Tetralogy of Fallot

What is a Tetralogy of Fallot?

Tetralogy of Fallot (TOF) is a congenital heart defect. Congenital means present at birth. They occur early in pregnancy often before a mother even knows she is pregnant. A defect of the heart can vary in severity from a simple problem to a more serious abnormality.

TOF includes four heart problems:

1. A large ventricular septal defect (VSD)

The heart has two ventricles, the left ventricle and the right ventricle. These are the lower pumping chambers of the heart. The right ventricle pumps blood to the lungs to receive oxygen and the left ventricle pumps this oxygenated blood out of heart to the body.

A VSD is a “hole” (an opening) in the wall between these two pumping chambers. This causes oxygen-rich blood to leak from the left side of the heart, where the pressure is higher, to the right side of the heart. The blood from the right side is pumped backed to the lungs.

2. Pulmonary Stenosis

Pulmonary stenosis is a narrowing of the passageway through which blood flows from the right ventricle to the lungs.

3. Right Ventricular Hypertrophy
Because of the pulmonary stenosis, not enough blood can get to the lungs so the heart has to work harder than normal to pump blood to the lungs and as a result the right ventricle thickens. This thickening is called ventricular hypertrophy.

4. Overriding Aorta

In a normal heart, the aorta, the main blood vessel taking blood to the body is directly attached to the left ventricle, which allows only the oxygen-rich blood to be taken to the body. In TOF, the aorta is shifted to between the left and right ventricles. As a result, the oxygen-poor blood from the right ventricle can flow into the aorta and out to the body instead of into the pulmonary artery to the lungs.

**What are the causes of TOF?**

For every 10,000 babies born, approximately 5 will have TOF.

Some congenital heart defects may have a genetic link causing heart problems to occur more often in certain families. Some babies with TOF have a chromosomal abnormality or other genetic problem.

But most of the time TOF occurs by chance and it appears to be a random event that can happen to anyone.

**What does this mean?**

During pregnancy, TOF is well tolerated by the baby as the baby does not need to use his/her lungs and the heart does not need to send blood to the lungs to pick up oxygen. The placenta and umbilical cord carries oxygen to the baby.

All babies born with TOF need heart surgery. You will meet with a paediatric cardiovascular surgeon (specialist in heart surgery for children) who will be able to describe the surgery in detail and outline the expected outcomes.

**What other tests should we consider?**

There is a chance that there are other heart defects and organ abnormalities so it is important to check the baby for other problems. Further tests include a detailed ultrasound that is used to assess the baby’s other organs. A fetal echocardiogram, which is a detailed ultrasound of your baby’s heart, will also be done.

An amniocentesis to look for genetic or chromosome problems will be offered. During an amniocentesis, a small amount of amniotic fluid is taken from the area around the baby and tested for genetic problems.

**What will happen around the time of the baby’s birth?**
Babies with TOF will need to be born at a hospital with a paediatric cardiovascular surgeon. Babies with TOF are closely monitored in the intensive care unit and further testing after birth will help the paediatric cardiologist (heart specialist for children) plan your baby’s care.

This level of care may not be available in your hospital and you may need to deliver at another centre. Your doctor will be able to tell you where your baby needs to be delivered.

**What does this mean for my baby’s future?**

The paediatric cardiologist will be able to discuss with you in detail what this means for your baby’s future and how it may affect his/her life.

**What do we do now?**

You will meet with a doctor that specializes in high-risk obstetrics. A referral to a paediatric cardiologist (heart specialist for children) will be made to discuss with you the care the baby will need after birth. These doctors will discuss with you in detail your options for further testing, discuss with you test results and provide you with treatment options. They will be able to answer any questions you may have.

**Where can I get more information?**

The Children’s Hospital of Philadelphia
Tetralogy of Fallot (TOF)
http://www.chop.edu/service/cardiac-center/heart-conditions/tetralogy-of-fallot.html

American Heart Association
Tetralogy of Fallot
http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Tetralogy-of-Fallot_UCM_307038_Article.jsp

National Heart Lung and Blood Institute
What Is Tetralogy of Fallot?
http://www.nhlbi.nih.gov/health/health-topics/topics/tof/