

Congenital Anomalies of the Aortic Arch: Evaluation with the Use of Multidetector Computed Tomography

Aysel Türkvtan, MD
Fatma Gül Büyükbayraktar, MD
Tülay Ölçer, MD
Turhan Cumhur, MD

Index terms:

Aortic arch anomalies
Multidetector computed
tomography
Angiography

DOI:10.3348/kjr.2009.10.2.176

Korean J Radiol 2009; 10: 176-184

Received April 11, 2008; accepted
after revision August 11, 2008.

All authors: Department of Radiology,
Türkiye Yüksek İhtisas Hospital, Ankara,
Turkey

Address reprint requests to:

Aysel Türkvtan, MD, Department of
Radiology, Türkiye Yüksek İhtisas
Hospital, Kızılay Street, No:4, 06100,
Sıhhiye, Ankara, Turkey.
Tel. +90-312-3061671
Fax. +90-312-3124120
e-mail: aturkvatan@yahoo.com

Abbreviations:

ALSA = aberrant left subclavian artery
ARSA = aberrant right subclavian artery
CAA = cervical aortic arch
IAA = interrupted aortic arch
LAA = left aortic arch
LSA = left subclavian artery
MDCT = multidetector computed
tomography
MinIP = minimum intensity projection
MIP = maximum intensity projection
MPR = multiplanar reformatting
RAA = right aortic arch
VR = volume rendering

Congenital anomalies of the aortic arch have clinical importance, as the anomalies may be associated with vascular rings or other congenital cardiovascular diseases. Multidetector computed tomography (MDCT) angiography enables one to display the detailed anatomy of vascular structures and the spatial relationships with adjacent organs; this ability is the greatest advantage of the use of MDCT angiography in comparison to other imaging modalities in the evaluation of the congenital anomalies of the aortic arch. In this review article, we illustrate 16-slice MDCT angiography appearances of congenital anomalies of the aortic arch.

Congenital anomalies of the aortic arch are uncommon and may be associated with other congenital cardiovascular diseases. A congenital anomaly of the aortic arch is usually an incidental radiological finding in asymptomatic patients, except when the anomaly constitutes a vascular ring that is formed when the abnormally patterned arch vessels completely encircle the trachea and esophagus (1). Multidetector computed tomography (MDCT) angiography has recently become a principal diagnostic method for the assessment of thoracic aortic abnormalities. The advantage of the use of MDCT angiography is that the modality is a noninvasive technique that enables evaluation of vascular anomalies and the status of tracheal or esophageal compression in the same study (2, 3). Disadvantages of the use of MDCT angiography, especially in pediatric patients, are the necessity to use iodinated contrast medium as well as an increase in patient radiation exposure. Magnetic resonance (MR) angiography is an alternative non-invasive tool without the need for contrast material or radiation exposure. However, the method is time-consuming and may require the need for prolonged sedation in pediatric patients. Although MR angiography may show vascular anomalies, the technique does not provide sufficient information regarding the esophagus and trachea.

Multidetector CT Scanning Technique

Multidetector CT angiography examinations were performed with a 16-row MDCT (LightSpeed Ultra, GE Healthcare, Milwaukee, WI). Patients were examined while in the supine position and all images were acquired during a single breath hold, extending from the base of the neck to the diaphragm. In adult patients, automatic tube current dose modulation was used. Imaging parameters were as follows: tube voltage, 100–120 kV; tube current, 140–300 mA, collimation, 16 × 1.25 mm; slice thickness, 1.25 mm; increment 0.6 mm; table feed, 27.5 mm/sec; rotation time, 0.5 sec. In pediatric patients, a body weight-based low-dose protocol (80–100 kV, 25–120 mA) was used to further reduce radiation exposure. A tube voltage of 80 kV was used for patients weighing less

MDCT Evaluation of Congenital Anomalies of Aortic Arch

than 50 kg and 100 kV was used for patients weighing more than 50 kg. A tube current of 25 mAs was used for patients weighing less than 15 kg. A tube current of 30 mAs was used for patients weighing between 15 and 24 kg. A tube current of 45 mAs was used for patients weighing between 25 and 34 kg. A tube current of 75 mAs was used for patients weighing between 35 and 44 kg. A tube current of 100 mAs was used for patients weighing between 45 and 54 kg and a tube current of 120 mAs was used for patients weighing more than 54 kg. Imaging data was acquired after an intravenous injection of 1.5–2 ml/kg iodinated contrast agent (Iodixanol, 320 mgI/ml Visipaque, GE Healthcare) at

a rate of 3–4 ml/sec. The scanning delay was determined with the use of a bolus tracking technique. The examination was initiated 4 seconds after the attenuation of a region of interest positioned in the ascending aorta reached 150 Hounsfield units (HU). For three-dimensional image reconstruction, the raw MDCT data was processed on a separate workstation (Advanced Workstation 4.2, GE Healthcare) with multiplanar reformatting (MPR), maximum intensity projection (MIP), minimum intensity projection (MinIP) and volume rendering (VR). MIP and VR techniques were used mainly for evaluation of cardiovascular structures, the MinIP technique was used to evaluate the

