Idiopathic Ventricular Tachycardia in Infancy and Childhood
A Multicenter Study on Clinical Profile and Outcome

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OBJECTIVES
The present study intended to evaluate the clinical profile and outcome in a large cohort of pediatric patients with idiopathic ventricular tachycardia (VT).

BACKGROUND
Ventricular tachycardia (VT) without underlying heart disease is rare in childhood. Limited information is available with regard to outcome and indications for long-term antiarrhythmic treatment.

METHODS
A retrospective multicenter study was conducted. Patient data were obtained from the individual centers using a standardized questionnaire.

RESULTS
Ninety-eight pediatric patients with episodes of VT in the absence of structural heart disease were included. Mean age at first manifestation of the arrhythmia was 5.4 years (range 0.1 to 15.1), with 27% of the patients having had VT already in infancy. Clinical symptoms or echocardiographic signs of left ventricular dysfunction were observed initially in 36% of the patients, of which one third (12% of the whole population) presented with severe symptoms (heart failure or syncope). After a mean follow-up of 47 months (range 12 to 182), no patient had died. Twenty-five patients had never been treated with antiarrhythmic drugs. Sixty-three patients were free of VT and did not take antiarrhythmic drugs at last follow-up. Prognosis was better when VT occurred during the first year of life (VT resolution in 89%) compared with VT occurrence beyond the first year of life (VT resolution in 56%; p < 0.01). The clinical profile was more favorable for patients with presumed right VT (VT resolution in 76%, symptoms in 25% of patients) compared with patients with presumed left VT, where VT resolution occurred in 37% and symptoms in 67% of patients (p < 0.01).

CONCLUSIONS
VT in children with a normal heart carried a good prognosis. Outcome was better after onset of VT during infancy and when VT originated in the right ventricle. A restrictive use of antiarrhythmic agents might be justified in a large proportion of these patients. (J Am Coll Cardiol 1999;33:2067–72) © 1999 by the American College of Cardiology

Idiopathic ventricular tachycardia (VT) refers to a clinical entity observed in children and adults, in whom conventional diagnostic evaluation fails to detect structural heart disease. For years, reports on small pediatric series have focused the attention on that arrhythmia (1–7). It was found that idiopathic VT basically carried a good prognosis, but due to the small numbers of patients reported, the risk profile of that condition still awaits clarification because the reports published have only included between 4 and 26 children with VT and normal hearts (3,5).

Guidelines for treatment of such patients are far from being established; it was advocated that therapy could be withheld in most patients (3,5) or that only symptomatic children should be treated (4). Conversely, radiofrequency ablation has been shown to be an effective treatment option in such patients, and even asymptomatic children had been treated that way (8), an approach that did not remain unchallenged (9).

We attempted to further elucidate the clinical profile and the outcome of infants and children with idiopathic VT. On the basis of a multi-institutional experience, we are reporting on the largest population of pediatric patients with idiopathic VT.

METHODS

Patient selection. The study was conducted by members of the Working Group on Arrhythmias and Electrophysiology of the Association for European Pediatric Cardiology (see Appendix for list of participating institutions). Children diagnosed and followed at one of the participating centers

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for the occurrence of ventricular tachycardia were studied. The VT diagnosis was based on accepted criteria (10,11) in any child showing at least three consecutive ventricular complexes at a rate clearly faster than the child’s sinus rhythm, and documented on surface electrocardiogram (ECG) or a Holter monitor. Episodes of VT were defined as sustained when lasting >30 s, otherwise, they were defined as nonsustained.

Children were included if they presented with documented episodes of VT between January 1985 and December 1995. Patients of the whole pediatric age range (neonatal up to ≤16 years) at the time of first manifestation of VT were included. Inclusion required a follow-up of each individual child of >1 year; shorter follow-up periods were only considered for inclusion in case the affected child had died. The children had to be studied with repetitive Holter monitors and echocardiograms. Exclusion of structural heart disease was required at least by a complete two-dimensional and Doppler echocardiographic study.

