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Research Article

Transposition of Great Vessels: Latin-American Postoperative Experience

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Abstract

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.

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.

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%.

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.

Keywords: Congenital disease, Cardiac Surgery, Anatomic Repair, Physiological repair, Outcomes.

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.

Methods

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.

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.

Results

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).

Table1. 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)
Urinarytractinfection	14.7 (5)
Gestationalhypothyroidism	8.82 (3)
Ischemiccardiopathy	2.94 (1)
Genital condyloma	2.94 (1)
Previousfamilialhistory	**% (n)**
None	88.2 (30)
Diabetes mellitus	5.66 (2)
Hypertension	2.94 (1)
Congenitalcardiopathy	2.94 (1)
Dilatedcardiopathy	2.94 (1)
Prenatal and perinatal history	**%**
None	79.4
Low birthweight	17.6
Oligohydramnios	2.9
Deliverymethod	**%**
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	55.8%

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.

Table2. Associated Cardiac Anomalies

AssociatedCardiacAnomalies	**%**
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 + SitusInversus	2.9
TGV + ASD + VSD + Aortic Coarctation	2.9
TGV + ASD + VSD + SitusInversus	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	

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.

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)

Table3. Type of surgical intervention

Typeofsurgicalintervention	%
Jatene	70.5 (24)
Pulmonaryarterycerclage	5.8 (2)
Glen	5.8 (2)
Contegra	5.8 (2)
Mustard	5.8 (2)
Damus Key Stansel	2.9 (1)
SystemicPulmonary Fistula	2.9 (1)

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)

Table4. Postoperativeoutcomes

Variable	Result
Intensive care unit	**100%**
Intensive care unit (stay)	10.4 days
Inotropictherapy	96%
Vasopressorsupport	83%
Acute renal injury	50%
Left ventricular ejection fraction compromise	50%
Arrhythmia*	16.6%
Mortality - 30 days	31%
Mortality - 1 year	7%
***Within 48 hours**	

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.

Conclusions

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.

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To our patients

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.

References

[1] Transposition of the great arteries | Orphanet Journal of Rare Diseases | Full Text [Internet]. [cited 2022 Sep 18]. Availablefrom: https://ojrd.biomedcentral.com/articles/10.1186/1750-1172-3-2

[2] Boy:girl ratio in children born with different forms of cardiac malformation: a population-based study - PubMed [Internet]. [cited 2022 Sep 18]. Available from: https://pubmed.ncbi.nlm.nih. gov/7997413.

[3] Bianca S, Ettore G. Sex ratio imbalance in transposition of the great arteries and possible agricultural environmental risk factors. ImagesPaediatrCardiol. 2001 Jul; 3(3):10–4.

[4] Cohen MS, Eidem BW, Cetta F, Fogel MA, Frommelt PC, Ganame J, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. J Am Soc Echocardiogr Off Publ Am Soc Echocardiogr. 2016 Jul; 29(7):571–621.

[5] Hogan WJ, Moon-Grady AJ, Zhao Y, Cresalia NM, Nawaytou H, Quezada E, et al. Fetal Cerebral Vascular Response to Maternal Hyperoxia in Congenital Heart Disease: Effects of Cardiac Physiology. Ultrasound Obstet Gynecol Off J Int Soc Ultrasound Obstet Gynecol. 2021 May;57(5):769–75.

[6] Sarris GE, Balmer C, Bonou P, Comas JV, da Cruz E, Chiara LD, et al. Clinical guidelines for the management of patients with transposition of the great arteries with intact ventricular septum. Eur J Cardio-Thorac Surg Off J Eur Assoc Cardio-Thorac Surg. 2017 Jan;51(1):e1–32.

[7] Williams WG, McCrindle BW, Ashburn DA, Jonas RA, Mavroudis C, Blackstone EH, et al. Outcomes of 829 neonates with complete transposition of the great arteries 12-17 years after repair. Eur J Cardio-Thorac Surg Off J Eur Assoc Cardio-Thorac Surg. 2003 Jul;24(1):1–9; discussion 9-10.

[8] Martins P, Castela E. Transposition of the great arteries. Orphanet J Rare Dis. 2008 Oct 13;3:27. doi: 10.1186/1750-1172-3-27. PMID: 18851735; PMCID: PMC2577629.

