



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



Displasia arritmogênica de VD

Rogério Braga Andalaft

Médico do Setor de Eletrofisiologia do Instituto Dante Pazzanese de Cardiologia
Médico do Centro de Arritmias do Hospital Israelita Albert Einstein
Grupo de Eletrofisiologia Pediátrica - GEP

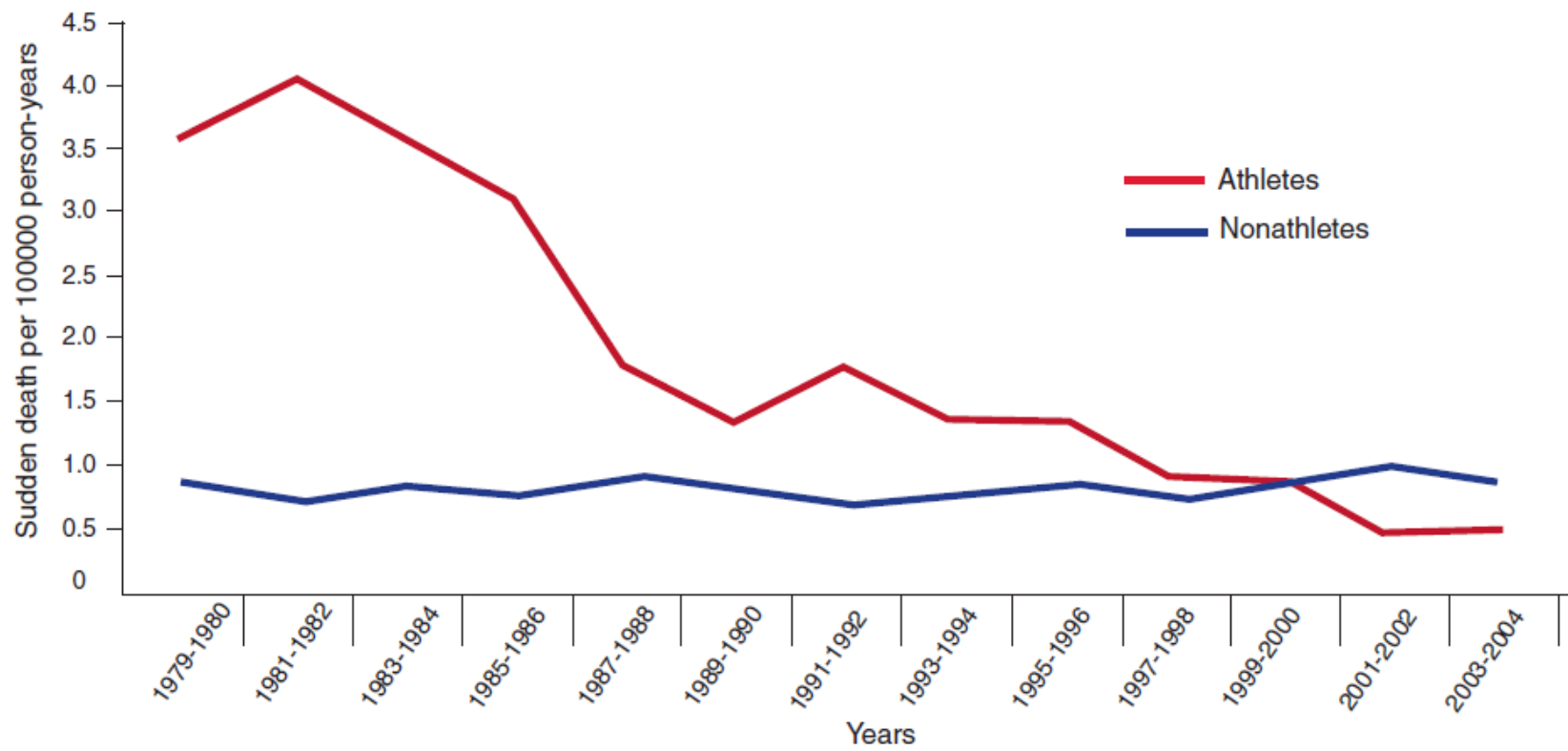
Aspectos gerais

- Doença degenerativa da musculatura miocárdica principalmente do VD
- Substituição fibro gordurosa
- Risco de arritmias e morte súbita
- Maior preocupação na população não síndrômica









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

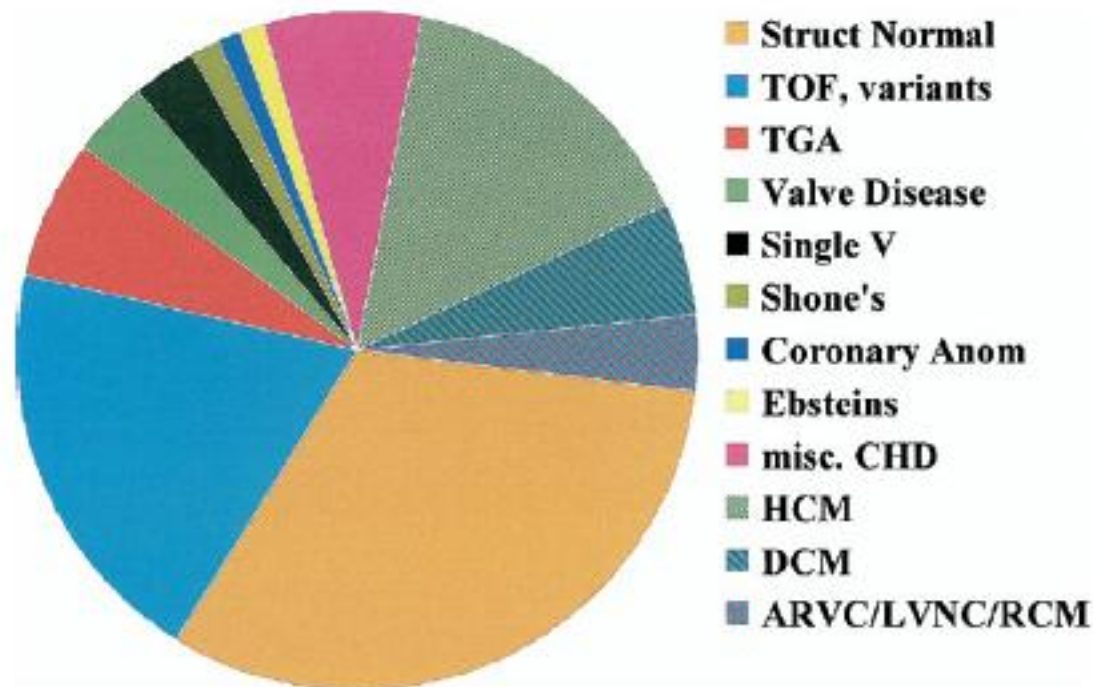
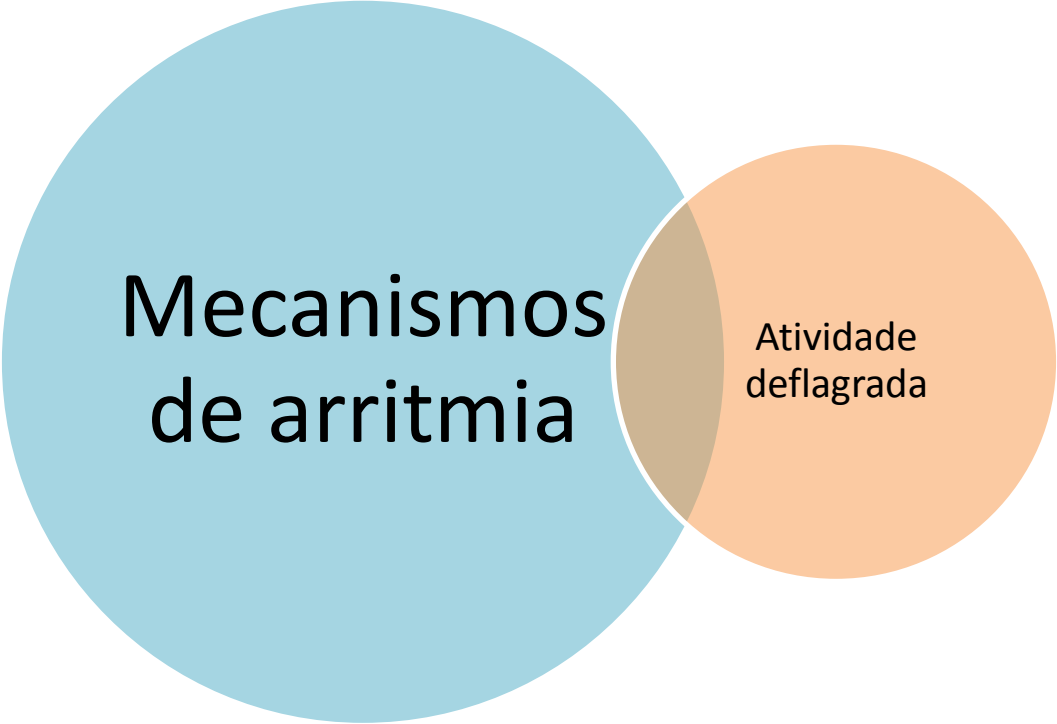


