



Transposition *of the* Great Arteries

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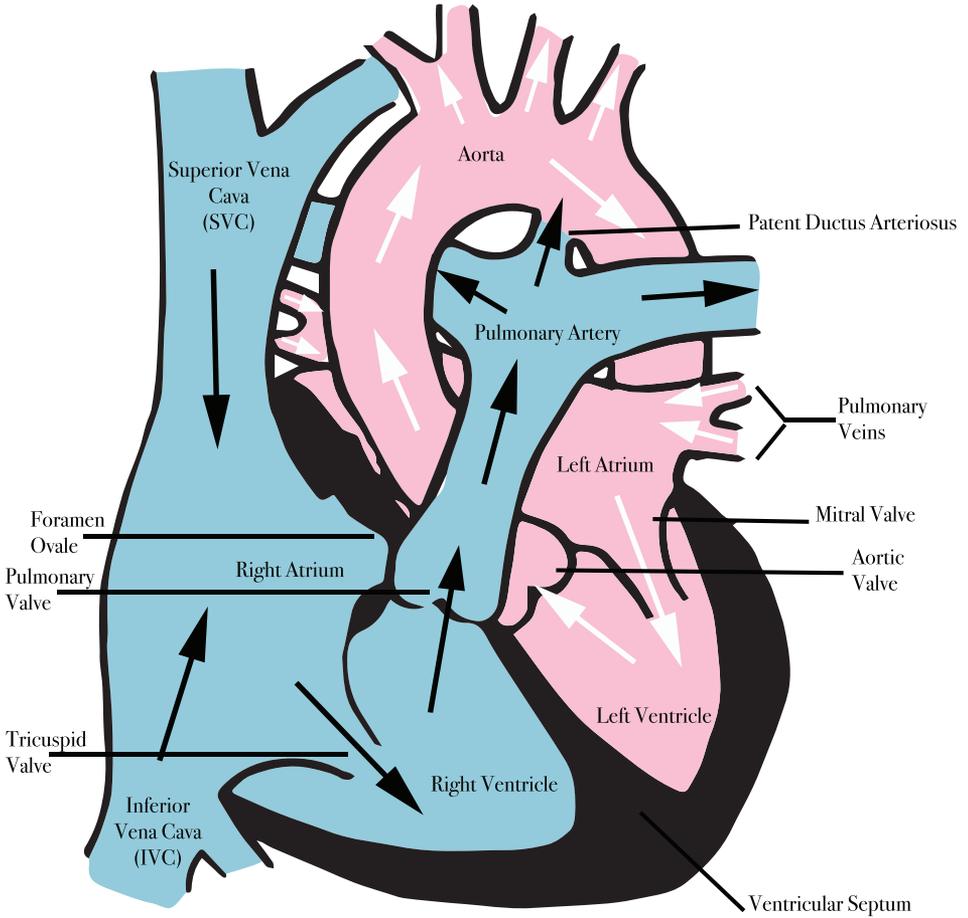
This guide will help you understand your child's heart. It is not a diagnosis and should never be used instead of medical advice.

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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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.

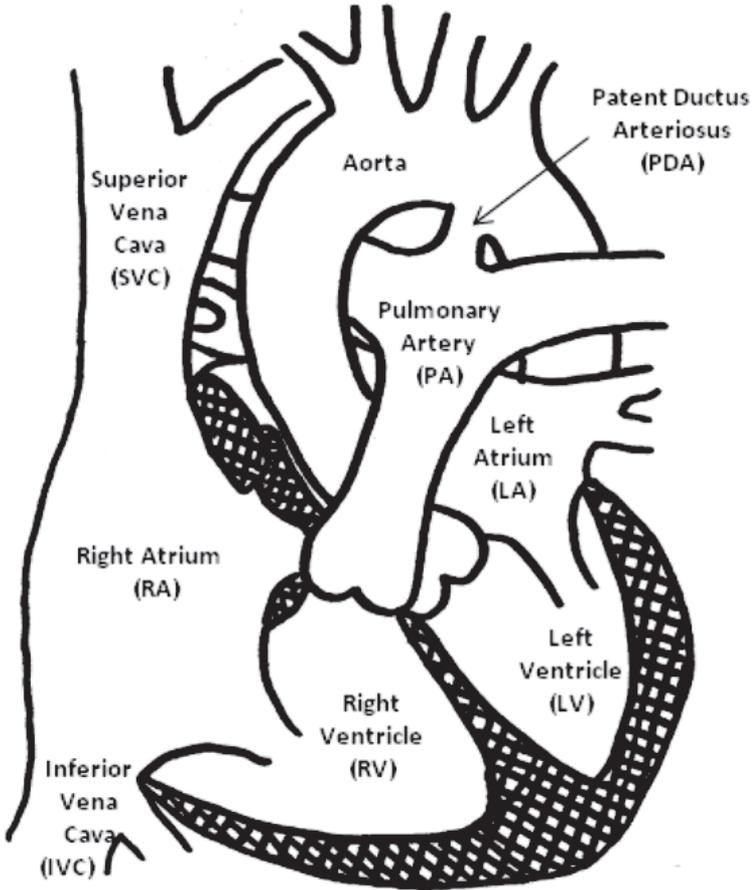
For more information, visit the fetal heart program online at: umm.edu/programs/fetalheart/patient-information/resources



