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IDIOPATHIC LONG Q–T SYNDROME*

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The association of Q-T interval prolongation, syncope and sudden death is known as the long Q-T syndrome. The syndrome may be familial, associated with congenital deafness, or idiopathic. The diagnosis is based on the electrocardiographic finding of a prolonged Q-T interval with or without T wave abnormalities. In this article, we present a nine-year-old boy admitted to the Hacettepe University Children's Hospital with complaints of syncopal episodes. The prolonged Q-T syndrome was diagnosed as the cause of the syncopal attacks. In addition, the same syndrome was detected in his father. The prolonged Q-T syndrome should be considered in the differential diagnosis of cases with recurrent, unexplained syncope in the pediatric age group. *Key words: Idiopathic long Q-T syndrome, syncopal attacks, family history.*

Idiopathic long Q-T syndrome is a hereditary condition with a fatal outcome[1,2]. The diagnosis of this syndrome is based on the electrocardiographic finding of a prolonged Q-T interval[1,3,4]. The treatment of choice for preventing syncopal attacks in this syndrome is either the administration of beta-adrenergic blocking agents or performing a left stellectomy[3,5,6]. In this article, we present a case who was admitted to the Hacettepe University Children's Hospital with ventricular tachycardia and syncope. The diagnosis of idiopathic long Q-T syndrome was made by electrophysiologic ambulatory Holter monitoring studies.

Case Report

A nine-year-old boy was admitted to the Hacettepe University Children's Hospital with complaints of syncopal attacks which were associated with urinary incontinence. However, there was no tonic-clonic movement or excessive salivation. About three years ago, he came to the Hacettepe University Children's

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Hospital with a history of fainting. He was then diagnosed as having epilepsy and treated with antiepileptic drugs but this therapy did not prevent the syncopal attacks. In addition, the attacks were induced by exercise and stress.
His family history revealed that his grandmother and grandfather were cousins. The son of the patient's uncle died at the age of fifteen following an epileptic seizure, and the patient's brother, died at age 12 at the Hacettepe University Children's Hospital, where he was under treatment for epilepsy.
His father suffered from palpitation and syncopal attacks induced by exercise but he did not take any drugs for these complaints (Fig. 1).

