Trans Pulmonary Closure of an Aorto-Pulmonary Window in a patient of Tetralogy of Fallot: A case report

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Abstract

We present a case of Tetralogy of Fallot accompanied by a type II Aorto-Pulmonary Window with severe pulmonary arterial hypertension in a pediatric patient. A successful repair of TOF with trans-pulmonary patch closure of APW was done.

Introduction:

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease (CHD) [(1)]. Its association with Aorto-Pulmonary window (APW) is very rare. Only 19 cases of TOF associated with APW have been reported of which 5 had associated pulmonary atresia [(2)]. Surgical management in this combination is indicated at the time of diagnosis to prevent the development of irreversible pulmonary vascular disease (PAH).

CASE REPORT

A 3-year-old girl weighing 9.7 kg presented with symptoms of recurrent chest infection with poor weight gain since birth. Clinical examination revealed a grade 3/6 ejection systolic murmur in the left sternal border, and oxygen saturation of 89% on room air. Chest x-ray revealed plethora covering 2/3rd of the lung field with a boot-shaped heart (Figure 1A). Transthoracic echocardiography showed a large mal-aligned peri-membranous Ventricular Septal Defect (VSD), 50% aortic override, good sized branch pulmonary arteries (PA) and a communication between distal ascending aorta and pulmonary artery likely to be an APW (Figure 2A). Computed tomography angiography was performed which suggested a Type II APW measuring 10 mm between ascending aorta and main pulmonary artery (MPA) at a distance of 20 mm from aortic valve. There was associated infundibular and valvar pulmonary stenosis. The Mc-Goon score was 4.1. Cardiac catheterization study indicated distal APW filling the PA's with findings of TOF and normal coronary arteries (Figure 1B). Cath data (post oxygen) revealed left ventricular pressure of 100/14 mm Hg, right ventricular pressure of 98/6 mm Hg and mean PA pressure of 62 mm Hg.

With a diagnosis of TOF with Type II APW and features suggestive of reversibility of PAH in TOF, surgical repair was decided. After appropriate consent she underwent an elective repair. A median sternotomy was done, and cardiopulmonary bypass was established with aorto-bicaval cannulation. The MPA and Aorta were dissected, branch PA's were looped separately and snugged. Aorta was cross clamped, and the heart was arrested by administering antegrade Del Nido (20 ml/kg) cardioplegia. Core temperature cooled to 28 degree centigrade. After snugging Inferior vena cava and Superior vena cava, Right Atrium was opened and anatomy inspected. Left heart was vented through Patent Foramen Ovale . As the annulus was narrow a

main pulmonary arteriotomy extending down across the infundibulum was made. Pulmonary valve appeared severely stenosed hence valvectomy was done. The RVOT (Right ventricular outflow tract) was cored out from atrium and MPA , bundles were cut and annulus was sized up to 10 Hegars, adequate for patient's body surface area. Large sized subaortic VSD with ˜50% aortic override was present. It was closed with a Dacron patch using 5-0 polypropylene. Doty's type II APW measuring 10 mm was present (Figure 3A). It was closed with 0.4 mm Gortex patch (Figure 3B) with a continuous 6-0 polypropylene suture through PA. Monocusp pulmonary valve was constructed using 0.1 mm Gortex membrame. The proximal MPA and RVOT were augmented with autologous pericardial patch. The heart was de-aired and gradually weaned off bypass in sinus rhythm with injection Milrinone @0.5 mic/kg/min and Adrenaline@ 0.1 mic/kg/min. Intraoperative trans esophageal echocardiography revealed intact VSD patch, no shunt across the great arteries and normal biventricular function. RVOT gradient was 20 mmHg with mild pulmonary regurgitation (PR). She was extubated after 24 hours. Her post-operative echocardiography revealed RVOT gradient of 23 mmHg, mild PR, no residual VSD, no residual APW and good biventricular function with RVSP of 28 mm Hg (Figure 2B) . At 1-month of follow-up, she was healthy and gaining weight.

DISCUSSION:

APW accounts for <1% of all CHD [(3).] Their clinical findings change according to the additional associated defect. These are aortic arch interruption (15-20%), especially type A; Patent Ductus Arteriosus (11%); VSD (8%); right aortic arch (7%); anomalies of the coronary arteries (8%); TOF (5%); subaortic stenosis (3%); bicuspid aortic valve[(4)] (3%). A study by Gowda et al. conducted in our Institute involving 55 patients of APW, over a span of 24 years no patient had associated TOF [(5)]. In one of the largest series spanning 40 years by Backer et al. out of 22 patients only 1 patient had associated TOF[(6)].

In TOF with APW, data of 13 out of 19 patients could be retrieved on extensive search of the literature, out of which 7 patients had associated PAH[(2,3,8)]. In view of raised PA pressures it was decided to operate as soon as this patient was diagnosed. Castaneda and Kirklin have also recommended repair of this combination as soon as the diagnosis is made.

