Catecholaminergic polymorphic ventricular tachycardia detected by an implantable loop recorder in a child

İmplante edilebilir loop recorder ile tanı konulan katekolaminerjik polimorf ventriküler taşikardili bir olgu

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Summary– We present a six-year-old boy with a history of recurrent syncope whose physical examination and family history were inconclusive. Laboratory findings, 12-lead ECG, chest radiography, Holter monitoring, event recorder monitoring, echocardiography, coronary computed tomography (CT) angiography, Brugada challenge test (ajmaline), cranial magnetic resonance imaging, and awake/sleep electroencephalogram were all unremarkable. Since syncope was exercise-induced, an electrophysiology study was also performed, but revealed no inducible ventricular arrhythmias. Implantable loop recorder (ILR) was implanted. Three weeks later, bidirectional ventricular tachycardia was found in ILR record during presyncope that was related to exercise. The patient, with the diagnosis of catecholaminergic polymorphic ventricular tachycardia, was started on high-dose beta-blocker therapy. Due to the recurrence of syncopes despite the presence of beta-blockers, an implantable cardioverter defibrillator was implanted.

Özet– Bu yazıda tekrarlayan egzersiz ile ilişkili senkop yakınması ile başvuran altı yaşında bir erkek hasta sunuldu. Fizik muayene ve aile öyküsünde özellik yoktu. Tanıya yönelik laboratuvar bulguları, 12 derivasyonlu elektrokardiyografi (EKG), göğüs röntgeni, Holter EKG, olay kaydedici, ekokardiyografi, koroner BT anjiyografi, Brugada sendromu tanısı için ajmalin testi, beyin magnetik rezonans incelemesi ve uyku/uyanıklık elektroensefalogramında özellik yoktu. Hastanın senkopları egzersiz ile ilişkili olduğundan elektrofizyolojik inceleme de yapıldı; ancak ventriküler aritmi uyarılamadı. Hastaya "implantable loop recorder (ILR)" takıldı ve üç hafta sonraki egzersiz ilişkili presenkop sırasında, ILR kaydında iki yönlü ventriküler taşikardi saptandı. Katekolaminerjik polimorf ventriküler taşikardi tanısıyla yüksek dozda beta bloker tedavi başlanan hastanın tedaviye rağmen senkopları devam edince hastaya yerleştirilebilen kardiyoversiyon defibrilatörü uygulandı.

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an inherited arrhythmia syndrome in young patients with structurally normal hearts characterized by bidirectional or polymorphic ventricular arrhythmias under conditions of increased sympathetic activity.[1-4] The baseline electrocardiography (ECG) is usually normal; however, bradycardia and 'borderline' QT interval have been reported.[1] Mutations in the genes encoding the cardiac ryanodine-calcium release channel, or infrequently, cardiac calsequestrin are identified in 50-62% of patients meeting the precise definition of CPVT.[4,5] Clinical diagnosis is made based on family history, exercise- or emotional stress-induced symptoms and -most important- response to exercise or catecholamine infusion. In children who are not able to perform exercise testing, Holter ECG, event recorder, and an implantable loop recorder (ILR) might be of additional help in detecting the typical ECG findings during exercise or emotional stress.[1,5]

Abbreviations:

CPVT	*Catecholaminergic polymorphic ventricular tachycardia*
CT	*Computed tomography*
ECG	*Electrocardiography*
ICD	*Implantable cardioverter defibrillator*
ILR	*Implantable loop recorder*
VT	*Ventricular tachycardia*

In this article, we report a case of CPVT detected by an ILR in a six-year-old boy with a syncopal history and no structural heart disease.

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CASE REPORT

A six-year-old boy was referred to our hospital with a history of recurrent exercise-induced syncope. There was no family history of syncope or sudden cardiac death. No other abnormalities were detected during the physical examination that included detailed cardiovascular and neurological evaluation; neuromotor and growth percentiles were unremarkable. Laboratory findings showed normal complete blood count, serum biochemistry, serum lipid profiles, and cardiac enzymes. A 12-lead ECG (Fig. 1a), chest radiography,

