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Persistent left superior vena cava –a benign or a sinister finding in fetal life-a review

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Introduction

Persistent left side superior vena cava (PLSVC) is an uncommon venous anomaly that occurs as a result of persistence of caudal part of left anterior cardinal vein (LACV). However, it is the most common anomaly of the thoracic venous system[1]. Its seen is about 0.2-0.5% of people with normal heart[1,2] and 5-9% in those with congenital heart disease (CHD).[2–5] When isolated, PLSVC is of no hemodynamic significance and is considered as a benign variant. However, a detailed fetal and post-natal echocardiogram is advised to rule out any structural heart defects. Its presence should be documented as it is important while doing cardiac procedures later in life.

Embryology

Heart is the first organ to develop in an embryo. There are three pairs of veins that drain the embryo: - umbilical veins, vitelline veins and cardinal veins. There are two pairs of cardinal veins – anterior and posterior- which drains the body of the embryo. The anterior ones drain the cranial parts and the posterior drains the caudal parts of the body. They both drain via common cardinal veins of either side into the heart. Each anterior cardinal vein (ACV) has a cephalic and a caudal part (Figure 1a). The cephalic part of the ACV forms the internal jugular vein. On the right side, the caudal portion of ACV and the corresponding common cardinal vein forms the right sided superior vena cava (SVC). The caudal portion of left anterior cardinal vein (LACV) obliterates[6] (Figure 1b). This forms the ligament of Marshalls, first described in 1850 as a "vestigial fold of pericardium"[7] (Figure 1b). The left common cardinal vein forms the coronary sinus. Failure of regression of the caudal portion LACV leads to the formation of PLSVC (Figure 1).

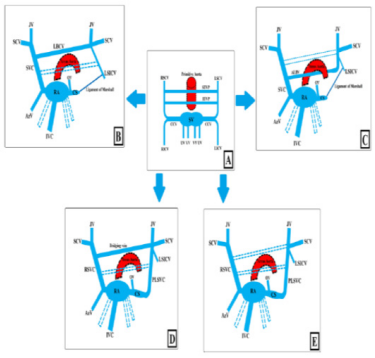

Figure 1 Development of venous system in the fetus.[8]

Anatomical variants

When the caudal portion of LACV fails to disappears, the left jugular and left subclavian vein forms LSVC. It runs down along the lateral border of the left atrium anterior to the aortic arch and left pulmonary artery.[2,8–10] PLSVC, when present, is approximately responsible for 20% of venous drainage from the left arm and the left half of the head and neck in the fetus.[2,8,11,12] It joins the coronary sinus (CS) in the atrioventricular groove and then drains into the right atria in majority of the cases (90%). PLSVC drainage into right atria is usually of no hemodynamic consequence. However, certain changes can occur due to its presence. The possible underlying mechanisms[2,8,11] for such changes are tabulated in the Table1.

Table 1 Mechanism of hemodynamic changes due to PLSVC

Changes	Mechanisms
Decrease in mitral valve area	Compression due to enlarged CS or increase in the size of right atria due to increased venous return
Decrease in the size of right superior vena cava	Decreased venous return through right superior vena cava

PLSVC can also directly drain into left atria or via left pulmonary veins or CS in 10-20% cases.[8] The drainage of PLSVC into CS is called as unroofed CS or CS atrial septal defect. The association of atrial septal defect and PLSVC draining into unroofed CS is called Raghib Syndrome.[8,11,12] Left atrial drainage leads to left to right shunt and is associated with cardiac anomalies. PLSVC and right SVC are present in 90% cases, called as double SVC. The right SVC can be smaller or larger in size in relation to LSVC. Usually, they run along their respective sides without any connection. Sometimes, there may be a bridging vein (LBCV) between the two[13] (Figure 1c&d). When there is regression of caudal portion of right ACV, a rare situation, there is only LSVC. This situation involves a detailed evaluation of fetal heart as it is associated with CHD[8,12,14] (Figures 2–4).

Antenatal diagnosis

With the advancement in the resolution of the ultrasound and the inclusion of three vessel trachea view (3VT) in the basic evaluation of the fetal heart, the detection rate of PLSVC has increased in the prenatal life over the years.[15–17] PLSVC is identified as an extra (4th vessel) in the 3 VT section to the left and anterior to the pulmonary artery on grey scale (Figure 5a).[2,9,10] A slightly cranial section (Figure 5b) to the 3VT shows both SVCs with the absence of left branchio-cephalic vein (LBVC).[2,18] In rare cases, right SVC may be absent,

hence there may be only 3 vessels (Figure 2–4) in the 3VT section.[2,14] A dilated coronary sinus can be seen in the 4-chamber view inferior to the mitral valve[2,13,19] which acts a clue to look for PLSVC (Figure 6).

