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Pediatric Nonpost-Operative Junctional Ectopic Tachycardia

Medical Management and Interventional Therapies

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Objectives To determine the outcomes of medical management, pacing, and catheter ablation for the treatment of nonpost-operative junctional ectopic tachycardia (JET) in a pediatric population.

Background Nonpost-operative JET is a rare tachyarrhythmia that is associated with a high rate of morbidity and mortality. Most reports of clinical outcomes were published before the routine use of amiodarone or ablation therapies.

Methods This is an international, multicenter retrospective outcome study of pediatric patients treated for nonpost-operative JET.

Results A total of 94 patients with JET and 5 patients with accelerated junctional rhythm (age 0.8 year, range fetus to 16 years) from 22 institutions were identified. JET patients presenting at age ≤6 months were more likely to have incessant JET and to have faster JET rates. Antiarrhythmic medications were utilized in a majority of JET patients (89%), and of those, amiodarone was the most commonly reported effective agent (60%). Radiofrequency ablation was conducted in 17 patients and cryoablation in 27, with comparable success rates (82% radiofrequency vs. 85% cryoablation, $p = 1.0$). Atrioventricular junction ablation was required in 3% and pacemaker implantation in 14%. There were 4 (4%) deaths, all in patients presenting at age ≤6 months.

Conclusions Patients with nonpost-operative JET have a wide range of clinical presentations, with younger patients demonstrating higher morbidity and mortality. With current medical, ablative, and device therapies, the majority of patients have a good clinical outcome. (J Am Coll Cardiol 2009;53:690–7) © 2009 by the American College of Cardiology Foundation

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Junctional ectopic tachycardia (JET) not associated with cardiac surgery is a rare arrhythmia that can present at any age (1). The mechanism of the tachycardia is thought to be abnormal automaticity arising from the region of the atrioventricular junction. Congenital JET presents in infancy and is associated with high morbidity and mortality (1–7). JET presenting beyond the first 6 months of life has a variable presentation. Most data concerning the treatment of JET are from single-center case reports and small case series (1–6,8–21). The most recent multicenter study on JET was published in 1990 (5), just as amiodarone was beginning to gain widespread use and before the era of catheter ablation as curative therapy. In addition, with improved technology for evaluation of the fetus, JET can now be diagnosed in utero, with the goal of earlier therapy and potentially improved outcome (15,22).

The purpose of this study is to describe outcomes of pediatric patients treated for JET, specifically addressing efficacy and safety of medical management, pacing, and catheter ablation.

Methods

Patient population. This is an international, multicenter retrospective review of nonpost-operative junctional tachycardias from the Pediatric and Congenital Electrophysiology Society. Approval for medical chart review was obtained from the Institutional Review Board at each institution. All patients ≤18 years of age evaluated and treated with the diagnosis of nonpost-operative JET or accelerated junctional rhythm (AJR) were included. Patients had either a normal structural heart or only minor cardiac defects that did not require cardiac surgery.

The diagnosis of JET was based on clinical, noninvasive evaluation of the patient by the attending electrophysiologist, using established electrocardiographic criteria (1,5) in addition to JET rate criteria (23,24). JET was defined as a tachycardia originating from the atrioventricular junction with gradual onset, gradual offset, and rate variability. The following features were observed: 1) a narrow QRS complex with either atrioventricular dissociation and sinus capture beats or 1:1 V:A association; or 2) a wide QRS complex with atrioventricular dissociation and sinus capture beats with the same wide QRS complex noted during tachycardia, the sinus capture beats, or sinus rhythm. JET was further defined by a junctional rate >95th percentile of heart rate for age (23,24). Patients with junctional rates ≤95th percentile were reported as having AJR. JET frequency on presentation was classified as incessant (a consistent tachycardia without conversion to sinus rhythm), sustained (JET occurring >50% of the time), or sporadic (predominantly sinus rhythm with episodic JET).

Abbreviations and Acronyms

AJR = accelerated junctional rhythm

JET = junctional ectopic tachycardia

RF = radiofrequency

Data collection. Medical charts were reviewed for the following: age at presentation, symptoms, family history of JET, antiarrhythmic therapies, details of radiofrequency (RF), or cryoablation procedures, the placement of a permanent pacemaker or defibrillator, and clinical outcomes. The finding of spontaneous atrioventricular block was recorded (10).

