

TRANSPOSITION OF THE GREAT ARTERIES

What is transposition of the great arteries (TGA)?

TGA is a congenital heart defect in which the large blood vessels which carry blood from the heart to the lungs (pulmonary arteries) and body (aorta) are coming off the wrong pumping chambers (ventricles). In the normal situation, the pulmonary artery comes from the right ventricle and carries blue blood (less oxygen) to the lungs, and the aorta comes from the left ventricle and carries red blood (more oxygen) to the body. In TGA the aorta comes from the right ventricle and the pulmonary arteries come from the left ventricle.

What causes TGA?

There is no known cause of TGA. Some patients with TGA have genetic disorders. TGA does not run in families, but there is an increased chance of having a congenital heart defect if a relative also was born with a heart defect.

How is TGA diagnosed? What are signs and symptoms?

TGA is sometimes diagnosed by fetal ultrasound before the baby is born. First trimester screening for chromosomal abnormalities is a good screening tool to identify patients who might be at an increased risk for cardiac defects. If TGA is not detected before birth, babies may initially appear healthy, although their oxygen levels will be decreased. They may appear bluish in color, or have difficulty breathing or fast breathing. If TGA or another heart defect is suspected, the baby will be evaluated by a pediatric cardiologist. That evaluation would include measuring the oxygen level, an electrocardiogram (ECG) and an echocardiogram (ultrasound of the heart). The echocardiogram would show the abnormal structure of the heart.

How will your pregnancy be managed?

If your baby is diagnosed with a TGA, a high-risk obstetrician will participate in your obstetric care. Overall care should be transferred to a specialized center where multidisciplinary care is available. The team should include a perinatologist, fetal and pediatric cardiologists, a genetic counselor, a neonatologist and a pediatric cardiac surgeon. Fetal well being will be followed closely by fetal ultrasound and nonstress tests. Towards the end of pregnancy, visits may be as often as two to three times a week. If there is no specific maternal or fetal reason for a C-section, vaginal delivery is often possible. Induction of labor is often scheduled for pregnancies affected with TGA to make sure that all of the team members are available at the time of delivery.

Why does TGA make babies sick?

Since the arteries are hooked up wrong, the blue and red blood go to the wrong places. The blue blood returns from the body to the right side of the heart. The blue blood is then pumped from the right ventricle back to the body, without ever getting more oxygen. Meanwhile the red blood returns from the lungs to the left side of the heart. It gets pumped from the left ventricle back out to the lungs. The two circuits are separate, and the body is not able to get the oxygen it needs. These makes babies turn blue, due to low oxygen levels, and could eventually lead to babies becoming very sick.

What can I expect after my baby is born?

Babies with these types of heart defects will need evaluation and stabilization in the neonatal intensive care unit after birth. The diagnosis will be confirmed with an echocardiogram, and babies may get large IV's placed. Some babies need a breathing tube placed to help manage their condition. The immediate treatment for newborns with TGA is to find a way for the blood to mix, so that the blood getting to the body will have at least some oxygen in it.

There are two parts of the heart that can allow blood to mix and keep babies from getting sick. One is called a patent ductus arteriosus (PDA). It is a vessel that is found in all babies before birth and connects the aorta and the pulmonary arteries. It normally closes in the first few days after birth, but there is a medication that we can give to babies to keep it open. This allows blue blood to go from the aorta to the pulmonary arteries, where it can get to the lungs and pick up oxygen.

Another way for the blood to mix is needed to get that red blood to the body. A patent foramen ovale (PFO) is also seen in all babies before birth. It is a hole between the two top collecting chambers (atria). After birth it will allow red blood coming back from the lungs to cross from the left atrium into the right atrium. That red blood will then be pumped out to the body. Usually the PFO remains open in babies, but in some babies it may not be large enough to allow for enough red blood to cross. In that case, the baby may need a procedure to enlarge the PFO. This procedure is called a balloon atrial septostomy.

Some babies with TGA also have a hole between the two ventricles, called a ventricular septal defect (VSD). This may add another area to allow red and blue blood to cross sides, much like the PDA and PFO.

Babies with TGA will need surgery to correct the defect. Initially after birth, the baby will be started on prostaglandins to keep the PDA open. Within the first week after birth, surgery will be done.

Because the care for children with congenital heart disease is complex, your baby will be cared for by a team of skilled clinicians. This starts before birth with maternal-fetal medicine and fetal cardiologists and continues with pediatric cardiologists and nurse practitioners, a pediatric cardiac surgeon, neonatal and pediatric intensive care physicians, pediatric anesthesiologists, pediatric cardiac operating room staff, pediatric nurses and many others.

