Case Report

Imaging of hypoplastic left heart syndrome –A rare antenatal cardiac anomaly

Dr. Gurivi Reddy.C [1] , Shrinuvasan.S*[1], Bruntha.D[1] , Chidambaram.R[1]

[1]Department Of Radiology , Sri Lakshmi Narayana Institute of Medical Sciences, Agaram Village, Affiliated to Bharath University, Chennai, Puducherry- 605502, India.

Corresponding author: Dr.S.Shrinuvasan, Assistant Professor

Abstract

A group of closely related congenital cardiac anomalies like underdeveloped left heart chambers, atresia or severe stenosis of aortic orifice and hypoplasia of ascending aorta constitutes Hypoplastic left heart syndrome (HLHS). Ultrasound and fetal cardiac echocardiography play an important role in detecting HLHS prenatally. HLHS can be detected by 18 – 22 weeks of gestation by USG. It is the most common cardiac malformation detected in fetal life .Here we present a case of the second gravida, came for anomaly scan, found to be carrying a fetus with the Hypoplastic left heart without any other anomalies.

Keywords: Hypoplastic left heart syndrome, congenital heart disease, Ultrasonography

Introduction

Hypoplastic left heart syndrome (HLHS) is defined as congenital hypoplasia of left ventricle in association with mitral atresia , aortic atresia , aortic stenosis and coarctation of the aorta. HLHS can be easily detected in the antenatal screening of 4 chamber view of fetal heart by 18 – 22 weeks of gestation. Nearly 4% of congenital heart diseases are due to HLHS and about one quarter deaths occur within 1 year of delivery [1]. In HLHS the underdeveloped left ventricle unable to support the systemic circulation creating pressure over the right ventricle .The ventricle takes over the function of underdeveloped left ventricle via a shunt through the ductus arteriosus resulting in ductus dependent systemic circulation.The pulmonary venous return is through patent foramen ovale resulting in mixing of both oxygenated and deoxygenated blood leading to cyanosis. Neonates die within days after birth due to the closure of foramen ovale resulting in cardiac failure due to inability to maintain systemic circulation.

Case report

31-year-old second gravid women with 20 weeks gestational age referred from obstetrics and gynecology department for fetal well-being and to rule out fetal anomalies. Her first pregnancy was uneventful and had full term normal vaginal delivery. No congenital anomalies detected in the first child. No significant family history recorded. she is a nondiabetic and nonhypertensive. No significant past medical and surgical history. Obstetric scan at 11 weeks shows no abnormality.

Present ultrasonography done at 20 weeks of gestation revealed hypoplastic left atrium and ventricle. Rest of the ultrasonographic examination showed normally grown fetus with no extracardiac manifestations.

Fetal echo revealed a 4 chamber asymmetry with altered axis and orientation. Hypoplastic left atrium

and left ventricle with dilated right sided heart noted. noted.
Hypoplastic LVOT with arch formed by the ductus

The mother and her attendants were informed about the poor prognosis of the baby by the gynecologist and advised for termination of pregnancy.

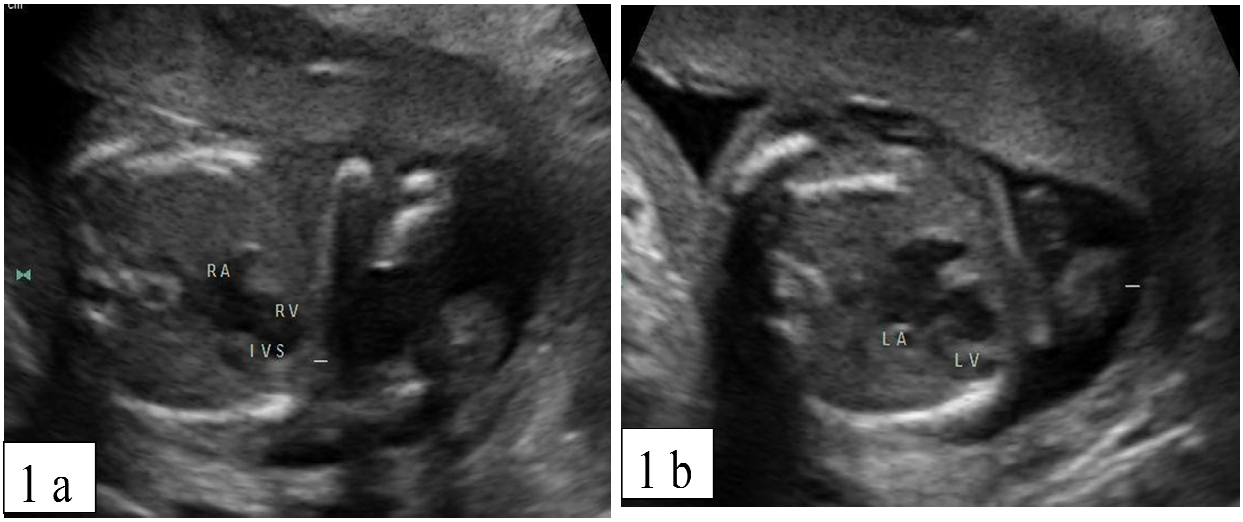

Figure 1: Fetal echocardiogram images **(a)** showing cardiac chamber asymmetry with dilated right atrium and right ventricle with displaced interventricular septum towards the left side and **(b)**showing hypoplastic left atrium and the left ventricular chamber.

