



TRANSPPOSITION OF THE GREAT VESSELS: A MAPPING OF BRAZILIAN LITERATURE

TRANSPPOSIÇÃO DOS GRANDES VASOS: UM MAPEAMENTO DA LITERATURA BRASILEIRA TRANSPPOSIÓN DE LOS GRANDES VASOS: UN MAPEO DA LITERATURA BRASILEÑA

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ABSTRACT

Objective: to map the Brazilian scientific literature on the transposition of the great vessels (TGV). **Method:** a descriptive study, based on a non-systematic review of publications on TGV in Brazil between 2004 and 2014 included in BVSsalud, Google Scholar and Scielo library. **Results:** it was included 28 articles (cross-sectional, reviews, case report and prospective and retrospective study) conducted in SP, DF, MG, RS, SC, CE and AL. In Brazil, the prevalence of TGV ranged between 2.6% and 11.8% and the incidence between 1% and 82.2%. About 47% of TGV diagnoses were performed during prenatal care and 30% after birth. **Conclusion:** the number and quality of studies on TGV are deficient given the prevalence of this disease among fetal malformations. There were no national baseline studies. It is necessary to study the TGV's real impact, prognosis, and the necessary organizational conditions (e.g., composition and integration teams) for effective assistance. **Descriptors:** Transposition of Great Vessels; Prenatal Care; Prognosis; Mortality; Brazil.

RESUMO

Objetivo: mapear a literatura científica brasileira sobre a transposição dos grandes vasos (TGV). **Método:** estudo descritivo, pautado numa revisão não sistemática das publicações sobre TGV no Brasil entre 2004 e 2014. Pesquisou-se as produções nas bases eletrônicas BVSsalud, Google Acadêmico e biblioteca Scielo. **Resultados:** incluem-se 28 artigos dos tipos transversal, revisão, relato de caso, estudo prospectivo e estudo retrospectivo conduzidos em SP, DF, MG, RS, SC, CE e AL. No Brasil, a prevalência de TGV variou entre 2,6% e 11,8% e a incidência entre 1% e 82,2%. Cerca de 47% dos diagnósticos de TGV foram realizados no pré-natal e 30%, tardiamente. **Conclusão:** o número e a qualidade dos estudos sobre TGV são deficientes, dada a prevalência dessa cardiopatia dentre as malformações fetais. Não se encontraram estudos de base nacional. Faz-se necessário estudar a real incidência e o prognóstico do portador de TGV, bem como as condições organizacionais necessárias (ex.: composição e integração de equipes) para uma efetiva assistência. **Descritores:** Transposição dos Grandes Vasos; Cuidado Pré-Natal; Prognóstico; Mortalidade; Brasil.

RESUMEN

Objetivo: mapear la literatura científica brasileña en la transposición de los grandes vasos (TGV). **Método:** estudio descriptivo, basado en una revisión no sistemática de publicaciones en TGV en Brasil entre 2004 y 2014 en las bases de datos electrónicas BVSALUD, Scielo y Google Académico. **Resultados:** se incluyen 28 artículos del tipo transversal, revisión, relato de caso y estudio prospectivo y retrospectivo desarrollados en SP, DF, MG, RS, SC, CE y AL. En Brasil, la prevalencia de TGV varió de 2,6% a 11,8% y la incidencia de 1% a 82,2%. Entre los diagnósticos, cerca del 47% se realizaron prenatal y el 30% en el púes parto. **Conclusión:** el número y la calidad de los estudios en TGV son deficientes debido a la prevalencia de esta enfermedad entre las malformaciones fetales. No hubo ningún estudio de línea de base nacional. Es necesario estudiar el impacto real y el pronóstico de portadores de TGV y las condiciones organizativas necesarias (e.g., composición e integración de equipos) para una asistencia efectiva. **Descritores:** Transposición De Los Grandes Vasos; Atención Prenatal; Pronóstico; Mortalidad; Brasil.