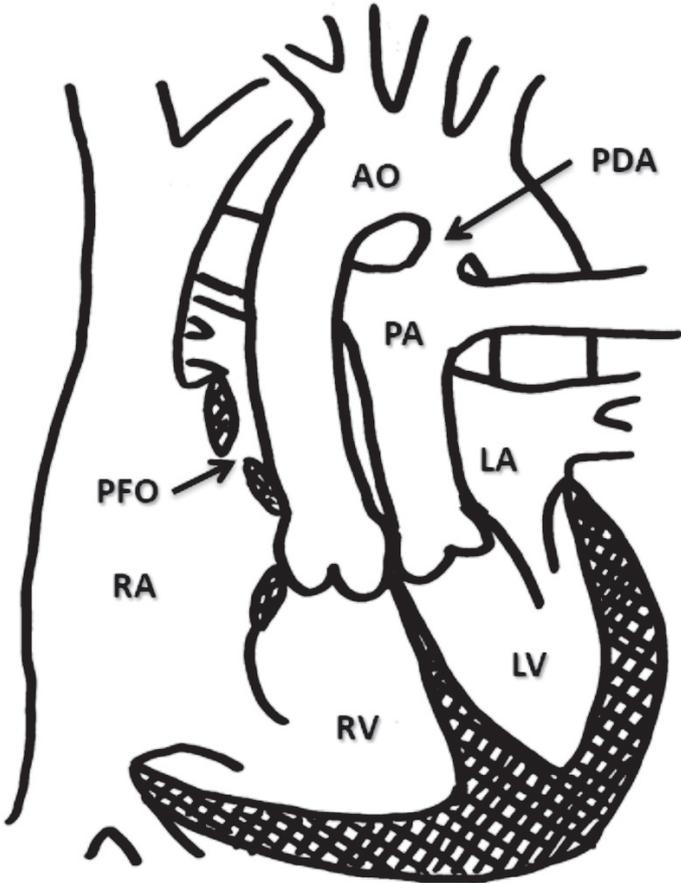
What is Transposition of the Great Arteries?

Transposition of the great arteries (TGA) is a congenital heart defect (CHD). TGA occurs when the main artery which carries blood to the lungs (pulmonary artery) and the main artery which carries blood to the rest of the body (aorta) are switched.

Normal Heart



Transposition of the Great Arteries



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

How is Transposition of the Great Arteries diagnosed?

TGA is diagnosed by an echocardiogram. A fetal echocardiogram (ultrasound of the fetal heart) is the first step in detecting if your baby demonstrates a possible congenital heart defect. A baby born with TGA may appear bluish in color. This is called cyanosis. A baby may have difficulty breathing. Postnatal evaluation for TGA includes an electrocardiogram (ECG), measuring the oxygen levels, and an echocardiogram. If further evaluation of the cardiac anatomy is needed, a cardiac MRI or cardiac catheterization may be performed.

What causes Transposition of the Great Arteries?

Like all congenital heart defects, the reason for TGA is often unknown. There is a small chance the defect may be associated with a genetic disorder. A baby with TGA and a chromosomal or genetic condition usually has physical and developmental problems, too. During pregnancy, either a chorionic villus sampling (CVS) or an amniocentesis can test for many chromosomal conditions. These procedures are associated with 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 newborn babies with TGA will be checked for genetic and chromosomal conditions.

What can I expect during my pregnancy?

If your baby is diagnosed before birth with TGA, 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 with TGA 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, with no intervention.

What can I expect after my baby is born?

After delivery, your baby will be taken to the neonatal intensive care unit (NICU). The specialists in the NICU will closely monitor your baby's vitals. An echocardiogram will be performed shortly after birth. The immediate treatment for a newborn with TGA is to find a way for the oxygenated and deoxygenated blood to mix. Once your baby is stable, surgery will be performed. Surgery typically takes place within the first week of life.

