

CARDIOVASCULAR IMAGES

A joint publication of the Department of Radiology and Heart Center

Truncus Arteriosus

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Clinical History

A 31 year old female with VSD, diagnosed as a child, presented for follow up of congenital heart disease. She was asymptomatic though on physical exam, she had mild cyanosis and mild bilateral clubbing. She was referred for echocardiography for surveillance. Echo showed findings consistent with VSD as well as ASD. The pulmonary arteries were not identified. RV and LV function were preserved. CT was ordered to evaluate the pulmonary vasculature and ascending aorta.

Findings

CT demonstrates a membranous VSD with an enlarged, overriding aorta. The pulmonary outflow tract is absent. There is calcification of the right and non coronary cusps as well as calcification of the remnant pulmonary outflow tract. There is a right-sided arch with mirror-image branching. Two large collateral vessels are identified arising from the descending aorta supplying the right lung and one vessel on the left supplies the left lung. There is proximal stenosis of the right superior collateral vessel with post-stenotic dilatation. Findings are consistent with a Collett and Edwards Type IV truncus arteriosus, also known as a pseudotruncus.

Discussion

Truncus arteriosus is a complex congenital anomaly, the embryology of which is still being elucidated. It is defined anatomically when one artery arises from a combined right and left ventricular outflow tract, giving rise to both the systemic and pulmonary circulation as well as the coronary arteries. Truncus arteriosus is usually associated with a ventricular septal defect (VSD) although uncommonly there is an intact ventricular septum. The Van Praagh classification system was introduced in 1965 and has largely replaced the earlier Collett and Edwards system. However, the Collette and Edwards system included a category which is not included in the Van Praagh classification.

The Collette and Edwards type IV truncus arteriosus demonstrates no identifiable pulmonary arteries. Instead, blood supply to the lung arises from hypertrophied bronchial arteries from the descending aorta. There is controversy whether this represents the extreme form of tetralogy of Fallot with pulmonary atresia. Some argue that with tetralogy of Fallot with pulmonary atresia, either the right, left, or distal main pulmonary artery is present (though hypoplastic) receiving blood from a patent ductus arteriosus or from bronchial arteries; whereas, in type IV truncus arteriosus, no true pulmonary arteries are identified.

The value of CT and MRI lies in reliable quantification of ventricular function and detailed assessment of the presence or absence of the pulmonary arteries. This is especially true in the case of newborns in whom surgical correction is planned. Additionally, MRI is useful for post-surgical follow up to evaluate right ventricular function, function of the RV to pulmonary artery homograft, and function of the neo-aorta.

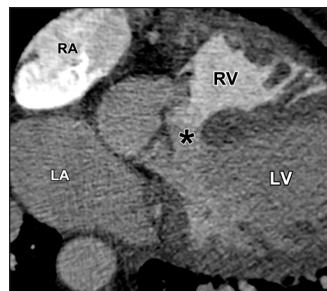


Figure 1

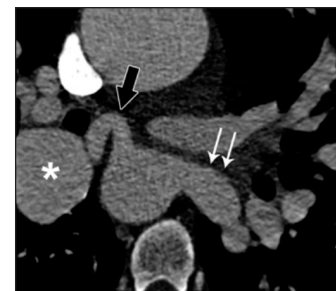


Figure 2

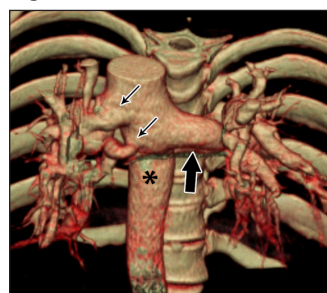


Figure 3



Figure 4

Figure 1: Axial image at the level of the ventricular outflow tract demonstrates a membranous VSD (asterisk). The aorta is large and overrides the septum.

Figure 2: Axial image demonstrates arteries arising from the descending aorta, perfusing the lung. On the right, there is an aneurysm arising from the superior right pulmonary artery (asterisk). The inferior right artery is smaller in caliber (black arrow). There is a single artery supplying the left lung (double arrow). The main pulmonary artery is absent.

Figure 3: 3D reformatted image with the heart removed demonstrates large arteries arising from the descending aorta (asterisk), perfusing the lung. On the right, two arteries are seen (small arrows). The superior artery is stenotic with post-stenotic dilatation. Only one artery is seen on the left (large arrow).

Figure 4: Curved reformatted image of the descending aorta shows a right aortic arch with right-sided descent. Two large arteries arise from the descending aorta, supplying the lung. The right pulmonary artery demonstrates a proximal stenosis (double arrow) with post-stenotic dilatation (asterisk). There is no stenosis of the left pulmonary artery (white arrow).

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