

## Pulmonary Atresia



### What Is It?

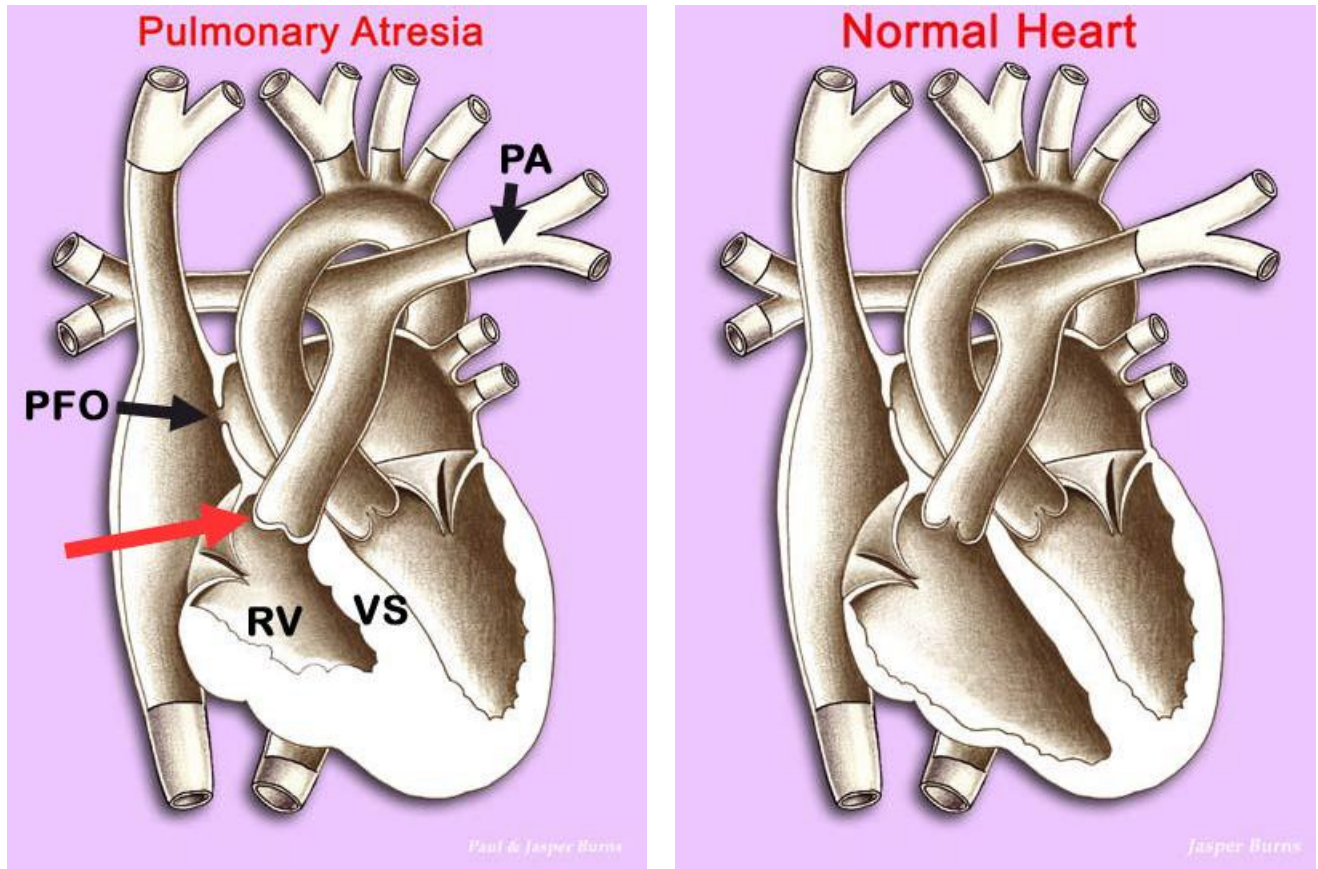
In Pulmonary Atresia, there is no Pulmonary Valve, or opening through which blood may enter the pulmonary artery (PA in illustration below) and be carried to the lungs. (An atresia is a blockage that separates a tube or passage into two separate sections.)

