

Total Situs Inversus and D- Transposition of Great Arteries Managed in 2 Surgical Stages. Case Report

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1. Abstract

A case report of a pediatric patient in whom multiple congenital cardiac malformations coexist is presented below. The presentation of D-transposition of the great arteries (TGA) associated with total situs inversus and pulmonary artery atresia presented a challenge in surgical management, however, it could be carried out successfully using the Rastelli technique.

2. Introduction

Situs inversus totalis is a rare congenital anatomic variant in which thoracic and abdominal organs are reflected mirror-like respecting the sagittal axis of the body. This is considered an anomaly with multiple factors involved including genetic factors and factors during the development of the pregnancy [1].

The prevalence of this entity is estimated between 1-2/10.000 new-borns and it is usually associated with other cardiovascular malformations [2]. In the National Birth Defects Prevention Study, the prevalence of congenital cardiopathies associated with a situs inversus totalis was 41%. From these, the most common were transposition of great arteries (TGA), pulmonary stenosis, ventricular septal defects (VSD) and atrial septal defects (ASD) [1].

On the other hand, TGA is a congenital malformation in which the aorta emerges from the right ventricle, whereas the pulmonary artery emerges from the left ventricle. This anomaly can be classified in D-transposition of the great arteries when the aorta is anterior and to the right from the pulmonary artery. Meanwhile, in L- transposition the aorta is located to the left of the pulmonary artery and it is considered physiologically corrected because of both

atrioventricular and ventricular-arterial discordance [3]. In order for this malformation to be compatible with life there must be a connection among both pulmonary and systemic systems, either an ASD, a VSD or a persistent ductus arteriosus (PDA) [4].

The following article seeks to demonstrate the surgical management of a patient with multiple coexisting congenital defects. Due to this morphological as well as functional malformation it is considered a high complexity level surgery managed in 2 stages.

3. Case presentation

3.1. Pre-surgical

This is a 2-year-old patient, first gestation, cesarean delivery due to unsatisfactory fetal status. A complex congenital cardiopathy was diagnosed postnatally, consisting of situs inversus with levocardia, D-TGA with pulmonary valve atresia, perimembranous VSD of 9.5 mm with extension to the inlet tract, ASD type sinus venosus defect of 7.4 mm and a right aortic arch. Secondary to the cardiac malformations congestive heart failure was present. On April 2019 a systemic-pulmonary Blalock Taussig shunt was made from the left subclavian artery to the left pulmonary artery that was normally functioning. On July 2019 PDA closure was made and in September of the same year the patient underwent a balloon angioplasty due to pulmonary stenosis.

The patient consults on March 2021 due to nocturnal diaphoresis, fatigue while feeding, perioral cyanosis and desaturation, which led to increase the oxygen requirement from 0.25 L/min to 0.5 L/min. It was considered a decompensated heart failure due to an underfunctioning shunt and right pulmonary artery stenosis.

Given this findings, the patient was taken to bilateral catheterization and pulmonary arteriography and right pulmonary artery angioplasty. With the catheterization results in a multidisciplinary meeting it was decided to take the patient to surgery and during the surgery decide between a Glenn Fontan-Kawashima approach versus clousure of the VSD with an intraventricular tunnel and Rastelli procedure creating an extracardiac conduit between the pulmonary ventricle (located on the left) and the pulmonary artery trunk, therefore ligating the sistemic-pulmonary shunt.

3.2. Surgical Procedure

On March 17th 2021 the patient was taken to surgery where we found a situs inversus totalis, with superior and inferior vena cava located at the left side, arriving into the physiological right atrium located on the left, with the tricuspid valve towards the pulmonary ventricle. From this ventricle the atresic pulmonary artery emerged. On the right side, was the physiological left atrium receiving the 4 pulmonary veins. Through this physiological left atrium, the mitral valve, the sistemic ventricle and the VSD were assessed (Figure 1). The sistemic pulmonary shunt was ligated with silk, the VSD was corrected with a Dacron patch of 18 x 9 mm and the ASD was closed with a pericardium patch. The atresic pulmonary artery trunk was ligated, and the Rastelli procedure was done with a Contegra graft of 18 mm creating a conduit between the pulmonary ventricle and the pulmonary artery (Figure 2). Heart activity was recovered with an atrioventricular block, which led to initiating stepmaker stimuli with good contractility. The patient exited extracorporeal circulation with acceptable hemodinamics, time of ischemia 135 minutes, time of perfusion 179 minutes, intrasurgical bleedeeing of 150 cc, required transfusion of one unit of blood and infusion of milrinone 0.5 mcg/kg/min, norepinefrine 0.05 mcg/kg/min, epinephrine 0.08 mcg/kg/min and nitroglycerin 0.5 mcg/kg/min. The procedure ended with no complications and the patient was taken to the pediatric intensive care unit.