Figure 1

Anatomic Diagnoses of Pediatric and Congenital Implantable Cardioverter-Defibrillator Recipients

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

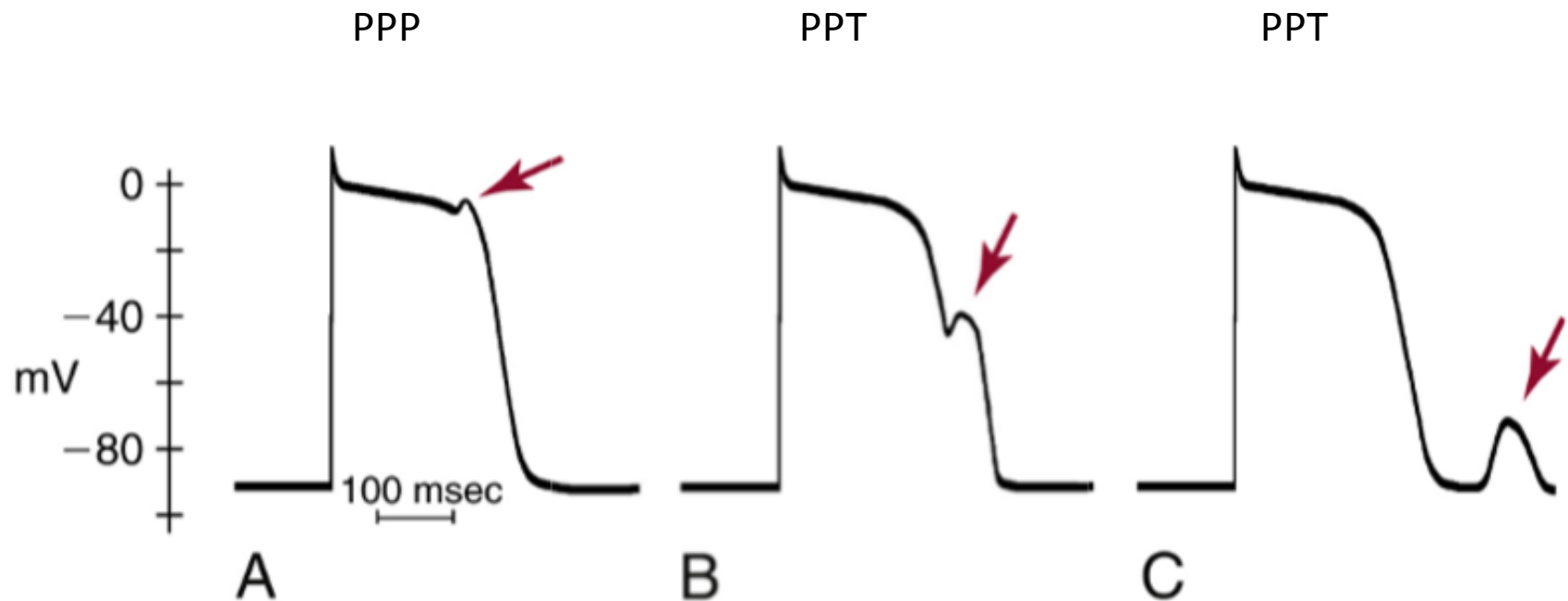
**PRINCIPAL DIAGNÓSTICO
ELETROFISIOLÓGICO COM
ARRITMIA IDIOPÁTICA DE VD**



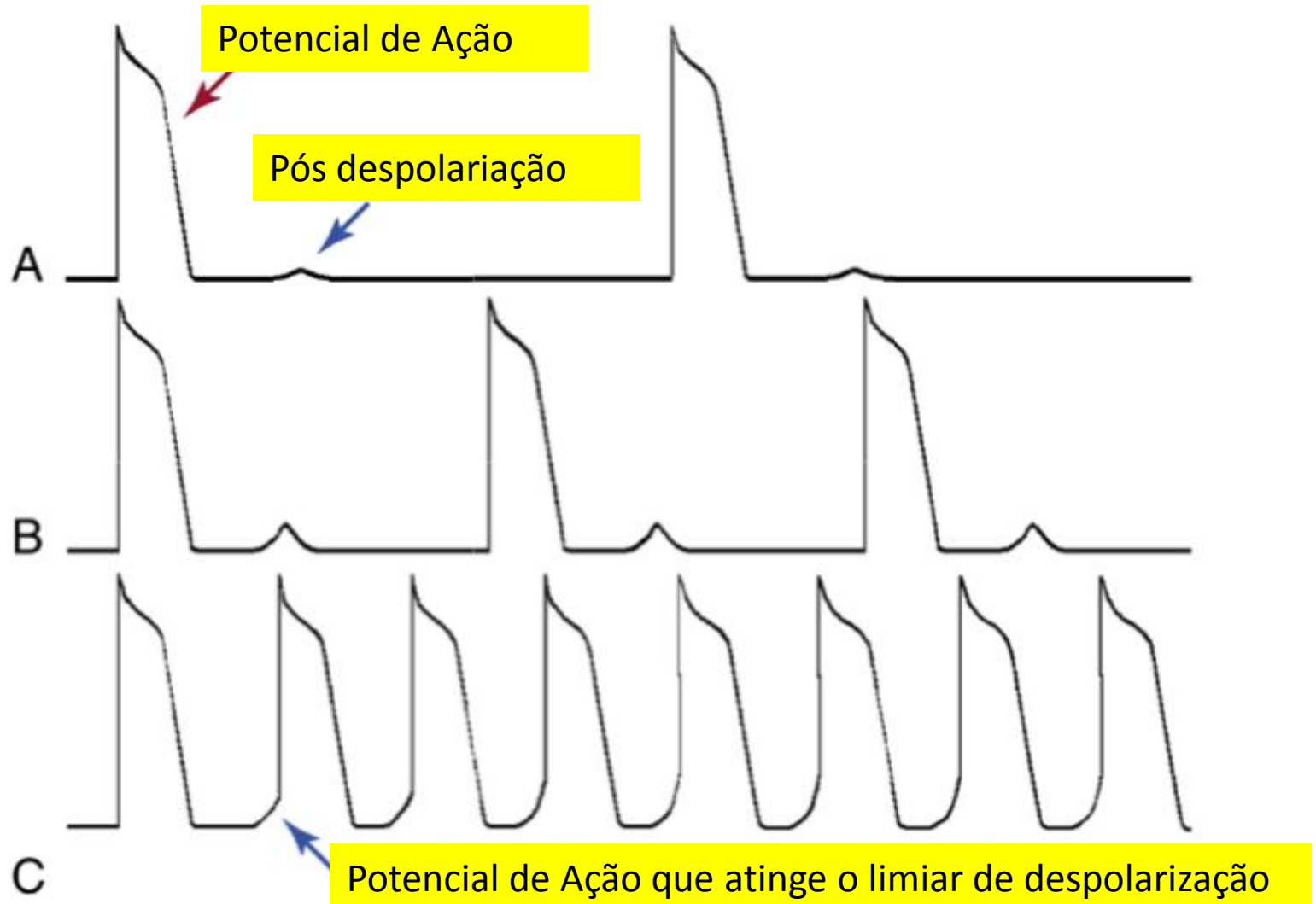
Mecanismos
de arritmia

Atividade
deflagrada

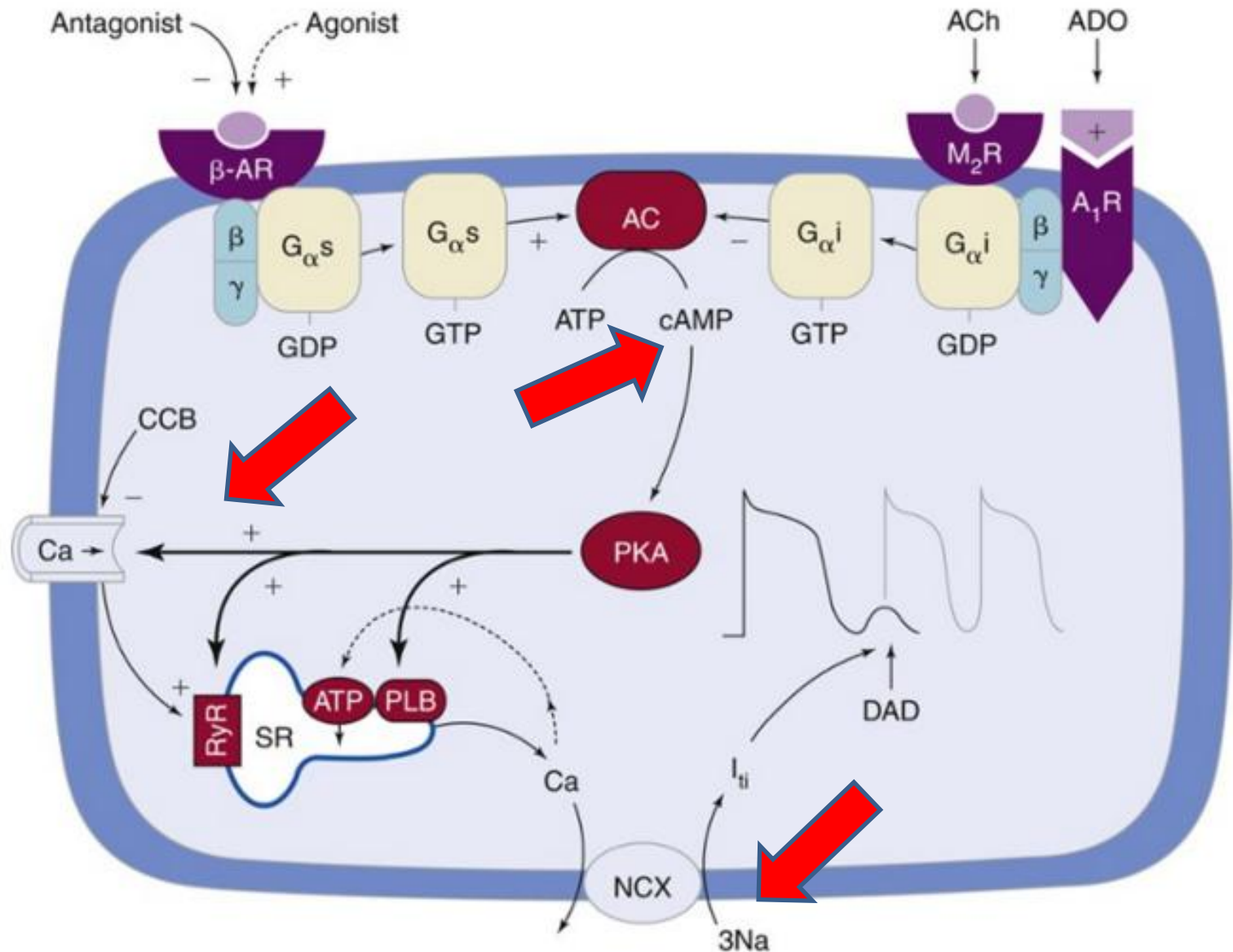
Atividade deflagrada



Pós despolarização e limiar de despolarização

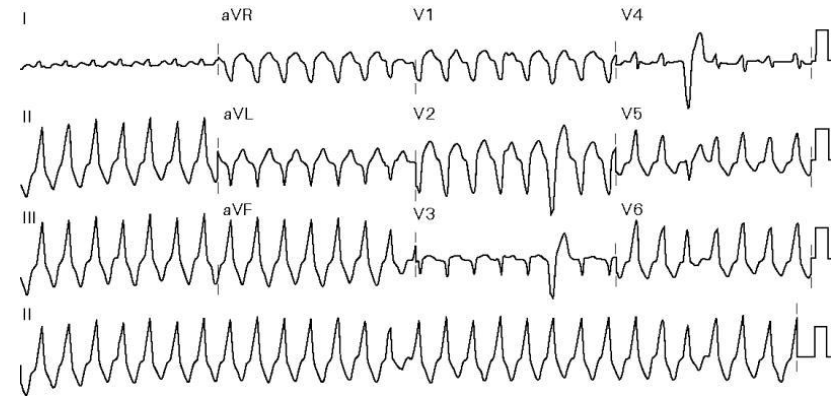


Mecanismo iônico da atividade deflagrada



Coração normal

