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CLINICAL CASE

Switch after Switch. Ventricular retraining as alternative to heart transplant

Switch después del Switch. Reentrenamiento como alternativa ante trasplante cardiaco

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What do we know about the subject matter of this study?

Systolic dysfunction of the systemic ventricle after atrial switch is a serious complication in the immediate postoperative period. Pulmonary artery banding may help stabilize and eventually retrain these patients until they can be considered candidates for late anatomical correction.

What does this study contribute to what is already known?

We present the case of a patient in the postoperative period following atrial switch surgery with severe early right ventricular (systemic) dysfunction, with heart transplantation being the only viable option. Ventricular retraining was considered to avoid heart transplantation and achieve an adequate quality of life. Given the low prevalence of this disease, its unusual presentation, and the few reports in the literature, this case is deemed important to report.

Abstract

Transposition of the great arteries (Dextro-TGA), repaired with physiological correction techniques (atrial switch - Mustard or Senning surgery), can present as a complication the failure of the right ventricle that acts as systemic and, at the same time, deconditioning of the left ventricle, leading to congestive heart failure. In these patients, treatment and recovery options are very limited. **Objective**: To describe successful late anatomical correction after ventricular retraining. **Clinical Case:** Patient diagnosed with Dextro-TGA, with multiple perinatal complications, treated in another institution with Mustard surgery at seven months, who developed refractory heart failure in the postoperative period. Given the option of a heart transplant, pulmonary banding was decided at 11 months of age as a stabilization measure, which subsequently allowed retraining of the left ventricle and led to anatomical correction with removal of the Mustard procedure and successful large artery switch at six years of age. Three years after surgery, the patient maintains good quality of life and functional class II. **Conclusions:** In selected cases, such as the one described, ventricular retraining can be a valid option for patients with transposition of the great arteries who have had a previous physiological correction and have developed right ventricular dysfunction (systemic).

Keywords:
Congenital Heart Disease;
Transposition of Great Arteries;
Heart Failure;
Arterial Switch Surgery

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Introduction

Simple d-transposition of great arteries (DTGA) is a congenital heart disease where there is ventriculoarterial discordance (the right ventricle is connected to the aorta, while the pulmonary artery emerges from the left ventricle). Worldwide, it has a prevalence of 4.7 per 10 thousand live births[1], and in Colombia 0.6 per 10 thousand live births [2].

The atrial switch or physiological correction with Mustard (figure 1) or Senning surgeries, consists of the creation of an intra-atrial baffle to redirect systemic venous blood flow to the left ventricle (connected to the pulmonary artery) and pulmonary venous blood flow to the right ventricle (connected to the aorta). In this way, physiological correction is achieved, but the right ventricle remains systemic.

This procedure was the mainstay of surgical repair during the '60s and '80s. However, this physiology imposes the right ventricle and tricuspid valve to support the systemic flow, therefore over time, right-sided heart failure occurs due to myocardial fiber dysfunction and/or tricuspid valve regurgitation, while the left ventricle is deconditioned, losing muscle mass, and therefore, its ability to withstand high pressures[3]. Survival of this procedure is close to 60% at 10 years[4].

Left ventricular retraining is a technique developed by Yacoub et al. for TGA with intact septum, in which the left ventricle becomes dysfunctional[5] (figure 2). Later, in 1986, Mee et al. used this technique for patients with TGA who had undergone an atrial switch and subsequently developed right-sided heart failure[6,7].

The technique consists of performing a pulmonary artery banding, which limits blood flow by placing a band around the circumference of the pulmonary artery trunk, then it is adjusted to reduce the vessel diameter, thereby limiting blood flow. Ventricular pressure is measured directly and is expected to be 70% of systemic pressure. The left ventricle, which in this type of physiology handles low pressures, will face the increased afterload with compensatory mechanisms such as increased contractility, hypertrophy with ventricular mass increase, and changes in ventricular geometry. This process is referred as ventricular retraining , and it is performed to prepare the left ventricle to withstand systemic arterial pressure[8].

