

## **Total Anomalous Pulmonary Venous Connection (TAPVC)**

### **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 cases, the cause isn't known. Some patients 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 TAPVC affect me?**

Symptoms may develop soon after birth. In other patients, 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. It's rare for an adult to have uncorrected TAPVC. Patients who survive until adulthood usually have only mild cyanosis and no evidence of blockage. Symptoms include shortness of breath and inability to exercise.

### **If my TAPVC was fixed in childhood, what can I expect?**

This defect is almost always surgically repaired in early infancy or childhood. At the time of open-heart surgery, the pulmonary veins are reconnected to the left atrium and the atrial septal defect is closed. The prognosis after surgery during childhood is excellent with uncommon late complications. Rarely, obstruction of the pulmonary veins at the site of reconnection can occur. Occasionally abnormal heart rhythms can also occur.

### **What if the defect is still present? Should it be repaired in adulthood?**

It's rare when the TAPVC hasn't been repaired in childhood, but most adults in this category are able to have surgery.

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### **Ongoing Care**

#### **What will I need in the future?**

Patients with a history of TAPVC should be seen periodically by a cardiologist with expertise in adult congenital heart disease to look for uncommon problems. The long-term outlook is excellent, and usually no medicines and no more surgery or catheterization are needed. You should also consult a cardiologist with expertise in caring for adults with congenital heart disease if you're undergoing any type of non-heart surgery or invasive procedure.

#### **Medical Follow-up**

Your cardiologist can monitor you with noninvasive tests if needed. These include electrocardiograms, Holter monitors, exercise stress tests and echocardiograms. They'll help show if more procedures, such as a cardiac catheterization, are needed. Medicines may only be required if there are abnormal heart rhythms.

#### **Activity Restrictions**

Most adults with repaired TAPVC will not need 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 patient has heart rhythm problems. Your cardiologist will help determine the proper level of activity for you.

#### **Endocarditis Prevention**

Not needed beyond six months after repair.

#### **Pregnancy**

There is little information about pregnancy in patients with repaired TAPVC since it is such a rare defect. Unless there is obstruction, or abnormal heart rhythms are poorly controlled, most women who have had repair should tolerate pregnancy provided that a pre-pregnancy evaluation doesn't reveal any problems.

#### **Will you need more surgery?**

Once the TAPVC has been repaired, it's unlikely that more surgery will be needed. Rarely, an adult patient may have obstruction that may need to be corrected depending on the severity.