

# Hypoplastic Left Heart Syndrome

A booklet for parents and families of patients being managed at the Royal Children's Hospital, Melbourne, Victoria. Australia

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- **Please Note:** The sections titled 'Coming to terms with HLHS' & 'Coping with the Operations' were co-written by parents of children with HLHS and Social Work staff at the Royal Children's Hospital Melbourne.

The contents of this booklet may not be appropriate for patients being managed in other centres. Parents with affected children who are being managed elsewhere should take advice from their treating Cardiologist and Surgeon.

## Introduction:

### The normal heart:

Before discussing the abnormalities associated with hypoplastic left heart syndrome, it may be useful to describe what the normal heart does.

The heart's primary role is to pump blood around the body. In achieving this, the heart has two separate sides.

The right side of the heart pumps blood to the lungs and the left side of the heart pumps blood to the body.

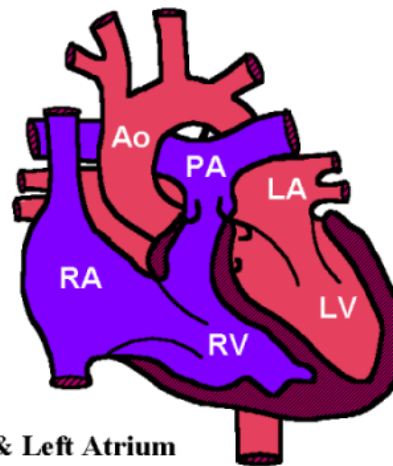
Because of this cooperation between the right and left sides of the heart, blood travelling around the body can take the following journey.

Once blood has given up its oxygen in order to nourish the body's tissues, it returns to the right side of the heart through veins. The millions of tiny veins gather into two large veins, the inferior vena cava (IVC), which drains all the blood from the lower half of the body and the superior vena cava (SVC), which drains blood from the upper half.

Blood from the superior and inferior cava enters a collecting chamber on the right side of the heart called the right atrium (RA). From there it is pumped into the right ventricle (RV), which is a muscular pumping chamber which pumps blood to the lungs where it can pick up oxygen. The right side of the heart

therefore has a primary role of receiving blood which has given up its oxygen in the tissues and transferring that blood to the lungs through the Pulmonary Artery (PA), where it can pick up more oxygen. Having picked up oxygen in the lungs, the blood then enters the left side of the circulation where the process occurs in reverse. In other words, blood drains back from the lungs into a collecting chamber on the left side of the heart, called the left atrium (LA), from where it enters the left ventricle (LV). The muscular left ventricular chamber then pumps the oxygenated blood out to the aorta (Ao) where it is then transmitted around the body through arteries. By this means oxygen is supplied to the body's tissues. The journey then begins again.

### Normal Heart and Arteries

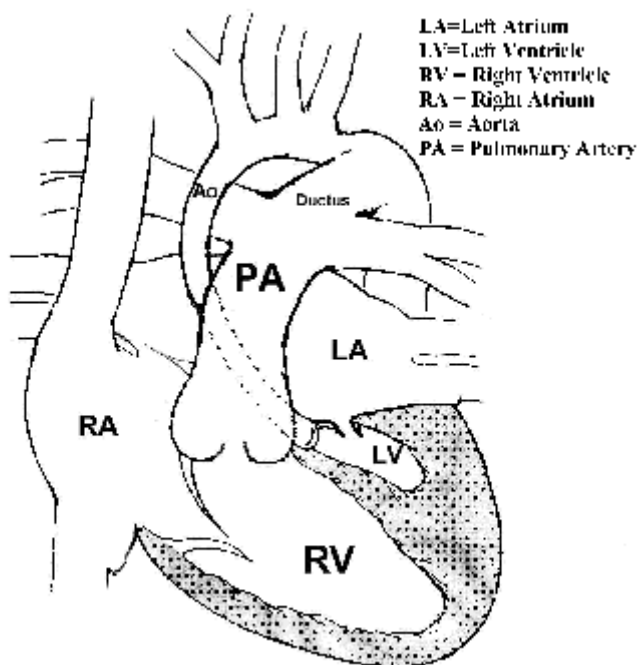


RA/LA = Right & Left Atrium

RV/LV = Right & Left Ventricle

Ao = Aorta; PA = Pulmonary Artery

## Hypoplastic left heart syndrome:



Hypoplastic left heart syndrome is one of the most serious forms of congenital heart defect. It basically describes the situation in which the left side of the heart is very poorly formed, so that it cannot support the circulation of oxygenated blood around the body. The left ventricle, which is the important pumping chamber on the left side of the heart is very small, as is the aorta, the main artery which arises from it.

