Cyanotic spells
Tetralogy of Fallot

Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust
This information sheet from Great Ormond Street Hospital (GOSH) explains about cyanotic spells that occur when a baby has a congenital (present at birth) heart condition called Tetralogy of Fallot. It also explains what action to take if your child has a cyanotic spell.

Normal heart anatomy

- Aorta
- Right atrium
- Tricuspid valve
- Pulmonary artery
- Left atrium
- Mitral valve
- Left ventricle
What is Tetralogy Of Fallot?

Tetralogy of Fallot (TOF) is a combination of four problems affecting the heart which happen while a baby is developing in the womb:

These are:

- **pulmonary stenosis** – a narrowing at, under or above the valve between the right pumping chamber (right ventricle) and the large blood vessel that carries blood to the lungs to pick up oxygen (pulmonary artery)

- **ventricular septal defect** – a hole between the right and left pumping chambers (ventricles)

- **over-riding aorta** – the entrance to the large blood vessel that takes blood away from the heart to the rest of the body (aorta) is next to the ventricular septal defect, allowing oxygen-poor blood to flow through it

- **thick right ventricle** – the heart has to work harder to pump blood through the narrowed pulmonary artery, causing the muscle to thicken
Most children are sent home to gain weight and grow soon after being born as normally they do not need any special treatment. Your baby will be monitored by the Cardiac Nurse Specialist team from GOSH alongside your community health visitor, community children’s nurse and local paediatrician.

Your local team will help to monitor your baby’s weight gain and general health. Your baby should be putting on around 150-200g a week to be able to grow at a safe rate and develop until they need their surgery at around six months to a year old (or sooner if they have any symptoms). We will give you a symptom diary so that you can record any spells or other changes in your child’s health. This will help us to monitor how your child is doing and watch out for any problems developing.

**What is a cyanotic spell?**

Children with Tetralogy of Fallot will sometimes appear blue around the lips, mouth or finger beds. This is called cyanosis. This is due to a low level of oxygenated blood flowing around the body. The level of the child’s cyanosis depends on the narrowness of the pulmonary valve. Children with Tetralogy of Fallot may have a period where they become very blue or pale and floppy, which is called a hypercyanotic episode or a ‘spell’.
Common contributing factors and how to prevent a cyanotic spell

- Pain
- Crying
- Feeding
- Passing stools & wind
- Dehydration
- Fever

Not all cyanotic spells can be prevented, but you can take small steps to reduce the risk of your child having a spell such as:

- Treating any causes of pain appropriately using comfort measures and/or medication if necessary.
- Treating any high temperatures by removing excess clothing and giving over the counter medicine such as paracetamol.
- Ensuring that your child has adequate fluid intake and monitor for any signs of dehydration such as sunken fontanelle (soft spot on skull) and dry skin and mouth. Fewer wet nappies than usual – most children have six to eight wet nappies a day – can also be a sign of dehydration.
- Contact your family doctor (GP) as soon as possible if you have any general concerns regarding your child’s health.

Most episodes will get better, but if they last longer than five minutes, they can be very serious and possibly life threatening so you should call 999 for an ambulance.
What to do if your child is ‘spelling’

- Try to remain calm as this reduces your child’s stress level and oxygen need.
- Bring your baby’s knees up to their chest or encourage young children to squat – this increases the pressure going around the body and forces blood flow to the lungs.
- If the spell continues for longer than five minutes or your child deteriorates further, call an ambulance to take your child to hospital.
- If you have a discharge summary, please remember to take it with you to hospital so the staff are aware of your child’s condition.

How your child’s spell will be managed in hospital

Initial steps may include introducing a medication called propranolol which relaxes the muscle around the tube going towards the lungs (pulmonary artery).

Your child may need to have a stent (a mesh-like cage) inserted into the heart to improve blood flow going towards the lungs. The procedure will involve a small tube being inserted into their groin and guided to the correct place via x-ray under general anaesthetic.

If your child’s cyanotic spells are becoming more frequent, they may need to come to GOSH for further assessment and review of the surgical plan and treatment by your cardiologist.

Please call the Cardiac Nurse Specialist team on 020 7405 9200 ext 5774 or the Cardiology Registrar on Bear Ward on 020 7829 8829 once you have been admitted to hospital for further advice.