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Clinical Course and Electrophysiological Characteristics of Permanent Junctional Reciprocating Tachycardia in Children

ORIGINAL INVESTIGATION

ABSTRACT

Background: In this study, we aimed to evaluate the clinical aspects, electrophysiological studies, and ablation results of permanent junctional reciprocating tachycardia in children.

Methods: The study comprised 29 pediatric patients diagnosed with permanent junctional reciprocating tachycardia between 2011 and 2021 in 2 pediatric electrophysiology centers. From the file records, the basic demographic characteristics of the patients, as well as electrocardiographic and echocardiographic findings, were acquired retrospectively. The medical treatment and responses of the patients throughout follow-up, as well as the electrophysiological study and ablation data of the patients who had electrophysiological study, were assessed.

Results: The mean age at diagnosis of the patients was 3.13 ± 4.43 (0-18) years and the mean weight was 18.22 ± 19.68 (3.8-94) kg. Eighteen patients (62.1%) were girls. Eleven patients (38%) developed tachycardia-induced cardiomyopathy. Tachycardia was incessant in 15 patients (51.7%). In total, 22 patients required 26 ablation procedures. Tachycardia-induced cardiomyopathy and multidrug-resistant tachycardia were the most prevalent indications for ablation. The right posteroseptal pathway was detected in 18 patients (81.8%). The acute procedure success rate was 100% (22/22). The recurrence rate was 18% (4/22) and 3 of them underwent successful ablation again. The overall success percentage was 95.4% (21/22). None of the patients had any complications. The mean follow-up period was 4.39 ± 3.05 years.

Conclusion: Although permanent junctional reciprocating tachycardia is uncommon, it is often persistent, resistant to medical treatment, and associated with a substantial risk of tachycardia-induced cardiomyopathy. Catheter ablation can be performed on these patients at any age, with minimal risk of complications and a high success rate. It is crucial to keep monitor of the patients' recurrence.

Keywords: Arrhythmia, catheter ablation, permanent junctional reciprocating tachycardia, supraventricular tachycardia, tachycardia-induced cardiomyopathy

Yakup Ergül [1]
Ayşe Sulu [1]
Bahar Çaran [1]
Hasan Candaş Kafalı [1]
Celal Akdeniz [2]
Volkan Tuzcu [2]

[1]*Department of Pediatric Cardiology, University of Health Sciences, İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey*
[2]*Department of Pediatric Cardiology, University of Mediopol, İstanbul, Turkey*

Corresponding author:
Ayşe Sulu
✉ suluayse@windowslive.com

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INTRODUCTION

Permanent junctional reciprocating tachycardia (PJRT) is a rare supraventricular tachycardia (SVT) caused by atrioventricular reentry, with the atrioventricular (AV) node representing the antegrade pathway and the slow-conducting accessory pathway as the retrograde pathway.[1-4] It is common among neonates and infants, and it is incessant.[5-10] Tachycardia-induced cardiomyopathy (TIC) is therefore common. The placement of the accessory pathway is frequently in the posteroseptal location, near the coronary sinus ostium.[2] In sinus rhythm, there is no preexcitation. Electrocardiography (ECG) demonstrates tachycardia with long RP with narrow QRS and reverse-sharp (sharp) P waves in the inferior leads during tachycardia. The occurrence of spontaneous resolution is extremely unusual, and it is frequently resistant to medical treatment. Since PJRT is a rare SVT, few studies have presented data and approaches.[2,5-9] The purpose of this study was to assess

