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Picture of the Month

Aortic arch with four vessels: aberrant right subclavian artery

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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: 1) the brachiocephalic artery, bifurcating into the right common carotid and right subclavian arteries, 2) the left common carotid artery and 3) the left subclavian artery (Figure 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 (Figure 2).

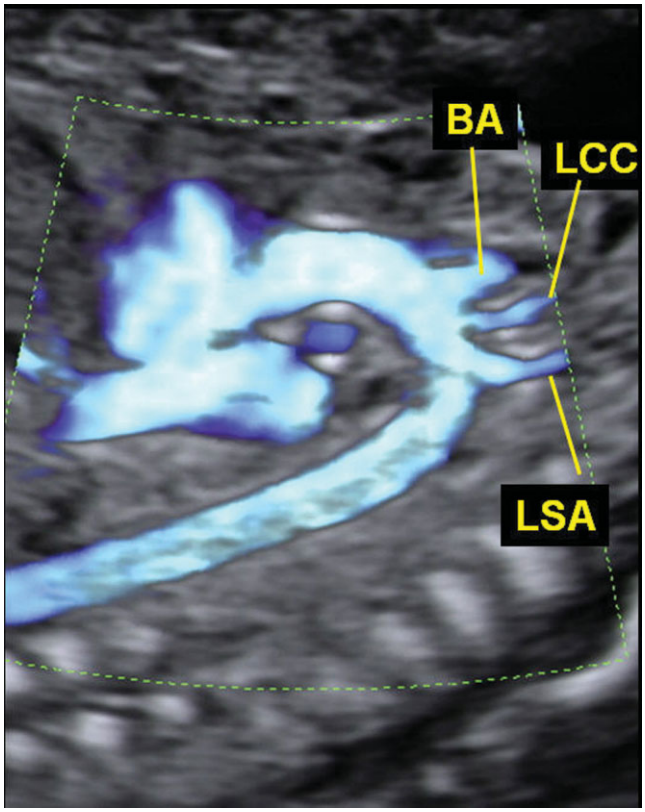

Figure 1 High-Definition® power Doppler image of a normal aortic arch with three vessels originating from it: the brachiocephalic artery (BA), the left common carotid artery (LCC) and the left subclavian artery (LSA).

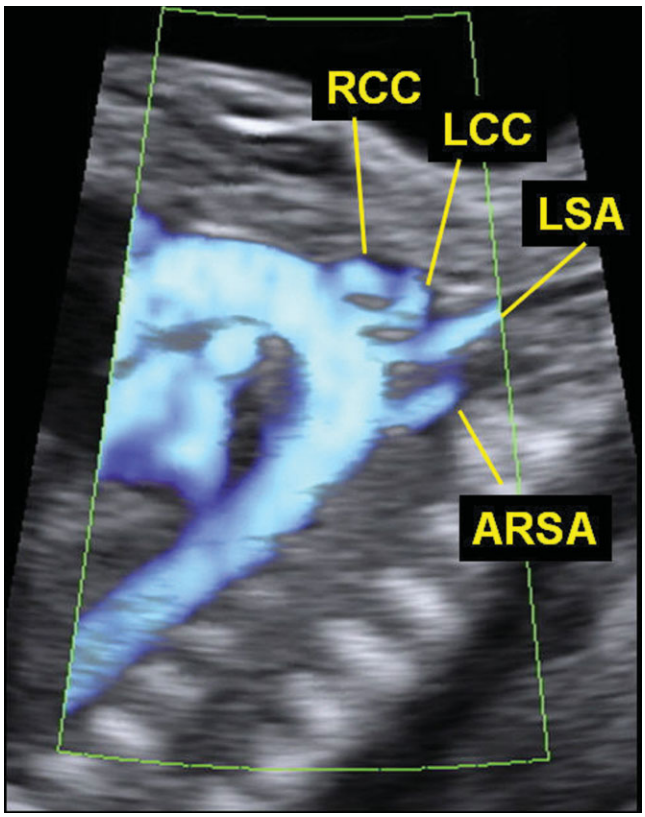

Figure 2 High-Definition® power Doppler image of left aortic with four vessels originating from it: the right common carotid artery (RCC), the left common carotid artery (LCC), the left subclavian artery (LSA) and an aberrant right subclavian artery (ARSA).

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Development of the left aortic arch with an aberrant subclavian artery

In the embryo, a double aortic arch first develops with a left and a right arch connecting the ascending with the neutral-positioned descending aorta, and with the trachea and esophagus situated between them (Figure 3). Each aortic arch gives rise to a common carotid artery and a subclavian artery. Under normal conditions, the right arch distal to the origin of the right subclavian artery regresses (Figure 3b), the right common carotid and subclavian artery merge to form the brachiocephalic artery, while the left aortic arch persists and descends on the left side of the spine. Most anomalies of the aortic arch are assumed to result from the abnormal persistence of parts that should have regressed, or abnormal regression of parts that should have persisted[1].

