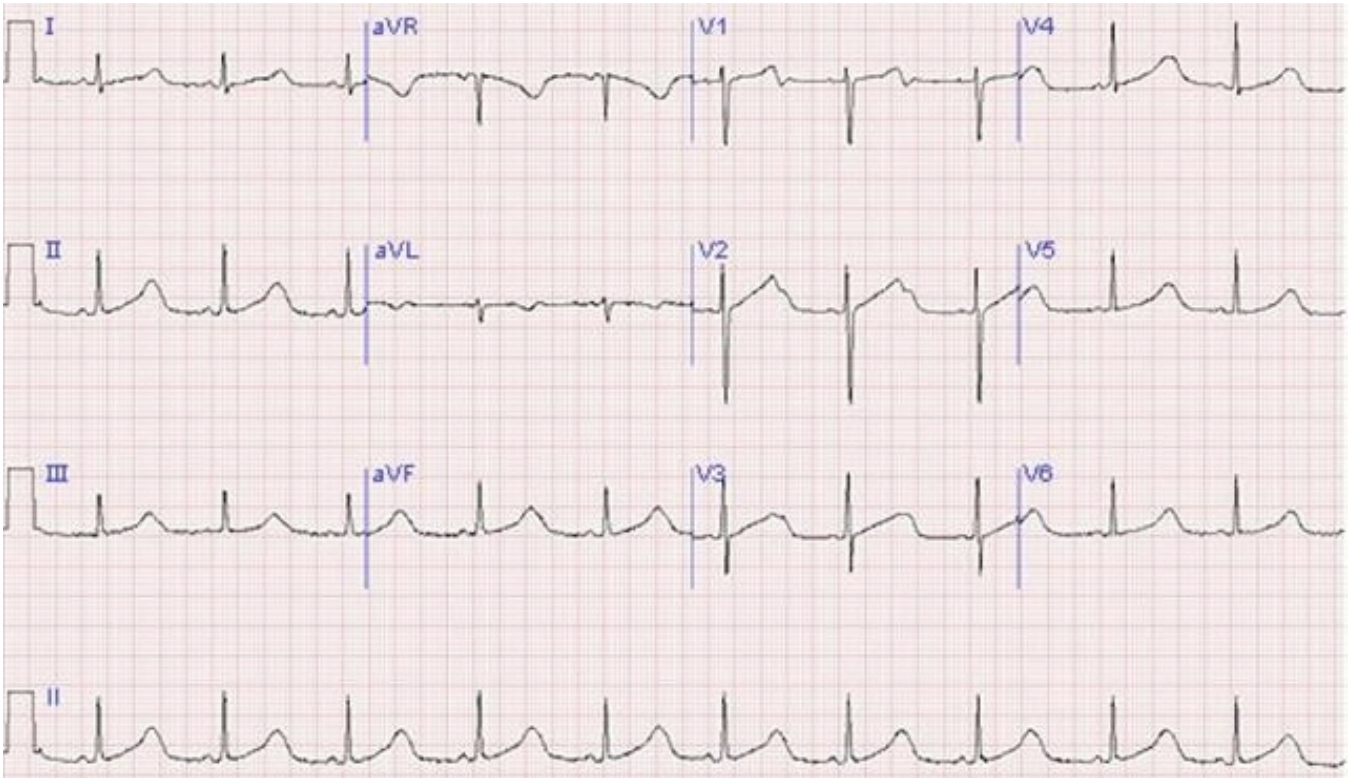
ED QUICK QUIZ

WHAT IS THE DIAGNOSIS?

BACKGROUND

20 year old female presents to the ED after a faint while using a home exercise bike yesterday. The faint was unwitnessed and without preceeding chest pain, shortness of breath, palpitations, dizziness, tongue biting or incontinence. She remembers awakening on the floor and briefly feeling dazed. She felt well afterwards but was worried this morning, and wants “to get checked out just to be safe”. She recalls a similar syncopal episode a few years ago again when exerting herself. She has no recent history of illness or fever and does not report any subsequent chest pains, shortness of breath or palpitations. She also denies recent dieting or use of any over-the-counter or illicit drugs.

Examination shows normal observations, HR 65 bpm and BP 110/73 mm Hg and is otherwise completely unremarkable. Her investigations including pregnancy test are normal. An ECG is obtained.



QUESTIONS

1. What is the diagnosis?
2. Any other questions in the history you would ask?

ANSWER & DISCUSSION

1. **Diagnosis**

Long QT syndrome (LQTS): The ECG demonstrates prolongation of the QT segment as demonstrated by a QT interval of 0.6 seconds. A QT interval greater than 0.44 seconds is generally considered to be prolonged, but as the QT interval is affected by the heart rate, and a corrected value referred to as the QT_c is calculated with the following formula: $QT_c = QT/\sqrt{RR}$ interval. In this case, for a heart rate of 65 bpm in a woman, the maximal normal QT interval is 0.42 seconds. Because the QT_c of 0.61 s is greater than the maximal allowed QT interval, the interval is prolonged.

The diagnosis of LQTS has been increasingly recognized as a cause of unexplained dizziness, syncope, and sudden cardiac death in otherwise healthy young individuals. The diagnosis should be considered in any patient with a history similar to this patient's.

