Hypoplastic left heart syndrome

Information for parents

Koala – Cardiac Surgery
The Royal Children’s Hospital Melbourne
50 Flemington Road, Parkville,
Victoria 3052 Australia
Telephone +61 3 9345 5702
www.rch.org.au/koala

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Your child has been diagnosed with hypoplastic left heart syndrome (HLHS). This information aims to help you understand how we treat this complicated condition at The Royal Children’s Hospital (RCH), and also how it might affect your child.

Hypoplastic left heart syndrome is due to a collection of problems which together mean that the left side of the heart is too small to pump enough blood to the body. We therefore have to use the right pumping chamber instead to pump the blood to the body.

Children with this condition go through a series of at least three operations to get through to the Fontan circulation.

The diagrams in this brochure show the structure of the heart and pictures of the operations in more detail.

The three operations are:
1. **Norwood procedure**, performed in the first few days of life.
2. **Bidirectional cavo-pulmonary connection/shunt**, performed at about three months of age.
3. **Fontan completion**, performed at about 4–5 years of age.

The aim of the three operations is to ensure that there is no obstruction to blood flow to the body through the aorta and to supply blood to the lungs in the most suitable way as your child’s heart and lungs mature.

There may be modifications of these operations to suit your child’s specific condition. Some children require interim steps with additional surgical or catheter treatment to deal with heart problems. Unfortunately some children may suffer complications which cause injury to other organs such as the kidneys, liver, gut or brain.

**Norwood (stage 1)**

This is usually performed in the first few days of life and often is the most complicated operation. The diagrams above show what is done surgically. The blood supply to the lungs is through either an arterial shunt (artificial tube connecting an artery to the body and the lung artery) or a tube directly from the right pumping chamber to the lung arteries (Sano/RV-PA conduit).

This circulation is very different to normal and is quite unstable. The amount of blood flow to parts of the body is easily affected by minor changes in your child’s condition.

The goal is to get used to your baby; to be familiar with how he/she breathes, their colour and feeding habits. Our nurses and allied health specialists will support you through this process, as colour and heart rhythm may change with how your baby breathes. Therapists will work with you to help your baby to eat and to get ready to return to your baby to be discharged home at the earliest possible time. The diagram below shows what is done surgically. The changes in your child’s condition may affect the amount of blood flow to parts of the body.

**Normal heart and circulation**

The operations are performed in two stages. The first operation is called the Norwood procedure (stage 1). The second operation is called the Fontan procedure (stage 2) and is performed when your child is about 4–5 years old.
Between Stage 1 and 2

Having some time away from the hospital

If your baby is well enough, we believe there are important benefits to being discharged from the hospital to your home/local accommodation, for example babies may settle and feed better once they are out of hospital. You and your baby will bond better outside the hospital environment and there is less chance your baby will get a hospital-acquired infection.

The need for local accommodation is determined by what emergency services are close by to your home. The follow-up arrangements after discharge are described in an accompanying information brochure, but essentially there are frequent appointments to see the cardiologist and your nurse care coordinator, as well as other specialists if required.

Keeping a close eye on your baby

Your child will not be like other babies and it will take some time for you to be comfortable with what is normal for your child’s heart condition. For example, your baby may be sleepier and less active than other children due to lower oxygen levels and reduced heart function. They may also have difficulty feeding and lose weight and may experience heart function problems, which means you may need medical intervention at home to provide your baby with the best possible care.

Bidirectional cavopulmonary connection (BCPC/BCPS)

This next operation is typically done around three months of age. Before this operation, we will usually arrange an MRI scan to check the blood vessels and heart function to decide if there are any additional things that need to be done at the time of surgery. This operation changes the blood flow in your baby to a very much more efficient, stable and safe system. The blood flow to the lungs is now provided by the connection between the upper body veins and the lung arteries, since this is the body’s own blood supply, it is much less likely to become blocked.

Shunt card

Although your baby will require medicines to keep the blood thin, there is still a risk of the shunt blocking, particularly if your baby becomes dehydrated. If your baby is getting sick, prompt treatment is necessary and so we will give you a card to provide medical staff information on how to care for your baby in an emergency.

If you are worried about your baby during this period you should bring your baby to the RCH Emergency Department for us to make an assessment. It may be that we feel your baby is well and does not need admission but we would prefer that we have the chance to see you and your baby.
Fontan Completion

Fontan completion is usually done around 4-5 years of age and takes most of the blood coming back from their lower body and also connects this to the lung arteries. After this operation the majority of the blood flow to the lungs bypasses the heart completely and gets there without any pumping action. This can only work if the heart and lungs are working well.

In preparation for this operation we will most likely perform a cardiac catheter to check that your child will tolerate this operation. There is a small percentage of children who we may consider to be unsuitable to go through this operation.

Long-term complications

Due to the abnormal pressures in blood vessels due to the Fontan circulation, there are concerns about the functioning of other parts of the body too. Whilst these complications do not occur in all patients, they can be a feature of poor outcomes due to the Fontan circulation.

Learning and behaviour

This heart condition has effects on brain development even before birth. The brain is typically more immature than that of children born at a similar time who don’t have a heart problem. Further brain injury can occur as a complication of the operations and other procedures that are done for your child. The severity of limitation depends on the severity of injury. We know however that the neurodevelopment of your child may be different to other children, for example there is double the risk of reduced attention and ability to concentrate in children with complicated heart problems such as HLHS. It is important to be aware of these possibilities so that extra help can be sought if needed.

Further information

You may have other questions after you have read this brochure, and we encourage you to raise these with your cardiologist and nurse coordinator to discuss further.

Inferior vena cava

Attached to Gore-tex conduit

Superior vena cava and Gore-tex conduit attached to pulmonary artery

A fenestration (small hole) allows blood flow between right atrium and conduit

Fontan operation

Fontan operation with extracardiac conduit

Fontan procedure consists of creating new pathways for blood to flow through the heart, allowing the right atrium to bypass the right ventricle and flow directly to the pulmonary arteries. The procedure involves connecting the inferior vena cava to the pulmonary arteries, creating a new pathway for blood to flow from the lower body to the lungs.