

CHAPTER 135

Upper Airway Obstruction

ETIOLOGY

Upper airway obstruction (UAO), which is defined as blockage of any part of the airway located above the thoracic inlet, ranges from nasal obstruction due to the common cold to life-threatening obstruction of the larynx or upper trachea. In children, nasal obstruction is usually more of a nuisance than a danger because the mouth can serve as an airway, but it may be a serious problem for neonates, who breathe predominantly through the nose. The differential diagnosis of airway obstruction varies with patient age and can be subdivided into **supraglottic**, **glottic**, and **subglottic** causes (Tables 135.1, 135.2, and 135.3).

TABLE 135.1 Age-Related Differential Diagnosis of Sub-Acute Upper Airway Obstruction

NEWBORN
Choanal atresia Rhinitis neonatorum Micrognathia (Pierre Robin syndrome, Treacher Collins syndrome, DiGeorge syndrome) Macroglossia (Beckwith-Wiedemann syndrome, hypothyroidism, Pompe disease, trisomy 21) Pharyngeal collapse Laryngeal web Vocal cord paralysis/paresis (idiopathic, birth trauma or central nervous system pathology) Congenital subglottic stenosis Nasal encephalocele
INFANT
Chronic or recurrent rhinitis (infection, acid reflux, irritant) Laryngomalacia (most common) Subglottic stenosis (congenital or acquired [e.g., after intubation]) Laryngeal web or cyst Laryngeal papillomatosis Airway hemangioma Vascular rings/slings
TODDLER
Chronic or recurrent rhinitis (infection, allergy, irritant) Hypertrophied tonsils and adenoids (most common) Spasmodic croup Laryngeal papillomatosis Vascular rings/slings
OLDER CHILDREN
Chronic or recurrent rhinitis (infection, allergic, irritants) Hypertrophied tonsils and adenoids (most common) Paradoxical vocal fold movement Infectious mononucleosis

TABLE 135.2 Common Causes of Acute Upper Airway Obstruction

DIAGNOSIS	AGE	HISTORY	EXAM	IMAGING AND LABS	TREATMENT
INFECTIOUS					
Croup (<i>parainfluenza</i> and other viruses)	6 mo–3yr	Fever, URI	Nontoxic, stridor, barking cough, hoarse	Steeple sign	Aerosolized epinephrine, systemic steroids, cool mist
Epiglottitis (<i>Streptococcus pneumoniae</i> , <i>Haemophilus influenzae</i> ; respiratory viruses)	2–6yr	High fever, rapid onset, no cough, unable to swallow	Toxic, agitated, tripod sitting, drooling, stridor	Thumb sign Leukocytosis	Intubation, antibiotics
Bacterial tracheitis (<i>Staphylococcus aureus</i> , <i>Moraxella catarrhalis</i>)	Any age	High fever, rapid onset, no URI symptoms	Toxic, anxious, $\pm$ stridor, $\pm$ cough	Ragged tracheal border Leukocytosis	Intubation, antibiotics
Retropharyngeal abscess (<i>S. aureus</i> , group A strep, oral anaerobes)	<6yr	Fever, insidious onset, sore throat, no URI/cough	Moderately toxic, drooling, arched neck, inflamed pharynx	Thickened retropharyngeal space Leukocytosis	Antibiotics, surgical drainage
Peritonsillar abscess (group A strep, oral anaerobes)	>8yr	Fever, sudden worsening, sore throat, trismus	Moderately toxic, “hot potato” voice, drooling, asymmetric tonsil swelling	Imaging not needed Leukocytosis	Antibiotics, surgical drainage
NONINFECTIOUS					
Angioedema	Any age	No fever, sudden onset, urticarial, facial swelling, $\pm$ allergen exposure	Nontoxic (unless anaphylaxis), $\pm$ stridor, hoarse, facial edema	Steeple sign	Aerosolized or intramuscular epinephrine, systemic steroids, antihistamines
Spasmodic croup	6 mo–6yr	Sudden onset, no fever/URI, recurrent, often nocturnal	Nontoxic, $\pm$ stridor, hoarse, barking cough	Often normal	Aerosolized epinephrine, antihistamines, antacids, systemic steroids
Foreign body	6 mo–5yr	Sudden onset, cough and choke	Nontoxic, anxious, stridor, aphonic, brassy cough	Radiopaque object may be seen	Rigid bronchoscopy

URI, Upper respiratory tract infection (with coryza, sneezing).

TABLE 135.3 Differentiating Supraglottic from Subglottic Causes of Acute Airway Obstruction

FEATURE	SUPRAGLOTTIC OBSTRUCTION	SUBGLOTTIC OBSTRUCTION
Common clinical syndromes	Epiglottitis, peritonsillar and retropharyngeal abscess	Croup, angioedema, foreign body, bacterial tracheitis
Stridor	Quiet	Loud
Voice	Muffled	Hoarse
Dysphagia	Yes	No
Tripod or arching posture	Yes	No
Barking cough	No	Yes
Toxic	Yes	No, unless tracheitis
Trismus	Some	No
Drooling	Yes	No

Modified from Davis H, Gartner JC, Galvis AG, et al. Acute upper airway obstruction: Croup and epiglottitis. *Pediatr Clin North Am.* 1981;28:859–880.

CLINICAL MANIFESTATIONS

UAO is more pronounced during inspiration because the negative pressure generated collapses the upper airway, increasing resistance and turbulent airflow that creates an inspiratory

noise. Children with UAO may have increased inspiratory work of breathing manifested by **suprasternal retractions**. The respiratory noise most commonly associated with UAO is **stridor**, a harsh sound caused by the vibration of the airway

structures. Stridor often decreases during sleep because of lower inspiratory flow rates, and increases during feeding, excitement, and agitation because of higher flow rates. Occasionally stridor may also be present on exhalation. Hoarseness or aphonia with stridor suggests **vocal cord** involvement. **Stertor** is a low-pitched sound like a snore and suggests implosion of pharyngeal soft tissue structures.

