1 General Considerations in Pediatric Otolaryngology

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KEY POINTS

- Children are physiologically different from adults, especially in the early stages of life.
- The practice of pediatric otolaryngology relies on close collaboration with other disciplines to manage children with complex medical issues and often with complicated family networks.
- Pediatric otolaryngology often requires the unique resources and facilities of a dedicated pediatric hospital, including access to intensive care, anesthesiology, and other allied health professionals with specific expertise.
- The ongoing physical, psychological, and functional development of the child plays a critical role in decision making for whatever investigations and treatments may be under consideration.
- Gaining the trust of the child and his or her guardians through providing a supportive environment is essential for optimal patient care.
- With practice, physical examination can be conducted in an efficient and complete manner that remains relatively atraumatic for the child. Involving the child in play may alleviate anxiety.
- The demographics of the child's family unit is becoming an increasingly important factor in facilitating successful communication.
- Genetic disorders, psychosocial issues, and comorbidities may affect the complexity and timing of the management of otolaryngologic disorders in children.

INTRODUCTION

Pediatric otolaryngologists often care for children with complex issues, often in a pediatric institution capable of providing multidisciplinary care. As with other pediatric subspecialties, the mission of pediatric otolaryngology is to develop benchmarks for the care of children to improve outcomes for illnesses faced by children and their families.[1] As stated by Robin T. Cotton, MD, in delivering the 2014 Wullstein Lecture to the German Otolaryngology Society, "The aim of pediatric otolaryngology is to set a standard and not to create a monopoly" (Fig. 1.1). Through high-quality training and meaningful research, the subspecialty of pediatric otolaryngology is focused on a small number of difficult problems not often encountered in general otolaryngologic practice, with the goal of continuous improvement in providing excellent care for children.

Pediatric otolaryngology as a surgical specialty has matured but is continuing to evolve, given the continued growth of the human population and the resources devoted to establishing pediatric institutions throughout the world.[2,3] Within these institutions, the emergence of specialties such as pediatric anesthesiology, pediatric critical care medicine, and neonatology has necessitated the development of surgeons who have the technical and clinical skills to care for children of various ages. Institutions that house multiple subspecialties allow pediatric surgical specialists with other skilled colleagues to manage children, including the youngest children and those with critical and complex life-threatening illnesses, in a safe medical environment. The experience of these institutions is then disseminated to the rest of the community in other health care facilities, academic medical centers, training programs, and the general public.

This chapter introduces a variety of topics that are covered comprehensively elsewhere in this volume, and it provides an overview of the multidisciplinary aspects of pediatric care and its influence in the field of otolaryngology.

History of Pediatric Otolaryngology

The evolution of pediatric otolaryngology, like that of most subspecialties, began when a group of like-minded colleagues decided to share their experiences with the challenging scenarios each had faced.[2] The Society of Ear, Nose and Throat Advances in Children (SENTAC) was founded in 1973 as an interdisciplinary professional organization.[3] Similar groups were established in Poland (1947), Hungary (1948), Japan (1979), South America (1979), and Australia/New Zealand (1985).[4–8] These groups helped to define subspecialty training to include an in-depth knowledge of developmental physiology, growth mechanisms, and pediatric pharmacotherapy.[4] The pediatric otolaryngologist must also have a demeanor conducive to caring for young patients and their parents. It is generally accepted that pediatric otolaryngology is based on shared decision making with an emphasis on the best outcomes for the children and their guardians.

Hirschberg and colleagues recorded the rise of the specialty in Hungary from the 1890s onward; it arose because of the need for tracheotomy in children.[8] Infections were the major killers of children in the late 1800s, and diphtheria was responsible for significant mortality from death by suffocation. Literature on acute purulent otitis media was published as early as the late 1890s and early 1900s, and the first successful retrieval of an aerodigestive foreign body occurred in 1908. Lye ingestion in the 1930s was common, and individuals such as Chevalier Jackson, in the United States, were instrumental in campaigning for the labeling of all poisonous or corrosive substances to prevent accidental ingestion.[2] The high mortality from crouplike illnesses and foreign body aspirations led to the development of the "airway foreign body service" in the late 1950s in Hungary.[8] Similarly, courses on pediatric airway management such as the Polish "Days of Pediatric Laryngology" began[4] and continue to this day (see endodays2017.org). The management of acute airway emergencies continues to be the sine qua non of pediatric otolaryngology.

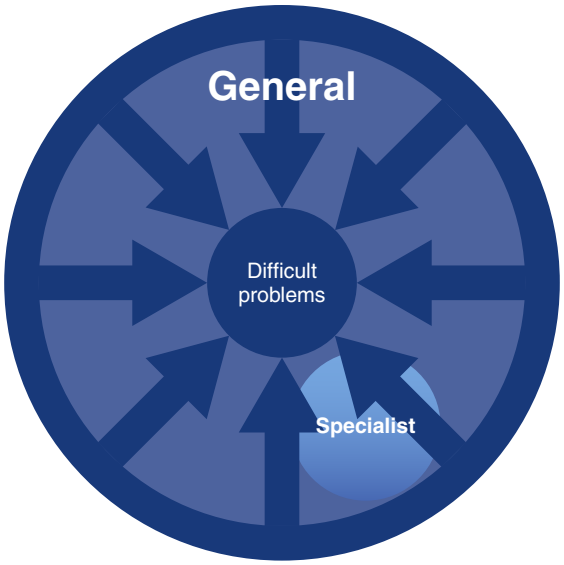

Fig. 1.1. Setting a standard of care. The aim is to set a standard and not to create a monopoly. (Adapted from Cotton R. *The Value of Pediatric Otolaryngology*. Wullstein lecture. Presentation to the German Society; May 2014.)

Severe neonatal and infantile rhinitis is frequently discussed at pediatric otolaryngology meetings, but better understanding came with the introduction, by Hounsfield and Cormack, of computed tomography (CT) in 1972. The developmental anatomy of the nose influences decisions made for treating conditions such as septal deformity, choanal atresia, cystic fibrosis, and genetic syndromes such as Treacher Collins and Crouzon. The ongoing evolution of imaging has enhanced the current use of telescopic miniaturization and camera enhancement in treating skull base lesions through the nostrils of tiny children, which was not possible 20 years earlier.

The Human Genome Project was completed in April 2003 (see www.genome.gov); it has led to the identification of the molecular etiology of conditions such as hereditary hearing loss (see www.hereditaryhearingloss.org), a better understanding of the chromosomal anomalies seen in Down syndrome and 22q11.2 deletion syndrome, and various craniofacial syndromes. Nearly 100 years after Gustave Crouzon first described a case in 1912, we now know that Crouzon syndrome results from mutations in the fibroblast growth factor receptor 2 (*FGFR2*) gene. Helpful Internet and established literature resources include www.genetics.org, www.orpha.net, *Online Mendelian Inheritance in Man* (see www.omim.org), and Smith's *Recognisable Patterns of Human Malformation*.

The role of the pediatric otolaryngologist in advocating for children's health is critically important in preventing illness. Pediatric otolaryngologists have participated in campaigns targeting excessive noise exposure and button battery ingestion, advocating for human papillomavirus (HPV) vaccination, and promoting screening for congenital human cytomegalovirus (HCMV) infection, childhood hearing loss, and speech and language delay. In a similar way, the Australian pediatric otolaryngology group promoted the benefits of installing swimming pools in remote Aboriginal communities to significantly reduce otitis media with tympanic membrane perforations and pyoderma in these communities.[9]

The Important Differences Between a Child and an Adult

Pediatric otolaryngologists may be called to evaluate children in the fetal stage after their initial ultrasound imaging, at the time of birth, as neonates or infants, or at any age thereafter up to transitional care in the middle teens. Children differ from adults in all four areas of development: craniofacial, physiologic, psychosocial, and intellectual. In addition, their underlying condition may be associated with developmental delay, presenting management challenges even into adulthood. The anxieties and concerns of the parents, along with their ability to cope and provide care, will also affect decision making. For children not fortunate enough to have parents to care for them, guardianship issues pertaining to cultural organizations, religious bodies, and governmental institutions add to the complexity.

TABLE 1.1 Arterial Oxygen Tension (Pao_2) in Healthy Infants and Children

Age	Pao_2 in Room Air (mm Hg)
0–1 week	70
1–10 months	85
4–8 years	90
12–16 years	96

Physiology

Physiologically, children have a greater ratio of body surface area to volume than adults, and as a result they are at greater risk of excessive loss of heat and fluids. Children are more easily affected by medication, toxins, and microorganisms. Children also have rapidly dividing cells, which assist in the rate of growth, making them quicker in healing but more sensitive to effects such as radiation or chemotherapy. With higher heart rates and respiratory rates, they have proportionately higher minute volumes; hence they are more susceptible to agents absorbed through the airway, responding more quickly to a variety of agents. With an immature blood-brain barrier, children have enhanced central nervous system receptivity; thus they have a higher prevalence for neurologic symptoms from illness and medication. Because of the immaturity of their neurologic system, children have exaggerated responses to insults such as laryngospasm and central apnea. The higher metabolic rate in children means a greater risk for increased fluid loss as a result of stress or illness. The dosage of most medications for children, unlike those for adults, is based on the child's weight and body size. Children generally are at greater risk for infection and have less herd immunity from certain infections because of their immature immune systems.

