

CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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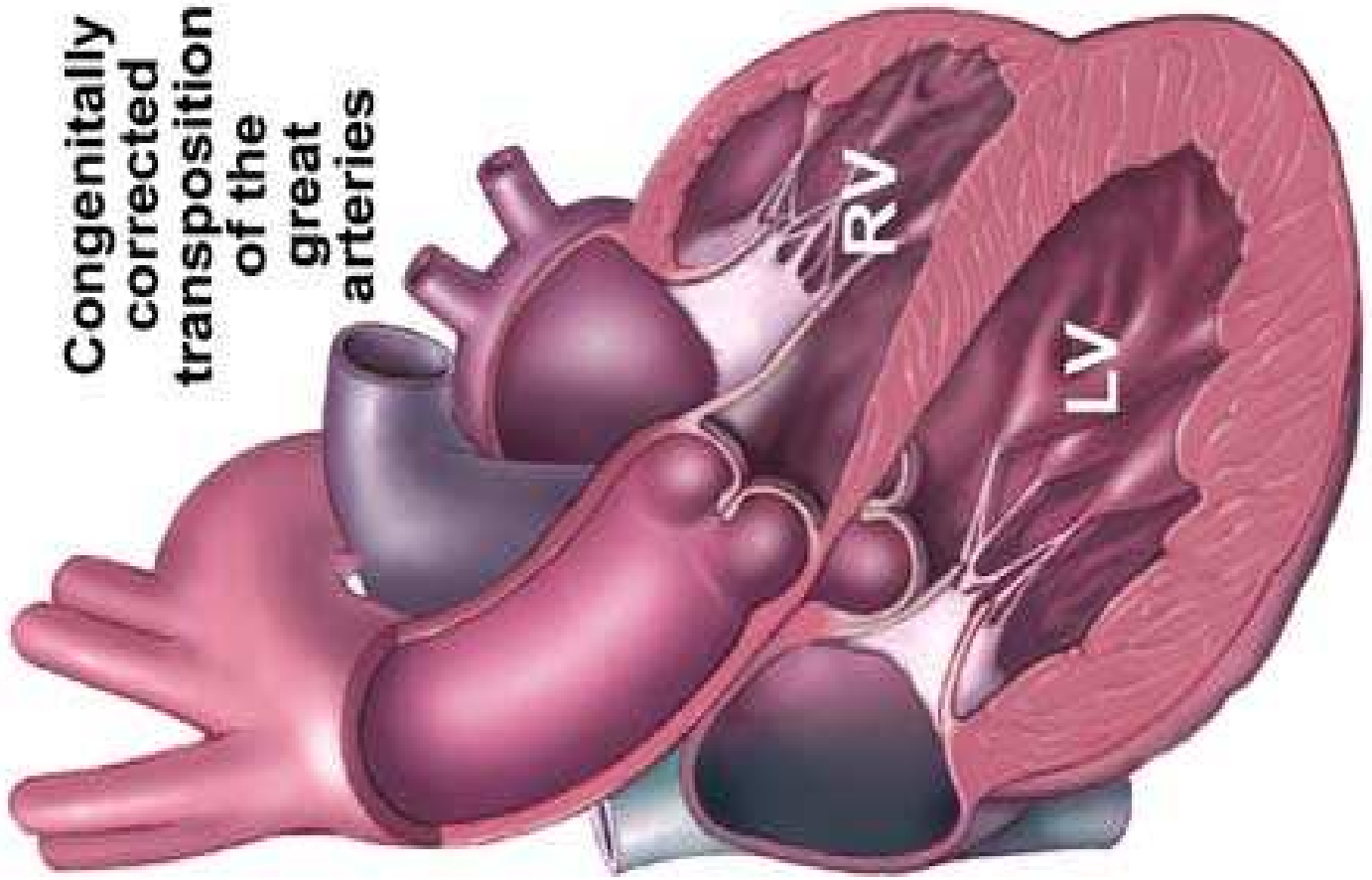
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- **Congenitally** -- existing at birth.
- **Corrected** -- blood flowing in correct direction.
- **Transposition** -- reversed or switched.
- **Great arteries** -- the aorta and pulmonary artery.

