



Total Anomalous Pulmonary Venous Connection (TAPVC)

Note: before reading the specific defect information and the image(s) that are associated with them, it will be helpful to review [normal heart function](#).

What is it?

In total anomalous pulmonary venous connection (drainage, return), the pulmonary veins that bring oxygen-rich (red) blood from the lungs back to the heart aren't connected to the left atrium. Instead, the pulmonary veins drain through abnormal connections to the right atrium.

What causes it?

In most children, the cause isn't known. Some children can have other heart defects along with TAPVC.

How does it affect the heart?

In the right atrium, oxygen-rich (red) blood from the pulmonary veins mixes with low-oxygen (bluish) blood from the body. Part of this mixture passes through the atrial septum (atrial septal defect) into the left atrium. From there it goes into the left ventricle, then into the aorta and out to the body. The rest of the blood flows through the right ventricle, into the pulmonary artery and on to the lungs. The blood passing through the aorta to the body doesn't have a normal amount of oxygen, which causes the child to look blue.

How does TAPVR affect my child?

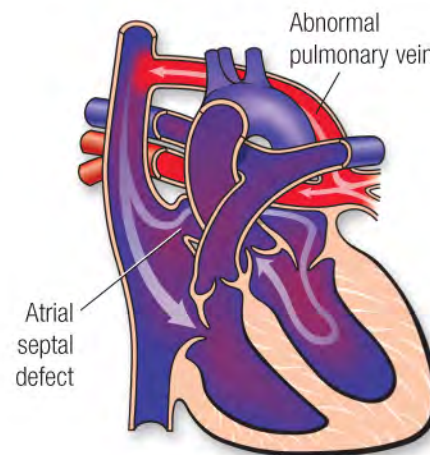
Symptoms may develop soon after birth. In other children, symptoms may be delayed. This partly depends on whether the lung veins are blocked as they drain toward the right atrium. Severe obstruction of the pulmonary veins tends to make infants breathe harder and look bluer (have lower oxygen levels) than infants with little obstruction.

What can be done about the defect?

This defect must be surgically repaired in early infancy. At the time of open-heart surgery, the pulmonary veins are reconnected to the left atrium and the atrial septal defect is closed.

What activities can my child do?

Children with repaired TAPVR may be advised to limit their physical activities to their own endurance. Some competitive sports may have greater risk if there is leftover obstruction in the pulmonary veins, or if the child has heart rhythm problems. Your child's pediatric cardiologist will help determine the proper level of activity.





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What will my child need in the future?

When surgical repair is done in early infancy, the long-term outlook is very good. However, your child will need regular follow-up with a pediatric cardiologist and, once your child reaches adulthood, lifelong regular follow-up with a cardiologist who's had special training in congenital heart defects. Follow-up is needed to make certain that any remaining problems, such as an obstruction in the pulmonary veins or irregularities in heart rhythm, are treated. Some children may need medicines, heart catheterization or even more surgery.

What about preventing endocarditis?

Children with TAPVR are at increased risk for developing endocarditis. Ask your pediatric cardiologist about your child's need to take antibiotics before certain dental procedures to help prevent endocarditis. See the section on [endocarditis](#) for more information.