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IMAGES IN MEDICINE

Does a Short QT Interval Presage a Short Life Span?

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ABSTRACT

A case of a 16-year old adolescent is presented who was referred for investigation of a syncopal episode. The ECG displayed a very short QT interval of 280 ms, compatible with the newly described short QT syndrome, which is a genetic primary electrical disease of abnormal potassium currents responsible for the repolarization process of the cardiac muscle. The prognosis is similar to the other well known familial cardiac channelopathies, such as the long QT or the Brugada syndromes, which are associated with a high risk of sudden cardiac death, requiring an implantable cardioverter defibrillator device for life-long protection.

KEY WORDS: *syncope; primary electrical disease, atrial fibrillation; sudden cardiac death; potassium channels; short QT syndrome*

A 16-year old adolescent was referred for investigation of a syncopal episode. The episode occurred while at an upright position, was preceded by dizziness and was non traumatic. There was no seizure activity reported, no tongue-bite or loss of bowel or bladder function. Family history was negative for sudden or unexplained deaths. Initial work-up was non-revealing. Physical examination was unremarkable. No orthostatic blood pressure changes were recorded. Carotid sinus massage was negative. However, the ECG displayed a very short QT interval of 280 ms (encircled, Fig. 1), compatible with the newly described short QT syndrome. There was also a relative absence of the ST segment with the J-point elevated in the lateral leads (early repolarization). Echocardiography disclosed no structural cardiac abnormalities. A stress and a tilting test to be followed by an electrophysiology study were recommended but the patient and his family refused further work-up or treatment. He remained well for the ensuing two years, but he was lost to further follow-up.

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The QT interval on an electrocardiogram encompasses the time required for the heart to depolarize and also complete its repolarization. It has been made abundantly clear over the years that a long QT interval mostly due to delayed and abnormal repolarization, a cardinal feature of the long QT syndromes, predisposes patients to life-threatening ventricular arrhythmia (torsades de pointes) and sudden cardiac death. The short QT syndrome is a newly recognized disease, first described in 2000,[1] characterized by a shortened QT interval and by episodes of syncope, paroxysmal atrial fibrillation or life-threatening arrhythmias. This autosomal dominant syndrome usually affects young and otherwise healthy individuals with no underlying structural heart disease and may be familial or present in sporadic cases. Over the last decade,

ABBREVIATIONS
ECG = electrocardiogram
ICD = implantable cardioverter defibrillator

