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Surgical Challenges of Cardiac transplantation in Dextrocardia with Congenital Heart Disease A Systemic Review

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Abstract

Pediatric cardiac transplant evolved in last decade. In preoperative investigation CT volumetry should be done to prevent donor recipient mismatch. In dextrocardia it is difficult to get same morphological donor so in case of dextrocardia donor heart should be tilted to right side and pericardium should be wide opened after opening of bilateral pleura. In case of left superior vena cava and situs inversus tube graft is used for vena caval connection. In post Glenn and Fontan patient extra length of donor pulmonary artery should be taken and in hypoplastic left heart syndrome donor aorta till isthmus should be taken. We have described different surgical strategies of 7 case reports and 1 case series of heart transplant with dextrocardia.

Keywords: Pediatric; Dextrocardia; Cardiac surgery

Introduction

Dextrocardia is a rare condition and only one percent of the population is affected. It is most often diagnosed incidentally. Dextrocardia is commonly associated with other congenital heart disease. Any kind of cardiac surgery is more challenging with dextrocardia [1]. Heart transplantation in a congenital heart disease itself is a technically challenging procedure which needs a team approach of surgeons, anaesthesiologists, nurses, cardiologists and intensivists and requires an appropriate preoperative planning. Appropriate planning and judicious understanding of the recipient anatomy is needed for procurement of donor organ. Most of the recipients have one or multiple previous open heart operations. In patients of Pulmonary and systemic venous anomalies, pulmonary branch defect, aortic arch anomalies, anomalies of the systemic venous return and heterotaxy syndrome establishing cardiopulmonary bypass is difficult and donor procurement technique should be planned accordingly. Incidence of systemic venous return abnormalities is 0.1 to 3 percent in the patients of congenital heart disease. Over the last several years incidence and prevalence of adult congenital heart disease has increased due to improved survival through childhood. 10-20% of adult congenital heart patients need heart transplantation in their life [2]. Very few case reports of dextrocardia with heart transplantation was reported heart transplantation in patients with dextrocardia. Finding a dextrocardia donor heart is also difficult. Placement of heart in right pleural cavity is barely reported [3].

Material and Methods

We have searched in Pub med with the key ward cardiac transplantation in dextrocardia, heart transplantation in dextrocardia. We have got 51 results. On the basis of Title and abstract we have selected 9 articles. After that on the basis of full text we have selected 8 articles. One article was excluded because operative technique was not mentioned. Our inclusion criteria were case report, case series, original article or review article on cardiac transplantation with dextrocardia. We have selected 7 case reports and 1 case series [4-10] (Table 1).

Discussion

Heart transplantation (HTx) is standard procedure for selected patients with end stage heart failure. Improvements in procurement of donor, surgical techniques, numerous immunosuppressive drugs and appropriate intensive care management have resulted in a substantial decrease in acute allograft rejection, which had previously significantly limited survival of HTx recipients [11].

First cardiac transplantation was performed by Christian Barnard from South Africa in December 1967. In the United States first cardiac transplant was performed by Dr. Shumway and colleagues at Stanford University in January 1968. However, initial outcomes of cardiac transplantation was not good because of the complex postoperative problems like graft rejection and infection. That's why total number of cases reduced after the initial phase. Number of transplant in every year was increased after discovery of cyclosporine in 1980. Currently, the number of cardiac transplant performed worldwide is estimated to be nearly 5,500 procedures annually [12]. Dextrocardia was first described by anatomist-surgeon Hieronymus Fabricus in 1606, and an account of situs inversus was given by Marco Aurelio Severinus in 1643. In Dextrocardia cardiac mass is displaced to the right of midline and the apex may or may not be on the same side. In adults, dextrocardia is usually associated with atrial situs inversus [13].

As it is very difficult to get a donor with dextrocardia most of the time levocardia heart is transplanted. During implantation excision of left sided pericardium is done for room of the heart and sometime lobectomy is also done. During pericardial excision care should be taken to preserve phrenic nerve. Another strategy is to tilt the apex 45 to 90 degree to the right side. In this perspective longer donor PA and aorta should be harvested for tension free anastomosis [6]. Dextrocardia is

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Table 1: List of Articles published on cardiac transplantation in dextrocardia and heart transplantation in dextrocardia.

