Institutional report - Congenital
Morphosurgical correlation of outcomes in complete double outlet right ventricle

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Received 6 April 2006; received in revised form 23 May 2006; accepted 29 May 2006

Abstract

Even though surgical experience with wide spectrum of double outlet right ventricle (DORV) is available, the experience with extreme form of complete DORV where both great arteries completely arise from the right ventricle is limited. We present our experience with this unique subset where the systemic outflow is far removed from the interventricular foramen and hence, the systemic ventricle. In this situation, biventricular repair can be technically demanding and challenging. Between June 2002 and February 2006, 12 patients underwent biventricular repair of this subset. The VSD was subaortic in all; eight patients had infundibular and valvar obstruction. Aorta was anterior and to the right, with the pulmonary artery far posterior and to the left. The interventricular foramen was patched with a long Gore-Tex patch to route the LV flow to the aorta. Eight patients had infundibular resection and right ventricular outflow tract (RVOT) enlargement with an autologous monocusp pericardial patch. No patient required a valved conduit. There was no operative mortality. In one patient, there was a small residual VSD that was not of haemodynamic significance. There was no RVOT and left ventricular outflow tract obstruction and no RV inflow obstruction. The early and mid-term results are good.

Keywords: Double outlet right ventricle; Biventricular repair; Congenital; Morphogenesis

1. Introduction

When both great arteries arise from the right ventricle in association with VSD, it is defined as double outlet right ventricle (DORV). The spectrum ranges from 50% override of the systemic artery to both arteries completely arising from the right ventricle [1, 2]. However, Lacour-Gayet et al. have tried to separate the more extreme forms of DORV as 200% depending on the degree of deviation of the posterior outlet towards the right ventricle [3]. Surgically, the experience with this complete DORV subset is limited as most studies have always described the DORV spectrum, which is variable. We present our experience with this unique subset among DORV with sub-aortic VSD, where the systemic outflow is far removed from the interventricular foramen and hence, the systemic ventricle. In this situation, the biventricular repair is technically demanding.

2. Material and methods

2.1. Patient profile

Between June 2002 and February 2006, 12 patients underwent biventricular repair for the extreme (complete) form of DORV variant. There were 6 males and 6 females. The age ranged from 2 months to 6 years (median 2.5 years). The weight ranged from 6–14 kg (median 8.5 kg). The clinical presentation was cyanosis in eight patients and failure to thrive with recurrent upper respiratory tract infection in four patients.

The diagnosis was established by 2D echocardiography in all patients. The aorta was anterior and to the right and pulmonary artery was far posterior and to the left. The VSD was sub-aortic in all patients. Assessment of distances relative to the arterial trunks and VSD, along with tricuspid valve to pulmonary artery and tricuspid valve to aorta were measured. Even though the VSD appeared sub-aortic and easily routable in the sagittal view (Fig. 1), it was found to be far removed from the aorta in the sub-costal four-chamber view (Fig. 2). Eight patients had infundibular and valvar obstruction. No patient required cardiac catheterization for establishing the diagnosis.

2.2. Operative technique

All the patients underwent repair through median sternotomy. On cardiopulmonary bypass (CPB) with moderate hypothermia and aortic root cold blood cardioplegia, the right atrium (RA) was opened and VSD inspected. All patients had an extreme form of DORV, wherein the aorta and pulmonary artery arose completely from the right ventricle (Fig. 3). The VSD was large and sub-aortic with
sub-aortic conus. The interventricular communication was closed transatrially with a long Gore-Tex (W.L. Gore & Associates, Inc., Arizona, USA) patch to re-route the left ventricle flow to the aorta (Fig. 4). In one patient, the VSD margin was not accessible through routine ventriculotomy for RVOT enlargement. So, a separate ventriculotomy was made on the superior aspect of the right ventricle, below the aorta, to complete the closure. The width of the patch was matched to be equal to the diameter of the aorta. The length was tailor-made to bridge the inferior margin of interventricular communication to the anterior margin of aorta. Care was taken to prevent tricuspid inflow, right ventricular and left ventricular outflow tract obstruction. In eight patients, the main pulmonary artery was hypoplastic and the pulmonary valve was stenosed and bicuspid (Fig. 5). In these patients, infundibular resection was done through RA and the main pulmonary artery (MPA) was incised longitudinally and extended across the pulmonary annulus onto the right ventricular outflow tract (RVOT). On beating heart, the RVOT was enlarged with an autologous monocusp pericardial patch in eight patients.

Fig. 2. Sub-costal four-chamber view of the same patient as seen in Fig. 1. Note the anterior and removed position of the aortic valve. Ao – Aorta; PA – Pulmonary Artery; STL – Septal Leaflet of Tricuspid Valve; TV – Tricuspid Valve.

