Chapter 124: Congenital Disorders of the Trachea

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Investigation of the larynx and pharynx may be incomplete in infants and children with congenital abnormalities without investigation of the tracheobronchial tree. In fact, the upper airways should be studied from the nasal cavities above to the bronchi below. This chapter discusses specific congenital tracheal anomalies.

Infants with congenital anomalies of the trachea other than the classic tracheoesophageal fistula and esophageal atresia have respiratory obstruction and stridor. The stridor may have not only an inspiratory component but also an expiratory phase. A barking cough, cyanotic attacks, apneic attacks, reflex apnea, aspiration, repeated, prolonged, or atypical croup, and sometimes hyperextension of the neck may all be features that indicate possible tracheal narrowing.

The tracheal lumen in the infant can be narrowed by compression from outside of the trachea itself, by wall weakness, webs, stenoses, developmental abnormalities, duplication cysts, and other rare congenital anomalies.

Tracheoesophageal Fistula

Tracheoesophageal fistula occurs with or without various types of esophageal atresia. Its presentation in the immediate neonatal period occurs with respiratory distress, excessive mucus, and cough, choking, and cyanosis associated with feeding.

Endoscopic examination is not necessary in all infants with tracheoesophageal fistula and esophageal atresia. The indications for endoscopy are diagnostic or therapeutic (Benjamin, 1981).

Diagnostic indications include evaluation in selected cases before operative repair; diagnosis of an H-fistula; assessment of tracheomalacia; identification of recurrent, residual, or a second fistula; evaluation of the presence of an associated anomaly; evaluation of esophageal stenosis, either congenital or acquired; and assessment of esophagitis caused by gastroesophageal reflux.

The therapy is dilatation of an established esophageal stricture after surgical anastomotic repair and removal of an impacted esophageal body.

Tracheoesophageal fistula and esophageal atresia

A newborn baby with rattling respirations, fine frothy bubbles of mucus at the lips or nostrils, and evidence of aspiration should have the patency of the esophagus tested clinically. The diagnosis of this common anomaly of esophageal atresia with lower-pouch fistula is established by failure to pass a catheter. With the less common varieties of malformation the exact diagnosis may be elusive, treatment may be delayed, or an incorrect operative procedure may even be performed. Preoperative diagnostic endoscopic evaluation in the neonate with esophageal atresia and a tracheoesophageal fistula before definitive surgery (Fig. 124-1) may

give valuable information to the surgeon before repair in selected cases. At endoscopy the site and size of the fistula and the state of the tracheal mucosa, whether soiled by gastric juice or having infected secretions, can be accurately judged. A fine catheter can be passed via the bronchoscope through the fistula into the lower esophagus and left in place so that the surgeon can quickly and confidently identify the fistula during a thoracotomy. The exact diagnosis and pathologic anatomy is established. Endoscopy also allows assessment of associated malformations that could be important in operative or postoperative care, for example, subglottic stenosis, vocal cord paralysis, tracheomalacia, tracheal or bronchial stenosis, an upper esophageal segment fistula, a second fistula, or an unusual fistula (Fig. 124-2).

H-type of tracheoesophageal fistula

The key to the eventual diagnosis of an H-type of fistula is to be aware of the possibility from clinical features, which include intermittent repeated aspiration, persistent cough, scattered segmental areas of atelectasis on the radiographic film, cyanotic attacks, and in severe cases, failure to thrive. Endoscopic identification of an elusive H-type of fistula or a recurrent fistula after primary definitive repair requires meticulous examination of the posterior wall of the trachea and of the anterior wall of the esophagus with angled telescopes. Such a diagnostic examination of the patient (who is under general anesthesia) is not only more reliable but is also safer than radiologic examination of the esophagus.

Neither rigid open-tube telescopic endoscopy nor contrast radiography are totally reliable in the detection of an H-type tracheoesophageal fistula. Each technique alone may miss a suspected fistula; the use of both techniques should ensure its identification. In addition, radiography will show neuromuscular incoordination of swallowing and assist in the diagnosis of gastroesophageal reflux. Endoscopy will reveal any associated abnormalities of the upper aerodigestive tract. Also, endoscopy immediately prior to corrective surgery will allow placement of a catheter to assist in localization of the fistula. Thus the two diagnostic techniques are complimentary.