DIAGNOSTIC STUDIES

Radiographic evaluation of a child with stridor is rarely helpful. On anteroposterior (AP) views of the neck taken with the head in extension, the subglottic space should be symmetrical with diameter continuing to increase down to the midtrachea. Asymmetry suggests subglottic stenosis or a mass lesion, whereas tapering suggests subglottic edema. However, these findings may be subtle. In the child with stertor or snoring, lateral views of the neck and nasopharynx may provide information about tonsil or adenoid hypertrophy. Computed tomography (CT) scans of the upper airway can help delineate the site of the obstruction when soft tissue mass (e.g., tonsillar abscess) is suspected, but may require sedation in younger children. Flexible nasopharyngoscopy/laryngoscopy, which can be done without sedation, is extremely useful in assessing airway patency, the presence of adenoid tissue or vocal cord and other airway lesions, and laryngomalacia.

DIFFERENTIAL DIAGNOSIS

Choanal Stenosis (Atresia)

Choanal stenosis/atresia is a congenital problem presenting in the neonatal period. It may be bilateral or unilateral and is relatively rare. Neonates are generally obligate nose breathers, so obstruction of nasal passages can cause significant respiratory distress, especially when feeding. Crying bypasses the obstruction because crying infants breathe through the mouth. Inability to easily pass a small catheter through the nostrils should raise the suspicion of choanal atresia. The diagnosis is confirmed by CT scan and by inspecting the area directly with a flexible nasopharyngoscope. An oral airway may be useful in the short term, but the definitive treatment is surgery.

Croup (Laryngotracheobronchitis)

See Chapter 107.

Epiglottitis

See Chapter 107.

Bacterial Tracheitis

See Chapter 107.

Laryngomalacia

Etiology

Laryngomalacia is due to exaggerated collapse of the glottic structures, especially the epiglottis and arytenoid cartilages, during inspiration. It is the most common cause of stridor in infants. It may be due to decreased muscular tone of the larynx and surrounding structures or immature cartilaginous

structures. It usually does not result in significant respiratory distress, but occasionally it is severe enough to cause apnea, hypoventilation, hypoxemia, and difficulty feeding.

Clinical Manifestations

The primary sign of laryngomalacia is **inspiratory stridor** with little or no expiratory component. The stridor is typically loudest when the infant is feeding or active and decreases when the infant is relaxed or placed prone, or when the neck is flexed. Any condition that increases upper airway inflammation will exacerbate laryngomalacia, including viral respiratory infections, dysphagia (swallowing dysfunction), and gastroesophageal reflux. Laryngomalacia normally peaks by 3–5 months of age and resolves between 6 and 12 months of age. However, occasionally it can persist in otherwise normal children up until 24 months of age, and even longer in children with underlying conditions, especially those with neurologic diseases affecting control of upper airway muscles (such as cerebral palsy).

Diagnostic Studies

In many infants with presumed laryngomalacia, the diagnosis can be tentatively established by history and physical examination. If the patient follows the typical course for laryngomalacia, then no further work-up is necessary. However, to firmly establish the diagnosis, which is important in more severe or atypical cases, the patient should undergo flexible nasopharyngoscopy to assess the patency and dynamic movement (collapse) of the larynx and surrounding structures. This procedure can also identify vocal cord abnormalities and airway lesions above the vocal cords.

Treatment

In most cases, no therapy is required for laryngomalacia. The infant should be observed closely during times of respiratory infection for evidence of respiratory compromise. Medical management for moderate to severe laryngomalacia can include acid suppression for those infants with suspected gastroesophageal reflux, feeding therapy, and optimization of nutritional status. Infants with severe laryngomalacia resulting in hypoventilation, hypoxia, or growth failure may benefit from a surgical procedure (e.g., supraglottoplasty) or, in extreme cases, a tracheostomy to bypass the upper airway.

Subglottic Stenosis

Etiology

Subglottic stenosis is the narrowing of the portion of the trachea immediately below the vocal cords. It may be congenital but more often is acquired. Endotracheal intubation, especially prolonged or repeated intubation required in some premature infants, can lead to inflammation and scarring of the subglottic space.

Clinical Manifestations

Subglottic stenosis can present as stridor that is **biphasic** (on both inspiration and expiration) but usually more prominent on inspiration. With increasing respiratory effort, the stridor becomes louder. Very small infants may not be able to breathe with enough force to generate a sound. Subglottic stenosis may also be associated with a barking cough similar to that noted with croup. Respiratory infections can cause subglottic edema, exacerbating the clinical manifestations of subglottic stenosis.

Diagnostic Studies

Definitive diagnosis requires endoscopic evaluation, either by flexible or rigid bronchoscopy.

Treatment

Mild subglottic stenosis can be managed conservatively and may improve sufficiently with airway growth alone. More severe cases require surgical intervention. Depending on the nature of the lesion, endoscopic laser treatment may be effective. Other surgical options include tracheoplasty and cricoid split procedures. A tracheostomy tube may be required to bypass the subglottic space until the airway is patent enough to allow adequate airflow.

Mass Lesions

Upper airway mass lesions are relatively uncommon. The most common laryngeal tumor in childhood is the **hemangioma**, which usually presents before 6 months of age and is often able to be treated medically (see [Chapter 194](#)). Definitive diagnosis requires endoscopy. If the obstruction is severe, a tracheostomy tube may be needed until the lesion spontaneously involutes or improves with therapy.

Laryngeal webs are either the result of failed recanalization of the glottic airway in utero or secondary to airway trauma (intubation, traumatic injury, prior surgical manipulation), whereas **laryngeal cysts** typically occur as a consequence of airway trauma. Laryngeal webs can present during infancy with a weak or high-pitched cry with respiratory distress. Both laryngeal webs and laryngeal cysts can produce biphasic stridor and are best identified by bronchoscopy. **Foreign body** should be considered in any infant or child capable of ingesting small objects who develops acute onset of stridor.

