Coexistence of Left-Sided Atrioventricular Accessory Pathways With a Common Inferior Pulmonary Vein Ostium

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Background—As the technique for radiofrequency catheter ablation for atrial fibrillation (AF) has progressed, so has our knowledge of both normal and abnormal anatomy of the left atrium and pulmonary veins (PV). We treated several AF patients with accessory conduction pathways (ACP) who were also found to have a common ostium of inferior PVs (CIPV), a relatively rare PV anomaly. No relation between ACP and PV anomalies has ever been reported, and the aim of our study was to study this association.

Methods and Results—This study included 137 consecutive patients (104 men; mean age, 60±9 years) who underwent AF ablation for paroxysmal and persistent AF at our institution from March 2009 to August 2010. We analyzed coexisting supraventricular tachycardias and left atrium and PV morphology by multidetector row CT. Thirty-eight of 137 patients (27.7%) were found to have some PV anomaly, consisting of 13 with a common trunk of left PV, 19 with right additional PV, 3 with a common trunk of right PV, and 3 with CIPV. Thirty-one patients (22.6%) had supraventricular tachycardias. They were 26 cases of atrial flutter, 4 of Wolff-Parkinson-White syndrome, and 3 of atrioventricular nodal reentrant tachycardia. The prevalence of a coexisting ACP was significantly higher in patients with CIPV than in those without CIPV (3 of 3 [100%] versus 1 in 134 [0.7%]; P<0.0001). All ACPs with CIPV were located in the left side. The other supraventricular tachycardias were not associated with any PV anomalies.

Conclusions—There is a possible association between CIPV and left-sided ACP in AF patients. This suggests that there is a likelihood of developmental association between them. (Circ Arrhythm Electrophysiol. 2011;4:310-317.)

Key Words: common ostium of the inferior pulmonary veins ■ accessory pathway ■ atrial fibrillation ■ pulmonary vein ■ computed tomography

The pulmonary veins (PVs) have been demonstrated to be an important source of atrial ectopy, initiating paroxysmal atrial fibrillation (PAF)1; this is why electric isolation of the PVs by radiofrequency catheter ablation (RFCA) is highly effective for treatment of PAF.2,3 Studies show that atrial ectopy in the PVs is associated with structural anomalies of the PVs,4–6 and both multidetector row computed tomography (MDCT) and magnetic resonance imaging (MRI) can provide precise localization of such structures and the left atrium (LA), guiding RFCA.7,8 Common ostium of the inferior PVs (CIPV) is a rare anomaly of the PV that has been shown to be an important trigger of PAF.9 However, an association between CIPV and other supraventricular tachycardias (SVT) has never been reported. Meanwhile, the atrioventricular accessory conduction pathway (ACP) is a common anatomic anomaly that causes atrioventricular reentrant tachycardia but is rarely accompanied by structural cardiac abnormalities, except for those that are age-related.10 We recently treated 3 patients with PAF who were found to have CIPV and coexisting ACP. The purpose of this study was to analyze the relationship between CIPV and ACP and their characteristics.

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Methods
This study included 137 consecutive patients who underwent AF ablation for drug-resistant paroxysmal and persistent AF at our institution from March 2009 to August 2010 and who underwent MDCT before the procedure. None had a history of prior LA ablation.

MDCT Imaging of the LA
All patients underwent MDCT imaging at our institution, as is our standard procedure before PVI. This involved nongated contrast-enhanced CT of the chest with a 64-detector helical CT scanner (Aquilion 64, Toshiba Medical Systems Co, Tokyo, Japan) using 64×0.5 mm collimation and a table feed of 45.0 mm/revolution, a
Electrophysiological Study and Catheter Ablation:

All antiarrhythmic drugs were withdrawn 5 half-lives before ablation. The CARTO system was used to guide the RFCA. Bilateral extensive encircling ipsilateral PV isolation was accomplished with the double LASSO technique for all PVs. The RFCA application was performed under guidance of the CARTO Merge image integration software (CARTO Merge Image Integration Module, Biosense Webster, Inc).

Statistical Analysis:

Statistical analysis was performed using SPSS version 18.0. A probability value of <0.05 was considered statistically significant.

Results:

Of 137 consecutive patients (104 men; mean age, 60 ± 9 years; 118 paroxysmal AF and 19 persistent AF), 38 patients (27.7%) had some thoracic vein anomalies in their MDCT image, consisting of 13 (9.5%) with common trunk of the left PV, 19 (13.9%) with right additional PV, 3 (2.2%) with common trunk of the right PV, 3 (2.2%) with CIPV, 1 (0.7%) with persistent left superior vena cava, and 1 (0.7%) with interruption of the inferior vena cava withazygos continuation (2 patients had 2 PV anomalies).

