



# ĐIỆN TÂM ĐỒ HỘI CHỨNG RỐI LOẠN NHỊP BẨM SINH

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Vietnam National Heart Institute.

# BIẾN ĐỔI GENE VÀ CÁC LOẠI RỐI LOẠN NHỊP TIM.

- Biến đổi gen và các rối loạn nhịp tim :
  - ✓ H.c Brugada.
  - ✓ H.c QT dài
  - ✓ H.c QT ngắn
  - ✓ Hc Tái cứu sớm.
  - ✓ Loạn sản thất phải (ARVD)
  - ✓ Tim nhanh thất đa hình
  - ✓ H.c W-P-W
  - ✓ Rung nhĩ

**HỘI CHỨNG BRUGADA**

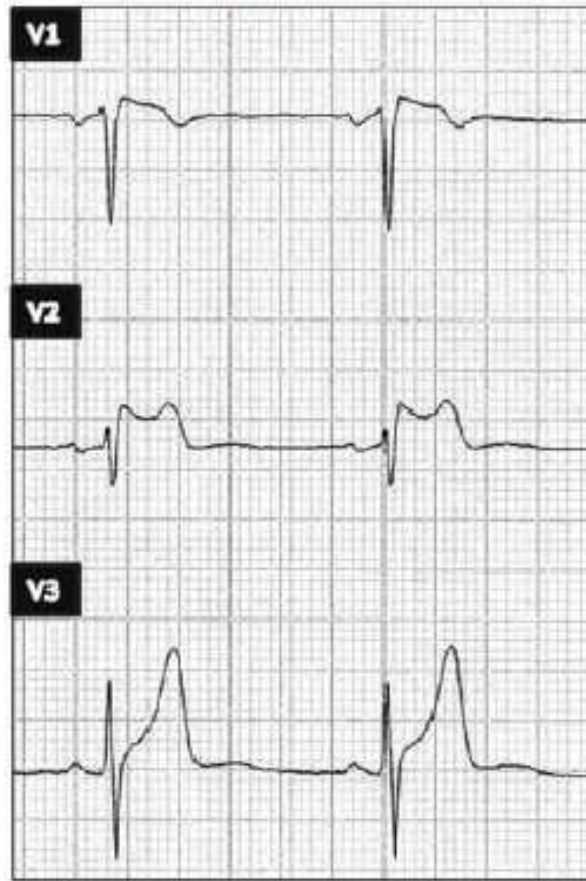
# ĐIỆN TÂM ĐỒ TRONG HỘI CHỨNG BRUGADA

Type I



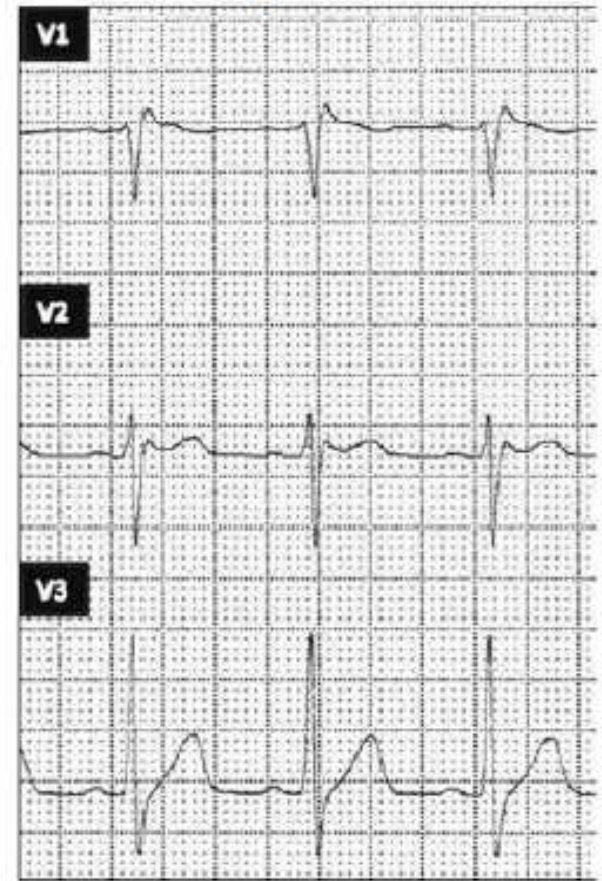
coved ST elevation  
with inverted T

Type II



saddled ST elevation  
 $\geq 1$  mm with upright or  
biphasic T

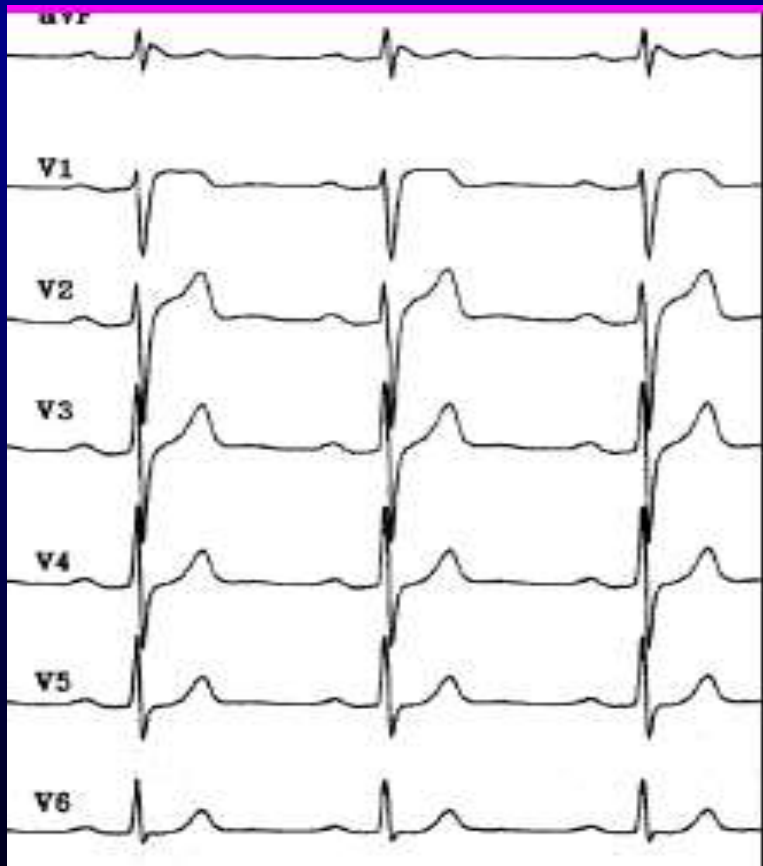
Type III



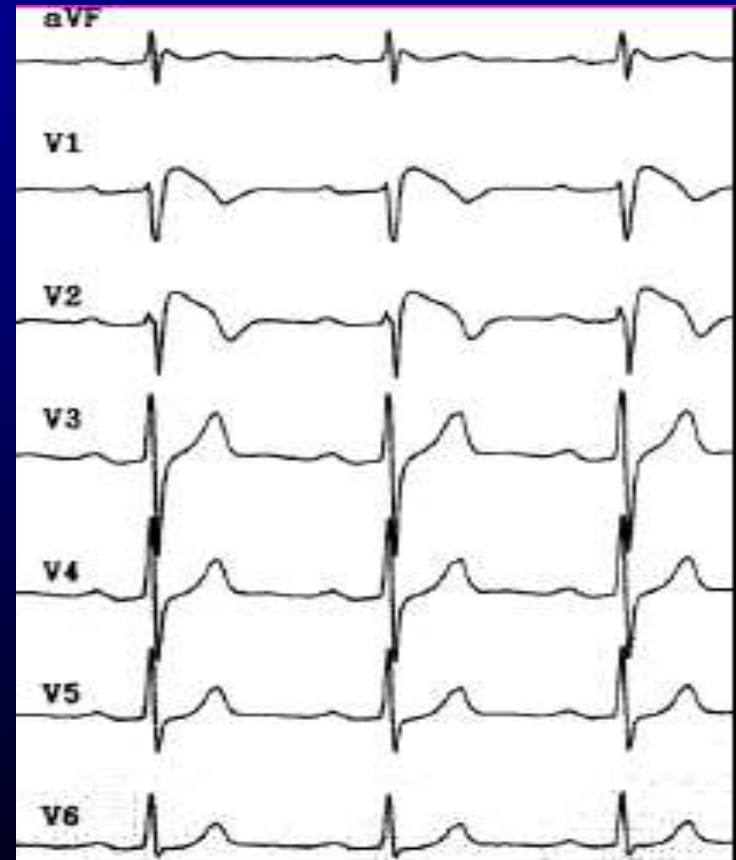
saddled ST elevation  
< 1 mm with upright or  
biphasic T

# VỊ TRÍ ĐẶT ĐIỆN CỰC

Vị trí V1-V3 bình thường



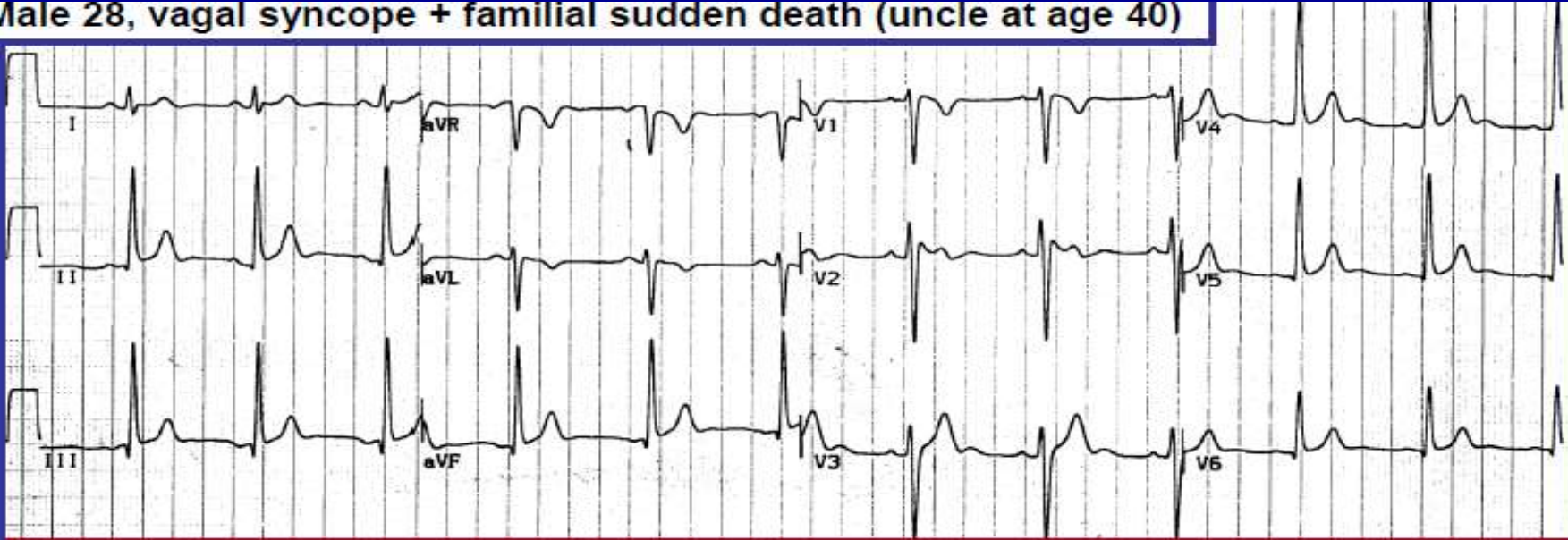
Vị trí V1-V3 cao



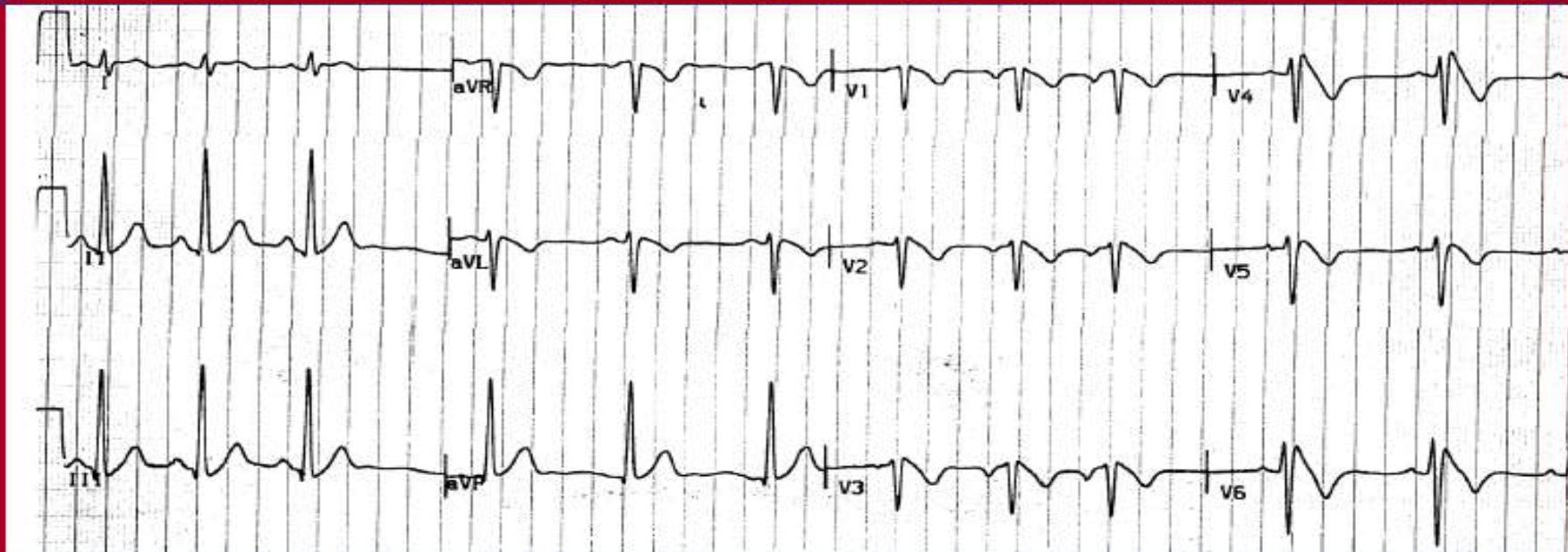
# HỘI CHỨNG BRUGADA

mnTvYkb, Male 28, vagal syncope + familial sudden death (uncle at age 40)

