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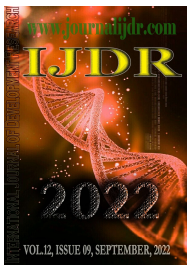

CASE REPORT OPEN ACCESS

DOUBLE-ENTRY UNIVENTRICULAR CONGENITAL HEART DISEASE WITH COARCTATION OF THE AORTA, TRANSPOSITION OF GREAT VESSELS AND GRAND ARTERIOUS CANAL: A CASE REPORT

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ABSTRACT

This study aims to report the case of a patient with double entry univentricular congenital heart disease with coarctation of the aorta, transposition of great vessels and great ductus arteriosus, whose treatment was specific and individualized surgery to correct anatomical and functional anomalies. The information was obtained by reviewing medical records, interviewing the patient's guardians, photographing the diagnostic and therapeutic methods to which the patient was submitted, and reviewing the literature. The case reported brings to light the therapeutic discussion of a complex situation that is univentricular heart disease and shows that, despite the degree of complexity of the surgeries, it is capable of obtaining satisfactory results that improve the patient's quality of life.

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INTRODUCTION

Conceived heart disease is one of the most prevalent diseases in neonates in the world. According to the Department of Pediatric Cardiovascular Surgery (DCCVP) of the Brazilian Society of Cardiovascular Surgery (SBCCV), the prevalence of heart disease is eight in each ten children for a thousand live births. Therefore, the number of new cases is estimated as twenty eight thousand eight hundred and forty sic (28.846) per year. Among them, aortic coarctation can be mentioned, one of the cardiopathies capable of developing the most severe cases of cardiac infusion and reactivating the circulation, with a systemic circulation dependent on the patency of the ductus arteriosus (Zielinsky, 1997). From a diagnostic point of view, aortic coarctation is difficult to recognize intrauterine echocardiography, developing a hypothesis based on some indirect propaedeutic data (Sharland, Chan & Allan, 1994). Thus, the most striking aspect is the disproportion in size between the two ventricles (RV), with the ventricle being larger and more hypertrophic than the left ventricle (LV). Likewise, the pulmonary artery is usually dilated, being of a much larger caliber than the ascending aorta.

On the other hand, the descending aorta is large, from the insertion of the ductus arteriosus. Often, there is a relative hypoplasia of the ascending aorta. Finally, Hypoplastic Left Heart Syndrome (HLHS) which despite its low incidence, 0.016 to 0.036% of births, causes 23% of cardiac deaths during the first week of life and 15% of cardiac deaths in the first month of life, followed by Hypoplastic Right Heart Syndrome (HRHS), poorly documented in the literature because it is an even rarer condition (Silva, Lopes & Silva, 2012). HRHS involves atresia of the pulmonary valve that has not formed, a very small right ventricle, a small tricuspid valve, and a small hypoplastic pulmonary artery. As the ventricle has failed to grow and develop the muscular structure of the ventricles, additional problems are encountered when the heart tries to pump blood to the pulmonic valve for transfer to the lungs. The adequate amount of blood pumped from the right atrium is not enough and this causes the blood to not be pumped efficiently to the lungs (Amaral et al., 1996). In this way, treatment is instituted, which must be performed soon after birth or after diagnosis and initially consists of intensive preoperative therapy with the objective of keeping the ductus arteriosus patent followed by palliative reconstruction divided into three stages or heart transplantation. The

first stage consists of the Norwood Sano operation, which must be performed in the first days of life, which consists of anastomosis of the pulmonary trunk to the aorta, atrial septectomy and systemic pulmonary graft of polytetrafluroethylene. The second stage is the bidirectional Glenn or hemi-Fontan operation, in which the flow from the superior vena cava is diverted to the pulmonary trunk. It is performed between two and six months of life. In the third stage (Fotan operation), the flow from the inferior vena cava to the lung is already directed. It must be performed between 18 and 24 months of age. In this context, the present report describes the case of a male patient with complex congenital heart disease, with a double inlet left ventricle, patent foramen ovale and ductus arteriosus, dilated confluent pulmonary arteries, dextrotransposition of the great arteries and coarctation of the aorta.