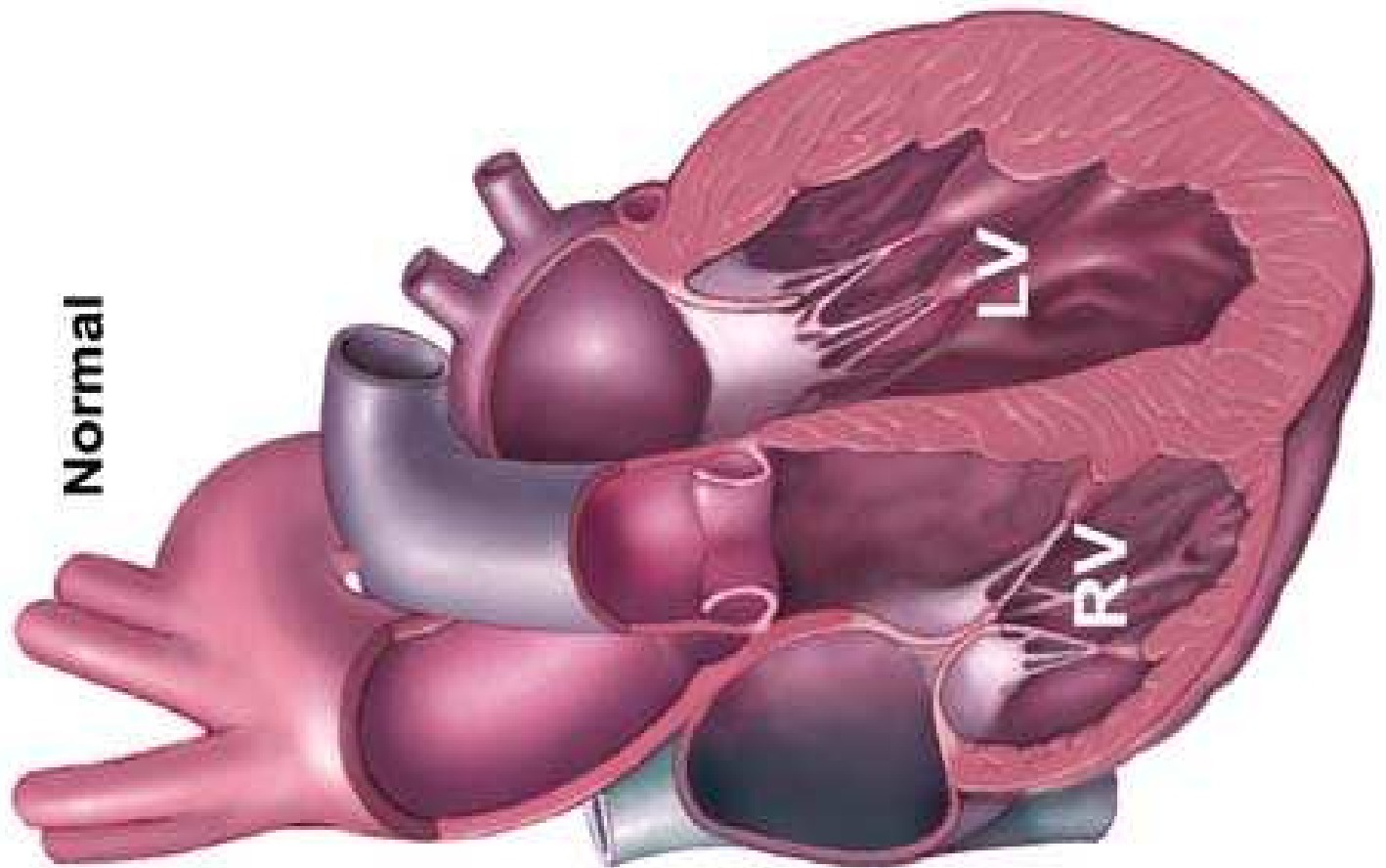
ccTGA

- Rare heart defect.
- Affects about 1 in 25,000 births.
- Occurs equally in boys and girls.

