Permanent junctional reciprocating tachycardia in children:

a multicenter study on clinical profile and outcome

Guy Vaksmann*, Christine D’Hoinne*, Vincent Lucet†, Sophie Guillaumont‡, Jean-Marc Lupoglazoff§, Alain Chantepie║, Isabelle Denjoy¶, Elisabeth Villain#, François Marçon**.

Brief Title: PJRT in Children
* department of pediatric cardiology, cardiologic hospital, Lille
† Department of pediatric dysrhythmias, Le chateau des côtes, Les Loges en Josas
‡ Department of pediatric cardiology, Montpellier
§ Department of pediatric cardiology, Robert Debré Hospital, Paris
║ Department of pediatric cardiology, Trousseau Hospital, Tour
¶ Department of electrophysiology, Lariboisiere Hospital, Paris
# Department of pediatric cardiology, Necker Hospital, Paris
** Department of pediatric cardiology, Pediatric Hospital, Nancy

Address for Correspondance: Dr Guy Vaksmann, Polyclinique de la Louviere, 69 rue de la Louviere, 59800, Lille, France.
Phone number : 00 33 680 92 02 72.
E-mail : guy.vaksmann@wanadoo.fr

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Abstract

Objectives. The aim of this study was to investigate the clinical profile, natural history and optimal management of permanent reciprocating tachycardia (PJRT) in children.

Background. Persistent or permanent junctional reciprocating tachycardia (PJRT) is an uncommon arrhythmia characterized by an incessant orthodromic tachycardia with anterograde conduction over the atrioventricular node, and retrograde ventriculoatrial conduction via an accessory pathway with slow and decremental conduction. The arrhythmia is commonly incessant from birth or infancy and may lead to a tachycardia-induced cardiomyopathy that is reversible with rate control. Due to the rareness of the reports on natural history of PJRT, little is known about the clinical course and the optimal management of this chronic tachycardia.

Methods and Results. Eighty-five patients meeting the ECG criteria for PJRT were included in a retrospective multicenter study. Age at diagnosis varied from birth to 20 years (median age: 3 months). Follow-up ranged from 0.1 to 26.0 (median 8.2) years. At the time of referral, 24 of 85 patients (28%) had congestive heart failure (CHF) that was resolved with medical treatment in all patients. Eighty three patients received drug treatment initially. Amiodarone and Verapamil were the most effective with a success rate of 84 to 94% alone or in association with digoxin. Radiofrequency ablation of the accessory pathway was performed in 18 patients. There was a trend for a relation between age at ablation and the result of the procedure, failures being more frequent in younger patients (p = .14). Two patients with persistent left ventricular dysfunction on echocardiography, but no symptoms of CHF, died suddenly 1 month and 3 years after diagnosis. Spontaneous resolution of PJRT occurred in 19 patients (22 %), age at diagnosis of PJRT was not predictor of spontaneous resolution.

Conclusions. PJRT is a potentially lethal arrhythmia in children with tachycardia-induced cardiomyopathy. Spontaneous resolution of tachycardia is not uncommon. Antiarrhythmic treatment is often effective. Radiofrequency ablation should be performed in older children or when rate control is not achieved, especially in patients with persistent left ventricular dysfunction.

Abbreviations:
PJRT: Persistent or permanent junctional reciprocating tachycardia
ECG: electrocardiogram
Persistent or permanent junctional reciprocating tachycardia (PJRT) is an uncommon arrhythmia, first described by Counel et al [1], characterized by an incessant orthodromic tachycardia with anterograde conduction over the atrioventricular node, and retrograde conduction via an accessory pathway usually located in the postero-septal region with slow and decremental conduction [2] [3] [4]. The arrhythmia is commonly incessant from birth or infancy, even though it may not be recognized until adulthood [5]. Its persistence over a long period of time may lead to a tachycardia-induced cardiomyopathy that is reversible with rate control [6] [7] [8] [9] [10]. PJRT is thought not to demonstrate the spontaneous resolution characteristics of other atrioventricular re-entry tachycardias in infancy but spontaneous resolution has been reported in some patients [8]. The arrhythmia is considered as usually refractory to drug therapy and various nonpharmacologic approaches have been utilized to prevent tachycardia or for rate control. Over the last few years radiofrequency catheter ablation of the accessory pathway has been reported to be very effective and safe as definitive treatment of PJRT [11] [12] [13] [14]. Although most patients having had catheter ablation had reach adulthood it has been proposed that the technique should be considered early in the management of these patients [14] [15]. However, due to the rareness of the reports on natural history of PJRT, little is known about the clinical course of this chronic tachycardia. Single institutional experience on PJRT usually is small, and generally accepted guidelines on how to manage these patients are still lacking. On the basis of a multi-institutional experience reporting the largest population of patients with PJRT, we attempted to determine the clinical profile, natural history and optimal management of this uncommon dysrhythmia.

