

# Levoatrial cardinal vein with normal left ventricle: A forgotten cause of pulmonary arterial hypertension

Onkar B Auti, Varun Shetty<sup>1</sup>, Vinay Belaval, Vimal Raj

Departments of Radio-diagnosis, <sup>1</sup>Cardiovascular Surgery, Narayana Health City, Bengaluru, Karnataka, India

**Correspondence:** Dr. Onkar B Auti, Department of Radio-diagnosis, Narayana Health City, Bengaluru, Karnataka, India.

E-mail: auti.onkar@gmail.com

## Abstract

Levoatrial cardinal vein (LACV) is anomalous connection between left atrium or pulmonary veins and systemic veins such as innominate vein or superior vena cava. This persistence of splanchnic circulation occurs when there is left-sided obstructive cardiac lesions such as hypoplastic left heart or mitral atresia. In this report we present three cases of LACV with well-developed left heart, without any obstructive lesions. All our cases presented with pulmonary arterial hypertension (PAH) and had associated intracardiac shunt such as ventricular/atrial septal defect and supracardiac partial anomalous pulmonary venous connection. Apart from the above shunts, LACV contributed to PAH in these cases. It is important to detect and report LACV as this may require surgical correction along with other defects. If LACV goes undetected during imaging workup, it may cause persistent PAH postoperatively.

**Key words:** Hypoplastic left heart; levoatrial cardinal vein; mitral atresia; pulmonary arterial hypertension

## Introduction

Levoatrial cardinal vein (LACV) is anomalous connection between the left atrium or pulmonary veins and any systemic vein which is derived from cardinal venous system.<sup>[1,2]</sup> Most often, this anomaly has been described in the context of left-sided obstructive cardiac lesions such as mitral atresia or hypoplastic left heart.<sup>[3]</sup> LACV is abnormal persistence of the splanchnic vein connecting the pulmonary venous plexus and the cardinal system.<sup>[4,5]</sup> Blood flow in LACV is from left atrium/pulmonary veins to right side of the heart amounting to left to right shunt. This connection may persist to provide alternative drainage of pulmonary venous blood in conditions where left heart is malformed.

Presence of LACV alone or in association with other shunts can cause pulmonary arterial hypertension (PAH). We present three cases (pediatric and adult) of LACV with associated other congenital heart defects and well-developed left side of the heart. A normal left ventricle without any obstruction in all these cases is an uncommon phenomenon.

## Case History

A 7-month-old male child presented with history of rapid breathing, poor feeding, and failure to thrive since birth. He was born preterm to a diabetic mother. He was on ventilator support for first 5 months of his life for respiratory distress and suffered through repeated seizures. He underwent

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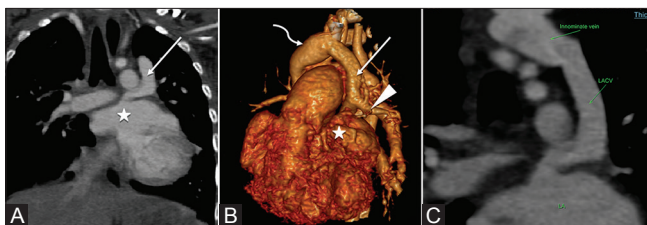


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surgery for necrotizing enterocolitis and was also diagnosed with a congenital heart defect and advised surgery. On examination, his motor milestones were grossly delayed, cardiovascular examination showed a loud second heart sound, and no murmurs. Electrocardiogram showed normal sinus rhythm with an axis of 90° and right ventricular hypertrophy. Chest X-ray showed a cardiothoracic ratio of 0.7 and increased pulmonary vascularity. Transthoracic echocardiogram showed a large inlet ventricular septal defect (VSD) with bidirectional shunt, parachute mitral valve with no stenosis, or insufficiency. A patent ductus arteriosus (PDA) and an anomalous communication between the left pulmonary vein and the innominate vein were also reported. There was severe PAH. A multidetector computer tomography (MDCT) (GE VCT, Lightspeed) examination was performed for further evaluation of the anomalous communication. This showed anomalous vessel between the left atrium and the innominate vein, i.e. LACV. The left superior pulmonary vein (LSPV) was shown to drain into the LACV, which contributed as partial anomalous pulmonary venous connection (PAPVC) [Figure 1]. MDCT also showed subaortic VSD [Figure 2] and features of PAH. PDA noted on echo was not seen on MDCT. He underwent surgical repair of the VSD, rerouting of the LSPV into the left atrium. The child was extubated the next day and had an uncomplicated recovery.

