Section 1 Basic Principles, Assessment, and Planning of Airway Management

Chapter 1

Developmental Anatomy of the Airway

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Expertise in airway management in infants and young children requires a comprehensive knowledge and understanding of the developmental anatomy of the human upper airway from birth through adolescence. This chapter will review these topics as well as the anatomical and developmental causes for common syndromes that are associated with difficult mask ventilation or difficult tracheal intubation.

Embryonic Development[1–5]

The upper airway consists of the air-conducting passages from the nose to the carina.[3] The structures of the upper airway continue to change their shape and properties until late into the first decade of life. Less is known about their developmental course during fetal life as compared with other organ systems because much of this understanding is gained from postmortem studies.

Upper airway structures develop with the cranium and the most cephalad boundaries of the digestive and respiratory systems. The lateral surface of a 5-week-old 4.0 mm embryo contains five or six pairs of narrow masses called *branchial (pharyngeal) arches*. Each branchial arch contains characteristic types of ectoderm and mesoderm, the primordial precursors of epithelial (e.g., skin) and mesothelial structures (e.g., muscle, bone), respectively. The structures of each branchial arch receive motor or sensory innervation from an adjacent cranial nerve. When the primordial muscle cell migrates, it retains its original embryonic innervation. The structures between the branchial arches are the *branchial (pharyngeal) clefts*, which, with the exception of the first cleft, disappear during the course of development. The tissue underlying the branchial clefts contains outpouchings of the foregut region called pharyngeal pouches. The pharyngeal pouches will develop into the corresponding endothelial structures of the upper digestive and respiratory organ systems.

The first branchial arch develops into the mandible, maxilla, and the muscles of mastication. It contributes to development of the bones of the middle ear and the muscles between the ear and mandible, such as the tensor tympani, tensor veli palatini, and the anterior belly of the digastric muscle. Motor and sensory innervation to the structures derived from the first arch are supplied by the trigeminal nerve (cranial nerve V).

The second branchial arch forms bony and muscular structures from the ear (proximally) to the hyoid bone (distally), including the muscles of the face and inner ear that are innervated by the facial nerve (cranial nerve VII). Skeletal contribution from the second branchial arch includes the styloid process and the lesser cornu of the hyoid bone.

The third branchial arch develops into the body and greater cornu of the hyoid bone and the stylopharyngeus muscle, which aids in elevating the pharynx during swallowing, and is innervated by the glossopharyngeal nerve (cranial nerve IX).

The fourth through sixth branchial arches contribute to the formation of the thyroid, cricoid, arytenoid, corniculate, and cuneiform laryngeal cartilages, as well as the muscles that form the pharynx, larynx, and upper half of the esophagus. These structures are innervated by the vagus (cranial nerve X) and accessory (cranial nerve XI) nerves. The earliest appearance of the future larynx is seen as a bud growing out from the ventral part of the foregut at approximately 4 weeks' gestation. The laryngeal and esophageal tracts are initially seen as one common tube, which eventually separates into two adjacent and functionally different conduits. By 16 weeks' gestation, the larynx contains all its definitive elements in their proper proportions. During fetal and postnatal growth, the development of the size of the larynx closely parallels the size of the surrounding bony and cartilaginous structures.[6]

The first branchial cleft becomes part of the external auditory canal, while the remaining clefts do not correspond to recognizable human structures. Nevertheless, abnormal formation here can lead to cysts or more significant malformations.

The first pharyngeal pouch becomes incorporated into the future temporal bone and forms the epithelial lining of the middle ear and the tympanic membrane. The second pharyngeal pouch develops into the tonsil. The superior portion of the third pharyngeal pouch differentiates into the inferior parathyroid, and the inferior portion migrates caudally to become the thymus. The fourth pharyngeal pouch forms the superior parathyroid gland; the area roughly corresponding to the fifth and sixth pharyngeal pouches is incorporated into the thyroid gland.

Postnatal Development

Airway management of infants and young children will be influenced by developmental differences in head and neck anatomy. These differences are influenced by two major growth spurts during childhood that contribute to the vertical growth of the facial structures: the first at the time of the acquisition of permanent dentition (i.e., 7–10 years of age), and the second during puberty in the teenage years.

When compared to the older child, the infant's skull (especially the occipital region) is relatively larger, such that neck flexion may not be required to attain the classic sniffing position that optimizes visualization of the glottic structures during laryngoscopy. At birth, the neurocranium-to-face size ratio is 8:1, and declines to 6:1 at 2 years of age, 4:1 at 5 years of age, and approximately 2:1 by adulthood.[7,8] The growth of the lower facial bones is proportionately linear from 1 to 11 years of age.[6]

The mandibular arch of the infant is U-shaped and becomes more V-shaped during childhood until it is completely developed during adolescence. The angle between the ramus and the body of the mandible is more obtuse in infants than in adults. This largely accounts for the relatively low incidence of difficult intubations in children compared with adults.