Juvenile laryngeal papillomatosis is a rare condition of benign tumors caused by human papillomavirus 6 (HPV-6) and HPV-11 acquired at birth from maternal genital warts. Clinical manifestations usually start in infancy and include biphasic stridor and hoarse voice/cry. The lesions are most commonly located in the larynx, but they can spread distally into the trachea, large bronchi, and even to the lung parenchyma. Treatment options, which are limited and rarely curative, include laser therapy and interferon. Tracheostomy may be required to ensure an adequate airway but should be avoided if possible as there is risk of seeding of the distal airways with tumor.

Vocal Cord Paralysis

Etiology

Vocal cord paralysis is an important cause of laryngeal dysfunction. Paralysis may be unilateral or bilateral and is more often caused by damage to the recurrent laryngeal nerve than by a central lesion. The left recurrent laryngeal nerve passes around the aortic arch and is more susceptible to damage than the right laryngeal nerve. Peripheral nerve injury may be caused by trauma (neck traction during delivery of infants or thoracic surgical procedures) and mediastinal lesions. Central causes include Arnold-Chiari malformation (meningomyelocele), hydrocephalus, and intracranial hemorrhage.

Clinical Manifestations

Vocal cord paralysis presents as biphasic stridor and alterations in voice and cry, including a weak cry (in infants), hoarseness, and aphonia. Children with vocal cord paralysis are at risk for

aspiration, often manifested as coughing/choking with drinking and coarse airway sounds audibly and by auscultation.

Treatment and Prognosis

Patients with traumatic injury to the recurrent laryngeal nerve often have spontaneous improvement over time, usually within 3–6 months. If the paralyzed vocal cord has not recovered within 1 year of the injury, then it is likely to be permanently damaged. In some cases, injection of biocompatible material into a paralyzed vocal cord can reposition the cord to improve phonation and airway protection. Patients with vocal cord paralysis resulting in severe airway obstruction and aspiration may require tracheostomy tube placement. Patients with vocal cord paralysis are at higher risk for descending aspiration and should be evaluated by a videofluoroscopic swallow study (VFSS) to assess safety for oral feeding.

Adenoidal and Tonsillar Hypertrophy

Etiology

The most common cause of chronic UAO in children is hypertrophy of the adenoids and tonsils. Adenoidal and tonsillar hyperplasia may be aggravated by recurrent infection, allergy, and inhaled irritants.

Clinical Manifestations

The signs of adenoidal and tonsillar hypertrophy are mouth breathing, snoring, and, in some patients, obstructive sleep apnea (see [Chapter 134](#)). The eustachian tubes enter the nasopharynx at the choanae and can be obstructed by enlarged adenoids, predisposing to recurrent or persistent otitis media.

Diagnostic Studies

Adenoidal hypertrophy is assessed by a lateral radiograph of the nasopharynx or by flexible nasopharyngoscopy.

Treatment

If the adenoids or tonsils are large and thought to be significantly contributing to UAO, then the most effective treatment is removal. Because the adenoids are not a discrete organ but rather consist of lymphoid tissue, regrowth is possible, especially in preschool children. Refractory OSA is less common after adenotonsillectomy than adenoidectomy or tonsillectomy alone.

PEARLS FOR PRACTITIONERS

See *The Respiratory System: Pearls for Practitioners* at the end of this section.

CHAPTER 136

Lower Airway, Parenchymal, and Pulmonary Vascular Diseases

ETIOLOGY

There are many causes of lower airway diseases (Table 136.1). Lower airway diseases often result in airway obstruction. The most common lower airway disease in children is **asthma**, which results in variable and recurring symptoms, diffuse

TABLE 136.1 Causes of Wheezing in Childhood

ACUTE
Asthma
Exercise-induced asthma
Hypersensitivity reactions
Infection (e.g., bronchiolitis)
Inhalation of irritant gases or particulates
Descending aspiration
Foreign body (airway or esophageal)
Aspiration of gastric contents (ascending aspiration)
CHRONIC OR RECURRENT
Asthma (see under acute)
Hypersensitivity reactions, allergic bronchopulmonary aspergillosis (seen only in children with either asthma or cystic fibrosis)
Vocal cord dysfunction
Recurrent aspiration/swallowing dysfunction (dysphagia)
Retained foreign body
Gastroesophageal reflux
Bronchopulmonary dysplasia
Bronchitis, bronchiectasis
Cystic fibrosis
Primary ciliary dyskinesia
Bronchiolitis obliterans
Pulmonary edema (congestive heart failure)
Bronchomalacia/tracheomalacia
Vascular ring
Pulmonary artery dilatation (absent pulmonary valve)
Bronchial or pulmonary cysts/masses
Lymph nodes (tuberculosis, lymphoma)
Endobronchial masses/tumors (carcinoid)
Bronchial or tracheal stenosis
Cardiomegaly

bronchial obstruction from airway inflammation, and hyperresponsiveness of bronchial smooth muscle leading to airway constriction. Virus-induced wheezing episodes are common, especially in children under 3 years of age. Wheezing that is localized to one area of the chest suggests focal airway obstruction (foreign body aspiration or extrinsic compression).

CLINICAL MANIFESTATIONS

A wheeze is a continuous sound that is produced by vibration of airway walls and generally has a more musical quality than does stridor. In contrast to upper airway obstruction, obstruction below the thoracic inlet causes more wheezing on expiration than on inspiration. Intrathoracic pressure is increased relative to atmospheric pressure during exhalation, which collapses the intrathoracic airways and accentuates airway narrowing on expiration. Expiratory airflow through obstructed intrathoracic airways leads to wheeze, prolonged expiratory phase, and increased abdominal work of breathing. In patients with chronic airway infection (e.g., cystic fibrosis), the bronchi become permanently damaged and dilated (bronchiectasis). Patients with bronchiectasis have episodes of cough, often productive of purulent sputum, and may have inspiratory crackles.

