Cardiology in the Young

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Arterial switch operation and aortic valve replacement for transposition of great arteries in adulthood: two cases from a tertiary care centre in India

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Brief Report

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Abstract

The surgical treatment of transposition of the great arteries, ventricular septal defect, and significant left ventricular outflow tract obstruction continues to evolve. The survival of an unrepaired transposition of the great arteries into late adulthood is a rarity. Even when large intracardiac shunts are present, it remains a lethal cyanotic CHD if it is not surgically corrected soon after birth. We present our experience of two cases, both of whom underwent a single-stage arterial switch operation and an aortic valve replacement for this defect.

Surgical decision-making and management of patients with transposition of the great arteries and left ventricular outflow tract obstruction with ventricular septal defect, who are not amenable for standard arterial switch operation, has been a significant challenge. Arterial switch operation with left ventricular outflow tract muscle resection may offer an immediate anatomical correction of the great arteries; however, there is a considerable risk of reintervention for the left ventricular outflow tract and neoaortic valve, if the mechanism of left ventricular outflow tract obstruction is complex.

The repair can be performed by different procedures: the Rastelli procedure, the Réparation à l'Etage Ventriculaire (REV) operation, the Metras modification, or the operations that were described by Bex and Nikaidoh. An atrial switch procedure (Senning/Mustard) may be an option depending on the degree of pulmonary stenosis that will remain. A left ventricle-to-pulmonary artery conduit has been described but is not a desirable option[1]. A bicuspid and obstructive pulmonary valve is sometimes amenable to valve repair after which an arterial switch operation may offer a good solution.[2] The anatomy usually allows for biventricular repair, but in some hearts only univentricular palliation is possible.

Case report

Case-1

A 20-year-old gentleman, presented to our outpatient department, with complaints of dyspnoea on exertion (NYHA-III) and cyanosis since birth interspersed with cyanotic spells occasionally but never took a formal medical opinion. There was no history of rheumatic fever or any significant cardiac history in the family. He was found to have uniform central cyanosis, Grade III clubbing with peripheral saturation of 85%. Blood pressure was 100/60 mmHg in the right arm. There was mild cardiomegaly, with the right ventricular type of apex. The first sound was normal with wide and fixed split-second sound. The pulmonary component was louder than the aortic component. There was no additional S3/S4. There was a grade 2/6 ejection systolic murmur at the aortic outflow.

A 12-lead Electrocardiogram showed sinus rhythm and right ventricular hypertrophy. Two-dimensional echocardiography images (Fig 1A-C) showed a large inlet ventricular septal defect and moderate pulmonary stenosis with a peak gradient across the pulmonary valve of 40 mmHg with good-sized ventricles. The right ventricle was giving origin to the aorta, and the left ventricle was giving origin to the main pulmonary artery (MPA). Although a catheter study (Fig 1G-I) is not routinely indicated in transposition of the great arteries, we proceeded with the catheter study in view of the age of the patient to assess the operability and confirmation of the diagnosis. Anterior ventriculogram showed aorta arising from the morphological right ventricle whereas the posterior ventriculogram showed MPA arising from the morphological left ventricle. There was a large subpulmonic ventricular septal defect noted with usual coronary