Overall, the most important difference in nasal anatomy between young children and adults is merely the smaller size. Small nasal passages are more likely to become obstructed with blood or secretions as a result of instrumentations during airway management. Children are less likely to have occult nasal polyps or septal deviations when compared with adults.[9] The anatomical dimensions of the nasopharynx increase linearly between 1 and 11 years of age.[6]

Early literature indicated that small infants were obligate nasal breathers, which predisposed them to breathing difficulties during periods of nasal obstruction. However, this has largely been disproven,[10] although infants with choanal atresia will often develop upper airway obstruction, which results in varying degrees of hypoxemia.[11]

The infant tongue is relatively larger in proportion to the oral cavity when compared with the adult. Tongue volume increases linearly between ages 1 and 11 years.[6] Magnetic resonance imaging (MRI) studies of the upper airway during general anesthesia have demonstrated that, as in adults,[12] upper airway obstruction occurs primarily at the levels of the soft palate and epiglottis, and not at the base of the tongue.[13]

The 20 primary teeth, identified by a lettering system, begin to erupt during the first year of life, and are shed between 6 and 12 years of age. The 32 permanent teeth begin to appear at the same time as the primary teeth are shed and are identified by a numbering system.

In newborns, the uvula and epiglottis are in close proximity within the oropharynx, which facilitates nasal breathing and oral ingestion of liquids simultaneously. This anatomical relationship is maintained throughout most of the first year of life, but during the second year, the larynx begins to descend as it adapts to its greater role in phonation.

Although the mechanisms have not been elucidated, the pharynx of premature newborns is susceptible to passive collapse, especially during apnea, but may also collapse as a result of cervical flexion or nasal obstruction.[14] These effects are exacerbated by the administration of general anesthesia or sedatives, which decrease pharyngeal muscle tone. Furthermore, pharyngeal collapse often occurs in premature infants during application of cricoid pressure.

Of interest, the upper airway of a normal infant is smaller in both inspiration and expiration at 6 weeks of age compared with the neonatal period. This relative narrowing may be caused by postnatal growth of adenoid tissue or thickening of the mucous membrane lining in response to infection or second-hand smoke exposure. The linear dimensions of the soft palate and oropharynx increase linearly between 1 and 11 years of age.[6,15]

Adenoidal and tonsillar tissue is minimal at birth, and then grows rapidly between 4 and 7 years of age. The growth of airway lymphoid tissue parallels the growth of the facial and cervical bony structures.[6] Hypertrophied tonsil and adenoid tissue is likely the most common cause of upper airway obstruction after administration of general anesthesia in children in this age group.

The epiglottis of infants is relatively narrow and short, and angled into the lumen of the airway. The lower portion of the oropharynx at the level of the epiglottis is particularly compliant and prone to collapse during anesthetic or sedative-induced loss of consciousness. Obstruction at the level of the epiglottis can be significantly decreased by placing the child in the lateral position.[16]

The effect of sex on oropharyngeal length has been studied, with particular reference to an association between relatively longer airway length and the predisposition to obstructive sleep apnea.[17] Prior to the onset of puberty, boys and girls have relatively similar oropharyngeal length, but after the onset of puberty, the oropharyngeal lengths in boys are greater than those of girls, even after correcting for height and weight. The relatively longer upper airway length in males has been implicated as a possible etiologic factor in the greater disposition in males toward obstructive sleep apnea.[18] Thus, postpubertal males may have a greater disposition toward upper airway collapse in response to administration of pharmacological agents that depress consciousness.

During infancy, the relative position of the larynx is slightly higher in the neck than in older children and adults. Although its position relative to the cervical spine is complete by 3 years of age (it descends from C2–C3 to C4–C5), it continues to descend relative to other facial structures such as the mandible.[19] The tip of the epiglottis proceeds in a gradual and linear descent from C2 to C3 from birth to 18 years of age.[20] This relative movement is unique to humans because of the shifting functionality from sucking and swallowing while breathing to the development of speech later in life. Early in life, a relatively high larynx facilitates simultaneous sucking and respiration due to the apposition of the epiglottis (as high as C1) and the soft palate. Additional differences in early life that protect against aspiration during feeding include relatively thicker aryepiglottic folds and larger arytenoids.

