



Hypoplastic left heart syndrome

Information for parents



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Murdoch Childrens Research Institute



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.

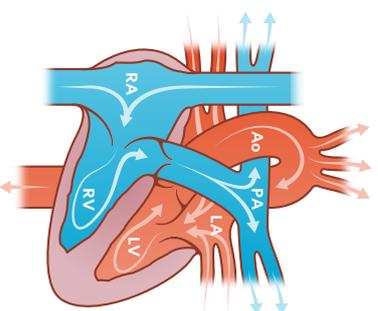
The three operations are:

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.

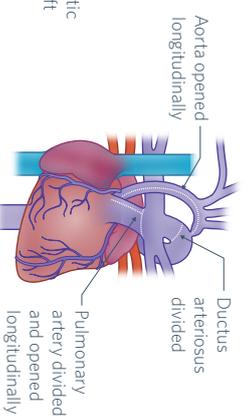
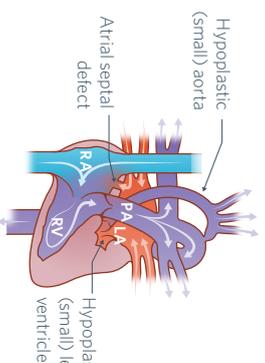
The aim of the three operations is to ensure that there is no obstruction to blood flow to the body through the aorta and to supply blood to the lungs in the most suitable way as your child's heart and lungs mature.

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.

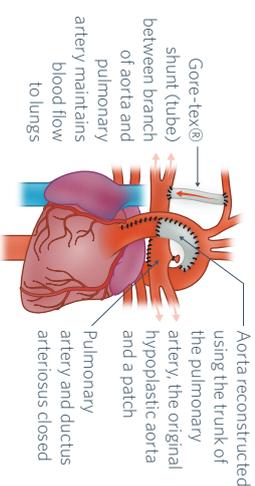
Normal heart and circulation



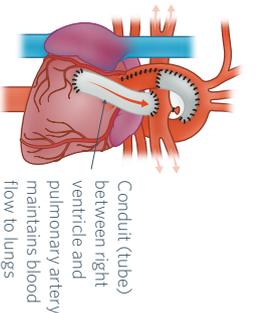
Pre-operative anatomy



Norwood operation



Norwood operation



Norwood operation (Sano)

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 such as dehydration, a high temperature or a chest infection. For these reasons we ask you to get used to your baby, to be familiar with how he/she breathes, their colour and their feeding habits. Our nurses and allied health specialists will support you through this process, as well as teach you how to give necessary medicines and use special feeding techniques or equipment if needed.

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 not be strong enough to take all their feeds by mouth and may need to be fed through a nasogastric tube.

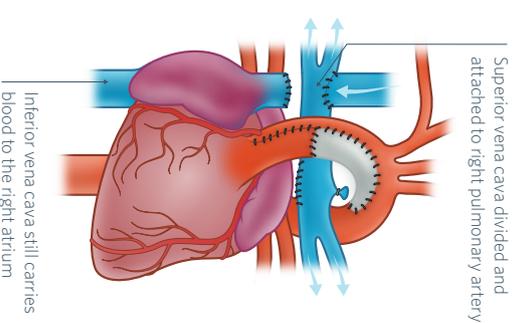
The period between Stage 1 and 2 is the most unstable and unsettling and this is when we worry most about your baby. The chance of rapid change, deterioration and death is usually highest in this period. It is the riskiest time because the heart is having to work much harder than usual and there is a possibility that the artificial tube supplying blood to the lungs can 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 as well as provide RCH contact details. Please try to keep this 'shunt card' with your baby at all times.

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.

Bidirectional cavo-pulmonary connection (BCPC)



Superior vena cava divided and attached to right pulmonary artery

Inferior vena cava still carries blood to the right atrium

Bidirectional cavopulmonary connection

(stage 2, 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 vein(s) and the lung arteries. Since this vein is the body's own tissue this is much less likely to become blocked. The heart does not have to work so hard and the circulation is much safer. Even though there are all these benefits with this type of operation in comparison with the artificial shunt in Stage 1, we have to wait for this time so that the lung vessels are mature enough for the blood to flow properly.

Fontan Completion (stage 3)

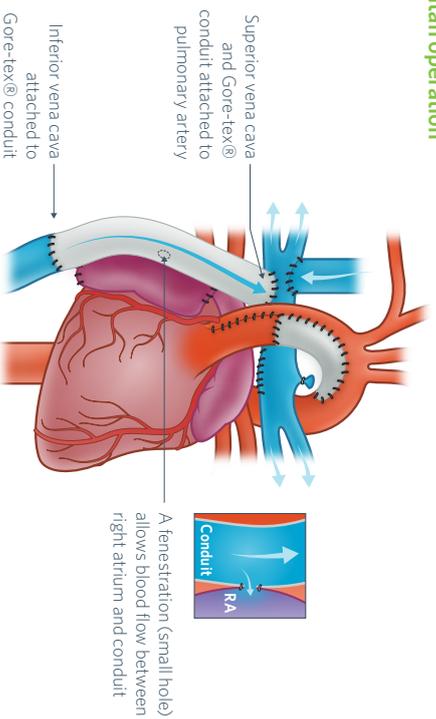
Fontan completion is usually done around 4–5 years of age and takes most of the blood coming back from their lower body and also connects this to the lung arteries. After this operation the majority of the blood flow to the lungs bypasses the heart completely and gets there without any pumping action.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

Physical activity

Most children who survive through to Fontan completion are able to take part in most activities. The majority of children go to mainstream schools with some children needing additional support in class.

Exercise capacity is typically lower than usual and whilst your child may not be able to take part in competitive sport we encourage them to take part in the usual physical education (PE) classes as much as possible. This has many physical but also psychological benefits by taking part with their classmates and peers. Certain medications and complications may mean that there are certain activities that your child should not take part in and so this should be discussed with your cardiologist.



Fontan operation with extracardiac conduit

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.

Longer-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 sign of poor future outcome if they do happen. These include problems with the gut (protein losing enteropathy), liver (scarring and cancer), kidneys and lung (bleeding and plastic bronchitis).

Unfortunately not all children survive through all three stages of surgical operations. The highest chance of dying is with the first operation and during the period between Stage 1 and 2.

As many as three out of ten children who have had a Norwood operation may die in this period. The cause of death is not always clear since it can happen abruptly and without warning. It is therefore important that you are used to the way that your baby behaves and come to the hospital for assessment if there is a change in behaviour.

The chance of dying with subsequent operations is lower but these are still important times of increased risk.

Overall, about four out of ten children will not survive through to ten years of age.

We are often asked if there is a normal life expectancy for patients with a Fontan circulation. Since this operation has only been performed since the early 1970s, it is difficult to be certain how effective the improvements in the way that we look after this complex condition will be in years to come. Due to current limitations however we believe that this patient group will not have a normal life expectancy.

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.