Cardiology in the Young (2012), **22**, 113–115
doi:10.1017/S1047951111001004

Brief Report

d-Transposition of the great arteries in a 12-year-old child: is arterial switch still an option?

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Abstract Arterial switch surgery for d-transposition of great arteries is usually performed in the first 2 weeks of life before the left ventricle regresses. The arterial switch surgery that helps achieve anatomic and physiologic correction of this condition has better long-term outcomes than other surgical approaches. The procedure may still be attempted at an older age where the left ventricle has not regressed. We report a rare case of a 12-year-old child with d-transposition of great arteries, a remote muscular ventricular septal defect and isolated valvar pulmonic stenosis, which was corrected by an arterial switch surgery.

Keywords: Arterial switch; pulmonary valve stenosis; older children

Received: 6 May 2011; Accepted: 24 May 2011; First published online: 21 July 2011

Case report

A 12-year-old girl, diagnosed with cardiac disease in infancy, was referred to our institute for further evaluation and management. She had a normal birth history and was the third child of non-consanguineous parents. Her parents had noticed cyanosis at 3 months of age. She had undergone an elective modified right Blalock–Taussig shunt with a 6-millimetre Gore-Tex tube (W.L. Gore & Associates, Inc., Flagstaff, Arizona, USA) at 1-year of age. Cyanosis had been less pronounced since then, but she had been complaining of exertional fatigue and dyspnoea over the past few years.

On examination, she was of moderate build, with central cyanosis – pulse oximeter oxygen saturation (84%) – and clubbing. There were no dysmorphic features. Precordial examination revealed a diffuse apex and a harsh ejection systolic murmur in the left second and third intercostal spaces. Chest radiograph showed normal pulmonary vascularity and no cardiomegaly. Electrocardiogram revealed normal sinus rhythm, right axis deviation, and an incomplete right bundle branch block pattern.

Transthoracic echocardiography showed normal viscero-atrial situs and atrioventricular concordance with ventriculo-arterial discordance (S, D, D). There was a large mid-muscular ventricular septal defect with a bidirectional shunt, a patent foramen ovale with left-to-right shunt, and a small patent ductus shunting from left to right. There was left ventricular outflow tract obstruction with a peak gradient of

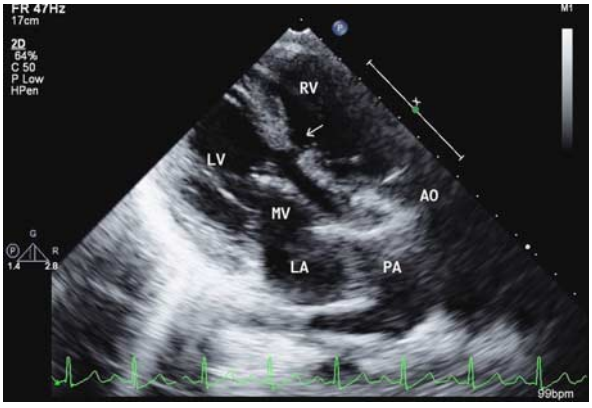

Figure 1.
Modified subcostal long axis view showing the remote mid-muscular ventricular septal defect and transposed great arteries.

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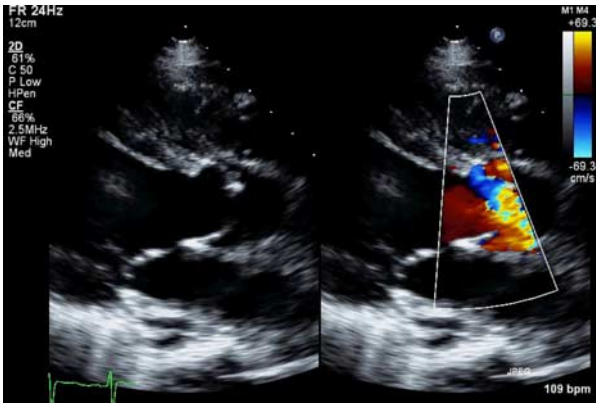

Figure 2.
Post-operative echocardiogram: parasternal long-axis view showing the repaired pulmonary valve in the neo-aortic position with mild neo-aortic regurgitation.