Thirty-one patients (22.6%) also had SVTs other than AF or atrial tachycardia. There were 26 cases of atrial flutter (19.0%), 4 cases of Wolff-Parkinson-White (WPW) syndrome (2.9%), and 3 cases of atrioventricular nodal reentrant tachycardia (2.2%) (2 patients had 2 of these arrhythmias). It is particularly worth noting that all 3 patients with CIPV had a coexisting ACP. The prevalence of a coexisting ACP was significantly higher in the patients with CIPV compared with those without CIPV (3 of 3 [100%] versus 1 of 134 [0.7%]; P < 0.0001, Table 1). The other SVTs were not associated with any PV anomalies.

The characteristics of these 3 patients are shown in Table 2, and we present their case reports below.

Case 1:

A 45-year-old man with paroxysmal narrow QRS regular tachycardia, not AF, was referred to our institution for RFCA. A
Delta wave had been found in his 12-lead surface ECG at the age of 36. He was taking medications for hypertension but had no history nor family history of other cardiac disease. His echocardiogram showed normal cardiac function and no obvious structural heart disease. A delta wave was apparent on his admission ECG (Figure 1A). Electrophysiological study revealed atrioventricular reentrant tachycardia complicated with PAF, and we performed RFCA only for the left lateral ACP by the conventional ventricular approach. However, we found it necessary to apply radiofrequency energy in the CS to eliminate retrograde ACP conduction. When we were done ablating the ACP, retrograde and antegrade ACP conduction were not observed even with adenosine triphosphate. Atrioventricular reentrant tachycardia (AVRT) could not be induced by programmed stimulation even with isoproterenol.

Some time later, the patient returned to the hospital with palpitations, and PAF was clinically documented for the first time. His PAF was drug-resistant, and we performed PV isolation after conducting MDCT to study LA and PV morphology. The MDCT showed dilation of the ostium of RSPV and CIPV (Figure 2A). Although delta waves were absent from his ECG at second admission, electrophysiological study revealed the recovery of retrograde conduction through the left lateral ACP and orthodromic atrioventricular reentrant tachycardia was easily induced by programmed stimulation. We unsuccessfully attempted RFCA of the ACP via an atrial approach (through transseptal puncture) and once again succeeded only after RFCA application in the CS. Ectopy initiating PAF frequently occurred from the CIPV (LIPV) and LSPV. We attempted PV isolation with “box isolation” using the original method of the double Lasso technique under guidance of the CARTO merge system, but this attempt was not successful. Therefore, we performed a “tricircle ablation,” with which we achieved PV isolation (Figure 3A).

**Case 2**
A 58-year-old man who had undergone RFCA for overt WPW syndrome at another institution 1 year previously was

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**Table 2. Characteristics of Patients With CIPV**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age, y</th>
<th>Complicating Disease</th>
<th>Type of AF</th>
<th>AF Trigger</th>
<th>Coexisting ACP</th>
<th>Type of WPW Syndrome</th>
<th>ACP Location</th>
<th>AVRT</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>46</td>
<td>Hypertension</td>
<td>Paroxysmal</td>
<td>LIPV, LSPV</td>
<td>+</td>
<td>Overt</td>
<td>Left lateral</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>58</td>
<td>None</td>
<td>Paroxysmal</td>
<td>RIPV, LSPV</td>
<td>+</td>
<td>Overt</td>
<td>Left posterolateral</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>56</td>
<td>Polycystic liver</td>
<td>Paroxysmal</td>
<td>SVC</td>
<td>+</td>
<td>Intermittent</td>
<td>Left posterior</td>
<td>-</td>
</tr>
</tbody>
</table>

ACP indicates accessory pathway; AVRT, atrioventricular reentrant tachycardia.

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**Figure 1.** Twelve-lead resting ECGs of 3 patients with common ostium of the inferior pulmonary veins. ECGs A, B, and C are from cases 1, 2, and 3, respectively, described in the text. Within each case, the 2 strips on the left are from before RFCA, and the 2 on the right are from after. Delta waves are visible in A and B before the RFCA but not after the ablation. Case 3 patient had intermittent type WPW syndrome, which was diagnosed for the first time at admission for catheter ablation of PAF (C).
referred to our institution for drug-resistant PAF. There were no delta waves on his admission ECG (Figure 1B). He had no other history and no family history of heart disease. His echocardiogram showed normal cardiac function and no

Figure 2. MDCT imaging of patients with CIPV. Posterior images of the LA in patients with CIPV are shown. A, B, and C represent cases 1, 2, and 3, respectively, as described in the text. In A and C, dilation of the ostium of the right superior PV is also observed.

