

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Persistent left superior vena cava: Case report ☆☆☆

Khadija Laasri, MD^{a,*}, Soumya El graini, MD^a, Hiba Zahi, MD^a,
 Ismail Mohamed Halfi, MD^a, Houda Bachri, MD^b, Emad Aldin Massri, MD^b, Sara Zebbar^b,
 Aida Soufiani^b, Nadia Fellat^b, Nesma Bendagha^b, Rokya Fellat^b, Ittimade Nassar^a,
 Nabil Moatassim Billah^a

^a Radiology Department, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco^b Cardiology A Department, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco

ARTICLE INFO

Article history:

Received 1 September 2022

Revised 16 September 2022

Accepted 21 September 2022

Keywords:

Persistent superior vena cava left
 Abnormal systemic venous return
 CT angiogram
 TTE

ABSTRACT

Persistent left superior vena cava (PLSVC) is a rare anomaly of the systemic venous circulation. We report the case of a 22-year-old female that had history of multiple repair surgeries for her esophageal atresia, as well as a right lobectomy for bronchiectasis 15 years prior. She was admitted to the surgical ward for complete resection of the right lung. A trans-thoracic echocardiography was performed as part of the pre-surgical work-up and it revealed a dilated coronary sinus which led us to suspect the presence of a PLSVC. The latter was confirmed by a simple “Bubble study” and confirmed by CT angiogram. We will discuss throughout this paper, the clinical and radiological features, as well as the embryology of this anomaly, so that the knowledge of the existence of this anatomical variant, especially if surgery or catheterization is at reach of the medical team, may lead to avoid serious complications.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

[\(http://creativecommons.org/licenses/by-nc-nd/4.0/\)](http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Persistent left superior vena cava (PLSVC) is a rare anomaly of the systemic venous circulation [1]. Its prevalence is estimated at 0.3% in subjects with a normal heart [2]. Often asymptomatic, PLSVC is discovered incidentally during car-

diovascular imaging. It may lead, however, to serious complications like arrhythmias and cyanosis. PLSVC also has serious implications in patients undergoing percutaneous venous interventions or heart surgery [3]. In this article, we report the incidental discovery of this anomaly during a routine pre-surgical echocardiography, the utility of contrast agents, and the confirmational role of CT angiogram.

☆ Guarantor of Submission: The corresponding author is the guarantor of submission.

☆☆ Competing Interests: The authors declare that they have no conflicts of interest.

* Corresponding author.

E-mail address: Kadjjalaasri93@gmail.com (K. Laasri).<https://doi.org/10.1016/j.radcr.2022.09.076>1930-0433/© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

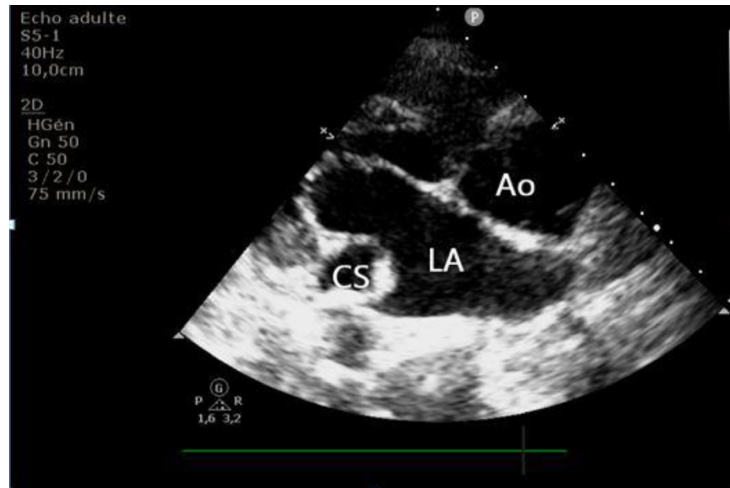


Fig. 1 – Transthoracic echocardiogram, parasternal long axis view, illustrating a dilated coronary sinus (CS).