All patients with signs of structural heart disease of any kind or with previous cardiac surgery, as well as patients with clinical or laboratory signs of systemic or inflammatory disease at initial presentation, were excluded. Patients with chronic systemic disease of any kind were also not eligible for the study.

Patients with so-called accelerated ventricular rhythm, defined as ventricular rate only ≤20% faster than the child’s concurrent sinus rhythm, were excluded (12). Prolongation of the corrected QT interval (≥440) in the resting ECG excluded patients from evaluation for the present study. Finally, patients with catecholaminergic VT (13), defined as occurrence of syncope due to polymorphic VT during exercise, were also not eligible for the present study.

Collection of data. The study was conducted as a retrospective, multicenter evaluation, and data for the individual patient were obtained from the participating centers by means of a standardized questionnaire. The questionnaires of all patients were evaluated by the study coordinators (JPP, TP). Surface ECGs and tracings of VT episodes, also requested for all patients, were reevaluated by the authors for decision of exclusion of patients in a blinded fashion.

Definitions. Resolution of VT was defined as lack of evidence of recurrences clinically and during Holter monitors. Complete success of drug treatment was defined as cessation of documented VT after any given newly introduced drug had reached its steady state. A partial effect of a drug was considered if episodes of VT were significantly less frequent (drop in number of VT episodes of >50%), if the heart rate during VT decreased by >20% or if sustained VT changed to only nonsustained VT episodes after introduction of a new drug.

Statistics. Descriptive data were characterized by the mean (median where appropriate) with standard deviation and the range. Patient groups were compared using the paired or unpaired t test where appropriate. Relative frequencies observed between different groups of patients were compared using the chi-square test; for small sample sizes, Fisher exact test was used. A p value <0.05 was considered statistically significant.

RESULTS

Patients’ characteristics. Data on 127 pediatric patients were collected from 13 European centers. After data evaluation, 98 children from 12 centers were included in the study. Reasons for exclusion of 29 patients were clearcut QTc prolongation in two children and diagnosis of catecholaminergic VT in 10 patients. Accelerated ventricular rhythm was present in four patients. In 10 children, VT was not adequately documented as required. Two children had evidence for arrhythmogenic right ventricular dysplasia and one child experienced one episode of VT only during a diagnostic tilt-table testing. The 98 patients included showed an unspecific sex distribution (53 male, 45 female). Mean age at initial presentation with VT was 5.4 years (range 0.1–15.1) for the whole study population. Twenty-seven patients (27%) were in their first year of life, of which 17 were in the newborn age at initial manifestation (Fig. 1).

Characteristics of VT. Sustained VT was observed in 36 of the patients (36%) and was equally distributed between the age groups (44% of infants and 34% of older children: p = NS). In eight patients (six infants and two older children) VT was almost incessant. No patient had documentation of polymorphous VT. Maximum heart rate
The LBBB pattern largely predominated and was seen which is adequate for VT in patients with normal hearts right bundle branch block (RBBB) morphology for left VT, of the QRS complexes in the ECG during tachycardia: left presumed origin of VT, as defined by the ECG morphology disease and the outcome of patients depending on the heart rate during VT and the occurrence of symptoms. In further analysis was aimed at assessing the course of the disease and the outcome of patients depending on the presumed origin of VT, as defined by the ECG morphology of the QRS complexes in the ECG during tachycardia: left bundle branch block (LBBB) morphology for right VT and right bundle branch block (RBBB) morphology for left VT, which is adequate for VT in patients with normal hearts (14). The LBBB pattern largely predominated and was seen in 71/98 patients (70%, p < 0.01), and was even more prevalent in the infant population (86% of infants) as compared with older children (68% of children older than one year). Ventricular tachycardia with LBBB pattern differed significantly from VT with RBBB pattern in the clinical profile, as only 25% of patients with right VT had symptoms, compared with 67% of patients with left VT (p < 0.01). Mean maximum heart rate in VT was not different between the two groups (175 ± 37 in right VT compared with 182 ± 38 in patients with left VT: p = NS). With regard to final outcome, again, VT with LBBB pattern showed a more favorable profile with resolution of VT at last follow-up in 76% of patients, as compared with VT resolution in 37% of children with RBBB morphology during VT (p < 0.01; follow-up periods not different in the two groups; Table 1).

