Vascular Ring and Sling, Diagnosis and Management

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ABSTRACT

Setting: Queen Alia Heart Institute-Amman -Jordan
Aim: The aim of the study is to present our experience in the diagnosis and the management of various kinds of vascular ring encountered in our institution over a ten year period.
Patients and methods: A total of 21 children operated between 2000 and 2010 with complete vascular ring were identified. Their medical charts were reviewed. Data regarding clinical presentation, age at diagnosis, anatomy, investigations, surgical interventions and postoperative outcome were evaluated.
Results: There were13 boys and 8 girls. The patients ranged in age from two months to one year and averaged five months. The severity of compression determines the severity of symptoms. 95.0% of patients have persistent respiratory difficulty, with only one patient presented with recurrent vomiting, and mild respiratory difficulty. Of the 21children: 12 had double aortic arches ,7.0 had a right aortic arch with left ligament arteriosum or small patent ductus arteriosus,and 2 had a pulmonary sling (the left pulmonary artery arising from the right pulmonary artery) .
Conclusion: The airways of the infant may be compressed by a variety of vascular anomalies.When a vascular ring is suspected from history and physical examination, routine chest radiograph and barium swallow can be performed with a high yield for the presence of a ring. CT angiography is a preferable modality of diagnosis for vascular ring and sling, as an alternative to conventional angiography. All vascular rings that cause symptoms should be surgically corrected.
Key words: Stridor, ring, sling, trachea.

INTRODUCTION

Vascular rings are congenital anomalies of the aortic arch system where vascular structures encircle and compress the trachea and esophagus. These rings are subclassified as double aortic arch, right arch/left ligamentum, pulmonary artery sling, and innominate compression. Double aortic arch is conveniently grouped into 3 categories, dominant right arch, dominant left arch, and balanced arches[1].
The embryonic development of vascular rings was detailed by Edwards[2]. A simplified version of the development of these anomalies is shown in (Figure 1). In the embryonic aortic arch system, the ventral and dorsal aorta are connected by six primitive aortic arches. The first, second, and fifth arches involute to form Edward's classic double aortic arch. If the right fourth arch involutes, a normal left arch is formed. If the left fourth arch involutes, a right aortic arch is formed[3].

The normal aortic arch crosses over the left mainstem bronchus, (figure 2). In the vascular ring malformations diagrammed in (figures 3 and 4), the aortic arch passes over the right mainstem bronchus instead. This is an abnormal right sided aortic arch. In (figure 3) (the double aortic arch), the aortic arch bifurcates such that one half of the aorta crosses over the left mainstem bronchus, while the other half of the aorta crosses over the right mainstem bronchus. In (figure 4) (the right sided aorta with anomalous left subclavian), the aortic arch crosses only over the right mainstem bronchus.

The other two tracheoesophageal compression syndromes that have been included under the heading, vascular ring, are innominate artery compression syndrome (IACS)[4] and pulmonary artery(PA) sling sling[5]. IACS occurs when there is an abnormally distal and posterior takeoff of the innominate artery and it compresses the trachea anteriorly as it courses from the left of the mediastinum to the right arm. A PA sling occurs when the left pulmonary artery (LPA) originates from the right pulmonary artery (RPA) and encircles the distal trachea and right mainstem branches as it courses between the trachea and esophagus to the left lung.(figure 5).

PATIENTS AND METHODS

A total of 21 children operated between 2000 and 2010 with complete vascular ring were identified. Their medical charts were reviewed. Data regarding clinical presentation, age at diagnosis, anatomy, investigations, surgical interventions and postoperative short-term outcome were evaluated. Vascular rings were classified according to the nomeclature established by the Congenital Heart Surgery Database Committee. Chest radiography, Barium oesophagography and two-dimensional echocardiography were performed in all patients. Computed tomography (CT) with three dimensional (3D) reconstructions were performed in 8.0 patients. Cardiac catheterization and angiography were performed in 13 patients, and bronchoscopy in 2.0 patients

Operative technique: Transpleural left postero-lateral thoracotomy was the preferred approach in all patients except the two patients with pulmonary slings. In case of double aortic arch, the ligature of the anterior/right arch was performed, followed by ligation of the ductus arterisus. In cases of a double aortic arch that was right arch dominant or a right aortic arch with left ligament or patent ductus arteriosus (PDA), a section of the left arch or ligament (or PDA) was performed and mediastinum masses were released in order to relieve esophageal and tracheal compression. In the two patients with pulmonary sling approached through midsternotomy

RESULTS

There were 13 boys and 8.0 girls. The patients ranged in age from two months to one year and averaged five months. The presenting symptoms and types of anomaly are summarized in tables 1,2.

Chest X-ray, varying degrees of atelectasis, pneumonic infiltrates, and hyperaeration were common while the presence of a right aortic knob in the chest radiograph, raised the suspicion of a right aortic arch and extrinsic tracheal compression in few patients. The site and extent of esophageal compression were visualised by barium swallow, where as tracheal compression were clearly visualized by contrast-enhanced CT with 3D reconstruction in 8 patients, aortic root angiography in 13 patients and bronchoscopy in two patients.

