# Repair of anomalous origin of right pulmonary artery in Tetralogy of Fallot with anomalous coronary artery

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**Abstract**

We present an unusual combination of lesions in an eight months old child diagnosed with Tetralogy of Fallot (TOF), Anomalous origin of Right Pulmonary artery (AORPA) and anomalous coronary artery (ACA) crossing the pulmonary annulus. The association of AOPA and TOF is extremely rare with an incidence of 0.4%. (1) The incidence of anomalous coronary artery in TOF is 10.3%. (3) However a combination of all three lesions poses challenges to surgical repair and has not been previously reported.

**Case Report**

An eight months old male child weighing 4.5 Kg presented with complains of breathlessness, repeated respiratory tract infections and failure to thrive. The child was malnourished and had tachycardia and tachyopnea. He had mild cyanosis with an oxygen saturation of 85% on room air. An ejection systolic murmur was heard over the pulmonary area on auscultation. Chest radiogram revealed cardiomegaly and differential pulmonary vascularity with features of increased pulmonary blood flow on the right lung field. The electrocardiogram suggested bi-ventricular hypertrophy. Echocardiogram examination revealed anomalous origin of right pulmonary artery (AORPA) from ascending aorta and hypoplastic pulmonary annulus. The main pulmonary artery (MPA) continued as left pulmonary artery. The interatrial septum was intact and there was a large mal-aligned ventricular septal defect. The obstruction in the right ventricular outflow tract was predominantly deemed to be valvular. A decision to disconnect right pulmonary artery (RPA) from aorta and create pulmonary confluence, close the VSD and enlarge the pulmonary outflow was made.

Surgical approach was median sternotomy. During intra-operative inspection, the pulmonary annulus was found to be severely hypoplastic and a large coronary artery was found crossing the pulmonary annulus. (Figure 1) It was a branch from right coronary artery crossing the right ventricular outflow tract and running parallel to the inter-ventricular groove as an accessory left anterior descending artery (LAD). Pulmonary arteries were extensively mobilised upto the lobar branches.. Cardiopulmonary bypass was initiated after aorto-bicaval cannulation. RPA was snared immediately prior to initiation of CPB. The ligamentum ductus was divided. Del Nido's Cardioplegia at 30ml/ Kg was administered to achieve cardiac arrest. RPA was disconnected from the side of aorta. Defect in the aorta was sutured primarily in two layers. RPA was anastomosed to MPA directly and pulmonary confluence was created. The ventricular septal defect (VSD) was routed to aorta using bovine pericardial patch. MPA was opened longitudinally without cutting across the pulmonary annulus in order to preserve the anomalous coronary artery. A tri-leaflet valved conduit was created using bovine pericardium (St. Jude Medical) and 0.1 mm Polytetrafluoroethylene (Gore-Tex, WL Gore and Associates Inc, Flagstaff, Ariz). The valve leaflets are created with 0.1mm PTFE membrane which is sutured to a sheet of bovine pericardium. The bovine pericardial sheet is rolled into a tube to make the valved conduit. The internal diameter of the conduit was 12mm and it was interposed between the right ventricle and MPA. (Figure 2) The native right ventricular outflow tract was also preserved making a double

barrel right ventricle outflow tract. The postoperative course was uneventful. The child was weaned from mechanical ventilation and extubated after 48 hours and was discharged on sixth postoperative day. The child is asymptomatic at 36 months after surgery. The native pulmonary valve has grown from 6 mm to 8 mm in diameter and the RV to PA conduit is functioning well with mild regurgitation.

**Comment**

Anomalous origin of branch pulmonary artery from aorta (AOPA) is an extremely rare condition accounting for only 0.1% of all congenital heart defects. (1) Aortopulmonary septum develops by fusion of right and left conotruncal ridges. Severe mal-alignment of these conotruncal ridges results in anomalous origin of the RPA from the ascending aorta. AOPA can be isolated or it may be associated with other cardiac defects. The most common associated lesion is patent ductus arteriosus, seen in 50% of cases. Rarely, it is associated with TOF, ventricular septal defect (VSD), Aortopulmonary window ( APW), Interrupted aortic arch and atrial septal defect. Anomalous origin of right pulmonary artery (AORPA) is far more common than anomalous origin of left pulmonary artery (AOLPA). (2) However, in tetralogy of Fallot AOLPA is far more common than AORPA. Clinical features are those of increased pulmonary blood flow and congestive heart failure. The cyanosis associated with tetralogy of Fallot may not be apparent due to increased blood flow to the ipsilateral lung. Differential lung vascularity on chest radiogram may be suggestive of the diagnosis. The repair should be performed immediately after diagnosis to prevent congestive heart failure and ipsilateral pulmonary hypertension. Our patient had an added complexity in the form of anomalous coronary crossing the pulmonary annulus. The incidence of anomalous coronary artery or a large conal artery crossing the RVOT is 10.2% based on a meta analysis of 28 studies. (3) The combination of AOPA and ACA with TOF has not been previously reported in the literature. ACA was circumvented by interposing a hand-sewn bovine pericardial tube with a tri-leaflet valve constructed from PTFE membrane. The integrity of the native pulmonary valve was maintained allowing future growth as has been proposed in literature. (4) This was demonstrated in our patient on echocardiogram examination. The native pulmonary annulus has increased in diameter from 6 mm to 8 mm over a period of 30 months. A promise of this growth potential combined with a larger than required RV to PA conduit will delay or perhaps prevent re-operations for the RVOT.

**Declaration of conflicting interests**

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**Consent**

The patient's family gave permission to publish this case report.

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**Legend for figures**

Figure 1 Sketch of preoperative anatomy: Ac LAD: Accessory Left Descending artery; AORPA: Anomalous origin of right pulmonary artery; LPA: Left pulmonary artery

Figure 2 Sketch of postoperative anatomy: Ac LAD: Accessory Left descending artery; AORPA: Anomalous origin of right pulmonary artery; LPA: Left pulmonary artery

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