Isolation of the left innominate artery: When to operate?

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ABSTRACT

A right aortic arch with an isolated left innominate artery from the pulmonary artery is an exceedingly rare congenital cardiac malformation. We describe the management and complex surgical timing considerations in two such cases, successfully operated on day 4 and 7 months of age, including the use of cranial ultrasound as a helpful tool to guide decision-making. We also describe the first reported association of this defect with a 4q25 deletion encompassing the *LEF1* gene.

Keywords: Cranial ultrasound, isolated innominate artery, *LEF1* gene

INTRODUCTION

A right aortic arch with an isolated left innominate artery from the pulmonary artery is an exceedingly rare congenital cardiac malformation. These patients can be diagnosed antenatally, diagnosed incidentally or present with symptoms of pulmonary over circulation, vertebrobasilar insufficiency, or left arm claudication.[1] The surgical repair is performed by re-implantation of the left innominate artery from the pulmonary artery to the aortic arch; however, there is no universally accepted age to perform this procedure, nor indeed, whether an operation is preferable over conservative management. We describe the management and complex surgical timing considerations in two such cases, successfully operated on day 3 and 7 months of age, including cranial ultrasound as a helpful tool to guide decision-making. We also describe the first reported association of this defect with a 4q25 deletion encompassing the *LEF1* gene.

CLINICAL SUMMARY

Patient 1 was born at 39 weeks with an antenatal diagnosis of a right aortic arch with bilateral arterial ducts; however, given uncertainty about the origin of the left subclavian, they were commenced on prostaglandin-E2 at birth. Their initial postnatal echocardiogram showed an atrial septal defect, small mid-muscular ventricular septal defects (VSD) and a right aortic arch with the left innominate artery arising from the main pulmonary artery through a left-sided arterial duct [Figure 1] which was subsequently confirmed on computed tomography (CT) imaging. A cranial ultrasound, importantly, showed an intact circle of Willis; however, diastolic flow reversal in the left intracranial internal carotid artery [Figure 2a]. Given the evidence of steal, with its potential for cerebral ischemia, the patient underwent successful surgical re-implantation on day 4 of life. There was a difficult postoperative course requiring multiple inotropes and blood products. Repeat cranial ultrasound following surgery demonstrated normal flow in the left intracranial internal carotid artery [Figure 2b]. The predischarge echocardiogram showed good flow across the aortic arch/innominate anastomosis with no evidence of stenosis. The patient was clinically well and symptom-free when most recently reviewed in the clinic at 5.5 years of age.

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Patient 2 was born at 38 with antenatal imaging suggesting a right aortic arch and small apical VSDs. Postnatal echocardiogram and CT imaging [Figure 3] demonstrated a right aortic arch with isolation of the left innominate artery from the pulmonary artery with bilateral arterial ducts. There were multiple additional apical VSDs, which later closed spontaneously. On cranial ultrasound, the circle of Willis was again intact; however, reassuringly, there was no evidence of diastolic reversal in the right and left anterior cerebral arteries [Figure 4]. In addition to the cardiac defects, the patient had a short-left forearm with ulnar bowing, radius deficiency, and a cleft left hand. They also had left-sided mixed hearing loss, additional pairs of ribs, and vertebral abnormalities. Genetic microarray identified 4q25 deletion, including the *LEF1* gene. The patient was commenced on prostaglandin-E2 on day 2 of life until the anatomy of the circle of Willis was established. They were accepted for surgical repair on an elective basis to minimize the potential of long-term sequelae, including vertebrobasilar insufficiency, left arm claudication, and cerebral ischemia.[1] The patient underwent successful surgical re-implantation of the left innominate artery to the aorta at 7 months of age with an uncomplicated postoperative period. The patient was well on assessment following surgery with good flow into a small left innominate artery.

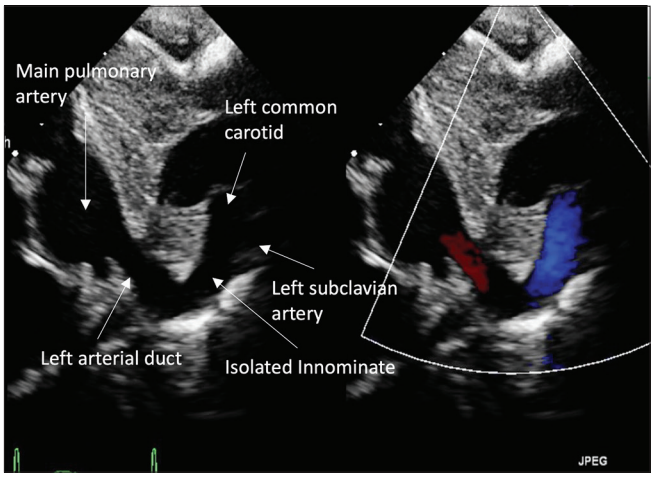

Figure 1: Patient 1 transthoracic echocardiogram with colour compare demonstrating the isolated left innominate artery arising from the main pulmonary artery

