



Tetralogy *of* Fallot

A GUIDE TO HELP UNDERSTAND YOUR BABY'S HEART



PROVIDED BY THE
CENTER *for* ADVANCED FETAL CARE
the FETAL HEART PROGRAM



UNIVERSITY *of* MARYLAND
MEDICAL CENTER

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This guide will help you understand your child's heart. It is not a diagnosis and should never be used instead of medical advice.

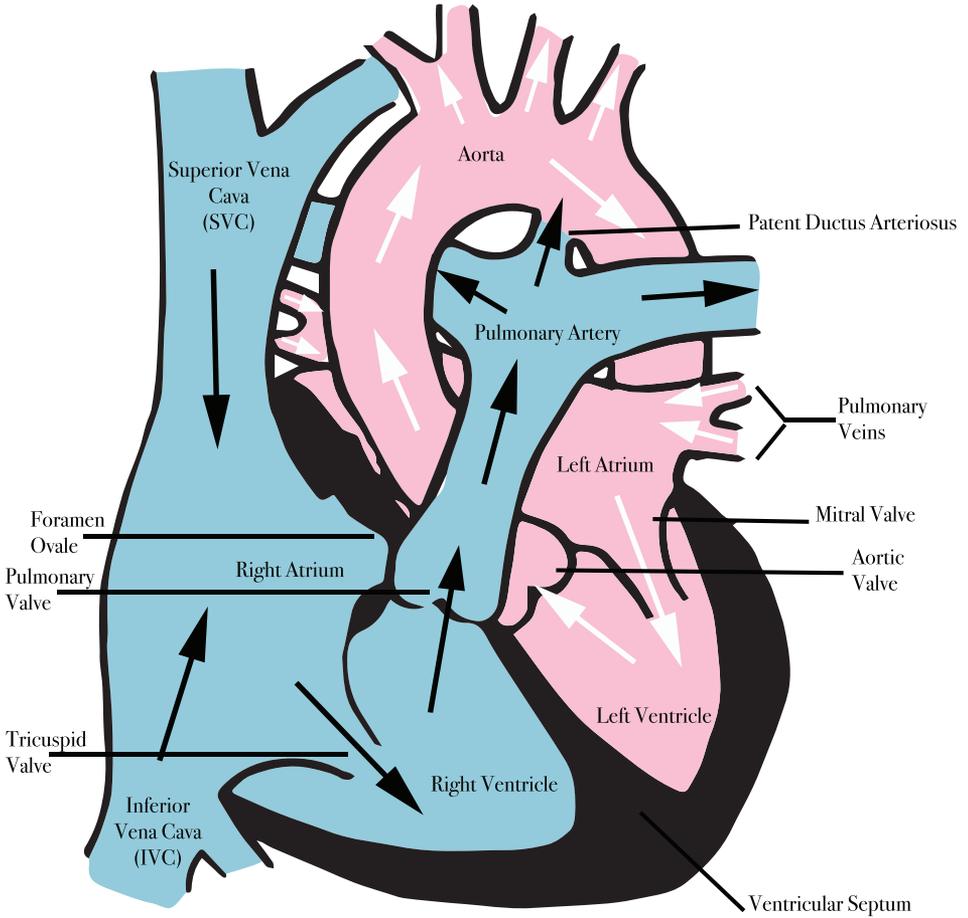
Our goal at the Center for Advanced Fetal Care is to assist you on the journey ahead and help educate you to better communicate with your team of physicians, friends and family.

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Illustrations by Dr. Alicia Chaves



The Normal Heart



The heart is a complex organ which pumps blood through the body. It drives the circulatory system, which carries oxygen and nutrients to the vital organs through a system of arteries and veins.

The heart has four chambers. The top two chambers are called the atria, which are separated by the atrial septum. The bottom two chambers are the ventricles, which are separated by the ventricular septum. As blood passes through each individual chamber, it exits through a valve. Each side of the heart works as its own pump.

