Right Aortic Arch and Isolated Left Subclavian Artery: Prenatal and Postnatal Echocardiographic and Tomographic Imaging

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INTRODUCTION

Right aortic arch (RAA) with isolated left subclavian artery (LSCA) is a rare congenital anomaly. In this anomaly, blood flow to the LSCA is commonly supplied by the ipsilateral vertebral artery, which has retrograde flow. It is typically associated with other congenital heart diseases, and clinical manifestation can widely vary.

We present the case of a newborn with RAA and isolated LSCA who underwent sequential prenatal and postnatal imaging, including echocardiography and computed tomographic angiography.

CASE PRESENTATION

A 29-year-old woman, G1P0, was referred to pediatric cardiology for a suspected cardiac anomaly found during a routine obstetric ultrasound examination at 20 weeks of gestation. Initial fetal echocardiography at 24 weeks of gestation showed RAA with bilateral ductus arteriosus and diastolic reversals in the left ductal arch (Figure 1). Follow-up fetal echocardiography was performed at 35 weeks of gestation, with no major changes.

A male newborn weighing 2.95 kg was born at 37 weeks' gestation via spontaneous vaginal delivery, with Apgar scores of 6 and 8 at 1 and 5 min. Vital signs were normal. On physical examination, he presented positive and symmetric femoral pulses, and pre- and postductal oxygen saturation levels were 98%. Transthoracic echocardiography was performed at 7 hours of life, showing RAA, bilateral ductus arteriosus with left-to-right shunt, and an undefined arterial branching pattern (Video 1, Figure 2A). Right and left ventricular sizes and systolic function were normal. All cardiac valves were normal, and there was no pericardial effusion.

Computed tomographic angiography with three-dimensional reconstruction was performed on the second day of life for better understanding of the aortic arch and branching. It confirmed the RAA. No patent ductus arteriosus was present, and an isolated LSCA was noted. The origin of the LSCA did not demonstrate a connection to the aortic arch, but it was seen perfused through the left vertebral artery and a small cervicothoracic collateral in a retrograde manner and a vascular ring was ruled out (Video 2, Figure 2B). A chromosomal microarray was also obtained at birth, with a normal result.

The patient was evaluated again in the pediatric cardiology clinic at 1 month of age. The family reported no symptoms. Physical examination revealed a weaker pulse in the left versus the right arm, and blood pressure could not be obtained from the left arm. However, perfusion was normal, and limb size was equal to the contralateral. After reviewing his case in a multidisciplinary cardiovascular care conference, conservative expectant management with close follow-up was advised.

DISCUSSION

The prevalence of RAA has been estimated at about 0.1% of the population.[1-3] RAA with isolated LSCA is the least common type of RAA, occurring in about 0.8% of RAA cases.[1,4-6] In 59% of isolated LSCA cases, it is associated with other congenital heart lesions, most commonly tetralogy of Fallot, ventricular septal defect, and other conotruncal defects.[5] As with other aortic arch abnormalities, RAA with isolated LSCA has been associated with chromosome 22q11 deletion, even in the absence of other intracardiac anomalies.[7,8]

Embryologically, the occurrence of RAA with isolated LSCA is clearly illustrated by the double aortic arch model, introduced by Edwards[9] in 1948. In this model, the aortic sac is connected to the dorsal aorta at each side via six paired aortic arches. Normally, the first two pairs of aortic arches form part of the vasculature of the face. The third pair of aortic arches forms the common carotid arteries, while the fourth aortic arches give rise to the part of the aorta between the common carotid and subclavian arteries. Each proximal sixth aortic arch forms the right and left pulmonary arteries, while the distal sixth arches form the ductus arteriosus on each side.[2,10] Finally, the seven intersegmental arteries connect the vertebral arteries to the ipsilateral dorsal aorta at each level during development, but only the seventh pair of intersegmental arteries persists at each side as the subclavian arteries. The normal left aortic arch develops from interruption of the dorsal segment of the right arch between the right subclavian artery and the descending aorta with regression of the right ductus arteriosus, along with persistence of the left aortic arch and left ductus arteriosus. Accordingly, RAA with a mirror-image branching pattern occurs when there is involution of the right ductus arteriosus and the left dorsal arch between the LSCA and descending aorta.[2]

In RAA with isolated LSCA, there is involution of the distal left dorsal aorta and the left fourth arch proximal to the origin of the LSCA, leaving the LSCA disconnected from the aortic arch but connected to the left pulmonary artery via the left sixth arch[1,2,11] (Figure 3). Finally, after spontaneous closure of the left ductus arteriosus, the LSCA is isolated and draws its blood supply through collateral flow such as the left

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VIDEO HIGHLIGHTS

Video 1: Suprasternal coronal view on transthoracic echocardiography with color Doppler on the first day of life. Normal pulmonary arteries and bilateral ductus arteriosus with left-to-right shunt.

