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Atrial Correction (Modified Senning) of Transposition of the Great Arteries and Intact Atrial Septum with Regressed Left Ventricle and Pulmonary Hypertension: A Video Presentation

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ABSTRACT

A 3-year-old male child diagnosed as D-transposition of the great arteries and intact atrial septum with regressed left ventricle, Yacoub's Type-B coronary arterial pattern, successfully underwent modified Senning operation under moderately hypothermic cardiopulmonary bypass and St. Thomas based cold blood cardioplegia. At 8 months of follow-up, there was no mitral or tricuspid regurgitation with good biventricular function in Ross clinical score of 2.

Keywords: Transposition of great arteries, Atrial correction, Pulmonary hypertension, Regressed

INTRODUCTION

Transposition of the great arteries is the congenital malformation which results from the abnormal chamber connections of atrioventricular concordance and ventriculoarterial discordance systemic diastolic dysfunction (SDD). It is generally classified as a type of conotruncal abnormalities, a group of abnormalities that has a common theme of deranged development of the cardiac outflow tract. In D-transposition, the aorta is anterior and to the right of the pulmonary artery. This pattern results in the systemic and pulmonary circulations occurring in parallel rather than in series.[1,2]

These patients are subdivided into those with intact ventricular septum (50%), ventricular septal defect (25%), and ventricular septal defect with pulmonary stenosis (25%).[1,2] Other associated cardiac anomalies include persistent ductus arteriosus, coarctation of aorta and transposition of the great arteries.[3,4] Transposition of the great arteries accounts for 9.9% of infants with congenital heart disease or 0.2/1000 live births.[5]

Many classification system have been used to describe the coronary anatomy in transposition of the great arteries.[4-6] The Leiden classification is most commonly used.[6-8] The most common coronary pattern in D-transposition of the great arteries (68%) consists of the left main coronary artery arising from the leftward coronary sinus giving origin to left anterior interventricular and circumflex coronary arteries. The right coronary artery arises from the right posterior sinus. In 20% of cases, the circumflex coronary arteries arise from the right coronary artery and passes behind the pulmonary artery. In 4.5% of cases, a single right coronary artery arises from the right posterior sinus or a single left coronary artery from the leftward coronary sinus.[9] Intramural coronary arteries that proceed in the aortic wall for a distance before exiting to the epicardial surface, single coronary ostium, or separate ostia have been described.[10] Inverted origin of the coronary arteries and inverted origin of the circumflex and right coronary arteries have been described in 3% and 7% of cases, respectively.[11]

Although anatomical repair of transposition of great arteries is the treatment of choice, total abandonment of the atrial baffle procedures has not taken place because of the technical problems and "learning curve" of the former, select clinical applications and good short-term results of the later.[11-14]

In the current era, the indications of the atrial switch operation have been limited to delayed/late presenters of D-transposition of the great arteries, transposition of the great arteries with complex coronary anatomy precluding an arterial switch operation, those who require atrial switch as a part of the anatomic repair of congenitally corrected transposition of the great arteries, those with isolated ventricular inversion, isolated atrioventricular discordance, D-transposition of the great arteries with the left ventricular outflow tract obstruction not amenable to a conventional Rastelli or Nikaidoh procedure and D- transposition of the great arteries with intact atrial septum, and severe pulmonary hypertension without a correctable cause.[12-20]

Without surgery, the natural history of patients with all variants of transposition of great arteries was dismal, with 55%, 85%, and 90% mortality rates at 1 month, 6 months, and 1 year, respectively.[11] After emerging as the only viable solution for patients with TGA, the Senning operation successfully enjoyed popularity, followed by abandonment in favor of the Mustard operation in the mid-1960s and early 70s, then an initial revival after modification introduced by Quaegebeur *et al*., before finally finding the current indications as enunciated above.[12-20]

At present, surgeons can perform the Senning operation with low mortality and minimal morbidity by applying several technical modifications and paying meticulous attention to large and unobstructed venous pathways.[11-20]

Following Senning's operation, symptomatic caval obstruction is relatively rare, generally observed within weeks to several months after Senning's operation and rarely beyond 1 year after surgery postoperatively.[21] Superior vena caval obstruction is observed more frequently than obstruction of the inferior vena cava.[22] Systemic venous obstruction has been reported more frequently after the Mustard operation (10–40%) than after the Senning operation.[22-24]

Pulmonary venous obstruction contrary to systemic-venous obstruction is usually symptomatic.[25] The reported incidence of this complication is 0–27%, much less frequent after the Senning operation.[21,24] Baffle leaks lead to either bidirectional or predominantly right-to-left shunt. Right-to-left shunting in the absence of elevated systemic venous pressures is due to streaming of blood underneath the interatrial baffle with an incidence ranging from 20% to 73% after the Mustard operation and from 0% to 50% after the Senning operation.[21]