Pump 1 - Right side pumps blood to the lungs. (blue on diagram) Blood travels from the right atrium through the tricuspid valve into the right ventricle. From the right ventricle, it travels through the pulmonary valve into the pulmonary artery to the lungs. Oxygenated blood from the lungs returns to the left atrium through the pulmonary veins.

Pump 2 - Left side pumps blood to the body. (pink on diagram) From the left atrium, oxygenated blood travels through the mitral valve to the left ventricle. Then, blood exits through the aortic valve to the aorta, the main artery, sending blood to the rest of the body. Once blood has supplied oxygen to the vital organs, it returns to the right atrium through the inferior vena cava (IVC) and the superior vena cava (SVC), the main veins of the body, to begin the process again.



What is a congenital heart defect?

A congenital heart defect is an abnormality of the structure and/or function of the heart. The defect typically develops during the early stages of pregnancy.

Why did this happen to my baby?

A congenital heart defect is the most common abnormality found in babies. Congenital heart disease may occur due to environmental factors, chromosome abnormalities, or genetic conditions; the majority of heart defects are multifactorial, which means that it occurs because of interactions between genes, chance, and the environment. In most cases, there is not an explanation for why a baby is born with a heart defect.

It is important to remember that any baby can have a congenital heart defect. Neither parent is to blame. Our hospital offers monthly support groups for families with children born with a congenital heart defect.

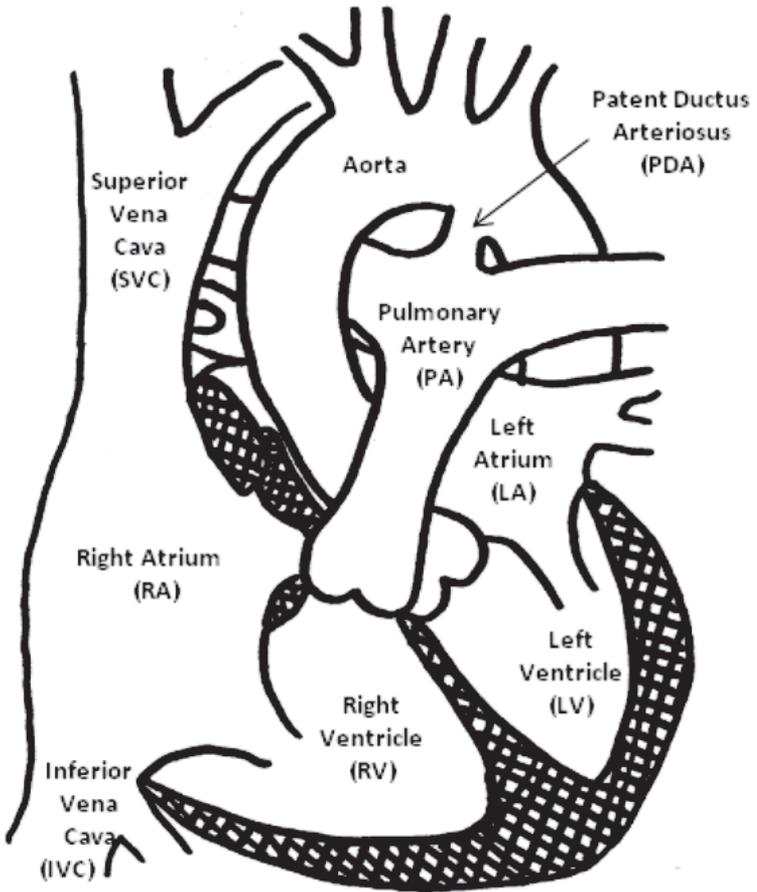
For more information, visit the fetal heart program online at: umm.edu/programs/fetalheart/patient-information/resources



What is Tetralogy of Fallot?

Tetralogy of Fallot (TOF) is a congenital heart defect (CHD). There are four defects that occur together: ventricular septal defect (VSD), overriding aorta, pulmonary stenosis, and right ventricular hypertrophy.

