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CASE REPORT

Case report: Corrected transposition of great arteries in joint housing routine

Bruno Dias Queiroz[1], Vinicius Dorighetto Ardisson[1], Sônia Maria Alves Andrade[2], Priascila de Castro Soares Barreto[2], Andrea Lube Antunes de S Thiago Pereira[1,2]

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Abstract

Introduction: We report a case of a newborn with Congenitally Corrected Transposition of the Great Arteries, congenital heart disease associated with pulmonary atresia, atrial septal defect, ventricular septal defect and situs inversus. Pacient successfully underwent modified Blalock-Taussig-Thomas surgery. **Objectives:** To describe the Corrected Transposition of the Great Arteries, reviewing the anatomy, elucidating the complementary confirmatory tests and the management of emergency care in the face of ductal-dependent congenital heart lesions. **Case report:** Full-term newborn, appropriate for gestational age, female, born by vaginal delivery without need for neonatal resuscitation. The mother received regular prenatal care, without complications. During the routine rooming-in visit, the patient had significant cyanosis, with no alteration in the physical examination that would explain the clinical picture. **Conclusion:** The importance of ultrasound during prenatal care is reinforced in the search for early diagnosis of congenital anomalies and the presence of a multidisciplinary team prepared to identify clinical signs indicative of critical cyanotic heart disease, allowing rapid and assertive care, which are fundamental for a favorable outcome for the newborn.

[1] Escola Superior de Ciências da Santa Casa de Misericórdia de Vitória, Medicina - Vitória - Espírito Santo - Brazil.
[2] Hospital Santa Casa de Misericórdia de Vitória - Unidade Pro-Matre, Neonatologia - Vitória - Espírito Santo - Brazil.

Correspondence to:
Bruno Dias Queiroz.
Escola Superior de Ciências da Santa Casa de Misericórdia de Vitória. Av. Nossa Sra. da Penha, 2190 - Santa Luíza, Vitória, ES, Brazil. CEP: 29045-402. E-mail: bruno@ciclo.com.br

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INTRODUCTION

Corrected transposition of the great arteries (CTGA) is a rare congenital heart defect characterized by double discordance (atrioventricular and ventriculoarterial), with a prevalence of approximately 0.5% to 1.4% of all congenital heart defects[1]. Among the malformations in newborns, cardiac defects have the highest mortality rate in the first year of life in Brazil, being the second leading cause of death within the first 30 days of life[2].

CASE REPORT

This article presents the case of a female full-term newborn, appropriate for gestational age, who was born via vaginal delivery, did not require resuscitation, and showed no apparent abnormalities during the initial physical examination. The mother received regular prenatal care without any complications.

On the first visit, held approximately ten hours after birth, while the newborn (NB) and the mother were in the rooming-in unit, significant central cyanosis was noted, which worsened when the newborn cried. Physical examination revealed no respiratory distress, and preductal pulse oximetry showed 20% saturation. The newborn was admitted to the Neonatal Intensive Care Unit (NICU), and even after the introduction of noninvasive ventilation with nasal Continuous Positive Airway Pressure (CPAP) and a fraction of inspired oxygen up to 100%, she remained hypoxic, raising suspicion for congenital heart disease. Intravenous prostaglandin infusion was initiated at 0.01 micrograms per kilogram per minute, improving saturation and reaching levels above 90%. On the same day, the newborn underwent a transthoracic echocardiogram that confirmed heart disease. The patient was then transferred to a referral center for treatment of pediatric congenital heart disease.

The newborn was hospitalized. One- and two-dimensional echocardiograms and pulsed- and continuous-wave Doppler tests were repeated. The patient was diagnosed with situs inversus with dextrocardia, atrioventricular and ventriculoarterial discordance, pulmonary valve atresia featuring a single aortic outlet connected to the right ventricle, wide misaligned interventricular communication (IVC), and patent ductus arteriosus. The heart's anatomy was consistent with L-TGA.

Due to pulmonary valve atresia, palliative systemic-pulmonary anastomosis surgery (modified Blalock-Taussig-Thomas shunt) was performed with satisfactory results. In the immediate postoperative period, the patient experienced increased bleeding, hypotension, and oliguria, which were resolved with packed red blood cell transfusions, volume replacement with 0.9% saline solution, and furosemide. She was extubated on the second postoperative day. After 24 days of hospitalization, with good progress and no new complications, the patient was discharged and referred to the cardiology outpatient clinic for ongoing medical monitoring.

DISCUSSION

Congenital heart disease can be classified by the presence or absence of cyanosis. Cyanosis is the leading indicator of cyanotic heart disease, which results from low levels of oxyhemoglobin in the arterial blood.