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INTRODUCTION

Congenital heart defects are the most common birth defects whose incidence ranges from 4: 1,000 and 50: 1,000 live births globally.¹ In Latin America, the average prevalence is 4.8: 1000 live births and in Brazil varies between 5.5 1000, and 13.2: 1000.² Brazil Ministry of Health's mortality information system showed in 2010, congenital anomalies as the second cause of death (11.2%) in children under one year in Brazil, of these, cardiovascular defects accounted for 39.4% of all child deaths malformation.³

Congenital heart defects are abnormalities resulting from anatomical defects in the heart or circulatory system, compromising its structure and / or function. These diseases are present at birth, and even if detected late result, in most cases, in an inadequate embryological development of a given structure or the failure of such structure in obtaining their full development, from early stage of tissue fetal.⁴

Approximately 30% of live births with congenital heart disease receive hospital discharge without diagnosis, and progress to shock, hypoxia or early death before receiving proper treatment^{5,6}. Most neonatal units discharge neonate between 36 and 48 hours of life. At this stage, cardiac auscultation can be apparently normal and the clinical manifestation of critical diseases may have not yet occurred. Early diagnosis is extremely important, since it can prevent shock, acidosis, cardiac arrest or neurological injury before cardiopathy treatment inception.⁷

Transposition of the great vessels (TGV) represents 5% to 7% of congenital heart diseases and affects mainly males (60% to 70%). The TGV is a structural abnormality of the heart characterized anatomically by atrioventricular concordance and ventricular arterial discordance.² It is manifested mainly by cyanosis, and the prognosis depends on shunt existence, the degree of tissue hypoxia, and on left ventricular ability to maintain systemic pressure.⁸

Despite the presence of shunts, most patients with TGV without correction die

in the first few months of life. Aiming to decrease mortality rates, advances in echocardiography have allowed more early diagnosis of congenital heart disease. Early diagnosis during prenatal reduces mortality rates, as it allows proper delivery planning and, if necessary, enables rapid transfer to pediatric intensive care unit. These precautions reduce risk factors including profound hypoxemia, which can influence the child future neurodevelopment, reducing morbi-mortality.⁹

In the treatment of TGV, atrial surgeries were the first to yield satisfactory results. However, nowadays the anatomic correction or Jatene's surgery in the neonatal period is the preferred surgical technique to treat this abnormality. This surgery aims to reverse the vessels, causing the left ventricle (LV) stay in communication with the aorta and the right ventricle (RV) with the pulmonary trunk (PT).⁹ Such intervention must be undertaken before the 15th day of life, but this period may be extended with relative safety until the end of the first month of life.¹⁰

The mortality of patients with TGV can be reduced by timely diagnosis and treatment. Therefore, to know its prevalence and main associated factors aid to prevent, treat and improve quality of life of those affected, especially when it is estimated increase in prevalence, as is the case of Brazil.

OBJETICVE

♦ To map the Brazilian scientific literature on Transposition of the great vessels (TGV).

METHOD

A non-systematic review of the Brazilian scientific literature regarding the transposition of the great vessels (TGV) was accomplished in May 2014 in various data bases including BVSalud, Scielo and Google Scholar, and request to paediatric experts. The following DeCS descriptors were used to search the databases: transposition of the great vessels, prenatal care, prognosis, mortality, Brazil, early diagnosis and children, in Portuguese, English, and Spanish. The

articles reviewed were selected using the following inclusion criteria: articles conducted with Brazilians and published in Brazil or abroad whose focus was transposition of the great vessels; the following exclusion criteria were used: articles that focus congenital malformations different of TGV; literature that do not reflect Brazilian studies, and paid articles. The following filters were used: full-text availability; new-borns; Portuguese; year of publication from 2010. There were no peer selection of independent researchers and methodological quality assessment of included articles in this review.

RESULTS AND DISCUSSION

Twenty-eight studies conducted in SP, DF, MG, RS, SC, CE and AL, a book⁸ and technical document of the Ministry of Education²⁷ were included. The methodological designs included prospective and retrospective study, case report, cross-sectional study and review article, published between 2004-2013 (Table 1). Results were categorized as follows: incidence and prevalence, diagnosis, complications and mortality, treatment, prognosis and services.

