

Tricuspid Atresia

What is Tricuspid Atresia?

Tricuspid refers to the Tricuspid Valve. Atresia means closed or absent.

So Tricuspid Atresia means:

- The tricuspid valve is closed or absent.
- The right ventricle is small.
- The pulmonary artery is narrow.
- The pulmonary valve may be blocked.

Because the tricuspid valve should act as a gateway between the right atrium and the right ventricle, when it is missing blue (deoxygenated) blood cannot be pumped through the right side of the heart to the lungs.

To survive your baby will need either:

- at birth, a hole between the atria – an ASD – and a hole between the ventricles – a VSD. The ASD allows blood to cross into the left atrium, through the mitral valve and into the left ventricle. Most of it will be pumped into the aorta, but some will cross the VSD and move into the pulmonary artery (if it is not blocked) and so to the lungs.

or

- a Ductus Arteriosus. The Ductus Arteriosus is a part of our circulation before we are born, and usually closes shortly after birth. It links the aorta to the pulmonary artery, allowing some blood to get to the lungs.

Fig 1 – Tricuspid Atresia

Tricuspid Atresia

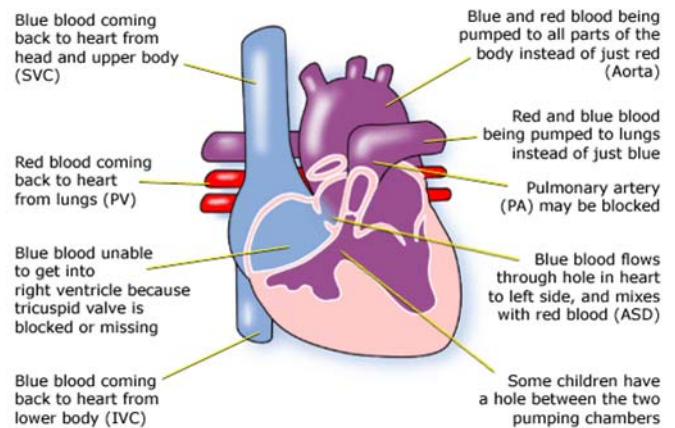
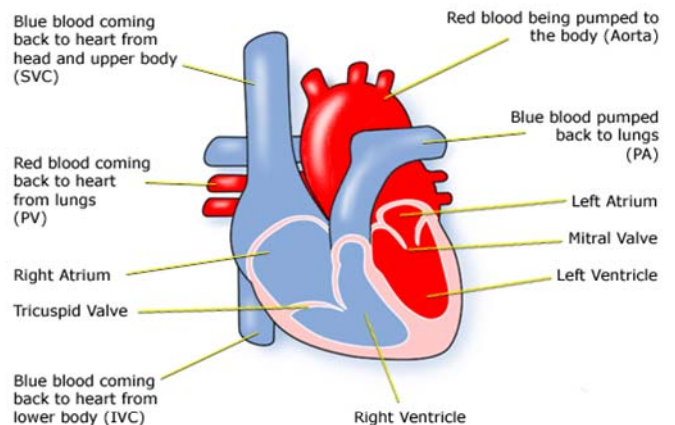


Fig 2 – Normal Heart

Normal Heart



Diagnosis

Tricuspid Atresia sometimes be picked up before birth on a scan when only three chambers can be seen.

If not, after birth a baby will be diagnosed early on – depending on how much blood

is getting to the lungs – because he or she will look blue, have problems feeding because of breathlessness, and the sound of blood moving in the wrong direction through the heart can be heard as a heart murmur.

When your baby is born how badly affected your baby is will depend on how much blood is getting to the lungs.

If the pulmonary artery is narrowed (stenotic) these symptoms will be apparent very early.

The tests used can be:

- pulse, blood pressure, temperature, and number of breaths a baby takes a minute
- listening with a stethoscope for changes in the heart sounds
- an oxygen saturation monitor to see how much oxygen is getting into the blood
- a chest x-ray to see the size and position of the heart
- an ECG (electrocardiogram) to check the electrical activity
- an ultrasound scan (echocardiogram) to see how the blood moves through the heart
- checks for chemical balance in blood and urine
- a catheter or Magnetic Resonance Imaging test may be needed.

Treatment

As soon as the diagnosis is made, the baby is moved to a paediatric cardiac unit, and medicine is given to keep the Ductus Arteriosus open.

The baby will need surgery when a few weeks or months old – depending on how he or she is coping. This could mean taking an artery which normally carries blood to the arm and diverting it to the pulmonary arteries so that a larger blood supply will pick up oxygen from the lungs (a BT shunt). This operation is done

through the side of the chest while the heart is still beating.

Further surgery will need to be carried out to improve the amount of blood picking up oxygen from the lungs. This may be:

- a Glenn / Cavopulmonary / HemiFontan Shunt - this involves making a passage between the superior vena cava – the big vein bringing blue blood from the upper body to the heart – to the right pulmonary artery. This means that a much larger amount of blue blood will flow into the lungs and pick up oxygen.
- a Fontan / Total Cavo-Pulmonary Connection. This operation joins the inferior vena cava to the pulmonary arteries as well.