3. Results

All patients were weaned off CPB with minimal inotropic support. The post CPB, RV to LV pressure ratio ranged from 0.4 to 0.6 (mean of 0.5). All patients were ventilated post-operatively, for a period of 20 to 68 h (median 25 h). The median ICU stay was 2.5 days (range 2.0–4.0 days). The median hospital stay was 8 days (range 7–12 days).

There was no operative mortality. 2D echocardiography was done before discharge in all patients. In one patient, there was a small residual ventricular shunt at the level of the attachment of the septal leaflet of the tricuspid valve. This was of no haemodynamic significance. There was no significant RVOT and LVOT obstruction or right ventricular inflow obstruction. All patients demonstrated normal ventricular function. The follow-up ranged from 2 months to 34 months (median of 20 months). All patients were in NYHA class I during follow-up.

4. Discussion

The definition of DORV remains controversial. According to Anderson et al., all of one great artery and 50% or more of the other artery must arise from the right ventricle to qualify for a diagnosis of DORV [1,2]. They contend that the development of this spectrum of hearts with abnormal ventriculoarterial connections is explained by departures from the normal development of the cushions of the bulbo-ventricular loop. Conal malrotation, changes in the position of
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Fig. 4. Patching of the inter-ventricular foramen with a long Gore-Tex patch to re-route left ventricle flow to aorta.

Fig. 5. Two hundred percent DORV with severe infundibular and valvular obstruction. DORV – Double Outlet Right Ventricle.

of the anterior portion of the muscular interventricular septum and differential conal absorption form the basis of their hypothesis [4].

Le Compte has proposed to change the terms DORV to malposition of the great arteries. Neufeld opined that, although the presence of aorto-mitral discontinuity and bilateral coni are important descriptors, they should not serve as absolute pre-requisites for the diagnosis of DORV [5]. The morphology of DORV is encompassed by a careful description of the ventricular septal defect (VSD) with its relationship to the semilunar valves, the great artery relationships to each other, and the tricuspid–pulmonary annular distance [6]. Recently, Lacour–Gayet has proposed a new nomenclature for this unique subset, where both great arteries arise totally from the RV (200%), in association with a VSD [3]. In essence, both arteries are completely arising from the right ventricle. However, in several earlier papers from their group they have clearly mentioned that the critical aspect of DORV has been the relationship of the VSD vis-a-vis the arterial trunks and whether there is sub-arterial narrowing related to the muscular outlet septum [7].

The anatomical feature in this rare subset is quite evident in our patients by the sub-costal sagittal and four-chamber planes by echocardiography. Interestingly, the VSD is sub-aortic and easily routable in the sagittal view while it is far removed from the aorta in the sub-costal four-chamber view. This feature has been well illustrated in Figs. 1 and 2. Also, measurements of the distances of the tricuspid valve to pulmonary valve/aortic valve and VSD to aorta/pulmonary valve should be carefully performed to assess if the VSD is routable to the aorta. These views were diagnostic of the extreme form of DORV, which was confirmed at the time of surgery. Surgical repair has to be entirely tailored according to this, as obstruction to the systemic ventricular outlet as well as pulmonary outlet is entirely dependent on this anatomical feature as well as patch closure of VSD.

The standard surgical procedure for DORV with a subaortic VSD is the intra-ventricular tunnel by re-routing LV to the aorta (as described above) or the pulmonary artery [3,7]. We have used Gore-Tex flat patch to re-route the left ventricular blood flow to the aorta, contrary to the standard surgical technique of using a half cut tube graft as a intra-ventricular tunnel [8]. In extreme forms of DORV, where both great arteries completely arise from the RV without any commitment to LV, the closure of the maligned VSD is impossible as the intention of surgical repair is to only re-route the interventricular communication to the subarterial muscular outlet septum (Anderson et al., personal communication). However, the margins of the interventricular communication are difficult to access, and along with poor visualization, results in residual interventricular shunts. During closure of the interventricular communication, the length and width of the patch should be adequate. If it is too short, it can cause left ventricular obstruction.

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outflow tract (LVOT) obstruction. If it is too large, it can cause RV inflow and outflow obstruction [9,10]. Since a vast majority of the complete DORV has associated RVOT obstruction, a variety of approaches such as infundibular patch, annular patch or conduits have been employed [11,12]. Even in our cohort, RVOT augmentation was necessary and we used an autologous monocusp pericardial patch. This provides a way to preserve the native valve and prevents free pulmonary regurgitation which helped during the immediate postoperative period. This strategy enabled us to avoid a valved conduit in our patients.

5. Conclusion

There exists a subset of DORV with sub-aortic VSD where both great arteries completely arise from the right ventricle (200% DORV). Because of its unique nature, it requires to be identified. Biventricular repair in this subset is technically demanding. Tailoring the VSD patch with the width being equal to the diameter of aorta helps to prevent RVOT, LVOT and RV inflow obstructions. The technique reduces the need for conduit. The early and mid-term results are good.

Acknowledgments

We thank Mr. Dhanabalan, the artist, and Mr. Ravi, the secretary for their help in preparing the manuscript.

References