Author	Year	Number	Diagnosis	Surgical Management
Rajab TK et al. [4]	2019	1	Dextrocardia, ASD, and Eisenmenger syndrome with V-V ECMO	Heart-lung transplantation.
Pradegan N et al. [3]	2019	1	DILV with right ventricular accessory outflow chamber, double-outlet from the outflow chamber, and pulmonary stenosis, LSVC present, Post BT shunt	Orthotropic heart transplantation
Boston U et al. [5]	2019	2	a. Visceral situs inversus, heterotaxy, dextrocardia, complete AVSD, DORV, severe pulmonary stenosis, bilateral superior vena cava (SVC) without a bridging innominate vein, interrupted IVC with azygous continuation to the left SVC, ipsilateral pulmonary veins, and congenital heart block, post B T shunt b. Visceral situs inversus, heterotaxy, dextrocardia, interrupted aortic arch, unbalanced complete AVSD with severe regurgitation, DORV, and bilateral SVC without a bridging innominate vein n. Initial palliation included interrupted aortic arch repair and pulmonary artery banding. A DKS procedure with BT shunt was performed at 5 months of age.	Orthotopic heart transplant with a. The LHV/LIVC was anastomosed to the donor right-sided IVC with tube graft(made of atrial tissue and porcine extracellular matrix). b. PA confluence deviated to left c. Bovine pericardium was to augment svc anastomosis. The donor to recipient right SVC anastomosis was constructed using a patch of bovine pericardium was used for the anterior wall to prevent stenosis. d. LSVC anastomosed to RA through bovine jugular vein anterior to ascending aorta e. In addition, for patient 2 the central right pulmonary veins were enlarged with a patch of porcine extracellular matrix
Matsuda H et al. [6]	2017	1	Left isomerism, single right ventricle, single atrium, CAVV, bilateral SVC, interrupted IVC with azygos connection, pulmonary stenosis (PS), dextrocardia, and s/p relief of PS and CAVV replacement	Orthotropic heart transplantation with a. Hepatic vein anastomosed to donor IVC. b. end to end anastomosis of RSVC. c. LSVC reconstruction with innominate vein posterior to ascending aorta.
Tobias D et al. [7]	2010	1	Situs inversus, dextrocardia, unbalanced AVSD, DORV, mitral and pulmonary atresia, and bilateral SVC.P/ BT shunt and BD glenn.	Heterotopic cardiac transplant. a. Apex rotated 120 degree to right. b. Donors SVC and innominate vein was anastomosed to recipient LSVC c. MPA was transposed to right side and large donors MPA patch was used. d. RSVC anastomosed to RA appendage.
Deuse T et al. [8]	2009	1	Situs inversus, dextrocardia, single ventricle, TGA, and pulmonic stenosis. P/Waterston shunt with Eisenmenger's physiology	Orthotopic heart lung transplantation with aortic repair at shunt side
Tector A J et al. [9]	2008	1	CCTGA, dextroversion of the left ventricle, situs solis, biventricular enlargement and failure, a large ventricular septal defect and unobstructed coronary arteries	Orthotopic heart transplant
Munoz G C et al. [10]	2008	1	Visceroatrial SI, dextrocardia, DTGA, short PM-VSD, ASD, SVC and IVC to LA, right aortic arch. P/Mustard.	Orthotopic heart transplantation a. Donors RA opened IVC to RA appendage and anastomosed to the right side of the atrial chamber that contained the drainage of the venae cavae through the Mustard baffle.

often associated with situs inversus. In dextrocardia with a normal situs solitus, then heart transplantation is challenging, as this may lead to a rightward rotation of the cardiac apex, as a result of that tricuspid valve and the inter ventricular septum may be distorted. If the pericardial space is dilated, this is not a major concern [14].

Appropriate donor selection is mandatory to prevent surgical difficulties during implantation. Size discrepancy between recipients and donors varies with age of the recipient. For neonates three times the weight of the recipient weight as donor's weight can be accepted. In case of older children donor's weight should be within 20% of the recipient weight. In case of significant weight discrepancy delayed sternal closure should be performed following diuresis till resolution of allograft myocardial edema. As much as donor tissue should be taken to in order to reconstruct at the time of implantation. Entire length of the superior vena cava, the left innominate vein, and the entire aortic arch, proximal portion of the descending thoracic aorta, branch pulmonary arteries from hilum to hilum, donor pericardium should be harvested as additional tissue for reconstruction [15].

Due to recipient factors and the increased complexity of the surgical implant procedure inspite of experienced surgical transplant team outcomes of heart transplantation for CHD is inferior to those for cardiomyopathy. Catastrophic mediastinal hemorrhage can be avoided during redosternotomy with proper planning including review of previous operative records and CECT chest. It is important to carefully plan for the operation, including review of prior operative reports and imaging studies. Exposure of groin vessels before sternotomy can be done if right ventricle or aorta is adhered to sternum as per preop CECT [16].