Statistical analysis. Data are presented as median (range). Chi-square and Fisher exact tests were utilized for categorical variables. The Wilcoxon rank sum test was used for nonparametric comparisons of continuous variables. Linear regression was utilized for the evaluation of JET rate as a function of patient age.

Results

Patient population. The study population consisted of 94 patients with JET and 5 patients with AJR from 22 institutions (Table 1). Several patients have been described previously (8,9,14,17,21). The median year of presentation was 2001 (1969 to 2008), with only 6 patients presenting before the availability of RF ablation. A familial association was noted in 20% of patients. The presence of maternal lupus antibodies was not routinely evaluated; however, 1 mother was known to be anti-Rho and anti-La positive. Minor heart defects were diagnosed in 24%, including 16 patent foramen ovale, 1 muscular ventricular septal defect, 4 patent ductus arteriosis, and 3 mitral valve prolapse.

Patients with accelerated AJR. The presenting symptom of patients with AJR was a rapid or irregular heart rate in 4 and unknown in 1. Rates of AJR were median 120 beats/min (117 to 170 beats/min). All 5 patients were given a trial of antiarrhythmic medications including digoxin (n = 3), atenolol (n = 2), verapamil (n = 1), and propafenone (n = 1). At last follow-up, all patients are doing well on no antiarrhythmic medications. One patient had rapid shift of

Table 1 Demographics of Pediatric Patients With JET and AJR

	All Patients	JET	AJR
Number	99	94	5
Male	58 (59%)	56 (60%)	2 (40%)
Median age, yrs (range)	0.8 (fetus–16)	0.9 (fetus–16)	0.8 (birth–7)
Age at presentation			
≤6 months	47 (47%)	44 (47%)	3 (60%)
>6 months	52 (53%)	50 (53%)	2 (40%)

AJR = accelerated junctional rhythm; JET = junctional ectopic tachycardia.

cardiac rhythm from AJR to complete atrioventricular block and required the placement of a permanent pacemaker. One patient had 2 cryoablation procedures, with no further tachycardia. Two patients had spontaneous resolution of their AJR, 1 after 2 electrophysiology studies without inducible tachycardia and with no ablation being performed. One patient continues to have episodic AJR but is asymptomatic.

Patients with JET. Patient age on presentation of JET was almost evenly divided between ≤6 months of life and >6 months of life (Table 1). Symptoms on presentation varied (Fig. 1). The majority of patients (60%; 56 of 94) had a rapid or irregular heart rate sensed by the patient or evaluated on physical examination. Congestive heart failure was found in 16% (15 of 94). Fetal tachycardia was diagnosed in 17% (16 of 94), with 4 of these patients having signs of hydrops.

JET frequency on presentation was more likely to be incessant in patients presenting at ≤6 months of age and sporadic for those presenting at >6 months (Fig. 2). The median JET rate on presentation was 209 beats/min (136 to 320 beats/min) and tended to be higher in younger patients (Fig. 3). Only 2 patients had a wide complex tachycardia (QRS 150 ms), which was diagnosed as JET during an invasive electrophysiology study. The relationship between ventricular and atrial contractions during tachycardia was ventricular-atrial dissociated in 56% (53 of 94), 1:1 ventricular/atrial relationship in 32% (30 of 94), periods of both in 3% (3 of 94), and not documented in 9% (8 of 94).

Antiarrhythmic medications in patients with JET. Antiarrhythmic medications were the initial therapy in 89% (84 of 94). Six patients (6%) with JET were monitored clinically and never required therapy. In these patients, JET rates were 140 to 211 beats/ min, and JET was episodic in 5 of the 6 patients. Another 4 (4%) patients underwent an ablation procedure as first-line therapy.

A wide variety of antiarrhythmic medications was utilized (Table 2). The majority of patients (62%; 52 of 84) required concomitant administration of ≥2 antiarrhythmic medications. Medications resulted in complete suppression of JET in only 11% (9 of 84). More commonly, medications were associated with a decrease in JET rate or frequency in 70% (59 of 84). In the remaining 19% (16 of 84), medications had no effect. Of the antiarrhythmic agents reported as

