

# Long QT Syndrome in Children

2019 LQTS Patients and Families Seminar



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# Agenda

## 1. Accurate diagnosis

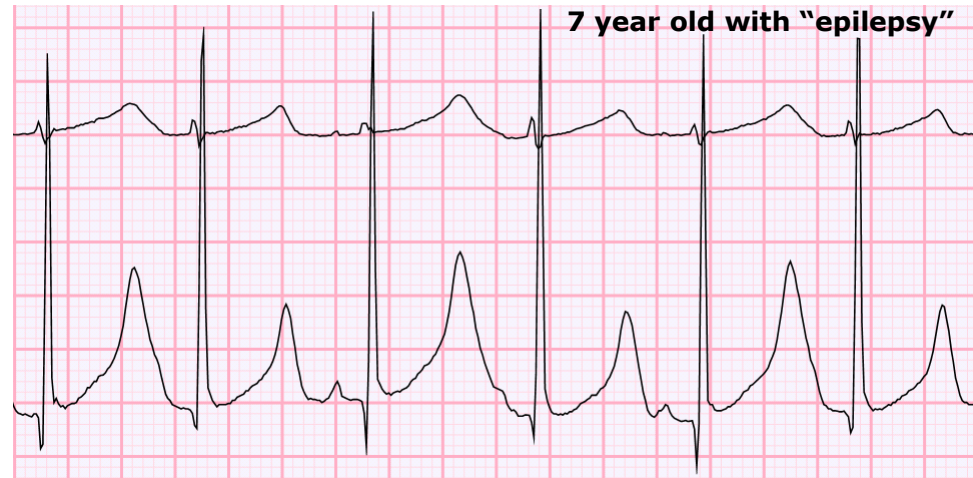
1. Symptomatic patients
2. Family members

## 2. Risk stratification

## 3. Treatment

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

## 4. Planning ahead



# Accurate Diagnosis



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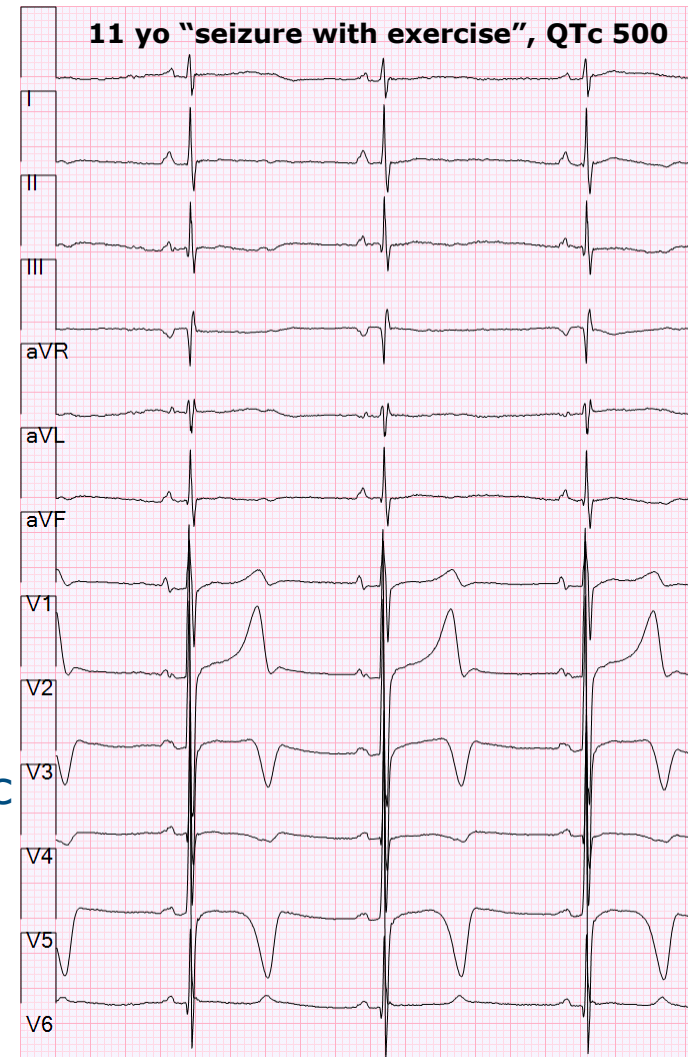
# Accurate diagnosis – symptomatic patients

