Volume-4, Issue-12, Dec-2015 • ISSN No 2277 - 8160

Research Paper | Medical Science

Persistent Left Superior Vena Cava Diagnosed Antenatally by Fetal Echocardiography – a Rare in Occurrence Case Report

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ABSTRACT

Persistent left superior vena cava is rare but important congenital vascular anomaly. It is the most common congenital thoracic venous anomaly with a prevalence of 0.3–0.5% in general population. It results when the left superior cardinal vein caudal to the innominate vein fails to regress. It is most commonly observed in isolation but can be associated with other cardiovascular abnormalities including atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum.

Here we describe a case of 29 year old female who reported to us for regular antenatal growth scan was found to have persistent left superior vena cavae draining into coronary sinus at 30 weeks and 5 days of gestational age.

KEYWORDS : PLSVC, Fetal, Antenatally.

Introduction :

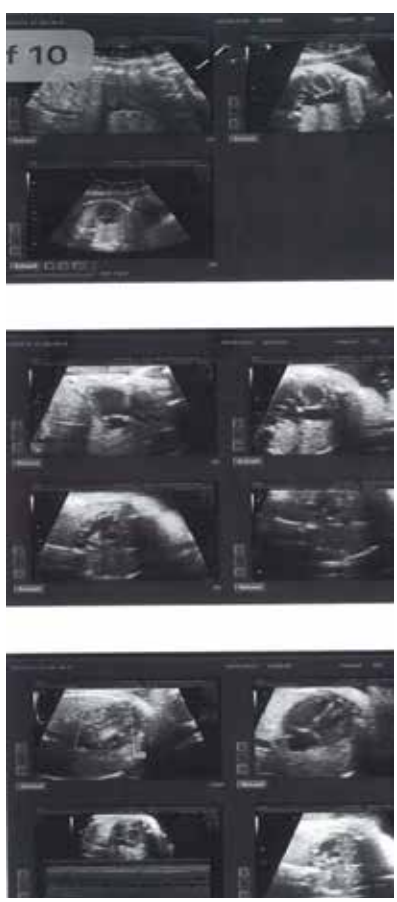
In this modern technical era, with the use of fetal ultrasound (US) and fetal echocardiography, diagnosis of various congenital heart defects (CHD) is more frequent. Although the descriptions of persistent LSVC in the adult date back to 1787, prenatal diagnosis of anomalous systemic venous return (ASVR)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

The presence of PLSVC can render access to the right side of heart challenging via the left subclavian approach, which is a common site of access utilized when placing pacemakers and Swan-Ganz catheters. Incidental notation of a dilated coronary sinus on echocardiography should raise the suspicion of PLSVC. The diagnosis should be confirmed by saline contrast echocardiography.

It is usually asymptomatic and is detected when cardiovascular imaging is performed for unrelated reasons. When a left subclavian approach is used for vascular access, its presence can complicate catheter placement within the right side of heart. Here we present a case that highlights the practical implications of PLSVC in a foetus of 30 weeks and 5 days gestational age diagnosed on fetal echo of 29 years old female.

Case Report:

A 29 year old G1P1 female of 30 weeks and 5 days gestational age reported to our out-patient department for routine ante-natal checkups. As a routine growth scan was advised. Incidentally fetal echocardiography showed normal cardio-thoracic ratio, normal situs, normal 4 chamber view, 3 vessel view showed left superior vena cava draining into coronary sinus(4mm in diameter), inter atrial septum with foramen ovale and flap valve were visualised to be normal, interventricular septum is normal in thickness, no evidence of defect noted, both ventricular outlets are normal, normal pulmonary veins, normal aorta and pulmonary arteries, heart pulsations were 143 beats per minute, no mass lesions in the heart noted, heart rate regular with no evidence of arrhythmias, no obvious lethal structural congenital anomalies noted. The fetal echocardiography findings were suggestive of



persistent left superior vena cava draining into coronary sinus.

Image 1: Showing PLSVC by Fetal Echocardiography.

