

Persistent left superior vena cava – considerations in fetal, pediatric and adult populations

Abstract

Persistent left superior vena cava (LSVC) is the commonest congenital anomaly of the thoracic venous system. It is within the group of anomalous systemic venous return (ASVR) and the group is subdivided in cephalic, involving the superior vena cava (SVC) and caudal, involving the inferior vena cava (IVC) types. It is also important to recognise that there can be a persistent LSVC with or without a normal right superior vena cava (RSVC). In most cases, a persistent LSVC drains into the right atrium via the coronary sinus without any clinical symptoms. In this article we discuss embryology, diagnostic and further management approaches and a review of the literature related to persistent LSVC.

Keywords: embryology, fetal echocardiography, inferior vena cava, superior vena cava.

Background

In this modern technical era, with the use of fetal ultrasound (US) and fetal echocardiography, diagnosis of various congenital heart defects (CHD) is more frequent. Although the descriptions of persistent LSVC in the adult date back to 1787, prenatal diagnosis of anomalous systemic venous return (ASVR) has only been reported recently and publications on it still limited.¹⁻⁵ We report three cases of the prenatal diagnosis of persistent LSVC and its practical implications.

Case presentations

A 33-year-old G1P0 was referred to a tertiary care Fetal Medicine Unit with a suspected transposition of the greater arteries. Our study demonstrated normal left ventricular and right ventricular outflow tracts, however, an abnormal three-vessel view. The configuration suggested a persistent LSVC without a RSVC. Detailed examination of the heart revealed a dilated coronary sinus. No other fetal anomalies were identified. The pregnancy was uneventful with an uncomplicated live birth; prenatal findings were confirmed on postnatal assessment.

Our second case was a 35-year-old, G2P1 referred for a growth scan at 36 weeks prior to consideration for vaginal birth after Caesarean section. An incidental finding of a persistent LSVC with RSVC was seen; the heart otherwise appeared normal. Again, an uncomplicated neonatal course was documented.

Our third case, a 31-year-old G2P0, was referred to our unit for management of a dichorionic diamniotic twin pregnancy. At 12 weeks, the nuchal translucencies were discordant,

with the measurement for Twin 2 on the 95th centile. Invasive testing was declined. At 19 weeks, an atrioventricular defect with persistent LSVC and absent RSVC was diagnosed in Twin 2. This twin had the additional findings of cleft lip and palate. Twin 1 developed polyhydramnios in the third trimester. Postnatal review revealed the twins were monozygotic and genetic testing revealed X-linked Opitz G/BBB syndrome, a rare syndrome affecting approximately 1 in 50,000 to 100,000 males. This is the first report on the association of this syndrome and PLSVC to the best of our knowledge. The other interesting observation in this case is also the variable phenotypical expression of this syndrome as is evident in this monochorionic gestation.

Discussion

In the general adult population, the prevalence of persistent LSVC is estimated to be approximately 0.3% to 0.5% in individuals with a normal heart.⁶⁻⁸ As systemic venous return to the heart is often unaffected by the persistent LSVC, the true incidence is unknown. The absence of the RSVC with persistent LSVC is less common and bilateral absent SVCs the least common. Persistent LSVC is associated with congenital heart disease (CHD) and other abnormalities, making the prenatal diagnosis of a persistent LSVC an indication for a detailed assessment of the fetus.

Persistent LSVC and CHD

Persistent LSVC is the most common venous anomaly associated with CHD and up to 3–8% of patients with CHD are reported to have persistent LSVC.^{4,9-12} Reported cardiac abnormalities include heterotaxy (left and right isomerism), with

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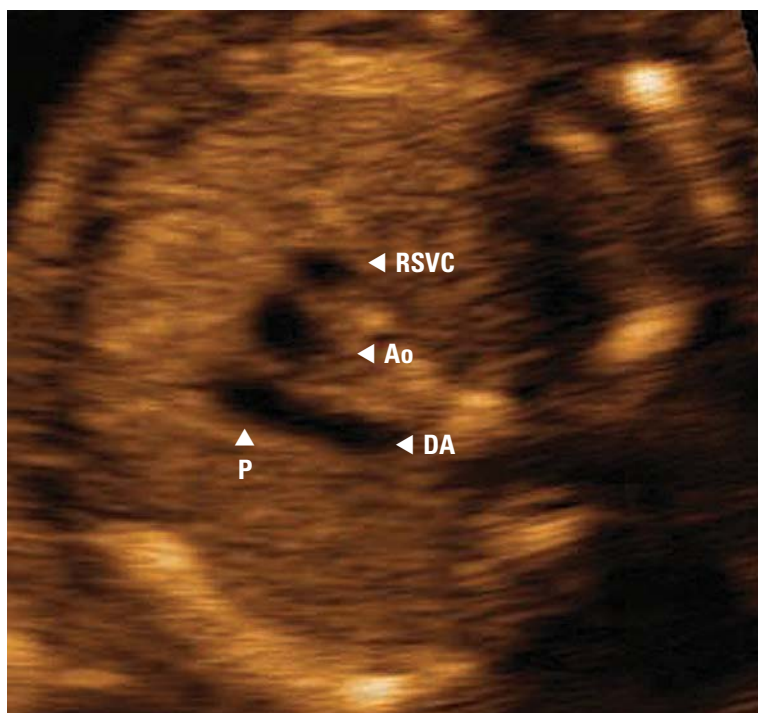


