CONGENITAL HEART DISEASE

Aortic Arch Anomalies

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ABSTRACT

Magnetic resonance imaging has become the gold standard for imaging the aortic arch with the ability to demonstrate not only the arterial branching pattern, but also the relationship of aorta and its branches to the trachea and bronchi. Examples of four major aortic arch types—left aortic arch, right aortic arch, double aortic arch and persistent fifth aortic—with most of their variations are demonstrated.

INTRODUCTION

Anomalies of the aortic arch were among the first congenital cardiovascular defects to be examined using clinical magnetic resonance imaging. While the clinical recognition of arch anomalies by plain chest radiograph and barium esophagography goes back decades, the precision of magnetic resonance imaging and computed tomography has virtually supplanted all other imaging techniques when surgical decision making requires an accurate anatomical diagnosis (1–7).

DEFINITIONS

In order to describe the various arch anomalies, some basic definitions are required.

Arch sidedness

An aortic arch is a vessel, whether patent or atretic, that connects the ascending and descending aorta. There are three basic types of aortic arch from the standpoint of the sidedness: left, right and double. Arch sidedness refers to the side of the trachea that the aortic arch passes as it crosses a mainstem bronchus.

This means that an ascending aorta that swings far to the right of the trachea before traveling back to the left and crossing the left mainstem bronchus is a left aortic arch. This may be difficult to appreciate with projection imaging such as angiography but is easily recognized on magnetic resonance imaging. Right aortic arches cross over the right mainstem bronchus. Double aortic arches consist of two aortic arches, one crossing the right and the other crossing the left mainstem bronchus.

Vascular ring

A vascular ring is an anomaly in which the trachea and esophagus (or its atretic remnant) are completely surrounded by vessels. These may include aortic arch or arches, arch vessels, pulmonary artery branch or branches (any of which may be patent or atretic), and ductus arteriosus or ligamentum arteriosum (Henceforth, ductus will be understood to mean ductus arteriosus and ligamentum, ligamentum arteriosum). Vascular rings may be sufficiently tight to cause clinical symptoms or may be loose enough to be asymptomatic. Symptoms are typically respiratory in nature at ages less than two years and relate to dysphagia in older children and adults although less frequently respiratory symptoms have been noted in older children and adults (8).

Since not all components of a vascular ring may be patent, and only vessels carrying blood may be visualized by magnetic resonance imaging (or any other currently available imaging modality other than direct inspection) there are three “Ds” for diagnosing vascular rings when the trachea and esophagus are not surrounded by patent vessels:

(Ductus) Dimple opposite the side of the arch
Diverticulum opposite the side of the arch
(Proximal) Descending Aorta opposite the side of the arch

Each of these will be described in more detail under the specific arch anomalies. The importance of these is that even in the
absence of visualization of vessels on all sides of the trachea and esophagus, one can make the diagnosis of vascular ring with certainty.

**Non-ring tracheo-bronchial or esophageal compression by arch or arch vessels**

Sometimes the trachea or bronchi may be partially compressed by the great arteries without a complete ring. Cases of innominate artery compression of the trachea (Fig. 1), anomalous origin of the left pulmonary artery from the right pulmonary artery, so-called pulmonary artery “sling (Fig. 2),” and the aorta wrapping part way around the trachea (Fig. 3) are examples of this. In cases where the aortic arch is oriented in a sagittal plane as is frequently the case with right aortic arches but can also occur in left arches with the ascending aorta in an unusual leftward position, the left mainstem bronchus may be compressed by the ascending and descending aorta plus the interposed right pulmonary artery (Fig. 4).

