

Hypoplastic Left Heart Syndrome and Obstructive Total Anomalous Pulmonary Venous Connection: A Rare and Severe Association

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Introduction

Hypoplastic Left Heart Syndrome (HLHS) is one of the most frequent cardiac anomalies diagnosed in utero¹. Its incidence is 1 to 5 for every 10,000 live births², and may be underestimated due to immediate fetal and neonatal deaths. It is a severe disease that requires early intervention in the neonatal period.

Total Anomalous Pulmonary Venous Connection (TAPVC) is also a rare congenital anomaly that accounts for 1% to 3% of congenital heart diseases³. Surgical mortality is related to obstructive connections and associations with univentricular hearts⁴.

The association of HLHS with TAPVC is very rare, with little information in the literature, particularly when diagnosed in prenatal life.

This report describes a favorable outcome in a case with the association of both diseases, where a precise prenatal diagnosis allowed therapeutic strategy planning of a less-invasive and effective early treatment.

Case Report

First gestation of a 21 years-old woman referred for fetal echocardiography because of HLHS identified during the second trimester screening ultrasound. A fetal echocardiogram performed in the 31st gestational week confirmed the diagnosis; however, it was associated with supracardiac obstructive TAPVC, with a high velocity flow observed at the level of the vertical vein (1.14 m/s - Figure 1). Elective caesarean section was performed in the 38th gestational week in a hospital with a well-developed pediatric cardiology program, and the catheterization laboratory was ready to receive the baby shortly after birth. The Apgar score was of 6/7/8 and birth weight 3,295 kg. Elective endotracheal intubation at the ICU and catheterization of the umbilical vein and artery were performed. Continuous infusion of prostaglandin was initiated. In the first hours of life, oxygen saturation ranged

from 30% to 40% and the chest x-ray revealed severe pulmonary edema (Figure 2-A). Postnatal echocardiography confirmed the diagnosis and identified diminutive left atrium (Figure 3). The Newborn (NB) was immediately taken to the catheterization laboratory for obstruction relief of the vertical vein. Two stents were deployed next to the connection of the vertical vein with the common pulmonary vein (Figure 4). After the procedure, oxygen saturation rose to 80% and there was a significant improvement of pulmonary congestion (Figure 2-B), hemodynamic and metabolic status. In the third day of life the baby underwent a hybrid procedure (selective pulmonary artery banding and ductal stenting). She had a good post-operative course and was transferred to the ward on the eighth day.

During her stay in the ward, the baby had runs of supraventricular tachycardia, needing a new period of intensive care for medical treatment. She was discharged home ten days later and came back every other week for clinical and echocardiographic evaluation.

Because of increased flow velocities in the vertical vein stent and reduced systolic and diastolic gradients across the pulmonary artery bandings, surgery was performed to connect the common pulmonary vein to the left atrium. This procedure was performed in the second month of life, with good postoperative outcome. After discharge, the baby was followed at the outpatient clinic and had an appropriate somatic and psychomotor development, with oxygen saturation ranging from 68% to 78% and preserved right ventricular systolic function. The comprehensive stage II – the Norwood/Glenn operation – was scheduled for the age of sixth month.

Discussion

HLHS is the most challenging neonatal cardiac disease, with a mortality rate of 90% in the first month of life if not surgically addressed⁵. Although the Norwood procedure have been described for more than thirty years, its mortality persists high, particularly in developing countries⁶. The hybrid procedure for HLHS was introduced by Gibbs et al⁷ in 1993 and was adopted as the management of choice in many centers^{5,7-9}. This technique aims to keep the circulation pattern similar to that in fetal life, by increasing the flow resistance to the lungs with selective pulmonary arteries banding and maintaining systemic circulation through the stented ductus arteriosus^{5,7-10}. To ensure free pulmonary venous return from the left atrium to the right chambers, a percutaneous atrioseptostomy is performed one or two weeks after the hybrid procedure^{7,10}. Less invasive procedure not requiring cardiopulmonary bypass and faster postoperative course are the most important advantages of the hybrid approach.

Keywords

Hypoplastic Left Heart Syndrome; Heart Defects, Congenital; Pulmonary Veins/Abnormalities; Prenatal Care.

