Persistent left superior vena cava complicating haemodynamic monitoring

A case report

C. P. ROOS, M. PLIT, J. LIPMAN, J. EIDELMAN, P. R. MARSHALL

Summary

Flow-directed, balloon-tipped catheterization of the right heart via a persistent left superior vena cava is described. The embryonic origin of the superior vena-caval system is briefly reviewed, and the left superior vena cava is discussed with respect to its clinical and surgical importance and methods of diagnosis.

A persistent left superior vena cava is one of the most common venous anomalies, occurring in 0.5% of the general population and in 3-10% of patients with congenital heart disease. It may therefore be an integral part of a more complex congenital cardiac malformation or an isolated anomaly having little haemodynamic significance.

With the steadily increasing use of pulmonary arterial catheterization in the diagnostic evaluation, haemodynamic monitoring and management of the critically ill patient, the clinical significance of this anomalous vein assumes great importance. When unsuspected it may complicate cardiac catheterization and the establishment of cardiopulmonary bypass during open-heart procedures.

A case is presented in which a flow-directed, balloon-tipped Swan-Ganz catheter was inserted via a left superior vena cava. The diagnosis was made from the chest radiograph by noting the characteristic pathway of the catheter.

Case report

In June 1981 a 68-year-old man was admitted to the medical Intensive Care Unit, J. G. Strydom Hospital, Johannesburg, with chest pain and associated features suggestive of acute myocardial infarction. He gave a history of two previous myocardial infarcts, in 1961 and 1965, followed by stable angina.

The diagnosis of acute anterolateral myocardial infarction was confirmed on electrocardiographic tracings and enzyme studies. At the outset the patient was in severe left ventricular failure and complained of persistent chest pain.

Pulmonary capillary wedge pressure was monitored by insertion of a No. 7F Swan-Ganz catheter, introduced percutaneously through the right subclavian vein. The catheter was advanced into the pulmonary artery by means of sequential pressure-pattern monitoring and the position confirmed by a portable chest radiograph (Fig. 1).

Fig. 1. Anteroposterior chest radiograph showing the Swan-Ganz catheter coursing from the right subclavian vein through the superior vena cava, right atrium, and right ventricle and terminating in the left pulmonary artery.

Seventy-two hours after its introduction, the Swan-Ganz catheter was removed because of balloon failure and a similar catheter inserted percutaneously via the left subclavian vein. The continued monitoring was required for the refractory left heart failure and the control of intravenous vasodilator therapy.

The catheter was again advanced under pressure-pattern monitoring, but this time difficulty was experienced in advancing it beyond the 35 cm mark and in obtaining a right ventricular pressure pattern. A portable chest radiograph showed the catheter...
ter traversing a persistent left superior vena cava down to the level of the coronary sinus.

The coronary sinus was catheterized by inflating the balloon with approximately 1 ml air (instead of 1.5 ml air) and representative pressure readings were then obtained from the right atrium, right ventricle, pulmonary artery and pulmonary capillary wedge pressure positions.

A radiograph taken after catheterization (Fig. 2) demonstrated the path of the catheter from the left subclavian vein to a left superior vena cava, through the coronary sinus into the right atrium, right ventricle and into the left pulmonary artery.

Discussion

Embryology2,3 (Figs 3 and 4)

The paired anterior cardinal veins, draining the cephalic capillary beds, and the paired posterior cardinal veins, which are formed by the coalescence of caudal lateral capillary beds anastomose with the umbilical veins and vitelline veins at the posterior end of the developing heart during the 4th week. The anastomoses of the anterior and posterior cardinal veins on either side form the common cardinal veins (ducts of Cuvier) which unite ventrally to open into the sinus venosus of the heart. From the 4th to the 8th week the sino-atrial orifice shifts to the right side of the yet undivided atrium and the ducts of Cuvier become incorporated into the sinus venosus. On the right side the sinus venosus is totally incorporated into the posterior wall of the right atrium. The left duct of Cuvier and the left horn of the sinus venosus atrophy to form the oblique vein (or ligament) of Marshall (ovm) and the coronary sinus (cs). Failure in the obliteration of the left common cardinal vein (ccv) results in a persistent left superior vena cava (plsvc) which drains into the right atrium (ra) via the coronary sinus (cs) (ijv = internal jugular vein; iv = right and left innominate veins; svc = superior vena cava; ivc = inferior vena cava; hiv = highest intercostal vein; av = ayzgos vein; svv = subclavian veins).

