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Picture of the Month

Right aortic arch with vascular ring and aberrant left subclavian artery: prenatal diagnosis assisted by three-dimensional power Doppler ultrasound

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The prenatal detection of a right-sided aortic arch achieved mainly by targeted visualization of the three-vessel and three vessels and trachea (3VT) view, with or without color Doppler, has been discussed recently in this journal[1–4].

Two typical forms of a right aortic arch can be distinguished[5,6]. In one condition a vascular ring is found around the trachea, the so-called U-sign prenatally (Figure 1)[2,3]. The trachea and esophagus are entrapped between the right aortic arch and the left ductus arteriosus and this abnormality is often an isolated incidental finding prenatally[2]. In the other condition, both the aorta and ductus arteriosus lie to the right of the trachea without a vascular ring. This condition is very commonly associated with cardiac anomalies[5].

The branching pattern of the great vessels arising from the aortic arch in both conditions is of major interest in pediatric cardiology[5,6]. The right aortic arch without a vascular ring usually exhibits mirror image branching of the arteries with the left innominate (brachiocephalic) artery arising first followed by the right common carotid and right subclavian artery[6]. By contrast, the right aortic arch with vascular ring very often has an association with an aberrant left subclavian artery. The left common carotid arises first from the aortic arch, followed by the right common carotid, right subclavian artery, and finally a retroesophageal vessel segment from which the left subclavian artery arises and the ductus arteriosus connects. The retroesophageal (and retrotracheal) vessel segment is known as the diverticulum of Kommerell. In other words, the left subclavian artery is connected ventrally to the ductus arteriosus arising from the left pulmonary artery, and dorsally through the Kommerell's diverticulum to the descending aorta. In postnatal life, after closure of the ductus arteriosus, blood enters the left subclavian artery via the descending aorta and Kommerell's diverticulum[4].

Prenatal assessment of a right-sided aortic arch and its branching pattern requires scanning in such planes as a transverse 3VT view (Figure 1), oblique cephalad planes and coronal longitudinal views ventrally from the spine. In the latter planes, the aortic arch lies to the right of the trachea, and the connection with the diverticulum of Kommerell is recognized as a Y-shaped structure dorsal to the trachea (Figure 2)[2,4]. The cephalic vessels arising from the aortic arch can be visualized in more ventral planes, but because of their horizontal course the use of power Doppler ultrasound, which is superior to color Doppler in such insonation conditions, is advised.

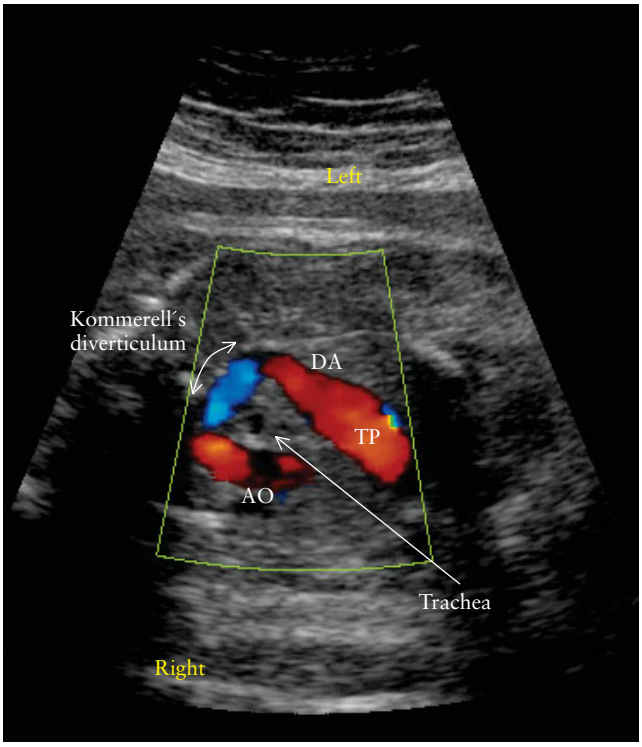

Figure 1 U-Sign in the 27-week fetus visualized from the left side in a cross-section of the three vessels and trachea view. The trachea is situated between the right-sided aortic arch (AO) and the left-sided pulmonary trunk (TP) and ductus arteriosus (DA). The connecting segment represents Kommerell's diverticulum.

