



Seção Médica de Eletrofisiologia Clínica e Arritmias Cardíacas  
Instituto Dante Pazzanese de Cardiologia

# O ECG como estratificador de risco em algumas cardiopatias congênitas e síndromes elétricas

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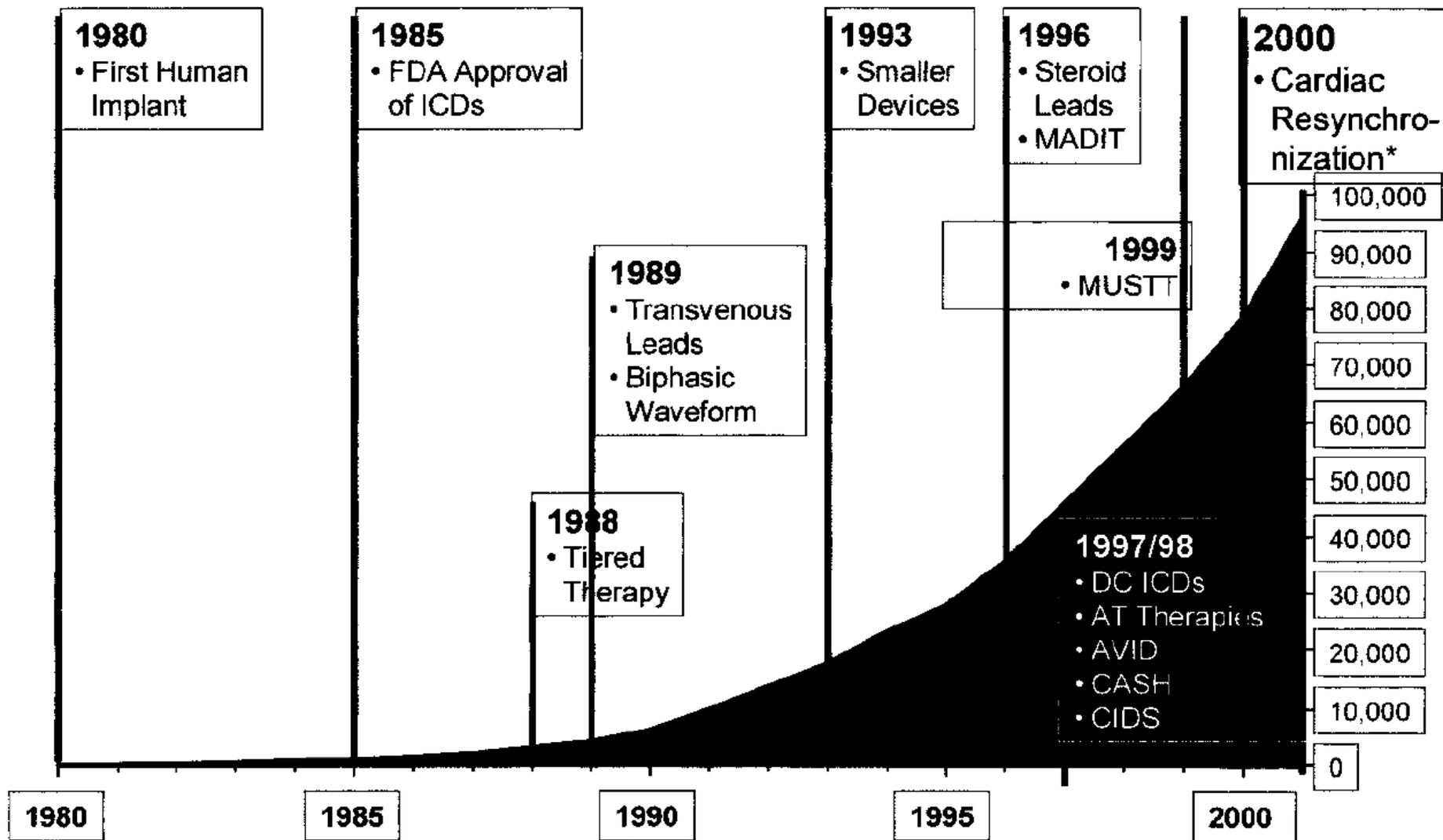


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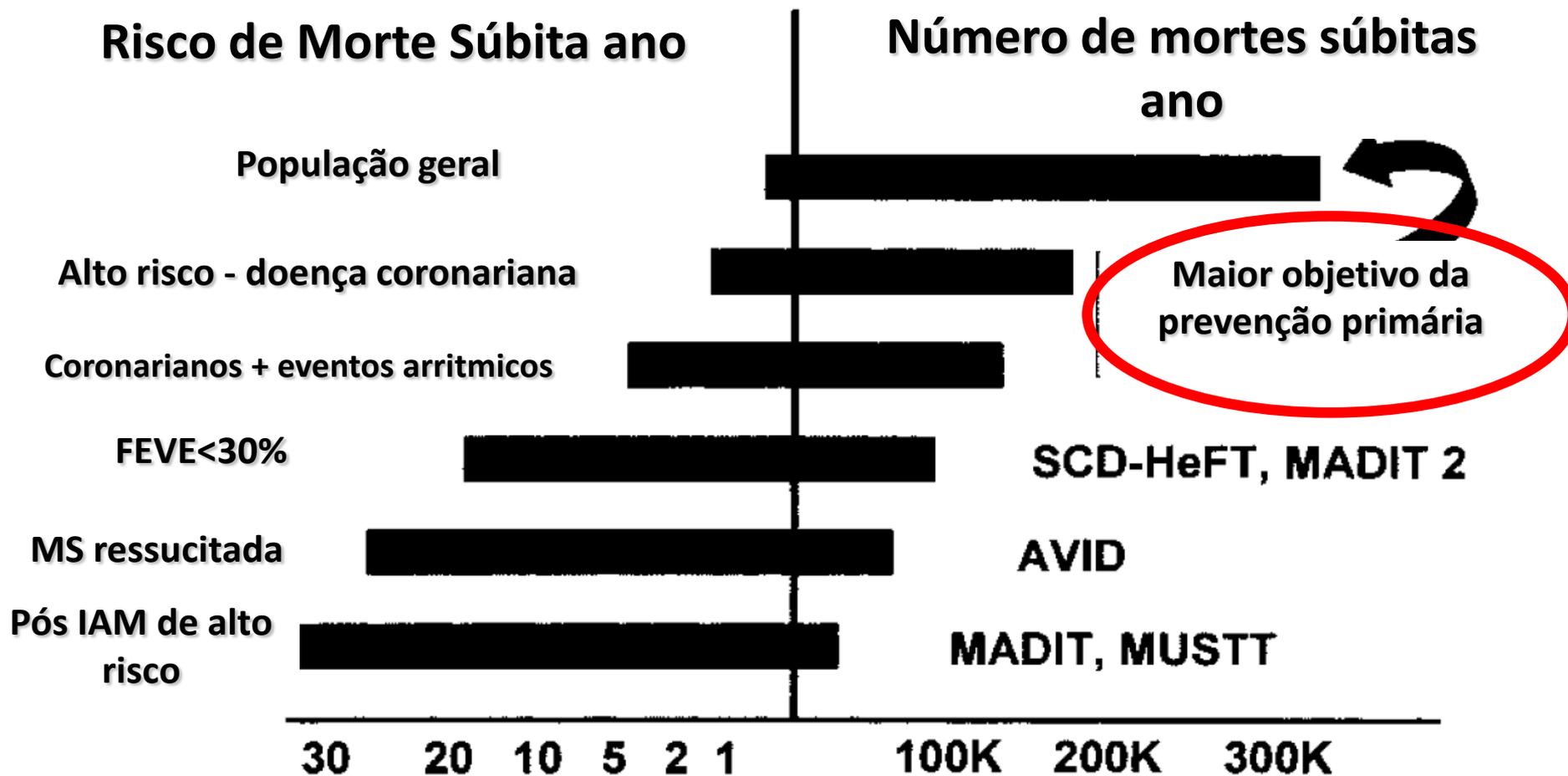




# Evolução da Indicação de CDI na população USA



# Morte cardíaca súbita



- **Profilaxia primária**
- **Profilaxia secundária**

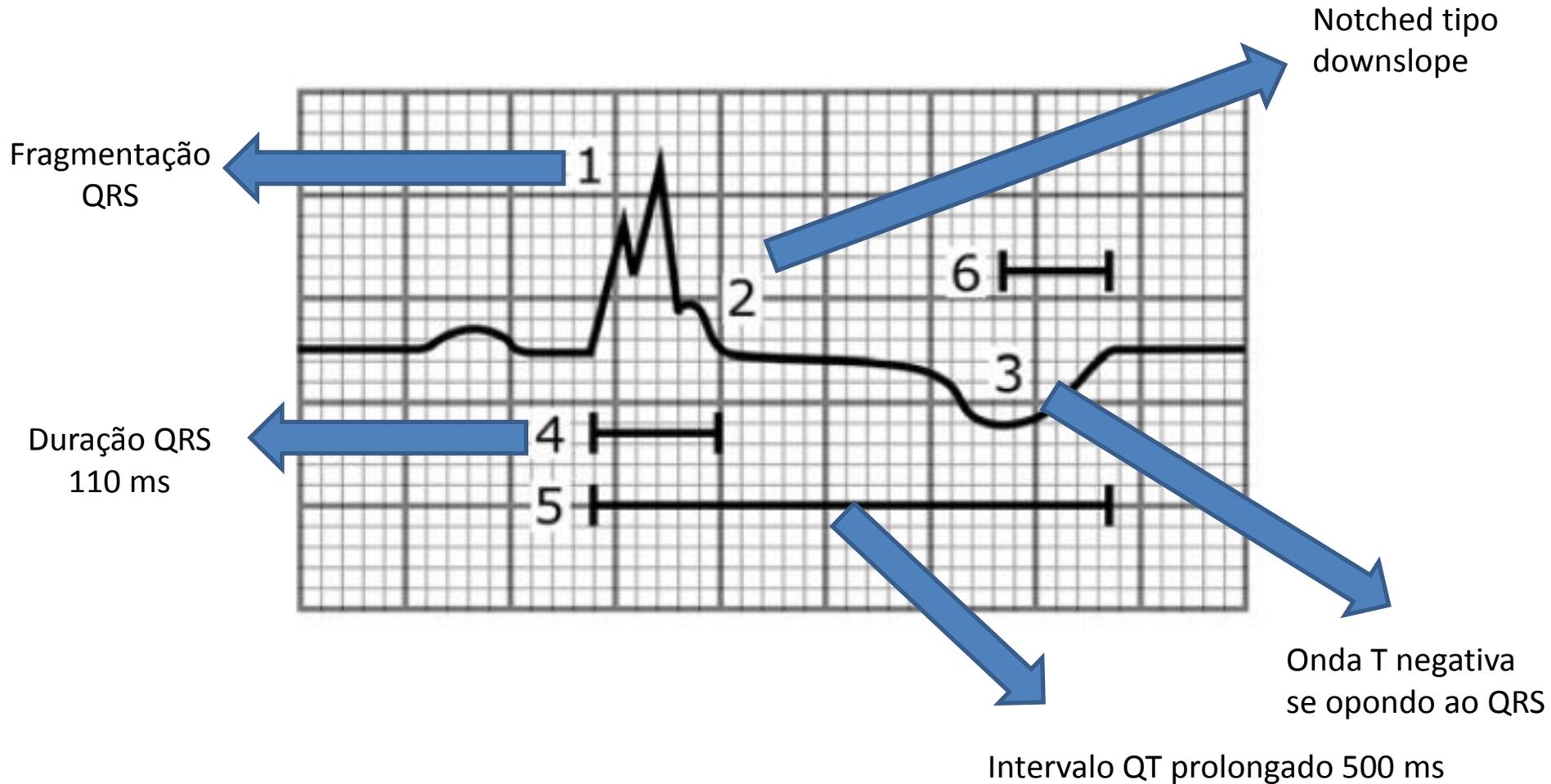
# Marcadores ECG de morte súbita em adultos

**Table 1.** Examples of studies assessing the SCD risk associated with electrocardiographic abnormalities in the general population.

Authors	Study population, follow-up	ECG parameter studied	Adjusted HR (95%CI) for SCD
Jouven et al. [20]	5713 men (42–53y); follow-up 23 years	Resting heart rate >75bpm	3.46 (1.60–7.44)
Aro et al. [8]	10,899 subjects (52% men, mean age 44 ± 8 y); follow-up 30 ± 11y	QRS ≥110ms	2.14 (1.38–3.33)
		IVCD (QRS ≥110ms without bundle-branch block)	3.11 (1.74–5.54)
		LBBB	2.71 (1.20–6.11)
Kurl et al. [9]	2049 men (age 42–60y); follow-up 19y	QRS >110ms	2.50 (1.38–4.55)
Aro et al. [21]	10,815 subjects (52% men, mean age 44 ± 8 y); follow-up 30 ± 11y	Delayed frontal QRS transition zone	1.89 (1.18–3.03)
Tikkanen et al. [17]	10,864 subjects (52% men, mean age 44 ± 8 y); follow-up 30 ± 11y	Inferior ER ≥0.1mV	1.43 (1.06–1.94)
		Inferior ER >0.2mV	2.92 (1.45–5.89)
		LVH (Sokolow-Lyon)	1.16 (1.05–1.27) <sup>a</sup>
Aro et al. [8]	10,713 subjects (52% men, mean age 44 ± 8 y); follow-up 30 ± 11y	QRS-T angle ≥100°	2.26 (1.59–3.21)
Aro et al. [8]	10,899 subjects (52% men, mean age 44 ± 8 y); follow-up 30 ± 11y	Inferior/lateral T-wave inversions	3.16 (1.86–5.36)
Straus et al. [13]	7983 subjects (61% women, age ≥55y)	Prolonged QTc (>450ms in men, >470ms in women)	2.5 (1.3–4.7)

<sup>a</sup>Cardiac death.

# Principais marcadores QRS com valor prognóstico



# Marcadores ECG na população pediátrica e em jovens



# Marcadores ECG na população pediátrica e em jovens



**Tabela 84.3** – Correlação entre tipo de cardiopatia congênita e possíveis arritmias

Condição clínica	Substrato	Arritmias pré ou pós-operatórias	Comentários
Comunicação interatrial	Dilatação do átrio direito (pré-operatório) e atriotomia e manipulação atrial (pós-operatório)	Taquicardia atrial (pré e pós) Flutter atrial (pré e pós) Fibrilação atrial (pré e pós) Lesão mecânica pós-operatória do sistema elétrico do coração (pós)	Fibrilação, <i>flutter</i> atrial e taquicardia atrial geralmente surgem na evolução natural tardia
Comunicação interventricular	Lesão cirúrgica do sistema de condução ou processo inflamatório pós-CEC	Bloqueios atrioventriculares (pós) Taquicardia juncional (pós) Taquicardias ventriculares (pré e pós com disfunção miocárdica)	Episódios de taquicardia juncional no pós-operatório são bem tratados com propafenona, se não houver disfunção
Transposição corrigida de grandes artérias (L-TGA)	Distorção do sistema de condução pela inversão ventricular, lesão cirúrgica	Bloqueios atrioventriculares até BAVT	Pode ocorrer espontaneamente ou no pós-operatório de ventriculoseptoplastia, por exemplo
Isomerismo atrial	Esquerdo: ausência de nódulo sinusal; direito: dois nódulos sinusais; presença de dois nódulos atrioventriculares	Arritmias atriais (ritmos atriais ectópicos, taquicardia atrial multifocal) Taquicardia por reentrada atrioventricular por feixe acessório ou pelos dois NAV	Pode ter diversas arritmias atriais primárias ou secundárias a procedimentos cirúrgicos
Tetralogia de Fallot	Disfunção do VD ou lesão pos-operatória	Arritmias ventriculares (simples ou complexas) Maior risco quanto mais tardio o pós-operatório e maior a idade da correção cirúrgica	Risco de morte súbita e arritmias malignas quando QRS > 180 ms, insuficiência pulmonar importante e disfunção de VD
Anomalia de Ebstein	Dilatação atrial e feixes anômalos	Arritmias atriais ( <i>flutter</i> e FA) WPW	Maior incidência de feixe anômalo que a população geral
Miocardiopatia hipertrófica	Feixes acessórios e reentradas na musculatura ventricular	TPSV por macrorreentrada atrioventricular	Septo maior que 30 mm, síncope, TVNS, TVS e história familiar





**Piora da função**

**Mecanismos compensatórios**

**Estresse adrenérgico**

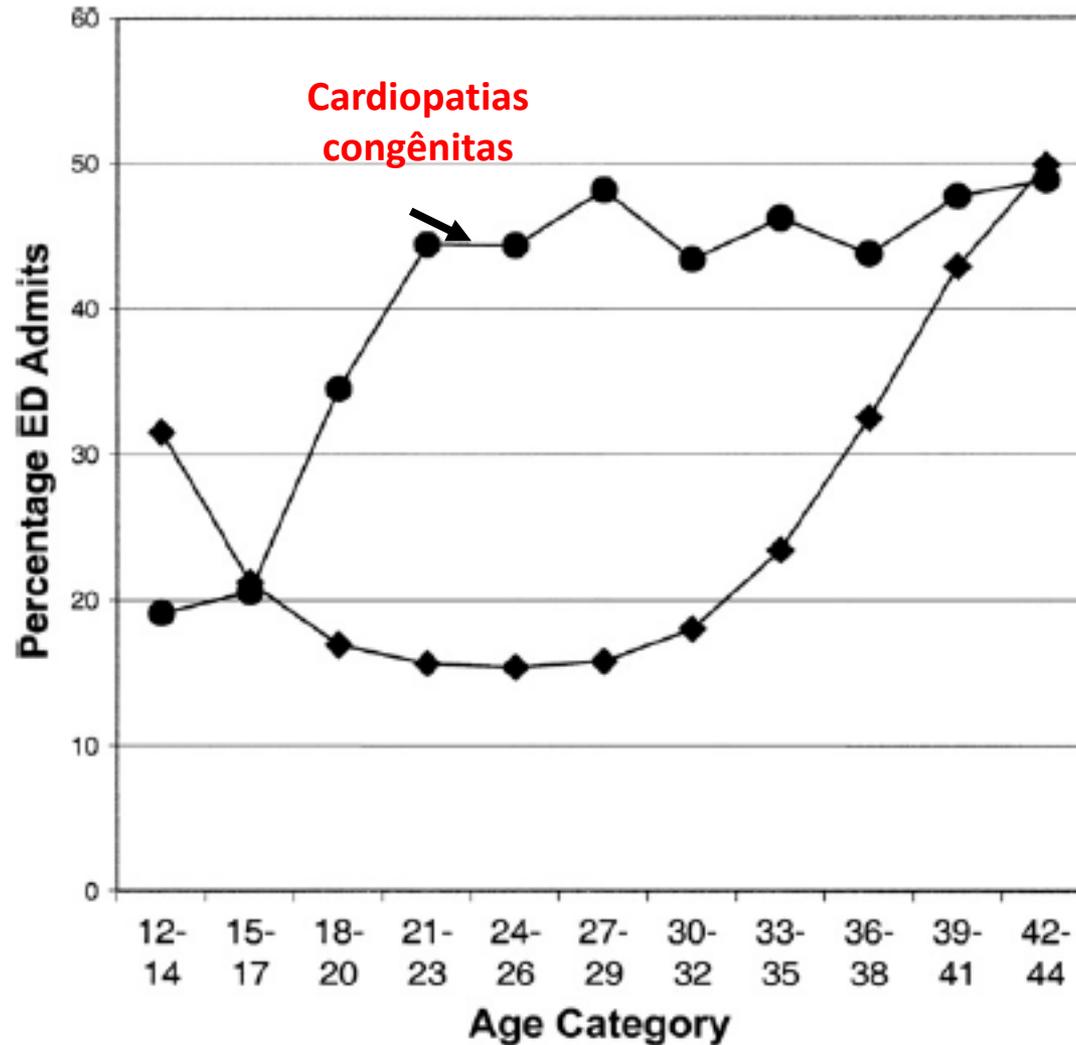
**Piora da função ventricular e aumento dos substratos**

**Eventos arrítmicos**

# Changes in Hospitalization Patterns Among Patients With Congenital Heart Disease During the Transition From Adolescence to Adulthood



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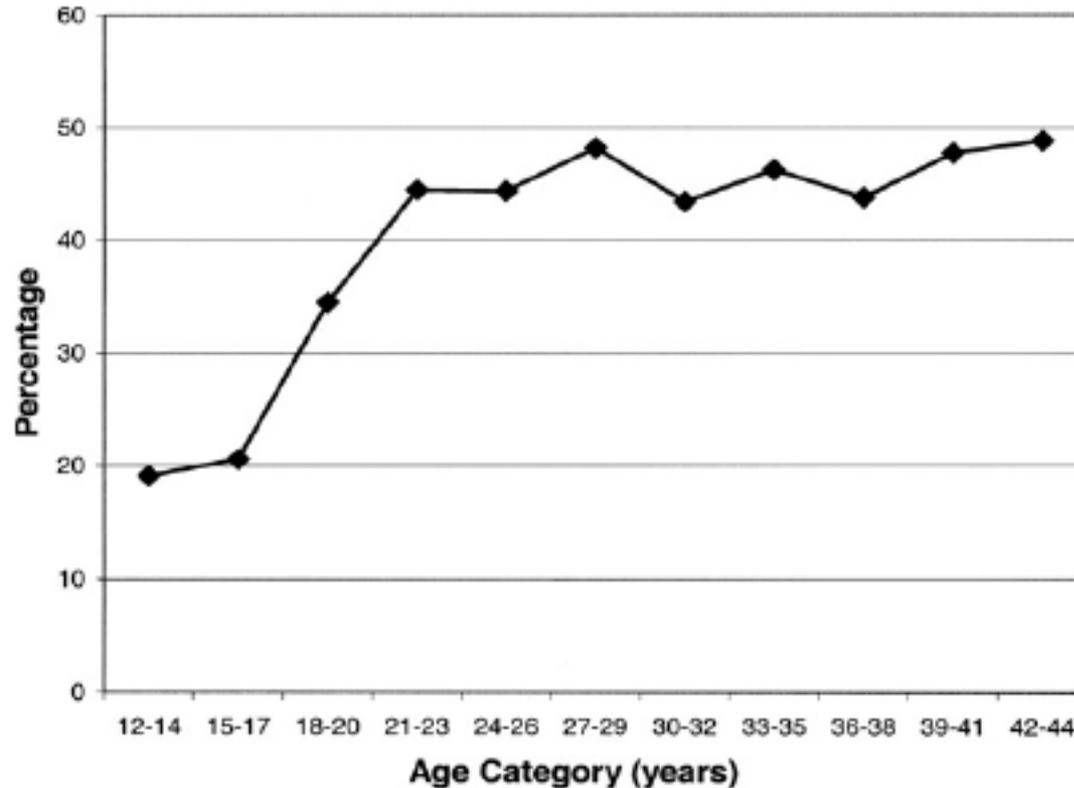


(J Am Coll Cardiol 2007;49:875-82)

# Changes in Hospitalization Patterns Among Patients With Congenital Heart Disease During the Transition From Adolescence to Adulthood



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**Porcentagem das admissões no PS entre  
2000 e 2003 na Califórnia**

(J Am Coll Cardiol 2007;49:875-82)

# The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period

## The Euro Heart Survey on adult congenital heart disease

Table 2 History of morbidity and functional status at baseline

	Endocarditis	CVA/TIA	MI/CABG/PTCA	Supraventricular arrhythmias	Ventricular arrhythmias	NYHA I	NYHA II	NYHA III/IV
ASD II	0 (3/879)	4 (36/876)	2 (16/880)	28 (243/861)	2 (16/861)	59 (504/825)	30 (257)	11 (91)
VSD	7 (44/621)	2 (14/622)	1 (5/626)	9 (56/617)	3 (18/617)	76 (472/619)	19 (118)	4 (29)
Tetralogy of Fallot	4 (33/810)	4 (32/807)	1 (5/806)	20 (159/796)	14 (115/796)	69 (551/793)	24 (189)	6 (53)
CoA	2 (11/547)	1 (6/545)	1 (4/546)	4 (21/547)	2 (9/547)	81 (441/543)	15 (84)	3 (18)
Transposition	1 (4/361)	6 (20/359)	0 (1/360)	26 (95/362)	2 (7/362)	67 (244/362)	27 (96)	6 (22)
Marfan syndrome	2 (5/284)	1 (4/285)	1 (2/286)	8 (23/281)	2 (6/281)	79 (223/282)	19 (54)	2 (5)
Fontan circulation	2 (3/198)	12 (23/198)	0 (0/198)	45 (90/198)	0 (0/198)	40 (78/193)	51 (98)	9 (17)
Cyanotic defect	6 (22/386)	10 (37/384)	0 (1/385)	16 (58/374)	6 (22/374)	12 (45/172)	44 (172)	44 (170)
Overall	3 (125/4086)	4 (172/4076)	1 (34/4088)	18 (745/4036)	5 (193/4036)	63 (2558/4031)	26 (1068)	10 (405)

All values are percentage followed by a fraction with frequency in the numerator and the number of patients with sufficient data to evaluate the item ( $k/n$ ) in the denominator. In the last two columns only the numerators are given, as the rows of the last three columns sum to 100%.

CVA, cerebrovascular accident; TIA, transient ischaemic attack; MI, myocardial infarction; CABG, coronary artery bypass graft; PTCA, percutaneous transluminal coronary angioplasty; NYHA, New York Heart Association functional class.



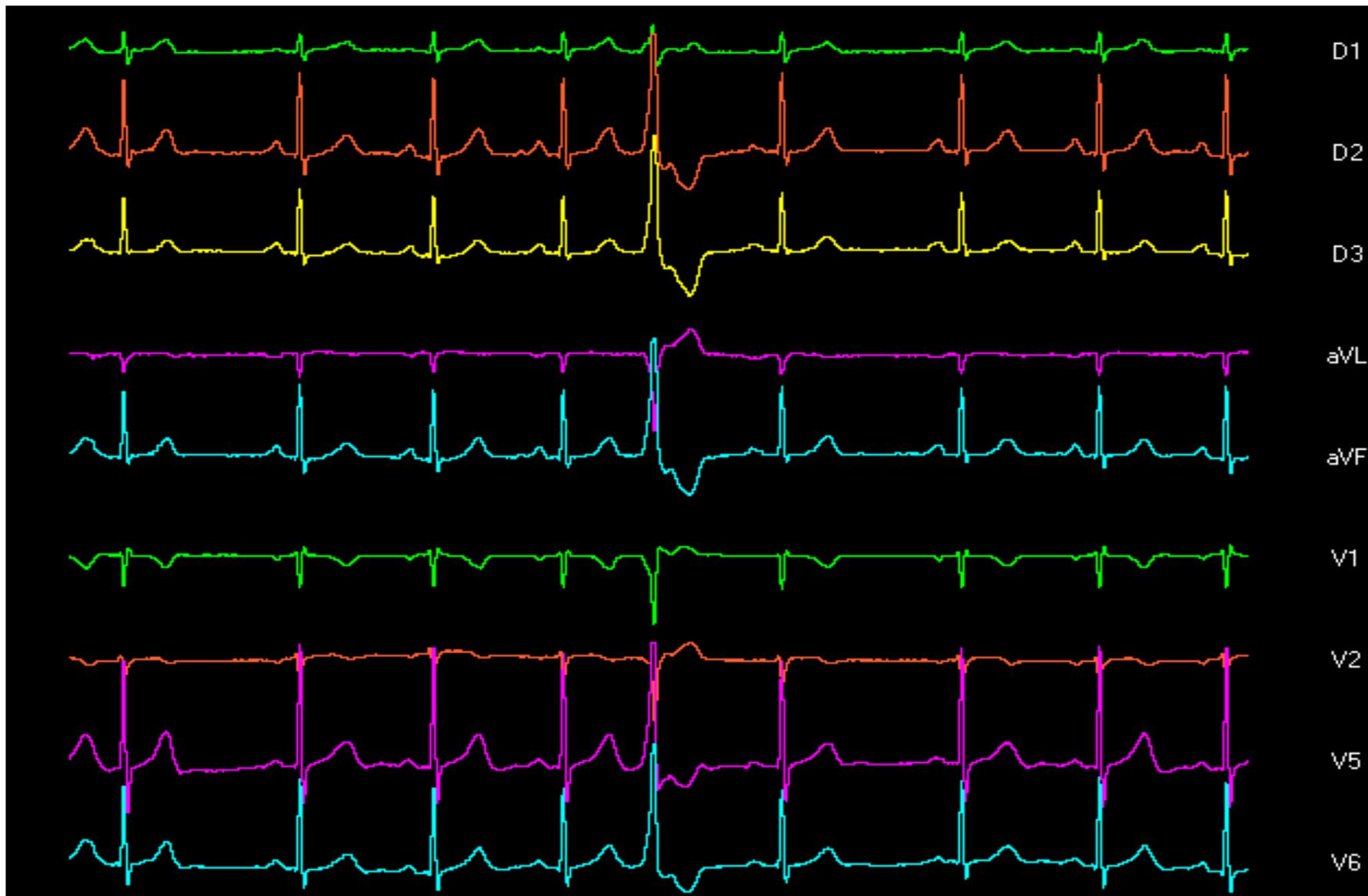
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SOCIETY OF  
CARDIOLOGY®

European Heart Journal (2005) 26, 2325–2333  
doi:10.1093/eurheartj/ehi396

# Mecanismos arritmia

- **Ventriculotomia**
- **Atriotomia**
- **Cicatrização irregular e ilhas de músculo cardíaco normal**
- **Dilatação camaras direitas e áreas de fibrose**

# ARRITMIA DE V.D.



安静時

08年 5月12日

ID: 0000007

氏名:

午前 9時29分

性別:

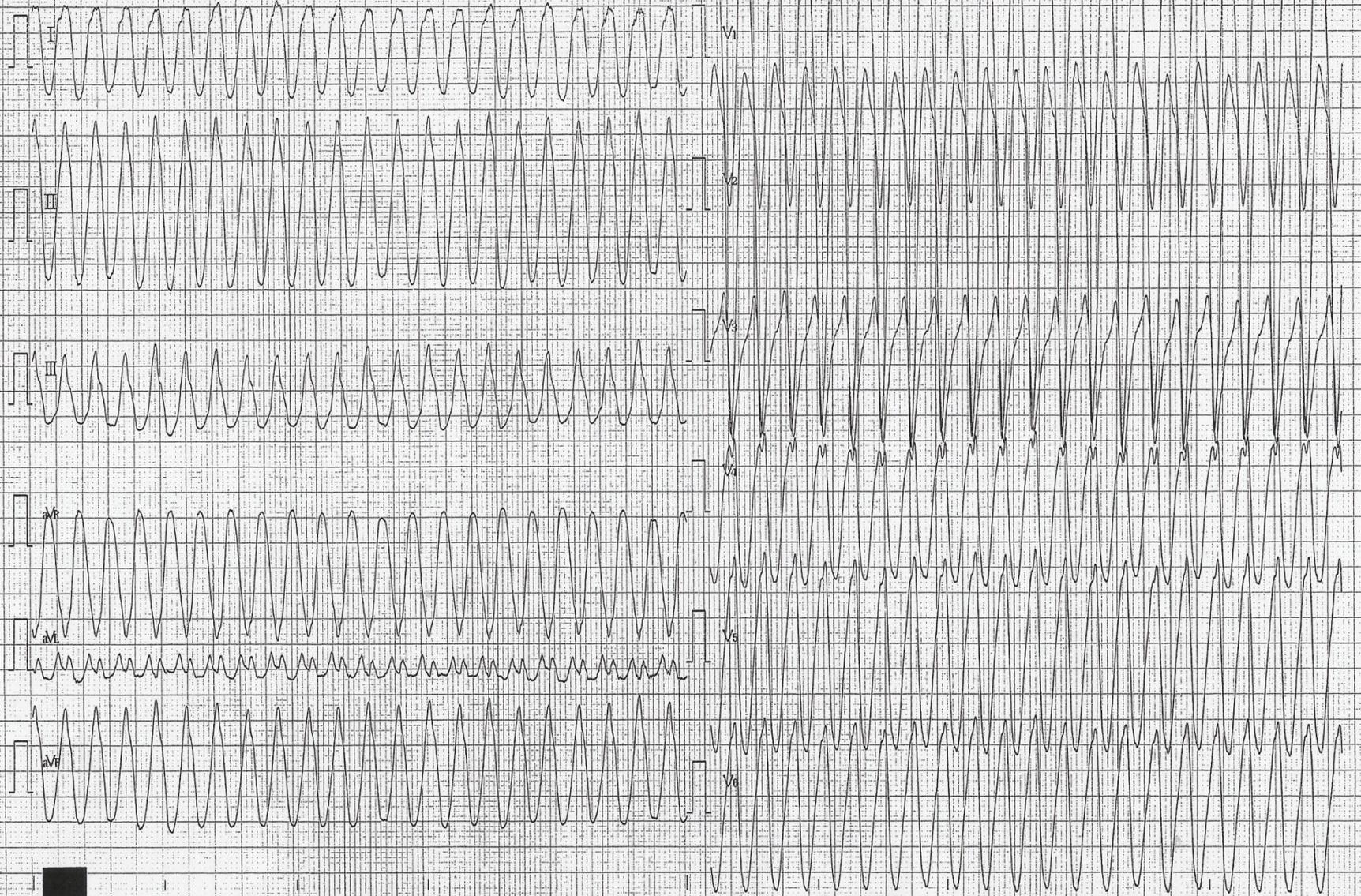
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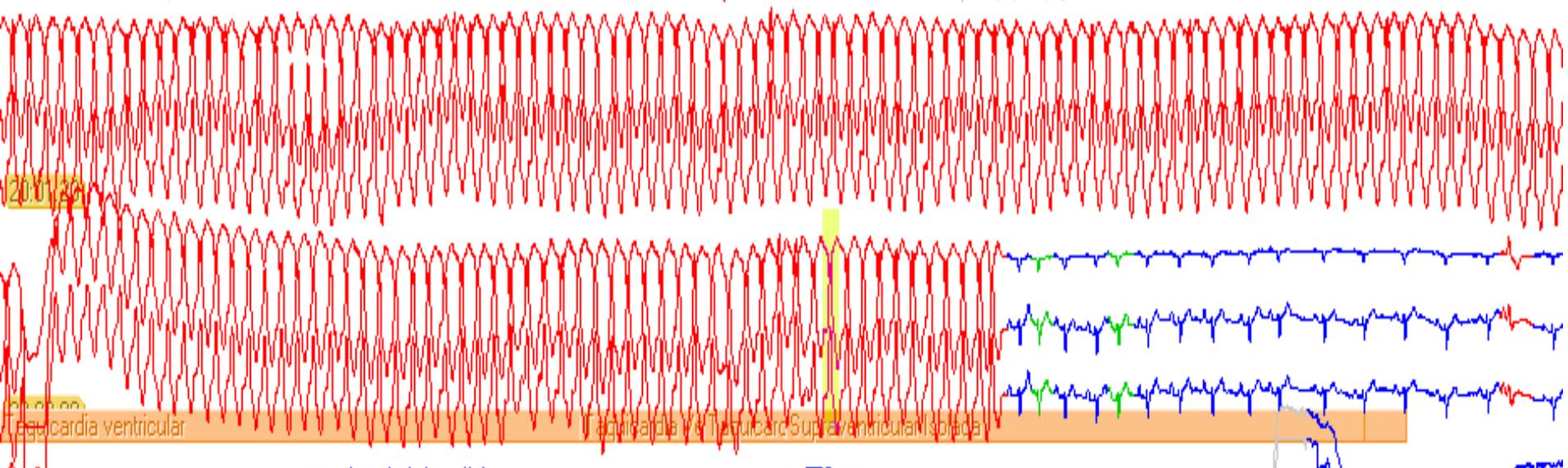
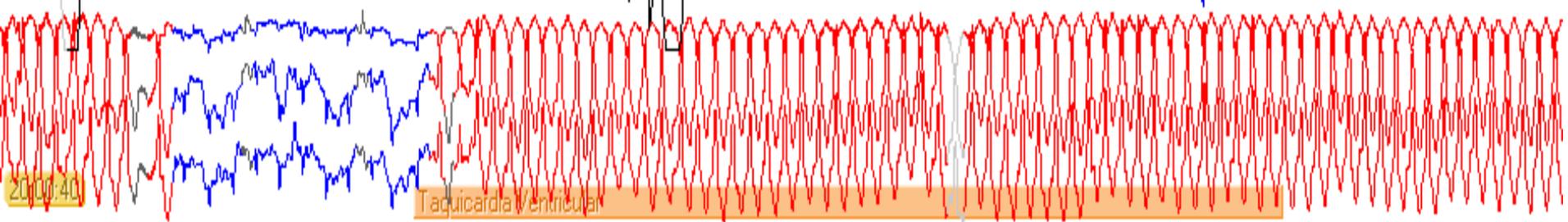
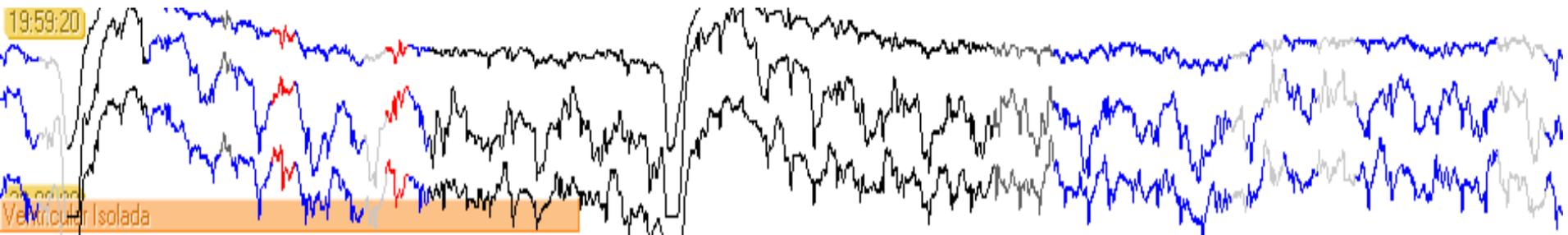
HR: \*\*\* BP:

フィルタ: DF, MF, AC

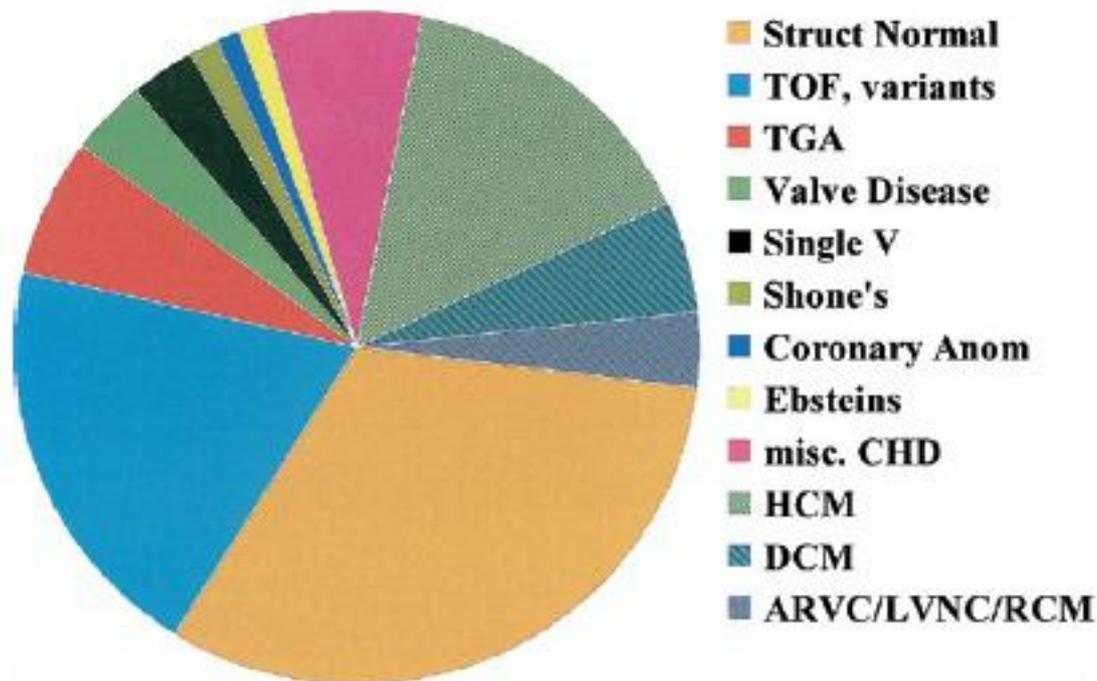
1cm/mV 25mm/S

1cm/mV





# Results of a Multicenter Retrospective Implantable Cardioverter-Defibrillator Registry of Pediatric and Congenital Heart Disease Patients

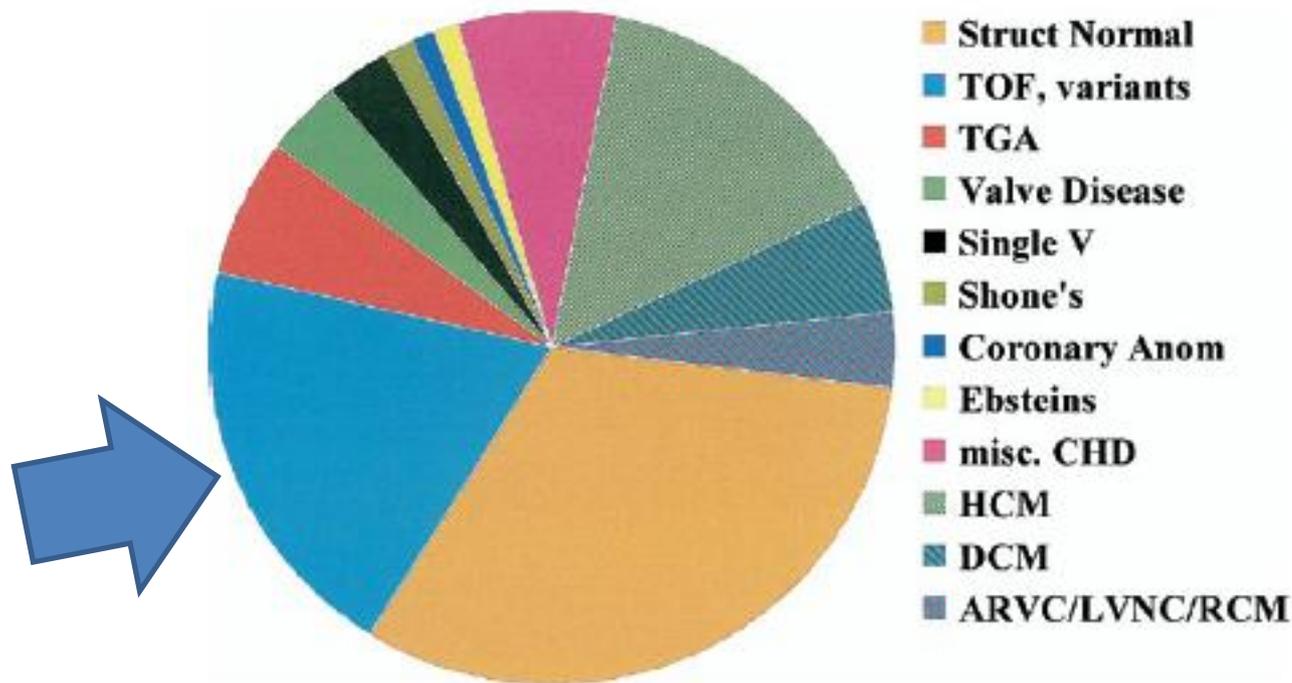


**Figure 1**

**Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients**

(J Am Coll Cardiol 2008;51:1685-91)

# Results of a Multicenter Retrospective Implantable Cardioverter-Defibrillator Registry of Pediatric and Congenital Heart Disease Patients



**Figure 1**

Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients

(J Am Coll Cardiol 2008;51:1685-91)

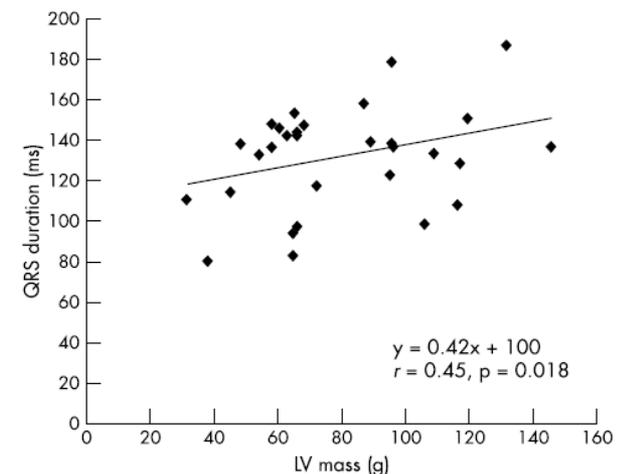
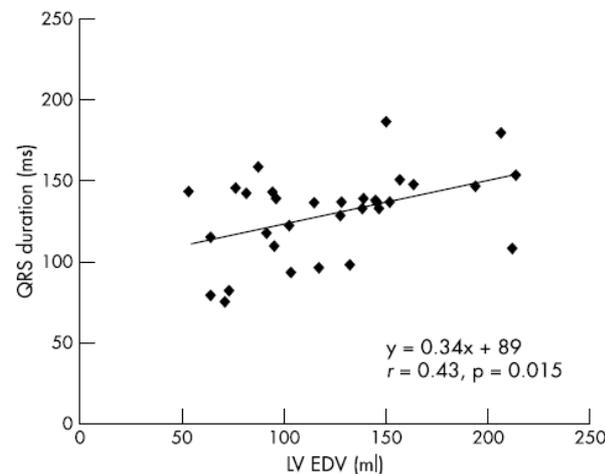
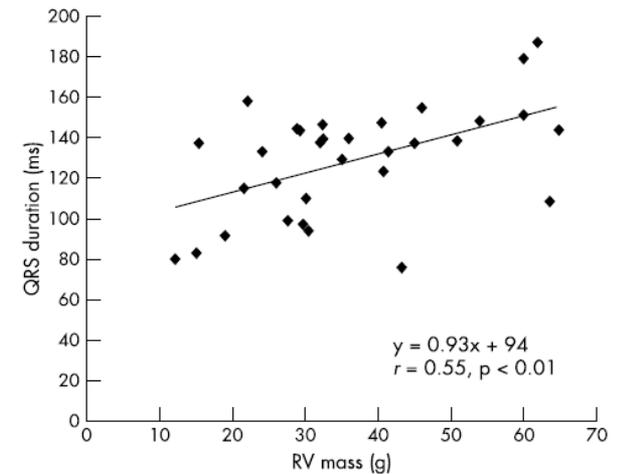
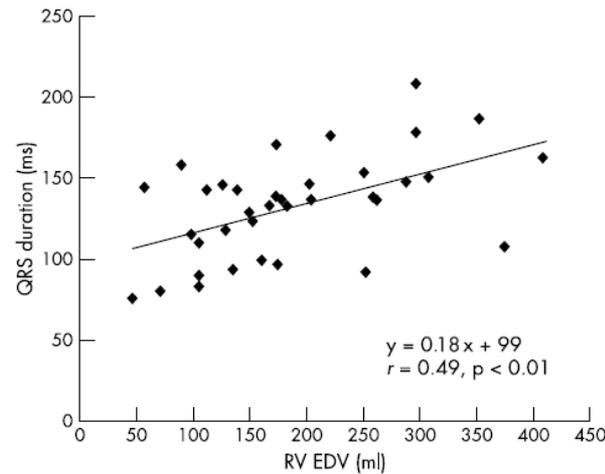
## CONGENITAL HEART DISEASE

# ECG predictors of ventricular arrhythmias and biventricular size and wall mass in tetralogy of Fallot with pulmonary regurgitation

W A Helbing, A A W Roest, R A Niezen, H W Vliegen, M G Hazekamp, J Ottenkamp, A de Roos, E E van der Wall

*Heart* 2002;**88**:515–520

**Correlação positiva entre aumento do QRS e aumento do volume diastólico final e massa ventricular**

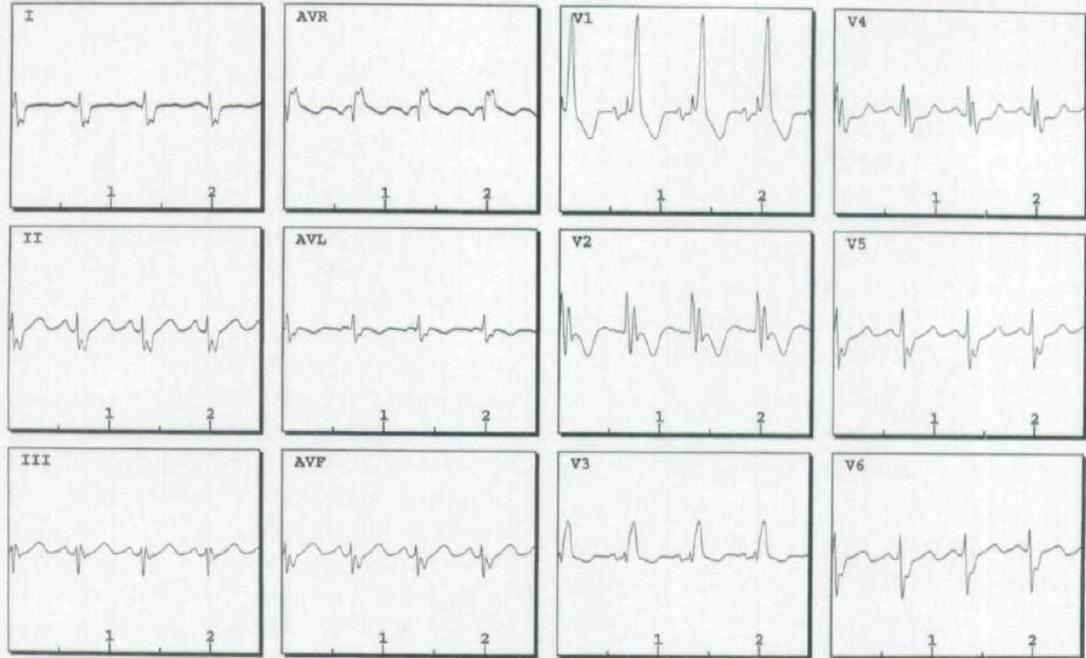


## Electrocardiographic Markers of Late Sudden Death Risk in Postoperative Tetralogy of Fallot Children

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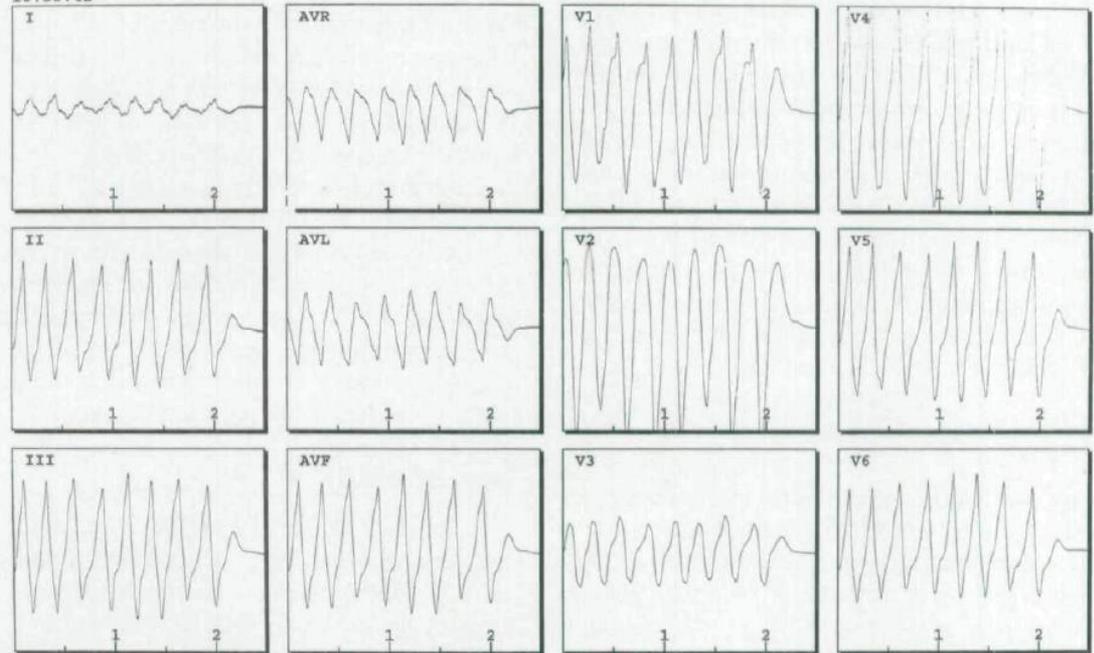
From the Division of Pediatric Cardiology, Boston Floating Hospital for Children-New England Medical Center, Tufts University School of Medicine, Boston, Massachusetts

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A

10:55:42

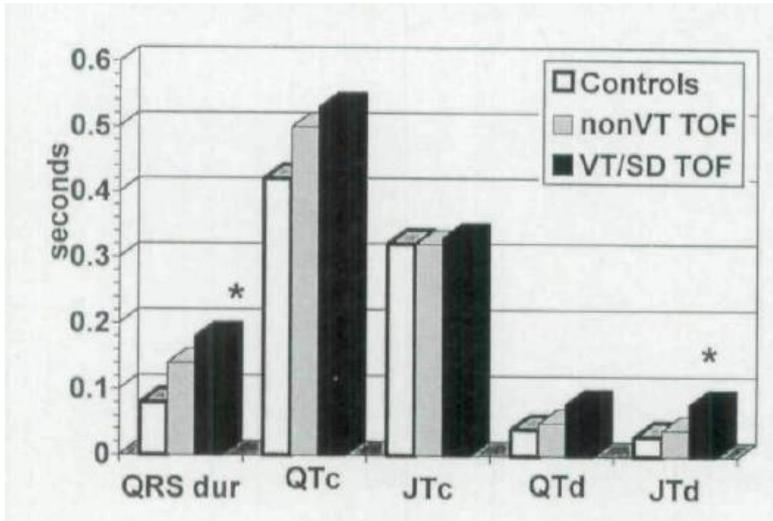


B

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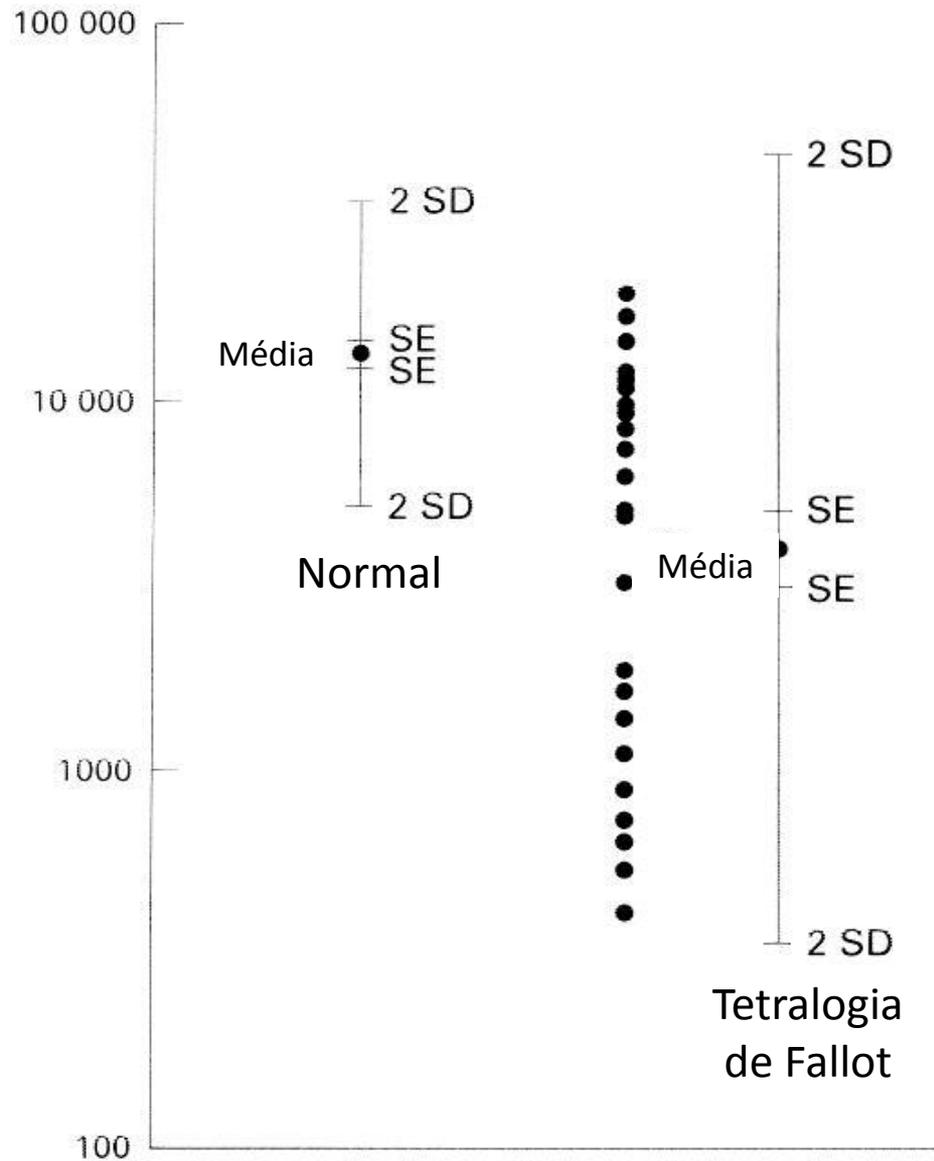
From the Division of Pediatric Cardiology, Boston Floating Hospital for Children-New England Medical Center, Tufts University School of Medicine, Boston, Massachusetts



**TABLE 1**  
Electrocardiographic Findings

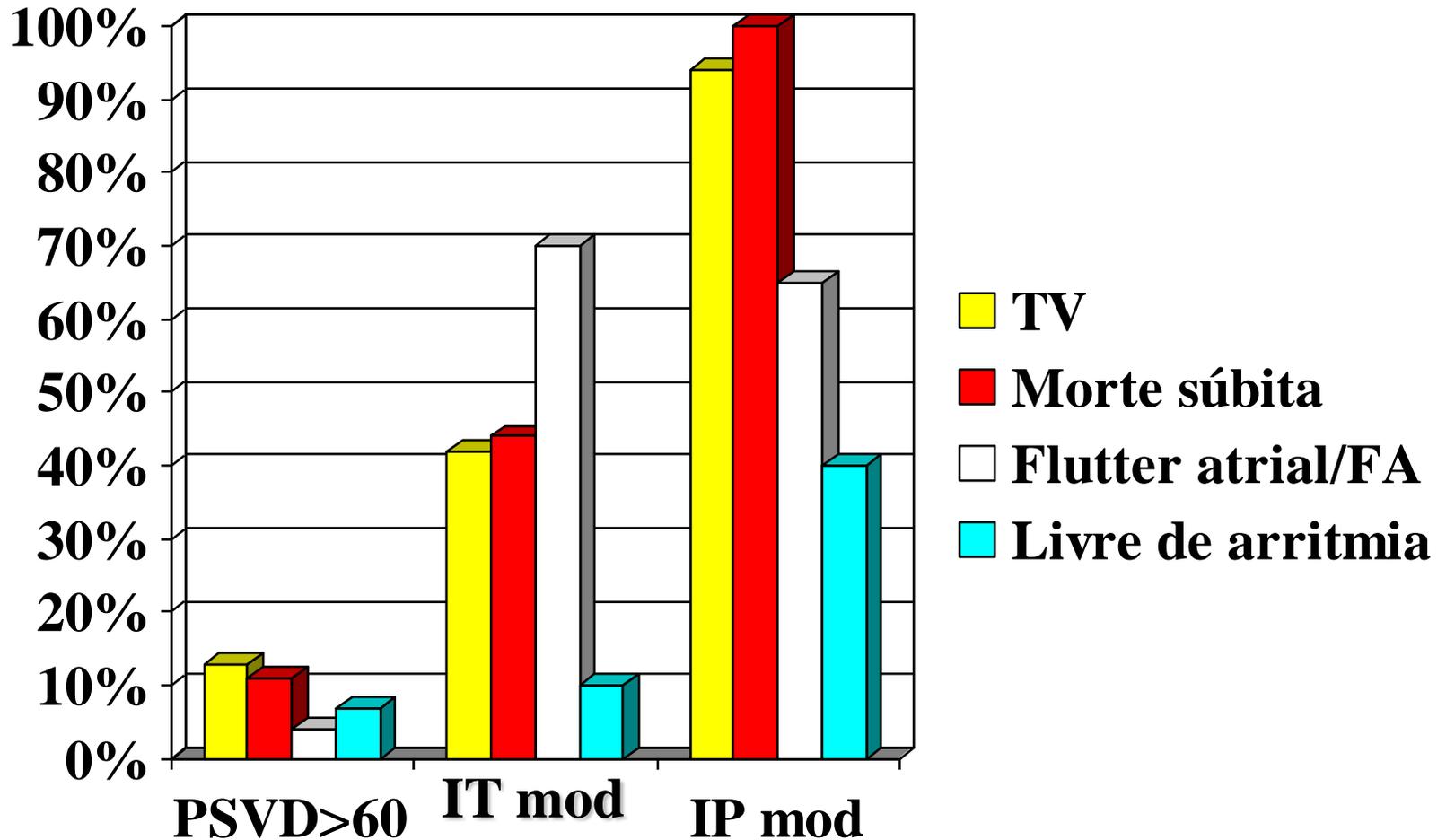
		Age	QRS	JT	JTc	QTc	JTd	QTd
Control Group (n = 1000)	Mean	12.5	0.08	0.23	0.32	0.42	0.03	0.04
	SD	4.7	0.01	0.04	0.02	0.02	0.02	0.02
TOF non-VT (n = 87)	Mean	11.1	0.14*	0.25	0.32	0.50*	0.04	0.05
	SD	5.7	0.02	0.03	0.03	0.03	0.02	0.03
TOF VT (n = 14)	Mean	14.9	0.18#	0.26	0.33	0.53*	0.08#	0.08*
	SD	4.7	0.02	0.03	0.03	0.05	0.03	0.03

# Variabilidade da FC na Tetralogia de Fallot



# Arritmias no Fallot

Avaliação ecocardiográfica de 456 pacientes – pré eventos

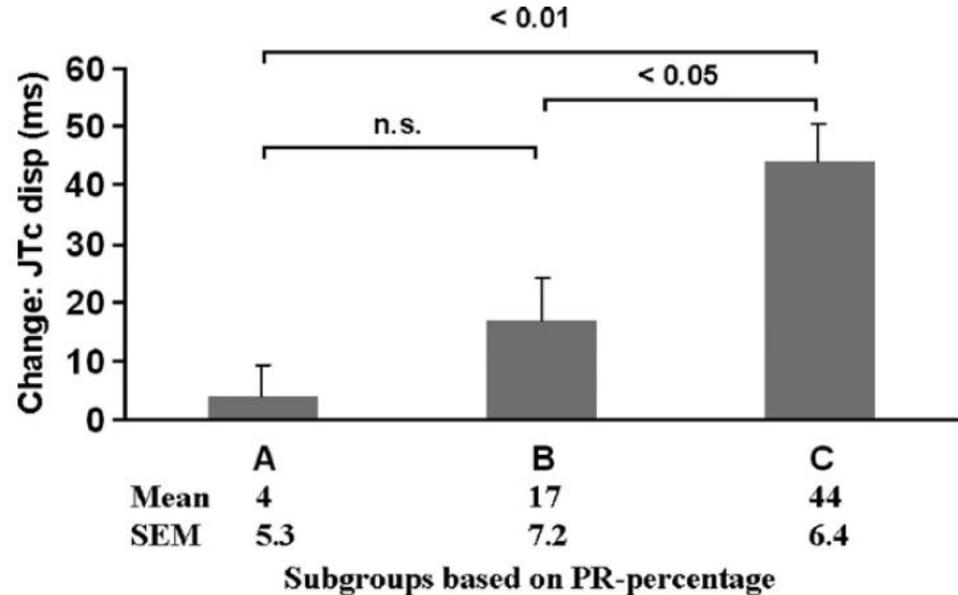
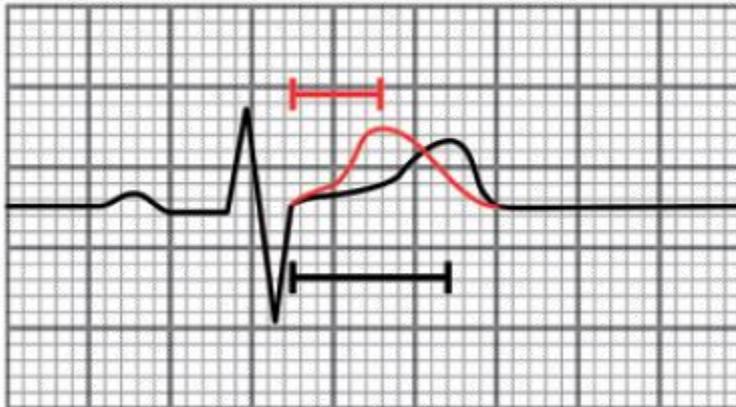


## Changes during exercise of ECG intervals related to increased risk for ventricular arrhythmia in repaired tetralogy of Fallot and their relationship to right ventricular size and function

Jochem van den Berg <sup>a,b</sup>, Sandra de Bie <sup>a</sup>, Folkert J. Meijboom <sup>c</sup>, Wim C. Hop <sup>d</sup>, Peter M.T. Pattynama <sup>b</sup>, Ad J.J.C. Bogers <sup>e</sup>, Willem A. Helbing <sup>a,b,\*</sup>