Usually, one or two more banding adjustments are required to increase intraventricular pressure to help the left ventricle withstand systemic pressures. Once the established objectives are achieved, the anatomical correction of the TGA can be performed with Jatene surgery or arterial switch[7], which consists of the pulmonary artery and aorta section and its connection with the due ventricle, reimplanting the coronary arteries, or also called two-stage arterial switch[9].

Currently, this type of case has a low prevalence, and there are few published in the literature; therefore, the objective of this manuscript is to describe a case of late anatomical correction after ventricular retraining.

Clinical Case

We present the case of a 2,5 kg, 36 weeks preterm male newborn, from a mother with regular prenatal care, , with no anomalies detected ,delivered in a primary care institution. He presented multiorgan dysfunction and an episode of cardiac arrest within the first 24 hours of life. He was transferred to a higher complexity institution where a DTGA diagnosis was made. After stabilization, he underwent an atrial septostomy on the second day of life. Due to the clinical condition arterial switch was not performed.

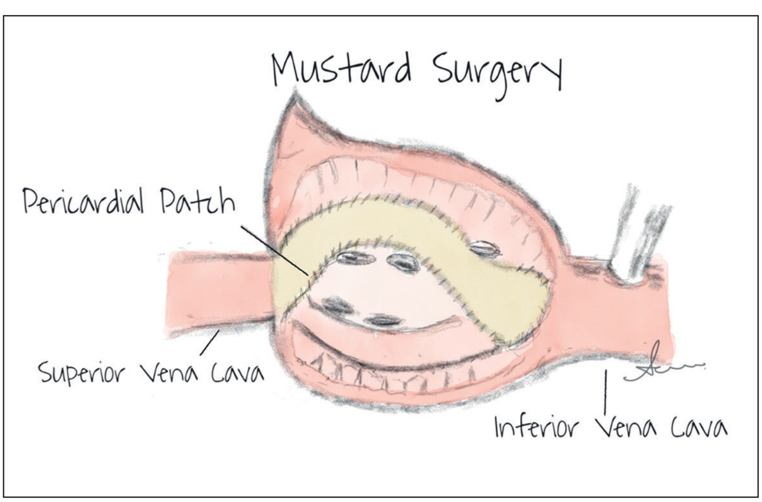

Figure 1. Mustard Surgery. Illustration for the Surgeon's View physiological correction of a DTGA with a Mustard Surgery. The tunnel was created with pericardial patch directing the venous blood flow form the SVC and IVC to the left ventricle which is connected to the pulmonary artery. The patch excludes the coronary sinus. The four pulmonary veins are draining to the right atrium that is connected to the right ventricle and to the aorta. The illustration shows the cannula for cardiopulmonary bypass in situ. Drawing done by: Sara Mendoza. MD.

He progressed with multiorgan dysfunction and pulmonary hypertension and required mechanical ventilation for two months with a prolonged stay in intensive care. The patient developed subpulmonary left ventricular dysfunction ("banana-shaped" ventricle), with severe systemic tricuspid valve regurgitation.

At 7 months of life, Mustard surgery and tricuspid valve plasty were performed. During the postoperative period, he presented with severe biventricular dysfunction and end-stage heart failure. Heart transplantation was considered the only viable option; therefore, he was referred to a high complexity hospital.

Admission evaluations confirmed the left "banana-shaped" ventricle and severe right systemic ventricular dysfunction, with moderate to severe tricuspid regurgitation. There was no evidence of a stenotic gradient in the Mustard baffles or coronary pattern alteration.

The patient remained inotrope-dependent and after 4 months on the transplant waiting list without available donor, a decision was made to perform pulmonary artery banding for left ventricular retraining.

