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Research Article

Successful Palliation in Monochorionic Twins with Hypoplastic Left Heart Syndrome

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Abstract

Hypoplastic left heart syndrome is a congenital heart disease which accounts for 8% of all congenital heart defects. Twin pregnancies have a significantly higher risk of structural heart disease than singleton ones, however the concordance rate has been found to be relatively low, even in monochorionic pregnancies. We describe here the favorable evolution of a monochorionic, diamniotic, twin pregnancy in which both fetuses were diagnosed with hypoplastic left heart syndrome by antenatal echocardiography. The present report suggests that at least in some cases there may be a genetic contribution to the etiology. The fetal diagnosis was imperative to a good postnatal outcome.

Keywords: hypoplastic left heart syndrome; congenital heart disease; twin pregnancy; Norwood-Sano palliation

Abbreviations

CT: Computed Tomography

HLHS: Hypoplastic Left Heart Syndrome

ICU: Intensive Care Unit

Introduction

Hypoplastic left heart syndrome (HLHS) is a congenital heart disease in which the left ventricle and aorta have varying degrees of hypoplasia associated with stenosis or atresia of the mitral and/or aortic valve [1]. It accounts for 8% of all congenital heart defects, with an estimated birth incidence of 0.1-0.25 in 1000 live births [2].

As the majority of congenital heart disease, HLHS presents complex genetic a etiology [3]. Twin pregnancies have a significantly higher risk of structural heart disease than singleton ones, however the concordance rate has been found to be relatively low, even in monochromic pregnancies [2].

We describe the favorable evolution of a monochorionic, diamniotic, twin pregnancy in which both fetuses were diagnosed with HLHS by antenatal echocardiography.

Sister 1

Patient 1 twin was born on 04/03/2020 by cesarean delivery, on 36 weeks, Apgar 9 and 10, birth weight of 1,980 grams. No resuscitation maneuvers were required. Patient was referred to the Cardiac Intensive Care Unit (ICU) where prostaglandin was initiated. Echocardiogram showed a 3.5 mm *ostium secundum* atrial septal defect, mild tricuspid insufficiency, mitral stenosis, aortic atresia, and a hypoplastic left ventricle. Pulmonary trunk was 11 mm (score Z: +2.86). Left aortic arch with retrograde flow. Ascending aorta was 4.3 mm (score Z: -6.06), transverse aortic arch 4.2 mm (score Z: -3.06), thoracic descending aorta was 6 mm (z score: +1.02) and isthmus was 3 mm (z score: -2.83). The wide arterial ductus measured 3.5 mm. There were normal coronary arteries and a preserved right ventricle function. Due to patient's low weight, it was chosen to perform

branch pulmonary artery banding, which happened on 04/07/2020. Two days later, patient was extubated. She was kept under prostaglandin infusion until Norwood palliation, which was indicated when patient achieved 3Kg. The procedure was performed on 05/02/2020, without complications. Cardiopulmonary bypass time: 190 minutes. Clamping time: 66 minutes. Total circulatory arrest: 44 minutes. She was transferred to the cardiac ICU with open chest, which was closed two days after cardiac surgery. She received prophylactic Cefuroxime and Vancomycin for 48h. After extubation, on 05/05, vasoactive drugs were progressively weaned until 05/09. On 05/12, she presented movements in the left upper limb and her electroencephalogram showed seizures, which ceased after Phenobarbital. Brain computed tomography (CT) showed multiple areas suggestive of ischemic events. She was transferred to the pediatric ward on May 13, 2020. On August 11th, patient returned to cardiac ICU after being submitted to cavopulmonary procedure. Surgery was performed without complications, and she was extubated at the same day. Discharge to Ward occurred three days later and from the hospital on September 1st.