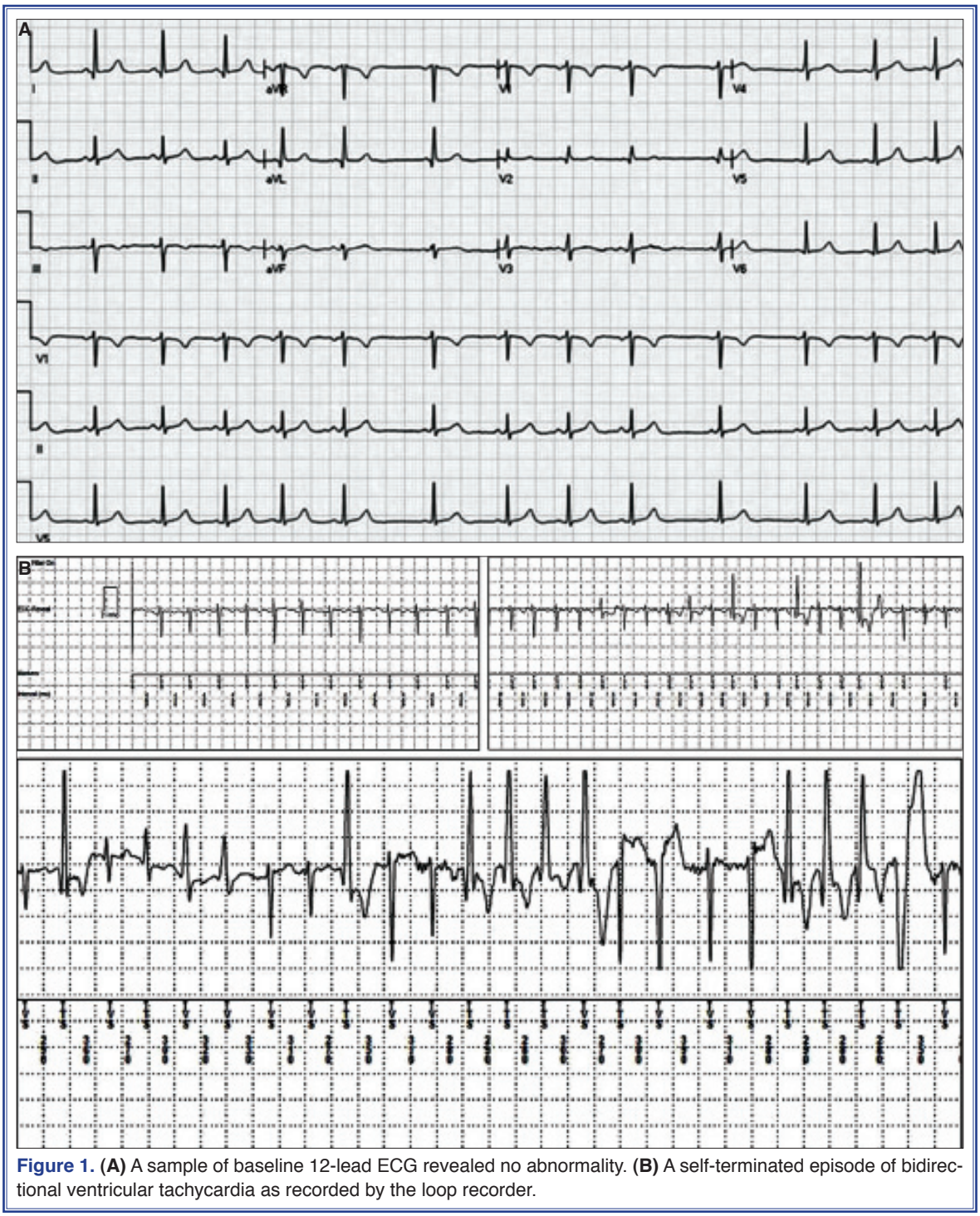

Figure 1. **(A)** A sample of baseline 12-lead ECG revealed no abnormality. **(B)** A self-terminated episode of bidirectional ventricular tachycardia as recorded by the loop recorder.