Figure 1: Three-vessel view with normal configuration of pulmonary trunk (P), ductus arteriosus (DA), aorta (Ao), and RSVC on the right of the aorta.

associated abnormalities such as dextrocardia, double outlet right ventricle, atrioventricular-septal defect, and associated polysplenia or asplenia. Other structural cardiac defects (not in the spectrum of heterotaxy) include coarctation of the aorta, ventricular septal defect, bicuspid aortic valves, tetralogy of Fallot and double aortic arch. A study by Galindo, *et al.* (2007) found that 9% of fetuses with CHD also had a persistent LSVC. In this group 41% were associated with heterotaxy, and 59% with other CHD. They concluded that a persistent LSVC was a strong marker for CHD.⁴

Persistent LSVC and arrhythmia

The persistent LSVC is associated with anatomical and architectural abnormalities of the pacemaker and conduction tissues. The atrioventricular node and sinus node both can show persistent fetal dispersion in the central fibrous body in subjects with persistent LSVC.¹³ Through its multiple anatomical and electrical communications with the atria the persistent LSVC may generate repetitive rapid discharges with shorter activation cycle length that promotes the initiation and maintenance of atrial fibrillation and sudden death.¹³⁻¹⁵

In more than 90% of cases, the LSVC drains into the right atrium via the coronary sinus and physiologically there are no clinical consequences.¹⁶ The clinical implications of a dilated coronary sinus include cardiac arrhythmia due to stretching of the atrioventricular node and bundle of His, and obstruction of the left atrioventricular flow because of partial occlusion of the mitral valve.¹⁷

In the remaining 10% of cases the LSVC drains directly into the left atrium, between the left atrial appendage and pulmonary veins. This anomaly is known as complete unroofing of the coronary sinus, or coronary sinus atrial septal defect, and results in a left to right shunt.^{18,19}

Persistent LSVC and other anomalies

In a postnatal series, Postema, *et al.* (2008) have also shown the

frequent association between a persistent LSVC and extracardiac anomalies.²⁰ The most common anomaly was oesophageal atresia. There was also a higher association with the VACTRL association (vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, limb anomalies) and CHARGE syndrome (coloboma, heart defects, atresia of the nasal choanae, retardation of growth, genital and/or urinary abnormalities, ear abnormalities and deafness).

Persistent LSVC in the asymptomatic adult

A RSVC is present in about 82% of the reported cases with or without a bridging innominate vein with persistent LSVC.²¹ In case of an absent RSVC, both right and left brachiocephalic veins drain into the persistent LSVC at the left internal side of the thorax.

Persistent LSVC has various practical implications when the left subclavian vein is used for access to the right side of the heart or pulmonary vasculature. Swan-Ganz catheter placement can be challenging. It can also complicate permanent pacemaker and implantable cardioverter defibrillator placement. Serious complications including angina, arrhythmia, cardiogenic shock, and even cardiac arrest have been reported when a guide wire or catheter is manipulated via persistent LSVC.²²

The clinical significance of a persistent LSVC has also been recognised by cardiothoracic surgeons. The presence of persistent LSVC is a relative contraindication to the administration of retrograde cardioplegia. Retrograde cardioplegia through persistent LSVC may lead to inadequate myocardial perfusion and therefore be ineffective.²³

Embryologic development and anatomy of Persistent Left SVC

Two pairs of cardinal veins constitute the main systemic venous drainage of the embryo. The anterior cardinal veins drain the cranial parts and the posterior cardinal veins drain the caudal parts of the embryo. Before entering the embryological heart,

