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Diagnosis of Hypoplastic Left Heart Syndrome

By Liji Thomas, MD

Hypoplastic left heart syndrome (HLHS) is a serious congenital heart condition in which the development of the left ventricle is abnormal. The normal heart has four chambers, two upper atria, and two lower ventricles. The atria receive blood from the body and the lungs.

The ventricles are pumping chambers which pump blood to the body and to the lungs. The left atrium receives oxygen-rich blood from the lungs through the four pulmonary veins, and empties it into the left ventricle. This, in turn, pumps it through the aorta into the systemic circulation, supplying oxygen to the cells and organs of the body.

The returning oxygen-poor blood empties into the right atrium through the superior and inferior vena cavae, the great veins that drain the upper and the lower parts of the body respectively. The blood travels through the tricuspid valve into the right ventricle which pumps it into the pulmonary circulation to be oxygenated in the lungs. From there it returns to the left atrium and the cycle repeats itself.

The left and right sides of the heart are normally separated from each other throughout their functional activity.

A person with HLHS has an undeveloped and small left side of the heart. Therefore, the left ventricle is tiny and the mitral valve between the left atrium and left ventricle is too small. The aortic valve and the first part of the aorta are also undeveloped. The two atria may also be connected by persistent communications through the wall that should separate them.

Diagnosis before birth

Prenatal diagnosis is made on the basis of ultrasound examinations of the fetus during intrauterine life. Abnormal findings on the routinely scheduled ultrasound scan may prompt the doctor to ask for a fetal echocardiogram between 18 and 22 weeks of life. Here the structure and function of the heart are visualized by ultrasound waves. This will show the exact nature and extent

of the defect. Future courses of action must be determined by the parents, and include proceeding with the pregnancy with delivery planned to be in a specialist health center, or termination of pregnancy. Genetic testing is also offered, as well as evaluation for other genetic syndromes such as Turner or Holt-Oram, which are often associated with this.

Diagnosis after birth

Post-natally, the infant may appear normal at first. This is because all babies have one or two normal communications or sites where oxygen-rich blood from the left side of the heart mixes with oxygen-poor blood from the right heart. These include the patent foramen ovale, in the septum that separates the right and the left atria (the interatrial septum), and the patent ductus arteriosus, a connection between the aorta and the pulmonary trunk.

The presence of these left-right shunts allows some oxygen to reach the cells of the infant with HLHS, despite the poor function of the right side of the heart. Oxygenated blood from the left atrium flows into the right atrium and the right ventricle. From there, it is pumped back to the lungs. Some blood flows from the pulmonary trunk into the aorta and thence to the systemic circulation.

However, once these close off, as normally occurs within a few days of birth, the baby suffers a severe cut-off of blood flow into the systemic circulation and becomes extremely symptomatic. Most affected infants are at serious risk of immediate death unless treated with some form of palliative surgery at once, or by a heart transplant. They will be put on medication to prevent the closure of the ductus, and the interatrial foramen will be allowed to persist.

Clinical features

The symptoms and signs with which the infant may present include:

- Breathlessness due to pulmonary congestion
- Pounding heartbeat
- A weak pulse
- Poor feeding
- Lethargy
- Hepatomegaly or enlargement of the liver
- Cyanosis or bluish discoloration of the lips and the area around the mouth

- A heart murmur or abnormal heart sound

Investigations

If these signs are present, an echocardiogram will be requested which will delineate the anatomy and the working of the heart. Other tests may be performed if they are needed to clarify the diagnosis, or add more information which will help to manage the child better. Genetic counseling and testing are always offered, and the child must be evaluated for other anomalies as well.

Commonly required additional investigations include:

- A chest X-ray
- An electrocardiogram (ECG)
- Cardiac catheterization (the passage of a flexible thin tube or catheter into the heart chambers, to visualize the various orifices and chambers)

Reviewed by Yolanda Smith, BPharm

References

- http://www.cdc.gov/ncbddd/heartdefects/hlhs.html
- https://medlineplus.gov/ency/article/001106.htm
- http://www.rch.org.au/cardiology/parent_info/Hypoplastic_Left_Heart_Syndrome/
- https://www.bhf.org.uk/-/media/files/publications/children-and-young-people/c9understanding-your-childs-heart-hypoplastic-left-heart0210.pdf
- http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1877799/

Further Reading

- Hypoplastic Left Heart Syndrome (HLHS)
- Causes and Symptoms of Hypoplastic Left Heart Syndrome
- Treatment of Hypoplastic Left Heart Syndrome
- Prognosis for Hypoplastic Left Heart Syndrome

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