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Prenatal diagnosis of persistent left superior vena cava

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ARTICLE INFORMATION

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Abbreviations
AVSD: atrioventricular septal defect
CHD: congenital heart disease
CS: coronary sinus
LV: left ventricle
PLSVC: persistent left superior vena cava
RSVC: right superior vena cava

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ABSTRACT

Introduction: The persistence of the left superior vena cava (PLSVC) is the most common anomaly of the thoracic venous system and may be associated with other cardiac malformations.
Objectives: To describe the echocardiographic characteristics of PLSVC and its relationship with risk factors and associated abnormalities.
Methods: A descriptive, longitudinal and retrospective study was carried out, in order to contribute to the study of the prenatal diagnosis of PLSVC through the analysis of different indicators. The sample consisted of 62 cases with a prenatal diagnosis of PLSVC, in fetuses between 22 and 35 weeks, examined in the Fetal Echocardiography Department of the Cardiocentro Pediátrico William Soler, during the period from January 2008 to May 2012.
Results: The PLSVC was associated with congenital heart disease in 33 of the 62 cases. Among these, the most frequent were conotruncal heart defects (38.46%).
Conclusions: The PLSVC is easily recognized by screening ultrasound and represents an important finding for the diagnosis of congenital heart disease; therefore, given its suspicion —due to its frequent association with congenital heart disease— it is suggested to offer the pregnant woman a detailed evaluation of the fetal heart.
Keywords: Superior vena cava, Left superior vena cava, Fetal echocardiography, Prenatal diagnosis

Diagnóstico prenatal de la vena cava superior izquierda persistente

RESUMEN

Introducción: La persistencia de la vena cava superior izquierda (VCSIP) es la anomalía más común del sistema venoso torácico y puede estar asociadas a otras malformaciones cardiacas.
Objetivos: Describir las características ecocardiográficas de la VCSIP y su relación con los factores de riesgo y anomalías asociadas.
Método: Se realizó un estudio descriptivo, longitudinal y retrospectivo, con el propósito de contribuir al estudio del diagnóstico prenatal de la VCSIP mediante el análisis de diferentes indicadores. La muestra estuvo conformada por 62 casos con diagnóstico prenatal de VCSIP, en fetos entre 22 y 35 semanas, examinados en el Servicio de Ecocardiografía Fetal del Cardiocentro Pediátrico William Soler, durante el período comprendido desde enero de 2008 hasta mayo de 2012.

Authors' contribution
CGG: Research conception and design, raw data collection-analysis and final report writing.
JAF and CMO: Research conception and design. Revision of final report.
WGL and YHM: Information research, raw data collection and analysis.
All authors read and unanimously approved the final manuscript.

Resultados: La VCSIP se presentó asociada a cardiopatías congénitas en 33 de los 62 casos. Dentro de estas, las más frecuentes fueron las anomalías troncoconales (38,46%).
Conclusiones: La VCSIP es de fácil reconocimiento mediante el ultrasonido de pesquisa y constituye un hallazgo importante para el diagnóstico de las cardiopatías congénitas; por lo cual, ante su sospecha —debido a su frecuente asociación con cardiopatías congénitas— se sugiere ofrecer a la gestante una evaluación detallada del corazón fetal.
Palabras clave: Vena cava superior, Vena cava superior izquierda, Ecocardiografía fetal, Diagnóstico prenatal

INTRODUCTION

At the dawn of prenatal cardiac diagnosis, most attention was paid to anomalies related to the heart and its great arteries. As ultrasonographic technology has advanced and operators have gained expertise, observation and analysis of venous vascular structures −both pericardial and those of the mediastinum and abdomen−, has taken giant steps forward. To this end, it is critical that research accuracy be based on knowledge of the development and variations of the normal venous system in humans, a subject that has been largely studied[1-3]. Isolated venous system anomalies are rare, but their incidence is high when associated with congenital heart disease (CHD). For example, persistent left superior vena cava (PLSVC) is the most common anomaly of the thoracic venous system, occurring in 0.3-0.5% of the general population and up to 10% in patients with congenital heart disease CHD[4]. Most of the time, the PLSVC connects to and drains into the coronary sinus (CS), a small tubular structure located above the posterior atrioventricular junction, which can be visualized in different echocardiographic views[5-7].

