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## OVERVIEW OF THE ECG

At first glance, there are no very obvious abnormalities, aside from some peaking of the T waves.

## MORE DETAILED ANALYSIS OF THE ECG

The rate is 60bpm and is regular. There are P waves before every QRS which are upright in Lead II and inverted in aVR, compatible with normal sinus rhythm. The PR interval is normal (130msec). The QRS complex is 80ms wide with a normal axis of +60°. There are tiny non-pathological Q waves in the inferior leads and septal Q waves in the lateral leads. There are small J waves in the inferior and lateral leads. The ST segments are normal, as is the T wave axis. The T waves are somewhat peaked in the chest leads. The QT interval is difficult to measure accurately, but is prolonged in all 12 leads (Figure 1). The longest is in V1 - 2 at 590ms. The average R-R interval is 1 second and the QTc is therefore 590ms (Figure 2) – considerably prolonged.

Measurement of QT can be difficult in the event of flat or inverted T waves or prominent U waves. Make use of multiple simultaneous leads to get the best estimate of the end of the T wave. Always measure the longest QT interval (usually V2 - V4). While some advocate using a tangent to the downslope of the T to estimate where it would reach the baseline (the tangent method), Professor Peter Schwartz, who has studied the congenital LQTS for more than 50 years, recommends trying to define the actual end of the T where it intersects the isoelectric line (the threshold method). Another problem is the individual variation in measurement of the QT; diurnal variation in the QT interval is also a factor. The QT interval must be corrected for heart rate. Bazett's formula is the most commonly used. A useful website is www.qtcalculator.org.

The causes of QT prolongation are legion (Figures 3 and 4).

Drugs are far and away the most common culprits. The QT drugs list.[1] (www.crediblemed.org) contains 535 drugs, 66 of which are proven to cause torsade de pointes ventricular tachycardia (Figure 5). Most of these drugs affect the $I_{Kr}$ potassium channel which has been labelled "promiscuous".

Metabolic causes include: Hypokalaemia, hypomagnesaemia, hypocalcaemia and hypothermia.

Cardiovascular causes include: Bradycardia, stroke or other cerebral injury, heart failure, and acute myocardial ischaemia.

The congenital long QT syndrome (LQTS) is a less common but vitally important cause, as it is associated with a significant risk of sudden cardiac death in young people.

In the absence of clinical information, it is difficult to reach a definitive diagnosis from this ECG alone. There are, however, features which make some diagnoses more or less likely.

Hypokalaemia causes T wave flattening and prominent U waves, and is therefore excluded. Isolated hypomagnesaemia is uncommon and is usually associated with hypokalaemia. The main findings are a prolonged QTc and increased P wave duration.[2] Hypocalcaemia causes a characteristic prolongation of the ST segment and the QTc, unlike this ECG. Hypothermia also prolongs the QTc, together with bradycardia and a prominent J (Osborne) wave – more marked than in this ECG.