Because the pulmonary valve (red arrow in diagram below) does not form during pregnancy, the right ventricle (RV), which normally pumps blood through this valve, may not develop normally and remain small (hypoplastic).

There are two main types of Pulmonary Atresia, distinguished by whether or not there is also a hole in the muscle wall (ventricular septum (VS)) that separates the right and left ventricles. This hole (not present in the diagram) is known as a Ventricular Septal Defect (VSD). If a Ventricular Septal Defect is present, it may promote growth of the right ventricle during fetal development as there is increased blood flow into this chamber through the hole in the septum.

A small hole in the muscle wall between the heart's upper chambers may also be present, known as an Patent Foramen Ovale (PFO). This is actually a feature of the fetal heart, which usually closes soon after birth but may remain open in this disorder.

Pulmonary Atresia is a rare defect, occurring with equal frequency in boys and girls, and is related to the congenital heart defect known as Tetralogy of Fallot. It develops during the first 8 weeks of pregnancy from unknown causes.



### What Are Its Effects?

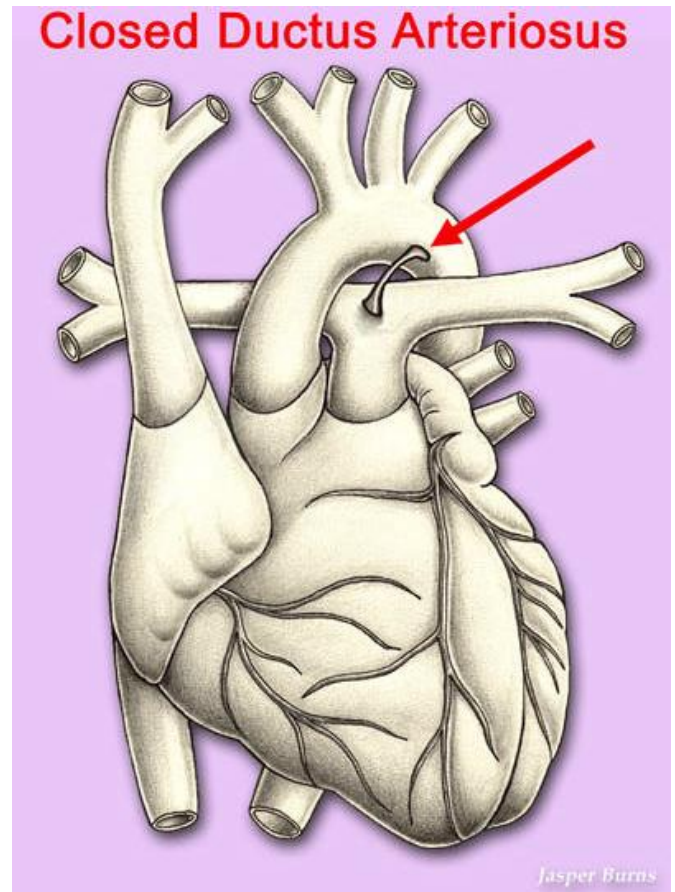
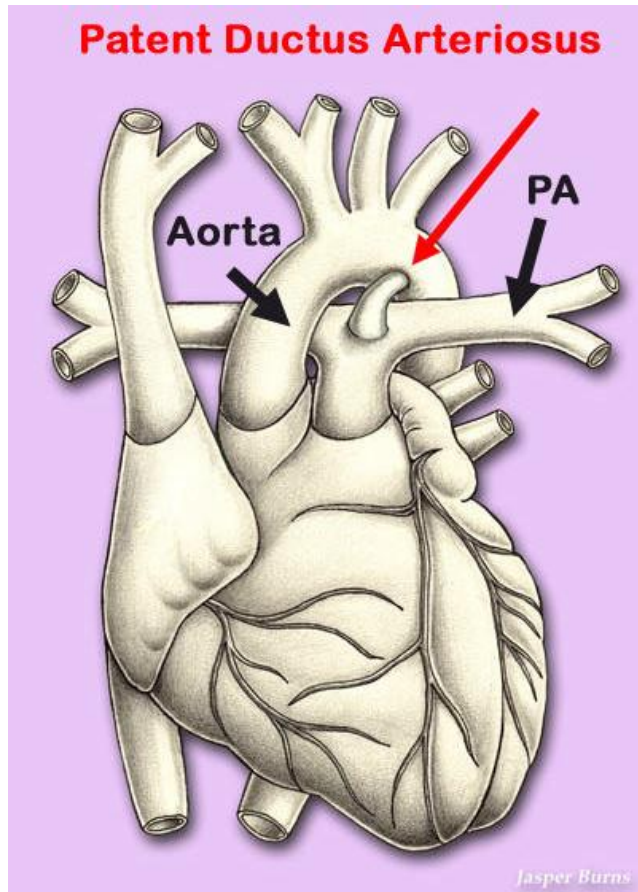
In the normal heart, oxygen-poor blood from the body's tissues is pumped by the right ventricle through the pulmonary artery to the lungs for reoxygenation. In Pulmonary Atresia, the route from the right ventricle to the lungs is blocked.