**Quanto maior a IP maior a dispersão da repolarização no esforço**

Grupo A – IP leve  
Grupo B – IP moderada  
Grupo C – IP grave

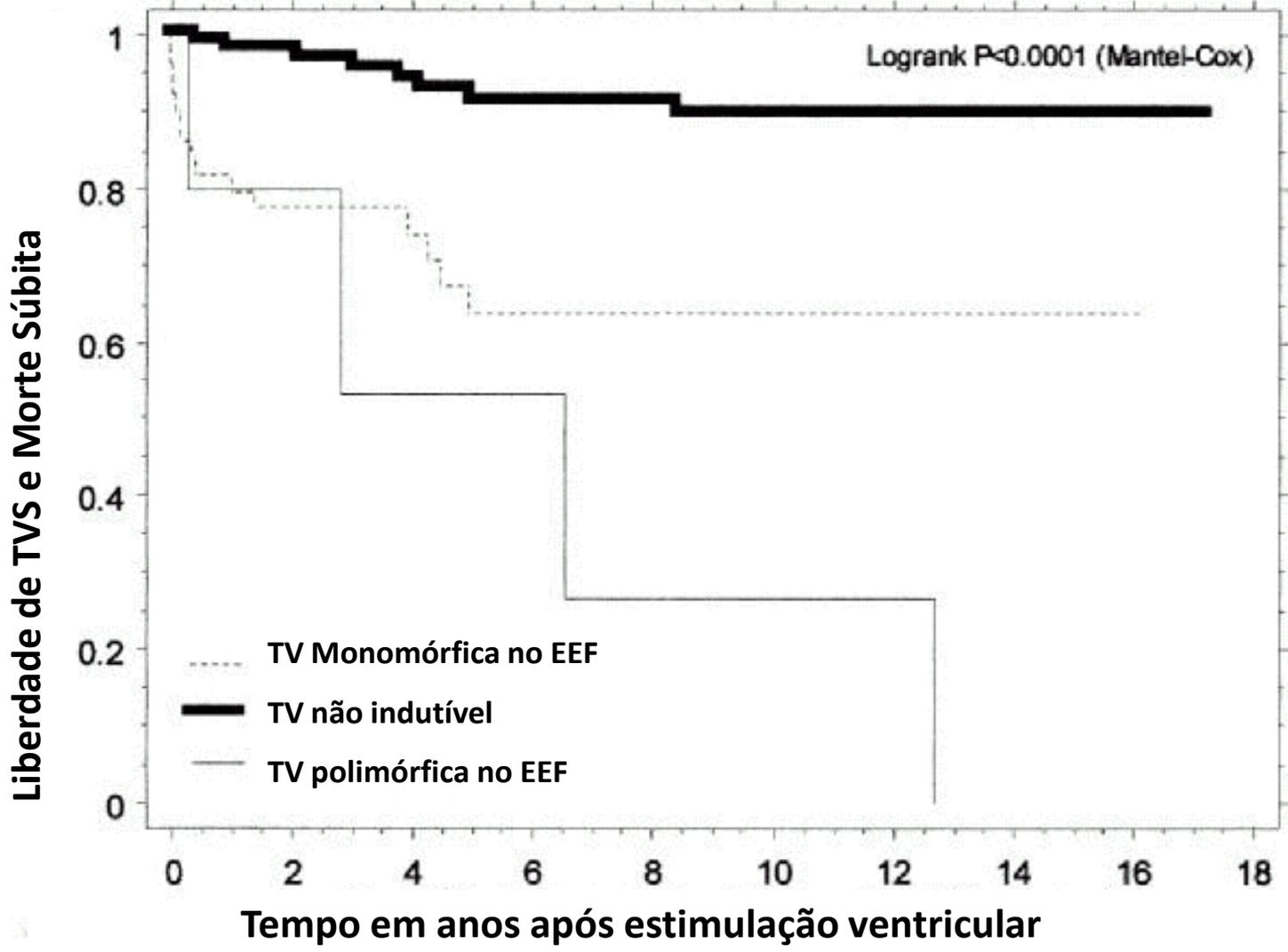


**Table 3** Predictors of appropriate and inappropriate therapies in adults with CHD (*n* = 64)

Variable	Appropriate shock therapy		Inappropriate shock therapy	
	HR (95% CI)	<i>P</i> -value	HR (95% CI)	<i>P</i> -value
Age at ICD implantation	0.96 (0.92–1.01)	0.12	0.98 (0.95–1.02)	0.29
Male gender	1.09 (0.37–3.20)	0.88	1.49 (0.62–3.57)	0.38
BMI >25	0.43 (0.14–1.34)	0.14	1.23 (0.55–2.75)	0.61
QRS width >180 ms	2.00 (0.60–6.70)	0.26	0.77 (0.26–2.34)	0.65
History of atrial arrhythmias	0.97 (0.33–2.84)	0.95	1.29 (0.57–2.88)	0.54
TOF	0.29 (0.10–0.84)	0.02	0.77 (0.34–1.76)	0.53
Single-chamber ICD device	1.68 (0.55–5.14)	0.36	1.90 (0.85–4.25)	0.12
Secondary prophylaxis <sup>a</sup>	0.47 (0.16–1.38)	0.17	1.75 (0.73–4.21)	0.21
Positive PES <sup>b</sup>	1.96 (0.25–15.4)	0.52	0.69 (0.19–2.48)	0.57
Impaired subpulmonary ventricular function	1.28 (0.42–3.84)	0.66	1.19 (0.50–2.84)	0.70
Impaired systemic ventricular function	2.34 (0.62–8.87)	0.21	1.22 (0.36–4.15)	0.76

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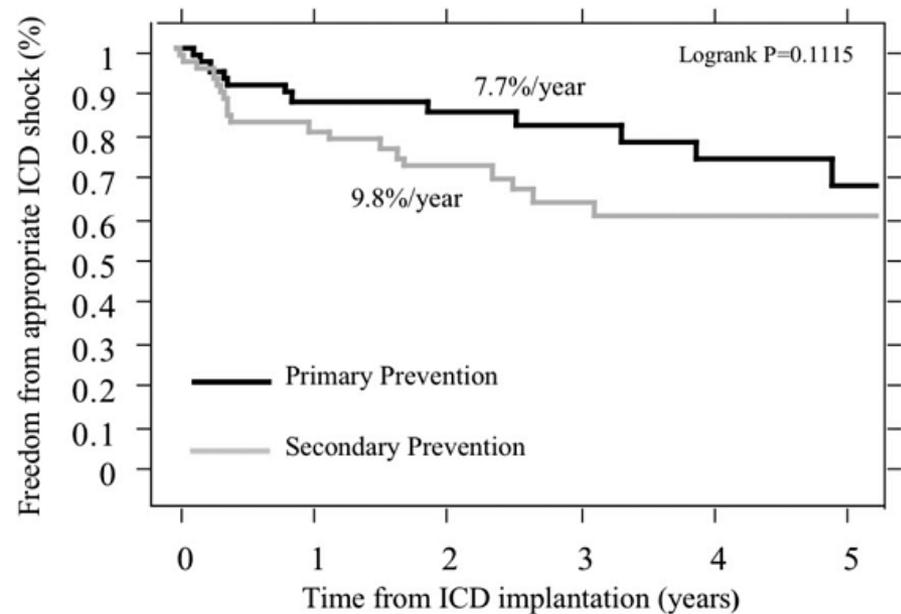
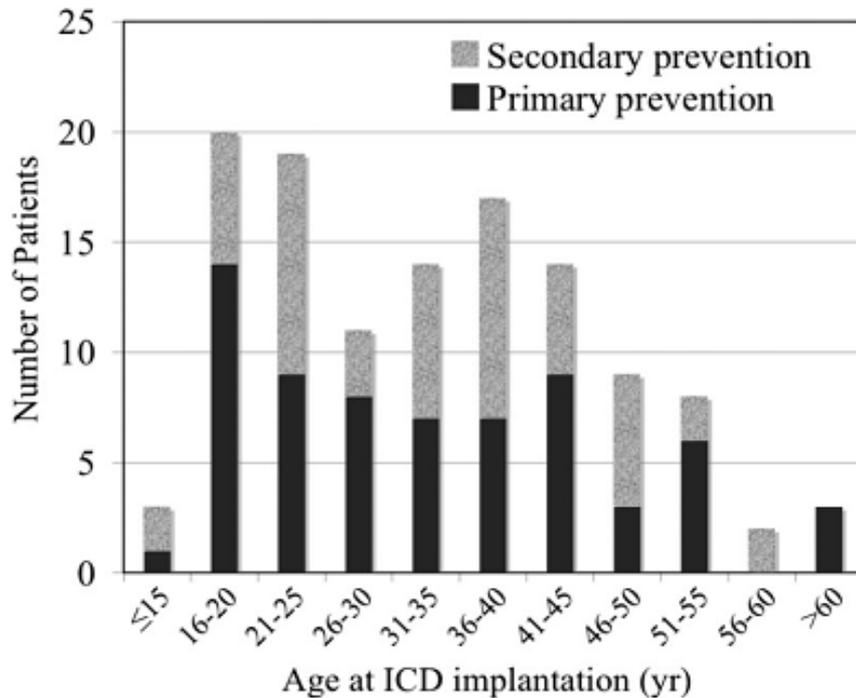
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Value of Programmed Ventricular Stimulation After Tetralogy of Fallot Repair-  
Circulation, 2004; 109: 1994-2000

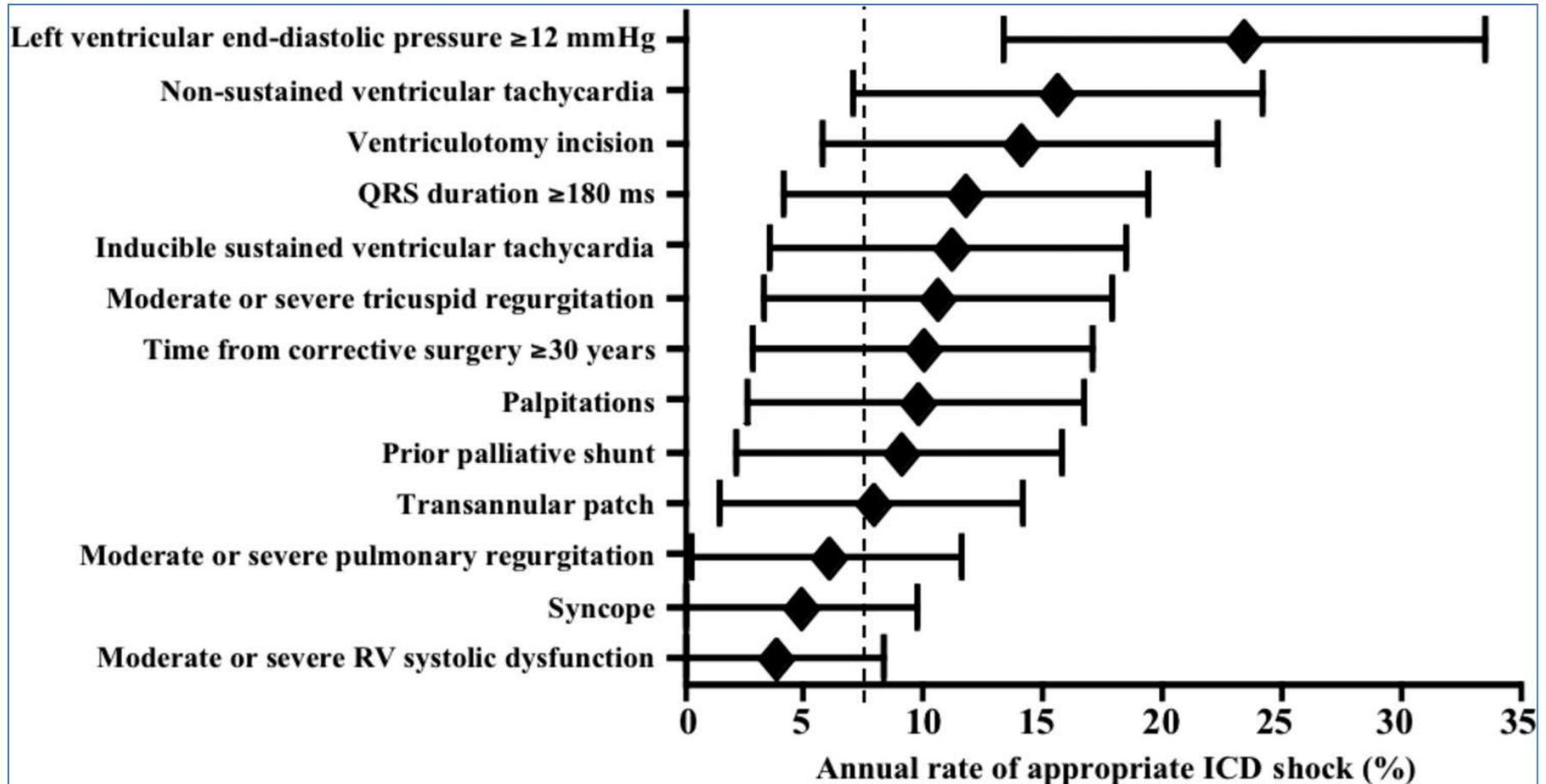
# Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

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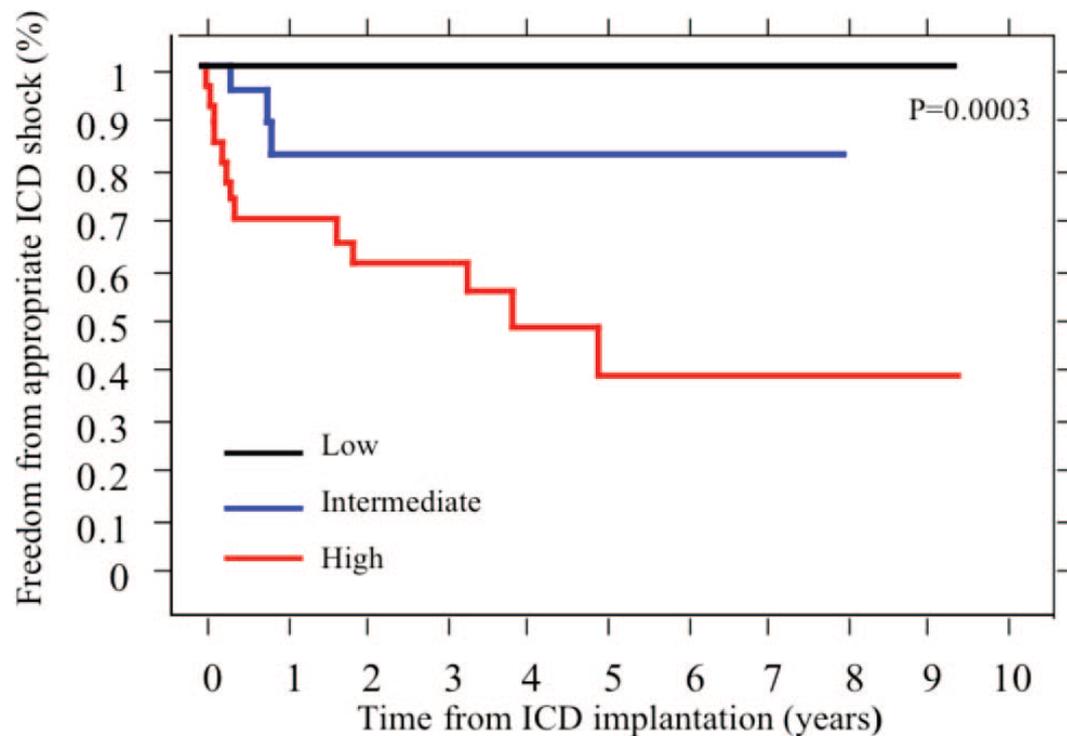
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**Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention**

Variable	Exp( $\beta$ )	Points Attributed
Prior palliative shunt	3.2	2
Inducible sustained ventricular tachycardia	2.6	2
QRS duration $\geq$ 180 ms	1.4	1
Ventriculotomy incision	3.4	2
Nonsustained ventricular tachycardia	3.7	2
LVEDP $\geq$ 12 mm Hg	4.9	3
Total points	...	0–12

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Susan M. Fernandes, MHP, PA-C; Luc Beauchesne, MD; Judith Therrien, MD; Philippe Chetaille, MD;  
Elaine Gordon, MD; Isabelle Vonder Muhll, MD; Frank Cecchin, MD



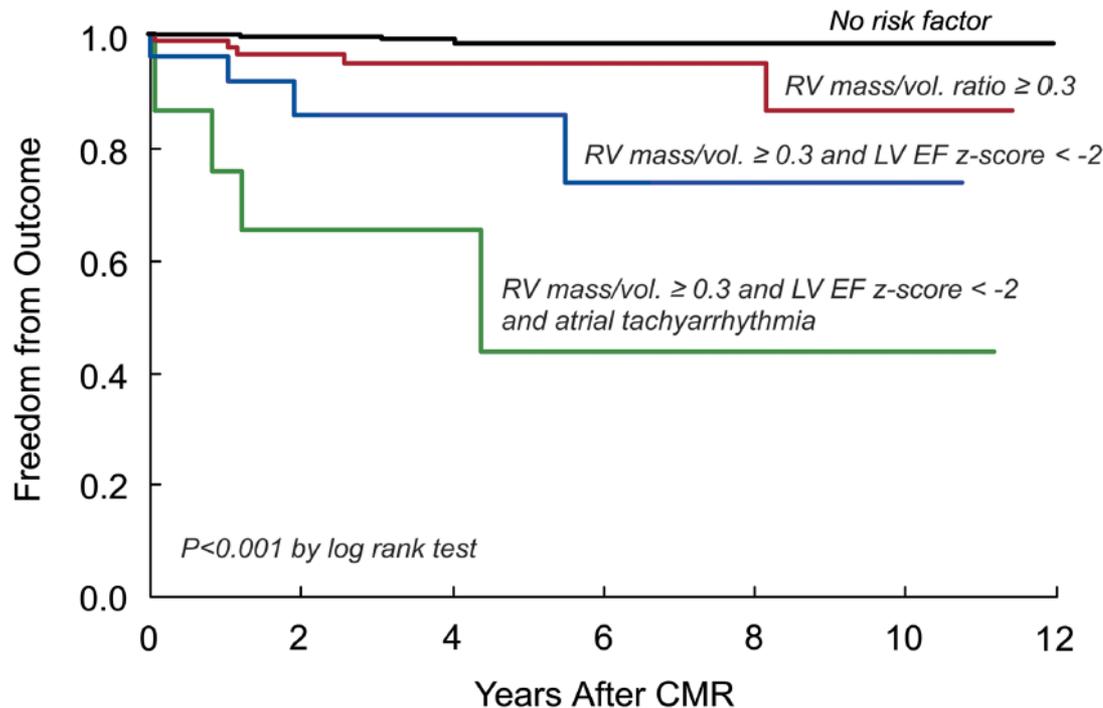
Risk score	Risk category	N	Annualized rate of appropriate shocks
0-2	Low	18	0%
3-5	Intermediate	24	3.8%
6-12	High	26	17.5%

# Contemporary predictors of death and sustained ventricular tachycardia in patients with repaired tetralogy of Fallot enrolled in the INDICATOR cohort

Anne Marie Valente,<sup>1</sup> Kimberlee Gauvreau,<sup>1</sup> Gabriele Egidy Assenza,<sup>1</sup> Sonya V Babu-Narayan,<sup>2</sup> Jenna Schreier,<sup>1</sup> Michael A Gatzoulis,<sup>2</sup> Maarten Groenink,<sup>3</sup> Ryo Inuzuka,<sup>2</sup> Philip J Kilner,<sup>2</sup> Zeliha Koyak,<sup>3</sup> Michael J Landzberg,<sup>1</sup> Barbara Mulder,<sup>3</sup> Andrew J Powell,<sup>1</sup> Rachel Wald,<sup>4</sup> Tal Geva<sup>1</sup>

873 pacientes  
Idade média 24,4 anos  
Média de seguimento 38 anos

Valente AM, et al. *Heart* 2014;**100**:247–253. doi:10.1136/heartjnl-2013-304958



## Mensagens

- Mortalidade aumenta entre adultos jovens
- Hipertrofia ventricular, disfunção VE ou VE e arritmias atriais aumentam mortalidade

# Morte súbita



# Evolução



209 cc



113 cc



80 cc



80 cc



72 cc



54 cc



62 cc



49 cc



39.5 cc



39 cc



39.5 cc



39 cc



39.5 cc

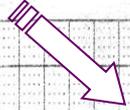


36 cc

Dec 18, 2003 13:47:23

CHART SPEED 25.0 mm/s

LEAD II 0.2 mV/mm

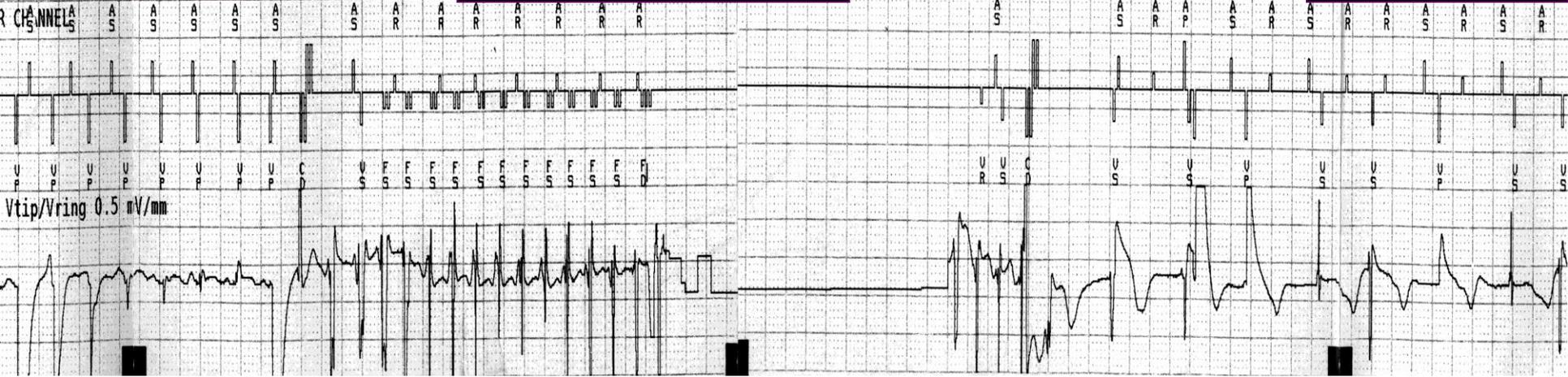


**Indução**

**Fibrilação ventricular**

**Choque**

**Reversão**



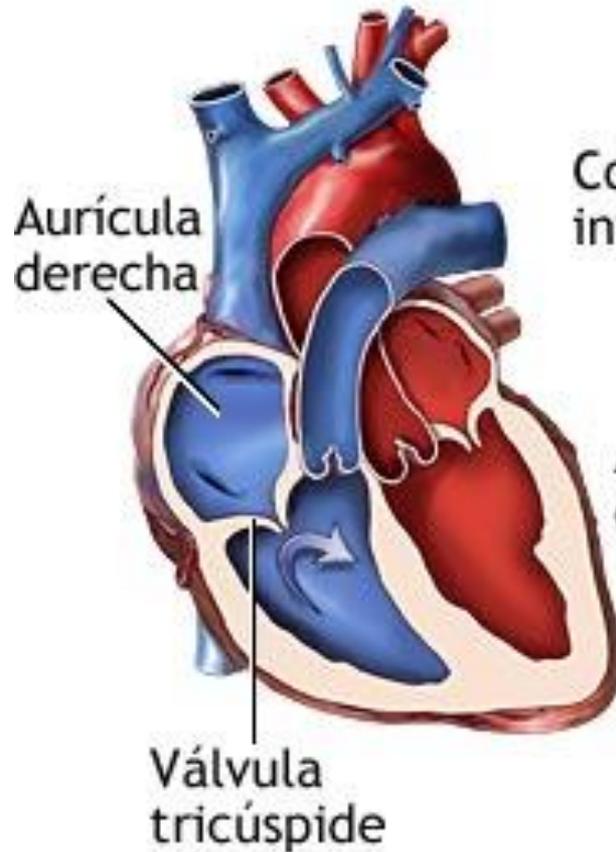
# Marcadores ECG na população pediátrica e em jovens



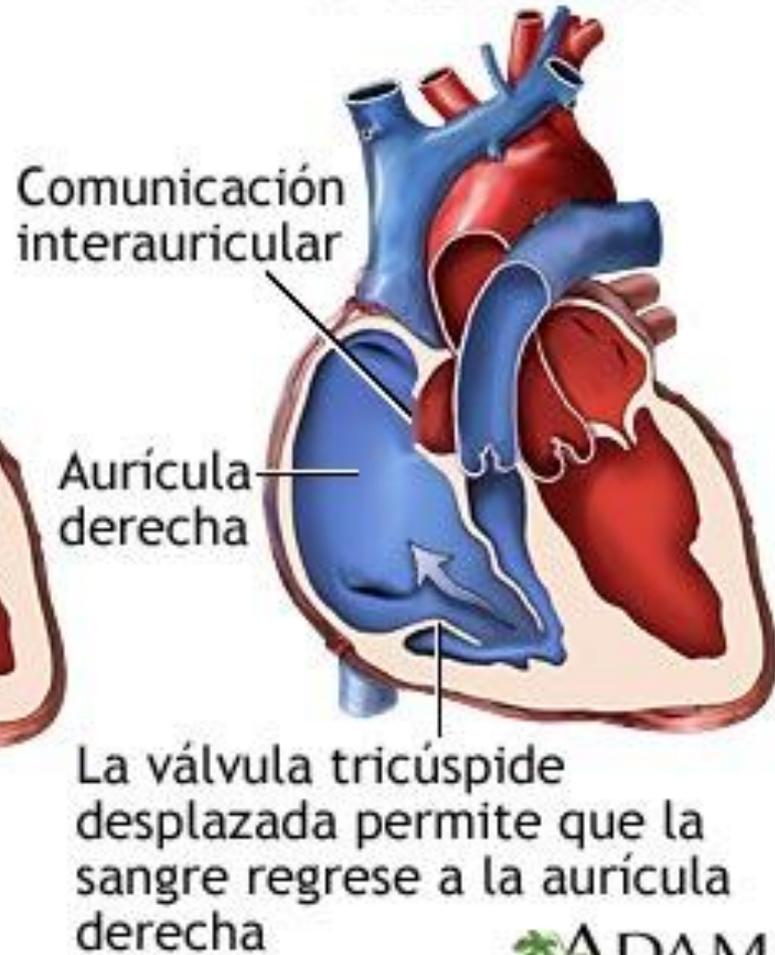
# Marcadores ECG na população pediátrica e em jovens



Corazón normal



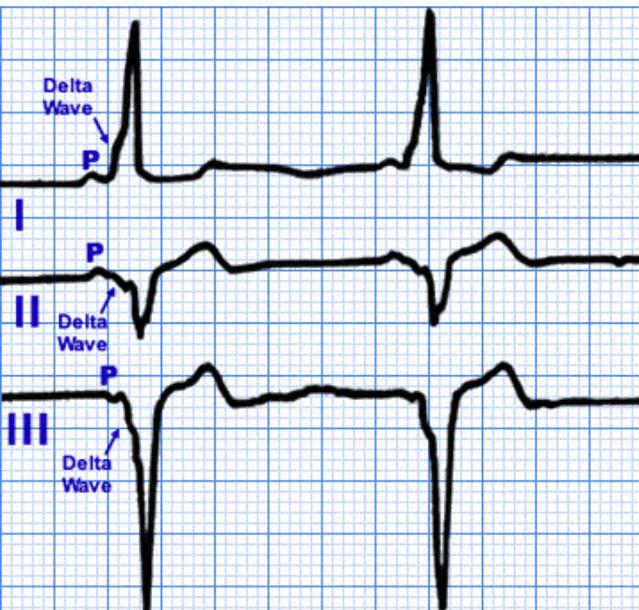
Anomalia de Ebstein



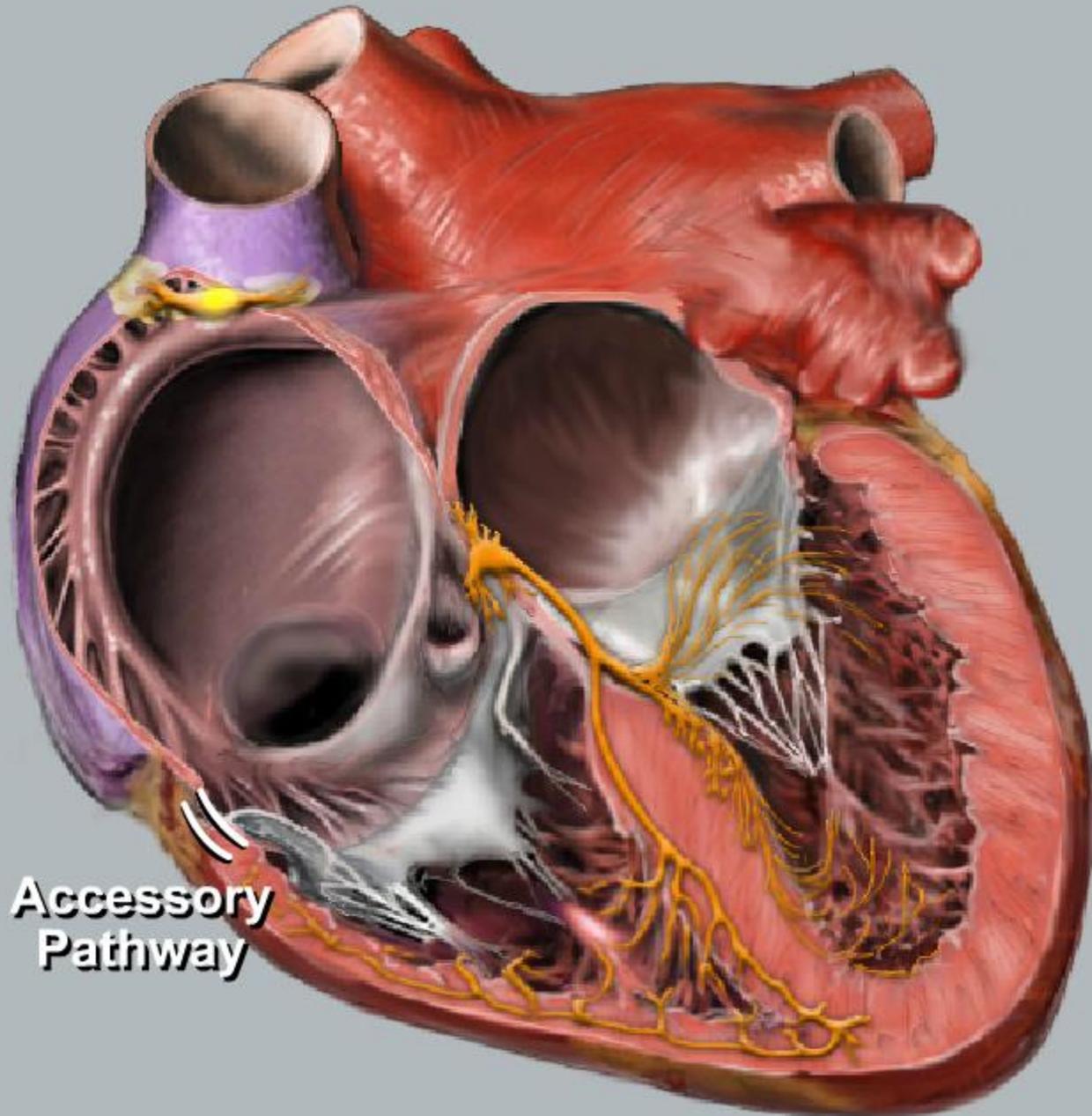
# Pré excitação ventricular

## Padrões do ECG

- PR curto < 120 ms
- Excluir ritmo juncional
- Presença de onda Delta
- Duração QRS > 100 ms



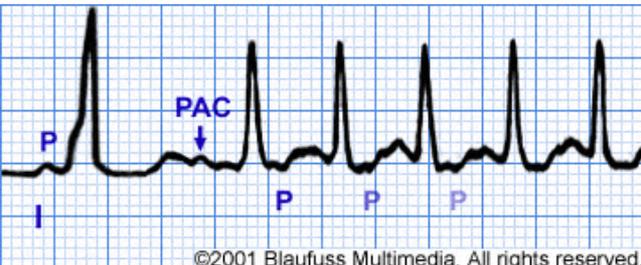
©2001 Blaufuss Multimedia. All rights reserved.