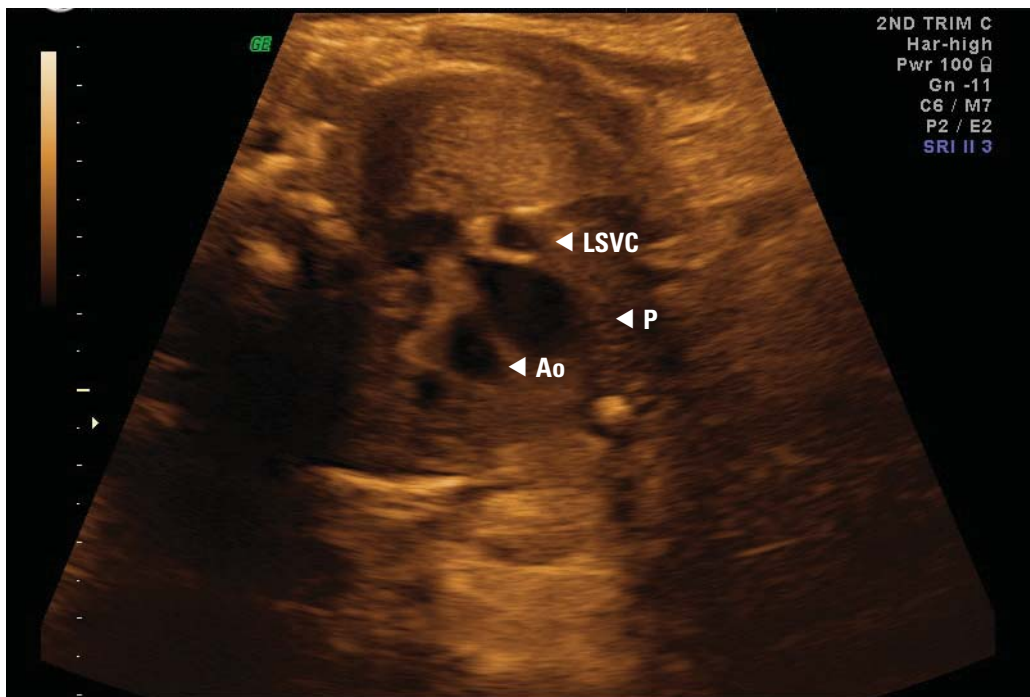


Figure 2: Three-vessel view with supernumerary vessel (LSVC) to the left of the pulmonary trunk (P) and aorta (Ao).

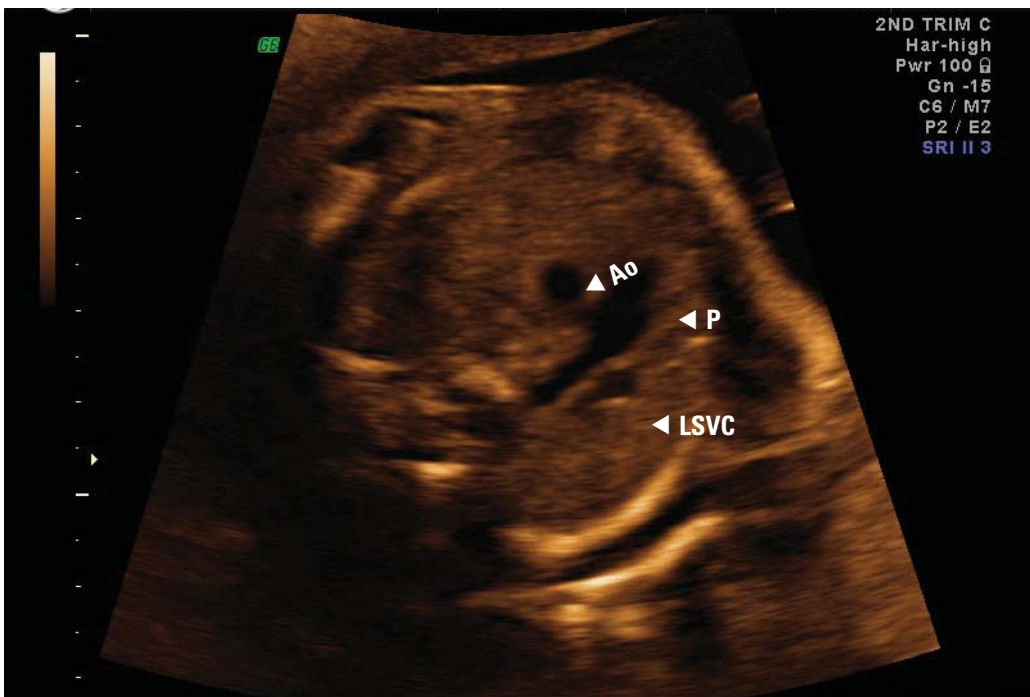


Figure 3: Three-vessel view with persistent LSVC on the left of the pulmonary trunk (P) with an absent RLSV.