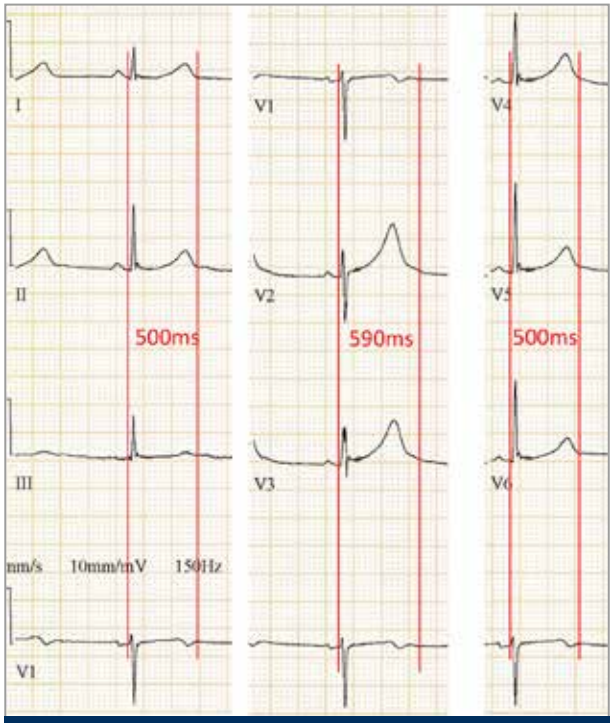


**FIGURE 1:** Measurement of the QT interval using the threshold method in different leads. It is possible that the longer QT measured in V1 - 3 incorporates a U wave, but the minimum QT/QTc (heart rate 60) is considerably prolonged at 500ms.

