Clinical spectrum and long-term course of sustained ventricular tachycardia in pediatric patients: 10 years of experience

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Abstract

Objective: Pediatric ventricular tachycardias (VTs) have heterogeneous etiology and different clinical features. This study aimed to evaluate the clinical spectrum and long-term course of pediatric sustained VTs.

Methods: Patients diagnosed as having sustained VT between 2010 and 2020 were evaluated retrospectively.

Results: A total of 129 patients with VT were evaluated; 74 patients were male, and the median age was 12.5 years (0.25–18 years). Patients were grouped as having idiopathic VT (IVT) [n=85 (65.9%)], cardiomyopathy-associated VT (CMP-VT) [n=24 (18.6%)], catecholaminergic polymorphic VT [n=17 (13.2%)], and myocarditis-associated VT [n=3, (2.3%)]. Palpitations (n=61) and syncope (n=24) were the most common symptoms. VT originated from the right ventricle in 53.6% of the patients. Half of the patients underwent electrophysiological study, 64 patients received radiofrequency ablation therapy, and 29 patients had implantable cardiac defibrillators. During the follow-up, 70.4% of all patients had complete resolution, whereas 19 patients had a partial resolution and 23 patients (19.5%) had stable disease. Monomorphic VTs and VTs with left bundle bunch block were more thriving controlled (p=0.02 vs. p=0.04). In terms of long-term results, no statistical difference was found among the VT groups (p=0.39). Deaths were observed only in IVT (n=1) and CMP-VT (n=8) groups (p<0.001), and the overall mortality rate of pediatric sustained VT was observed at 6.9% in this study.

Conclusion: VTs, which can cause sudden cardiac arrest, are potentially life-threatening arrhythmias. Identifying the heterogeneity of this VT and its peculiar characteristics would facilitate appropriate diagnosis and therapy.

Key words: cardiomyopathy, children, sustained ventricular tachycardia, structural heart disease

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Introduction

Ventricular tachycardia (VT) is an uncommon, potentially life-threatening cardiac arrhythmia that can cause sudden cardiac arrest in the pediatric population (1) and may occur with or without structural heart disease (SHD). Although VT is often associated with ischemic heart disease in adults, pediatric VT is mostly idiopathic, without any underlying heart disease (2). Because the management and prognosis differ according to the underlying etiology, the clinical presentation of pediatric VT is exceptionally variable and requires extensive diagnostic evaluation (3, 4).

This study aimed to define the clinical characteristics, treatment strategies, and long-term course of pediatric patients who were diagnosed as having sustained VT according to the clinical category.

Methods

This single tertiary cardiac center study was performed in children with sustained VT between January 2010 and January 2020. Patients were evaluated for a family history of sudden cardiac death, demographic findings, symptoms, cardiac structure and function, electrocardiogram (ECG), 24-hour Holter monitoring, exercise test, management (acute or long-term)

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HIGHLIGHTS

- Most cases are idiopathic.
- Catheter ablation could be effective in pediatric patients with recurrent VT episodes.
- The long-term outcome achieved only by medical treatment in the patients with the age of < 2 years in IVT group is very satisfying.
- The prognosis is especially good with monomorphic VT and also VT with LBBB.
- The mortality rate of sustained VT was higher in cases with underlying SHD than in others.
- Many cases with underlying SHD are permanent, and some are resistant to treatment.

modalities, and follow-up data. Patients with congenital heart disease, only ventricular premature beats, nonsustained VT (12-lead and/or Holter ECG), and torsades de pointes/ventricular fibrillation were excluded from the study. The 12-lead ECGs and all available 12-lead Holter monitoring data of the patients were reviewed to determine whether VT originated from the right ventricle or the left ventricle. VTs were classified as left bundle branch block (LBBB) pattern, which originates from the right ventricle, and right bundle branch block (RBBB) pattern, which originates from the left ventricle.

VT was determined by the presence of ventriculoarterial dissociation, sinus capture, and/or fusion beats in a surface ECG. VT was defined as monomorphic when it showed similar QRS configuration from beat to beat and as polymorphic when VT showed at least 2 or more different QRS patterns. Sustained VT was described as a tachycardia with consecutive beats that last ≥30 seconds or that requires intervention for termination and originates from the ventricles independent of atrial or atrioventricular nodal conduction (5, 6). Incessant VT was defined as a continuous sustained VT, which recurs promptly despite repeated intervention for termination over several hours (6). Bidirectional VT, which is often seen in catecholaminergic polymorphic VT (CPVT), was defined as VT with a beat-to-beat alternation in the QRS axis (6).

After VT was diagnosed, transthoracic echocardiography (TTE), exercise testing, and cardiac magnetic resonance imaging (MRI) were performed to explore underlying heart disease. To determine structural cardiac abnormalities and cardiomyopathy (CMP) and to evaluate the left ventricle function, TTE was performed as recommended by the American Society of Echocardiography guidelines (7). Modified Bruce treadmill testing (GE Healthcare CASE Cardiac Stress Test device) was performed in suitable patients. Moreover, to determine myocarditis, arrhythmogenic right ventricular dysplasia (ARVD), and other suspected CMPs, cardiac MRI was performed in selected patients.

