Anomalies of cardiac venous drainage associated with abnormalities of cardiac conduction system

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The embryological development of the superior vena cava (SVC) is complex. If the left common cardinal vein fails to occlude it can, along with the left duct of Cuvier form a left SVC, which frequently drains into the coronary sinus. This may result in abnormalities in the anatomy of this structure. A persistent left SVC occurs in 0·5% of the normal population, and 3% to 4·3% of patients with congenital heart anomalies.

The pacemaking tissue of the heart is derived from two sites near the progenitors of the superior vena cava. The right-sided site forms the sinoatrial node, the left-sided site is normally carried down to an area near the coronary sinus.

Out of 300 patients with cardiac rhythm abnormalities, who have undergone electrophysiological studies (EPS), or permanent pacemaker insertion (PPI), we identified 12 patients with cardiac conduction abnormalities and anomalies of venous drainage. Anomalies of the coronary sinus may be associated with abnormalities of the conduction system of the heart. This may be due to the close proximity of the coronary sinus to the final position of the left-sided primitive pacemaking tissue. In our series of 300 patients, 4% had an associated left SVC, a similar incidence to that found in previous studies of congenital heart disease.

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Introduction

Abnormalities of the venous drainage of the heart and great vessels have previously been shown to be associated with conduction system abnormalities[1]. We present a case series of 12 patients with conduction system abnormalities who were found to have anomalies of their venous drainage.

Patients

Three patients were noted to have persistent LSVC, plus normal right-sided SVC, in association with atrio-ventricular re-entry tachycardia.

The first was a 26-year-old male with intermittent atrioventricular tachycardia found to be secondary to a left-sided accessory pathway during electrophysiological (EP) study. Resting ECG showed pre-excitation (Fig. 1). Dual SVCs were noted during EP study. He underwent successful treatment with an Intertach II antitachycardia pacemaker, programmed in AAI mode.

Patient 2 was a 35-year-old female with intermittent palpitation, 12 lead ECG showing pre-excitation. During EP study an accessory pathway was mapped from a markedly dilated pulmonary vein, to the mitral valve annulus. Again dual SVC were present. The accessory pathway was successfully ablated.

The third patient was a 29-year-old lady with documented atrioventricular (AV) nodal re-entry tachycardia. Resting ECG showed right axis deviation, and transoesophageal echo demonstrated an ostium secundum atrial septum defect (ASD), with a left to right shunt. Left-sided SVC was noted, and contrast injection revealed the presence of a right SVC. The ASD was successfully closed, and the patient has declined further studies.

