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Anomalous retroaortic paravertebral course of the left innominate vein in a child with atrial septal defect

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Abstract

We report the case of a 2.5-year-old boy with atrial septal defect in whom anomalous position of the left innominate vein was detected on preoperative ultrasound examination. Before joining the right brachiocephalic vein, the vessel extended from the left to the right and downward beyond the descending aorta. It was considerably flattened by the thoracic vertebral column, and was invisible on ultrasonography in this section. The appearance of the visible segments raised a suspicion of an anomalous course of persistent left superior vena cava draining into the left atrium, dilated azygos vein in a case of interrupted inferior vena cava, or partial anomalous pulmonary venous return. Since all doubts had to be resolved before open heart surgery, a decision was made to expand the diagnostic work-up to include computed tomography angiography. We present the echocardiographic and computed tomography findings of this unusual and previously unreported case of anomalous venous return.

Case report

A boy with Down syndrome, aged 2.5 years, was referred to the Department for corrective cardiac surgery of a large ostium secundum atrial septal defect (ASD II) (Qp: Qs = 2.8 : 1). The general condition of the child was good, with no signs of overt heart failure.

Echocardiography

Preoperative echocardiographic evaluation (Fig. 1, Fig. 2, Fig. 3, Fig. 4, Fig. 5, Fig. 6) additionally revealed the presence of a wide vein with an atypical course in the posterior mediastinum. The vessel was adjacent posteriorly and laterally on the left to the descending aorta, and slightly below the junction with the left pulmonary artery it disappeared into the aerated lung tissue, most likely in the posterior (retroaortic) region. The flow in the vessel was considered typical for a systemic vein with a downward direction of flow. The left brachiocephalic vein was not visualized in its usual location. In this area, the persistent left superior vena cava (LSVC) can be seen, with a similar direction of flow, though it passes anteriorly from the aorta, crosses the left pulmonary artery anteriorly and then, between the left atrial appendage and the left superior pulmonary vein (posteriorly), descends behind the wall of the left atrium, draining into the wide coronary sinus. It is considerably less common for the LSVC to drain directly into the left atrium above the opening of the left superior pulmonary vein and the appendage. The vein studied in this case, however, had a different course. Its inferior segment was located at a considerable distance from the left atrium, close to the posterior chest wall, disappearing behind the aorta, probably crossing it posteriorly. The coronary sinus had a normal diameter of approximately 3 mm.

The right parasternal view place reveals the opening of a wide, intensely color-filled venous vessel in the region where the azygos vein (*vena azagos*, VA) drains into the

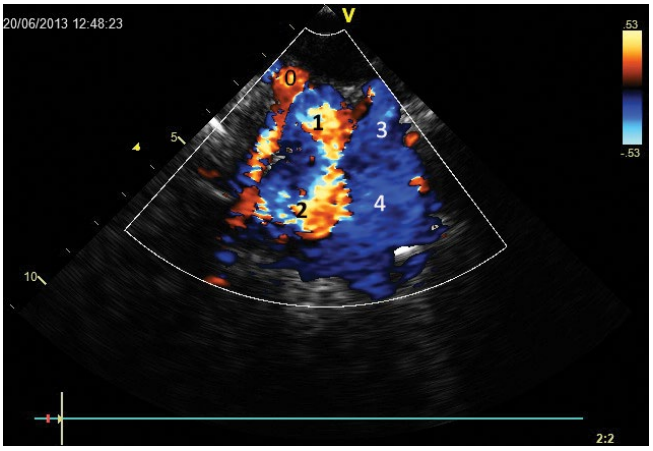

Fig. 1. *Cross-section showing the upper mediastinum in an inclined plane close to the plane where the aortic arch passes. Visible distal transverse portion of the arch (1) and the initial segment of the descending aorta (2). Systole. The scale of representation of flow velocities with color is adjusted to the visualization of relatively low velocities (max. 53 cm/s, so the aorta is filled with mosaic color dominated by blue (downward flow). A wide vessel (3) runs laterally to the left and posteriorly from the aorta, with a continuous downward flow much at a velocity considerably lower than in the aorta. The inferior segment of the vessel (4) crosses the thoracic aorta posteriorly; the further course of the vessel cannot be traced. Multiple color artefacts caused by proximity to the vessels of aerated lung tissue (0)*