both pairs of veins join to form right and left common cardinal veins. By the eighth week of gestation the innominate (or left brachiocephalic) vein connects the right and left anterior cardinal veins. The cephalic portion of superior cardinal veins form the internal jugular veins while the right anterior and common cardinal veins form the right SVC. The part of the left anterior cardinal vein caudal to the innominate vein normally regresses to become the ligament of Marshall.^{8,24,25} Failure of this normal regression can be attributed to the formation of a

persistent LSVC.²⁶ This persistent LSVC runs between the left atrial appendage and the left pulmonary veins, and almost always runs down the back of the left atrium, entering the right atrium through the orifice of an enlarged coronary sinus.⁹

Diagnosis in fetus and other considerations

The three vessel view was first described by Yoo and co-workers in 1997 and is now an integral part of the standard examination of the fetal heart.²⁸ The addition of the three vessel view at the

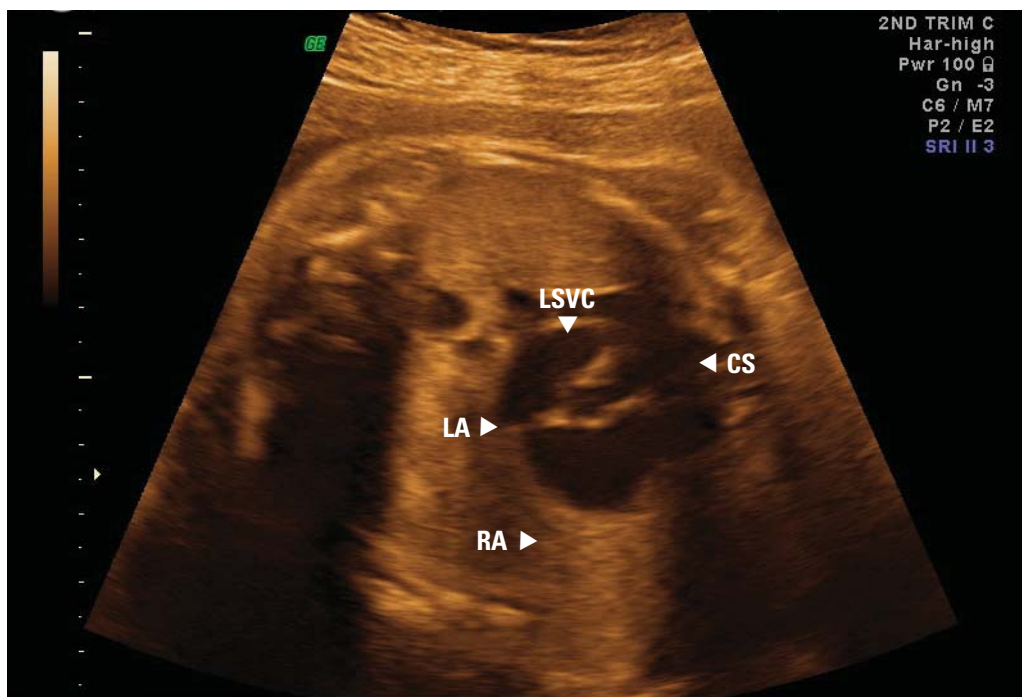


Figure 4: Oblique parasagittal view of the persistent LSVC and dilated coronary sinus (CS). Left atrium (LA), right atrium (RA).