We decided to carry out this research as it is frequently associated with other CHD and because there is no previous research in our country. Our objective was to describe the echocardiographic features of PLSVC and its relationship with risk factors and associated anomalies.

METHOD

A descriptive, longitudinal and retrospective study was conducted aiming to contribute to the study of prenatal PLSVC diagnosis through the analysis of different indicators. The sample consisted of 62 cases with prenatal diagnosis of PLSVC, in fetuses between 22-35 weeks, examined in the Fetal Echocardiography Department of the *Cardiocentro Pediátrico William Soler*, from January 1, 2008 to May 31, 2012. Data were taken from the records designed for this purpose in our Department, obtaining the following variables: gestational age, risk factors, description of echocardiographic views and associated anomalies. Transverse, longitudinal and angled scanning planes with their corresponding views were used in the echocardiographic study allowing assessment of the different structures. Standard criteria described in the international literature for this type of examination were considered[5-9].

Presence of an echolucent image within the left atrium (dilated CS) was taken as a sign of possible PLSVC and its diagnosis was based on the confirmation of the presence of PLSVC to the left of the main pulmonary artery, in the three-vessel view; to the left of the aorta, in the transverse aortic arch view; and in demonstrating this benign vascular anomaly entering the dilated CS, in the short-axis view after tilting the probe.

The diagnosis of PLSVC was confirmed by postnatal ultrasonography or pathological study when the parents opted for termination of pregnancy after cardiogenetic counseling.

Aloka 5500 and HDI 5000, with 3 and 5 MHz transabdominal probes, with the use of B Mode and color Doppler were employed for examination of the fetal heart; ALOKA 5500 system and 3.5 to 7 MHz sector probes, transthoracic, in the supine decubitus position was used for newborns. B-mode, pulsed, continuous and color Doppler were utilized.

RESULTS

A total of 62 fetuses with PLSVC were diagnosed within the research period. Suspected CHD as pri-

mary risk factor for referral was present in 57/62 (91.93%) cases. In 1/62 cases the risk factor was related to increased nuchal translucency. Other causes were: a mother with Noonan syndrome and a case of gestational diabetes. No risk factors were found in the other two fetuses. Most frequent possible diagnoses in the cases referred for suspected CHD were the presence of PLSVC in (26,31%), atrioventricular septal defects (AVSD) (21.05%) and dilation of right chambers (19.28%) (**Table 1**). Imaging planes —transverse (four-chamber/three-vessel/transverse aortic arch views), angled (left ventricular [LV] long-axis) and longitudinal (short-axis of the aorta with probe tilting)— showed disorders in all of the cases studied. Absent right superior vena cava (RSVC) with persistent LSVC was evidenced in 4/62 fetuses. Presence of PLSVC was associated with congenital anomalies in almost half of cases (46.77%); of these, heart disease was the most significant group (89.65%) (**Table 2**) while conotruncal heart defects (38.46%) were the most frequent.

Fourty nine out of 62 couples opted to continue the pregnancy and 13 (12 with CHD and 1 with diaphragmatic hernia) opted for its termination after genetic counseling.

Out of the 49 cases that decided to continue the pregnancy, 2 were fetal deaths. In the 13 terminated fetuses (4 tetralogy of Fallot, 4 left heart hypoplasia, 1 truncus arteriosus communis, 1 AVSD, 1 subaortic stenosis, 1 diaphragmatic hernia, 1 double-outlet right ventricle) the presence of PLSVC was confirmed by pathological anatomy and 12 of them presented the above-mentioned anomalies. One of the cases with prenatal AVSD with PLSVC diagnosis was false-positive, as a wide ventricular septal defect with PLSVC was eventually confirmed, supporting the above-mentioned differential diagnosis of this disease.

