Ultrasound Obstet Gynecol 2005; **25**: 296–298
Published online in Wiley InterScience (www.interscience.wiley.com). **DOI:** 10.1002/uog.1841

Perinatal management of right aortic arch with aberrant left subclavian artery associated with critical stenosis of the subclavian artery in a newborn

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KEYWORDS: aberrant left subclavian artery; ductus arteriosus; perinatal management; prenatal diagnosis; power Doppler; right aortic arch

ABSTRACT

A right-sided aortic arch with an aberrant left subclavian artery is a congenital vascular anomaly that is easily detectable in utero *at the level of the three vessels and trachea view, but which is rarely symptomatic in the neonate. We present a newborn with prenatally diagnosed right-sided aortic arch and aberrant subclavian artery who showed a clinically relevant stenosis of the subclavian artery during the first week of life. An intravascular stent was implanted into the stenosis of the aberrant left subclavian artery by catheterization. This case report demonstrates that a right-sided aortic arch with an aberrant subclavian artery can be diagnosed prenatally, that in these patients a stenosis of the subclavian artery can occur in early infancy and requires awareness of the neonatologist or pediatrician, and that stent implantation represents a minimally invasive therapeutic approach.* Copyright © 2005 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORT

A 31-year-old woman was referred at 23 weeks of gestation due to the abnormal course of the aortic arch detected on routine ultrasound. Targeted fetal echocardiography showed normal four- and five-chamber views. The three vessels and trachea view was abnormal, with the typical U-sign[1], the aorta on the right of the trachea and the ductus arteriosus on the left with a connection behind the trachea (Kommerell's diverticulum)[2]. The ductus arteriosus seemed to be very narrow.

The finding of a right aortic arch with a sling was confirmed at 29 and 34 weeks. An aberrant left subclavian artery arising from the junction of Kommerell's diverticulum and the narrow ductus arteriosus region was visualized on power Doppler ultrasound (Figure 1). Antegrade, non-turbulent flow was demonstrated in the ductus arteriosus and the left subclavian artery. A healthy boy was delivered at 38 weeks.

Postnatal clinical course and diagnostic evaluation

After postnatal adaptation, no difference in peripheral oxygen saturation or blood pressure was found between

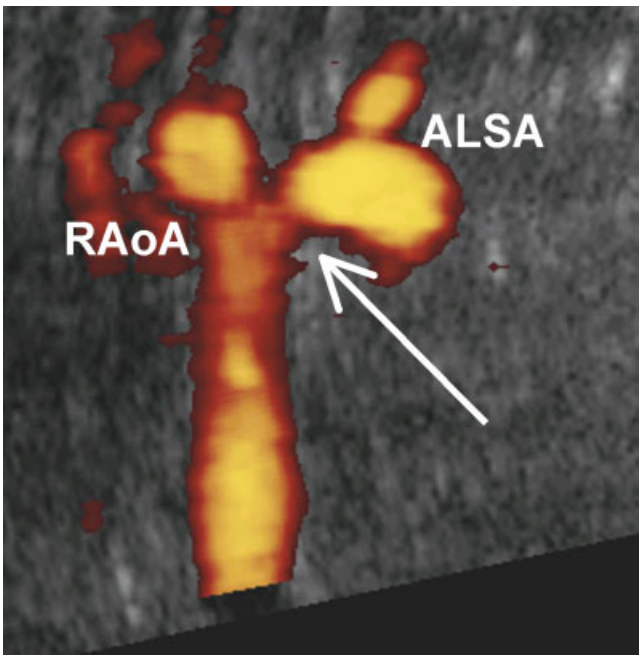

Figure 1 Power Doppler ultrasound image showing the frontal view of the descending aorta with the right aortic arch (RAoA) with Kommerell's diverticulum (arrow) and aberrant left subclavian artery (ALSA). (Same projection as the postnatal angiogram in Figure 2.)