Figure 1: Cardiac anatomy prior to surgical intervention

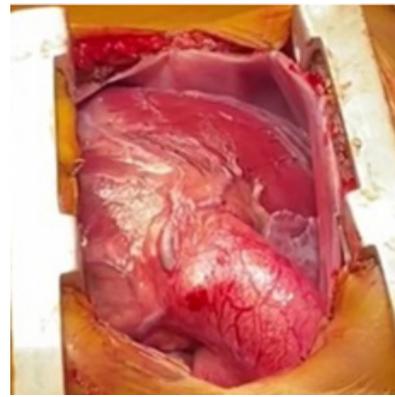


Figure 2: Insertion of extracardiac conduit with Contegra graft

3.3. Post-surgery

The postoperative ecocardiogram showed both left and right centricular systolic function was maintained with LVEF of 50%, right pulmonary artery stenosis with gradient of 49 mmHg, right ventricular overload and moderate tricuspid regurgitation. The patient was breathing through a mechanical ventilator with adecuate pulse oximetry, however he developed metabolic acidosis, increased requirement of vasopresor and inotropic support, pacemaker dependent and elevated lactate levels. In third day POP the patient had elevated temperature, altered coagulation times and bad general stage. It was considered a multisistemic failure associated to an increased right pulmonary artery gradient. A meeting was held between cardiology, hemodinamics, cardiovascular surgery, pediatric intesive care and anesthesia departments and it was decided to take the patient to a second catheterization to review named artery due to the risk of compromising in the long term the Contegra graft and overload the pulmonary ventricle.

3.4. Second Catheterization

The second catheterization showed a gradient between the right pulmonary artery and the right ventricle of 50 mmHg. A bilateral pulmonary arteriography was made and it demonstrated a 4 mm stenosis in the medial and distal third of the right pulmonary artery, which led to passing a coronary balloon 4x20 through the stenosis with adecuate opening. The procedure ended with no complications, the stent in adecuate position and a good blood flow. After the procedure the patient remained in bad conditions, with requirement of low dose vasopresors (norepinephrine 0.12 mcg/kg/day, epinephrine 0.17 mcg/kg/day, milrinone 0.3 mcg/kg/day), pacemaker dependat, on mechanical ventilation with a moderate oxigenation disorder and developed diarrhea and fever, norovirus was isolated and caspofungin therapy was initiated.

3.5. Hemopericardial Drainage

A chest X-ray was taken with evidence of cariomegaly, and the ecocardiogram showed septate pericardial effusion, that within the spam of 3 days increased by 6 mm, so a pericardial window was made on april 2021. During the procedure a hemoperical colection of 100 cc was found coming from one of the sutures from

the graft. A culture of the drained fluid identified *Klebsiella*, that led to starting cefepime and vancomycin therapy in addition to the caspofungin. Later on, we switched cefepime for meropenem under infectology recommendations.

The patient continued with stationary evolution, hemodynamically stable, until he developed arrhythmias associated with malfunctioning of the temporary epicardic pacemaker that led to deterioration. A multidisciplinary meeting concluded it was best to take the patient to a definitive pacemaker insertion posterior to the infectious process. The patient remained stable, without new fever episodes and negative cultures for 48 hours, and on April 19th through a thoracotomy he got a St. Jude Assurity unicameral pacemaker inserted. After the procedure the patient had good evolution, with adequate response, however the fever re-started and antibiotic therapy was reinstated.

