Unidirectional ventricular septal valved patch for repair of late presenting ventricular septal defect with aortopulmonary window

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ABSTRACT

Management of long standing left to right shunt lesion resulting in elevated pulmonary vascular resistance (PVR) is challenging. Limited surgical options are further complicated by an unpredictable postoperative period. Unidirectional valve patch (UVP) closure has shown to be useful in cases of the large ventricular septal defect (VSD) who present late. We report a case of large aortopulmonary window coexisting with a large VSD with severe pulmonary artery hypertension and significantly elevated PVR that was managed surgically by closure of the window by sandwich technique and closure of the septal defect with a UVP. This report emphasizes the importance of UVP in the management of such patients.

Keywords: Aortopulmonary window, pulmonary hypertension, valved patch, ventricular septal defect

INTRODUCTION

Late presentation of left to right shunts with severe pulmonary arterial hypertension (PAH) is a common problem in the low- and middle-income countries. We present a patient who was diagnosed with a large aortopulmonary window (APW) and a ventricular septal defect (VSD) with severe PAH at the age of 9 years. The child underwent successful closure of APW by sandwich technique and unidirectional valve patch (UVP) closure of the VSD through the right atrium using the technique described by us previously.[1]

CASE REPORT

A 9-year-old girl presented to the outpatient department with a history of recurrent respiratory tract infections since early childhood, delayed milestones, and easy fatigability. There was no cyanosis on examination. The pulse oximetry saturation was measured as 94% at rest which declined to 90% after exercise. There was no difference in the upper limb and lower limb saturations. Cardiomegaly was evident on cardiovascular examination. S1 was normal whereas S2 was loud with a narrow split; a grade 3/6 pansystolic murmur was audible at the left lower sternal border. In addition, a faint early diastolic murmur was audible at the left upper sternal border. Chest radiograph confirmed the presence of cardiomegaly and increased the pulmonary blood flow. Electrocardiogram showed normal sinus rhythm and biventricular hypertrophy. Narrow q waves were present in leads V5-V6. Transthoracic echocardiogram revealed situs solitus, levocardia with normal venoatrial connections, and concordant atrioventricular and ventriculoarterial connections. A large VSD was seen in the perimembranous region. In addition, a large APW was present. Color Doppler interrogation revealed low velocity bidirectional flow across VSD suggesting the presence of severe PAH. Low moderate tricuspid valve regurgitation was present with the predicted right ventricular (RV) systolic pressure measured at 90 mmHg. The left ventricle (LV)

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measured 38 (z score 1.3) and 28 mm (z score 2.0) during diastole and systole, respectively. The RV and LV systolic function was normal. Considering older age of the patient and inconsistencies in the clinical data, cardiac catheterization was performed which revealed elevated pulmonary vascular resistance (PVR) index which measured at 6.6 wood units on room air and reduced to 4.6 after administration of 100% oxygen. Qp/Qs increased from basal 2.5 to 3.3 following oxygen administration [Table 1]. In summary, catheterization data suggested possibly operable hemodynamics and therefore, surgical repair was planned. Nonetheless, considering the age of the patient complete closure of APW with UVP closure of VSD was contemplated.

Surgical repair

Anesthetic management

The child was premedicated with intramuscular injections of morphine (1.5 mg) Phenergan (7.5 mg) 30 min prior to shifting to the operating room. Inhalational anesthesia was induced using oxygen and air mixture with incremental inhaled sevoflurane. Subsequent maintenance was by using intermittent boluses of injection fentanyl, midazolam, vecuronium, and oxygen air mixture with 1-2% sevoflurane. Intraoperative transesophageal echocardiography (TEE) was performed using a S7-3t transducer on an iE33 system (Philips Medical System, Andover, Mass). The midesophageal four chamber view with color flow Doppler showed a perimembranous VSD measuring 1.9 cm with left to right shunt [Figure 1]. The RV inflow outflow view in color compare mode showed the APW 1.86 cm in size with flow across the defect into the main pulmonary artery (PA) [Figure 2] and the VSD with left to right shunt. High pressure tricuspid regurgitation (TR) jet was noted with a peak gradient of 86 mmHg. The coronary sinus measured 1.45 cm, and the presence of a left sided vena cava opening into the right atrium via the coronary sinus was speculated which was subsequently confirmed using bubble contrast echocardiography.