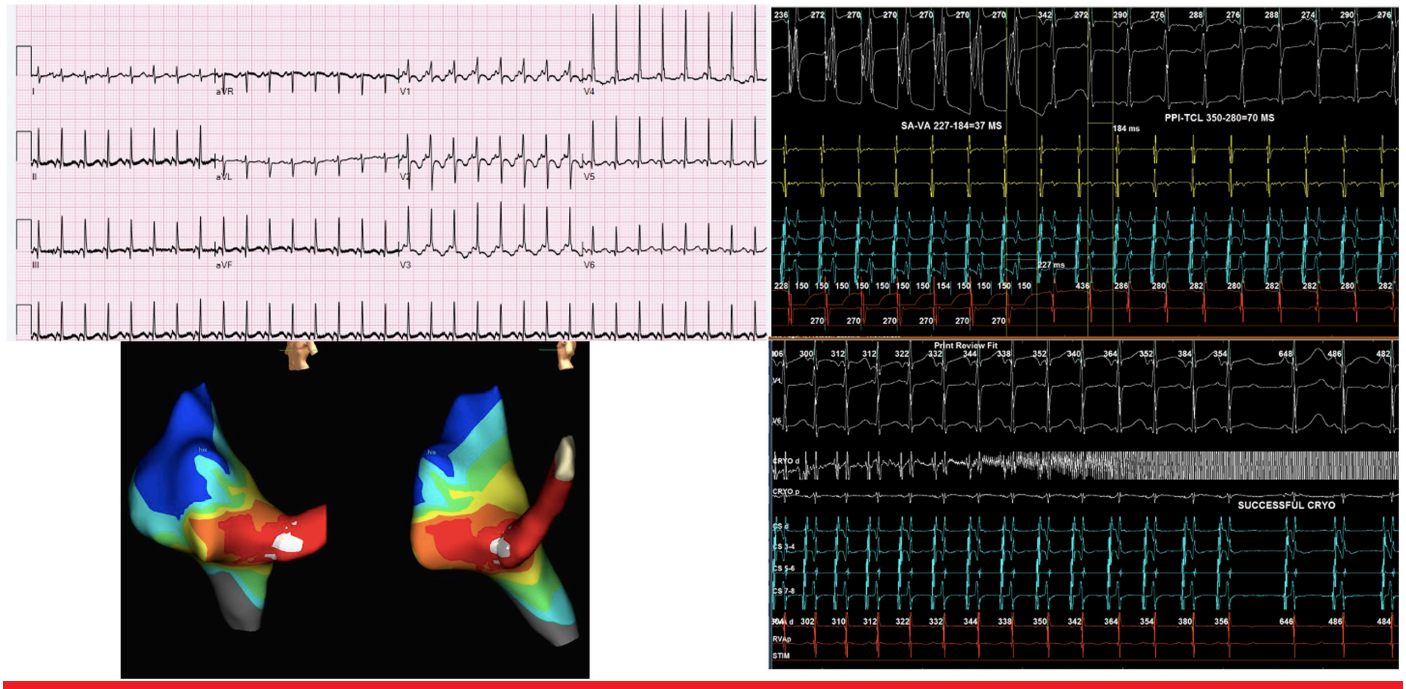

Figure 1. Top right: superficial electrocardiography example showing long RP tachycardia; top left: intracardiac electrocardiogram with PPI-TCL115 ms and SA-VA < 85 ms after ventricular entrainment; bottom right: 3D map showing accessory pathway localization; and bottom left: after successful cryoablation, intracardiac recordings show sinus rhythm. PPI, post-pacing interval; TCL, total cycle length; SA, stimulus–A interval; VA, ventricle–atrium interval.

the clinical, ablation, and follow-up data of our patients diagnosed with PJRT at our tertiary pediatric arrhythmia centers.

METHODS

Between 2011 and 2021, data from patients diagnosed with PJRT in 2 pediatric electrophysiology centers were analyzed retrospectively from file records. Approval for the study was obtained from the Local Ethics Committee (2022.01.04). The research involved a total of 29 patients. Patients' age, height, weight, symptoms, electrocardiography, rhythm Holter and echocardiography, medical treatment and responses, and electrophysiological study data were acquired from the file records of patients who had electrophysiological study (EPS) ablation.