| Author / Journal /Year | Objective | Type of study/ Number of participants | Results |
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| Rivera IR1 et al./Arquivos Brasileiros de Cardiologia/2007. | Analyze the importance of symptoms as a reason for referral to pediatric cardiologists in the diagnosis of congenital heart disease (CHD) in newborns (NB). | Cross-sectional study/3,716 live births in the maternity ward of the UFAL's Hospital between August 1999 and July 2002. | It was detected 49 cases of congenital heart disease (CHD) and 128 of patent ductus arteriosus (PDA). The prevalence of CHD was 13.2: 1000 NB. The main reason for referral to the cardiologist was blowing in 256 (72%) NB, of which 39 (15%) had CHD and 91% of the 128 cases, PCA. 14 (4%) NB, the reason for referral was cyanosis, 8 (57%) of whom had CHD. Heart failure was the reason for referral in 37 (10%) NB, 17 (46%) of whom carried CHD. Arrhythmia, associated congenital malformations or chromosome disorders were the reasons in 14% of cases. |
| Oliveira RP2 et al./Arquivos Brasileiros de Cardiologia/2008. | Case report with clinical presentation of corrected transposition of the great arteries in the fifth decade of life, with tricuspid insufficiency, occasion that the patient underwent a valve replacement. | Case report/A female patient, 47 years old, diagnosed with congenitally corrected transposition of the great arteries (CCTGA) done at 18 years of age. | CCTGA, a rare congenital heart disease, is associated with higher incidence of cardiac complications. |
| Leite DL3 et al./Arquivos Brasileiros de Cardiologia/2010. | To determine the prevalence and characteristics of congenital cardiac malformations in pediatric necropsies performed at the Hospital Regional da Asa Sul, Brasilia, DF, from January 1996 to December 2007. | Cross-sectional study/People who passed away in the Hospital Regional da Asa Sul from January 1996 to December 2007. | It was noted cardiac anomalies in neonatal deaths (117 / 61.9%), stillbirths (35 / 18.5%), infants (30 / 15.9%) and preschool (7 / 3.7%). There were no high school or college students. The main changes detected were: atrial septal defect in 96 patients (27%); interventricular communication in 66 (18.5%) and patent ductus arteriosus in 51 (14.3%), with no gender predominance. In 133 patients (70.4%), heart |

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| | | | disease were multiple and in 96 (50.8%) were associated with anomalies of other organs and systems; among these, 45 (23.8%) had heart disease as syndrome components, highlighting the high prevalence of chromosomal diseases, especially trisomies in all age groups. |
| Jansen D4 et al. / Revista SOCERJ/2000. | Create a script before, during and after surgery to contribute to the systematization of nursing care in Cardiopediatrics; Conduct a survey of major surgeries that took place in 1998. | Review article. | The script systematized nursing care guides the team to the prevention and early diagnosis of complications, favoring the early recovery of the child and, consequently, reducing the length of stay in the hospital. |
| Kuehl KS5 et al. / Pediatrics/1999. | Identify the misdiagnosis of predictors of congenital heart disease in newborns. | Retrospective study /4390 children. | Children's characteristics (weight, gestational age, intrauterine growth retardation, and chromosomal abnormality) are associated with the death of newborns with congenital cardiovascular malformations and death of these children before diagnosis. The diagnosis of coarctation of the aorta, Ebstein's anomaly, atrial septal defect, and truncus arteriosus are represented in babies found by community research, especially in children without malformations associated. |
| Mellander M6 et al. / Acta Paediatr/2006. | Determine the proportion of NB with critical heart defects not diagnosed at the hospital. | Retrospective study / Children who were born between 1993-2001 submitted to surgery catheter before 2 months of age for critical heart defects. | 259 term infants had critical heart defects. Systemic circulation duct dependent was present in 129, the pulmonary circulation dependent on duct in 106, and 24 children had no circulation duct dependent. In 51 children (20%) were not suspected any heart defects before hospital discharge. |
| Abu-Harb M7 et al. / Archives of Disease in Childhood/1994. | Identify patients with congenital heart disease in children to evaluate the treatment of those who die before the diagnosis. | Retrospective study /1074 children. | Of 1,074 children diagnosed in infancy, 185 of them died whereas 56 (30%) were undiagnosed. Serious noncardiac malformations were present in 29 of the 56. Cardiovascular anomalies were grouped into groups: complex in 13 out of 27 and in 14 out of 27 significant. |
| Schoen FJ8. Jatene MB9 et al. / Arquivos Brasileiros de Cardiologia/2008. | Cardiology book Study and analyze the prevalence of stenosis, as well as describe the surgical treatment and propose technical maneuvers to prevent its onset. | Don't apply Cross-sectional study / 553 children with transposition of the great arteries undergoing surgery type Jatene between April 1975 to December 2000 at Hospital da Real e Benemérita Sociedade Portuguesa de | Don't apply There was good progress in 20 patients, with death in one case. |