**PARTICULAR ADVANTAGES OF MRI FOR ARCH ANOMALIES**

Magnetic resonance imaging is clearly one of many imaging modalities by which arch sidedness and branching pattern may be discerned. However, the importance of simultaneous visualization of the trachea and bronchi along with vessels degrades the potential contributions of angiography and ultrasound examinations. Recognition of airway compression by vascular structures is a valuable adjunct to determination of arch anatomy. Furthermore, even in those cases where arch sidedness alone is the goal of imaging, such as in planning for surgical repair of esophageal atresia, artifacts introduced by the lungs and airway negatively affect the ability to accurately assess the relationship of aortic arch to trachea. Therefore, branching pattern is often used as a surrogate for arch-sidedness. As a general rule the first arch vessel (typically carotid artery or innominate artery) travels to the side opposite the side of the aortic arch. However, there are exceptions to this rule: 1) Rarely a retroesophageal innominate artery arises as the last vessel from the arch rather than the first. Thus, the first arch vessel, a carotid artery, is ipsilateral to the arch. 2) More commonly, both carotid arteries may arise adjacent to each other so that one cannot distinguish which is first.

**Figure 1.** Innominate artery compression of trachea. Axial black blood images showing distortion of the trachea (T) from its nearly round shape in Fig. 1A to a more triangular shape in Fig. 1B at the point where the right innominate artery (RIA) crosses it. The trachea is also abnormally flattened in Fig. 1C, and some distortion of the left bronchus is noted in Fig. 1D at the level of the carina (Car).

**Figure 2.** Anomalous origin of the left pulmonary artery from the right, so-called pulmonary artery sling. The left pulmonary artery arises from the right (RPA) instead of from the main pulmonary artery (MPA) and passes between T and esophagus (E). AAo = ascending aorta; DAo = descending aorta. Reproduced with permission (14).

**Figure 3.** Tracheal compression by aortic arch without ring. Case of marked dextrocardia with extreme rightward displacement of AAo. Note coronal orientation of aortic arch (Arch) in Fig. 3A and narrowing of T in Fig. 3B. LPA = left pulmonary artery. Reproduced with permission (15).
3) If one can distinguish which is first, right or left arch may be diagnosed incorrectly when, in fact, there is a double aortic arch. In all of these cases significant errors in diagnosis can result from inappropriate application of the branching rule. Another rule that we believe has no exceptions states that a retroesophageal or an isolated subclavian artery is always opposite the side of the aortic arch. While this rule is more reliable than the previously noted one, it is also more difficult to apply. Retroesophageal vessels are difficult to see with ultrasound since they run behind the air-filled trachea. Furthermore the aorta coursing behind the esophagus, as noted on barium swallow, can be mistaken for a retroesophageal subclavian artery.

While barium esophagography and echocardiography can accurately determine arch-sidedness and presence of a vascular ring in the majority of cases, neither modality can reliably delineate the optimal site for division of a ring, particularly in those cases with atretic or hypoplastic segments. Therefore, the surgeon is required to do more extensive dissection in order to find the best location for division. It is also noteworthy that errors in determining arch sidedness from barium swallow or echocardiography are not that rare, and their occurrence may result in bilateral thoracotomies. In the current era of minimally invasive surgery, precise definition of arch anatomy, such as that provided by magnetic resonance imaging noninvasively, is superior to the aforementioned techniques as well as to the more invasive angiographic methods.

**MR TECHNIQUES FOR EVALUATION OF ARCH ANOMALIES**

The protocol for imaging the aortic arch and related structures is designed to accomplish the following: 1) rapid identification of the basic arch anatomy so that necessary surgery can be carried out, even if the patient becomes unstable from a respiratory standpoint before completion of a comprehensive study; 2) determination of tracheo-bronchial compression and its relationship to vascular structures; and in the case of vascular rings 3) optimal imaging of the aortic arch to determine the ideal site for division.

