

What is Tetralogy of Fallot?

Tetralogy of Fallot (T.O.F.) is the most common type of heart defect that causes a baby to turn blue. There are four components to T.O.F. 1) a large hole between the ventricles, 2) a relatively large aorta that straddles the hole, 3) narrowing of the pulmonary artery (the artery that goes to the lung), and 4) abnormal thickening of the right ventricle.

About one third of T.O.F. will turn blue during the first three months of life. These are the most severe forms and usually need to have surgery early on. At the present time, most major pediatric heart centers are fixing children with Tetralogy of Fallot within the first three months of life depending on when they begin to turn blue or have oxygen saturations of less than 84%. Best results are obtained if the baby can grow as much as possible. We try to avoid surgery in the first month if possible because babies that young tend to have more problems with swelling and more difficulty with post-op care. Surgery can be performed on babies as small as 3 Kg if need be. About one third of babies will turn blue during the next three to six months of life and these children may have a complete repair at the time of diagnosis. The remainder may not ever turn blue, but have symptoms of a big hole between the ventricles, and can be repaired electively around 3 to 6 months of age. These days surgeons use BT shunts only in very small babies or where the pulmonary arteries are very small and need to grow. There is a condition call **Tetralogy of Fallot with Pulmonary atresia** in which there is a complete separation of the main pulmonary artery from the right ventricle and no forward blood flow occurs from the right ventricle to the pulmonary artery. These cases are handled by placing shunts early on to the pulmonary arteries and trying to establish a connection with the right ventricle, also leaving the VSD open temporarily. These babies are somewhat blue until the repair is completed.

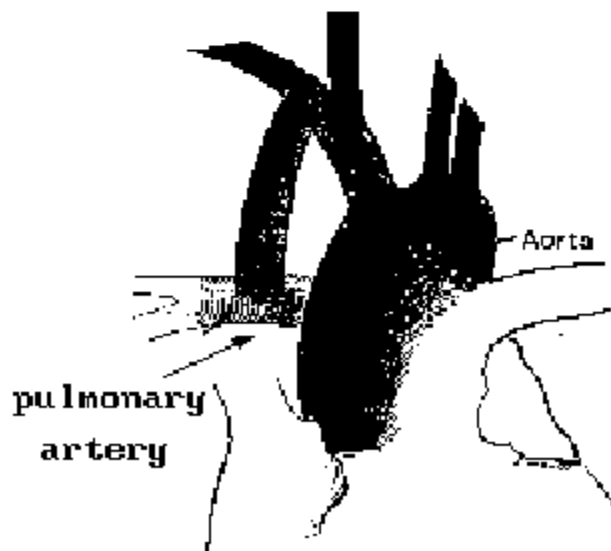
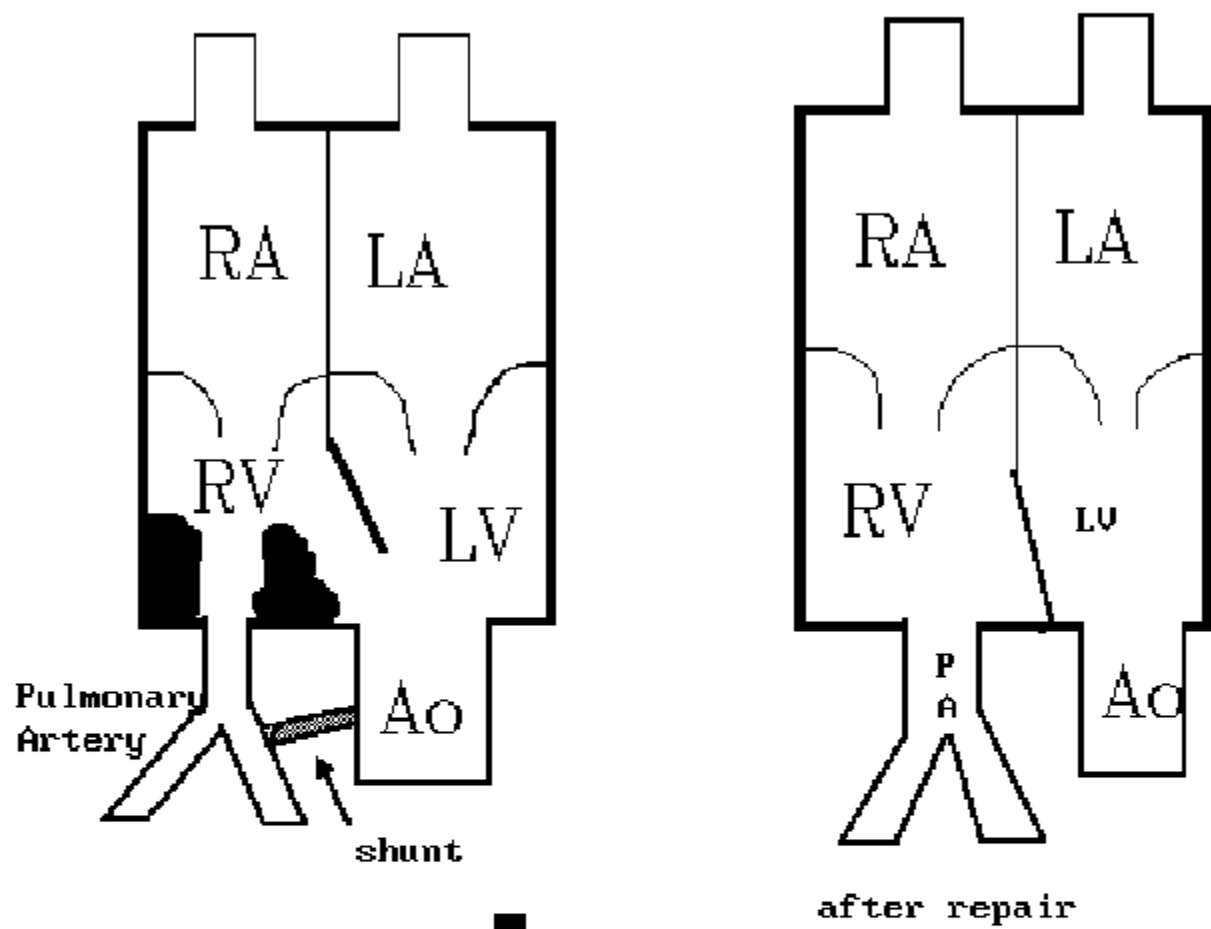
The most serious problem that happens with T.O.F. is that the child can turn blue. This is because not enough blood is getting to the lungs. When this happens, not enough oxygen goes to the body and in particular the brain. Patients with T.O.F. can gradually become blue because of progressive thickening of the right ventricle or they may suddenly turn blue when having a "TET spell". This occurs when the muscle of the right ventricle and pulmonary arteries go into spasm, or tighten down. When patients turn very blue it is a medical emergency and needs to be treated right away.