In the diagnostic evaluation, the patients were grouped into the following categories: idiopathic VT (IVT), CMP-associated VT (CMP-VT), CPVT, and myocarditis-associated VT (MC-VT) (3). Idiopathic VT was defined as VT that occurs in the absence of clinically apparent SHD or any identifiable predisposing causes. Patients who had polymorphic VT triggered by catecholamine during emotional stress or physical activity and who did not have SHD or known associated syndromes were diagnosed as having CPVT. The CMP-VT group comprised patients diagnosed as having ARVD, hypertrophic CMP (HCMP), left ventricle noncompaction CMP (LVNC), and dilated CMP (DCMP).

The treatments for patients were determined as an acute or chronic treatment. In acute treatment, the methods used to terminate incessant or life-threatening VT were determined as intravenous (IV) drug therapy, external electrical cardioversion (CV), or IV drug + external electrical CV. In chronic treatment, methods used for VT treatment were determined as antiarrhythmic drug (AAD) therapy, implantable cardiac defibrillator (ICD), and radiofrequency ablation (RFA) for children with recurrent and drug-refractory VT. Electrophysiological study (EPS) and RFA were performed in severely symptomatic patients with drug-resistant VT. An ICD was placed to patients according to the indications in the current guidelines on the treatment of ventricular arrhythmias (6, 8, 9).

The complete resolution (CR) of VT was defined as an absence of evidence of recurrences clinically or on the recent Holter recording in patients with sustained VT at the initial presentation. The partial resolution (PR) of VT was defined as the sustained VT that changed to nonsustained VT episodes or dropped in the number of VT episodes of >50%, and stable disease (SD) was characterized as VT that persisted and did not change during follow-up (10). Follow-up was defined from the time of the presentation to the time of death or the final clinical follow-up.

Statistical analysis

The one-sample Kolmogorov-Smirnov test was used to test the distribution of each continuous variable for normality. Nonnormally distributed variables were performed using the Kruskal-Wallis test and are expressed as median value (interquartile range). The categorical variables are expressed in frequencies and percentages. The Pearson's chi-square test was used to compare categorical variables. $P<0.05$ was considered statistically significant. SPSS 25 software (SPSS Inc., Chicago, IL, USA) was used for statistical analysis.

Results

General findings

The clinical characteristics of the patients are presented in Table 1. A total of 129 patients with sustained VT were evaluated; 74 patients were male, the median age of initial presentation was 12.5 years (range, 3 months to 18 years), and the median weight was 45 kg (range, 4.6–117 kg). Notably, 8 patients were admitted under the age of 1 year. VT was frequently observed after the age of 5 years (Fig. 1). No statistically significant difference was found among the presentation ages of clinical categories ($p=0.202$) (Table 1). The median presentation age was between 10 and 15 years in all patient groups (Table 1).

Table 1. Clinical and electrocardiographic characteristics and diagnostic tests of the patients with VT

Clinical category	All patients	IVT	CMP-VT	CPVT	MC-VT	*P*-value
Patients, n (%)	129 (100)	85 (65.9)	24 (18.6)	17 (13.2)	3 (2.3)	
Median age (IQR), years	12.5 (0.25-18)	11.9 (0.25-18)	13.7 (0.25-17.6)	12.8 (5.75-17)	14 (10.5-17.2)	0.202
Median weight (IQR), kg	45 (4.6-117)	45 (6-117)	48.5 (4.6-70)	42 (20-70)	40 (40-60)	0.791
Male gender, n (%)	74 (57.4)	46 (54)	17 (71)	9 (53)	2 (66.6)	0.496
Symptoms						
None, n (%)	30 (23.2)	25 (29.4)	3 (12.5)	2 (11.8)	0	0.137
Palpitation, n (%)	**61 (47.3)**	**48 (56.5)**	**10 (41.6)**	1 (5.9)	**2 (66.7)**	**0.002**
Syncope, n (%)	24(18.6)	9 (10.6)	4 (16.7)	**11 (64.7)**	0	**<0.001**
Cardiac arrest, n (%)	8 (6.2)	2 (2.3)	3 (12.5)	3 (17.6)	0	0.05
Fatigue, n (%)	4 (3.1)	0	4 (16.7)	0	0	<0.001
Chest pain, n (%)	2 (1.6)	1 (1.2)	0	0	1 (33.3)	<0.001
Family history: sudden deaths at young age, n (%)	14 (10.8)	4 (4.7)	6 (25)	4 (2.4)		0.099
Median VT rate (IQR), bpm/min	174.5 (120-260)	170 (120-250)	166 (120-260)	190 (170-230)	200 (190-260)	**0.013**
VT type						
Monomorphic, n (%)	97 (75.2)	81 (95.3)	13 (54.2)		3 (100)	<0.001
Polymorphic, n (%)	32 (24.8)	4 (4.7)	11 (45.8)	17 (100)		
QRS morphology of monomorphic VT						
LBBB, n (%)	**52 (53.6)**	**42 (51.9)**	**8 (61.5)**		**2 (66.7)**	<0.001
RBBB, n (%)	45 (46.4)	39 (48.1)	5 (38.5)		1 (33.3)	
LBBB—inferior axis, n (%) (%) (%)	49 (38)	42 (49.4)	6 (25)		1 (33.3)	
LBBB—superior axis, n (%)	3 (2.3)		2 (8.3)		1 (33.3)	
RBBB—inferior axis, n (%)	7 (5.4)	5 (5.9)	2 (8.3)			
RBBB—superior axis, n (%)	38 (39.2)	34 (42)	3 (12.5)		1 (33.3)	
Treadmill test, n (%)			70/129 (54.2)			
VT with exercise, n (%)			31/70 (44.3)			
Cardiac MRI, n (%)			30/129 (23)			
Normal, n (%)			13/30 (43.3)			
ARVD, n (%)			8/30 (26.7)			
LVNC, n (%)			6/30 (20)			
Myocarditis, n (%)			3/30 (10)			

