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Original article

Permanent form of junctional reciprocating tachycardia in adults: peculiar features and results of radiofrequency catheter ablation

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KEYWORDS
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Aim The aim of this study is to describe the clinical presentation, electrophysiological characteristics, feasibility and safety of radiofrequency ablation, and the long-term prognosis in a large group of adult patients with the permanent form of junctional reciprocating tachycardia (PJRT). PJRT occurs predominantly in infants and children and are limited to small series in adults.

Methods and results Forty-nine adult patients (22 male and 27 female; mean age 43 ± 16) with a diagnosis of PJRT confirmed at electrophysiological study were included. Eight patients (16%) presented with tachycardia-induced cardiomyopathy (TIC). Ventricular rate was 146 ± 30 b.p.m. The arrhythmia was permanent or incessant in 23/49 cases (47%) and paroxysmal in 26/49 (53%). A significant correlation was found between symptom duration and tachycardia rate ($r^2 = 0.12$, $P = 0.01$). The accessory pathway (AP) was located in the right posteroseptal region in 37 cases (76%) and in atypical sites in 12 cases (24%).Patients with the incessant or permanent form of JRT had longer duration of symptoms, more frequently TIC and a slower tachycardia rate. Radiofrequency catheter ablation was initially successful in 46 cases (94%) without any serious complication. Long-term success rate was 100% (49/49 patients) in the absence of any antiarrhythmic drug treatment (mean follow-up 49 ± 38 months). Regression of TIC was observed in all cases (8/8).

Conclusion PJRT in adults is often paroxysmal (53%), and the retrograde slowly conducting, decremental AP is not infrequent in a non-posteroseptal location. Radiofrequency catheter ablation is highly effective and should be considered as the treatment of first choice in adult patients with PJRT.

Introduction

Permanent junctional reciprocating tachycardia (PJRT) is a rare form of nearly incessant supraventricular tachycardia occurring predominantly in infants and children and characterized by a long RP′ interval and, in the typical form, by negative P-waves in leads II, III, and aVF on the surface ECG. During sinus rhythm, the surface ECG is normal, without manifest pre-excitation. PJRT is caused by an atrioventricular (AV) re-entry using the AV node as the antegrade limb and a slowly conducting accessory pathway (AP) as the retrograde limb.[1–3] The location of the AP is commonly right posteroseptal with an atrial insertion close to the ostium of the coronary sinus, but other locations have been reported.[4–7] Chronic uncontrolled tachycardia has been reported to result in tachycardia-induced cardiomyopathy (TIC), which usually recovers with adequate ventricular rate control.[8,9] As PJRT is most of the time refractory to drug therapy, radiofrequency catheter ablation of the AP has become the treatment of first choice.[10–13]

The aim of the present multicentre retrospective study, the largest so far published, is to describe the clinical presentation of PJRT in an adult population, with special interest focused on the characteristics of the AP, on the feasibility and safety of radiofrequency ablation, and on the long-term follow-up for arrhythmia recurrence and improvement in left ventricular systolic function.

Methods

Patients (*Table 1*)

This retrospective study included 49 patients (27 female and 22 male) with a mean age of 43 ± 16 (range 18–72 years), referred

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Table 1 Clinical features, radiofrequency catheter ablation characteristics and follow-up data of the study population