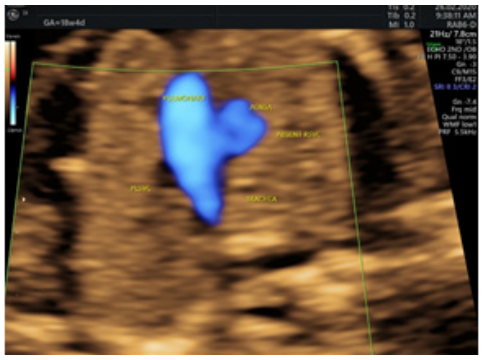

Figure 2 Colour 3VT view shows a case of LSVC without right SVC. You can see a vessel on the left of pulmonary artery but right SVC could not be seen.

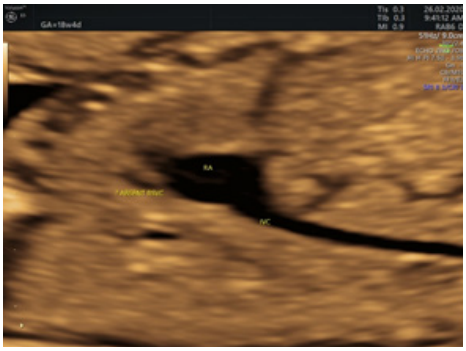

Figure 3 Hammock view when right SVC could not be traced draining into right atria.

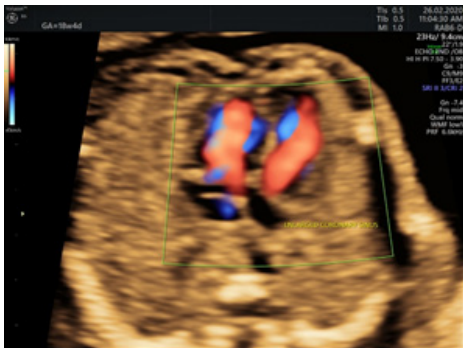

Figure 4 Colour 4 Chamber view shows presence of PLSVC. You can see the presence of enlarged coronary sinus which can give false impression of AVSD.

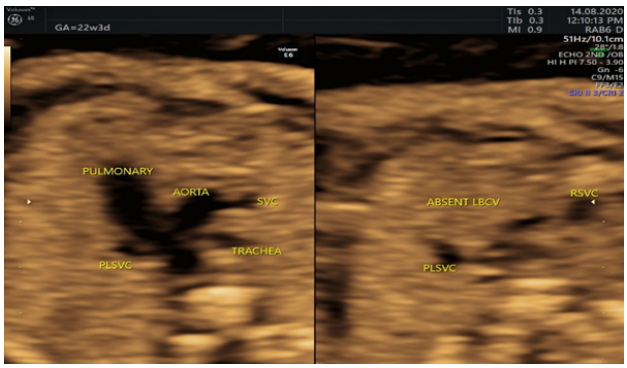

Figure 5 a) 3VT section shows PLSVC as the 4th vessel on left of pulmonary artery. b) shows absent LBCV with right and left SVC.

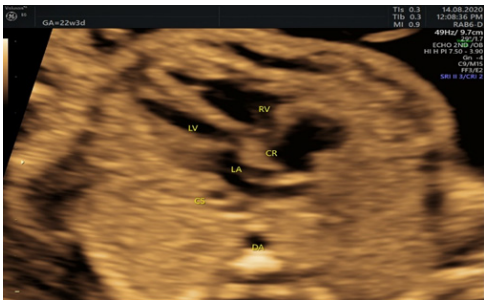

Figure 6 4 Chamber view shows enlarged coronary sinus (CS).