Normal Heart



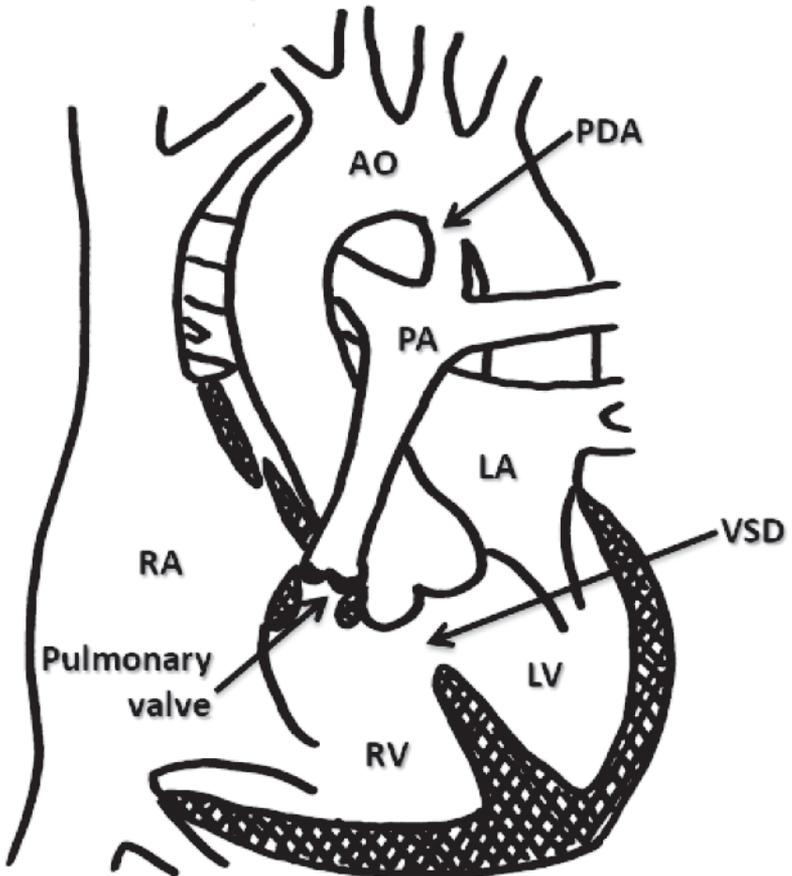
VSD: a hole between the 2 ventricles of the heart. (sometimes referred to as a "hole in the heart").

Overriding aorta: condition when the aorta is shifted over the VSD instead of the left ventricle.

Pulmonary stenosis: narrowing of the pulmonary valve, making it more difficult to pump blood to the lungs.

Right ventricular hypertrophy: thickening of the right ventricular wall caused by pulmonary stenosis.

Tetralogy of Fallot



For more information visit the fetal heart program online at:
umm.edu/programs/fetalheart/health-information/services/tof

How is Tetralogy of Fallot Diagnosed?

TOF is diagnosed with an echocardiogram (ultrasound of the heart). A severe case of TOF may be diagnosed as early as 12 weeks gestation. In general, targeted anatomy ultrasound at 18-23 weeks is the best time to identify TOF. A milder case of TOF might not be diagnosed before birth. TOF may be diagnosed after birth if the pediatrician hears a murmur (abnormal heart sound) or if the baby has bluish lips and skin. This bluish appearance is called “cyanosis”, which is the result of the baby’s blood not having enough oxygen.

What causes Tetralogy of Fallot?

The exact cause of TOF is unknown. There are some chromosome conditions and genetic syndromes that can be associated with TOF, like Down syndrome or DiGeorge syndrome (22q11 deletion syndrome). A baby who has TOF because of a chromosomal or genetic condition usually has physical and developmental problems in addition to their heart defect. During pregnancy, either a chorionic villus sampling (CVS) or an amniocentesis can test for many of these chromosomal conditions. These procedures carry a small risk of miscarriage; however, many providers and patients believe that the benefits outweigh the risks. If a family chooses not to have these tests during pregnancy, all babies with TOF will be checked for genetic and chromosomal conditions as a newborn. Most people with TOF do not have a genetic syndrome. TOF does not typically run in families. In general, congenital heart defects are slightly more common if there is a close relative with any kind of congenital heart defect.