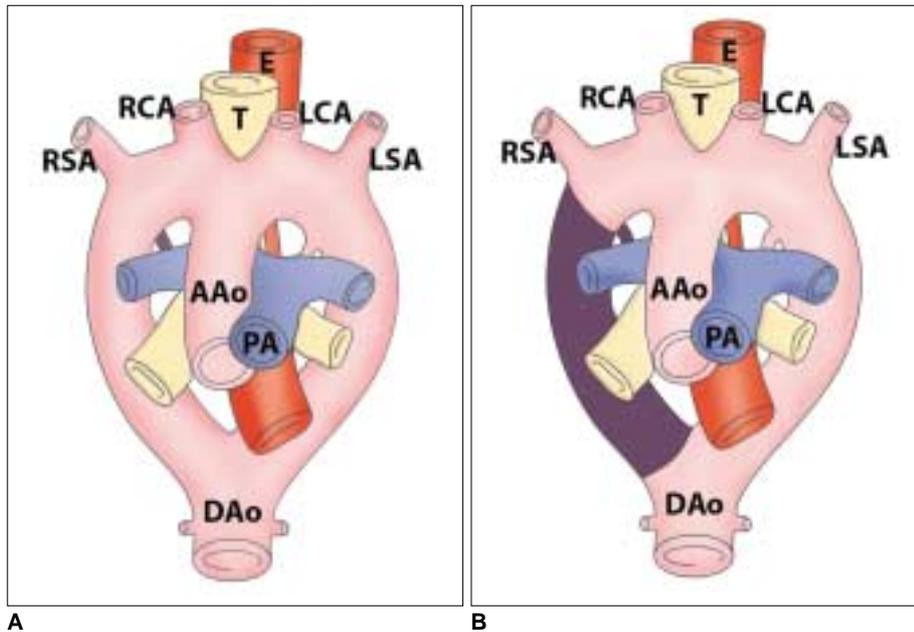


Fig. 1. Diagram of Edward's developmental model of aortic arch. **A.** According to hypothetical double aortic arch system in which there is aortic arch and ductus arteriosus on each side, carotid and subclavian arteries arise from their respective arches. Descending aorta is in midline. **B.** Normal branching arch results from interruption of dorsal segment of right arch between right subclavian artery and descending aorta, with regression of right ductus arteriosus. AAo = ascending aorta, DAo = descending aorta, PA = pulmonary artery, T = trachea, E = esophagus, RSA = right subclavian artery, RCA = right carotid artery, LCA = left carotid artery, LSA = left subclavian artery

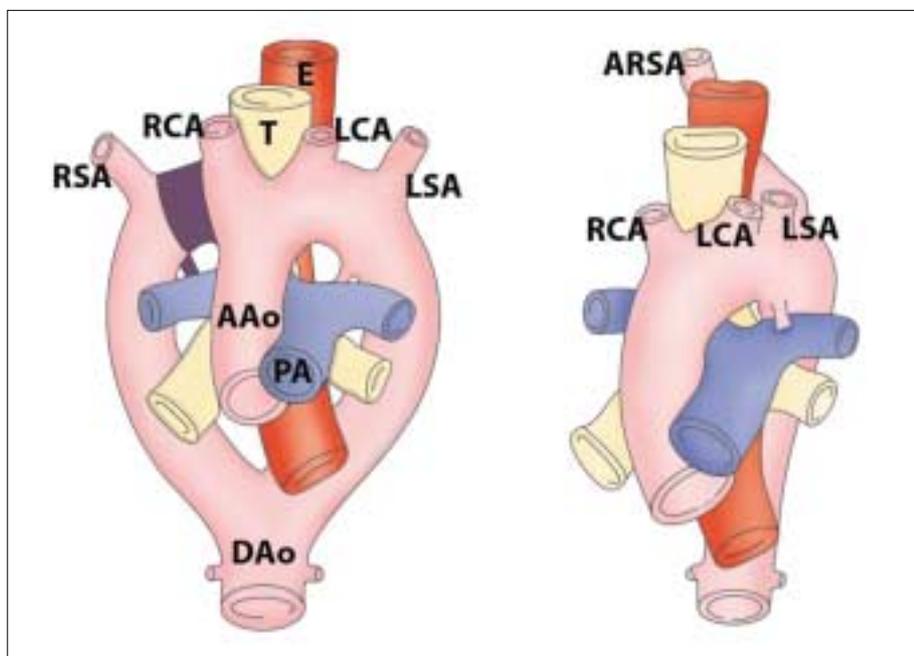


Fig. 2. Diagram of development of left aortic arch with aberrant right subclavian artery is presented. This anomaly results from interruption of dorsal segment of right arch between right carotid artery and right subclavian arteries with regression of right ductus arteriosus in hypothetical double aortic arch. AAo = ascending aorta, DAo = descending aorta, PA = pulmonary artery, T = trachea, E = esophagus, RSA = right subclavian artery, RCA = right carotid artery, LCA = left carotid artery, LSA = left subclavian artery, ARSA = aberrant right subclavian artery

tracheobronchial air column and the MPR technique was used to evaluate both cardiovascular structures and the status of tracheal or esophageal compression.

Embryology

According to Edward's hypothetical double aortic arch system in which there is an aortic arch and a ductus arteriosus on each side, the right carotid and subclavian arteries arise from the right arch and the left carotid, and subclavian arteries originate from the left arch. The descending aorta is in the midline (Fig. 1A). Interruption of this arch system at different locations can explain the various aortic arch anomalies. The normal arch system results from interruption of the dorsal segment of the right arch between the right subclavian artery and descending aorta, with regression of the right ductus arteriosus (4) (Fig. 1B).

Congenital anomalies of the aortic arch include the

following. The left aortic arch (LAA) with an aberrant right subclavian artery (ARSA), the right aortic arch (RAA) with an aberrant left subclavian artery (ALSA), the RAA with mirror image branching, the RAA with isolation of the left subclavian artery (LSA), the double aortic arch, the cervical aortic arch (CAA) and the interrupted aortic arch (IAA).

Left Aortic Arch with an Aberrant Right Subclavian Artery

An ARSA originating from normal left sided aortic arch is the most common aortic arch anomaly, with an incidence of 0.5-2% (5). This anomaly results from interruption of the dorsal segment of the right arch between the right carotid artery and right subclavian artery with regression of the right ductus arteriosus in the developing double aortic arch (4) (Fig. 2). In this anomaly, the right carotid artery arises as the first branch directly from the aortic

