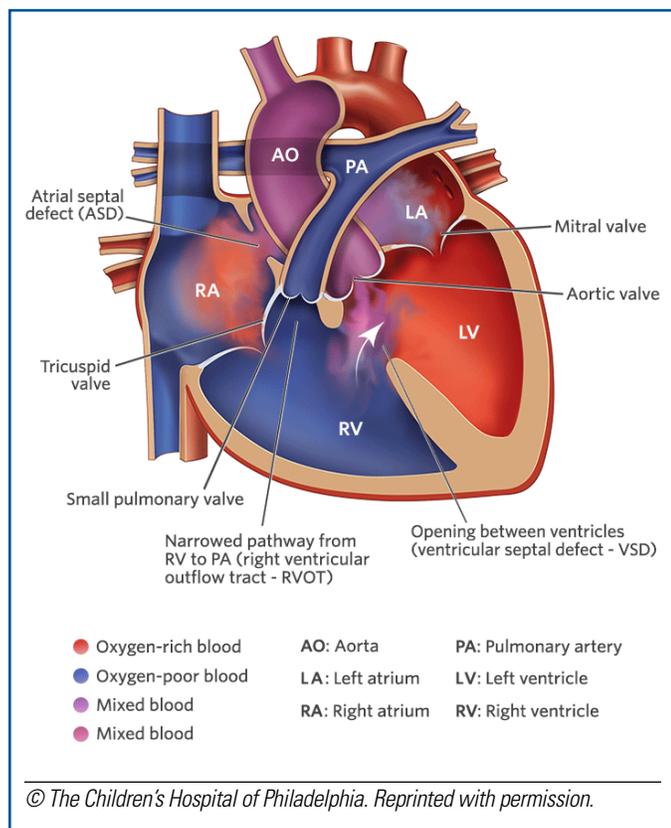




Cardiac Defects: Tetralogy of Fallot



Tetralogy of Fallot has four characteristics:

- Ventricular septal defect (VSD)—There is a hole between the two bottom chambers (the ventricles) of the heart that eject blood to the body and lungs.
- Overriding aorta—The aorta, the large artery that takes blood to the body, is on top of both ventricles, instead of just the left ventricle as in a normal heart.
- Pulmonary stenosis—There is a narrowing of the pulmonary valve, the area below the valve, or the pulmonary arteries, which carry blood from the heart to the lungs.
- Hypertrophy—The right ventricle becomes thicker and more muscular than normal as a result of working harder to pump blood through the narrow pulmonary valve.

Symptoms

The symptoms of tetralogy of Fallot include

- blue or purple tint to lips, skin, and nails (cyanosis)

- heart murmur—the heart sounds abnormal when a doctor listens with a stethoscope
- abnormal shape of the fingertips (“clubbing”) in older children
- spells in which oxygen levels drop—lips and skin will become bluer, and the child will become fussy or irritable and then sleepy or unresponsive.

How is tetralogy of Fallot diagnosed?

Tetralogy of Fallot may be diagnosed with fetal echocardiogram (ultrasound). Your baby’s provider will prepare a plan for delivery and care immediately after birth.

Your baby’s providers might make the tetralogy of Fallot diagnosis before your baby leaves the hospital if they hear a murmur or see a blue tint to the skin. A primary care provider might detect the same symptoms during a checkup, or you might notice the symptoms and bring your baby to a doctor or hospital.

Diagnosis of tetralogy of Fallot may require some or all of these tests:

- pulse oximetry—a painless way to monitor the oxygen content of the blood
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- echocardiogram (also called echo or ultrasound)—sound waves create an image of the heart
- chest X ray
- cardiac magnetic resonance imaging—a three-dimensional image shows the heart’s abnormalities
- cardiac catheterization—a thin tube (catheter) is inserted into the heart through a large vein in the leg.

A number of children with tetralogy of Fallot also have genetic syndromes such as DiGeorge syndrome (22q11 deletion syndrome), Trisomy 21 (Down syndrome), or Alagille syndrome. Genetic testing (a blood test) may be part of the evaluation.



What are the treatment options for tetralogy of Fallot?

Surgery is required to repair tetralogy of Fallot. Typically, in the first few months of life, surgeons will perform open-heart surgery to patch the hole and widen the pulmonary valve or artery. In some cases, depending on the unique needs of the patient, they will perform a temporary repair until a complete repair can be done. The temporary repair involves connecting the pulmonary arteries (which carry blood from heart to lungs) with one of the large arteries that carry blood away from the heart to the body. This increases the amount of blood that reaches the lungs, and so increases the amount of oxygen in the blood. Both the primary complete surgical repair and the staged repair give comparable outcomes, but infants undergoing the staged repair might have better neurodevelopmental outcomes (associated with early surgery and shorter length of stay in the hospital; Bailey, Mercer-Rosa, Mascio, Elci, & Goldmuntz, 2016).

What is the follow-up care for tetralogy of Fallot?

Through Age 18

A child who has had surgical repair of tetralogy of Fallot will require lifelong care by a cardiologist.

Pediatric cardiologists follow patients until they are young adults, coordinating care with the primary care provider. Parents will need to carefully follow the advice of their baby's provider, including staying on any medications prescribed and, in some cases, limiting exercise.

Sometimes children with tetralogy of Fallot experience heart problems later in life, including a leaky heart valve and irregular heartbeat (arrhythmia). Medicine or repeat surgery may be required.

Into Adulthood

Because of enormous strides in medicine and technology, today most children born with heart conditions like tetralogy of Fallot go on to lead healthy, productive lives as adults.

References

- Bailey, J., Mercer-Rosa, L., Mascio, C., Elci, O., & Goldmuntz, E. (2016). Staged versus complete repair in the symptomatic neonate with tetralogy of fallot: in search of the optimal strategy. *Circulation, 134*, A16954.

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