Chapter 99: Congenital Disorders of the Larynx

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The study of congenital abnormalities of the upper airway in premature babies, neonates, and infants may require consultation among pediatric specialists in the fields of medicine, surgery, neonatology, pulmonology, anesthesiology, and radiology. The pediatric laryngologist is the focal point for definitive endoscopic diagnosis and for determining management of airway obstruction. Although the majority of patients undergoing diagnostic examination are seldom seriously ill, many demanding problems confront the endoscopist and anesthesiologist, sometimes unexpectedly. Mutual understanding and teamwork are essential for the safe sharing of access to the airway. Moreover, when airway obstruction exists, anesthesia and endoscopic instrumentation may precipitate serious or total obstruction, so facilities to overcome this emergency must always be readily at hand.

Anatomy

Adult and child differences

Embryology is covered in Chapter 95. Both structural and functional differences exist between the pediatric and the adult larynx.

The size of the internal larynx at birth is approximtaly one third of its adult size, and the infant larynx is smaller in proportion to the rest of the body than it is in the ault. The vocal cors are 4.0 to 4.4 mm long at birth. The laryngeal saccule is variable in size and may be large enough to be seen on a lateral raiograph (Fig. 99-1). The supraglottic airway makes an angle with the subglottic airway at the vocal cords.

The larynx grows until puberty, at which time it is fully developed. Whereas the thyroid cartilage in an infant is short and broad, the adult larynx, especially in the male, develops the laryngeal prominence and thyroid notch.

The thyroid cartilage is closer to the hyoid in the infant. The fetal larynx is positioned high in the neck, usually at the level of the second and third vertebrae, descending to the fourth vertebra at birth, to the fifth at approximately 6 years of age, and to the sixth or seventh at puberty.

The higher larynx of the infant is softer, more easily displaced, and more easily irritated. The epiglottis projects into the oropharynnx, and its shape has a considerable range of variation. The so-calle infantile epiglottis is longer, narrower, and more tubular. This Omega-shaped structure is seen more often in laryngomalacia but is by no means a constant finding nor necessarily a diagnostic feature in this condition. The aryepiglottic folds are disproportionately large, and the arytenoids themselves vary in shape, being more prominent in the infant than in the adult larynx. The epiglottis is softer and less rigid than in the adult, as indeed is the entire cartilaginous framework of the larynx, which has less resistance to changing pressures. Although this mobility of the musculature and soft tissues of the pharynx and larynx in the newborn is greater, the capacity is also greater for stabilization by the sucking pa in the cheeks and the skeletal muscular action in fixing the mandible and hyoid

bones. The normal pharyngeal airway is maintained from the moment of birth despite variations in head and neck position, opening of the jaw, and various powerful sucking movements. During inspiration there is constriction and during expiration the upper respiratory chamber of the pharynx normally expands.

The mucous membrane and underlying connective tissue of the immediate subglottic region are loosely attached. Since the subglottic space is bouned by the rigid ring of the cricoid cartilage, the tissues enlarge at the expense of the airway when swelling and edema occur.

Nerve supply

The nerve supply of the larynx, both sensory an motor, is through the tenth cranial nerve (cranial nerve X) via the superior an inferior laryngeal nerve branches. Laryngeal paralysis is virtually always caused by a peripheral nerve lesion, since each side has bilateral representation in the cerebral cortex. Because the left recurrent laryngeal nerve runs under the arch of the aorta in a longer course than the right nerve does, it is liable to be affected by a greater variery of disease; thus motor paralysis is commoner on the left side than on the right.

Physiology

Laryngeal reflexes

A number of vital laryngeal reflexes exist. Laryngeal spasm, bronchial constriction, slowing of the heart rate, and apnea are serious, life-threatening effects on the cardiovascular and respiratory systems, mediated by well-known laryngeal reflexes affecting the cariovascular and respiratory system. The administration of vagolytic drugs and the application of topical anesthesia reuce the effect of these powerful laryngeal reflexes during endoscopic manipulation.

Protection

The protective function of the larynx prvents contamination of the airway by food and fluid during swallowing as the vocal cords an the laryngeal sphincter mechanism completely close. The shape and backward movement of the epiglottis divert the bolus to each side into the piriform fossae before it enters the esophagus. The cough rflex, activate by receptors in the larynx and upper trachea, triggers an explosive cough against closed vocal cords to remove any foreign substance from the airway before serious aspiration to the lower airways can occur - a vital protective function of the larynx.

Phonation

Phonation is the secon primary function of the larynx, and an abnormality of this function in the newborn chil prouces an abnormal or feeble cry or no cry at all.

History of Clinical Features

Significant facts relating to the congenital abnormality are obtained from the parents, the medical attendants (including the obstetrician, pediatrician, or neonatologist), and the nursing staff.

The nature of the mother's pregnancy (hydramnios is associated with tracheoesophageal fistula) and the course of the perinatal period (a traumatic or forceps delivery may cause nasal trauma) are important. Some clinical conditions, such as severe micrognathia, are obvious as soon as the baby is born. Neonatal respiratory distress caused by major airway obstruction may be apparent immediately after birth: bilateral choanal atresia, bilateral vocal cord paralysis, large congenital cyst, or major laryngeal web. Periods of cyanosis can accompany the stridor associated with total nasal obstruction, but if the infant cries and takes a breath through the mouth, the airway obstruction is momentarily relieved.

If both vocal cords are paralyzed, stridor is always present, the cry may be weak, and mucus may be aspirated. A muffled or absent cry suggests supraglottic pharyngeal obstruction caused by a cyst or a mass. A harsh barking cough or biphasic inspiratory and expiratory stridor, especially with retraction of the head and neck, suggests tracheal obstruction caused by stenosis, compression, or collapse.

A history of respiratory distress and aspiration with feeding in the neonatal period signals the possibility of a communication between the respiratory and digestive tracts, such as a tracheoesophageal fistula or a posterior laryngeal cleft.

Some congenital lesions do not present their features at birth, and the features are delayed for some days or weeks. Examples include laryngomalacia, in which the stridor may not appear for several days or even a week or more after birth; vascular compression of the trachea, which may appear weeks or months after birth; and subglottic hemangioma, the symptoms of which may not appear for several months.

Physical Examination

General examination

An estimated 10% of all newborn babies, especially those who are premature, have some form of respiratory difficulty in the first few hours of life, but most recover quickly. Most cases of respiratory distress are caused by hyaline membrane disease; some are caused by respiratory depression from drugs. Probably less than 5% of persistent neonatal respiratory distress is caused by congenital anomalies of the upper respiratory tract, but they are of great significance. When one anomaly exists, one must remember that other anomalies may also exist, either in the respiratory tract or in other organ systems.

Examination may show respiratory distress, a rapid or even slow respiratory rate, and signs of hypoxia or cyanosis. The normal respiratory rate is rapid in a newborn baby and slower in older children. Physical signs found in a general examination of the baby may lead to suspicion of a specific condition.

Complete nasal obstruction may result in death from asphyxia. For example, bilateral posterior choanal atresia is associated with periods of respiratory obstruction. Vigorous inspiratory efforts produce marked chest retraction and cyanosis, but if the infant cries and takes a breath through the mouth, the airway obstruction is relieved until the crying stops. The mouth closes until the cycle of obstruction is repeated. These features should alert the attending physician to the possibility of serious nasal obstruction and the probability of bilateral choanal atresia.

A baby with obvious signs of Pierre Robin sequence may have serious upper airway problems caused by oropharyngeal obstruction from micrognathia and a retroposed tongue. Similar signs are seen in babies with Treacher Collins' syndrome, Crouzon's syndrome, Apert's syndrome, and macroglossia, whether it is idiopathic caused by lymphangioma or hemangioma or associated with cretinism.

Other conditions obvious at a general examination include large neck masses such as a hemangioma or lymphangioma, a large thyroglossal duct cyst, or even a branchial cyst, which may compress the airway in the pharynx or the trachea. Most of the conditions causing respiratory obstruction in the newborn baby are congenital, but not all (for example, trauma from an endotracheal tube), whereas respiratory difficulties in older patients are usually acquired.

In older children the larynx and hypopharynx can often be inspected by indirect examination with the laryngeal mirror, but this examination is not possible in infants. The larynx and related structures can be adequately inspected only by direct laryngoscopy, which allows accurate visualization of all structures. The laryngologist's clinical examination relies on evaluation of the facies, the nasal airways, the tongue, the oropharynx, and the neck, as well as on chest examination and auscultation. The neck and the laryngeal structures are palpated for position and genetal outline.

The description, duration, and nature of the features described from the history must naturally be correlated with the findings from the physical examination. Although these features or combination of features may suggest the possibility of the underlying pathologic condition, few are absolutely diagnostic. Usually the clinician obtains radiologic studies before proceeding to thorough endoscopic examination with the patient under general anesthesia.