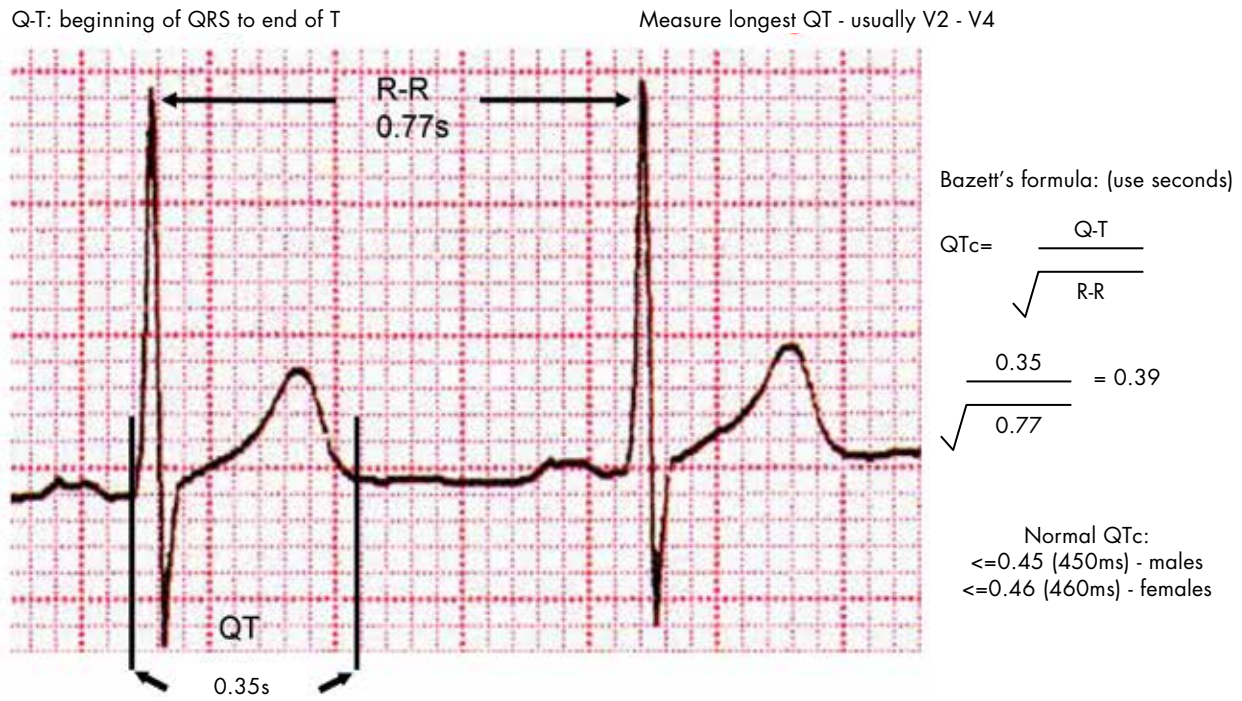


**FIGURE 2:** Measuring and correcting the QT interval (threshold method).

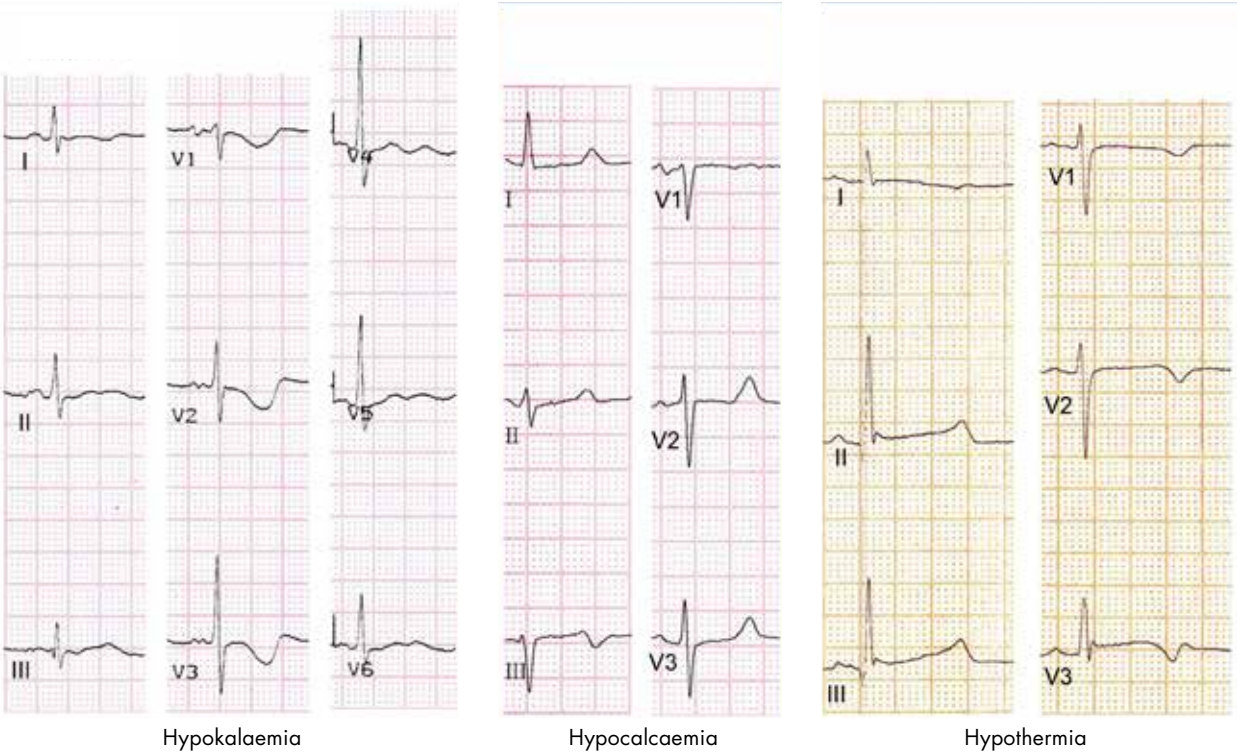


**FIGURE 3:** Metabolic causes of QT prolongation.
Hypokalaemia: QT/QTc 430/490ms; hypocalcaemia: QT/QTc 670/540ms; hypothermia: QT/QTc 670/540ms. The QT is particularly difficult to measure in hypokalaemia because of the prominent U wave.

