Transposition of the Great Arteries, L-Type

What Is It?

This is an abnormality of the positions of the two ventricles, or pumping chambers of the heart, and the two great arteries, the aorta (which carries blood to the body) and the pulmonary artery (which carries blood to the lungs).

In this condition, the atria, or upper chambers of the heart, are in their normal positions. Blood from the right atrium crosses a mitral valve and enters a morphologic left ventricle that is on the right side. The blood leaves this ventricle and enters the pulmonary artery that is also in the wrong position. It is posterior or behind the aorta, which is opposite of the normal heart. Blood from the left atrium crosses a tricuspid valve and enters a morphologic right ventricle that is on the left side. The blood leaves this ventricle and enters the aorta that is also in the wrong position. It is anterior (in front of) to the pulmonary artery which again is opposite of the normal heart.

Transposition of the Great Arteries: L-Type may be associated with Ventricular Septal Defects (VSDs), or holes in the muscle wall that separates the two ventricles. In addition, the pulmonary artery (PA), which carries blood from the heart to the lungs, may be narrowed (Pulmonary Stenosis). Abnormalities in the form of the tricuspid valve and with the heart's natural pacemaker may also occur.
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What Are Its Effects?

Unlike the other form of Transposition (Transposition of the Great Arteries: D-Type), the blood flow in this defect is normal. Oxygen-rich blood from the lungs is pumped through the aorta to the body and oxygen-poor blood from the body is pumped through the pulmonary artery to the lungs. Therefore, the abnormal positions of the ventricles and great arteries do not cause problems per se.

However, the defects that may be associated with this condition, such as ventricular septal defects, narrowing of the pulmonary artery, defective tricuspid valve, and a defective pacemaker, many case difficulties.

Perhaps the most serious complication that may arise stems from the abnormal conduction system. (Please see Electrophysiology for a discussion of normal conduction.) Because the conduction pathways differ significantly from the normal arrangement, the beating of the heart may be abnormal. In extreme cases, there may be the development of third degree, or complete, heart block, in which the atrial and ventricular pumping become completely out of synch.
How Is It Treated?

Though Transposition of the Great Arteries: L-Type itself does not require treatment, the defects that may be associated with it often do. If necessary, ventricular septal defects may be sutured or patched, an abnormal tricuspid valve may be repaired or replaced, or a narrowed (stenotic) pulmonary artery may be widened with a patch.

In cases where the heart's pacemaker is not functioning properly because of the abnormal conduction pathways, an artificial pacemaker may be inserted.

The most common postoperative difficulties involve heart block (the atria and ventricles do not pump in the proper sequence with each other), which may occur despite extreme care to avoid interruption of the conduction pathways. External or internal pacemakers may be implemented to correct these problems.

Another long term concern is the function of the ventricles. In this condition, the left ventricle is pumping under low pressure to the lungs, which is tolerated well. The right ventricle, however, is under a high pressure work load and in some children can begin to fail over time.

The length of the postoperative hospital stay depends on the nature of the repairs undertaken.