Corrected Transposition of the Great Arteries (C-TGA)

Patient Information Series – What you should know, what you should ask.

What is corrected transposition of the great arteries (C-TGA)?

C-TGA is a disease of abnormal heart development where the right and left pumps (ventricles) of the heart can be reversed and develop on the opposite side of the heart from where they are usually found. In addition, the major blood vessels leaving these chambers are also on opposite sides which balances, or "corrects" the reversal of the chambers to some extent.

C-TGA is rare, present in only 1% of all children with congenital heart defects. In the absence of other serious abnormalities in the heart, symptoms in fetal and neonatal life can be mild and surgical repair may only be required later in life.

In some cases, C-TGA is associated with other cardiac abnormalities. In 50% of cases, the heart is located on the right side of the chest (dextrocardia). Ventricular septal defect (VSD), or "hole in the heart" is noted in 80% of cases. Narrowing of the pulmonary artery (stenosis), bringing blood to the lungs, is noted in 50% of patients. In 30% of cases, defects (dysfunction) are noted in the valve of the system pump. Sometimes the pump chambers may be underdeveloped and abnormalities of heart rhythm may occur. The presence of additional cardiac abnormalities in this disease increases the likelihood of surgery.

How does a C-TGA happen?

A normal heart has two lower chambers (ventricles), one on the left and the other on the right. Both function together to form a pump to distribute blood flow to the lungs (right) and around the body (left). The left pump provides a strong pump action to distribute blood flow around the body (systemic circulation), while the right a somewhat weaker pump, provides blood flow to the lungs (pulmonary circulation.)

In C-TGA the right and left pumps are reversed so that the weaker pump has to create more pressure for the systemic circulation (systemic ventricle), while the stronger pump has less work to supply blood to the pulmonary circulation. If only pump reversal occurs during C-TGA, and no other cardiac abnormalities are noted, then no significant changes to blood flow (hemodynamic changes) are observed. But over time, if the weaker pump is unable to keep up and supply blood to the systemic circulation, the heart can become increasingly weaker, leading to heart failure, and in this case, surgery is required.

How are chromosomes relevant to a C-TGA?

The mechanism whereby C-TGA develops is unknown and is considered multi-factorial. Familial incidence is rare and has a recurrence risk of about 2% in first-degree relatives. Associated chromosomal abnormalities are also uncommon.

Should I have more tests done?

If C-TGA is suspected on routine ultrasound screening, a detailed ultrasound of the heart (fetal echocardiography) by a specialist is indicated. In addition to congenital heart defects, genetic syndromes can be noted in 40-50% of cases. In this regard, genetic counseling should be considered, especially if there are additional features that may suggest genetic or chromosomal abnormalities.

What are the things to watch for during the pregnancy?

Given the characteristics of the fetal cardiovascular system, the structural changes seen in this condition do not cause special adverse events in fetal life. However, sometimes different types of abnormal heart rhythms can be encountered and the fetal heart rhythm of the fetus should be carefully evaluated.

What does it mean for my baby after it is born?

The course of the disease differs depending on the structural and functional changes encountered in C-TGA. In some cases, there is no need for surgical intervention. In severe cases, several surgeries may be required in the first months of life.

Will it happen again?

C-TGA may not be sporadic and can recur in subsequent pregnancies. Therefore pregnancies should be examined earlier with a detailed ultrasound of the fetal heart and careful follow-up.

What other questions should I ask?

- Is the baby's heart rate and rhythm normal?
- Are there are other structural differences present in the baby besides the heart?
- Should I have an amniocentesis?
- Where my baby should be born?
- Do I need a Caesarean Section?
- What is the risk that C-TGA will recur in another pregnancy?

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