Respiratory distress is a common symptom in the newborn. Surfactant deficiency in the preterm infant and meconium aspiration and transient tachypnea in the term newborn are among the most common causes of respiratory distress, but primary airway abnormalities can also present with respiratory distress in the newborn. Delay in diagnosis of airway abnormality can be life threatening. Radiologists are among the first to be consulted in the evaluation of a neonate with respiratory distress, and knowledge of the spectrum of airway anomalies is essential for making the correct diagnosis. This article describes airway abnormalities that can present as respiratory distress in a neonate.

Choanal Atresia and Stenosis

Choanal atresia or stenosis is the result of buccopharyngeal and nasopharyngeal membrane resorption failure. The posterior choanae are narrowed with either atresia or stenosis. Choanal atresia occurs in 1 in 5000-8000 neonates and is more common in girls. Overall, 50%-60% of cases are unilateral. Respiratory distress relieved by crying in a neonate is the typical presentation. Unilateral choanal atresia can go undiagnosed until later in life, but bilateral obstruction is life threatening. Osseous obstruction occurs in 90% of cases with membranous, and osseomembranous obstruction accounting for the remaining cases. Coloboma, Heart defect, Atresia choanae, Retarded development, Genital anomalies, and Ear anomalies syndrome (CHARGE syndrome) is the most common syndrome associated with choanal atresia. Other associations include Treacher Collins syndrome, amniotic band syndrome, DiGeorge syndrome, and bowel malrotation. CT is the imaging modality of choice. Narrowing of the posterior choanae to less than 34 mm in children younger than 2 years is diagnostic of choanal atresia (Fig. 1). Other features on CT include inward bowing of the posterior maxilla, fusion or thickening of the vomer, and the presence of a bone or soft tissue septum across the posterior choanae. Treatment is delayed in unilateral obstruction to allow for midface growth. Membranous obstruction is treated by endoscopic perforation. Treatment options for osseous choanal obstruction include reconstruction and stent placement.

Laryngomalacia

Laryngomalacia is the collapse of supraglottic structures into the airway during inspiration. This produces the characteristic inspiratory stridor. Congenital laryngomalacia is the most...
common cause of newborn stridor and airway obstruction.\(^5\) Stridor worsens with feeding, crying, agitation, and supine position. Symptoms are usually mild with resolution by 12-24 months of age. The etiology is unknown, but the most accepted theory is that laryngomalacia results from underdevelopment of the nervous system controlling the airway.\(^5\)

Airway fluoroscopy can be used to diagnose laryngomalacia. Changes in the airway caliber during inspiration and expiration are assessed during the fluoroscopic examination.\(^6\) Direct flexible laryngoscopy can confirm the diagnosis in infants suspected of laryngomalacia.\(^7\)

Dynamic pulmonary CT examination is a new technique of assessing the airway (Fig. 2). A continuous scan acquisition during a respiratory cycle is performed and multiple dynamic reconstructions created. Cine loops can then be created allowing for qualitative and quantitative assessment of the larynx throughout the respiratory cycle.\(^8,9\)

Most infants have mild symptoms and are conservatively managed. Patients with associated gastroesophageal reflux or laryngopharyngeal reflux or both frequently respond to acid suppression therapy. However, in severe cases, supraglottoplasty may be necessary.\(^5\)

**Tracheobronchomalacia**

Tracheobronchomalacia results from weakness or deficiency of the tracheal skeleton\(^10\) causing increased tracheal and bronchial wall collapse during expiration.\(^10\) Tracheomalacia is the most common congenital anomaly of the trachea with an incidence between 1 in 500 and 1 in 2500.\(^11\) This is an underestimation as evaluation has previously required invasive techniques such as bronchoscopy for diagnosis.\(^11\) Tracheobronchomalacia can occur as an isolated condition, be part of a syndrome, or be associated with other anomalies.\(^10\) Syndromes associated with tracheomalacia include Down syndrome, DiGeorge syndrome, Larsen syndrome, and Williams-Campbell syndrome.\(^10\) Tracheomalacia can occur in association with tracheoesophageal fistula or gastroesophageal reflux. It can be associated with extrinsic compressions such as vascular rings and masses.\(^11\) Primary tracheomalacia can occur in both term and preterm neonates. Tracheomalacia can be acquired in premature babies with chronic lung disease.

