

# Cardioverter defibrillator implantation in a patient with absent right superior vena cava

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**Left-sided superior vena cava (LSVC) is the most common venous thoracic anomaly. Absence of the right superior vena cava (RSVC) on the other hand is very rare. We describe a patient with this abnormal venous system, who was admitted to our centre for an implantation of a cardioverter defibrillator (ICD). (*Neth Heart J* 2006;14:255-7.)**

Keywords: superior vena cava (left), superior vena cava (right, absent), anomaly, thorax (upper), cardioverter defibrillator (implantable)

**A** 53-year-old man was admitted with crescendo angina pectoris. The electrocardiogram showed a short PR interval, diffuse T wave inversions and deep Q waves in V<sub>1</sub> and V<sub>2</sub> consistent with an old anterior wall myocardial infarction (figure 1).

Echocardiography showed poor left ventricular function (left ventricular ejection fraction 25%) and absent anterior wall movement. Coronary angiography was performed and revealed an occluded proximal left anterior descending coronary artery and a significant stenosis of the right coronary artery for which a percutaneous transluminal coronary angioplasty (PTCA) procedure with coronary stent implantation was performed. A week later he was re-admitted for prophylactic insertion of an implantable cardioverter

defibrillator (ICD). Because the patient was left handed, the approach was made through the right subclavian vein. Because the inserted lead had an abnormal course during fluoroscopy, a venogram was performed. The venogram revealed that the route to the entrance of the right atrium was via an LSVC. The contrast material failed to show the presence of a RSVC; it was therefore concluded that the RSVC was, in fact, absent (figure 2).

The ICD lead was inserted with active tip fixation in the right ventricular apex. The patient also had sinus node dysfunction with episodes of AV-junctional rhythm. Therefore an additional right atrial lead was implanted in the lateral wall of the right atrium for AAI pacing. The defibrillation threshold of the ICD was tested. Ventricular fibrillation was induced twice and successfully terminated with 14 Joule. A chest X-ray the following day revealed the remarkable position of the leads on the left side of the mediastinum (figure 3). At two-month follow-up, stable right ventricular pacing and sensing parameters were seen.

## Discussion

Many case reports have been published describing an LSVC, often diagnosed incidentally with trans-venous procedures such as insertion of pulmonary pressure wires, pacemaker leads or dialysis lines. Although a rare finding, an LSVC is the most common anomaly of the venous system with an incidence of 0.3% in the healthy general population and 4.3% in patients with congenital heart disease.<sup>1</sup> Absence of the RSVC is much rarer, with an incidence of 0.07 to 0.13%. In 92% of cases an LSVC drains via the coronary sinus into the right atrium. In 7 to 8%, however, the LSVC drains into the left atrium; in this condition the coronary sinus may or may not be absent. As a right-left shunt is created there is an increased risk of systemic embolisation of air or thrombus.<sup>1</sup> Most commonly associated malformations are atrial septal defects (16%), endocardial cushion defects (11%) and Fallot's tetralogy (9%).<sup>2</sup> Early in embryonic development, the common cardinal veins, which become the superior

