Long QT case
History of current illness

• The 44 year-old female patient presumed to be normal until presenting to hospital because of aborted cardiac arrest.
• 30 minutes before admission, she suddenly collapsed without any prodrome while sitting on the bus on the way to go home.
• At local ER, the doctor noted ventricular fibrillation and performed cardioversion successfully. Then she was transferred to Tam Duc hospital with ventilator and high dose of inotropic agent infusion.
ECG on admission

QTc= 530ms
After spending 3 weeks in ICU department, she recovered incompletely. She could speak naturally but unable to recognize her relatives. Her EF had improved from 20% to 60%. Coronary and pulmonary arteries angiogram was done with normal result.

QTc = 500ms

Physical examination and clinical investigations are normal
Score $\geq 4$.
Repeated ECG after correcting other factors: $\text{QTc} \geq 500\text{ms}$

Long QT syndrome – aborted sudden cardiac arrest

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**Table 1. Long QT Syndrome (LQTS) Diagnostic Criteria**

<table>
<thead>
<tr>
<th>ECG Findings</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. QT: $\geq 480$ msec$^{10}$</td>
<td>3</td>
</tr>
<tr>
<td>B. Torsade de pointes$^{3}$</td>
<td>2</td>
</tr>
<tr>
<td>C. T-wave alternans</td>
<td>1</td>
</tr>
<tr>
<td>D. Notched T-wave in 3 leads</td>
<td>1</td>
</tr>
<tr>
<td>E. Low heart rate for age$^{4}$</td>
<td>0.5</td>
</tr>
</tbody>
</table>

**Clinical History**

| A. Syncope$^{4}$ With stress | 2 |
| A. Syncope$^{4}$ Without stress | 1 |
| B. Congenital deafness | 0.5 |

**Family History$^{6}$**

| A. Family members with definite LQTS$^{2}$ | 1 |
| B. Unexplained sudden cardiac death below age 30 among immediate family members | 0.5 |

*In the absence of medications or disorders known to affect these electrocardiographic features.

**SCORING:**

1 point, low probability of LQTS; 2-3 points, intermediate probability of LQTS; 4 points, high probability of LQTS.

**EXPLANATIONS:**

$^{1}$ QT calculated by Bazett's formula, where $\text{QT} = \text{QT}/\sqrt{RR}$.

$^{2}$ Mutually exclusive.

$^{3}$ Resting heart rate below the second percentile for age.

$^{4}$ Mutually exclusive.

$^{5}$ The same family member cannot be counted in A and B.

$^{6}$ Definite LQTS is defined by a LQTS score $\geq 4$.

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**ICD implantation with the use of beta-blockers is recommended in LQTS patients with previous cardiac arrest.**

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**2. Long QT Syndrome (LQTS) Expert Consensus Recommendations on LQTS Diagnosis**

1. LQTS is diagnosed:
   a. In the presence of an LQTS risk score $\geq 3.5$ in the absence of a secondary cause for QT prolongation and/or
   b. In the presence of an unequivocally pathogenic mutation in one of the LQTS genes or
   c. In the presence of a QT interval corrected for heart rate using Bazett's formula (QTc) $\geq 500$ ms in repeated 12-lead electrocardiogram (ECG) and in the absence of a secondary cause for QT prolongation.

2. LQTS can be diagnosed in the presence of a QTc between 480–499 ms in repeated 12-lead ECGs in a patient with unexplained syncope in the absence of a secondary cause for QT prolongation and in the absence of a pathogenic mutation.
ICD implantation (Aug, 2015)

- After ICD implantation, although beta blocker (propanolol) combining with phenytoin dose were optimized, she still complained dizziness and near-syncope
- ICD check revealed many polymorphic VTs which were triggered by PVC. PVC burden was 22% on 24H holter ECG.
- She sometimes had 30 VTs in just one day. Luckily, many VTs were terminated by ATP
- The patient was once hospitalized because of these arrhythmia.
Session File

Device Model: Fortify™ ST VR 1255-40Q
Serial No.: 827536
Session Date and Time: Sep 30, 2015, 7:29 am
Operation: Capture Test Results, 7:43 am

1 Alert

Test Results (Today's most recent results shown on top)

Capture

V
- Today: 1.0V @ 0.5mA (8)
  Jul 28, 2015: 1.25V @ 0.5mA (8)
- Today: 11.7mV (8)
  Jul 28, 2015: 11.7mV (8)
- Today: 400c (8)
  Jul 29, 2015: 430c (8)

Sense

V
- Today: 500mV (RV-SVCCAP)
  Jul 29, 2015: 590mV (RV-SVCCAP)

Lead Impedance

HV

Tachy: 3 Zone Configuration

VT-1  VT-2  VF
150 mmHg  181 mmHg  214 mmHg
Minim: ATP2  ATP1  ATP1
Maxim: ATP3  ATP3  ATP3
VT/VF Episodes: 3

VT-1  VT-2  VF
0  0  0
SVT Episodes: 0

Diagnostics Summary

Since Jun 8, 2015

VT/VF Episodes: 21
Since Sep 30, 2015

VT-1  VT-2  VF
Episodes
0  0  21
ATP Delivered
0  0  9
Shocks Delivered
0  0  8

SVT Episodes: 0
Issue

• Long QT syndrome – Electrical storm

• Further treatment:
  1. ICD did a good job. No more intervention
  2. Left sympathetic cardiac nerve denervation – increased medications
Left Cardiac Sympathetic Denervation in the Management of High-Risk Patients Affected by the Long-QT Syndrome

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Background—The management of long-QT syndrome (LQTS) patients who continue to have cardiac events (CEs) despite β-blockers is complex. We assessed the long-term efficacy of left cardiac sympathetic denervation (LCSD) in a group of high-risk patients.