Specific presenting features

Stridor

The cardinal sign of airway obstruction is stridor - well known to all medical practitioners. The sound, abnormal and distinctive, is produced by turbulence of the airflow in the upper airways. Stridor is usually inspiratory but occasionally expiratory; it is caused by some degree of respiratory obstruction. It may be accompanied by physical signs of airway obstruction, a weak or absent cry, repeated aspiration, cyanotic or apneic attacks, cough, swallowing difficulty, or lower respiratory tract manifestations such as pneumonia or bronchitis. Stridor is heard by anyone listening beside the patient. A wheeze like the expiratory accompaniment of asthma may also be heard if it is loud but is usually heard by auscultation of the chest with the stethoscope. The vibration set up by severe stridor, for

example in laryngomalacia, may vibrate the surrounding tissues in the neck or chest. The parent can often feel it by placing a hand on the child's chest. The parent may state that the child's chest "rattles".

Stridor's cause may be found in the nose or nasal cavities (as in bilateral posterior choanal atresia), in the pahrynx (as in micrognathia and glossoptosis), in the larynx (as in laryngomalacia), in the trachea (as in compression of the trachea by a vascular ring), or even in the main bronchi (as in congenital bronchial stenosis). The congenital abnormalities of the pharynx and larynx that cause airway obstruction are listed as follows:

Nasopharyngeal obstruction

--> Bilateral posterior choanal atresia

--> Turbinate hypertrophy ("stuffy nose syndrome")

--> Deviated septum caused by birth trauma

--> Encephalocele, dermoid, chordoma, hamartoma, and so on

Facial skeletal abnormalities

--> Pierre Robin sequence (micrognathia, glossoptosis, cleft palate)

--> Treacher Collins' syndrome (mandibulofacial dysostosis)

--> Apert's syndrome (acrocephalosyndactyly)

--> Crouzon's syndrome (craniofacial dysostosis)

Oropharyngeal obstruction

--> Macroglossia

--> Lingual thyroid or internal thyroglossal duct cyst

--> Pharyngeal tumors, such as cystic hygroma, dermoid, aberrant thyroid tissue

Laryngeal obstruction

1. *Supraglottic*

--> Laryngomalacia

--> Ductal retention cyst (eppiglottic or vallecula cyst)

--> Saccular cyst

--> Cystic hygroma

--> Bifid epiglottis

--> Hypoplasia of epiglottis

--> Pharyngolaryngeal web (probably a variant of normal)

2. *Glottic*

--> Vocal cord paralysis

--> Web and atresia

--> Interarytenoid web

--> Posterior laryngeal cleft

--> Artrogryposis multiplex congenita

--> Cri-du-chat syndrome

--> Anterior laryngeal cleft

Congenital neurovascul dysfunction (Plott's syndrome)

3. *Subglottic*

--> Congenital subglottic stenosis

--> Subglottic hemangioma

--> Web and atresia

--> Cysts.

Congenital abnormalities of the trachea that cause airway obstruction are listed as follows:

--> Tracheoesophageal fistula

--> Tracheomalacia

--> Vascular ring compression

--> Innominate artery compression

--> Stenosis

--> Tracheal compression

--> Hemangioma

--> Webs

--> Atresia

--> Tracheogenic duplication cysts.

The characteristic of the stridor may be a clue to the location of the obstruction. A harsh, high-pitched, crowing noise during inspiration often indicates an abnormality in the larynx, in the subglottic region, or more often in the supraglottic tissues. Stridor of lower pitch with snoring and excessive secretions may indicate a pharyngeal or nasopharyngeal obstruction. Stridor that is both inspiratory and expiratory with a prolonged low-pitched expiratory phase suggests obstruction of the trachea or even main bronchi from compression oor collapse.

Stridor and respiratory distress may also be caused by conditions outside the respiratory tract. For instance, a large abdominal mass, a diaphragmatic hernia and eventration, a reduplication cyst, a hemangioma, or a lymphangioma can narrow the major airways.

The clinician must know when the stridor commenced, how long it has persisted, whether it is constant or intermittent, and what has been its progress and degree. Its relation to inspiration and expiration, the effect of sleeping, eating, crying, posture, and possible association with aspiration may all be important. Observation and examination over a period of hours or days may give useful information not detectable at a single examination.

Correlation of the clinical features, physical signs, radiologic abnormalities, and finally the endoscopic changes is essential to localized the cause and arrive at a correct diagnosis. Endoscopic examination of a patient under general anesthesia always includes the oropharynx, larynx, trachea, and bronchi, often the esophagus, and, when indicated, the nasal cavities and nasopharynx. Laryngoscopy alone is complete because pathologic conditions in the tracheobronchial tree may remain undiagnosed. Over 10% of patients have lesions at more than one anatomic site in the upper aerodigestive tract (Friedman et al, 1984).

Stridor or *congenital laryngeal stridor* are all-inclusive terms but *never* diagnoses. If possible the term should be qualified as to its primary cause and its anatomic location, since many reasons exist for stridor in an infant and not all are congenital. Laryngoscopy alone in the investigation of "congenital laryngeal stridor" without examination of the tracheobronchial tree and, if necessary, the esophagus must be regarded as an incomplete examination.

Indications for Endoscopy

Diagnostic endoscopic examination to investigate stridor is indicated when one of the following exists:

Severe stridor

Progressive stridor

Stridor associated with unusual features, such as cyanotic attacks, apneic attacks, dysphagia, aspiration, failure to thrive, or a radiologic abnormality

Stridor causing undue parental anxiety.

Signs of obstruction

Airway obstruction may be present from birth and persistent, recurring with every breath, as in bilateral vocal cord paralysis. It may also be variable and phasic, as in laryngomalacia. Finally, it may be relentlessly progressive as with an enlarging cyst.

Major congenital airway obstruction in a newborn baby produces stridor, rapid breathing with increased effort indicated by retraction of the chest, epigastric in-drawing, tracheal tug, suprasternal and intercostal retraction, and flaring of the nasal alae during inspiration. As obstruction progresses, even vigorous use of the accessory respiratory muscles and tachypnea might not prevent respiratory failure with impaired pulmonary ventilation, cyanosis, and subsequent bradycardia as the levels of hypoxia and hypercarbia increase.

With chronic airway obstruction, sternal retraction caused by persistent, negative pleural pressure and high compliance of the rib cage may be exaggerated. With a long-standing obstrucion, a permanent pectus excavatum may result.

In seriously ill patients with lower airway obstruction and lung parenchymal lesions causing respiratory failure, blood-gas and blood-pH studies may be required as single or serial examinations to assess the degree of respiratory failure and to assist in the management of respiratory or metabolic acidosis. In patients with obstruction of the upper airways, blood gases may remain normal or near normal even with severe obstruction. Clinical assessment is much more important.

Cry abnormalities

Normal phonation depends not only on the subglottic air pressure but also on the length, tension, and mass of the vibrating vocal cords and the ability of the straight free medial margin of the vocal cord to vibrate freely. Any change in these variables produces an abnormality of vocalization.

When both vocal cords are paralyzed, stridor always exists, and the cry, although usually weak, is clear. With unilateral cord paralysis the cry may be weak and feeble, but usually no serious ariway obstruction exists. Depending on the size of the laryngeal web, complete aphonia or a weak, breathy, and feeble cry may exist; with smaller webs minimal impairment of phonation may occur. A muffled or absent cry in infants may relate to pharyngeal involvement, for example, from a cyst or other pharyngeal obstruction. Cysts in or about the larynx affect the voice as well as obstruct the airway.

The cri-du-chat syndrome (Ward et al, 1968) is a rare but interesting cause of a weak and high-pitched abnormal cry resembling the meowing of a cat.

Aspiration

Congenital anomalies and neurologic conditions of the oral cavity, pharynx, larynx, trachea, and esophagus are the most common causes of repeated aspiration and aspiration pneumonia with segmental atelectasis. Recurrent aspiration frequently causes failure to thrive.

Feeding difficulty and incoordinated swallow and aspiration are often features in patients with cleft palate, mandibular hypoplasia and glossoptosis, disorders in the mid-facial skeleton, and choanal atresia, as well as esophageal abnormalities such as stenosis, webs, and duplication.

The differential diagnosis of repeated aspiration in infants requiring endoscopic investigation includes gastroesophageal reflux with aspiration, neuromuscular incoordination of swallowing in central nervous system disorders, bulbar paralysis, tracheoesophageal fistula, cleft larynx, and vagal paralysis with insensitivity of the superior half of the larynx with ipsilateral recurrent laryngeal nerve paralysis.