58 millimetres of mercury and turbulence beginning at the level of a bicuspid, thickened pulmonic valve. There was no subvalvar obstruction, and the branch pulmonary arteries were normal. The right ventricular outflow tract and aorta were normal, with preserved biventricular function. The left coronary artery arose from the leftward sinus and the right coronary from the rightward sinus of Valsalva. The modified right Blalock–Taussig shunt was functional.

A cardiac catheterisation was performed for better delineation of the coronary anatomy and location of the ventricular septal defect in relation to the aorta. Right and left ventricular pressures were 95/8 and 94/6 millimetres of mercury, respectively. The main pulmonary artery pressure was 28/12 millimetres of mercury. The left ventriculogram showed a mildly dilated, well-contracting left ventricle; a large mid-muscular ventricular septal defect opacifying the right ventricle and aorta; and no additional septal defects. There was no subvalvar narrowing. Doming of the pulmonic valve was noted and the branch pulmonary arteries were of a fair size. The aortogram showed a small ductus supplying the left pulmonary artery and a functional, modified right Blalock–Taussig shunt. The origin and course of the coronaries were similar to that noted on echocardiography – 1L Cx; 2R, Congenital Heart Surgeons Society nomenclature.

She underwent anatomic correction of the defect, involving closure of the ventricular septal defect with a Sauvage patch (Bard Inc., Tempe, Arizona, USA) and an arterial switch. Repair of the bicuspid pulmonary valve comprised excision of the pseudoraphe in the anterior leaflet, suturing of the small incomplete scallop in the anterior leaflet, and a Trusler hitch on the left side with a left commissurotomy. The tiny duct was ligated and the systemic pulmonary shunt was taken off. Post-operative echocardiography showed good biventricular function, a tiny residual leak across the ventricular septal patch, and a neo-aortic valve gradient of 29/19 millimetres of mercury with mild aortic regurgitation. She was discharged on the fifth post-operative day.

Discussion

Transposition of the great arteries is a frequent cardiac malformation, which is amenable to near-total corrective repair. Ventricular septal defect is the most commonly associated anomaly. Transposition of the great arteries with a ventricular septal defect causes left ventricular outflow obstruction in about 30% of cases. The ventricular septal defect may be perimembranous (33%), muscular (27%), malaligned (30%), inlet type (5%), or associated with conal septal hypoplasia (5%).[1] Most muscular defects are in the mid-septum.

Left ventricular outflow obstruction is most often subvalvar and may be a fibrous ring, a tunnel type of narrowing, or a muscular obstruction secondary to outlet septal malalignment. Pulmonary valvar stenosis is rare. Other uncommon causes of obstruction include malattachment of the anterior mitral valve to the outlet septum by anomalous chordae, redundant tricuspid valve tissue protruding through the ventricular septal defect, subpulmonic membrane, or aneurysmal dilation of the membranous septum.[2]

Surgical approaches to management of patients in this subgroup have usually included the Rastelli or *Réparation à l'étage ventriculare* procedures.[3] The Rastelli operation achieves anatomic correction and bypasses the left ventricular outflow obstruction. Conduit replacement is inevitable and long-term results with the Rastelli repair have not been good, with mortality rates of 59% at 20 years.[4] The *Réparation à l'étage ventriculare* procedure avoids a prosthetic extracardiac conduit and intracardiac tunnel obstruction. Transpulmonary or transmitral resections of a fixed fibromuscular subvalvar shelf, membrane or fibrous tags were also performed, followed by an arterial switch.

The problems unique to our case were older age at presentation, isolated pulmonary valvar stenosis, which was amenable to repair, and the mid-muscular location of the septal defect, which prevented rerouting of the left ventricle to the aorta. Complete anatomic correction with an arterial switch may still be the best option in d-transposition with ventricular septal defect and left ventricular outflow obstruction, if the outflow tract obstruction can be repaired and the left ventricle has not regressed.[5] The problems

https://doi.org/10.1017/S1047951111001004 Published online by Cambridge University Press

related to the conduit in a Rastelli procedure can be avoided. Patients need to be followed up for residual left ventricular outflow tract obstruction, neo-aortic regurgitation, myocardial perfusion abnormalities, and left ventricular function.

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