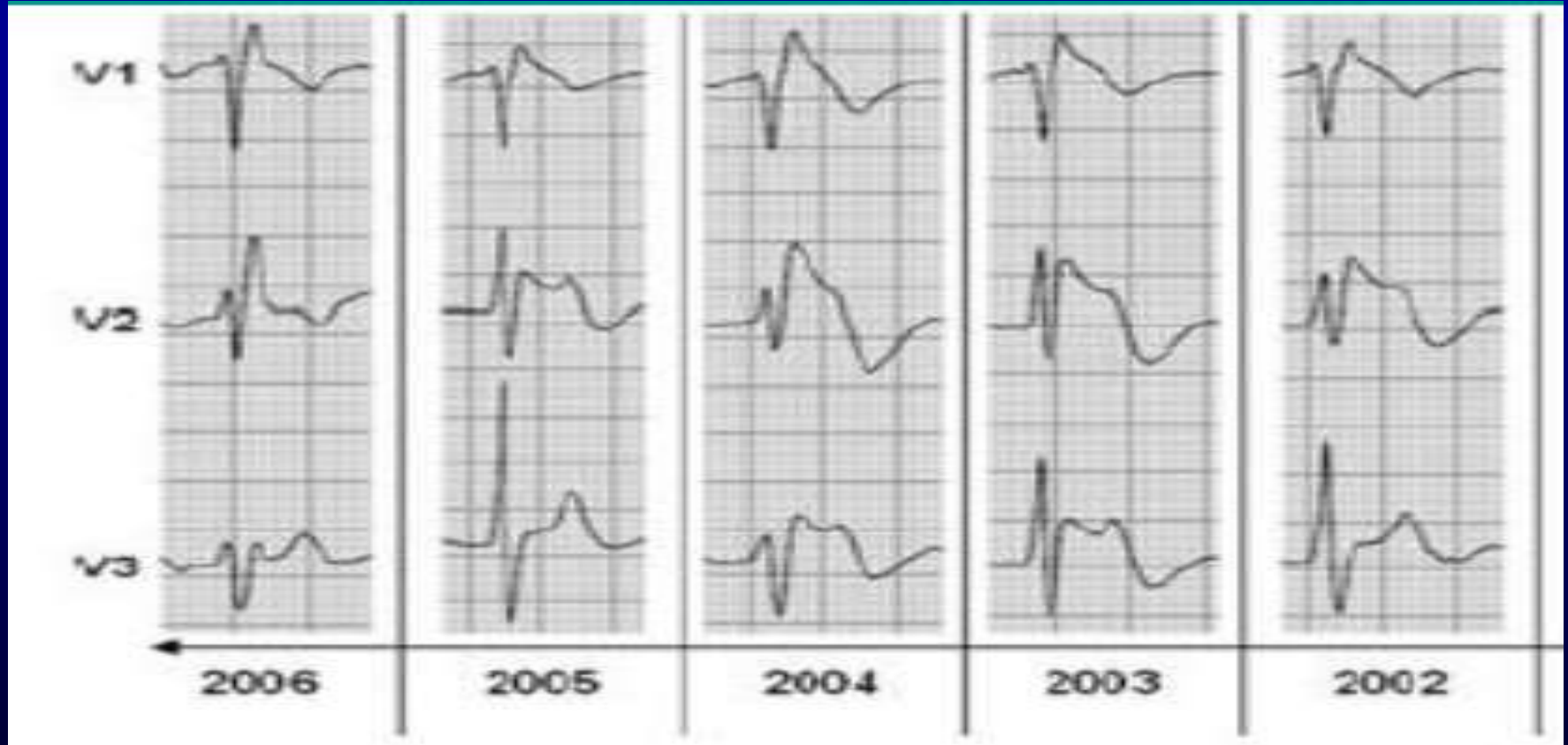
Baseline



V3-V4 = high, V5-V6 = high+  
+ vagal !



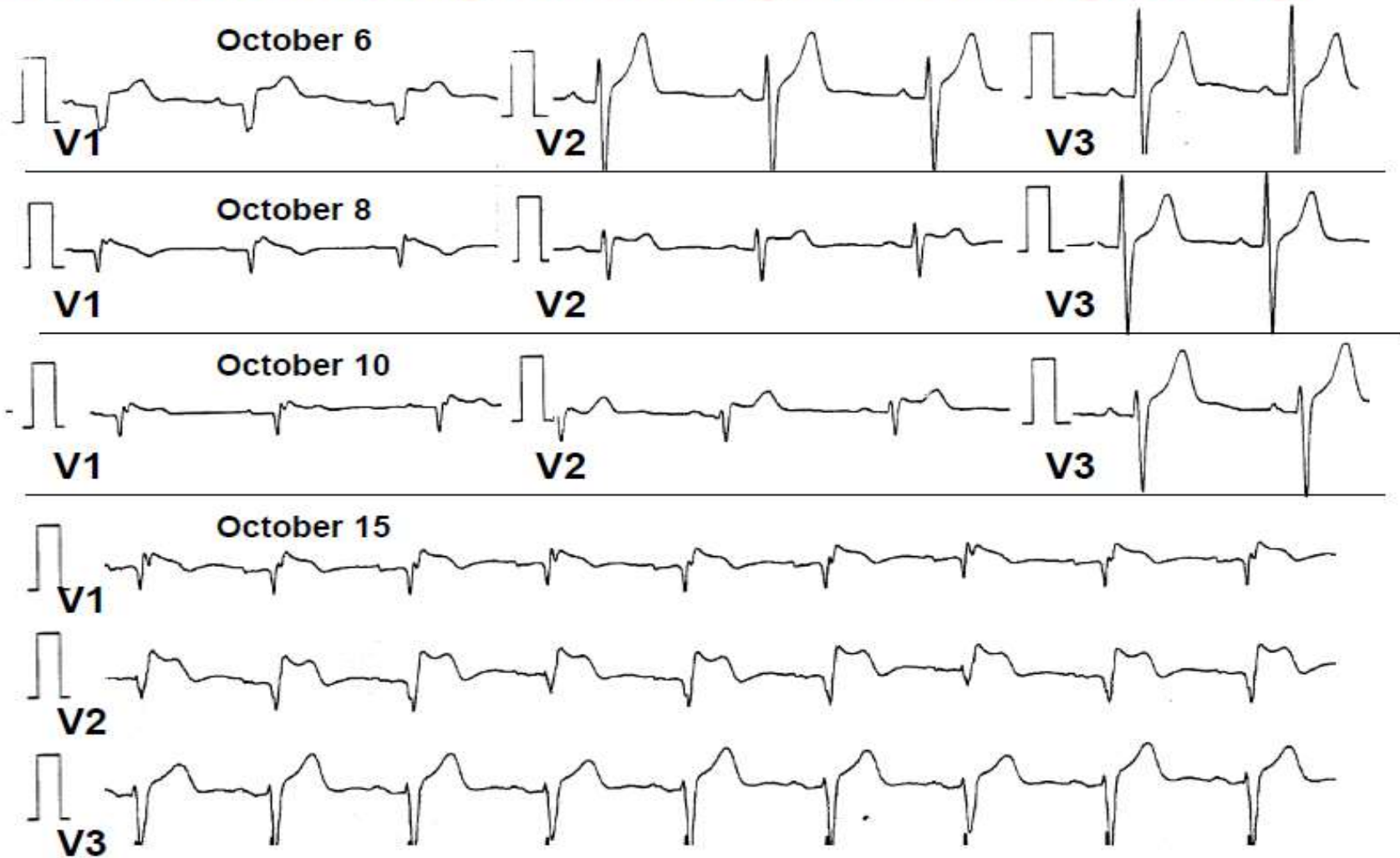
# THAY ĐỔI HÌNH ẢNH ĐTĐ QUA NĂM THÁNG



Richter JCE 2009

# HỘI CHỨNG BRUGAGA

The electrocardiogram changes from day to day





# HỘI CHỨNG BRUGADA

## Inferior and Lateral Electrocardiographic Repolarization Abnormalities in Brugada Syndrome

Andrea Sarkozy, MD; Gian-Battista Chierchia, MD; Gaetano Paparella, MD; Tim. Boussy, MD;  
Carlo De Asmundis, MD; Marcus Roos, MD; Stefan Henkens, RN; Leonard Kaufman, PhD;  
Ronald Buyl, MSc. Ramon Brugada, MD, PhD; Josep Brugada, MD, PhD; Pedro Brugada, MD, PhD



observed in the inferior leads. (*Circ Arrhythmia Electrophysiol.* 2009;2:154-161.)

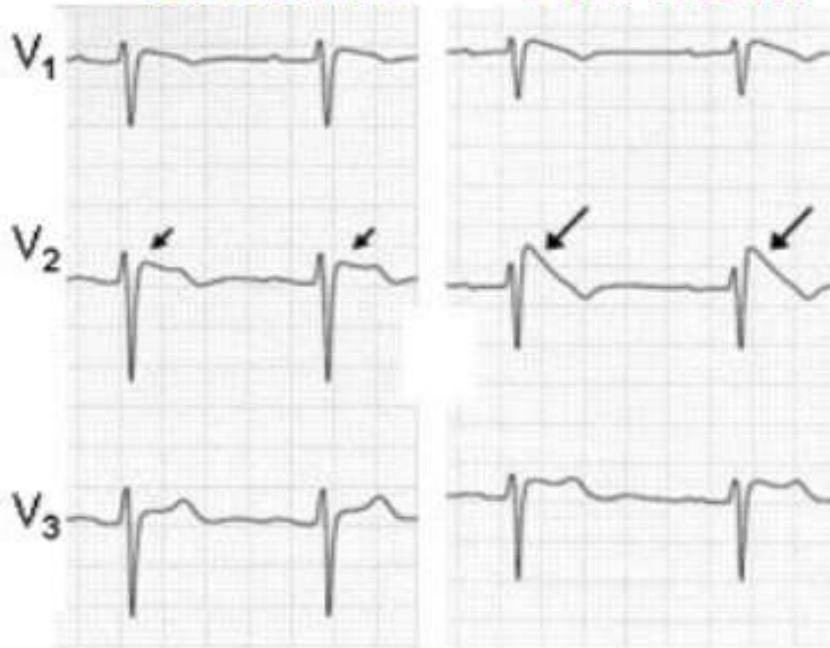
# HỘI CHỨNG BRUGADA

## Effects of a large meal on the ST elevation in patients with Brugada syndrome

Patient 1.

**Baseline**

**Full-meal**



Patient 2.