Residual lesions of recipient's heart identified during surgery should be repaired as it is one reason for the higher mortality seen in earlier eras for patients transplanted for CHD. In most univentricular lesions, reconstruction of the pulmonary arteries may be required typically with patch material.

Large amount of pulmonary venous return entering the left atrium in cyanotic congenital heart disease is due to aortopulmonary collateral and it may cause allograft rewarming and obscuring surgical field. In addition to obscuring the surgical field, this increased return also rewarms the allograft during implantation. This can be managed with a left ventricular vent, cooling to a lower temperature and decreasing the flow rate on cardiopulmonary bypass [17].

Three options are present for implantation of the allograft in the recipient:

a. Bicaval technique: This is the most common method and

lower risks of tricuspid insufficiency, improved right atrial transport function and less atrial arrhythmias are found in this method. In case of SVC stenosis patients are managed with balloon dilation and stent implantation.

b. Biatrial technique: In infant donor RA is anastomosed with recipient RA because there is chance of SVC anastomosis stricture due to small and fragile SVC in infant.

c. Individual pulmonary vein technique: In adult this can be done due to less incidence of postop LA thrombus. In paediatric population it is not commonly done due to higher incidence of atrial arrhythmia and individual pulmonary vein stenosis [18].

A left SVC commonly drains into the right atrium through a dilated coronary sinus. In case of normal innominate vein ligation and division of LSVC creates normal venous return. In case of small or absent innominate vein end-to-side anastomosis of the left SVC to the right SVC, end-to-end anastomosis of the left SVC to the right atrial appendage (direct anastomosis or anastomosis using a conduit), and anastomosis of both right and left SVCs to the innominate vein of the donor graft or keep the coronary sinus and inferior vena cava together en block and sutured to the donor inferior vena cava are the different options.. In rare scenario LSVC may drain directly into the roof of the left atrium, known as a Raghib association. In this situation, the left SVC is repaired either by an anastomosis to the left innominate vein or by construction of an intra-atrial baffle from the roof of the left atrium to the right atrium [19].

In CC-TGA heart transplantation is commonly done after previous operation like senning, mustard or double switch operation. During transplantation, the recipient's ascending aorta is transected superiorly, where the aorta is closer to the midline. The MPA is transected at the confluence and the incision is extended into the proximal origin of the left pulmonary artery. The rightward aspect of the pulmonary arteriotomy is closed to move the pulmonary artery bifurcation leftward which avoids PA kinking by the midline ascending aorta. During implantation aortic anastomosis is done after PA anastomosis for clear surgical field. The donor ascending aorta is kept long and anastomosed to the distal aspect of the recipient ascending aorta which moves the position of the ascending aorta rightward [20].

Situs inversus is a rare anomaly and it is often associated with dextrocardia. Rerouting of the systemic venous return is the main surgical challenge in transplanted heart. During cardiectomy, the atrial septum is not resected because it is used for reroute pulmonary venous drainage. The aorta and main pulmonary artery are transected distally. Mobilization of atrial septum is done by dividing it superiorly and inferiorly for and then anastomosed to the left atrial free wall, anterior to the right pulmonary veins. Anterior to the left pulmonary veins separate atriotomy is made to serve as the recipient neo-left atrium, which is now left-sided, corresponding to the donor heart. The leftward aspect of the right atrial cuff is closed, which moves the right atrial cuff rightward. The RA anastomosis is performed after the LA anastomosis. The PA anastomosis is performed next, using a similar technique as described above for CCTGA. Heterotaxy syndromes (also known as right or left atrial isomerism) often accompany situs inversus lesions and abnormalities of systemic and pulmonary venous return. The recipient operation combines elements of CCTGA and situs inversus [21].

Surgical challenges in transplantation of the fontan patient are due to multiple previous sternotomy and longer ischemic time due to fontan takedown and pulmonary artery reconstruction. During donor cardiectomy extra length of PA should be taken and during recipient exploration pericardium should be taken for PA reconstruction after Glenn or fontan [22]. In transplantation after Norwood procedure extra length of donor aorta and PA should be taken [23].

Conclusion

Cardiac transplantation is challenging and gradually improving with emerging techniques and critical care management. For transplantation in dextrocardia appropriate preoperative planning is needed and preoperative surgical record should be checked and CECT thorax should be done to prevent meditational haemorrhage after resternotomy. CT volumetric analysis is proffered to avoid donor recipient mismatch. As it is very difficult to get dextrocardia donor heart is implanted with excision of pericardium and lobectomy may be required in rare scenario and apical rotation of the heart is required.

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