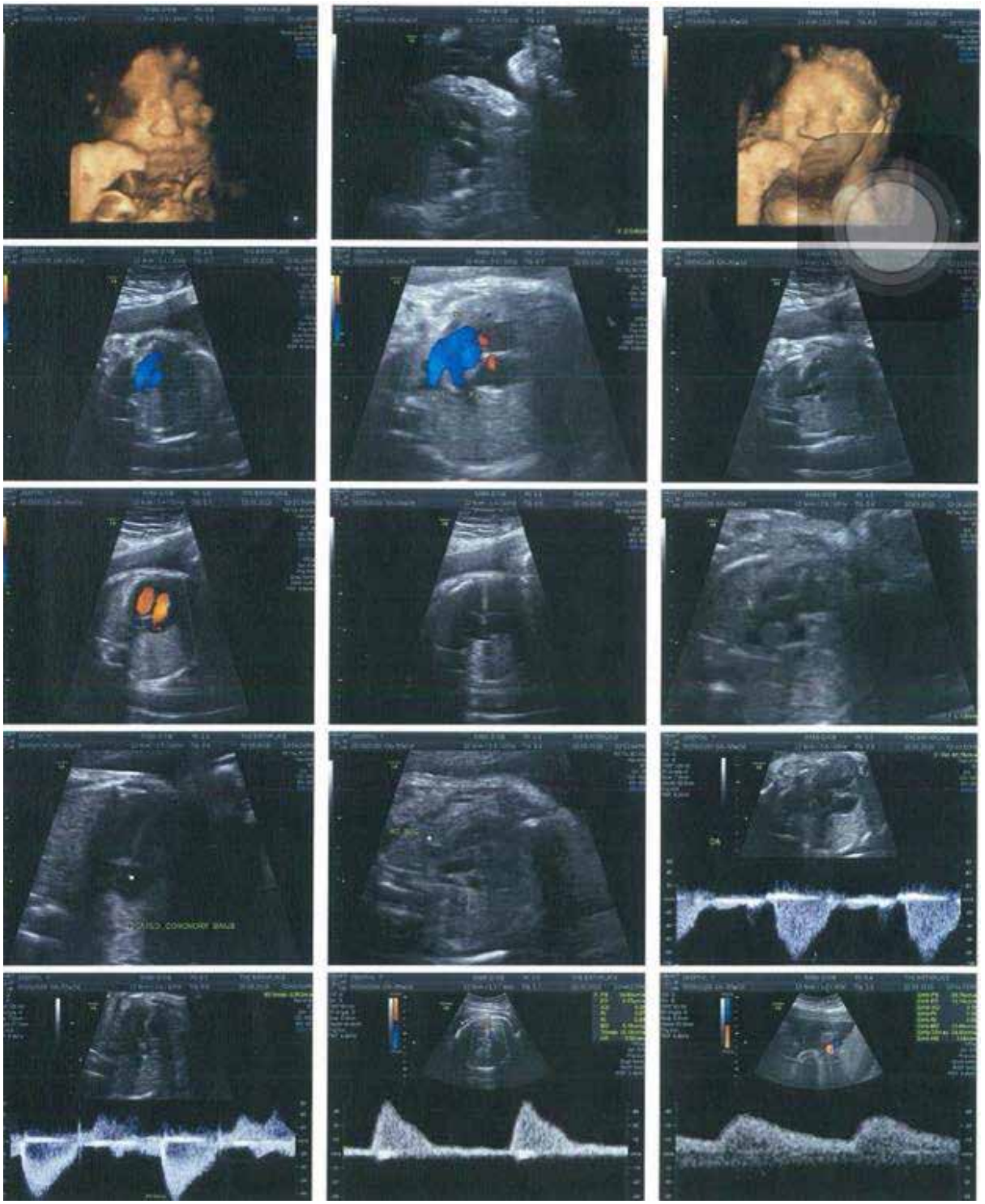


Image 2: Showing the total view in sections of PLSVC in Fetal Echocardiography.

Discussion:

Persistent left SVC is the most common congenital thoracic venous anomaly with a prevalence of 0.3–0.5% in general population.[6] The thoracic embryonic venous system is composed of two large veins (the superior cardinal veins) which return blood from cranial aspect of embryo, and the inferior cardinal vein, which returns blood from the caudal aspect. Both pairs of veins join to form right and left common cardinal veins before entering the embryological heart. The left common cardinal vein persists to form coronary sinus and oblique vein of left atrium. During the 8th week of gestation, an anastomosis forms between right and left superior cardinal veins resulting in the innominate (or brachiocephalic) vein. The cephalic portion of superior cardinal veins form the internal jugular veins. The caudal portion of right superior vein forms the normal right-sided superior vena cava, while the portion of the left superior cardinal vein caudal to the innominate vein normally regresses to become "ligament of Marshall". If this normal regression of the left superior cardinal vein fails to occur, a persistent left-sided vascular structure that empties into the coronary sinus, results (the PLSVC). The innominate vein may or may not degenerate in these cases leading to variations in anatomy.