Holter monitoring, event recorder monitoring, echocardiography, coronary computed tomography (CT) angiography, Brugada challenge test (ajmaline), cranial magnetic resonance imaging, and awake/sleep electroencephalogram were all unremarkable. Due to the young age of the patient, exercise testing (Bruce protocol) could not be considered completely reliable. Since syncope was exercise-induced, an electrophysiology study was also performed. The baseline intervals were as follows: cycle length 710 msec, AH interval 86 msec, HV interval 42 msec, and QT interval 410 msec. Corrected sinus node recovery time was 350 msec, and Wenckebach cycle length was 380 msec. No supraventricular or ventricular arrhythmias were induced during the stimulation protocols (with basic and orciprenaline infusion). Subsequently, an ILR (Reveal DX 9528, Medtronic Inc., Minneapolis, MN, USA) was implanted subcutaneously in the left pectoral region using a small incision to minimize scarring. Surface mapping was done in order to identify the maximum recorded R-wave amplitude and to optimize the quality of the ECG recording. The patient was also started on a beta-blocker treatment for prophylactic purposes. Twenty days after implantation and while the patient was stair-climbing, he suffered a pre-syncopal episode, and his mother activated the device. The stored electrogram, which was retrieved by telemetry, showed a self-terminated episode of bidirectional ventricular tachycardia (VT) (Fig. 1b). The patient was diagnosed as CPVT with the help of ILR record findings and the patient's current medical history. Epicardial implantable cardioverter defibrillator (ICD) was implanted with the written informed consent of the patient's parents in view of persisting syncopes (two more) despite the presence of high-dose beta-blockers (metoprolol, 4 mg/kg/day). The patient remains under observation with exercise limitation and high-dose beta-blocker therapy.

DISCUSSION

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare but highly malignant inherited arrhythmia disorder. It is characterized by VT, which is polymorphic or bidirectional, and induced by catecholamines triggered by physical exercise or emotional stress, typically in the absence of structural heart disease.[1,2] Exercise testing is essential when suspecting an arrhythmogenic origin of syncope, and in the case of CPVT, it may be even more sensitive than Holter monitoring. However, some CPVT patients during early childhood may not have arrhythmias in the exercise stress test, and arrhythmias are seldom inducible by programmed electrical stimulation in the electrophysiologic study.[1-5]

Indeed, CPVT is considered to be a malignant inherited arrhythmia syndrome. In untreated patients, eight-year overall arrhythmic event rates of 58% and fatal or near-fatal event rates of 25% have been reported. The risk of arrhythmic events is considered to be higher in patients with aborted cardiac arrest, younger age, and in patients carrying recessively inherited CASQ2 mutations.[6] However, it is important to conclude that there is a lack of data to identify CPVT patients with such a low risk of arrhythmic events that would make treatment unnecessary. Thus, all phenotypically and/or genotypically diagnosed CPVT patients should receive appropriate therapy.[1,4] In addition, advising against participation in competitive sports and emphasizing the great importance of drug compliance are essential. CPVT patients should also be informed that the use of sympathomimetic agents is contraindicated.[1,4,5] The first step in treating a CPVT patient should be a beta-blocker in the highest tolerable dose.[1-6] Verapamil may be added to beta-blocker therapy, but addition of flecainide is preferred and more effective when beta-blocker therapy fails.[4,7] In patients resistant to combination therapy with beta-blocker and flecainide, either left cardiac sympathetic denervation should be performed or an ICD should be implanted.[1,4] In the 2006 ACC/AHA/ESC Guidelines for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death, a Class I recommendation was given for implantation of an ICD in addition to beta-blocker therapy for CPVT patients who are survivors of aborted cardiac arrest. A Class IIa recommendation was given for ICD implantation in CPVT patients with syncope and/or documented sustained VT despite beta- blocker therapy.[8] ICD treatment without concomitant use of beta-blockers is dangerous because of the risk of electrical storm induced by the adrenergic surge related to a shock.[1]

The ILR can be used as a diagnostic tool in cases of recurrent syncope in which an etiological origin cannot be found. Currently, there are no guidelines for the use of ILRs in pediatric patients. The American College of Cardiology guidelines for ambulatory

ECG suggest that the ILR can be particularly useful in monitoring patients with infrequent symptoms.[9] The European Society of Cardiology guidelines for the management of syncope advise that the conventional route of investigations is preferable and recommend the ILR as a last resort, with the exception of cases with structural heart disease, a family history of sudden death, exercise-induced syncope, chest pain or palpitations, or presence of an ECG that suggests a risk of arrhythmic syncope.[10]

In conclusion, this case demonstrates the value of ILR in diagnosing arrhythmic events in patients with inherited arrhythmogenic diseases associated with recurrent syncope.

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Key words: Child; defibrillators, implantable; tachycardia, ventricular/diagnosis.

Anahtar sözcükler: Çocuk; defibrilatör, takılabilir; taşikardi, ventrikül/tanı.