



Supraventricular tachyarrhythmias in patients with a persistent left superior vena cava

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Aims

A persistent left superior vena cava (PLSVC) is the most common thoracic venous anomaly. This venous anomaly can impact the evaluation and treatment of supraventricular tachyarrhythmia (SVA). The aim of this study was to assess the proportion and characteristics of PLSVC in adult SVA patients.

Methods and results

From July 2002 to July 2012, clinical and procedural data from databases of 10 cardiac electrophysiology laboratories in the Yeungnam region of the Republic of Korea were reviewed. Of 6662 adult SVA patients who underwent an EP study or catheter ablation of SVA during the 10-year study period, 18 patients had PLSVC (mean age 47.6 ± 14.8 years, 10 men). The proportion of PLSVC in adult SVA patients was 0.27% (18/6662). SVA type and procedural outcomes of radiofrequency (RF) catheter ablation in these patients were investigated and the results were as follows: successful slow pathway modification in six of seven patients with atrioventricular nodal reentrant tachycardia (AVNRT), successful ablation of accessory pathway in three of four patients with atrioventricular reentrant tachycardia, and successful ablation of atrial tachycardia (cavotricuspid isthmus-dependent in two, septal macroreentry in one, focal from the PLSVC in one) in three of four patients. In one patient with junctional tachycardia, catheter ablation failed. In two patients with atrial fibrillation, catheter ablation was successful.

Conclusion

Among adult SVA patients who underwent an EP study or RF catheter ablation during the 10-year study period, 0.27% had PLSVC. The most common type of SVA was AVNRT. The success rate of catheter ablation was 82% in SVA patients with PLSVC. There were no procedure-related complications.

Keywords

Superior vena cava • Supraventricular tachycardia • Catheter ablation

Introduction

A persistent left superior vena cava (PLSVC) is the remnant of the left cardinal vein, an embryological vessel, that is present during the

early developmental period.¹ As the most common thoracic vein anomaly, the incidence of PLSVC is between 0.3 and 2% in individuals with a normal heart and 4.5–9% in those with a concomitant congenital heart disease.^{1–3} PLSVC typically drains into the right atrium (RA)

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What's new?

- Persistent left superior vena cava (PLSVC) was observed in 18 of 6662 patients (0.27%) with supraventricular tachyarrhythmia (SVA) during a 10-year study period.
- Ectopy from the PLSVC was found in 5 of 18 patients, and decremental property of PLSVC potential was found.

via the coronary sinus (CS), which can become enlarged due to volume overload.⁴ This enlarged CS can complicate mapping and ablation procedures of supraventricular tachyarrhythmias (SVA) arising from the CS ostia and the triangle of Koch.⁵ Also, excessive motion of a CS mapping catheter has been shown to preclude accurate electrogram localization.⁶ Furthermore, PLSVC itself can be a source of ectopy that initiates atrial fibrillation (AF), but catheter ablation targeting PLSVC has a risk of major complications.^{7,8} Thus, the presence of PLSVC can impact the evaluation and treatment of SVAs. Despite its clinical importance described above, only single case report or case-series studies have been previously reported and the research on the exact proportion of PLSVC in adult patients with SVA undergoing catheter ablation are scarce.^{6–17}

Therefore, the aims of this study were (i) to evaluate the proportion of PLSVC in SVA patients and (ii) to investigate the clinical and electrophysiological (EP) characteristics as well as the procedural outcomes of radiofrequency (RF) catheter ablation in this group of patients.

Methods

Study population

This retrospective multicentre study included all consecutive adult SVA patients who underwent an EP study and/or catheter ablation between July 2002 and July 2012 in the Yeungnam region of the Republic of Korea. The Yeungnam region includes the southeastern metropolitan cities of Busan, Ulsan, and Daegu as well as the Gyeongsangnam-do and Gyeongsangbuk-do provinces of Korea (see Supplementary material online, Figure S1). The area is 32 266.92 km² and had a total population of 13 153 705 in July 2002 and 13 199 622 in July 2012, as indicated by resident registration-based population statistics, thereby accounting for approximately 24% of the total population of Korea.¹⁸ There are 13 EP centres located in this region, 10 of which participated in this study. A total of 6662 patients were reviewed, and from this population, 18 patients with PLSVC were selected. Clinical characteristics, EP characteristics, outcomes of catheter ablations, and follow-up data for these patients were collected and retrospectively analysed.

