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HYPOPLASTIC LEFT HEART SYNDROME (HLHS): A CHALLENGE FOR CARDIOFETAL MEDICINE

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INTRODUCTION

Hypoplastic Left Heart Syndrome (HLHS) is a rare heart disease, whose worldwide incidence ranges from 0.016 to 0.036%, representing only 1.5% of congenital heart diseases.

HLHS is characterized as a morphological underdevelopment, resulting in very reduced dimensions of the left heart chambers, represented by the left ventricle, mitral valve, aortic valve and ascending aorta. Cardiac malformations are prevalent associated with chromosomal disorders. However, the etiology is multifactorial and can be found without genetic factors (image 1).

The fetus has anatomical and physiological resources, such as the ductus arteriosus and the foramen ovale, to remain hemodynamically stable. However, at birth, if it does not obtain immediate clinical-surgical intervention, it evolves to death.

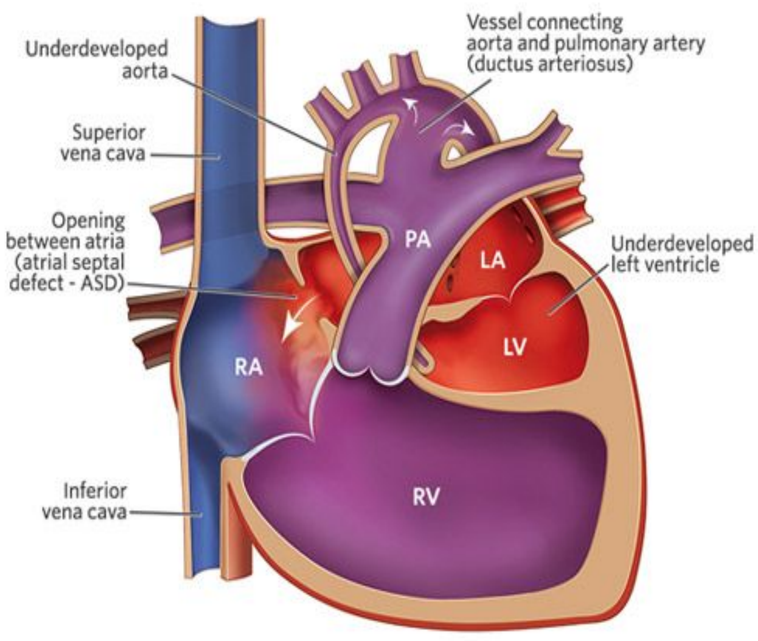

Image 1: HLHS adapted. 2014 The Children's Hospital of Philadelphia

Clinical-surgical therapy for HLHS is not curative, but palliative. It is based on prostaglandin E1 or interatrial catheterization shortly after birth and a 3-step surgical sequence: Norwood, Glenn and Fontan operation. Even with surgical therapy, the mortality rate remains high, around 82.4%. Heart transplantation is the only curative treatment, but, due to the reduced possibility, many newborns end up dying before the surgery.

Therefore, the prognosis remains unfavorable. Studies have shown incidence of microcephaly, seizures, delay in neuropsychomotor development, recurrent hospitalizations and a maximum life expectancy around 20 years of age.

OBJECTIVE

The objective of this study was to report the case of a patient, pregnant with a fetus diagnosed with HLHS, followed in by the high-risk prenatal care at the Hospital Universitário São Francisco (HUSF), in Bragança Paulista - SP, Brazil, in January 2023. Beyond that, this study aims to raise the current knowledge about HLHS: its etiology, epidemiology and pathophysiology, as well as its therapeutic, prognostic and procedural challenges in this region.

METHODS

The study was carried out through quantitative and descriptive research, analyzing two cases of HLHS at the HUSF in Bragança Paulista and their respective outcomes at another hospital, PUC-Campinas. In addition, an extensive bibliographical review in the main data platforms such as Scielo, PubMed, Google Scholar and Lilacs whose selection criterion was the keywords Hypoplastic Left Heart Syndrome and to be between the years 2019 and 2023.

