Dual ICD therapy on a long QT syndrome patient along with pseudo block A-V 2:1

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SUMMARY

A 10-year-old girl was admitted to the hospital after experiencing multiple episodes of loss of consciousness due to ventricular tachycardia on the background of congenital long QT syndrome. The patient's ECG reveals two distinct and unusual features: T wave alternation and 2:1 atrio-ventricular block. After admission, she continued to appear unconscious with monitoring images of ventricular tachycardia and torsades de pointes. The patient was resuscitated and a temporary pacemaker was implanted. Finally, the patient was appointed to implant a dual ICD device. However, when analyzing the device's tracking, it was found that:

Her native rhythm was sinus rhythm with a long QT of 680 ms when the sinus rhythm cycle longer than 1400 ms, its can lead to atrial-ventricular conduction of 2:1. Finally, the VVI anti-bradycardia mode was set to decrease ventricular pacing, which causes biventricular synchronization primitives, as well as the phenomenon of ventricular pacing on the T wave, which causes polymorphic VT. This leads to optimal ICD in minimizing ATP and electrical shock.

BACKGROUND

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. The extreme prolongation of ventricular action potential duration that occurs in some of the long QT syndromes may result in two forms of alternating activity of the heart included of T wave alternation and a "pseudo" 2:1 atrio-ventricular block, both of which are rate dependent. 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). Futhermore, a "pseudo" 2:1 atrio-ventricalar (AV) block can hide a condition of asymptomatic sinus node dysfunction (sick sinus syndrome) or high degree AV block. Electrophysiology studies is essential before deciding to implant an ICD, which is extremely important for optimal patient outcomes, epesically patient have AV block in their surface cardiogram.

Keywords: Long QT syndrome, sudden cardiac death, T-wave alternans, a "pseudo" 2:1 atrio-ventricular (AV) block, ICD.

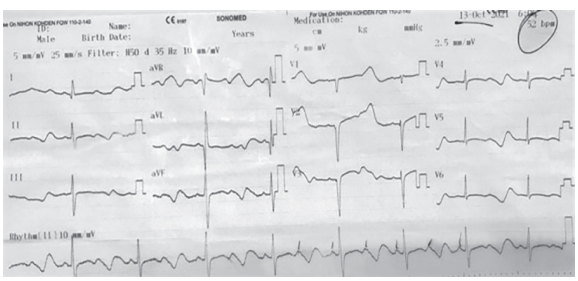
CASE REPRESENTATION

A 10-year-old girl had been transported

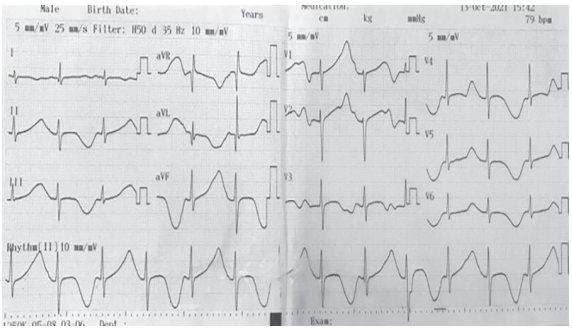
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to the emergency room after suffering head trauma. She has a history of 3-4 occurrences of loss of consciousness in the previous 2 days, each lasting one minute. She had a headache and feels dizzy before each incident. She had no seizures or agina throughout each incident. She awoke after this event. **Medical history:** a diagnosis of bradycardia six years previously. She had no history of drug ingestion in recent time. **Family history:** There were no episodes of unexplained sudden death in her family. Her vital signs at time of administration were in normal range: (1) A temperature of 36.8 degrees Celsius, (2) A heart rate of 52 cycles per minute, (3) A respiratory rate of 20 breaths per minute with her oxygen saturation of 98% (at rest in room air), and (4) Blood pressure of 120/80 mmHg. Her examination had revealed no positive findings. She had a further episode of loss of consciousness after 30 minutes of hospitalization. Her monitoring had showed she has complete AV block, torsades de pointes, and non-sustained VT. The patient was resuscitated and a temporary pacemaker was implanted. Finally, the patient was appointed to implant a dual ICD.

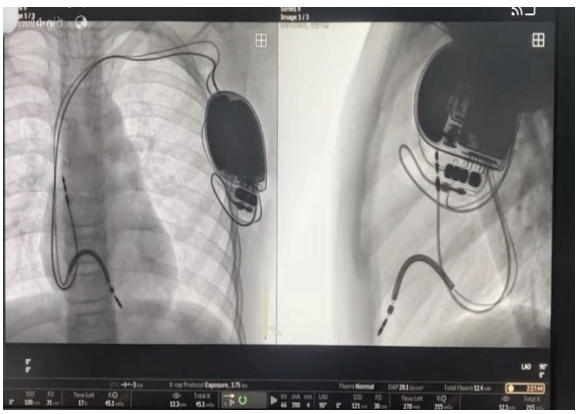
INVESTIGATIONS



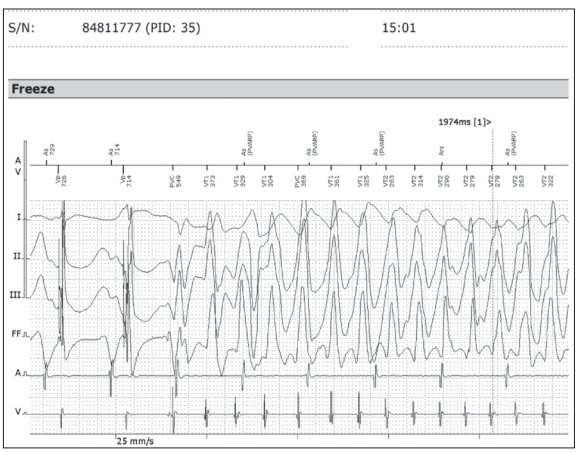
Picture 01. ECG when the patient is admitted to the hospital: Sinus rhythm, QTc = 720 ms and conduction of 2 atria followed by 1 ventricle activation.



