Original Article

Hypo-Plastic Left Heart Syndrome (Hlhs) : Norwood Surgery

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INTRODUCTION

A genetic condition known as the hypoechoic left atrial syndrome makes it difficult for the heart to pump effectively, often known as HLHS. One of the most difficult disorders is hypoplastic left heart syndrome. cardiac conditions that may develop in a newborn (HLHS). It is one of the most challenging congenital cardiac abnormalities to cure. It is one of a number of cardiac malformations classified as single ventricle defects. The Centers for Prevention and Control of Diseases predict that In the US, 1,025 infants are born each year with hypoechoic left heart malfunction (CDC). This means that 3,841 of every 100,000 births in the US each year develop benign granulomas left heart disease. Drawing oxygenated blood from the lungs and delivering it to the body is the responsibility of the "left heart" area of the heart. Warmed blood passes through the mitral valve and into the left ventricle before from the lungs reaches the healthy heart's left atrium (higher collecting chamber) (lower pumping chamber). The left ventricle releases this blood through the arteries either through the aortic valve, from which it is carried by the arterial network to every area of the body. The blood picks up oxygen as it passes via the body's tissues and organs. Prior to passing through tricuspid valve and reach the intended ventricle, this oxygen - poor right atrium of the heart once again. The pulmonary circulation transports this blood first from heart to the lungs. system, where it is reoxygenated. pulmonary artery valve. The cycle of this oxygenated blood starts when it enters the left atrium anew.

History of Hypo-plastic Left Heart Syndrome

Lev first referred to the congenital heart abnormality as In 1952, it was first referred to as hypoplasia of the thoracic tract complex; it is now more often known as agenesis left heart condition (HLHS). After many specimens were investigated, it was discovered that they had solitary aortic hyperkeratosis, with or without vsd and aortic ectasia Hypoplasia of the arch with or without cardiogenic shock or atresia, and aortic hypoplasia with or without aortic stenosis or an atrial septal defect. After Lev's series, Noonan and Nadas conducted their study. They coined the phrase "hypoechoic left heart syndrome" in 1958 to refer to their assortment of samples with various left-sided cardiac issues..

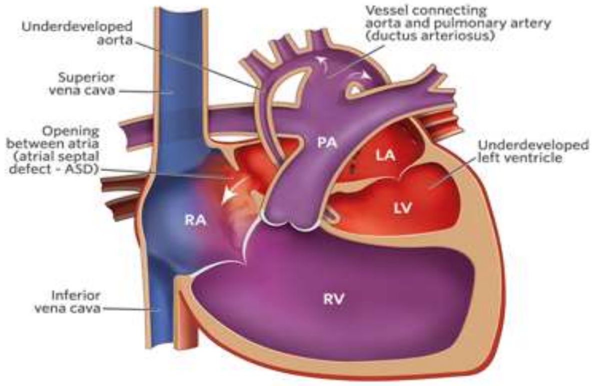
What is Hypo-plastic Left Heart Syndrome?

The congenital defect known as hypoplastic left ventricular syndrome disrupts the heart's typical blood flow (HLHS) (HLHS). The left half of the heart is still forming as the foetus is growing. grows during pregnancy. One kind of congenital heart abnormality is hypoplastic left heart syndrome. Congenital means existing at conception. HLHS is regarded as a serious congenital heart abnormality since a newborn with this condition requires procedures performed immediately after birth (CCHD).

Several of the heart's left lateral chambers underdeveloped structures are impacted by the hypo-plastic left heart syndrome, including:

- The left ventricle, which is undersized and undeveloped.
- The mitral valves are either absent or very tiny.
- There is no aortic valve or there is very little.
- The ascending aorta is abnormally small or has an undeveloped ascending segment.
- A Infants with short stature Here between left and right heart chambers, frequently exists a hole. in people with left heart syndrome. chambers (atria).

What happens in hypo-plastic left heart syndrome.



Through an atrial septal defect, blood first from lungs is transferred from the left atrium to the right atrium. As it enters the right ventricle, the blood that has lost oxygen joins it. This mixed blood with oxygen is discharged from the heart via the pulmonary artery. While some of this blood travels to the lungs, the bulk comes into the body via the aorta's patent ductus arteriosus (PDA). All foetuses have the arterio, a blood flow that runs from the vena cava to the aorta. If the frontal sinus ovale and the artery shut, as they regularly do in the first days after birth, Blood cannot be pumped every region of the body, starting with the right side of the heart. Prior to surgery, infants with issues can be addressed left heart malfunction must take medication cardiac surgery to keep these different approaches and maintain blood flow to the body.

What are the Causes & Risk Factors of Hypoplastic Left Heart Syndrome?