Methods and Results—We identified 147 LQTS patients who underwent LCSD. Their QT interval was very prolonged (QTc, 543 ± 65 ms); 99% were symptomatic; 48% had a cardiac arrest; and 75% of those treated with β-blockers remained symptomatic. The average follow-up periods between first CE and LCSD and post-LCSD were 4.6 and 7.8 years, respectively. After LCSD, 46% remained asymptomatic. Syncope occurred in 31%, aborted cardiac arrest in 16%, and sudden death in 7%. The mean yearly number of CEs per patient dropped by 91% (P < 0.001). Among 74 patients with only syncope before LCSD, all types of CEs decreased significantly as in the entire group, and a post-LCSD QTc <500 ms predicted very low risk. The percentage of patients with >5 CEs declined from 55% to 8% (P < 0.001). In 5 patients with preoperative implantable defibrillator and multiple discharges, the post-LCSD count of shocks decreased by 95% (P = 0.02) from a median number of 25 to 0 per patient. Among 51 genotyped patients, LCSD appeared more effective in LQT1 and LQT3 patients.

Conclusions—LCSD is associated with a significant reduction in the incidence of aborted cardiac arrest and syncope in high-risk LQTS patients when compared with pre-LCSD events. However, LCSD is not entirely effective in preventing cardiac events including sudden cardiac death during long-term follow-up. LCSD should be considered in patients with recurrent syncope despite β-blockade and in patients who experience arrhythmia storms with an implanted defibrillator. (Circulation. 2004;109:1826-1833.)
LCSD and ICDs

Progressively larger numbers of LQTS patients are receiving an ICD, often even without previous cardiac arrest. Although ICDs can prevent SD, they do not prevent arrhythmic episodes requiring shocks. The present data show that when LCSD was performed after ICD implant because of excessively frequent shocks, it reduced the median number of shocks per patient from 25 to 0 with a reduction of 95%. Thus, in patients with previous cardiac arrest, ICD and LCSD are not mutually exclusive and may complement each other by providing prevention of SD and by improving quality of life through reduction the number of shocks.
LCSD and the QT Interval

LCSD shortened QTc duration by an average of 39 ms. The patients who continue to have a QTc ≥500 ms 6 months after surgery remain at higher risk for subsequent events, and an ICD implant might be considered. Thus, LCSD seems to act in some patients not only by interfering with the “trigger,” represented by sympathetic activation, but also by favorably changing the arrhythmogenic substrate, represented by QT interval duration.

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Left cardiac sympathetic denervation should be considered in patients with symptomatic LQTS when
(a) Beta-blockers are either not effective, not tolerated or contraindicated;
(b) ICD therapy is contraindicated or refused;
(c) Patients on beta-blockers with an ICD experience multiple shocks.

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death
LSCND was done by the end of 2015
QTc = 450ms (on phenyltoin)
PVC burden/holter: 10 to 22.6%
Follow-up after LSCND

- Numbness on the left arm and self-termination within 2 weeks
- Stable hemodynamic status
- No disorder of sweat secretion at the upper part of her body
- Denied syncope or pre-syncope or dizziness
- ICD check for the first 3 months did not record VT/VF.
3 months later, her symptoms recurred. ICD check detected many VT/VF. Some was suppressed by ATP, others required electrical shock.
Frequent PVC
PVC trigger ablation
Thermal trigger during RF
PVC 2
ECG after ablation
QTc = 480ms
• There is no ventricular arrhythmia has been detected.

• No more PVC on 24H Holter ECG after 1 month
Session File

Device Model: Fortify™ ST VR 1235-40Q
Serial No: 827336
Session Date and Time: 23 Jan 2018, 7:30
Operation: Capture Test Results, 7:36

2 Alerts

Device upgrade for battery performance & cybersecurity is available

ST Thresholds at 100%, limiting ST Episode detection. Suggested Thresholds available.

Tachy: 3 Zone Configuration

<table>
<thead>
<tr>
<th>Mode</th>
<th>VT-1</th>
<th>VT-2</th>
<th>VF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Base Rate</td>
<td>150 min⁻¹</td>
<td>181 min⁻¹</td>
<td>214 min⁻¹</td>
</tr>
<tr>
<td>V Pulse Amp</td>
<td>ATPx3, 25.01, 36.01, 40.0x2</td>
<td>ATPx1, 25.01, 36.01, 40.0x2</td>
<td></td>
</tr>
<tr>
<td>V Pulse Width</td>
<td>0.5 ms</td>
<td>0.5 ms</td>
<td></td>
</tr>
</tbody>
</table>

VT/VF Episodes: 0

VP: <1 %
QTc: 480ms
Follow-up

- No AAD
- ICD check every 6 months
- Free of symptoms
CONCLUSIONS

• Long QT is one of the most common causes of SCD in adult patient
• Beta blocker and ICD are the cornerstone in management long QT syndrome.
• LSCND and PVC trigger ablation can be an adjunctive treatment for recurrent ICD shocks
Thank you very much