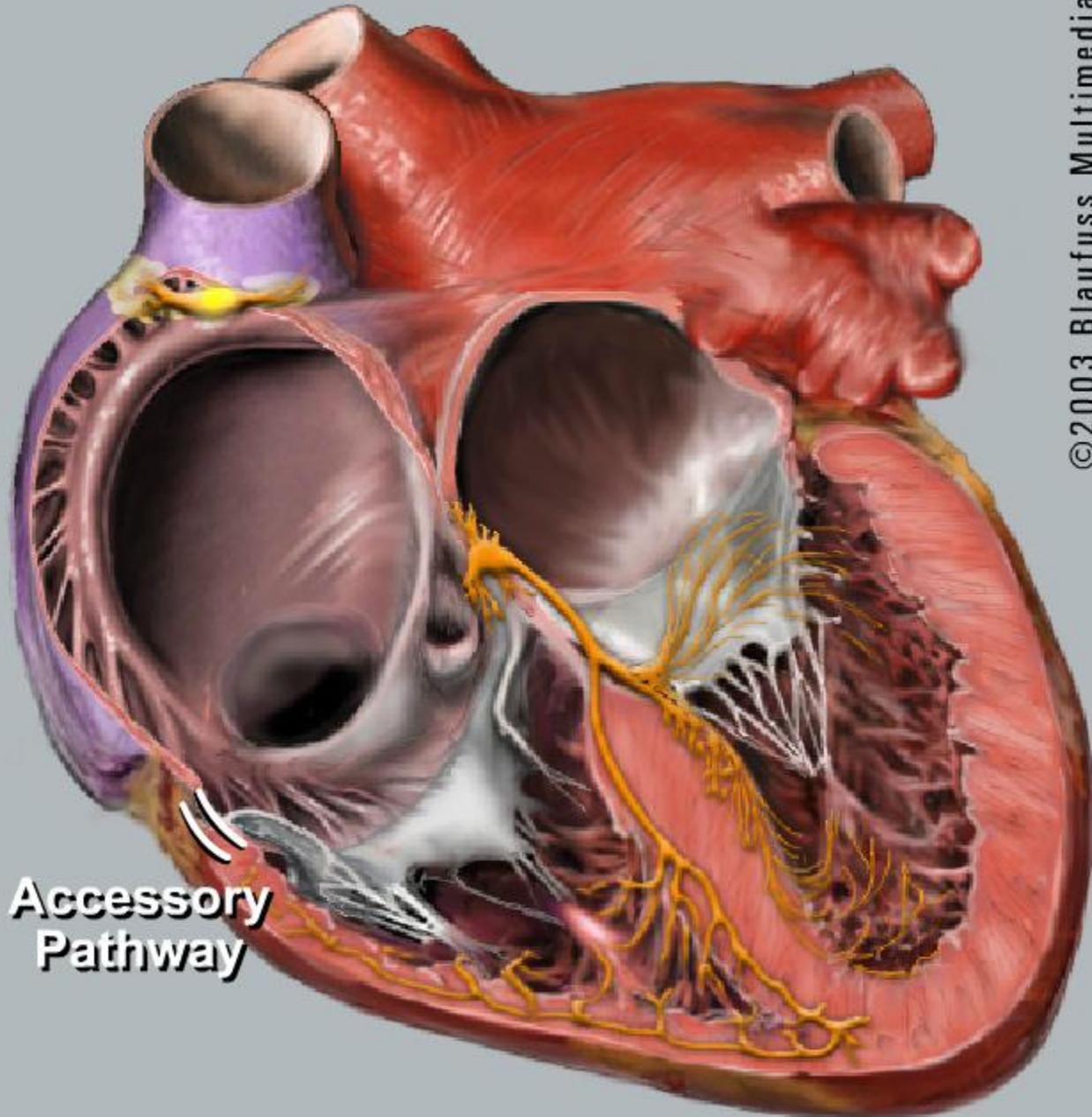
# Início da taquicardia no WPW

## Taquicardia por reentrada

- Pode ser iniciado por
- Bloqueio da via acessória
- Condução pelo NAV
- Condução retrógrada pela via acessória
- Onda P retrógrada visível

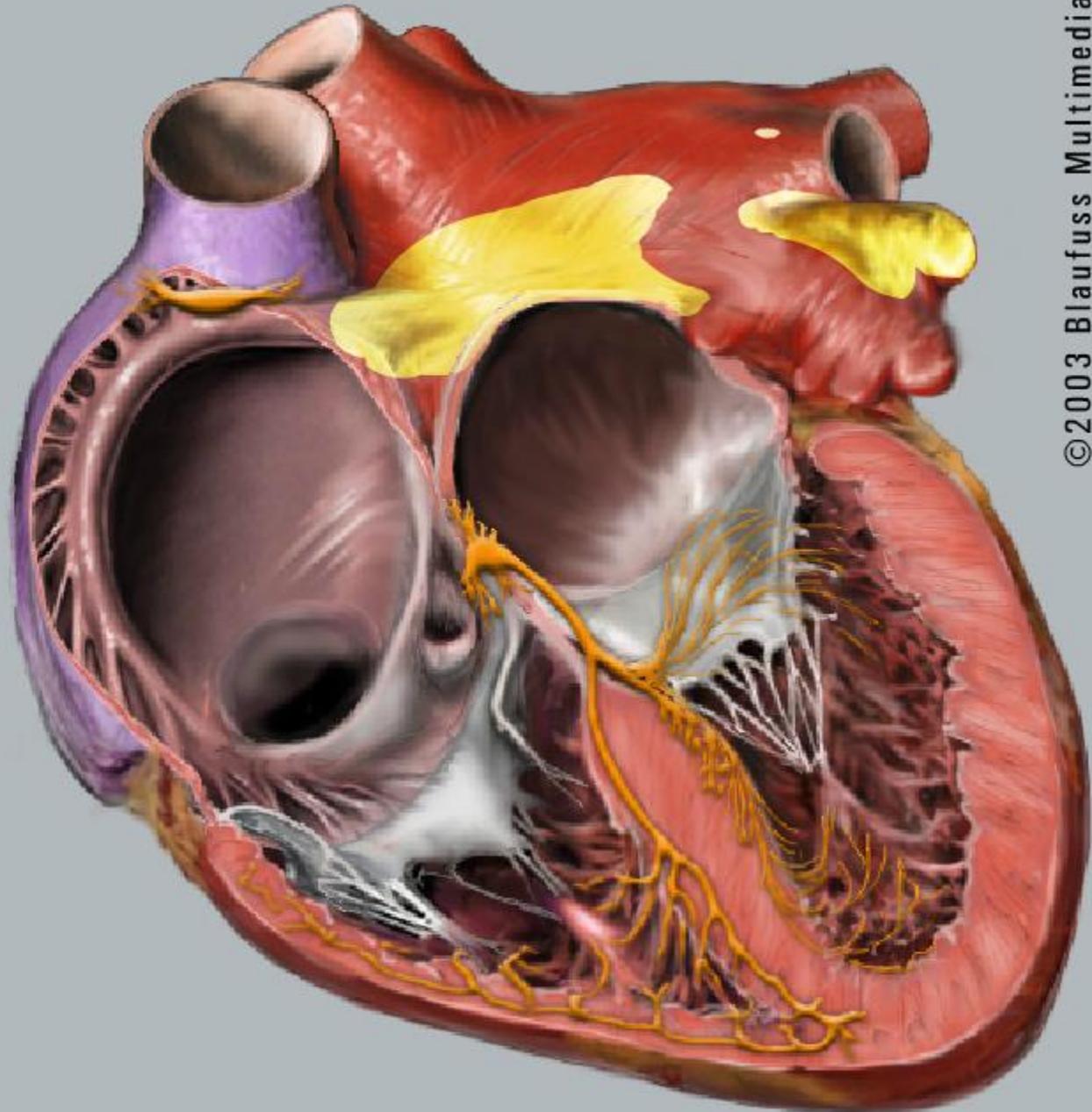


Accessory Pathway



# FA + pré excitação

**Condução AV rápida  
inferior a 240 ms risco  
real de morte súbita**



# Taquicardias automatismo

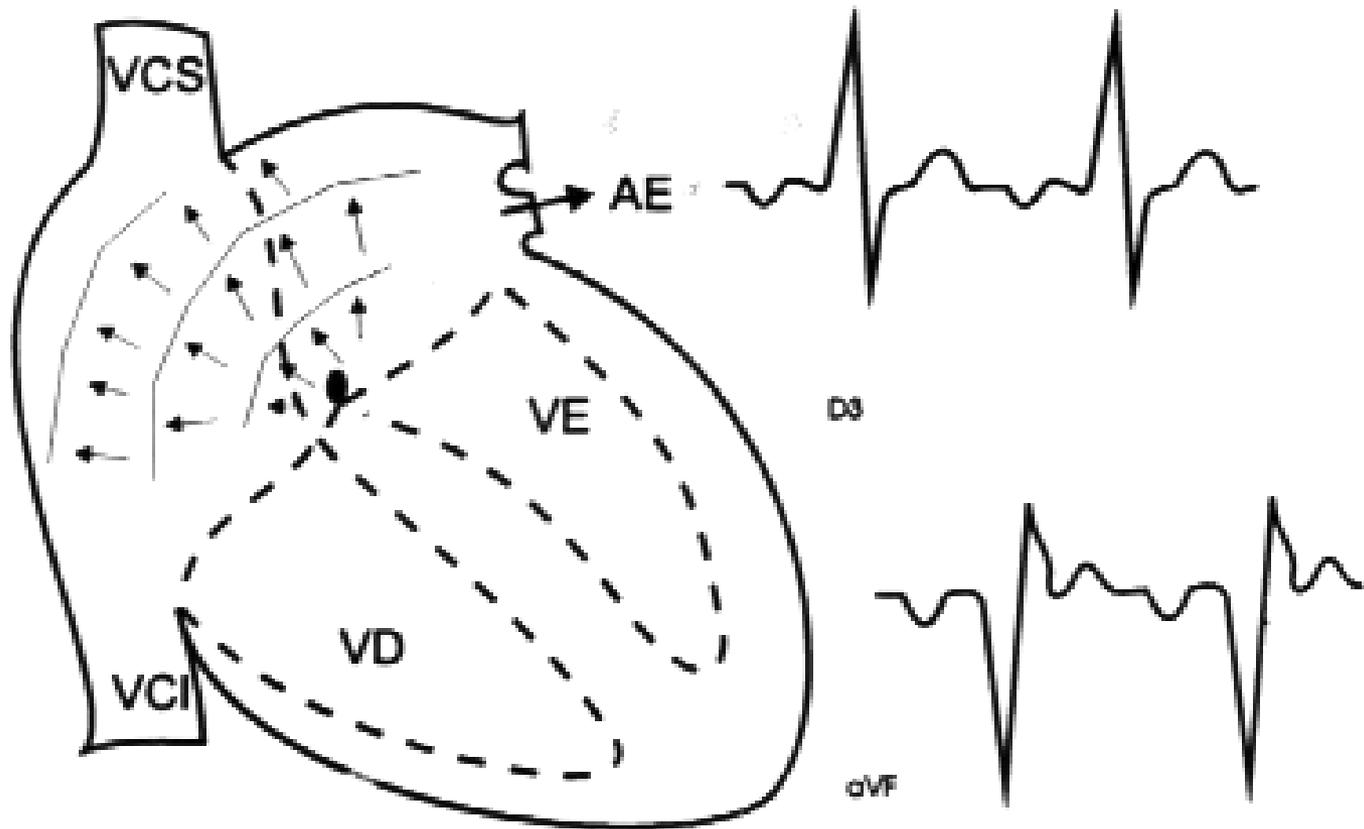
➤ **Taquicardia sinusal**

➤ **Taquicardia atrial**

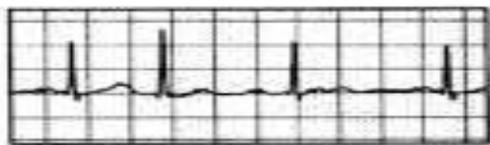
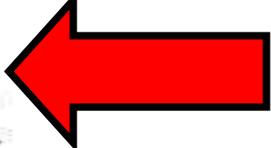
➤ **Taquicardia juncional**

# Taquicardia Atrial

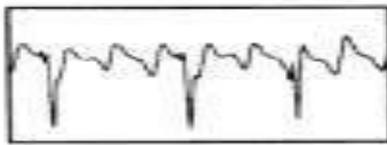
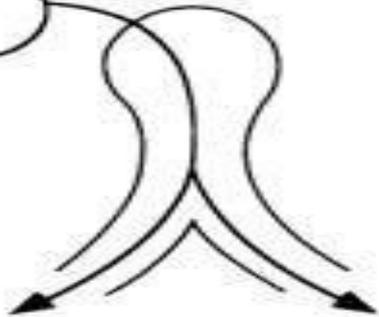
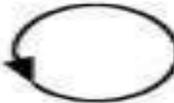
## TAQUICARDIA ATRIAL ECTÓPICA



**G. EAT**



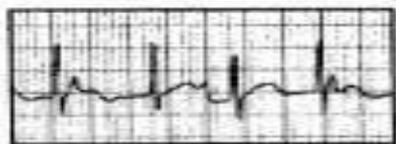
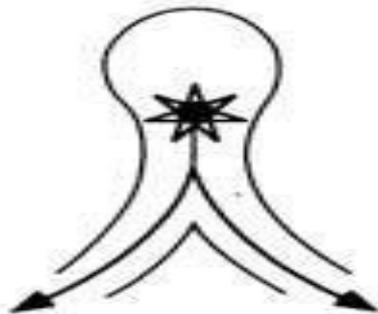
**H. IART**



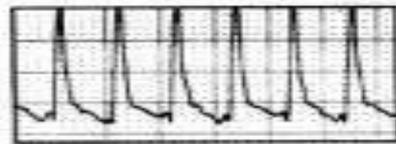
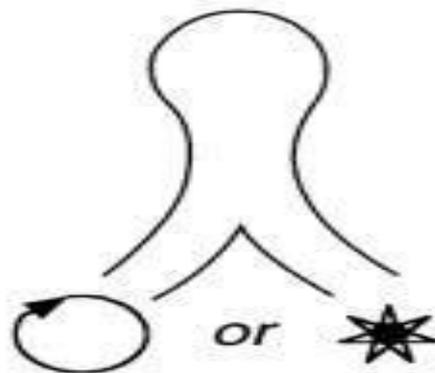
**I. AVNRT**

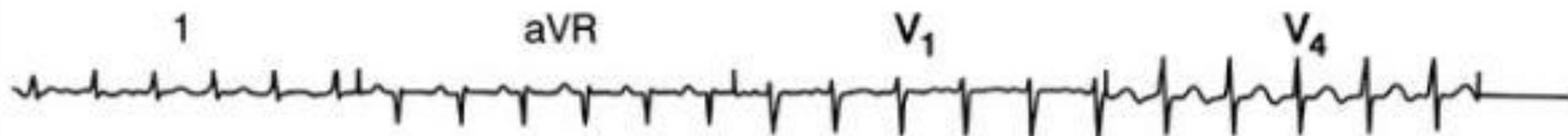


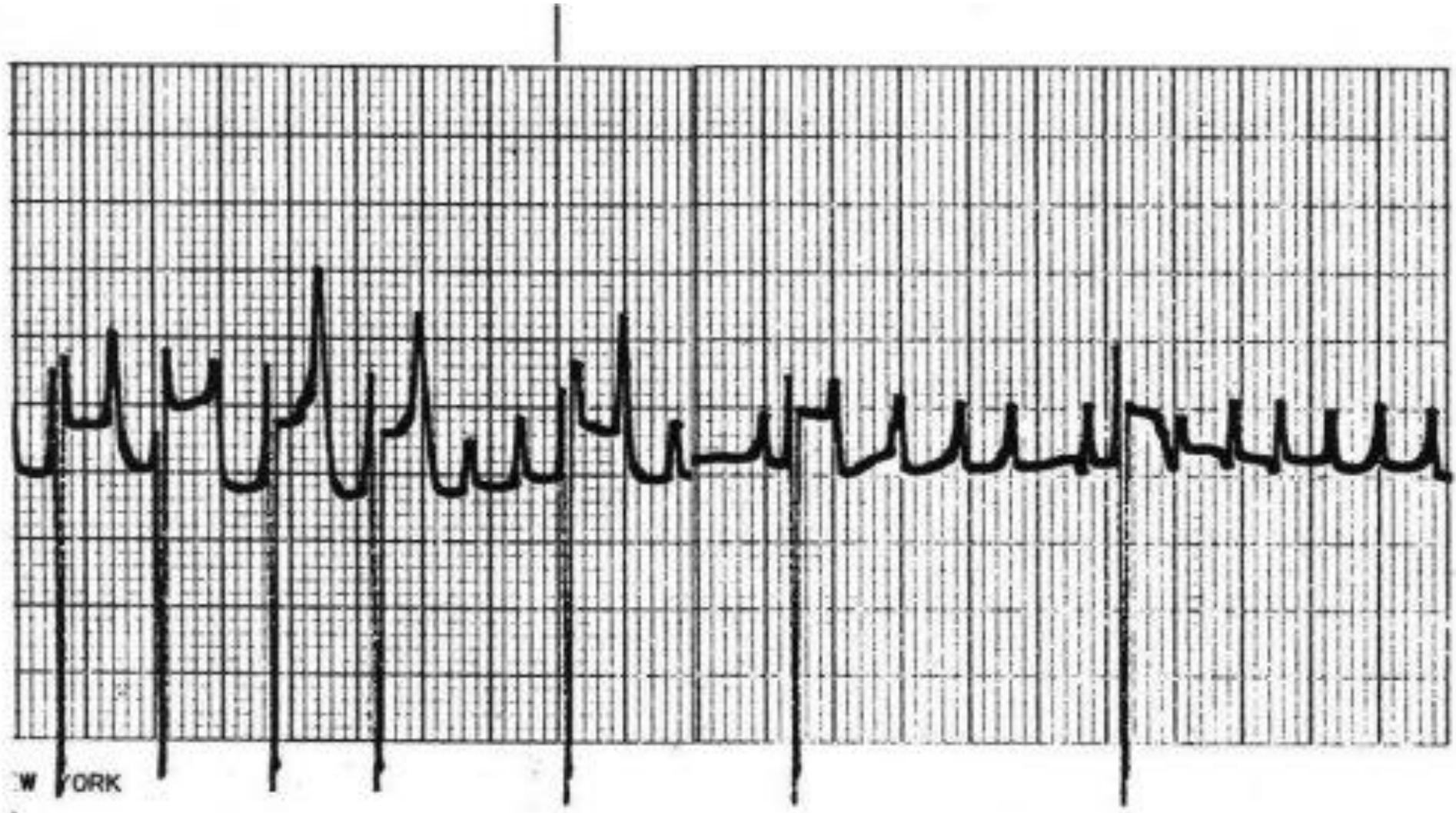
**J. JET**



**K. VT**



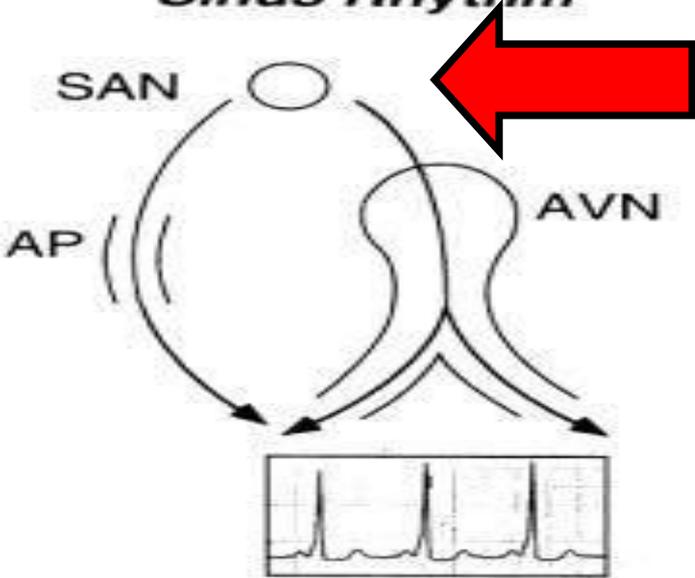




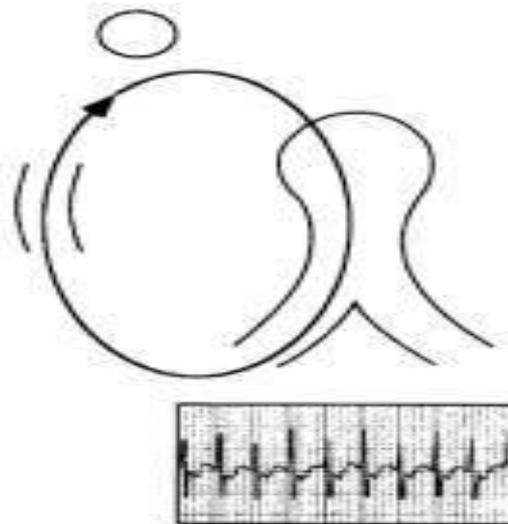
W YORK

**Pré excitação + taquicardia = WPW**

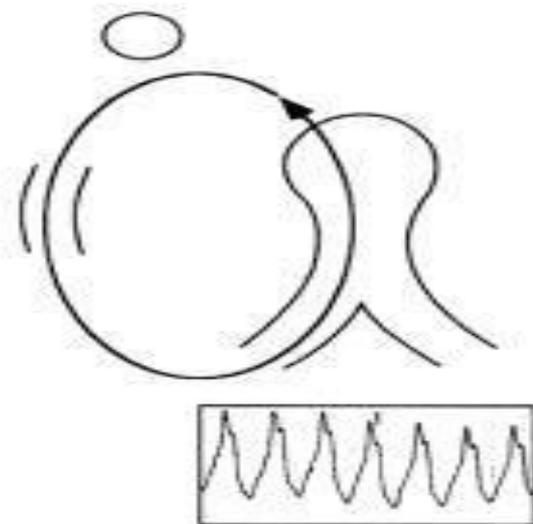
**A. WPW - Sinus Rhythm**



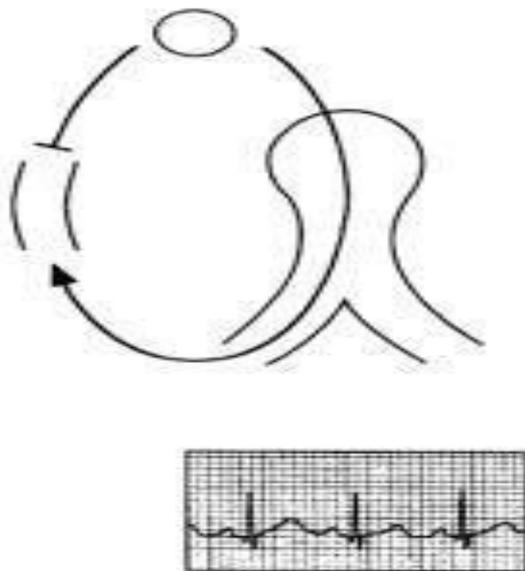
**B. ORT**



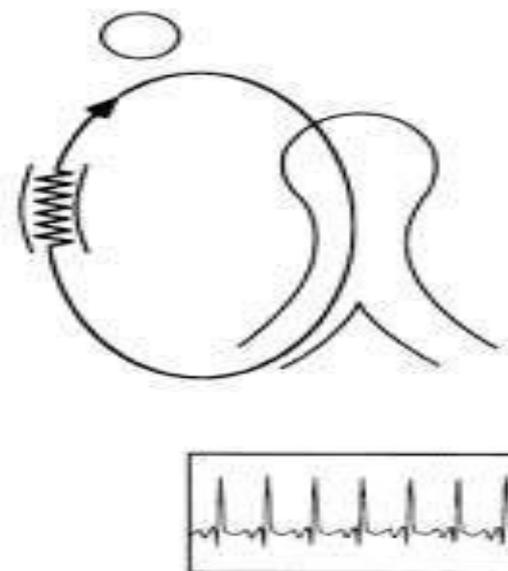
**C. ART**



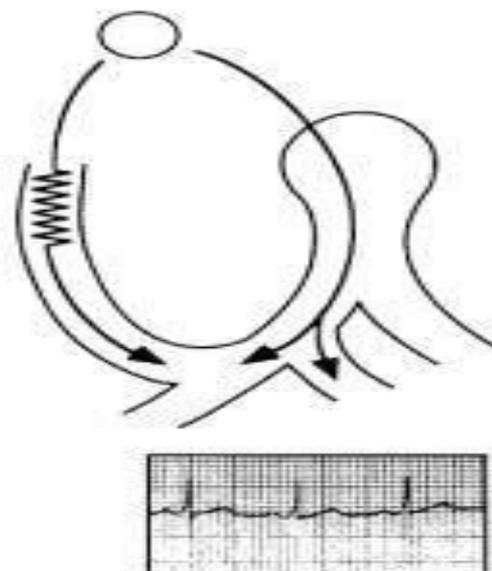
**D. Concealed AP**



**E. PJRT**



**F. Mahaim**

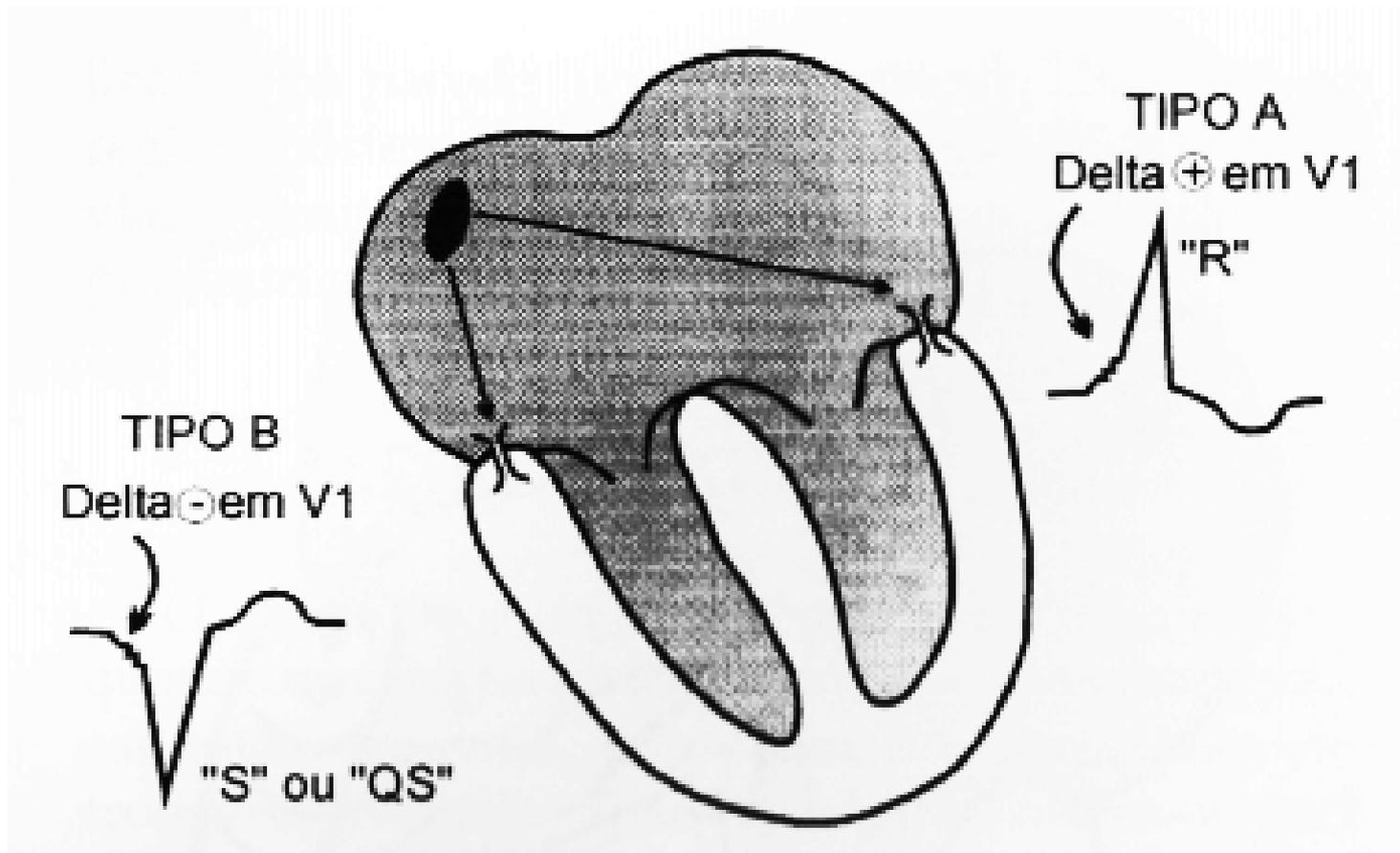


# Anomalia Ebstein e WPW

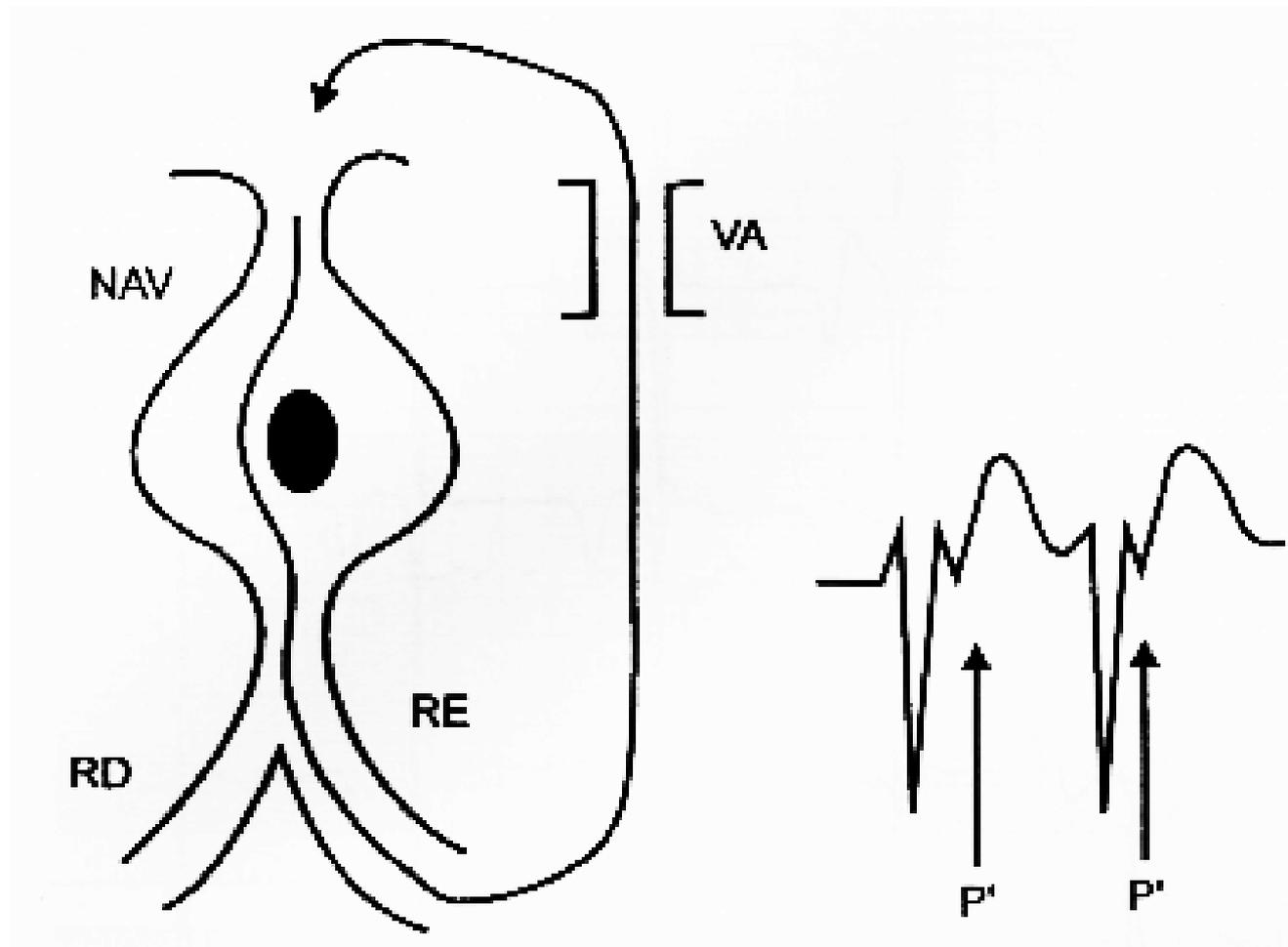
- ✓ 20 a 30% dos pacientes apresentam TRAV
- ✓ Vias acessórias a direita
- ✓ 50% dos casos multiplas vias

