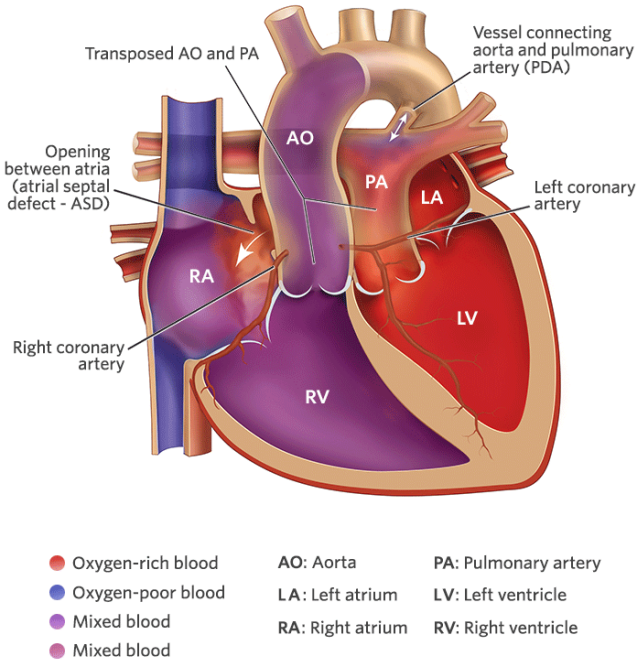
Cardiac Defects: Transposition of the Great Arteries



Transposition of the great arteries (TGA) is a complex congenital heart defect in which the two large arteries that carry blood out of the heart are connected to the heart abnormally:

- The aorta is attached to the right-sided pumping chamber (ventricle), instead of the left.
- The pulmonary artery is attached to the left-sided pumping chamber (ventricle), instead of the right.

Normally, blood flows in this pattern: body to right side of heart, to lungs, to left side of heart, and back to body. The pulmonary artery carries blood from the right side of the heart to the lungs, and the aorta carries blood from the left side of the heart to the body. In children with TGA, the normal pattern of flow does not exist, and the body doesn't get enough oxygenated blood.

What are the symptoms of TGA?

The symptoms of TGA include

- blue or purple tint to lips, skin, and nails (cyanosis)
- rapid breathing
- difficulty feeding, poor appetite, and poor weight gain.

How is TGA diagnosed?

TGA may be diagnosed before birth with a fetal echocardiogram (ultrasound). Your baby's provider will prepare a plan for delivery and care immediately after birth.

TGA may be diagnosed when an infant is a few hours or days old, or in some cases, infants may not have visible symptoms for weeks or months. Pediatricians refer newborns to a pediatric cardiologist when they notice symptoms or abnormal values on screenings such as pulse oximetry.

Diagnosis of TGA may require some or all of these tests:

- echocardiogram (also called echo or ultrasound)—sound waves create an image of the heart
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- chest X ray
- pulse oximetry—a noninvasive way to monitor the oxygen content of the blood
- cardiac catheterization—a thin tube is inserted into the heart through a vein or artery in either the leg or through the umbilicus ("belly button")
- cardiac magnetic resonance imaging—a three-dimensional image shows the heart's abnormalities.

What are the treatment options for TGA?

TGA is unpredictable. Approximately one-third of newborns with the condition will require an urgent intervention called a *balloon atrial septostomy* (BAS) within hours after birth. This procedure creates or enlarges a hole between the upper chambers of the heart to allow blood to mix.

For babies requiring a BAS procedure, access to immediate expert care is essential.

All children with TGA will require open-heart surgery to treat the defect. Without surgical repair, the overwhelming majority of patients with TGA will not survive their first year. The surgery, known as the *arterial switch operation*, is typically performed within a few days of birth. Pediatric cardiac surgeons reconstruct the heart so that the aorta is attached to the left ventricle and the pulmonary artery is attached to the right ventricle.

After surgery your child will recover in a neonatal or pediatric intensive care unit.

What is the follow-up care for TGA?

Through Age 18

Children who have had surgical repair of TGA require lifelong care by a cardiologist. Ongoing medication use is uncommon. More surgery may be required as the child grows.

Pediatric cardiologists follow patients until they are young adults, coordinating care with the primary care physicians.

Into Adulthood

The pediatric cardiologist will help your child transition to an adult cardiologist.

Until approximately 25 years ago, infants with TGA were managed by alternative surgical procedures, sometimes referred to as the *Senning* or *Mustard* operations. As a result, the long-term effects of the arterial switch operation beyond young adulthood are not known. Although some studies have shown that a proportion of patients display a slight cognitive and psychosocial disparity, there still is a need for more prospective research on the long-term health of these patients (Heinrichs et al., 2014; Kalfa et al., 2017). It is anticipated that the overwhelming majority of children born with TGA will go on to lead healthy, productive lives. Limitations to day-to-day activities, including sports, are rare.

References

Heinrichs, A. K., Holschen, A., Krings, T., Messmer, B. J., Schnitker, R., Minkenberg, R., & Hovels-Gurich, H. H. (2014). Neurologic and psycho-intellectual outcome related to structural brain imaging in adolescents and young adults after neonatal arterial switch operation for transposition of the great arteries. *The Journal of Thoracic Cardiovascular Surgery, 148*(5), 2190–2199.

Kalfa, D., Kasmi, L., Geronikola, N., Calderon, J., Lambert, V., Belli, E., . . . Montreuil, M. (2017). Cognitive outcomes and health-related quality of life in adults two decades after the arterial switch operation for transposition of the great arteries. *The Journal of Thoracic Cardiovascular Surgery, 154*(3), 1028–1035.