ARVD - arrhythmogenic right ventricular cardiomyopathy; VT - ventricular tachycardia; CMP-VT - cardiomyopathy-associated VT; CPVT - catecholaminergic polymorphic VT; IVT - idiopathic VT; LBBB - left bundle branch block; LVNC - left ventricle noncompaction cardiomyopathy; MC-VT - myocarditis-associated VT; MRI - magnetic resonance imaging; RBBB - right bundle branch block

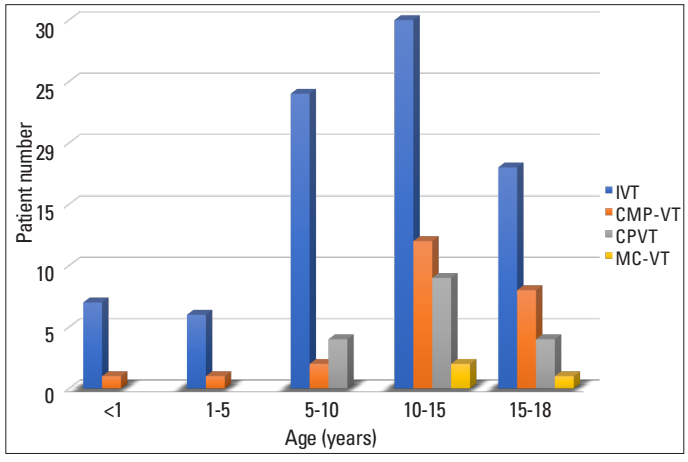

Figure 1. The distribution of the presentation age of ventricular tachycardia
VT - ventricular tachycardia; CMP-VT - cardiomyopathy-associated VT; CPVT - catecholaminergic polymorphic VT; IVT - idiopathic VT; MC-VT - myocarditis-associated VT

Palpitations (n=61) and syncope (n=24) were the most common initial symptoms. Palpitations were seen mainly in children at the age of >9 years (95%). Among 8 patients who presented with resuscitated/aborted cardiac arrest, 2 patients were diagnosed as having IVT, 3 with CPVT, and 3 with CMP-VT (1 patient with HCMP, 1 with ARVD, and 1 with DCMP) (p=0.05). A total of 30 patients (23.3%) had no symptoms at admission.

Family history and diagnostic evaluation

A review of familial history revealed that 14 patients had sudden cardiac deaths in their families at a young age; 4 patients, whose siblings died from sudden cardiac death, presented with syncope and were diagnosed as having CPVT. Although 1 of the 3 patients with a family history of sudden death was presented with palpitations, 2 of them were asymptomatic, and patients were diagnosed as having HCMP. Moreover, 4 patients with a family history of sudden cardiac death were diagnosed as hav-

