The Role of 3-Dimensional Ultrasound for the Diagnosis of LPSVC and A Review of the Literature

Wael El Guindi1*, Ahmed Ellithy2, Sondos Salem3, Islam Badr1, Sameh Abdellatif3, Sherif Elmekkawi4 and Mahmoud Alalfy2

1El Guindi Clinic, 6 October City, Egypt
2Kaser El Ainy Teaching Hospital, Cairo, Egypt
3National Research Centre, Cairo, Egypt
4Ain Shams University

Abstract

Objective: The purpose of this article is to review the published literature and determine the role of 3-dimensional ultrasonography (3DUS) in the diagnosis of LPSVC.

Methods: The volume data sets were acquired with transverse sweeps through the fetal chest. Volumes were stored in the ultrasound machine's hard drive and subsequently analyzed offline.

Results: LPSVC was unequivocally diagnosed and the diagnosis confirmed by postnatal US.

Conclusion: 3D ultrasonography especially power Doppler imaging made it possible to unequivocally confirm the diagnosis of the LPSVC and demonstrate LPSVC entering the dilated coronary sinus. The images obtained made it easy to explain this anomaly to the patient. This case of isolated LPSVC in a 30-week fetus demonstrates the added value of 3D ultrasonography and especially 3D power colour in fetal echocardiography.

Case Presentation

A 39-year-old woman, gravida 2 para 1, presented at 30 weeks' gestation for detailed targeted organ scanning with fetal echocardiography, after having received contradictory opinions concerning a suspected atroventricular canal defect during previous ultrasound examinations. Karyotype, which had been tested because of advanced maternal age, was normal. Targeted organ scanning for exclusion of fetal anomalies was performed, including complete fetal echocardiography 2 and 3D US and coulor Doppler and 3D Power Doppler power imaging (Figures 1-3).

In the present case, the initial diagnosis of LPSVC was made on observation of a dilated coronary sinus and an extraneous vessel identified to the left of the ducal arch in the three-vessels and trachea (3VT) view of the fetal heart (Figures 1E, F and 3D).

We performed ultrasound examinations using a GE Voluson 730 pro. (Voluson 730, GE Healthcare, Milwaukee, WI, USA) using a 3-5-MHz mechanical volume transabdominal transducer. The volumes were stored and subsequently analyzed offline.

Meticulous inspection of the fetal anatomy did not reveal any associated cardiac or extra cardiac anomalies and fetal growth was on the 50th centile. A male neonate was spontaneously delivered at 38 weeks of gestation with a birth weight of 3150 g with Apgar scores of 8, 9 and 10 at 1.5 and 10 min, respectively. Postnatal adaptation was uneventful. Postnatal echocardiography confirmed the prenatal diagnosis.

3D power Doppler imaging made it possible to unequivocally confirm the diagnosis of the LPSVC and demonstrate LPSVC entering the dilated coronary sinus (Figures 1 and 3). The images obtained made it easy to explain this anomaly to the patient. This case of isolated LPSVC in a 30-week fetus demonstrates the added value of power Doppler 3D in fetal echocardiography.

It appears that 3D power Doppler imaging applications will make a significant contribution to our understanding of the developing fetal heart in both normal and anomalous cases, to interdisciplinary management team consultation, to parental counselling, and to professional training.

Comment

Persistent left superior vena cava is a rare but is the most common congenital anomaly of the systemic thoracic venous return. It is found in 0.3-0.5% of the general population [1-7]. The reported prevalence of persistent in CHD patients varies widely according to the diagnostic tool utilized it ranges from 1.3%, 2%, 3 to 10% and in up to 12% in association with congenital heart diseases [2,5,7-8].

In normal fetal development, the paired anterior cardinal veins drain the upper body and extremities of the fetus and become connected by an oblique vessel. During the 8th week of embryological development, a communication forms between the veins (future innominate [brachiocephalic] vein). The distal end of the left anterior cardinal vein degenerates (forms the ligament of Marshall), and is represented at birth by a small vestigial vein adjacent to the posterior wall of the left atria (Marshall’s oblique vein) and by a fibrous band attached to this vein (Marshall’s ligament), the remaining right anterior cardinal vein ultimately becomes the Right Superior Vena Cava (RSVC).