The most common anomaly of branching of the aortic arch is left aortic arch with ARSA. In this condition, the right aortic arch regresses between rather than distal to the common and right subclavian arteries (Figure 3c), thus hindering their fusion to form the brachiocephalic artery. This results in a left aortic arch giving rise to four arteries (Figures 2 and 3c): the right common carotid, the left common carotid, the left subclavian and the (aberrant) right subclavian arteries. The latter, arising most distally from the aortic arch, has to course from the left side of the spine behind the esophagus and trachea to the right upper arm and is also known as aberrant retroesophageal right subclavian artery or formerly *arteria lusoria*.

If the regression occurs within the left arch, between the common and left subclavian arteries, this results in a right aortic arch also giving rise to four vessels: the left common carotid, the right common carotid, the right subclavian and the (aberrant) left subclavian arteries. This condition has been presented extensively in a previous issue of the Journal[1,2].

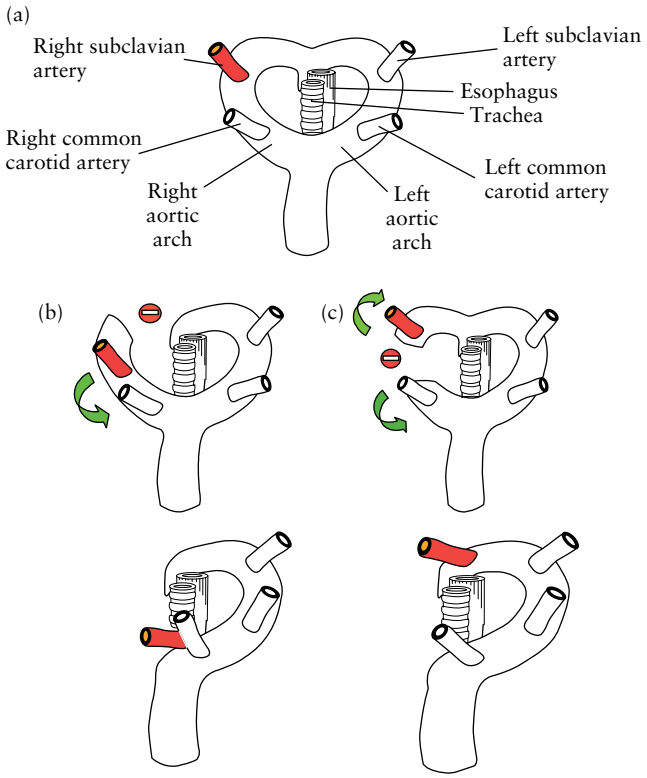

Figure 3 Diagram showing embryological development of a normal left aortic arch and a left aortic arch with an aberrant right subclavian artery (ARSA). The image of a (hypothetical) double aortic arch (a) is helpful for better understanding. The double aortic, with esophagus and trachea between both right and left arches, shows right (RSA, in red) and left (LSA) subclavian arteries as well as right and left common carotid arteries arising from the right aortic arch and the left aortic arch. In order to form the normal left aortic arch (b), the distal part of the right arch regresses and the RSA (red) and right common carotid artery merge to form one artery, the brachiocephalic artery. In (c), the right aortic arch regresses between the common carotid artery and RSA rather than being distal of them, hindering the fusion of these arteries. The left aortic arch can develop, but only the right common carotid artery will arise as the first vessel, while the right subclavian artery will be aberrant and arise distally from the aortic arch, and course behind the esophagus and trachea.

Frequency and clinical significance of aberrant right subclavian artery

The true incidence of ARSA is unknown, but in the general population it seems to range between 0.5 and 1.5%. In a previous preliminary study on 905 fetuses referred for screening, we found the incidence of the ARSA to be 1.4%[3], a frequency recently confirmed by other authors[4]. ARSA may be associated with cardiac anomalies[5], but in the majority of cases it is isolated. In clinical medicine, an aberrant subclavian artery that is located between the esophagus and the spine is reported to occasionally cause pressure on the esophagus and may cause dysphagia. This is probably an extreme and rare clinical symptom, since the high incidence of this vascular variant does not tally with the low incidence of dysphagia leading to medical care. It was reported that care should be taken in patients with ARSA, when transesophageal echocardiography or gastroscopy are performed, to avoid pressure on the retroesophageal vessel which could potentially reduce perfusion of the right arm.

