CASE REPORTS

EXERCISE-PROVOKED BIDIRECTIONAL VENTRICULAR TACHYCARDIA IN A YOUNG WOMAN

Yen-Hung Lin,[1] *Ling-Ping Lai,*[1] [redacted][2] *and Jiunn-Lee Lin*[1]

林廷光（等三作者）

Abstract: Exercise-induced ventricular tachycardia (VT) is rare in children and young adults without structural heart disease. Catecholaminergic polymorphic VT (CPVT) is among the possible causes and carries a poor prognosis. The QRS morphology of CPVT can be bidirectional, polymorphic or even ventricular fibrillation. We report a case of CPVT initially presenting as sudden collapse in an 18-year-old Taiwanese woman. Family history was negative for arrhythmias and sudden death. Laboratory analyses, transthoracic echocardiography, magnetic resonance imaging, electrophysiological study including procainamide and isoproterenol test were all negative. Bidirectional VT was induced by treadmill exercise test. She responded well to β-blocker therapy. Some cases of CPVT are sporadic and some occur in patients with a family history. The treatment of choice for this disease is β-blocker and implantation of an internal cardioverter defibrillator.

Key words: Adrenergic beta-antagonists; Diagnosis; Electrocardiography; Ventricular tachycardia

J Formos Med Assoc 2004;**103**:780-3

Exercise-induced ventricular tachycardia (VT) in children and young adults without structural heart disease is rare. Generally, the prognosis is relatively benign if the morphology is monomorphic.[1] Several entities of exercise-induced polymorphic VT have been identified, including congenital long-QT syndrome, Brugada syndrome, short-coupled variant of torsade de pointes VT and catecholaminergic polymorphic VT (CPVT).[2] CPVT is a special disease entity with multiform QRS morphology during VT and carries a worse prognosis.[1,2] Leehardt et al reported clinical characteristics in 21 patients with CPVT.[2] Most reported cases of CPVT were in Caucasians and Japanese.[1-6] To the best of our knowledge, CPVT has not been reported in Taiwanese. We report a case of CPVT that presented as sudden collapse. Typical bidirectional VT was induced during treadmill exercise test.

Case Report

An 18-year-old Taiwanese girl was evaluated due to an unexplained episode of sudden collapse during an outdoor teaching trip. She lost consciousness suddenly when bending down to adjust her shoes. She was resuscitated by local medical personnel in an ambulance and was sent to the emergency service immediately. Ambulatory electrocardiogram (ECG) recorded by automatic external defibrillator revealed polymorphic and bidirectional VT (Fig. 1). She recovered fully after the resuscitation.

She was otherwise healthy and had never experienced syncope or exercise-induced dizziness. There was no history of heart disease, syncope, or sudden death in her family. Physical examination was unremarkable, as well as laboratory analyses, including biochemical profile and serum electrolytes. The ECG after resuscitation is shown in Fig. 2. Signal-averaged ECG, transthoracic echocardiography, and magnetic resonance imaging revealed no abnormalities. Programmed ventricular stimulation failed to induce sustained or non-sustained VT even with isoproterenol infusion. Procainamide provocation test was negative for Brugada syndrome.

Treadmill exercise test was performed to evaluate the possibility of exercise-induced arrhythmia. After

[1]Division of Cardiology, Department of Internal Medicine, National Taiwan University Hospital, Taipei; [2]Division of Cardiology, Department of Internal Medicine, Buddhist Dalin Tzu Chi General Hospital, Chia Yi, Taiwan.
Received: 1 December 2003 Revised: 6 February 2004 Accepted: 9 March 2004
Reprint requests and correspondence to: Dr. Jiunn-Lee Lin, Department of Internal Medicine, National Taiwan University Hospital, 7, Chung-Shan South Road, Taipei, Taiwan.