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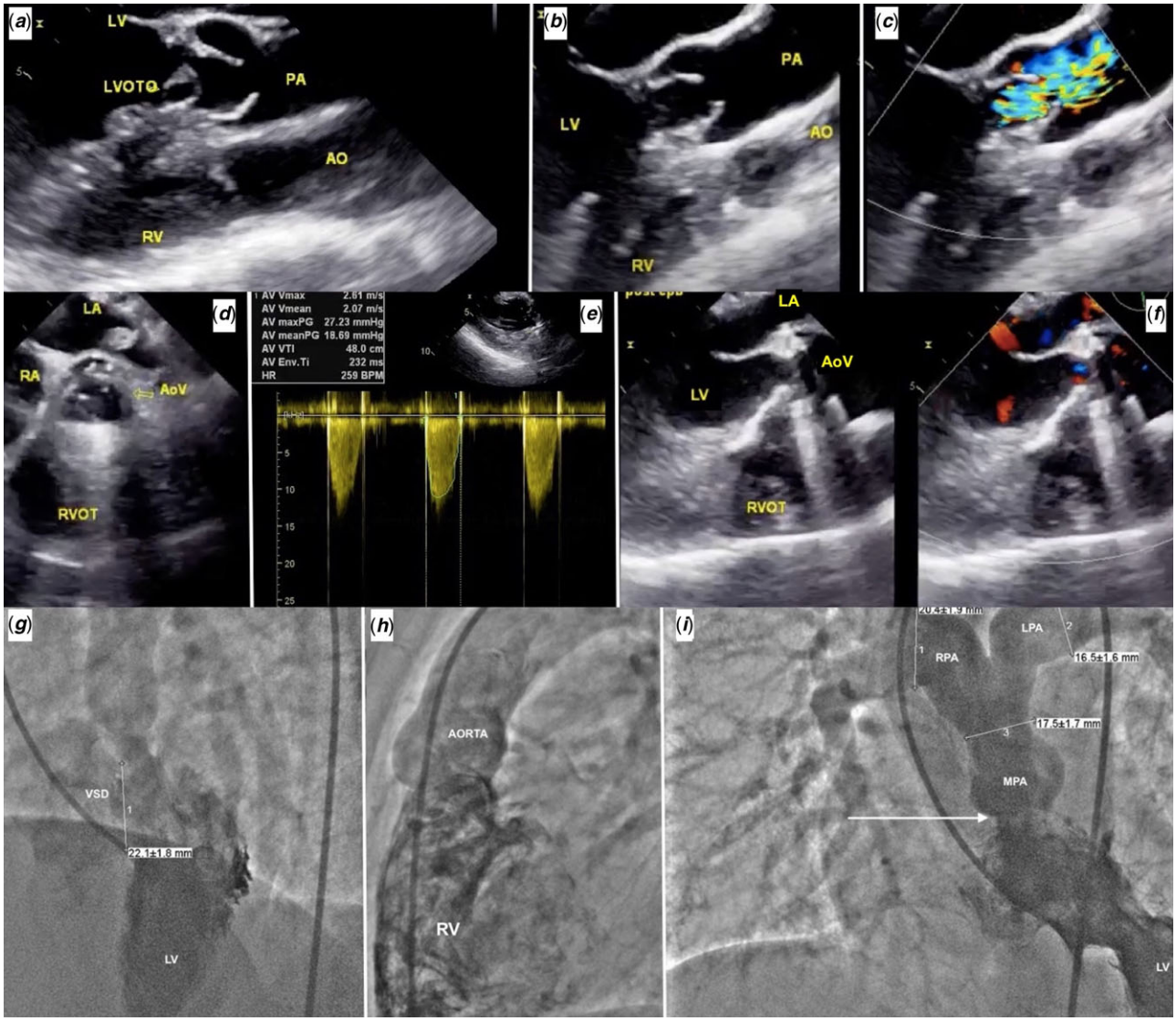

Figure 1. Pre-operative trans-esophageal echocardiography: (***a***) Mid-esophageal Long axis view showing a membrane in left ventricular outflow tract just below the pulmonary valve, (***b* and *c***) Mid-esophageal Long axis view showing turbulence at the pulmonary valve in systole, suggestive of stenosis, Post-operative transesophageal echocardiography images – (***d***) Mid-esophageal aortic valve short axis view showing mechanical aortic valve in place, (***e***) Trans-gastric long axis view showing a mean gradient of 18.69 mmHg on applying continuous wave Doppler across mechanical aortic valve, (***f***) Mid-esophageal valve long axis view showing washing jets across the mechanical aortic valve and no turbulence on applying colour Doppler, Pre-operative catheter angiography: (***g***) LAO Cranial view showing the large VSD (22m) with the finely trabeculated left ventricle (LV), (***h***) Coarsely trabeculated ventricle (RV) giving origin to the aorta anteriorly, (***i***) LAO Cranial view showing pulmonary artery arisisng from the fine trabeculated ventricle (LV) with confluent branch pulmonary arteries and narrowing at the valvular level (arrow).

artery pattern for transposition of the great arteries. The pulmonary angiogram showed confluent and good-sized branch pulmonary arteries and valvular pulmonary stenosis. Left ventricle and right ventricle angiogram were suggestive of normal biventricular systolic function with pressures of 100/21 mmHg and 120/16 mmHg, respectively. The pulmonary artery pressure was 52/26 mmHg with a peak systolic gradient of 48 mmHg across the left ventricular outflow tract. There were no major aortopulmonary collaterals. The operation was performed under hypothermic (28 °C) cardiopulmonary bypass with aortic and bicaval cannulation. The heart was arrested in diastole using Del Nido cardioplegia. The neo-aortic valve was bicuspid, thickened, and stenosed necessitating neo-aortic valve replacement with 17 mm mechanical valve prosthesis (St. Jude Medical Regent™ Mechanical Heart Valve) (Fig 1D-F) Jatene's arterial switch operation with the Lecompte maneuvre was performed. The right atrium was opened and the ventricular septal defect repaired through the tricuspid valve with a PTFE patch (PolyTetra Fluoro Ethylene) (Bard, Impra® ePTFE Cardiovascular Patch) using a continuous 5–0 polypropylene suture. The sternum was closed and the patient was shifted to ICU on standard inotropes. The patient was discharged on post-operative day 7. The patient is doing well in the follow-up clinic.