**Symptoms.** Thirty-six of the 98 patients (36%) showed symptoms at presentation of VT or during follow-up. Symptoms were more frequent in children (30/71 or 38%) than in infants (6/27 or 22%, p < 0.05). In 12 of these symptomatic patients, symptoms were considered as severe: 4 patients experienced syncope due to documented VT, and heart failure was observed in 8 patients; 4 of the infants (15%) as compared with 4 of the older children (6%, p = NS). Mild symptoms in the other 24 patients consisted of dizziness (n = 6), palpitations (n = 12) and exercise intolerance (n = 6). Left ventricular function as assessed by echocardiography during sinus rhythm (decreased ejection fraction or shortening fraction and/or left ventricular dilatation) was impaired in 11 of these patients (6 children and 5 infants) but in none of the asymptomatic patients. It was assessed whether there was any relation between maximum heart rate during VT and the occurrence of symptoms. In children with a VT rate <150/min, 75% (18/24) were asymptomatic, whereas in children with VT rates between 150 and 200, 50% (17/35) were asymptomatic. Of those with heart rates >200/min, again, 50% (6/12) were asymptomatic. Statistically, this gave only a trend but was not significant (p = 0.07). For the infant group, the same analysis showed a significant difference, with one out of 16 infants with VT rates <200/min showing symptoms, compared with 5 out of 11 infants with VT rates >200 (p < 0.01). In symptomatic infants, mean maximum heart rate during VT (226 ± 43 beats/min) was significantly higher than in asymptomatic infants (196 ± 25; p < 0.05).

Compared with the whole study population, where sustained episodes of VT occurred in 36% of patients, sustained VT was observed in a significantly larger proportion of symptomatic patients (20/36 or 56%; p < 0.01).

**Diagnostic workup.** Besides a complete echocardiographic study and a chest X-ray, which was done in all patients, additional diagnostic methods to exclude structural heart disease were used in 31 patients. Nine children had exclusion of structural abnormalities of the heart by magnetic resonance imaging and 22 children had cardiac catheterization for hemodynamic evaluation and angiography. In only 14 patients was a diagnostic electrophysiologic study performed, initially with VT inducible in only five children.

Of the patients presenting with symptoms (n = 36) 16 (44%), in addition to the echocardiographic study, had further diagnostic tests such as magnetic resonance imaging or cardiac catheterization for confirmation of a structurally normal heart.

**Drug treatment.** Of all patients, 25 never received drug therapy; 9/27 infants (33%) and 16/71 children (22%; p = NS). Forty patients received drug treatment initially, which was withdrawn during the course of the disease after a mean time of 23 months (1–142 months). Only three of these patients had a relapse of VT after drug withdrawal.

At last follow-up, 23 patients still took antiarrhythmic drugs, and there was a clear difference with regard to age at first presentation. Only 2/27 patients (7%) with VT manifestation in infancy still required treatment, compared with 21/71 children (30%; p < 0.05) with first manifestation of VT resolution in 37% of children with RBBB morphology during VT (p < 0.05), and was even more prevalent in the infant population (86% of infants) as compared with older children (68% of children older than one year). Ventricular tachycardia with LBBB pattern differed significantly from VT with RBBB pattern in the clinical profile, as only 25% of patients with right VT had symptoms, compared with 67% of patients with left VT (p < 0.01). Mean maximum heart rate in VT was not different between the two groups (175 ± 37 in right VT compared with 182 ± 38 in patients with left VT: p = NS). With regard to final outcome, again, VT with LBBB pattern showed a more favorable profile with resolution of VT at last follow-up in 76% of patients, as compared with VT resolution in 37% of children with RBBB morphology during VT (p < 0.01; follow-up periods not different in the two groups; Table 1).

