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Brief Report

Innominate vein vascular ring provides novel insight into systemic venous embryogenesis

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Abstract Anomalies of the innominate vein are uncommon in congenital cardiac disease. We report a case of duplicate innominate veins forming a vascular ring encircling the ascending aorta. We postulate that this vascular ring represents the failure of both a dorsal and ventral precardinal anastomosis to regress.

Keywords: Brachiocephalic vein; cardiac defects; congenital; embryology; vein development

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Case report

A 2-DAY-OLD MALE INFANT PRESENTED WITH respiratory distress. Echocardiography revealed situs solitus, levocardia, normally related great vessels, an aortopulmonary window, abnormal origin of the right pulmonary artery from the aorta, a secundum atrial septal defect, a hypoplastic left aortic arch, severe juxtaductal coarctation of the aorta, and a patent ductus arteriosus. In addition, this patient had an unusual innominate venous vascular ring (Fig 1). To further define the patient's anatomy, cardiac magnetic resonance imaging was performed.

Magnetic resonance imaging revealed a large aortopulmonary window measuring 9 millimetres. At the level of the aortopulmonary window, the right pulmonary artery was arising from the posterior aspect of the ascending aorta. The left pulmonary artery arose normally from the main pulmonary artery. Distal to the aortopulmonary window, the aortic arch was hypoplastic and there was a discrete juxtaductal coarctation. Both a normally situated innominate vein and a retroaortic innominate vein were present. Both innominate veins drained into the single right-sided superior caval vein, which drained normally to the right atrium. These innominate veins effectively created a vascular ring, which encircled the ascending aorta.

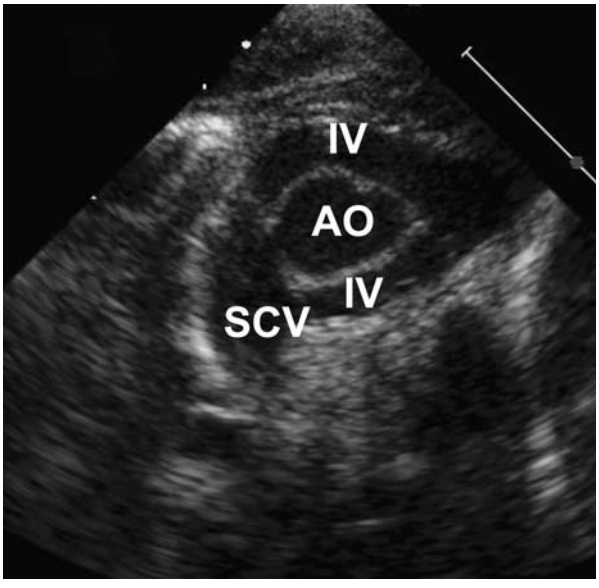

Figure 1.
Echocardiogram. High parasternal short-axis view of the ascending aorta (Ao), innominate vein ring (IV), and superior caval vein (SCV).

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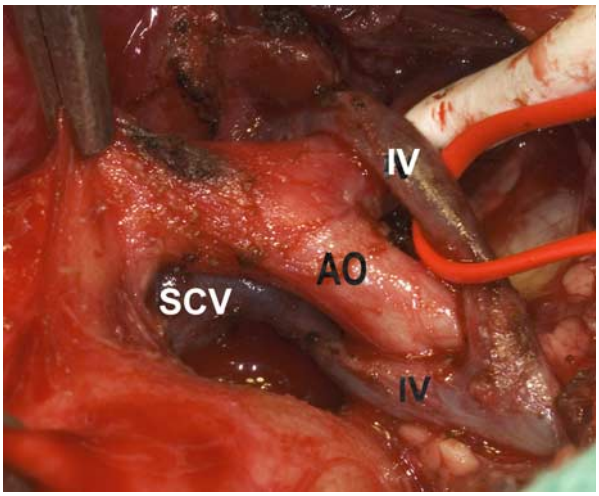

Figure 2.
Intra-operative image. Ascending aorta (Ao), innominate vein (IV), superior caval vein (SCV).

The patient underwent surgical repair at 2 weeks of age (Fig 2). This included an aortic arch reconstruction with a pulmonary artery homograft, repair of the aortopulmonary window, suture closure of the atrial septal defect, ligation and division of the patent ductus arteriosus, and division of the retroaortic portion of the venous vascular ring. The patient had an uncomplicated recovery post-operatively. He subsequently developed mild right pulmonary artery stenosis for which he has not required intervention. No extracardiac congenital anomalies have been identified. Testing for 22q11 deletion was negative and the karyotype was normal.

