Coarctation of the Aorta (CoA)

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

What is a Coarctation of the Aorta (CoA)?

Coarctation of the Aorta is a congenital heart defect (CHD). Our hearts are comprised of four chambers, two upper chambers called atria, 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 blood to the lower body and legs.

During fetal life, a small vessel called the ductus arteriosus allows blood flow from the pulmonary artery to be directed to the descending aorta. In coarctation of the aorta, a narrowing forms along the vessel that constricts (limits) blood flow. This narrower portion of the aorta can occur anywhere, but most often it is found just past the arch, in the descending aorta. The type of coarctation is defined by its location in relation to the ductus arteriosus. The narrowing limits the blood flow to the lower part of the body and the heart has to pump harder to get past this "bottleneck". In addition, other types of CHD may be diagnosed along with CoA. The most common is a ventricular septal defect, or VSD. A VSD is an opening in the septum, the wall of muscle that separates the two ventricles of the heart. This opening allows blood from the two sides of the heart to mix.

How does a CoA happen?

There are different theories as to why CoA happens. It seems that during the embryonic period (the first weeks of life in the womb), as the complex anatomy of the heart and blood vessels develops, CoA seems to occur from abnormal growth in some vessels that alters blood flow in the aortic arch. There may be a genetic link to CoA, but in most cases no specific cause is found. Some cases of CoA may be associated with a genetic syndrome called Turner Syndrome, which is caused by the absence of one set of genes from the short arm of one of the X-chromosomes.

Should I have more tests done?

Your caregiver may refer you for genetic counseling and genetic testing. This can provide essential information regarding your individual case. Your caregiver may refer you to specialists in fetal cardiac disease, such as an obstetrician specialising in fetal imaging, a maternal-fetal medicine specialist and/or a pediatric cardiologist. They can monitor your baby's

progress with fetal echocardiography: ultrasound scans focused on the fetal heart and cardiovascular system. 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 CoA is unique. Your caregivers will thoroughly examine your fetus'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 baby and your family.

What are the things to watch for during the pregnancy?

Your caregiver will probably order serial ultrasound scans, to watch carefully to assess whether the CoA is affecting your baby's growth and well-being, and to monitor for any signs of distress. 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?

This depends on many factors. Treatment will be individualised according to your baby's needs. The severity of the coactation (how narrow the artery is at the area of the defect) and any symptoms that develop, as well as the presence of any other defects, will guide your caregivers in developing a treatment plan. In some cases of CoA there are no symptoms and no intervention is necessary. If symptoms are severe, there are ways to repair the affected segment. One approach is cardiac catheterisation. In this procedure a narrow tube is threaded into the narrow portion of the aorta and a tiny balloon is inflated there to stretch the vessel, to make it wider. Then a small device called a stent is placed to keep the segment unrestricted while the balloon is removed. In other cases, an open heart surgery may be necessary, in which the narrow segment will be repaired or replaced. If other defects are present, these are often repaired at this time if possible.

Will it happen again?

This depends whether or not a genetic cause was found to explain the CoA, and what other anatomic findings were present. All these factors affect the likelihood of having another baby with CoA. Your caregiver will likely order early targeted fetal scanning to rule out fetal heart defects in subsequent pregnancies.

What other questions should I ask?

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

- What degree of coarctation does my fetus 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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