**2D imaging**

The study begins with simple transverse/axial imaging from a level in the neck just below the larynx, down to the level of the diaphragm. Trachea and esophagus are identified, and any area of narrowing or distortion of the nearly circular trachea is noted. Each arch vessel is followed from its respective termination down to the level of the aortic arch. If time permits, simple coronal imaging affords rapid cross-sectional imaging of the aortic arch(es) on either side of the trachea, which is helpful in deciding which arch is larger in the case of double aortic arch. This view is also useful in recognizing a diverticulum (discussed below) in vessels running more vertically than horizontally. Also, if time permits, a series of simple sagittal images is particularly useful for assessing the linear extent and degree of tracheal narrowing (Fig. 5).

**3D imaging**

After 2D imaging in one or more standard planes, gadolinium injection is performed to provide a 3D shaded surface display of the aorta. In the case of a vascular ring, this further facilitates decisions about where best to divide it. The other application of 3D imaging is to allow curved cuts through the aorta for cases where tortuosity obscures areas of narrowing. By essentially
re-slicing the aorta along its major axis, areas of stenosis or hypoplasia become more evident.

**Flow assessment**

Finally, in cases of double aortic arch where there is a question of which arch to divide, velocity mapping in the two arches will show which carries the lesser blood flow and therefore would be the better one to divide.

**NORMAL LEFT AORTIC ARCH**

The normal left aortic arch (Fig. 6) is characterized by a rightward ascending aorta that courses leftward and posterior crossing over the left mainstem bronchus and to the left of the trachea with the descending aorta to the left of the spine. The first arch vessel is a right innominate artery that gives rise to the right subclavian and right carotid arteries. The right subclavian in turn gives rise to the right vertebral artery. The second vessel is a left carotid artery, and the third is a left subclavian artery. In 10% of normal people, the left vertebral artery arises directly from the aortic arch as the third vessel just anterior to the left subclavian, which would then be fourth. If one only looks at the origins of the arch vessels, this variation could be confused with anomalous right subclavian (see below); however, the marked size discrepancy (vertebral smaller than subclavian) helps distinguish these two conditions.

**LEFT ARCH ANOMALIES**

**Retroesophageal right subclavian artery**

(Fig. 7)

The most common aortic arch anomaly is left aortic arch with anomalous right subclavian artery. The sequence of arch vessels is right carotid artery, left carotid, left subclavian, with the last vessel being a retroesophageal right subclavian artery. This does not form a vascular ring since the trachea and esophagus are only bordered on three sides by vascular structures – nothing passes to the right. In the older literature, this anomaly was referred to as dysphagia lusoria: dysphagia due to a “trick” of nature. This symptom is rare in children but is seen in a small number of adults.

**Left arch, right descending aorta**

(Fig. 8)

This is a rare arch anomaly (9) in which the aorta passes to the left of the trachea, swings immediately to the right behind the trachea and esophagus, and then turns inferiorly to become the descending aorta. A right ductus or ligamentum connects the rightward descending aorta to the right pulmonary artery, thus completing a vascular ring. The branching pattern is usually similar to that seen above with anomalous right subclavian artery although a normal branching pattern with right innominate artery is theoretically possible. The difference from the previously described retroesophageal right subclavian artery is that while the branching sequence is the same, it is the aortic arch itself that is retroesophageal; the right subclavian artery arises from the aorta after it has become right-sided. Therefore the right subclavian
Left aortic arch, right descending aorta. Rare vascular ring. Axial images in Figs. 8A, C, and D, and a shaded surface display in Fig. 8B created from semiautomated reconstruction from standard black-blood spin echo images. The branching sequence is similar to that in Fig. 7, but the aortic arch itself passes retroesophageally – circumflex aortic arch (CircAA). This pattern only occurs when there is a ductus or ligamentum connecting the right descending aorta to the right pulmonary artery. Note the flattening of the T in Fig. 8C. Reproduced with permission (16).