Video 2: Volume-rendered three-dimensional reconstruction of computed tomographic angiography in the oblique axial plane on the second day of life. Remnants of bilateral ductus arteriosus at the superior aspect of the main pulmonary artery and remnant of the ductal ampulla at the inferior aspect of the aorta. Isolated LSA is supplied by retrograde flow in the vertebral artery.

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vertebral artery. Alternatively, the LSCA may be also supplied by mediastinal or thoracic collateral vessels.[2,12]

Most patients with isolated LSCA are asymptomatic, although symptoms can present during the neonatal period or later in life.[2,12] Patients can present with symptoms related to decreased perfusion of the left arm, such as pain, weakness, paresthesia, coldness,[2] or even underdevelopment of the limb.[12] On the contrary, they can also present with subclavian steal phenomenon, with retrograde runoff of vertebral artery flow, causing vertebrobasilar insufficiency.[1,2,13] This can manifest with vertigo, headache, or syncope. RAA with isolated LSCA can also manifest with pulmonary steal phenomenon, either during the neonatal period, after the pulmonary resistance drops, or later in life, in the presence of a persistent left ductus arteriosus, manifested by both underperfusion of the limb and vertebrobasilar insufficiency.[1,2,14] In their review in 1990, Luetmer and Miller[5] reported that among 30 cases with symptoms, only 17% of patients had presented symptoms of ischemia of the left upper extremity, and 17% had symptoms suggestive of vertebrobasilar insufficiency. The mean age at diagnosis of the symptomatic patients was 37.3 years (range, 22–53 years), with a duration of symptoms before diagnosis from <1 to 11 years, whereas the mean age at diagnosis of the asymptomatic patients was 7.7 years (range, 0–34 years).[5] We hypothesize that this difference was because asymptomatic patients might have been diagnosed either incidentally or by imaging obtained for other anomalies.

Diagnosis of this lesion requires a high index of suspicion when symptoms are present. Also, it should also be considered in asymptomatic patients with differences in pulse intensity and blood pressure between the upper extremities.[2]

As shown in our case, we believe that this lesion could be increasingly found incidentally in asymptomatic patients. Although still rare, isolated LSCA may be in fact underreported.

Echocardiography is currently the preferred initial diagnostic tool. Besides providing detailed information on both intra- and extracardiac anatomy and function, aortic arch sidedness and branching pattern can be assessed using echocardiography, including prenatally. If echocardiography is performed early enough, bilateral patent ductus arteriosus can be seen. However, once these undergo spontaneous closure, their presence cannot be ascertained. Computed tomographic angiography and magnetic resonance imaging can be the second-line imaging modalities to better delineate the course and connections of the LSCA. These are useful even after the closure of ductus arteriosus. The remnants of closed ductus arteriosus are marked by "bird-beaking" on the pulmonary arteries or aorta.

Regarding treatment, if other associated congenital heart lesions need to be surgically addressed, repair is indicated.[1] However, the indication of surgery in asymptomatic patients is controversial. Although different reports have supported surgical repair in asymptomatic patients,[1,12,13] we believe that close monitoring might also be an appropriate strategy, weighing the risk and benefits of surgery in every case.

When indicated, the preferred surgical approach is reimplantation of the subclavian artery, either through reanastomosis or autologous saphenous vein graft placement, to the aortic root[2] or the common carotid artery.[1] The recurrent laryngeal nerve lies in close proximity

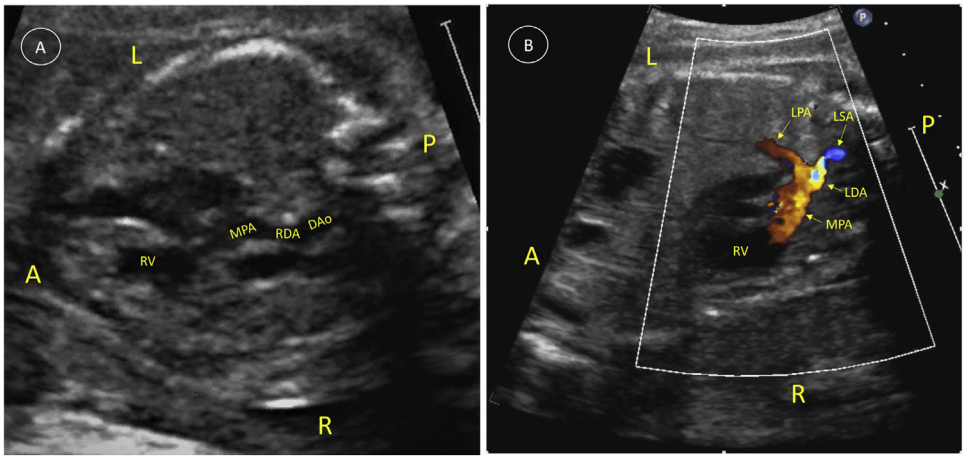

Figure 1 **(A)** Four-chamber view on fetal two-dimensional echocardiography. The main pulmonary artery (MPA) was connected to the descending aorta (DAo) via the right ductus arteriosus (RDA). **(B)** Sagittal view with color Doppler on fetal echocardiography. Normal MPA and left pulmonary artery (LPA), with color aliasing at the left ductus arteriosus (LDA), which is supplying the LSCA. *RV*, Right ventricle.

