

Extended Abstract

Total Anomalous Pulmonary Venous Connection With Atrioventricular Septal Defect: A Rare Association

Ketak Nagare¹ MBBS, MS, MCh, Balaji Aironi² MBBS, MS, MCh, Abhishek Joshi² MBBS, MS, MCh, Mohammed Ibrahim¹ MBBS, MS, MCh, Mohammed Idhrees¹ MBBS, MS, MCh, FAIS

¹Institute of Cardiac and Aortic Diseases, SIMS hospital, Chennai, India²Seth G S Medical College and KEM hospital, Mumbai, India

Email id : ketak29kejal@gmail.com

Abstract:

Total anomalous pulmonary venous connection along with atrioventricular septal defect is a rare pathology and becomes a different subset compare to TAPVC alone. The combination of two separate life-threatening congenital heart defects makes the management of these patients challenging. View towards the surgical management is changing after reporting few cases treated successfully with an anatomical repair. We report a case of an eight-month infant diagnosed as supracardiac TAPVC associated with the complete atrioventricular septal defect. The patient underwent early single-stage intracardiac repair.

Key Words: TAPVC, AVSD, Congenital cyanotic heart disease, double patch technique.

Introduction :

Total anomalous pulmonary venous connection (TAPVC) is a congenital cyanotic heart disease (CCHD) in which all four pulmonary veins fail to connect directly to the left atrium. The incidence of TAPVC is 0.7-1.5% of all congenital heart diseases. Atrioventricular septal defect (AVSD) is also CCHD

characterized by a common atrioventricular junction and with deficient atrioventricular septation. The incidence of AVSD is 4% of all congenital cardiac malformations. But the incidence of both these conditions together is 1% of all CCHD. It is tough to manage patients with TAPVC with AVSD due to corrective surgery is technically demanding, co-morbid conditions presenting along with it, and difficult postoperative management.

We report a case of an eight-month-old infant having supracardiac TAPVC with mesocardia & Complete AVSD.

Case report :

8-month infant presented with progressive central cyanosis and repeated respiratory tract infections from the age of 15 days. The infant had a body mass index of less than 5th percentile. On examination, there was central symmetrical deep cyanosis with room air saturation of 78%. On auscultation, she had an ejection systolic murmur (grade III/VI) all over the precordium. The chest X-ray revealed cardiomegaly, mesocardia along with pulmonary plethora. Electrocardiogram showed left axis deviation. Echocardiogram showed supracardiac TAPVC with complete AVSD. There was ostium primum atrial septal (OP-ASD) defect with an obligatory right to left shunt along with a single atrioventricular valve. There was a large-sized non-restrictive inlet ventricular septal defect (VSD). The patient had severe pulmonary hypertension & moderate dysfunction of both ventricles.

Midline sternotomy was done. Mesocardia was confirmed. The pulmonary artery was tense and dilated. Inferior vena cava (IVC) was on the left side. The pulmonary veins formed a confluence that drained

into the innominate vein via a vertical vein. The proximal part of the vertical vein was aneurysmal. Cardiopulmonary bypass was established by cannulating aorta & both cavae. Antegrade cold cardioplegia (del Nido) was used. It was difficult to expose the heart for TAPVC correction due to left-sided IVC and mesocardia. The anastomosis of pulmonary confluence was done with the left atrial appendage with the help of retracting sutures which were taken into the myocardium with caution not to injure it. The inlet VSD was closed through right atrium (RA) with interrupted polypropylene pledgetted sutures using the Dacron patch (Sauvage Filamentous dacron - BARD). OP-ASD closed with a self-untreated pericardial patch (autograft). Except in the area near conduction tissue, sutures for OP-ASD closure were taken in a contentious manner. The total cardiopulmonary bypass time was 300 minutes. The patient was weaned off cardiopulmonary bypass and was shifted to the recovery room on supports of injection milrinone 0.4 mcg/kg/min, injection adrenaline 0.03 mcg/kg/min, and injection noradrenaline 0.04 mcg/kg/min.

It was difficult to wean off the patient from a ventilator. The patient developed pneumonia during the recovery period. An echocardiogram demonstrated pulmonary anastomosis patency and normal bi-ventricular function. The patient was discharged on postoperative day 21. The patient was asymptomatic at a 6month follow-up.

Discussion :

TAPVC with AVSD is one of the rare forms of congenital heart defects and is even rarer when a left-sided IVC and mesocardia present along with it. In TAPVC patients there is a mixing of oxygen-rich blood from the pulmonary system and deoxygenated blood from the systemic venous system. Clinical presentation in

AVSD depends upon the type of AVSD. In complete AVSD, patients develop congestive heart failure earlier. Both TAPVC and AVSD are the cause of pulmonary overflow and cardiac volume overload. Hence when both of these conditions present together it results in early congestive cardiac failure and pulmonary arterial hypertension.

Surgical correction of TAPVC with AVSD is technically demanding. Cardiac structural deformities like heterotaxia, right isomerism are commonly associated with AVSD patients. It makes exposure of the heart difficult to perform an anastomosis between pulmonary venous confluence and Left atrium. Sachin et al described a bi-atrial approach. In a 24-year patient rerouting of the left Superior vena cava into RA was done through left atrium and AVSD correction performed via RA . We used retraction stitches over myocardium for better exposure of the heart. AVSD can be corrected using single or two separate patches. Self-pericardium is easy to harvest, inexpensive, easy to handle, and decreases the risk of thromboembolism compared to prosthetic material patches. AVSD correction with a double patch technique is convenient when cardiac anatomy is unusual.

TAPVC with other cardiac pathology cases is difficult to manage even in the postoperative period. In a single case study from Denmark, even after successful surgical correction of TAPVC with AVSD along with Truncus Arteriosus, the patient took 25 days for recovery. During the initial period, early single-stage anatomical repair was under question. But subsequently, reporting of successful management of such cases with early single-stage cardiac correction surgery has changed the mind-set of surgeons .

Our 8-month-old patient presented with deep central cyanosis and repeated upper respiratory tract

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infections. Echocardiography diagnosed her having TAPVC and AVSD along with left-sided IVC. She underwent single-stage intracardiac repair with a double patch technique for AVSD correction using self-pericardium. The patient had a difficult postoperative course, but she recovered and asymptomatic at 6 months follow up.

Conclusion :

TAPVC with AVSD is a rare combination. In combination, these pathologies have additive negative effects on cardiac hemodynamic making early surgery a must. Surgeons may have intra-operative difficulties due to associated pathologies and cardiac structural deformities. Preoperative planning is required. Post-operative management of these cases is also difficult and it requires a large scale study with similar cases to make perioperative management protocol. Still, early single-stage correction gives good results.