

Left superior vena cava: revisited

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The persistence of a left-sided superior vena cava is the most common variant of systemic venous drainage. Increased utility of cardiac imaging, in particular cross-sectional techniques such as computed tomography and magnetic resonance (MR), will result in increased detection of the anomaly and its variants. Whilst in the typical form it is often haemodynamically insignificant, its discovery may have clinical significance nonetheless, and its mimics require exclusion. During cardiac development the anomaly results from a failure of the left anterior cardinal vein to obliterate. Recognized anatomical variants include the absence of the right superior vena cava and of an innominate bridging vein. Typical drainage is to the coronary sinus, dilatation of which may be the first hint to the anomaly. Clinical implications with respect to vascular access and arrhythmia are well described. A significant minority drain into the left atrium, potentially creating a haemodynamically significant lesion. Additionally, differentiation from anomalous left upper pulmonary venous drainage via a vertical vein is mandatory. A newly discovered variant runs an intra-atrial course with subsequent typical drainage, and if not recognized as such, may be confused with a left atrial mass. The use of 3D contrast-enhanced MR venography has proven extremely helpful in characterizing anomalous vasculature, and we demonstrate how such techniques can help delineate the anomaly and differentiate from its mimics.

Keywords

Persistent left superior vena cava • Intra-atrial course • Magnetic resonance venography

Introduction

A persistent left-sided superior vena cava (LSVC) is the most common variant of systemic venous drainage. It is present in ~0.5% of the general population, and up to 10% of those with established congenital heart disease.¹ It results from a failure of obliteration of the left common cardinal vein, and typically drains the left subclavian and jugular veins into the right atrium via the coronary sinus (CS). It may be identified incidentally by echocardiography, often indirectly through recognition of a dilated CS (Figure 1). On cross-sectional imaging with computed tomography (CT) or magnetic resonance (MR), it is apparent as a vessel coursing vertically in the mediastinum, lateral to the aortic arch. In the majority of individuals with this anomaly, a right superior vena cava is present, although this may be smaller than usual and the left innominate vein is typically absent or small. There is an increased likelihood of congenital heart disease if there is no right-sided superior vena cava or if the LSVC drains into the left atrium (LA).

Typically it is regarded as an incidental anomaly with little functional importance, although it may be significant if it is unilateral or drains into the LA, particularly as the incidence of congenital cardiac anomalies is increased.

An LSVC should be carefully differentiated from another relatively common congenital venous abnormality, that of partial anomalous pulmonary venous drainage of the left upper lobe.

This latter condition has a similar appearance on cross-sectional imaging with an anomalous vertical vein coursing in the mediastinum lateral to the aortic arch. This drains the left upper lobe pulmonary veins into the left innominate vein.

Embryological origins of LSVC

During development of the right atrium, the sinu-atrial chamber differentiates into the sinus venosus and the atrial chamber. The right and left anterior cardinal veins drain the cranial regions into the respective ducts of Cuvier, which in turn drain into the sinus venosus. The sinus venosus is eventually absorbed into the structure of the right atrium, with the bridging connection between the anterior cardinal veins forming the left innominate vein. The right cardinal vein and right duct of Cuvier form the SVC, whereas the left cardinal vein typically obliterates. If occlusion of the left cardinal vein fails to occur, then the vessel 'persists' as a left-sided SVC, typically draining into the CS via the vein of Marshall² (Figure 2).

Anatomical considerations

LSVC of 82–90% are duplicate SVC, with a co-existent right-sided vessel^{2–4} (Figure 3). In duplicate SVC, the prevalence of an innominate bridging vein is 30%,⁵ although this vessel may be small.⁶

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Figure 1 Echocardiogram, parasternal view in left-sided superior vena cava. A typical dilated coronary sinus (CS) is apparent.

When the isolated LSVC is encountered, dextrocardia or complete situs inversus may be present.

Typically the LSVC descends vertically, anterior, and to the left of the aortic arch and main pulmonary artery. It runs adjacent to the LA before turning medially, piercing the pericardium to run in the posterior atrio-ventricular groove.² About 90% of cases drain into the CS; alternative sites include the inferior vena cava, hepatic vein and LA.

