30 de setembro de 2017

coração curumim

Morte súbita em lactentes, crianças e adolescentes

Palestrante Rogério Braga Andalaft

rogerio.andalaft@einstein.br

ALBERT EINSTEIN HOSPITAL ISRAELITA

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

American Heart Association®
life is why™

PACES

Siga instruções
TRATAMENTO
Eletrodos
Liga/desliga
Desfibrilador Externo Automático
DEA

Morte súbita

- Profilaxia primária

- Profilaxia secundária

ALBERT EINSTEIN
HOSPITAL ISRAELITA

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

Fisiopatologia

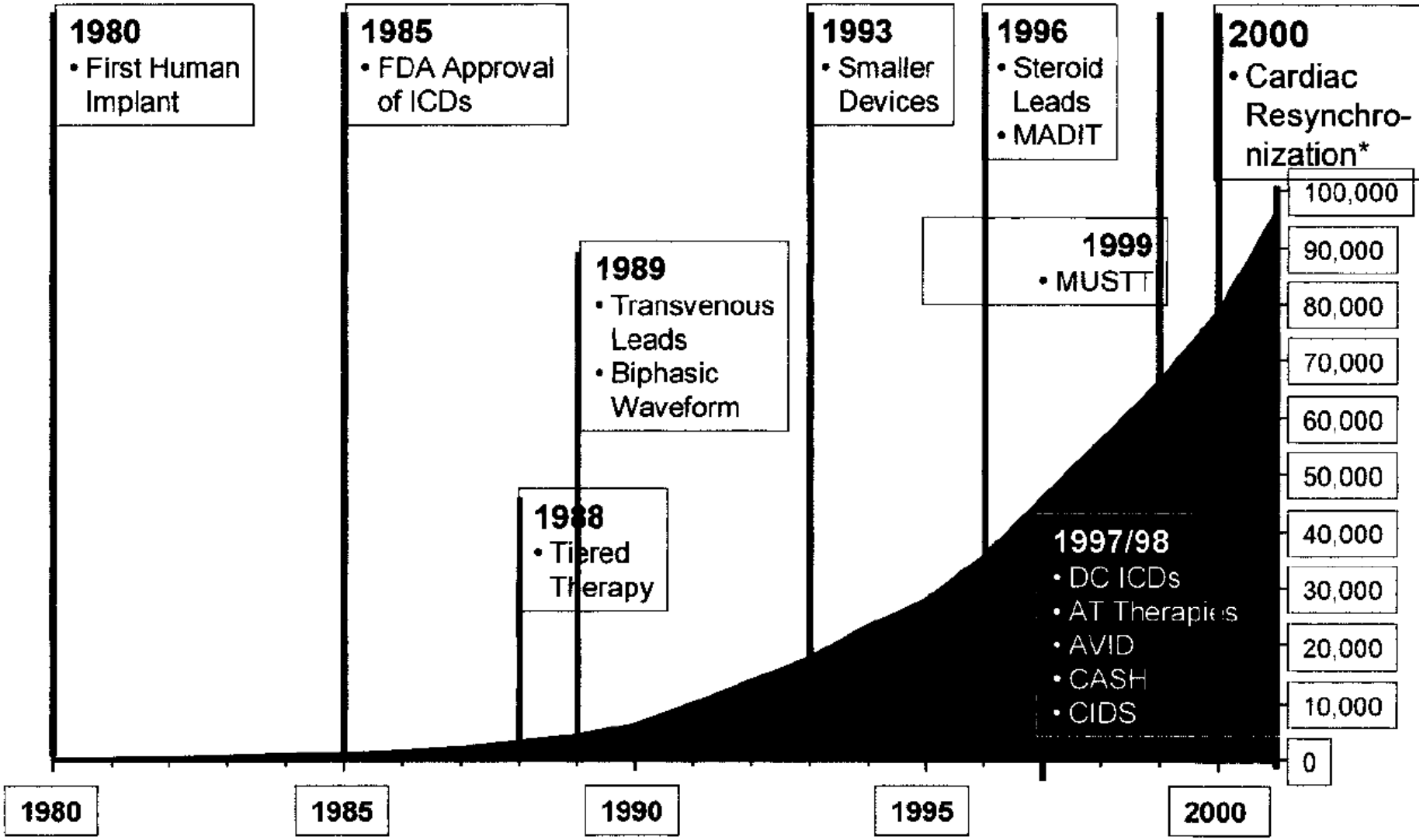
Gatilho + substrato

Instabilidade elétrica

Taquicardia ventricular

Fibrilação ventricular

Evolução da Indicação de CDI na população USA

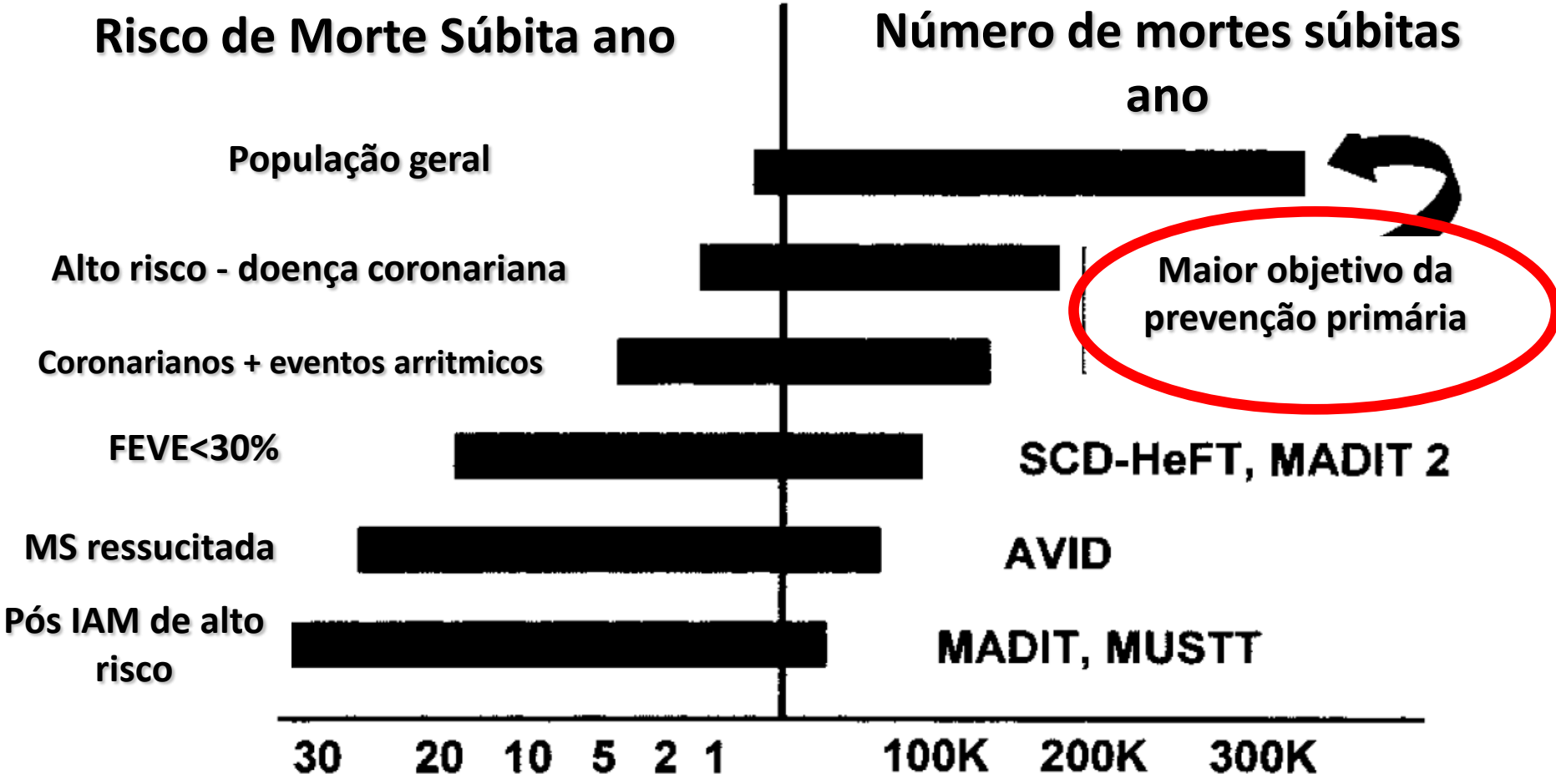


Circulation -2004; 109: 2685-2691

Morte cardíaca súbita

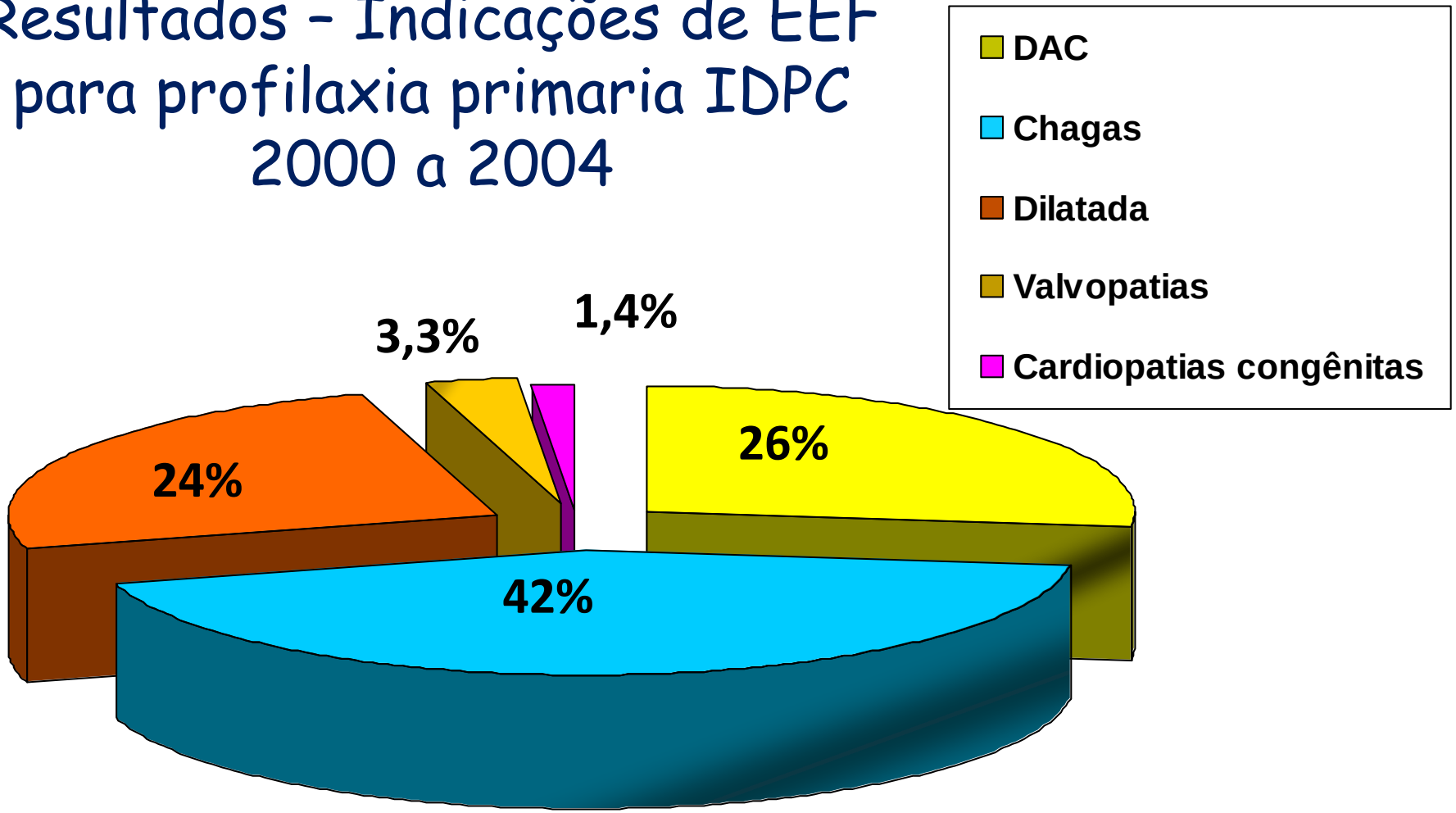
ALBERT EINSTEIN
HOSPITAL ISRAELITA

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



Circulation -2004; 109: 2685-2691

Resultados – Indicações de EEF para profilaxia primaria IDPC 2000 a 2004

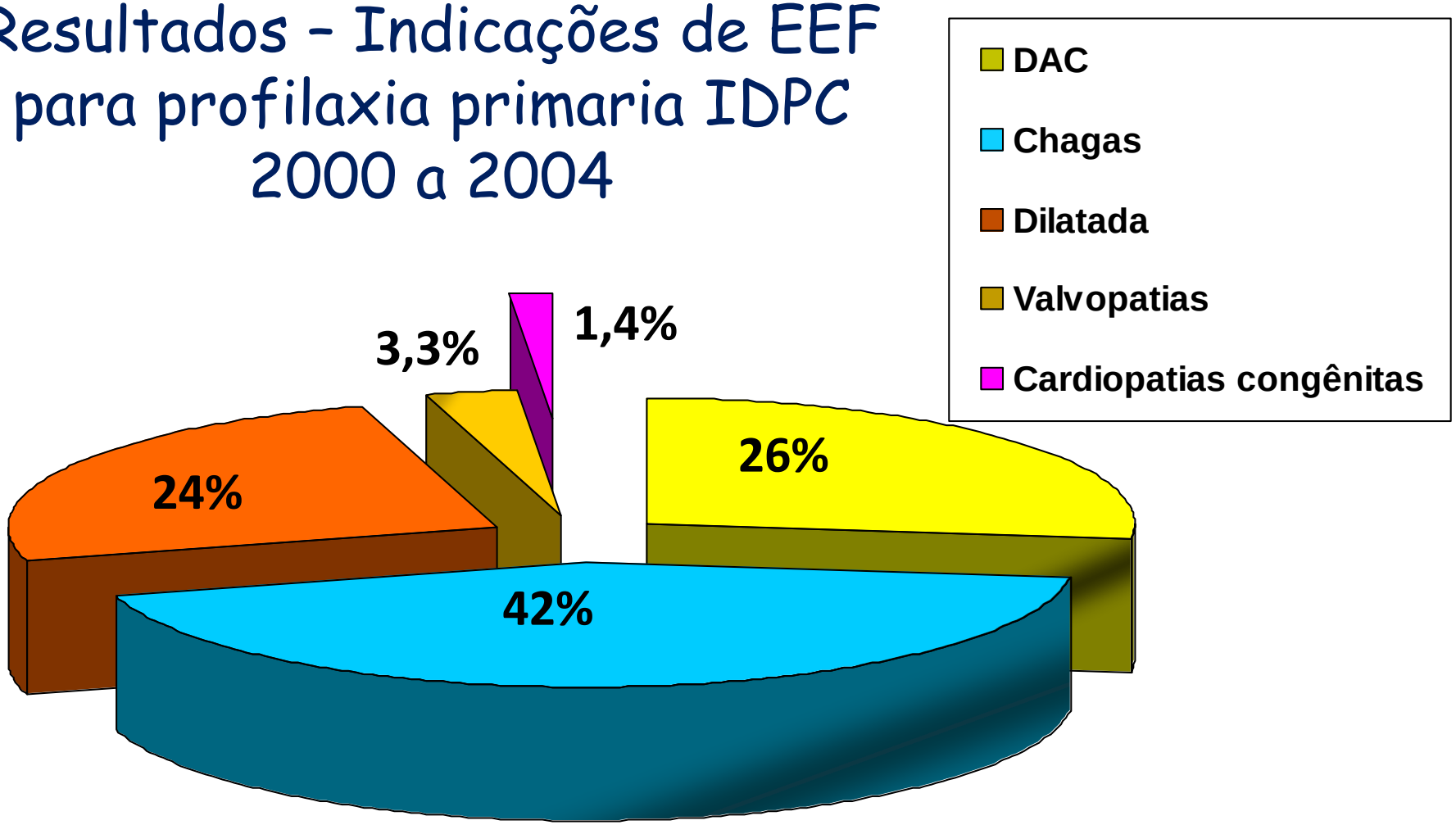


ALBERT EINSTEIN
HOSPITAL ISRAELITA

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

Andalaft et al Socesp 2004

Resultados – Indicações de EEF para profilaxia primaria IDPC 2000 a 2004



A presença de beta bloqueadores e amiodarona não influenciou na indução de TVS e FV (p=0,252), assim como não melhorou a tolerabilidade destes pacientes à arritmia (p=0,99).

Andalaft et al Socesp 2004

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

Table 1 Previous Pediatric ICD Retrospective Series

First Author (Ref. #)	Year Published	No. of Patients	Study Type
Silka (1)	1993	125	Manufacturers' datasets and multicenter retrospective
Kron (2)	1994	17	Manufacturers' datasets
Hamilton (3)	1996	11	Single-center
Link (4)	1999	11 (vs. 309 adults)	Single-center
Stefanelli (5)	2002	27	Single-center
Alexander (6)	2004	76 (included here)	Single-center
Gradaus (7)	2004	25	Single-center
Goel (8)	2004	12	Single-center
Werner (9)	2004	7	Single-center
Korte (10)	2004	20	Single-center
Ten Harkel (11)	2005	23	Multi-center
Apitz (12)	2006	12	Single-center
Eicken (13)	2006	16	Single-center
Kaski (14)	2007	22	Single-center

(J Am Coll Cardiol 2008;51:1685–91)

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

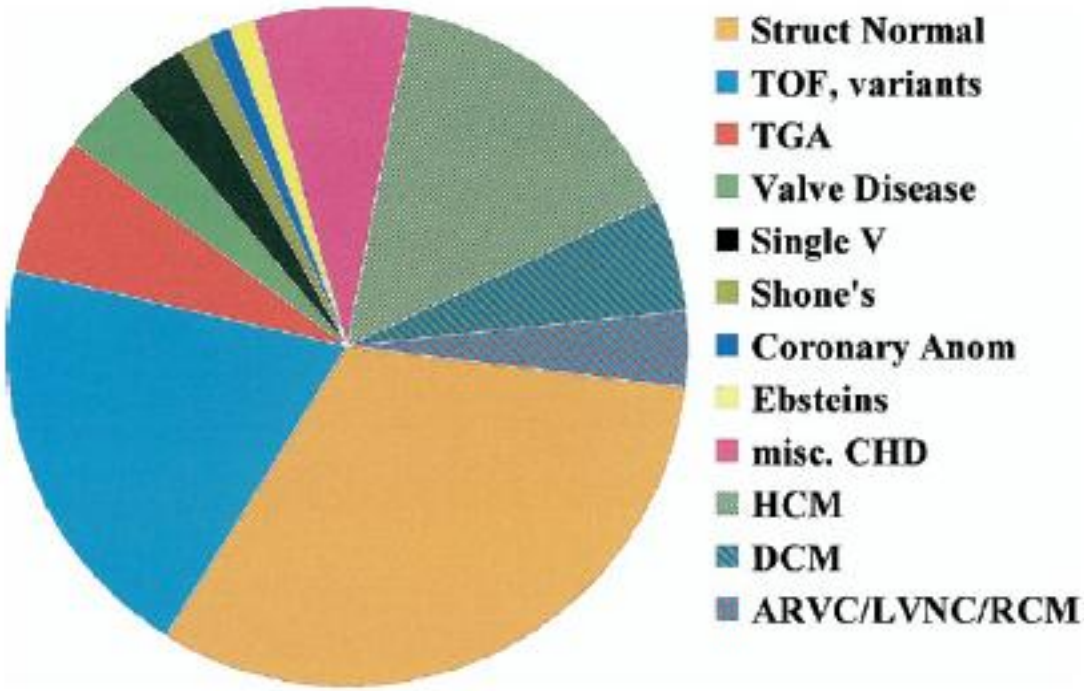
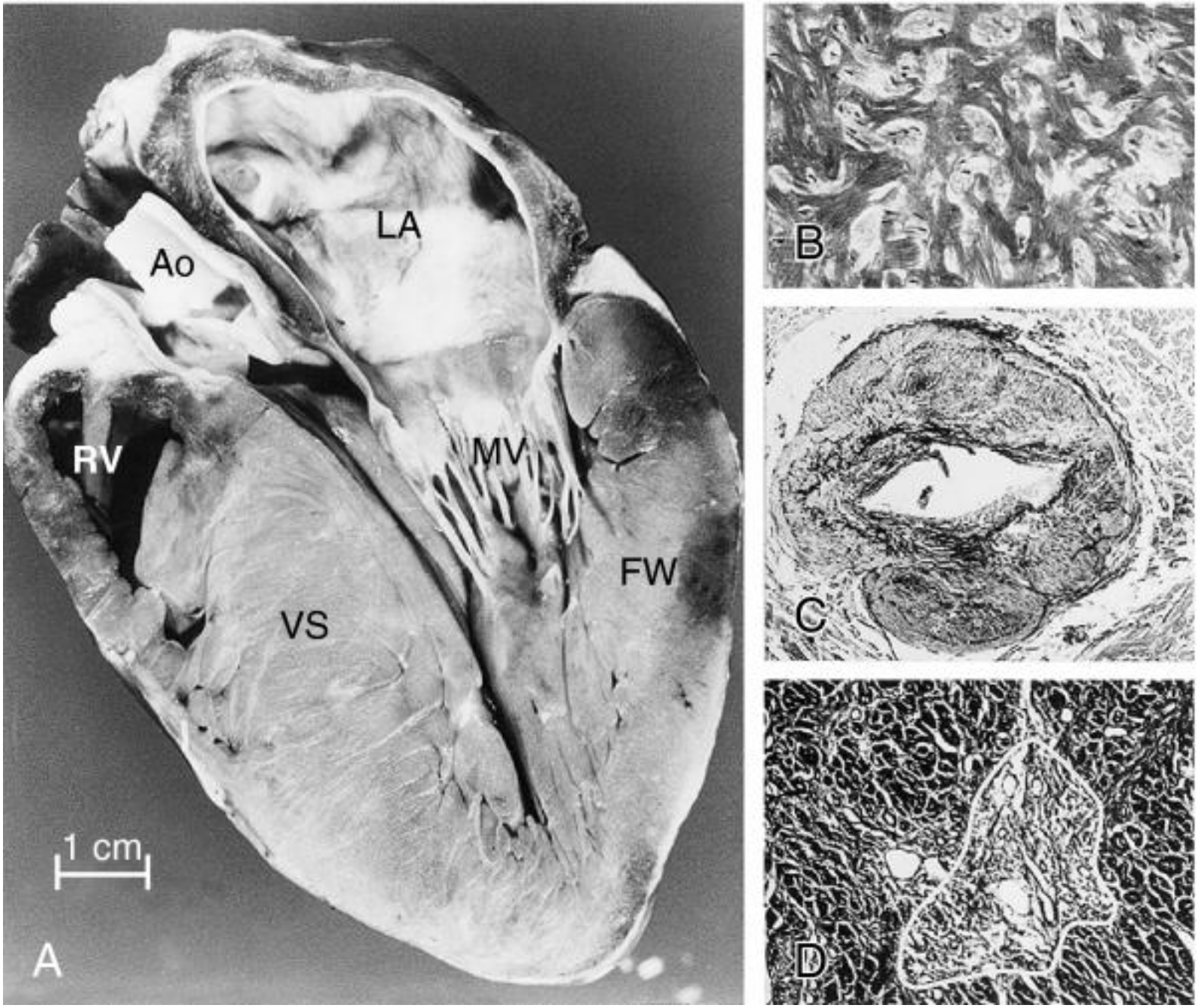
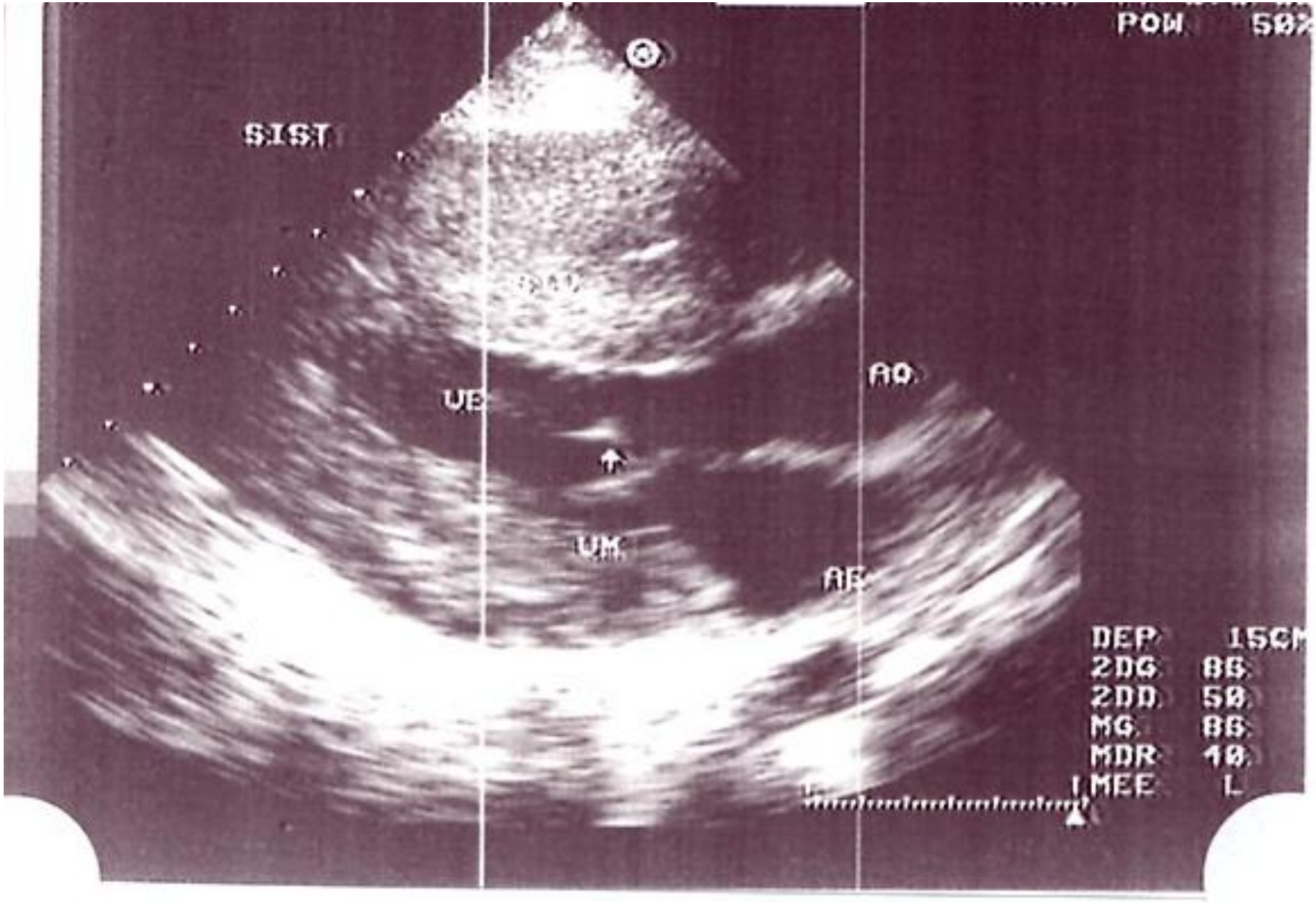


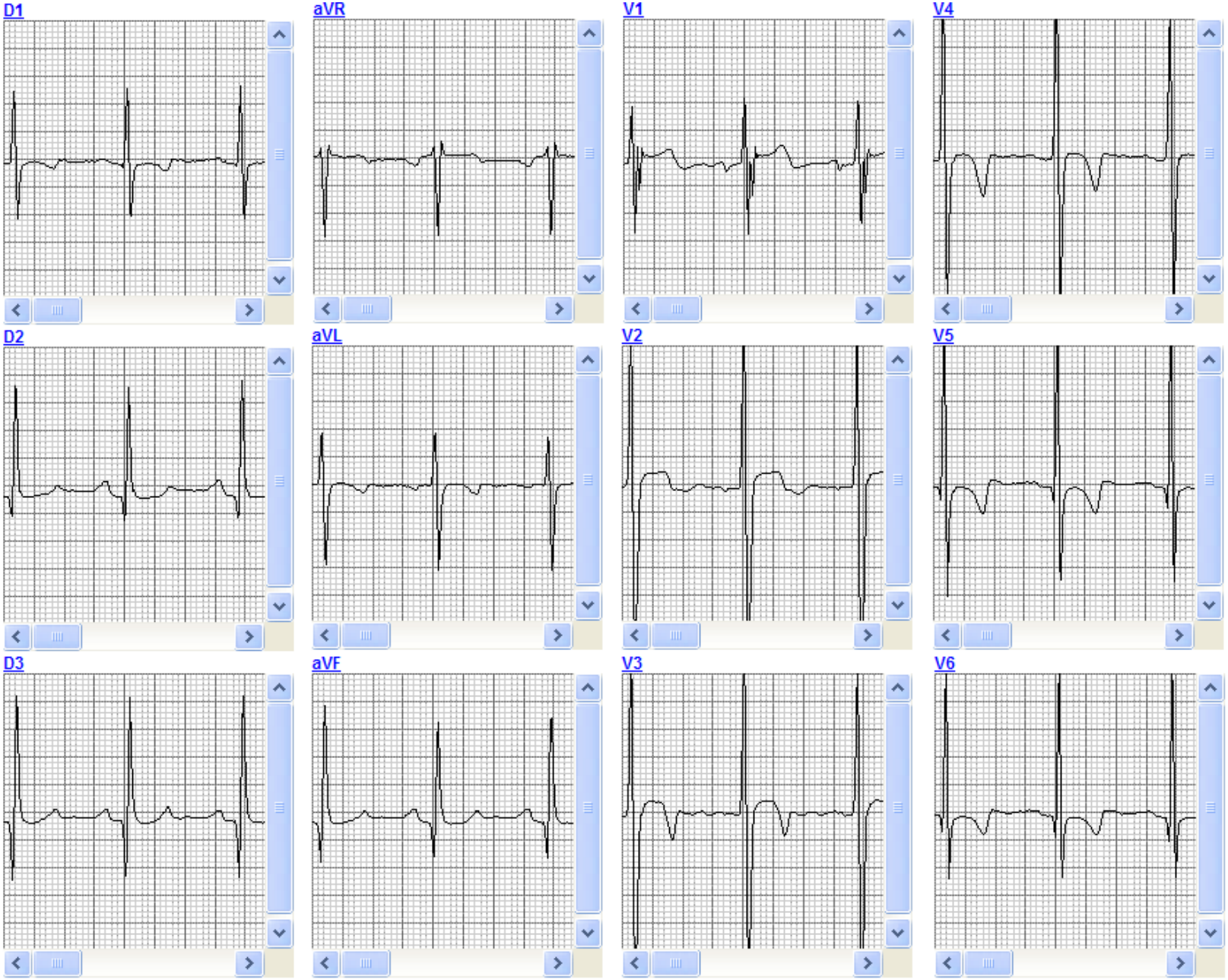
Figure 1 Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients

(J Am Coll Cardiol 2008;51:1685–91)

Miocardiopatia Hipertrófica

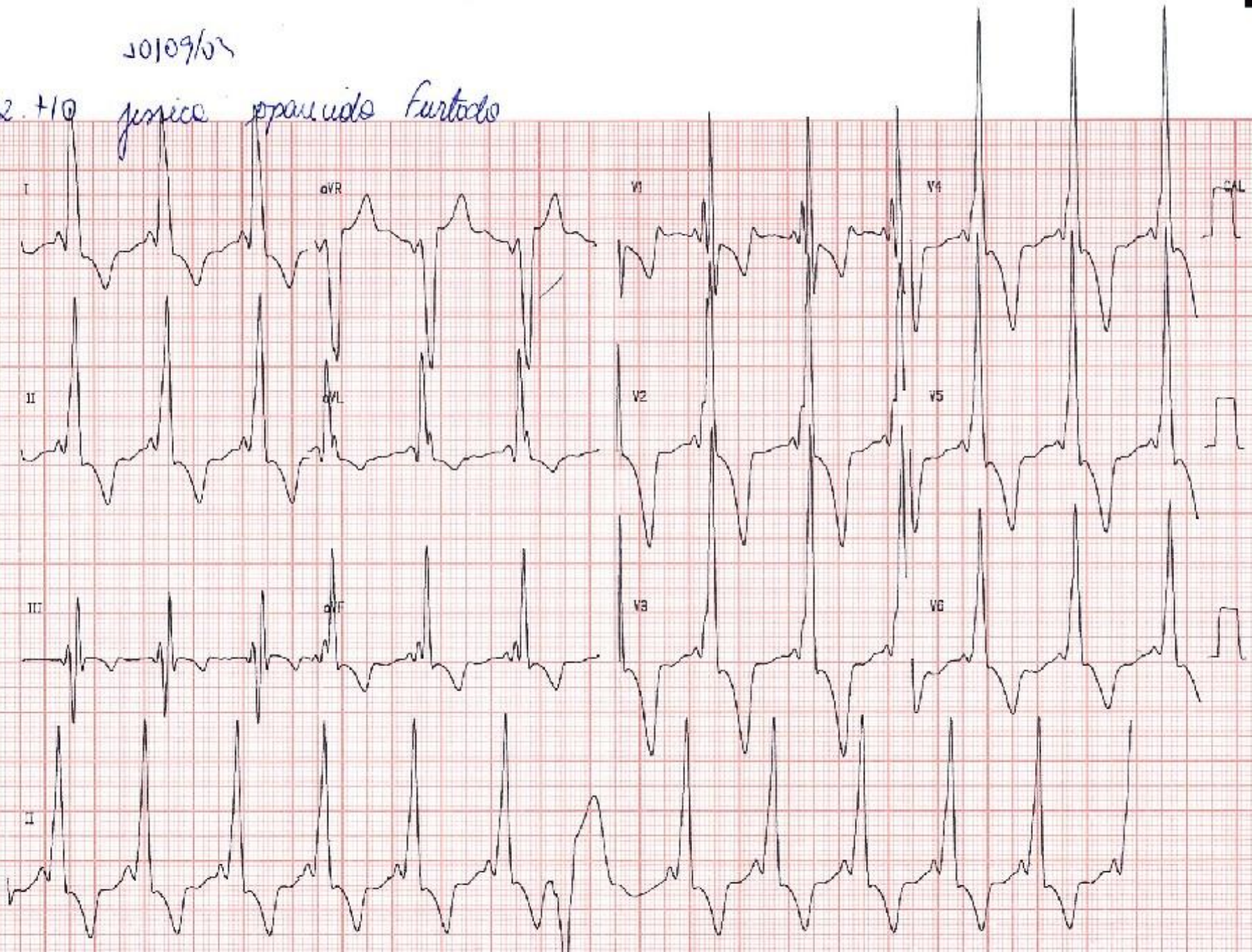

LA
Ao
RV
MV
FW
VS
1 cm
A
B
C
D


POW 50%
SIST
AO
UE
UM
AE
DEP 15CM
2DG 86
2DD 50
MG 86
MDR 40
MEE L

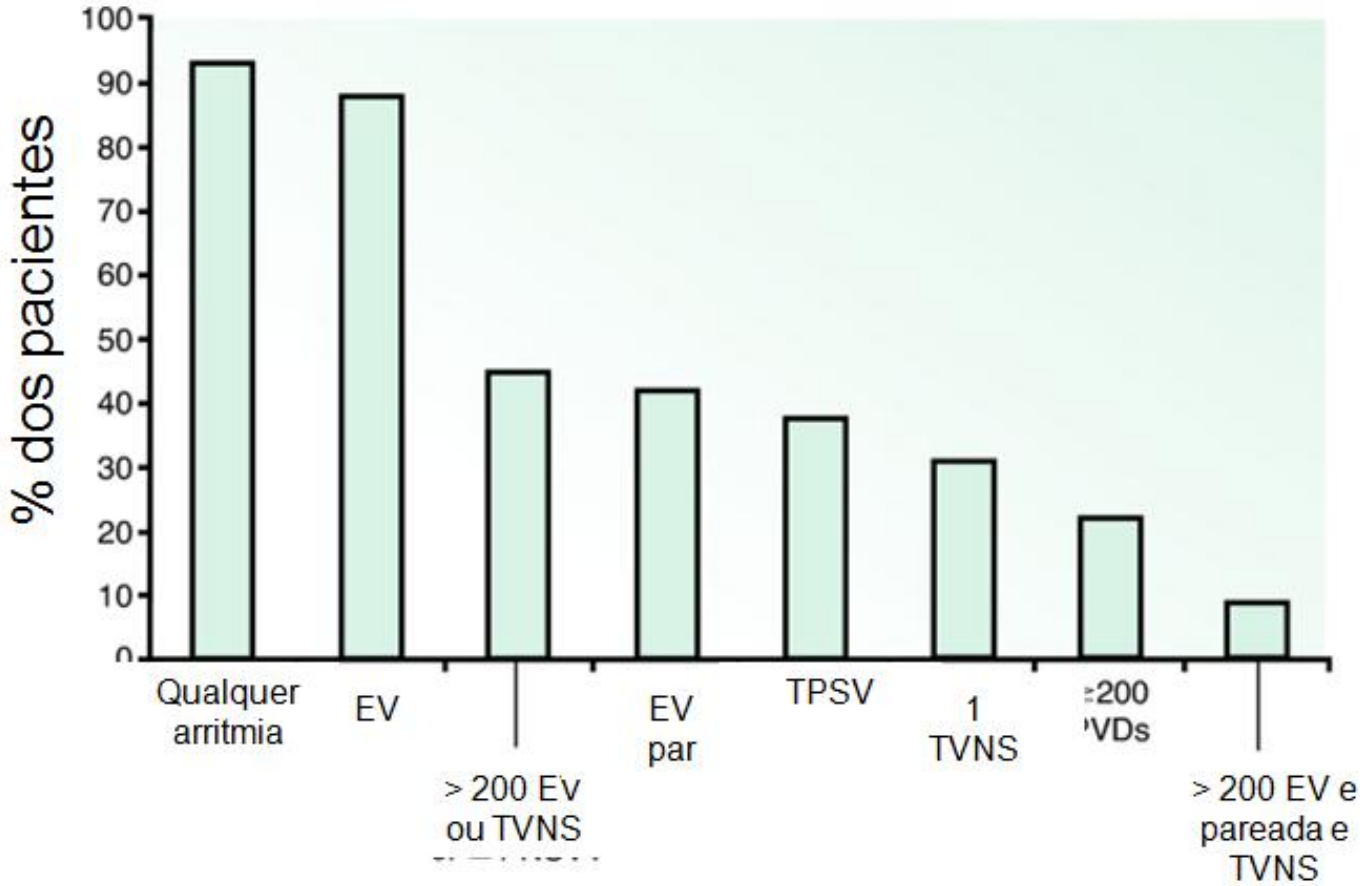

D1
aVR
V1
V4
D2
aVL
V2
V5
D3
aVF
V3
V6

30/09/03

532.710 Jessica Aparecida Furtado



J Am Coll Cardiol 45:697-704, 2005



Morte súbita e cardiomiopatia hipertrófica

Parada cardíaca

- Arritmia ventricular
- FA paroxística
- BAVT
- Rápida condução atrioventricular por via acessória
- Isquemia miocárdica

Eur Heart J, 2001; 22 (4): 1397-9

Morte súbita e cardiomiopatia hipertrófica

- Estratificação de risco
  - História familiar
  - História de TV sustentada ou parada cardíaca ressucitada
  - Sintomas
  - Preditores ecocardiográficos
  - Resposta da PA ao teste de esforço
  - Holter – TVNS
  - Anormalidades genéticas

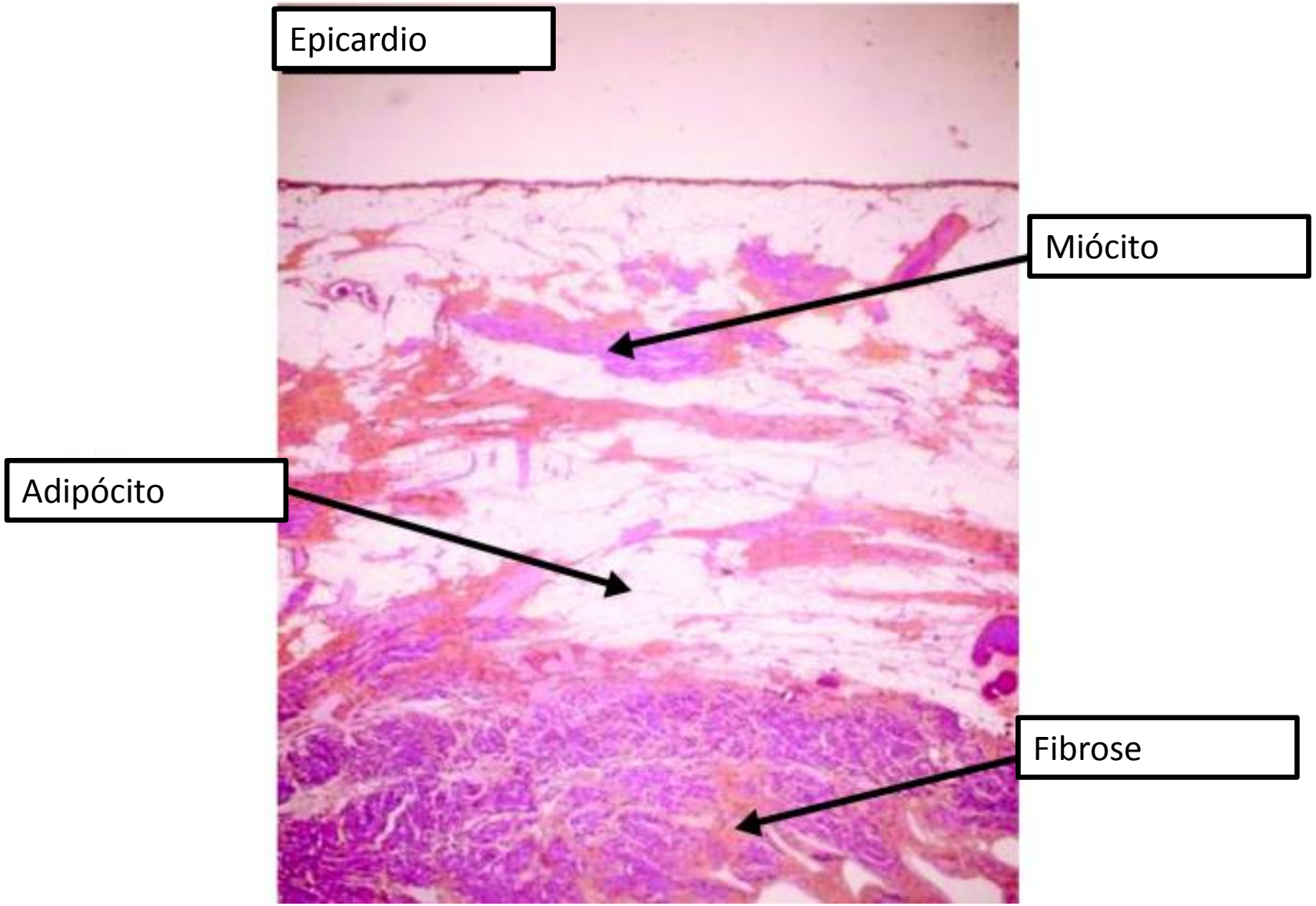
Eur Heart J, 2001; 22 (4): 1397-9

Estratificação de risco miocardiopatia hipertrófica

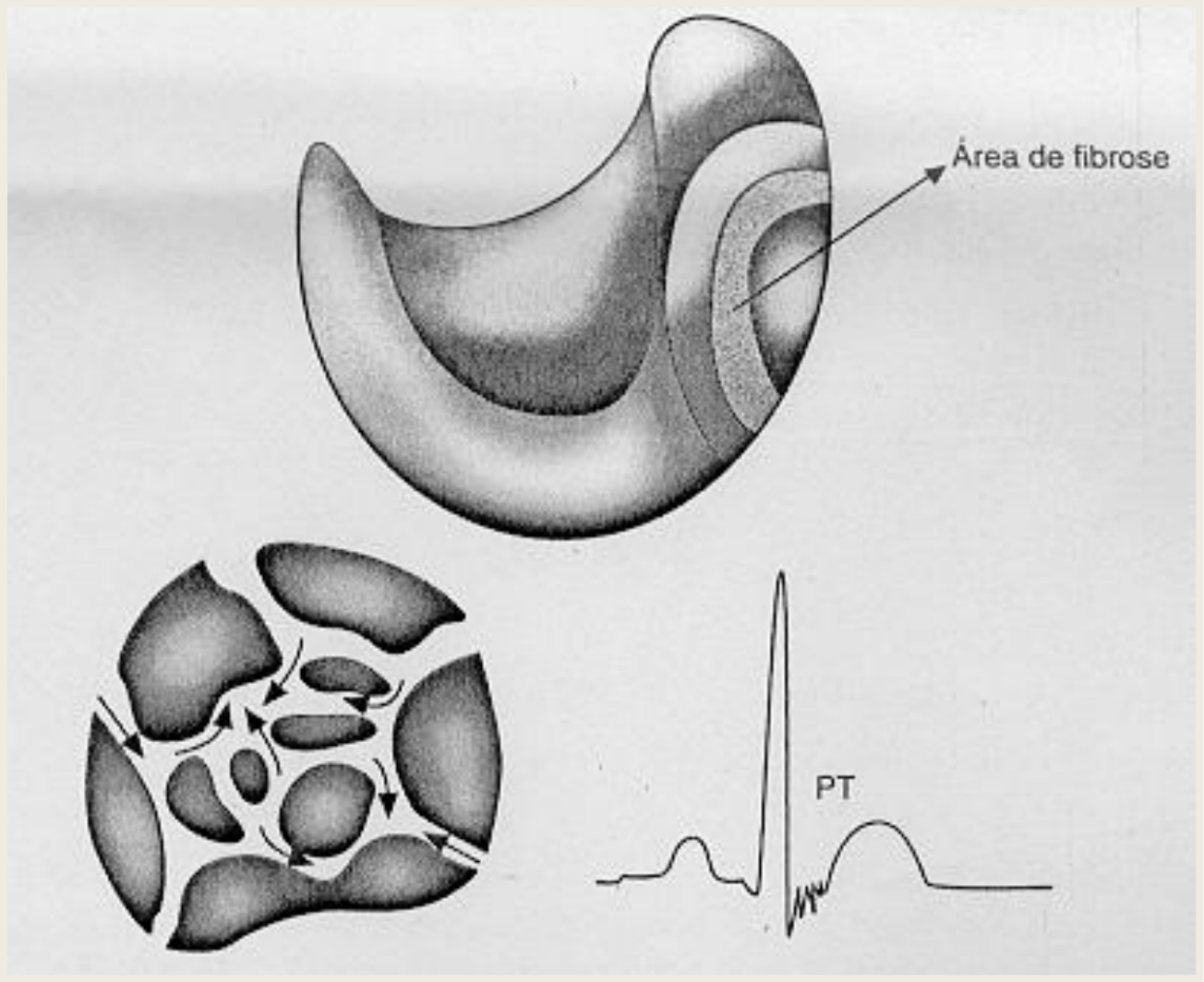
	I	IIa	IIb
Estratificação de Risco	TV/FV	História fam MS Síncope Septo>30mm TVNS Hipotensão TE	Mutações de alto risco
Prevenção Primária	-	CDI	Amiodarona
Prevenção Secundária	CDI	-	-

Europace 2002; 4:3-18

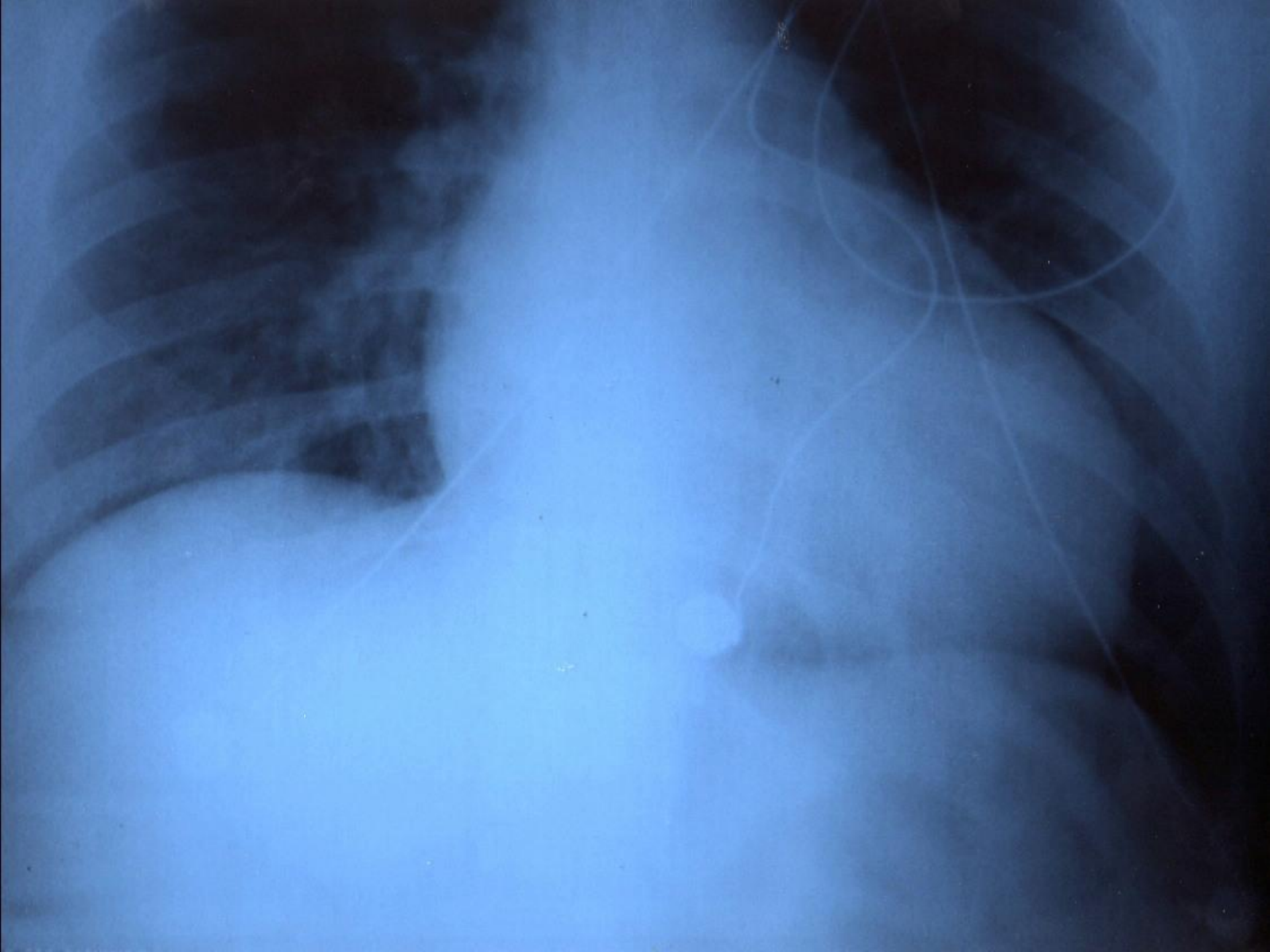
Displasia arritmogênica do VD


Epicardio
Miócito
Adipócito
Fibrose

Áreas de fibrose ou gordura entremeadas com tecido sadio



Moffa PJ, Sanches PCR. Eletrocardiograma p.881-90



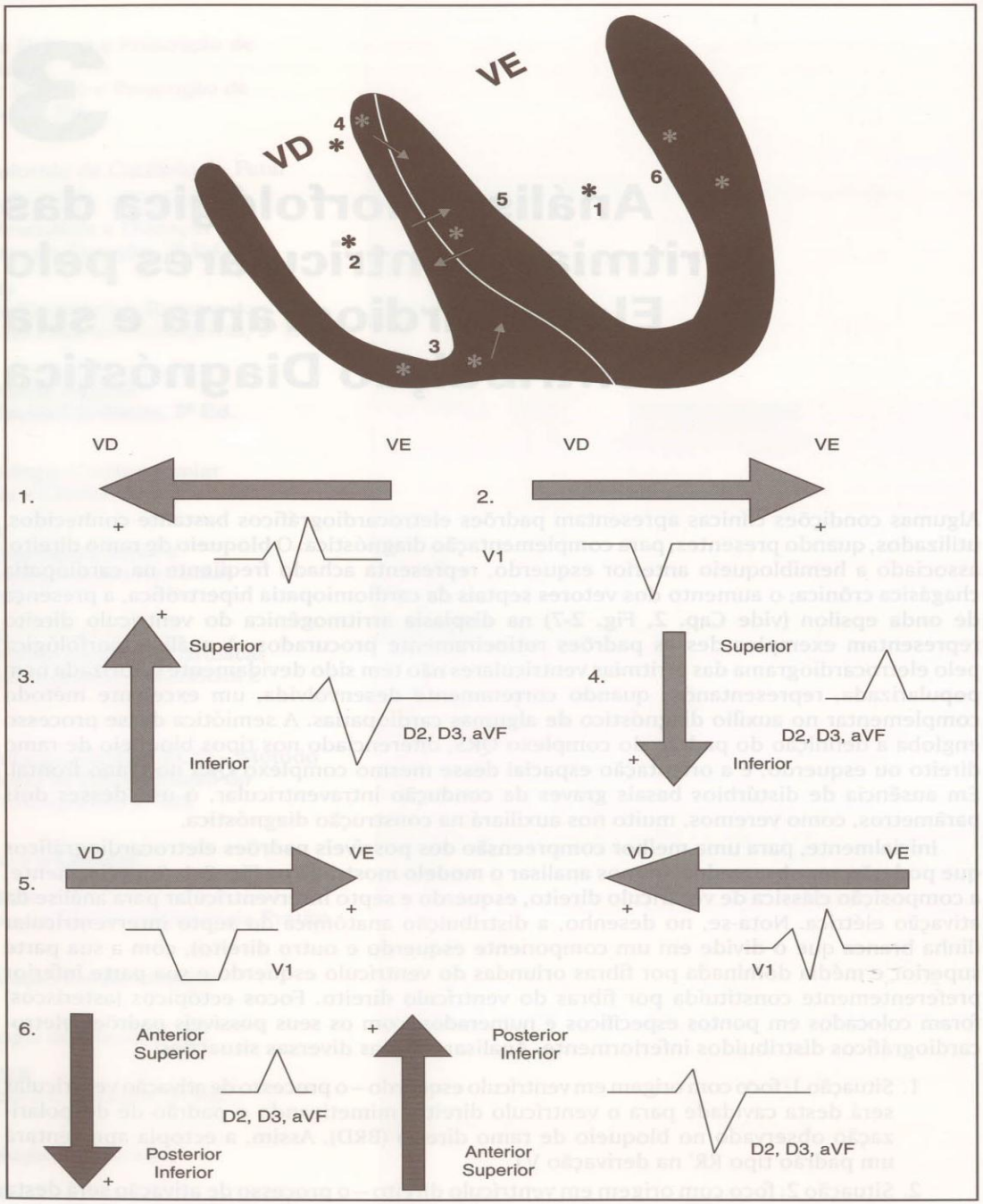


Fig. 3-1. Relações entre ectopias ventriculares e local de origem.

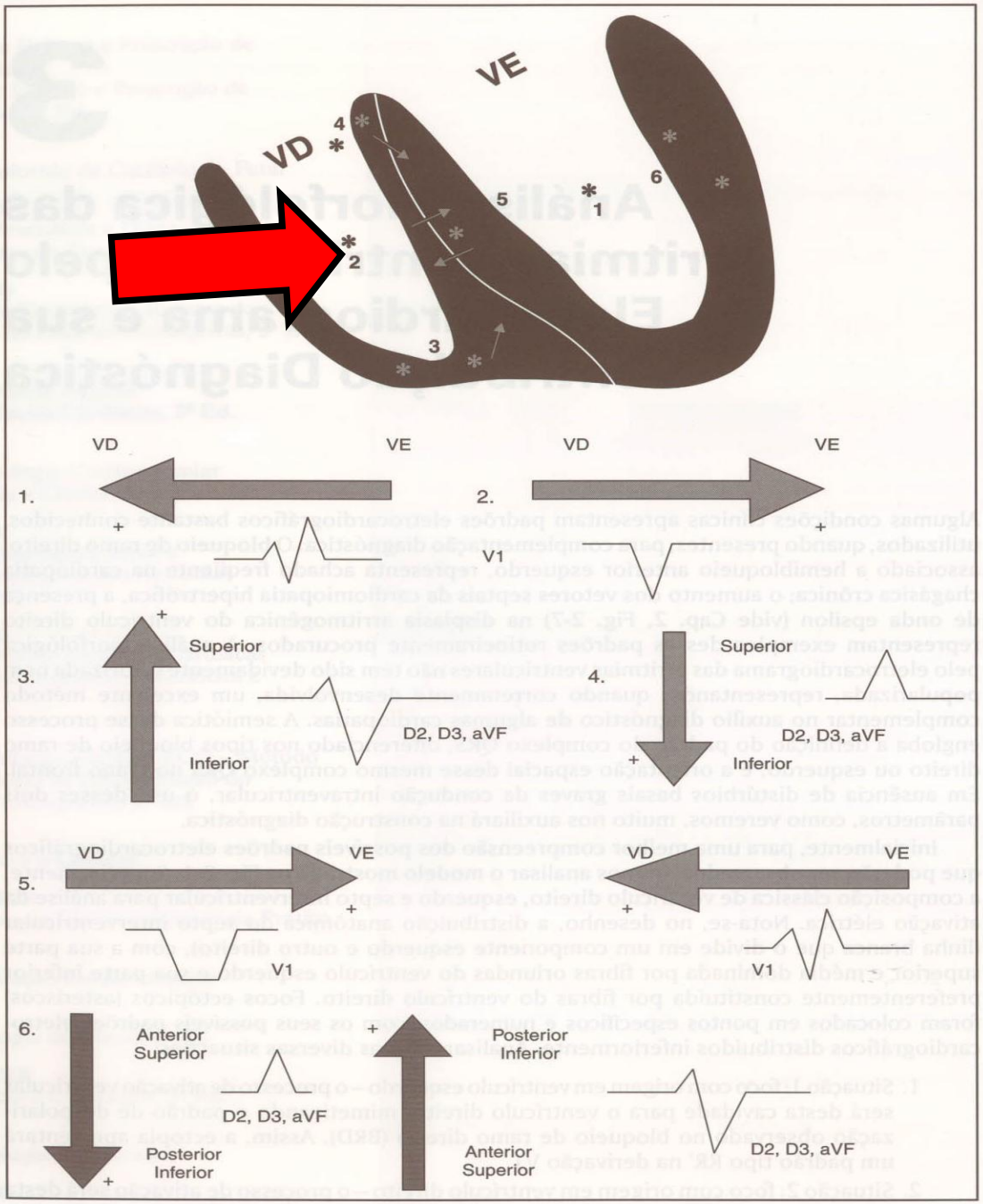
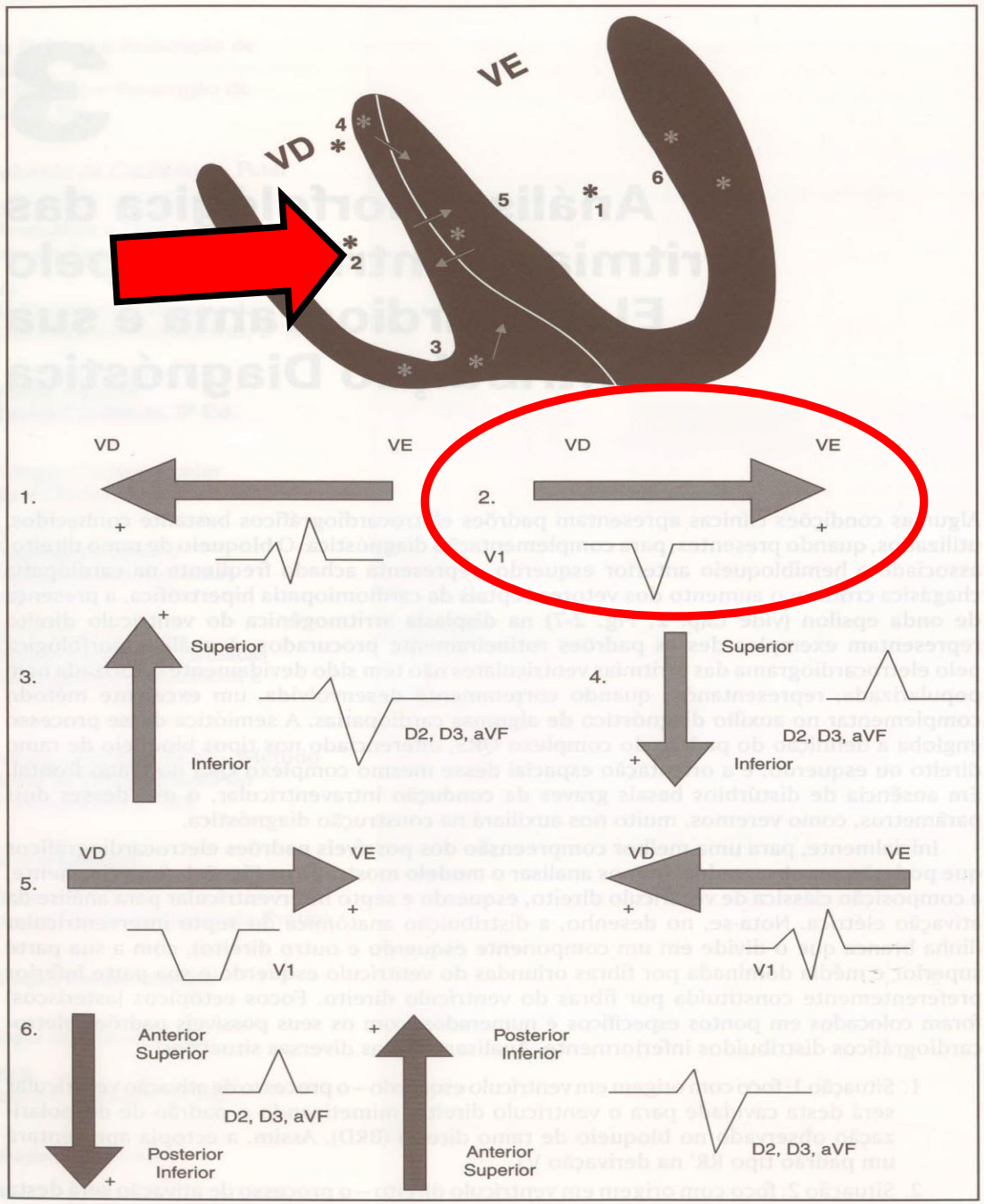
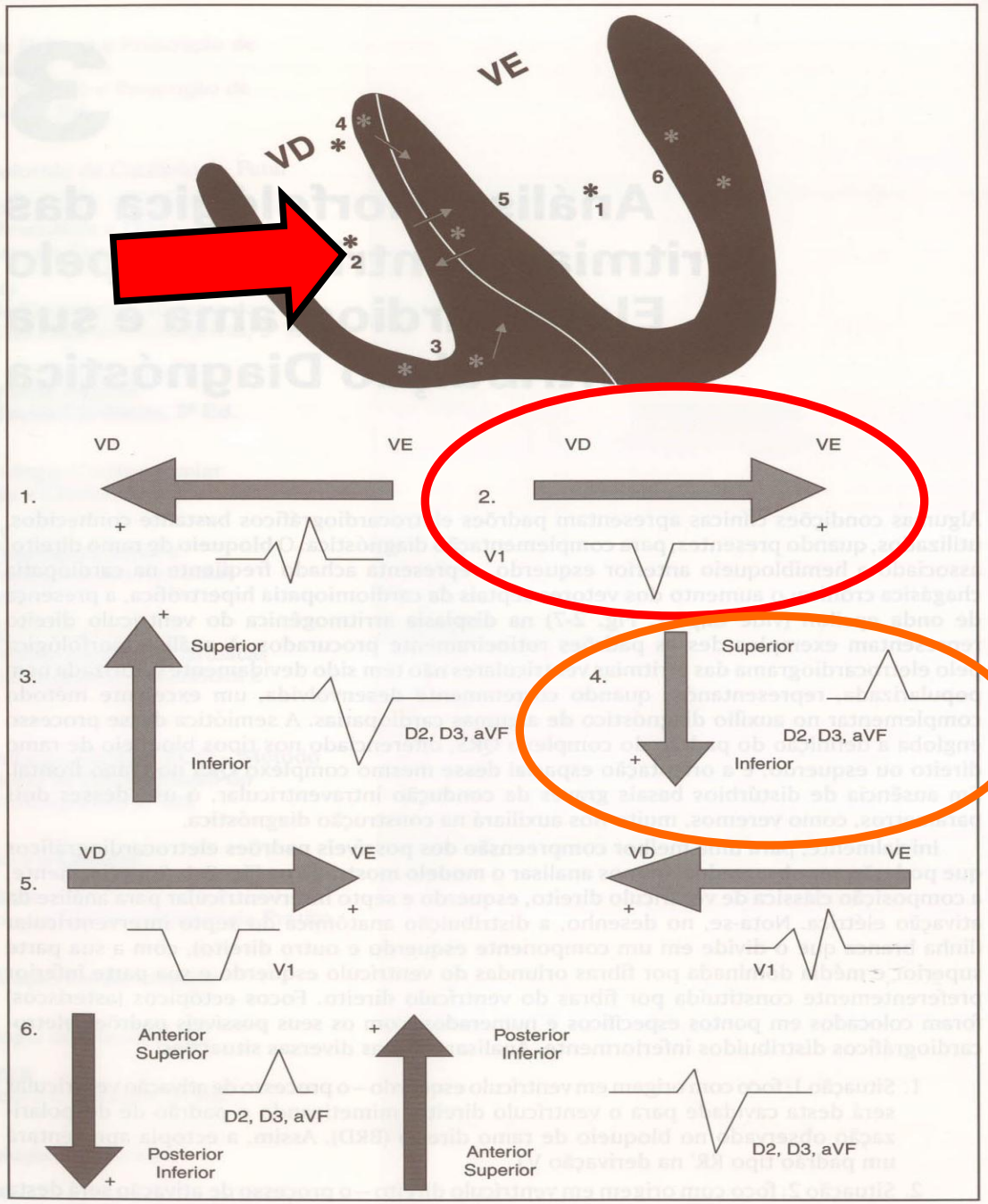


Fig. 3-1. Relações entre ectopias ventriculares e local de origem.