At 11 months of life weighing with 6.9 kg of weight, pulmonary artery banding (PAB) was performed, obtaining a peak gradient of 20 mmHg and a ventricular mass of 19.3 g/m^2, after it was possible to wean off theinotropic support.. Once stabilized, management of heart failure and outpatient levosimendan boluses were continued. Brain natriuretic peptide (BNP) changed from 330 pg/mL before the first banding, to a minimum value of 48 pg/mL after the procedure.

Given the adequate clinical response, a new banding adjustment was performed at 20 months of life with 9 kg of weight, achieving 50 mmHg of gradient increase, with a ventricular mass of 50 g/m^2, and a change in left ventricular morphology. BNP increased to 272 pg/mL in the first year after the second banding, although the trend was downward afterward (figure 3).

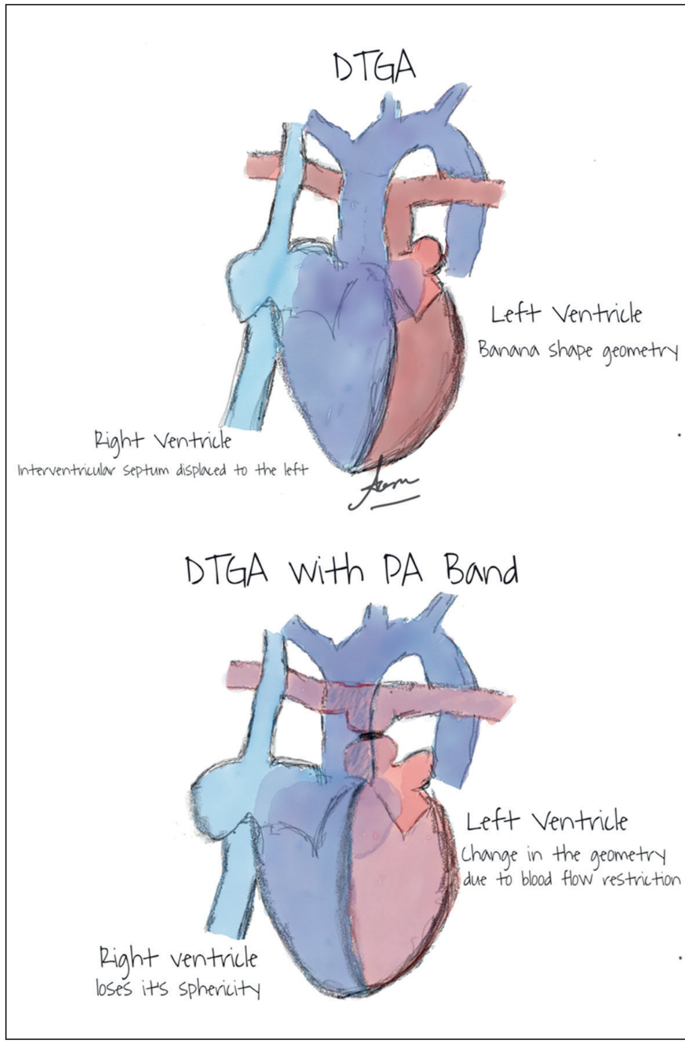

Figure 2. DTGA (Top Drawing):The great vessels are connected invertedly. The pulmonary artery to the left ventricle and the aorta to the right ventricle. Image of the training band (bottom drawing), on the pulmonary artery of a drawing of the same heart disease, method used for ventricular retraining described by Yacoub. Drawing done by: Sara Mendoza. MD.

Four months after the second PAB, the last bolus of levosimendan was given. He remained on oral medicationand strict clinical follow-up for pediatric heart failure for 6 years, with a progressive response, meeting some of Stanford's criteria for adequate left ventricular retraining (table 1). The BNP dropped to 104 pg/ml, which was considered quite acceptable for the physiology at the moment and supported the decision to proceed to full correction (figure 3). Figures 4 to 7 show the evolution of LV re-training. Based on previous data, it was decided that he was a candidate for anatomical correction. Jatene surgery was performed with coronary reimplantation and LeCompte maneuver, Mustard removal, atrial reconstruction, banding removal, and reconstruction of the pulmonary artery. After this procedure, the patient continued outpatient follow-up for pediatric heart failure for 3 years, with functional class II and normalized BNP.