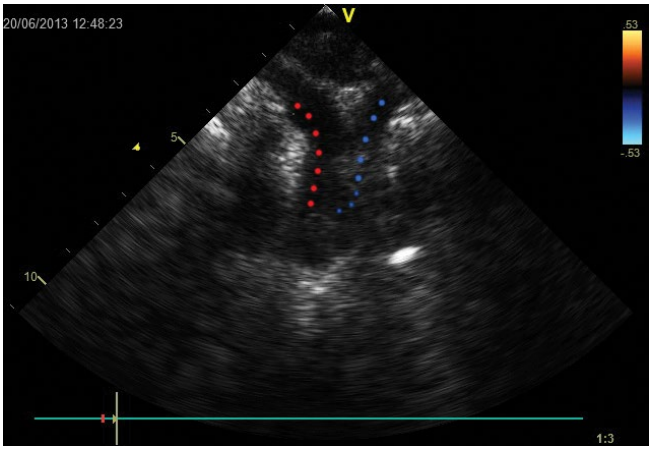

Fig. 2. *Image presented in Fig. after color removal. Without color, the vein which passes retroaortically (red dotted line) is far less clearly visualized, but its lumen can still be identified (blue dotted line)*

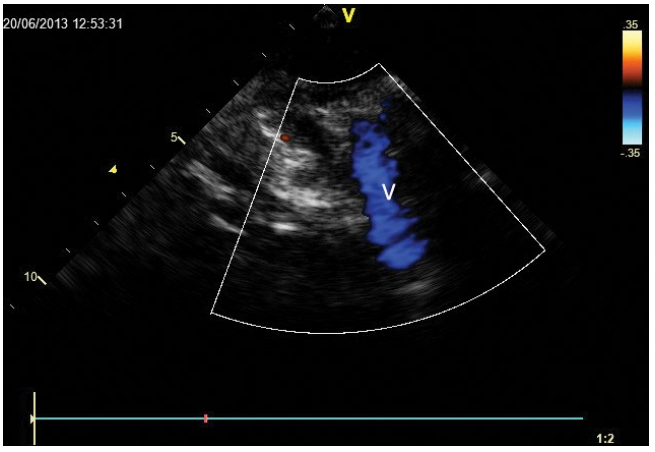

Fig. 3. *Corresponding view during diastole, with flow noted only in the venous vessel (V) adjacent to the aorta*

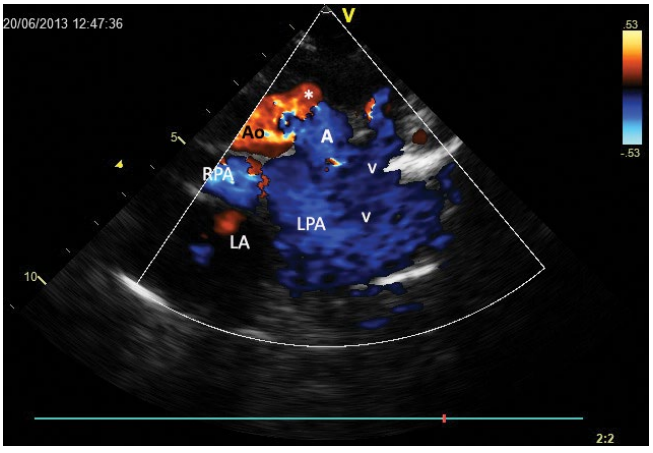

Fig. 4. *Cross-section in a plane close to frontal (with slight head rotation). Visible proximal part of the aortic arch (Ao) and its transverse section; a vertically running vein (V) crosses posteriorly the left pulmonary artery (LPA) and the descending aorta. Multiple respiratory artefacts obscuring the view of vascular flow. LA – left atrium, RPA – right pulmonary artery*

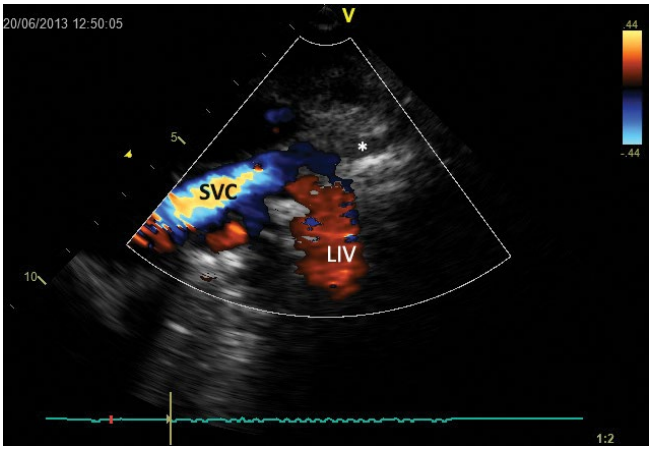

Fig. 5. *High parasternal view (right) visualizing mediastinal structures in the sagittal plane. Superior vena cava (SVC) with a wide venous vessel (LIV) draining into it slightly above the junction with the right pulmonary artery. It is the typical drainage site of the azygos vein, though in normal conditions it is markedly less intensely color-filled. Another important observation is a significant increase in flow rate in the proximal segment of the superior vena cava. (*) right brachiocephalic vein*