After general anesthesia, intubation/laryngeal mask, decapolar diagnostic catheter insertion into the coronary sinus, and quadripolar high right atrium and right ventricle catheters, an electrophysiological study, mapping, and ablation were performed using the EnSite 3D mapping system (St. Jude Medical, Inc., St. Paul, Minn, USA). The following electrophysiological findings were used to establish the diagnosis of PJRT: (Figure 1)

- 1 : 1 AV relationship, long RP SVT;
- Post-pacing interval (PPI)–total cycle length <115 ms [corrected (c) PPI<110 ms in some cases];
- Entrainable from the ventricle and stimulus–A interval and ventricle–atrium interval< 85 ms;
- His refractory premature ventricular contraction (PVC) atrium resets (advance or delay) or tachycardia stops with VA block;
- His-A interval entrainment from ventricle–SVT His-A interval was defined as <0 ms detection.[1]

The ablation age, weight, indication, methods and energy type, accessory pathway localization, procedure time, fluoro duration, dose, and success rate were obtained from the procedure records of patients who underwent ablation. All patients waited for half an hour after ablation. After the waiting period, isoproterenol was used in addition to the basal planned stimulations for EPS-based tachycardia stimulations. File records were used to examine the recurrence rate, complications, cardiomyopathy (CMP) recovery time, and post-procedure follow-up duration records.

Statistical Analysis

The data were analyzed with the Statistical Package for the Social Sciences version 25.0 (SPSS Inc., Chicago, Ill, USA), with percentages for categorical data and mean and standard deviations for numerical data.

HIGHLIGHTS

- Permanent junctional reciprocating tachycardia is an uncommon and frequently incessant supraventricular tachycardia.
- Despite the use of multiple medical therapies, the remission rate is poor.
- Ablation may be done at any age and has a low complication rate.
- The recurrence rate following ablation is high, and further intervention may be needed.

RESULTS

The study included a total of 29 cases from 2 different centers. The mean age at diagnosis of the patients was 3.13 ± 4.43 (0-18) years and their body weight was 18.22 ± 19.68 (2.85-94) kg. Eighteen patients (62.1%) were female and 11 (37.9%) were male. Four of the patients had a history of tachycardia that began during their prenatal. It was diagnosed in the neonatal period in 7 patients and under 1-year-old in 16 patients. While 24% of the patients were asymptomatic at the time of presentation, the other patients had dyspnea (31%), palpitation (24%), and chest discomfort (6.9%). At the time of admission, 31% of the patients had TIC, with a mean shortening fraction of 17 ± 7% and an ejection percentage of 34 ± 14% in these patients. Tachycardia was incessant in 15 (51.7%) patients. All patients except 1 patient were given medical treatment. Twenty-two patients were using 2 or more drugs. The majority of the patients (41.4%) were receiving triple combination therapy. Amiodarone, beta-blockers, class 1 c drugs, and their combinations were the most widely utilized treatments. Medical therapy was frequently just partially effective (62.1%) or completely ineffective (17.2%). With medical treatment, tachyarrhythmia was controlled in 6 cases. Except for hypothyroidism in one patient and hepatotoxicity in another, there were no adverse effects.