CASE REPORT

Male patient, born on 03/06/2018, term, by cesarean section (3.66 kg and 50 cm), with APGAR 8/9, completedprenatal care performed, without any complications, with 7 days of life, he returned maternity due to persistent physiological jaundice, being referred to the Intensive Care Unit to undergo phototherapy. On the same day of admission to the ICU, the patient presented with acute central cyanosis, and oxygen therapy was performed. Chest radiography ruled out possible obstruction by a foreign body of the bronchial tree. On the same day, a transthoracic echocardiogram was requested, which indicated a left ventricle with double inlet, with discordant ventriculo-arterial connection (aorta anterior to the pulmonary artery) and interruption of the aortic arch with patent ductus arteriosus (Figure 1).

tachycardia, cold sweating, metabolic acidosis, Hb saturation 76%, making it impossible to carry out air transport to a final destination. Therefore, after minimal stabilization, on march seventheenth (03/17/2018), with 11 days of life, he was referred to a local referral hospital, where he was maintained on artificial mechanical ventilation (FiO2 21% RR = 18) and on vasoactive drugs (epinephrine and alprostadil). Through these measures, referred stability was achieved, maintaining Hb saturation at 89%, BP 73/37 mmHg (mean 49 mmHg), HR 149 bpm, temperature 36.6°C. He had a urinary output of 3.7 ml/kg/hour in association with furosemide 1mg/kg/day and the last blood gas analysis showed no metabolic acidosis. In view of this situation of stability, air transport to a referral hospital in another state was possible, being admitted to the Intensive Care Unit on 03/18/2018, 12 days after his birth. For diagnostic verification, CT angiography of the heart was performed, which confirmed the diagnosis of complex congenital heart disease (double inlet of a single left ventricle, with transposition vessels, important coarctation of the aorta, crossed pulmonary arteries and large ductus arteriosus) (Figure 2). In addition, a cranial tomography was performed, which showed an area of ischemia in the border region and intraparenchymal hemorrhage (data not shown). On 03/23/2018, 5 days after admission to a referral hospital, the patient was evaluated by clinical neurology, which cleared him for a surgical procedure. However, the patient suffered from sepsis, initiating antibiotic therapy with Meropenem, later associated with Amikacin, Vancomycin and Micafugine until blood cultures became negative. Thus, after 21 days of antibiotic therapy, a surgical procedure was performed on 04/13/2018, with Norwood-type correction with Blalok with n4 tube. During the surgery, he presented bleeding that was difficult to control, and he was kept with an open chest. After controlling the condition, one day after the Norwood operation, he underwent thoracotomy with open drainage for revision and closure.

