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.

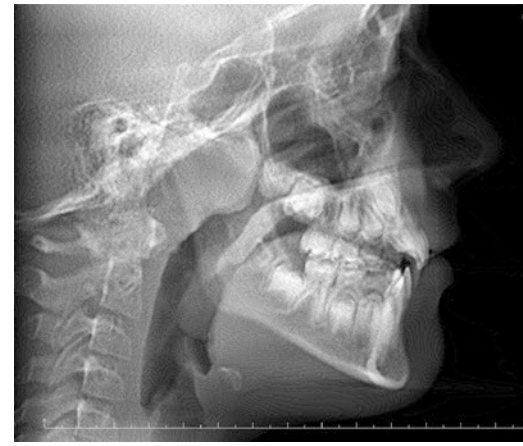

Fig. 1.1 Large adenoidal pad filling the nasopharynx in this patient

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

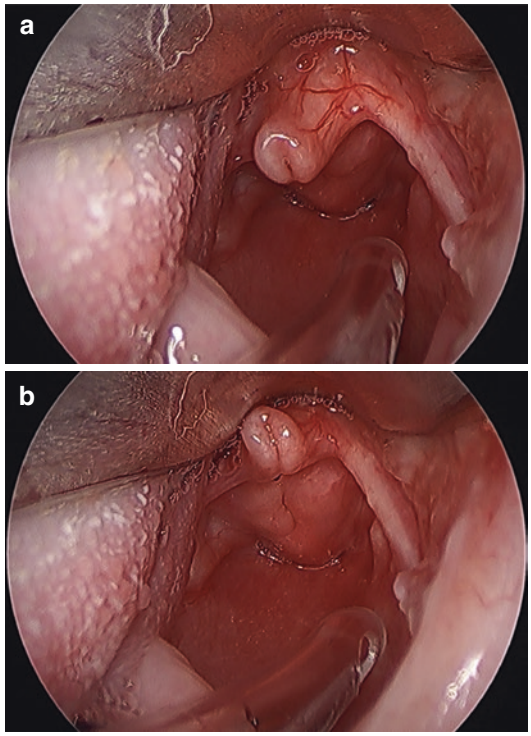

Fig. 1.2 (**a**, **b**) Operative direct laryngoscopy in a 6-week-old child with signs and symptoms of progressive noisy breathing and retractions since birth. Her symptoms were worse during activity of crying and feeding. She has a diagnosis of laryngomalacia. The infantile larynx is curled and obstructed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

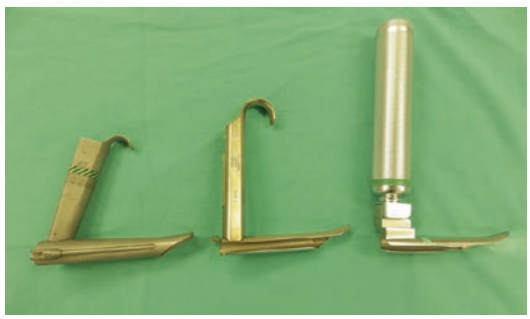

Fig. 2.1 Examples of pediatric laryngoscopes, including the Lindholm (left), Parsons (middle), and Philips (right)

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

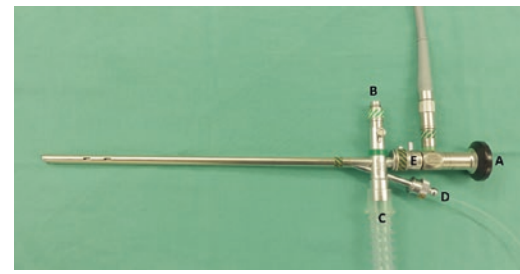

Fig. 2.2 Ventilating bronchoscope with attachments including telescope (**a**), light prism (**b**), ventilatory circuit attachment (**c**), and suction (**d**). A bridge (**e**) connects the telescope to the bronchoscope