This situation would prove fatal soon after birth if not for two remnants of the fetal circulatory system that allow some blood from the body tissues to be pumped to the lungs. These are the Patent Ductus Arteriosus (PDA), a small tube that connects the pulmonary artery and aorta and which usually closes soon after birth (see illustrations below), and the Patent Foramen Ovale.

The Patent Ductus Arteriosus allows blood to pass from the aorta to the lungs. However, blood returning from the body into the right atrium and right ventricle would be blocked if not for the presence of the small Patent Foramen Ovale and, in some case, a Ventricular Septal Defect. These allow the mixing of blood between the right and left chambers of the heart and permit the passage of blood from the body into the left ventricle, which pumps it to the lungs by way of the aorta and Patent Ductus Arteriosus.

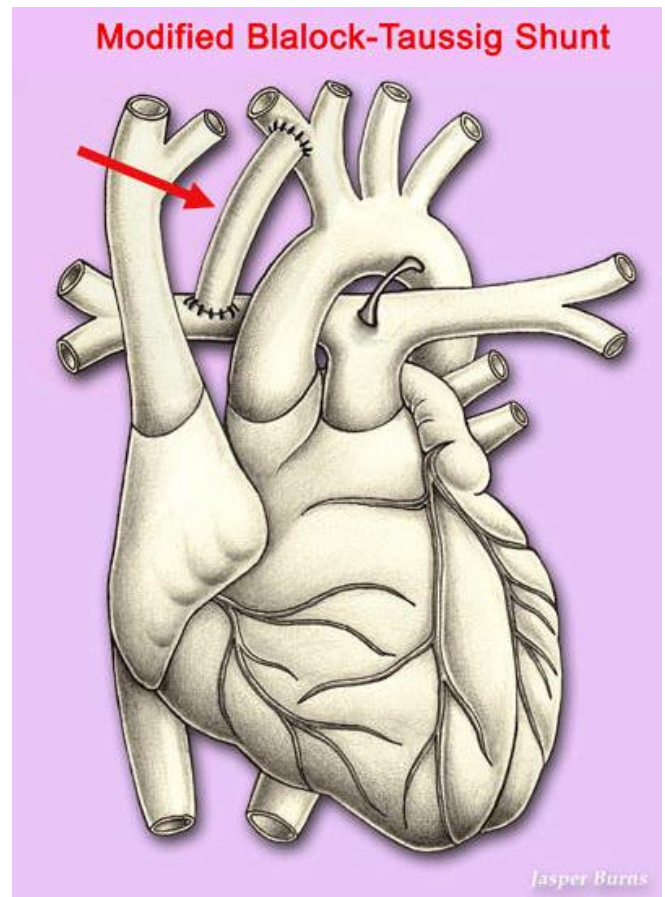
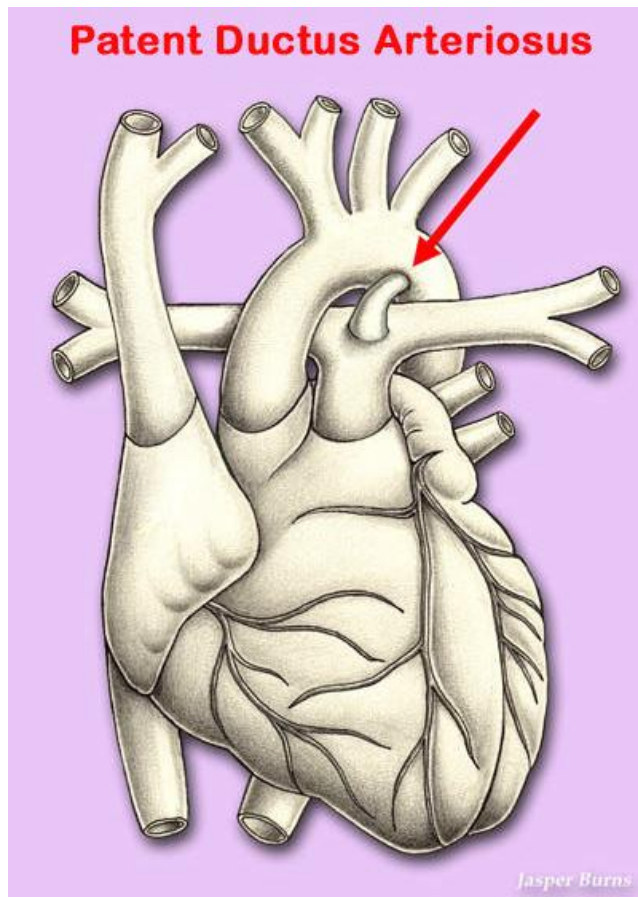
As long as the Patent Ductus Arteriosus, Patent Foramen Ovale, and Ventricular Septal Defect (if present) remain open, the infant can survive, though cyanosis ("blue baby syndrome") may develop because less oxygen than normal is carried to the body tissues. Other symptoms that may develop soon after birth include a heart murmur, rapid or labored breathing, lethargy, and irritability.

In cases where the right ventricle is undersized (hypoplastic), the formation of the coronary arteries, which are on the outer surface of the heart and carry oxygen-rich blood to the heart muscle, may be adversely affected.



Immediately after birth, it is imperative to keep the Patent Ductus Arteriosus open. Medications, such as Prostaglandin E1, may be given for this purpose.

