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Anatomical Correction of Transposition of the Great Arteries at the Arterial Level with Dacron Patch Closure of Multiple Ventricular Septal Defects under Integrated Extracorporeal Membrane Oxygenation: A Video Presentation

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ABSTRACT

A 5-week-old male child, weighing 4 kg diagnosed with d-transposition of the great arteries with multiple muscular ventricular septal defects, Yacoub's type-A coronary arterial pattern, successfully underwent arterial switch operation with Dacron patch closure of ventricular septal defects under moderately hypothermic cardiopulmonary bypass and St. Thomas based cold blood cardioplegia under integrated extracorporeal membrane oxygenation. Postoperatively, he required mechanical circulatory assistance for 72 h. At 12 months of follow-up, there was no mitral or tricuspid regurgitation, no neoaortic valve insufficiency with good biventricular function in Ross clinical score of 2.

Keywords: Transposition of the great arteries, Regressed left ventricle, Ventricular septal defect

INTRODUCTION

Transposition of the great arteries is a congenital cardiac malformation that results from the abnormal chamber connections of atrioventricular concordance and ventriculoarterial discordance (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]

Transposition of the great arteries accounts for 9.9% of infants with congenital heart disease or 0.206/1000 live births.[3] 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 and coarctation of the aorta.[4,5]

The ventricular septal defects are classified based on the surgical anatomy of the interventricular septum and the morphologic definitions as published by Soto *et al*.[6] In patients with transposition of great arteries, because of the surgical difficulties that are encountered for ventricular septal defects located at the trabecular septum, Serraf *et al*. divided the trabecular septum into three components: high (an anterior), mid (centered by the moderator band), and low (or apical).[7]

Thus, the interventricular septum is composed of six components: Perimembranous, inlet muscular, outlet muscular, and the three trabecular muscular components.[8,9] The definition of multiple ventricular septal defects is based on the involvement of at least two out of six components of the interventricular septum.[8,9]

In 1999, Seddio *et al*. defined "Swiss-Cheese" septum as a localized or general lack of compaction of the interventricular septum.[10] In 1995, Houyel *et al*. described the relationship of the great arteries in cases of transposition of great arteries with ventricular septal defect.[11] In 76% of cases, the aorta and pulmonary artery were in an anteroposterior relationship. A side-by-side relationship was observed in 20% and l-malposition of the aorta was present in 4% of cases.[12]

Coronary arterial anomalies are more frequent in the transposition of great arteries in ventricular septal defects. Many classification systems have been used to describe the coronary anatomy in transposition of the great arteries.[5,12-14] The Leiden classification is most commonly used.[12-16] 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 pass 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.[17] 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.[16] The 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.[18]

Despite advances in pre-operative diagnosis, improved surgical techniques, and perioperative care, significant mortality still exists in patients undergoing arterial switch operations with multiple ventricular septal defect closure.[19-21]

Although the spectrum of ventricular septal defect in the transposition of great arteries is different from that of hearts with ventriculoarterial concordance, the principles of surgical repair are nearly similar.[7-11,20,22,23]

There remains considerable controversy regarding the optimal management of these patients with transposition of great arteries and multiple ventricular septal defects.[7-11,20,22,23] Although the present practice is to perform early anatomical repair, the ideal time of surgical intervention, the ideal surgical approach, and one versus two-stage management remains controversial. At present, in patients with transposition of great arteries and multiple ventricular septal defects, the presence of associated aortic arch obstruction without severe hypoplasia, and "Swiss Cheese" defects, a preliminary pulmonary artery banding may be the preferred option.[7,20-24] The presence of aortic and/or subaortic stenosis, coronary artery coursing between the aorta and pulmonary artery, and closing ductus with a restricted atrial septal defect are contraindications for pulmonary artery banding as initial palliation.