What is the treatment/surgery for TGA?

The surgery that is done for TGA is called an arterial switch. The surgeon cuts the aorta and pulmonary artery off of the heart. They are cut above the valves, so that a small part of the blood vessel and the valve remain attached to the heart. The aorta is then sewn onto the former pulmonary valve and left ventricle. The pulmonary artery is sewn onto the former aortic valve and right ventricle. The coronary arteries are small blood vessels that come from the aorta and carry blood to the muscle of the heart. They must be removed from their original location and sewn back onto the aorta in its new position. Once the surgery is complete, the arteries will be coming off of the correct ventricles and will carry blue blood to the lungs and red blood to the body.

Babies need to stay in the intensive care unit after surgery, and usually remain in the hospital for about 2 weeks. Once they are eating well, they are able to go home and often don't require medication.

What other procedures or follow up will my baby need?

People with TGA will need to be followed by a cardiologist for their whole life. It is possible that they will never require another procedure or surgery, but there are some complications that can occur. Visits will be frequent when they are babies, and will become every 6 to 12 months as they get older. An echocardiogram and electrocardiogram will be done at follow up visits. Exercise stress tests and cardiac MRI's may also be done as children get older.

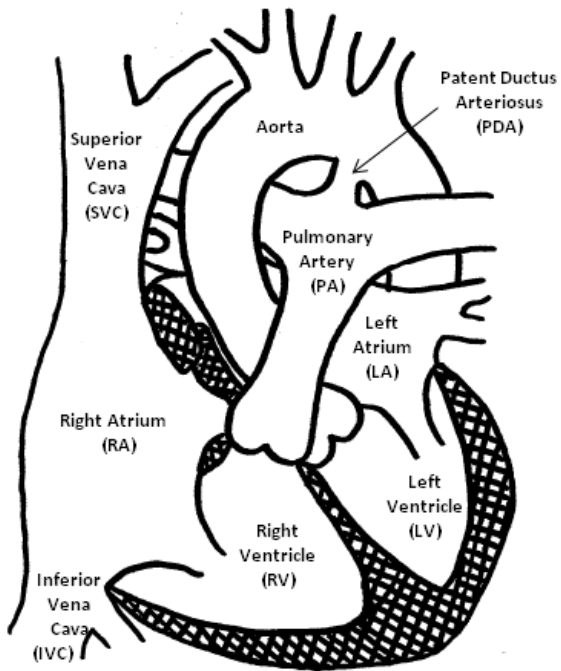
After the arterial switch surgery, some children develop narrowing in the aorta, pulmonary arteries, or where the coronary arteries are reattached to the aorta. If it is mild, this may be something to monitor with echocardiograms. Sometimes these narrowed areas can be fixed by using a balloon or a stent in the cardiac catheterization laboratory. Other times, repeat surgery may be needed.

What is the long term outcome for children with TGA?

The long term outcome for TGA is very good. Almost all babies survive the arterial switch surgery. Children are usually able to participate in activities, including sports. The arterial switch surgery was first performed about 30 years ago, so there are now adults who have TGA and had this type of repair.

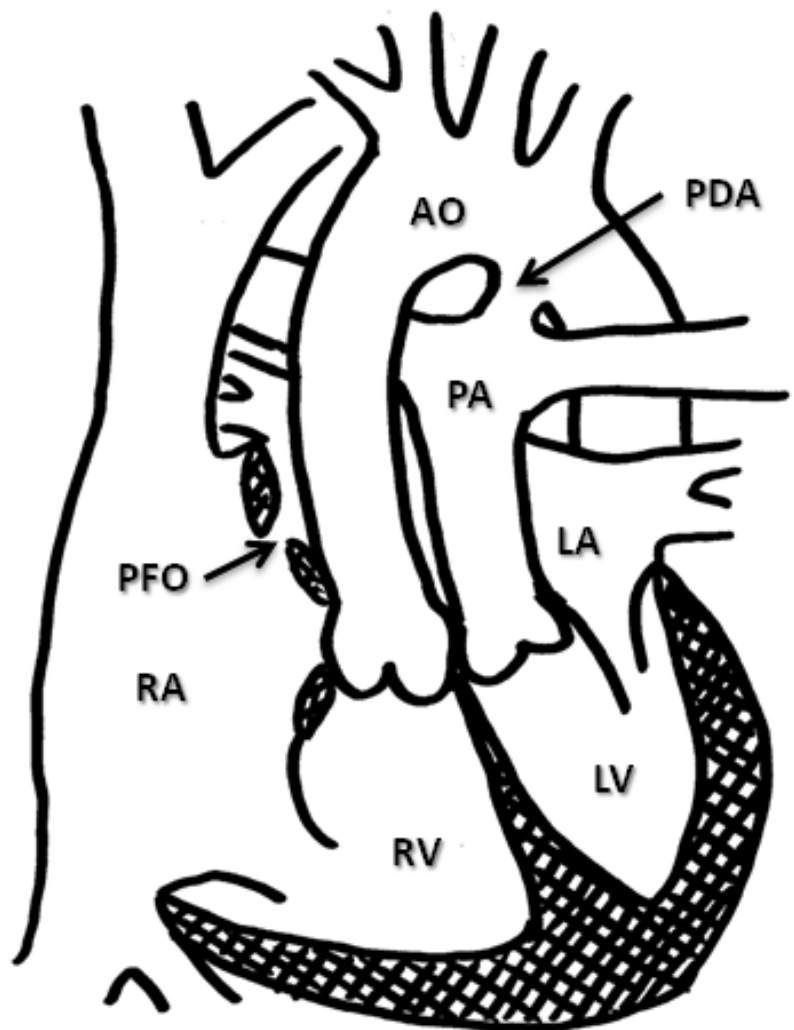
If I have had one baby with a TGA, am I more likely to have other babies with TGA?

