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Original Article

Anomalous subaortic course of the left brachiocephalic (innominate) vein: echocardiographic diagnosis and report of an unusual association

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Abstract Subaortic left brachiocephalic, or innominate, vein is an uncommon finding in congenital heart disease, usually associated with obstruction of the right ventricular outflow tract. We describe our experience with 14 patients in whom the lesion was identified echocardiographically, 12 of them with right ventricular obstruction, one with totally anomalous pulmonary venous connection in the absence of obstruction to the right ventricular outflow tract, and the final one with a normal heart. A precise diagnosis of this venous anomaly is of great importance, since it needs to be differentiated from a central pulmonary artery, a pulmonary venous confluence, or an ascending vertical vein in totally anomalous pulmonary venous connection. In patients referred for surgery without catheterization, an incorrect echocardiographic diagnosis could lead to disastrous surgical results.

Keywords: Anomalous left brachiocephalic vein; echocardiography; total anomalous pulmonary venous connection

The left brachiocephalic, or innominate, vein usually courses anterior to the aortic arch, where it joins the right brachiocephalic vein to form the superior caval vein. Rarely, the left vein takes an anomalous course, crossing from left to right beneath the aortic arch, and forming a short superior caval vein, with the azygos vein arising from the right brachiocephalic vein above its site of communication with the left brachiocephalic vein. It is important not to confuse this arrangement with a central pulmonary artery, a pulmonary venous confluence, or an ascending vertical vein draining pulmonary venous return. In this report, we describe 14 patients with this anomaly diagnosed by echocardiography, including an unusual association of cardiac malformations, and review the current literature.

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Patients and methods

From August 1999 to June 2001, we performed 9897 echocardiographic studies in patients suspected of having cardiac disease. The identification of the anomalous pathway by echocardiography was done by insonating from the suprasternal notch in the coronal plane. Unlike the normal course, in which the vein passes anterior to the aortic arch (Fig. 1), no vessel was seen in this position in these 14 patients. This finding first produced suspicion of an anomalous course of the vein. Without any rotation of the transducer, it proved possible to visualize a vessel beneath the aortic arch, parallel and anterior to the right pulmonary artery, which joined the right brachiocephalic vein at a more inferior site, forming a short superior caval vein. With the addition of pulsed Doppler and color flow mapping, non-pulsatile venous flow with blue coloration was detected (Fig. 2). When the transducer was rotated to produce a long axis view of the aortic arch, the vein could be seen in a transverse plane, anterior to the right pulmonary artery.

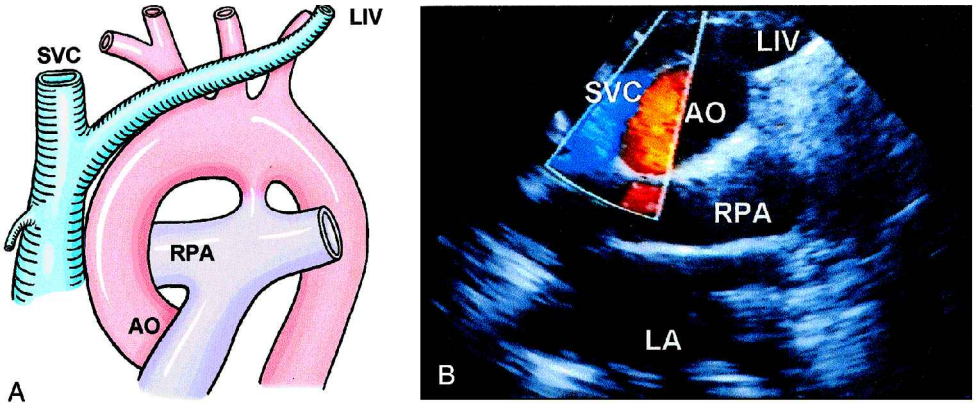

Figure 1.
A: Diagram illustrating the normal course of a left brachiocephalic vein anterior to the aortic arch. B: cross-sectional echocardiographic view from the suprasternal notch showing the normal course of the vein. AO = aorta; LIV = left brachiocephalic vein; RPA = right pulmonary artery; SVC = superior caval vein.