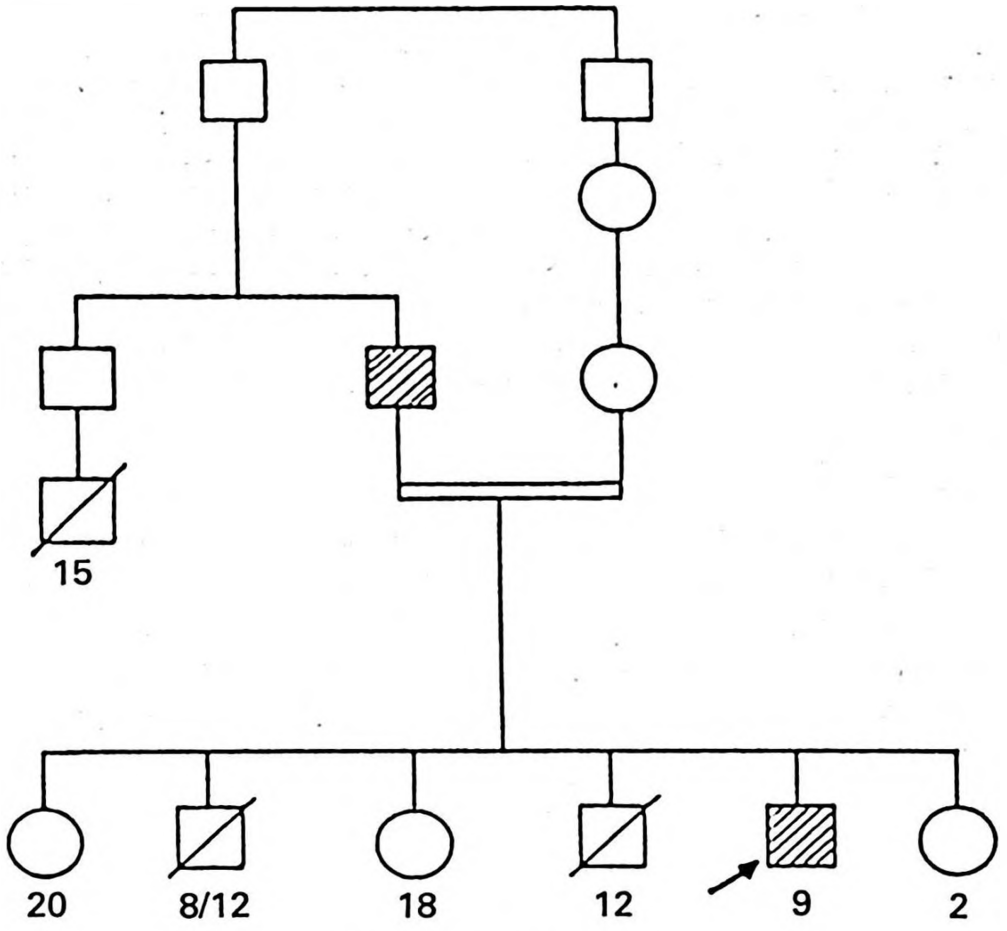

Fig 1: Family pedigree.

Physical examination at admission revealed that the boy's weight was 25 kg, height 133 cm, and arterial blood pressure 120/80 mm Hg. On auscultation the heart rate was regular at 80 per minute and there was no significant murmur. Laboratory studies revealed a hemoglobin of 13.65 g/dl, white blood cells 4200 per/mm^3, and plasma calcium 9.6 mg/dl. The electrocardiogram revealed that the sinus rhythm and the QRS mean vector was +15° in the frontal plane, PR 0.16 sec, and the corrected Q-T interval was 0.42 sec (Fig. 2). Two-dimensional echocardiographic examinations showed normal left ventricular function and