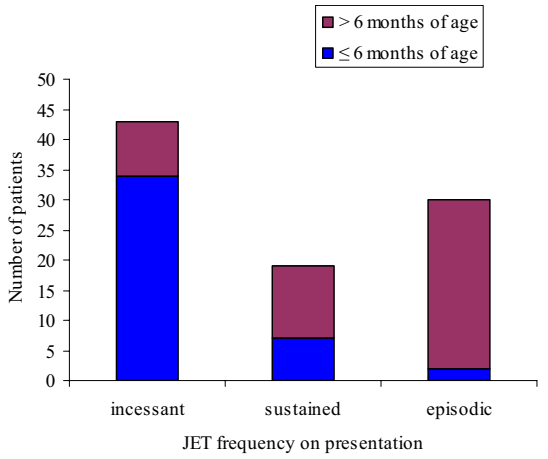

Figure 2 Frequency of JET on Presentation

This graph depicts the number of patients with incessant, sporadic, and episodic junctional ectopic tachycardia (JET). The **bars** are further divided by age at presentation of ≤6 months **(blue area)** and >6 months **(pink area)**. Patients who were age ≤6 months were more likely to have incessant JET, whereas patients age >6 months were more likely to have episodic JET ($p < 0.0001$).

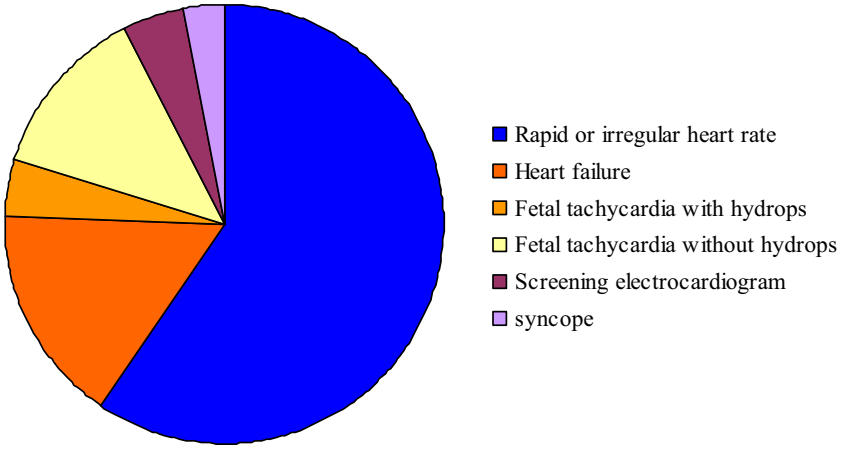

Figure 1 Presenting Signs or Symptoms for Nonpost-Operative Junctional Ectopic Tachycardia

The presenting signs or symptoms are shown for 94 pediatric patients with nonpost-operative junctional ectopic tachycardia. **Blue** = rapid or irregular heart rate; **red** = heart failure; **orange** = fetal tachycardia with hydrops; **yellow** = fetal tachycardia without hydrops; **pink** = screening electrocardiogram; **lavender** = syncope.

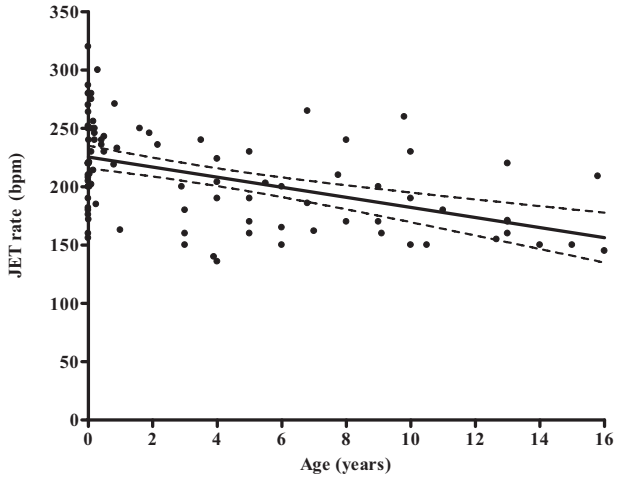

Figure 3 JET Rate on Presentation as Function of Patient Age

This graph shows the relationship of junctional ectopic tachycardia (JET) rate (beats/min) on presentation with the patient's age (years) at presentation. Patients who presented at a younger age had higher JET rates ($r^2 = 0.232$, $p < 0.0001$).

successfully affecting the rate or frequency of JET, amiodarone alone or in combination was the most frequently cited medication, at 60% (41 of 68) (Table 3).

