Tetralogy of Fallot Spells

What is Tetralogy of Fallot?

Tetralogy of Fallot is a congenital heart defect, which changes the way blood flows through the heart and to the lungs. The condition includes 4 main abnormalities:

1. **Ventricular septal defect (VSD)**
   
   VSD is a hole in the wall that separates the two pumping chambers (ventricles) of the heart. This hole allows oxygen-poor blood in the right ventricle to mix with oxygen-rich blood from the left ventricle. This usually results in less blood flow to the lungs.

2. **Pulmonary stenosis**
   
   The pulmonary artery is the blood vessel that carries oxygen-poor blood from the heart to the lungs. Blood picks up oxygen from the lungs and becomes oxygen-rich blood. Pulmonary stenosis is where there is a narrowing or thickening of the door (valve) that connects the right ventricle to the pulmonary artery.
   
   The narrow pathway from the right ventricle to the lungs means it is easier for oxygen-poor blood to go back around the body. As a result, less blood gets to the lungs and your child’s lips, tongue, skin or nails may appear blue (cyanosed).

3. **Right ventricular hypertrophy**
   
   This is where there is a thickening of the muscular wall of the right ventricle. The narrowing of the pathway from the right ventricle to the lungs and thickening of the pulmonary valve causes more pressure against the right ventricle to pump. The heart is a muscle and if it has to work harder, it can become thicker.

4. **An overriding aorta**
   
   The artery that carries oxygen-rich blood to the body (aorta) usually arises from the left ventricle. In Tetralogy of Fallot, the aorta sits on top of the VSD so it comes off the left ventricle and part of the right ventricle. This is another reason that oxygen-poor blood gets pumped back around the body through the overriding aorta.

How is Tetralogy of Fallot repaired?

1. **Temporary surgery:**
   
   Minor repairs can improve blood flow to the lungs and reduce cyanosis. This is usually done when an infant is too small to
have full surgery. Temporary surgery involves one of the following.

- Using a tiny wire mesh tube (called a stent) to prop open the part of the heart that carries blood out of the right ventricle to the lungs.
- Alternatively, inserting a small tube (called a shunt) between a large artery branching off the aorta and the pulmonary artery. As a result, blood can travel to the lungs for oxygen via the shunt.

2. Complete repair:
Corrective surgery is usually performed at about 4 to 6 months of age. It involves:

- Closure of the VSD with a patch.
- Enlargement of the narrowed area of the right ventricle and pulmonary artery, often needing a further patch.
- If your child has had a shunt or stent, it would be removed at this stage.

Spells are most common in young infants, around 2 to 6 months of age, while the infant is awaiting surgery. Spells often occur during feeding, crying, straining, or on waking early in the morning.

What do spells look like?

- Increased depth, rate and effort of breathing
- Worsening blue discolouration typically noticed around the mouth, nose, hands and feet
- Extreme paleness
- Prolonged irritability and crying
- Lethargy
- Severe or untreated episodes may progress to loss of consciousness or limpness

How to manage spells at home

Your child’s doctor and Cardiac Clinical Nurse Consultant (CNC) will explain the following strategies to help you understand what is expected of you when you are at home and to explain what steps to take should you need further assistance:

- Promt management: Attending to your child whilst they are having a spell as early as possible is essential as they may become unwell quickly.
- Minimise distress: Help to calm your child (e.g. offering a feed). Spells will often resolve when your child is soothed and comforted. This is because it reduces their heart rate and oxygen consumption.
- Knee-to-chest position: Pick up and cuddle your child, with your child’s knees tucked against their chest. This helps to increase the amount of blood exiting the right ventricle to the lungs.

What to do if you are worried

It is normal for any baby to change colour when crying or cold. In normal colour changes, a baby will appear well and will only change colour during crying. Their colour will return to normal after they have stopped crying. This is normal and is not a spell. If you are worried your child is not returning to normal after a few minutes, it is safest to call an ambulance (call 000).

It is helpful to notice spells and to let your Cardiac CNC, or if after business hours, the Cardiology Fellow on-call know via the hospital switchboard so that this can be factored into treatment planning.

What are “spells”?

Tetralogy of Fallot or hypercyanotic “spells” occur when your child has a quick drop in the amount of oxygen in his or her blood caused by reduced blood flow to the lungs.