superior vena cava (SVC), slightly above the junction with the right pulmonary artery. Similarly to the VA, the vein passed in the sagittal plane. Spectral Doppler examination showed the characteristic pattern of systemic venous flow (typical respiratory variation, S-wave velocity markedly greater than D-wave velocity). It should be noted, though, that under normal conditions the VA is much less intensely color-filled. A notable observation was that the flow rate was higher in the proximal segment of the superior vena cava, suggesting an increased volume of blood draining into the SVC. The findings might be consistent with interrupted inferior vena cava (IVC) with its continuation through the azygos vein, but echocardiography revealed no abnormalities of the intrahepatic segment of the inferior vena cava (IVC). The differential diagnosis also had to include partial anomalous pulmonary venous return but, firstly, the four pulmonary veins were found to drain into the left atrium; secondly, the pulmonary veins draining into the SVC run in the frontal rather than sagittal plane; and thirdly, they show a different flow pattern with practically no respiratory variation, and mostly similar S- and D-wave velocities. It was hypothesized that the left-sided vertical vein might cross the posterior mediastinum and drain into the SVC together with the azygos vein, but echocardiography failed to visualize its entire course. In the context of scheduled cardiac surgery, it was considered necessary to clarify the existing doubts, as atypical anatomy of the venous system could pose the risk of complications associated with systemic venous cannulation as well as undiagnosed intersystemic leaks. A CT angiography scan was performed, revealing abnormal location of the left innominate vein joining the right brachiocephalic vein right above the right main bronchus and the right pulmonary artery. The vessel passed behind the descending aorta and on the anterior surface of the thoracic vertebral bodies (at Th 5–6) posteriorly from the tracheal bifurcation and esophagus, where it became manifestly flattened (Fig. 6 and Fig. 7).

Standard uncomplicated cardiosurgical correction of the defect with direct cannulation of the venae cavae was performed. In postoperative echocardiography, attempts were undertaken to visualize the transverse, most posterior segment of the left superior vena cava. However, despite an in-depth knowledge of the anatomy, no clearly satisfactory images were obtained.

Discussion

Under normal anatomical conditions, the left innominate vein (LIV), also referred to as the left brachiocephalic vein, is formed by the confluence of the left internal jugular vein and the left subclavian vein. The LIV passes to the right, crossing the midline, with a course anterior and superior to the vessels of the aortic arch. The left and right brachiocephalic veins merge to form the superior vena cava.

The retroaortic course of the LIV (posteriorly from the ascending aorta) is found in 0.2–1% of patients with congenital heart defects. In 70% of cases, it coexists with tetralogy of Fallot and the right-sided aortic arch[1,2]. Other

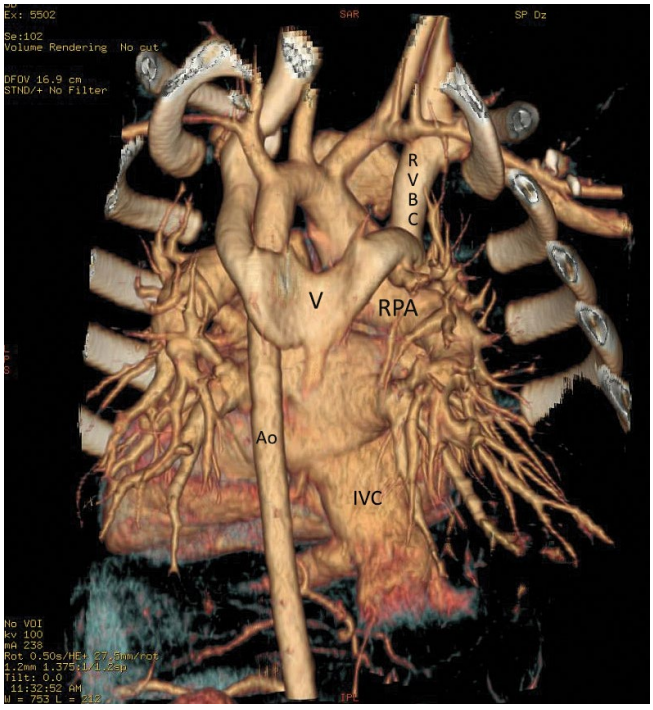

Fig. 6. *3D angiotomography reconstruction – posterior view, with removed bone elements of the posterior chest wall: vertebral column and ribs. In the foreground, a wide flattened venous vessel (V) located posteriorly from the descending aorta (Ao) is visualized. The vein is a continuation of the atypically running left brachiocephalic vein. Very clear deformation by adjacent thoracic vertebral bodies is seen; at this level, a relatively wide venous channel, probably the azygos vein, drains into the vessel from the bottom. Anomalous left innominate vein (V) joins the right brachiocephalic vein (RVBC) slightly above the junction with the right pulmonary artery (RPA). Normal drainage of pulmonary veins into the left atrium is clearly visualized. IVC – inferior vena cava*