Patient number	Gender	Age (years)	Symptoms duration (months)	Tachycardia induced DCM	Type of PJRT	Tachycardia CL (ms)	Successful ablation site	Procedure time (min)	Fluoroscopy time (min)	Follow-up duration (months)	Recurrences	Successful second RF ablation
1	F	45	400	0	I	480	Right PS	120	21	120	0	
2	M	59	360	0	I	490	Right PS	300	48	100	0	
3	M	49	240	0	I	450	Right PS	155	27	86	1	1
4	M	54	120	0	I	380	Mid-septal	130	52	84	0	
5	M	21	144	0	I	580	Right PS	80	13	38	0	
6	M	49	360	1	I	590	Right PS-MCV	140	74	5	1	1
7	F	24	36	0	I	540	Right PS	120	12	24	0	
8	F	54	300	1	I	440	Left posterolateral	170	30	48	0	
9	M	18	96	0	P	300	Left posterior	105	16	60	0	
10	F	18	12	0	P	428	Right PS	45	8	120	0	
11	F	41	2	1	P	340	Right PS	150	22	48	0	
12	M	51	120	0	P	410	Left lateral	180	24	48	0	
13	M	35	300	0	P	320	Left anterior	210	70	10	0	
14	F	45	12	0	I	428	Right PS	250	89	30	1	1
15	F	72	18	0	P	430	Left posteroseptal	245	70	36	0	
16	F	57	24	0	P	338	Right PS	200	38	40	0	
17	M	54	9	0	P	476	Right PS	100	8	5	0	
18	F	37	2	0	I	500	Right PS	100	7	40	0	
19	M	33	6	0	P	320	Mid-septal	110	13	36	0	
20	F	20	192	0	P	340	Right PS-MCV	230	45	30	0	
21	M	47	240	0	P	417	Right PS	110	18	30	1	1
22	F	26	48	0	P	375	Mid-septal	155	30	20	1	1
23	F	66	180	0	P	500	Right PS	70	5	36	0	
24	F	62	36	0	P	360	Left anterior	165	35	24	0	
25	F	27	18	0	P	340	Right PS	135	18	15	0	
26	F	63	480	0	P	420	Right PS-MCV	180	35	13	0	
27	M	72	360	0	P	428	Right PS	120	8	13	1	1
28	F	63	1	0	P	335	Right PS	150	10	48	0	
29	F	26	24	0	P	380	Right PS	125	12	15	0	
30	F	78	120	1	I	400	Right PS	65	13	72	0	
31	M	29	120	1	I	440	Right PS	N/A	N/A	110	1	1
32	F	31	13	0	P	280	Right PS	260	63	4	0	
33	M	57	220	0	P	340	Left posteroseptal	165	42	96	0	
34	F	38	12	0	I	430	Left anterolateral	235	65	110	0	
35	M	37	20	0	I	410	Right PS	152	46	132	0	
36	M	62	132	1	I	500	Right PS	165	34	132	0	
37	F	51	4	0	P	310	Left posterolateral	210	30	42	0	
38	F	18	120	0	I	450	Right PS	125	7	60	0	
39	M	44	36	0	I	260	Right PS	180	16	66	0	
40	M	30	240	0	I	340	Right PS	60	15	72	0	
41	F	48	120	0	I	520	Right PS	60	18	10	0	
42	M	58	498	1	I	400	Right PS	60	10	6	0	

Continued

Table 1 *Continued*

Patient number	Gender	Age (years)	Symptoms duration (months)	Tachycardia induced DCM	Type of PJRT	Tachycardia CL (ms)	Successful ablation site	Procedure time (min)	Fluoroscopy time (min)	Follow-up duration (months)	Recurrences	Successful second RF ablation
43	M	37	240	0	I	380	Right PS	90	N/A	30	0	
44	F	25	24	0	P	600	Right PS	70	N/A	30	0	
45	M	47	240	0	P	400	Right PS	60	N/A	13	0	
46	M	27	180	0	I	380	Right PS	60	12	1	0	
47	F	36	360	1	I	460	Right PS	60	10	72	0	
48	F	27	300	0	I	420	Right PS	45	8	8	0	
49	F	35	360	0	I	380	Right PS	120	25	102	0	

M, male; F, female; DCM, dilated cardiomyopathy; CL, cycle length; PS, posteroseptal; MCV, mid-cardiac vein; N/A, not available.

for radiofrequency catheter ablation in six electrophysiological referral centres. Because the aim of the present study is to focus on PJRT in an adult population, infants, children, and adolescents are excluded from analysis.

The diagnosis of PJRT was based on typical surface ECG criteria (long RP′ interval, RP′ greater than P′R, 1:1 AV ratio, and inverted P′ wave in leads II, III, and aVF during tachycardia) (*Figure 1*) and on specific electrophysiological characteristics [exclusion of an atrial tachycardia by demonstrating that single ventricular extrastimuli introduced during tachycardia can reproducibly terminate the tachycardia without activating the atria; exclusion of an atypical form of AV nodal re-entrant tachycardia (fast–slow or slow–slow) by demonstrating the ability to pre-excite the atria with single ventricular extrastimuli applied during tachycardia at a time when the His bundle is refractory]. Only electrophysiological criteria were used when the AP was not typically located in the right posteroseptal region.