The symptoms of tracheobronchomalacia are nonspecific making the diagnosis difficult. Children with tracheobronchomalacia are prone to lower respiratory tract infections and with a prolonged course of recovery.\(^11\)

Though expiratory wheeze is a classic sign in an older child, the presentation in neonatal life is that of respiratory distress, difficulty weaning off the ventilator, feeding difficulties, and recurrent life-threatening events.\(^11\) Inspiratory and expiratory radiographs have been used to diagnose tracheobronchomalacia with a reported sensitivity of 62%. Cine fluoroscopy unlike static radiographs allows for dynamic airway assessment.\(^6\) The fixed narrowing of stenosis can be differentiated from malacia. Fluoroscopy examination does not require the same degree of patient cooperation as radiography, and assessment of the airway can be done while the patient is crying or feeding. The entire airway from nasopharynx to the

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**Figure 1** Maxillofacial CT without IV contrast in a full-term, 1-day-old female patient with history of respiratory distress soon after birth and failure to pass nasogastric tube. (A) Axial image demonstrates bilateral choanal stenosis (arrows). (B) Sagittal reformatted image demonstrates a bony septum on the right side (arrow). IV, intravenous.

**Figure 2** A 3D reformatted sagittal image in a preterm male infant with history of stridor and ventilator dependence demonstrates supraglottic airway narrowing (arrow). (Color version of figure is available online.)
bronchi can be imaged. The reported specificity is 94%-100%
for diagnosing tracheobronchomalacia. However, there are
limitations of this technique. The assessment is subjective. The
resolution of the distal airway is particularly poor. The
anatomical details of trachea and especially the paratracheal
structures are suboptimal.

Dynamic pulmonary CT with or without intravenous
contrast can objectively evaluate the airway. A wide-detector scanner is required, and scanning is performed in continuous mode for 1 respiratory cycle. This can be performed in a self-ventilating or intubated child. Multiple
dynamic phases are reconstructed, and cine loops are created
d for evaluation. Qualitative changes in airway diameter
can be evaluated or quantitative reduction in the cross-
sectional area can be measured allowing for objective diagnosis
de of tracheobronchomalacia. Tracheobronchomalacia can be
defined as greater than 28% reduction in airway cross-
sectional area during expiration. This technique allows
for the assessment of both intrinsic airway abnormalities and
extrinsic compression of the airway.

Mild to moderate tracheomalacia is managed conservatively
as the tracheal cartilage grows and symptoms improve by
approximately 2 years of age. Treatment of infections, gastro-
esophageal reflux, and chest physiotherapy help to alleviate
the symptoms in mild to moderate cases. Positive pressure
ventilation is used in children with moderate or intermittent
severe obstruction. Tracheostomy can bypass the malacic
segment. Airway stents may be placed to maintain patency.
Tracheoplasty may be needed for diffuse tracheomalacia.

Tracheal Stenosis

Congenital tracheal stenosis is defined as tracheal narrowing,
which is most often caused by complete cartilaginous rings.
Submucosal glands and connective tissue hypertrophy com-
plicate the stenosis. Severe congenital stenosis and atresia are
rare malformations occurring with an estimated rate of 2 per
100,000 live births. Most cases of tracheal stenosis are
acquired with congenital tracheal stenosis representing only
0.3% to 1% of all laryngotracheal stenosis. Congenital
tracheal stenosis is frequently associated with other anomalies
with the most common association being cardiovascular
anomalies. Isolated congenital tracheal stenosis accounts for
only 10%-25% of patients. Cardiovascular anomalies asso-
ciated with congenital tracheal stenosis include left pulmonary
artery sling, vascular rings, patent ductus arteriosus, and
ventricular septal defects. Other associations include pulmo-

Figure 3 Dynamic pulmonary CT in a male patient with history of bradycardia and reflux since birth and subsequent
development of apneas demonstrates collapse of trachea as well as more distal airways with expiration. (A) Axial images in
inspiration (left) and expiration (right) show collapse of trachea during expiration. (B) The 3D reformatted coronal images
in inspiration (left) and expiration (right) demonstrate malacia of both main stem bronchi and right upper and middle lobar
bronchi (arrows). (Color version of figure is available online.)
Congenital tracheal stenosis has been classified into 3 types by Cantrell and Gould. Type I is generalized hypoplasia, type II is funnel type stenosis with one normal end and the other stenotic, and type III is segmental stenosis with involvement of 2 or 3 cartilage rings.14

Symptoms depend on the trachea stenosis severity. An infant with severe stenosis is usually symptomatic soon after birth with respiratory distress, is difficult to intubate with an appropriate-sized endotracheal tube, and has increased ventilator requirements. Patients with less severe stenosis present with stridor, wheezing, failure to thrive, and cyanosis.17

Rigid bronchoscopy is the gold standard for the diagnosis of tracheal stenosis. The technique requires general anesthesia, and narrowing may be so severe that the smallest pediatric endoscope cannot penetrate the stenosis. Rigid bronchoscopy is invasive and can lead to mucosal damage and further stenosis.14

Nonspecific findings on chest radiograph include air trapping (Fig. 4A); the region of airway narrowing and abnormal airway branching may be apparent on plain films but is often subtle. CT is widely accepted as an excellent imaging modality to evaluate tracheal stenosis.

