

## **Tetralogy of Fallot**

### **Introduction**

Tetralogy of Fallot (teh-TRAL-uh-jee of fuh-LOW) is a rare condition caused by a combination of four heart defects that are present at birth (congenital).

These defects, which affect the structure of the heart, cause oxygen-poor blood to flow out of the heart and to the rest of the body. Infants and children with tetralogy of Fallot usually have blue-tinged skin because their blood doesn't carry enough oxygen.

Tetralogy of Fallot is often diagnosed during infancy or soon after. However, tetralogy of Fallot might not be detected until later in life in some adults, depending on the severity of the defects and symptoms.

With early diagnosis followed by appropriate surgical treatment, most children and adults who have tetralogy of Fallot live relatively normal lives, though they'll need regular medical care throughout life and might have restrictions on exercise.

### **What Is Tetralogy of Fallot?**

The main function of your heart is to pump blood through the rest of your body. This blood gives your body nutrients and oxygen. If your heart doesn't work properly, it may not be efficient in pumping blood to other organs. This affects the amount of oxygen getting to other bodily tissues. Sometimes, there are defects or problems with the heart when a person is born. These defects are known as congenital heart defects.

Tetralogy of Fallot (TOF) is a congenital heart defect that can be fatal if it's left untreated. It's also known as "tet." The "tetra" in the name of the condition comes from the four problems associated with it. The condition is named after Dr. Etienne Fallot.

The four heart defects associated with TOF are:

- a hole between the right and left ventricles, which is also called a ventricular septal defect (VSD)
- a narrow pulmonary outflow tract, which connects the heart with the lungs
- a thickened right ventricle
- an aorta that has a shifted orientation and lays over the VSD

The condition causes cyanosis. This means it causes the skin to have a bluish hue due to the lack of circulating oxygen. Typically, oxygenated blood gives the skin its pinkish hue.

## Symptoms

Tetralogy of Fallot symptoms vary, depending on the extent of obstruction of blood flow out of the right ventricle and into the lungs. Signs and symptoms may include:

- A bluish coloration of the skin caused by blood low in oxygen (cyanosis)
- Shortness of breath and rapid breathing, especially during feeding or exercise
- Loss of consciousness (fainting)
- Clubbing of fingers and toes — an abnormal, rounded shape of the nail bed
- Poor weight gain
- Tiring easily during play or exercise
- Irritability
- Prolonged crying
- A heart murmur

### *Tet spells*

Sometimes, babies who have tetralogy of Fallot will suddenly develop deep blue skin, nails and lips after crying or feeding, or when agitated.

These episodes are called tet spells and are caused by a rapid drop in the amount of oxygen in the blood. Tet spells are most common in young infants, around 2 to 4 months old. Toddlers or older children might instinctively squat when they're short of breath. Squatting increases blood flow to the lungs.

### *When to see a doctor*

Seek medical help if you notice that your baby has the following symptoms:

- Difficulty breathing
- Bluish discoloration of the skin
- Passing out or seizures
- Weakness
- Unusual irritability

If your baby becomes blue (cyanotic), place your baby on his or her side and pull your baby's knees up to his or her chest. This helps increase blood flow to the lungs. Call 911 or your local emergency number immediately.

## Causes

Tetralogy of Fallot occurs during fetal growth, when the baby's heart is developing. While factors such as poor maternal nutrition, viral illness or genetic disorders might increase the risk of this condition, in most cases the cause of tetralogy of Fallot is unknown.

The four abnormalities that make up the tetralogy of Fallot include:

- **Pulmonary valve stenosis.** Pulmonary valve stenosis is a narrowing of the pulmonary valve — the valve that separates the lower right chamber of the heart (right ventricle) from the main blood vessel leading to the lungs (pulmonary artery).

Narrowing (constriction) of the pulmonary valve reduces blood flow to the lungs. The narrowing might also affect the muscle beneath the pulmonary valve. In some severe cases, the pulmonary valve doesn't form properly (pulmonary atresia) and causes reduced blood flow to the lungs.

- **Ventricular septal defect.** A ventricular septal defect is a hole (defect) in the wall (septum) that separates the two lower chambers of the heart — the left and right ventricles. The hole allows deoxygenated blood in the right ventricle — blood that has circulated through the body and is returning to the lungs to replenish its oxygen supply — to flow into the left ventricle and mix with oxygenated blood fresh from the lungs.

Blood from the left ventricle also flows back to the right ventricle in an inefficient manner. This ability for blood to flow through the ventricular septal defect reduces the supply of oxygenated blood to the body and eventually can weaken the heart.

- **Overriding aorta.** Normally the aorta — the main artery leading out to the body — branches off the left ventricle. In tetralogy of Fallot, the aorta is shifted slightly to the right and lies directly above the ventricular septal defect.

In this position the aorta receives blood from both the right and left ventricles, mixing the oxygen-poor blood from the right ventricle with the oxygen-rich blood from the left ventricle.

- **Right ventricular hypertrophy.** When the heart's pumping action is overworked, it causes the muscular wall of the right ventricle to thicken. Over time this might cause the heart to stiffen, become weak and eventually fail.

Some children or adults who have tetralogy of Fallot may have other heart defects, such as a hole between the heart's upper chambers (atrial septal defect), a right aortic arch or abnormalities of the coronary arteries.

## Risk factors

While the exact cause of tetralogy of Fallot is unknown, various factors might increase the risk of a baby being born with this condition. These risk factors include:

- A viral illness during pregnancy, such as rubella (German measles)
- Alcoholism during pregnancy
- Poor nutrition during pregnancy
- A mother older than age 40
- A parent who has tetralogy of Fallot
- The presence of Down syndrome or DiGeorge syndrome

## Complications

All babies who have tetralogy of Fallot need corrective surgery. Without treatment, your baby might not grow and develop properly.

Your baby may also be at an increased risk of serious complications, such as infective endocarditis — an infection of the inner lining of the heart or heart valve caused by a bacterial infection.

Untreated cases of tetralogy of Fallot usually develop severe complications over time, which might result in death or disability by early adulthood.

## Components of tetralogy of fallot?

**Pulmonary stenosis** – narrowing of the right ventricular outflow tract

**Ventricular septal defect** – a hole between the left and right ventricle

**Overriding aorta** –the aortic valve is situated on top of both the left and right ventricle - due to the VSD

**Right ventricular hypertrophy** – an increase in muscle mass of the right ventricle as a compensatory mechanism due to the cianotic (undersaturated) blood flow

## What Is the Treatment for Tetralogy?

Treatment of TOF requires surgery. This usually occurs within the first few months of life. The surgery includes closing the VSD and enlarging the pulmonary valve. According to the Children's Hospital of Philadelphia, if doctors can't perform a complete repair, they'll perform a temporary repair until a complete surgery is possible.

If TOF is left untreated, it can cause problems with heart rhythms, developmental delays, and seizures. If the condition is never fixed, which is rare, it typically causes death by the age of 20 years old. Usually, a doctor will notice the condition early on and perform surgery to correct the problem.

After having surgery for TOF, a person will need to see a cardiologist for the rest of their life. A cardiologist will perform regular follow-up exams and consult with the person's primary care physician on any medications or health problems that are present. Some people who have surgery for TOF develop heart problems as time goes on, which makes consistent and continuous care important.

### REFERENCES

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