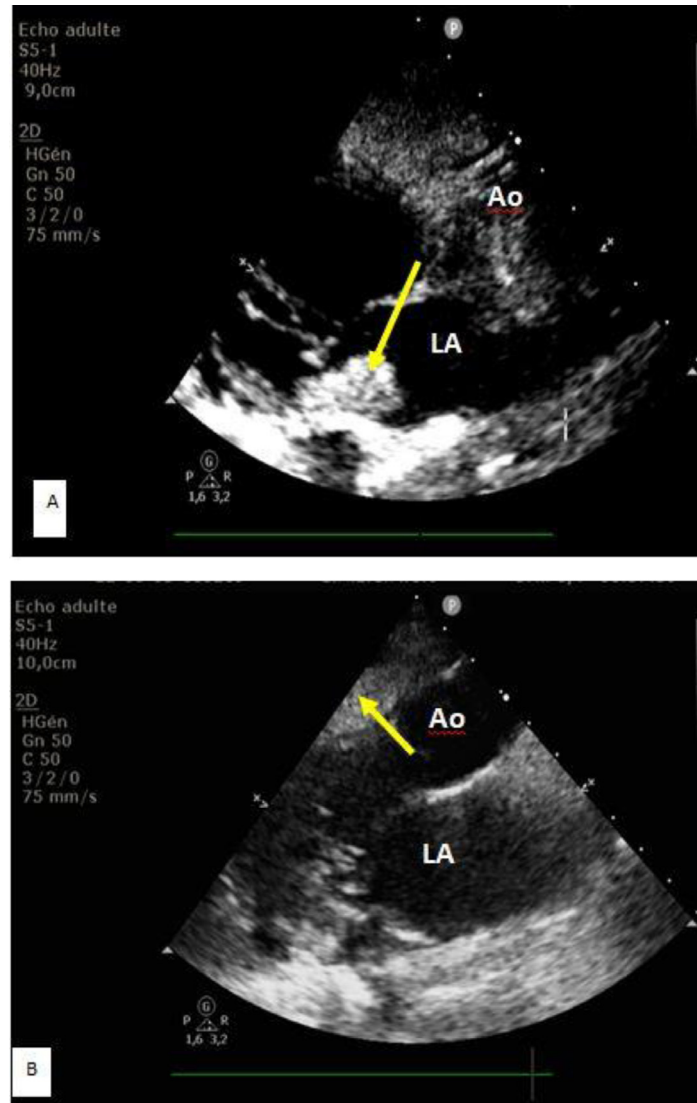


Fig. 2 – Injection of agitated saline in the left arm with bubbles (yellow arrow) appearing first in the coronary sinus (A), and later into the right atrium. (B) Coronary sinus draining into an enlarged right atrium.

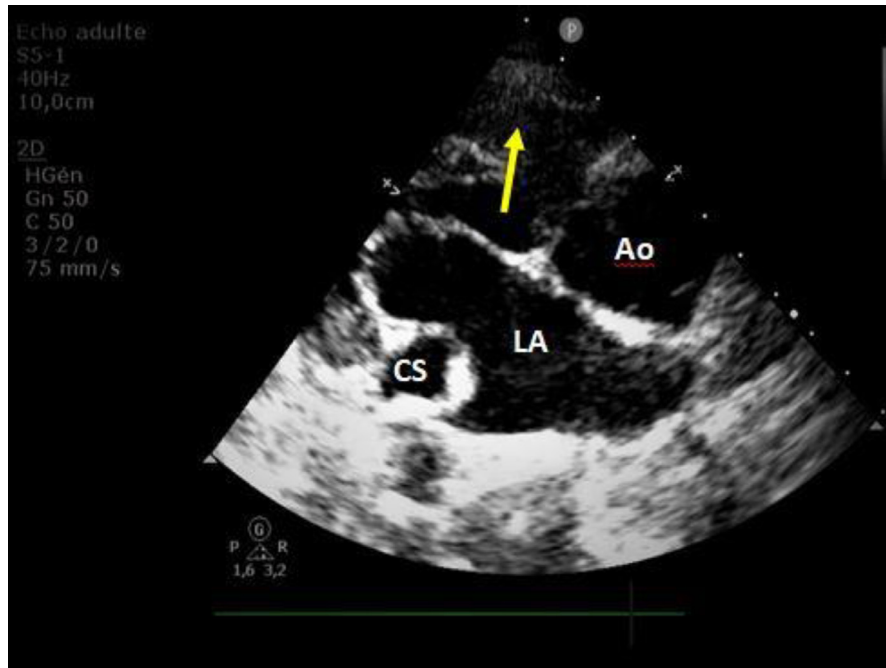


Fig. 3 – Bubble study showing the normal connection of right superior vena cava (RSVC) by early appearance of saline contrast in the right ventricle (RV) (yellow arrow) –right arm injection (parasternal long axis view).