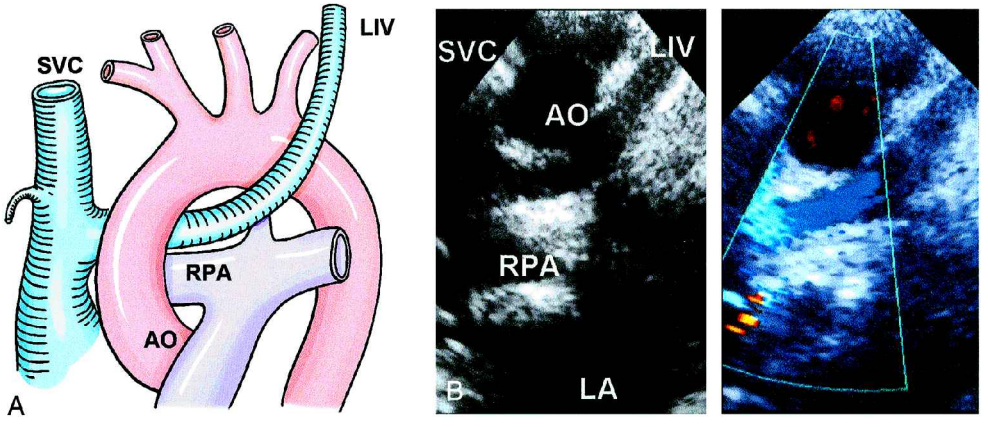

Figure 2.
A: Diagram illustrating the anomalous course of the left brachiocephalic vein, beneath the aortic arch and anterior to the right pulmonary artery. B: **left:** *cross-sectional echocardiographic view from the suprasternal notch* **right:** *color flow mapping (blue) showing the anomalous course of the left brachiocephalic vein. AO = aorta; LIV = left brachiocephalic vein; RPA = right pulmonary artery; SVC = superior caval vein.*

Results

We found 14 patients with an anomalous subaortic left brachiocephalic vein, accounting for 0.14% of the total echocardiograms performed in the period. Their ages ranged from 1 month to 36 years at the time of the diagnosis. Of the patients, 10 were female and 4 male (Table 1).

Additional congenital cardiac malformations were found in 13 patients. Of these, 12 had obstruction of the right ventricular outflow tract. There were 11 patients with tetralogy of Fallot, 9 with stenosis and 2 with pulmonary atresia. The other patient had an atrial septal defect and pulmonary stenosis.

One patient had complex malformations with increased pulmonary flow. Specifically, there was right isomerism, totally anomalous pulmonary venous connection, double inlet right ventricle through a common atrioventricular valve, and large atrial and ventricular septal defects. The right-sided morphologically left ventricle was hypoplastic, with the dominant morphologically right ventricle to the left, indicative of left hand ventricular topology. Both arterial trunks arose from the left-sided right ventricle, with the aorta anterior and to the left of the pulmonary trunk. The central pulmonary arteries were confluent and of normal size, and the aortic arch was left-sided. The pulmonary veins connected to the right superior caval vein via the anomalous

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Table 1. Clinical characteristics, laterality of aortic arch, associated anomalies and diagnostic confirmation.

