Atrial Fibrillation Originating From Persistent Left Superior Vena Cava

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Background—The left superior vena cava (LSVC) is the embryological precursor of the ligament of Marshall, which has been implicated in the initiation and maintenance of atrial fibrillation (AF). Rarely, the LSVC may persist and has been associated with some organized arrhythmias, though not with AF. We report 5 patients in whom the LSVC was a source of ectopy, initiating AF.

Methods and Results—In 5 patients (4 men; age, 46±11 years) with symptomatic drug-refractory AF, ectopy from the LSVC resulting in AF was observed after pulmonary vein isolation. The ectopics were spontaneous in 2 and induced by isoproterenol in the others and preceded P-wave onset by 67±13 ms. During multielectrode or electroanatomic mapping, venous potentials were recorded circumferentially at the proximal LSVC near its junction with the coronary sinus (CS), but at the mid-LSVC level, they were recorded only on part of the circumference. The LSVC was electrically connected to the lateral left atrium (LA) and through the CS to the right atrium, with 4.1±2.3 CS-LSVC and 1.6±0.5 LA-LSVC connections per patient. Catheter ablation in the LSVC targeting these connections resulted in electrical isolation in 4 of the 5 patients without complications. After 15±10 months, the 4 patients with successful isolation, including 1 who had successful reablation for LA flutter, remained in sinus rhythm without drugs.

Conclusions—The LSVC can be the arrhythmogenic source of AF with connections to the CS and LA. Ablation of these connections resulted in electrical isolation. (Circulation. 2004;109:828-832.)

Key Words: catheter ablation ■ fibrillation ■ mapping

The major thoracic veins, with their specific electrical properties, have an established role in the genesis and maintenance of atrial fibrillation (AF).1,2 These include the vein of Marshall (VOM),3–5 which drains into the coronary sinus (CS). The VOM is located within a vestigial fold of pericardium, the ligament of Marshall (LOM), which is the developmental remnant of the embryonic left superior vena cava (LSVC).1,6 Rarely, the LSVC can persist, especially with congenital heart disease, and has been previously associated with some arrhythmias7 but not with AF. In the present investigation, we studied 5 patients in whom the LSVC was demonstrated to be a source of AF.

Methods

Patients

Five patients (4 men; age, 46±11 years) with symptomatic drug-refractory AF (4 paroxysmal, 1 persistent) of 146±77 months’ duration were studied at 3 different centers. Three of the patients presented to a single center over a 3-year period, during which a total of 851 patients had undergone catheter ablation for AF. Two patients had surgically corrected congenital heart disease, and 2 had previously successful ablation for other arrhythmias (Table). The presence of an LSVC was known in the 2 patients with previous surgery but was detected before the procedure by transesophageal echocardiography (n=1) or during the procedure (n=2) in the others.

Procedure

After written informed consent was obtained, multipolar catheters were introduced into the CS, right atria, and transseptally into the left atria (LA) for pacing and recording. Contrast venography was performed to delineate the pulmonary veins (PVs), CS, and LSVC. Before LSVC mapping, the PVs were electrically isolated by ablation in all patients, and in case 5, an atypical LA flutter was also ablated. If no ectopy was observed after PV ablation, provocative measures were attempted. LSVC mapping was performed in sinus rhythm.

Multielectrode Circumferential Mapping

In 3 patients, an 8F circumferential decapolar catheter (Lasso, Biosense-Webster) was introduced retrogradely through the CS into the LSVC. In sinus rhythm, a local double potential was recorded, comprising an initial far-field LA component followed by a discrete rapid deflection/spike, which was the local LSVC potential.8 This sequence was reversed during ectopy (Figure 1A). Mapping was started proximally near its junction with the CS, and the catheter was advanced distally into the LSVC until no further potentials could be recorded.
CS-L SVC connections were defined as the earliest activation recorded on 1 bipole (or >1 adjacent bipole) of the Lasso catheter placed at the proximal L SVC (at its junction with the distal CS) during sinus rhythm (Figure 1B, proximal L SVC). LA-L SVC connections were defined as the sites of earliest L SVC activation recorded on the Lasso catheter placed at mid-L SVC, corresponding to the fluoroscopic level of the left superior PV (Figure 1B, mid-L SVC) and confirmed by pacing at these sites from both the L SVC and adjacent lateral LA. Great care was taken during the pacing maneuvers to avoid direct capture of the LA while pacing from the LSVC, or vice versa. To achieve this, the pacing output was progressively reduced to demonstrate that both structures were activated with constant timing up to the loss of capture of the local structure. Thus, pacing from within one structure at or near a connection easily captured the other structure at low outputs, usually up to the local threshold, resulting in fusion of LA and L SVC potentials (Figure 2, top panels). In contrast, if the pacing site did not correspond to the site of a connection, capture was limited to the local structure, with the other structure being activated passively with a delayed potential (Figure 2, bottom panels). Multiple connections were suspected if a change in activation sequence occurred after ablation of the first connection and were confirmed after repeating the pacing maneuvers at the new site.

