

# Hypertrophic Cardiomyopathy

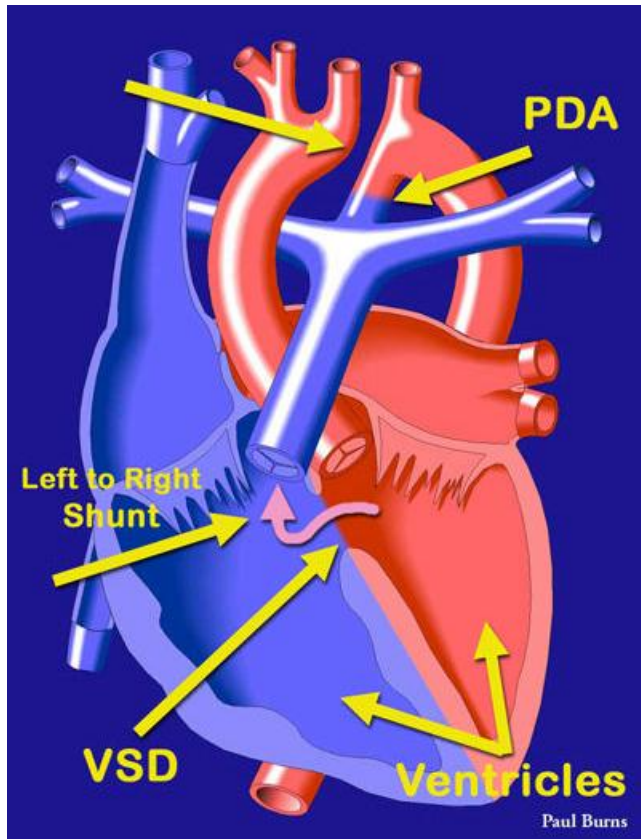


## What Is It?

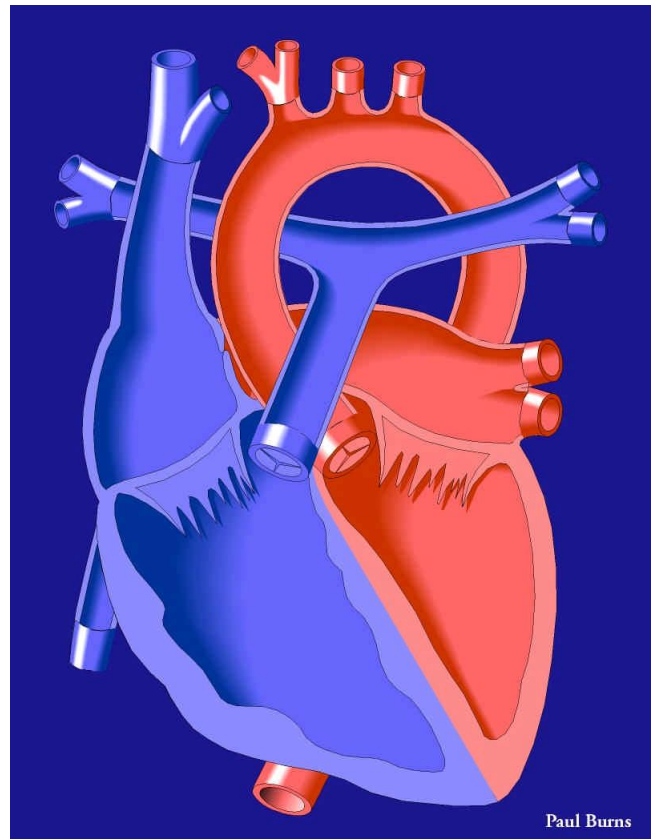
This rare genetic disorder involves two defects. First, the Aortic Arch is divided, or "interrupted" (see upper arrow in diagram). The Aortic arch is the part of the aorta (the major vessel that carries oxygen-rich blood from the heart to the body's tissues) that curves directly above the heart and begins the descent to the lower body. Second, there is a hole, called a Ventricular Septal Defect (VSD in diagram), 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) that 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 much oxygen to the lower body. However, in this case the blood carries more oxygen than usual because of the Ventricular Septal Defect (VSD), which allows the leakage of oxygen-rich blood from the left ventricle into the right ventricle, which pumps blood into the pulmonary artery.



