

Tetralogy of Fallot

What is it?

Tetralogy of Fallot has four key features. A ventricular septal defect (VSD; a hole between the ventricles) and obstruction from the right ventricle to the lungs (pulmonary stenosis) are the most important. Also, the aorta (the major artery from the heart to the body) lies directly over the ventricular septal defect, and the right ventricle develops thickened muscle.

What causes it?

In most children, the cause of tetralogy of Fallot isn't known. It's a common type of heart defect. It may be seen more commonly in children with Down syndrome or DiGeorge syndrome. Some children can have other heart defects along with tetralogy of Fallot.

How does it affect the heart?

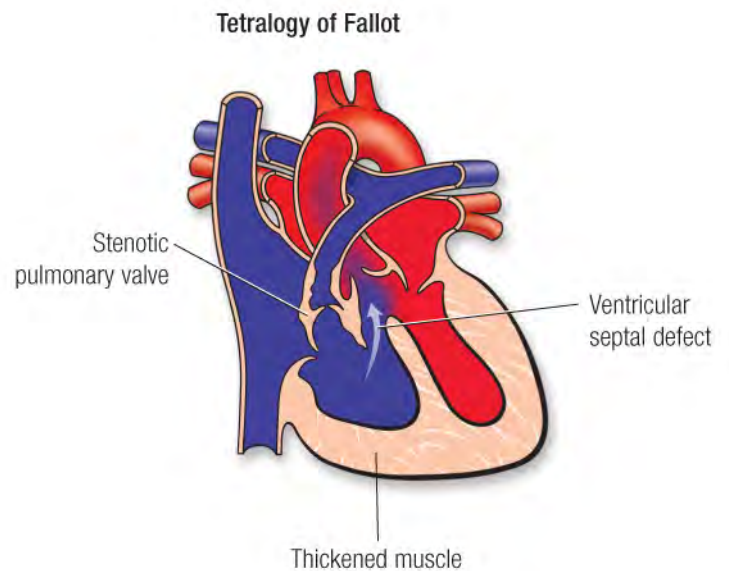
Normally the left side of the heart only pumps blood to the body, and the heart's right side only pumps blood to the lungs. In a child with tetralogy of Fallot, blood can travel across the hole (VSD) from the right pumping chamber (right ventricle) to the left pumping chamber (left ventricle) and out into the body artery (aorta). Obstruction in the pulmonary valve leading from the right ventricle to the lung artery prevents the normal amount of blood from being pumped to the lungs. Sometimes the pulmonary valve is completely obstructed (pulmonary atresia).

How does tetralogy of Fallot affect my child?

Infants and young children with unrepaired tetralogy of Fallot are often blue (cyanotic). The reason is that some oxygen-poor blood is pumped to the body through the hole in the wall between the right and left ventricle instead of being pumped to the lungs.

What can be done about tetralogy of Fallot?

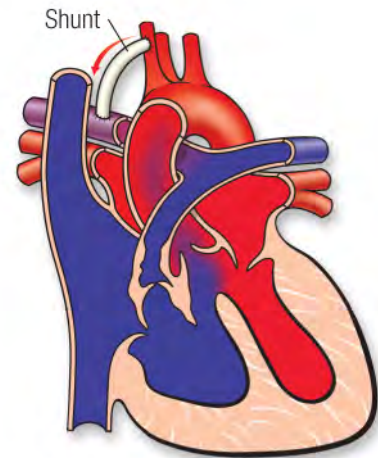
Tetralogy of Fallot is treated surgically. A temporary operation may be done at first if the baby is small or if there are other problems. Complete repair comes later. Sometimes the first operation is a complete repair.



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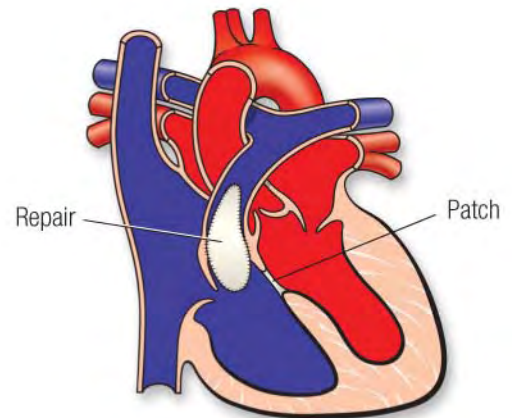
Temporary Operation

In some infants, a *shunt* operation may be done first to provide adequate blood flow to the lungs. This is not open-heart surgery and doesn't fix the inside of the heart. The shunt is usually a small tube of synthetic material sewn between a body artery (or the aorta) and the pulmonary artery. The shunt is closed when a complete repair is done later.



Complete Repair

Complete repair tends to be done early in life. The surgeon closes the ventricular septal defect with a patch and opens the right ventricular outflow tract by removing some thickened muscle below the pulmonary valve, repairing or removing the obstructed pulmonary valve and, if needed, enlarging the branch pulmonary arteries that go to each lung.



Sometimes a tube is placed between the right ventricle and the pulmonary artery. This is sometimes called a Rastelli repair. It's similar to the type of repair used for some other heart defects.

Will my child's activities be limited?

Your child may need to limit physical activity, particularly for competitive sports, if there is leftover obstruction or leak in the pulmonary valve, which is common after repair. Children with decreased heart function or rhythm disturbances may need to limit their activity more.

If the tetralogy has been repaired with surgery, and there's no obstruction or leak in the pulmonary valve, your child may be able to participate in normal activities without much increased risk.

Your child's pediatric cardiologist will help decide if your child needs limits on physical activity.

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What will my child need in the future?

If your child has had tetralogy of Fallot repaired, he or she will need regular follow-up with a pediatric cardiologist. As an adult, your child will need lifelong regular follow-up with a cardiologist who's had special training in congenital heart defects.

Some long-term problems can include leftover or worsening obstruction between the right pumping chamber and the lung arteries. Children with repaired tetralogy of Fallot have a higher risk of heart rhythm disturbances called arrhythmias. Sometimes these may cause dizziness or fainting.

Generally, the long-term outlook is good, but some children may need medicines, heart catheterization or even more surgery.

What about preventing endocarditis?

Children with tetralogy of Fallot are at increased risk for endocarditis. Some children, including those who have had a valve replacement, still have a shunt or have leaks around surgical patches, and need to take antibiotics before certain dental procedures to help prevent endocarditis.