

What is Hypoplastic Left Heart Syndrome?

Hypoplastic Left Heart Syndrome (HLHS) is a name given to a developmental condition of the heart where the Left Ventricle is felt to be too small to support adequate cardiac output for the body. It is usually but not always accompanied by a very small Aorta which is the blood vessel that supplies blood to all of the body. Up until the mid 1980's this was felt to be a uniformly lethal condition and no surgical options were given to parents having such a child. Two areas of surgical advancement have changed our approach to this problem. The first was a surgery to combine the aorta and pulmonary artery in what has come to be known as the Norwood Procedure. The Second has been the advancement of heart transplantation in infants.

First it is important to understand the problem with HLHS. Normally there are two roughly equal sized ventricles in the heart. These are the pumping chambers. Normally the Right ventricle pumps blood to the lungs and the Left Ventricle pumps blood to the rest of the body. Normally the two vessels coming out of the ventricles are of equal size. In the case of HLHS the Aorta is usually very small as is the left ventricle. The problem is that not enough blood can be pumped out and through the aorta to supply the needs of the rest of the body. When the body does not receive enough blood (and nutrients and oxygen) the cells of the body begin to die. When we are first born there is a blood vessel called a patent ductus Arteriosus (PDA) that connects the aorta with the pulmonary artery. As long as this vessel is open then an adequate amount of blood will go to the body because the right ventricle is able to supply what the left Ventricle can't. Unfortunately this PDA can only stay open temporarily and usually closes within a few days. Medication can be given to keep the PDA open for several more days, but this is not permanent.

The Norwood Procedure is done shortly after the diagnosis is made, usually within the first week of life. In this operation the Pulmonary Artery and the Aorta are combined and the right ventricle becomes the pump for the body. Blood flow to the lungs is provided by a small tube (shunt) that comes from the new Aorta. This is the first stage. The second and third stages of the Norwood operation involve connecting the systemic veins (that normally drain the blue blood without oxygen back to the heart) directly to the pulmonary arteries (vessels that supply blood to the lungs). All of these operations are done within the first two years of life.

Heart transplantation involves putting a heart removed from a baby who is brain dead in place of the defective heart. The advantage to transplantation is that the new heart is normal and has two pumping chambers. The disadvantage is that there is a possibility of rejection (the body's immune system fights and kills the new heart) and so the patient will need to be on life long medications to suppress the immune system. This increases the possibility of infections and also cancer.

As you can see this is a very complex issue and your doctor will be happy to answer your questions.

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