Electrophysiological study and catheter ablation procedure

Before the procedure, careful evaluations were performed, including patient history and physical examination, as well as a detailed 12-lead surface electrocardiographic (ECG) examination and two-dimensional transthoracic echocardiogram (TTE) to assess left ventricular (LV) ejection fraction (EF) and to rule out any structural and/or valvular disease. All patients signed informed consent for the procedure. The procedures were performed under a fasting state with light sedation after withdrawal

from antiarrhythmic drugs for a period of time equal to five-times the half-lives of the drugs. Multipolar electrode catheters were inserted percutaneously into the femoral vein, and, using single-plane fluoroscopy, were deployed into the CS, His bundle, right ventricular apex, respectively. The EP study included: (i) measurement of the conducting properties of the atrium, AV node, ventricle, and accessory pathways (if present); (ii) initiation of SVAs; and (iii) determination of the mechanism of tachycardia. If tachycardia could not be induced in the baseline state, isoproterenol or atropine was used to facilitate induction. Induced SVAs were classified as atrioventricular nodal reentrant tachycardia (AVNRT), atrioventricular reentrant tachycardia (AVRT), typical atrial flutter (AFL), atrial tachycardia (AT), junctional tachycardia (JT), and AF. A three-dimensional electroanatomic mapping system (CARTO, Biosense Webster Inc., Diamond Bar, CA, USA or NavX Ensite Velocity, St. Jude Medical Inc., Milwaukee, WI, USA) was used in selected patients. RF catheter ablation was tried if indicated. Catheter ablation was performed using a 4 mm tip non-irrigated ablation catheter. External irrigation catheter (3.5 or 4 mm tip) was used in AF patients and in some AT or AFL patients. Endpoints of catheter ablation were: (i) slow pathway modification in AVNRT, (ii) bidirectional absence of accessory pathway conduction in AVRT, (iii) bidirectional conduction block across the critical isthmus in macro-reentrant AT, (iv) successful ablation of ectopic foci in focal AT or JT, (v) entrance and exit block of the pulmonary veins (PV) and non-PV foci ablation in AF, and (vi) elimination or dissociation of PLSVC potentials if it causes clinically significant SVA.

The presence of PLSVC was suspected if (i) CS dilatation was observed on preprocedural TTE or (ii) catheter for recording for CS electrogram was positioned in an unconventional manner (directed to the left main bronchus rather than the anterior basal left ventricle). Subsequently, contrast venography using a long sheath or pigtail catheter was performed to confirm and delineate the PLSVC.

Statistics

Continuous variables are expressed as the mean value \pm standard deviation. Categorical variables are expressed as numbers and percentages. Only descriptive statistics are provided.

Ethics statement

The study protocol was reviewed and approved by the institutional review board of Pusan National University Yangsan Hospital (IRB No. 05-2015-141), and performed in accordance with the Declaration of Helsinki.

Results

Baseline characteristics of the study subjects

Table 1 summarizes the characteristics of SVA patients with PLSVC. The mean age was 47.6 ± 14.8 years and 10 patients (55.5%) were male. Three patients (16.7%) had prior operations for congenital heart disease: one patient had atrial septal defect (ASD) closure, one patient had ASD with Cor triatriatum correction operation, and one patient had an operation for double outlet right ventricle, subaortic ventricular septal defect, pulmonic stenosis, and ASD with redo closure of ASD later. Surgical patch closure was performed in all ASD. There were no residual shunts or stenoses observed after surgical repair in these patients.

Table 1 Baseline clinical and echocardiographic characteristics

Parameters	Total (n = 18)
Age (years)	47.6 ± 14.8
Gender (female/male)	8/10
Body mass index (kg/m ²)	25.2 ± 3.5
LVEDD (mm)	50.7 ± 6.5
LVESD (mm)	33.7 ± 7.1
Left ventricular ejection fraction (%)	61.0 ± 10.7
Left atrium anteroposterior diameter (mm)	36.5 ± 7.2
Coronary sinus dilatation	13 (72.2)
Congenital heart disease	3 (16.7)

Data are expressed as the mean ± standard deviation or as a number (%). LVEDD, left ventricular end-diastolic dimension; LVESD, left ventricular end-systolic dimension.

