



Prevalence of left superior vena cava in patients with congenital heart disease

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Abstract

Background: Persistent Left Superior Vena Cava (PLSVC) is an anomaly of systemic venous return with a prevalence of 0.1% to 0.3% in general population and about 3 to 10% in case of Congenital Heart Disease (CHD). This congenital anomaly is without hemodynamic consequence if it's isolated. Its recognition is important in many circumstances including cases of open heart cardiac surgery during cannulation and in case of central venous catheterization maneuvers by upper left channel.

Goal: The aim of this study was to evaluate the prevalence of PLSVC among patients with Congenital Heart Disease (CHD).

Methods: In this retrospective study, 1923 patients with CHD were collected from all patients who underwent transthoracic echocardiography at our institution between December 2008 and January 2014. Variables are presented as average, median and number of patients observed (percentage). A p-value <0.05 was used as level of significance

Results: Among 4034 patients examined, 1923 had CHD. The persistent LSVC was present in 76 patients of which 71 had CHD with a prevalence of 3.7%. The most common cardiac defect associated with persistent LSVC was atrial septal defect (ASD) (32.3%) (OR 1.60, 95% CI 0.95-2.68, $p = 0.07$). Among the five patients having LSVC in the absence of cardiac malformation (prevalence 0.2%), one had iridocoloboma, a second had a mucopolysaccharidosis and the other patients had no extra-cardiac anomalies.

Conclusion: The persistence of LSVC is a rare entity that should be sought systematically, especially in case of any heart defects or extracardiac malformation.



Introduction

The persistence of Left Superior Vena Cava (LSVC) was reported for the first time in literature by Chat in 1738 [1]. Its prevalence in general population is 0.1-0.3% [2,3]. The prevalence of LSVC was little understood and vary depending on the diagnostic tool used for patients with congenital heart disease. Its frequency is 5% by echocardiography and 11% by angiography [4-7]. Persistent left superior vena cava results from the failure of the left cardinal vein to regress in the embryo. In normal development, the left superior vena cava disappears, only its cardiac termination remaining as the coronary sinus.

The importance of the diagnosis of LSVC is the possibility of occurrence of complications in patients requiring cardiac surgery or a central venous catheterization. Thus, in case of surgery the LSVC cannulation can cause difficulties for extracorporeal circulation or realization of retrograde cardioplegia [8]. Technical difficulties may be encountered when first venous subclavian or internal jugular left during application of Swan-Ganz catheter, a myocardial biopsy or the establishment of a pacing probe in the right ventricle by LSVC especially in the absence of right superior vena cava [9].

They are due to the acute angle formed between the orifice of the coronary sinus and the tricuspid valve. The purpose of this study was to evaluate the frequency of LSVC in population admitted for congenital heart disease screening.

Patients and methods

All patients referred to the unit of pediatric cardiology of our institution for echocardiography, from December 2008 to January 2014, were included. 4034 patients were evaluated by trans-thoracic echocardiography (using GE Vivid 7 system and transducers 3S 4MHz and 7S 8MHz) using standard views according to ASE echocardiography guidelines. Patients were referred from peripheral hospitals, intensive care units of neonatology, pediatric intensive care units, units of general pediatrics and cardiology. Classification of congenital heart disease was made based on anatomical [10], physiopathological [10,11] and embryological classifications [12-14]. LSVC screening was systematic in each of our echocardiographic assessment. The identification of LSVC was established by the combination of echocardiographic cuts. The LSVC was visualized by suprasternal and long parasternal incidence. The diagnosis of LSVC was suspected in the presence of coronary sinus dilatation. The characteristic aspect of the dilated coronary sinus corresponds to a rounded picture, anechoic, located in atrio-ventricular groove, at the back of the left atrium, clearly seen in long parasternal axis cut. It can be also visualized in the apical 2 chambers view, or between the right and the left atrium in apical 4 chambers, orientated in a posterior plan.

In addition to two-dimensional echocardiogram the color mapping and the Doppler has an important role to spot it and to be able to follow it until sometimes its termination

In adults with unsatisfactory transthoracic window. an injection of agitated saline solution during the echocardiogram can be done to search the PLSVC.

Statistical analysis

The analysis of data was made possible with the statistical software SPSS (SPSS inc, version 21). Variables are presented as average, median and number of patients observed (percent-

age). A p-value <0.05 was used as level of significance. Odds ratios were calculated at a 95 confidence interval (CI) for the presence of persistent LSVC or absence of CHD, and for an association to a specific diagnosis.