# Pré-Excitação Ventricular

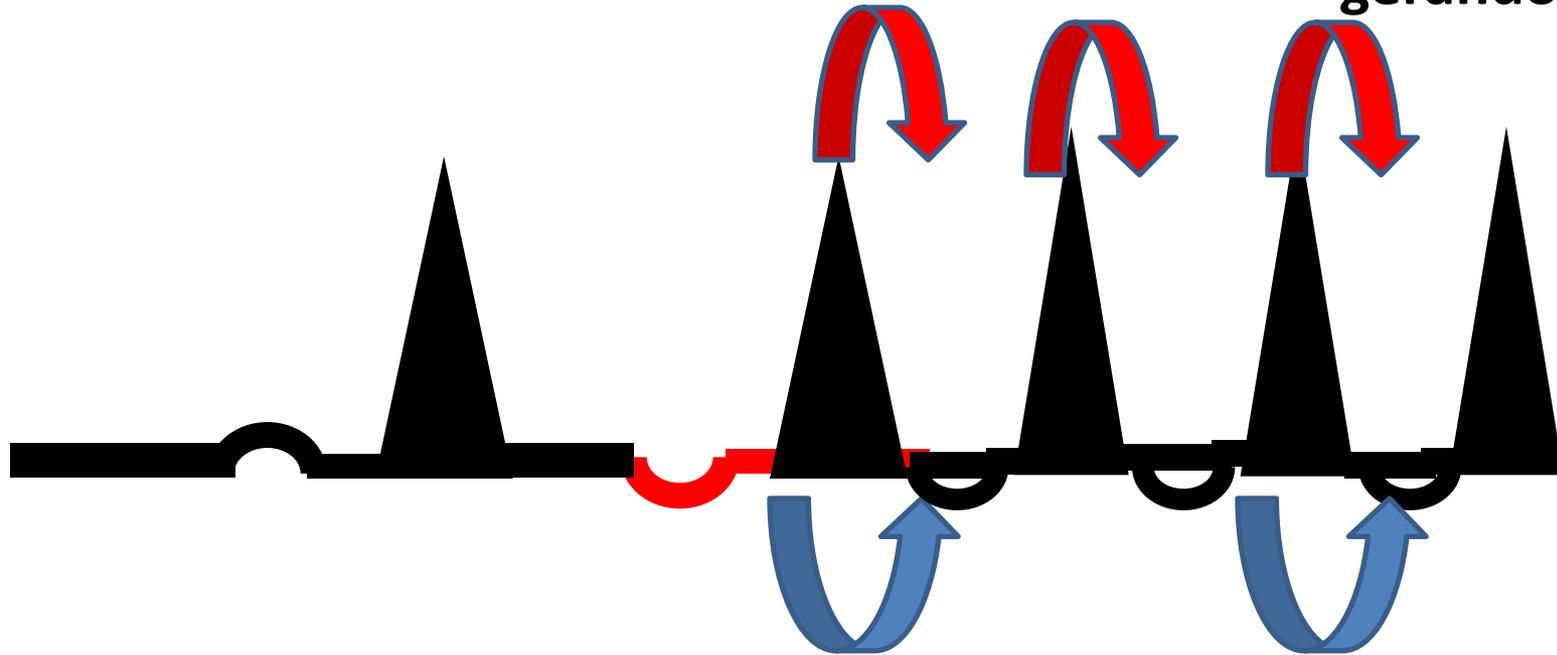
## Classificação Eletrocardiográfica



# Pré-Excitação Ventricular TPSV Ortodrômica

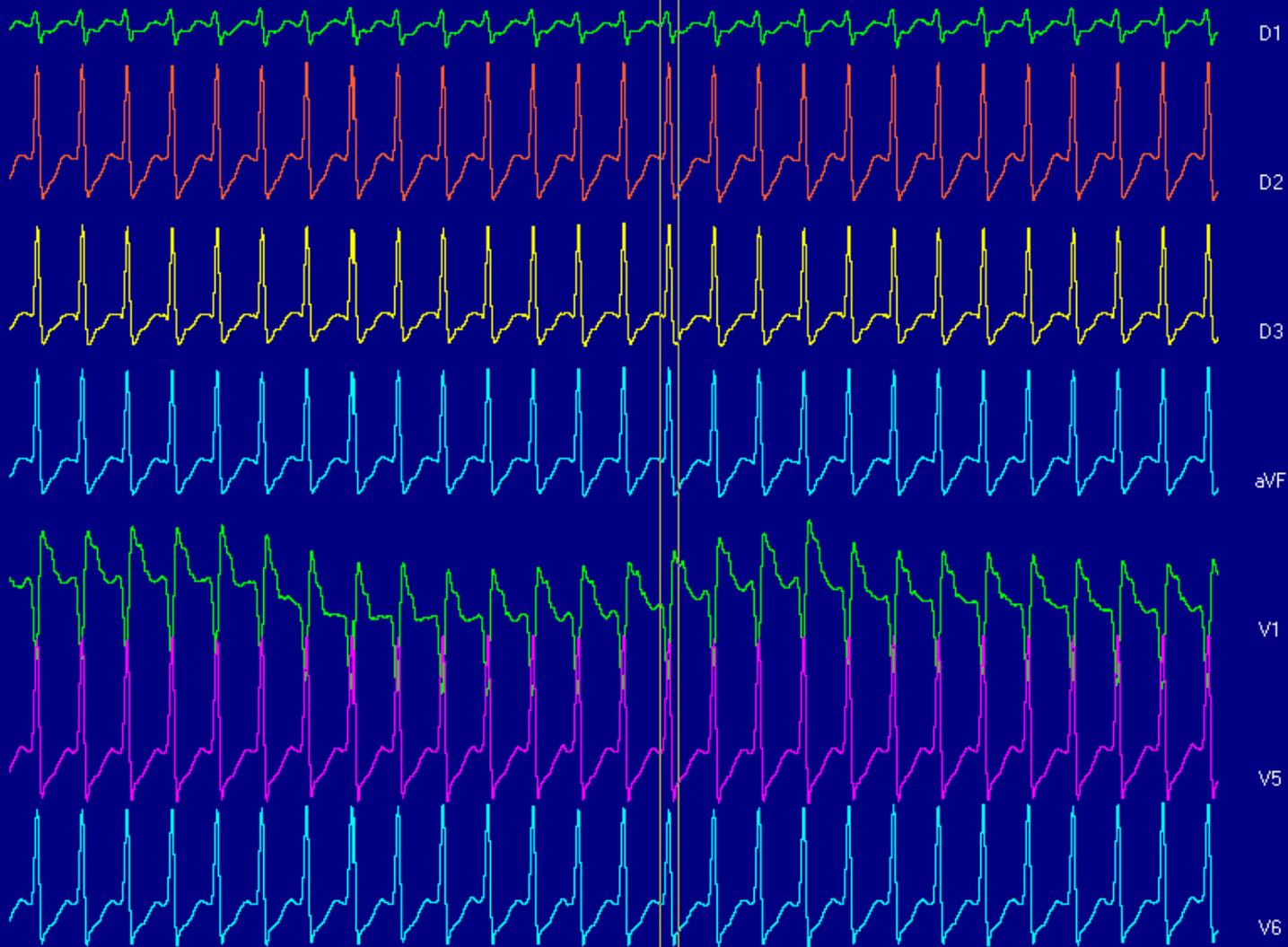


**Complexos QRS  
gerando P retrógrada**



**Início QRS início p  
retrógrada maior  
que 70 ms**

t= 110



**TEB**<sup>®</sup>

ECGPC Versão 2.19

Título:

TRECHO 11

Janela N

25

< Adjacente >

Imprimir Janela

Imprimir por derivação:  
Use F1 a F12

Arquivo BMP

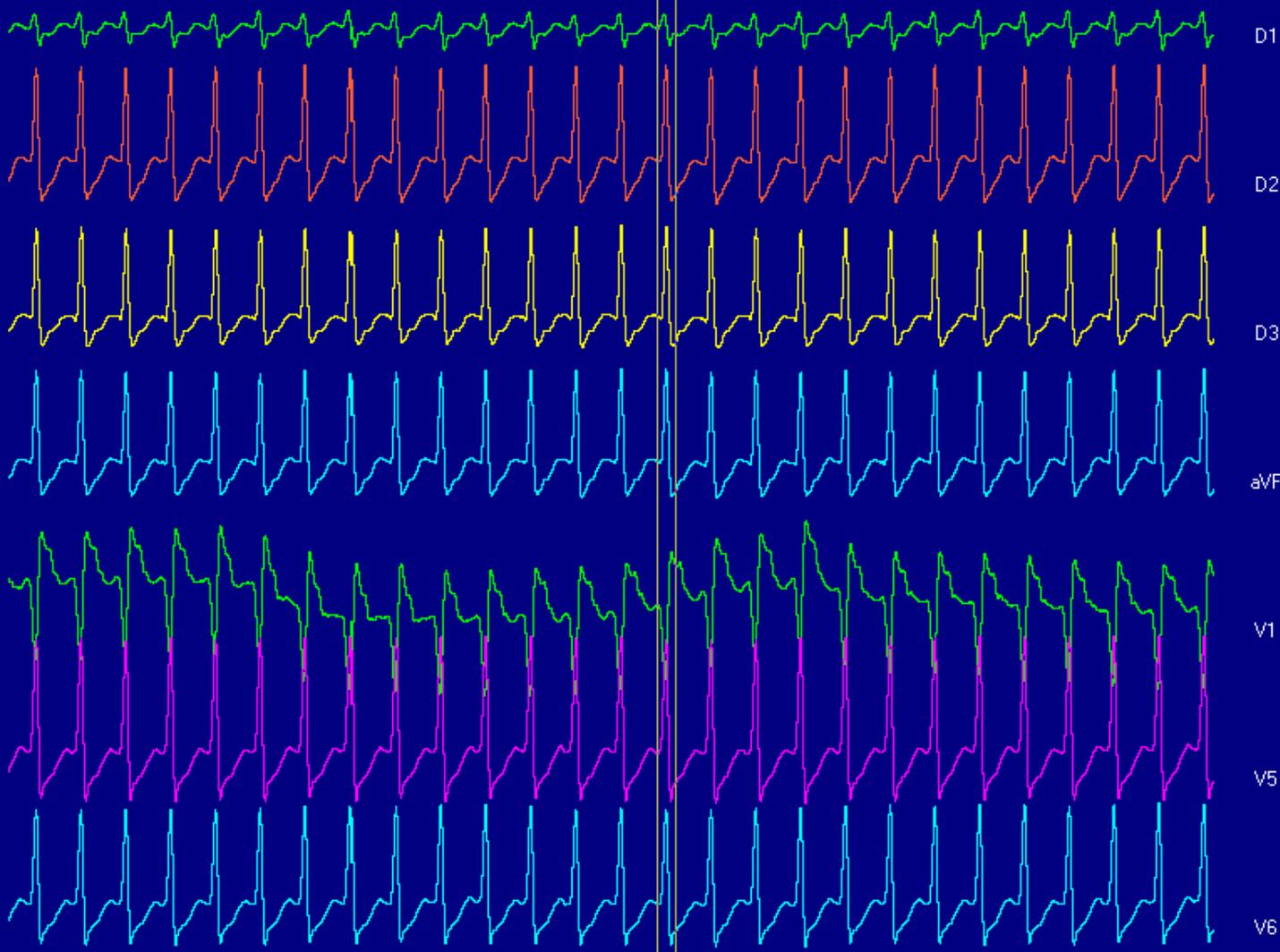
Medidas

Laudo/Salvar

Voltar

Português (Brasil)

t= 110



**TEB**  
ECGPC Versão 2.19

Título:  
TRECHO 11

Janela N  
25

< Adjacente >

Imprimir Janela

Imprimir por derivação:  
Use F1 a F12

Arquivo BMP

Medidas

Laudo/Salvar

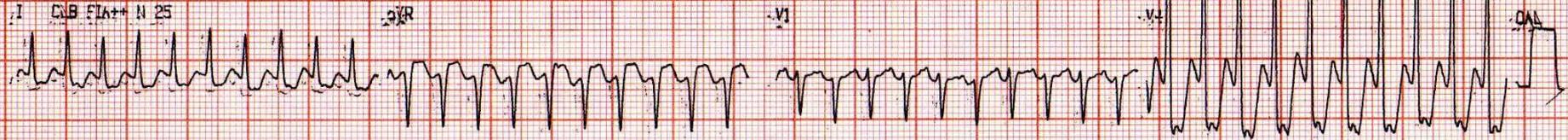
Voltar

Português (Brasil)

14:38

Ganon

I CB FIA++ N 25



aVR

aVL

aVF

Cal

II aVI



aV2

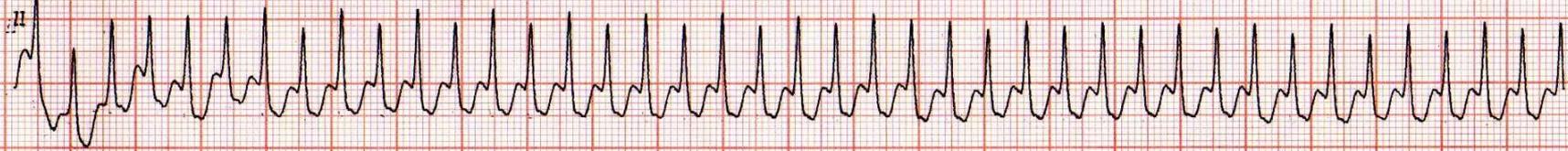
aV5

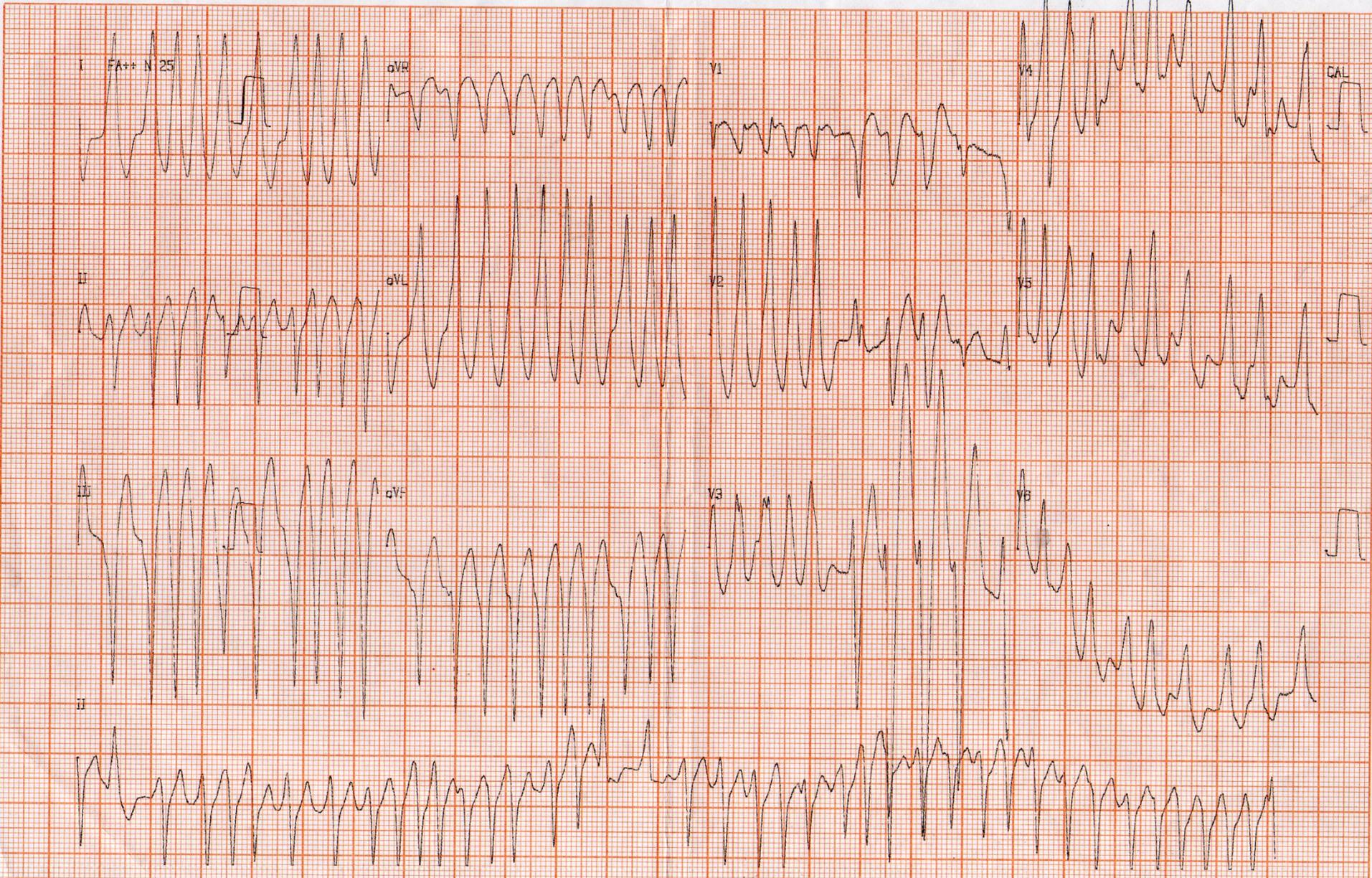
III aVF



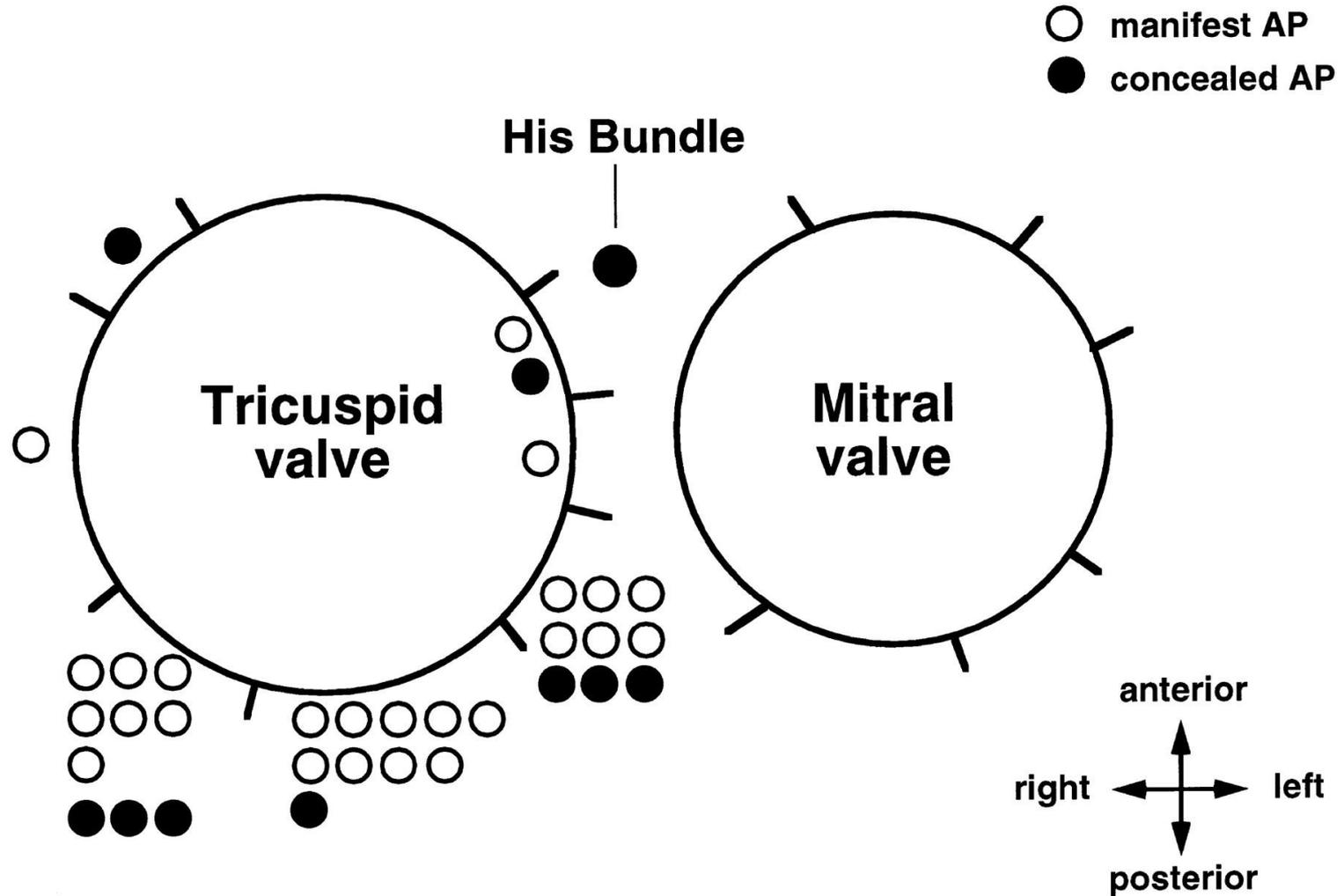
aV3

aV6





Schematic of the tricuspid and mitral valve annuli, as viewed in the 30° left anterior oblique projection, illustrates the locations of the 34 accessory pathways.

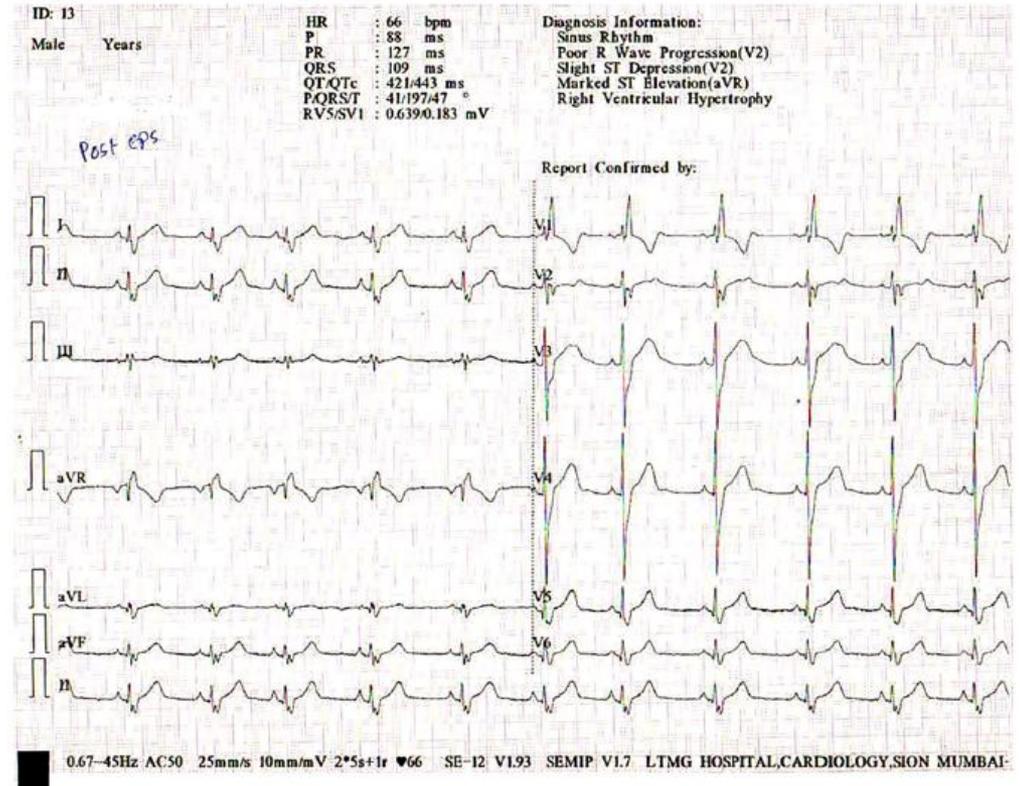


Cappato R et al. Circulation 1996;94:376-383

Case Report

**Absent right bundle branch block: Is it a clue of pre-excitation in Ebstein's anomaly?**

Pranil Bhalchandra Gangurde<sup>a,\*</sup>, Abhay Tidake<sup>b</sup>, Hetan Shah<sup>c</sup>,  
Ajay Mahajan<sup>d</sup>, Yash Lokhandwala<sup>e</sup>, Pratap Nathani<sup>f</sup>



# Surgery in a contemporary adult cohort of patients with Ebstein malformation of tricuspid valve and relation with ECG markers, atrial fibrosis and arrhythmic load

Yumi Shiina <sup>a,\*</sup>, Taka-aki Matsuyama <sup>b</sup>, Iki Adachi <sup>b</sup>, Wei Li <sup>a</sup>, Michael A. Gatzoulis <sup>a</sup>, Hideki Uemura <sup>c</sup>

<sup>a</sup> Adult Congenital Heart Centre and National Centre for Pulmonary Hypertension, Royal Brompton Hospital, NIHR Cardiovascular Biomedical Research Unit, Royal Brompton Hospital and National Heart and Lung Institute, Imperial College London, United Kingdom

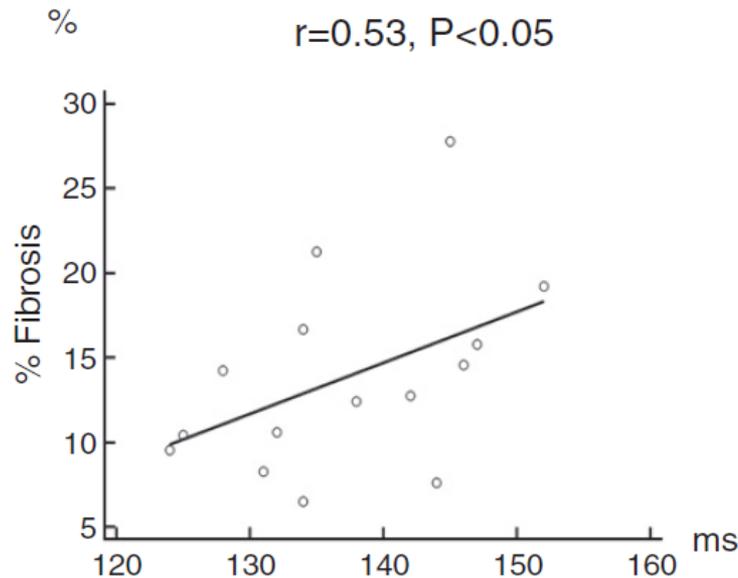
<sup>b</sup> Cardiac Morphology Unit, Royal Brompton Hospital, NIHR Cardiovascular Biomedical Research Unit, Royal Brompton Hospital and National Heart and Lung Institute, Imperial College London, United Kingdom

<sup>c</sup> Department of Cardiothoracic Surgery, Royal Brompton Hospital, NIHR Cardiovascular Biomedical Research Unit, Royal Brompton Hospital and National Heart and Lung Institute, Imperial College London, United Kingdom

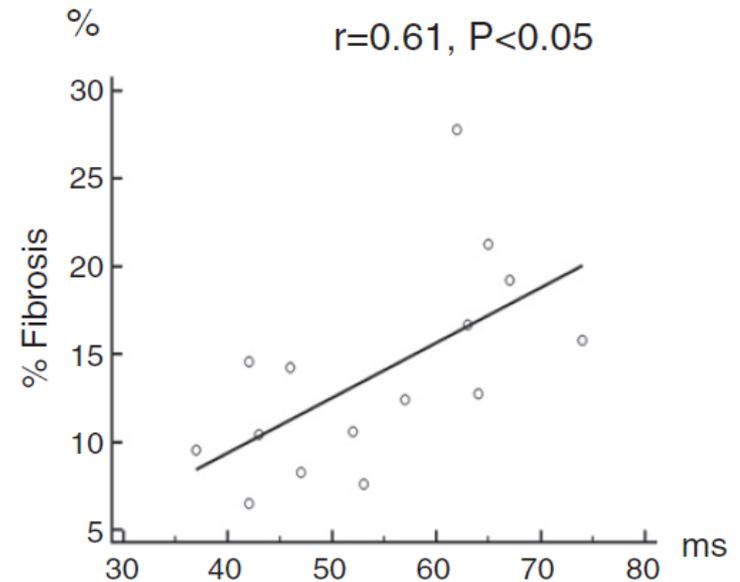
<http://dx.doi.org/10.1016/j.ijcard.2012.12.025>

## Fibrose e características da onda P no Ebstein

Maximum P-wave duration



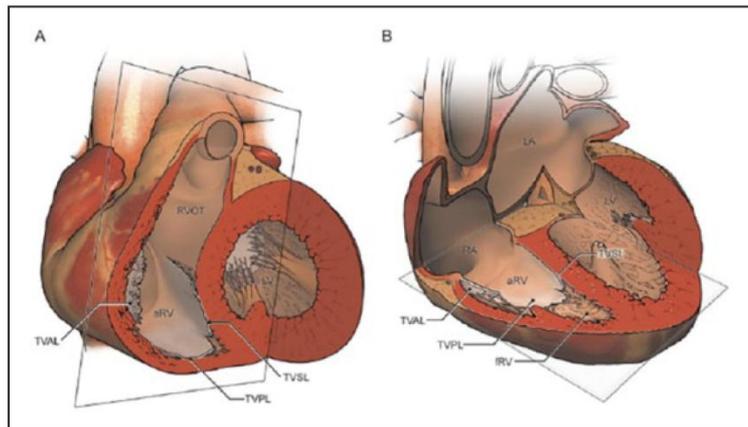
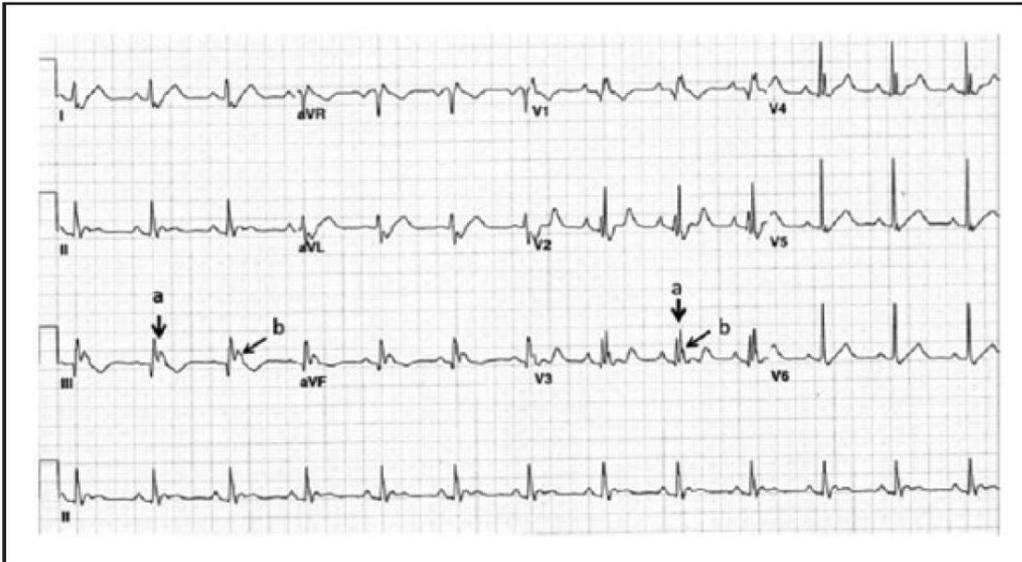
P-wave dispersion

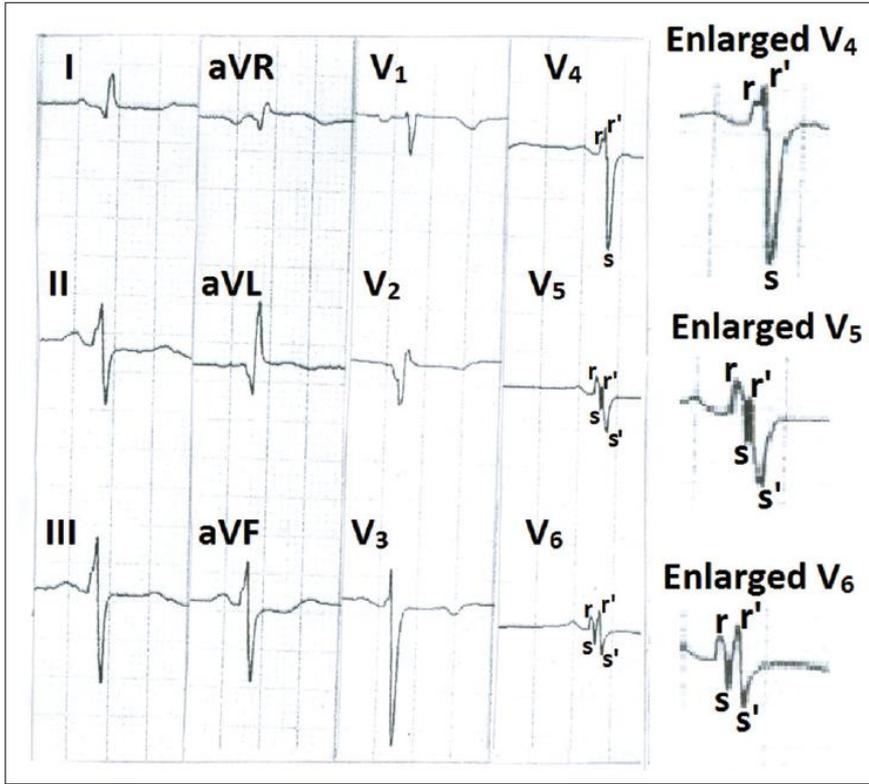


# Fragmentação QRS anomalia Ebstein

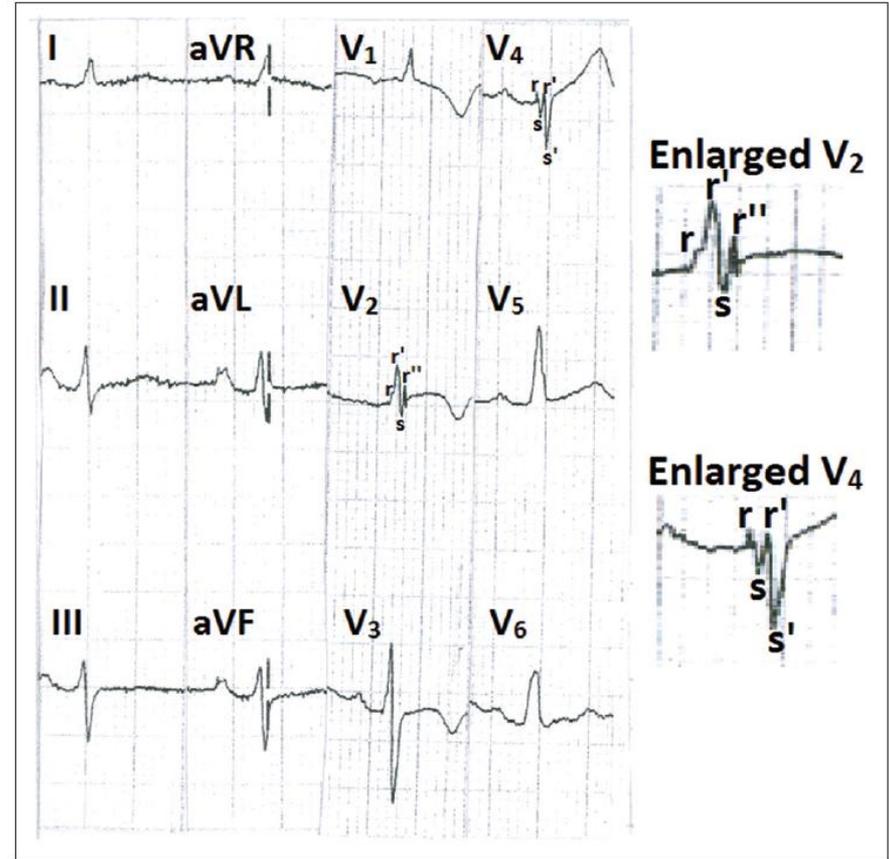
← Pré operatório

↓ Pós operatório





**Figure 1:** Electrocardiogram showing fragmented QRS in leads V<sub>4</sub> to V<sub>6</sub>. Lead I, aVL, and V<sub>2</sub> also show Q-waves

