Vascular Abnormalities

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Although extrinsic compression of the trachea or a bronchus resulting from an abnormal vascular structure is uncommon, it is an important cause of serious, persistent respiratory problems in infancy. Since the aorta, brachiocephalic vessels, and pulmonary arteries are closely related to the trachea and bronchi, minor aberrations of these vascular structures may produce extrinsic compression of the respiratory tract. A number of different anatomical vascular abnormalities may cause airway compromise. During the past 10 years, 26 patients with various vascular abnormalities has surgical intervention to alleviate airway obstruction at the Children's Hospital of Pittsburgh. Twenty-one were caused by aortic arch anomalies and the remaining 5 were related to pulmonary arterial anomalies (Table 1).

Table 1. Surgical Experiences with Vascular Abnormalities at the Children's Hospital of Pittsburgh (1969 to 1980)

Type of Anomalies	No of Patients
Double aortic arch	13
Both arches patent	9
One arch atretic	4
Aberrant innominate artery	4
Right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum	3
Left aortic arch with right descending aorta and right ligamentum arteriosum	1
Distal origin of the left pulmonary artery	2
Tetralogy of Fallot with absent pulmonic valve	3
Total	26

Symptomatology

The clinical manifestations of these vascular abnormalities may vary considerably depending on the type of lesion and severity of encroachment of trachea, bronchi, or esophagus. The most common presentation is the picture of airway obstruction, mainly stridor, wheezing, or excessive secretion. In the newborn period respiratory symptoms are usually mild and rarely recognized. However, symptoms usually become apparent during the first few months of life. These patients often have a history of recurrent respiratory embarassment, or infection, requiring frequent medical attention. Some have been referred for evaluation of a possible allergic problem such as asthma. Unless there is severe airway compromise, symptoms usually vary from time to time. During sleep and quiet moments some patients show little sign or respiratory difficulty. However, their symptoms are frequently exacerbated by crying or exertion. Upper respiratory infection also may trigger further compromise of the airway as a result of inflammatory changes in the tracheobronchial lumen.

Although dysphagia with solid food ingestion may be seen in some patients with large and/or tight retroesophageal vascular compression, it is a rather uncommon manifestation in the pediatric age grou. Some patients may have excessive oropharyngeal secretions that result in frequent aspiration pneumonia. Despite chronic respiratory difficulties, the majority of patients with vascular abnormalities maintain an adequate weight gain. Only those with frequent pulmonary infection and/or feeding problems may have problems of failure to thrive and poor development.

Types of Vascular Abnormalities

Double aortic arch is the most common vascular abnormality requiring operation. In this condition, the trachea and esophagus are completely encircled by the bilateral aortic arches owing to persistence of both right and left embryologic fourth aortic arches. Both aortic arches may be patent, or may be atretic and remaining as a fibrotic cord. In either condition, it usually causes marked tracheal compression with severe respiratory symptoms in early infancy.

In patients with a right aortic arch, an aberrant left subclavian artery and left ductus or ligamentum arteriosum is a rather common abnormality. This often forms a loose vascular ring, and thus a relatively small portion of the patients with this anomaly have significant encroachment upon the trachea or esophagus which requires surgical intervention. In some patients with this anomaly, significant dysphagia may develop when solid food is added. We have also seen a case of rare aortic arch anomaly consisting of a left aortic arch and right descending aorta with a right ligamentum arteriosum which caused tracheal compression.

Pulmonary sling (distal origin of the left pulmonary artery or vascular sling) is a rare but serious congenital anomaly in which the left pulmonary artery arises abnormally from the right pulmonary artery. It then takes a course between the trachea and esophagus, resulting in severe encroachment of the proximal right main bronchus. Patients with this condition usually develop respiratory symptoms early in infancy and manifest persistent wheezing and stridor.

Various congenital cardiac lesions, primarily those with a large left to right shunt and a dilated pulmonary artery, may cause external compression of the trachea and bronchial tree and the resultant symptoms of airway obstruction. Of particular interest, patients with tetralogy of Fallot and absent pulmonary valve usually have a markedly dilated pulmonary artery which frequently causes severe airway obstructive symptoms.

Another important cause of airway obstruction by a vascular structure is an aberrant innominate artery. Some patients in whom origin of the innominate artery is from the aortic arch distal to the midline and posteriorly oriented may have compression along the anterior aspect of the trachea. On occasion, the innominate artery may arise abnormally from the aortic arch as a common trunk with the left carotid artery. This anomaly may also cause anterior compression of the trachea. Although clinical symptomatology is quite variable, the tracheal compression can be severe enough to cause apneic spells and even necrosis of the trachea. According to a large series from the Hospital for Sick Children in Toronto, only a small percentage of patients with this abnormality require surgical intervention and the majority of patients improve spontaneously with age.

