

Dept of Neonatology

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HYPOPLASTIC LEFT HEART SYNDROME

What is hypoplastic left heart syndrome?

Hypoplastic means small or underdeveloped. Hypoplastic left heart syndrome is where all of the structures on the left side of the heart are underdeveloped to some extent.

What are the structures on the left side of the heart?

The normal heart is made up of four chambers: two atria and two ventricles. The atria are smaller than the ventricles. The atria receive blood and pump it through to the ventricles. The ventricles are the large pumping chambers of the heart. The heart has valves that help control blood flow from the atria to the ventricles and then to the main blood vessels leaving the heart. The mitral valve is situated between the left atrium and left ventricle. The aortic valve is situated between the left ventricle and the aorta. The left side of the heart receives oxygenated blood from the lungs which is then pumped to the body via the aorta. (The right side of the heart receives blood from the body to pump to the lungs to be oxygenated.) In hypoplastic heart syndrome the left ventricle, aortic valve, mitral valve and the beginning of the aorta are underdeveloped.



Hypoplastic Left Heart Syndrome

How is hypoplastic left heart syndrome diagnosed?

Hypoplastic left heart syndrome may be detected on an antenatal ultrasound, but may also be found after a baby is born. As the left side of the heart and aorta is underdeveloped it cannot provide the body with enough blood supply. The right side of the heart must try and pump for both sides of the heart. Blood to the body is dependent upon the ductus arteriosus (DA) remaining open. The DA is a vessel between the pulmonary arteries (arteries going to the lungs) and aorta which bypasses the left side of the heart The DA normally closes after the baby is born. The baby will often appear well in the first few hours to days of life as the DA remains open. When the DA closes the rest of the body can no longer get enough blood supply and the baby becomes shocked and ill. The baby's pulses will be difficult to feel and they will have a low blood pressure. An echocardiogram (ultrasound of the heart) will diagnose hypoplastic left heart syndrome.

How is hypoplastic left heart syndrome treated?

The most important initial step is to keep the DA open so that there is blood supply to the rest of the body. Your baby will be commenced on a drug called prostin to keep the DA open. One of the side effects of this medication is that it can make babies stop breathing properly. Your baby will be intubated (a tube placed into the airway) and attached to a ventilator to help them breathe while they are on this medication. Your baby may also be started on medications to help maintain their blood pressure.

Intravenous lines will be placed in the umbilical artery and vein. This allows us to monitor your baby's blood pressure, check the oxygen levels in your baby's blood and provide fluid and nutrition.

Hypoplastic left heart syndrome is a life threatening condition and requires specialised care. This care is unavailable at the Canberra Hospital. If your baby is diagnosed with this condition antenatally you will be referred to a paediatric Cardiologist (heart specialist) in Sydney who will discuss the options for treatment. Surgery for this condition is available only in Melbourne. If surgical treatment is an option the Obstetric staff here in Canberra will arrange for you to deliver your baby in Melbourne. If your baby is diagnosed after delivery he/she will be transferred to Melbourne after initial stabilisation, to be assessed by Cardiologists and cardiac surgeons. This will be arranged by the staff here at the Canberra Hospital in conjunction with the hospitals in Melbourne.

If you have any further questions please ask the medical and nursing staff.