Ventrículo Direito

Fig. 3-1. Relações entre ectopias ventriculares e local de origem.

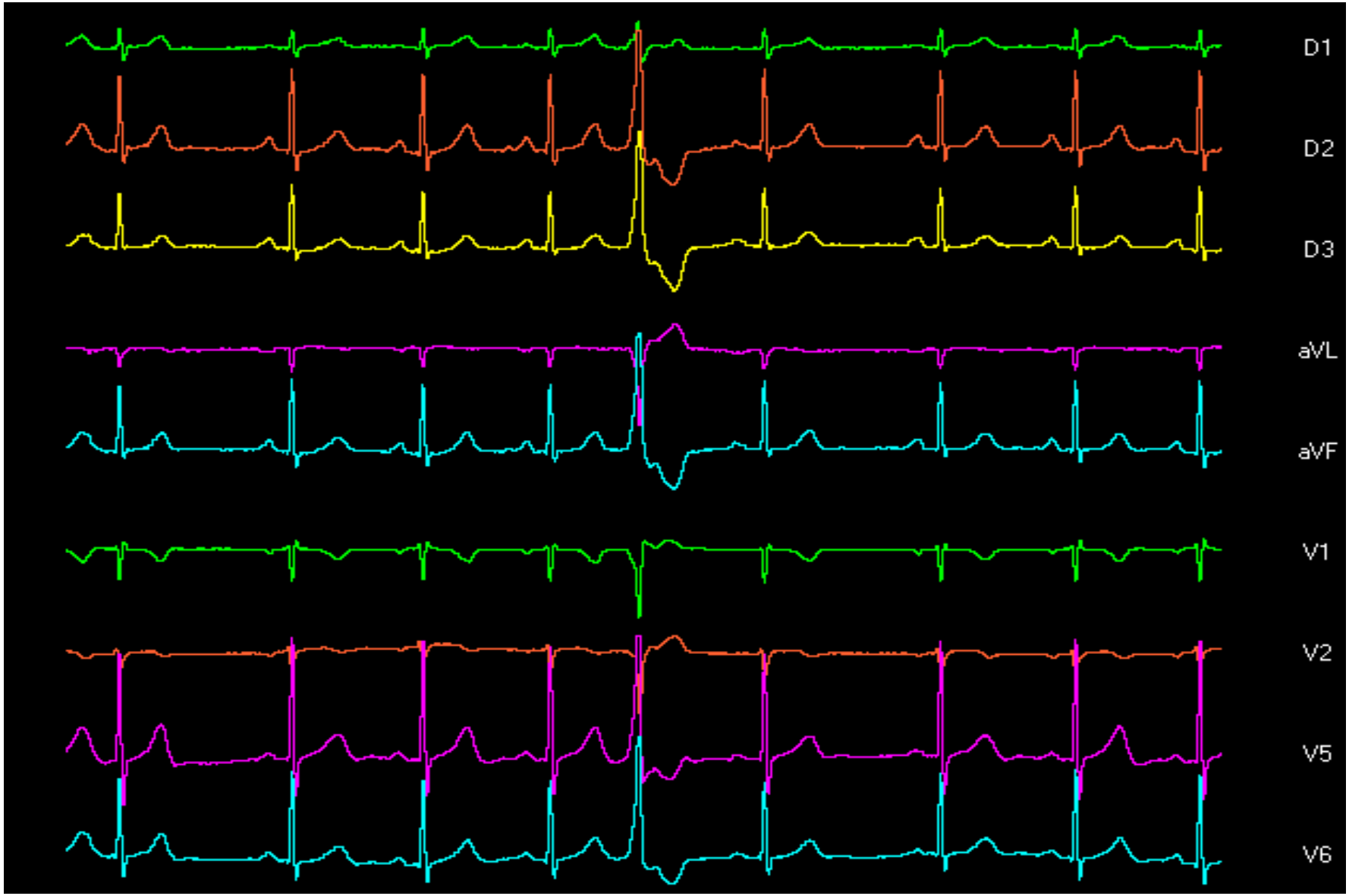


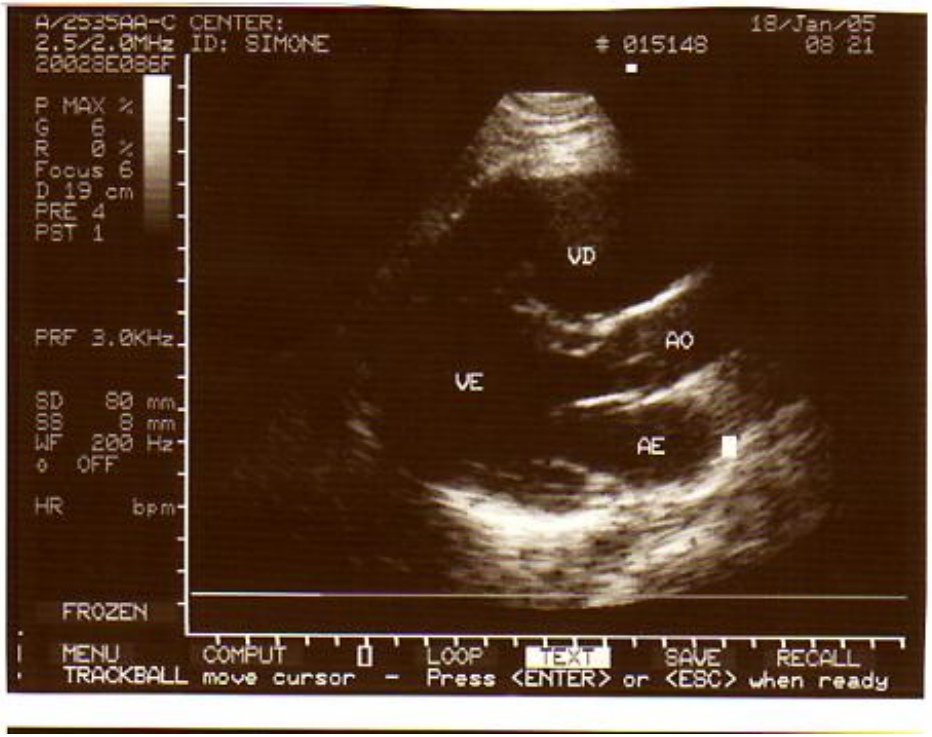
Ventrículo Direito

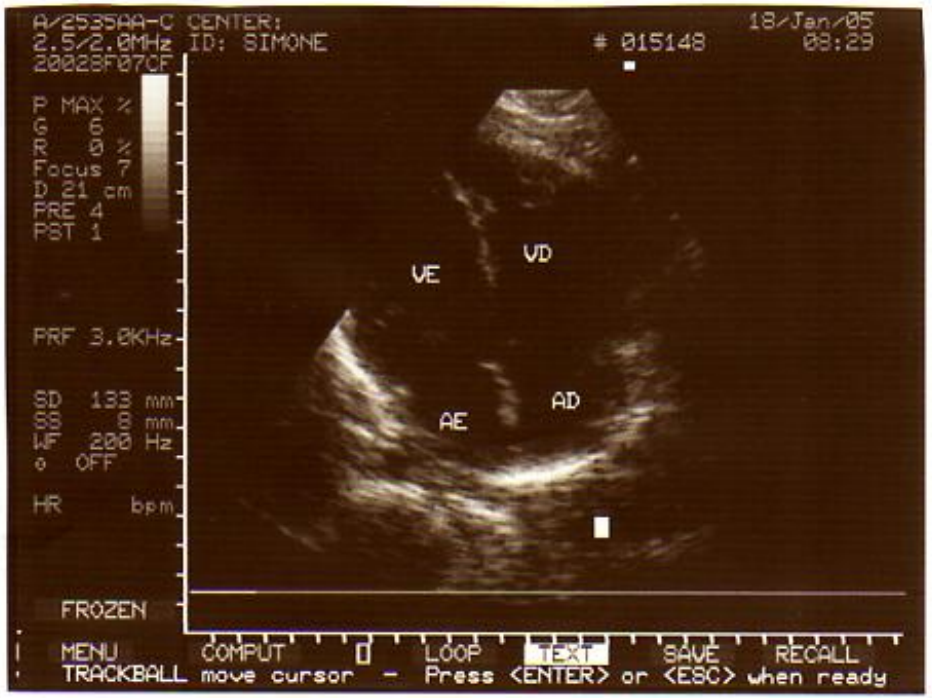
Via de saída

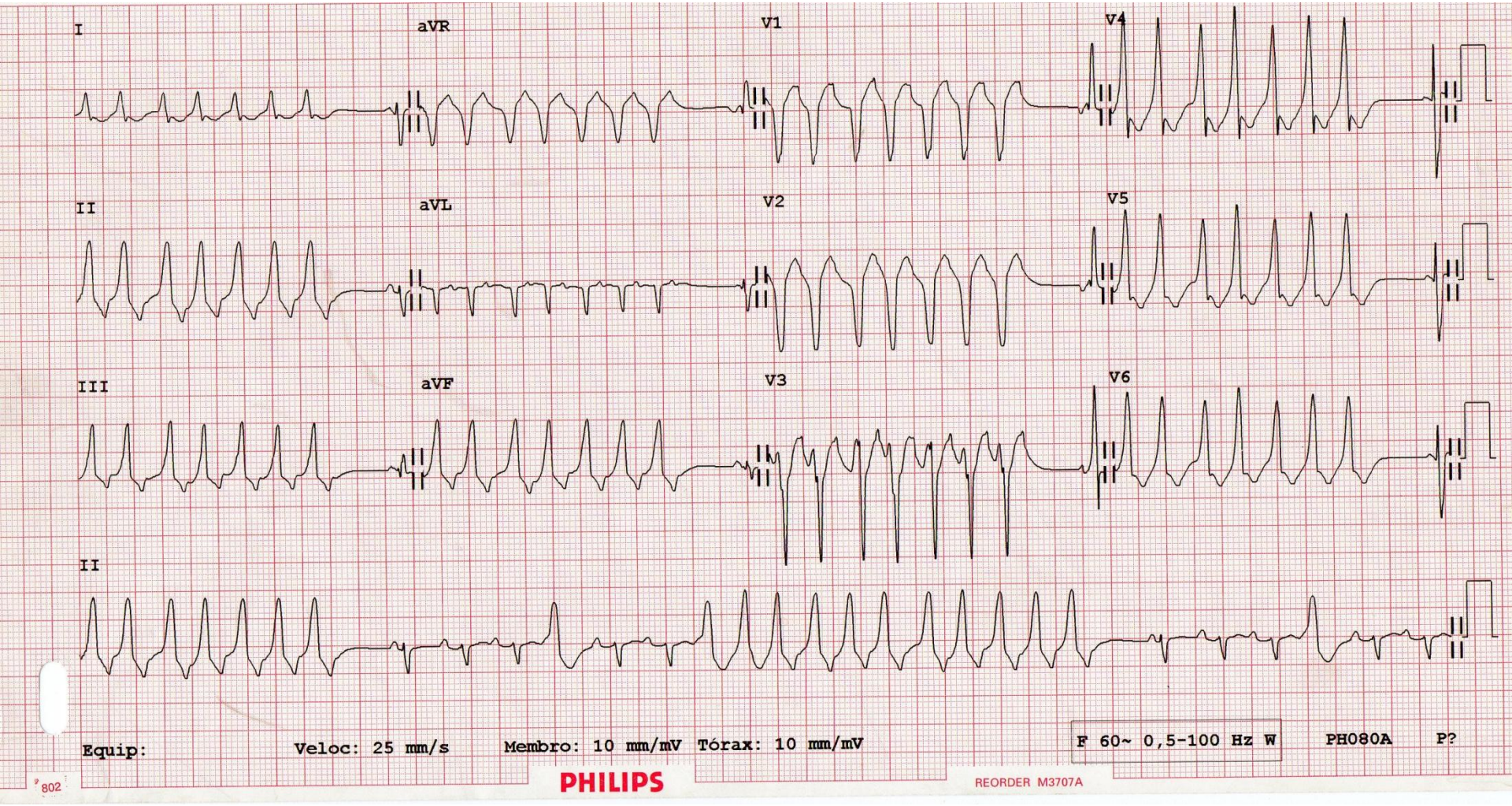
Fig. 3-1. Relações entre ectopias ventriculares e local de origem.

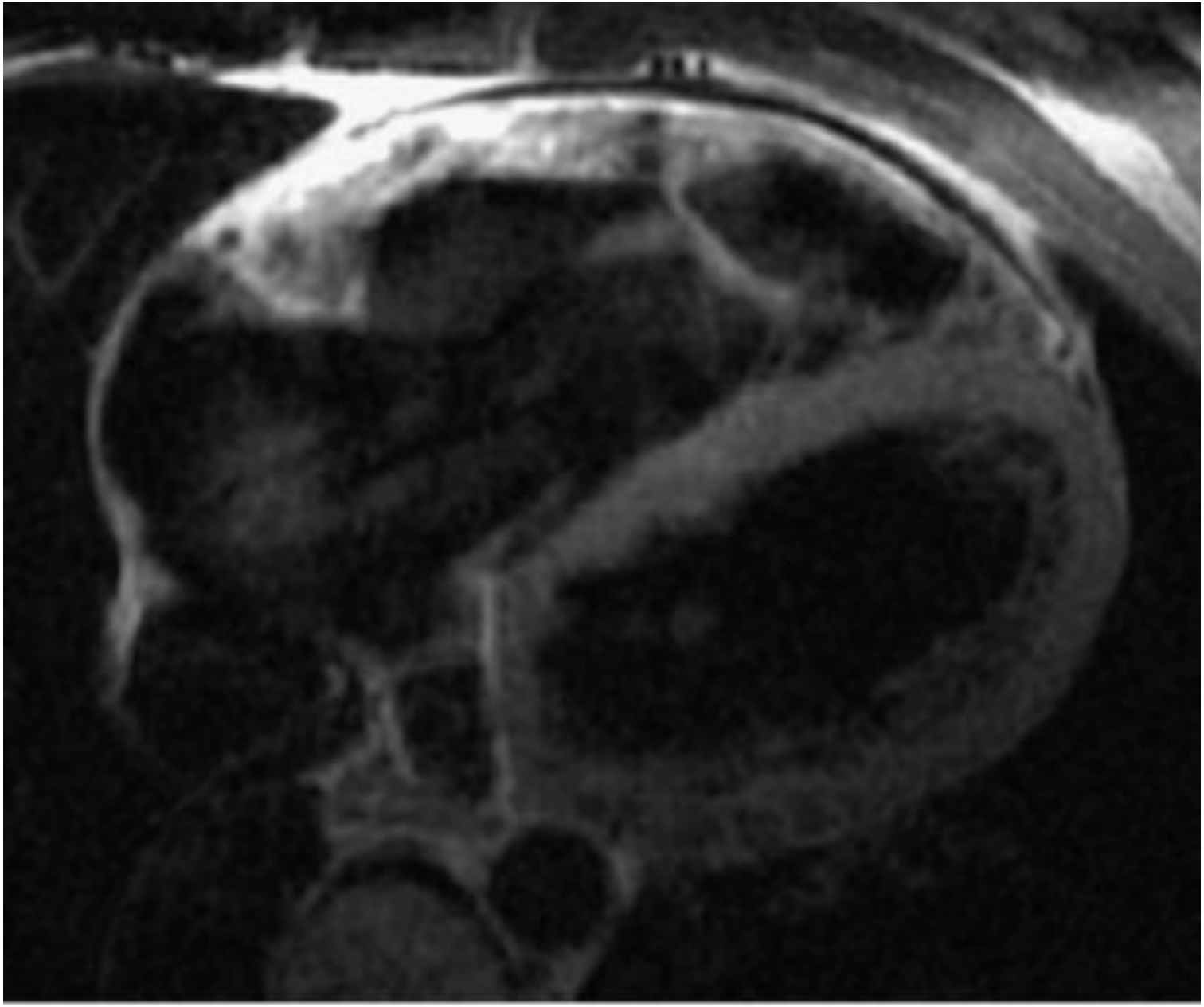
ARRITMIA DE V.D.




A/2535AA-C CENTER:
2.5/2.0MHz ID: SIMONE
20028E086F
015148
18/Jan/05
08 21
P MAX %
G 6
R 0 %
Focus 6
D 19 cm
PRE 4
PST 1
PRF 3.0KHz
SD 80 mm
SS 8 mm
WF 200 Hz
OFF
HR bpm
VD
AO
VE
AE
FROZEN
MENU
COMPUT
LOOP
TEXT
SAVE
RECALL
TRACKBALL move cursor - Press <ENTER> or <ESC> when ready


A/2535AA-C CENTER:
2.5/2.0MHz ID: SIMONE
20028F07CF
015148
18/Jan/05
08:29
P MAX %
G 6
R 0 %
Focus 7
D 21 cm
PRE 4
PST 1
PRF 3.0KHz
SD 133 mm
SS 8 mm
WF 200 Hz
OFF
HR bpm
VD
VE
AD
AE
FROZEN
MENU
COMPUT
LOOP
TEXT
SAVE
RECALL
TRACKBALL move cursor - Press <ENTER> or <ESC> when ready


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
Equip:
Veloc: 25 mm/s
Membro: 10 mm/mV
Tórax: 10 mm/mV
F 60~ 0,5-100 Hz W
PH080A
P?
802
PHILIPS
REORDER M3707A



Cine RM - Eixo Curto 4

Diagnóstico diferencial de Displasia arritmogênica do VD

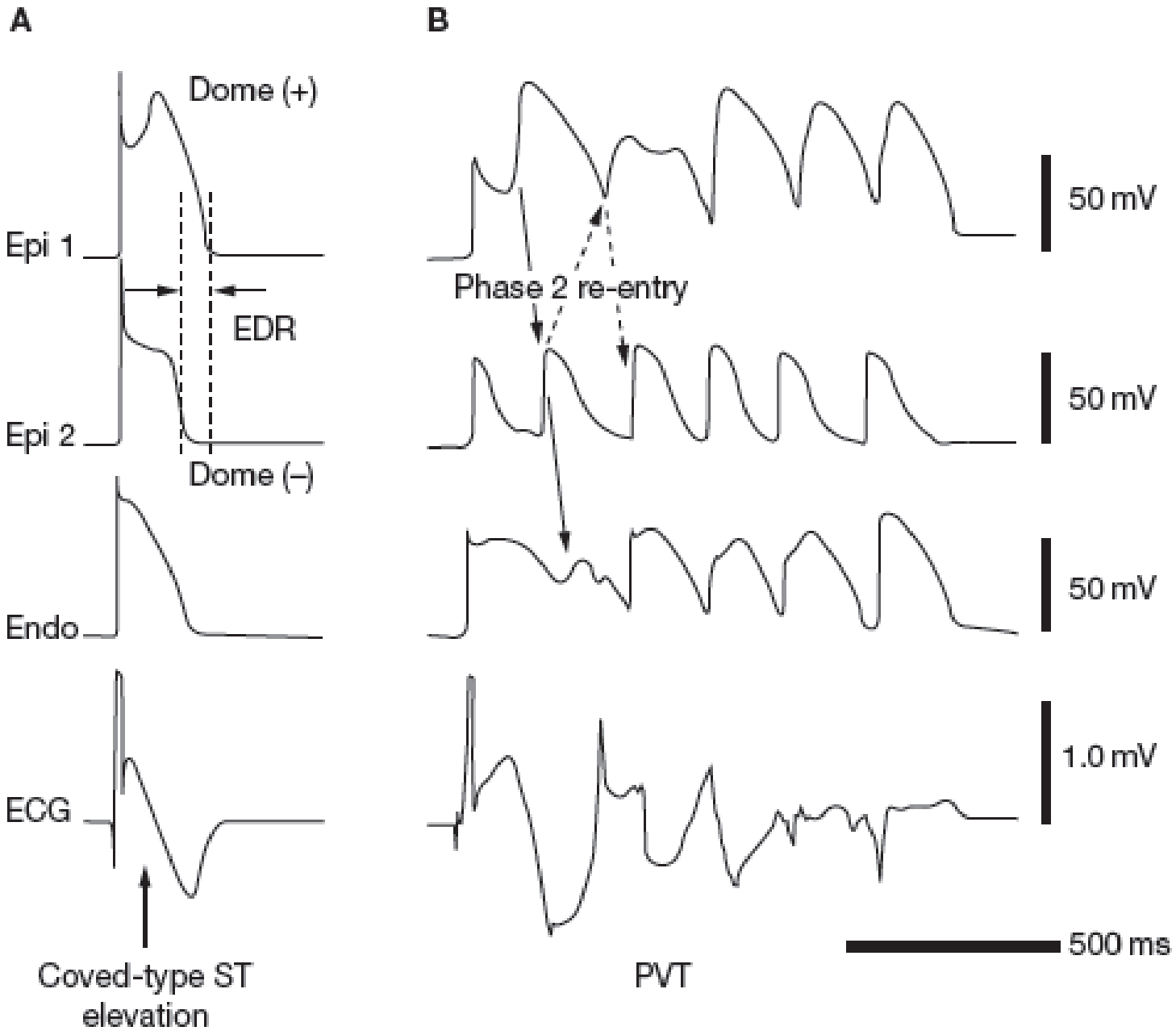
	TV idiopática de VD		DAVD
História Familiar de MS	Não		•Frequentemente sim
Arritmias	EV, TVNS, TVS Em repouso ou exercício		•idem
Morfologia onda T	T normais		•T negativas V1 a V3
Duração do QRS	Menor que 110 ms		•Maior que 110 ms
Onda Epsilon	ausente		•Presente em 30%
Nadir S final QRS > 55 ms	Raramente positivo		•Positivo em mais que 95%
ECG AR	normal		•Geralmente anormal
Ecocardiograma	normal		•Aumento ou alterações do VD
Ventriculografia	Geralmente normal		•Aumento ou alterações do VD
RNM	Geralmente normal		•Aumento ou alterações do VD

Estratificação de risco Displasia arritmogênica do ventrículo direito

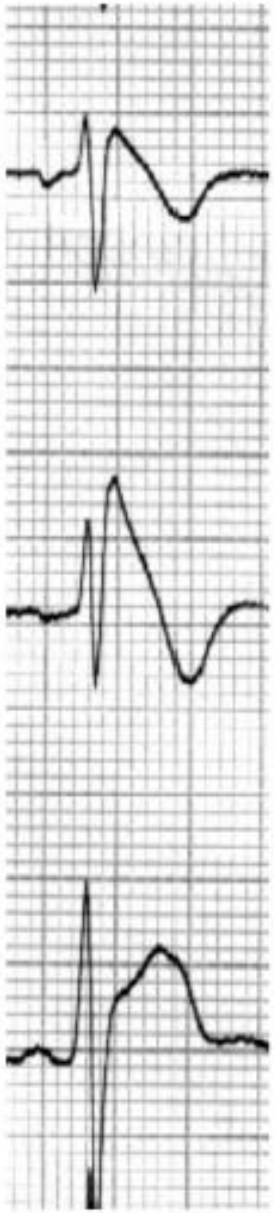
	I	IIa	IIb
Estratificação de Risco	-	•TV/FV •Dilatação do VD •Indutibilidade no EEF	•História familiar de MS •Potenciais tardios + disfunção do VD •TV •Indutibilidade no EEF
Prevenção Primária	-	CDI	•Medicações antiarrítmicas
Prevenção Secundária	CDI	-	-

Europace 2002; 4:3-18

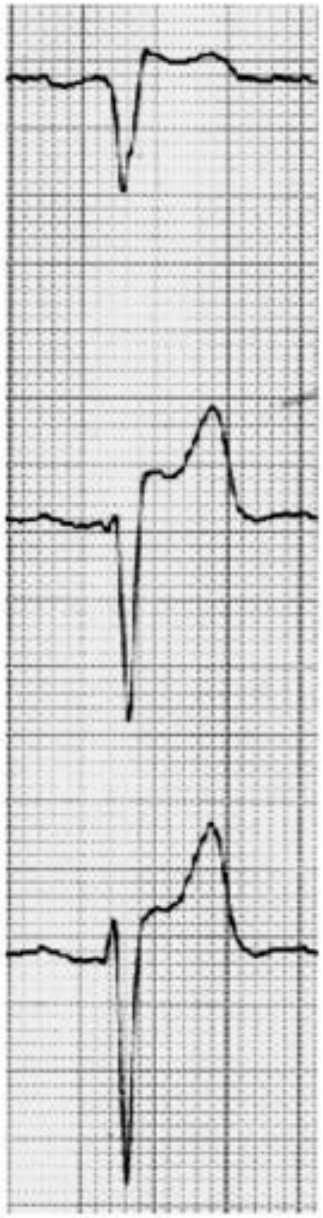
Síndrome de Brugada


A
B
Dome (+)
Epi 1
EDR
Epi 2
Dome (–)
Endo
ECG
Coved-type ST elevation
Phase 2 re-entry
50 mV
50 mV
50 mV
1.0 mV
500 ms
PVT

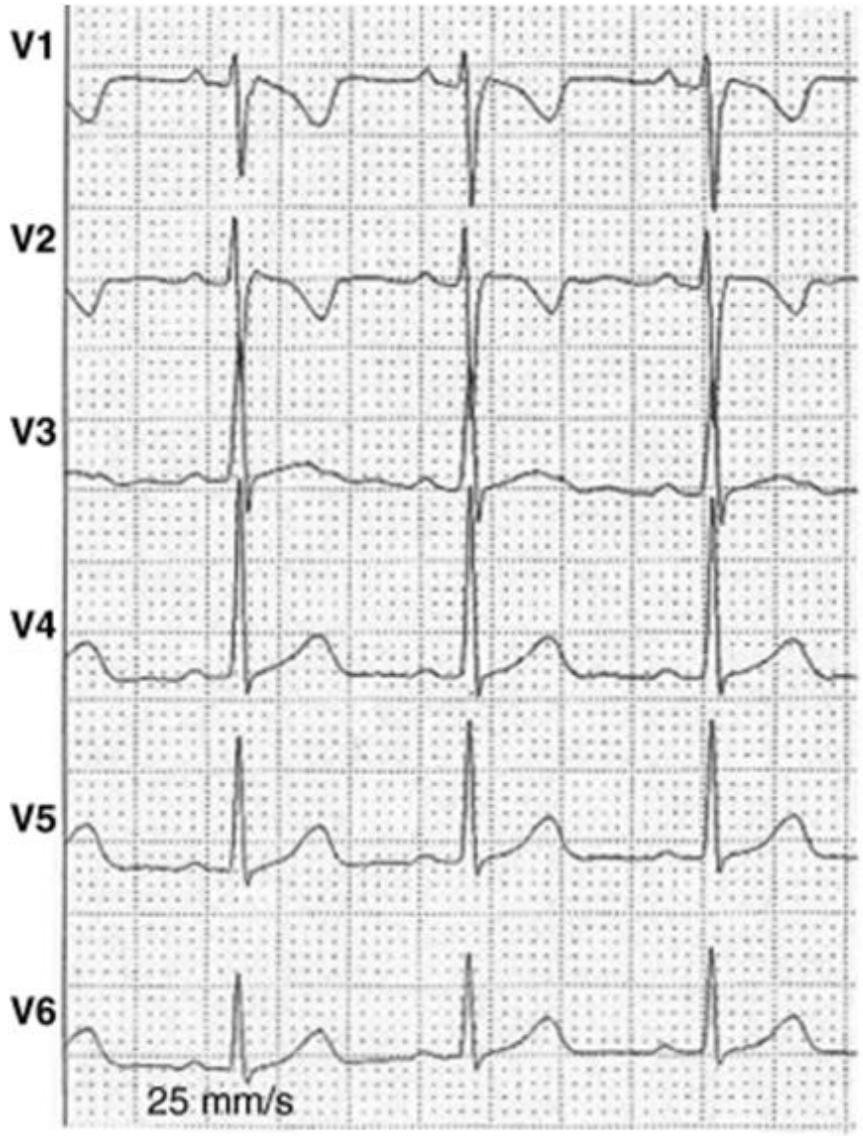
Tipo 1



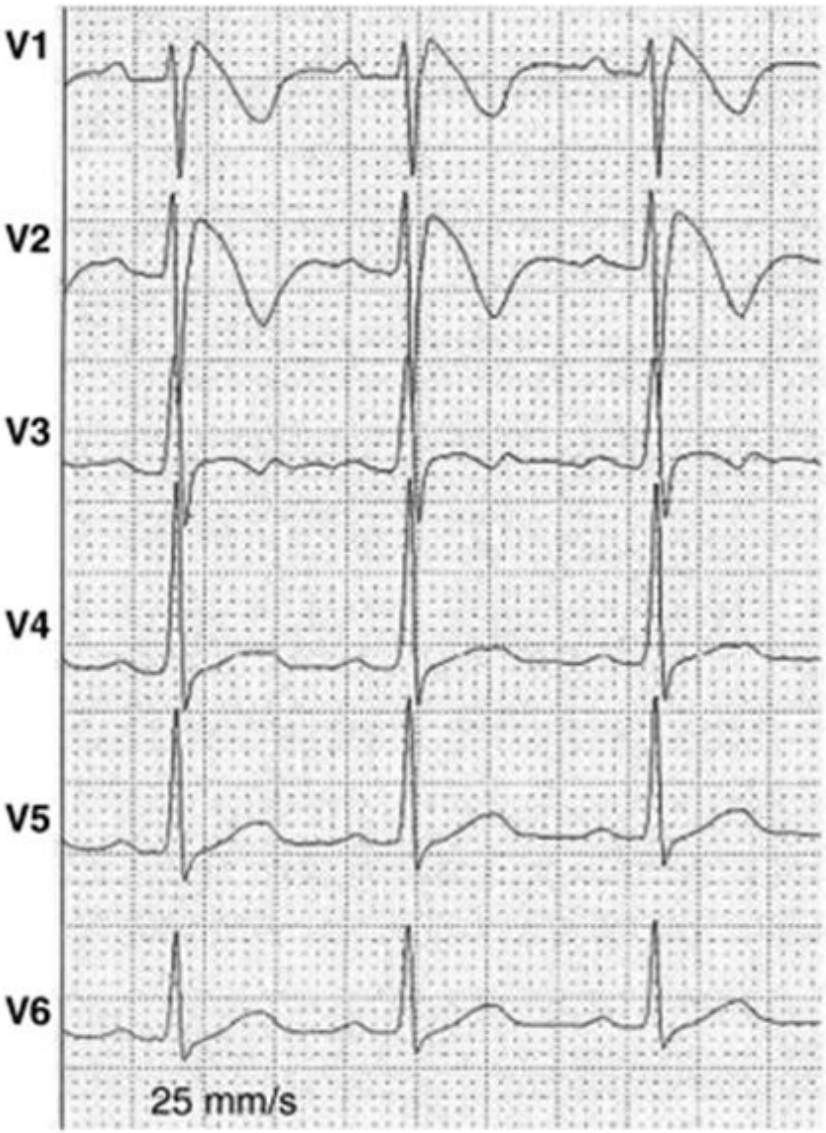
Tipo 2



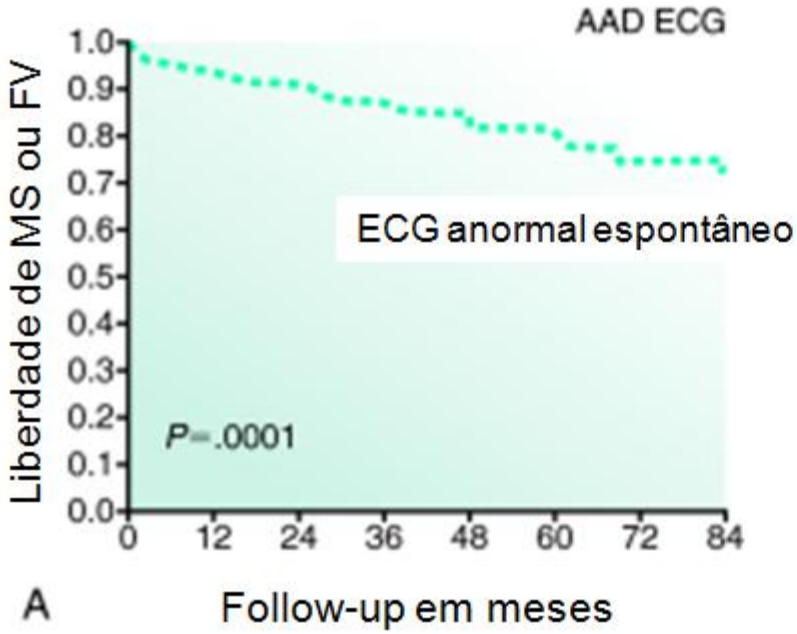
Tipo 3

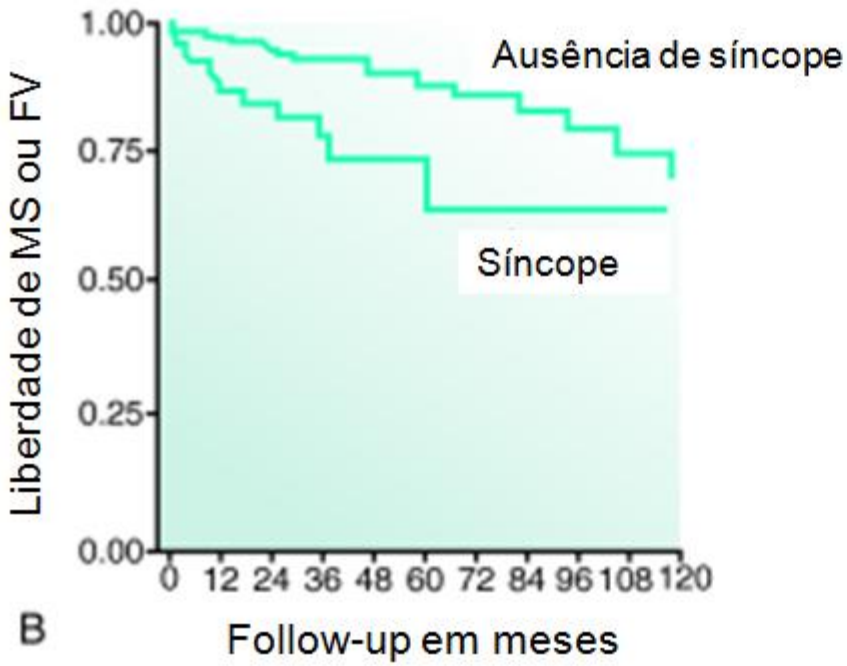


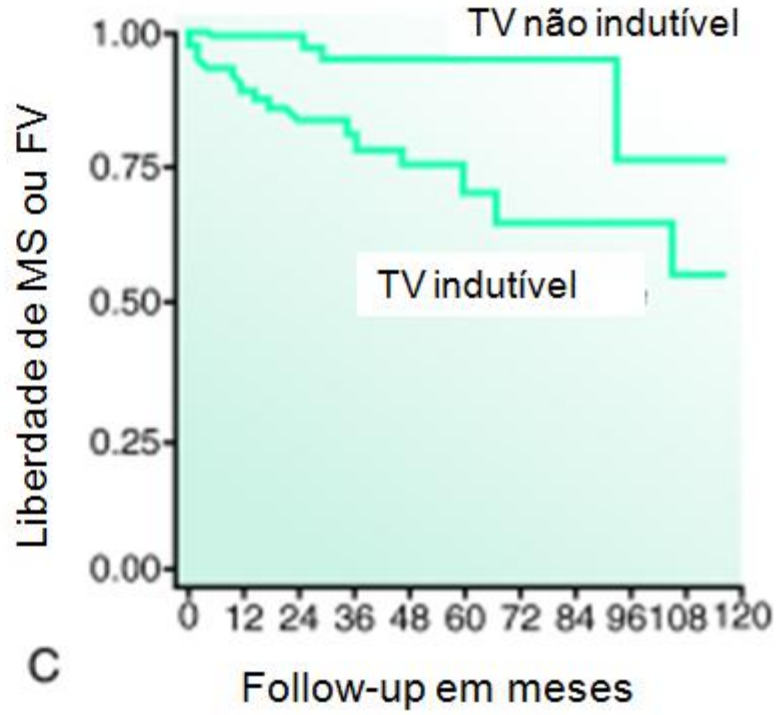
ECG basal



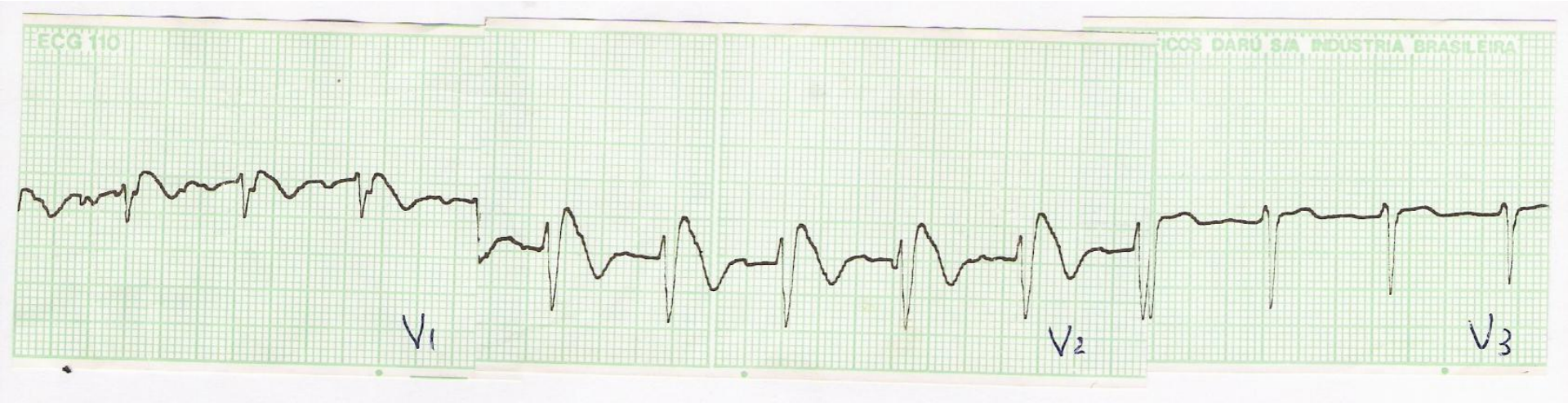
ECG após prova farmacológica

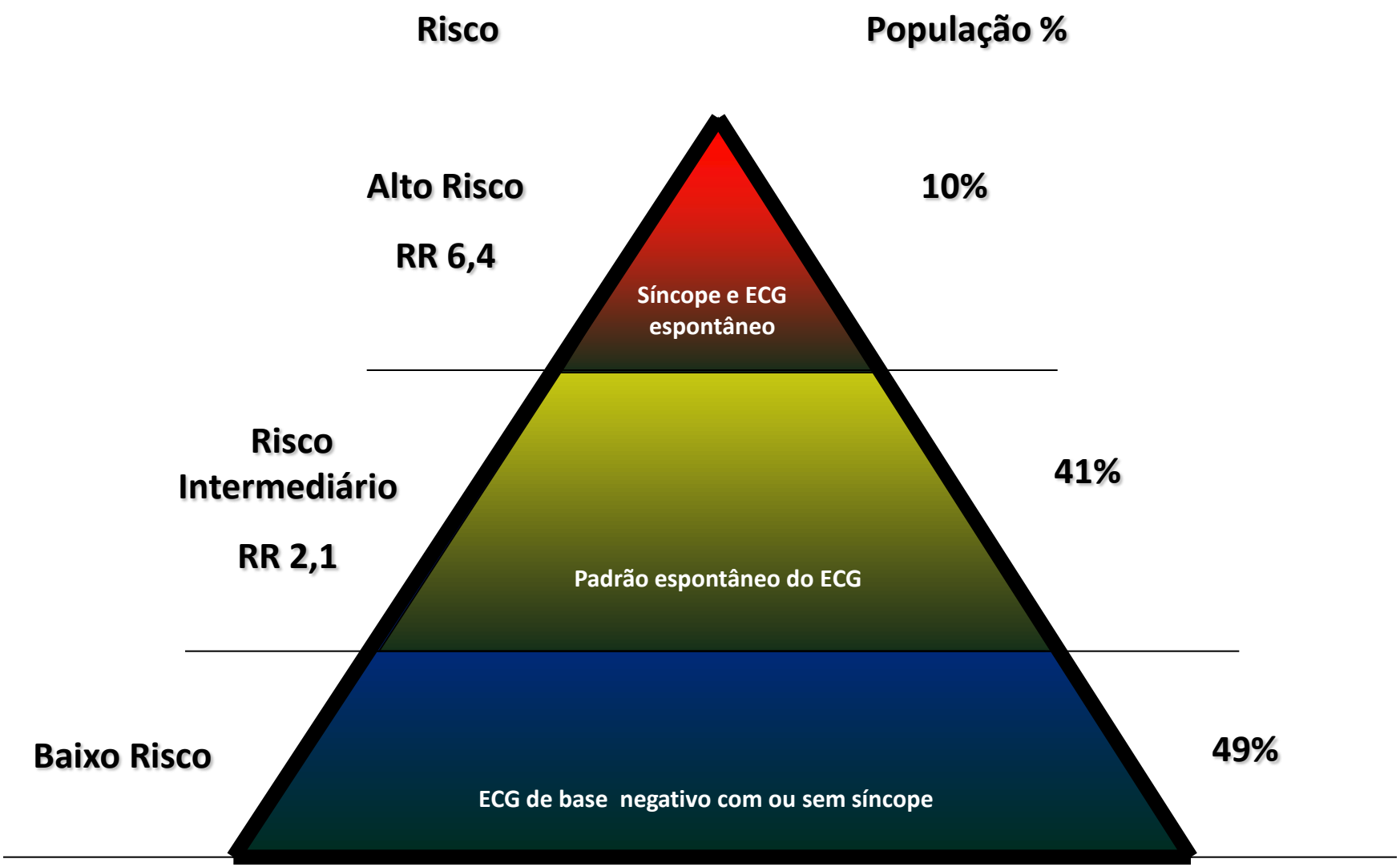

AAD ECG
Liberdade de MS ou FV
1.0
0.9
0.8
0.7
0.6
0.5
0.4
0.3
0.2
0.1
0.0
ECG anormal espontâneo
P=.0001
0 12 24 36 48 60 72 84
A
Follow-up em meses


Liberdade de MS ou FV
1.00
0.75
0.50
0.25
0.00
Ausência de síncope
Síncope
0 12 24 36 48 60 72 84 96 108 120
B
Follow-up em meses


TV não indutível
Liberdade de MS ou FV
1.00
0.75
0.50
0.25
0.00
TV indutível
0 12 24 36 48 60 72 84 96 108 120
C
Follow-up em meses

ECG Brugada –V1- V3

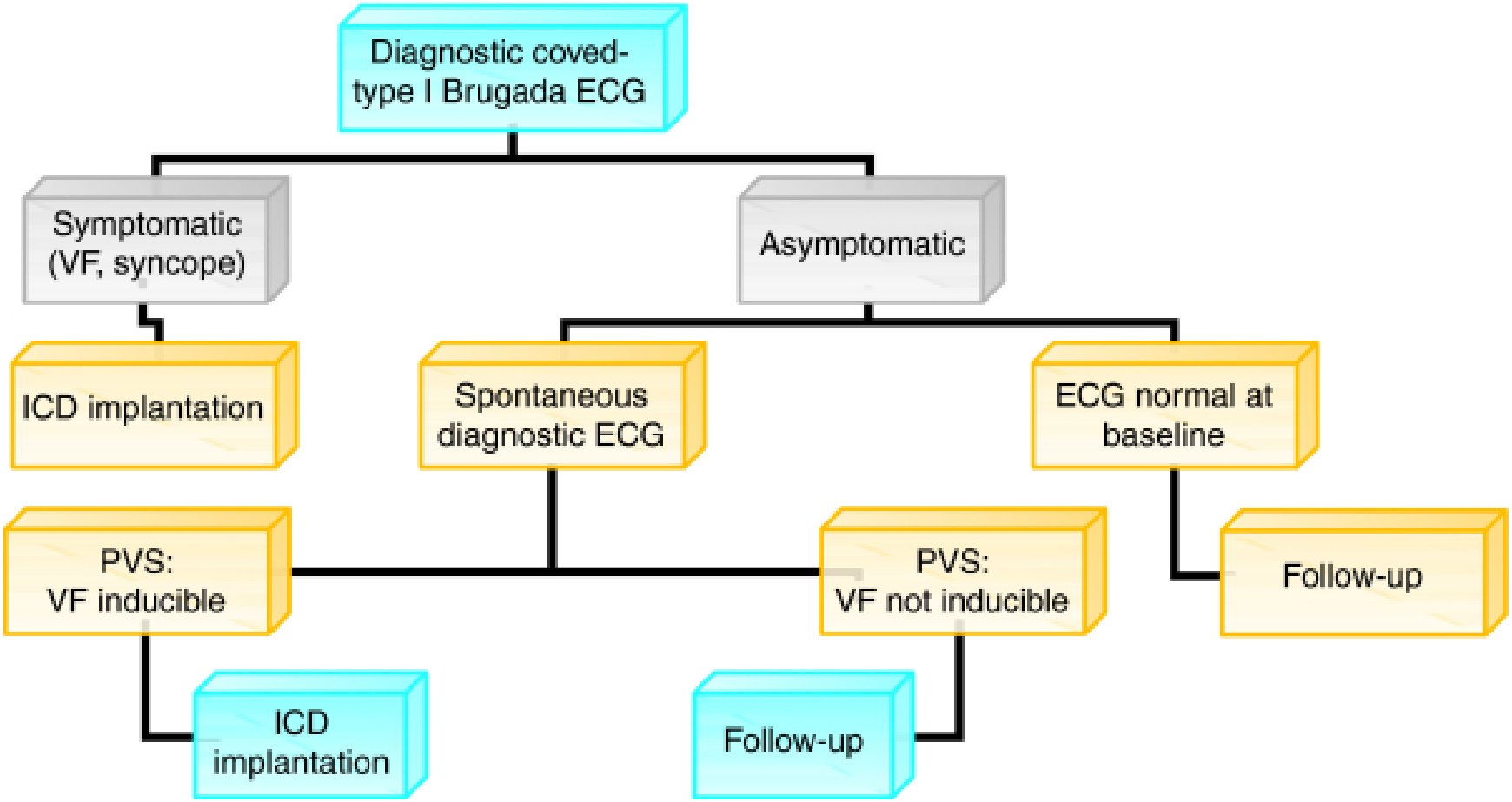



Risco
População %
Alto Risco
RR 6,4
10%
Síncope e ECG espontâneo
Risco Intermediário
RR 2,1
41%
Padrão espontâneo do ECG
Baixo Risco
49%
ECG de base negativo com ou sem síncope

Estratificação de risco Síndrome de Brugada

	I	IIa	IIb
Estratificação de Risco	TV/FV	•História fam MS •Síncope	•TV ou FV no estudo eletrofisiológico
Prevenção Primária	CDI (em pacientes com síncope e TV)	-	•CDI (em paciente assintomático indutivel no EEF)
Prevenção Secundária	CDI	-	-

Europace 2002; 4:3-18


Diagnostic coved-type I Brugada ECG
Symptomatic (VF, syncope)
Asymptomatic
ICD implantation
Spontaneous diagnostic ECG
ECG normal at baseline
PVS: VF inducible
PVS: VF not inducible
Follow-up
ICD implantation
Follow-up

Síndrome do QT longo

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

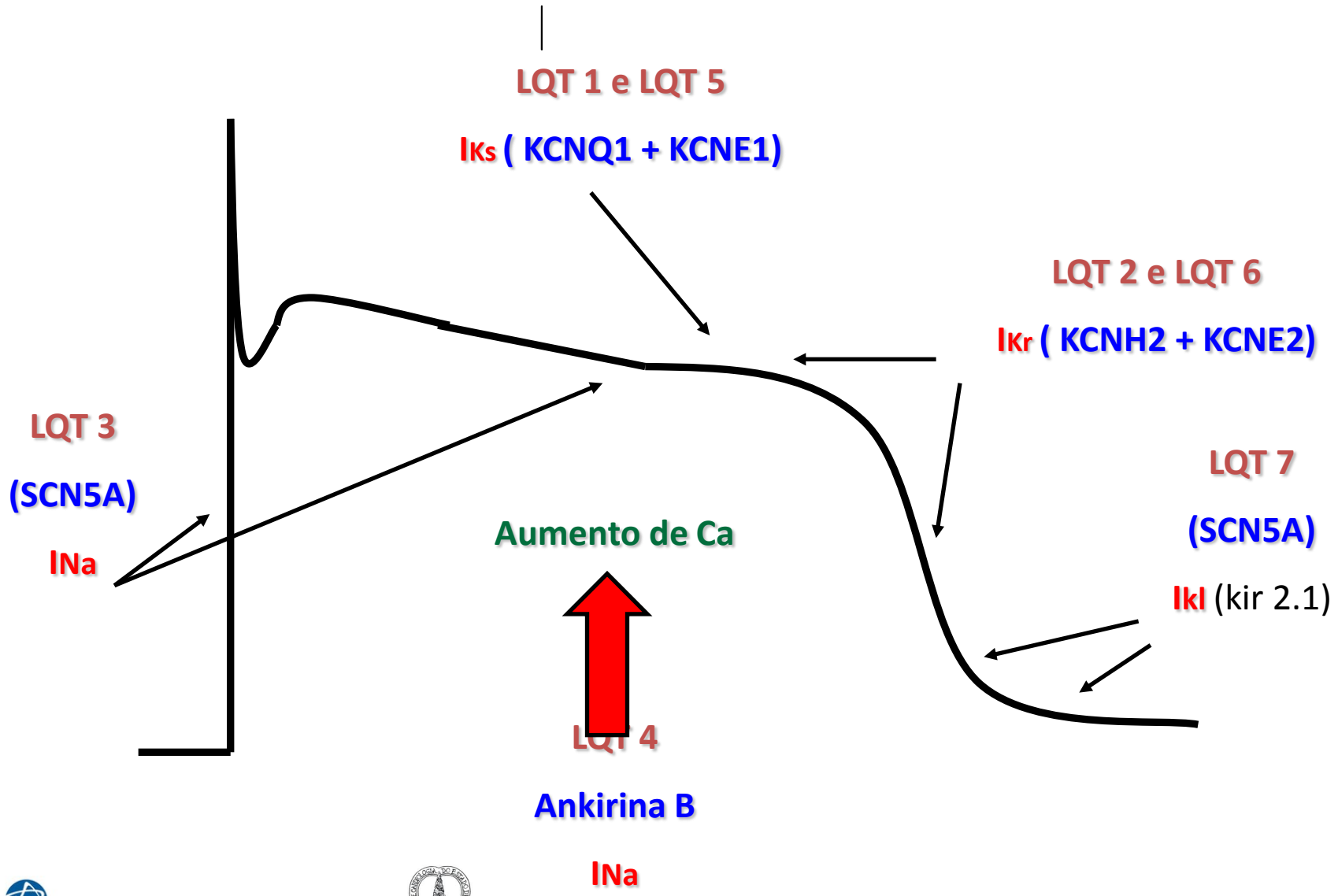
- Sentimento de palpitação ou “vibrar” são incomuns porque a TDP é muito rápida
- Sincope

Critérios diagnósticos Síndrome do QT longo (Schwartz)

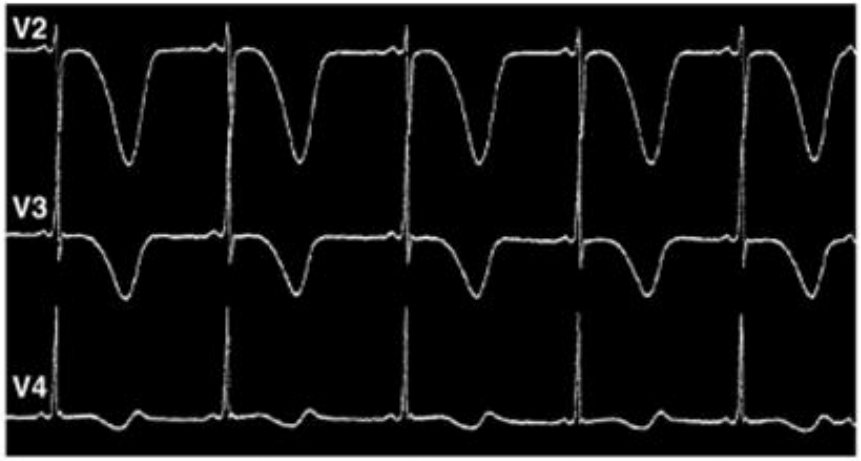
	Pontuação
QTc	
>480 ms	3
460 – 470 ms	2
450 ms (masculino)	1
Torsade pointes	2
Alternância de ondas T (TWA)	1
Baixa frequência cardíaca	0,5
Síncope	
Com estresse	2
Sem estresse	1
Surdez congênita	0,5
Membros da familia com LQTS	1
Morte cardíaca inexplicada na família (imediatos) antes dos 30 anos	0,5

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

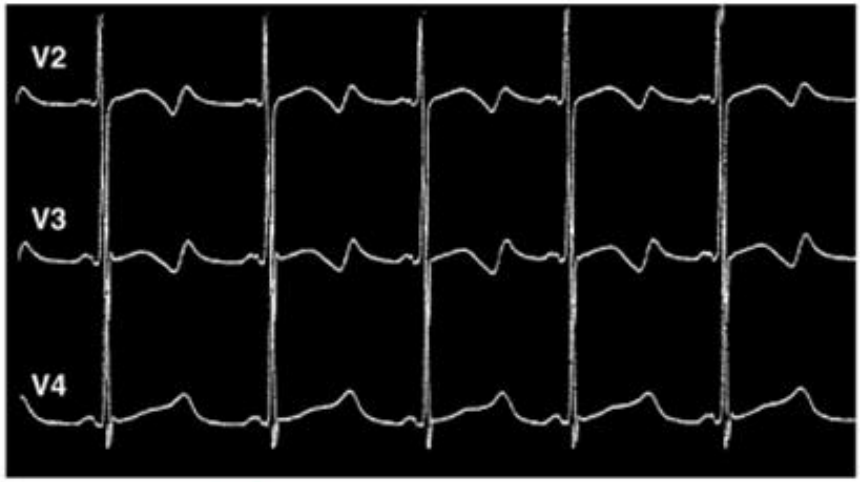
J Intern Med 2006; 259:39-47


LQT 1 e LQT 5
IKs (KCNQ1 + KCNE1)
LQT 2 e LQT 6
IKr (KCNH2 + KCNE2)
LQT 3
(SCN5A)
INa
LQT 7
(SCN5A)
IkI (kir 2.1)
Aumento de Ca
LQT 4
Ankirina B
INa

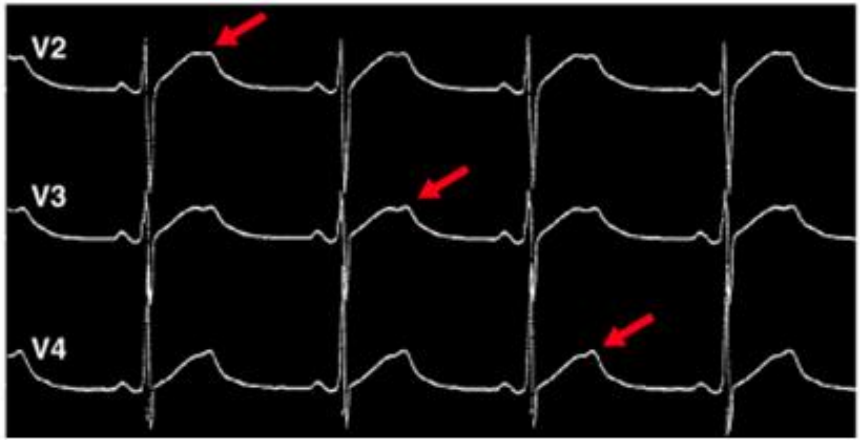
Caso índice
7 anos



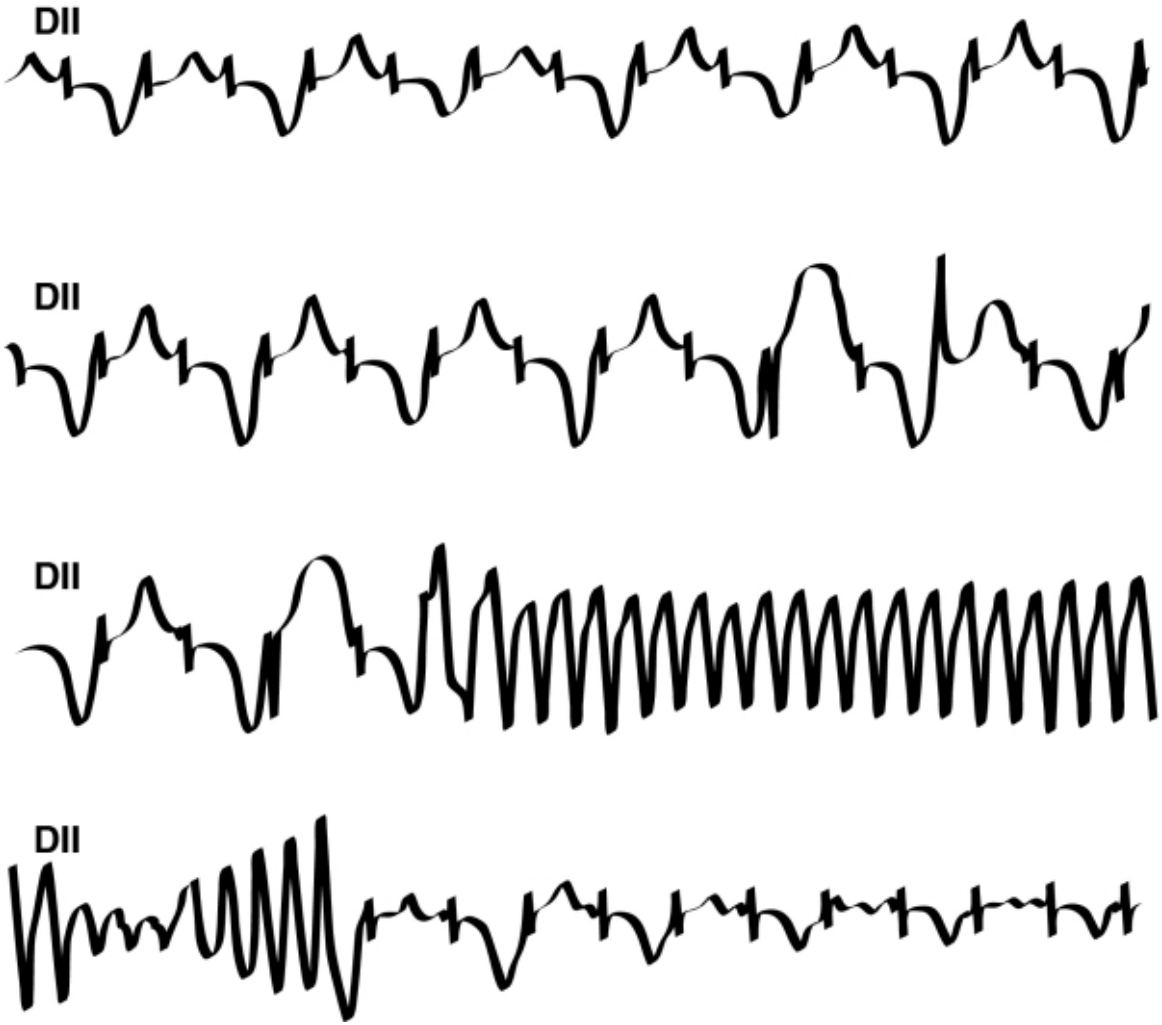
Irmã
10 anos



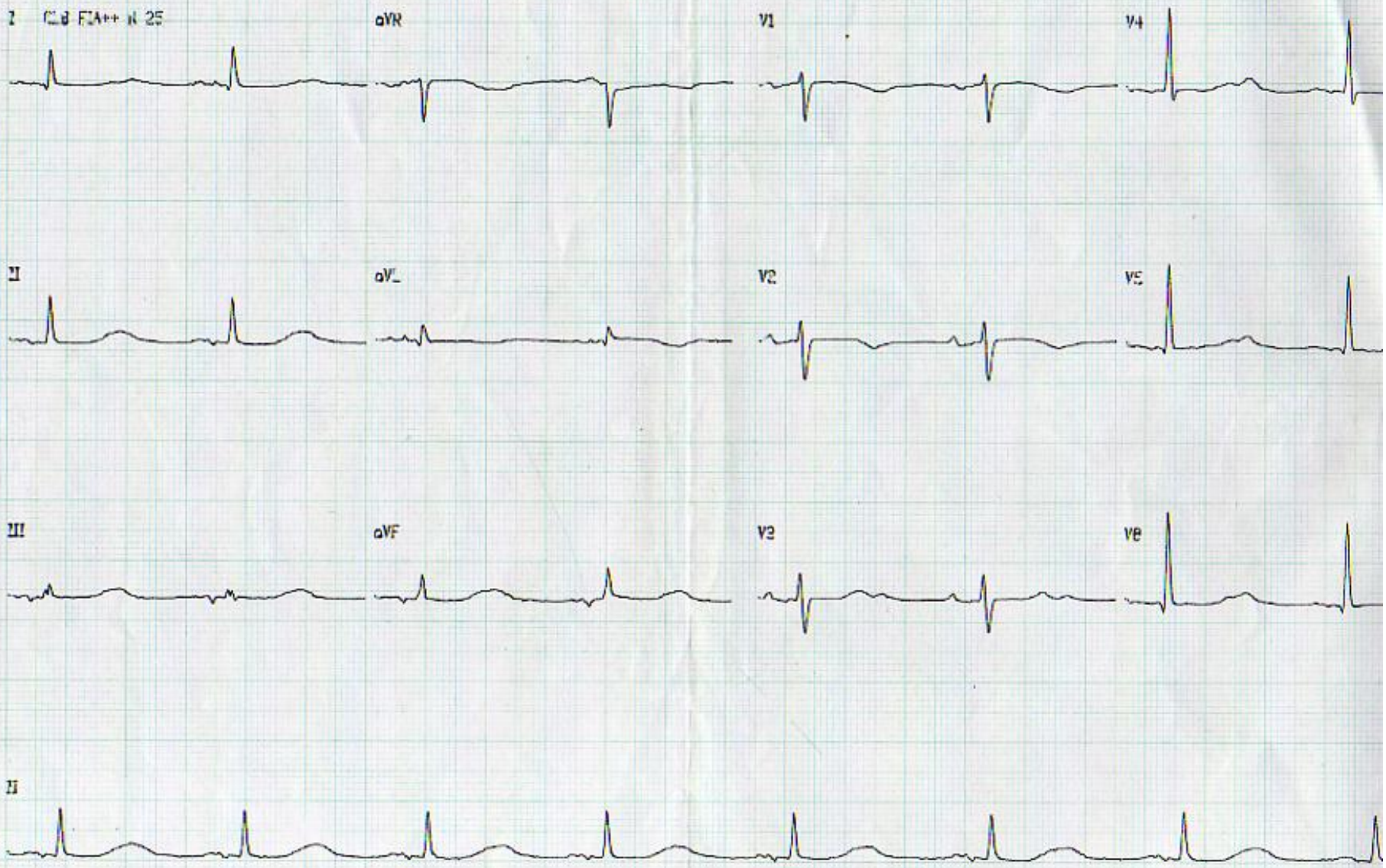
Pai
37 anos



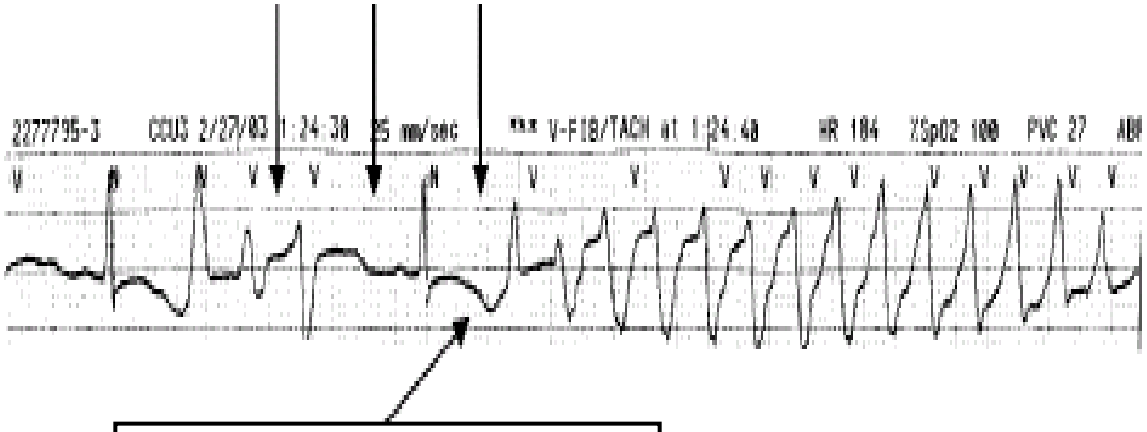
Alternancia de onda T no QT longo



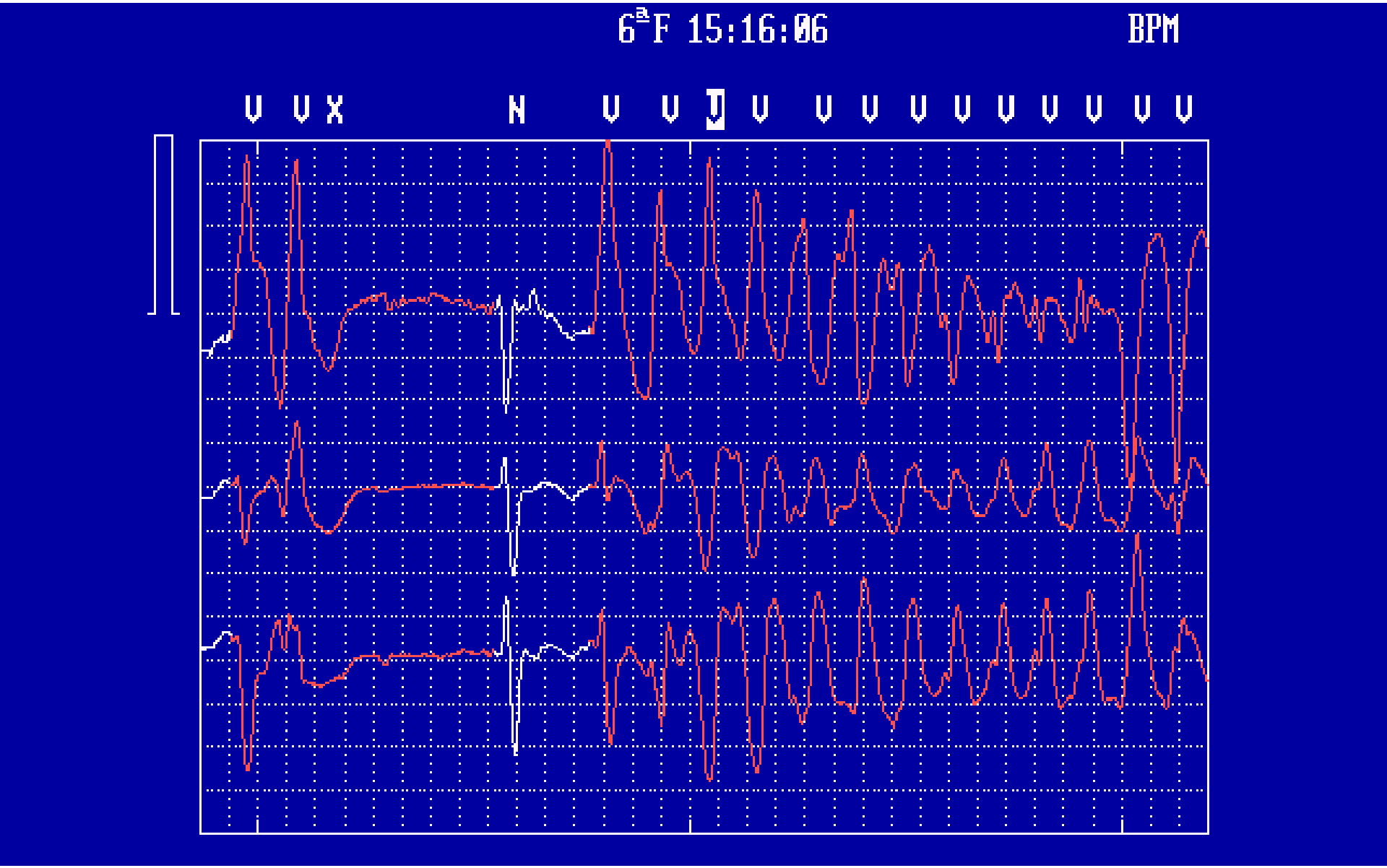
Europ Paediatr Cardiol 8:28-36, 1972.