Ablation therapy for JET. Of the patients with JET, 49 patients underwent invasive electrophysiology studies with the intention of catheter ablation of the junctional focus, with 10 patients requiring more than 1 procedure. Radiofrequency ablation of JET was performed in 17 and cryoablation in 26 patients. Other procedures included both RF and cryoablation of JET at the same procedure (n = 2), RF ablation for an accessory pathway (n = 2), and electrophysiology study without ablation (n = 2). Catheter ablation of the atrioventricular junction was performed in 3 patients, 1 as a primary procedure and 2 after JET recurrences. Two patients had ablation for atypical atrioventricular nodal reentrant tachycardia, 1 at the same procedure as JET and 1 as an initial procedure with JET diagnosed later in the course.

Table 2 Antiarrhythmic Medications Utilized for the Treatment of JET

Medication	No. of Patients on Medication
Beta-blockers	53
Atenolol	20
Carvedilol	1
Esmolol	6
Metoprolol	2
Propranolol	24
Amiodarone	50
Digoxin	44
Flecainide	22
Procainamide	11
Sotalol	11
Propafenone	6
Verapamil	5
Lidocaine	4
Diphenylhydantoin	1
Mexiletine	1
Quinidine	1

JET = junctional ectopic tachycardia.

Table 3 Medications Reported to be Successful in the Treatment of JET

Medication	Number of Patients
Amiodarone (total)	41
As a single agent	20
Plus beta-blocker	6
Plus digoxin	3
Plus digoxin plus procainamide	5
Plus digoxin plus beta-blocker	1
Plus flecainide	4
Plus propafenone	2
Digoxin	1
Beta-blocker	9
Digoxin plus beta-blocker	2
Flecainide	3
Flecainide plus verapamil	1
Propafenone	1
Propafenone plus digoxin	1
Sotalol	2
Procainamide	1
Not specified	6
Total	68

JET = junctional ectopic tachycardia.

Data comparing RF ablation to cryoablation reflect information for the first ablation procedure performed at the institution. In the cryoablation group, the 1 patient with AJR and cryoablation is also included in the analysis (Table 4). The indication for ablation was largely heart failure or refractory JET for patients undergoing RF ablation and elective for those undergoing cryoablation. A 3-dimensional mapping system was utilized in 4 patients with cryoablation. Radiofrequency or cryoablation lesions were placed during sinus or atrial-paced rhythm in approximately one-third of the patients and in JET for two-thirds. Cryoablation catheter tip size was evenly divided between a 4- and 6-mm tip. A freeze-thaw-freeze was utilized in 7 patients.

Initial success (82% to 85%) and recurrence rates (13% to 14%) were similar for RF and cryoablation. In 2 patients who received both RF and cryoablation during the same procedure, 1 had JET recurrence and the other has been tachycardia-free. Inadvertent permanent third-degree atrioventricular block occurred in 3 of 17 (18%) patients who underwent RF ablation. There was no permanent atrioventricular block with cryoablation. Transient third-degree atrioventricular block occurred in 1 patient with RF ablation and in 5 patients with cryoablation (1 after a successful cryomapping).

Permanent pacemakers in patients with JET. Permanent pacing systems were placed in 14% (13 of 94) patients.

Table 4 Outcomes of Radiofrequency Ablation and Cryoablation for Junctional Tachycardias

	Radiofrequency	Cryoablation*	p Value
n	17	27	
Year of ablation	2002 (1989–2008)	2006 (2003–2008)	<0.0001
Age, yrs	7 (0.05–38)	9 (2.4–36)	0.31
Ablation indication			0.003
Heart failure, JET, medications unsuccessful	5	2	
Refractory JET, medications unsuccessful	10	7	
Medication side effects	0	2	
Elective	2	12	
General anesthesia	12 (71%)	22 (82%)	0.47
JET during electrophysiology study			
Spontaneous	10	20	0.57
Inducible	6	6	
None	1	1	
JET frequency during electrophysiology study			
Incessant	5	9	0.18
Sustained	10	10	
Nonsustained	1	7	
Isuprel needed for JET	6 (38%)	8 (31%)	0.74
Fluoroscopy time, min	9 (3–195)	14 (2–50)	0.28
Procedure time, min	151 (45–220)	165 (20–362)	0.69
Ablation lesions, n	4 (1–32)	7 (1–67)	0.62
Total duration of ablation lesions, s	145 (11–647)	1,247 (240–4,472)	0.0004
Initial success	14 (82%)	23 (85%)	1.00
Transient third-degree AV block	1 (6%)	5 (18%)	
Inadvertent permanent third-degree AV block	3 (18%)	0	0.05
Recurrence after an initially successful procedure	2/14 (14%)	3/23 (13%)	1.00

Data are presented as median (range) or n (%). *Cryoablation group includes 26 patients with junctional ectopic tachycardia and 1 with accelerated junctional rhythm.
AV = atrioventricular; JET = junctional ectopic tachycardia.

