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## Gradual diagnosis and clinical importance of prenatally detected persistent left superior vena cava with absent right superior vena cava – a case report and literature review

Oskar Sylwestrzak<sup>1,2</sup>, Maria Respondek-Liberska<sup>2,3</sup>

<sup>1</sup> Medical Faculty, Medical University of Lodz, Lodz, Poland

<sup>2</sup> Department for Prenatal Cardiology, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland

<sup>3</sup> Department for Congenital Malformations and Prevention, Faculty of Public Health, Medical University of Lodz, Poland

Correspondence: Oskar Sylwestrzak, Department of Prenatal Cardiology, Polish Mother's Memorial Hospital Research Institute in Łódź, 281/289 Rzgowska, 93-338 Lodz, Poland; tel. +48 42 271 15 56, e-mail: sylwestrzakoskarptryk@gmail.com

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### Keywords

fetal echocardiography, persistent left superior vena cava, ultrasound

### Abstract

Persistent left superior vena cava is a malformation of cardinal veins. We report a case of a secundigravida who had many fetal ultrasound examinations – first performed by an obstetrician (who described fetal mediastinum as “abnormal”), second by other obstetrician, who performed basic fetal echocardiographic examination and diagnosed persistent left superior vena cava. The woman was referred to a tertiary center for detailed fetal echocardiography. The diagnosis of persistent left superior vena cava with agenesis of the right superior vena cava was confirmed. The anomaly had no influence on fetal hemodynamic stability, fetal life, delivery and early postnatal period. After delivery, the neonate was under observation for further anomalies, aortic coarctation in particular. Prenatal and postnatal management was summarized. Literature review is presented.

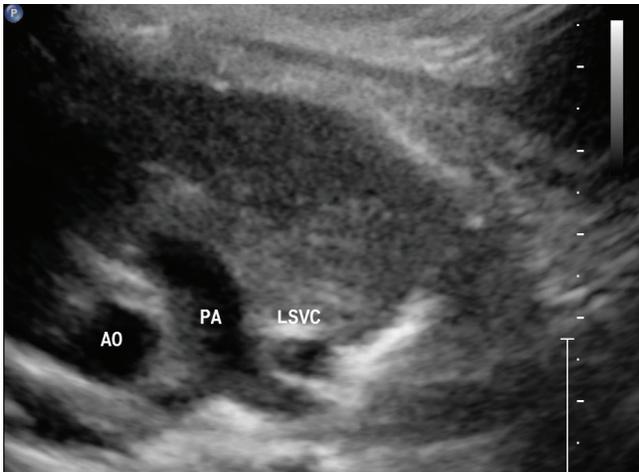
## Introduction

At week 4 of gestation, the blood of the upper half of a fetal body is drained by the bilateral symmetrically arranged anterior cardinal veins. By week 8 they connect by an oblique anastomosis (future left brachiocephalic or innominate vein) and the part of the left anterior cardinal vein regresses. Persistent left superior vena cava (PLSVC) seems to be a remnant of this regressed vessel<sup>(1)</sup>. Achiron *et al.* proposed a classification of fetal venous system anomalies, where PLSVC is categorized as isolated malformations of cardinal veins, and, among others, PLSVC is the most common variant of anomalous systemic venous return in adults<sup>(2,3)</sup>. The aim of our article was to present a case report

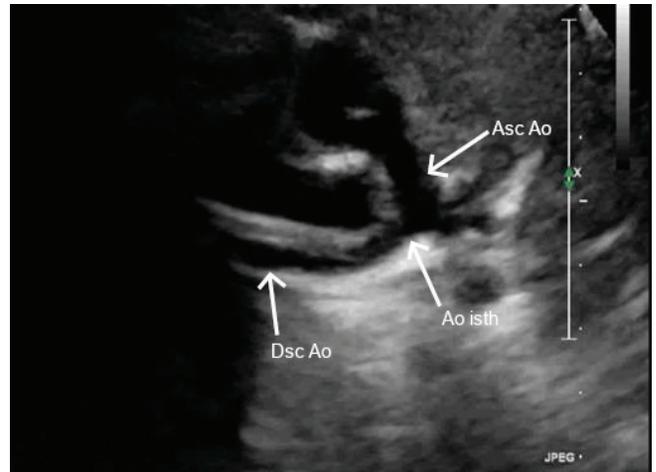
of a fetus with PLSVC and agenesis of the right superior vena cava (SVC), as well as to emphasize the diagnostic role of a prenatal cardiology center. We also wish to show the clinical significance of PLSVC based on a literature review.