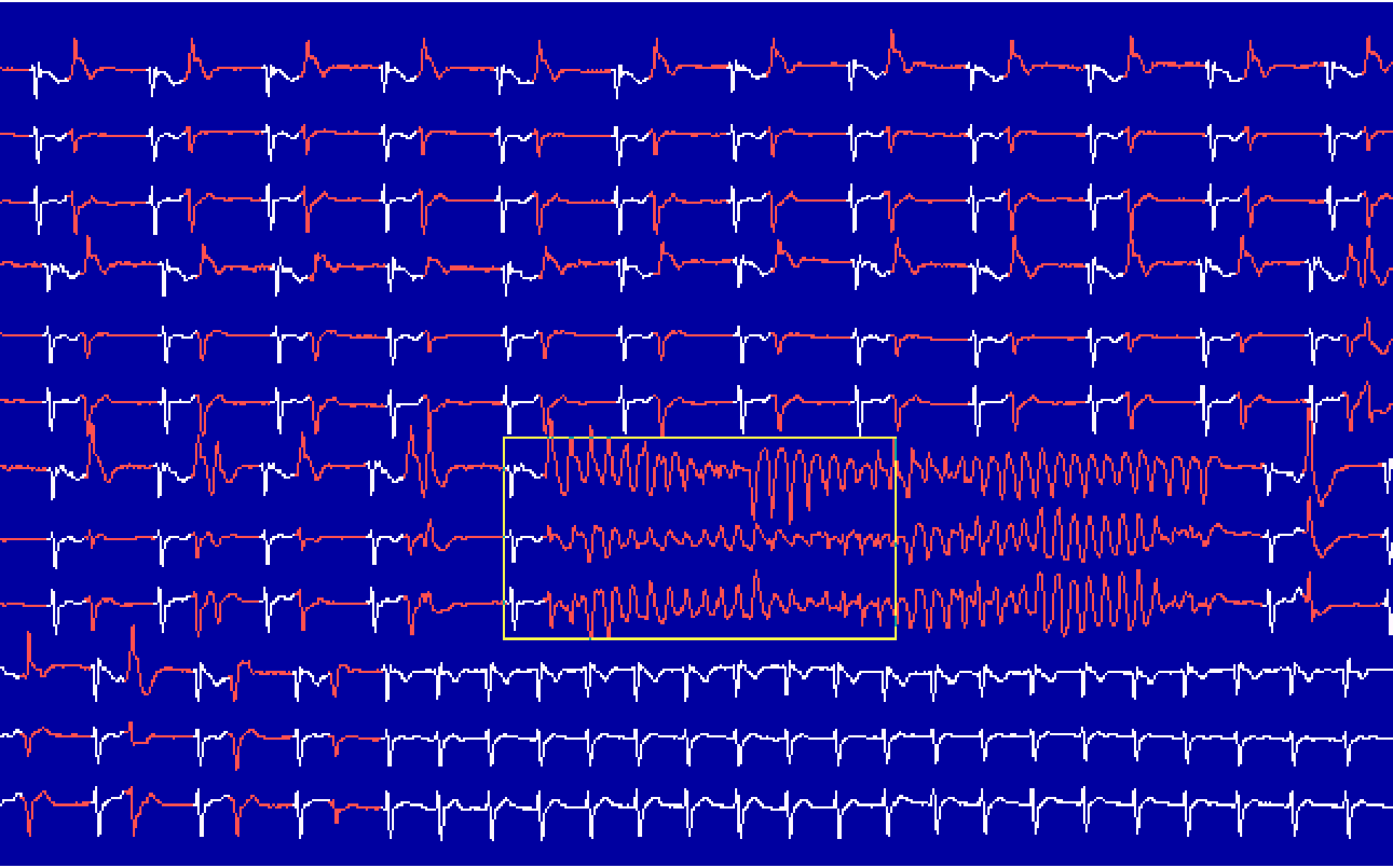

aVR
V1
V4
aVL
V2
V5
aVF
V3
V6
II

Curto Longo Curto

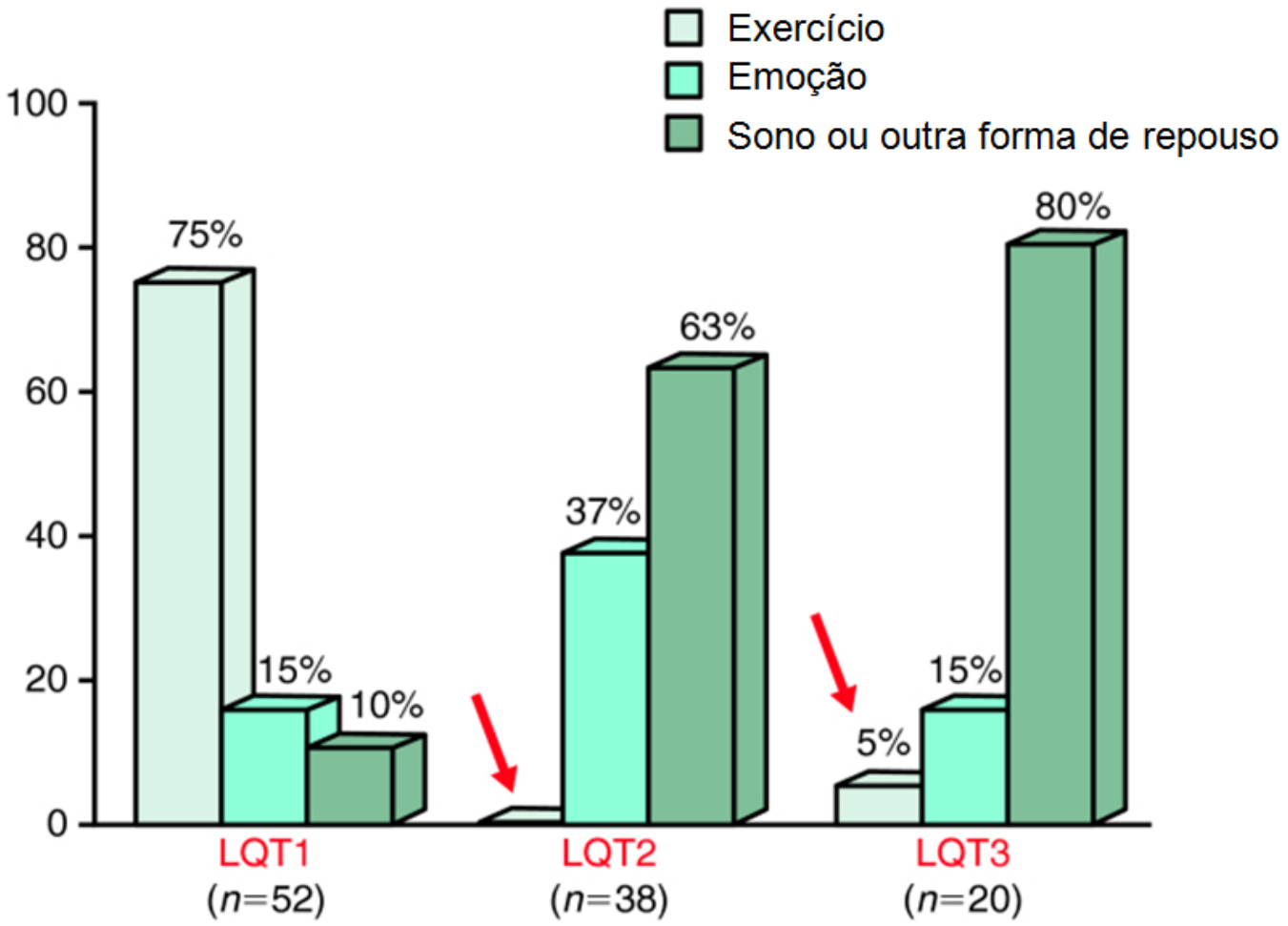


Intervalo QT longo e onda T bizarra

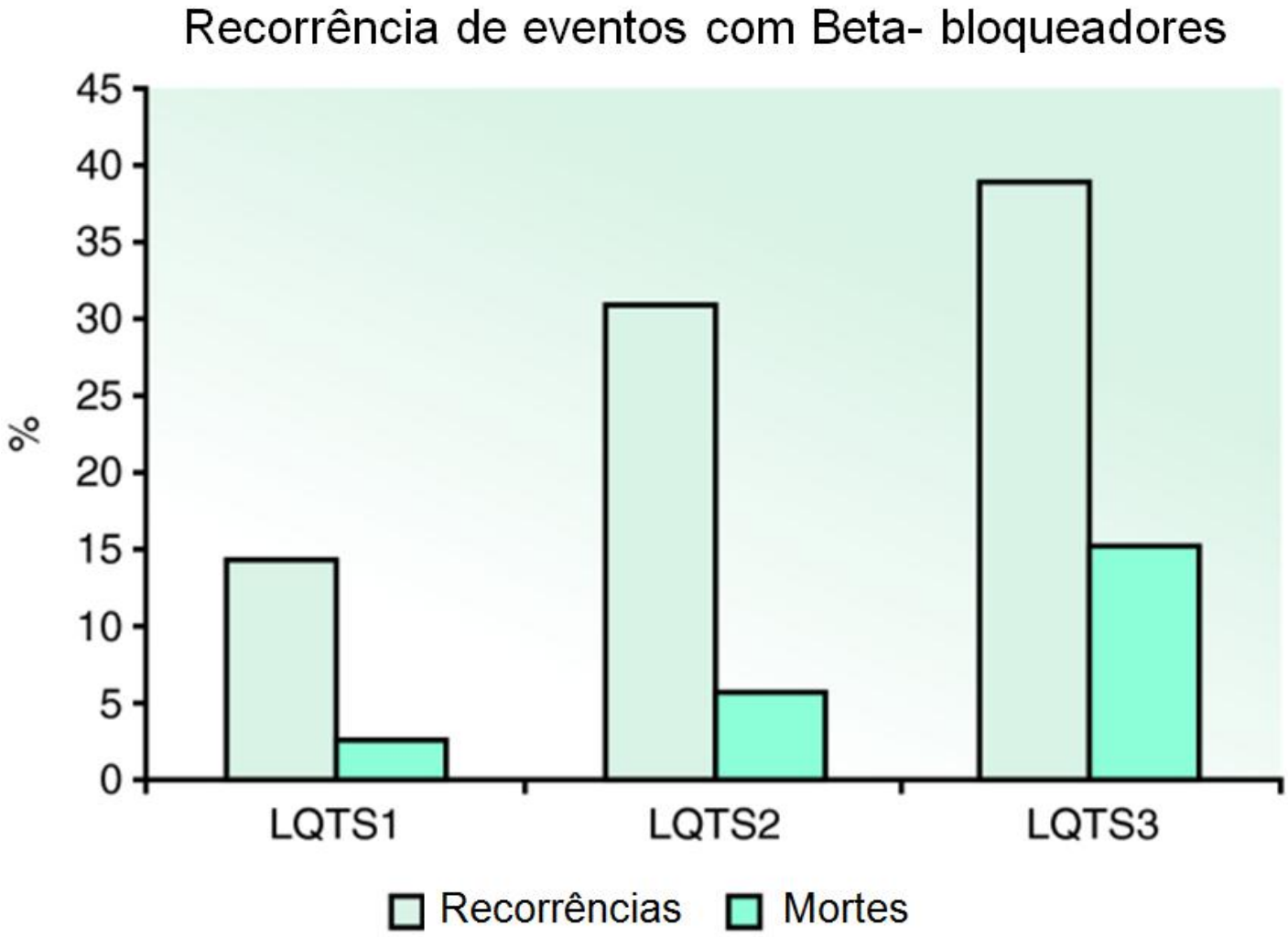

6ªF 15:16:06
BPM
V V X N V V J V V V V V V V V V V



Gatilho para eventos súbitos na LQTS

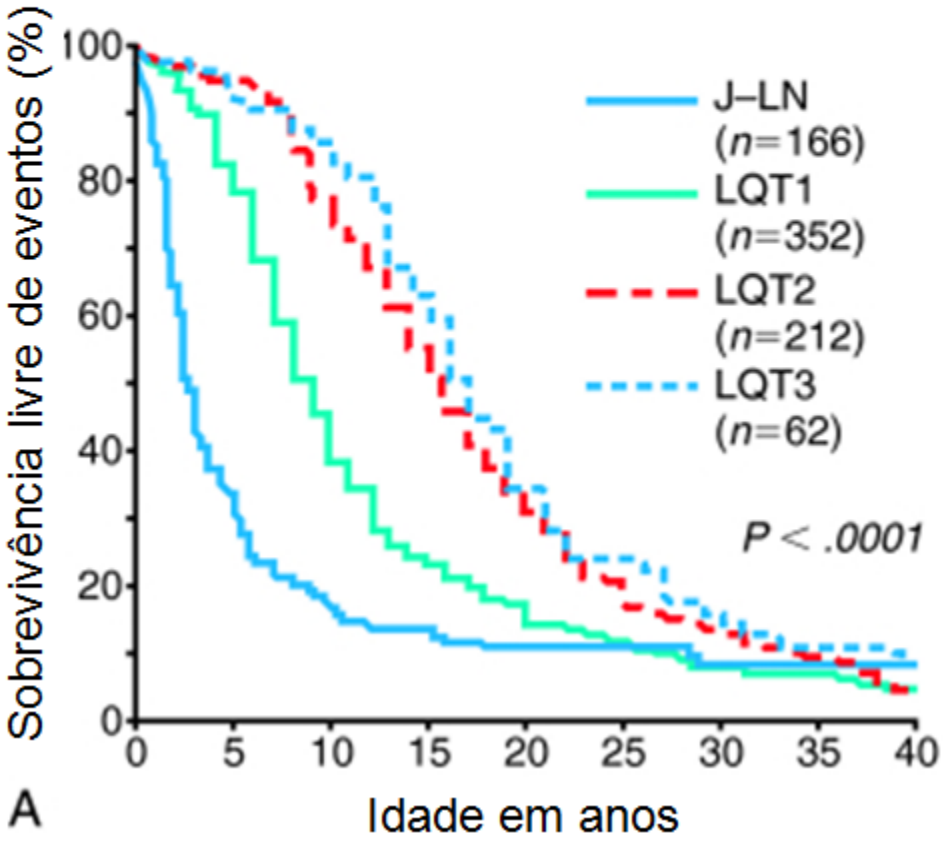


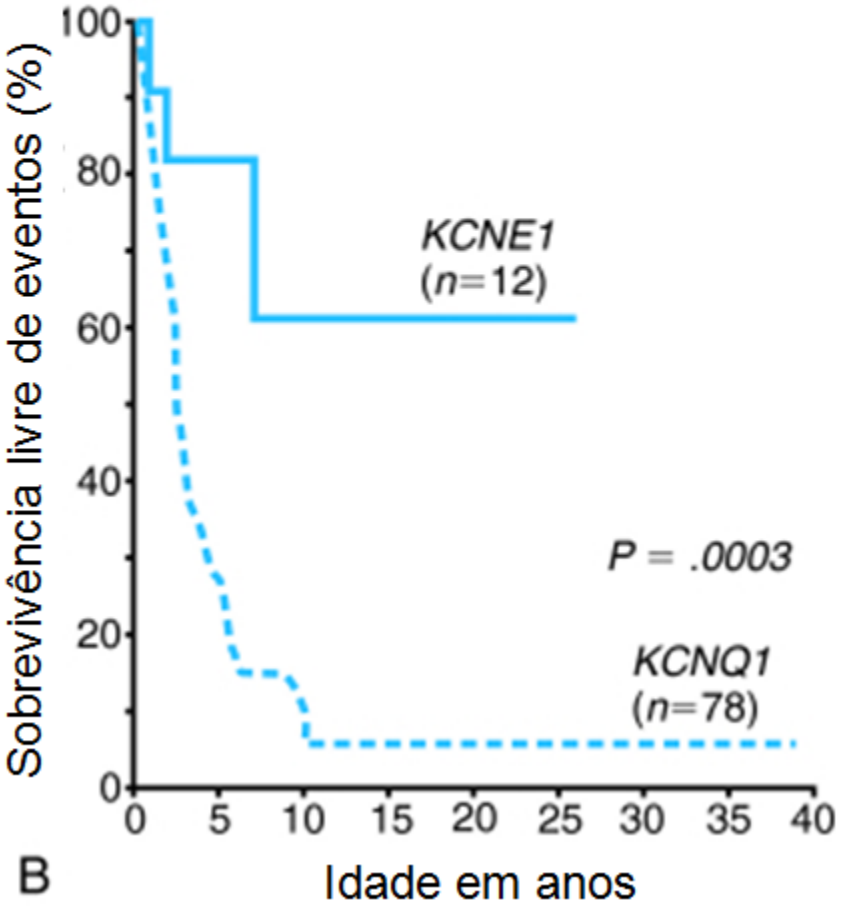
Circulation 103:89-95, 2001



JAMA 2004; 292:1341-1344

J Med Genet 2003; 40:141-145.





Circulation 113:783-790, 2006.

Estratificação de risco Síndrome do QT longo

	I	IIa	IIb
Estratificação de Risco	•Torsade pointes/FV •Parada cardíaca •JLN •LQT3	•QTc > 600 ms •Evento cardíaco na infância •Sindactilia + BAVT •Alternância de T (TWA) •Sexo Feminino	•História familiar de MS •Aumento da dispersão do QT
Prevenção Primária	• Suspender medicamentos que prolongam o QT •Suspender atividade física •Betabloqueadores	CDI	•Denervação cardíaca
Prevenção Secundária	•CDI •Betabloqueadores •Suspender medicamentos que prolongam o QT •Suspender atividade física	-	-

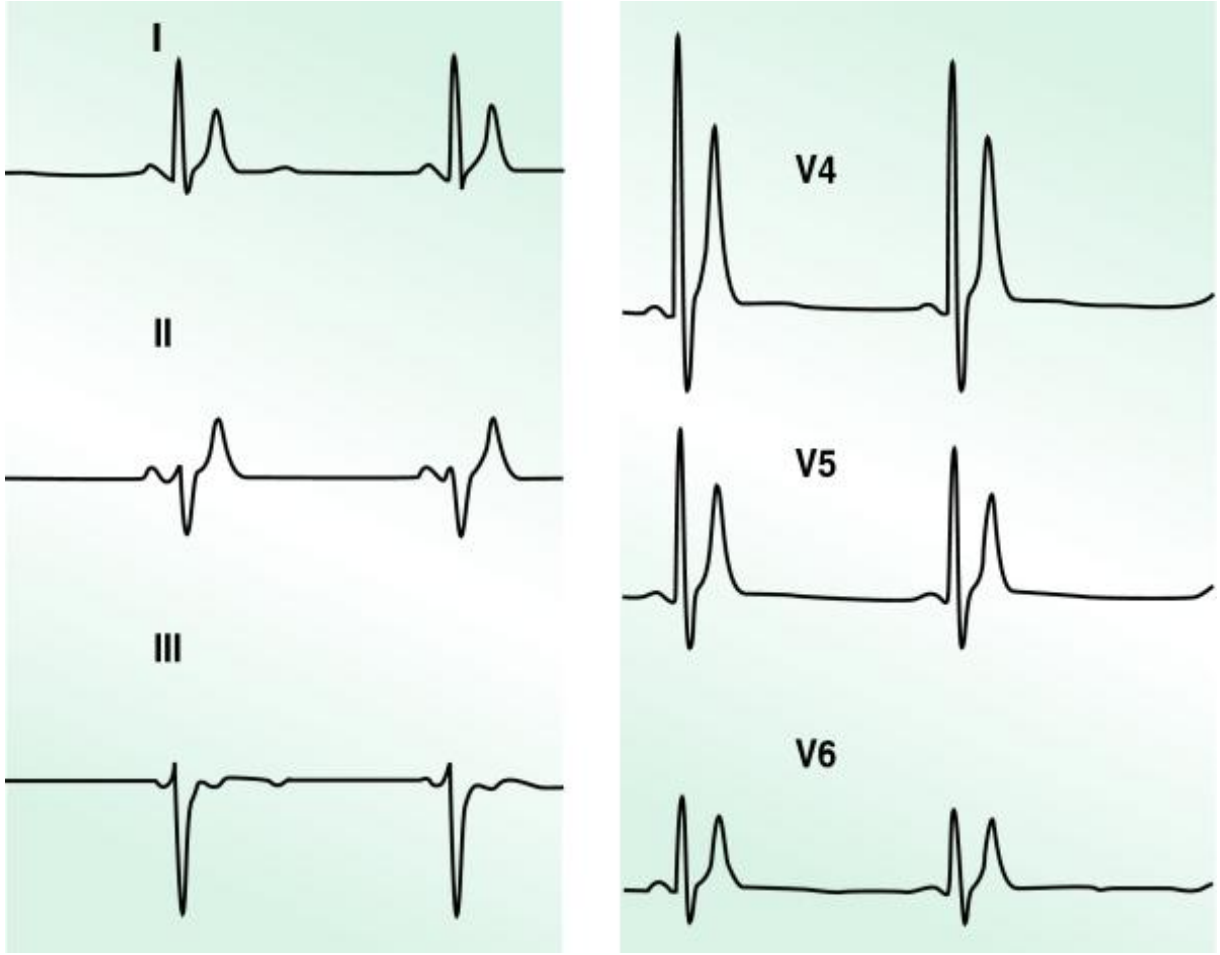
Europace 2002; 4:3-18

Síndrome do QT Curto

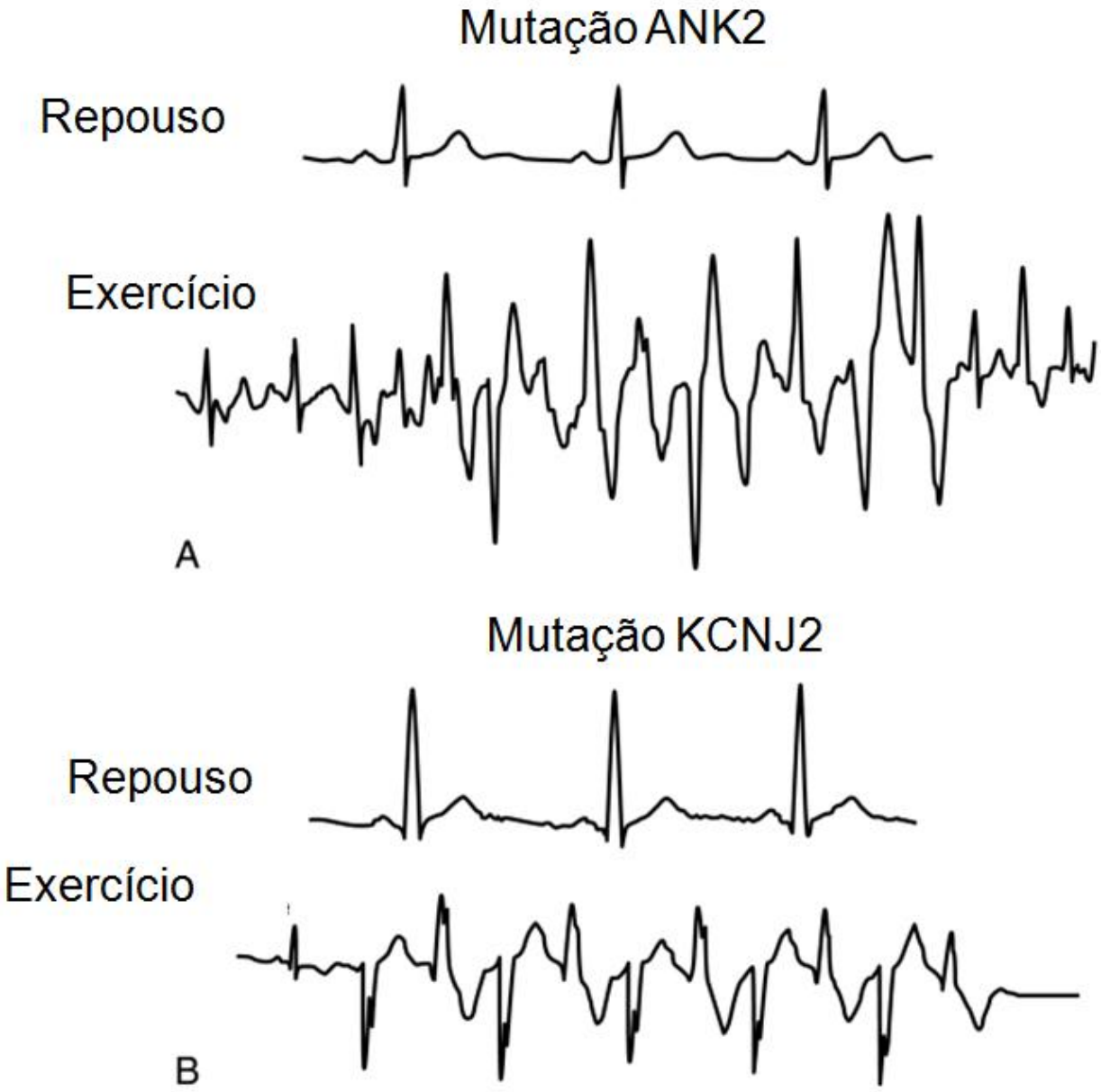
Síndrome do QT curto

- Asoociado a FV, TV e fibrilação atrial
- Associação com morte súbita berço
- Mutação no gene KCNH2
- 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

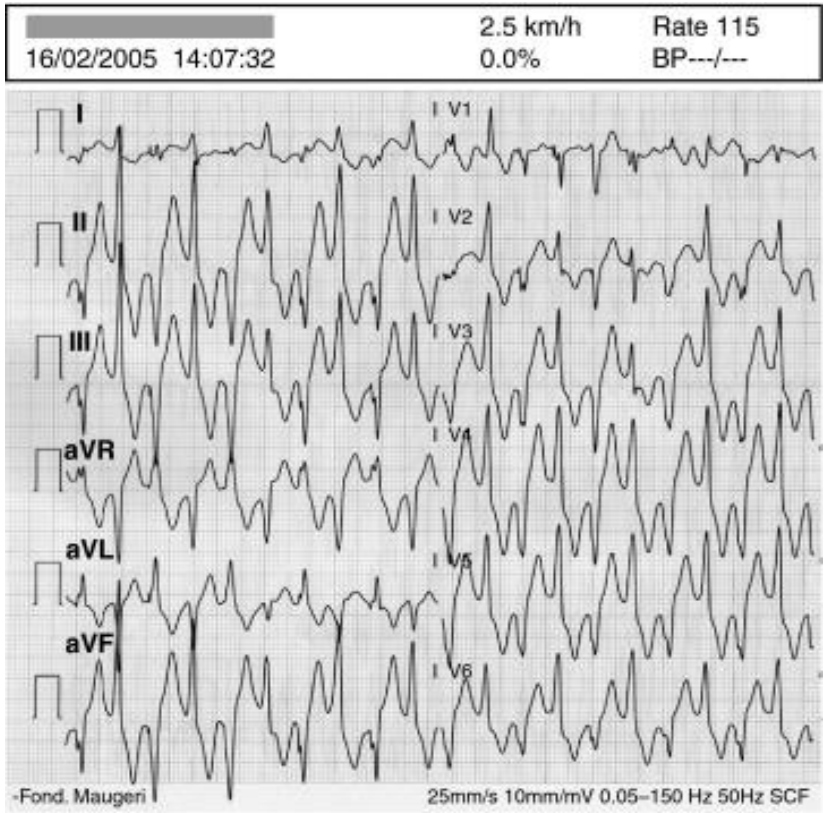
Diagnóstico = QTc < 320 ms

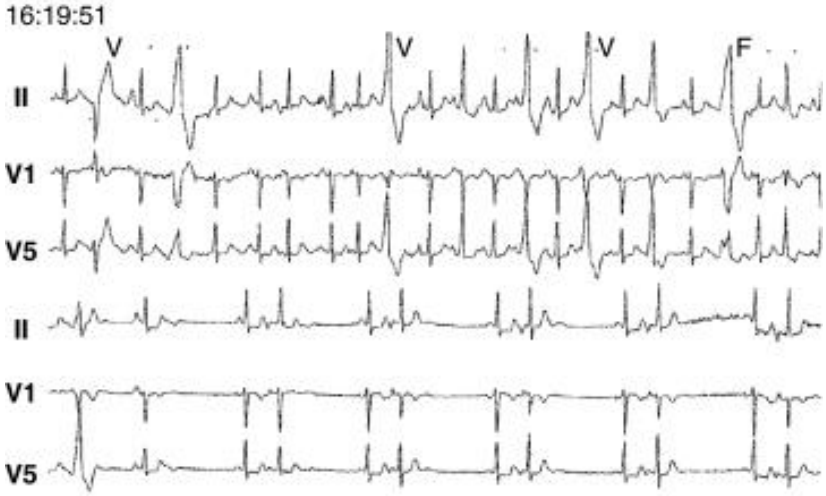


Taquicardia ventricular catecolaminérgica



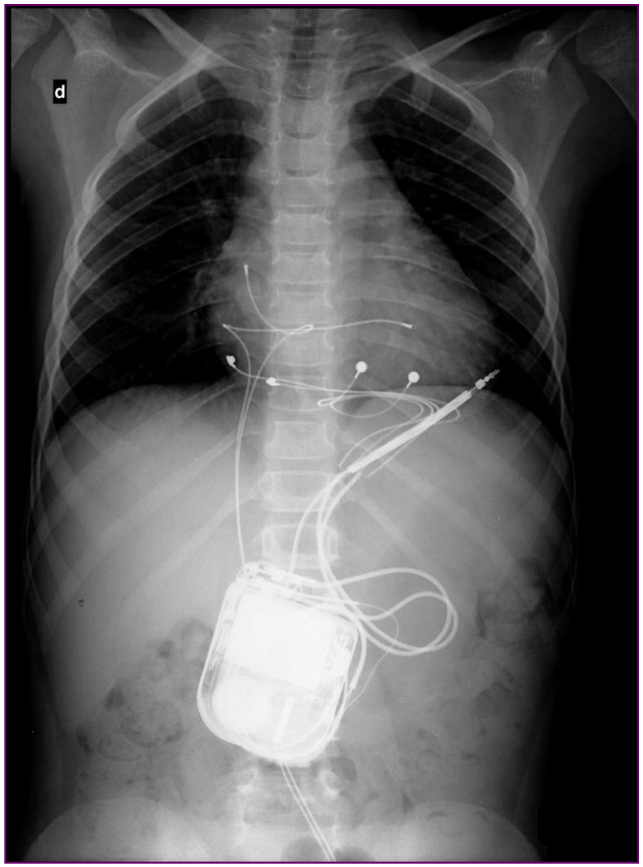
Heart Rhythm 2:1218-1223, 2005.

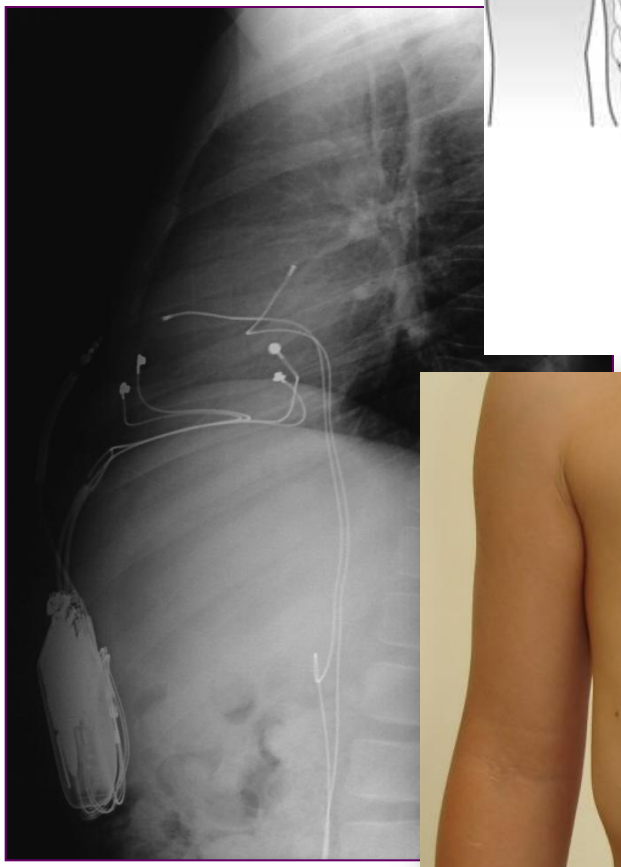

2.5 km/h
Rate 115
16/02/2005 14:07:32
0.0%
BP---/---
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
-Fond. Maugeri
25mm/s 10mm/mV 0.05–150 Hz 50Hz SCF

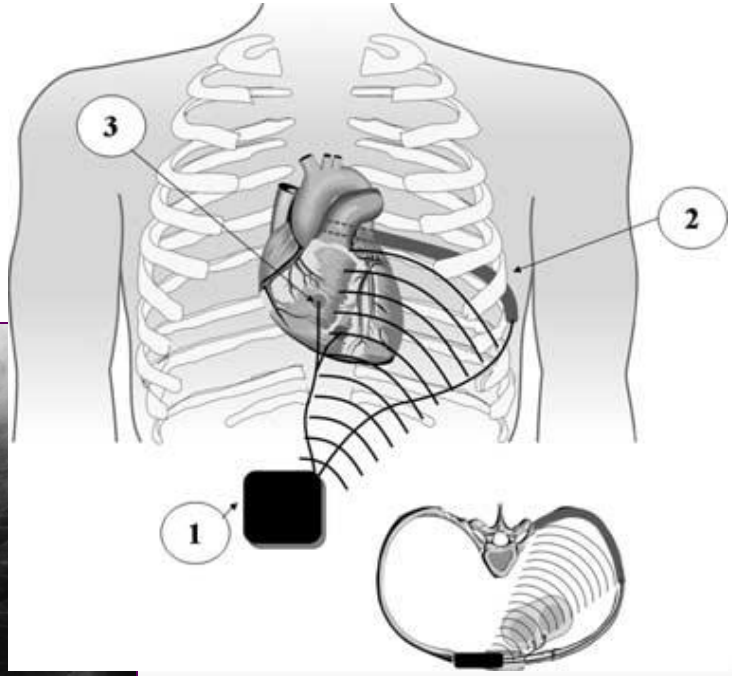

16:19:51
V
V
V
F
II
V1
V5
II
V1
V5

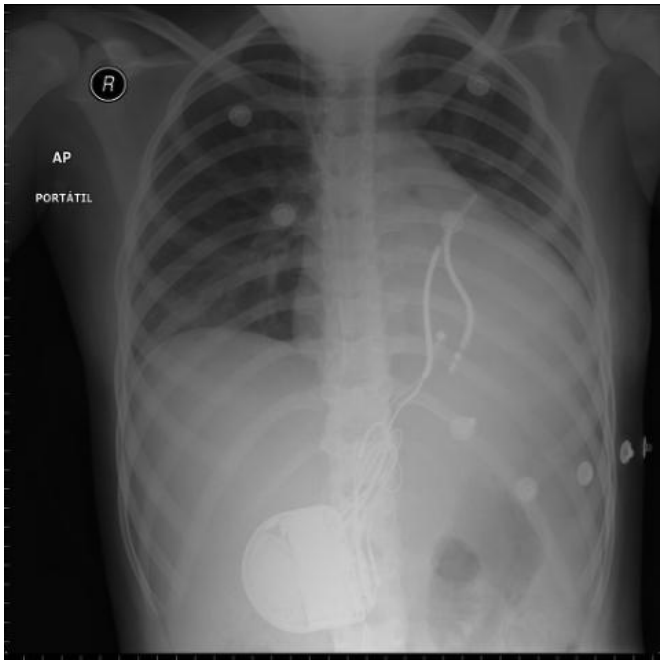
Evolução

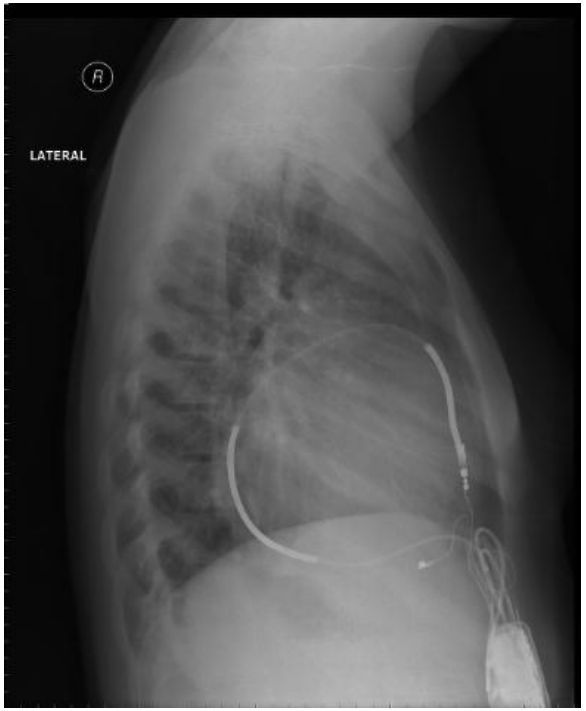
209 cc
113 cc
80 cc
80 cc
72 cc
54 cc
62 cc
49 cc
39.5 cc
39 cc
39.5 cc
39 cc
39.5 cc
36 cc


d




3
2
1

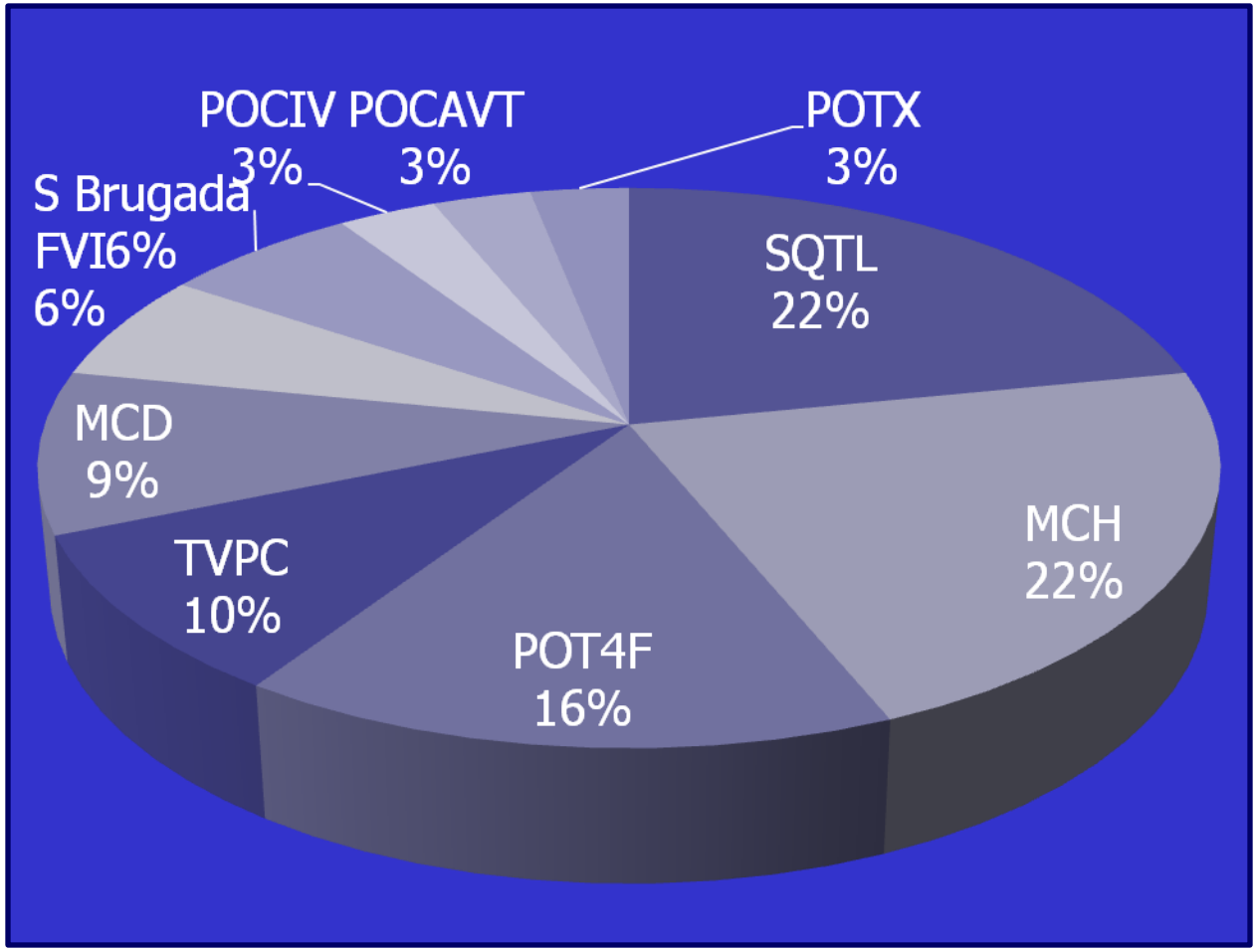

R
AP
PORTÁTIL


R
LATERAL

32 pacientes

Idade
10 meses a 18 anos
m 6,5 anos

Sexo
25 masculino



- Prevenção 2ª
17 pacientes = 53%

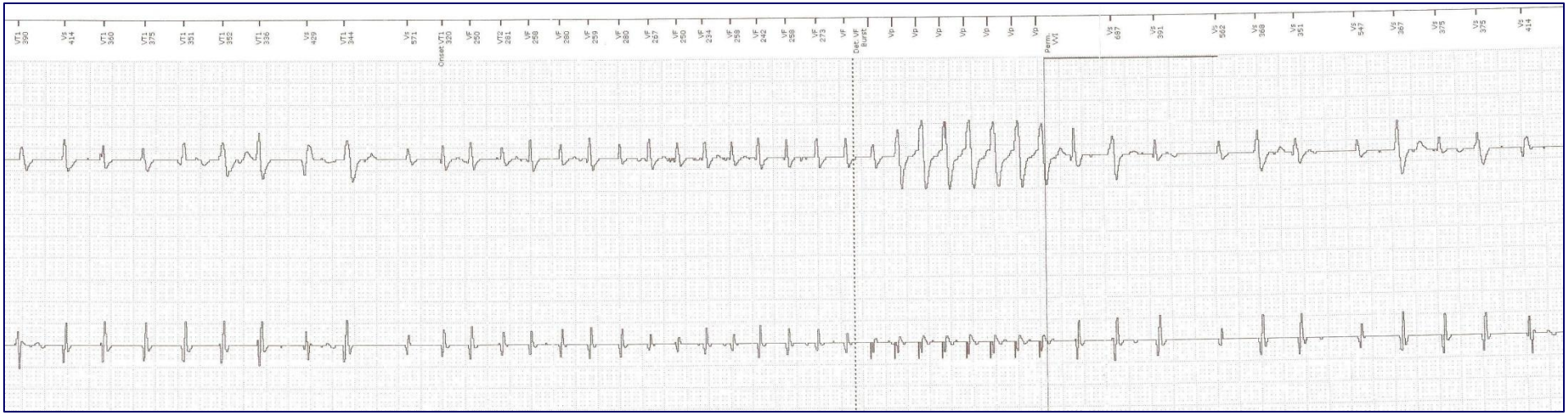
Técnica

↓

- 22 epimiocardicos = 62%

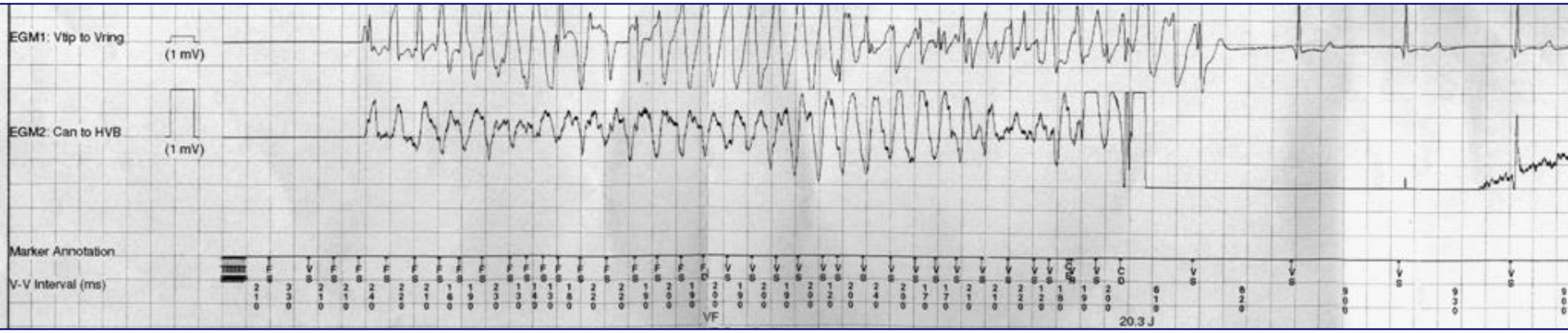
Miocardiopatia dilatada

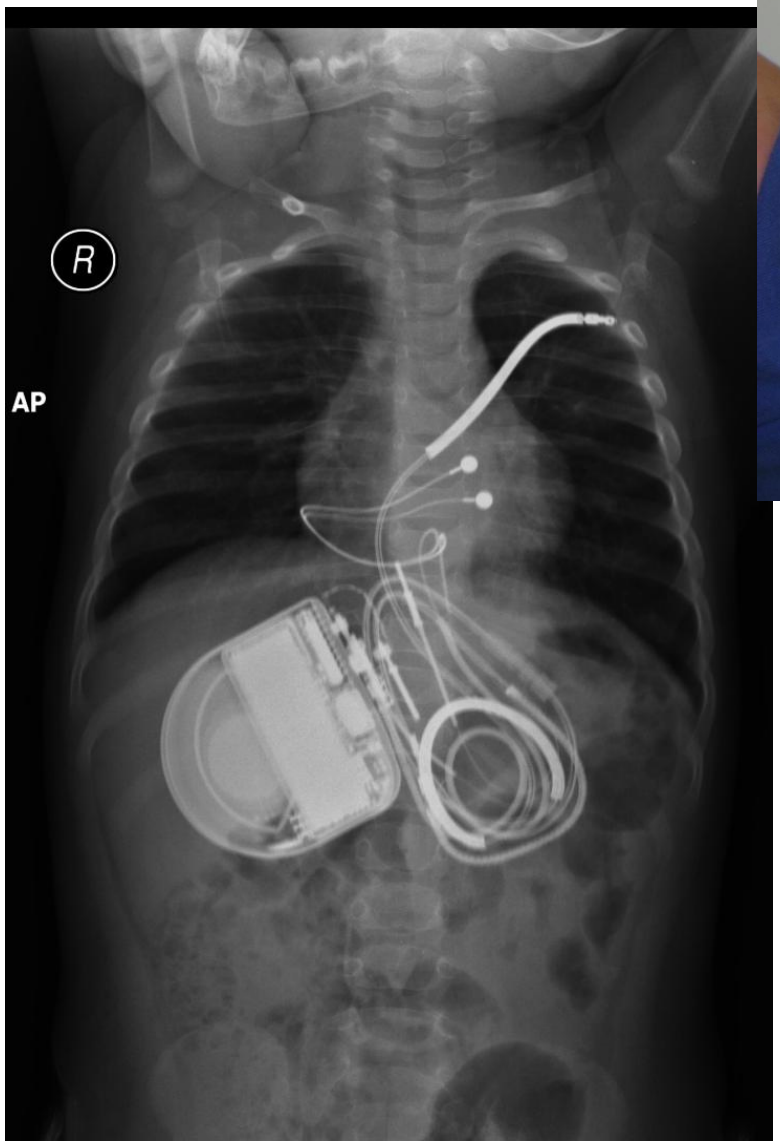
Burst

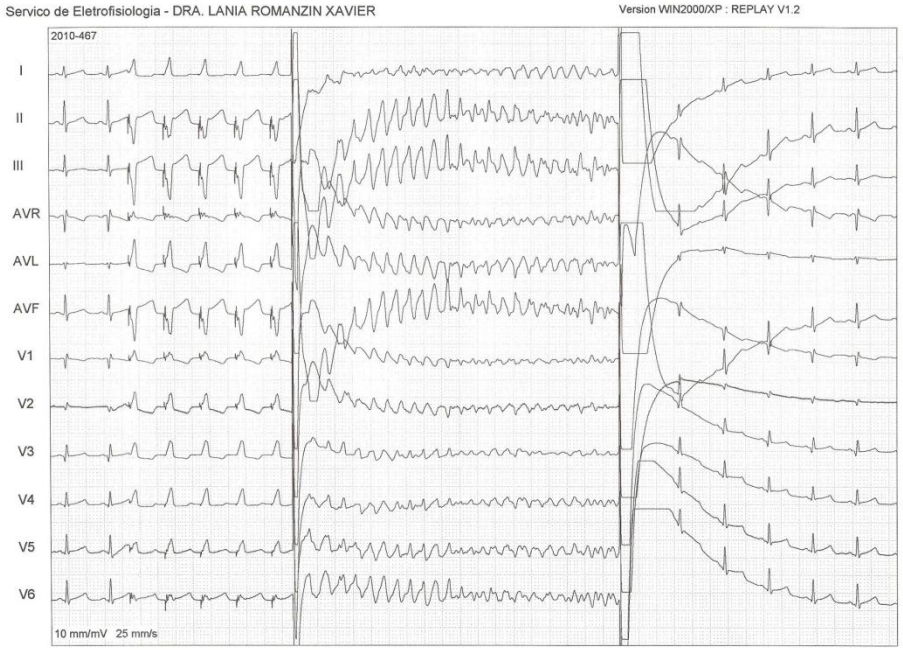


Sindrome do QTL

Choque




R
AP


Servico de Eletrofisiologia - DRA. LANIA ROMANZIN XAVIER
Version WIN2000/XP : REPLAY V1.2
2010-467
I
II
III
AVR
AVL
AVF
V1
V2
V3
V4
V5
V6
10 mm/mV 25 mm/s

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

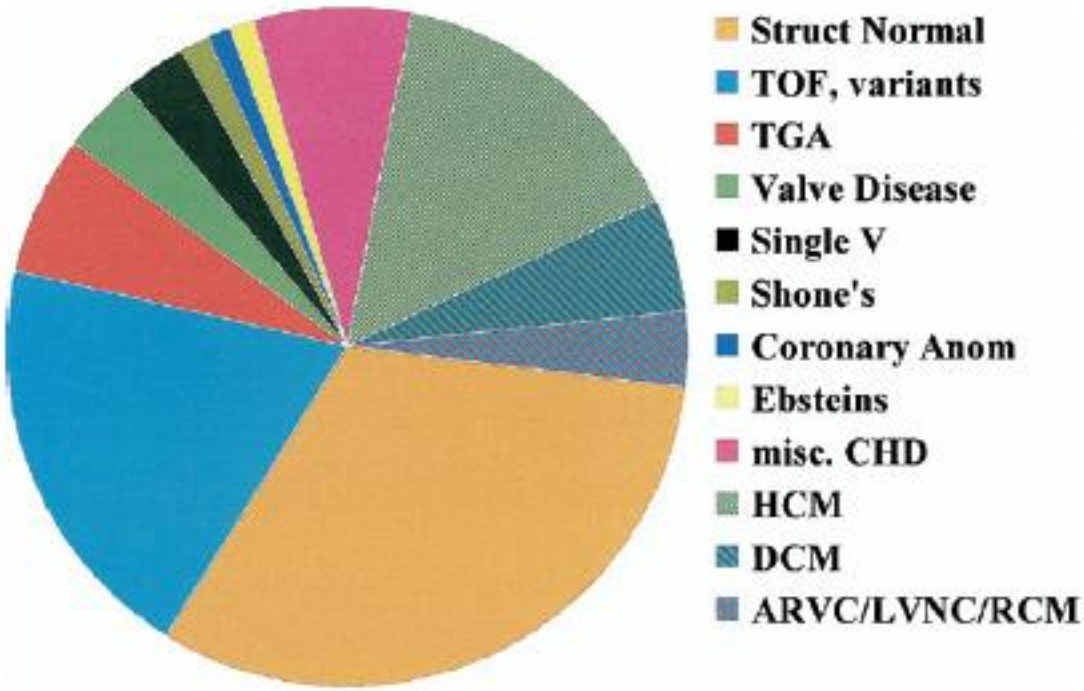


Figure 1 Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients

(J Am Coll Cardiol 2008;51:1685–91)

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

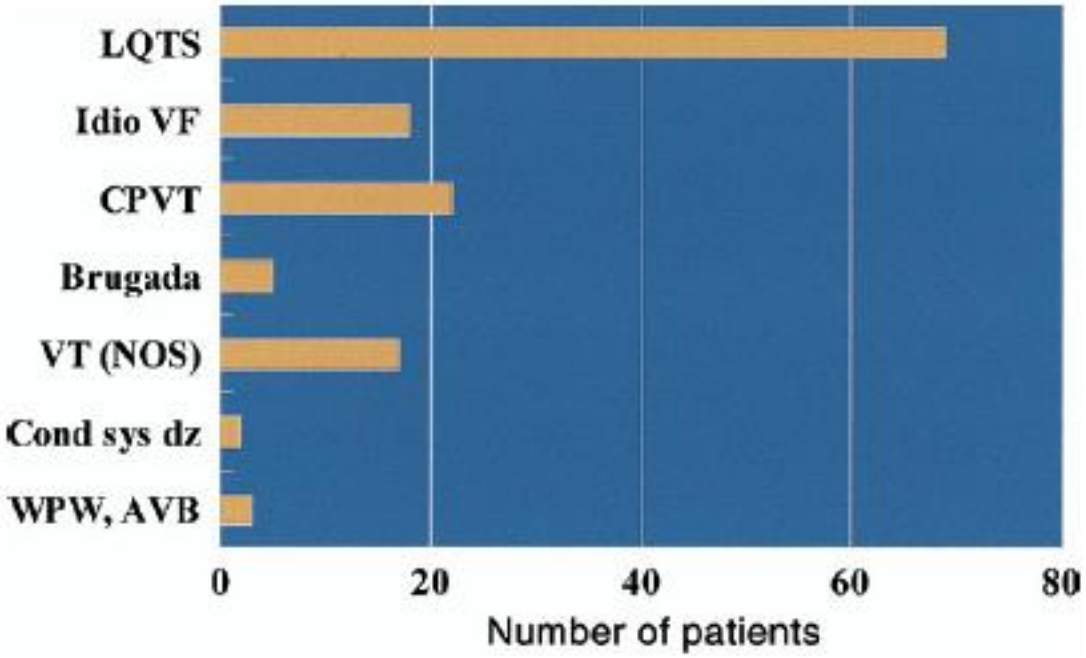


Figure 2 Electrical Diagnoses of Pediatric ICD Recipients With Structurally Normal Hearts

(J Am Coll Cardiol 2008;51:1685–91)

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

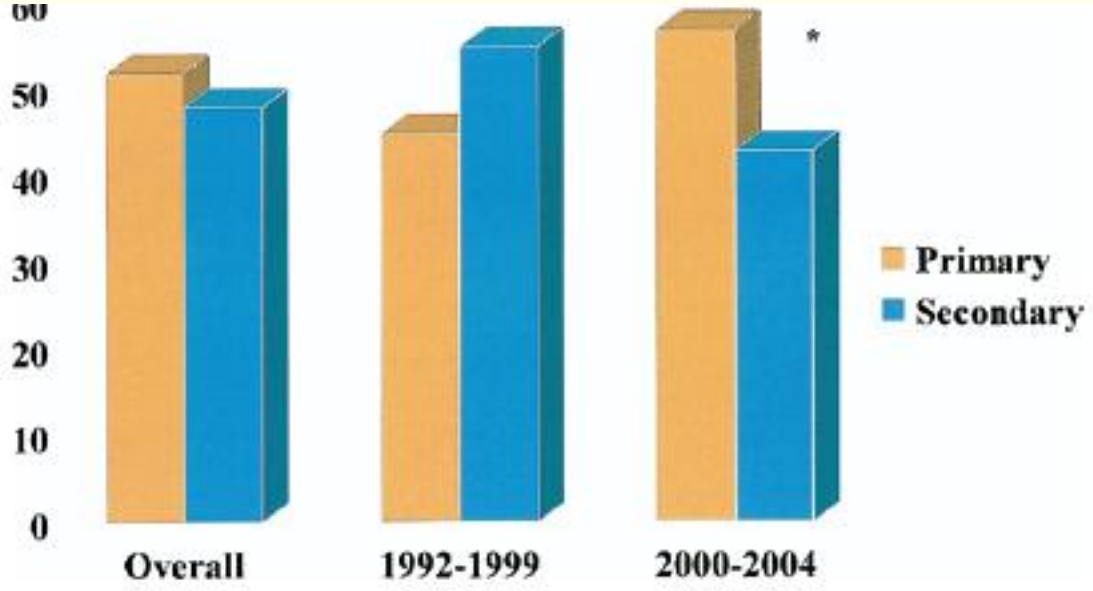
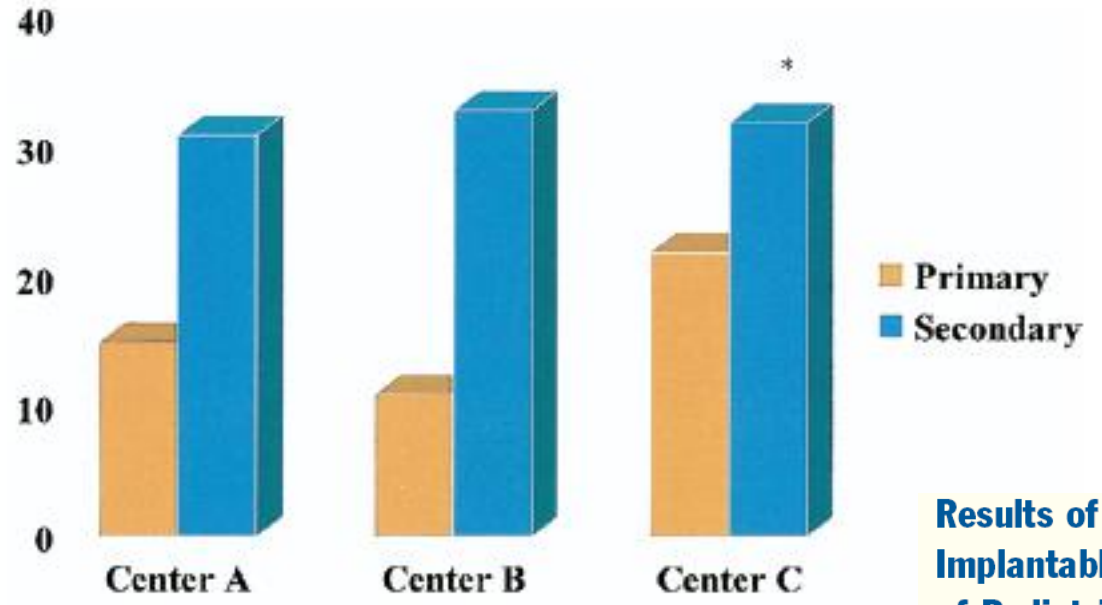
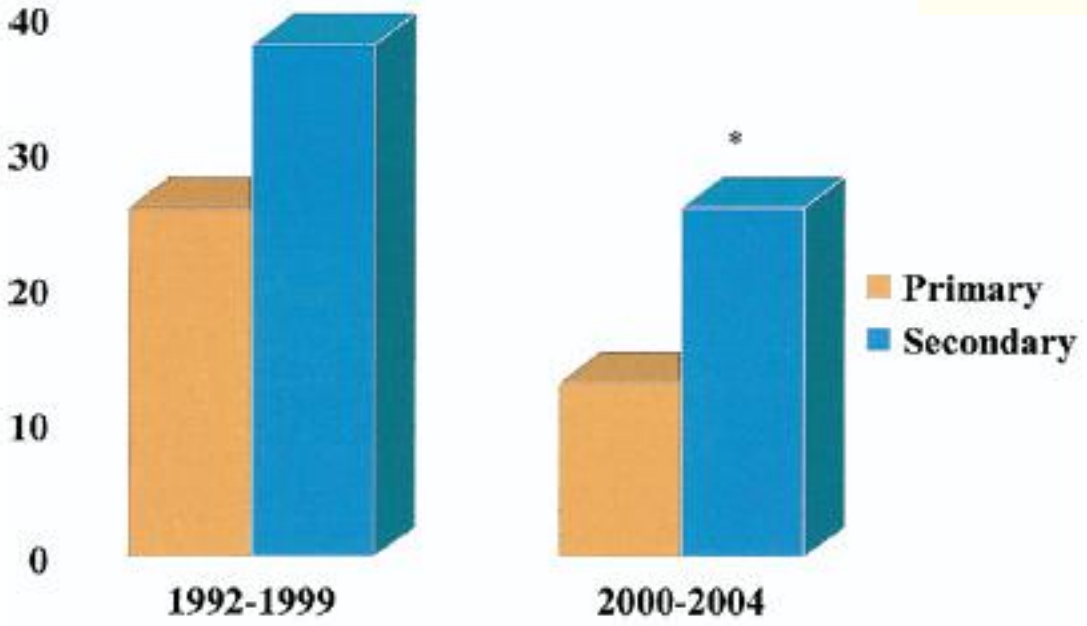


Figure 3 Indications for Implantable Cardioverter-Defibrillator Implantation, Overall and by Era

(J Am Coll Cardiol 2008;51:1685–91)



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



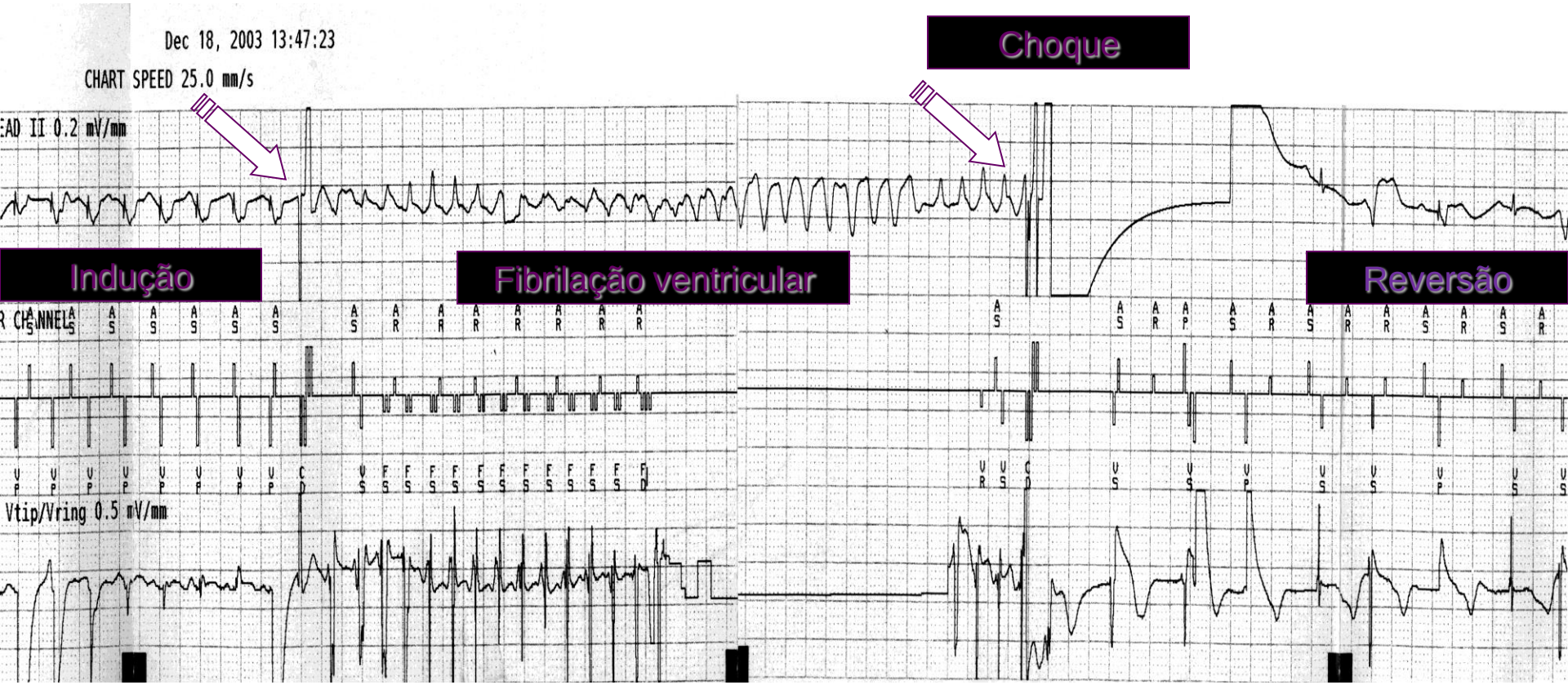
Número Total de Choques apropriados

(J Am Coll Cardiol 2008;51:1685–91)

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

Número Total de Choques inapropriados

(J Am Coll Cardiol 2008;51:1685–91)


Dec 18, 2003 13:47:23
CHART SPEED 25.0 mm/s
EAD II 0.2 mV/mm
Choque
Indução
Fibrilação ventricular
Reversão
R CHANNEL
Vtip/Vring 0.5 mV/mm

Defibrillator indications and implantation in young children

Charles I. Berul, MD, FHRS

From the Department of Cardiology, Children's Hospital Boston, Harvard Medical School, Boston, Massachusetts.

