

T-wave alternans in long QT syndrome

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ABSTRACT

Long QT syndrome (LQTS) is a congenital disorder characterized by prolongation of QT interval in the electrocardiogram (ECG) and a propensity to develop ventricular arrhythmias, which may lead to syncope, cardiac arrest or sudden death. T-wave alternans (TWA), a phenomenon of beat-to-beat variability in the repolarization phase of the ventricles, has been closely associated with an increased risk of ventricular tachyarrhythmic events (VTE) and sudden cardiac death (SCD).

Keywords: Long QT syndrome, sudden cardiac death, t-wave alternans

CLINICAL SUMMARY

This is a case of long QT syndrome (LQTS) with macroscopic T wave alternans (TWA) in the ECG.

A 5-year-old boy, second born of nonconsanguineous parents, was admitted with history of repeated episodes of loss of consciousness, each episode lasting for few minutes, in the past 6 months. There were at least three episodes of loss of consciousness every month, each lasting for few minutes after the initial episode. There was history of sudden cardiac death in the older sibling.

There was no history of head trauma or drug ingestion. He did not have syncope or seizures in the past. At the time of admission, he was drowsy, with no other positive physical findings. Seizure disorder was considered as the first possibility in the pediatric emergency room. The blood sugar, serum sodium, potassium, calcium and magnesium were normal. Electroencephalogram, echocardiogram and a CT scan of the brain were normal. Electrocardiogram on the day of hospitalization revealed marked prolongation of corrected QT interval [Figure 1] along with macroscopic T wave alternans [Figure 2].

Screening ECGs of the parents were normal. Based on the symptoms and ECG findings a diagnosis of LQTS was made

and the child was started on oral propranolol which was gradually escalated to 3mg/kg/dose every 6 hours over 7 days. He had further episodes of loss of consciousness despite maximal betablocker therapy. He therefore underwent left thoracoscopic cervical sympathectomy.^[1] Left sided Horner's syndrome is confirmed. The patient is also scheduled for an implantable cardioverter defibrillator device as soon as the finances are arranged.

DISCUSSION

Long QT syndrome (LQTS) is a congenital disorder characterized by prolongation of QT interval in the electrocardiogram (ECG) and a propensity to develop ventricular arrhythmias, which may lead to syncope, cardiac arrest or sudden death. TWA may be visible to the naked eye (macroscopic TWA) or only detected by special equipment that detect minute changes in the T wave voltages not visible to the naked eye (micro volt TWA). Macroscopic TWA may precede dangerous arrhythmias like polymorphic ventricular tachycardia (torsade de pointes) and ventricular fibrillation.^[2-6] Very few cases of macroscopic TWA in LQTS have been reported from India.^[4]

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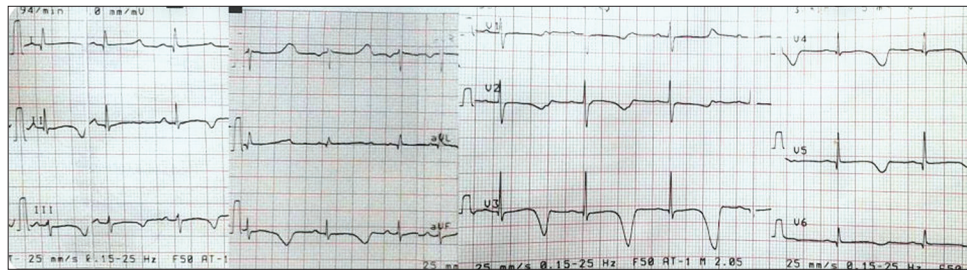


Figure 1: Electrocardiogram showing markedly prolonged corrected QT interval

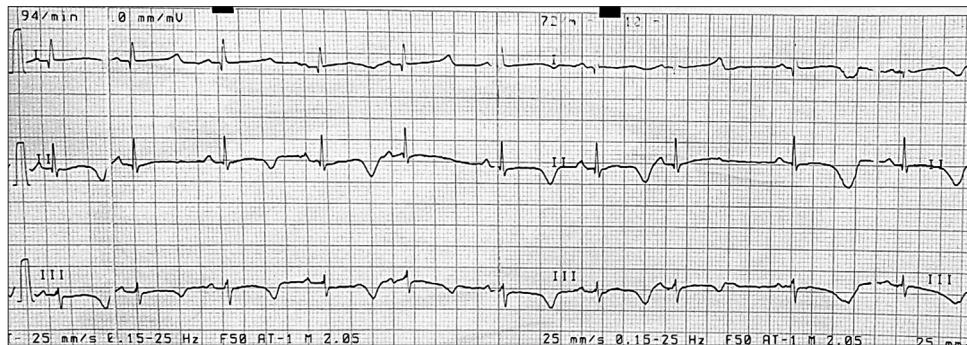


Figure 2: Electrocardiogram showing markedly prolonged corrected QT interval with macroscopic T Wave alternans

Conditions associated with T-wave alternans

In normal hearts T wave alternans is a physiological rate related phenomenon. It is considered to be pathological when it occurs at heart rates <110bpm.^[7,8] TWA is seen in many disease states like congestive heart failure, acute ischemia, drug effects, Brugada syndrome, and hypertrophic cardiomyopathy. Though the mechanism of TWA may be different in these various diseases, generally 2 major hypotheses exist to explain the mechanism of TWA.

Mechanism of T-wave alternans

The Action Potential Duration Restitution Hypothesis states that when heart rate exceeds the ability of a repolarization channel to fully recover from time-dependent inactivation, the ion channel is partially open upon initiation of the next beat. This shortens the APD of the next beat, and its diastolic recovery interval is greater. Subsequently, the channel is more recovered and thus is less open upon initiation (less net repolarization current), resulting in longer APD and shorter diastolic interval, perpetuating action potential alternans.^[8]

The Calcium-Cycling (Calcium Restitution) Hypothesis states that cellular alternans occurs when heart rate exceeds the capacity of the myocyte’s intracellular machinery to cycle calcium on an every-beat basis. Consequently, sarcoplasmic reticular and cytoplasmic calcium flux alternates secondary to alterations in intracellular calcium.^[8]

T-wave alternans and arrhythmogenesis

T wave alternans can be concordant or discordant, and this difference is important as it forms the basis of arrhythmogenesis. When action potential alternans is first initiated above a particular threshold heart rate, there is no phase difference between the various regions /cells in the myocardium. This is known as spatially concordant action potential and is not arrhythmogenic.^[8] With further acceleration of heart rate or a premature impulse, spatial discordance sets in. Here, myocytes in neighboring regions of myocardium exhibit action potentials that alternate with opposite phase. Discordant alternans has a significant impact on the spatial organization of repolarization across the ventricle and creates significant gradients of repolarization that are sufficient to cause conduction block, promoting reentrant excitation.^[9] It serves as a mechanism for dynamically amplifying electrophysiologic heterogeneities in the heart to produce conditions for re-entrant, and is always a pre-requisite for initiating re-entry.^[2]

T-wave alternans in long QT syndrome

Long-QT syndrome is different unusual in that T-wave alternans and arrhythmias are provoked at slow rather than increased heart rates. The role of calcium handling as the primary mechanism of TWA is also unclear though it is considered that it might contribute. The autonomic nervous system seems to play an important modulatory role in cardiac alternans in long-QT syndrome.^[2,5]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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