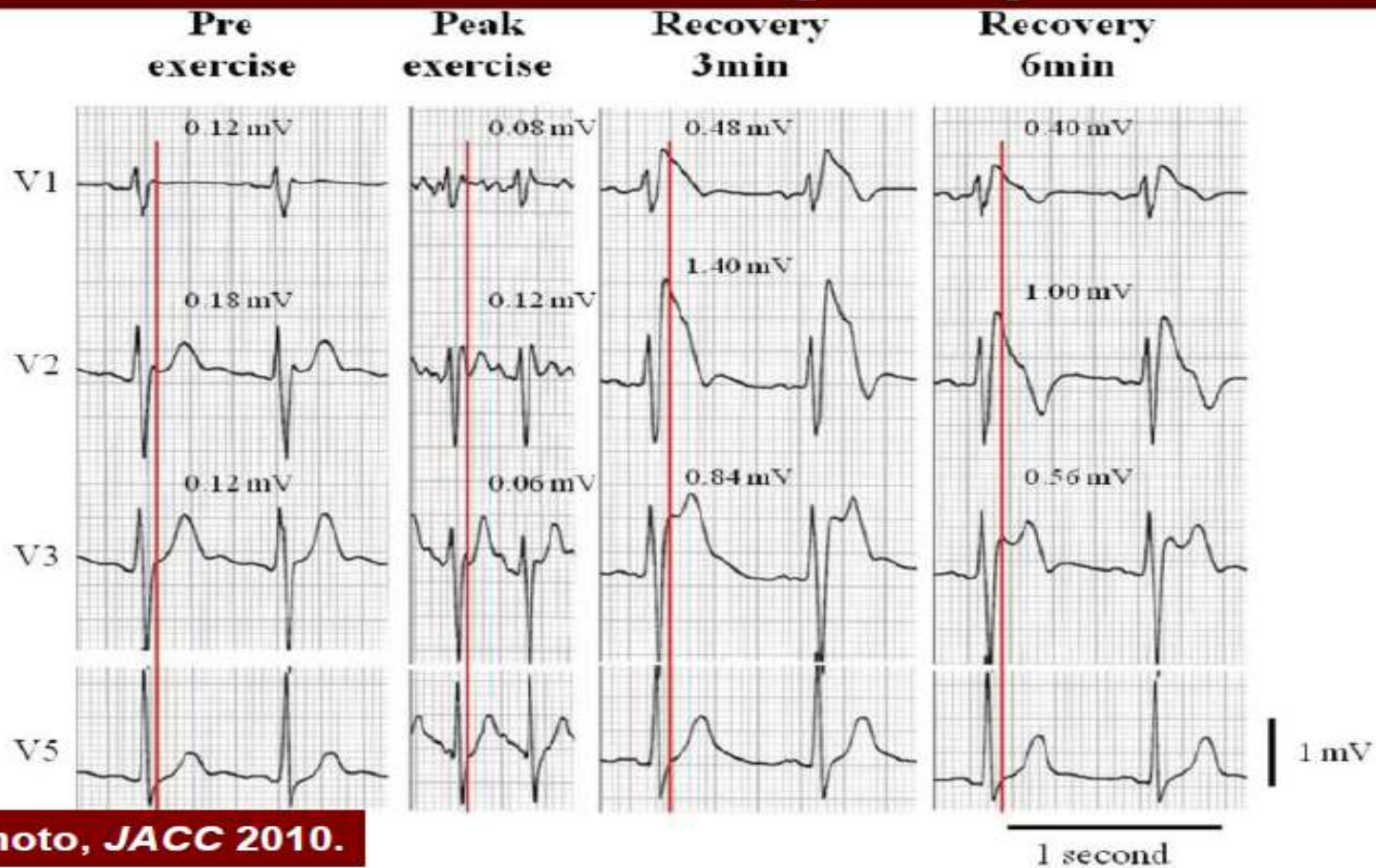
**Baseline**

**Full-meal**



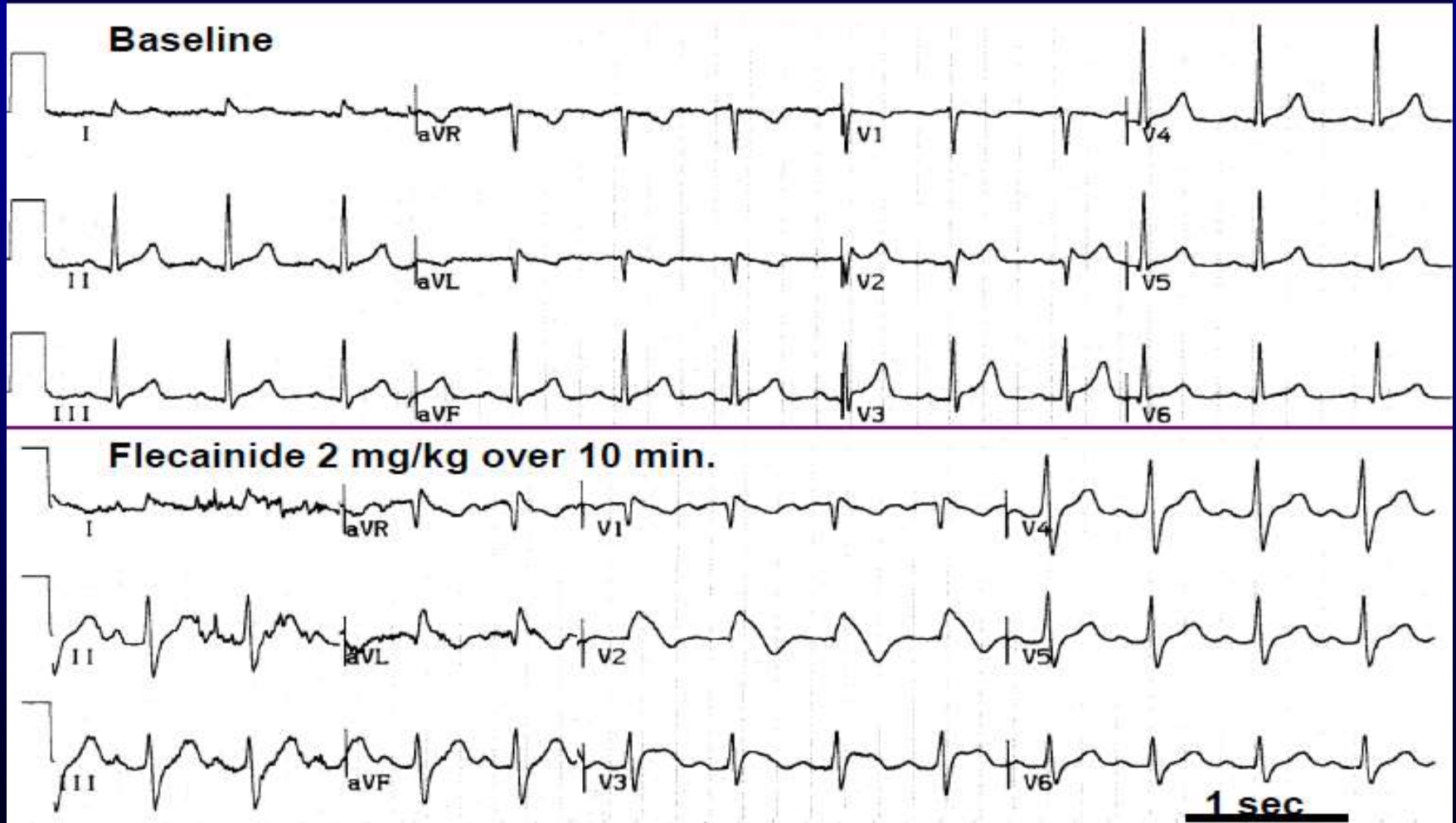
# HỘI CHỨNG BRUGADA

## ST-segment elevation during the recovery phase of exercise test in Brugada syndrome



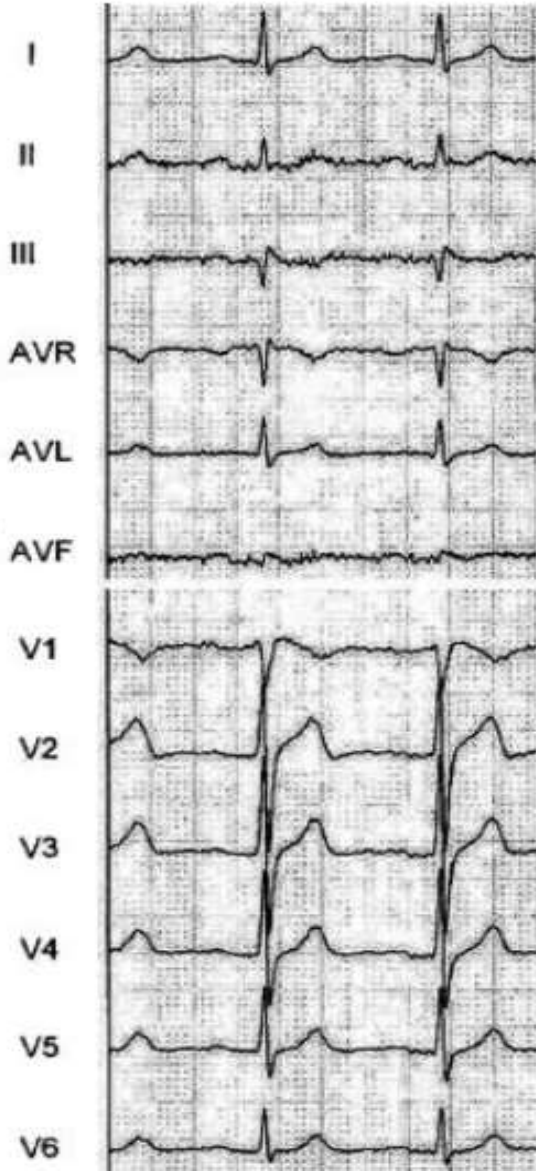
Makimoto, JACC 2010.

# HỘI CHỨNG BRUGADA

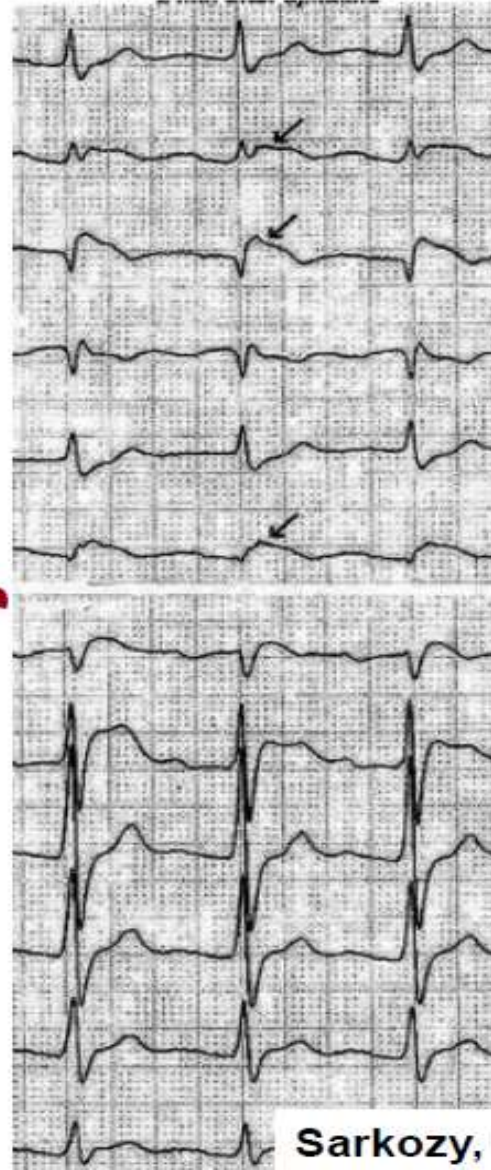


# HỘI CHỨNG BRUGADA

**Before ajmaline**

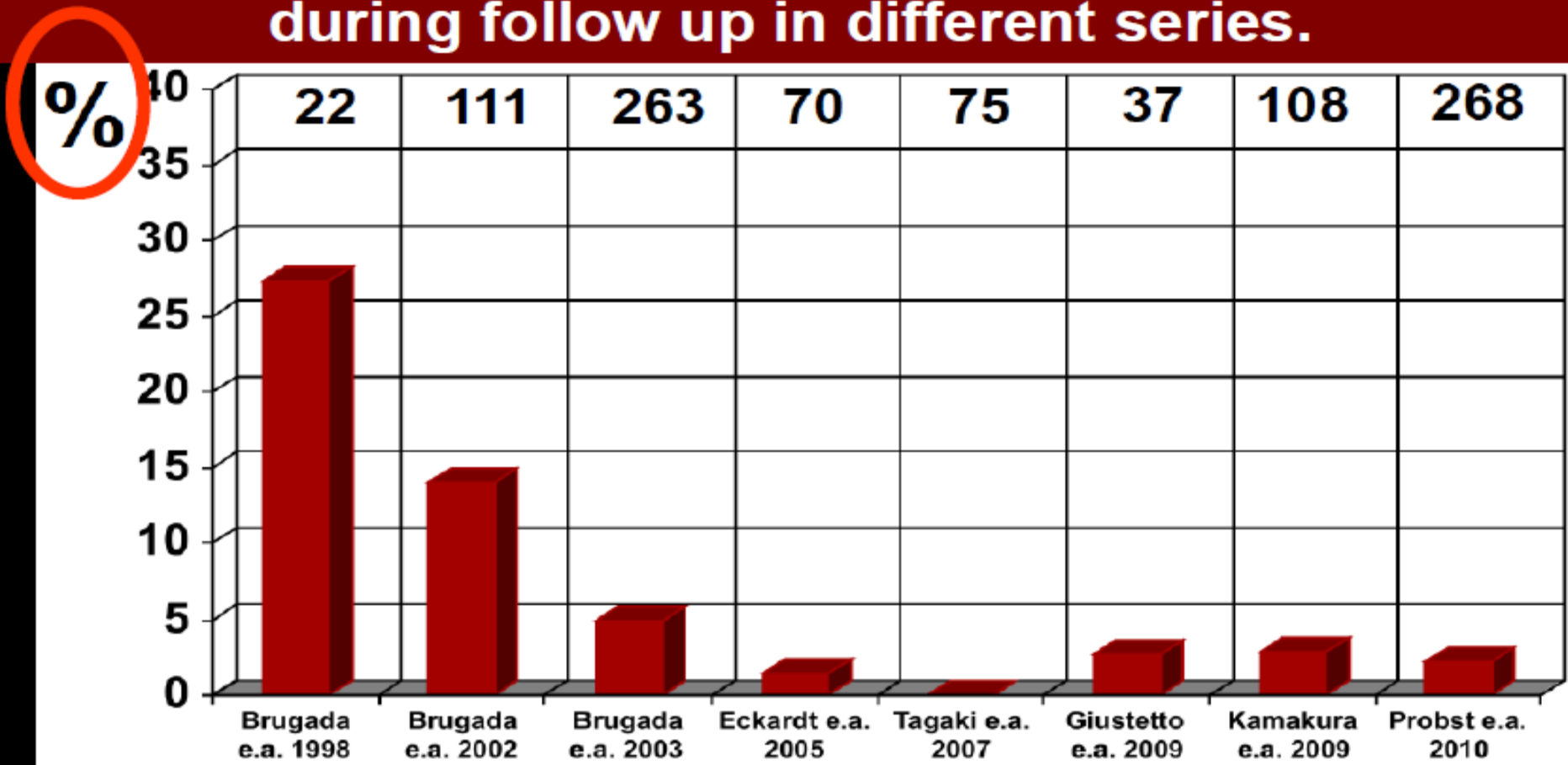


**After ajmaline**



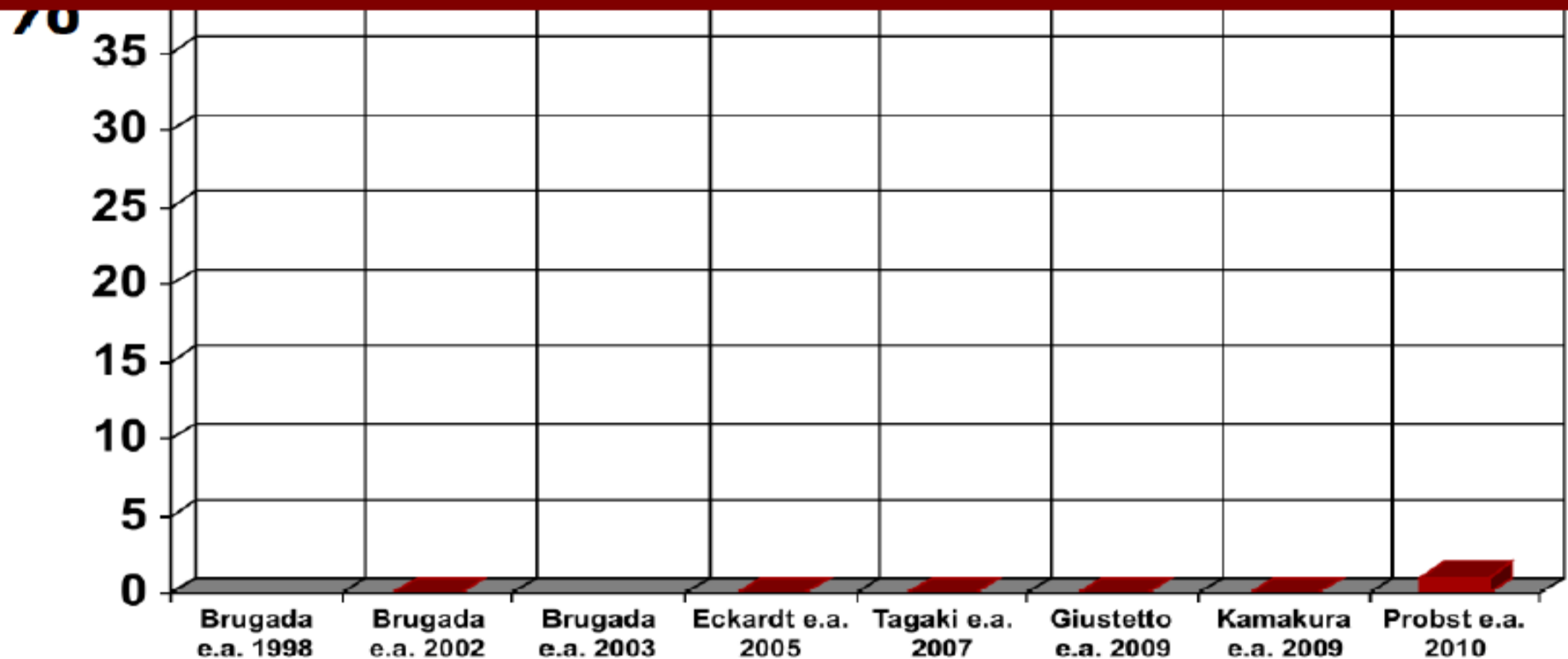
# HỘI CHỨNG BRUGADA

Percentage of patients with initially asymptomatic Brugada syndrome who developed spontaneous VF during follow up in different series.



# HỘI CHỨNG BRUGADA

**Percentage of patients with initially asymptomatic Brugada syndrome (drug-induced Type I) who developed spontaneous VF during follow up in different series.**



## Brugada Syndrome

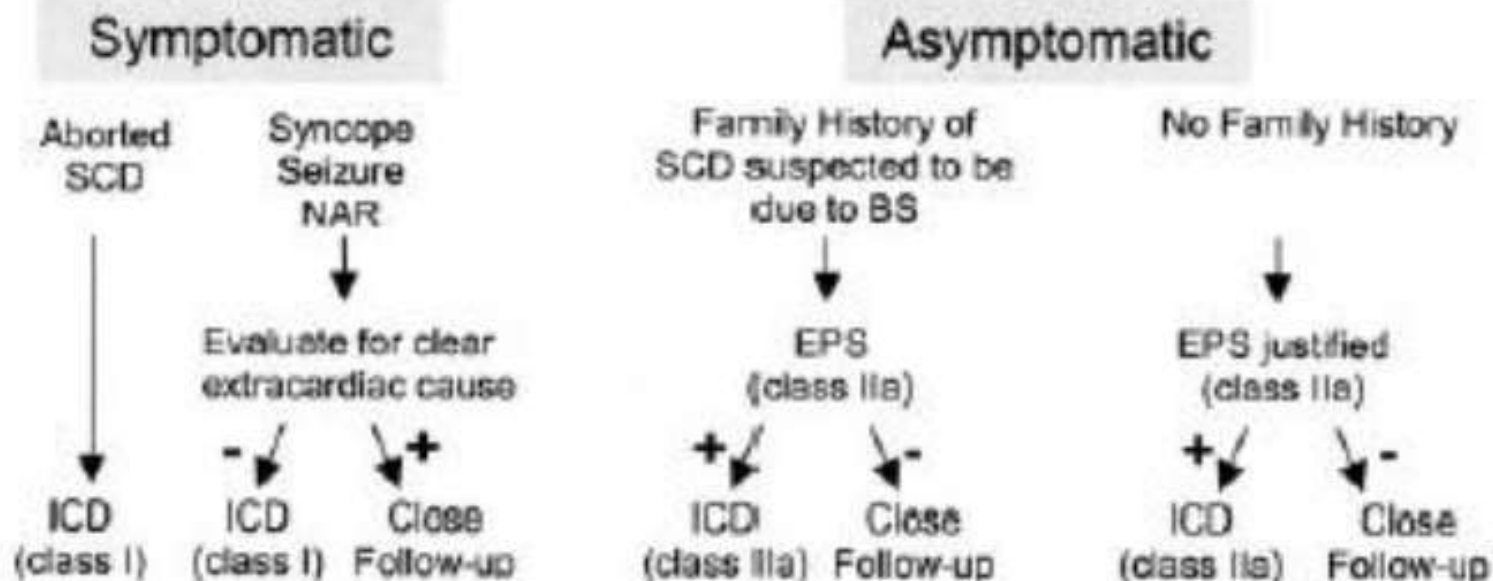
### Report of the Second Consensus Conference

*Endorsed by the Heart Rhythm Society and the European Heart Rhythm Association*

Charles Antzelevitch, PhD; Pedro Brugada, MD, PhD; Martin Borggrefe, MD, PhD; Josep Brugada, MD; Ramon Brugada, MD; Domenico Corrado, MD, PhD; Ihor Gussak, MD, PhD; Herve LeMarec, MD; Koonlawee Nademanee, MD; Andres Ricardo Perez Riera, MD; Wataru Shimizu, MD, PhD; Eric Schulze-Bahr, MD; Hanno Tan, MD, PhD; Arthur Wilde, MD, PhD

**Abstract**—Since the discovery of Brugada syndrome as a genetic disease to one of the major causes of sudden cardiac death (SCD) associated with a specific ECG pattern in young adults and children. The syndrome is primarily a primary disorder, but can be secondary to various conditions, including congenitally concealed and acquired long QT syndrome, catecholaminergic polymorphic  $\alpha$ -adrenergic stimulation, and electrolyte abnormalities such as hypokalemia, hypomagnesemia, and hypocalcemia. In 2003, an international consensus conference on diagnostic criteria and management of Brugada syndrome was held in Brno, Czech Republic. This report summarizes the consensus on diagnostic and management criteria for Brugada syndrome. (Circulation. 2005;111:659-670.)