In 1999, Conte *et al*. reported internal pulmonary arterial band placement to reduce the risk of pulmonary arterial distortion and coronary compression.[25] An accurate pre-operative diagnosis is essential for a satisfactory outcome.[7-10,20,22-24]

Although the atrial baffle procedures are associated with a low mortality rate (<5%), they are plagued by long-term complications, namely, baffle leak, supraventricular and ventricular dysrhythmias, systemic venous pathway obstruction, tricuspid valve insufficiency, and right ventricular (RV) failure.[26-28] The arterial switch operation has largely replaced atrial baffle procedures with excellent mid-term and long-term results.[26-28] At present, the indications of atrial baffle procedures are limited and are institutionally dependent.[26-28]

The anatomical correction at the arterial level is the procedure of choice. Improved techniques of coronary transfer, myocardial protection, and neogreat vessel reconstruction have resulted in improved survival statistics that compare favorably with the atrial baffle procedures.[29-31] Anomalies of the coronary arterial origin and course in transposition of great arteries are common and various innovative techniques have been employed for coronary artery transfer during arterial switch operation.[29-31]

Numerous attempts have been made to extend the boundaries of the atrial switch operation in children presenting late with intact ventricular septum or restrictive ventricular septal effects and regressed left ventricle. Literature is divided on the usefulness of elective institution of mechanical circulatory support, such as extracorporeal membrane oxygenation (ECMO) or left ventricular assist device (LVAD) for late presenters of d-transposition.[32-37] Published data support that a primary atrial switch operation can safely be performed as a primary procedure up to 2 months of age, and above this age, left ventricular (LV) mass and LV mass to LV end-diastolic volume ratio may orient toward rapid two-stage or primary atrial switch operation up to 2 years

of age. Beyond 2 years, an atrial switch procedure should be considered.[33-37]

The decision to perform a primary atrial switch operation in our center is based on the values of LV mass >35 g/m^2 and when between 25 and 35 g/m^2 on the value of LV mass/LV end-diastolic volume ratio >1.2, LV posterior wall thickness >4 mm in diastole and absence of "D" or "banana" shaped LV cavity.[34-37]

However, the above-mentioned echocardiographic criteria to calculate LV mass assume the LV to be either spherical or prolate ellipsoid in shape.[38] However, in transposition, the regressed left ventricle is either "banana" or "D" shaped. Therefore, the criteria of LV mass index <35 g/m^2 should be used with caution in labeling the regressed left ventricle. The other indications used in our institutions, as well as by others for retraining the regressed left ventricle have been an LV/RV pressure ratio of <0.6 for children more than 3 months or <0.5 in those <3 months.[39,40]

We have incorporated ECMO in the cardiopulmonary bypass (CPB) circuit as an integrated ECMO – CPB circuit to reduce the cost, minimize the time lag, initiate ECMO to enable LV retraining under normoxemia and controlled loading, avoid low cardiac output state and surgical asepsis.

Several investigators in recent times have used LVAD in the postoperative period after delayed ASO.[41-43] T-stage ASO although being practiced in several institutions is fraught with its own set of complications with higher interstage mortality and morbidity from neoaortic regurgitation and LV diastolic dysfunction due to subendocardial ischemia.[44]

The arterial switch operation restores LV to aortic continuity and has not been associated with atrial baffle-related complications as enumerated above. The mid-and long-term results of patients undergoing arterial switch operation are encouraging. In the congenital heart surgeon study on 513 infants (transposition of great arteries with intact ventricular septum, n = 385; transposition of great arteries with ventricular septal defect, n = 129), the 1-month, 1-year, and 5-year survival rates were 84%, 82%, and 82%, respectively.[45,46] The largest series on 1095 patients from a single institution with long-term follow-up was reported in 2001. At a mean follow-up of 5 years, hospital mortality was 8.6%, survival was 89% at 1 year, and 88% at 10 and 15 years.[45]

SURGICAL TECHNIQUES

Surgical planning and the position

Following median sternotomy, the thymus was subtotally excised taking care not to expose the brachiocephalic vein. The pericardium was incised about 5–6 mm in front of and parallel to the phrenic nerve, thus exposing the two great arteries, the right atrium, and the superior caval vein. The pericardium was opened using scissors and not cautery to avoid inadvertent cautery-induced ventricular fibrillation. A rectangular segment of pericardium was harvested and fixed in 10% glutaraldehyde for 10 min for later use of RV outflow tract reconstruction.