Case-2

An 18 years of age lady presented to the hospital with similar complaints of cyanosis and dyspnoea. Two-dimensional echocardiography images showed a large subpulmonic ventricular septal defect and moderate pulmonary stenosis with a peak gradient across the pulmonary valve of 60 mmHg. The right ventricle was giving origin to the aorta, and the LV was giving origin to

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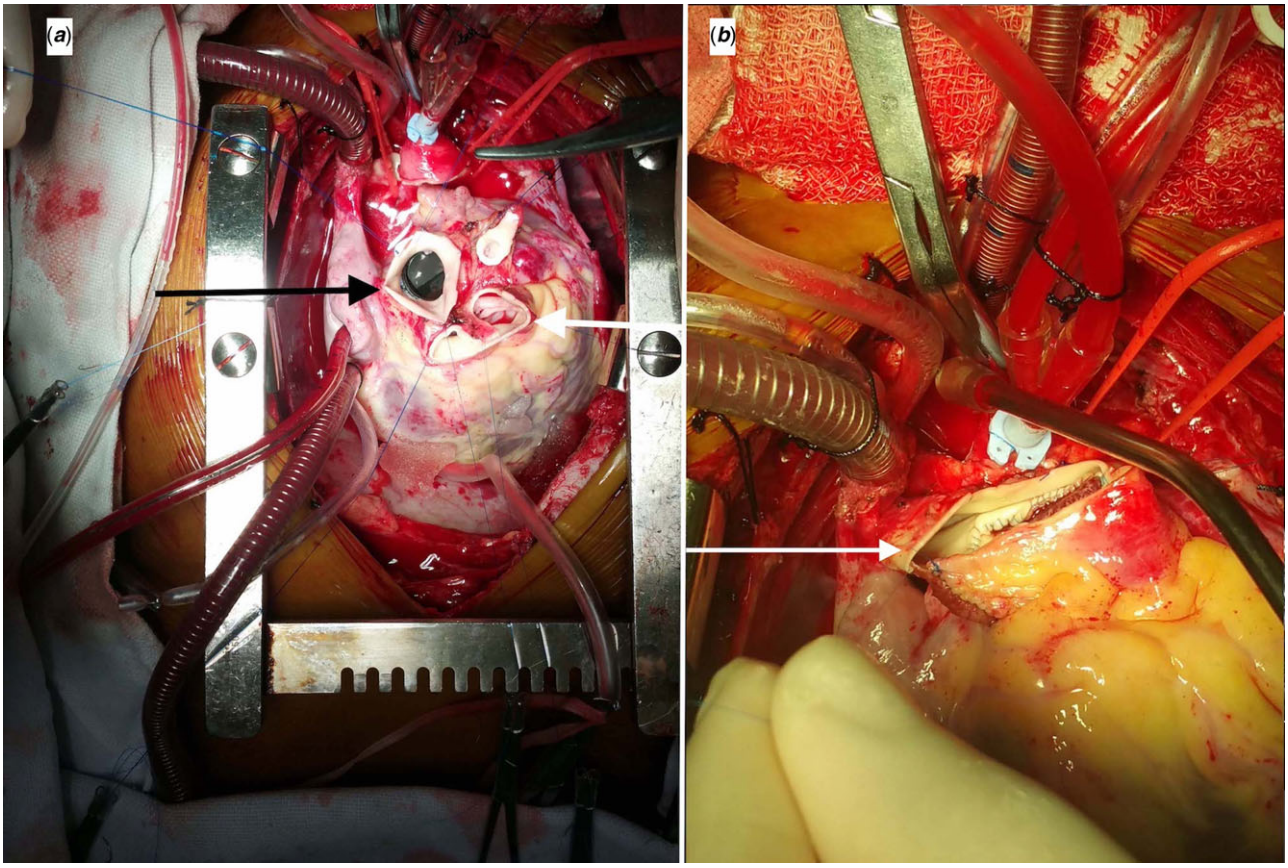