Radiologic studies with contrast material used for the esophageal swallow and cineradiography are traditional investigations. Scanning for radioactive-labeled milk may also be useful, especially for gastroesophageal reflux. Reflux may occur after the time of swallowing, with return of gastric contents and aspiration into the tracheobronchial tree. Esophagoscopy with biopsy and pH monitoring may give useful information.

In some cases of repeated aspiration, gastric tube feedings are an essential part of the management during investigation and observation to prevent further deterioration and to provide the required nutritional support.

Cough and apnea

Coughing is an essential component of the body's defense mechanism and is a primitive reflex that is most reliable in protecting the lower respiratory tract. As a symptom, cough represents a physiologic response to a variety oof respiratory tract stimuli. Repeated, persistent coughing in a baby is distressing to parents.

Cough is not a common feature of disorders of the respiratory tract in babies. When it does occur, however, cough is important as diagnostic aid. The onset, frequency, and force of the cough and the presence of secretions should be noted. For example, coughing associated with feeding or with drinking may be caused by aspiration. A persistent barking cough with or without stridor is a prominent characteristic of tracheal narrowing and is suggestiive of tracheomalacia or tracheal compression.

In tracheal compression or collapse, the head and neck may be held in hyperextension, and cyanotic and/or apneic attacks may occur. Apneic attacks occurring with inominate artery compression of the trachea or with tracheomalacia associated with tracheoesophageal fistula have been called "reflex apnea" attacks or "dying spells". A prolonged apneic attack leads to

cyanosis, bradycardia, or even cardiac arrest.

Radiologic Examination

After correlation of evidence obtained from a careful history and physical examination, an appropriate radiologic examination should be considered in infants with possible congenital abnormalities of the hypopharynx, larynx, and trachea, especially if stridor and airway obstruction exist.

Anteroposterior chest radiographs are a routine requirement. An anteroposterior view of the tracheal air column using a high-kilovolt technique to enhance the air column may be useful, for example, in subglottic hemangioma or tracheal stenosis. The film should be taken, if possible, during both inspiration and expiration, since changes in the mobile soft tissues of the pharynx change the appearance (Benjamin, 1975).

A lateral study of the upper airways with a radiograph or xeroradiogram is particularly useful. A well-exposed film with the patient's head and neck in the hyperextended position consistently provides worthwhile information, often of essential diagnostic value. Many variations are seen in a normal lateral-airway radiograph in infants. Experience in interpretation distinguishes the normal from the abnormal. The most reliable landmark in the larynx is the laryngeal ventricle, which is normally distended with air. It must be emphasized that apparent abnormalities in an otherwise normal airway can be produced by poor positioning or exposure technique. When possible, the radiographic investigations should be undertaken, except in an extreme emergency, before a patient is examined by endoscopy under a general anesthetic.

Xeroradiography has been used for many years in evaluating the upper respiratory tract and has a very useful application in the larynx. Its unique properties of enhancing the air-soft tissue edge and having a wide exposure latitude are distinct advantages over conventional radiologic technique, and in many patients the airway can be seen from the nasal cavities above to the bifurcation of the trachea below. The higher radiation dosage associated with xeroradiography must be carefully considered and balanced against the clinical needs for the particular patient and the possibility of added useful information (Fig. 99-2).

Contrast laryngography has no place in the investigation or documentation of infant laryngeal problems.

The contrast esophagogram is essential in the investigation of some infants and may demonstrate gastroesophageal reflux. Although the barium study has traditionally been regarded as a diagnostic radiologic investigation, the unreliability of a single barium swallow examination, even when performed by an experienced radiologist, is well known. However, the esophagogram may help in the diagnosis of a vascular ring, H-type tracheoesophageal fistulas, swallowing difficulties caused by neuromuscular incoordination, mediastinal cysts or tumors, and even a posterior laryngeal cleft. There are no studies to show that computed tomography (CT) has additional advantages in assessing the laryngeal airway, but it is of great advantage in investigating a pulmonary or mediastinal mass, whether solid or cystic - for example, neurogenic tumor, teratoma, lymphosarcoma, and bronchogenic or duplication cyst (Fig. 99-3).

In selected cases contrast tracheobronchography (as for congenital stenosis of the trachea or bronchi) and occasionally angiocardiograpphy (as for a vascular ring) are helpful. Clearly then, investigation of the larynx and pharynx alone without study of the tracheobronchial tree and, if necessary, the esophagus must be regarded as an incomplete investigation of some congenital abnormalities of the upper airways.

Care must be taken in the interpretation of a single film because the soft tissue structures are subject to changes in caliber and appearance from moment to moment. The pharynx, larynx, and trachea may be visualized during any phase of respiration or deglutition. Although two or more films are taken, they may not necessarily represent the phases of respiration, since ensurijng an inspiratory and expiratory film in infants is often difficult.

Symptoms relating to the airway and to the esophagus often coexist in infants who have aspiration caused by gastroesophageal reflux, tracheoesophageal fistula, or a repaired tracheoesophageal fistula. In some cases secondary lower respiratory tract disease is either clinically or radiologically apparent.

Endoscopy

Anesthesia

Cooperation, mutual understanding and teamwork between the surgeon and the anesthesiologist are imperative for the safety of the baby during endoscopic examination of the upper airways. Rarely do patients undergo examination without general anesthesia. Occasionally in sick neonates up to a few weeks of age or in those with suspected vocal cord paralysis, no anesthetic is used, but the anesthesiologist is in attendance. Intubating a baby without general anesthesia may sometimes be preferable in the Pierre robin sequence, in which a difficult airway problem exists.

The most common general anesthetic technique for neonatal endoscopy (Benjamin, 1984) relies on spontaneouos respiration using insufflation of nitrous oxide, oxygen, and halothane. Some anesthesiologists prefer to add methoxyflurane to the gaseous mixture to provide additional analgesia of longer duration, thus "smoothing out" the procedure. In the case of an ill child, oxygen with only halothane is used.

Atropine as a single agent is used for premedication in babies. It can be given by intramuscular injection properatively or intravenously at or soon after induction of anesthesia. Atropine affords protection against bradycardia and helps minimize secretions in the respiratory tract. Topical anesthesia using lidocaine (up to 5 mg/kg) in a dilute solution minimizes unwanted reflex activity and is regarded as most important, since it usually abolishes laryngeal spasm during endoscopy. However, laryngealspasm can occasionally be a complication in endoscopy with spontaneous respiration. A venipuncture is always performed with endoscopy, and a muscle relaxant is ready to be given if required.

Instruments

Several sizes and types of pediatric laryngoscopes should be available, including the Holinger anterior commissure infant laryngoscope. A complete range of rigid bronchoscopes, starting with the 2.5 mm instrument and with adaptations to allow ventilation, is available. A variety of small-diameter Hopkins fiberoptic rigid telescopes, whose light may be directed straight ahead or at various angles, is necessary. The smallest has an outside diameter of 1.9, then there are 2.8 and 4.0 mm telescopes, they are used constantly in the larynx, tracheobronchial tree, and esophagus for detailed examination. Telescopes with viewing angles of 0, 30, and 70 degrees can be conveniently used to examine not only all parts of the larynx but also the tracheobronchial tree with previously unavailable clarity and precision (Fig. 99-4).

We have found few routine uses for the flexible fiberoptic pharyngoscope, laryngoscope, or bronchoscope in the investigation of congenital disorders of the larynx. It may be useful, however, to observe laryngeal dynamics in the infant with laryngomalacia, whether the infant is awake or under general anesthesia and breathing spontaneously. A flexible neonatalscope, such as that described by Silberman et al (1984), designed for the examination of intubated neonates, should have valuable and interesting applications in the placement of endotracheal and tracheotomy tubes and for localizing areas of obstruction.

Technique

A preliminary survey of the larynx and pharynx with the naked eye is accomplished wiith the hand-held laryngoscope (Fig. 99-5). Hopkins rigid telescopes provide a magnified image for detailed examination of anatomic structure and mucosa. Although direct laryngoscopy is used principally to examine the larynx, other specific anatomic areas are always evaluated. These areas include the oropharynx, the base of the tongue and valleculae, the piriform fossae, the postcricoid region, the epiglottis, the arytenoids, the false cords, the ventricles, the vocal cords (including the anterior and posterior commissure), the subglottic region, and the trachea. External pressure and manipulation of the larynx with a finger or fingers on the neck, with gentle counterpressure from the distal beak of the laryngoscope, rotate or displace the larynx sufficiently to make the other side of the endolarynx or subglottic region more prominent and easier to visualize.