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Accepted: 2 August 2004

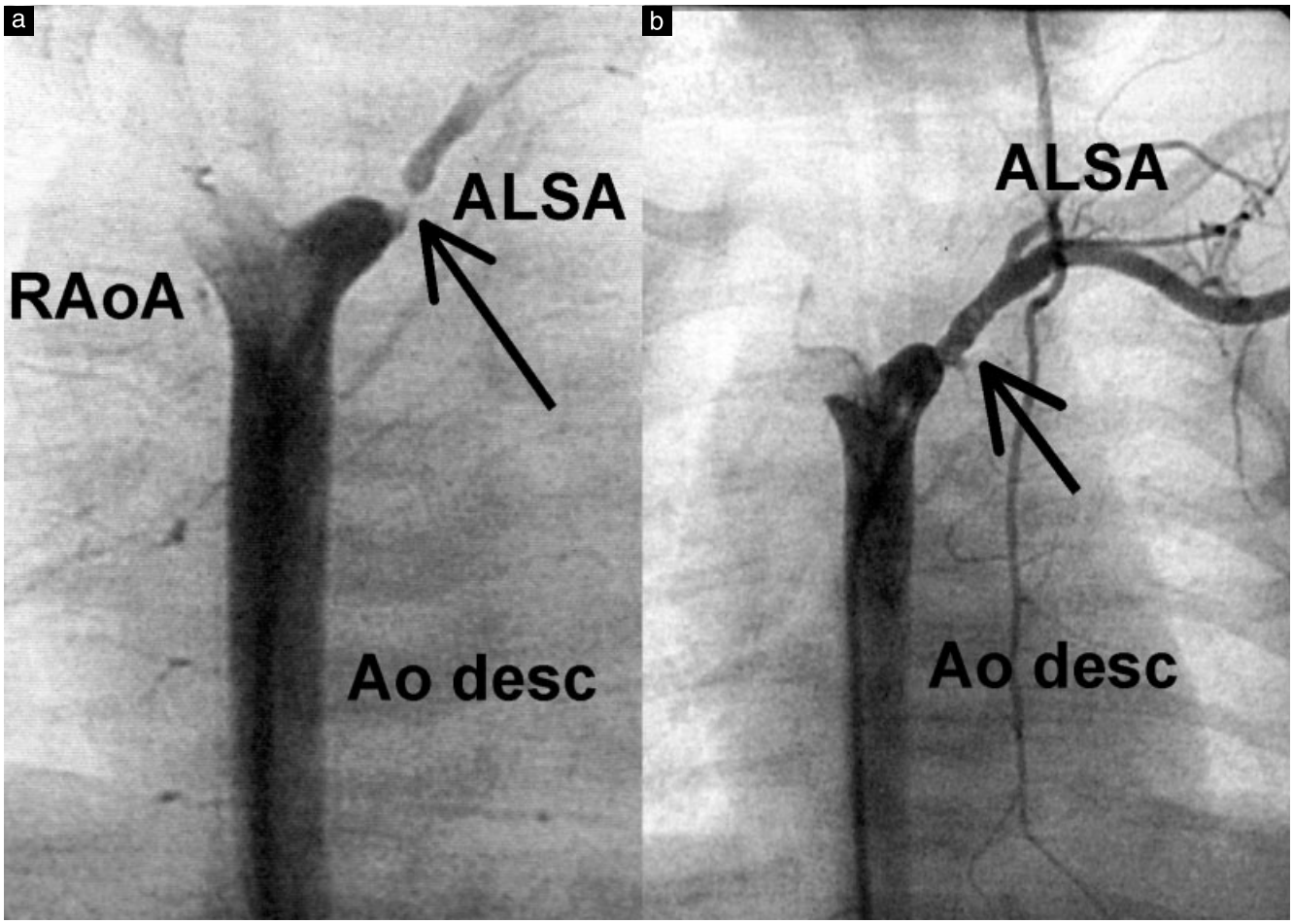

Figure 2 Left ventricular angiograms: (a) confirming the right-sided aortic arch (RAoA) with a high-grade stenosis of the left subclavian artery (ALSA) and poor blood flow past the stenotic segment (arrow); (b) after deployment and expansion of a 2 multi-link pixel stent (8 × 2.5 mm) across the stenotic segment (arrow), fully patent left subclavian artery (ALSA) can be seen with good antegrade flow. Ao desc, descending aorta.

the left and right upper limbs. Two-dimensional echocardiography on day 1 of postnatal life demonstrated a right-sided aortic arch, an aberrant left subclavian artery with normal arterial flow pattern and a non-patent ductus arteriosus.