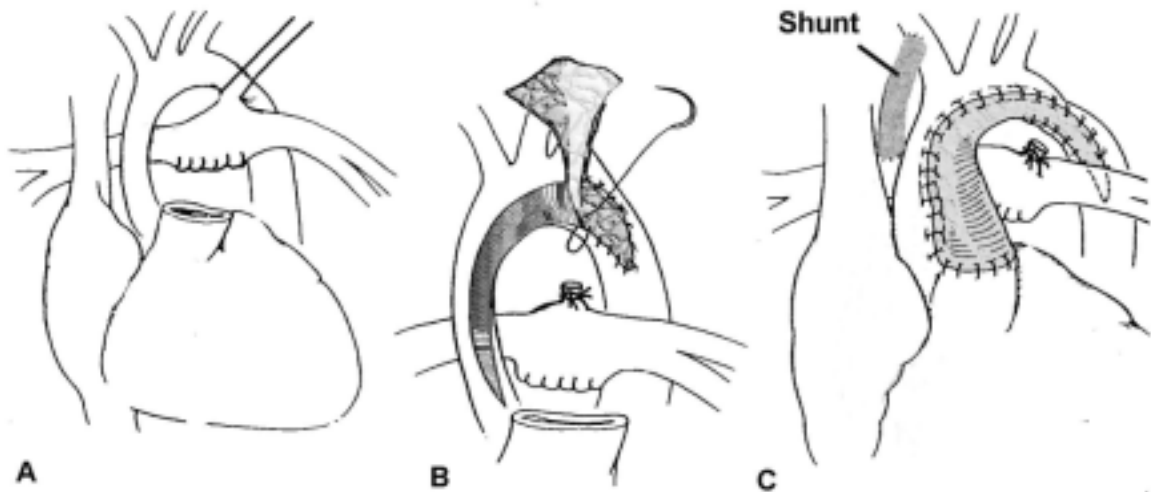
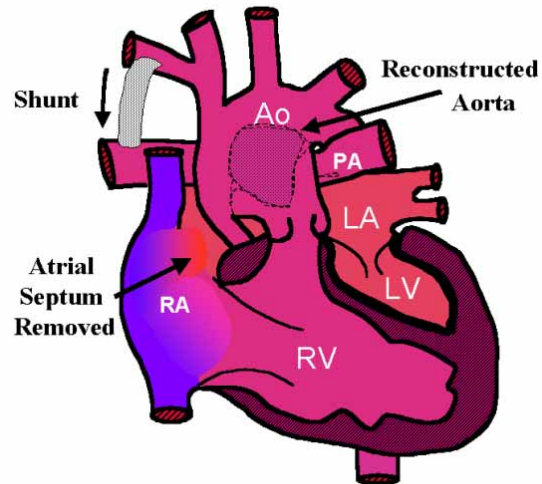
One question which many parents have is how a baby can be apparently well throughout the pregnancy and then rapidly deteriorates after birth. This is because when the baby is in the womb it does not need to breathe for itself, as the mother's circulation provides all the oxygen that is necessary for the foetus to survive. Furthermore because of a series of shunts, or communications within the heart of the foetus, the circulation is able to compensate for the underdeveloped left side. As a result, when the baby is born and needs its own circulation to provide oxygen and once some of the channels which are present in the heart in utero, begin to close, the baby can become extremely unwell.

Without surgical treatment almost all babies with hypoplastic left heart syndrome die in the newborn period. Two treatment approaches are available which will be outlined below. Some parents may choose not to put their child forward for surgical treatment and to allow their baby to die peacefully, either in hospital or at home. We at Royal Children's Hospital respect this opinion and would make every endeavour to provide any support that we could to these families.

Two surgical approaches are potentially available to families who wish to continue with treatment. It must be emphasised however that currently there is no cure available for hypoplastic left heart syndrome and both approaches are palliative. The first approach attempts to use the baby's own heart in order to create a viable circulation. A number of procedures are required in order to achieve this, beginning with the Norwood operation and eventually achieving a Fontan operation. The second approach is to perform heart transplantation. It must be emphasised however that it is extremely unusual for us to be offered a heart for transplantation that would be suitable for a newborn baby. In general therefore we do not tend to promote this approach.

## Norwood operation:

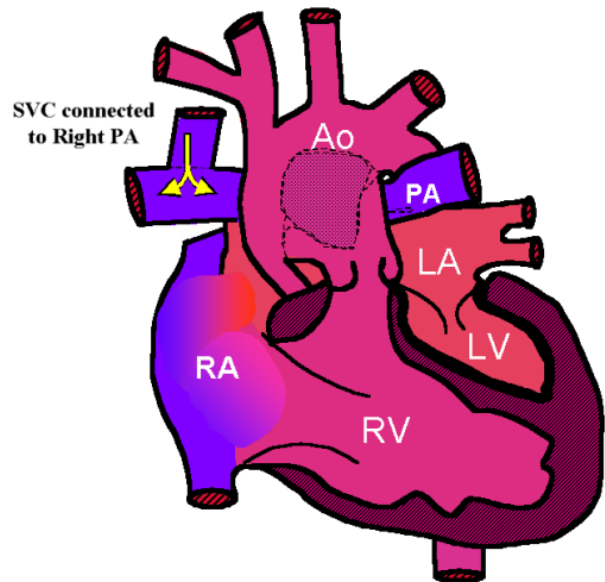
While most children with hypoplastic left heart syndrome are potentially suitable for a Norwood operation, we must emphasise that, because of slight variations in the anatomy or because of the clinical condition of the baby, some will be at much higher risk than others. Assuming that a Norwood operation proceeds, this involves connecting the origin of the pulmonary artery to the aorta, which is reconstructed with a patch, to allow the right ventricle to pump blood to the main circulation. The atrial septum is removed to allow blood to pass freely from the left atrium to the right side of the heart. A 'Shunt' operation is also done at the same time. This involves insertion of a tiny piece of artificial tube (made from Goretex) between the right arm artery and the right pulmonary artery, to maintain blood flow to the lungs.



