Unilateral absence of left pulmonary artery with absent pulmonary valve syndrome

Sachin Mahajan[1], Sudhansoo Khanna[1], Sanjeev Naganur[1], and JAVID RAJA[1]

[1]Post Graduate Institute of Medical Education and Research

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Abstract

Tetralogy Of Fallot (TOF) with absent pulmonary valve (APV) and unilateral absence of pulmonary artery (UAPA) is an extremely rare congenital cardiac anomaly. "Complete" unilateral absence implies absence of both intrapericardial and hilar segments. Co- existence of both free pulmonary regurgitation (PR) and decrease in the cross sectional area for right ventricular outflow tract (RVOT) in this substrate, are detrimental for right ventricular (RV) function. Early intervention to preserve RV function is imperative. Durable and competent valve in RVOT can be extremely helpful in improving the overall prognosis. We present a case of a two year old child, TOF with APV, absent left pulmonary artery (ALPA) who underwent RVOT reconstruction, with a pulmonary valve (PV) made from polytetrafluorethylene (PTFE) membrane. To our knowledge construction of PV using Graham Nunn technique, has not been reported with this morphology.

Text

Introduction

Tetralogy Of Fallot (TOF) with absent pulmonary valve (APV) and unilateral absence of pulmonary artery (UAPA) is a rare congenital cardiac anomaly. This syndrome was first reported in 1930.[1] McCaghan et al reported a 14.3% incidence of absent LPA in a group of 35 patients with APV syndrome (APVS).[2] Surgical correction carries a very high mortality, with sudden cardiac deaths reported in the majority of patients.[3] Construction of durable valve in RVOT with total correction can prevent future dilatation of proximal pulmonary artery (PA), and at the same time preserve RV function in the long term. We present a patient with this combination, who underwent total correction with construction of PV using a PTFE membrane.

Case Report

Informed written consent was taken from the guardian for publication. A two year old male child was brought with the history of dyspnea and cyanosis on mild exertion since 3 months of age. Patient had cyanosis and a to and fro murmur was present at the upper left sternal border. Chest X-ray revealed mild cardiomegaly, hypoplastic left lung and prominent right hilar shadow. Echocardiography revealed TOF morphology, large subaortic ventricular septal defect (VSD), severe pulmonary stenosis (PS) and dilated main pulmonary artery (MPA) with continuation only as a dilated right pulmonary artery (RPA), suggestive of absent left pulmonary artery (LPA). Neither PDA nor any major collateral supplying the left lung could be appreciated on aortography. Pulmonary vein wedge injection also confirmed its absence. No obvious compression of airway on the right side could be appreciated on CT scan. Child underwent successful total correction. Intra-operatively, pulmonary annulus was mildly stenotic, RPA was aneurysmally dilated and pulmonary valve was rudimentary (Figure 1 and 2). After minimal infundibular resection, VSD was closed with 5/0 polypropylene, TF needle, using continuous suture technique. As the annulus was small with rudimentary leaflets, hence for RVOT reconstruction the infundibular incision was extended onto MPA to a limited extent. In view of absence of airway symptoms, no compression of airways and absence of LPA,

the dilated RPA was only partially plicated. Completion of RVOT reconstruction was done by creating a bicuspid valve from 0.1-mm PTFE membrane using Graham Nunn technique[4] (Figure 3) and augmentation of RVOT by a rectangular patch of treated pericardium. Trans-esophageal echocardiography (TEE) revealed mild pulmonary regurgitation (PR) with peak gradient of 22 mm Hg. Postoperative course was uneventful.

Discussion

APVS is characterized by a rudimentary PV and aneurysmal dilatation of central pulmonary arteries.[5] It occurs in 2.4% to 6.3% of patients with TOF.[6] Compression of airways secondary to pulmonary dilatation can cause early presentation and portray dismal prognosis. Co-existence of absent APV with complete absence of the LPA in TOF is an extremely rare combination.

The absence of LPA 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.[7] Only 18 patients with APVS and TOF with complete absence of left PA has been reported.[3, 8] Complete absence of RPA in association with APVS has not been reported.[3]

Asymmetric lung field with reduced vascularity on left side can provide a clue to the diagnosis. Cardiac catheterization and angiography are usually diagnostic. Combination of pulmonary regurgitation and a single pulmonary artery may ultimately result in pulmonary hypertension. Pulmonary hypertension will in turn worsen pulmonary regurgitation, creating a vicious cycle ultimately leading to early onset of right ventricular dysfunction.[9] We expect increased overall mortality in this subset of TOF patients owing to high pulmonary vascular resistance (PVR), free PR, and significantly less cross sectional area of RVOT.

Timing of surgical intervention is usually dictated by clinical status and onset of RV dysfunction. As the entire right ventricular stroke volume is directed to the unilateral pulmonary artery, aneurysmal dilatation of connected PA can occur early and be more pronounced, leading to increased symptomatology. In symptomatic newborn and infant, with involvement of airway, morbidity and mortaility rise considerably.[2]

Total correction, including construction of a durable PV, will be the most appropriate surgical approach. TOF with absent pulmonary valve with dilated PA, have PVR on the higher side. Considering unusual spectrum of PA morphology in which RV has to eject against higher PVR, we strongly believe in construction of valve in RVOT. This would have both short term and long term benefits.

Conduit repair is possibly indicated in patients with airway obstruction, but it has inherent problems. For those without airway obstruction, valves created from pericardium or 0.1-mm PTFE membrane is always an option. The competence of valve created depends upon surgeon's skill. Long term durability of the pericardium is extremely doubtful. Graham et al reported that all the pericardial valves constructed by them for RVOT developed free PR whereas the bileaflet polytetrafluoroethylene (0.1-mm) valves have remained competent with regurgitant fractions of only 5% to 30% (as evaluated on magnetic resonance imaging) and this has remained stable with time.[4] To our knowledge, creation of a bileaflet PV using PTFE membrane in a TOF patient with absent LPA and APV syndrome has not been reported in the literature until now. In our patient, with two years follow up, there is only mild PR with preserved RV function.

To conclude, TOF with APV with UAPA is a very rare congenital anomaly, with high mortality rate. Early surgical intervention, with provision of durable valve in RVOT, can preserve RV function. Onset of RV dysfunction in the follow-up mandates aggressive intervention, which might include conduit replacement, to eliminate any degree of PR.

Author contributions

Dr. Sachin Mahajan- Critical Revision of article.

Dr. Sudhansoo Khanna - Data analysis, Drafting article.

Dr. Sanjeev Naganur- Concept/ design.

Dr. Javid Raja- Data collection.

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Figure legends

Figure 1 Intra-operative photograph showing MPA continuing as single dilated RPA with absent LPA

Figure 2 Intra-operative photograph after trans-annular incision showing absent PV and single dilated RPA.

Figure 3 Intra-operative photograph showing hand sewn bileaflet PV created (from 0.1-mm PTFE membrane) in the RVOT.