

Total anomalous pulmonary venous drainage to persistent left superior vena cava: a unique configuration

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DESCRIPTION

A 1-year-old child underwent cardiac CT angiography (CTA) for further characterisation of a complex congenital cardiac defect. Review of CTA multiplanar reconstructed and volume-rendered images revealed thoracoatrial right isomerism with the presence of bilateral superior vena cavae (SVC) and an intercommunicating vein. The right superior pulmonary vein, right inferior pulmonary vein and left inferior pulmonary vein formed a common channel (*) which received the left superior pulmonary vein just before draining into the left superior vena cava (LSVC) (figure 1A–D). The LSVC was seen to directly drain into the left atrium with the absence of coronary sinus (figure 1E,F). The other concomitant structural defects included the presence of a double-outlet right ventricle, ventricular septal defect and pulmonary stenosis.

Embryologically, the pulmonary venous plexus (which surrounds the lung bud and is contiguous with the splanchnic venous plexus) forms a connection with the pulmonary veins which arise from the dorsal wall of sinus venosus and in this process, its connections with the cardinal and umbilicovitelline veins disappear. Abnormal

persistence of these communications gives rise to partial or total anomalous pulmonary venous connection (TAPVC).¹ If these connections coexist along with abnormal persistence of the left precardinal vein, an extremely rare configuration of anomalous (total or partial) pulmonary venous drainage into the persistent LSVC may arise. Though various patterns of communication of pulmonary veins and persistent LSVC have been explained embryologically and classified by Snellen *et al*, there are only anecdotal reports demonstrating this anomalous drainage pattern on cardiac CTA.² Although Maki *et al* recently described a case of partial anomalous pulmonary venous drainage into persistent LSVC on cardiac CTA, to the best of our knowledge, TAPVC into the LSVC on CTA is hitherto unreported.³ Also, even though TAPVC is the most common type of pulmonary venous drainage anomaly associated with right isomerism, its connection with the persistent LSVC is only rarely reported in the literature.

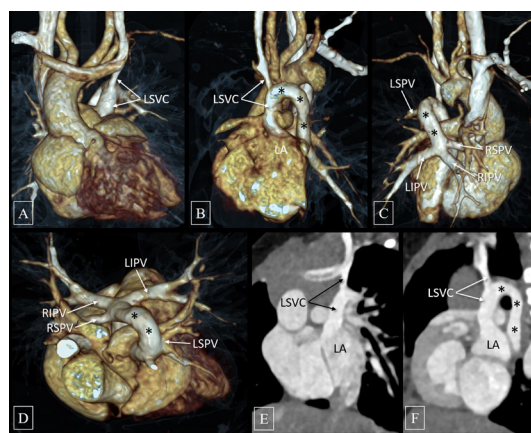


Figure 1 Volume rendered images (A–D) depict the RSPV, RIPV and LIPV forming a common channel (*) which receives the (LSPV just before draining into the LSVC. Maximum intensity projection images in the oblique coronal (E) and oblique sagittal (F) planes demonstrate the LSVC draining directly into the LA. LA, left atrium; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; LSVC, left superior vena cava; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein

Learning points

- ▶ Abnormal persistence of embryological connections between the pulmonary venous plexus and the cardinal and umbilicovitelline veins give rise to partial or total anomalous pulmonary venous connections.
- ▶ Although total anomalous pulmonary venous connection is the most common type of pulmonary venous drainage anomaly associated with right isomerism, its connection with the persistent left superior vena cava is rare.

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