### Spontaneous Type 1 ECG





# Amid the fourth lustrum after the description of Brugada syndrome: controversies over?

Pedro Brugada\*

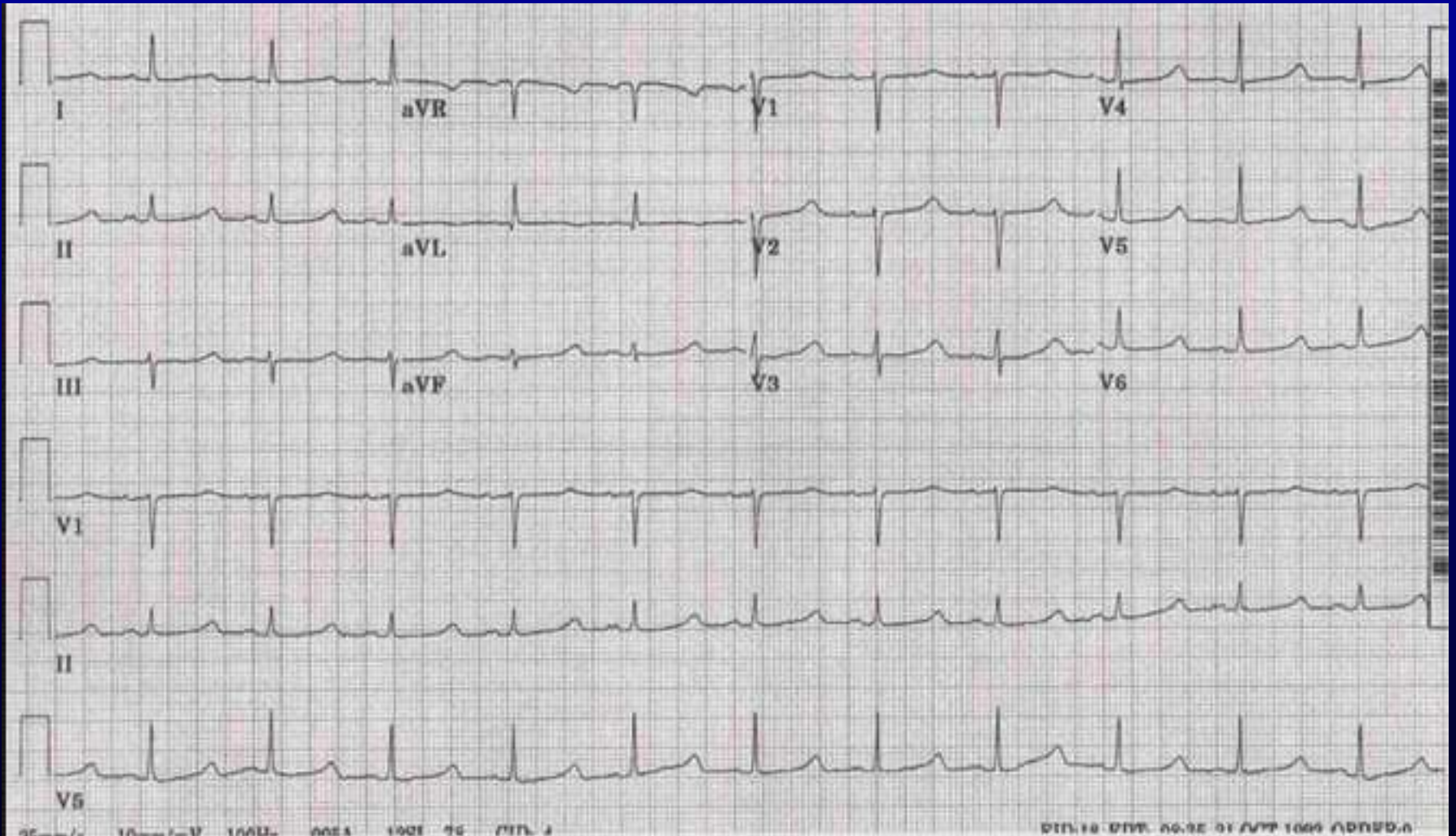
The stratification scheme is now more valid than it ever was.

**Table 1** Risk stratification of patients with a Brugada electrocardiogram

Clinical presentation	Results EP	Decision
Aborted sudden death or syncope	Not needed	High risk, ICD
Asymptomatic with spontaneous Brugada ECG	Inducible Not inducible	High risk, ICD Low risk, follow-up
Asymptomatic with Brugada ECG only after drugs	Not needed	Low risk, follow-up

HỘI CHỨNG QT DÀI

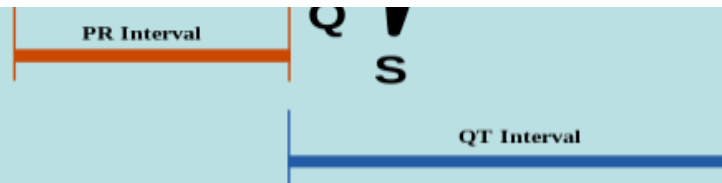
# HỘI CHỨNG QT DÀI



# ĐO KHOẢNG QT NH THẾ NÀO?

In clinical practice, the QT interval is corrected for the heart rate by the Bazett formula:

$$\text{Corrected QT (QTc)} = \left[ \text{QT interval} / \text{square root of the RR interval} \right]$$



# KHOẢNG QTc ?

<http://biostat.mc.vanderbilt.edu/ECGPredictionInterval>

Age (years)	<input type="text" value="57"/>	
Height	<input type="text" value="1.8"/>	<input type="radio"/> Inches <input checked="" type="radio"/> Meters
Weight	<input type="text"/>	<input type="radio"/> Lbs <input checked="" type="radio"/> Kg
BMI (kg/m <sup>2</sup> )	<input type="text" value="25"/>	
Gender	<input type="radio"/> Female <input checked="" type="radio"/> Male	
Race	<input type="text" value="White"/>	
History of Type II Diabetes	<input checked="" type="radio"/> No <input type="radio"/> Yes	
Heart Rate	<input type="text" value="70"/>	<input checked="" type="radio"/> Unknown The heart rate is based on <input type="radio"/> Known calculations
<input type="button" value="CALCULATE"/>		

## Predicted Value [95% Normal Region]

PR Interval (msec)	160 [127, 193]
QRS Duration (msec)	88 [73, 104]
Heart Rate (bpm)	70 [48, 91]
QTc Interval (msec) (Bazett correction <sup>1</sup> )	404 [359, 448]

# TIÊU CHUẨN CHẨN ĐOÁN QT DÀI

Khả năng mắc LQTS:

$\leq 1$  : thấp.

2-3: nguy cơ trung bình.

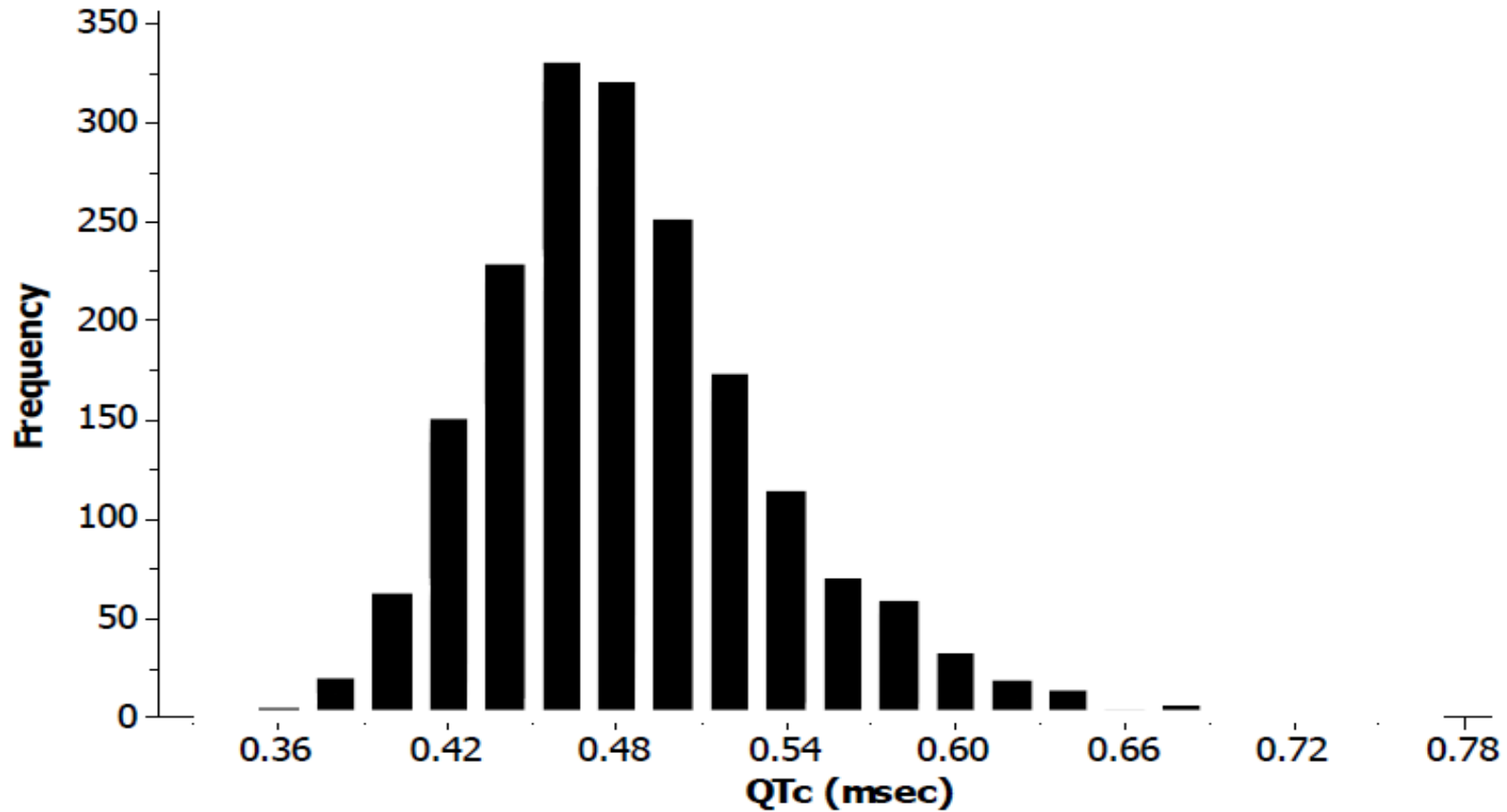
$\geq 4$ : nguy cơ cao

Schwartz PJ, Circulation 1993

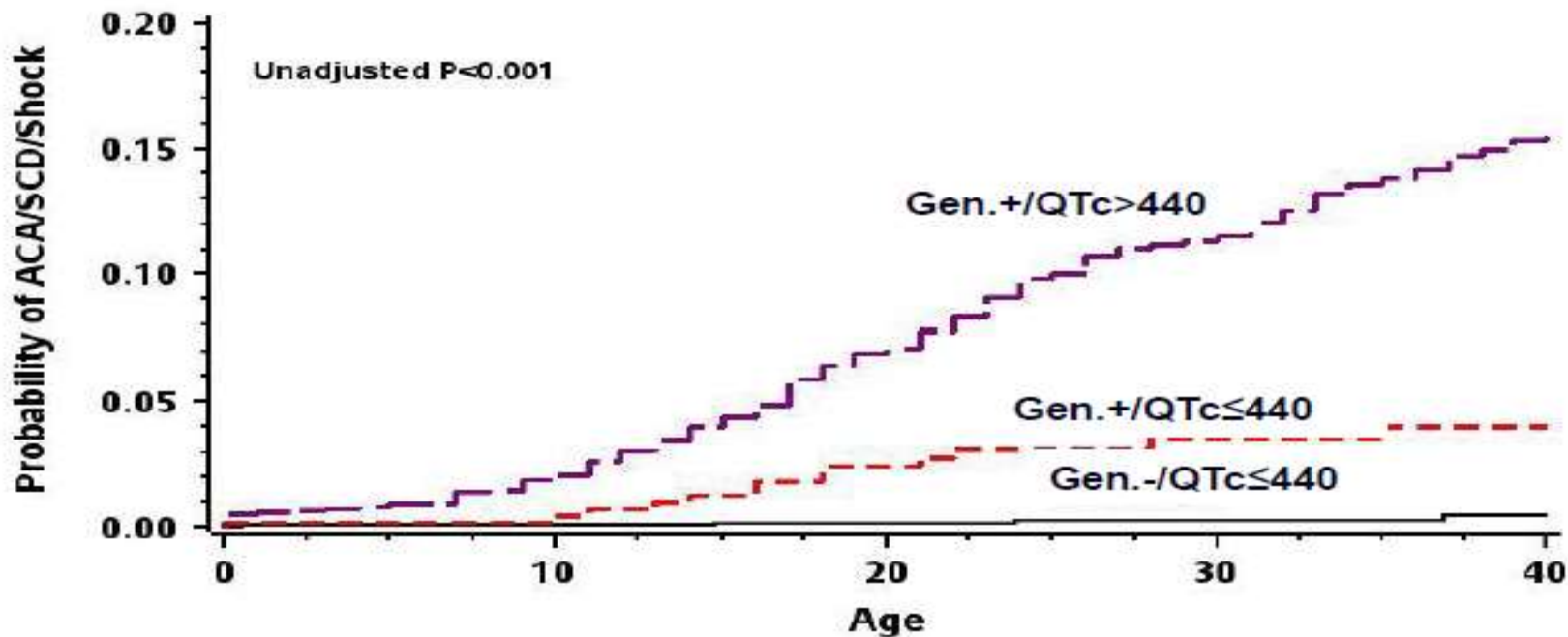
Finding	Score
Electrocardiographic†	
Corrected QT-Interval, ms	
$\geq 480$	3
460-470	2
450 (in males)	1
Torsades de pointes‡	2
T-wave alternans	1
Notched T-wave in 3 leads	1
Low heart rate for age§	0.5
Clinical history	
Syncope‡	
With stress	2
Without stress	1
Congenital deafness	0.5
Family history	
Family members with definite LQTS	1
Unexplained SCD in immediate family members <30 yrs old	0.5