Surgical management

Surgical approach was via a standard median sternotomy. The external cardiac anatomy was typical for an APW. The right and the left PAs were dissected and looped. After systemic heparinization, the aorta was cannulated as high as possible, and both the vena cavae were cannulated. Both the pulmonary arteries were tightly snugged to prevent run off and cardiopulmonary

Table 1: Data obtained on cardiac catheterization

Chamber	Room air	Postoxygen
Saturations (%)		SaO_2 (PaO_2)
SVC	65	67 (54)
RA	65	
RV	77	
PA	87	92 (154)
PV (assumed)	98	100 (250)
LV	91	
FA	91	98 (203)
Pressures (mmHg)		
RA	Mean 7	Mean 6
RV	110 ed 12	110 ed 12
PA	102/52/78	112/42/70
PCWP	14	14
LV	120 ed 14	120 ed 14
Aorta	120/58/88	116/49/84
FA	120/55/87	122/54/88
Calculated variables		
Qpi (L/min/m^2)	9.7	12.6
Qsi (L/min/m^2)	3.9	3.7
Qpi/Qsi	2.5	3.4
PVRI (WU/m^2)	6.6	4.4
SVRI (WU/m^2)	20.5	20.9
PVRI/SVRI	0.3	0.2

SVC: Superior vena cava, RA: Right atrium, RV: Right ventricle, PA: Pulmonary artery, PV: Pulmonary vein, LV: Left ventricle, FA: Femoral artery, PCWP: Pulmonary capillary wedge pressure, SaO_2: Saturation (%), PaO_2: Partial pressure of oxygen (mmHg), ed: End diastolic, Qpi: Indexed pulmonary flow, Qsi: Indexed systemic flow, PVRI: Pulmonary vascular resistance index, SVRI: Systemic vascular resistance index, WU/m^2: Wood units per m^2

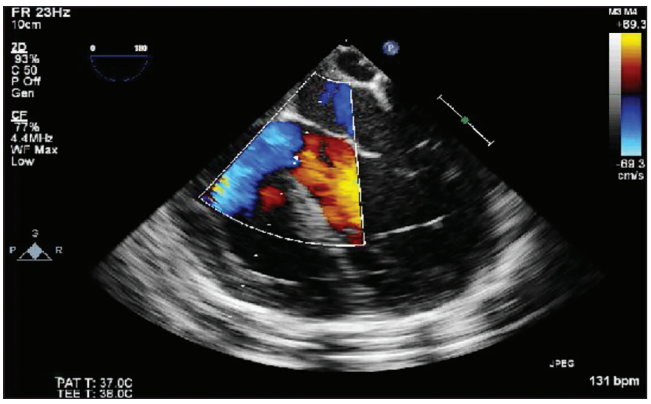

Figure 1: Midesophageal four chamber view with color flow Doppler showing sub aortic ventricular septal defect with a left to right shunt

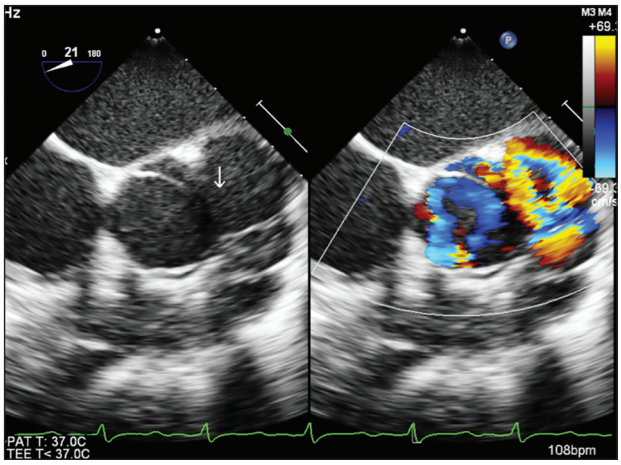

Figure 2: Midesophageal right ventricular inflow outflow view in color compare mode showing the aortopulmonary window and the left to right flow across the defect into the main pulmonary artery

bypass (CPB) was instituted with core cooling to 32°C. Del Nido's cardioplegia was administered antegradely through the aortic root. The window was then opened and the defect was identified [Figure 3]. The window was closed using a Gortex (WL Gore and Assoc, Flagstaff, AZ) patch with running sutures by sandwich method taking care not to damage the aortic and pulmonary valves and the origin of the right coronary artery that was very close to the window. Following this, the right atrium was opened and the large perimembranous VSD was located. The return from the left superior vena cava was managed by placing a cardiotomy sucker into the opening of the coronary sinus. In view of the presence of severe PAH, the VSD was closed with a UVP using the technique described by us earlier.[1,2] The tricuspid valve was repaired by placing interrupted stitches at the anteroseptal and posterior commissures. The right atrium was de-aired and closed. CPB was weaned slowly and the patient was separated from CPB on injection dobutamine 10 mcg/kg/min and injection nitroglycerine 0.5 mcg/kg/min. CPB and aortic cross clamp time were 102 min and 84 min, respectively. Immediately after surgical repair, TEE was performed to confirm the adequacy of the surgical repair and this revealed no residual flow across APW. There was mild RV dysfunction with low moderate TR and predicted RV systolic pressure measured at 50 mmHg. The simultaneous aortic pressure was 80/38 mmHg while the pressure in the right atrium was 7 mmHg. The flow across UVP showed right to left shunt that was limited to diastole [Figure 4]. The saturation in the postoperative period remained consistently above 95% although there were episodes of systemic desaturation with the saturation dipping to 85-90%; these episodes were transient and correlated with right to left shunting through the UVP on transthoracic echocardiography. At 1 month of follow-up, the patient is in NYHA class I and is on diuretics and oral sildenafil. Echocardiograms reveal normal biventricular function with no residual defect. And no left to right or right to left shunt. The systemic saturation at rest and on exercise is above 95%.