The unique design of the Lindholm laryngoscope (Karl Storz, Tuttlingen, Germany) provides a panoramic view of the laryngopharynx by placing the beak of the laryngoscope at the base of the tongue in the vallecula in front of the epiglottis. Although designed for adults, it can be used satisfactorily, without trauma, in most infants over 6 to 9 months of age. An infant size instrument is now available. These laryngoscopes are especially useful for microlaryngoscopy and laser surgery. The standard slotted Storz pediatric laryngoscopes of 8.0, 9.5, 11.0, and 13.5 cm in length or the Jackson pediatric laryngoscopes with a slide are used routinely. If the larynx is difficult to expose or examine, a special-purpose laryngoscope such as the Holinger pediatric anterior commissure laryngoscope, the Benjamin pediatric operating microlaryngoscope, or the smaller Tucker-Benjamin slotted laryngoscope and subglottiscope for newborn and premature infants are useful. They are helpful, for instance, in exposing the larynx in micrognathia or abnormalities of the middle third of the face. The anterior and posterior commissures and the interarytenoid region can be seen and displayed by deliberately separating the vocal folds; the subglottic region is more satisfactorily exposed.

Laryngoscopy with telescopes gives a magnified image of the anatomic structures using the standard 0-degree straight-ahead rigid Hopkins telescope and the 30- or 70-degree angled telescope for the laryngeal ventricles, the anterior and posterior commissures, and the anterior subglottic region.

Examination of the trachea, main bronchi, and segmental openings is an integral part of endoscopy for congenital airway problems. The patient continues to breathe the anesthetic mixture through the ventilating bronchoscope while the topical analgesic can be applied in measured doses to the tracheal mucosa or to the carina if necessary. Again, the rigid Storz-Hopkins telescopes are used alone, passed through a laryngoscope only or through a bronchoscope to examine the contour, caliber, and color of the airways as the dynamics of inspiration and expiration continue. Secretions may be aspirated and collected in a trap bottle for examination and culture. Any abnormal compression, collapse, or pulsation is noted. Biopsy of a lesion is necessary only very occasionally.

It is important that pressure from the tip of the laryngoscope blade or the direction of its introduction does not disturb the assessment of vocal cord function and general laryngeal dynamics. Such interference may lead to a false diagnosis of vocal cord paralysis. With the laryngoscope blade in the vallecula, and sufficient time for complete evaluation during inspiration and vocalization, the examination will be reliable. Assessment of vocal cord movement and examination of the typical changes of laryngomalacia are best performed when the anesthesia has been discontinued at the end of the procedure and the patient is recovering pharyngeal and laryngeal movements.

Endoscopic examination of the larynx, pharynx, tracheobronchial tree, and esophagus can be successfully and safely performed in a patient at any age - if necessary, even in premature infants weighing less than 1000 g. The hospital must have the necessary equipment for investigation, together with experienced anesthetic and endoscopic personnel. A fully staffed postoperative recovery ward and intensive care area with facilities to recognize, diagnose, and manage potential complications immediately are necessary. A multidisciplinary approach with close cooperation between the pediatrician, endoscopist, anesthesiologist, and others involved in the child's care is essential.

Specific Abnormalities

Pierre Robin sequence

The Pierre Robin sequence consists of mandibular hypoplasia (micrognathia), glossoptosis (retroposed tongue), and an incomplete midline cleft of the palate, although an occasional patient may not have one of these features. The condition usually presents as an isolated anomaly or as part of a syndrome, commonly Stickler's syndrome. Estimates of incidence range from 1:2.000 to 1:50.000 births. The cause is unknown, but some patients have a family history of micrognathia or cleft palate.

Upper airway obstruction is common. The severity is less in the prone position as the tongue falls forward, partly relieving pharyngeal obstruction. Severe obstruction may cause cyanotic episodes, cerebral hypoxia, aspiration pneumonia, right-sided heart failure, or fatal asphyxia. In an infant with micrognathia and glossoptosis, the larynx is under the base of the

tongue, making direct laryngoscopy and intubation difficult and sometimes impossible (Fig. 99-6).

The onset of obstruction is usually within a few hours of birth, but it is not possible at first to assess the future severity of the airway obstruction. Therefore repeated observation in the neonatal intensive care ward is needed to determine the best method of airway management. Monitoring by pulse oximetry is performed in every case; a reasonable guide is to have the oxygen saturation in room air over 85% for at least 90% of the time. Airway management should be individualized until successful control is achieved, using the following progressive sequence:

--> Posturing in the prone position

--> Nasopharyngeal tube

--> Endotracheal intubation

--> Tracheotomy.

The method may partly depend on the severity of associated cardiac and other abnormalities that commonly coexist. Posturing prone is the most important initial part of treatment; the tongue falls forward, partly clearing the oropharyngeal airway. Posturing can be discontinued when the child is able to maintain an airway sleeping supine, at about 5 or 6 months of age in most children.

Nasogastric tube feeding may be needed if aspiration occurs during feeding.

In cases where posturing is unsuccessful, a nasopharyngeal tube is passed through one nasal cavity and positioned so the tip is below the level of the base of the tongue but just above the larynx to provide an alternate artificial airway. Posturing may still be necessary.

A variety of operative techniques to secure the tongue forward have been described, but most are no longer used. In an emergency, traction using a toothed forceps or a suture to pull the tongue forward will immediately improve the airway.

If positioning prone or use of a nasopharyngeal tube does not provide a reliable airway, emergency endotracheal intubation or definitive tracheotomy should be considered. Endotracheal intubation performed under controlled conditions will provide an excellent temporary airway, but there are disadvantages. Laryngoscopy for intubation is always difficult and sometimes impossible without special-purpose laryngoscopes such as the Holinger pediatric anterior commissure laryngoscope or the smaller Tucker-Benjamin laryngoscope for premature or small babies. The patency of the tube must be carefully maintained. Accidental extubation may occur with failure to reintubate, and laryngeal trauma from prolonged intubation is a distinct possibility. Tracheotomy is the preferred long-term artifical airway.

If other techniques fail, tracheotomy is safe and dependable to successfully bypass the obstruction and should be performed for episodes of cyanosis observed clinically and desaturation measured objectively. There should be no reluctance or delay in performing a

tracheotomy because of the relatively high chance of hypoxic cerebral damage or death. The tracheotomy can usually be removed between 6 to 18 months of age.

The patients can be divided into three distinct groups according to the airway management found to be necessary for the severity of airway obstruction and the eventual outcome.

Mild	Posture alone
Moderate	Nasopharyngeal tube
Severe	Short-term intubation, tracheotomy, or death.

Experience shows that no one method of airway management is effective for every patient. Nasotracheal intubation in many cases can secure a certain, safe airway that can be maintained for many weeks with painstaking nursing care if necessary over a prolonged period. The airway obstruction inevitably improves with time as the anatomic structures assume a more normal anatomic relationship and the tone in the tongue and pharyngeal musculature improves.

The death rate can be from 10% to 20%. The deaths from respiratory obstruction indicate the prime importance of airway management and the need for tracheotomy in severe cases.

Abnormalities of midfacial skeleton

Infants with Treacher Collins' syndrome (mandibulofacial dysostosis), Apert's syndrome (acrocephalosyndactyly), Crouzon's syndrome (craniofacial dysostosis), and other diseases may have nasal airway obstruction, retrognathia, and malocclusion, so that posterior displacement of the midfacial structures causes oropharyngeal upper airway obstruction. In older children with these anomalies sometimes adenoid hypertrophy exists, making the chronic nasopharyngeal obstruction worse and causing episodes of obstructive sleep apnea. In these patients removal of the adenoids and tonsils may be dramatically effective.

In the neonatal period, however, conservative management of the airway obstruction by vigilant nursing maintenance of a suitable posture and supportive measures is usually sufficient. In severe or refractory cases, a tracheotomy is indicated.

Oropharyngeal obstruction

A cause of oropharyngeal obstruction is macroglossia, which may be primary (cretinism, Beckwith-Wiedemann syndrome), secondary (lymphangioma, hemangioma), part of another congenital syndrome, or idiopathic. Reduction of tongue size by surgical excision offers an acceptable functional result with minimal morbidity (Rizer et al, 1985).

Aberrant thyroid tissue is most commonly seen as lingual thyroid at the foramen cecum in the base of the tongue but has been reported elsewhere in the pharynx as a cause of airway obstruction. Great care must be taken before a lingual thyroid is considered for removal, since it may be the only functioning thyroid tissue in the body. A radioactive thyroid scan detects any other functioning thyroid tissue that is present.

Mucous-retention cysts or ductal cysts most frequently occur in the vallecula. Other pharyngeal masses include dermoid cysts, teratomas, and chordomas. The treatment of these various conditions is symptomatic for each case. With a ductal mucous-retention cyst, aspiration, "deroofing", or marsupialization may be necessary.

