Learning objectives

1. To illustrate the embryologic development of the aortic arch and describe the variations of anomalies.

2. To present computed tomography (CT) and magnetic resonance (MR) imaging appearances of the frequently encountered anomalies and some unusual variants.
Background

Congenital anomalies of the aortic arch, which take many forms, result from aberrant development of one or more components of the embryonic pharyngeal arch system. There are five primary groups of anatomic arch anomalies: double aortic arch, right aortic arch with mirror-image branching, right aortic arch with abnormal branching, left aortic arch with abnormal branching, and cervical aortic arch. Clinically, aortic arch anomalies can be divided into that cause physiologic abnormalities, and those that do not. These anomalies are fairly easy to identify and distinguish from other mediastinal pathologies. However, the classification of the rare and complicated variants can be confusing. Familiarity with these variations is essential for correct interpretation of cross-sectional images and to avoid erroneous diagnosis.
Imaging findings OR Procedure details

In early human embryo, six pairs of primitive aortic arches appear in one after another. As some of them regress and the others persist and develop, the mature aortic arch system of fetus is formed 1). It is possible to describe most anomalies of the aortic arch by postulating regression of a segment of the arch that would normally persist or, conversely, persistence of a segment of the arch that would normally regress 2). Within this framework, Edwards proposed a hypothetical double aortic arch with bilateral aortic arches and bilateral ductus arteriosus 3). According to this concept, the anatomy of the normal left arch and each abnormality of the aortic arch are described by persistence or regression of segments of the hypothetical aortic arch (Fig.1).

However, actual developments undergo considerably complicated processes and therefore it is also important to recognize that this hypothesis is imperfect to explain all issues.
Fig.: Edwards' hypothetical double aortic arch system with bilateral aortic arches and bilateral ductus arteriosus. Regression of each segment indicated as dotted line can explain the corresponding configuration of normal or malformed aortic arches. 1: ascending aorta 2: descending aorta 3: right common carotid artery 4: right subclavian artery 5: right innominate artery 6: left common carotid artery 7: left subclavian artery 8: left innominate artery 9: pulmonary artery 10: trachea 11: esophagus 12: ductus arteriosus 13: aortic diverticulum (Kommerell's diverticulum)

References: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

Fig.1: Edwards' hypothetical double aortic arch system with bilateral aortic arches and bilateral ductus arteriosus. Regression of each segment indicated as dotted line can explain the corresponding configuration of normal or malformed aortic arches.


# DOUBLE AORTIC ARCH

This anomaly represents persistence of both the right and left aortic arches and results from no regression of segment of Edwards' schema (Fig.2). It is the most common cause of a symptomatic vascular ring; the trachea and esophagus are encircled and compressed by the two arches 4).

Clinical manifestations, usually dating from birth, include wheezing, stridulous breathing that is constant but exacerbated by crying, tachypnea or cyanosis, and dysphagia in infants and children—though these symptoms may become lighter with growth because of increase of tracheal diameter.

In the usual courses, the right arch is higher and larger than the left arch and the descending aorta is left-sided. Chest radiographs show deviation of the trachea to the left, and may show distal narrowing.

In some cases, segmental atresia of one arch, usually the left, occurs with fibrous continuity of the interrupted segment. In this type, an aortic diverticulum may be present at upper descending aorta which is more often right-sided. Differentiation of this incomplete double aortic arch with left arch atresia from right aortic arch with mirror-image branching can be difficult. Since double aortic arch (either with or without partial atresia) has bilateral common carotid and subclavian arteries originating from each arch without innominate artery, symmetric appearance of common carotid and subclavian arteries are depicted at the transverse section above the level of these arches. This finding on CT is known as 'four artery sign' and helps the accurate diagnosis 5), 6).
Multidetector CT (MDCT) is especially able to display the detailed anatomy of vascular structures and the spatial relationships with adjacent organs.

Furthermore, MRI can provide noninvasive assessments about patency of these vessels without using contrast material 7).

**Fig. 2**: Double aortic arch in a 83-year-old man A: Chest radiograph shows bilateral aortic arches. The right aortic knob (R) is higher than the left aortic knob (L) and the descending aorta is left-sided (arrow). B/C: Contrast enhanced CT images show the left arch (L) presenting at caudal level of the right arch (R). D: Volume rendered three-dimensional CT image shows hypoplastic left aortic arch (L) and normal-sized right aortic arch (R). Bilateral common carotid and subclavian arteries arise from each arch.

