

Pulmonary Atresia / Intact Ventricular Septum

What is it?

In pulmonary atresia, no pulmonary valve exists. Blood can't flow from the right ventricle into the pulmonary artery and on to the lungs. The right ventricle and tricuspid valve are often poorly developed.

What causes it?

In most children, the cause isn't known. Some children can have other heart defects along with pulmonary atresia. (Children with tetralogy of Fallot who also have pulmonary atresia may have treatment similar to others with tetralogy of Fallot.)

How does it affect the heart?

An opening in the atrial septum lets blood exit the right atrium, so low-oxygen (bluish) blood mixes with the oxygen-rich (red) blood in the left atrium. The left ventricle pumps this mixture of oxygen-poor blood into the aorta and out to the body. The infant appears blue (cyanotic) because there's less oxygen in the blood. The only source of lung blood flow is the patent ductus arteriosus (PDA), an open passageway between the pulmonary artery and the aorta.

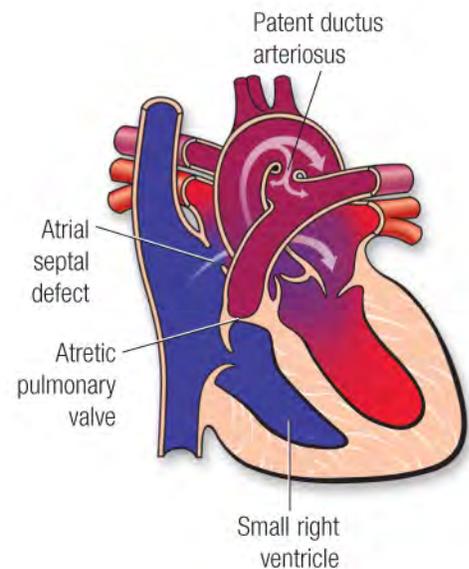
How does pulmonary atresia affect my child?

If the PDA narrows or closes, the lung blood flow is reduced to critically low levels. This can cause very severe cyanosis. Symptoms may develop soon after birth.

What can be done about the defect?

Temporary treatment includes a drug to keep the PDA from closing. A surgeon can create a shunt between the aorta and the pulmonary artery that may help increase blood flow to the lungs.

A more complete repair depends on the size of the pulmonary artery and right ventricle. If the pulmonary artery and right ventricle are very small, it may not be possible to correct the defect with surgery. In some children, abnormal channels (sinusoids) form between the coronary arteries and the right ventricle. These sinusoids can also limit the type of surgery a child can have. In children where the pulmonary artery and right ventricle are more normal in size, open-heart surgery may help the heart work better.



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If the right ventricle stays too small to be a good pumping chamber, the surgeon can connect the body veins directly to the pulmonary arteries. The atrial defect also can be closed to relieve the cyanosis. These surgeries are called the Glenn and Fontan procedures.

What activities can my child do?

Children with pulmonary atresia may be advised to limit their physical activities to their own endurance. Some competitive sports may pose greater risk. Your child's pediatric cardiologist will help determine the proper level of activity.

What will my child need in the future?

Children with pulmonary atresia need regular follow-up with a pediatric cardiologist and, once they reach adulthood, lifelong regular follow-up with a cardiologist who's had special training in congenital heart defects. Some children may need medicines, heart catheterization or additional surgery.

What about preventing endocarditis?

Children with pulmonary atresia 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.