The fetus with subaortic stenosis also showed ventricular septal defect. The fetus with left heart hypoplasia associated with other anomalies (holoprosencephaly, microcephaly and polydactyly) underwent fetal karyotyping with a diagnosis of chromosomal aberrations (trisomy 13). All 47 newborns

Table 1. Clinical diagnosis in cases referred with suspected congenital heart disease.

Clinical diagnosis	Total of cases	%
Presence of PLSVC	15	26,31
Atrioventricular septal defect	12	21,05
Right ventricular dilation	11	19,28
Tetralogy of Fallot	5	8,77
Left heart hypoplasia	5	8,77
Abnormal 3-vessel view	3	5,26
Aortic overriding	2	3,50
Ventricular septal defect	2	3,50
Truncus arteriosus communis	1	1,75
Total anomalous pulmonary venous connection to the coronary sinus	1	1,75
Total	57	100

Source: Fetal Echocardiography Department Records of the *Cardiocentro Pediátrico William Soler*.

Table 2. Congenital heart disease associated with persistent left superior vena cava.

Type of congenital heart disease	Total of cases	%
Double-outlet right ventricle	4	15,38
Tetralogy of Fallot	4	15,38
Left heart hypoplasia	4	15,38
Coarctation of the aorta	3	11,53
Ventricular septal defect	3	11,53
Aortic stenosis	2	7,69
Atrial septal defect	1	3,84
Total anomalous pulmonary venous connection	1	3,84
Type B interrupted aortic arch	1	3,84
Right isomerism	1	3,84
Atrioventricular septal defect	1	3,84
Truncus arteriosus communis	1	3,84
Total	26	100

Source: Fetal Echocardiography Department Records of the *Cardiocentro Pediátrico William Soler*.

were studied echocardiographically and the diagnosis of PLSVC –associated in 5 cases with other disorders– was confirmed in all of them. One case exhibited phenotypic anomalies and was confirmed with Williams syndrome at 6 months of age; hence, he was followed-up and supravalvular aortic stenosis was found at 1 year of age. Another patient was diagnosed with total anomalous pulmonary venous connection to the right atrium, a difficult-to-diagnose CHD in the prenatal stage. Two of them developed septal defects (ventricular/atrial septal defect). The newborn from a mother with Noonam syndrome also showed phenotypic compatibility with this disease

DISCUSSION

In the embryo, the cardinal vein system is responsible for the superior vena cava. As it grows, the left anterior cardinal vein will shunt its blood towards the right cardinal vein, through the innominate vein. This implies an increasingly important development of the future RSVC, at the expense of the left anterior cardinal vein. The presence of a left superior vena cava (LSVC) can be attributed to the persistence of the proximal part of the left anterior cardinal vein. If flow through the innominate vein is reversed, the right cephalic region drains into the left anterior cardinal vein resulting in an atretic or absent RSVC. This PLSVC drains either into the coronary sinus, right atrium or to both atria via an unroofed coronary sinus[10-12].

Pregnant women are frequently referred to the cardiologist for evaluation as risk factors for cardiac abnormalities are identified on prenatal ultrasound. In our case, suspected CHD was the primary risk factor for referral. We believe this is due to two main reasons: the operational structure of the national health system and high quality training of sonographers in recent years. Cuba runs a National –Prenatal Diagnosis of Congenital Defects and Genetic Diseases– Program; pregnant women between 18 and 24 weeks are offered dating ultrasound examination: echocardiographic four-chamber view, three-vessel view, left ventricular long-axis and short-axis of the aorta views are evaluated, which is highly convenient when identifying signs of such anomalies. Therefore, targeted screening during routine fetal ultrasonography along with systematic observation of the fetal heart would be the natural way to diagnose CHD. The low number of pregnant women without risk factors attending our hospital (only two) owes to the fact that it is a national reference center; therefore, all pregnant women received are previously evaluated at the different healthcare levels. The most frequent diagnoses in the cases referred for suspected CHD were: presence of PLSVC (26.31%), AVSD (21.05%) and right ventricular dilation (19.28%) (**Table 2**).