Methods
Patient selection
This multicenter retrospective study included 7 French pediatric institutions involved in the care of pediatric dysrhythmias. The participating centers and the number of contributed patients are as follows: Les loges en Josas (27 patients), Lille (18 patients), Montpellier (13 patients), Nancy (12 patients), Paris-Necker hospital (5 patients), Paris-Robert Debré hospital (6 patients), Tours (4 patients). Twenty-one of the 85 patients have been the subjects of previous reports [9] [10]. A questionnaire addressing the patients’ clinical course, therapy and follow-up was given to each center to complete. Patients were included if they presented with PJRT prior to their twentieth birthday. Follow-up of each individual patient had to be longer than 1 year unless the affected patient died earlier.

Diagnosis of PJRT was based on surface ECG criteria including the presence of an almost incessant narrow QRS tachycardia with negative P waves in inferior leads, a P'R interval shorter than the RP' interval and an atrioventricular ratio of 1:1. During episodes of sinus rhythm PR interval had to be normal with no evidence of delta wave. During tachycardia, no episode of functional atrioventricular block had to be documented on 24-hour ambulatory recordings or vagal maneuvers.
All patients were followed as outpatients. An intracardiac electrophysiologic study was performed in 21 patients with subsequent catheter ablation attempt in 18.

Criteria for treatment efficiency:
The results of treatment were assessed by repeated ECGs and 24-hour ambulatory recordings. Treatment success was defined as conversion to stable permanent sinus rhythm or to predominant sinus rhythm alternating with non sustained (< 30 seconds) low rate PJRT (< 110 bpm) with normal left ventricular function on echocardiography; partial success was defined as significant decrease in heart rate (PJRT rate reduction > 30 %) associated with periods of sustained sinus rhythm on Holter monitoring and normalization of left ventricular function on echocardiography if dysfunction was initially present; failure was defined as non significant decrease of heart rate or persistence of left ventricular dysfunction if initially present; spontaneous resolution of PJRT was defined as no recurrence of PJRT after cessation of antiarrhythmic treatment with permanent sinus rhythm on Holter monitoring and exercise test in patients older than 8 years.

Statistics: Data were compared using Student’s T test for continuous variables. Relative frequencies observed between different groups of patients were compared using Fisher exact test.

Results:
Patients characteristics at presentation:
The 85 patients (43 females, 42 males) were admitted for incessant tachycardia in 1966-1998. The initial age at diagnosis varied from birth to 20 years (median age: 3 months); 57 of 85 patients
presented before 1 year of age, 19 of them had a history of intrauterine tachycardia (3 with hydrops fetalis) although the type of tachycardia could not be ascertained from the fetal echocardiogram. (figure 1). One patient with Williams syndrome had a mitral valve prolapse with grade 2 mitral insufficiency. No other patient had structural heart defect.

At the time of referral, 24 of 85 patients (28%) presented with varying degrees of congestive heart failure. Congestive heart failure was more frequent in infants (21/57 or 37%) than in children older than 1 year (3/28 or 11%, p<.05). Infants had a higher tachycardia rate than older children (221 ± 40 v 180 ± 44 bpm, p<.01). However, the ventricular rate in those patients with congestive heart failure was not significantly different than in those without congestive heart failure (217 ± 30 v 205 ± 50 bpm, p>.05).