Table 1 Implantable defibrillator lead route options in pediatric patients

	Advantages	Disadvantages
Transvenous	Relatively easy implant; common use; approved indication	Lead fractures; extractions difficult; vascular obstruction
Epicardial patch	Long history, follow-up; approved use; surgeons familiar; good DFT	Patch failure; buckling; restrictive pericardial physiology
Subcutaneous array or coil*	No transvenous coil or epicardial patch; minimally invasive	Limited long-term data; higher DFT
Pericardial coil*	No need for transvenous access or epicardial patch; low DFT	Requires surgeon; adhesions may limit video-assisted thoracoscopic surgery; limited follow-up data
Subcutaneous leadless implantable* cardioverter-defibrillator	No need for transvenous or epicardial access; minimally invasive	Limited long-term data; higher DFT; no chronic pacing or antitachycardia pacing

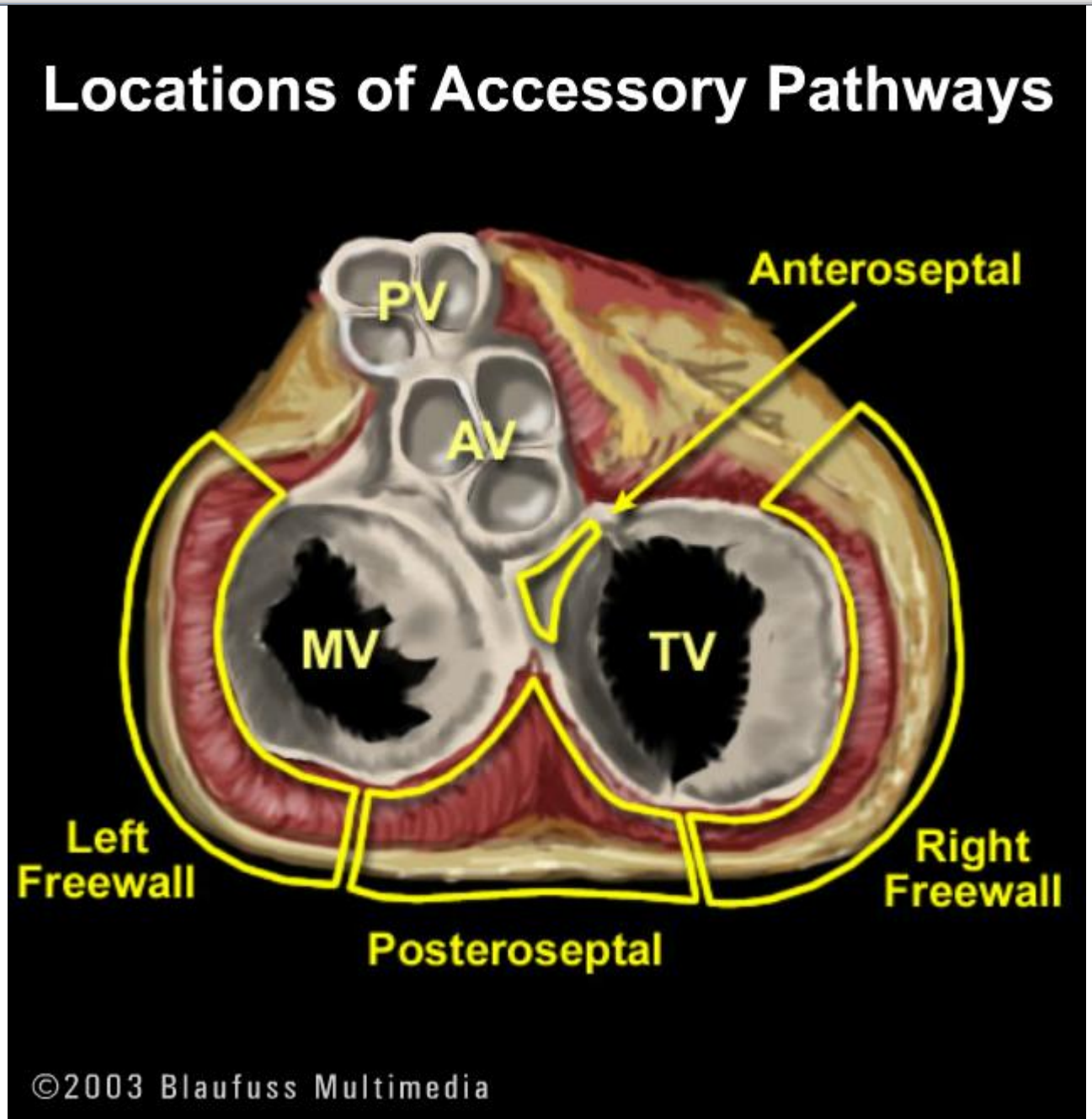
Results of a Multicenter Retrospective Implantable Cardioverter-Defibrillator Registry of Pediatric and Congenital Heart Disease Patients

Charles I. Berul, George F. Van Hare, Naomi J. Kertesz, Anne M. Dubin, Frank Cecchin, Kathryn K. Collins, Bryan C. Cannon, Mark E. Alexander, John K. Triedman, Edward P. Walsh, and Richard A. Friedman

J. Am. Coll. Cardiol. 2008;51;1685-1691

	No. of Complications
Acute Complications (Perioperative or Within 30 Days of Implant)	
Lead dislodgement	13
Inability to defibrillate or unacceptable DFT	9
Bleeding or pocket hematoma	8
Infection	7
Unsuccessful transvenous lead placement	6
Electrical storm	5
Hemothorax or pneumothorax	4
EMD/PEA	4
Skin erosion	3
SVC syndrome	2
Skin burns	2
Pneumonia and ileus	1
Total acute complications	64 (in 55 patients)

Possibilidades de localização da via acessória

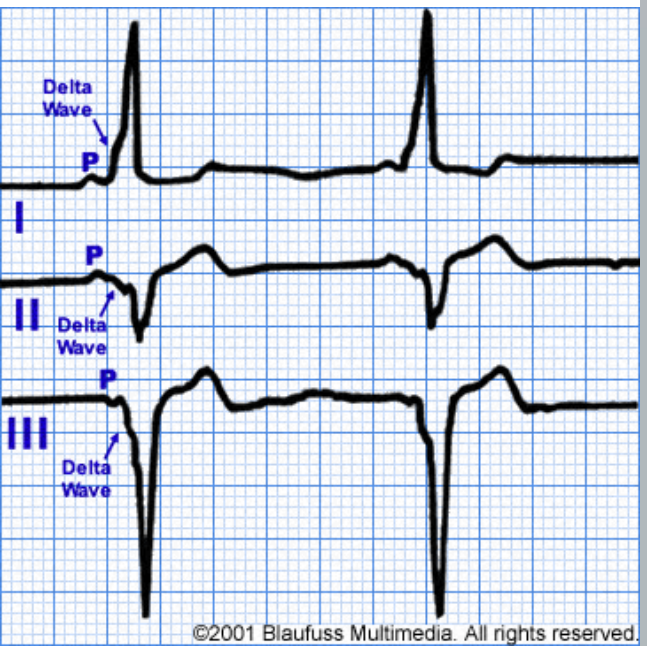


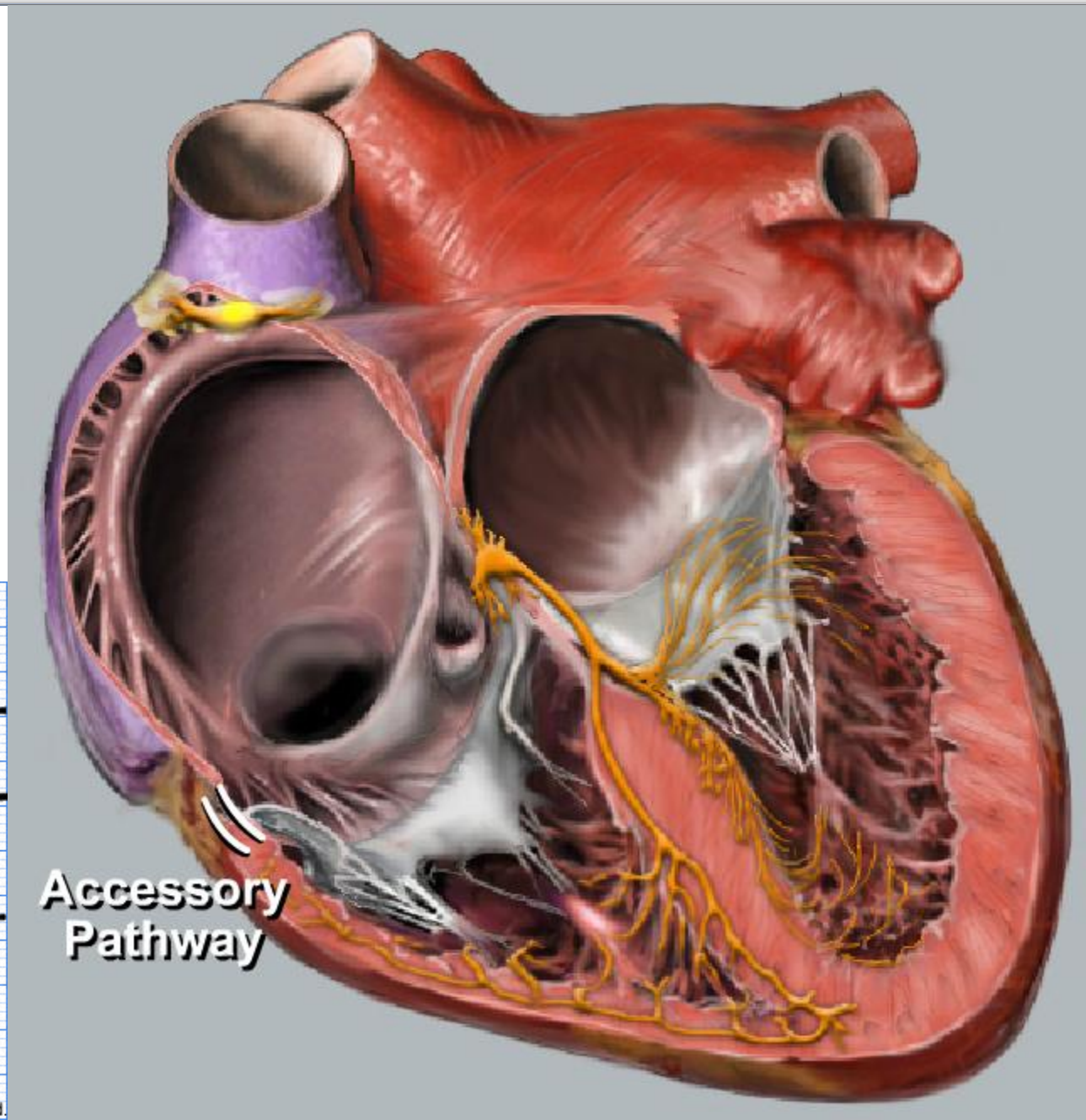
Pré excitação ventricular

PR curto

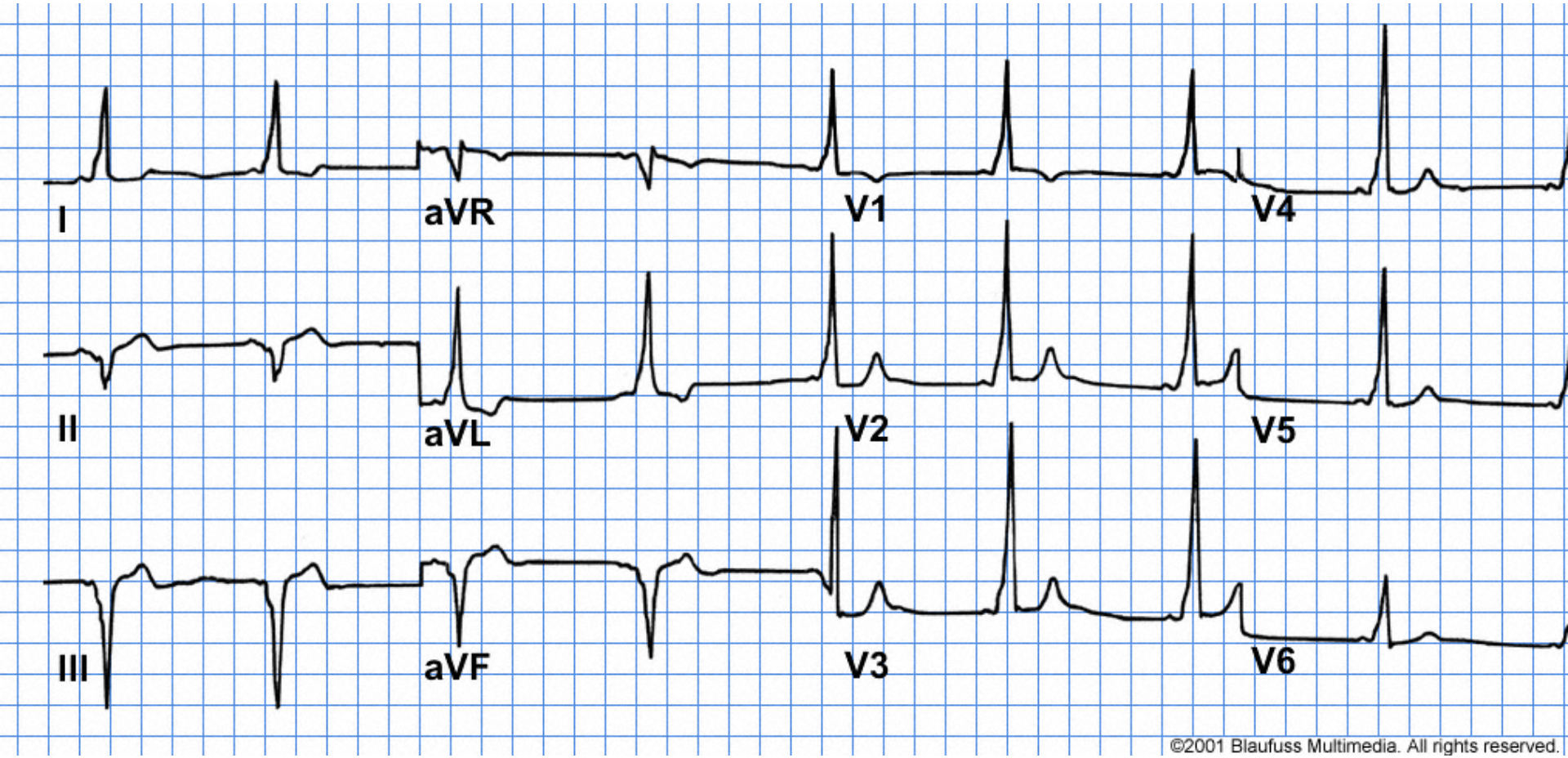
Onda delta

Alteração da repolarização

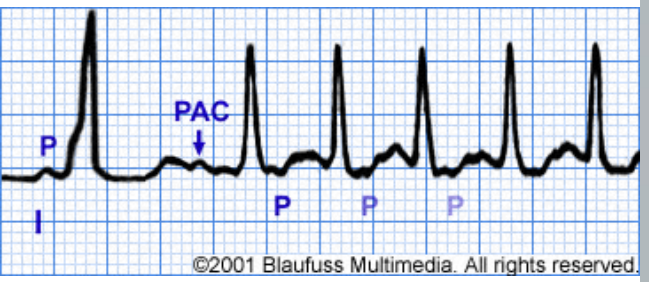


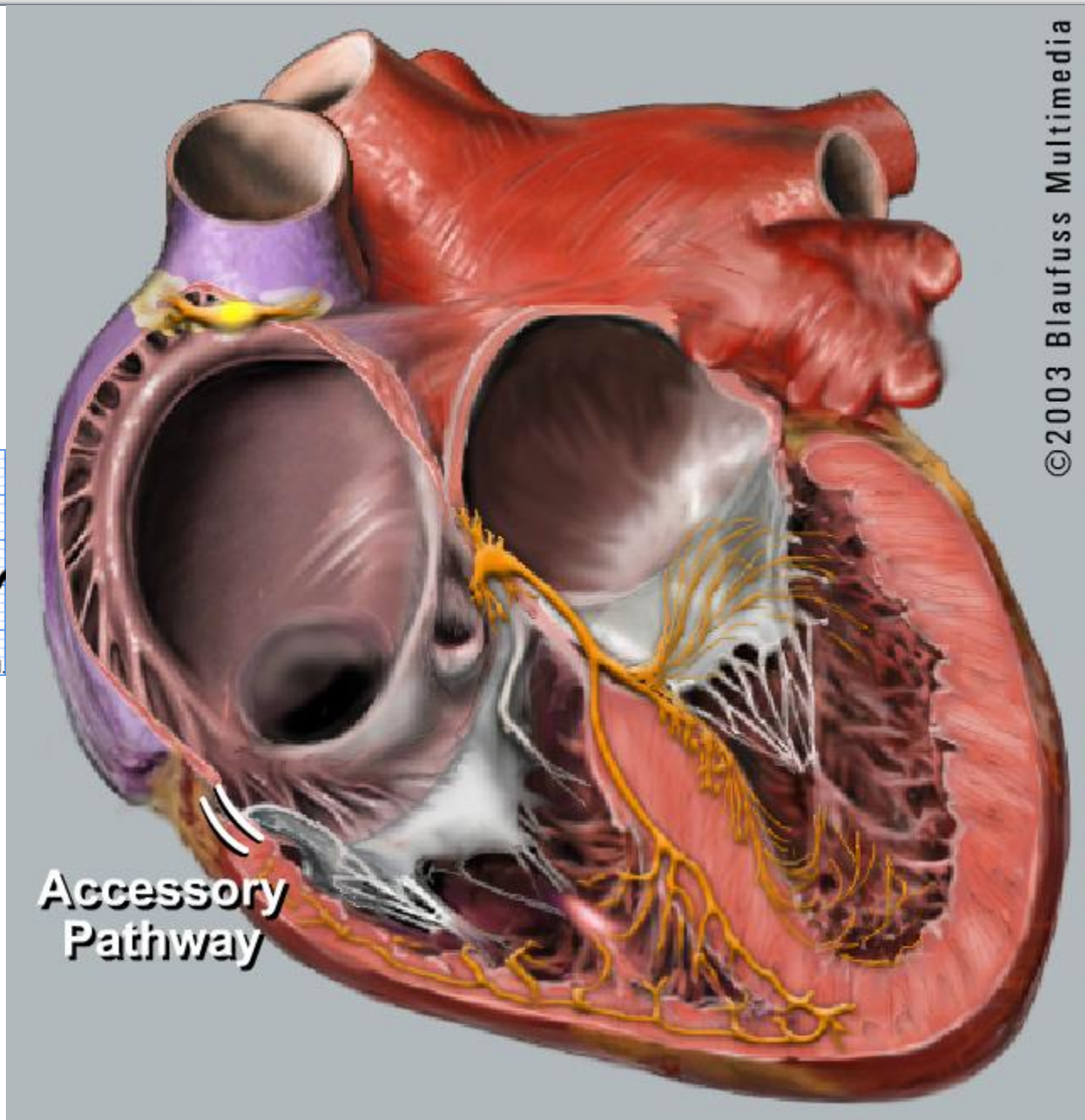


ECG Pré excitação

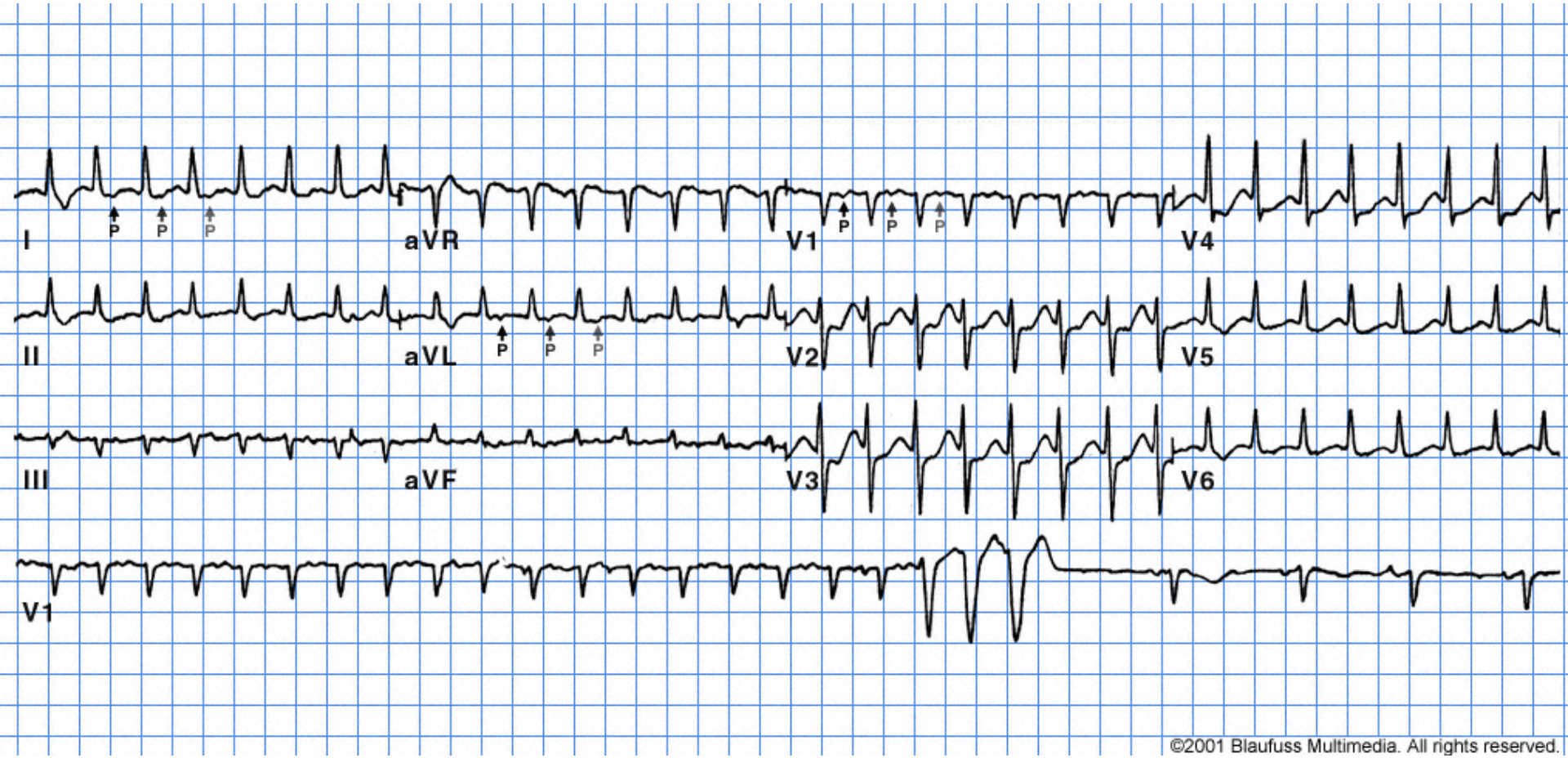


TRAV início

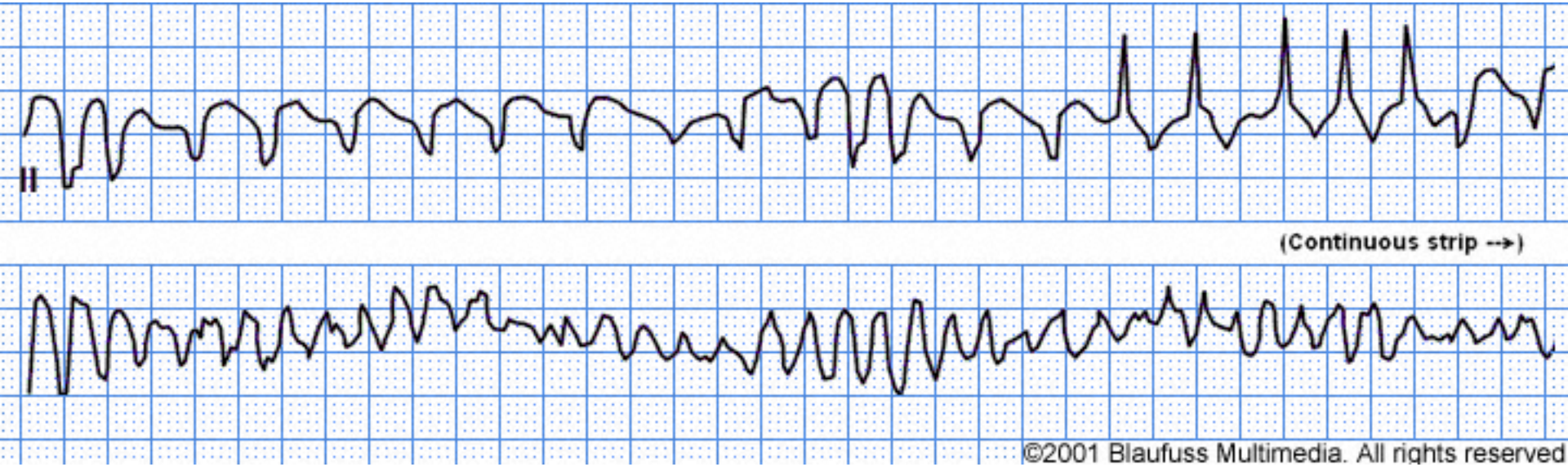

PAC
P
I
P
P
P
©2001 Blaufuss Multimedia. All rights reserved.


Accessory
Pathway
©2003 Blaufuss Multimedia

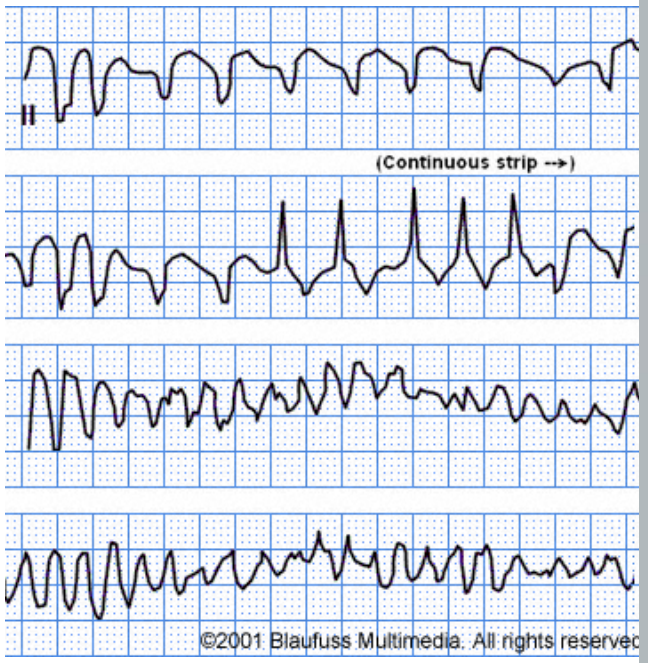
TRAV ECG da crise

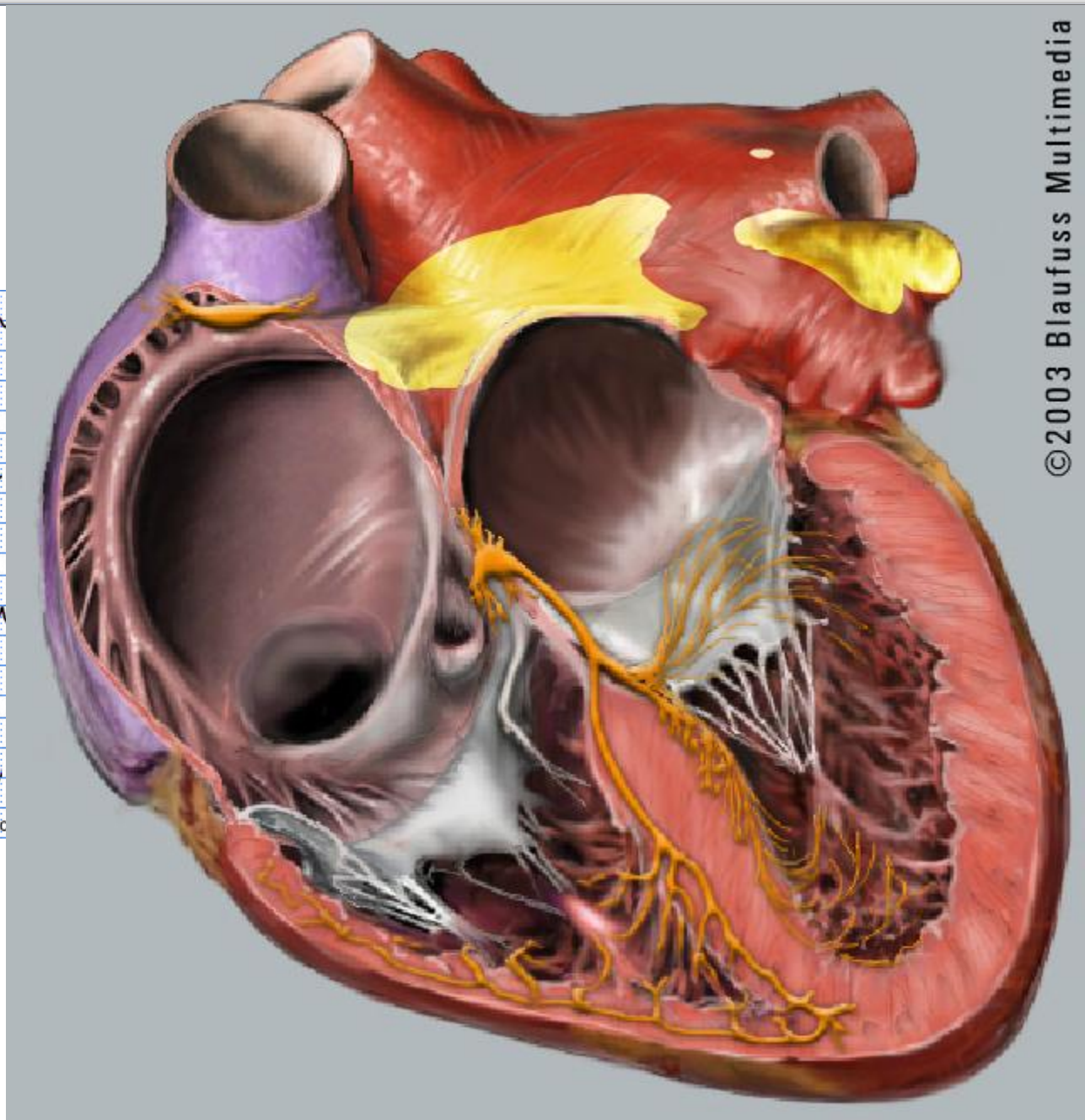


FA + pré excitação

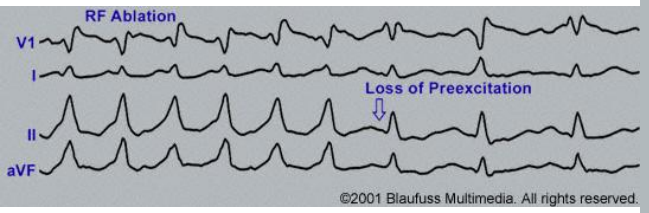


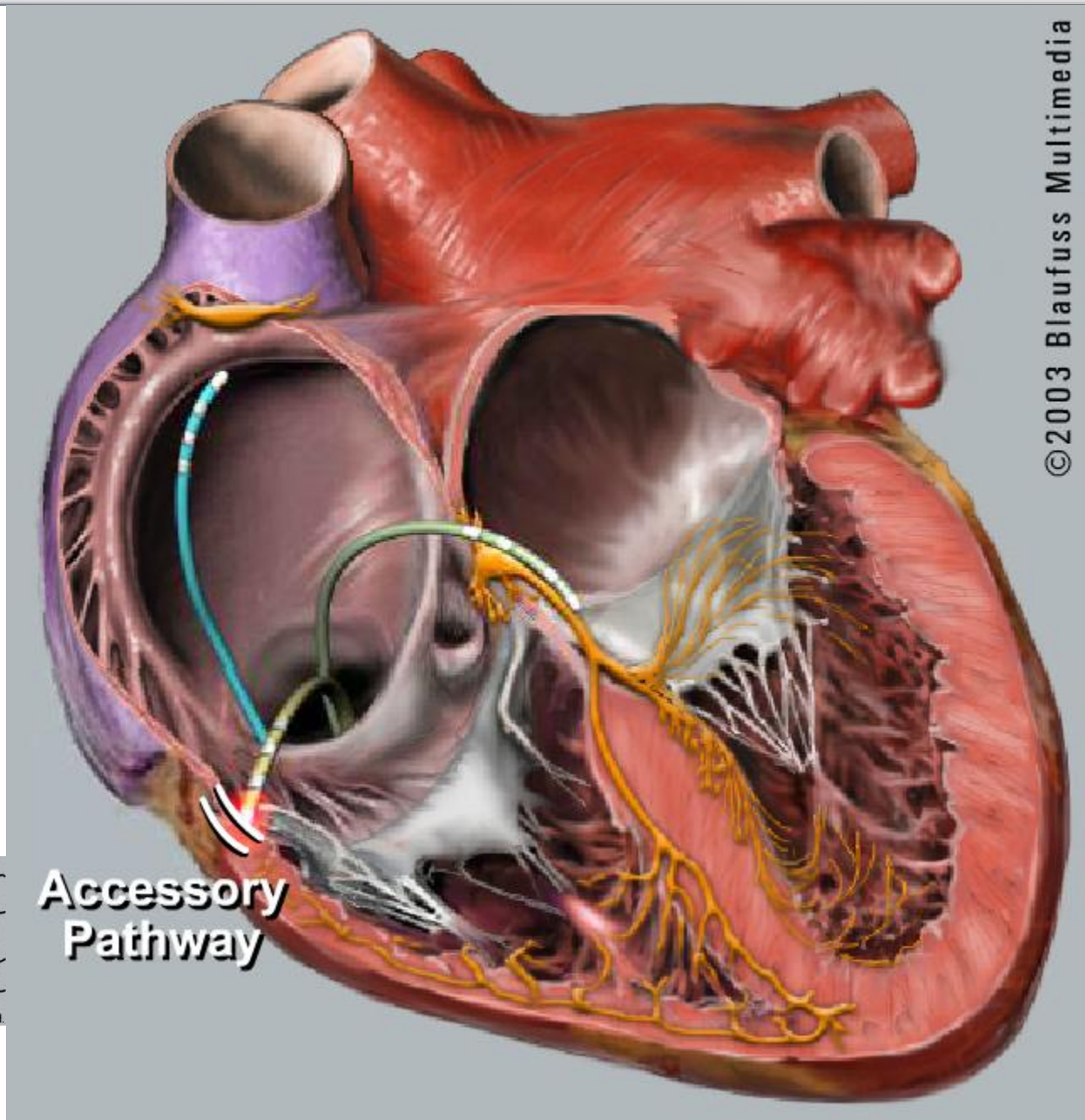
FA + pré excitação

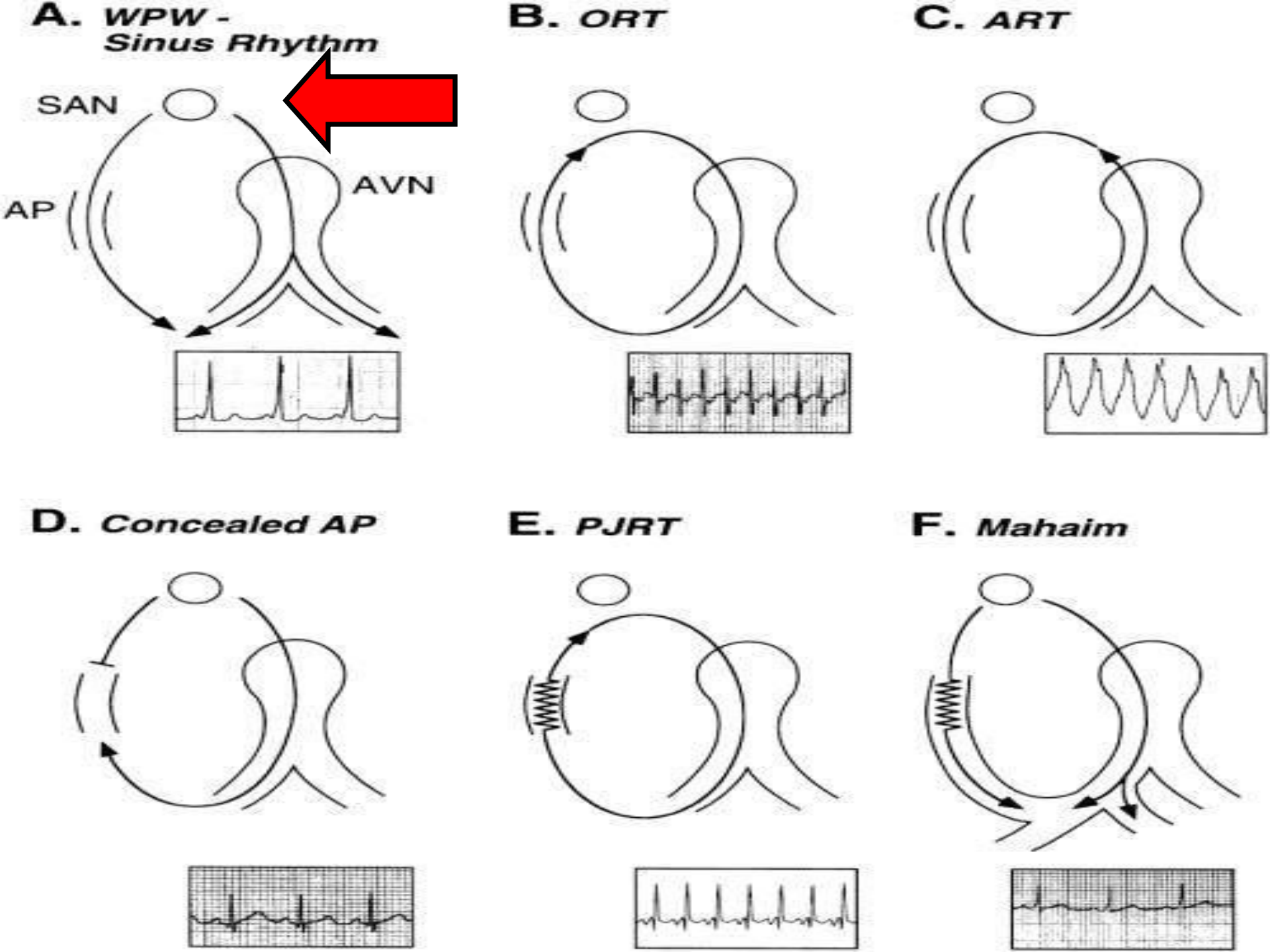


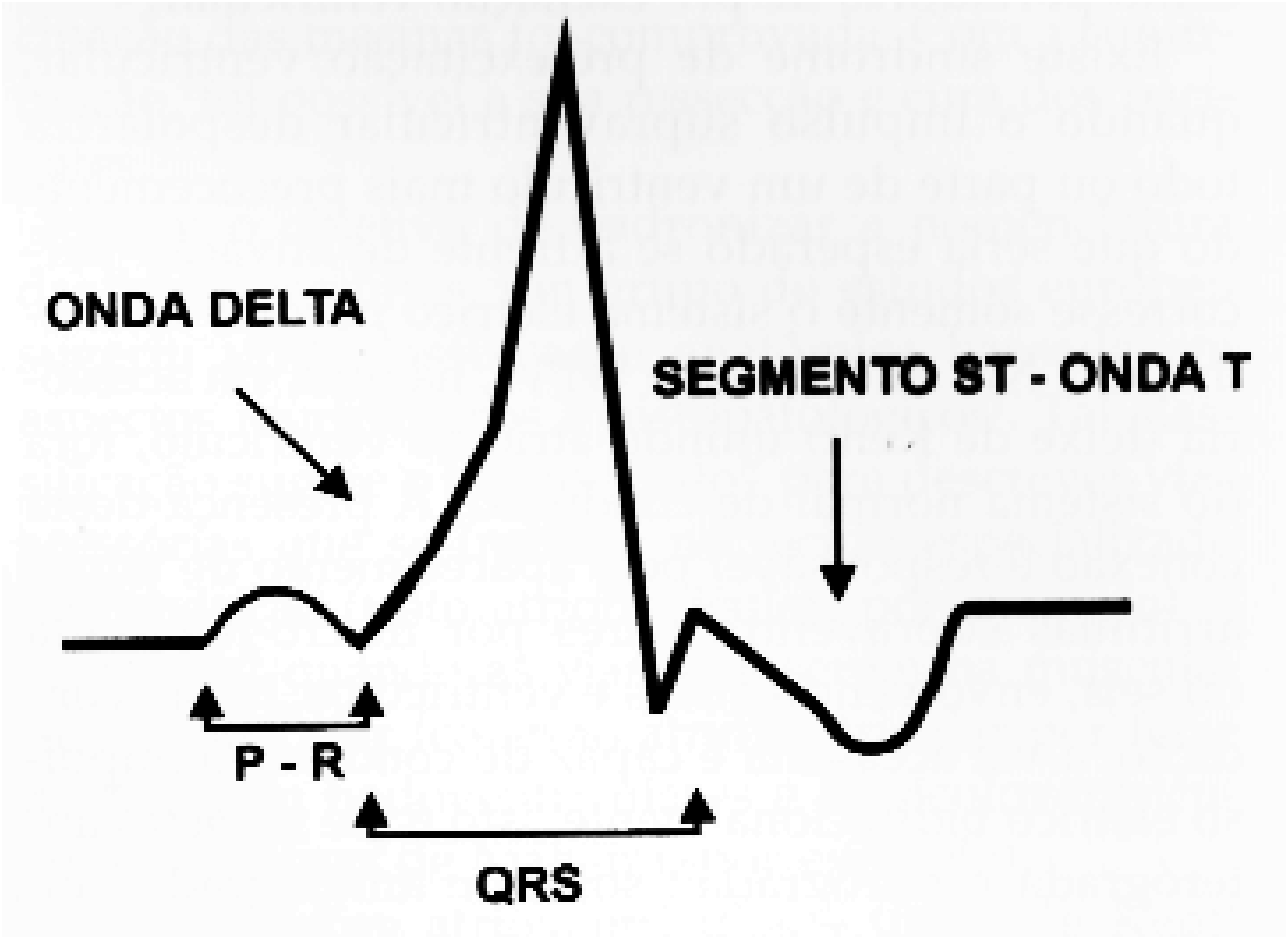


Ablação por radiofrequência

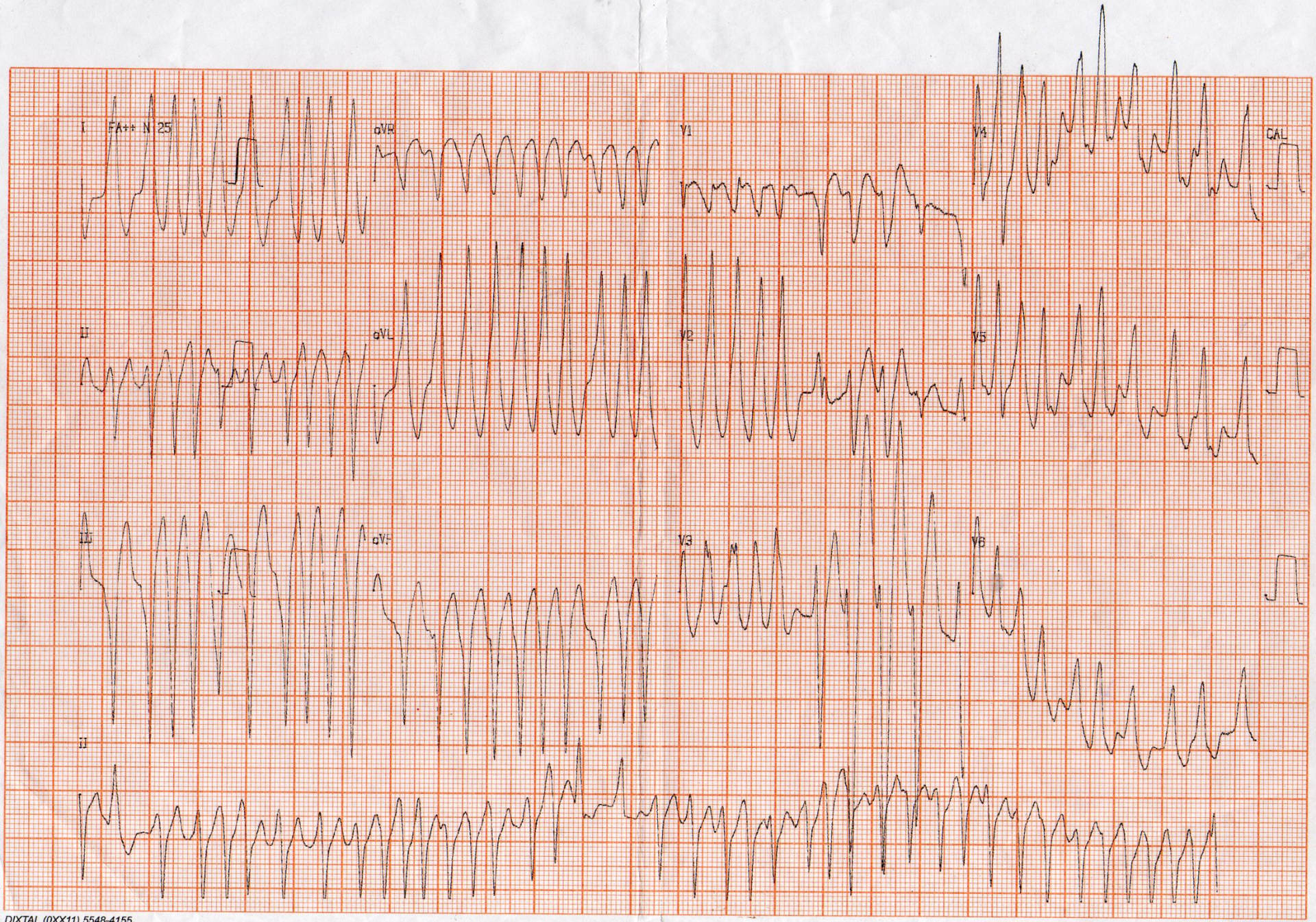




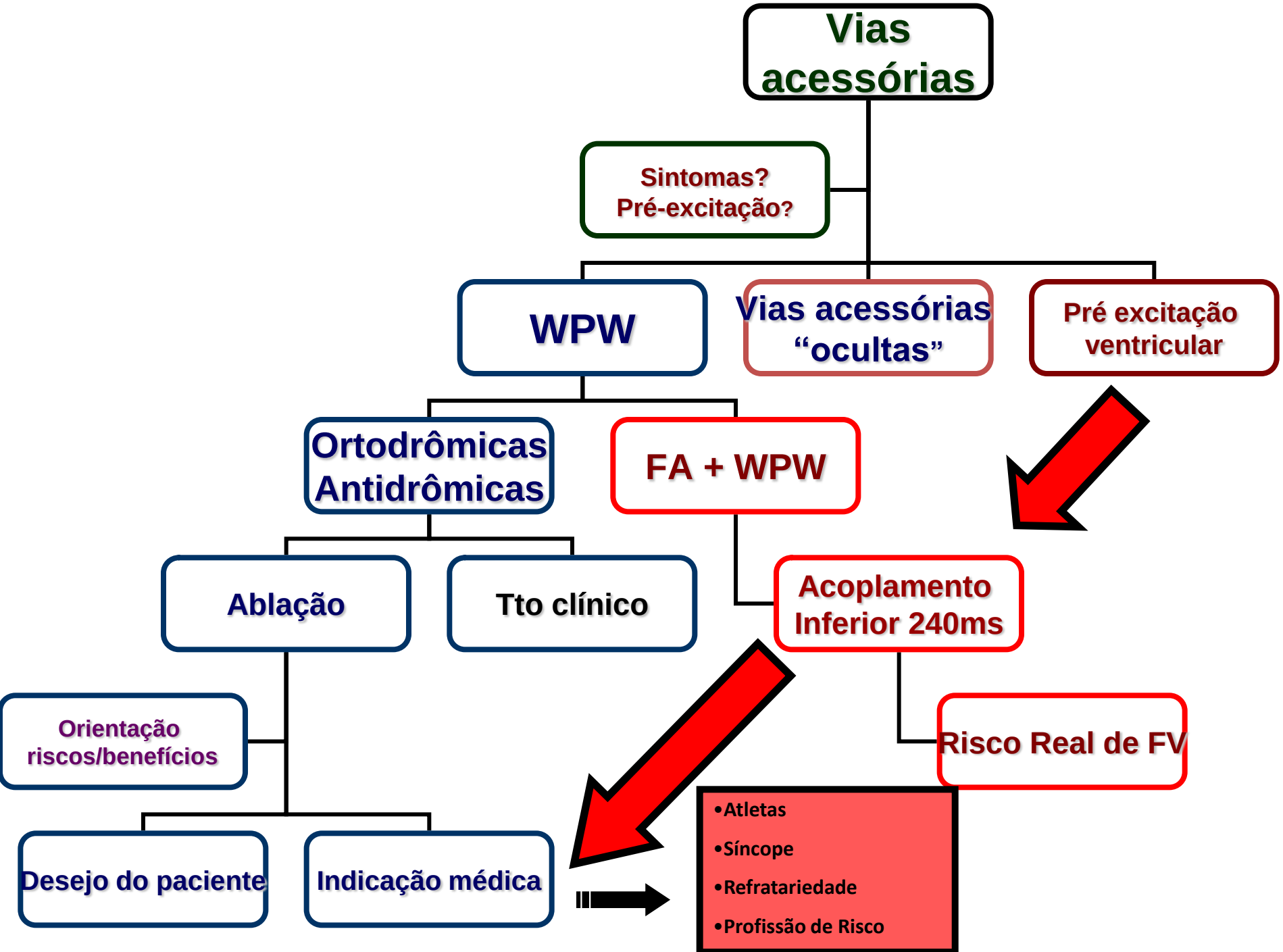

A. WPW - Sinus Rhythm
SAN
AP
AVN
B. ORT
C. ART
D. Concealed AP
E. PJRT
F. Mahaim


ONDA DELTA
SEGMENTO ST - ONDA T
P - R
QRS

I
aVR
V1
V4
CAL
II
aVL
V2
V5
III
aVF
V3
V6
II



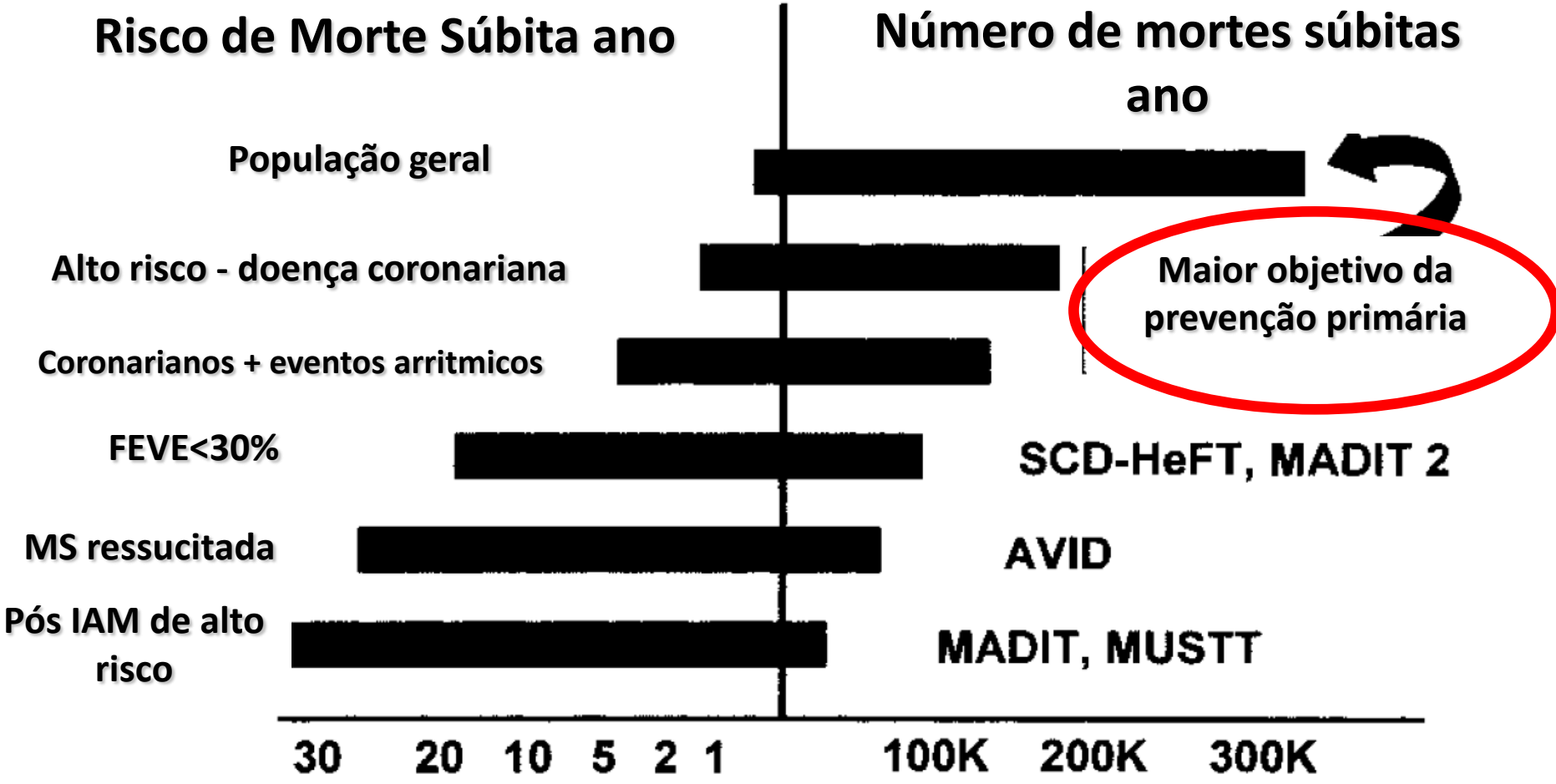
DIXTAL (0XX11) 5548-4155
DIXTAL (0XX11) 5548-4155


Vias acessórias
Sintomas?
Pré-excitação?
WPW
Vias acessórias "ocultas"
Pré excitação ventricular
Ortodrômicas
Antidrômicas
FA + WPW
Ablação
Tto clínico
Acoplamento Inferior 240ms
Orientação riscos/benefícios
Risco Real de FV
Desejo do paciente
Indicação médica
•Atletas
•Síncope
•Refratariedade
•Profissão de Risco

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

Morte cardíaca súbita



Circulation -2004; 109: 2685-2691

- **Profilaxia primária**

- **Profilaxia secundária**

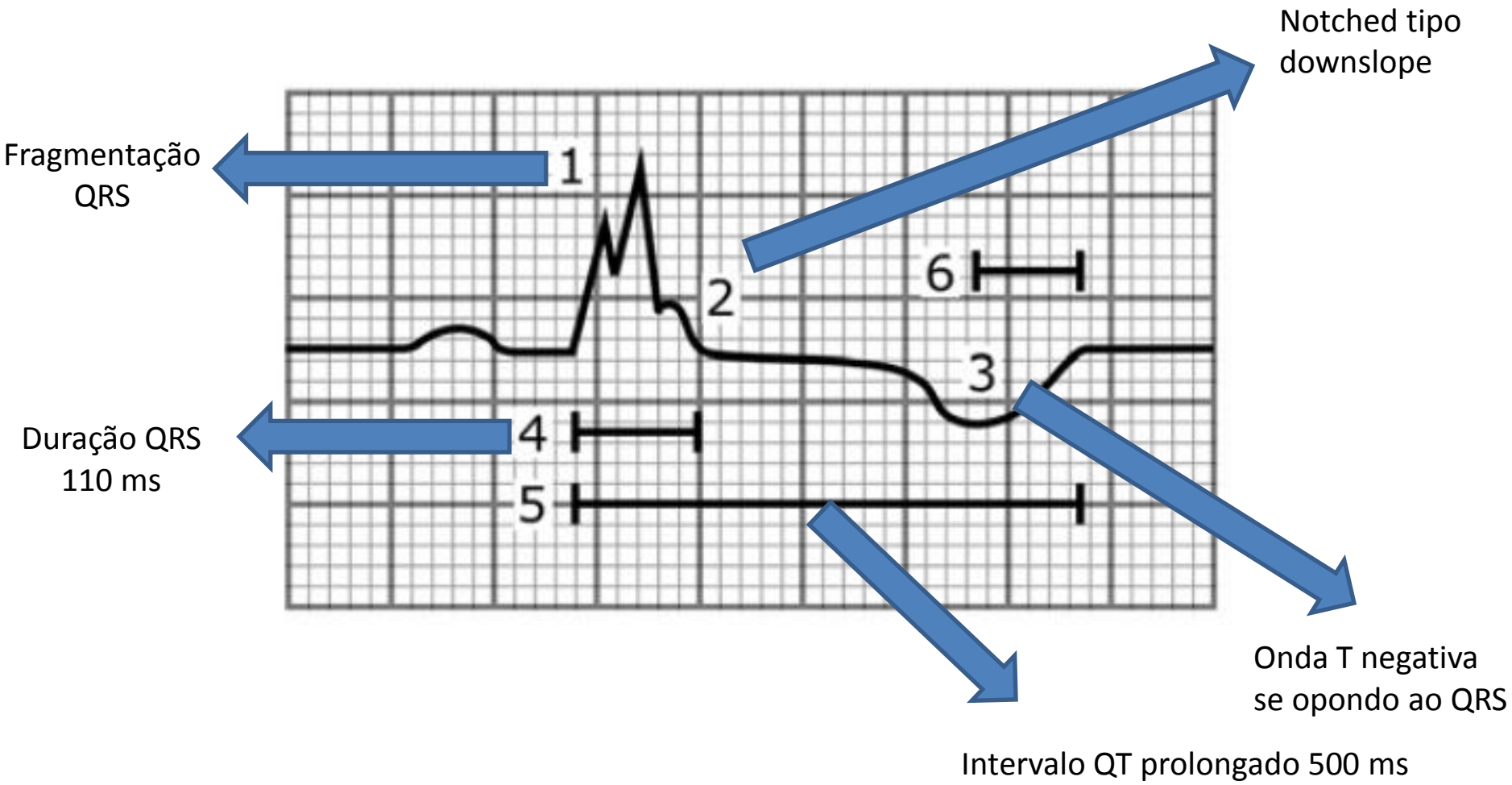
Marcadores ECG de morte súbita em adultos

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)[a]
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)

[a]Cardiac death.

