Original paper

Med Ultrason 2016, Vol. 18, no. 2, 214-217
DOI: 10.11152/mu.2013.2066.182.fet

Ultrasonographic features of the persistence of superior left vena cava and pathological cardiac associations in fetus. Cases serie.

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Abstract

The persistence of superior left vena cava (PLSVC) is a pathological condition in fetus with risk of association with abnormalities like fetuses heterotaxy, cardiac abnormalities – atrioventricular septum defect, and conotruncal anomalies. In this paper we report 23 cases of fetuse with PLSVCs, reviewing their diagnosis, co-morbidities, and evolution in the newborns.

Keywords: fetal, persistence of superior left vena cava, echocardiography

Introduction

Persistent left superior vena cava (PLSVC) is the proximal segment of anterior left cardinal vein, which results from the lack of atrophy that normally follows the formation of left brachiocephalic vein [1]. PLSVC is the most frequent encountered anomaly of the thoracic systemic venous return (0.3%- 0.5% in the general population) [2,3]. This congenital anomaly can be associated with other cardiac or extra-cardiac anomalies. PLSVC passes between the left atrium appendix and left pulmonary veins, almost always gets at the back of the left atrium and enters the right atrium through the foramen of an enlarged coronary sinus [4]. In 65% of the cases the innominate vein or left brachiocephalic vein is absent or with a decreased diameter [5].

Received 08.01.2016 Accepted 19.02.2016
Med Ultrason
2016, Vol. 18, No 2, 214-217
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Cases analysis

Between the 1st of February 2007 and the 10th of November 2015, in a tertiary center (2 medical units: the Clinic of Obstetrics-Gynecology I and the Clinic of Pediatrics Cardiology from Tîrgu Mureș, Romania), 259 fetuses were diagnosed with cardiac anomalies (236 with structural anomalies and 23 with pathological arrhythmias). In this group of fetuses we diagnosed 23 cases of PLSVC, cases that were grouped according to the type of cardiac malformations.

The 9 groups are described in table I. The representative figures for the PLSVC associations are illustrated in figures 1-8. Out of the total number of cases, in 5 cases (2 with medical abortions and 3 newborns who died soon after birth) we do not have postpartum or postabortion confirmation of the diagnosis.

Discussions

During the fetal cardiac screening suggestive signs for a venous anomaly can appear: dilated coronary sinus or anechoic cyst on the wall of the left atrium in the same place with the entrance of the superior left vena cava in the coronary sinus. The section "three vessels" is the most important for the identification of a fourth supernumerary vessel on the left from the ductal arch or three vessels

with abnormal arrangement in the absence of the superior right vena cava [6-8]. For the diagnosis of the PLSVC is useful to demonstrate the absence of the brachiocephalic vein [1] and a "putter" appearance of the supernumerary vessel with trajectory behind the heart in connection with the coronary sinus. The Color Doppler examinaton is useful for the follow-up of the vessels confluence.

The PLSVC is an anomaly of venous return which appears in 0.3% of adults without cardiac abnormalities and in 4% of those with cardiac malformations [9]. Galindo et al [10] found a frequency of 0.2% in fetuses with normal heart and 9% in fetuses with cardiac abnormalities. Nsah et al [11] found an incidence of 9% on 1208 autopsied specimens of fetuses with cardiac abnormalities. The incidence of this venous anomaly in the general population may be even higher, up to 12% due to the fact that it usually is asymptomatic [3]. In our series of fetal cardiac malformations or fetal arrhythmias, the incidence of the

Table I. The nine groups of associate pathology of the 23 cases with persistent left superior vena cava.

Group	Number of cases	Family history	Age of diagnosis (weeks of gestation)	Outcome, neonatal period
Left ventricular outflow tract anomalies	5	unremarkable	22- 35	1 medical abortion 2 AoCo including one with surgical correction 2 asymptomatic children
Double outlet right ventricle	3	1 patient with spontaneus abortion in the first trimester	21-31	2 newborns underwent surgery – sistemicopulmonary shunt in the first week of life 1 newborn is planed for surgical intervention
Tetralogy of Fallot	2	unremarkable	29-37	1 newborn with surgical intervention planed 1 newborn with extreme form of Fallot died shortly after birth
Complex fetal cardiac malformations	3	unremarkable	21-34	1 medical abortion 2 newborns died in the first day of life
Common arterial trunk	1	unremarkable	36	Cheilognatopalatoschisis, 18 trisomy, the newborn died at two months of age
Premature constriction of the arterial duct	1	unremarkable	37	Essential pulmonary hypertension, the newborn improves rapidely
Cardiac arrythmias	1	brothers and parents with ASD and arrythmias	20	Newborn treated 2 years with antiarrythmic drugs.
PLSVC with absence of the right superior vena cava	4	unremarkable	22-36	Healthy children between 2 to 12 months
Presence of PLSVC without associations	3	unremarkable	19-25	Healthy children at the age of 6 to 12 months