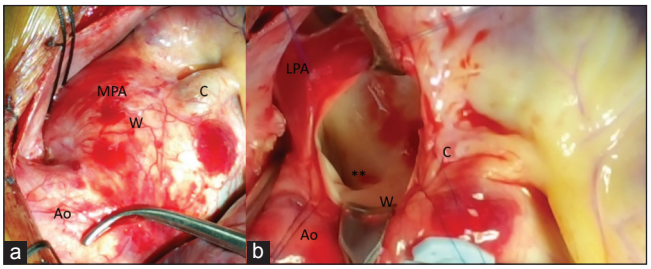

Figure 3: Intraoperative surgical image (a) external anatomy (b) after opening the window. Figure shows posterior margin of the aortopulmonary window (W), openings of the Right pulmonary artery (**), left pulmonary artery (LPA), aorta (Ao). Origin of the right coronary artery is also seen (C)

DISCUSSION

Surgical correction of a large left to right shunts in children with elevated PVR is associated with significant morbidity and mortality secondary to long standing severe PAH which leads to either bidirectional or reversed shunting across the defect. Conventional methods of complete patch closure may not be tolerated by such patients due to elevations of PA pressures and RV decompensation in the postoperative period.[3] Conventional options for these patients have been the use of nitric oxide and/or extracorporeal membrane oxygenation rescue.[2-4] These are expensive and not universally available. Surgical options in these patients include leaving a small atrial septal defect open, partial closure of the VSD with a fenestrated patch and a subsequent closure of the remaining defect by surgical or interventional methods.[5] or a single staged unidirectional flap-valve patch repair.[1-4,6-9] We have successfully demonstrated the favorable effect of UVP on the immediate,[1] early,[7-9] and midterm clinical outcomes[6] and hemodynamic parameters[9] in patients with borderline operability. It has become a standard clinical practice for the senior author (ST) to offer this method of management to all patients where operability may be questionable. In our case the presence of a large AP window in addition to the large sub aortic VSD further aggravated the degree of PAH, but irreversible pulmonary vascular disease was probably absent as demonstrated by the cardiac catheterization data. However, the patient showed the evidence of the right to left shunting through the UVP after terminating CPB that justified the use of the UVP in this situation. It is anticipated that the UVP will function as a safety-valve and allow decompression of the RV in the event of elevations of PA pressures. Moreover, because the patch is unidirectional and allows flow only from right to left, there would be no left to right shunt

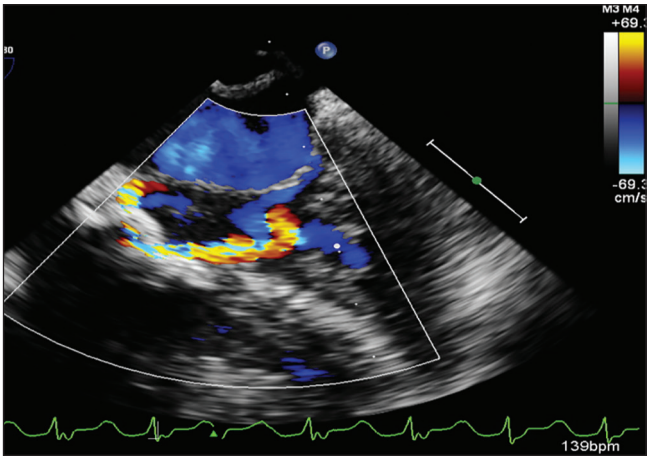

Figure 4: Postoperative midesophageal four chamber view with color flow Doppler shows right to left shunt across the unidirectional flap valve used to close the ventricular septal defect

in the follow-up after the PA pressures normalize. This would prevent a reintervention that would otherwise be needed with a fenestrated patch. TEE played a pivotal role in determining the adequacy of surgical repair, appropriate functioning of the UVP, assessment of RV function, and tailoring inotropic/medical management appropriate to the patient condition.

Although the strategy of a UVP in the setting of severe PAH may be argued as not being novel, we believe that such patients will continue to test our diagnostic and management acumen and every such case needs to be reported in detail to add to the body of knowledge of the anticipated benefits versus accrued benefits with the use of UVP which is still not universally practiced and questioned by many. This may be helpful in disseminating this knowledge to more members of the cardiovascular community and encourage them to practice this further.

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Conflicts of interest

There are no conflicts of interest.

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