Laryngomalacia

Laryngomalacia is the most common cause of stridor in infants. The term *congenital laryngeal stridor* should never be used as a definitive diagnosis - it is no more a diagnosis than "fever" or "pain" or "anemia" in general medicine. Stridor has multiple causes in an infant; not all are congenital nor is the stridor necessarily generated from the larynx region. The cause of stridor in a particular patient must be properly determined. If stridor is used as a diagnostic term and associated with a specific disease, diagnosis of other laryngopharyngeal anomalies may be delayed, forgotten, or inadvertently disregarded.

The cause of laryngomalacia is not known with certainty. The abnormal flaccidity of the laryngeal tissues is probably a temporary physiologic dysfunction that resolves with growth. Most cases resolve spontaneously within 6 to 18 months. Immaturity of tissues or histopathologic abnormality of the cartilage with increased softness has never been proven. The role of cartilaginous rigidity and elasticity is not known. Some claim that laryngomalacia is a manifestation of delayed development of neuromuscular control or is caused by an anatomic abnormality (Belmont and Grundfast, 1984).

Diagnostic evaluation

Physical findings. The important features of laryngomalacia include variable inspiratory stridor, signs of intermittent upper airway obstruction, a normal cry, and normal general health and development. Although persistent, noisy stridor from laryngomalacia is distressing to anxious parents, the condition seldom affects the baby's general state of health and progress. It is twice as common in boys as in girls.

Stridor very seldom exists at birth but usually begins in the first few days or weeks of life and persists thereafter as a variable inspiratory accompaniment. It is usually described as harsh and crowing, but at times it is low pitche an fluttering. Sometimes vibrations may be felt by a hand on the infant's chest. The stridor is usually worse with crying and feeding, during periods of excitement or activity, or when the child is lying on his back with the head and neck flexed. On the other hand, stridor may improve when the child lies prone or when the head and neck are extended. Elevation of the mandible and submental soft tissues with a finger may partly improve the airway obstruction. The stridor is intermittent and variable: at times the child sleeps quietly and at other times he seems "mucousy", but aspiration of mucus from the pharynx does not relieve the symptoms. Observation and reexamination over a period of days or weeks may give useful information that is not apparent at a single examination. If chest retraction is severe and prolonged, pectus excavatum (sternal retraction) may develop.

Laryngomalacia appears to be caused by flaccidity or incoordination of the supralaryngeal structures, including the cartilages and the soft tissue. Some patients have an associated micrognathia, but this is not consistently seen. The cry is clear, strong, and normal.

Cyanotic attacks are so uncommon that if they occur, the presence of some condition other than laryngomalacia must be suspected. Similarly, feeding is occasionally slow and noisy; if dysphagia exists, especially if it is associated with aspiration, then the clinician should be alerted to some other condition.

Endoscopic examination. A confident diagnosis of laryngomalacia can be made only by direct endoscopic examination during respiration. Not only does complete endoscopic investigation positively confirm the diagnosis, but also the possibility of any associated abnormality in the tracheobronchial tree is excluded. The clinician completes a careful endoscopic examination of the upper respiratory tract with and without telescopes. As the administration of anesthesia is discontinued and the patient is regaining muscular tone, the examiner reintroduces the laryngoscope to watch for the typical changes, with the blade at the base of the tongue above and in front of the epiglottis. The epiglottis may be abnormal: tall, narrow, and folded in on itself so that its lateral margins lie close together. Traditionally endoscopists have described the "infantile" epiglottis with laryngomalacia, but many patients who have neither laryngomalacia nor stridor have an omega-shaped or tubular epiglottis, indicating that the shape of the epiglottis itself is not as important as the flaccidity and tendency of the supraglottic tissues to collapse. The aryepiglottic folds and arytenoids are tall, thin, pale, and flaccid, appearing lax and redundant. During each inspiration they are sucked into the larynx. On expiration they are blown up and out, so that expiration is unimpeded. When the epiglottis and the aryepiglottic folds of the supraglottic laryngeal introitus are splinted outward by a laryngoscope, the stridor is immediately corrected and the airway is unimpeded. Sometimes the characteristic changes of laryngomalacia are seen during the initial laryngoscopy, but more often they are seen at the end of the procedure as the administration of anesthesia is discontinued (Fig. 99-7).

Flexible fiberoptic laryngoscopy can be used to confirm the abnormality of the supraglottic structures in laryngomalacia, but the examination is inadequate unless the subglottic region and tracheobronchial tree are also examined endoscopically, since some patients have associated pathologic findings in other areas.

Radiologic examination. Although chest and lateral airway radiographs are routinely obtained in the investigation of stridor, radiologic examination of laryngomalacia is normal. No diagnostic radiologic features can be seen in single films.

Management

Most infants with laryngomalacia gain weight and mature normally. With satisfactory general progress, diagnosis by endoscopic examination usually allays the parents' concern. Active treatment is unnecessary. The parents should receive reassurance that the symptoms will subside as the months go by and with growth and development of the larynx. Only very rarely is surgical treatment required. In extreme cases a tracheotomy may be necessary when the respiratory obstruction is severe or associate with feeding difficulties and failure to thrive. In the few cases that require tracheotomy, the infants can be intubated for a week or so to observe the effect on their nutrition and weight and to observe the improvement that occurs when each inspiration is not a struggle to survive.

Recently, severe cases of stridor caused by laryngomalacia, judged to be causing hypoxia and failure to thrive, have been treated by surgical means. The carbon dioxide laser is used to divide and "release" the aryepiglottic fold on one or both sides. The indications and the efficacy of the procedure requires further assessment.

Laryngeal cysts

Cysts of the larynx and pharynx can be classified as follows:

--> Ductal cysts, which occur anywhere a mucous-producing gland exists.

--> Saccular cysts, either lateral or (less commonly) anterior; the saccule is normally small and contains air.

--> Thyroglossal duct cysts, which occur at the tongue base at the foramen cecum.

--> Cystic hygroma or lymphangioma, which is a multilocular cystic developmental abnormality arising from lymph vessels.

Ductal cysts

Ductal cysts are also known as mucous-retention cysts. They are the cysts found in the valleculae within the larynx or the subglottic region. They result from retention of mucus in dilated collecting ducts of the submucosal glands and are usually less than 1 cm in diameter, being quite superficial and remaining within the mucous membrane. Thus the common vallecular cyst is merely a ductal cyst in the anatomic region of the vallecula. Similarly, a congenital subglottic cyst is a ductal cyst in the subglottic region of the larynx.

In infants who have undergone prolonged intubation, ductal cysts are occasionally seen in the subglottic region (Fig. 99-8) and may be multiple. These cysts are caused by irritation and obstruction of mucous gland ducts. They might not be clearly seen on a lateral airway radiograph but are diagnosed at direct endoscopic examination. They are treated by removal either by forceps or laser, with care being taken for reexamination to treat possible recurrence. A small intracordal cyst (Fig. 99-9) requires removal by laryngeal microsurgery.

Saccule cysts

The saccule of the laryngeal ventricle (Fig. 99-1) is a normal structure seen as an out-pouching of mucous membrane that contains mucous glands, between the false and true vocal cords at the anterior third of the laryngeal ventricle. Delicate intrinsic laryngeal muscles lie meial and lateral to the saccule; by compressing the saccule they are thought to control expression of its secretions onto the vocal cords for lubrication.

A congenital saccular cyst in the newborn (Fig. 99-10) is a dilated saccule filled with mucus that does not communicate with or drain into the laryngeal lumen. Saccular cysts have also been called "congenital cysts of the larynx", "laryngeal mucoceles", and "saccular mucoceles". Saccular cysts are of two types. Although both are rare, the more common is the lateral saccular cyst that extends up to disten the false vocal cord and the aryepiglottic fold

(Fig. 99-11). A lateral saccular cyst is the same thing as an internal laryngomucocele. The anterior saccular cyst is smaller and extends medially into the laryngeal lumen between the true and false cords (Fig. 99-12).

A laryngocele (most often seen in adults) is an abnormal dilation of a saccule. It is distended with air, which causes it to become pathologic and temporarily symptomatic (Fig. 99-13). Apparently, then, congenital laryngeal cysts in the region of the ventricle are saccular cysts and are the counterparts of the ault laryngoceles. In the neonate and infant the saccular cyst is the common pathologic finding. A laryngocele is distinguished from a cyst in that its lumen may be filled with air at times and at other times temporarily distended with mucus that then discharges into the lumen of the larynx through the normal communication, which has been temporarily obstructed. Holinger et al (1978) pointed out that a developmental spectrum exists among the normal saccule, large saccule, laryngocele, and saccular cyst.

In infants a saccular cyst may exist at birth and cause severe respiratory distress with inspiratory stridor, episodes of cyanosis, inaudible or muffled cry, and occasionally dysphagia. Although very rare, an external swelling may exist in the neck; if so, investigation is required for both an external and an internal laryngocele.