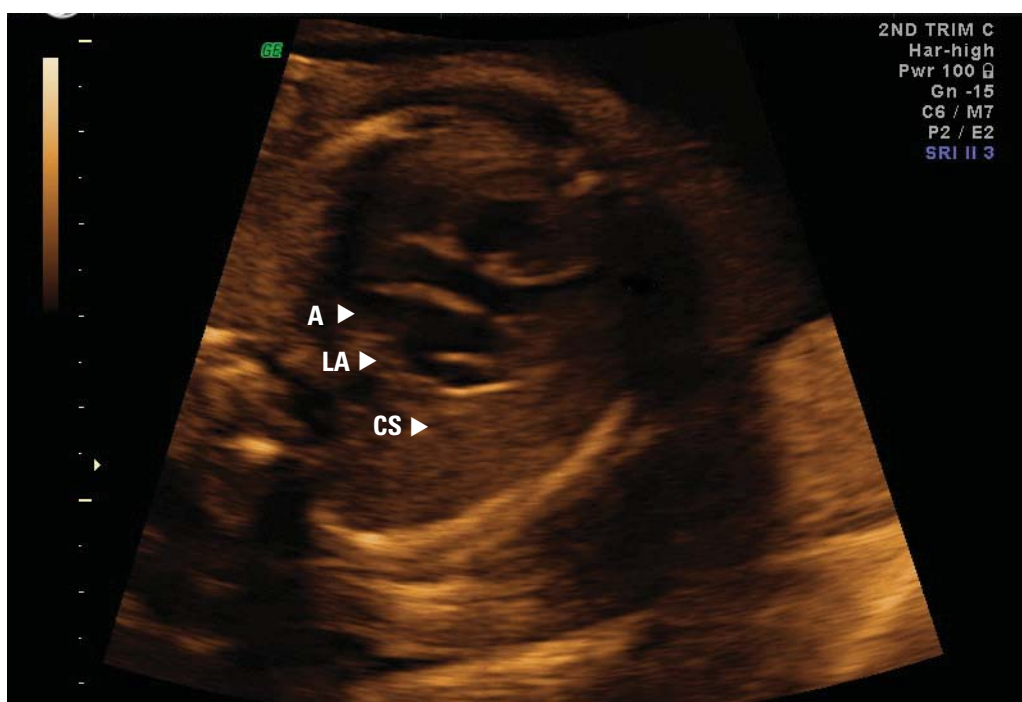


Figure 5: Dilated coronary sinus. Aortic root (A), left Atrium (LA), coronary sinus (CS).

upper mediastinum to fetal cardiac examination has facilitated easy and accurate prenatal diagnosis of persistent LSVC. In the normal fetus the three vessels that are usually seen going from left to right in the scan are arterial duct, aortic arch and RSVC (Figure 1). If a persistent LSVC is present, four instead of three vessels are seen, with a supernumerary vessel to the left of the pulmonary trunk and arterial duct (Figure 2). In case of an absent RSVC, there will again be three vessels but 'abnormally' arranged; from right to left, aortic arch and arterial duct and persistent LSVC (Figure 3).

A recent case series study by Barrea, *et al.* (2011) strongly recommended that the three axial parallel views (four-chamber view, three-vessel view and abdominal plane) should be part of the systemic ultrasound examination of the fetal heart.²⁷

The diagnosis of a persistent LSVC can also be characterised by the 'tobacco pipe' sign in a slightly oblique left parasagittal view, showing the LSVC draining into a dilated coronary sinus⁵ (Figure 4) This view is described in pediatric echocardiography and was reproduced in fetal echocardiography by Freund, *et al.* (2008).⁵ An isolated enlarged coronary sinus (Figure 5) is highly

Cardiac anomalies	Heterotaxy (left and right isomerism); coarctation of the aorta, ventricular septal defect, bicuspid aortic valves, tetralogy of Fallot and double aortic arch
Extra-cardiac anomalies	Oesophageal atresia
Genetic syndromes	VACTRL, CHARGE and Opitz G/BBB syndrome
Chromosomal abnormalities	Trisomy 21; Turner syndrome; microdeletion 22q11.2
Other	Increased nuchal translucency

Table 1: Summary – Associated findings in PLSVC.

suggestive of persistent LSVC, although this finding may have both false positive and false negative diagnoses.^{3,28–35}

With the aid of colour and spectral Doppler further diagnostic precisions can be obtained. For example, in cases with unroofed coronary sinus the left to right shunt can be easily identified.

It is accepted practice that an increased nuchal translucency (above the 95th centile) is an indication for fetal echocardiography, as the prevalence of major CHD for these fetuses is higher than in the general population.³³ A retrospective case series review by Galindo, *et al.* (2007) noted that up to 29% of the fetuses with persistent LSVC had an increased NT in the first trimester.⁴

The association of aneuploidy (particularly Trisomy 21, Turner syndrome and microdeletion 22q11.2) and persistent LSVC has been reported, with some authors advocating that fetal karyotype should be routinely offered whenever persistent LSVC is detected prenatally.^{30,32} Others contend that persistent LSVC itself should not constitute an indication for fetal karyotype, as the cardiac defects themselves are the true conditions associated with chromosomal disorders and not the persistent left SVC.⁴ Table 1 summarises all the associations with this finding.

Conclusion

The key to the prenatal diagnosis of a persistent left superior vena cava (LSVC) is the three vessel view. It is a finding with a high association with cardiac, extra-cardiac and genetic syndromes and for that reason it is an important diagnosis. The defect in isolation is however generally associated with a favourable prognosis (as in our cases) but it might be an important finding later in life when cardiac procedures are required.

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