Our second case was that of a 40-year-old female, who presented with shortness of breath, class II New York Heart Association in the last 6 months. Her symptoms had gradually worsened and she had developed paroxysmal nocturnal dyspnea and syncope. In the past she was suspected to have acute pulmonary embolism following her first pregnancy and was put on oral anticoagulation for 6 months. On examination, her jugular venous pulse was elevated and an ejection click was heard in the pulmonary area. Her electrocardiogram showed right axis deviation with a strain pattern. Chest X-ray showed enlargement of the right atrium and pulmonary arteries. Transthoracic echocardiogram reported a moderate-sized ostium secundum atrial septal defect (ASD), dilated right heart, moderate tricuspid valve regurgitation, and severe PAH. MDCT (GE VCT, Lightspeed) was advised to exclude



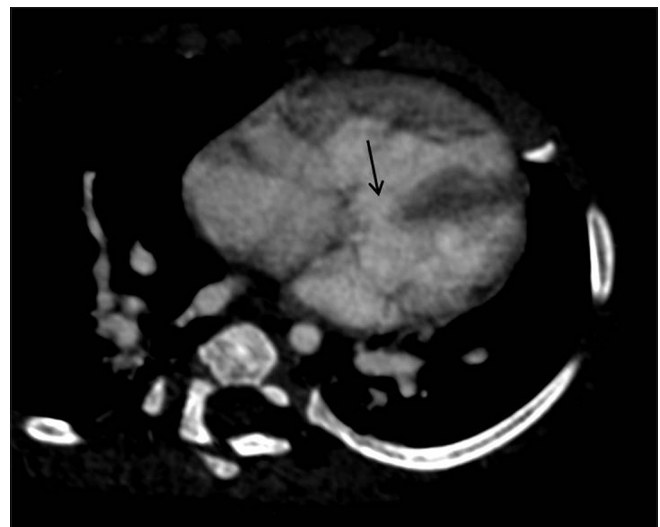
**Figure 1 (A-C):** Case 1; (A) Coronal MDCT image showing anomalous LACV (straight arrow) draining into left atrium (star). (B) 3D reconstruction showing LACV (straight arrow) connecting left atrium (star) and innominate vein (curved arrow). Note the left superior pulmonary vein (arrowhead) draining into LACV. (C) Curved reformats showing anomalous LACV connecting left atrium and innominate vein

pulmonary thromboembolism. MDCT demonstrated LACV connecting left atrium and the innominate vein. PAPVC was noted where LSPV drained into the LACV [Figure 3]. It also showed ASD [Figure 4]. There were no signs of acute or chronic pulmonary thromboembolism. It was decided to treat the patient medically.

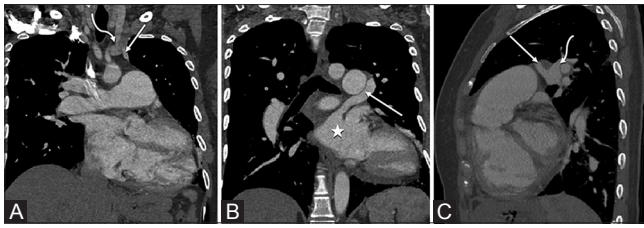
Our third case was of a 5-month-old female child who presented with rapid breathing with suck rest suck cycle. Patient also had history of recurrent lower respiratory tract infections. On examination, cyanosis was noted with regular pulse and tachypnea. On auscultation, early systolic murmur was heard. Echo demonstrated large ASD with bidirectional shunt, PAPVC in which left pulmonary veins were draining into the innominate vein via left ascending vein. There was bicuspid aortic valve and PAH. MDCT (GE VCT, Lightspeed) showed LACV connecting left inferior pulmonary vein to innominate vein. The left superior and middle pulmonary veins also drained into LACV [Figure 5]. Left inferior pulmonary veins were also communicating with the left atrium. MDCT also confirmed presence of ASD and bicuspid aortic valve [Figure 6].