Left atrial drainage is almost always associated with other congenital abnormalities.⁷ The LSVC enters the LA between the pulmonary veins and the atrial appendage,⁸ and is invariably associated with an atrial septal defect in published series.^{4,8}

Clinical relevance of LSVC

Although an LSVC draining into the CS has no adverse haemodynamic effects, it may impact on procedures which require upper limb venous access. Challenges and complications have been widely reported during transvenous pacing for bradycardia^{9–11} and advanced device implantation.^{12–14} Central venous cannulation may result in unusual catheter positions,⁵ and inadvertent CS cannulation may result in cardiac perforation.¹⁵ Cannulation of the heart for cardiopulmonary bypass may result in ineffective retrograde cardioplegia.¹⁶

LSVC has been associated with an increased risk of arrhythmias, most commonly atrial fibrillation.⁵ Recent studies highlighting the arrhythmogenic potential of the ligament of Marshall may be relevant, as this structure contains the vein of Marshall, whose muscular sleeves continue into the CS.¹⁷

The presence of LSVC should prompt the clinician to search for other congenital cardiac defects. Associations include atrial and ventricular septal defects, endocardial cushion defects, tetralogy of Fallot, CS ostial atresia, and cor triatriatum.^{5,18,19} Atypical LSVC drainage into the LA results in a right to left shunt. This scenario may cause cyanosis,^{20,21} and has been implicated in cases of paradoxical thromboembolism,²² air and septic embolism.^{23,24}

Case studies

Case 1

A 19-year-old male with a recurrent atrial arrhythmia underwent transthoracic echocardiography, which reported a left atrial mass. Cardiovascular magnetic resonance (CMR) was performed to delineate left atrial anatomy and pulmonary venous drainage. An LSVC was identified on axial fast spin echo [Half-Fourier Acquisition Single Shot Turbo Spin Echo (HASTE)] imaging (Figure 4).

Steady-state free precession (SSFP) cine sequences revealed unusual appearances (Figure 5). In the two-chamber view, an apparent band was seen traversing the LA, raising the possibility of cor triatriatum. A vertical vessel was visualized entering the LA cranio-caudally behind the left atrial appendage, initially presumed secondary to partial volume effects. The CS was dilated, consistent with typical LSVC drainage. The possibility of an intra-atrial course of the LSVC was then considered. The four-chamber view revealed appearances consistent with the description of a cystic mass adjacent to the Coumadin ridge.

However, the structure exhibited features of slow flow that was apparently perpendicular to the predominant LA flow direction, accounting for the distinct signal difference within the structure.

Transoesophageal echocardiography (TOE) subsequently confirmed the CMR findings. In short axis, the vessel mimicked a cystic mass. Bubble contrast injection from the left arm using orthogonal views conclusively revealed the true nature and connections of the vessel (Figure 6).

The LSVC in this case takes a highly unusual intra-atrial course along the medial border of the Coumadin ridge. Rather than coursing lateral to the LA, this vessel enters the chamber via its roof. The anatomical point of entry is exactly as described when an LSVC drains into the LA,⁸ however no left atrial communication was seen, and no atrial septal defect was present.

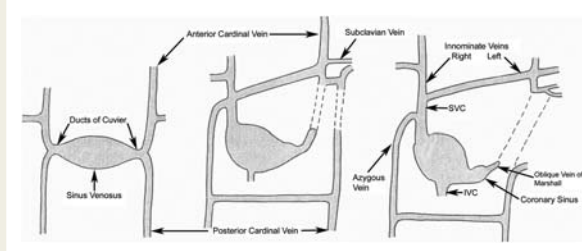


Figure 2 Embryological development of left-sided superior vena cava (SVC). Initially, the anterior cardinal vein drains into the primitive sinu-atrial chamber (left image). The left cardinal vein later obliterates (centre image), leaving only the oblique vein of Marshall as a remnant (right image). The failure of obliteration leads to a 'persistent' left-sided superior vena cava (adapted from Campbell and Deuchar⁴).