After the Norwood operation the baby will remain mildly cyanosed (blue) and will require very close monitoring for several weeks or months to ensure that heart function is adequate and that the repair of the aorta and the shunt are working well. Any problem with feeding or intercurrent infection, such as a 'cold' or an episode of 'diarrhoea' may necessitate admission to hospital to make sure that the baby does not develop more serious complications. The family will need to have close medical or nursing support and the baby will need to be seen frequently by the hospital specialist during this time.

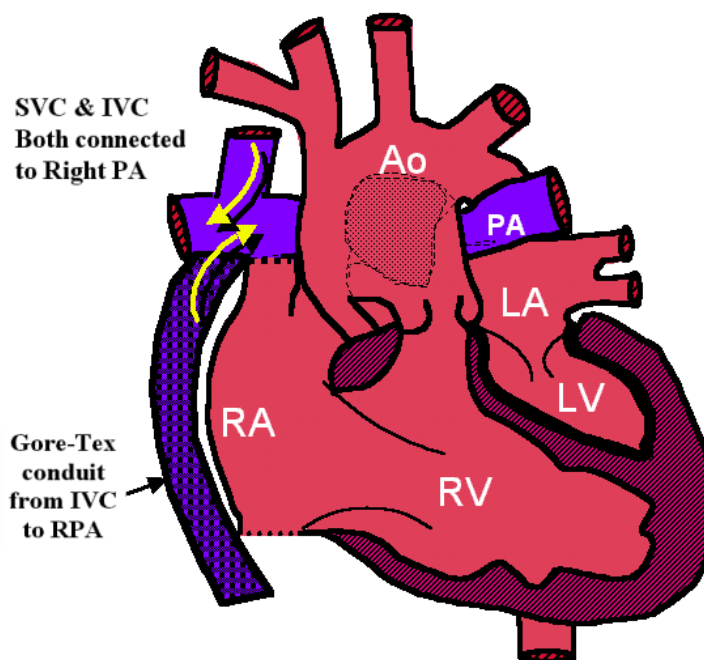
### **Bidirectional Cavopulmonary Shunt:**

At around three to four months, further surgery will be required. This is likely to involve a 'Bidirectional Cavo-Pulmonary Shunt' operation (sometimes called a 'BCPS' or 'Glenn'). In this operation the superior Vena Cava is connected to the right pulmonary artery and the previous shunt is removed. This allows blood from the upper part of the main circulation to get into the lung circulation directly. The baby will still be blue, but is usually better able to withstand infections and no longer needs such intense monitoring.



### **Fontan operation:**

The final operation is called the 'Fontan Operation', which is often done at between two and four years of age. The main result of this is that the blood returning from the lower circulation via the Inferior Vena Cava (IVC) is also connected to the pulmonary arteries. This leads to all the 'blue blood' (low in oxygen) being channelled through the lungs, without any 'pump' driving it. The right ventricle then pumps the red blood (high in oxygen), round the body. This operation makes the child 'Pink'.



Not all affected children need to have this final 'Fontan' operation. If they are well, after the BCPS, they may not require this last operation.

Some children may be unsuitable for a Fontan operation and alternative forms of treatment may be offered.

## **Coming to terms with HLHS:**

Finding out that your baby has a heart condition is very difficult, whether during the pregnancy or after the birth of your child. At first you may ask yourself “Did I do something wrong?” You may go through many different emotions having both ‘good’ and ‘bad’ days. It is important not to keep your emotions bottled up as to express your feelings can be a ‘good’ coping mechanism.

Although the people around you may not understand how you feel or what you are going through, they will be able to offer you support. You might find that talking to those around you is a good way of coming to terms with HLHS. It is important to be true to yourself and your emotions. If you are upset, sad, nervous or concerned, make sure you express your feelings.

If you have any questions while your child is being treated at the Royal Children’s Hospital it is important that you ask the hospital’s medical staff. Being worried about your child’s progress is only natural. Therefore, expressing these concerns to your Consultant, or other relevant staff, may ease some of your anxiety.

## **Coping with the Operations:**

When your child is about to undergo an operation it is hard to know how to pass the time as the duration of the procedure may be unpredictable. Although it may be difficult, it is important to occupy yourself during this time. If possible it may be valuable to leave the hospital for a while. Even if you just go for a walk or sit outside in the park you might find that the fresh air will help you relax. While this may be a time that you prefer to be alone, calling a friend may be a welcome distraction and even help the time to pass a bit quicker. Having a meal may also help settle your nerves.

