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A case of antenatally diagnosed hypoplastic left heart syndrome

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Abstract

The term hypoplastic left heart syndrome describes a diminutive left ventricle with underdevelopment of the mitral and aortic valves.

Because of its small size, the left ventricle is incapable of supporting the systemic circulation. There is hypoplasia of the ascending aorta and the aortic arch.

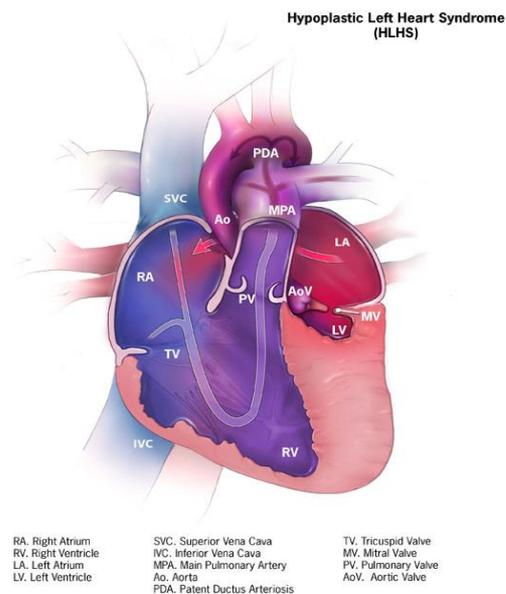
Prevalence: varies between 0.21 and 0.28 per 1000 live births. 4, 5 with male preponderance. Infants with this disease presents early in life with tachypnea and cyanosis.

Diagnosis is done by two dimensional echo-cardiography. Afflicted children require surgery as neonates, as they have duct-dependent systemic circulation. Surgical therapy for hypoplastic left-heart syndrome is associated with improving survival rates, reported as high as 90-95% for the 1st-stage palliation in experienced centers.

Keywords: antenatally diagnosed, hypoplastic heart, surgical therapy

Introduction

The term hypoplastic left heart syndrome describes a diminutive left ventricle with underdevelopment of the mitral and aortic valves [1]. Because of its small size, the left ventricle is incapable of supporting the systemic circulation. There is hypoplasia of the ascending aorta and the aortic arch [2].



The left atrium is small reflecting the limited blood flow in utero. The atrial septum is thickened; the foramen ovale may be small and, occasionally, may be closed. A patent ductus arteriosus is required for survival.

As the arterial duct closes, the systemic perfusion becomes decreased, resulting in hypoxemia, acidosis, and shock. Hypoplastic left heart syndrome is also associated with extra cardiac anomalies like of the brain (holoprosencephaly, agenesis of corpus callosum) Prevalence: varies between 0.21 and 0.28 per 1000 live births [4, 5]. With male preponderance. Infants with this disease presents early in life with tachypnea and cyanosis. Diagnosis is done by two dimensional echo-cardiography.

Case Report

A male baby with birth weight 2.5kg was born by normal vaginal delivery, cried immediately after birth, with H/O antenatally performed USG at 33wks of gestation suggestive of asymmetry in the cardiac chambers; left atrium and ventricle appeared to be small. This was again confirmed at 37wks of gestation. The baby was shifted to NICU with APGAR score of 6 at 1 and 8 at 5mins respectively for observation and further investigation. In the NICU, saturation was recorded as 100% at half hour of birth.



On Examination

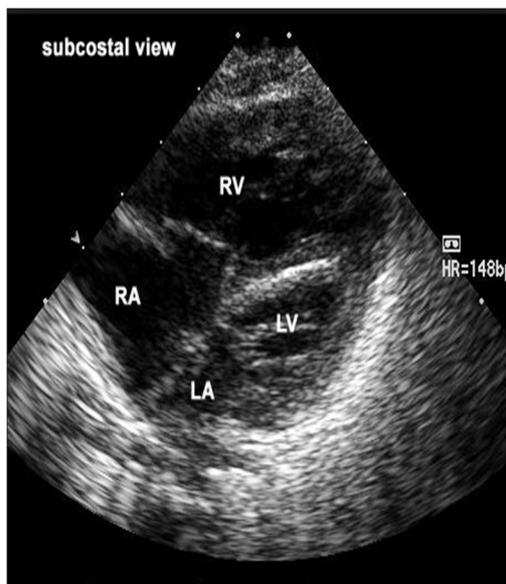
VITALS
 Temperature- 98.8°F
 HR 146bpm
 RR 52/min
 Mean BP- 38mmHg
 CRT = 3secs, PP felt
 No cyanosis, pallor
 BSL- 102mg/dl
 urine passed within 6 hour of life

SYSTEMIC EXAMINATION
 Respiratory: Air entry heard bilaterally.
 Cardiovascular: S1 S2 +
 Abdomen: Soft, Bowel Sounds +
 CNS: Tone, suck, cry, reflexes
NORMAL

At six hour of life the child developed distress in the form of intercostal retraction and nasal flaring and child was not maintaining saturation, SpO2 on O2 was 82-85%. The child further deteriorated and was put on mechanical ventilation.

2 dimensional echo-cardiography was done s/o 6mm ASD, left to right shunt, hypoplastic mitral valve and left ventricle, aortic atresia, severe coarctation of aorta, severe pulmonary artery hypertension.