- TV adenosina sensível
- TV verapamil sensível



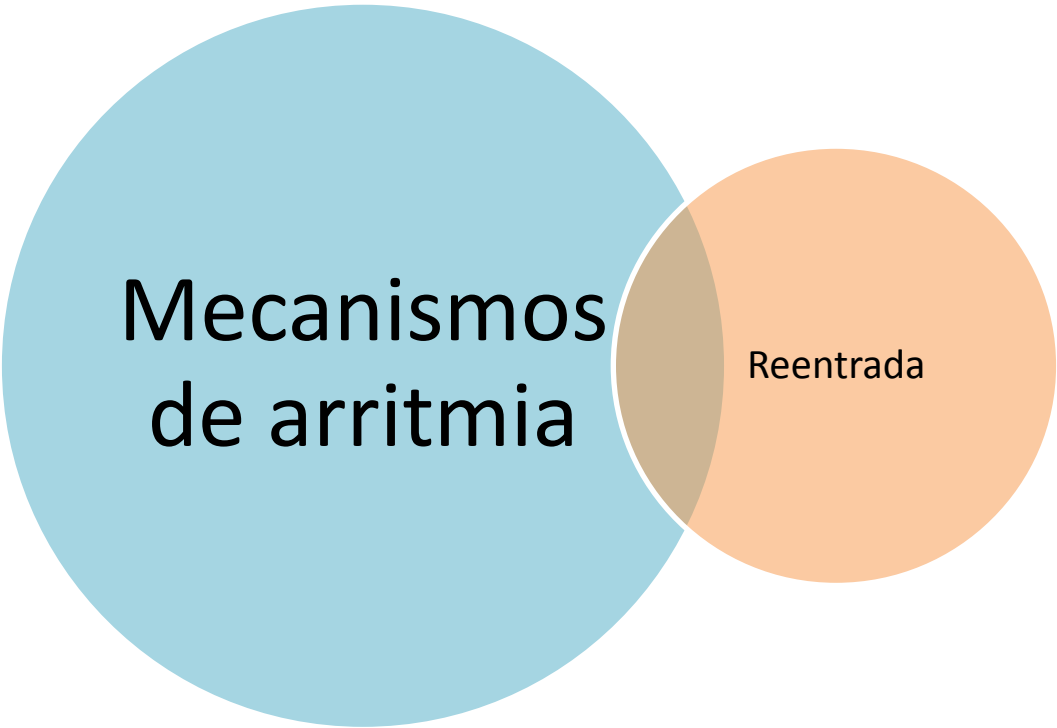
TV com Morfologia
BRE
(principalmente as
de VSVD)

Habitualmente
Sensível a
adenosina

Tratamento crônico
Betabloqueador
Bloqueador canal
de Ca



**E QUAL O MECANISMO DA
ARRITMIA NA DISPLASIA
ARRITMOGÊNICA DE VD**



A Venn diagram consisting of two overlapping circles. The left circle is light blue and contains the text 'Mecanismos de arritmia'. The right circle is light orange and contains the text 'Reentrada'. The overlapping area in the center is a brownish-tan color.