DIAGNOSTIC STUDIES

When asthma is suspected, empiric trials of therapy (bronchodilators, short courses of oral corticosteroids, long-term use of inhaled corticosteroids) are useful in arriving at a diagnosis. In children older than 6 years, pulmonary function tests (spirometry) can assess airflow obstruction and response to bronchodilators. **Radiographic evaluation** is not needed with each episode of wheezing, but those with significant respiratory distress, fever, history consistent with foreign body aspiration, or focal auscultatory findings should have posteroanterior (PA) and lateral chest radiographs obtained. Generalized **hyperinflation**, indicated by flattening of the diaphragms and an increased anteroposterior (AP) chest diameter, suggests diffuse obstruction of the small airways. Localized hyperinflation, especially on expiratory views, suggests localized bronchial obstruction (foreign body or an anatomic anomaly). Dysphagia leading to aspiration and airway inflammation can present with persistent wheezing. This is best assessed with a **videofluoroscopic swallowing study**. Gastroesophageal reflux may aggravate asthma and may lead to wheezing in very young children, especially if it is associated with aspiration. Infants with persistent wheezing despite empiric treatment with asthma therapy may benefit from an airway evaluation with **flexible bronchoscopy**.

DIFFERENTIAL DIAGNOSIS

Asthma

See Chapter 78.

Tracheomalacia

Etiology

Tracheomalacia is a floppy trachea due to lack of structural integrity of the tracheal wall (Fig. 136.1). The tracheal cartilaginous rings normally extend through an arc of approximately 320 degrees, maintaining rigidity of the trachea during changes in intrathoracic pressure. With tracheomalacia, the cartilaginous rings may not extend as far around the circumference (leaving the membranous posterior trachea wider than usual), may be completely absent, or may be present but damaged. These abnormalities can result in excessive collapse of the trachea, most pronounced during expiration. Tracheomalacia is the most common congenital tracheal abnormality and may be congenital (tracheoesophageal fistula or bony dysplasia syndromes) or acquired (long-term mechanical ventilation). Tracheomalacia must be differentiated from extrinsic tracheal compression by masses or vascular structures. Localized

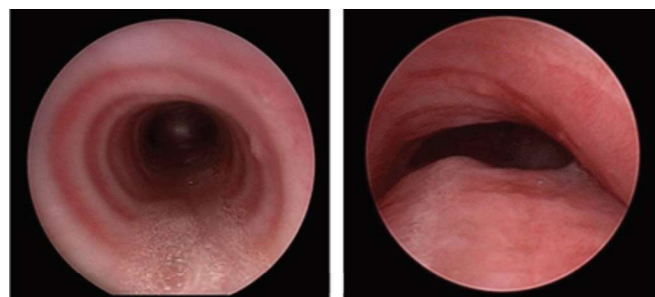


FIGURE 136.1 Normal tracheal anatomy (A) and tracheomalacia (B) showing loss of anterior tracheal ring integrity and collapse of the tracheal posterior wall membrane. (Courtesy Anne & Robert H. Lurie Children's Hospital of Chicago.)

tracheomalacia may persist after the trachea has been relieved of extrinsic compression.

Clinical Manifestations

With tracheomalacia, the tracheal collapse may only be apparent during forced exhalation or with cough. It is commonly aggravated by respiratory infections. The airway collapse causes a harsh, monophonic wheeze compared to the polyphonic wheeze more often seen in asthma. Secretions may be retained behind the segment of malacia, predisposing to infection. Infants with severe tracheomalacia may completely collapse the trachea during agitation, resulting in cyanotic episodes that resemble breath holding spells. The voice is normal, as is inspiratory effort. In older children, the hallmark sign is a brassy, barking cough due to the vibration of the tracheal walls. The expiratory noises of tracheomalacia are often mistakenly ascribed to asthma or bronchiolitis, and the barking cough is often misdiagnosed as croup.

Treatment

Infants with mild to moderate tracheomalacia usually require no intervention. Tracheomalacia improves with airway growth as the lumen increases in diameter and the tracheal wall becomes more firm. The treatment of older symptomatic children is geared toward treating the precipitating cause for cough and providing supportive care. Chest physiotherapy is essential to improve mucociliary clearance, and antibiotics are often necessary to treat concurrent infection.

Children, especially infants, with severe tracheomalacia may require tracheostomy tubes to administer continuous positive airway pressure (CPAP), which serves to stent open the airway. Custom-length tracheostomy tubes are used to bypass the site of collapse. Surgical techniques such as slide tracheoplasty may benefit short segments of severe malacia. Aortopexy has been used to limit anterior collapse but does not treat posterior tracheal membrane collapse. Historically, surgically placed airway stents have been problematic in children because airway stents may erode and migrate, do not grow with children, and serve as a source of fixed stenosis and obstruction. Newer absorbable airway external splints and internal stents with three-dimensional printing technology are under active investigation for treatment of severe pediatric tracheomalacia.

Tracheoesophageal Fistula

See Chapter 128.

Extrinsic Tracheal Compression

Compression of the trachea by vascular structures or masses can cause significant respiratory compromise. Tracheal compression by aberrant great vessels (aorta, innominate artery) may cause wheezing, stridor, cough, and dyspnea. Incomplete vascular rings are often asymptomatic and incidentally found, but anterior tracheal compression can occur from an **anomalous innominate artery** that arises more distally than normal from the aortic arch. This usually results in mild respiratory symptoms. Surgical correction is rarely necessary. Complete vascular rings, which compress the esophagus posteriorly and the trachea anteriorly, include the double aortic arch and a right aortic arch with a persistent ligamentum arteriosum (most common). Both lesions have right-sided aortic arches, which may be visible on chest radiograph. In addition to respiratory symptoms, complete vascular rings

may cause dysphagia as a result of esophageal compression. The diagnosis of vascular anomalies can often be made by a barium swallow, which identifies the esophageal compression (Fig. 136.2). Bronchoscopy will identify a pulsatile compression of the airway, but the diagnostic procedures of choice are a computed tomography (CT) angiogram of the chest and great vessels or magnetic resonance angiogram (MRA). Definitive treatment of vascular rings requires surgical repair. Other causes of extrinsic tracheal compression include enlarged mediastinal lymph nodes (tuberculosis), mediastinal masses (teratoma, lymphoma, thymoma, germ cell tumors), and, rarely, cystic hygromas.