Electrophysiological testing and radiofrequency catheter ablation procedure

The procedure was performed in the fasting state under local anaesthesia with pre-medication with lorazepam (1–2.5 mg) in selected cases. All antiarrhythmic medications were discontinued at least 72 h before the procedure. Two to four multipolar electrode catheters (4F, 6F, or 7F) were inserted percutaneously through the femoral vein and positioned in the high right atrium, His bundle position, coronary sinus, and right ventricle. For left-sided APs, an additional 7F sheath was introduced into the femoral artery, and the ablation procedure was performed using a retrograde transaortic approach. The diagnostic electrophysiological study was performed using standard techniques (multichannel digital recorder; analysis at a paper speed of 100 or 200 mm/s, with filter settings of 0.5–1000 Hz for the surface ECG and 0.5–500 Hz for intracardiac recordings; stimulation using 1-ms-duration pulses at twice diastolic threshold). Mapping and radiofrequency catheter ablation were performed using a 7F quadripolar temperature-controlled electrode catheter with a 4-mm tip and a deflectable curve. Location of the AP was assessed by intracardiac mapping and successful ablation on the basis of the shortest VA interval during tachycardia. Accessory pathway locations were classified as right anteroseptal, para-Hisian, right lateral, mid-septal, right posterolateral, left posteroseptal, left posterolateral, left lateral, left anterolateral, and left anterior. Radiofrequency current was delivered from the tip on a 7F deflectable temperature-controlled ablation catheter with a standard 4-mm tip; the radiofrequency generator was set so as to achieve a temperature of 50–65°C and a power of 45–55 W. A 1000–3000 IU bolus of heparin was given intravenously after catheter placement, followed by 1000 IU/h thereafter. Atropine or isoprenaline was used in selected cases. On completion of the procedure, all catheters and sheaths were removed and manual compression was maintained to achieve complete haemostasis. All patients received aspirin 100 mg/day for 4 weeks after the procedure.

Follow-up

A clinical follow-up evaluation was performed by the attending physician 2–3 weeks after the procedure and then at 6-month intervals. Effectiveness of radiofrequency catheter ablation was established by history and 24-h Holter recording to exclude recurrences of PJRT; echocardiography was used to assess recovery of systolic left ventricular function.

Statistical analysis

Values are expressedas mean $\pm$1 SD and $\pm$SEM when appropriate.- Differences in continuous variables between groups were analysed using Student's *t*-test. Differences in categorical data were performed using χ^2 analysis. Pearson's correlation and