Regarding echocardiographic characteristics, he continued with 53% left ventricular ejection fraction, 45% right ventricular ejection fraction, and no fibrosis, although with criteria for impaired diastolic relaxation (figure 8). On clinical evaluation, he was stable, attending school, performing routine activities and sports, and he continued closely monitored at the pediatric heart failure clinic. Heart transplantation was not considered again.

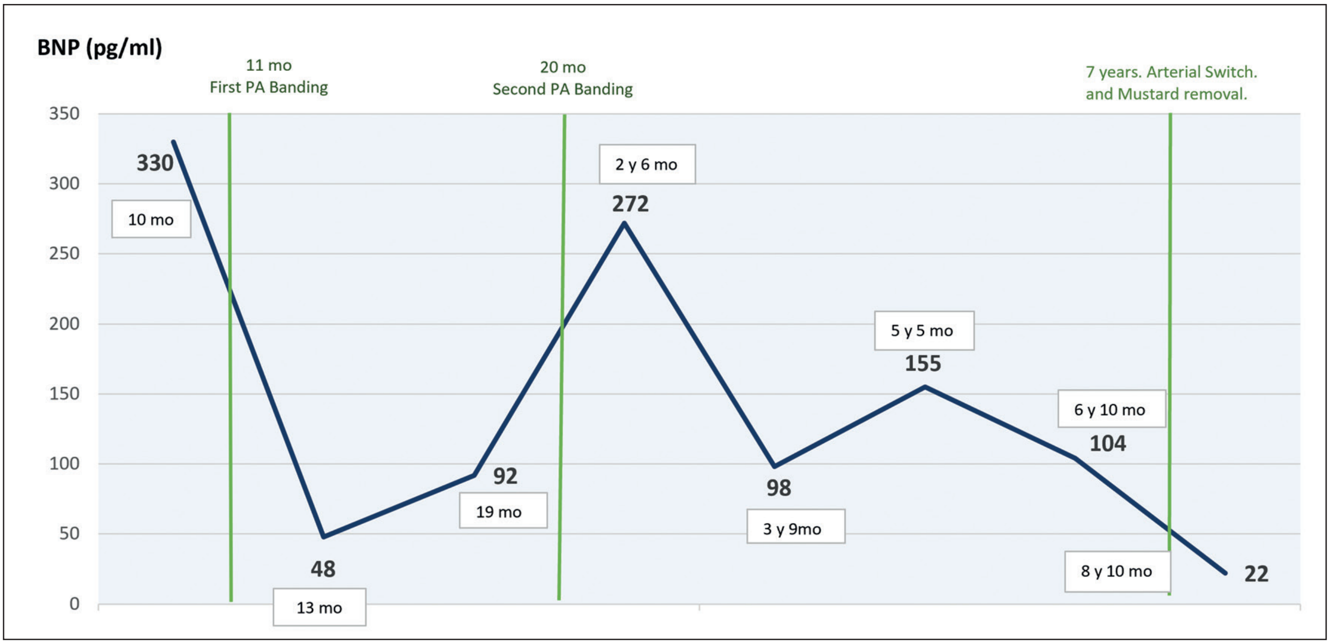

Figure 3. Historic BNP levels.

Table 1. Stanford´s Criteria Met by the Patient

Measurement	Stanford´s Criteria	Patient	Agreement
Left Ventricular Pressure (LV)	90% of systemic pressure	> 90% of systemic pressure	Yes
LV systolic Function	EF > 55 %	42 %	No
LV End Diastolic Pressure	< 12 mmHg	14 mmHg	No
Mitral Valve Function	Less than mild regurgitation	No regurgitation	Yes
LV mass	> 50 gr/m^2	112 gr/m^2	Yes

Left Ventricle (LV), Ejection Fraction (EF). Modified from Mainwaring RD., et al[11].