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| | | Beneficência, in Instituto Dante Pazzanese de Cardiologia of Hospital do Coração da Associação do Sanatório Sírio and at Instituto do Coração da Faculdade de Medicina da Universidade de São Paulo. | |
| Filho BG10 et al. / Arquivos Brasileiros de Cardiologia/2005. | To assess the surgical outcome in patients with transposition of the great arteries with intact ventricular septum undergoing surgery after the neonatal period. | Prospective study / 121 children with transposition of the great arteries with intact ventricular septum seen between January 1998 and March 2004. | In the group undergoing primary anatomic correction, it happened one (8.3%) hospital death from sepsis. In the correction group in two stages, five patients underwent slow preparation with correction 3-6 months after the 1st stage, and four deaths occurred after this stage. There was a change in the protocol adoption of rapid preparation technique in seven patients, all having reached the 2nd stage. Of the eight children who underwent the 2nd stage there was one death and one late death. The evolution late clinical presentation of children in both groups was favorable. |
| Costa MAT11 et al. Revista Brasileira de Ecocardiografia/ 2006. | Analysis of prenatal detection of congenital heart disease with fetal echocardiography, the risk factors and intrauterine and neonatal outcome. | Retrospective study / 1,546 fetuses with mean gestational age of 28 weeks. | They were detected 101 cases of congenital heart disease in 1,546 fetuses studied (6.5%). The most frequent malformations were atrioventricular septal defect (21.7%), hypoplastic left heart (15.8%), double outlet right ventricle (9.9%) and tricuspid valve abnormalities (5.9%). In 46.5% of cases, there was no evidence of a risk factor for congenital heart disease; 19 fetuses were intrauterine death (18.8%) and 33 in the neonatal period (32.6%). |
| Aragão JÁ 12 et al. / Revista Brasileira de Ciências da Saúde/ 2013. | To describe the epidemiological profile of patients with congenital heart disease undergoing heart surgery in hospital. | Cross-sectional study/Patients under 18 years of age, patients with congenital heart defects, operated between August 1, 2005 and July 31, 2010. | Of the 300 patients with congenital heart disease surgery, 209 (69.7%) were acyanotic type, and between these the most frequent was the ventricular septal defect (VSD) with 63 (21%). Among cyanotic, tetralogy of Fallot was 42 (14%). Cyanotic patients died in 24 (82.7%) cases, while acyanotic in five (17.3%). |
| Amorim LFP13 et al. / Jornal de Pediatria/2008. | To estimate the prevalence and clinical features and factors associated with congenital heart disease diagnosed at birth between August 1990 and December 2003, at Maternidade do Hospital das Clínicas de Minas Gerais. | Retrospective study / Live births (LBs) and stillbirths with congenital heart diseases diagnosed by postnatal echocardiogram or necropsy. | There were 29,770 births (28,915 LBs and 855 stillbirths). Among the LBs, the prevalence of heart diseases was 9.58: 1.000 (277 / 28,915), and among stillbirths to 87.72: 1.000 (75/855). The isolated heart disease occurred in 37.2% of cases between LBs and 18.7% between |