Right aortic arches are a diverse set of anomalies including the second and third most common vascular rings. Because the aorta usually arises to the right of the midline (irrespective of arch sidedness) and, with one exception, right aortic arches aorta begin descending on the right, there is a chance that the right mainstem bronchus will be compressed between the sagittally oriented ascending and descending aorta as described above. In situs solitus (normal arrangement of asymmetrical body organs and atria) the descending aorta is left sided at the diaphragm irrespective of arch sidedness. Thus with right aortic arches the aorta must go from being right-sided at the level of the bronchi to left-sided at the diaphragm. In most cases this is a gradual diagonal passage with no perceptible indentation on the esophagus. However, in one situation – right aortic arch with left descending aorta – similar to left arch right descending above, the crossing is abrupt and associated with a ring.

Mirror image right aortic arch (Fig. 9)

Mirror image right aortic arch is the mirror image of the normal left aortic arch. That means the branching pattern consists of a left innominate artery as the first branch, followed by right carotid, then right subclavian artery. The descending aorta begins on the right. There are several variations on this theme with regard to ductus/ligamentum location. A true mirror image includes a right ductus passing between the aortic arch or proximal descending aorta and the right pulmonary artery. This is the only variation possible in the absence of a major intracardiac abnormality. A left ductus with a right ductus can occur with or without heart disease. A left ductus alone or congenital absence of the ductus can only occur in the presence of a major intracardiac anomaly in which right ventricular blood can reach the aorta without traversing the main pulmonary artery, such as by way of a large ventricular septal defect. This is because a left ductus with mirror image right aortic arch arises from the base of the left innominate artery. If the only egress of blood from the fetal right ventricle were via the pulmonary artery and left ductus, the majority of the cardiac output would take a very tortuous path before reaching the aorta. Furthermore, one would expect to see marked dilatation of the innominate artery because of the high percentage of right ventricular contribution to cardiac output. In reality, this is not seen. Obviously absence
of the ductus requires major intracardiac shunting for the same reason.

**Right aortic arch with retroesophageal left subclavian artery (non-ring) (Figs. 10, 11)**

This anomaly is the mirror image of left aortic arch with retroesophageal right subclavian artery. The branching sequence is left carotid, right carotid, right subclavian and anomalous left subclavian arteries. If there is no major intracardiac anomaly, there must be a right ductus or ligamentum. With intracardiac shunt there can be congenital absence of the ductus. From a diagnostic standpoint, the caliber of the anomalous left subclavian artery is relatively constant from its origin in the proximal descending aorta for most of its intrathoracic course. There is no vascular ring since there is no vessel passing to the left of the trachea and esophagus. An indentation on the barium filled esophagus will be noted.

**Right aortic arch with diverticulum of Kommerell (Fig. 12)**

The branching pattern for this anomaly is virtually identical to the retroesophageal left subclavian artery described above with one important difference. Instead of the subclavian artery having a constant caliber, the retroesophageal portion is notably larger than the remainder of the vessel. The larger caliber proximal portion is referred to as an aortic diverticulum. There is an abrupt tapering of the vessel best appreciated on coronal imaging. This pattern is associated with the presence of a left ductus or ligamentum connecting left pulmonary artery to the diverticulum forming a vascular ring. This is the second most common vascular ring after double aortic arch. The so-called diverticulum is due to fetal ductal flow to the descending aorta by way of the proximal subclavian artery. While the ligamentum is not visible with any imaging modality, the aortic diverticulum assures the
Figure 13. Right aortic arch with left descending aorta. Axial TrueFISP images in Figs. 13A–C. RAA that passes behind the trachea and esophagus seen in Figs. 13B&C, so-called circumflex aortic arch (CircAA). This anomaly is much more common than its mirror image shown in Fig. 8 above. The tortuosity of the aorta makes it difficult to evaluate in any single plane. Fig. 13D is the result of a "curved cut" along the path described by the line shown in Fig. 13B. The aorta is effectively uncurled for better appreciation of its caliber throughout. As with the case shown in Fig. 8, the presence of a CircAA assures the presence of a vascular ring. LVrt = left vertebral artery, which joins the LSCA more inferiorly.