What can I expect during my pregnancy?

If your baby is diagnosed before birth with TOF, a team of specialists will care for you and your unborn baby. The team at the University of Maryland Medical Center includes a maternal fetal medicine specialist (MFM), cardiologists (fetal and pediatric), genetic counselor, neonatologist, and a pediatric cardiac surgeon. Your baby will be monitored closely by fetal ultrasounds and a delivery plan will be discussed among you, your obstetrician, and the various other specialists. Induction of labor may be scheduled for a pregnancy affected by TOF to ensure that your care team is present at delivery. If there are no maternal or fetal issues other than the baby’s TOF, our goal is for you to have a normal delivery.

What can I expect after my baby is born?

Surgery is necessary for babies born with TOF. The timing of the surgery is dependent on how your baby adjusts as a newborn. After delivery, your baby will be taken to the NICU, where specialists will closely monitor his/her condition. An echocardiogram will be performed shortly after birth. Based on the results, a plan will be made for your baby's needs.

Most babies with TOF do well after delivery. If the oxygen level is normal, and the baby is breathing well, he/she will be monitored for a few days, and then go home. In this case, surgery will not be necessary until your baby is approximately 4-6 months old.

If the oxygen level is low, your baby may need a medication called prostaglandins. This medication keeps open the blood vessel patent ductus arteriosus (PDA). The PDA is a vessel that connects the pulmonary artery and aorta. In all babies, it is open before birth and then closes within the first few days after birth. When open, it can supply extra blood to the lungs. If prostaglandins is necessary, your baby will most likely require surgery before going home. A baby with pulmonary atresia, the most severe form of TOF, will require surgery.

Any baby with TOF can turn blue for short periods of time, especially when crying or pooping. Babies having a "blue spell" may become very irritable, sleepy or even unresponsive. Most of these spells last a few minutes and can be treated by comforting the baby or flexing the baby's knees towards the chest. Sometimes, the spells are more severe and do not go away. A severe spell is rare, but may be an emergency.

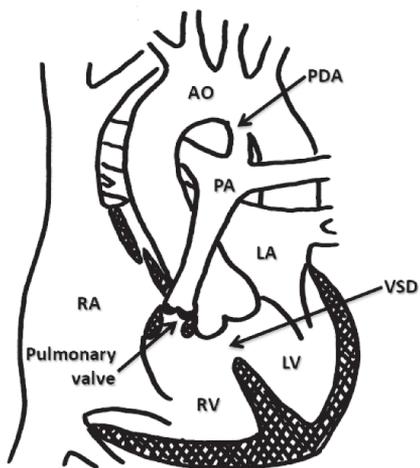
How is Tetralogy of Fallot treated?

Your baby's oxygen level will determine when surgery occurs. A baby without enough blood flow to the lungs right after birth will be placed on prostaglandins. After a few days, they may need surgery for placement of a shunt. A shunt is a small tube that connects the aorta to the pulmonary artery. It acts much like the PDA, and it provides extra blood flow to the lungs. Although it is not a final solution, it will give the baby time to grow and mature before more extensive surgery. The surgery for TOF is usually performed between 4-6 months. The surgeon removes the obstruction of blood flow to the lungs. The surgery is performed by cutting out the extra muscle tissue and enlarging the narrow areas with a transannular patch repair, so the pulmonary valve opens easier. The ventricular septal defect (VSD) is also closed with a patch.