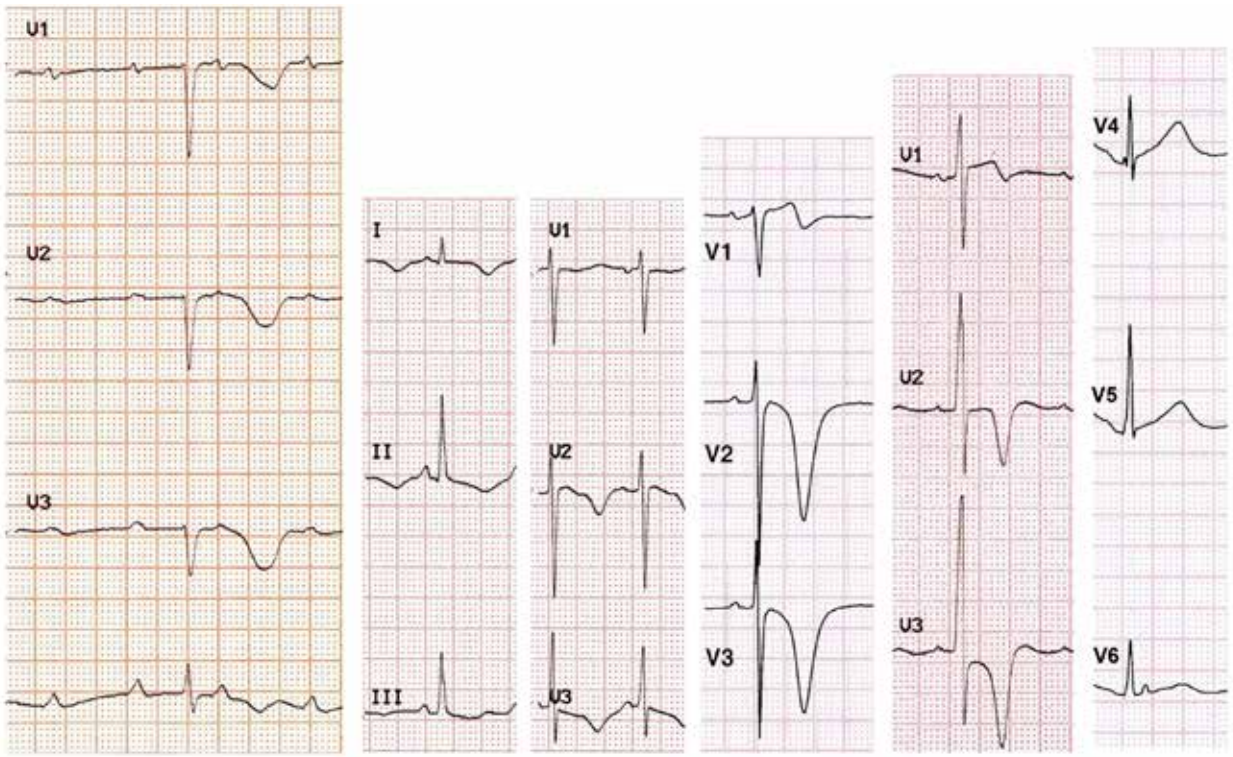


**FIGURE 4:** Miscellaneous causes of QT prolongation.
A: Bradycardia due to complete heart block: QT/QTc 720/540. The extremely long QT makes torsade de pointes very likely (see Figure 5). B: Subarachnoid haemorrhage: QT/QTc 475/590. C: Wellens' syndrome: QT/QTc 510/505ms. D: Apical hypertrophic obstructive cardiomyopathy. QT/QTc 450/490ms. E: Sotalol, one of many drugs known to prolong the QT and cause torsade de pointes: QT/QTc 580/630ms.

Bradycardia, especially when caused by heart block, prolongs the QT and often the QTc. A QT of 590ms at a rate of 60bpm cannot be explained by bradycardia. There is no other evidence of acute ischaemia. Cerebral insult may cause bradycardia and QT prolongation but is usually accompanied by T wave inversion and / or ST segment deviation.

Drug-induced QT prolongation must always be strongly considered, with females more susceptible than males. Often, more than one drug is involved, either because both prolong the QT or one interferes with the metabolism of the other. Additional hypokalaemia may be an aggravating factor and may precipitate torsade de pointes.

**The answer to question 1 is therefore (e): All of the above is an acceptable answer, the most likely are drug-induced or the congenital Long QT Syndrome (LQTS).**

The patient was a young woman with genetically proven LQTS type 1.