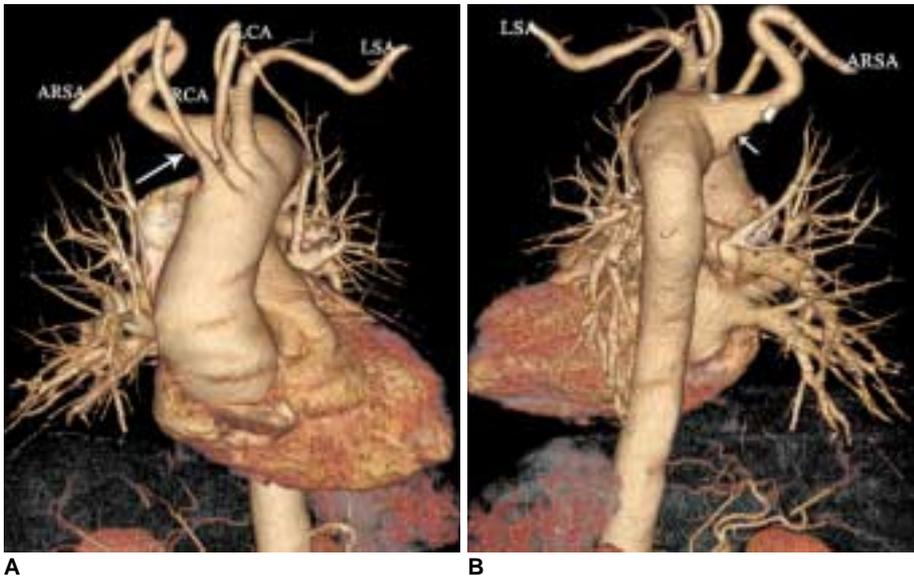
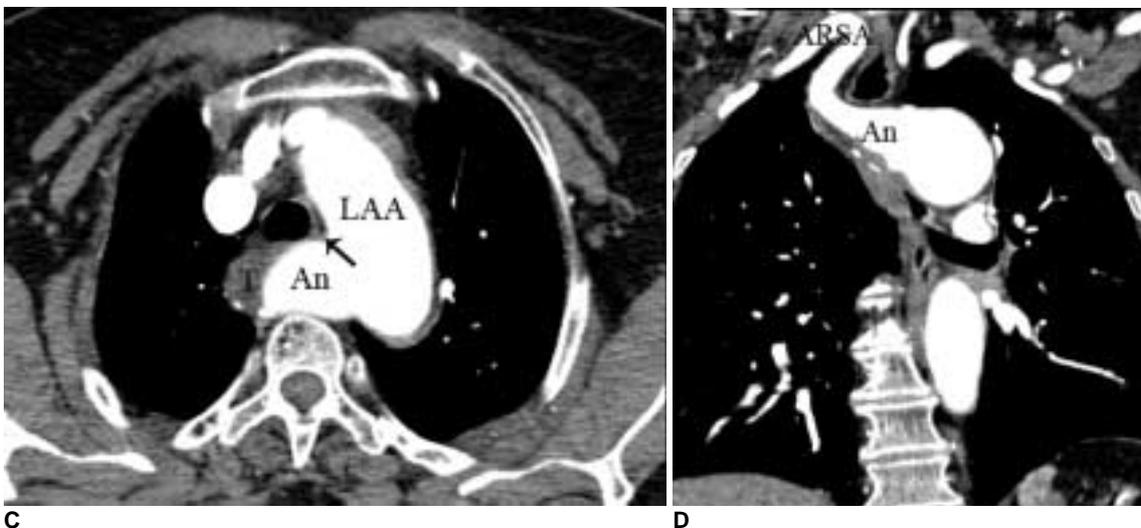


Fig. 3. Left aortic arch (LAA) with aberrant right subclavian artery (ARSA) in 66-year-old woman with dysphagia is shown. Anterior (A) and posterior (B) volume rendering images show ARSA with Kommerell's diverticulum at its origin (white arrows). Right carotid artery (RCA) arises as first branch from aortic arch, which is followed by left carotid artery (LCA), left subclavian artery (LSA) and ARSA. Axial (C) and coronal multiplanar reformatted (D) images show presence of aneurysm (An) of Kommerell's diverticulum with mural thrombus (T) and calcifications. There is compression of esophagus (black arrow).



MDCT Evaluation of Congenital Anomalies of Aortic Arch

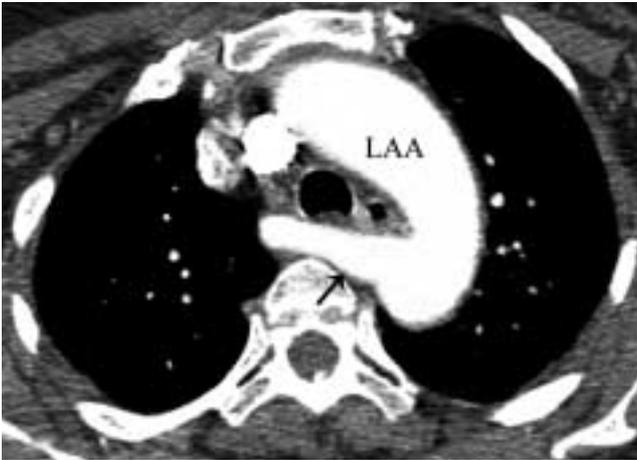
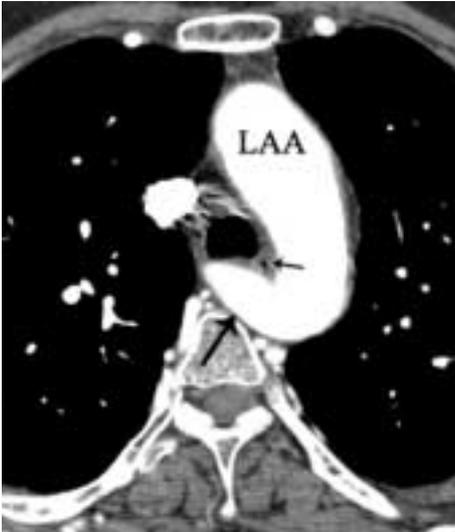
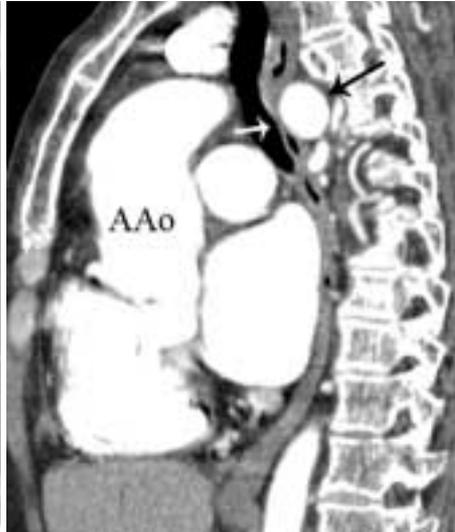


Fig. 4. Left aortic arch (LAA) with aberrant right subclavian artery in 48-year-old woman with anomalous origin of right vertebral artery from right common carotid artery is shown. Axial image shows retroesophageal course of aberrant right subclavian artery (arrow).

