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CASE REPORT

Transposition of great arteries in 1-year-old child

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Abstract

Objective: To report the case of a pediatric female patient with late diagnosis of transposition of the great arteries associated with atrial septal defect and ventricular septal defect, 1 year old. And relate this story with literary basis emphasizing the clinical and imaging diagnosis, and treatment of temporary and permanent. **Case report:** Report the case of a girl with late diagnosis of transposition of the great arteries. **Conclusions:** Transposition of the great arteries is a major evolution of cyanotic heart disease if not treated early. When it comes associated with atrial and ventricular septal, the clinical picture may appear more bland and cyanosis not be as evident as in the reported case, which leads to late diagnosis. The arterial switch surgery generally good clinical outcome when well executed and provides an increased survival of the disease by 96%.

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INTRODUCTION

Transposition of the great arteries (TGA) is a form of congenital cyanotic heart malformation that is classically characterized by the location of the aorta anteriorly and laterally (to the right) of the pulmonary artery. In some patients, anatomical variations, such as the location of the aorta posteriorly, may occur. Shunts between the pulmonary and systemic circulation are necessary, and the interventricular septum is open in 40% of cases[1].

The incidence of TGA is 1 for every 3,000 live births, corresponding to 8% of all cases of congenital heart disease. The male: female ratio of TGA is 3:1. Because of the severity of the condition, this condition requires surgical correction in childhood, particularly in the neonatal period[2].

Classical or simple TGA occurs when the integrity of the interventricular septum is maintained, there is no obstruction of the left ventricle outlet, but a patent foramen ovale and a patent ductus arteriosus are common. By contrast, the complex form may be associated with interventricular communication (IVC), pulmonary stenosis (in 30% of cases), and coarctation of the aorta, which hinders the output flow from the left ventricle[3].

Hemodynamically, there is complete separation between pulmonary and systemic circulations, which occur in parallel. This separation is because venous blood returns to the body through the aorta from the right ventricle. Similarly, arterial blood returns to the lungs through the pulmonary artery from the left ventricle[4].

The interatrial communication (IAC) and IVC that are present in the fetal period allow for satisfactory blood mixing and allows fetal survival. However, after birth, such communications tend to close, leading to two separate and parallel circulations, which is incompatible with life. The severity of symptoms is related to the dimensions of the persisting communications: the smaller they are, the more severe the disease is. Cases in which the condition is associated with pulmonary, valvar, or infundibular stenosis, or even pulmonary hypertension present worse clinical outcome and prognosis[4].

In TGA, the aorta and coronary arteries originate from the right ventricle, and the pulmonary trunk originates from the left ventricle. The discovery of the anatomy of the coronary arteries in TGA by Jatene was decisive for the success of the surgical procedure he devised. The anomalous origin of the coronary arteries increases surgical complexity and risk, and there are several anatomical presentations (single coronary ostium and intramural coronary artery, among others) that usually make an anatomical correction impossible[4].

In the classic form of TGA, the main signs and symptoms observed in the newborn are cyanosis, which presents within the first hours of life, and a single loud S_2 in the pulmonary area on physical examination. When present, heart murmur has grade 1 intensity and is more easily audible in the pulmonary or tricuspid area.[3] In the complex form, cyanosis is mild because of blood mixing. In this case, the increase in pulmonary flow can lead to heart failure at the end of the first month of life[3].

TGA is associated with high mortality. Without surgical treatment, 28.7% of TGA patient's progress to death within the first week of life, 51.6% within the first month, and 89.3% do not survive the first year[3]. The surgical technique that allows for a definitive correction of TGA is the Jatene procedure, also known as arterial switch. The first patient to be successfully operated using this technique had IVC, which allowed the patient to survive long enough for the surgery to be performed later at the age of a few months[4,5].

The procedure should be performed early, in the first 2-3 weeks of life. Later procedures may cause a decrease in pulmonary pressure and an involution of the left ventricular mass, and there is an increased risk of the left ventricle not adapting to the postoperative systemic resistance[3-5].

CASE REPORT

Our patient, S.A.S., was aged 1 year and 21 days, was female, and was born in Mallet, state of Paraná, Brazil. She was referred from a hospital in the nearby city of Ponta Grossa, where she had been admitted for pneumonia and had been administered ceftriaxone for 8 days. Central cyanosis was diagnosed on admission, and the parents had not noticed this condition before admission. The results of echocardiogram indicated IAC and IVC. Nonetheless, because this condition did not explain the cyanosis, the test was repeated with suspicion of TGA.