Ventilation/Perfusion Relationships

Ventilation and perfusion are imperfectly matched in the neonatal lungs because of persistent anatomic shunts in the newborn circulatory system and a relatively high closing volume in the lungs. A newborn breathing room air has a normal arterial oxygen tension of 50 mm Hg. During the first 24 hours of life, the arterial oxygen tension increases dramatically with changes in the fetal circulation and the maturation of lung parenchyma; then, during the ensuing months and years, it continues to change slowly (Table 1.1).

Newborn Heart and Cardiac Output

The heart of a healthy neonate is quite different from that of an adult. The thickness of the right ventricle exceeds that of the left, as seen by the normal right-axis deviation on the neonatal electrocardiogram. Shortly after birth, with closure of the fetal circulation, the left ventricle enlarges disproportionately. By the age of 6 months, the adult right/left ventricular size ratio

TABLE 1.2 Normal Heart Rate for Children by Age

Age	Heart Rate (Beats/Min)	
	Average	Range
Newborn	120	100–170
1–11 months	120	80–160
2 years	120	80–160
4 years	100	80–120
6 years	100	75–115
10 years	90	70–110

is established. The newborn myocardium is also significantly different from that of the adult. Cardiac output is rate dependent in the neonatal heart, which has reduced compliance and contractility. The low compliance of the relaxed ventricle limits the size of the stroke volume; therefore increases in preload are not as important in neonatal physiology as the heart rate. Bradycardia invariably equates with reduced cardiac output because the infant heart cannot achieve the increased contractility needed to maintain cardiac output. It is extremely important to recognize this distinction during surgical and anesthetic procedures that may induce bradycardia. Autonomic innervation is also incomplete in the neonatal heart, with its relative lack of sympathetic elements; this relative underdevelopment may further compromise the ability of the less contractile neonatal myocardium to respond to stress.

Heart rate is crucially important in the very young. The normal range for the newborn is 100 to 170 beats/min and the rhythm is regular. As the child grows, the heart rate decreases (Table 1.2). Sinus arrhythmia is common in children, but all other irregular rhythms should be considered abnormal. The average newborn systolic blood pressure is 60 mm Hg; the diastolic pressure is 35 mm Hg.

Blood Volume

Because the total blood volume of an infant is small, relatively minor surgical blood loss may be hemodynamically significant. It has been observed during exchange transfusions that withdrawal of blood parallels a decline in systolic blood pressure and cardiac output. Replacement of the same blood volume that was removed can reverse this decline to restore normal parameters. When the heart rate is normal, changes in arterial blood pressure are thus proportional to the degree of hypovolemia. A newborn's ability to adapt the intravascular volume to the available blood volume is limited because of less efficient control of capacitance vessels and immature or ineffective baroreceptors.

Neonatal blood volume is approximately 80 mL/kg at term and is notably 20% higher in preterm infants. The hematocrit is 60%, and the hemoglobin content is 18 g/100 mL, although these values vary by infant and depend on when the umbilical cord is clamped. After remaining stable during the first week of life, the hemoglobin level declines, with the change occurring more rapidly in preterm infants. Approximately 70% to 90% of the hemoglobin in a full-term infant is of the fetal type. Fetal hemoglobin has a higher affinity for oxygen than that of an adult. It combines with oxygen more readily but also releases oxygen less efficiently at the tissue level compared with adult hemoglobin. The increase in hemoglobin content in neonates is required to overcome this higher affinity of fetal hemoglobin for oxygen. A concentration less than 12 g/100 mL constitutes anemia. Correction of anemia by blood transfusion is indicated if the infant requires oxygen or experiences apnea.

During the first weeks of life, the hematocrit drops as a result of early suppression of erythropoiesis; the fetal type of hemoglobin is replaced with the adult type of hemoglobin with more optimal oxygen-carrying capacity. This physiologic anemia reaches its nadir at 2 to 3 months, with a hemoglobin content of 9 to 11 g/100 mL. Provided that nutrition is adequate, the hemoglobin level will then gradually rise over several weeks to 12 to 13 g/100 mL, which is maintained throughout childhood.

TABLE 1.3 Ideal Electrolyte Composition for Infants

	Na^+ (mEq/L)	K^+ (mEq/L)
Intracellular	10	150
Extracellular	140	4.5

Response to Hypoxia

Because neonates have a relatively high metabolic rate and low reserve for gas exchange, hypoxemia develops rapidly, first manifesting as bradycardia. During surgery, any unexplained episode of bradycardia should initially be treated with oxygen and increased ventilation. Pulmonary vasoconstriction and hypertension occur more dramatically in response to hypoxemia in the neonate than in the adult. With a patent foramen ovale and/or a persistent patent ductus arteriosus (PDA), the increase in pulmonary vascular resistance may favor a shift into fetal circulation with right-to-left shunting, which compounds the problem. Changes in cardiac output and systemic vascular resistance also differ in neonates compared with older children and adults. During hypoxemia in adults, the principal response is systemic vasodilation; this, together with an increase in cardiac output, helps to maintain oxygen transport to the tissues. Fetuses and some neonates respond to hypoxemia with systemic vasoconstriction. In the fetus, hypoxemia shifts blood to the placenta to improve gas exchange and oxygenation. After birth, however, hypoxemia may lead to decreased cardiac output, further limiting oxygen delivery and increasing cardiac work. In infants, early and pronounced bradycardia may result from myocardial hypoxia and acidosis. Neonates exposed to hypoxemia suffer pulmonary and systemic vasoconstriction, decreased cardiac output, and bradycardia. Rapid recognition and intervention are necessary to prevent cardiopulmonary collapse, cardiac arrest, and death.

Fluids and Fluid Management

As in adults, preoperative, intraoperative, and postoperative fluid management is extremely important in children; extreme vigilance, early recognition, and tight control are required. Some of the physiologic differences outlined earlier make fluid administration even more critical. Because of their small intravascular volume (70 to 80 mL/kg), infants who experience small changes in fluid balance can easily become either dehydrated or overloaded with fluid. The compartmentalization of total body water changes with age, but intracellular and extracellular electrolyte composition remains stable (Table 1.3). Maintenance fluid requirements for a child can be calculated by relatively simple formulas that vary according to metabolic and physical activity. The calculation of water loss per calorie is described in Table 1.4. The correspondence of necessary fluid intake proportional to weight is described in Table 1.5. Complex fluid and electrolyte deficits are beyond the scope of this discussion. In most cases, consultation with pediatric medical specialists is advised.

Pain Management

The management of pain in infants and children has undergone tremendous advances in recent years. It had been

TABLE 1.4 Expected Fluid Losses in Children

System	Fluid Loss (mL/100 cal/day)
SENSIBLE LOSSES	
Kidneys	55
INSENSIBLE LOSSES	
Lung	15
Skin	30
Total	100

TABLE 1.5 Children's Maintenance Fluid Intake Calculated by Weight

Weight	Estimated Fluid Intake
0–10 kg	4 mL/kg/h
11–20 kg	2 mL/kg/h (for second 10 kg) + 40 mL/h
>20 kg	1 mL/kg/h (for every kg above 20 kg) + 60 mL/h

Data from the American College of Surgeons. *Optimal Resources for Children's Surgical Care, V.1*; 2015.

commonly believed that because of their immature nervous systems, infants and newborns did not perceive pain and would not remember any pain that occurred. However, direct physiologic consequences have been observed in infants in response to pain. Changes in heart rate, blood pressure, and respiration rate have been documented in infants experiencing painful stimuli, and such changes can be physiologically and emotionally deleterious to the child.

The perception of pain depends on both sensory and emotional experiences that may be altered by various psychologic factors. These factors may be specific for each individual patient based on his or her expectations and past experiences. Efforts to reduce stress, anxiety, and fear will help to decrease the apprehension and perception of pain during procedures in the office or in the operating room. In patients of appropriate age, relaxation techniques, such as guided imagery, deep breathing, and hypnosis, may diminish the emotional component of pain. An adequate and age-appropriate explanation of expectations will also reduce anxiety, increase cooperation, and decrease perceived pain. The caregiver or parent should also be coached and prepared, because children often look to the psychological state of a parent for cues. An anxious parent often increases the anxiety of the child. Conversely, a calm and collected parent can help calm the child during uncomfortable procedures. The propagation of child life programs and patient-centered/family-centered care principles in pediatric institutions has transformed care in this regard.

