NICCS Network for Inherited Cardiac Conditions Scotland

Investigation & referral pathway for suspected inherited Long QT syndrome (LQTS)*

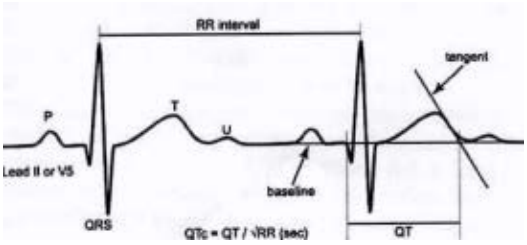
Inherited LQTS is a rare (~1 in 2000) genetic abnormality of cardiac repolarisation. There are several genetic types with differences in certain clinical features. All types are associated with ventricular arrhythmia and patients may present clinically with:

- Asymptomatic QTc prolongation on 12-lead ECG (♂ >460ms , ♀ >470ms)
- Syncope (or seizures)
- Polymorphic VT (Torsades de Pointes [TdP], which may present in bursts)
- Resuscitated VF**
- Sudden cardiac death (SCD) in a family member**

Syncope, TdP and SCD in LQTS may have ***potentially avoidable*** triggers, including: stress, exertion, QTc-prolonging drugs (www.crediblemeds.org), electrolyte imbalance, post-partum state, swimming (LQT1), auditory stimuli (LQT2) and rest/sleep (LQT3).

QTc intervals vary in individuals, so (i) an isolated prolonged QTc may be normal (usu. <500ms) and (ii) patients with LQTS may have normal QTc intervals.

1) Assess QTc using standard 12-lead ECG



a) Automated QT measurements are generally accurate, however it is useful to verify manually† - use tangent method (as shown) and Bazett's‡ correction formula, available at https://www.mdcalc.com/corrected-qt-interval-qtc or https://www.qtcalculator.org

b) Consider causes of acquired LQTS & if present, reassess QTc after correction

- drug effects (www.crediblemeds.org)
- electrolyte imbalance, hypothyroidism, acute coronary syndrome

c) Arrange repeat 12-lead ECG (within days-weeks) with sit-stand if possible

2) Consider high-risk features

Cardiac arrest, TdP*** (inc. non-sustained arrhythmia) or QTc > 500ms with syncope/seizures

→ discuss with cardiologist/heart rhythm specialist, take a blood sample for genetic testing‡

3) Refer for evaluation

a) QTc prolongation (♂ >460ms , ♀ >470ms) on ≥ 2 ECGs, without acquired cause

⟶ refer to ICC clinic, consider LQTS advice‡‡ and taking a blood sample for genetic testing‡

In addition, if high index of suspicion (QTc >500ms, QTc >480ms with syncope/seizures, family history LQTS/SCD, or congenital deafness)

⟶ give LQTS advice‡‡ pending evaluation, take a blood sample for genetic testing‡

b) If QTc prolongation outwith above criteria refer to cardiology for further evaluation

For cardiologists, where the phenotype is unclear consider sit-stand ECG, ETT†† and echo please use appropriate coding in clinical correspondence:

Long QT syndrome	**ICD-10**	**I45.81**	**READ**	**G56y5**
ECG shows prolonged QT interval (not yet diagnosed)	**ICD-10**	**R94.31**	**READ**	**G3217.00**

** See specific NICCS investigation & referral pathways:
† if ventricular pacing or QRS duration (QRSd) > 120ms use adjusted QTc (adQTc) = QTc – (QRSd – 100ms)
‡ Fridericia if over 100 bpm and Framingham if under 60bpm (formulae available in both online calculators)
‡ EDTA sample labelled as per blood transfusion to clinical genetics, if consent or type of testing is unclear DNA can be extracted and saved
*** stop QTc-prolonging drugs, correct K^+ to 4-5mmol/L, consider IV Mg^{2+} ± pacing / isoprenaline for TdP
†† for assessment of QTc at 4 minutes into recovery (>480ms abnormal)
‡‡ Available at https://www.niccs.scot.nhs.uk/