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Table 1. Clinical presentation of vascular ring & sling

Stridor	20(96%)
Noisy Breathing	15(55%)
Brassy Cough	12(67%)
Choking episodes	11(52%)
Wheezes(misdiagnosed asthma)	16(74%)
Interrupted Feeding	13(63%)
Recurrent chest infection	8(37%)
Attacks of cyanosis	9(44%)
Recurrent vomiting	7(33%)
Apnea episode	3(14%)

Table 2.Typesof vascular rings

Type	number	Associated cardiac anomaly(no.-%)
Double Aortic Arch	12	-ASD (1) -VSD (1) *17%*
Right aortic Arch	7	TOF(1)- DCRV(1) *28%*
Pulmonary sling	2	*0%*

ASD:Atrial septal defect.
VSD:ventricular septal defect.
TOF:teralogy of Fallote
DCRV :double chamber right ventricle.

Figure 1:A simplified version of the development of vascular ring anomalies.

Figure 2: normal aorta.

Although echocardiography is a useful and safe technique for the diagnosis of vascular anomalies, contrast-enhanced CT with 3D reconstruction angiography is usually necessary for definitive anatomical evaluation, especially before surgery

Of the 21 children: 12 had double aortic arches, 7.0 had a right aortic arch with left ligament or left PDA, and 2 had a pulmonary artery sling. In patients who had double aortic arches, the posterior arch was the dominant arch in all patients.

Figure 3 - Double aortic arch.

Figure 4 - Right sided aorta with anomalous left subclavian artery.

Fig 5. The anomalous left pulmonary artery (*LPA*) is seen arising from the right pulmonary artery (*RPA*) and coursing between the esophagus and the trachea

Figure 6.This esophagram demonstrates extrinsic compression of the esophagus consistent with a vascular ring

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DISCUSSION

The phrase "vascular ring" was first used by Dr. Robert Gross in his report describing the first successful division of a double aortic arch in 1945.[6]In that manuscript, Gross recalled his observations at the time of an autopsy he performed in 1931 on a five-month old baby who had wheezing respiration since birth.

Vascular rings and pulmonary slings are very important but rare causes for common respiratory symptoms. Children with vascular rings are often diagnosed during infancy. Presenting symptoms are usually due to tracheal or esophageal compression such as stridor, noisy breathing, frequent respiratory infections, wheezing, interrupted feeding, choking, apnea, wheezing, croupy cough, hoarse cry, etc[7]. A typical presenting history is that of noisy breathing since birth. It is common for these infants to have more

severe symptoms during a respiratory infection. A young infant with a history of "asthma" is another typical way that these patients present[8,9]. Sometimes these children are diagnosed with cardiac disease, but the vascular ring may be occult until an appropriate imaging study is performed.

In infants, the most common cause of stridor is laryngomalacia. In addition, children with congenital tracheal stenosis can present with symptoms similar to children with anatomically complete vascular rings such as: stridor, retractions, dyspnea, apnea, and respiratory distress. The severity of symptoms depends on the degree of compression on the trachea and esophagus.

The interesting fact of our group of patients is that all were infants with respiratory symptoms predominate in initial presentation.These symptoms occur from tracheobronchial compression from the vascular ring or pulmonary sling. In this study 95% of patients have respiratory symptoms, with only one patient presented with recuurent vomitting, with mild respiratuory difficulty. Tracheal compression causes airflow obstruction and decreased mucociliary clearance of secretions, leading to recurrent bronchopulmonary infections.[(10)]Reflex apnea is hypothesized to be a type of respiratory arrest that occurs when vagal afferent nerves are stimulated[10,11,12].

An even, charachteristic finding in some cases, is that stridor and other respiratory symptoms were more pronounced during feeding or activity. Difficulty in swallowing and régurgitation are likely to set off an attack of coughing, stndor, difficulty in breathing and cyanosis. Esophageal compression by the DAA causes it to bulge forward, indenting the membranous trachea and exacerbating respiratory distress. Esophageal compression also leads to regurgitation and aspiration pneumonia[10,11,12].

The age at presentation ranged from one month to 12 months.Detailed history revealed the onset of symptoms appear shortly after birth, may be relatively mild or severe, and worsened when the infant lying supine.Some of the patients have a delay in diagnosis due to attribution of symptoms to other more common etiologies**.** The absence of response to treatment, was the leading cause for referral to cardiology department. Tracheobronchial compression by vascular structures in childhood is uncommon and may be masked by non-specific respiratory symptoms. Early investigations to look for airway and high index of suspicion are essential to confirm the vascular compression of trachea[13].

DAA is the most frequently encountered vascular ring malformation (57%) that inevitably completely encircles the trachea and esophagus. The DAA, together with the right aortic arch with left ligamentum (33%), and pulmonary sling 10%, is defined anatomically as the complete vascular ring.