Patient	Age*	Sex	Diagnosis	Aortic arch	Associated anomalies	Diagnosis confirmation
1	9 mo	F	TOF + PS	R	–	Surgery
2	1 yr 3 mo	M	TOF + PS	R	–	Surgery
3	2 yr 1 mo	F	TOF + PS	L	–	–
4	7 yr 5 mo	F	TOF + PA	R	Absence of LPA; hypoplastic RPA; collaterals	Surgery
5	1 yr 1 mo	F	TOF + PS	L	Supravalvar PS	Surgery
6	7 yr	F	TOF + PS	R	–	Surgery
7	1 mo	M	TOF + PS	R		Surgery
8	8 mo	F	TAPVC AVSD DORV	L	Right isomerism	Surgery Necropsy
9	20 yr	M	TOF + PS	R	–	Surgery
10	36 yr	F	ASD + PS	L	–	Surgery
11	18 yr	F	TOF + PS	L	–	Surgery
12	8 mo	M	TOF + PS	R	–	Surgery
13	15 yr	F	Normal heart	L	–	–
14	1 mo	F	TOF + PA	L	Collaterals	–

*Age at time of echocardiographic diagnosis

Abbreviations: ASD: atrial septal defect; AVSD: atrioventricular septal defetc; DORV: double outlet right ventricle; F: female; L: left; LPA: left pulmonary artery; M: male; mo: month; PA: pulmonary atresia; PS: pulmonary stenosis; R: right; RPA: right pulmonary artery; TAPVC: totally anomalous pulmonary venous connection; TOF: tetralogy of Fallot; yr: year

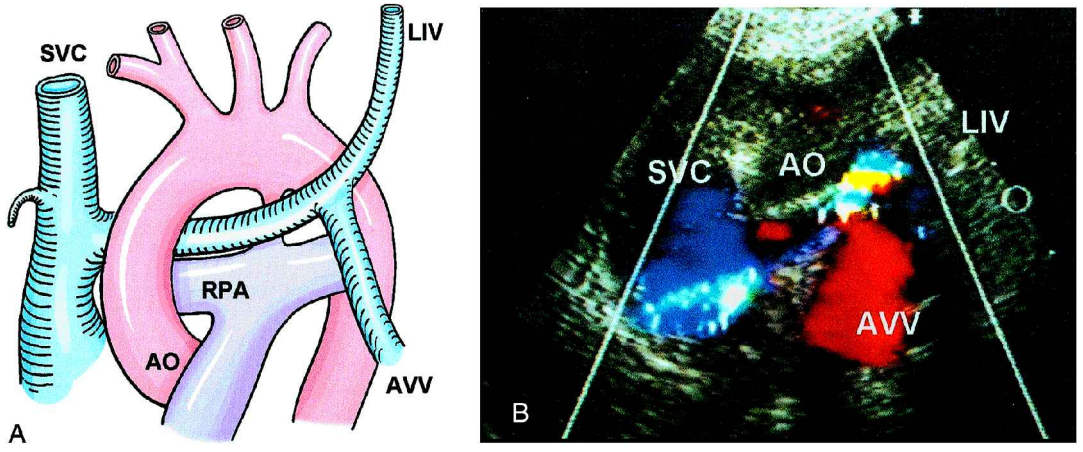

Figure 3.
A: Diagram illustrating the ascending vertical vein draining into the anomalous left brachiocephalic vein. B: cross-sectional echocardiographic view from the suprasternal notch in our patient with right isomerism showing the ascending vertical vein draining into the anomalous left brachiocephalic vein, with a turbulent flow at their junction. AO = aorta; AVV = ascending vertical vein; LIV = left brachiocephalic vein; RPA = right pulmonary vein; SVC = superior caval vein.

left subaortic brachiocephalic vein, with an obstruction at the junction of the pulmonary and systemic venous channels (Fig. 3). In the outstanding patient, the heart itself was normal, the echocardiogram having been indicated to investigate a heart murmur.

The aortic arch was to the right in half of the patients. The diagnosis was confirmed by surgery in 11, and by surgery and autopsy in one (Table 1).

Discussion

It was Kershner[1] who first described this anomaly at autopsy one hundred years ago. Since then, to the best of our knowledge, a total of 59 patients have

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been reported.[1–8] Its incidence is difficult to assess. The veins usually are not routinely studied in detail by any diagnostic exam, even at autopsy.

