

Transposition of the Great Arteries (TGA)

What is TGA?

Transposition means 'in each other's place'.

The **Great Arteries** are:

- The pulmonary artery, which should carry blue (deoxygenated) blood from the right ventricle to the lungs.
- The aorta, which should carry red (oxygenated) blood from the left ventricle to the body.

So Transposition of the Great Arteries means that the pulmonary artery is where the aorta should be, carrying red blood from the left ventricle to the lungs, and the aorta is where the pulmonary artery should be, carrying blue blood around the body.

There may be a Ventricular Septal Defect: Ventricular means 'of the ventricles' - the pumping chambers of the heart.

Septal means 'of the septum' – the wall between the right and left sides of the heart.

Defect means a hole.

So a VSD is a hole in the wall between the ventricles. This means that blood can leak from one side to the other.

There may be other associated defects, such as coarctation of the aorta or pulmonary stenosis.

Fig 1 – TGA

Transposition of the Great Arteries

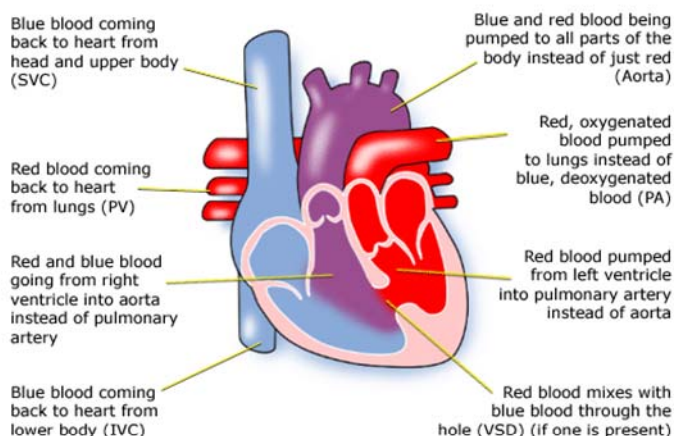
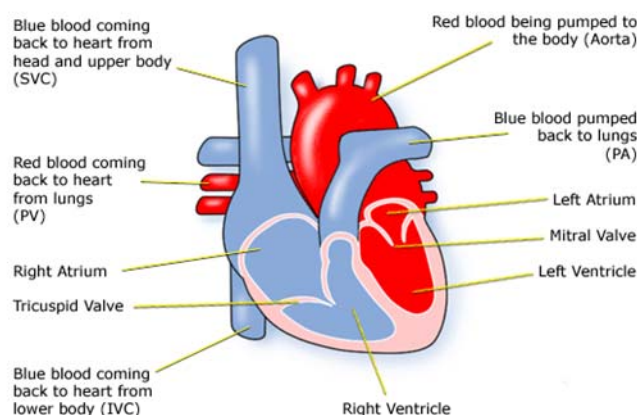


Fig 2 – Normal Heart

Normal Heart



Diagnosis

Your child's TGA may have been diagnosed on a scan during pregnancy. If so, he or she would have been taken into hospital shortly before birth to see if the prenatal assessment of the defect was accurate, and if the heart needs treatment in the shorter term

Your child may have been diagnosed after birth. Your baby might have been quite well before the foetal circulation system started to close down. This is because red blood was passing between the left and right atrium, from where it was pumped into aorta and around the body. Some of the blue blood in the aorta passed through the ductus arteriosus into the pulmonary artery and was carried to the lungs.

As the foetal circulation started to close down, your baby may have become increasingly blue (cyanosed), and so breathless that he or she was very difficult to feed.

If your child has a VSD this will allow red blood to flow from the left ventricle into the aorta, and the symptoms will not be so severe.

When a heart problem 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.

At home

You may be at home while your baby grows stronger for surgery.

- 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.

Treatment

Medication: your baby may be given medicine (Prostaglandin) to keep the foetal circulation open. This allows some deoxygenated blood to flow from the aorta into the pulmonary artery.

Balloon septostomy: Another way of increasing red blood to the aorta is to make the hole between the atria bigger. This involves threading a fine tube – a catheter – through the umbilicus (belly button) or groin, into the heart and through the hole between the atria. A balloon on the end is inflated and pulled back, so that the hole is made bigger.

Corrective surgery: this is 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 normally does – the heart bypass machine. The aim of the operation is to make the circulation of blood through the heart and lungs normal.

