PARENT GUIDE: TETRALOGY OF FALLOT





WHAT IS TETRALOGY OF FALLOT (TOF)?

Tetralogy of Fallot (TOF) is a congenital heart condition made up of four different heart defects.

Ventricular septal defect (VSD)

A defect where a hole forms between the bottom two chambers (ventricles) of the heart.

Overriding aorta

The aorta sits between the right and left ventricle, over the VSD, instead of off the left ventricle.

Pulmonary stenosis

The pulmonary valve is more narrow than normal and doesn't open properly. This makes it harder for the right ventricle to pump blood to the lungs, causing the baby to have low oxygen levels.

Right ventricular hypertrophy

The right part of the heart has to work harder, causing the right pumping ventricles to get thicker.

HOW IS TETRALOGY OF FALLOT DIAGNOSED?

During pregnancy, tests can be conducted by a doctor to check if your baby has any heart problems. However, some problems may not be present until the baby is born.

DURING PREGNANCY

A **prenatal ultrasound** can be used during pregnancy to check for any complex heart defects. This test will show if your baby's heart is not shaped correctly. If this is found, you will usually be scheduled for a foetal echocardiogram to help gather more information about your baby's heart.

A **foetal echocardiogram** is a special type of ultrasound used by paediatric doctors to look closely at your baby's heart and surrounding blood vessels. Once your baby is born, a cardiologist will assess your baby's heart again with an echocardiogram to confirm the diagnosis.

Foetal MRI's can also be used, which is a type of imaging that gives a more detailed picture of your baby's organs. It can be used to find problems with the heart or lungs.

AFTER BIRTH

As we've mentioned, after your baby is born, an **echocardiogram** can be used to get a closer look at your baby's heart. However, there are other ways in which TOF can be diagnosed after birth.

Heart catheterisation is a test to measure pressure in the heart. It can take pictures of the heart and open narrowings with a balloon or small metal coil, called a stent. This process is conducted while your baby is under anaesthesia.

Another way to diagnose TOF is by using a **cardiac MRI**. This test uses radio waves, magnets and a computer to make a detailed image of the heart and the surrounding blood vessels. Cardiac MRI's can provide detailed information on the type and severity of the heart disease.

OTHER TESTS AFTER BIRTH INCLUDE:

- **Pulse oximeter-** This involves a small sensory being clipped onto the babies fingertip, toe or ear to measure how much oxygen is in the blood.
- **Electrocardiogram-** This test records the electrical activity of the heart.
- Chest X-Rays

HOW DOES TETRALOGY OF FALLOT AFFECT THE HEART?

As we've discussed, TOF is made up of four different heart conditions. However, you may be wondering how these conditions actually affect the heart.

VENTRICULAR SEPTAL DEFECT (VSD)

VSD is a hole in the separating walls (septum) between the two lower chambers (ventricles) of the heart. The septum normally acts as a barrier that prevents oxygen-rich and oxygen-poor blood from mixing. But when there is a VSD, the two types of blood can mix. This can lead to either too much or too little blood flowing into the lungs. Children born with TOF usually have a large VSD.

OVERRIDING AORTA

An overriding aorta means the artery that carries oxygenrich blood to the body is out of place and sits above both ventricles, instead of just the left ventricle. This allows some oxygen-poor blood to flow into the aorta and out to the body instead of the pulmonary artery, which would normally take it to the lungs and collect oxygen.

PULMONARY STENOSIS

Pulmonary stenosis is the narrowing or thickening of the valve that connects the right ventricle to the pulmonary artery (the blood vessel that carries oxygen-poor blood from the heart to the lungs). With pulmonary stenosis, the heart has to work harder than a normal heart to pump blood to the lungs. There is also less blood travelling in the lungs because the pathway is narrower, resulting in less blood becoming oxygen-rich. Sometimes babies with TOF are born with pulmonary atresia, in which the pulmonary valve is completely sealed off.

RIGHT VENTRICULAR HYPERTROPHY

Right ventricular hypertrophy is a thickening of the muscular wall of the right ventricle. The thickened wall can contribute to the blocking of blood flow through the pulmonary valve, which allows blood from the heart to flow into the lungs.