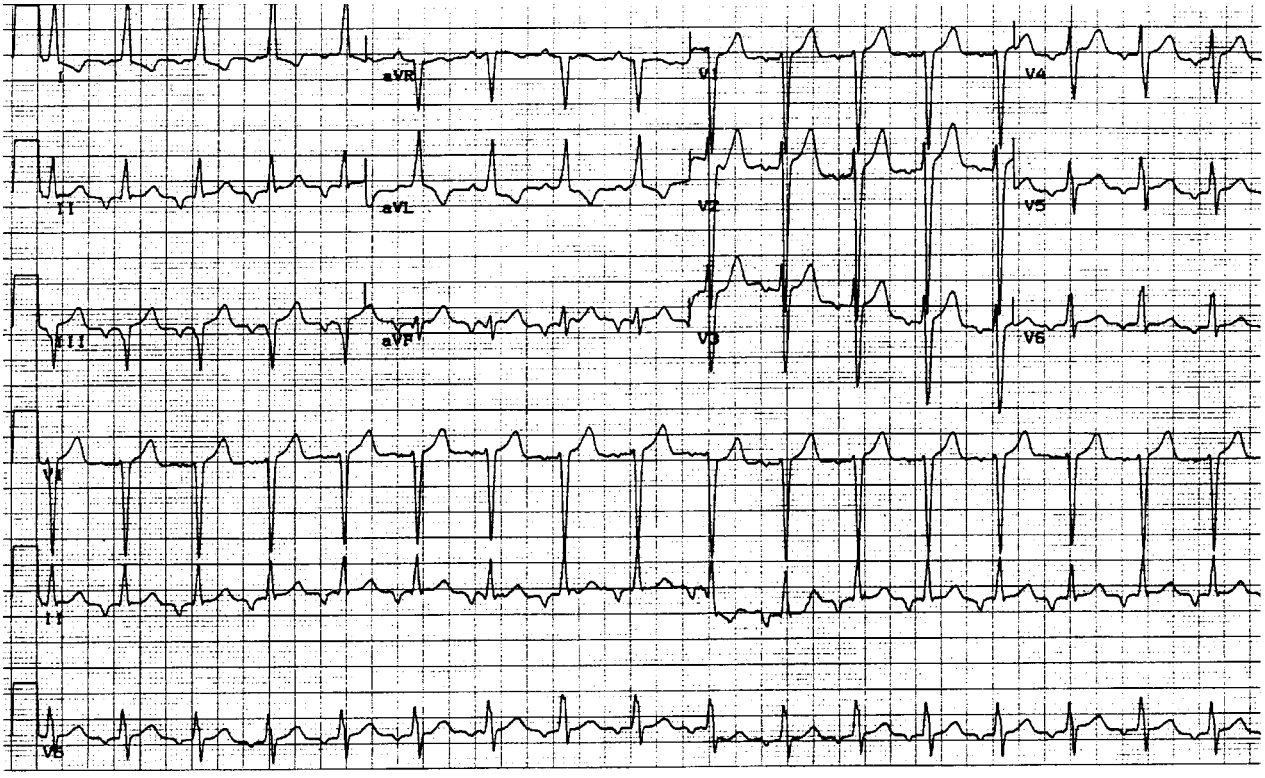

Figure 1 12-lead ECG from Patient 5 showing a narrow QRS complex tachycardia at a rate of 110 b.p.m., with long RP interval, a negative P-wave in leads II, III, and aVF, and a positive P-wave in lead aVL.

linear-regression analysis were performed between tachycardia cycle length and age, duration of symptoms, and AH intervals. A *P*-value less than 0.05 was considered statistically significant.

Results

Patients characteristics (*Table 1*)

The mean age of the study population was 43 ± 16 years with a range from 18 to 72 years. Eleven patients (22%) had an associated cardiovascular abnormality: dilated cardiomyopathy, presumed to be related to the rapid ventricular rate, was present in eight (14%), coronary artery disease in two (4%), and sick sinus syndrome in one (2%). Forty-five patients (92%) complained of palpitations and six of malaise or dizziness (12%), but none had experienced syncope. Symptoms have been present for a median of 10 years (mean 153 ± 144 months, range 1–498 months). Ventricular rate during tachycardia was 146 ± 30 b.p.m. (range 100–240 b.p.m.). No patient had manifest pre-excitation during sinus rhythm on the 12-lead resting ECG. In 23/49 cases (47%), the arrhythmia was permanent or incessant, but in 26/49 (53%), the arrhythmia was paroxysmal with prolonged periods of normal sinus rhythm. Twenty patients (41%) never received any specific antiarrhythmic treatment; in the remaining 29 patients, a mean of 1.9 ± 1.3 drug trial has been attempted without success (beta-blocker 24; verapamil 13; digoxin 7; Class Ic 9; amiodarone 4).

Electrophysiological characteristics (*Table 1*)

AV re-entrant tachycardia was inducible in all patients during electrophysiological study. The diagnosis of PJRT was confirmed in all cases on the basis of predefined criteria (Methods). The mean cycle length of the induced tachycardia was 407 ± 89 ms. There was a significant correlation between symptom duration and tachycardia cycle length ($r^2 = 0.12$, $P = 0.01$) but not between tachycardia cycle length and age ($r^2 = 0.001$, $P = 0.79$) or tachycardia cycle length and AH interval ($r^2 = 0.02$, $P = 0.29$). During sinus rhythm, the mean AH interval was 81 ± 22 ms, and the mean HV interval was 46 ± 10 ms. Although difficult to assess properly in several patients because of immediate tachycardia induction, the effective refractory period of the retrogradely conducting AP was 293 ± 42 ms. Manifest ventricular pre-excitation was never demonstrated either during sinus rhythm or during programmed stimulation. A typical right posteroseptal location of the AP was present in 37 cases (76%). In 12 cases (24%), the location of the decremental slow conducting AP was atypical: mid-septal in three (6%), left posteroseptal in three (6%), left posterolateral in two (4%), left lateral in one (2%), left anterolateral in one (2%), and left anterior in two (4%). In three cases (6%), multiple APs were present (right posteroseptal + left lateral in one; left anterior + left lateral in one; right posteroseptal + right mid-septal in one). In one patient, slow–slow AVNRT was also present, and in one patient, atrial flutter and atrial fibrillation were induced during the electrophysiological evaluation.

