Congenital anomalies of the Aortic arch

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Abstract

Aortic arch anomalies refer to congenital abnormalities of the position or branching pattern, or both of the aortic arch. Insight into the hypothetical arch model is crucial to understanding anomalies of the aortic arch in the fetus and recognition of the trachea, three major vessels, ductus arteriosus and descending aorta in the axial views of the upper mediastinum is necessary for a complete fetal cardiac assessment. Clues to aortic arch anomalies include abnormal position of the descending aorta, absence of the normal 'V'-shaped confluence of the ductal and aortic arches, a gap between the ascending aorta and main pulmonary artery in the three-vessel view, and an abnormal vessel behind the trachea with or without a vascular loop or ring around the trachea.

Meticulous attention to anatomic landmarks will lead to successful prenatal diagnosis of important vascular rings making early postnatal management possible.

Keywords: aortic arch, coarctation, right arch

1. Introduction

In early gestation, the aortic arch undergoes complex development [1] that normally results in the formation of a left aortic arch from which three arteries originate: the brachiocephalic artery, bifurcating into the right common carotid and right subclavian arteries, the left common carotid artery and the left subclavian artery (Fig. 1). Development of the aortic arch is abnormal in approximately 1-2% of human fetuses, and may involve complex cardiac defects (e.g., interruption or tubular hypoplasia of the aortic arch, aortic coarctation or double aortic arch) or more subtle findings, classified as normal variants, which are rarely associated with clinical signs in later life (e.g., the right aortic arch either with mirror-image branching of the brachiocephalic arteries or with an aberrant left subclavian artery (ALSA), or left aortic arch with an aberrant right subclavian artery (ARSA)). Some of these findings are associated with an abnormal number of brachiocephalic arteries. Anomalies of the aortic arch may be associated with congenital heart defects, or present as incidental findings in asymptomatic infants. Some anomalies of the aortic arch, particularly those with aberrant branching, may form vascular rings, causing clinical symptoms from tracheal or esophageal compression [1, 2]. Various types of congenital aortic arch abnormalities have been described; however, most have been reported in the pediatric or radiological literature using magnetic resonance imaging (MRI) and spiral computed tomography (CT). [3, 5]

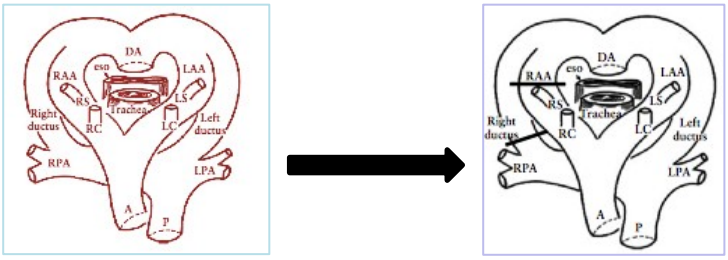


Fig. 1. Hypothetical double aortic arch model of Jesse E. Edwards. A, ascending (ventral) aorta; DA, descending (dorsal) aorta; eso, esophagus; LAA, left aortic arch; LC, left common carotid artery; LPA, left pulmonary artery; LS, left subclavian artery; P, main pulmonary artery; RAA, right aortic arch, RC, right common carotid artery; RPA, right pulmonary artery; RS, right subclavian artery.

2. Fetal Sonographic Approach and Normal Anatomy

The left- or right-sidedness of the aortic arch refers to the position of the aortic arch relative to the trachea. It does not refer to which side of the midline the aorta ascends [6]. The left- and right-sidedness of the ductus can also be defined by its position relative to the trachea. As Achiron *et al*., [7] have shown, location of the aortic arch in relation to the trachea is possible because the fetal airway is normally filled with fluid allowing sonographic visualization. It can be best achieved by obtaining the orthogonal transverse views of the upper mediastinum. The examination of the aortic arch should start from the three-vessel view where the ascending and descending aorta and main pulmonary trunk can be identified. Then, the transducer should be moved cephalad along the vertical axis of the fetal thorax until the aortic arch and the ductus arising from the pulmonary trunk form, on the left side of distal trachea, a 'V'- or 'Y'-shaped confluence at the descending aorta, designated by Yagel *et al*., [8] as three vessels and trachea (3VT) view. When the transducer is moved further cephalad, only the aortic archis seen as a sausage-shaped structure on the left side of the trachea. In these views, the descending aorta is located at the left anterior corner of the spine, and there is no vascular structure crossing the midline behind the trachea. Any branch coursing behind the trachea should be considered as an abnormal aberrant branch.