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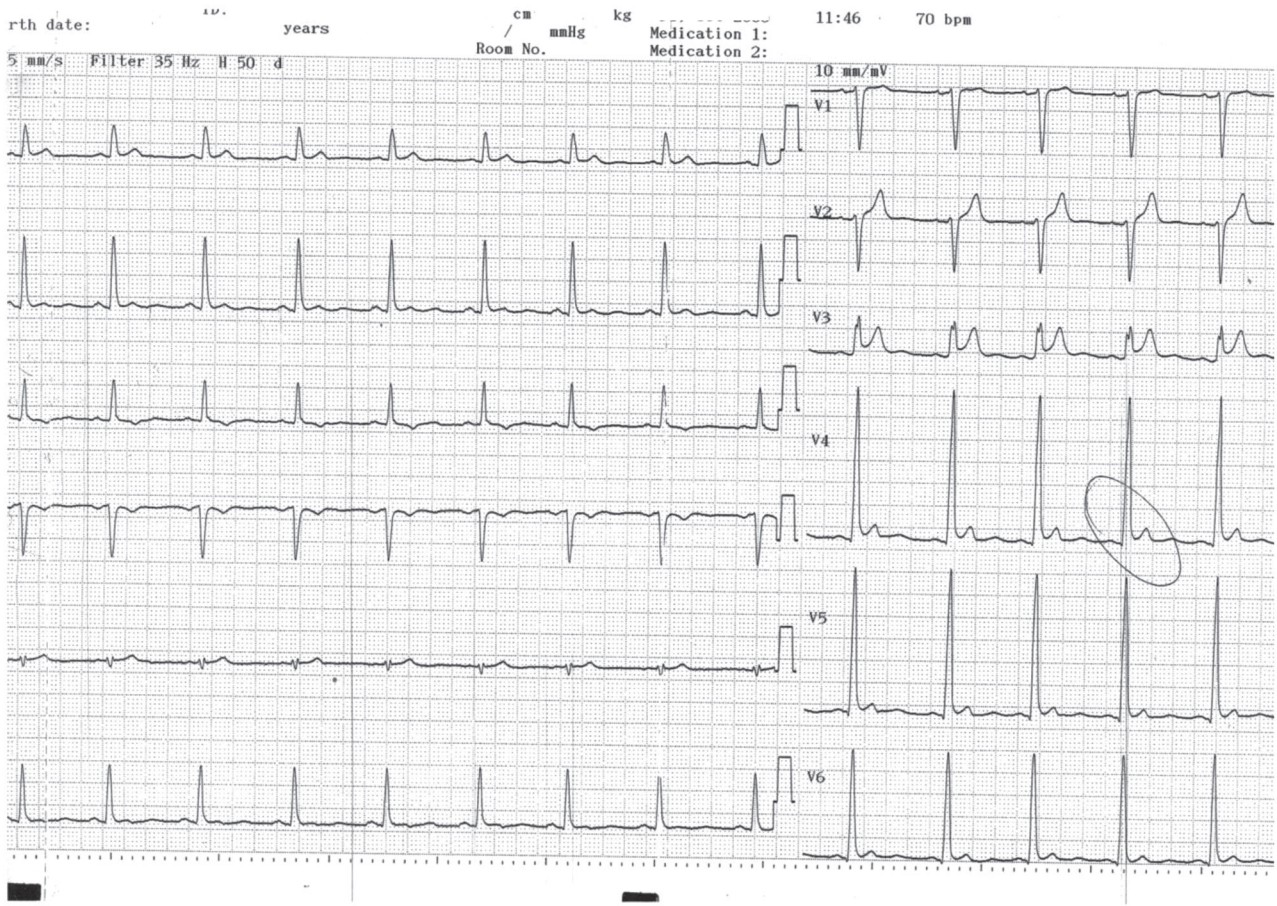

FIGURE 1. See text for discussion.

3 different genes leading to this disease have been described, and treatment has been proposed.[2,3]

The ion currents responsible for cardiac depolarization are mediated mainly by channels that allow the entry of sodium and calcium ions into the cell, while the repolarizing currents are mediated by channels that allow the exit of potassium ions. The short QT syndrome, as is the case with most primary electrical diseases (long QT syndrome, Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia) is caused by mutations in genes that encode for cardiac ion channels participating in repolarization.[2-4] To date 3 genes, KCNH2 (HERG), KCNQ1 and KCNJ2, encoding different repolarizing potassium channels have been detected in this syndrome. Although there may be benign variants of QT abbreviation, most patients with short QT syndrome have a family history of syncope, sudden death or atrial fibrillation. The clinical picture is highly variable; individuals afflicted by the syndrome may be asymptomatic or report episodes of syncope or atrial fibrillation or sustain episodes of ventricular tachyarrhythmias which may lead to sudden death. The ECG characteristically shows a very short QT interval ($\leq$320 ms), virtual absence of the ST segment (Fig. 1); and tall, peaked, narrow-based T waves. In some patients, as in our case, early repolarization has been concurrently noted (Fig. 1).[5-7]

For patients with aborted sudden death, or symptomatic individuals with syncope or those with a family history of sudden death, implantation of a cardioverter-defibrillator (ICD) device is the standard treatment. Antiarrhythmic agents that prolong the QT interval, such as sotalol or quinidine or disopyramide, have been suggested for the first form of the syndrome, however, no known potassium blocking agent can specifically target the other 2 forms.[3,4] This, of course, implies that there is a need to identify the form, by either genetic screening or by clinical means, so that the right therapy can be selected, which is usually not an easy task. Some investigators have proposed that early repolarization concomitant with short QT interval indicates a potential for sudden cardiac death.[5-7] In the largest series in the literature of long-term follow-up (over 5 years) of 53 patients with short QT syndrome, 24 patients received an ICD which was effective in preventing sudden death, while no arrhythmic events occurred in 12 patients receiving hydroquinidine.[8]

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