Long QT Syndromes – Congenital & Acquired

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Objectives

• Demonstrate how to measure the QTc
• Identify patients at risk for acquired long QT
• Discuss nursing considerations in caring for Long QT syndrome patients.

Measurement of the QT Interval

• Encompasses ventricular depolarization and repolarization
• Locate beginning of QRS complex
• Locate the end of T wave
• Measure R to R interval (for QTc)

Trouble knowing where to measure?

• If the T wave is notched, then the end of the T wave should be considered the end of the entire complex.
• Discrete U waves, which arise after the T wave has returned to baseline, should not be included in the QT interval.
• If difficult to determine the end of the T-wave, suggest drawing a line with the most consistent down slope of the T-wave and measuring to where this bisects the baseline.
**QTc Interval**

- Measure QT & adjust for heart rate:
  - Bazett’s formula:
    \[ QTc = \frac{QT}{\sqrt{R-R \text{ interval}}} \]
  - Example: \( QT = 400 \text{ msec} \)
    
    \[ \frac{\sqrt{R-R}}{0.8} = 0.89 \text{ sec} \]
    
    so \( QTc = 449 \text{ msec} \)

**Normal & Abnormal QT Interval**

<table>
<thead>
<tr>
<th>Age</th>
<th>Men</th>
<th>Women</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt;0.44</td>
<td>&lt;0.43</td>
</tr>
<tr>
<td>Borderline</td>
<td>0.44-0.46</td>
<td>0.45-0.47</td>
</tr>
<tr>
<td>Prolonged</td>
<td>&gt;0.46</td>
<td>&gt;0.45</td>
</tr>
</tbody>
</table>

Consider anything >0.50 abnormal
If BBB or pacemaker >0.60 abnormal

**Causes of QT Abnormalities**

- Acquired Prolonged QT:
  - drug therapy
    - Antiarrhythmics (quinidine, procainamide, amiodarone)
    - Antipsychotics (haloperidol)
    - Antibiotics (erythromycin)
      
      [www.torsades.org](http://www.torsades.org)
  - electrolyte imbalance
    (hypokalemia, hypomagnesemia, hypocalcemia)
  - hypothermia

**At risk for Acquired QT Prolongation**

- Prolonged rapid heart rates
- Following ablation
- Significant bradycardias, heart blocks or frequent pauses

Other warning signs for Torsades

- Polymorphic PVCs, bigeminy, couplets, triplets, V-runs
- T-wave alternans
Case Study
78 y/o woman in ICU being treated with IV erythromycin for pneumonia

Case Study
same patient one hour later.....

T-wave alternans

Congenital Long QT Syndrome

- Congenital disorder
- Prolonged QT interval
- High incidence of lethal ventricular arrhythmias
  - 50% of LQTS patients present with SCD
  - SCD in as many as 80% of affected patients

Two Well-described Syndromes

- Jervell and Lange-Nielsen syndrome
  - long QT
  - congenital nerve deafness
  - syncope or sudden death
  - family history of sudden death
  - autosomal recessive inheritance
- Romano-Ward syndrome
  - same features, except no deafness
  - autosomal dominant inheritance

Pathophysiology

- Inherited abnormality in membrane protein involved in either potassium or sodium channel function
  - action potential prolonged
  - causes early after depolarizations

Torsades de Pointes

- Polymorphic Ventricular tachycardia

- QRS complexes gradually change back and forth from one shape and direction to another - “twisting around a point”
Pathophysiology

Sympathetic imbalance
- Low levels of right cardiac sympathetic activity
- Imbalance causes areas of action potential prolongation
- Sudden sympathetic discharge from left triggers arrhythmia