Anatomical delineation of the aortic arch, its branches and ductal arch can be facilitated by using color and power Doppler examination [9]. The position of the aortic arch relative to the trachea can also be evaluated in a slanted coronal plane through the tracheal bifurcation. In this plane the cross-section of the aortic arch is seen on the left side of the trachea above the left main bronchus. The proximal left pulmonary artery or ductus is seen lateral and slightly inferior to the aortic arch. When investigating the aortic arch it is important to obtain an oblique sagittal view of the aortic arch to confirm that it gives rise to the branches to the head and neck. Visualization of the oblique sagittal views of the aortic arch and ductal arch can be facilitated by referencing the three-vessel or 3VT plane [10, 11]. For the aortic arch view, the transducer head is positioned so that the ascending aorta and the descending aorta are aligned with the sonographic beam axis in the three-vessel plane. Then, the transducer is rotated 90 and the aortic arch is seen as a candy cane-shaped structure arising from the center of the mediastinum between the right and left atria and giving rise to the head and neck branches. For the ductal arch view, a three-vessel view is obtained with the pulmonary trunk and the descending aorta aligned with the sonographic beam.

From this position the transducer is rotated 90 and the ductal arch appears as a hockey stick-shaped structure arising from the anterior mediastinum immediately behind the anterior chest wall. (Fig. 2)

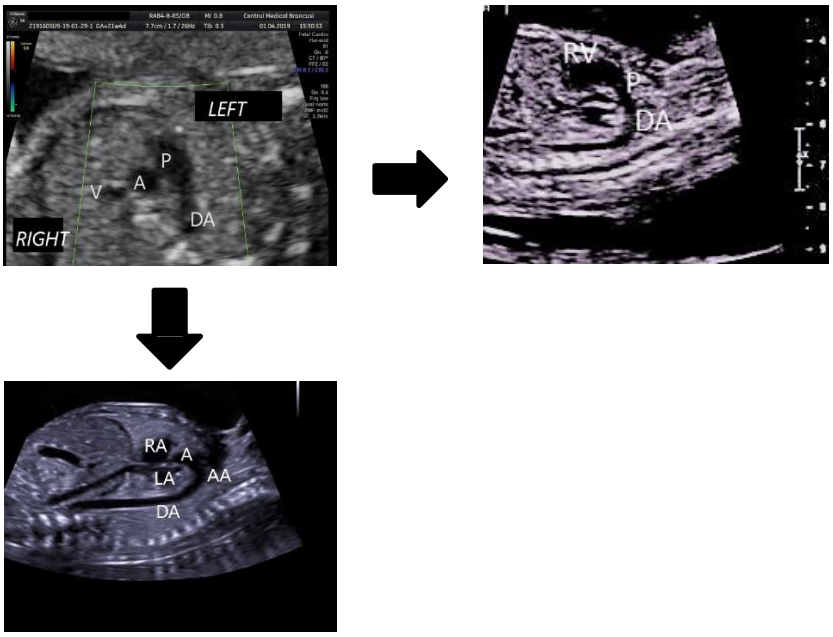


Fig. 2. Maneuvers for visualization of the long-axis views of the aortic and ductal arches. For the aortic arch (AA), a three-vessel view is obtained so that the ascending aorta (A) and descending aorta (DA) are aligned vertically with the sonographic beam axis (left upper panel). Then the transducer is rotated 90. The aortic arch is shown as a candy cane-shaped structure arising from the junction of the right (RA) and left (LA) atria (left lower panel). For the ductal arch, a three-vessel view is obtained so that the main pulmonary artery (P) and descending aorta are aligned vertically with the sonographic beam axis. Then the transducer is rotated 90. The ductal arch is shown as a hockey stick-shaped structure

3. Right Aortic Arch with Mirror-Image Branching

The most common arch anomaly in pediatric patients is a right aortic arch that gives rise to the left innominate, right carotid and right subclavian arteries in sequence, which is a mirror-image of a normal left aortic arch. This variant is formed with regression of the left aortic arch distal to the origin of the left subclavian artery. Conversely, the ductus is not mirror-imaged in most cases.

