

Interrupted Aortic Arch (IAA)

What is it?

Interrupted aortic arch (IAA) is a rare **congenital**, meaning present at birth, heart defect. IAA is a defect of the aorta, which is the main blood vessel that carries oxygen-rich blood from the heart to the rest of the body. IAA occurs when part of the aortic arch that carries blood to the head and neck is missing. Oxygen-rich blood is not able to reach all the areas of the body. There are three types of IAA based on where the missing part of aorta is:

1. **Type A** is when the missing part occurs after the left subclavian artery. About 30% – 40% of infants with IAA have type A.
2. **Type B** is when the missing part occurs between the left carotid artery and the left subclavian artery. This is the most common form with about 53% of infants with IAA having type B.
3. **Type C** is when the missing part occurs between the innominate artery and the left carotid artery. Only about 4% of babies with IAA have this form.

IAA is considered a **critical congenital heart defect**, meaning it can cause serious health problems or even death if left untreated. A child with IAA also might have an opening between the two lower chambers of the heart called a **ventricular septal defect (VSD)**. Blood may skip the missing part of aortic arch if the ductus arteriosus stays open. The **ductus arteriosus** is a blood vessel present in a developing baby that lets blood skip the baby's fluid-filled lungs. Normally, the ductus arteriosus closes shortly after birth. In babies with IAA, though, doctors might want to keep it open. If the ductus arteriosus is kept open, it is called a **patent ductus arteriosus (PDA)**. This helps the blood flow around the missing part of aortic arch. Children with IAA often have a chromosomal abnormality called DiGeorge syndrome. Your child's doctor should discuss this possibility with you.

How common is it?

IAA is extremely rare. About 2 per 100,000 babies are born with IAA in the United States.

What causes it?

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



How is it diagnosed?

IAA can be diagnosed during pregnancy or after. Screenings are done during pregnancy to check for birth defects. After birth a baby may have rapid breathing, a pounding heart rate, poor feeding, and blue skin color and lips (called **cyanosis**). If a baby has symptoms of a heart defect, then a doctor might do a diagnostic test. A screening test called a **pulse oximetry screen** is done shortly after birth to check for critical congenital heart defects, too. 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 the screening, then the doctor should perform a diagnostic test called an **echocardiogram** to check for defects in the heart.

How is it treated?

Medication may be given to keep the ductus arteriosus open to allow blood flow until surgery. The goal of surgery is to reconnect the aortic arch to create one unbroken tube. The VSD will be closed, and the PDA will be tied off. Surgery is usually urgent and will be done within the first few days of life. 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

National Heart, Lung, and Blood Institute

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