A ring with double-arch anatomy tends to have more severe symptoms and to present earlier than other types of vascular rings.[(14,15,16)]In this tudy, patients with a double aortic arch had earlier onset of symptoms (1-3 months)than the patients with a right aortic arch and left ligamentum(>6 months).compression of the trachea and esophagus from double aortic arch appeares to be more severe than that with other types of vascular rings.However we have a patient with right aortic arch who have earlier and more severe presentation. The severity of compression and the onset of symptoms are determined by the space between the tracheoesophageal axis and the components of the ring, and tracheoesophageal dimensions[12].

One type of vascular ring, the pulmonary artery (PA) sling, is very commonly (almost 50% of cases) associated with congenital tracheal stenosis. As bronchoscopy has become more widely applied, two patients have undergone bronchoscopy preoperatively. However none of the two patients had a tracheal stenosis.

Patients with PAS have clinical symptoms secondary to tracheal compression and, to a lesser degree, esophageal compression. The two patients with PAS have acute episodes of near lethal dyspnea and cyanosis and symptoms present in early infancy. Reflex apnea is hypothesized to be a type of respiratory arrest that occurs when vagal afferent nerves are stimulated.[(10,11,12)] The cause of near lethal dyspnea and cyanosis as in these cases should not be given undue prominence when dealing with this rather common problem in early infancy. So many other causes are far commoner. However, the remediable obstruction in these children does suggest that when dyspnea and cyanosis are severe and a point of obstruction can be demonstrated the infant deserves an appropraite investigation.

Radiology has played a crucial role in delineating vascular causes of airway obstruction in infants, from the 1940s with conventional radiographs and barium esophagrams, through the 1960s–1970s with angiography, to the 1980s and 1990s with CT and MR Imaging. These modalities have facilitated more precise diagnosis and thus led to earlier treatment and better outcome[17].

In the early time period of this study, barium esophagram and aortic angiography was often the only diagnostic procedure performed before surgical intervention. Frequently, the initial diagnosis of a vascular ring is made by a contrast study that shows an indentation of the posterior esophagus (figure 6). The

diagnosis of our patients was also first suspected by barium esophagography findings. Barium esophagography has been advocated as a valuable investigation for patients with suspected vascular rings, because it can be safely and rapidly performed to diagnose DAA with high sensitivity. However, this method is no longer considered a sufficient evaluation of the patient before proceeding with treatment by vascular ring repair[10]. In PAS a marked indentation of the anterior wall of the esophagus was seen just below the aortic arch with no posterior displacement[18].

Although echocardiography is often the initial diagnostic method used in patients with suspected congenital cardiovascular disease. In our cases echocardiography appears to be of little value in the diagnosis of a vascular ring, but is essential to exclude associated cardiovascular malformations. The suprasternal notch approach can image the area of interest in the near field. In the hands of experienced investigators, transthoracic echocardiography permits an accurate imaging of DAA and any associated congenital heart diseases as well as clear identification of the sidedness of the aortic arch. The major problem resides in its limited ability to image clearly the atretic aortic arch structures and a ligamentum arteriosum. Nonetheless, echocardiography remains a powerful, useful, and non-invasive tool that is capable of evaluating patients with suspected DAA**[19].**

Conventional angiography is an invasive technique that has several disadvantages including long procedure time, need for sedation, arterial puncture and rare potential complications such as dissection and occlusion. With advances in imaging technology, the introduction of multidetector row CT (MDCT) has dramatically expanded the applications of CT in the evaluation of vascular and airway diseases Three-dimensional images allow excellent display of vascular anomalies that can be used as a vascular road map by surgeons. CT angiography is a preferable modality of diagnosis for arterial disease as an alternative to conventional angiography, because it is safer, less time consuming and also more cost-effective. In addition to displaying vascular anatomy, thoracic CT angiograms provide information about both airway and lung parenchyma, which is important in patients who have thoracic vascular anomalies[19].

Operative risks are relatively small. Intuitively, early recognition and surgical treatment of symptomatic vascular rings may allow the greatest possibility of normal growth of the trachea, esophagus, and relief of aerodigestive symptoms; however, when severe early airway symptoms are present, this may be indicative of a worse pathology and patient substrate. After surgical treatment and ring division, all patients showed a significant improvement in symptoms. However, some patients with conventional ring division will have long-term issues of the aerodigestive tract[20]. Fortunately all of our group of patients became free of symptoms after surgery, which is probably due to early recognition and surgical division of the rings. While only few patients in our group

CONCLUSION

The airways of the infant may be compressed by a variety of vascular anomalies. When a vascular ring is suspected from history and physical examination, routine chest radiograph and barium swallow can be performed with a high yield for the presence of a ring. CT angiography is a preferable modality of diagnosis for vascular ring and sling, as an alternative to conventional angiography. All vascular rings that cause symptoms should be surgically corrected.

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