Once your child has come out of surgery you will be able to see them in ICU. Seeing your child for the first time after their operation is difficult and if you do not want to go alone it is ok to ask for some support. You may like a friend or relative to accompany you into ICU, if not a hospital social worker can be contacted. If you do not want to be accompanied into ICU to see your child it might be a good idea to talk to someone about the experience afterwards. Once again, a hospital social worker can be contacted.

# Cardiac Medical Staff:

## Cardiac Surgeons

- Dr C. Brizard (Director)
- Dr A. Cochrane
- Dr R. Soto

## Cardiologists

- Dr D. Penny (Director)
- Dr J. Wilkinson
- Dr T.H. Goh
- Dr B. Edis
- Dr S. Menahem
- Dr L. Fong
- Dr R. Weintraub
- Dr A. Davis

## Nursing and Allied Health Staff:

- |                        |                |                       |
|------------------------|----------------|-----------------------|
| • Unit Manager         | Janette McEwan | Ext 5702              |
| • Care Manager         | .....          | Ext 5702              |
| • Lactation Consultant | .....          | Ext 5702              |
| • Dietitian            | .....          | Ext 5663              |
| • Physiotherapist      | .....          | Ext 5411              |
| • Speech Pathologist   | .....          | Ext 5549              |
| • Social Worker        | .....          | Ext 6111              |
| • Play/Music Therapist | .....          | Ext 5702/5424         |
| • Chaplain             | .....          | Via switchboard (x91) |
- Please fill in your allocated workers names in the spaces provided above.

## **Navigating the Hospital**

**The Royal Children's Cafe** - 1st Floor, Main Building

**Carparking** - entry from Gatehouse Street

Concession parking available, after the 3rd day of admission

Enquires can be made with the unit manager, care managers or ward clerk

**Card phone**- Located on Ground floor

**Chemist** - 1st Floor, Front Entry Building

**Chapel** - 1st Floor, Main Building. Ext# 5194

There is a range of denominations available

Regular services are held in the chapel

Baptisms and blessings can be arranged

**Deli** - 1st Floor, Front Entry Building

**Equipment Distribution Centre (EDC)** - Lower Ground Floor,  
South East Building. Ext# 5325

Hires breast pumps

Provides feeding supplies & equipment for discharge, plus much more

**Formula Room** - 1st Floor, Main Building. Ext# 5124

Stores fresh and frozen EBM

Makes formulas

**Gift Shop** - 1st Floor, Front Entry Building

**Information Desk** - Front entrance (Flemington Road)

Provides general information

Access to wheelchairs for new mothers

**Lunch Trolley** - Available at approximately midday on the ward

**McDonalds** - Front Entry (Flemington Road)

**Met Tickets** - Available from the Chemist, 1st Floor, Front Entry Building

**Parent Accommodation / Reception** - 28 Gatehouse Street. Ext# 5359

Provides a variety of accommodation for parents and families

**Public Phones** - 7th Floor outside lifts, and similarly on each floor.  
Phone cards are available on the 1st floor near the Chemist

**Public Toilets** - Ground, 3rd, 6th and 9th Floors

**Safety Shop, Travel Agent** - 1st Floor, Front Entry Building

**Sibling Care** - 1st Floor, North West Building. Ext# 6795  
Care for siblings, by volunteers, from 9am to 3pm Monday to Friday  
Free service although lunch needs to be provided for your child/children

**Social Work Department** - 5th Floor, South East Building. Ext# 6111  
Provide advocacy and support to parents/families

**Starlight Room** - 1st Floor, North West Building. Ext# 7991  
Free entertainment for all ages  
Open Monday - Friday 10am - 4pm

## **Looking After Yourself While Your Child is in Hospital:**

### **Advice from parents**

It is very easy to become totally focused on the care that your child needs, to the detriment of your own health. To be able to care for your child over what may be a long stay in hospital, it is very important that you remain in good mental and physical health.

Some of the points listed below seem obvious but their importance should not be underestimated.

- Eat well and drink plenty of water particularly if breastfeeding or expressing.
- Ensure that you get eight hours sleep in every twenty-four. A lack of sleep over an extended period of time can be very debilitating.
- Cook a good meal whenever possible.
- Accept any help that is offered and do not try to do everything on your own.
- Consider taking multivitamins (but not as an alternate to good quality food).
- Go for a walk every day to get some fresh air and exercise. It is also important to have a break from the institutional environment of the hospital for a short while.
- And mums, don't forget to organise and have your six-week postnatal checkup with an obstetrician.

## **Living Away from Home:**

### **Advice from parents who have been there before.**

Depending on the progress of your child you are likely to be faced with a long stay in Melbourne. In its self this is a big upset in your life and that of your family's.

In general it is recommended that you plan for a four-month absence from home and your normal environment. Routinely, your child will be required to remain in Melbourne, until the 2<sup>nd</sup> stage surgery, afterwhich they are likely to be more stable. Second stage surgery normally occurs when your child is 3-4months old.

