gender-expansive people. Of note, multiple children's hospitals and clinics across the country have developed coordinated clinical care programs for transgender and gender-expansive children [14]. These multidisciplinary programs generally include pediatricians, psychologists, psychiatrists, endocrinologists, urologists, surgeons, nurses, and social workers. Some programs incorporate other professionals, such as ethics consultants, school consultants, or legal consultation services on their teams. Speech-language pathologists are directly involved in some programs and will likely become increasingly included as part of these multidisciplinary teams over time. The demand for transgender and gender-expansive services for youth will likely soon push the need beyond these established programs to other hospitals, university clinics, community clinics, private practices, and schools.

Research

There are limited evidence-based research studies published for transgender and gender-expansive voice and communication services, with the field being further limited by the fact that research that currently exists is primarily based on transgender women clients and is almost exclusively based on adult participants. There are limited randomized controlled trials (RCT), and most published studies have limited sample sizes. Limited evidence-based information exists for effectiveness of communication services for aspects of communication beyond voice, such as language and nonverbal communication, and there is little data on the optimal dose for frequency and duration of sessions.

A companion document to the WPATH Standards of Care, entitled "Voice and Communication Change for Gender Nonconforming Individuals: Giving Voice to the Person Inside," summarizes the research and outlines current best practice for speech-language pathologists [15]. The document contains important information about best practice for voice and communication parameters and methods, evaluation, and clinical competence among other topics. It also covers the topics of voice masculinization and surgery for pitch elevation. Shelagh Davies introduces and summarizes the results of the article in "The Evidence Behind the Practice: A Review of WPATH Suggested Guidelines in Transgender Voice and Communication" [16]. Selected conclusions include:

- Speech-language pathology interventions for voice feminization are safe and effective [16].
- While many clients may initially be focused on modifying pitch, a combination of parameters for voice results in better outcomes for voice feminization or masculinization [16].
- It is generally agreed that increasing speaking fundamental frequency (SFF) is an important component of voice feminization, but the definitive target has not yet been determined with estimates ranging from 155 to 220 Hz [16–22].
- Resonance is an important parameter in voice feminization. The most effective way to achieve voice feminization may be to combine an increase of average SFF with raising vocal tract resonances [15, 16, 23–25].
- Intonation can be another important feature of services, including use of wider intonation contours and more upward gliding [16, 26].
- Education about vocal health and hygiene as well as about vocal anatomy and physiology are necessary parts of services [27].

While there are limited data to support other communication parameters, some clinicians may include work on semioccluded vocal tract exercises, speech rate, vocal quality or intensity, articulation, and other areas of language and nonverbal communication as targets of overall communication modification. Published research is available on potentially feminine and masculine characteristics of some of these parameters, but clinicians should be wary of stereotypes and discuss goal areas with clients carefully and openly [15, 16].

Evaluation/Assessment

As previously discussed, speech-language services are optimally delivered as part of an overall team of providers, supporting each individual in their medical, psychological, and social needs [16]. While ASHA guides speech-language pathologists that prior to initiating voice therapy, "All patients/clients with voice disorders are examined by a physician, preferably in a discipline appropriate to the presenting complaint," [28] transgender and gender-expansive individuals seeking voice modification are considered to be non-disordered populations, and a screening for voice disorder is generally accepted, with referral provided for further examination only if needed [16]. However, the practice of screening versus requiring a physician examination is debated, with some SLPs in this practice area requiring the physician exam prior to initiating services.

Davies, Papp, and Antoni state, "To date there are no minimal standards for the assessment of voice and communication in transgender clients. The field of practice is recent and the evidence base, is still weak" [15]. Evaluations should be comprised of multiple assessments spanning various voice and communication parameters. Careful attention needs to be paid to client goals as they relate to measures undertaken in an initial evaluation.

An initial interview should include a discussion of client goals including the client's perception of their voice and communication use. SLPs need to be aware that some clients may be open to discussing their past, while some may not. Questionnaires, such as the Transsexual Voice Questionnaire for Male-to-Female Transsexuals (MtF)-TVQ(MtF), may be used to gain further insight into impact that voice presentation has for clients [29]. Acoustic measurements should include "average SFF, SFF range, maximum phonation range, and the first, second and third formants of vowels, in particular the corner vowels /a/, /i/, and /u/" [15] elicited across a variety of speaking tasks including phonation of prolonged vowel, oral reading, picture description, and conversational speech [16, 30–32]. Video or audio recording may be used to aid in assessment of voice and communication [15].

Hancock and Helenius Study

Adrienne Hancock and Lauren Helenius published a study in the *Journal of Communication Disorders* in 2012 documenting results of an adolescent's response to a speech-language voice and communication intervention [9]. In the only peer-reviewed case study of a pediatric transgender client for voice and communication services, the paper by Hancock and Helenius provides rationale, procedures, and outcomes for one 15-year-old MtF transgender client, who attended 15 sessions conducted over 7 months in a university clinic. The client had strong family support, no history of voice issues or abuse, and no previous work with SLP on transgender voice and communication, but she had attempted some voice modifications on her own. The client participated in sessions which included many elements of a typical transgender voice and communication program, including work on: vocal hygiene, relaxation techniques, breath support, fundamental frequency, intonation, resonance, vocal quality, and rate [9]. Positive results were reported across multiple domains and included client report of increased self-confidence as result in change in communication and progress during SLP services. Perceptually, improvements were documented in multiple areas, including breathiness; pitch, which increased to within typical female limits; increased use of feminine intonation (greater pitch contours);

forward-focused resonance in oral cavity; relaxed and aligned posture; and use of primarily abdominal-diaphragmatic breathing. Unfamiliar listeners gave high ratings for the femininity and "softness" of the client's voice [9]. The conclusions of the study indicated that voice and communication services helped the client to achieve feminine voice and communication, using both objective and subjective data, and that methods used for MtF transgender adults were effective for a younger voice [9]. Though the scope of this study was limited, it is an important start to our understanding of the ways in which voice and communication methods that are used for the adult population can be effective for adolescent clients as well.

Numbers of Transgender and Gender-Expansive Children/Adolescents

With the growing number of out and transitioning youth, there is an increasing need for speech-language pathology services for voice and communication [33]. There is limited literature about voice and communication services for children and adolescents. Since children have not yet experienced pubertal voice changes, a speech-language pathologist's role with transgender or gender-expansive children and families, if needed, is most typically as an educator or consultant. Transgender or gender-expansive adolescents may be eager to seek out education and services for voice and communication modification, especially when experiencing voice changes during puberty.

