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Clinical Case

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Fetal left ventricular hypoplasia syndrome

Abstract

Left ventricular hypoplasia is a severe congenital heart disease with a currently reserved prognosis, even for patients who can be treated in the newborn period, either because they have been diagnosed early at birth or because the diagnosis was made in the fetal period. The latter is important because it allows the medical team to be prepared and to offer the patient the best management conditions in the neonatal period and a surgical treatment appropriate to their pathology and in the best situation in a tertiary level cardiac surgical center.

We present a case with an antenatal diagnosis where all the stages were carried out to provide an adequate treatment, but due to the complexity of the case the parents opted for compassionate treatment.

Keywords: hypoplastic left heart syndrome, atretic stenosis, protein-losing enteropathy, cardiac malformation

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Introduction

Fetal hypoplastic left heart syndrome (HLHS) is a rare and complex congenital heart disease. It is a categorical diagnosis of structural deficiency and insufficiency in the capacity of the left ventricle (LV) to sustain and provide systemic perfusión.[1]

Its rarity and high mortality rate have made it one of the most complex congenital heart diseases. In the past, when there was no treatment, all patients with LVHS died. Nowadays, thanks to different investigations and procedures, the life of a patient with hypoplastic left heart syndrome is not only able to survive beyond one month of age, but also has a high probability of reaching adulthood.[2] However, this does not take away from the fact that the main surgical interventions that are performed are extremely invasive and complex, added to the fact that later it is highly probable that the patient will present certain complications both due to the surgery itself and due to the physiology that he or she presents.

Clinical case

We present the case of a pregnant woman with a 35-week fetus referred for suspicion of fetal heart malformation. The fetal echocardiogram showed severe hypoplasia of the left ventricle (Figure 1). A very small donut-shaped left ventricle with severe mitral valve stenosis is observed. The patient was managed by being referred to a tertiary center where the diagnosis of LVHS was confirmed and he remained under control at said center until birth. A 39-week-old newborn was born at the tertiary level, small for gestational age, and the diagnosis of LVHS, mitral-aortic hypoplasia, accentuated aortic arch hypoplasia was confirmed. His parents decided not to operate on their son and to have compassionate management due to the severity of the cardiac malformation.

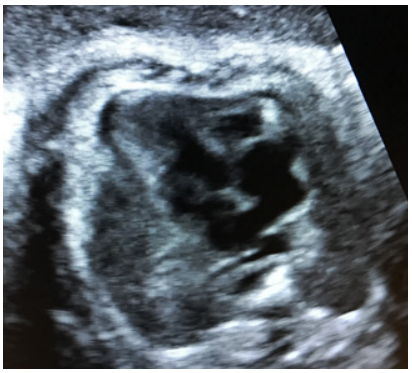

Figure 1 The image shows a left ventricle with severe hypoplasia and the mitral valve with severe, almost atretic stenosis.

Discussion

Fetal left ventricular hypoplasia syndrome encompasses a large number of congenital malformations and is associated with malformations of the left heart components such as the mitral valve, aortic valve, aorta and left ventricle. Examples of these malformations are mitral atresia with aortic atresia, in which there may be no identifiable left at all, and mitral stenosis with aortic atresia or stenosis, in which a small but inadequate left ventricle is present. In addition, atrioventricular septal defects and even double outlet right ventricles with a small left ventricle may be found. All of these conditions have in common the inability of the left ventricle to fulfill its natural function of providing systemic perfusion.[1,3]

Congenital heart defects have an incidence rate of 8 out of 1000 live births, while HLSH is found in 2 out of 10,000 pregnant women, which contrasts with its extremely low incidence rate. Despite this, it

is responsible for 23% of cardiovascular deaths during the first week of life and 15% of cardiovascular deaths in the first month. 7 out of 10 cases correspond to men.[2]

The fetus with LVHS is very stable. Intrauterine death is very rare, but if it occurs, it is usually associated with a genetic or chromosomal abnormality. The single right ventricle of LVHS usually functions well and drives the combination of fetal systemic venous return and placental umbilical venous return through the fetoplacental circulation.[3,4]

Nowadays, the vast majority of LVHS are detected before birth, and their presence is usually suspected in the obstetric ultrasound at 20 weeks of gestation. The 4-chamber view easily reveals the abnormality.[3,4]

There are situations where babies with LVHS are born without being diagnosed, managing to remain stable for a time, while pulmonary vascular resistance remains high and the ductus arteriosus remains wide open. However, as pulmonary vascular resistance decreases, more blood is diverted through the pulmonary vascular bed, causing tachypnea, relative hypotension, and acidosis. This, together with the progressive closure of the ductus arteriosus, further compromises systemic circulation.[3,4]

Without intervention, LVHS is lethal. Through a series of three complex surgeries consisting of the Norwood operation in the first days of life, the Glenn shunt within 4 months after the first surgery and finally the Fontan operation, survival is now possible in 63-74% of patients during the first year of life.[1,2]

The Norwood operation consists of three consecutive procedures; an atrial septectomy, reconstruction of the aorta using tissue from the pulmonary artery and the Blalock-Taussing (BT) shunt that allows pulmonary perfusion. The objective of the Norwood operation is to ensure that the right ventricle carries out correct perfusion at the systemic level, of the neoaorta and the lungs, the latter by means of BT.[1,3]

Three months after the Norwood operation and with the patient constantly growing, the Blalock-Taussing shunt is replaced by a direct connection between the superior vena cava (SVC) and the branches of the pulmonary arteries, a process known as Glenn shunt.[1,3,5]

Finally, during the preschool period, the Fontan operation is performed, which consists of connecting the inferior vena cava to the pulmonary arteries in order to divert all systemic deoxygenated blood to the lungs by means of changes in intrathoracic pressures.[2,6]

It is important to mention that the Fontan circulation itself imposes long-term consequences on the patient with single ventricle LVHS. It has been observed that Fontan patients present multiple comorbidities, among which the most important are protein-losing enteropathy (PLE), cardiac complications, where we find arrhythmias, heart failure and pulmonary hypertension and the main complication of the Fontan circulation, which is liver dysfunction.[1,2,6]

Cardiac: Arrhythmias, mostly intra-atrial and reentrant, which are an independent risk factor for late Fontan failure and reduce survival to 70%. Heart failure usually occurs due to late sequelae of aortic reconstructions which can increase afterload on the systemic right ventricle, potentially contributing to heart failure and ischemia.[2,7]

Hepatic dysfunction: Fontan surgery causes a large amount of systemic venous congestion, where the liver, located between the pulmonary and splenic vascular beds, acts as a reservoir, resulting in fibrosis and sinusoidal dilation. In fact, almost all adult patients develop Fontan-associated liver disease, characterized by hepatic fibrosis and associated with cirrhosis, hepatocellular carcinoma and failure of Fontan physiology.[2,8] Other less common complications are thrombotic events caused by hepatic venous congestion and endothelial dysfunction; and plastic bronchitis, which is extremely rare with a prevalence of 4-14% but with a transplant-free survival of approximately 8 years.[2]

Conclusion

Left ventricular hypoplasia syndrome is a rare disease that requires complex, non-curative treatment. Prenatal diagnosis is vitally important in this disease, as it will allow referral to a tertiary level for confirmation, initial management and surgical treatment, thus giving the patient the best treatment conditions and achieving good surgical results and low early and late complications. In this case, due to the complexity and severity of left ventricular hypoplasia syndrome, the family opted for compassionate management.

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Conflicts of interest

The authors declare there is no conflict of interest.

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