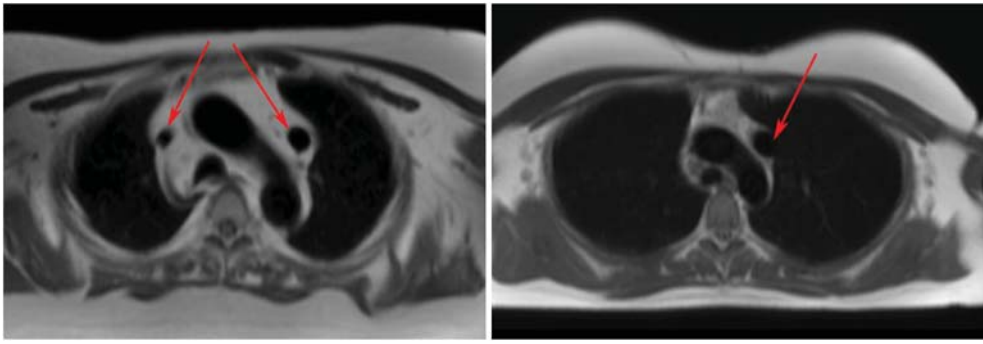


Figure 3 Transaxial turbo spin echo (HASTE) magnetic resonance images at the level of the aortic arch. The left image demonstrates bilateral superior vena cava (arrowed). The right-sided superior vena cava is smaller than usual, and the left innominate vein absent. The right image demonstrates a rare isolated left-sided superior vena cava.

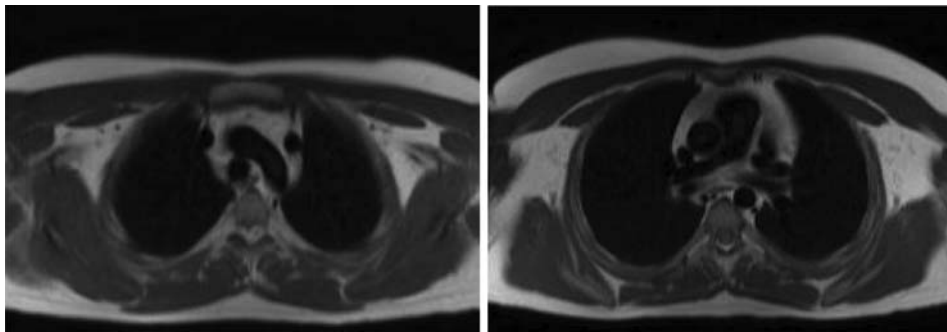


Figure 4 Transaxial HASTE images. An anomalous vessel is seen in a typical left-sided superior vena cava position.

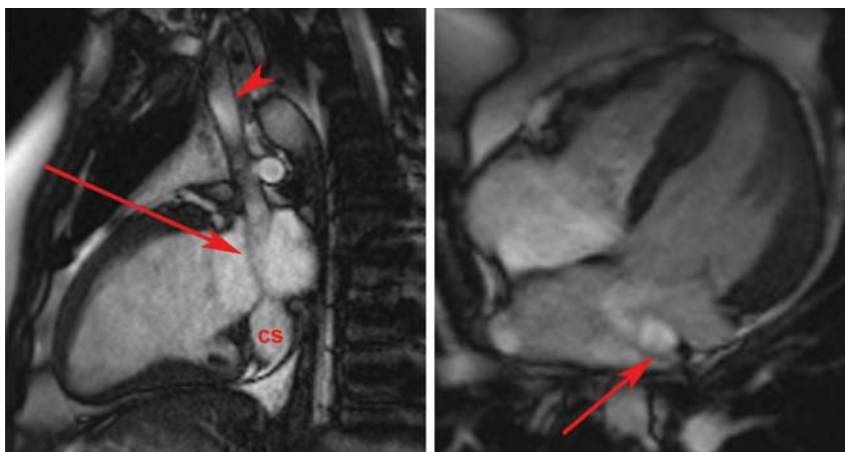


Figure 5 Steady-state free precession imaging in long-axis planes. Two-chamber view (left image) revealed a dilated coronary sinus (CS), an unusual band in the left atrium (red arrow), and a vertical vessel above the left atrium (red arrowhead). In the orthogonal four-chamber view (right image), this vessel is identified running within the left atrium (red arrow), near the Coumadin ridge.

