SAM - A Saviour in Transposition of Great Arteries

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Abstract

The arterial switch operation (ASO) is the procedure of choice for the management of d-transposition of the great arteries (TGA). However, the surgical management of infants older than 6 weeks with TGA and intact ventricular septum (IVS) remains contentious. We report a case of late presenting TGA, IVS with Systolic anterior motion(SAM) of mitral valve with preserved left ventricle(LV) and its management.

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Abstract:

The arterial switch operation (ASO) is the procedure of choice for the management of d-transposition of the great arteries (TGA). However, the surgical management of infants older than 6 weeks with TGA and intact ventricular septum (IVS) remains contentious. We report a case of late presenting TGA, IVS with Systolic anterior motion(SAM) of mitral valve with preserved left ventricle(LV) and its management.

Key words: Transposition of great arteries, Arterial switch operation, Systolic anterior motion

Introduction:

Patients with TGA, IVS presenting after the neonatal period are at high risk for ASO in view of concerns regarding the preparedness of LV to support the systemic circulation. In such cases, either an atrial switch operation or a rapid two-stage arterial switch procedure has been usually performed, though the former is inferior to ASO[1,2]. Some centers advocate a primary ASO with standby extracorporeal membrane oxygenation (ECMO) support[3]. We report our experience with a 2 year old girl with TGA, IVS with preserved LV mass and its management.

Case report:

This 2-year-old girl presented with the complaints of breathlessness and cyanosis and was diagnosed to have cyanotic heart disease and referred to our centre for management. On examination, her height was 65 cm, body weight 5 kg, pulse 124/ min, BP 103/64 mm Hg, and respiratory rate 35/min. There was clinical cyanosis, clubbing, but no pallor or jaundice. Oxygen saturation was 70% in room air. Echocardiography revealed normal situs, atrioventricular concordance and ventriculoarterial discordance, two fossa ovalis atrial

septal defect 8 mm and 4 mm each, muscle bundle seen bulging into left ventricular outflow tract(LVOT) causing Systolic Anterior Motion(SAM) of anterior leaflet of mitral valve with peak gradient 12 mmHg (Fig. 1) and normal bi-ventricular function. Since the LV posterior wall thickness and LV mass was normal(Fig. 2) and there was circular cross-sectional appearance of LV in the short-axis view, patient was planned for primary ASO.

With parental consent, a primary ASO was performed with closure of the ASD. Cardiopulmonary bypass time was 241 min and ischemic time of 127 min with two doses of del Nido cardioplegia and moderate hypothermia. The child was successfully weaned from CPB with supports of milrinone 0.8 micrograms/kg/min, norepinephrine 0.1 micrograms/kg/min, epinephrine 0.05 micrograms/kg/min and returned to ICU with sternum left open in view of severe LV systolic dysfunction. ECMO was kept standby, However, by the simple intervention of vasodilatation and accepting low systemic blood pressure (mean BP of 45 mm Hg), its use was avoided. Chest closure was done on day 2. The poorly contractile LV necessitated prolonged ventilation and sedation along with extended inotropes and vasodilator infusion. She was extubated on postoperative day 7 and was discharged on 14th postoperative day and is on regular follow-up.

Discussion:

In spite of many published reports, little is understood about the origin of left ventricular outflow tract obstruction in simple TGA, IVS. Aziz and colleagues[4] described two types of obstruction in TGA and IVS namely, "fixed" in which there was no SAM of the mitral valve and in which the diameter of the LVOT was reduced compared with the diameter of the pulmonary artery through out the cardiac cycle and "dynamic", in which the left ventricular outflow tract was wide open during diastole but narrowed in systole and was associated with SAM of the mitral valve. In the index case there was solely dynamic obstruction of LVOT that has contributed in preservation of LV mass.

In TGA with IVS, LV cavity appears ellipsoid at birth but becomes banana shaped sooner corresponding to a decline in PVR[5]. The assessment of adequacy of LV to tolerate systemic circulation is one of the major evaluation criteria for ASO. LV end-diastolic volume calculated using area length method (Bullet method) on Transthoracic Echocardiography in subcostal or parasternal short axis and subcostal or apical long axis view is useful in calculating LV mass.[5] LV mass of <60% of predicted or of <35 gm/m2 with respect to body surface area suggests a regressed LV. Posterior wall thickness of LV measured at the mid-cavity level if <4 mm is also suggestive of regressed LV.[6] Furthermore, assessment of LV cavity from subcostal short axis views of the heart at end-systole can yield additional information on favourable LV for single-stage repair. LV geometry was classified as "favourable" or type I if the superoinferior/ anteroposterior dimension ratio was <2; "acceptable" or type II if the ratio was between 2 and 3 and "unfavorable" or type III if the ratio was >3.[7] In the present case it was favourable and LV posterior wall thickness was >4 mm.