Multidetector CT allows 3D reformations and virtual endoscopy (Fig. 4B and C). Additional advantages of CT include delineation of mediastinal vascular anatomy and assessment of external airway compression. MRI including cine MRI has also been used in the diagnosis of tracheal stenosis.14

Surgery is the primary treatment for tracheal stenosis. Patients with mild symptoms are conservatively managed as symptoms are likely to improve with growth. Tracheal resection with end-to-end anastomosis is the procedure of choice for short-segment stenosis. Longer stenoses are best managed by tracheoplasty. Slide tracheoplasty by using native tracheal tissue is a popular technique as granulation tissue is

![Figure 4](https://clinicalkey.com/assets/images/figure/4.png)
minimal owing to the use of native tissue and hence the need for stenting is reduced.\textsuperscript{16}

**Extrinsic or Combined Vascular-Airway Abnormalities**

These include the following:
- Vascular rings
- Anomalous innominate artery syndrome
- Pulmonary sling

**Vascular Rings**

Vascular ring is defined as abnormal encirclement of the trachea and esophagus by the aortic arch and its branches.\textsuperscript{16} The 2 most common vascular rings are double aortic arch and right arch with an aberrant left subclavian artery. Vascular rings represent 1\% of the cardiovascular congenital anomalies.\textsuperscript{19} Vascular rings can be patent or have atretic segments.\textsuperscript{20} Some vascular rings can be associated with other cardiac anomalies, whereas others occur in isolation.\textsuperscript{20} Symptoms vary depending on the tightness of the vascular ring.

Symptoms include dyspnea, wheezing, stridor, failure to extubate, and frequent respiratory tract infections. Cyanosis and acute life-threatening events such as reflex apnea are rarely encountered. Compression of the esophagus presents with feeding difficulties.\textsuperscript{21}

*Double aortic arch*: Double aortic arch is the most common cause of vascular compression of the airway in children.\textsuperscript{22} The trachea and esophagus are encircled by the right and left arches (Fig. 5). The right arch is typically higher and dominant in 70\% of cases. The left arch can be dominant but is usually equal or

**Figure 5** A 9-day old full-term female patient with respiratory distress and stridor. (A) Lateral chest radiograph shows tracheal narrowing (arrow). (B) Axial CT of chest with IV contrast demonstrates double aortic arch (arrow) compressing the trachea. (C) A 3D coronal reformatted image demonstrates the tracheal narrowing (arrow) at the level of the vascular ring. IV, intravenous. (Color version of figure is available online.)
smaller than the right aortic arch. An atretic segment composed of a fibrous cord can complete the ring (typically on the left side). The descending aorta may be on the right, left, or midline. Right aortic arch with aberrant left subclavian artery is the second most common vascular ring. A diverticulum of Kommerell connects the right aorta arch to the true but aberrant left subclavian artery. A ligamentum arteriosum completes the ring. Double aortic arch and right arch with aberrant left subclavian artery can be isolated or associated with other cardiac anomalies.

Chest radiography and contrast esophagography can demonstrates a vascular ring. These studies demonstrate the mass effect on adjacent structures such as the trachea and esophagus. Chest radiography will demonstrate the laterality of the aortic arch by observing deviation and mass effect on the trachea (Fig. 6A). Pickhardt found at least 1 abnormal finding on chest radiograph in every patient in a study of 41 frontal and 39 lateral radiographs of patients with vascular rings. A midline trachea is a sign of a double aortic arch. The aortic arch may be difficult to directly identify in a neonate, but the pedicles will be denser on the side of the aortic arch.

Computed tomography angiography (CTA) and MRI are more definitive diagnostic imaging tests for vascular rings. Both CTA and MR angiography accurately depict the vascular anatomy and allow visualization of the airway (Fig. 6B). CTA is faster than MRI and can be performed in infants using less than a millisievert of ionizing radiation. MRI frequently requires sedation and carries greater risk than CTA in a neonate with respiratory distress. Both techniques allow for 3D reconstructions which are helpful for surgical planning. Focal intrinsic airway abnormalities such as tracheobronchomalacia are associated with vascular rings. Evaluation of airway dynamics in patients with mediastinal vascular anomalies may be helpful. Surgery includes ligation of the vascular ring. Aortopexy may be necessary to relieve airway compression and is followed by airway caliber improvement.

