

NICCS Network for Inherited Cardiac Conditions Scotland

Primary Care investigation & referral pathway for suspected inherited

Long QT syndrome (LQTS)*

Inherited LQTS is a rare (~1 in 2000) genetic abnormality of cardiac repolarisation associated with a risk of ventricular arrhythmia, which may present as:

- Asymptomatic QTc prolongation (♂ >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**

1) Check automated QTc interval using standard 12-lead ECG

If QTc prolonged (♂ >460ms , ♀ >470ms) consider causes of acquired LQTS

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

If **YES**, address cause and repeat ECG

If **NO** (or other) refer for evaluation (ideally arrange repeat ECG)

2) Consider high-risk features

If QTc > 500ms with syncope or seizures

→ refer for urgent in-patient assessment

3) Refer for evaluation

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

→ refer direct to ICC clinic, if available locally

b) If QTc prolongation outwith above criteria (or direct ICC referral not available)

→ refer to cardiology

** See specific NICCS investigation & referral pathways:

‡ 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.*