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Transposition of Great Vessels: Latin-American Postoperative Experience

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1. Abstract

1.1. Background: The transposition of great vessels (TGV) is a congenital cardiac malformation where atrioventricular concordance is conserved with ventriculoarterial discordance. Regarding the management of this pathology, surgery is the cornerstone treatment. We aimed to describe the surgical characteristics of the pediatric population with transposition of great vessels surgically managed by our cardiovascular surgery group.

1.2. Methods: A retrospective analysis of a prospectively collected database was conducted for patients who underwent surgical management for TGV between 2009 and 2021 in our institution. We described preoperative, intraoperative, and postoperative characteristics, morbidity, mortality, early and late follow-up.

1.3. Results: A total of 34 patients were included. The mean maternal age was 23.8 years. Majority of the patients were male (67.6%). Types of transposition were distributed as: 73.5% complex dextro-transposition, 17.6% non-complex dextro-transposition, 5.88% complex levo-transposition and 2.9% non-complex levo-transposition with no patients presenting simple transposition. The most prevalent malformation associated was atrial septal defect with persistent ductus (32.3%). The mean ischemia and perfusion times were 125 and 179 minutes respectively. Cardiogenic shock was present in 41% of the patients, 71.4% of whom died intraoperatively. All patients were admitted to the intensive care unit. Average length of stay was 10.4 days. Mortality at 30 days was 41% and at one year 7%.

1.4. Conclusion: The characterization of patients with TGV is essential to specify and recognize the sociodemographic and clinical characteristics of this pathology, this aimed at establishing associations, within the natural course of this congenital heart disease, and its clinical outcomes.

2. Introduction

TGV is defined as a congenital cardiac malformation where there is atrioventricular concordance with ventriculoarterial discordance [1-3]. The most frequently mentioned are D-TGV and L-TGV, referring to the positioning of the aorta in relation to the trunk of the pulmonary artery. TGV in general represents the sixth most frequent congenital heart disease (CHD), behind ventricular septal defect (VSD), atrial septal defect (ASD), aortic stenosis (AS), pulmonary artery stenosis (PS), and aortic coarctation (AC) [2]. This pathology corresponds to 5-8% of all congenital heart diseases [1-5]. Before 1964, approximately 90% of infants with this condition died within the first year of life [2-6] and 45% of untreated infants died within the first month [3]. Due to the evolution and advances in diagnosis, the morbimortality of infants has been reduced over the years [3,4].

Different diagnostic methods have been described for TGV [6-8]. The main goal of imaging is to provide accurate characterization of the pathology to allow physicians to plan a respective surgical intervention [4]. This, associated with other diagnostic modalities such as hyperoxia test, cardiovascular magnetic resonance, cardiovascular computed tomography, nuclear scintigraphy, exercise

and stress imaging, catheterization and angiography have been described [4-9] to fully understand each patient. Despite each imaging modality having different strengths and limitations, echocardiography remains the preferred diagnostic modality secondary to its widespread availability and portability [9-11]. The main echocardiographic findings assessed by transthoracic echocardiography allow us to evaluate levels of shunting, the relationship between great vessels, ventricular function, the severity of outflow tract obstruction, valve function, and additional abnormalities [4,11]. Other diagnostic methods such as magnetic resonance imaging (MRI) and computed tomography (CT) have been proposed as diagnostic options, especially when concomitance with other cardiac abnormalities is suspected [1,12].

In terms of management, the main focus of palliative treatment is to ensure the correct exchange of blood between the cardiac cavities. Among the proposed strategies is prostaglandin E1 (PGE1) to maintain ductal patency until the comprehensive series of postnatal echocardiograms is complete [6,13-15]. In the same line, corrective treatment includes arterial switch operation (ASO), a procedure showing 5-, 10- and 15-year survival of up to 85% [6,7]. Regarding this, the European Associations for Cardio-Thoracic Surgery (EACTS) and the Associations for European Pediatric and Congenital Cardiology (AEPC) recommended that a primary ASO in neonates with TGV should be performed from the first few days to 3 weeks of life [6]. The characterization of the population and surgical performance is well documented in developed countries. Nevertheless, in developing countries such as Colombia, there is scarce information on this subject. Therefore, we aimed to describe the preoperative, intraoperative, and postoperative characteristics of the pediatric population with the transposition of great vessels surgically repaired by our cardiovascular surgery group.