## 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



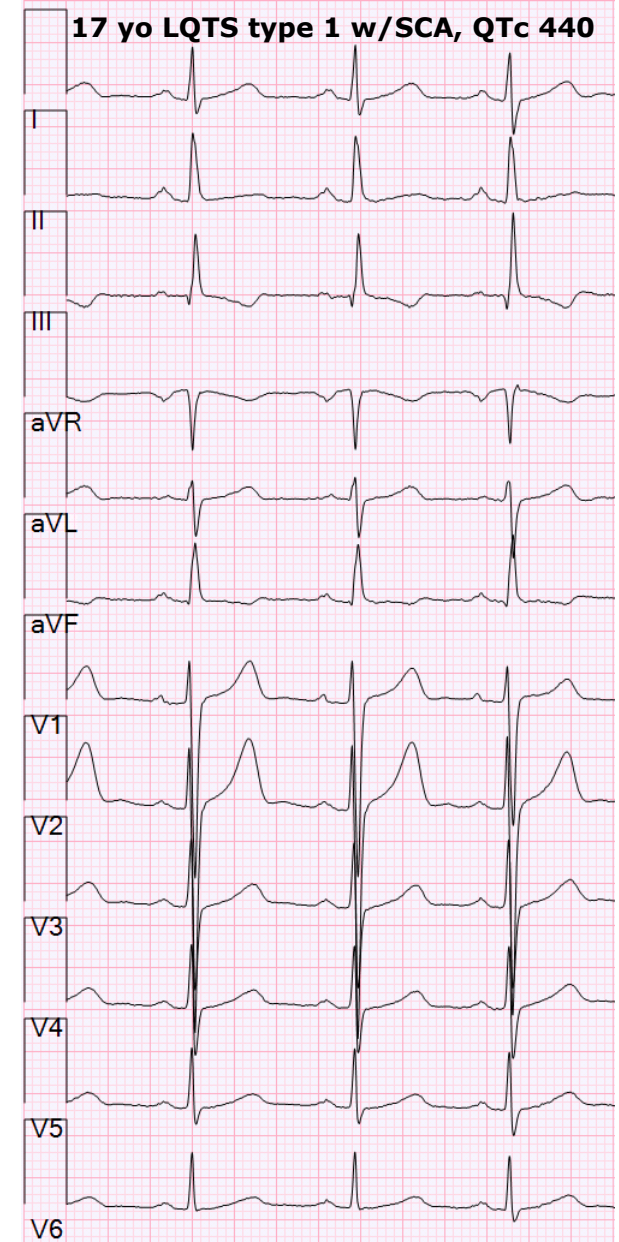
# Accurate diagnosis – symptomatic patients

Education (doctors, nurses, athletic trainers, etc)

- Recognition of symptoms
- Recognition of family history

Accurate interpretation of ECG [EKG]

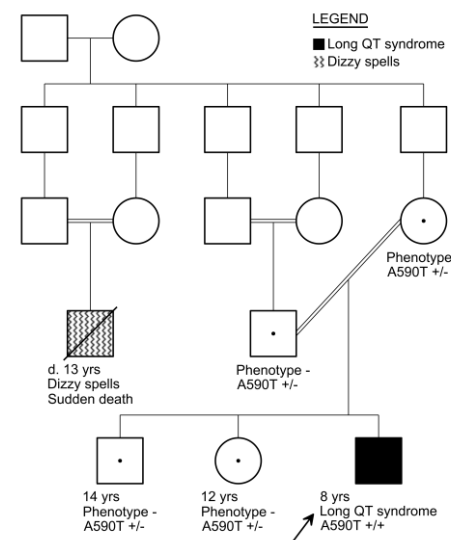
- Abnormal ECGs are often overlooked
- Normal ECG never excludes LQTS
- Referral if suspicious symptoms



# 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



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# 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