arch, which is followed by the left carotid artery, left subclavian arteries and ARSA (Fig. 3). The ARSA arises from the descending aorta as a last branch and crosses the mediastinum from left to right, passing behind the esophagus and trachea (Figs. 3, 4). An aortic diverticulum, also known as Kommerell's diverticulum, may be present at the origin of this vessel and has been reported in up to 60% of cases, representing the remnant of the distal RAA (6) (Fig. 3). An ARSA is generally asymptomatic and has been diagnosed incidentally, but about 10% of adults with this anomaly have symptoms of dysphagia due to extrinsic compression of the esophagus due to its retroesophageal course (Fig. 5). Rarely, with failure regression of the right ductus, a loose vascular ring may be formed by the LAA with an ARSA, right pulmonary artery and right ductus arteriosus (1). Aneurysmal dilatation of the proximal portion of an ARSA is uncommon. Such an aneurysm may



A

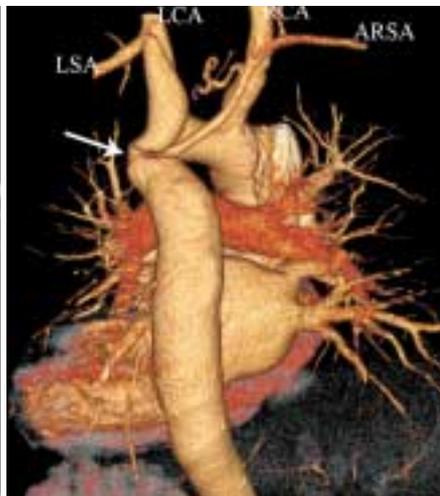


B

Fig. 5. Aberrant right subclavian artery in 53-year-old woman with severe aortic coarctation presenting with hypertension and dysphagia. Axial (A) and sagittal multiplanar reformatted (B) images show left aortic arch (LAA) with aberrant right subclavian artery (long arrows) causing esophageal compression (short arrows). Paravertebral collaterals and prominent internal mammary arteries are also seen. AAO = ascending aorta



A



B

Fig. 6. Aberrant right subclavian artery in 48-year-old man with aortic coarctation presenting with hypertension. Axial oblique (A) and posterior volume rendering (B) images show aberrant right subclavian artery (ARSA) arising just distal to aortic coarctation (arrows). LAA = left aortic arch, LSA = left subclavian artery, LCA = left carotid artery, RCA = right carotid artery

or may not be associated with Kommerell's diverticulum, and is believed to be the result of atherosclerotic disease (6) (Fig. 3). Generally, this anomaly is isolated, but may be associated with other cardiovascular anomalies, principally with coarctation of the aorta (Fig. 6), a patent ductus arteriosus, intracardiac defects, anomalous pulmonary artery circulation and carotid or vertebral artery anomalies (5, 7) (Fig. 7).

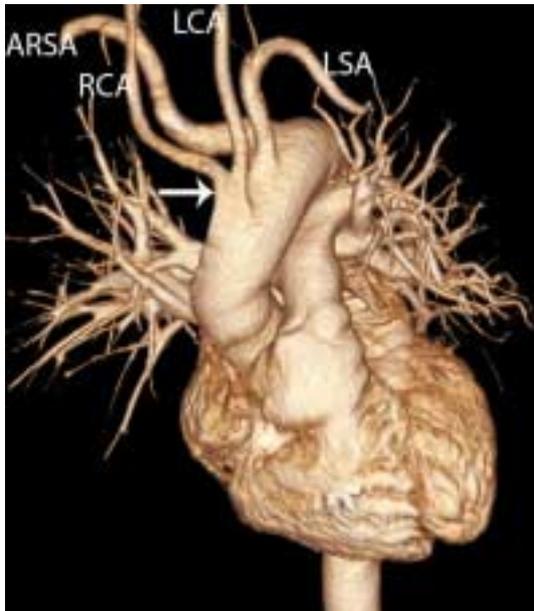


Fig. 7. Aberrant right subclavian artery in 51-year-old woman with dysphagia is shown. Anterior volume rendering image shows aberrant right subclavian artery (ARSA) and common origin of carotid arteries (arrow). RCA = right carotid artery, LCA = left carotid artery, LSA = left subclavian artery

Right Aortic Arch

The RAA is a relatively common anomaly, occurring in approximately 0.05% of the population (5). RAAs have been classified into three types according to the branching pattern of the arch vessels: an RAA with an ALSA, an RAA with mirror image branching and an RAA with isolation of the LSA.

An RAA with an ALSA is the most common type (5). This anomaly results from interruption of the dorsal segment of the left arch between the left common carotid and left subclavian arteries with regression of the right ductus arteriosus in the hypothetical double aortic arch (4) (Fig. 8). In this anomaly, the first branch arising from the aortic arch is the left carotid artery, which is followed by the right carotid artery, right subclavian arteries and ALSA in order (Fig. 9). The descending aorta is usually on the right side or near the midline. An ALSA may arise from a remnant of the left dorsal aortic root (Kommerell's diverticulum) (4) (Figs. 9, 10). This anomaly rarely produces symptoms and is usually an incidental radiological finding. Rarely, a right arch with an ALSA forms a complete vascular ring, left pulmonary artery and left ductus arteriosus. Symptoms of esophageal compression may develop in older individuals with ectasia, tortuosity or an aneurysm of the ALSA (1) (Figs. 10, 11). Respiratory symptoms due to tracheal compression may be present in pediatric patients (8). This anomaly is rarely associated with other cardiovascular abnormalities (5).