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### Table 1. Comparison of Clinical Characteristics of Patients Depending on the QRS Morphology During VT

<table>
<thead>
<tr>
<th></th>
<th>RBBB (Presumed Left VT)</th>
<th>LBBB (Presumed Right VT)</th>
<th>p Value</th>
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</thead>
<tbody>
<tr>
<td>Infants</td>
<td>4/27 (14%)</td>
<td>23/71 (32%)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Symptoms</td>
<td>18/27 (67%)</td>
<td>18/71 (25%)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Max. heart rate in VT</td>
<td>182 ± 38 beats/min</td>
<td>175 ± 37 beats/min</td>
<td>NS</td>
</tr>
<tr>
<td>VT resolution</td>
<td>10/27 (37%)</td>
<td>54/71 (76%)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Ablation</td>
<td>4/27 (14%)</td>
<td>5/71 (7%)</td>
<td>NS</td>
</tr>
</tbody>
</table>

LBBB = left bundle branch block morphology; RBBB = right bundle branch block morphology; VT = ventricular tachycardia.
beyond infancy still treated at last observation. Follow-up time was not different between the two age groups.

Of the 73 patients who received antiarrhythmic drugs in the course of the disease, 44 (60%) were managed with one single antiarrhythmic drug, whereas 29 patients had more than two consecutive or concomitant antiarrhythmic agents. Of these, six (all from the same center) even received four to six antiarrhythmic drugs (Fig. 2).

A comparison of the efficacy (complete and partial success) of the antiarrhythmic agents most widely used is shown in Figure 3. The reported success rates were highest for amiodarone (89% of treatments). Sotalol, the most widely used single agent, showed a success rate of 62%. The overall efficacy of beta-adrenergic blocking agents was lowest of all drugs (35%).

Radiofrequency ablation. Ablation was attempted in nine children in four centers. Ablation was conducted by pace mapping. Success was defined as noninducibility of VT immediately and 30 min after ablation. Mean age of the patients at the time of the procedure was 8.4 years (range 2 to 13); there was no ablation attempted in an infant. In five children, the indication for ablation was severity of symptoms together with failure of drug therapy to control VT. In three additional patients, ablation was performed for the presence of almost incessant VT (with left ventricular dysfunction in one child but absence of symptoms in the other two). In one asymptomatic child, ablation was done because of failure of antiarrhythmic drug therapy. Mean heart rate during VT in the patients that underwent ablation (168/min) did not differ from the mean rate of VT in the other children (172/min; p = NS). Of the 9 ablation attempts, 7 were successful. The site of ablation was located in the right outflow tract (n = 5; two failures of ablation) and in the left ventricular aspect of the interventricular septum (n = 4; all successful). Ablation was performed with a mean interval of 20 months (1–58 months) after the first VT manifestation.

Final outcome. After a mean follow-up of 47 months (12–182 months), all the patients included in the study were alive. Of the 98 children, 63 (including the 7 patients successfully ablated) were free of all drugs and free of VT recurrences (Fig. 4). Sixteen additional patients were also free of VT but were still treated with antiarrhythmic drugs. Nineteen patients (19%) still experienced episodes of VT, and out of these children, 10 were not treated at the time of last follow-up, and the remaining 9 patients still had antiarrhythmic prophylaxis.

With regard to overall final outcome, there was no difference between patients with sustained VT compared with patients with nonsustained VT (VT resolution and no medication in 53% of children with sustained and in 60% of patients with nonsustained VT, p = NS). The only significant difference between these two groups was that all attempts at ablation were performed in patients who had experienced sustained VT (p < 0.01).

