Long QT Syndrome (LQTS)

Long QT Syndrome (LQTS) is a heart rhythm disorder in which the QT interval (representing ventricular depolarization and repolarization) is prolonged. The most common "cardiac channelopathy"

PRESENTATION

- **Asymptomatic** *(most common)*
- Palpitations, dizziness, syncope
- ***Sudden Cardiac Death*** *(may be presenting symptom)*

ETIOLOGY

Congenital

Many genes identified in pts, with more being discovered. The most common are:

- LQTS Type 1 (45%) – *KCNQ1 mutation (K+ channel).* Subtypes:
  - Jervell and Lange-Nielsen Syndrome – *Autosomal recessive; Associated with* ***congenital deafness***
  - Romano-Ward Syndrome – *Autosomal dominant*
- LQTS Type 2 (25-40%) – *KCNH2 mutation (K+ channel)*
- LQTS Type 3 (5-10%) – *SCN5A mutation (Na+ channel)*

Acquired (Other)

- Electrolyte imbalances (and several conditions leading to imbalances) →
  - ↓K+,
  - ↓Mg++
  - ↓Ca++ *(ex. Severe vitamin D deficiency)*
- Acute CNS insult – ischemic stroke or intracranial hemorrhage
- Post-myocardial injury or cardiac arrest of any cause

Acquired (Drugs)

- Antiarrhythmics (Class Ia and III)
- Antibiotics (eg. macrolides, fluoroquinolones)
- Antihistamines (eg. diphenhydramine)
- Antidepressants (most TCAs, some SSRIs, lithium)
- Antipsychotics (eg. haloperidol, ziprasidone)
- Anticonvulsants (eg. fosphenytoin, felbamate)
- Others (eg. Ondansetron, azoles, opioids, protease inhibitors)

***** For up to date comprehensive list of QT prolonging medications, visit https://www.crediblemeds.org*****

PATHOPHYSIOLOGY

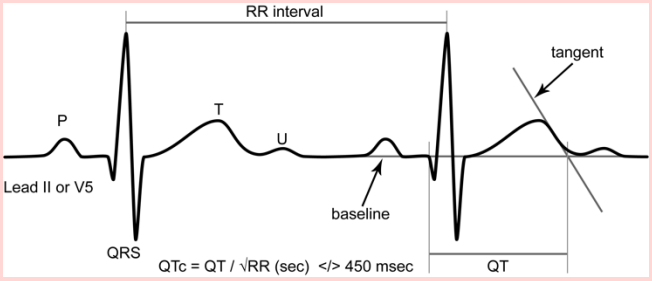
Congenital → Gene mutations = cardiac ion channel dysfunction → **Loss of function** in repolarizing K+ channels **Gain of function** in depolarizing Na+ or Ca++ channels

Acquired →

Prolonged action potential duration in ventricular myocytes, increasing the likelihood of early afterdepolarizations, which can trigger ventricular arrythmias (torsades de pointes (TdP), palpitations, dizzy spells, syncope, or sudden death

DIAGNOSIS

- **Prolonged QT interval on ECG, corrected for HR (QTc)**
  - Males >440ms
  - Females > 460ms
  - Using Bazett formula $QTc = \frac{QT\ (msec)}{\sqrt{RR\ (msec)}}$
  - Usually best to measure Lead II or V5
- Genetic testing (esp with family history)(may not always yield results)



Risk factors for sudden death:

- ECG findings – bradycardia for age, QTc >550ms, abnormal T-wave morphology, T wave alternans, previous TdP or VF
- Symptoms at presentation (syncope, seizures, arrest)
- Young age at presentation (<1m)
- Noncompliance with medications

MANAGEMENT

- If acquired → remove offending agent
- **First line =** Beta blockers (Nadolol or Propanolol preferred)
- High risk patients **NOT** responsive to first-line → Implantable cardioverter defibrillator (ICD) or left cardiac sympathetic denervation surgery

May 2025

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