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Two-Patch Repair of Rastelli's Type-A Complete Atrioventricular Septal Defect, Pulmonary Valvotomy and Relief of Right Ventricular Outflow Tract Obstruction under Mild Hypothermic Extracorporeal Circulation and Cardioplegic Arrest: A Video Presentation

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Abstract

The association of right ventricular outflow tract obstruction with complete atrioventricular septal defect is rare and poses surgical challenge at the time of intracardiac repair. We report here-in a 30-months-old female child diagnosed with severe right ventricular outflow tract obstruction, complete atrioventricular septal defect and severe left atrioventricular valvar regurgitation undergoing successful pulmonary valvotomy, resection of the right ventricular outflow tract with reconstruction of the complete atrioventricular septal defect by two-patch technique. The pulmonary valve ring was sutured directly. Postoperative period was uneventful.

Introduction

The atrioventricular septal defects encompass a spectrum of cardiac malformations unified by a deficiency of the atrioventricular membranous and muscular septa¹⁻³. The deficiency of the atrioventricular septum results in an ostium primum atrial septal defect and a scooped-out area in the inlet (basal) portion of the ventricular septum¹⁻³. Abnormal differentiation and remodelling of the cushion mesenchyme into valvulo septal tissue results in a deficiency or absence of the atrioventricular septum⁴.

At the extreme end of the spectrum lies the complete atrioventricular septal defect characterized by large deficiencies of the atrial and ventricular septa, and a common atrioventricular valvar orifice¹⁻³. Anatomically, all types of atrioventricular septal defects share the following features. These includes: i) absence of wedging of the aortic annulus due to a common atrioventricular valvar ring causing an elongated left ventricular outflow tract and a "goose neck" appearance; ii) lengthened outlet septum-to-ventricular apex ratio; iii) apical displacement of the attachments of the atrioventricular valves to the ventricular crest; and iv) posterior and inferior displacement of the coronary sinus resulting in inferior displacement of the atrioventricular node and elongation of the common atrioventricular bundle^{1-3,5-7}.

Published literature documents a number of morphological factors that complicate the "usual" form of atrioventricular septal defect. These include: i) double outlet left or right atrium⁸; ii) double orifice left atrioventricular valve⁹; iii) parachute deformity of the left atrioventricular valve¹⁰; iv) variable degrees of underdevelopment

of either the right or left ventricle (prevalence of 10%)^{11,12}; and v) variable degrees of left ventricular outflow tract obstruction^{10,13}. Right ventricular outflow tract obstruction complicating complete atrioventricular septal defect has rarely been reported.

Based on the variable anatomical relationship between the superior bridging leaflet with its chordal attachments and the underlying ventricular septum, Rastelli from Mayo Clinic classified complete atrioventricular septal defect into three types of severity¹⁴. In type A defect (69%), the common superior bridging leaflet is committed almost entirely to the left ventricle, and its commissure with the anterior tricuspid leaflet lies along the right anterosuperior rim of the ventricular septum. In the rare type B (9%), the superior bridging leaflet is larger and overhangs the ventricular septum. An anomalous papillary muscle attaches from the right side of the ventricular septum to the left side of the superior bridging leaflet, thereby creating a free interventricular communication. In type C (22%) defects, the superior bridging leaflet is larger, "free-floating", overhangs the ventricular septum more than in type A or type B without chordal attachment to the ventricular septal crest. The anterior tricuspid leaflet is generally small, supported by the medial papillary muscle¹⁴⁻¹⁶.

Patients with complete atrioventricular septal defect without associated cardiac anomalies presenting with congestive heart failure should be operated between 3 and 6 months of age to prevent progressive pulmonary arterial hypertension, congestive heart failure and Eisenmenger's syndrome¹⁷. The principles of surgical repair of complete atrioventricular septal defect include closure of the atrial septal defect, closure of the ventricular septal defect, creation of non-obstructive competent atrioventricular valves, and avoidance of damage to the atrioventricular node and bundle of His^{15,17,18}.