### **Firstly accommodation.**

Long-term accommodation in Melbourne can be arranged through the hospital. This accommodation can take a number of forms depending on you and your family's requirements. You may of course wish to make your own arrangements but it is preferable to be reasonably close to the hospital in case of emergency.

Your child is going to require parental care from at least one parent for the entire length of his or her stay at the hospital. Also for the first few weeks at least it is important that this parent has the support of as many family members as possible during what is a very stressful time. Organising your family at this time is not easy but help is available. There are limited resources for sibling care through the hospital but it is strongly advised that you have another person stay with you to help with sibling care. This may be your partner, a grandparent or close friend.

As time progresses some of your family members may have to return home (work commitments perhaps). If this is the case some weekend visits should be organised if possible.

If your home is going to be unoccupied remember to ask a neighbour to keep an eye on it and forward your mail etc. You may need to make some banking, and/or social service arrangements prior to leaving home to ensure these services can be accessed from Melbourne.

Stay in contact with your normal circle of friends.

Most employers are very supportive of families faced with difficulties such as this. Do keep them informed of your child's progress and your leave requirements.

Avail of the services that the social workers at the hospital provide, they are very willing to help and although the heart condition your child has is relatively uncommon, the difficulties you are facing as a parent are not and help is available. Remember to keep your local doctors informed of your child's progress.

## **Making Medical Jargon Accessible: A Glossary of Commonly Used Terms:**

The following are terms that you may hear whilst your child is in the hospital. Always ask your cardiac team to explain things if they use terms you do not understand.

**ACYANOTIC** – Not looking blue

**ASD** - Hole between the top two chambers of the heart

**ANTIBIOTICS** – A medicine, which may be taken by mouth as a syrup, tablet or capsule, or may be given through a drip into a vein, to prevent or treat a bacterial infection.

**ANTICOAGULANT** – A drug used to reduce blood clotting, e.g. warfarin, aspirin.

**AORTA** – The main artery which takes blood from the heart in the circulation around the body (systematic circulation).

**AORTIC ATRESIA** – Complete obstruction to the aortic valve or the aorta.

**AORTIC STENOSIS** – Narrowing of the aortic valve resulting from a congenital defect of the valve leaflets or disease of the valve.

**AORTIC VALVE** – The valve at the origin of the aorta, which controls flow of blood out of the heart and prevents back flow.

**APYREXIAL** – Normal temperature

**ARRHYTHMIA** – A disturbance in the normal heart rhythm.

**ARTERIAL VALVE** – One of the valves which control flow from the ventricles into the circulation, during each heart beat, and prevents back flow.

**ATRESIA** – Complete obstruction to a valve or blood vessel, so that blood cannot get past this point in the circulation and has to take an alternative route.

**ATRETIC** – Obstructed.

**ATRIUM** – One of the two blood – collecting (reservoir) chambers at the top of the heart; (plural = atria or atriums).

**BLOOD TESTS** – Many blood tests may be needed before and after heart surgery, or at other times. They may include blood counts to detect anaemia or infection, or chemical analysis to detect an imbalance in the blood or build-up of harmful acid or waste products. During the early days after surgery and while patients are having drug treatment for congestive heart failure, chemical imbalances in the blood are frequent and may require medications to be altered. Some medicines need to be checked to see how much of it is in the child's blood in order to assess whether the dose needs to be increased or decreased. Other tests may measure the effectiveness of a medication (e.g. anticoagulants). The presence of fever or other indication of possible infection will also need investigation with blood counts or blood cultures.

**BLOOD PRESSURE** – A measure of the pressure with which the heart pumps blood into circulation, usually referred to by two numbers. The 'systolic' pressure refers to the maximum pressure in the artery during each heart beat, while the 'diastolic' pressure is the minimum pressure between heart beats. The left side of the heart and arteries are operating as a high pressure system, while the veins, the right side of the heart and the pulmonary circulation operate at a much lower pressure.

**BRADYCARDIA** – A slow heart rate.

**BYPASS OPERATION** – An operation needing 'Heart-Lung Bypass'. This refers to the use of the heart-lung bypass machine which take over the function of the heart and the lungs, pumping blood round the body and supplying oxygen to the blood. (When people refer to a 'double bypass' or 'triple bypass' operation etc., they are usually referring to an operation to provide a bypass for blocked coronary arteries, usually in adults with Coronary Heart Disease. This is not the same as a Bypass Operation, although most operations for Coronary Heart Disease do use 'Heart-Lung Bypass').

**CANNULA** – A very small tube introduced into a vein to provide access for a 'drip'.

**CAPILLARY** – A microscopic blood vessel connecting arteries to veins. The blood constituents seep through these tiny vessels in the systemic (body) circulation and pulmonary (lung) circulation. Their walls are extremely thin and allow oxygen, carbon dioxide, glucose and other chemicals to pass to and fro between the blood and the surrounding cells in the body organs, or air sacs in the lungs.

**CARDIAC** – A term used for anything to do with the heart (e.g. cardiac muscle, cardiac specialist, cardiac rhythm etc.).

**CATHETER (CARDIAC)** – A ‘tube’ used to perform heart tests, by introducing it through an artery or vein into the heart (Catheter Tests).