Congenitally corrected transposition of the great arteries



Normal



- Since blood is bumped to the right places, ccTGA is missed until adulthood.

The pumping chambers of the heart are reversed.

- The right ventricle pumps blood to the body.
- The left ventricle pumps blood to the lungs.

RIGHT VENTRICLE

- The ventricle that ended up on the left ,now has to do the job of pumping blood out to the body, was not really "made" to do this job. It is much more suited to pump blood to the lower pressure lung circuit.
- Since the right ventricle is not equipped to pump blood to the higher pressures of the body as opposed to the lower pressures in the lungs, extra work needed to pump blood for 20 or 30 years or more, it can then begin to weaken, leading to symptoms of congestive heart failure.
- Often this is due to deterioration of the tricuspid valve.

WHY?

Early in fetal life the heart first forms in the shape of a tube which bends and folds in on itself, creating the four heart chambers and the four heart valves. If the tube bends to the left instead of the right, the ventricles are reversed: the right ventricle is on the left and the left ventricle is on the right. Two of the heart valves "follow" the ventricles so they are also reversed: the mitral valve is on the right and the tricuspid valve is on the left. Although the two heart valves and the two great arteries, the pulmonary artery and the aorta, are transposed or exit from the "wrong" ventricle, the blood flows to the correct place because the ventricles are also reversed. That is why this heart defect is called "corrected" transposition

The reversal of the ventricles alone does not usually have an adverse effect.

The early health effects of corrected transposition result from the other associated heart problems :

VSD (60-80%)

PS (30-50%)

Tricuspid Valve abnormalities (14-56%).

Heart Block (30-45% post surgery, 2% per year in the absence of surgery).

The effects of an abnormal **Tricuspid Valve** vary based on how much the valve leaks. Mild leakage is tolerated quite well and does not need to be treated. If the leakage is moderate to severe, the forward flow of blood out to the body is limited and symptoms of congestive heart failure occur including low energy levels, clammy sweating, fast breathing, poor appetite . If the valve is too leaky, heart medicines may help but sometimes an operation to either repair or replace the valve is needed.

Heart Block : an abnormally slow heart rate caused by a block in the heart's electrical system where the impulse passes from the heart's upper chambers to the lower chambers.

If there are no other problems such as a VSD, pulmonary stenosis, and/or a leaky tricuspid valve, then the slow heart rate may not cause symptoms, at least not early on. If there are other heart problems the slow heart rate further impairs the heart's ability to meet the body's needs, adding to the symptoms of congestive heart failure. In this case a pacemaker may be needed.

Given current improvement in medical and surgical treatments, the outlook for patient with corrected transposition and associated heart problems is good.

Long-term there may be problems with heart failure and abnormal heart rhythms, so ongoing follow-up by a heart specialist is needed.

PREGNANCY

Careful consideration is needed for women with corrected transposition prior to pregnancy as congestive heart failure can result from the extra demands pregnancy places on the heart.

Pre-pregnancy counseling is crucial for all patient to anticipate and treat possible complication.

Connolly HM, J Am Coll Cardiol 1999

SBE

Subacute Bacterial Endocarditis :is an infection of the heart caused by bacteria in the blood stream. patient with heart defects are more prone to this problem because of the altered flow of blood through the heart. SBE can occur after dental work or medical procedures on the GI or respiratory tract because these procedures almost always result in some bacteria entering the blood. Fortunately, the problem can usually be prevented by giving an antibiotic before these procedures.

Functional Status

An individual exercise program is best planned with the doctor so that all factors can be included.

There are few restrictions for patient with corrected transposition but this depends on the presence and severity of other heart defects.

If a pacemaker or valve replacement is needed, the patient will usually be restricted from vigorous or competitive sports but can participate in non-contact sports.

The doctor may recommend that the patient be able to self-limit their activity, which means that they should be allowed to rest whenever they feel the need to do so.

Functional Status

ccTGA with associated lesion:
60% no symptom, 25% mild symptom.
ccTGA with no associated lesion:
73% no symptom, 22% mild symptom

Graham J Am coll cardiol 2000