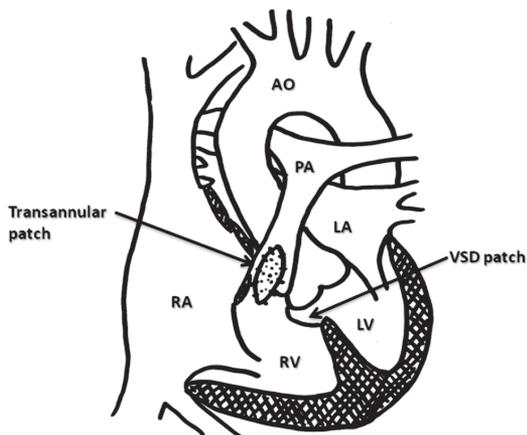
If your baby has pulmonary atresia, a severe obstruction of the pulmonary valve, the surgeon may not be able to remove the obstruction as described above. Instead, your baby will have a conduit inserted. A conduit is a tube which bypasses the pulmonary valve and connects the right ventricle to the pulmonary artery. This procedure is called a Rastelli repair. Your baby could be in the hospital for approximately 5-10 days after the surgery. The surgery will close the VSD and remove most of the obstruction of blood flow to the lungs.

A baby with a conduit will eventually develop obstructed blood flow due to physical growth while the conduit stays the same size. The conduit needs to be replaced a few times during childhood. When the surgeons remove the blood flow obstruction to the lungs, the pulmonary valve is altered. The pulmonary valve continues to open but does not seal when closed, which causes the valve to leak. Although this leakage does not cause immediate problems, eventually the right ventricle may enlarge and not pump blood as well. A baby with valve leakage will eventually require a replacement valve.

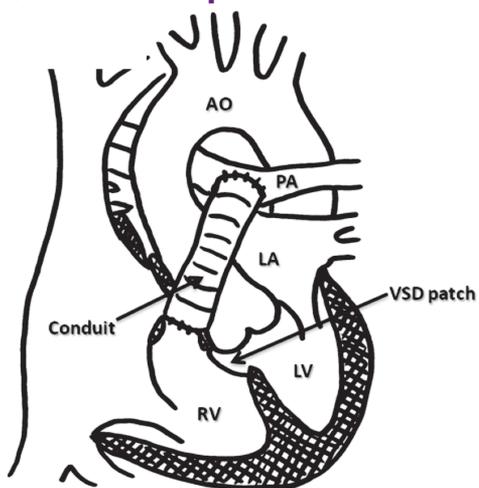
Before Surgery



Transannular Patch Repair



Rastelli Repair



Will my baby have a normal childhood?

Most children born with TOF are able to participate in normal activities, including sports. The long-term expectation for a child with TOF is good and the survival rate from surgery is high. Immediately after surgery, there will be frequent visits with your cardiologist. After the first year, the appointments will become less frequent, eventually occurring only once a year. At follow-up visits, echocardiograms, electrocardiograms (EKGs, which look at the electrical activity of the heart), and other types of monitoring will be done. As your child gets older, there may be exercise stress tests, 24-hour heart monitors, or cardiac MRIs. A patient with TOF may develop abnormal heart rhythms, which sometimes means medications or pacemakers. If there is blood flow obstruction to the lungs or significant valve leakage, your child may require surgery or a catheterization procedure. Your cardiologist will decide if your child should avoid any activities.

Will we have another child with a congenital heart defect?

Studies suggest that if you have one child with a congenital heart defect, your risk of having another child with a heart defect is about 2%-3%. If your baby's congenital heart defect is associated with a chromosome abnormality or a genetic syndrome, a genetic counselor can discuss with you and your family the risk of having another baby with the same condition. In any future pregnancies, we highly recommend nuchal translucency screening with an early fetal echocardiogram during your first trimester. Then, we recommend a targeted anatomy ultrasound between 18-20 weeks, and a fetal echocardiogram between 22-24 weeks.

How can the Fetal Heart Program help?

The Fetal Heart Program at the University of Maryland Medical Center is dedicated to the care and support of you and your unborn child. Our world class program aims to diagnose congenital heart defects as early, and as accurately as possible. We strive to create personalized prenatal care and optimize your delivery plan. Our multidisciplinary team is devoted to you and your baby's needs before and after birth.

It has been
our privilege
to care for you and your child.



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