Patients with the incessant or permanent form of PJRT had a longer duration of symptoms, more frequently TIC, and a longer tachycardia cycle length, but AH interval was not statistically different between the two groups (*Table 2*).

Patients with TIC showed no statistical difference when compared with patients without cardiomyopathy, but they

Table 2 Comparison of patients with permanent or incessant PJRT and patients with paroxysmal PJRT

	Permanent or incessant PJRT ($n = 23$)	Paroxysmal PJRT ($n = 26$)	*P*-value
Male/female	11/12	11/15	0.15
Mean age (years)	42 ± 14	43 ± 18	0.85
Duration of symptoms (months)	197 ± 149	114 ± 130	0.04*
TIC	7/23	1/26	0.001*
Tachycardia cycle length (ms)	459 ± 72	372 ± 61	0.0001*
AH interval (ms) during SR	82 ± 22	81 ± 24	0.87
Posteroseptal location of the AP	18/23	19/26	0.67
Recurrences after RF ablation	4/23	3/26	0.55

RF, radiofrequency; SR, sinus rhythm.

Table 3 Comparison of patients with and without tachycardia-induced cardiomyopathy

	TIC present ($n = 8$)	TIC absent ($n = 41$)	*P*-value
Male/female	4/4	18/23	0.95
Mean age (years)	51 ± 16	41 ± 16	0.12
Duration of symptoms (months)	237 ± 117	137 ± 135	0.07
Tachycardia cycle length (ms)	444 ± 75	407 ± 79	0.22
Posteroseptal location of the AP	7/8	30/41	0.93
Recurrences after RF ablation	2/8	5/41	0.92

tended to be older and to have a longer duration of symptoms (*Table 3*).

Radiofrequency catheter ablation data

Mapping was performed using the shortest VA interval during tachycardia (*Figure 2*). Radiofrequency catheter ablation was initially successful in 46 cases (94%) with a mean of 7±7 radiofrequency current applications (range 1–32). The mean interval between radiofrequency current application and interruption of tachycardia was 4.1 ± 3.8 s (*Figure 3*). The mean power delivered was 43 ± 14 W, and the mean duration of radiofrequency current application was 70 ± 25 s. The mean procedure duration was 137 ± 63 min, and the mean fluoroscopy time was 28 ± 21 min. *Figure 4* shows the site of successful radiofrequency catheter ablation for each of the 49 patients included in this study. The three cases (6%) with initial failure (one right posteroseptal AP and two posteroseptal AP inside the coronary sinus) were successfully ablated during a second procedure. No serious complication was observed during the ablation procedure. Only one minor complication occurred (first degree AV block) without any clinical consequences.

Follow-up data

The mean follow-up was 49 ± 38 months. In four cases (8%), a recurrence of PJRT was observed during follow-up (mean delay between the ablation procedure and recurrence was 2.4 ± 2.1 months) and these four patients were successfully re-ablated in a second procedure. The long-term success rate was 100% (49/49 patients) in the absence of any anti-arrhythmic drug treatment. Regression of left ventricular systolic dysfunction was observed during follow-up in all cases (8/8).