Literature documents three different surgical techniques for repair of complete atrioventricular septal defects. The techniques include: i) the single-patch technique, as described in 1962 by Maloney and Gerbode^{18,19}; ii) the two-patch technique as described by George Trusler in 1976²⁰; and iii) the modified single-patch technique of Graham Nunn in 1995^{21,22}.

Classic single-patch technique

Although a pericardial, polytetrafluoroethylene or Dacron patch have been used as the patch material, use of pericardium is associated with pericardial patch aneurysm at the ventricular level and the Dacron patch carries the risk of postoperative hemolysis^{18,19,23}.

Two-patch technique

In this technique, the ventricular septal defect is closed

using either a Dacron or polytetrafluoroethylene patch and the atrial septal defect is closed using a separate pericardial patch²⁰.

Modified single-patch technique

In this technique, a series of pledget supported 5-0 polypropylene sutures are placed on the right-side of the interventricular septum. These sutures are then passed sequentially through the middle of the superior and inferior bridging leaflet at a site predetermined to separate the mitral and tricuspid valves. Subsequently, these sutures are then placed through the edge of the autologous pericardial patch^{21,22}.

The operative mortality reported in the published literature varies between 3% and 15%^{17,24,25}. The reoperation rates for left atrioventricular valve regurgitation varies between 6% and 9% in different institutions^{17,24,25}.

In 1995, Backer and associates performed a meta-analysis of results among 794 patients undergoing operation by two-patch technique from five institutions⁷. The operative mortality and the incidence of reoperation for mitral regurgitation were 7% and 8% respectively⁷. Graham R Nunn and Nicholson reported their results using modified single-patch technique among 72 consecutive patients. The operative mortality was 2.8%. Postoperative mild and moderate left atrioventricular valve regurgitation was present in 29% and 5% of patients respectively^{21,22}.

We report here-in the surgical reconstruction of the complete atrioventricular septal defect using two-patch technique, pulmonary valvotomy, and resection of the right ventricular outflow tract obstruction. A 30-months old female child presented with poor weight gain, recurrent episodes of chest infection and cyanosis since birth. Clinically, she had cardiomegaly, left parasternal heave, and systolic ejection murmur of grade IV/VI intensity over the 2nd and 3rd left intercostal spaces. Oxygen saturation at rest was 84%-85% as determined by pulse oximetry. Two-dimensional color Doppler echocardiography established the diagnosis of bilateral superior vena cavae, complete atrioventricular septal defect, severe left atrioventricular valvar regurgitation, and severe right ventricular outflow tract obstruction with peak systolic gradient of 110 mmHg across the right ventricular outflow tract. The obstruction was at the level of pulmonary valve and right ventricular outflow tract.

The child underwent successful pulmonary valvotomy, resection of right ventricular outflow tract with reconstruction of complete atrioventricular septal defect by two-patch technique. The pulmonary valve ring was directly sutured.

Although our institution is a tertiary level center, the

socioeconomic profile of the patients and lack of health insurance benefit led to delayed referral and surgery.

Surgical Techniques

Patient position

The patient was placed in the supine position.

Intraoperative transesophageal echocardiography

Intraoperative transesophageal echocardiography was performed using a Philips echocardiographic machine to assess the morphology of the atrioventricular valves, size and location of the ventricular septal defects, severity of left atrioventricular valvar regurgitation, degree and level of right ventricular outflow tract obstruction, the sizes of both ventricles and ventricular function (Philips iE 33 echo machine; Philips X7-2T probe, Bothell, WA, USA).

The incision

Following median sternotomy, the thymus was subtotally excised taking care not to expose the brachiocephalic vein. The pericardium was incised on the left side using scissors. We did not use cautery to avoid inadvertent cautery-induced ventricular fibrillation. Three 4-0 silk stay sutures were placed on the incised pericardial edge thus creating a right sided pericardial flap.

Surgical planning

The operation was performed under moderately hypothermic cardiopulmonary bypass with an aortic infusion cannula, angled venous cannulae into both superior vena-cavae and a straight venous cannula into the inferior vena cava. St. Thomas (II) based cold hyperkalemic blood cardioplegia (1:4) and topical ice cooling was used for myocardial preservation.