Conclusion

The American Speech-Language-Hearing Association (ASHA) provides resources and guidance via their web page "Providing Transgender Voice Services" [34] and is working toward publication of a new practice portal for transgender voice and communication. WPATH provides guidelines for minimal credentials for SLPs working in this practice area [5]. ASHA also instructs SLPs to refer to another provider with special expertise, if one does not feel they have the skill set to address the needs of a client [35].

With the growing number of children and adolescents identifying as transgender and gender diverse, voice and communication modification will be an increasing need. Adolescents in some school districts have begun to ask school speech-language pathologists for help accessing these services. Because of the non-disordered nature of services, speech-language pathology does not currently qualify for inclusion on an individualized education plan (IEP), but SLPs in this practice area are beginning to explore how voice and communication modification can be extended to the school setting. This quickly changing and emerging area of practice is both challenging and rewarding. Speech-language pathologists who provide these essential services will surely benefit from helping transgender and gender-expansive clients achieve their goals and become aligned with their gender.

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Weird Wonders of the Larynx

44

J. Scott McMurray and Matthew R. Hoffman

As with any profession or specialty, tedium and monotony can threaten as experience is gained and expertise achieved. Pediatric voice and swallow disorders are all too often cast as a set of problems that will resolve once puberty and maturation occur. I believe this is far from the truth, although I also believe we must first do no harm and therefore think before we incur risk by altering structures that may otherwise resolve without undue or prolonged sequelae. If every dysphonic child had the resources to receive an assessment and treatment, the world would be a better place with less discrimination and a higher quality of life. In this profession, however, not all children will present with vocal nodules that need some voice therapy to change their maladaptive behaviors and set them toward a path of euphonia. The following are a few case experiences of weird, wonderful, and unexpected things that stimulated, challenged, and required creative troubleshooting. They are presented here as a potpourri to energize the clinician and affirm the great variety of problems we may face in the field of pediatric voice and swallowing.

Infant with Noisy Breathing

NK was a few weeks old and had noisy breathing since birth. It was described as inspiratory stridor that was worse with activity and crying. His cry was not usual and could be described as breathy or weak. He was term and had been gaining weight well. He had never been intubated. He had no visible birthmarks. He had been seen by an otolaryngologist who had looked at his larynx with flexible laryngoscopy and had made the diagnosis of unilateral left vocal fold paralysis and was sent for further evaluation and management. His breathing difficulties were slowly progressing, and he was beginning to develop retractions with agitation but no cyanosis. He had no known cardiac anomalies and had not had any surgery.

Repeated office nasopharyngolaryngoscopy was performed to assess the laryngeal structures and function. There was indeed asymmetry of motion of the vocal folds. The vocal folds were difficult to visualize due to supraglottic hooding. The right larynx seemed to be moving well and there was little to no motion on the left.

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Figure 44.1 shows the transnasal view of the larynx. It revealed decreased motion of the left hemilarynx. There were a supraglottic web and a very small left hemilarynx. The right arytenoid seemed relatively large compared to the small left hemilarynx. It was very difficult to see the vocal folds due to the supraglottic hooding. The child had a very breathy cry and inspiratory stridor with prolapse of the supraglottic structures during inhalation. A diagnosis of decreased left laryngeal motion and a supraglottic web was made. Operative endoscopy for further physical examination and an accurate diagnosis was recommended.

The child was taken to the operating room where inhalational anesthesia with spontaneous ventilation and insufflation. It was difficult to visualize the larynx due to the small glottic inlet posed by the supraglottic web. The child was easily intubated with a small endotracheal tube. Figure 44.2 shows the operative endoscopic findings with the supraglottic web and a left hemilaryngeal hypoplasia. The bottom right panel shows the initial visualization after intubation. The web can be seen anterior to the endotracheal tube. It appears to extend from false vocal fold to false vocal fold. The bottom left panel shows the web as it was incised with a sickle knife. The right upper panel shows the lysed web and exposure of the glottis and right vocal fold. The left upper panel shows the end result after lysis of the web has occurred. The left vocal fold is atretic. There is poor development of the arytenoid cartilage and membranous vocal fold.

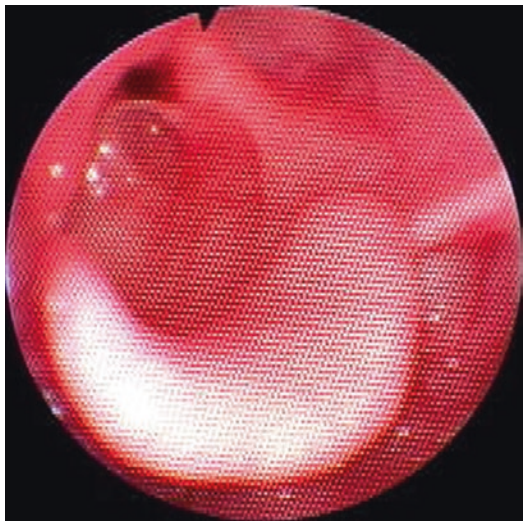

Fig. 44.1 Nasopharyngolaryngoscopy of this child revealed decreased motion of the left hemilarynx. The epiglottis is at the bottom of the photo and the posterior glottis is at the top. There were a supraglottic web and a very small left hemilarynx. The right arytenoid seemed relatively large to the small left hemilarynx. It was very difficult to see the vocal folds due to the supraglottic hooding

Although there was left hemilaryngeal hypoplasia and therefore a smaller subglottis, after release of the supraglottic web, it appeared that there would be an adequate airway. The child was extubated. Over the years, he has had some dysphonia characterized by breathiness with poor projection. He has also had some exercise intolerance but now plays football. Figure 44.3a, b shows his laryngeal development at the age of 5. He has severe hypoplasia of the left vocal fold and little or no motion. He has been able to compensate and achieve glottic closure with his right glottis. The hypoplasia of the left vocal fold in relation to the right vocal fold at the anterior commissure can be seen in Fig. 44.3b.

His larynx has grown with him over the years, and no other reconstructive surgery has been required. He has undergone voice therapy to maximize his voicing with the laryngeal structures that he has. His initial diagnosis was difficult, and it was easy to ascribe his difference in laryngeal motion to a paralysis. It was not easy to tell without the operative endoscopy the nature of his true anatomy. It did appear at first glance to be a left-sided paralysis, but in reality, his motion impairment was from underdevelopment of the left hemilarynx and not a paralysis or fixation.