Gerlis and Ho,[2] in their review of published reports, found an incidence of one case among 2010 angiographic studies of superior caval vein and brachiocephalic veins, and 6 cases out of about 2500 autopsied cases with congenital heart disease. With the advent of cross-sectional echocardiography, the anomalous course of the left brachiocephalic vein could readily be identified noninvasively from the suprasternal notch. Smallhorn et al.[3] were the first to describe the echocardiographic identification of the anomaly. They described seven cases, but the total number of patients studied was not mentioned.

Choi et al.[4] found 24 cases of the venous anomaly among 2457 echocardiograms carried out in patients with congenital cardiac disease, this proportion of 0.98% being the highest frequency thus far reported. In our series, we identified the lesion in 0.14% of 9897 echocardiograms performed, a frequency similar to the one found by Gerlis and Ho,[2] but much lower than the number of cases seen by Choi et al.[4] The latter authors opined that their higher proportion of cases when compared with the results described by Gerlis and Ho[2] could be explained by the difference in the populations studied, and the method used for the diagnosis. They also commented that their echocardiographers were well aware of the lesion, and were therefore unlikely to miss its existence, or make overdiagnoses.[4] We applied the same method of diagnosis reported by Choi et al.[4] We would not deny that the anomaly could escape notice in a routine evaluation, but their high incidence has not been replicated in several other reports.[3,5–7]

The anomalous vein is seen most frequently with tetralogy of Fallot, usually with pulmonary stenosis but also with pulmonary atresia.[2–7] Others associations are listed in Table 2. Our findings are similar to these previous reports. Thus, 11 of our patients had tetralogy of Fallot, either with pulmonary stenosis or atresia. As far as we know, however, the association with right isomerism has not previously been reported. Interestingly, in this patient the anomalous pulmonary venous connection drained through an ascending vertical vein which joined the subaortic left brachiocephalic vein, with obstruction at the junction. Precise echocardiographic diagnosis was crucial in this case, since surgical ligation of the brachiocephalic channel could have proved disastrous.

The aortic arch has been right-sided in 28 of the 38 cases reported with tetralogy of Fallot and pulmonary stenosis or atresia[2–8] (Table 2). We found a right-sided aortic arch in 7 of our 11 patients with tetralogy of Fallot (Table 1).

Table 2. Reported cases of subaortic left brachiocephalic vein with associated cardiovascular malformation and laterality of the aortic arch.

N	Associated anomalies	Right AA
38	TOF/PA + VSD[2–6]	28
2	Common arterial trunk[2,4]	1
1	DORV + "absence" of the pulmonary valve[2]	–
1	VSD[2]	?
1	IAA + APW[2]	
1	TGA[2]	?
1	Tricuspid atresia[7]	–
1	Mirror-imagery + discordant atrioventricular connexion + PA[4]	–
1	Left isomerism[8]	1
2	Minor vascular anomalies*[2]	–
1	RSPV drainage into the superior caval vein[2]	–
1	Anomalous isolated LSA arising from arterial duct[2]	1

*1 case of a small subthyroid transverse venous anastomosis, and another one of left jugular vein anomaly

Abbreviations: AA: aortic arch; APW: aortopulmonary window; DORV: double outlet right ventricle; IAA: interrupted aortic arch; LSA: left subclavian artery; N: number of cases; PA: pulmonary atresia; TGA: transposition of the great arteries; TOF: tetralogy of Fallot; VSD: ventricular septal defect; RSPV: right superior pulmonary vein

Of the 59 patients thus far reported, only 8 had the anomalous venous channel as an isolated lesion. Gerlis and Ho[2] could find no example of the anomalous vein in their review of approximately 5000 autopsies of individuals with normal hearts. Our final patient, nonetheless, had the vein as an isolated finding in an otherwise normal heart.

Although the anomaly usually has no clinical implications, it is important for the echocardiographer to be aware of its existence, and not to miss its presence, or more importantly to mistake the structure for a central pulmonary artery, a venous confluence, or a vertical ascending vein in patients with anomalous pulmonary venous connection.

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