Figure 2. Intra-operative image – (***a***) Showing the neo-aorta with the mechanical aortic valve in situ (black arrow), neo-pulmonary artery with the harvested coronary buttons (white arrow), (***b***) neo-pulmonary artery (white arrow) anastomosis being completed using autologous pericardial patch.

the MPA. Conventional catheter angiography was suggestive of transposition of the great arteries, large outlet ventricular septal defect, and valvular pulmonary stenosis. Descending Thoracic Aorta angiogram showed one significant MAPCA which was coiled 1 day prior to surgery. The patient underwent an arterial switch operation with aortic valve replacement using 17 mm mechanical valve prosthesis (St. Jude Medical Regent™ Mechanical Heart Valve) in a similar manner as described above (Fig 2). The patient was discharged on post-operative day 8 and is currently doing well, 7 years after her surgery.

Discussion

The combination of TGA, pulmonary stenosis, and ventricular septal defect encompasses a wide spectrum of anatomic relationships and physiologic presentations. The ventricular size and function; great vessel dimensions and spatial relationships; size and location of the ventricular septal defect; the presence, severity, and constitution of left ventricular outflow tract obstruction; atrioventricular valve structural abnormalities; and coronary anatomy must be understood and considered. Each of the above variables guides the optimal choice of intervention.

In transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction, the position of the great arteries and the coronary anatomy can vary. Ventricular septal defects may be committed or non-committed and can be small or large. Left ventricular outflow tract obstruction can vary greatly.[3] When the pulmonary valve is big enough and functional, an arterial switch operation with ventricular septal defect closure and left ventricular outflow tract obstruction resection is possible. With pulmonary valve hypoplasia or severe valvular dysfunction, an arterial switch is no longer an option. The left ventricle must be either tunnelled to the aorta, or the aorta must be translocated to the left ventricular outflow tract. The Rastelli operation was the first procedure (1968) to correct transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction, with the left ventricle as systemic ventricle.[4]

Although the Rastelli operation has been the most widely performed surgical procedure during the past decades, several studies have shown its suboptimal long-term prognosis. Several authors have reported that the long-term results of the Rastelli procedure are not satisfactory. The two main issues are the left ventricle to aortic tunnel leading to subaortic stenosis and reduced right ventricle volume and the extracardiac right ventricle to pulmonary artery conduit which is less durable than the orthotopically placed conduit.

Following the Rastelli operation, various procedures have been described by Bex and Nikaidoh, to correct this anomaly. The REV (Réparation à l'Etage Ventriculaire) was developed by Lecompte and modified by Metras and Kreitmann later on. More radical techniques have been reported by Yamagishi and by Mair: an autologous half-turned truncal block that involves both semilunar valves, which are fully detached from the heart and turned 180° before they are re-inserted with the aortic valve on top of the left ventricular outflow tract and the pulmonary valve over the right ventricular outflow tract obstruction.

Aortic valve competence represents the most important complication in the long-term patient's outcome after arterial

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switch operation and aortic root translocation procedures. The timing of the operation, however, is a big concern, given that the left ventricle rapidly regresses in the presence of low pulmonary artery pressure, which was a major challenge in our cases. In both our cases, the pressure in the left ventricle was maintained owing to the unrestrictive ventricular septal defect and some degree of left ventricular outflow tract obstruction. Also, the left ventricular outflow tract obstruction was helpful in protecting the pulmonary circulation. Surgical correction in a neonate is preferred and the delay was found to be associated with increased mortality. Even when large intracardiac shunts are present, it still remains a lethal cyanotic CHD if it is not surgically corrected soon after birth.[5,6] In our patients, the neo-aortic valve was stenotic with thickened leaflets which precluded repair, and we had to proceed with mechanical valve replacement. The ultimate success is determined by long-term follow-up with specific attention paid to potential long-term complications, such as pulmonary, aortic, or coronary artery stenosis and aortic or pulmonary valve regurgitation.

Conclusion

Transposition of the great arteries with a ventricular septal defect and left ventricular outflow tract obstruction surviving into adulthood without any intervention is next to impossible. Patients benefit most from an individualised surgical approach. There is not one single technique that fits all patients with this anomaly but aortic valve replacement with arterial switch operation is an option in such cases.

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Conflicts of interest. None.

Ethical standards. As per our institutional protocol, ethical clearance is not required for a case report.

Consent. Written and informed consent was taken from the patients and their parents regarding the publication of this case

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