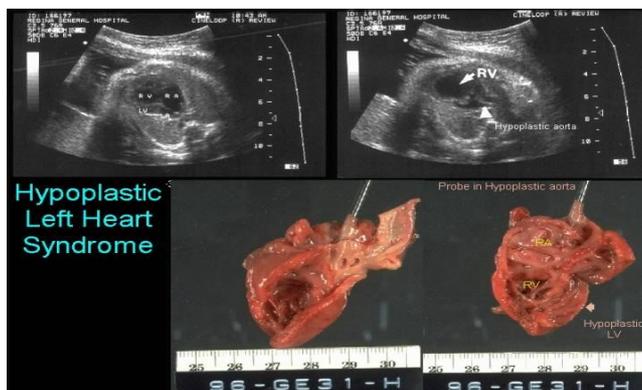
ABG: pH 7.2, PO2 35, PCO2 40, HCO3 15



Treatment Strategies

Prevention of hypothermia
 Double lumen UVC
 IV Fluid as per NICU protocol
 IV Dobutamine
 PGE2 was considered but could not be given due to affordability issues. USG cranium and abdomen revealed no abnormality. We did not have a neonatal cardiovascular surgery center. Twelve hours after birth, the baby died due to cardiopulmonary failure. The parents refused autopsy on the fetus. Our patient didn't follow the routine course of hypoplastic left heart syndrome.

Antenatal Diagnosis



Vaginal delivery is often recommended, as long as the foetus has no signs of cardiac failure. Most health care professionals advise that the birth of the infant should occur in a cardiac surgical centre.

Discussion

Hypoplastic left heart comprises 7% to 8% of symptomatic heart disease in the first year of life [8] and is responsible for 25% of cardiac deaths in the first week of life [9]. A conclusion underscored by an average lifespan of only 5 to 14 days [10-12]. Chromosomal aberrations account for about 6% of all congenitally malformed hearts. Many genetic and hereditary diseases are associated with such congenital malformations, although the causative mechanism is

unknown. Precarious survival depends on three tenuous variables: patency of the ductus arteriosus, pulmonary vascular resistance, and an adequate interatrial communication [12]. Tachypnea, tachycardia, and cyanosis are present during the brief interval of ductal patency [13]. Risk is greatest during the period of normal ductal closure when systemic blood flow and coronary blood flow decrease or cease altogether. A fall in pulmonary vascular resistance diverts blood from the systemic circulation into the pulmonary circulation and augments flow into the obstructed left atrium. A rise in pulmonary vascular resistance improves systemic blood flow, but at the price of hypoxemia. Ninety-five percent of afflicted infants die within the first month of life [9, 14]. Fetal echocardiography permits the diagnosis as early as the 24th week of gestation [16]. Flow patterns in the fetal ductus can be monitored, [17] and the condition of the atrial septum can be determined. Echocardiography with color flow imaging and Doppler interrogation establishes the diagnosis of hypoplastic left heart with aortic atresia and a hypoplastic but perforate mitral valve [15]. A hypoplastic but perforate mitral valve communicates with a small left ventricle that gives rise to an atretic aortic valve and a tubular hypoplastic ascending aorta. The ventricular septum and the free wall of the hypoplastic left ventricle are thick and immobile, and the small cavity is lined with endocardial fibroelastosis. Coarctation is identified as a thin discrete posterior ledge extending across the lumen of the aorta at the level of the ductus arteriosus or as kinking and narrowing at the site of ductal insertion. Doppler interrogation establishes retrograde flow into the hypoplastic ascending aorta and occasionally identifies biphasic flow in the proximal coronary arteries. Direct ventriculo-coronary arterial communications can be identified.

Management

Afflicted children require surgery as neonates, as they have duct-dependent systemic circulation. Currently, there are two major treatment modalities. These are primary cardiac transplantation, or a series of staged functionally univentricular palliations [18]. Functionally univentricular palliations: The functionally univentricular palliation typically includes three operations.

1. The first stage of palliation, or the Norwood operation, is performed at birth.
2. The second stage is a bi-directional Glenn operation, usually undertaken at 6 to 8 months of age.

The third, and final, stage is the Fontan operation, which can be performed between the ages of 18 months and 4 years. For patients undergoing functionally univentricular palliation, leading to creation of the Fontan circulation, the highest risk of mortality is following the initial operation, with up to three-tenths of patients dying in some reported series.¹⁹ The Norwood operation consists of constructing a new aortic root and arch, disconnecting the pulmonary trunk from the pulmonary circulation, and incorporating it into the systemic outflow tract. A modified Blalock-Taussig shunt, of 3 to 4 millimeters in diameter, is constructed to supply blood to the lungs. Nowadays, the operative mortality for conversion to the Fontan circulation is also less than 5%. Cardiac catheterization is usually undertaken prior to both the second and third stages of palliation to study anatomical and physiological details, and to perform corrective interventions

Prognosis and Outcome

Untreated patients most often succumb during the 1st few months of life, usually during the 1st or 2nd wk. Occasionally, unoperated patients may live for months or, rarely, years. Up to 30% of infants with hypoplastic left-heart syndrome have evidence of either a major or minor central nervous system abnormality. Surgical therapy for hypoplastic left-heart syndrome is associated with improving survival rates, reported as high as 90-95% for the 1st-stage palliation in experienced centers.

Genetic Counselling

Upon diagnosis, both genetic counselling and testing should be offered to both parents. Multiple genetic syndromes have been reported, including Turner's syndrome, Noonan's syndrome, Smith-Lemli-Opitz syndrome, Holt-Oram syndrome, and many others (20, 21). Therefore genetic counselling forms an important part of it.

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