In TGA, the main arteries are switched, creating 2 parallel circuits. One pump is only circulating oxygenated blood to the lungs and the second pump is only circulating deoxygenated blood to the body. The vital organs of the body are deprived of oxygen rich blood. The parts of the heart that allow blood to mix prior to surgery are the patent ductus arteriosus and the patent foramen ovale.

The patent ductus arteriosus (PDA) is a connection between the aorta and pulmonary arteries. It normally closes in the first few days after birth. A medication, prostaglandins, is given through an IV to maintain the PDA. This creates a path for the deoxygenated blood to go from the aorta to the pulmonary arteries, where it can pass through the lungs and pick up oxygen.

The patent foramen ovale (PFO) is a hole between the two atria. When oxygenated blood from the lungs drains into the left atrium, the PFO lets blood enter the right atrium. This oxygenated blood is then pumped out to the rest of the body. After birth, the foramen ovale eventually closes. To ensure that it stays open, a balloon atrial septostomy procedure is performed. A catheter with a small balloon tip is guided into the heart and across the atrial septum through either a vessel in the groin or belly. The purpose is to create an opening in the atrial septum. This procedure may be performed either at bedside, with guidance from an echocardiogram, or in the catheterization laboratory.

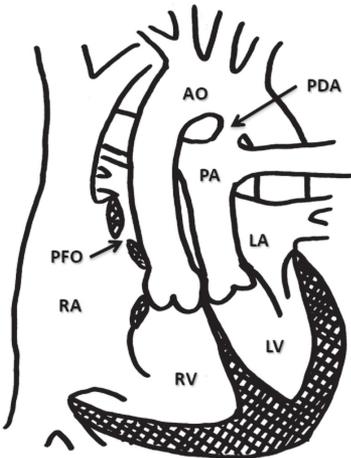
A ventricular septal defect (VSD) is seen in approximately 25% of patients with TGA. A VSD is a hole between the 2 ventricles. Depending on the size, the hole may allow oxygenated and deoxygenated blood to mix, similar to the patent ductus arteriosus and the patent foramen ovale.

How is Transposition of the Great Arteries treated?

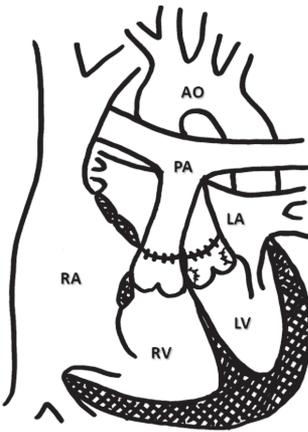
Surgery, called an arterial switch, is needed in the first couple weeks after birth to establish normal blood flow. The surgeon cuts the aorta and pulmonary artery above the valve and then carefully reconnects them to the proper ventricle. The aorta is attached to the left ventricle. The pulmonary artery is attached to the right ventricle. The next step in the arterial switch surgery is to properly attach the coronary arteries. The coronary arteries are small blood vessels that come from the aorta and carry blood to the heart. They must be removed from their original location and sewn back onto the aorta in its new position. After surgery, your baby will remain in the pediatric intensive care unit (PICU) for about 2 weeks. Your baby will be able to go home when he/she is eating well and has stable vital signs.

After the arterial switch surgery, narrowing in the aorta, pulmonary arteries, or where the coronary arteries are reattached to the aorta may develop at any stage in your child's life. If it is mild, this may be monitored with echocardiograms. Sometimes, these narrowed areas can be fixed by using a balloon or a stent in the cardiac catheterization laboratory. Other times, repeat surgery is needed. While the arterial switch surgery is not without risk, outcomes are typically positive, with a greater than 95% success rate.

Before Surgery



After Surgery



Normal



TGA



Repair of TGA



Will my baby have a normal childhood?

Most children with a successful arterial switch surgery can participate in normal activities, including sports. A person with TGA will be monitored by a cardiologist during their entire life. It is possible they will never require another procedure or surgery, but there are some complications that can occur. Follow ups will be frequent for a baby with TGA. An echocardiogram and electrocardiogram will be done at each follow up visit. Exercise stress tests and cardiac MRI's may also be done as your child grows older. Your cardiologist will decide if your child should avoid any specific types of 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.



UNIVERSITY *of* MARYLAND
MEDICAL CENTER

Center For Advanced Fetal Care
22 South Greene Street
Room N6E16
Baltimore, MD 21201
410-328-3865

Visit us online
umm.edu/programs/fetalheart