Discussion

A retroaortic innominate vein in the absence of a normally situated innominate vein occurs in approximately 1% of patients with congenital

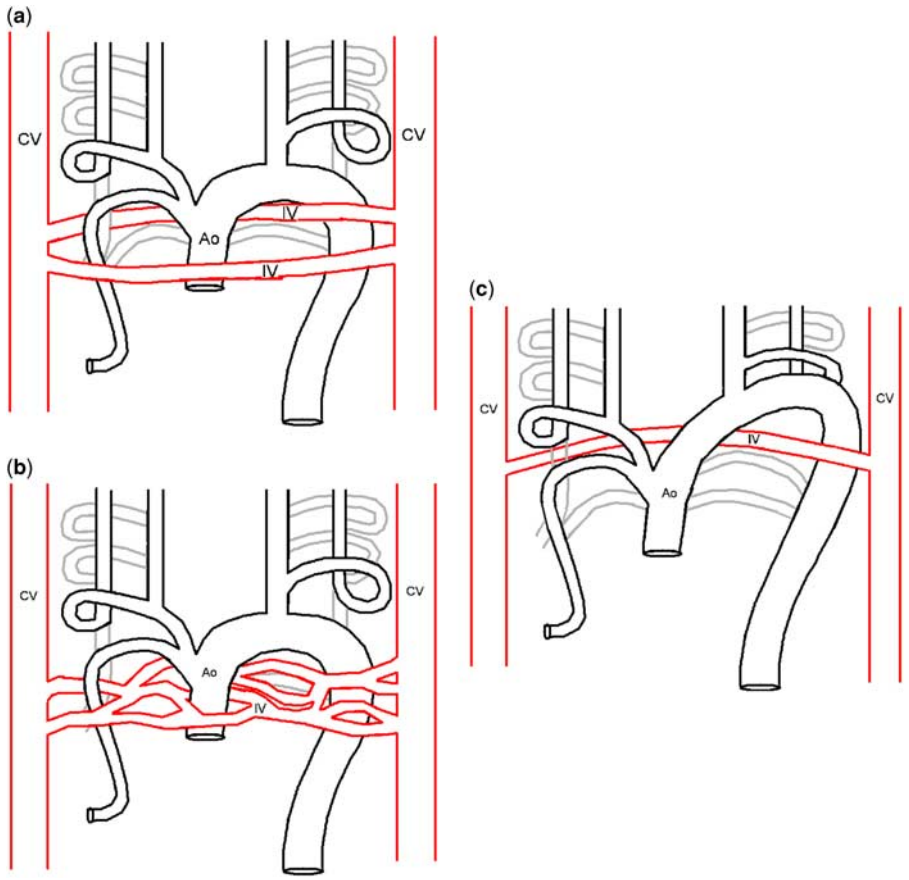

Figure 3.
*Theories of embryological development of the innominate vein. (**a**) Adachi hypothesis – ventral and dorsal double precardinal anastomoses exist with eventual regression of the dorsal anastomosis resulting in a normally situated innominate vein. (**b**) Kim hypothesis – minor mediastinal interconnections between paired cardinal veins with eventual dominance of one ventral vein in normal hearts. (**c**) Location of the developing arterial system influences the pathway the innominate vein is obliged to course. Ao = ascending aorta; CV = cardinal vein; IV = innominate vein.*

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cardiac disease and is most commonly associated with tetralogy of Fallot, pulmonary atresia with ventricular septal defect, and right aortic arch.[1] Duplicate innominate veins were first described by Takada et al[2] in 1992, and to date only four cases have been reported worldwide.[2–4]

The development of the innominate vein occurs in the seventh embryonic week.[3] Several theories regarding the embryogenesis of the innominate vein have been proposed. Adachi[5] hypothesised that double precardinal anastomoses exist, one ventral to the aortic sac and the other dorsal (Fig 3a). In normal development, the dorsal precardinal anastomosis regresses and leaves a normally located innominate vein. According to Adachi's hypothesis, the retro-aortic left innominate vein forms as a consequence of regression of the ventral precardinal anastomosis. Kim et al[6] speculated that minor mediastinal interconnections might exist between the paired cardinal veins during foetal development (Fig 3b). Should a normal brachiocephalic vein fail to develop, partial arrest of regression of a left superior caval vein combined with development of one of these minor mediastinal interconnections might result in a retro-aortic innominate vein. However, other collateral vessels have not been described in patients with a retroaortic innominate vein.

A third theory relates to the development of the arterial system between the fourth and seventh embryonic weeks.[7,8] Although the aortic arch is shortening and the great vessels enlarge, the venous system develops. High aortic arches have been associated with retroaortic left innominate veins.[7,8] It has been postulated that the innominate vein is obliged to course dorsally – in other words, retroaortic – as a result of a high or abnormally elongated aorta obstructing any possible ventral course (Fig 3c).[7,8] However, patients with normal aortic arches have been described with retroaortic innominate veins.[7,9]

We propose that our patient represents the failure of both a dorsal and ventral precardinal anastomosis to regress resulting in this innominate vein ring. It is difficult to conclude what role the complex anatomy of our patient's aortic arch and pulmonary arteries might have had during embryogenesis. Regardless, this case provides strong anatomical evidence for Adachi's theory.

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