Case Report: Favoring biventricular repair beyond the conventional boundaries with primary arterial switch operation in a young adult [version 1; peer review: awaiting peer review]

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Abstract
Transposition of great arteries (TGA) presents in neonates or in infancy. We report a case of TGA with ventricular septal defect (VSD) and pulmonary stenosis (PS) in an adult male patient of 23 years age. Arterial switch operation with VSD closure and neo-aortic valve replacement was done. The patient recovered well in the post-operative period. In adult patients, conversion from atrial to arterial switch has been widely reported, both directly and after prior pulmonary artery banding in two stages, but primary arterial switch for TGA has not been reported previously. In this patient there was a benefit of having a large VSD and severe PS.

Keywords
Grown Up Congenital Heart disease, arterial switch operation

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Introduction

Transposition of great arteries (TGA) presents in neonates or in infancy\(^1\). Recently, we managed an adult patient with cyanotic congenital heart disease [TGA with sub pulmonic ventricular septal defect (VSD) with severe pulmonary stenosis (PS)] with complete biventricular repair by performing an arterial switch operation.

Case report

A 23-year-old man presented with complaints of bluish discoloration of skin and nails from childhood, associated with frequent attacks of pneumonia and dyspnea on exertion, off and on from school going age. He was deeply cyanosed and had grade 3 clubbing. Pulse oximetry showed 76% saturation in room air, not improving on oxygen supplementation. Auscultation revealed an ejection systolic murmur over the upper left sternal border, and normal heart sounds. Bilateral vesicular breath sounds were heard equally, and the abdomen was soft with no palpable organomegaly.

Echocardiography revealed ventriculoarterial discordance: aorta arising from right ventricle (RV) and pulmonary artery (PA) from left ventricle (LV); aorta found to the right and anterior to PA; evidence of severe valvular PS (gradient-83 mm Hg), with large sub-pulmonic VSD; normal biventricular function. The initial plan of management was for Rastelli procedure with valved conduit, and the patient was planned for cardiac catheterization study which suggested 92% PA saturation and 66% femoral artery saturation, PA pressure of 73/31 mm Hg and PVRI of 7.9 wood units, pulmonary flow Qp 4.3, systemic flow Qs 5.3, Qp/Qs-0.8. LV angiography suggested large S/A routeable VSD, no additional VSD, and confluent PAs. Prior to surgery, an echocardiogram was repeated and the patient reassessed: VSD was found to be non-routeable, with normal biventricular function. Therefore, the patient was planned for arterial switch operation (ASO) with VSD closure and pulmonary valve replacement.

Surgical correction was performed via median sternotomy, on cardiopulmonary bypass using high ascending aortic and bicaval cannulation under mild hypothermia. Cold blood cardioplegia (Delnido) with no topical cooling was used.

Peroperative findings were as follows: aorta was present anterior to and to the right of PA and arising from RV (anterior ventricle) and PA from LV (posterior ventricle); moderate sized sub pulmonic VSD; pulmonic valve was bicuspid with calcification; no atrial septal defect (ASD).

The PAs were dissected up to the lung hila after division of the ductus arteriosus. The aorta was cross-clamped, cardioplegia given to the aortic root, and the aorta transected. Coronary buttons were harvested from the aortic sinuses together with a variable amount of sinus wall using minimum dissection. The main PA was transected 3 to 4 mm below the bifurcation. Bicuspid and calcific cusps excised and valved sized and seated with 17 MM SJM Regent valve. One of the pulmonary valve cusps had a thrombus, which was completely removed. This was causing an outlet obstruction for the systemic ventricle (future LV) at the valvular level. The coronary buttons were re-implanted into the neo-aortic sinus rather than above the sino-tubular junction. Trans aortic PTFE patch VSD closure was done. Neo-aortic anastomosis was completed after LeCompte maneuver. Right atrium opened to check for ASD, with no ASD was present. Autogenous fixed pericardial patch reconstruction of the neo-pulmonary root was done. Aortic cross clamp was released, root vented and neo-pulmonary anastomosis was carried out during rewarining phase. Patient was weaned off from bypass. Aortic decannulation was done after giving pro-tamine (single dose of 1.5mg/kg in order to neutralize Heparin given at 1mg/kg).

Postoperative recovery was uneventful in the ICU. Inotropes (Dobutamine at 10mic/kg/min slowly tapered and stopped within 24hours), antibiotics (Inj. Cefotaxime 1g twice a day for first 24hrs postoperatively) and supplements (nutrition) were followed as per unit policy. On the third postoperative day, with active mobilization, the patient had complaints of occasional dizziness and headache. Detailed ENT and neurological evaluation showed no anatomical substrate for his symptoms. His symptoms gradually subsided over a period of few days.

Discussion

The conventional age boundaries for cardiac surgery for complex cyanotic cases have been pushed previously\(^2\). We report a singular experience of a 23-year-old young adult with congenital TGA (with VSD and PS) who successfully underwent a complete repair – ASO with transaortic VSD closure with neo-aortic valve replacement.

Whereas most case reports of ASO in adults\(^3\)–\(^5\) prior to ours have reported cases of converting and atrial switch to arterial switch operation after prior LV retraining as a two stage ASO, we report a successful primary arterial switch in this adult patient.

Massimo et al. in 2000\(^6\) reported a case (Table 1) with moderate PA hypertension and preserved LV function operated for ASO. In their case, moderate pulmonary hypertension played...

<table>
<thead>
<tr>
<th>Case</th>
<th>N</th>
<th>Age at 1st operation</th>
<th>Initial procedure</th>
<th>PA Band</th>
<th>Arterial switch operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Massimo et al.(^3)</td>
<td>1</td>
<td>Infancy</td>
<td>Mustard operation + ventricular septal defect closure</td>
<td>Yes</td>
<td>6 months after PA banding + at age of 23 years</td>
</tr>
<tr>
<td>Cochrane et al.(^4)</td>
<td>24</td>
<td>14 months</td>
<td>Atrial switch (Senning/Mustard)</td>
<td>In 15 cases</td>
<td>12 cases: staged switch 4 cases: direct switch</td>
</tr>
<tr>
<td>De Jong(^6)</td>
<td>2</td>
<td>6 weeks and 4 months</td>
<td>Balloon atrial septostomy + Mustard</td>
<td>No</td>
<td>Conversion to arterial switch</td>
</tr>
</tbody>
</table>
a role in preserving some degree of LV hypertrophy. Di Donato reviewed the literature on ASO in older children and adults post PA banding. The longer the morphological LV works at low pulmonary pressure, the lower is the capability of inducing LV hypertrophy by PA banding. Hence the mature myocardium may have poor response to pressure overload.

Our patient had good LV morphology and function due to the presence of two favorable factors – a large VSD and presence of severe PS that provided afterload to morphologic LV, which is otherwise not achieved in cases without severe PS. Thus, these two factors ensured that a primary ASO with VSD closure and neo-aortic valve replacement was adequate in this young man.

Consent
Written informed consent was obtained from the patient for the publication of the report of his case.

Data availability
All data underlying the results are available as part of the article and no additional source data are required.

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References

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