Transposition of the Great Arteries

What is it?
In transposition of the great arteries, the aorta and pulmonary artery are reversed. The aorta receives the oxygen-poor blood from the right ventricle, but it’s carried back to the body without receiving more oxygen. Likewise, the pulmonary artery receives the oxygen-rich blood from the left ventricle but carries it back to the lungs.

What about surgical treatment?
Patients with transposition of the great arteries require surgery early in life to survive. Many infants undergo a procedure in the catheterization laboratory to “buy time” and delay the surgery until they can handle it better. The procedure enlarges a naturally occurring connection between the right and left upper chambers (the atria). This lets the blood mix so some oxygen-rich and oxygen-poor blood can be pumped to the correct side.

Two major types of surgery can correct the transposition. The first creates a tunnel (a baffle) between the atria. This redirects the oxygen-rich blood to the right ventricle and aorta and the oxygen-poor blood to the left ventricle and the pulmonary artery. This operation is called an atrial or venous switch. It’s also called the Mustard procedure or the Senning procedure.
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The second type is called the arterial switch operation. The aorta and pulmonary artery are switched back to their normal positions. The aorta is connected to the left ventricle, and the pulmonary artery is connected to the right ventricle. The coronary arteries, which carry the oxygen-rich blood that nourishes the heart muscle, also need to be re-attached to the new aorta.

What type of problems might my child have?

Heart function problems
Patients who’ve had an atrial switch (e.g., Mustard or Senning operation) may have a serious decline in heart muscle or heart valve function. This is because the right ventricle is pumping blood to the entire body instead of just the lungs. Medications to help the heart pump better, control fluid accumulation (diuretics) and control blood pressure may help. Patients who’ve had the arterial switch operation don’t seem to have as great a risk of heart muscle decline. They may have valve leakage or coronary artery problems, however.

Heart rhythm problems (Arrhythmias)
People with repaired transposition, especially those who’ve had the Mustard or Senning operation, are at risk of developing heart rhythm abnormalities (arrhythmias). These arrhythmias often arise in the heart’s upper chambers. Your child’s heart rate may be too slow or too fast. If the heart rate is too slow, an artificial pacemaker can speed it up. If your child’s heart rate is too fast, medication can slow it down. At times, your child may need a cardiac catheterization to study and treat these rhythm disturbances.

Will my child need more surgery?
Some patients need more surgery to help their heart pump better, repair abnormal valves or control heart rhythm disturbances. Patients who’ve had the Mustard or Senning operation may need surgery to correct abnormalities of the tunnel in the atria, repair abnormal valves or control rhythm disturbances.

Patients who had the arterial switch operation may need more surgery to relieve narrowings in the aorta or pulmonary artery where the original surgery was done, or to fix leaky valves.

Will my child’s activities be limited?
Most cardiologists recommend that patients limit their physical activities to their endurance. They don’t recommend competitive sports for high school and college students. Your child’s cardiologist will help determine the proper level of activity restriction.
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What will my child need in the future?
Patients with transposition will require lifelong follow-up with a cardiologist trained to care for patients with congenital heart disease. Your child may need to take medications to improve how his or her heart works. The cardiologist will track your child with a variety of non-invasive tests. These include electrocardiograms, Holter monitors, exercise stress tests and echocardiograms.

What about preventing endocarditis?
Children who have transposition of the great arteries are at increased risk for endocarditis. Some children will need to take antibiotics before certain dental procedures.