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Intracardiac Repair of Intermediate Atrioventricular Canal by Nunn's Technique: A Video Presentation

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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 atrioventricular septum [1-6]. This deficiency of the atrioventricular septum results in an ostium primum defect immediately above the atrioventricular valves and a scooped-out area in the inlet (basal) portion of the ventricular septum. 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 [7,8]. Approximately, 50-75% of these patients have trisomy 21 [7-9]. When viewed from a reverse perspective, approximately 35-

40% of patients with Down's syndrome have an atrioventricular septal defect [7-9].

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. 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 [1-6].

Transitional atrioventricular septal defect consist of an ostium primum atrial septal defect, a left-sided cleft atrioventricular valve, and a restrictive ventricular septal defect. The

ventricular septal defect is small, centrally located and partially closed by attachments of chordae from the superior and inferior bridging leaflets to the crest of the interventricular septum. It is caused by incomplete fusion of the points of coaptation between the left superior and left inferior leaflet. Additional small ventricular septal defects may be found under the superior and inferior leaflets by retraction of the right superior leaflets and probing with a small 1-2mm probe.

There are two atrioventricular valves forming two separate orifices. The ventricular septal defect in some cases is small and located underneath the superior and inferior bridging leaflets. In other cases, it may be larger, yet restrictive to flow with a gradient between left and right ventricle.

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 [1-6,10].

Abnormal differentiation and remodeling of the cushion mesenchyme into valvuloseptal tissue is the mechanism for development of atrioventricular septal defect [11]. The wide variability in the degree of development of the endocardial cushions explains the variability in size and extent of the septal defects and varying

grades of malformed atrioventricular valves [12].

Patients with transitional atrioventricular septal defects fall between partial and complete atrioventricular septal defects. The restrictive ventricular septal defect usually protects against the harmful sequelae of excessive pulmonary blood flow. The restrictive ventricular septal defect allows transitional atrioventricular septal defects to be repaired between 1-2 years of age [13]. If the restrictive ventricular septal defect is moderate in size, the repair should be done earlier to prevent progression of pulmonary vascular obstructive disease [14,15]. Moderate atrioventricular valvular regurgitation is present in 20-40% of patients with intermediate atrioventricular septal defects and severe regurgitation in about 15%. Atrioventricular valvular regurgitation may be more prevalent in older patients [1-6,16].

One of the keys to successful repair of atrioventricular septal defect by any technique is intraoperative assessment using transesophageal echocardiography and after cardiectomy. There are two techniques of closing a transitional atrioventricular septal defects: a) A pledgeted suture is taken through the crest of the interventricular septum and then through the left superior and left inferior leaflet, to close the ventricular septal defect. The mitral cleft is closed. Repair is completed by closure of the ostium primum atrial septal defect using a pericardial patch; b) Graham Nunn's modified single-patch technique [17,18]. This technique may be utilized, whereby the atrioventricular valve leaflets are brought to the crest of the interventricular septum to obliterate the restrictive interventricular communications [17,18]. The ostium primum atrial septal defect is closed using a pericardial patch.

We report here-in the surgical repair of the intermediate type of atrioventricular canal. A 2 month-old male child diagnosed with intermediate type of atrioventricular septal defect with mild pulmonary arterial

hypertension and severe left atrioventricular valvular regurgitation underwent successful reconstruction of the intermediate atrioventricular septal defect by Graham Nunn's technique.

Surgical Techniques

The operation

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 mild hypothermic cardiopulmonary bypass with an aortic infusion cannula and angled venous cannulae into the 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.

The left ventricle is being vented through the inter-atrial septum using a DLP vent through the patent foramen ovale. The analysis of the lesions is carried out. There are two atrioventricular valves with two separate orifices. There is an ostium primum atrial septal defect and the junction between the right and left atrioventricular valves is clearly delineated. As opposed to partial atrioventricular septal defect, the junction is not adherent to the muscular ventricular septum but connected to it by a fragile, flaccid membranous septum. There is a small perforated restrictive ventricular septal defect in the central portion as well as underneath the superior and inferior bridging

leaflets with multiple chordae being attached to the crest of the ventricular septum. The coronary sinus and margin of the atrioventricular septal defect delineate the boundaries of the atrial level defect.

The atrioventricular valve is distended by injecting cold saline with a bulb syringe into the left ventricular cavity so as to assess the morphology of the valves in the closed position and to assess the site(s) of leakage.

The repair is undertaken utilizing the principles of Graham Nunn's technique. Two elastomer vascular loops are used to retract the left superior and left inferior bridging leaflets. 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.

Multiple pledget supported 5-0 polypropylene suture (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA) are placed on the right side of the ventricular septum. The sutures are then passed through the substance of the superior and inferior bridging leaflets at the projected intercept of the ventricular septum and valve leaflets and a patch of pericardium to be used to close the atrial septal defect. Extreme precautions are taken at both superior and inferior portion of the defect to make certain that the ventricular septal defect is closed at

both the corners so as not to have any residual defect.

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.

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 transitional atrioventricular septal defect should be done in early childhood. 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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