

Congenital Heart Disease Factsheets: **SINGLE VENTRICLE DEFECTS**

What is congenital heart disease?

Congenital heart disease is the term used to describe a number of different problems that affect how well the heart works. The word “congenital” means the problem occurs while the fetus is being formed, meaning the condition is present at birth.

There are a large number of different defects, some are mild and don’t need treatment, while some are life-threatening and need open heart surgery or other medical interventions. Sometimes the term congenital heart “defects” is used instead of “disease” – either way, the name is commonly shortened to CHD.

CHD is the most common congenital birth defect – affecting around 1 in every 125 babies – and around 3,000 surgeries or catheter procedures take place on babies under one year of age in the UK every year.

What are single ventricle heart conditions?

In a normal heart there are four chambers. The two lower chambers are ventricles and they pump blood out of the heart.

A child with a single ventricle heart condition has only one ventricle that is large enough to work effectively. The other ventricle may be small, underdeveloped or missing a valve. Single ventricle defects are rare. They occur in about five out of every 100,000 live births.

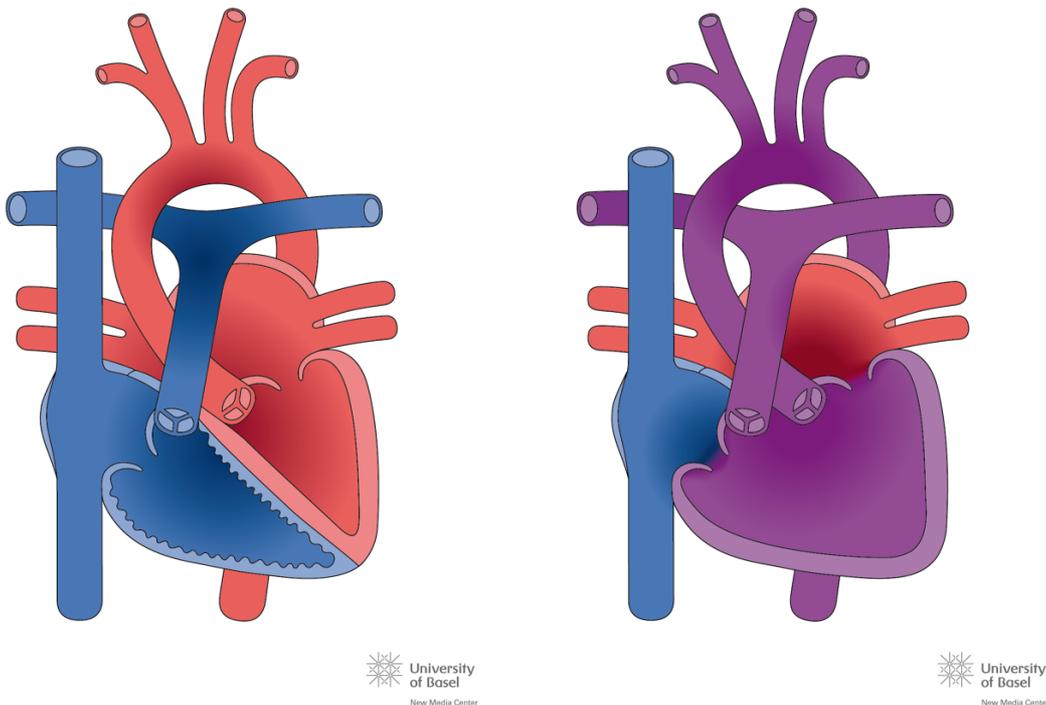
Different types of single ventricle heart conditions

There are different types of single heart conditions, some of which are:

- **Hypoplastic Left Heart Syndrome (HLHS):**
This defect comprises a number of problems on the left side of the heart. The left ventricle is small (hypoplastic) and the mitral and/or the aortic valve may be narrow, blocked or not formed at all. The aorta is often small.
- **Tricuspid atresia:**
A defect where the valve between the right atrium and right ventricle doesn’t form at all.