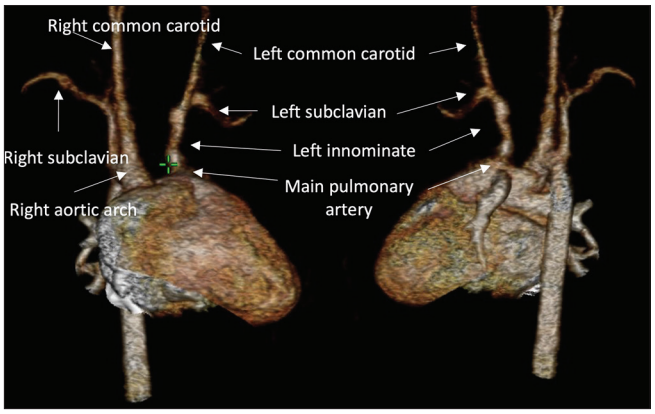

Figure 3: Computed tomography 3D reconstruction on patient 2 demonstrating the isolated left innominate artery from the main pulmonary artery

DISCUSSION

The Edwards hypothetical double-arch model helps us to understand this rare malformation with an interruption at two positions: proximal to the left common carotid and distal to the left arterial duct.[2] Malakan Rad and Pouraliakbar described a novel way to classify patients with isolation of the innominate artery according to the potential sources of steal.[1] According to this classification, both of our cases fall into the worse prognosis category of

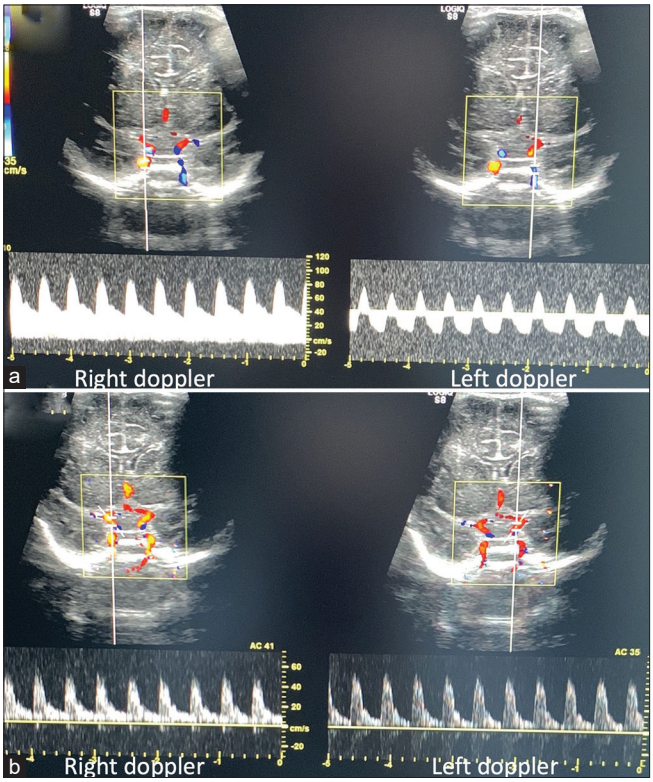

Figure 2: Cranial ultrasound in patient 1 (a) before and (b) after re-implantation demonstrating resolution of diastolic flow reversal in the left intracranial internal carotid artery

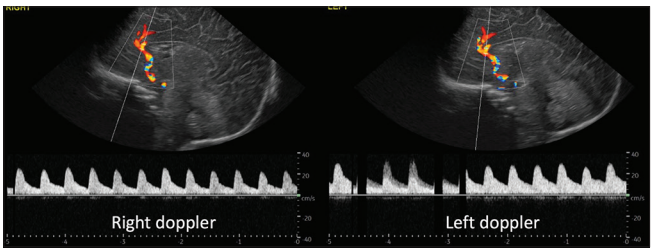

Figure 4: Normal cranial ultrasound dopplers in patient 2

the "triple-steal" type. Blood from the cerebral circulation can be "stolen" to the left subclavian artery, left arterial duct, or right arterial duct. In this circulation, the left cerebral hemisphere and left arm have retrograde perfusion from the right-sided arteries through the circle of Willis. Cranial ultrasound is crucial in these patients to demonstrate an intact circle of Willis and quantify the degree of cerebral steal. To the author's knowledge, this is the first case report showing such a technique to help guide surgical timing. Diastolic steal in the first patient helped inform the decision for immediate repair versus delayed repair in the second patient, for whom this was not observed. There is limited evidence to validate immediate versus delayed re-implantation or even conservative versus surgical management and their impact on the long-term outcomes. Conservative management should be balanced with the potential sequelae of pulmonary over circulation or steal phenomena.[3,4] Surgical considerations should also include the distance between the isolated innominate artery and the aortic arch. In both cases, we describe similarly good outcomes despite the differing ages at repair. However, we observed that the re-implanted left innominate was small in the second patient, possibly due to their later operation, but we postulate that this will grow in time.

Although isolation of the left innominate artery has been associated with Down syndrome, 22q11 microdeletion and Prader-Willi syndrome, we are not aware of any prior report of an association with a microdeletion of the 4q25 region encompassing the *LEF1* gene.[5-7] A 4q25 deletion is usually associated with radial ray defects, hand abnormalities, and ectodermal dysplasia.[8] Although its association with an isolated innominate artery could be a coincidence in our patient, *LEF1* is strongly expressed in the embryologically developing heart in mouse models, and a deletion in this region has been demonstrated in a patient with an aberrant right subclavian artery, suggesting that this gene is potentially involved in aortic arch development.[9,10]

We describe the two cases of a right aortic arch with an isolated left innominate artery arising from the pulmonary artery, successfully operated immediately and at 7 months of age. There is a lack of evidence to guide surgical management and timing; however, cranial ultrasound to detect cerebral steal, as described in this article, can be helpful to assist in decision-making. We further describe the first reported association of a deletion in the 4q25 region encompassing the *LEF1* gene with this rare congenital malformation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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