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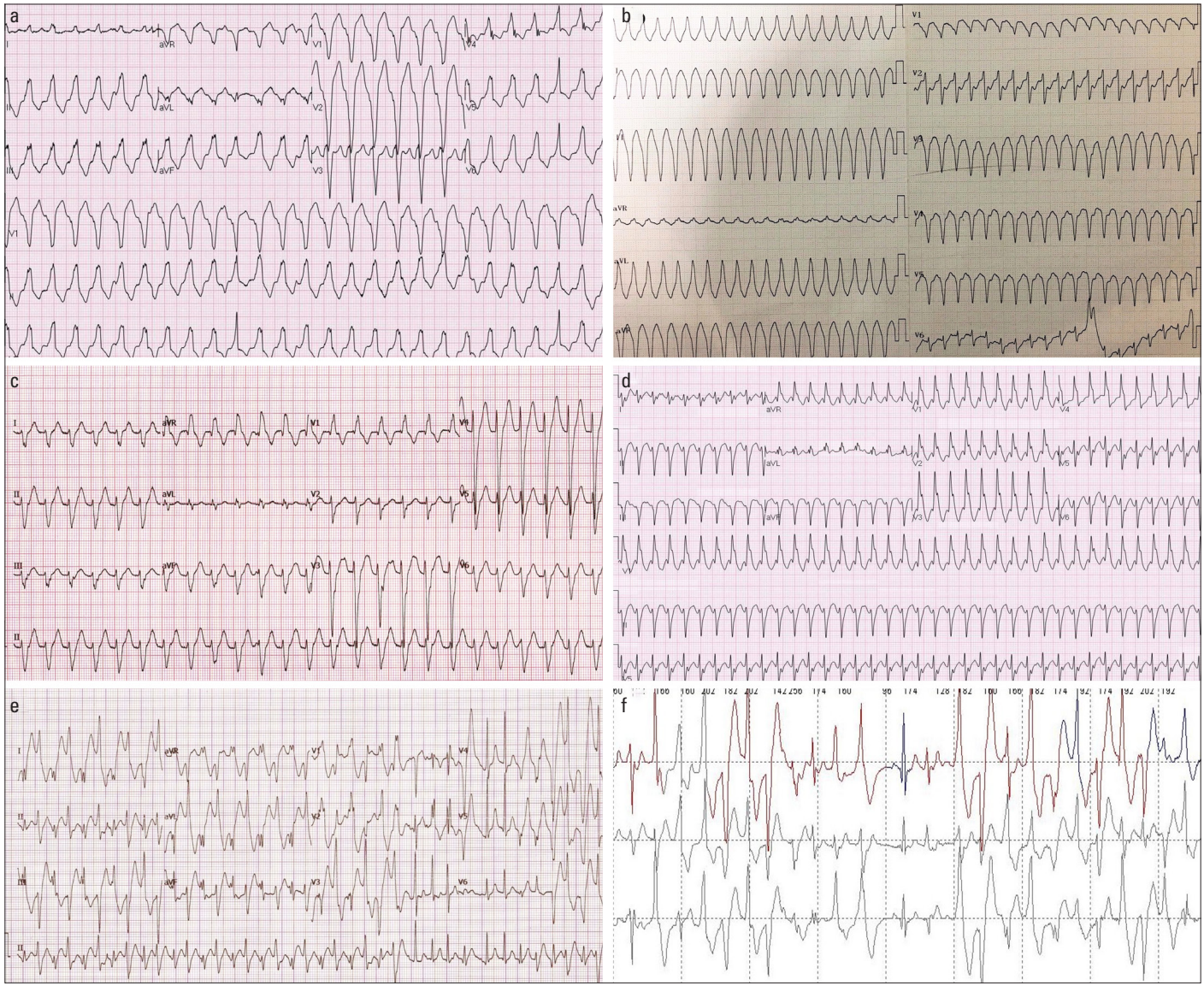

Figure 2. Examples of patients with sustained VT.
(a) Ventricular tachycardia with LBBB and inferior axis in patient with idiopathic RVOT VT.
(b) Ventricular tachycardia with LBBB and superior axis in patient with ARVD.
(c) Posterior fascicular ventricular tachycardia.
(d) Ventricular tachycardia with RBBB and superior axis in patient with HCMP.
(e) Polymorphic VT images of a patient with CPVT on ECG recording.
(f) Polymorphic VT images of a patient with CPVT on Holter monitoring.
VT - ventricular tachycardia; CMP-VT - cardiomyopathy-associated VT; CPVT - catecholaminergic polymorphic VT; IVT - idiopathic VT; LBBB - left bundle branch block; MC-VT - myocarditis-associated VT; RBBB - right bundle branch block; RVOT - right ventricle outflow tract

ing IVT, 2 patients with LVNC, and 1 patient with ARVD; 3 patients with the diagnosis of CPVT were previously diagnosed as having epilepsy, 2 patients were siblings, and 1 patient's brother also had CPVT.

All patients underwent echocardiograms, and the cardiac structure was detected normally in the IVT and CPVT groups. Structural cardiac abnormalities were detected in 44 patients (34.1%). Exercise testing was performed for suitable 70 patients (54.2%), and VT was induced by exercise stress test in 31 cases (44.3%). Medical records revealed that 30 patients underwent cardiac MRI; 8 patients had findings to diagnose ARVD, and 6 patients had LVNC, 3 patients had myocarditis findings. A cardiac MRI of 13 patients was found to be normal. Diagnostic groups and tests of VT patients are presented in Table 1.

Ventricular tachycardia characteristics

The mean heart rate during VT was 175 beats/min (range, 120–260 beats/min). Monomorphic VT and polymorphic VT were detected in 97 (75.2%) and 32 (24.8%) patients, respectively. The type of VT was found to be statistically different among the

Table 2. Treatment and follow-up characteristics of patients

	All patients	IVT	CMP-VT	CPVT	MC-VT
Antiarrhythmic drug, n (%)	129/129 (100)	85/85 (100)	24/24 (100)	17/17 (100)	3/3 (100)
Monotherapy, n (%)	61 (47.3)	51 (60)	6 (25)	2 (11.8)	2 (66.7)
Combined drug therapy, n (%)	68 (53.7)	34 (40)	18 (75)	15 (88.3)	1 (33.3)
Electrophysiological study, n (%)	69 (53.5)	59 (69.4)	8 (33.3)	1 (5.9)	1 (33.3)
Radiofrequency catheter ablation, n (%)	64 (49.6)	56 (65.9)	7 (29.2)	-	1 (33.3)
Implantable cardioverter defibrillator, n (%)	29 (22.5)	2 (2.3)	14 (58.3)	12 (70)	1 (33.3)
Median follow-up period, months (range)	22 (1-120)	11 (1-116)	16 (1-77)	42 (3-94)	9 (1-24)

VT - ventricular tachycardia; CMP-VT - cardiomyopathy-associated VT; CPVT - catecholaminergic polymorphic VT; IVT - idiopathic VT; MC-VT - myocarditis-associated VT

