

Dilated coronary sinus due to persistent left superior vena cava in a healthy athlete: A case report with brief review

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Abstract

The left superior vena cava is a persistent embryo-genetic variant (PLSVC), in which the venous blood of the upper half of the body is drained through a left vena cava into the coronary sinus and/or through the atrio-ventricular groove in the right atrium. PLSVC is an uncommon but clinically relevant venous congenital abnormality which is encountered by non-invasive cardiovascular imaging. Its recognition is important when an enlarged coronary sinus occurs and consequently a differential diagnosis must be made. PLSVC should always be suspected in presence of a dilated coronary sinus, incidentally discovered by echocardiography. Here the author describes the incidental discovery of a dilated coronary sinus due to persistent left superior vena cava in a healthy 46-year-old male athlete during sports pre-participation screening, and discuss the importance and usefulness of the combination of non-invasive cardiovascular examinations, i.e. echocardiography and multi-slice computed tomography, to make an optimal diagnosis and illustration of congenital cardiovascular anomalies.

Introduction

Persistent left superior vena cava (PLSVC) is an uncommon congenital abnormality but it is the commonest congenital venous anomaly of the thorax cause of dilated coronary sinus (CS), with a prevalence of approximately 0,5% in the general population [1]. The coronary sinus is subject to a variety of congenital abnormalities [2]. A persistent LSVC should be suspected when a dilated coronary sinus is identified via echocardiography. Additional etiologies of a dilated coronary sinus include elevated right atrial pressure, partial anomalous pulmonary venous return to the coronary sinus, and coronary arteriovenous fistula [3]. PLSVC carries coronary sinus blood retrograde into the left innominate vein [4], and in most cases it drains into the right atrium through the coronary sinus in the absence of congenital heart disease [5]. The coronary sinus enlarges when it receives a left superior vena cava or a hepatic vein, when it is joined by a left superior vena cava that receives blood from the inferior vena cava via the hemiazygos vein. This venous anomaly is often present in asymptomatic individuals and is discovered incidentally during cardiovascular imaging [6], device implantation [7], or surgery [8]. Here the author describes the incidental discovery of a dilated coronary sinus due to persistent left superior vena cava in a healthy 46-year-old male athlete during sports pre-participation screening, and discusses the importance and usefulness of the combination of non-invasive cardiovascular examinations to make an optimal diagnosis and illustration of congenital cardiovascular anomalies.

Case report

A 46-year-old male athlete was referred to our sports center for pre-participation screening. The athlete's past family and medical history was unremarkable. The cardio-respiratory physical examination was normal except for a mild diastolic murmur at the lower left sternal border. Resting ECG was normal and an exercise stress test showed sporadic premature ventricular contractions (PVCs) without ST-segment changes and good coronary reserve. The athlete underwent bi-

dimensional transthoracic echocardiography (2D-TTE) utilizing harmonic imaging and Doppler evaluation by commercially available ultrasound equipment (MyLab 30 Gold Esaote). Echocardiographic examination showed normal size and function of the left cardiac chambers, but revealed a marked dilated coronary sinus, well visualized both in classic left parasternal long axis view (Figure 1 Panel A) and in apical 4 chamber view with posterior tilt (Figure 1 Panel B), and also showed mild enlargement of the right atrium. Apparent inter-atrial shunt or significant valvular disease was not detected. In addition, a mild aortic root dilatation with minimal aortic insufficiency was seen (Video 1). The athlete refused the injection of agitated saline solution to demonstrate opacification of the coronary sinus, so it was decided to make a multi-slice computed tomography (MSCT) with angiography to confirm the suspected diagnosis of persistent left superior vena cava and to rule out other abnormalities, such as an atrial septal defect, coarctation of the aorta, and anomalous pulmonary venous return. Subsequently an MSCT – angiography (Figure 2/Figure 3 Panel B) completed by 3-D volume-rendered reconstruction and coronal image (Figure 2/Figure 3 Panel A) showed an opacified persistent left sided superior vena cava which drains into the coronary sinus that appeared dilated (max diameter 14 mm). Also the innominate vein and mild aortic root dilatation with patent coronary artery are visualized. No other anomalies were detected. These findings did not affect the physical activity, therefore the athlete was judged fit and eligible for sports competition, with annual follow up.