Diagnostic evaluation. Soft tissue lateral radiographic studies of the neck and airway show a cyst in all cases. A definitive diagnosis can be made only at the time of direct laryngoscopy. If the radiograph reveals the presence of such a problem, the surgeon should be fully prepared - for example, to decompress a cyst by sucking out its contents or by incising it. Attempts at anesthesia induction can precipitate severe airway obstruction. Endoscopy will reveal a large distended bluish or pink fluid-fille cyst occupying one side of the supraglottic tissues. Consequently, aspiration via a large-bore needle, incision with a sharp instrument, or removal of the dome of the cyst permits drainage of the thick fluid. This procedure may have to be repeated if the cyst re-forms. Very rarely does an infant require an external incision or an excision of the external laryngocele of a persisting cyst. Commonly recurrence requires repeated aspiration or removal of part of the cyst.

Thyroglossal cyst

Thyroglossal duct cysts (Fig. 99-14) can occur anywhere from the foramen cecum to the thyroid gland and the persisting thyroglossal duct. An internal thyroglossal duct cyst occurs at the base of the tongue in the region of the foramen cecum. Such a cyst, proximal to the hyoid bone, may push the epiglottis backward and down, producing severe obstruction of the supraglottic laryngeal lumen. Treatment is by incision, drainage, and removal of the roof.

Cystic hygroma

Cystic hygromas (lymphangiomas) are multilocular developmental congenital malformations rather than true neoplasms and are most common in soft tissues of the lateral neck as diffuse, compressible, smooth, nontender masses that transilluminate. They might be obvious at birth or in the first year or two of life. Histologically they consist of widely dilated lymph vessel spaces. A mass that contains both abnormal lymphatics and blood vessels is a lymphangiohemangioma.

Cystic hygromas are usually asymptomatic except for large masses causing cosmetic deformity or masses that stretch or compress tissues of the pharynx and larynx to produce airway obstruction. A cystic hygroma is virtually always a supraglottic mass, whereas a congenital hemangioma causing airway obstruction is usually subglottic.

Treatment of cystic hygromas is moderately controversial. Aspiration of the contents of the cystic spaces may relieve airway compression if tension within the hygroma is excessive. Treatment of symptomatic lateral neck lesions is by surgical excision, which may be difficult because of undefined surgical planes. External excision of the bulk by laser or some other modality may be required. For a severe chronic obstruction a long-term tracheotomy may be necessary.

Vocal cord paralysis

Vocal cord paralysis, either unilateral or bilateral, accounts for about 10% of all congenital laryngeal lesions (Holinger et al, 1976a). Unilateral vocal cord paralysis probably goes undiagnosed in some infants, and with later recovery of laryngeal function the condition is never documented.

The lesion affecting the motor nerve supply can be anywhere from the nucleus ambiguus in the brainstem to the neuromuscular junction in the larynx, involving the vagus nerve or its recurrent branch. Consequently, each case must be carefully investigated to locate the causal lesion (Cohen et al, 1982).

Unilateral vocal cord paralysis is seen more often on the left side than the right. The etiology is often unknown. However, a variety of congenital cardiovascular anomalies can affect the left side, including ventricular septal defect, tetralogy of Fallot, abnormalities of the great vessels, and patent ductus arteriosus. Surgical damage to a nerve can occur with such events as operation for an H-fistula, cervical esophagostomy, and surgery for a congenital heart defect.

On the other hand, bilateral vocal cord paralysis with both vocal cords unable to abduct results in consistent severe stridor an cyanotic attacks. Aspiration is common with recurrent chest infections and scattered small areas of atelectasis on the chest radiograph. Although one can see bilateral vocal cord paralysis in an otherwise normal infant, a strong association exists with central neurologic abnormalities such as meningomyelocele, hydrocephalus, Arnold-Chiari malformation, bulbar palsy, and birth trauma. Bilateral vocal cord paralysis may occur associate with birth trauma related to difficult, prolonged second-stage labor with difficult forceps delivery and undue traction on the cervical spine. In these cases recovery in 6 to 9 months can be expected. Hereditary bilateral vocal cord paralysis is extremely rare (Gacek, 1976).

Diagnostic evaluation

Clinical presentation. The cardinal clinical features of unilateral vocal cord paralysis in a newborn baby are a weak, breathy cry, often aspiration of pharyngeal secretions, and sometimes cyanotic attacks and choking during feeding. Loss of sensation of the involved hemilarynx contributes to aspiration. Stridor is selom a prominent feature.

A tracheotomy is almost always necessary in cases of bilateral vocal cord paralysis. When a neurologic abnormality exists, however, even though a tracheotomy may relieve the airway obstruction, the abnormal respiratory function, apneic attacks, and cyanotic episodes may persist. Caudal displacement of the brainstem with the medulla and the cerebellum is thought to stretch the tenth cranial nerve (cranial nerve X). It may also cause disturbed function of the displaced respiratory center.

Laryngoscopic examination. Confirmation of the clinical suspicion of unilateral or bilateral vocal cord paralysis must be made by direct laryngoscopic examination. Pressure from the laryngoscope blade or the direction of introduction must not distort assessment of vocal cord function and the shape or general dynamics of the larynx. Such interference may cause a false diagnosis of lost vocal cord movement.

The laryngoscope blade should be placed in the vallecula (in front of the epiglottis) at the end of the procedure, when anesthesia has been discontinued. As the patient recovers pharyngeal and laryngeal movement, time is ample for a complete, reliable evaluation during inspiration and vocalization. Vocal cord movement and the mobility of the cricoarytenoid joint are compared with that of the vocal cord on the other side to detect cricoarytenoid joint function. It is generally easy to make a confident diagnosis of unilateral vocal cord paralysis but not so easy to be certain of bilateral paralysis. For neonates in whom bilateral vocal cord paralysis is suspected but for whom a confident definitive diagnosis cannot be made at the first examination, a nasotracheal tube can be left in situ and reexamination performed some days or a week later. If bilateral vocal cord paralysis is confirmed, then a tracheotomy is performed. A tracheotomy is virtually always necessary for bilateral vocal cord paralysis and sometimes necessary even in cases of unilateral vocal cord paralysis with stridor and partial airway obstruction or troublesome aspiration.

Vocal cord paralysis can usually be reliably assessed with a flexible fiberoptic laryngoscope, either in the unanesthetized child or during spontaneous respiration anesthesia.

Management

Spontaneous resolution of the paralysis sometimes occur. In unilateral cases this results in improvement of the cry. In bilateral cases the airway improvement is usually sufficient for removal of the tracheotomy tube.

Correction of the airway obstruction in unremitting bilateral paralysis is a major problem, and the optimal age for further surgical procedures is conjectural.

No reports have been made of the results of nerve-muscle pedicle reinnervation in the pediatric age group. Reasonably, however, this operation can be considered for patients over the age of 4 years. The surgery is performed exclusively in the soft tissues of the neck; the integrity of the larynx is not breached; and should the operation be unsuccessful, an arytenoidectomy can be performed later at a suitable age. The results of an arytenoidectomy (Cohen, 1973) are more predictable in maintaining a good voice and obtaining a good airway so that decannulation is possible. Some of the patients continue to have stridulous breathing at night and do not obtain normal exercise tolerance.

Laryngeal webs and atresia

Laryngeal webs are not uncommon. They are formed during embryogenesis of the laryngotracheal groove. Actively proliferating epithelium temporarily obliterates the developing laryngeal opening, but the lumen is normally reestablished as the vocal cords appear separately on each side. Laryngeal web, subglottic stenosis, and congenital laryngeal atresia result from different degrees of failure of the epithelium's resorption during the seventh and eight weeks of intrauterine development.

The most common web is at the glottic level and affects the vocal cords (Fig. 99-15). Other types include the posterior glottic web, causing interarytenoid vocal cord fixation (Cox and Simmons, 1974); subglottic webs, which may occur with or without cricoid cartilage involvement and subglottic stenosis; and supraglottic webs. Laryngeal atresia, a rare life-threatening anomaly, appears to represent a lack of recanalization of the embryonic larynx. Prompt diagnosis in the delivery room followed by a tracheotomy may allow the infant to survive. There is an association with tracheoesophageal fistula.

Diagnostic evaluation

Physical findings. The presenting clinical features of a laryngeal web are often multiple; the major features are an abnormality of the cry or voice, "respiratory distress", and stridor. The cry may be absent or husky. Respiratory distress includes cyanosis at birth or an unexplained airway obstruction, which requires either immediate intubation or a tracheotomy to provide an artificial airway before further assessment can be undertaken. Stridor is not often a feature. Recurrent or atypical "croup" occurs in some patients, especially those whose web are associated with subglottic stenosis. Many of the patients have major congenital anomalies of other systems, principally of the upper respiratory tract - most often subglottic stenosis. About one chance in three exists of having an associated abnormality of the respiratory tract (Benjamin, 1983). Congenital subglottic stenosis is commonly seen when the glottic web is severe.