Case report

We report the case of a 22-year-old female admitted to the surgical ward for right lung pneumonectomy for chronic lung infection, with history of multiple surgical interventions for her esophageal atresia in addition to a history of right lobectomy for bronchiectasis 15 years prior. Transthoracic echocardiogram (TTE) was performed to evaluate the right heart as part of the pre-surgical work-up which end up revealing a dilated coronary sinus leading to suspect a PLSVC (Fig. 1).

Then we performed a Bubble study, consisting of injecting an agitated saline solution into the left antecubital vein. Following the trajectory of the saline solution, as it went through an abnormal sequence; a clear inflow of the solution in the coronary sinus first then in the right heart chambers (Fig. 2). As such we were able to confirm the existence of a left superior vena cava that drains into the coronary sinus and joins the right atrium. Whereas the right superior vena cava drains directly into the right atrium as shown by the contrast injection in the right antecubital vein (Fig. 3).

A CT angiogram was performed to confirm the diagnosis, showing a normal sized heart, normal caliber of the pulmonary arteries, aorta, and a modal distribution of the supra-aortic trunks. The superior vena cava and the azygos system was without abnormality. However, in the left para aorta, a vascular structure was identified, following the left subclavian vein. This structure, descending along the left side of the aortic arch, crossing in front of the pulmonary artery and the junction of the left pulmonary veins to drain into the coronary

sinus which led to the right atrium, consistent with PLSVC (Fig. 4), a collateral was present connecting it to the right superior vena cava (Fig. 5). Parenchymal window shows a destroyed right lung with foci of dilated supra infarcted branches (Fig. 6). We retained as a diagnosis a double superior vena cava by persistence of a left vena cava and a destroyed right lung with foci of dilated supra infarcted branches.

Discussion

Knowledge of the normal anatomy of the large vessels of the neck and thorax is fundamental for any clinician. The variants of the superior vena cava are not well known. John Marshall (1818–1891), an English surgeon and professor of anatomy at the University College Hospital in London, published the first description of the large anterior veins of the thoracic region in humans and mammals in 1850 [4]. Since the majority of these congenital venous malformations are asymptomatic, it is difficult to estimate their prevalence in the general population [4,5]. According to several authors, it is estimated to be about 0.3%-0.5% in the general population [6,7], it is increased in patients with congenital cardiovascular disease and ranges from 1.3% to 12.9% [7].

The congenital cardiac disorders found in case of persistence of the left superior vein are atrial septal defect and ventricular septal defect, followed by coarctation of the aorta, transposition of the great vessels, tetralogy of Fallot and abnormal pulmonary vein connections [7]. On the other hand,



Fig. 4 – CT angiogram of chest in axial section (A), sagittal reformatted scan (B), and coronal reformatted scan (C), showing a vascular structure following the left subclavian vein (Fig. 2A). This structure, descending along the left side of the aortic arch (Fig. 4B), consistent with PLSVC (Yellow arrow). Note the right superior vena cava in place (Fig. 2C) (Red arrow).

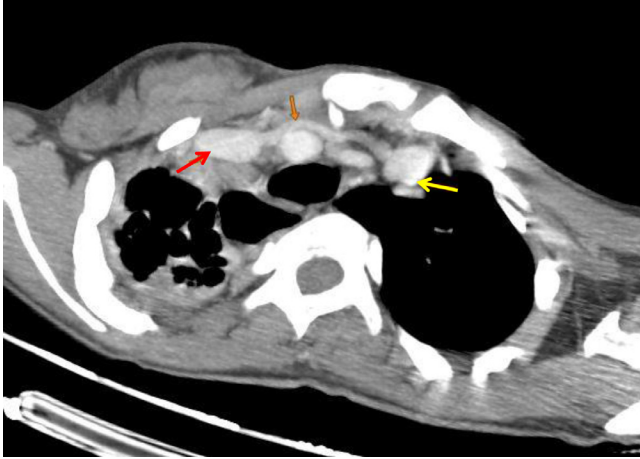


Fig. 5 – CT angiogram of chest in the mediastinal parenchyma window, in axial section showing a collateral (orange arrow), connecting the 2 superior vena cava left (yellow arrow) at right (red arrow).

the most common extracardiac anomaly is esophageal atresia [5], as it is the case with our patient.