The distal end of the left anterior cardinal vein drains into the left horn of the sinus venosus (future coronary sinus) before it degenerates; this explains the drainage pattern of a PL SVC into the coronary sinus Failure of closure of the left anterior cardinal vein during cardiac development results in PL SVC [1,4,9-11].

Failure of the distal left anterior cardinal vein to degenerate is possible with or without degeneration of the right cardinal veins (Figure 3A). A genetic culprit may be genes for left- right signalling [12]. In general, Patients with left superior vena cava (LSVC) usually

*Corresponding author: Wael El Guindi, El Guindi Clinic, 6 October City, Egypt, Tel: 202 3837 2255; E-mail: welguindi@yahoo.com

Received December 30, 2013; Accepted January 20, 2014; Published January 31, 2014

Citation: El Guindi W, Ellithy A, Elmekkawi S, Badr I, Abdellatif S, et al. (2014) The Role of 3-Dimensional Ultrasound for the Diagnosis of LPSVC and A Review of the Literature. Gynecol Obstet (Sunnyvale) 4: 202. doi:10.4172/2161-0932.1000202

Copyright: © 2014 El Guindi W, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
A, B, C: Normal three-vessel view. The 3-V V and trachea.
RA, right atrium; Left ventricle, right common carotid artery; RV, right ventricle; SVC, superior vena cava; T, trachea; Ao, aorta, PA, pulmonary artery.

D: 3D power Doppler ultrasound showing the LPSVC drains into the dilated coronary sinus and to the right atrium. RA, right atrium; Left ventricle, right common carotid artery; RV, right ventricle; SVC, superior vena cava; T, trachea; Ao, aorta, PA, pulmonary artery, RSVC, right superior vena cava, PLSVC, persistent left superior vena cava, RA, right atrium, UV, umbilical vein, CS, coronary sinus.

Figure 1: Transverse view of the upper mediastinum

Figure 2: A, B and C: Four vessel view showing a left persistent superior vena cava (LPSVC). D: 3D power Doppler ultrasound showing the LPSVC drains into the dilated coronary sinus and to the right atrium. RA, right atrium; Left ventricle, right common carotid artery; RV, right ventricle; SVC, superior vena cava; T, trachea; Ao, aorta, PA, pulmonary artery, RSVC, right superior vena cava, PLSVC, persistent left superior vena cava, RA, right atrium, UV, umbilical vein, CS, coronary sinus.

have a normal right superior vena cava are asymptomatic, and the condition is infrequently detected a casual finding during an imaging test (CAT, NMR or venography) or during intervention (catheterization, pacemaker or defibrillator implantation, or surgery), and it has only practical implications when considering a left superior approach for implantation of a pacemaker, a cardioverter-defibrillator, or positioning of a central venous line [13,14].

When isolated, LPSVC usually has no clinical significance; however, it has been reported to occur in isolation in only 9% of cases [15]. Associated congenital cardiac defects (atrial septal defects, ventricular septal defects, endocardial cushion defects, and tetralogy of Fallot) and high incidence of rhythm disturbances may be detected in patients with PLSVC [16]. Presence of a LPSVC was first reported by Le Chat in 1787, and the first account of foetal structures comprising LPSVC was published by Marshall in 1850 [17].

According to Schummer, the superior vena cava system can be classified as type I, normal anatomy; type II, only LPSVC; type IIIa, right and left superior vena cava with connection; and type IIIb, right and left superior vena cava without connection [18,19]. The most frequent variant (82%) of the normal anatomy is the simultaneous presence of both right and left cava veins (double superior vena cava, type III) and so this anomaly is often missed as central line catheters are more commonly inserted on the right side [20,21]. Persistence of Left SVC with, absent right superior vena cava (type II) occurs in only 0.09-0.13% of patients with congenital heart disease [11]. It is often an isolated anomaly [21].