Table 3. Long-term course of ventricular tachycardia

	Total	CR	PR	SD	*P*-value
VT type					
Monomorphic VT, n (%)	91 (100)	64 (70.3)	14 (15.4)	13 (14.3)	0.020
Polymorphic VT, n (%)	27 (100)	12 (44.4)	5 (18.5)	10 (37)	
Total	118 (100)	76 (64.4)	19 (16.1)	23 (19.5)	
Site of origin					
LBBB, n (%)	47 (100)	32 (35.2)	11 (12.1)	4 (4.4)	**0.040**
RBBB, n (%)	44 (100)	32 (35.2)	3 (3.3)	9 (9.9)	
Total, n (%)	91 (100)	64 (70.3)	14 (15.4)	13 (14.3)	

P-values were examined among VT type groups, and among VT origin groups in patients with monomorphic VT.
VT - ventricular tachycardia; CR - complete resolution of VT; LBBB - left bundle branch block; PR - partial resolution of VT; RBBB - right bundle branch block; SD - stable disease

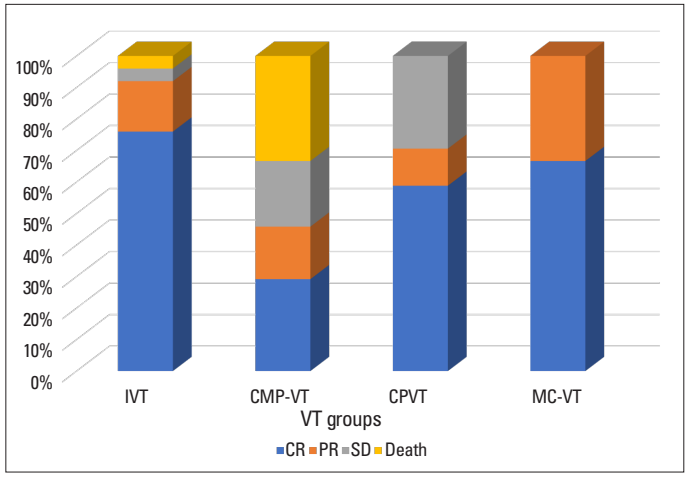

Figure 3. Outcomes of the patients according to the clinical groups
VT - ventricular tachycardia; CMP-VT - cardiomyopathy-associated VT; CPVT - catecholaminergic polymorphic VT; IVT - idiopathic VT; LBBB - left bundle branch block; MC-VT - myocarditis-associated VT; PR - partial resolution of VT; SD - stable disease

clinical groups (p<0.001). VTs with LBBB were observed in 53.6% of patients, with RBBB in 46.4%. This difference was statistically significant among the clinical groups (p<0.001). The ECG characteristics of the VT are presented in Table 1, and examples of VT on 12-lead ECG and Holter recordings are shown in Figure 2. The detailed evaluation of VT subgroups is as follows:

Idiopathic VT group [n=85 (65.9%)]

The median age of IVT was 11.9 years (range, 0.25–18 years), 12 patients were under 2 years of age (infant VT), and 46 patients were male. Polymorphic VT was noted in 4 patients. According to the evaluation of the QRS axis of VT, 42 patients had an LBBB pattern with an inferior axis, which indicates the right ventricular outflow tract (RVOT); 34 patients had an RBBB pattern with a superior axis, which means the left ventricle posterior fascicle; and the remaining 5 patients had RBBB pattern with an inferior axis, which indicates the left ventricle anterior fascicle (Table 1). The left ventricular functions of 5 patients declined at admission and improved by treatment over time. In this clinical group, 31 patients required acute intervention to terminate the incessant or life-threatening VT. Of the 85 patients with IVT, 51 patients received monotherapy, whereas 29 patients received combined AAD therapy with 2 drugs, and 5 patients received combined AAD therapy with 3 prescriptions. A total of 59 patients underwent EPS, and consequently, 56 had RFA. A CR was observed in 8 patients with infant-onset IVT after taking AADs. The median time to disappearing was 19.5 months (range, 1–40 months), and the median age of CR was 2.2 years (range, 0.5–3.8 years). RFA was applied owing to the incessant VT in an infant whose VT was resistant to AADs. An ICD was placed in 2 patients to prevent sudden cardiac death, and the CR was observed in 69.5% of all IVT patients during

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follow-up. Two of the patients died of noncardiac causes. We made telephone conversations with the family of the infant who had been previously treated at our clinic with RFA for multidrug-resistant, recurrent VT episodes. We learned that the patient's palpitations continued despite 3 more sessions of EPS and ablations that were performed at another center, and he died suddenly at home shortly thereafter. We considered this case as sudden death.

Cardiomyopathy-associated VT group [n=24 (18.6%)]

This VT group consisted of 9 patients with ARVD (37.5%), 8 with LVNC (33.3%), 5 with HCMP (20.8%), and 2 patients with DCMP (8.4%). Notably, 11 patients required acute intervention to terminate the life-threatening VT. In the EPS, which was performed in patients with CMP-VT, arrhythmia origin was detected in the left ventricle in 5 patients and the right ventricle in 3. ICDs were implanted in 6 patients with ARVD, 4 with HCMP, 2 with LVNC, and 2 patients with DCMP for a history of syncope, aborted sudden death, or prevention from cardiac arrest. Moreover, 2 patients died suddenly, 3 patients died with heart failure, 2 ARVD patients died with incessant VT and heart failure, and 1 patient who was diagnosed as having ARVD died with multiorgan dysfunction after biventricular assist device implantation.