In most cases, a tube, or shunt, made of a synthetic material called Gore-Tex, will be inserted between the pulmonary artery and the aorta or their branches to ensure continued blood flow to the lungs. This is known as a modified Blalock-Taussig Shunt.



### How Is It Treated?

The Blalock-Taussig Shunt procedure is only a temporary measure in the treatment of this disorder. At one to three years of age, a repair operation is usually performed.

In most cases of pulmonary atresia the right ventricle is quite small and cannot function effectively as the pumping chamber that sends blue blood to the lungs. This condition falls into the category of "single ventricle" heart defects, which are discussed elsewhere. The ultimate operation for these defects is the Fontan procedure.

If the right ventricle is of normal or near normal size, then the Pulmonary Atresia Repair operation involves patching the ventricular septal defect (if present) and removing the Gore-Tex shunt that was inserted during the Blalock-Taussig Shunt procedure. Sometimes, it is also necessary to enlarge parts of the pulmonary artery through the insertion of one or more patches. (See illustrations)

In more severe cases, a conduit, or tube with a valve at one end, is inserted between the branches of the pulmonary artery and the right ventricle. In essence, the closed segment of the pulmonary artery is replaced by an artificial but normally functioning pulmonary artery base and pulmonary valve.

Postoperative recovery is variable, depending on the individual's anatomy and the nature of the surgery. The average hospital stay is from one to two weeks.