# HỘI CHỨNG QT DÀI

**Distribution of QTc Duration in Genotype-Positive Patients**



# KHẢ NĂNG GÂY ĐỘT TỬ HỘI CHỨNG QT DÀI



## Patients at Risk

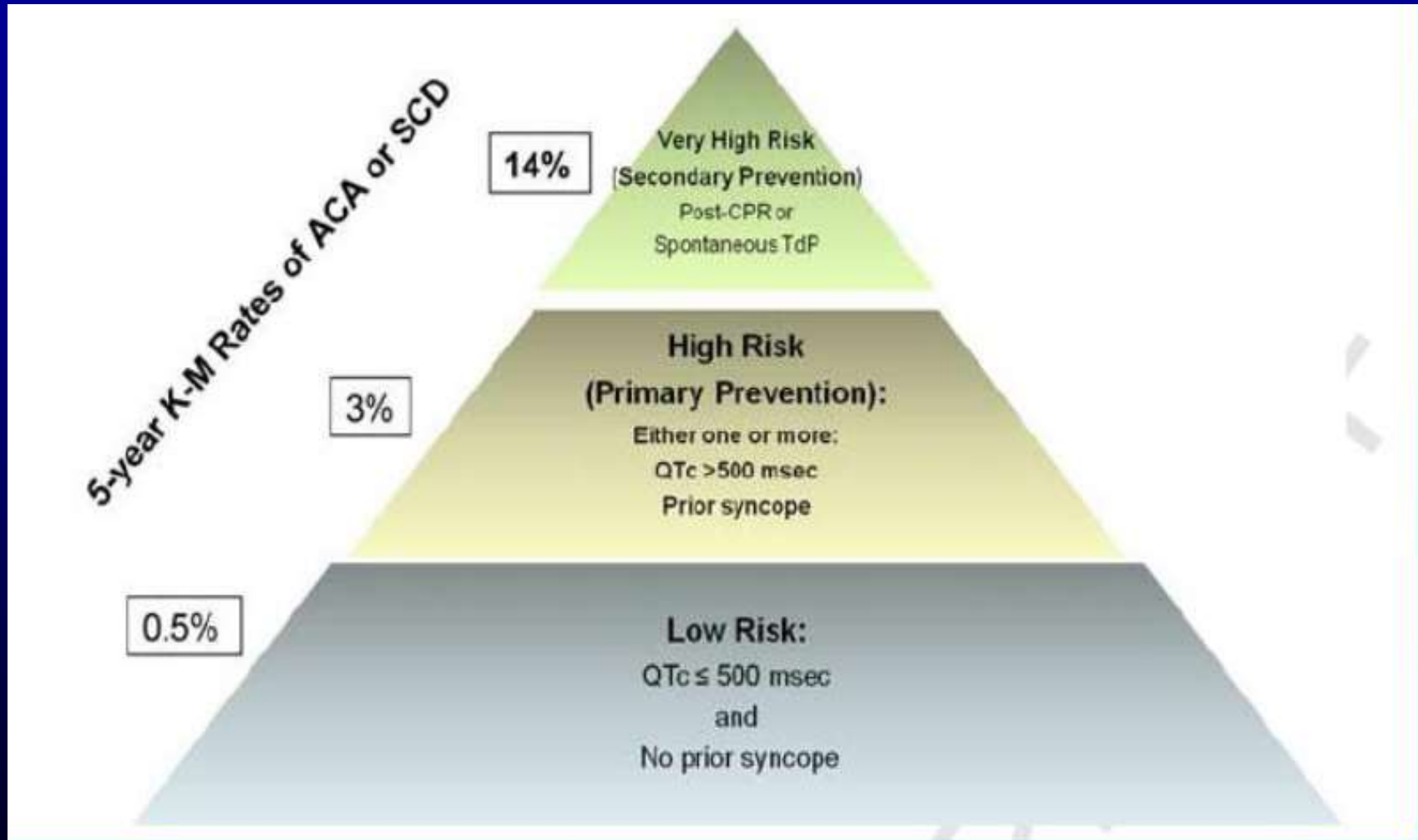
	0	10	20	30	40
Gen-/Phen-	1525	1377 (0)	996 (0)	728 (0)	547 (0)
Gen+/Phen-	469	406 (0)	294 (0.02)	236 (0.04)	177 (0.04)
Gen+/Phen+	1392	1235 (0.02)	945 (0.07)	729 (0.11)	543 (0.15)



# NGUY CƠ ĐỘT TỬ HỘI CHỨNG QT DÀI

Age Group (Ref. #)	Risk Factor	Hazard Ratio (p Value)	Beta-Blocker Efficacy, % Reduction (p Value)
Childhood (1-12 yrs) (33)	Male gender	3.96 (<0.001)	73% (0.002)
	QTc > 500 ms	2.12 (0.02)	
	Prior syncope:		
	Recent (<2 yrs)	14.34 (<0.001)	
	Remote (≥2 yrs)	6.45 (<0.001)	
Adolescence (10-20 yrs) (28)	QTc > 530 ms	2.3 (<0.001)	64% (0.01)
	Syncope		
	≥2 syncopal events in past 2 yrs	18.1 (<0.001)	
	1 syncopal event in past 2 yrs	11.7 (<0.001)	
	≥2 syncopal events in past 2-10 yrs	5.8 (<0.001)	
	1 syncopal events in past 2-10 yrs	2.7 (<0.001)	
Adulthood (18-40 yrs) (29)	Female gender	2.68 (<0.05)	60% (<0.01)
	QTc duration		
	QTc ≥500 ms	6.35 (<0.01)	
	QTc 500-549 ms	3.34 (<0.01)	
	Prior syncope	5.10 (<0.01)	
Adulthood (41-60 yrs) (53)†	Recent syncope (<2 yrs)	9.92 (<0.001)	42% (0.40)‡
	QTc > 530 ms	1.68 (0,06)	
	LQT3 genotype	4.76 (0.02)	

# ĐÁNH GIÁ NGUY CƠ ĐỘT TỬ HỘI CHỨNG QT DÀI



HỘI CHỨNG QT NGẮN

## Idiopathic Short QT Interval: A New Clinical Syndrome?

Ihor Gussak<sup>a</sup> Pedro Brugada<sup>b</sup> Josep Brugada<sup>c</sup> R. Scott Wright<sup>a</sup>  
Stephen L. Kopecky<sup>a</sup> Bernard R. Chaitman<sup>d</sup> Preben Bjerregaard<sup>d</sup>

<sup>a</sup>Mayo Physician Alliance for Clinical Trials, Mayo Clinic and Mayo Foundation, Rochester, Minn., USA  
<sup>b</sup>Cardiovascular F, Barcelona, Spain,  
<sup>c</sup>Cardiovascular F, Barcelona, Spain,  
<sup>d</sup>Cardiovascular F, Copenhagen, Denmark

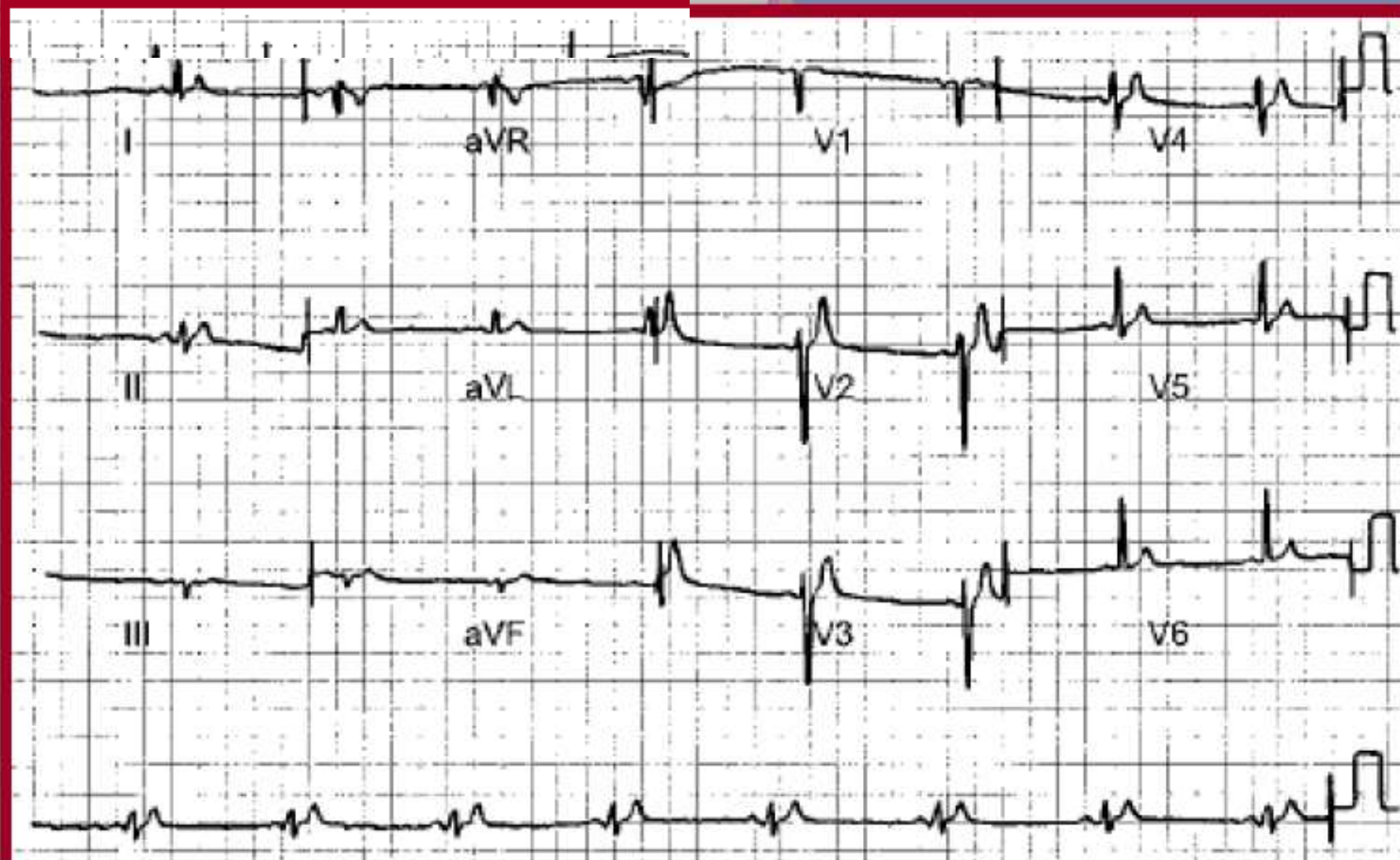


TABLE 1

**Diagnostic criteria for short QT syndrome**

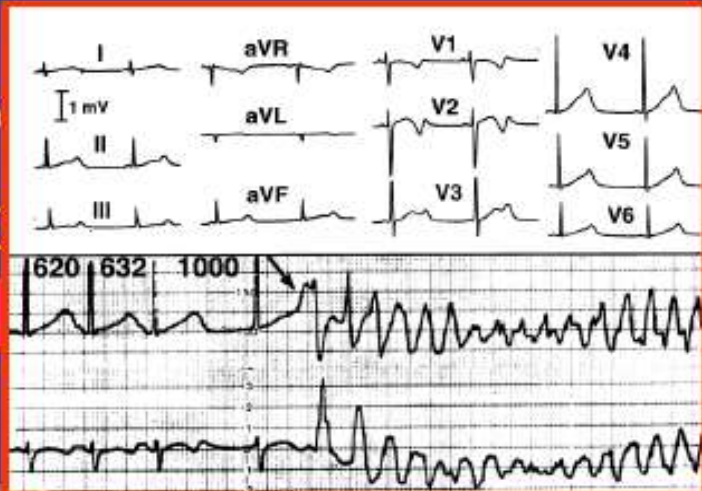
CRITERIA	POINTS
<b>Corrected QT interval <sup>a</sup></b>	
< 370 ms	1
< 350 ms	2
< 330 ms	3
<b>Interval from J point to T peak &lt; 120 ms <sup>a,b</sup></b>	1
<b>Clinical history <sup>c</sup></b>	
History of sudden cardiac arrest	2
Documented polymorphic ventricular tachycardia or ventricular fibrillation	2
Unexplained syncope	1
Atrial fibrillation	1
<b>Family history <sup>d</sup></b>	
First- or second-degree relative with high probability of short QT syndrome	2
First- or second-degree relative with autopsy-negative sudden cardiac death	1
Sudden infant death syndrome	1
<b>Genotype</b>	
Genotype-positive	2
Mutation of undetermined significance in a culprit gene	1
<b>TOTAL</b>	_____
<b>Probability of short QT syndrome</b>	
High	4 points
Intermediate	3 points
Low	2 points

# HỘI CHỨNG QT NGẮN

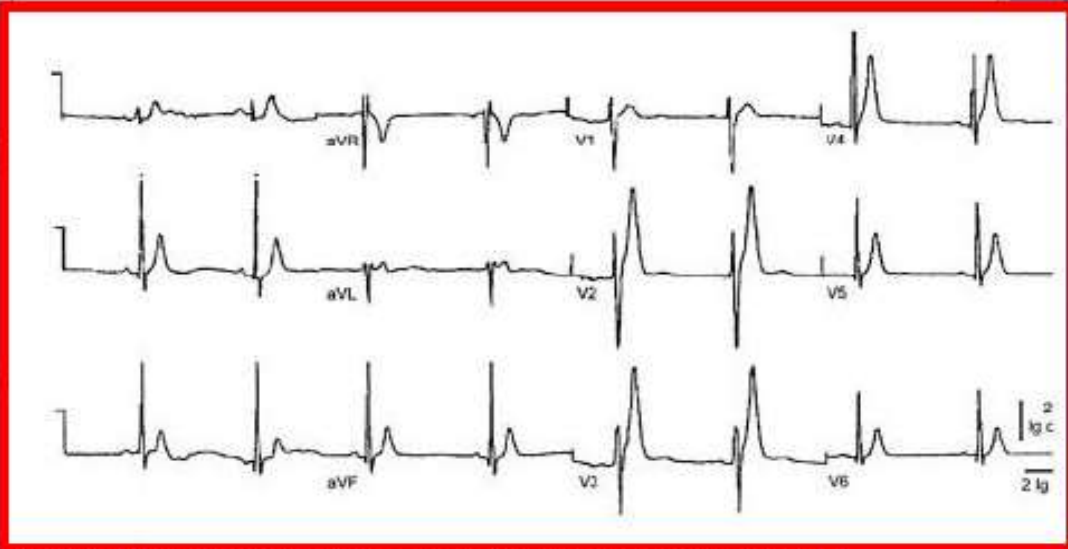
## Sudden Death Associated With Short-QT Syndrome Linked to Mutations in HERG