Left SVC drains either into right atrium via coronary sinus or into left atrium. 92% of Left SVC drains into right atrium via left portion of sinus venous or coronary sinus and are asymptomatic [22]. They are considered to be anomaly of coronary sinus [23]. In this case, the ostium of the coronary sinus opens directly into the right atrium in close proximity to the insertion of the atroventricular valves [24]. If the coronary sinus is dilated, it can create the appearance of an atroventricular canal defect (as in the present case) Park et al presented three cases in which the correct diagnosis avoided an unnecessary termination of pregnancy [25]. In 8% of cases, it connects to the left atrium in such variants with absent or unroofed coronary sinus or normal coronary sinus and so creates a right-to-left shunt and
paradoxical emboli and [26,27]. 3D power Doppler imaging made it possible to unequivocally confirm the diagnosis of the LPSVC and demonstrate LPSVC entering the dilated coronary sinus (Figures 1 and 3). The images obtained made it easy to explain this anomaly to the patient. This case of isolated LPSVC in a 30-week fetus demonstrates the added value of power color Doppler 3D in fetal echocardiography. In addition, 3D/4D image processing is advantageous compared with 2D ultrasound because detailed offline evaluation is possible; spatial relationships between lesions are easily demonstrated; blood vessel courses can be followed completely; volume calculations provide more accurate information on areas; and volumes can be inspected from different angles [28]. The digital storage capabilities of 3D/4D imaging enable remote diagnosis. The virtual volume can be stored and reloaded to allow follow-up examinations, comparisons, video conferencing, or revision of initial diagnosis. Furthermore, the volume dataset can be analyzed offline or reviewed by experts in another centre connected by the internet [29].

References

1. Nay T Tun, Thein T Aun, Swamaralatha Kannegant, Koroush Khalighi (2013) Persistent Left Superior Vena Cava. Cath Lab Digest. 22. http://www.cathlabdigest.com/articles/Persistent-Left-Superior-Vena-Cava

2. Iimura A, Oguchi T, Shibata M, Matsuo M, Takahashi T (2011) Double superior vena cava and anomaly of cardiovascular system with a review of the literature. Okajimas Folia Anat Jpn 88: 37-42.

3. Demos TC, Posniak HV, Pierce KL, Olson MC, Muscato M (2004) Venous anomalies of the thorax. AJR Am J Roentgenol 182: 1139-1150.

4. Ramos N, Fernández-Pineda L, Tamariz-Martel A, Villagrá F, Egurbide N, et al. (2005) [Absent right superior vena cava with left superior vena cava draining to an unroofed coronary sinus]. Rev Esp Cardiol 58: 984-987.

5. Vizzardi E, Fracassi F, Farina D, Nardi M, D’Aloia A, et al. (2008) Persistence of left superior vena cava, absence of coronary sinus and cerebral icus. Int J Cardiol 126: e39-41.

6. Goyal SK, Punnam SR, Verma G, Ruberg FL (2008) Persistent left superior vena cava: a case report and review of literature. Cardiovasc Ultrasound 6: 50.

7. Pucelikova T, Kautznerova D, Vedlich D, Tintera J, Kautzner J (2007) A complex anomaly of systemic and pulmonary venous return associated with sinus venosus atrial septal defect. Int J Cardiol 115: e47-48.

8. Shiekh Eldin G, El-Segaier M, Galal MO (2013) High prevalence rate of left superior vena cava determined by echocardiography in patients with congenital heart disease in Saudi Arabia. Libyan J Med 8: 21675.

9. Maithili Shenoy, Omaira Ali, Tushar Tuliani, Nour Jurafli2, Mahir Elder (2011) Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava: Out of Mind is Out of Sight?. J Clinic Case Reports. 2: 1.