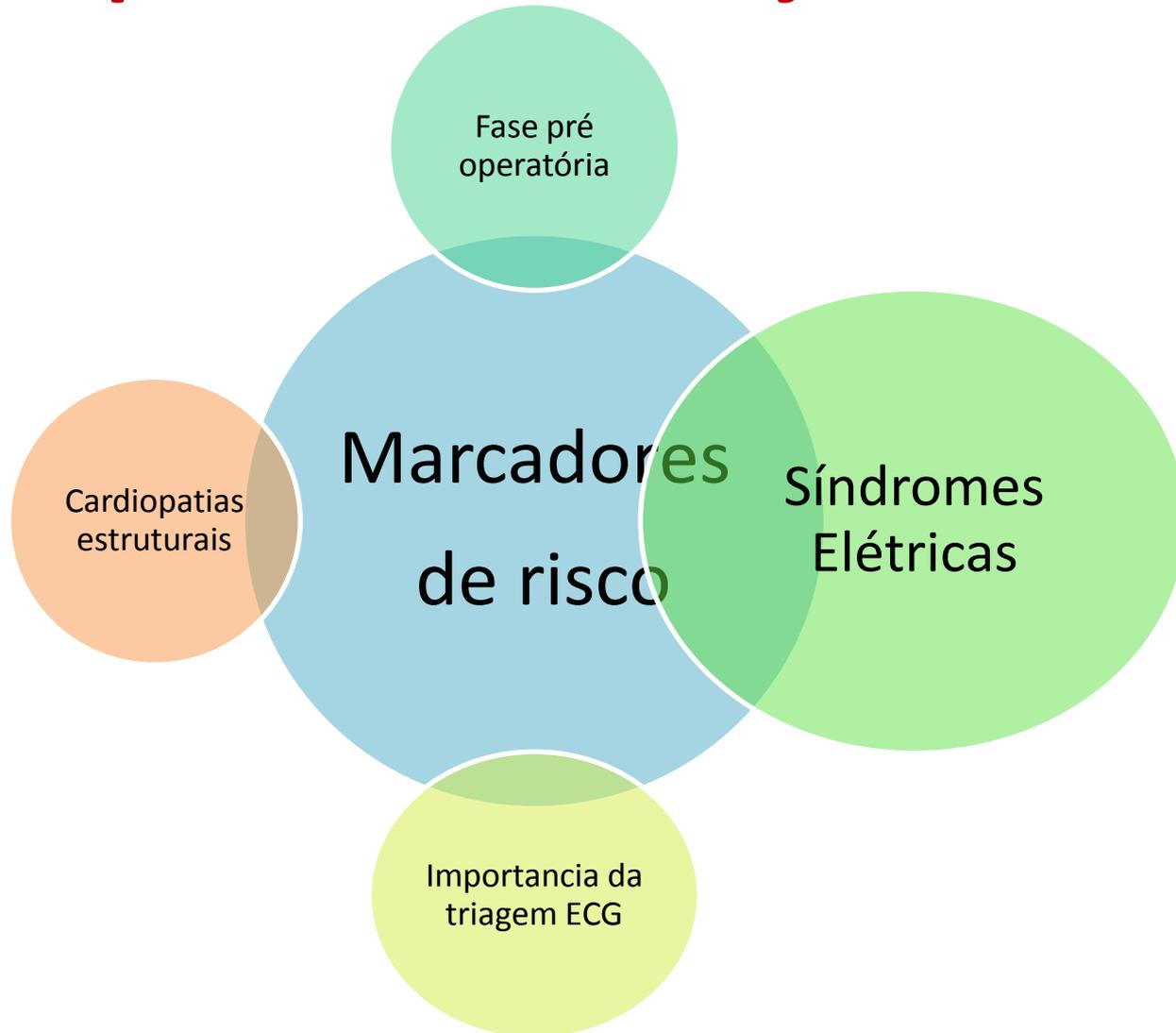


**Figure 2:** Electrocardiogram showing fragmented QRS in leads V<sub>2</sub> and V<sub>4</sub>

# Marcadores ECG na população pediátrica e em jovens



# Marcadores ECG na população pediátrica e em jovens



# **Síndrome do QT longo**

## **Síndrome do QT longo congênito (LQTS):**

- **Doença rara com história de síncope recorrente e morte súbita**
- **Primeira descrição em 1957 em uma família**
  - **desordem familiar autossômica recessiva e menos comum (síndrome de Jervell e de Lange-Nielsen)**
- **1960 - forma mais comum e autossômica dominante a síndrome Romano-Ward**

### **Epidemiologia**

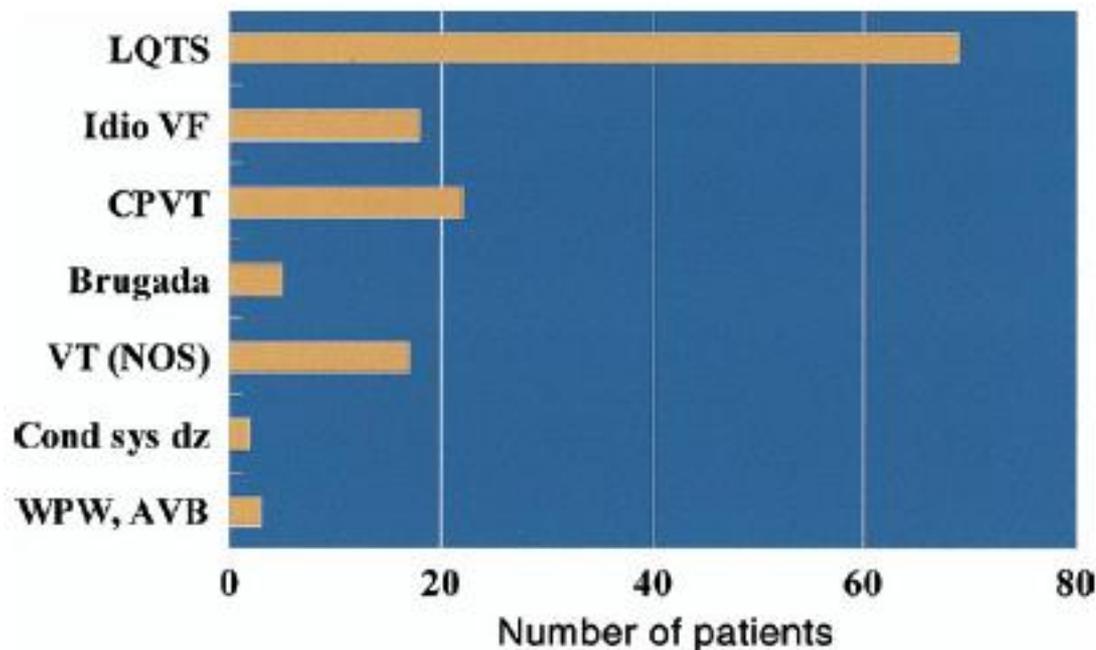
- **Síndrome de Romano-Ward - 1 de 5000 até 1 em 10000 indivíduos**
- **Síndrome de JLN - 1,6 a 6 por milhão de indivíduos .**
- **Causa de 3 a 4 mil mortes de causa repentina em crianças e adultos jovens cada ano e associada a uma de mortalidade em 10 anos de 70%**

# Anormalidades eletrocardiografica

## QTc prolongado

- O intervalo QT deve ser medido do inicio do QRS à extremidade final da onda T e ser calculado a média de 3-5 batimentos ;as ondas U devem ser incluídas na medida se forem grandes e unidas a onda T e deve ser ajustado para FC.
- Os ECGs de repetição são necessários quando a suspeita
- QTc e onda T normal não exclui LQTS
- TDP é mais freqüente em QTc > 600ms
- Complexo QRS largo aceita-se que um QTc 500ms como longo

# Results of a Multicenter Retrospective Implantable Cardioverter-Defibrillator Registry of Pediatric and Congenital Heart Disease Patients



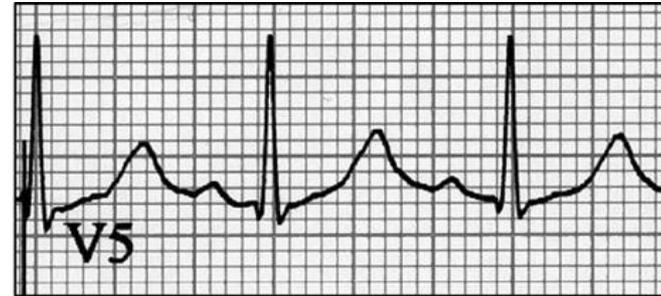
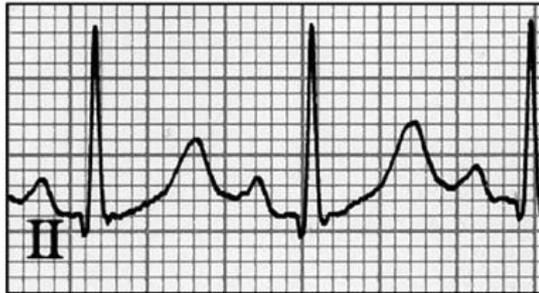
**Figure 2**

Electrical Diagnoses of Pediatric ICD Recipients With Structurally Normal Hearts

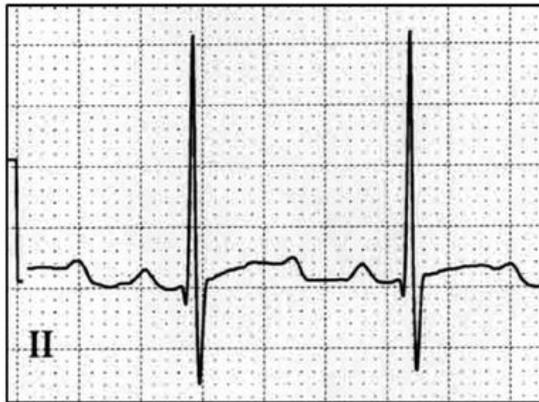
(J Am Coll Cardiol 2008;51:1685-91)

# Padrões eletrocardiográficos QT longo

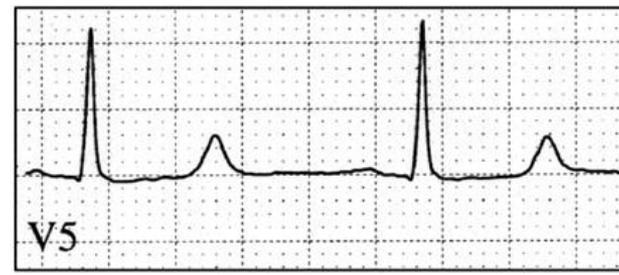
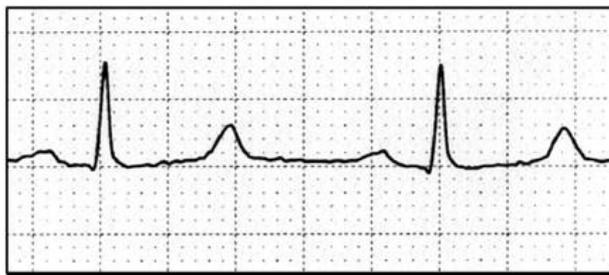
LQT1



LQT2



LQT3



# Como calcular o intervalo QTc

Different methods of QT correction.

QT correction method	Formula
Bazett	$QT/RR^{1/2}$
Fridericia	$QT/RR^{1/3}$
Framingham	$QT + 0.154 (1000 - RR)$
Hodges	$QT + 105 (1/RR - 1)$

## **Torsades de Pointes**

- Sequência curto-longo-curto
- Taquicardia polimórfica associada ao intervalo QT longo

## **Dispersão de QT**

- Esta aumentada nos pacientes LQTS (QTmax-QTmin)

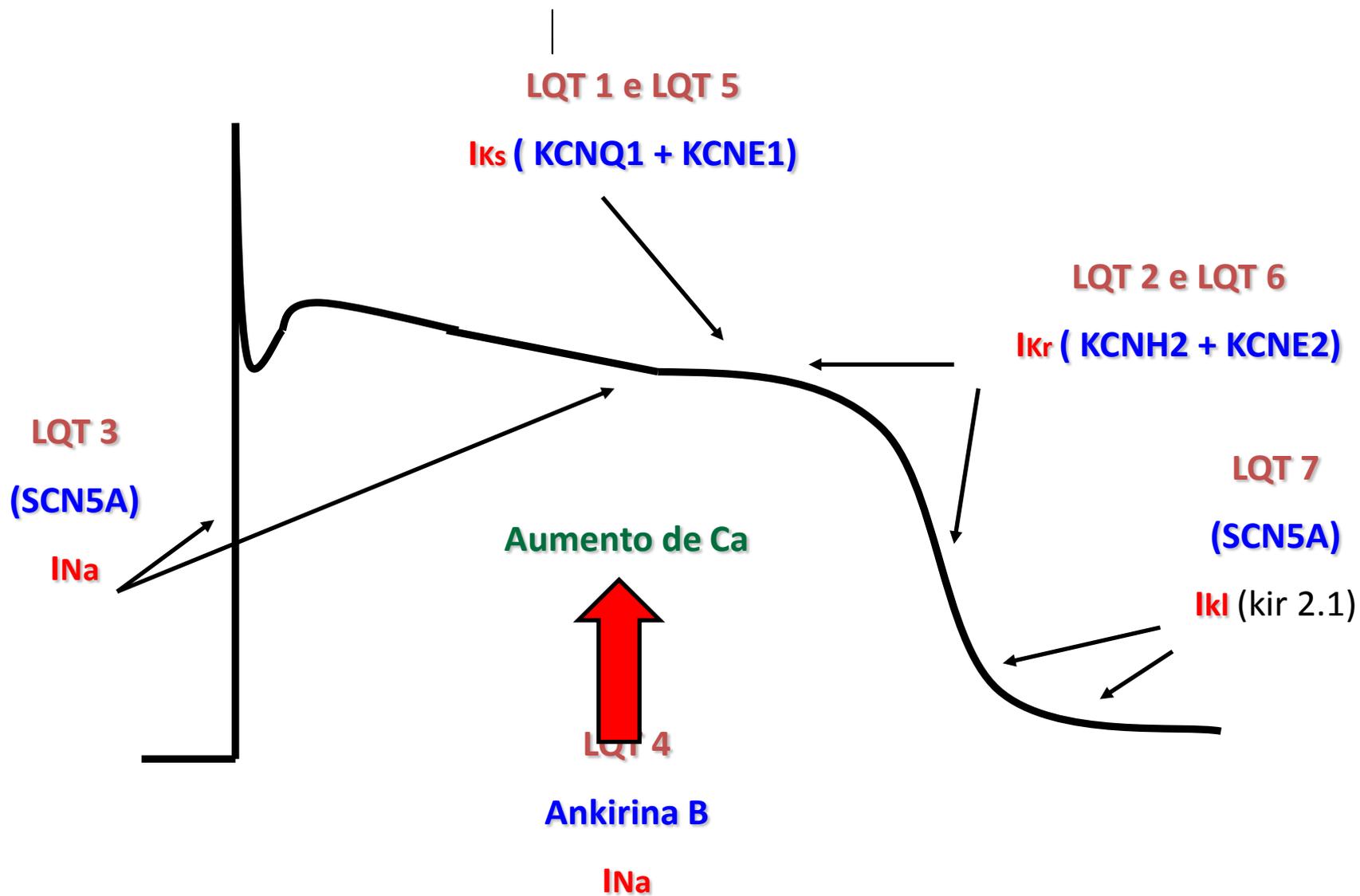
## **Sintomas**

- Sentimento de palpitação ou “vibrar” são incomuns porque a TDP é muito rápida
- Sincope

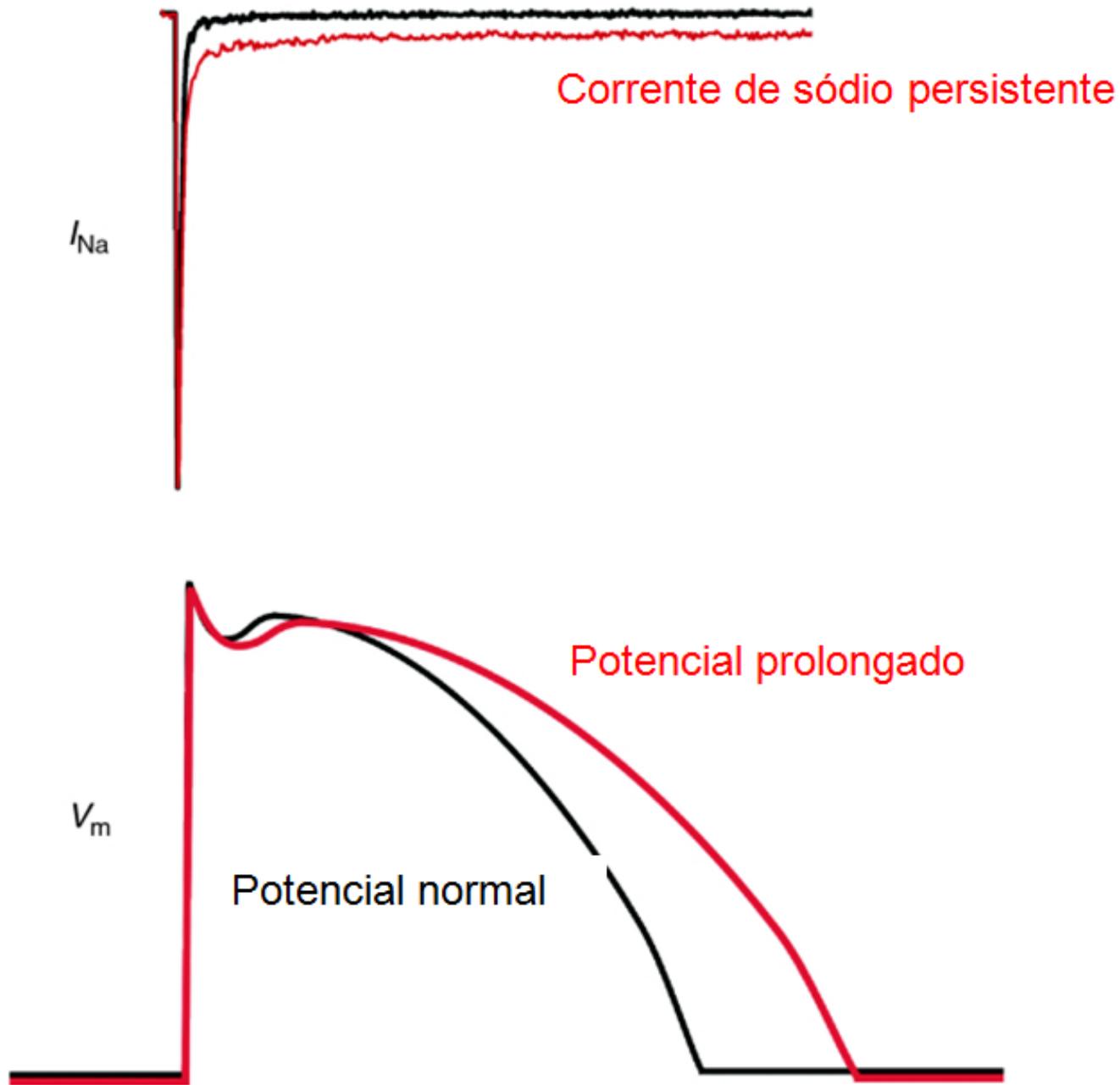
# **Critérios diagnósticos Síndrome do QT longo (Schwartz)**

	<b>Pontuação</b>
<b>QTc</b>	
>480 ms	<b>3</b>
460 – 470 ms	<b>2</b>
450 ms (masculino)	<b>1</b>
<b>Torsade pointes</b>	<b>2</b>
<b>Alternância de ondas T (TWA)</b>	<b>1</b>
<b>Baixa frequência cardíaca</b>	<b>0,5</b>
<b>Síncope</b>	
Com estresse	<b>2</b>
Sem estresse	<b>1</b>
Surdez congênita	<b>0,5</b>
<b>Membros da família com LQTS</b>	<b>1</b>
<b>Morte cardíaca inexplicada na família (imediatos) antes dos 30 anos</b>	<b>0,5</b>

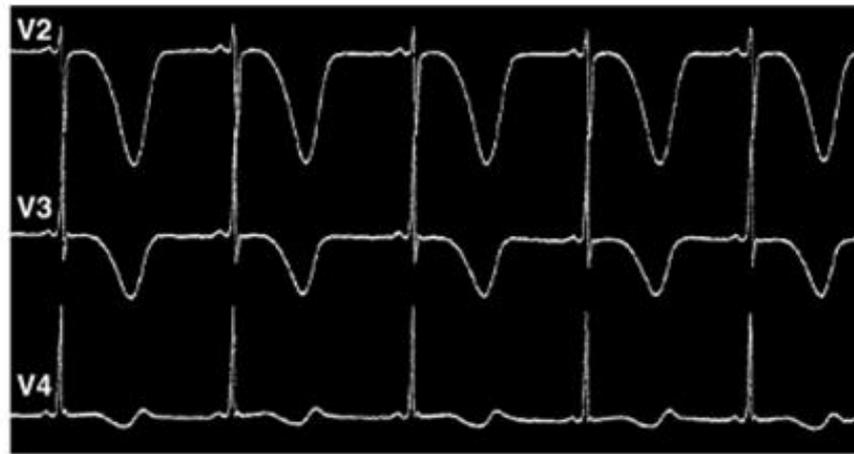
**<1 = baixa probabilidade; 2-3 = probabilidade intermediária; >4 alta probabilidade**