Discussion

Clinical aspects

Classically, the permanent form of PJRT occurs in children or in young adults and is characterized by an incessant (sometimes permanent) supraventricular tachycardia. However, PJRT may be diagnosed at any age and can sometimes express itself as a paroxysmal form of supraventricular tachycardia with a long RP interval.[14,15] The present data in an adult population confirm that retrograde slowly conducting, decremental APs can present at any age. In this series, the paroxysmal form of PJRT was more common than previously reported,[14,15] and this difference may be due to different patient populations or to different diagnostic criteria (clinical vs. electrophysiological). In infants and children, PJRT is most often incessant or permanent as shown by Lindinger *et al.*[16] *Table 4* summarizes clinical and electrophysiological data of major series published on the subject. In the present series as in others,[14–16] there was a huge variability in tachycardia cycle length (from 260 to 600 ms) and tachycardia rate was significantly higher in patients with the paroxysmal form of PJRT when compared with patients with incessant or permanent tachycardia despite comparable AH intervals during sinus rhythm. However, there was no correlation between tachycardia rate and the presence of TIC, although the number of patients with this type of complication was too small to draw definite conclusions. TIC was observed in only eight cases (16%), and this figure is in accordance with previous observations.[14–16] This type of complication was more frequently observed in patients with the incessant/permanent form of PJRT than in patients with the paroxysmal form, and it appears that the duration of tachycardia more than the duration of the intrinsic rate is responsible for left ventricular dysfunction. The paroxysmal form of PJRT appears to be more frequent in adults when compared with children, and this feature may explain why the diagnosis of PJRT may remain unrecognized until adulthood.[9] Regression of left ventricular dysfunction after successful elimination of the AP was the rule in this series as in others.[9,14–18]

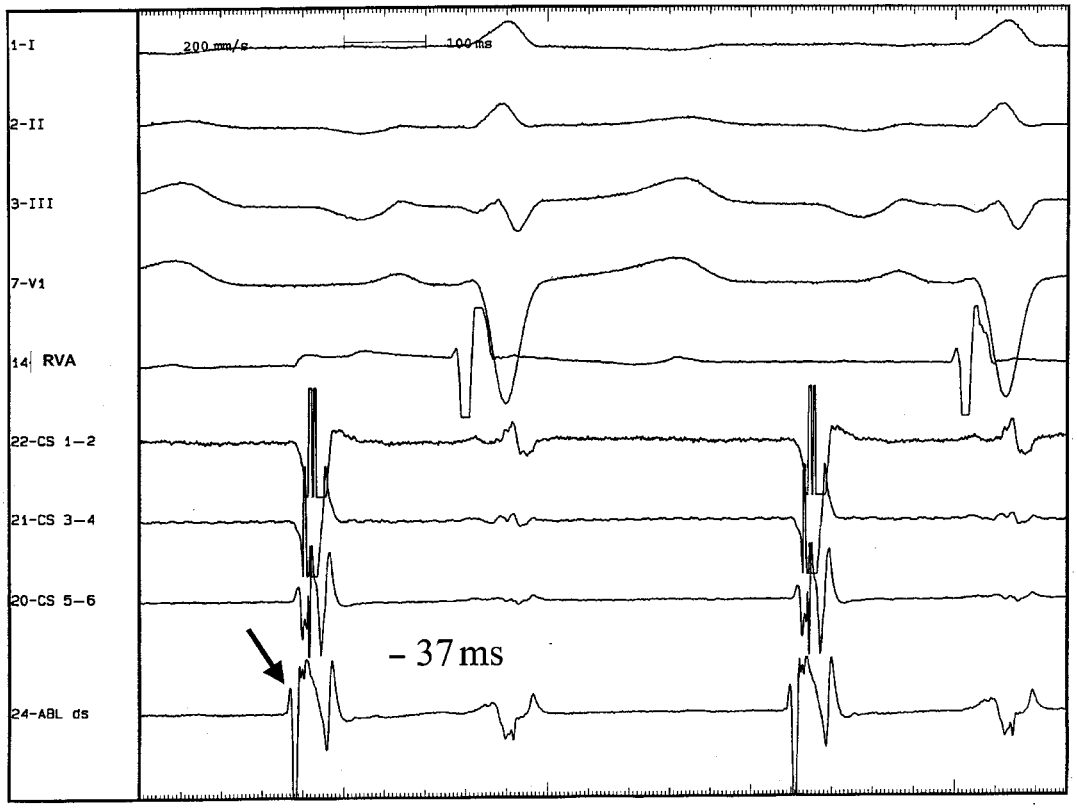

Figure 2 Successful ablation site in Patient 6: four surface ECG leads are displayed (I, II, III, and V1), together with five bipolar intracavitary recordings (RVA: right ventricular apex; CS 1–2, 3–4, and 5–6: coronary sinus distal to proximal; ABL ds: tip of the ablation catheter). Paper speed 100 mm/s. During tachycardia, the earliest retrograde activation was recorded at the lower margin of the CS os, and in that site, the local activation was 37 ms ahead of the onset of the P-wave on the surface ECG (arrow).