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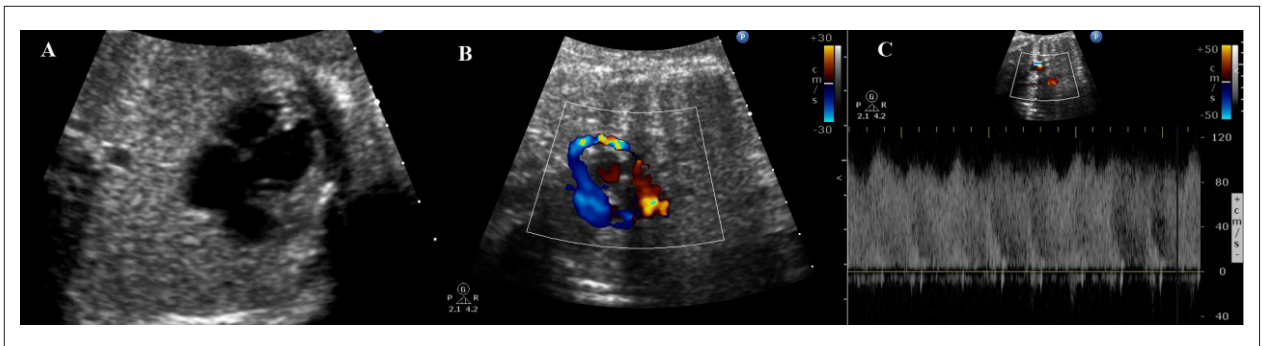


Figure 1 – Fetal echocardiography. A) Apical four-chamber view, with increasing distance between the descending thoracic aorta and the left atrium. B) Venous collector and ascending vertical vein showing flow acceleration with (mosaic). C) Doppler in the ascending vertical vein at the site of greatest flow acceleration showing increasing velocity (1.14 m/s).

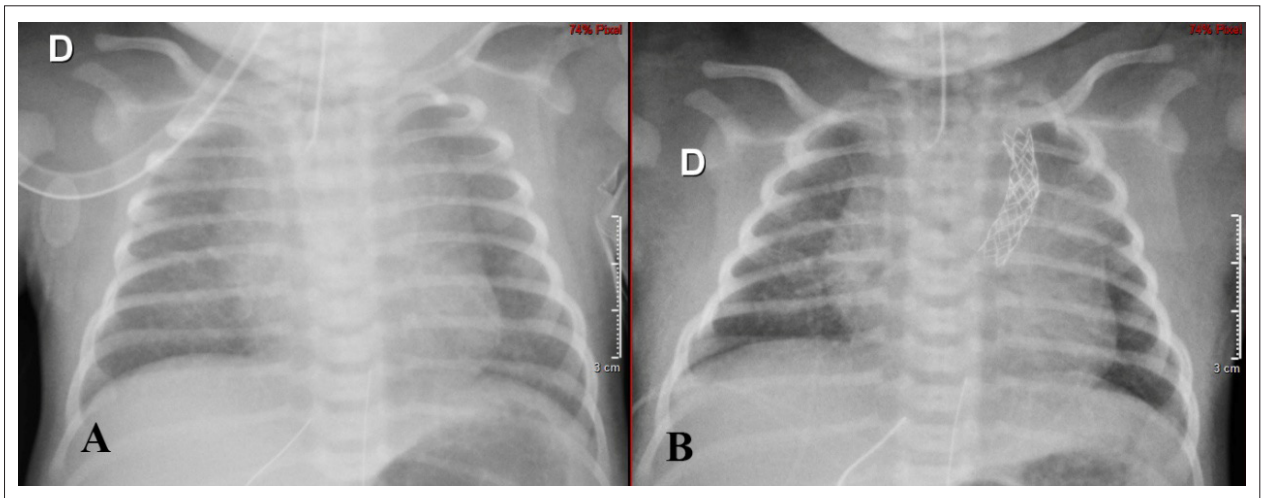


Figure 2 – Chest X-ray. A) After birth showing severe pulmonary edema. B) After stenting in the ascending vertical vein with improvement of pulmonary edema.

The association of HLHS with obstructive TAPVC has high morbidity and mortality, and its clinical presentation is similar to HLHS with critically restrictive atrial septal defect or intact interatrial septum^{4,7-12}. Pulmonary edema determines severe hypoxemia and low cardiac output, leading to circulatory collapse shortly after birth. Therefore, the fetal diagnosis of this association is essential for immediate and proper treatment planning.

