Permanent junctional reciprocating tachycardia in children: A multicenter experience

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BACKGROUND Permanent junctional reciprocating tachycardia (PJRT) is an uncommon form of supraventricular tachycardia in children. Treatment of this arrhythmia has been considered difficult because of a high medication failure rate and risk of cardiomyopathy. Outcomes in the current era of interventional treatment with catheter ablation have not been published.

OBJECTIVE To describe the presentation and clinical course of PJRT in children.

METHODS This is a retrospective review of 194 pediatric patients with PJRT managed at 11 institutions between January 2000 and December 2010.

RESULTS The median age at diagnosis was 3.2 months, including 110 infants (57%; aged <1 year). PJRT was incessant in 47%. The ratio of RP interval to cycle length was higher with incessant than with nonincessant tachycardia. Tachycardia-induced cardiomyopathy was observed in 18%. Antiarrhythmic medications were used for initial management in 76%, while catheter ablation was used initially in only 10%. Medications achieved complete resolution in 23% with clinical benefit in an additional 47%. Overall, 140 patients underwent 175 catheter ablation procedures with a success rate of 90%. There were complications in 9% with no major complications reported. Patients were followed for a median of 45.1 months. Regardless of treatment modality, normal sinus rhythm was present in 90% at last follow-up. Spontaneous resolution occurred in 12% of the patients.

CONCLUSION PJRT in children is frequently incessant at the time of diagnosis and may be associated with tachycardia-induced cardiomyopathy. Antiarrhythmic medications result in complete control in few patients. Catheter ablation is effective, and serious complications are rare.

KEYWORDS Permanent junctional reciprocating tachycardia; Antiarrhythmic medications ok; Catheter ablation; Pediatrics

ABBREVIATIONS **AV** = atrioventricular; **ECG** = electrocardiogram/electrocardiography; **EF** = ejection fraction; **IQR** = interquartile range; **PJRT** = permanent junctional reciprocating tachycardia; **RFA** = radiofrequency ablation; **SF** = shortening fraction; **TIC** = tachycardia-induced cardiomyopathy

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Introduction

Permanent junctional reciprocating tachycardia (PJRT) is a rare form of supraventricular tachycardia that occurs predominantly in infants and children.[1] The arrhythmia substrate is an accessory pathway with slow, decremental retrograde conduction[2] that is commonly located in the

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posteroseptal region of the atrioventricular (AV) junction.[3] In the majority of affected patients, PJRT is incessant and may lead to tachycardia-induced cardiomyopathy (TIC), which is reversible with a sustained period of rate or rhythm control.

There are several studies on the course and treatment of PJRT in children,[4–9] but they do not reflect management and outcomes in the current era. In previous case series, PJRT was controlled with antiarrhythmic medications in up to 25% of the pediatric patients, and spontaneous resolution was uncommon.[5,8,9] Disappointing outcomes from medical management have resulted in the use of catheter ablation as primary therapy and for PJRT that is refractory to antiarrhythmic medications.[4–8] Pediatric catheter ablation has evolved rapidly over the past decade, and modern technologies such as cryoablation and electroanatomic mapping have improved outcomes and decreased the risk of adverse events, the latter primarily reducing the need for fluoroscopy use during the procedure.[10]

The objective of this study was to describe the clinical course and outcomes of treatment of PJRT in children in the modern era to reflect these therapeutic advances in catheter ablation technology.

Methods

This is a retrospective cohort study of patients with PJRT from 11 pediatric centers. Participating centers were solicited through the Pediatric and Congenital Electrophysiology Society. Approval from the local ethics committee was obtained at each center.

Inclusion criteria

Patients were included if their diagnosis occurred from age 0 to 18 years with at least 1 follow-up visit between January 2000 and December 2011. The PJRT diagnosis was based on electrocardiographic (ECG) criteria described previously[2]: (1) narrow QRS tachycardia, (2) negative P waves in inferior leads, (3) PR interval to RP interval ratio <1, (4) 1:1 AV relation during tachycardia with no evidence of functional AV block, and (5) absent delta waves in the PR segment during sinus rhythm. Other evidence to support the diagnosis included termination with vagal maneuvers, adenosine, or cardioversion; spontaneous termination with an atrial event; and absence of a warm-up or cooldown.[11] For patients who underwent electrophysiologic testing, confirmation of the diagnosis of PJRT and identification of an accessory pathway were required.[12]

Data collection

Data were collected from existing medical records using a standardized data collection form. Study data were collected and managed using REDCap electronic data capture tools hosted by the Child and Family Research Institute at British Columbia Children's Hospital.[13]

Definitions

PJRT was considered incessant if present for $>90\%$ of the time monitored by 24-hour Holter ECG or telemetry, sustained if present 50%–90%, episodic if present 10%–50%, and sporadic if present $<10\%$ of the time. Cardiomyopathy was defined as a left ventricular ejection fraction (EF) of $<40\%$ or a shortening fraction (SF) of $<28\%$. Rhythm control was defined as normal sinus rhythm on 24-hour Holter ECG; rate control was defined as an age-appropriate normal heart rate and variability on 24-hour Holter ECG. Acute success with catheter ablation was defined as normal sinus rhythm and noninducibility of PJRT at the completion of the procedure. Recurrence was defined as documented PJRT after successful catheter ablation. Resolution was defined as normal sinus rhythm on 24-hour Holter ECG with no recurrence of symptoms after discontinuing antiarrhythmic therapy for at least 1 month. Spontaneous resolution was defined as resolution not requiring ongoing antiarrhythmic medications or ablation therapy. Clinical benefit was defined as any improvement in PJRT arrhythmia burden, including rate control, less incessant PJRT, or complete suppression. Determination of these was left to the discretion of the attending pediatric electrophysiologist.

Statistical analysis

Frequency tables were generated for all categorical data. Differences between groups were analyzed using either the χ^2 test or the Fisher exact test. Continuous data were analyzed using a univariate procedure. Data are presented as median (interquartile range [IQR]). Differences between the infant (<1 year at the time of diagnosis) and the older (≥ 1 year) patient cohorts and those diagnosed with PJRT before and after 2006 were analyzed using a Wilcoxon rank-sum test. All tests were 2-sided, and a P value of $<.05$ was considered statistically significant. All statistical analyses were completed using SAS statistical software (version 9.3, SAS Institute, Cary, NC).

Results

Clinical presentation and PJRT at diagnosis

There were 194 patients (98 male patients) included in the study. The median age at diagnosis was 3.2 months (IQR 0.1–116.1 months). PJRT cases were observed in a unimodal distribution peaking in infancy (Figure 1). There were 110 patients (57%) diagnosed with PJRT in infancy (aged <1 year), including 53 patients (27%) presenting with fetal tachycardia. PJRT presented with symptoms in 134 of 194 patients (69%), while in the remaining 31% of the patients it was an incidental finding on physical examination or ECG. The most common presenting symptoms were palpitations in 44 of 194 patients (23%) and symptoms of heart failure in 36 of 194 patients (19%). Fetal tachycardia was present with and without hydrops in 7% and 21% of the patients, respectively. Most patients (67%) were hospitalized at the time of diagnosis.