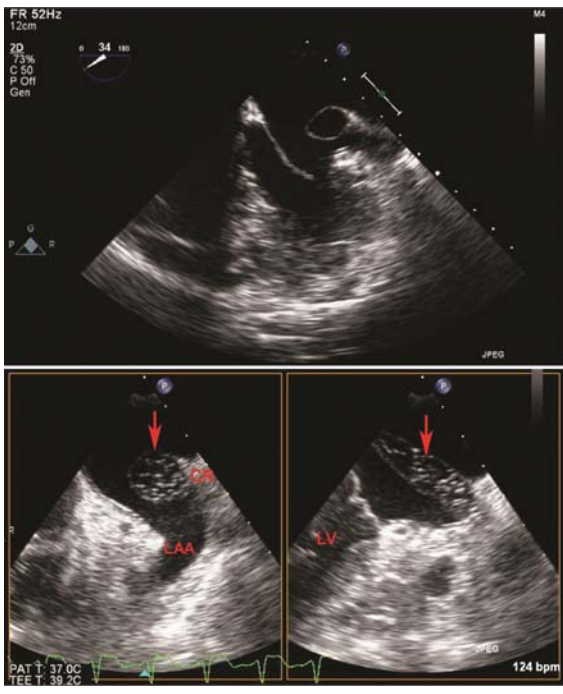


Figure 6 Transoesophageal echocardiography. Top image: a cyst-like structure is evident. Lower left: bubble contrast injection from the left arm. Left image: the vessel lies in continuity with the Coumadin ridge (CR), posterior to the left atrial appendage (LAA). Right image: in the orthogonal view, the vessel is seen to traverse the left atrium (LV, left ventricle), with bubble contrast passing inferiorly (right to left in this image).

Case 2

A 22-year-old male underwent CMR following the repair of a double outlet right ventricle with residual VSD. Axial HASTE imaging revealed a small anomalous vessel in a typical LSVC position. A normal right SVC and innominate vein were evident, and the CS was not dilated. A 3D contrast-enhanced (CE)-MR venography (MRV) sequence demonstrated the intra-thoracic course, and SSFP sequences in a four-chamber stack identified the vessel at the medial edge of a prominent Coumadin ridge (Figure 7). Subsequent transthoracic echocardiography (TTE) with bubble-contrast injection from a left antecubital vein was performed. The anomalous vessel was noted coursing vertically through the LA in the two-chamber view (Figure 7). Opacification of the vessel was demonstrated in the four-chamber view following bubble contrast, proving an LSVC with intra-atrial course (Figure 8).

This case demonstrates a vessel with almost identical anatomical course to Case 1, albeit of a lesser calibre. In both cases ultimate drainage was to the right atrium via the CS (characteristically absent in left atrial drainage). With increasing utility of advanced cardiac imaging, in particular cross-sectional techniques, this abnormality may become increasingly recognized. Unless recognized, such appearances may result in misdiagnosis or lead to further potentially unnecessary investigation.

Case 3

A 43-year-old male with dyspnoea was referred for CMR imaging. TTE had revealed dilated right heart chambers, but no atrial septal defect. The patient was referred primarily to exclude a sinus venosus defect. Routine axial HASTE sequences showed an anomalous vessel arising from the junction of the left innominate and left subclavian veins; the appearance was consistent with a persistent LSVC (Figure 9).

SSFP cine sequences were used to interrogate the intra-atrial septum, with no evidence of an intra-atrial defect. Further SSFP sequences were used to delineate the pulmonary veins, all of which except the left upper were clearly identified (Figure 10). Right ventricular volumes were raised (end-diastolic volume of 102 mL/m²).