3. Methods

3.1. Study Population

With the Institutional (Pontificia Universidad Javeriana and Hospital Universitario San Ignacio) Review Board's approval and following Health Insurance Portability and Accountability Act (HIPAA) guidelines, a retrospective review of a prospectively collected database was conducted. All patients with the diagnosis of TGV who received surgical management between 2009 -2021 were included. Patients with missing data, and follow-up < 1 month were excluded. Ethical compliance with the Helsinki Declaration, current legislation on research Res. 008430-1993 and Res. 2378-2008 (Colombia), and the International Committee of Medical Journal Editors (ICMJE) were ensured under our Ethics and Research Institutional Committee (IRB) approval.

3.2. Data Assessment

Patients' demographics and associated comorbidities were evaluated. Prenatal assessment, clinical variables include preoperative weight and height, duration of symptoms until surgical procedure, heart failure at the time of surgery, type of preoperative diagnosis, left ventricular function, and stenosis gradient (based on preoperative echocardiogram). Operative variables including, type and urgency of surgical procedure, type of bicuspid aortic valve, perfusion, and ischemic time were evaluated. Postoperative mortality was evaluated at 30 days follow-up, and survival rate up to 1 year after surgery was defined according to the national database.

Descriptive statistics of all study parameters were provided according to the nature of the variable. The distribution of the variables was assessed according to the Kurtosis/Skewness test. Continuous variables were summarized by means or medians and standard deviation or interquartile ranges according to their nature and distribution. Categorical data were summarized by their frequency and proportion. Data were analyzed using STATA 17 licensed version.

4. Results

4.1. Demographics

A total of 34 patients were included. Male patients corresponded to 67.6% (n=23) and the men to women ratio was 2.09:1. The mean age of admission to the institution was 2 months with a mean parity of 1.9 gestations and a median maternal age of 23.8 years. The median number of weeks gestation at birth was 38 weeks.

Prematurity at birth was present in 29.4% of the cases, of which 90% and 10% were at 36 and 35 weeks respectively. Likewise, 55.8% reached 38 or more weeks of gestation. Regarding maternal history, we found that the most common pathologies were preeclampsia (17.6%), urinary tract infection (14.7%), gestational hypothyroidism (8.82%), ischemic heart disease (2.94%), and genital condylomatosis (2.94%).

On the other hand, regarding prenatal and perinatal history, 79.4% of the cases had no pathological history, 17.6% presented low birth weight, of which 33% were diagnosed with intrauterine growth restriction, and 10% had miscarriage threat during the second trimester of gestation. Oligohydramnios was documented in 2.9%, without intrauterine growth restriction or low birth weight. As for prenatal care, 11.4% of the patients had no control, and therefore the diagnosis of heart disease was postnatal; however, 26.4% of the patients had no prenatal diagnosis, in spite of controls during the gestational period. Of the births, 64.7% were by cesarean section and the median weight of the patients was 3,161 grams (Table 1). The types of transposition found were distributed as shown in (Table 2). None of the patients had TGV exclusively. Therefore, the association with other types of malformations was described in (Table 2). Moderate to severe pulmonary hypertension was documented in 26.4% of the patients and in 17.6% of the cases there was evidence of decreased left ventricular function (LVEF) in the preoperative echocardiogram. These findings were consistent with the intraoperative findings in 76.4% of all patients.