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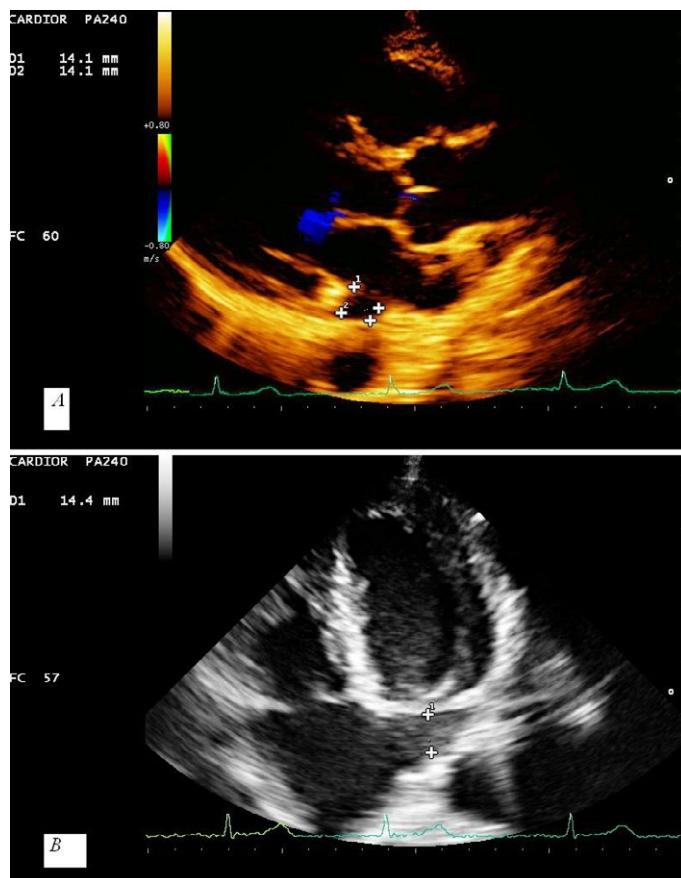
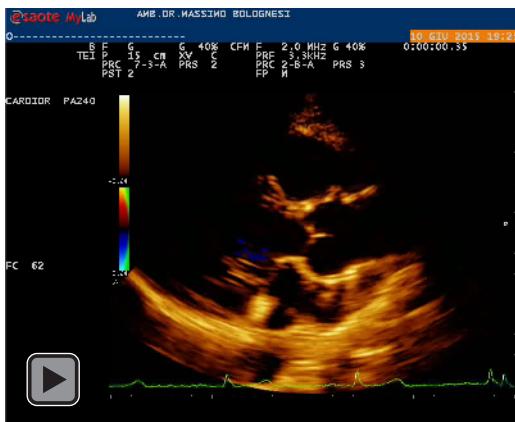


Figure 1. (Panel A) TTE in left parasternal long-axis view showing a dilated coronary sinus (diameter >14 mm) and a mild aortic root dilatation; (Panel B) From this apical 4 chamber view with posterior tilt, a dilated coronary sinus is clearly evident (*).



Video 1. TTE with colour Doppler in left parasternal axis view shows a marked dilated coronary sinus, a mild aortic root dilatation and a mild aortic insufficiency.

Discussion

Persistent left superior vena cava (PLSVC) is a rare congenital anomaly but it is the most common abnormality of thoracic venous system and the commonest cause of dilated coronary sinus [1]. Two types of PLSVC are known in literature: in 92% of cases, PLSVC is connected to the right atrium through the coronary sinus without significant hemodynamic consequences, while in 8% of cases, PLSVC connects directly or through the pulmonary veins to the left atrium,

causing a right to left shunt [9]. It is most commonly an isolated finding but can be associated with other congenital heart defects. Normally, during the 8th week of gestation, portions of the left cardinal venous system regress to become the ligament of Marshall. If this regression fails, a persistent LSVC occurs and drains, typically without hemodynamic consequence, to the right atrium via the coronary sinus [10].