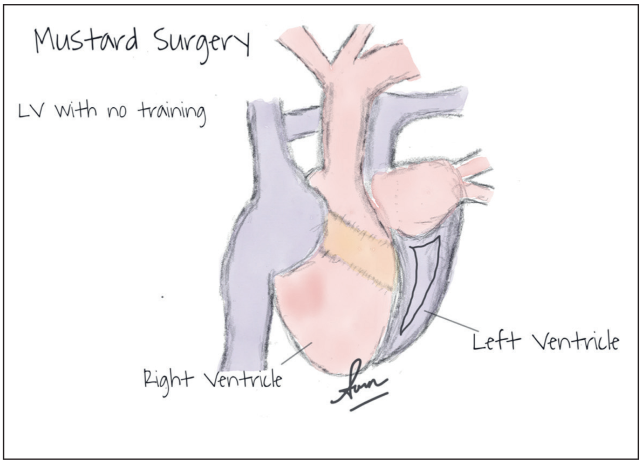

Figure 4. Illustration (Coronal plane) depicting the initial anatomy of the patient with Mustard surgery. The untrained left ventricle (banana shape) is represented. Venous blood in violet color and arterial blood in pink color. Drawing done by: Sara Mendoza. MD.

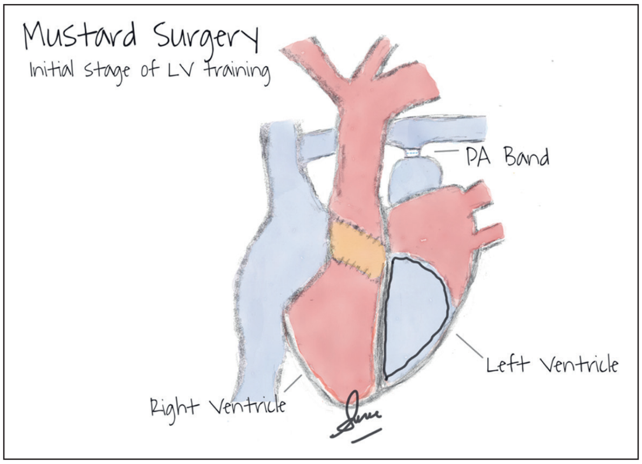

Figure 5. Coronal view diagram representing the cardiac anatomy after the first pulmonary band, performed at eleven months of age. A band of PTFE (Polytetrafluoroethylene) knotted in the trunk of the pulmonary artery can be seen. Displacement of the interventricular septum and improvement of left ventricular morphology should be noted. In addition, the morphology of the right ventricle is modified with a reduction in the size of the ventricle that favors a better coaptation of the tricuspid valve. Drawing done by: Sara Mendoza. MD.

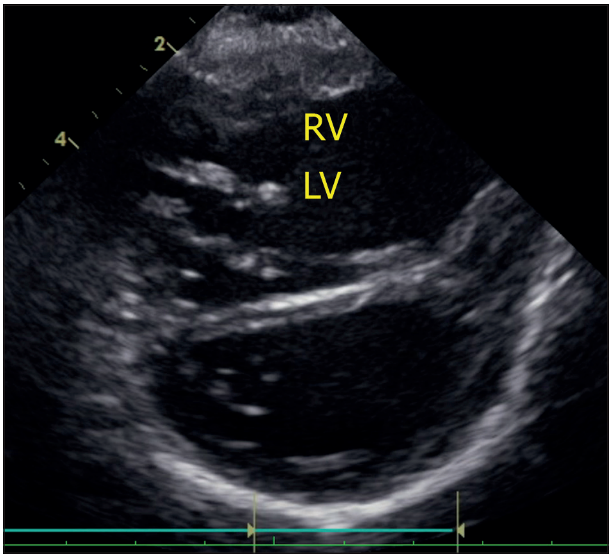

Figure 6. Echocardiogram image in short-axis view at the level of the ventricles showing the late evolution after the second pulmonary artery band during the retraining period (Age 4 years). The image shows the rectification of the septum, left ventricle (LV) on the lower wall of the image and right ventricle (RV) on the upper wall.

