A Teenager With Syncope

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Vignette

An 18-year-old girl is brought by ambulance to the emergency department. Her mother, a nurse, says she found her unconscious and pulseless at home and immediately began cardio-pulmonary resuscitation. There is no history of prior illness, drug ingestion, or cardiac disease, and no family history of syncope or sudden death.

The patient's cardiovascular exam is normal. Routine hematology and biochemistry lab values are normal, with the exception of mild hypokalemia (3.5 mmol/L). An electrocardiogram (ECG) is obtained (Figure 1).

Questions

1. **Does the ECG provide any clue to the diagnosis?**
2. **What condition might the patient have?**

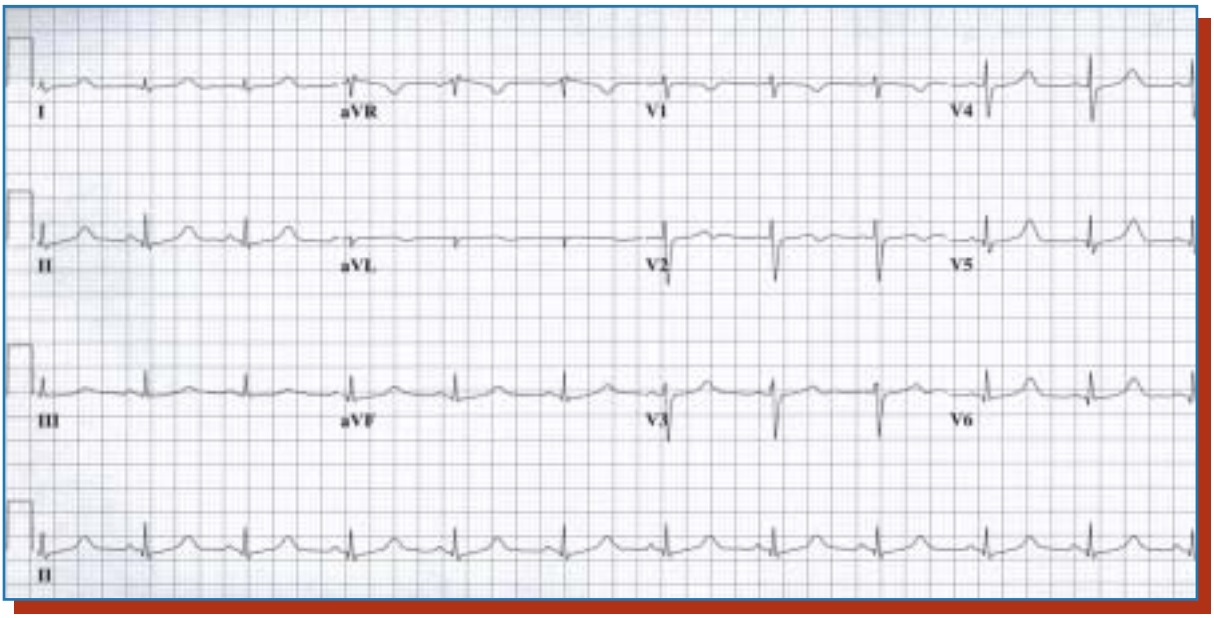

Figure 1. ECG upon presentation.

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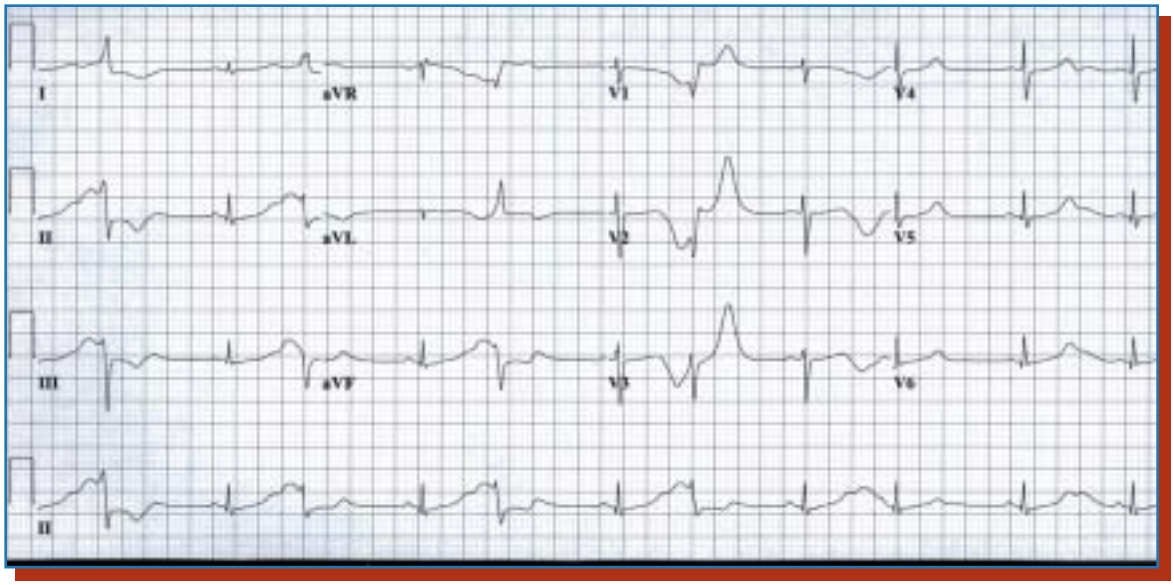

Figure 2. A repeat ECG one hour after presentation.

Answers

1. The ECG obtained shortly after presentation shows sinus rhythm at a rate of 70 beats per minute. The QRS configuration is normal; there is no evidence of ischemia or injury and no conduction disturbances are apparent. At first glance, the recording appears to be normal. However, upon closer scrutiny, the QT interval appears marginally prolonged. It actually measures 0.47 seconds and when corrected for heart rate (QTc = measured QT interval divided by the square root of the R-R interval), the QTc interval is 0.5 seconds. The upper limit of normal for the QTc interval in a woman is 0.46 seconds.

2. The presence of a somewhat prolonged QTc interval in this clinical setting raises concern about a congenital long QT syndrome (LQTS). At least six different forms of the syndrome have been identified.

The syndrome is caused by mutations in cardiac ion channel genes. These mutations generally result in a decreased potassium outward current or a persistent inward sodium current. In either case, the overload of myocardial cells with positively charged ions results in abnormally prolonged repolarization. Drugs which prolong the QT interval and electrolyte abnormalities (especially hypokalemia) may aggravate the situation. The occurrence of syncope and sudden cardiac death in LQTS patients is due to polymorphous ventricular tachycardia of the torsade de pointes variety.

One hour after being placed on cardiac telemetry, ventricular ectopic activity was noted. A repeat ECG (Figure 2) showed dramatic QT prolongation with a measured QT interval of approximately 0.75 seconds. Note also the presence of transient T wave alternans in the last three or four beats on the rhythm strip; this finding is considered an indicator of increased arrhythmia risk.

The presumptive diagnosis in this case was syncope due to torsade de pointes in a patient with LQTS. The patient was referred for electrophysiologic and genetic assessment and subsequently received an implantable cardiac defibrillator. Dx