A difference in pulse quality between the right and left radial arteries increased from the third day onwards with a maximum difference in mean blood pressure of 10 mmHg. Duplex ultrasound revealed a marked decrease in maximum systolic flow in the left subclavian artery (36 cm/s vs. 67 cm/s in the right subclavian artery). The maximum systolic flow in the left axillary artery was decreased to nearly half (23 cm/s vs. 56 cm/s on the right side). Magnetic resonance angiography confirmed a right-sided aortic arch with a median descending aorta, separate origin of right subclavian and right common carotid arteries and a high-grade stenosis of the left subclavian artery near its origin.

Therapeutic management

Percutaneous transluminal angioplasty was performed at 18 days postnatally. A femoral arterial approach was used and a 2 multi-link pixel stent (8 × 2.5 mm) was implanted into the stenosis of the left subclavian artery (Figure 2). The procedure was uncomplicated, and the infant was maintained on low-molecular weight heparin to prevent coagulation and discharged at the age of 25 days.

Follow-up duplex sonography revealed an improvement in the flow pattern in the left subclavian and axillary arteries, but a small stenosis remained with a peak flow of 72 cm/s (vs. 88 cm/s in the right subclavian artery). The maximum systolic flow in the left axillary artery was 60 cm/s (vs. 73 cm/s on the right side).

At the age of 8 weeks the stent was dilated to a diameter of 2.9 mm. On follow-up at 5 and 18 months of age the patient showed good physical and mental development. Blood pressure in both upper limbs was equal (right limb, 103/61 mmHg; left limb, 100/74 mmHg). Duplex sonography revealed a normal arterial flow pattern in the left subclavian artery.

DISCUSSION

A right-sided aortic arch represents a common variant of the thoracic vascular anatomy and occurs in approximately 0.1% of the population[3,4]. A vascular

ring around the trachea can be present and the origin and course of the brachiocephalic vessels varies[5]. Several reports in this journal[2,6,7] have emphasized the prenatal visualization of this condition and have reported good postnatal outcome.

A right-sided aortic arch results from abnormal organogenesis of the primitive aortic arches[6,8]. By definition a right aortic arch crosses the right main stem bronchus whereas the aorta can descend to the right, left or midline. Shuford *et al.*[5] classified right aortic arch into three subgroups: (1) those with mirror-image branching, (2) those with aberrant left subclavian artery, and (3) those with an isolated left subclavian artery.

Our patient had the second form with an aberrant left subclavian artery (Figure 1). If the aberrant left subclavian artery originates from the junction between the distal arch and the descending thoracic aorta, dysphagia, airway compression and aneurysmal complications can occur[9–11] in infancy and adulthood, but complications are rare in the neonatal period. Our patient did not show any signs of airway compression or swallowing difficulties. After spontaneous closure of the ductus arteriosus, clinical signs suggested a constriction of the aberrant left subclavian artery. Subsequent echocardiography demonstrated the stenosis of the left subclavian artery at the origin of the closed ductus arteriosus (Figure 2a).

To our knowledge such a complication has not yet been described in a neonate with a right aortic arch and an aberrant left subclavian artery. However, it is possible that it occurs more often than suspected, with no clinical symptoms being evident at the time of discharge. In these patients the upper arm could be perfused by developing collaterals and signs of a steal syndrome could occur during later life[12–14]. Therefore, careful fetal echocardiography – preferably using three-dimensional power Doppler – is required to detect anomalies of the aortic arch[2]. Furthermore, a prenatal diagnosis requires close follow-up in order to detect not only airway compression syndromes but also stenosis of the subclavian artery early in neonatal life.

Percutaneous transluminal angioplasty was successfully used in our patient to implant a stent into the high-grade stenosis. Until now, management of aberrant subclavian arteries has required surgical reconstruction, since vascular compression is the predominant symptom[15]. Percutaneous techniques are less invasive and carry a lower surgical risk. Thus, stent implantation provides an attractive alternative to conventional surgical strategies, particularly in newborns without vascular compression but with stenosis of the subclavian artery. However, stent administration could be associated with long-term problems such as re-stenosis[16]. Follow-up of our patient at 18 months of age showed good physical and mental development and no symptoms of re-stenosis.

In summary, the case described here demonstrates that: 1) an aberrant subclavian artery in association with a right-sided aortic arch can be prenatally diagnosed; 2) stenosis of the aberrant subclavian artery can develop early in neonatal life and requires an awareness of the neonatologist/pediatrician; 3) percutaneous transluminal angioplasty with stent implantation into the stenotic subclavian artery represents a less invasive therapeutic option than does surgical intervention.

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