**Mecanismos
de arritmia**

Reentrada

Tipos de reentrada



Reentrada anatômica

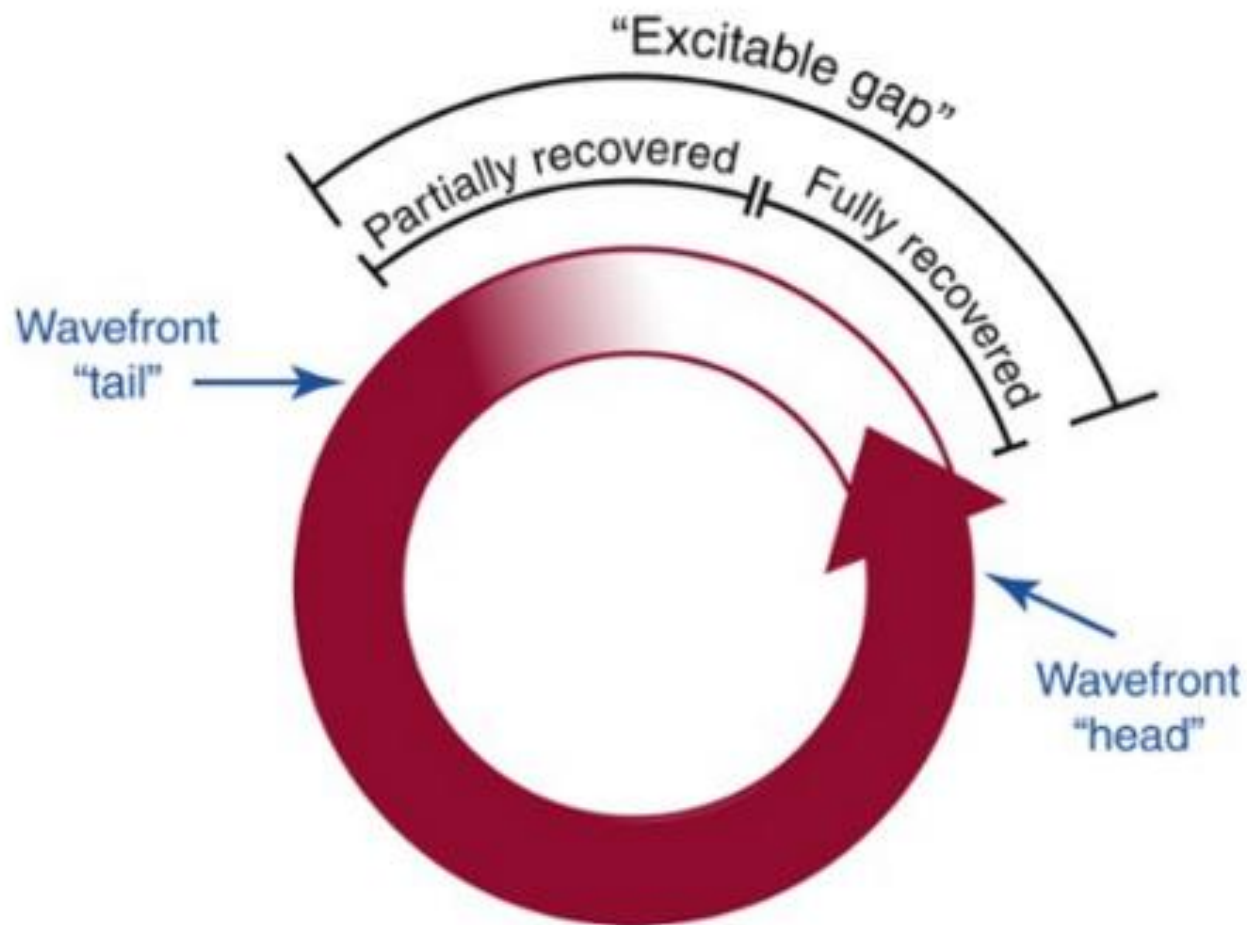


Reentrada funcional



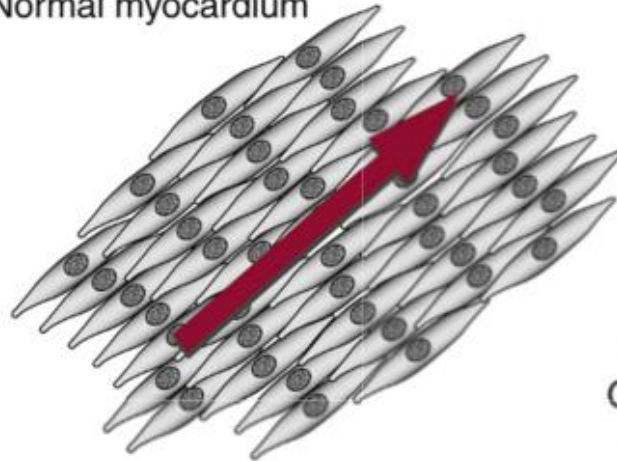
Reentrada deflectida

Gap excitável para encarrilhamento



Cicatrices e reentrada

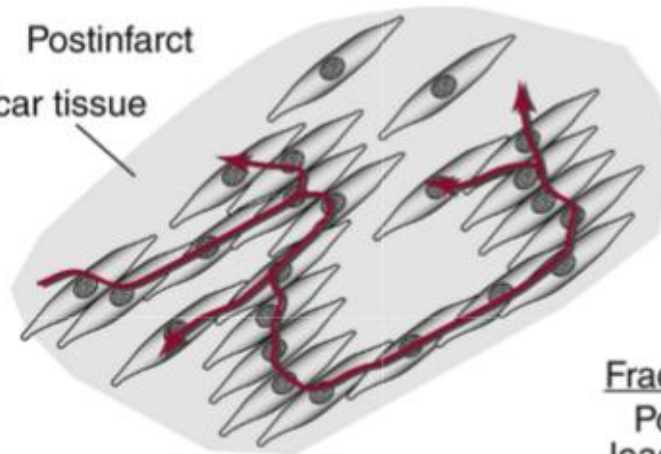
