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Dual Left Pulmonary Venous Drainage in a Child with Right Atrial and Right Ventricular Dilatation

Nikolaos G Eleftherakis* and Nikolaos D Andreou

Department of Pediatric Cardiology, 'Agia Sofia' Children's Hospital, Athens, Greece

Abstract

We report the case of dual pulmonary venous drainage, normal to left atrium and abnormal to left innominate vein, as a very rare cause of right ventricular dilatation in an otherwise normal and asymptomatic child.

Keywords: Dual drainage; Left pulmonary vein

Introduction

Anomalies of pulmonary venous drainage result in increased preload of the right heart thus causing right atrial and ventricular dilatation. Total anomalous drainage with obstruction causes symptoms in early infancy and usually need urgent surgical correction [1]. If there is no obstruction and there is interatrial communication or if the anomalous drainage is partial, symptoms are not present until childhood or adulthood [2]. There are five subtypes of partial anomalous pulmonary venous connection as described by Alsoufi et al. [3]. The case we present is rarer because there is not only anomalous connection but coexists with normal drainage. Clinical and echocardiographic findings are similar.

Case Presentation

An 8 year old boy was examined in our department in terms of screening for participation in sports. He only complaints of very slight fatigue on exercise. A mild systolic murmur was audible in the left upper sternal edge and also wide split of the second cardiac tone. There was right QRS complex axis deviation on resting electrocardiogram and increased cardiothoracic ratio on chest X-ray. Transthoracic echocardiogram with color Doppler imaging showed dilatation of the right cardiac chambers with intact atrial septum (Figure 1). The pulmonary veins appeared to drain normally to the left atrium. There was however increased flow in the superior vena cava and at suprasternal view a vertical vein with flow to the left innominate vein was observed (Figure 2).

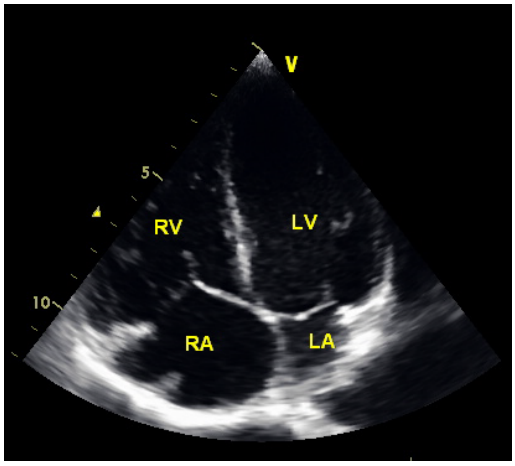

Figure 1: Two dimensional transthoracic echocardiographic four chamber apical view of the child showing dilated right atrium and ventricle. RV=Right Ventricle, RA=Right Atrium, LV=Left Ventricle, LA=Left Atrium.

The boy underwent a diagnostic right cardiac catheterization. Angiography revealed that the left common pulmonary vein connected normally to the left atrium (Figure 3). There was however in addition anomalous drainage to the left innominate vein through a vertical vein (Figure 4). Pulmonary artery pressure was normal (systolic-diastolic -mean) 24-12-17 millimeters of mercury, as was the pulmonary vascular resistance (2.1 Wood units per square meter of Body Surface Area). Significant left to right shunt was found with a pulmonary to systemic blood flow ratio (Qp/Qs) 1.7: 1, thus explaining dilatation of right atrium and ventricle.

Discussion

There is no classification system proposed for double pulmonary venous connection. Two anatomic subtypes have been reported in literature:

- Drainage of right middle and lower pulmonary vein into inferior vena cava and left atrium in scimitar syndrome [4,5].
- Drainage of left upper lobe to the innominate vein via a large

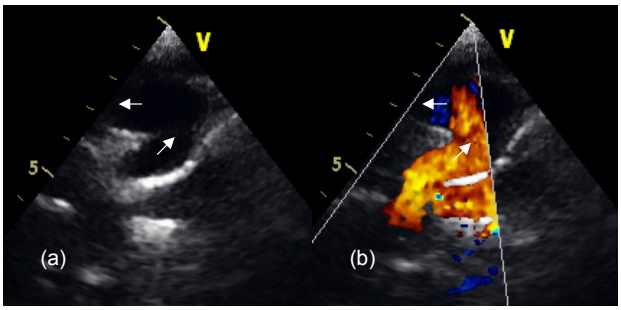

Figure 2: Suprasternal two dimensional echocardiographic image of the abnormal connection to the left innominate vein (a). Color Doppler image of the abnormal drainage to the left innominate vein (b). Arrows show the direction of blood flow. These echocardiographic images are the respective of the angiographic images of Figure 3.

***Corresponding author:** Nikolaos G Eleftherakis, Department of Pediatric Cardiology, Agia Sofia Children's Hospital, Thivon and Papadiamantopoulou St, 115 27 Goudi, Athens, Greece, Tel: 00302107467105; Fax: 00302107797649; E-mail: ngeleftherakis@hotmail.com

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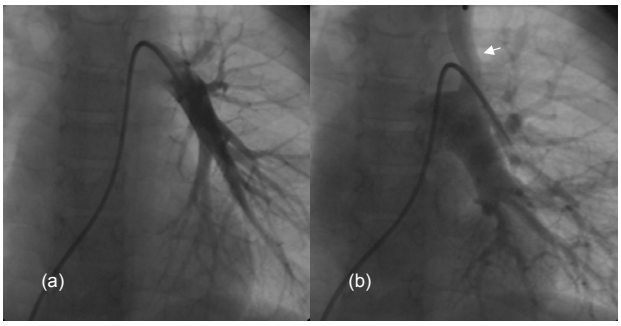

Figure 3: Angiogram of the left pulmonary artery (a) and left pulmonary vein (b). A vertical vein arising from the pulmonary vein is obvious (arrow).

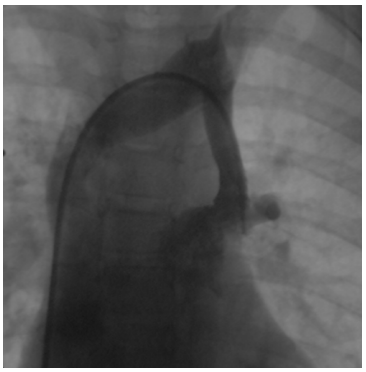

Figure 4: Angiogram of the left pulmonary vein through the left innominate vein. There is normal drainage to the left atrium (single arrow) and abnormal to the superior vena cava (double arrow).

vertical vein (left superior cardinal vein) and to the left atrium via the left upper pulmonary vein [5].

The case described is less common because there is dual venous drainage of the whole left lung. Cardiac sequelae are that of a significant interatrial shunt. Pediatric patients usually have no symptoms but patients over 40 years of age are symptomatic at presentation [5].

Thus in the absence of atrial septal defect this anomaly should be considered in case of right heart dilatation in asymptomatic children. The vast majority of such anomalies were treated surgically in the past. Currently, most of cases undergo transcatheter repair with the use of vascular occlusion devices [6-8].

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