



Cardiac Defects: Total Anomalous Pulmonary Venous Return

Arteries carry blood away from the heart; veins carry it toward the heart. The pulmonary veins are very important. They carry the “red” blood that has picked up oxygen in the lungs back to the heart to be pumped out to the body.

Typically, there are four pulmonary veins, two from each lung, and normally they all connect directly into the heart’s left atrium (left upper chamber). From the left atrium, blood travels into the left ventricle (left lower chamber), which pumps it out to the aorta and to the body.

Anomalous means abnormal. In a child born with total anomalous pulmonary venous return (TAPVR), the pulmonary veins connect to other veins and ultimately drain their blood into the right atrium. You also may hear the term *total anomalous pulmonary venous connection* (TAPVC). Ordinarily, the right atrium only receives “blue” blood coming back from the body without oxygen, which then passes into the right ventricle and is pumped to the lungs. In TAPVR, the right atrium receives both “blue” blood from the body and “red” blood from the lungs.

As a result, the right side of the heart is overworked, pumping the extra blood it receives. Frequently, there is obstruction (blockage) to the veins that receive blood from the pulmonary veins, which causes a backup of blood in the lungs. This disrupts the normal flow of blood between the lungs and the body. In addition, the blood leaving the heart for the body doesn’t have as high a level of oxygen as it should.

Children with TAPVR also have other heart defects. They have a hole in the wall separating the two upper chambers of the heart (atrial septal defect) and may have a patent ductus arteriosus, an extra blood vessel between the pulmonary arteries and the aorta. These heart defects actually can help a child with TAPVR survive by allowing

more blood to get from the right side of the heart to the left side and out to the body.

Your baby’s cardiologist will explain your baby’s heart anatomy in detail. Generally, there are four types of TAPVR:

- Supracardiac TAPVR: The pulmonary veins drain into the right atrium through the superior vena cava. (Normally this large vein carries only deoxygenated, or “blue,” blood into the right atrium.)
- Infracardiac TAPVR: The pulmonary veins drain into the right atrium through the liver (hepatic) veins and the inferior vena cava (another large vein that normally carries only deoxygenated blood).
- Cardiac TAPVR: In one type, the pulmonary veins can directly enter into the right side of the heart, into the right atrium. In the second type, the pulmonary veins can drain into the coronary sinus, a vein that usually only carries blood coming out of the heart muscle. This vein is usually very small but becomes quite large with this abnormal amount of blood.
- Mixed TAPVR: The pulmonary veins split up and drain partially to more than one of these options.

Symptoms

TAPVR symptoms include

- blue or purple tint to lips, skin, and nails (cyanosis)
- rapid breathing or working harder while breathing, especially while eating
- heart murmur (an extra heart sound when a doctor listens with a stethoscope).

The severity of TAPVR symptoms varies.

How is TAPVR diagnosed?

In some cases, newborns with TAPVR have difficulty breathing and quickly become very ill. This occurs when the pulmonary veins are too narrow or are obstructed at some point, and blood can’t flow from the lungs as quickly as it should. This is called *TAPVR with pulmonary obstruction*.



In other cases, TAPVR is diagnosed in the first few months of life after a child demonstrates milder symptoms such as a heart murmur or cyanosis.

Diagnosis of TAPVR may require some or all of these tests:

- echocardiogram—sound waves create an image of the heart
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- chest X ray
- pulse oximetry—a noninvasive way to monitor the oxygen content of the blood
- cardiac catheterization—a thin tube is inserted into the heart through a vein or artery in either the leg or through the umbilicus (“belly button”)
- cardiac magnetic resonance imaging—a three-dimensional image shows the heart’s abnormalities.

What are the treatment options for TAPVR?

TAPVR requires open-heart surgery in all cases. Critically ill newborns will have surgery immediately. If the child is not critically ill, surgeons may wait up to 2 months to perform surgery, depending on the strength of the child and on the heart anatomy.

To understand the surgery, one important thing to know about TAPVR is that the pulmonary veins, despite their abnormal connections to other veins, all end in a collection (called a confluence) at the back of the left atrium. The surgeon opens the confluence so that the veins can drain into the left atrium and then ties off all abnormal connections between the pulmonary veins and other veins, so that blood can follow only the path to the left atrium. The surgeon also closes septal defects (the abnormal holes) with tiny patches or stitches and closes the

patent ductus arteriosus. As the child ages, the lining of the heart grows over the stitches.

Another surgical option is a sutureless open-heart surgery, which may provide certain benefits and decreased mortality for patients with a pulmonary valve obstruction (PVO; Zhang et al., 2016).

What is the follow-up care for TAPVR? Through Age 18

Children who were born with TAPVR and had it repaired must continue to see a pediatric cardiologist regularly. Children who were critically ill as newborns may have a longer road to recovery. In most cases, however, children won’t experience long-term effects—they won’t have to remain on any medicines or limit physical activity.

Rarely, the pulmonary veins become obstructed later in life and additional surgery or a catheterization procedure is required. Also rarely, children experience arrhythmia (abnormal heart rhythm), which may be treated with medicines, radiofrequency ablation, or a pacemaker.

Into Adulthood

It is important that children who were born with TAPVR continue to see a cardiologist. Because of enormous strides in medicine and technology, today most children born with TAPVR go on to lead productive lives as adults.

References

- Zhang, C., Ou, Y., Zhuang, J., Chen, J., Nie, Z., & Ding, Y. (2016). Comparison of sutureless and conventional techniques to repair total anomalous pulmonary venous connection. *Seminars in Thoracic and Cardiovascular Surgery*, 28(2), 473–484.

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