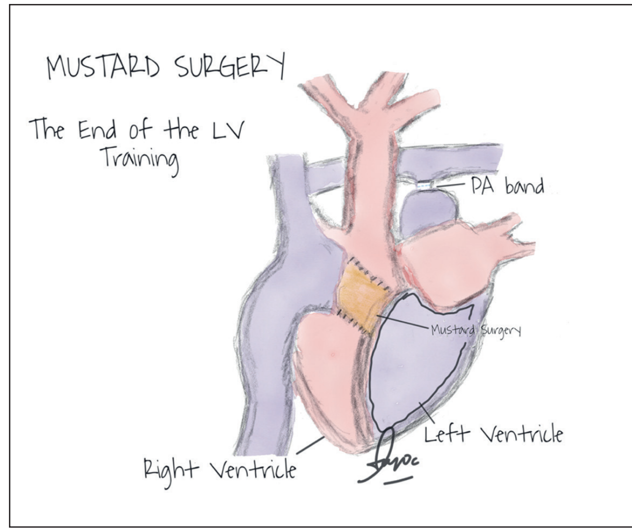

Figure 7. Coronal view illustration depicting cardiac anatomy at the end of ventricular retraining: After a second adjustment of the pulmonary band and medical management for six years. The left ventricular morphology was recovered. The interventricular septum adopts a position close to normal, which is slightly displaced towards the right ventricle. Drawing done by: Sara Mendoza. MD.

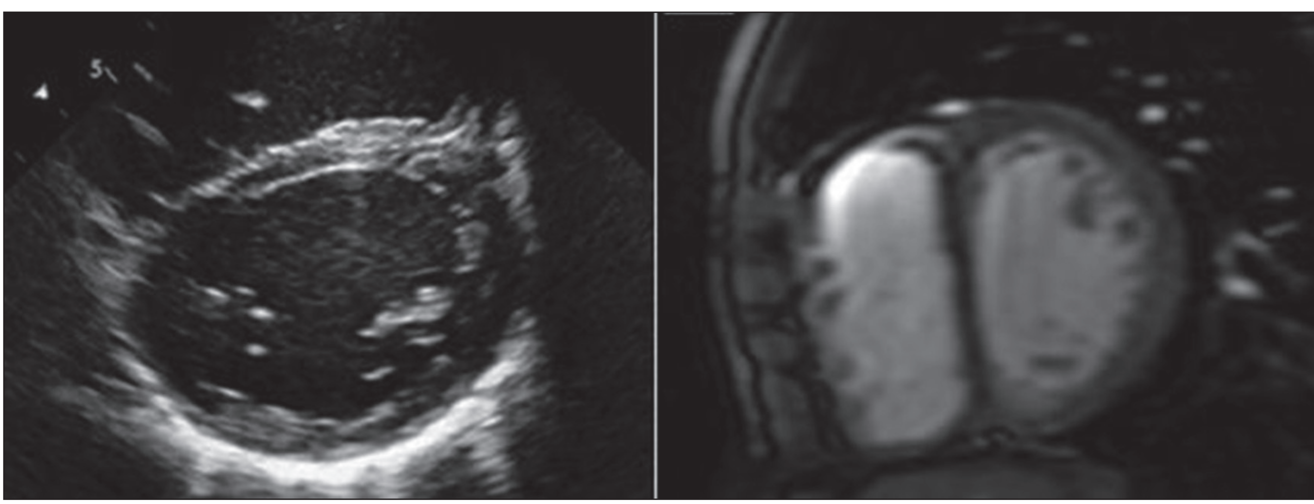

Figure 8. Short-axis echocardiogram image at the level of the left ventricles and MRI in the same view performed at 9 years of age, where the left ventricular morphology can be seen one year after anatomical correction.