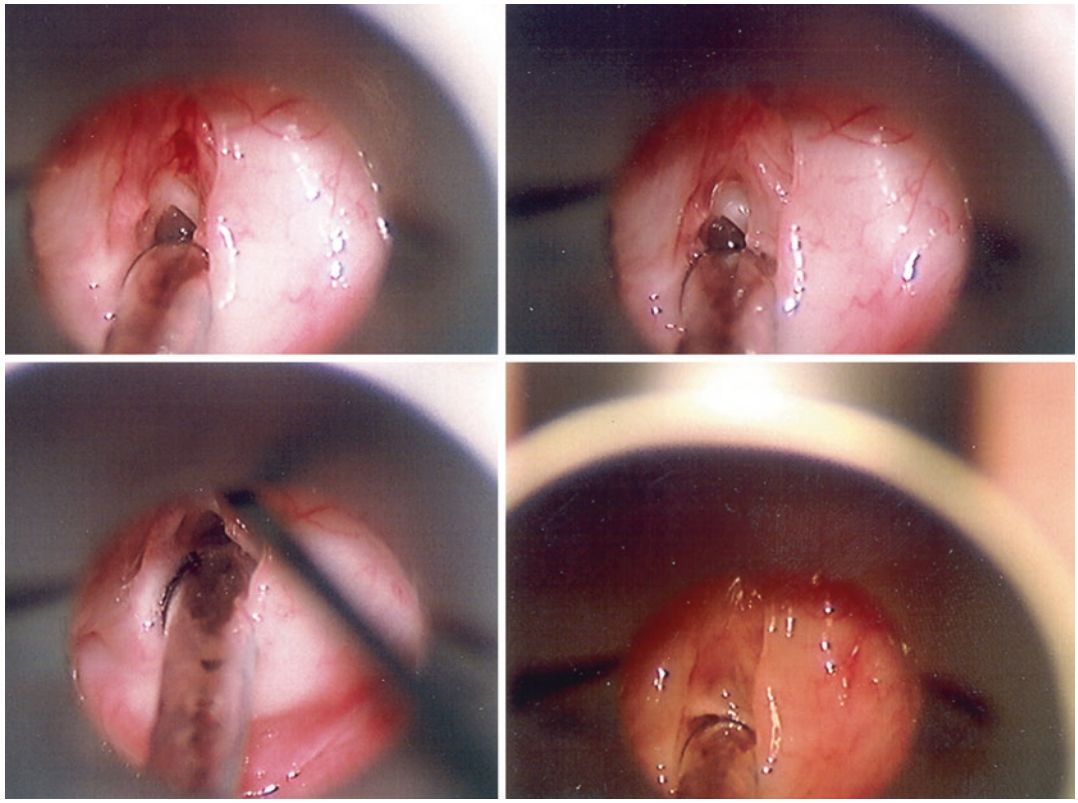

Fig. 44.2 Operative endoscopy of the child with supraglottic webbing and a left hemilaryngeal hypoplasia. Anterior is at the top of the panel and posterior is at the bottom. The bottom right panel shows the initial visualization after intubation. The web can be seen anterior to the endotracheal tube. The bottom left panel shows the web as it is incised with a sickle knife. The right upper panel shows the lysed web and exposure of the glottis and right vocal fold. The left upper panel shows the end result after lysis of the web has occurred. The left vocal fold is atretic. There is poor development of the arytenoid cartilage and membranous vocal fold

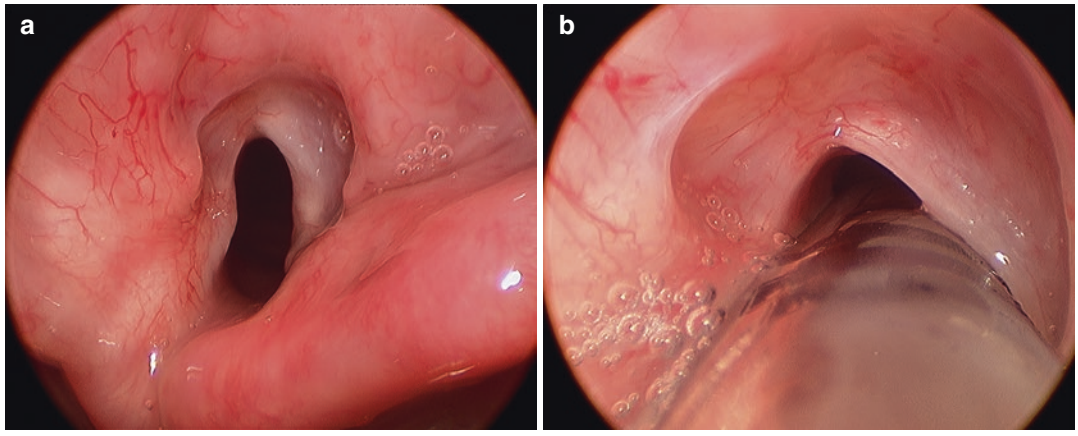

Fig. 44.3 (**a**, **b**) Operative endoscopy of NK at the age of 5 years. He has a left hemilaryngeal hypoplasia best seen in (**a**). The release of the supraglottic web can be seen above the vocal folds. (**b**) The same view after intubation with an age-appropriate endotracheal tube to demonstrate the anterior commissure and the hypoplastic nature of the left glottis. There is no motion on his left side. He has compensated to achieve near total closure with his right side

Infant Choking in the Back of a Car

A distraught mother drove to the nearest emergency room with her infant who had been choking and had hemoptysis. While driving, her rear window spontaneously shattered, covering the rear seat, the infant carrier, and her child. She pulled over and cleared the broken glass from the car seat and her child. The child appeared fine. As she drove for a few more miles, she heard the infant choking and then saw that there was some bloody froth coming from the baby's mouth. The mom decided to drive to the closest emergency department. The infant was drooling but did not have respiratory distress. With the history of a witnessed cough choke, the ED obtained a chest radiograph seen in Fig. 44.4. A large shard of glass was seen in the infant's cervical esophagus.

The infant was sent by helicopter to our institution for management of the esophageal glass shard. It was uncertain how the shard would be removed and what damage might occur to the esophagus. Other surgical services were notified that the child would be taken to the operating room for removal of the glass shard. The child had a general anesthetic with endotracheal intubation. Rigid esophagoscopy allowed for visualization of the upper part of the glass shard in the cervical esophagus. The shard was too large to bring into the rigid esophagoscope to sheath the sharp edges of the glass. The glass was secured with an optical grasper and gently removed from the esophagus. The piece slid out of the esophagus easily through the cricopharyngeus. It was taken out as a single piece although it was safety glass and had multiple fractures. Repeated esophagoscopy did not reveal any mucosal injury or other pieces of glass. The retrieved glass shard can be seen in Fig. 44.5. Although the piece was large, close inspection of the broken edges revealed obtuse angles of fracture. This is a very fortunate feature of safety glass. Over the years, automobile manufacturers have moved from plate glass to laminated glass and now tempered glass as a safety feature in cars such as seat belts. Safety glass was added to the rear windows some years after it was introduced for the front windscreen.