Nonnarcotic analgesics are helpful for pain management. Acetaminophen in doses that range from 10 to 15 mg/kg every 4 hours is useful. Nonsteroidal antiinflammatory drugs such as ibuprofen are also excellent for pain management but may inhibit platelet function and should be used only at the discretion of the surgeon.

Narcotic analgesics are indicated for moderate to severe pain in all age groups. Optimal use requires consideration of the needs of the individual patient, and neonates require special observation during the administration of narcotics. Ventilation responses to hypoxia and hypercarbia are diminished in this age group. Narcotics may further decrease these responses to potentially life-threatening levels. The metabolism and half-lives of narcotics are different in neonates than those in older children and adults, and the permeability of the blood-brain barrier may also be increased in neonates. However, the use of intravenous, intramuscular, and oral narcotics is safe in the appropriately monitored setting and with proper dose. Unlike adults, who usually self-administer and therefore self-regulate narcotic analgesics, pediatric patients often rely on caregivers to administer pain medication, which can lead to under dosing or over dosing.

Sedation

More often today, professional certification and credentialing for pediatric sedation are required at institutions that care for children. The development of sedation teams, often staffed by pediatric anesthesiologists or critical care specialists, has increased safety and monitoring for these patients. Otolaryngologists can participate in the team effort by assessing the airway of the patient referred for sedation. Occasionally the best way to ensure the safety of a sedated patient is by general anesthesia with a secured airway. To achieve a successful outcome as measured by family satisfaction and patient safety, it is important to adhere to the guidelines of the institution and to work together as a team.

The Faces of Children

The development of the human face results from 500 million years of progressive evolution. In the embryo and fetus, the face takes shape in an impossibly rapid sequence, and any interference with this process can have catastrophic consequences. Thus an understanding of the structural and physiologic processes underlying normal development is required to evaluate and manage craniofacial anomalies. Parents and caregivers are greatly relieved on meeting a clinician who is familiar with the constellation of special clinical features associated with their child's syndrome.

Rapid differential growth of the head and neck in four dimensions presents challenges for the pediatric otolaryngologist. The frontonasal prominence and the three paired structures—the lateral nasal and maxillary and mandibular prominences—all develop at differing rates according to a preprogrammed sequence, with rate changes affected by tissue interactions and environmental events. The timing of surgical intervention must consider the normal course of development in all affected systems to minimize the long-term functional and cosmetic sequelae, particularly for cleft lip and palate (Chapter 188) and congenital anomalies of the nose and nasopharynx (Chapter 190). For example, the solitary median maxillary central incisor syndrome (Fig. 1.2A) is associated with pyriform aperture stenosis (see Fig. 1.2B), midnasal stenosis, choanal stenosis, holoprosencephaly, absence of the corpus callosum, diabetes insipidus, microcephaly, hypotelorism, cervical hemivertebrae, congenital heart disease, cervical dermoid, hypothyroidism, and intellectual delay.[10]

Development of the face, head, and neck involves cellular differentiation, proliferation, and migration. The branchial arches and their associated vasculature, neural structures, and musculature develop quickly in the fetus but continue to change in the postnatal period until the individual reaches adulthood. Recognizing the two types of bone, endochondral (of cartilaginous origin) and mesenchymal (of membranous origin), provides insight into hereditary disorders. For example, achondroplasia is primarily a disease of endochondral bone; it results from mutations in the *FGFR3* gene; and manifestations include disproportionate dwarfism, frontal bossing, flattened nasal bridge, and a small face.

In the postnatal individual, ongoing growth and development of the human head and neck is largely governed by the need to maintain an airway for respiration and an aerodigestive tract for nutrition. The development of other organ systems of eye, ear, nose, and mouth—which determine vision,

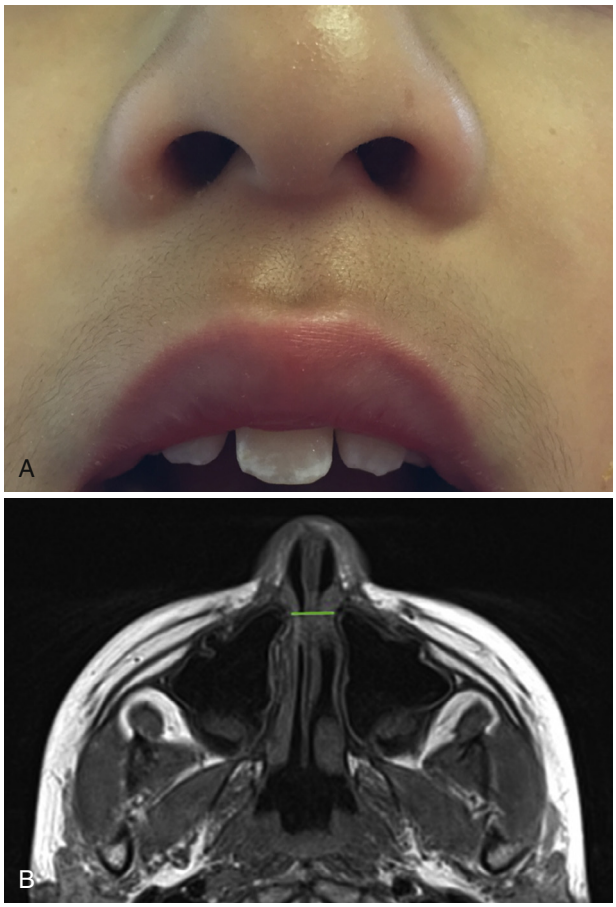

Fig. 1.2. (A and B) Midline incisor associated with confirmed computed tomography evidence of pyriform aperture stenosis (<11 mm).

hearing, balance, smell, and speech—is programmed by developmental control genes, peptide growth factors, and cytoskeletal elements that govern displacement and remodeling. The various growth centers within the quadrilateral cartilage of the nasal septum illustrate the importance of understanding how trauma or surgery can lead to differential growth of the nose, influence the surrounding bony structures, and subsequently functionally affect the rest of the facial skeleton.[11] It is important to appreciate the role of the septodorsal cartilage and how simple fractures or incisions may affect cartilage growth and the normal development of the premaxillary-maxillary sutures. An age-specific anatomy of the nasal skeleton has been defined in children and adolescents, and peak growth velocity differs between boys and girls. Verwoerd and colleagues[11] reflected on the importance of subcategorizing the pediatric age group to define optimal timing for surgical intervention and the appropriate surgical technique that will not affect long-term function detrimentally.

Respiratory System

Control of Ventilation

Emergence from an underwater world, in which the child relied on placental exchange of gases and nutrition, induces tremendous changes in the neonate's physiology and anatomy. Pulmonary blood flow is relatively restricted in utero by relative fetal hypoxia, which radically changes when the establishment of respiration improves oxygen content and increases pulmonary blood flow. Consequently, unusual blood vessels may develop and cause extrinsic compression of the airway (e.g., vascular rings or slings).

The chest wall of the neonate stabilizes a compliant rib cage whereas the neonatal lung accounts for only 10% to 15% of the total lung capacity, especially in the premature child. The functional residual capacity of the neonatal lung may be increased in several ways, including "expiratory braking" (the use of active glottic expiration), active use of inspiratory muscles during expiration, and rapid respiratory rates.[12] Lack of these reflexes, as in the child with bilateral vocal fold impairment, produces biphasic stridor and rapid decompensation. This may require immediate intubation or possible noninvasive ventilation (NIV) to maintain a patent airway. However, in a neurologically functioning airway, a child's physiology also permits general anesthesia with spontaneous respiration without endotracheal intubation, a common practice in pediatric units during the performance of a microlaryngoscopy and bronchoscopy (see Chapter 184).[13]

Measurements of ventilation to assess respiratory drive depend on the assumption that the respiratory muscles are capable of converting this drive into work, which is not always the case in infants and neonates. Muscle fibers are classified as type I fibers, which are red in color, slow twitch, highly oxidative, and fatigue resistant; type IIA fibers, which are also red in color but are fast twitch and oxidative in nature; and type IIB fibers, which are white in color, fast twitch, and glycolytic in nature. Type IIB fibers are most easily fatigable, and type IIA are less fatigable.[14] Newborns have a paucity of type I muscle fibers but develop them shortly after birth. The muscle fibers in the diaphragm of a preterm infant are composed of less than 10% of type I fibers, whereas the muscle fibers of a full-term infant may be 30% type I fibers; the percentage of type I fibers increases to 55%, the expected adult level, during the first year of life.[15,16] Preterm infants are more prone to respiratory fatigue, although this predilection disappears as they reach maturity.