Supraventricular dysrhythmias are frequent after an atrial switch operation. Sinus node dysfunction is present in 30% of operative survivors, more frequently in patients aged younger than 5 months at the time of surgery.[26]

Deanfield *et al*. reported normal sinus rhythm in 84% of their patients in the immediate post-operative period, falling to 56% in sinus rhythm after a Senning correction, and to 66% after a Mustard operation at a mean follow-up of 7 years.[27]

Intra-atrial re-entry tachycardia occurs in 2–10% of patients after an atrial switch operation and may be responsible for 3–15% incidence of post-operative sudden death.[28] Atrial tachyarrhythmias are induced by re-entrant circuits from extensive atrial suture lines involved in a Senning or Mustard operation.[28]

The incidence of post-operative tricuspid regurgitation following an atrial switch reaches as high as 52% and is more frequent in patients with transposition of the great arteries and ventricular septal defect.[25,28-30]

Eventual right ventricular failure is not a time-related event.[26] There is evidence in the published literature that onset or degree of right ventricular failure or exercise tolerance may be reduced when the Senning operation is performed earlier, particularly before the age of 1 year.[31]

The operative mortality of the Senning correction in the published literature ranges from 0% to 10%.[14,22,29-39] The incidence of late mortality ranges from 0% to 16.1% in recent series, most often in the form of sudden death.[14,22,29-39]

At 25-years follow-up, the Senning operation had survival advantage as compared to Mustard operation (late cardiac mortality 2% at 10 years and 15% at 15 years, respectively).[40]

Moons and associates in a multicenter study from Belgium compared the long-term outcome in 339 patients up to 30 years after Senning and Mustard operations.[41] Both groups had a relatively high mortality rate, but actuarial survival at 10, 20, and 30 years was satisfactory at 91.7%, 88.6%, and 79.3%, respectively. The survival was slightly better for the Senning group.[29-40]

We present here-in a 3-years old male child diagnosed as D-transposition of the great arteries and intact atrial septum with regressed left ventricle, Yacoub's Type-B coronary arterial pattern, undergoing modified Senning operation under moderately hypothermic cardiopulmonary bypass, and St. Thomas based cold blood cardioplegia. Post-operative recovery was uneventful.

SURGICAL TECHNIQUES

The operation

Survival planning and the position

Following median sternotomy, the thymus was subtotally excised taking care not to expose the brachiocephalic vein. The pericardium was incised 1 cm in front of and parallel to the phrenic nerve, thus exposing the two great arteries, the right atrium and the superior vena cava. The pericardium was opened slightly on the left side using scissors and not cautery to avoid inadvertent cautery-induced ventricular fibrillation. During dissection, extreme precautions were taken not to injure the pericardium with cautery or scissors for possible later usage as an *in situ* pericardial flap for pulmonary venous pathway. The extrapericardial segments of the superior and inferior caval veins were dissected for the purpose of venous cannulation.

Extracorporeal circulation

The operation was performed with moderately hypothermic cardiopulmonary bypass at 32°C. Angled venous cannulas were inserted into the superior and inferior caval veins and with distal aortic cannulation. Measures were taken to avoid excessive manipulation.

Dissection and ligation of the ductus arteriosus

The persistent ductus arteriosus was dissected on the superior surface of the pulmonary artery by McGoon's technique and was doubly ligated using No.2 silk suture (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA).

Cross-clamping of the ascending aorta and administration of cardioplegia

The aorta was cross-clamped. Antegrade St. Thomas based (1:4) cold blood cardioplegia and topical cooling was used for myocardial preservation.

Incision in the right and left atrium

Access to the inside of right atrium was gained through a longitudinal incision 3–4 mm anterior and parallel to the sulcus terminalis. Precautions were taken to ensure placement of the incision well away from the sinoatrial node. The cranial extent of the incision was limited to 0.5 cm from the superior margin of the right atrium.

Only after inspecting inside of the right atrium, the incision was extended caudally so that it could be directed toward the lateral insertion of the Estachian valve. The pericardial reflection of the junction of inferior vena cava and right atrium was left intact for later suturing.

Formation of left atrial outlet and insertion of left atrial vent

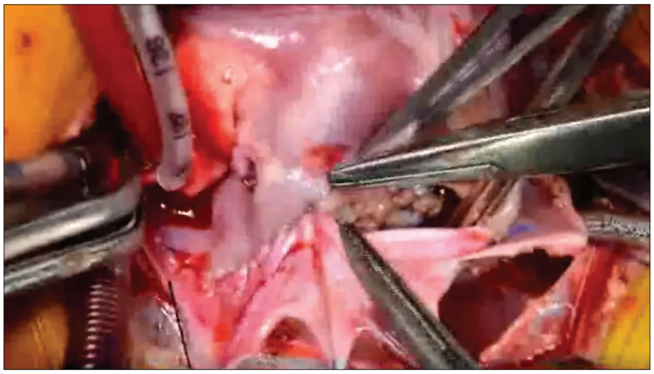
The interatrial groove was dissected in front of the right pulmonary veins. The left atrium was opened by a craniocaudal incision posterior to the interatrial groove, ensuring an adequate, non-restrictive future left atrial outlet. The left atrium was vented through the left atriotomy using a No. 13 DLP suction vent (Medtronic Inc., Medtronic Parkway N.E., Minneapolis, MN, USA).