AoCo – aortic coarctation; ASD – atrial septal defect; PLSVC – persistent left superior vena cava

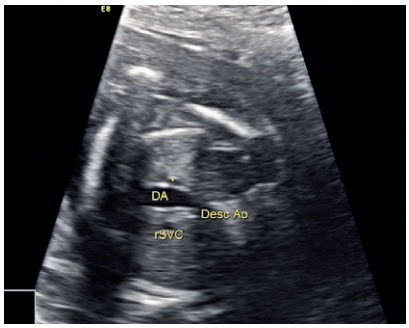


Fig 1. The section "three-vessels" identifies the fourth vessels, suspicion of AoCo. SVC – superior vena cava, Ao-aorta, asterix – persistent left superior vena cava.

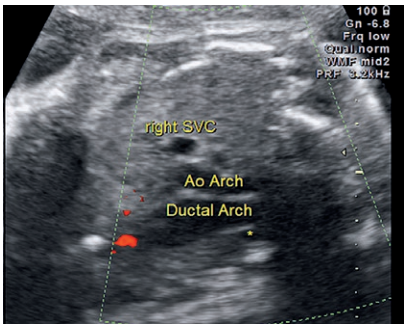


Fig 2. Hypoplasic left heart syndrome, with dilation of coronary sinus (CS) given by the presence of evident hypoplasia of left ventricle (LV) versus right ventricle (RV).

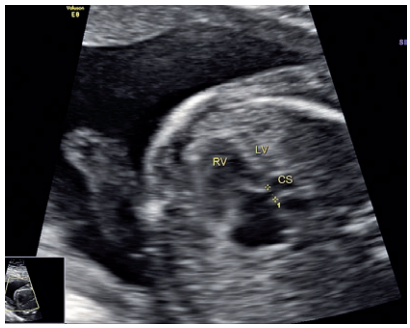


Fig 3. Hypoplasic left heart syndrome, the "three vessels" section in the absence of the aortic arch. rSVC – right superior vena cava, DA – ductus arteriosus, Desc Ao – descending aorta, asterix – persistent left superior vena cava

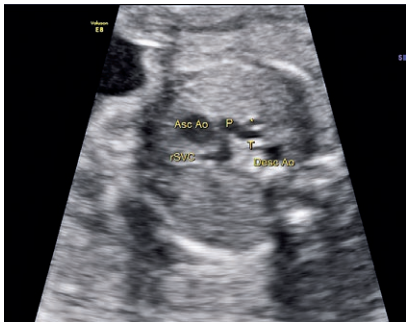


Fig 4. Double outlet right ventricle with pulmonary stenosis, "three vessels" section. T – trachea, Asc Ao – ascending aorta, rSVC –right superior vena cava, P-pulmonary trunk, asterix – persistent left superior vena cava

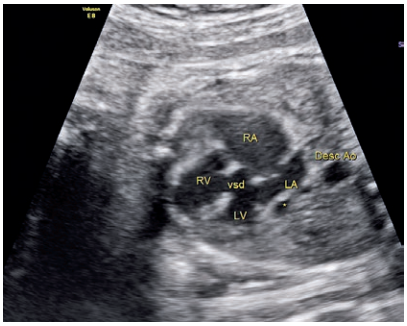


Fig 5. Tetralogy of Fallot, four chambers view. RV – right ventricle, LV – left ventricle, LA – left atrium, RA – right atrium, VSD – ventricular septal defect, Desc Ao-descending aorta, asterix – the entrance of the persistent left superior vena cava in the coronary sinus, on the left atrium