Usually a left ductus, instead of a right ductus, is patent [12, 14]. The diagnosis of a right aortic arch can be made in a transverse view in which a sausage-shaped aortic arch is located to the right of the trachea (Fig. 3).

The clue to the diagnosis is also present in three-vessel and four-chamber views in which the descending aorta is seen on the right side or in the midline, although this finding can also be seen in double aortic arch and circumflex retroesophageal aortic arch. As the ascending aorta courses to the right in the upper mediastinum, a three-vessel view shows a gap between the ascending aorta and the main pulmonary artery. In contrast to the normal left aortic arch, the right aortic arch does not form a 'V'-shaped confluence with the ductus because the patent ductus is usually a left ductus between the left innominate artery and the left pulmonary artery. Even if the ductus is patent on the right side, the confluence of the ductus and the aortic arch is not in a horizontal plane because the right ductus is located below the aortic arch to connect to the right pulmonary artery.

When the right aortic arch is associated with tetralogy of Fallot, the patent ductus tends to be small and the blood flow through it can be reversed or bidirectional [10]. Therefore, it is often difficult to identify the ductus in this situation.

4. Right Aortic Arch with Aberrant Left Subclavian or Innominate Artery

A right aortic arch can also be formed with regression of the left aortic arch segment between the origins of the left common carotid and subclavian arteries of the double aortic arch model. As a consequence, the right aortic arch gives rise to the left common carotid artery as the first branch followed by the right common carotid, right subclavian and left subclavian arteries in sequence.

As the left subclavian artery arises from the descending aorta and courses leftward behind the trachea and esophagus, it is called an aberrant retroesophageal left subclavian artery. Rarely, the left aortic arch is interrupted proximal to the origin of the left common carotid artery, resulting in a right aortic arch with an aberrant retroesophageal left innominate artery. The reported incidence of association with congenital heart disease varies from 10% to 50% [12, 14].

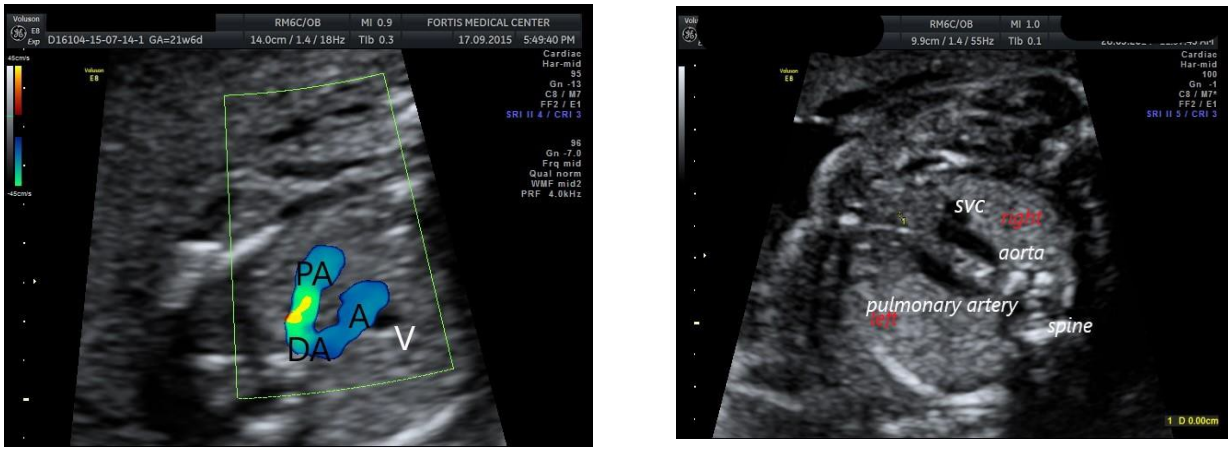


Fig. 3. Right aortic arch (A). Transverse sonogram through the aortic arch shows that it is on the right side of the trache. Three-vessel view shows that the descending aorta is on the right side, SVC superior vena cava