Picture 02. ECG 5 hours after admission: Sinus rhythm, QTc = 680 ms and T wave alternation.



Picture 03. X-ray image of a dual ICD after implantation



Picture 04. Ventricular pacing induced VT shortly after connection to the atrial and ventricular electrodes of ICD.

S/N:	84811777 (PID: 35)	15:35
Parameters - Bradycardia		
Mode		VVI
Basic rate		
Basic rate [bpm]		40
Rate hysteresis [bpm]		OFF
Scan/Repetitive		
Night rate [bpm]		OFF
Night begins		
Night ends		

Picture 05. VVI - The last ICD setting pacing mode is selected to suit the patient

Other investgations:

TTE: LVEF = 35% and LV diameter (Dd) = 55 mm.

MSCT cerebral vascular: No sign of occlusion, no cerebrovascular malformation.

Her blood sugar level, serum sodium, potassium, calcium, and magnesium were in a normal range.

DIAGNOSIS

Based on the symptoms and ECG findings a diagnosis of LQTS and high-grade AV block was made.

TREATMENT

Her ESP in previous 6 years show that she had non-persistent AV block with HV interval of 74 ms. She had been indicated to temporary cardiac pacing. Because Beta-blocker therapy was contraindicated, so she had been scheduled for a dual implantable cardioverter defibrillator (ICD).

DISSCUSION

Congenital long QT syndrome (LQTS) is characterized by heart rate corrected QT interval prolongation and potentially fatal arrhythmias, resulting in syncope and sudden death. (1) QTc in this patient was determined as 680 ms using the Bazett formula for measuring QT duration. It meets the criteria for long-QT syndrome (QTc > 460 ms in women). According to Schwartz criteria, (2) this patient has: QT duration (as determined by the Bazett's formula) = 680 ms >= 480 ms (1 point) this patient also had confirmed that no drug consumption. She had a normal blood test with K, Mg in serum. Torsades de Ponites (2 points) Low resting heart rate (1 points) and Syncope (0.5 points). This patient's total score = 4.5 points > 3.5, where this score suggests a strong probability of LQTS. According to the Hobbs et al, (3) in LQTS, the timing and frequency of syncope, QTc prolongation, and sex were predictive of risk for aborted cardiac arrest and sudden cardiac death during adolescence. Additional approaches for more accurately assessing LQTS on the ECG have been developed in recent years, including the use of artificial intelligence in establishing the diagnosis. (4)

T-wave alternans is an uncommonly observed electrocardiographic (ECG) finding of beat-to-beat alternation in T-wave shape or amplitude. "Macroscopic" T-wave alternans, visible to the naked eye, is associated with a prolonged QT interval and long QT syndrome (LQTS). T-wave alternans may portend a rapidly developing and life-threatening scenario, and its recognition is crucial to prevent progression to lethal

arrhythmias. (5) TWA can be found in a variety of diseases, including congestive heart failure, acute ischemia, medication side effects, Brugada syndrome, and hypertrophic cardiomyopathy. Though the mechanism of TWA may vary in these different disorders, there are typically two basic hypotheses to describe the causes of TWA: the action potetial duration restitution and the calcium-cycling (Calcium Restitution). Long-QT syndrome is distinct in that T-wave alternans and arrhythmias are triggered at low rather than high heart rates. The involvement of calcium handling as the major mechanism of TWA is similarly unknown, while it is thought to have a role. (6), (7) This patient was one of a few cases of macroscopic TWA in LQTS documented in Vietnam. T-wave alternans were difficult to detect in her administration's ECG (Picture 01). However, the second ECG (Picture 02) after 5 hours of hospitalization showed negative T-waves alternating with positive T-waves. The occurrence of T-wave alternans in electrocardiographic signals was recently linked to susceptibility to ventricular arrhythmias and sudden cardiac death. (8)

According to the main treatment principle: Beta blockers are the mainstay of therapy for the prevention of cardiac events and implantable defibrillators for secondary prevention. (9) The patient in this case with an indication for atrial pacing included a history of symptomatic sinus high degree AV block, sinus arrest with syncope and a history of Torsades de Ponites. Dual-chamber ICDs are indicated for this patient. Dual-chamber ICD was considered to be an effective therapy for this condition. When the device's tracking was examined, it was discovered that her native rhythm was sinus rhythm with a long QT of 680 ms. When the sinus rhythm cycle is longer than 1400 ms, it can lead to atrial-ventricular conduction of 2:1. Finally, the VVI anti-bradycardia mode was adjusted to reduce ventricular pacing, which creates biventricular synchronization primitives, as well as ventricular pacing on the T wave, which generates polymorphic VT. This results in ideal ICD in terms of ATP and electrical shock.

LEARNING POINTS

1. In summary, congenital LQTS is an inheritable entity characterised by a prolonged heart-rate corrected QT interval, and it associates with malignant arrhythmias at young age.

2. The extreme prolongation of ventricular action potential duration that occurs in some of the long QT syndromes may result in two forms of alternating activity of the heart: a T wave alternation and a "pseudo" 2:1 atrio-ventricular (AV) block.

3. Electrophysiology studies are required before determining to implant an ICD, which is critical for optimal clinical outcomes if indeed the patient demonstrates AV block on their surface electrocardiogram.

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 her consent for her 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.

FINANCIAL SUPPORT AND SPONSORSHIP

Nil.

CONFLICTS OF INTEREST

There are no conflicts of interest.

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