Ramon Brugada, MD\*, Kai Hong, MD, PhD\*, Robert Dumaine, PhD; Jonathan Cordeiro, PhD;  
 Fiorenzo Gaita, MD; Martin Borggrefe, MD; Teresa M. Menendez, MD; Josep Brugada, MD, PhD;  
 Guido D. Pollevick, PhD; Christian Wolpert, MD; Elena Burashnikov, MS;  
 Kiyotaka Matsuo, MD, PhD; Yue Sheng Wu, MD; Alejandra Gnerchicoff, PhD;  
 Francesca Bianchi, MD; Carla Giustetto, MD; Rainer Schimpf, MD;  
 Pedro Brugada, MD, PhD; Charles Antzelevitch, PhD

## LQT2 = Loss of function of HERG



## SQT1 = Gain of function of HERG



# HỘI CHỨNG QT NGẮN

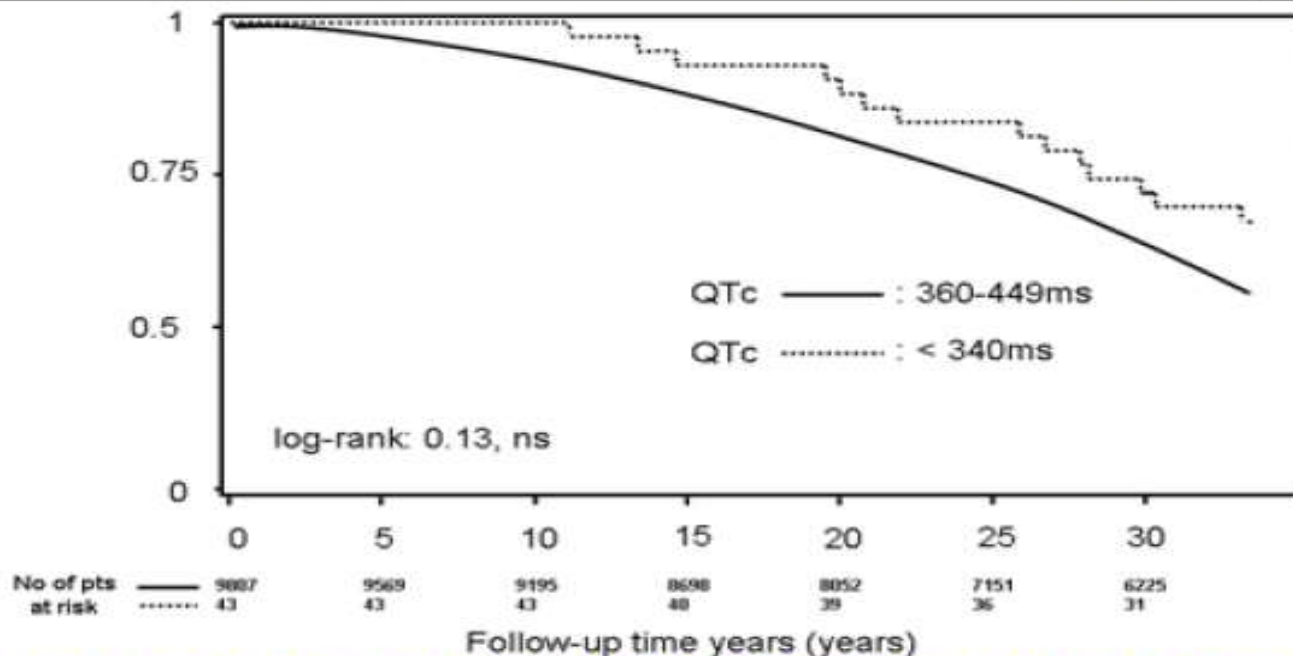
Based on 10,822 ECGs (Age 44 ± 8 years):

95% CI for QTc <320 is <0.2%

**4 in 1000 healthy men have QTc <340 msec**

**2% of men had QTc 340-360 msec**

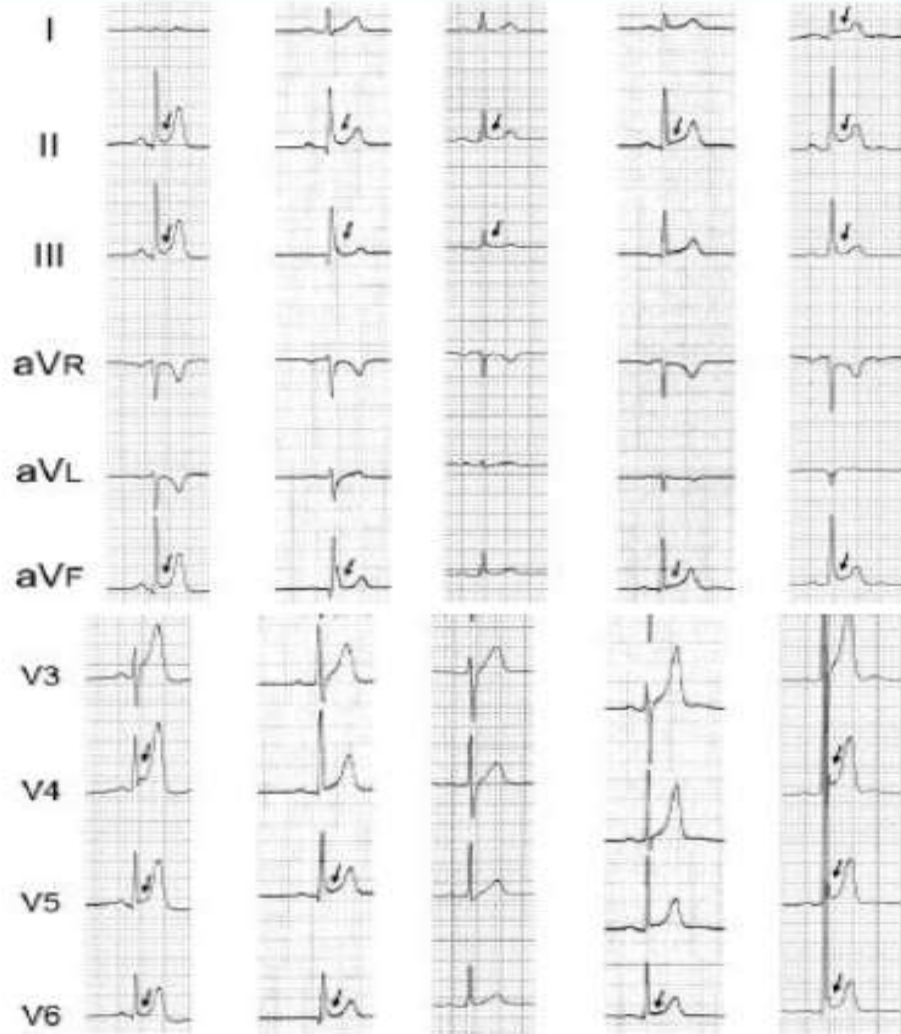
Anttonen, *Circulation* 2007.



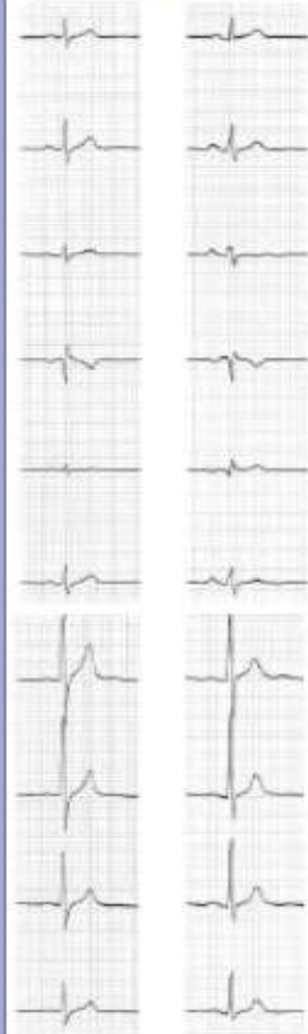
***Survival of healthy individuals with QTc <340 msec***