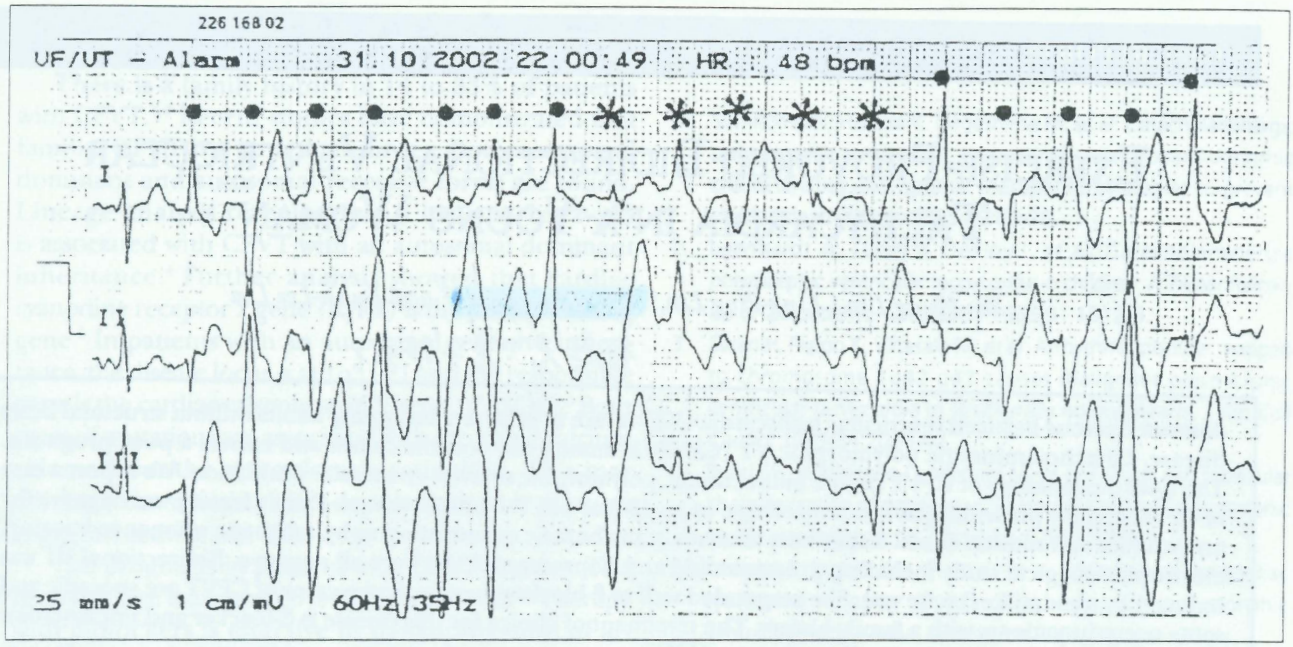

Fig. 1. Ambulatory electrocardiogram recorded by automatic external defibrillator showing alternating bidirectional (black dots) and polymorphic (star) ventricular tachycardia.

