Case report - Congenital

Biventricular repair of complete atrioventricular canal, double-outlet right ventricle and common atrium using a modified double switch technique. A valid alternative to univentricular procedure

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Abstract

An eleven month-old child underwent a successful modified double switch operation for total correction of complete atrioventricular canal, double outlet right ventricle, noncommitted ventricular septal defect, pulmonary stenosis, common atrium and anomalous left superior vena cava to an unroofed coronary sinus. We describe the technique of modified double switch operation, utilizing an atrial switch combined with a Rastelli type reconstruction between the venous ventricle and the pulmonary artery.

Keywords: Double outlet right ventricle; Double switch; Non-committed

1. Introduction

Biventricular repair of double-outlet right ventricle (DORV) with noncommitted ventricular septal defect (VSD) suppose a formidable surgical challenge. The results are conditioned mainly on the feasibility to perform complex baffle procedures connecting the left ventricle to the aorta through an enlarged VSD [1]. The results are poor and a high risk of reoperation exists to treat complex subaortic stenosis. Moreover a subgroup of patients exist with restrictive inlet VSD and complex tricuspid attachments in the outlet septum in which these intraventricular baffles repairs must be abandoned after surgical exploration. They need to be converted to an univentricular repair [2]. We present a successful single stage repair using a modified double switch operation in a patient with complete atrioventricular canal (CAVC), DORV with noncommitted VSD, pulmonary stenosis (PS), common atrium, anomalous systemic and pulmonary veins and unroofed coronary sinus. This technique allowed a single-stage biventricular repair avoiding a univentricular operation.

2. Case report

A male infant 11-month-old, weighing 7 kg was referred to us with mild cyanosis and the diagnosis of CAVC, DORV with complete origin of the great vessels from the right ventricle, aorta right anterior oblique with regard to the pulmonary artery trunk, pulmonary valvar and subvalvar stenosis, common atrium with double superior vena cava, continuation inferior vena cava-azygous-left superior vena cava (SVC) to an unroofed coronary sinus, suprahepatic veins draining independently to the floor of the common atrium and moderate insufficiency of the common valve (visceral heterotaxy syndrome).

At cardiac catheterisation, the pulmonary artery pressure was 24/15 mmHg with a mean of 18 and the Qp/Qs was 1.8/1. The pulmonary arteries were smaller than normal with a Nakata index of approximately 165 mm²/m² and arterial oxygen saturation of 86%.

A nuclear magnetic resonance was deemed necessary to best visualize venous drainages to the common atrium and assess the possibility of atrium septation. Echocardiography revealed a long distance between the inlet VSD and subaortic conus, with numerous cordal attachments of the left structures of the common valve crossing the subaortic pathway (Rastelli type C).

The operation was performed on cardiopulmonary bypass and hypothermia to 18 °C. Both SVCs and suprahepatic veins were cannulated. After cross-clamping the aorta, the atrium was opened and the intracardiac anatomy explored. There was an inlet type VSD with a common valve straddling the ventricular crest and abnormal chordae attachments to the outlet septum, a heavy conal septum and a long and restrictive subaortic conus. All these anatomical arrangements render the intraventricular baffle repair unfeasible. There was also a common atrium with a left SVC draining between the left pulmonary inferior vein and left lateral leaflet of the common valve, with no coronary sinus. A right SVC and suprahepatic veins were draining anterior to the right pulmonary veins (Fig. 1). It was impossible to perform the septation of the atrium without left SVC.
translocation. This translocation was impossible due to a huge azygous vein draining the blood from the inferior vena cava, which could not be sacrificed.

A modified double switch operation was planned. The CAVC was repaired by closing the ventricular component with a patch of heterologous pericardium, implanting the superior and inferior components of the atrioventricular valve in the superior edge of the patch, but leaving more valve tissue on the right side. The cleft on the right valve was completely closed and competence tested. An autologous pericardial patch (ostium primum patch), was sutured to the fibrous rafe of the common valve just over the ventricular patch and around the walls of the common atrium. The suture was extended only 5 mm over the valvular plane. With a second autologous pericardial patch (Mustard patch), a Mustard procedure was performed allowing the pulmonary veins to drain into the right atrioventricular valve and systemic veins into the left atrioventricular valve. The atrial septation was then completed suturing this patch over the superior edge of the ostium primum patch. Finally a modified Rastelli operation was performed: The main pulmonary artery was transected and proximally oversewn. A 14 mm jugular bovine heterograft conduit (contegra®), was interposed between the left ventricle and distal pulmonary trunk (Fig. 2).

The aortic cross-clamp time was 172 min and cardiopulmonary bypass 235 min. A second degree atrioventricular heart block was alternating with sinus rhythm. Postoperative recovery was uneventful. The infant was discharged from the hospital on postop day 15 in good clinical condition and permanent sinus rhythm. Echocardiogram showed trivial right atrioventricular valve regurgitation, normal pulmonary and systemic veins drainages and a mean gradient of 23 mmHg across the left ventricle-pulmonary artery connexion.

3. Discussion

Reports of surgical repairs of patients with CAVC associated to DORV with noncommitted VSD are, to our knowledge, very scarce. They are frequently treated by an univentricular repair [2,3]. Complex baffle procedures to connect the left ventricle to the aorta suppose a formidable surgical challenge [4], with a considerable number of cases being converted to a univentricular repair after intracardiac exploration. Moreover, a high rate of reoperation exists for progressive complex subaortic stenosis [5].

Our patient was thought to be a bad candidate for univentricular repair. A double Glenn operation could invert the flow through the azygous vein towards the hepatic veins in the near future, with the risk of progressive desaturation. The completion of a Fontan circulation would be less than attractive because of the small size of the pulmonary arteries and independent drainages of the suprahepatic veins in the middle of the floor of the common atrium.

We present a successful single physiologic biventricular repair with a modified double switch operation, as a valid alternative to univentricular repair, in those patients in which an anatomical biventricular repair is unfeasible or very challenging. The anatomical characteristics of our patient precluded an anatomical biventricular repair, not only at ventricular level, but also at auricular level, because the left SVC could not be either detached from the left atrium and anastomosed to the right atrium or directly tunnelled to the right atrium, without causing pulmonary vein obstruction and left atrioventricular valve distortion.

Since the functional state of the patients following univentricular repairs decline with time, we elected this physiologic biventricular repair as a better alternative [6]. Of course, long-term follow-up will be necessary to assess the function of the systemic right ventricle and the performance of the heterograft conduit, to finally validate this technique over univentricular repairs in this complex group of patients.

References

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Appendix A. ICVTS on-line discussion

Author: Jose I. Aramendi (Hospital de Cruces, Spain)

eComment: The paper of Caffarena and colleagues deals with biventricular repair in complex DORV, complete AV canal in the setting of heterotaxy syndrome. Those children are usually not amenable to biventricular repair and are driven to the Fontan pathway. In this report they use the concept of double switch repair in the opposite way as in L-TGA, that is, not to rescue the left ventricle for the systemic circulation but to achieve a biventricular repair at the expense of a systemic circulation driven by the right ventricle. In my opinion this is not an ideal solution; it is a complex operation and offers little if any advantage over the Fontan palliation. There is something even worse than a univentricular heart and that is a biventricular heart with a RV in the systemic circulation as in L-TGA. In this situation if there is not a subpulmonary obstruction, the LV becomes hypotensive and there is a shift of the IV septum towards LV producing RV dilatation and so-called left sided Ebstein’s disease. This situation rapidly leads to intractable right ventricular failure. I suggest reading the excellent paper by Koh et al.

References