How to identify long QT syndrome and individuals most likely to benefit from referral to genetics and a cardiac arrhythmia specialist

Long QT syndrome (LQTS) is one of several inherited heart disorders than can lead to sudden cardiac death (SCD). LQTS is a rhythm disorder that can predispose to fast, chaotic heart rhythm which may trigger a sudden fainting spell, seizure or SCD. It is treatable if diagnosed. The ECG is neither sensitive nor specific for hereditary LQTS. Individuals with clinical features or family history shown in Box 1 should be referred to a cardiac arrhythmia specialist and a genetics clinic, for assessment and genetic testing where indicated. A QTc ≥ 500ms is considered high risk for LQTS. Family physicians can play a crucial role in referring first degree relatives to cardiac genetics specialist services following the death of a young person in whom autopsy did not identify cause of death or in whom a heritable cardiac disorder was suspected.

- Syncope (*loss of consciousness*) or near syncope spells triggered by:
  1. Physical exertion
  2. Auditory stimuli e.g. fire alarm
  3. Emotional stress/distress
  - Repetitive events more concerning
  - Excluding events that are likely due to vasovagal events is difficult but necessary (e.g. those occurring during abrupt postural changes, exposure to heat and dehydration, emotional reactions to events such as blood draw, etc.)
- Family history of unexplained sudden death in otherwise healthy persons at a young age (< 40 years)
  - Attention to: unexplained death during swimming, death during seizures, a family history of "seizure" disorders and other unexplained deaths
    - Sodium-channel abnormalities may be precipitated by fever. These cardiac events may appear seizure-like and may be mislabelled as epilepsy.
- Corrected QT interval of:
  - Men: >450ms
  - Women: >470ms

Box 1. Clinical symptoms, signs and family history to prompt referral to cardiac arrhythmia specialists and genetics clinic for assessment.

For a comprehensive, up-to-date list of QT-prolonging medications see www.crediblemeds.org
Access is free, requiring a one-time registration.