Should inhalation of contrast material occur in the radiography department, choking cyanosis and apnea may occur and facilities for resuscitation are not always readily available.

Positive-pressure ventilation of the trachea through the endotracheal tube, after a few milliliters of saline have been instilled into the upper esophagus with an esophagoscope in place, has proved to be a useful endoscopic technique in detecting a small fistula. Bubbles of air will be seen coming from the fistula itself. Endoscopic catheterization of the fistula with a fine catheter facilitates surgical treatment.

The incidence of associated abnormalities of the trachea and of the esophagus in patients with an H-type of fistulas is high. These anomalies include tracheal stenosis, esophageal stenosis, and tracheomalacia.

Postoperative symptoms

Symptoms relating to the airway and to the esophagus after surgical repair of esophageal atresia and a tracheoesophageal fistula often exist in the postoperative period and may be caused by tracheomalacia, bronchomalacia, tracheal stenosis, esophageal anastomotic stricture, gastroesophageal reflux with or without aspiration, and occasionally, recurrent or residual fistula. In the past, features such as respiratory distress, infection, obstruction, stridor, retention of secretions, barking cough, aspiration, cyanotic attacks, and apneic attacks following repair of esophageal atresia were rather loosely considered as postoperative pulmonary complications. It is now known that tracheomalacia is the major factor producing these symptoms, whose severity can be contributed to by tracheal compression by the innominate artery; food or an impacted foreign body that fills the dilated upper segment of the esophagus above the esophageal anastomosis; or aspiration into the trachea, which may be a result of disordered physiology or gastroesophageal reflux and disordered mucociliary mechanism. The clearing action may be impeded by the wide mouth of the residual pouch of the fistula, which crosses the posterior wall. The pouch occasionally contains a suture, granulation tissue, infected discharge, or soft-tissue reaction. Emery and Haddadin (1971) demonstrated squamous epithelium in the trachea of patients with tracheoesophageal fistulas who clinically had sputum retention.

Tracheomalacia

Tracheomalacia is a condition affecting part or all of the tracheal wall when there is weakness caused by softening of the supporting cartilage and an abnormal widening of the posterior wall. The trachea lacks its usual degree of firmness, and the anterior and posterior walls come closer together during respiration and coughing, with a resulting reduction of the tracheal lumen.

Wailoo and Emery (1979) showed that the structural changes in tracheomalacia include deficiency of the cartilage and increase in the width of the posterior membranous tracheal wall. The ratio of the length of the cartilage to the width of the transverse muscle is abnormally reduced. The less the proportion of the tracheal wall that is cartilage, the more collapsible is the trachea, and the wide floppy posterior wall balloons forward into the lumen, especially during expiration.

Clinicians have an increased perception of the relationship between respiratory disease in infants and children and abnormal weakness and collapse of the trachea. The diagnosis of tracheomalacia requires awareness on the part of the clinician, skilled interpretation of the lateral airway radiographic films, and expert endoscopic evaluation.

In general the clinical features include inspiratory and/or expiratory stridor, wheezing, a cough that sometimes has a peculiar barking quality, hyperextension of the neck, recurrent respiratory infections, difficulty clearing endobronchial secretions, and sometimes attacks of reflex apnea. The latter are dramatic episodes of respiratory arrest, occasionally progressing to cardiac arrest; they have been aptly referred to as "dying spells". They indicate the need to consider surgical suspension of the trachea.

Tracheomalacia in infants and children has been classified into primary and secondary (Benjamin, 1984b). Primary tracheomalacia occurs in premature infants, in otherwise normal, full-term infants, and in infants with one of the rare dyschondroplasias. Secondary tracheomalacia occurs with tracheoesophageal fistula and innominate artery compression or other forms of external compression.