Table 1: Demographic variables

Variable	Result
Gender	**% (n)**
Male	67.6 (23)
Female	32.3% (11)
Men:Women ratio	2.09:1
Previous Maternal History	**% (n)**
None	58.8 (21)
Pre-eclampsia	17.6 (6)
Urinary tract infection	14.7 (5)
Gestational hypothyroidism	8.82 (3)
Ischemic cardiopathy	2.94 (1)
Genital condyloma	2.94 (1)
Previous Familial History	**% (n)**
None	88.2 (30)
Diabetes mellitus	5.66 (2)
Hypertension	2.94 (1)
Congenital cardiopathy	2.94 (1)
Dilated cardiopathy	2.94 (1)
Prenatal and Perinatal History	**%**
None	79.4
Low birth weight	17.6
Oligohydramnios	2.9
Delivery method	**%**
Vaginal	35.3
C-section	64.7
Admission Variables	**Results**
Median age of admission (months)	2
Median maternal age (years)	23.8
Median parity (gestations)	1.9
Median number of gestational age (weeks)	38
Prematurity	29.4% (10)
35 weeks	10%
36 weeks	90%
38≥ weeks	55.80%

Table 2: Associated Cardiac Anomalies

Associated Cardiac Anomalies	%
TGV + Patent Ductus Arteriosus	2.9
TGV + ASD	2.9
TGV + ASD + Aortic Coarctation + Single Ventricle	2.9
TGV + Aortic coarctation + Situs Inversus	2.9
TGV + VSD + Ebstein's anomaly + Double outlet right ventricle	2.9
TGV + VSD + Pulmonary Atresia + Situs Inversus	2.9
TGV + ASD + VSD + Aortic Coarctation	2.9
TGV + ASD + VSD + Situs Inversus	2.9
TGV + Single Ventricle + Pulmonary Atresia	2.9
TGV + Aortic coarctation + Situs Inversus	5.8
TGV + VSD + Patent ductus arteriosus	5.8
TGV + Aortic coarctation + Single ventricle	8.8
TGV + ASD + VSD	8.8
TGV + ASD + VSD+ Pulmonary Atresia	11.7
TGV + ASD + Patent ductus arteriosus	32.3

TGA= Transposition of Great Vessels; ASD= Atrial septal defect; VSD= Ventricular septal defect

4.2. Preoperative Management

Regarding preoperative management, 94.1% of patients were admitted to the Neonatal Intensive Care Unit (NICU), where 38% of cases required ventilatory support, 17% support with inotropic therapy, and 14.7% vasopressor support. Only 1 patient was managed with concomitant vasopressor support and inotropic therapy. Prostaglandin infusion therapy was started in 85% of the patients, which was maintained until the surgical intervention was performed, 50% required diuretic therapy. In addition, 2.9% of the patients required atrioseptostomy as a vital emergency procedure.

4.3. Operative Management

Patients underwent surgery with a median of 11 days of life. A median of 14.5% of the patients received surgery without extracorporeal circulation, this given that the elected management was surgical procedures of a palliative type. The mean ischemia and perfusion time were 125 and 179 minutes respectively. The presence of reperfusion arrhythmias was reported in 47% of the cases, with an average bleeding volume of 111 cc and autotransfusion required in 47% of the patients. Regarding intraoperative evolution, 41% of the patients presented cardiogenic shock, of which 3 patients (21%) had pulmonary hypertensive crisis and 2 of these presented alveolar hemorrhage. Of the patients who presented cardiogenic shock, 71.4% died intraoperatively (Table 3).

Table 3: Type of surgical intervention

Type of surgical intervention	%
Jatene	70.5 (24)
Pulmonary artery cerclage	5.8 (2)
Glen	5.8 (2)
Contegra	5.8 (2)
Mustard	5.8 (2)
Damus Key Stansel	2.9 (1)
Systemic Pulmonary Fistula	2.9 (1)

4.4. Follow-Up

Regarding postoperative care, all patients were managed in the Neonatal Intensive Care Unit, with an average stay of 10.4 days. All patients required ventilatory support, with a median duration of 5 days (1 - 15 days). 96% of the patients required management with inotropic therapy and 83% with vasopressor support. Acute renal injury was present in 50% of the patients and LVEF compromise was documented in 50% of them. In addition, 16.6% presented some type of arrhythmia within the first 48 hours of the surgical procedure. Likewise, mortality at 30 days of follow-up was 31% and 7% at one year (Table 4).