Discussion

Left ventricular retraining has been described as a method to recondition the ventricle to systemic pressures by banding the pulmonary artery. In some patients, the creation of a systemic artery-to-pulmonary artery fistula is necessary if cyanosis prevents adequate training. This procedure is used in three specific cases: i) DTGA presenting late to arterial switch; ii) patients who have undergone atrial switch surgery who developed severe right ventricular dysfunction and/or tricuspid valve regurgitation; and iii) patients with congenitally corrected transposition with untrained subpulmonary left ventricle, when the goal is anatomical repair[4,10].

This surgery was developed by Dr. Mee et al in 1986[6]. Despite the historical nature of this procedure and the low number of patients who may require this management, there was no consensus on the retraining method, until 2018, when the Stanford criteria were published[11]. These criteria were used for the surgical decision in the case described. Our patient did not meet two criteria: end-diastolic pressure, which was slightly elevated comparedt to the normal range, and ejection fraction, which should have been normal according to the criteria, however, in our patient, was in the range of mild dysfunction (42%).

The decision to proceed with surgical correction was based on the patient´s good clinical condition concordant with the evolution of BNP during follow-up (figure 3), a biomarker that is produced in ventricular cardiomyocytes in the presence of pressure and volume overload, and its elevation correlates with systolic dysfunction of the adult left ventricle[12]. Price et al. predicted worse 90-day survival in children with left ventricular systolic dysfunction[13]; In another study, Auerbach et al. demonstrated that a BNP level ≥ 140 pg/mL identified a poorer prognosis risk for children older than 2 years with moderate symptoms of heart failure[14]. It has been reported than in children with congenital heart disease and heart failure, elevated levels of this biomarker correlates with the severity of heart failure[15].

In this patient, right ventricular dysfunction occurred very early after the Mustard procedure , limiting available therapeutic options. In patients with DTGA, the subpulmonary left ventricle naturally tends to lose its geometry, ventricular mass, and function around the second postnatal week, this phenomenon is termed ‘defunctionalization’[8]. Meanwhile, the right ventricle, connected to the aorta, is physiologically unprepared to withstand systemic pressure. The patient´s postnatal clinical deterioration, including cardiac arrest, likely contributed to greater deterioration. By the time he underwent the Mustard procedure, he had developed severe tricuspid valve regurgitation. The impact of this complex surgery may have contributed to severe biventricular dysfunction in the postoperative period.

Transplantation in children under one year of age is a challenge due to the scarcity of donors. The waiting list mortality rate has been reported up to 25% for this age range in the United States before a suitable donor becomes available[16]. In addition to the multiple risks inherent to post-transplant immunosuppressive management, the decision was made to retrain the left ventricle. The patient required two adjustments of the pulmonary banding with an interval of nine months, along with both intravenous and oral management of heart failure andprolonged, strict clinical follow-up. Although there is a risk of progression of diastolic dysfunction, thoroughfollow-up for pediatric heart failure, has allowed the patient to remain in functional class II. We consider that left ventricular retraining was an excellent option for this patient, faced with the prospect of heart transplantation, it became a bridge to recovery and successful anatomical repair.

Conclusion

Ventricular retraining, usually employed for DTGA with a detrained left ventricle, may be an option for late correction in patients with a primary physiological approach (Mustard or Senning), in whom systemic ventricular function has deteriorated. To the authors’ knowledge, we report the first successful case in Colombia of this strategy in a patient previously considered end-stage and referred for heart transplantation.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

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