Results

A total of 4034 patients ranging in age from 1 day to 75 years were examined. All had a complete cardiac evaluation. From this total, 1923 had congenital heart disease. The persistence of LSVC was present in 76 patients of which 71 had congenital heart disease or a prevalence of 3.7%. The age range of patients with LSVC was 2 days to 42 years (median age of 3 years). The sex ratio was 1. The most associated congenital heart disease with persistence of LSVC was isolated atrial septal defect (32.3%) (OR 1.60, 95% CI 0.95-2.68, $p = 0.07$). Persistence of LSVC was significantly associated with tetralogy of Fallot (TOF) (OR 2.06, 95% CI 1.03-4.12, $p = 0.035$). Any congenital heart disease was likely to be associated with LSVC as shown in Table 1. All patients with LSVC had a right superior vena cava. One case of LSVC draining directly into the left atrium was found. Five patients had no heart defect. Fifteen patients had extracardiac anomalies including trisomy 21, anorectal malformation, esophageal atresia, iridocoloboma, convergent strabismus, club foot, Hurler syndrome (Table 2).

Table 1: Congenital heart disease associated with left superior vena cava

Cardiac malformations	Frequency of LSVC(n)	Frequency of LSVC(%)
AVSD	20	28,1
VSD	20	28,1
ASD	23	32,3
PDA	15	21,1
PA, PS, DCRV	16	22,5
TOF, DCRV avec TOF	9	12,6
TGA, DCRV avec TOF	9	12,6
CoA	2	2,8
BAV, subaortic membrane	2	2,8
MA, congenital mitral stenosis	2	2,8
PAPVD, TAPVD	7	9,8
Interrupted IVC (isolated)	2	2,8
Supramitral ring	2	2,8
Cor triatriatum	1	1,4
Interruption of the aortic arch	1	1,4
PA-VSD	1	1,4

AVSD: Atrioventricular Septal Defect; VSD: Ventricular Septal Defect; ASD: Atrial Septal Defect; PDA: Patent Ductus Arteriosus; PA: Pulmonary Atresia; PS: Pulmonary Stenosis, DCRV: Double Chambered Right Ventricle; TOF: Tetralogy Of Fallot; TGA: Transposition Of The Great Arteries; Co. A: Coarctation Of The Aorta; BAV: Bicuspid Aortic Valve; MA: Mitral Atresia; PAPVD: Partial Anomalous Pulmonary Venous Drainage; TAPVD: Total Anomalous Pulmonary Venous Drainage; IVC: Inferior Vena Cava; PA-VSD: Pulmonary Atresia With Ventricular Septal Defect

Table 2: Extra-cardiac anomalies associated with left superior vena cava

Extra-cardiac anomalies	Frequency of LSVc(n)	Frequency of LSVc(%)
Trisomy 21	6	5,4
Anorectal malformation	4	100
Esophageal atresia	1	100
clubfoot	1	100
Convergent strabismus	1	100
Iridocoloboma	1	100
Hurler syndrome	1	25

Discussion

The importance of the diagnosis of the LSVc lies in:

- Increased risk of paradoxical embolism when venous return is at the left atrium [19,20]
- Very frequent association with other congenital heart disease like VSD, ASD, TOF and TGA .
- Retrograde cardioplegia impossible because the perfusate leaks into the superior vena cava and does not perfuse the heart.
- Risk during central venous catheterization, access to the coronary sinus can cause angina, hypotension or cardiac arrest.
- Difficulty in setting up a pacemaker.

The prevalence of LSVc in congenital heart varies according to the usual diagnostic tool. Buirski et al reported a prevalence of 11% with angiographic diagnosis. Huhta et al reported a prevalence of 1.3% to 5% by collecting all cases of LSVc diagnosed by angiography, echocardiography, or autopsy [10]. This prevalence has doubled from the work of Postema et al made by echocardiographic tool. Our Study shows that the prevalence of LSVc was 3,7% and that was associated with congenital heart disease particularly atrial septal defect, Tetralogy of Fallot, ventricular septal defect and atrioventricular septal defect. The limitation of these studies on the prevalence of LSVc was due to the fact that the main inclusion criterion was the congenital heart disease carriers but in our study we expand the screening to general population. This result is consistent with studies by Ghada and al who found that persistent LSVc was associated to AVSD (11%), ASD (10%), VSD (7%), TOF (5%) [17], Postema and al respectively in 10%, 19%, 35% and 12% [18].

The most frequent lesion association is with the ASD and the TOF. Normal venous development is a process of progressive appearance and regression of a series of paired venous structures. Initially, three venous channels drain into the sinus venosus: the omphalomesenteric (vitelline) veins, the umbilical vein, and the common cardinal veins (ducts of Cuvier). Common cardinal veins form by the join of the superior (anterior) and inferior (posterior) cardinal veins. The left sinus horn and the transverse portion of the sinus venosus, initially horizontal and posterior to the atrium, become separated from the left side of the primitive atrium by the development of a prominent fold, while connecting channels develop between the right and left veins. Regression of the left-sided vitelline and cardinal veins