Foreign Body Aspiration

Epidemiology

Aspiration of foreign bodies into the trachea and bronchi is relatively common. The majority of children who aspirate foreign bodies are under 3 years of age. Patients with developmental delay or with older siblings are at increased risk. Because the right mainstem bronchus takes off at a less acute angle than the left mainstem bronchus, foreign bodies tend to lodge in right-sided airways. Some foreign bodies, especially nuts, can also lodge more proximally in the larynx or subglottic space, totally occluding the airway. Many foreign bodies are not radiopaque, which makes them difficult to detect radiographically. The most common foreign bodies aspirated by young children are food (especially nuts) and small toys. In younger children, coins and button batteries more often lodge in the esophagus than in the airways. Button batteries can cause esophageal erosion and be life threatening. Older children have been known to aspirate rubber balloons, which also can be life threatening.



FIGURE 136.2 Vascular ring. Indentation of the posterior esophagus seen on barium swallow, lateral view. This child had a double aortic arch.

Clinical Manifestations

Many children who aspirate foreign bodies have clear histories of choking, witnessed aspiration, or physical or radiographic evidence of foreign body aspiration. However, a proportion of patients have a negative history because the aspiration went unrecognized. Physical findings observed with acute foreign body aspiration may include cough, localized wheezing, unilateral absence of breath sounds, stridor, and, rarely, bloody sputum.

Most foreign bodies are small and quickly expelled, but some may remain in the lung for long periods of time and may come to medical attention because of persistent cough, sputum production, or recurrent unilateral pneumonia. Foreign body aspiration should be in the differential diagnosis of patients with persistent wheezing unresponsive to bronchodilator therapy, persistent atelectasis, recurrent or persistent pneumonia, or chronic cough without another explanation. Foreign bodies may also lodge in the esophagus and compress the trachea, thus producing respiratory symptoms. Therefore esophageal foreign bodies should be included in the differential diagnosis of infants or young children with persistent cough, stridor, or wheezing, particularly if dysphagia is present.

Diagnostic Studies

Radiographic studies will reveal the presence of radiopaque objects and can also identify focal air trapping, especially on expiratory views or decubitus films. Many foreign bodies are not radiopaque. Thus, when foreign body aspiration is suspected, expiratory or lateral decubitus chest radiographs may identify air trapping on the affected, dependent side. If history or exam is suggestive of foreign body aspiration, the patient should undergo bronchoscopy. Flexible bronchoscopy can be used to locate an aspirated foreign body and may be useful when the presentation is not straightforward, but foreign body removal is best performed via rigid bronchoscopy.

Prevention

Common foods, including peanuts or other nuts, popcorn, uncooked carrots, or any foods difficult to break into small pieces, are at risk to be aspirated by infants and children, particularly before molar teeth have erupted. Older siblings should be counseled to keep small toys separate and not present younger children with small parts.

Bronchiolitis

See Chapter 109.

Bronchopulmonary Dysplasia

See Chapter 61.

Congenital Lung Anomalies

Congenital lobar emphysema (congenital lobar overinflation) consists of hyperinflation of one or more lobes of the lung. This overdistention may cause severe respiratory distress in the neonatal period due to compression of surrounding normal lung tissue, but it can also be asymptomatic and remain undiagnosed for years. Radiographically, it may be mistaken for a pneumothorax. Lobectomy may be required if respiratory

distress is severe or progressive, but if the patient is asymptomatic then surgical resection may not be indicated.

Congenital pulmonary airway malformations (CPAMs) are a spectrum of lesions arising during early lung development with histologic appearance ranging from simplified bronchial to complex cystic alveolar tissue, connected to the tracheobronchial tree. They are often first identified on antenatal ultrasound, and after birth can be inconspicuous on chest radiograph. CPAMs are nonfunctioning lung tissue and can have hybrid features of **pulmonary sequestration** (presence of systemic circulation connections). Although CPAM lesions can be stand-alone lesions, a substantial proportion of pulmonary sequestrations are hybrid CPAM lesions. Sequestrations without hybrid features are separate from the tracheobronchial tree and are classified as intrapulmonary (located in a normal lobe without its own pleura) and extrapulmonary (outside normal lung). CPAMs and sequestrations can coexist with other chest lesions including **congenital diaphragmatic hernia**. Sequestration is also associated with **Scimitar syndrome** (partial anomalous pulmonary venous return). Symptomatic neonates with respiratory distress, large lesions, or family history of features concerning for malignancy require further imaging. Although large malformations can cause neonatal respiratory distress, many are asymptomatic. Chest CT with contrast is the diagnostic test of choice (Fig. 136.3) but may be delayed if the child is asymptomatic with close clinical follow-up for symptom development. Surgical resection is curative, but risk of infection or malignancy is not completely understood in these children. Conservative management with observation is a reasonable option for families with low risk.

Pulmonary hypoplasia, or a relative decrease in number and size of alveoli, may be the result of bronchopulmonary dysplasia or due to congenital differences such as congenital heart disease, trisomy 21, **congenital diaphragmatic hernia** (see Chapter 61), or unilateral congenital absence of a pulmonary artery. Alveolar growth occurs after birth, predominantly up to 3 years, but has been documented beyond 8 years of age.

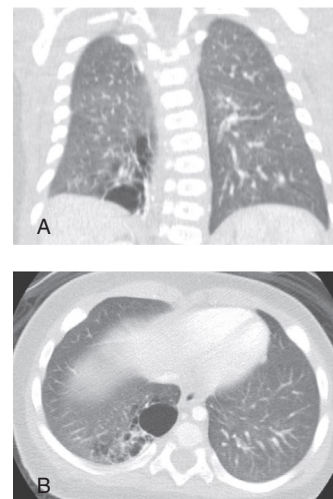


FIGURE 136.3 Congenital pulmonary airway malformation. Coronal (A) and axial (B) computed tomography images of a right lower lobe congenital pulmonary airway malformation in a child who presented with recurrent pneumonias. The entire left lung and upper and middle lobes of the right lung are normal.