Right aortic arch with left descending aorta (Figs. 13, 14)

This anomaly, also known as circumflex aortic arch, is the third most common form of vascular ring. There are two branching patterns associated with this anomaly. One has an anomalous left subclavian artery that is not retroesophageal—essentially the mirror image of the rare left aortic arch with right descending aorta described above. The other has a mirror image right aortic arch branching pattern—left innominate, right carotid, right subclavian arteries—with the aortic arch itself passing retroesophageally to the left of the spine where it is joined by the left ductus or ligamentum. Both of these patterns are indistinguishable from double aortic arch with atretic left arch described below.

Right aortic arch with retroesophageal left innominate artery (non-ring) (Fig. 16)

This is a very rare arch anomaly (11) in which the first arch vessel is a right carotid artery in "violation" of the usual rule of vessel branching.

Figure 14. Right aortic arch with left descending aorta. 3D shaded surface display of same case shown in Fig. 13. The marked tortuosity of the aorta is again appreciated. Fig. 14A is an anterior view, 14B left lateral, 14C posterior, and 14D right lateral. In this case a diverticulum (Div) receives both the left subclavian artery, which is aberrant but not retroesophageal, and the ligamentum arteriosum.

Figure 15. Right aortic arch with left ductus or ligamentum. Axial (Figs. 15A-C) and coronal (Figs. 15D-F) black-blood images demonstrate a rare vascular ring. The branching pattern appears to be a mirror image right aortic arch, but the ductus dimple (Dimp) seen heading leftward from the descending aorta (Figs. 15C and F) indicates a left-sided ligamentum, which completes a vascular ring with a right aortic arch. Of note is the fact that a left ductus or ligamentum from a left innominate artery would not create a ring, but this vessel courses posterior to the trachea and esophagus; whereas a ligamentum from a left innominate artery does not.
that the first arch vessel contains a carotid artery opposite the side of the arch. The second branch is the right subclavian, and the last is a vessel that gives rise to both the left carotid and left subclavian arteries – designated innominate artery. The anomalous innominate, while producing a relatively large indentation on the barium-filled esophagus, does not create a vascular ring as there is no left-sided vessel completing the ring.

**DOUBLE AORTIC ARCH**

Double aortic arch is a persistence of both the left and right embryonic fourth arches and left and right dorsal aortas. Typically the descending aorta is on the left although a midline or right-sided position of the proximal descending aorta are possible. In most but not all cases, the right arch is larger than the left (dominant right arch), but equal sized arches and even dominant left arch are known.

If there is no associated major intracardiac anomaly, as is usually the case, there must be a ductus. Typically it is left-sided. Right-sided ductus can occur. Bilateral ductus is theoretically possible, but its occurrence is extremely rare. The importance of ductus or ligamentum location is that it may need to be divided at the same time as division of one of the arches for relief of the vascular ring. If it is not, the ductus or ligamentum can still complete a ring, even though the arch is successfully divided.
Double aortic arch, atretic left arch distal to left subclavian artery. Axial TruFISP images (Figs. 19A–D) and black-blood coronal image (Fig. 19E) show the more common double aortic arch with atretic left arch. While there is no connection between the LAA in Fig. 19B and the DAo, the small diverticulum from the DAo in Fig. 19C assures that there is a complete ring. Fig. 19E shows the markedly larger RAA.

It is usually relatively easy to determine which of the two arches is smaller and therefore which should be divided in the symptomatic patient. However, in cases where the caliber of the two arches appear similar, one can measure blood flow in the two distal arches using phase encoded velocity mapping. The arch with lesser flow would be the ideal one to divide.

**Dominant right arch**

By far the most common double arch is a dominant right aortic arch. This is usually evident on coronal imaging in a plane passing through the trachea where the two arches are seen on either side. Typically, the right arch is superior to the level of the left. Occasionally the arches will be of equal size in this plane, but the more posterior portion of the left arch will show more obvious diminution in caliber or may even disappear (i.e., have atresia).