Genetic Background

<table>
<thead>
<tr>
<th>Type</th>
<th>Gene</th>
<th>Chromosome</th>
<th>Locus</th>
<th>Ion</th>
<th>% of LQTS Population</th>
</tr>
</thead>
<tbody>
<tr>
<td>LQT1</td>
<td>KVLQVT1</td>
<td>11p15.5</td>
<td>I</td>
<td>Ks</td>
<td>45%</td>
</tr>
<tr>
<td>LQT2</td>
<td>HERG</td>
<td>7q35-35</td>
<td>I</td>
<td>Kr</td>
<td>46%</td>
</tr>
<tr>
<td>LQT3</td>
<td>SCN5A</td>
<td>3p21-24</td>
<td>I</td>
<td>Na</td>
<td>6%</td>
</tr>
<tr>
<td>LQT4</td>
<td>Unknown</td>
<td>4q25-27</td>
<td>??</td>
<td>??</td>
<td>6%</td>
</tr>
<tr>
<td>LQT5</td>
<td>KCNE1</td>
<td>21q22.1-.2</td>
<td>I</td>
<td>Ks</td>
<td>12%</td>
</tr>
<tr>
<td>LQT6</td>
<td>MiRP1</td>
<td>21q22.1-.2</td>
<td>I</td>
<td>Kr</td>
<td>12%</td>
</tr>
</tbody>
</table>

Incidence & Significance

- 1 in 3000 - 15000 individuals
  - Jervell and Lange-Nielsen = 10% of cases
  - Romano-Ward is more common
  - Up to 30% are idiopathic
- Mortality rate 6% by age 40
  - Untreated mortality rate up to 71-80%
  - Annual 4.5% mortality rate during treatment
  - After 1st syncope the mortality rate is 20% within 1 year, if untreated, and 50% in 10 years

Presentation

- Unexplained syncope (26%)
  - Related to exercise, noise, or emotion
- Seizures (10%)
- Sudden death or cardiac arrest (9%)
- Presyncope (6%)
- Palpitations (6%)
- Up to 39% LQTS patients are asymptomatic
- Usually in childhood to young adulthood

Diagnosis

- Physical exam may find bradycardia (20%)
- QTc >480 msec
  - Treadmill exercise test
  - Holter monitor
- Documented Torsade de pointes
- History of syncope (especially with stress)
- Family History

Variations Among LQTS Groups

- LQT1
  - 50% are symptomatic by age 10
  - 88% of events are at times of high SNS outflow
  - Swimming is a significant trigger
- LQT2
  - Often auditory trigger when asleep
  - About 50% are asymptomatic till early adulthood
- LQT3
  - Bradycardia is a more frequent finding
  - Less commonly symptomatic
  - Most arrhythmias & lethal events with rest/sleep
Treatment of Torsades de Pointes
• Stop any offending drugs
• Correct electrolyte abnormalities
• Acute and long-term pacing is recommended for patients presenting with torsades de pointes due to heart block, symptomatic bradycardia or pause-dependent torsades
• IV magnesium sulfate is reasonable for patients who present with LQTS but not likely to be effective in patients with a normal QT interval.
• Beta blockade combined with pacing if torsades is related to sinus bradycardia.
• Isoproterenol as temporary treatment in acute patients who present with recurrent pause-dependent torsades de pointes associated with prolonged QT.

Chronic Management of LQTS
• Beta blockers – effective in preventing cardiac events in 70%
• ICD if recurrent events despite treatment
• Pacemaker for arrhythmogenic bradycardia – decreases repolarization heterogeneity
• High left thoracic sympathectomy

Patient Teaching
• Factors that trigger arrhythmias – vigorous exercise – water activities – strong emotions – auditory stimuli (alarm clock)
• Importance of medication compliance
• Family members/school training in CPR
• Avoid high-risk occupations

Support Group
• The Sudden Arrhythmia Death Syndromes Foundation (SADS)
  • 540 Arapeen Drive, Suite 207, Salt Lake City, UT 84108, 800-786-7723.
  • E-mail: sads@mail.aros.net.
  • Internet: www.sads.org.*

Take –Home Message
• QTc should be part of routine rhythm analysis (as is PR interval).
• Be aware of conditions and medications that can prolong the QT interval.

Title
• Text