What is Total Anomalous Pulmonary Venous Return?

Total anomalous pulmonary venous return (TAPVR) is a rare congenital heart defect that causes cyanosis or blueness. The basic problem is that the blood flow coming back from the lungs (which is full of Oxygen) through pulmonary veins is diverted to the right atrium so that there is complete mixing of blood (red and Blue) within the heart. These abnormalities are divided into two major groups - obstructed and unobstructed. Unobstructed is the most common and this frequently involves the common pulmonary vein being connected by an abnormal vertical vein to the superior vena cava and emptying into the right atrium. There can also be a direct connection of the common pulmonary vein to the right atrium. Either way this causes complete mixing of the blood within the right atrium. These babies are usually a little blue at birth, but not necessarily very sick. Many times they will go home from the nursery and be found to be breathing fast by the pediatrician. This is because the mixing of blood causes the heart to work harder than normal. This is usually relatively easy to fix. It does involve open-heart surgery. Once fixed, the heart is for the most part normal.

Obstructive veins are another matter. These babies are usually very ill at birth. In this situation the blood flow leaving the lungs through the pulmonary veins is not only going to the wrong place but is at least partially blocked, this makes it harder for blood to enter the lungs. Most of the time these abnormal connections occur in the liver. They can occur in the upper part of the chest as well. When the veins are obstructed It also makes the pressure in the right ventricle higher than normal. When blood cannot easily enter the lungs the babies are very blue because not enough blood goes to the lungs and there is a lot of mixing of blood (blue blood with red blood) at the level of the PDA and Foramen Ovale (opening in wall between the upper chambers of the heart). Because not enough oxygen is being circulated in the body the baby becomes acidotic and the muscle tissue and kidneys, etc. do not work well. When the oxygen level is low, the arteries in the lungs tend to tighten up, making it harder for blood to be pumped through the lungs and thus the situation even worse. When the lungs are full of blood they become stiff, making it harder to breathe. Very frequently in this condition the actual veins draining back from the lungs are smaller than normal. This is because they never developed normally in the first place. This condition is lethal unless it can be fixed quickly after diagnosis.

Most of the time we are able to diagnose this condition by echocardiogram. One can see the abnormal flow patterns of blood coming from the lungs into the SVC or we can see abnormal patterns in the liver, which is where most of the obstructed veins return. We also look to see if we can see the normal pulmonary veins returning to the left atrium. We can measure oxygen levels in the right atrium with an umbilical catheter and this will be abnormally high.

TAPVR can closely mimic a condition called persistent fetal circulation. In this case the pulmonary veins are normal but very little blood flow is going to the lungs. This can sometimes occur with stressed or infected babies and can be very difficult to treat. The pressures in the lungs can be very high and little blood flow goes through the lungs. As with TAPVR this is a lot of mixing of blood within the heart and the oxygen level can be quite low. This condition can be treated with a ventilator, pulmonary vasodilators, nitric oxide and sometimes ECMO. It can be very difficult to distinguish these two conditions as they have many similar clinical features.

TAPVR is not something that can be treated medically. The longer the delay in surgery, the worse the outcome. This condition is frequently associated with other rare conditions such as dextrocardia, and ambiguous situs. In this situation a baby's internal organs can have either two right sides or two left sides.

The outcome for unobstructive TAPVR is usually quite good. The outcome of obstructive TAPVR can vary and ultimately depends on the overall size of the pulmonary veins. Many attempts have been made to enlarge these surgically and with balloons and stents but the results are seldom satisfactory.

If you have any questions, please ask one of the doctors.

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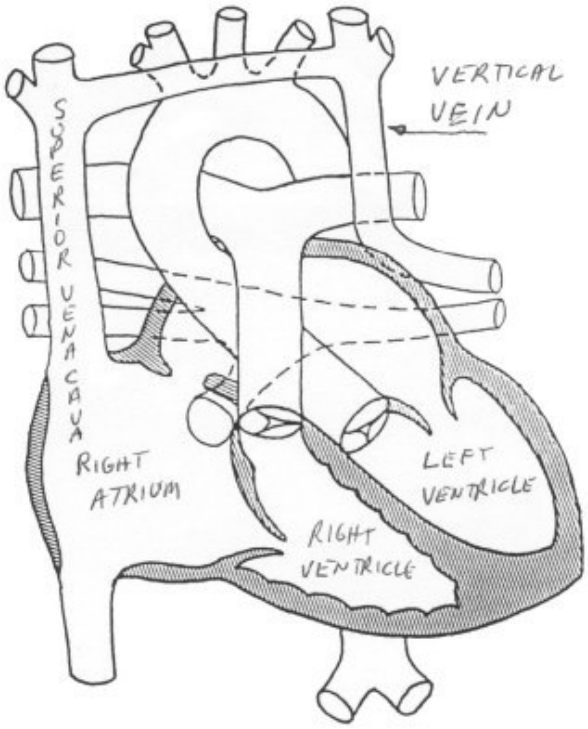
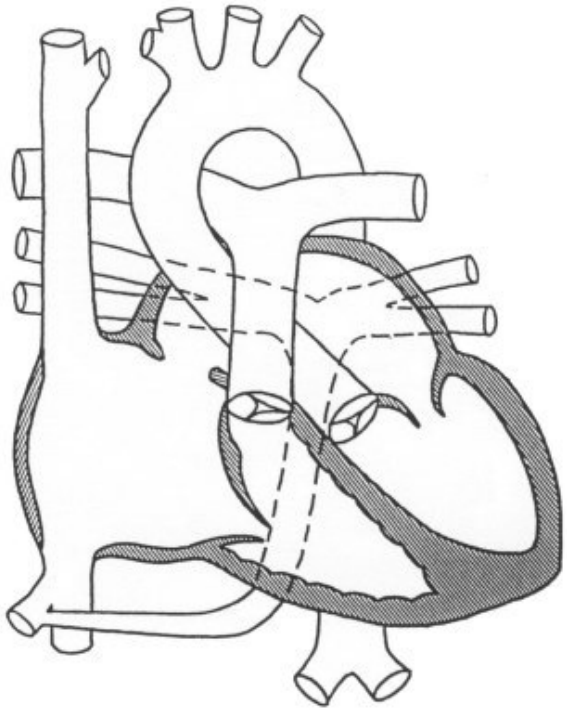


Total anomalous pulmonary venous connection
Mixed drainage with:
Left upper pulmonary vein to left vertical vein
Left lower and all of right pulmonary veins to orifice of coronary sinus
Atrial septal defect



Total anomalous pulmonary venous connection to ductus venosus (infradiaphragmatic)
Atrial septal defect