Endoscopic examination. Most laryngeal webs are located at the level of the glottis. In all cases definitive diagnosis requires direct endoscopy to assess the web and detect any associated abnormalities of the upper respiratory tract. The larynx is examined with magnification from both straight and angled lenses because they allow better evaluation of the site, extent, and thickness of the web than can be obtained with the operating microscope. Assessing the thickness at and below the anterior commissure is essential. A large, thick web may cause life-threatening obstruction, or there may merely be a small, thin membrane at the anterior commissure. The free posterior edge of the web is thin, round, regular, concave posteriorly, and sharply outlined; the anterior part is usually thicker, depending on the size of the membranous web itself. The presence and severity of congenital subglottic stenosis can best be assessed by a combination of endoscopic examination and radiographic studies. A lateral radiograph or xeroradiogram may give valuable information about the anterior thickness of the web, the presence or absence of congenital subglottic stenosis, and the site of a subglottic web (Fig. 99-16).

Congenital interarytenoid web is a rare laryngeal anomaly whose distinctive feature is a band of tissue joining the medial surfaces of the arytenoids and restricting abduction of the vocal cords (Fig. 99-17). In addition to the interarytenoid web, which is present to some degree in all patients, associated anomalous features may include subglottic stenosis, enlarged bulky arytenoids, and difficulty exposing the larynx and maintaining the airway during anesthesia and endoscopy. Inspiratory stridor is the cardinal feature; the obstruction may be severe at birth or may worsen over weeks or months. The stridor is episodic and sometimes associated with obstructive cyanotic attacks. Diagnosis depends on direct laryngoscopy with particular attention to the posterior larynx without an endotracheal anesthesia tube in place and preferably utilizing a technique of insufflation anesthesia with spontaneous ventilation. Direct inspection of the larynx is difficult to some degree in every case and special-purpose hand-held laryngoscopes such as the Holinger pediatric anterior commissure laryngoscope or the Tucker-Benjamin infant laryngoscope and subglottiscope are helpful. They are used with rigid telescopes both straight ahead and angled so that the posterior commissure and arytenoids are deliberately separated and palpated to determine the consistency and depth of the web. The arytenoids are often large and bulky, and often a congenital subglottic stenosis is present. It seems likely that some cases have been misdiagnosed as bilateral vocal cord paralysis and others as cricoarytenoid joint fixation or posterior glottic stenosis. The association with other congenital anomalies, especially those of the respiratory tract, is high.

Management

Many forms of treatment for congenital laryngeal webs have been advocated, including dilation, which may be done deliberately (knowing that a web is present) or inadvertently (while passing an endotracheal tube or a bronchoscope); simple enoscopic microsurgical division with scissors; endoscopic division with an attempt to prevent recurrence by using sutures through the free cut edge of the web or the repeated use of dilators; endoscopic insertion of a keel; laser treatment; or laryngofissure to allow removal of redundant soft tissues in the subglottic area with or without the use of a keel.

In general, the simpler the web and the simpler the treatment required, the better the result. Thin glottic webs alone respond well to simple incision or rupture. For the remainder, obtaining a satisfactory result as judged by improvement of the voice is difficult, as is improving the airway to achieve decannulation when a tracheotomy has been performed.

Infants with congenital interarytenoid web alone usually do not require an artificial airway. Infants with congenital interarytenoid web and associated congenital subglottic stenosis or other congenital anomaly (involving dysmorphic facies or midface hypoplasia) usually need a tracheotomy, but decannulation is possible after 3 to 5 years with a very good long-term prognosis.

Posterior laryngeal cleft

A posterior cleft of the larynx is difficult to diagnose and a frequently missed anomaly (Cohen, 1975). The posterior cleft may be limited to the interarytenoid region, or it may involve the cricoid lamina. When the cleft is more extensive, the term *laryngotracheoesophageal cleft* should be used. A classification into four types has recently been proposed (Fig. 99-18; Benjamin and Inglis, 1989).

A posterior cleft of the larynx often has other associated anomalies, such as esophageal atresia, tracheoesophageal fistula, and stenosis of the trachea or bronchi.

Diagnostic evaluation

Depending on the extent of the cleft, the carinal presenting feature, especially with oral feedings, is aspiration, which causes choking and cyanosis. Although some change in the cry may occur, there is no specific characteristic. Stridor is unusual; when present, it is usually caused by tracheobronchial obstruction from aspirated secretions. There may be radiographic evidence of aspiration pneumonitis (Fig. 99-19).

Final diagnosis depends exclusively on the endoscopic examination, especially for minor interarytenoid clefts. Failure to diagnose the anomaly may result from inexperience of the endoscopist.

A laryngeal cleft should be specificially looked for at endoscopy in any patient with aspiration and stridor. A suitable laryngoscope, such as the Holinger anterior commissure pediatric laryngoscope, can be used to deliberately separate the posterior glottis and examine the posterior commissure while the patient is under general anesthesia and breathing spontaneously with no endotracheal tube in use (Fig. 99-20). Visualizing a small or large cleft can be difficult because of an unstable, collapsing larynx and redundant esophageal mucosa, especially when the cleft has not been suspected clinically. A laryngeal cleft may be found in patients with the VATER associated, with tracheoesophageal fistula, and with G syndrome.

Although the clinical features of a minor supraglottic interarytenoid cleft may suggest the presence of laryngomalacia, final definitive diagnosis can be made only by a careful examination of the larynx with particular attention to the interarytenoid region.

Management

A posterior cleft limited to the larynx might not require surgical treatment. Endogenous secretions are cleared by careful nursing. When feedings are thick, the tendency exists to aspirate and choke. With careful medical management and the child's growth and development, symptoms of the condition may completely subside. Some minor clefts with persistent aspiration despite conservative treatment can be managed by endoscopic microsurgical repair. In larger clefts, surgical repair either by the lateral pharyngotomy approach or by the laryngofissure approach has been successful.

Arthrogryposis multiplex congenita

Arthrogryposis multiplex congenita is an uncommon congenital (sometimes familial) disorder characterized by multiple joint deformities and other congenital defects, such as facial diplegia (Möbius syndrome), difficulty in swallowing, and other central nervous system abnormalities.

Cohen and Isaacs (1976) fully reviewed manifestations of arthrogryposis multiplex congenita. They described Pierre Robin-like features (micrognathia, glossoptosis, cleft palate), severe dysphagia, aspiration pneumonitis, aphonia or changes in the cry, vocal cord paralysis,

supraglottic changes similar to those of laryngomalacia, and hypertrophy of the cricopharyngeus and upper third of the esophagus.

Central nervous system dysfunction rather than simple muscle weakness causes the dysphagia and respiratory difficulties. A tracheotomy and feeding gastrostomy are part of the management, but the long-term prognosis is poor.

Cri-du-chat syndrome

Cri-du-chat syndrome (Ward et al, 1968) is a chromosomal disorder in which the newborn infant has a cry quite similar to the meowing sound of a cat. Partial deletion of the short arm of chromosome 5 exists. There is a characteristic congenital high-pitched stridor with endoscopic abnormalities seen in the larynx. The endolarynx is said to be elongated and curved with a floppy epiglottis, changes similar to laryngomalacia. On phonation the posterior part of the larynx shows an open triangle, probably caused by paralysis of the interarytenoid muscle.

Other associated abnormalities include microcephaly, hypertelorism, generalized hypotonia, severe mental retardation, and low-slung ears with a beaklike profile. In general, the prognosis is poor.

Neuromuscular dysfunction

(Plott's syndrome)

Plott (1964) described a syndrome that is a hereditary disorder with congenital laryngeal adductor paralysis as a feature. The disorder is X-linked and is associated with mental retardation. The prognosis for life is poor.

Subglottic stenosis

Congenital subglottic stenosis is the third most common congenital laryngeal anomaly after laryngomalacia and vocal cord paralysis. However, it is the most common congenital laryngeal abnormality causing serious respiratory obstruction and requiring a tracheotomy in infants.

The subglottic larynx is the region extending from the insertion of the conus elasticus into the free edge of the vocal cords above to the inferior margin of the cricoid cartilage below. By definition, congenital subglottic stenosis is considered to exist when this anatomic area has a caliber less than 4.0 mm in a full-term newborn infant. All conditions - such as neoplasms, inflammatory conditions, and acquired strictures following endotracheal intubation - are excluded from the definition. The distinction between congenital and acquired subglottic stenosis must be clear, although the latter can be superimposed on the former. The prognosis is better in purely congenital cases.