Both arches are patent in the majority of double aortic arches. On axial imaging, one may not appreciate the complete ring in any single slice (Fig. 17). The right arch is almost always superior to the left. Furthermore, the proximal bifurcation is typically superior to the distal confluence of the two arches. Three-dimensional displays facilitate the viewing of the complete ring in a single image. A left or right posterior oblique view with cranial angulation usually eliminates overlapping of structures to best appreciate the ring and the relative sizes of the arches.

Double aortic arch with atretic left arch can occur in either of two configurations: atretic segment between carotid and subclavian arteries (Fig. 18) or distal to the left subclavian artery (Fig. 19) (12). The former may be indistinguishable from right aortic arch with retroesophageal diverticulum of Kommerell in the unusual case of a right descending aorta. However, both patterns may be indistinguishable from the two forms of right aortic arch with left descending aorta. Sometimes there is noticeable tethering of the left carotid or left subclavian artery posteriorly from the atretic arch, but this is not a consistent finding. Therefore, it is best to remind the surgeon who is about to divide a vascular ring thought to be due to right aortic arch with retroesophageal diverticulum or right arch with left descending aorta that there could be an atretic left aortic arch in addition to a left ligamentum.

**Equal sized arches (Fig. 20)**

These have two well-developed arches that are both patent. Similar to dominant right arch, the right arch is typically superior to the left, sometimes a cervical arch, i.e., superior to the head of the clavicle.

**Dominant left arch (Fig. 21)**

Double aortic arch with a dominant left arch is uncommon but not rare. This anomaly along with the rare left aortic arch with right descending aorta (noted above) are the two situations where a vascular ring should be divided by way of a right thoracotomy rather than a left. These two anomalies are not usually recognized by barium swallow or echocardiography. Careful attention must
Figure 21. Double aortic arch, dominant left arch. Axial TruFISP images (Figs. 21A-C) and coronal black-blood image (Fig. 21D) demonstrate a relatively uncommon double aortic arch with a larger left arch seen in both axial (Fig. 21B) and coronal (Fig. 21D) view. This appears to be a relatively loose ring with minimal tracheal distortion.

be paid when evaluating a double aortic arch with two patent arches because the caliber may vary in different parts of the arch. As discussed previously, simple axial or transverse imaging for comparison of arch caliber is complicated by the fact that the ring is not seen in any single transverse plane. In addition, as mentioned above, the narrowest portion of the right arch may not occur in the coronal plane where the two arches straddle the trachea. Three-dimensional rendering is especially useful in this situation. Shaded surface displays are particularly helpful in comparing the two arches. In those cases where a discrete coarctation or relatively short hypoplastic segment of the right arch is present, a curved cut along the longitudinal axis of the aorta may be the best way to assess the sizes of the two arches.

PERSISTENT FIFTH AORTIC ARCH

Persistent fifth aortic arch is really two different anomalies in which there are two aortic arches but not a vascular ring. The first—so-called double-barreled aortic arch or “subway” aortic arch—is an asymptomatic curiosity; there are two parallel arches with the brachiocephalic vessels arising from the more superior one (presumably the derivative of the normal embryonic fourth branchial arch [Fig. 22]) (13). Its importance is that it not be confused with a dissecting aneurysm of the aortic arch. The other anomaly consists of atresia of the distal portion of the “normal” aortic arch with patency but coarctation of the distal end of the persistent fifth embryonic arch (Fig. 23). This anomaly is recognized by the apparent origin of all four brachiocephalic vessels from a single stalk—the embryonic fourth arch. The atretic portion is distal to the left subclavian artery. While not visualized by any imaging modality, the unusual appearance of a truly common brachiocephalic trunk identifies this anomaly.

REFERENCES