Normal myocardium



Normal electrogram:
Good cell-cell coupling
allows synchronous
depolarization

A

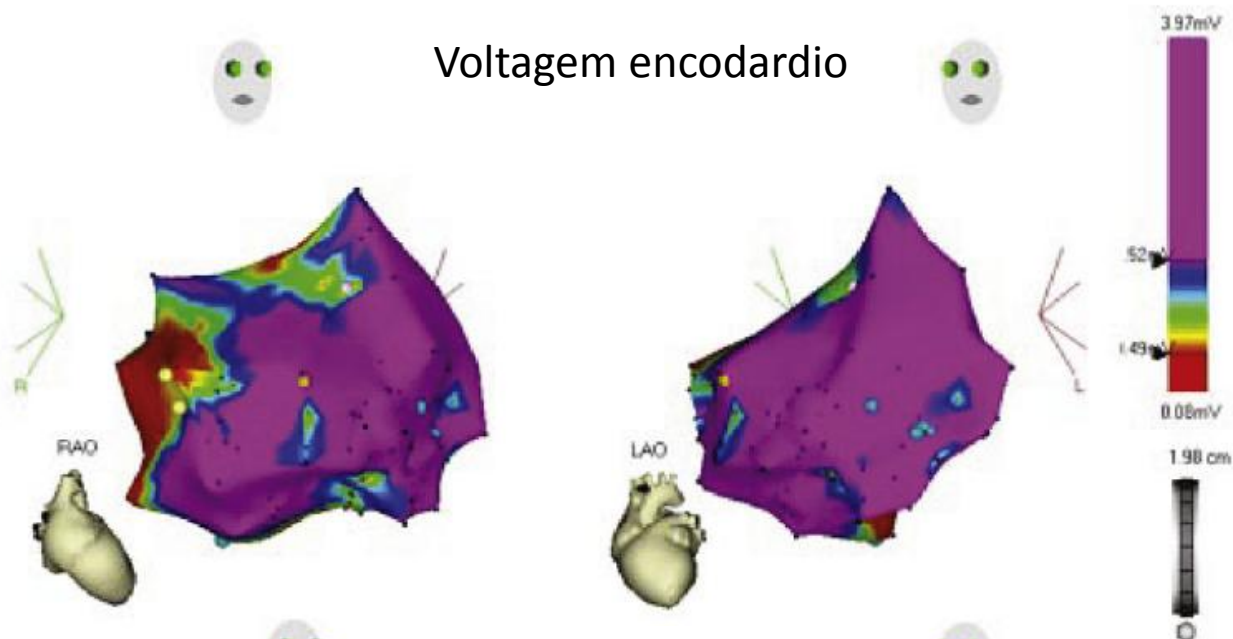
Postinfarct
Scar tissue



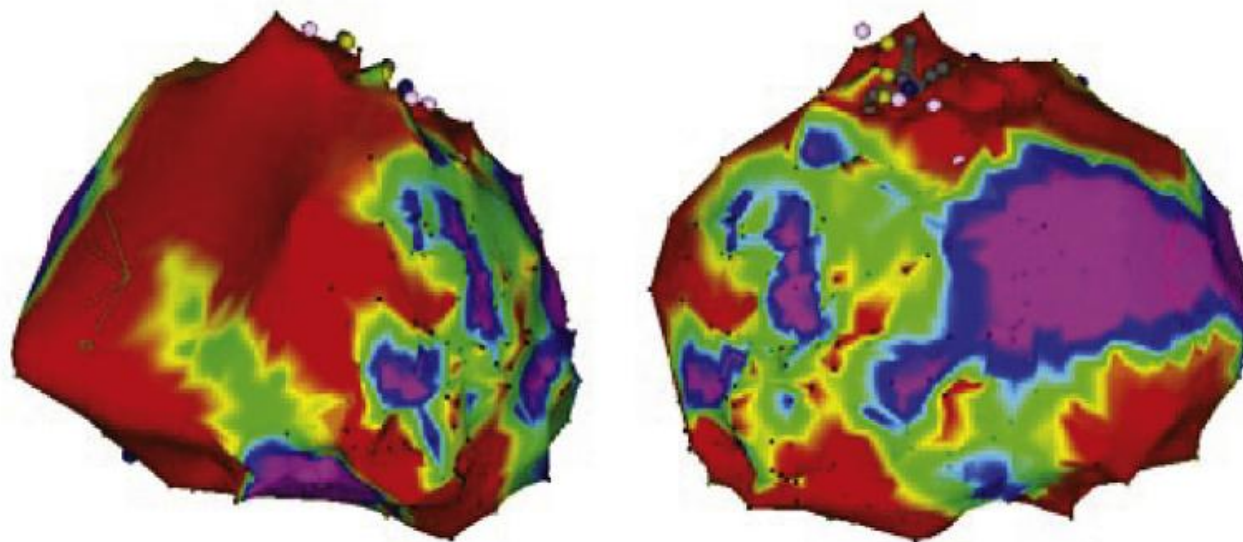
Fractionated electrogram:
Poor cell-cell coupling
leads to dyssynchronous
depolarization

