What is Tetralogy of Fallot?
In this condition there are four defects in the heart. These defects are:
1) Ventricular septal defect
2) Pulmonary stenosis
3) Right ventricular hypertrophy
4) Overriding aorta

What are atria and a ventricle?
There are four chambers in the heart. The two small chambers are called atria and the two large chambers are called ventricles. The right atrium receives deoxygenated blood from the body and drains in to the right ventricle, which pumps the blood to the lungs to be oxygenated. The left atrium receives oxygenated blood from the lungs and drains in to the left ventricle, which pumps the oxygenated blood to the body.

What is a ventricular septal defect (VSD)?
A septal defect is a hole in the membrane separating either the atria and/or ventricles. A ventricular septal defect is a hole between the two ventricles.

Tetralogy of Fallot (TOF or "Tet")

What is pulmonary stenosis?
The right ventricle pumps deoxygenated blood into the pulmonary artery through the pulmonary valve. Pulmonary stenosis is a narrowing of the entry into the pulmonary artery, which may involve the pulmonary valve.

http://www.schneiderchildrenshospital.org/peds_html_fixed/peds/cardiac/tf.htm
What is right ventricular hypertrophy?
Because of the narrowing of the pulmonary artery, the right side of the heart has to work harder. As a consequence the muscle of the heart gets thicker (hypertrophy).

What is an overriding aorta?
This occurs when the aorta (vessel that carries oxygenated blood from the left side of the heart to the body) sits above the VSD (hole in the heart between the two ventricles). Some of the deoxygenated (blue) blood from the right side of the heart flows through the hole in to the aorta mixing with the oxygenated (red) blood from the lungs going to the body. This may mean that your baby may carry less oxygenated blood around their body and be cyanosed (blue in colour).

Do they occur commonly?
Tetralogy of Fallot is a common cyanotic heart defect and accounts for 10% of all congenital heart defects.

How will this affect my baby?
The large hole in the middle of the heart allows oxygen rich blood from the left side of the heart to pass into the right side of the heart. This can cause more blood than usual to go to the right side of the heart to the lungs to be oxygenated. This in time can lead to the left side of the heart working too hard and failing (congestive heart failure), as well as the lung blood vessels becoming damaged due to the high pressure (pulmonary hypertension). Alternatively, if there is significant pulmonary stenosis deoxygenated (blue) blood from the right side of the heart will mix with oxygenated (red) blood from the left side of the heart which is then pumped to the body resulting in your baby being cyanosed (blue in colour). Some babies may have “blue spells” because of the pulmonary stenosis.

How is a Tetrology of Fallot diagnosed?
In some babies with this condition, the diagnosis is made on an antenatal ultrasound scan. This helps prepare the medical and nursing staff when your baby is born and to ensure that appropriate care is obtained.

After birth, when medical or nursing staff listens to your baby’s heart they may hear a heart murmur. A murmur is a noise. Like water flowing in a river, when it comes to a bend the water then makes noise. Blood is similar. When blood flows along smooth surfaces it makes little noise, but if it then flows through an opening such as a hole in the septa or a poorly functioning valve, it will make a sound. If a murmur is heard an echocardiogram (ultrasound of the heart) will be performed, which will show the defects.

How is Tetrology of Fallot treated?
The majority of babies will require surgical management in infancy and/or childhood.

Babies with Tetrology of Fallot require close observation. Signs of heart failure include a fast heart rate, fast breathing and poor weight gain. Your baby may be treated with medications that improve the function of the heart until the baby grows big enough to have heart surgery.

Sometimes the ventricular defect is so large that it causes significant heart failure which cannot be controlled with medications. In these cases early surgery may be required to band the pulmonary artery to decrease the amount of blood flowing to the lungs. This is a temporary measure until the baby is then big enough for more complex heart surgery. This will be discussed with you by the medical staff. Surgery to manage Tetrology of Fallot is done in Sydney by paediatric heart surgeons.

Until the hole is closed there is a slight risk of infected blood clots attaching to the edges of the hole (bacterial endocarditis). This can make a baby or child very unwell. These infected clots are usually associated with dental or other surgical procedures. You should inform your dentist or surgeon of the Tetrology of Fallot and your baby should be given antibiotics at the time of the procedure. If there is ongoing pulmonary valve involvement after your baby has had all of their corrective surgery, lifelong antibiotic prophylaxis for surgery and dental procedures may be required.

If you have any further questions please ask the medical and nursing staff.

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