I-Transposition of the Great Arteries (Congenitally Corrected
Transposition of the Great Arteries)

What is it?
I-transposition of the great arteries (also known as levo-transposition or “Congenitally Corrected” transposition of the great arteries) is the less common type of transposition. The right and left ventricles are reversed (ventricular inversion). The aorta and pulmonary artery are also connected to the wrong ventricles. Unlike in d-TGA, the aorta receives the oxygen-rich blood from the right ventricle, and oxygen-poor blood is carried back from the body. Likewise, the pulmonary artery receives the oxygen-poor blood from the left ventricle, which pumps it to the lungs. Because the blood flows normally despite the inverted ventricles, this lesion is also called “congenitally corrected TGA.” Some children may also have ventricular septal defects, obstruction to flow into the pulmonary artery, or leakage of the valve tricuspid valve.

What causes it?
The cause is unknown, but genetic factors may contribute to it.

How does it affect the heart?
In this condition, the blood is normally routed but the right ventricle must pump at higher pressure than is normal. The right ventricular function may decline over time.

How does it affect my child?
Babies born with I-transposition usually aren’t blue. The congenital heart defect may go undetected for a long time. It might not be diagnosed until well into adulthood when congestive heart failure, heart murmurs and abnormal heart rhythms can develop. When there is a ventricular septal defect and pulmonary valve obstruction, the baby may be blue and murmurs are usually heard.

Is surgery needed?
Most children without a VSD or pulmonary valve obstruction won’t need surgery. Children with these problems may require surgery to close the hole, relieve the blockage of blood flow to the lungs, and in some cases repair or replace the leaky tricuspid valve. In patients with VSDs, it may be recommended
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to undergo a more complex surgical procedure known as a “double switch” which reroutes the red blood to the left ventricle and aorta and blue blood to the right ventricle and pulmonary artery.

Problems your child may have may have
Patients may have problems with heart muscle, tricuspid valve leakage or heart rhythm as they enter adulthood, whether or not they had prior surgery. Like patients with atrial repair of d-TGA, the right ventricle may weaken, leading to congestive heart failure. Abnormally slow heart rhythms may cause fainting or fatigue. The most worrisome slow heart rhythm, complete heart block, is common in l-transposition. Rapid heart rhythms are less common than in d-TGA patients.

Ongoing Care:

What will my child need in the future?
Medications to help the heart pump better, control fluid accumulation (diuretics) and control blood pressure may help with congestive heart failure symptoms. A pacemaker may be required in the event of abnormally slow heart rhythms. You should also consult a cardiologist with expertise in caring for adults with congenital heart disease if you are undergoing any type of non-heart surgery or invasive procedure.

Medical Follow up
Routine evaluation may include an EKG, echocardiogram, Holter monitor and stress testing. Your cardiologist may recommend other testing such as an MRI or heart catheterization.

Activity Restrictions
Many patients don’t need to limit their activity, except to avoid activities like weight lifting that cause a rise in blood pressure. However, if your child’s heart doesn’t pump normally or if you have heart rhythm issues, you may need to limit your activity to your endurance. Your cardiologist will help determine if your child needs to limit your activity.

Preventing Endocarditis
People who have l-transposition of the great arteries may require endocarditis prophylaxis if they have a prosthetic valve. In the majority of patients, there is no prosthetic valve and the cardiologist will be able to determine if the patient needs to keep taking routine antibiotics before certain dental work.