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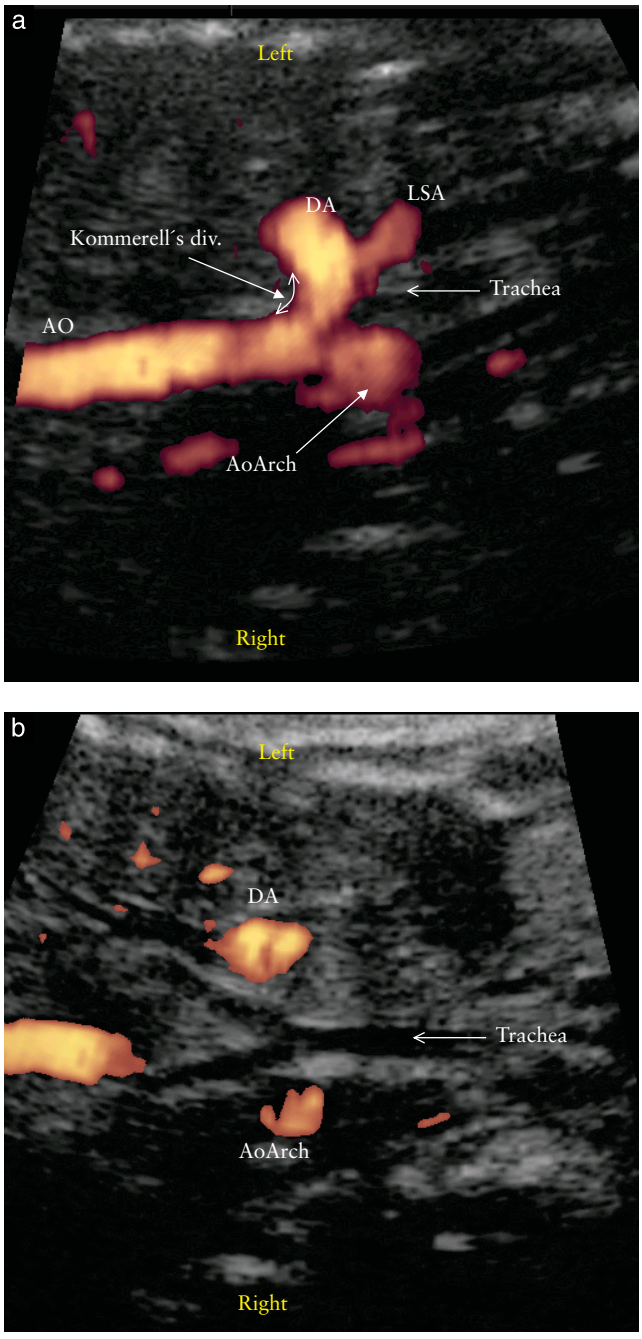

Figure 2 (a) Coronal view with power Doppler of the descending aorta. The aortic arch course is on the right side of the trachea and the descending aorta (AO) is in the middle. The ductus arteriosus (DA) and the aberrant left subclavian artery (LSA) are seen connecting with the descending aorta through Kommerell's diverticulum in a Y-shape. (b) In a more ventral plane the trachea can be seen with its bifurcation. The branching vessels can be followed by tilting the transducer ventrally and dorsally to the trachea. From our experience, this is the best plane to use for the three-dimensional reconstruction.

The Picture of the Month demonstrates a prenatal three-dimensional power Doppler ultrasound (3D-PDU) rendering of a right aortic arch with a vascular ring (around the trachea and esophagus) and the associated aberrant left subclavian artery (Figure 3).

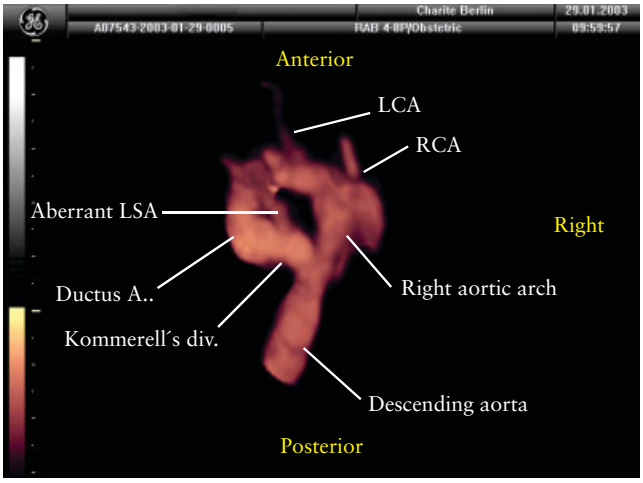

Figure 3 Three-dimensional power Doppler reconstruction of the vascular ring formed by the right-sided aortic arch and the ductus arteriosus with Kommerell's diverticulum as seen from the posterior. The heart was not rendered in this image. Three-dimensional rendering allowed the visualization of some branching arteries such as the aberrant left subclavian artery (LSA) arising at the junction between the ductus arteriosus and Kommerell's diverticulum. The trachea and esophagus are entrapped in the gap between ductus arteriosus and right aortic arch (compare with Figure 1). LCA, left carotid artery; RCA, right carotid artery.