Complete atrioventricular block was the indication for pacing in 10 patients: 3 subsequent to ablation of the atrioventricular junction, 4 secondary to inadvertent atrioventricular block during RF ablation with their initial or subsequent procedure, and 3 with spontaneous atrioventricular block. The remaining 3 patients had a placement of a permanent pacemaker for the following indications: 1 had a pacemaker implanted at age 5 weeks for "back-up" pacing, and this was explanted 10 years later; 1 had sinus bradycardia on antiarrhythmic therapy; and 1 had a possible asystolic cardiac arrest.

Comorbidities and complications in patients with JET. Comorbidities or complications were reported for 13 patients during their treatment for JET and none for patients with AJR (Table 5). One of the patients had sustained a cardiac arrest and had an implantable cardioverter-defibrillator placed at age 9 years; this was a patient with possible myocarditis.

Outcomes of patients with JET. Patients were followed up for a median of 4.5 years (range 0.03 to 28 years). Most patients (75%; 70 of 94), were doing well on no current therapy for JET. These patients included 32 with successful ablation procedures, 21 with spontaneous resolution of the JET, 11 with continued JET but with acceptable heart rates, and 6 with permanent pacing and no further JET. Those treated with medications alone were able to be weaned off the medications after a median duration of 3 years (range 0.2 to 10 years).

Twenty patients (21%) continued antiarrhythmic therapy, but with simplification of their medical regimens. Patients remained on regimens of amiodarone (n = 7), amiodarone plus beta-blocker (n = 2), amiodarone plus flecainide (n = 1), beta-blocker (n = 4), digoxin (n = 3),

Table 5 Comorbidities or Complications During the Treatment of Patients With JET

Comorbidity or Complication	Number
Patients presenting age <6 months	
Cardiac arrest	2
Possible myocarditis	1
Prematurity	1
Extracorporeal membrane oxygenation	1
Hemopericardium after transvenous pacemaker implant	1
Possible lung hypoplasia and copious ascites	1
Short gut syndrome from arrest bowel ischemia	1
Tuberous sclerosis	1
Vitamin D deficiency, rickets	1
Insulin-dependent diabetes and hypertension	1
Patients presenting age >6 months	
Cardiac arrest, possible myocarditis	1
Postural orthostatic tachycardia syndrome	1

JET = junctional ectopic tachycardia.

beta-blocker plus digoxin (n = 2), flecainide plus digoxin (n = 1), and propafenone plus digoxin (n = 1).

Deaths. Death occurred in 4 (4%) patients, all of whom presented with JET at age ≤6 months. The presenting JET rate was 205 to 270 beats/ min. Two infants died at ≤1 week of age after presenting with fetal tachycardia and hydrops. A third patient died at age 11 months with complications from heart failure. The fourth infant died suddenly at age 7 months. After a prenatal diagnosis, his JET rate was well controlled on amiodarone and digoxin. Laboratory evaluation indicated hypocalcemia and vitamin D deficiency rickets, for which he was being treated. His sudden death suggested the possibility that electrolyte imbalances in addition to the antiarrhythmic medications could have provoked an acute lethal arrhythmia.

Discussion

This international multicenter study documents the outcomes of 94 pediatric patients with nonpost-operative JET and 5 patients with AJR. To our knowledge, this is the largest published series that reflects the current era management strategies for this problem. As in previous smaller series, JET continues to be difficult to treat. Patient outcomes, however, have improved with medical management (primarily amiodarone), permanent pacing, RF ablation, and cryoablation. Interventions such as ablation of the atrioventricular junction were required less frequently than in prior reports.