Professional training of ultrasonographers in a CHD diagnostic center and design of didactic materials to enhance continuous updating are key aspects that have led to increased rates of prenatal diagnosis of heart disease; thus giving the parents the chance of detailed cardiogenetic counseling.

Dilated CS due to PLSVC may be regarded as right ventricular disproportion or VSD (partial variety) proving a false positive diagnosis, as occurred in a group of patients (23/57). This is because the CS lies along the posterior wall of the left atrioventricular groove, which, being dilated, distorts the posterior wall of the left atrium, displacing the septum primum and foramen ovale; resulting in a modified four-chamber view where the ultrasound beam cuts transversely across the dilated CS mimicking absence of the septum primum.

The higher number of cases with diagnosis of PLSVC referred to our service in the years 2010-2012 (5, 6 and 4 cases, respectively) compared to those received in 2008-2009 (no cases) is relevant. This is due to the development of skills for the diagnosis of this benign venous anomaly as part of ultrasonographers training, who perform screening and diagnosis at different care levels, based on its value as a predictive marker for CHD.

Among the anatomical anomalies with predictive value detected by ultrasonography, there is a number of lesions that –although may be normal variants in the general population– have been found to be associated with cardiac structural defects[13]; These include, in addition to PLSVC, increased nuchal translucency between 11-13 weeks of gestation, interruption of the inferior vena cava with azygos continuation, and cardiac chambers disproportion.

It is worth remembering that the technique of fetal echocardiography depends on professional expertise. The more experienced the professional, the greater the degree of diagnostic accuracy.

In all cases, disorders were seen in different imaging planes: transverse (four-chamber/three-vessel/transverse aortic arch views), angled (left ventricular [LV] long-axis) and longitudinal (short-axis of

the aorta with probe tilting).

Four-chamber and left ventricular long-axis views showed dilated CS as an element of suspicion in all the fetuses studied (**Figure 1**), while in the remaining echocardiographic views the condition itself was evident. The other views described in fetal echocardiography showed no changes suggestive of this anomaly.

The CS, a venous structure where the coronary veins drain, is observable in the fetus. An angled movement of the probe allows for assessment of the posterior wall of the left atrium revealing a low-echogenicity tubular image of 3 mm maximum diameter, running from the left atrium to the interatrial septum[5-8]. In cases of PLSVC draining into CS, there was an increase in the size of the latter structure, which was evident in all fetuses.

Although the above form of presentation is the most common, PLSVC may sometimes drain into the left atrium (as published by Raghib in 1965[14]), associated with an atrial septal defect and complete unroofing of the CS, a rare anomaly, but one to be considered.

Other forms described in the literature are those PLSVC draining to the roof of the left atrium and left superior or inferior pulmonary vein connection to the PLSVC itself, completely absent among the cases analyzed[10].

Total anomalous pulmonary venous connection to coronary sinus allows dilation of this structure, an aspect to be taken into account for accurate diagnosis[15].

Several authors point out the importance of transverse three-vessel view in the diagnosis of PLSVC, as it is easily identified as a fourth vessel to the left of the pulmonary artery[5-9]. This work coincides with that described by other researchers (**Figure 2A**).

Slight movements of the probe, starting from the aforementioned view, enabled us to obtain a more anterior plane of the fetus and visualize the PLSVC, but on the left side of the aortic artery (trans-verse aortic arch view) (**Figure 2B**). This view also made it possible to specify the position of the aortic arch (left) and determine the size of the thymus (normal), both of which were found in all fetuses.