WHAT CAUSES TETRALOGY OF FALLOT?

The specific cause of TOF is not known, however in some cases genetics are thought to be the main reason. For example, babies born with Down Syndrome or DiGeorge syndrome are more likely to have TOF.

Babies of mothers who have Rubella or other viral illnesses during the pregnancy are also at a higher risk of TOF.

Other pregnancy-related risks that are thought to be associated with TOF include poor nutrition, alcohol use, poorly controlled diabetes and the mothers age.

External environmental factors are also thought to increase the chance of TOF, such as air pollution.



TETRALOGY OF FALLOT SYMPTOMS

One of the most common signs of TOF is **cyanosis**, which refers to a bluish-purple colour to the skin.

Sometimes, a baby with TOF will have sudden episodes of deep cyanosis during crying or feeding. This is known as **'hypercyanotic spells'** or **'Tet spells**'.

Heart murmurs	
Fussiness	
Tiring easily	
Difficultly breathing	
Heart palpitations	
Fainting	
Clubbing of nails	

TREATMENTS FOR TETRALOGY OF FALLOT

TOF can be treated through open-heart surgery soon after the baby is born or later on during infancy. This will depend on the baby's health and weight and the severity of the defects and symptoms.

COMPLETE REPAIR

The surgeon widens the passage between the right ventricle and pulmonary artery to improve blood flow to the lungs. The VSD is patched to stop the mixing of oxygen-rich and oxygen-poor blood.

These repairs also fix the two other defects of the overriding aorta and right ventricle hypertrophy. This is because the right ventricle doesn't have to work as hard to pump blood into the lungs and the thickness of the walls will decrease. The patched VSD prevents oxygen-poor blood from flowing into the aorta.

TEMPORARY OR PALLIATIVE SURGERY

Minor repairs can improve blood flow to the lungs. This is usually only done when a baby is too weak or small to have full surgery. Your baby's surgeon will create a secondary route for blood to travel to the lungs for oxygen by placing a small tube, a shunt, between a large artery branching off the aorta and the pulmonary artery.

Once the baby is stronger, they can then be considered for the full repair surgery.

Most babies who have TOF repaired do very well after surgery, however they will need regular follow ups with a heart specialist throughout their whole life.



WHAT IS THE LIFE EXPECTANCY OF A Child with tetralogy of fallot?

It can be difficult to predict the life expectancy of a child with a TOF. However, data suggests that those who have had it repaired will **live up to 30-40 years after complete repair.**

Knowledge about the survival rates is made difficult since recent advances in surgical techniques and intensive care management have had a positive effect on the survival rates early on and late late after surgery.

OTHER CONDITIONS ASSOCIATED WITH TETRALOGY OF FALLOT

Most babies who are born with TOD don't have any other

conditions, but some do. Some babies may have a syndrome called '22q11 deletion' or DiGeorge syndrome, where a small piece of genetic material is missing from their DNA. Others may have syndromes such as Down's Syndrome.

If your baby is diagnosed with TOF, your doctor will discuss with you the options of having a test to find out if they have chromosomal abnormalities. If the TOF is diagnosed before birth, these tests can be carried out before your baby is born.

MEDICAL TERMS & WHAT THEY MEAN

AORTA The main artery of the heart that supplies oxygen-rich blood to the body.
ATRIA Either of the two upper chambers in the heart.
ATRIUM one of the two upper chambers of the heart.
CONGENITAL Present from birth.
ECHOCARDIOGRAM A scan to look at the heart and nearby blood vessels.
HEART MURMUR A whoosing or rasping sound heard during a heartbeat.
VENTRICLE One of the two lower chambers of the heart.



Respite and support of any kind is invaluable.

Lagan's Foundation, a Charitable Incorporated Organisation in England and Wales (1154208) aims to physically help support families who have infants and children up to 19 years old, diagnosed with complex health conditions specialising in heart defects and feeding difficulties. Lagan's Foundation's purpose is to provide unique at home and in hospital care to support parents in their caring role.

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