Principais marcadores QRS com valor prognóstico

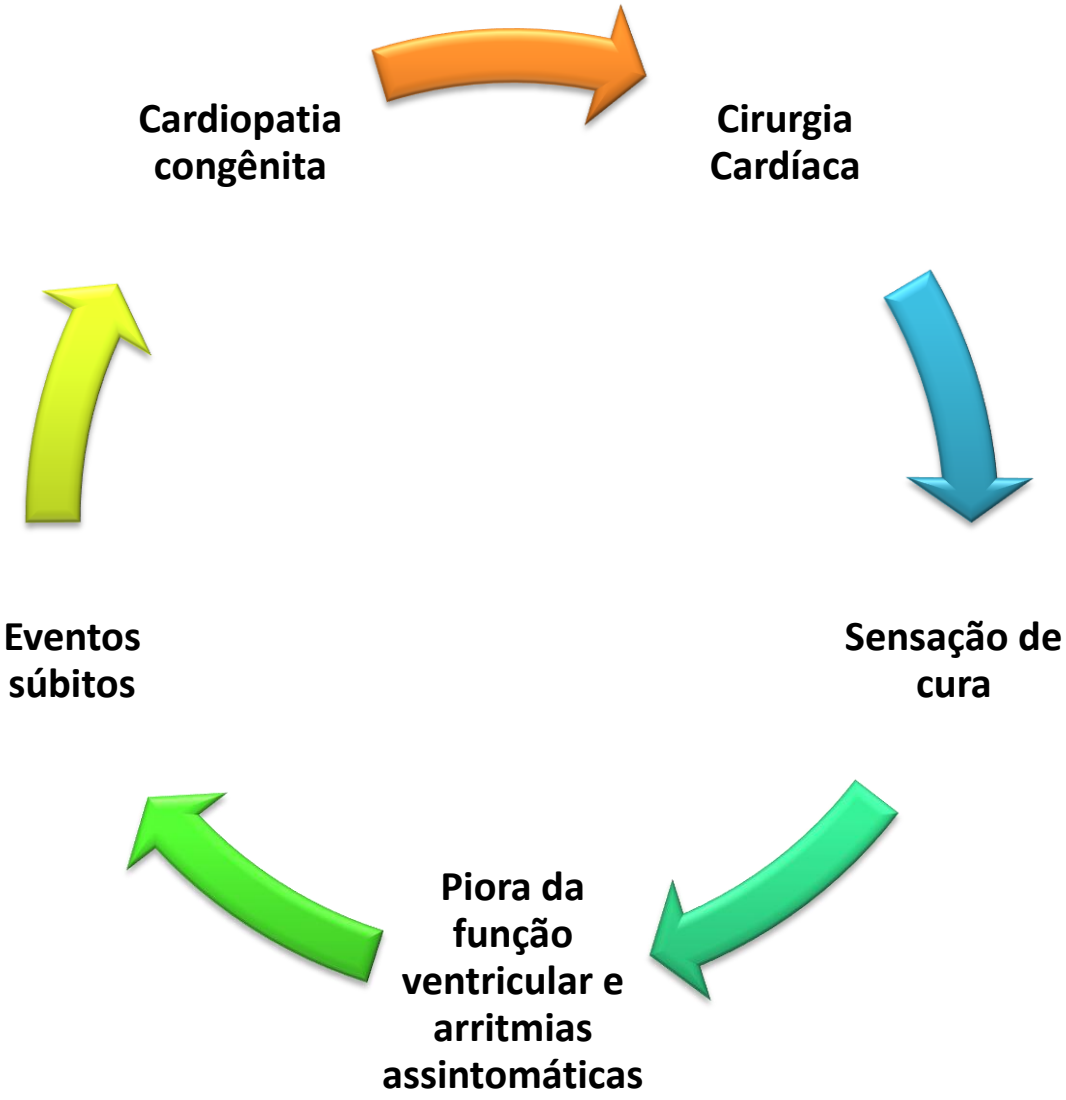


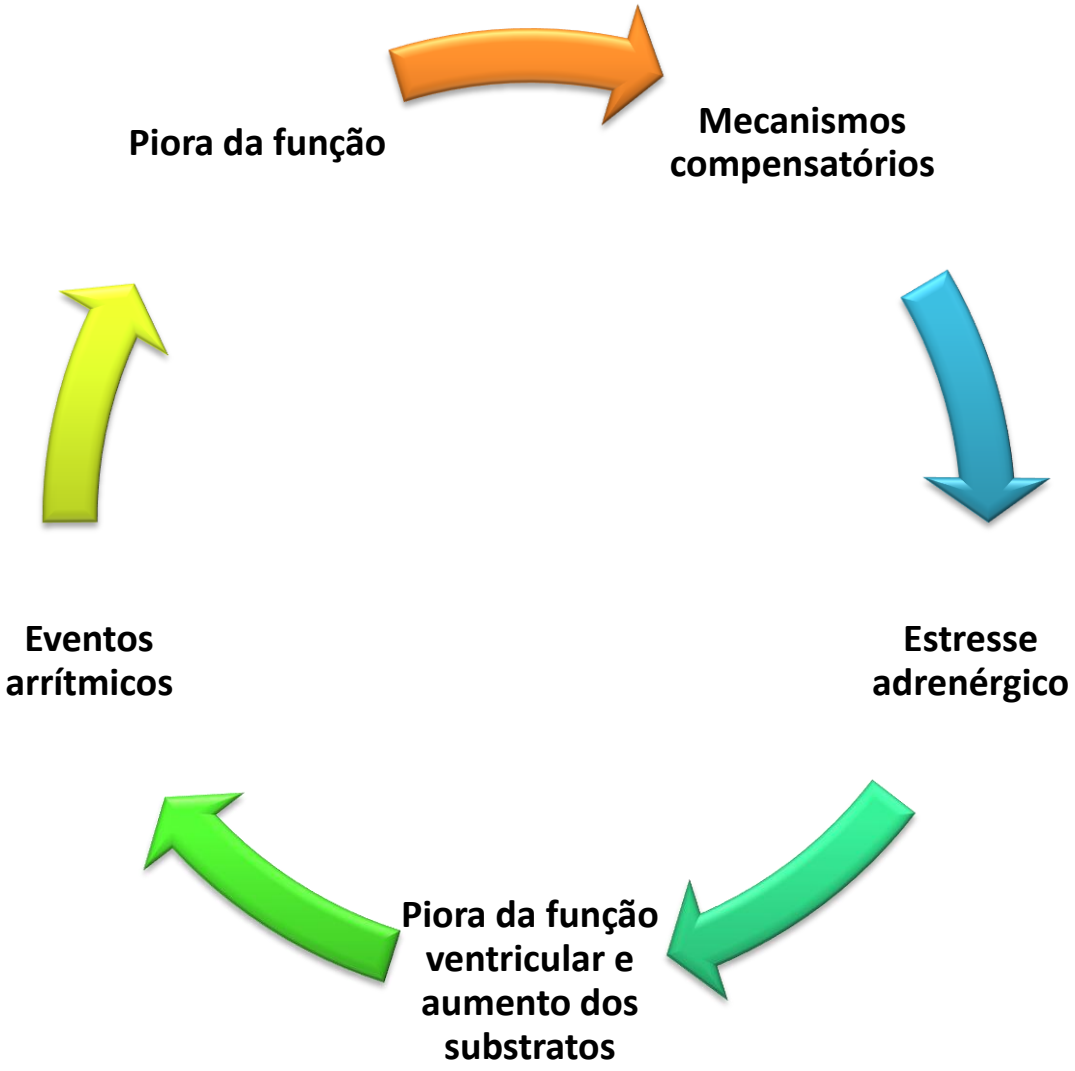
Marcadores ECG na população pediátrica e em jovens

Marcadores ECG na população pediátrica e em jovens

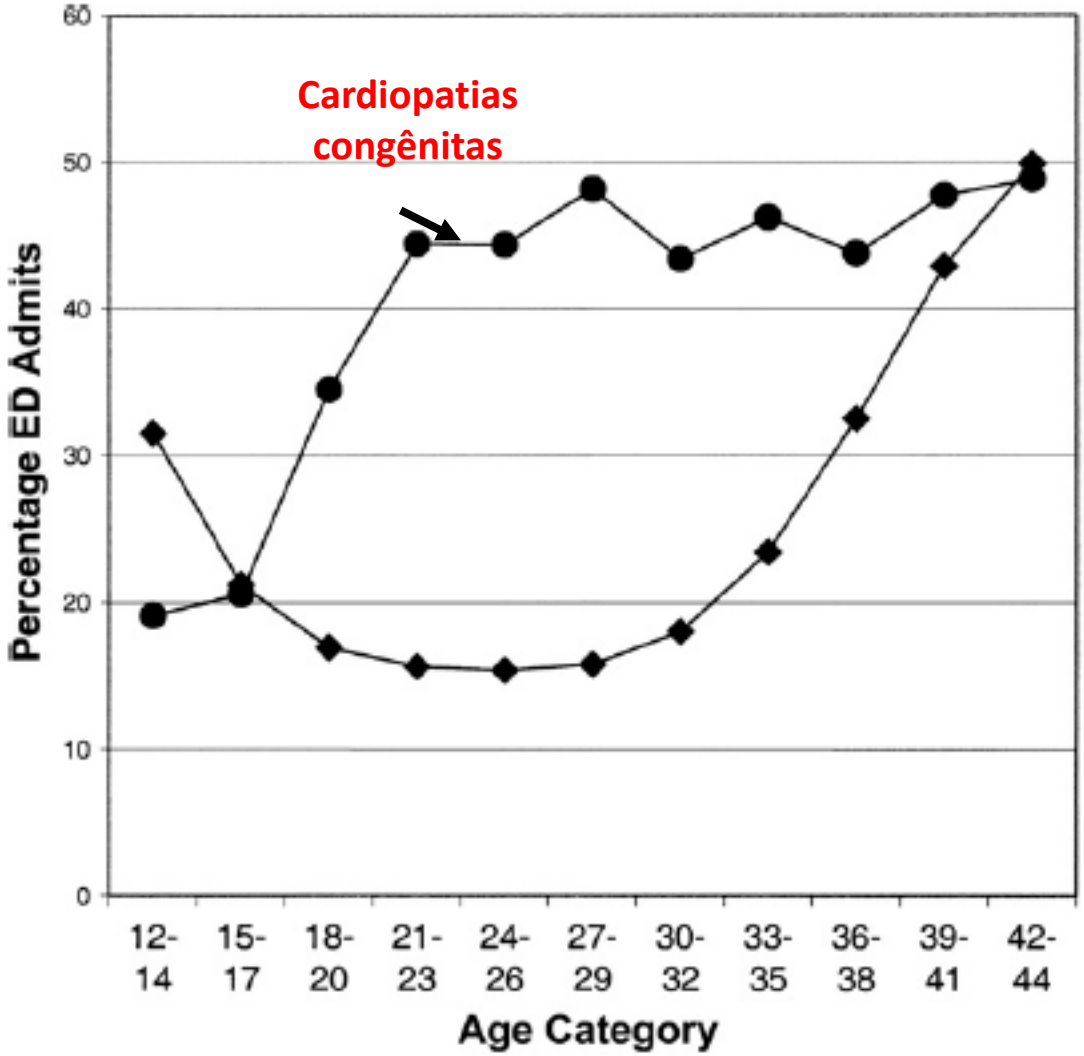
Tabela 84.3 – Correlação entre tipo de cardiopatia congênita e possíveis arritmias

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, *flutter* 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 ventriculosseptoplastia, 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 (*flutter* 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


Cardiopatia congênita
Cirurgia Cardíaca
Sensação de cura
Piora da função ventricular e arritmias assintomáticas
Eventos súbitos

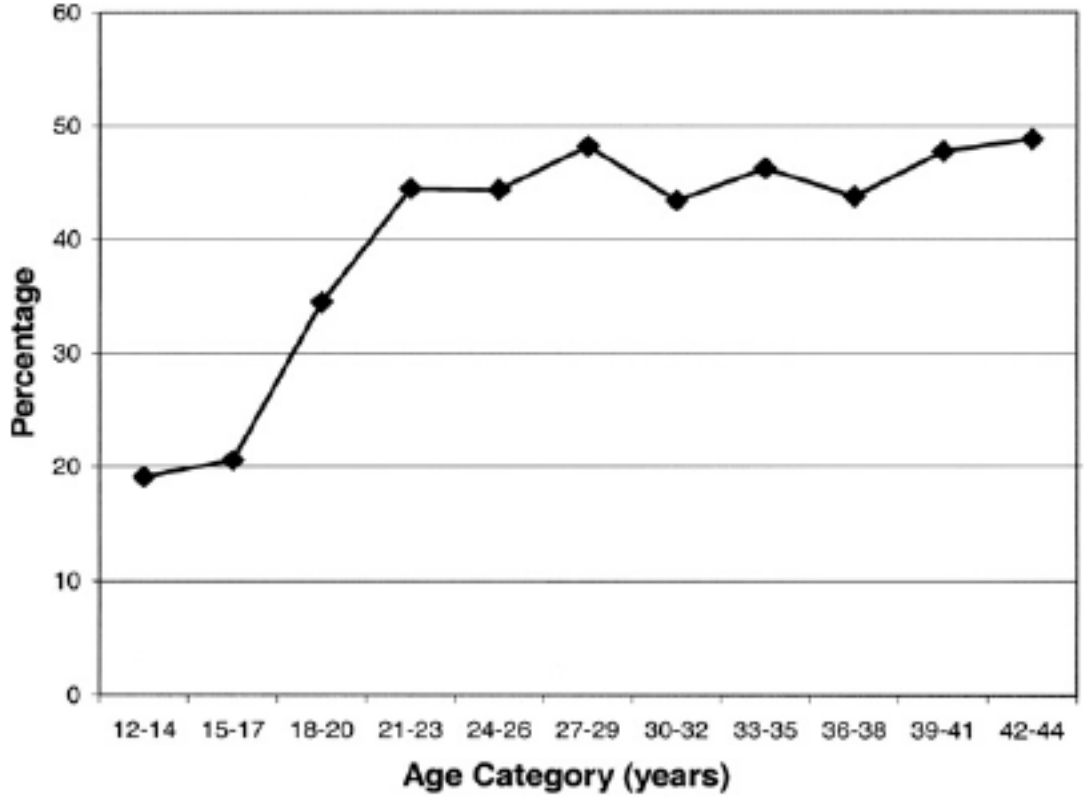

Piora da função
Mecanismos compensatórios
Estresse adrenérgico
Piora da função ventricular e aumento dos substratos
Eventos arrítmicos

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



(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



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.

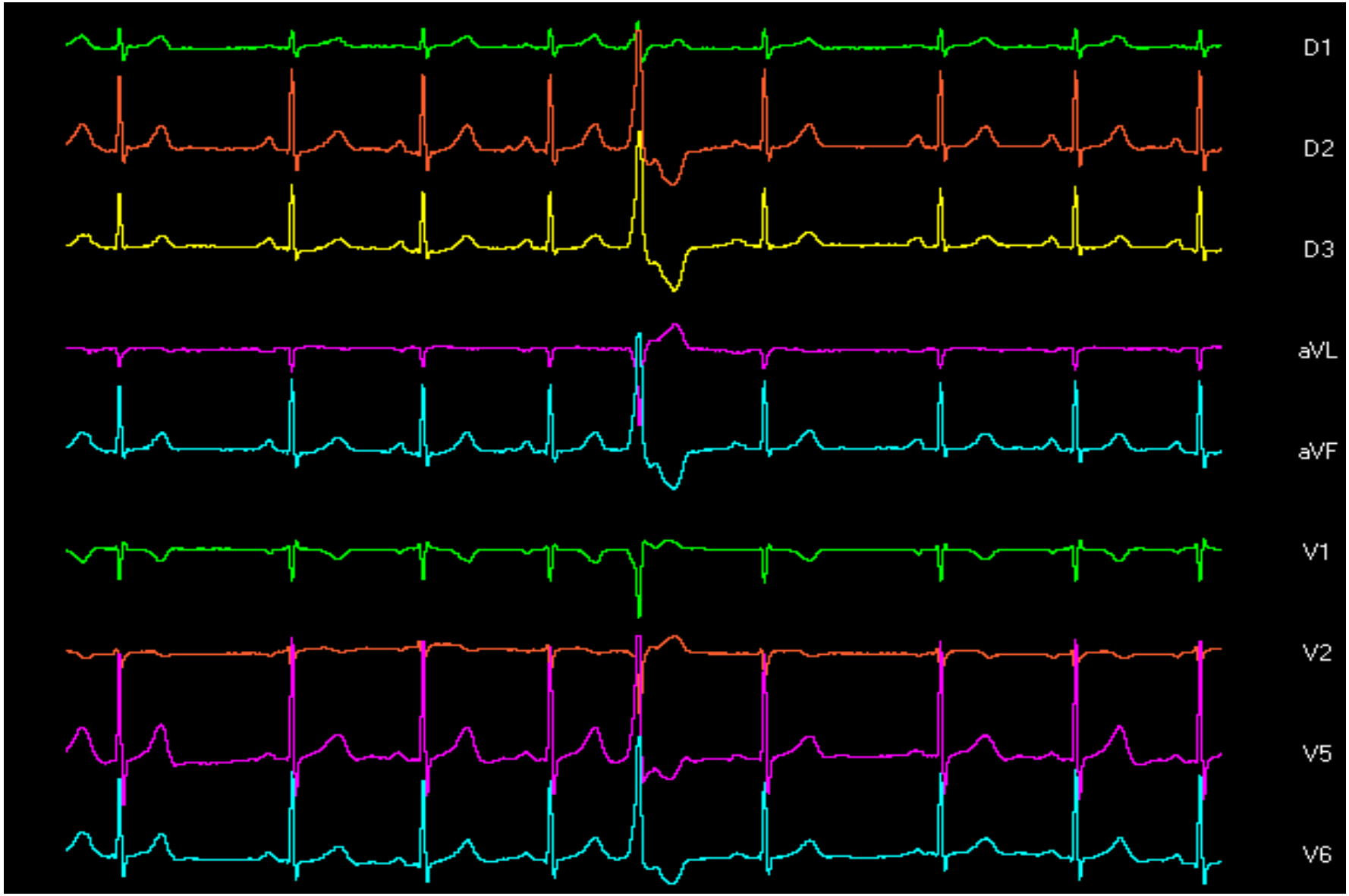
EUROPEAN SOCIETY OF CARDIOLOGY®

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.



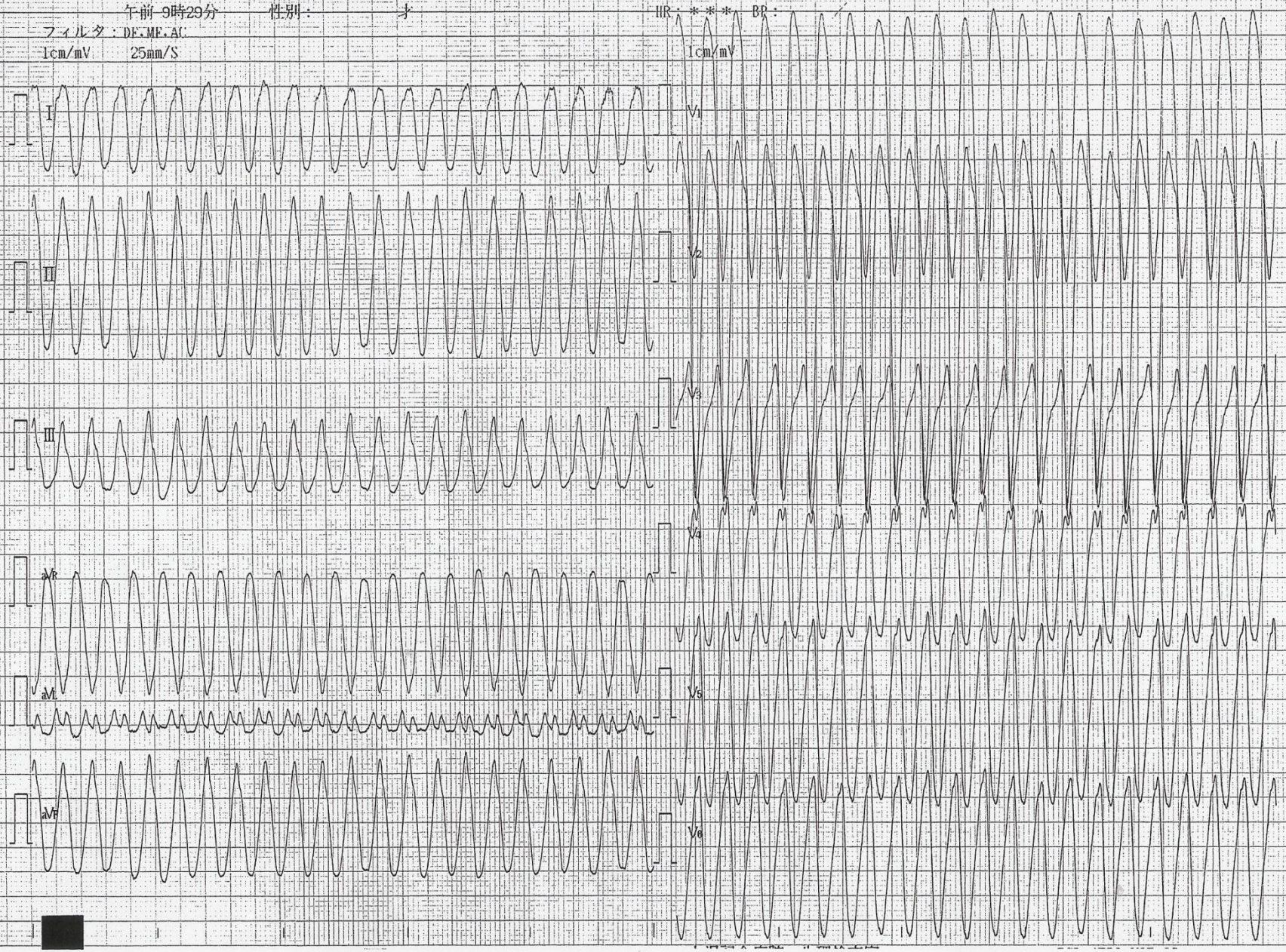
[redacted]

安静時　08年 5月12日　ＩＤ：０００００００７　氏名：

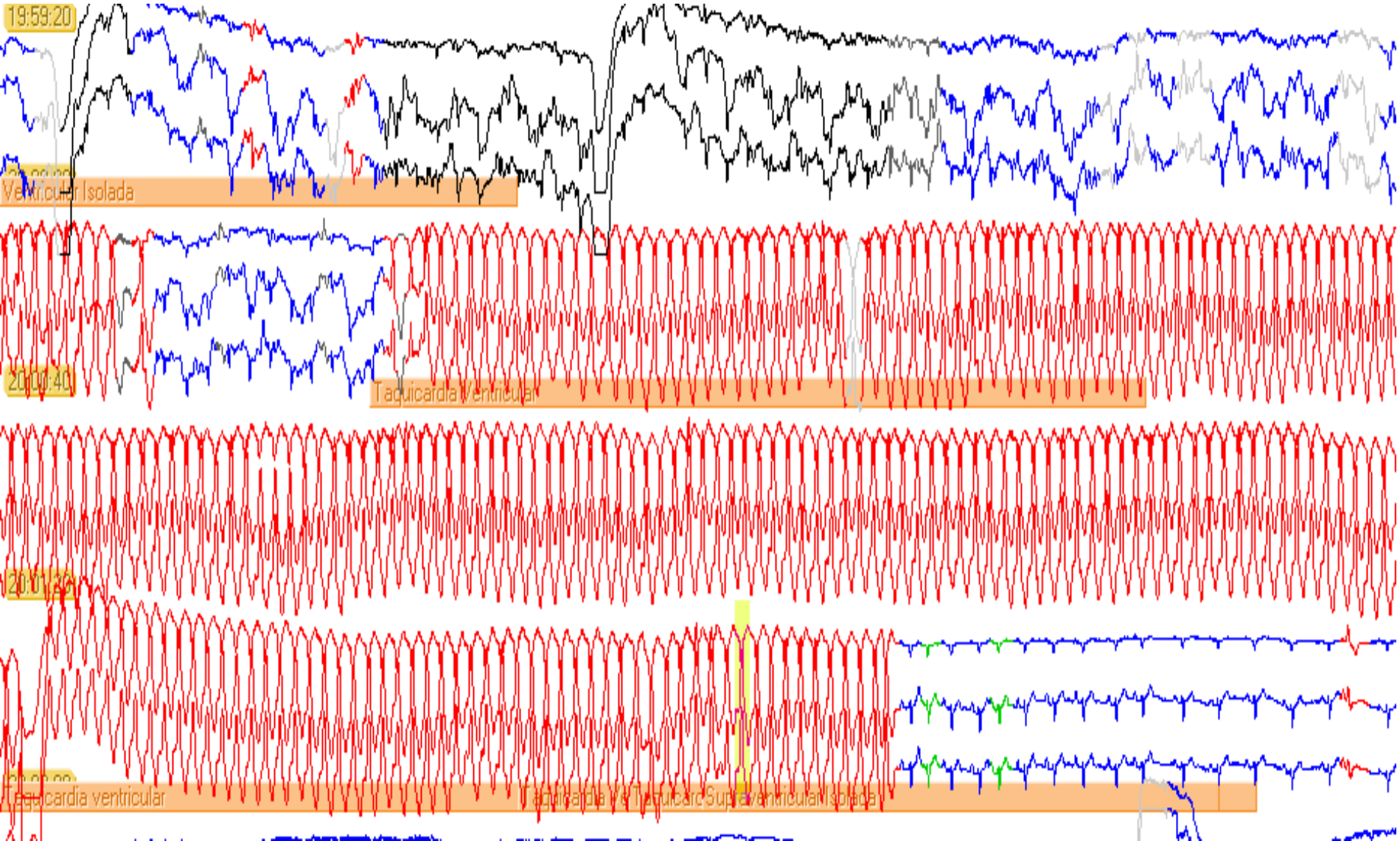
午前 9時29分　性別：　才　HR：＊＊＊　BP：

フィルタ：DF.MF.AC

1cm/mV　25mm/S



[redacted]


19:59:20
Ventricular Isolada
20:00:40
Taquicardia Ventricular
20:01:20
Taquicardia ventricular
Taquicardia Supraventricular Isolada

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

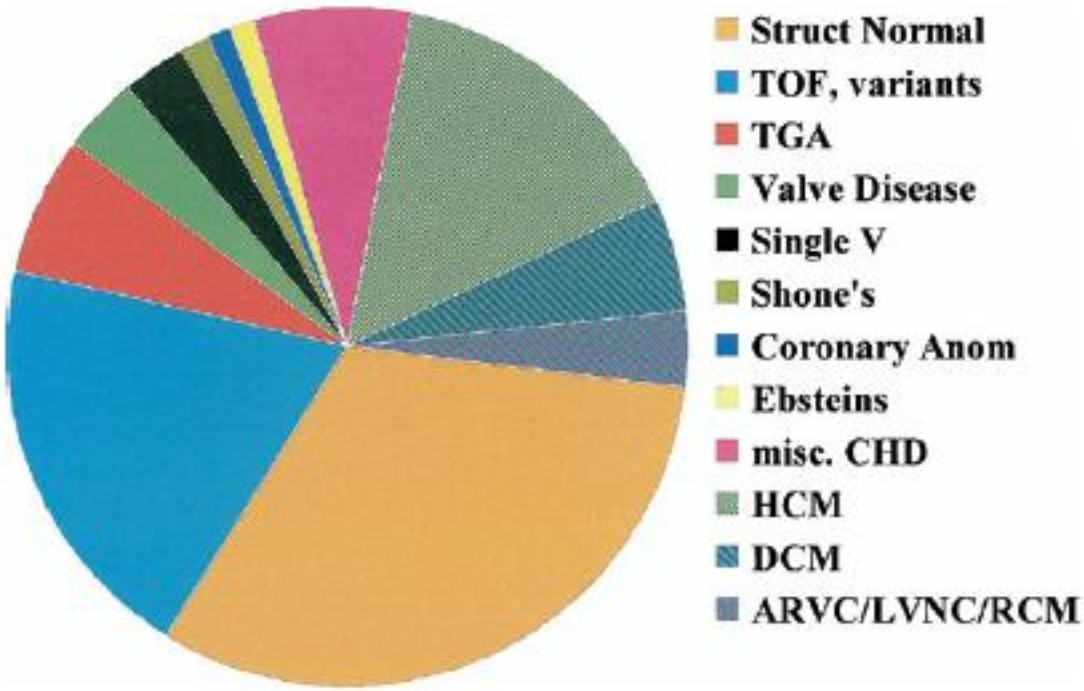


Figure 1 Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients

(J Am Coll Cardiol 2008;51:1685–91)

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

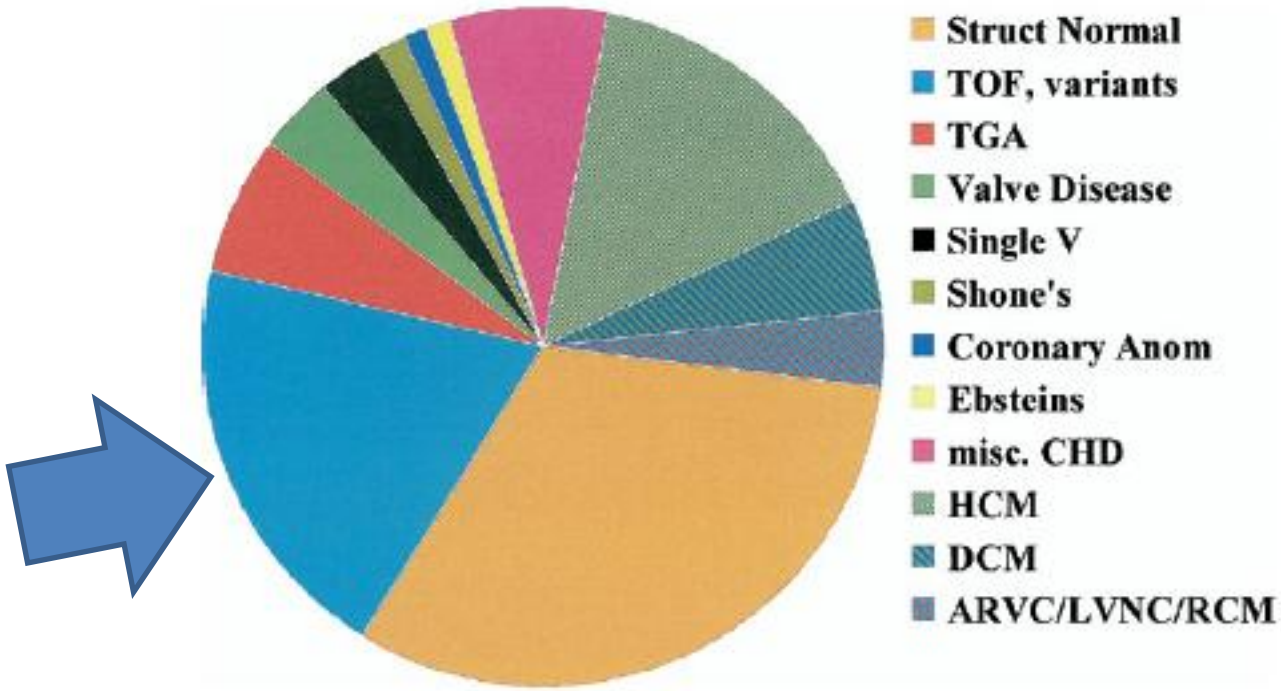


Figure 1 Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients

(J Am Coll Cardiol 2008;51:1685–91)

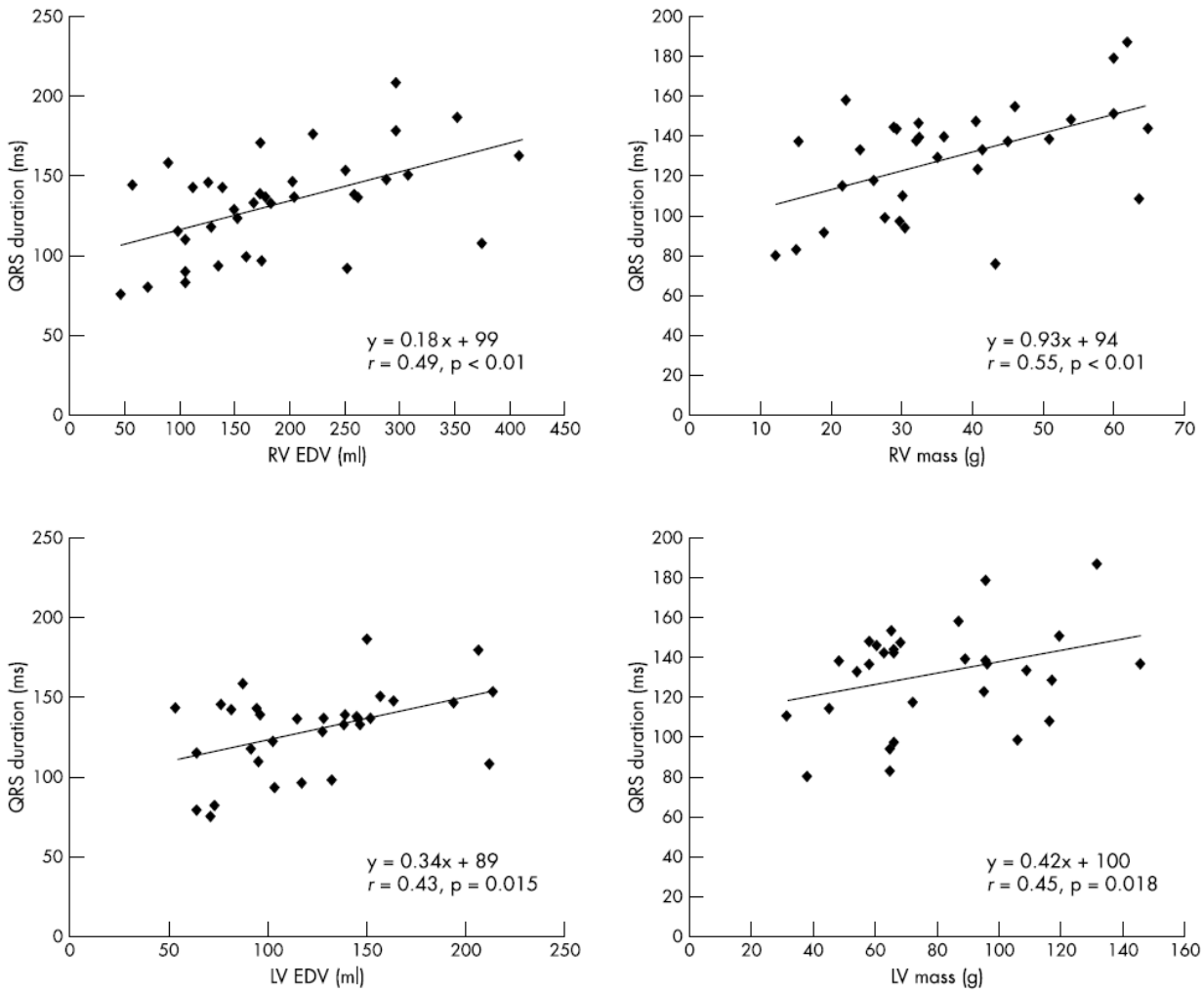
CONGENITAL HEART DISEASE

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

Heart 2002;**88**:515–520

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

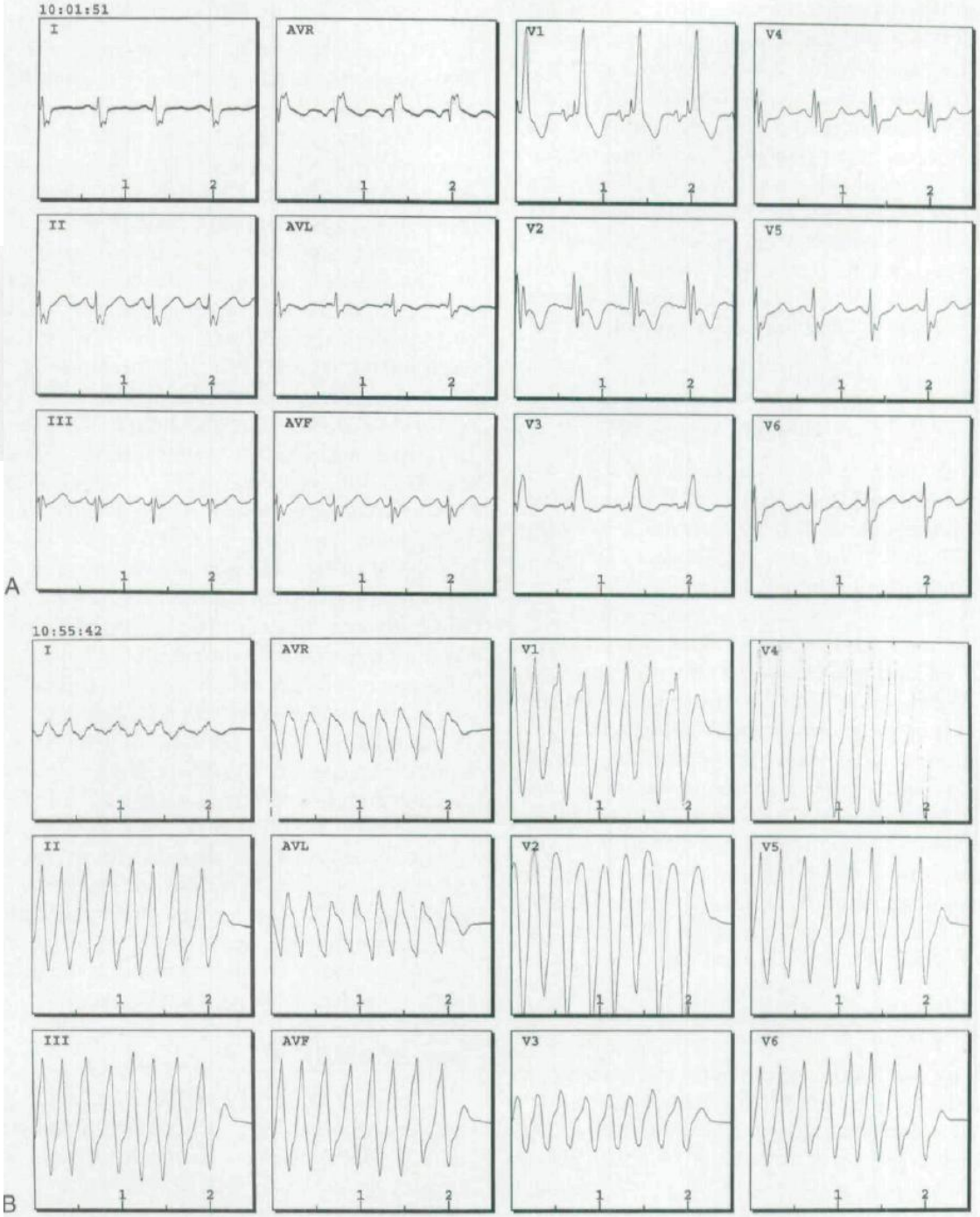
Correlação positiva entre aumento do QRS e aumento do volume diastólico final e massa ventricular



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

CHARLES I. BERUL, M.D., SHARON L. HILL, B.S.N., ROBERT L. GEGGEL, M.D., ZIYAD M. HIJAZI, M.D., GERALD R. MARX, M.D., JONATHAN RHODES, M.D., KERRI A. WALSH, R.C.V.T., and DAVID R. FULTON, M.D.

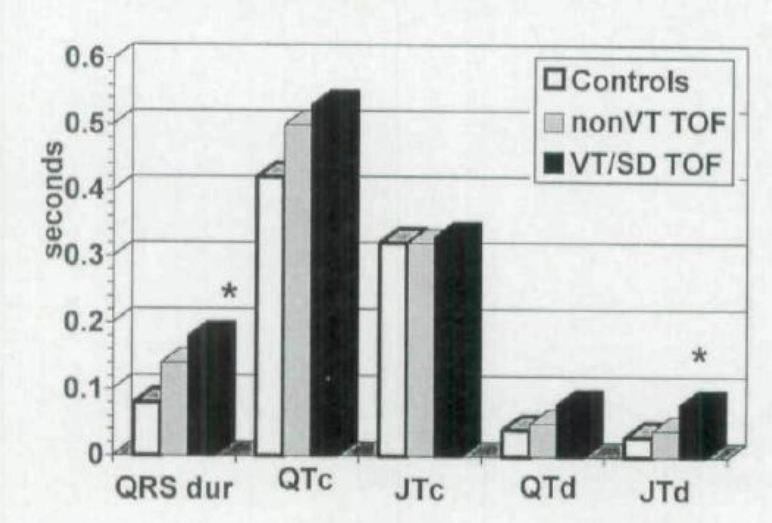
From the Division of Pediatric Cardiology, Boston Floating Hospital for Children-New England Medical Center, Tufts University School of Medicine, Boston, Massachusetts



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

CHARLES I. BERUL, M.D., SHARON L. HILL, B.S.N., ROBERT L. GEGGEL, M.D., ZIYAD M. HIJAZI, M.D., GERALD R. MARX, M.D., JONATHAN RHODES, M.D., KERRI A. WALSH, R.C.V.T., and DAVID R. FULTON, M.D.

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



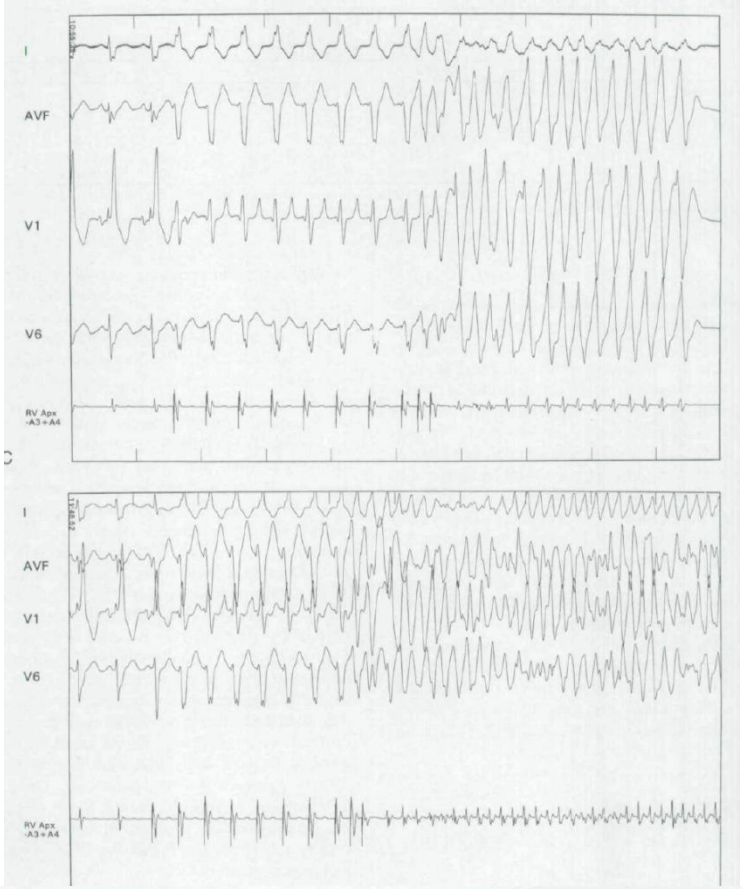


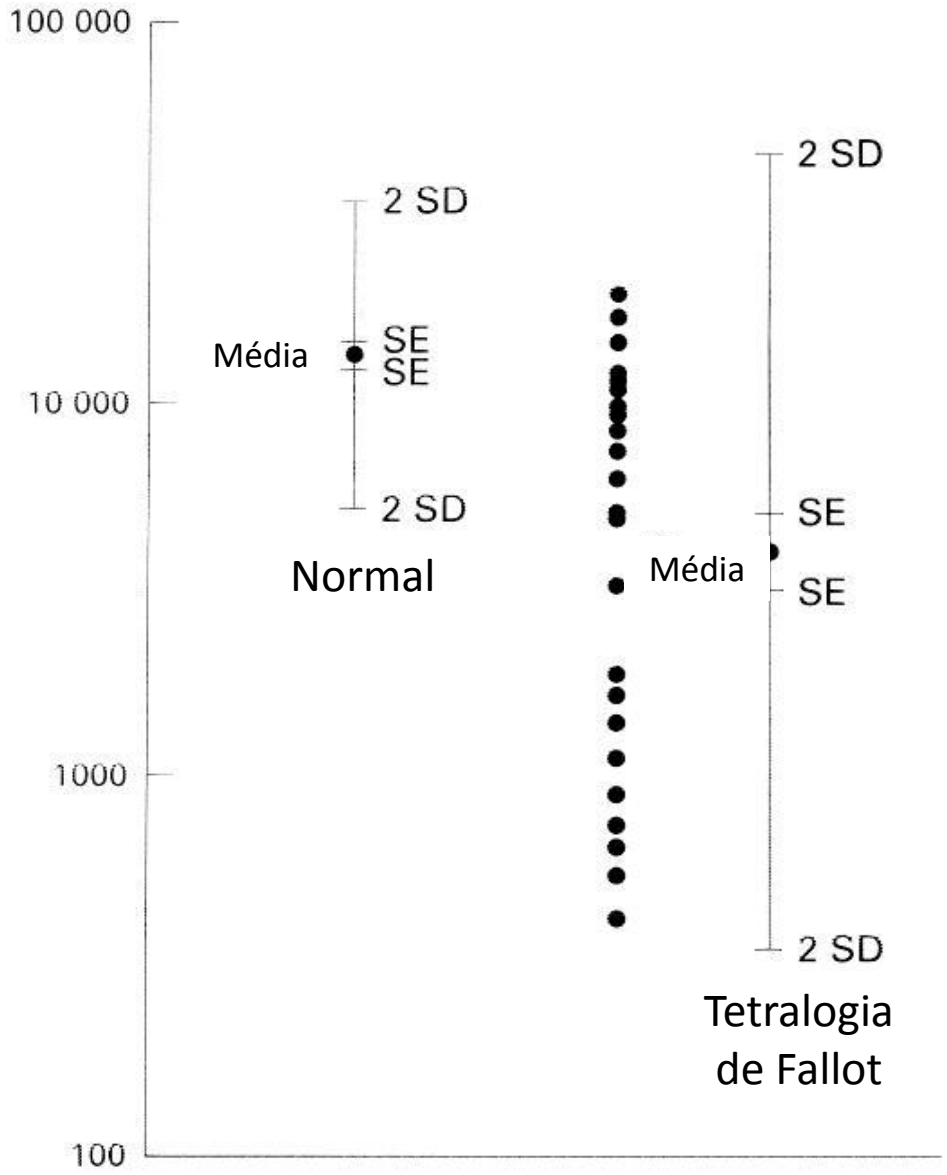
TABLE 1

Electrocardiographic Findings

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

(J Cardiovasc Electrophysiol, Vol. 8, pp. 1349-1356, December 1997)

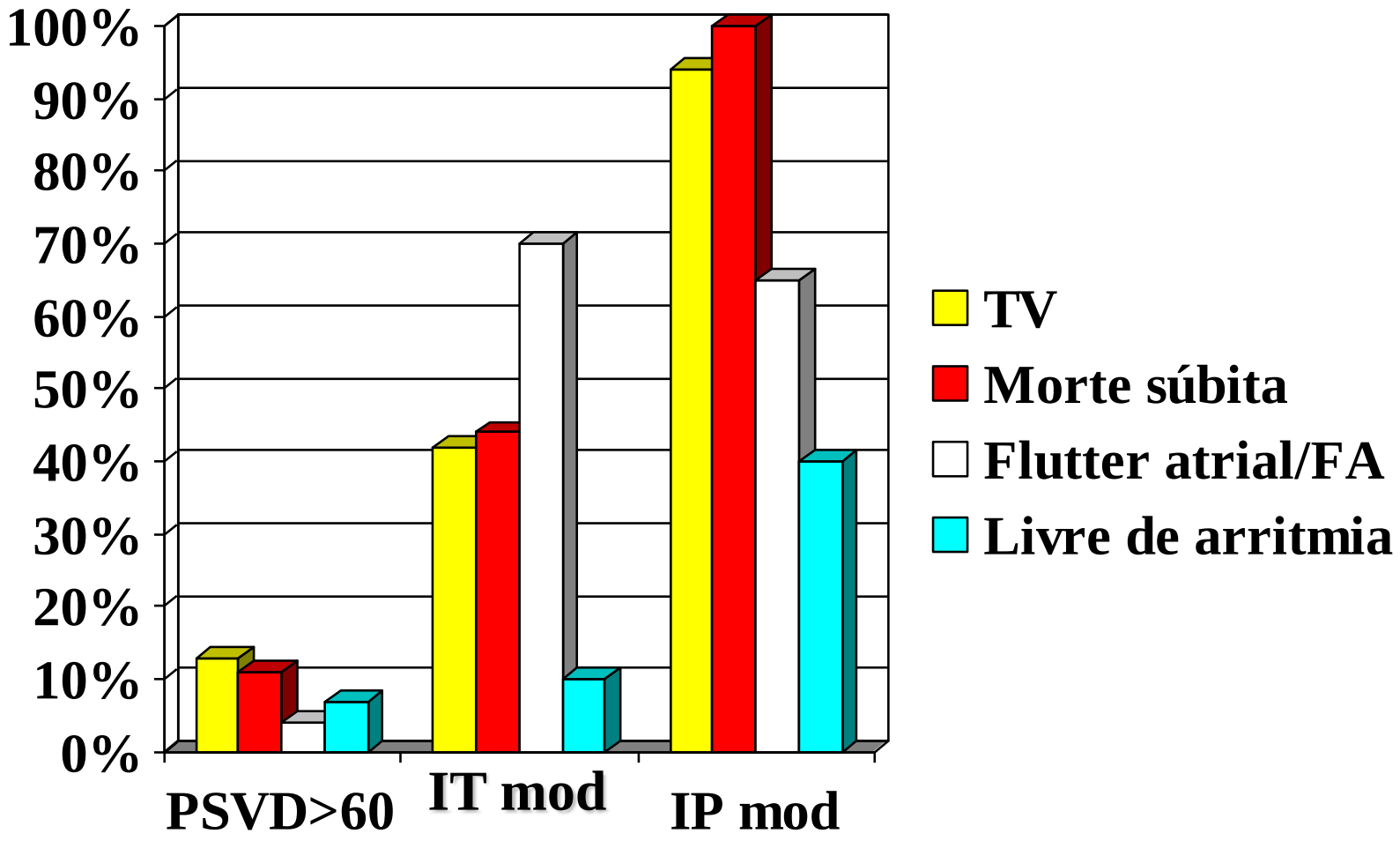
Variabilidade da FC na Tetralogia de Fallot



Reduced heart variability fallowing repair of tetralogy Fallot – Heart 1999; 81: 656-660

Arritmias no Fallot

Avaliação ecocardiográfica de 456 pacientes – pré eventos



Risk factors for arrhythmia and sudden death late after repair of tetralogy of Fallot: a multicenter studyLancet setembro 2000

ELSEVIER

International Journal of Cardiology 124 (2008) 332 – 338

International Journal of Cardiology

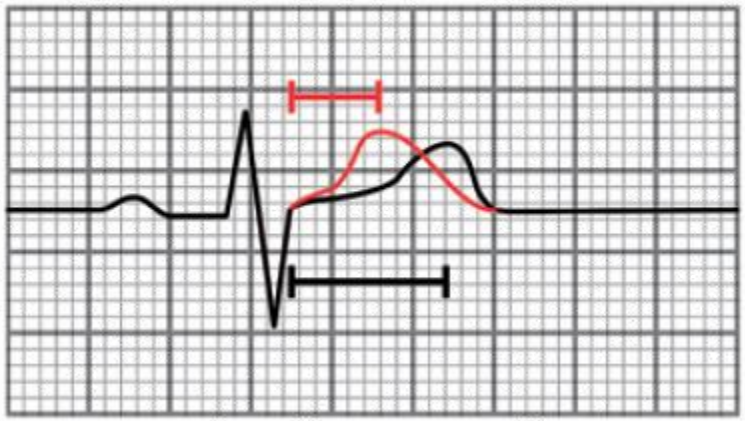
www.elsevier.com/locate/ijcard

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 [a,b], Sandra de Bie [a], Folkert J. Meijboom [c], Wim C. Hop [d], Peter M.T. Pattynama [b], Ad J.J.C. Bogers [e], Willem A. Helbing [a,b,*]

Quanto maior a IP maior a dispersão da repolarização no esforço

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



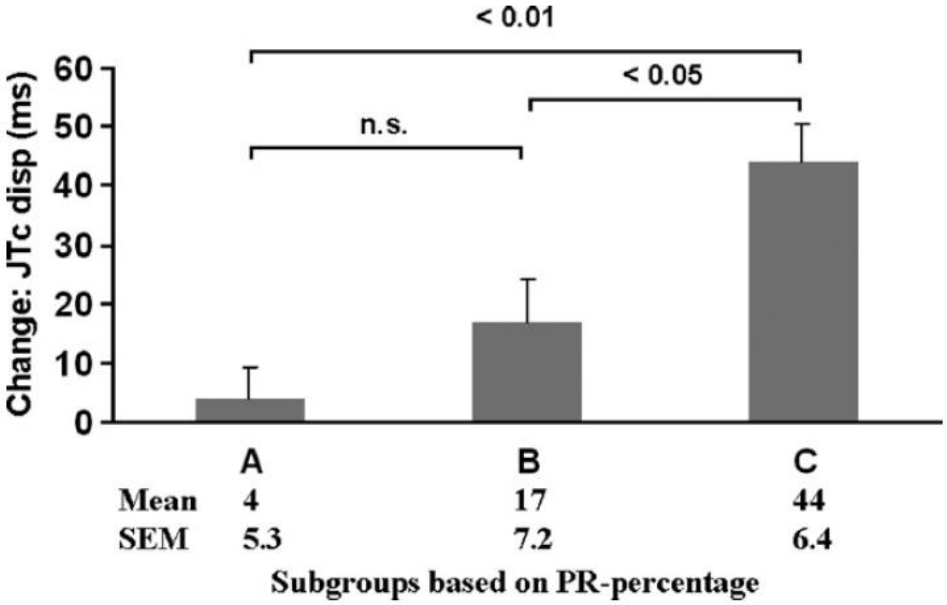


Table 3 Predictors of appropriate and inappropriate therapies in adults with CHD ($n = 64$)

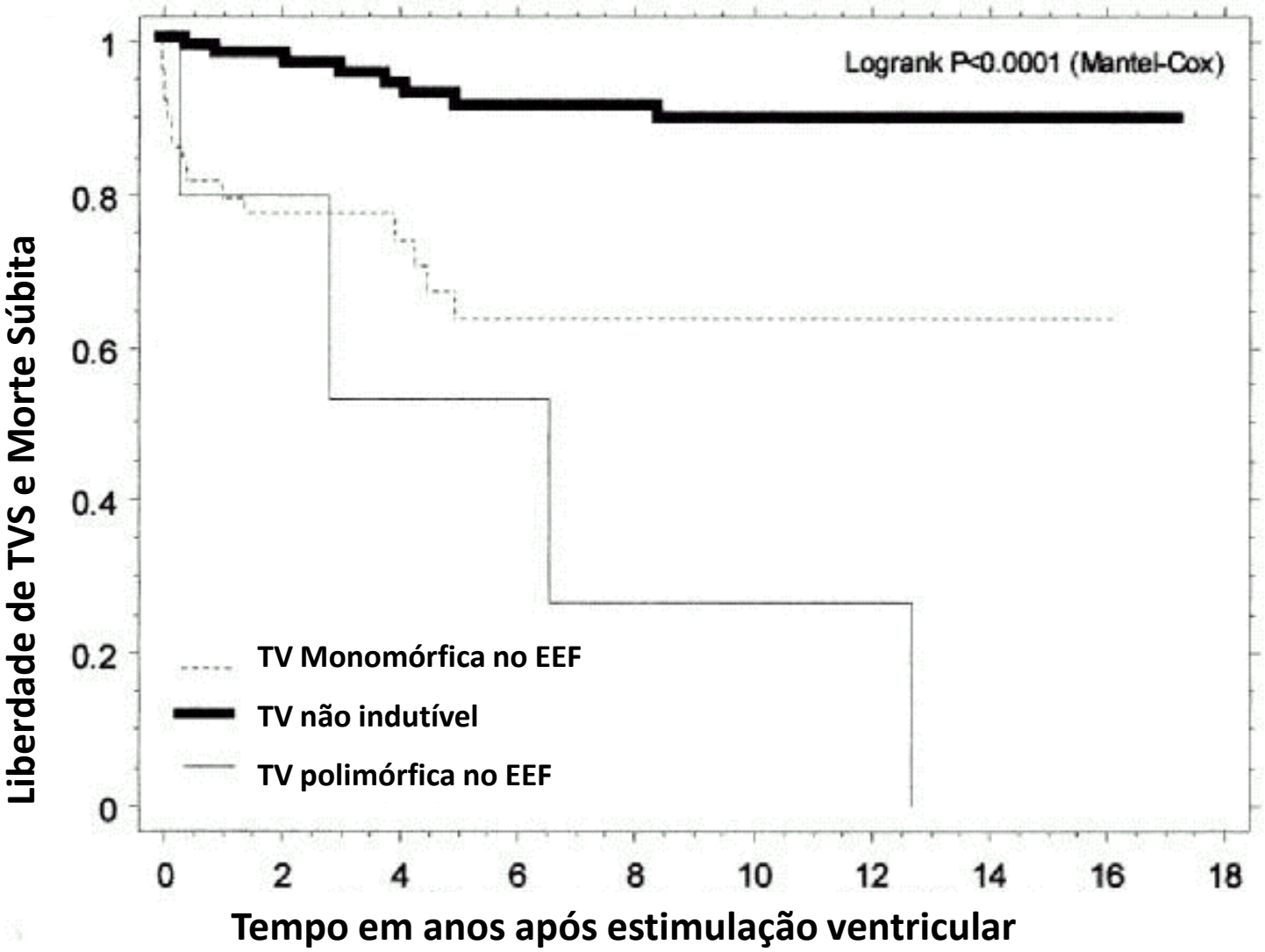
Variable	Appropriate shock therapy		Inappropriate shock therapy	
	HR (95% CI)	*P*-value	HR (95% CI)	*P*-value
Age at ICD implantation	0.96 (0.92–1.01)	0.12	0.98 (0.95–1.02)	0.29
Male gender	1.09 (0.37–3.20)	0.88	1.49 (0.62–3.57)	0.38
BMI >25	0.43 (0.14–1.34)	0.14	1.23 (0.55–2.75)	0.61
QRS width >180 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[a]	0.47 (0.16–1.38)	0.17	1.75 (0.73–4.21)	0.21
Positive PES[b]	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

European Heart Journal (2007) **28**, 1854–1861
doi:10.1093/eurheartj/ehl306

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

Variable	Appropriate shock therapy		Inappropriate shock therapy	
	HR (95% CI)	*P*-value	HR (95% CI)	*P*-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[a]	0.47 (0.16–1.38)	0.17	1.75 (0.73–4.21)	0.21
Positive PES[b]	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

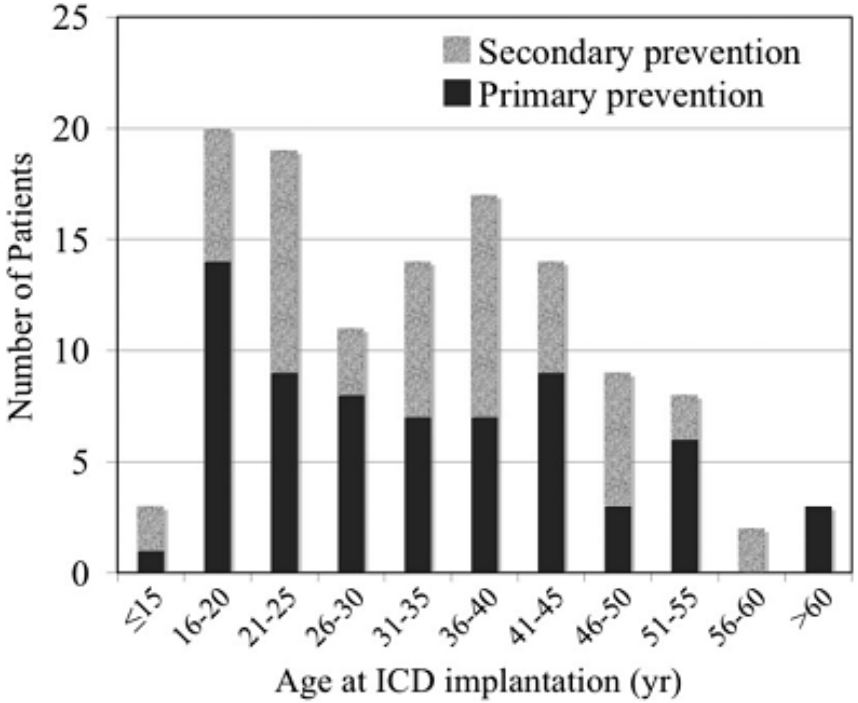
European Heart Journal (2007) **28**, 1854–1861
doi:10.1093/eurheartj/ehl306

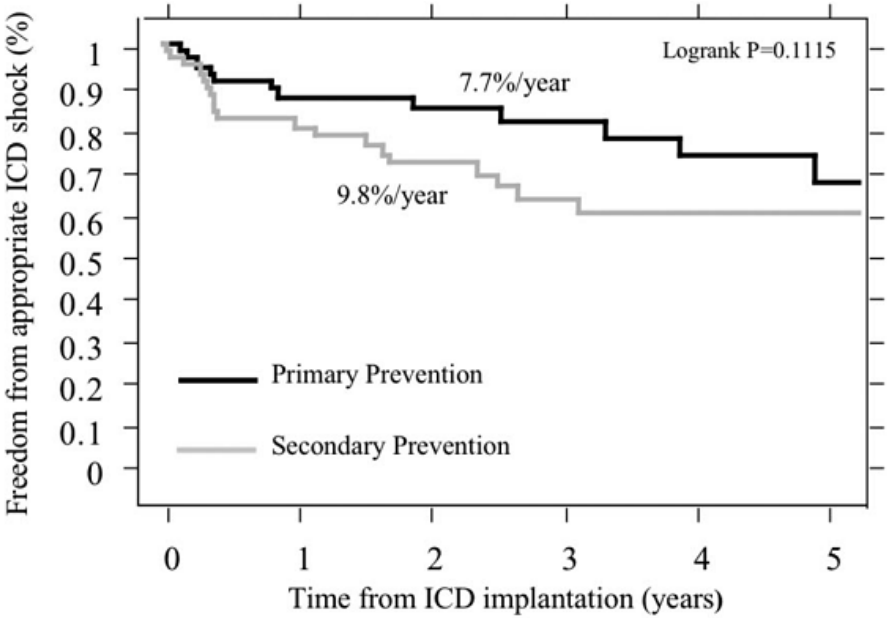


Value of Programmed Ventricular Stimulation After Tetralogy of Fallot Repair-
Circulation, 2004; 109: 1994-2000

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;
Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD;
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

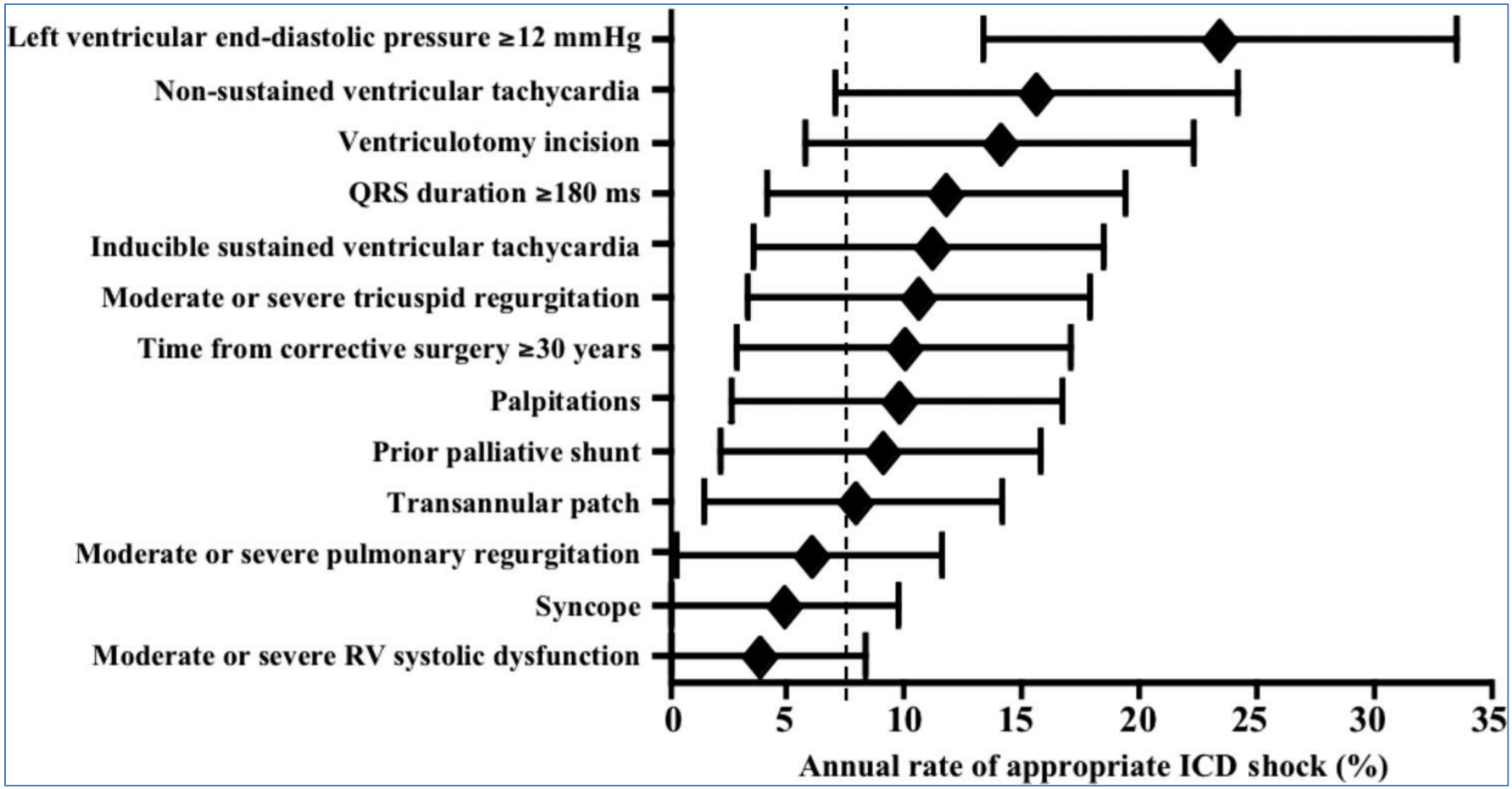




Circulation. 2008;117:363-370

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;
Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD;
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