# HỘI CHỨNG QT NGẮN

## Short QT syndrome

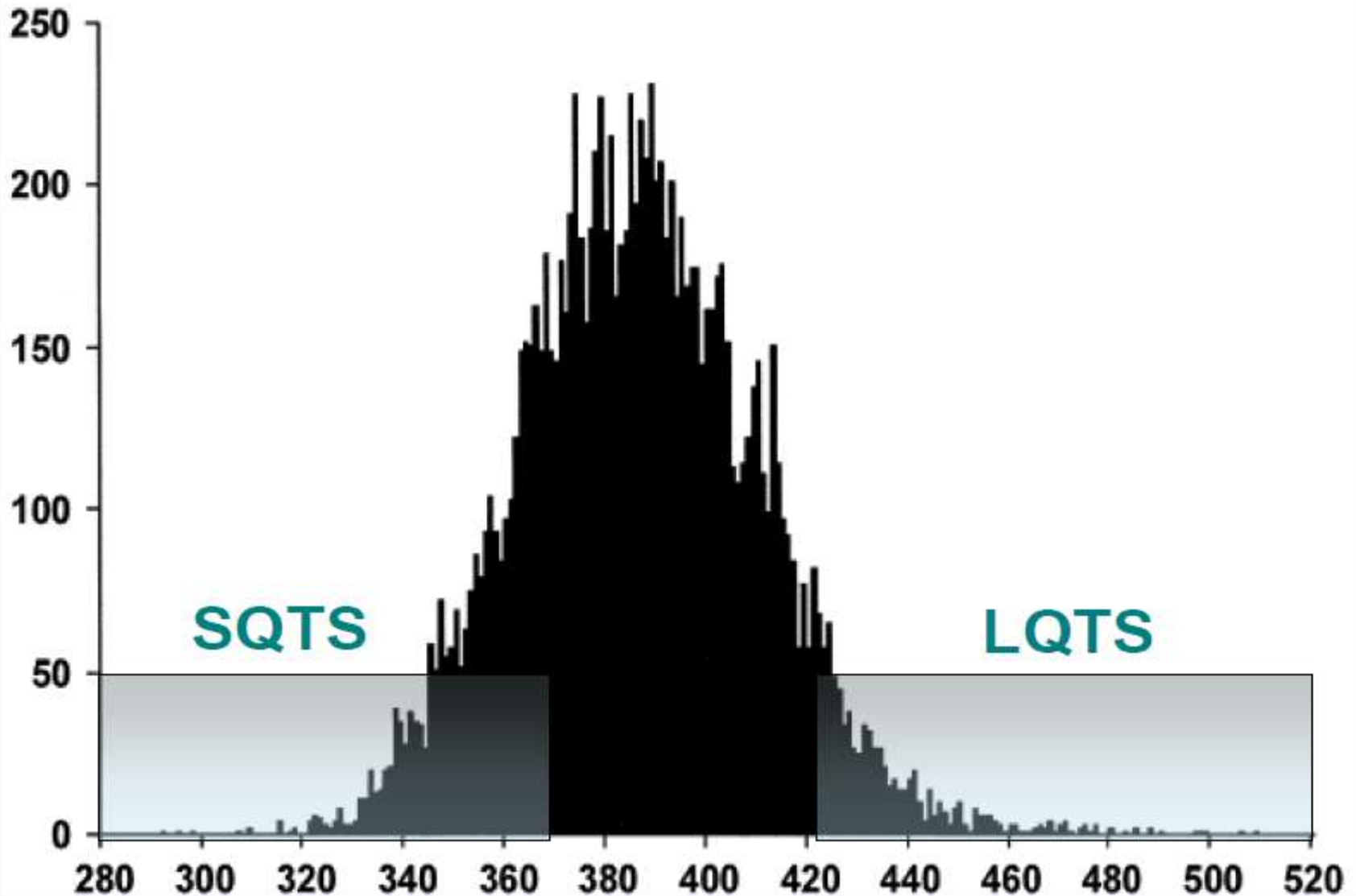


## Asymptomatic short QT



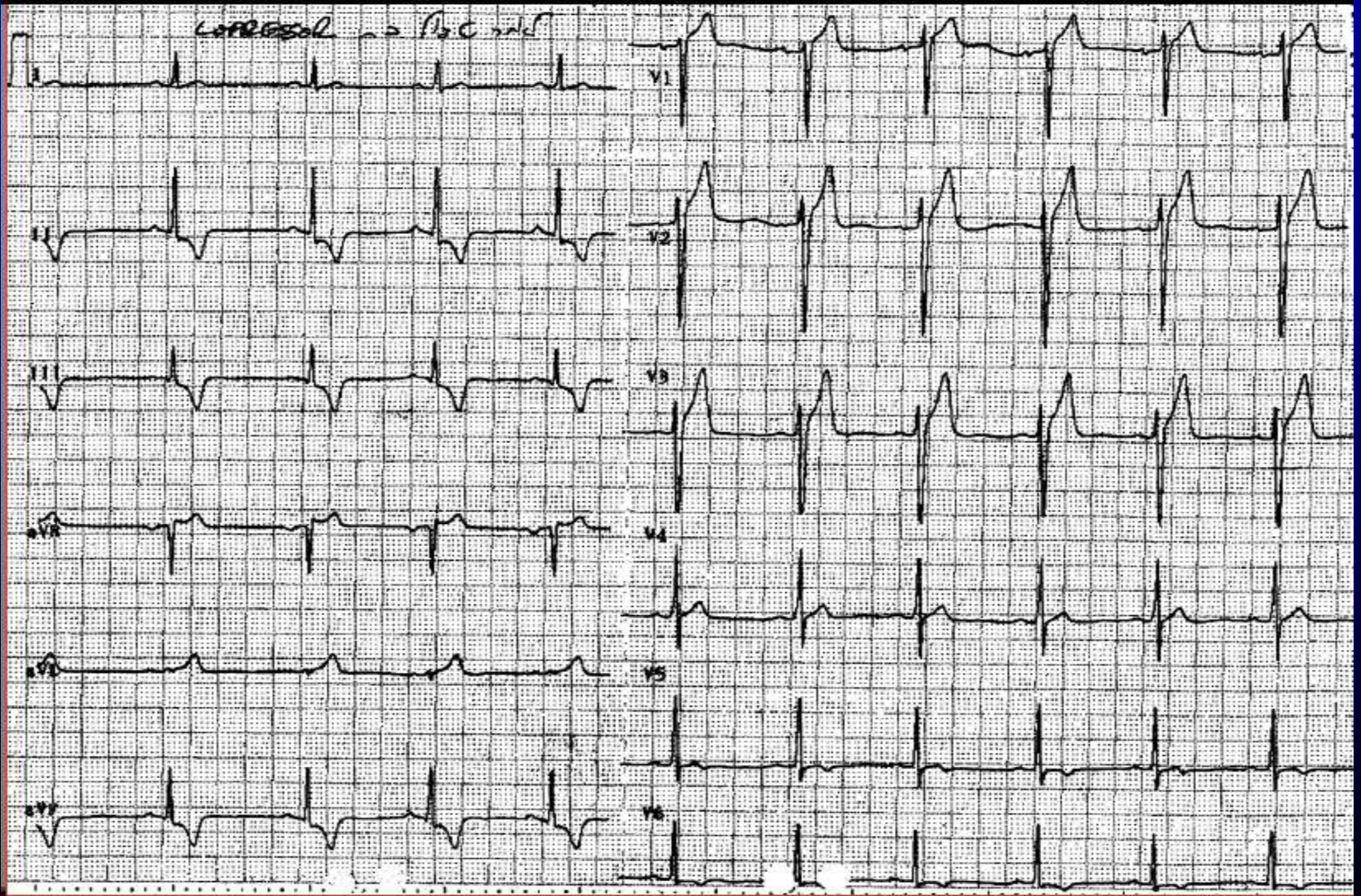


# HỘI CHỨNG QT NGẮN



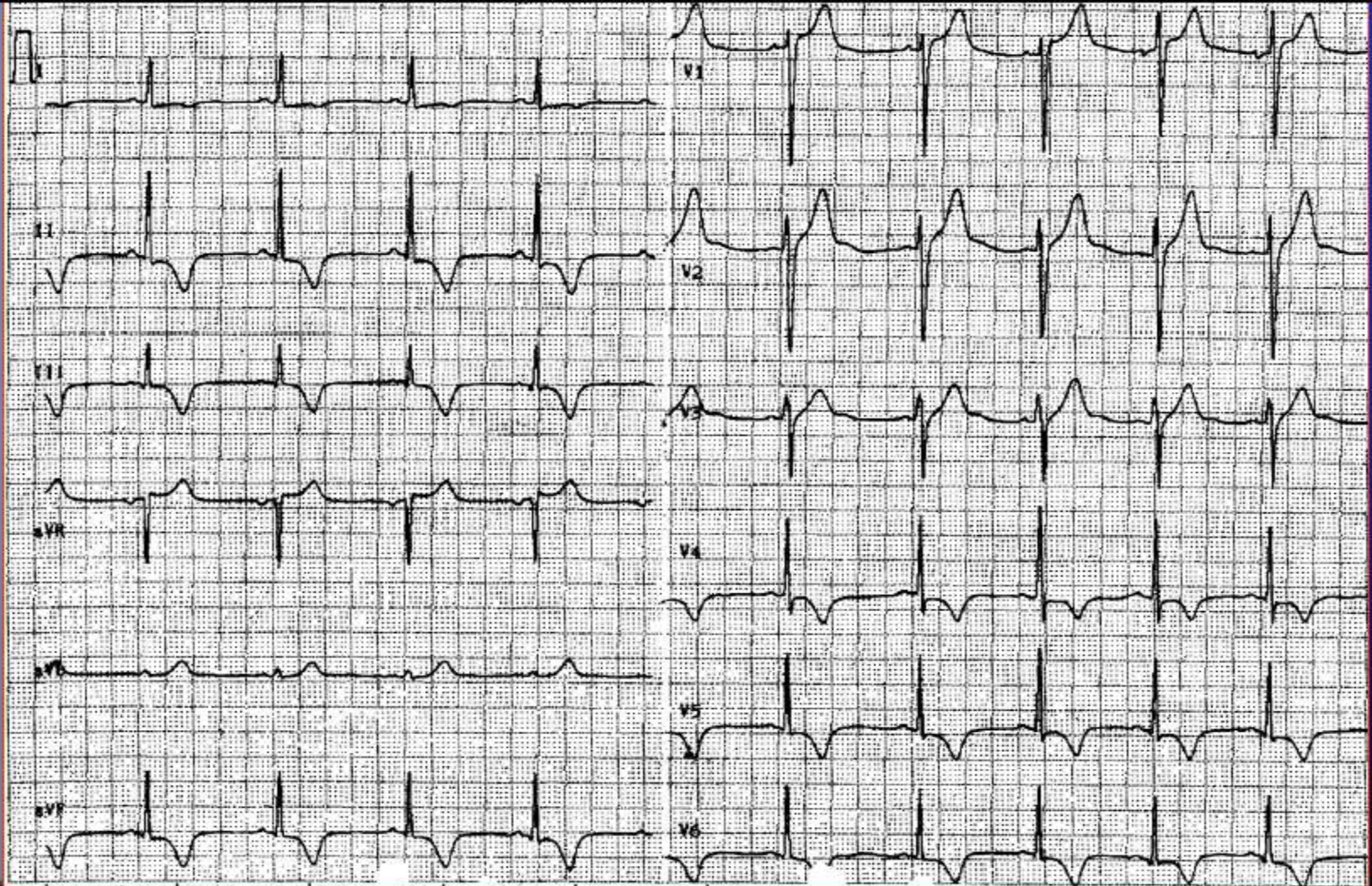
# HỘI CHỨNG QT NGẮN

Male/17. Palpitations and presyncope: Heart rate 72/min, QT 280; QTc 310 msec



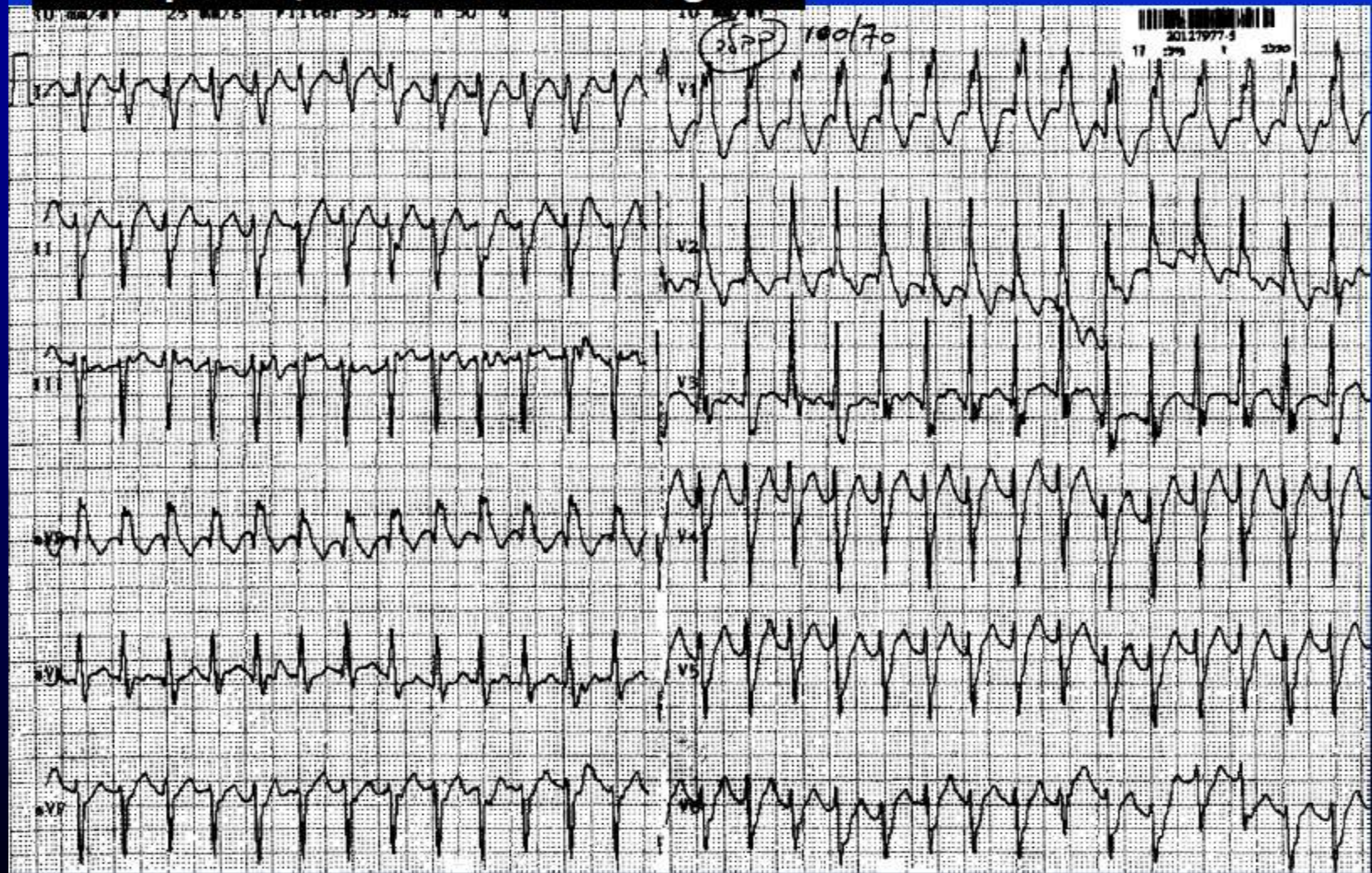
# HỘI CHỨNG QT NGẮN

Same patient, shortly thereafter: Heart rate 70/min, QT 400, QTc 440 msec.



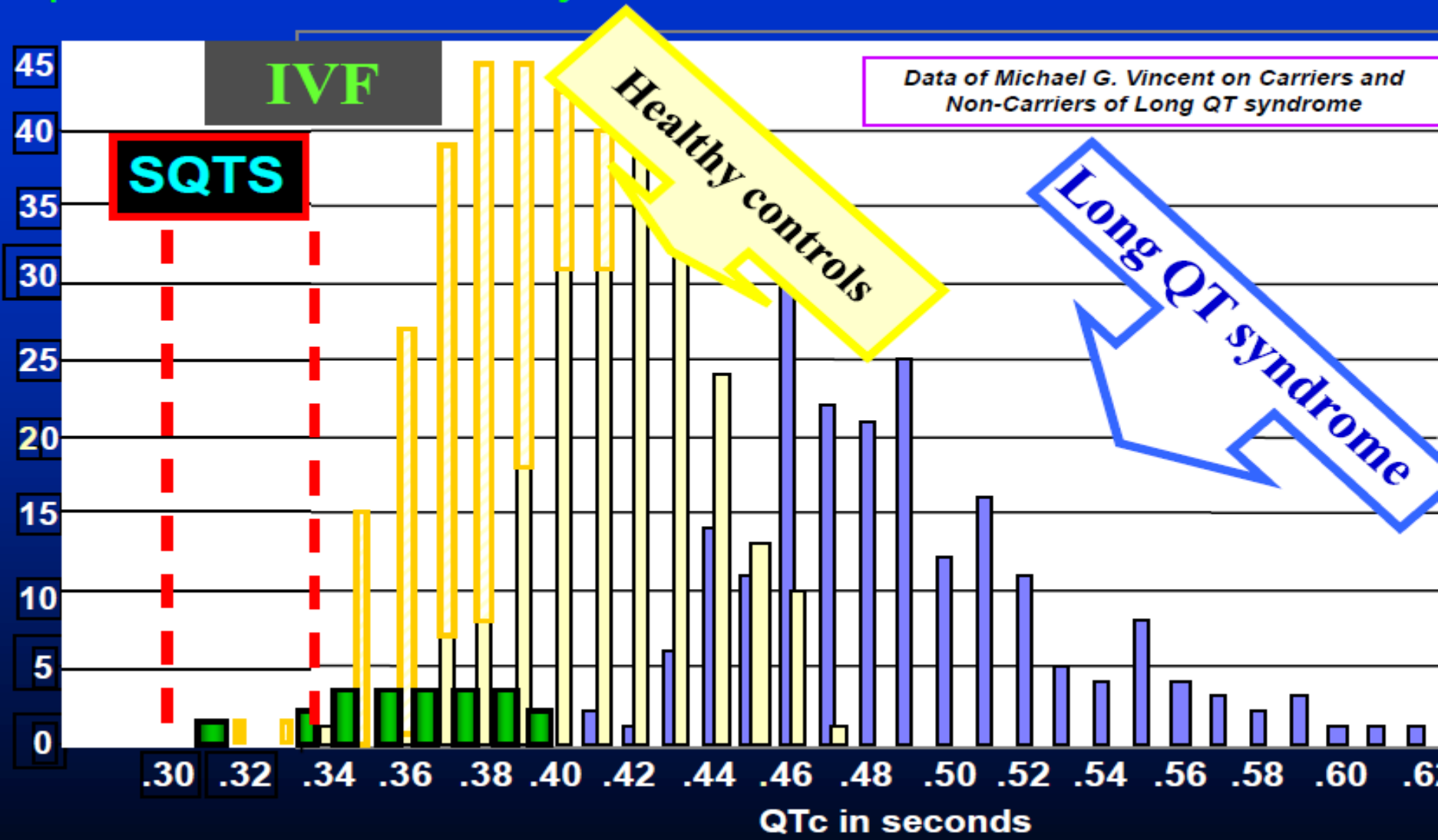
# HỘI CHỨNG QT NGẮN

Same patient, initial electrocardiogram.



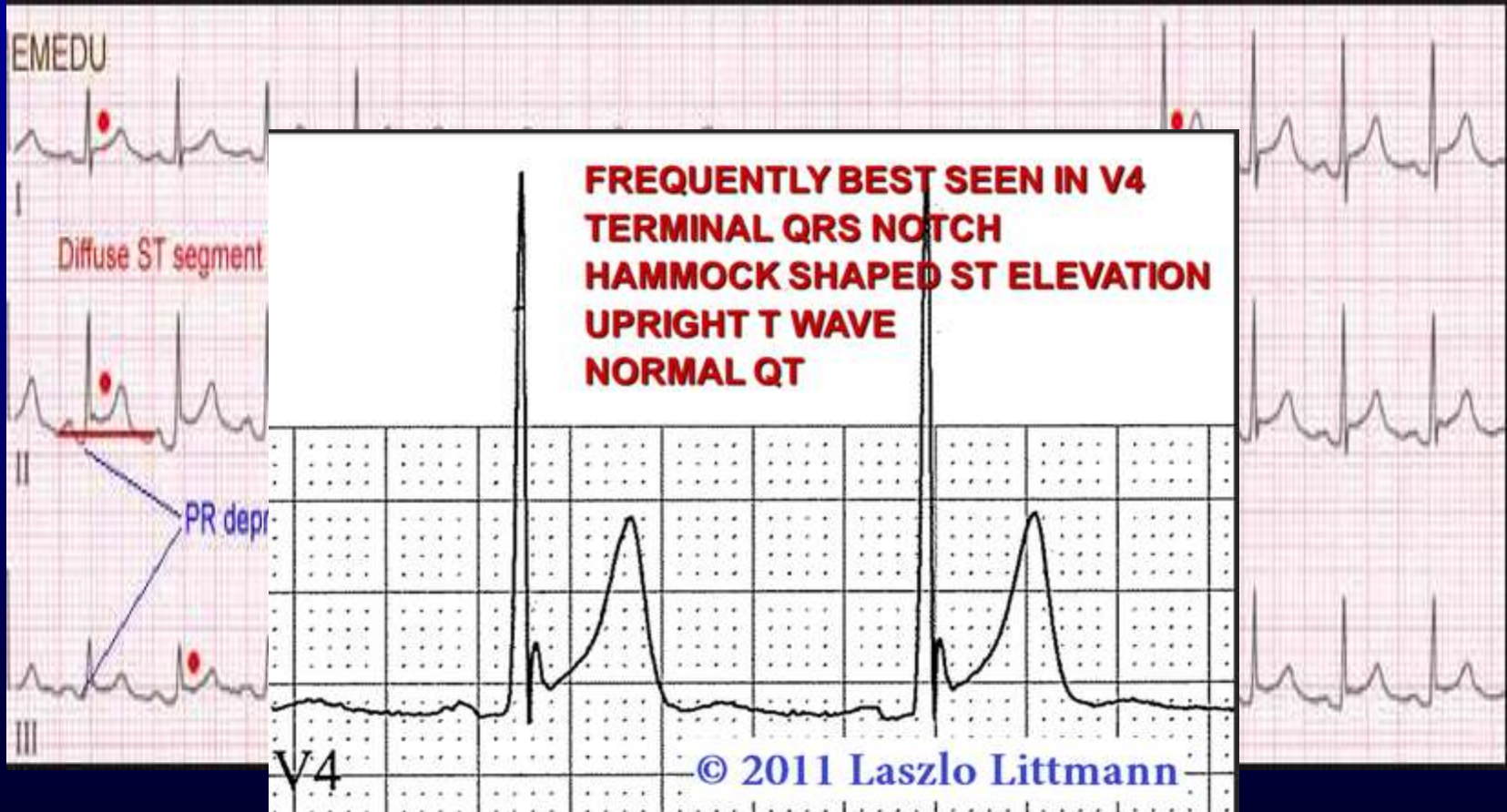
# HỘI CHỨNG QT NGẮN

Idiopathic VF: A short QT syndrome with not-so-short QT interval.