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| | | | stillbirths; associated with anomalies of other organs and systems without a syndromic diagnosis in 31.4% between LBs and 48.0% between stillbirths; It was part of syndromes in 23.1% of LBs and 32.0% of stillbirths. Multivariate analysis showed an association between heart disease and birth weight 2500 g in all types of clinical presentation between maternal age 35 years and heart disease syndromes and among females in isolated heart disease. |
| Hagemann LL14 et al./ Arquivos Brasileiros de Cardiologia/2004. | To study the morphological and functional abnormalities of the fetal cardiovascular system detectable by ultrasound. | Cross-sectional study/3,980 fetuses of pregnant women in the city of Porto Alegre. C) There were 103 diagnoses of fetal cardiovascular abnormalities, corresponding to 103 out of 3,980 (2.5%) of the study population, or 25.8 out of 1000, of which 47 were related to morpho-functional alterations of the fetal heart, overall prevalence of 11, 8 out of 1000 (47 / 3,980), and 56 showed changes in Refrangibility (golf ball). Three false-negative and no false positive results increased the overall prevalence of 12.5 out of 1000 (50 / 3,980), or 26.6 out of 1000 (106 / 3,980), while also including Refrangibility changes. | |
| Bastos LF15 et al./ Revista de enfermagem UFPE online/ 2013. | Identify the clinical and epidemiological profile of children with congenital heart disease undergoing cardiac surgery. | Prospective study / Children with congenital heart disease hospitalized in the pediatric postoperative unit of Hospital Dr. Carlos Alberto Gomes Studart. | Most were female (55.1%), the infant life (44.94%) and previous diagnosis and no treatment (47.2%). The most present congenital heart defect was interatrial communication with 100 cases (56.2%), followed by VSD 40.4%. |
| Diretrizes16/Arquivos Brasileiros de Cardiologia/2009 | Guide clinically surgical practice in fetal cardiac, pediatric cardiology and congenital heart diseases in adults. | Clinical-surgical Guideline | |
| Richmond S17 et al./Arch Dis Child Fetal Neonatal/2002. | To evaluate the effect of routine measurements of post-ductal oxygen saturation as an adjunct to routine clinical examination of the newborn. | Prospective study /6,166 babies in a general hospital in the district. | Measurements were made at 98% of eligible babies. SO2 less than 95% was found in 5% but persisted in only 1%. Structural heart defects were found in 50 (8.1 / 1000), 26 of which with isolated ventricular septal defects. Of the 24 remaining with other heart defects, six showed low SO2 and four showed for the first time (for other |

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| | | | reasons), low SO ₂ . SO ₂ also occurred for the first time in 13 other sick babies for other reasons. |
| Koppel RI18, et al. / Pediatrics/ 2003. | Determine the sensitivity, specificity, predictive value, and accuracy of oximetry screening program pulse in asymptomatic newborns with critical congenital cardiovascular malformation (CCCM). | Study of clinical epidemiology /11282 Newborns asymptomatic two hospitals. | 11281 oximetry held in asymptomatic infants with detection of three cases of critical congenital cardiovascular malformation. Nine births of babies with CCCM occurred during the study interval in a group of 15 fetuses with CCCM detected by fetal echocardiography. Six children with CCCM were symptomatic before the screening. There was a false positive case. Two children with false negative were readmitted. Other non-urgent diagnoses were obtained, including cases of patent foramen ovale, peripheral pulmonary stenosis and ventricular septal defect. The prevalence of CCCM among all live births was 1 in 564 and between the protected population was 1 in 2256 (sensitivity: 60%; specificity: 99.95%; positive predictive value: 75%; negative predictive value: 99, 98%; precision: 99.97%). |
| Wahl AG19 et al. / ActaPaediatr/2005. | Assess the feasibility of detection of congenital heart disease by pulse oximetry in dependent conduct cases before discharge. | Case-control study /200 normal newborns and 66 children with critical congenital heart disease. | Normal infants showed an average of saturation postductal 99 %. A group showed significantly greater proportion of post-ductal saturation values in normal infants compared with each other. |
| Meberg A20, et al. / Pediatr. / 2008 | Evaluate the effectiveness of pulse oximetry on the first day of life to detect congenital heart defects. | Multicentre prospective study /50,008 / 57,959 or 86% of infants born alive. | Of screened infants, 324 (0.6%) failed the test. Of these, 43 (13%) had congenital heart disease (27 in a critical condition), and 134 (41%) had pulmonary diseases and other disorders. The remaining 147 children (45%) were healthy with transition movement. To identify critical congenital heart defects, pulse oximetry had sensitivity of 77.1% (95% Interval of confidence (IC), 59.4 to 89.0), specificity rate of 99.4% (95% IC, 99.3 to 99, 5), and a rate of 0.6% false positives (95% IC, 0.5-0.7). |
| Ewer AK21, et al. /Lancet/2011. | To assess the accuracy of pulse oximetry as a screening test for congenital heart defects. | Prospective study /20,055 newborns at six maternity units in the UK. | Of the 20,055 screened newborns, 53 had congenital heart disease (24 in a critical condition) totalling prevalence of 2 to 6 per 1,000 live births. The sensitivity of pulse oximetry was 75% for critical cases and 49.06% for all major congenital |