The most common subtype of PLSVC results in the presence of both left and right SVCs. A bridging innominate vein may or may not be present. Webb et al[7] reported that a PLSVC is associated with absence of the innominate vein in 65% cases. More rarely, the caudal right superior cardinal vein regresses leading to an absent right SVC with PLSVC. In this case, the left SVC returns all the blood from cranial aspect of the body. Variations have also been reported in the insertion of left SVC. In 80–90% of individuals, the persistent LSVC drains into the right atrium via the coronary sinus and is of no hemodynamic consequence. In the remaining cases, it may drain in left atrium resulting in a right to left sided shunt.

Diagnosis of left SVC is usually made as an incidental finding during cardiovascular imaging or surgery. In this case we report you PLSVC draining into coronary sinus in a foetus of 30 weeks and 5 days age which is rare in occurrence to be diagnosed at this age.

Almost 40% of patients with PLSVC can have a variety of associated cardiac anomalies,[8,9] such as atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum. The presence of associated anomalies is more common with concomitant absence of right SVC the notation of which warrants appropriate investigation to rule out other anomalies. The PLSVC has been associated with anatomical and architectural abnormalities of the sinus node and conduction tissues. Both sinus and AV node can have persistent fetal dispersion in the central fibrous body in subjects with PLSVC.[10]

PLSVC 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 as it is performed without imaging under many circumstances, such as at the bed- side. PLSVC can also complicate permanent pacemaker and implantable cardioverter defibrillator (ICD) placement (the latter of which is always done under fluoroscopic guidance thus the anomaly is typically detected during the procedure). Serious complications such as arrhythmia, cardiogenic shock, cardiac tamponade, and coronary sinus thrombosis have been reported when pacemaker leads or catheters have been inserted via PLSVC. Fortunately, the incidence of such complications is relatively low, and permanent pacemaker leads for single chamber pacing have been successfully placed via PLSVC as early as 1971.[11]

During cardiac surgery, the presence of PLSVC is a relative contraindication to the administration of retrograde cardioplegia. It may be possible to clamp the PLSVC to pre- vent the cardioplegia solution from perfusing retrograde up the PLSVC and its branches with inadequate myocardial protection.[12] However, there is a possibility that there may be some steal of cardioplegia solution through an accessory vein. During heart transplantation in a patient with PLSVC, the coronary sinus must be dissected carefully to permit re-anastomosis of PLSVC to right atrium.[13]

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.[4,14–17] Reported cardiac abnormalities include heterotaxy (left and right isomerism), with 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, (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.[18] 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.[18–20] In more than 90% of cases, the LSVC drains into the right atrium via the coronary sinus and physiologically there are no clinical consequences.[21] 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.[22] 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.[23,24]

Persistent LSVC and other anomalies:

In a postnatal series, Postema, (2008) have also shown the frequent association between a persistent LSVC and extracardiac anomalies.[25] 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).

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, 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.[26,27,28] Failure of this normal regression can be attributed to the formation of a persistent LSVC.[29] The is 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.[14]

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.[31] The addition of the three vessel view at the upper mediastinum to fetal cardiac examination has facilitated easy and accurate prenatal diagnosis of persistent LSVC. A recent case series study by Barrea, (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.[30] 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 sinus. This view is described in pediatric echocardiography and was reproduced in fetal echocardiography by Freund, (2008).[5] An isolated enlarged coronary sinus is highly suggestive of persistent LSVC, although this finding may have both false positive and false negative diagnoses.[3,31–38]

Conclusion :

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. We describe here a case of PLSVC which is very rare in occurrence and was diagnosed at regular antenatal growth scan. The key to diagnosis of PLSVC is the three vessel view. The defect in isolation is however generally associated with favourable prognosis as in this case but it might be an important finding later in life where the necessity of cardiac intervention is to be required.

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