A tale of Syncope, Prolonged QT and an ICD

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BACKGROUND

- Long QT syndrome (LQTS) was first described in the 1960s.
- It is clinically manifested as syncope, cardiac arrest or sudden cardiac death.
- LQTS can be caused by 15 different genes.
- These gene mutations lead to action potential prolongation by causing impaired repolarizing currents.

CASE DESCRIPTION

- A 29-year-old previously healthy Caucasian woman was admitted after recurrent episodes of syncope that happened within 1-month prior to presentation.
- She was hemodynamically stable with normal vitals.
- She had an episode of sustained monomorphic ventricular tachycardia (VT) and underwent cardioversion. (Figure 1 & 2)
- She was put on amiodarone infusion.
- Serial ECGs showed prolonged QTc.
- She had another episode of pulseless VT that terminated without defibrillation.
- She was transferred to our facility for possible ablation and/or implantable cardioverter defibrillator (ICD) placement.
- Her family history was significant for paternal aunt who had died unexpectedly at the age of 39.
- All her lab work including electrolytes, thyroid panel, cardiac enzymes, inflammatory markers and extended drug screen was unrevealing.
- Transthoracic echocardiogram showed normal biventricular size and function.

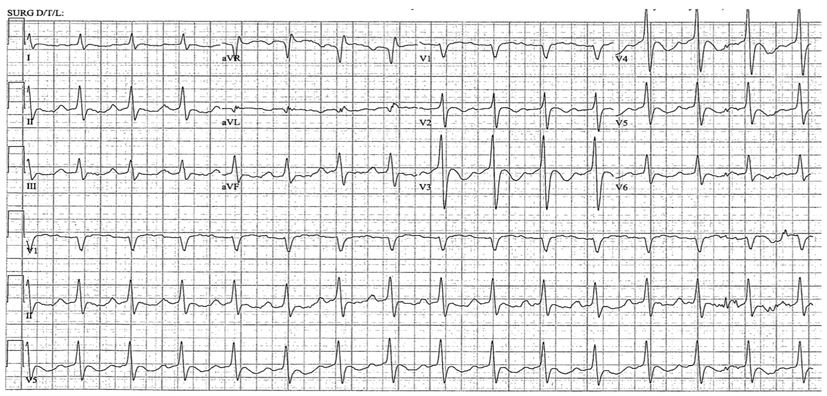


Figure 1: Normal Sinus rhythm, Idioventricualr conduction delay, PR 176msec, QRS 166msec, QTc 598msec