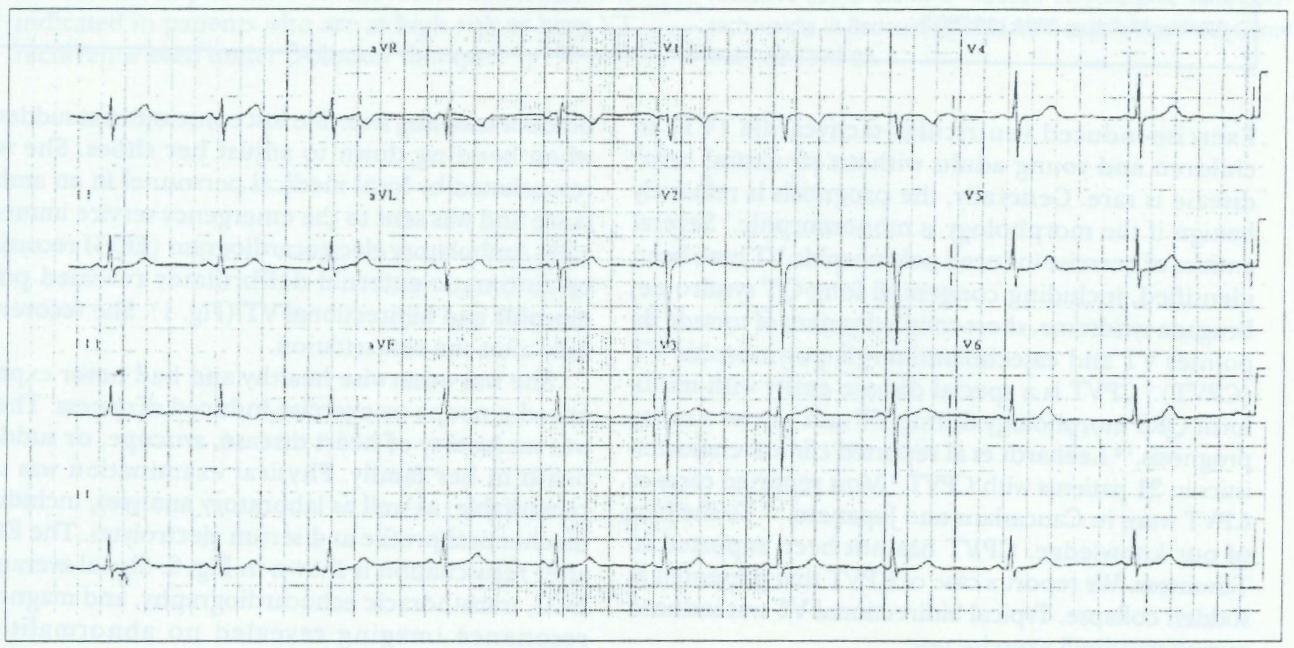

Fig. 2. Resting 12-lead electrocardiogram after successful resuscitation shows normal QRS morphology, no QT prolongation, nor abnormal ST-T change.

1 minute of exercise, the patient developed short runs of bidirectional VT (Fig. 3) and the exercise was terminated. VT regressed spontaneously 4 minutes later. She remained asymptomatic throughout the exercise test.

CPVT was diagnosed and the patient was placed on long-acting β-blocker (nadolol) therapy. Implantation of an automatic internal cardioverter defibrillator was suggested, but refused. She was free of symptoms at 6 months' follow-up. Her parents and younger sister also underwent a treadmill exercise test, but arrhythmia was not induced.

Discussion

The diagnosis of CPVT depends on 3 criteria: 1) the ability to induce more than 2 types of VT morphologies