**Follow-up and treatment**

Follow-up ranged from 0.1 to 26.0 (median 8.2) years. Of all patients, 2 never received drug therapy because of low rate tachycardia of 100 bpm, normal left ventricular function and absence of symptoms in the first patient and because of primary radiofrequency ablation procedure in the other patient with a well tolerated tachycardia of 150 bpm.

Eighty three patients received drug treatment initially. Among them, only 17 were managed with one single antiarrhythmic drug, whereas 66 received at least 2 consecutive antiarrhythmic agents, but none received more than one antiarrhythmic drug simultaneously, except in combination with digoxin. Of these, 24 even received consecutively four to nine antiarrhythmic drugs (figure 2).

A comparison of the efficacy (complete and partial success) of the antiarrhythmic agents used is shown in figure 3. The reported initial success rate were highest for amiodarone and verapamil which were effective or partially effective alone or in association with digoxin in 84% to 94% of patients. Verapamil was given if patients were older than 2 years. Digoxin alone had a success rate of 52%. Relapse of PJRT after initial success occurred in 6 patients; 4 were on digoxin, 1 received digoxin and amiodarone and 1 received verapamil. The 4 patients who relapsed on digoxin were successfully treated with amiodarone (3 patients) or verapamil (1 patient). The 2 other patients underwent successful radiofrequency ablation of their accessory pathway.

Treatment side effect occurred in 12 patients. Two had chemical evidence of hyperthyroidism and 4 had chemical evidence of hypothyroidism. None had clinical manifestation of thyroid dysfunction and lab values normalized after cessation of the drug. Photosensitivity occurred in 3 other patients who received amiodarone. Corneal microdeposits were found in only one patient receiving amiodarone but were not systematically searched. Two patients receiving flecainide or propafenone had post tachycardic pauses on Holter monitoring of more than 3 seconds suggesting sinus node dysfunction. Radiofrequency ablation of the accessory pathway was performed in 18 patients by experienced operators from 4 institutions in 1988 – 1998 for a total of 24 procedures. Median age at ablation was 17.5 years (range 4 – 25). Ablation was successful at first attempt in 14 patients including the patient with mitral valve prolapse, at second attempt in 3 patients and at the fourth attempts in 8 years in one patient. There was a trend for a relation between age at ablation and the result of the procedure, failures being more frequent in younger patients. Three on 6 procedures performed in children younger than 10 years were successful compared to 15 on 18 procedures performed in older children (p = .14).

When initially present, congestive heart failure resolved with medical treatment in all patients. However, 2 patients with persistent decreased left ventricular ejection fraction on echocardiography, but no symptoms of congestive heart failure, died suddenly. The first one was a 4 year-old boy who suffered from pulmonary edema at admission. He was treated with amiodarone and died 1 month after onset of treatment despite rate control and improvement of clinical symptoms and of left ventricular ejection fraction. The second one was a 9 year-old girl who received digoxin for 3 years, thought in good control under treatment but with persistent left ventricular dilatation on echocardiography with an ejection fraction of the left ventricle of 45 %.

Spontaneous resolution of PJRT occurred in 19 patients (22 p cent) 5.4 years (2 months to 16 years) after diagnosis of tachycardia. Median follow-up after spontaneous resolution was 2.2 years (range .3 – 11.9). Age at diagnosis of PJRT was not predictor of spontaneous resolution. Spontaneous resolution occurred in 14/57 (25 p cent) of patients younger than 1 year and in 5/28 (18p cent) of patients older than 1 year at diagnosis (p=NS).

**Status at last follow-up.**
At last follow-up, 36 patients (42%) were free of PJRT on Holter monitoring and/or exercise test and free of pharmacologic treatment including the 19 patients who demonstrate spontaneous resolution and the 17 with successful radiofrequency ablation. Twenty-six patients (31%) were also free of PJRT on Holter monitoring but were still treated with antiarhythmic drugs. Five patients (6%) had a low rate PJRT or had an initially incessant tachycardia that became non sustained and were not treated, among them is the patient who was never treated. Only 18 patients (21%) had PJRT on drugs, among them were the 2 patients who died suddenly. None of the remaining 16 patients had clinical symptoms or impairment of left ventricular function on echocardiography.