Other subtle differences in sleep patterns also affect the control of ventilation (see Chapter 186). Preterm infants spend as much as 50% to 60% of their sleep state in the rapid eye movement (REM) state. During REM sleep, the intercostal muscles are inhibited, as are most of the other skeletal muscles, which places a greater burden on diaphragmatic activity. Much of this activity may be wasted because of paradoxic motion in the very young; purposeless movement readily leads to hypoventilation, increased respiratory drive, and diaphragmatic fatigue.

Full-term neonates have biochemical and reflex controls of ventilation that are similar to those of adults, although incompletely developed.[17] Neonates have a higher basal metabolic rate than adults, reflected in higher ventilatory rates relative to body mass at any given partial pressure of carbon dioxide ($PaCO_2$). In term infants and adults, an increased $PaCO_2$ results in a proportionately similar increase in ventilation; however, preterm infants do not exhibit this response. Compared with full-term infants and adults, preterm infants have a blunted response to increases in $PaCO_2$ and an altered response to changes in partial pressure of oxygen. The administration of 100% oxygen decreases ventilation in the very young, suggesting the existence of chemoreceptor activity not generally seen in adults.[17]

Gestational age, postnatal age, body temperature, and sleep state affect the ventilatory response of newborns to hypoxia. Preterm and full-term infants less than a week old who are awake and euthermic usually demonstrate biphasic breathing patterns. They often experience tachypnea followed by hypoventilation. Hypothermic infants demonstrate a blunted response to hypoxia with respiratory depression without initial hyperpnea. The central effects of hypoxia on the respiratory center may cause depression of ventilation. Active peripheral

chemoreceptors are unable to maintain a significant influence over this response. REM sleep also may decrease the response to hypoxia in these infants, whereas sleep states other than REM sleep are associated with an increase in the ventilatory response to hypoxia. Arousal from sleep during hypoxia is not seen in newborns, but with further maturation of the chemoreceptors during the first few weeks of life, the ventilatory drive to hypoxia is increased. Of interest, a decreased response to hypercarbia associated with hypoxia also occurs in newborns but not in older infants and adults.

Ventilation Reflexes. Reflexes that arise from the lung and chest wall are essential in maintaining ventilation in newborns and in determining the respiratory tidal volume. Periodic breathing is thought to result from dyscoordination of the feedback loops that control ventilation. It is characterized by periods of alternating rapid ventilation followed by apnea and is common in both preterm and full-term infants. During the apneic part of periodic breathing, the $Paco_2$ may increase but changes in heart rate do not occur. Generally, periodic breathing is considered normal without serious physiologic consequences and typically resolves by 6 years of age. Some preterm infants, however, may demonstrate serious and potentially life-threatening apnea episodes, which may be accompanied by bradycardia and last longer than 20 seconds. Apneic episodes may represent a failed response to hypoxia. Because these episodes are more commonly seen during REM sleep, they may be caused by ventilatory fatigue and an impaired chemoreceptor response to hypoxia. Usually stimulation is all that is required to terminate the apneic event. Aminophylline or caffeine treatment generally decreases the apneic episodes through central stimulation. Continuous positive airway pressure (CPAP) may also be helpful to decrease apneic episodes by modifying the lung and chest wall reflexes.

Laryngeal Sensitivity. The primary function of the larynx is to protect the lungs from aspiration. Hence the laryngeal adductor response is a very strong reflex that may even be lethal in some instances. The robustness of the reflex has been shown to change with age and maturity in animal models. When laryngeal adduction is coupled with tachycardia, hypertension, and apnea, it is termed the *laryngeal chemoreflex* (LCR). The LCR can be induced by laryngeal exposure to acid, base, and pressure, but it is most sensitive to water and is ablated by saline. These properties of the LCR may have implications for sudden infant death syndrome (SIDS). Responses to hypoxia, hypercarbia, and other stimuli evolve with age. Deaths attributed to SIDS have dramatically decreased with the change in recommended sleep position from prone to supine. Elimination of hypercarbia from rebreathing may be the mechanism behind the decrease in SIDS deaths, and hypercarbia is a known potentiator of the LCR. An increased awareness and more aggressive treatment of infant reflux may have reduced the incidence of infant apnea as mediated by the LCR by mitigating one potential stimulus. Decreased bolus size, increased feeding frequency, frequent burping, and positioning to reduce regurgitation have been helpful in this regard.

Laryngospasm is produced by the forced closure of the aryepiglottic folds, false vocal cords, and true vocal cords. Prolonged spasms can be serious, causing fatal cardiac and cerebral complications. The incidence of laryngospasm is quoted at 8 to 9 per 1000 in adults and 27 to 28 per 1000 in children undergoing anesthesia.[18] The risks are greater in certain situations such as those involving infants 1 to 3 years of age; during certain airway procedures such as tonsillectomy; and in the presence of airway inflammation, such as respiratory tract infections (RTIs).[19] The preventive measures commonly employed include (1) avoidance of intubation anesthesia in elective cases for children who have had a recent RTI or asthmatic attack, allowing 4 to 6 weeks for airway hyperreactivity to abate; (2) use of topical lidocaine 4% to the airway prior to short oral endoscopic procedures; (3) the "no-touch" technique on emergence, avoiding any disturbance or stimulation of the child until he or she is fully awake; (4) preventing buckling or kinking of an endotracheal tube in situ; (5) placing the child in the lateral recovery position after careful suctioning; and (6) using positive airway pressure during extubation, hence reducing the adductor response of the laryngeal muscles and inducing a forced exhalation and expulsion of secretions or blood at the larynx.

In emergency situations involving smaller children, who tend to deteriorate rapidly, initial management includes removing the irritant stimulus, deepening the level of anesthesia, applying the jaw thrust maneuver, and inserting an oral or nasal airway. The general steps advocated include the "four Ps": positive airway pressure with 100% oxygen; painful stimulus with forward displacement of the mandible and tongue resulting in the relaxation of the vocal cords via the autonomic nervous system; propofol in subhypnotic doses of 0.25 to 0.8 mg/kg for rapid onset of action; and paralysis with succinylcholine (suxamethonium) 0.1 to 0.3 mg/kg intravenously or 4 mg/kg intramuscularly/intralingually, followed by mask ventilation or tracheal intubation. If these methods fail, emergency tracheotomy or cricothyrotomy may be required.

Lung Volumes

In proportion to body size, the total lung capacity, functional residual capacity, and tidal volume are roughly equivalent in adults and infants. In the full-term infant, the total lung volume is approximately 160 mL with functional residual capacity at half this volume. Tidal volume is approximately 16 mL, and dead space is approximately 5 mL. Because of infants' small lung volumes, any increase in dead space is much more significant than that in adults. In contrast with static lung volumes, however, alveolar ventilation is proportionately much greater in newborns (100 to 150 mL/kg body weight) than in adults (60 mL/kg body weight). This higher alveolar ventilation in infants results in an alveolar–functional residual capacity ratio of 5:1, compared with 1.5:1 in adults. Consequently the functional residual capacity is unable to provide the same buffer in infants, such that changes in the concentration of inspired gases are much more rapidly reflected in alveolar and arterial levels. Thus induction of anesthesia using inhalational techniques is easier in infants than in adults.

The differences in lung volumes also explain why infants have a smaller ventilatory reserve. Time from apnea to oxygen desaturation is much shorter in infants than in adults. Accordingly, surgical maneuvers requiring short apneic periods pose more of a challenge in infants than in adults. The total surface area of the air-tissue interface of alveoli in infants is small (2.8 m^2). When this relatively small gas exchange area is combined with a higher relative metabolic rate, infants have a reduced reserve capacity for gas exchange. When congenital defects interfere with lung growth and development or if lung parenchyma becomes damaged, the remaining healthy lung may not be adequate to sustain life.

Respiratory Rate

The most efficient respiratory rate for newborns has been calculated to be around 37 breaths/min, which is close to that observed for average newborns. Full-term infants are similar to adults in requiring approximately 1% of their metabolic energy to maintain ventilation. The "cost" of breathing is then 0.5 mL/0.5 L of ventilation, whereas it nearly doubles to 0.9

mL/0.5 L of ventilation in preterm infants. The cost of respiration dramatically increases if the lung parenchyma is diseased or damaged by processes other than prematurity. In either case, it results in higher caloric and nutritional demands. Respiratory rate can directly affect the infant's ability to complete the suck-swallow-breathe cycle as well. If gas exchange is poor, ventilation rates may increase, which may not allow adequate time for the suck-swallow portion of feeding. Caloric intake decreases precipitously as the infant expends energy toward breathing over feeding, which may lead to a vicious cycle resulting in failure to thrive.