Severe heart disease was suspected, and therefore, the child was referred for examination, diagnostic confirmation, and hemodynamic studies. The patient was treated with captopril, furosemide, spironolactone, and hydrochlorothiazide. On physical examination, the patient was in good general condition and was eupneic, cyanotic, and anicteric. Cardiovascular auscultation indicated two heart sounds with a loud S_2 at the superior left sternal border (pulmonary area), which was louder than that at the superior right sternal border (aortic area), a systolic murmur of ++/4+ at the inferior left sternal border and +/4+ at the superior left sternal border. Peripheral pulse was full and symmetric. Edema was present. Extremities were cyanotic, but digital clubbing was absent. Other organs were unremarkable.

ECG, cardiac catheterization, and another echocardiogram were performed at the Pequeno Príncipe Hospital. The diagnosis was confirmed as TGA, large IVC, small IAC, slight infundibular-valvar pulmonary stenosis, moderate pulmonary hypertension, and coronary arteries with no anomalies. The chosen treatment was total surgical correction.

Echocardiogram: Ostium secundum, IAC, 5.7 mm; IVC, 10 mm; ventriculoarterial discordance; mitral, 13 mm; tricuspid, 18 mm; maximum pulmonary pressure gradient, 25 mmHg; pulmonary ring, 11 mm; and aorta, 12 mm. Vessels in parallel, enlarged right ventricle, larger than the left ventricle. Conclusion: TGA + IAC + IVC (figure 1).

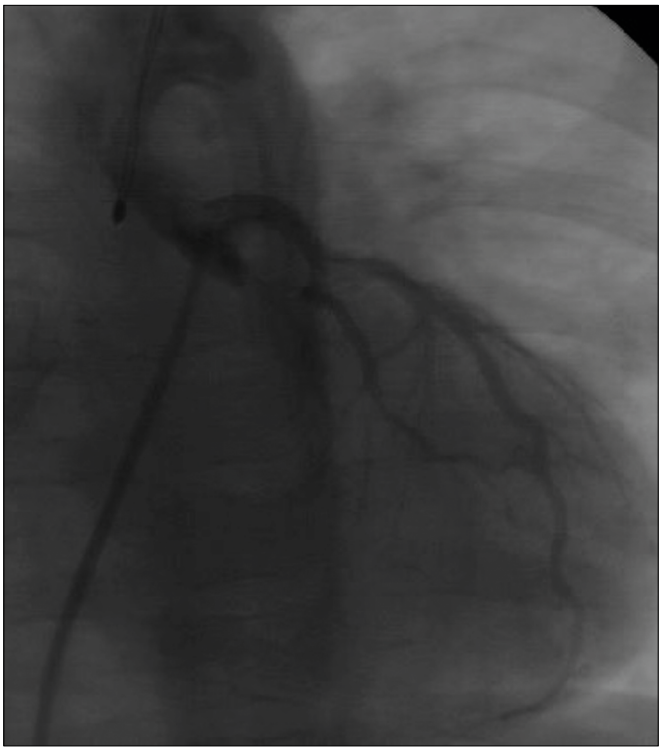


Figura 1. Echocardiogram.

Cardiac catheterization: TGA + large perimembranous IVC + moderate ostium secundum IAC + mild infundibular-valvar pulmonary stenosis. Coronary arteries were unremarkable. Balanced ventricles; pulmonary ring, 14.9 mm; moderate pulmonary hypertension; pulmonary vascular resistance, 1.78; pulmonary vascular resistance/systemic vascular resistance ratio, 0.22; right ventricular pressure, 62; left ventricular pressure, 60; pulmonary arterial pressure, 45 (figure 2).

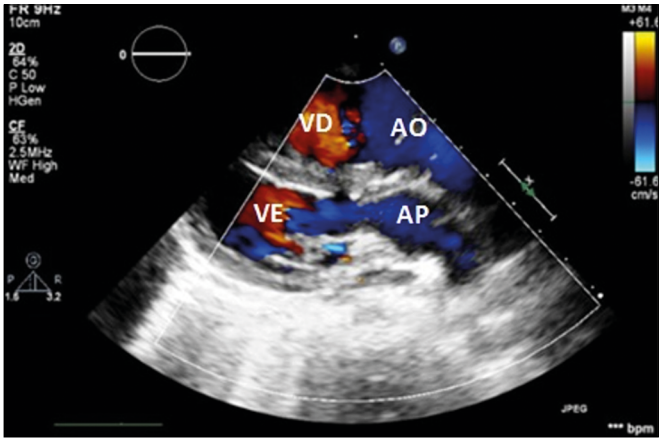


Figura 2. Cardiac catheterization.

The patient was admitted for surgical correction at the age of 13 months. Laboratory and imaging preoperative examinations were requested before surgical procedure.

The Jatene procedure was performed, IVC was closed with bovine pericardium and IAC was corrected with direct suture. After surgery, the patient presented with intravascular volume depletion, polyuria, and hyperglycemia. She needed a red blood cell concentrate transfusion and several courses of hydration with Ringer's lactate but presented with good ventricular function.