In most cases with a right aortic arch with an aberrant left subclavian or innominate artery, the ductus persists on the left side between the aberrant artery and the left pulmonary artery. In this setting, the ascending aorta,right aortic arch, aberrant left subclavian or innominate artery, left ductus and pulmonary arterial trunk,together with the heart,form a vascular ring around the trachea and esophagus. On transverse view of the aortic arch, the aortic arch, proximal part of the aberrant left subclavian artery, left-sided ductus and pulmonary artery forma 'U'-shaped vascular loop around the trachea. In a coronal view of the trachea and bronchi, the aortic arch is seen on the right side of the trachea, and the ductus on the left side, which is not different from the finding seen with double aortic arch . In fetal life, most of the main pulmonary arterial blood flow passes into the descending aorta through the ductus. When a right aortic arch is associated with an aberrant subclavian or innominate artery and a left ductus, the proximal part of the aberrant artery carries the bloodflow from the ductus into the descending aorta. Therefore, the proximal part of the aberrant artery is as wide as the ductus and descending aorta, and the blood flow through it is reversed. With closure of the ductus immediately after birth, the aberrant artery loses its blood flow from the pulmonary artery and gets blood flow from the descending aorta. The proximal part of the aberrant left subclavian or innominate artery that carried the ductal blood flow to the descending aorta remains dilated as a diverticular pouch. This dilated segment is known as the aortic diverticulum of Kommerell [6, 12]. Persistence of the right ductus is uncommon when the right aortic arch occurs with an aberrant branch. In this combination, a sling or an incomplete

vascular ring is formed around the trachea and esophagus. In this setting, the diameter of the aberrant left subclavian or innominate artery is uniform throughout.

5. Left Aortic Arch with Aberrant Right Subclavian or Innominate Artery

This type is a mirror-image pattern of the right aortic arch with aberrant left subclavian or innominate. It is formed by regression of the right aortic arch segment between the origins of the right common carotid and subclavian arteries of the double aortic arch model. The consequence is the abnormal sequence of the head and neck branches; the right common carotid being the first branch followed by the left common carotid,left subclavian and aberrant retroesophageal right subclavian arteries in sequence. Rarely, the right aortic arch is interrupted proximal to the origin of the right common carotid artery, resulting in a left aortic arch with an aberrant retroesophageal right innominate artery. Association with congenital heart disease is common, especially in Down syndrome cases [12, 14]

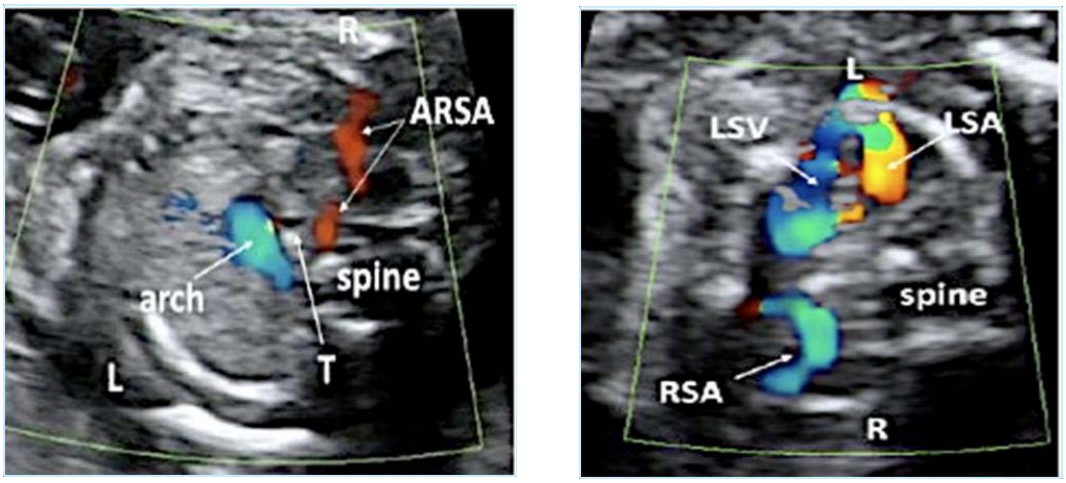


Fig. 4. Color Doppler of the upper mediastinum with ARSA going behind the trachea (left) comparative with the normal course of LSA (right)