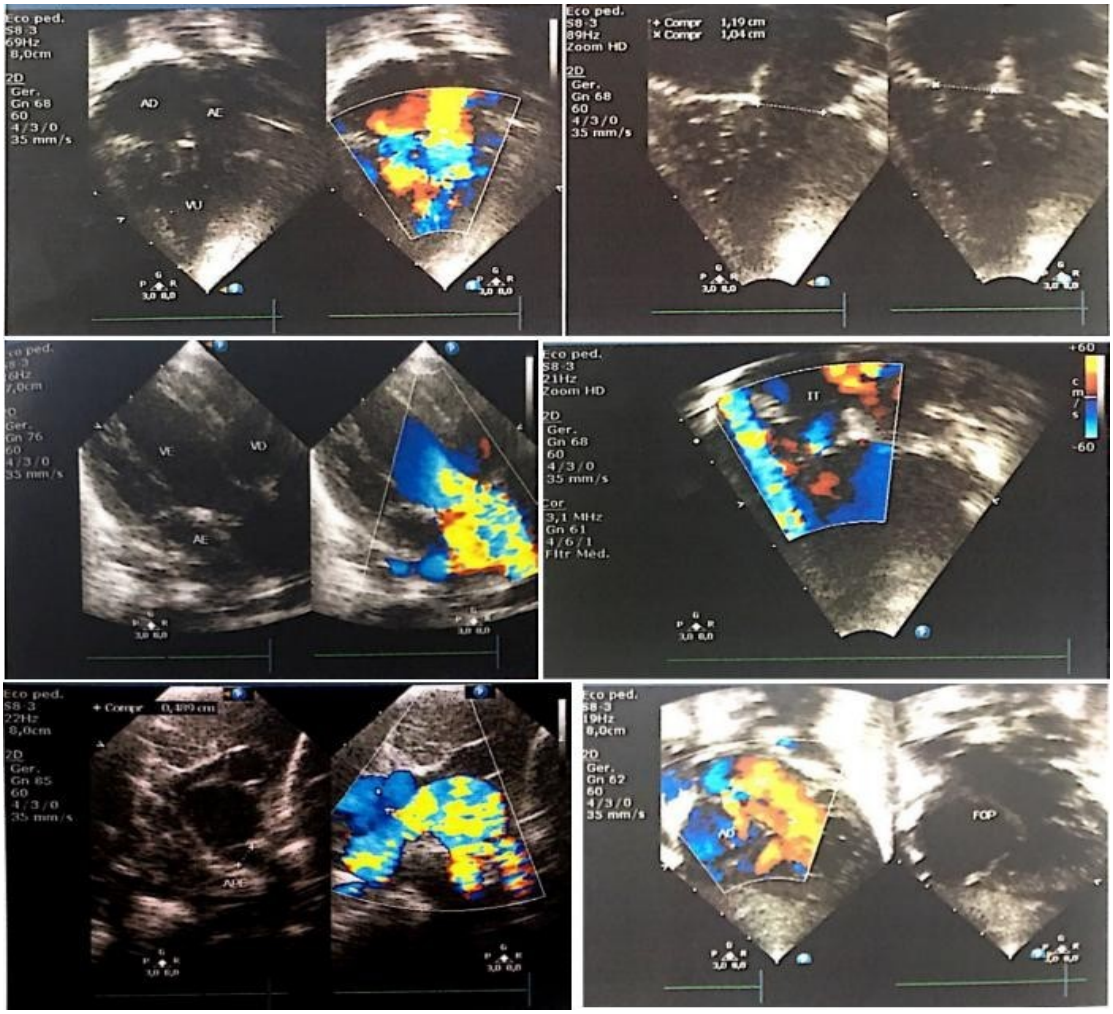

Figure 1. Echocardiogram performed at hospital admission showing a left ventricle with double inlet (top and center images), with discordant ventriculo-arterial connection (aorta anterior to the pulmonary artery) and interruption of the aortic arch with patent ductus arteriosus

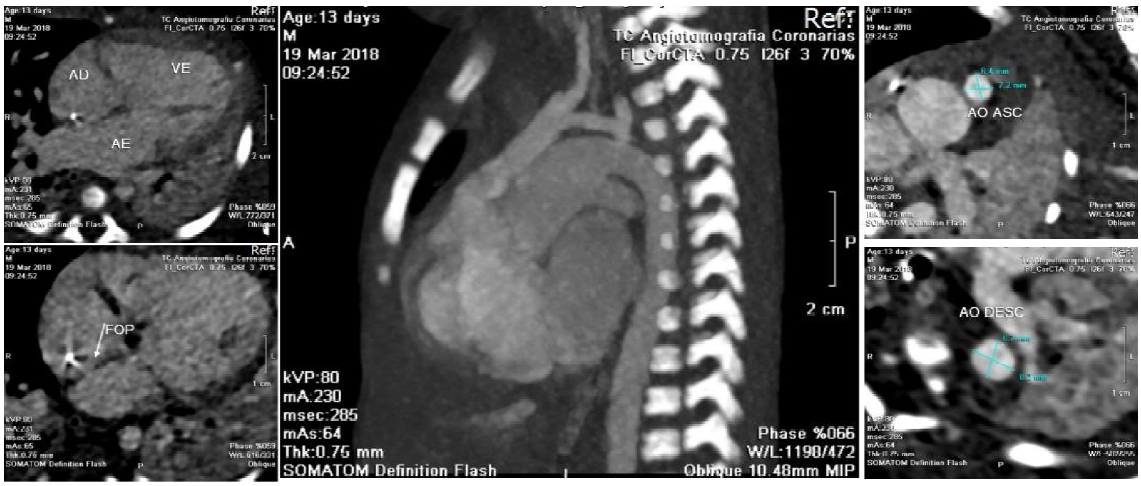

Figure 2. Angiotomography of the heart, showing complex congenital heart disease: double inlet of a single left ventricle (top left image), with transposition vessels and crossed pulmonary arteries (center image), aortic coarctation (right images), and large ductus arteriosus