These are open heart surgeries – the heart will need to be stopped and opened to repair it. This means that a machine will have to take over the job that the heart and lungs normally do– the heart bypass machine.

The aim of the operations is to increase the amount of blood picking up oxygen from the lungs. The success will depend on how well the blood can flow into the pulmonary arteries from the low pressure veins.

Which of these operations is used will depend on the size of the pulmonary arteries and the pressures within them. Your child may need catheter tests to measure these pressures inside the heart.

Some children will need to continue on diuretic medicines to help them get rid of excess fluid. In some cases an anticoagulant medicine, such as aspirin or warfarin, will be prescribed. This is because blood flowing at low pressure (there is no pumping from the right ventricle) could cause a blood clot (a thrombosis).

The length of time in hospital can be several weeks and a longer period may be needed in intensive care or high dependency unit while the heart and lungs adjust to the different pressures.

How the child is affected

After the major surgeries your child may take a long period to recover. He or she will have a scar down the middle of the chest, and there will be small scars where drain tubes were used. These fade very rapidly in most children, but they will not go altogether. Smaller scars on the hands and neck usually fade away to nothing.

If the child is on warfarin the level of anticoagulation has to be monitored frequently using a blood test (INR test – this stands for International Normalisation Ratio).

Some children develop a fast heart beat (a tachycardia) after surgery. This may be tackled with antiarrhythmic drugs if it is serious.

Your child may develop more slowly than others and tire quickly – due to the lack of oxygen in the bloodstream. After the last surgery to redirect blood, he or she should be able to catch up with others of his or her age, and take part in normal activities.

Most parents are amazed at how quickly their child recovers from surgery and starts to gain weight.

Some of these problems can occur after surgery or later in life:

- It is common for the valves to leak a little, but if this becomes severe, they may need further repair or even replacement with an artificial valve. If this happens the child will need to take an anticoagulant medicine to stop blood clots forming. The anticoagulant effect has to be monitored frequently using a blood test.

- A very fast heart beat (tachycardia) can happen every now and then. This may need to be corrected by medicines – antiarrhythmic drugs. If the problem continues your child may be offered an ablation.
- Hearts that are not normal are more likely to have an infection called endocarditis. Although rare this is a difficult disease to treat. People born with Tricuspid Atresia will need to take antibiotics if there is a chance that a large number of bacteria will get into the blood stream. The most common way for this to happen is during ear-piercing or tattooing, or surgery or a dental procedure such as de-scaling of teeth or an extraction.

These problems may not become serious until the teen years or adulthood.

Children on warfarin may not be able to take part in contact sports, and their INR will need monitoring. Girls should be warned against pregnancy when taking warfarin, and should be aware that carrying a baby to term may be difficult and pregnancy may endanger their health.

Some children have other health problems and learning difficulties. It is natural to think that this is due to the surgery that the child had when he or she was very young, but they may be part of the same syndrome that caused the heart defect. Make sure that your child is referred to a Child Development Centre so that other problems can be diagnosed as early as possible and before school-age.

Your child's heart condition will be carefully monitored throughout his or her life.

Parents' stories

Melissa

They should have known that Melissa was ill – I told the midwife that she wasn't right

straightaway. I was told all babies are a dusky colour, but her feet were navy blue. It was when the night staff came on that one of them looked at her, and carried her off to the Special Care Baby Unit straightaway. I was rigid with fear and tried to find her – they said they were doing tests and then came and said they thought she had a heart problem. That really, really frightened me, because I had a little sister who died as a baby from a heart problem. I wasn't told much about it, but I thought babies couldn't survive if their hearts aren't alright.

Then she had to go to another hospital miles away – yet more tests they said. They took her in an ambulance, but I had to go separately, and my husband was working so I got my brother to come and get me.

They took her straight in to the hospital and said she would need an operation the next day, but then they decided it would have to be that night. Dave had come in by then, and he signed a form, although we were told that she might die – well, we knew that.

It's all a blur now, but we got to take Melissa home, and she had two more lots of surgery before she was two. The hardest part really, apart from the operations, is that she won't eat – or only a very little bit. She had a tube when she was very young, and we got extra strength

formula, and supplements from the hospital for her, but she doesn't like eating.

Freddie

Angela was sent for an extra scan at 22 weeks pregnant. They brought in a paediatric cardiologist who spent a long time watching images on a machine – then he said that half of the foetus' heart wasn't working properly. We were devastated, and didn't hear what was being said – although I'm sure we asked the right questions. We were given a lot of material and invited back to talk about the course the pregnancy should not take.

We had to consider whether we could take on a child with Tricuspid Atresia – I work abroad a lot, and our daughter travels with us. Also we are a very active family – our daughter Chelsea already rides, and has tried skiing.

I contacted family who had a child with a similar heart problem – he was nine and you could hear the pride in his mother's voice when she said that no-one would be able to tell just by looking at him what he had been through.

We went ahead, and Freddie is a great little boy. I'm looking for a move to London with my company, and I think we are strong enough to give Freddie the best chance in life.

Please contact CHF if you have suggested amendments or changes as we like to keep our information sheets relevant and up-to-date.