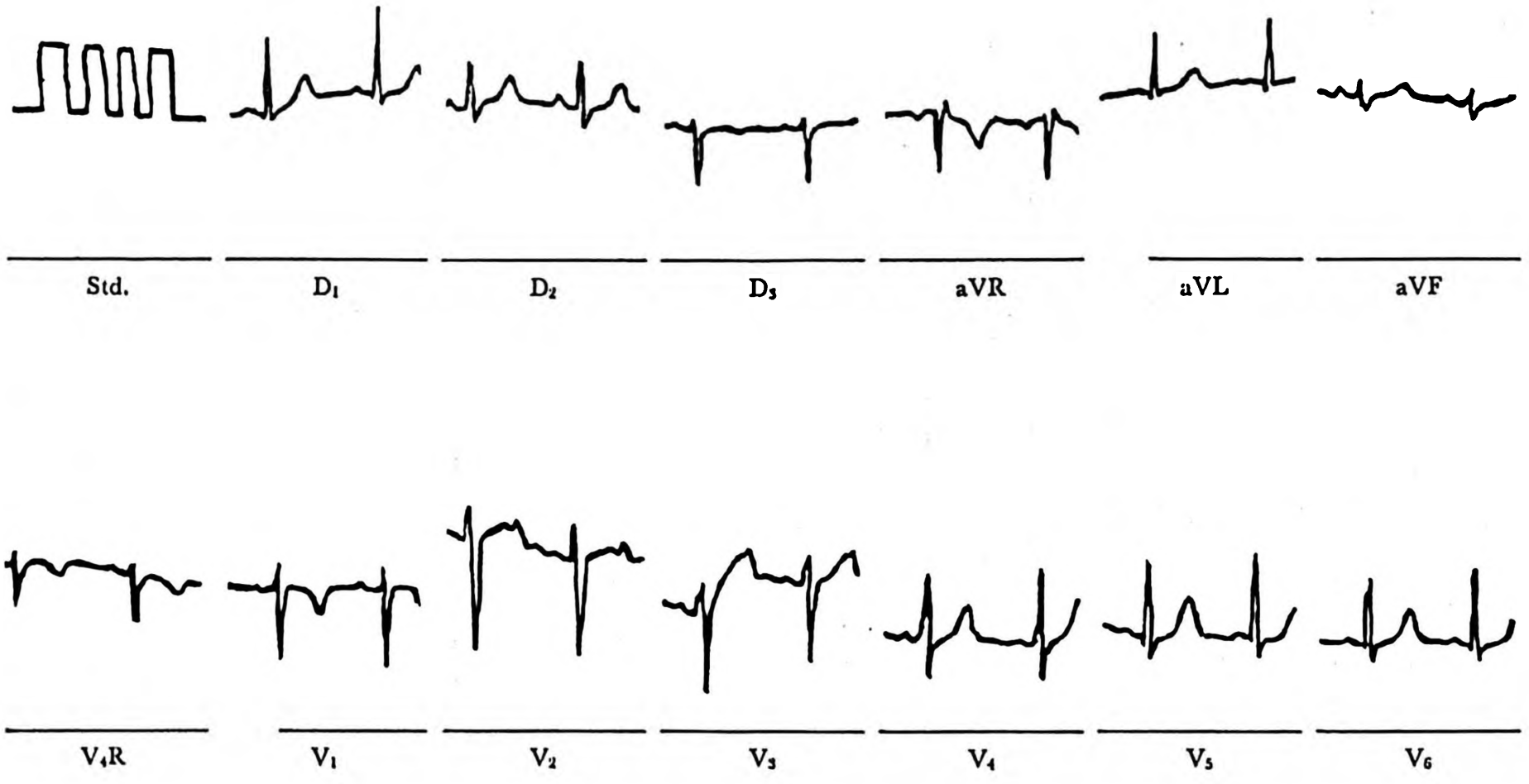

Fig 2: Electrocardiogram of the patient.

neither cardiomyopathy nor mitral valve prolapse was demonstrated. Pindolol (Visken^{R}) 2.5 mg/day was used to treat the tachycardia. After discontinuing the drug for one day electrophysiological studies were performed. Following local anesthesia, both femoral veins were catheterized and multipolar electrodes were placed in the high right atrium, low right atrium, His and right ventricular positions. The study was carried out according to a strict protocol, and Q-T interval 360 msec and corrected Q-T interval of 430 msec were measured respectively. The other parameters (RR, PR, QRS, QT, AH, HV, maximum sinus node recovery time, maximum corrected sinus recovery time, sinoatrial conduction time, Wenckebach point, effective atrial refractory period, effective atrioventricular node refractory period, effective ventricular refractory period) were normal, and despite several attempts, ventricular tachycardia could not be produced. A 24-hour ambulatory Holter-monitor study did not reveal any ventricular tachycardia (Fig. 3). Beta-adrenergic blockade as the choice of treatment in this case was very beneficial. After treatment with 2.5 mg pindolol, no syncopal attacks occurred.

