RIGHT VENTRICULAR OUTFLOW TRACT TACHYCARDIA IN CHILDREN

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Objective To assess the clinical spectrum of right ventricular outflow tract tachycardia and its management in children.

Study design Five centers identified patients for retrospective review. Patients (age <18 years) demonstrating ventricular tachycardia with an inferior axis and left bundle branch block were included. Patients with structural heart disease, myocarditis, cardiomyopathy, or long QT syndrome were excluded. Demographics, clinical presentation, investigations, and treatment were analyzed. Holter data were used to quantify ectopy.

Results Patients (n = 48) were referred for evaluation of incidental findings (39/48), near syncope or syncope (7/48), or other (2/48). Investigations included magnetic resonance imaging (51%), endomyocardial biopsy (25%), and angiography (23%). Medical treatment was initiated in 26 of the 48 patients. The most common indications for treatment were frequent ectopy and symptoms. Medical treatment (P <.007) and observation alone (P <.02) were both associated with a reduction in ectopy. Symptoms persisted in 3 of 13 patients who were treated medically and in all untreated patients. At follow-up, there were no deaths and no difference in ectopy (P <.46) between patients who were treated medically and patients who were observed. Ablation was attempted in 6 of the 48 patients (successful in 4/6).

Conclusion The clinical spectrum and management of right ventricular outflow tract tachycardia in children are diverse. Both medical therapy and observation alone were associated with a reduction in ectopy. *(J Pediatr 2006;149:822-6)*

Ventricular tachycardia (VT) may occur with a broad range of circumstances in children. Accordingly, VT has a variable clinical course, ranging from benign asymptomatic ectopy to malignant tachycardias causing life-threatening hemodynamic compromise. VT occurring in patients with structurally normal hearts is reported to be associated with a favorable prognosis.[1,2] Such idiopathic VT tends to arise in the left ventricular apex or in the right ventricular outflow tract (RVOT). In children with structurally normal hearts, right ventricular outflow tract tachycardia (RVOT VT) is the most commonly recognized VT and is characterized by findings on an electrocardiogram (ECG) of a left bundle branch block pattern and an inferior QRS axis.[3-5] Although generally considered to have a good prognosis,[6] the natural history of RVOT VT in children is unknown.

The limited literature on RVOT VT in children, consisting primarily of case reports, small series, and retrospective cohort studies,[2,4,7-9] has demonstrated that fewer than half of these patients have symptoms at presentation. Currently no standard diagnostic approach exists, and management is heterogeneous. However, these studies confirm that in a limited follow-up period RVOT VT carries a good prognosis in children. However, the etiology of ventricular tachycardias originating in the RVOT is diverse and includes: RVOT VT, acute myocarditis, cardiac tumors, and arrhythmogenic right ventricular cardiomyopathy (ARVC).[10] We sought to assess the clinical spectrum of RVOT VT and its current evaluation and management in Canadian children.

METHODS

A joint venture between Canadian pediatric electrophysiologists was established to facilitate collaborative research. The Canadian Pediatric Electrophysiology Working Group (CPEWG) includes centers in Quebec City, Montreal (Sainte Justine and Montreal Children's Hospitals), Ottawa, Toronto, Edmonton, and Vancouver. Members of

ARVC	Arrhythmogenic right ventricular cardiomyopathy	MRI	Magnetic resonance imaging
CPEWG	Canadian Pediatric Electrophysiology Working Group	RV	Right ventricular
ECG	Electrocardiogram	RVOT	Right ventricular outflow tract
EP	Electrophysiology	RVOT VT	Right ventricular outflow tract tachycardia
		VT	Ventricular tachycardia

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this group receive referrals from across Canada for care of pediatric rhythm disorders. All members of the CPEWG were invited to participate in the study. Five of 7 eligible centers reviewed patients for enrollment. Each center selected patients from pre-existing coded databases for review; at some centers, electrophysiologists identified cases from their clinical practice for which coded databases were not available or recently implemented. Ethics approval was obtained by clinical research ethics boards at each of the institutions involved in the study.

The inclusion criteria were: patients <18 years old at presentation, absence of structural heart disease (patent ductus arteriosus and patent foramen ovale were not considered structural heart disease), ECG evidence of monomorphic VT with an inferior axis, and left bundle branch block pattern (Figure 1; available at www.jpeds.com). Patients with acute myocarditis, cardiomyopathy, electrolyte abnormalities, or long QT syndrome at presentation and patients who had an established diagnosis of ARVC according to the McKenna criteria[11] were excluded.