## Case report

A 23-year-old secundigravida was referred to our tertiary center due to suspected PLSVC in her male fetus. There was no relevant family history, but the patient complained of flu-like symptoms, leukorrhoea and infections of urinary tract earlier during pregnancy. Many ultrasound examinations were performed during pregnancy.



**Fig. 1.** Fetal mediastinum- additional vessel, left to the main pulmonary artery. (AO – aorta, PA – main pulmonary artery, LSVC – left superior vena cava)



**Fig. 2.** Aortic arch view (AscAo – ascending aorta, Aoisth – aortic isthmus, DscAo – descending aorta)

### 22 + 2 weeks of gestation

During a routine ultrasound examination at 22 + 2 weeks of gestation, an obstetrician described amniotic fluid index (AFI), central nervous system, face, abdominal organs, skeleton and limbs as normal. He measured biometric parameters and estimated weight ( $\pm 645$  g). There was a 3-vessel umbilical cord. The axis and size of fetal heart, 4-chamber view, LVOT (left ventricular outflow tract) and RVOT (right ventricular outflow tract) were also described as normal. Nevertheless, the 3-vessel view of the mediastinum was defined as 'abnormal'.

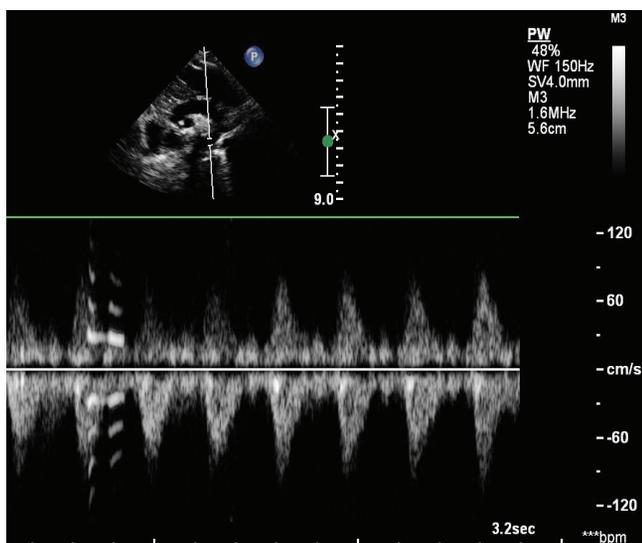
### 23 weeks of gestation

At 23 weeks of gestation, the obstetrician who performed basic fetal echocardiographical examination confirmed

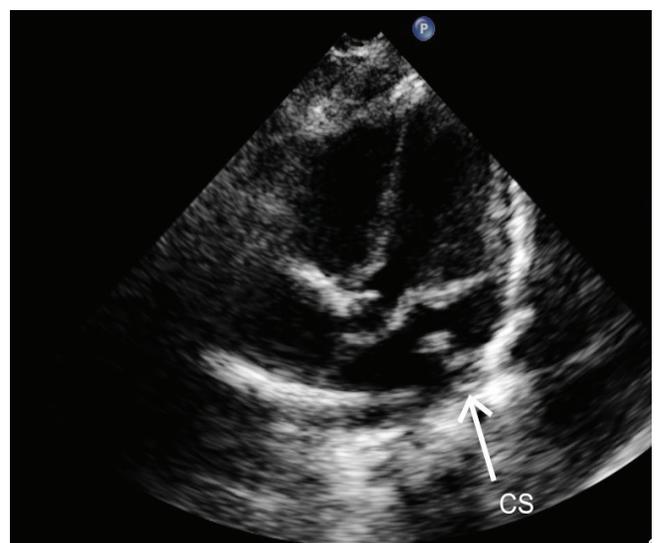
correct position size, axis, LVOT and RVOT of the fetal heart. Sinus rhythm was described. The sizes of both atria and ventricles were equal. However, he noticed a widened coronary sinus, an abnormal arrangement of the mediastinal vessels, with the absence of the right SVC and an additional vessel, left to the MPA (main pulmonary artery). He suspected PLSVC.

