

Coarctation of a Right Aortic Arch

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ABSTRACT Right-sided aortic arch with a concomitant coarctation is an exceedingly rare congenital cardiac anomaly. We report of a 4-year-old boy who presented with a history of a stenotic bicuspid aortic valve who upon further evaluation was found to have a coarctation of a right-sided aortic arch. The frequency with which other anomalies exist in either of the above conditions requires thorough cardiac evaluation and detailed imaging. Surgical repair of this anomaly can safely be undertaken through a right thoracotomy. doi: 10.1111/j.1540-8191.2005.00123.x (*J Card Surg* 2006;21:261-263)

Coarctation of the aorta occurs in 0.2 to 0.6 per 1000 live births and accounts for 5% to 8% of all congenital cardiovascular malformations.¹ Coarctation may occur at any point from the transverse arch to the iliac bifurcation. However, 98% occur just below the origin of the left subclavian artery at the origin of the ductus arteriosus. Numerous concomitant cardiac processes have been associated with coarctation including subaortic stenosis, bicuspid aortic valves, mitral valve abnormalities, atrial and ventricular septal defects.

Right-sided aortic arch in the setting of normal cardiac situs is uncommon. In this anomaly, the aorta curves to the right and descends on the right side of the vertebral column. It occurs in less than 0.1% live newborns,² and like coarctation, there is an association with a wide variety of cardiac anomalies, specifically tetralogy of Fallot.

CASE REPORT

The combination of right-sided aortic arch with concomitant coarctation is extremely rare. We report the case of a 4-year-old boy who presented with a history of stenotic bicuspid aortic valve. He underwent balloon aortic valvuloplasty at 16 months of age at an outside facility. His valvular gradient improved significantly; however, he developed mild aortic insufficiency. In follow-up, he had a large blood pressure differential between his right upper extremity and the other three extremities, suggestive of coarctation. His vital signs were otherwise normal. On physical examination, he was a healthy, well-nourished, noncyanotic

young boy with a normal pulmonary exam. On cardiac examination, he had a normal first heart sound with a harsh, grade III/VI systolic ejection murmur. The murmur was heard best at the middle left sternal border and the second intercostal space on the right. A decrescendo grade II/VI diastolic murmur could also be heard at the left sternal border. Femoral pulses were diminished with warm distal extremities. His electrocardiogram was consistent with left ventricular hypertrophy. An echocardiogram demonstrated an aortic coarctation and aortic valve stenosis (gradient 45 mmHg). His left ventricular function and a left ventricular end-diastolic diameter were within normal limits. A subsequent cardiac catheterization demonstrated a 34 mmHg systolic gradient across the coarctation and a 19 mmHg gradient across the aortic valve. Normal coronary anatomy as well as a right-sided aortic arch with an aberrant left subclavian artery arising off the distal aorta was also noted (Fig. 1).

The patient was taken to the operating room for repair of the coarctation. Through a right thoracotomy (Fig. 2), the patent ductus arteriosus was clamped and ligated. The patient was heparinized prior to clamping the aorta proximal and distal to the coarctation. The coarctation was resected and the proximal and distal ends of the aorta were broadened. A primary end-to-end anastomosis was completed creating a tension-free, size-matched repair. The postoperative recovery was uneventful with restoration of symmetric blood pressures in all four extremities.

DISCUSSION

The combination of right aortic arch and coarctation of the aorta is a rare morphologic finding. McEnlhinney et al. reviewed the worldwide literature of this rare

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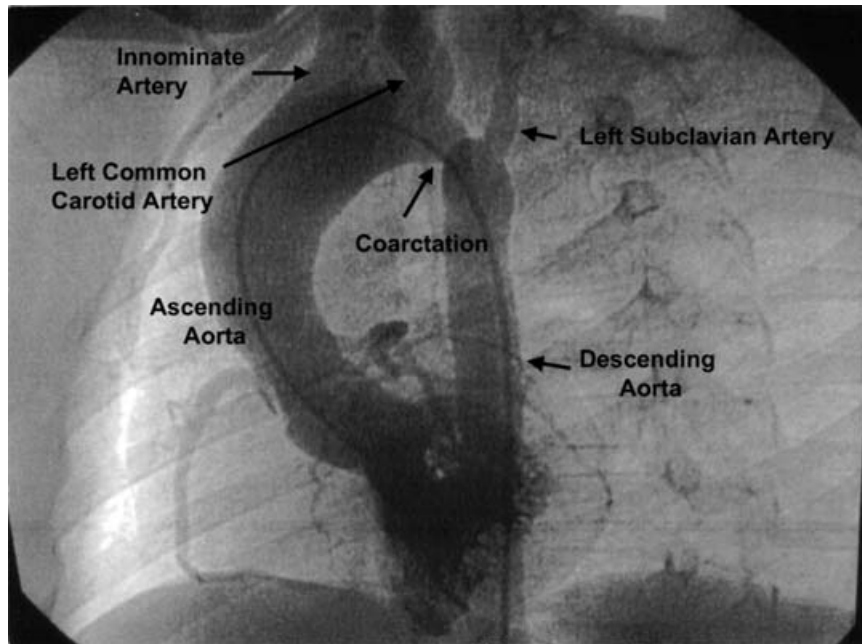


Figure 1. Aortogram demonstrating right-sided aortic arch, coarctation, and aberrant left subclavian artery.

phenomena describing 23 published reports, including his own series of 4 patients.³ Six of these cases involved mirror image branching and 12 patients had aberrant origin of the left subclavian artery from the descending aorta with a left ductus. Our patient also had an aberrant subclavian artery; however, a right-sided ductus was present.