Primary Ciliary Dyskinesia

Etiology

Primary ciliary dyskinesia (PCD) is a heterogeneous, usually recessive genetic disorder of motile cilia and is characterized by a diverse array of clinical features, including chronic/recurrent upper and lower respiratory tract infections, laterality defects (including *situs inversus totalis* [SIT] and heterotaxy), and male infertility. This disorder affects approximately 1 in 15,000–20,000 live births, although the actual incidence may be higher because of a limited ability to definitively diagnose affected patients.

Clinical Manifestations

The clinical features of PCD reflect defective motile cilia function in the respiratory tract, sinuses, middle ear, and reproductive tract, as well as laterality defects. Early pediatric PCD clinical features include (1) history of unexplained neonatal respiratory distress, with supplemental oxygen requirement for more than 24 hours in term infants, (2) early-onset, year-round wet cough, (3) early onset, year-round nasal congestion, and (4) laterality defects. Patients develop progressive bronchiectasis that is universal by adulthood and can lead to end-stage lung disease. Cilia dysfunction in embryonic nodal cells can result in laterality defects and congenital heart disease. Other clinical manifestations include impaired growth, recurrent middle ear effusions with associated hearing loss, chronic sinusitis, nasal polyps, male infertility, and decreased female fertility.

Genotype-phenotype relationships are emerging in PCD, particularly regarding variability in severity and progression of lung disease. Patients with a distinct ultrastructural defect (inner dynein arm defect with microtubular disorganization), which is usually linked to mutations in *CCDC39* and *CCDC40*, typically present earlier in life, have worse pulmonary function by both functional and structural assessment (spirometry and CT scans), and poorer nutritional status compared to similar-aged patients with ODA defects.

Diagnostic Studies

PCD diagnosis has historically relied on a combination of clinical features and ultrastructural analysis of respiratory cilia by electron microscopy, obtained from scrapings/biopsy of nasal or airway epithelium. Results may be difficult to interpret as chronic infection and inflammation may also lead to ultrastructural abnormalities in nasal cilia. The most common ciliary ultrastructural defect is the absence of outer dynein arms (ODA). However, a third of affected individuals have normal ciliary ultrastructure. The measurement of nasal nitric oxide is the most sensitive and specific diagnostic test for PCD, and a low nasal nitric oxide level (<77 nL/min) is consistent with PCD. PCD is usually autosomal recessive; however, it has been associated with X-linked cri du chat syndrome and other overlapping ciliopathies. Currently 50 genes have been identified to explain approximately 75% of cases. High-speed videomicroscopy assesses ciliary beat frequency and pattern but requires specialized equipment, and, currently, there is an absence of standardized reporting methods.

Treatment

Treatment is geared toward treating infections and improving clearance of respiratory secretions. High-resolution chest CT scans are useful to confirm and monitor bronchiectasis.

Surveillance cultures help identify organisms involved and guide antibiotic therapy. Chest physiotherapy and prompt treatment of bacterial infections are helpful, but the course of the lower airway disease tends to be slowly progressive. Inhaled hypertonic saline improves cough clearance and has been shown to improve lung function. Antiinflammatory antibiotics, such as low-dose macrolides, may reduce the number of exacerbations per year. Sinus surgical procedures are often done to manage chronic sinusitis, but their benefit is questionable. Most children require placement of pressure equalization (PE) tubes for management of recurrent otitis media. Multidisciplinary care with pulmonology, otolaryngology, audiology, respiratory therapy, and nutrition is recommended.

Pneumonia

See Chapter 110.

Pulmonary Edema

Etiology

Pulmonary edema is the seepage of fluid into the alveolar and interstitial spaces. Capillary hydrostatic forces and interstitial osmotic pressures tend to push fluid into the air spaces, whereas plasma osmotic pressures and tissue mechanical forces tend to move fluid away from the air spaces. Under normal circumstances, the sum of these forces favors absorption, so the alveolar and interstitial spaces remain dry. Fluid entering the alveoli is normally removed by pulmonary lymphatics. Pulmonary edema forms when transcapillary fluid flux exceeds lymphatic drainage. Reduced left ventricular function leads to pulmonary venous hypertension and increased capillary hydrostatic pressure, and fluid moves into the interstitial space and alveoli. Fluid initially enters the interstitial space around the terminal bronchioles, alveoli, and arterioles (**interstitial edema**), causing increased lung stiffness and premature closure of bronchioles on expiration. If the process continues, fluid then enters the alveoli, further reducing compliance and resulting in **intrapulmonary shunting** (alveolar units that are perfused but not ventilated). Decreased ventilation (hypercarbia) is a late finding.

Pulmonary edema is most commonly due to heart failure from left ventricular or biventricular dysfunction. Pulmonary hypertension (PH) and associated cor pulmonale (right ventricular dysfunction) do not usually cause pulmonary edema, as the increased vascular resistance is proximal to the capillary bed. Increased capillary permeability, seen in disease states such as sepsis and acute respiratory distress syndrome, can lead to pulmonary edema. Pulmonary edema may also occur with excessive swings in intrathoracic pressure, as seen after tracheal foreign body aspiration or severe obstruction from hypertrophied tonsils and adenoids (**postobstructive pulmonary edema**). Pulmonary edema may also be present in conditions with decreased serum oncotic pressure (hypoalbuminemia); after administration of large volumes of intravenous (IV) fluids, especially if there is capillary injury; with ascent to high altitude (**high-altitude pulmonary edema**); and after central nervous system injury (**neurogenic pulmonary edema**).

Clinical Manifestations

The clinical manifestations of pulmonary edema are **dyspnea**, tachypnea, and cough (can occur with frothy, pink-tinged

sputum). As the edema worsens, there is increased work of breathing and hypoxemia; diffuse inspiratory crackles can be heard on auscultation.

Diagnostic Studies

Chest radiographs may reveal diffuse hazy infiltrates, classically in a perihilar pattern, but these findings may be obscured by underlying lung disease. Interstitial edema (**Kerley B lines**) may be seen, especially at the lung bases.