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| | | | heart defects. In 35 cases, there was already suspected congenital heart disease after completion of prenatal ultrasound, so, with the exclusion of these cases, there was a reduction of test sensitivity to 58.33% for critical cases and 28.57% for all cases of major congenital heart defects. False-positive results were observed in 169 (0.8%) infants. |
| Mahle WT22, et al. /Pediatrics./2009. | Addressing the routine use of pulse oximetry in newborns to detect congenital heart disease critical. | Review article. | The evaluation performed by pulse oximetry after 24 hours of life has estimated sensitivity of critical congenital heart disease detection of 69.6%, positive predictive value 47.0%, with wide variation in sensitivity (between 0% and 100%). False-positive cases requiring further evaluation occurred in only 0.035% of infants screened after 24 hours. |
| Kemper AR23, et al. /Pediatrics/2011. | Develop strategies for the implementation of safe, effective and efficient screening. | Review article. | It was found enough evidence to recommend the beginning of the screening of low oxygen saturation in the blood through the use of monitoring pulse oximetry as congenital heart disease critical signal. Public health agencies have an important role in quality assurance and monitoring. |
| Zerbini EJ24, Revista Brasileira de Cirurgia Cardiovascular/ 2010. | - | Memory - Symposium / 1,000 patients undergoing cardiac surgery with extracorporeal circulation, and 680 at the Hospital da Faculdade de Medicina da Universidade de Sao Paulo and 320 in the Instituto de Cardiologia do Estado de São Paulo. | |
| Silva MEM25et al. / Revista Brasileira de Cirurgia Cardiovascular/2011. | Gather and present scientific evidence on the physiotherapist's role in pre-, during and postoperative period of cardiac surgery. | Review article. | It was evident the respiratory system involvement (specifically the bronchial secretion clearance and ventilatory adaptation) after surgery and the effectiveness of physical therapy will be shown to reduce rates of pneumonia, atelectasis, hospitalization, deleterious sequelae time restriction the bed, in addition to clinical improvement. There is a positive contribution of physical therapy intervention in the postoperative period of congenital corrections. |
| Lopes LM26 et al. /Arquivos Brasileiros de Cardiologia/2010. | Evaluate atrioseptostomy results with balloon catheter in cyanotic neonates. | Cross-sectional study/102 patients with cyanotic congenital heart disease in need of atrial shunt to ensure | Of the 102 cases of Rashkind performed, 98 met the inclusion criteria , divided in: 90 newborns at Group A (Rashkind |

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| | <p>survival to palliative or definitive surgical treatment. There was a predominance of males (75%), the mean age was 8.3 ± 9.3 days and the average weight was $3,100 \pm 1,100$ g.</p> | <p>procedure in the preoperative phase) and 8 at Group B (postoperative phase procedure). The transposition of the great arteries was the most frequent congenital heart disease ($n = 74$). Comparing the values of pre saturations and post-procedure ($65.9 \pm 19.5\%$ and $86 \pm 9.7\%$) and the diameter of the pre septal and post-procedure (2.3 ± 1.0 mm and 5.5 ± 1.3 mm) there was a statistically significant difference ($p < 0.001$). Comparing the values of saturation and the diameter of the ASD in groups of survivors and there was no statistically significant difference ($p > 0.05$).</p> |
| <p>Ministério da Educação27. Fundação Universidade Federal do Mato Grosso do Sul. Plano de Reestruturação Hospital Universitário Maria Aparecida Pedrossian. Mato Grosso do Sul: 2010.</p> | Government Document. | |
| <p>Caneo LF28 et al. / Rev Bras Cir Cardiovasc./2012.</p> | Review article | |

Figure: characteristics of included studies.

Legend: features not found or not reported in the article.