Table 2 Proportion of PLSVC in patients with SVA

Type of arrhythmia	Number of patients	Number of PLSVC patients	Proportion of PLSVC in SVA patients (%)
AVNRT	2380	7	0.29
AVRT	2724	4	0.15
Typical atrial flutter	499	2	0.40
Atrial tachycardia	239	2	0.84
Junctional tachycardia	23	1	4.35
Atrial fibrillation	734	2	0.27
Total	6662	18	0.27

PLSVC, persistent left superior vena cava; SVA, supraventricular tachyarrhythmia; AVNRT, atrioventricular nodal reentrant tachyarrhythmia; AVRT, atrioventricular reentrant tachycardia.

The mean LV EF was 61.0 ± 10.7%. The mean LA anteroposterior diameter measured by echocardiography was 36.5 ± 7.2 mm. Dilated CS was found in 13 patients (72.2%) by two-dimensional TTE.

ECG examination during clinical tachycardia revealed narrow QRS tachycardia in 10 patients, wide QRS tachycardia in 1 patient, ventricular pre-excitation in 2 patients, AFL in 2 patients, and AF in 2 patients. One patient had paroxysmal palpitations without documented SVA.

Types of supraventricular tachyarrhythmia and proportion of persistent left superior vena cava

SVA diagnosis was confirmed by an EP study. Among the 18 SVA patients with PLSVC, 7 had AVNRT, 4 had AVRT, 2 had typical AFL, 1 had AT from septal macroentry, 1 had AT from the PLSVC, 1 had JT, and 2 had AF. The proportion of PLSVC in SVA patients was 0.27% (18/6662) during the 10-year study period. *Table 2* shows the detailed proportion of PLSVC in patients with SVA.

Catheter ablation and follow-up results

Catheter ablation was tried in all but one patient. The patient number (No.), types of SVA, and procedures performed in individual patients are listed in *Table 3*.

Atrioventricular nodal reentrant tachycardia

Six patients (Patient Nos. 1–6) exhibited narrow QRS tachycardia on ECG, and EP studies revealed typical AVNRT. The mean cycle length was 405.7 ± 69.2 ms. Successful slow pathway modification was performed in these six patients. The mean procedure time was 92.5 ± 30.8 min and the mean fluoroscopy time was 21.4 ± 14 min. Patient No. 18, who had an operation for congenital heart disease (double outlet right ventricle, subaortic ventricular septal defect, pulmonary stenosis, and ASD), presented with a wide QRS tachycardia on ECG, and an EP study revealed typical AVNRT with bundle branch block. The ablation of AVNRT in this patient was unsuccessful due to difficulty in mapping the triangle of Koch. After 4 months, the patient visited the emergency room due to typical AFL. Ablation of AFL was successful.

Atrioventricular reentrant tachycardia

The EP studies of two patients with narrow QRS tachycardia (Patient Nos. 7 and 8) and two patients with ventricular pre-excitation (Patient Nos. 9 and 10) revealed AVRT. The mean cycle length was 310.0 ± 99.6 ms. Successful ablation of the left lateral accessory pathway was performed in three patients. Patient No. 7 exhibited a left posterolateral bypass tract and underwent a failed catheter ablation. In this patient, the irrigation catheter was not used by the operator at the time of the procedure and it was the main reason for failure. The mean procedure time was 83 ± 28.7 min and the mean fluoroscopy time was 17.9 ± 13.1 min.

Typical atrial flutter

Two patients exhibited a typical AFL and underwent successful ablation at the cavotricuspid isthmus (CTI) (Patient Nos. 11 and 12). The tachycardia cycle length was 300 and 220 ms. Patient No. 11 had undergone a previous ASD patch closure operation. The procedure time was 180 and 100 min, respectively, and the fluoroscopy time was 35 and 18.4 min, respectively.