Fetal diagnosis of HLHS is not difficult and may be suspected when a single right ventricle is associated with a hypoplastic ascending aorta. However, due to physiologic low pulmonary cardiac output in the fetal circulation, TAPVC may be easily missed. Therefore, finding at least two pulmonary veins connected to the left atrium is part of the routine assessment of the fetal heart. Doppler examination of the pulmonary vein flow is mandatory in fetuses with HLHS, and reverse “A” wave can be significantly increased when the atrial septal defect (ASD) is restrictive. In cases with critical flow restriction at the atrial level, the “D” wave (passive diastole), is reduced or abolished, with a bidirectional pulmonary vein flow pattern. Thus, assessment of

pulmonary venous drainage in HLHS is essential for neonatal treatment planning^{3,4,12}. One of the markers of TAPVC in fetal life is the increased distance between the left atrium and the descending thoracic aorta in the four-chamber view, as seen in this case (Figure 1-A). This occurs because the common pulmonary vein is usually located behind the left atrium, which can be identified with color flow Doppler evaluation with low pulse repetition frequency (PRF). In addition, color Doppler can demonstrate common pulmonary vein connection with the vertical vein which connects to the innominate vein and superior vena cava. The vertical vein obstruction was noticed exactly where it crosses the left pulmonary artery, and was identified because of flow acceleration with color and pulsed Doppler (Figure 1-C). In this case, although the superior vena cava receives the systemic and pulmonary venous return, it does not look dilated. The association of low pulmonary output during fetal life (lower than 10% of total fetal cardiac output), and significant obstruction at the vertical vein may justify this echocardiographic finding.

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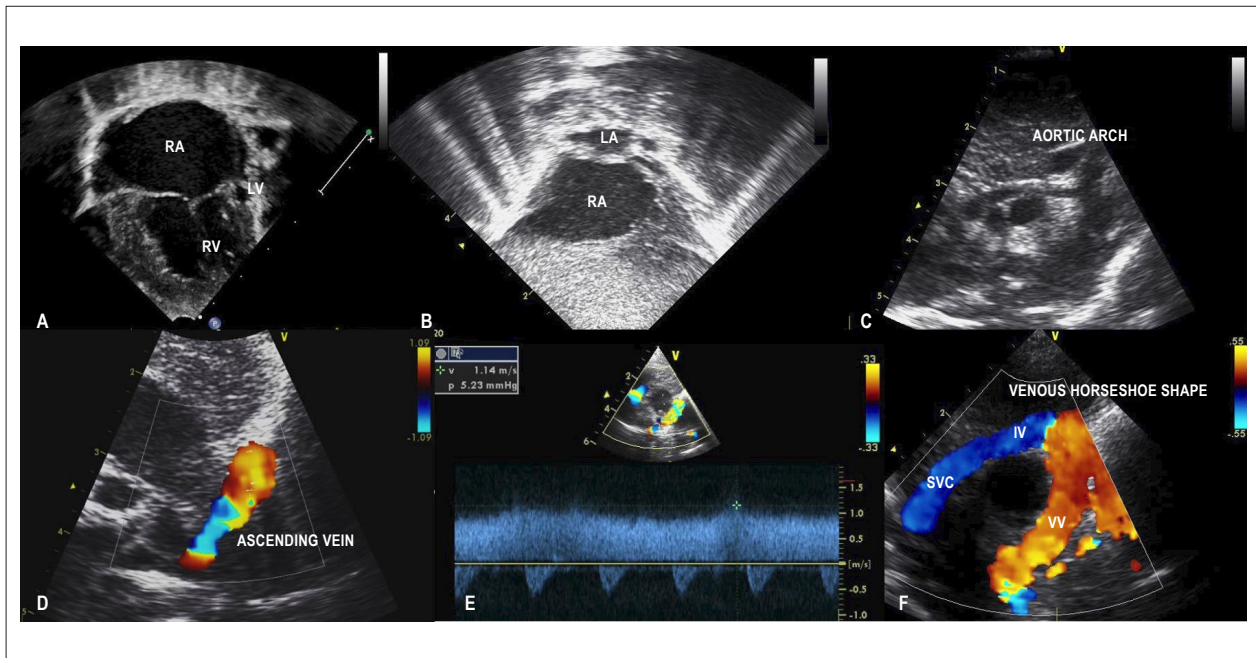


Figure 3 – Fetal echocardiography. A) Four-chamber apical view with hypoplastic left ventricle and dilated direct cardiac cavities. B) Subcostal view, tiny left atrium and intact atrial septum. C) Suprasternal view showing severe hypoplasia of the ascending aorta. D) Ascending vertical vein identified with color flow mapping showing flow acceleration at the site that intersects with the left pulmonary artery. E) Increased flow velocity in the ascending vertical vein (1.14 m/s). F) Supracardiac anomalous venous connection with vertical vein draining into the innominate vein and flowing into the superior vena cava (evaluation after stenting with relief of obstruction). RA: right atrium; RV: right ventricle; LA: left atrium; SVC: superior vena cava; IV: innominate vein; VV: vertical vein.