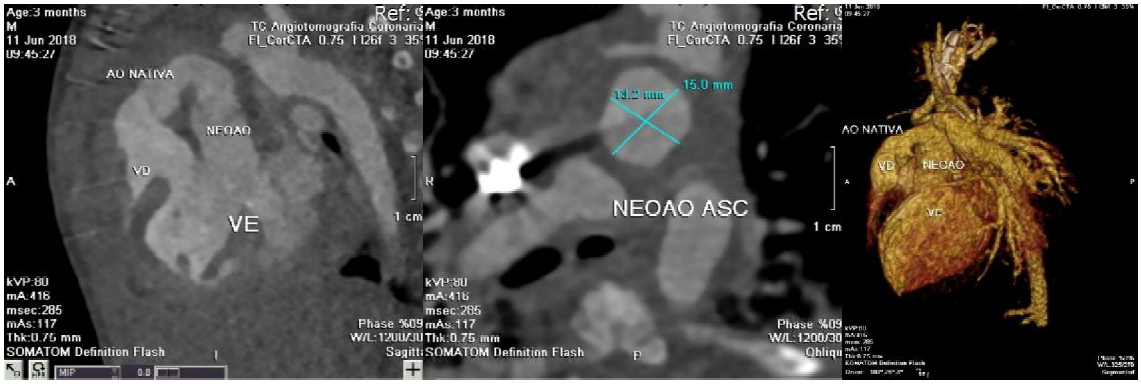

Figure 3. Angiotomography showing neo-aorta

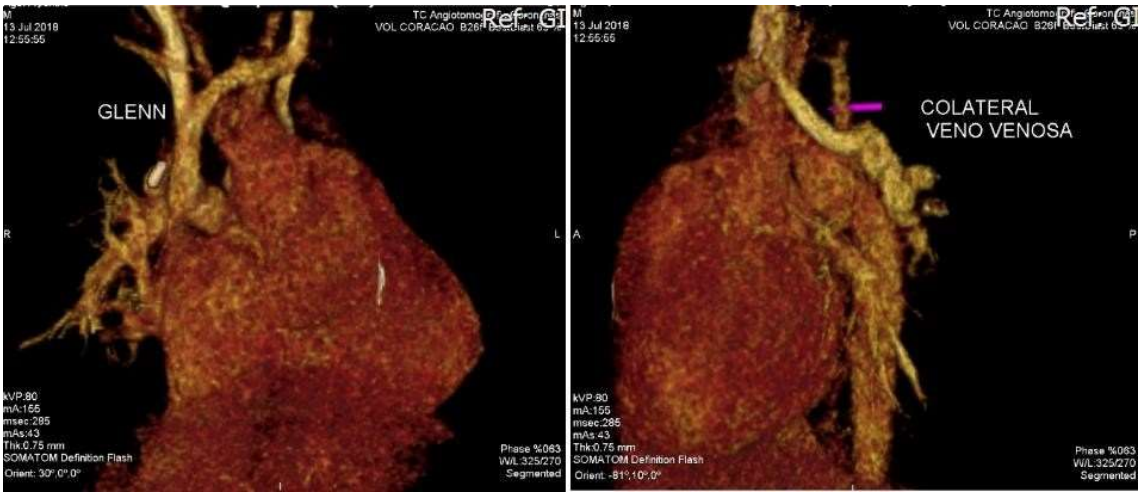

Figure 4. Post-Glenn CT angiography (left), showing venovenous fistula (right)