An RAA with mirror image branching is uncommon, but not rare (5). This anomaly results from interruption of the dorsal segment of the left arch between the LSA and the

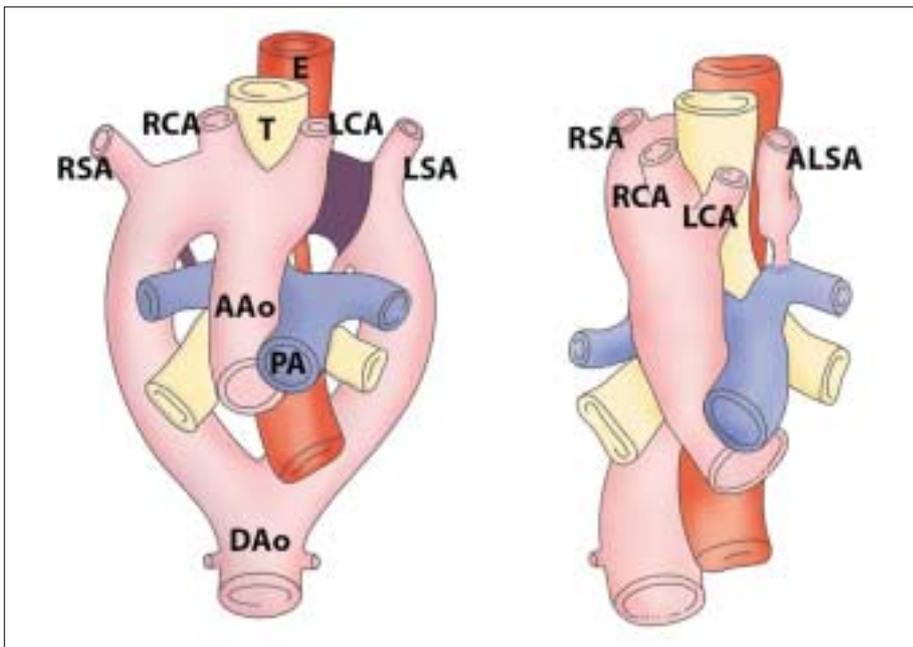


Fig. 8. Diagram of development of right aortic arch with aberrant left subclavian artery is shown. Anomaly results from interruption of dorsal segment of left arch between left common carotid artery and left subclavian arteries with regression of right ductus arteriosus in hypothetical double aortic arch. AAO = ascending aorta, DAO = descending aorta, PA = pulmonary artery, T = trachea, E = esophagus, RSA = right subclavian artery, RCA = right carotid artery, LCA = left carotid artery, LSA = left subclavian artery, ALSA = aberrant left subclavian artery

MDCT Evaluation of Congenital Anomalies of Aortic Arch

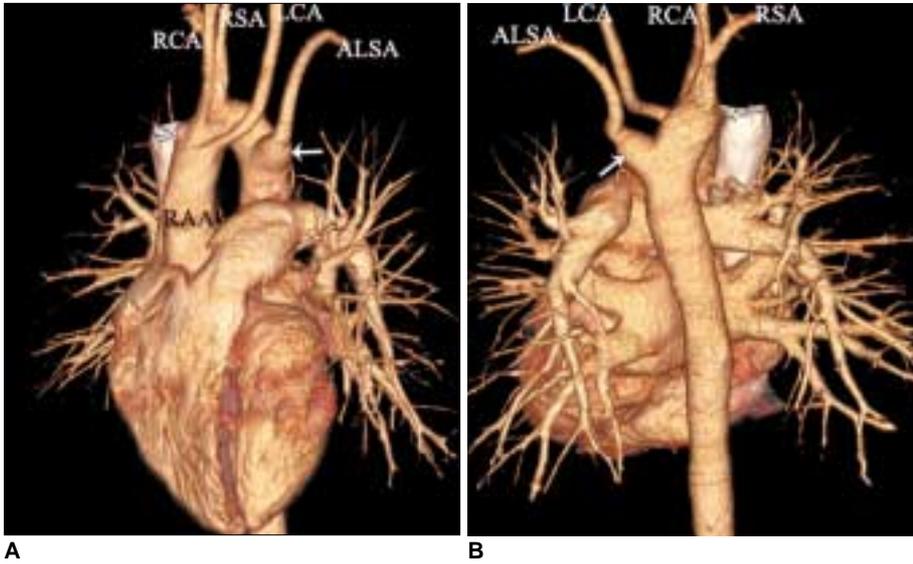


Fig. 9. Right aortic arch with aberrant left subclavian artery in 45-year-old asymptomatic woman is shown. Anterior (A) and posterior (B) volume rendering images show right aortic arch (RAA) with aberrant left subclavian artery (ALSA) associated with Kommerell's diverticulum (arrows) at its origin. First branch arising from aortic arch is left carotid artery (LCA), which is followed by right carotid artery (RCA), right subclavian artery (RSA) and ALSA.

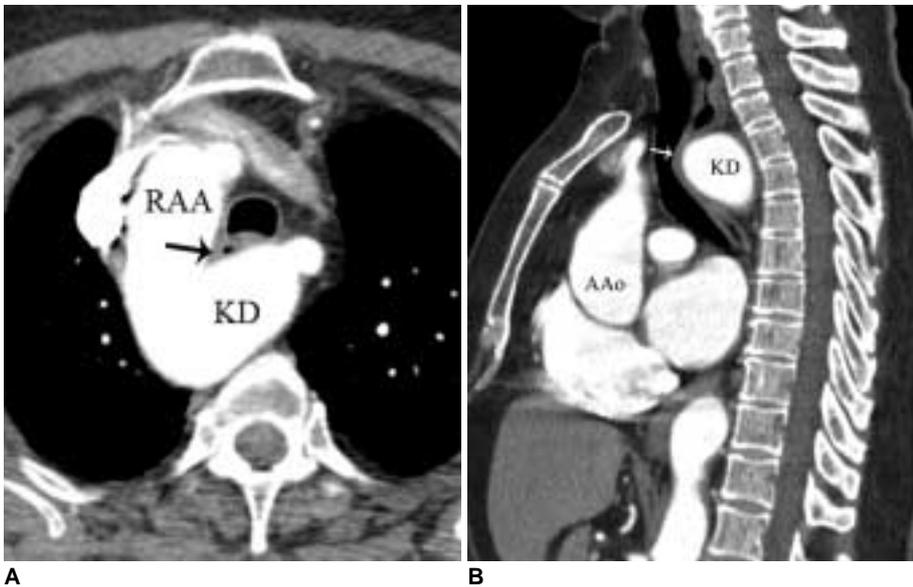


Fig. 10. Right aortic arch (RAA) with aberrant left subclavian artery in 63-year-old woman without dysphagia. Axial (A) and sagittal multiplanar reformatted (B) images show aneurysmal Kommerell's diverticulum (KD) with 3.2 cm diameter causing esophageal compression (arrows). AAo = ascending aorta

descending aorta, with regression of the right ductus arteriosus in the hypothetical double aortic arch (Fig. 12). The left innominate artery is the first branch arising from the arch, which is followed by the right carotid artery and right subclavian arteries (Fig. 13). This anomaly is usually associated with cyanotic congenital heart disease, especially tetralogy of Fallot and truncus arteriosus (9).

