

# Double aortic arch (DAA)

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

**This leaflet is to help you understand what Double aortic arch is, what tests you need and the implication of being diagnosed with Double aortic arch for you, your baby and your family.**

## **What is a Double aortic arch (DAA)?**

Double aortic arch is an anatomic variation of the aorta, a large blood vessel that carries blood from heart to the rest of the body that has an arch-like configuration at its top, it is a congenital variation which means it is present since before birth. Normally a newborn will have only one aortic arch on left side of trachea and in case of double aortic arch, an additional arch on the opposite side persists in addition to the normal arch.

The reasons for this are unknown. This leads to two aortic arches, one on either side of trachea and esophagus of the baby forming a circle of vessels. If this circle is too tight, it can cause symptoms in your baby due to the compression of trachea or esophagus.

## **How does a DAA happen?**

The exact cause of this entity is not known. About 1 in 10,000 babies are born with Double Aortic Arch. Most of the time it is the only anomaly in the baby and rarely it can be associated with other congenital cardiac defects. There are several variations of the double aortic arch which can arise in fetal life. Rarely it can be associated with a genetic cause such as a deletion on one chromosome, called “22q11 deletion”.

## **Do I need more tests done?**

Once a diagnosis of Double aortic arch is made in your baby it's important to rule out whether the problem is isolated or associated with other cardiac or genetic conditions. The genetic makeup of the baby can be determined by doing amniocenteses (procedure of taking out small amount of amniotic fluid around fetus with the help of ultrasound and thin needle) and genetic testing of the amniotic fluid.

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You will need consultation with the fetal medicine specialist to look for any other anatomic defects in the baby on ultrasound. Fetal echocardiography, a specialised and detailed ultrasound of the baby's heart during the pregnancy is also suggested, with consultation by a Pediatric Cardiologist who will care for your baby after birth.

## **What are the things to watch for during the pregnancy?**

During pregnancy, a double aortic arch will not cause any problems to the baby. A detailed ultrasound examination from a fetal medicine specialist is suggested. During pregnancy scan the fetal medicine specialist, in conjunction with a Pediatric Cardiologist will look for the branching pattern of the double aortic arch, variations in the configuration of the double aortic arch, evaluate the size of the thymus gland (a gland present in upper thorax) and check if there are any associated heart and other anatomic defects in the baby.

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

The appearance of symptoms in the baby related to the double aortic arch is usually depends upon tightness of the vascular ring formed around the trachea and esophagus by the double aortic arch. Prior to birth this structural difference will generally not cause problems for the baby. Even in first few years of life your baby may be asymptomatic. Symptoms, if present, are related to the compression of trachea or esophagus or both by the double aortic arch. Compression of the trachea causes symptoms related to the respiratory system like cough, asthma, wheezing and recurrent episodes of pneumonia. Compression of the esophagus causes symptoms like difficulty in swallowing, choking episodes and reflux disease (food comes back up from the esophagus).

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## Will it happen again?

If there is no other genetic disease is found with double aortic arch, the risk of this condition occurring again in your baby is very low. If an underlying genetic condition is found the risk of recurrence depends on the nature of the genetic condition. In this event consultation with a geneticist will be helpful to know more about this.

## How is it managed after delivery and what tests done?

To confirm the diagnosis of double aortic arch after delivery various tests can be done like chest X ray, CT scan, MRI and Echocardiography (echo). Passing a scope down the trachea, (bronchoscopy) may be done to evaluate the patency of the airway of your baby. This condition is managed surgically. The surgeon makes cuts on lateral aspect of chest of your baby between two ribs, enters thorax and divides and ligates the compressing segment of double aortic arch on trachea. This will release pressure and compression over the trachea. Surgical repair has excellent outcomes and the risk of complications from double aortic arch repair is low. Rarely, a baby may have trouble with feeding or have persistent breathing problems. You can discuss with pediatric thoracic surgeon regarding complications and outcome of the surgical treatment in detail.

## What other questions should I ask?

- How often will I have ultrasound examinations done?
- Does the baby's heart look normal?
- Does my baby have other anatomic defects?
- Where should I deliver so that baby will receive best care after it is born?
- What symptoms should I look out for at home?
- When will my baby need surgery?
- Where will the surgery take place, and who will provide it?
- Should my baby or others in our family have genetic testing?

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