One of the differential diagnoses to consider is the presence of a levoatrial cardinal vein[16]. When atrial pressures are elevated, during the development of left ventricular outflow tract obstruction in early gestation, the patency of the levoatrial cardinal vein, a vessel draining into the innominate vein or the right superior vena cava, is favored. This vein

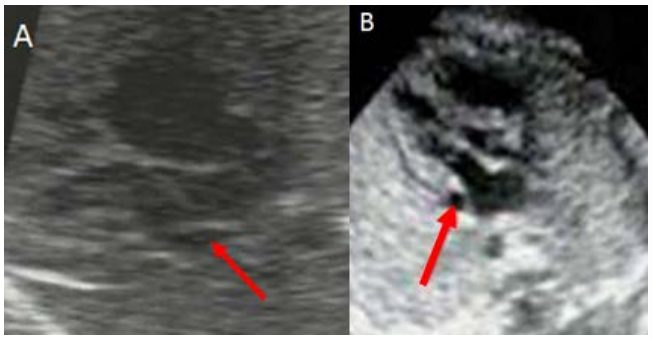

Figure 1. Four-chamber echocardiographic view (**A**) left ventricular long-axis view (**B**). The arrow highlights the dilated coronary sinus in both cases.

(levoatrial cardinal vein) is an embryological connection between the capillary plexus of the embryological foregut (origin of the pulmonary veins) and the cardinal venous system[17,18]. It differs from the normal course of the PLSVC by running behind the left pulmonary vein and in front of the left bronchus, which may cause compression or obstruction of the structures it passes through. Left atrial outflow obstruction occurs when there is mitral stenosis or atresia, a restrictive atrial defect, and an intact atrial septum with or without left heart hypoplasia. When this is diagnosed, retrograde flow into the innominate vein or right superior vena cava can be demonstrated[16-18].

Researchers point out that the aortic origin of one of the branches of the pulmonary artery is another malformation that must be ruled out. In the case studied, malalignment of the three-vessel view was appreciable, in addition to finding, to the left of the pulmonary artery, a vessel that appeared to correspond to the left branch originating from the ascending aorta. The presence of an anteriorly and slightly to the right descending aorta led to consider the possibility of a right aortic arch. In the transverse aortic arch view, from the origin of the vessel (aortic artery) a two-branch bifurcation, one downward and to the right (aortic arch) and the other to the left, was observable, in correspondence with the left branch of the pulmonary artery, directed toward the left lung[19].

Another differential diagnosis to bear in mind is congenital pulmonary airway malformation, where cysts in the left lung may actually mimic PLSVC. The slightly angled longitudinal view showed the PLSVC entering the CS as an intraluminal circular structure, which reaches the lateral wall of the left atrium (**Figure 2C**).