[9] Samánek M: Boy girl ratio in children born with different forms of cardiac malformation: a population-based study. Pediatr Cardiol 1994, 15:53-7

[10] Sampayo F, Pinto FF: The sex distribution of congenital cardiopathies. Acta Med Port 1994, 7:413.

[11] Güçer S, Ince T, Kale G, Akçören Z, Ozkutlu S, Talim B , Cağlar M : Noncardiac malformations in congenital heart disease: a ret- rospective analysis of 305 Pediatric autopsies. Turk J Pediatr 2005, 47:159-66.

[12] Alfarhan A, Alquayt M, Alshalhoub M, Alnahdi MA, Masuadi E, Alhabshan F. Risk factors for transposition of the great arteries in the Saudi population. SaudiMed J. 2020 Oct; 41(10):1054-1062. doi: 10.15 537 /smj.2020.10.25418. PMID: 33026 04 5; PMCID: PMC7841512.

[13] Unolt, M., Putotto, C., Silvestri, L. M., Marino, D., Scarabotti, A., Valerio Massaccesi, Caiaro, A., Versacci, P., & Marino, B. (2013). Transposition of great arteries: new insights into pathogenesis. Frontiers in pediatrics, 1, 11.

[14] Goor DA, Edwards JP. The spectrum of transposition of the great arteries: with special reference to developmental anatomy of the conus. Circulation (1973) 48:406–1510.

[15] De la Cruz MV, Arteaga M, Espino-Vela J, Quero-Jiménez M, Anderson RH, Diaz GF. Complete transposition of the great arteries: types and morphogenesis of ventriculoarterial discordance. Am Heart J (1981) 102:271 -- 8110.1016/S0002-8703 (81)80018-X.

[16] Turon-Viñas A, Riverola-de Veciana A, Moreno-Hernando J, Bartrons-Casas J, Prada-Martínez FH, Mayol-Gómez J, Caffarena-Calvar JM. Characteristics and outcomes of transposition of great arteries in the neonatal period. Rev Esp Cardiol (Engl Ed). 2014 Feb;67(2):114-9. doi: 10.1016/j.rec.2013.06.017. Epub 2013 Nov 23. PMID: 24795118.

[17] J.A. García-Hernández, C. Montero-Valladares, A.I. Martínez-López, M. Gil-Fournier, J.M. Praena-Fernández, J. Cano-Franco, *et al*. Valoración pronóstica del *switch* arterial en la transposición de grandes arterias.AnPediatr (Barc), 74 (2011), pp. 174-181.

[18] Hraska V, Podnar T, Kunovsky P, Kovacikova L, Kaldarárová M, Horvathova E, Masura J, Mayer JE Jr. Is a learning curve for arterial switch operation in small countries still acceptable? Model for cooperation in Europe. Eur J Cardiothorac Surg. 2003 Sep;24(3):352-7. doi:10.1016/s 1010-7940(03)00439-1. PMID:129653 04.

[19] Duncan BW, Poirier NC, Mee RB, Drummond-Webb JJ, Qureshi A, Mesia CI, Graney JA, Malek CL, Latson LA. Selective timing for the arterial switch operation. Ann Thorac Surg. 2004 May;77(5):1691-6; discussion 1697. doi: 10.1016/ j.athoracsur.2003.10.042.PMID:15111168.

[20] Dibardino DJ, Allison AE, Vaughn WK, McKenzie ED, Fraser CD Jr. Current expectations for newborns undergoing the arterial switch operation. Ann Surg. 2004 May;239(5):588-96; discussion 596-8. doi: 10.1097/01.sla.0000124293.52814.a7. PMID: 15082962; PMCID: PMC1356266.

[21] Prêtre R, Tamisier D, Bonhoeffer P, Mauriat P, Pouard P, Sidi D, Vouhé P. Results of the arterial switch operation in neonates with transposed great arteries. Lancet. 2001 Jun 9;357 (9271):1826-30. doi: 10.1016/S014 0-6736(00)04957-6. PMID: 11410190.

[22] Jaggers JJ, Cameron DE, Herlong JR, Ungerleider RM. Congenital Heart Surgery Nomenclature and Database Project: transposition of the great arteries. Ann Thorac Surg. 2000 Apr;69(4 Suppl):S205-35. doi: 10.1016/s0003-4975(99)01282-5. PMID: 10798431.

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