The repair of T.O.F. requires open-heart surgery. The surgeon must first place a dacron (plastic) patch over the hole between the ventricles. He must then remove any excess muscle in the right ventricle and sometimes put a dacron patch over the area where the pulmonary artery connects to the right ventricle. Sometimes he may need to replace or repair the pulmonic valve and enlarge the branch pulmonary arteries with a patch. It is not unusual to need to replace the pulmonic valve with a pulmonary Homograph " which is a pulmonary valve and artery harvested from a cadaver. Sometimes it is necessary to delay complete repair if the coronary arteries cross the area that needs to be cut open. This happens in less than 5% of the cases.

We do not know the cause of T.O.F. Usually it is a sporadic event and not related to anything the mother did. On occasion it may be related to a syndrome called 22P Deletion or other chromosomal abnormalities. It is sometimes related to **DiGeorge syndrome** which also has immune deficiencies associated with it. T.O.F. is sometimes associated with a medicine used to treat acne called Acutane. The good news is that most of the time children with T.O.F. can have a successful repair and lead useful, active lives. Girls can have babies. There is a 10-13% chance of them having a child with some type of heart defect. The Tetralogy heart is never made normal with surgery and follow up over a lifetime with a cardiologist is necessary. Sometimes children with repaired Tetralogy of Fallot will develop irregular heartbeats that may require treatment with medication or catheter ablation. As these children get older we are finding that many of them will need another operation to replace the pulmonic valve if it leaks significantly to prevent the right ventricle from enlarging. Many times the pulmonic valve can be replaced with an artificial valve. Sometimes it is necessary to further enlarge the pulmonary arteries, especially if they were small to start with.. The main goal is to preserve the function of the right ventricle. Patients with T.O.F. whether repaired or not need to take antibiotics before dental treatments (including cleaning) and surgeries because they are at risk of developing an infection of the heart.

If you have any questions, please ask one of the doctors.

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Modified Right B-T shunt