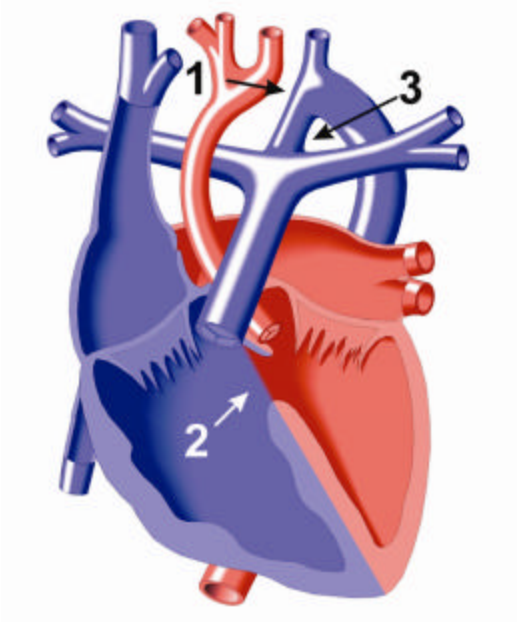
Interrupted Aortic Arch

This rare genetic disorder involves two defects. First, the Aortic Arch does not form a complete tube and is divided, or "interrupted." The aortic arch is the part of the aorta (the major vessel that carries oxygen-rich blood from the heart to the body's tissues) that curves directly above the heart and begins the descent to the lower body. Second, there is a hole, called a Ventricular Septal Defect (VSD), in the muscle wall (septum) that separates the two ventricles, or pumping chambers of the heart.



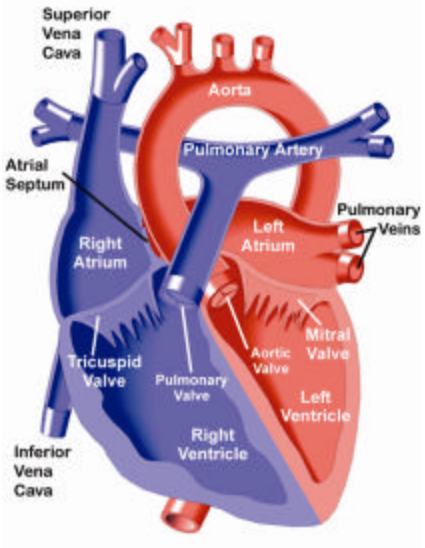
Because the aorta is interrupted and cannot carry blood from the left ventricle to the lower body as in a normal heart, it might seem that the child with this anomaly could not survive. However, some blood does enter the lower part of the aorta because of a small vessel, known as the Patent Ductus Arteriosus (PDA), which connects the lower part of the aorta with the pulmonary artery. (The Patent Ductus Arteriosus is a feature of the fetal circulatory system that usually closes soon after birth.)

The pulmonary artery normally carries oxygen-poor blood to the lungs, so it might seem that blood entering the lower aorta from this vessel (through the PDA) would not carry enough oxygen to the lower body. However, in this case the Ventricular Septal Defect (VSD) allows mixing of oxygen-rich blood from the left ventricle into the right ventricle, which pumps blood into the pulmonary artery.

An infant with this anomaly is usually quite sick immediately after birth.

If steps are not taken to keep the Patent Ductus Arteriosus (PDA) open, no oxygen will make its way to the lower body tissues and the patient will go into shock.

Interrupted aortic arch usually occurs with other cardiac anomalies, such as ventricular septal defects, patent ductus arteriosus, transposition of the great arteries, aortic stenosis, truncus arteriosus, bicuspid aortic valve, and atrioventricular canal defects. The prognosis and treatment for the patient with IAA will be affected by the presence of any associated defects.



Interrupted aortic arch is commonly seen as a result of a genetic chromosomal microdeletion (chromosome 22q11) and often referred to as DiGeorge syndrome. Patients with DiGeorge syndrome can have cranio-facial, immune and electrolyte abnormalities in addition to their congenital heart disease. Patients with interrupted aortic arch are evaluated by a geneticist to determine if they have DiGeorge syndrome.

Top:
1. Interrupted aortic arch.
2. Ventricular septal defect.
3. Patent ductus arteriosus.

Left: Normal Heart