Double Aortic Arch Repair: An Interesting Case Scenerio

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Abstract:
The term “VASCULAR RINGS” refers to any vascular anomaly causing tracheoesophageal compression. Double aortic arch is the most common of the complete vascular rings, causing tracheo-esophageal compression. We present this rare case report of a double aortic arch with dominant right aortic arch, presenting with extrinsic trachea-bronchial obstruction. A 11-month-old female child was referred to our institution with stridor since 1 month of age and recurrent respiratory infection. Chest x-ray showed no obvious indentation of the tracheal shadow. Echocardiographic evaluation showed double aortic arch with dominant right arch and right descending aorta, causing vascular ring and tracheal obstruction. Patient was further evaluated with cardiac catheterization and angiography which revealed double aortic arch. The patient underwent division of the left aortic arch through a left postero-lateral thoracotomy approach. The left aortic arch was divided at its junction with the descending aorta and the cut edges were oversewn.

In summary, vascular rings, more commonly the double aortic arch, are an important cause of tracheo-esophageal compression, the clinical suspicion and diagnosis of which can lead to early surgical intervention and relief.

Keywords:
Vascular Rings; Double Aortic Arch; Thoracotomy

1. INTRODUCTION

Aortic ring, "bifid aortic arch," and "split aortic arch" are names given to a congenital malformation where, as a result of persistence of both fourth branchial arches, the aorta, instead of being a single tube, divides into two, one branch going in front of the trachea, the other behind and almost invariably behind the oesophagus as well, the two limbs uniting again to form one descending aorta. Complete vascular rings represent about 0.5-1% of all congenital cardiovascular malformations. The majority of these are double aortic arches. There is no known gender preference, i.e. males and females are about equally affected. There is also no known ethnic or geographic disposition. Associated cardiovascular anomalies are found in 10-15% of patients. This is a case report of a double aortic arch with dominant right arch, presenting with extrinsic tracheo-bronchial obstruction. We present this case because it is a rare
presentation which is not seen very often in general practice and can be kept in mind when diagnosing a pediatric case presenting with respiratory distress.

2. CASE REPORT

A 11-month-old female child was referred to our institution with stridor since 1 month of age and recurrent respiratory infection. She presented with non improvement of respiratory symptoms requiring mechanical ventilation. Patient was being treated by her physician for respiratory complaints. Our pediatric cardiologist examined and further evaluated her. Chest x-ray showed no obvious indentation of the tracheal shadow. Echocardiographic evaluation showed nearly closed down ventricular septal defect and double aortic arch with dominant right arch and right descending aorta. Patient was further evaluated with cardiac catheterization and angiography which revealed double aortic arch, with right arch giving rise to left carotid, right carotid and right subclavian, while left arch giving rise to left subclavian with both arches widely patent (Figure 1). The patient underwent division of the left aortic arch through a left posterolateral thoracotomy approach. The ligamentum arteriosum was attached to the left aortic arch and the left pulmonary artery. The ligamentum was divided (Figure 2).

![Catheterization picture of double aortic arch.](image)

The anterior left aortic arch was clamped just proximal to its junction with the descending aorta. There was no decrease in femoral arterial blood pressure, no gradient between the radial and the femoral arterial blood pressures, and both radial and carotid pulses were palpable. The anterior left aortic arch was divided at its junction with the descending aorta and the cut edges were oversewn (Figure 3).
The mediastinal tissue around the aortic arches was dissected, allowing the left aortic arch to retract. Strands of tissue around the trachea and esophagus were divided to relieve any potential residual compression or fibrosis.

The child was extubated on the second postoperative day. The child was stable and comfortable without any clinical evidence of respiratory obstruction. She was transferred to the ward on the third postoperative day. The remaining postoperative recovery was uneventful, with no respiratory or feeding problems.
Complete vascular rings represent about 0.5-1% of all congenital cardiovascular malformations. The majority of these are double aortic arches. There is no known gender preference, i.e. males and females are about equally affected. There is also no known ethnic or geographic disposition. Associated cardiovascular anomalies are found in 10-15% of patients. In 1945 Gross accomplished the first successful surgical intervention for a Double Aortic Arch [1]. The development of vascular rings was detailed by Edwards. In the embryonic aortic arch system, the ventral aorta is linked by six pairs of arches to two dorsal aortae, which join to form the single dorsal aorta. The right fourth arch usually involutes, leaving a normal left arch. Left fourth arch involution results in a right aortic arch. Classical Double Aortic Arch anatomy develops when both fourth arches persist.