Embryologically, in the normal state, the latter is the result of 2 pairs of symmetrical venous systems: the common cardinal veins and the anterior cardinal veins. Persistent CSGV results from the absence of involution of the left anterior cardinal vein [8] (Fig. 7 is a schematic representation of the developmental anatomy [9]). The disposition of the azygos and hemiazygos veins can also be variable in these situations. There are 3 primary categories for vena cava anatomical abnormalities. Type I relates to the anatomy of the normal central vein. Type II includes the 10%-20% of patients with persistent CSGV who do not have a right superior vena cava [5,6]. However, a right superior vena cava, which is type III and known as double vena cava, is present in 80%-90% of cases. This implies that the CSGV and the GSV coexist. In about 35% of these patients, these 2 veins are connected by the innominate left brachiocephalic vein, which is classified as type IIIa. This innominate vein is absent in type IIIb [5]. In our case, it was type IIIa (Fig. 3). In terms of anatomical position, the persistent CSGV descends vertically in front of the arch of the aorta and lat-

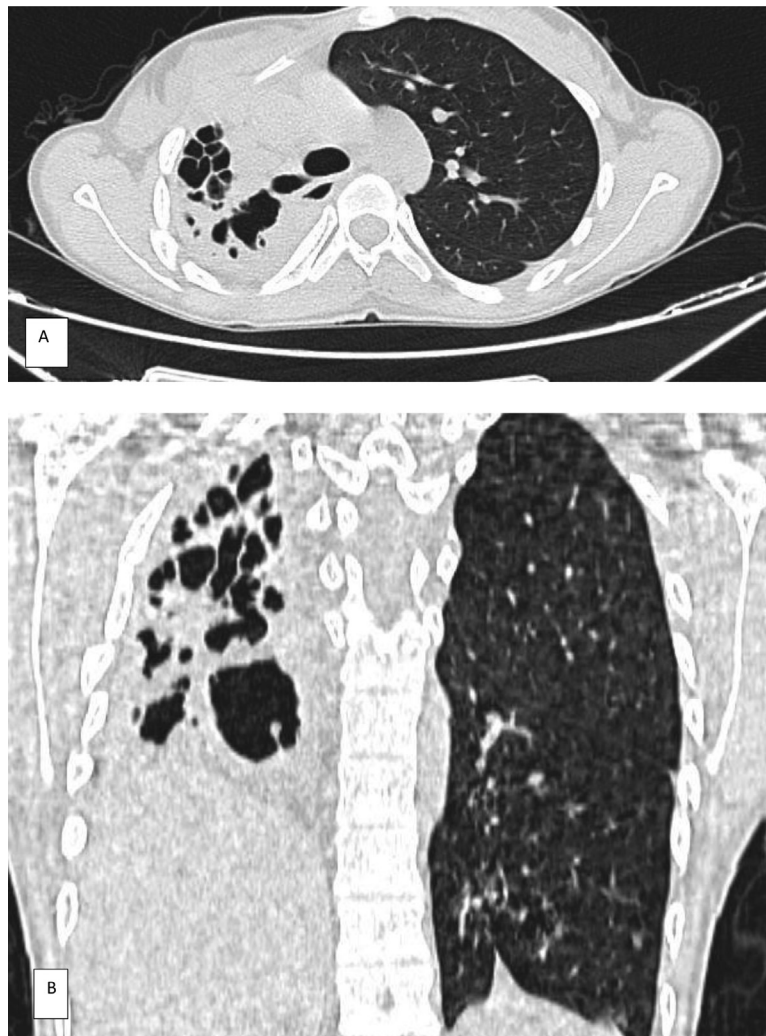


Fig. 6 – Angio computed tomographic scan of chest in the parenchyma window, in axial section (A), and coronal reformatted scan (B), showing destroyed right lung with foci of dilated supra infarcted branches.

