

Tetralogy of Fallot with Absent Pulmonary Valve Syndrome with Absent Left Pulmonary Artery - A Rare Presentation

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Abstract:

Absent left pulmonary artery with Tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS), is a rare congenital cardiac anomaly. Here we present such a case of A 2 year 11 month old girl with cyanosis, exertional dyspnoea. Her diagnosis is confirmed by echocardiography and CT angiogram. There are very few cases have been reported till date with high post-operative mortality. Although per operative decision

making was challenging regarding pulmonary valve and size of the RPA, we performed ICR with RPA reductionplasty and creation of monocuspid pulmonary valve with success. As it is a rare association and we have overcome the hindrance we came across per operatively, we are reporting this case.

Keyword: Tetralogy of Fallot, Absent Left pulmonary artery, absent pulmonary valve syndrome.

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Introduction:

Absent pulmonary valve syndrome (APVS) is an uncommon form of congenital heart disease. It occurs in 2.4% to 6.3% of patients with tetralogy of Fallot (TOF).¹⁻³ The primary symptoms are recurrent wheezing and dyspnea, which occur due to compression of the bronchi and trachea by aneurysmal pulmonary arteries. This syndrome was first reported in 1830.^{4,5} APVS may occur alone or in association with other forms of congenital heart disease, particularly TOF.^{6,7} Absence of the pulmonary artery in association with an absent

pulmonary valve is an extremely rare scenario.⁵ McCaughan et al.⁸ found a 14.3% incidence of true (complete) absence left pulmonary artery (ALPA) in a group of 35 patients with APVS. We have presented one patient with TOF and APVS with true (complete) absence of the left pulmonary artery.

Case report:

2 Yrs 11 Months old a pretty girl who is the only child of her non consanguinal parent came to our hospital with a diagnosed case of congenital heart disease and presented with complaints of bluish discoloration of skin & palpitation. She was cyanosed and dyspnic. NYHA class-II, no clubbing, and oxygen saturation at room air was 86%, heart rate 120beats/min. She had systolic murmur at the middle of left sternal border. Chest bilaterally Clear.

Echocardiography revealed Tetralogy of Fallot dysplastic pulmonary valve with severe PS with moderate to severe PR Hugely dilated MPA & absent LPA Left aortic arch Good biventricular function.

CT angiography of heart and great vessel show situs solitus levocardia. Pulmonary infundibulum severely

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Fig.-1: Absent LPA, dilated RPA, dysplastic PV

stenosed with absent pulmonary valve, left pulmonary artery. Hugely dilated right pulmonary artery. Perimembranous type of Ventricular septal defect. Bovine type of aortic arch. Aberrant origin of right sub-clavian artery. Hypertrophied right ventricle.

McGoon Ratio 3.54 Nakata Index 1293 mm²/BS. With proper counseling with her parent we planned for intra cardiac repair.

Routine induction of General Anesthesia and placement of monitoring lines. Standard median sternotomy done. Thymus dissected off. Pericardium harvested. Systemic heparinization (300 U/kg) and aorto-bicaval cannulation established. CPB temperatur cooled to 28°C. Aorta cross-clamped and heart arrested with cold Del Nido cardioplegia delivered antegradely through the aortic root. Cavae snared . LV vented through RA.

