Right aortic arch
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This leaflet provides information for parents of children with a right aortic arch. The leaflet does not replace the need for personal advice from a qualified healthcare professional. Please ask your doctor or clinical nurse specialist if you have any questions.
What is a right aortic arch?

A right aortic arch occurs when the aorta, the large vessel that takes oxygenated (red) blood from the left side of the heart to the body, branches off to the right rather than the left of the trachea (windpipe).

Some babies with a right aortic arch may have other heart defects. If this is the case, your cardiologist (specialist heart doctor) will explain this to you.

Your baby may have symptoms of this condition after birth (Please refer to the Symptoms section on page 8).

Normal fetal heart (before birth)
Two of the most common types of right aortic arch

1. Right aortic arch with aberrant left subclavian artery

The features of a right aortic arch with aberrant left subclavian artery are:

- The aorta, the large body vessel, branches to the right rather than the left.
- The ‘left subclavian artery’ – the large vessel that supplies oxygenated blood to the left arm, is positioned abnormally but flow to the arm is not usually affected.
• The way the aorta and the left subclavian artery are positioned creates a vascular ring around the trachea and oesophagus (feeding tube). Vascular refers to the body’s network of blood vessels. The condition is called ‘vascular ring’ because of the ‘ring’ shape formed by the right aortic arch, the left subclavian artery and the ‘ductus arteriosus’.

The ductus arteriosus is a blood vessel that connects the pulmonary artery (main vessel supplying the blood to the lungs) to the aorta (main vessel supplying the blood to the body). This connection is present in all babies in the womb, but should close shortly after birth.

• A baby will not usually have any symptoms of this condition at birth.

The structure of a vascular ring
2. Right aortic arch with mirror image branching pattern

The features of a right aortic arch with mirror image branching pattern

- The aorta, the large body vessel, branches to the right rather than the left.
- All the head and neck vessels are connected in the same position and provide blood to the correct part of the upper body.
- The pattern does not usually form a ‘vascular ring’.
- A baby will not usually have any symptoms.
How is a right aortic arch diagnosed?

A right aortic arch is often identified during a 20-week ultrasound pregnancy scan. If that happens, parents are referred to a cardiologist who will confirm the diagnosis after an echocardiogram is carried out. An echocardiogram (or echo) is a scan which uses sound waves to build up a moving picture of the heart in a similar way to an ultrasound scan.

Once born, a baby is referred for another echocardiogram at Royal Brompton Hospital or to one of our outreach cardiology clinics at a local hospital.

If the symptoms of a right aortic arch show after birth (whether or not the condition has been diagnosed during pregnancy) a baby will be seen by a cardiologist at Royal Brompton Hospital.

Parents will also meet with one of our clinical nurse specialists (CNSs) in children's cardiac care who explain more about a baby’s condition and provide support.

On rare occasions, a right aortic arch is linked to a genetic defect such as a microdeletion of chromosome 22q11 (a missing piece of chromosome) which is present at birth. If that is the case, a cardiologist will discuss the finding with parents and the options for further testing.

How common is a right aortic arch?

A right aortic arch is thought to happen in approximately one in 1,000 people.

Does a right aortic arch affect the way the heart works?

A right aortic arch itself will not affect the function of the heart muscle, or the blood flow in and out of the heart. The heart will work normally.
Symptoms of a right aortic arch

Most babies diagnosed antenatally (before birth) with a right aortic arch will not have any symptoms after birth or during childhood.

However, some babies will develop symptoms after birth. This is more likely if a baby also has an aberrant left subclavian artery.

Symptoms are more likely with an aberrant left subclavian artery because the ring that forms around the breathing (trachea) and feeding (oesophagus) tubes can compress (squash) them.

This may cause:

- noisy breathing
- difficulty swallowing – especially solid food
- additional effort of breathing when feeding. If this happens, a baby’s ribs and the centre of his/her chest will be pulled in (known as recession)
- frequent chest infections
- poor feeding
- poor weight gain

Symptoms rarely happen after birth and are more likely within the first year of life and when your child begins eating solid foods (weaning).

If you have any concerns or recognise any of these symptoms while at home, please contact your clinical nurse specialist for advice. If there are immediate concerns with your baby’s breathing or colour, please go to your local emergency department (A&E).
Please remember that Royal Brompton Hospital does not have an accident and emergency (A&E) department.

Follow-up appointments

We will arrange for your child to have echocardiogram/s and check on his/her progress at follow-up appointments with a cardiologist throughout childhood.

In many cases, children have no symptoms. If the cardiologist feels that it is necessary, we will carry out tests to show whether your child’s breathing and feeding tubes are being compressed.

Will my baby need surgery?

If the breathing and feeding tubes are compressed, surgery may be recommended to ‘release’ the vascular ring so the aorta no longer circles round or puts pressure on the trachea or oesophagus.

This is usually carried out by making an incision (cut) in the side of the chest (a thoracotomy) rather than open heart surgery, and needs a short stay in hospital. The surgeon will explain the surgery. In most cases, the risks of surgery are low, and babies recover well.

Everyday activities

Children with any type of right aortic arch can lead normal lives. Your child can take part in everyday activities, including sports at school.
Information and support

A cardiologist and clinical nurse specialist (CNS) will explain more about your child’s right aortic arch.

Please contact your CNS if you have any worries or questions when you are at home.

Further family support can be offered by The Brompton Fountain Charity.

The charity provides vital support for children who are being cared for at Royal Brompton and Harefield hospitals. It aims to improve the quality of life for our young patients and their families. The charity works closely with paediatric teams to provide activities, medical equipment and services that are not normally supplied by the NHS.

Website:  www.thebromptonfountain.org.uk

British Heart Foundation

The British Heart Foundation offers support to families with children who have a cardiac condition.

Helpline:  0300 330 3311 (Monday to Friday, 9am to 5pm)
Website:  www.bhf.org.uk

Useful contacts

If you need more information, please contact a member of the CNS team.

Royal Brompton clinical nurse specialists team  0330 128 7727
(Monday to Friday, 9am to 5pm)
If you have concerns about any aspect of the service you have received in hospital and feel unable to talk to those people responsible for your care, call the Patient Advice and Liaison Service (PALS) on:

- Royal Brompton Hospital – 020 7349 7715
- Harefield Hospital – 01895 826 572

You can also email pals@rbht.nhs.uk. This is a confidential service.
Royal Brompton Hospital
Sydney Street
London
SW3 6NP
Tel: 0330 12 88121

Harefield Hospital
Hill End Road
Harefield
Middlesex
UB9 6JH
Tel: 0330 12 88121

Website: www.rbht.nhs.uk

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