Usually an operation called the **Arterial switch** will be carried out in the first few days of life. The pulmonary artery and aorta are cut and reattached to the correct side of the heart. The arteries which supply the heart with red blood (coronary arteries) also need to be reattached.

The holes between the two sides of the heart are closed.

If your child has other heart defects, the kind of surgery needed will depend on how the heart can best be modified to cope with all the problems he or she has.

For most children this surgery is low risk, but it can depend on how well your child is otherwise. The doctors will discuss risks with you in detail before asking you to consent to the operation.

The length of time in hospital will usually be only 10 to 12 days, of which one or two will be spent in the intensive care and high dependency unit. Of course this depends on how well your child is before and after the surgery, and whether any complications arise.

How the child is affected

Cases of TGA can be more complicated than this description. In some cases the arterial switch operation cannot be performed, and other kinds of surgery can be offered (Rastelli, Senning, Fontan).

After surgery it is not uncommon for a baby 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 pulse rate (called tachycardia), which may need medication to keep it stable.

But most babies are completely well, pink, 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, but they will not go altogether. Smaller scars on the hands and neck usually fade away to nothing.

A child who has had an arterial switch operation will be monitored as there can be problems in the development of the arteries to the lungs (pulmonary) and to the heart itself (coronary).

Children with complicated forms of TGA who have had other procedures will often need surgery later in life.

After the first year, the child will be monitored infrequently by a cardiologist. Some children with TGA have other health problems as they get older, which may not be related to their heart condition. You will need to make sure that these are properly

diagnosed and not put down to the trauma of surgery.

- 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
- The electrical system of the heart is sometimes damaged, causing a fast or slow heartbeat – these may need to be corrected by using medicines, a pacemaker or 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 TGA 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.

Parents' stories

Emma

Fifteen years ago, I was expecting my second child and went along for a scan at about 22 weeks. I remember that the echo machine was new, and being demonstrated to some pupil midwives – I was quite happy to show off my unborn infant, but it was irritating that I couldn't see the screen.

To begin with there was lots of chortling – oh, you can see the head ... I didn't realise

they moved so much ... I can count the fingers.

The radiographer was saying look, there are the kidneys and we can get a really good look at the heart. Then she went silent. When one of the others asked her what she was looking at she said 'I'm just looking to see if I can show you all four chambers – but the baby is not in a good position.' Then she asked me if I would take a walk around so that the baby would change position. I was quite flattered that she was using me and my perfect baby to teach what foetal organs should look like.

When I went back after 15 minutes, there was a man in the room with the radiographer and the midwives had all left. She muttered something about the doctor being interested, and off we went again. They muttered between themselves, and took lots of pictures, and I asked if I could see. Then I was told that someone who specialised in hearts would want to see them, and I wasn't to worry. Do you know, my first feeling was one of betrayal. How could they have pretended to be admiring the baby when in fact they thought there was something wrong?

In fact, Emma had TGA without a VSD, and after several more scans, I was booked to give birth at a time convenient to the surgical team, so that they could operate on her as soon as she was born. As it happened I went into labour a couple of weeks early and had to have a Caesarean, Emma was whisked off, and the next time I saw her she was post op, having had her arteries switched around, been through intensive care and back on the Baby Ward.

She is checked every year, and has a mild leak through her aortic valve. She has just taken 10 GCSEs a year early and got high grades in them all. I sometimes think that

my scars from that time are much worse than hers.

Cameron

It seemed such an appalling defect to have that we simply didn't expect Cameron to grow up at all. We didn't find out until after he was born – it was difficult to persuade our GP that there was a problem, and I had to ask my sister, who works for the NHS, to come in to casualty to convince them that he was ill.

Anyway, it was a relief to know that there was a name for it, and I didn't have to watch him just fade away at home. Apparently the surgery was difficult, because of his coronary arteries, and we know that he will need surgery on his valve before he is much older.

We couldn't find any information about the kind of health problems that he might have – it was suggested that we were creating the problem by worrying about him, but we just wanted to be prepared.

Cameron was slow to talk, and still plays with his very young toys. He is immature and at 7 years old he can't read or write. Also he has problems distinguishing between fantasy and reality.

We are finding it hard to provide him with the kind of support he needs to stay at school, as apparently he does not need a special school, but we are worried that some time soon he will feel different and unhappy.

Reading through what I have written I can see it sounds as if our family relationships are very cold. I have to write objectively, because otherwise I would get emotional - he is my precious boy who still snuggles up to me to watch 'his' cartoons, and loves to play football with his Dad and younger brother.

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