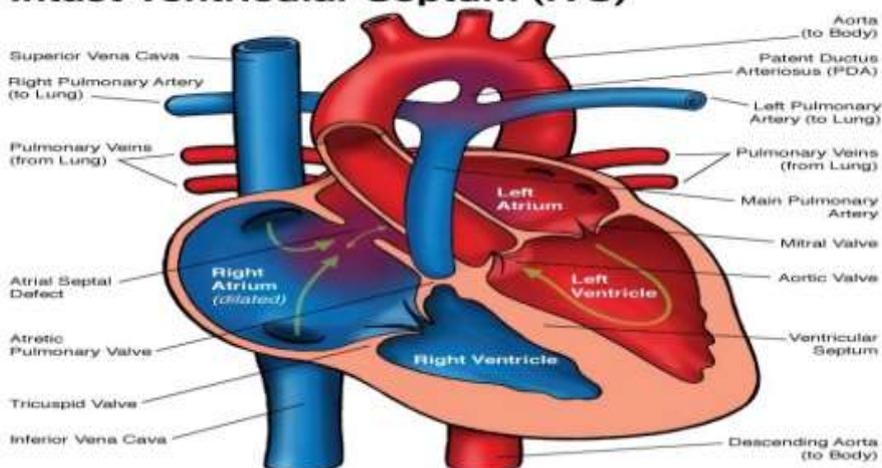


Pulmonary Atresia Intact Ventricular Septum (IVS)



Pulmonary Atresia with Intact ventricular septum

Definition:

In pulmonary atresia, the pulmonary valve simply does not exist, so blood cannot pass from the right ventricle to the lungs. This in turn causes the right ventricle and the tricuspid valve to be poorly developed.

What causes it?

Cause is unknown, and it can be associated with other heart defects.

How does it affect the heart?

The right ventricle is unable to transport blood to the pulmonary artery for oxygenation. This means that the baby will be born blue and cyanosis. The child is able to survive birth however, due to the presence of an ASD (hole between the two atria) and a PDA (connection between the pulmonary artery and the aorta). The deoxygenated blood will mix with the oxygenated blood and be pumped to the aorta via the left ventricle where some of it will be shunted via the PDA to the lung arteries for oxygenation.

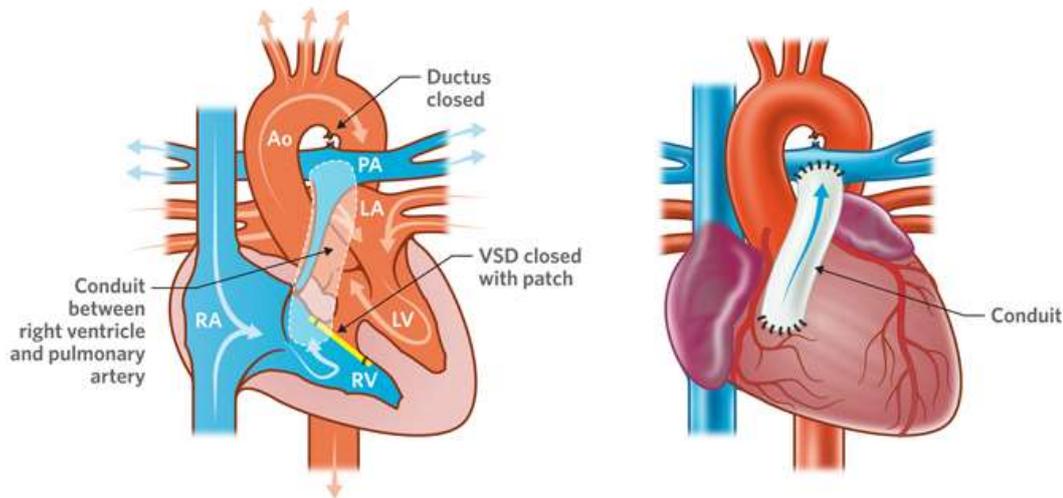
How does it affect the child?

If the PDA narrows or closes, oxygenation of blood will not occur and the child's oxygen levels will be critically low which will cause the baby to be severely blue and death. Symptoms will develop immediately after birth.

Management options:

PDA must be kept open by using a specific medication to preserve life. Sometimes the surgeon can create a connection between the aorta and pulmonary artery to help increase blood flow to the lungs. There are generally two types of surgery that can be done and are carried out according to the size of the right ventricle and pulmonary artery. If they are of adequate size, open heart surgery can be done to repair and essentially create a valve conduit allowing blood to pass from the right ventricle to the lungs.

Repair of pulmonary atresia with VSD



If the right ventricle and the pulmonary artery are small sized, the surgeon can connect the veins directly to the pulmonary artery and bypass the right side of the heart entirely. These surgeries are known as Glenn and Fontan and are done in 2 stages (Glenn done can be done after 3 months and Fontan after 18 months)

What activities can your child do?

Physical activity must be limited as dictated by the pediatric cardiologist.

Follow up in the future:-

Regular follow up with the pediatric cardiologist will be necessary even into adulthood, as medication, catheterization or surgical interventions might be necessary.

Children may need to receive antibiotics before any surgical or dental procedures to prevent infective endocarditis.