VASCULAR RING- LEFT AORTIC ARCH WITH RIGHT DESCENDING THORACIC AORTA AND RIGHT LIGAMENTUM ARTERIOSUM

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ABSTRACT

BACKGROUND
The Gerbode left ventriculo-right atrial defect is a very rare congenital anomaly. Gerbode described two routes for blood to travel from the left ventricle to the right atrium, a direct or supravalvular or (type A) and an indirect (type B) or infravalvar. Case Characteristics- A male born to nonconsanguineous couple and had indirect type C Gerbode VSD. Observation- The child had midmuscular VSD. Blood flow from left ventricle to right ventricle through midmuscular VSD and then through tunnel formed by tricuspid to right atrium. Outcome- Parent advised to go for further investigation. Message-A rare congenital malformation to be suspected when any child with midmuscular VSD have tricuspid regurgitation.

KEYWORDS
Gerbode Left Venticulo-Right Atrial Defect, Vascular Ring, Left Aortic Arch, Ligamentum Arteriosum.


BACKGROUND
The vascular ring is a congenital condition in which the anomalous configuration of the aortic arch, its associated vessels or both forming a complete or incomplete ring around the trachea and/or oesophagus. Several other related vascular anomalies involving arch vessels do not form a complete ring, but have been grouped with vascular rings because they produce similar symptoms related to compression of the trachea or oesophagus. The first vascular ring described was a double aortic arch noted by Hommel in 1737. Bayford reported retrooesophageal right subclavian artery in 1794 after performing an autopsy on a woman who had experienced dysphagia for years and died of starvation. Maude Abbott described five cases of double aortic arch in 1932 and made the suggestion that surgical intervention should be undertaken in such cases. Finally, in 1945, Gross used the term vascular ring after performing the first successful division of a double aortic arch. Since that time, numerous reports of successful treatment have been reported. We are reporting a case because of its rarity.

CASE REPORT
A female child delivered by normal vaginal delivery at 38 weeks of gestation to fourth gravida women. She had nonconsanguineous marriage and uneventful antenatal course. Baby had delayed cry at birth required bag and mask ventilation for resuscitation. The baby was admitted in NICU. Her birth weight was 2.4 kg, head circumference 32.5 cm and length 44cm. She had an episode of tonic-clonic convulsion at twelve hours of life and responded to parenteral phenobarbitone. On fifth day of life, baby developed respiratory distress, feed intolerance, became lethargic and developed systolic murmur in left third and fourth intercostal space. Examination revealed normal anterior fontanelle, blood pressure was normal for age and SPO2 was 95% in all four limbs. Her x-ray chest PA view revealed cardiomegaly and ECG revealed RAD with RVH, normal for age. The child's septic screen was positive and was managed as early onset sepsis.

For cardiac evaluation, child was referred to paediatric cardiac clinic. While performing echocardiography, we noticed barking cough. This led to suspicion of tracheal vascular ring. Echocardiograph revealed muscular ventricular sepal defect with left to right flow, much early origin of relatively narrow left pulmonary artery from main pulmonary artery. Aortic arch was visualised from suprasternal and left infraclavicular area and also revealed normal ascending aorta with descending aorta going towards right side of the thorax, Figure 1.

CT thorax revealed normal ascending aortic arch (left ventricle) with right descending aorta and circumflex course of isthmus. The right carotid, left carotid, left...
subclavian artery and right subclavian artery were seen as first, second, third and fourth branch respectively with right ligamentum arteriosum. The right subclavian artery is narrow at its origin. The left pulmonary artery is relatively narrow as compared to right subclavian artery and originating much early from main pulmonary artery Figure 2, 3 and 4.

Figure 1. 2D Colour Doppler Study Depicting Muscular VSD, Narrow and Early Origin of Left Pulmonary Artery, Right Side Descending Aorta and Normal Both Coronary Artery

Figure 2. CT Thorax Revealed Normal Left Aortic Arch with Right Descending Aorta, Circumflex Course of Isthmus, Right Carotid, Left Carotid, Left Subclavian Artery and Narrowing of Right Subclavian Artery at its Origin as First, Second, Third and Fourth Branch of Aorta Respectively
DISCUSSION

The phrase "vascular ring" was coined by Robert E. Grossin 1945. Vascular rings are uncommon anomalies and make up less than 1% of all congenital cardiac defects. It occurs with about equal frequency in both sexes and had no geographical or racial predominance. Gross described the two types of vascular rings, true (complete) vascular rings and partial vascular ring (divided or split aortic arch). The true vascular ring makes up 85-95% of all cases. The two most common true or complete vascular rings are double aortic arch and right aortic arch with left ligamentum arteriosum. The other vascular rings that are extremely rare (<1%) include (a) Right aortic arch with mirror image branching and left ligamentum arteriosum and (b) A left-sided aortic arch with a retrooesophageal component, a right-sided descending aorta in conjunction with an anomalous right subclavian artery and a ligamentum arteriosum joining the origin of it to the right pulmonary artery. In type b anomaly, the first, second, third and fourth branch of arch are the right common carotid, which passes anterior to the trachea, the left carotid, the left subclavian artery and the right subclavian artery, respectively. The right subclavian artery is branch of the proximal right side descending aorta. The ligamentum arteriosum arises from the base of the right subclavian artery or a nearby diverticulum and travel towards the right pulmonary artery. The rightward tethering of the vascular structures that is aberrant right subclavian and right ductus/ligamentum may result in a "circumflex" course of the isthmus and proximal descending aorta. Berman et al described hypoplastic left pulmonary artery with such anomaly.

The various forms of conotruncal anomaly occur very early in embryologic development as a result of abnormal
or incomplete regression of one of the six embryonic branchial arches. The abnormal regression of right fourth aortic arch results in development of right subclavian artery from right 7th intersegmental artery (fourth branch of aorta). Abnormal retention of right dorsal aortic arch leads to right aortic arch and right ductus arteriosus or ligamentum arteriosum. The deletion of chromosome region 22q11 appears to be closely associated with these anomalies. Patients having such anomaly often have associated congenital cardiac defects while others may have vascular ring as isolated defect.1% of the general population and 40% of patients with trisomy 21 and CHD have right subclavian artery and 13-35% of patients with TOF and 8% of patients with TGA may have right side aortic arch.

No medical therapy exists for the definitive treatment of vascular rings. To avoid serious complications such as sudden death or significant tracheal or bronchial damage, surgery should not be delayed, especially in patients with symptoms of airway compression. Surgical division of a vascular ring is indicated in all symptomatic patients and should not be delayed in the presence of a respiratory tract infection, because the division of the ring allows more adequate and complete clearing of respiratory secretions.

**CONCLUSION**

Preoperatively, the patient should be given adequate nutritional support as well as general respiratory care and appropriate treatment of any respiratory tract infection. Most surgeons either approach by right thoracotomy or by midline sternotomy and laparotomy or by left thoracotomy with aim to divide arterial ligament to relieve the compression produced by the vascular ring.

Our patient had classic ascending left aorta, right descending aorta, aberrant right subclavian artery and ligamentum arteriosum (Figure 2, 3, 4). Because the ductus arteriosus was not patent, all the structures that contributed to the vascular ring could not be visualised on CT angiography. Patient was having persistent barking cough, respiratory distress and hypothetical and autopsy reports by Edwards, however, prompted us to the possibility of vascular ring. Thus, patient was referred to tertiary care paediatric cardiac center for surgery.

**REFERENCES**