He showed clinical improvement-slow and progressive-allowing weaning from mechanical ventilation and vasoactive drugs with discharge from the ICU to the ward on 05/16/2018. Before undergoing the second surgery, a new CT angiography was performed to verify the changes obtained with the first surgical intervention (Figure 3). During the interstage period, the patient did not present any complications, and thus, on 06/28/2018, approximately 4 months after his birth, he underwent Glenn's surgery, with mitral valve plasia and tricuspid valve plasia, evolving with hypoxemia aggravated by systemic and pulmonary infection, and later with urinary tract infection by Meropen-sensitive Klebsiella pneumoniae 2 days after surgery. After negative cultures, he was kept on mechanical ventilation until 07/09/2018, remaining in the ICU using a high-flow catheter, with borderline O2 saturation.

Two weeks after Glenn's surgery, he underwent computed tomography angiography to analyze the Glenn for a better assessment of hypoxemia, showing the presence of a large important veno-venous fistula with paravertebral ramifications without the possibility of percutaneous closure (Figure 4). After extubation, he developed severe hypoxemia worsened by CHF and respiratory infection, requiring reintubation and mechanical ventilation. Two weeks after the diagnosis of the fistula, he performed a new cardiac surgery for ligation of a veno-venous fistula. He evolved with hemodynamic stability and was referred to the ward on 08/07/2018. In the ward, he evolved with clinical improvement, weight gain, good acceptance of oral diet. An echocardiogram was performed on 08/22/2018, which concluded by the evolutionary control of the fistula closure

postoperative period. The minor evolved in good general condition and was discharged on the same day, with outpatient follow-up guidance with cardiology, hematology, endocrinology, neurology, physical therapy, speech therapy and occupational therapy.

DISCUSSION

Treatments for congenital heart defects, in general, have developed substantially in recent decades, being adapted to the needs of each baby and, consequently, depending on the degree of complexity presented in each case. Therefore, recognizing the patterns of malformations and their complexity is always important, so that adequate decision-making can be carried out. In the present report, we present a case of complex congenital heart disease, presenting a series of patterns rarely seen together: double inlet left ventricle, patent foramen ovale and ductus arteriosus, dilated confluent pulmonary arteries, dextrotransposition of the great arteries and coarctation of the aorta. Using descriptors of patterns of congenital cardiac malformations, isolated or combined by Boolean operators, we performed a search in the PUBMED article database, and found the following amount of studies reporting the presence of these malformations together, in the same patient or group of patients: patients: “atrial septal defect”, 21187 results; patent ductus arteriosus; 12671 results; “coarctation of the aorta”, 12156 results; “transposition of the great arteries”, 9,859 results; “patent foramen ovale”, 5979 results; “double-entry left ventricle”, 409 results; “dextrotransposition of the great arteries”, 152 results; “double-entry left ventricle AND aortic coarctation”, 29 results; “dextrotransposition of the great arteries AND atrial septal defect”, 16 results; “dextrotransposition of the great arteries AND patent ductus arteriosus”, 7 results; “double-entry left ventricle AND coarctation of the aorta AND transposition of the great arteries”, 14 results; “transposition of great arteries AND confluent pulmonary arteries”, 7 results; “double-entry left ventricle AND confluent pulmonary arteries”, 4 results; “double-entry left ventricle AND coarctation of the aorta AND transposition of the great arteries AND atrial septal defect”, 3 results; “double-entry left ventricle AND coarctation of the aorta AND transposition of the great arteries AND patent foramen ovale”, 1 result; “double-entry left ventricle AND coarctation of the aorta AND transposition of the great arteries AND patent ductus arteriosus”, 1 result; “double-entry left ventricle AND coarctation of the aorta AND right-hand transposition of the great arteries”, 0 results; “double-entry left ventricle AND coarctation of the aorta and transposition of the great arteries AND atrial septal defect AND confluent pulmonary arteries”, 0 results; “double-entry left ventricle AND transposition of the great arteries AND confluent pulmonary arteries”, 0 results.

CONCLUSION

Thus, we conclude that the case reported in the present work will certainly contribute to the mapping of potential joint occurrences of congenital cardiac malformations and, consequently, to the development of strategies for their effective treatment.

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