Twenty-two of the 29 patients (22/29, 75.8%) had EPS/ablation, and 22 of them had a total of 26 ablation procedures. Five patients were under 1 year of age when EPS was performed. Table 1 shows the EPS/ablation statistics. Before being admitted to our clinics, 3 patients had failed catheter ablation at other institutions. In 2 patients, the operation was repeated twice, and in 1 patient, it was repeated 3 times. Indications for ablation of the patients were TIC resistant to medical therapy in 7 patients, incessant tachycardia unresponsive to medical therapy in 9 patients, elective in 6 patients. After infancy follow-up under elective conditions, ablation was done in 6 patients owing to tachycardia episodes that continued to require medical care until they were >5 years and 20 kg. The acute success rate was 22 out of 22 (100%). Only 1 procedure was performed from the right jugular vein. All other attempt procedures were conducted via right and left femoral veins. Accessory pathway localization was right posteroseptal in 18 (81.81%) patients, the intracoronary sinus-middle cardiac vein in 2 patients, midseptal in 1 patient, and posterior-posteroseptal oblique in 1 patient. In 16 of the procedures, cryoablation (6 mm in 13 patients, 8 mm in 3 patients), Radiofrequency (RF) ablation (7 patients), and irrigated RF (4 patients) were applied. Cryoablation and RF ablation were employed as the same procedure in 1 patient. The average number of lesions was 5.65 ± 2.1 during cryoablation at −80°. In patients who had RF ablation, the mean power was 27.4 ± 11.09 W, the impedance was 104.9 ± 13.9 Ω , and the lesion duration was 232.5 ± 159.41 seconds. Fluoro was employed in 24 operations, with

Table 1. Demographic and Ablation Data of Patients Who Underwent Ablation

	Mean ± SD (Min-Max)
Diagnosis age (years)	4.07 ± 4.73 (0-18)
Weight (kg)	21.47 ± 21.2 (2.85-94)
Ablation age (years)	5.47 ± 4.34 (0.07-18.19)
EF (%)	54.43 ± 18.62 (22-85)
FS (%)	27.57 ± 9.89 (11-46)
Rate (ms)	339.47 ± 48.25 (260-440)
RP interval (ms)	231.83 ± 53.76 (155-350)
PR interval (ms)	109.00 ± 39.13 (50-170)
PPI-TCL (ms)	90.77 ± 18.97 (67-124)
SA-VA (ms)	62.40 ± 14.98 (34-85)
Fluoro time (minutes)	2.97 ± 5.91 (0-26.1)
Fluoro dose (mgray/m^2)	248 ± 894 (0-3775)
Procedure time (minutes)	148.67 ± 41.36 (90-300)
Follow-up (years)	4.43 ± 3.06 (0.25-10.1)

EF, ejection fraction; FS, shortening fraction; PPI, postpacing interval; TCL, total cycle length; SA-VA: stimulus to A- ventricle to A interval.

Table 2. Clinical and EPS Characteristics of Patients with Recurrence

	Case 1	Case 2	Case 3	Case 4
Age (years)/sex	11/M	6/F	4.25/ F	1.5/ F
Weight (kg)	50	22	13	10
Fetal tachycardia (+/−)	−	−	−	+
Incessant	+	+	+	−
Ejection fraction (%)	69	28	30	75
Medication	Flecainide Metoprolol	Amiodarone Flecainide Propranolol	Amiodarone Flecainide Propranolol	Amiodarone Propafenone Propranolol
Ablation history	3 times	2 times	Once	-
Rate	420	410	352	300
Ablation	First: Irrigated RF Second: CR (8 mm)	First: CR (6 mm) Second: RF Third: Irrigated RF	First: CR (6 mm) Second: Unsuccessful	First: RF Second: CR (6 mm)
Localization	First: RPS Second: MCV ostium	RPS	RPS	RPS

CR, cryoablation; RF, radiofrequency ablation; RPS, right posteroseptal; MCV, middle cardiac vein.