Atrial tachycardia

Patient No. 13, who had a corrective operation for ASD with cor triatriatum, exhibited AT. The EP study revealed that the mechanism was septal macroentry around the septal patch. The tachycardia cycle length was 285 ms. Linear ablation at the RA posterior septum was performed using an irrigation catheter. The procedure time was 210 min and the fluoroscopy time was 69.3 min. Patient No. 14 received an EP study due to paroxysmal palpitations without documented arrhythmia. In the EP study, the existence of the PLSVC was confirmed and an AT from the PLSVC was noted. The cycle length was 365 ms. We did not ablate the PLSVC because there was an unclear correlation between the induced arrhythmia and palpitations.

Table 3 Electrophysiological characteristics and procedural outcomes of individual patients

Patient no.	Sex	Age	Structural heart disease	Tachycardia mechanism	Tachycardia cycle length	Arrhythmogenicity of PLSVC	Procedure	Success	Recurrence	Follow-up duration (months)
1	F	52		AVNRT, typical	360	No	SP modification	Yes	No	5
2	M	46		AVNRT, typical	380	No	SP modification	Yes	No	24
3	M	75		AVNRT, typical	295	No	SP modification	Yes	No	12
4	F	66		AVNRT, typical	385	No	SP modification	Yes	No	1
5	F	47		AVNRT, typical	465	Yes	SP modification	Yes	No	29
6	F	23		AVNRT, typical	430	No	SP modification	Yes	No	18
7	M	65		AVRT	305	No	AP ablation	No	No	7
8	F	42		AVRT	450	No	AP ablation	Yes	No	6
9	M	50		AVRT	265	No	AP ablation	Yes	No	36
10	M	23		AVRT	220	Yes	AP ablation	Yes	No	36
11	M	26	ASD closure op.	AFL, typical	300	No	CTI ablation	Yes	No	60
12	M	56		AFL, typical	220	No	CTI ablation	Yes	No	4
13	F	66	ASD & Cor triatriatum correction op.	Septal macroreentry	285	No	RA septum ablation	Yes	No	12
14	F	40		AT from PLSVC	365	Yes	EPS only		No	12
15	F	37		JT	545	Yes	Focus ablation	No	Yes	56
16	M	47		AF		No	PV isolation CTI ablation	Yes	No	36
17	M	59		AF		Yes	PV isolation CFAE in CS ablation	Yes	Yes (AFL)	3
18	M	37	Correction op. for DORV, subaortic VSD, PS, ASD. Redo ASD closure	AVNRT, typical	505	No	SP modification	No	Yes (AFL)	8

AVNRT, atrioventricular nodal reentrant tachycardia; SP, slow pathway; AVRT, atrioventricular reentrant tachycardia; AP, accessory pathway; AFL, atrial flutter; CTI, cavotricuspid isthmus; ASD, atrial septal defect; RA, right atrium; AT, atrial tachycardia; PLSVC, persistent left superior vena cava; EPS, electrophysiological study; JT, junctional tachycardia; AF, atrial fibrillation; PV, pulmonary veins; CFAE, complex fractionated atrial electrogram; CS, coronary sinus; DORV, double outlet right ventricle; VSD, ventricular septal defect; PS, pulmonic stenosis.

Junctional tachycardia

Patient No. 15 exhibited JT from an anterior high septal site. The cycle length was 545 ms. RF catheter ablation of this patient was unsuccessful.

Atrial fibrillation

There were two patients with AF. Patient No. 16 successfully underwent pulmonary vein isolation with CTI ablation. The procedure time was 445 min and the fluoroscopy time was 116 min. Patient No. 17 was a 59-year-old man who exhibited paroxysmal AF. During an EP study after pulmonary vein isolation, complex fractionated electrograms were observed at the PLSVC, and a three-dimensional electroanatomical map demonstrated that the PLSVC acted as an arrhythmogenic substrate for AF. After focal ablation at the site at which fragmented potentials were observed, AF changed to a typical AFL. Additional ablation of the AFL was successfully executed. The procedure time in this patient was 328 min, and fluoroscopy time, 72 min. However, after 3 months, AT had occurred and another EP study exhibited reentrant AFL at the mitral isthmus. At this time, PLSVC was not directly involved in the pathogenesis of reentrant

AFL, and was activated from the entrance site at a proximal part that faced the LA. Thus, in this patient, the PLSVC presented as an arrhythmogenic substrate of paroxysmal AF and a bystander of AT.