The persistent ductus arteriosus was clipped using a Hemacclip (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA) after pulling down the superior surface of the pulmonary artery at the commencement of cardiopulmonary bypass as described by Dwight McGoon. The pump flow was temporarily lowered at the time of interruption of the ductus arteriosus.

Pulmonary arteriotomy, right atriotomy, insertion of a left ventricular vent

The main pulmonary artery was opened in between stay sutures prior to cross-clamping the aorta to avoid distortion of the main pulmonary artery. The right atrium was opened longitudinally 1 cm posterior to the right atrioventricular groove. Four stay sutures were placed on either side of the right atrial edges. The left ventricle was vented through the atrial septal defect using a DLP vent (Medtronic Inc., Minneapolis, MN).

Relief of right ventricular outflow tract obstruction

The pulmonary valve was tricuspid and stenosed. The stenosed pulmonary valve was incised at the level of pulmonary valve commissures. The obstructing muscle of the right ventricular outflow tract was excised (parietal insertion of the infundibular septum) via combined transpulmonary and transtricuspid approach. Adequacy of right ventricular outflow tract resection was checked by visual assessment and insertion of an appropriately sized Hegars dilator.

Assessment of the atrioventricular valves

The anatomical details of the malformation were examined. The atrial septal defect was of ostium primum type with the posteriorly positioned coronary sinus and margin of the atrioventricular septal defect delineating its boundaries.

The atrioventricular valve was distended by injecting cold saline into the ventricular cavities. This enabled us to delineate the optimal coaptation point of the left superior and inferior bridging leaflets and identify any surgical issues that would require attention during reconstruction. There was a prominent cleft between the left and right superior bridging leaflets of Rastelli's type A variety and a large unrestrictive ventricular septal defect with scooped out ventricular septum.

Dacron patch closure of the ventricular septal defect

The exposure of the ventricular septal defect was facilitated by use of two elastomer vascular loops. It is imperative to measure the length and height of the ventricular septal defect. The distance between the crest of the ventricular septum and the point of coaptation of the left superior and inferior bridging leaflets was considered as the height of the ventricular septal defect.

In order to minimize postoperative left atrioventricular valve regurgitation, it is important to maintain a smaller height of the ventricular septal patch to prevent/ minimize iatrogenic postoperative left atrioventricular valvar regurgitation, which is classically "D-shaped". A series of pledget supported interrupted 5-0 polypropylene sutures (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA) were placed 0.5 cm away from the ventricular septal crest.

At the superior and inferior corners of the ventricular septal defect, extreme precautions were taken to prevent residual ventricular septal defect. In both superior and inferior corners, the sutures were brought through the annulus and the substance of the superior and inferior bridging leaflet to prevent residual ventricular septal defect. The ventricular septal defect was closed using a "D-shaped" Dacron polyester patch (Bard® Savage® filamentous knitted polyester fabric, Bard Peripheral

Vascular Inc., Tempe, AZ, USA), slightly smaller than the size of the ventricular septal defect.

Reconstruction of the left atrioventricular valve

The septal commissure was reconstructed using Alain Carpentier's technique. A stay suture of 6-0 polypropylene was placed at the free leaflet margin opposing the atrial edge of the coaptation border. The septal commissural cleft was repaired using multiple interrupted, non-pledgeted 6-0 polypropylene sutures, taking precautions to take the bites through the atrial edge and not the ventricular edge, thus ensuring perfect competence¹⁵.

The newly constructed left atrioventricular valve opening was sized and matched with the indexed mitral valve orifice appropriate for the patient, thus avoiding iatrogenic mitral stenosis¹⁵. Cold saline was injected into the left ventricular cavity to ensure competence of the left atrioventricular valve and any leaking commissures, if present.

Pericardial patch closure of the atrial septal defect

A series of interrupted 5-0 polypropylene sutures were used between top of the ventricular septal patch and pericardial margin thus sandwiching the left atrioventricular valve. Extreme precautions were exercised at the superior and inferior corners of the defect, ensuring that the ventricular septal patch was brought into the corners of the superior and inferior bridging leaflets, thus preventing residual defects.