Clinically, 2/98 patients (2%) were symptomatic at last follow-up: one patient had syncope and the other child

Figure 2. Antiarrhythmic drug treatment. Number of concomitant or consecutive antiarrhythmic agents given to the patients over the whole study period is shown.

Figure 3. Effectiveness of the antiarrhythmic agents most frequently used. Bars indicate the relative success rates (hatched bars, partial effect; filled bars, complete success). Amio = amiodarone; Bbl = beta-blocker, Flec = flecainide; Prop = Propafenone; Sot = sotalol; Vera = verapamil.

Figure 4. Status of the study patients at last follow-up. Sixty-three percent of patients were free of ventricular tachycardia and free of drug treatment, including the seven patients who underwent successful catheter ablation.
showed exercise intolerance; both were scheduled for an ablation procedure. The other 96 patients (98%) were free of symptoms, whether or not they still experienced VT attacks. Echocardiographic evaluation showed that left ventricular function was normal in all patients at last follow-up.

Comparison of age groups. The characteristics and the outcome of the patients were studied with regard to the age at first manifestation of VT: infants (<1 year of age at presentation) (n = 27) were compared with the older children (>1 year at first manifestation: n = 71). Table 2 shows a comparison of relevant data. A significantly larger proportion of children were symptomatic (38% vs. 22% of infants, p < 0.05). The two age groups did not differ in the proportion of patients showing sustained VT. There was also no significant difference in the proportion of patients that were always left untreated. The most significant difference was observed in the outcome of patients: at last follow-up, only two of the infants (7%) were still taking antiarrhythmic drugs, whereas that proportion in the group of older children was 30% (p < 0.05). With regard to resolution of VT, it was observed that in 89% of the infants, VT had disappeared during follow-up, whereas in the older children, resolution of VT was observed in only 56% of patients (p < 0.01).

**DISCUSSION**

Idiopathic VT in the presence of an apparently normal heart is observed in adults and rarely in children, and usually is a monomorphic tachycardia. Different pathophysiological mechanisms (reentry, triggered activity, automaticity) have been shown to be involved (14,15). Single institutional experience usually is small (1,2,5), and generally accepted guidelines on how to manage these children are still lacking. Published pediatric studies on idiopathic VT have shown a favorable prognosis for the children affected (2,3,5,6). The results of the present study confirm the favorable overall prognosis of this condition, and the lack of mortality is an important finding of this large series. After a mean follow-up of almost four years, no child with idiopathic VT had died, only 19% of our patients continued to have episodes of VT and most (98%) of the children were asymptomatic.

**Clinical profile.** There were two main findings of the present study. In contrast with what had been observed in the smaller series of pediatric patients reported so far, age at first manifestation of VT influenced the final outcome, and onset of VT in infancy was associated with a higher proportion of patients in whom VT resolved as compared with older age (>1 year) at initial presentation. Furthermore, in the infant population, symptoms were less frequent. This was proven in a previous study, but in a mixed population of children with and without heart disease and VT (6). Another factor influencing the final outcome was the presumed site of origin of VT. The present study showed that over an equal period of follow-up, VT with a RBBB morphology of the QRS complexes (presumably originating from the right ventricle) was associated with a significantly smaller proportion of symptomatic patients, and that resolution of VT was observed in a higher proportion of patients compared with VT presumably originating from the left ventricle (RBBB pattern of QRS complexes).