A 3D CE-MR angiography (CE-MRA) sequence was performed with a gadolinium-based contrast agent (Magnevist, Bayer, Berlin, Germany). Following left antecubital fossa venous injection, two phases were acquired. The arterial phase acquisition revealed the diagnosis of partial anomalous pulmonary venous drainage (PAPVD) via a 'vertical vein' (Figure 11).

It is clear how PAPVD of the left upper lobe can mimic LSVC on cross-sectional imaging, so care is required to make the correct diagnosis. In the former, blood drains cranially via a 'vertical vein' into the left innominate vein. The resulting haemodynamic effect is a left to right shunt above the atrio-ventricular valves. Consequent right heart dilatation is a useful clue to the diagnosis, and such a finding should prompt a detailed search for the rarer cases where multiple anomalous connections lead to clinically significant shunts.

Other differentiating imaging features are worthy of mention. LSVC manifests as two vessels anterior to the left main bronchus on axial imaging, whereas in PAPVD there will be no aberrant vessel or superior pulmonary vein evident⁶ (see Figure 12). Absence of an innominate vein is inconsistent with left upper lobe APVD, although the presence of this vessel is not a helpful discriminator. The presence of a dilated CS is often a marker of LSVC, although one should not forget that other possibilities exist, and importantly it does not discriminate between SVC and APVD.²⁰

The differential diagnosis of a dilated CS includes partial anomalous hepatic venous drainage, or continuity of the IVC with an LSVC via the hemiazygos vein. Various shunts may result in CS dilatation, either low pressure in the case of unroofed CS and APVD, or high pressure in the form of a coronary artery to CS fistula.²⁵ Finally any cause of raised right atrial pressure may cause dilatation.

Imaging of the central veins with MRA

CE-MRA combines a gadolinium-based contrast agent with a gradient echo sequence to create 'bright-blood' images of a vessel lumen. This can be extremely helpful in characterizing unusual vasculature, however certain technical factors are worthy of note. Because of susceptibility-induced signal drop off caused by a compact contrast bolus, first pass imaging of the central thoracic veins is often poor. Alternative strategies such as dilution of