**RCCA**: right common carotid artery **RSCA**: right subclavian artery **LCCA**: left common carotid artery **LSCA**: left subclavian artery

**References**: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

**Fig.2**: Double aortic arch in a 83-year-old man

A: Chest radiograph shows bilateral aortic arches. The right aortic knob (R) is higher than the left aortic knob (L) and the descending aorta is left-sided (arrow).
B/C: Contrast enhanced CT images show the left arch (L) presenting at caudal level of the right arch (R).

D: Volume rendered three-dimensional CT image shows hypoplastic left aortic arch (L) and normal-sized right aortic arch (R). Bilateral common carotid and subclavian arteries arise from each arch.

RCCA: right common carotid artery RSCA: right subclavian artery LCCA: left common carotid artery LSCA: left subclavian artery

#RIGHT AORTIC ARCH

This malformation represents the right aortic arch passing the right side of the trachea, contrary to normal development. There are many complicated subtypes classified according to position of descending aorta or ligamentum arteriosum, or existence of aortic diverticulum, etc (8). We review some variations of them encountered frequently.

(1) Right aortic arch with mirror-image branching

This anomaly results from regression of segment (d') of Edwards’ schema and is commonly associated with cyanotic CHD: tetralogy of Fallot, truncus arteriosus, tricuspid atresia, and transposition of the great arteries with pulmonary valvular stenosis (Fig.3).

After left innominate artery, right common carotid artery followed by right subclavian artery arises from right aortic arch. The right arch passes between the superior vena cava (SVC) and the right side of the trachea and esophagus. It doesn’t create a true vascular ring inasmuch as the ligamentum arteriosum arises from the anteriorly positioned left innominate artery and descends to the left pulmonary artery (4).

Rarely, usually without CHD, a vascular ring may be created if the left ligamentum arteriosum extends from the left pulmonary artery to retroesophageal aortic diverticulum of the right-sided upper descending aorta (Fig.4).

Chest radiographs show right-sided aortic arch at somewhat higher level than the position of the normal left arch. When the aortic knob is unclear, left deviation of the trachea at the arch level may help the diagnosis. Superior Mediastinal widening may appear because of lateral deviation of SVC. CT and MRI can delineate coexisted CHD.
**Fig.**: Right aortic arch in a 10-year-old girl with double outlet right ventricle A: Although the laterality of the arch is not visualized clearly, the tracheal air column is indented on the right side and deviated to the left (arrowhead). B/C: Contrast enhanced CT images. Aortic arch (Ao) is positioned at the right side of the trachea (T) and gives mirror-image branching of a normal left arch. LIA: left innominate artery RCCA: right common carotid artery RSCA: right subclavian artery

**References:** Department of Radiology, Kyorin University School of Medicine - Tokyo/ JP

Fig.3: Right aortic arch in a 10-year-old girl with double outlet right ventricle

A: Although the laterality of the arch is not visualized clearly, the tracheal air column is indented on the right side and deviated to the left (arrowhead).

B/C: Contrast enhanced CT images. Aortic arch (Ao) is positioned at the right side of the trachea (T) and gives mirror-image branching of a normal left arch.

LIA: left innominate artery RCCA: right common carotid artery RSCA: right subclavian artery
**Fig.** Right aortic arch with an aortic diverticulum in a 67-year-old man without CHD  
A: Chest radiograph shows aortic arch likely to be positioned at the right side of the trachea (arrowhead). B,C,D: Contrast enhanced CT images show an aortic diverticulum (Di) protruding behind the trachea (T) and esophagus (E). The mirror-image branching of normal left aortic arch is demonstrated. LIA: left innominate artery LCCA: left common carotid artery LSCA: left subclavian artery SVC: superior vena cava Ao: aortic arch

**References:** Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

Fig.4: Right aortic arch with an aortic diverticulum in a 67-year-old man without CHD

A: Chest radiograph shows aortic arch likely to be positioned at the right side of the trachea (arrowhead).