with increasing prominence of the right sinus horn follows, and the left horn shifts to the right becoming inferior to the right sinus horn. The right sinus horn and a part of the transverse portion of the sinus venosus become incorporated into the wall of the right atrium, allowing the future caval veins to drain directly into the right atrium. A vessel connecting the right and left superior cardinal veins enlarges and becomes the left brachiocephalic (innominate) vein. Consequently, the left superior cardinal vein below this connection gradually obliterates. The proximal portion of the left sinus horn persists as the coronary sinus and the left common cardinal vein persists as the ligament or oblique vein of Marshall. The right superior and common cardinal veins persist and form the right superior caval vein. In the lateralized atrial situs, persistence of the left superior cardinal vein connected to the left atrium is an infrequent occurrence, this vein usually draining into the right atrium via coronary sinus. In atrial isomerism, conversely, the left superior cardinal vein usually persists, draining either into the roof of the left-sided atrium (mainly in right atrial isomerism) or into the coronary sinus (usually in left atrial isomerism). The PLSVC draining into the coronary sinus causes an enlargement of this vessel, and this may interfere with the formation of the posterior wall of the left atrium causing either a coronary sinus-left atrium fenestration (unroofing) or an interatrial communication through the mouth of the coronary sinus (coronary sinus type atrial septal defect). In this situation, the PLSVC either drains into the left atrium, because of the 'unroofing of coronary sinus', or overrides the coronary sinus type septal defect. A possible explanation embryological this is the part isomerism superior cardinal veins.

The persistence of LSVc is usually asymptomatic when venous return is at the level of the right atrium and is discovered incidentally during central venous catheterization. When the return is at the left atrium, it may be the cause of cyanosis and exposed to a risk of paradoxical embolism. In our series, a case of LSVc draining into the left atrium was suspected in a partial absence of the partition between the coronary sinus and the left atrium. In 80-90% of cases LSVc drains into the right atrium through the coronary sinus, causing no symptoms (incidental finding during heart surgery or during imaging tests). In other cases it is brought together into the left atrium causing a right to left shunt with cardiac hemodynamic consequences requiring surgical treatment.

Venous return from the cranial part of the embryo is represented by the two cardinal veins right and left common (right chest and left channel) born from the meeting of cardinal veins upper and lower right and left. The common veins drain into the venous sinuses. During the eighth week of gestation anastomosis forms between the two cardinal veins above (innominate venous trunk). The portion of the upper left cardinal vein downstream of this anastomosis will regress. The anomalies of this step will cause the persistence of LSVc with regression or not the right superior vena cava. The persistence of LSVc is related to the lack of involution of the cardinal vein that normally occurs at the sixth month of uterine life. This anomaly may be isolated or most often associated with congenital heart disease (ASD, TOF, anomalous pulmonary venous drainage or left IVC).

GUSTAPANE et al. have searched for the PSLV on prenatal ultrasound in the fetus showed that PLSV is associated with intra-cardiac and extra-cardiac and chromosomal anomalies in a significant proportion of cases with coarctation of the aorta which was associated with PLSVC in 15% of cases, thus requiring serial follow-up scans during pregnancy [22].

The presence of a LSVC in the absence of right superior vena cava is extremely rare (0.05%) and usually associated with situs inversus [11]. All patients with LSVC had a right superior vena cava (RSVC). In 90% of cases LSVC drains into the coronary sinus, much more rarely in the left atrium either directly or through a pulmonary vein [16]. In our case a series of LSVC draining directly into the left atrium was suspected. The persistence of LSVC is usually asymptomatic when venous return is at the level of the right atrium. When the return is at the left atrium, it may be the cause of cyanosis and exposed to a risk of paradoxical embolism.

In our study there was a high prevalence of LSVC in ASD patients and there was no strong association between ASD and trisomy 21. Six patients (5.4%) had LSVC associated with AVSD. This combination joins the work of Ghada et al. [13]. The low prevalence of LSVC outside a cardiac disease in our study is similar to that of Postema and et al [7].

The LSVC can be viewed directly trans-thoracic echocardiography in certain privileged cases in adults by suprasternal track. Most often, the diagnostic LSVC is suspected in the presence of a dilation of the coronary sinus. The characteristic feature of dilated coronary sinus is a rounded picture anechoic, located in the atrioventricular groove behind the left atrium, well displayed Cup parasternal long axis. It can also be displayed in section between the two cavities or left ventricle and left atrium apical four cavities oriented in a posterior plane. This structure must be differentiated from the descending aorta is more posterior and unlike the coronary sinus located outside the pericardium. Other pathological structures giving an anechoic rounded picture can be observed in the atrioventricular groove: Vascular (aneurysm of the circumflex artery), infectious (abscess of the mitral annulus), tumor (cyst, mediastinal lymph nodes) [21]. However, the clinical context, the increasing impact of TEE and rarely TOE allow an accurate diagnosis in most cases.

The retrospective nature of our study has limitations. As a result, data were missing with incomplete or missing files that were excluded.

Conclusion

The persistence of LSVC is a rare entity that should be sought systematically to heart defects and / or extracardiac and even in normal situation. Echocardiography can be a sensitive tool for the detection of active LSVC. Its diagnosis remains important especially in patients who will benefit from cardiac surgery or a central venous catheterization as well as look for an associated intra-cardiac or extra-cardiac malformation.

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