

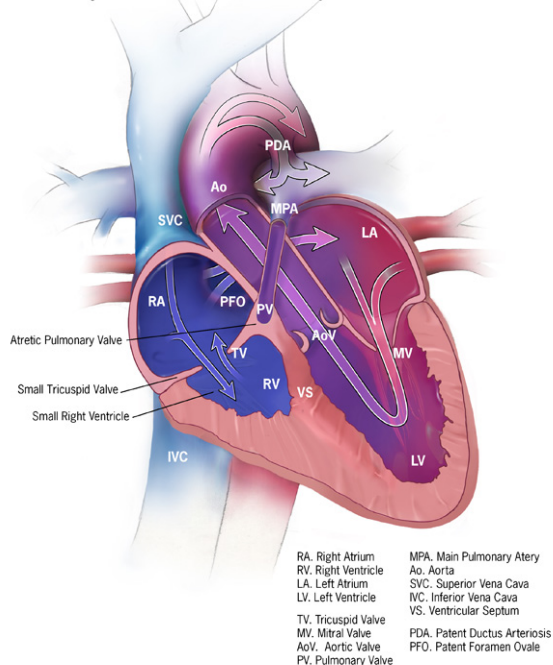
Pulmonary Atresia

What is it?

Pulmonary atresia (pronounced PULL-mun-airy ah-TREE-sha) is a congenital, meaning present at birth, heart defect of the pulmonary valve. The **pulmonary valve** controls blood flow from the heart to the lungs. **Pulmonary atresia** occurs when the pulmonary valve does not form, causing blood to have trouble flowing to the lungs to pick up oxygen for the rest of the body. Pulmonary atresia is considered a **critical congenital heart defect**, meaning it can cause serious health problems or even death if left untreated. Blood must use a different way to get to the lungs. The **foramen ovale** is a normal opening between the upper chambers of the heart during pregnancy that usually closes shortly after birth. In a baby with pulmonary atresia, the foramen ovale will stay open. When it stays open, it is called a **patent foramen ovale (PFO)**. The ductus arteriosus is a blood vessel that lets blood move around the lungs before the baby is born, and it normally closes after birth. When it remains open, it is called a **patent ductus arteriosus (PDA)**. Medicine may be given to keep the ductus arteriosus open. Keeping the PFO and PDA open will let blood flow to the lungs. There are two types of pulmonary atresia:

1. **Pulmonary atresia with an intact ventricular septum.** This type involves a missing pulmonary valve but no opening between the lower chambers of the heart.
2. **Pulmonary atresia with a ventricular septal defect.** This type involves a missing pulmonary valve and an opening between the lower chambers of the heart, called a **ventricular septal defect (VSD)**.

Pulmonary Atresia with Intact Ventricular Septum



Severe TOF/Pulmonary Atresia

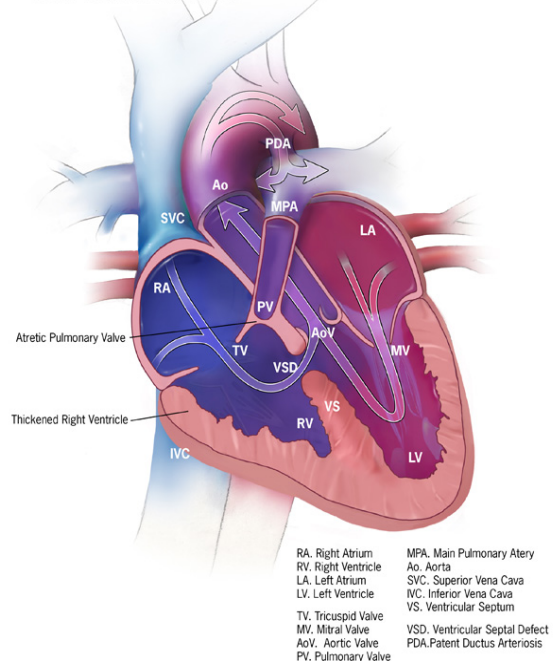


Image courtesy of the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities

How common is it?

Pulmonary atresia is rare. About 1 per 10,000 babies will be born with pulmonary atresia.

What causes it?

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

How is it diagnosed?

Pulmonary atresia can be diagnosed during pregnancy. Screenings are done during pregnancy to check for birth defects. A baby also can be diagnosed with pulmonary atresia after birth if he or she has rapid breathing, poor feeding, blue skin and lip color, and extreme sleepiness. A screening test called a **pulse oximetry screen** is done 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 this screening, then the doctor should perform a diagnostic test called an **echocardiogram** to check for defects in the heart.

How is it treated?

Most babies will need medication to keep the PDA open until the pulmonary valve is repaired. The severity of the defect will determine the treatment. Doctors may try to improve the blood flow by inserting a small tube called a **stent** to keep the PDA open. That would be done by cardiac catheterization. Many babies will need surgery to repair the pulmonary valve. If a VSD is present, a patch is placed over the VSD to help improve blood flow to the lungs and the rest of the body. 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 watch for any other heart conditions.

For more information:

American Heart Association

http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/About-Congenital-Heart-Defects_UCM_001217_Article.jsp#.Wv2YtPnwbc

Centers for Disease Control and Prevention

<https://www.cdc.gov/ncbddd/heartdefects/pulmonaryatresia.html>

National Heart, Lung, and Blood Institute

<https://www.nhlbi.nih.gov/health-topics/congenital-heart-defects>