PEDIATRIC OTOLARYNGOLOGY PRACTICE

Referral Sources

Compared with adults, pediatric patients are more likely to have a primary care physician, the pediatrician or family practice physician, who may refer patients for specialty evaluation. Pediatric medicine is a specialty that legitimately considers its practitioners to be advocates for their patients. The child's primary care physician is responsible for his or her overall health and well-being, including preventive medical procedures such as vaccination. The primary care provider can serve as a tremendous ally to help identify and refer problems early on and help educate the family regarding issues related to surgical management. Although every patient encounter requires an element of trust building and rapport development, a long-term relationship usually has already developed between the child's doctor and the family.

The culture of pediatric medicine and its interface with surgery differs somewhat from its counterpart in the typical adult world. The perioperative care of the child more often requires a multidisciplinary approach, which can enhance the care of the patient as long as there is open communication and clear delineation of duties and responsibilities. All parties involved must understand that the surgeon is ultimately responsible for the care of any patient undergoing surgery, even if the caregivers have developed expectations for surgery prior to evaluation by the otolaryngologist. The parents and the referring physician should support the decision to proceed.

Patients

Although the entire family is involved in the care of a child, it is important to focus on the child as the patient. Children experience the same fears of pain and discomfort as adults but may lack the maturity and skills to overcome these fears. In particular, children may not understand the concept of delayed gratification. Whereas adults often bring themselves to the doctor and therefore have overcome the first hurdle in obtaining medical care, children are brought by their parents and often have no idea why they are there. Establishing rapport with the child is important for many reasons, not only to put the child at ease but also to put the parents at ease. This is accomplished when they see that the physician has a genuine interest in the well-being of their child. It is important to explain what is to be expected during the history and physical examination, not only to the parent but also to the child, on a level that the child can understand. Procedures that may be uncomfortable are best left to the end of the examination, to be performed quickly but thoroughly. Speed and experience allow for a brief but relatively complete physical examination without undue stress on the child or the parent. Restraining an older child who may have been traumatized by a previous forceful examination is not advisable and could be dangerous. In these cases, sedation or a general anesthetic is appropriate to complete the required examination.

Parents

Parents may experience unease or anxiety when they are required to make decisions for their sick child. It is important for the surgeon to be open and honest regarding the options and risks involved and the potential discomfort. Although there may be a single best option from the surgeon's point of view, alternative treatments with their respective risks and benefits and expected outcomes should be explained to the family. The reasons why a surgeon considers one option better than others should be shared, taking the time needed to answer questions. Parents often have questions that may be biased by previous experiences, advice of family members or friends, or exposure to the media.

Often, families have been referred by the primary care physician, who usually has answered many of the relevant questions. Having been referred to a specialist, the parents often identify the otolaryngologist as a person with expertise and added ability. Some parents may have difficulty making a decision, even after all of their questions have been answered; in this case, it may be helpful to suggest that they contact their primary care physician for his or her perspective. Finally, it may be more comfortable for families to defer a decision regarding surgery until they have been able to discuss it at home, with the understanding that they can call if more questions arise and that they will schedule an appointment if they wish to proceed.

History

It is important to obtain a general family history, a history of any diseases or illnesses that affect the child's siblings, and any issues involving the mother's pregnancy and/or the subsequent birth of the child. Insight into family stresses or school problems may influence the child's psychosocial development. As the provider who cares for children must rely on the parents as proxies to provide information, historical data should be carefully obtained. A referral letter from the primary care physician will often accompany the child, and though this is important, it does not replace the history taken from the parents or the child. Historical data from the primary physician verify the information provided by others and ensure that they and the data gathered during the examination are in agreement. The parents are usually very astute regarding their child's behaviors or symptoms and should therefore be listened to carefully, recognizing that one parent may embellish the symptoms whereas the other may play them down. When appropriate in terms of the child's age, the child should be questioned as well, although he or she may not understand or be able to identify a problem. Often chronic symptoms such as velopharyngeal insufficiency may be lifelong symptoms and are not seen as problems by the child or sometimes even by the parents. Sometimes the pediatric otolaryngologist may have to synthesize conflicting data into a coherent story. Occasionally admission to the hospital for observation is required to allow for the further collection of data, where symptoms described by the caregiver can then be documented and perhaps better understood. Further investigations may document whether the child's safety is compromised.

Physical Examination

Physical examination of a child should be done in a nonthreatening and calm manner. Rapport and trust are the key elements, and engaging the child as an active participant often leads to more cooperation. Letting the child hold or examine the instruments before they are used may reduce the child's anxiety. Because many well-child examinations are accompanied by vaccinations, children may unfortunately often equate any visit to

the doctor with a shot. The resulting apprehension, and in some cases an attempt at maintaining independence or control, may underlie seemingly irrational behavior during an examination.

The majority of the head and neck examination should be painless, and otoscopy generally is managed without difficulty; pneumatic otoscopy can be performed but should be explained before proceeding so as not to surprise the child (Fig. 1.3A). Explanations build a child's trust in the examining physician. The use of a head mirror or headlamp during the examination is the surgeon's choice. Equipment that covers the examiner's face often frightens children, which can decrease cooperation. Anterior rhinoscopy can be performed with the otoscope (see Fig. 1.3B) and a larger speculum, eliminating the need to introduce a new instrument to the child. Oral examination can often be accomplished without a tongue depressor, although occasionally one may be required to view the posterior pharynx. Palpation of the neck can be accomplished without any significant discomfort to the child, noting any masses or tenderness.

During the examination, further observations of the child's facial features can be made, which include assessment of the auricles (for shape, pits, or fistulas) and the eyes for symmetry in shape and alignment without apparent hypertelorism or heterochromia. The skin may be assessed for café-au-lait spots, a hallmark of neurofibromatosis type I, or vascular anomalies. The nasal airway should be assessed for chronic mouth breathing or hyponasality, and speech should be assessed for hypernasality to help identify any abnormality of the palate. Listening to the child's breathing for the sounds of stridor or stertor can help to identify the level or site of potential airway obstruction. Auscultation of the neck and chest may also be performed. The otolaryngologist will concentrate the physical examination on the head and neck, but elements of the general physical examination should be performed as indicated.

Microscopic Examination of the Ear

Otomicroscopy is an invaluable tool for assessing the ears, although children may have anxiety about this portion of the examination. Often, allowing the child to look at the examiner's thumb through the microscope will show that the microscope is simply a tool to obtain a magnified view of the ear. The parent can assist in reminding the child to hold still for the examination while cerumen or debris is removed from the ear canal. With the parent reclined on the examination chair or table, the child may lie on the parent's lap. The parent can then hug the child to hold the arms and body still while an assistant helps to steady the head. Use of a papoose board is possible with smaller children but often is not successful with older children. Sedation is usually not necessary but may be used in certain cases in accordance with the sedation policies of the institution. Occasionally a general anesthetic is required to permit a complete and thorough examination. Although usually not painful, suctioning is loud and can be frightening. The suction device is a very useful tool to quickly clean away debris and drainage, although some clinicians prefer to use cotton-tipped applicators instead. It is important to have a number of techniques in reserve and to use the one that will quickly and effectively accomplish the required task without undue stress and discomfort to the child. Other techniques to allay fears include warming one's hands by washing them with warm water, getting on the floor to examine the child, or distracting the child with toys or devices that play videos, such as smartphones or electronic tablets.

Flexible Endoscopy

Nasopharyngoscopy is an invaluable tool for the otolaryngologist. With advances in technology, smaller-caliber

Fig. 1.3. (A and B) Otoscopy and anterior rhinoscopy performed in an unhurried and nonthreatening manner with a well-illuminated otoscope may visualize the eardrum and nasopharynx, respectively, allowing the eye to accommodate to the initially dark environment.

endoscopes with improving optics are being developed. The fiberoptic endoscope allows for a dynamic examination of the nose, velum, pharynx, hypopharynx, and larynx. When this is combined with video recording, slow-motion replay is possible. As in adults, topical anesthesia with lidocaine gel or tetracaine spray and nasal decongestion with oxymetazoline is very useful. However, excessive doses of lidocaine can impair protective airway reflexes in young children or in those with hypotonia at risk for aspiration, such as children with cerebral palsy. Cocaine and stronger doses of lidocaine in young children are not advisable because of variable absorption rates, producing unwanted irritability and nervousness. The parents should be counseled that the child should not eat or drink for approximately 30 minutes after receiving the topical anesthetic. Sedation generally is not necessary for this examination. Complications from nasopharyngoscopy are rare and nasal bleeding, if it occurs, is often self-limited or requires little intervention. The patient care area should have an emergency crash cart available, as specified by regulatory agencies.

Audiology

There are a variety of techniques to evaluate hearing, which can be accomplished in any child at any age. Pediatric otolaryngologists can help to educate both families and primary care physicians to help avoid delays in diagnosis. Audiologists with expertise in pediatric testing are generally able to evaluate children as young as 6 months of age with visual response audiometry, which in experienced hands can be highly reliable. Other techniques for younger children or children who are unable to conform to the demands of the task include distortion product otoacoustic emissions and auditory brainstem response testing. If a screening test identifies that the child requires further investigation, it is essential for the otolaryngologist to provide any necessary referrals and follow-up evaluations.

