

## Wild Card Cases IV

### **All in the Family: Managing Relatives of LQTS SCD Victims**

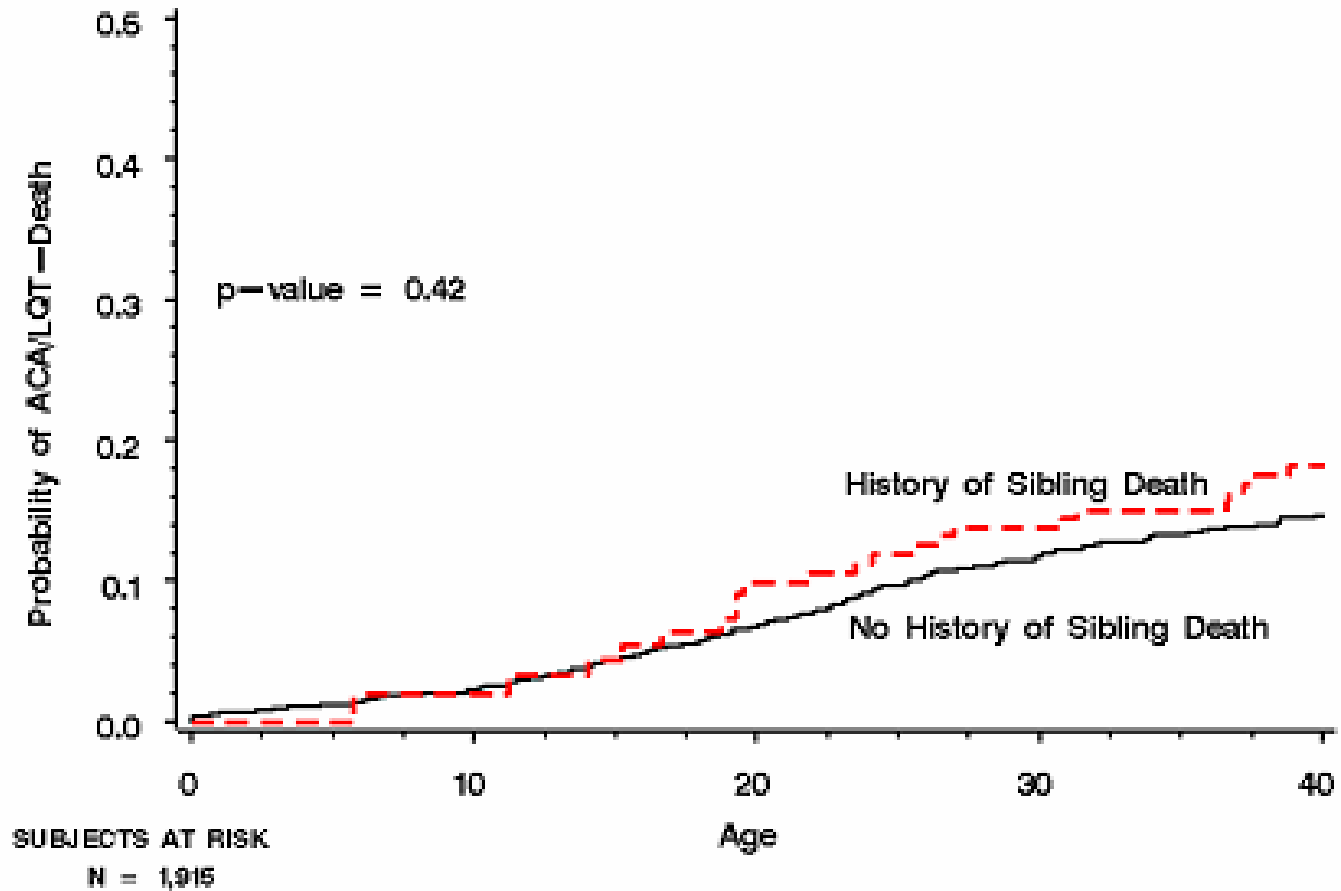
**QUESTION: In an LQTS family in which one member has died suddenly or had ACA at a young age, what is the risk to affected family members, and how should they be treated?**