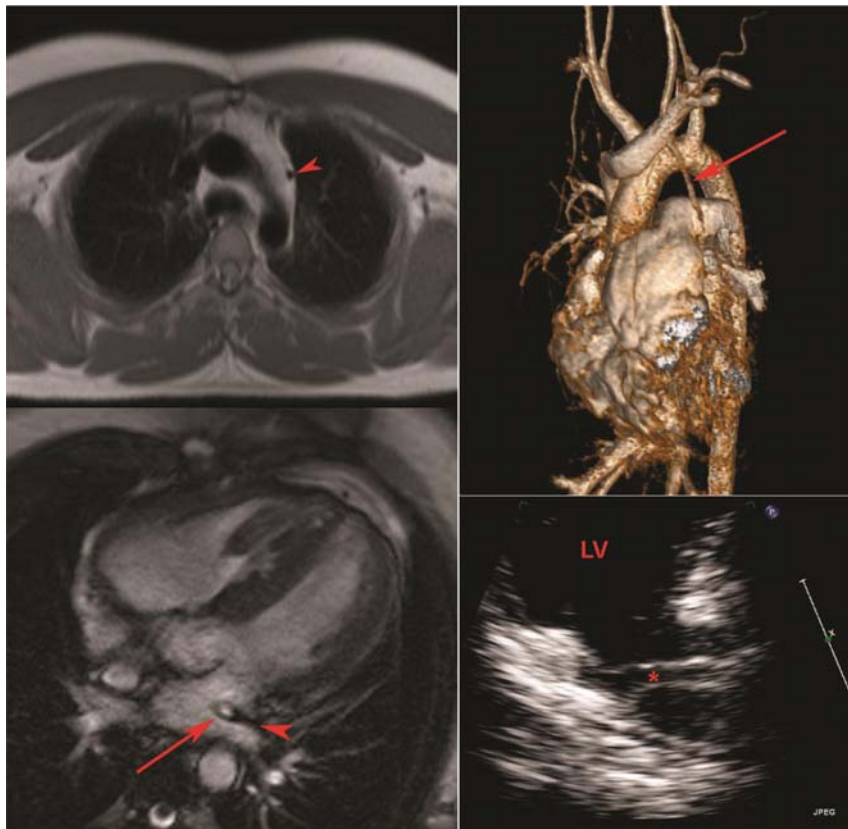


Figure 7 Top left: transaxial HASTE image, an anomalous vessel seen in left-sided superior vena cava position (arrowhead). Top right: contrast-enhanced magnetic resonance venography with the vessel highlighted (red arrow). Bottom left: still frame of steady-state free precession cine sequence from a four-chamber stack. The vessel is noted passing through the left atrium (LA, red arrow) adjacent to a prominent Coumadin ridge (arrowhead). Bottom right: transthoracic echocardiography 'two-chamber' image of the vessel (*) traversing the left atrium from cranial to caudal.

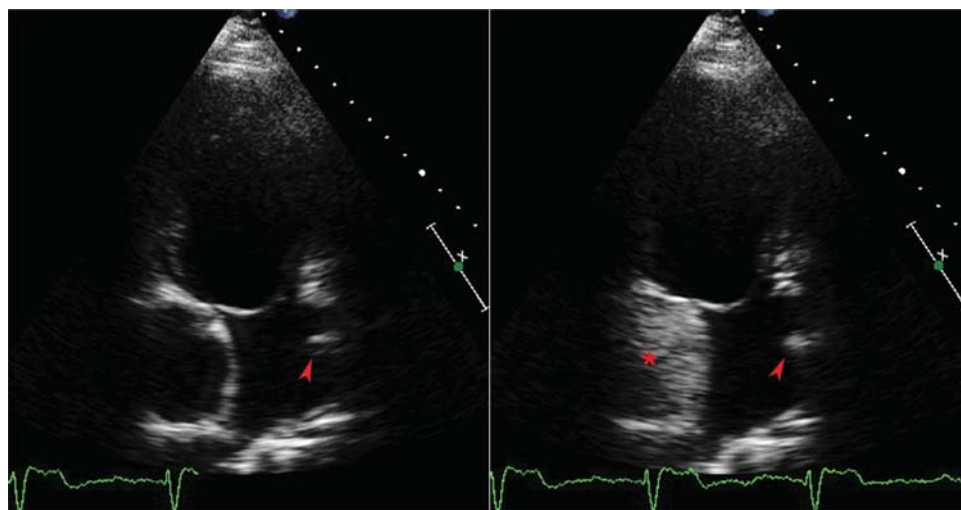


Figure 8 Transthoracic echocardiography with bubble contrast. Note opacification of right atrium (*) and anomalous left-sided superior vena cava (arrowhead) following injection from a left antecubital fossa vein.

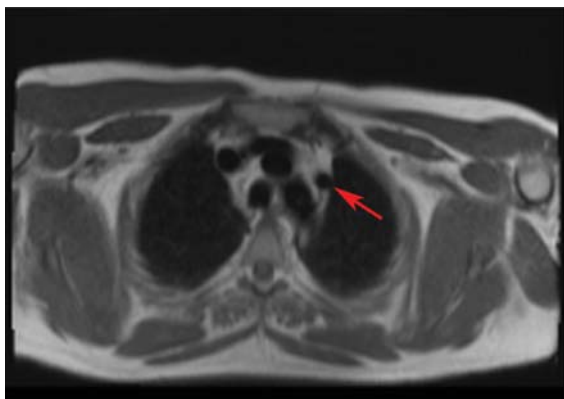


Figure 9 Trans-axial HASTE showing an anomalous vessel (red arrow).

