Right Aortic Arch (RAA)

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

What is a Right Aortic Arch (RAA)?

Right aortic arch (RAA) is an anatomic variant of the Aorta, the great blood vessel that carries blood from the heart toward the body and brain. In most people, the aorta forms an arch to the left of the trachea (the windpipe). In RAA, the aortic arch is on the right of the trachea.

How does a RAA happen?

Isolated RAA occurs in about 1 in 1000 babies. The exact cause is not known. Almost 20% of the babies with a RAA will have other problems, most often with their heart. One in 10 babies with RAA has an alteration in their chromosomes. Chromosomes are where most of our genetic information is stored. The chromosomal abnormality most commonly associated with RAA is Di George syndrome, where a small fragment of chromosome 22q11.2 is missing. This is called a microdeletion.

Should I have more tests done?

Many women will choose to have more tests and/or consultations performed, in order to learn more about the condition of their fetus. For example, you might consult with a Geneticist, a specialist in genetic diseases and congenital defects. You might undergo genetic tests such as amniocentesis (where a thin needle in used to take a little amniotic fluid from the womb) or CVS to look for chromosomal abnormalities, or chromosomal microarray (CMA, or "chip") or exome sequencing, to investigate your fetus's chromosomal make-up further. You might be referred to fetal echocardiography, a specialised ultrasound of the fetal heart during the pregnancy, to determine whether the RAA is isolated (the only heart defect) or associated with other abnormalities. You might also request a detailed fetal scan by a fetal medicine specialist.

What are the things to watch for during the pregnancy?

Babies with isolated RAA do not usually develop any problem during the pregnancy, although an ultrasound examination from a fetal medicine specialist is recommended. The ultrasound will help identify if the RAA is associated with a different course of another arterial vessel, which is called aberrant left subclavian artery (ALSA), departing from the RAA. This combination will create a vascular ring that encircles the trachea (windpipe) and esophagus (the tube connecting the mouth and stomach).

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

The majority of babies with isolated RAA will not experience problems because of the RAA. Babies with a vascular ring can have symptoms that can be quiet varied. Rarely they may be noted in the newborn period but more often are recognised later. Symptoms are usually of airway obstruction such as noisy breathing or breathing difficulties during feeding. They may have frequent upper respiratory tract infection and such symptoms as wheezing, coughing, feeding problems and vomiting.

Will it happen again?

When no other genetic reason is found to explain the RAA, the risk of this happening again is extremely rare. If a genetic cause was identified, then the risk of recurrence depends on that cause. A consultation with a geneticist may be helpful to shed light on this.

What other questions should I ask?

- Does the baby's heart look normal?
- How often will I have ultrasound examinations done?
- Is a vascular ring present?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet in advance with the team of doctors that will be looking after my baby when it is born?

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