

**Department Of Health and Mental Hygiene
Prevention and Health Promotion Administration
Office for Genetics and People with Special Health Care Needs**

Pulmonary Atresia

What is Pulmonary Atresia?

- Heart defect which is present at birth (congenital)
- Condition in which the pulmonary valve does not form properly. The pulmonary valve connects the right side of the heart with the pulmonary artery that carries blood from the right ventricle to the lungs to pick up oxygen
- A solid sheet of tissue forms where the valve opening should be, and the valve is very narrow or stays closed. As a result of this defect, blood from the right side of the heart does not travel to the lungs to pick up oxygen

What is the cause of Pulmonary Atresia?

- It occurs during fetal growth and development but the cause is unknown
- Pulmonary Atresia can occur with or without a Ventricular Septal Defect (VSD)
 1. Without VSD - the condition is known as Pulmonary Atresia with intact Ventricular Septum (PA/IVS)
 2. With VSD - the condition is known as Pulmonary Atresia with VSD. This may be called an extreme form of Tetralogy of Fallot

Signs and Symptoms

- Bluish skin color, especially around lips and fingernails
- Fast and difficult breathing
- Poor feeding
- Lethargy (tiredness)

How is Pulmonary Atresia treated?

- Medication called prostaglandin is used to keep the Ductus Arteriosus open. (Ductus Arteriosus is a blood vessel between the pulmonary artery and aorta that allows blood to flow to the lungs to pick up oxygen)
- Surgical treatment is needed and varies depending on the extent of the problem:
 1. Cardiac Catheterization - a thin flexible tube is threaded into the heart to open up the valve
 2. Open Heart Surgery - used to repair or replace the valve or to place a tube between the right ventricle and the lung arteries

Support Groups and Online Resources

- Congenital Heart Disease Information and Resources
<http://www.tchin.org/>
1-609-822-1572
- Little Hearts Inc.
www.littlehearts.org
1-866-435-4673
- March of Dimes
www.marchofdimes.com
1-410-546-2241
- Office for Genetics and People with Special Health Care Needs
phpa.dhmh.maryland.gov/genetics/
1-855-535-5655
- The Parents' Place Of Maryland
www.ppmmd.org
1-410-768-9100