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). These include the vein of Marshall (VOM), 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). Rarely, the LSVC can persist, especially with congenital heart disease, and has been previously associated with some arrhythmias 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 11 years’ 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. 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.
Arrhythmias

After PV isolation, ectopy was observed spontaneously in 2 patients and with isoproterenol infusion in the rest (Table). The earliest conducted ectopic activity preceded the onset of the P wave by 67±13 ms. Repetitive beats originating from the LSVC had short cycle lengths (mean, 159±11 ms), and AF was initiated in all patients (Figure 1A).

Distribution of LSVC Potentials

In all patients, LSVC potentials were recorded along the entire proximal circumference at its junction with the distal CS (Figure 1B, proximal LSVC). These potentials were not synchronous, with activation starting at a discrete site and spreading circumferentially. Proceeding distally into the LSVC, the circumferential distribution was lost, with local potentials recorded from only part of the perimeter. At the mid-LSVC level, the potentials covered 53±6% of the circumference (Figure 1B, mid-LSVC).

Connections

There were 4.1±2.3 CS-LSVC (range, 1 to 6) and 1.6±0.5 LA-LSVC connections (range, 1 to 2) per patient. The latter connected the lateral LA region near the anterior aspect of the left PV ostia to the anteromedial aspect of the LSVC and were located between the proximal and mid LSVC levels.

Ablation

For the CS-LSVC connections, ablation was started at the proximal LSVC 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-LSVC connections, ablation was performed at the anteromedial or medial part of the LSVC, 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-LSVC and 9±3 minutes for LA-LSVC 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 ablation

Figure 2. A, CS and LA connections, together with ablation site (red markers) are highlighted on electroanatomic activation map of LSVC in sinus rhythm (right anterior oblique projection). LSVC is activated from both CS and LA, with wave fronts colliding in the middle. Pacing at low output from earliest activation site at anteromedial LSVC (which abuts the lateral LA near the left PV ostia) resulted in fusion of both potentials, consistent with LA-LSVC connection. In contrast, during pacing from anterior and lateral LSVC (distant from LA-LSVC connection) in the same patient, LA potentials appeared significantly later after local LSVC potentials (stars). B, Pacing from lateral LA captured LSVC at low outputs up to LA threshold without significant delay. This resulted in near-fusion of LSVC and LA potentials, suggesting proximity of LA-LSVC connections. Right atrial (RA) activation was late. In contrast, pacing from anterolateral LA (LA appendage) captured both LA and LSVC at high output (8 mA). When output was lowered (<5 mA), direct LSVC capture was lost. However, the LSVC was still activated through the CS, with resultant delay in LSVC potentials (stars).
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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