The patient was referred at 23 weeks' gestation because of an abnormal 3VT view. No additional cardiac or extracardiac anomalies were found. Abnormal chromosomes including microdeletion 22q11 were excluded by amniocentesis. The aberrant left subclavian artery was diagnosed by color and power Doppler. 3D-PDU rendering was performed at 35 weeks using commercially available equipment (Voluson 730 Expert, Kretztechnik, Zipf, Austria) with a 4–8-MHz transducer and 3D-static mode combined with power Doppler ultrasound. The examination was performed from the fetal left side after the left aberrant subclavian artery was visualized on two-dimensional power Doppler (Figure 2a).

3D-PDU has been shown to be useful in fetal vascular malformations, but its application for the fetal heart is still considered limited due to lack of cardiac cycle triggering[7]. In a previous issue of this journal we demonstrated the crossing of the great vessels in a normal human fetus using 3D-PDU, explaining how to obtain clear signals without heart rate gating[8]. The example presented with this case emphasizes the possible clinical usefulness of 3D-PDU, in facilitation of the spatial orientation of the aortic arch, pulmonary artery with ductus arteriosus and Kommerell's diverticulum, as well as the demonstration of the origin of the cephalic vessels (Figure 3). Unfortunately, clear demonstration of all cephalic vessels arising from the aortic arch was not possible, the limiting factors being chiefly ribs shadowing as well as the overlapping from neighboring veins when the low-velocities preset is chosen. The trachea and esophagus are not seen on the 3D image but the reader can imagine them within the ring formed by the aortic arch on the right and the

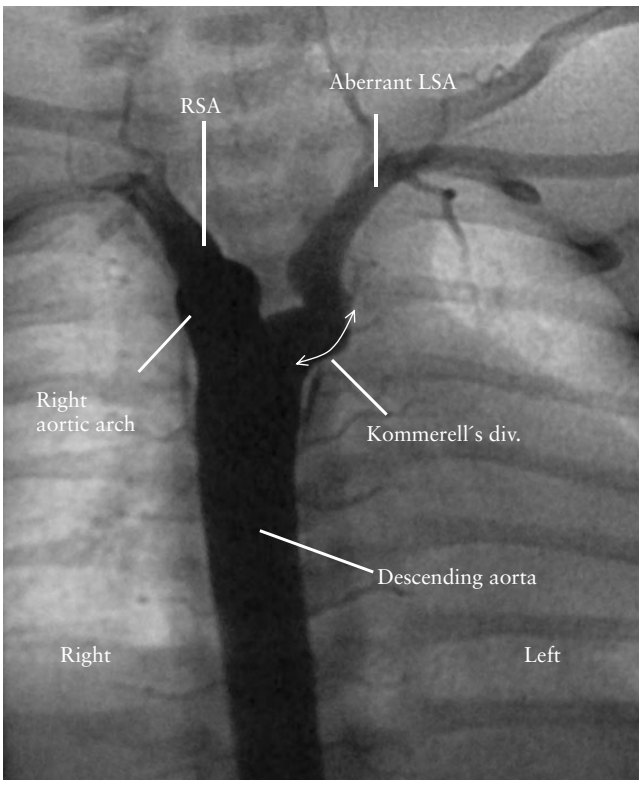

Figure 4 Neonatal X-ray angiography of the aortic arch with descending aorta (in the middle) showing both the right (RSA) and left (LSA) subclavian arteries. The left subclavian artery is perfused from the aorta through the vascular remnant referred to as Kommerell's diverticulum (shaped arrow) after the ductus arteriosus is closed.

ductus arteriosus and diverticulum of Kommerell on the left.

The 3D image was presented to the pediatric cardiologists and neonatologists at our institution and they found the image to be comparable with postnatal 3D images acquired by spiral computed tomography[2,4]. Follow-up after birth was decided upon.

Spontaneous birth occurred at 39 weeks, and echocardiography as well as angiography (Figure 4) confirmed the diagnosis with no additional anomalies. At 6 months of age, no complications were observed, and cardiological and clinical follow-up are planned.

Occasionally, this anomaly may present either with stridor or dysphagia due to the compression of the trachea[9] or esophagus[10]. Left subclavian steal syndrome due to constriction of the aberrant artery may also occur[11]. Achiron *et al.*[2] reported that only 1/18 children with right aortic arch was symptomatic first at the age of 6 months. In a similar case seen recently at our institution, constriction of an aberrant left subclavian artery occurred at 10 days of age, necessitating catheter dilatation and stent insertion.

In summary, 3D-PDU can be utilized in fetal cardiology even without cardiac cycle triggering. The new method provides the clinician with information similar to that of postnatal angiography and facilitates the spatial understanding of the anomaly. Future work will show which further cardiac anomalies are amenable to 3D-PDU rendering.

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