**The correct answer to question 2 is therefore (a): A detailed history is clearly vital.**

In her case, there was a family history of sudden cardiac death. An uncle had died suddenly as a child and a cousin had died while water-skiing. She had had episodes of syncope related to exertion; the most recent occurred while swimming from which she was resuscitated from near drowning.

Examination was normal. A stress ECG was not performed. The expected response during effort would be the failure of the QT interval to shorten appropriately.

## COMMENT

The congenital LQTS first came to clinician's attention in the 1957 with the description of the Lange-Jervell-Nielson syndrome.[3] They described syncope and sudden death in children with congenital deafness and noted the long QT on their ECGs. Later, Romano and Ward described a similar dominantly inherited syndrome without deafness.[4,5] Much work followed

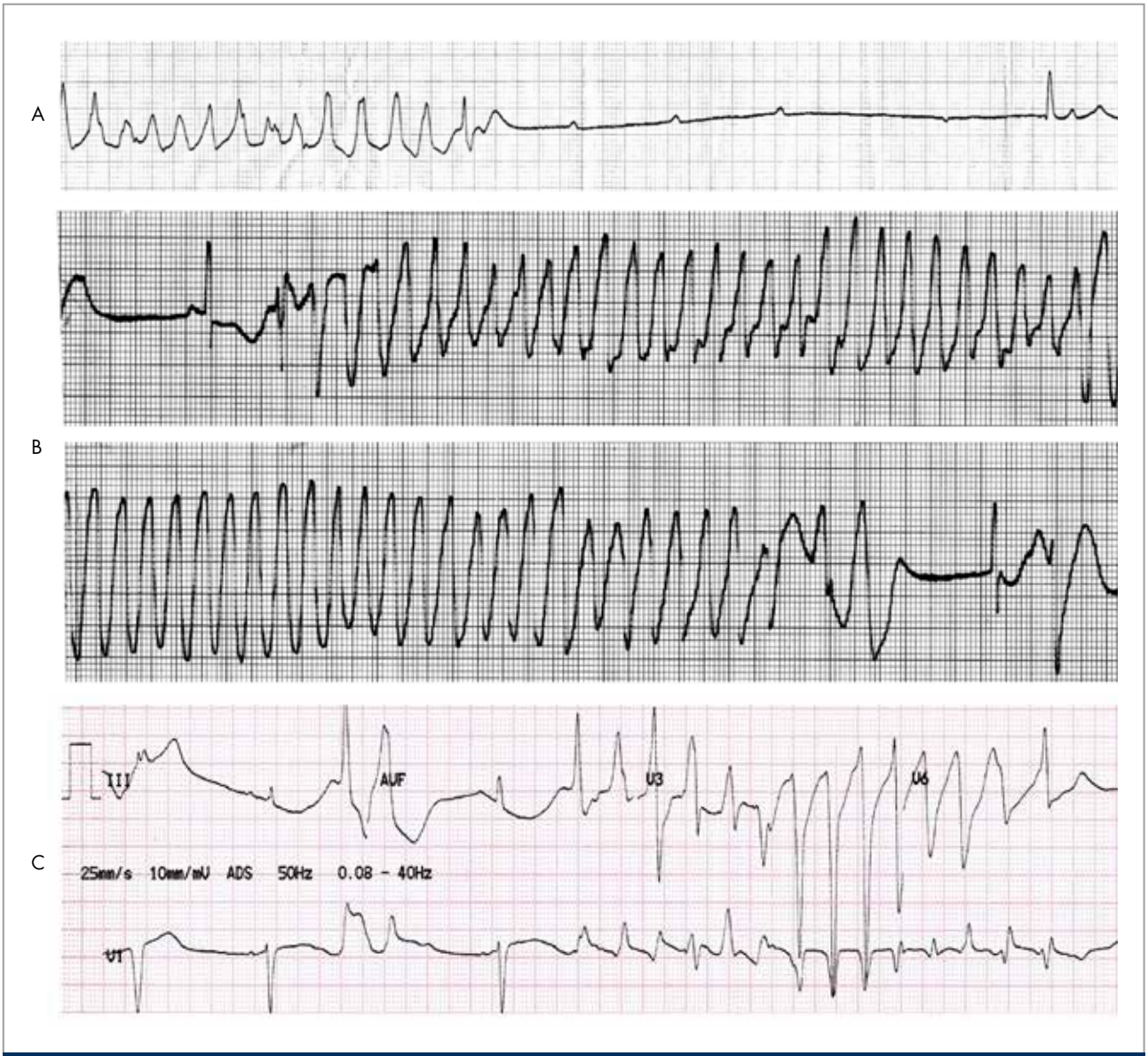


**FIGURE 5:** Examples of torsade de pointes (TdP) ventricular tachycardia related to QT prolongation. Note the twisting pattern of the QRS complexes.
A: complete heart block; TdP is followed by ventricular asystole, either of which may be fatal. B: TdP due to quinidine used to cardiovert atrial fibrillation. C: TdP caused by sotalol, a Class III antiarrhythmic agent with a relatively high risk of TdP, particularly at higher doses, in the presence of hypokalaemia or when another QT prolonging drug is added.

to elucidate the cause. Professor Peter Schwartz and colleagues from Pavia, Italy, demonstrated that QT prolongation and electrical instability could be induced by stimulation of the left stellate ganglion in cats.[6] Subsequent research established the efficacy of both beta blockade and left cervical sympathectomy in preventing syncope and reducing the risk of sudden death.

The condition has proved a Rosetta Stone in the elucidation of the function of cardiac ion channels and their mutations. There are currently 16 known types of congenital LQTS, the most common of which is LQTS type 1. Most of the South African patients are type 1, the result of a founder effect from a Portuguese man who landed here in the late 17th century. Occasional local patients with LQTS types 2 and 3 have been seen. Subtle differences in the ECG findings, other than the QT interval, exist between the different types (Figures 6 and 7).

An international LQTS registry was set up in 1979 by Arthur Moss and Peter Schwartz.[7] This has been an important source of ongoing information about all aspects of the syndrome.