3.6. Evolution and Discharge

On May 2021 the patient was stable without support, with oxygen through nasal cannula 2 L/min, the antibiotic therapy ended. He was transferred from the pediatric ICU to the pediatrics floor to continue recovery. He developed post extubation croup, received IM dexamethasone and micronebulizations with budesonide, with no extra measures required. During the rest of his hospital stay he remained stable, with surgical wound healthy and no signs of infection, blood pressure within normal, less oxygen requirements, normal urinary output, no new fever episodes, with no neurological damage.

He was discharged on June 23rd 2021 with oxygen 0.5 L/min, ASA, hydrochlorothiazide, sildenafil, furosemide and prophylactic amoxicillin. Final ecocardiogram showed POP D- transposition of great arteries with Rastelli procedure, situs inversus with levocardia, Contegra graft permeable, biventricular function preserved. LVEF 60%.

4. Discussion

On patients with TGA with no VSD the surgical technique of choice is the arterial switch, in which the aorta including the coronary arteries and the pulmonary artery are cut and reconnected to their respective ventricles. However, TGA in patients with other associated defects, like the case of our patient with situs inversus totalis and pulmonary artery atresia the preferred surgical repair is the Rastelli procedure, designed in 1969. This technique consists of creating an extracardiac conduit from the pulmonary ventricle to the pulmonary artery, this allows to reconstruct both ventricle outlet tracts and separate the systemic and pulmonary circuits [5]. Even though this procedure has had plenty of successful outcomes, there is one long term disadvantage, and is the need of re-intervention due to obstruction of the right ventricular outflow tract. [5]. In a study made in Riley Hospital for Children at Indiana University Health, the investigators evaluated long term results from 47 patients that underwent Rastelli procedure from 1988 to 2017.

This study revealed that the Kaplan-Meier freedom from death or cardiac transplantation was 93% in 5 to 10 years and 84% in 15 to 20 years. From the 47 patients in the study, 51% required conduit replacement, the Kaplan-Meier calculation from these population was 85% in 5 years and 24% in 15 years. The main causes were conduit stenosis (92%), severe pulmonary regurgitation (8%) and only one case of bacterial endocarditis. 3 additional patients required a second conduit replacement due to endocarditis and stenosis [6].

Because of this complications in 1994 the Nikaidoh technique was developed to treat this anomaly [5]. In this procedure the ascending aorta and the pulmonary trunk are sectioned, the aorta is translocated in the left ventricular outflow tract and the pulmonary trunk to the right ventricular outflow tract with re-implantation of the coronary arteries [5].

This particular patient was a high complexity case with a previous systemic pulmonary shunt, as well the multiple malformations coexisting (D-TGA, VSD, ASD, pulmonary artery stenosis and situs inversus totalis) in which a biventricular reconstruction was proposed. However, with the interdisciplinary work of multiple specialties both anatomic and physiological difficulties were overcome with a good final outcome, leaving a patient with biventricular physiology and a better short and long term prognosis.

References

1. Chen W, Guo Z, Qian L, Wang L. Comorbidities in situs inversus totalis: A hospital-based study. *Birth defects research*. 2020; 112(5): 418-426.
2. Urquia H, Discua L, Valenzuela R. Situs inversus totalis y cardiopatía congénita: diagnóstico casual en un lactante con neumonía. *CIMEL Ciencia e Investigación Médica Estudiantil Latinoamericana*. 2010; 15(2): 85-88.
3. Valente A, Landzberg M. Capítulo 264: Cardiopatía congénita en el adulto. En: Jameson J, Fauci A, Kasper D, Hauser S, Longo D, Loscalzo J. *Harrison. Principios de Medicina Interna*. 20° edición. McGraw-Hill Education. 2018.
4. Warnes C. Congenital heart disease for the adult cardiologist: Transposition of the great arteries. *Circulation*. 2006; 114(24): 2699-2709.
5. Susana A. Cirugía de Rastelli en anomalía de la conexión ventriculoarterial asociada con defecto septal ventricular y obstrucción del tracto de salida ventricular. *Revista Argentina de Cardiología*. 2010; 78(4): 302-304.
6. Huang E, Herrmann J, Rodefeld M, Turrentine M, Brown J. Rastelli Operation for D-Transposition of the Great Arteries, Ventricular Septal Defect, and Pulmonary Stenosis. *World Journal for Pediatric and Congenital Heart Surgery*. 2019; 10(2): 157-163.