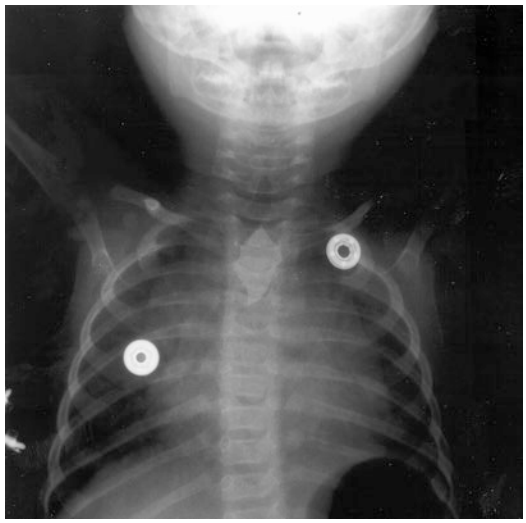

Fig. 44.4 Chest radiograph of an infant with choking and hemoptysis. The rear window he was facing in his infant carrier had spontaneously exploded and covered him and the rear seat with shards of glass. His mother had pulled over and cleared the glass from his seat, but a few miles later, he was witnessed to choke and have hemoptysis. This is the initial chest radiograph upon presentation to the ED. A large shard of glass can be seen in the proximal esophagus

Fig. 44.5 Large shard of safety glass retrieved as an esophageal foreign body. The piece was too large to sheath inside the rigid esophagoscope to protect the esophageal mucosa. The special features of safety glass, however, helped to protect this infant. Safety glass fracture with mostly obtuse or square edges, decreasing the number of sharp knifelike edges that would have lacerated the esophagus. There was no injury to the esophagus upon retrieval of this foreign body

Newborn with Imperforate Anus Repair Unable to Extubate with Square Flow Volume Loop on Ventilator

An infant with truncus arteriosus congenital cardiac anomalies underwent repair of an imperforate anus on the second day of life. The child was not ready for extubation at the end of the procedure. He was transferred to the PICU (pediatric intensive care unit) for further care. There was difficulty with ventilation, and his flow volume loop on his ventilator was square with a clipped inspiratory and expiratory loop consistent with a fixed obstruction. Consultation for airway evaluation was requested due to the flow volume loop. After discussion with the cardiothoracic and intensivist team, diagnostic rigid endoscopy was performed. He would need cardiac surgery in the near future, and it was hoped that he would be able to extubate after the imperforate anus repair. It was easy to believe, however, that the clipped flow volume loop was an artifact. At endoscopy, however, this infant was found to have long-segment complete tracheal rings spanning from above the thoracic inlet to just above the carina. There was swelling and granulation tissue developing where the tip of the endotracheal tube had been resting against the tracheal stenosis. The tracheal narrowing can be seen in Fig. 44.6.

A plan for corrective tracheal and congenital heart surgery was formulated. He had a long-segment slide tracheoplasty to repair his complete tracheal rings. While the trachea was split, the aberrant pulmonary artery was repositioned. His truncus arteriosus was also repaired at the same time. He eventually has done very well and is now 6 years old and very active.

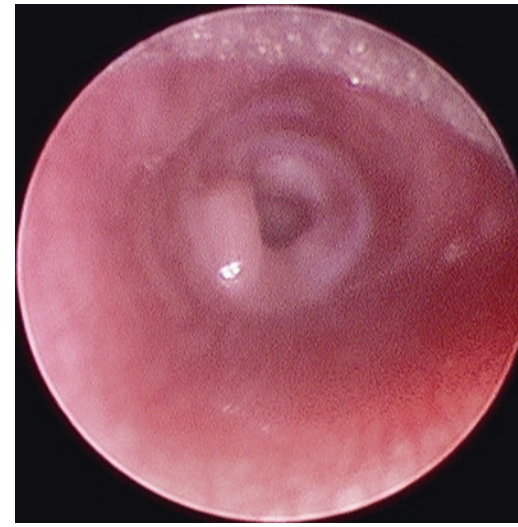

Fig. 44.6 Operative endoscopy showing a circular pattern to the cervical trachea consistent with complete tracheal rings. This child was unable to extubate after repair of an imperforate anus and prior to his complex cardiac surgery repair. His ventilator showed a square and clipped flow volume loop suggesting a fixed obstruction

Laryngeal Star

A 31-month-old girl was helping her mother wrap Christmas presents the weekend after Thanksgiving. She had a cough choke episode and her mom took her urgently to the ED. She was evaluated and her cough stopped, and no foreign body was seen by radiograph. Her voice was normal, and she was able to swallow well. She was discharged from the ED. Over the next month, she developed intermittent coughing and symptoms consistent with upper respiratory tract infections. She had been treated with an oral antibiotic for rhinosinusitis. This helped her some of her symptoms, but they continued to wax and wane over the next weeks. She went to several urgent care settings, and then her breathing started to be consistently labored, and her voice began to become consistently hoarse. She was sent to otolaryngology around St. Patrick's Day. An otolaryngologist did flexible nasopharyngoscopy who saw granulation tissue at the posterior glottis consistent with possible changes from reflux. The otolaryngologist was not comfortable with the clinical picture and so sent her to pediatric otolaryngology for evaluation. During office nasopharyngoscopy with pediatric otolaryngology, the granulation was also seen. There were also granulation at the anterior commissure and what appeared to be a foreign body wedged from the anterior commissure to the interarytenoid gap. The cough choke event was then remembered by her mother. She was taken to the operating room for further endoscopy and removal of the foreign body. As the foreign body had been

there for 3–4 months, there was great concern about the airway at induction. An inhalational anesthetic was performed with spontaneous ventilation. When she was in a stable plan, laryngoscopy was performed confirming a foreign body lodged in the laryngeal inlet. This can be seen in Fig. 44.7. The foreign body was grasped by an optical grasper and was removed from the glottic inlet. The foreign body, seen in Fig. 44.8, was metallic coated plastic star that could be attached to Christmas presents. There was significant granulation tissue at the glottic inlet as seen in Fig. 44.9. This was not disturbed. There was no bleeding. She was given steroids and remained un-intubated but was observed in a monitored setting. She did well and her voice returned to normal.

