

## 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:

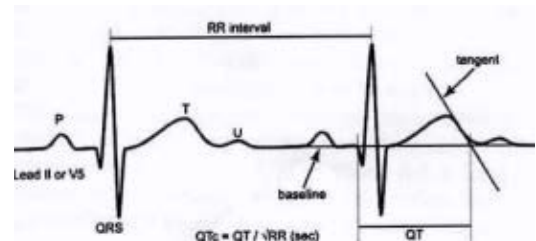
- Asymptomatic QTc prolongation on 12-lead ECG (♂ >460ms , ♀ >470ms)
- Syncope (or seizures)
- Polymorphic VT (Torsades de Pointes [TdP], which may present in bursts)
- Resuscitated VF\*\*
- Sudden cardiac death (SCD) in a family member\*\*

Syncope, TdP and SCD in LQTS may have **potentially avoidable** triggers, including: stress, exertion, QTc-prolonging drugs ([www.crediblemeds.org](http://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<sup>†</sup> - use tangent method (as shown) and Bazett's<sup>‡</sup> correction formula, available at <https://www.mdcalc.com/corrected-qt-interval-qt-c> or <https://www.qtcalculator.org>



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

- drug effects ([www.crediblemeds.org](http://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<sup>\*\*\*</sup> (inc. non-sustained arrhythmia) or QTc > 500ms with syncope/seizures

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

### 3) Refer for evaluation

a) QTc prolongation ( $\text{♂} >460\text{ms}$  ,  $\text{♀} >470\text{ms}$ ) on  $\geq 2$  ECGs, without acquired cause

→ refer to ICC clinic, consider LQTS advice<sup>††</sup> and taking a blood sample for genetic testing<sup>†</sup>

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

→ give LQTS advice<sup>††</sup> pending evaluation, take a blood sample for genetic testing<sup>†</sup>

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<sup>††</sup> 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

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\*\* See specific NICCS investigation & referral pathways:

† if ventricular pacing or QRS duration (QRSd)  $> 120\text{ms}$  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<sup>+</sup> to 4-5mmol/L, consider IV Mg<sup>2+</sup> ± pacing / isoprenaline for TdP

†† for assessment of QTc at 4 minutes into recovery ( $>480\text{ms}$  abnormal)

†† Available at <https://www.niccs.scot.nhs.uk/>

*\* This pathway is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to pathway recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.*