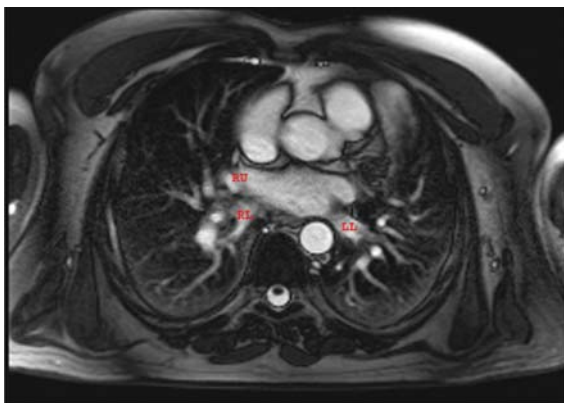


Figure 10 Steady-state free precession sequence showing pulmonary venous drainage. The left upper pulmonary vein could not be clearly identified. RU, right upper, RL, right lower, LL, left lower.

contrast, slow, and prolonged injection, second pass or recirculation imaging must be applied in order to image such vessels.^{26–28} Timing the acquisition of central k-space according to the expected pathology is paramount. In practice it is recommended to acquire both arterial and venous phases if abnormal vascular arrangements are suspected.

Two clinically competing MR protocols should be considered for central venous imaging. Firstly 3D CE-MRV, and secondly a time-resolved 2D MRV approach. Whilst the former generally produces crisper anatomic images with a better spatial resolution, the latter technique has other advantages.²⁸ 2D MRV is almost identical in philosophy to an X-ray venogram following injected iodinated contrast. Beyond the important issue of obtaining temporal data with this sequence, advantages include the elimination of the need for bolus tracking and post-processing, and the ability to use lower contrast doses (e.g. <1 mL vs. >3 mLs at 1.5T). Moreover, scan time is shorter, making the technique more suitable for patients with



Figure 11 Arterial phase magnetic resonance angiography showing the anomalous vessel (vertical vein) draining into the venous system (blue arrow). In, innominate vein; SVC, (right) superior vena cava.

limited breath hold ability.²⁹ The ultimate decision which technology to apply will be driven by the clinical question to be answered. For example, assessment of venous anatomy prior to pacemaker implant in a patient with suspected subclavian vein thrombosis/stenosis may be best performed as a 3D CE-MRV, whilst the patient with upper limb lymphoedema may benefit from the temporal information obtained by a time-resolved 2D MRV.

MR in comparison to other imaging modalities

MR has a number of attributes which make it an ideal modality to image pulmonary and systemic venous anomalies. The ability to encompass a large field of view and image extra-cardiac anatomy is a key strength. The ability to scan in any plane is a further advantage of CMR over existing echocardiographic and catheterization-based techniques. TTE is additionally limited by acoustic windows, and TOE represents a semi-invasive technique. CT angiography allows 3D volume rendering of vascular abnormalities, and similar to CMR it affords a large field of view and multiple-plane analysis. It is potentially limited by the need for iodinated contrast and the need for ionizing radiation. Particularly in the younger patient, or those having multiple examinations, MRA may be favoured.

Gadolinium-enhanced 3D MRA sequences have been an important advance for thoracic and cardiac MR. The ability to accurately and non-invasively image pulmonary and systemic venous anomalies has been proved.³⁰ Using 3D MRA techniques, Prasad *et al.*³¹ found 23% of PAPVD diagnosed by CMR were undetected on either echocardiography, or invasive cardiac catheterization.

As we have demonstrated, supplementary findings may guide the clinician to the true nature of an anomalous venous structure. To help exclude PAPVD, CMR allows highly accurate shunt quantification using phase-contrast velocity mapping sequences.³² Additionally, it provides the gold standard in measuring right ventricular volumes.³³ CMR has an established role in the evaluation of congenital heart disease, allowing a comprehensive assessment of