Circulation. 2008;117:363-370

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD; Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD; 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

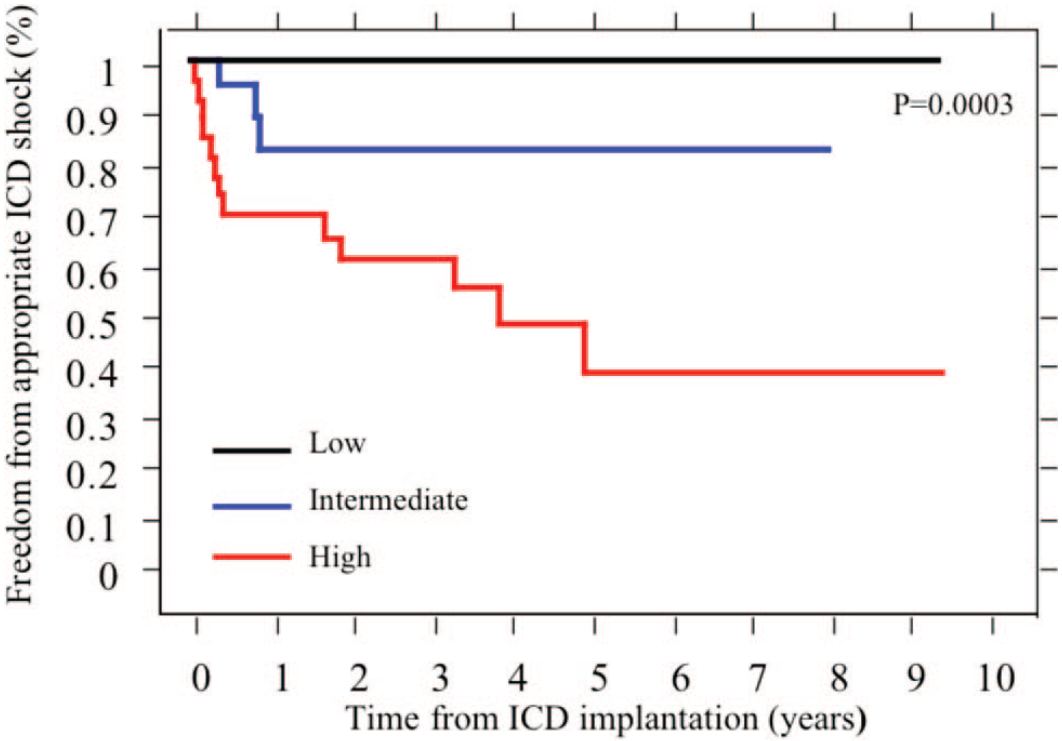
Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention

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

Circulation. 2008;117:363-370

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;
Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD;
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%

Circulation. 2008;117:363-370

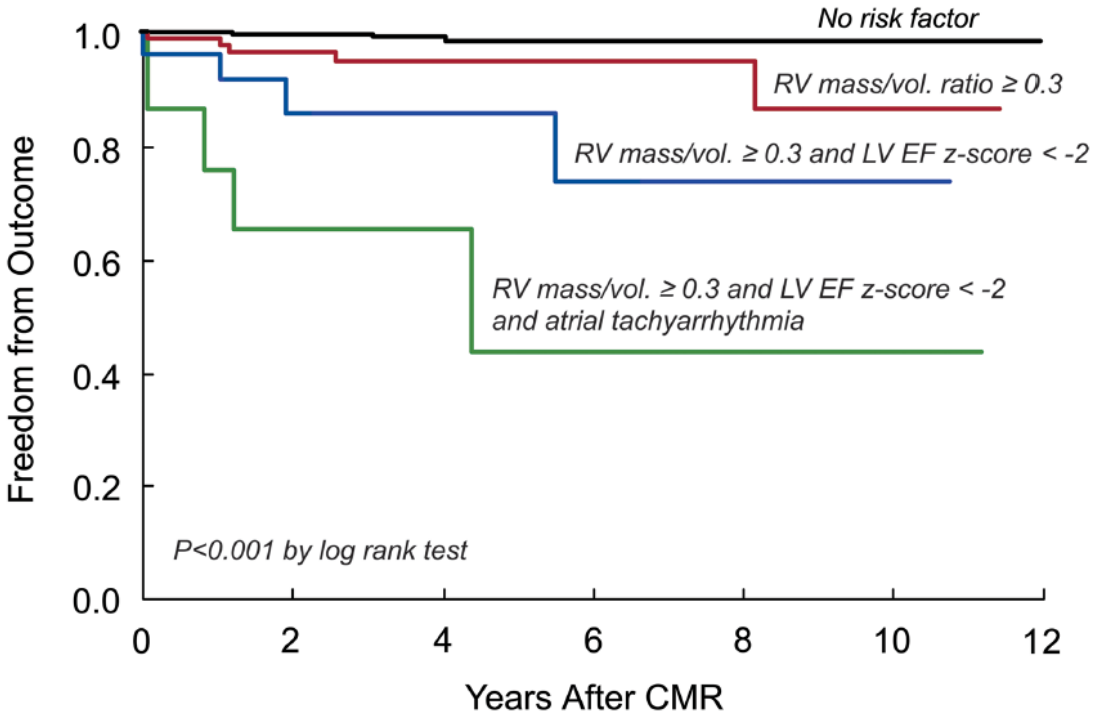
ORIGINAL ARTICLE

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

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

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

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

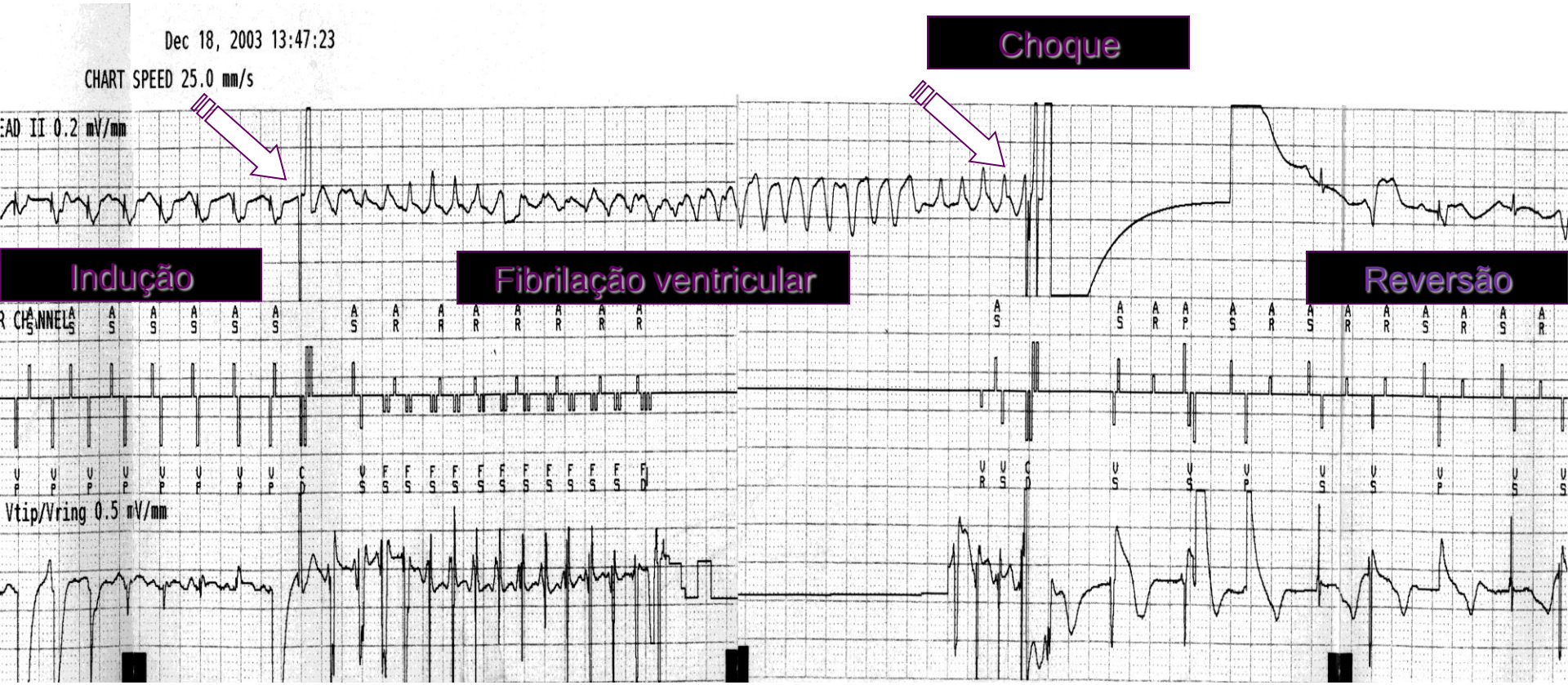


Mensagens

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

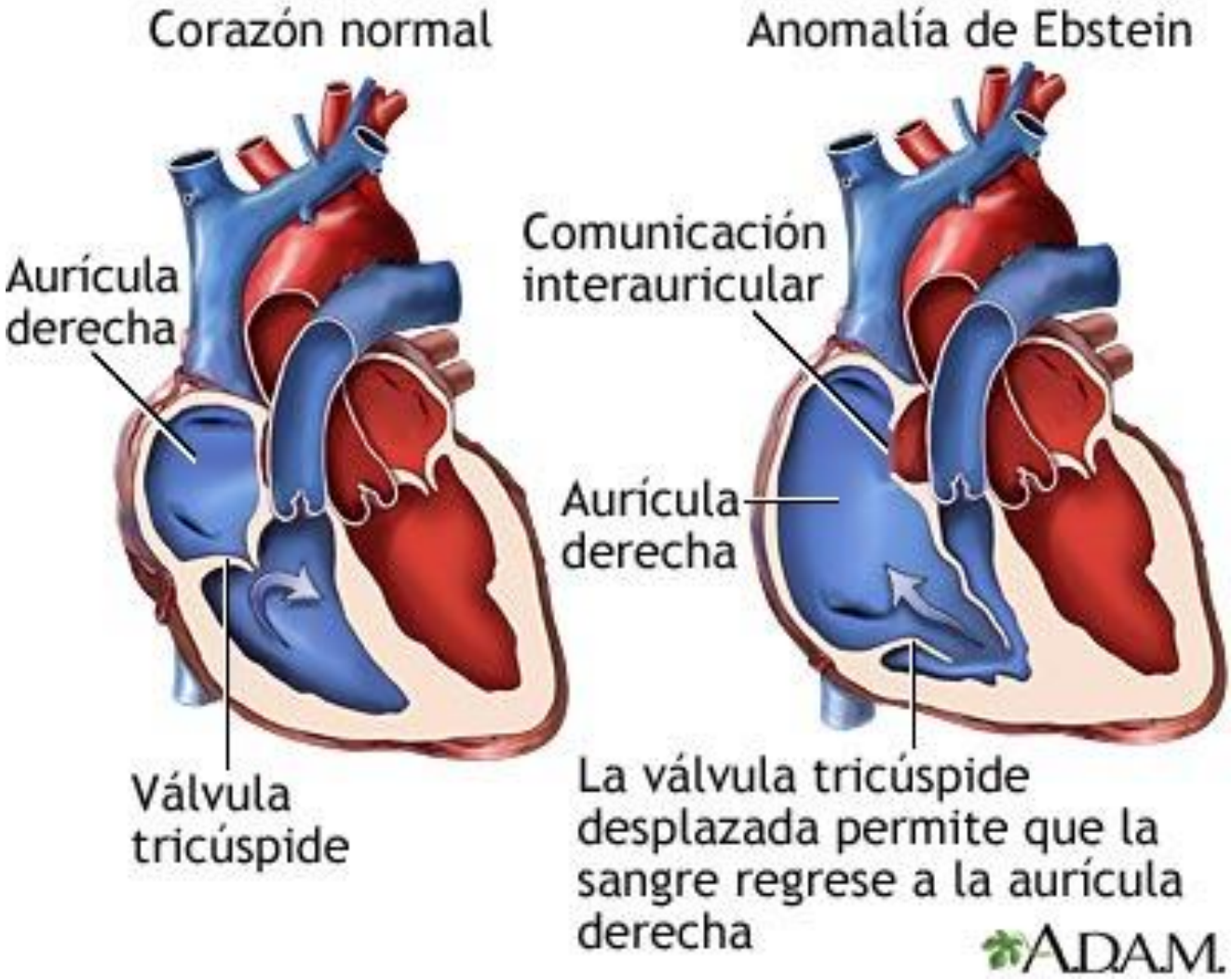
Evolução

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
EAD II 0.2 mV/mm
Choque
Indução
Fibrilação ventricular
Reversão
R CHANNEL
Vtip/Vring 0.5 mV/mm

Marcadores ECG na população pediátrica e em jovens

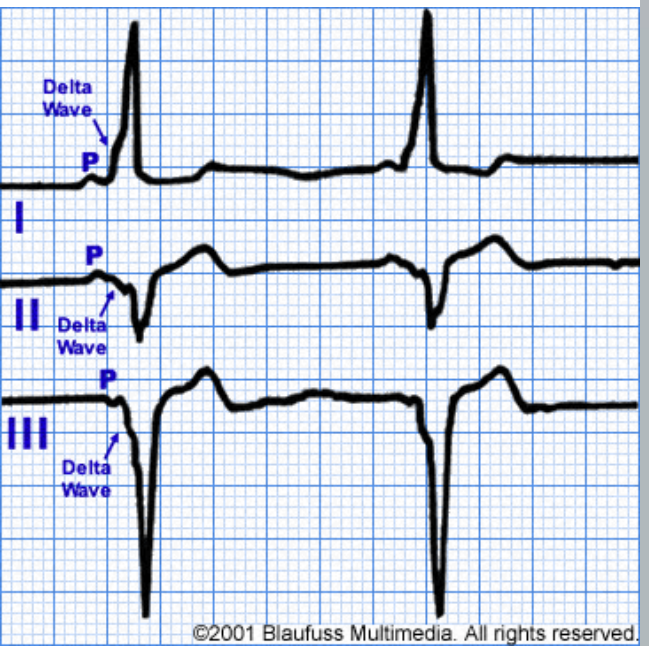
Marcadores ECG na população pediátrica e em jovens

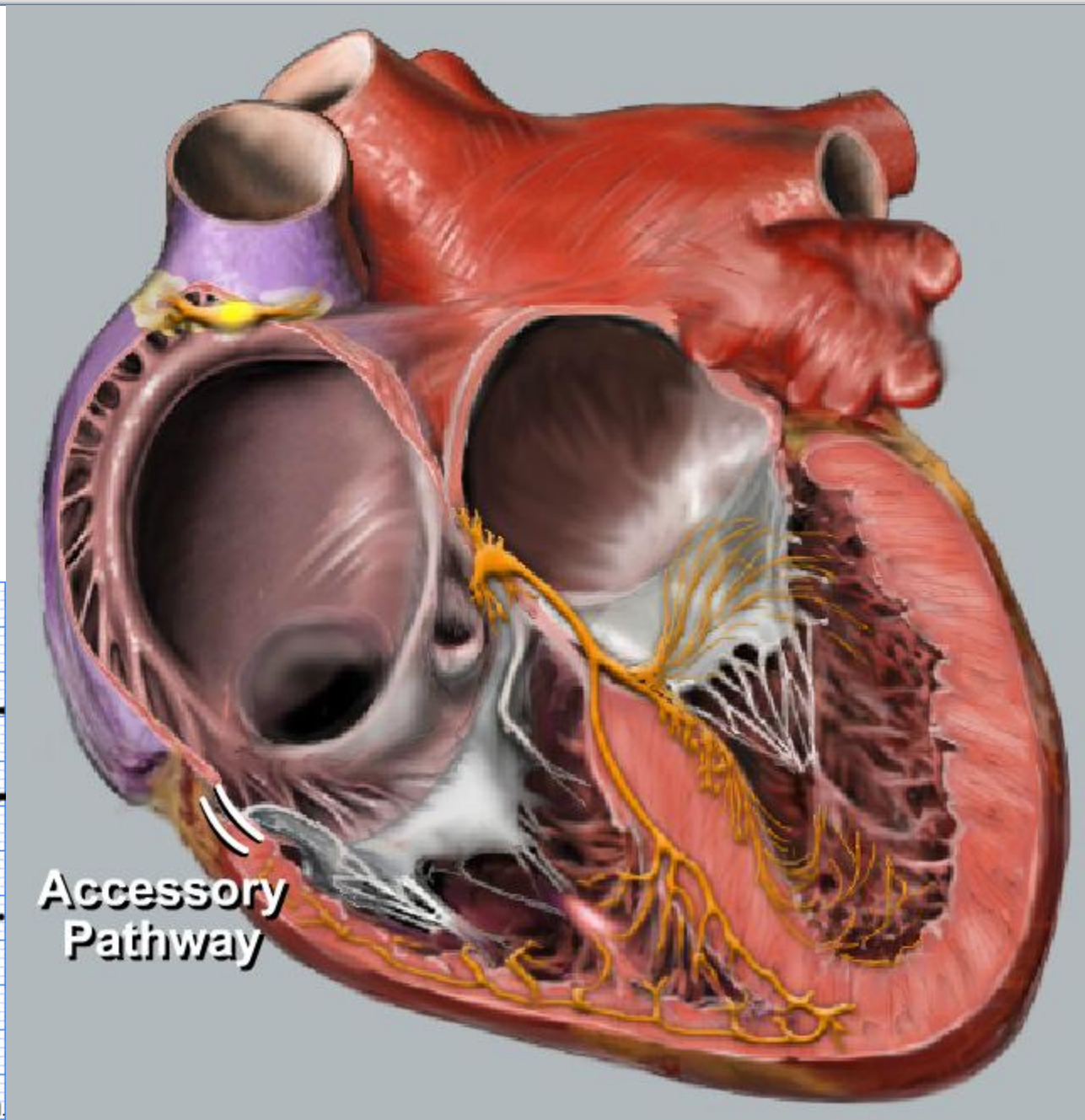

Corazón normal
Anomalía de Ebstein
Aurícula derecha
Válvula tricúspide
Comunicación interauricular
Aurícula derecha
La válvula tricúspide desplazada permite que la sangre regrese a la aurícula derecha
A.D.A.M.

Pré excitação ventricular

Padrões do ECG

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

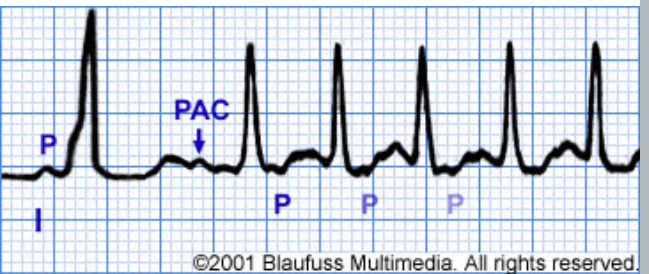


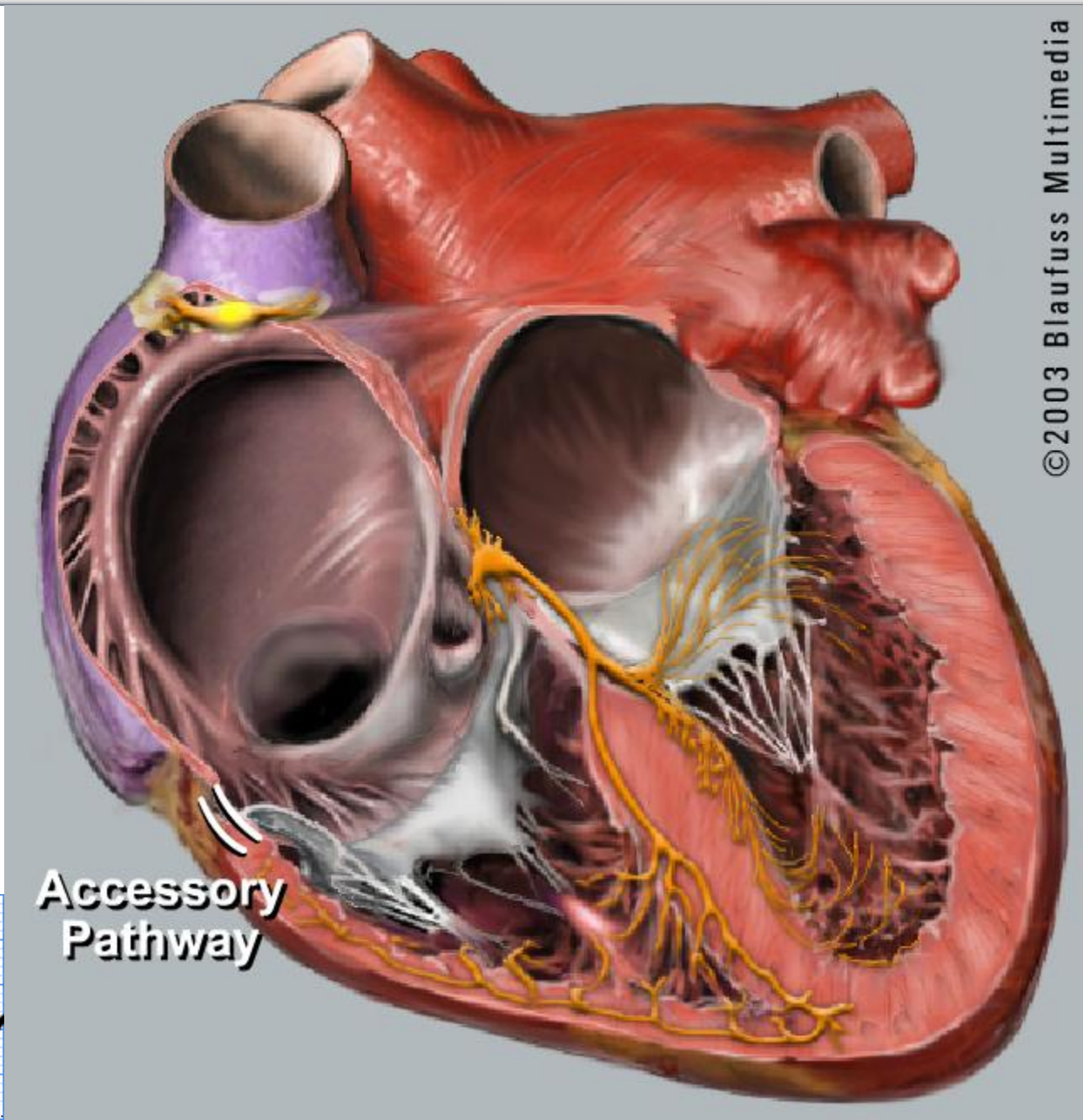


Início da taquicardia no WPW

Taquicardia por reentrada

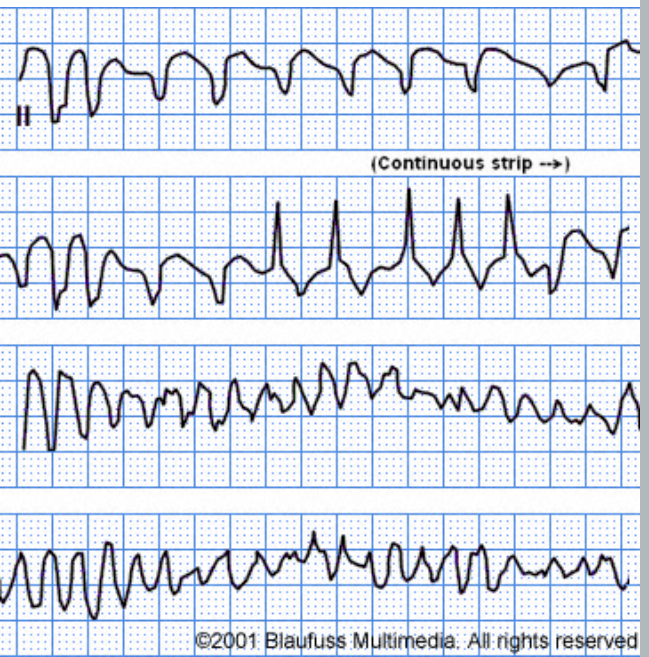
- Pode ser iniciado por
- Bloqueio da via acessória
- Condução pelo NAV
- Condução retrógrada pela via acessória
- Onda P retrógrada visível

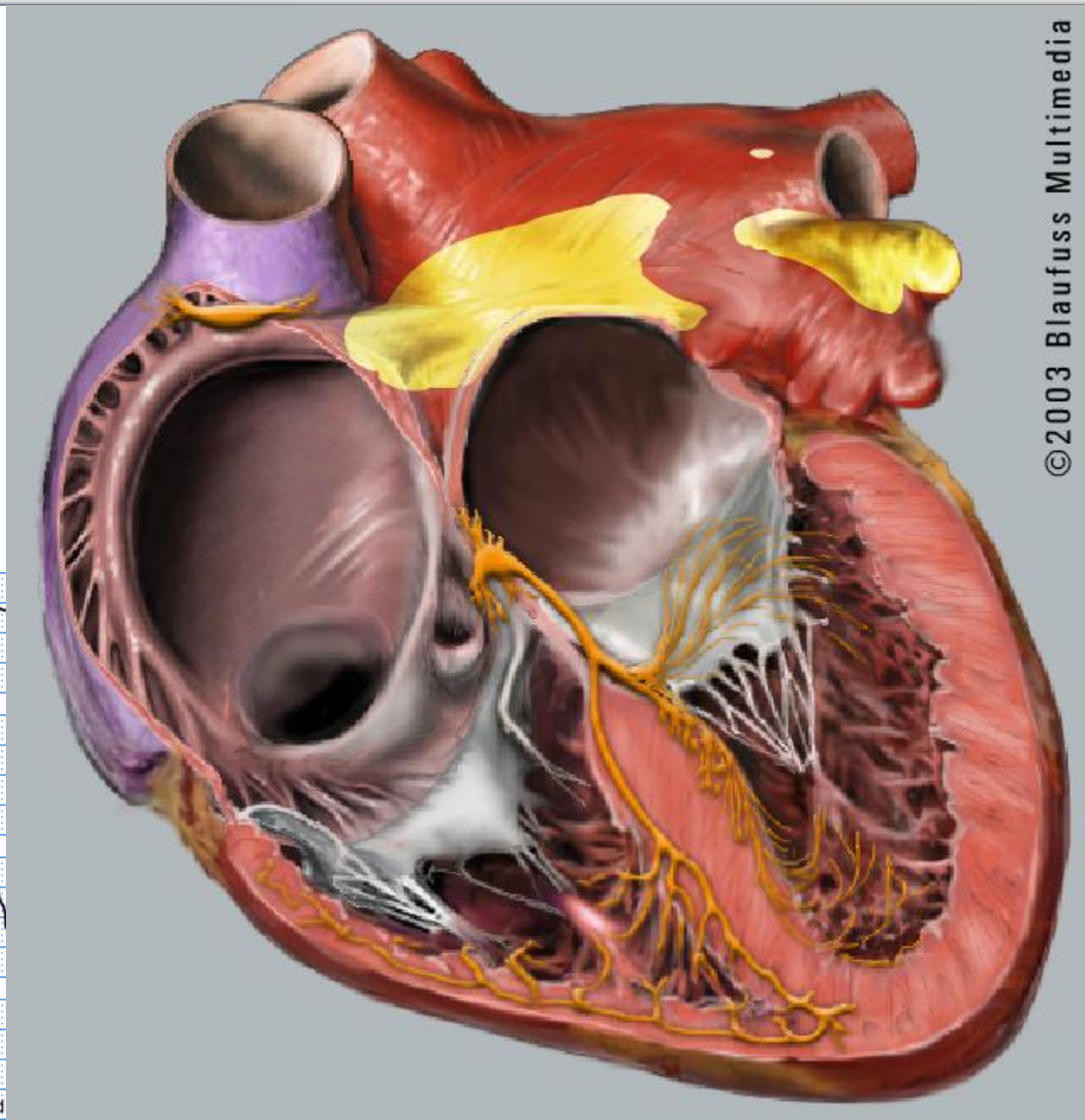




FA + pré excitação

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

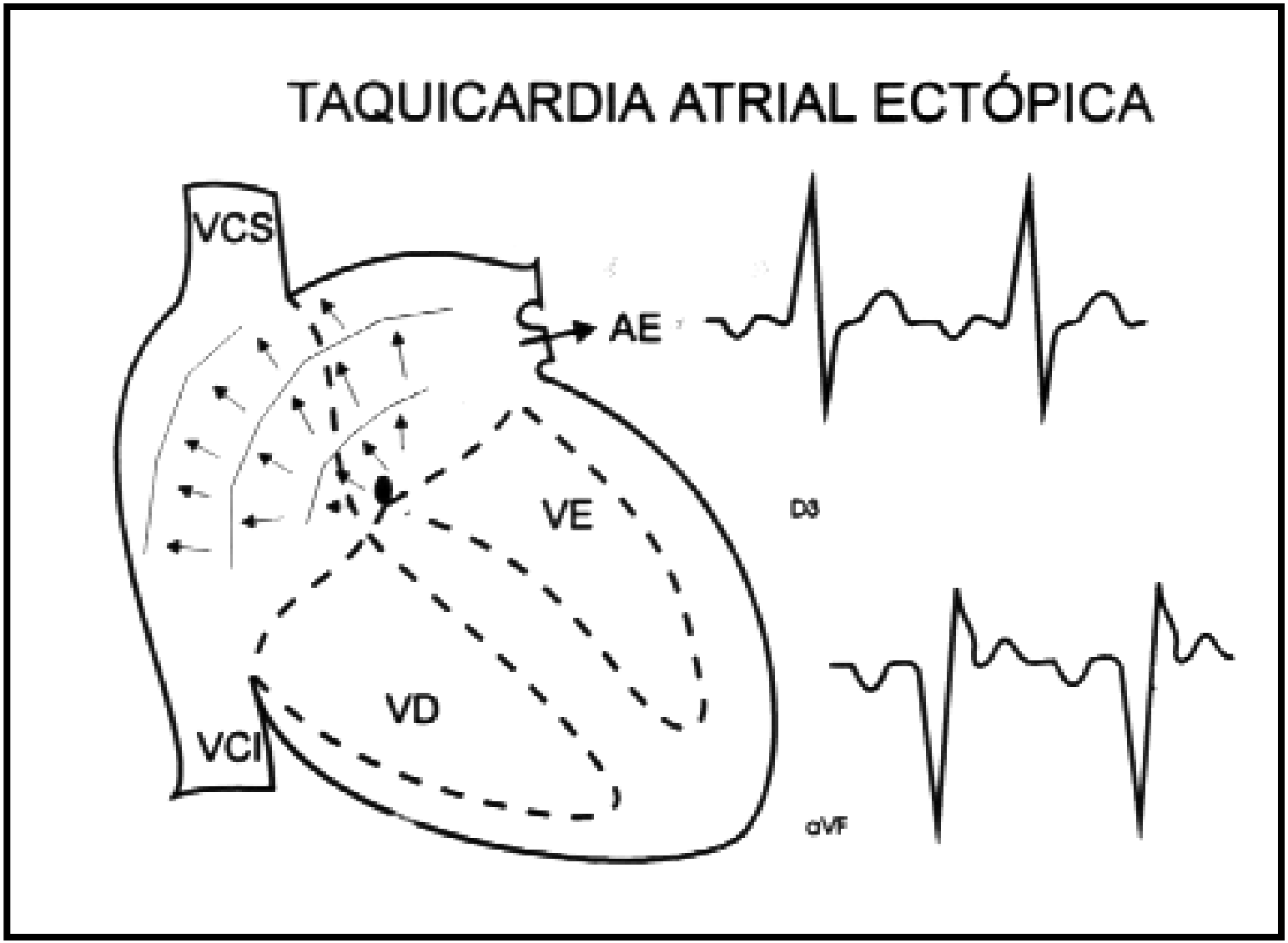


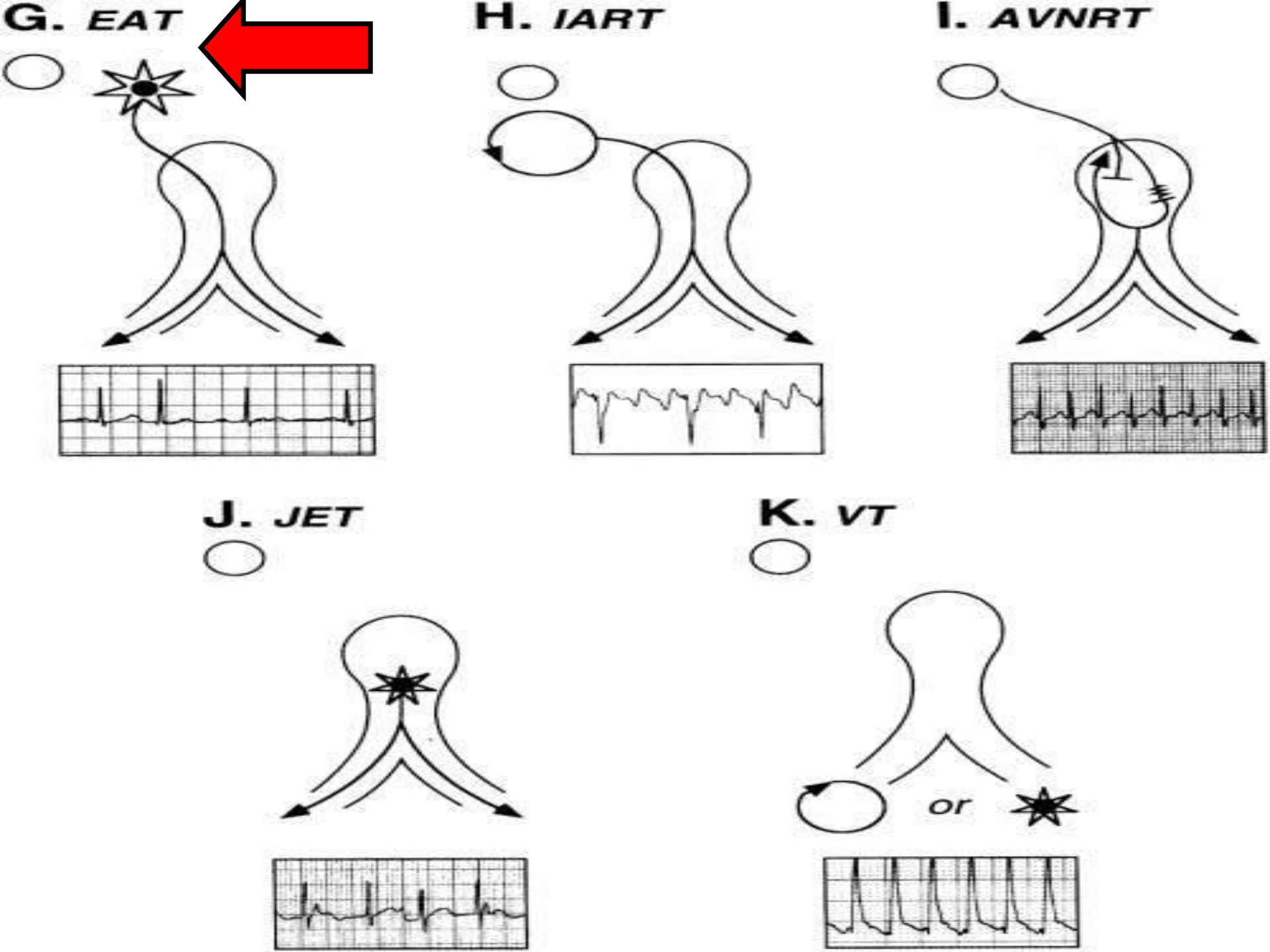


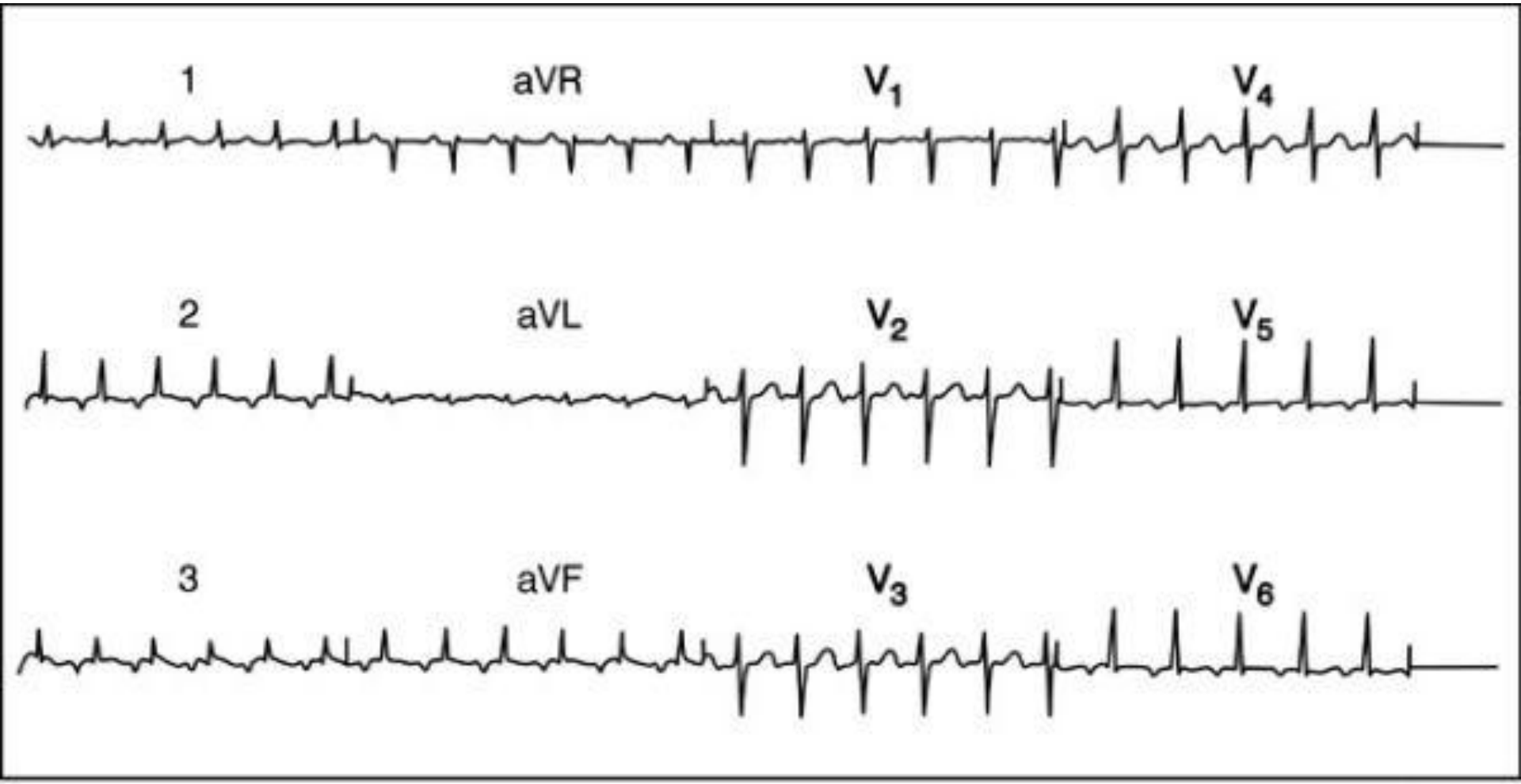
Taquicardias automatismo

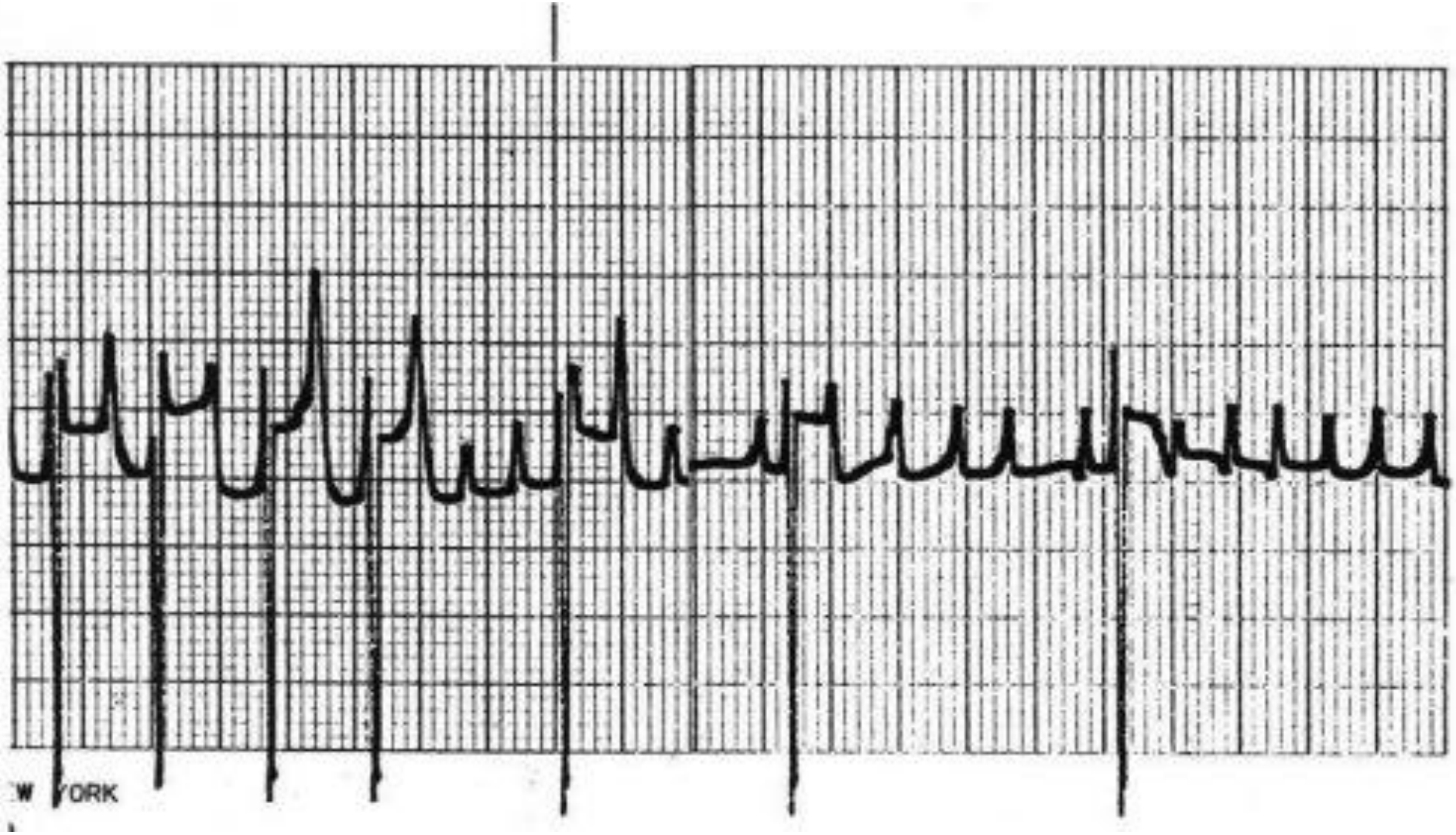
- **Taquicardia sinusal**
- ***Taquicardia atrial***
- **Taquicardia juncional**

Taquicardia Atrial

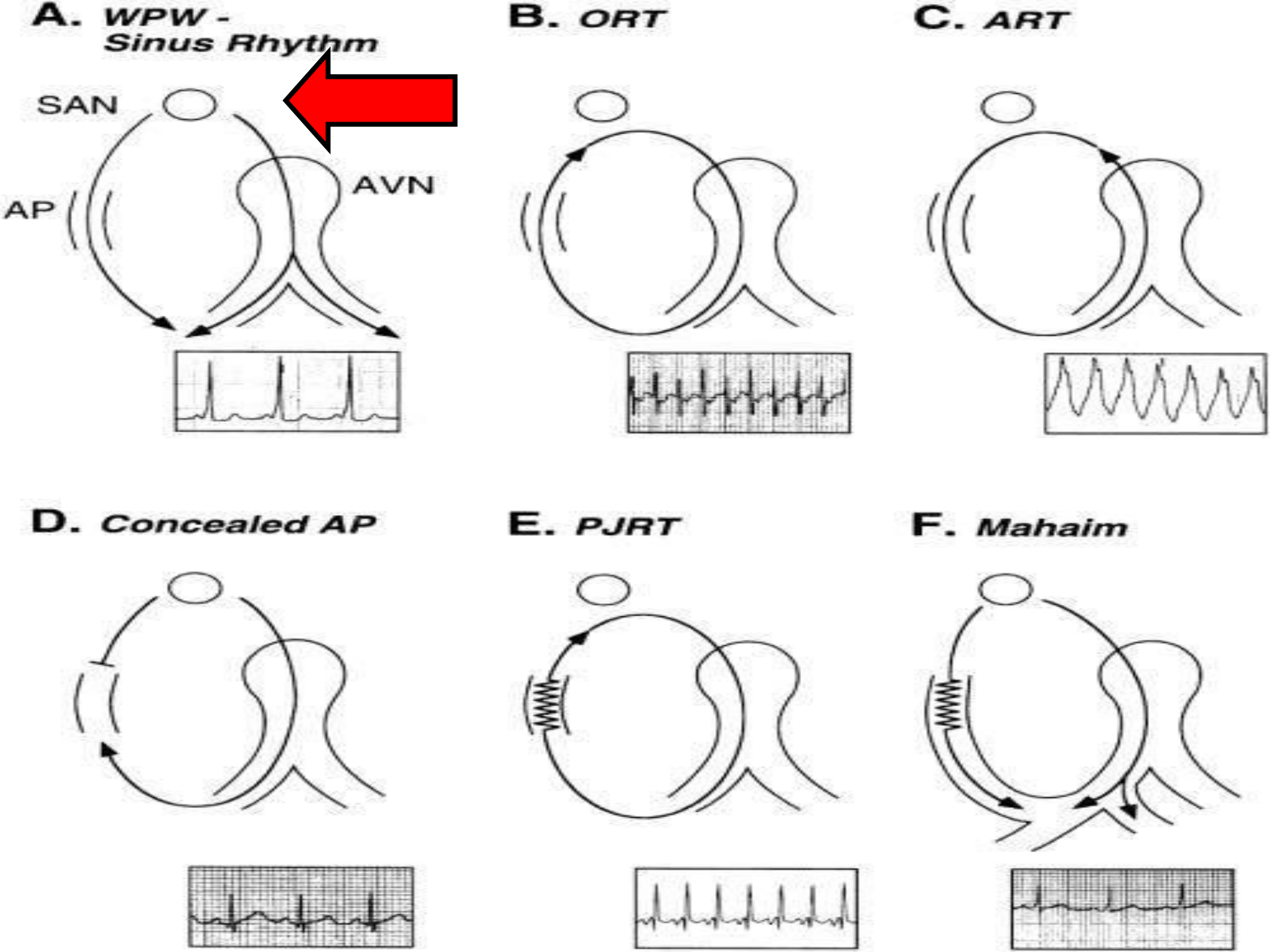



G. EAT
H. IART
I. AVNRT
J. JET
K. VT
or


1
aVR
V1
V4
2
aVL
V2
V5
3
aVF
V3
V6


W YORK

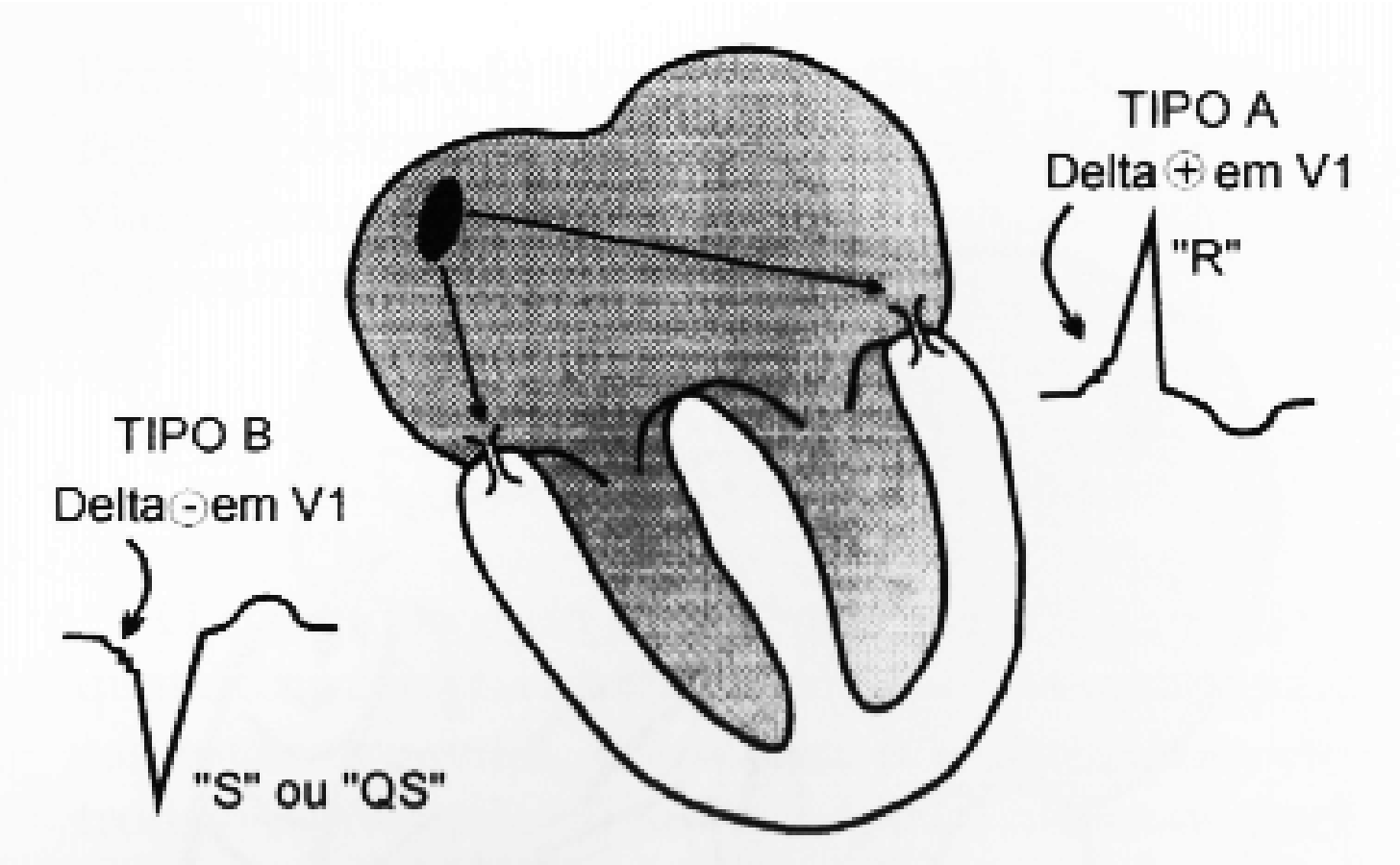
Pré excitação + taquicardia = WPW


A. WPW - Sinus Rhythm
SAN
AP
AVN
B. ORT
C. ART
D. Concealed AP
E. PJRT
F. Mahaim

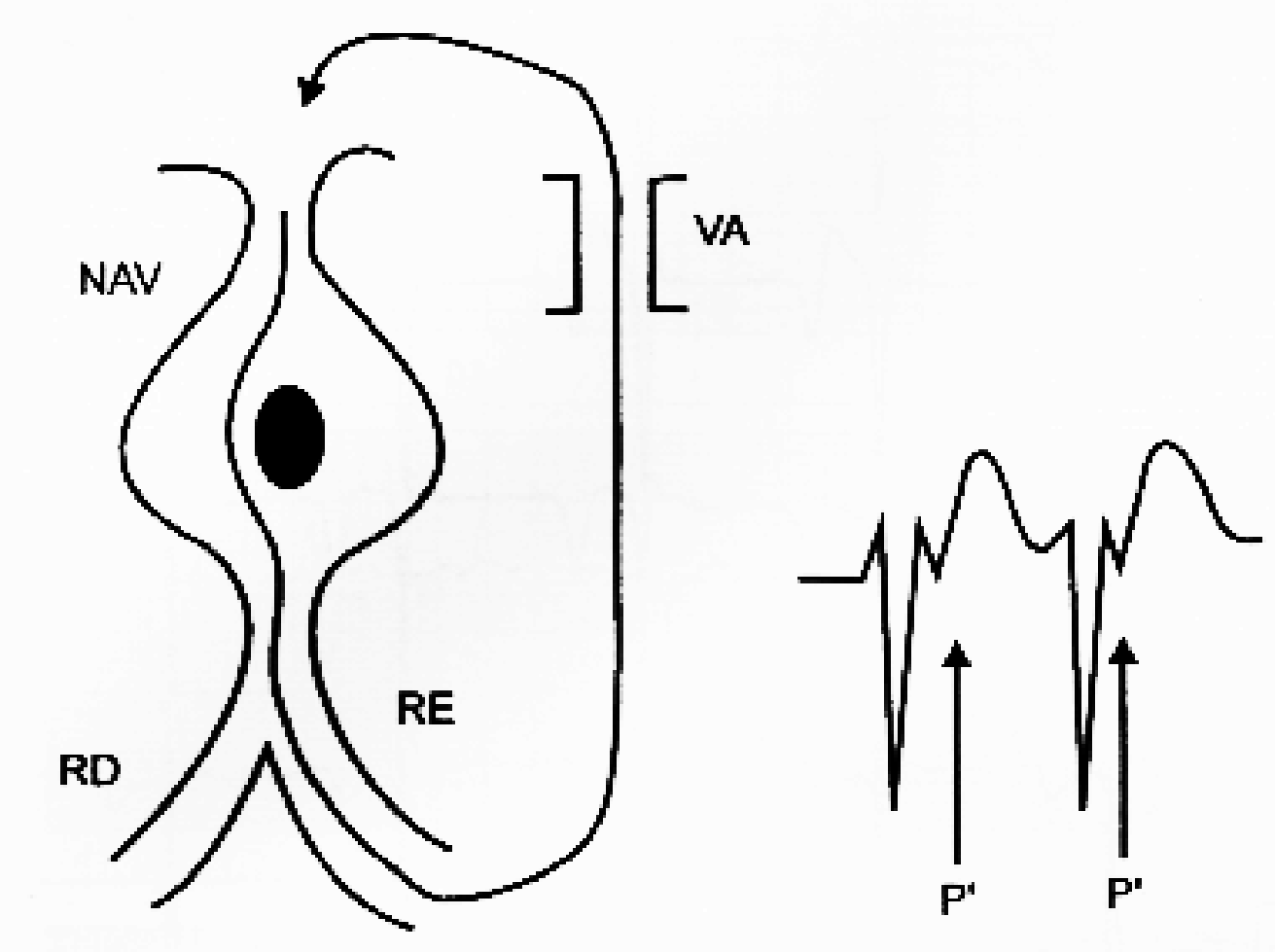
Anomalia Ebstein e WPW

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

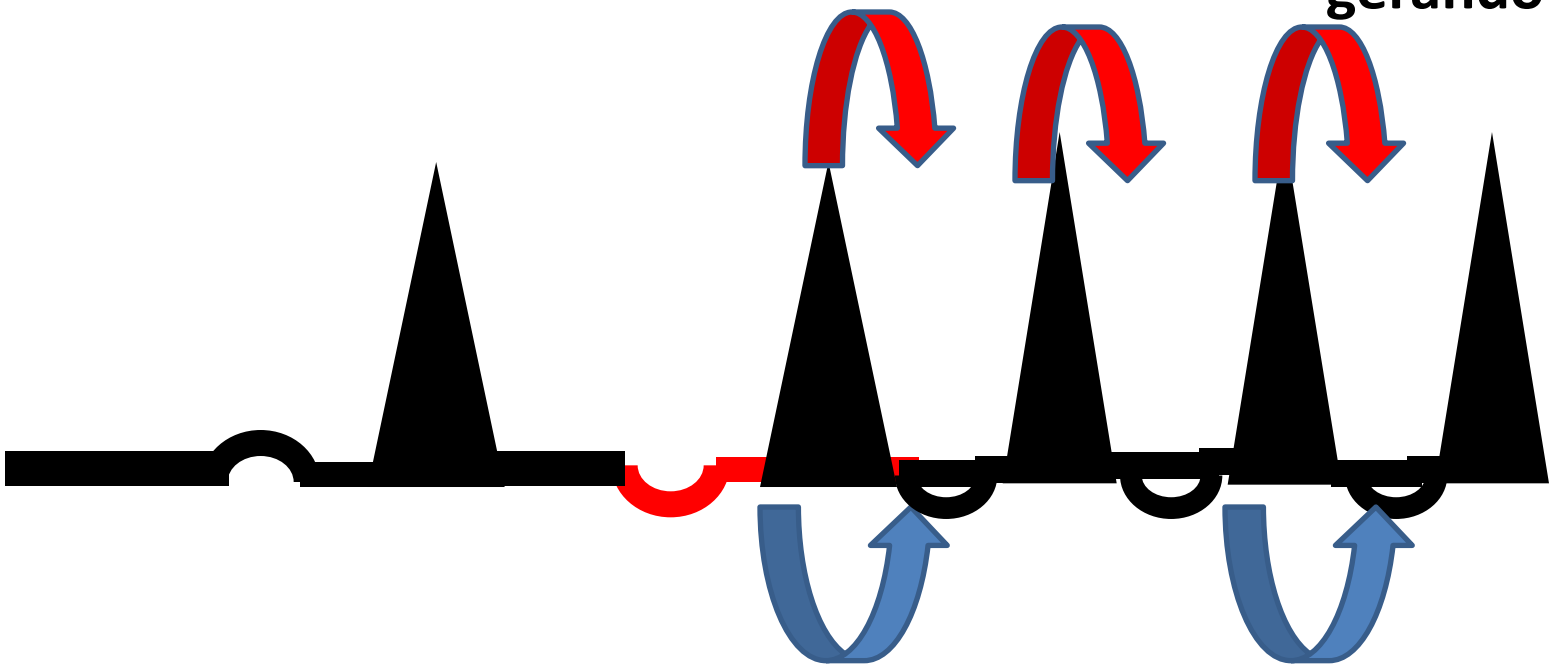
Pré-Excitação Ventricular
Classificação Eletrocardiográfica



Pré-Excitação Ventricular
TPSV Ortodrômica



Moreira DAR, Arritmias Cardíacas: Clínica, Diagnóstico e Terapêutica, 1995


Complexos QRS
gerando P retrógrada
Início QRS início p
retrógrada maior
que 70 ms

ECGPC - Gravação : 033

t= 110

D1

D2

D3

aVF

V1

V5

V6

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)

Iniciar ECGPC Microsoft PowerPoint ... imagem - Paint PT 18:16

ECGPC - Gravação : 033

t= 110

D1

D2

D3

aVF

V1

V5

V6

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

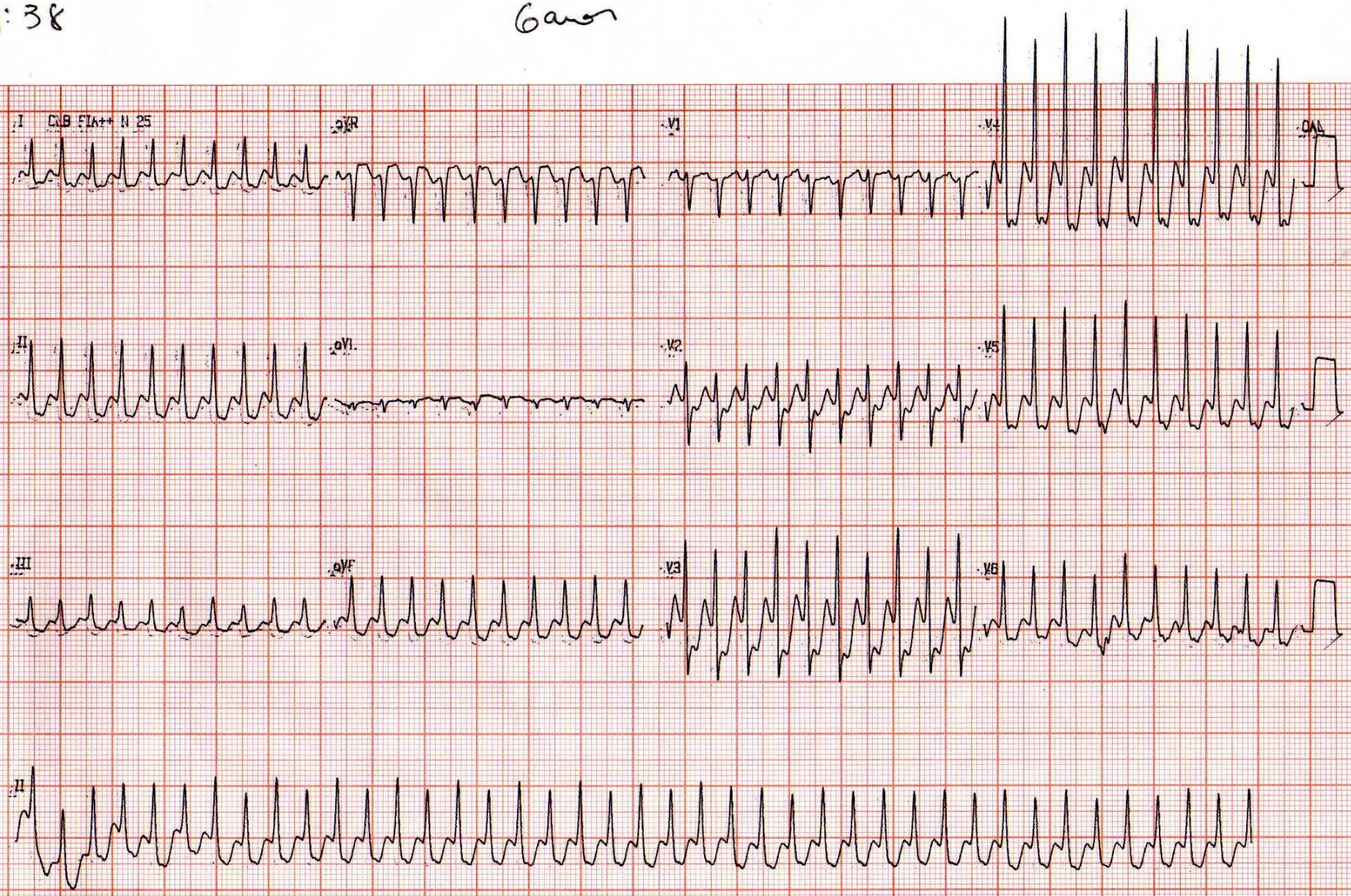
Voltar

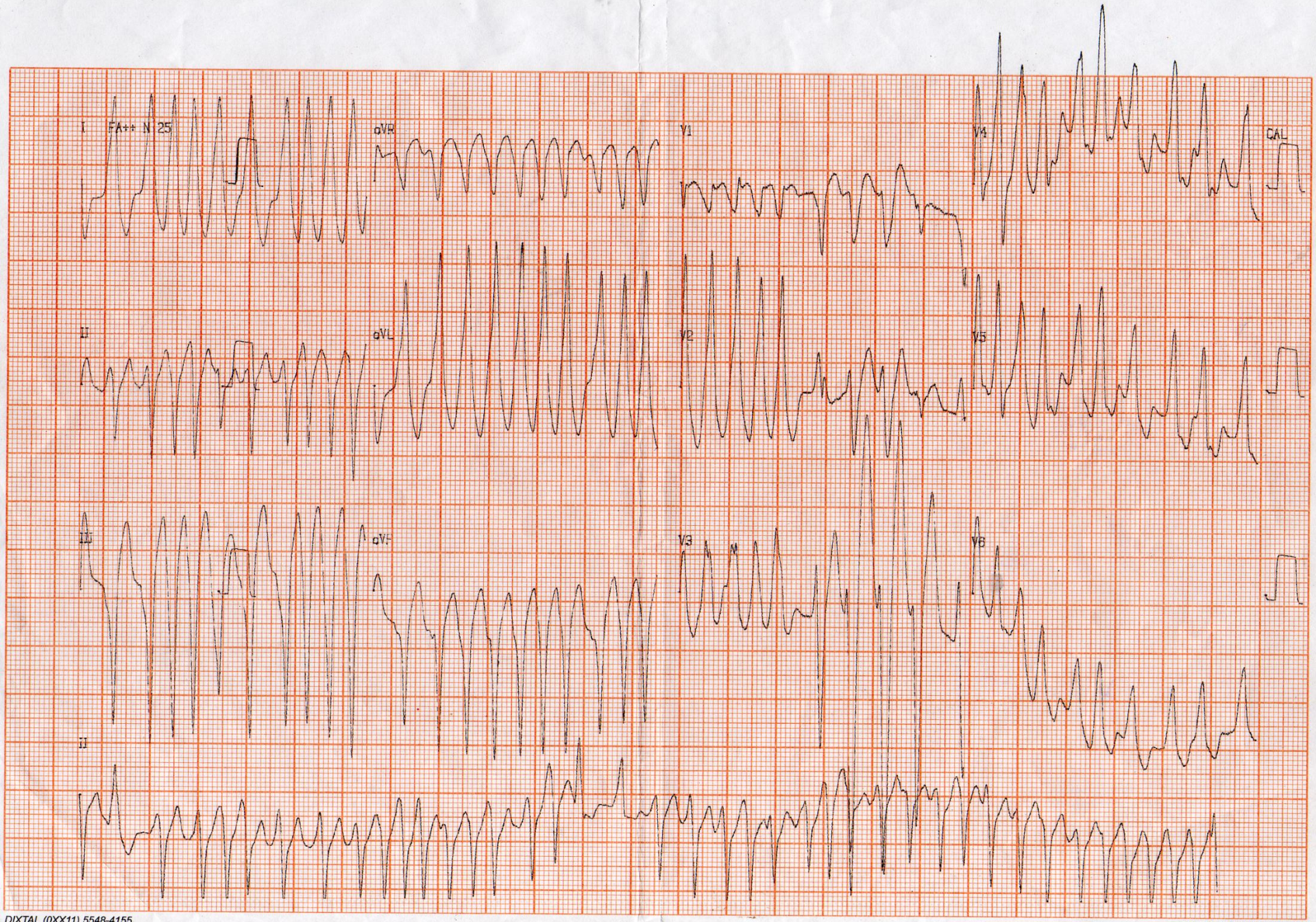
Português (Brasil)