A further group of 5 patients was found to have abnormalities of the coronary sinus, in association with arrythmias.
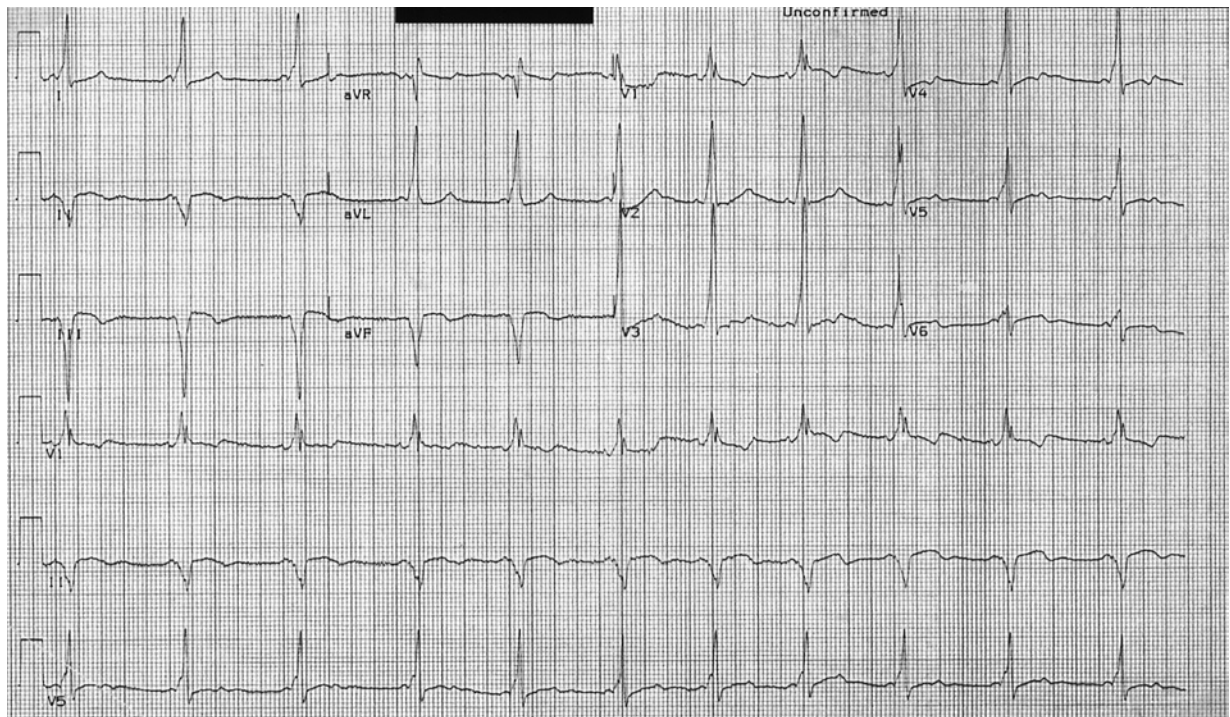
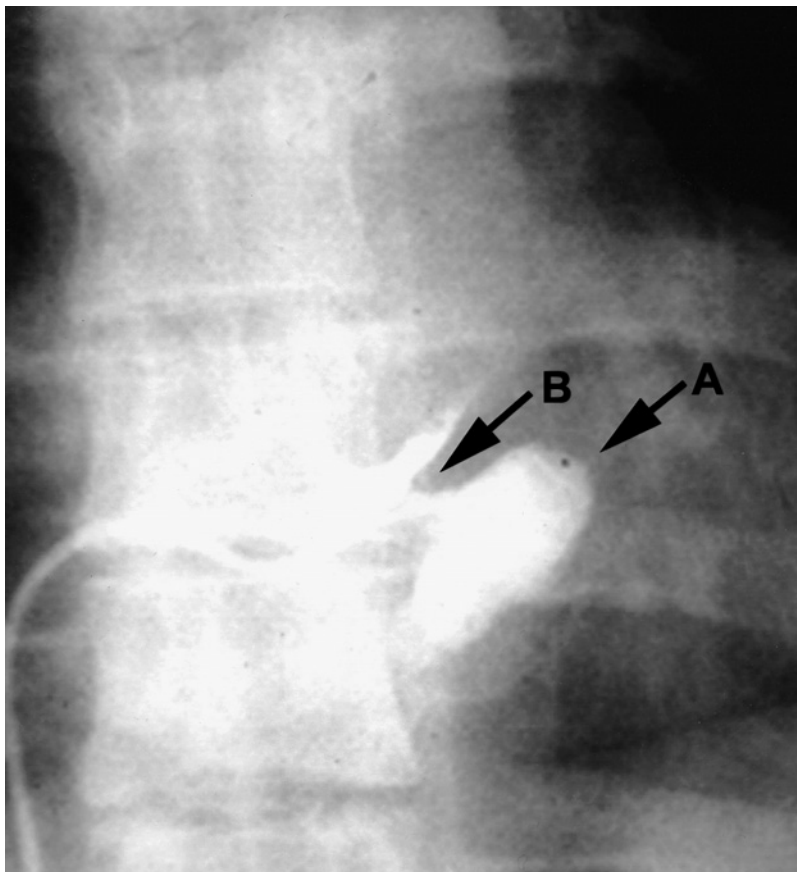***Figure 1*** **Twelve lead ECG showing Type A Wolff-Parkinson-White syndrome.**

Figure 2 **Contrast injection showing (A) coronary sinus diverticulum, (B) neck of diverticulum where accessory pathway was located.**

Figure 3 Contrast injection showing (A) sole left superior vena cava, draining into (B) grossly dilated coronary sinus.

The first was a 60-year-old female with intermittent palpitation, of left bundle branch morphology. An AV nodal re-entry tachycardia (AVNRT) was stimulated during EP study. A left SVC was noted, draining into a markedly dilated coronary sinus. A right SVC was also present (Fig. 2). Again the patient underwent successful radiofrequency ablation.

