Aberrant Right Subclavian Artery

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

What is an Aberrant Right Subclavian Artery (ARSA)?

ARSA is a relatively common variation of the vessels of the Aortic Arch. The Aorta is one of the great blood vessels that deliver blood from the heart: it supplies oxygen-rich blood to the head and body. As the aorta exits the heart it runs upward towards the head, then forms the Aortic Arch as it turns downward toward the chest and abdomen. In most people, three vessels branch out of the aortic arch going towards the head, the neck and the arms. Usually, the right subclavian artery arises from one of them. In about 1% of people, the aortic arch has four branches, and the right subclavian artery arises directly from the aorta as it turns down toward the body. It goes behind the trachea (windpipe) and the esophagus (tube that connects the mouth to the stomach), and then turns towards the right shoulder and arm.

How does an ARSA 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, certain vessels that begin as pairs, merge, while others disappear. In ARSA, the usual pattern of development is slightly changed, preventing the typical formation of the three blood vessels. This leads to there being four branches rather than the normal three arising from the left aortic arch, one being the ARSA.

How are chromosomes relevant to ARSA?

Chromosomes are where most of our genetic information is kept. We usually have 46 of them matched in pairs: 23 come from one parent and the other 23 come from the other parent. For example, people with Down syndrome have an extra chromosome number 21. People with DiGeorge syndrome have a change in the information in a fragment of the chromosomes number 22. Both of those conditions are seen more frequently in babies with an ARSA. If such a change in information within the chromosomes occurs, it would be the cause for the ARSA. However in most cases of ARSA, no chromosomal defect is identified and no genetic cause of the ARSA is determined.

Should I have more tests done?

When ARSA is identified during your scan, your caregiver can refer you for a targeted or expanded scan of the baby's anatomy, particularly the heart and blood vessels. If the ARSA is isolated and no other anatomic defects or markers are identified, you will probably not need any further testing. However if other worrying findings are seen, you may be referred for genetic counseling and genetic testing, such as amniocentesis or chorionic villus sampling (CVS) with chromosomal microarray or whole exome sequencing, can help to rule out possible chromosomal conditions in your baby, such as Down syndrome and DiGeorge syndrome. Your individual risk for having a fetus with Down syndrome or other genetic condition can be derived from a battery of tests. The management approach in your case will depend on several factors, including your age, the week of pregnancy when ARSA was diagnosed, and the results of any other screening tests for Down syndrome and other genetic testing you have had.

What are the things to watch for during the pregnancy?

If the ARSA is isolated, the rest of your pregnancy care will most likely continue as planned. Your caregiver can advise you regarding further antenatal visits and ultrasound scans.

If there were anatomic and/or genetic findings in addition to the ARSA, your caregiver may refer you to the appropriate specialists, for example a genetic counselor and a maternal-fetal medicine specialist or pediatric cardiologist, for further follow-up and to develop a management plan with you.

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

Most people with ARSA have no symptoms, and are most likely not aware that they have this variant. By itself, ARSA is not a very worrying finding. If the ARSA is isolated, your caregiver or your baby's pediatrician can advise you what to watch for in your newborn. In some cases the extra blood vessel might press on the trachea (windpipe) or esophagus (food path from the mouth to the stomach) and create problems with breathing or swallowing. If the ARSA is found to be associated with other anatomic or chromosomal findings, the specialists consulting with you can discuss your baby's individual prognosis in light of these findings. Every baby is unique. You and your doctors and other caregivers can work together to develop the best management plan for you, your baby and your family.

Will it happen again?

There seems to be a genetic component in ARSA. Genetic counseling can help to determine your individual risk of recurrence, which depends on whether a cause for the ARSA was found in the first place as well as your age and other factors.

What other questions should I ask?

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

- Is this an isolated ARSA?
- Were any other anatomic defects or markers seen during the scan?
- Should I have genetic counseling or genetic testing?
- How often will I have ultrasound examinations done?
- Where should I deliver my baby?

Last updated June 2024