B,C,D: Contrast enhanced CT images show an aortic diverticulum (Di) protruding behind the trachea (T) and esophagus (E). The mirror-image branching of normal left aortic arch is demonstrated.

LIA: left innominate artery LCCA: left common carotid artery LSCA: left subclavian artery SVC: superior vena cava Ao: aortic arch
(2) Right aortic arch with aberrant left subclavian artery

This anomaly results from regression of segment (b') of Edwards’ schema and is the most common variation of right aortic arch (Fig.5). This type is ordinarily not associated with CHD.

The distal position of the rudimentary left arch often persists as a diverticulum(Kommerell's diverticulum) giving origin of the left subclavian artery.

This anomaly is a common cause of a vascular ring because the ligamentum arteriosum typically attaches to the aortic diverticulum or left subclavian artery 4).

In this anomaly the first branch arising from the aortic arch is the left common carotid artery, which is followed by the right common carotid, right subclavian artery in that order. Finally left subclavian artery arises from aortic diverticulum as the last branch of the arch and passes behind the esophagus from caudal right to cephalad left.

The aortic diverticulum may appear a mass-like shadow at the left side of the trachea and may be mistaken as a mediastinal tumor.

On the esophagogram, the aortic diverticulum and aberrant left subclavian artery may produce an impression running obliquely from caudal right to cephalad left.
**Fig.**: Right aortic arch with aberrant left subclavian artery in a 72-year-old woman.

A: Chest radiograph shows right-sided aortic knob (arrow). There also seems to be a protrusion at the left side of the superior mediastinum (arrowhead).

B, C, D: Contrast enhanced CT images (C: saggital and D: coronal reformatted images) show right-sided aortic arch (Ao) and left aberrant subclavian artery (LSCA) originating from aortic diverticulum (Kommerell's diverticulum; Di) and passing behind the esophagus (E).

T: trachea PA: pulmonary artery

**References**: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

Fig.5: Right aortic arch with aberrant left subclavian artery in a 72-year-old woman

A: Chest radiograph shows right-sided aortic knob (arrow). There also seems to be a protrusion at the left side of the superior mediastinum (arrowhead).

B, C, D: Contrast enhanced CT images (C: saggital and D: coronal reformatted images) show right-sided aortic arch (Ao) and left aberrant subclavian artery (LSCA) originating from aortic diverticulum (Kommerell's diverticulum; Di) and passing behind the esophagus (E).

T: trachea PA: pulmonary artery
(3) Right aortic arch with left descending aorta (circumflex aorta)

It is characterized by a right aortic arch passing behind the esophagus and descending left side of the spine, typically with an aberrant left subclavian artery. Chest radiographs show upper mediastinal widening easily mistaken for a mediastinal tumor or aortic aneurysm. Displacement of the trachea to the left and bilateral symmetric widening of the superior mediastinum should suggest this anomaly 9).

#LEFT AORTIC ARCH

(1) Normal branching

The normal aortic arch develops by regression of segment (d) of Edwards' schema.

The first branch arising from the aortic arch is the right innominate artery, which is followed by the left common carotid, left subclavian artery in that order. There are some variations in which the left common carotid artery arises from or very closely to the right innominate artery, the left vertebral artery arises directly from the aortic arch, etc.

(2) Aberrant origin of the right subclavian artery

This anomaly represents the right subclavian artery arising as the most distal branch of the aortic arch (Fig.6). It results from regression of segment (b) of Edwards' schema and is the most common congenital anomaly of aortic arch occurring in approximately 0.5% of normal autopsies. The vessel runs a typical oblique course retroesophageally from caudal left to cephalad right. It may make esophageal compression and be a cause of dysphagia, though in most cases asymptomatic.
Fig.: 3D MR Angiography of aberrant right subclavian artery in a 82-year-old woman pointed out side-to-side differentiation of blood pressure between upper extremities. Right subclavian artery (RSCA) arises as the last branch of the left aortic arch with stenosis (arrow) at its proximate region.

References: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

Fig.6: 3D MR Angiography of aberrant right subclavian artery in a 82-year-old woman pointed out side-to-side differentiation of blood pressure between upper extremities. Right subclavian artery (RSCA) arises as the last branch of the left aortic arch with stenosis (arrow) at its proximate region.

