Interrupted Aortic Arch (IAA)

Patient Information Series – What you should know, what you should ask.

What is an interrupted Aortic Arch (IAA)?

Interrupted Aortic Arch (IAA) is a rare birth defect of the heart. Our hearts are comprised of four chambers, two upper chambers, the right atrium and left atrium, and two lower chambers, the right and left ventricles. The ventricles are the heart's pumping chambers. Blood from the heart flows into the Great Arteries, the Aorta and the Pulmonary Artery. The left ventricle pumps blood rich in oxygen through the Aorta to the body and brain, while the right ventricle pumps oxygen-poor blood out through the Pulmonary Artery to the lungs. The Aorta runs upward toward the head then turns downward toward the body, to form the Aortic Arch. The part of the aorta below the arch is called the Descending Aorta, it supplies the lower body and legs.

In most people, three blood vessels branch out of the aortic arch to supply the head and brain and the arms with oxygen-rich blood. In Interrupted Aortic Arch (IAA), the aorta is divided somewhere along the arch into separate blood vessels. Three variations of this problem exist, according to the place of the interruption. The upper portion of the aorta still supplies the head and arms, but the flow to the lower body and legs is supplied by blood flowing from the pulmonary artery through a special fetal vessel and then into the Descending Aorta.

Many fetuses with IAA also have another heart defect, called a ventricular septal defect or VSD. The right and left ventricles of the heart are separated by a wall of muscle called the ventricular septum; a hole in this wall is called a VSD. VSD's are quite common, in fact they are the most common form of heart defect at birth. A VSD allows blood to cross between the two ventricles of the heart, causing oxygen-rich blood to mix with the oxygen-poor blood that is pumped by the right ventricle to the lungs. Other congenital heart defects, as well as other malformations, may also occur together with IAA.

How does an IAA happen?

During the embryonic period (the first weeks of life in the womb), as the complex anatomy of the heart and blood vessels is developing, IAA seems to occur from a failure of earlier small arches to grow and merge properly. There seems to be a genetic component to IAA in some cases. The most common is DiGeorge syndrome, which is caused by a deletion of a small portion of chromosome, one of the 23 pairs of chromosomes that store our genetic information. In many cases, however, no genetic cause is found.

Should I have more tests done?

Your caregiver may refer you for genetic counseling and genetic testing. This may include tests like amniocentesis or chorionic villus sampling (CVS) to check for chromosomal abnormalities, and other genetic diagnostic tests such as chromosomal microarray testing or whole exome sequencing. Such testing can provide essential information regarding your individual case. Your caregivers and the genetic counselors can answer questions about what to expect from testing and understanding the results.

In addition, your caregiver may refer you to specialists in fetal heart problems, such as a maternal-fetal medicine specialist and/or a pediatric cardiologist. They can monitor your baby's progress with ultrasound scans focused on the fetal heart and the blood vessels surrounding it. Those specialized ultrasounds are called fetal echocardiography. You may also meet with specialists in pediatric heart surgery before delivery, to consult with you regarding the best way to manage your baby's birth and care in the newborn nursery. Each baby with IAA is unique. Your caregivers will thoroughly examine your baby's heart and other organs to get the most complete picture of his/her anatomy, and to consult with you to work out a care plan that best suits your individual case.

What are the things to watch for during my pregnancy?

Your caregiver will probably order serial ultrasound scans, to watch carefully to assess whether the IAA is affecting your baby's growth and well-being. You may be asked to monitor your baby's movements throughout the day. Your caregivers will advise you as the pregnancy progresses.

What does it mean for my baby after it is born?

While inside the womb, your baby has a small vessel with a long name, the ductus arteriosus, that allows blood flow from the pulmonary artery to be directed to the descending aorta so the lower part of the body can receive blood. After delivery, the ductus arteriosus normally closes within a few days and all the blood from the pulmonary artery flows to the lungs, to be supplied with oxygen and returned to the heart. In IAA, it is important to keep the ductus arteriosus open, to maintain a supply of blood to the portion of the aorta that brings blood to the lower part of the body. Your baby will be given a medication called prostaglandins to keep this vessel open until heart surgery can be performed. IAA always needs to be repaired by heart surgery. The goal of surgery is to create a connection between the two segments of the aorta and to close the hole in the heart (the VSD), if present.

Will it happen again?

This depends whether or not a genetic cause was found to explain the IAA, and what other problems were present. All these factors affect the likelihood of having another baby with IAA. You can discuss your individual risks with the genetic counselor. Your caregiver will likely order early targeted fetal scanning to rule out fetal heart defects in future pregnancies.

What other questions should I ask?

You might ask your caregiver these questions, or any other question regarding your pregnancy:

- What type of IAA does my baby have?
- How often will I have ultrasound examinations done?
- Should I have genetic counseling? What genetic testing should we consider?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet the team of doctors that will be assisting my baby when it is born, in advance of my delivery?

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