Electroanatomic Mapping

In 2 patients, electroanatomic mapping (CARTO, Biosense-Webster) of the L SVC was performed with the use of a 7F quadrupolar catheter with a location sensor (NAVI-STAR, Biosense-Webster). A bipolar recording from the proximal CS was used as the timing reference. The mapping catheter was advanced until no signals were recorded, then pulled back with multiple sequential recordings of the LSVC potential or if a change in activation sequence occurred after ablation of the first connection, which was localized and ablated as described. The mean duration of RF applied was 11±3 minutes for CS-L SVC and 9±3 minutes for LA-L SVC disconnection, respectively. After ablation, the LA could not be captured by pacing from the LSVC and vice versa in 4 patients, confirming electrical isolation, and ectopy and AF were no longer inducible with isoproterenol.

Ablation

For the CS-L SVC connections, ablation was started at the proximal L SVC at the site of earliest activation. In 1 patient, this resulted in elimination of all local potentials, whereas in the rest, other connections were unmasked, requiring additional RF delivery, including full circumferential ablation in 2. For LA-L SVC connections, ablation was performed at the anteromedial or oral part of the L SVC, starting at the site of earliest local activation. If potentials were still present after the initial RF application, the pacing maneuvers were repeated. An unchanged activation sequence implied that the connection still persisted, necessitating further local RF delivery, whereas a different sequence suggested the presence of a second connection, which was localized and ablated as described. The mean duration of RF applied was 11±3 minutes for CS-L SVC and 9±3 minutes for LA-L SVC disconnection, respectively. After ablation, the LA could not be captured by pacing from the LSVC and vice versa in 4 patients, confirming electrical isolation, and ectopy and AF were no longer inducible with isoproterenol.

Follow-Up

No complications were observed. During follow-up of 15±10 months, 3 patients remained in sinus rhythm without drugs,
Figure 1. A, Different forms of LSVC ectopy. In preceding sinus beats, double potentials were recorded in the LSVC by the Lasso catheter—an initial far-field LA component followed by sharp local LSVC potential (stars). This sequence was reversed in the conducted ectopic with and without AF initiation (earliest activation with polarity reversal indicated by arrows). Both LA and proximal CS were activated later. B, Distribution of LSVC potentials recorded on Lasso catheter (earliest activation denoted by arrows). Recording positions indicated on venogram (anteroposterior view) by long arrows. At the proximal LSVC (where it joined the distal CS), potentials were recorded along entire venous circumference. At mid-LSVC (level of the left superior PV), distribution of venous potentials was limited to 4 bipoles, corresponding to anteromedial aspect of the vein.
whereas AF recurred in the patient with unsuccessful LSVC isolation. The last patient had no recurrence of AF but required 2 further ablation procedures for LA flutter.

**Discussion**

This report presents new evidence about the LSVC as a source of ectopy that can initiate AF. These ectopics were conducted through connections to the lateral LA near the left PVs and through the CS. Ablation of these connections resulted in electrical isolation.

In the embryonic heart, bilateral pacemaking areas are present near the sinus horns and common cardinal veins. Whereas the right side takes over cardiac pacemaking function as the sinoatrial node, persistence of the left common cardinal vein as the LSVC may be associated with continuing presence of pacemaker tissue and hence ectopic pacemaker activity.

The presence of electrical potentials within the LSVC, consistent with the presence of muscle bundles, has been demonstrated with conventional and electroanatomic mapping. These potentials closely resembled the double potentials recorded in all thoracic veins, including PVs, SVC, and VOM. Although the exact mechanism for arrhythmogenicity could not be evaluated in the present study, the ability of the LSVC to generate rapid discharges (mean cycle length of repetitive beats was 159±11 ms) is a major factor for AF induction and maintenance. A similar mechanism has been observed in PVs and the LOM.

Our findings have implications for ablation of the more common LOM. Myocardial tracts inserting into the CS and LA free wall have been described in the LOM, and ectopy arising from this structure can be spontaneous or induced by isoproterenol, as in our patients. On the basis of anatomic studies of the LOM, it was suggested that endocardial ablation in the region of the lateral LA could sever both its LA and CS connections. This was performed by Hwang et al, guided by cannulation of the VOM, and resulted in AF termination in 4 of 6 patients but did not completely eliminate all LOM signals. In a different study, combined endocardial
and distal CS ablation, resulting in abolition of all LOM signals, was associated with better clinical outcome than endocardial ablation alone. In our patients, the presence of separate connections to the LA and distal CS necessitated ablation of both sites for LSVC isolation, which was electrophysiologically proven by pacing without capture of adjacent structures, and noninducibility of AF. Hence, a combination of endocardial and epicardial approaches may likewise be required for successful ablation of the LOM.

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References

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