A standardized database was developed to collect data pertaining to demographics, reason for referral, symptoms, clinical findings, investigations (including ECG, Holter monitor, echocardiogram, exercise test, magnetic resonance imaging (MRI), angiography, and biopsy), management, and outcomes (for which follow-up data were available). This database includes all patients with RVOT VT born after Jan 1, 1980, who have been seen at participating CPEWG centers. Data collections were conducted at each site by using a standardized database and an associated coding document to limit variability in data entry. A single investigator reviewed all data and made the final decisions about inclusion and exclusion.

Holter monitor data were used to quantify ectopy and monitor progression of ectopy (using the percentage of QRS complexes that were ectopic). Holter monitor reports were analyzed for maximum ventricular rate, number of premature ventricular contractions, number of couplets and episodes of VT, and percent of ectopic beats. Echocardiogram reports were analyzed for septal thickness, right ventricular diameter, left ventricular end-diastolic and end-systolic dimensions, left ventricular ejection fraction and subtle right ventricular abnormalities, such as dilatation of the right ventricular (RV) apex or outflow tract. MRI reports were analyzed for descriptions of fatty infiltrate, fibrosis, aneurysm, ventricular wall thinning, or excavation. Biopsy reports were reviewed for evidence of fibrosis or fatty infiltrate. Angiogram reports were analyzed for trabeculation, dilatation of the pulmonary outflow tract, focal dyskinesia, and apical hypokinesia.

In patients requiring medical or interventional treatment, the indication for treatment was determined. The effect of treatment was assessed for change in ectopy on the basis of Holter monitor data and change in symptoms. Adverse effects of treatment were recorded.

Statistical Analyses

The data were entered into a Microsoft Excel spreadsheet (Microsoft Corporation, Redmond, Wash) and then imported into Statistical Analysis System software (SAS version 8.02, SAS Institute, Cary, NC) for analysis. Frequency tables were generated for categorical data. When appropriate, 2-by-2 contingency tables were created, and χ^2 analysis was used. Univariate analysis was used to analyze continuous variables with the median (range) of values reported. Group differences were assessed by using a Wilcoxon Rank Sum Test. An alpha level of <0.05 ($P < .05$) was considered to be statistically significant.

RESULTS

Forty-eight patients (25 female) with RVOT VT were identified from the 5 participating centers. The median age at presentation was 8.2 years (0.1-17.0 years). Three patients were discharged after the initial visit, 12 patients await follow-up, and 33 patients were seen on multiple occasions, and their complete follow-up data are presented. The median follow-up period was 22 months (1-210 months). The most common reason patients were referred for a cardiology evaluation was an incidental finding of an irregular heart rhythm or abnormal results on an ECG (82%). However, 7 of 48 patients (15%) were referred because of syncope or near-syncope. Nineteen patients (40%) had a history of exercise intolerance, and 5 patients (10%) had a family history of arrhythmia.

Twenty-two patients (46%) reported symptoms when initially examined by a cardiologist. In addition to conventional cardiac symptoms, myriad non-descript symptoms were reported, including abdominal pain, asthma-related symptoms, fatigue or malaise, fever, limb pain, and rash. Cardiac symptoms persisted in 7 of the17 patients with initial symptoms for whom complete follow-up data was available. Symptoms resolved in 10 of the 17 patients with initial symptoms, all of whom were treated medically. There was no association between the presence of symptoms and arrhythmia burden (% ectopy on Holter; $P > .05$). Overall, there was a significant reduction in the prevalence of symptoms ($P < .04$) from initial presentation to the most recent follow-up. However, new symptoms did develop in 2 patients during the course of follow-up. Apart from the findings associated with ectopy or a flow murmur, the cardiovascular examination was normal in all 48 patients.

A number of clinical investigations were undertaken at the time of initial presentation, with the median length of time of the investigation reported in Table I (available at www.jpeds.com). Predictably, an ECG, echocardiogram, and Holter monitor were routinely ordered on the day of initial presentation. The echocardiogram results were normal in 46 of 47 patients (98%). One patient had reduced ventricular function that normalized on resolution of the ectopy.

Holter monitoring was performed in 88% of patients, with a median length of monitoring of 24 hours (8.1-24.7 hours). Overall, there was a reduction in ectopy from a median of 19.0% (0-87.0%) at the initial visit to 0.3% (0-40.0%)