Patient 5, a 26-year-old male with Wolff-Parkinson-White (WPW) syndrome, was found to have a malignant accessory pathway, mapped from the neck of an abnor-mal diverticulum of the coronary sinus during EP study (Fig. 3). The pathway was successfully ablated.

A further patient, a 35-year-old male, again with intermittent palpitation secondary to WPW, was found during EP study to have an abnormal stricture in the coronary sinus. The accessory pathway was mapped from the stricture, and successfully ablated.

Patient 7, a 54-year-old female, presented with palpitation and syncope. Twelve lead ECG demonstrated periods of up to 3 s of sino-atrio block. EP study
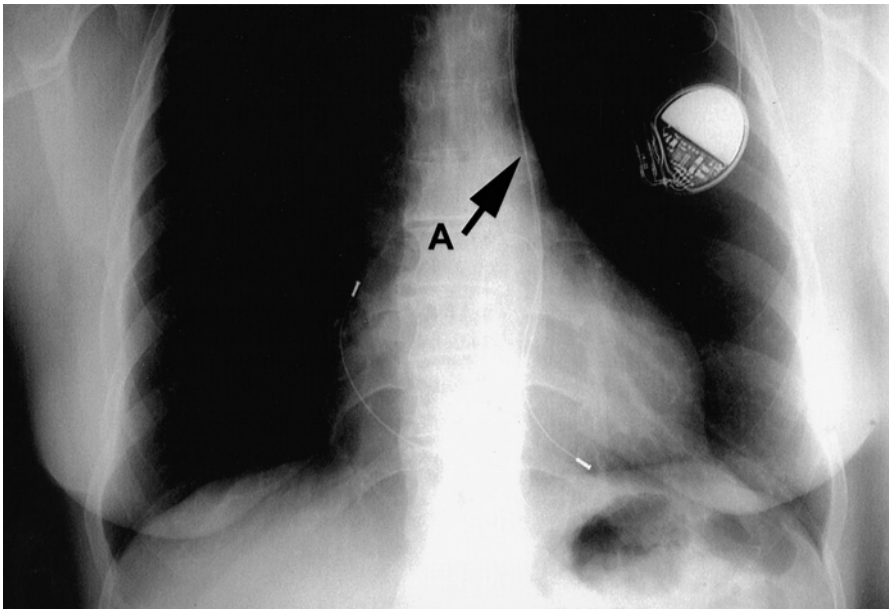
Figure 4   Chest radiograph showing (A) dual chamber pacemaker leads inserted via a sole left superior vena cava.
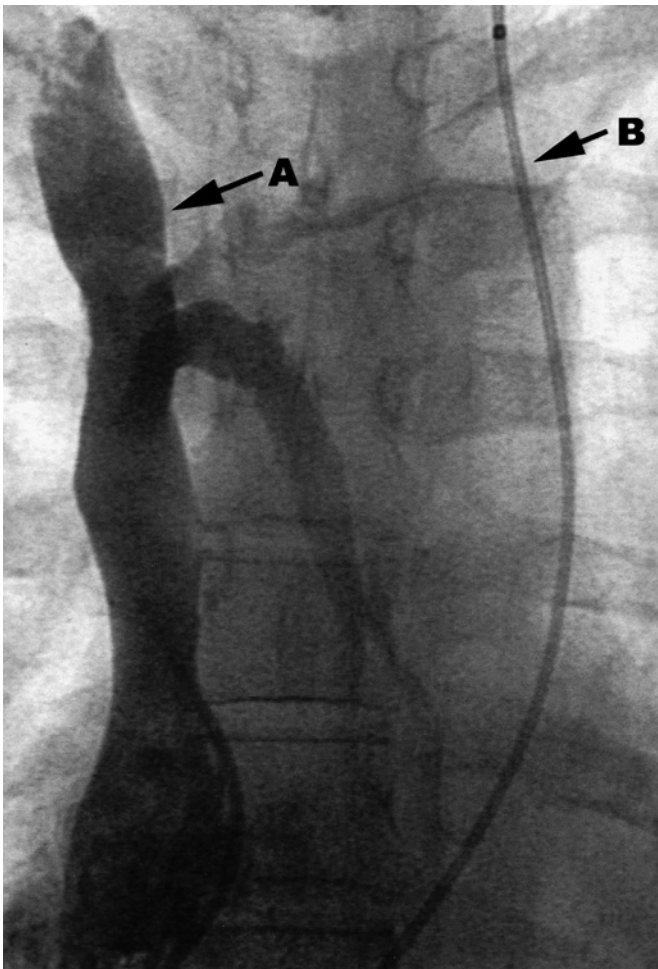

Figure 5   Contrast injection showing (A) right superior vena cava, (B) catheter in left superior vena cava.

