

Levoatriocardinal Vein Associated with Pulmonary Stenosis and Aortic Hypoplasia in Young Adult

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Introduction

The levoatriocardinal vein (LACV) was first described in 1926 by McIntosh as a persistent abnormal connection between the pulmonary and the systemic venous system through the splanchnic plexus. It is most commonly associated with left obstructive lesions such as mitral atresia, cor triatriatum, left heart hypoplasia, aorta atresia or coarctation.^{1,2} It is believed that the persistence of this communication occurs to provide an alternative in the pulmonary venous drainage in a scenario where the left heart is malformed and under increased pressures.³

We report the case of a young adult with levoatriocardinal vein determining pulmonary hyperflow with consequent overload of the right chambers associated with pulmonary stenosis and ascending aortic hypoplasia.

Case Report

A 19-year-old white male patient was referred to the cardiology outpatient clinic to investigate hypertension and cardiac murmur. The patient reported ventilatory dependent pinching on the chest during deep inspiration, dyspnea to mild exertion and sensation of fatigue and prostration. On physical examination, there was symmetrical 170/80 mmHg blood pressure in the upper and lower limbs. Cardiac auscultation revealed normal heart sounds, regular rhythm, split S2 and ejection sound with systolic murmur of 3+/6+ in pulmonary focus. Additional tests were requested for investigation. Resting electrocardiography presented sinus rhythm, clockwise rotation and axis around +120°, no overloads or blockages. Echocardiography showed major increase in right chambers with preserved right ventricular systolic function, intact atrial septal defect bulging towards the left, normal pulmonary venous drainage through four pulmonary veins draining in the left atrium and presence of an anomalous ascending vertical vessel (levoatriocardinal vein) originating near the pulmonary hilum in the left upper lobe with abundant flow, flowing into the innominate vein and later into the superior vena cava and

Palavras-chave

Congenital cardiomyopathy with increased pulmonary blood flow; Pulmonary Stenosis; Anomalous pulmonary venous drainage; Aortic Hypoplasia.

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right atrium (Figure 1). Pulmonary valve slightly thickened with funnel-shaped systolic dynamics, mild stenosis with maximum systolic gradient estimated at 37 mmHg, overestimated due to the hyperflow. Aorta with slight hypoplasia of the ascending tubular portion, measuring 17 mm in diameter, reference value 26-34 mm.⁴ Left ventricle with normal dimensions and ejection fraction of 76%.

After evaluation of the initial tests, the patient was referred for admission and evaluation concerning a potential surgery. The patient underwent computed tomography angiography of the chest showing the four pulmonary veins properly draining into the left atrium (Figure 2) and an anomalous vessel originating in the left superior pulmonary vein flowing into the innominate vein (Figure 3). The patient underwent surgical correction by median sternotomy and cardiopulmonary bypass connecting the levoatriocardinal vein to the innominate vein and redirection of this vessel to the left atrial appendage. After a good clinical evolution, the patient was discharged with prescription of outpatient follow-up and maintenance of hypertension therapy, which was interpreted as secondary to aortic hypoplasia.

Discussion

In the embryogenesis of the first two months of fetal development, the lungs drain into the systemic veins and the pulmonary veins get formed from a pocket on the dorsal wall of the primitive left atrium. When the common pulmonary vein merges into the primitive lungs, the pulmonary drainage system obliterates into the systemic veins. Then the common pulmonary vein is incorporated into the left atrium and is normally differentiated into four vessels, two of which for drainage of each lung.⁵

Based on the above, the development of pulmonary drainage anomalies begins early during embryogenesis and may occur when any of these stages does not progress adequately.⁶ The types of abnormal drainage of pulmonary veins can be divided into four: right anomalous pulmonary venous drainage (APVD), left APVD, scimitar syndrome and levoatriocardinal vein.⁵ Anomalous drainage can occur through the superior vena cava, right atrium, inferior vena cava, azygous veins, innominate vein, coronary sinus and hemiazygos veins.⁷ When obstructive lesions of the left heart occur, the pulmonary veins may develop normal communication with the left atrium, but may retain some form of primitive communication with the cardinal system. This abnormal connection serves as an alternative outflow route for pulmonary venous system drainage into the cardinal system through the innominate vein, jugular vein or superior vena cava and forms the base of the levoatriocardinal vein.⁸