## Discussion

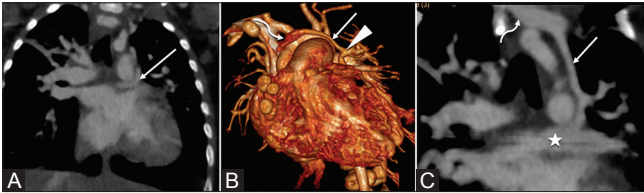
LACV is an abnormal connection between the pulmonary venous and systemic venous drainage system. LACV was first reported by McIntosh in 1926 and the term was used by Edwards and DuShane in 1950 in a case of mitral atresia.<sup>[1,2]</sup> Embryologically, LACV constitutes a persistent connection between pulmonary and cardinal venous system particularly in obstructive left heart lesions to facilitate drainage of left atrium to right side.<sup>[5]</sup> In cases of left ventricular outflow obstruction, LACV acts as a “pop off valve” and allows pulmonary venous drainage into



**Figure 2:** Case 1; Reformatted MDCT image (four-chamber view) demonstrating subaortic VSD (arrow)



**Figure 3 (A-C):** Case 2; (A and B) Coronal images MDCT images showing LACV (straight arrow) connecting left atrium (star) and innominate vein (curved arrow). (C) Sagittal image showing left superior pulmonary vein (curved arrow) draining into LACV (straight arrow)



**Figure 5 (A-C):** Case 3; (A) Coronal images showing LACV (straight arrow). (B) Three-dimensional reconstruction showing LACV (straight arrow) connecting innominate vein (curved arrow) to left superior pulmonary vein (arrowhead). (C) Curved reformations showing anomalous LACV (straight arrow) connecting left inferior pulmonary veins (star) and innominate vein (curved arrow)

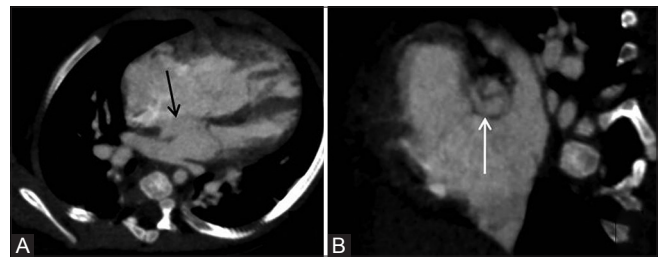
the systemic venous system without raising the left atrial pressure.

There are many cases of LACV reported with left-sided obstructive lesion, and the largest study was done by Bernstein *et al.*, who reported 25 patients of LACV with left-sided obstructive lesions such as mitral atresia, aortic stenosis, aortic coarctation, and cor triatrium.<sup>3</sup> Presence of LACV is not very common in the setting of normal left heart morphology. A handful of cases have been reported in the literature, with this report being the largest series. Fujiwara *et al.*<sup>[6]</sup> reported a case of LACV in a boy being operated for Tetralogy of Fallot. Another case was report of a 38-year-old woman with an otherwise normal heart, but required surgery because of the increased shunt across the LACV<sup>5</sup>. Recently, Genc *et al.*<sup>[7]</sup> have reported it as an incidental finding in a patient undergoing MDCT. LACV has also been described in cases with acquired rheumatic mitral stenosis.<sup>[8]</sup>

Differentiating LACV from left SVC is important as left SVC has no clinical significance. Usually, left SVC drain into coronary sinus, which further joins right atrium. On contrary, LACV connects to left atrium or pulmonary vein. Direction of blood flow in left SVC is toward heart (vertically downwards), while that of LACV is away from heart (vertically upwards). Direction of blood flow in anomalous vessel can be assessed by color Doppler or phase-contrast MR imaging. The left innominate vein is either absent or hypo plastic and works as a bridging vein in patients with left SVC. In diagnosis of LACV, presence of left innominate vein is essential.<sup>[3]</sup>



**Figure 4:** Case 2; Axial MDCT image demonstrating ASD (arrow)



**Figure 6 (A and B):** Case 3; (A) Reformatted MDCT image (four-chamber view) demonstrating ASD (black arrow). (B) Reformatted MDCT image of aortic valve showing bicuspid valve (white arrow)

All of our patients were detected to have a LACV in the absence of left heart obstruction with MDCT. All of these patients had another associated intracardiac shunt, such as VSD or ASD. Supracardiac type of PAPVC was noted in all cases where left pulmonary veins were draining into LACV, which further drains into innominate vein, SVC, and right atrium. LACV along with these associated shunts contributed to the PAH in our cases. Therefore, it is important to address LACV also if surgical management is being considered. If LACV goes undetected during imaging workup, it may cause persistent PAH postoperatively.<sup>[6]</sup> If symptomatic, patients can also be treated noninvasively with device closure.<sup>[9]</sup>

MDCT is an excellent tool for delineating the anatomy of anomalous venous drainage systems. The high spatial resolution, multiplanar reconstruction (MPR) and three-dimensional volume-rendering technique and maximum intensity projection provided by this technique enable a detailed evaluation of vascular anomalies and pathological cardiac structures.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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