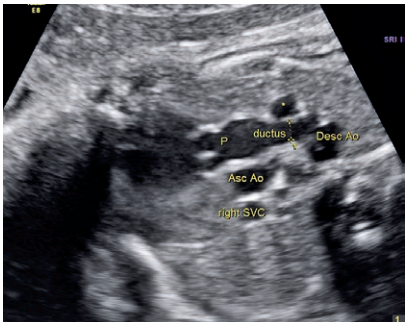


Fig 6. Premature constriction of the arterial duct at 36 weeks. In the "three vessels" wiev: P – pulmonary trunk, Desc Ao – descending aorta, Asc Ao – ascending aorta, SVC- superior vena cava, asterix – persistent left superior vena cava. Ductus arteriosus was minus 1.35 on Z score

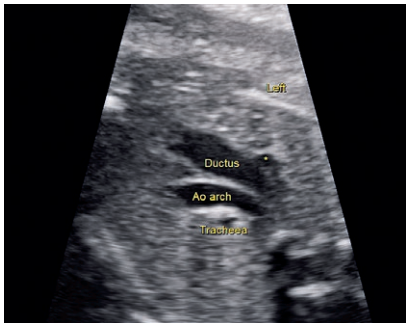


Fig 7. Three vessels view in the absence of the right superior vena cava. Asterix – left superior vena cava

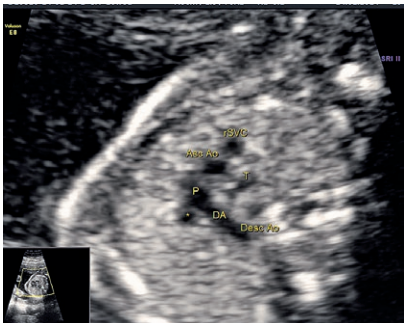


Fig 8. Typical appearance of a simple persistent left superior vena cava (with asterix*) in the "three vessels" trachea view: P – pulmonary trunk, DA – ductus arteriosus, Desc Ao – descending aorta, T – trachea, Asc Ao – ascending aorta, rSVC – right superior vena cava

PLSVC was 8.88%, similar to the data in the literature. The complex fetal cardiac anomalies are associated in 23% of the cases with the presence of PSLVC [12] and in 9% of cases the PLSVC can be the single anomaly [13]. In our study the isolated cases of PLSVC represent 13% of the cardiac anomalies. As reported, the most frequent cardiac abnormalities associated with PSLVC are atrial septal defect, ventricular septal defect, aortic coarctation, transposition of great vessels, tetralogy of Fallot, and anomalous connections of the pulmonary veins, in this order of incidence [3,14,15]. PLSVC is not only associated with cardiac anomalies, but also with extracardiac malformations [12,13].

Regarding the prognosis, if PLSVC is isolated the prognosis is excellent, the anomaly being asymptomatic [16]. Otherwise, there is a significant morbidity and mortality, which results especially from the associated abnormalities [10,12,13,17]. Identification in the fetal life of the PLSVC increases the risk of cardiac malformation by 50 times [16]. Identification of an associated complex fetal cardiac anomaly is a sign of bad prognosis [18]. Usually the outcome of the fetuses with PLSVC depends on the presence of an associated heart anomaly, and if present, also on its severity, similar as we observed in our study [10].

In our newborn data, the prognosis of the 3 isolated cases and of the 4 cases with PLSVC in the absence of right superior vena cava was good. In 2 newborns the suspicion of aortic coarctation was not confirmed The rest of our cases had the following postnatal evolution: 5 newborns underwent surgical therapy or had a planned surgical therapy for cardiac associations, one is under cardiologic monitoring with possible surgical intervention in the future for aortic coarctation, one is under therapy for supraventricular tachycardia, 4 newborns died (2 with complex cardiac malformations and 2 with multiple malformations), 2 medical abortions were done for severe malformations, and one with "in utero" premature ductal arch constriction had a good evolution after birth.

A review of 238 cases reported from almost 20,000 fetal and pediatric echocardiograms, justifies why we cannot consider PLSVC as a variant of normality [17].

In **conclusion,** even though it is rare, persistent superior left vena cava can present different associations;

identification of this anomaly in the section of "three vessels" needs supplementary fetal cardiac evaluation.

Acknowledgement: This paper was partially supported by internal research grants from the University of Medicine and Pharmacy of Târgu Mureș, România, under contract number 1093/1/30.01.2013.

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