

Truncus Arteriosus (or Common Arterial Trunk)

What is Truncus Arteriosus?

When a baby's heart is developing in the womb, one large blood vessel, the truncus arteriosus, should divide to become the two main arteries: the aorta, and the pulmonary artery.

The aorta should carry red (oxygenated) blood from the heart to the body, and the pulmonary artery should carry blue (deoxygenated) blood to the lungs.

If the aorta and pulmonary artery do not divide, the child is born with a persistent truncus arteriosus or common arterial trunk. This one big artery has one large valve instead of there being pulmonary and aortic valves

A part of the wall between the left and right ventricles is missing (VSD) so that this single blood vessel is able to receive blue blood from the right side of the heart, and red blood from the left side of the heart.

The Truncus Arteriosus divides into the aorta and the pulmonary arteries, which carry this mixture of blood to the lungs and to the body:

In Type 1 the pulmonary artery branches off just above the valve.

Type 2 the pulmonary arteries to the lungs branch separately but close together directly off the truncus.

Type 3 the pulmonary arteries branch separately and are further apart.

There may be other differences that affect how your child is treated – the VSD may be missing, the valve may not work well, and may leak or be narrowed.

The result of Truncus Arteriosus is usually that too much blood is going to the lungs and too little red (oxygenated) blood is reaching the body.

Fig 1 – Truncus Arteriosus

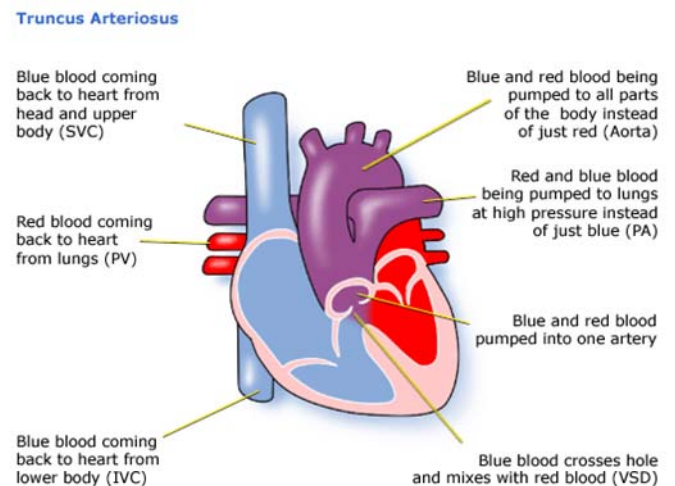
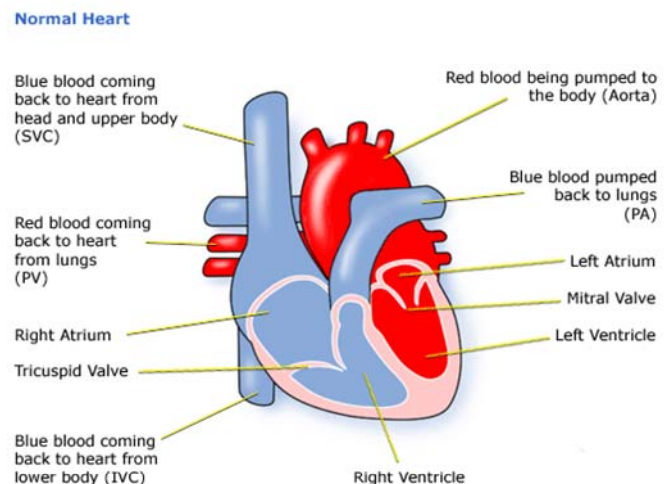


Fig 2 – Normal Heart



Diagnosis

Truncus Arteriosus can sometimes be seen on a scan during pregnancy.

After birth your baby may have been diagnosed because a heart murmur was

heard. The child becomes increasingly blue (cyanosed), because of the blue blood going being pumped to the body instead of the lungs.

Because of the high pressure he or she will develop congestive heart failure as blood flow to the lungs increases, be breathless, difficult to feed and 'fail to thrive'.

When a heart defect is suspected 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

The aim of the treatment is to separate the circulation of the blood so that blue blood goes to the lungs, and red to the body.

Risk of Procedures

There is a risk to your child in the procedures described, but how great that risk is depends on how severe the heart defect is, and how well your child is otherwise. The doctors will discuss risks with you in detail before asking you to consent to any of the operations.

Although the time is often very short to take decisions on behalf of a newborn, you

will find that later decisions often leave you and your child several months or even years to decide the best time for more treatment.

Surgery

Surgery will be needed before the increased pressure irreparably damages the lungs (pulmonary vascular disease). This will be open heart surgery. 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 usual procedure is to place a donor lung artery from the right ventricle to the branch lung arteries and a plastic patch is usually used to close the hole (VSD). There is no risk of rejecting the donor lung artery, since it is not living blood vessel.