Preparation for Hospitalization and Surgery

Once the decision to proceed with hospitalization or surgery has been made, successful outcomes are optimized by preparing the child and parents emotionally. The details and expectations should be reviewed with the family by knowledgeable personnel. Most children's hospitals and some other institutions have programs to introduce the family to the hospital, operating room, and operating room procedures. A sense of familiarity and an understanding of what to expect is helpful to all involved. Children can then role play before hospital admission or surgery. With a clear understanding of what is expected, parents also are less apprehensive and will be able to help calm the child. It is important to clarify arrival times and the nothing-by-mouth requirements to the family. Patient-friendly special films and booklets are valuable tools for education. The parents must be encouraged to be truthful, to the best of their ability, about preparation for the upcoming surgery. It is also important that the surgeon communicate directly with both the parents and the child so that all questions are answered completely.

Hospital

The staff involved with pediatric surgical care should be comfortable working with children and their caregivers. Ensuring an optimal environment for pediatric treatment encourages the best outcomes and leads to the greatest satisfaction among patients and their families.[20] The concept of a team of professionals who deal strictly with children is not new; moreover, a team approach is becoming increasingly accepted as the standard of care. When complications arise, as they inevitably do in the course of even the best medical care, having support facilities and personnel available to promptly handle any need is vital for the best possible outcomes.

Selecting Anesthesia

Pediatric anesthesia is a subspecialty in itself. Reaching a degree of proficiency to deliver an anesthetic to children of all ages requires considerable training and experience. The younger the patient, the more complex the problems encountered. As in all aspects of medicine, this subspecialty includes practitioners with special interests and expertise in dealing with children, who have smaller body volumes and lung fields to handle anesthesia. Specialists with expertise in safely delivering anesthesia to children should be sought when assistance is required, and reliance on these specialists benefits both the patient and surgeon. Clear communication and attitudes of respect among the surgeon, the pediatric anesthetist, and the rest of the operating room staff about the exact procedure planned is critical prior to the procedure, and the exact techniques that are employed by each member of the team should be made clear. Thus the best outcome for the patient is achieved and any complications will be readily managed with efficiency and effectiveness.

Postoperative Management

The most important part of postoperative management is preoperative education. The parent and child will have been adequately prepared regarding the effects of surgery so that they can approach the postoperative period with decreased stress and optimal adherence to the recommended postoperative regimen. Detailed teaching about the expected clinical course and unexpected possibilities is important. Providing written instructions is very helpful, along with contact telephone numbers if the parents should have questions. Knowing what to expect is the single most important factor in achieving an uneventful recovery from a surgical procedure without the need for multiple phone calls and excessive concern from both patient and physician. If a child must be hospitalized, it has been shown that the presence of a parent makes the stay less fearsome and stressful for the child. Whenever possible, parents should be encouraged to stay with the child while he or she is in the hospital. The length of hospital stay is determined by the requirements for safety, recognizing that it is best for the child to return to the home setting as soon as possible.

Modifications in surgical technique may be required in the management of pediatric problems. Dressing changes, suture removal, and postoperative manipulations should be kept to a minimum so as not to provoke fear and discomfort in children who must undergo minor procedures. Children may require sedation or a general anesthetic for packing or suture removal, which is rarely required for adults. In addition, it is prudent to plan ahead of time to use absorbable suture material and dressings that need only infrequent changing.

Genetics and Pediatric Otolaryngology

It is becoming imperative that pediatric otolaryngologists be familiar with genomic medicine to appropriately counsel and refer their patients. Most otolaryngologists understand the basics of Mendelian inheritance and the genetic basis of conditions such as sensorineural hearing loss (SNHL). SNHL displays remarkable genetic heterogeneity, with at least 45 genes and over 80 genetic loci currently identified. The *GJB2* gene encodes the connexin 26 protein, and *GJB2* mutations are responsible for about one third of sporadic prelingual SNHL and more than 50% of cases with an autosomal recessive

inheritance pattern.[21] Thus the otolaryngologist should also have some knowledge of genetic testing to answer questions such as "Why is my child deaf?" "What is my child's prognosis?" "How likely is hearing loss in my future children?" Imaging that demonstrates cochlear dysplasia, lateral semicircular canal dysplasia, and an enlarged vestibular aqueduct may partly answer the "why," but genetic testing offers the possibility of more accurate recurrence risk information compared with estimates based on empirical data.

Although referral for genetic counseling would be preferred, it is not always available.[21] Ideally, children with SNHL would receive treatment at an institution with a pediatric genetics service. Appropriate genetic counseling should be provided before and after genetic testing. It is important to clearly explain the consequences of positive and negative results, as misunderstanding among parents, caregivers, and even some providers is not uncommon. Genetic results have the potential for long-term consequences to the individual, the family, and the clinician involved, which mandates proper training on what is still an evolving field in terms of ethics. Presymptomatic testing of minors and the reporting of incidental findings continue to pose particular challenges.

Genetic factors in pediatric otolaryngology are also important in a variety of head and neck lesions. Thyroid cancer is the most common type of neck tumor in childhood, and the majority are well-differentiated cancer. Five percent of pediatric thyroid cancer cases are medullary thyroid carcinoma (MTC), which is mainly familial. The diagnosis is made on clinical suspicion and studies based on fine-needle aspiration; there is a 25% association with multiple endocrine neoplasia type 2. It is important to rule out pheochromocytoma, and prophylactic total thyroidectomy is recommended for genetic carriers. Genetic testing for mutations in the *RET* proto-oncogene should be considered.[22] A 1995 court judgment, *Pate v. Threikel*,[23] found that offering genetic testing and genetic counseling is considered the standard of care for highly treatable/preventable disease in genetic carriers. Radioactive iodine is not effective for the treatment of MTC. Therefore the best hope for cure is complete surgical removal of the thyroid gland, which for the aggressive form should occur before the age of 5 years, prior to the development of a tumor.

These examples of genetic conditions highlight the ethical implications of testing in children who are unable to consent for genetic tests; the importance of cascade testing of family members in certain conditions; and the need to provide information to help in reproductive decisions in the situation of carrier status, in vitro fertilization, and/or prenatal testing.

Illustrative Case 1

A baby girl with an antenatal diagnosis of craniosynostosis suggestive of Crouzon syndrome was born at term via normal vaginal delivery to nonconsanguineous parents. She developed nasal obstruction requiring oxygen therapy and was transferred to a tertiary pediatric center. At 2 weeks of age, a polysomnogram (PSG) showed severe obstructive sleep apnea (OSA) that improved with CPAP therapy. Nasal endoscopy and imaging confirmed bilateral choanal stenosis/atresia and hydrocephalus. Further evaluation of her airway showed multilevel obstruction with glossoptosis, palatal apposition to the posterior wall, impairment of bilateral vocal cord motion, and tracheomalacia.

Because of her multilevel obstruction, she was managed initially with a nasopharyngeal airway and CPAP therapy with high pressures. Obstructive episodes were challenging to manage, especially during periods of respiratory viral illnesses. She had also undergone multiple cranial surgeries and multiple ventriculoperitoneal shunt revisions. The CPAP support interfered with straps needed for headgear, and the pressure of CPAP combined with proptosis produced chemosis. Failure to thrive and reflux were later definitively managed by percutaneous gastrostomy feedings and fundoplication. After careful consideration of options, the family made an informed decision to proceed with a tracheostomy at 18 months of age, with significant improvement in her overall health and resolution of her failure to thrive.

Learning points: Congenital conditions often present with a host of clinical problems for the pediatric otolaryngologist to evaluate and manage, including airway distress, sleep apnea, maintenance of normal hearing, avoiding oral aversion, and identifying delays in the development of language and speech. The medical and psychosocial concerns of the family and child are greatly assuaged by the participation of a surgeon who is familiar with the underlying genetic syndrome. This patient's management required the coordination and cooperation of multiple medical (general pediatrics, pulmonary/sleep, genetics, ophthalmology), surgical (otolaryngology neurosurgery, craniofacial), and allied health (dietitians, speech pathology, physiotherapist, and occupational) teams.

Illustrative Case 2

Presenting symptom: A pregnant mother at 30 weeks of gestation presented with polyhydramnios on ultrasound. Subsequent investigation including magnetic resonance imaging (MRI) confirmed a jaw index less than 23 and suspicion of severe micrognathia in the fetus. Otolaryngology is consulted for a possible ex utero intrapartum treatment (EXIT) procedure.

Socioeconomically, although both parents work, the family lives at the poverty line, with another child at home. The parents are young with strong religious beliefs. They are now expecting a second child who, on the basis of a prenatal MRI, may have a condition that is almost impossible to survive.

