

UNDERSTANDING TRANSPOSITION OF THE GREAT ARTERIES

CARMEN AND JOHN THAIN CENTER FOR PRENATAL PEDIATRICS

What is transposition of the great arteries?

In the normal heart, the right side of the heart pumps blood into the lungs where the blood can be filled with oxygen. Blood returns from the lungs into the left side of the heart and is then pumped into the body through the aorta.

In transposition of the great arteries (TGA) the outflow tracts - the major blood vessels coming out of the right and left sides of the heart - are reversed. Blood without oxygen is pumped by the right side of the heart into the body, and blood with oxygen is pumped by the left side of the heart to the lungs. (See the diagram for an illustration of this.)

Babies with TGA may not have enough oxygen going to the body, which is referred to as 'cyanotic'. There are various connections between the right and left sides of the heart in fetal life that may help a baby be less cyanotic at birth. Approximately 1/3 to 1/2 of babies with TGA also have a ventricular septal defect. Although this defect will need to be repaired, it will help the baby with oxygenation after birth

How common is TGA and what causes it?

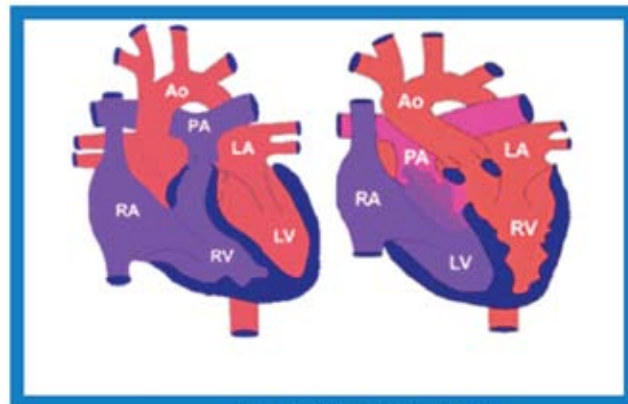
Transposition of the great arteries is seen in 2-4 out of every 10,000 live births. It represents about 4 to 6% of all cases of heart defects present at birth. It occurs equally in boys and girls.

No one knows for sure what causes TGA, but it likely happens very early in pregnancy when the heart is developing, around 4 to 5 weeks gestation.

TGA is slightly more common in babies born to mothers with diabetes mellitus, however, most cases are due to chance alone.

How is TGA detected during pregnancy?

TGA is usually detected during pregnancy through the sonogram performed to check fetal development in the second trimester. The diagnosis is made by identifying the aorta coming out of the right ventricle, and identifying the pulmonary artery coming out of the left ventricle. (Normally the pulmonary artery should come out of the right ventricle, and the aorta should come out of the left ventricle.) A ventricular



The Royal Children's Hospital Melbourne
(r) Illustration of a normal heart, (l) "Corrected" Transposition (with VSD)

septal defect may also be seen on this sonogram, although these may be small and difficult to see. The diagnosis of TGA may also be made or confirmed with a fetal echocardiogram. This is a specialized sonogram of the fetal heart. At NewYork-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center, all fetal echocardiograms for TGA are performed by Pediatric Cardiologists with expertise in this technique. Other cardiac defects can sometimes be seen in addition to TGA. This sonogram will assess for any additional cardiac abnormalities.

Of cases of TGA diagnosed prenatally, essentially all are confirmed after birth.

How will my pregnancy be managed now that TGA has been diagnosed?

An ultrasound will carefully examine the fetal development and assess for any additional birth defects. Although TGA is only rarely associated with additional abnormalities, it is important to investigate for other abnormalities that may affect the baby's prognosis. Your prenatal care will be managed by a Maternal-Fetal Medicine specialist, an obstetrician with special training and expertise in high-risk pregnancies. A fetal echocardiogram will be performed by a Pediatric Cardiologist to confirm the diagnosis and exclude other heart problems. Over the course of your pregnancy you will have additional ultrasounds to make sure that the fetus is growing well.