Among the heart diseases that cause cyanosis, Dextro-Transposition of the Great Arteries (D-TGA) is characterized by ventriculoarterial discordance (the aorta emerges from the right ventricle, and the pulmonary artery arises from the left ventricle), which places the systemic and pulmonary circulations in parallel rather than in series. Additionally, the presence of shunt sites between the two circulations (such as interatrial communication, VSD, and patent ductus arteriosus) is essential for maintaining life. The term "right-handed connotation" of D-TGA refers to the looping of the ventricles during cardiac morphogenesis, which positions the right ventricle to the right of the morphological left ventricle. This provides a concise nickname to help differentiate this form of transposition from Levo-Transposition of the Great Arteries (L-TGA)[3].

In the L-TGA anomaly shown in Figure 1 below, the morphologically right atrium connects to the morphologically left ventricle, which subsequently gives rise to the pulmonary artery. The morphologically left atrium connects to the morphologically right ventricle (RV), from which the aorta originates[4]. This results in a double discordance, meaning both atrioventricular and ventriculoarterial discordance form[5].

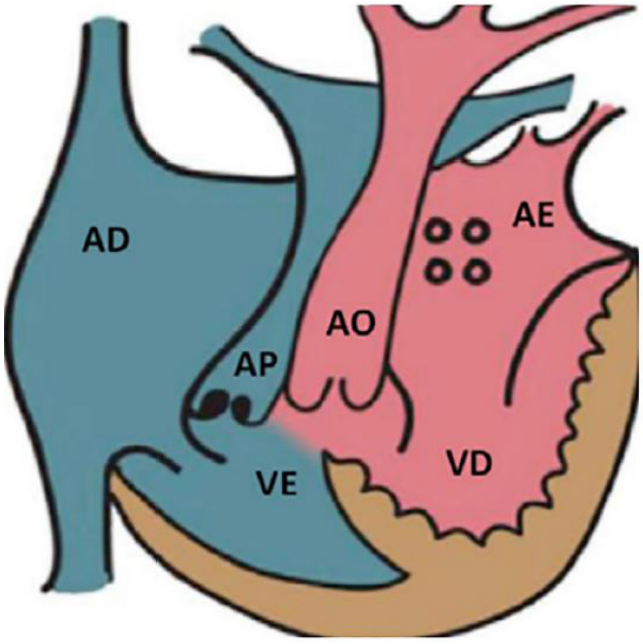


Figure 1. Schematic drawing of a heart with L-TGA. Source: Amaral (2023)[6].

Congenitally or physiologically corrected TGA refers to the standard physiological return of deoxygenated systemic venous blood to the heart and the transport of oxygenated pulmonary venous return to the systemic circulation through the discordant ventricles and large arteries[7]. However, this condition is not a "benign heart disease" without consequences. It is typically associated with several congenital defects, the most common of which, in order of occurrence, are VSD, pulmonary outflow obstruction, and tricuspid and mitral valve anomalies. There is often a high risk of progression to total atrioventricular block, systemic valve regurgitation, congestive heart failure, and right ventricular dysfunction. Since the right ventricle is connected to the aorta, it pumps blood against systemic pressure, a task it is not suited for.

In most patients with this rare congenital disease, if there are no other significant associated anomalies, cardiac symptoms might manifest in the fourth or fifth decade of life[8]. However, the pulmonary valve atresia in the patient described in the case causes high pressure in the right ventricle, creating a right-to-left shunt through the VSD. This results in low pulmonary flow and hypoxemia. Therefore, prostaglandin is essential to keep the ductus arteriosus patent, allowing the maintenance of a satisfactory minimum pulmonary flow until surgery is performed.

The modified Blalock-Taussig-Thomas procedure creates a shunt using a polytetrafluoroethylene tube that connects the subclavian artery to the ipsilateral pulmonary artery, facilitating pulmonary flow and promoting the development of the pulmonary arteries and arterial circulation. This procedure is a proven, reliable, and safe method for the palliative care of patients with heart diseases that cause low pulmonary flow[9]. Compared to its classic counterpart, a disadvantage of this procedure is that the graft does not grow as the child develops, as it involves anastomosing the patient's branch of the subclavian artery. Figure 2 below presents the results of the procedures described.

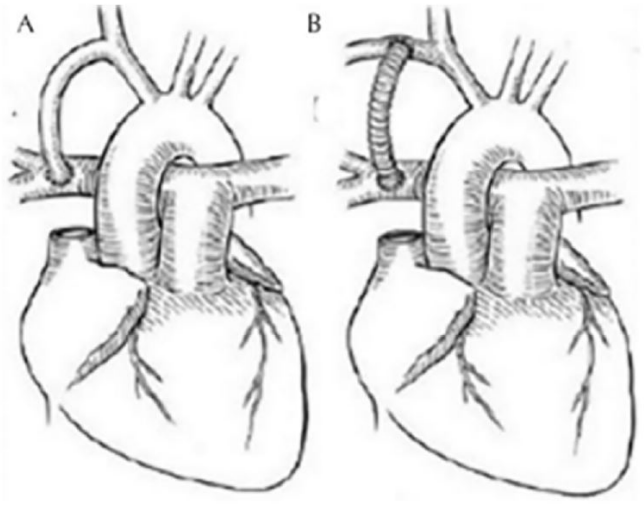


Figure 2. Classic (A) and modified (B) Blalock-Taussig-Thomas shunt. Source: López-Terrazas (2016)[10].