#CERVICAL AORTIC ARCH

A cervical aortic arch is characterized by an aortic arch extending abnormally high in the upper mediastinum-low cervical region on right or left side. A right cervical arch is more common than a left cervical arch. The branching of the aortic arch vessels is variable 4).

Some hypothetical theories to explain the cause of a cervical aortic arch have been proposed; (1) Failure of descent of the otherwise normal fourth branchial arch, (2)
Development of the third or second branchial arches into the aortic arch, and (3) Fusion of the third and fourth branchial arches on the involved side 10).

Radiographic findings include upper mediastinal widening, tracheal displacement to the contralateral side and anteriorly, a large oblique impression on the esophagogram, and a contralateral-sided descending aorta.

Clinically a pulsatile mass which is easily mistaken for an aneurysm may be present in the supraclavicular region. Operation is indicated in patients with symptomatic vascular ring.
Fig. 0: Edwards' hypothetical double aortic arch system with bilateral aortic arches and bilateral ductus arteriosus. Regression of each segment indicated as dotted line can explain the corresponding configuration of normal or malformed aortic arches. 1: ascending aorta 2: descending aorta 3: right common carotid artery 4: right subclavian artery 5: right innominate artery 6: left common carotid artery 7: left subclavian artery 8: left innominate artery 9: pulmonary artery 10: trachea 11: esophagus 12: ductus arteriosus 13: aortic diverticulum (Kommerell's diverticulum)

© Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
**Fig. 0:** Double aortic arch in a 83-year-old man A: Chest radiograph shows bilateral aortic arches. The right aortic knob (R) is higher than the left aortic knob (L) and the descending aorta is left-sided (arrow). B/C: Contrast enhanced CT images show the left arch (L) presenting at caudal level of the right arch (R). D: Volume rendered three-dimensional CT image shows hypoplastic left aortic arch (L) and normal-sized right aortic arch (R). Bilateral common carotid and subclavian arteries arise from each arch. RCCA: right common carotid artery RSCA: right subclavian artery LCCA: left common carotid artery LSCA: left subclavian artery

© Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
**Fig. 0:** Right aortic arch in a 10-year-old girl with double outlet right ventricle A: Although the laterality of the arch is not visualized clearly, the tracheal air column is indented on the right side and deviated to the left (arrowhead). B/C: Contrast enhanced CT images. Aortic arch (Ao) is positioned at the right side of the trachea (T) and gives mirror-image branching of a normal left arch. LIA: left innominate artery RCCA: right common carotid artery RSCA: right subclavian artery

© Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
Fig. 0: Right aortic arch with an aortic diverticulum in a 67-year-old man without CHD A: Chest radiograph shows aortic arch likely to be positioned at the right side of the trachea (arrowhead). B, C, D: Contrast enhanced CT images show an aortic diverticulum (Di) protruding behind the trachea (T) and esophagus (E). The mirror-image branching of normal left aortic arch is demonstrated. LIA: left innominate artery LCCA: left common carotid artery LSCA: left subclavian artery SVC: superior vena cava Ao: aortic arch

© Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
**Fig. 0:** Right aortic arch with aberrant left subclavian artery in a 72-year-old woman. 
A: Chest radiograph shows right-sided aortic knob (arrow). There also seems to be a protrusion at the left side of the superior mediastinum (arrowhead). B,C,D: Contrast enhanced CT images (C; saggital and D; coronal reformatted images) show right-sided aortic arch (Ao) and left aberrant subclavian artery (LSCA) originating from aortic diverticulum (Kommerell's diverticulum; Di) and passing behind the esophagus (E). 
T: trachea  PA: pulmonary artery

© Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
Fig. 0: 3D MR Angiography of aberrant right subclavian artery in a 82-year-old woman pointed out side-to-side differentiation of blood pressure between upper extremities. Right subclavian artery (RSCA) arises as the last branch of the left aortic arch with stenosis (arrow) at its proximate region.

© Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
Conclusion

The correct identification of aortic arch anomalies is necessary to avoid diagnostic pitfalls and to prevent clinical serious complications. CT and MR imaging has proved to be excellent for evaluating the anomalies of aortic arch. MDCT is especially able to display the detailed anatomy of vascular structures and the spatial relationships with adjacent organs.
References