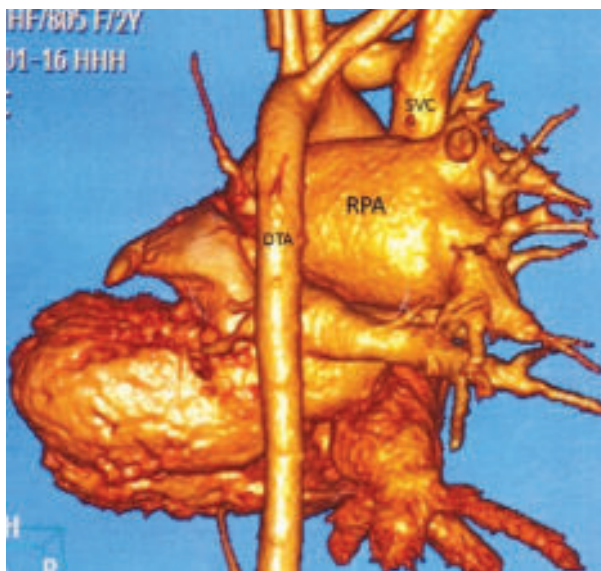


Fig.-2: Absent LPA

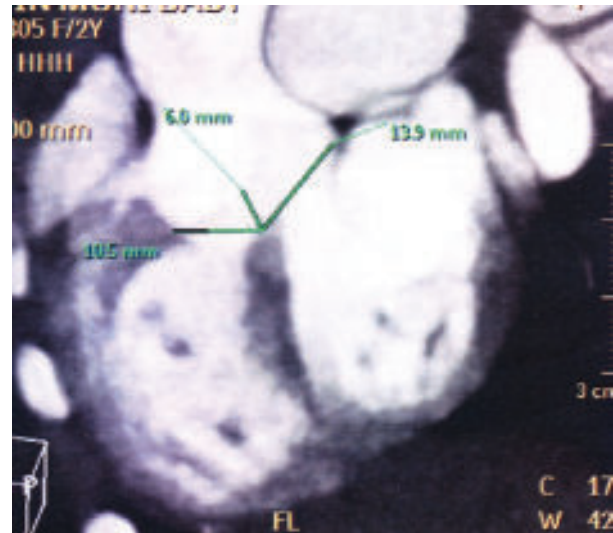


Fig.-3: large VSD, aortic overriding

PA tomy & Right ventricular outflow tract tomy done. VSD repair with Dacron patch. Hypertrophied bands of RVOT excised through RVOT. RPA reduction plasty done at posterior & anterior wall. Augmentation of RVOT & MPA by transannular patch of pericardium with creation of PTFE monoleaflet PV. Fenestration was kept in IAS. Rewarming started. Deairing the heart done. Cross clamp released. RA closure with 6-0 prolene. Patient weaned off CPB with NSR. Postoperative PA pressure was high. We allow the high PA pressure and give protamine. Decannulation done. 2 pacing wire attached with RV, 2 with RA. Chest closure done in layers keeping O2 mediastinal and right pleural drain in situ. She was shifted to ICU with stable hemodynamics and on inotrope (Dobutamine and milrinone) support.

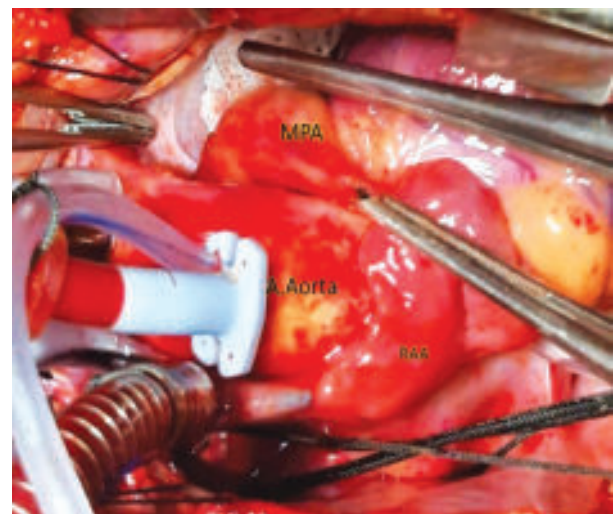


Fig.-4: Absent LPA

Her ICU stay was a bit stormy initially but she recovered well. She was extubated on 2nd POD total ventilation time was 45 hrs 30 min.

She was shifted to ward on 6th POD and discharged from hospital on 9th POD. She was doing well on her first follow up one month after discharge.