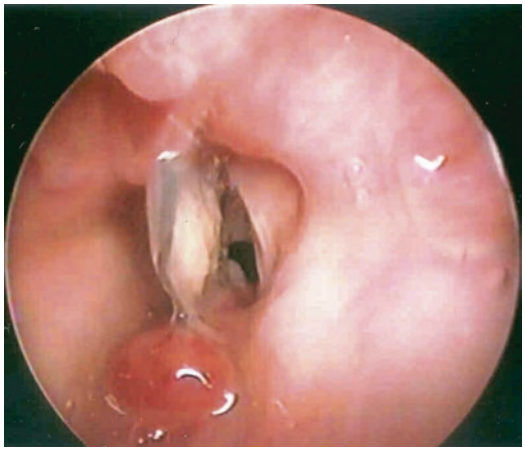

Fig. 44.7 Laryngoscopy of a 31-month-old with a foreign body wedge from the anterior commissure to the posterior interarytenoid gap. The anterior commissure is at the top, and the granulation tissue that can be seen is in the interarytenoid gap

Fig. 44.8 The laryngeal foreign body was a plastic star that could be attached to a package for Christmas

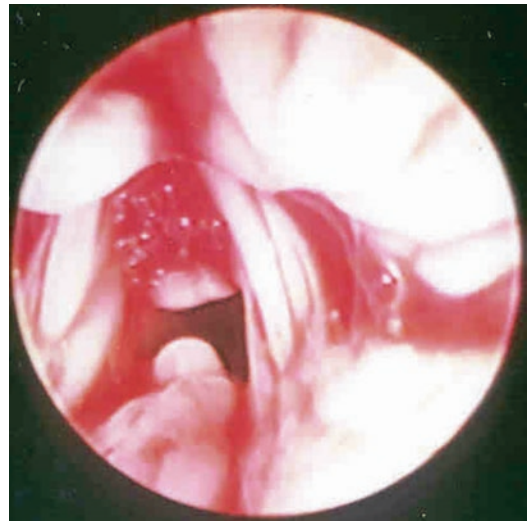

Fig. 44.9 Granulation tissue at the anterior commissure and the posterior glottis as a result of a foreign body that was wedged there for 3 months

Hoarseness Refractory to Voice Therapy

A 9-year-old young man with severe dysphonia was diagnosed with vocal nodules and laryngeal inflammation consistent with laryngopharyngeal reflux. He was treated with voice therapy and escalating antireflux measures. He was treated with a proton pump inhibitor and H2 blockers. He mastered his voice therapy but continued to have fluctuating voice quality which interfered with scholastics and caused a significant decrease in quality of life. He was difficult to examine as he did not tolerate rigid laryngoscopy and also had a hard time with flexible nasopharyngoscopy, which can be seen in Fig. 44.10. Despite these hardships, he was very committed to improving his voice and followed his voice therapy regimen and was compliant with his medication.

As he had not made progress over the previous year, despite adequate treatment for reflux laryngitis and vocal nodules, the diagnosis was ques-

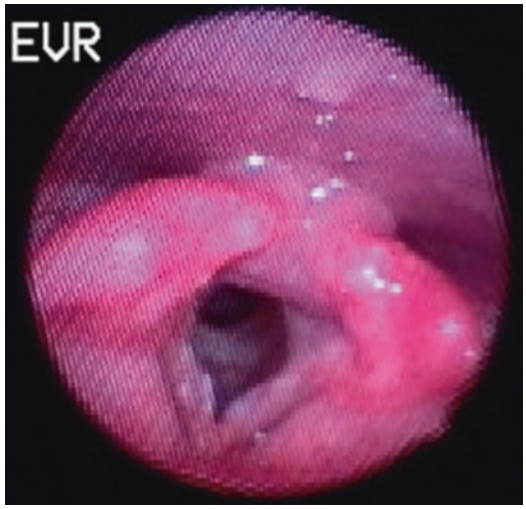

Fig. 44.10 Nine-year-old with severe dysphonia refractory to voice therapy and antireflux treatment. There are erythema and edema as well as corrugation of the interarytenoid area suggestive of laryngopharyngeal reflux. There are bilateral thickenings that were diagnosed as vocal nodules

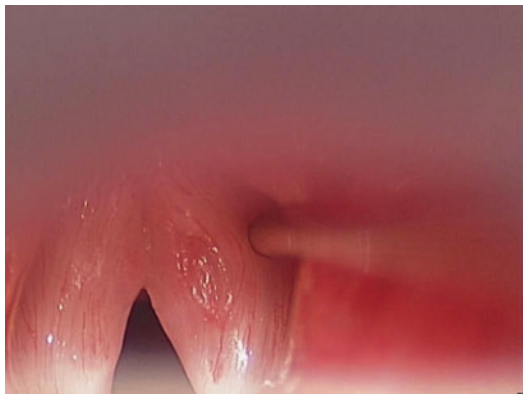

Fig. 44.11 Right-sided sulcus vocalis seen in a 9-year-old young man with persistent dysphonia. This lesion was not appreciated earlier. He was taken for operative endoscopy and further physical examination as his voice was not improving despite appropriate treatment for the presumed diagnosis. He needed surgical correction of the sulcus vocalis to progress in his vocal quality

tioned, and so further physical examination with direct laryngoscopy was recommended. At direct laryngoscopy seen in Fig. 44.11, a new previously unrecognized lesion was identified. A right-sided sulcus vocalis was seen. This was excised and the mucosa was reapproximated. There was a contralateral lesion consistent with a reactive

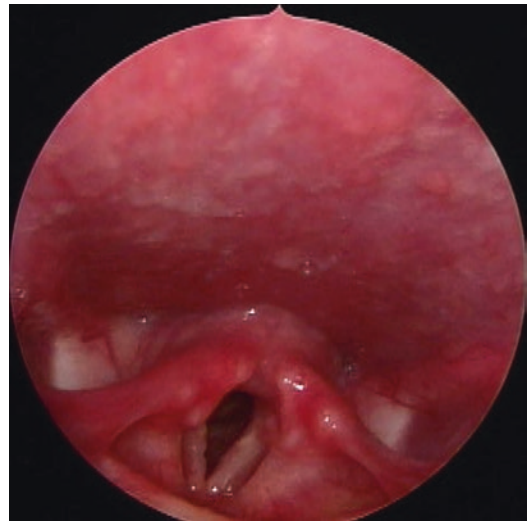

Fig. 44.12 Nasopharyngoscopic view of a 9-year-old young man after an excision of a sulcus vocalis and voice therapy. The laryngeal lesions are much smaller. He is happy with his vocal quality and durability

nodule. The patient's voice dramatically improved after healing from the microlaryngeal surgery and voice therapy. The antireflux medication was weaned off. He continued to have an improved and durable voice and was very happy with improved scholastics and social acceptance. His postsurgical result can be seen in Fig. 44.12. If results are not achieved as expected based on the diagnosis, challenge the assumptions and reassess.