The ductus usually persists on the left side [12, 14]. Uncommonly, the right ductus persists between the aberrant right subclavian or innominate artery and the right pulmonaryartery, forming a complete vascular ring. In this situation, the proximal part of the aberrant artery is a wide channel carrying the blood from the ductus into the descending aorta. After birth, with closure of the ductus, the dilated proximal part persists as the aortic diverticulum of Kommerell.

6. Double Aortic Arch

Double aortic arch represents a persistence of both right and left fourth aortic arches that form a complete vascular ring around the trachea and esophagus . Although the aortic arches may be symmetrical, one arch is usually larger and higher than the other, the right arch being the larger one in approximately 75% of cases [12, 14]. Rarely, one arch is atretic. The common carotid and subclavian arteries arise separately from each arch and are usually symmetrically arranged. In most cases, only one ductus is patent, the left ductus being patent in the majority of cases. Almost always, the descending aorta is deviated to one side or another, usually to the side of the patent ductus. Double aortic arch is associated with congenital heart disease in approximately 20% of cases [13]. On transverse view, the vascular ring of double aortic arch can be seen to encircle the

fluid-filled trachea. When one arch is significantly higher than the other, it is necessary to tilt the transducer to one side or another to visualize both arches. The vascular ring and ductus can be imaged in a single imaging plane, giving rise to an appearance of a figure of '6' or '9'. When one of the two arches is atretic it is almost impossible to differentiate it from a unilateral arch with an abnormal branching. In coronal view of the trachea and bronchi, cross-sections of the aortic arches are seen on both sides of the trachea. However, the coronal view of the double aortic arch is not distinguishable from that of right or left arch with an aberrant artery and a patent ductus on the side of the aberrant artery.

7. Coarctation of the Aorta

Coarctation of the aorta is a common congenital heart defect. It accounts for approximately 8% of cardiac defects. Coarctation of the aorta is characterized by narrowing of the distal aortic arch.

This obstructive lesion may reduce the blood flow in the fetal aortic arch, leading to arch hypoplasia, although in some cases this may only be clinically evident after birth, or even in later life.

It remains one of the most difficult cardiac defects to diagnose before birth. Antenatal diagnosis of coarctation is critically important for early treatment of the neonate. Suspicion is usually raised when there is a ventricular disproportion, with a disproportionately smaller left ventricle than right ventricle (Fig. 6). But a discrepant ventricular size has only a moderate sensitivity and a low specificity and low positive predictive value for the diagnosis. Other measurements such as the isthmus diameter, ductal diameter, isthmus/ductal ratio, z-scores derived from measurements of the distal aortic isthmus and arterial duct, the presence of a shelf and flow obstruction over the isthmus in a sagittal view can be of aid in the diagnosis of coarctation. (Fig. 5)