**Structurally normal heart.** The definition of a normal heart is a matter of debate. It may be argued that by defining the normal heart based only on echocardiography findings, there might be a number of children included in the present study with subtle structural changes of the heart undetected by echocardiography. For instance, one recent study in adults with only single but frequent premature right ventricular contractions found subtle structural abnormalities in the right ventricular myocardium by magnetic resonance imaging in most patients (16). Several reports found an association between VT in otherwise normal hearts of pediatric patients and biopsy findings compatible with silent myocarditis, and immunosuppressive treatment resulted in improvement or resolution of VT (17,18). Moreover, it has been observed that small intramyocardial tumors, undetected by either echocardiography and hemodynamic assessment at catheterization, were the source of permanent VT in infants (19). Such subtle structural abnormalities cannot be excluded by the design of the present study. On the other hand, 44% of the symptomatic children in the present study had additional diagnostic tests (magnetic resonance imaging or cardiac catheterization), and no such subtle structural changes had been observed. This is in contrast to the findings of a previous study in children where subtle structural changes could be diagnosed at cardiac catheterization in most of the symptomatic patients with idiopathic VT and normal echocardiographic findings (4).

**Treatment.** Whether or not children with idiopathic VT needed drug treatment or even radiofrequency catheter ablation has so far been based on (mostly limited) institutional and individual experience. This was again observed in the present study with quite divergent treatment strategies.
in the centers involved. In adult patients, a cautious use of antiarrhythmic agents directed at the relief of symptoms has been advocated (14). In light of the favorable course and outcome of most patients reported in the present evaluation, it may as well be advised that in children with idiopathic VT, the potential benefit of antiarrhythmic drug therapy should be carefully weighed against its inherent risks. On the basis of the results presented, it may be concluded that the asymptomatic child (including normal left ventricular function and dimensions) with VT in a normal heart should be followed closely, but could be managed without drug treatment. If empiric drug treatment is required, it was found, in accordance with a previous report (4), that the use of amiodarone and verapamil resulted in suppression of VT in a considerable number of patients, whereas treatment with beta-blockers resulted in the lowest success rates. As an alternative option, tailored drug therapy based on the results of an electrophysiologic study could be used (15). As in adults (20), radiofrequency catheter ablation has also been successfully performed as a treatment for idiopathic VT in children (8). The use of this treatment in asymptomatic children has been debated (9), because the long-term effects of ablation in the ventricles of the growing myocardium are not well known at the present time. For the child with symptoms who is not responsive to drug therapy, this option must be considered.

**Study limitations.** Several points need to be addressed. Because of the design as a retrospective and multicenter study, the reported patients reflected different and sometimes divergent treatment strategies, especially with regard to decisions for initiation and withdrawal of drug trials, thus, medical treatment as outlined in the study resulted from different clinical approaches to children with VT, making a comparison of treatments difficult. Diagnosis of VT origin was based in most cases on the analysis of QRS morphology on the surface ECG during tachycardia. Although this has been shown to be adequate for patients with normal hearts, there might be some patients (e.g., septal origin of VT) where origin of VT was misdiagnosed.

**Conclusions.** Ventricular tachycardia in children with a normal heart carried a good long-term prognosis, and in most of the patients affected, VT resolved. The clinical profile and the prognosis were better after onset of VT already in infancy and for VT with a LBBB morphology and thus presumably originating from the right ventricle. As long as there are no symptoms and the left ventricular dimensions and function remain normal, a restrictive use of antiarrhythmic treatment may be justified. If refractoriness of VT to antiarrhythmic therapy occurs, catheter ablation may be indicated in the symptomatic child.

**APPENDIX**

List of participating institutions: Bauersfeld U, MD, Zurich, Switzerland; Von Bernuth G, MD, Aachen, Germany; Bieganowska K, MD, Warsaw, Poland; Ebere T, MD, Ulmer HE, MD, Heidelberg, Germany; Gillor A, MD, Köln, Germany; Hentrich F, MD, Essen, Germany; Lubbers LJ, MD, Amsterdam, Netherlands; Vaksman G, MD, Lille, France; Vagnati G, MD, Milano, Italy; Villain E, MD, Paris, France; Paul T, MD, Hannover, Germany; Pfammatter JP, MD, Berne, Switzerland.

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