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

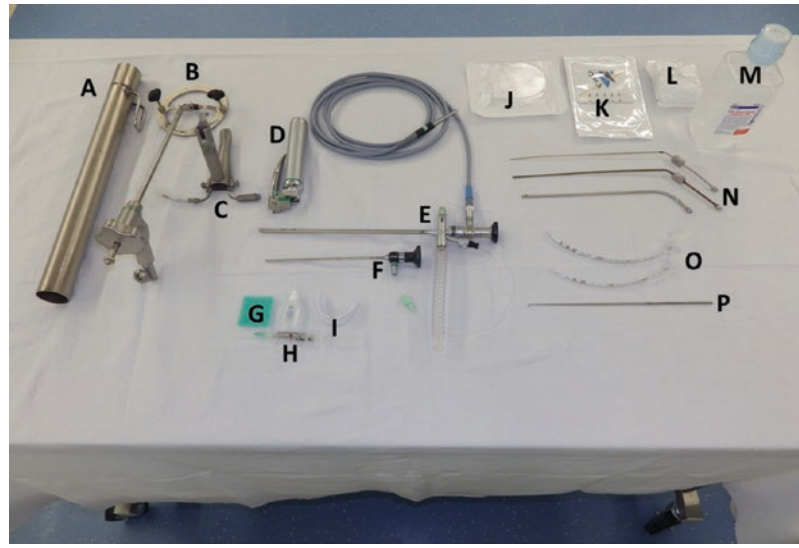

Fig. 2.3 Table setup for direct laryngoscopy and bronchoscopy for purpose of upper airway exam. Equipment shown includes quiver (**a**), suspension arm (**b**), Lindholm laryngoscope (**c**), Phillips laryngoscope (**d**), bronchoscope (**e**), additional telescope (**f**), defog (**g**), topical lidocaine (**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

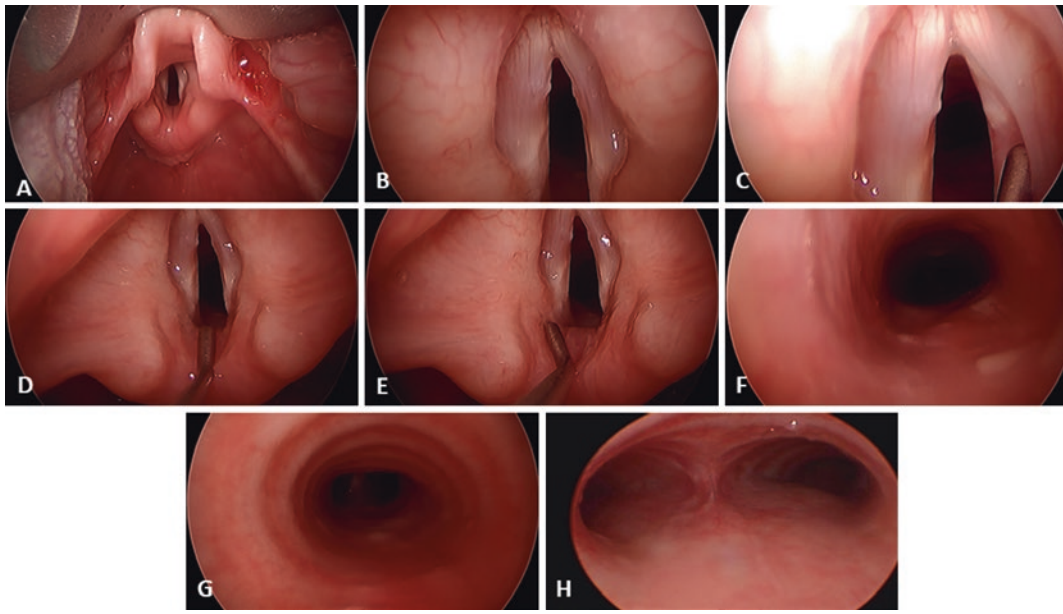

Fig. 2.4 Series of images recorded during standard direct laryngoscopy and bronchoscopy, including intubating view (**a**), close-up of glottis (**b**), use of right-angle probe to visualize infraglottic surface of right true vocal fold (**c**), palpation of interarytenoid region to rule out cleft (**d**),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