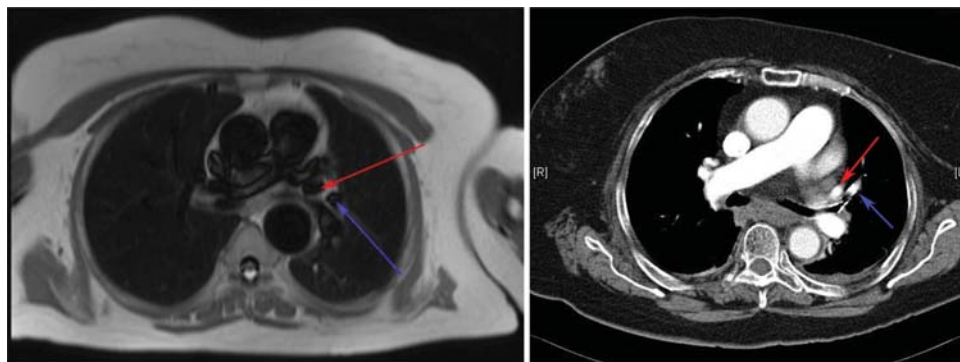


Figure 12 Axial HASTE cardiovascular magnetic resonance (left image) with computed tomography at a corresponding level (right image). Two vessels are visible anterior to the left main bronchus, left-sided superior vena cava (red arrow) and left upper pulmonary vein (blue arrow) on both images.

anatomy and function. The incidental finding of LSCV should prompt the search for associated anomalies, and CMR is ideally suited to achieving this in a single examination.

Conclusion

Anomalies of the superior vena cava are frequently identified as incidental findings on cross-sectional imaging and echocardiography, and are occasionally associated with important clinical sequelae. Cardiac imaging specialists should be aware of their imaging features and associations, and be able to differentiate the persistent left-sided SVC from partial anomalous pulmonary venous drainage of the left upper lobe. If the latter is present and the right heart dilatation/strain is evident, additional right-sided venous drainage abnormalities should be actively searched for and excluded. MRA techniques are extremely useful in depicting anomalous venous anatomy, although certain technical considerations should be noted. A rare variant of LSCV with an intra-atrial course has been identified, and may become increasingly reported as the use of advanced cardiac imaging techniques becomes more widespread.

Conflict of interest: none declared.

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IMAGE FOCUS

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Idiopathic dilatation of the right atrium

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A 66-year-old woman was referred to our institution suffering from progressive dyspnoea and asthenia. Her medical history included palpitations without cardiac investigation. She presented clinical manifestations of right-sided heart failure. Auscultation revealed a systolic murmur of tricuspid regurgitation. EKG recorded sinus rhythm with a complete right bundle branch block. Chest radiography showed a cardiac enlargement (cardiothoracic index of 0.74; Panel A). Echocardiogram demonstrated a highly enlarged right atrium, a functional tricuspid regurgitation, and a dilated right ventricle. The left cavities were compressed by the right cavities (Panel B). Systolic pulmonary artery pressure was estimated at 45 mmHg. Agitated saline injection did not show atrial shunt. Idiopathic dilatation of the right atrium (IDRA) was diagnosed. Magnetic resonance imaging confirmed the echocardiographic findings and the volume of the right atrium was measured at 700 mL (Panel C). The patient refused further extensive examinations. She was discharged with diuretics.

IDRA must be evoked on the finding of a disproportionately enlarged right atrium in the absence of other cardiac or haemodynamic abnormalities and must be distinguished from other anomalies such as right atrial diverticula or Ebstein's anomaly. Right atrial enlargement may be associated with tricuspid annular dilation responsible for functional regurgitation, which may worsen the right atrium dilation and led to right ventricular dysfunction. IDRA has been reported to occur at all stages of life and is often diagnosed fortuitously. Atrial fibrillation, systemic embolism, and heart failure are the classical complications that can be managed medically in most of the cases. Surgical resection of the right atrium may be proposed in patients with incessant arrhythmia, congestive heart failure, and left ventricular compression that might cause sudden cardiac death.

Panel A. Chest radiography: cardiac enlargement with a cardiothoracic index of 0.74 and slight congestion of the bilateral lung fields.

Panel B. An enlarged right atrium with severe functional tricuspid regurgitation, dilated hypokinetic right ventricle, and compression of the left cavities by the right cavities.

Panel C. Magnetic resonance imaging: a highly enlarged right atrium with a volume measured at 700 mL.