B

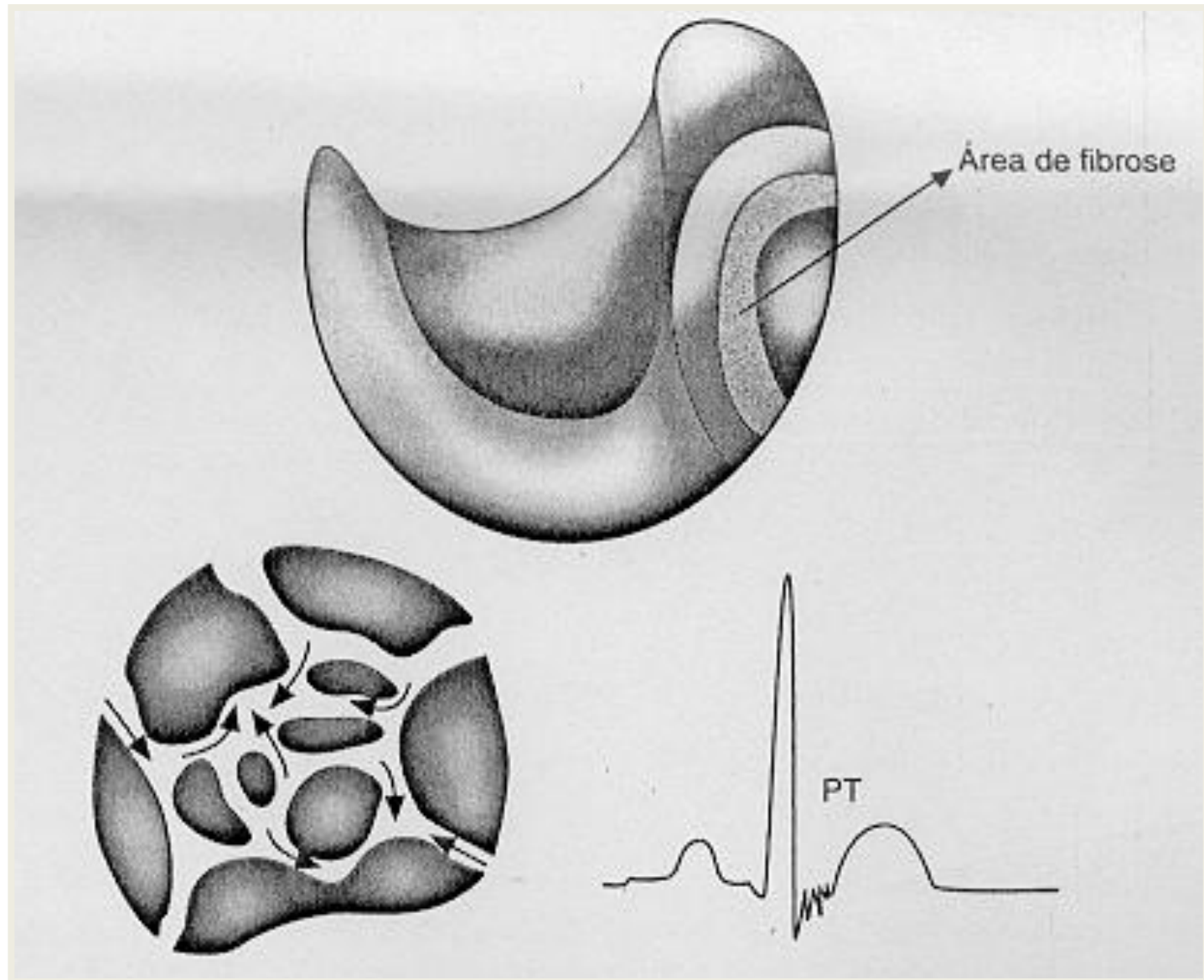
Voltagem encodardio

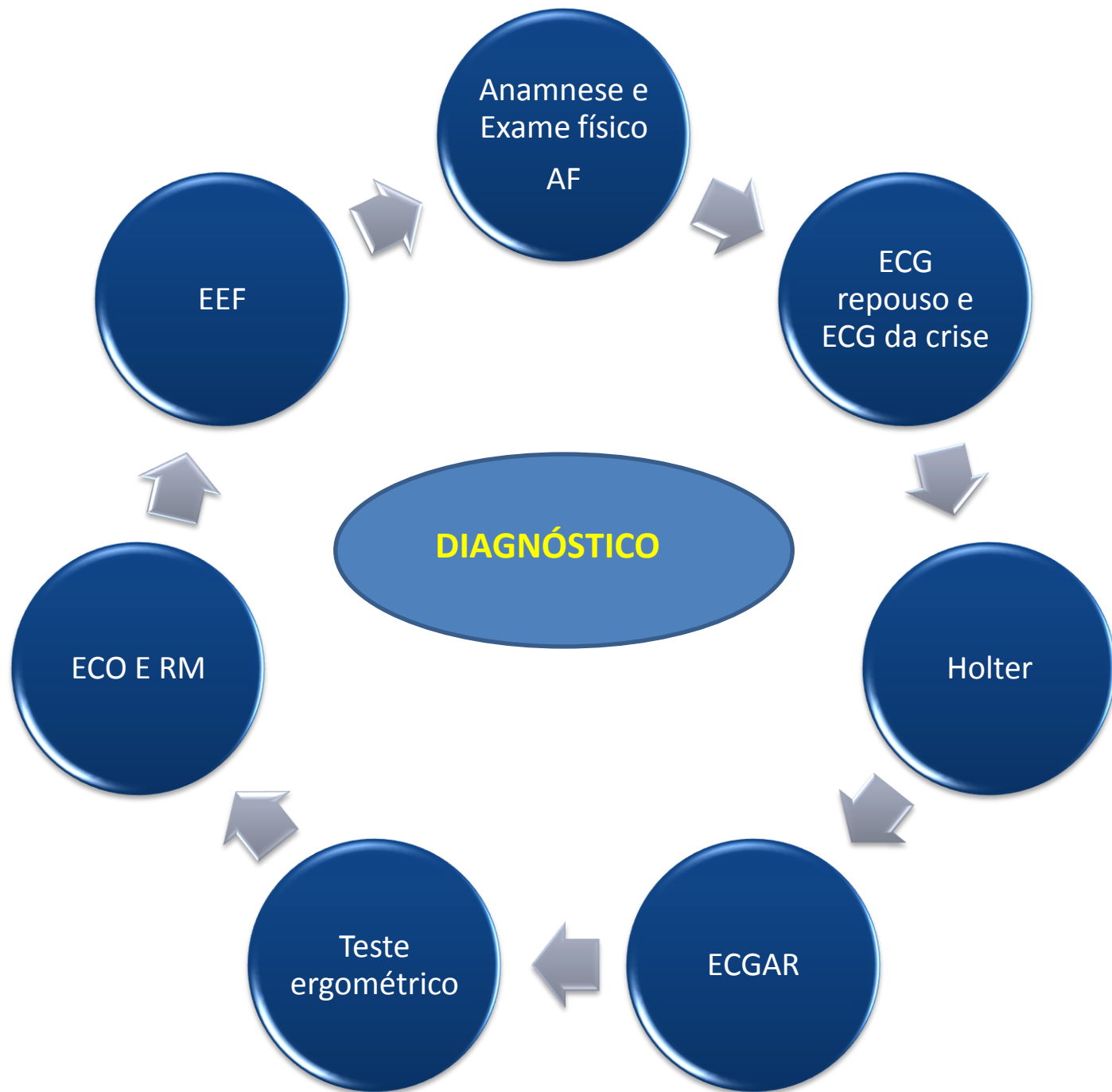