Mobilization of the interatrial septum and creation of a interatrial septal flap

After the right atrium was opened in between stay sutures, the interatrial septum was carefully inspected. An incision was made in the most anterior part of the foramen ovale and extended cranially for a distance of approximately 7 mm. The direction of the incision was then changed dorsally toward the superior margin of the right superior pulmonary vein and extended to the base of the interatrial septum. Similarly, an incision from the lower part of fossa ovalis was continued downward toward the inferior margin of the right inferior pulmonary vein. While creating the atrial septal flap, precautions were taken not to deviate the incision anteriorly toward the muscular aortic mound and not to injure the vascular supply to the sinoatrial node. The raw margins of the septum were then endothelialized with interrupted sutures of 5-0 polypropylene taking superficial bites and approximating the endocardium. The atrial septal flap was now connected only at the base, which corresponds outside the atrium to the interatrial groove.

Assessment of the adequacy of flap size

The distance between the tip of the trapezoid septal flap and left atrial wall between the left atrial appendage and pulmonary veins was measured. An appropriate sized expanded polytetrafluoroethylene patch (Impra e PTFE Cardiovascular Patch, M/s Bard Peripheral Vascular Inc., West 3rd Street, Tempe, Arizona) was used to augment the native interatrial septal flap to avoid excessive traction, and flattening of the neosystemic venous pathway.



Video 1: Ujjwal-Senning (Video).

Creation of the posterior wall of the neosystemic venous pathway

The trapezoidal interatrial flap was sutured to the left atrial wall between the base of the left atrial appendage and the pulmonary veins using 5-0 polypropylene suture. The suture line was continued both superiorly and inferiorly along the posterior wall of the left atrium to the base of the flap. During suturing of the flap, we ensured that the flap was without any tension and not causing pulmonary venous obstruction. The suture line was checked for leaks with a nerve hook to prevent any post-operative shunting.

Sewing the anterior edge of the posterior segment of the right atrium

The anterior edge of the posterior segment of the right atrium was sutured to the anterior part of the septal defect between the mitral and tricuspid valves. Suturing was continued superiorly and inferiorly around the lateral margins of the orifices of the superior and inferior venae cavae. A 24-Fr venous cannula was inserted through the wound to ensure proper sizing of the systemic venous pathway. The atrial wall flap was then sutured into the place using the cannula as a stent. The suture line, when continued down toward the inferior caval vein passed anterior to the coronary sinus to avoid injury to the atrioventricular node [Video 1].

Creation of the neopulmonary venous pathway

Few interrupted 5-0 polypropylene sutures were used to oppose the lateral edge of the wall of the left atrium and adjacent pericardium taking care not to make any stitch holes. The posterior edge of the anterior segment of the right atrial wall was sewn to the *in situ* pericardium on the right side and to the right atrial wall around the caval tunnel. To prevent injury to the sinoatrial node, suturing was done about 1.0 cm above the superior cavoatrial junction. The caval snares were loosened so that both venae cavae were filled, fully distended, and stressed thus avoiding caval constriction.

The child was weaned off cardiopulmonary bypass with stable hemodynamics, in sinus rhythm, on dobutamine and nitroglycerine infusion. At 8 months of follow-up, there was no mitral or tricuspid regurgitation with good biventricular function (left ventricular ejection fraction 0.60), in Ross clinical score of 2.

CONCLUSION

The modified Senning operation currently is the treatment of choice in late presenters of transposition of great arteries with regressed left ventricle and/or pulmonary hypertension. The advantages include avoidance of use of large akinetic pericardial patch which is prone for shrinkage, use of maximal viable native tissue in the neosystemic and pulmonary venous pathway having growth potential, potential functional capacity with muscular contraction of the atrial chambers, and avoidance of akinetic patches that can scar, shrink, thicken, and further obstruct inflow, such as that seen with Mustard operation. Meticulous attention should be exercised at every step of the operation to avoid sinoatrial nodal injury, baffle leaks, and obstruction of the systemic and pulmonary venous pathway. An *in situ* pericardial patch for the neopulmonary venous pathway as was originally described by John Waldhausen should avoid any future pulmonary venous obstruction.[42]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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