Analysis of idiopathic ventricular tachycardia in childhood

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Idiopathic ventricular tachycardia (VT) is a relatively benign and rare form of VT. It is seen in young people without demonstrable cardiac pathology. The aim of our study was to review the clinical picture of idiopathic VT, before evaluating the indications for antiarrhythmic treatment and the efficacy of radiofrequency ablation (RFA). The notes of patients diagnosed with idiopathic VT in the last 13 years (n: 22) were included in the study. The median age of onset was 11 years (1 month-16 years). We evaluated the findings regarding the diagnosis, treatment and prognosis of these patients. The most common initial symptom was palpitation, in 15 cases. Five children with idiopathic VT were symptom-free. VT was of right ventricular origin in 10 patients and left ventricular origin in 8 patients. β-blockers were the mainstay of medical treatment in right VT and calcium channel blockers (Ca-channel blocker) were mostly used in left VT cases. The success rate of RFA was 57% in right VT and 100% in left VT. The median follow-up was 41 months (9 months-60 months), and all patients are alive currently with no symptoms. VT without demonstrable cardiac pathology is associated with a good prognosis. Treatment is unnecessary for asymptomatic non-sustained VT. RFA is useful in patients with symptomatic drug-refractory idiopathic VT arising from the left or right ventricle.

Key words: idiopathic ventricular tachycardia, childhood, radiofrequency ablation.

Ventricular tachycardia (VT) refers to a rhythm that originates from a ventricular ectopic focus as myocytes or Purkinje cells below the bifurcation of the bundle of His. VT is defined as three or more consecutive ectopic beats at a rate greater than normal range for age (in most cases >120 beats/min) of ventricular origin on electrocardiogram. In pediatric patients, VT is less common than supraventricular tachycardia. The incidence of VT was reported as 0.2-0.8 per 10,000 children in a school-based heart disease screening. VT carries a more serious prognosis than supraventricular tachycardia, not only from the hemodynamic disadvantage of the fast ventricular rate, but also, more importantly, because VT typically occurs in abnormal myocardium with suboptimal function or a channelopathy that may be vulnerable to ventricular fibrillation. Most pediatric VT cases are idiopathic without underlying heart disease. If underlying pathology has been carefully excluded, idiopathic VT is a relatively benign form of VT. The aim of this study was to define the clinical characteristics, treatment results and long-term course of patients who were diagnosed with idiopathic VT.

Material and Methods
We retrospectively analyzed the documents of 22 patients who were diagnosed with idiopathic VT between 1997 and 2008 because of the lack of previous data. The patients who had cardiac structural or electrical abnormality or cardiac surgery were excluded. We included all of the patients diagnosed with idiopathic VT in the study. We assessed past medical history, presenting symptoms, 12-lead electrocardiography (ECG), exercise stress test, chest X-ray, echocardiography, Holter monitoring and electrophysiologic study (EPS), treatment modalities, and response to therapy. A positive response was defined as being symptom-free and absence of VT on Holter or exercise stress test.
The exercise stress test was performed using a free protocol with manual increase of speed and incline of the treadmill, step by step, until the patient was unable to continue. Observation of more than three consecutive monomorphic ventricular ectopic beats during any part of the test indicated VT, and we accepted that the treadmill exercise test induces VT. The test was also stopped when the tachycardia began. The EPS was performed under sedation with midazolam. After some measurements at basic intervals, programmed extrastimuli from the high right atrium were given. Extra stimulus of up to three beats were delivered from the right ventricular apex, right ventricular outflow tract (RVOT), and from the left ventricle apex in selected patients to try to induce tachycardia. Isoproterenol infusion was also used in patients in whom tachycardia was not inducible at basal conditions.

**Results**

Twenty-two patients (13 male, 9 female) were diagnosed with idiopathic VT. All subjects had normal cardiac structure and function on echocardiography. The median age of the patients was 11 years (range: 0-16). Twelve of the 22 patients (54%) were older than 10 years. The most frequent initial complaint was palpitation in 15 cases (68%), and one had syncope as well. In addition, one infant was diagnosed with the complaint of prolonged crying. Five patients (22%) had no complaint before VT was documented.

Ventricular tachycardia (VT) was detected by 12-lead ECG in 5 of the 22 cases. Seven cases had frequent premature ventricular contractions on ECG. The VT was induced by exercise stress test in 10/22 cases, and in 8 patients, this was sustained VT. VT was detected in 12/22 cases by 24-hour Holter monitoring. All patients were evaluated with echocardiographic examination and 4 patients with magnetic resonance imaging. The right and left ventricle functions were normal in all. The VT was right ventricular in origin in 10 cases and left ventricular in origin in 8 cases. In 4 cases, the location of VT was not determined.