Embryological Structures

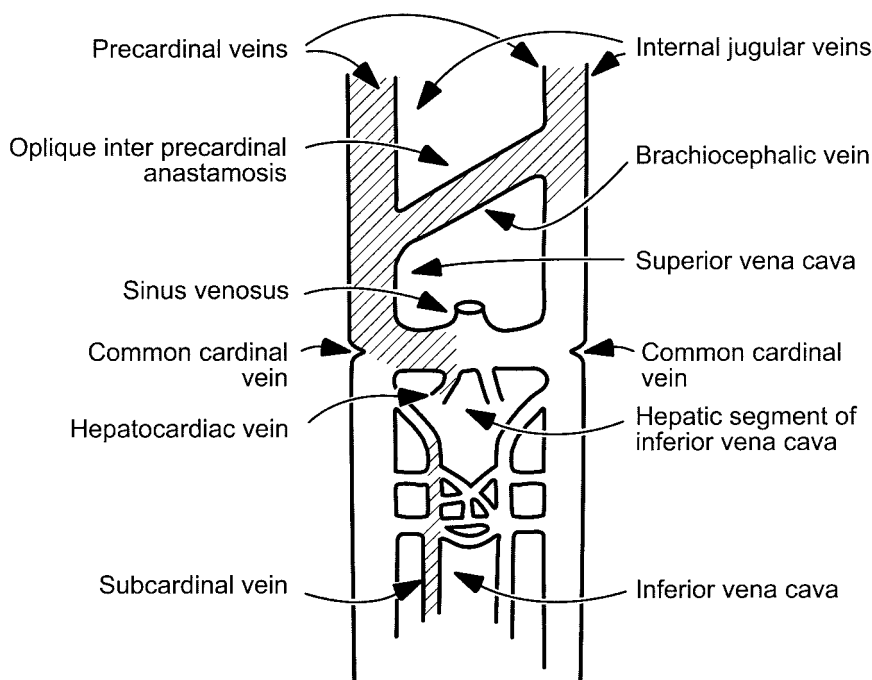## Structures persisting into adulthood (shaded)

Figure 6   **Diagram of development of the great vessels.**

demonstrated the presence of an AVNRT. A sole LSVC was noted, draining into a markedly dilated coronary sinus (Fig. 5). A dual chamber pacemaker was inserted via the LSVC (Fig. 4).

A 72-year-old female presented with sick sinus syndrome, and no history of ischaemic heart disease. During pacemaker insertion she was found to have a sole LSVC, opening into a giant coronary sinus. PPI was successful.

Sole LSVC, along with a normal coronary sinus was detected in three patients with conduction abnormalities. The first was a 17-year-old female with type A Wolff Parkinson White syndrome. During EP study she was found to have an LSVC, and normal coronary sinus. The left lateral wall accessory pathway was successfully ablated.

The next was a 61-year-old male who developed atrial fibrillation, and subsequently complete heart block following an anterior non-Q-wave myocardial infarction. During pacing he was noted to have a sole LSVC, draining into a morphologically normal coronary sinus. It may however be possible in this case, that the disordered conduction was secondary to acute myocardial infarction, and the LSVC a coincidental finding.

Patient 11, a 53-year-old lady with symptomatic bradycardia, and syncope underwent attempted pacemaker insertion. She was again found to have a sole LSVC. Unfortunately due to technical difficulties PPI was unsuccessful.

Finally, patient 12 was a 78-year-old female with carotid sinus hypersensitivity, and pre-syncope. Permanent pacemaker insertion demonstrated the presence of dual superior vena cavae.