Management: A multidisciplinary meeting is called by the maternal/fetal medicine team, along with neonatology, pediatric and adult anesthesiology, and pediatric otolaryngology. Parents are counseled preoperatively about the potential clinical outcomes, possible risks and complications, and informed of the clinical care pathways and procedures necessary to achieve a satisfactory outcome. Special mention is made of the possibility of a tracheostomy being required.

Resulting scenario: The baby is born by cesarean section at 37 weeks of gestation. While the child was still on placental support (Fig. 1.4), direct laryngoscopy was attempted via several techniques but without success. As the larynx could not be identified, a vertical tracheostomy was performed while the child lay on the mother's abdomen and was then successfully delivered with a tracheostomy and transported to neonatal intensive care. Evaluation by the clinical genetics service confirmed a diagnosis of cerebrocostomandibular syndrome. Pediatric pulmonology introduced NIV. After multidisciplinary consultation, mandibular distraction osteogenesis was performed. The tracheostomy was removed at 12 months of age, and the child is now developing well in all aspects. Ongoing management for otitis media with effusion was required. Subsequent microlaryngoscopy has confirmed that the child still has a difficult laryngoscopy for routine intubation, and it is necessary to inform subsequent anesthetists that a flexible bronchoscopy may be necessary to manage this child's airway safely.

Learning points: Preoperative and postoperative planning among multiple disciplines was necessary for a successful outcome. Having a radiologist familiar with three-dimensional ultrasound and prenatal MRI was imperative, and the pediatric

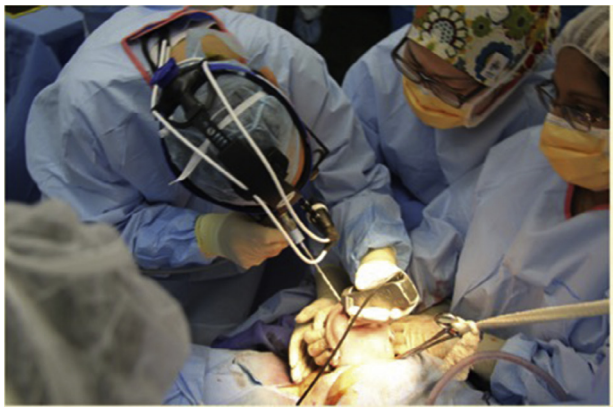

Fig. 1.4. During an ex utero intrapartum treatment procedure, the baby is partially delivered to establish airway control. Standard oral intubation proved unsuccessful, and it was necessary to proceed to a tracheotomy while the patient was still on placental support.

otolaryngologist worked closely with the pediatric anesthetist and neonatologists to deliver the baby successfully. It is important to note that all EXIT procedures involve two patients, and partnering with the adult obstetrics and anesthesia team is essential. The hospital must have appropriate resources for both adult and pediatric advanced care. The expertise of the pediatric otolaryngologist in performing tracheotomy in the smallest of infants is essential.

Illustrative Case 3

A child was diagnosed at birth with bilateral choanal atresia and bilateral retinal coloboma. Her echocardiogram showed a ventricular septal defect (VSD) and an atrial septal defect (ASD), both small. She had a large sliding hiatal hernia, and transpyloric feeding was required. After a failed choanal atresia repair with stent placement in a rural general otolaryngology setting, the child experienced ongoing respiratory distress and was facing possible tracheostomy. Head imaging confirmed hypoplastic vestibules, bilateral absent semicircular canals, small internal auditory canals, abnormal ossicles, and opacified sinuses. She suffered from recurrent respiratory infections and chronic lung disease requiring blow-by oxygen. When she was months old, ligation of the PDA was performed. She had failure to thrive, growth retardation, and poor development in swallowing on repeated swallow studies and required a high caloric intake via a transpyloric tube. Her sliding hiatal hernia was treated by fundoplication and insertion of a gastrostomy tube at 3 weeks of age. She was severely visually impaired. Electrocochleography as a neonate had confirmed near-normal hearing in the right ear but profound deafness in the left. Genetic assessment confirmed CHARGE syndrome (coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, and ear abnormalities).

After hospital transfer from a rural setting to a pediatric institution, the child's choanal atresia was successfully managed with revision surgery. Over subsequent years, she was maintained on NIV while sleeping, repeated ventilation, tube insertion for persistent middle ear effusions and mastoiditis, and tube fed until she was 3½ years old. Her reactive airway disease was managed by her pediatrician with inhaled and systemic steroids, her sleep physician regularly monitored her NIV, and her otolaryngologist treated her middle ear disease. Audiology managed her bilateral behind-the-ear hearing amplification devices to rehabilitate the moderately severe hearing loss of the right ear and profound hearing loss of the left ear (Fig. 1.5).

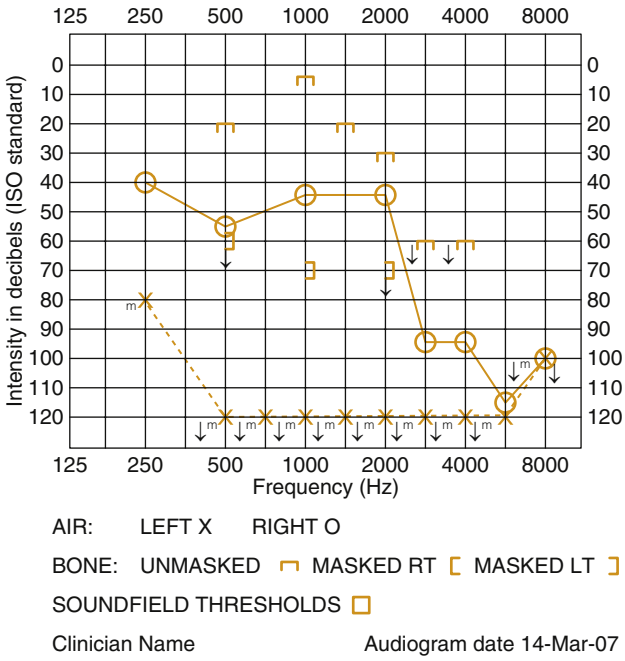

Fig. 1.5. Preoperative audiogram of patient described in Illustrative Case 2.

The patient received itinerant and visual support from her teachers, eventually becoming a teacher's aide and achieving a black belt in martial arts. Her visual acuity was 3/60 on the left and 6/20 on right, and she was able to learn to read with the help of glasses. Nystagmus and a left-sided coloboma persisted. After undergoing speech and articulation therapy, she clearly expressed her needs. She wanted an otoplasty to correct her bilateral lop-ear deformity. She also developed eardrum perforations, which frequently discharged after her ventilation tubes extruded. She then developed a cholesteatoma in the only-hearing right ear with an atelectatic eardrum adherent to the promontory. The patient underwent surgical removal of a cholesteatoma with placement of another ventilation tube. At 10 years of age, she had a tonsillectomy to correct her ongoing sleep apnea. This was successful, with a subsequent apnea-hypopnea index (AHI) of 4.0 per hour, normal CO_2 levels, no oxygen desaturations below 93%, and no further need for NIV. She managed to reach the 70th centile for weight and 40th centile for length.

The patient's speech therapist concentrated on her speech intelligibility and noted that she had severe hyponasal resonance. With a series of simple exercises, improved speech intelligibility was achieved. At the age of 16 years, she underwent an external approach septorhinoplasty and endoscopic sinus surgery to successfully correct her severe septal deformity, chronic sinusitis, and nasal obstruction. A cartilage graft tympanoplasty was performed to repair the atelectatic eardrum. A second cholesteatoma between the incus and stapes was discovered and excised at this time. She has a residual mild hearing loss in the right ear that remains stable and dry (Fig. 1.6). At the age of 20, she is working in the retail food industry, enjoying life, and planning her next holiday at the beach.