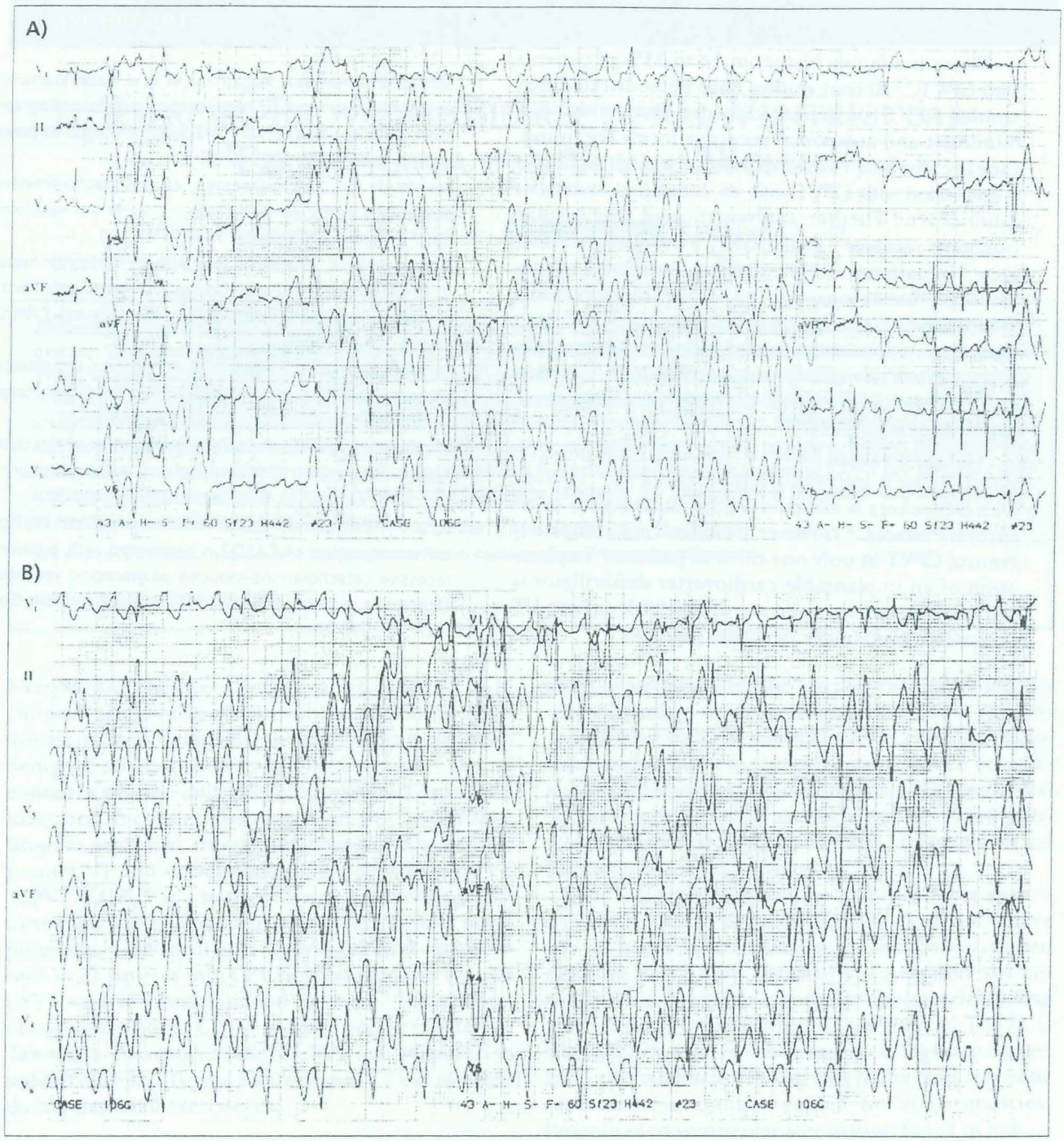

Fig. 3. Bidirectional ventricular tachycardia (VT) induced by treadmill exercise test: panel A, near the end of the exercise protocol; panel B, the beginning of the recovery phase. Electrocardiogram after 3 minutes of exercise shows bidirectional VT (A), which persisted for another 2 minutes after termination of exercise protocol (B, recovery phase). Panels A) and B) are continuous electrocardiogram strips.

for more than 3 consecutive beats by exercise or catecholamine infusion; 2) the absence of electrolyte imbalance, drug or organic heart disease; and 3) the absence of primary electrical disease such as long QT syndrome or Brugada syndrome.[1] The initial manifestations of CPVT include syncope (70-90%) and sudden death (10-30%).[1,2,4,5] The ECG morphology of CPVT is polymorphic, bidirectional VT, ventricular fibrillation, or their combinations.[1,3] CPVT is 100% inducible by exercise and 75% by isoproterenol infusion.

By contrast, programmed electrical stimulation can only induce CPVT occasionally, and the late potential of signal-averaged ECG is always negative. These

findings suggest that a catecholamine sensitive mechanism is responsible for CPVT.[1]

There is a family history in 14 to 33% of patients with CPVT.[1,2] Recent studies have demonstrated that familial CPVT is a genetic disorder. Both autosomal dominant and autosomal recessive forms are found. Lineage analysis revealed that a locus at 1q42-q43 is associated with CPVT with an autosomal dominant inheritance.[4] Further analysis showed that cardiac ryanodine receptor 2 gene (RyR2) is the disease-causing gene.[5] In patients with an autosomal recessive inheritance, the disease locus is at 1q31-21 and the responsible gene is the cardiac calsequestrin 2 gene (CASQ2).[6] Both types of mutations are associated with increased calcium release from the sarcoplasmic reticulum. This may enhance the delayed after-depolarization and cause triggered activity and VT.

The prognosis in patients with CPVT is grave. The mortality rate is around 30% by the age of 30.[4,5] Therapy with β-blockers is effective in the suppression of arrhythmic attacks.[1,2] However, β-blockers can completely control CPVT in only one-third of patients.[1] Implantation of an implantable cardioverter defibrillator is indicated in patients who are at high risk or have VT recurrence even under β-blocker therapy.

References

1. Sumitomo N, Harada K, Nagashima M, et al: Catecholaminergic polymorphic ventricular tachycardia: electrocardiographic characteristics and optimal therapeutic strategies to prevent sudden death. *Heart* 2003;89:66-70.
2. Leenhardt A, Lucet V, Denjoy I, et al: Catecholaminergic polymorphic ventricular tachycardia in children: A 7-year follow-up of 21 patients. *Circulation* 1995;91:1512-9.
3. Swan H, Piippo K, Viitasalo M, et al: Arrhythmic disorder mapped to chromosome 1q42-q43 causes malignant polymorphic ventricular tachycardia in structurally normal hearts. *J Am Coll Cardiol* 1999;34:2035-42.
4. Priori SG, Napolitano C, Memmi M, et al: Clinical and molecular characterization of patients with catecholaminergic polymorphic ventricular tachycardia. *Circulation* 2002;106:69-74.
5. Laitinen P, Brown KM, Piipo K, et al: Mutation of the cardiac ryanodine receptor (RyR2) gene in familial polymorphic ventricular tachycardia. *Circulation* 2001;103:485-90.
6. Lahat H, Pras E, Olender T, et al: A missense mutation in a highly conserved region of CASQ2 is associated with autosomal recessive catecholamine-induced polymorphic ventricular tachycardia in Bedouin families from Israel. *Am J Hum Genet* 2001;69:1378-84.