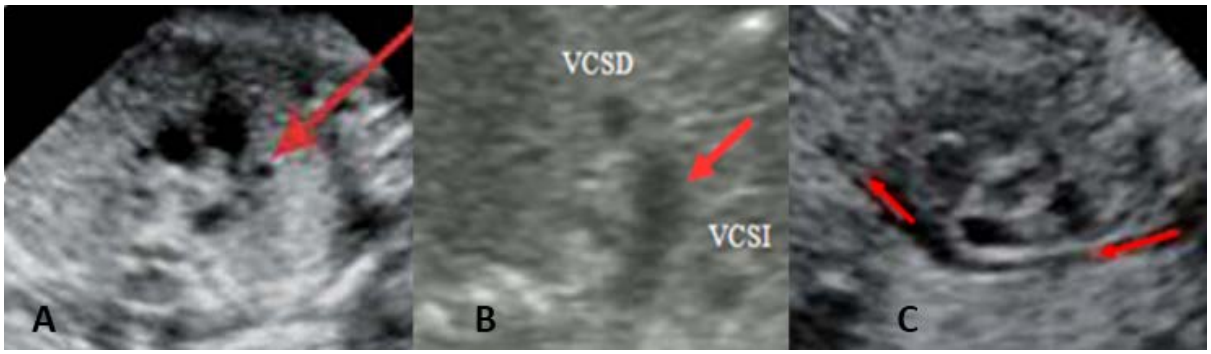

Figure 2. A. Echocardiographic view of the three vessels. A fourth vessel corresponding to the persistent left superior vena cava is indicated (arrow). **B.** Transverse aortic arch view (arrow). **C.** Echocardiographic longitudinal short-axis view of the aorta, with probe tilting, showing the arch-shaped entry of the PLSVC into the coronary sinus.
VCSD (acronym in Spanish): right superior vena cava. VCSI (acronym in Spanish): left superior vena cava.

Persistent left superior vena cava with absent RSVC was found in 4 of our cases. The combination of PLSVC with absent RSVC, first described in 1862, is extremely rare[20] and is therefore incidentally found in some patients during pacemaker implantation, cardiac catheterization, surgery or autopsy[21-23].

The incidence of PLSVC and absence of RSVC in fetuses is unknown a study by Lenox and collaborators in 1980 —cited by Pasquini *et al.*[22] and Bernal *et al.*[23]—, during a series of autopsies in children, found an incidence of 0.05%, with the peculiarity that all cases had associated CHD. Other authors (Phoon and Neill, 1994; according to Bernal *et al.*[23]) described —in 487 patients with right atrial isomerism— persistence of both caval veins in 46% of cases, while in 11% they found PLSVC with absent RSVC[22,23].

In the embryo, the cardinal vein system gives rise to the superior vena cava; as the embryo grows, the left anterior cardinal vein shunts its blood into the right anterior cardinal vein through the innominate vein, and causes an increasingly important development of the future RSVC to the detriment of the left anterior cardinal vein. The presence of PLSVC can be attributed to the persistence of the proximal part of the left anterior cardinal vein. If flow through the innominate vein is reversed, the right cephalic region drains into the left anterior cardinal vein resulting in an atretic or absent RSVC[10-12,23].

Three-vessel transverse and longitudinal vena cava views allow imaging of the RSVC in the fetus. When the RSVC is absent only three vessels will be visible; the first, to the left, smaller in diameter and located anteriorly, corresponds to the PLSVC, followed by the pulmonary and aortic arteries (**Figure 3A**). A more anterior echocardiographic plane shows the innominate vein entering the left superior vena cava, which collects blood from the right side of the neck and head in these cases (**Figure 3 B and C**). Longitudinal echocardiographic view of the caval veins revealed absent RSVC.

A PLSVC draining into the coronary sinus is an asymptomatic condition of no hemodynamic significance. However, its association with CHD, will be the main determinant of outcome.

The presence of PLSVC was associated with congenital anomalies in almost half of the cases; of these, heart disease was the most representative group. Pasquini *et al.*[22] reported a diagnosis of heart disease in 12 of 16 fetuses analyzed, with coarctation of the aorta being the most frequently observed. A study by Berg *et al.*[24] in two tertiary centers in Germany showed that of 82 fetuses with PLSVC, 83% had CHD (47% corresponded to positional [situs] anomalies) and conotruncal heart defects were the most frequent.

In 2007, Galindo *et al.*[2] reported PLSVC in 48% of fetuses with isolated heart disease. The most frequently observed anomalies in this series were LV outflow tract obstructions and conotruncal heart disease, results similar to ours.

Although no arrhythmias were found in the cases studied, it is important to highlight that the embryological development of the sinus/atrioventricular node, His bundle and conduction system may be highly influenced by failure of regression of cardinal veins, especially the left one, since these structures are located at the junction of both cardinal veins with the venous sinus.