**CATHETER (URINARY)** – A ‘tube’ used to drain urine from the bladder (often used after heart surgery).

**COARCTATION** – A area of stenosis (narrowing) in the aorta.

**CONDUIT** – A term referring to a tube used to connect one part of the circulation to another surgically (e.g. to bypass an obstructed valve or blocked artery).

**CONGENITAL** – Describes a condition which develops during formation, before birth, and so already exists when a baby is born.

**CONGENITAL HEART DISEASE** – A defect or disease affecting the heart from birth (compare with ‘Acquired Heart Disease’).

**CONGESTIVE HEART FAILURE** – A condition which occurs when there is a build-up of fluid (i.e. congestion) in the lungs or other organs such as the liver. This congestion usually results when the heart is unable to work efficiently. Children with heart failure may have symptoms such as marked shortness of breath and difficulty with feeding. The term ‘heart failure’ **does not** mean that the heart will suddenly stop beating.

**CONTRACTION** – When the muscle in the heart wall works (squeezes) to push the blood through the heart and out into the arteries.

**CORONARY ARTERIES** – These small arteries carry the blood supply to the heart muscle itself, and are the first arteries to branch off the aorta.

**CROSS MATCH** – The test to ensure that blood, for transfusion, is compatible with the patient and will not cause an unwanted reaction.

**CYANOSIS** – The condition of the skin and nails looking a purplish-blue colour due to lowered oxygen level in the blood. A child with this condition may be described as being cyanosed.

**DEFECT** – A physical abnormality, e.g. of the position or structure of the heart or main blood vessels.

**DIASTOLE** – The time during which the ventricular heart muscle relaxes after each contraction, between each heart beat.

**DIASTOLIC PRESSURE** – The lowest point of blood pressure in the arteries.

**DIGOXIN** – A medicine given to strengthen the heart contraction or reduce the heart rate.

**DIURETICS** – Medicine to help the body to get rid of the excess fluid which may build up in the lungs or elsewhere in the body in congestive heart failure, e.g. aldactone, lasix. It does this by causing the kidneys to increase their production of urine. Other medicine, e.g. potassium, may be given at the same time to maintain the balance of salts in the body, which may tend to be affected adversely by the diuretic.

**DUCTUS** – The blood vessel connecting the aorta with the pulmonary artery before birth. The full name is ‘Ductus Arteriosus’. It usually closes within the early days or weeks after birth. If it remains open after this time it is called a Patent (or Persistent) Ductus Arteriosus (PDA).

**ECHOCARDIOGRAM (ECHO)** – An ultrasound scan of the heart. Very high frequency sound waves (ultrasound) are used to create a moving picture of the heart and of blood flowing through it, using a sophisticated computer. The test detects most heart defects and can provide detailed information about the nature and severity of heart problems of many kinds. The test is not painful, but requires that the child remain still for 20 minutes (usually longer). Some children may benefit from mild sedation to help them cooperate during the scan.

**E.C.G.** - Electrocardiogram

**EFFUSION** – A collection of fluid in an area such as the pericardium or the pleural cavity. Such a collection, if it is large, may lead to build-up of pressure on the heart or lungs, and often results in deterioration in their function, and so it may need to be drained.

**ELECTROCARDIOGRAM (ECG)** – A test to measure the heart’s electrical activity with each heart beat. Wires are attached to the skin of the arms, legs and chest, using soft, stick-on discs (called ‘electrodes’). A tracing is printed on paper and gives information about the heart rate and regularity, as well as providing data about enlargement of the heart chamber and thickening of heart muscle, which may provide useful information about the nature and severity of heart problems.

**FONTAN OPERATION** – An operation to connect the main veins from the systemic circulation (SVC and IVC) to the lung arteries. Blood then flows directly into the lung circulation, after returning from the body, without going through the right ventricle as

would happen in a normal heart. This operation is named after a French surgeon – Francis Fontan.

**FORAMEN OVALE** – The hole between the two atriums present at birth.

**GENERAL PRACTITIONER (GP)** – Your family doctor. It is helpful to first visit your GP when your heart child is reasonably well. Then, when your child is ill, the doctor can see how skin colour, breathing, heart sounds, etc. have changed.

**HAEMOGLOBIN** – The red blood pigment which carries oxygen in the red blood cells.

**HEART-LUNG BYPASS** – A technique employed for nearly all open heart operations by which the circulation is supported by a machine. The machine takes over the function of both the heart and lungs (the ‘heart-lung bypass machine’), while the surgeon opens the heart to perform an operation inside it.

**HYPO** – Prefix meaning smaller or less than usual.

**HYPOPLASTIC** – Smaller than normal or underdeveloped.

**HYPOPLASTIC LEFT HEART SYNDROME** – A condition in which the left side of the heart is poorly developed and unable to pump blood into the systemic circulation effectively.

**HYPOTENSION** – Low blood pressure (in the systemic circulation) – a problem quite often encountered after heart surgery and may need treatment with medication to raise the blood pressure.