An issue with the baby's development during pregnancy leads to HLHS. Boys are more likely than females to experience it. The following genetic disorders have been linked to HLHS:

- Noonan syndrome
- Turner syndrome
- Trisomy 21
- Holt-Oram syndrome
- Trisomy 13
- Smitp-Lemli-Opitz syndrome
- Trisomy 18

Some families are genetically predisposed to HLHS development. So far, some HLHS instances have been found to contain mutations in the genes NOTCH1 and NKX2.5.

Other danger factors can be:

- Medications consumed during pregnancy
- Certain environmental elements that have an impact on the expectant mother
- Diet of the mother during pregnancy

Signs and symptoms of HLHS

At birth or a few days later, the following signs of hypoplastic left core syndrome could be present:

- Blue color of the skin, lips, and nails (cyanosis)
- Pale skin
- Sweaty, clammy, or cool skin
- Trouble breathing
- Fast heart rate
- Cold feet
- Poor pulses in the feet
- Poor feeding

Diagnosis of Hypoplastic Left Heart Syndrome

Healthcare professionals use non-invasive, painless imaging techniques during pregnancy, including:

- **Ultrasonography, foetal echocardiography to assess the baby's heart before birth.**

During a procedure, a device known as a transducer emits high-frequency sound pulses into the abdomen. ultrasound or ultrasonography. High-frequency sound waves are sent by the transducer into the body, where they bounce off the baby's internal organs. The transducer picks up the sound waves or echoes that return and converts them into images that appear on a screen. It is possible to print these images.

- Chest X-ray: This displays the Baby's heart and lungs: size and structure.
- Echocardiogram: This ultrasound shows internal heart structures.
- An electrocardiogram (EKG) analyses the electrical alterations that occur during a heartbeat.

• Pulse oximetry testing: This determines the amount of oxygen in the baby's blood.

Treatment
A heart transplant or a variety of surgical techniques may be applied to treat left heart hypoplastic condition.

Medication. The drug alprostadil (Prostin VR Pediatric) keeps the ductus arteriosus open and aids in blood vessel expansion.
• **Breathing support.** A breathing machine may be necessary for infants who are having respiratory difficulties (ventilator).
• **Tube for feeding.** Feeding tubes can be used to feed infants who struggle to eat or who become fatigued while doing so.
• **The atrial septostomy.** In order to facilitate greater Through this surgery, an opening between the two upper chambers of the heart is made or enlarged. by allowing blood to flow first from atrioventricular node to the left atrium. This is carried out if the sinus ovale closes or shrinks too much. If a newborn has a gaping wound in their heart (called an atrial septal defect), they might not need this treatment.

Surgery
1. The Norwood Procedure
Either a heart transplant or the three-step surgical procedure known as phased palliation are required for therapy for the condition of left heart hypoplasia. Among the most popular forms of important advancements in congenital heart surgery in recent years.. At age 5, children have a survival rate of almost 70%, and the majority of these kids grow and develop normally. This three-step surgical technique aims to restore regular heart in order that the body may get the air blood it needs, blood flow requires.

The Norwood Procedure's past
For the treatment of HLHS or other heart conditions affecting one working ventricle, Fontan and Baudet created a procedure in the 1970s that is being performed today. Norwood recorded the first occurrence of palliative surgical treatment for HLHS. et al. in 1980.The first newborn heart transplant for HLHS was successfully accomplished in 1986 by Bailey et al., providing an alternative to the palliative reconstructive strategy.

Completely new systemic circuit in means (e.g. left heart syndrome patients is created using the three-stage Norwood technique. Atrial septectomy, transection, and closure of the Stage includes the distal major pulmonary artery in each case 1 of the Norwood surgery.

Stage 1
Soon after delivery, the operation is begun. It transforms the brain into the primary ventricle, which circulates blood to the body's organs and lungs. both thoracic arteries branches that would normally provide blood to the lungs on each side of the primary pulmonary artery are separated from one another. side by the aorta. An alternative is the placement of a shunt in the gap between aorta and indeed the pulmonary arteries.. The lungs get blood as a result..

Stage 2 (Bi-directional Glenn Operation)
This procedure to redirect half of the alveoli into the circulation is often carried done six months following the Norwood. once the blood moves through the organs no longer necessitates as much tension from the ventricle. The right atrium artery, which no longer has a The vena cava, the blood channel that delivers anaemic blood to the shunt here to the pulmonary arteries and the flow of the blood from the body into the heart. Bypassing the ventricle, the deoxygenated blood is sent directly to the lungs.
Stage 3 (Fontan Operation)
This final phase is often finished 18 to 36 months after Glenn. It creates a pathway into or just beyond the chest for blood to get to the respiratory system by connecting the pulmonary vein to the vena cava, the principal blood artery that transfers anoxic blood from the lower body to the atrium. The whole bloodstream is now passively going to the lungs.

Prognosis

According to the UK's National CHD audit, the Norwood, Glenn, and Fontan surgeries have 30-day post-operative survival rates that are higher than 90%. Outside of the UK, recent five-year survival rates varied from 18.8% in New Zealand to 72% in Denmark.

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