Iniciar ECGPC Microsoft PowerPoint ... imagem - Paint PT 18:16

14:38

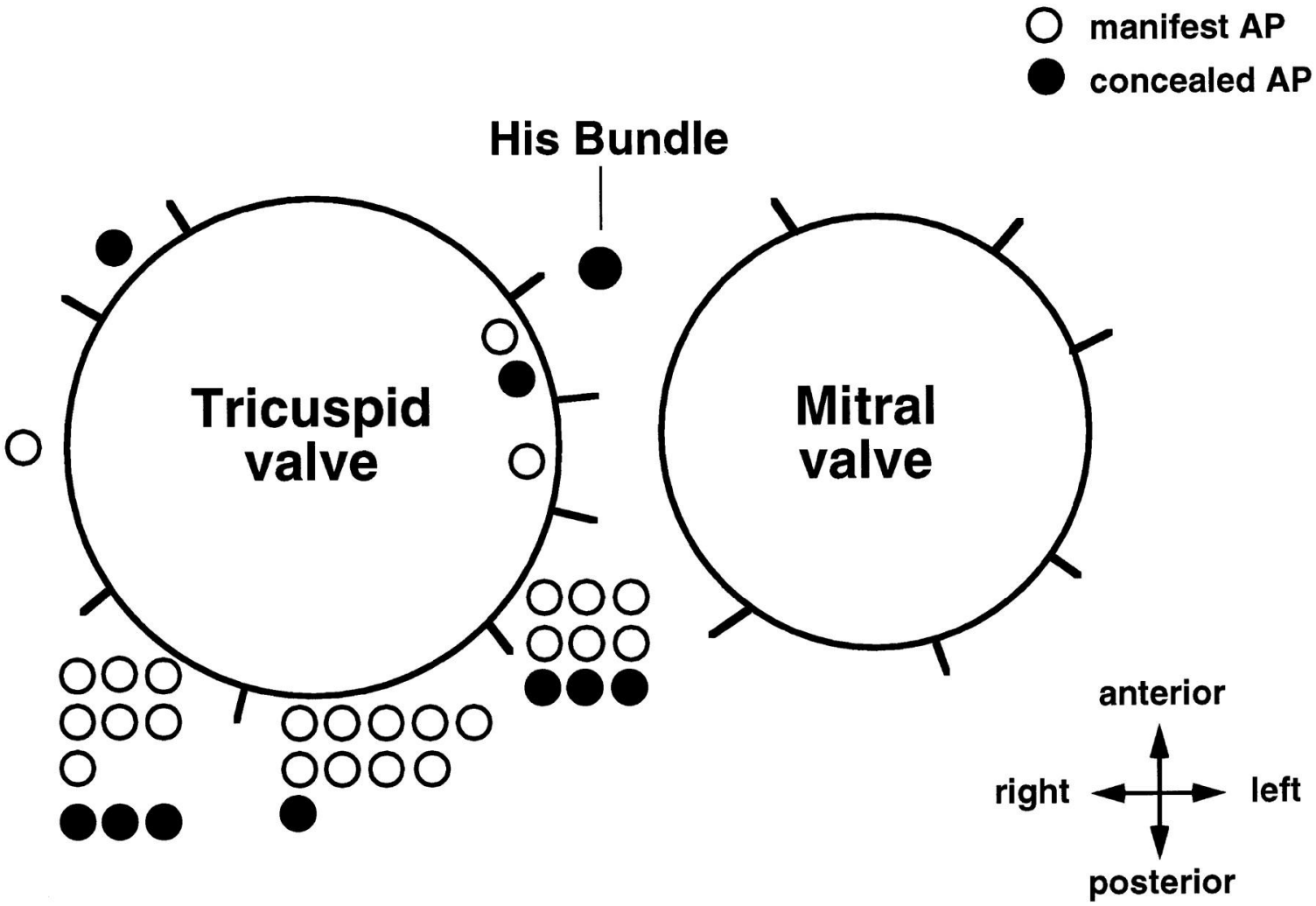
6 años





DIXTAL (0XX11) 5548-4155
DIXTAL (0XX11) 5548-4155

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

American Heart Association
Learn and Live

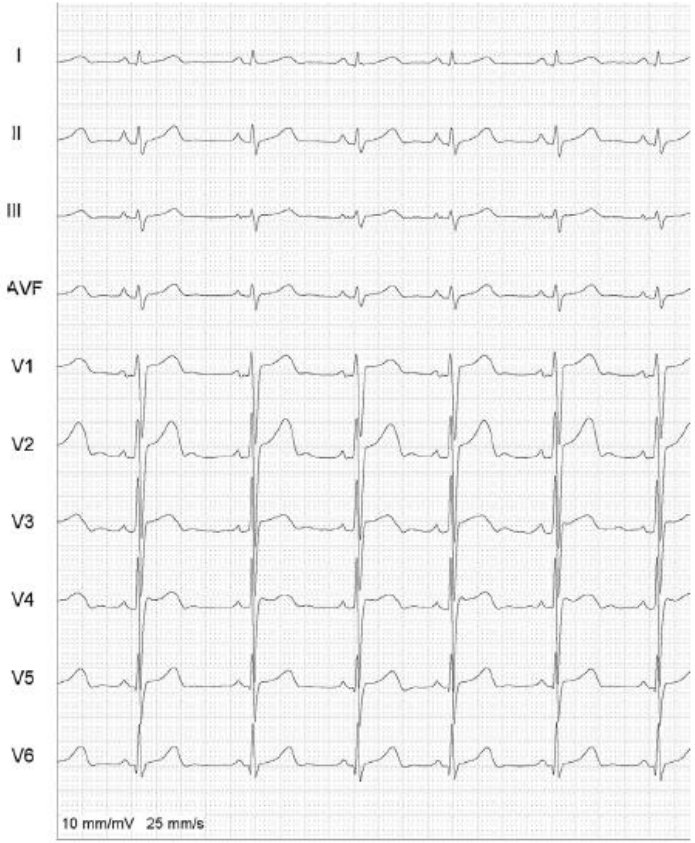
Copyright © American Heart Association

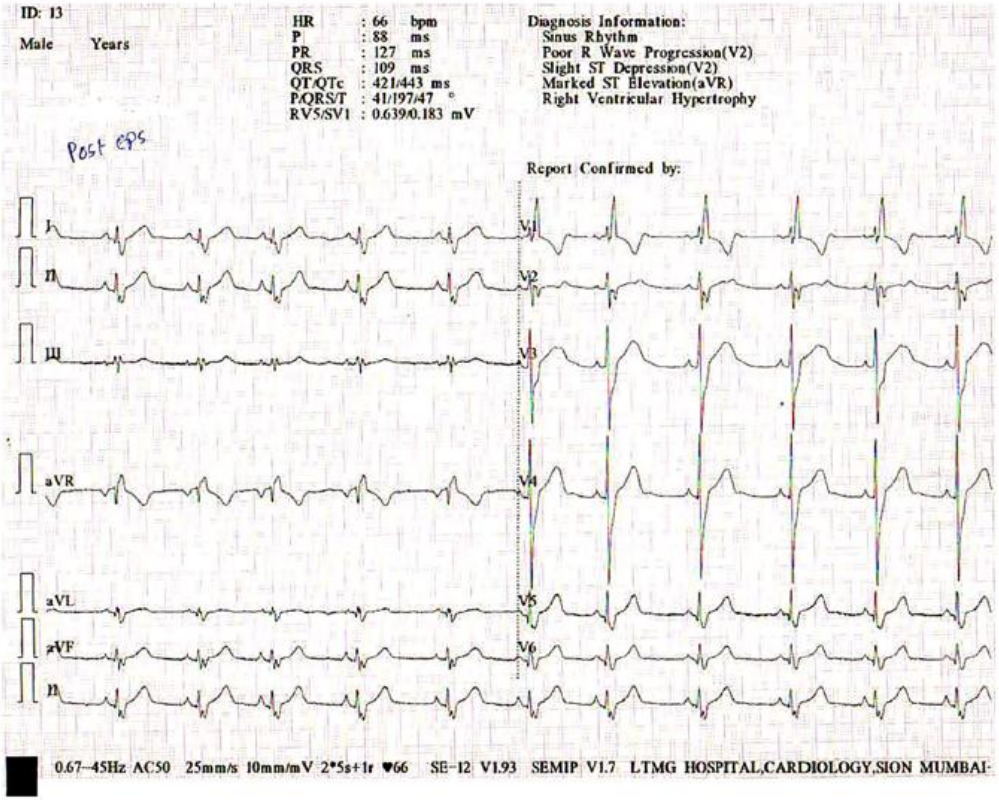
Available online at www.sciencedirect.com
ScienceDirect
journal homepage: www.elsevier.com/locate/ihj

Case Report

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

Pranil Bhalchandra Gangurde[a,*], *Abhay Tidake*[b], *Hetan Shah*[c], *Ajay Mahajan*[d], *Yash Lokhandwala*[e], *Pratap Nathani*[f]





http://dx.doi.org/10.1016/j.ihj.2015.11.001

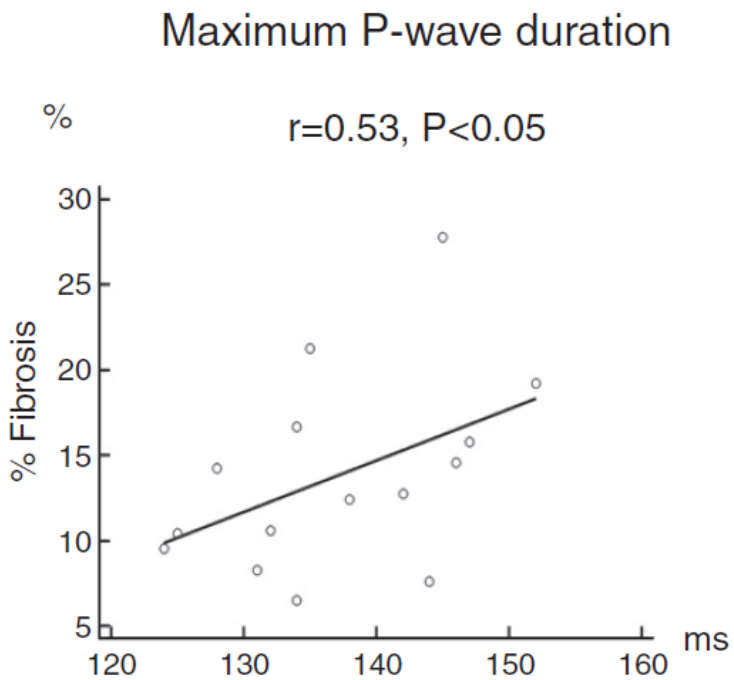
Surgery in a contemporary adult cohort of patients with Ebstein malformation of tricuspid valve and relation with ECG markers, atrial fibrosis and arrhythmic load

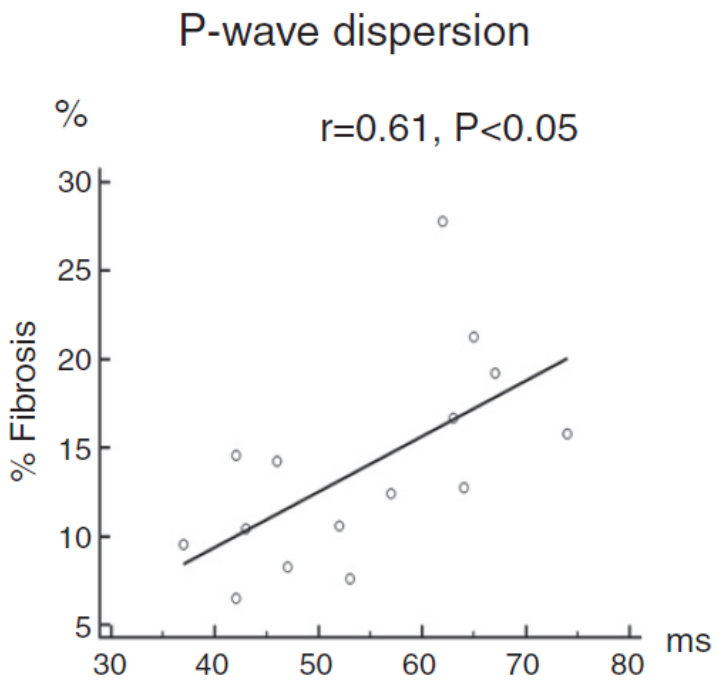
Yumi Shiina [a,*], Taka-aki Matsuyama [b], Iki Adachi [b], Wei Li [a], Michael A. Gatzoulis [a], Hideki Uemura [c]

[a] 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
[b] 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
[c] 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

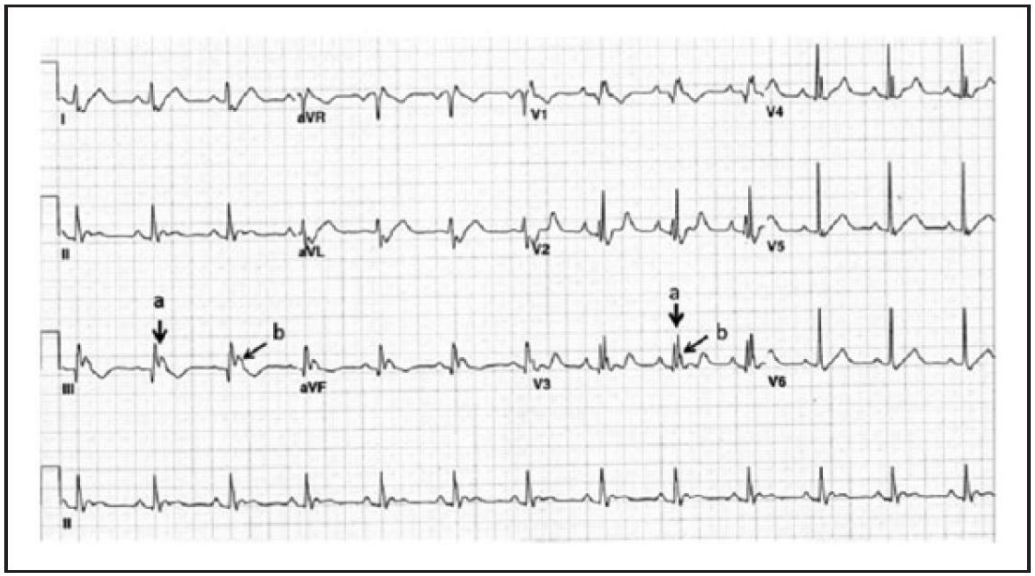
Fibrose e características da onda P no Ebstein





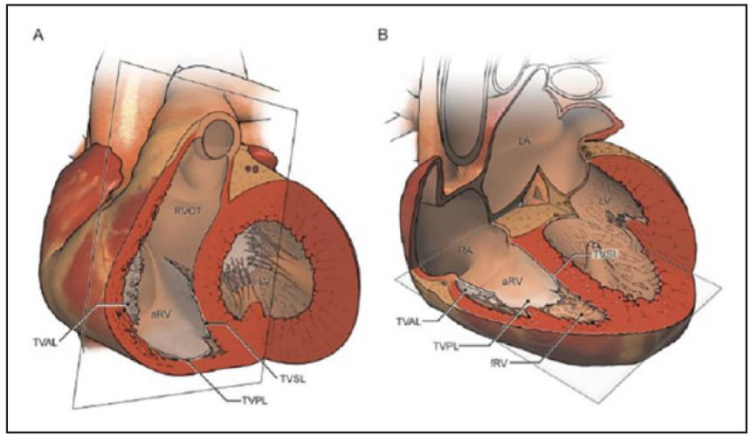
Journal of Investigative Medicine High Impact Case Reports
January-March 2017: 1–3
© 2017 American Federation for Medical Research
DOI: 10.1177/2324709616688710
journals.sagepub.com/home/hic
SAGE

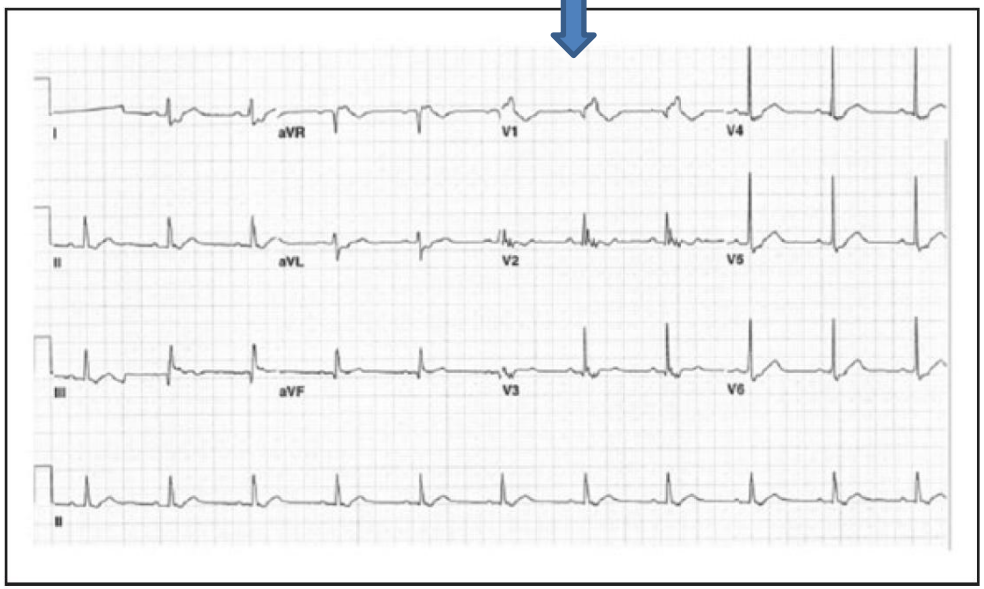
Fragmentação QRS anomalia Ebstein



Pré operatório

Pós operatório





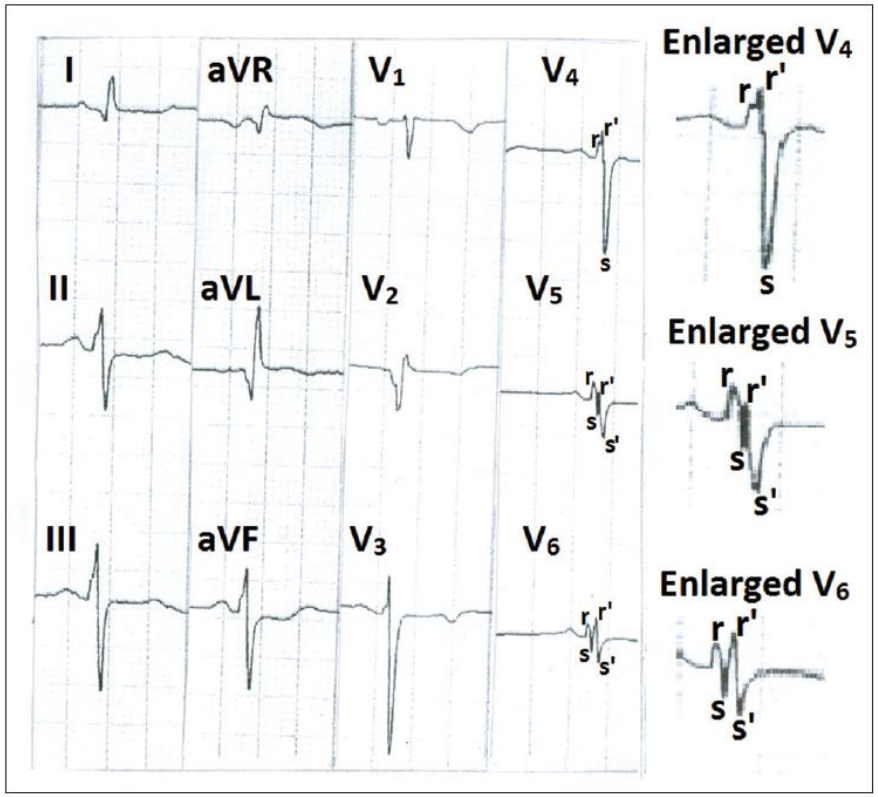


Figure 1: Electrocardiogram showing fragmented QRS in leads V_4 to V_6. Lead I, aVL, and V_2 also show Q-waves

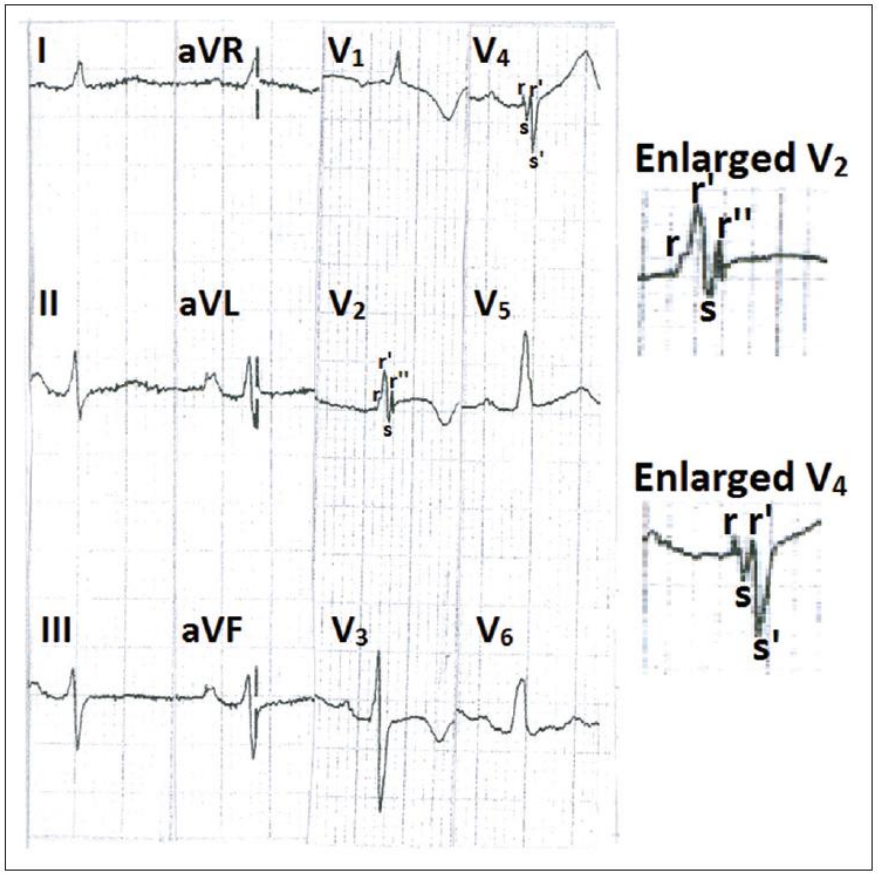
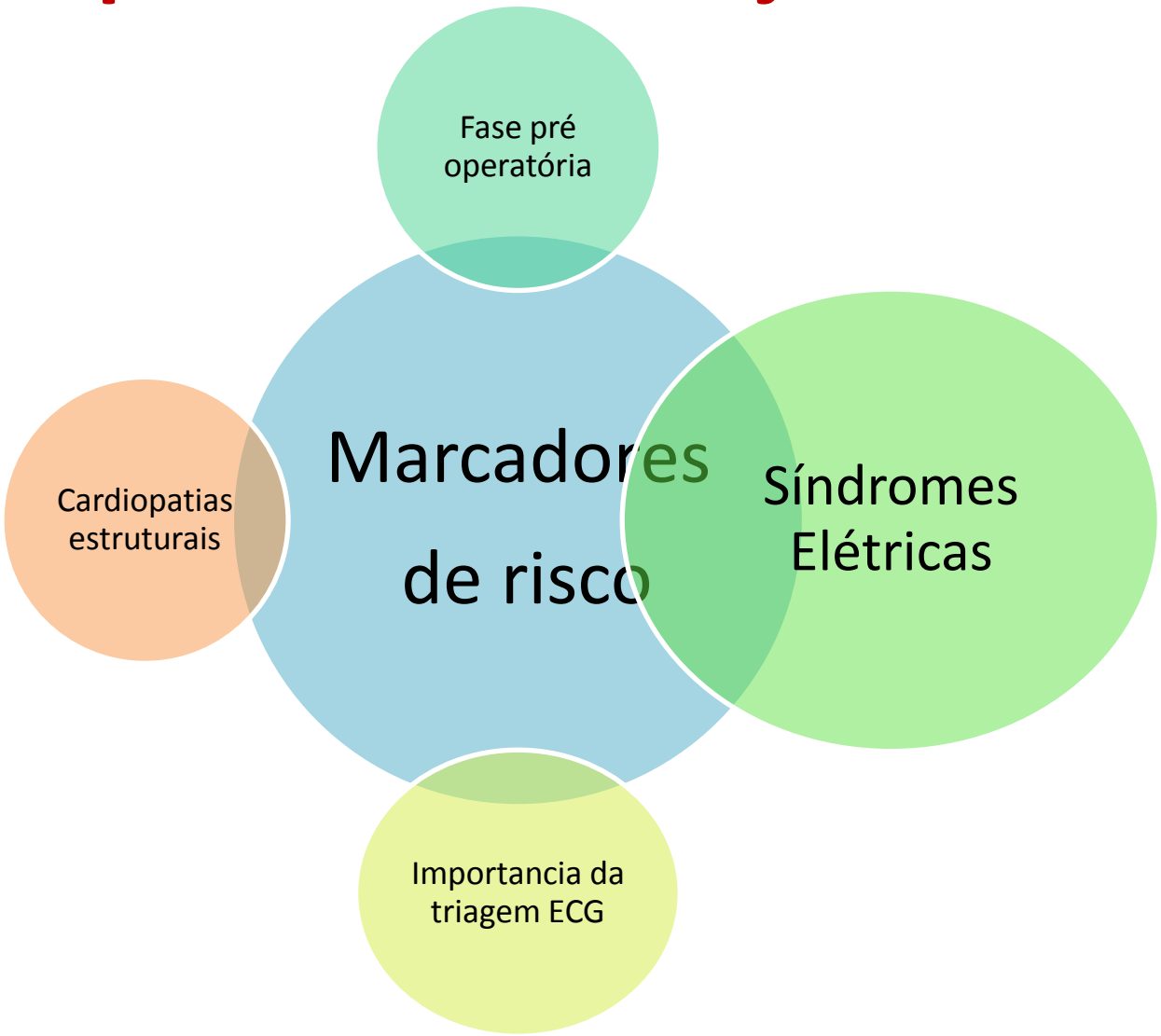


Figure 2: Electrocardiogram showing fragmented QRS in leads V_2 and V_4

Marcadores ECG na população pediátrica e em jovens

Marcadores ECG na população pediátrica e em jovens



Síndrome do QT longo

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

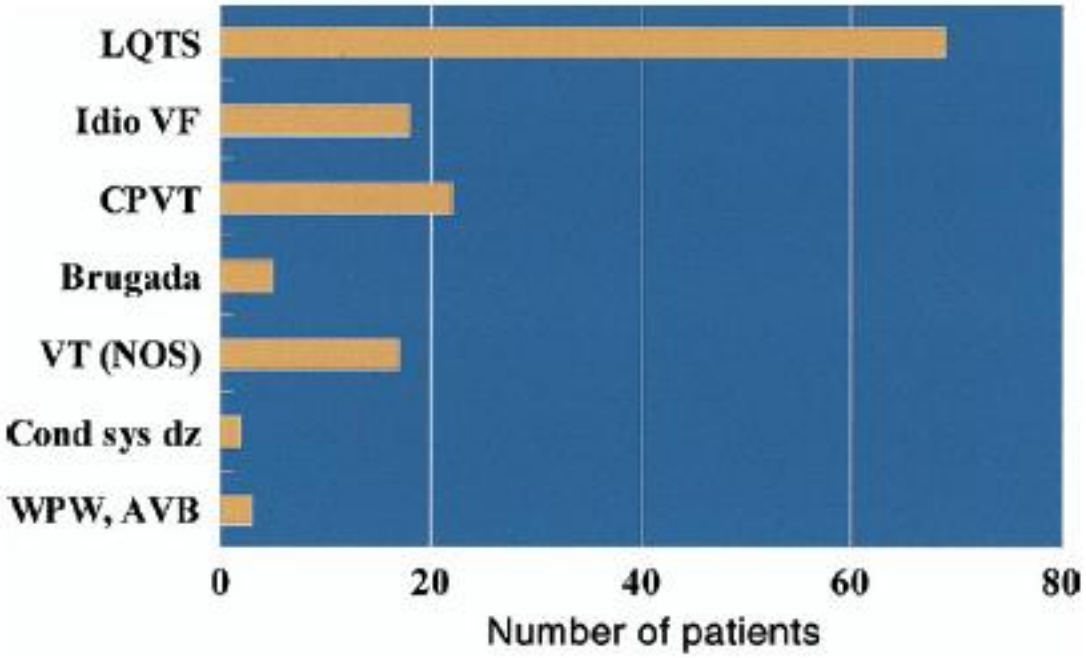
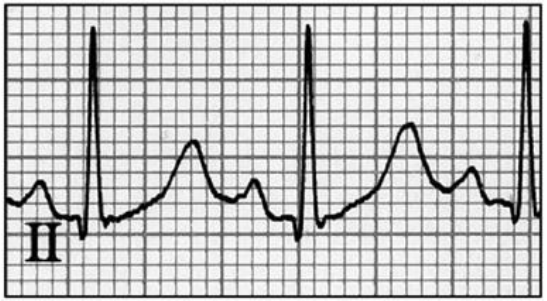


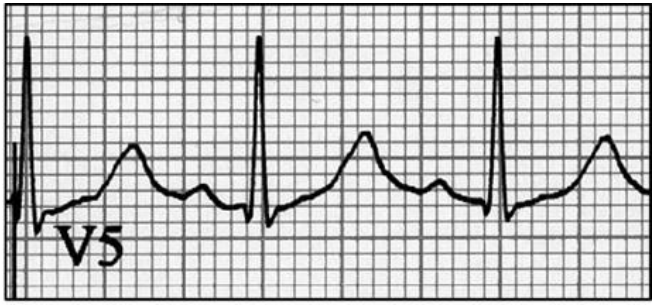
Figure 2 Electrical Diagnoses of Pediatric ICD Recipients With Structurally Normal Hearts

(J Am Coll Cardiol 2008;51:1685–91)

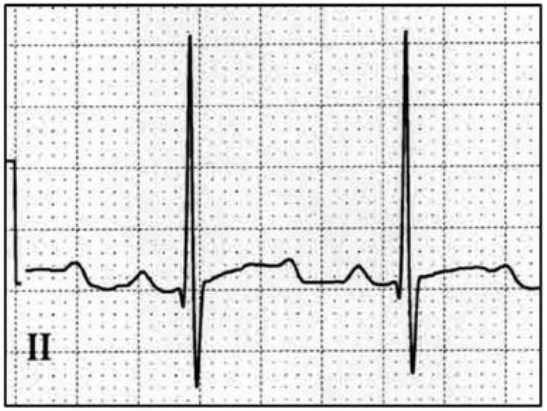
Padrões eletrocardiográficos QT longo

LQT1

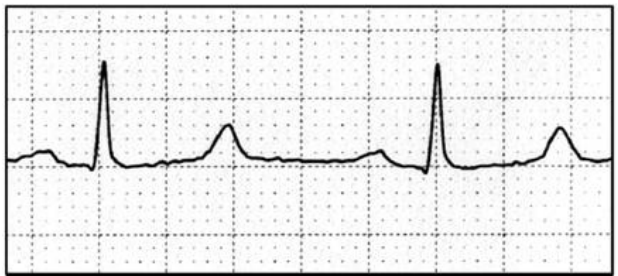




LQT2



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

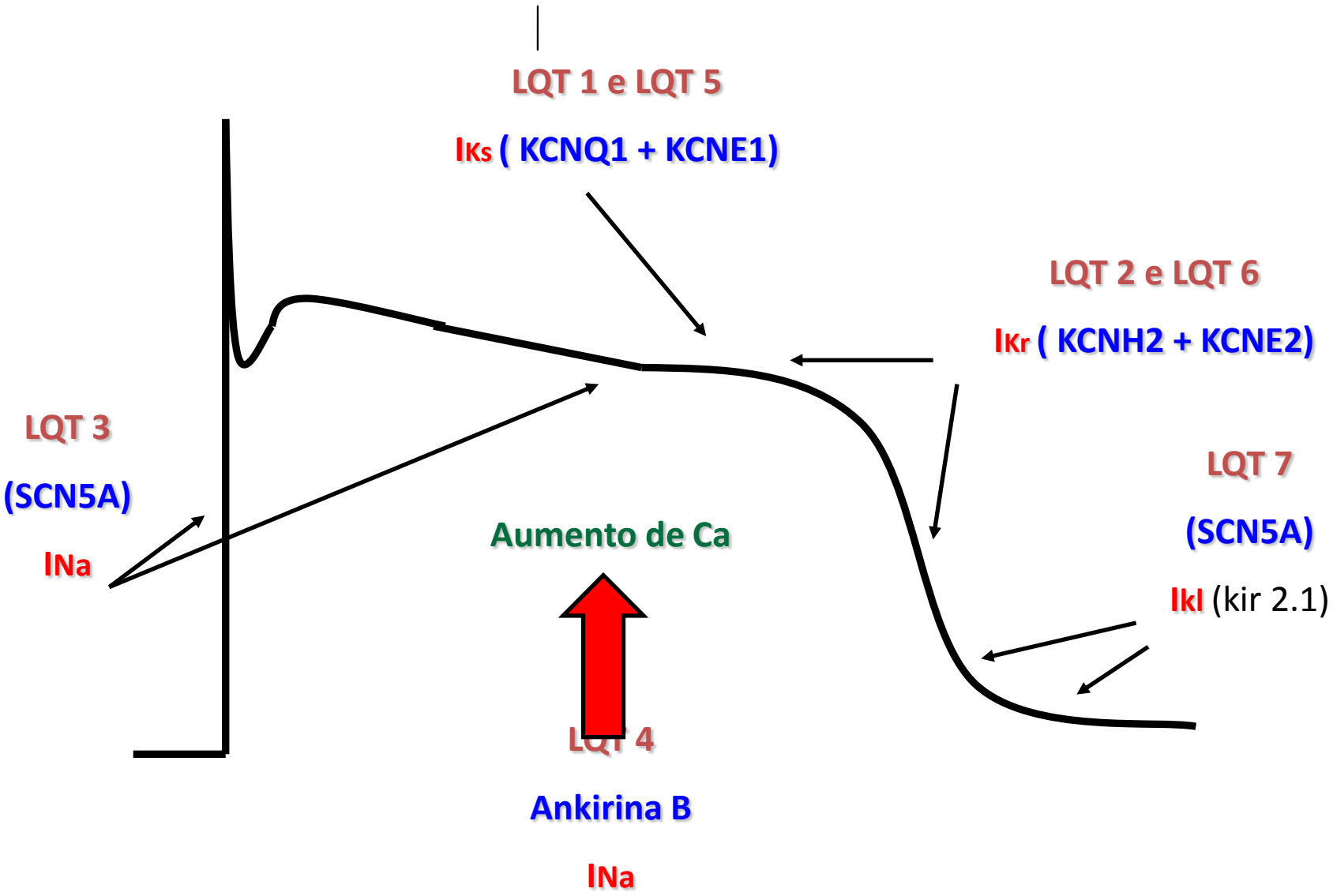
- Sentimento de palpitação ou “vibrar” são incomuns porque a TDP é muito rápida
- Sincope

Critérios diagnósticos Síndrome do QT longo (Schwartz)

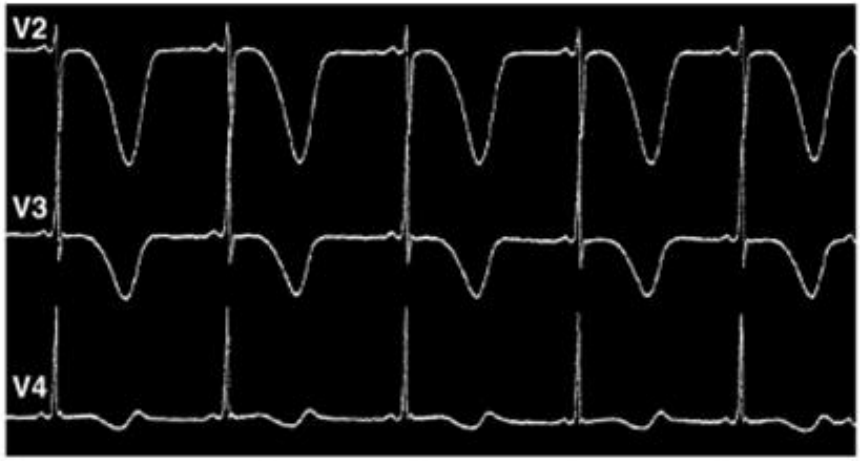
	Pontuação
QTc	
>480 ms	3
460 – 470 ms	2
450 ms (masculino)	1
Torsade pointes	2
Alternância de ondas T (TWA)	1
Baixa frequência cardíaca	0,5
Síncope	
Com estresse	2
Sem estresse	1
Surdez congênita	0,5
Membros da familia com LQTS	1
Morte cardíaca inexplicada na família (imediatos) antes dos 30 anos	0,5

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

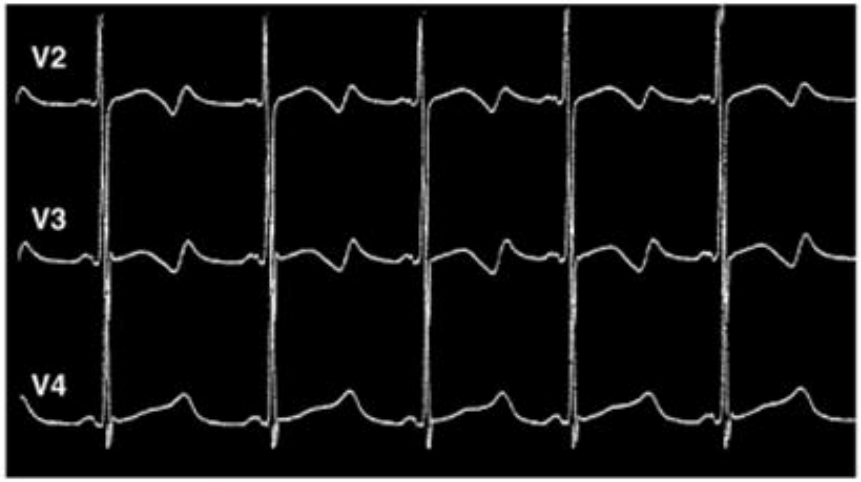
J Intern Med 2006; 259:39-47


LQT 1 e LQT 5
IKs (KCNQ1 + KCNE1)
LQT 2 e LQT 6
IKr (KCNH2 + KCNE2)
LQT 3
(SCN5A)
INa
Aumento de Ca
LQT 7
(SCN5A)
IkI (kir 2.1)
LQT 4
Ankirina B
INa

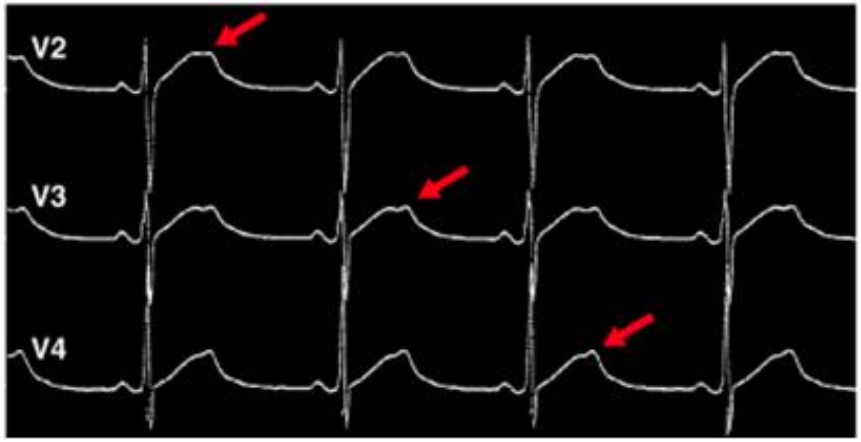
Caso índice
7 anos



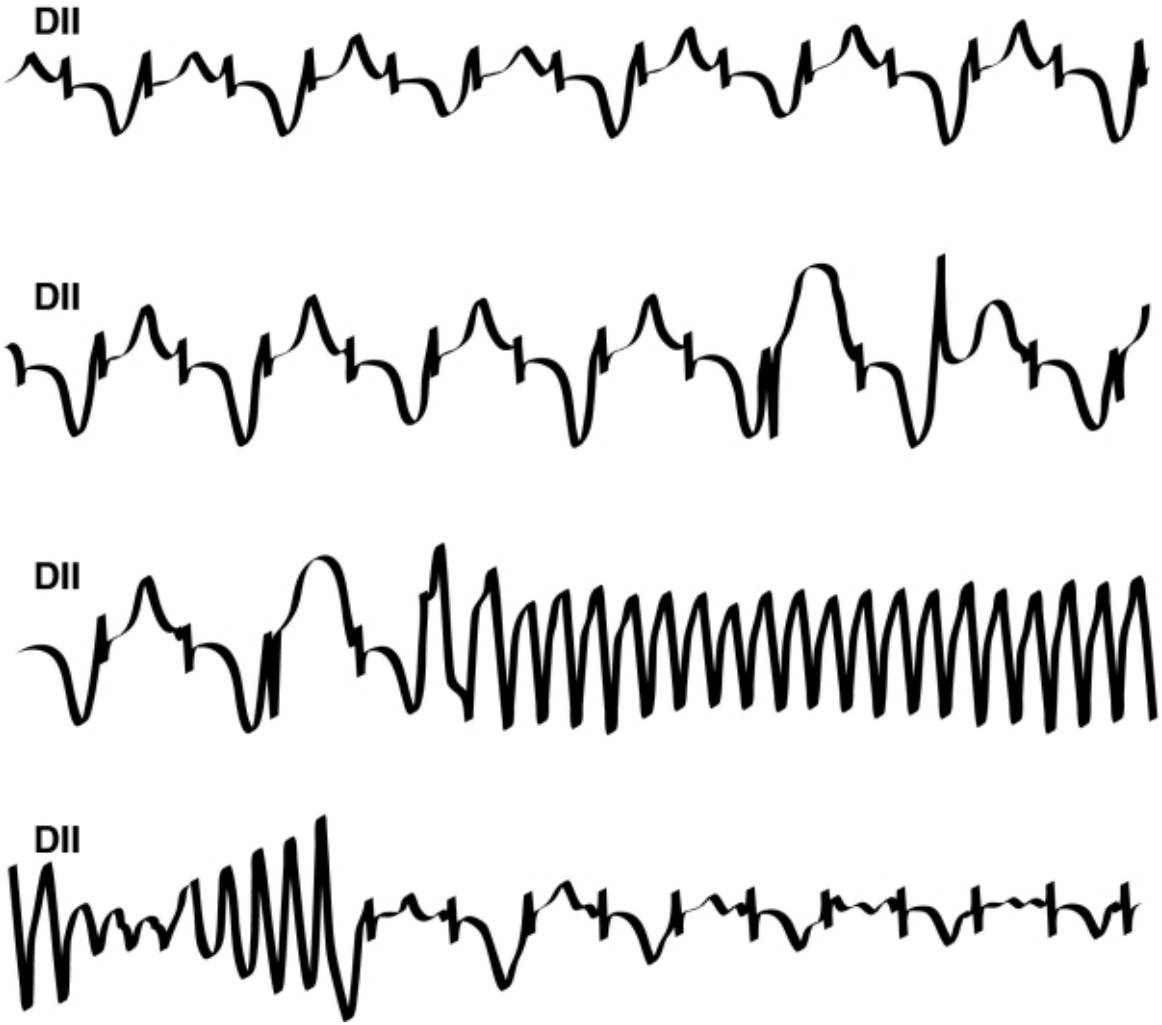
Irmã
10 anos



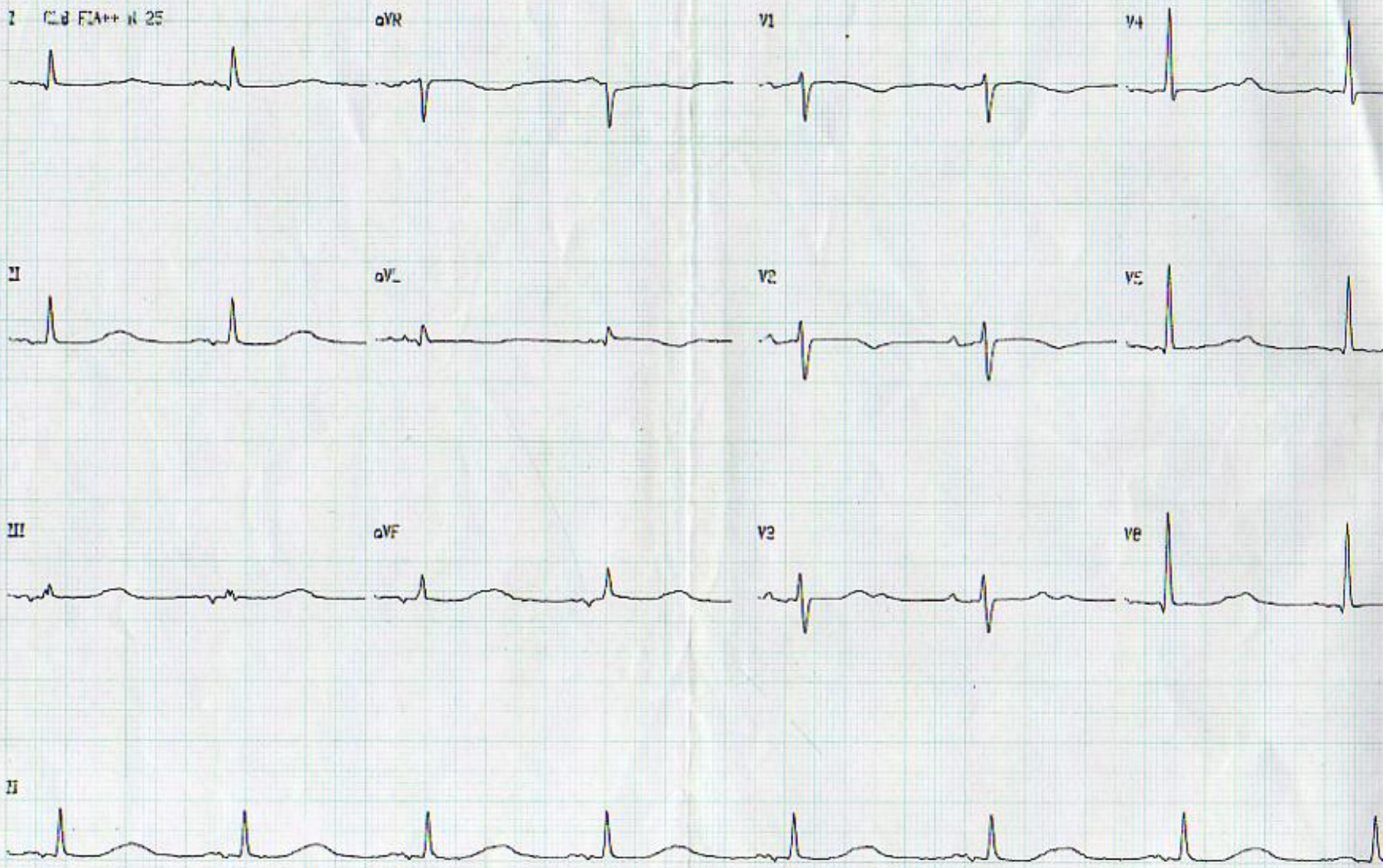
Pai
37 anos



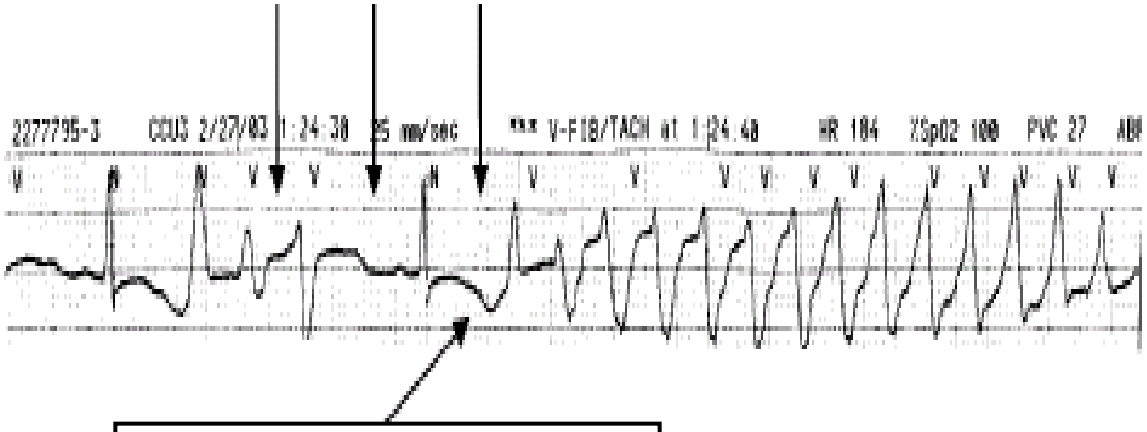
Alternancia de onda T no QT longo



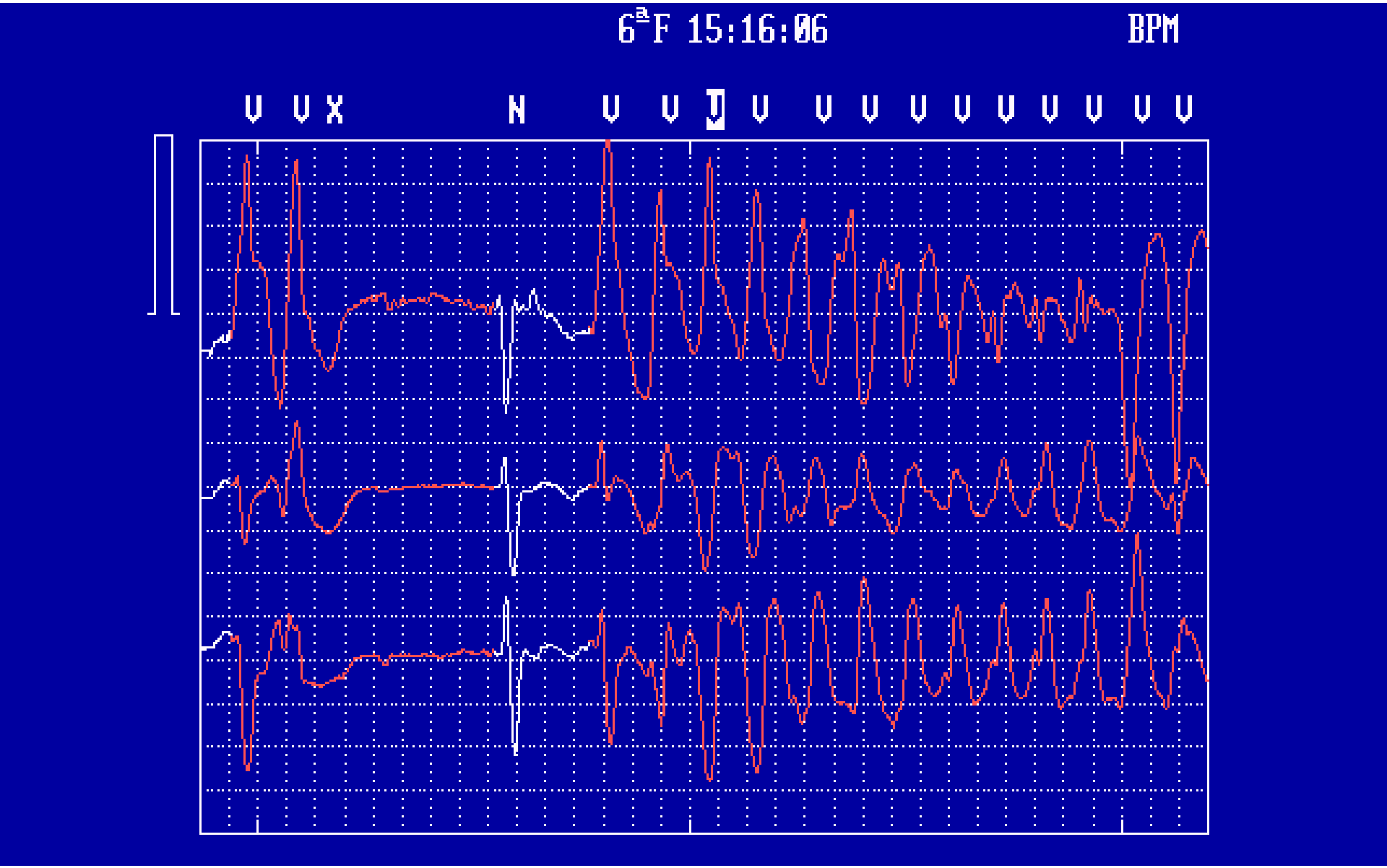
Europ Paediatr Cardiol 8:28-36, 1972.

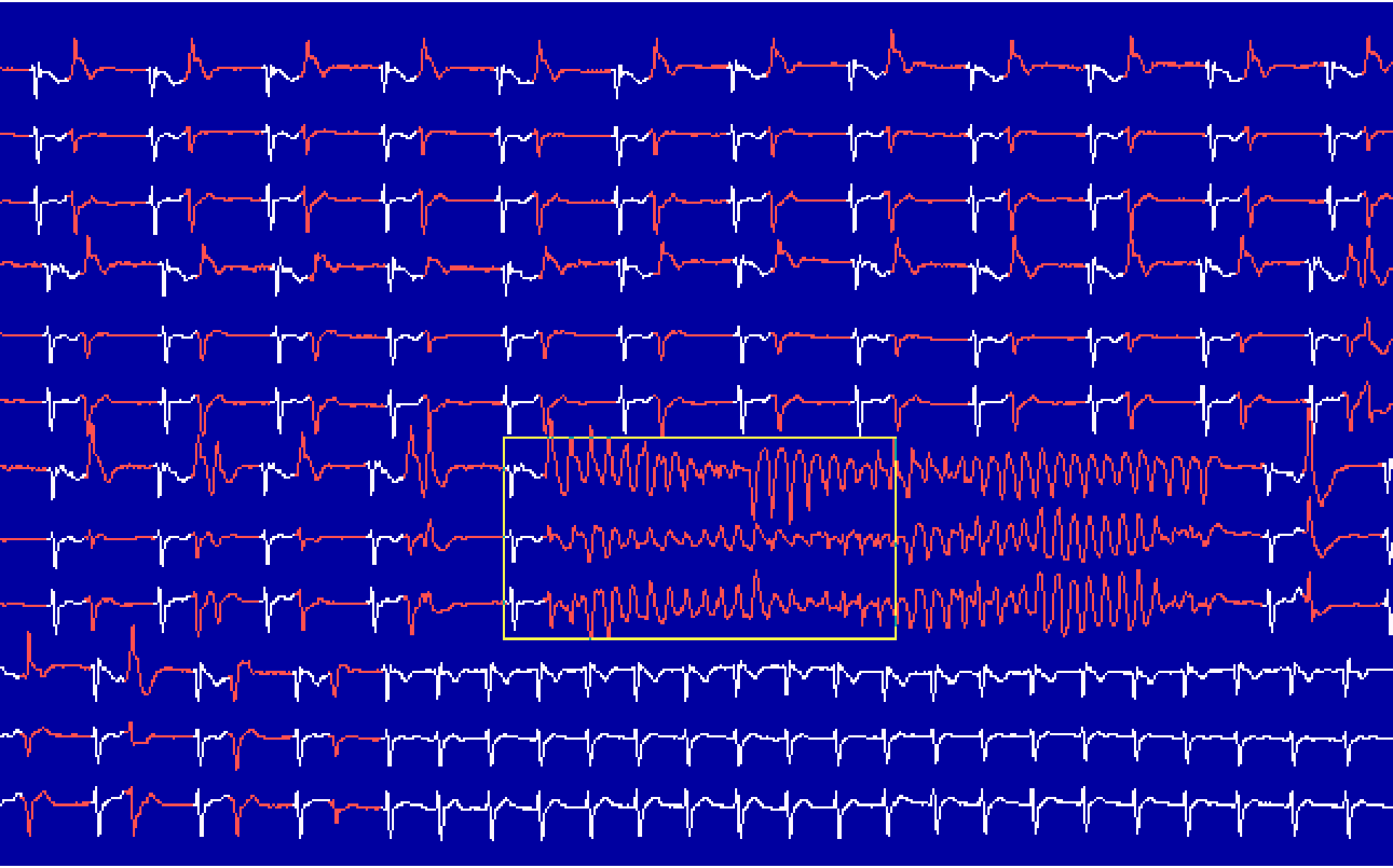

aVR
V1
V4
aVL
V2
V5
aVF
V3
V6
II

Curto Longo Curto

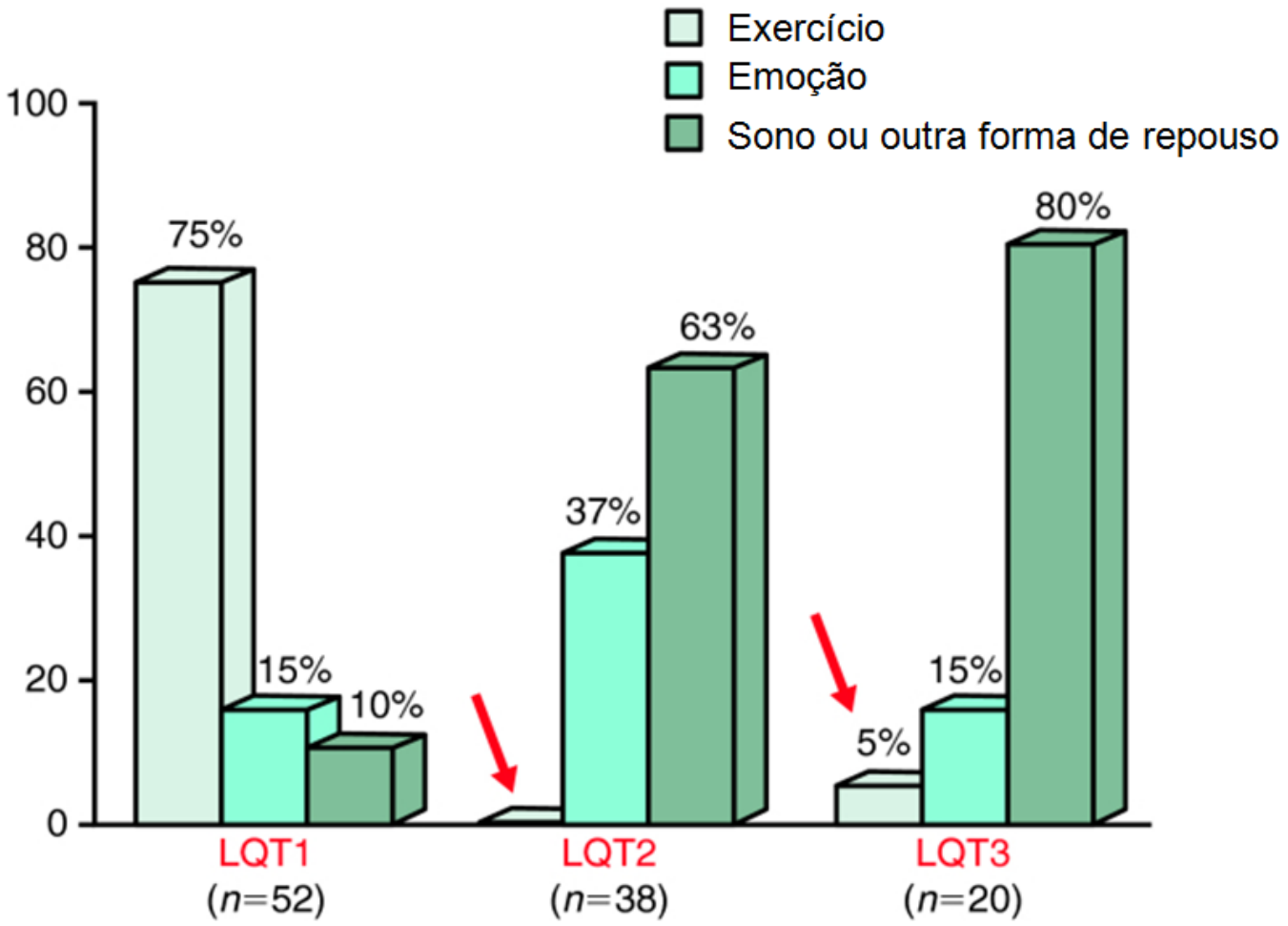


Intervalo QT longo e onda T bizarra

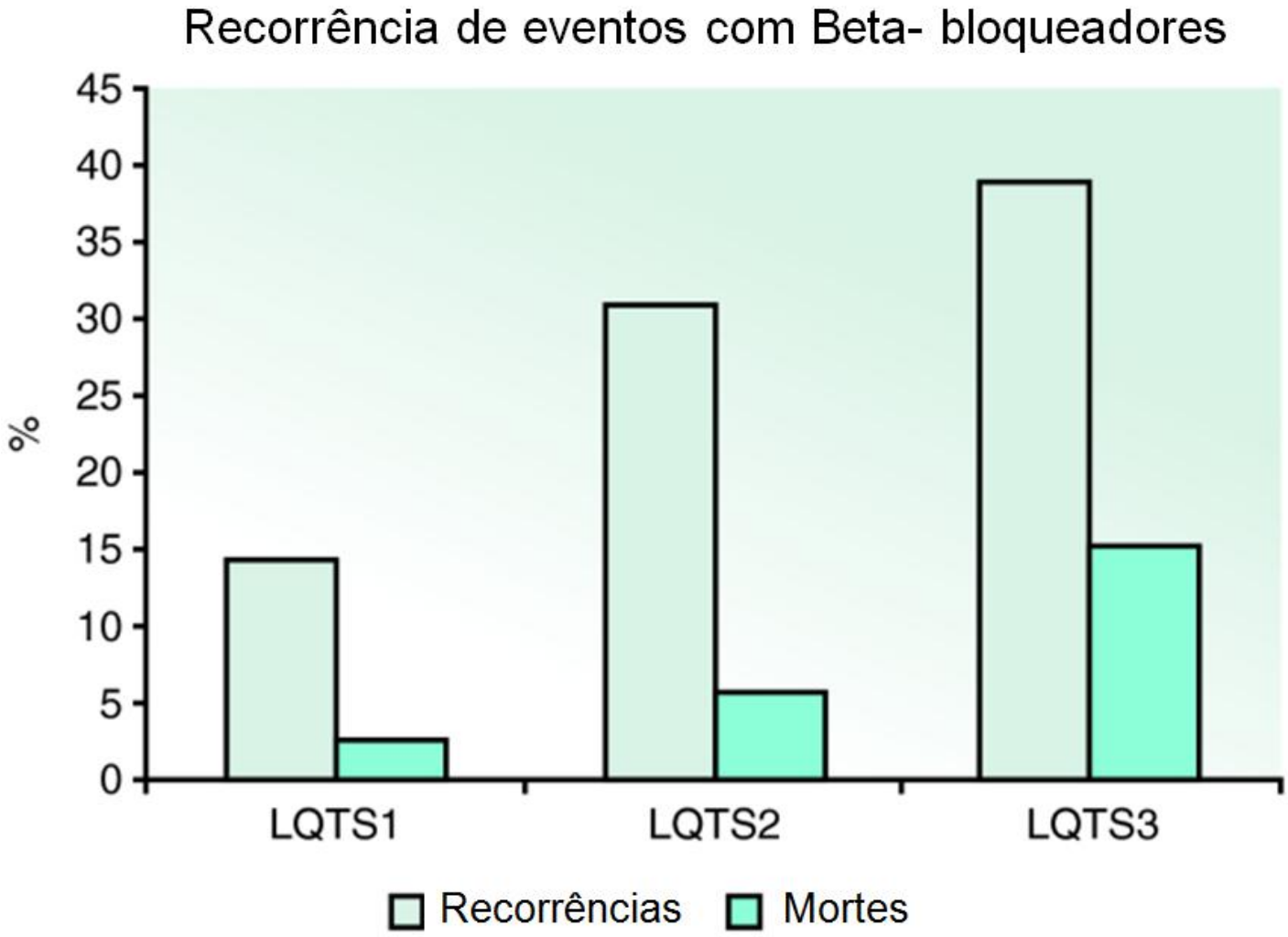

6ªF 15:16:06
BPM
V V X N V V J V V V V V V V V V V



Gatilho para eventos súbitos na LQTS



Circulation 103:89-95, 2001



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

	QTc	Sexo	Risco
LQT1	>= 500 ms	•Masculino e feminino	•Alto (>50%)
	<500 ms		•Baixo (<30%)
LQT2	>= 500 ms	•Masculino e feminino	•Alto (>50%)
	<500 ms	•Feminino •Masculino	•Intermediário (30 a 49%) •Baixo (<30%)
LQT3	>= 500 ms	•Masculino •Feminino	•Alto (>50%) •Intermediário (30 a 49%)
	<500 ms	•Masculino e Feminino	•Intermediário (30 a 49%)

Estratificação de risco Síndrome do QT longo

	I	IIa	IIb
Estratificação de Risco	•Torsade pointes/FV •Parada cardíaca •JLN •LQT3	•QTc > 600 ms •Evento cardíaco na infância •Sindactilia + BAVT •Alternância de T (TWA) •Sexo Feminino	•História familiar de MS •Aumento da dispersão do QT
Prevenção Primária	• Suspender medicamentos que prolongam o QT •Suspender atividade física •Betabloqueadores	CDI	•Denervação cardíaca
Prevenção Secundária	•CDI •Betabloqueadores •Suspender medicamentos que prolongam o QT •Suspender atividade física	-	-

Europace 2002; 4:3-18

Diferenças fisiopatológicas QT longo e QT curto

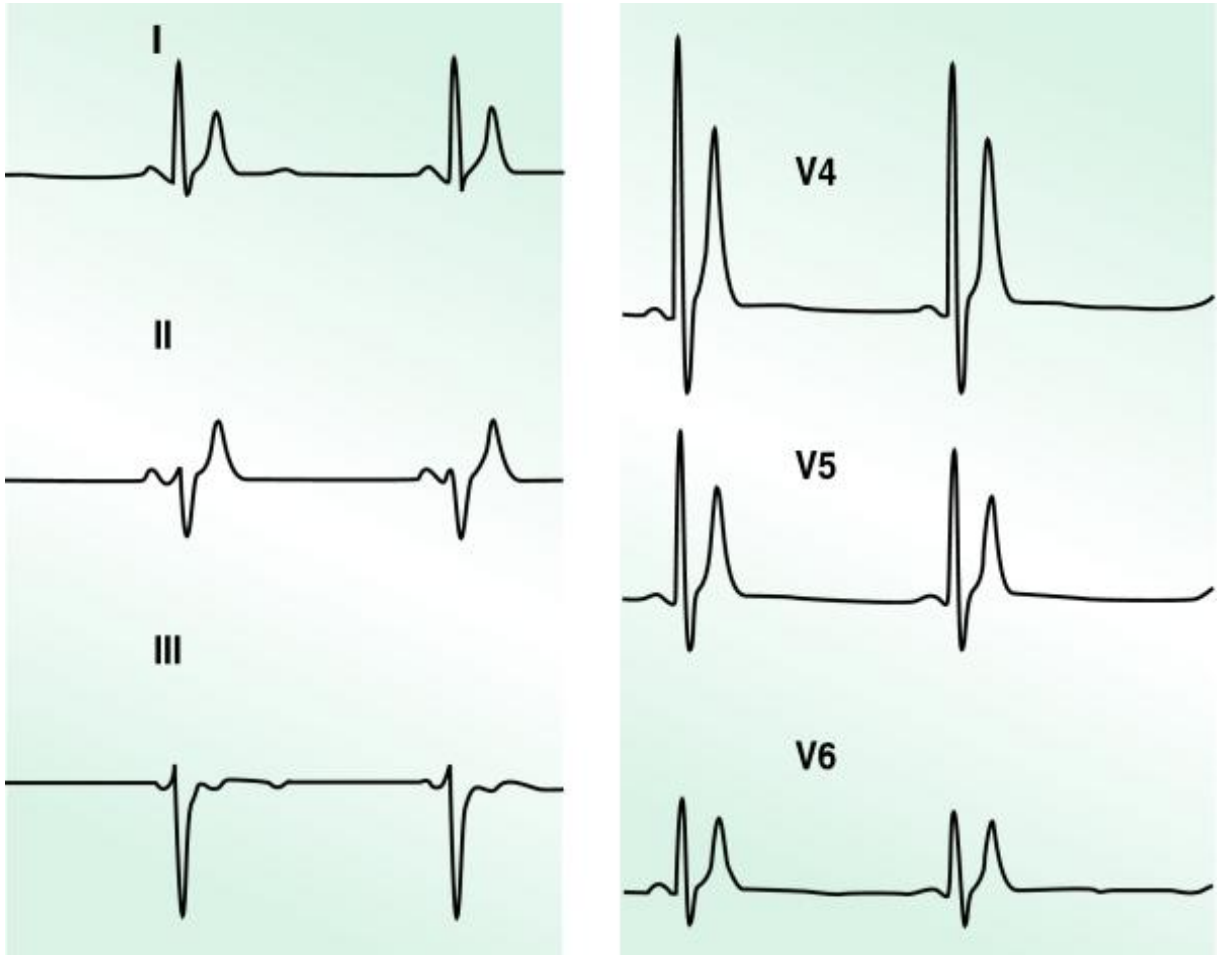
Electrophysiological changes observed in long and short QT syndromes (LQTS and SQTS). CV: conduction velocity. APD: action potential duration. ERP: effective refractory period. TDR: transmural dispersion of repolarization. λ: excitation wavelength (CV × ERP).