The chest wall of neonates and small infants is highly compliant and tends to collapse inward, thus reducing functional residual capacity (FRC) and promoting atelectasis. To preserve FRC, the adductor muscles of the larynx act as an expiratory "valve," and restrict exhalation in order to maintain positive end-expiratory pressure (PEEP). This is referred to as "laryngeal braking."[21,22]

The higher position of the larynx during infancy influences airway management to the extent that the glottic opening is more easily visualized using a straight, rather than a curved, laryngoscope. In infants of less than 1 year of age, elevation of the base of the skull during direct laryngoscopy is usually not necessary.[23]

In children who have received neuromuscular blockade, the fixed-diameter cricoid cartilage is the narrowest structure of the upper airway because of its inability to distend in a similar manner to the vocal cords.[24–26] A tracheal tube that easily passes through the relatively compliant vocal cords may compress surface mucosa at the subglottic level or the cricoid cartilage and predispose to inflammation, edema, and subsequent scarring and stenosis.[27–29] Tracheal edema is more likely to increase airway resistance in smaller diameter airways since the resistance to flow through a tube is related to the fifth power of the radius of the tube (since this flow is largely turbulent). In non-intubated, sedated children without neuromuscular blockade, the adductor muscles of the vocal cords are tonically active, and are the basis for the narrowest portion of the upper airway to occur at this level.[30–32] The relationship between the sizes of the structures along the upper airway remains relatively stable throughout growth and development.[32] There is no specific age during childhood at which the use of an uncuffed tracheal tube would be beneficial.

Tracheal lengths (distance between glottis and carina) increase linearly during childhood.[25,33] A familiarity with these distances in infants will facilitate proper placement of the tracheal tube midway between the glottis and carina to minimize the risk of displacement (either too high out of the larynx or too low into the main bronchus) with changes in head or neck position.

Anatomical Basis for Syndromes Associated with Difficult Airways

Up to 3% of children have congenital or acquired upper airway abnormalities, frequently associated with a craniofacial anomaly, which may result in

difficulty in mask ventilation or tracheal intubation.[1] Successful airway management requires an understanding of the anatomical basis of these abnormalities and the relationship to normal development. To organize these craniofacial anomalies, the Committee on Nomenclature and Classification of Craniofacial Anomalies of the American Cleft Palate Association has organized them into five distinct categories: *hypoplasia*, *clefts*, *synostosis*, *hyperplasia*, and *unclassified*.[34,35] The first three of these are often implicated in difficult airways.

Anomalies within the *hypoplasia* category are characterized by hypoplasia or atrophy of a portion of the craniofacial skeleton. Micrognathia, or mandibular hypoplasia, is a common cause of difficulty with either ventilation or intubation in the neonatal period and beyond.[36] The finding of micrognathia along with glossoptosis and resultant airway obstruction defines Pierre Robin sequence, which occurs in up to 1:8500 live births. PRS can occur in isolation, associated with additional congenital malformations, or as part of a clinical syndrome with a specific chromosomal anomaly (in approximately 60% of cases).[37] The wide range of clinical conditions demonstrating PRS suggests a diverse developmental pathogenesis of the sequence, with a few leading hypotheses elucidated.[38]

Intrauterine constraint, leading to compression of the chin and limitation of jaw growth prenatally has been implicated in both asymmetric and symmetric micrognathia. Support for this theory includes findings of pressure indentations on the chest at birth, associated with muscular torticollis on the same side as unilateral micrognathia, and an increased incidence of PRS in twin gestations.[39] The hypothesis that PRS develops as a primary failure of mandibular outgrowth traces development of the mandible back to its origin in the first branchial arch. Unlike the maxilla, which forms as bone in close association with the developing facial bones, the mandible forms in relative isolation, initially as cartilage, called Meckel's cartilage. Micrognathia, in this theory, is due to defective generation and/or growth of this cartilage.[38] In either case, mandibular hypoplasia prevents fusion of the palatal shelf, which normally occurs between the eighth and tenth weeks of gestation.[37] This causes the retrognathia, glossoptosis, and airway obstruction that characterizes PRS, and explains the frequent occurrence of cleft palate with PRS.[38]

Aberrant development of the first and second branchial arches is implicated in the etiology of *oculoauriculovertebral spectrum* (OAVS; hemifacial microsomia, Goldenhar syndrome), which involves unilateral hypoplasia of the craniofacial skeleton. As the names imply, there is a wide clinical spectrum associated with this disorder. There is also no consensus on minimum diagnostic criteria; however, the majority of patients have asymmetric microsomia and external ear abnormalities.[40] The reported prevalence varies from as common as 1 in 5600 to 1 in 45 000.[41] While most cases of OAVS are sporadic, there is evidence that genetic, epigenetic, and environmental factors all contribute to the complex etiology of this disorder, likely also involving the neural crest cells in the first and second branchial arches.[40]

Clefting most frequently involves the lip and/or palate, typically not associated with a difficult airway, but also may involve the midface, as in Treacher Collins syndrome (Figure 1.1). This is an autosomal dominant inherited disorder associated with severe airway obstruction, and inability to provide mask ventilation or tracheal intubation. It is associated with a genetic mutation on chromosome 5 and occurs in