If your baby's TGA is related to a chromosome abnormality or a genetic syndrome, a genetic counselor can tell you what the chances are that a future pregnancy would have the same condition. When TGA is not associated with an underlying genetic problem, future children are at a slightly increased risk for heart defects. In future pregnancies, nuchal translucency ultrasound (at the end of the first trimester), targeted anatomy ultrasound (between 18-20 weeks) and fetal echocardiography are recommended.

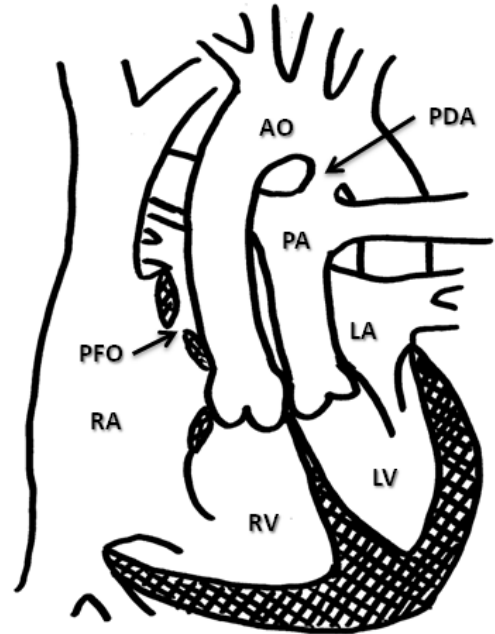


Normal Newborn Heart

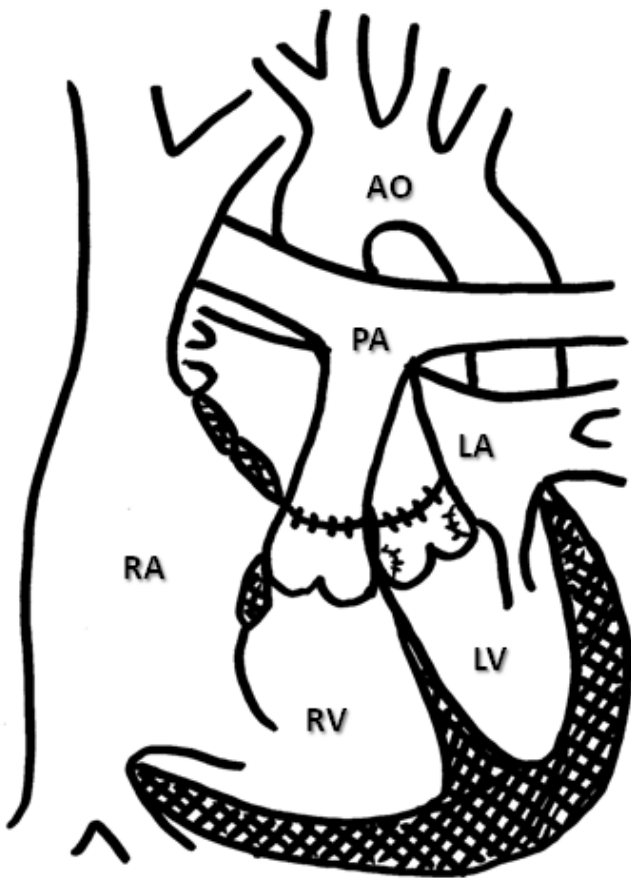
Transposition of the Great Arteries (TGA)



Before Surgery



After surgery



Normal



TGA



Repair of TGA

