

CLINICAL IMAGE

Pulmonary artery catheter following an aberrant course

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Case history

A 43-year-old woman was admitted with dyspnoea, a cough and peripheral oedema, rapidly followed by respiratory insufficiency necessitating mechanical ventilation. Bilateral pulmonary embolism and chronic thrombo-embolic pulmonary hypertension were diagnosed. After sufficient anticoagulant therapy the patient could not be weaned from mechanical ventilation. Because of the difficult weaning process the patient was referred to the Intensive Care Unit (ICU) at our university hospital. To rule out any cardiac causes that could be responsible for the difficult weaning process, a pulmonary artery catheter (PAC) was introduced through the right internal jugular vein. After several attempts, right ventricular curves and, finally, pulmonary artery curves were obtained. Apart from mild pulmonary hypertension to be explained by the existence of pulmonary emboli, no additional cardiac abnormalities were found. However, her chest X-ray, showed the pulmonary artery catheter to be following an aberrant course (Figure 1).

Repeat echocardiography was performed which showed normal left and right ventricular function with moderate tricuspid valve regurgitation. A wide coronary sinus was found, endorsing the presumed diagnosis of a persistent left superior vena cava (PLSVC). Additional CT-scanning with intravenous contrast confirmed the diagnosis PLSVC, draining into the right atrium via a wide coronary sinus. The right superior vena cava was absent (Figure 2). No other cardiac anomalies were found.

Background

PLSVC is the most common venous anomaly in humans, occurring in 0.5% of the normal population and in 0.47% of patients undergoing implantation of a pacemaker or cardioverter defibrillator (ICD) [1]. A frequency of 5 to 10% has been reported among patients with congenital heart defects. The right superior vena cava is absent in only 1% of patients with PLSVC [2,3].

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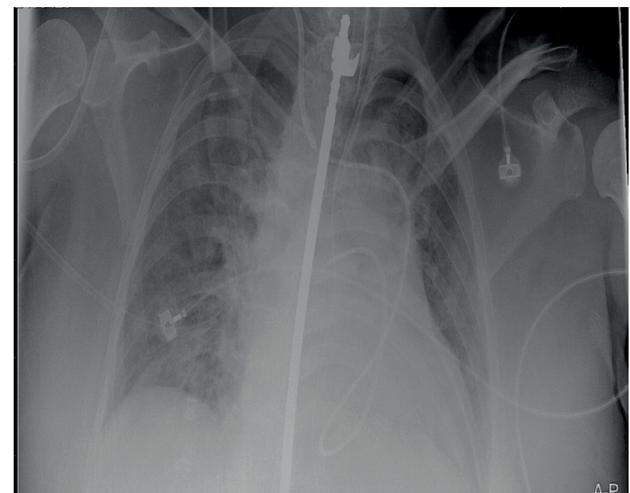
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(Figure 3) In a small percentage of cases, the left superior vena cava drains into the left atrium; this is usually associated with an atrial septal defect. Usually PLSVC drains into the right atrium via the coronary sinus which is wide. Associated cardiac anomalies are diverse e.g. atrial septal defect, ventricular septal defect, tetralogy of Fallot, situs inversus and coarctatio aorta. Extra-cardiac anomalies include VACTERL*-association and CHARGE^-association [4]. These combinations of anomalies might be explained by disorders in the development of the systemic veins starting at 4 weeks gestation, as discussed in the embryology and pathophysiology of this venous anomaly [5].

Conclusion

While right heart catheterization or pacemaker placement is performed frequently in ICU patients, the intensivist may be confronted with PLSVC, frustrating proper positioning. In cases

Figure 1. Pulmonary artery catheter following an aberrant course



of PLSVC, right heart catheterization or pacemaker placement may alternatively be performed via the inferior vena cava, guided by fluoroscopy or echocardiography .

*VACTERL = vertebral defects, anal atresia, cardiac malformations, tracheo-esophageal fistula with esophageal atresia, radial and renal dysplasia and limb abnormalities;
 ^CHARGE = coloboma, heart defects, atresia of choanae, retardation, genital and ear abnormalities.

Figure 2 Angiography showing a persistent left superior vena cava (PLSVC), draining into the coronary sinus. The right superior vena cava is absent.

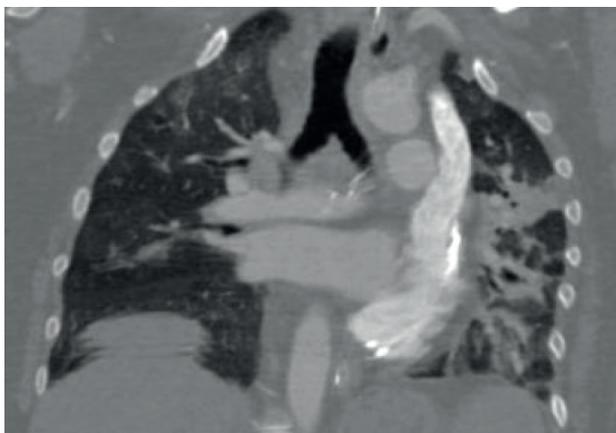
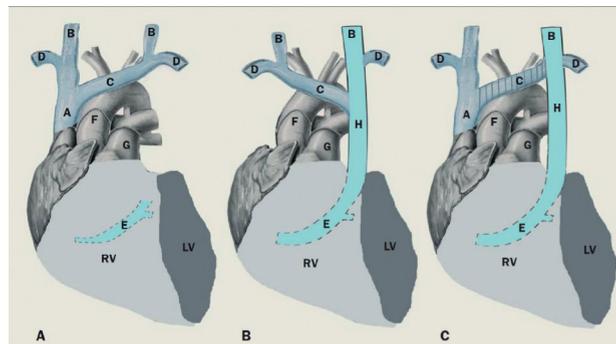


Figure 3. (from Schreve-Steensma et al. Neth Heart J 2008; 16: 272-274.(ref .3), reprinted with permission from the authors): Schematic drawing of the anatomical variations of the superior vena cava. 3A) The normal situation. 3B) Single left superior vena cava. This is the situation of the presented patient. 3C) Doubled superior vena cava. The right superior vena cava is either connected to the left by the left brachiocephalic vein or is without any connection.



A = superior vena cava; B = internal jugular vein; C = left brachiocephalic vein; D = subclavian vein; E = coronary sinus; F = aorta; G = main pulmonary artery; H = left superior vena cava; RV = right ventricle; LV = left ventricle.

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