Long QT Syndrome in Children

2019 LQTS Patients and Families Seminar

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Agenda

1. Accurate diagnosis
   1. Symptomatic patients
   2. Family members

2. Risk stratification

3. Treatment
   1. Trigger avoidance & safety plan & impact on sports
   2. Medication
   3. Surgery

4. Planning ahead
Accurate Diagnosis
Accurate diagnosis – symptomatic patients

Symptoms of LQTS

- Fainting, especially with exercise or emotion
- Seizures, especially with exercise or emotion
- Near-drowning
- Cardiac arrest

Rarely symptoms of LQTS

- Chest pain
- Palpitations
- Lightheadedness/fainting upon standing up, etc.
- Symptoms that persist for long periods of time
Accurate diagnosis – symptomatic patients

**Education** (doctors, nurses, athletic trainers, etc)
- Recognition of symptoms
- Recognition of family history

**Accurate interpretation of ECG [EKG]**
- Abnormal ECGs are often overlooked
- Normal ECG never excludes LQTS
- Referral if suspicious symptoms

**17 yo LQTS type 1 w/SCA, QTc 440**
Accurate diagnosis – family members

Extremely important to screen families
- LQTS is dangerous without treatment
- LQTS is usually very manageable with treatment

Testing relatives by ECG is **not** enough
- Genetic testing for the most severely affected person in the family
  - Then chase relatives near & far
- If no clear gene:
  - Contemplate the affected person (QT longer on stress test?)
  - Evaluate relatives appropriately

**Families are complicated** – but these efforts save lives
Risk stratification

Risk for future event without treatment varies *widely*

- LQTS genetic type (and even specific mutations)
  - Age/gender effect
- QT length
- Other medical issues that can interact

**Key principle:** treatment intensity should match level of risk

- More risk → more treatment
- Less risk → less treatment

Risk can change over time, always re-evaluate
Risk stratification example #1

6 year old borderline QT on screening ECG
  • No symptoms, no family history

Genetic testing → LQTS type 1
  • Specific mutation common in part of Sweden
  • Known to be mild (SCA 1% untreated)

Options:
  • Trigger avoidance only
    or
  • Non-aggressive beta-blockade

Figures from Winbo et al 2014
Risk stratification example

Father – mildly long QT on routine ECG
  • No symptoms, genetic testing → LQT2
  • Daughter then in preschool
    • Gene positive, mildly long QT
    • Considered low risk, low-dose nadolol

Years later, daughter entering puberty
  • Faints while excited
  • Walks in with QTc>600 +/- Torsades
  • Now very high risk
    • Maximize meds + defibrillator
Treatment

(more on this later in the day)
Treatment – trigger avoidance & safety plan

**Lifestyle modification re triggers**
- Adrenaline – sports, emotion
- Sudden noises – phones, alarm clocks, fire alarm at school

**Avoiding QT-prolonging medications**
- CredibleMeds (smartphone app or [www.QTdrugs.org](http://www.QTdrugs.org))
- Advocate for yourself (*always* double-check)
- Exceptions can be made
  - Need input from EP
  - Some meds are *much* worse than others (even in the same class)

**Avoiding electrolyte disturbances, fever**
- Overheating/dehydration
- GI illness (“stomach bug”)

Treatment – trigger avoidance & safety plan

Safety plan for home

- CPR training
- AED?
- Adult supervision especially with exercise

Safety plan for school

- CPR training
- Adult supervision
- Written emergency plan (and practice it!)  [www.heart.org/CERP](http://www.heart.org/CERP)
- May need 504
Treatment – sports considerations

Adrenaline is a trigger, especially for LQT1

But sports are very important for physical and emotional health

At diagnosis – wait until evaluation complete & treatment plan in place

• Then shared decision with child, parents, doctor

Pick your battles:

• Must have adult supervision and AED
• Where will exercise be? (school gym vs neighbor’s backyard)
• How bad is fainting? (swimming, climbing, skiing)
• How easy is rescue? (track vs cross-country)
Treatment - medications

Beta blockers

- Nadolol most common  (strong and long-acting)
- Propranolol also good  (but awkward pill sizes)
- Nothing else is reliable
- Most people tolerate nadolol or propranolol if started very slowly

Who needs treatment (in childhood)?

- Depends on risk profile
- High-risk patients definitely
- Low-risk patients
  - Usually try and see if tolerated
  - Carefully chosen patients may be okay with trigger avoidance only
Treatment - medications

Other medications can sometimes be added

- LQT1
  - Not really needed – we have denervation surgery as “plan B”

- LQT2
  - Spironolactone/potassium
  - Mexiletine

- LQT3
  - Mexiletine & others
Treatment - surgery

Left cardiac sympathetic denervation ("sympathectomy")

- Minimally invasive
- Works very well for LQT1 and pretty well for LQT2 when:
  - Beta blocker not tolerated
  - Beta blocker not enough (breakthrough events, high-risk features)

Pacemaker alone

- Very rare now

Defibrillator (w/pacemaker)

- Cardiac arrest survivors (except special cases)
- High-risk LQT2 and LQT3
- For high-risk LQT1, prefer denervation surgery
Planning ahead
Planning ahead

Sports planning
- Sway young kids towards supervisable activities on the ground

Career planning
- Medical clearance issues: police, fire, military, pilot, etc
- Difficult to get help: forestry, marine biology, etc

Reproductive planning
- Usually 50% recurrence risk
  - *Child may have more or less severe case*
- Medical team input prior to reproduction
  - Education for partner who may be new to this
  - Opportunity to optimize mother’s status/meds (if affected)
- Plan in place for fetal monitoring
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