An RAA with isolation of the LSA is the rarest type (5). This anomaly results from interruption of the left arch at two levels, with one level between the left common carotid and left subclavian arteries and the other level distal to the attachment of the left ductus (Fig. 14). The left carotid artery arises as the first branch of the right arch, followed by the right carotid artery and right subclavian arteries. The LSA does not have a connection with the aorta, but is



Fig. 11. Right aortic arch (RAA) with aberrant left subclavian artery in 52-year-old woman with dysphagia is presented. Axial image shows aneurysmal Kommerell's diverticulum (KD) with 4 cm diameter.

connected to the pulmonary artery by a left ductus arteriosus. This rare anomaly may cause congenital subclavian steal syndrome and vertebrobasilar insufficiency. Rarely, the anomaly is associated with congenital heart disease, especially tetralogy of Fallot (10).

Double Aortic Arch

In a double aortic arch, the ascending aorta divides into two separate arches. The left or anterior arch has a course similar to that of the normal LAA, and the right or posterior arch has a course to the left behind the esophagus

and joins the left arch. The descending aorta is usually on the left side. The subclavian and carotid arteries arise from their respective arches. The right and left arches may be symmetric, but usually the right arch is larger, extends more cephalad and is more posterior than the left arch (Fig. 15). This anomaly is most common cause of a complete vascular ring (5). Clinically, the onset and severity of symptoms of esophageal and tracheal obstruction are variable and depend on the tightness of the ring. Most patients present with stridor, recurrent respiratory infections or dysphagia within the first six months of life.

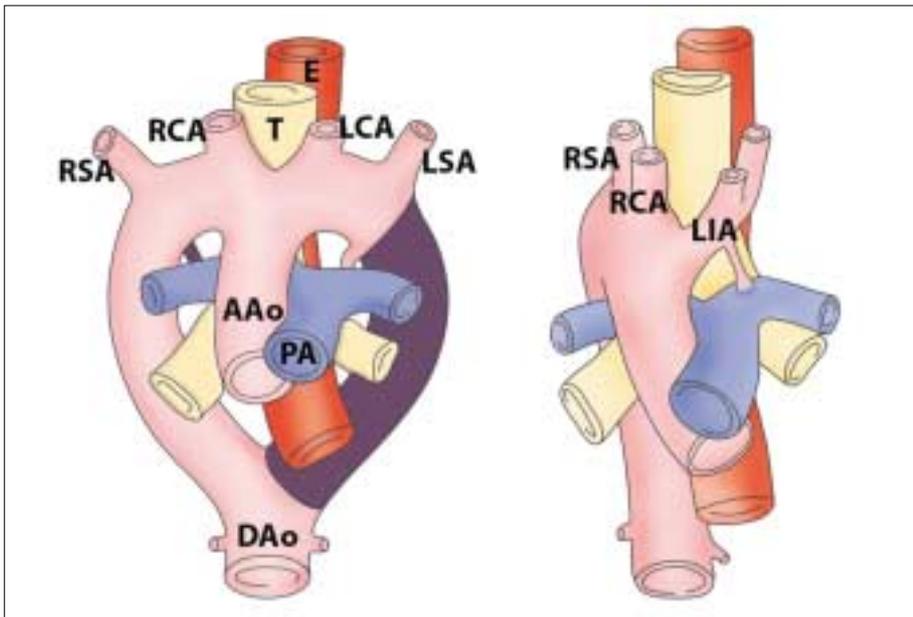


Fig. 12. Diagram of development of right aortic arch with mirror image branching. Anomaly results from interruption of dorsal segment of left arch between left subclavian artery and descending aorta with regression of right ductus arteriosus in hypothetical double aortic arch. AAO = ascending aorta, DAo = descending aorta, PA = pulmonary artery, T = trachea, E = esophagus, RSA = right subclavian artery, RCA = right carotid artery, LCA = left carotid artery, LSA = left subclavian artery, LIA = left innominate artery