Table 3. Data of Studies Conducted on PJRT and Our Study

Study	n	Age	TIC Rate (%)	Medical Treatment Response	Spontaneous Resolution	Adverse Event	Ablation Age/Rate	Success Rate of Ablation (%)	Localization	Complication	Follow-Up Time	Recurrence
This study (2 center)	29	3.13 years	31	13.7% full 62.1% partial	3.44%	6.88%	5.4 years 22/29 26 procedure 15 CR 6 RF 4 irrigated RF 1 CR + RF	100	81.8% RPS 6.8% CS	No	4.43 years	First ablation: 18% Second ablation: 3.44%
Kang[5] (11 center)	194	3.2 month	18	23% full 64% partial	12%	4%	N: 140 (175) 7.9 years 138 RF 27 CR 10 RF + CR	90	66% RPS 8.7% R intermediate septal 7.6% LPS	9% RF (7%) CR (12%)	3.8 years	First ablation 13% Second ablation 19%
Valksmann[2] (7 center)	85	3 month	28	50%-80%	22% (5.4 years) 2 ex 1 TIC	14%	N: 18 (24) 17.5 years	77	?	?	8.2 years	? 3 patients 2 times 1 patient 3 times
Barbero [8]	<2 month 16	<2 month	?	43%	6% (3 years)		N: 9 (neonatal) N: 9 (8 month)	93	81.25% RPS 12.5% LPS 6.25% LL	5.5%	10.4 years	First ablation: 11% Second ablation: %0
Meiltz[6] (6 center)	Adult 49	43 ± 16 years	30	?	?	?	N: 49 43years	94	75% RPS	2%	49 ± 38 years	8%
Gökoğlan [7]	Adult 62	30.5 ± 11 years	9.7				N: 62	90.3 98.4	59.7% RPS 11.3% LP 8.1% LL 4.8% MS		23 ± 22 month	8%
Dorostkar[9] (2 center)	21	0.54 years	28.5	52%	0	4.7%	N: 20 (21) 12.8 years 20 RF 1 CR	96.3	81% RPS 19% freewall	4.7%		23.8%

RPS, right posteroseptal; LPS, left postreoseptal; LL, left lateral; MS, midseptal; RF, radiofrequency ablation; CR, cryoablation; TIC, tachycardia-induced cardiomyopathy.

a mean fluoro duration of 3.07 (0.3-26.1) minutes and dosage of 212.67 (0.19-3775) mgray/m^2. The total procedure time (sheath to sheath) was 148.67 (90-300) minutes.

None of the patients had any complications. The rate of recurrence was 18% (there were a total of 5 recurrences in 4 patients). Table 2 summarizes the characteristics of patients with recurrence. Three of the relapsed patients already had recurrent ablation at other centers before presenting to our clinic. Patients who relapsed often had an early recurrence within 24 hours and 2 of them had TIC. Re-intervention was performed in 3 of 4 patients, and 2 patients did not recur after the second procedure, while 1 patient required the third ablation procedure. Cryoablation was used in the first procedure of this patient, classical RF was used in the second procedure, and finally, irrigated RF was used and this ablation was performed through the mouth of the coronary sinus and no recurrence was observed. Possible epicardial localization in the coronary sinus was considered as the cause of recurrence in this patient. Because in the first 2 procedures, tachycardia ended in the first lesion with the standard waiting time of 30 minutes after the procedure. Afterward, VA conduction was controlled and stimulation with basal and isoproterenol was performed. Cryoablation was performed in 2 of the patients with relapse (2 of them 6 mm), and 1 of them was made RF.

Patients were followed for a mean of 4.43±3.06 years, including after medical or ablation procedures. Cardiomyopathy improved in all patients who had ablation during the post-procedure follow-up (7 days-1 year). The mean shortening fraction (SF) was 33.19 ± 6.44% and the EF was 66.57 ± 12.18% at the follow-up. Without EPS/ablation, only 1 out of 7 patients followed up with medical treatment recovers spontaneously, while 6 patients continue medical treatment.

DISCUSSION

Our study is part of a wide series of pediatric PJRT cases published in the literature. This is the country's first pediatric research study about PJRT clinical course and EPS/ablation results. There are some differences in the studies published in the literature, despite some similarities. Table 3 summarizes the studies performed on adults and children, as well as the main findings of our study.