The position and size of the great arteries, as well as the site of origin and distribution of the coronary arteries, were determined. All four chambers were inspected and pressures were recorded. Note the anteroposterior disposition of the aorta and the main pulmonary artery with the left main coronary artery arising from the left posterior aortic sinus and the right coronary artery arising from the right posterior aortic sinus. The left main coronary artery gives origin both to the left anterior coronary artery and circumflex coronary artery.

The operation was performed with moderately hypothermic CPB 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 division of the ductus arteriosus

The persistent ductus arteriosus was dissected on the superior surface of the pulmonary artery by McGoon's technique, transfixed at both aortic and pulmonary arterial ends using 6-0 polypropylene suture (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA) and was divided.

Mobilization of the aorta and the pulmonary arteries

Both great arteries were separated from each other by dividing the pericardial reflection between them, starting from the level of the top of the commissures and pulmonary artery till the level of pulmonary arterial branching on either side. Low voltage cautery (10–15 mv) was used during dissection. The aorta, right, and left pulmonary arteries were looped using elastomer vessel loops.

Placement of marking sutures at the proposed neoaortic location of coronary transfer

Two marking sutures were placed over the pulmonary artery at the proposed neoaortic location. Note the site of neoaortic coronary button, a little above the sinuses to avoid waisting/kinking of the coronaries.

Cross-clamping of the ascending aorta and administration of cardioplegia

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

Venting of the left ventricle

Right atriotomy was done after snugging the inferior caval vein and the left heart was vented through the atrial septal defect using a No.13 DLP suction vent (Medtronic Inc., Medtronic Parkway N.E., Minneapolis, MN, USA).

Dacron patch closure of the ventricular septal defects

There were two ventricular septal defects: One measuring 2 × 1.5 cm was located in the apical trabecular septum and the second ventricular septal defect was located in the mid-muscular septum above the trabecula septomarginalis. The apical ventricular septal defect was closed using an appropriately sized Dacron polyester patch (Bard® Savage® filamentous knitted polyester fabric, Bard Peripheral Vascular Inc., Tempe, AZ, USA) and 10 pledgeted 5-0 polypropylene sutures (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA). The other ventricular septal defect was closed using 7 pledgeted 5-0 polypropylene sutures. The tricuspid valve was checked for competence in injecting cold saline through the tricuspid valve into the right ventricle.

Administration of second dose of cardioplegia

The second dose of antegrade root cardioplegia was administered after closing the ventricular septal defect.

Dissection of the aorta and pulmonary trunk

The aorta was divided between stay sutures at the level of pulmonary arterial bifurcation. Two stay sutures of 6-0 polypropylene were placed on the top of each coronary button for retraction and mobilization. The transected distal aortic end was retracted superiorly for optimal visualization and uncluttering of the surgical field.

The pulmonary trunk was next divided about 1–2 mm below the level of pulmonary arterial bifurcation. The vessel loops on the right and left pulmonary arteries were placed on traction for the LeCompte maneuver.

Inspection of the pulmonary valve

The pulmonary valve and subpulmonary region were inspected through the transected end of the pulmonary artery.

Identification of the coronary artery

The position of the coronary ostia and their relation to the sinuses of the valsalva of the aortic and pulmonary valves was determined. Note the origin of both coronary arteries from the left and right posterior sinuses, respectively. In addition, the course and mode of branching of the proximal 5–6 mm of each artery were inspected.