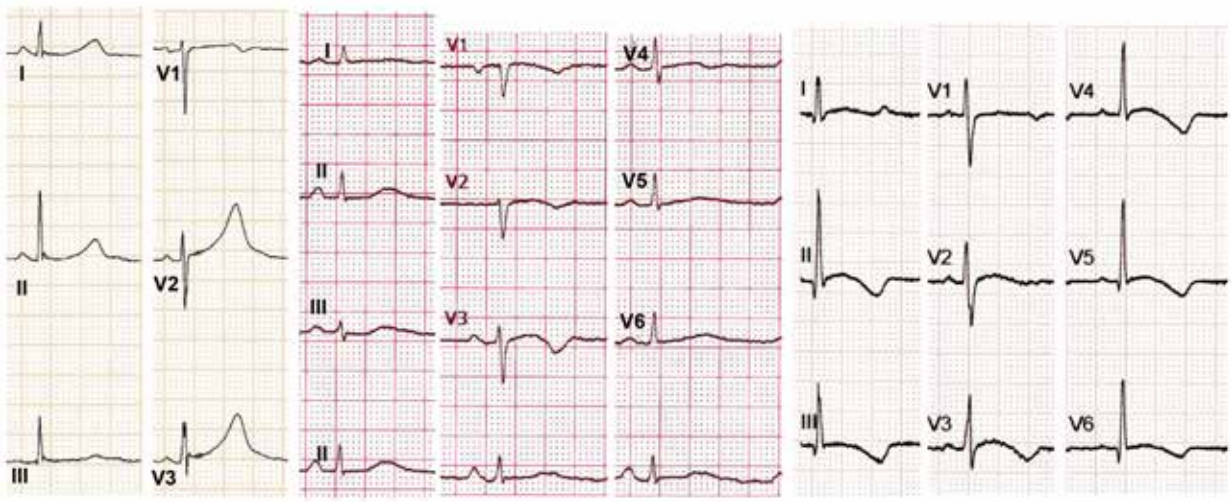


**FIGURE 6:** Examples of the 3 most common types of LQTS from Groote Schuur Hospital (genetically proven).
LQT1 QT/QTc 590ms; LQT2 QT/QTc 550/570ms; LQT3 QT/QTc 550/595ms.

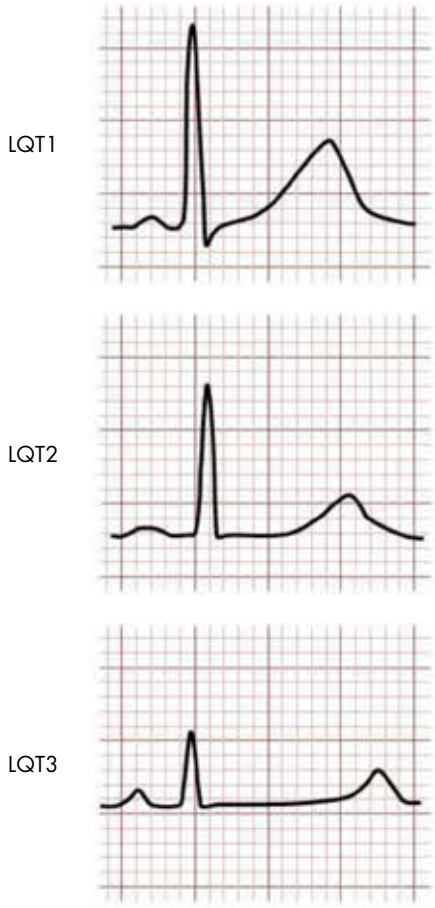


**FIGURE 7:** The classical patterns of the ST segment and T waves of LQT1-3. (Wikipedia creative commons)

**TABLE I:** Diagnostic criteria for congenital LQTS (after Schwartz 2012)

| | | Points |
|---|---|---|
| **ECG findings** | | |
| A. QTc (Bazett's formula) | ≥480 | 3 |
| | 460 - 479 | 2 |
| | 450 - 459 (men) | 1 |
| B. QTc, 4th minute of recovery from exercise stress test | ≥480ms | 1 |
| C. Torsade 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 |
| OR | | |
| B. Unexplained sudden cardiac death younger than age 30 among immediate family members. | | 0.5 |

*LQTS: Long QT Syndrome.*
*QTc in the absence of drugs or other causes of prolonged QT.*
*Torsade de pointes and syncope are mutually exclusive.*
*Resting heart rate below the 2nd percentile for age.*
*The same family member cannot be counted in A and B*
***Score:*** *<1 point: low probability of LQTS. 1.5-3 points: intermediate probability. >3.5 points: high probability of congenital LQTS.*

A scoring system for diagnosing congenital LQTS was developed in 1993, updated in 2012[8] (Table I). Her score is 5.5, which is an unequivocal diagnosis of LQTS, later confirmed to be type 1 by genetic testing.

A comprehensive review of LQTS was published in 2022.[9] I would recommend all cardiologists to read it because of the importance of this relatively rare condition. A heightened awareness of this potentially lethal ECG / clinical diagnosis should help to prevent misdiagnosis, particularly as "epilepsy" in a child.

## LESSONS AND CONCLUSIONS

- QT prolongation is a common and important ECG finding.
- Numerous causes and aggravating factors exist.
- Drugs are the most common culprits, many of which can cause torsade de pointes and sudden death.
- The congenital LQTS is a vital diagnosis to make as a potential cause of sudden cardiac death in children and young adults.

**Conflict of interest: none declared.**

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