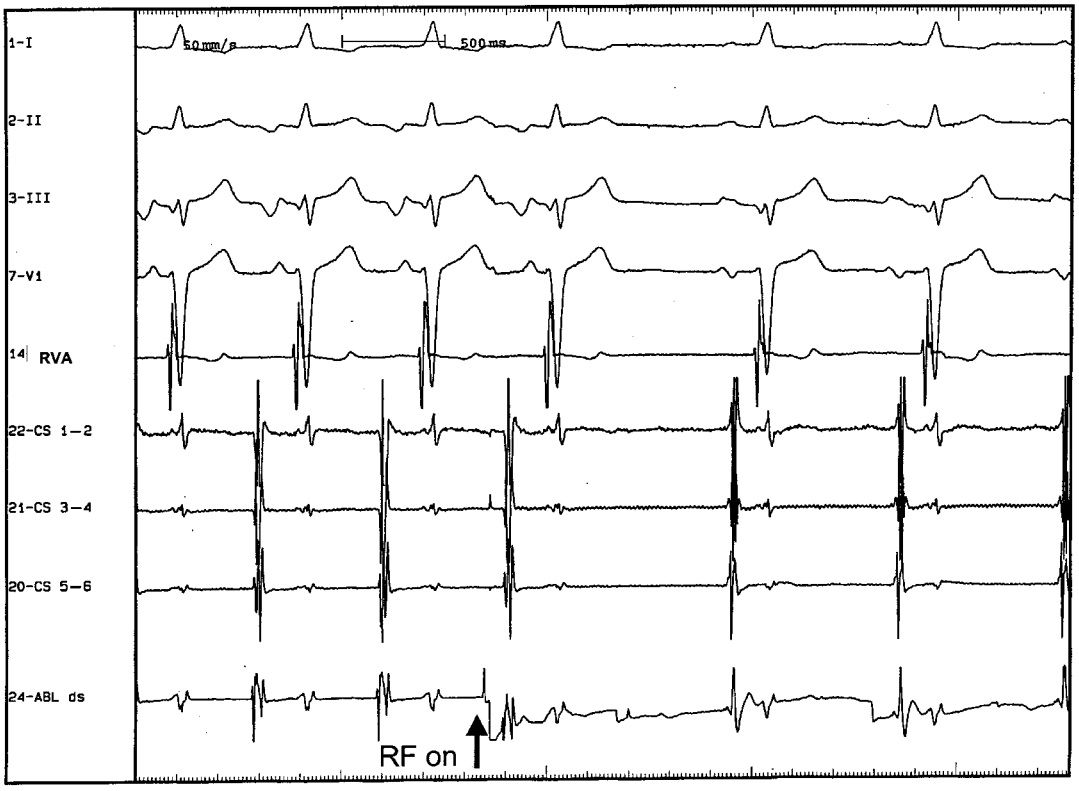

Figure 3 Same patient as in *Figure 2*. Radiofrequency current application (RF on, arrow) at the site of earliest activation immediately terminated the tachycardia, which could no longer be induced. Same abbreviations as in *Figure 2*.

Electrophysiological data

In the vast majority of PJRT cases, the retrograde slowly conducting, decremental AP is located in the posteroseptal region,[1,3,12,14–16] and therefore, catheter ablation is usually performed using a right-sided approach.[14] However, other locations of the AP have been described in 12–24% of cases,[7,14–15] and according to Gaïta *et al.*,[14] a non-posteroseptal location appears more frequently in patients with paroxysmal PJRT than in patients with incessant or permanent tachycardia. This observation was confirmed in the present study (27 vs. 22%), but the difference was not statistically significant. The AP in PJRT can be observed in almost every location from mid-septal to left anterior, even though most of the APs are located around or just within the ostium of the coronary sinus.[5,6,14]