Definition of JET versus AJR. The mechanism of JET is thought to be an automatic focus originating at or near the atrioventricular junction. Junctional rates can be quite variable, and range from AJR to the more rapid JET. In review of prior literature pertaining to nonpost-operative JET, there appears to be no specific heart rate cut-off for the diagnosis of AJR versus JET (3–5,14,17). In creating a precise definition of JET for this manuscript, a survey of the authors was conducted with 3 proposed definitions for JET versus AJR based on prior studies in similar but different tachyarrhythmias: 1) JET defined by a junctional rate >95th percentile of heart rate for age; this definition was utilized in a study of the efficacy of amiodarone in a largely post-operative congenital heart disease population (24); 2) JET defined by a junctional rate of >170 beats/min; this definition was utilized in a study evaluating the treatment of JET in the post-operative congenital heart disease population (25); or 3) JET defined by >10% to 20% of the surrounding sinus heart rate; this type of definition was utilized to differentiate accelerated ventricular rhythm from ventricular tachycardia (26). Ultimately, the authors agreed upon the definition of JET that included >95th percentile of heart rate because it factored in both tachycardia rate and patient age and also had clinical significance, with patients having AJR likely being less affected than those with JET.

Clinical outcomes comparison with prior reports. The largest prior report of nonpost-operative JET was published by Villain et al. (5) and described 26 infants with JET between 1970 and 1987. In that article, atrioventricular junction ablation was performed in 23%, pacemakers were placed in 15%, and death occurred in 35%. In comparison, our patients had improved clinical courses, with atrioventricular junction ablation performed in only 3%, a similar number of pacemaker placements at 14%, and a lower risk of death at 4%. There are, however, several differences in the patient populations. In the prior study (5), only infants age ≤6 months were included. Of these, 61% had decreased ventricular function and congestive heart failure. Secondary to the different eras of the studies, none of the patients in that study had RF or cryoablation for JET. Also, several of the atrioventricular junction ablations were completed surgically. In the present study, we included all patients ≤18 years of age who were diagnosed with JET. Only 20% presented with decreased ventricular function or hydrops secondary to fetal tachycardia.

Consistent with the study by Villian et al. (5) is the findings that patients who present at age ≤6 months are more likely to have incessant JET and faster JET rates. These youngest patients were also more likely to have associated comorbidities and a higher risk of death. At last follow-up, 75% of the patients in the current study were doing well on no medications for JET. This outcome is primarily due to ablative therapies, but also includes cases of spontaneous remission and patients in whom the JET persists at a slower rate that is not causing clinical symptoms or hemodynamic compromise. A smaller case series (3) reported 9 patients with JET, all of whom presented at age ≤6 months with incessant JET and were able to be medically managed. Despite a long follow-up period of 3 to 21 years, none was able to be weaned from medications. While there may be some differences in patient population between the prior studies and ours, the differences in outcomes may likely be due to the more widespread use of amiodarone and the advancement in ablative technologies.

Antiarrhythmic medications. Reflecting the difficulties in treating JET and the multicenter nature of the present study, patients were prescribed a variety of antiarrhythmic therapies. Many patients required >2 medications, either sequentially or in combination, to eventually see a salutary drug effect on JET rate. As in prior studies, our findings support the use of amiodarone for the treatment of JET (3,5). For patients for whom medications were thought to be effective for tachycardia conversion or rate control, amiodarone was the most frequently cited medication. Still, many patients required amiodarone in combination with a second agent.

Ablation. The first reported RF catheter ablation in a child was in 1990 (21), in a patient with JET who had an initial successful ablation but required subsequent atrioventricular junction ablation. Subsequently, other case reports have shown successful ablation of JET with preservation of atrioventricular nodal conduction (2,8,11–13,18–21,27,28). With radiofrequency ablation in close proximity to the

normal conduction system, however, there remains a significant risk of complete atrioventricular block, which would require the placement of a permanent pacing system. In the multicenter Prospective Assessment After Pediatric Cardiac Ablation study (29), only 9 cases of RF ablation of JET were reported in the "not cohort-eligible" registry. In these patients, there was a 100% success rate and an 11% rate of atrioventricular block. By comparison, in the present study, 17 patients underwent RF ablation for JET, with an initial success rate of 82% and a risk of inadvertent atrioventricular block of 18% (3 of 17).

In the early 2000s, catheter cryoablation became available as an alternative energy source to RF ablation. Cryoablation has specific advantages of increased safety when used near the normal conduction system. Previously, cryoablation of JET has been reported in 8 pediatric patients (14,17,30), with an initial success rate of 100% and no reported inadvertent atrioventricular block. In the present study, 27 patients have undergone cryoablation procedures for JET. When compared with RF ablation, cryoablation was more likely to be an elective procedure in patients with sporadic or sustained JET. Cryoablation resulted in an initial success rate of 85%, with no atrioventricular block. The recurrence rate for both RF ablation and cryoablation of JET were similar at 13% to 14%.