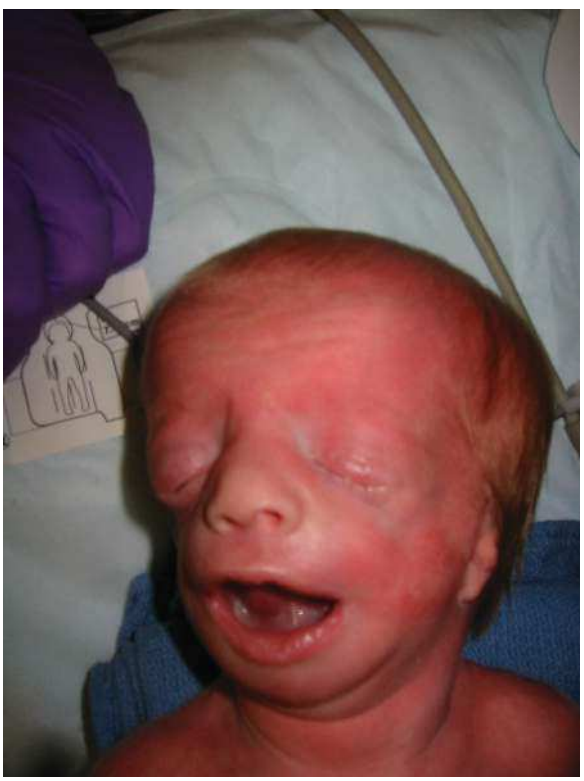

Figure 1.1 Treacher Collins syndrome. (From *Basics of Pediatric Anesthesia*, Litman, RS, ed., 2016, with permission.)

approximately 1:50 000 live births.[42] Specific findings include symmetric maxillary and zygomatic hypoplasia, a high arched and/or cleft palate, downward sloping palpebral fissures, and microtia often associated with hearing loss.[38,42] Most of these findings occur in tissues that arise from the first and second branchial arches and are due to mutations (in the *treacle ribosome biogenesis factor 1 [TCOF1]* gene) associated with a decreased number of cranial neural crest cells in these regions.[42]

Craniosynostosis refers to the abnormal closure of one or more of the cranial sutures, and is commonly associated with midface hypoplasia in syndromes that also have difficult airways. Airway issues are usually related to mask ventilation, with poor mask fit, combined with choanal stenosis and excessive nasopharyngeal soft tissue leading to multilevel airway obstruction. Intubation may also prove difficult and may be worsened by fibrosis or mechanical limitations following midface advancement surgery.[37,43]

Apert syndrome is inherited in an autosomal dominant fashion and is associated with mutations in the *fibroblast growth factor receptor 2* (*FGFR-2*) gene on chromosome 10, causing abnormalities in suture closure and bone formation of structures originating from the first branchial arch.[44] Mutations in *FGFR* genes also cause additional craniosynostosis syndromes associated with midface hypoplasia and potential difficult airways, such as Pfeiffer syndrome (Figure 1.2) and Crouzon syndrome.[45] Evaluation of the hands of these patients will help distinguish between these otherwise similarly appearing syndromes. Patients with Apert syndrome have syndactyly, while Pfeiffer syndrome is associated with a broad thumb, and patients with Crouzon syndrome have completely normal hands.[35]

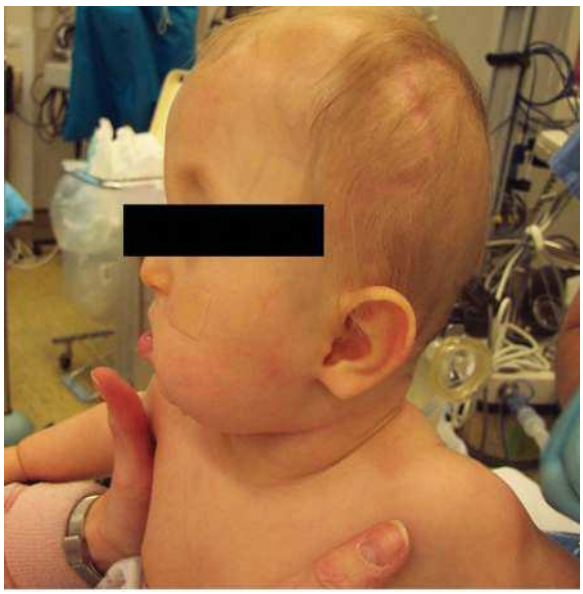

Figure 1.2 Pfeiffer syndrome. (From *Basics of Pediatric Anesthesia*, Litman, RS, ed., 2016, with permission.)

Understanding the changing airway anatomy as children grow, as well as the anatomical causes of difficult airway syndromes is the first step in developing safe and effective airway management plans in the difficult pediatric airway.

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