Subsequently, the pericardial patch was sutured to the edge of the ostium primum defect. In the area of the bundle of His, the sutures were passed through the superior edge of the coronary sinus, thus diverting the coronary sinus to the left side. Before completing the suture line, the left side of the heart was filled with cold

saline and gentle ventilation was resumed to evacuate residual air, if any.

Assessment of the right atrioventricular valve

Cold saline was injected into the right ventricle to ensure competence of the right atrioventricular valve. The right atrioventricular valve was competent. No additional reconstructive procedures were required.

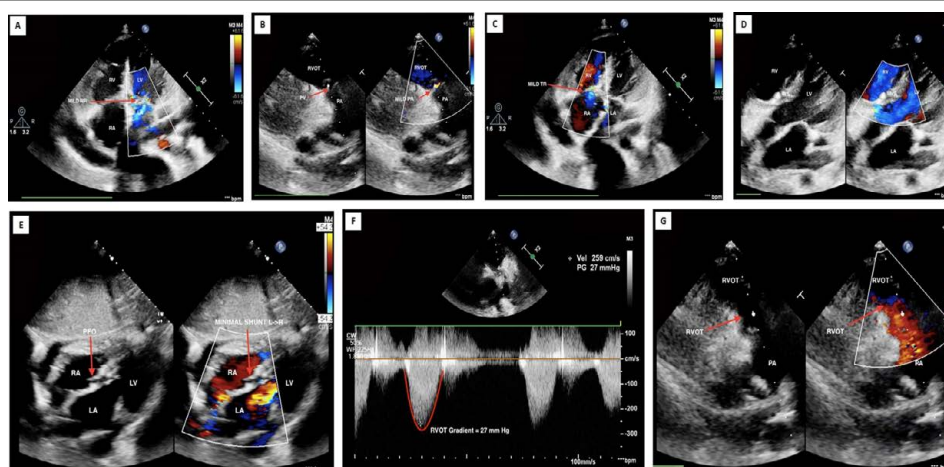
Closure of the right atriotomy and weaning off bypass

The right atrium is closed in two layers using 5-0 polypropylene suture. The pulmonary valve ring was closed directly in two layers using 5-0 polypropylene sutures. The patient was weaned off cardiopulmonary bypass with stable hemodynamics. The aortic cross clamp and cardiopulmonary bypass times were 62 minutes and 84 minutes respectively.

The postoperative peak systolic right ventricle-to-left ventricle pressure ratio was 0.5. Intraoperative transesophageal echocardiography did not demonstrate any residual ventricular septal defect. There was no left or right atrioventricular valvar regurgitation and no significant residual right ventricular outflow tract gradient.

Short- and Long-term Results

Postoperatively, the child was in normal sinus rhythm and recovery was uneventful. At 24th month follow-up the child was asymptomatic, no clinical evidence of cardiac failure, with Ross's clinical score of 2. Two-dimensional color flow Doppler echocardiography revealed normal biventricular function with minimal atrioventricular valvar regurgitation, intact atrial and ventricular septation with mild mitral and tricuspid regurgitation. The peak systolic right ventricular outflow tract gradient was 27 mmHg with minimal pulmonary regurgitation (Figures 1A-1G).



Figures 1A-1G: Series of post repair intraoperative transesophageal two-dimensional color flow Doppler echocardiography showing normal biventricular function with minimal atrioventricular valvar regurgitation, intact atrial and ventricular septation with mild mitral and tricuspid regurgitation. The peak systolic right ventricular outflow tract gradient was 27 mmHg with minimal pulmonary regurgitation

Study Limitations

Because of limited availability of slots for magnetic resonance scanning in the institute and the small age of the child which would have necessitated heavy sedation or general anaesthesia on account of longer scan duration in magnetic resonance imaging, magnetic resonance angiography could not have been performed.

Conclusions

Surgical repair of severe right ventricular outflow tract obstruction and left atrioventricular valve regurgitation should be undertaken in early childhood to prevent further anatomical deterioration/deformation of the left atrioventricular valve and to preserve ventricular function. Adequate resection of the right ventricular outflow tract without creating pulmonary regurgitation is of paramount importance to prevent right ventricular dysfunction, thus providing good long-term outcome.

Declaration of conflicting interests

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