Total anomalous pulmonary venous return (TAPVR) occurs when the veins bringing blood back from the lungs (pulmonary veins) do not connect to the left upper chamber like normal. The pulmonary veins go to the heart in an abnormal (anomalous) way. Sometimes not all the veins have an abnormal connection. If there are some abnormal connections, but one or more veins return normally to the left upper chamber, this is called partial anomalous venous return (PAPVR). Oxygen-rich blood mixes with oxygen-poor blood in the right side of the heart, so less oxygen is gets to the body. A baby with TAPVR often has an opening between the two upper chambers of the heart. This is called an atrial septal defect (ASD), and it lets the mixed blood get to the left side of the heart and be pumped to the rest of the body. TAPVR is a critical congenital heart defect, meaning it can cause serious health problems or even death if left untreated. PAPVR is not as critical as TAPVR.

Image courtesy of the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities
How common is it?

TAPVR is rare. About 1 out of every 10,000 babies is born with TAPVR.

What causes it?

The cause of TAPVR is unknown in most babies. There may be many factors that cause TAPVR, but more research is needed to understand the exact cause for it.

How is it diagnosed?

TAPVR can be diagnosed during pregnancy or after. During pregnancy screenings are done to check for birth defects. After birth a doctor will do a physical examination to see whether the baby has blue-colored skin and lips, called cyanosis. This can be a sign of low levels of oxygen in the blood. The doctor will listen to the baby’s heart. If the doctor hears a heart murmur, or a “whooshing” sound, that can be a sign of a heart defect. A doctor also might see that a baby is having trouble breathing, a pounding heart, and poor feeding, all of which could be signs of a heart defect. Symptoms are often seen soon after birth in a baby with TAPVR. A screening test called a pulse oximetry screen is done shortly after birth to check for critical congenital heart defects. A pulse oximeter is a tool that detects oxygen levels in blood. Low levels of oxygen in the blood could mean there is a heart defect. If a baby fails the screening, then the doctor should perform a diagnostic test called an echocardiogram to check for defects in the heart.

How is it treated?

Treatment for a TAPVR requires surgery. The goal of surgery is to restore normal blood flow through the heart. Your child’s doctor should discuss treatment options with you. Regular visits to a cardiologist, a doctor who specializes in the heart, will be necessary to avoid problems and watch for any other health conditions.

For more information:

American Heart Association
http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/About-Congenital-Heart-Defects_UCM_001217_Article.jsp#.Wv2YtPnwbcs

Centers for Disease Control and Prevention
https://www.cdc.gov/ncbddd/heartdefects/tapvr.html

National Heart, Lung, and Blood Institute
https://www.nhlbi.nih.gov/health-topics/congenital-heart-defects