**Discussion:**

This multicenter study having collected a large number of patients with PJRT confirmed some findings and brought some new characteristics useful for the management of this rare dysrhythmia.

**Clinical profile:**

PJRT may be diagnosed at any age during childhood but a majority of patients are diagnosed during infancy or prenatally. Symptoms of congestive heart failure were more common in younger patients before introduction of medical therapy. It is likely that older patients at diagnosis of PJRT have had unrecognized slower tachycardia less sustained during early childhood explaining their better tolerance and their late diagnosis. Congestive heart failure always resolved with antiarrhythmic treatment. Nevertheless 2 patients with persistent left ventricular dysfunction on echocardiography, although clinically improved by treatment, died suddenly 1 month and 5 years after diagnosis. Unlike accessory pathways of wolff-Parkinson-White syndrome in children that are associated with structural heart defect in approximately one third of patients \[17\] \[18\] \[19\] \[20\] \[21\], accessory pathways of PJRT are generally isolated. Only one patient of this series with Williams syndrome had a structural heart defect. In addition, no patients from 2 other large series published having collected 53 patients had structural cardiac anomaly \[8\] \[22\].

One of the main finding of the present study is the relatively high rate of spontaneous resolution of PJRT which exceeded 20%. In contrast to what has been observed in smaller series of pediatric patients reported so far, spontaneous resolution of PJRT was not so uncommon \[8\] \[22\]. Such an evolution happened sometimes many years after diagnosis of tachycardia. Another interesting finding of this study is the remarkable efficiency of some antiarhythmic drugs on dysrhythmia, especially amiodarone and verapamil that had a success rate of more than 80%. Even patients receiving digoxin alone had a success rate of more than 50%. In our series, RF ablation had a greater success rate in older patients. In addition, although RF ablation can be performed at any age, even in infancy, complications in this age group are more frequent \[23\] \[24\] \[25\]. Furthermore, experimental data have shown that radiofrequency lesions appear to increase in size with somatic growth with replacement of normal myocytes with fibrous and elastic tissue representing a potential arrhythmogenic substrate \[26\]. Thus, considering high success rate of antiarhythmic drugs, it is our recommendation that patients with PJRT should be treated initially with amiodarone or verapamil in combination with digoxin. Usual efficiency of these drugs may allow delay of RF ablation until patients have reached an adequate growth.

**Limitations of the study:**

First, the present study is a retrospective and multicenter investigation. However it included a large number of patients with a relatively uniform management. In all the centers, medical treatment was tried first, especially in younger patients, and radiofrequency ablation had been a first choice procedure in only one patient. The other patients have been ablated after failure of medical treatment or occurrence of side effects or in patients having reached adulthood.

Second, electrophysiological study was not mandatory for patient inclusion. However, ECG and Holter monitoring studies allowed to exclude the diagnosis of ectopic atrial tachycardia. Intrnodal reentry, on its rarer form (fast-slow) could be another possibility but such a tachycardia is very uncommon in children and we do not believe that conclusions of our study could be modified by the absence of electrophysiological study for patient inclusion.

Third, exercise test was not mandatory to consider that antiarrhythmic treatment was successful. However, the goal of treatment was not to obtain a permanent sinus rhythm but to avoid sustained high frequency (> 110 bpm) tachycardia on Holter monitoring. However, exercise test was performed in...
children older than 8 years who did not receive antiarhythmic treatment to confirm either spontaneous resolution of the tachycardia or success of radiofrequency ablation.

**Conclusion:**

PJRT is a potentially lethal arrhythmia in children with tachycardia-induced cardiomyopathy. Although rarely reported, spontaneous resolution is not uncommon. Antiarhythmic treatment is often effective especially amiodarone and verapamil. Radiofrequency ablation should be reserved for older patients but may be indicated even in the very young when rate-control is not achieved and especially in patients with persistent left ventricular dysfunction.
References


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