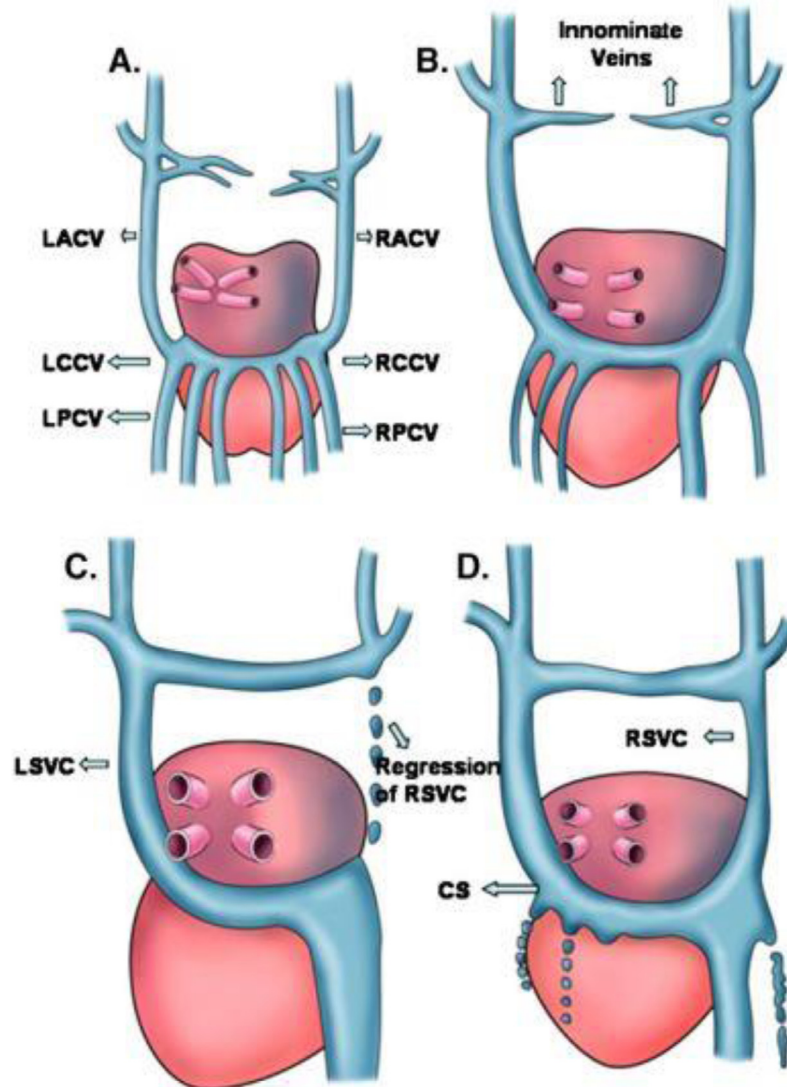


Fig. 7 – Developmental anatomy of the persistent left superior vena cava as viewed from the posterior aspect of the heart. (A) Pairs of anterior and posterior cardinal veins draining into the embryonic heart via the right and left common cardinal veins. (B) Development of bridging innominate vein connecting left and right anterior cardinal veins during eighth week of gestation. (C) Regression of the right-sided superior vena cava with persistence of left-sided superior vena cava as a single superior vena cava that drains the cephalic portion of the body including upper extremities. (D) Right-sided superior vena cava connected with the persistent left superior vena cava via innominate vein in the post-natal heart. CS, coronary sinus; LACV, left anterior cardinal vein; LCCV, left common cardinal vein; LPCV, left posterior cardinal vein; LSVC, left superior vena cava; RACV, right anterior cardinal vein; RCCV, right common cardinal vein; RPCV, right posterior cardinal vein; RSVC, right superior vena cava [9].

erally to the left to the left vagus nerve and courses anterior to the left pulmonary hilum to drain into the right atrium through the coronary sinus in 92% of cases, but in the other 8% of cases, it opens into the left atrium, even though the coronary sinus is normally developed, creating a right-left shunt or into the right superior vena cava [6,8].

Depending on the drainage site and associated cardiac anomalies, the clinical significance of a PLSVC will vary from asymptomatic to severe cyanosis, arrhythmias, and paradoxical embolism [10]. The coronary sinus is usually dilated when the PLSVC drains into the right atrium, leading to the com-

pression of the atrio-ventricular node and his bundle and may cause arrhythmias [1]. In patients with associated congenital anomalies, the symptoms are more related to these associated anomalies [11]. In the rare cases of direct draining into the left atrium, it can cause cyanosis [8] and a risk of paradoxical embolism [6].

Awareness of a PLSVC is capital in advance in patients undergoing invasive interventions such as cardiac resynchronization therapy (CRT), pacemaker implementation or central venous catheterization as it may complicate these procedures [12].

Some cardiac surgeries may also be hindered by the PLSVC requiring special management. For example, during cardiopulmonary bypass, PLSVC will be responsible for a blood surplus return and insufficient venous return to the pump [1].

The standard radiograph is most often normal or may show an enlargement of the left upper mediastinal space [15]. VSCG is often found incidentally during a thoracic CT scan with enhancement. Thus, the diagnosis is established by the presence of a left mediastinal tubular opacity that draining into the coronary sinus in front of the aortic arch and shows vascular contrast enhancement dynamics [8,13].