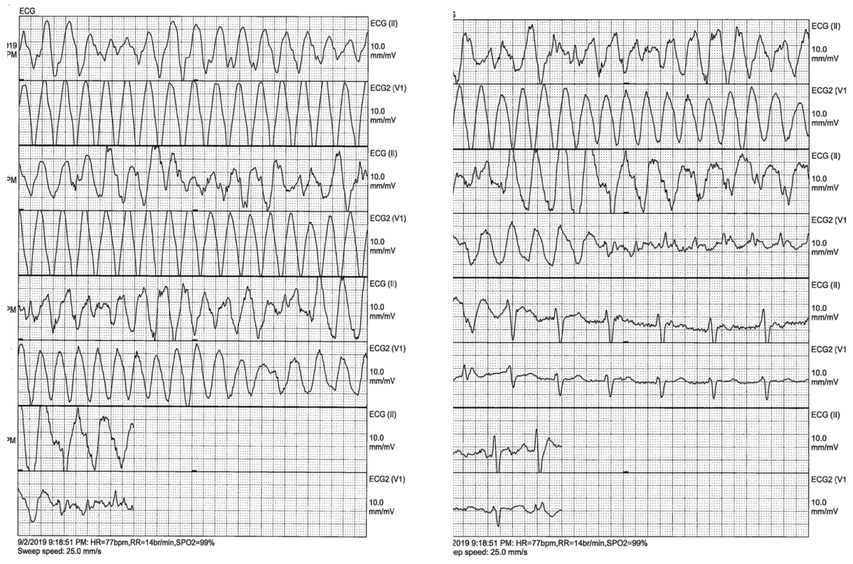
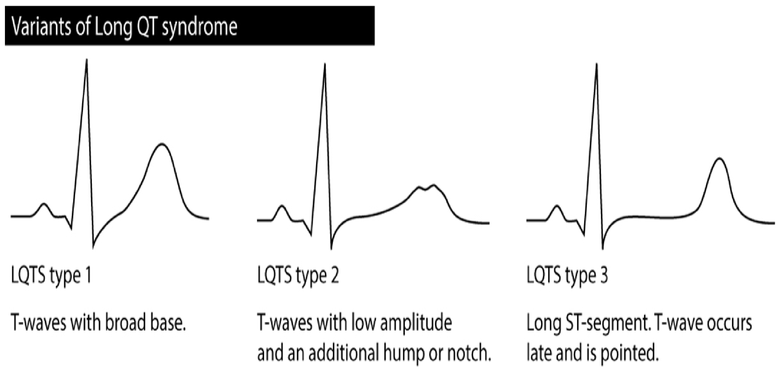
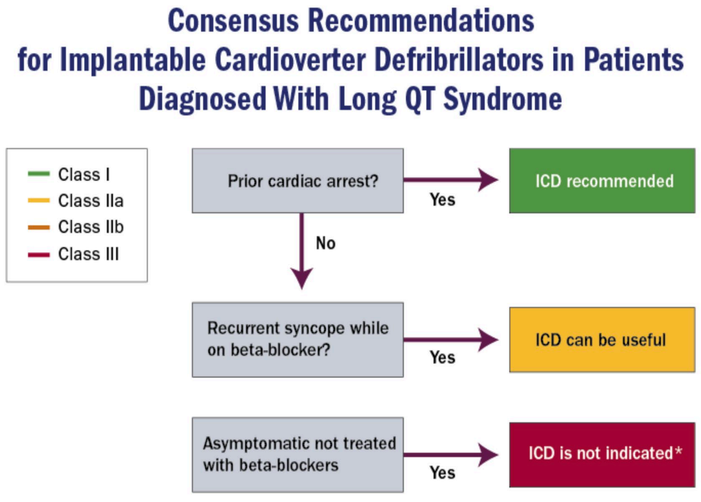


Figure 2: Polymorphic sustained ventricular tachycardia terminated by DCCV



DECISION MAKING

- She was started on propranolol for possible LQTS.
- Cardiac MR did not show any evidence of structural abnormalities.
- Genetic panel was sent.
- Since myocarditis or familial LQTS could not be ruled out, we proceeded with ICD implantation for secondary prevention.
- She was discharged home on nadolol.



DISCUSSION

- In the absence of genetic information, LQTS can be diagnosed in symptomatic patients with QTc >480msec on serial ECGs after excluding secondary causes.
- Schwartz score comprising of ECG findings, symptoms, clinical & family history is diagnostic when greater than 3.5.
- Beta-blockers are indicated in all patients with a clinical diagnosis.
- Patients must avoid any QT prolonging agents and strenuous exercise.
- An ICD is indicated in patients who suffered cardiac arrest.
- ICD may also be considered for primary prevention in high risk patients.

Schwartz score diagnostic criteria for long QT syndrome (LQTS)

	Points
Electrocardiographic findings*	
A. QTc¶	
▪ ≥480 ms	3
▪ 460 to 479 ms	2
▪ 450 to 459 ms (in males)	1
B. QTc¶ fourth minute of recovery from exercise stress test ≥480 ms	1
C. Torsades de pointes△	2
D. T wave alternans	1
E. Notched T wave in 3 leads	1
F. Low heart rate for age◇	0.5
Clinical history	
A. Syncope△	
▪ With stress	2
▪ Without stress	1
B. Congenital deafness	0.5
Family history	
A. Family members with definite LQTS§	1
B. Unexplained sudden cardiac death below age 30 among immediate family members§	0.5