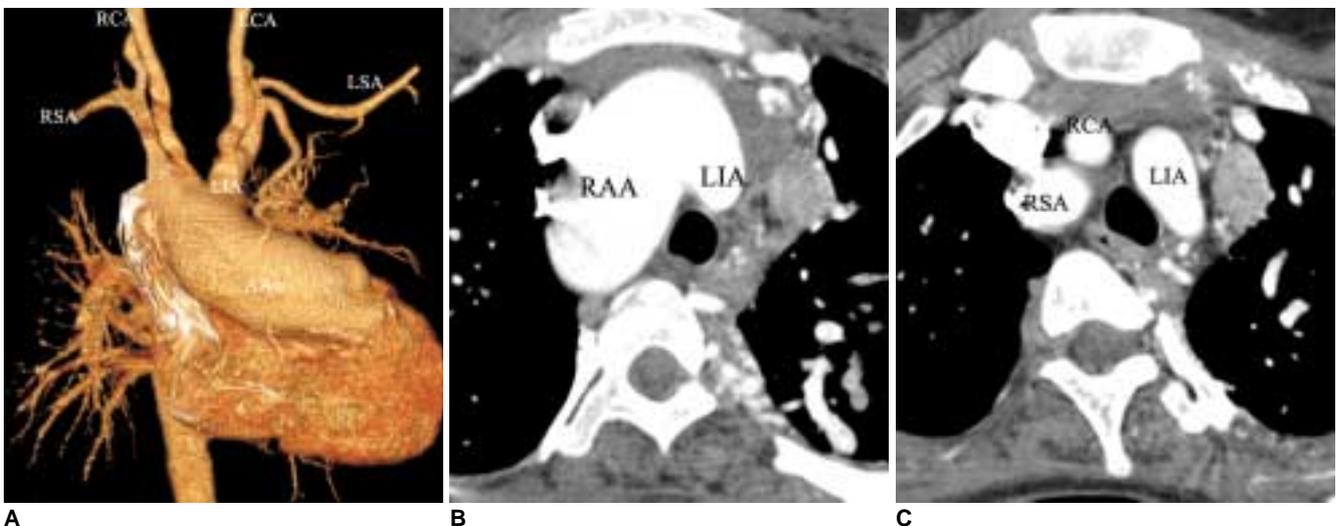


Fig. 13. Right aortic arch with mirror image branching in 20-year-old woman with tetralogy of Fallot, aortopulmonary collateral arteries, partial anomalous pulmonary venous connection, double superior vena cava and double inferior vena cava with azygos continuation. Coronal volume rendering (A) and axial (B, C) images show left innominate artery (LIA) is first branch arising from arch, which is followed by right carotid artery (RCA) and right subclavian artery (RSA). Prominent left internal mammary artery, mediastinal collateral vessels and dilatation of ascending and arcus aorta are also seen. AAO = ascending aorta, LCA = left carotid artery, LSA = left subclavian artery, RAA = right aortic arch

MDCT Evaluation of Congenital Anomalies of Aortic Arch

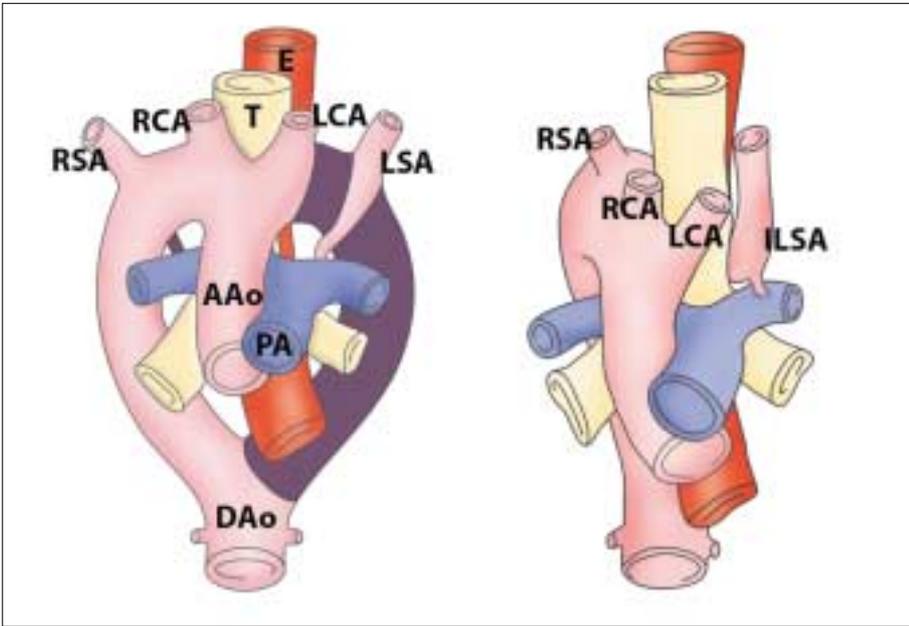


Fig. 14. Diagram of development of right aortic arch with isolated left subclavian artery is shown. Anomaly results from interruption of left arch at two levels; one level is between left common carotid and left subclavian arteries and other level is distal to attachment of left ductus. Left subclavian artery does not have connection with aorta, but is connected to pulmonary artery by left ductus arteriosus. AAO = ascending aorta, DAo = descending aorta, PA = pulmonary artery, T = trachea, E = esophagus, RSA = right subclavian artery, RCA = right carotid artery, LCA = left carotid artery, LSA = left subclavian artery, ILSA = isolated left subclavian artery

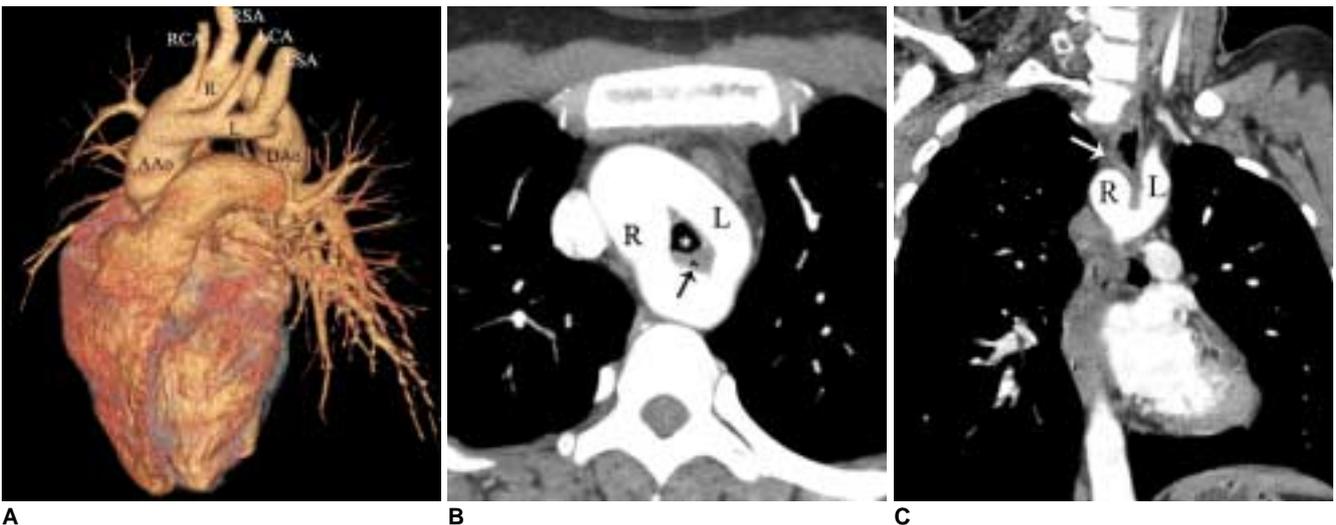


Fig. 15. Double aortic arch in 31-year-old woman presenting with dysphagia is shown. Volume rendering (A) image shows that right arch is larger and extends more cephalad than left arch. Brachiocephalic arteries arise from their respective arches. Axial maximum intensity projection images (B), coronal multiplanar reformatted images (C, D) and volume rendering image with lower opacity values (E) show presence of complete vascular ring causing esophageal (arrows) and tracheal (T) compression. AAO = ascending aorta, DAo = descending aorta, R = right arch, L = left arch, RCA = right carotid artery, RSA = right subclavian artery, LCA = left carotid artery, LSA = left subclavian artery