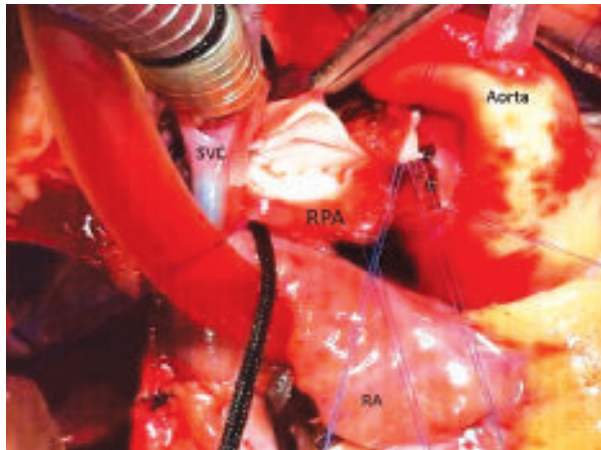


Fig.-5: posterior wall of the RPA sutured for reduction

Discussion:

APVS is a rare congenital cardiac malformation characterized by a rudimentary or dysplastic pulmonary valve and the aneurysmal dilatation of the main pulmonary artery and one or both of its proximal branches.⁹ Pulmonary regurgitation plays an important role in the expansion of the main pulmonary artery and its proximal branches. Turbulence due to severe pulmonary regurgitation at the narrow pulmonary annulus and poststenotic dilatation result in expansion of the pulmonary arteries.¹⁰ Pulmonary expansion results from increased pulse pressure in the pulmonary artery due to pulmonary regurgitation, as well as increased RV stroke volume and increased blood volume in the RV during diastole and subsequent turbulence at the stenotic pulmonary annulus, followed by post-stenotic dilatation.¹⁰ These findings are supported by the subsequent decrease in pulmonary artery diameter following surgery for pulmonary regurgitation.¹¹ APVS patients are separated into two groups based on their ages and clinical courses: The patients in the first group are newborn babies or infants who present with dyspnea, recurrent lung infections, pulmonary emphysema, and atelectasis as a result of compression of bronchi secondary to pulmonary artery dilatation. The second group includes older patients with mild symptoms who survive infancy. For these patients, closure of their VSD

and relief of their pulmonary stenosis may be performed later on an elective basis with minimal risk.^{12,13} Absent pulmonary valve with true (complete) absence of the left pulmonary artery represents an extremely rare combination in TOF. The absence of the left pulmonary artery is believed to be the result of a continued link between the fetal ductus arteriosus and the intrapulmonary aspect of the pulmonary artery during the in utero phase, as well as the involution of the 6th aortic arch (the extrapulmonary portion of pulmonary artery). Severe hypoplasia occurs in the pulmonary artery due to closure of the ductus after birth.¹⁴ Surgical correction of APVS is both extremely challenging and controversial.³ The types of surgical correction used have evolved over time and should be individualized based on the patient's age and clinical symptoms. In a literature review by Calder et al.,⁵ 13.3% to 40% mortality rate was reported following total corrective surgery involving patients in all age groups. The operative mortality may be as high as 58.3% in symptomatic infants.⁸ Patients with minimal symptoms often present later in life and have a relatively low risk of mortality (5%) with elective surgery.²⁴

Surgical correction of APVS is even more challenging when the former is associated with ALPA because it is associated with a higher mortality rate. Total correction is possible without reconstruction of the pulmonary valve, but the long-term results of this procedure are unknown.³ McCaughan et al.⁸ recommend valve insertion in all patients with APVS and ALPA. While the unilateral absence of the pulmonary artery will ameliorate the development of pulmonary hypertension, it may also worsen pulmonary regurgitation and right ventricular dysfunction.

Conclusion:

Cases of APVS and TOF with unilateral absence of the pulmonary artery are very rare and have high mortality rates. Per operative decision making also crucial regarding size of the RPA, competency of pulmonary valve and early development of pulmonary hypertension because all are contribute to the good post-operative outcome.

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