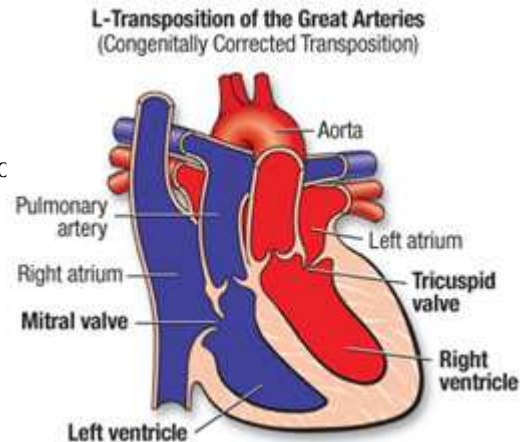


I-Transposition of the Great Arteries

What is it?

I-transposition of the great arteries (also known as levo-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 or obstruction to flow into the pulmonary artery.



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 me?

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. Unless these problems are fixed in childhood, an adult patient may still occasionally be blue.

If I-transposition was repaired in childhood what can I expect?

Most children without a VSD or pulmonary valve obstruction won't need surgery. Even if the repair was performed, most are still left with a right ventricle that pumps to the aorta.

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Can further repair be done in adulthood?

A first surgery or repeat surgery may be needed in adulthood. For example, the tricuspid valve may become leaky and need to be replaced. There are rare patients who may benefit from a complex procedure called a “double switch.”

Problems you may have

Patients may have problems with heart muscle, tricuspid valve leakage or heart rhythm as they enter the third and fourth decades of life, 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 I-transposition. Rapid heart rhythms are less common than in d-TGA patients.

Ongoing Care:

What will I need in the future?

A cardiologist with expertise in managing adults with congenital heart disease should evaluate you regularly, at least once a year and more often if you have symptoms. 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. If you have a pacemaker, you'll need more frequent visits to monitor the pacemaker function.

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 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 you need to limit your activity.

Preventing Endocarditis

People who have I-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.

Pregnancy

Most women with I-transposition can carry a pregnancy with only mildly increased risk. The risk from pregnancy to the mother increases if heart failure or abnormal heart rhythms (arrhythmias) is present. It's important to consult with a cardiologist experienced in caring for patients with congenital heart defects before pregnancy to find out the health risks.

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Will I need surgery as an adult?

Most patients with I-transposition have not had prior surgery and will never require surgery. Some patients need surgery to repair abnormal valves or control heart rhythm disturbances. Patients who still have a ventricular septal defect and pulmonary stenosis may benefit from surgery, which should be performed by a heart surgeon with a great deal of experience in congenital heart defects.