Discussion

The association between left superior vena cava and abnormal rhythm patterns may be more than chance. The early embryonic heart has two symmetrically placed pacemaking areas at the junction of the common cardinal veins with the horns of the sinus venosus (see fig. 6). The right pacemaker area takes over pacemaker function to become the sino-atrial node. The left pacemaking area appears to be carried down to its final location at or near the coronary sinus. Whilst this is the normal pattern it is possible to speculate that where a left superior vena cava occurs, this may be accompanied by functionally well developed pacemaker tissue derived originally from the left sinus horn[2].

A persistent left superior vena cava has been reported in 0·5% of the general population and 4·3% of patients with congenital heart disease[3,4]. A left-sided superior vena cava may also exist along with a normal right-sided vena cava, or it may be the sole superior vena cava[5]. Some other venous anomalies may be associated with left-sided vena cava[5]. Thus left-sided superior vena cava has been described along with complete and partial

situs inversus[6]. The most frequent situation, however, is when there are bilateral superior vena cavae.

It has been suggested that left-sided superior vena cava, although associated with other cardiac malformations more frequently than expected by chance, is usually an innocent finding[5–8], though it can cause problems with pacemaker lead implantation[7]. This view is disputed by others who state that left superior vena cava is usually associated with other structural cardiac anomalies, such as atrial septal defect, tetralogy of Fallot, transposition of the great vessels[9,10]. Anderson *et al.* found that the presence of a LSVC, although not affecting the presence of cell types within the AV node, did lead to a significant alteration in the arrangement of cells, which could translate into conduction abnormalities[12].

Many case reports have noted left-sided superior vena cava in patients with abnormalities of cardiac rhythm or conduction. Other case series have also described the coexistence of left superior vena cava with a fistulous connection to the coronary sinus associated with Wolff Parkinson White syndrome, atrio ventricular tachycardia or atrio ventricular nodal re-entry tachycardia[1]. The use of a persistent superior vena cava to introduce pacemaker leads in a patient with junctional rhythm has also been described previously[11]. It is possible to postulate, therefore, that the abnormal development which leads to the presence of a left-sided superior vena cava could also result in abnormalities in conduction tissue, due to the close association of these areas during early intrauterine development. These abnormalities could then become manifest in some cases as abnormal rhythms.

Abnormalities of the anatomy of the great veins and coronary sinus could result in anomalies of the left pacemaking area close to the coronary sinus. Chiang *et al.* found major coronary sinus abnormalities in 4·7% of patients with accessory pathways, and found that they are often anatomically related to the localization of the accessory pathways[13]. Ninety-two per cent of patients with major coronary sinus abnormalities were found to have accessory pathways located exclusively in the left free wall and posteroseptal area. During cardiogenesis the coronary sinus develops from the proximal left sinus horn and the transverse portion of the sinus venosus[14]. Most accessory AV pathways are thought to be composed of ordinary myocardium[15] which is the remnant of incomplete separation of atrial and ventricular myocardium by the annulus fibrosis[16] at the same stage as the development of the coronary sinus in the embryo[14]. The same developmental stage and the proximity of the coronary sinus to the left AV ring and posterior septum may account for the high concurrence of left free wall and posteroseptal accessory pathways and major coronary sinus abnormalities. In contrast only 2·9% of patients with supra ventricular tachycardia had major coronary sinus abnormalities.

In our practice in electrophysiology we have seen, in approximately 300 successive cases 12 patients with abnormalities of cardiac rhythm and anomalies of venous drainage. This is considerably higher than the incidence of 0·5% suggested in the general community and similar to the frequency of 3% and 4·3% found in two major series of patients with congenital heart anomalies[4,6].

We would propose two different associations between anomalies of venous drainage and conduction system abnormalities. There may be an association between anomalies which lead to distortion of the coronary sinus and atrioventricular re-entry tachycardias. These tachycardias could be related to stretching of the atrioventricular nodal tissue, such as occurs when a left-sided superior vena cava drains into a dilated coronary sinus. A further association may be between left-sided superior vena cava, either alone, or with right-sided superior vena cava, and sino atrial node dysfunction. This relationship might be related to the embryological abnormalities which produced the persistent superior vena cava also being associated with abnormalities of the early conduction tissue which is in close proximity to the cardinal venous tissue.

It would be interesting to see if other workers have found similarly high rates of anomalies of venous anatomy in association with rhythm abnormalities similar to our findings.

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