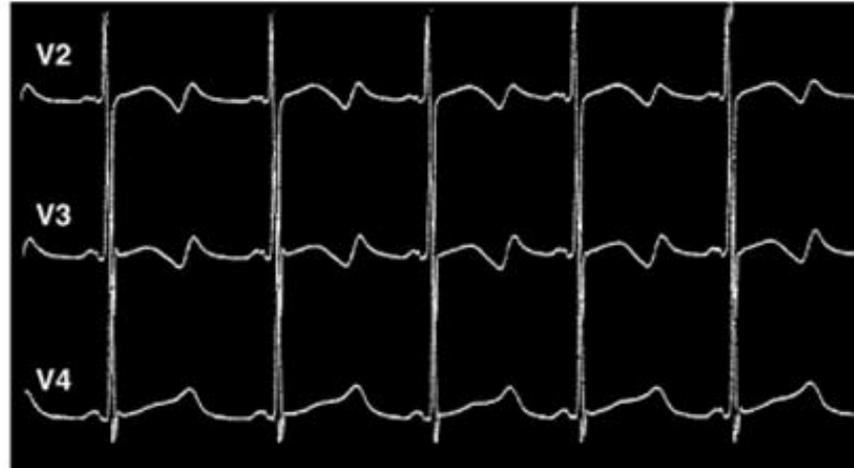
# LQTS 3



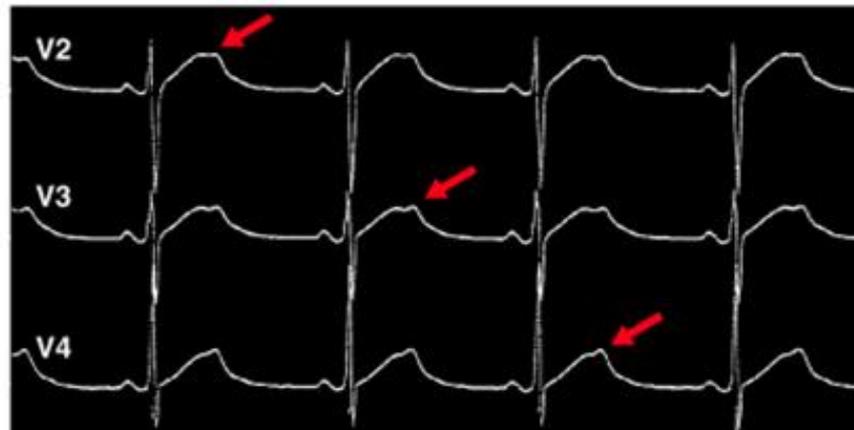
Caso índice  
7 anos



Irmã  
10 anos



Pai  
37 anos



# Alternancia de onda T no QT longo



I CB FA++ N 25

aVR

V1

V4



II

aVL

V2

V5



III

aVF

V3

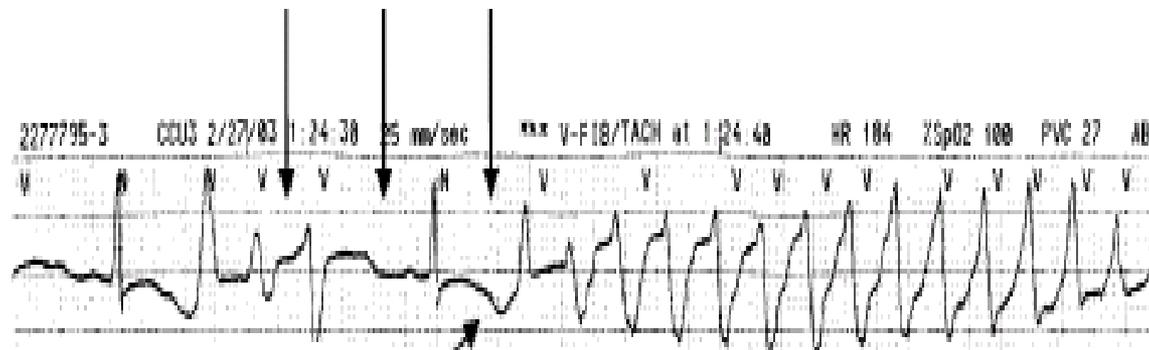
V6



II



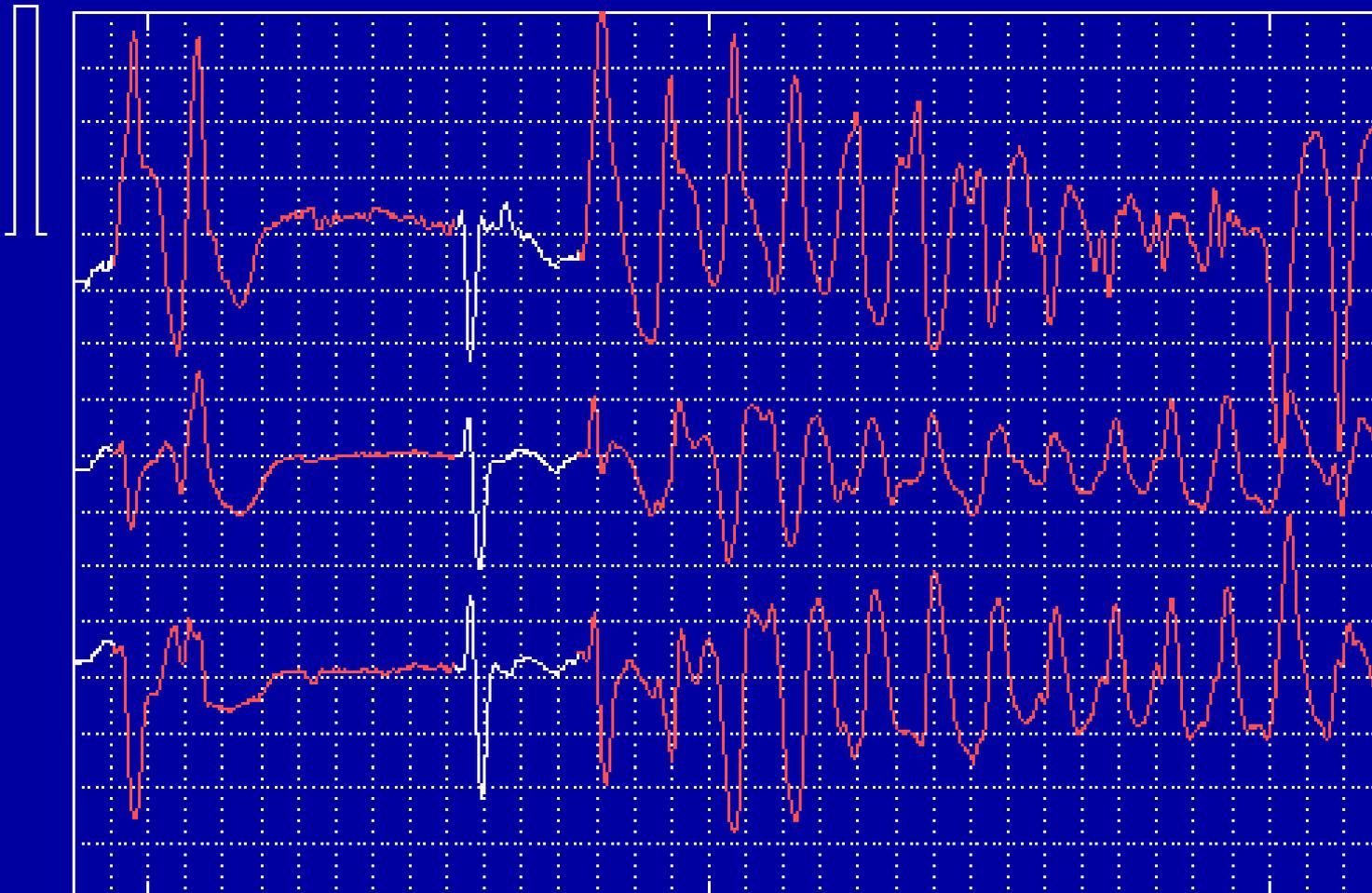
Curto Longo Curto

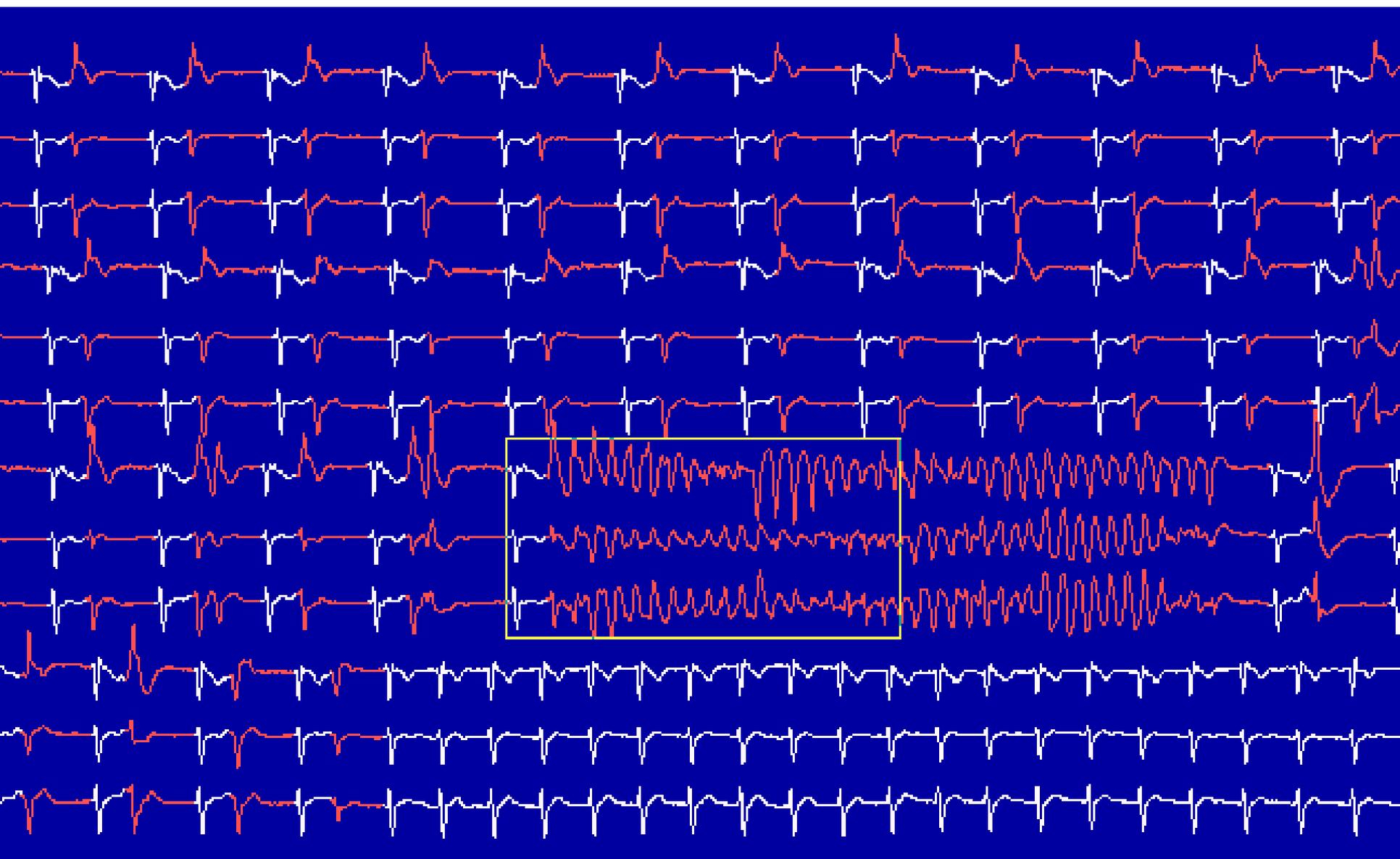


6<sup>a</sup>F 15:16:06

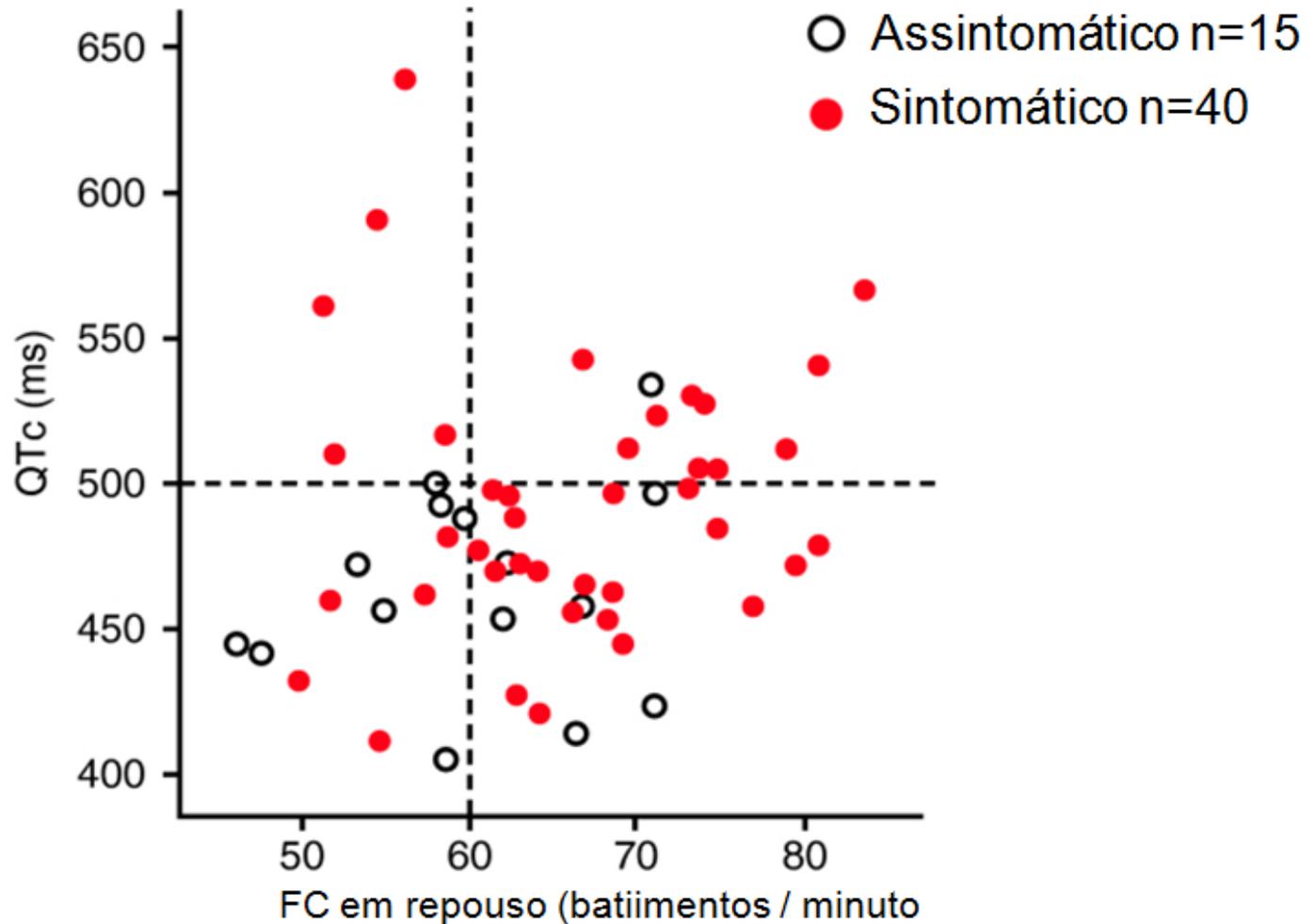
BPM

V V X N U U U U U U U U U U U U

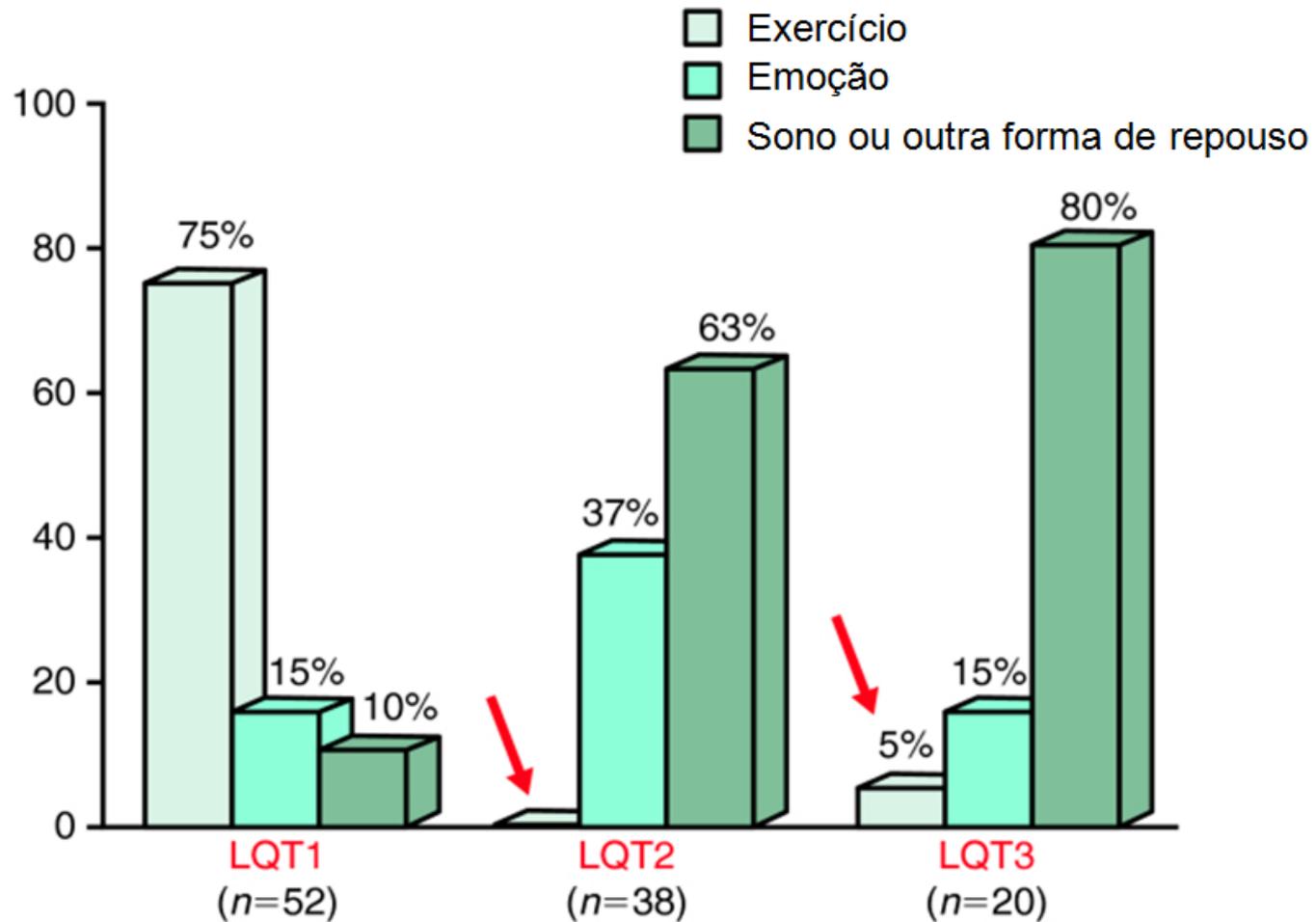




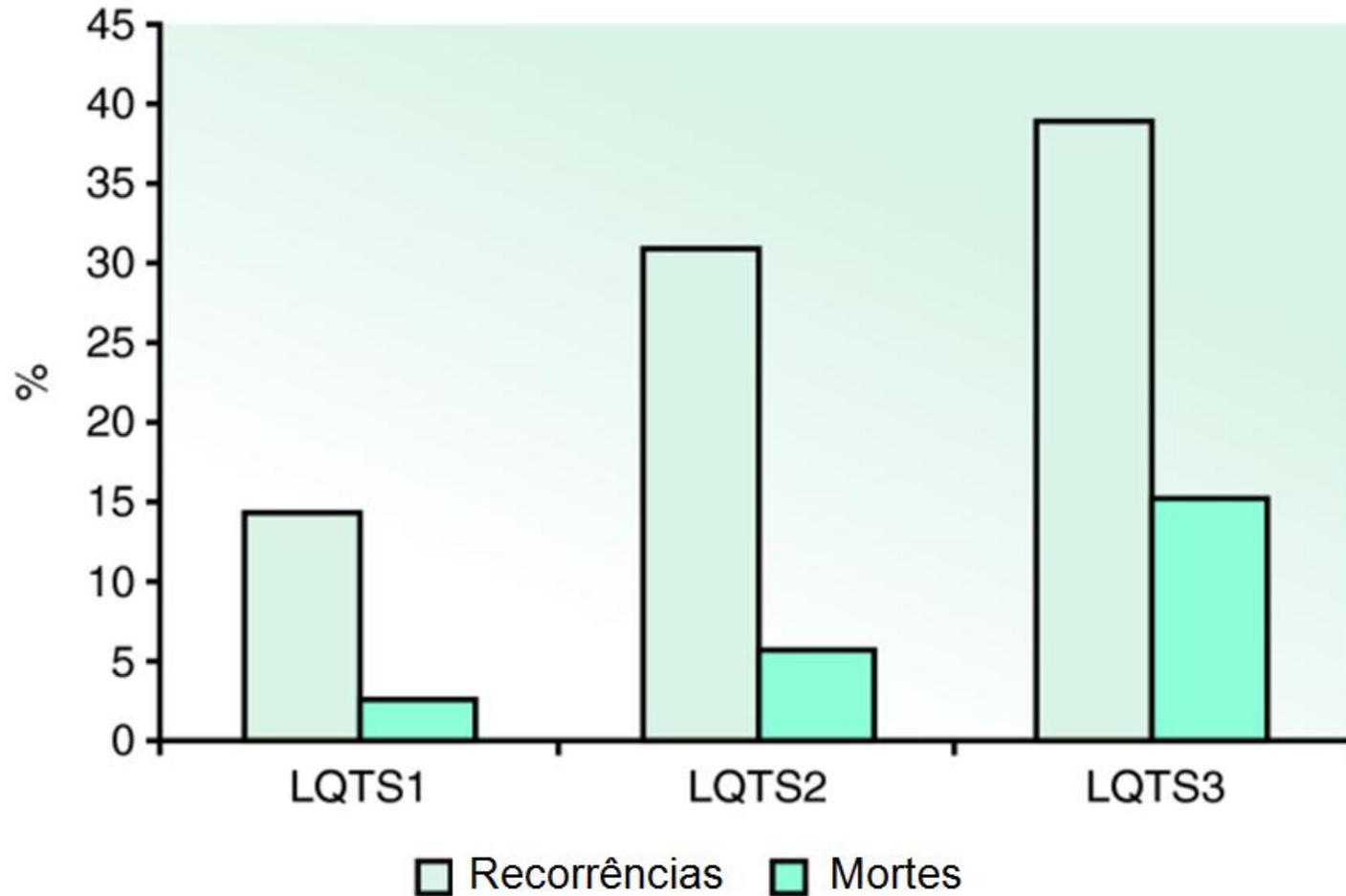
## Número de eventos dependente da FC e do QTc



# Gatilho para eventos súbitos na LQTS



## Recorrência de eventos com Beta- bloqueadores



*JAMA* 2004; 292:1341-1344

*J Med Genet* 2003; 40:141-145.

## Probabilidade de primeiro evento cardíaco (síncope, parada cardíaca e morte súbita) baseado no diagnóstico do tipo de LQT

	<b>QTc</b>	<b>Sexo</b>	<b>Risco</b>
<b>LQT1</b>	<p style="text-align: center;">≥ 500 ms</p> <p style="text-align: center;">&lt;500 ms</p>	<p>•Masculino e feminino</p>	<p>•Alto (&gt;50%)</p> <p>•Baixo (&lt;30%)</p>
<b>LQT2</b>	<p style="text-align: center;">≥ 500 ms</p> <p style="text-align: center;">&lt;500 ms</p>	<p>•Masculino e feminino</p> <p>•Feminino</p> <p>•Masculino</p>	<p>•Alto (&gt;50%)</p> <p>•Intermediário (30 a 49%)</p> <p>•Baixo (&lt;30%)</p>
<b>LQT3</b>	<p style="text-align: center;">≥ 500 ms</p> <p style="text-align: center;">&lt;500 ms</p>	<p>•Masculino</p> <p>•Feminino</p> <p>•Masculino e Feminino</p>	<p>•Alto (&gt;50%)</p> <p>•Intermediário (30 a 49%)</p> <p>•Intermediário (30 a 49%)</p>

# Estratificação de risco Síndrome do QT longo

	<b>I</b>	<b>Ila</b>	<b>Ilb</b>
<b>Estratificação de Risco</b>	<ul style="list-style-type: none"><li>•Torsade pointes/FV</li><li>•Parada cardíaca<ul style="list-style-type: none"><li>•JLN</li><li>•LQT3</li></ul></li></ul>	<ul style="list-style-type: none"><li>•QTc &gt; 600 ms</li><li>•Evento cardíaco na infância</li><li>•Sindactilia + BAVT</li><li>•Alternância de T (TWA)</li><li>•Sexo Feminino</li></ul>	<ul style="list-style-type: none"><li>•História familiar de MS</li><li>•Aumento da dispersão do QT</li></ul>
<b>Prevenção Primária</b>	<ul style="list-style-type: none"><li>• Suspende medicamentos que prolongam o QT</li><li>•Suspende atividade física</li><li>•Betabloqueadores</li></ul>	<b>CDI</b>	<ul style="list-style-type: none"><li>•Denervação cardíaca</li></ul>
<b>Prevenção Secundária</b>	<ul style="list-style-type: none"><li>•CDI</li><li>•Betabloqueadores</li><li>•Suspende medicamentos que prolongam o QT</li><li>•Suspende atividade física</li></ul>	-	-

# Diferenças fisiopatológicas QT longo e QT curto

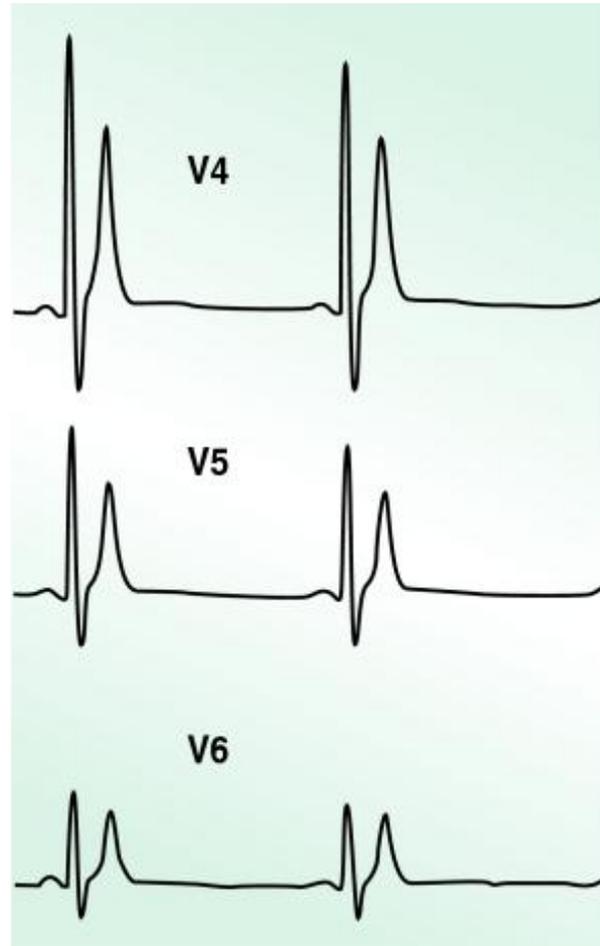
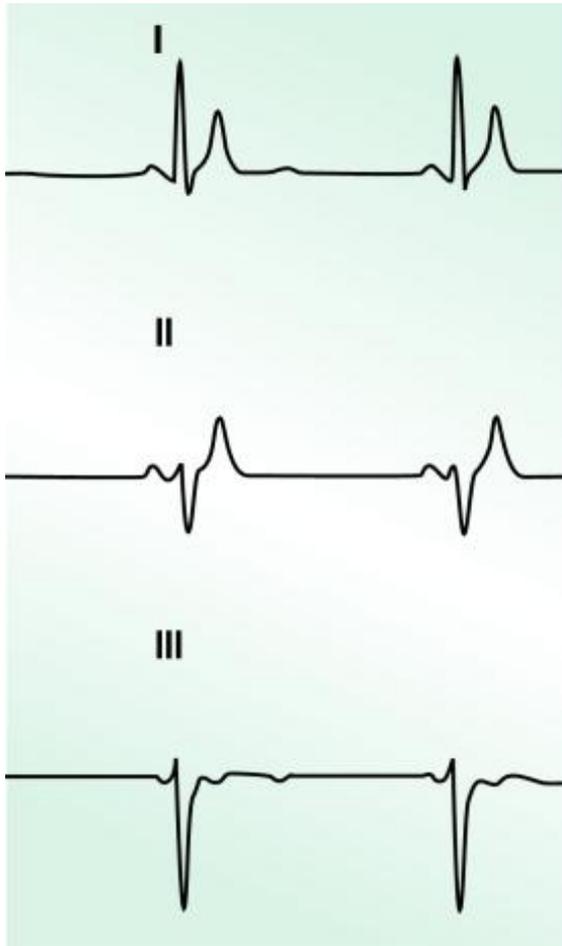
Electrophysiological changes observed in long and short QT syndromes (LQTS and SQTS). CV: conduction velocity. APD: action potential duration. ERP: effective refractory period. TDR: transmural dispersion of repolarization.  $\lambda$ : excitation wavelength ( $CV \times ERP$ ).

Abnormalities	LQTS	SQTS	References
Molecular mechanisms	Increased inward currents or reduced outward currents	Reduced inward currents or reduced outward currents	[15,30]
Triggered activity	Early afterdepolarizations from LTCC reactivation	Not observed	[86]
Substrates for reentry			
CV	$\leftrightarrow/\downarrow$	$\leftrightarrow/\uparrow$	[63]
APD	$\uparrow$	$\downarrow$	[53]
ERP	$\uparrow$	$\downarrow$	[63]
TDR	$\uparrow$	$\uparrow$	[54,87]
$\lambda$	$\downarrow$	$\downarrow$	[37,63]
APD alternans	$\uparrow$	N/A	[88,89]
CV restitution	$\leftrightarrow$	N/A	[29,90,91]
APD restitution	$\uparrow$ gradient	N/A	[29,92,93]
VERP restitution	$\uparrow$ gradient	N/A	[94]

# Síndrome do QT curto

- Associado a FV, TV e fibrilação atrial
- Associação com morte súbita berço
- Alterações nos canais de K
  - Redução na inativação da corrente de K
  - Interferência na fase 2 do potencial de ação
  - Encurtamento do QT
- PCR ocorre em repouso ou no estresse
- Tres formas clínicas
  - SQT1, SQT2, SQT3

Diagnóstico = QTc < 340 ms



# Aspectos clínicos QT curto

- Intervalo QTc abaixo 340 ms (zona nebulosa entre 330 e 360ms)
- Único marcador eficaz é clínico: história de PCR prévia
- Dois picos de mortalidade
  - Primeiro ano de vida
  - Entre 20 e 40 anos

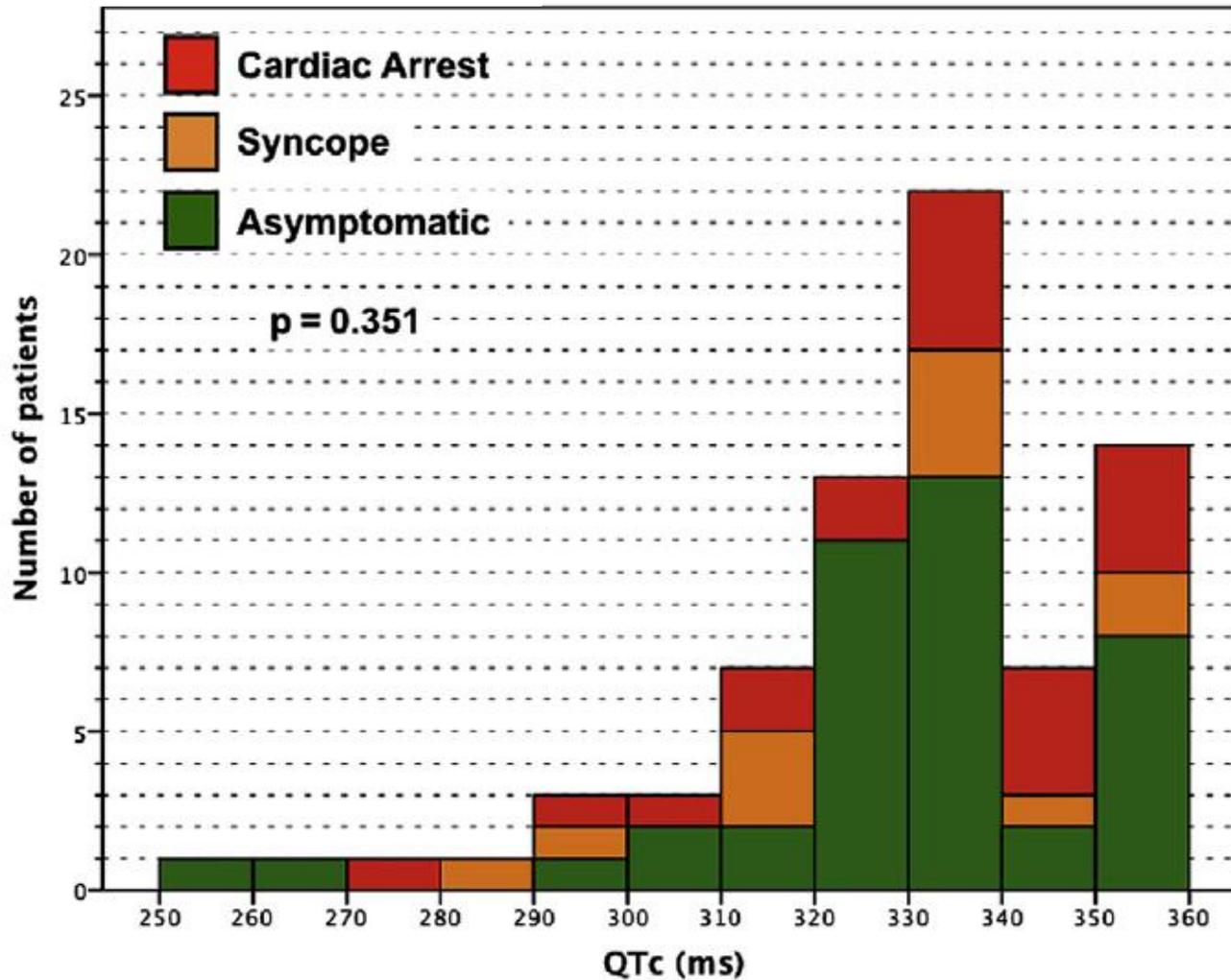
## Novel Insight Into the Natural History of Short QT Syndrome



Andrea Mazzanti, MD,\* Ajita Kanthan, MBBS, PhD,\* Nicola Monteforte, MD,\*  
Mirella Memmi, PhD,\* Raffaella Bloise, MD,\* Valeria Novelli, PhD,\* Carlotta Miceli, MS,\*  
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Antonio Curcio, MD, PhD,\* Andreea Elena Surducan, MS,§ Mario Colombo, MS,§  
Carlo Napolitano, MD, PhD,\*† Silvia G. Priori, MD, PhD\*†||

*Pavia, Italy; Gdansk, Poland; and New York, New York*

(J Am Coll Cardiol 2014;63:1300–8)

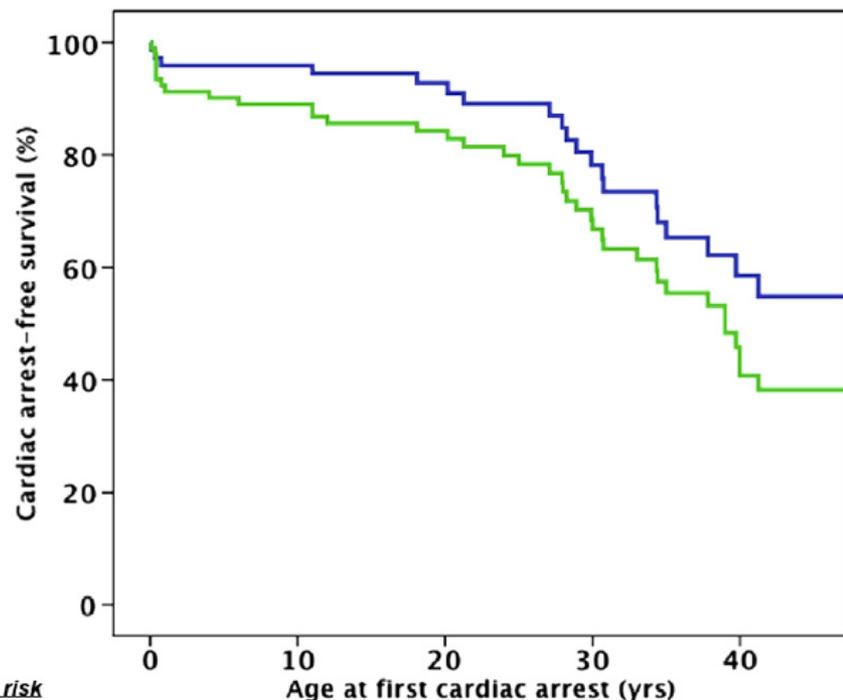


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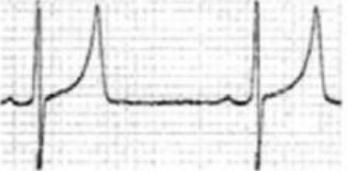
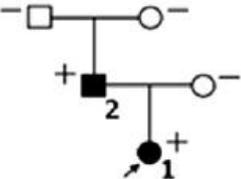
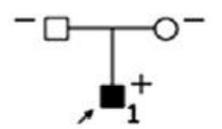
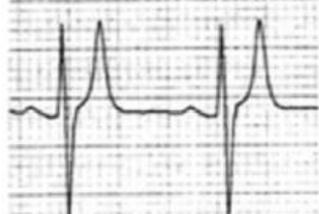
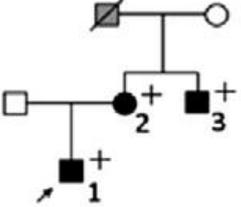
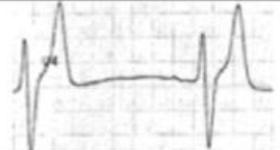
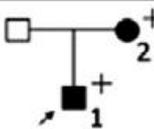
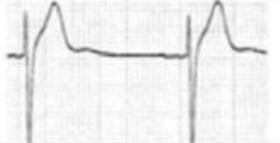
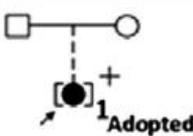
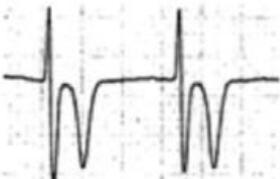
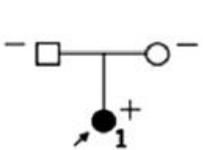
*Number at risk*

	0	10	20	30	40
Patients with ECG	73	69	52	34	16
Also pts without ECG	91	80	61	40	18

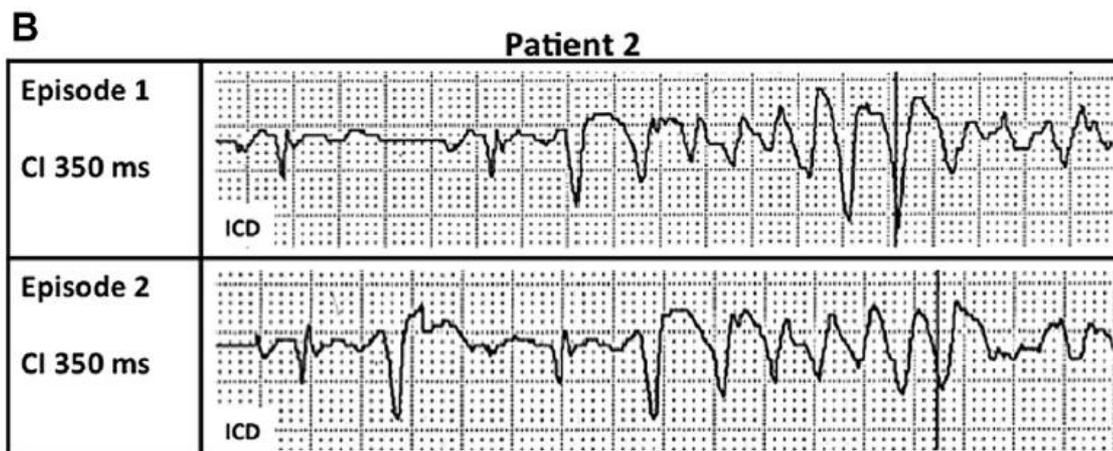
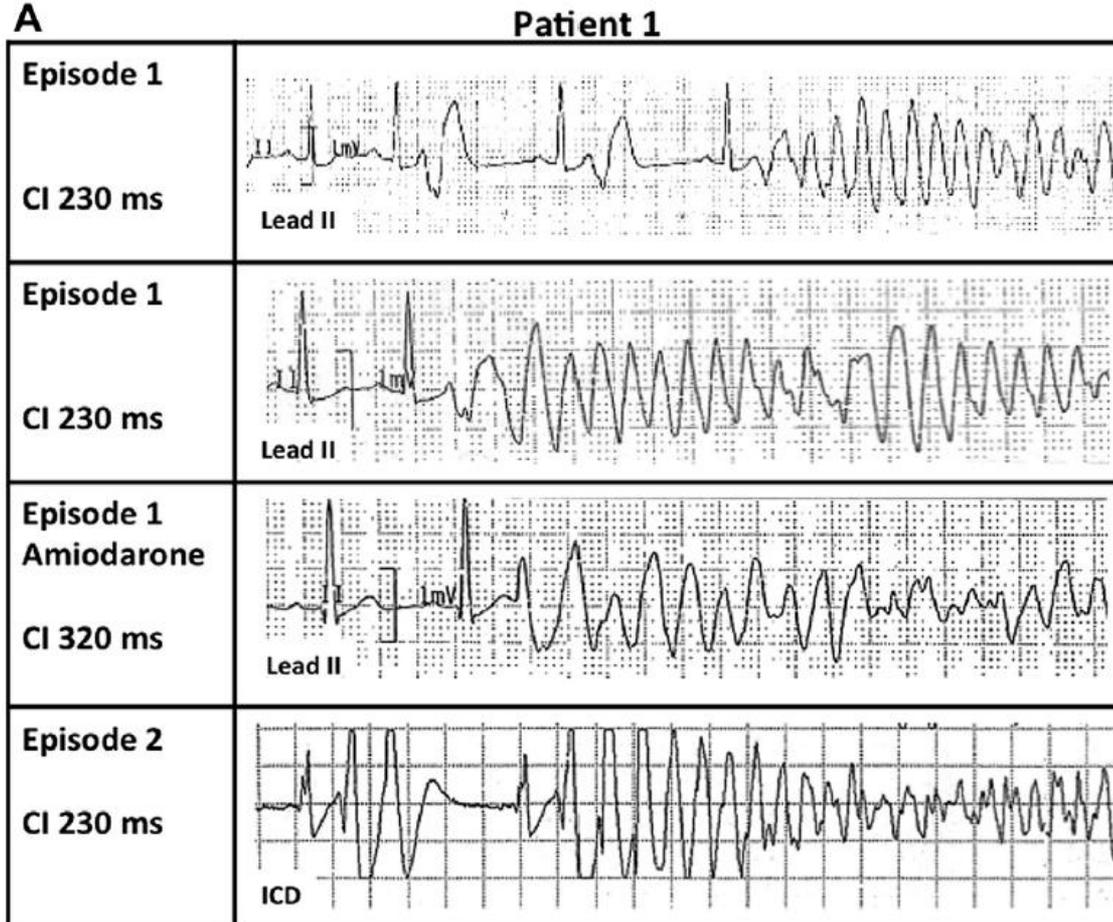
	Asymptomatic (n = 41)	Syncope (n = 12)	CA (n = 20)	p Value*
Male	32 (78)	11 (92)	18 (90)	0.353
Age at event, yrs	—	21 ± 11	25 ± 13	0.453
RR interval, ms	948 ± 93	938 ± 153	920 ± 132	0.682
QT interval, ms	321 ± 30	314 ± 29	317 ± 32	0.765
QTc interval, ms	330 ± 22	326 ± 24	331 ± 22	0.802

Values are mean ± SD or n (%). \*p values are for comparisons of all 3 groups together.

CA = cardiac arrest; QTc = corrected QT (interval).

Gene Mutation	ECG (precordial leads)	Pedigree	QTc	Symptoms at enrollment	Follow-up Events
KCNJ2 D172N			1 = 321 ms 2 = 332 ms	1 = None 2 = None	1 = None 2 = None
KCNJ2 E299V			1 = 256 ms	1 = AF	1 = None
KCNH2 N588K			1 = 291 ms 2 = 299 ms 3 = 283 ms	1 = Aborted CA 2 = None 3 = Syncope, AF	1 = None 2 = None 3 = None
KCNH2 T618I			1 = 264 ms 2 = 320 ms	1 = None 2 = None	1 = NSVT 2 = None
KCNQ1 R259H*			1 = 316 ms	1 = Aborted CA	1 = ICD Shock, NSVT
CACNA1C R1977Q*			1 = 316 ms	1 = Aborted CA	1 = Aborted CA

# Evolução para FV em 2 casos distintos



# Recorrência de PCR vs Pacientes assintomáticos

Journal of the American College of Cardiology  
 © 2014 by the American College of Cardiology Foundation  
 Published by Elsevier Inc.