Catheter ablation

In the present study as in others,[14,15,17,18] radiofrequency catheter ablation proved to be safe and highly effective for obtaining a definitive cure in patients with PJRT whatever the location of the AP (*Table 4*). Recurrences are not rare (from 13 to 23% in the literature; 8% in the present series), but long-term success is usually obtained after a second ablation procedure and ranged from 92 to 100%. The higher recurrence rate in PJRT compared with what is observed in non-decremental APs may be explained by the long, tortuous course of the AP along the AV sulcus.[3] Complication rate is low, but cautious application of radiofrequency current is mandatory when the AP is located above the ostium of the coronary sinus or in the mid-septal region, because the risk of second- or third-degree AV block is in the range of 5–7%. According to our data, we believe that radiofrequency catheter ablation should be considered as the treatment of first choice in adult patients with PJRT. The situation is quite different in infants and children, because antiarrhythmic drug treatment appears to be more effective in this age group and because complication rate of radiofrequency catheter ablation is inversely related to body weight.[16,19,20] Cryoablation, as recently reported by Gaïta *et al.*,[21] could represent a safe alternative in children by reducing the risk of AV block.

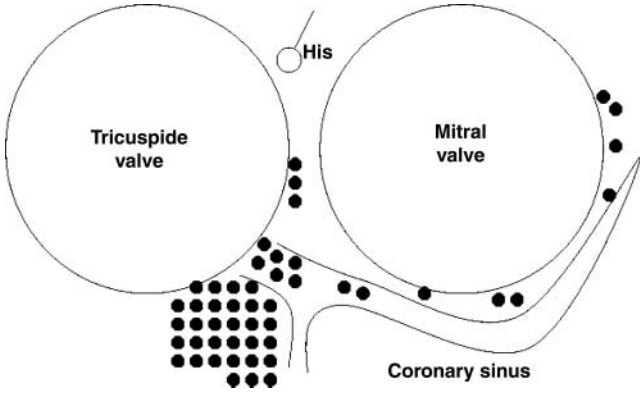

Figure 4 Schematic representation of successful radiofrequency catheter ablation site for each of 49 patients (60° left anterior oblique view).

Limitation of the study

This study is retrospective and observational, collecting data from six different institutions. However, great efforts have been made to fulfil criteria and to confirm the specific electrophysiological features of PJRT. The population is a selected one, all patients being referred for catheter ablation after unsuccessful pharmacological management.

Conclusion

PJRT in adults may have various clinical presentations and is often paroxysmal (53%), and the retrograde slowly conducting, decremental AP is not infrequently in a non-posteroseptal location. This study confirms that radiofrequency catheter ablation should be considered as the treatment of first choice in adult patients with PJRT for the following reasons: catheter ablation is highly successful,

Table 4 Comparison of published series of RF ablation for PJRT

	Present study	Gaïta[14]	Aguinaga[15]	Lindinger[16]
Number of patients	49	32	36	32
>18 years (%)	100% (49/49)	78% (25/32)	NA	3% (1/32)
Mean age (years)	43 ± 16	29 ± 15	44 ± 21	NA
Incessant/paroxysmal	23/26	25/7	23/13	29/3
TIC	8/49 (16%)	7/32 (22%)	7/36 (19%)	9/32 (28%)
Right posteroseptal location	37/49 (76%)	25/32 (76%)	32/36 (88%)	13/15 (87%)
Successful first RF ablation	46/49 (94%)	32/32 (100%)	35/36 (97%)	14/14 (100%)
Recurrences	4/49 (8%)	4/32 (13%)	8/35 (23%)	3/14 (21%)
Successful second RF ablation	7/7 (100%)	3/3 (100%)	5/8 (63%)	3/3 (100%)
Long-term success	49/49 (100%)	31/32 (97%)	33/36 (92%)	13/14 (93%)
Regression of LV dysfunction	8/8 (100%)	7/7 (100%)	7/7 (100%)	9/9 (100%)
Complication of RF ablation	1/49 (2%)	2/32 (6.3%)	2/36 (5.6%)	1/14 (7.1%)
Type of complication	First-degree AV block	Second- and third-degree AV block (transient)	Third-degree AV block (permanent, w/PM)	1sr + second-degree AV block

LV, left ventricular; AV, atrioventricular; w/PM, with pacemaker implantation; NA, not available.

the complication rate is low, and PJRT may lead to TIC, which is reversible.

Conflict of interest: none declared.

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