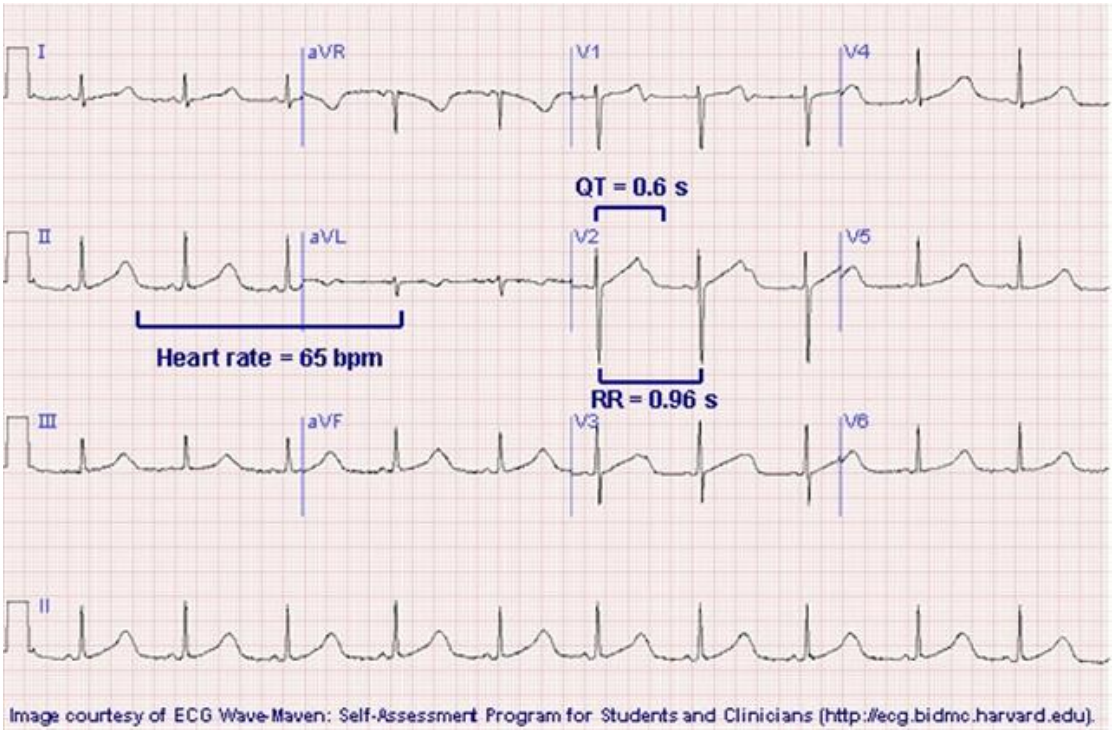


Image courtesy of ECG Wave-Maven: Self-Assessment Program for Students and Clinicians (http://ecg.bidmc.harvard.edu).

2. Other Questions

Have any family members ever had syncopal episodes or sudden death? In this case her brother had suffered from syncopal episodes. Such a presentation of syncope with a similar history in an immediate family member is suggestive of **congenital** LQTS.

Congenital LQTS is now considered to be an inheritable abnormality in cardiac sodium and potassium channels. This "channelopathy" predisposes patients to episodes of torsades de pointes, especially when they are exposed to increased catecholamine levels (adrenergic dependent or tachycardia

dependent). A related important point to assess in patients with a familial history of unexplained syncope or sudden death is an associated history of hearing loss. Some forms of LQTS (eg, Jervell and Lange-Nielsen syndrome) are accompanied by congenital neuronal deafness. Other forms (eg, Romano-Ward syndrome) do not have an associated hearing loss.

The danger of a prolonged QT segment is the potential for degeneration to a specific type of polymorphic ventricular tachycardia known as torsades de points, characterized by a ventricular rate greater than 200 bpm in which the QRS structure has an undulating axis that shifts polarity about the baseline. This rhythm can spontaneously convert to a sinus rhythm or degenerate into ventricular fibrillation. The rhythm itself or a brief degeneration into ventricular fibrillation can account for a clinical presentation of syncope in patients with LQTS.

Acquired forms of LQTS are not uncommonly encountered in the ED. Acquired QT prolongation is usually precipitated by a slow heart rate and therefore called pause dependent in contrast to adrenergic dependent. Acquired forms are often the result of drug therapy with a variety of antiarrhythmic medications, phenothiazines (eg, haloperidol), cyclic depressants, high dose methadone, antihistamines, and some antimicrobials. Resultant torsade de pointes is usually observed within 2 weeks of the start of the QT-altering medication, however delayed presentations can also occur if other medications that affect the QT interval are added to the patient's regimen. Other causes of pause-dependent prolongation of the QT interval are electrolyte disturbances (↓K, Mg, Ca), myocardial ischemia, hypothyroidism, use of drugs (eg, cocaine, amphetamines), and haemorrhagic CVA or SAH.

Patients with congenital LQTS require long-term treatment. The cornerstone of therapy is life-long adrenergic blockade with beta-blockers.

- No participation in competitive sports for patients with the diagnosis established by means of genetic testing only
- Beta-blockers should be given to patients who have QTc-interval prolongation (>460 ms in women and >440 ms in men) and are recommended (class IIa) for patients with a normal QTc interval
- An implantable cardioverter-defibrillator (ICD) should be used in survivors of cardiac arrest and is recommended (class IIa) for patients with syncope while receiving beta-blockers; ICD therapy can be considered (class IIb) for primary prevention in patients with characteristics that suggest high risk (including LQT2, LQT3, and QTc interval >500 ms).

Acquired LQTS, withdrawal of the offending agent and/or electrolyte repletion is often all that is necessary to prevent recurrences in most patients. The exception is in patients with sick sinus syndrome or AV blocks in whom a pause or bradycardia precipitates torsades. These patients require permanent pacemakers.