Diagnostic Approach

Although there are a number of causes of chronic stridor in infancy and childhood, the possibility of a vascular abnormality causing extrinsic airway compression should be kept in mind in the differential diagnosis. *A chest roentgenogram with barium filling the esophagus is the simplest and yet the most informative procedure in the evaluation of patients who have airway symptoms and are suspected of having a vascular abnormality.* Since most patients with respiratory problems often have already had chest roentgenograms, careful review of the films may be helpful initially to evaluate tracheal or bronchial abnormalities such as an abnormal shift or constriction. If dysphagia and aspiration problems coexist, a barium esophagram with simultaneous fluoroscopic examination is indicated. However, a simple four view cardiac series is often sufficient to evaluate this condition. The radiographic studies facilitate determining the situs of the aortic arch. An abnormal indentation of the esophagus caused by an abnormal vessel is also readily recognized. Most aortic arch anomalies causing vascular rings have a posterior esophageal indentation. An anterior esophageal indentation is typical of a pulmonary vascular sling but on rare occasions an aberrant subclavian artery may also pass between the trachea and esophagus. Patients with aberrant innominate artery usually have a normal barium esophagram. However, in these patients an anterior indentation of the trachea 1 to 2 cm above the tracheal bifurcation can be seen on the lateral view of chest and neck roentgenograms.

Although an abnormal esophagram frequently provides diagnostic information as to the presence of a possible anatomic derangement, several different anomalies may show similar esophagraphic findings. Therefore precise anatomic diagnosis cannot be certain without angiographic studies. Tonki et al have advocated simultaneous angiographic and barium esophagram studies during the catheterization to evaluate the anatomic relation between the blood vessels and the esophagus. We have found that leaving a small radiopaque nastogastric line, such as a Swan Ganz catheter, in the esophagus during the study is helpful to determine spatial relationships since the trachea is located just anterior to this esophageal line. At the termination of the study, the balloon can be inflated with contrast material and slowly withdrawn from the stomach to the oropharynx to evaluate the site and severity of esophageal narrowing. A cine-fluoroscopic record of this catheter withdrawal is made to correlate the site of esophageal narrowing with angiographic information.

Certain vascular structures such as an atretic aortic arch segment or a ligamentum arteriosum cannot be visualized by angiography. If tenting or distortion of the brachiocephalic vessels or a blind aortic diverticulum in the proximal portion of the descending aorta is observed, then an atretic aortic arch and/or ligamentum on the contralateral side of the descending aorta (and thus a possible vascular ring) should be suspected.

Biplane angiography with multiple views using selective injection in the vessel suspected of causing compromise is essential for adequate evaluation of these abnormalities. Although the morbidity of bronchoscopy in the hands of a skilled otolaryngologist is insignificant, this procedure should be reserved for patients with special indications or diagnostic problems since it might exacerbate symptoms in patients with a compromised airway. For instance, bronchoscopy in the patient with an aberrant innominate artery is helpful to demonstrate a discrete pulsatile compression on the anterior aspect of the trachea, 1 to 2

cm above the carina. Tracheobronchographic studies in these patients should be discouraged in view of their associated high morbidity.

Management

Simple existence of a vascular ring is not an indication for surgical intervention. However, when airway compromise from an abnormal vascular structure is significant, surgical intervention should be undertaken without delay. A double aortic arch invariably requires surgery in early infancy, and in these patients the smaller or atretic arch should be divided. In patients with a right aortic arch, aberrant left subclavian artery, and left ductus (or ligamentum) arteriosus, division of the ductus usually relieves airway symptoms. Occasionally division of the aberrant subclavian artery is necessary to alleviate dysphagia. In the case of distal origin of the left pulmonary artery, the left pulmonary artery is divided at its origin from the right pulmonary artery and anastomosed to the main pulmonary artery using cardiopulmonary bypass. An aberrant innominate artery that causes respiratory compromise can usually be suspended against the sternum with a satisfactory postoperative result.

Postoperative recovery in these patients is considerably variable. Although prompt symptomatic improvement may occur shortly after surgery, persistent respiratory symptoms are not unusual in many patients. Some patients may have such significant damage or distortion of the tracheobronchial tree owing to chronic compression by an abnormal vascular structure that severe respiratory symptoms may persist postoperatively. In some of these patients severe associated tracheomalacia may require a tracheal graft with autologous rib in order to alleviate life-threatening respiratory problems.

The prognosis for survival is generally good in most patients with aortic arch anomalies. However some patients, especially those with distal origin of the left pulmonary artery, may succumb before the diagnosis is made. Patients with this anomaly frequently have associated cardiac as well as noncardiac and additional tracheal abnormalities, and as a result their prognosis is poor.

Increased awareness of these vascular abnormalities as a cause of respiratory problems in infants and children will prompt appropriate diagnostic and surgical management and even further improve the morbidity and mortality.