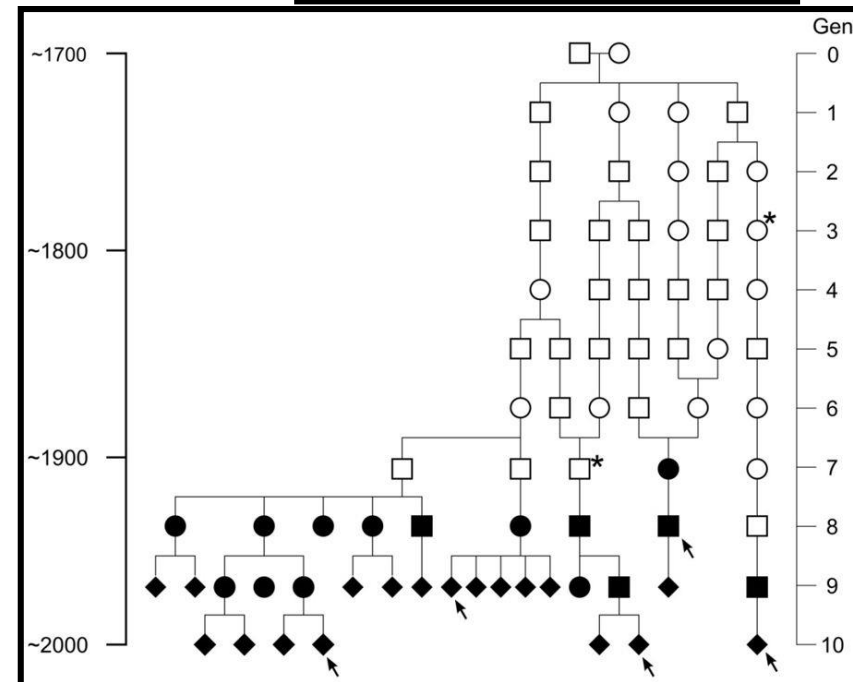
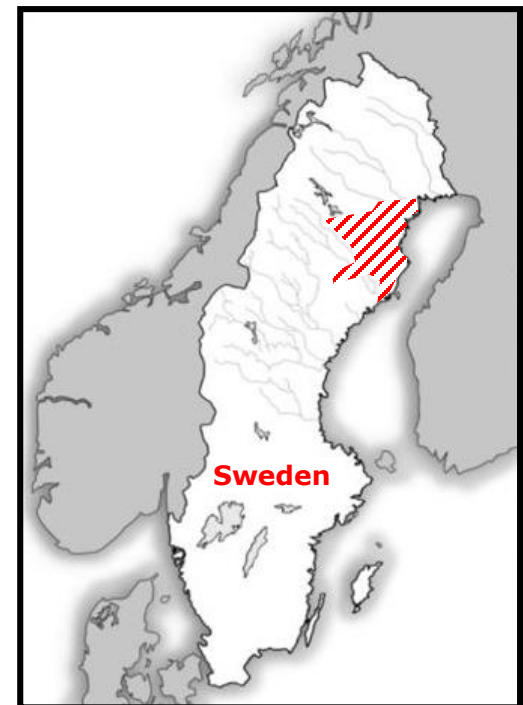
- No symptoms, no family history