### 30 weeks of gestation

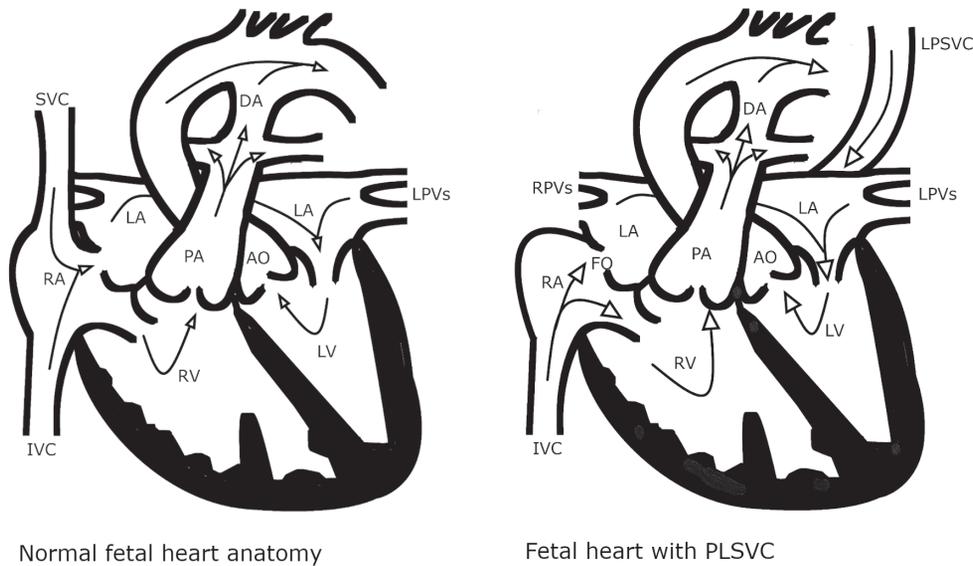
The patient was admitted to our department at 30 weeks of gestation and monitored even after delivery. Prenatal cardiologists thoroughly assessed many fetal heart parameters and evaluated the previously described abnormality (Fig. 1). Agenesis of the right SVC and PLSVC were confirmed. The aortic arch (Z-score and Vmax for aortic arch were within normal limits throughout pregnancy) and LVOT were



**Fig. 3.** Flow through aortic arch



**Fig. 4.** Left ventricular outflow tract. No disproportion between left and right heart (CS – coronary sinus)



**Fig. 5.** Sketches of normal fetal heart (left) and fetal heart with PLSVC and agenesis of the right SVC. Arrows – flow through fetal heart (RA – right atrium, LA – left atrium, RV – right ventricle, LV – left ventricle, PA – main pulmonary artery, AO – aorta, DA – ductus arteriosus, RPVs – right pulmonary veins, LPVs – left pulmonary veins, FO – foramen ovale, IVC – inferior vena cava, LPSVC – left persistent vena cava)

examined in detail (Fig. 2, Fig. 3, Fig. 4) and no abnormality of aorta was detected. The fetus was described as cardiovascularly efficient, so the anomaly was inconsequential from cardiological point of view. Other organs were also evaluated during pregnancy.