Functional Dysphonia

A 5-year-old boy had a history of intermittent upper respiratory tract infections that also caused dysphonia. His voice waxed and waned with a slowly progressive course of worsening hoarseness. He was treated for URIs a few times at urgent care and then was sent for a voice evaluation and voice therapy. He had worsening dysphonia and began to have periods of aphonia. He was diagnosed with a functional voice disorder as he could have normal voicing if he tried very hard but easily fell into severe dysphonia and aphonia. He then began to develop respiratory compromise with worsening URI symptoms and noisy breathing. He was sent for and evaluation with

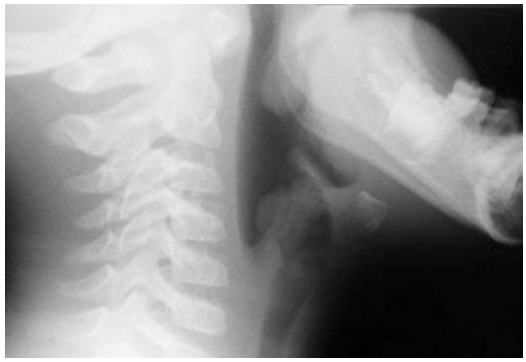

Fig. 44.13 Lateral neck radiograph in a 5-year-old boy with slowly progressively worsening dysphonia and respiratory symptoms. A mass lesion like a cluster of grapes can be seen at the glottic inlet

an otolaryngologist who obtained a lateral neck radiograph seen in Fig. 44.13. The lateral neck radiograph showed a mass lesion like a cluster of grapes at the glottic inlet. The boy was then sent for evaluation by pediatric otolaryngology. Nasopharyngoscopy was performed and he was found to have severe obstructing papillomatosis. The laryngeal inlet could not be visualized because of the size of the glottic papilloma. The boy was taken urgently to the operating room to debulk the papilloma. He was intubated to maintain his airway, while the papillomata were debrided with the laryngeal skimmer. His initial operative laryngoscopy can be seen in Fig. 44.14.

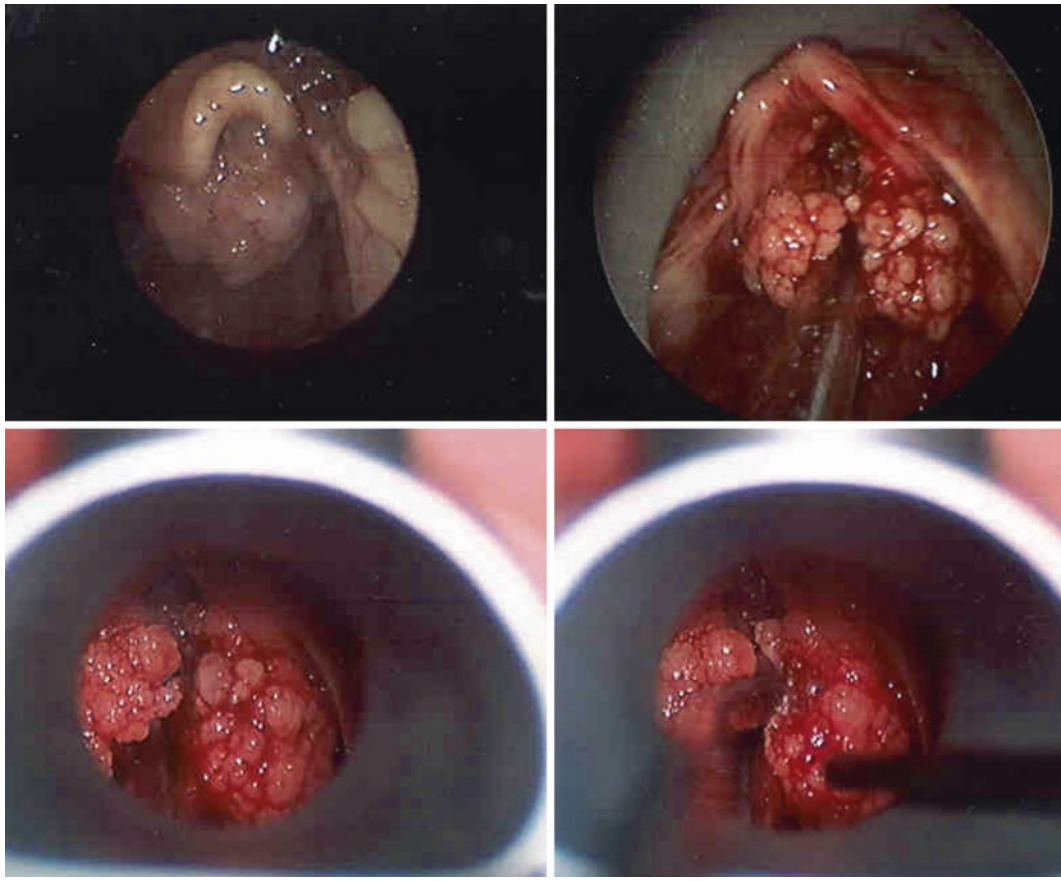

Fig. 44.14 Operative laryngoscopy in a 5-year-old with worsening voicing and breathing symptoms. He was originally diagnosed with functional dysphonia as he could produce good voicing if he tried very hard. His larynx had not been visualized previously. It is very important to have an accurate diagnosis prior to therapy for voice disorders. His laryngeal inlet is filled with papilloma. The tip of the epiglottis can be seen at the top of the endoscopic images

This case demonstrates the importance of visualizing the larynx prior to instituting voice therapy. Accurate treatment requires an accurate diagnosis.

Safety Pin Ingestion

An 11-month-old was playing with his father when he had a witnessed cough choke episode followed by crying and drooling. The child was taken to an emergency room where radiographs were obtained to rule out foreign body. The radiographs can be seen in Fig. 44.15 and