The etiology of both right aortic arch and coarctation is complex and not fully understood. Goldmuntz et al. described a correlation between patients with dele-

tions of the chromosomal region 22q11 in patients with isolated anomalies of the aortic arch.⁴ These microosomal deletions are thought to play a role in neural crest cell migration. However, the pathogenic nature of this gene deletion has not been fully described. Subsequent genotyping of this patient demonstrated no chromosome abnormality.

The persistence or regression of the six pairs of embryonic aortic arches results in the normal pattern of aorta, pulmonary artery, and great vessels. The

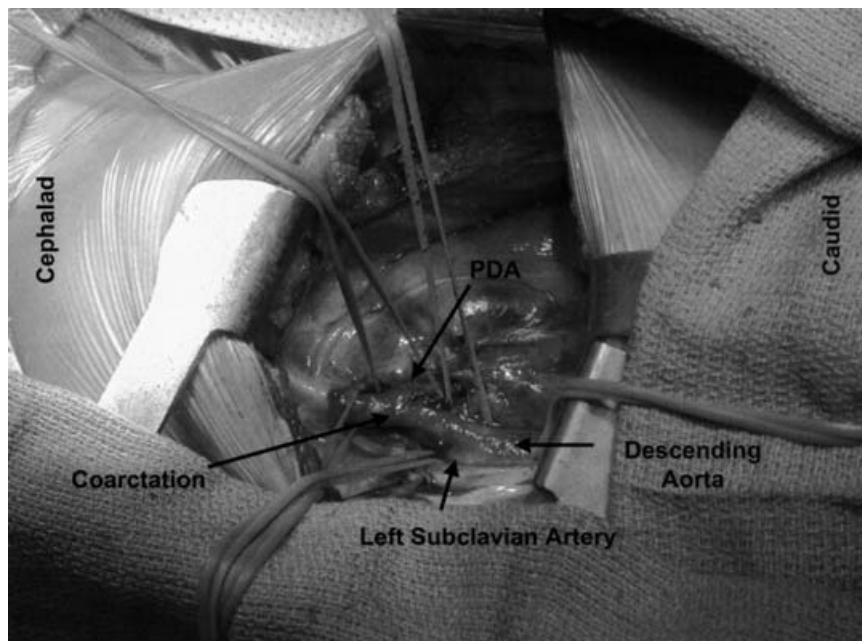


Figure 2. Intraoperative photograph of right-sided arch exposed through a right thoracotomy. The coarctation is labeled. Left aberrant subclavian artery is marked. The patent ductus arteriosus (PDA) is evident.

hemodynamic conditions in a normally developing cardiovascular system favor right ventricular flow through the left ductus and into the left descending aorta.⁵ Cardiac anomalies associated with right ventricular outflow obstruction diminish the right-to-left ductal flow and subsequently diminish the flow-related impetus for persistence of the left ductus and dorsal aorta. It is postulated that coarctation is initiated in fetal life by the presence of a cardiac anomaly that results in decreased antegrade flow through the aortic valve as seen in bicuspid aortic valves and VSDs. This flow principle of increased right-to-left ductal flow and decreased flow across the aortic arch is in sharp contrast to the flow principles associated with right aortic arch. These conflicting hemodynamic principles most likely account for the rarity of coarctation of a right aortic arch; however, there are undoubtedly other factors that influence the process of aortic arch development.

The importance of accurately defining the anatomy of the arch and great vessels cannot be overemphasized. In this particular case, angiography clearly defined the arch, brachiocephalic, and ductal anatomy which then confirmed our surgical approach. Angiography, however, is not always necessary. While echocardiography is not the gold standard for identifying the great vessels, at our institution, echocardiography is usually reliable. If there is any question as to the specificity of echocardiography, we recommend cardiac MRI to further define the arch and great vessels.

Our most common surgical repair of coarctation, with or without right aortic arch, has been to perform an end-to-end anastomosis through a posterolateral thoracotomy. The right-sided arch of this patient was approached through a right thoracotomy and was easily exposed. In our experience with coarctation, we almost always find the aorta able to be mobilized sufficiently to allow complete resection of abnormal tissue and a tension-free primary repair. We successfully performed a spatulated end-to-end anastomosis to augment the diameter of the aorta. Other techniques of coarctation repair, such as patch aortoplasty or a subclavian flap, could have been implemented in this case. Our experience with aneurysmal dilation of the posterior aortic wall⁶ with these techniques has limited their use in our institution. We described that the hemodynamic nature of the prosthetic material most likely adds to this phenomenon with resultant aneurysm formation, as the tensile strength of the prosthetic patch differs from that

of the aorta. When the aortic pulse wave reaches the stiff, less-compliant patch, turbulence is transmitted to the more elastic aorta opposite and adjacent to the patch. This transmitted turbulence subsequently leads to progressive weakening and dilation of the aortic wall. We have recently described that the incidence of aneurysm formation following coarctation repair with patch aortoplasty is significantly higher in patients with concomitant arch hypoplasia.⁷ Because of the morbidity associated with postoperative aneurysm formation, we encourage coarctation resection with extended end to undersurface of aortic arch anastomosis to address both the aortic arch hypoplasia and juxtaductal stenosis.

CONCLUSION

In summary, the combination of right aortic arch and coarctation is extremely rare. The rarity of these combined entities most likely is explained by the contrasting fetal hemodynamics that tend to cause one or other of these anomalies. The frequency of associated cardiovascular anomalies found with either condition warrants extensive evaluation and detailed preoperative imaging. Surgical repair of the coarctation is imperative to avoid late morbidity and can be safely performed through a right thoracotomy.

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