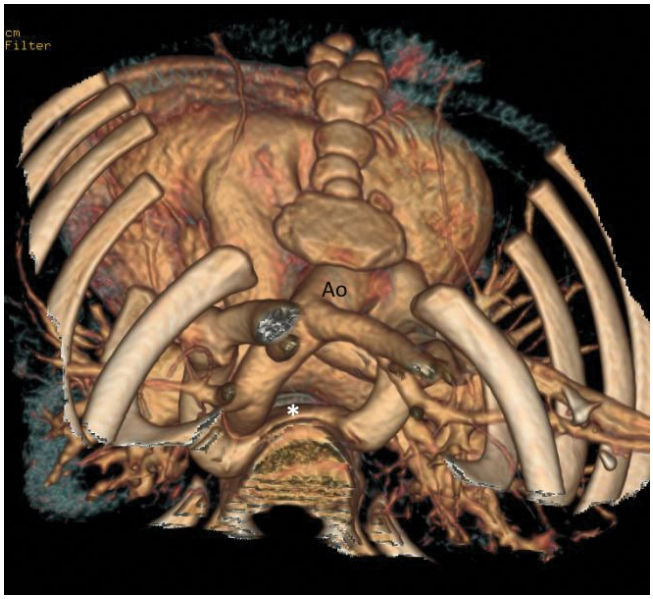

Fig. 7. *CTA – anterior and top view. Preserved features of vertebral bodies with a wide venous channel extending tightly on the spine (*)*

congenital abnormalities, by the frequency of coexistence, include ventricular septal defect with pulmonary atresia, common arterial trunk and interrupted aortic arch[3,4]. The first report, authored by Kershner, was published over 120 years ago[4]. The available literature[1–6] discusses retroaortic course as the position of the left innominate vein (LIV) posteriorly from the aortic arch or the ascending aorta. This case report presents what appears to be the first published case of a "truly retroaortic" course of the LIV, in which the vein is flattened on the thoracic vertebrae and it is located posteriorly from the descending aorta (retroaortic left innominate vein, RAoLIV). It is important to note that a comprehensive review of anomalies of the brachiocephalic (innominate) vein presented by Chen *et al*.[5] does not include the anomaly discussed in this case report.

The cause of the anomalous location of the LIV was not fully elucidated. It is probably an effect of abnormal development of the system of paired anterior and posterior cardinal veins draining into the venous sinus, with the anterior cardinal veins being connected via the superior and inferior transverse venous plexuses. Anomalies in the development of these junctions lead to an atypical course of the resulting vessels (LIV)[1–3,5].

The anomaly does not cause overt clinical manifestations, but it poses a problem to diagnosticians, precluding precise assessment of vascular structures[2]. The diagnosis of RAoLIV can be verified by transthoracic echocardiography. The differential diagnosis should consider persistent left superior vena cava, ascending vertical vein in total anomalous pulmonary venous return and left-sided partial anomalous pulmonary venous return[5]. The middle section of the LIV requires differentiation with the pulmonary trunk, and in its retroaortic course it may be misinterpreted as enlarged lymph nodes[3]. An anomalous course of the left brachiocephalic vein can be determined precisely by CT angiography[5].

Accurate diagnosis of atypical LIV morphology is a prerequisite for performing safe corrective cardiac surgery under extracorporeal circulation, establishment of central venous access via the left jugular vein, and transvenous placement of a pacing electrode from the access via left brachial vessels. Precise determination of anatomy allows safe dissection of the mediastinal vessels, and cannulation of the SVC[5,6].

Conclusions

1. Echocardiographic examination is not always sufficient to resolve all doubts concerning extracardiac vascular structures, especially if they are obscured by aerated lung tissue or bone structures. However, it is usually possible to detect abnormalities indicating a potential anomaly.
2. This case report is probably the first published case of a truly retroaortic course of the left innominate vein which not only runs posteriorly from the aortic arch, but also posteriorly from the descending aorta.

Conflict of interest

The authors do not declare any financial or personal links with other persons or organizations that might adversely affect the content of the publication or claim any right to the publication.

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