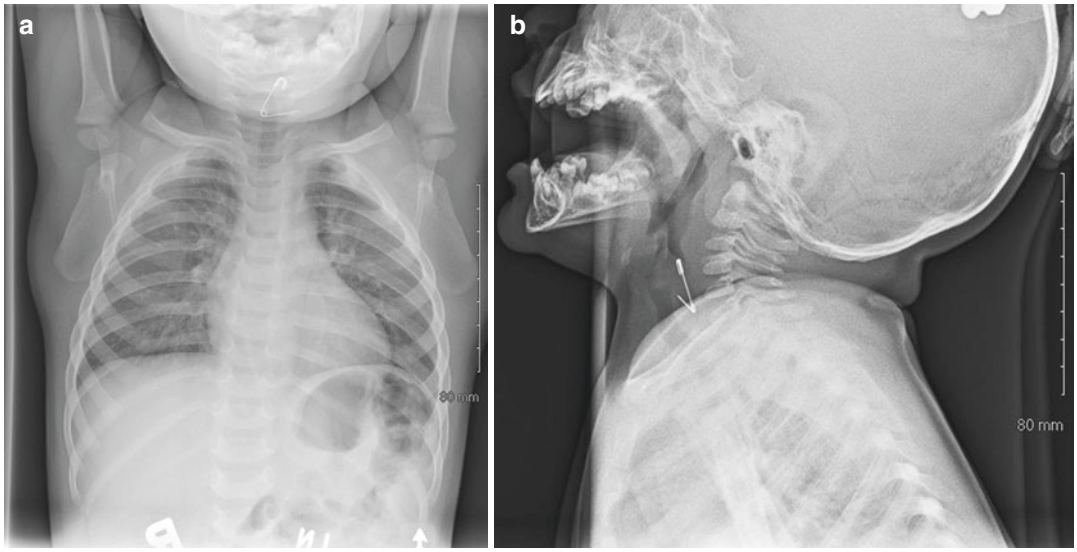

Fig. 44.15 Posteroanterior chest (**a**) and lateral neck (**b**) radiographs demonstrating an open safety pin in the cervical esophagus in an 11-month-old infant

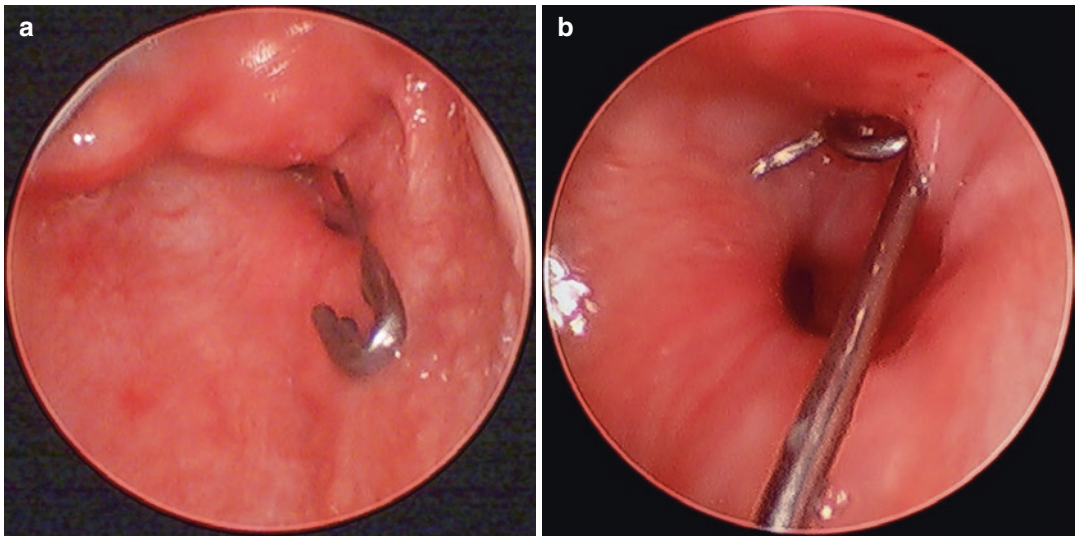

Fig. 44.16 (**a**, **b**) The open safety pin can be seen during operative endoscopy. The safety pin is open, and the needle penetrated the hypopharyngeal mucosa. To retrieve the safety pin, it must be pulled from the mucosa and the tip sheathed and protected

demonstrated an open safety pin at or below the cricopharyngeus. The child was transferred to a pediatric hospital for management. He was taken to the operating room for removal of the foreign body. His airway was secured, and the safety pin was found in the postcricoid area penetrating the posterolateral mucosa as seen in Fig. 44.16. Retrieval of the safety pin was challenging because of its location and the opened nature. The pin was located in the postcricoid area and so the larynx had to be lifted to expose it. The pin had to be pushed away and down into the cervical esophagus to disengage the needle from the hypopharyngeal wall. This put the safety pin below the cricopharyngeus. The esophagoscope had to be repositioned to visualize the safety pin again. The pin could not be closed, and so the sharp point was sheathed into the esophagoscope tube to protect the mucosa. The safety pin was then removed in a single motion. The only injury was the needle puncture seen in Fig. 44.17. The difficulty resulted from the position of the pin requiring losing visualization briefly and then re-exposure. As the pin could not be closed, the point was sheathed leaving a small portion of the pin outside the esophagoscope.

Plastic Bottle Collar Calamity

A 9-month-old was playing with a water bottle and had a witnessed cough choke episode. She was taken to an ED and examined. Her coughing decreased and she had no respiratory symptoms. Radiographs were reportedly normal. She was discharged but was told to return if symptoms returned. Over the next 6 weeks, she had intermittent recurrence of upper respiratory symptoms and intermittent hoarseness. She was treated with several rounds of oral antibiotics with only transient results. As her respiratory symptoms were progressing, she was eventually admitted to a local hospital as a transfer from a neighboring ED. During her admission, the history of the recent cough choke was re-established, and the hospitalist called the otolaryngology service for a transfer to the pediatric hospital and for a consult to rule out a foreign body. Radiographs had been negative. Nasopharyngoscopy upon transfer revealed a white plastic foreign body impaled in the nasopharynx (Fig. 44.18) and hanging down toward the larynx. The tip of the plastic piece poked at the larynx with swallow-

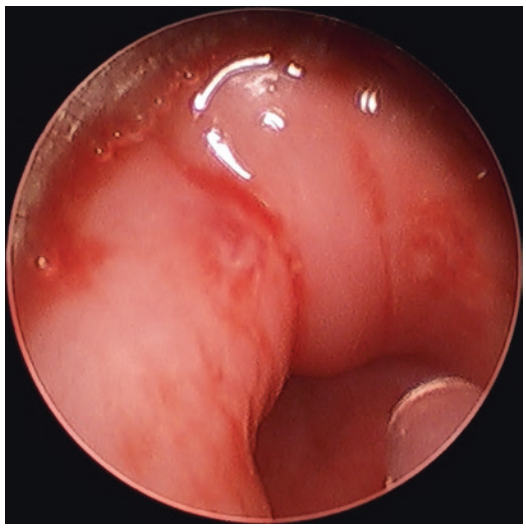

Fig. 44.17 Residual needle puncture after retrieval of an open safety pin. The needle was disengaged from the mucosa and then sheathed into the tube of the esophagoscope to protect the mucosa from further injury