ASO is found to be a better operation in patients of TGA, IVS with dynamic obstruction of LVOT unlike atrial repairs in which dynamic LVOT obstruction is likely to persist or progress, and can even develop postoperatively in some patients [8,9]. The other important thing is that the child was managed with adequate inotropes and vasodilator infusion without the use of ECMO. Very few cases have been reported in late presenting TGA, IVS in which the LV mass is preserved solely due to dynamic LVOT obstruction caused by SAM of mitral valve.

Conclusion:

Our experience supports the notion that the LV in TGA-IVS maintains the potential for systemic work well beyond the neonatal period even with dynamic LVOT obstruction. Though transient postoperative LV dysfunction almost always results in a prolonged postoperative course, its always reversible and therefore primary ASO should be the first surgical option in such cases.

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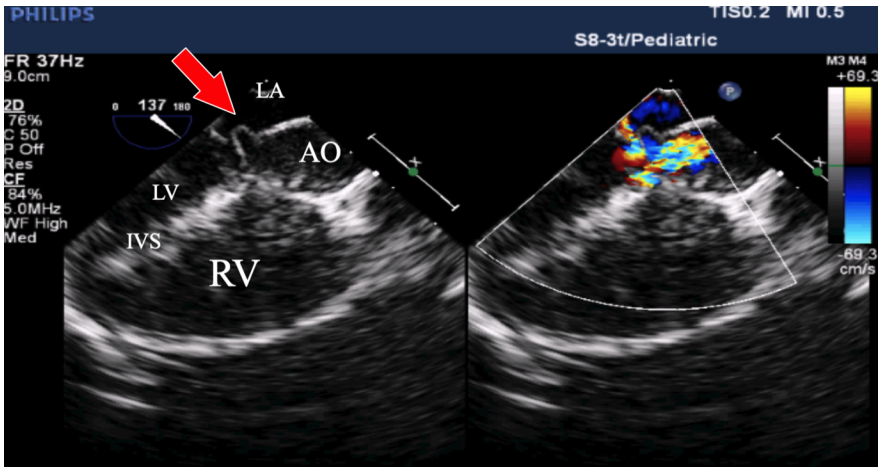

Fig. 1: Midesophageal long axis view image showing Systolic anterior motion of Mitral valve(Red arrow)

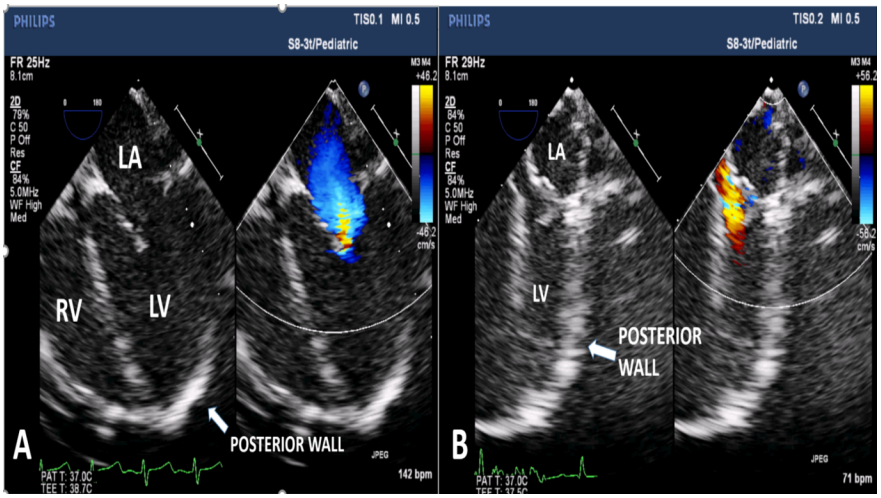

Fig. 2: Midesophageal 4 chamber view (A) and Modified Midesophageal view image showing preserved left ventricle with posterior wall thickness