Although PJRT can be found at any age, beginning in the fetal period, the majority of research and case reports in the children's age range are under the age of 1 year.[8-11] However, there have been studies conducted on adults. The mean age in our study was found to be greater than in other pediatric age group studies. However, a specific age group was selected in some of the reported studies. Because all pediatric age groups were covered and our study was done in electrophysiology centers; one of the reasons for the high mean age of our patients might be that the patients who were followed up medically were not or delayed referred.

Tachycardia-induced cardiomyopathy has been observed to occur in 9.7%-30% of PJRT patients in different studies. The rate of 31% in our study was comparable to the high rates reported in the literature. Tachycardia-induced cardiomyopathy improved rapidly following ablation, as reported in the literature. It was considered that the high rate was due to the fact that our centers are the reference centers, and patients who can not be managed by medical therapy were referred there. Although Vaksmann et al[2] reported a rate of spontaneous resolution as high as 22%, another investigation revealed no resolution. Vaksmann et al[2] on the other hand, had a 5.4-year follow-up period, in which 2 patients died and 1 patient had TIC. In our investigation, only 1 patient achieved spontaneous resolution, no mortality was observed in any patient, and CMP improved in all patients. Although rates of response to medical treatment have been reported as high as 50%-80%, the complete response rate of 23% and partial response rate of 64% in the largest pediatric series published by Kang et al[5] are similar to our study. In pediatric trials, the rate of adverse effects from medicinal therapy was 4%-14%, and it was comparable in our patients (6.8%).

While the ablation success rate in the pediatric age group was reported to be 77%-96.4 %, the acute procedure success rate in our patients was 100%.[2,5,8,9] There have been reports of complication rates ranging from 2% to 12%. Despite the fact that the ablation age in our study was lower than that reported in other studies, no complications were observed in any of our patients, and the acute success rate was also high. Ablation localization was most common in the right posteroseptal region, as reported in all studies. Our study had a high recurrence rate, which was similar to the Dorostkar et al[9] and Kang et al[5] series in the literature; however, our patients' ablation age and complication rate were lower than in these studies. Most of the patients who relapsed in our study had a history of intervention before they applied to our center. Croyoablation was employed in 2 of the 5 relapse surgeries, and Rf ablation was used in 2 of them, 1 of which used irrigated RF. The procedures were not compared since the number of relapsed patients was so small. Although Kang et al[5] reported that cryoablation had a higher recurrence rate, there is no study in the literature that compares the procedures. Repeated ablation of our patients with recurrences has been successful using different modalities.

Study limitations

The limitations of our study are its retrospective design, inadequate follow-up information of some patients, a small number of patients, and the fact that EPS was not conducted on all of them. Despite the fact that patient recruitment from different centers may result in disparities in patient treatment, the 2 centers' patient management in our study is largely identical.

CONCLUSION

In conclusion, despite the rarity of PJRT, cardiomyopathy has a significant prevalence because of its frequent incessant character. Although it is most commonly observed in infants, it can occur at any age beginning with prenatal life. Patients who had undergone cryoablation and had a history of recurrent procedures made up the majority of those who relapsed. The response to medical therapy is not sufficient, despite the use of multiple drugs. The resolution of spontaneous resolution is

really poor. Although ablation has a high (18%) recurrence rate, it has a low complication rate and may be performed safely, especially in patients with TIC who are resistant to medical treatment and develop side effects, even at a young age and low weight. Different modalities should be explored on a case-by-case basis in recurrent procedures. Prospective research comparing different ablation techniques are required.

Ethics Committee Approval: University of Health Sciences İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital Non-Interventional Researches Ethics Committee approval was obtained (2022.01.04).

Informed Consent: The data were obtained retrospectively from electronic file records.

Peer-review: Externally peer-reviewed.

Author Contributions: Study conception and design: YE, AS; data collection: AS, BÇ; analysis and interpretation of results:AS, HCK, CA, draft manuscript preparation:YE, AS, BÇ, HCK, CA, VT, Supervisor; YE, CA, VT. All authors reviewed the results and approved the final version of the manuscript.

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