Catecholaminergic polymorphic VT group [n=17 (13.2%)]

All patients with CPVT had polymorphic, bidirectional VT induced by effort or emotional stress; 11 patients experienced syncope, and 3 patients presented with resuscitated/aborted cardiac arrest. All patients (n=14), except 2, received combined AAD therapy. In our study cohort, ICD implantation was required in 5 patients, left cardiac sympathetic denervation (LCSD) in 2 patients, and ICD implantation with LCSD in 7 patients.

Myocarditis-associated VT group [n=3 (2.3%)]

Three patients diagnosed as having myocarditis presented with sustained VT in the acute stage of the disease. The left ventricular functions of all patients declined at admission. Cardiac functions did not improve in 1 patient after the acute stage, and ICD implantation was performed after the diagnosis of DCMP. Successful left posterior fascicular VT RFA was applied to the patient with improved cardiac functions.

Management and long-term course

Acute intervention in VT was required in 45 patients (34.8%); in 32 patients (24.8%), an intervention was made with IV medication, 6 (4.6%) with the external electrical CV, and 7 (5.4%) with IV drug + external electrical CV. A total of 61 patients (47.3%) were taking 1 drug, and 68 patients were taking combined therapy with 2 or 3 drugs (Table 2). In 64 of 129 patients, the VT was not controlled by antiarrhythmic medication, and consequently, they underwent catheter ablation. In the present study, more than 50% of patients underwent either left posterior fascicle or RVOT RFA. Notably, 29 patients had ICDs to prevent sudden death. The median follow-up time was 22 months (range, 1–120 months) (Table 2). During the follow-up, 70.4% of all patients had CR, whereas 19 patients had a PR, and 23 patients (19.5%) had SD. The outcomes of the patients according to the clinical groups are shown in Figure 3. Monomorphic VT and VT with LBBB were more thriving controlled (p=0.02 vs. p=0.04) (Table 3). In terms of long-term results, no statistical difference was found among the VT groups (p=0.39). The overall mortality rate of pediatric sustained VT was 6.9%, and deaths were observed only in the IVT (n=1) and CMP-VT (n=8) groups (p<0.001).

Discussion

The results of this study describe the clinical characteristics and long-term course of 129 pediatric sustained VT patients from a single tertiary cardiac center. To the best of our knowledge, although there are many studies on IVT (11–13), this is the first study to report about all pediatric sustained VT evaluated with clinical groups from our country. The most remarkable findings in this study are as follows:

1. Most cases are idiopathic.
2. Catheter ablation could be effective in pediatric patients with recurrent VT episodes.
3. Although there were only 12 small patients with the age of <2 years in the IVT group in this study, the long-term outcome achieved only by medical treatment in this unique population is very satisfying, with a CR of 66.7%.
4. The prognosis is especially good with monomorphic VT and also VT with LBBB.
5. The mortality rate of sustained VT was higher in cases with underlying SHD than in others.
6. Many cases with underlying SHD are permanent, and some are resistant to treatment.
7. ICD implantation should be kept in mind as an alternative therapy, especially in sustained VTs that develop in channelopathy and CMPs.

The differential diagnosis for pediatric VT is spacious, and VT may result from electrolyte abnormalities (hypocalcemia, hyperkalemia, and hypomagnesemia), myocarditis, CMPs (DCMP, HCMP, arrhythmogenic right ventricular CMP, LVNC), congenital heart disease, myocardial ischemia, cardiac tumors, and operated congenital heart disease with ventricular scarring. Besides, long QT syndrome, CPVT, and Brugada syndrome are primary electrical diseases that can cause VT in anatomically normal hearts (2). In this study, we described the clinical characteristics and prognosis of pediatric sustained VT, and we demonstrated that these varied according to the VT type, QRS morphology of VT, and clinical categories, which are classified as IVT, CMP-VT, CPVT, and MC-VT. The clinical presentations of patients with VT encompass a broad spectrum, ranging from asymptomatic to incessant VT or sudden cardiac death (6). One-fifth of our patients were asymptomatic, more than half of the patients presented with palpitations, 24 patients presented with syncope, and 8 patients with resuscitated/aborted cardiac arrest.

VT is mostly associated with ischemic heart disease in adults, and IVT accounts for 10% to 20% of VT cases (14, 15). In this study, it was found that most patients (65.9%) diagnosed as having sustained VT were in the IVT group, which was similar to Song et al.'s (3) report. Two different types of VT have been defined: VT that originated from either the right or left ventricle (10). In this report, 53.6% of all patients with monomorphic VT and 50.6% of patients with IVT had LBBB morphology, contrary to a study of pediatric VT patients in which VTs with RBBB configuration were mostly detected (3). Previous studies in adults revealed that the most common form of IVT had LBBB configuration (14, 16). The LBBB pattern of the VT observed quite more than an RBBB pattern, among the clinical groups ($p<0.001$). It is demonstrated that the most common IVT in children originated from the right ventricle, mainly arising from the RVOT, as in those of previous studies in pediatric patients and adults (10, 16, 17). Although IVT is stated to have a good prognosis (10, 11) in our study, 3 patients, including 1 infant, died suddenly, and life-threatening events such as syncope and resuscitated/aborted cardiac arrest occurred in 9 and 2 patients, respectively. Besides, 5 patients had tachycardia-induced CMP at presentation. Idiopathic VT is not an innocent arrhythmia, as mentioned in previous studies (10, 11), and therefore, treatment should be considered as soon as possible after the diagnosis. There are studies indicating that RFA therapy is a good option in addition to AAD therapy in IVT treatment (3, 18). In the present study, RFA was confirmed anew as an effective treatment method in IVT management.