### Discussion

PLSVC (Fig. 5) was described for the first time in 1950 by Edwards and DuShane<sup>(4)</sup>. This is the most common variant of anomalous systemic venous return in adults<sup>(3)</sup>. PLSVC is observed in 0,21% of the general population and 4–8% of patients with congenital heart disease (CHD)<sup>(5,6)</sup>. There is no sex difference in the incidence of PLSVC<sup>(6)</sup>.

Since in most cases PLSVC drains into the right atrium via the coronary sinus (CS), even in 93%<sup>(7)</sup>, dilation of CS is usually the first sign detected during fetal echocardiography, but it can also indicate anomalous pulmonary venous drainage<sup>(3,8)</sup>. In the case of agenesis of the right SVC, CS may be markedly dilated as it receives the whole venous drainage from the upper body<sup>(9)</sup>. Moreover, dilated CS might be misdiagnosed as an ostium primum atrial septal defect or as mitral atresia<sup>(8,10)</sup>. The diagnosis must be confirmed in the three-vessel trachea view (3VT) showing the supernumerary vessel to the left of the pulmonary trunk and arterial duct<sup>(1)</sup>. Measuring the distance between the right SVC and the aorta at 3VT might be also used for PLSVC detection as this distance is increased in bilateral SVC<sup>(11)</sup>. However, this technique would be inadequate in our patient as the fetus was diagnosed with agenesis of the right SVC. PLSVC and agenesis of the right SVC is uncommon, with an incidence of 0.09–0.13%<sup>(9)</sup>; therefore, four vessels are most often seen in the mediastinum<sup>(4,12)</sup>. Of 222 fetuses with PLSVC, only one had agenesis of the right SVC<sup>(7)</sup>. Sometimes PLSVC with agenesis of the right SVC is termed ‘isolated PLSVC’ in the literature. For this article, the term ‘isolated PLSVC’ means

PLSVC without any other abnormal finding (no intracardiac and extracardiac anomaly).

The incidence of PLSVC is significantly higher among fetuses with chromosomal anomalies (7.8%) compared to those clinically normal (0.4%), and the odds ratio is 27.5. The highest incidence was observed in fetuses with trisomy 18 (12.1%). Among chromosomally abnormal PLSVC fetuses, PLSVC was an isolated defect only in 10% (2/20)<sup>(13)</sup>. Most aneuploid fetuses with PLSVC exhibit additional intra- or extracardiac defects<sup>(14)</sup>. There is low likelihood of chromosomal anomalies in fetuses with apparently isolated PLSVC<sup>(3)</sup>. Tertiary prenatal cardiology centers seem to be an adequate facility to confirm PLSVC before delivery due to their experience. Additional intra- and extracardiac

**Tab. 1.** Differences between fetal echocardiography and obstetrical ultrasound<sup>(8)</sup>

	Additional fetal echocardiography elements
Duration of examination	30–45 minutes
Separate elements of examination	Techniques: Color Doppler, Spectral Doppler, Power Doppler, Power angio, M-Mode, tissue M-Mode, 3D, 4D  Detailed examination of: atria, ventricles, foramen ovale and its flap, valves: mitral, tricuspid, aortic, pulmonary, pulmonary trunk, aortic arch, isthmus, descending aorta, pulmonary arteries, superior vena cava, inferior vena cava, intraventricular septum, ductus venosus, ductus arteriosus, umbilical arteries and vein, upper mediastinum, thymus  Parameters: AP (transverse heart diameter), shortening fraction, TEI index, Pulsatility index, Resistance index, TAPSE, MAPSE

anomalies were missed at the prenatal ultrasound only in 2.4% of fetuses with PLSVC<sup>(3)</sup>.