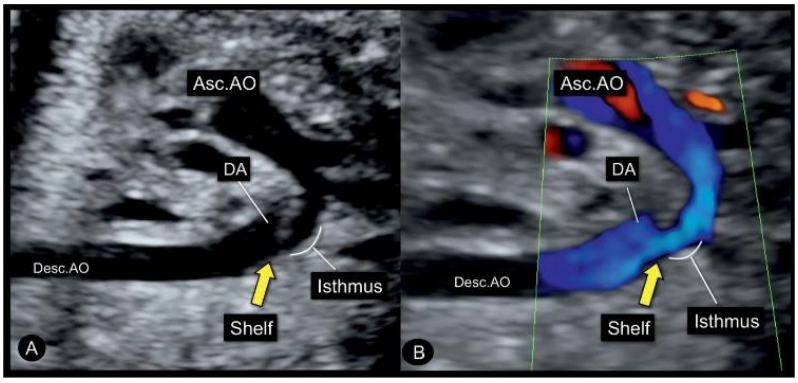
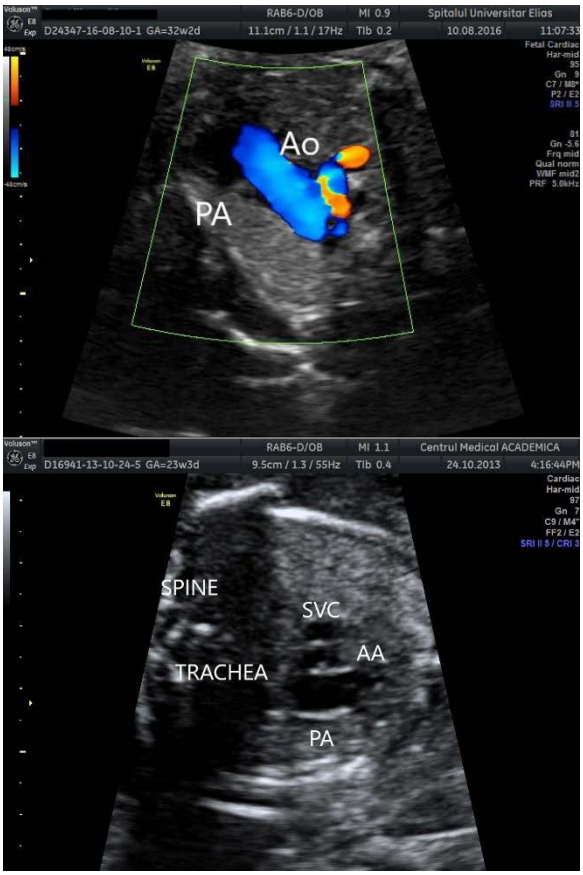


Fig. 5. A. Sagittal view of the aortic arch with the presence of a shelf.
B. Color Doppler of the isthmus in a sagittal view showing flow obstruction

Suspicion for coarctation of the aorta is usually raised when there is ventricular disproportion in fetal life (with a smaller left than right ventricle).



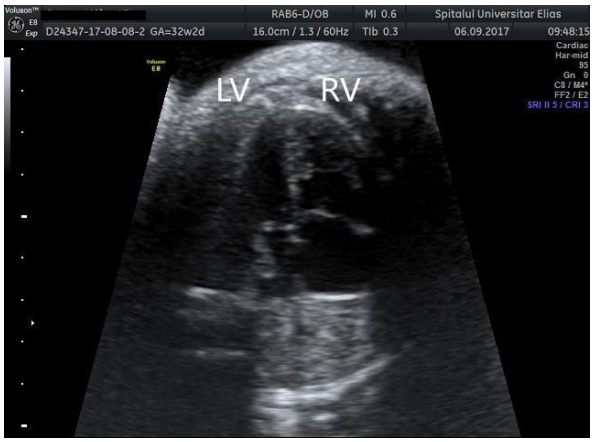


Fig. 6. 3V trachea view with Color Doppler –reversed flow in the aortic arch (left upper corner). Ventricular disproportion with the prevalence of the right ventricle – four chamber view (right upper corner). Disproportion of the ductal/isthmal ratio

In 1997 Brown *et al*., have already demonstrated a moderate sensitivity (62%) and a mediocre positive predictive value for coarctation (33%) for various forms of left-sided structural heart disease. A ventricular disproportion is the most sensitive in the second trimester, before 25 weeks of gestational age, and less in the third trimester. In the third trimester there is already a slight degree of fysiological disproportion (normal: LV/RV < 1.5). There is a high false positive rate, especially after 34 weeks (up to 80%). False-positive diagnosis can result in parental anxiety and the differential diagnosis of isolated 4-chamber cardiac disproportion is also wide.

8. Summary

Axial views of the upper fetal mediastinum and recognition of the trachea, three major vessels, ductus and descending aorta are necessary steps of a complete fetal cardiac assessment. Clues to arch anomalies include abnormal position of the descending aorta, absence of the normal 'V'-shaped confluence of the ductal and aortic arches, a gap between the ascending aorta and main pulmonary artery in the three-vessel view, and an abnormal vessel behind the trachea with or with out a'U'-, '6'- or '9'-shaped vascular loop or ring around the trachea.

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