HỘI CHỨNG TÁI CỤC SỐM

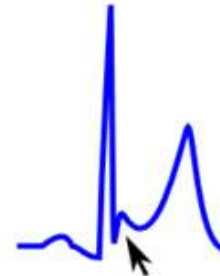
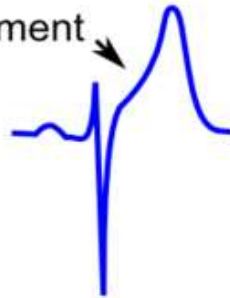
# HỘI CHỨNG TÁI CỨU SỚM



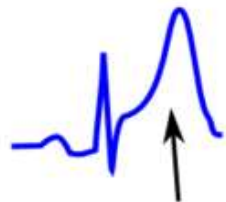
# HỘI CHỨNG TÁI CỨU SỚM

ST elevation due to early repolarization

concave ST segment



Notched or slurred J point



Symmetrical large T wave

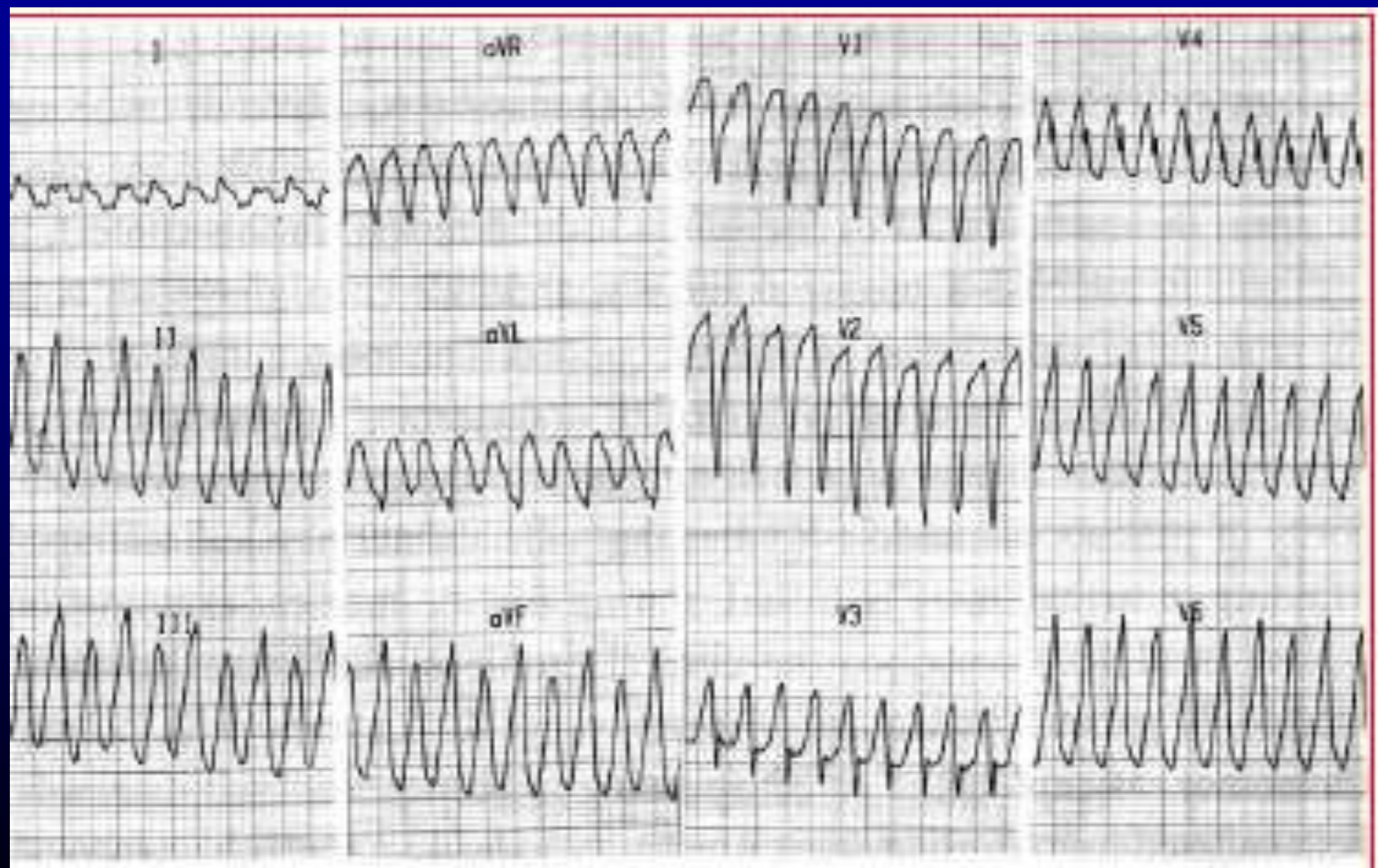


LOẠN SẢN THẤT PHẢI

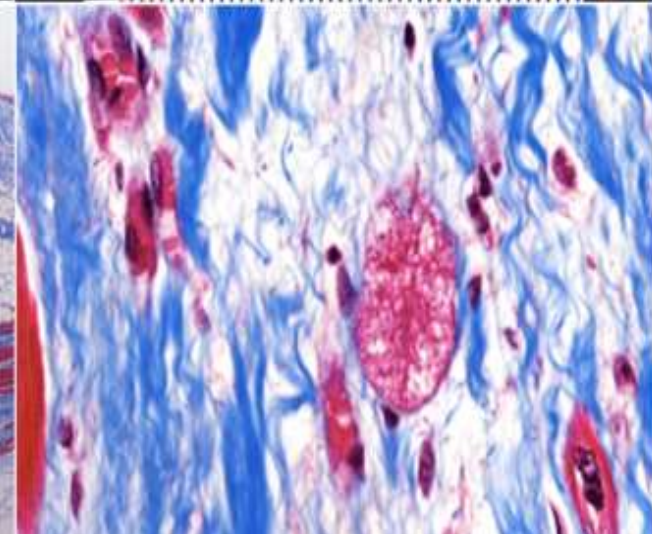
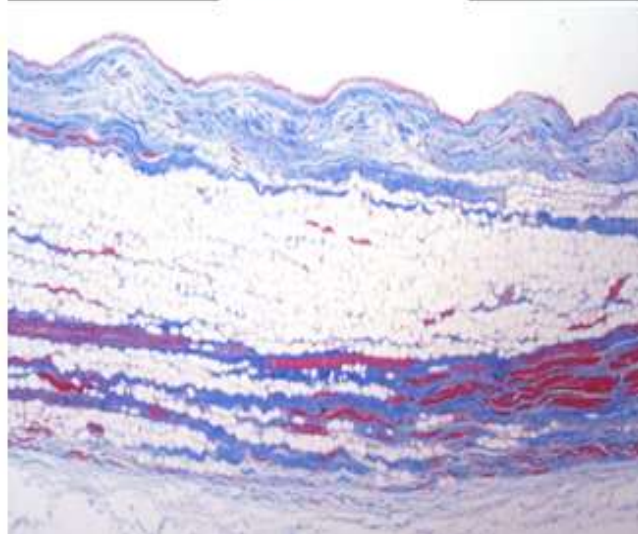
# NTT/T TRÊN BỆNH NHÂN ARVD



# TIM NHANH THẮT TRÊN BỆNH NHÂN ARVD



# BỆNH NHÂN ARVD

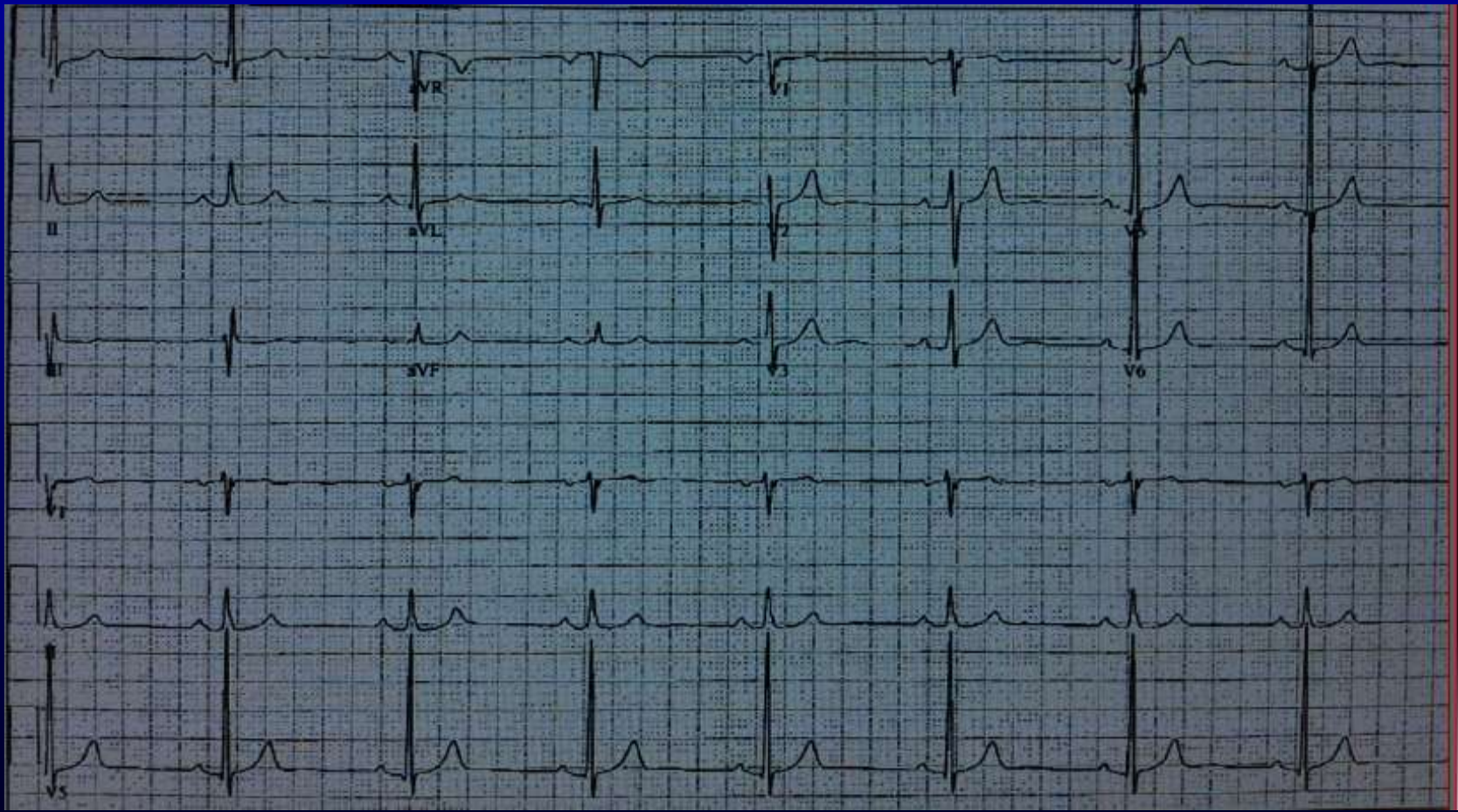


TIM NHANH THẮT ĐÁ HÌNH

# NTT/T ĐA Ổ



# BN NAM 38 TUỔI

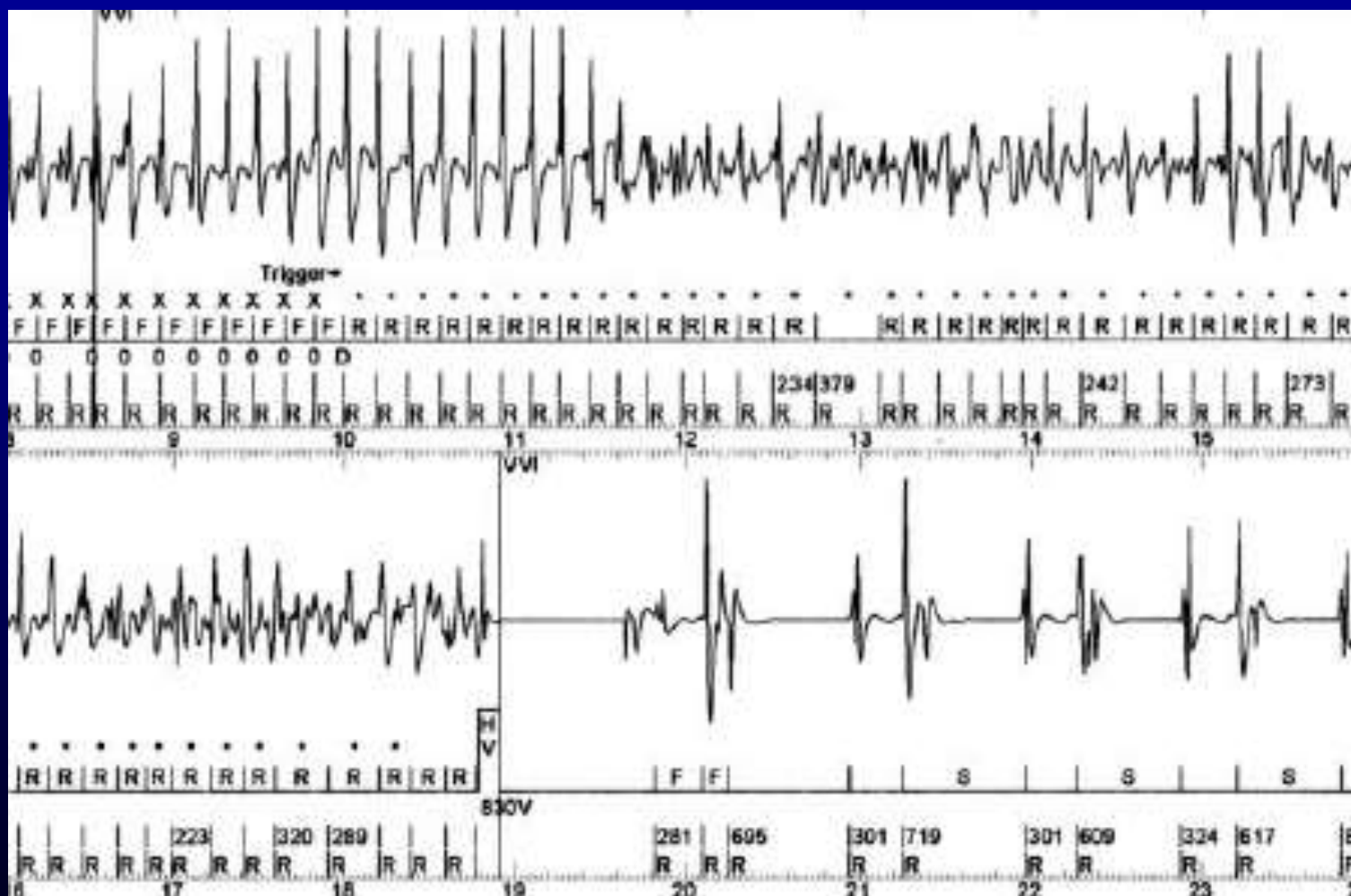


# TIM NHANH THẮT ĐA HÌNH





# TIM NHANH THẮT ĐA HÌNH



# XIN CẢM ƠN SỰ CHỦ Ý

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Tel:0913225648

