Stenting of ventricular septal defects to retrain the left ventricle in patients with transposition of the great arteries and restrictive ventricular septal defect

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The arterial switch operation (ASO) is the standard treatment for patients with dextrotransposition of the great arteries (d-TGA) with a prepared left ventricle (LV). We recently encountered an 8-month-old infant with d-TGA, restrictive ventricular septal defect (VSD), and a regressed LV who underwent a successful ASO after an initial VSD stenting procedure. This novel approach is described.

CLINICAL SUMMARY

An 8-month-old infant weighing 5.5 kg presented with cyanosis since birth. Pertinent clinical findings included central cyanosis, a loud aortic component of the second heart sound, and a grade 4/6 pansystolic murmur best heard at the apex. Chest radiograph showed the typical “egg on side” appearance of the heart with evidence of increased pulmonary blood flow. Transthoracic echocardiography was consistent with a diagnosis of d-TGA with a 3- to 4-mm size restrictive muscular VSD with a gradient of 70 mm Hg across it. The LV was regressed as suggested by its typical banana shape with the posterior wall measuring only 2.7 mm (Figure 1). On cardiac catheterization the LV pressure was 33/7 mm Hg compared with the right ventricular (RV) pressure of 83/10 mm Hg. Because there was no patent arterial duct (PDA) and the VSD was restrictive we decided to place a stent across it in an attempt to prepare the LV for an eventual ASO.

Approach was through the right femoral vein. A straight tip Terumo guide wire (Terumo Corporation, Tokyo, Japan) of 0.025-in diameter was passed via the femoral vein into the right atrium and then across the atrial septal defect into the left atrium and the LV. From LV, the VSD was identified, and the guide wire was negotiated through the VSD and entered into the RV. Over the Terumo wire, an RCA Judkin's JR4 catheter (Cordis de Mexico, Chihuahua, Mexico) was passed into the RV. The RCA catheter was removed and exchanged with a 6F renal guiding catheter (having a 4F

Postventricular septal defect stent echocardiographic image.

Central Message

A new approach to retrain the left ventricle in patients with d-TGA and a restrictive VSD is described.

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multipurpose catheter inside as mother-and-child technique to smoothly negotiate the crossing) parked into the RV. The straight-tip Terumo catheter was removed and exchanged with a BMW 0.014 guide wire (Abbot Vascular, Santa Clara, Calif) required for the stent. Next a 5 × 15 mm Express vascular SD stent (Boston Scientific, Natick, Mass) was introduced and placed across the VSD. Position was confirmed with fluoroscopy and echocardiography and the stent was deployed at 10 atm pressure then postdilated at 12 atm. After stent deployment, echocardiography showed patent flow through the stent and the gradient across the VSD was 49 mm Hg (previously it was 70 mm Hg). LV systolic pressures improved from 33 mm Hg to 45 mm Hg. Saturation of peripheral oxygen improved from 65% to 85%.

Before attempting the VSD stenting, PDA stenting was considered but the occluded ductus could not be crossed by the straight tip Terumo 0.025 guide wire, hence the procedure was abandoned in favor of the VSD stenting procedure. The patient received 10 mg clopidogrel as a loading dose the night before the procedure and after the procedure continued to receive 4 mg clopidogrel once daily. Low-molecular-weight heparin was administered for 10 days after stenting.

LV parameters gradually improved. The Doppler gradients did not change remarkably from the day of stent, but the LV shape appeared more circular with time. The

FIGURE 1. A, Prestent echocardiogram (parasternal long axis view) shows the typical banana-shaped left ventricle (LV). B, same view immediately after the stent procedure. C, The stent (*) is seen positioned across the ventricular septum. M-mode echo 1 day after the stent procedure shows a reduction in gradient across the ventricular septal defect. RV, Right ventricle.

posterior wall thickness of the LV started increasing and was 4.48 mm at 3 weeks and the cavity became more circular with the interventricular septum moving with the LV (Figure 2). It was therefore decided to perform the ASO 39 days after VSD stent placement.

At surgery the coronary pattern was 1L2RCX. Through the right atrium and after retracting the tricuspid valve, the stent was pulled out from the septum and the VSD was closed. Considerable difficulty was encountered during removal of the stent and it took nearly 20 minutes of ischemia time. This was followed by the ASO. The cardiopulmonary bypass time was 186 minutes and the aortic crossclamp

time was 106 minutes. Postoperative recovery was uneventful except for need for prolonged ventilatory support due to chest infection that necessitated a tracheostomy. At 3 months follow-up, the patient is off all cardiac medication. Follow-up echocardiograms have demonstrated no residual defects and biventricular function is normal.

DISCUSSION

This 8-month-old infant presented difficult management options. The simplest option would have been to perform an ASO. Although the early results of this procedure are excellent, it is associated with a definite incidence of progressive RV dysfunction and arrhythmias.¹ The other option is to retrain the LV by performing a pulmonary artery band and a systemic to pulmonary shunt and then performing a rapid or delayed 2-stage arterial switch.² This may necessitate multiple reoperations to achieve a balance between the preload and afterload to the LV, thereby increasing the costs. Finally ASO followed by mechanical LV assist³ is another method that has found increasing application in recent times. In developing countries, however, the prohibitive costs of this procedure and standards of intensive care unit care are limiting factors.

The concept used with our patient is quite similar to that of stenting of the PDA that was first reported by Sivakumar and colleagues⁴ and subsequently used by us as well.⁵ However, patients in both these reports had a PDA that could be

FIGURE 2. M-Mode echocardiogram 3 weeks after a stent procedure shows marked increase in left ventricle dimensions.

stented and had an intact interventricular septum. In contrast, our patient did not have a PDA and the small muscular VSD was restrictive.

It is possible that the patient could have undergone a primary ASO with a similar outcome. But the improvements in LV parameters afforded a certain degree of safety that we believe helped the outcome. The intraoperative difficulties in removing the stent were substantial and perhaps might decrease with more experience. The appropriate size of the stent remains unclear. A larger VSD could perhaps be more helpful. Creating or increasing the VSD size by using cutting balloons and stent has been reported for different clinical applications. The Boston group⁶ reported in detail stenting of the systemic ventricular outflow tract and creation or enlargement of a VSD using stents as potential therapeutic options in rare patients with congenital heart disease who develop significant ventricular hypertension due to outflow obstruction. We are not aware of any previous report of use of this strategy to retrain the LV in d-TGA.

CONCLUSIONS

A regressed LV may be prepared for ASO by stenting a muscular VSD in carefully selected patients. To the best of our knowledge, this approach has not been reported earlier.

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EDITORIAL COMMENTARY

A new approach to retrain the deconditioned left ventricle in transposition of the great arteries

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Delayed presentation of transposition of the great arteries can be a challenging issue when the patient is seen late with a deconditioned left ventricle. These patients require a fair bit of judgment regarding how best to proceed, and every patient needs to be treated as an individual case. The authors Talwar and colleagues¹ from New Delhi present a novel solution that uses stenting of a restrictive ventricular septal defect (VSD) to retrain the left ventricle before proceeding with an arterial switch procedure.

Typical solutions for these patients include banding of the pulmonary artery for “retraining” of the left ventricle and planning for temporary mechanical support after an arterial

Paul J. Chai, MD

Central Message

A new method is presented for retraining the deconditioned left ventricle in transposition of the great arteries by stenting the VSD.

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