The anterior cardinal veins (acv) and posterior cardinal veins (pcv) unite into two short common cardinal veins (ccv), or ducts of Cuvier, which empty into the sinus venosus (sv) of the heart (vv = vitelline veins; uv = umbilical veins; svv = subclavian veins).
The left superior vena cava enters the atrium on its own side. There may or may not be a brachiocephalic vein connecting the two venae cavae. In these primitive cases there are important cardiac defects, usually septation, in addition to the doubling of the vena cava. More frequent than the primitive anomalies described is the pattern in which the anomalous left superior vena cava enters the coronary sinus and drains into the right atrium, as does the normal right superior vena cava. An anastomosing brachiocephalic vein may or may not be present.

**Diagnosis**

A persistent left superior vena cava may be suspected clinically by the presence of abnormal left jugular venous pulsations. The chest radiograph often suggests the diagnosis by demonstrating a widened aortic shadow or vascular pedicle, a definite strip of lessened density along the upper left cardiac border. A paramedial bulge below the aortic arch or a crescentic vascular shadow passing from the left upper border of the aortic arch toward the middle third of the left clavicle.

During catheterization of the right heart or coronary sinus, the catheter may pass into the anomalous vein, with direct angiographic demonstration of a left superior vena cava. Angiography via the left arm vein will demonstrate the anomalous left superior vena cava as opacification along the left paramediastinum and cardiac border on the posterior-anterior chest radiograph and as a vertically descending vein lying in front of the aortic arch and along the posterior border of the heart on lateral chest radiograph. Selective catheterization of the coronary sinus will also demonstrate a persistent left superior vena cava. Owen and Urquhart describe a notch in the inferior border of the left atrium (due to a dilated coronary sinus receiving blood directly from a persistent left superior vena cava) with left ventricular angiography.

Echocardiographic studies of a persistent left superior vena cava reveal a large, circular, echo-free space behind the posterior left ventricular wall, representing a dilated coronary sinus through which the anomalous vein drains. The injection of contrast medium via a left arm vein obliterates this echo-free space, thus differentiating it from a pericardial effusion. Cross-sectional echocardiography reveals the persistent left superior vena cava as a narrow tubular echo cavity posterior to the left atrium and left ventricle (corresponding to the left atrioventricular sulcus) which fluctuates in size during systole and diastole.

Table I summarizes the criteria for diagnosis of a persistent left superior vena cava.

**Clinical significance**

The incidence of left superior vena cava varies from 0.5% in subjects with otherwise normal hearts to between 3% and 10% in patients with congenital heart disease. The clinical significance of the left superior vena cava is that it is a pointer to associated anomalies, the most common being atrial septal defect with anomalous pulmonary venous drainage.

Recognition of a persistent left superior vena cava draining into the right atrium, although usually having no haemodynamic significance, may be important for the correct interpretation of cardiac catheterization, angiographic, radiological and echocardiographic findings.

An anomalous vein may complicate the insertion of transvenous pacemaker electrode catheters as well as the catheterization of the pulmonary artery. This is due to the difficulty in negotiating the coronary sinus and in manoeuvring the catheter into the right ventricle as a result of the unfavourable direction of the emerging catheter from the coronary sinus. This applies particularly with the use of the non-flow-directed stiff-walled catheters and pacemaker electrodes.

Catheterization of the coronary sinus has been associated with chest pain, electrocardiographic changes consistent with myocardial ischaemia, cardiorespiratory arrest due to vagal stimulation by the catheter in the left superior vena cava, and electrical instability and arrhythmias. Pre-operative diagnosis of a persistent left superior vena cava in patients undergoing open-heart surgery with cardiopulmonary bypass necessitates certain technical procedures in order to prevent excessive blood loss due to troublesome inflow of blood into the right atrium.

In summary, a relatively uncommon cardiac anomaly, a left superior vena cava, was found during Swan-Ganz catheterization. Intensive-care personnel utilizing pulmonary arterial catheter monitoring of critically ill patients should be aware of this anatomical variant and its associated lesions, and of its clinical significance.

**REFERENCES**


**Table I. Diagnosis of a Left Superior Vena Cava**

1. Clinical inspection
2. Abnormal left jugular pulsation
3. Plain chest radiograph
4. Left paramediastinal strip or crescent
5. Widening of the vascular pedicle
6. Aortic bulge
7. Position of catheter in the left superior vena cava on chest radiograph
8. Angiocardiography
9. Left arm veins
10. Catheterization of coronary sinus
11. Left atrial notch sign on left ventricular angiography
12. Two-dimensional echocardiography
13. Contrast echocardiography
14. Cross-sectional echocardiography
15. Surgery
16. Autopsy