A persistent LSVC should be suspected when a dilated coronary sinus is identified via echocardiography. This is often discovered incidentally during an echocardiographic examination or other cardiovascular imaging, because it is almost always asymptomatic. Therefore the evidence of a dilated coronary sinus must always be investigated in search of a congenital malformation and especially the persistence of a left superior vena cava. In the setting of sports medicine trans-thoracic echocardiography (TTE) is an excellent non-invasive method to identify a dilated coronary sinus and therefore suspect PLSVC, as demonstrated by the case report above described. Usually, agitated saline injected into the brachial left vein drains through the abnormal connection between the left-sided venous return and the CS, where the bubbles can be clearly seen first in the CS and in the successive opacification of the right atrium. Contrast echocardiography can also be performed [11]. However, considering that physicians should attempt to rule out causes of right atrium pressure/volume overload such as atrial septal defect, coronary fistulas and other anomalous

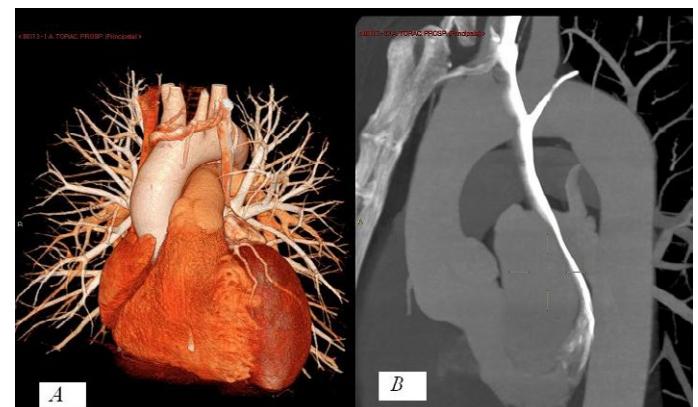


Figure 2. (Panel B) MSCT angiography and (Panel A) from 3-D volume rendered reconstruction showing opacified persistent left sided superior vena cava which drains into the coronary sinus.

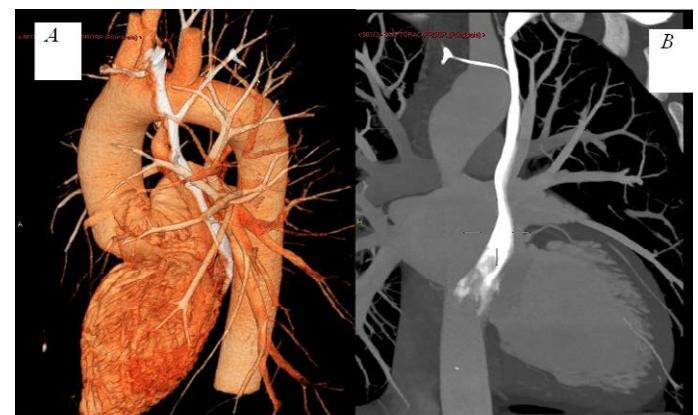


Figure 3. (Panel B) MSCT angiography and (Panel A) from 3-D volume rendered reconstruction showing opacified persistent left sided superior vena cava which drains into the coronary sinus.

drainage pattern into the CS, nowadays MSCT is more frequently applied by cardiologists and radiologists to visualize coronary anatomy, coronary anomalies and aortic pathology. PLSVC has important clinical implications, although it is a benign condition. First of all, it can be associated with a variety of congenital malformations of the heart and great vessels, and in the second instance it may technically complicate some endovascular and surgery procedures. For example during cardiac surgery, administration of retrograde cardioplegia is hampered by the presence of a persistent left superior vena cava, which could complicate the surgery. Otherwise, when the left subclavian vein is used to access, serious complications such as arrhythmias, shock, and coronary sinus thrombosis may occur [12]. Furthermore, pacemaker (PM) or implantable cardioverter defibrillator (ICD) placement can be difficult and dangerous.

Conclusion

For the above mentioned reasons this case-study review provides another contribution to the knowledge of this congenital anomaly and supports the importance of echocardiography in the diagnostic setting of sports cardiology medicine. Furthermore, it attests the utility of the combination of non-invasive cardiovascular imaging, namely echocardiography and computed tomography, for a better and more complete definition of the anatomical and functional cardiovascular abnormalities [13-16].

Conflict of interest

The author does not report any conflict of interest regarding this work

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