Figure 3. Anterior endoscopic view of the LA in patients with CIPV and their ablation site. The anterior endoscopic LA images of cases 1, 2, and 3 (as described in the text) produced by the CARTOmerge system are shown in A, B, and C, respectively. The CIPV can be seen from inside the LA. In each image, the RFCA application sites for PV isolation (red dot) and the AP location (white dot and arrow) are shown, except that in case 3, a white dot is placed at the site where ACP was thought to exist, because ablation for the ACP was not performed. In case 1, tricircle ablation (A) was performed for PAF ablation and in case 2, box isolation (B).
obvious structural heart disease. His MDCT image showed CIPV (Figure 2B). His electrophysiological study revealed that retrograde ACP conduction remained (but not antegrade ACP conduction), and orthodromic AVRT was easily induced. Spontaneous firing also occurred frequently from the CIPV (RIPV) and LSPV. We achieved ablation of the ACP by the transseptal atrial approach, and PV isolation for the CIPV was completed by a “box isolation” (Figure 3B) with double Lasso technique under guidance of CARTO merge system. Complete isolation of the whole of the LA posterior wall and 4 PVs was confirmed by the absence of local electric activity in them and the inability to capture outside the box during pacing in each of them.

Case 3
A 56-year-old man was referred to our institution for symptomatic drug-resistant PAF. His symptoms had always occurred with PAF without delta wave, and narrow regular tachycardia had never been documented. He had polycystic liver, for which he was receiving no specific therapy. He had no other history and no family history of heart disease. His echocardiogram showed normal cardiac function and no obvious structural heart disease. We scheduled his PV isolation, and MDCT was performed before the procedure, which revealed CIPV and dilation of the RSPV ostium (Figure 2C).

A delta wave was observed on his ECG at admission (Figure 1C), and the patient was diagnosed with intermittent WPW syndrome for the first time. Electrophysiological study suggested an ACP located in the left posterior area (Figure 3C), but no retrograde ACP conduction was observed, and antegrade ACP conduction was rarely observed (intermittent). AVRT was not inducible even with isoproterenol. We did not perform RFCA for ACP because his ACP was considered a bystander and because we could not determine its location due to its intermittency. This patient’s PVs including the CIPV showed no ectopic activity. Instead, frequent spontaneous firings initiating PAF were observed repetitively from the SVC. Therefore, we performed only SVC isolation. After SVC isolation, AF could not be induced with isoproterenol and atrial burst pacing.

Follow-Up After Catheter Ablation
Over the mean 6 months of follow-up after the RFCA procedure, there were no obvious complications related to the procedure. All 3 cases remained free from AF and paroxysmal supraventricular tachycardia.

Discussion
The present study is the first to describe a possible association between a PV anomaly and ACP. The incidence of ACP detected electrocardiographically has been reported to be 0.1 to 3.1 in 1000 in people of all ages, and atrial flutter-fibrillation may be the presenting arrhythmia in 5% to 10% of patients with ACP. The majority of patients with ACP have no heart disease, but some ACPs are associated with certain congenital heart abnormalities, particularly Ebstein anomaly of the tricuspid valve, hypertrophic obstructive cardiomyopathy, and some genetic defects have been also reported to accompany ACP. Recently, some reports attempted to demonstrate the incidence and characteristics of paroxysmal supraventricular tachycardia in AF patients. According to those reports, AVRT could be induced during electrophysiological study for PAF ablation in 1.7% to 1.9% of the patients. Furthermore, successful catheter ablation of accessory pathways was found to prevent further recurrence of AF in 91% of patients in one study. However, no reports have ever analyzed an association between a PV anomaly and ACP.

Conversely, MDCT and MRI of the LA and PVs are now widely performed to assess atrial anatomy before procedures for AF, and physicians are becoming increasingly familiar with normal and abnormal anatomy of these structures. In previous reports, a common trunk of the left or right PV, and the right PV were occasionally described, but CIPV was thought to be a rare condition observed in 0.9% to 1.5% of AF patients. Furthermore, in those reports, the relation between CIPV and SVTs other than AF was not assessed, and some of those included patients with a history of prior RFCA application for LA, which may have affected LA and PV morphology. Our study consisted entirely of patients who underwent first-time LA ablation. Given the rareness of ACPs on their own and the rareness of CIPVs on their own the presentation of 3 patients having both ACP and CIPV triggered our interest.

Characteristics of the ACPs With CIPV
The patients with CIPV were all middle-aged men without any structural heart disease. ECG delta waves were recorded in all 3. All ACPs were located in the left side (Figure 3), and there were no specific macroscopic structures around the ACP in their MDCT images. In the only WPW syndrome patient without CIPV, the ACP was located on the right side.

Some previous studies have demonstrated coronary vein anomalies related to accessory atrioventricular pathways, especially the posteroseptal epicardial ACPs. In this study, all 3 patients underwent coronary venography during the procedure, but no anomalies of the coronary sinus were detected. In case 1, the ablation of ACP failed with both a venicular and atrial approach, and RFCA application in the CS was needed to eliminate his ACP. In endocardial RFCA of ACPs, ablation failure may be due partly to the epicardial insertion of the ACP.

