



Cardiac Defects: Truncus Arteriosus

When the fetus develops during pregnancy, the heart has a single large blood vessel coming from the heart called the *truncus arteriosus*. If fetal development progresses normally, the truncus divides into two arteries that carry blood out of the heart:

- The pulmonary artery, which is attached to the right bottom chamber (ventricle) of the heart, divides into two arteries carrying oxygen-poor (“blue”) blood to each side of the lungs.
- The aorta, which is attached to the left bottom chamber (ventricle) of the heart, carries oxygen-rich (“red”) blood to the body.

Sometimes the single large blood vessel fails to divide during fetal development, and the baby is born with a heart that has one artery carrying blood out of it. This condition is known as *truncus arteriosus* or *persistent truncus arteriosus* (the trunk “persists”).

The undivided trunk is attached to the heart as one artery straddling the bottom chambers and then divides into arteries taking blood to the lungs and body. The oxygen-poor blood from the right ventricle (bottom chamber) and the oxygen-rich blood from the left ventricle (bottom chamber) mix when ejected out into the trunk, and more blood than normal goes back to the lungs, making it harder for the infant to breathe.

In almost all cases, children with the congenital heart defect truncus arteriosus also have a large hole between the bottom chambers of the heart. This is called a *ventricular septal defect* (VSD).

As a result of these abnormalities, the baby’s blood isn’t as oxygenated as it should be when it circulates through the body.

Signs and Symptoms

Signs and symptoms of truncus arteriosus include

- blue or purple tint to lips, skin, and nails (cyanosis)
- poor eating and poor weight gain
- rapid breathing or shortness of breath

- profuse sweating, especially with feeding
- more sleepiness than normal
- unresponsiveness (the baby seems “out of it”)
- heart murmur—the heart sounds abnormal when a doctor listens with a stethoscope.

How is truncus arteriosus diagnosed?

Truncus arteriosus is a life-threatening congenital heart defect; most babies won’t live for more than a few months without treatment.

Usually truncus arteriosus is diagnosed before the baby leaves the hospital if the doctor hears a murmur or sees a blue tint to the lips or skin. In some cases, a primary care pediatrician might detect the symptoms of truncus arteriosus during a check-up, or a parent might notice symptoms and bring the baby to a doctor or hospital.

Diagnosis of truncus arteriosus may require some or all of these tests:

- pulse oximetry—a painless way to monitor the oxygen content of the blood
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- echocardiogram (also called “echo” or ultrasound)—sound waves create an image of the heart
- chest X ray
- cardiac MRI—a three-dimensional image shows the heart’s abnormalities
- 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”).

Sometimes truncus arteriosus is diagnosed on a fetal ultrasound or echocardiogram. Your baby’s providers can prepare a plan for delivery and care immediately after birth.

A number of children with truncus arteriosus also have a genetic syndrome called 22q11 deletion syndrome (also known as *DiGeorge*, *velocardiofacial*, or *conotruncal*



anomaly face syndromes). Genetic testing (a blood test) for this syndrome may be part of the evaluation.

Truncus Arteriosus Treatment

Open-heart surgery is required to treat truncus arteriosus, usually before the baby is 2 months old. More than one operation may be required.

Cardiothoracic surgeons place a patch to close the hole (the ventricular septal defect). They separate the pulmonary arteries from the trunk and then connect the pulmonary arteries to the right bottom chamber (ventricle) of the heart using different kinds of conduits (tubes). They repair the trunk so that it becomes a complete, functioning aorta. Other repairs may be needed, based on each child's unique needs.

What is the follow-up care for truncus arteriosus?

Through Age 18

A child who has had surgical repair of truncus arteriosus will require life-long care by a cardiologist.

Pediatric cardiologists follow patients with truncus arteriosus until they are young adults, coordinating care with the primary care provider. Patients will need to carefully follow providers' advice, including staying on any medications prescribed and, in some cases, limiting certain types of exercise.

Sometimes children with truncus arteriosus experience heart problems later in life, including irregular heartbeat (arrhythmia), a restricted conduit or pulmonary artery, or a leaky aortic valve. Medicine, surgery, or cardiac catheterization may be required.

Into Adulthood

Your baby's pediatric cardiologist will help patients who have had truncus arteriosus treatment transition to an adult cardiologist.

Because of enormous strides in medicine and technology, today most children with heart defects go on to lead productive lives as adults.

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