**Interrupted Aortic Arch**



**Normal Heart**

### **What Are Its Effects?**

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

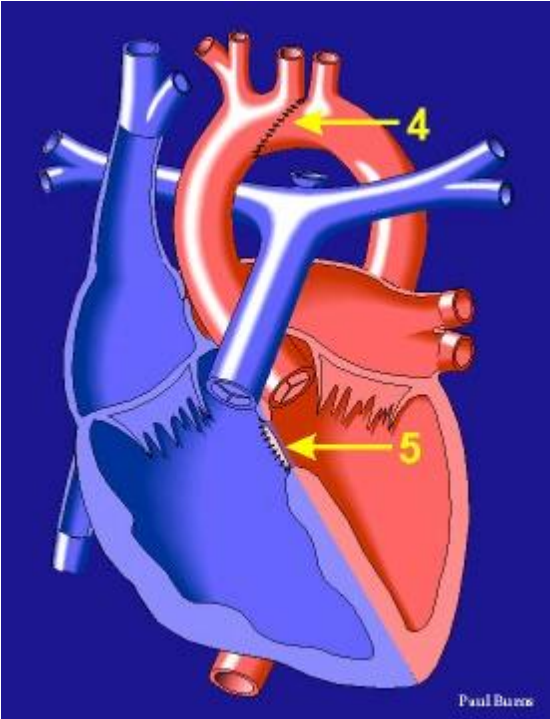
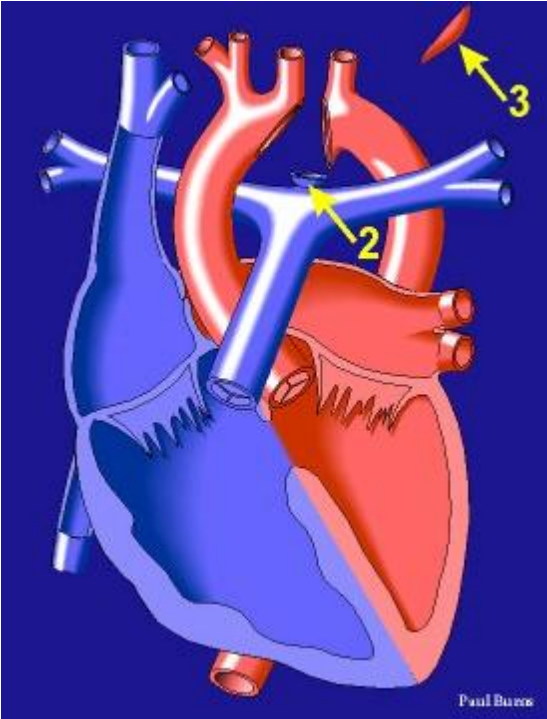
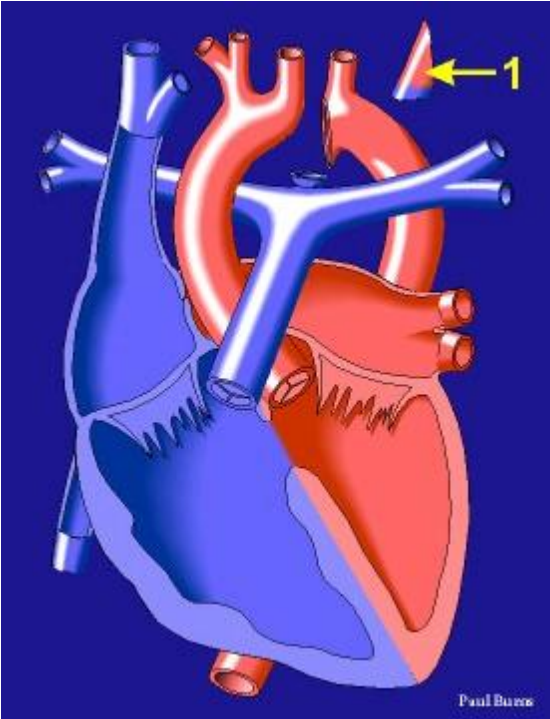
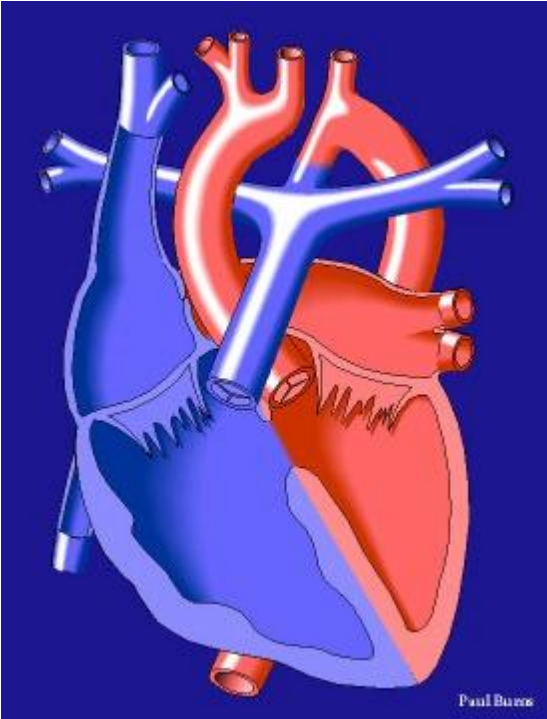
If steps are not taken to keep the Patent Ductus Arteriosus (PDA) open, no oxygen will make its way to the lower body tissues and the patient will go into shock.

### **How Is It Treated?**

As the only source of blood for the lower body after birth in the case of Interrupted Aortic Arch is through the Patent Ductus Arteriosus, the medication Prostaglandin E1 is administered soon after birth to keep that vessel open. It is important to diagnose this defect as soon after birth as possible.

Surgery to correct the defect is straightforward. The Patent Ductus Arteriosus is partially removed (1, 3 in diagram) and its opening from the pulmonary artery closed (2). The lower portion of the aortic arch is connected to the upper portion with the use of sutures (4). Also, the Ventricular Septal Defect (VSD) is patched (5) with a piece of pericardium (the membrane that covers the heart) or with silk or a synthetic material such as Dacron or Teflon.

A hospital stay after corrective surgery for Interrupted Aortic Arch of two weeks or more should be expected.



**Surgical Repair of Interrupted Aortic Arch**