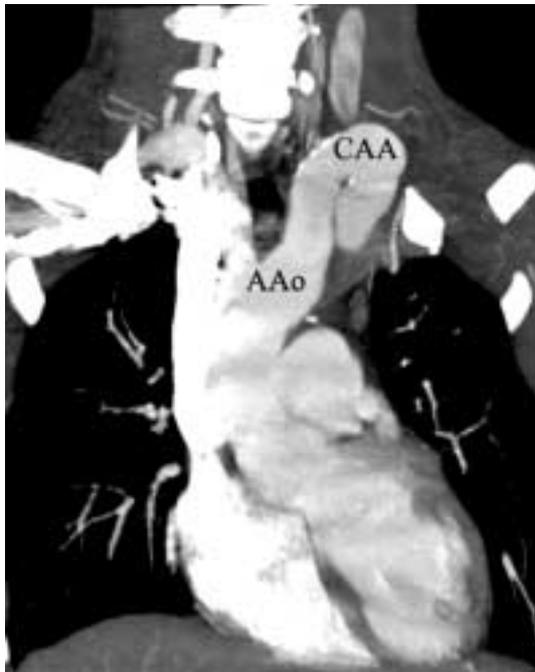


Fig. 16. Left cervical aortic arch (CAA) in 27-year-old-woman presenting with pulsatile mass in left neck. Coronal maximum intensity projection image shows apex of left CAA is above level of clavicle. AAo = ascending aorta

Occasionally, the ring is so loose that is discovered incidentally in asymptomatic adults (1, 8). It is rarely associated with congenital heart disease (5).

Cervical Aortic Arch

A CAA is an uncommon congenital anomaly with a prevalence of less than 0.01%, in which the aortic arch is situated cervically above the clavicle (Fig. 16). Though this condition is often an isolated anomaly, in some cases it may be associated with other cardiac and aortic abnormalities. A CAA is associated with an aneurysm, more often left sided, in 20% of cases (11).

Interrupted Aortic Arch

An IAA is rare congenital anomaly and is characterized by the separation between the ascending and descending aorta, and is differentiated from severe coarctation and aortic atresia by the absence of any structural connection between the ascending and descending aorta. An IAA is classified based on the site of interruption relative to the brachiocephalic arteries: type A occurs distal to the LSA, type B occurs between the LSA and the left carotid artery and type C occurs between the left carotid artery and the innominate artery. Each of these three types is further subdivided as follows: subtype 1, a normal subclavian artery; subtype 2, an aberrant subclavian artery; subtype 3, an isolated subclavian artery that arises from the ipsilat-

eral pulmonary artery by way of the ductus arteriosus. An IAA is associated with additional cardiovascular anatomic defects in up to 98% of cases (12, 13).

In conclusion, aortic arch anomalies may be associated with other developmental cardiovascular defects and may cause symptomatic esophageal or tracheal compression. MDCT is able to display the detailed anatomy of the vascular structures and their spatial relationships to adjacent organs. This ability, combined with the availability of various post-processing options such as VR, MIP and MPR that are applicable to all structures in the scanned volume, give MDCT a significant advantage in comparison to the use of other imaging modalities in the evaluation of aortic arch anomalies.

References

1. Grathwohl KW, Afifi AY, Dillard TA, Olson JP, Heric BR. Vascular rings of the thoracic aorta in adults. *Am J Surg* 1999;65:1077-1083
2. Lee EY, Siegel MJ, Hildebolt CF, Gutierrez FR, Bhalla S, Fallah JH. MDCT evaluation of thoracic aortic anomalies in pediatric patients and young adults: comparison of axial, multiplanar, and 3D images. *AJR Am J Roentgenol* 2004;182:777-784
3. Gilkeson RC, Ciancibello L, Zahka K. Multidetector CT evaluation of congenital heart disease in pediatric and adult patients. *AJR Am J Roentgenol* 2003;180:973-980
4. Edwards JE. Anomalies of the derivatives of the aortic arch system. *Med Clin North Am* 1948;32:925-949
5. Stewart JR, Kincaid OW, Edwards JE. *An atlas of vascular rings and related malformations of the aortic arch system*. Illinois: Springfield, 1964:12-131
6. Fisher RG, Whigham CJ, Trinh C. Diverticula of Kommerell and aberrant subclavian arteries complicated by aneurysms. *Cardiovasc Intervent Radiol* 2005;28:553-560
7. Tsai IC, Tzeng WS, Lee T, Jan SL, Fu YC, Chen MC, et al. Vertebral and carotid artery anomalies in patients with aberrant right subclavian arteries. *Pediatr Radiol* 2007;37:1007-1012
8. Backer CL, Ilbawi MN, Idriss FS, DeLeon SY. Vascular anomalies causing tracheoesophageal compression. Review of experience in children. *J Thorac Cardiovasc Surg* 1989;97:725-731
9. Hastreiter AR, D'Cruz IA, Cantez T, Namin EP, Licata R. Right-sided aorta. Part I. Occurrence of right aortic arch in various types of congenital heart disease. II. Right aortic arch, right descending aorta, and associated anomalies. *Br Heart J* 1966;28:722-725
10. Luetmer PH, Miller GM. Right aortic arch with isolation of the left subclavian artery: case report and review of the literature. *Mayo Clin Proc* 1990;65:407-413
11. Pearson GD, Kan JS, Neill CA, Midgley FM, Gardner TJ, Hougen TJ. Cervical aortic arch with aneurysm formation. *Am J Cardiol* 1997;79:112-114
12. Goo HW, Park IS, Ko JK, Kim YH, Seo DM, Yun TJ, et al. CT of congenital heart disease: normal anatomy and typical pathologic conditions. *Radiographics* 2003;23:S147-S165
13. Dillman JR, Yarram SG, D'Amico AR, Hernandez RJ. Interrupted aortic arch: spectrum of MRI findings. *AJR Am J Roentgenol* 2008;190:1467-1474