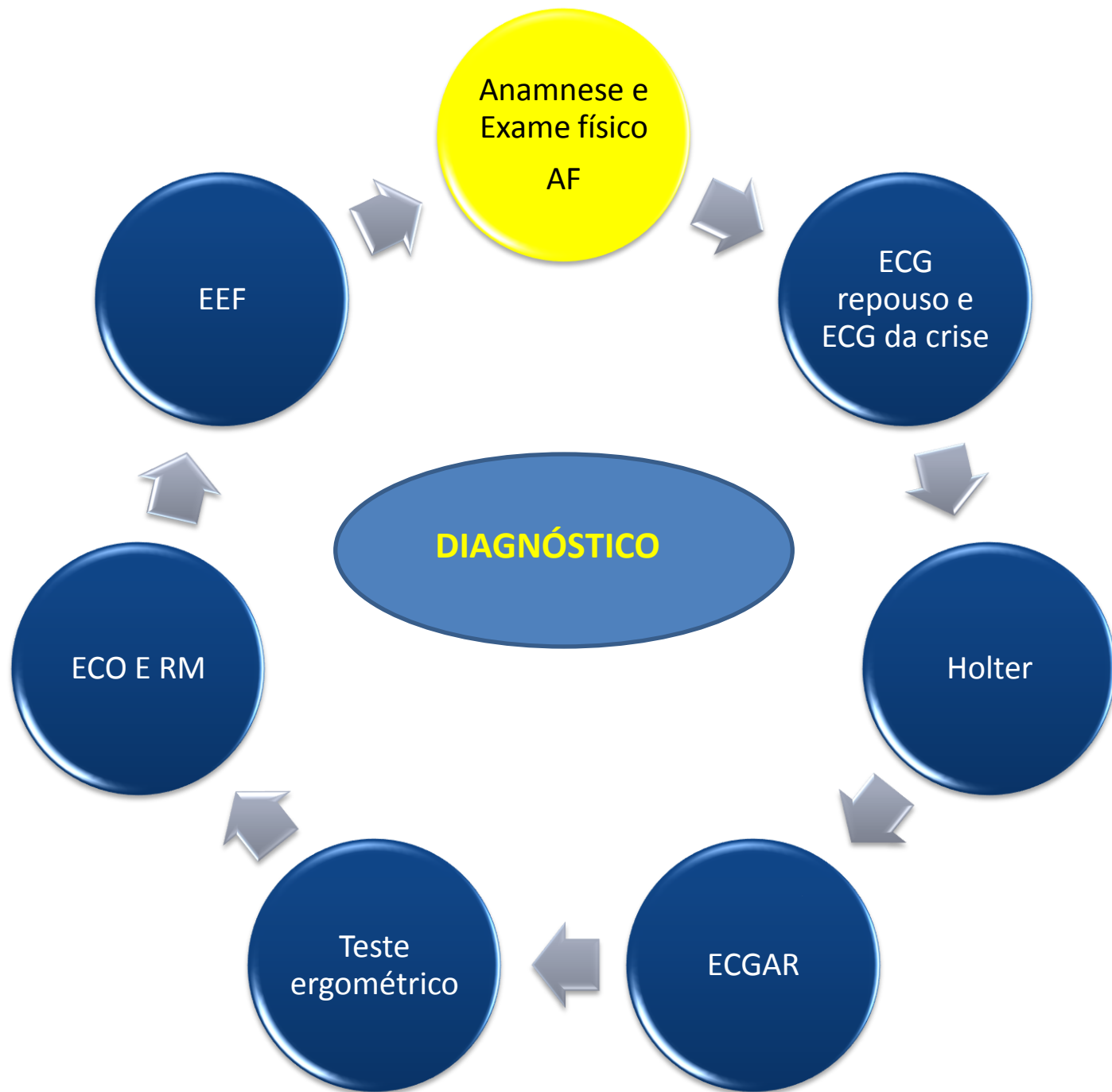
Voltagem epidardio



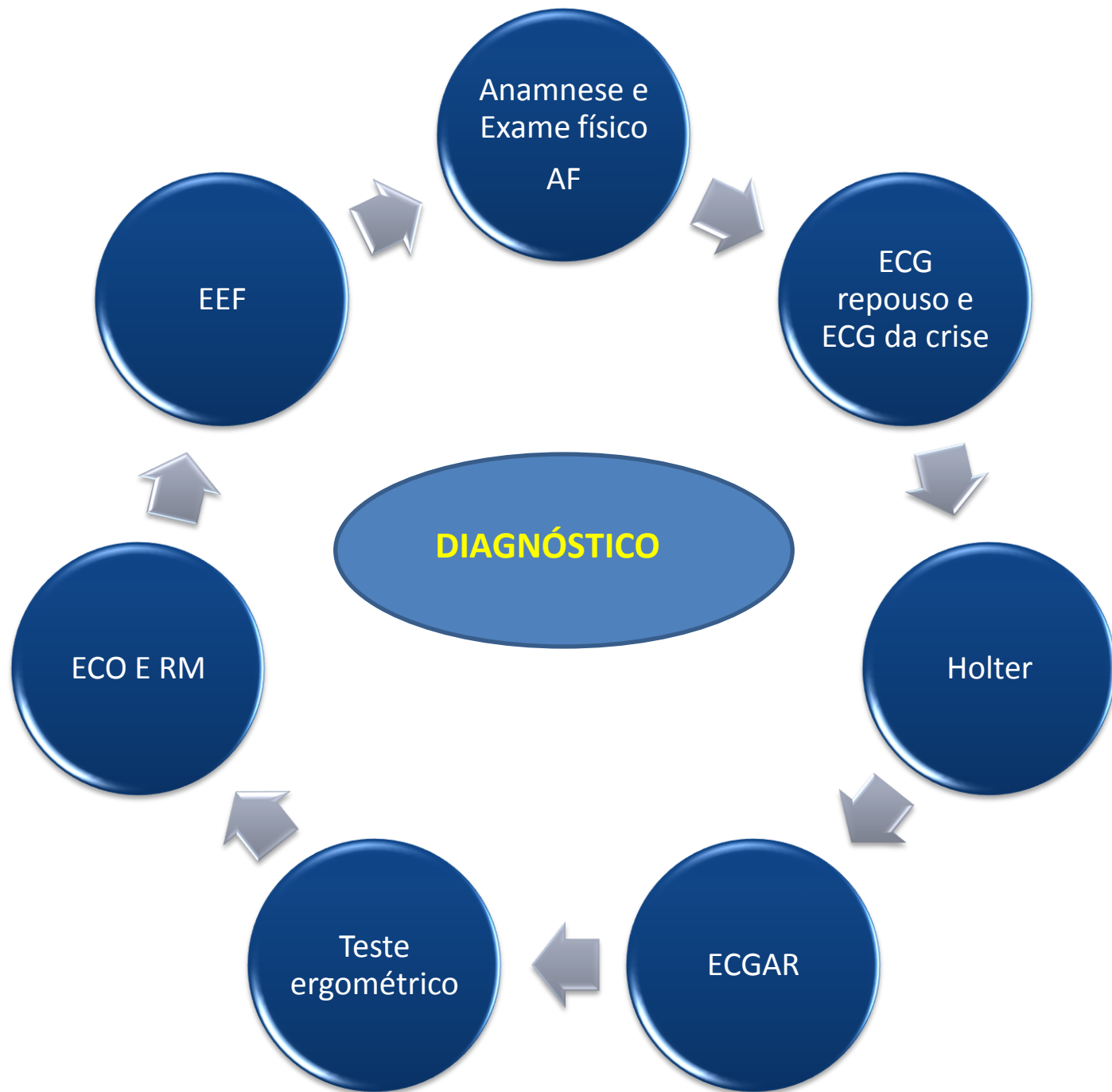