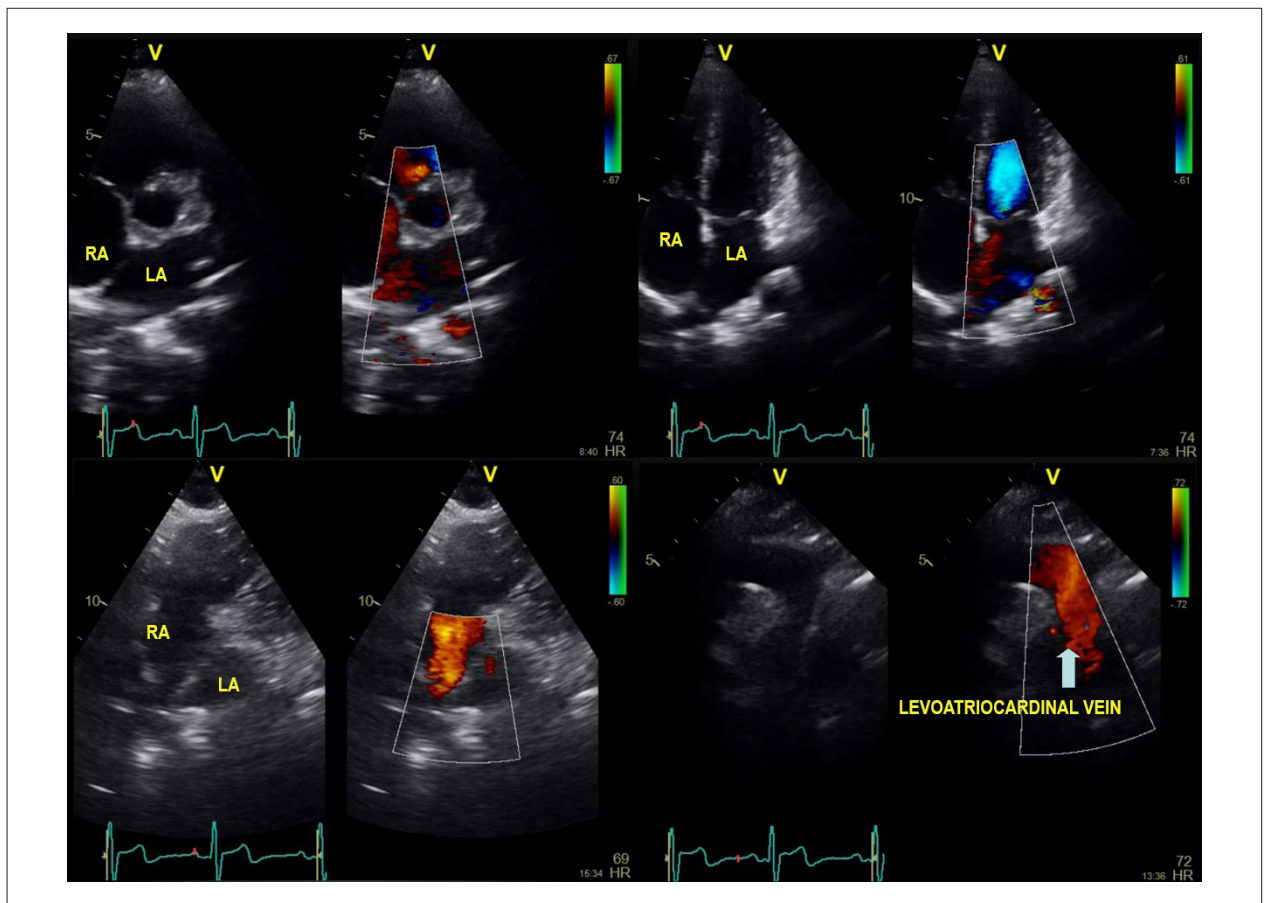


Figure 1 – Echocardiographic images showing intact interatrial septum, pulmonary veins draining into the left atrium and levoatriocardinal vein draining into the innominate vein.

The levoatriocardinal vein is a rare entity distinct from anomalous pulmonary venous drainage, in which the primary abnormality is the absence of connection of one or more pulmonary veins in the left atrium.⁸ The LACV is a collateral venous vessel that connects the left atrium or one of the pulmonary veins to the systemic venous system, usually The innominate vein.⁹

In a case series reported by Bernstein et al., 25 LACV patients were studied. The occurrence of LACV associated with intracardiac obstruction of pulmonary venous return was generally associated with intact atrial septum. Only in 4 cases (16%) there was a hemodynamically significant atrial defect, showing that a functionally intact atrial septum was not absolutely necessary for the formation of the levoatriocardinal vein.¹⁰

The levoatriocardinal vein phenomenon may occur in a structurally normal heart, in isolation, and clinical findings may mimic partial anomalous pulmonary drainage or atrial septal defect. Patients with intracardiac anatomy and normal pulmonary venous return may have symptoms of low output and overload of right chambers during life.²

Conclusion

The levoatriocardinal vein is a rare cause of left-right shunting and should be remembered in differential diagnosis

in patients with right-chamber overload after evaluation of atrial septal defects and pulmonary venous drainage.

Echocardiography is an efficient noninvasive test capable of diagnosing levoatriocardinal vein.

Authors' contributions

Research creation and design: Pretto JLCS; Data acquisition: Pretto JLCS, Baldissera DMB, Hoppen GR; Data analysis and interpretation: Pretto JLCS; Manuscript drafting: Pretto JLCS, Baldissera DMB, Roman RM; Critical revision of the manuscript as for important intellectual content: Pretto JLCS, Ferreira MO, Menegotto ET, Roman RM.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate program.

Case Report



Figure 2 – Computed tomography angiography showing 4 pulmonary veins draining into the left atrium.



Figure 3 – Reconstruction of computed tomography angiography showing the levoatriocardinal vein draining into the innominate vein.

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