Tricuspid Atresia

What is it?
In this condition, there’s no tricuspid valve so blood can’t flow from the right atrium to the right ventricle. As a result, the right ventricle is small and not fully developed. The child’s survival depends on there being an opening in the wall between the atria (atrial septal defect) and usually an opening in the wall between the two ventricles (ventricular septal defect). As a result, the low-oxygen (bluish) blood that returns from the body veins to the right atrium flows through the atrial septal defect and into the left atrium. There it mixes with oxygen-rich (red) blood from the lungs. Most of this partially oxygenated blood goes from the left ventricle into the aorta and on to the body. A smaller-than-normal amount flows through the ventricular septal defect into the small right ventricle, through the pulmonary artery, and back to the lungs. Because of this abnormal circulation, the child looks blue (cyanotic).

What can be done to treat it?
Often it’s necessary to do a surgical procedure, called a shunt, to increase blood flow to the lungs. This improves the cyanosis. Some children with tricuspid atresia have too much blood flowing to the lungs. They may need a different type of surgery, called pulmonary artery banding, to decrease blood flow to the lungs. This is important to protect the lung blood vessels.
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Can it be repaired?
Most children with tricuspid atresia can have surgery to allow their hearts to work more like normal. Connections are created between the body veins and the lung (pulmonary) arteries. This is usually done in two stages. First, the large vein from the upper half of the body (the superior vena cava) is connected to the lung arteries in a procedure called a Bidirectional Glenn Operation.

Later, the large vein from the lower half of the body (the inferior vena cava,) as well as the veins from the liver, are connected to the lung arteries in a surgery called a Fontan Operation. Sometimes, at the time of the Fontan surgery, an opening is purposely left between the bluish (low-oxygen) and red (high-oxygen) sides of the blood flows.

The Fontan operation may eliminate or greatly improve the cyanosis but, without a right ventricle that works normally, the heart doesn’t work like a normal heart, which has two pumps. The Fontan procedure can be performed using a tube that goes around the heart as shown in the picture or with a path (baffle) that goes inside the heart. Both types of Fontan operations route the blue blood from the lower half of the body and liver to the lungs.

What will my child need in the future?
Children with tricuspid atresia require lifelong follow-up by a cardiologist for repeated checks of how their heart is working.
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What about preventing endocarditis?
Children with tricuspid atresia are at increased risk for developing endocarditis. Ask your pediatric cardiologist about your child’s need to take antibiotics before certain dental procedures to help prevent endocarditis.