MRI can easily, on axial and coronal images, demonstrate a left superior vena cava as an abnormal vascular structure attached to the left lateral side of the aortic arch. The fine-axis sections, moreover, confirm the drainage of the aberrant vessel into a dilated coronary sinus [14].

Conclusion

The presence of a dilated coronary sinus on echocardiography should alert the clinician towards the possibility of PLSVC. In addition to saline contrast echocardiography, angio scan is crucial in establishing the diagnosis of this entity when it is suspected or can be discovered accidentally while performing a CT scan for a further pathology.

Patient consent

Written informed consent for publication was obtained from patient.

REFERENCES

- [1] Azizova A, Onder O, Arselan S, Harizolan T. Persistent left superior vena cava: clinical importance and differential diagnoses. *Insights Imaging* 2020;11(1):110. doi:10.1186/s13244-020-00906-2.
- [2] Perles Z, et al. Prevalence of persistent superior vena cava and association with congenital heart anomalies. *Am J Cardiol* 2013;112(8):1214–18. doi:10.1016/j.amjcard.2013.05.079.
- [3] Hutyra M, et al. Persistent left superior vena cava connected through the left upper pulmonary vein to the left atrium: an unusual pathway for paradoxical embolization and a rare cause of recurrent transient ischaemic attack. *Eur Heart J Cardiovasc Imaging* 2010;11(9):E35. doi:10.1093/ejehoccard/jeq079.
- [4] Ruesch S, Walder B, Tramer MR. Complications of central venous catheters: internal jugular versus subclavian access a systematic review. *Crit Care Med* 2002;30:454–60.
- [5] Waldemar I, Piotr G, Jaroslaw K. Haemothorax as a complication of subclavian vein cannulation with a haemodialysis: a case report. *Anaesthesiol Intensive Ther* 2013;45(2):89–92 ISSN 1642–5758.
- [6] Mballa Amougou JC, Mbo Amvene J, Jemea B, Magny Tiam E, Menanga A, Kaze Folefack P, et al. Veine Cave Supérieure Gauche : à Propos de Trois Cas. *Health Sci Dis* 2018;19(4). Retrieved from <http://www.hsd-fmsb.org/index.php/hsd/article/view/1211>.
- [7] Sarodia BD, Stoller JK. Persistent left superior vena cava: case report and literature review. *Respir Care* 2000;45(4):411–16.
- [8] Laamrani FZ, Lahkim M, Laabioui R, El Quessar AJ, Cherkaoui MM. Veine cave supérieure gauche persistante. *Feuill Radiol* 2014;54(3):168–70. doi:10.1016/j.frad.2014.01.004.
- [9] Ratliff HL, Yousufuddin M, Lieving WR, Watson BE, Malas A, Rosencrance G, et al. Persistent left superior vena cava: case reports and clinical implications. *Int J Cardiol* 2006;113(2):242–6. doi:10.1016/j.ijcard.2005.08.067.
- [10] Nagasawa H, et al. Incidence of persistent left superior vena cava in the normal population and in patients with congenital heart diseases detected using echocardiography. *Pediatr Cardiol* 2018;39(3):484–90. doi:10.1007/s00246-017-1778-3.
- [11] Erdoğan M, Karakaş P, Uygur F, Meşe B, Yamak B, Bozkır MG. Persistent left superior vena cava: the anatomical and surgical importance. *West Indian Med J* 2007;56:72–6.
- [12] Demos TC, et al. Venous anomalies of the thorax. *Am J Roentgenol* 2004;182(5):1139–50. doi:10.2214/ajr.182.5.1821139.
- [13] Cosson S, Kévorkian JP, Roux O, Fressonnet R, Leenhardt A, Beaufile P. Veine cave supérieure gauche : un diagnostic simple par échocardiographie transthoracique. A propos d'un cas. *Ann Cardiol Angiol* 2002;52:194–7.
- [14] Kastler B, Livolsi A, Germain P, Bernard Y, Michalakakis D, Rodiere E, et al. Apport de l'IRM dans l'exploration des anomalies cardiaques congénitales et des gros vaisseaux. *J Radiol* 2004;85(10):1821–50. doi:10.1016/s0221-0363(04)97754-2.
- [15] Ghadiali N, Teo LM, Sheah K. Bedside confirmation of a persistent left superior vena cava based on aberrantly positioned central venous catheter on chest radiograph. *Br J Anaesth* 2006;96(1):53–6. doi:10.1093/bja/aei272.