PLSVC is associated with cardiac defects in more than 80% of cases<sup>(8)</sup>. Increased risk of coarctation of aorta (CoA) was noticed in up to 21% of fetuses with PLSVC<sup>(3)</sup>. The ratio of the incidence of PLSVC in CHD patients to that in the normal population was very high (over hundredfold) in CoA and DORV group<sup>(6)</sup>. There is a high prevalence of ventricular septal defect, atrial septal defect, endocardial cushion defect, cor triatriatum and tetralogy of Fallot in patients with PLSVC and agenesis of the right SVC<sup>(15)</sup>. Additionally, in patients with pulmonary valve stenosis, where the incidence of PLSVC was the lowest, it was 7.0 times higher than that of the normal population<sup>(8)</sup>. It is noteworthy that the majority of extracardiac anomalies occurred in association with cardiac anomalies in some studies<sup>(13)</sup>. PLSVC is more often seen in association with esophageal atresia, diaphragmatic hernia, or hypoplastic thymus (the latter one in autopsy)<sup>(2,7)</sup>. At this point, we need to note that significant morbidity and mortality might arise from the associated anomalies and other heart defects rather than the PLSVC itself<sup>(2,16)</sup>. Autopsies of cases with PLSVC revealed associated anomalies in all these cases, with extra cardiac anomalies in 96%<sup>(7)</sup>.

After delivery, due to the relatively devious course, central venous catheterization, placement of pacemaker leads and pulmonary artery catheter may be difficult. That is why PLSVC should be reported in patient's medical history. We even recommend an entry in a special fetal health record book for better postnatal healthcare. Central venous line insertion into the right internal jugular vein in the case of PLSVC with agenesis of the right SVC in a child or an adult may be impossible. Arrhythmias and conduction abnormalities are common in PLSVC patients. Draining PLSVC to the left atrium may increase the risk of paradoxical embolism due to the right to left systemic shunt. Also, intravenous drugs may bypass the right heart and directly reach the systemic circulation. There is a need for draining PLSVC with right SVC agenesis by a separate venous cannula, when performing cardiac surgeries involving opening of right heart. In electrocardiographic record, left axis deviation of the P wave with shortened PR interval may be present. X-ray might show additional shadow of the PLSVC<sup>(15,17,18)</sup>.

Considering these facts, prenatal detection of PLSVC should prompt referral for detailed echocardiography<sup>(19)</sup>. In a tertiary center, an echocardiographic examination differs from basic ultrasound screening (Tab. 1). More extensive

examination is crucial for providing better healthcare, especially when additional intra- or extracardiac defects occur; for example fetal CoA is still a considerable challenge due to the very difficult prenatal diagnosis and fatal consequences if undetected<sup>(10)</sup>. Postnatal follow-up of prenatally detected PLSVC seems reasonable. Experienced prenatal cardiologists deal with congenital heart defects and abnormalities in their everyday practice. Multistage diagnosis enables proper healthcare, especially in a third grade hospital, where many specialized departments cooperate. A tertiary center of prenatal cardiology should not only detect fetal heart abnormalities, but also offer information about treatment options, prognosis, and counsel about the way and place of delivery, as well as about future procreative plans. Moreover, many experienced clinics may provide care for a neonate, an infant and a child if necessary. We recommend neonatal echocardiographic examination after birth and at the age of 3 and 12 months for possible CoA detection. Fortunately, in our case, the postnatal follow-up revealed asymptomatic course and no CoA. Chao *et al.* noted that more extensive newborn screening using echocardiography can allow for an early diagnosis of PLSVC<sup>(20)</sup>. Specialized centers of prenatal cardiology could allow the detection of PLSVC with high rate even earlier, before delivery. Rarely, but still PLSVC could be unmentioned in patients' medical records, which may have catastrophic consequences during invasive procedures<sup>(20,21)</sup>.

## Conclusions

PLSVC with agenesis of the right SVC may be an anomaly with no influence on fetal hemodynamic stability, fetal life, delivery and early postnatal period. Despite the fact that PLSVC does not affect fetal circulation in most cases, it is always an indication for the extension of echocardiographic, ultrasound and sometimes genetic diagnosis in fetuses. Reference centers of prenatal cardiology are adequate for further diagnosis, after detection of PLSVC by an obstetrician. Postnatal echocardiographic examination should be performed to exclude CoA. PLSVC should be recorded in patient's medical history.

## Conflict of interest

*Authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.*

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