Differential diagnosis

The differential diagnosis for the 4th vessel seen in the 3 VT section includes the vertical vein in the supra-cardiac variety of total/partial anomalous pulmonary venous connection (TAPVC/ PAPVC) and rarely, double aortic arch (DAA) could also be seen as 4 vessels. The other rare differentials for vessel on the left side of the mediastinum includes levoatriocardinal vein, aberrant LBCV, left superior intercostal vein and pericardiophrenic vein.[8]

In TAPVC type I, the 4th vessel seen in the 3VT section is the vertical vein. In vertical vein, the colour Doppler flow is upwards towards the thorax and a dilated LBCV is present. The vertical vein ascends upwards drains into LBCV which drains into right SVC (Figure 7). However, in PLSVC, the colour flow is down towards the heart and the LBCV is absent. PLSVC is the most common cause of dilated coronary sinus (CS).[20] Coronary ostial stenosis can also cause its enlargement but is also associated with PLSVC. The anomalous pulmonary drainage into coronary sinus also causes enlarged CS, difficult to be diagnosed in utero. This effect is minimal as the pulmonary venous flow is very less in the fetal life. Other causes are like coronary or ventricular fistula draining into CS, but these have not been diagnosed in utero.[20] DAA will be differentiated by means of presence of right sided aortic arch and lambda sign formed by right and left aortic arch. It can be seen on slightly cranial and oblique views on colour Doppler.

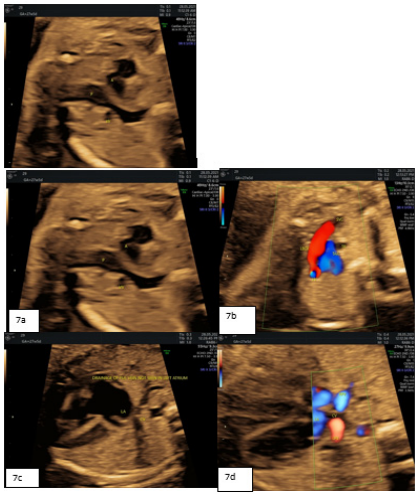

Figure 7 shows supra-cardiac variety of TAPVC. a 3VT section shows a 4th vessel on the left of pulmonary artery identified as vertical vein. B. section slightly cranial to 3VT shows presence of vertical vein draining into LBCV which in turn is draining into SVC. c. grey scale 4 chamber view shows that the pulmonary veins do not drain into left atria. You can see that the pulmonary veins form a common vein (CV) which ascends as vertical vein to drain into LBVC. Also, there is presence of enlarged coronary sinus as indicated by solid black arrow (◄). d. 4 chamber view in power Doppler.

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Levoatriocardinal vein is a rare anomaly associated with hypoplastic left heart with intact atrial septum. In these cases, there is abnormal persistence of anastomosis that connects the embryonic foregut to the cardinal veins. It helps to decompress the left atria into brachiocephalic or azygous veins.[21] It is considered as a distinct entity from PLSVC.[22] Aberrant course of LBCV when it courses inferiorly along the left side of mediastinum and behind the aorta or behind the oesophagus to drain into right brachiocephalic vein. It is a rare condition and is associated with CHDs. Its location in the 3VT view can help to easily differentiate it from PLSVC.[8,23] However, the other rare causes are difficult to differentiate in the fetal life but they gain importance in the postnatal life, if interventions like cardiac catheterizations are required.

Associations

LSVC is known to be associated with cardiac, extra-cardiac and chromosomal abnormalities.

PLSVC and CHD

Presence of PLSVC warrants a detailed fetal cardiac evaluation as there is 61% risk of having a cardiac abnormality. Even in isolated cases, there is 2.4% risk of having a cardiac abnormality being detected later or after birth.[24] The wide variety of cardiac abnormalities have been reported in literature as shown in Table 2 in association with PLSVC.[1,8, 25,26]

Table 2 Variety of cardiac abnormalities reported in literature in association with PLSVC

Main groups	Subgroups
Shunt lesions	Ventricular septal defects, Atrial septal defects, Atrio-ventricular septal defects
Conotruncal malformations	Tetralogy of Fallot, Pulmonary atresia, Double Outlet right ventricle, Transposition of great arteries
Left sided obstructive lesions	Coarctation of aorta, Mitral Stenosis, Bicuspid aortic valve
Right sided obstructive lesions	Ebsteins anomaly, Pulmonary stenosis or atresia, Tricuspid atresia
Aortic arch anomalies	Right aortic arch with or without Aberrant left subclavian artery, Double aortic arch, Aberrant right subclavian artery

Hence, presence of PLSVC is a strong marker for CHDs. Galindo had reported increased frequency (41%) of heterotaxy syndromes in cases with PLSVC.[1] Also, the prevalence of aortic anomalies especially Coarctation of aorta (CoA) deserves a special mention in this case. 2016 meta-analysis shows that in 21% of isolated cases of PLSVC, coarctation of aorta was found.[24] Another systematic review did not find an increased risk of association of CoA with PLSVC, but they still recommend screening for CoA in such cases on follow up.