◆ Incidence and prevalence

The prevalence and incidence of congenital anomalies are difficult to measure because these are the main causes of miscarriages. Birth defects occurrence depends on many risk factors that can be of fetal or maternal origin. The main risk factors for congenital malformations are: maternal diabetes mellitus (type I or gestational); maternal age over 35 years; repeat abortions; maternal exposure to drugs or viral agents; family history of congenital heart disease; twin; fetal arrhythmia and extracardiac malformations.¹¹

In several articles selected, the prevalence of cardiac abnormalities in Brazil ranged from 2.6% to 11.8% among all malformations. Most studies show that among heart diseases, the acyanotic are more prevalent than cyanotic. However, in all of them, TGV presents among the most frequent of cyanotic.¹² Among the set of congenital heart disease, the incidence of TGV ranged from 1% to 82.2%, making an average of 20.47%. Several studies included reported the prevalence of heart disease and TGV, but only two considered statistically significant results ($p < 0.05$).^{13,14} In addition, only one study expresses the incidence of heart disease and TGV concomitantly.¹¹

◆ Diagnosis

The results of these studies show that 47.2% of TGV diagnostics are performed during prenatal care, 30.3% of diagnoses are lately reached, and the remaining 22.5%, cannot be said when it was diagnosed¹⁵ including diagnostic during necropsy studies.³

It is recommended that the diagnosis of TGV should be firmed as early as possible, preferably in the gestational phase by echocardiography with color flow mapping for fetal heart assessment, but its use as a screening tool is unfeasible. The intrauterine TGV cardiac diagnosis can guide health team and family planning related to fetus therapeutic, propedeutic and continuous care to be taken in the immediately postpartum.¹⁰

Echocardiography can be performed in the first trimester of pregnancy by transvaginal and transabdominal vias. However, only from the 18th week of gestation to term, that all the structural or functional abnormalities of the fetal heart can be seen. Early diagnosis in this period is crucial for the clinical and / or effective surgical treatment are subsequently instituted in a timely manner, thus changing the natural history of disease and sometimes allowing their total healing.⁶

About the diagnosis made in the postnatal period it is extremely important to track cardiac abnormalities in seemingly healthy

newborns with gestational age over 34 weeks using pulse oximetry measurement, 'Teste do coraçãozinho'. When this screening is routinely performed on all newborns, shows significant sensitivity and specificity for early detection of these diseases, that often require in most of the time intervention and immediate treatment.^{17,18,19,20,21,22,23} The hospitals where this test is not offered should recommend parents to seek healthy childcare services in primary healthcare units and if any signs or symptom of cardiovascular compromise is detected, immediate hospital support, where realization of further clinical examination, chest radiography, electrocardiography and echocardiography will subsidize diagnosis and possible surgery. At physical examination, attention should be given to heart murmur, cyanosis or signs of ventricular dysfunction.

Indications for laboratory tests in the diagnosis of heart diseases were found. The first is the suspicion of cardiac malformations in obstetrical ultrasound, which emphasizes the importance of training professionals who perform obstetrical ultrasound. The second indication refers to existing and above mentioned risk factors for these anomalies. After an appropriate indication, echocardiography should be performed to evaluate the atrial *situs*, systemic and pulmonary venous drainage, atrioventricular and ventricular-arterial connections, aortic arch, ductal arch and interatrial septa, atrioventricular and interventricular.^{11,15}

◆ Complications and mortality

The complications and mortality affect patients with and without surgery to treat TGV. In contrast, the rate of complications in patients undergoing surgery was 2%, while in non-operated patient, complication rate was 67%. Regarding mortality, patients undergoing surgery, the mortality rate ranged from 4 to 18%.²⁴ It is inconclusive whether the surgery improved the prognosis of patients because no study reported mortality rates in patients without surgical correction.

Complications vary according to the surgical technique used. In Rashkind's technique, performed in 98 patients, two patients presented clinical complications (i.e., atrial fibrillation followed by ventricular tachycardia and cardiac tamponation), and eight died in the postoperative period.²⁵ Jatene's technique did not cause clinical complications in the first postoperative month in 121 patients, but after this period, two patients had pulmonary complications and one patient was re-operated. A third surgery technic was reported by Blalock and Hanlon that was performed by 28 patients with TGV

who did not presented postoperative complications, but resulted in a 18% of death rate.²⁴

◆ Treatment

The most common causes of morbidity and mortality in cardiac surgery in all heart diseases in adults are pulmonary complications; the incidence varies between 6% and 76%. In the pediatric age this incidence remains undefined. It was found that the earlier happen the surgery, the lower the physical and psychological sequelae in children.²⁵

Before the diagnosis of cyanotic congenital heart disease, only contraindication to surgery in two eventualities: 1) surgical technique of absence that may benefit the patient (example: people with common arterial trunk, single ventricle with large pulmonary circulation and atresia aortic); 2) development or the stage of the disease to prevent surgical treatment as the disease with abnormal communication between arterial and venous industry at auricular, ventricular or pulmonary, with large pulmonary hypertension and "shunt" venoarterial pure or predominant.²⁴

Treatment of TGV depends on the clinical presentation and associated comorbidities. Therefore, it can be a medical treatment with drugs (example: angiotensin converting enzyme inhibitors, diuretics, digitalis) or palliative surgical treatment (physiological) or definitive (anatomical) with blood exchange.