Learning points: This case illustrates how children with congenital syndromes such as CHARGE, who have needs that change throughout development, require long-term care. It highlights the multidisciplinary nature of the condition and the increasing survivorship of these children. The otolaryngologist should take an active role in identifying speech and language delays and in making appropriate referrals. In subsequent years, the expectations of the child and guardians

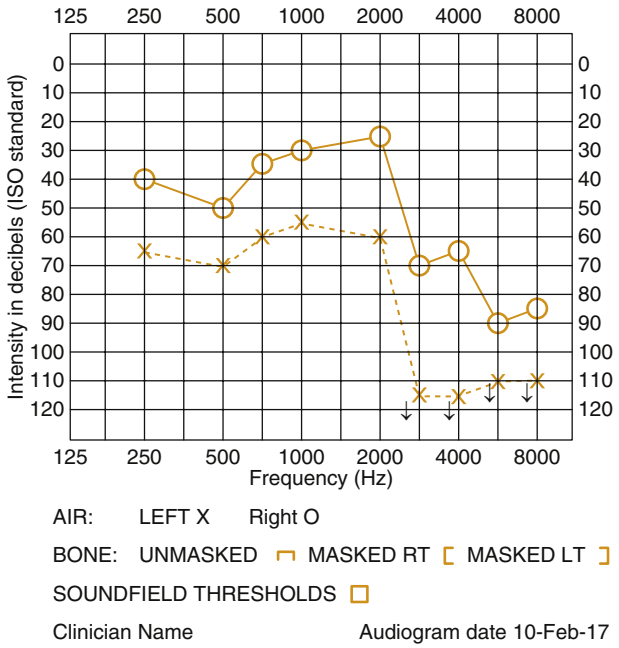

Fig. 1.6. Postoperative audiogram of patient described in Illustrative Case 3.

evolve, requiring ongoing attention to hearing and breathing, autonomy, self-image, and societal acceptance. Parents or caregivers need to know why things are being done, what they can expect, and—most importantly—what is expected of them. With ongoing care by their supportive caregivers and family members, these children can enjoy a regular life and become significant contributors to their communities.

Biosocial Issues in Pediatric Otolaryngology

The multifactorial model characterizes illnesses as a combination of four major factors: disease diagnosis, patient characteristics, psychological stressors, and developmental issues. Experts have advocated for managing disease with the multifactorial model[23] as opposed to the previous dichotomous model of organic versus functional, particularly for the pediatric population. A child's development may be negatively influenced by illness and how it was managed, whether in an acute or chronic setting, a hospital institution or home environment, or with caregivers or guardians with different cultural backgrounds.[24]

As discussed by Erikson and Piaget in Chan et al.,[25] adults continue to develop psychosocially until their behaviors, values, and social circles are mature and stable. Children and adolescents, in comparison, continue to develop rapidly around their world environment, using its cues to help them develop. It is important that health professionals interact and communicate with the children and their caregivers/guardians carefully and to be aware of the broader context of a child's or adolescent's life, as this affects the way they experience and recover from illness or injury.

Infants (ages 1 to 18 months) are dependent on their caregivers, who in turn aim to gain their children's trust that no harm will come to them, establishing a sense of stability and security in their lives. An illness at this stage can jeopardize this trust such that children may appear to regard the world outside of their home as an unsafe and unpleasant environment. The toddler (ages 1 to 3 years) engages in developing communication and motor skills to allow interaction with the environment. An illness requiring surgery, which may be simple or complex, puts a great strain on the child's relationship with his or her environment. A child may demonstrate difficulty with separation, emerging with a poor self-image and fearful of interactions with peers and adults.

Similarly, preschool children (ages 3 to 6 years) enter a preoperational period, recognizing that events and things have causes. Illness at this stage limits the development of social and motor competence, whereas physical restrictions can produce a child who becomes passive and fearful of achieving goal-directed behaviors. Children of school age (ages 6 to 12 years) aim to develop mastery and industry, but illness will produce issues such as feelings of inadequacy, a sense of being different from others, and a poor self-concept. These issues may result in withdrawal from school and delay in the schooling process. Adolescents (13 to 18 years) hope to achieve a personal identity, true independence, and a mature mind. Adolescents understand the physiologic basis of disease, and they have a strong need to be accepted by family, peers, and society at large. Adolescents may regard a minor anatomic deformity as devastating, and the need to comply with medical treatment may be met with a pathologic desire to be independent of families and health professionals.[25]

An illness requiring surgery puts a great strain on the relationship of the child with his or her environment, whether simple or complex. As pediatric physicians/surgeons, we must work to promote the child's continual growth and development in a harmonious way lest the child subsequently avoid continued care needed for a return to health.

Autism Spectrum Disorders

As these children display delayed speech and language development along with social isolation and unusual behavior, autism has become a common condition faced by pediatric otolaryngologists. First described by Leo Kanner at the Johns Hopkins hospital in 1943, pervasive developmental disorders occur along a spectrum that includes Asperger syndrome, attention-deficit disorders, attention-deficit/hyperactivity disorder, learning disabilities, and autism. Autism affects 6 of every 1000 children and affects boys more commonly than girls by a ratio of 5:1.[26,27] The conditions may present with some or all of the following features: (1) limited social interaction, (2) reduced imagination and unusual interests and activities, and (3) alteration of verbal and nonverbal communication.

Hearing impairment, speech delay, and suspected sleep apnea often manifest in this patient group, and involvement of the otolaryngology service is frequently requested. Autistic children also exhibit unusual responses to differing sensory stimuli, including a heightened or depressed response to hearing, sight, smell, taste, and touch, with reactions such as emotional instability. The otolaryngologist may often be the first physician to suggest the diagnosis of autism spectrum disorder, particularly once conductive hearing loss and SNHL have been ruled out. The expertise of a pediatric audiologist may be required to assess hearing in these children. Snoring and chronic mouth breathing may be attributed to the presumed behavioral disorder without complete assessment of airway or sleep because of the difficulty of interacting with these children.

Children with autism may require extra time and patience to complete the clinic visit. Examination is facilitated by using play techniques to establish rapport, performing the exam on the floor of the office in the sector of toys, where the child feels most at ease and is most likely to allow a successful physical examination. It is important to prepare such a child for surgery appropriately, with the assistance of an anesthetist, to help the child get prepared in clothes of his or her own choosing, and to have a trusted caregiver with him or her at all times.

Children with behavioral disorders will benefit from improved quality of life in terms of maintaining adequate hearing, good-quality sleep, and less frequent illnesses. The family unit often requires additional care, support, and predetermined management strategies during doctor and hospital visits and to prepare for surgical procedures, which then translates into better social integration over the long term.

Ethical Dilemmas in Pediatric Otolaryngology

The current era is notable for pluralistic values, such that individuals hold a wide range of beliefs and values that are important to them. Widespread immigration and the effect of global communication via the Internet, social media, and television allow these diverse values to be distributed widely. The Western tradition of philosophical thought, which originated with Plato and Aristotle, has traditionally guided moral decision making in providing the principles of autonomy, nonmaleficence, beneficence, and justice. It offers moral insight into the patient's illness and the realities of his or her situation.[28]

A collaborative family meeting with open disclosure and discussion prior to decision making will enable a noncoercive consensus-seeking dialogue. This dialogue/consensus approach, also referred to as the shared decision-making model, currently drives the case conferences among clinicians, the family, and other stakeholders in determining what will best contribute to the welfare of the ill child. The purpose of the family case conference is to explain the facts of the medical condition, convey any uncertainty about the exact diagnosis and prognosis, and allow the various participants to speak truthfully and noncoercively in a way that does not aim to direct or influence other participants in the discussion. The ultimate aim is to achieve consensus, ensuring all involved understand the reality of the situation along with the values held by the patient's caregivers and the clinicians. Habermas refers to the truth in the dialogue, the data collection, and explanation,[29] which is aimed at maximizing the "good." Emphasizing a forthright and honest approach to the truth of the situation helps to achieve a level of consensual dialogue that is meaningful.[28]

The cases described earlier illustrate some of the competing values faced by families and medical professionals alike. The joy of the expectant parents is acknowledged along with the personhood and dignity of the child. At the same time, the possibility of no intervention and its consequences may also be discussed. Children with congenital anomalies may require long-term care and multistage surgeries to improve their well-being in terms of respiration, feeding, and swallowing. Normal speech and swallowing cannot be assumed, and the family must be prepared for the possibility of other comorbidities not yet diagnosed, such as hearing loss and central neurologic dysfunction. A child who needs specialized care may be ineligible for day care or an untrained babysitter, which will affect the parents' ability to work, with potential financial implications. The family's dynamics are affected, even to the point of divorce from the stress involved in caring for a disabled child. The pediatric otolaryngologist must have the medical training to care for critically ill children and to use new technology to perform complex procedures while also exhibiting professionalism and expertise in patient- and family-centered care. On the basis of the consensus gained via dialogue, parents are supported in their decision making, which is all the more important when outcomes may be painful.

CONCLUSION

The care of children can be an extremely rewarding experience. Children may become ill quickly but often recover just as quickly if set on the right path. Watching children grow and develop makes for lasting memories and personal satisfaction. For many clinicians, helping children with chronic illness to overcome disabilities and difficulties and watching them reach their full potential as they grow to adulthood is highly satisfying. Pediatric otolaryngology continues to evolve. Along with this process, it is important to foster communication among trained professionals who may share their experiences at appropriate institutions and in small group meetings. Moreover, it is important to support the general otolaryngologist who rightfully continues to manage children, sharing his or her knowledge and expertise as previously rare and complex conditions become more prevalent. In an ever-changing medical landscape, the need for innovation is pressing.

For a complete list of references, visit ExpertConsult.com.

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