Mobilization of coronary buttons

The left and right coronary ostia were mobilized with a surrounding rim of the aortic wall including almost the full-thickness wall of the sinus of valsalva without causing injury to the valve leaflets. The process was started at the edge of the transected aortic wall for the left coronary ostium followed by the right coronary ostium. Low voltage cautery (5–6 mv) was used for hemostasis at the harvested site.

Creation of medial-based pulmonary arterial flaps for coronary artery translocation

A site on the pulmonary artery for the left and right coronary anastomoses was chosen. This neoaortic location was higher than the top of the sinus of valsalva, to avoid tension/kinking/waisting of the coronary arteries and to avoid distortion of the aortic valve. Two medially based pulmonary arterial flaps as described by Roger Mee were created accordingly.[15,46] The sites chosen allowed for rotation of the mobilized coronary buttons through an angle not more than 30°. No portion of the pulmonary arterial wall was excised.

Coronary anastomoses

The coronary anastomoses were performed using a 6-0 polypropylene suture, taking care not to injure the coronary ostia and distortion of the mobilized disc.

The LeCompte maneuver

The pulmonary arterial bifurcation, right, and left pulmonary arteries were mobilized beyond the site of the divided ductus into the pulmonary hilum on the left side and behind the superior caval vein to the branching of the right pulmonary artery on the right side. The distal transected end of the aorta was then threaded behind the mobilized pulmonary arterial bifurcation by the LeCompte maneuver and the second aortic controlling clamp was transferred onto the aorta at its new site behind the pulmonary artery bifurcation.

Reconstruction of the aorta

The small distal end of the aorta was matched to the larger proximal end for anastomosis using a 6-0 polypropylene suture.

Repair of the defects in the aortic sinus

Two defects that were produced in the facing sinuses of the pulmonary trunk were repaired using a single patch of the autologous pericardium and a 6-0 polypropylene suture. The size of the patch is approximately one and a half times the size of the defect, thus enlarging the diameter of

the proximal aorta to match the size of the distal pulmonary artery. At this stage, we used topical thrombin (Tisseel, Baxter AG, Vienna, Austria) as an additional topical hemostatic agent.

Release of the aortic cross-clamp and restoration of myocardial perfusion

The aortic cross-clamp was released, thus restoring myocardial perfusion. Note the distended left and right coronary arteries perfusing the myocardium without any kinking or torsion.

Reconstruction of the pulmonary artery

The last stage of the LeCompte maneuver consists of direct anastomosis between the reconstructed proximal aorta and the distal pulmonary artery using a 6-0 polypropylene suture. Precautions were taken to anastomose the two ends without compressing the proximal parts of the coronary arteries, without causing narrowing of the right and left pulmonary ostia, and without stretching the branches of both pulmonary arteries.

Closure of the right atriotomy

The atrial septal defect was directly closed using a 5-0 polypropylene suture. The right atrium was closed in two layers using a 5-0 polypropylene suture.

RESULTS

The child was electively placed on integrated ECMO and dopamine 5 μg/kg/min, dobutamine 5 μg/kg/min, adrenaline 0.01 μg/kg/min, and nitroglycerine 0.5 μg/kg/min for 72 h. Subsequently, the child was weaned off ECMO support and discharged home on the 20th post-operative day. Postoperatively, the child is asymptomatic, on nil medications, and not in cardiac failure, with Ross clinical score of 2. Echocardiographically, the child has normal biventricular function without any residual anatomical lesions.

CONCLUSION

Anatomical correction of transposition with concomitant closure of multiple ventricular septal defects as a one-stage procedure with integrated ECMO is an expedient, safe, and effective surgical strategy, for late presenters beyond 4 weeks of age. The large left-to-right shunt and pulmonary hypertension resulting from the multiple ventricular septal defects (and from the patent arterial duct) may obscure the presence of magnitude of the smaller ones, especially located in the high and mid-trabecular septum. Thus, an accurate pre-operative diagnosis and intraoperative systemic examination are crucial for a successful outcome.

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.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of Artificial Intelligence (AI)-Assisted Technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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