The technique of choice for the correction of TGV with intact ventricular septum is the Jatene surgery, which should be performed until the 15th day of life and may extend this period until the end of the first month of life. After that date, the progressive decrease in pulmonary vascular resistance reconditions the performance of the left ventricle (LV) to a low-pressure circuit with progressive reduction of its muscular mass.¹⁰

Support physical therapy begins with the arrival of the child to the intensive care unit (ICU). The patient's positioning in bed ensures proper location of vascular access, drains and tracheal tube, known the risk of displacement of these during transport of the operating room. Physical therapy for these patients reduces the length of stay in the ICU of these aids in ambulation as early as possible and reduces hospital stay by improving oxygenation, preservation of favorable conditions of ventilation and maintenance of airway patency. There is also a trend towards improvement in expiratory tidal volume, lung

compliance and resistance, preventing complications.²⁵

◆ Prognosis

The mapping of scientific papers conducted in Brazil on TGV showed significant gap on the prognosis of these patients. Some articles report the need of diagnosis in the prenatal period or before the first month of life to ensure effective intervention, with less likelihood of complications, and correction of heart disease. This recommendation suggests that in patients diagnosed and operated on time seems to be better prognosis and longer life expectancy.

◆ Services

A major barrier for assistance to people with TGV is the insufficient availability of information coupled with inadequate infrastructure for the provision of services. Many hospitals do not hold adequate infrastructure, equipment and technology for complementary exams and treatment (surgical or ambulatory), satisfactory conditions for admission and ICU postoperative, and conditions and ideal surgical materials for surgery that long and hard.²⁷

This mapping also identifies important gap regarding the organizational arrangements of health services where people with TGV are assisted. No data found as tracking test (example: Teste do coraçõzinho), health team composition, remuneration schemes, the existence and degree of implementation of protocols or clinical and surgical guidelines, monitoring and evaluation of assistance, technology used, access, quality, length of stay, hospital infection rate, cost, and satisfaction of those involved in regard to the TGV.

Some papers categorize hospitals as referral centers for the care of cardiac patients, but not conceptualize such a definition. According to information obtained from a professional center reference in the care of patients with cardiac abnormalities - Biocor Hospital of Belo Horizonte, to receive such a designation the service must have a cardiovascular surgeon with experience in surgery for congenital heart defects, ICU postoperative clinic of pediatric cardiology, echocardiography, hemodynamics service and arrhythmology services.

◆ Health team

For the effective care of a patient with TGV is fundamental action of a multidisciplinary team of physicians, nurses, physiotherapists, nutritionists, psychologists and social workers. It should be noted that these professionals need to be adequately

capacitated to provide comprehensive care, humane and the most timely possible to these patients.

One of the analyzed studies shows that the technical quality of Brazilian surgeons is indisputable. Thus, it is notable that a number of Brazilian contributions helped in the development of pediatric surgery, among them there is the Jatene operation for correction of the TGV. However, satisfactory diagnostic and surgical results depend on an entire hospital system, technical and non-technical.²⁸

In literature mapping, no articles were found that addressed the multidisciplinary team needed to follow, more appropriately, the carrier TGV from the moment of his birth. Unfortunately, it is a particular concern in the national scene, since it is evident the importance of working together to minimize possible damage and consequences arising from the TGV, as death and impaired quality of life.

CONCLUSION

The number and quality of studies on TGV are insufficient when compared to the prevalence of this disease among fetal malformations. Virtually no studies conducted in Brazil report the composition of organizational conditions necessary for the satisfactory care of the patient, and resulting from this, no studies conducted include a sample of the entire national territory. Thus, it is necessary to increase the quality and quantity of studies regarding the actual incidence of TGV, health team composition, required organizational conditions and the TGV carrier prognosis.

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