The truncus will now act as the aorta, carrying red blood from the left ventricle to the body. Surgeries will need to be repeated as your child grows.

The more complicated types of Truncus may be treated with a Fontan-style of operation

How the child is affected

The surgery changes the pressures inside the heart, and it may take your baby a while to adapt to the new ones. It is not uncommon for a child to pick up an infection, such as a chest infection or infected wound, while undergoing treatment. Some children react badly to some kinds of medicines. The kind of surgery needed can sometimes cause a very fast (tachycardia), or very slow (bradycardia) pulse rate, which may need medication to keep it stable.

Children with Truncus Arteriosus often have other health problems, which can include Di George syndrome, 22q deletion and a small or absent thymus gland which leaves them open to infections

Most babies are well, active, and gaining weight a few days after surgery. He or she will have a scar down the middle of the

chest, and there may 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.

Unfortunately, because the conduit that has been put in doesn't grow with the child, further surgery will be needed as he or she gets bigger – perhaps another two or three open heart surgeries.

Because the flow of blood is slower than it should be in the conduit, there may be a danger of blood clotting – if this is thought to be a problem in your child's case, he or she may need to take aspirin or Warfarin. Aspirin has an anticoagulant effect (stops blood cells sticking together) and Warfarin is an anticoagulant medicine. Warfarin does need to be checked by a blood test every few weeks.

At home

While you are at home:

- You, your GP and Health Visitor should have details of your baby's condition from the heart doctor (paediatric cardiologist). If not, call the hospital at which your baby was treated, ask for the name of the paediatric cardiologist and their telephone number. Call and explain that you need the information to pass on to, for example, your local casualty Department should he or she have a sudden illness.
- You should have the number of a cardiac liaison nurse or outreach nurse to call should you have questions or any fears about your baby's heart problem.
- You should have the number of a parent support group.

The child will be monitored regularly by a cardiologist.

You may find that your child has many health problems associated with the heart defect or with a syndrome. Take advice about Disability Living Allowance if your child needs much more care than a child of his or her age, or is having problems walking.

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

- The Truncal valve may need 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.
- The electrical system of the heart is sometimes damaged, causing a very fast heart beat – a Supraventricular Tachycardia, or SVT – to occur. This may be improved with medicine or 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 Truncus arteriosus 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.

Parents' Stories

Ryan

Getting Ryan into nursery was really hard. He was born with Truncus Arteriosus, and had to have two valves put in his heart. We all got over it alright – of course we were upset to start with, but once he'd had his second operation to put a valve in to replace the leaking one we thought all the fuss was over.

He has to have warfarin every day to stop blood clots on his valve, so he tends to look a bit battered – he's a busy little boy, given to jumping off chairs, falling down steps and rolling about with his big brother, so he gets bruises.

When I took him down to the nursery to sign him up, they said they didn't have a place for him, which was a bit odd as they knew I wanted him to go with his brother. So I went to see the head, and she said that he would need special watching, and she didn't have the extra staff. What would she do if he suddenly had a heart attack.

Well, what would you do if any of the kids had a heart attack? I asked, and anyway it was very unlikely that Ryan would have a heart problem as he is seen every six months and checked out.

She said it wasn't just his heart, but that he might bleed. Fair enough, but since all children bleed at school, and they have plasters and everything, I couldn't see the point of that.

Eventually, I had to get the nurse from the hospital to talk to the school, and then they agreed to take him.

Daisy

We lost our little girl when she was just over a year old. When we discovered she had a heart problem, we thought it was the end of the world.

Through our support group we met two other families whose children had been born with Truncus Arteriosus and are now fine.

So when Daisy had surgery at three weeks old we were optimistic. The surgery seemed to go well, but the promised recovery just didn't happen. Endless infections and kidney failure kept her in intensive care for month.

We were told that she had a DiGeorge Syndrome, and that her immunity was very poor it was unlikely that she would ever be a healthy child.

We looked up DiGeorge Syndrome and were thrown into a state of misery by the number of problems Daisy could have. Mostly we feared the predicted retardation – although she didn't develop at the rate of a normal baby that was never an issue during her short life.

Came the day that we finally brought her home from the hospital – her heart function was quite good, but she still needed a nasogastric tube and oxygen and a huge range of medicines. My family were able to help out and for two blessed weeks I did everything for Daisy, and felt for the first time like our little girl's mother.

But she had a relapse, back into hospital, and this time she didn't make it home again.

Now that the first shock of losing her is over, I can smile at the memory of her smiling and waving as we came towards her hospital cot, and of the pride we felt at the place she made for herself in so many of our lives.

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