Vol. 63, No. 13, 2014  
 ISSN 0735-1097/836-0  
<http://dx.doi.org/10.1016/j.jacc.2013.09.078>

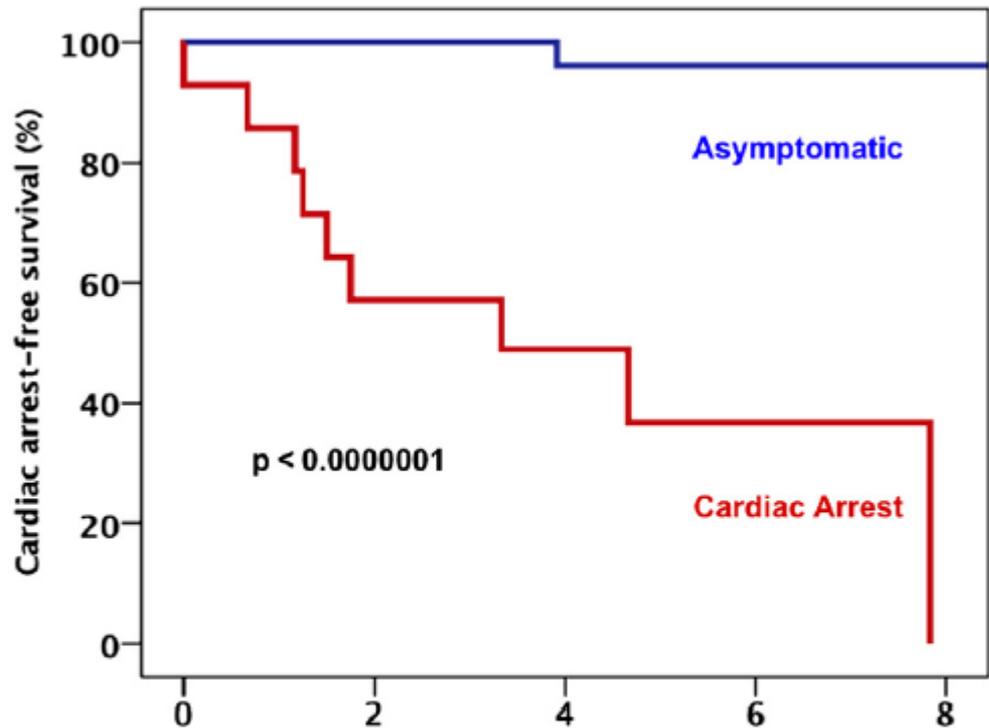
Heart Rhythm Disorders

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(J Am Coll Cardiol 2014;63:1300-8)



*Number at risk*

	0	2	4	6	8
Asymptomatic	48	36	25	14	7
CA	14	8	6	2	0

Figure 4

CA-Free Survival at Follow-up by CA Occurrence Before Enrollment

# **Taquicardia ventricular catecolaminérgica**

#### 5.4.5. *Taquicardia bidirecional*<sup>18</sup>

Trata-se de taquicardia de origem ventricular que, ao conduzir-se para o ventrículo, apresenta um sistema de condução com o ramo direito bloqueado constantemente e as divisões anterossuperior e posteroinferior do ramo esquerdo bloqueadas alternadamente, batimento a batimento. Eletrocardiograficamente, um batimento com QRS positivo e outro com QRS negativo se alternam sucessivamente, dando aspecto bidirecional. Esta arritmia se relaciona a quadros de intoxicação digitálica, doença miocárdica grave por cardiomiopatia avançada e doença sem cardiopatia estrutural, como a taquicardia catecolaminérgica familiar.

## Mutação ANK2

Repouso



Exercício



A

## Mutação KCNJ2

Repouso



Exercício



B

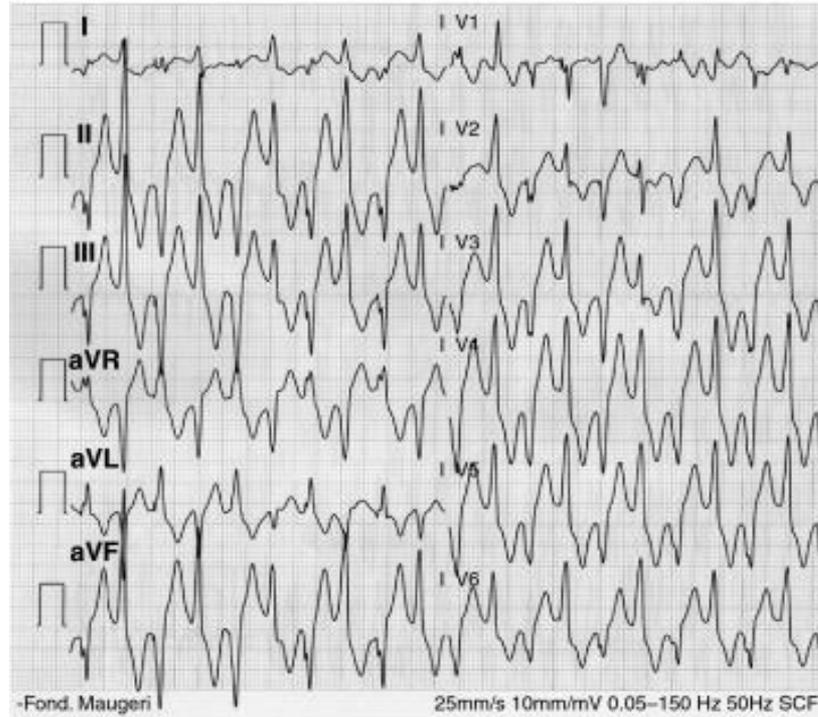
16/02/2005 14:07:32

2.5 km/h

Rate 115

0.0%

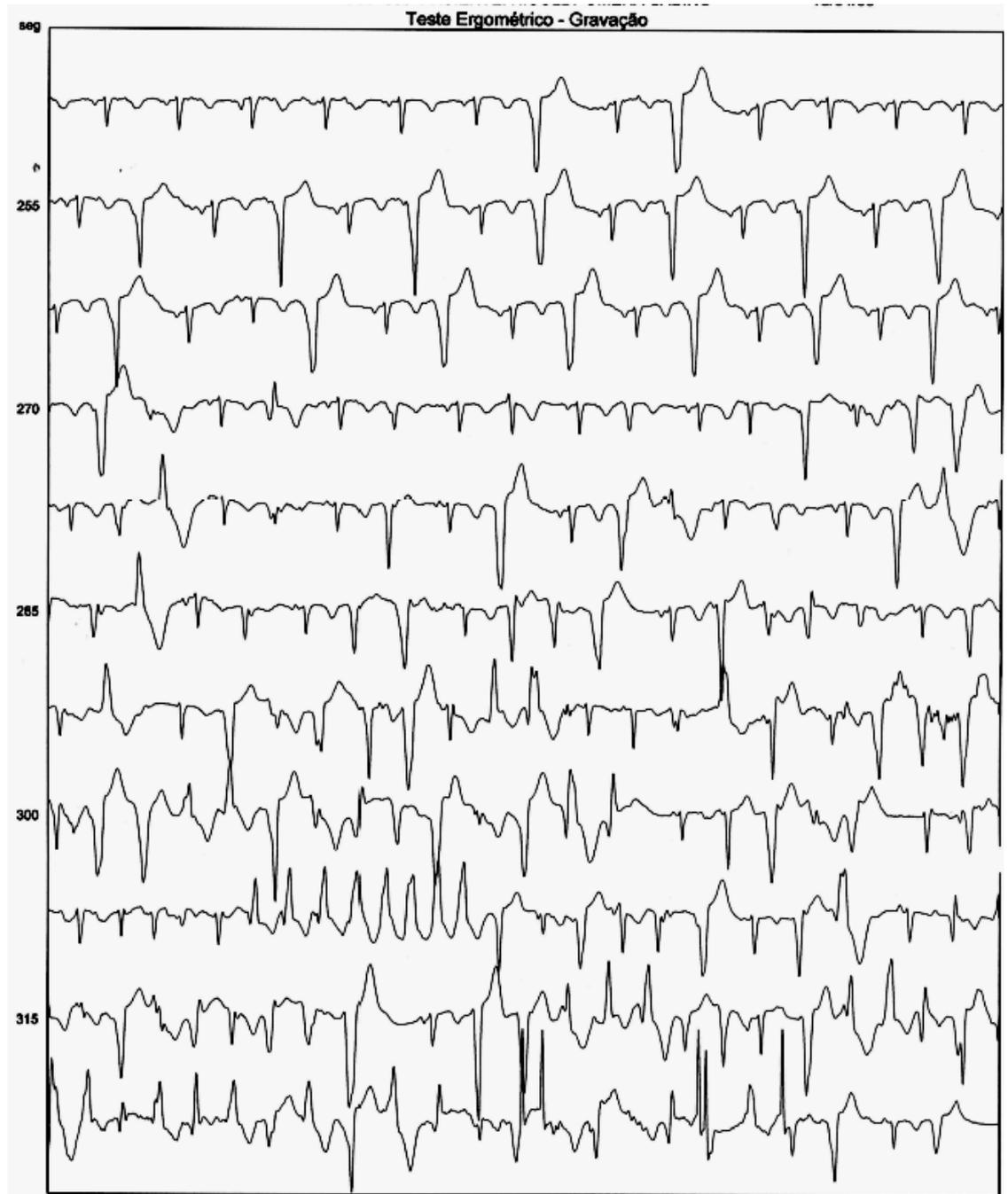
BP---/---



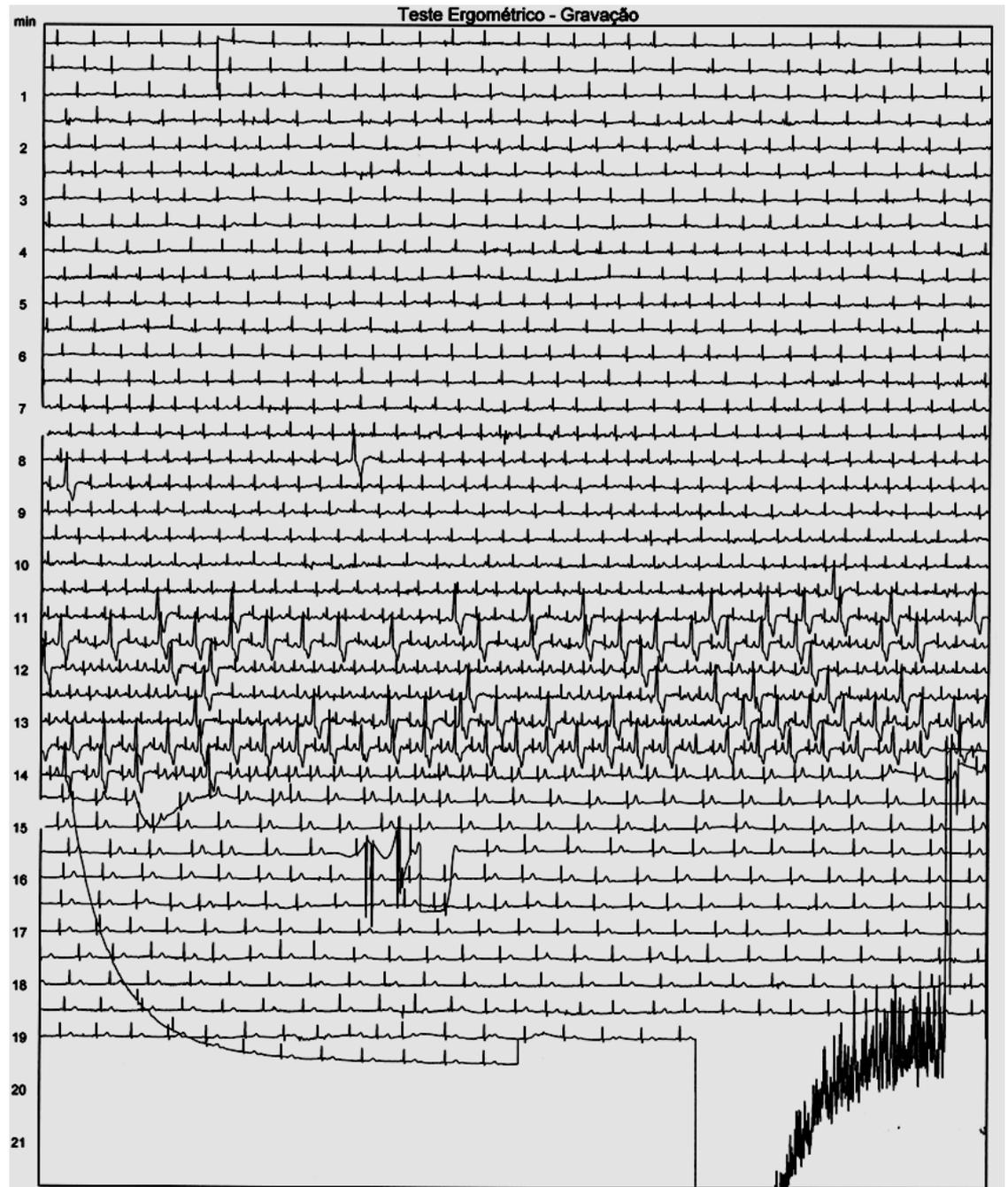
16:19:51



**Menina 4 anos  
síncope aos  
esforços  
pré medicação**



**Menina 4 anos  
síncope aos  
esforços após  
medicação**



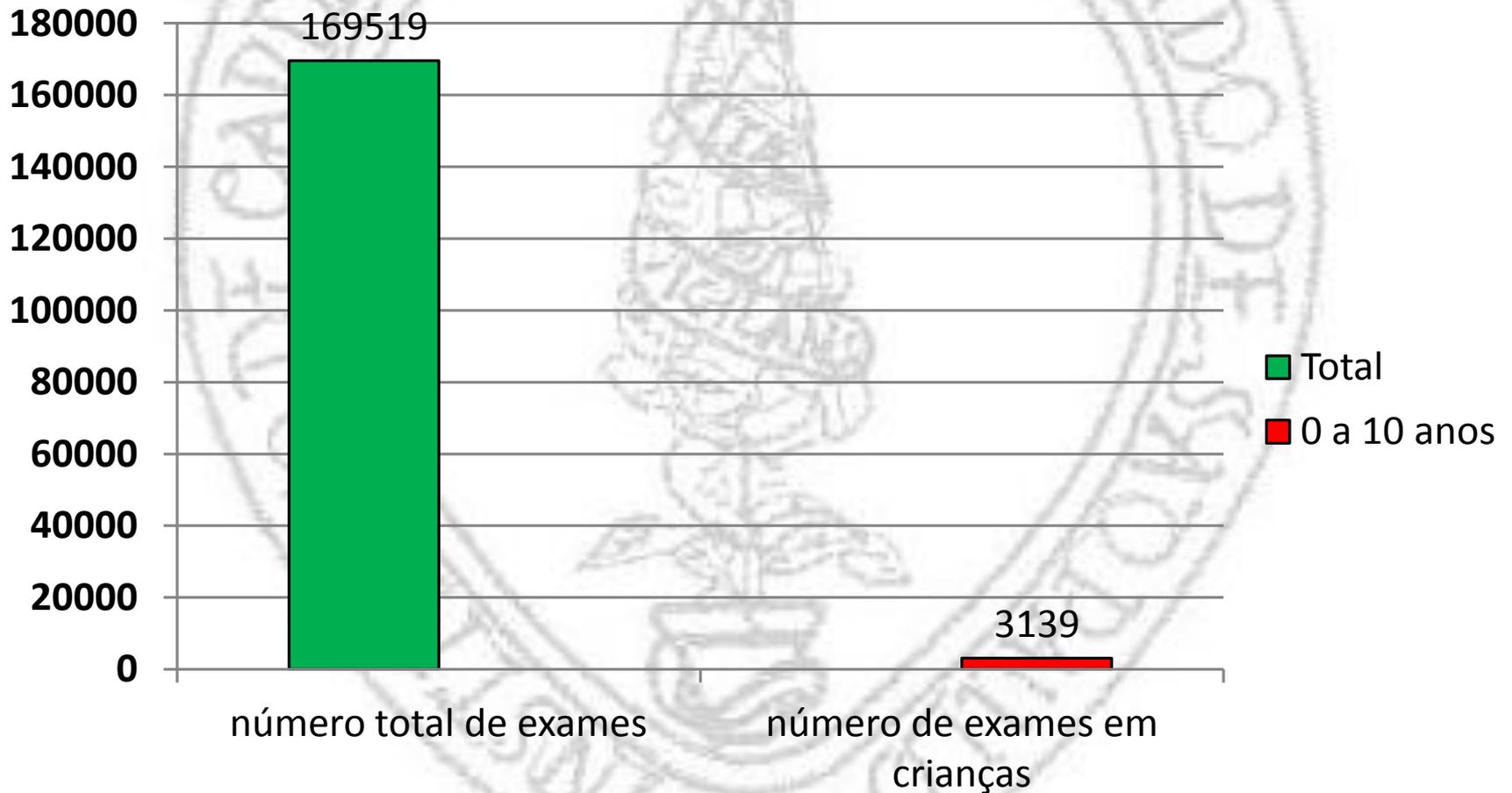
# Marcadores ECG na população pediátrica e em jovens



# Marcadores ECG na população pediátrica e em jovens

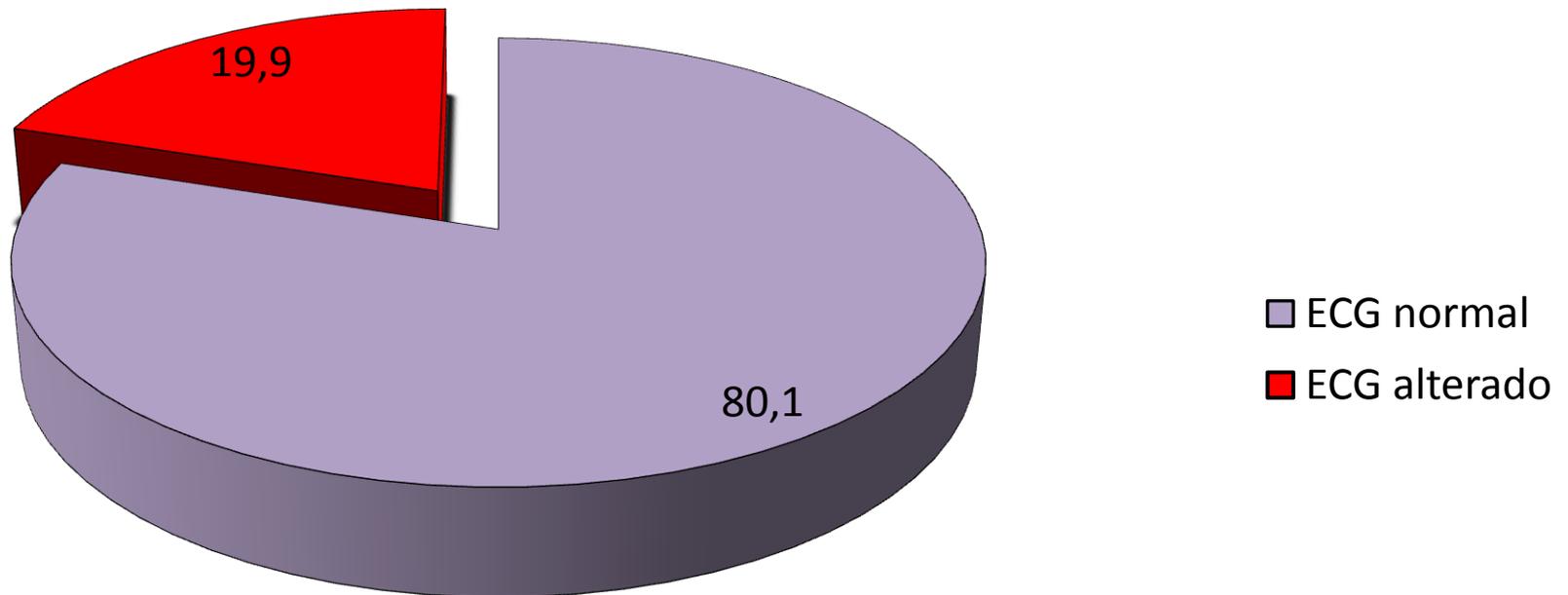


# Julho 2007 – Dezembro 2010



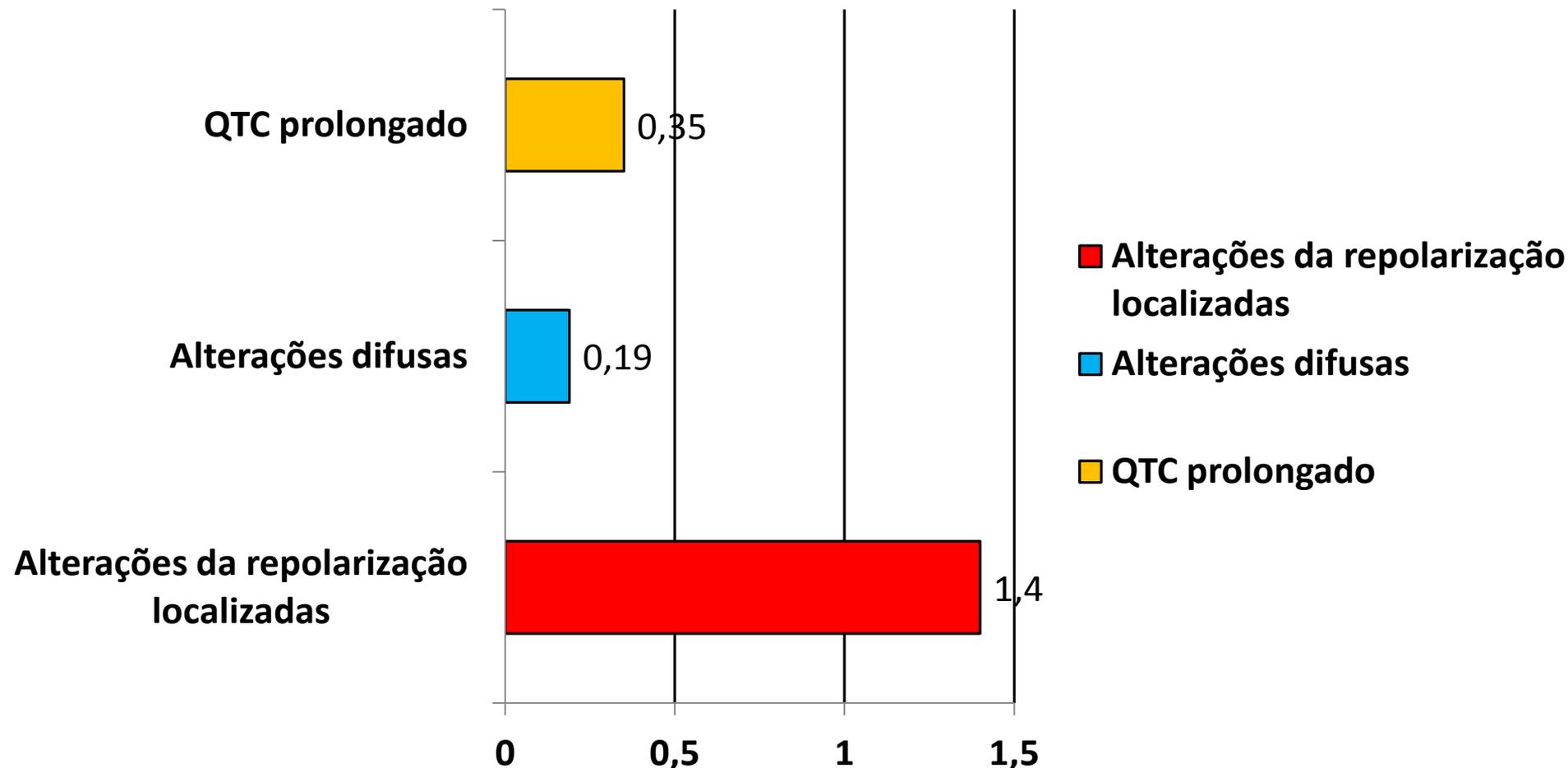
# Perfil geral dos exames em crianças 0 a 10 anos

% de exames - total 3139 exames



# Análise da repolarização ventricular

% dos exames - total 3139 exames





Seção Médica de Eletrofisiologia Clínica e Arritmias Cardíacas  
Instituto Dante Pazzanese de Cardiologia

# O perfil eletrocardiográfico de adolescentes assintomáticos submetidos ao eletrocardiograma pelo sistema TELE ECG: análise de 11058 pacientes

.

*Rogério Andalaft, MARIANA FUZUY, VIRGINIA CERUTTI, MARCELO MORETTI, SANDRO FELICIONI, RINALDO FERNANDES, ADIMÉIA SANTOS, BENTO GOMES, DALMO A R MOREIRA, FRANCISCO FAUSTINO DE A.C. FRANCA*

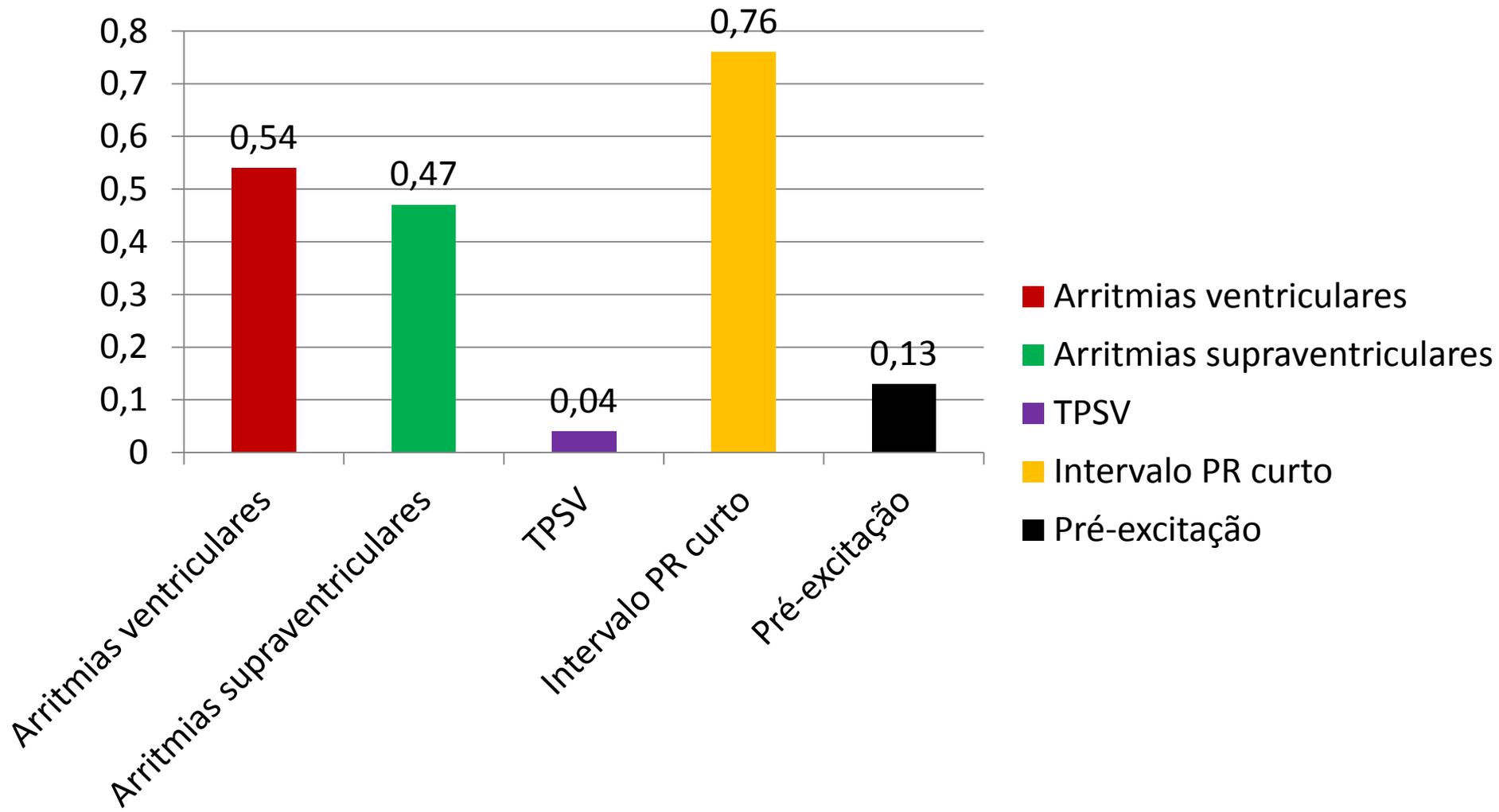
**Setor de Tele- ECG**

**Seção Médica de Eletrofisiologia Clínica e Arritmias Cardíacas do  
Instituto Dante Pazzanese de Cardiologia**

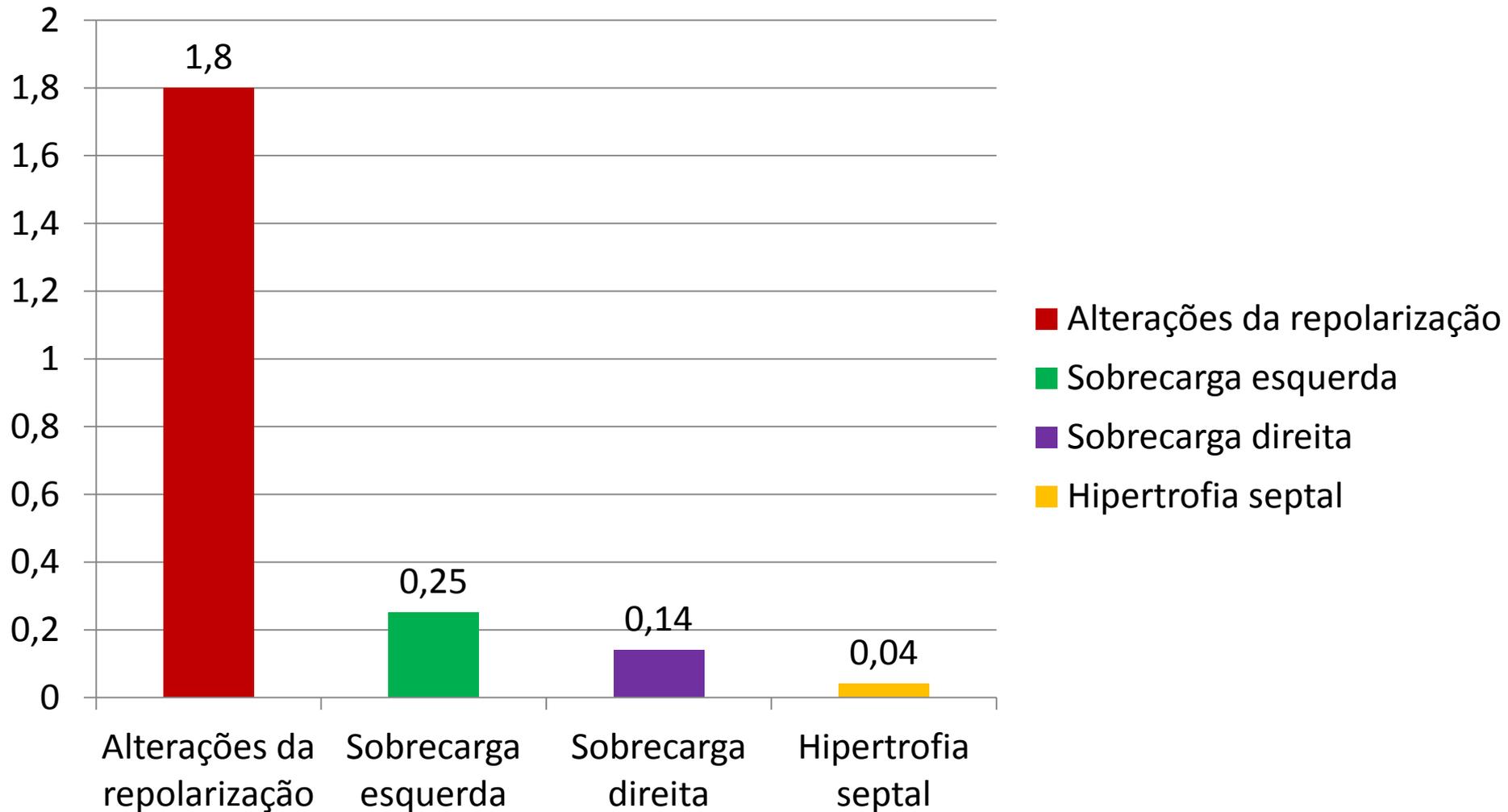
# Material

- Foram analisados 11058 pacientes (55% sexo masculino)
- Idade entre 10 e 20 anos de idade
- 
- Base de dados de 797115 p
- Período de análise entre julho de 2007 e agosto 2014

## Arritmias assintomáticas ao ECG (%)



## Alterações da repolarização e sobrecargas(%)





**São Paulo**  
**7 e 8 de abril de 2017**

## **Curso de Eletrocardiograma em crianças e cardiopatias congenitas**



**ECG normal em pediatria**  
**Sobrecargas atriais e ventriculares**  
**Bloqueios de ramo**  
**ECG nas cardiopatias não cianogênicas**  
**ECG nas cardiopatias cianogênicas**  
**O ECG no pronto socorro pediátrico**  
**Síndromes elétricas e morte súbita**  
**Diagnóstico de arritmias no pós operatório**  
**O uso do eletro em condições especiais**



### **Informações**

**Data: 07 e 08 de abril de 2017**  
**Local: Rua Loefgren, 1057**  
**(próximo ao Shopping Santa Cruz)**

**07/04: das 18:00h às 22:00h**  
**08/04: das 08:00h às 18:00h**



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**[www.arritmiaonline.com.br](http://www.arritmiaonline.com.br)**

<b>Categoria</b>	<b>Investimento</b>
Médicos	R\$ 350,00
Residentes	R\$ 280,00
Outros profissionais	R\$ 250,00
No local	R\$ 500,00



# Obrigado!!!

[rogerioandalaft@arritmiaonline.com.br](mailto:rogerioandalaft@arritmiaonline.com.br)



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