Treatment and Prognosis

Treatment and prognosis depend on the cause of the pulmonary edema and response to therapy. Patients should be positioned in an upright posture and given supplemental O₂. Diuretic therapy and rapidly acting IV inotropic agents may be helpful in cardiogenic pulmonary edema. CPAP or intubation with positive pressure ventilation using high positive end-expiratory pressures (PEEP) may be required.

Acute Respiratory Distress Syndrome

See Chapter 39.

Pulmonary Arterial Hypertension and Cor Pulmonale

Etiology

PH is defined as a mean pulmonary artery pressure of 20 mm Hg or higher in children over 3 months of age at sea level. Pediatric PH is categorized into five groups based on the World Health Organization classification, similar to adult PH. Group 1 is pulmonary arterial hypertension (PAH) due to diseases within veins and small pulmonary muscular arteries such as persistent PH of the newborn, congenital heart disease (including those with excessive pulmonary blood flow due to left-to-right cardiac shunting), exposure to various drugs (e.g., cocaine, amphetamines), and **idiopathic pulmonary arterial hypertension** (IPAH, previously called primary PH). Hereditary IPAH is also in this group and has been linked with the bone morphogenetic receptor-2 (*BMPR2*) gene. Group 2 is caused by dysfunction of the left heart such as valvular disease. Group 3 is secondary to lung disease or chronic hypoxia, including upper airway obstruction resulting in obstructive sleep apnea and hypoxemia. In Group 4, PH is secondary to chronic pulmonary thromboembolism. Group 5 includes PH of unclear but multifactorial diseases, including some autoimmune diseases and metabolic diseases. The Panama classification is a new pediatric-specific system that includes 10 categories to capture the heterogeneity of pulmonary hypertensive vascular disease from the fetus to adolescent (e.g., perinatal pulmonary vascular maladaptation, multifactorial pulmonary hypertensive vascular disease in congenital malformation syndromes, diffuse pediatric lung disease). This classification system is promising but has yet to be tested and validated for utility and accuracy.

Prolonged PH may lead to irreversible changes in the intima and media of the pulmonary arterioles. PH strains the right side of the heart, which leads to hypertrophy and dilation of the right ventricle. Altered structure and/or function of the right heart from PH is known as **cor pulmonale**. The most common causes of cor pulmonale in children are chronic lung diseases, especially severe bronchopulmonary dysplasia

and severe, untreated obstructive sleep apnea (with chronic hypoxia).

Clinical Manifestations and Diagnostic Studies

PAH should be suspected with developmental anatomic abnormalities or historical exposures (e.g., congenital heart disease, bronchopulmonary dysplasia, congenital diaphragmatic hernia, prolonged hypoxemia). Exertional dyspnea and unexplained syncope are presenting symptoms. In addition to the other physical findings associated with pulmonary and cardiac diseases, an accentuated pulmonary component of the second heart sound may be heard. Definitive diagnosis is made by cardiac catheterization, but echocardiography may confirm the presence of right ventricular hypertrophy, ventricular dysfunction, interventricular septal flattening, and tricuspid insufficiency, which can be used to estimate the pulmonary artery pressures. Cardiac magnetic resonance imaging can be useful in diagnosis and to assess changes in ventricular function and size. Lung biopsy may be considered for children with PAH suspected of diffuse lung disease, pulmonary venoocclusive disease, or vasculitis.

Treatment and Prognosis

Treatment should focus on the underlying condition of known causes of PH, such as lung volume optimization in persistent PH of the newborn, or therapy with diuretics and salt/fluid restriction in heart failure. In all conditions of PH and PAH, the relief of hypoxemia with supplemental O₂ therapy can be therapeutic as a pulmonary vasodilator. Medications for pulmonary vasodilation such as short-term inhaled nitric oxide may be helpful in some patients. Continuous infusions of prostacyclin may be helpful, both acutely and chronically, and is available in inhaled, subcutaneous, and IV forms. Endothelium antagonists may be beneficial. Unfortunately, many patients with the idiopathic form of PAH have progressive courses, and lung or heart-lung transplantation may be the only treatment option.

Pulmonary Hemorrhage

Etiology

Pulmonary hemorrhage is a rare but potentially life-threatening condition in children. It can be due to bleeding from the airways (hemangiomas, bronchial vessel bleeds) or from diffuse capillary bleeding (alveolar hemorrhage). Alveolar hemorrhage is usually due to diffuse capillary disruption/inflammation.

Clinical Manifestations

The presentation of pulmonary hemorrhage is variable with a spectrum of asymptomatic radiographic abnormalities to life-threatening respiratory failure. **Hemoptysis**, although experienced by most children with pulmonary hemorrhage, can be absent in a third of individuals. It is important to rule out extrapulmonary sources of bleeding, including hematemesis and bleeding from the nasopharynx or mouth, as these are more common than true pulmonary hemorrhage. In addition to hemoptysis, the presenting signs and symptoms of pulmonary hemorrhage include cough, wheeze, shortness of breath, pallor, fatigue, cyanosis, and fever. Episodic pulmonary hemorrhage frequently manifests as recurrent respiratory symptoms associated with pulmonary infiltrates on chest radiographs.

Symptomatic **airway hemorrhage** may result in significant hemoptysis with few radiographic changes, whereas **alveolar bleeding** often causes profound respiratory symptoms, hypoxemia, diffuse infiltrates on radiographs, and minimal hemoptysis. Some patients experience a localized bubbling sensation in the chest, which may be helpful in differentiating local from diffuse sources of pulmonary bleeding. Physical examination findings may include locally or diffusely decreased breath sounds, cyanosis, and crackles on auscultation.