Few studies have described the prenatal diagnosis of CTGA. Establishing a diagnosis before birth is essential for parent counseling and improving interdisciplinary postnatal care[11]. The importance of prenatal care and fetal echocardiography in identifying, diagnosing, and promptly referring cases of congenital heart disease to specialized centers cannot be overstated. Patients without a prenatal diagnosis have a greater chance of preoperative complications, with increased morbidity and mortality[12].

It is essential to emphasize the significance of this report for the maternity care team responsible for newborn care, as it reaffirms necessary emergency procedures and the need for differential diagnosis in cases of central cyanosis in neonates. The necessity of performing proper bedside physical examination, which includes assessing airway permeability, chest expansion, signs of respiratory failure, cardiac auscultation, and measuring pre- and post-ductal oxygen saturation, must be emphasized. Definitive diagnosis is confirmed through echocardiographic evaluation. Clinical suspicion of congenital heart disease dependent on the ductus arteriosus arises when there is a lack of response to oxygen therapy, a critical factor in deciding to initiate prostaglandin infusion, an essential medication for managing hypoxemia in these patients.

BIBLIOGRAPHIC REFERENCES

1. Oliveira RP de, Agorianitis P, Vegni R, Nobre G, Kalichsztein M, Kezen J. Transposição corrigida das grandes artérias: apresentação clinica tardia, na quinta década de vida. Arq Bras Cardiol. 2008;91(4): e35-7
2. Soares AM. Mortality in Congenital Heart Disease in Brazil - What do we Know? Arq Bras Cardiol. 2020;115(6):1174-5
3. Fulton DR, Kane DA. D-transposition of the great arteries (D-TGA): Anatomy, clinical features, and diagnosis [Internet]. In: UpToDate. Alphen aan den Rijn: Wolters Kluwer; c2023 [citado em 1 Jan 2024]. Disponível em: https://www.uptodate.com/contents/d-transposition-of-the-great-arteries-d-tga-anatomy-clinical-features-and-diagnosis
4. Sousa PA, Cavaco D, Adragão P, Teixeira A, Ribeiras R, Martins M, Mendes M. Ressincronização cardíaca em doente com transposição congenitamente corrigida das grandes artérias. Rev Port Cardiol. 2014;33(6): 387.e1-387.e6
5. Warnes CA. Transposition of the great arteries. Circulation. 2006; 114:2699-709
6. Amaral F. O paciente com ventrículo direito sistêmico. Rev Soc Cardiol Estado de São Paulo. 2023;33(1):11-6
7. Kane DA. L-transposition of the great arteries: Anatomy, clinical features, and diagnosis. In: UpToDate. Alphen aan den Rijn: Wolters Kluwer; c2023 [citado em 1 jan 2024]. Disponível em: https://www.uptodate.com/contents/l-transposition-of-the-great-arteries-l-tga-anatomy-clinical-features-and-diagnosis?search=7.%09Kane%20DA.%20L-transposition%20of%20the%20great%20arteries:%20Anatomy,%20physiology,%20clinical%20features,%20and%20diagnosis.%20&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1
8. Sayuti KA, Azizi MYSB. Incidental congenitally corrected transposition of the great arteries (ccTGA) in an adult with suspected coronary artery disease: review on radiological features and pathophysiology. BMJ Case Rep. 2020;13(4):e234225.

9. Maluf MA, Andrade JCS, Carvalho A, Catani R, Veja H, Andrade JL, et al. Operação de Blalock-Taussig modificada para o tratamento paliativo de cardiopatias congênitas com hipofluxo pulmonar. Braz J Cardiovasc Surg. 1995;10(3):126-32
10. López-Terrazas JH, Colín-Ortiz JL, López-Corella E. Niño con cardiopatía congénita controlada y muerte inesperada. Acta Pediatr Mex. 2016;37(6):333-340
11. Krummholz A, Gottschalk I, Geipel A, et al. Prenatal diagnosis, associated findings and postnatal outcome in fetuses with congenitally corrected transposition of the great arteries. Arch Gynecol Obstet. 2021;303(6):1469-81
12. Binotto CN, Alves SC, Turra MLM, Malanche RM. Transposição de grandes vasos em criança de 1 ano de idade. Resid Pediatr [internet]. 2018[citado 1 jan 2024];8(1):41-4. Disponível em: https://residenciapediatrica.com.br/detalhes/308/transposicao-de-grandes-vasos-em-crianca-de-1-ano-de-idade