The patient left surgery with drainage tubes in the left and right chest, as well as nasogastric and vesical tubes, and was under mechanical ventilation. She was discharged from the cardiology ICU 6 days after the surgery. An echocardiogram was performed 7 days after the surgery, which showed good ventricular function, mild residual aortic stenosis and insufficiency, moderate pulmonary hypertension, and enlarged right chambers. The patient showed good recovery and was discharged from the hospital 10 days after the TGA correction in good general condition and with good oral acceptance of food. Digoxin, sildenafil, and furosemide were prescribed. The patient's legal guardian was instructed to bring the patient back to the cardiology outpatient facility for a clinical follow-up.

COMMENTS

The patient in this case report was a 1-year-old child, with a clinical presentation of mild central cyanosis, and was diagnosed late with TGA and associated IVC and IAC. Although TGA is more common in boys (male: female ratio of 3:1),[2] the patient in this case report was a girl.

TGA is a very severe cyanotic heart disease and usually requires surgical repair in the neonatal period. However, in some patients, including the one in this report, the presence of IVC and IAC allowed blood to be mixed and the patient to survive. The severity of the condition is inversely related to the size of the communications.[4] In the present case, the patient had a large IVC and was diagnosed late due to a lack of perception of the mild cyanosis and probably absence of neonatal cardiology screening.

According to Jatene *et al.*[4], a definite diagnosis can only be made with echocardiography. Catheterization is indicated for a better evaluation of the heart anatomy, particularly regarding the insertion of the coronary arteries[5]. In this patient, catheterization did not show any anatomical variations. A similar case was reported by Atik *et al.*[6], in which a 7-month-old male patient did not present cyanosis and diagnosis was confirmed with echocardiography after heart failure. The patient had been treated clinically at first, without clinical improvement. The definitive treatment was the Jatene procedure, with a good clinical improvement after the procedure.

With respect to treatment, the surgical technique used for correcting TGA has varied over time. The postoperative outcome changed from a short survival period to high long-term survival rates[7]. Jatene *et al*.[4] and Dodge-Khatami *et al.*[7] analyzed different surgical techniques performed over time, emphasizing the Jatene procedure, which is now the first-choice treatment for correcting TGA. The correction was partial at first by applying palliative treatments, such as atrioseptostomy, and even more invasive procedures, such as those developed by Senning and Mustard.

Although the evolution of definitive treatment currently leads to complete TGA correction, some residual complications may occur. Such problems are related to the patient's TGA anatomical condition or the chosen surgical technique and include supraventricular arrhythmias, right ventricular dysfunction, tricuspid valve insufficiency, and stenosis of the pulmonary or systemic venous system (or both).

These conditions may require further surgical intervention. In patients with Taussig-Bing syndrome or TGA + IVC, the interventricular septal deviation anteriorly and laterally (to the right) may result in right ventricular hypoplasia affecting the subaortic region. Furthermore, subpulmonary pressure gradients may be generated by hypertrophic septoparietal trabeculations and abnormal insertion of the atrioventricular valves[8,9].

Early diagnosis is crucial because pulmonary hypertension is directly affected by patient age when surgery is performed. Other complications may appear, even if they do not directly influence the patient's prognosis; these include pulmonary stenosis, which occurs less frequently, and aortic insufficiency, which affects the patient's hemodynamic function in only 5% of cases. Moreover, such patients may develop functional problems such as decreased exercise capacity, diffuse coronary insufficiency, and neurologic development deficits[7,8].

With regard to preoperative complications, patients who are not diagnosed before birth develop more complications in the short- and long-term. The incidence of metabolic acidosis and low oxygen saturation is higher, which may lead to a greater risk of multiple organ failure and increased mortality rate[10]. Untreated patients with large IVCs may present with hepatomegaly and signs of congestive heart failure, which may lead to different outcomes, including acute pulmonary edema[5]. In contrast, an early intervention, especially in simple TGA, may lead to lower incidence of postoperative complications and lower morbidity. It may also positively affect the child's neuromotor and psychological development[10].

Similar to the first successful surgery performed by Jatene, the case presented herein involved TGA with IVC[4]. The Jatene procedure was chosen for the definitive treatment of the patient and correction of the IVC and IAC. The procedure was uneventful and the patient had a good postoperative evolution. She remains under follow-up as an outpatient.

The Jatene procedure is highly complex and therefore should be performed by an experienced surgeon, particularly when anatomical variations are present. However, when well executed, it has a good clinical outcome and has allowed an increase in TGA survival rate to up to 96%. For this reason, this technique is now considered the definitive first-choice treatment for TGA[4].

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