10. Sergey G Toshinskiy et al. (2013) Pediatric Surgery for Unroofed Coronary Sinus Medscape. 4
11. Pálinkás A, Nagy E, Forster T, Morvai Z, Nagy E, et al. (2006) A case of absent right and persistent left superior vena cava. Cardiovasc Ultrasound 4: 6.

12. Jacob M, Sokoll A, Mannherz HG (2010) A case of persistent left and absent right superior caval vein: An anatomical and embryological perspective. Clin Anat 23: 277-286.

13. Delicia I, Gentille Lorente (2013) Persistent Left Superior Vena Cava in a Patient with Cardiac Conduction Abnormality; Rev Fed Arg Cardiol. 42: 219-221.

14. Biffi M, Boriani G, Frabetti L, Bronzetti G, Branzi A (2001) Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation: a 10-year experience. Chest 120: 139-144.

15. Yagel S, Kivilcivitch Z, Cohen SM, Valsky DV, Messing B, et al. (2010) The fetal venous system, Part II: ultrasound evaluation of the fetus with congenital venous system malformation or developing circulatory compromise. Ultrasound Obstet Gynecol 36: 93-111.

16. Recupero, P. Pugliatti, F. Rizzo, et al. (2007) "Persistent left-sided superior vena cava: Integrated noninvasive diagnosis," Echocardiography. 24: 982-986.

17. Sarodia BD, Stoller JK (2000) Persistent left superior vena cava: case report and literature review. Respir Care 45: 411-416.

18. Schummer W, Schummer C, Fröber R (2003) Persistent left superior vena cava and central venous catheter position: clinical impact illustrated by four cases. Surg Radiol Anat 25: 315-321.

19. Soward A, ten Cate F, Fioretti P, Roelandt J, Serruyts PW (1986) An elusive persistent left superior vena cava draining into left atrium. Cardiology 73: 368-371.

20. Ghadiali N, Teo LM, Sheah K (2006) Bedside confirmation of a persistent left superior vena cava based on aberrantly positioned central venous catheter on chest radiograph. Br J Anaesth 96: 53-56.

21. Pasquini L, Belmar C, Seale A, Gardiner HM (2006) Prenatal diagnosis of absent right and persistent left superior vena cava. Prenat Diagn 26: 700-702.

22. Vladimir Boshkov, Lidiya Poposka, Ilina Danilovska (2013) Vascular anomaly - persistent left superior vena cava, Cardiologia Croatica. (9):301. http://www.kardio.hr/pdf/Cardiologia%20Croatica_2013_9_301.pdf

23. Sharma O P, Sethil S (2010) Left sided Superior Vena cava: (A case report and review of literature). Asian Journal of Medical Sciences. 1: 18-19

24. Paval J, Nayak S (2007) A persistant left superior vena cava. Singapore Med J 48: e90-93.

25. Park JK, Taylor DK, Skeels M, Towner DR (1997) Dilated coronary sinus in the fetus: misinterpretation as an atrioventricular canal defect. Ultrasound Obstet Gynecol 10: 126-129.

26. Biffi M, Boriani G, Frabetti L, Bronzetti G, Branzi A (2001) Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation: a 10-year experience. Chest 120: 139-144.

27. Hutyra M, Skala T, Sanak D, Novotny J, Köcher M, et al. (2010) Persistent left superior vena cava connected through the left upper pulmonary vein to the left atrium: an unusual pathway for paradoxical embolization and a rare cause of recurrent transient ischaemic attack. Eur J Echocardiogr 11: E35.

28. Zhang M, Pu DR, Zhou QC, Peng QH, Tian LQ (2010) Four-dimensional echocardiography with B-flow imaging and spatiotemporal image correlation in the assessment of congenital heart defects. Prenat Diagn 30: 443-448.

29. El Guindi W, Dreyfus M, Carles G, Lambert V, Herliciovecz M, et al. (2013) 3D ultrasound and Doppler angiography for evaluation of fetal cardiovascular anomalies. Int J Gynaecol Obstet 120: 173-177.