Ectopic potentials from the persistent left superior vena cava

Five of 18 patients exhibited ectopic potentials and one patient among them showed arrhythmogenicity at the PLSVC. Patient No. 5, who had an AVNRT, showed atrial premature complex arising from the PLSVC. In Patient No. 10, who had an AVRT, AT was induced from the PLSVC after isoproterenol infusion. Fractionated potentials with decremental property under atrial pacing were seen at the PLSVC (Figure 1). Nevertheless, we did not treat the AT considering the risk of complication. During the follow-up period of 36 months, the patient had no recurrence of SVA. Patient No. 14 showed AT from the PLSVC in the EP study. We did not ablate the AT from PLSVC as described above. The patient did not complain of further palpitations following treatment with a low dosage of a beta-blocker. Patient No. 15, who had JT, showed AT from the PLSVC, but ablation at the PLSVC was not attempted. In Patient No. 17, who exhibited

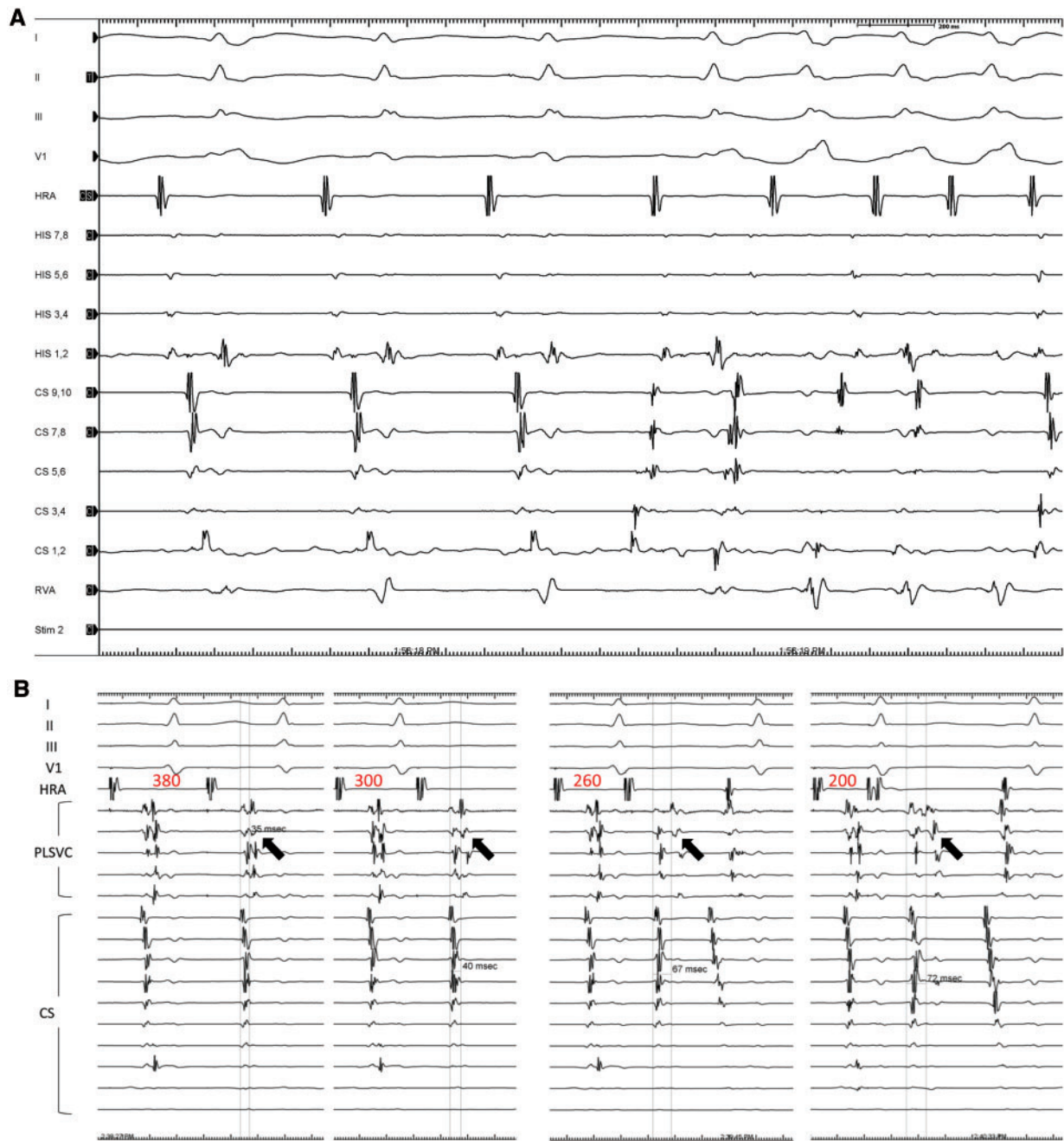


Figure 1 Cardiac tracings from a 23-year-old man with WPW syndrome. (A) AT was induced from PLSVC after isoproterenol infusion. Cardiac tracings are displayed from top to bottom: surface electrograms (I, II, III, and V1), HRA, proximal to distal His bundle electrograms (His 7–8, 5–6, 3–4, 1–2), unipolar electrograms of the proximal to distal CS (designated as CS 9–10, 7–8, 5–6, 3–4, 1–2), and electrograms from the RVA. (B) Response of PLSVC potentials (arrows) upon atrial extrastimulation pacing. With an extrastimulus coupling interval decremented from 380 to 200 ms, decremental property of the PLSVC could be demonstrated. Cardiac tracings are displayed from top to bottom: surface electrograms (I, II, III, and V1), HRA, unipolar electrograms of the PLSVC, unipolar electrograms of the proximal to distal CS and electrograms from the RVA. WPW, Wolff–Parkinson–White; AT, atrial tachycardia. PLSVC, persistent left superior vena cava; HRA, high right atrial electrogram; CS, coronary sinus; RVA, right ventricular apex.

AF, the PLSVC showed complex fragmented potential. Ablation at these foci resulted in organization of AF into AFL.

Procedural outcomes