Characteristics of CIPV
As in past reports, the MDCT imaging (Figure 4) revealed no structures around the common ostia external to the LA and PVs that could possibly have obstructed development of the PV, forcing CIPV to remain undifferentiated. The common trunks of the inferior PVs projected in a posterior direction and were very close to the esophagus. It is very important to avoid ablating these areas or to reduce radiofrequency energy to reduce the risk of damage to the esophagus.

Some PV anomalies have been associated with the initiation of AF. In a past report, focal ectopy followed by AF was observed in 4 of 11 CIPVs. In this study, 2 of 3 patients with CIPVs had spontaneous firing from the CIPV during...
their procedure session. CIPVs may play an important role in triggering AF, similar to other type of PV anomalies.

Development of PV and Atrioventricular Septation

Embryologically, the common PV joins to the primary atrial component of the heart and starts to bifurcate, dilate, and incorporate into the LA at a gestational age of around 30 days. By 7 to 8 weeks of gestation, contemporaneously with septation of the atriums and ventricles, the common PV separates into the RPV and LPV and positions itself in the most inferior part of the LA, adjacent to the atrioventricular groove and CS. At the age of 15 weeks, it is possible to identify 4 PV ostia draining into the LA.31 During this developmental process, incomplete PV incorporation can result in a unilateral common PV ostium, whereas extreme PV incorporation can lead to more than 4 PV ostia.32

The atrioventricular sulcus and cushions develop concurrently with PV development, and the continuity of atrial and ventricular muscle is interrupted gradually. At the end of the third month of gestation, the atrial myocardium and ventricular myocardium are almost completely separated in normal subjects.33 ACPs in WPW syndrome are thought to be derived from abnormal remnants of muscle fibers that provided direct continuity between atrial and ventricular myocardium.34 Normal regression of muscle fibers during atrioventricular septation may be inhibited by a molecular defect. We note that in the present study, the CIPV were in the inferior LA, and all of the ACP were in the posterior to lateral LA atrioventricular border, near the inferior LA. We think that it is of significant interest that developmentally at the time of common PV bifurcation, the common PV is adjacent to the atrioventricular groove, suggesting a possible common etiology. Furthermore, the other types of PV anomalies were not related to ACPs or other supraventricular tachycardias, suggesting that CIPV may belong to a different class of PV anomalies from the others, perhaps due to its proximity to the AV border.

Clinical Implications

Some consideration is required in treating AF with CIPV. First, CIPV is a potential target vein of PV isolation. However, the standard ablation strategy of PV isolation, extensive encircling ipsilateral PV isolation,3 is very difficult in CIPV, especially the making of a longitudinal line on the LA posterior wall. Although one report recommends “tricircle” ablation,8 we consider box isolation to be a preferable strategy for CIPV because box isolation does not need a transverse RFCA line to the posterior aspect of the LA, which is thought to be closer to the esophagus than other LA and PV morphologies.13 Second, the association of CIPV with ACP should be considered. In treating AF associated with CIPV, the coexistence of ACP should be carefully investigated.
Limitations
There are several limitations to the study. First, it was a retrospective analysis with a small number of patients, and the study group consisted solely of patients who underwent AF ablation, that is, it was a selected population and different from the general population. Second, this study did not include patients who underwent ablation only for WPW syndrome because MDCT is not routinely performed for WPW patients at our institution. Therefore, we could not assess the prevalence of CIPV in WPW syndrome. A prospective and larger study is needed to investigate the precise relationship between CIPV and ACP.

Conclusions
We found a possible association between a unique pulmonary vein morphology, CIPV, and left-sided accessory pathway in AF patients. This suggests a developmental association between them.

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Disclosures
None.

References


CLINICAL PERSPECTIVE

The majority of patients with atrioventricular accessory pathway (ACP) have no heart disease. Recently, we treated several patients for atrial fibrillation who had both an ACP and a common ostium of the inferior pulmonary veins (CIPV). Among 137 consecutive patients who underwent first time atrial fibrillation ablation and multidetector row computed tomography of the left atrium and pulmonary veins at our institution, 4 were found to have coexisting ACP and 3 had CIPV. Surprisingly, all 3 patients with CIPV had a coexisting ACP. Furthermore, the other types of PV anomalies were not related to ACP or other supraventricular tachycardias. Considering the rareness of both ACP and CIPV in atrial fibrillation patients, this fascinating finding prompted us to conduct a statistical analysis, which produced a probability value of <0.0001. The literature on cardiac development indicates that the development of pulmonary veins and atrioventricular septation overlap both temporally and spatially. To the best of our knowledge, this is the first study describing an association between a PV anomaly, namely CIPV, and an ACP. This finding may offer new insights into the mechanism of cardiac development, especially the formation of ACPs.
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