**Introduction**

The atrioventricular septal defects encompass a spectrum of lesions in which the common etiology appears to be abnormal development of the superior and inferior endocardial cushions, resulting in a deficiency or absence of the the atrioventricular septum [1,2]. Data from the New England Regional Infant Cardiac Program and the Baltimore-Washington infant study defined a prevalence of 0.118 and 0.352 per 1000 live births [3,4]. Approximately, 50-75% of these patients have trisomy 21 [5]. When viewed from a reverse perspective, approximately 35-40% of patients with Down’s syndrome have an atrioventricular septal defect [5].

Atrioventricular septal defects include a spectrum of malformations. At one end of the spectrum is the partial atrioventricular septal defect characterized by an interatrial communication, but no interventricular communication and a connection of variable width between the left superior and left inferior leaflets [6]. At the other end is the most extreme form with large deficiencies in the atrial and ventricular septa, and a common atrioventricular valvular orifice, known as complete atrioventricular septal defect [1-5].
Several anatomical features are shared among all types of atrioventricular septal defects [1-6]. These include: a) absence of the usual wedged position of the aortic valve due to a common atrioventricular valve ring; b) lengthened outlet septum-to-ventricular apex ratio, resulting in an elongated left ventricular outflow tract and a “goose-neck” appearance; c) shortened dimension of the inlet septum-to-ventricular apex giving the interventricular septum a “scooped out” appearance; d) apical displacement of the attachments of the atrioventricular valves to the ventricular crest; e) inferior displacement of the atrioventricular node and coronary sinus; and f) variable degrees of underdevelopment of the inlet septum, resulting in absence of a ventricular septal defect, a restrictive ventricular septal defect, or a large ventricular septal defect [1-5].

In complete atrioventricular septal defect, there is a common atrioventricular valve that bridges both the left and right sides of the heart, creating superior and inferior bridging leaflets. These patients always have a “bare area” at the crest of the ventricular septum [1,2,7]. The Rastelli classification describes 3 types of complete atrioventricular septal defects based on the morphology of the superior bridging leaflet, its degree of bridging, and its chordal attachments. The classification does not relate to anatomy of the inferior bridging leaflet. In a Rastelli type A defect (69%), the common superior bridging leaflet is effectively split into two “halves” at the septum. Rastelli type B (9%) is rare and describes an anomalous papillary muscle attachment from the right side of the ventricular septum to the left side of the common superior bridging leaflet. In type C defects (22%) there is marked bridging of the ventricular septum by the superior bridging leaflet. The superior bridging leaflet is not divided but floats freely over the ventricular septum without chordal attachment to the crest of the ventricular septum [8].

There are a number of morphological factors that complicate the “usual” form of atrioventricular septal defect. These include: a) significant underdevelopment of either the right or left ventricle (prevalence of 10%) [9,10]; b) left ventricular outflow tract obstruction [11,12]; c) parachute deformity of the left atrioventricular valve [6,12]; d) double orifice left atrioventricular valve [13]; and e) double outlet left or right atrium [14].

There is definite association of complete atrioventricular septal defect and other conotruncal anomalies, particularly tetralogy of Fallot, double outlet right ventricle and transposition of the great arteries [1,2]. In patients with complete atrioventricular septal defect, the large left to right shunt causes right ventricle and pulmonary arterial pressures to be the same as the systemic pressure. These patients can have a relatively rapid progression of their pulmonary vascular disease [15,16]. Down syndrome appears to be associated with a rapid progression of pulmonary hypertension [15,16].

Patients with complete atrioventricular septal defect often present with severe congestive heart failure in the first 2 to 4 months of infancy and should be operated on between the ages of 3 and 6 months [17]. Presently, early primary repair is the procedure of choice in the majority of the centres. The principles of surgical management of complete atrioventricular septal defect include closure of the atrial septal defect, closure of the ventricular septal defect, creation of two non-stenotic competent atrioventricular valves and avoidance of damage to the atrioventricular node and bundle of His [7,17-19].

Repair of the complete atrioventricular septal defect has been described using 3 different techniques: a) the single-patch technique, as described in 1962 by Maloney and Gerbode [19,20]; b) the two-patch technique as described by George Trusler in 1975 [21]; c) the
modified single-patch technique of Graham Nunn in 1995 [22,23].

One of the keys to successful repair of atrioventricular septal defect using any technique is intraoperative assessment using transesophageal echocardiography and after cardiotomy. This includes an assessment of the right and left ventricular size, the size and shape of the atrial and ventricular septal defect, the number and location of the papillary muscles, the arrangement and attachment of the chordopapillary apparatus [17-22].

**Classic single-patch technique**

A pericardial, polytetrafluoroethylene or Dacron patch have been used in the single-patch technique. Use of pericardium as the ventricular septal defect patch is associated with the risk of development of aneurysm at the ventricular level [24]. The Dacron patch carries the risk of postoperative hemolysis [18-20].

**Two-patch technique**

A patch of Dacron or polytetrafluoroethylene is used to close the ventricular septal defect and a separate pericardial patch is used to close the atrial septal defect [21].

**Modified single-patch technique**

The ventricular septal defect is closed by placing a series of pledget supported 5-0 polypropylene sutures in the right-side of the interventricular septum. These sutures are then passed sequentially through the superior and inferior bridging leaflets at a site predetermined to separate the mitral and tricuspid valves. The sutures are then placed through the edge of the patch of autologous pericardium [22,23].

Literature documents almost similar results of all 3 surgical techniques. The single-patch technique has been extensively employed at Children’s Hospital, Boston [17], the University of California, Los Angeles [25] and Vanderbilt University [26]. The operative mortality was 3%, 7% and 15% and the reoperative rates for mitral regurgitation was 9%, 6% and 6% respectively among 3 institutions [17,25,26].