As your delivery date approaches, induction of labor or a cesarean section may be planned to optimize the delivery and ensure that the medical specialists are available to care for the baby immediately after birth. However, this may not be necessary in all cases, and labor management does not need to be changed because your baby has TGA.

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How will the TGA be treated after birth?

All patients with TGA require surgery. Typically surgery is performed within the first week of life.

The **arterial switch** is most commonly performed to repair transposition. In this surgery the great vessels (the main pulmonary artery and aorta) are disconnected and reconnected to the correct ventricles (main pulmonary artery to the right ventricle, and aorta to the left ventricle). The coronary arteries, the blood vessels supplying the heart, must also be transplanted to the left side of the heart.

A **Rastelli operation** is performed when transposition of the great arteries is associated with a ventricular septal defect and subvalvular pulmonary stenosis. The arterial switch cannot be performed for infants with these defects, since it would result in aortic stenosis after surgery. In the Rastelli operation, an intraventricular baffle is used to close the septal defect and direct the pulmonary venous blood to the aorta. In addition, a valved homograft conduit is placed to connect the right ventricle to the pulmonary artery.

What is the long-term outlook for babies with TGA?

The operative mortality rate for the arterial switch procedure is reported to be between 3 and 5%. Subsequent reoperation of catheterization procedures may be required in up to 10% of patients. Importantly, long term survival for patients with corrected TGA appears to be excellent. Outcomes do seem to be much better after arterial switch than for the Rastelli operation.

There may be some late complications of the arterial switch procedure. Patients with a corrected TGA should continue to be followed by a cardiologist throughout life. The most common late complications are right ventricular dysfunction and tricuspid regurgitation. Other late complications may include: supralvalvular pulmonary stenosis, neo-aortic root enlargement, aortic regurgitation, left ventricular dysfunction, branch pulmonary artery stenosis, aortic anastomotic stenosis, and coronary artery stenosis or distortion.

What are the chances I could have another baby with TGA?

The chance for a second pregnancy affected by TGA is very low. When one sibling is affected that risk is approximately 1.5%. Even when two siblings are affected the risk is only 5% for another pregnancy.

TGA is very rarely associated with chromosomal abnormalities or genetic syndromes. Speak to your genetic counselor about the options for further testing.

What can I expect from the specialists at Morgan Stanley Children's Hospital?

The specialists at Morgan Stanley Children's Hospital are among the most experienced with TGA in the United States. The Center approach includes a whole team in your prenatal care to optimize the chances for your baby's well being after birth. You might expect to meet the following specialists:

- **Maternal-Fetal Medicine (MFM)** — you will see one of a team of MFMs, specialized obstetricians, throughout your pregnancy.
- **Pediatric Cardiology** — these doctors perform fetal echocardiograms in the prenatal setting to diagnose TGA. The diagnosis will be confirmed after birth, and these physicians will continue to follow children with corrected TGA.
- **Pediatric Cardiothoracic Surgery** — these doctors perform the surgery to correct TGA. A recently released report by the New York State Department of Health has shown that NewYork-Presbyterian Hospital is a leader in the state for best outcomes of surgeries performed for children with congenital heart defects.
- **Genetics** — the genetic counselors will discuss the diagnosis of TGA with you in more detail as well as recurrence risk and options for additional testing.
- **Neonatology** — our top-ranked neonatology unit regularly cares for children with congenital heart defects before and after surgery.

About the Carmen and John Thain Center for Prenatal Pediatrics

Complex pregnancies receive better care when specialists collaborate. The Carmen and John Thain Center for Prenatal Pediatrics is dedicated to helping pregnant women and their families when a birth defect or genetic syndrome is detected before the baby is born. The Center offers sensitive, complete, up-to-date information and testing, and an integrated approach to care that begins in the prenatal period and continues after birth with pediatric follow-up. A collaborative, coordinated program of care is created among specialists in perinatology, neonatology, genetics, pediatric cardiology, pediatric surgery and all pediatric subspecialties.