Anatomically, the ascending aorta arises normally and, as it exits the pericardium, it divides into two – right and left aortic arches which encircle the trachea and the esophagus and reunite posteriorly to form the descending aorta. Hypoplasia of one of the aortic arches is common with one arch, more commonly the right aortic arch, being dominant. Atresia can be uncommonly present in any of the segments of either of the aortic arches, resulting in various subtypes of double aortic arch with atresia [2]. The posterior descending aorta formed by the union of the two aortic arches can be on the left or on the right of the thoracic vertebrae. A right dominant double aortic arch has been more commonly observed with left descending aorta and vice versa. The descending aorta has a tendency to be more midline than normal. This abnormal position can cause compression of the airway between the pulmonary artery and the descending aorta postoperatively, leading to persistence of symptoms following surgery [3]. The ductus or ligamentum arteriosum, which is not a part of the vascular ring, runs between the left pulmonary artery, inferior to the junction between the left aortic arch and the descending aorta [4]. The aortic arches give rise to the ipsilateral common carotid and subclavian arteries and the innominate artery is absent.

Patients with double aortic arch can be asymptomatic or present with symptoms ranging from non-specific complaints to life threatening respiratory distress. The symptoms of stridor, dysphagia, a “bark” chronic cough, susceptibility to bronchopneumonia, head retraction, malnutrition, onset during early infancy, and an increase of respiratory distress during feeding were described by Wolman et al [5]. An esophageal foreign body at the site of compression by the vascular ring is rarely the presenting feature of double aortic arch. Life threatening episodes of respiratory arrest and apnea have also been described [6].

Conventional chest x-ray may show indentation of tracheal shadow, retro tracheal opacity, and anterior tracheal bowing. Specific radiological signs have been described for barium esophagography [7]. These include bilateral persistent extrinsic compressions of esophagus in AnteroPosterior view, with the dominant arch causing a deeper and superior indentation and a deep posterior indentation in lateral and oblique views. Barium swallow is diagnostic in the majority of cases. However, Computed Tomography and Magnetic Resonance Imaging have become increasingly utilized in the diagnosis and evaluation of aortic arch anomalies, including double aortic arch [8]. The assessment of the arch dominance and surrounding tissues in the mediastinum has improved with these radiological modalities, especially when the barium swallow is negative, there is innominate artery compression, or in complex cases. The four-vessel sign in the superior mediastinum can be seen due to the presence of separate subclavian and common carotid arteries on both sides. Presently angiography is rarely indicated or necessary for adequate evaluation of aortic arch anomalies. Echocardiography is recommended to rule out associated congenital cardiac defects [6, 8–10].

Repair is achieved through a left posterolateral thoracotomy approach, especially those with a dominant
right aortic arch [6, 8, 9, 11]. The Mayo Clinic recommends a right posterolateral thoracotomy approach in patients with left aortic arch, right-sided descending thoracic aorta, and right ductus or ligamentum arteriosus; a double arch with atresia of the right posterior segment; or when anastomosis of an aberrant right subclavian artery to the ascending aorta is performed [12]. Backer and Mavroudis [13] recommend that innominate artery compression be approached from the right side, with suspension of the innominate artery to the sternum. Repair through a median sternotomy is recommended when concomitant repair of intracardiac defects is performed [10, 12].

The principles of surgery are essentially the same as described by Gross [2]. The surgical repair includes adequate dissection of the aortic arches and descending aorta, division of the non-dominant aortic arch, division of the ductus or ligamentum arteriosum, and the dissection and division of the mediastinal adventitial bands that may compress the trachea or esophagus.

Post-operative complications include bleeding, vocal cord paralysis, pneumonia, pneumothorax, chylothorax, feeding difficulties, and residual respiratory obstruction. Residual respiratory complaints have been noted in up to 54% of patients [6, 9, 11]. While the residual respiratory symptoms are usually due to tracheobronchomalacia, anatomic compression of the trachea by the postoperative arch and the anterior remnant of the divided arch, or by midline descending aorta, can be present [14]. Due to the presence of the dual-sidedness of the balanced aortic arches, these patients have a more midline descending aorta. This results in abnormal stacking of the structures anterior to the spine and leads to extrinsic compression of the left main bronchus between the midline descending aorta posteriorly and the pulmonary artery anteriorly.

In summary, vascular rings, more commonly the double aortic arch, are an important cause of tracheoesophageal compression, the clinical suspicion and diagnosis of which can lead to early surgical intervention and relief or avoidance of immediate or long term respiratory complications.

References


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