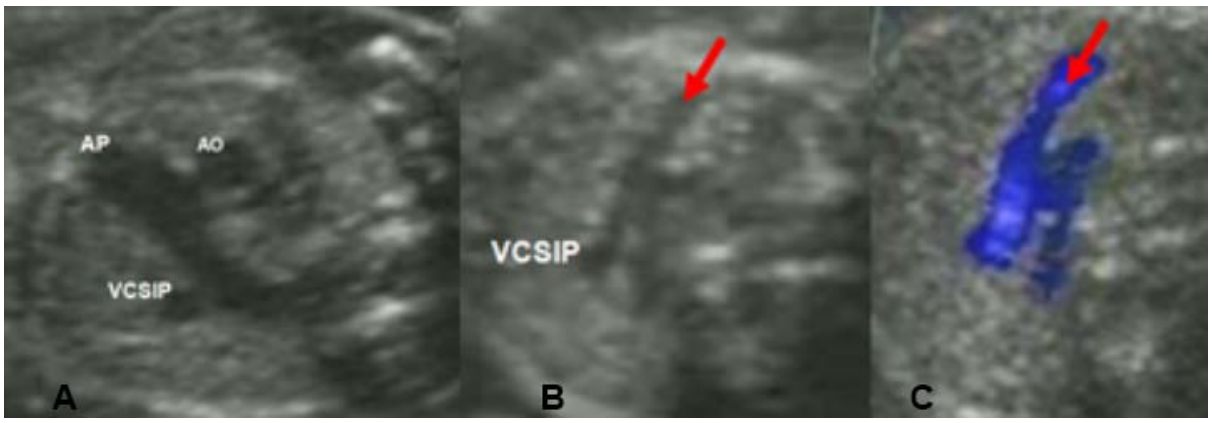

Figure 3. A. Three-vessel echocardiographic view. From left to right: left superior vena cava (VCSIP), pulmonary (AP) and aorta (AO) arteries. **B.** Transverse echocardiographic view (anterior) showing the innominate vein (arrow) entering the left superior vena cava (VCSIP). **C.** Color overlay on the two-dimensional image reveals the color flow path (blue) from right to left, characteristic of cases with absent right superior vena cava.

Persistent left superior vena cava with absent RSVC have been reported to modify the location and histological organization of the sinus node and the atrioventricular junction, resulting in: poor sinus node formation, fetal dispersion of the atrioventricular node and His bundle within the central fibrous body, small diameter of the His bundle, and poor blood supply to both nodes.

The association between PLSVC and chromosomal aberrations has been frequently described and some authors consider it necessary to carry out routine fetal karyotyping whenever this marker is detected[4,24-27]. However, most fetal aneuploidies are associated with cardiac, extracardiac defects, or both[4]. In the study by Galindo *et al.*[2], all chromosomal aberrations were diagnosed in fetuses with CHD, which suggested that cardiac defects are conditions truly associated with chromosomal aberrations, but not with PLSVC; therefore, we agree that this marker alone cannot be an indication for fetal karyotyping.

Persistent left superior vena cava was diagnosed in each of the cases assessed, which justifies the possibility of in utero diagnosis by means of the above-mentioned echocardiographic views. These views are standard for screening for congenital defects at primary care level and have proven useful since, although infrequent, PLSVC is a valuable marker that allows us to suspect the presence of associated anomalies, mainly CHD.

Fetal echocardiography, as a key tool for in utero cardiological diagnosis, has become essential for fetal assessment both for the obstetric sonographer, who has the first contact with the fetus, and for the specialist, who will confirm or exclude the presence of heart disease. Now, a good number of serious heart diseases requiring emergency clinical-surgical care after birth can be diagnosed from in utero life, which allows the medical team for planning immediate postpartum management in advance. The introduction of cardiac therapeutic measures targeting the fetus is becoming more and more widespread throughout the world.

CONCLUSIONS AND RECOMMENDATIONS

This study clearly indicates that persistent left superior vena cava is easily recognized by screening ultrasound and that it is a key finding in the diagnosis of congenital heart disease. Therefore, if suspected or identified, given its frequent association with congenital heart disease, a detailed assessment of the fetal heart should be offered to the pregnant woman.

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