The first transaortic patch closure was reported by Deverall et al. from Great Ormond Street in 1969. The first series of patients having a Dacron patch closure through a transaortic approach was reported by Clarke and Richardson in 1976. They noted that the transaortic approach is easy to close, allows the origin of the coronary arteries to be accurately visualized, and allows right PA reconstruction if necessary. The patch placed from the aortic side does not distort either the aorta or PA and is naturally self-sealing [(6)]. Erez et al completely separated the aorta and pulmonary artery and closed the defect in each artery with a different pericardial patch in majority of his patients. In this manner he carried out the repair under direct vision of the nearby structures in each artery. This approach was found to be safe as it precludes recurrence of the defect and minimizes the chance of distorting adjacent structures[(7)]. Crawford et al closed the communication with a patch from within the aorta followed by repair of TOF[(8)]. We preferred closing the APW through PA since it was easily accessible and pulmonary valve pathology had to be addressed. There was no deformity of great arteries noted after completion of repair. By this approach an incision and suturing of the aorta was also avoided.

CONCLUSION:

APW is associated with other defects in 50% of the cases, leading to delay in diagnosis. It should be excluded in children with other CHD's[**(4)**] . Missing the diagnosis may lead to death due to heart failure or PAH in a child who could undergo an early successful surgery and become normal. Early surgery i.e. before advancement of PAH is recommended as the outcome is good. As in our case, if the TOF repair requires opening the PA, we recommend closing the APW through PA incision thus avoiding aortotomy and an additional suture line.

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Critical revision of article, Approval of article: Dr Neelam B Desai, Dr J.Nageshwar Rao

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Figure Legends

Figure 1.(A) Chest Radiograph. (B) Aortic root angiogram showing distal APW. APW: Aorto-Pulmonary Window; MPA: Main Pulmonary Artery

Figure 2. Transthoracic short-axis view echocardiogram showing: (A) Preoperative image showing APW in relation to Aorta and pulmonary artery. Turbulent flow across the right ventricular outflow tract and APW is seen. (B) Post op transthoracic echocardiography showing intact APW patch. VSD: Ventricular Septal Defect

Figure 3: Intra operative photograph showing: (A) APW in relation to great vessels. (B) Gortex closure of APW.

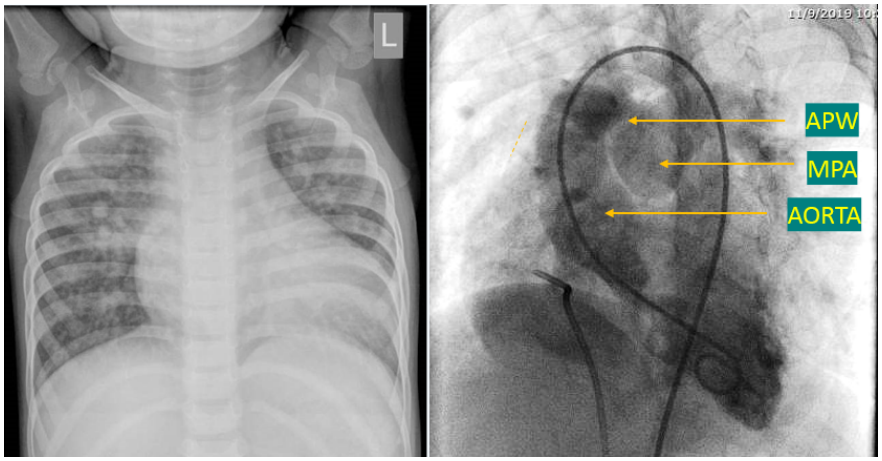

Figure 2A- Pre-operative ECHO

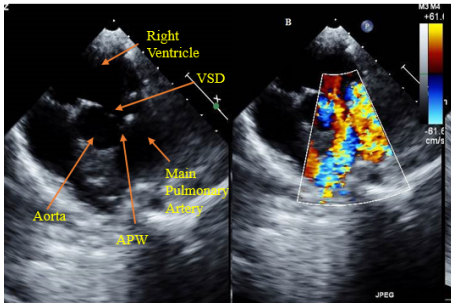

Figure 2B- Post-operative ECHO

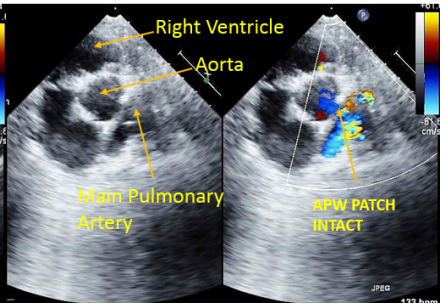

Figure 3A- Intraoperative Finding

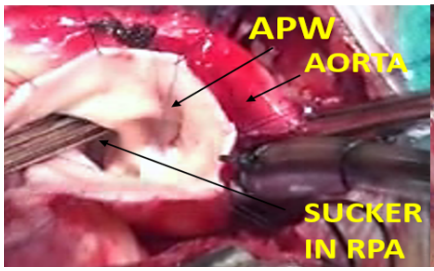

Figure 3B- Post Trans-PA APW patch closure