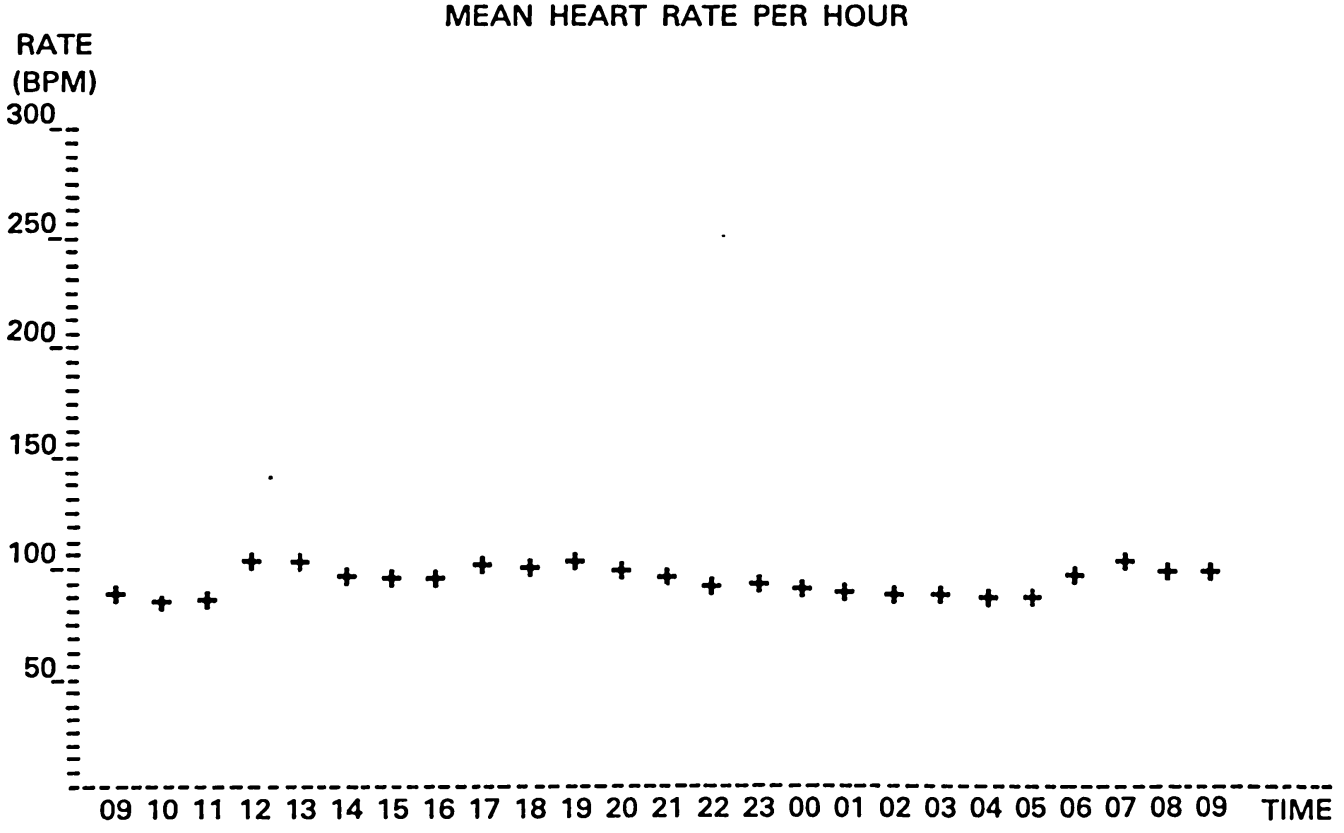

Fig 3: Mean heart rate per hour on the ambulatory Holter monitor.

The electrocardiograms of the patient's family members were evaluated for this syndrome, and a Q-T interval of 0.40 sec and a corrected Q-T interval of 0.43 sec were detected on the father's electrocardiogram (Fig. 4). Because of this finding, we ascertained that the father also had the idiopathic long Q-T syndrome.