Prenatally, ARSA has been reported to be more frequent in trisomy 21 fetuses[4,6–8] and in fetuses with other chromosomal aberrations, including microdeletion 22q11, than it is in chromosomally normal fetuses[7]. The incidence differs in these groups, since the detection of ARSA in prenatal and in postmortem studies is related to a targeted examination of this vessel, which is not always part of the study protocol. In the first reported prenatal series on trisomy 21 fetuses, we found ARSA in 34% of the 14 fetuses with Down syndrome[6]. In a second study including another 14 cases of Down syndrome, the rate was around 28.5%[7]. We think that in a large number of fetuses with trisomy 21, the rate of ARSA would be around 15–20%, with some having no other cardiac anomaly but additional extracardiac findings. In a French autopsy study on fetuses with trisomy 21 following termination of pregnancy, the incidence of ARSA was found to be 14%[8]. Compared to the rate of 1.4% in the

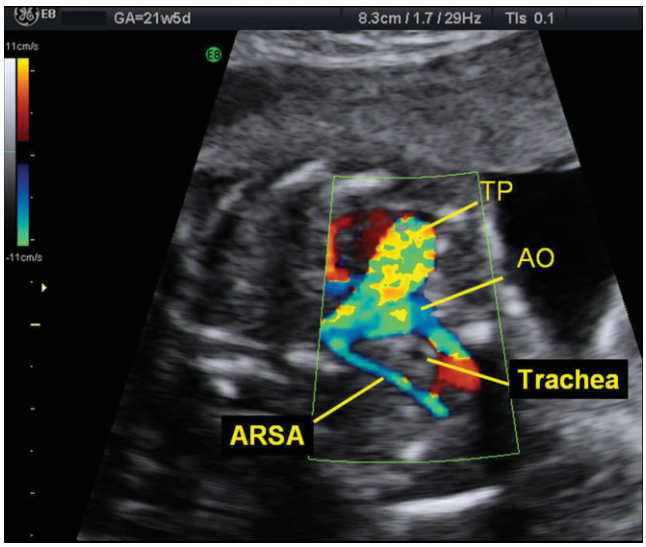

Figure 4 Demonstration of an aberrant right subclavian artery (ARSA) in a cross-sectional plane. The three vessels and trachea view is visualized with a transverse view of the aortic (AO) and pulmonary trunk (TP) arches pointing to the left, with the trachea on their right. The ARSA arises from the distal aortic arch and courses behind the trachea towards the right arm.

normal population, this is at least a 10-fold higher risk. Whether this variant can be used as an isolated marker of a chromosomal aberration or only in combination with additional cardiac and extracardiac markers is yet to be established.

Demonstration of ARSA on prenatal ultrasound

We have reported in detail how to demonstrate or rule out ARSA in the fetus[6], a technique that is feasible as early as the 11–14-week scan and that is easily achieved at the 16-week genetic scan or the 22-week anomaly scan. In the three vessels and trachea view, a vessel leading from the junction of the aortic arch and the ductus arteriosus, behind the trachea and towards the right clavicle and shoulder, is sought by lowering the color Doppler velocity range to 10–15 cm/s (Figure 4). In this plane, one must be careful not to confuse ARSA with the azygos vein, which appears to course in the direction of the superior vena cava. When ARSA is suspected, its presence can be confirmed by demonstrating arterial flow on pulsed Doppler. If the right subclavian artery has a normal origin, it is found in a plane of the transverse aortic arch that is more cranial than the three vessels and trachea view, and it courses ventrally from the trachea[3].

Figure 2 shows that ARSA can be demonstrated in a longitudinal view of the aortic arch, with the four vessels arising from it. To obtain this image, we did not use conventional color images but used the more sensitive bidirectional High Definition (HD) Flow® (Voluson 730, GE Kretztechnik, Zipf, Austria). HD Flow, also known as 'Advanced Dynamic Flow®', is a highly sensitive technique based on power Doppler, but using broad-band digital signals[9]. The presets that enable such an image to be achieved are adjusted to the maximum color filter, a low velocity range (velocities at 7 cm/s) and a very low balance of gray-scale/color.

Theoretically, a longitudinal view of the aortic arch with four vessels can also be demonstrated in the case of a right aortic arch with ALSA, although we have not yet been able to obtain such a view, probably due to the course of the aortic arch.

In some institutions, demonstration of the aortic arch is part of the cardiac examination protocol. We recommend that, in these institutions, attention is paid to the size of the first vessel and that the examiner checks whether three or four vessels arise from the aortic arch, facilitating the detection of an aberrant subclavian artery. Confirmation, however, is best achieved in the suggested three vessels and trachea view (Figure 4).

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