- **Pulmonary atresia with intact ventricular septum:**
A defect where the valve that controls the flow of blood between the right ventricle and the main pulmonary artery doesn't form properly and there is no hole between the right and left ventricles
- **Univentricular heart (Double inlet or outlet ventricle):**
In these conditions there is a single large pumping chamber (ventricle) which both collecting chambers (atria) empty their blood into. There may be a valve at the end of each collecting chamber or just one between the two. The two major blood vessels leaving the heart may leave from either the left or right sides of the ventricle and they may be swapped around (transposed).

How a single ventricle heart is different



The diagram on the left above shows a normally developed heart, with two similar-sized ventricles at the bottom. The diagram on the right above shows a heart with a single ventricle condition – you can see that the one of the ventricles has not developed fully.

Please note, this is a basic diagram to help show the differences that can occur with a single ventricle condition. Not all single ventricle issues are the same and every child's heart is different. Your cardiologist will be able to tell you more about how your child's heart is developing.

How can they be spotted?

Most single ventricle hearts can be detected at the 20 week anatomy scan or after birth by echocardiogram.

If it is not picked up in the antenatal period, once the baby is born they will display the following signs:

- Bluish skin colour
- Difficulty breathing
- Difficulty feeding
- Lethargy (sleepy or unresponsive)

In most cases, children with single ventricle heart defects require intensive medical intervention soon after birth.

How can they be treated?

Single-ventricle heart defects cannot be corrected. However, the symptoms can be improved by surgery. Normally several stages of surgery are required to balance and optimise blood flow to the lungs and the body.

These will be open heart surgeries performed under general anaesthetic, the timing of which will depend on the severity of the anomaly, and the general condition of the baby. Once baby has been born, the cardiologist will assess whether:

- There is too much blood going to the lungs
- Not enough blood going to the lungs

If there is too much blood flow to the lungs pulmonary banding will be needed to restrict this excessive flow. If there is not enough blood flow to the lungs then a shunt or a stent will be needed to increase the blood going to the lungs.

Some babies with univentricular hearts balance themselves out regarding blood flow and don't need any intervention in the immediate neonatal period.

Other surgeries offered can include:

- **Norwood procedure:**
This is the first surgery that is usually done in the first few weeks after birth. The surgeons will build a new larger aorta and use a small tube to direct blood flow to the lungs.
- **Glenn procedure:**
The second surgery is usually done when the baby is around 6 months of age and the surgeons direct the blood flow from the upper body to the pulmonary artery

so that the blood can collect oxygen from the lungs without having to go through the heart

- **Fontan procedure:**

This is the third surgery that is usually done when the child is 2 – 6 years old. The surgeons will now be separating the circulation so that no mixing of blood occurs. This also decreases the workload on the single ventricle. This procedure does not create a normal blood circulation in the body but it does improve the circulation.

General advice & information for the future

Babies and children with a single ventricle will need lifelong follow up. Most will be on medicines for the rest of their lives.

They will need repeat tests like echocardiogram, ECG and sometimes cardiac Magnetic Resonance Imaging (MRI) scans. The aim of these check-ups is to monitor your child's heart function so that any future heart problems are diagnosed and treated quickly.

Your cardiologist will advise you about any restrictions on activity that you should be aware of. As complicated as these conditions are, children are encouraged to be as active as they possibly can be.

As your baby will have had open heart surgery, they will have a scar down the middle of the chest, and there will be small scars where drain tubes were used, but these will fade over time.

How Tiny Tickers can help you

Tiny Tickers is here to help you and your family when you receive a diagnosis of congenital heart disease. Our website – www.tinytickers.org – has lots of information and experience of other families. We have a private Facebook forum with hundreds of other parents who you talk to. We have booklets about congenital heart disease and about going into hospital for surgery that are free to order.

Hearing the news that your baby has a poorly heart is very upsetting. Please look after yourself, and only read information from trusted sources. Congenital heart disease may be rare, but you are not alone. Please contact us via our website or info@tinytickers.org if you need further information or support.