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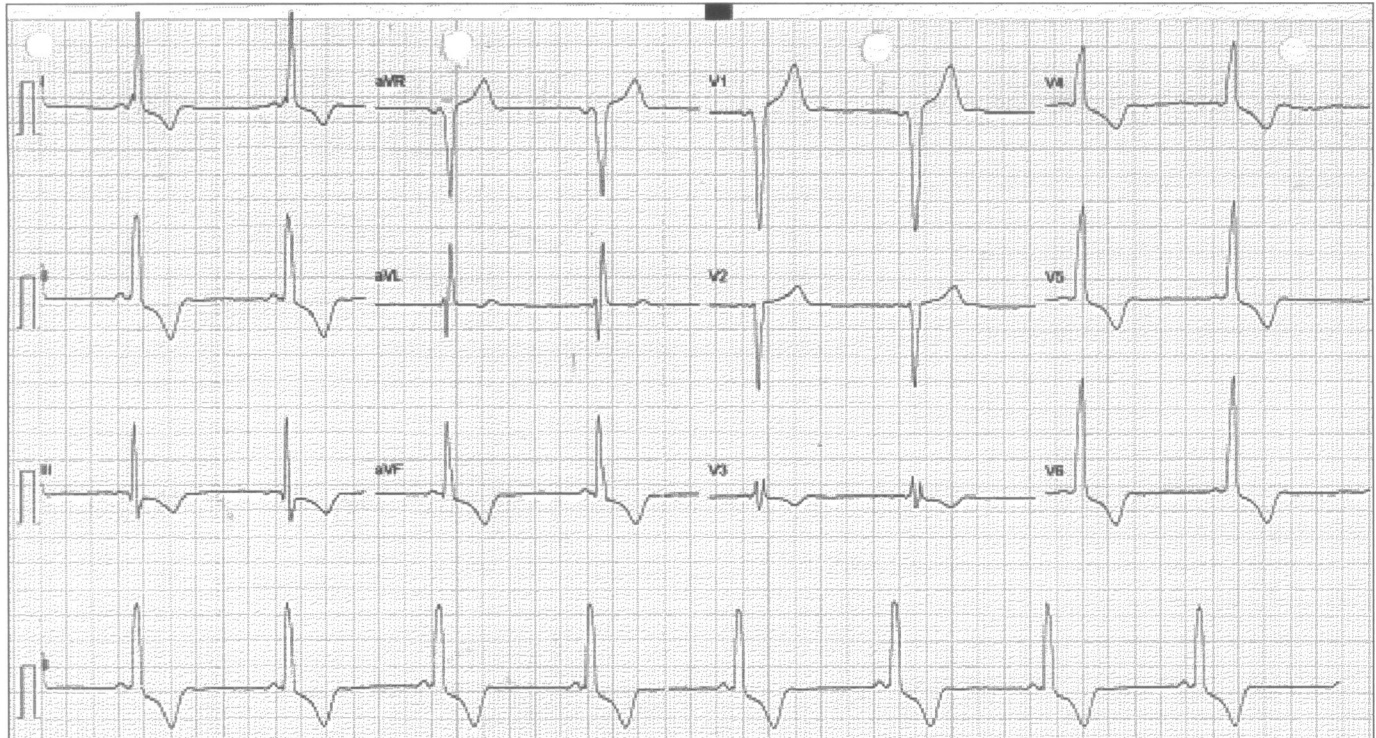
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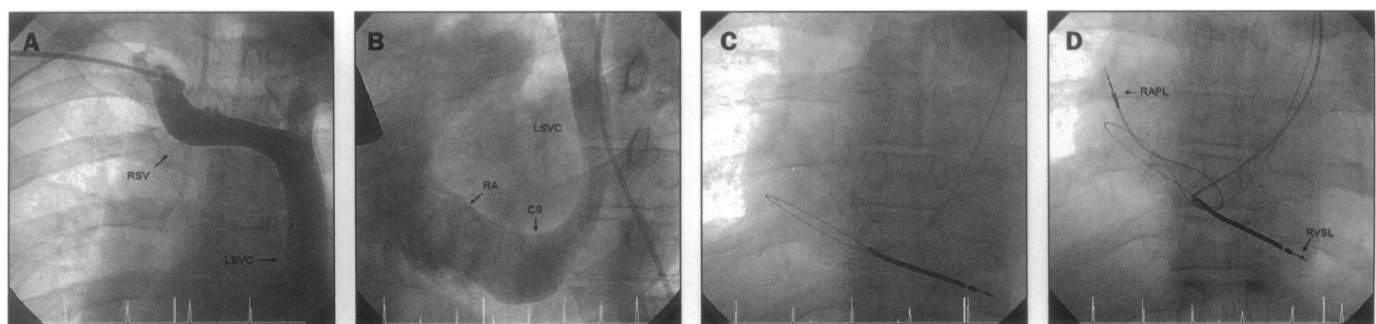