A total of 17 RF catheter ablations were performed and 14 procedures were successful (Patient No. 14 only received an EP study). An attempt to perform catheter ablation on three patients failed to ameliorate the AVNRT of Patient No. 18, AVRT of Patient No. 7, and JT of Patient No. 15. The procedural success rate was 82% (14/17). There were no procedure-related complications.

In patients with successful ablation of SVA, there was no recurrence of atrial arrhythmias during the median follow-up duration of 18 months (1–60 months).

Discussion

Major findings

The major findings of this study are the following: First, PLSVC was observed in 0.27% of adult SVA patients who underwent EP studies or RF catheter ablations during the 10-year study period. This incidence rate is similar to that seen in the general population. Second, repaired congenital heart disease was present in a relatively small portion (3 of 18, 16.7%) of patients. Third, the most common type of SVA was AVNRT, which is consistent with results of previous reports. Fourth, ectopy from the PLSVC was found in 5 of 18 patients.

Proportion of persistent left superior vena cava in supraventricular tachyarrhythmia patients

In our report, 6662 adult SVA patients underwent an EP or RF catheter ablation procedure in the Yeungnam region of Korea during the 10-year study period. In this period, PLSVC was observed in 18 SVA patients. Therefore, the prevalence of PLSVC in our SVA population is 0.27%. This is similar to the prevalence of PLSVC in the general population: between 0.3 and 2% in individuals with a normal heart and 4.5–9% in those with congenital heart defects.^{1,3,19} Furthermore, this trend was maintained in each subtype of SVA as the rate of PLSVC was 0.29% in AVNRT, 0.15%, in AVRT, 0.84% in AT, 0.4% in AFL, and 0.27% in AF. Unlike other arrhythmias, PLSVC was observed in 4.35% of JT patients. However, this may be due to the small total number of JT patients.

Recently, Ouchi *et al.*²⁰ reported that the incidence of PLSVC was 0.27% (19 out of 6936 patients) in adult patients who underwent cardiac CT images for cardiac diseases or preprocedural planning. This reported incidence rate exactly matches with that of our study results.