VTs in the setting of CMP are also common forms of VT and form one of the most common electrical mechanisms that cause of sudden cardiac death in CMPs, which can be challenging to manage clinically (19, 20). According to the varying hemodynamic stability of VT, patients with CMP may present with syncope or cardiac arrest to cardiology clinics, and sustained VT is associated with significant morbidity and mortality (19, 21). In this study, 4 patients presented with syncope (ARVD=3, LVNC=1) and 3 with resuscitated/aborted cardiac arrest (ARVD=1, LVNC=1, HCMP=1) in accordance with the abovementioned information. CMP-VT patients are a population whose management is troublesome owing to the heterogeneous underlying diseases. Although there are many treatment options, unfortunately, there is no single treatment method that can cause excellent results in these patients. Pharmacologic therapy has not been shown to decrease mortality in this population but can help to reduce the VT burden. Therefore, combination therapies are often needed for arrhythmia control (22). In our institution, we also use combination therapy in our patients when necessary. Although all patients with CMP-VT were receiving AADs, in 14 patients, ICD was placed to prevent sudden death, and 5 of them underwent catheter ablation for arrhythmia control. However, despite all treatment strategies, 8 patients in this group died because of the CMP's high worsening potential. In the present study, the median follow-up duration for CMP-VT was 19.5 months (range, 1–81 months); hence, a longer follow-up is necessary.

CPVT is a rare inherited arrhythmogenic disorder characterized by stress-induced bidirectional and polymorphic VT in patients with a structurally normal heart and normal resting ECG. This is a life-threatening syndrome, in which the first clinical episode often manifests in the first or second decade of life, usually prompted by physical activity or emotional stress (23). Supporting this information, the median age of the patients in our study was 12.8 years (range, 5.75–17 years); 3 patients presented with aborted sudden death, and 11 patients with syncope. Family history of exercise-related syncope, seizure, and cardiac arrest has been reported in 30% of the patients, and a significant number of patients are followed up with a misdiagnosis of epilepsy before being diagnosed as having CPVT (23). It is observed that 4 patients had a sudden death at a young age in the family, 2 patients had a family history of CPVT, 1 patient had a family history of epilepsy, and 3 patients were misdiagnosed as having epilepsy before the diagnosis of CPVT. According to the current guidelines, beta blockers and lifestyle changes such as exercise restriction and avoiding stressful situations are the first-line treatment options for CPVT patients (23). As soon as the diagnosis was made for all of our patients, we recommended exercise restriction by starting beta-blocker therapy. Two of our patients were using beta blockers only, 14 patients received combined therapy with 2 drugs (beta blocker, flecainide), and 1 patient with 3 drugs (beta blocker, flecainide, calcium-channel blockers). In CPVT patients whose symptoms persist despite optimal medical therapy, ICD (23) and/or LCSD treatments should be considered (24). In our study, in which recurrent syncope occurs despite pharmacologic therapy, ICD implantation was required in 5 patients, LCSD in 2 patients, and ICD implantation with LCSD in 7 patients. De Ferrari et al. (24) announced the LCSD as an effective antifibrillatory intervention for patients with CPVT in their report. During follow-up, we observed that in 10 patients, sustained VT attacks disappeared, 2 patients had a PR, and 5 patients had SD, whereas no mortality was detected in this patient group.

Myocarditis is an "inflammatory disease of the heart muscle," which is commonly caused by viral and autoimmune etiologies, although many cases are idiopathic (25). Children with myocarditis may present with a variety of symptoms, ranging from prodromal symptoms to heart failure, and it remains a diagnostic challenge in the clinical setting (25, 26). Myocarditis can also present in conjunction with ventricular arrhythmias caused by myocardial inflammation and remodeling (26). In our study, we documented that sustained VT was developed in 3 patients diagnosed as having myocarditis. The first patient was a 14-year-old girl; although she was treated with a diagnosis of myocarditis in another hospital's inpatient clinic, she had refractory VT and had received extracorporeal membrane oxygenation support. After the treatment was completed and discharged, the patient started to be followed up from our clinic, DCMP developed, and ICD implantation in addition to antiarrhythmic therapy was performed to prevent sudden cardiac death. A 17-year-old male patient was admitted for chest pain

Anatol J Cardiol 2021; 25: 313-22
DOI:10.14744/AnatolJCardiol.2020.95759

and diagnosed as having myocarditis. Successful left posterior fascicular VT ablation was applied to the patient who underwent CV as an acute treatment but had persisted ventricular arrhythmia. The last patient with the diagnosis of myocarditis was a 10-year-old boy, who received an IV amiodarone, lidocaine, and CV for acute termination of VT. The cardiac functions of all 3 patients declined at the beginning of the diagnosis, and the systolic functions of the patients other than the first patient improved after treatment.