Antiarrhythmic treatment was instituted in 21 cases. Thirteen of 21 patients had been treated with ß-blockers (1-3 mg/kg/day). ß-blockers were mostly used for RVOT-VT. Eight of 10 patients with RVOT-VT had been treated with ß-blockers, and 5 of these 8 patients reported a positive response to the drug. Two of the 8 left fascicular VT patients and 3 of 4 patients with undetermined VT had been treated with ß-blockers and had a positive response (Table I). Finally, ß-blocker treatment was successful in 10 of 13 cases (77%). Eight of 21 cases had been treated with calcium (Ca)-channel blockers (5-8 mg/kg/day) (2 were right ventricular, 6 left ventricular in origin). All of the 8 cases treated with Ca-channel blockers had a positive response to the drug (100% success rate). Ca-channel blockers were effective in all cases of left fascicular VT. It is of note that Ca-channel blockers are mostly used for left fascicular VT. One patient, who had short nonsustained monomorphic VT periods, was not treated.

Electrophysiologic study (EPS) was performed in 17 of 22 patients. Twelve patients (7 with RVOT-VT, 5 with left VT) underwent radiofrequency ablation (RFA). For VT of right ventricular origin, 4 of 7 RFA procedures were successful. In VT of left ventricular origin, 4 of 5 were successful on the first procedure, while one was treated on the second procedure. In total, 9 of 12 patients (75%) were treated successfully. The success rate was 57% in right VT and 100% in left VT. Noncontact mapping, using EnSite balloon, was performed.

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<th>Table I. Medical Treatment Characteristics of Patients with Idiopathic Ventricular Tachycardia</th>
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<td>Origin of VT</td>
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<td>Right VT</td>
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VT: Ventricular tachycardia. Ca: Calcium.
in 8 patients (4 RVOT-VT, 4 left VT). We had no major complications with RFA therapy. Defibrillator implantation was not necessary in any of the patients.

All 22 patients are alive with no clinical complaints after medical or RFA treatment.

Discussion
Although rare in childhood, VT is an important arrhythmia and should be treated promptly. The hemodynamic consequences of VT depend largely on the presence or absence of myocardial dysfunction\(^1\,^5\). Childhood VT may be a result of cardiac muscle or ion channel diseases like long QT or Brugada syndromes, electrolyte disturbances, drug toxicity, metabolic abnormalities, or as a consequence of cardiac surgery\(^1\). Idiopathic VT is defined as VT arising in a patient without demonstrable structural or electrical heart disease. Most pediatric cases of VT are idiopathic in origin without underlying heart disease compared with that in adults, in whom most VT is associated with coronary artery disease\(^6\). Idiopathic VT was found incidentally in a school-based heart disease screening\(^3\). Idiopathic VT is a relatively benign condition seen occasionally in young patients. In this study, none of the patients had signs of myocardial or electrical disorders related with VT.

Two distinct types of idiopathic VT have been described, as either of right or left ventricular origin\(^1\). The most common form of idiopathic VT is right VT, and approximately 90% arises from an automatic focus on the RVOT, with slow rates (140-190 beats/min) and monomorphic left bundle branch block morphology\(^7\,^8\). RVOT-VT is associated with a good prognosis in those without structural heart disease\(^6\). Severe symptoms are uncommon, syncope is rare, and some patients are unaware of this arrhythmia\(^8\). In the most recent clinical studies on idiopathic VT, no tachycardia-related mortality was observed\(^9\,^13\). In this study, nearly half of the patients who had RVOT-VT had no complaint and were asymptomatic, none of them had syncope, and they were unaware of the arrhythmia before VT was documented. Benign RVOT-VT was distinguished from the arrhythmogenic right ventricular dysplasia based on clinical characteristics, echocardiography and cardiac magnetic resonance imaging. Symptoms tend to be mild and syncope is rare in RVOT-VT\(^14\). If all tests are normal and the patient is asymptomatic, no specific therapy is required for benign RVOT-VT\(^6\). In the present study, for symptomatic RVOT-VT cases, \(\beta\)-blocker therapy was considered. RFA is highly successful for this disorder especially with non-contact mapping method\(^6\). The unsuccessful cases could be due to the relatively small anatomy and aortic cusp focus in our patient group.

A second form of benign idiopathic VT originates from a reentrant circuit along the left ventricular septum, which appears to involve the posterior fascicle of the left bundle branch and is referred to as left posterior fascicular tachycardia\(^15\). This form of VT presents as abrupt-onset VT with superior axis right bundle branch block. Left posterior fascicular VT is typically a reentrant tachycardia and may respond to Ca-channel blockers. It can be called verapamil-sensitive VT\(^15\). In our patients, treatment with Ca-channel blockers was successful. Alternatively, RFA can be performed along the left ventricular septal surface. The success rate of left posterior fascicular VT was approximately 90%, which is the highest for any form of VT in young patients\(^16,^17\). Our results were consistent with this finding.

In conclusion, drug therapy with \(\beta\)-blockers and/or Ca-channel blockers can be useful in patients with idiopathic symptomatic VT. EPS and RFA are useful in patients for symptomatic idiopathic VT arising from the right or left ventricle, who are drug-intolerant or do not desire drug therapy, or for drug-refractory VT. The technique of catheter ablation has been proven safe enough for use in children even if they are asymptomatic. For asymptomatic cases of idiopathic non-sustained monomorphic VT, treatment is not essential if there is careful observation. This is safe enough even in children without symptoms to confirm the type of VT is benign, so they can have an active physical life.

REFERENCES