# **PBS: The Mysterious Heart (2/08)**

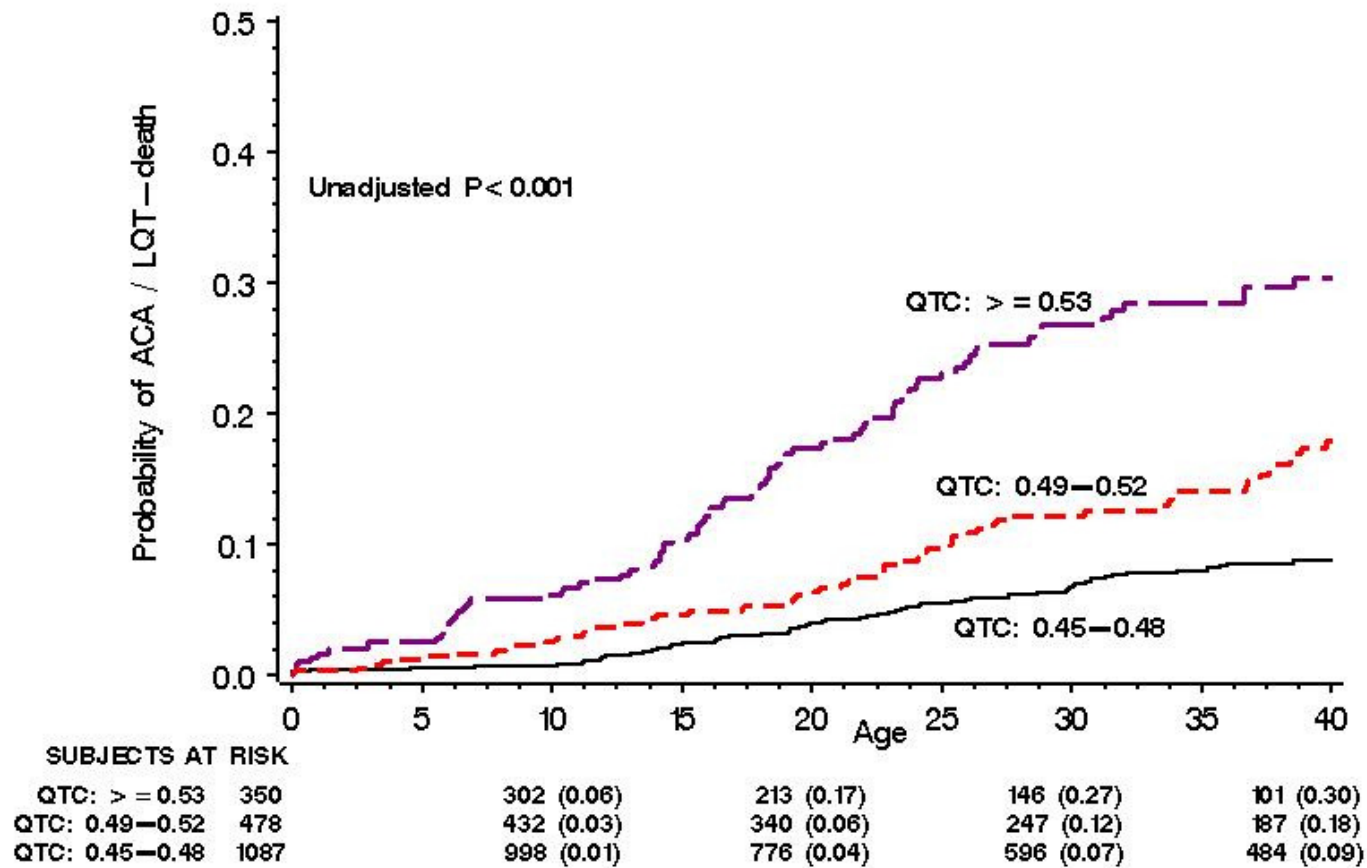
## **ACA in a College Student**

- **19y/o college student with a few episodes of lightheadedness but no syncope arrested in his dorm and resuscitated by roommates; Rx BB+ ICD**
- **Work-up in Rochester revealed LQTS (LQT2), and two younger siblings and mother were affected (QTc ~0.50s), but asymptomatic**
- **Question: Rx family members only with BB, or also add ICD? What would you do?**

# LQTS: Risk of ACA/Death in LQTS Families With and Without a Hx of Sibling Death



# LQTS: Risk of ACA/Death by QTc



Kaufman, Moss, et al. Heart Rhythm 2008

# LQTS: RISK OF ACA/LQTS DEATH IN SURVIVING SIBS AFTER ONE SIB HAS DIED

	<u>Hazard Ratio</u>	<u>P-value</u>
QTc <sub>u</sub> ≥0.53	2.54	<0.01
Syncope <2 yrs	11.26	<0.01
Beta-blocker Rx	0.47	<0.01
ICD Rx	0.13	<0.05
Sib death yes:no	1.14	0.58

**What Did We Do In The Family  
of the College Student?**

# THE CLINICAL CHALLENGE

- In a LQTS family with a SCD, the risk of SCD in surviving affected siblings is related to the presence of LQTS risk factors; in sibs without high-risk factors, the mortality risk is small, but not negligible.
- BBs and life-style changes indicated
- Risk/benefit considerations regarding a “prophylactic” ICD is a judgment call
- Liability issues are a consideration
- Get a 2<sup>nd</sup> opinion from an LQTS expert

