Total Anomalous Pulmonary Venous Return (TAPVR)

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

What is a TAPVR?

TAPVR accounts for 1% of all children with congenital heart defects. It occurs when there are structural differences in the blood flow to the heart. Usually, oxygen-rich blood from the lungs enters the left upper chamber (atrium) of the heart. This blood then enters the lower chamber (ventricle) where it is pumped to the body through the aorta.

In TAPVR, oxygen-rich blood mistakenly enters other structures, usually the right atrium where it mixes with less oxygen-rich blood coming from the rest of the body. There are several anatomical types of structural differences in the blood vessels in TAPVR. Surgical correction is required in each case. Sometimes, there is an additional problem of “narrowness “of the pulmonary veins, along with differences in where these veins drain back into the heart. This can cause variation in the clinical symptoms. Children with narrowing (stenosis) of the pulmonary veins in addition, usually have more severe symptoms and can have a duskiness to their skin colour called cyanosis. When this is present, earlier surgical correction is required.

How does a TAPVR happen?

There are two types of blood circulation in the human body - systemic and pulmonary circulation. The systemic circulation starts from the left atrium, left ventricle, and aorta, where oxygen-rich blood gets delivered around the body, before ending with oxygen-poor blood from the tissues returning to the heart through the systemic veins.

Pulmonary circulation involves oxygen-poor blood entering the right atrium and then into the right ventricle, where it gets directed to the lungs through the pulmonary artery. It is here that the blood picks up oxygen from the lungs to send around the body. First, it enters the left atrium, usually through the pulmonary veins. If all the pulmonary veins are not draining into the left atrium, then oxygen-rich blood is not delivered to the systemic circulation and to the body. All structural differences in this arrangement whereby there is no direct connection between the pulmonary veins and the left atrium is called Total Anomalous Pulmonary Venous Return (TAPVR).

How are chromosomes relevant to a TAPVR?

The precise way TAPVR develops in fetal life is unknown. Chromosomal abnormalities are uncommon in this disorder.

Should I have more tests done?

If TAPVR is detected in the fetus, a detailed fetal ultrasound of your baby's heart (fetal echocardiography) is recommended. You will likely be referred to a Maternal Fetal Medicine Specialist and Pediatric Cardiologist to have this assessment. A direct association between TAPVR and chromosomal abnormalities is not noted however several gene defects have been found in association with this disorder. If there are additional ultrasound indicators to suspect other abnormalities, your doctor will likely suggest a consultation with a geneticist for further discussion.

What are the things to watch for during the pregnancy?

In fetal life, the fetus does not need to use its lungs in a significant way as the placenta plays a key role in delivering oxygen. Hence, the mother and the fetus will likely not demonstrate any special symptoms related to this condition. Serial fetal echo studies may be required, however, as the pregnancy advances, to assess any changes in the blood flow in the heart as a consequence of this structural difference. A plan for delivery in a center equipped to care for these babies after birth will be required.

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

After birth, your baby will be watched carefully for any signs of severe disease. An echocardiogram will be performed to verify the diagnosis and establish the type of TAPVR and rule out other structural changes in the heart such as the presence and degree of obstruction in the venous system.

The baby will be watched for complications such as pneumonia and acute heart failure. Surgical intervention is required to correct this condition, the timing of which will be decided by the pediatric cardiologist and cardiovascular surgeon, after newborn examination and investigations are completed. TAPVR is a serious heart anomaly. Without corrective surgery, many babies will die in the first year of life.

Will it happen again?

Recurrence of TAPVR in siblings and other family members has been reported. Because of this recurrence risk, future pregnancies should be evaluated by careful ultrasound examination of the heart with fetal echocardiography.

What other questions should I ask?

- What type of TAPVR does my baby have?
- Are there any other problems in the heart or elsewhere in the baby?
- Where should my baby be delivered?
- Is there any special testing I should do?
- Can I meet the Pediatric Cardiologist who will care for my baby before its born?

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