Primary tracheomalacia

Premature or very premature infants treated for respiratory distress syndrome by prolonged endotracheal intubation with assisted positive-pressure ventilation may have wheezing, stridor, barking cough, and susceptibility to lower respiratory tract infections when the endotracheal tube is removed. Tracheomalacia and bronchomalacia can be recognized at endoscopic examination.

Mature infants who are otherwise normal occasionally have severe primary tracheomalacia in the first few weeks of life, with respiratory distress, cyanosis, wheezing, and inspiratory or expiratory stridor. Early bronchoscopy is essential for a definitive diagnosis. Tracheomalacia is seen with the typical lack of cartilaginous support and widened posterior membrane wall, allowing collapse during expiration. In severe cases, respiratory support by means of prolonged intubation with high positive pressures for assisted ventilation is necessary for weeks or months. If death does not occur, spontaneous resolution may take many months or even several years.

Secondary tracheomalacia

Tracheoesophageal fistula

Tracheomalacia causes symptoms that can be classified as mild, moderate, or severe, depending on the age of the child and presentation of the symptoms, their severity, the amount of respiratory obstruction, sputum retention, and whether or not there are attacks of reflex apnea. In severe cases the symptoms develop within the first 2 months of life. If they are life-threatening, then surgical suspension of the trachea should be considered (Fig. 124-3).

Tracheomalacia is the major cause of respiratory problems after repair of a tracheoesophageal fistula. Diagnostic endoscopy is mandatory. The tracheal cartilages have an indented half-circle shape rather than the normal horse-shoe shape, the posterior membranous wall is markedly widened, and there is forward bulging, narrowing the anteroposterior lumen. This triad affects several centimeters of the lower trachea - usually the lower third, a longer segment than is affected in innominate artery compression (Fig. 124-4).

Innominate artery compression

Innominate artery compression is well recognized as a cause of respiratory symptoms in infants (Fearon and Shortreed, 1963). The clinical features include inspiratory and sometimes also expiratory stridor, a cough that sometimes has a barking quality, recurrent pneumonia or bronchitis, retained secretions, attacks of atypical croup, and attacks of reflex apnea. The endoscopic observations (Fig. 124-5) in moderate or severe innominate artery compression are similar to those in tracheomalacia associated with tracheoesophageal fistula,

with two notable exceptions: a shorter segment of the lower trachea is involved, and the associated finding of a wide posterior membranous wall with anterior bulging does not always exist. Thus innominate artery compression appears to occur with or without tracheomalacia. The degree of innominate artery compression can be recorded as a percentage of the loss of tracheal lumen. A relationship exists between the severity of stridor and the loss of the tracheal lumen. On the other hand, relatively mild compression may exist in patients with reflex apnea. Again, life-threatening attacks of reflex apnea warrant surgical suspension of the trachea.

External compressions

Tracheomalacia in these cases is more localized and is secondary to external compression by congenital anomalies of the great vessels or heart, a bronchogenic or other duplication cyst, teratoma, abscess, cystic hygroma, hemangioma, or other tumor (Fig. 124-6).

A localized segment of soft, floppy tracheal cartilaginous wall remains as an area of weakness. For example, after division of a vascular ring or removal of bronchogenic cyst, the features of tracheal weakness and narrowing tend to persist. There is a localized segment of soft tracheal wall that may remain after surgical removal of the cause. Endoscopic evaluation shows the area subject to collapse from increased intrathoracic pressure. It may be months or years before this area becomes firm and the patient is free of symptoms.

Direct endoscopic examination remains the only reliable diagnostic examination giving definitive information about the dynamics of the tracheal caliber. The diagnosis of tracheomalacia should not be made by exclusion. No amount of inferential supposition or logical interpretation of chest radiographs can supply the information obtained during endoscopy with slim, rigid fiberoptic telescopes, which can be inserted without distortion or distension of weak or collapsing areas. The slim rigid telescopes alone, without a bronchoscope, can be used as a tracheoscope to obtain a perfectly clear, enlarged image beyond the tip of the examining instrument, so that a final and definitive diagnosis of tracheomalacia can be made.

Vascular Compression of the Tracheobronchial Tree

Congenital malformations of the heart and great vessels may cause compression of the tracheobronchial tree and stridor. Clinical radiologic and endoscopci assessment is necessary; the latter is especially needed for assessment of the obstruction's site and extent.