# LQTS: CASE REPORT

- **12 y/oM with known LQTS**
- **Hx recurrent syncope – Rx effectively with beta-blockers for several years**
- **Aborted cardiac arrest – June 1998**

# What Was Done

**ICD Implanted.**

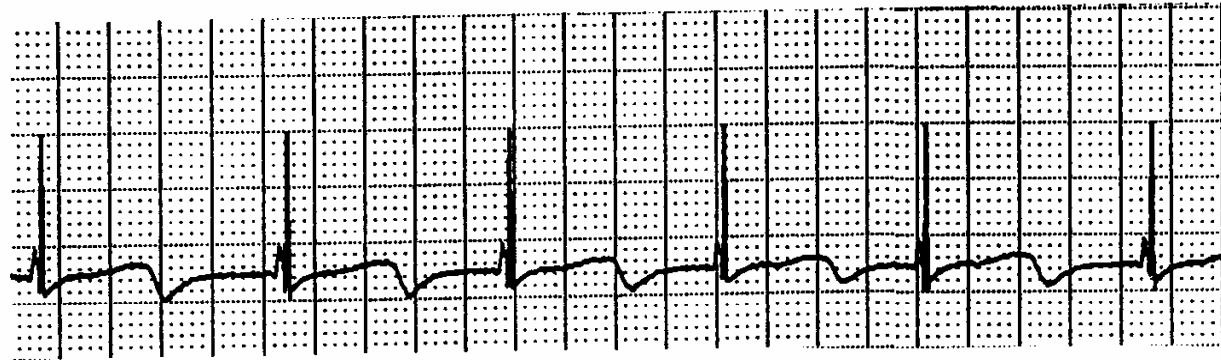
**What was the follow-up?**

## **3 Months Later: 1<sup>st</sup> Day of school**

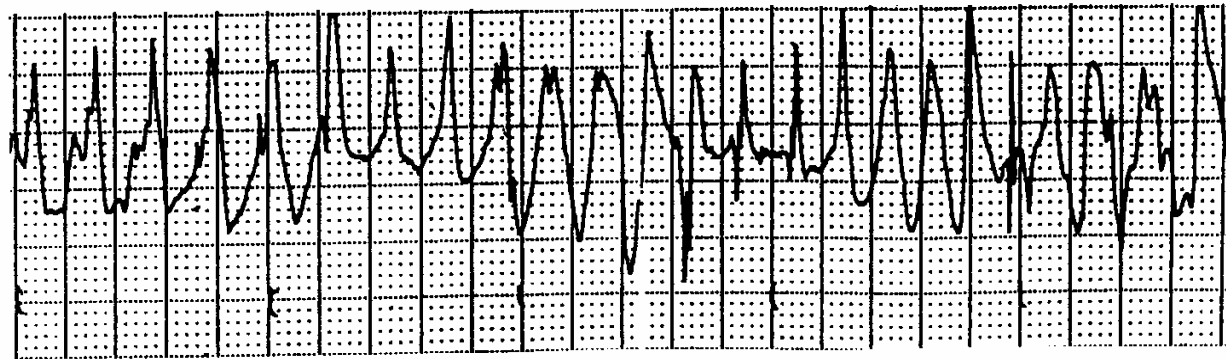
- **Teacher called on student**
- **Student collapsed – abrupt syncope with quick recovery**

