



Cardiac Defects: Tricuspid Atresia

The right side of the normal heart receives oxygen-poor blood (“blue” blood) from the body’s veins and pumps it to the lungs to receive oxygen. The oxygen-rich blood (“red” blood) returns from the lungs to the left side of the heart, which pumps the blood to the body. The tricuspid valve is the opening between the right atrium (the upper chamber) and the right ventricle (the lower chamber). A heart with tricuspid atresia is characterized by poorly developed right heart structures and

- no tricuspid valve
- a smaller-than-normal, or hypoplastic, right ventricle
- a hole between the right atrium and left atrium, so that oxygen-poor and oxygen-rich blood mix inside of the heart
- a hole between the right ventricle and left ventricle.

Tricuspid atresia in children often is associated with pulmonary stenosis or narrowing of the pulmonary valve, or pulmonary atresia, where the pulmonary valve is completely closed. Tricuspid atresia also can be associated with transposition of the great arteries, where the aorta, the large artery that carries blood to the body, is connected to the small right ventricle.

Tricuspid atresia is a single-ventricle lesion because the heart has only one functioning ventricle (the left ventricle).

Symptoms

Tricuspid atresia symptoms in children include

- blue or purple tint to lips, skin, and nails (cyanosis)
- heart murmur—the heart sounds abnormal when a doctor listens with a stethoscope
- shortness of breath
- difficulty feeding
- poor weight gain
- fatigue
- abnormal shape of the fingertips (“clubbing”) in older children.

How is tricuspid atresia diagnosed?

Tricuspid atresia in children may be diagnosed before birth with a fetal echocardiogram. Your baby’s providers

can prepare a plan for delivery and care immediately after birth.

Tricuspid atresia is usually diagnosed a few hours or days after birth. Pediatricians refer newborns to pediatric cardiologists when they notice symptoms and signs such as a “blue baby with a heart murmur.” Pulse oximetry is a painless way to monitor the oxygen level of the blood.

Some or all of these tests may be required for diagnosis of tricuspid atresia in children:

- chest X ray
- blood tests
- electrocardiogram (EKG)—this test shows the electrical activity of the heart
- echocardiogram (also called echo or ultrasound)—sound waves create an image of the heart. This test usually confirms the diagnosis.
- 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”).

What are the treatment options for tricuspid atresia?

Your baby will be admitted to the cardiac intensive care unit. The baby may require oxygen and a medication called prostaglandin to maintain adequate oxygen levels in the blood. *Prostaglandin* is an intravenous medication that keeps open the connection between the pulmonary artery (the artery that normally carries blue blood to the lungs to receive oxygen) and the aorta (the artery that carries red blood to the body). This connection, called patent ductus arteriosus (PDA), is open in the fetus and closes soon after birth. When the PDA closes, some babies with tricuspid atresia turn quite blue (cyanosed). An infusion of prostaglandin can reopen the PDA and is a lifesaving intervention. Not all babies with tricuspid atresia require prostaglandin.

If the baby has labored breathing or poor effort, he or she may need help with a breathing machine or ventilator.



It is not uncommon for babies to have poor respiratory effort or apnea while on prostaglandin infusion.

At least two and possibly three surgeries will be required:

Blalock-Taussig Shunt

Babies who require prostaglandin to maintain adequate oxygen levels will require surgery soon after birth. The surgery involves the creation of a shunt, which is a tube that connects one of the branches of the aorta and the pulmonary artery, and thus replaces the PDA. This operation is called the *Blalock-Taussig* (BT) shunt. Many babies with tricuspid atresia are well enough to be discharged home soon after birth. However, some of these babies may require the shunt operation at a few weeks of life if the level of oxygen in their blood is decreasing.

Some babies with tricuspid atresia are too “pink” or have too much blood flow to the lungs and will require an operation called *pulmonary artery banding* to narrow the pulmonary artery and regulate blood flow to the lungs. Babies with tricuspid atresia and transposition of great arteries may require the Norwood procedure if the aorta is too small (see *Hypoplastic Left Heart Syndrome* in Diagnoses).

Hemi-Fontan/Glenn

The second operation, called the *hemi-Fontan/Glenn*, usually occurs within 6 months of birth. During this surgery, the superior vena cava, one of the two large veins attached to the heart to return deoxygenated or blue blood from the upper half of the body, is disconnected from the heart and attached to the pulmonary artery. During this operation, the surgeon also removes the BT shunt. After this operation, deoxygenated or blue blood from the upper body goes to the lungs without passing through the heart.

Fontan

The third operation, called the Fontan, occurs at approximately 18 months to 3 years of age. During this surgery, the inferior vena cava, the other large vein that returns deoxygenated blood to the heart from the lower half of the body, is disconnected from the heart and attached to the pulmonary artery. This means that deoxygenated or

blue blood from the whole body goes to the lungs without passing through the heart.

The cardiac team will explain the surgical procedures to you in more detail, based on your child’s heart anatomy.

What is the follow-up care for tricuspid atresia?

Between the Norwood and Glenn Procedures

Although early outcomes for patients with single ventricle heart defects after staged reconstruction have improved dramatically, the period between the Norwood and Glenn procedures remains a very vulnerable time for infants.

Through Age 18

Children with tricuspid atresia require lifelong care by a cardiologist. Many remain on medications for life. Additional surgeries may be required.

As single ventricle survivors get older, doctors are recognizing that, although some do fine, many experience complications, including lung, liver, and gastrointestinal diseases.

In addition, as a group, children with complex congenital heart defects who have had open-heart surgery as infants are at a higher risk for neurodevelopmental issues compared with children without congenital heart defects. Pediatric cardiologists follow patients until they are young adults, coordinating care with the primary care physicians.

Into Adulthood

It’s important that your child continues to see a cardiologist as an adult. The pediatric cardiologist will help with the transition to an adult cardiologist. Because of enormous strides in medicine and technology, today many children born with tricuspid atresia go on to lead healthy, productive lives as adults.

Adapted with permission. © The Children’s Hospital of Philadelphia.