Histopathology

Holinger et al (1976b) emphasized that the more accurate the histopathologic diagnosis of subglottic stenosis, the more precise and appropriate will be the treatment. The most common abnormality is generalized circumferential stenosis of the cricoid cartilage, so that the subglottic diameter is small. However, the lumen may be eccentric, and the abnormal shape of the cricoid cartilage may be associated with a large anterior lamina, an oval or elliptic shape, a large posterior lamina, or generalized thickening. A submucosal (occult) cleft or a trapped first tracheal ring (Tucker, 1982; Tucker et al, 1979) is a rare deformity. Soft subglottic stenoses of a congenital nature may be caused by submucosal mucinous gland hyperplasia and must be distinguished from a congenital subglottic hemangioma or, less commonly, from ductal cystic disease and from acquired soft tissue subglottic narrowing caused by submucosal fibrosis and/or granulation tissue. The latter may be acquired and caused by endotracheal intubation, a complication more likely to occur in the patient who has an underlying congenital subglottic stenosis.

Diagnostic evaluation

Typical findings. The manifestations of congenital subglottic stenosis usually occur in the first few weeks or months of life and are not always evident at birth or in the neonatal period. Stridor is the most common feature, followed by respiratory distress, a harsh "croupy" barking cough, or a hoarse cry. The condition affects males twice as often as females. Having associated congenital abnormalities of the upper respiratory tract or other organs is not uncommon.

Clinically, congenital subglottic stenosis may present in the following ways:

1. "Subacute", or persistent, croup, in which the stridor lasts for 1 to 3 weeks (whereas the usual attack of acute viral laryngotracheitis lasts 1 to 3 days).

2. Recurrent atypical croup, with each attack persisting for longer than usual.

3. Difficulty introducing an endotracheal anesthetic tube of average size. Narrowing of the subglottic region is detected when the child is being anesthetized for some unrelated surgical condition.

4. Decannulation difficulty after tracheotomy or extubation difficulty after prolonged intubation.

A tight endotracheal tube without a leak of gas around the subglottic region may cause reactive mucosal edema after the tube is removed. Stridor and a croupy cough may occur minutes or hours after extubation. Congenital subglottic stenosis is often the underlying lesion in patients in whom one of the following is true: (1) difficulty removing an endotracheal tube exists after an ill-managed, prolonged intubation; or (2) tracheotomy tube extubation fails after the tracheotomy has been performed for prolonged or recurrent croup.

Endoscopic findings. A lateral airway radiographic study usually shows the congenital subglottic stenosis clearly (Fig. 99-21). However, the definitive diagnosis is made at endoscopy; the diameter of the subglottic region is assessed carefully and gently, especially if the airway is marginal. The usual finding is an isolated circumferential subglottic stenosis, the diameter of which is then measured. The outside diameter of a bronchoscope or an endotracheal tube that passes comfortably through the subglottic region represents the internal diameter of the cricoid, which is therefore calibrated and recorded for future reference. The endoscopic findings are less severe than those in patients with acquired subglottic stenosis.

Occasionally diagnostic endoscopy results in minimal but significant mucosal swelling in the subglottic region, caused by the trauma of excessive manipulation, the passage of too large a bronchoscope, or repeated manipulation. This is more significant when there is a preexisting subglottic stenosis. Subglottic edema occurs within the limits of the inexpansible cricoid cartilage in the immediate postendoscopic period. There may be croupy cough, stridor, and increasing airway obstruction. Careful technique should reduce the incidence of this complication to an absolute minimum.

Management

Management is based on the knowledge that children outgrow congenital subglottic stenosis. Most cases produce few or minimal symptoms, and no definitive treatment is required unless acute inflammation or trauma from intubation or endoscopy precipitates serious obstruction. However, when a major degree of congenital subglottic stenosis exists or when acquired fibrous tissue granulations and edema produce a serious obstruction, a tracheotomy may be require. Small patients with a stenosis severe enough to require a tracheotomy may require repeated endoscopic examination over months or years, awaiting sufficient relative improvement of the subglottic diameter to allow decannulation. This conservative management prevents the potential complications of external laryngeal surgery in the infant. On the other hand, Cotton and Evans (1981), among others, have advocated a more aggressive approach by laryngotracheal surgical reconstruction to obtain a lumen large enough to permid decannulation. Repeated "dilation treatment" of hard cartilaginous stenosis is not successful, and ill-judged forced "dilations" inevitably cause further trauma and scarring. Laser resection has a limited application and must be restricted to treatment of the soft tissue swelling in selected cases where the stenosis is less than 5 mm thick. There is no uniform management of subglottic stenosis and no universally successful endoscopic or external surgical technique. The tracheotomy tube usually should be worn for an extended period of time to allow the cartilaginous stenosis to grow along with the child's general rate of growth until the problem is overcome. Decannulation is usually possible even in difficult cases by the age of 3 to 4 years.

Laryngeal hemangioma

Hemangiomas are congenital malformations that form from mesodermal rests of vasoformative tissue. In most anatomic sites they can be managed conservatively, but narrowing of the airway is an exception. Congenital hemangioma in the larynx usually occurs in the subglottic region, developing in the submucosa and following a typical growth pattern, with increased size usually causing symptoms within the first 8 weeks or so of life. A review of autopsy cases (Brodsky et al, 1983) showed that the lesion sometimes extended into the

perichondrium and even into surrounding tissues between the tracheal rings and beyond the trachea, showing the possibility of regrowth after treatment. The more extensive lesions may require an alternate treatment plan. About half of the patients have cutaneous hemangiomas of the head, face, or neck, but no correlation exists with the site of the laryngeal lesion. All patients have some form of respiratoy distress, usually variable and fluctuant. The clinical features are more clearly defined as inspiratory stridor (intermittent at first and then persistent), harsh "barking" cough, altered cry or hoarseness, and failure to thrive. An initial erroneous diagnosis of croup is often made. A 2:1 female-to-male preponderance exists. Most cases appear by 6 months of age.

Diagnostic evaluation

Lateral radiographic study of the upper airway shows subglottic soft tissue swelling and usually clearly demonstrates a subglottic abnormality consistent with hemangioma (Fig. 99-22).

Diagnosis is made by endoscopic examination. The appearance is sufficiently characteristic for an experienced observer to make a macroscopic diagnosis without biopsy, especially if there are associated cutaneous hemangiomas. The lesion is usually localized to the subglottic region on one side, as a sessile, fairly firm, and compressible pink, red, or bluish lesion that is poorly delineated from the surrounding tissues. A biopsy may be performed when the diagnosis is uncertain, with precautions taken to maintain the airway in the unlikely event of excessive bleeding. Although infantile subglottic hemangioma has generally been regarded as a unilateral lesion, this is not always so. With modern anesthesia and sophisticated endoscopic techniques using magnification, an exact evaluation of the site and distribution of the lesion can be made. Classification of congenital laryngeal hemangiomas by anatomic site (Benjamin and Carter, 1983) includes lesions that are unilateral, bilateral, unilateral with posterior extension, and upper tracheal (Fig. 99-22).

Management

The airway obstruction is usually severe and life threatening. Some patients have acute obstruction and require immediate relief, either by a tracheotomy performed at the time of diagnosti endoscopy or by temporary intubation followed later by a tracheotomy. Most cases require a tracheotomy both for the patient's safety and to enable treatment to be performed.

Many forms of treatment have been advocated. They include tracheotomy and no active treatment, awaiting spontaneous regression; temporary, intermittent, or prolonged endotracheal tube intubation with or without steroids; injection of sclerosant; or injection of intralesional steroids for prolonged or repeated periods; excisional surgery; cryosurgery; external beam irradiation; or laser endoscopic surgery or endoscopic placement of a radioactive gold grain. Because of the relatively small numbers of patients treated and the varying attitudes to therapy, controlled studies have not been possible.

Success has been repeated with each of the various modalities of treatment outlined above, but generally, expectant treatment, steroid treatment, or laser excision is favored. Radiation therapy has previously been an acknowledged, reliable treatment used for many years to achieve resolution of the lesion and removal of the tracheotomy tube. A tracheotomy

combined with external radiation has a 93% cure rate (Brodsky et al, 1983). There are no reports of untoward effects, despite the theoretic possibility of radiation-induced malignancy of the thyroid gland. However, for this reason external radiation therapy is no longer generally favored. With placement of a radioactive gold grain in the substance of the hemangioma, the radiation dose to surrounding tissues (including the thyroid gland) is minimal, and this is often the treatment used in my institution. A unilateral lesion can usually be satisfactorily treated with the carbon dioxide laser with minimal chance of causing subglottic stenosis. The choice of treatment in an individual case may depend on the variation in anatomic exposure, surgical visibility in the subglottic region, and age of the patient. Unrecognized, untreated, or poorly treated cases have a high mortality, and active treatment, even though it may be a tracheotomy alone, is always justified.