VT is a life-threatening arrhythmia, and urgent management is required. Acute termination of nontolerated VT should be considered in the patient presenting with hemodynamic instability (27). An early termination using defibrillation or R-wave synchronous CV is crucial, as recommended by all available guidelines (8, 9). In patients with hemodynamically compromising VT, which persist after CV/defibrillation, IV amiodarone should be administered to attempt to achieve a stable rhythm (class of recommendation I). In the current guidelines, IV lidocaine has been recommended as beneficial for ventricular fibrillation/polymorphic VT unresponsive to defibrillation (class of recommendation IIb). In patients with hemodynamically stable VT, IV amiodarone or sotalol have been recommended as an effective AAD to attempt to terminate VT (class of recommendation IIb) (6). In the present study, immediate termination of VT was required in 45 patients. Intravenous AADs (amiodarone, lidocaine, calcium-channel blockers, beta blockers) were used in 32 patients, 6 patients received external electrical CV, and 7 patients' acute VT termination was made by pharmacologic and electrical CV. In the chronic management of VT patients, an essential factor is preventing VT recurrences. Discontinuation of proarrhythmic drugs, appropriate antiarrhythmic therapy with drugs, and ablation are the pillar of the management of a manifest arrhythmia (8). All of our patients have received medications, and in the chronic treatment of the patients, pharmacologic treatment, including monotherapy (47.3%) or combined therapy with 2 (45.7%) or 3 (7%) drugs, was used. Beta blockers are the first-line drugs that we preferred in management of VT patients. If the patient needs a second drug, we mostly use flecainide(or sotalol) in combination with the beta blockers. If we think that VT is a posterior fascicular VT, we mostly start treatment with verapamil (if the age is appropriate). Catheter ablation is considered an option for patients with drug-refractory VT and has an essential role in patients with manifest arrhythmias (8, 9, 19, 21). In 64 of 129 patients (49.6%), the tachycardia could not be controlled with antiarrhythmic therapy, and consequently, patients underwent catheter ablation. It was demonstrated that VT was originated from the left posterior fascicle in 45% of the patients, in whom we performed RFA, and from RVOT in 20%. Even though pharmacologic and RFA treatment were the primary therapies for ventricular tachyarrhythmias in the children, ICD is the mainstay treatment and might be the most useful prevention from sudden cardiac death for the patients with a high risk of sudden death as in our study. ICDs were placed in a total of 29 patients, and 23 patients received the device for primary prevention. Although the decision to implant ICDs was given for 4 additional patients, they could not be placed in because familial consent could not be obtained. Song et al. (3) revealed a logistic regression analysis and reported that CPVT, CMP-associated VT, polymorphic VT, and sustained VT were strongly associated with sudden death or cardiac arrest. In the present study, the outcome of CMP-VT was more flawed than the other clinical groups. For the VT type, the monomorphic VT and VT with an LBBB had a better prognosis. The overall CR of the study population was noted as 70.4%. No statistically significant difference was found among clinical groups when compared in terms of long-term results described as CR, PR, and SD (p=0.39).

Although there are many reports on VT, there are few publications on sustained VT in the pediatric population in the literature. Chiu et al. (28) evaluated the clinical characteristics of 116 patients who developed sustained VT diagnosed as having IVT, CMP-VT, SHD-associated VT, and VT associated with channelopathies. In their study, VT with CMP was found to be associated with the highest mortality rate, similarly as in ours (28). Song et al.'s (3) study describes the clinical characteristics and prognostic factors of VT in Korean children. The patients were classified as having IVT, CMP-VT, CPVT, MC-VT, and VT associated with congenital heart disease, which was different from our study. The authors found the overall mortality rate of pediatric VT (nonsustained and sustained) was 7.4% (3). In our study, we demonstrated that the mortality rate of pediatric sustained VT was 6.9%. In our study, we found that 24 patients presented with syncope, 11 of whom were diagnosed as having CPVT, 9 with IVT, and 4 with CMP-VT. Only 1 patient who presented with syncope and were diagnosed as having ARVD died, and the other patients presented with syncope are still alive. We did not find a relationship between death and syncope presentation, because there was only 1 patient who died among the patients who presented with syncope and were diagnosed as having sustained VT.

Study limitations

Our study evaluated the patients with sustained VT through retrospective reviews in a single institution, and the sample size of patients with individual arrhythmias in clinical groups, except the IVT group, is relatively small. Another limitation of this study is that the follow-up time of these patients is short because there are patients who have recently applied and are newly diagnosed.

Conclusion

Sustained VTs, which can cause sudden cardiac arrest, are potentially life-threatening arrhythmias. Pediatric VT is different from adult VT in terms of its features, clinical spectrum, and management. Identifying the heterogeneity of this VT and its peculiar characteristics would facilitate appropriate diagnosis and therapy. The best results can be achieved with the correct

diagnosis and proper treatment according to clinical and tachycardia features.

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Anatol J Cardiol 2021; 25: 313-22
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