**Anomalous Innominate Artery Syndrome**

The anomalous innominate artery syndrome is anterior compression of the trachea by a crossing innominate artery that causes respiratory symptoms. Up to 30% of normal children have mild to moderate tracheal narrowing by the innominate artery without causing symptoms. The mild tracheal compression in these children is a normal developmental variant. However, tracheal compression by the innominate artery can result in severe respiratory distress that is relieved by aortopexy (Fig. 7A and C). Tracheomalacia due to the innominate artery compression is a frequent finding in these symptomatic children. A lateral chest radiograph may demonstrate anterior tracheal indentation. CTA and MRI are definitive in demonstrating the mass effect. Dynamic airway CT and cine MRI are particularly useful in demonstrating tracheomalacia.

**Pulmonary Sling**

Pulmonary sling accounts for approximately 10% of airway abnormalities associated with vascular anomalies. The left pulmonary artery arises from the right pulmonary artery and courses between the trachea and the esophagus. Most patients are symptomatic soon after birth. Airway narrowing may be caused directly by vascular compression, but most also have intrinsic airway abnormalities (Fig. 8). Associated anomalies include tracheobronchomalacia, tracheal stenosis, and complete tracheal rings. A separate right tracheal bronchus can be associated with pulmonary sling. Pulmonary arterial slings are classified into 2 types. In type 1, the carina is at the T4-T5 level. The anomalous left

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**Figure 6** History of stridor in a term male patient with facial and airway hemangiomas. At bronchoscopy, there was right tracheal wall external compression with pulsation. (A) Chest radiograph demonstrates the right aortic arch (arrow) impression on the trachea. (B) Axial postcontrast CT of chest demonstrates a right arch with aberrant left subclavian artery (curved arrow) with associated mild narrowing of the trachea (asterisk) and severe compression of the esophagus (arrow). (Color version of figure is available online.)
pulmonary artery courses around the distal trachea and right bronchus before coursing to the left lung. Type 1 pulmonary sling is associated with distal tracheal and right bronchus narrowing and malacia with resultant right lung air trapping. The type 2 pulmonary sling has a more caudal-positioned carina, which is T shaped. The sling is at T5-T6 and is more often associated with abnormal bronchial branching such as a bridging right bronchus as well as long-segment tracheal stenosis.

Tracheal narrowing, right lateral indentation of the lower trachea, and an inverted “T”-shaped carina may be apparent on conventional radiography. Anterior deviation of the lower trachea and carina can be seen on lateral views. Other findings on chest radiography include unilateral or bilateral lung hyperinflation. Unlike vascular rings, an anterior esophageal indentation is present on esophagography. CTA and MRI are definitive in the assessment of the left pulmonary artery course in patients with pulmonary sling. The tracheobronchial tree is best evaluated by CT. Dynamic airway assessment with CT or MRI is particularly helpful in obtaining additional information on tracheobronchial stenosis and tracheobronchomalacia for surgical planning.

Treatment is by surgical reimplantation of the anomalous left pulmonary artery to the pulmonary artery, anterior to trachea. Tracheoplasty or tracheal resection with end-to-end anastomosis is performed to correct associated airway anomalies.

Summary

Radiologists are among the first to be consulted in the evaluation of a neonate with respiratory distress. Surfactant deficiency in the preterm infant and meconium aspiration and transient tachypnea in the term newborn are among the most common causes of respiratory distress, but primary airway

Figure 7 A 1-month-old male patient with history of wheezing. (A). Axial CT of chest with IV contrast demonstrates the right brachiocephalic artery (asterisk) crossing anterior to trachea with associated mild narrowing of the trachea (arrow). Patient was treated conservatively. (B). A 3D coronal reformatted image of CT chest in a different patient with anomalous innominate artery syndrome (arrow). (C) A 3D coronal CT image of the airway in the same patient as in B demonstrates severe tracheal narrowing (arrow). Patient underwent innominate artery pexy to the sternum. (Color version of figure is available online.)
abnormalities can also present with respiratory distress in the newborn, and knowledge of the airway anomalies is essential for making the correct diagnosis. Chest radiographs may show indirect signs such as hyperinflation or atelectasis of lungs, abnormal position of aortic arch, poorly visualized or narrowed airway, and abnormal position and shape of carina. These findings should alert the radiologists to an associated airway abnormality. CT or MRI allows an accurate diagnosis and influences patient management.

References


Figure 8 A full-term female patient with antenatal diagnosis of duodenal atresia with difficult intubation. (A) Axial CT chest with IV contrast at 13 days of age demonstrates left pulmonary artery sling (arrow) partially encircling the trachea and extending between the trachea and the esophagus. (B) A 3D coronal reformatted image demonstrates long-segment tracheal stenosis (arrow) and a low T-shaped carina. IV, intravenous. (Color version of figure is available online.)