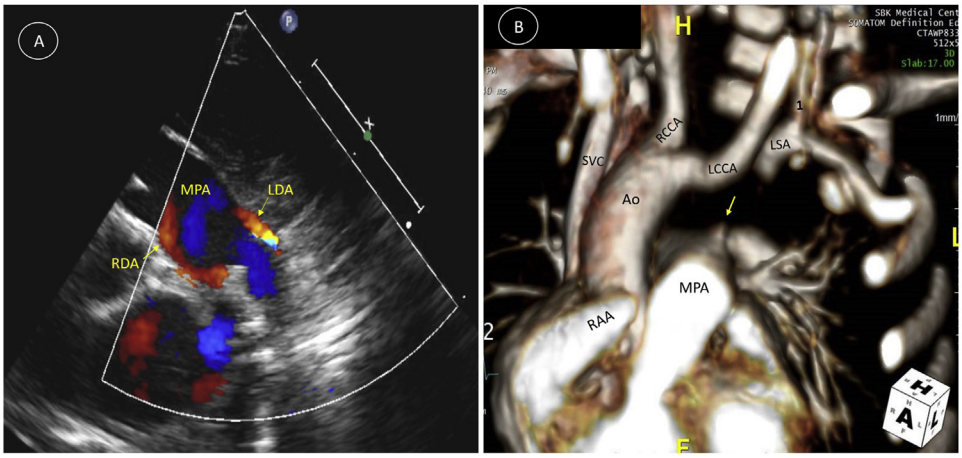

Figure 2 **(A)** Bilateral ductus arteriosus entering the main pulmonary artery (MPA). **(B)** Volume-rendered three-dimensional reconstruction of computed tomography angiography in the oblique coronal plane on the second day of life. The LSCA is supplied by the left vertebral artery (1). The arrow shows the bird-beaking remnant of left ductus arteriosus. *Ao*, Ascending aorta; *LCCA*, left common carotid artery; *LDA*, left ductus arteriosus; *RAA*, right atrial appendage; *RCCA*, right common carotid artery; *SVC*, superior vena cava.

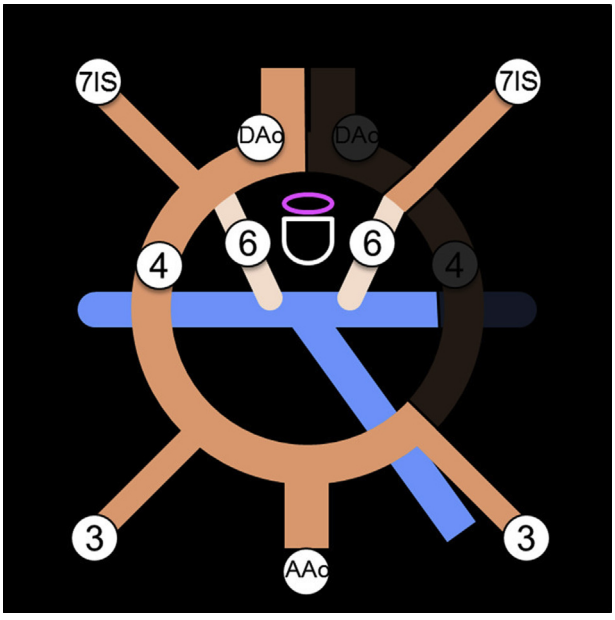

Figure 3 Diagram of theoretical double aortic arch, showing involution of left fourth aortic arch and left descending aorta (DAo), leaving the isolated LSCA supplied by the left ductus arteriosus. *AAo*, Ascending aorta.

to both the subclavian and common carotid arteries, and its careful identification is essential for a successful surgery.[1,13]

CONCLUSION

RAA with isolated LSCA is a rare aortic arch anomaly. Although predominantly asymptomatic, it can manifest at any age with symptoms related to decreased perfusion of the left arm, vertebrobasilar insufficiency, or pulmonary steal. The role of intervention remains case dependent, and in this particular patient without associated congenital cardiac anomalies, the patient has been managed expectantly.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.04.010.

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