



# Isolated right aortic arch (with left arterial duct)

This leaflet explains how we diagnose and treat a right aortic arch with left arterial duct.

# What is a right aortic arch (with left arterial duct)?

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 windpipe (trachea). This is known as a left arterial duct.

About 1 in 1,000 people are thought to have a right aortic arch, but this number is likely to be higher as more babies are being diagnosed during prenatal screening (before the baby is born). A right aortic arch can be an isolated finding, which means that no other conditions occur with it. However, some babies who are diagnosed with a right aortic arch will also have other heart conditions that are present before they are born. Your specialist heart doctor (cardiologist) will have explained any other heart issues with you.

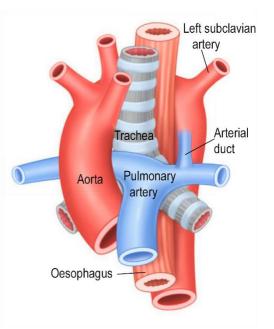
There are usually 3 blood vessels that rise from the aortic arch. However, in many babies with a right aortic arch, there are 4. The fourth vessel is called an aberrant left subclavian artery and it supplies the left arm with blood. The picture shows a right aortic arch with a left duct and an aberrant left subclavian artery.

# Symptoms of a right-sided aortic arch (with a left-sided arterial duct)

A right-sided aortic arch with a left-sided arterial duct forms a ring around the windpipe (trachea) and food pipe (oesophagus). The blood pulsating through the aortic arch, might squash the windpipe and food pipe.

If this happens, symptoms can include:

- noisy breathing
- frequent chest infections
- slow weight gain
- difficulty swallowing and/or feeding in your baby





These symptoms might develop slowly, over a course of years, and not necessarily in the first months after birth.

If your child has any of these symptoms, you should speak to your child's doctor or our clinical nurse specialists (cardiac outreach nurses).

In many cases, children have no symptoms, but when we directly look at their windpipe it is very squashed. Therefore, even if your baby has no symptoms we would discuss the option of a bronchoscopy or CT scan for further assessment.

#### Other conditions associated with a right aortic arch

About 15 out of 100 babies who have been diagnosed with a right aortic arch also have conditions which affect their other organs. If your child has not had a detailed scan (anomaly scan), we would recommend that they have one. We will make a referral to a fetal medicine specialist if we think this is needed.

Babies who have been diagnosed with a right aortic arch might also have other chromosomal conditions such as DiGeorge syndrome (22q11 deletion) and Down's syndrome (Trisomy 21). Your child's doctor will be able to speak to you about this, and the results of other tests which your child has had.

# Confirming your baby's diagnosis

In the majority of babies, we can confirm the diagnosis based on the ultrasound scans done during pregnancy. We might offer another ultrasound appointment in your third trimester (around week 30) to check for any minor heart defects which may not have been seen on the 20-week scan.

A fetal cardiac MRI (magnetic resonance imaging) scan might be offered as an additional test in the third trimester. This is offered if more information is needed about the aortic arch, because in some babies there are 2 aortic arches (double aortic arch).

When you see the fetal medicine specialist, you can choose to have a test to see if your baby might have a chromosomal condition. There are 2 ways this can be done, it will depend on how far you are into your pregnancy. The tests are called an amniocentesis, and chorionic villus sampling (CVS). The fetal medicine team will be able to support you through this decision.

# After your baby is born

We would recommend that your baby is seen by a specialist heart doctor (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, they won't need to be seen urgently.

#### Tests needed after birth

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 your baby.

Depending on the results, your baby might also need:

- bronchoscopy to look at the inside of the windpipe with a small camera to see if it is being squashed
- CT scan to show how the blood vessels and windpipe are arranged in the chest
- barium swallow an X-ray that looks at how food moves down the food pipe to the stomach

If the results of these tests are normal then a follow-up might not be needed.

#### **Treatment**

The need for treatment depends on the results from the bronchoscopy. If we can see that the windpipe is very squashed, we would recommend an operation to release the ring, or move the vessels away from other structures. The results of this surgery are good if it is done early enough.

#### **Useful information**

**ECHO**, web https://echo-uk.org/ gives support to parents with children with heart conditions. **arc**, web 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, **phone** 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, phone 020 7188 4546, Monday to Friday, 9am to 5pm.

For more information on conditions, procedures, treatments and services offered at our hospitals, please visit web 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, **phone** 020 7188 3003, Monday to Friday, 10am to 5pm **email** letstalkmedicines@gstt.nhs.uk

#### Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS), phone 020 7188 8801 email pals@gstt.nhs.uk. To make a complaint contact the resolution department phone 020 7188 3514 email complaints2@gstt.nhs.uk

#### Language and accessible support services

If you need an interpreter or information about your care in a different language or format, please contact the department your appointment is with.



Leaflet number: 4556/VER2 Date published: August 2023 Review date: August 2026 © 2023 Guy's and St Thomas' NHS Foundation Trust A list of sources is available on request