Intentional atrioventricular junction ablation was performed infrequently in our JET population. Only 3 patients underwent this treatment, and these were earlier in the study. Unlike prior reports of JET therapy, the atrioventricular junction ablations were carried out with a transcatheter technique.

The patients in the present report appeared to benefit significantly from ablation procedures. While the choice between RF ablation and cryoablation should be at the discretion of the electrophysiologist, it seems prudent to err on the side of safety with cryoablation technology.

Pacemakers and atrioventricular block. A total of 14% of patients in the present study required permanent pacemakers. This outcome is similar to that in the 1990 study by Villain et al. (5). In our patients, the indication for pacing was largely atrioventricular block secondary to RF ablation of JET or atrioventricular junction ablation. Only a few patients had spontaneous atrioventricular block. Dubin et al. (10) described a possible association between congenital JET and congenital atrioventricular block with anti-Ro and anti-La antibodies in the mother. In our population, the history of maternal lupus or maternal anti-lupus antibodies was largely unknown.

Deaths. The mortality rate for JET has been previously reported by Villain et al. (5) to be 34%, with 2 of 9 being sudden unexplained deaths in infants thought to have the JET rate controlled with medications. In contrast, Cilliers et al. (3) showed no deaths among 9 patients with JET. In our population, we report a 4% mortality; all deaths occurred among patients presenting at age ≤6 months. Only 1 death was sudden and may have been secondary to possible electrolyte imbalances in a patient being treated with antiarrhythmic agents.

Study limitations. This was a retrospective review, with associated biases of retrospective studies. Among the 22 institutions, there was no standard approach to JET management. With medical management, the dose and duration of therapies were not specifically recorded.

Conclusions

This international multicenter study of nonpost-operative JET in pediatric patients demonstrates improved outcomes with the current available therapies of medical management (primarily amiodarone), permanent pacing, RF ablation, and cryoablation.

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REFERENCES

1. Garson A Jr., Gillette PC. Junctional ectopic tachycardia in children: electrocardiography, electrophysiology and pharmacologic response. Am J Cardiol 1979;44:298–302.
2. Bae EJ, Kang SJ, Noh CI, Choi JY, Yun YS. A case of congenital junctional ectopic tachycardia: diagnosis and successful radiofrequency catheter ablation in infancy. Pacing Clin Electrophysiol 2005;28:254–7.
3. Cilliers AM, du Plessis JP, Clur SA, Dateling F, Levin SE. Junctional ectopic tachycardia in six paediatric patients. Heart 1997;78:413–5.
4. Sarubbi B, Musto B, Ducceschi V, et al. Congenital junctional ectopic tachycardia in children and adolescents: a 20 year experience based study. Heart 2002;88:188–90.
5. Villain E, Vetter VL, Garcia JM, Herre J, Cifarelli A, Garson A Jr. Evolving concepts in the management of congenital junctional ectopic tachycardia. A multicenter study. Circulation 1990;81:1544–9.
6. Gillette PC, Garson A Jr., Porter CJ, et al. Junctional automatic ectopic tachycardia: new proposed treatment by transcatheter His bundle ablation. Am Heart J 1983;106:619–23.
7. Gillette PC, Garson A Jr., Hesslein PS, et al. Successful surgical treatment of atrial, junctional, and ventricular tachycardia unassociated with accessory connections in infants and children. Am Heart J 1981;102:984–91.
8. Berul CI, Hill SL, Wang PJ, Marx GR, Fulton DR, Estes NA III. Neonatal radiofrequency catheter ablation of junctional tachycardias. J Interv Card Electrophysiol 1998;2:91–100.
9. Darst JR, Kaufman J. Case report: an infant with congenital junctional ectopic tachycardia requiring extracorporeal mechanical oxygenation. Curr Opin Pediatr 2007;19:597–600.
10. Dubin AM, Cuneo BF, Strasburger JF, Wakai RT, Van Hare GF, Rosenthal DN. Congenital junctional ectopic tachycardia and congenital complete atrioventricular block: a shared etiology? Heart Rhythm 2005;2:313–5.
11. Ehlert FA, Goldberger JJ, Deal BJ, Benson DW, Kadish AH. Successful radiofrequency energy ablation of automatic junctional tachycardia preserving normal atrioventricular nodal conduction. Pacing Clin Electrophysiol 1993;16:54–61.
12. Emmel M, Sreeram N, Brockmeier K. Catheter ablation of junctional ectopic tachycardia in children, with preservation of atrioventricular conduction. Z Kardiol 2005;94:280–6.