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	↔/↓	↔/↑	[63]
APD	↑	↓	[53]
ERP	↑	↓	[63]
TDR	↑	↑	[54,87]
λ	↓	↓	[37,63]
APD alternans	↑	N/A	[88,89]
CV restitution	↔	N/A	[29,90,91]
APD restitution	↑gradient	N/A	[29,92,93]
VERP restitution	↑gradient	N/A	[94]

Síndrome do QT curto

- Asoociado 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

Journal of the American College of Cardiology
Vol. 63, No. 13, 2014
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2013.09.078
© 2014 by the American College of Cardiology Foundation
Published by Elsevier Inc.

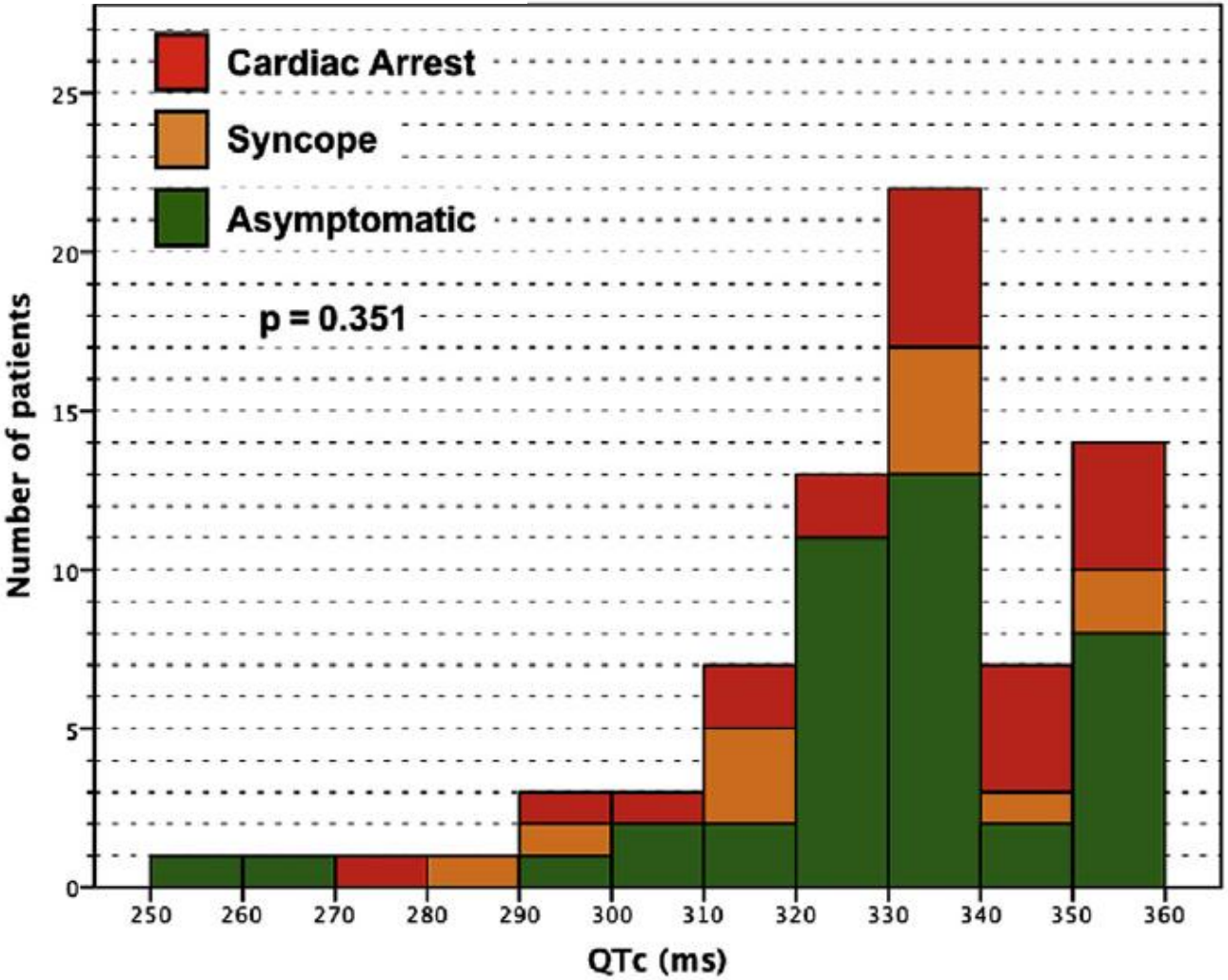
Heart Rhythm Disorders

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,* Sean O'Rourke, BS,† Gianluca Borio, MS,* Agnieszka Zienciuk-Krajka, MD,‡ 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)



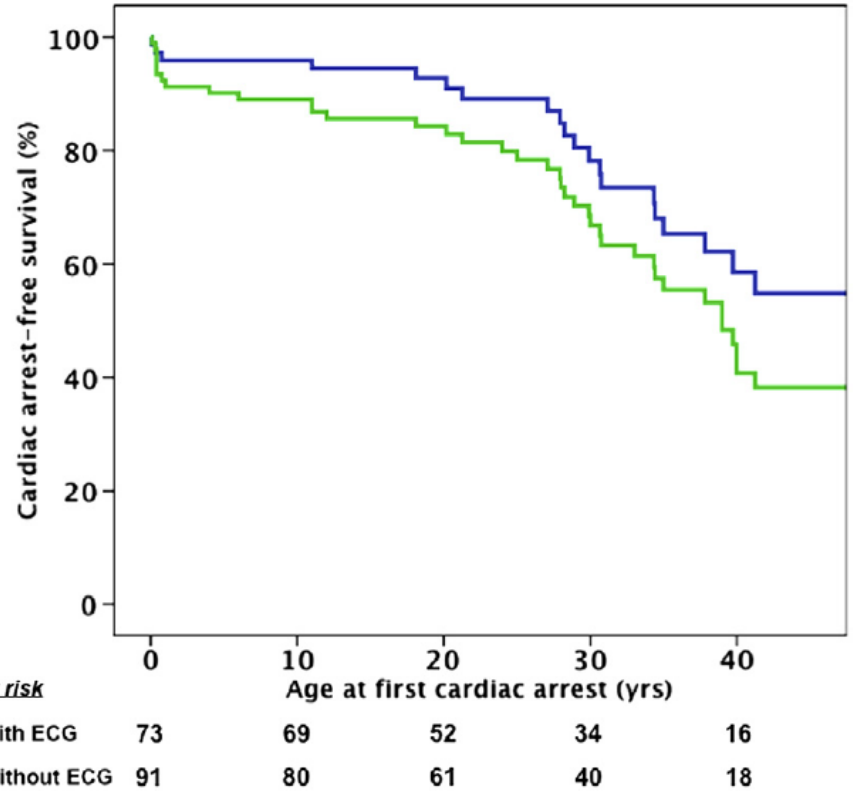
Journal of the American College of Cardiology
Vol. 63, No. 13, 2014
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2013.09.078
© 2014 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Heart Rhythm Disorders

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,*
Sean O'Rourke, BS,† Gianluca Borio, MS,* Agnieszka Zienciuk-Krajka, MD,‡
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

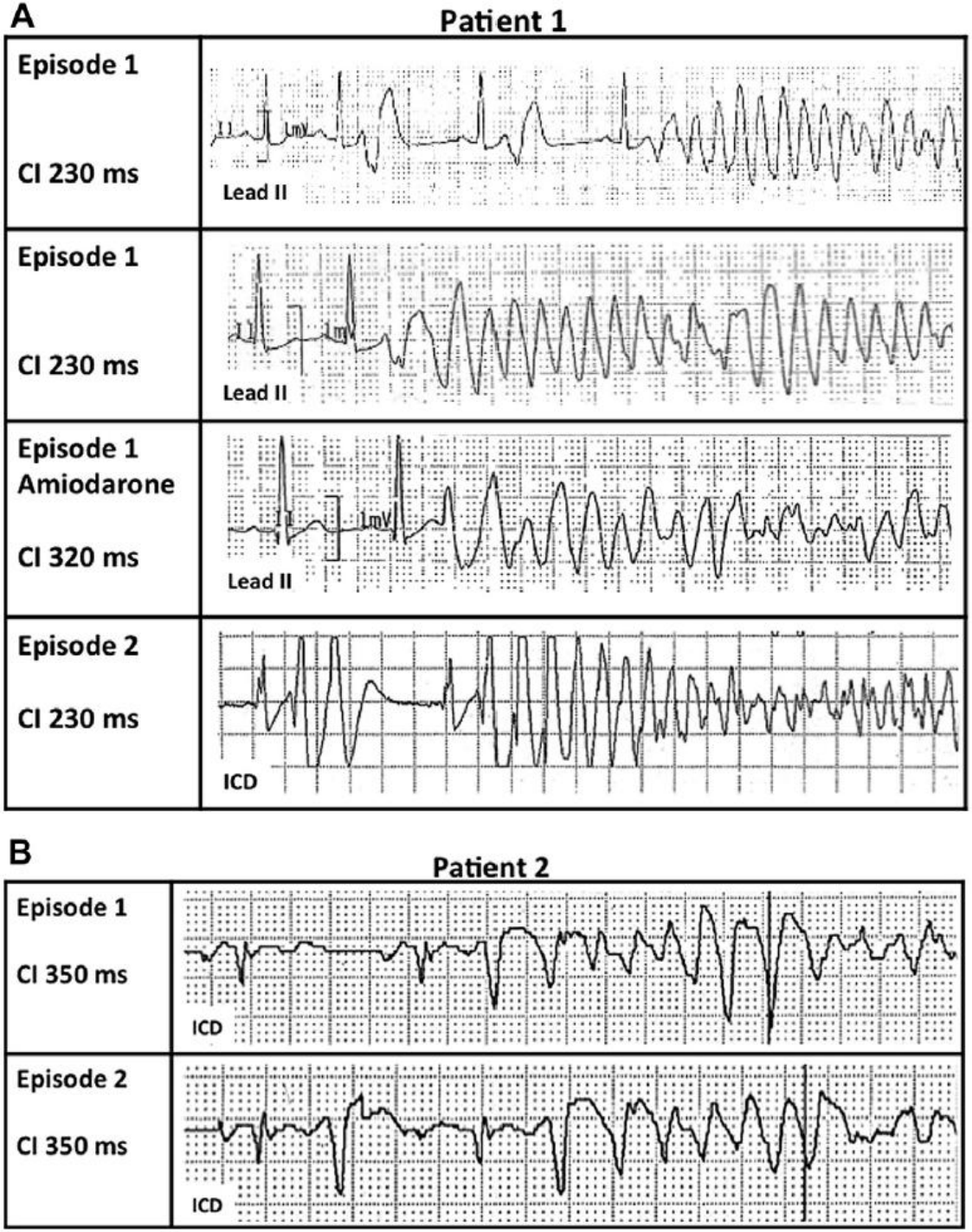


	Asymptomatic (n = 41)	Syncope (n = 12)	CA (n = 20)	p Value*
Male	32 (78)	11 (92)	18 (90)	0.353
Age at event, yrs	—	21 ± 11	25 ± 13	0.453
RR interval, ms	948 ± 93	938 ± 153	920 ± 132	0.682
QT interval, ms	321 ± 30	314 ± 29	317 ± 32	0.765
QTc interval, ms	330 ± 22	326 ± 24	331 ± 22	0.802

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*		Adopted	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
Vol. 63, No. 13, 2014
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2013.09.078
Published by Elsevier Inc.
© 2014 by the American College of Cardiology Foundation

Heart Rhythm Disorders

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,* Sean O'Rourke, BS,† Gianluca Borio, MS,* Agnieszka Zienciuk-Krajka, MD,‡ 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)

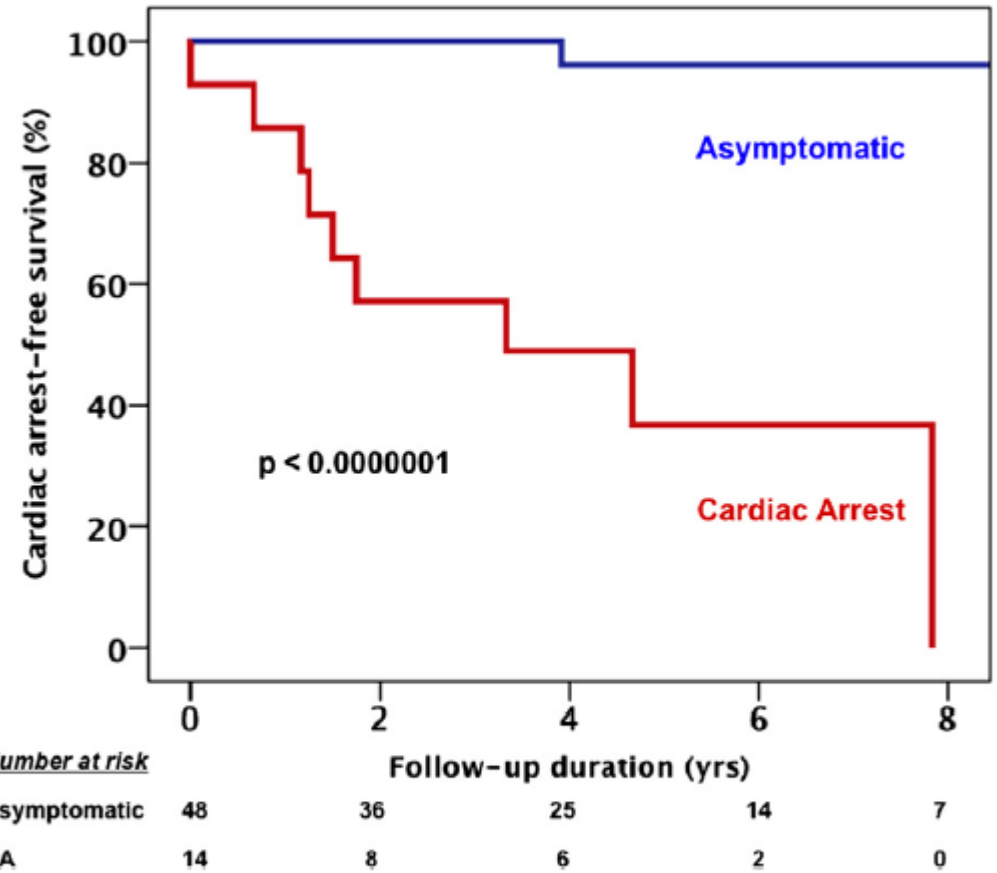
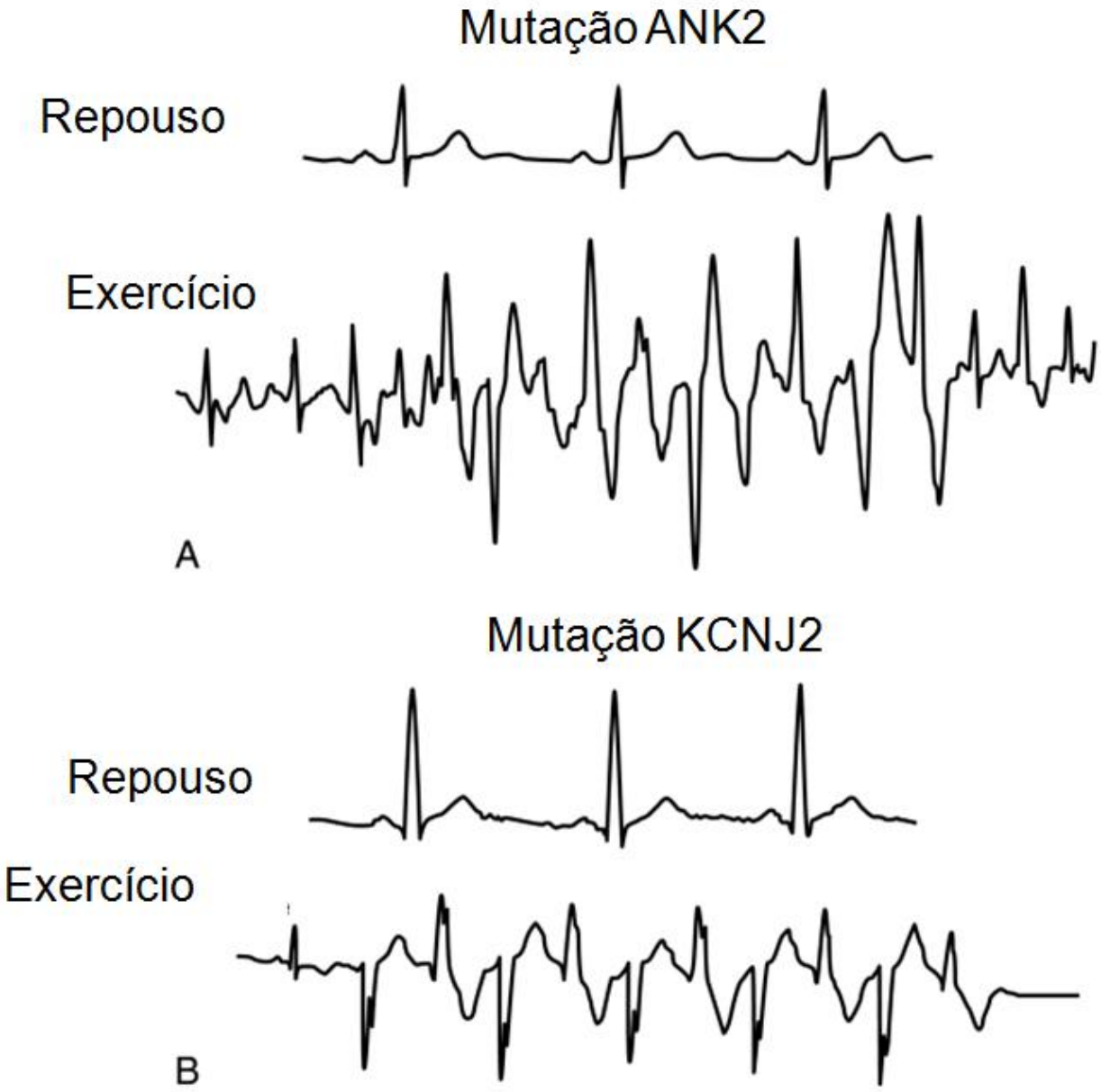


Figure 4 CA-Free Survival at Follow-up by CA Occurrence Before Enrollment

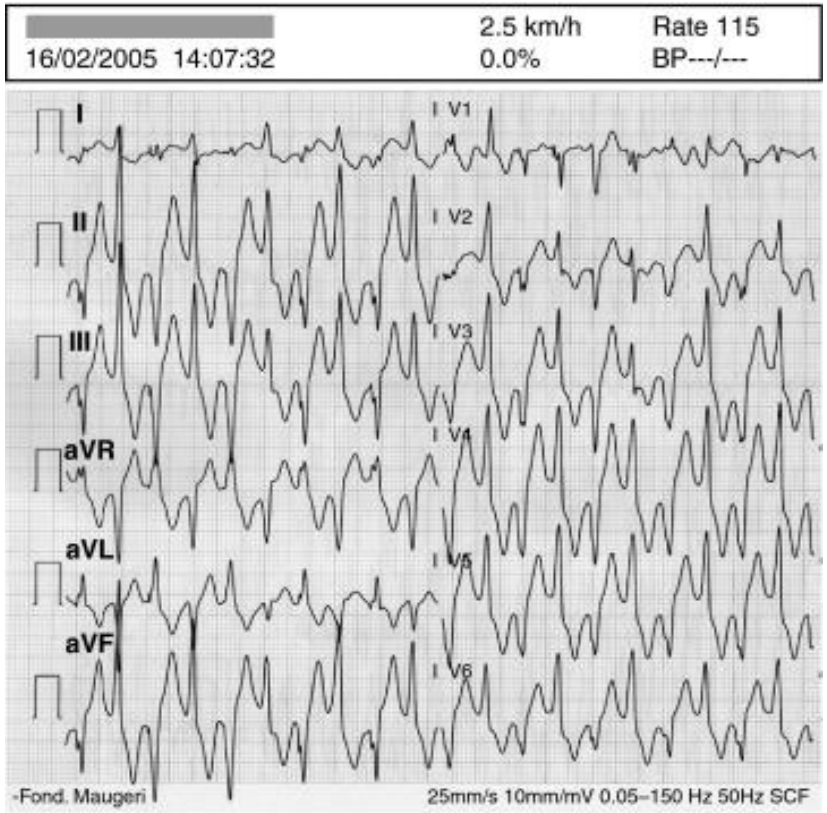
Taquicardia ventricular catecolaminérgica

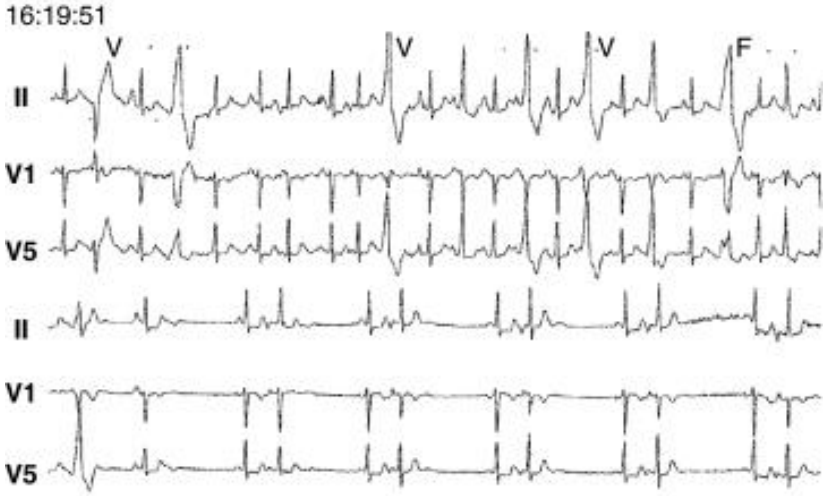
5.4.5. *Taquicardia bidirecional*[18]

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.



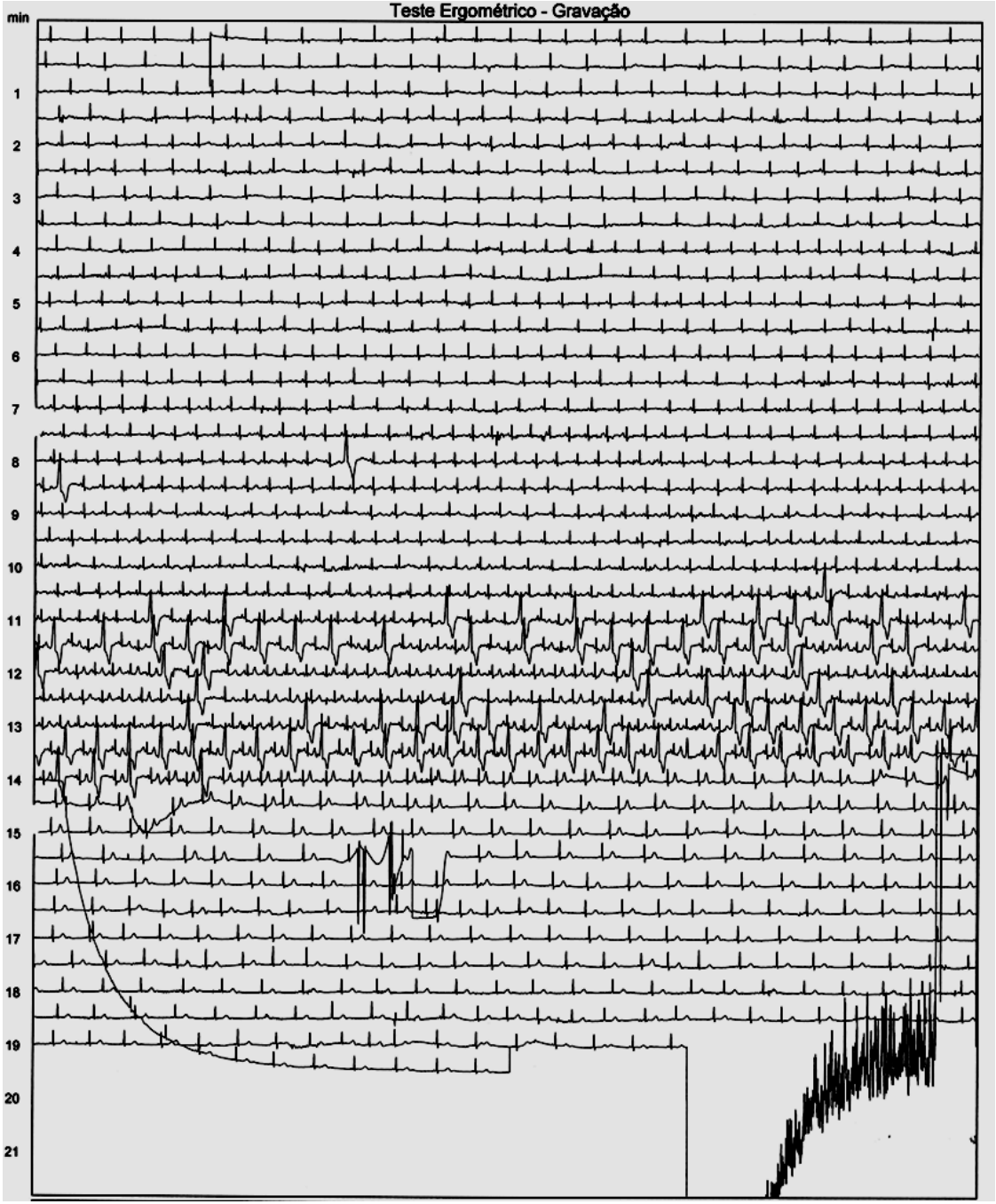
Heart Rhythm 2:1218-1223, 2005.


2.5 km/h
Rate 115
16/02/2005 14:07:32
0.0%
BP---/---
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
-Fond. Maugeri
25mm/s 10mm/mV 0.05–150 Hz 50Hz SCF


16:19:51
V
V
V
F
II
V1
V5
II
V1
V5

Menina 4 anos síncope aos esforços pré medicação

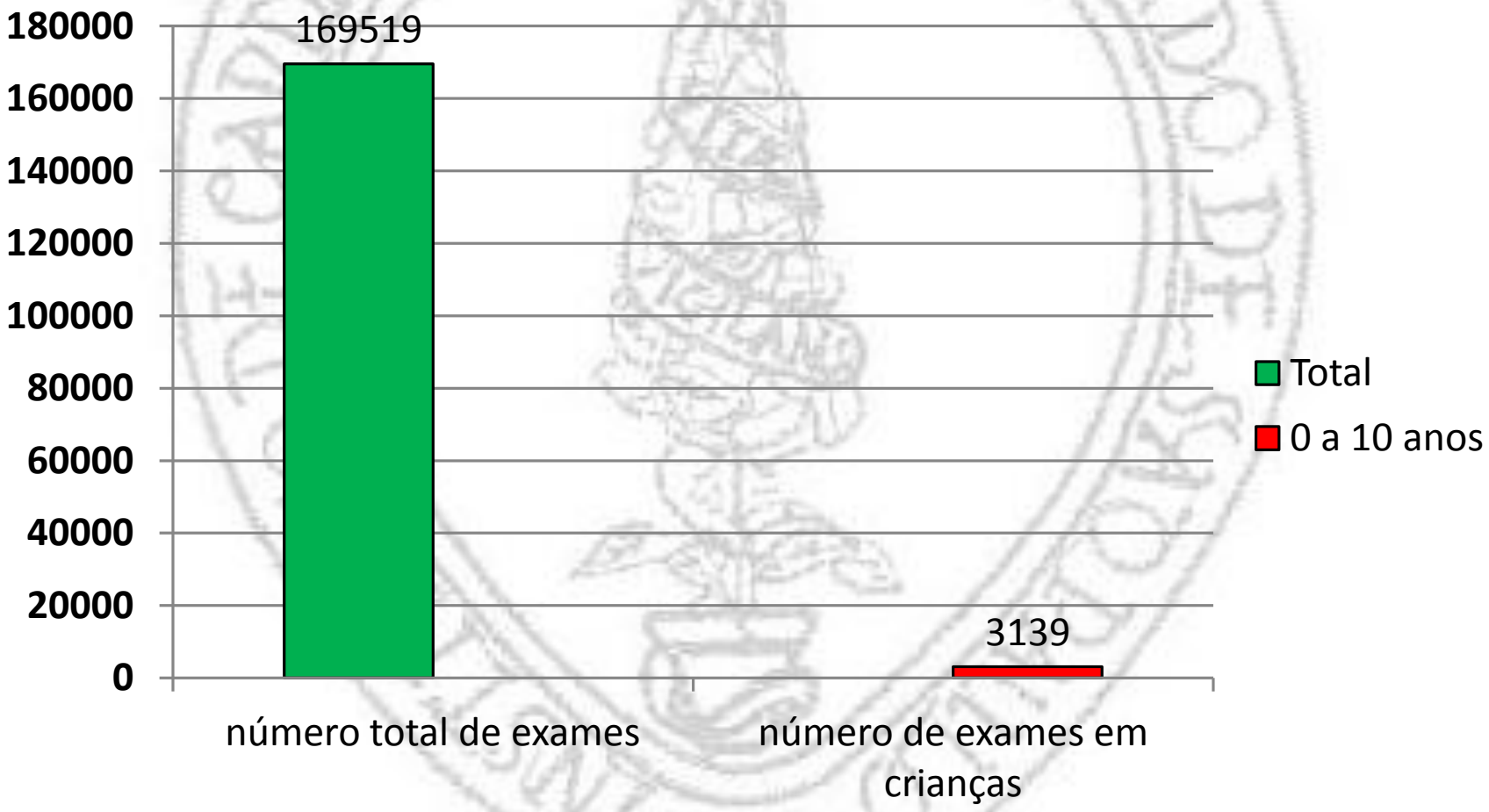
Menina 4 anos síncope aos esforços após medicação



Marcadores ECG na população pediátrica e em jovens

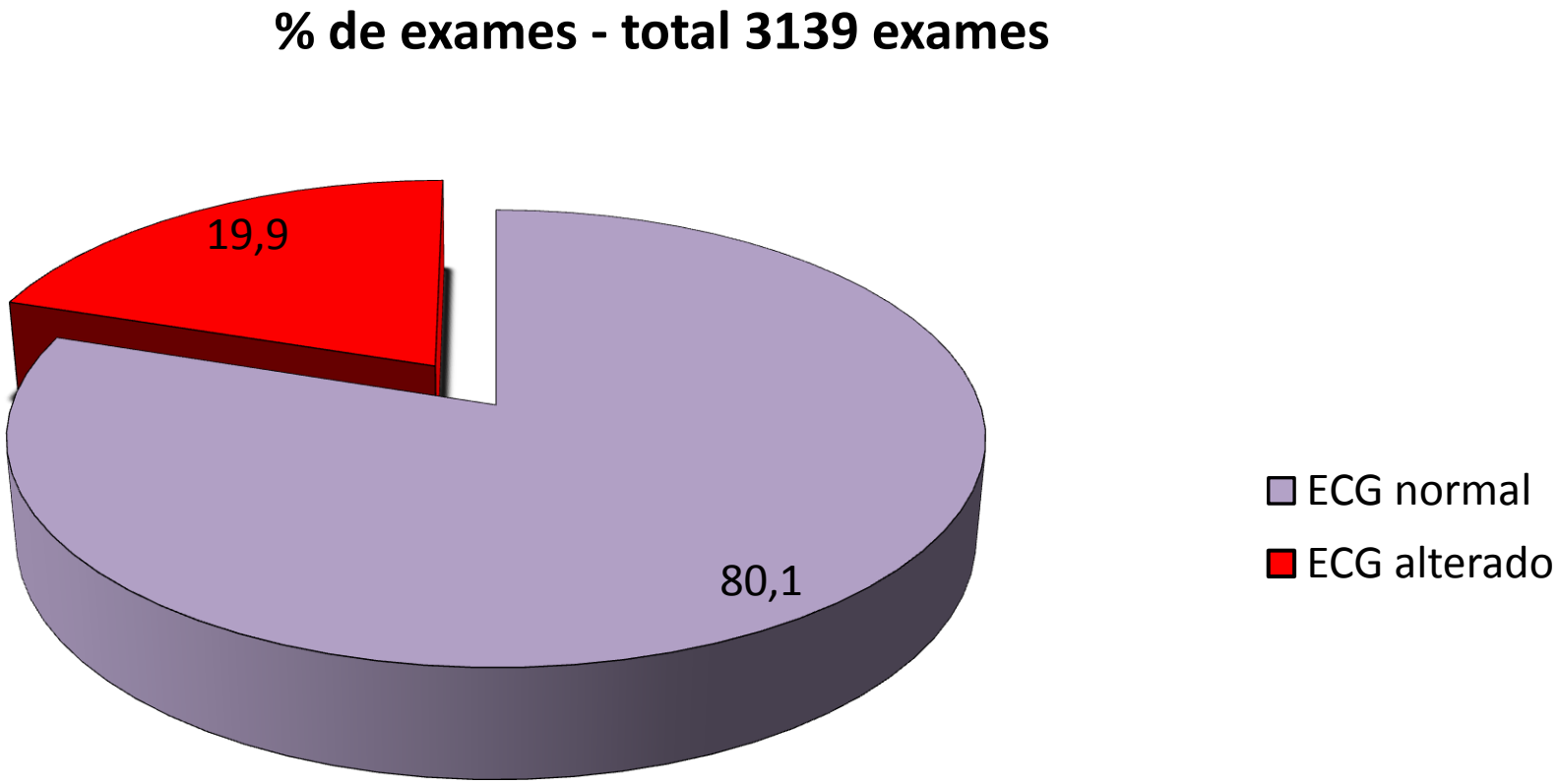
Marcadores ECG na população pediátrica e em jovens

Julho 2007 – Dezembro 2010



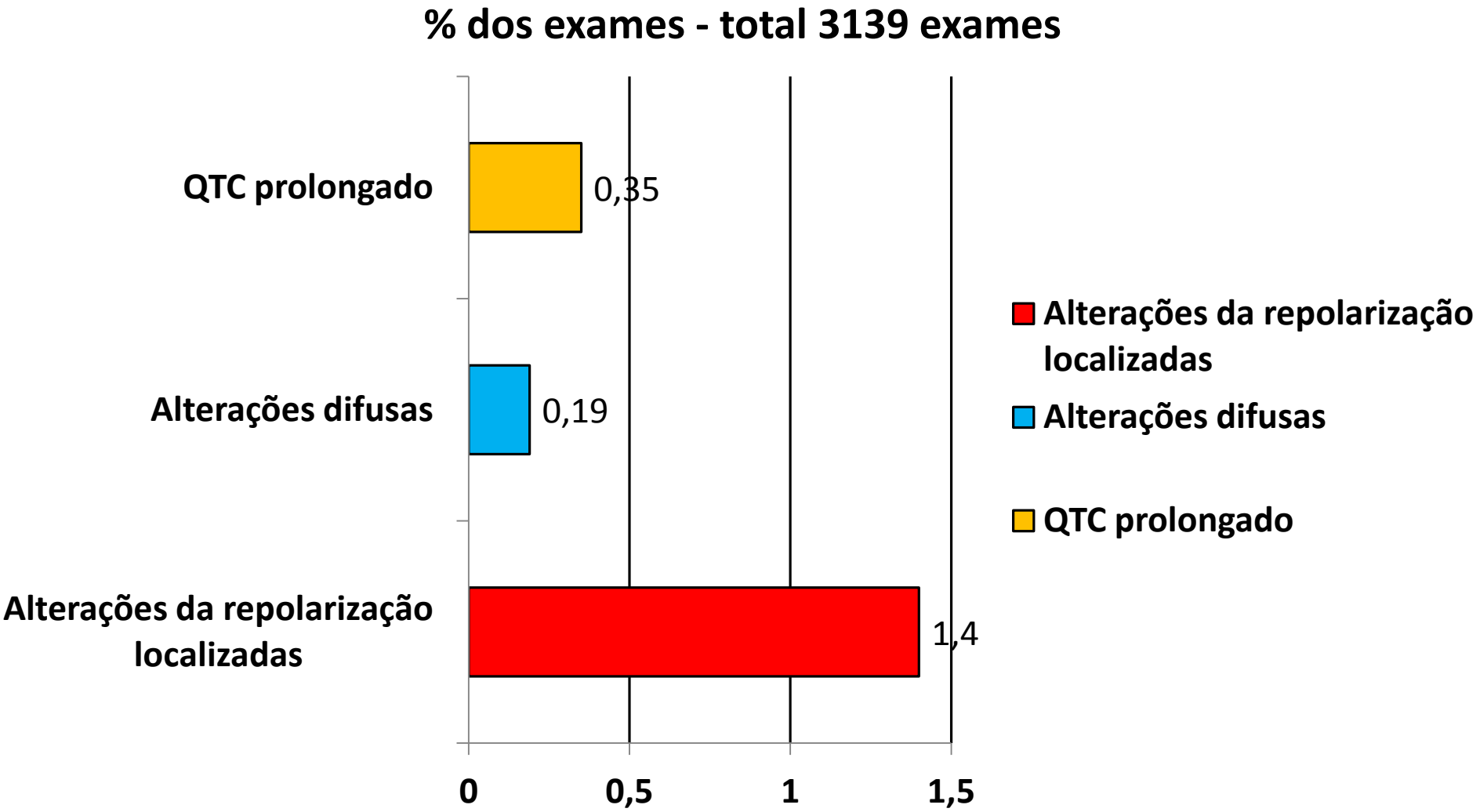
Andalaft RB, Almeida C, Fuziy M . Diagnósticos de ECG na população pediátrica com o uso de um sistema de Tele ECG. Relampa 2011; 24(4):360

Perfil geral dos exames em crianças 0 a 10 anos



Andalaft RB, Almeida C, Fuziy M . Diagnósticos de ECG na população pediátrica com o uso de um sistema de Tele ECG. Relampa 2011; 24(4):360

Análise da repolarização ventricular



Andalaft RB, Almeida C, Fuziy M . Diagnósticos de ECG na população pediátrica com o uso de um sistema de Tele ECG. Relampa 2011; 24(4):360

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

.

Rogerio Andalaft, MARIANA FUZIY, 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

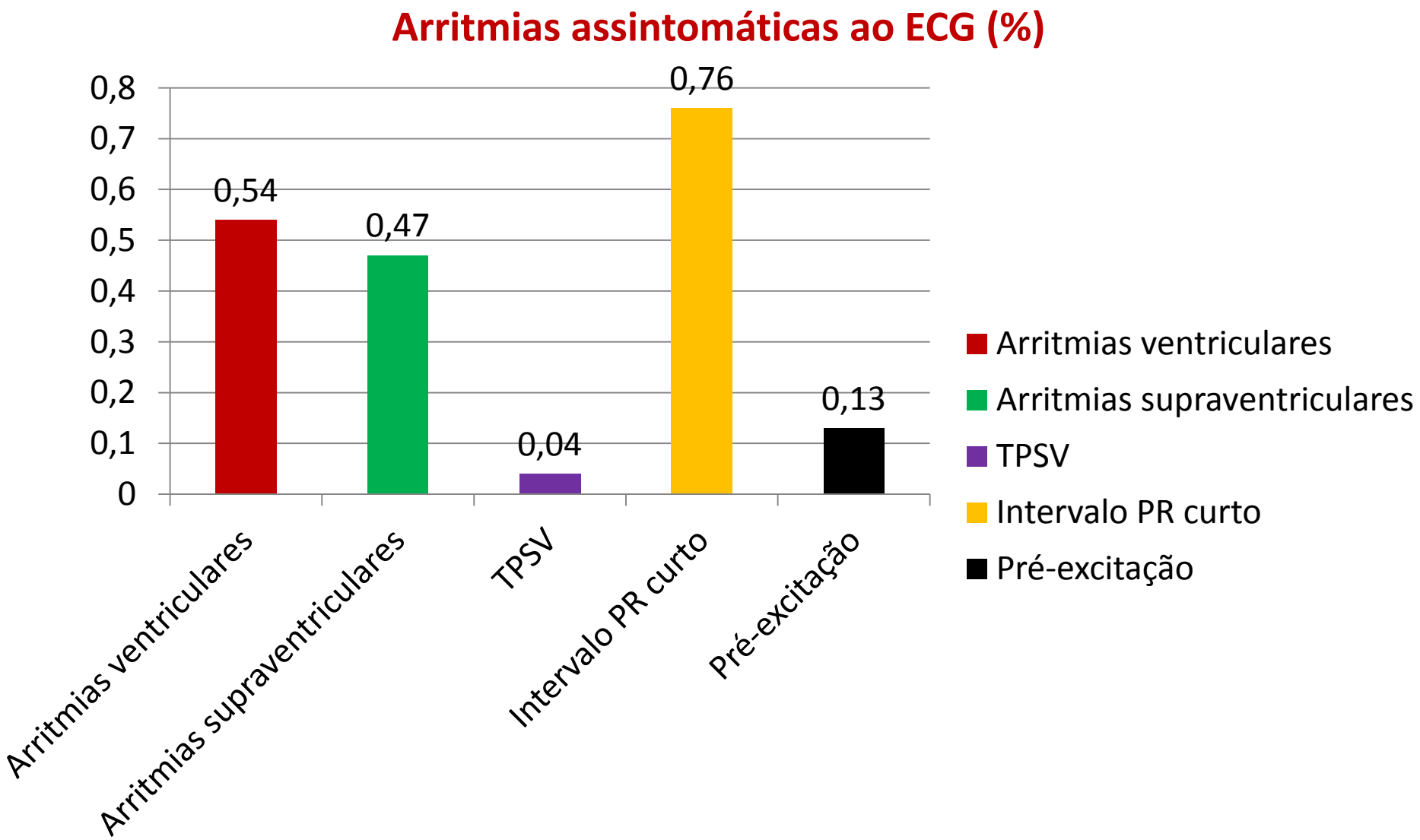
Instituto Dante Pazzanese de Cardiologia

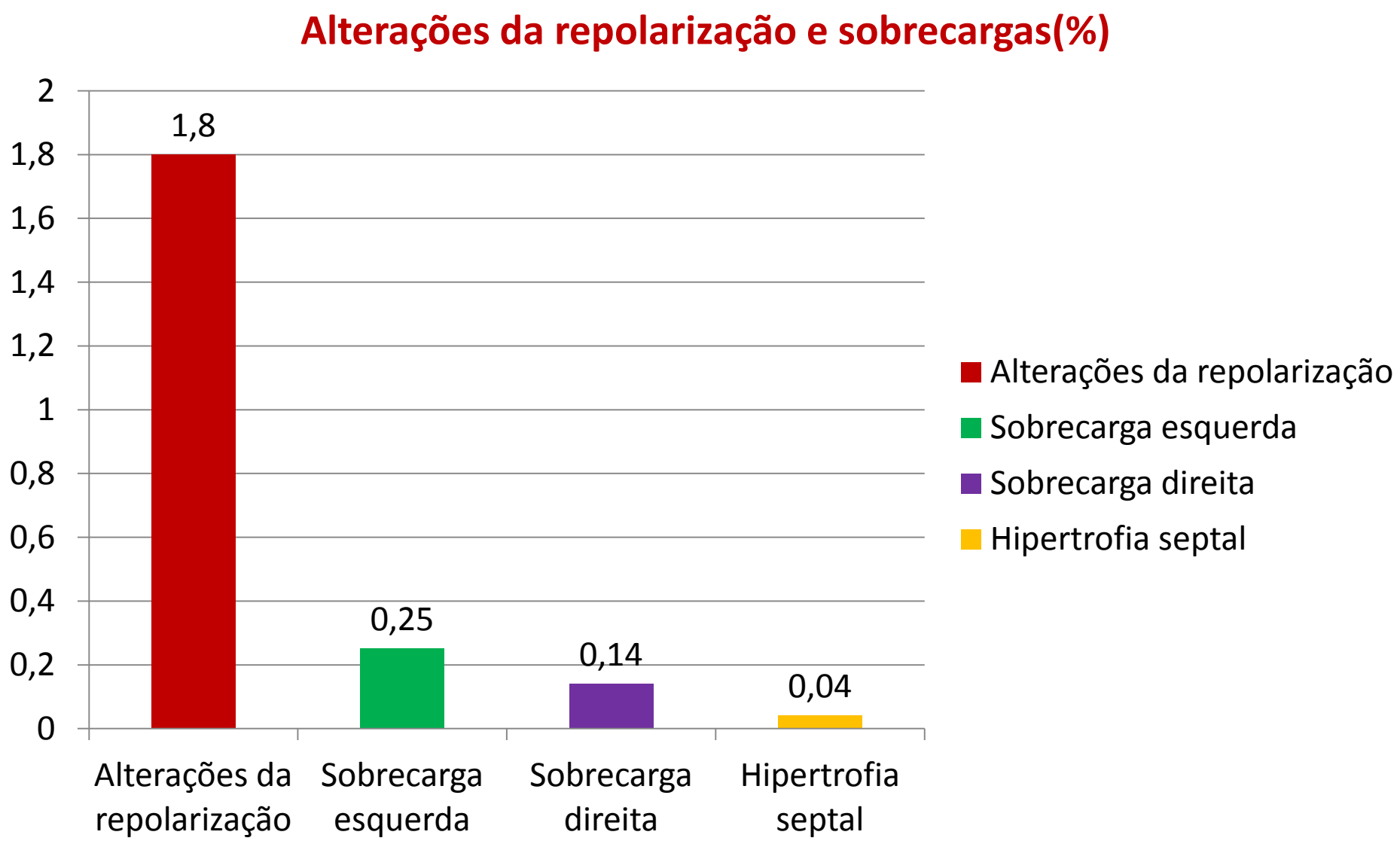
Material

- Foram analisados 11058 pacientes (55% sexo masculino)

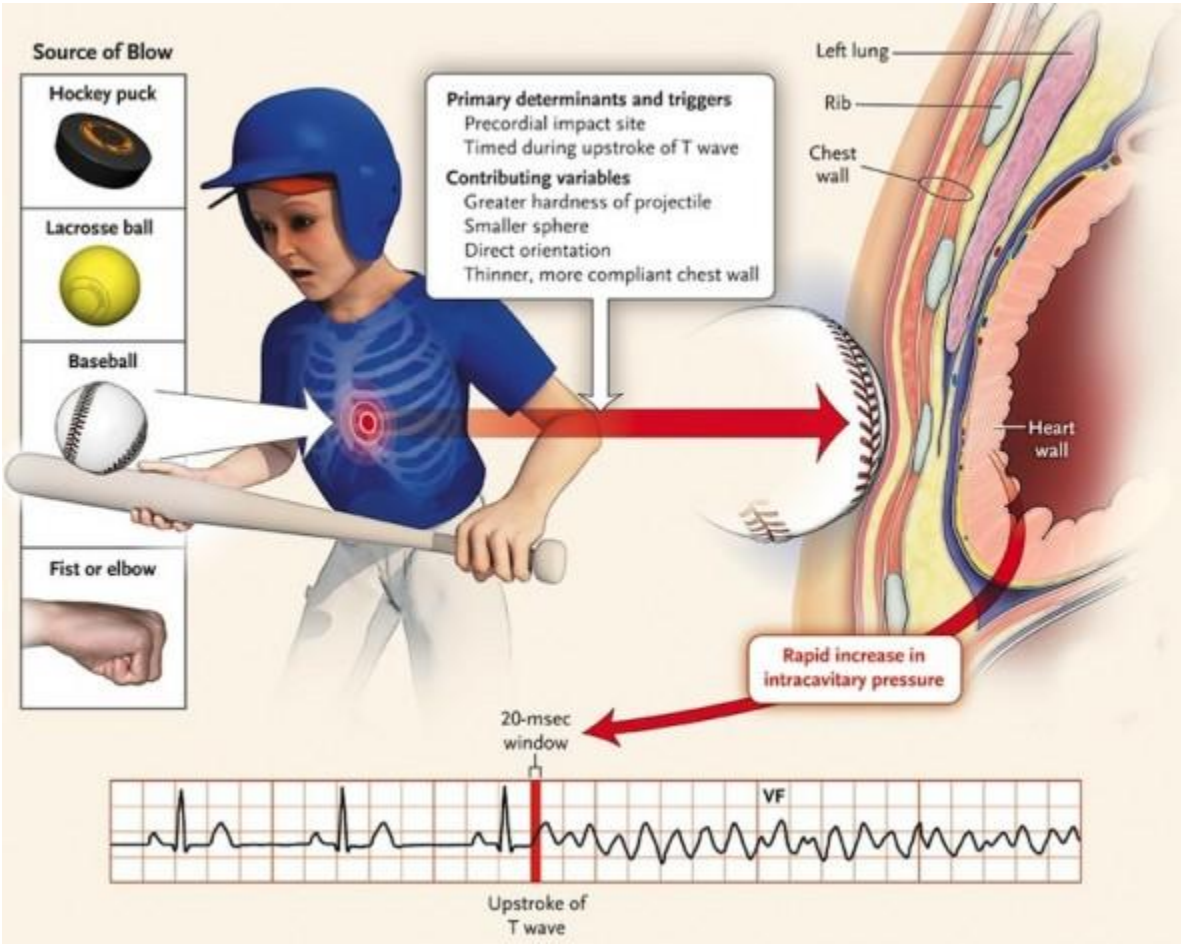
- Idade entre 10 e 20 anos de idade
-
- Base de dados de 797115 p

- Período de análise entre julho de 2007 e agosto 2014

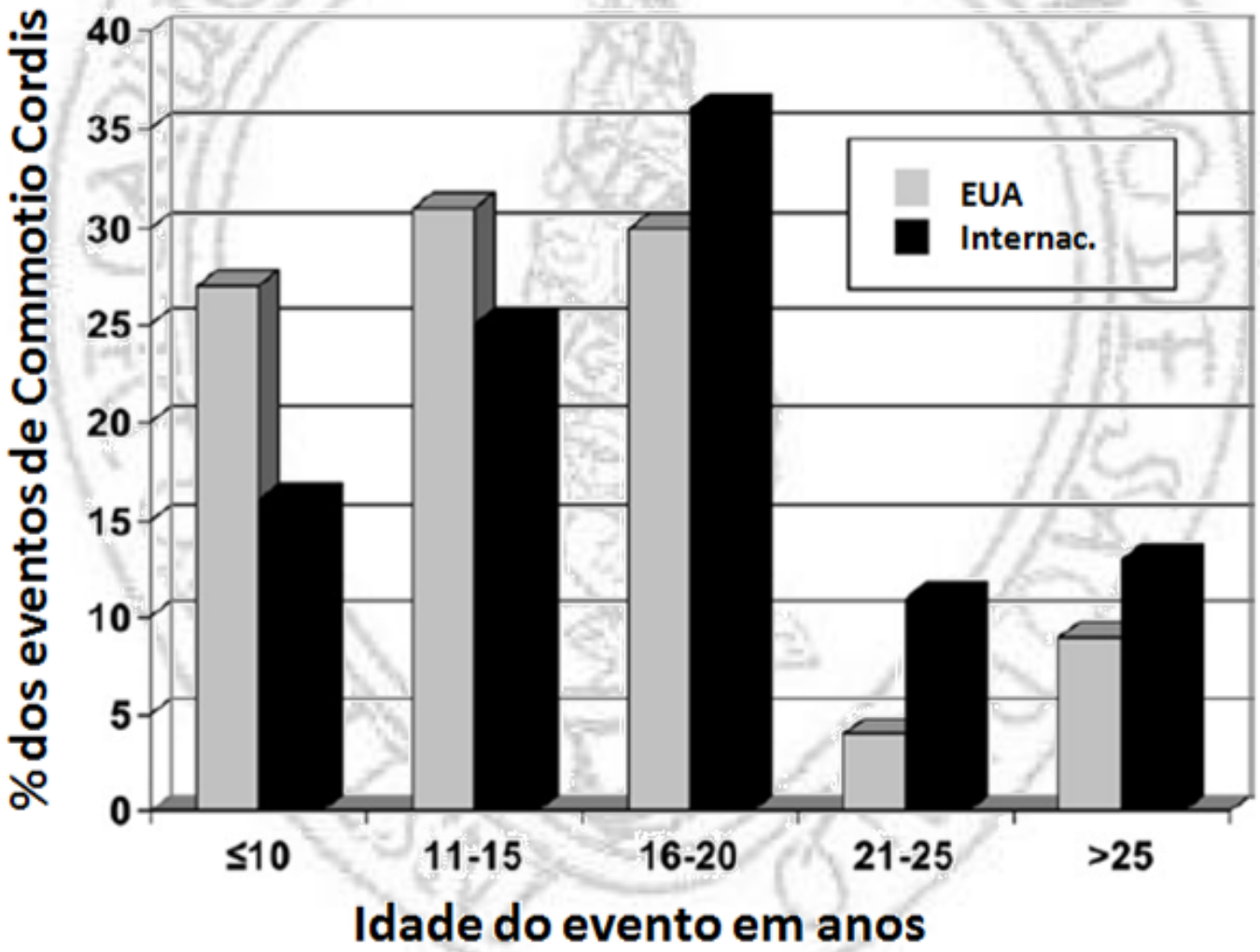

Arritmias assintomáticas ao ECG (%)
0,8
0,7
0,6
0,5
0,4
0,3
0,2
0,1
0
0,54
0,47
0,04
0,76
0,13
Arritmias ventriculares
Arritmias supraventriculares
TPSV
Intervalo PR curto
Pré-excitação
Arritmias ventriculares
Arritmias supraventriculares
TPSV
Intervalo PR curto
Pré-excitação


Alterações da repolarização e sobrecargas(%)
2
1,8
1,6
1,4
1,2
1
0,8
0,6
0,4
0,2
0
1,8
0,25
0,14
0,04
Alterações da repolarização
Sobrecarga esquerda
Sobrecarga direita
Hipertrofia septal
Alterações da repolarização
Sobrecarga esquerda
Sobrecarga direita
Hipertrofia septal

Commotio cordis



Distribuição Commotio cordis por idade



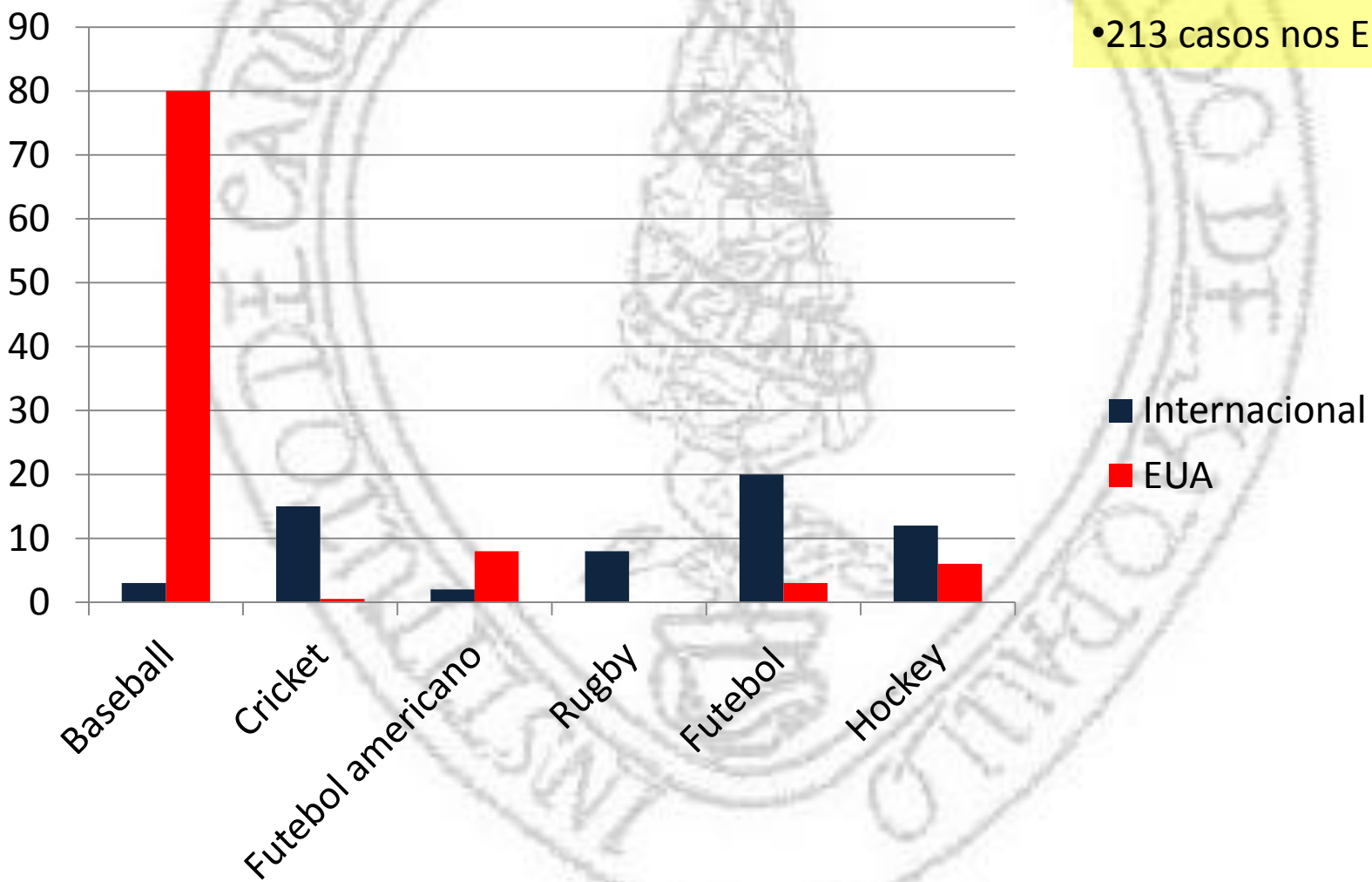
(Heart Rhythm 2011;8:1969 –1971)

Global epidemiology and demographics of commotio cordis

Barry J. Maron, MD,* Aneesha Ahluwalia*, Tammy S. Haas, RN,* Christopher Semsarian, MBBS, PhD,†
Mark S. Link, MD, FHRS,‡ N. A. Mark Estes, III, MD, FHRS‡

*From The Hypertrophic Cardiomyopathy Center, Minneapolis Heart Institute Foundation, Minneapolis, Minnesota;
†Anges Ginges Centre for Molecular Cardiology, Centenary Institute, Sydney Medical School, University of Sydney and Department of Cardiology, Royal Prince Alfred Hospital, Camperdown, Australia; and
‡Division of Cardiology, Tufts Medical Center, Boston, Massachusetts.

- 60 casos internacional
- 213 casos nos EUA



(Heart Rhythm 2011;8:1969–1971)

São Paulo
Fevereiro de 2018

Curso de Eletrocardiograma em crianças e cardiopatias congênitas

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 életricas e morte súbita
Diagnóstico de arritmias no pós operatório
O uso do eletro em condições especiais

Informações e inscrições:
www.arritmiaonline.com.br

pagseguro

Contato: sac@arritmiaonline.com.br ou **telefone:** +5511 55752849

www.arritmiaonline.com.br

Obrigado!!!

rogerioandalaft@arritmiaonline.com.br

rogerio.andalaft@einstein.br

ALBERT EINSTEIN
HOSPITAL ISRAELITA

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

AULA DISPONÍVEL EM:

www.arritmiaonline.com.br