

Isolated right aortic arch (with left arterial duct)

This leaflet explains about prenatal diagnosis of right aortic arch with left arterial duct.

What is a right aortic arch and why does my baby have it?

The aorta is the body's main artery that leaves the heart and delivers blood, carrying oxygen, around the body. The aorta forms an arch in the chest and usually travels inside the chest on the left side. In some people the arch of the aorta turns to the right and travels down the chest on the right side. This is known as a right aortic arch. The arterial duct is a blood vessel which closes off after birth, forming a tight string, and in most cases passes to the left of the trachea (windpipe).

A right aortic arch is thought to occur in around one in 1,000 people, but is likely to be higher as identification during prenatal screening is increasing. A right aortic arch can be an isolated finding and some babies may have additional heart defects that are present before birth. Your cardiologist (specialist heart doctor) will have explained any other heart issues with you.

There are usually three blood vessels that arise from the aortic arch. In many fetuses with a right aortic arch, there are four blood vessels. This fourth vessel is called an aberrant left subclavian artery and it supplies the left arm with blood. The additional implications of this are detailed below.

Why is this condition important?

A right sided aortic arch with a left sided arterial duct form a ring around the trachea and oesophagus (food pipe). Due to the blood pulsating through the aortic arch, it may squash these pipes. This may show itself as noisy breathing, recurrent chest infections, slow weight gain or difficulty swallowing and/or feeding in your baby. These symptoms may develop slowly, and not necessarily in the first months after birth. If any of these symptoms develop then you should discuss this with your doctor or our paediatric clinical nurse specialists.

A right aortic arch can be associated with abnormalities in other organs in about 15 out of 100 cases. If you have not had a detailed scan of your baby (anomaly scan), then we would recommend that this is done. We will make a referral to a fetal medicine specialist.

A right aortic arch can occasionally be associated with chromosomal abnormalities such as microdeletion of chromosome 22q11.2 and Trisomy 21. Your doctor will be able to advise you about this, taking into account the results of other tests which you may have had.

Does my baby need any tests to confirm the diagnosis?

In the vast majority of babies, we can be certain of the diagnosis from the ultrasound scans performed in pregnancy. We may offer a further ultrasound appointment in your third trimester (around week 30) to check for any minor heart anomalies which may not have been apparent at the 20 week scan.

A fetal cardiac MRI (magnetic resonance imaging) may be offered to you as an additional investigation in the third trimester. This may be offered if further information is required about the anatomy of the aortic arch because in some babies there are two aortic arches (double aortic arch).

When you see the fetal medicine specialist you can choose to have a test to look for a chromosomal abnormality. There are two ways this can be done, and this depends on how far you are into your pregnancy. The tests are called amniocentesis and chorionic villus sampling (CVS).

Should my baby see a doctor or nurse after birth?

We would recommend that your baby is reviewed by a cardiologist after birth. Someone from our team will contact you to arrange this appointment either at Evelina London Children's Hospital or at one of our outreach clinics. If your baby is well this does not need to be performed urgently.

Does my baby need any tests after birth?

Yes. We recommend that your baby has a heart scan after birth – this is the same type of ultrasound scan you have during pregnancy to look at the baby. It is not invasive or harmful to the baby.

Depending on the findings, your baby may also need some of the following tests:

- bronchoscopy – this test is to look inside the trachea with a small camera to see if it is being squashed
- computerised tomography (CT) or MRI – these tests show in detail how the blood vessels and windpipe are arranged in the chest.
- barium swallow – this is an x-ray that looks at how food moves down the food pipe to the stomach.

If these are normal then follow-up may not be required.

What are the signs and symptoms?

Most babies diagnosed in the womb with a right aortic arch will be free of symptoms after birth and during childhood. However, due to the different position of the aortic arch in the chest, it may cause some symptoms in your baby after birth. This is more likely if there is also an aberrant left subclavian artery. As these structures form a ring around the trachea and oesophagus, they can squash these structures and cause:

- noisy breathing
- frequent chest infections
- poor feeding
- poor weight gain
- difficulty swallowing – especially lumps/solids

These symptoms may not be evident immediately after birth and may develop over the course of years. If you are concerned that your child has any of these symptoms, your GP should refer your child to a specialist paediatrician for investigation.

In many cases, children have no symptoms, but when we directly look at their trachea it is very squashed. Therefore, even if your baby has no symptoms we would discuss the option of bronchoscopy and/or CT scan and refer your baby on to our respiratory/airway team for further assessment.

What treatments are available?

The need for treatment depends on the bronchoscopy findings. If there is evidence that the trachea is significantly squashed, we would recommend an operation to release the ring or move the vessels away from other structures. The outcome of these procedures, if performed in a timely manner, is good.

Useful sources of information

w: www.echo-evelina.org.uk give support for parents with children with heart abnormalities.

w: www.arc-uk.org provides information about making choices in pregnancy.

Contact us

Before birth: If you have any questions or concerns about a right aortic arch, please contact our fetal cardiology clinical nurse specialists, **t:** 020 7188 2307, Monday to Friday, 9am to 5pm.

After birth: If you have any questions or concerns about symptoms your baby is showing, please contact our paediatric cardiology clinical nurse specialists, **t:** 020 7188 4546, Monday to Friday, 9am to 5pm.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.evelinalondon.nhs.uk/leaflets.

Evelina London Medicines Helpline

If you have any questions or concerns about your child's medicines, please speak to the staff caring for them or contact our helpline.

t: 020 7188 3003 10am to 5pm, Monday to Friday **e:** letstalkmedicines@gstt.nhs.uk

Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.

t: 020 7188 8801 (PALS) **e:** pals@gstt.nhs.uk

t: 020 7188 3514 (complaints) **e:** complaints2@gstt.nhs.uk

Language Support Services

If you need an interpreter or information about your care in a different language or format, please get in touch:

t: 020 7188 8815 **e:** languagesupport@gstt.nhs.uk

NHS Choices

Provides online information and guidance on all aspects of health and healthcare, to help you make choices about your health.

w: www.nhs.uk

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