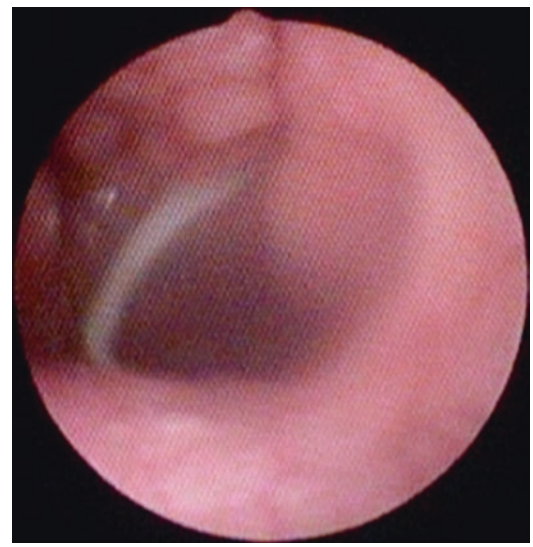

Fig. 44.18 This is a view into the left nasopharynx of a 9-month-old with a nasopharyngeal foreign body. The white plastic foreign body can be seen coming from the left lateral nasopharyngeal wall extending to the patient's right and heading down into the hypopharynx

ing and crying. This had caused swelling and granulation tissue on the left vocal fold as seen in Fig. 44.19. Figure 44.20 shows a closer view of the laryngeal injury caused by the intermittent piercing by the plastic foreign body. The foreign body can be seen to the extreme left in the figure. The child was taken to the operating room for removal of the foreign body. Her airway was compromised by the swelling and granulation from the prolonged intermittent injury from the nasopharyngeal plastic. She required intubation to maintain her airway. The foreign body was

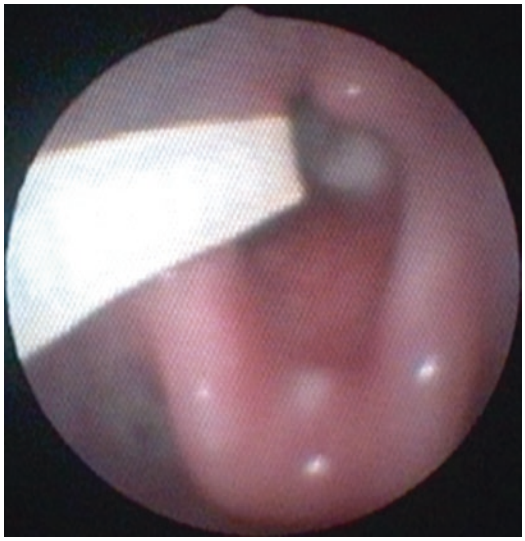

Fig. 44.19 Hypopharyngeal view of a 9-month-old with a nasopharyngeal foreign body. The white plastic foreign body can be seen hanging down into the glottic introitus

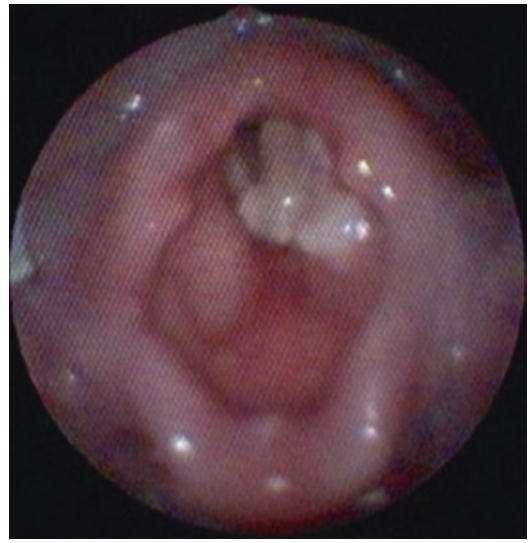

Fig. 44.20 Laryngeal view of the injury caused by a nasopharyngeal foreign body hanging down and causing intermittent contact and trauma to the left vocal fold during crying and swallowing in a 9-month-old infant with a delayed diagnosis of nasopharyngeal foreign body

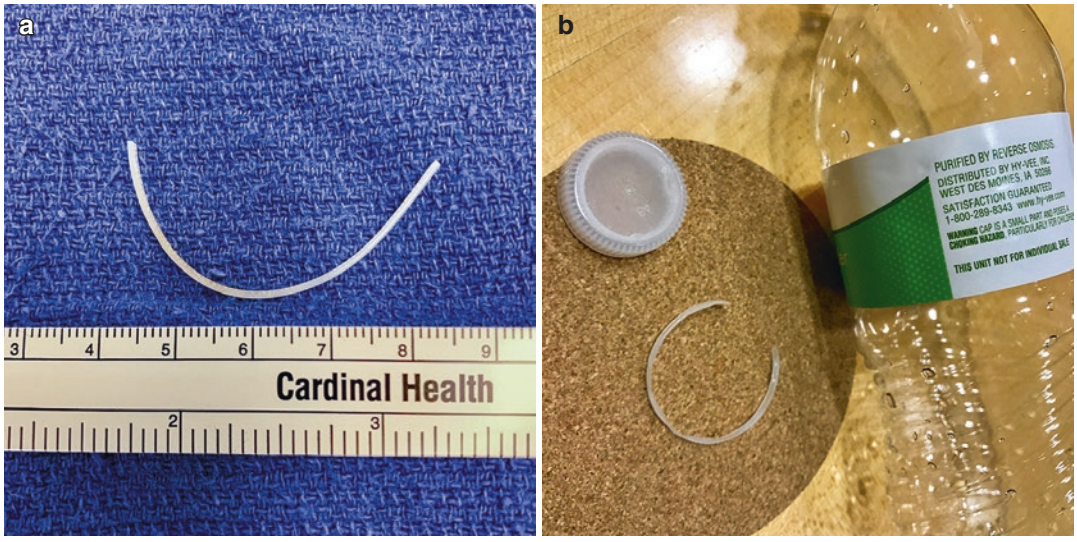

Fig. 44.21 (**a**) The retrieved plastic foreign body is retrieved from the nasopharynx of a 9-month-old who had a cough choke episode 3 weeks prior to final identification and retrieval. (**b**) The plastic collar from a water bottle with a choking warning label is seen

then pulled from the nasopharynx. Complete endoscopy did not reveal other foreign bodies. Some of the granulation tissue was debrided to improve on her airway and allow for extubation. It was determined that the foreign body was the plastic collar from the top of a water bottle. The foreign body retrieved can be seen in Fig. 44.21a. Figure 44.21b shows a bottle from which the plastic collar may have come. There is a warning on the label of the bottle. When symptoms do not make sense, always start from the beginning and have a high index of suspicion. The hospitalists that readmitted this child started the history over again and rediscovered the cough choke as a sign for a foreign body.

These vignettes are presented to highlight the wide variety of potential difficult problems that can be seen in treating children with voice and swallowing problems. They require creativity in diagnosis and treatment. The impact on these patients' lives is very rewarding.

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