Vascular ring

The ring may be purely vascular. A double aortic arch is a ring that encircles the trachea and esophagus. Two arches compress the trachea and esophagus as one arch passes to the right and one arch to the left (Fig. 124-7). They join to form a descending aorta. The arches may be of the same size, or one may be larger.

The ring may be fibrovascular. In this cases the aorta does not divide but passes, together with an arterial ligament, to the right side of the trachea and the arch to connect with the left pulmonary artery and to compress the trachea and the esophagus.

Vascular sling

The left pulmonary artery passes between the trachea and esophagus, causing pressure on both; symptoms are caused only by narrowing of the tracheal airway (Fig. 124-8).

Posterior anomalies

An aberrant right subclavian artery passing behind the esophagus usually causes no clinical features but can be seen as a posterior indentation on the barium swallow radiograph.

Anomalies of branches of the aortic arch

The anterior wall of the trachea may be compressed when an anomalous innominate artery arises to the left of its usual source and passes from left to right, compressing the anterior wall of the trachea as it does so. There are several variations of this condition involving an aberrant left common carotid, and abnormal right subclavian artery passing anterior to the trachea, and a left subclavian artery arising from a right aortic arch.

Pulmonary artery dilatation

Compression of the pliable cartilage of the tracheobronchial tree by dilated, hypertensive pulmonary arteries can also occur in infants with either acyanotic congenital heart disease involving large left-to-right shunts (such as ventricular septal defects and patent ductus arteriosus) or the anomalies causing cyanosis, such as the Eisenmenger syndrome and transposition of the great vessels (Berlinger et al, 1983).

Although congestive cardiac failure, pneumonia, or compression of lung parenchyma often causes respiratory difficulties in these infants, in some cases the dilated pulmonary arteries compress the tracheobronchial tree, leading to recurrent or intractable atelectasis, pneumonia, or sometimes lobar emphysema. A dilated left pulmonary artery can compress the upper aspect of the left main bronchus or the posterior aspect of the left upper lobe bronchus. A dilated right pulmonary artery can compress the superior aspect of the right intermediary bronchus and middle lobe bronchus.

Dilated pulmonary arteries may displace the aorta against the left lateral aspect of the trachea and cause compression. When left atrial enlargement occurs, compression of the left bronchus by the dilated pulmonary arteries is made worse, pushing up both bronchi and thereby increasing the angle of the tracheal bifurcation.

Clinical features

The clinical features include not only nonspecific respiratory distress, tachypnea, unresolved pneumonia, and atelectasis, but also specific features that suggest compression of the large airways below the larynx, such as inspiratory and expiratory stridor, barking cough, retained secretions, arching of the head and neck, and segmental lobar emphysema. Difficulty swallowing is seldom a feature.

Radiologic examination

Radiologic studies include a plain anteroposterior chest film, lateral views of the laryngopharynx and trachea, and a contrast esophagogram. Narrowing or indentation of the normal air column and compression or notching of the esophageal outline occur according to which malformation is present. For instance, a vascular ring caused by a double aortic arch may show compression of the lower trachea (Fig. 124-9) and notching of the posterior esophagus (Fig. 124-10) at the same level; a vascular sling shows separation of the posterior wall of the lower trachea from the anterior wall of the esophagus (Fig. 124-8). Lower tracheal and bronchial compression at one of the sites of predilection can be suspected in cases of hypertensive pulmonary artery compression. Cardiac catheterization, angiocardiography and ultrasonography are necessary in selected cases, especially before a thoracotomy for double aortic arch or one of its vascular ring variants. Sometimes ventilation and perfusion studies are indicated. Contrast tracheobronchography has no role.

Endoscopy

Endoscopic investigation allows accurate assessment of the anatomic abnormality, the site, nature, and extent of the compression, and degree of secondary tracheomalacia. If a large vessel is to be divided (double aortic arch), a tracheopexy planned (innominate artery compression), or a pulmonary arteriopexy performed (dilated pulmonary artery), then preoperative evaluation of the airways allows rational postoperative assessment. Examination of the esophagus is an essential part of the examination. The endoscopist must look for other congenital abnormalities. An awareness that hypertensive dilated pulmonary arteries can occur with acyanotic congestive heart disease and can cause compression improves endoscopic diagnostic accuracy.