Embryologic/anatomic considerations of persistent left superior vena cava

Embryologically, PLSVC results from the abnormal development of the sinus venosus during foetal life. Usually, the proximal portion of the left sinus horn persists as the CS, whereas the distal left horn and the left cardinal vein obliterate, thereby forming the ligament of Marshall in the adult. When the left cardinal vein persists, it becomes the PLSVC and generally drains into the RA through an enlarged

CS.^{2–4} A PLSVC is usually suspected when an unusually large CS on two-dimensional TTE views is observed (appears as dilated, echo-free space posteriorly in the AV groove between the LA and left ventricle).²¹ In our study, 13 of the 18 PLSVC patients (72%) exhibited CS dilatation on preprocedural TTE. Thus, echocardiography can be an appropriate method for screening PLSVC patients in catheter ablation candidates. In addition, PLSVC can be suspected when the catheter for recording for CS electrogram is positioned in an unconventional manner (directed to the left main bronchus rather than anterior basal left ventricle). Therefore, if the conditions described above are met, it is recommended to look for the presence of PLSVC before or during the procedure.

Clinical impact of persistent left superior vena cava in supraventricular tachyarrhythmias

Distorted CS anatomy due to PLSVC can be an obstacle to treating AVNRT and AVRT. Okishige *et al.*¹² reported the difficulty of RF ablation for AVNRT associated with PLSVC. The authors described that the patients with PLSVC have significantly enlarged CS ostia, and therefore, the course of the slow pathway may be displaced. The His bundle may also be displaced, increasing the risk of damaging the AV node during an ablation of the slow pathway. Moreover, the contact of the ablation catheter may have been insufficient.¹⁵ In AVRT, a CS electrode catheter cannot be used for guidance during ablation of the left-sided accessory pathway and may not allow for a stable electrode position at an endocardial target site.^{10,14}

Despite these difficulties, successful catheter ablation of AVNRT and AVRT was performed in previous case reports.^{5,13,14,22–24} Our study also showed an acceptable ablation success rate (82%, 9/11). Our two failed patients had additional reasons for failure: the surgery of congenital heart disease contributed to the failure in the AVNRT patient, and inappropriate catheter selection was responsible in the failed AVRT patient. Therefore, catheter ablation can be a reasonable method in the treatment of AVNRT and AVRT patients with PLSVC.

In our study, five patients showed ectopic potentials from the PLSVC, and one patient among them who demonstrated PLSVC acted as an arrhythmogenic substrate for AF. It has previously been well described that the major thoracic veins have an established role in the genesis and maintenance of AF.^{25,26} These include the vein and ligament of Marshall, which are the developmental remnants of the embryonic left superior vena cava.²⁷ Furthermore, PLSVC has been demonstrated to be an arrhythmogenic source of paroxysmal or persistent AF in patients.¹⁶ In our study, one patient exhibited arrhythmogenic potentials from PLSVC, which induced AF, and this patient successfully received RF catheter ablation at the PLSVC. Four patients showed ectopy from the PLSVC. Ablation was not performed due to the uncertain role of these electric potentials. During the follow-up period, SVA recurrence due to the PLSVC did not occur in these patients.

Study limitations

There are several limitations to our study. First, this is a retrospective study, which has inherent limitations. Second, because of the relatively small number of study subjects, we could not perform a subgroup analysis for each SVA. Third, a pharmacologic provocation test

using multiple drugs or a high dose infusion of isoproterenol to evaluate arrhythmogenic property was not conducted in all patients. If those maneuvers were performed, arrhythmogenic property of the PLSVC could have been determined in more patients. Fourth, since venography was not routinely performed in all patients, the incidence of PLSVC in SVA patients who underwent EP studies or RF catheter ablations might not be true incidences. Also, the classification of the subtype of PLSVC could not be assessed.

Conclusions

In our study, the proportion of adult SVA patients with a PLSVC who underwent an EP study and RF catheter ablation procedure was 0.27% during the 10-year study period. The most common type of SVA was AVNRT. The success rate of catheter ablation in SVA patients with PLSVC was 82%. There were no complications related to the procedure. In patients with successful ablation, the recurrence of SVA was not observed during the median follow-up period of 18 months. In some patients, electrical potentials of the PLSVC were demonstrated.

Supplementary material

Supplementary material is available at *Europace* online.

Conflict of interest: none declared.

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