13. Fishberger SB, Rossi AF, Messina JJ, Saul JP. Successful radiofrequency catheter ablation of congenital junctional ectopic tachycardia with preservation of atrioventricular conduction in a 9-month-old infant. Pacing Clin Electrophysiol 1998;21:2132–5.
14. Law IH, Von Bergen NH, Gingerich JC, Saarel EV, Fischbach PS, Dick M Jr. Transcatheter cryothermal ablation of junctional ectopic tachycardia in the normal heart. Heart Rhythm 2006;3:903–7.
15. Lupoglazoff JM, Denjoy I, Luton D, Magnier S, Azancot A. Prenatal diagnosis of a familial form of junctional ectopic tachycardia. Prenat Diagn 1999;19:767–70.
16. Paul T, Reimer A, Janousek J, Kallfelz HC. Efficacy and safety of propafenone in congenital junctional ectopic tachycardia. J Am Coll Cardiol 1992;20:911–4.
17. Shah MJ, Wieand T, Vetter VL. Cryoablation of congenital familial ectopic tachycardia with preservation of atrioventricular nodal function in an infant. J Cardiovasc Electrophysiol 2007;18:773–6.
18. Simmers TA, Sreeram N, Wittkampf FH, Derksen R. Radiofrequency catheter ablation of junctional ectopic tachycardia with preservation of atrioventricular conduction. Pacing Clin Electrophysiol 2003;26:1284–8.
19. Wu MH, Lin JL, Chang YC. Catheter ablation of junctional ectopic tachycardia by guarded low dose radiofrequency energy application. Pacing Clin Electrophysiol 1996;19:1655–8.
20. Young ML, Mehta MB, Martinez RM, Wolff GS, Gelband H. Combined alpha-adrenergic blockade and radiofrequency ablation to treat junctional ectopic tachycardia successfully without atrioventricular block. Am J Cardiol 1993;71:883–5.
21. Van Hare GF, Velvis H, Langberg JJ. Successful transcatheter ablation of congenital junctional ectopic tachycardia in a ten-month-old infant using radiofrequency energy. Pacing Clin Electrophysiol 1990;13:730–5.
22. Villazon E, Fouron JC, Fournier A, Proulx F. Prenatal diagnosis of junctional ectopic tachycardia. Pediatr Cardiol 2001;22:160–2.
23. Davignon A, Rautaharju P, Boiselle E, Soumis F, Megelas M, Choquette A. Normal ECG standards for infants and children. Pediatr Cardiol 1979;1:123–31.
24. Saul JP, Scott WA, Brown S, et al. Intravenous amiodarone for incessant tachyarrhythmias in children: a randomized, double-blind, antiarrhythmic drug trial. Circulation 2005;112:3470–7.
25. Walsh EP, Saul JP, Sholler GF, et al. Evaluation of a staged treatment protocol for rapid automatic junctional tachycardia after operation for congenital heart disease. J Am Coll Cardiol 1997;29:1046–53.
26. Van Hare GF, Stanger P. Ventricular tachycardia and accelerated ventricular rhythm presenting in the first month of life. Am J Cardiol 1991;67:42–5.
27. Hamdan M, Van Hare GF, Fisher W, et al. Selective catheter ablation of the tachycardia focus in patients with nonreentrant junctional tachycardia. Am J Cardiol 1996;78:1292–7.
28. Rychik J, Marchlinski FE, Sweeten TL, et al. Transcatheter radiofrequency ablation for congenital junctional ectopic tachycardia in infancy. Pediatr Cardiol 1997;18:447–50.
29. Van Hare GF, Javitz H, Carmelli D, et al. Prospective assessment after pediatric cardiac ablation: demographics, medical profiles, and initial outcomes. J Cardiovasc Electrophysiol 2004;15:759–70.
30. Papez AL, Al-Ahdab M, Dick M Jr., Fischbach PS. Transcatheter cryotherapy for the treatment of supraventricular tachyarrhythmias in children: a single center experience. J Interv Card Electrophysiol 2006;15:191–6.

Key Words: junctional ectopic tachycardia ■ arrhythmia ■ amiodarone ■ radiofrequency catheter ablation ■ cryoablation ■ child.