Cooperation among the neonatologist, cardiologist, endoscopist, and pediatric surgeon permits earlier and more exact diagnosis, leading to better management of these difficult problems.

Tracheal Stenosis

Congenital tracheal stenosis is an uncommon serious congenital abnormality and is very often associated with other major or minor anomalies of the respiratory tract, skeleton, or other organ systems. The association with the H-type of tracheoesophageal fistula, pulmonary hypoplasia (Fig. 124-11), and vascular sling (Benjamin et al, 1981; Cohen and Landing, 1976) is common.

The malformed segment of the trachea has an intrinsic narrowing of the lumen, the walls of which are rigid and nondistensible. The condition affects part or all of the trachea and usually extends into the main bronchi. The cartilages are firm, nonpliable, and usually complete rings (Fig. 124-12) and therefore lack the normal structure of the posterior membranous trachea (Benjamin et al, 1981).

The age of presentation is usually the first weeks or months of life. Males and females are equally affected. An undue delay in diagnosis often occurs because the clinician does not consider the possibility of congenital tracheal stenosis. The presenting clinical features are often multiple and include respiratory distress, persistent wheeze or stridor, atypical croup or bronchiolitis, cough, cyanotic attacks, and failure to respond to the usual treatment. In undiagnosed patients treated symptomatically by intubation and ventilation for respiratory failure, introduction of an endotracheal tube often compounds the problem. The distal tip of the tube is very likely to cause edema and form granulations where it traumatizes the upper aspect of the stenosis, leading to further narrowing.

The presentation does not occur in any set pattern. All patients have some form of respiratory obstruction. If the attending clinician is aware of the condition and alert to its clinical manifestations, endoscopic examination is mandatory to make the diagnosis. Endoscopic examination, even by an experienced laryngologist with expert pediatric anesthesiology, is fraught with danger in these cases. The drying effect of the premedication and the anesthetic gases and the trauma of the endoscopic instruments' passage can easily worsen an already critical situation. In most cases the diameter of the stenosed segment is less than 3 mm, and the smallest bronchoscope should be passed only into the upper trachea proximal to the stenosis. Even the 2.8-mm rigid fiberoptic telescope is difficult or impossible to pass to make a satisfactory evaluation of the lower trachea and bronchi. A 1.9-mm Storz-Hopkins telescope is now available.

Contrast tracheobronchography (Fig. 124-13) can be performed in selected cases but is potentially dangerous. Where possible, it should be performed when the patient is in remission. The procedure must be carefully planned with meticulous attention to detail, extreme care with anesthesia, and expert postoperative intensive care. Documentation of the nature and extent of the tracheal stenosis and any associated bronchial stenosis (Fig. 124-14) is important to identify the presence of other associated anomalies, to evaluate the possibility of surgical correction, to plan medical therapy, and to assist in the assessment of the prognosis. Although successful segmental surgical resection has been reported, it is only very rarely feasible, the limiting factors being the length of the tracheal stenosis, the degree of the accompanying bronchial stenosis, and the associated abnormalities of other major organs.

In most cases conservative management is indicated, consisting of regular physiotherapy (intensified when there is an intercurrent respiratory infection), humidification of the inspired air, administration of antibiotics when appropriate, and avoidance of the use of an endotracheal tube. Admission to the hospital for observation and treatment is occasionally necessary. Some children seem to live reasonably well with their disease, the stenosis apparently widening as the child grows. In recognized cases it is good practice, when endotracheal intubation is necessary for assisted ventilation, to position carefully the distal end of the tube at least 1 cm above the proximal end of the tracheal stenosis. Suction with catheters is kept to an absolute minimum, and the endotracheal tube is removed as soon as possible. Approximately half of the patients with congenital tracheal stenosis die of either that condition or major congenital anomalies associated with it.