**HYPER** – Prefix meaning larger or more than usual.

**HYPERTENSION** – High blood pressure. The term usually refers to high pressure in the systemic circulation. However ‘pulmonary hypertension’ means elevation of pressure in the lung circulation.

**INCOMPETENT** – A term used to refer to ‘leakage’ at a heart valve. The valve is incompetent if it fails to close effectively and does not prevent ‘back-flow’. The flow of blood backwards through a valve, which should be closed, is referred to as ‘regurgitation’ or ‘incompetence’.

**INFECTION** – Occurs when a micro-organism which can produce disease invades a living tissue. **Inflammation** is one of the body’s reactions to infection.

**INFERIOR VENA CAVA (IVC)** – The main vein from the lower part of the body which returns deoxygenated blood to the heart.

**INFLAMMATION** – Occurs when a living tissue is reacting to an injury or an infection. Swelling, pain or redness in the tissue are signs of inflammation.

**INTENSIVE CARE UNIT (ICU)** – The special medical and nursing unit to which patients go for a few hours (or days) after major heart surgery or if they are extremely ill. This area is used to ensure very close monitoring, and ready availability of medical and nursing staff and equipment to cater for any emergency. It is available for seriously ill children, with a wide variety of medical problems, and for those who have had open heart operation, or other major heart surgery, especially if they need ‘ventilator’ treatment.

**INTRAVENOUS DRIP (IV or DRIP)** – A method of providing medications, fluids or nutrition into a vein. The fluid usually flows from a polythene bag or bottle and can be seen to ‘drip’ into a small container (chamber), which is connected by a tube to a cannula in the vein. Most children will have a ‘drip’ following an anaesthetic or after heart surgery. In small babies and in children who need continuous medication by ‘drip’, the medications may be given by a motorised syringe pump, which controls the rate of administration very precisely.

**IVC** – Inferior Vena Cava

**MURMUR** – A noise, heard with the doctor’s stethoscope (or occasionally with the naked ear), which results from turbulence (eddies) in the flow of blood through the heart or blood vessels. The noise often has a ‘blowing’, ‘swishing’ or ‘cooing’ character and is quite different the ‘heart sounds’ – though many normal children have soft murmurs.

**OEDEMA** – Fluid retention

**OPEN HEART OPERATION** – An operation which requires that the heart be opened in order to perform surgery inside it. These operations necessitate the use of heart-lung bypass.

**PACEMAKER (ARTIFICIAL)** – An electronic device used to stimulate the heart and regulate the heart rhythm. Many children are attached to a pacemaker temporarily after heart surgery, in case their heart rhythm becomes abnormal. Rarely, children need a permanent artificial pacemaker (often because of heart block), in which case the pacemaker is put in, under the skin and upper abdomen or in front of an armpit, at an operation.

**PAEDIATRICIAN** – A specialist in children's disease or problems. Some paediatricians are experts in particular types of disease or defects (e.g. Paediatric Cardiologist, or Paediatric Neurologist [specialist in brain and nerve disease]). Others take a more general interest in the whole development of the child, physically and intellectually, and in a wide range of disease and problems of infancy and childhood.

**PATENT** – 'Open' (as in Patent Ductus Arteriosus).

**PATENT DUCTUS ARTERIOSUS (PDA)** – Refers to the situation when the ductus remains open after the early days or weeks of life, resulting in a shunt of blood between the aorta and the pulmonary artery.

**PERICARDIAL EFFUSION** – A collection of fluid in the pericardium (sac around the heart). Such a collection, if it is large, may lead to a build-up of pressure on the heart, which may result in a deterioration in its function.

**PLEURAL EFFUSION** – A collection of fluid in the pleural cavity. Such a collection, if it is large, may lead to build-up of pressure on the affected lung. This often results in deterioration in its function, with breathlessness and a fall in oxygen levels in the blood.

**PNEUMOTHORAX** – Presence of a collection of air in the pleural cavity. It may result from a tiny puncture in the outside wall of the lung or can follow heart or lung surgery. Such a collection, if it is large, may lead to a build-up of pressure on the affected lung. This may result in a deterioration in its function, with breathlessness and a fall in oxygen levels in the blood.

**PULMONARY** – A term for anything to do with the lungs.

**PULMONARY ARTERIES** – The arteries carrying blood into each lung. There are two large branches from the (main) pulmonary artery, ('left pulmonary artery' and 'right pulmonary artery') and many smaller branches within each lung. All these are called 'pulmonary arteries'.

**PULSE** – The 'beat' felt in an artery, for example in the wrist, groin, neck, or other sites and measured in beats per minute. It shows the rate at which the heart is beating, or contracting.

**PYREXIAL** – High temperature

**RED BLOOD CELLS** – The blood cells which carry oxygen.

**REGURGITATION** – See 'Incompetent'.

**RESPIRATION** – Breathing.

**RESPIRATOR (VENTILATOR)** – A machine for helping to maintain adequate respiration when the patient is unable to breathe adequately on their own.