**Figure 1.** *Electrocardiogram with sinus rhythm, short PR interval and signs of an old anteroseptal myocardial infarction.*

vena cava, are bilaterally symmetric. During the normal course of morphogenesis, the left common cardinal vein drains into the left portion of the sinus venosus, i.e. the coronary sinus. In late embryonic or early foetal life the LSVC becomes obliterated. Failure of this obliteration results in persistent connection to the coronary sinus.<sup>3</sup>

In previous case reports technical difficulties have been described with pacemaker procedures although, in most cases, the use of the abnormal vein did succeed.<sup>4</sup> Because the lead tip enters the right atrium through the coronary sinus rather than the usual entrance of

the superior vena cava, one can imagine that the tip of the lead is deflected away from the tricuspid valve. Using the right subclavian vein rather than the left, curving the stylet into a pigtail or a L-shape and using a wide loop in the right atrium might overcome this problem.<sup>6-8</sup> Some authors describe an association between an absent RSVC (with persistent LSVC) and arrhythmias. The pathological substrate is an isolated LSVC which causes dilatation of the coronary sinus, leading to stretching of the atrioventricular (AV) node and His bundle. Therefore the AV node may be stretched out over a large coronary sinus. The sinus node often appears to be extremely small and hypo-



**Figure 2.** *Venography during ICD lead insertion through the right subclavian vein. A. Frontal view: approach through the right subclavian vein (RSV) showing absence of the right superior vena cava and a persistent left superior vena cava (LSVC). B. Lateral view: an enlarged coronary sinus (CS) is visible posterior to the right atrium (RA). C. The ICD lead curves anteriorly and superiorly before crossing the tricuspid valve. D. Additional pacemaker lead in right atrium (RAPL). RVSL= right ventricular shock lead.*

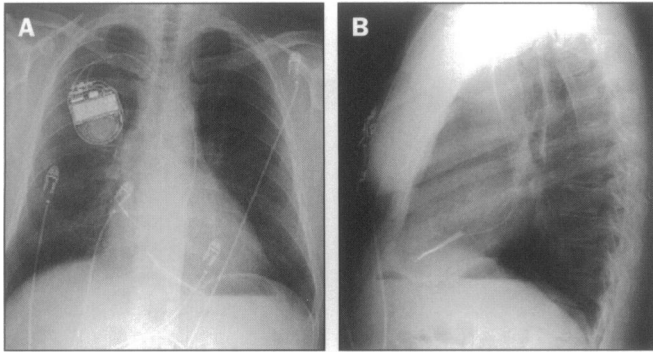


Figure 3. Chest X-ray after ICD insertion. Note position of the lead on left side of the upper mediastinum.

plastic.<sup>4,8</sup> The electrocardiogram often shows a leftward deviation of the P-wave axis and a shortened PR interval. The differential diagnosis should include pre-excitation syndrome.<sup>9</sup> Associated rhythm abnormalities are sick sinus, ectopic atrial rhythm, wandering pacemaker, first-degree AV block, complete heart block and tachyarrhythmias.<sup>5</sup>

### Conclusion

LSVC is a relatively common venous anomaly. Association with an absent RSVC is very rare. The majority of people with this anomaly do not have other malformations and on its own, it is usually of clinical insignificance. However this condition might cause difficulties with transvenous procedures such as insertion of pacemaker or ICD leads and carries an increased risk of sinus node dysfunction and other arrhythmias associated with the absence of the RSVC. We therefore emphasise the importance of recognising an LSVC. If, for instance, a central venous line appears on the chest X-ray on the left hand side of the heart, it is advisable proceeding to (contrast) echocardiography,

computed tomography or venography to obtain exact information of the upper venous system. If the patient does require a pacemaker or ICD implantation the additional information would be of great benefit: entrance of the superior vena cava in the left atrium and other structural abnormalities need to be excluded; the operator might consider a right-sided subclavian approach, the use of curved leads or longer leads and is aware of the associated arrhythmias. ■

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