Thoracic Surgery Considerations in the Child and Young Adult

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KEYWORDS

• Vascular ring • Pulmonary artery sling • Tracheobronchial compression • Congenital heart surgery

KEY POINTS

- Various pathologic aortic arch anomalies or cardiomegaly from congenital heart disease can lead to significant compression of the trachea, bronchii, and/or the esophagus.
- As a general rule, neonates and infants present with airway issues, whereas dysphagia tends to occur more widely in older children and adults.
- A vascular ring may be suggested on chest radiography by the nonspecific findings of a pulmonary infiltrate, atelectasis, unilateral or bilateral hyperinflation, or the aortic arch position.
- Barium esophagogram has been replaced by computed tomography angiography and cardiac gated MRI in the assessment of aortic arch anomalies.
- Vascular rings or slings may result in important obstructive airway or esophageal symptoms, necessitating division of the ring or relocation of the sling.

INTRODUCTION

Thoracic surgery in the pediatric population encompasses a vast array of pathologies, including problems of the trachea and airway, esophagus, diaphragm, lungs, chest wall, heart, and great vessels. Many of these conditions are managed by the general pediatric surgeon and represent a core component of that specialty. For example, disorders such as esophageal atresia, tracheoesophageal fistula, congenital diaphragmatic hernia, chest wall deformities, and bronchopulmonary malformations (congenital pulmonary airway malformation, pulmonary sequestration, congenital lobar emphysema and bronchogenic cyst) fall under the purview of the general pediatric surgeon. As such, these disorders are not covered in this issue of *Thoracic Surgery Clinics*.

The congenital heart surgeon manages all disorders in the pediatric population involving the heart, great vessels, and central airway, including compression of the trachea/bronchii and esophagus from vascular structures or their remnants and congenital tracheal stenosis. Vascular pathology involving the trachea and bronchi are typically managed in conjunction with either the pediatric surgeon or the pediatric otolaryngologist. Because many of these neonates and children with primary tracheal and bronchial pathology also have associated congenital heart defects, most often as a part of a syndromic disorder (such as VACTERL [vertebral, anal, cardiac, tracheoesophageal, renal, limb defects]), a congenital heart surgeon is typically involved in their care.

Disclosure Statement: Neither author has any financial or commercial interest or disclosures relevant to the content of this article.

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Thorac Surg Clin ■ (2017) ■–■

https://doi.org/10.1016/j.thorsurg.2017.08.005

This article reviews commonly encountered pathologic vascular anomalies associated with tracheobronchial disorders as well as primary tracheal disorders that are encountered in a pediatric cardiothoracic practice.

EMBRYOLOGY

By 5 weeks of fetal development, the primordial heart tubes have fused and 6 paired aortic arches form connecting the dorsal and ventral aortae. The arches develop serially. That is, the first and second arches followed by the third and fourth, then the fifth and sixth. In normal development, the first and second arches undergo near complete involution with minor contributions to the facial arteries. Septation of the conotruncus forms the proximal ascending aorta and the proximal pulmonary artery. The distal ascending aorta, the aortic arch until the left common carotid artery, and the innominate artery derive from the aortic sac. The third aortic arch forms the bilateral carotid arteries. The left fourth aortic arch forms the distal aortic arch, aortic isthmus, and joins the truncus arteriosus contributing to the ascending aorta. Normally a portion of the right fourth aortic arch involutes, leaving the standard leftward aortic arch. The remaining components of the right fourth arch contributes to the development of the right subclavian artery. The ventral portion of the sixth arch interacts with the lung bud to form the remaining pulmonary artery. The dorsal right sixth aortic arch involutes, whereas the dorsal left sixth aortic arch forms the ductus arteriosus. The left subclavian artery forms from the left intersegmental artery. The descending thoracic aorta is formed from the left dorsal aorta (Fig. 1).

This complex interaction between the involution or programmed cell death, and migration or persistence of the various portions of the aortic arches and dorsal or ventral aortae results in variant anatomy. The outcome can range from an inconsequential variant to life threatening with partial or complete vascular rings causing severe tracheal compression. Aortic arch anomalies producing tracheoesophageal constriction account for 1% to 2% of all congenital heart defects.

Diagnosis

Significant airway compression may occur owing to such pathologic vascular anomalies or even from underlying congenital heart disease and cardiomegaly itself. Airway compression is often unrecognized on prenatal ultrasound imaging. Neonates can present with dyspnea, wheezing, and stridor that may be life threatening. There may also be symptoms of feeding difficulties and dysphagia with cardiovascular-related compression. As a general rule, neonates present with airway issues, whereas dysphagia tends to occur more widely in older children and adults.

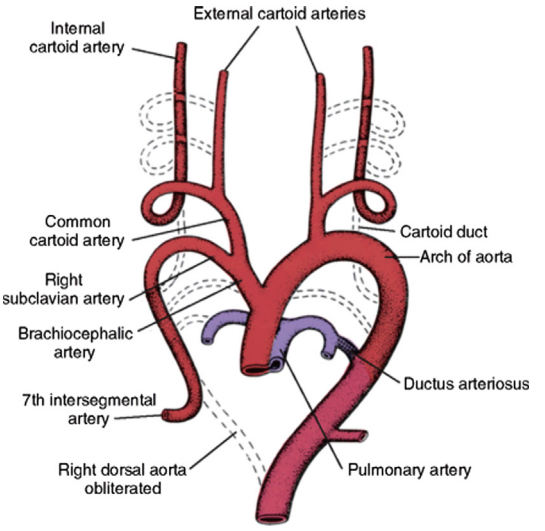

Fig. 1. Normal left aortic arch development from the 6 paired aortic arches that connect the dorsal and ventral aortae. The opaque arches represent normal programmed cell death and involution.

Evaluation of the neonate or child with suspected tracheobronchial compression may include anteroposterior and lateral chest radiographs, barium esophagram, echocardiogram, computed tomography (CT) angiography (CTA) or cardiac MRI, and direct microlaryngoscopy and bronchoscopy. When fine resolution of the airway is not necessary, cardiac MRI is preferred to CTA as because it avoids ionizing radiation in children.[1,2] A disadvantage of MRI is the significant time needed for image acquisition and the necessity to provide sedation, especially in the neonate and infant. A normal frontal and lateral chest radiograph may increase the likelihood of finding a vascular ring, because the vast majority of vascular rings are associated with a right aortic arch.[3] Microlaryngoscopy and bronchoscopy can also help to identify synchronous primary airway lesions that may be present, as well as help document vocal cord motion before cardiovascular surgery. Often it is helpful in distinguishing tracheomalacia from true compression.

By far, the majority of vascular rings are associated with a right-sided aortic arch. The most prevalent vascular anomalies that result in airway or esophageal compression are (in decreasing order of frequency): (1) double aortic arch, (2) right aortic arch with aberrant left subclavian artery originating from a retroesophageal diverticulum (diverticulum of Kommerell), (3) innominate artery compression,