In a meta-analysis of results from five institutions among 794 patients undergoing operation by two-patch technique, the operative mortality was 7% and the incidence of reoperation for mitral regurgitation was 8% [18]. Among 72 consecutive repairs using modified single-patch technique, the operatively mortality was 2.8%, mild left atrioventricular valve regurgitation 29% and moderate regurgitation in 5% [22,23]. A trivial ventricular septal defect that did not require reoperation was noted in 20% of patients [23].

We report here-in the surgical repair of the complete atrioventricular septal defect using two-patch technique. A 26 months-old male child diagnosed with complete atrioventricular septal defect with moderate pulmonary arterial hypertension and severe left atrioventricular valvular regurgitation underwent successful reconstruction of the complete atrioventricular septal defect by two-patch technique.

**Surgical Techniques**

Following median sternotomy, the thymus is subtotally excised taking care not to expose the brachiocephalic vein. The pericardium is opened on the left side raising a right-sided flap in between stay sutures using scissors and not cautery to avoid inadvertent cautery-induced ventricular fibrillation.

The operation is performed under moderately hypothermic cardiopulmonary bypass with an aortic infusion cannula and angled venous cannulae into superior and inferior caval veins. St. Thomas based cold hyperkalemic blood cardioplegia (1:4) and topical ice cooling is used for myocardial preservation.
The persistent ductus arteriosus is ligated using No.2 ductus silk suture pulling down the superior surface of the pulmonary artery at the commencement of cardiopulmonary bypass as described by Dwight McGoon. The pump flow is temporarily lowered at the time of ligation of the ductus arteriosus.

Exposure is achieved through an oblique incision in right atrium 1 cm posterior to the interatrial groove. Four stay sutures are placed on the right atrial edges. The left ventricle is being vented through the atrial septal defect using a DLP vent (Medtronic Inc., Minneapolis, MN). The anatomic features of the malformation are examined. The coronary sinus and margin of the atrioventricular septal defect delineate the boundaries of the atrial level defect. There is a prominent cleft between the left and right superior bridging leaflets, consistent with Rastelli type A classification and a large ventricular septal defect with scooped out ventricular septum.

The atrioventricular valve is distended by injecting cold saline into the ventricular cavities. This enables us to delineate the optimal coaptation point of the left superior and inferior bridging leaflets and identify any abnormalities in valve coaptation that require attention during reconstruction. Two elastomer vascular loops are used to retract the left superior and inferior bridging leaflets. The length and height of the ventricular septal defect is measured. The height of the septal patch is determined between the crest of the ventricular septum and the optimal point of coaptation of the left superior and inferior bridging leaflets. The shape of the ventricular septal patch is classically “D-shaped” with its convexity along the edge of the ventricular septum. It is important to maintain a smaller height of the ventricular septal patch to prevent iatrogenic postoperative left atrioventricular valvular regurgitation.

Multiple pledget supported interrupted 5-0 polypropylene sutures (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA) are placed 0.5 cm away from the ventricular septal crest. Extreme care is taken at superior and inferior corners of the ventricular septal defect, where the septum meets the annulus to prevent residual ventricular septal defect. At these points, the sutures are brought through the annulus and the substance of the superior and inferior bridging leaflets to prevent residual ventricular septal defects in these locations.

The 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 is used to close the defect. For reconstruction of the leaking septal commissure, a stay suture of 6-0 polypropylene is placed at the free leaflet margin opposing the atrial edge of the coaptation border. Multiple interrupted, non-pledgeted 6-0 polypropylene sutures are used to repair the septal commissure taking precautions to take the bites through the atrial edge and not the ventricular edge, as recommended by Alain Carpentier, thus ensuring perfect competence.

Cold saline is injected again into the left ventricular cavity to check for a competent left atrioventricular valve and to identify any additional leaking commissures. Following repair of the left atrioventricular valve, we ensured that the left atrioventricular valve opening commensurate with the indexed mitral valve orifice, thus avoiding iatrogenic mitral stenosis. The left atrioventricular valve is then “sandwiched” between the top of the ventricular septal patch and a second patch of pericardium to be used to close the atrial level defect. Throughout the operation, interrupted mattress sutures are used. Precautions are taken at both superior and inferior portion of the defect to make certain that the ventricular septal patch is brought into the corner of the superior
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and inferior bridging leaflets and then along its border so as not have any residual defects.

The pericardial patch is sutured to the edge of the ostium primum defect with the exception of the area of bundle of His, where the sutures are displaced towards 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 is filled with iced saline and gentle ventilation to evacuate residual air, if any. Cold saline is injected into the right ventricle to ensure competence of the right atrioventricular valve. The right atrium is closed in two layers using 5-0 polypropylene suture. The patient is weaned off cardiopulmonary bypass with stable hemodynamics.

Short- and Long-term Results

The postoperative recovery was uneventful. At 26th month follow-up the child was asymptomatic, no clinical evidence of cardiac failure, with Ross’s clinical score of 2. Echocardiography revealed normal biventricular function without atrioventricular valvular regurgitation. There were no residual atrial or ventricular septal defects.

Conclusions

Surgical repair of complete atrioventricular septal defect should be done in early childhood. before the onset of severe pulmonary vascular obstructive disease and severe deformation of the atrioventricular valves. In cases of severe left atrioventricular valve regurgitation, the repair should be undertaken earlier to prevent further deterioration of valve function, left ventricular dilatation and function.

Declaration of conflicting interests

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