The **differential diagnosis** of pulmonary hemorrhage includes alveolar and airway bleeding. Airway bleeding can be due to airway hemangiomas, pulmonary arteriovenous malformations (e.g., hereditary hemorrhagic telangiectasia), and bronchial artery collaterals, which develop in some patients with chronic lung infections (e.g., cystic fibrosis). The causes of alveolar (capillary) bleeding can be divided into immune-mediated and non-immune-mediated categories. Immune-mediated causes include antineutrophil cytoplasmic autoantibodies (ANCA)—associated vasculitides, isolated pulmonary capillaritis, antiglomerular basement membrane disease, drug-induced vasculitis, and other connective tissue diseases. Nonimmune mediated causes include idiopathic pulmonary hemosiderosis, clotting disorders, infection, inhalation injury, and cardiac conditions associated with elevated pulmonary venous and capillary pressures (Table 136.2). Alveolar

hemorrhage following bone marrow transplant can occur from immune-mediated causes and from complications of diffuse alveolar damage. Rarely, a previously well infant will present with life-threatening acute alveolar hemorrhage. Often no cause is found, and, once the acute episode resolves, the infant returns to normal. Most of these infants never have a second bleeding episode. **Idiopathic pulmonary hemosiderosis** is a rare disorder characterized by recurrent alveolar bleeding, iron-deficiency anemia, and hemosiderin-laden macrophages in the lung, which can be identified microscopically with the use of special iron-staining techniques in bronchoalveolar lavage or lung biopsy specimens. Although the term *hemosiderosis* is sometimes used interchangeably with pulmonary hemorrhage, it is a pathologic finding that results from bleeding anywhere in the lung, airway, pharynx, nasopharynx, or mouth leading to hemosiderin accumulation in the lung.

Diagnostic Studies

It is important to perform a thorough upper airway examination to rule out epistaxis. Sometimes this requires nasopharyngoscopy. If extrapulmonary sources of bleeding have been excluded, then a chest radiograph, CT angiogram of the chest, bronchoscopy, echocardiogram, and evaluation for rheumatologic/autoimmune diseases follow, especially to consider antiglomerular basement membrane disease, granulomatosis with polyangiitis, Henoch-Schönlein purpura, and systemic lupus erythematosus. Approximately one-third of children with hemoptysis will have a normal chest radiograph, and additional diagnostic evaluation is warranted.

Treatment

The management of acute episodes of pulmonary bleeding includes the administration of supplemental O₂, blood transfusions, and, often with acute alveolar hemorrhages, mechanical ventilation with PEEP to tamponade the bleeding. Treatment is directed toward the underlying disorders and providing supportive care. In bronchial arterial bleeding, arteriography with vessel embolization has been shown to be successful.

Pulmonary Embolism

Etiology

Pulmonary embolism (PE) is rare in children. When it occurs, it is often associated with indwelling central venous catheters, oral contraceptives, or hypercoagulable states. Other risk factors, such as trauma, surgery, immobilization, systemic lupus erythematosus, systemic infection, and cancer, may lead to deep vein thromboses (DVTs) and pulmonary embolism.

Clinical Manifestations

In the distensible pulmonary vascular bed, small emboli may be asymptomatic unless they are infected (septic emboli). Large emboli may cause acute dyspnea, pleuritic chest pain, cough, and hemoptysis. Tachypnea, tachycardia, and hypoxia are common, as are nonspecific ST-segment and T-wave changes on the electrocardiogram (ECG). The P₂ heart sound may be increased. Unilateral calf swelling may be present in the setting of DVT.

Diagnostic Studies

Imaging plays an important role in the evaluation and management of acute PE. The diagnostic approach in children

TABLE 136.2 Differential Diagnosis of Hemoptysis-Pulmonary Hemorrhage

CARDIOVASCULAR DISORDERS
Heart failure with pulmonary edema
Pulmonary hypertension with Eisenmenger syndrome
Mitral stenosis
Venooclusive disease
Arteriovenous malformation (Osler-Weber-Rendu syndrome)
Pulmonary embolism
Portal vein obstruction
PULMONARY DISORDERS
Bronchogenic cyst
Bronchopulmonary sequestration
Pneumonia (bacterial, mycobacterial, fungal, parasitic, or viral)
Bronchiectasis (cystic fibrosis, primary ciliary dyskinesia, immunodeficiency, retained foreign body)
Tracheobronchitis
Lung abscess
Tumor (adenoma, carcinoid, hemangioma, metastasis)
Trauma (contusion, laceration)
IMMUNE DISORDERS
Henoch-Schönlein purpura
Pulmonary capillaritis
Idiopathic pulmonary hemosiderosis
Antiglomerular basement membrane disease (Goodpasture disease)
Granulomatosis with polyangiitis (formerly known as Wegener granulomatosis)
Systemic lupus erythematosus
Polyarteritis nodosa
OTHER CONDITIONS/FACTORS
Coagulopathy (Von Willebrand factor)
Toxic inhalation (nitrogen dioxide, pesticides, crack cocaine)
Post-bone marrow transplantation
Catamenial hemoptysis (females)

differs somewhat from the approach in adults in that D-dimer testing and clinical prediction tools lack utility in the diagnosis of PE in children. CT pulmonary angiography (CTPA) is the current standard of care and provides accurate diagnosis with rapid turnaround time. CT also provides information on other potential causes of acute chest pain. Chest radiograph has limited utility, occasionally showing findings of PE or infarction, but is useful in evaluating other potential causes of chest pain. Ventilation-perfusion scans may reveal defects in perfusion without matching ventilation defects, but they are difficult to perform in young children. Catheter pulmonary angiography is no longer used for diagnosis and is used only for interventional management. Doppler or compression ultrasonography can be useful in assessing patients for lower extremity DVTs. Children with pulmonary embolism without an obvious cause should be evaluated for hypercoagulable states, the most common of which is factor V Leiden.

Treatment

The patient should be anticoagulated, usually with low-molecular-weight heparin or unfractionated heparin. Although rare, if pulmonary embolism is extensive and hemodynamically compromising, catheter-directed thrombolytic agents such as tissue plasminogen activator should be considered. All patients should receive supplemental O₂, and it is important to treat the predisposing factors.

PEARLS FOR PRACTITIONERS

See *The Respiratory System: Pearls for Practitioners* at the end of this section.