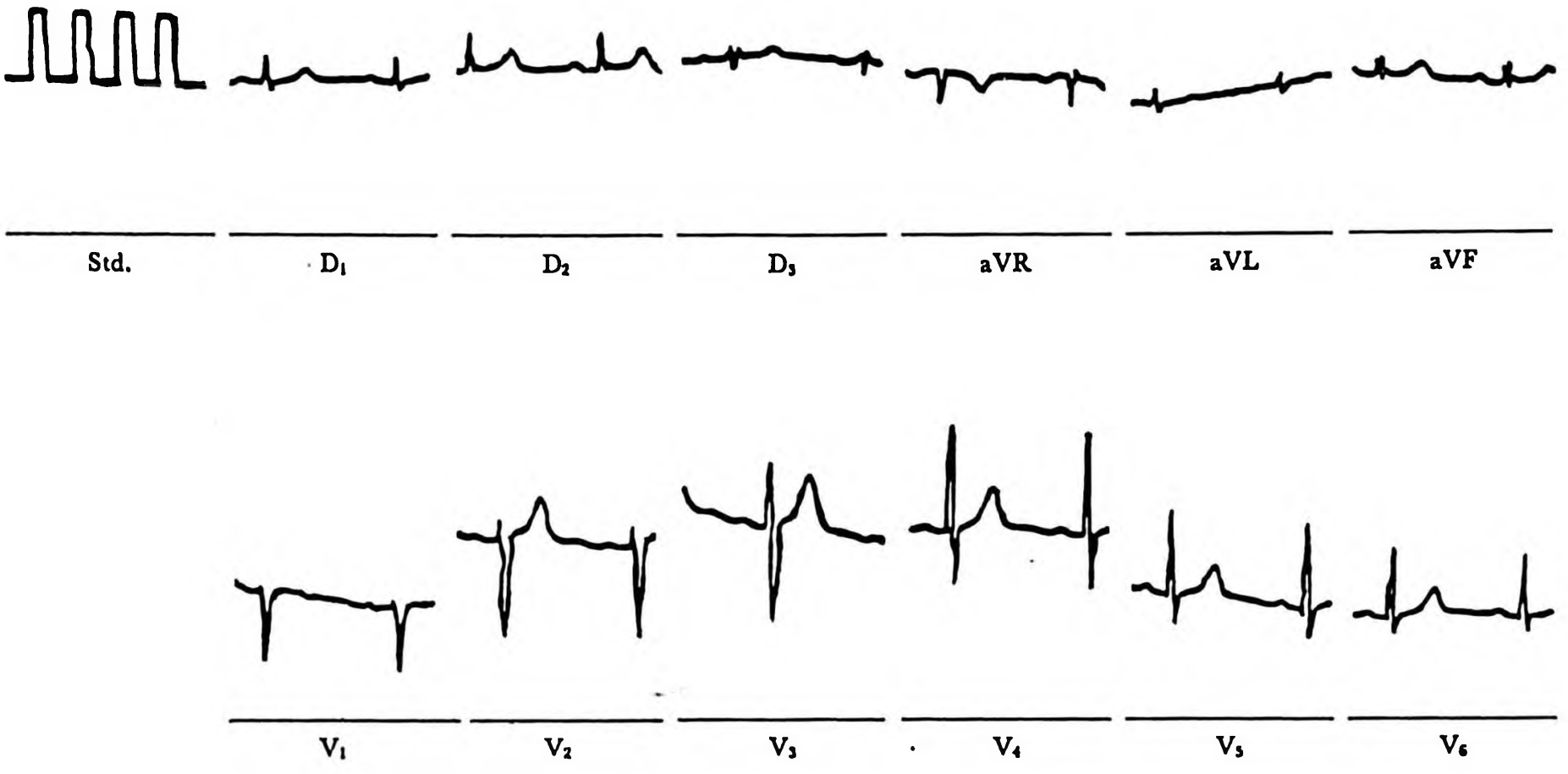

Fig 4: The father's electrocardiogram.

Discussion

Idiopathic long Q-T syndrome is a ventricular repolarization abnormality characterized by syncopal attacks and sudden death[1,2]. Some authors have investigated the characteristics of this syndrome[2,3], and Schwartz et al[1] have described the complete syndrome.

The Jervell and Lange-Nielsen syndrome is accompanied by congenital deafness and has an autosomal recessive pattern of inheritance. In the Romano-Ward syndrome, hearing is normal and an autosomal dominant pattern of inheritance has been confirmed[1,2]. A recent report suggests a probable close linkage between the HLA A 9 BW 54 and the Romano-Ward syndrome[4,7].

The Q-T interval varies with the heart rate. The corrected Q-T interval can be found by using Bazett's formula[4,7] given below:

$$QTc = \frac{QT}{\sqrt{R\text{-}R}}$$

The corrected Q-T interval is abnormal if it is prolonged beyond 0.40 sec in children under 12 years of age or is greater than 0.43 sec in children 12 years of age and over[4]. The affected individuals have ventricular tachycardia and syncope induced by exercise and stress[1,3,5].

Schwartz proposed some diagnostic criteria for this syndrome[1] (Table I). The diagnosis of idiopathic long Q-T syndrome should be made in the presence of two major criteria or one major and two minor criteria[1]. Bhandari et al[8] studied 15 patients with the idiopathic long Q-T syndrome with a thorough electrophysiologic protocol, and concluded that electrophysiologic studies were useless.

TABLE I. Diagnostic Criteria

Major	Minor
– Prolonged Q-T interval (QTC > 440 msec)	– Congenital deafness
– Stress-induced syncope	– Episodes of T wave alternans
– Family members with long Q-T syndrome	– Low heart rate (in children)
	– Abnormal ventricular repolarization

When a patient with the idiopathic long Q-T syndrome is identified, treatment with beta blockers should be instituted immediately. If the syncopal episodes continue despite full-dose beta-adrenergic blockade, a high thoracic left sympathectomy should be performed[1,3,5,6]. With adequate treatment, a reduction in the occurrence of syncope and sudden death has been achieved[3,5].

The clinical and electrocardiographic studies of both our patient and his father confirmed the diagnosis of the idiopathic long Q-T syndrome. In addition, we suggest that the patient's brother, who had suffered from syncopal attacks, may have died from the same syndrome.

Beta-blocker therapy was instituted in our case, and as a result, the syncopal attacks did not occur. We are planning a high thoracic left sympathectomy if tolerance develops.

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