Role of magnetic resonance imaging in diagnosis of complex congenital aortic arch anomalies

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Over the last few years the role of CMR in the diagnosis of aortic arch abnormalities is becoming increasingly important.¹

Today, cardiovascular magnetic resonance (CMR) is routinely employed in detaching and evaluating aortic arch abnormalities. CMR and CT scan are the techniques commonly used when a detailed examination of aortic arch and its branches is required.

However, complex congenital anatomical abnormalities of the aortic arch and its branches remain a challenging situation to diagnose, image and treat.

Usually, patients are referred to MRI for a suspicion of aortic arch anomaly at ultrasound. At ultrasound examination, the aortic arch and its branches may be extremely difficult to visualise. In fact, the acoustic window is a major limitation when extra-cardiac anatomy need to be examined.

CMR not only allow for an accurate anatomical and functional intra-cardiac examination, but also provides a detailed visualisation of extra-cardiac structures.

Three-dimensional (3D) volume-rendered and multi-reformatted images of gadolinium-enhanced MRA allows for an excellent visualisation of thoracic vessels and structures.

A complete MR examination should include 3D axial volume images of heart and great vessels, cine images, short axis ventricular volumes, phase contrast flow images of great vessels and 3D gadolinium MRA.

There is a wide spectrum of anomalies of aortic arch and its branches, from severely symptomatic to asymptomatic forms. Symptomatic forms usually present with stridor, breathlessness and dysphagia of unknown origins. However, symptoms presentation is often misleading and diagnosis may be difficult. In our experience most of the patients referred to CMR were asymptomatic for airways or oesophageal compression and arch abnormalities were noticed during investigation for associated conditions.

We describe three of the commonest types of anomalies of aortic arch and its branches.

**Double aortic arch** is an anomaly in which both right and left aortic arches are present and is the commonest symptomatic condition. Several variants are described, depending on the size of the two arches. Incomplete double aortic arch is a variant in which one of the two arches, usually the left, is partially or completely atretic and persisting as a fibrous cord connecting to the descending aorta.² Usually, the double aortic arch is complete and the right-sided arch is dominant in approximately 70% of the cases.³

Technically, all double aortic arches forms a complete vascular ring around the trachea and the oesophagus; however, clinical manifestations are related to the tightness of the ring. Symptoms vary from with stridor to breathlessness, recurrent respiratory infections and dysphagia in the newborn period or early in life. Surgical division is indicated in these patients. In our experience, the vast majority of patients with symptoms of airway or oesophageal compression had a double aortic arch. This abnormality is caused by the persistence of both right and left fourth branchial arches.

**Right-sided aortic arch** is due to the regression of the left arch in the embryonic development. This form of aortic arch anomaly is usually asymptomatic; however, it is often associated with major intra-cardiac diseases. Rarely, these patients have a vascular ring; however, it is more likely in those with no intra-cardiac anomalies.² Right-sided aortic arch may present in many forms, the most common of which is with mirror image branching of head and neck vessels.³ The other three major types of right aortic arch are: with retro-oesophageal left subclavian artery, with retro-oesophageal diverticulum and with left descending aorta. Right-sided aortic arch with mirror image branching is nearly always associated with intra-cardiac abnormalities. Particularly, there is an high incidence of right-sided aortic arch with mirror image branching in cono-truncal anomalies such as pulmonary atresia with intact ventricular septum, tetralogy of Fallot and double outlet right ventricle with atrial isomerism. In most of the cases, right-sided aortic arch with mirror image branching may be considered as an anatomical variant associated with major cardiac disease. However, this anomalous arrangement of aortic arch and head and neck vessels may have an implication in the surgical management of these patients, especially in cyanotic cases requiring a BT shunt.⁴

**Aberrant right sub-clavian artery** is due to an anomalous regression of the fourth aortic arch between the right common carotid artery and the right sub-clavian artery. It originates from the proximal descending aorta and passes posterior to the oesophagus, forming an incomplete vascular ring.³ Also this anomaly is usually asymptomatic, however, it may present with symptoms of stridor or dysphagia, due to the compression of the trachea or oesophagus. If symptoms occur, usually there is a typical presentation early in

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life with a tendency to regress after a few months. A surgical correction is almost never required.1,8

Usually, aberrant right sub-clavian artery is a single anomaly of the aortic arch. It may, however, be associated with coarctation, complete interruption of the aortic arch or with a right-sided aortic arch.11 In our experience an aberrant right sub-clavian artery has been often detached as an incidental finding during the follow-up of patients with coarctation of the aorta. However, only around 1% of patients with aortic coarctation have an associated anomalous right sub-clavian artery. The aberrant vessel may arise either proximal or distal to the coarctation site; however, post-stenotic origin is the most common.12,13 In patients with aortic coarctation, usually the aberrant vessel may be considered as an anatomical variant. However, when surgery of the coarctation is required, an aberrant right sub-clavian artery may represent an increased risk of postoperative complications such as paraplegia.1,9 MRI, defining in detail the aortic arch anatomy, can add helpful information in planning the management of these patients.

In conclusion, CMR is an important diagnostic tool in evaluating anomalies of aortic arch and its branches. CMR allow for an accurate visualisation of anomalous vessels and their relationship with airways and oesophagus. For this reason, CMR is extremely helpful in identifying those patients requiring a surgical correction, in planning their management and in their follow-up.

In contrast, to CT scan has the well-known advantage of avoiding ionising radiation. This is particularly important in children in whom we strive to avoid radiation exposure and in all the patients in which periodic examinations are required. Moreover, CMR by virtue of providing high-resolution three-dimensional images offer also the advantage of a detailed assessment of intra-cardiac anatomy and function. Interventional MRI is the present and the future of MRI in the management of patients with aortic arch anomalies. Interventional MRI has been employed already in animals models; however, its use on human remains difficult. Although excellent real-time MR imaging sufficient to guide investigational therapeutic procedures already exists, many problems remain with the use of compatible catheter and devices.15

Conclusions
CMR can be considered the gold standard technique in the evaluation of anomalies of aortic arch and its branches. Moreover, it is extremely helpful in planning (when necessary) the surgical management of these patients and in their follow-up. In the near future, real-time MRI could be employed to guide interventional procedures.

References