# LQTS: ICD

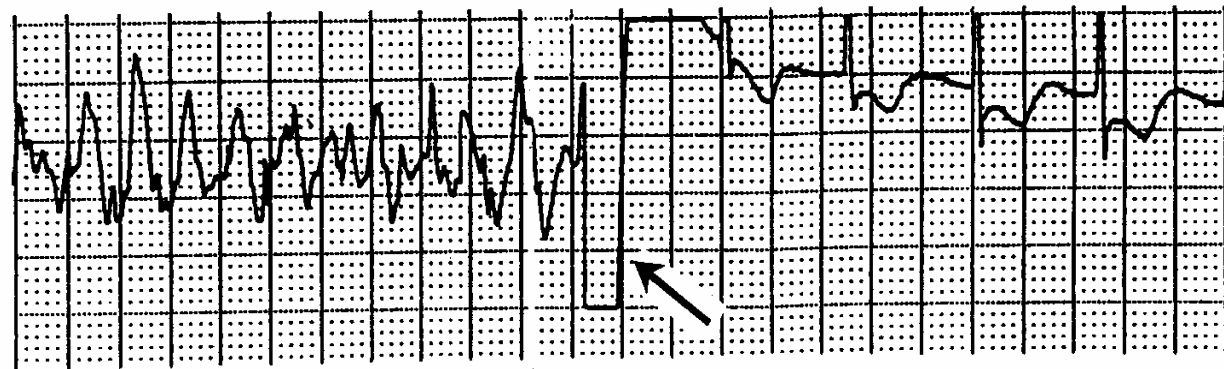
NSR



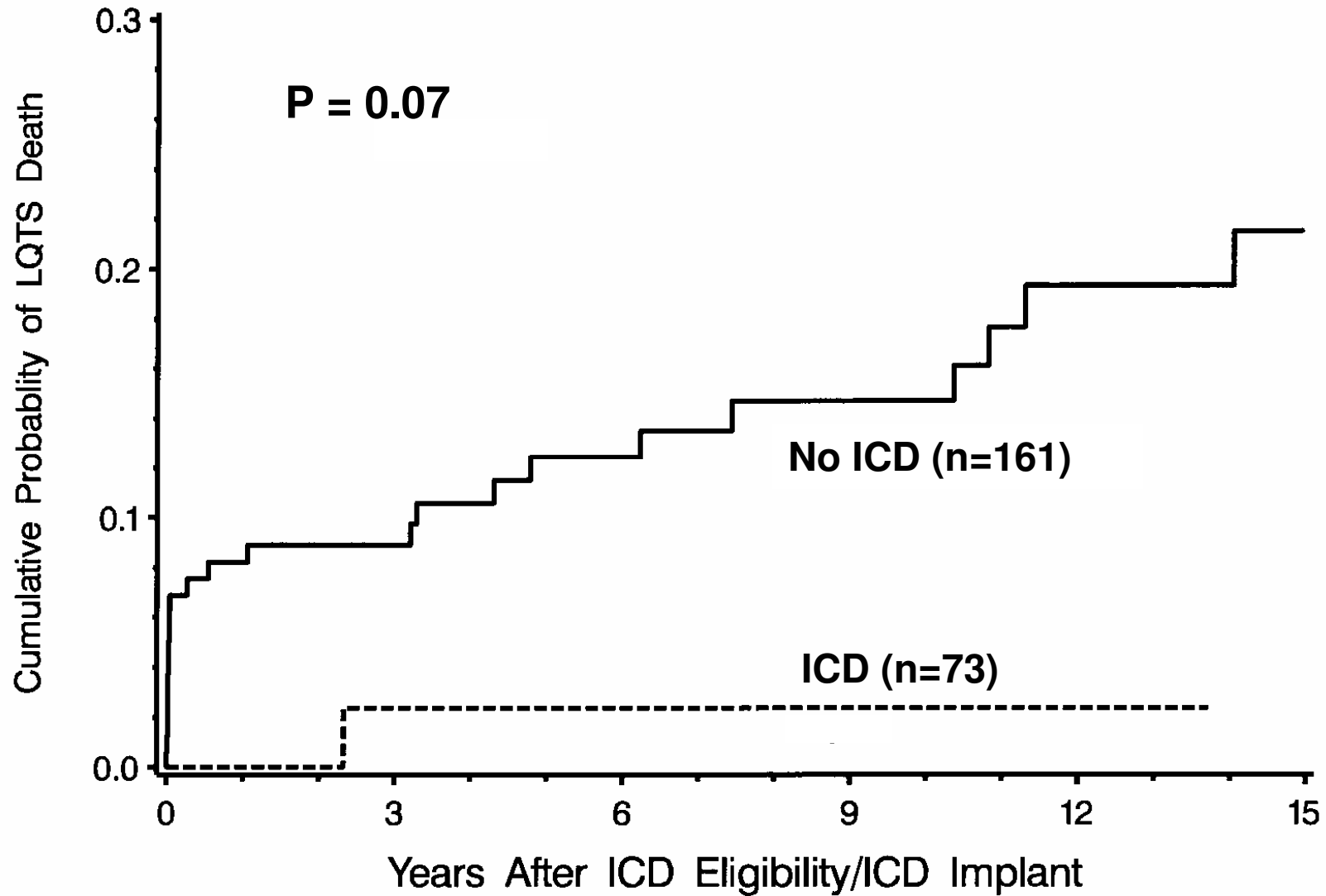
TdP



VF-CD-NSR



# LQTS: ICD Rx



# **ACC/AHA/ESC GUIDELINES (2007): LQTS THERAPEUTIC RECOMMENDATIONS**

## **Class I (good evidence and agreement)**

- 1. Life style modifications**
- 2. Beta-blockers in those with clinical Dx**
- 3. BB + ICD in those with aborted cardiac arrest**

## **Class IIa (weight of evidence favors Rx)**

- 1. BB in those with genetic Dx and normal QT**
- 2. ICD in those with recurrent syncope on BB**

## **Class IIb (weight of evidence less well established)**

- 1. LCTSG in those with recurrent syncope on BB**
- 2. ICD in asymptomatic high-risk patients ( $QTc > 0.53s$ )**