**RESPIRATORY** – Anything to do with ‘respiration’.

**RESPIRATORY FAILURE** – Implies that the lungs are unable to perform adequate gas exchange.

**SEPTUM** – The wall within the heart separating the left and right sides. The atriums are separated by the ‘atrial septum’ and the ventricles by the ‘ventricular septum’.

**SHUNT** – A term which may refer to either 1) blood flow from an area of higher pressure to an area of lower pressure, through an abnormal communication (e.g. septal defect), or 2) a surgically created communication between two blood vessels usually achieved via the insertion of an artificial tube (conduit). This improves the circulation through the lungs, in cyanosed infants or children, who have reduced blood flow in the lungs.

**STENOSIS** – Narrowing (usually of a heart valve or an artery).

**SUB-AORTIC STENOSIS** – Narrowing below the aortic valve. This usually results from the presence of abnormal tissue or muscle below the valve.

**SUPERIOR VENA CAVA (SVC)** – The main vein from the upper part of the body which returns deoxygenated blood to the heart.

**SVC** – See ‘**Superior Vena Cava**’

**SYSTEMIC** – Something which involves the whole body, e.g. systemic circulation.

**SYSTOLE** – The time during which the ventricular heart muscle contracts to pump blood.

**SYSTOLIC BLOOD PRESSURE** – The highest point of blood pressure in the arteries.

**TACHYCARDIA** – A fast heart rate.

**TRICUSPID VALVE** – The valve at the junction of the right atrium with the right ventricle, which controls flow of blood into the ventricle before each contraction, and

prevents back-flow.

**VALVE** – A structure in a blood vessel or the heart which ensures blood flows only one way. They are constructed of single or multiple flaps which swing open to allow blood to flow forwards and swing shut to prevent back flow. The valve flaps are referred to as ‘leaflets’ or ‘cusps’.

**VEINS** – Blood vessels which carry blood back towards the heart, after it has circulated around the body. Veins usually carry deoxygenated blood, except in the pulmonary veins where oxygenated blood is carried back to the heart from the lungs.

**VENTILATE** – To use a ventilator (respirator) to help a patient who cannot breathe adequately for themselves.

**VENTILATOR** – See ‘**Respirator**’.

**VENTRICLE** – One of the two powerful muscular chambers at the bottom of the heart which pump blood out to the body with each heart contraction.

**VENTRICULAR SEPTAL DEFECT (VSD)** – A defect (hole) in the ventricular septum which allows blood to shunt from one ventricle to the other.

**VENTRICULAR SEPTAL** – The septum (partition or wall) between the two ventricles.

**VSD** – See ‘**Ventricular Septal Defect**’.

**X-RAY** – An X-ray of the chest will often be performed as part of an initial assessment or a follow up appointment. This will show the size and shape of the heart and also helps to demonstrate effects of heart problems on the lungs (e.g. congestion of the lungs). Enlargement of the heart and abnormalities of its shape may give valuable information about many heart defects and their severity.

### **Please note**

All terms used in the above glossary were taken from:

Children with Heart Problems: A booklet for parents of children with a heart condition. Produced for the Cardiology Department Royal Children’s hospital Melbourne Australia. 1997 Heartkids VIC Inc.

## **Contacts and Supports**

### **HEARTKIDS**

Heartkids Victoria is a support organisation for families and children affected by congenital or childhood-acquired heart defects.

There are similar support groups for families of children with heart conditions in each Australian State and Territory.

There are a number of families around Australia who have had children diagnosed with HLHS and who are willing to share their experiences.

These families may be contacted through Heartkids Victoria, by phoning Lynette McCoullough on 03 9333 1586.

### **HLHS WEB PAGE LINKS**

<http://www.lhm.org.uk>

<http://www.hlhsinfo.homestead.com/>

[www.choa.org/library/conditions/hlhs\\_whatishlhs.shtml](http://www.choa.org/library/conditions/hlhs_whatishlhs.shtml)

<http://www.geocities.com/aussiehearts/>

## Acknowledgments

The Royal Children's Hospital Melbourne would like to acknowledge the following for their help in the compellation of this resource book.

- Royal Children's Hospital Cardiologists Dr D. Penny (Director of Cardiology) & Dr J. Wilkinson.
- Sweekum Ho & Danielle Peuker - Care Managers of 7West at the Royal Children's Hospital Melbourne.
- Margaret Patullo, President of Heartkids Vic.
- Isabel Alessi, Social Worker, Royal Children's Hospital Melbourne.
- Daniel Taylor, Social Work student, Royal Children's Hospital Melbourne.
- Many thanks to other Royal Children's Hospital Melbourne staff for supporting the parents group and the compellation of this book.

In early 2001 a group for parents of children with HLHS was run at the Royal Children's Hospital Melbourne. Parents in this group wanted to provide a resource book for future parents whose children are diagnosed with HLHS. Without their contribution, this book would not have been possible.

- Slavica & Edward Butler, parents of Kieran
- Deanne Dumigan & Andrew Jewell, parents of Sam
- Trudy Stewart, mother of Riley
- Simone & Stewart Downs, parents of Chloe
- Simone Boundey & Peter Whiting, parents of Bradley
- Catherin & Mark Worsnop, parents of Thomas