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



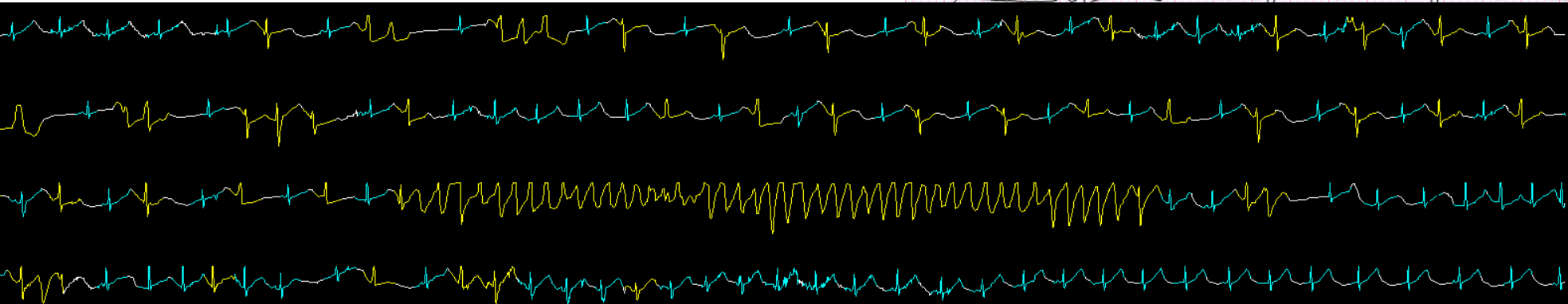
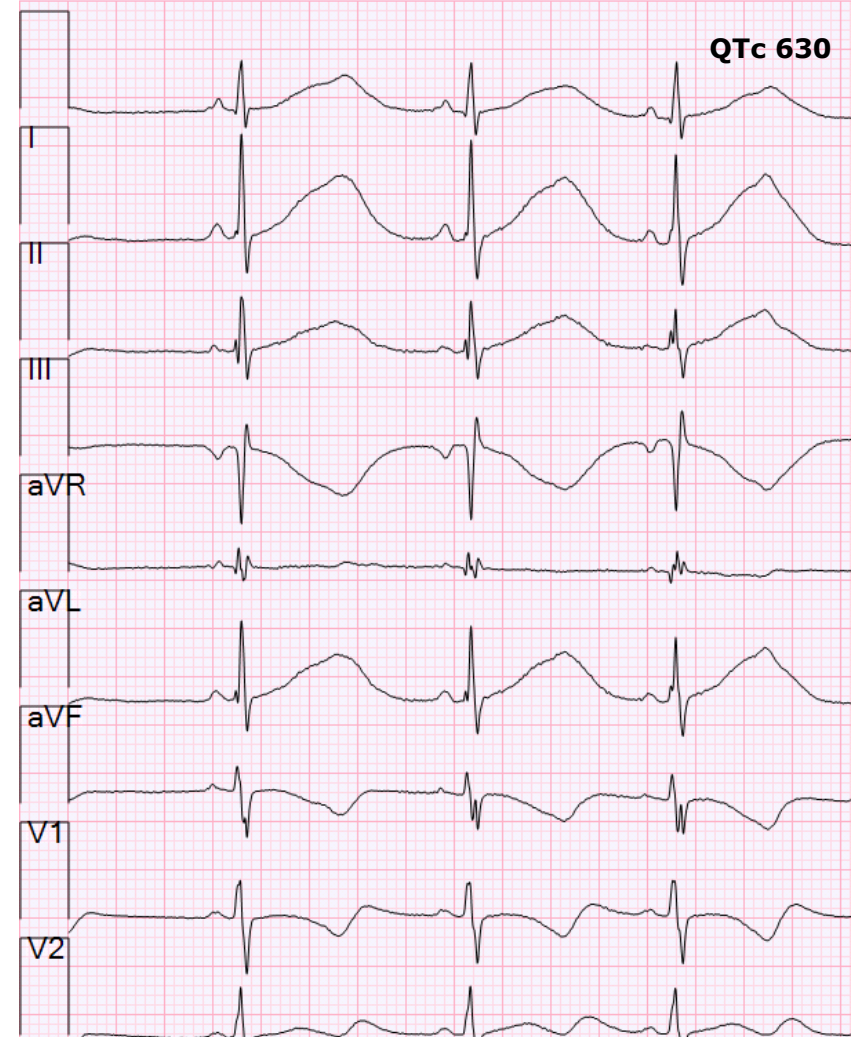
# Risk stratification example

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)



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# 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

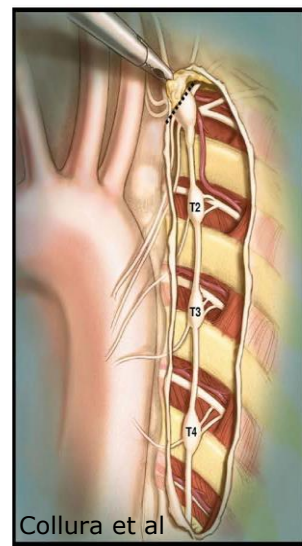
- LQT1
  - Not really needed – we have denervation surgery as “plan B”
- LQT2
  - Spironolactone/potassium
  - Mexiletine
- LQT3
  - Mexiletine & others



# Treatment - surgery

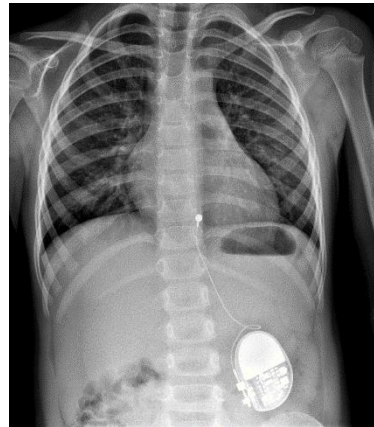
## Left cardiac sympathetic denervation ("sympathectomy")

- 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)



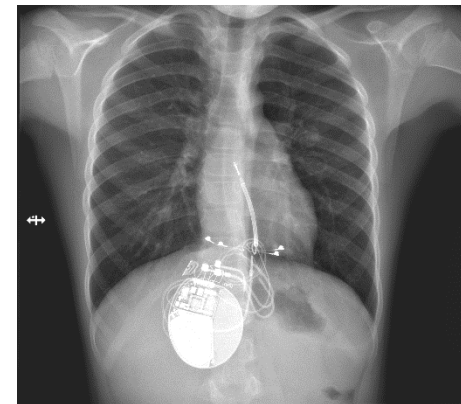
## Pacemaker alone

- 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



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# 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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