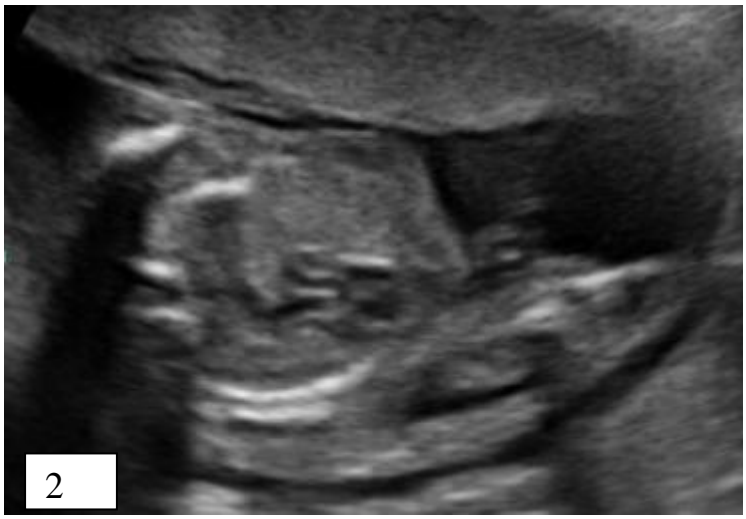

Figure 2: Fetal echocardiographic image showing three vessel view with hypoplastic left ventricular outflow tract(LVOT) and the arch formed by ductus.

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Discussion

Hypoplastic left heart syndrome (HLHS) , the most severe congenital cardiac anomaly is characterized by hypoplasia or complete atresia of ascending aorta, aortic valve ,left ventricle and mitral valve . Incidence is 1 – 3 in 10,000 live births and males are affected twice more commonly than females Recurrence is seen in 0.5 % [2,3] . HLHS /left sided obstructive cardiac lesion leads to cyanosis and severe cardiac failure within the first week of life.

Due to hypoplasia or complete atresia of ascending aorta , entire cardiac output enters a dilated pulmonary trunk and supplies to the systemic circulation via large ductus arteriosus. Blood flow in the aortic arch and hypoplastic ascending aorta is in the retrograde direction. This leads to congestive heart failure due to increases blood flow in the pulmonary arteries.

Extra-cardiac defects frequently associated with hypoplastic left heart includes two-vessel umbilical cord, and craniofacial, gastrointestinal, genitourinary and central nervous system abnormalities [4]. The risk of aneuploidy associated with fetal cardiac anomalies is much greater (ranging from 13 to 32%) than that associated with advanced maternal age [5]

Fetal ultrasonography with 4 chamber view plays an important role in diagnosing Hypoplastic left heart syndrome. The sensitivity of sonographic detection of isolated left heart syndrome is reported as 61.9% in a study wherein cardiac defects affecting the size of the ventricles had the highest detection rate [6].

HLHS has an extremely poor prognosis with 25% of mortality in the first week of life and if untreated infants die in 6 weeks[7]. Prenatal diagnosis of HLHS is beneficial for preventing ductal shock and keeping affected infants stable in the preoperative stage[8-10]. Monophasic blood flow across the mitral valve, restricted flow through the foramen ovale, and retrograde flow through the aorta are all considered poor prognostic signs in utero. Several palliative procedures including atrial septectomy, banding of the pulmonary artery, and the creation of aortopulmonary shunt have been used hoping for a better prognosis [11,12]. In the management of hypoplastic left heart syndrome, no obstetrical interventions are needed during pregnancy [13], apart from determining the karyotype and investigating of associated anomalies . There are many surgical procedures to correct HLHS, most important among them are Norwood procedure, Bi-directional Glenn shunt procedure, Fontan procedure. Inspite of these surgeries,infants have life-long complications.

In our case, anomaly scan and fetal echocardiography done at 20 weeks of gestational age shows asymmetry of cardiac chambers with dilated right ventricle and right atrium and hypoplastic LVOT. The patient has advised termination of pregnancy without undergoing karyotyping.

Conclusion

Antenatal anomaly scan plays an important role in early detection of fetal congenital anomalies specially of the cardiac origin like Hypoplastic left heart syndrome group of disorders. Thus, helping us in providing the patients with the option to either continue with the pregnancy or terminate it to prevent a bad outcome.

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