RESULTS

We present the case of a 34-year-old tercigest with a gestational age of 34 weeks and 3 days who was referred to the high-risk prenatal care at Hospital Universitário São Francisco (HUSF) in Bragança Paulista, SP, Brazil, in January 2023 with a cardiac abnormality seen on fetal echocardiography. A second ultrasound was requested and confirmed the diagnosis of HLHS (Image 2).

Once the diagnosis was confirmed and knowing the level of complexity of care the baby would need to receive at birth, we made numerous requests for transfers to major centers in the state of São Paulo. Many were denied, alleging that the woman's prenatal care should be completed in the high-risk service where she started.

Despite the regional bureaucracy, in about 2 weeks we managed to get the PUC-Campinas hospital to accept her. The woman was transferred at 37 weeks to that reference hospital in the region, where a cesarean birth was performed and subsequent catheterization in the newborn (NB). Five days later, Norwood surgery was performed in the NB. Even so, the NB did not have a good recovery, needing to be placed on ECMO. Unfortunately, after six days, the baby died.

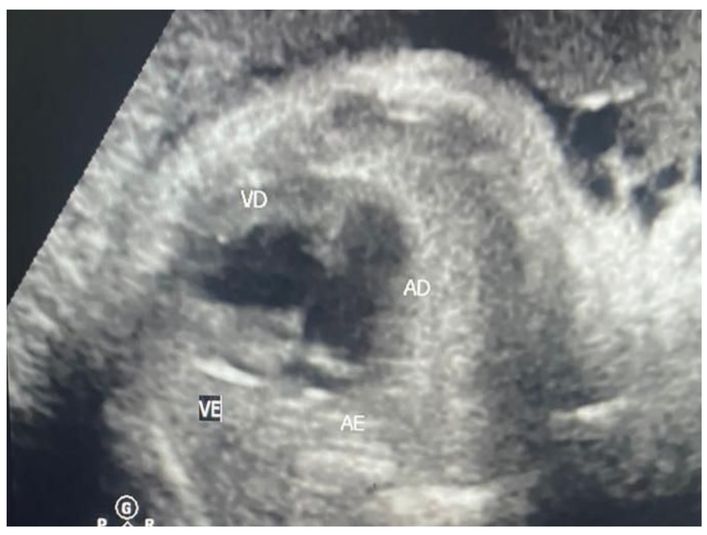

Image 2: ultrasound made with 36 weeks showed the asymmetry of the left and right cavities of the heart

We also present the case of a 25-year-old secundigravid with gestational diabetes and a gestational age of 33 weeks and 6 days who was referred to the high-risk prenatal care at HUSF in July 2020, already diagnosed with HLHS. This patient was also transferred to PUC-Campinas where several surgeries were performed and her son is alive at 2 years old, but with sequels of the disease. As the case was transferred to another hospital, we have few details of the case and the follow-up was unfortunately lost.

Both cases of HLHS at the HUSF were diagnosed and referred to the high-risk group late, around 34 weeks of gestation. This scenario demonstrates the problem of the current Unified Health System (SUS) in Brazil, because, while it allows access to medical care and exams, the lack of knowledge, structure, system overload and excessive bureaucratization makes it difficult to monitor and treat these patients. Also the access to regional surgical treatment is difficult, as there are only 3 centers that are referenced and trained to receive a newborn with HLHS and perform the procedures in the region. In other words, the transfer of prenatal care for follow-up and subsequent scheduling of delivery in these places proves to be extremely bureaucratic, limited and ineffective.

CONCLUSION

Within the Brazilian reality, the referral of patients with fetuses with malformations to high-risk prenatal care is late. This leads to difficulty in transferring patients to specialized centers in advance, plan the necessary interventions and little time for adequate psychological support to the family.

It is also clear that there is a great lack of specialized services and trained professionals to perform heart surgeries and invasive procedures in newborns with heart disease in the region. It is up to the health managers responsible for administering the system to hire pediatric surgeons specialized in cardiac surgeries in newborns for this region. Probably, there is no initiative because they believe that cardiac malformations, such as HLHS, are rare diseases with little incidents. However, with this work, it is evident that the incidence has a significant value, greater than expected and greater than the world average. It is important to alert health professionals and the health administrative system that there is a significant demand and, therefore, to take such measures.