Table 4: Postoperative outcomes

Variable	Result
Intensive care unit	100%
Intensive care unit (stay)	10.4 days
Inotropic therapy	96%
Vasopressor support	83%
Acute renal injury	50%
Left ventricular ejection fraction compromise	50%
Arrhythmia*	16.60%
Mortality - 30 days	31%
Mortality - 1 year	7%

*Within 48 hours

5. Discussion

TGV remains to be a pathology with elevated morbidity and mortality rates, literature regarding socio demographic, clinical and operative characteristics is scarce. In our environment, we do not have studies that describe sociodemographic variables, prenatal, gestational or maternal history. In Spain, the cohort described by Turon - Viñas [16] presented characteristics with some degree of similarity. A higher prevalence is described in men, and the most frequent type of malformation was D - Transposition, which is concordant with our results [16,17]. In terms of preoperative management, the use of prostaglandin E1 in our study was 85%, whereas in the Turon-Viñas cohort it was 88%. In terms of the requirement of atrioseptostomy, in our study only 2.9% patients required this intervention, unlike what was reported in previous cohorts, such as the study performed by Hraska [18], who reported 80% of patients who required atrioseptomy.

In the intraoperative aspects, the mean age of the intervention (11 days), the ischemia and perfusion time (125 and 179 minutes respectively) were similar to those reported by the Garcia-Hernandez series, who showed a mean intervention age of 11 days (8-16 days), with a mean ischemia time of 215 minutes and a perfusion time of 120 minutes [17], all the patients required vasopressor support during the intraoperative period and around half of the patients required concomitant use with inotropic which was maintained during the postoperative period with a mean time of 5 days for vasopressor and 7 days for inotropy similar to the ones reported by Garcia-Hernandez and Turón-Viñas [16,17].

Regarding intrahospital mortality, our study shows a higher ratio than the ones described in the literature [16,17], we consider this due to the socioeconomic context and pre-surgical clinical conditions of the patients such as low weight at birth, malnutrition, important gestational history such as intrauterine growth restriction

and prematurity, absence of prenatal controls in 11.4%, absence of prenatal diagnosis in 26.4% and type of transposition established as complex in 79.3% of patients. However, the follow-up at one year, of 7%, is similar to the reported with values ranging from 0.8 to 9% [16-22]. With the exception of Brazil with large series, there is no characterization of the population in Latin America. There is a deficit in the diagnosis and especially in the follow-up of these patients in terms of valve involvement, type of stenosis and need for reintervention, among others factors such as the great barriers of access to health and follow-up within a congenital heart disease program.

Regarding the limitations of our study, we include the retrospective nature of itself, and the small sample size. However, strengths include a 10-year follow-up, and to the best of our knowledge this is the first study reporting the sociodemographic, clinical characteristics and the management of great vessel transposition in Colombia and contribute to fulfilling the lack of information in the literature regarding this pathology in South-America.

6. Conclusion

Great vessel transposition remains to be a high mortality pathology, and requires complex and multidisciplinary management in order to offer an appropriate surgical or medical option to each patient. The characterization of patients with TGV is essential to specify and recognize the sociodemographic and clinical characteristics, this aimed at establishing associations within the natural course of this congenital heart disease and its clinical outcomes.

7. Acknowledgement

8. Funding Statement

9. Author Contribution

PLT, SV, DM, SNC, MPH, JRC, GR: Idea conception, manuscript writing, data recollection and analysis.

CERC, RENC: Manuscript writing, critical revision of the manuscript, data analysis, statistical analysis.

10. Data Avaliability

The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

11. Ethical Approval

Institutional approval by Pontificia Universidad Javeriana and Hospital Universitario San Ignacio was obtained prior to publication.

12. Statement of Conflict of Interest

There is no conflict of interest of any of the listed authors.

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