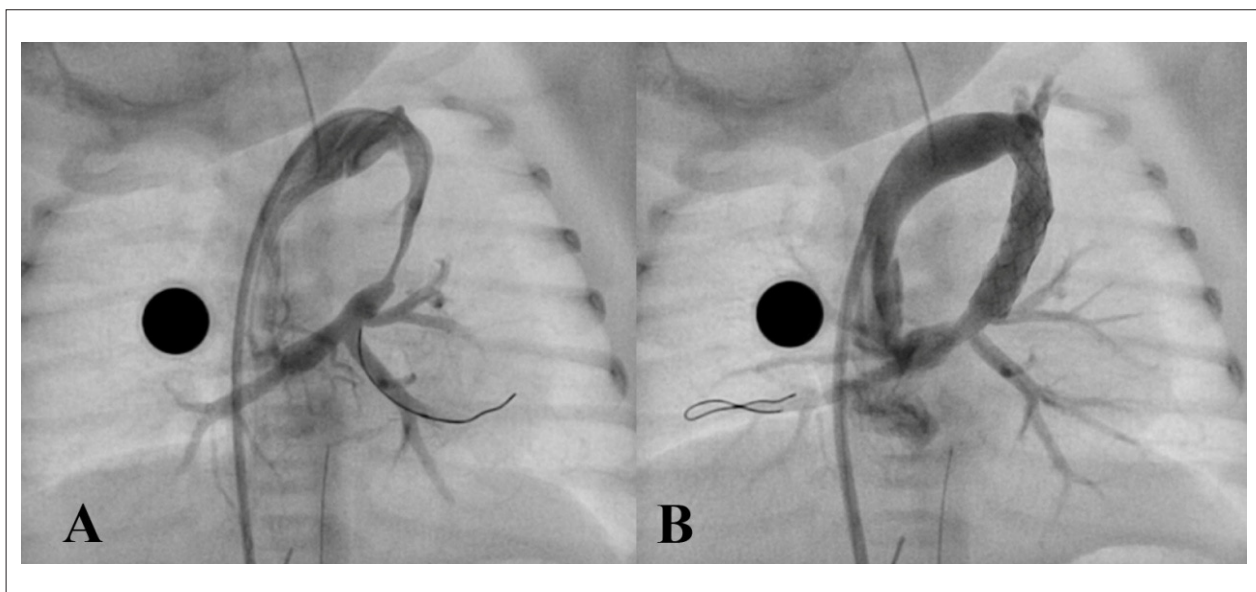


Figure 4 – Cardiac catheterization. A) Total anomalous pulmonary venous connection in venous lake that opens into the ascending vertical vein with critical narrowing at its proximal portion. B) Angiography performed after deployment of the two stents in the stenotic path of the ascending vertical vein.

When HLHS is associated with obstructive TAPVC in fetal life, two different therapeutic strategies can be planned: balloon dilation and/or stenting the vertical vein at the site of obstruction or common pulmonary vein connection to the left atrium. Schranz et al showed good results in connecting the common pulmonary vein to the left atrium in the catheterization laboratory¹². In the case here described, this procedure was not possible because the left atrium was significantly small. For this reason, we opted to stent the vertical vein first, allowing the baby to recover from the critical clinical condition developed soon after birth. However, due to the low flow velocity at the vertical vein, a rapid neointimal proliferation partially obstructed again the pulmonary venous return, requiring surgical procedure to connect the common pulmonary vein to the left atrium. Although the operation was performed under pulmonary bypass, because of the previous hybrid palliation, the baby had a very favorable outcome.

Another point to be discussed is the importance of prenatal diagnosis for planning birth in a pediatric cardiology center, which has all diagnostic and therapeutic resources available. These complex heart diseases do not allow any delay in diagnosis and inter-hospital transportation. It has become increasingly common to deliver babies with this kind of congenital heart malformation in the pediatric cardiology center allowing specific and specialized treatment immediately after birth.

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Conclusion

This case report illustrates the important role of prenatal diagnosis of critical heart diseases. Although the association of HLHS and obstructive TAPVC has a very high mortality rate, this baby had a good outcome because the therapeutic strategy was planned beforehand, and allowed immediate and adjusted neonatal treatment.

Authors' contribution

Research creation and design: Cosentino CM, Shiraishi KS, Stella AKS, Pedra SF; Data acquisition: Cosentino CM, Shiraishi KS, Stella AKS, Pedra SF; Analysis and interpretation of data: Cosentino CM, Pedra SF; Manuscript drafting: Cosentino CM, Pedra SF; Critical review of the manuscript as to relevant intellectual content: Cosentino CM, Martins TC, Pedra CAC, Pedra SF.

Potential Conflicts of Interest

No relevant potential conflicts of interest.

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Academic Association

This study is not associated with any graduate programs.

Case Report