PSLVC can lead to disproportionate ventricles (Figure 3,4). It could be due to decrease in the size of left atria and mitral valve area because of coronary sinus enlargement. However, in certain cases, increased drainage into the right atrium can also cause enlargement in the size of the right atria and ventricle as well.[8,11] However, the possibility of its association with CoA which can also lead to discrepant ventricles, should also be kept in mind (Figure 3,4).

PLSVC and extra-cardiac abnormalities

38% of cases with PLSVC had extra-cardiac abnormalities. In 6% cases, such abnormalities might be detected later or even after birth.[24] The extra-cardiac abnormalities reported are increased nuchal translucency, single umbilical artery, agenesis or aberrant drainage of ductus venosus, hemivertebrae, esophageal atresia.[1,26] Galindo had single umbilical artery as the most common extra-cardiac abnormality[1]. The association with VACTERL and CHARGE has also been reported. It is not known whether it is a co-incidence or the part of association.[16,26,27]

PLSVC and chromosomal and genetic abnormalities

The overall risk of chromosomal abnormalities is 12.5 %.[24]

The various syndromes associated are VACTERL association (vertebral defects, anal atresia, cardiac defects, tracho-esophageal fistula, renal and limbs abnormalities), CHARGE association (coloboma, heart defects, atresia of choanae, retardation, genital and ear anomalies) and Turner's, Trisomy 21, 18, 22q 11 deletion, deletion in chromosome 7, 8, 13q, 15, Leber optic hereditary neuropathy, Smith Lemli Optiz or Goldenhar syndrome.[1,26,28]

However, in isolated PLSVC, the risk of having a chromosomal abnormality could be as high as 7%.[24] It is still not clear whether invasive testing should be offered in isolated cases or not. Even if, invasive testing is offered, it is better to offer chromosomal microarray as it is the investigation of choice these days. We still do not have enough data whether the investigation of choice should be karyotype or chromosomal micro-array.

PLSVC and arrythmias

It could be cause of arrythmias due to compression of atrioventricular node by enlarged coronary sinus, seen later in life. If found to be trigger of arrythmias, then needs to be ablated.[29]

Prenatal follow up

Isolated PLSVC generally do not cause hemodynamic consequence. Hence, no increase in follow up is required but attention must be made to look for development of disproportionate ventricles. It could be due to right dominance due to drainage of PLSVC into coronary sinus or enlarged coronary sinus obstructing mitral valve or development of CoA. Usually, fortnightly follow up is advised in cases with associated congenital heart defects. However, the exact condition and the progression of congenital heart defects may increase the frequency of follow up.

Delivery

In isolated cases, the delivery should be as per obstetric indications. For isolated cases, the delivery should be in a centre where facilities for neonatal echocardiography or paediatric cardiology is available.

If associated with malformations, then delivery should be in a tertiary care set up where paediatric cardiology and cardiac surgery facilities are available. The timing of delivery should be as far as near to term as possible in such cases.

Postnatal management

All cases with PLSVC should undergo postnatal echocardiography. This is to confirm the findings and rule out any other associated cardiac malformations. The knowledge about presence of PLSVC, even if isolated, is important. The knowledge about its presence is essential as it may be a cause of arrythmias[28] or may complicate cardiac procedures like central venous catheterization, pacemaker implantation or cardiac surgeries.

My experience

In a span of 3 years, from 2019 till date, I have had 12 cases of PLSVC. The incidence of PLSVC in our centre was 0.26%. 6

were isolated with good postnatal outcome. None of the cases had chromosomal nor any other structural – cardiac or extra-cardiac-abnormality. Two cases on follow up developed disproportionate ventricles and none had Coarctation of aorta. Out of six cases, two had normal karyotype and one had low risk non-invasive prenatal screening test.

Rest 6 had major cardiac malformations. The list ranged from atrioventricular septal defect (AVSD), hypoplastic left heart, double outlet right ventricle, mitral stenosis or transposition of great vessel. The most common was AVSD in the series.

Conclusion

Isolated PLSVC is a benign finding with usually an uncomplicated course. Follow up is essential to look for Coarctation of aorta. Its knowledge is important when various cardiac procedures are planned later in life.

Declaration

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Conflict of interest

There was no conflict of interest.

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