Sister 2

Patient 2 twin, was born on 36 weeks, Apgar 8 and 9, birth weight of 1810 g. No resuscitation maneuvers were required. After admission in Cardiac ICU, prostaglandin and milrinone were started. Echocardiogram showed mitral atresia and aortic stenosis. *Ostio secundum* atrial septal defect measured 4.2 mm, with left-right shunt and no restriction. A muscular ventricular septal defect was present in the outflow tract, measuring 3.2 mm with right-to-left shunt and no significant gradient. Right chambers presented moderate dilation and left ventricle was hypoplastic. There were moderate tricuspid insufficiency and mild pulmonary insufficiency. Pulmonary trunk was dilated, measuring 9 mm (score Z: +1,67). Aortic valve was thickened, trivalvular but with bivalvular opening and without significant systolic gradient. Anterograde blood flow was of difficult evaluation, since there was severe reduction of aortic arch between left carotid artery and left subclavian, isthmus measured 1,7 mm. Aortic root was 6.86 mm (score Z: -1.83), synotubular region 5.2 mm (score Z: -1.66), ascending aorta 4.1 mm (score Z: -6.2). and transverse arch 1.7 mm (score Z: -8.9). The wide arterial ductus measured 4.4 mm. There were normal coronary arteries and a preserved right ventricle function. To exclude the hypothesis of interrupted aortic arch, a CT scan was performed and confirmed the diagnosis of HLHS. The exam also showed presence of persistent left superior vena cava draining in dilated coronary sinus, ascending aorta with preserved dimensions, severe aortic arch hypoplasia and isthmic coarctation, and no interrupted aortic arch. The bulb had a caliber of 7.2 x 7.1 mm, ascending aorta 6.2 x 5.2 mm, transverse arch 2.1 x 2 mm, isthmus 2.5 x 2.4 mm. Descending aorta presented a caliber of 6.8 x 6.4 mm and thoracoabdominal transition 6.4 x 5.8 mm. Branch pulmonary artery banding was the procedure initially chosen due to the low weight at birth. Surgery was performed without complications on 04/07/2020 and patient presented a good evolution at cardiac ICU. On 05/14/2020, she underwent Norwood-Sano surgery. Cardiopulmonary bypass time was 181 minutes, clamping time 74 minutes and total circulatory arrest was 52 minutes. She was admitted to the ICU with open chest, hemodynamically stable. Thoracorrhaphy was performed on May 16th, without complications. She was extubated on May 17th and all vasoactive drugs were removed on May 19th. Three days later, she evolved to surgical wound infection and on May 23rd, surgical cleaning was performed. Enterococcus was evidenced and treatment was completed with piperacillin + tazobactam and vancomycin. She was extubated again on May 27th and was discharged to pediatric ward on June 2nd. On August 18th, patient was submitted to bicaval cavopulmonary connection and tricuspid valve plasty. She was extubated at the same day and was discharged to Ward on August 21st. On August 29th, she returns to cardiac ICU after clinical seizures, confirmed on electroencephalogram. Brain CT from August 30th demonstrated extensive ischemic cerebral stroke in middle cerebral artery territory. She was reintubated on August 31st after a new episode of seizure and brain CT was repeated to exclude the presence of hemorrhagic stroke. She was extubated three days later and on September 23rd she was discharged to ward again. Discharge home occurred on September 28th.

Discussion

Although rare, HLHS has previously been reported in monochorionic twin pregnancies affecting both fetuses. The risk of structural heart disease is higher in twin than in singleton pregnancies, but the concordance rate is relatively low, implicating non-genetic factors in the etiology [4]. However, the present report suggests that at least in some cases there may be a genetic contribution to the etiology.

The fetal diagnosis was imperative to a good postnatal outcome. It allowed planning the delivery in a reference center, which is familiarized with preoperative and postoperative care, since HLHS is nearly always fatal without surgical palliation [5].f Besides, prenatal recognition of disease allows families to prepare for a child with a life-threatening defect. Evaluation for extracardiac anomalies is also important for these patients, as well as genetic counseling [2].

Although the disease is still associated with a high mortality rate in some centers worldwide, both infants presented a good evolution and were submitted to surgical second stage (cavopulmonary or Glenn procedure) and were discharged home. Our team is one with the best results with this type of pathology in Brazil [5].

Acknowledgments

None.

Conflicts of interest

None.

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