Áreas de Fibrose Entremeadas com Tecido Sadio

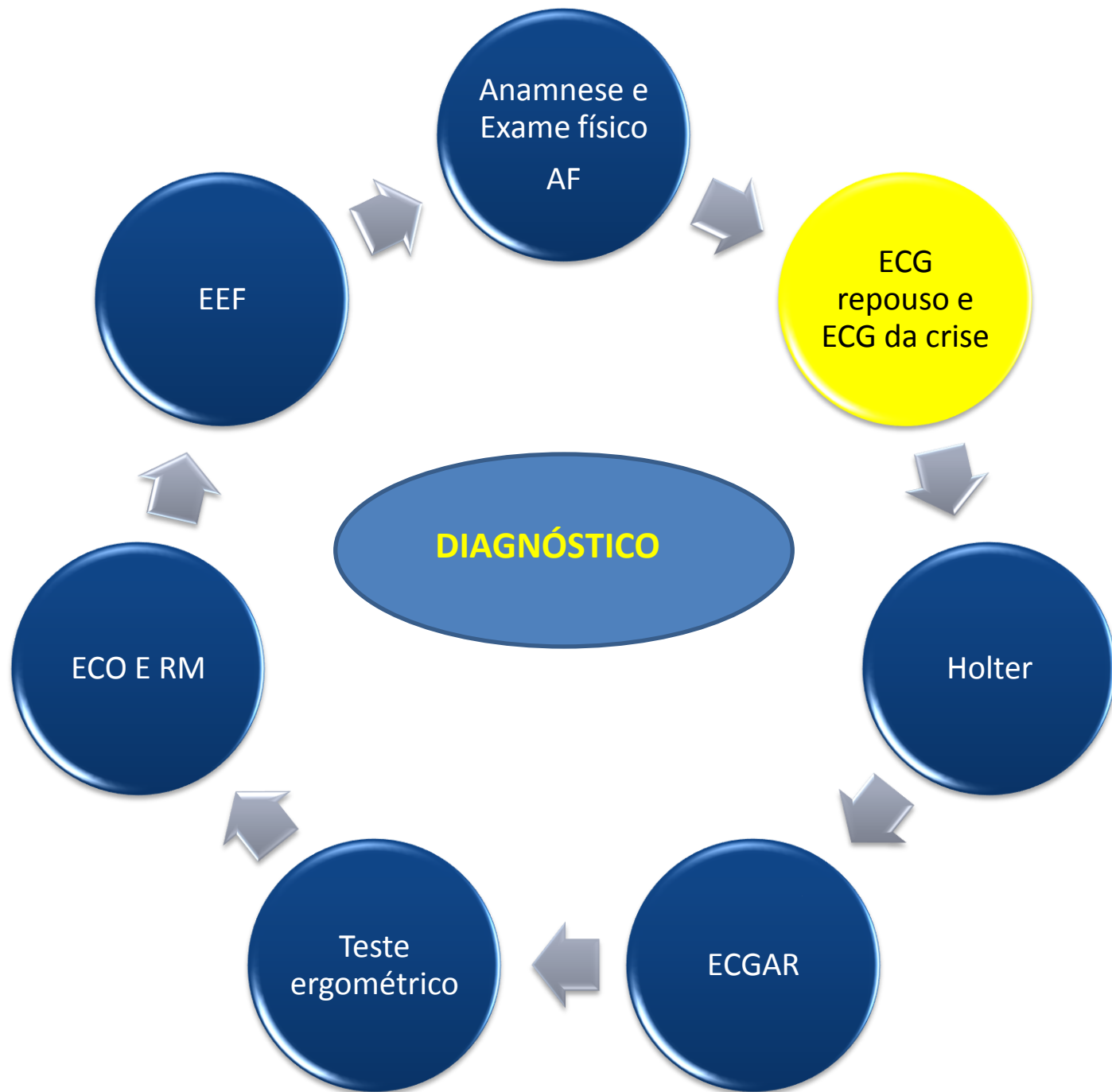




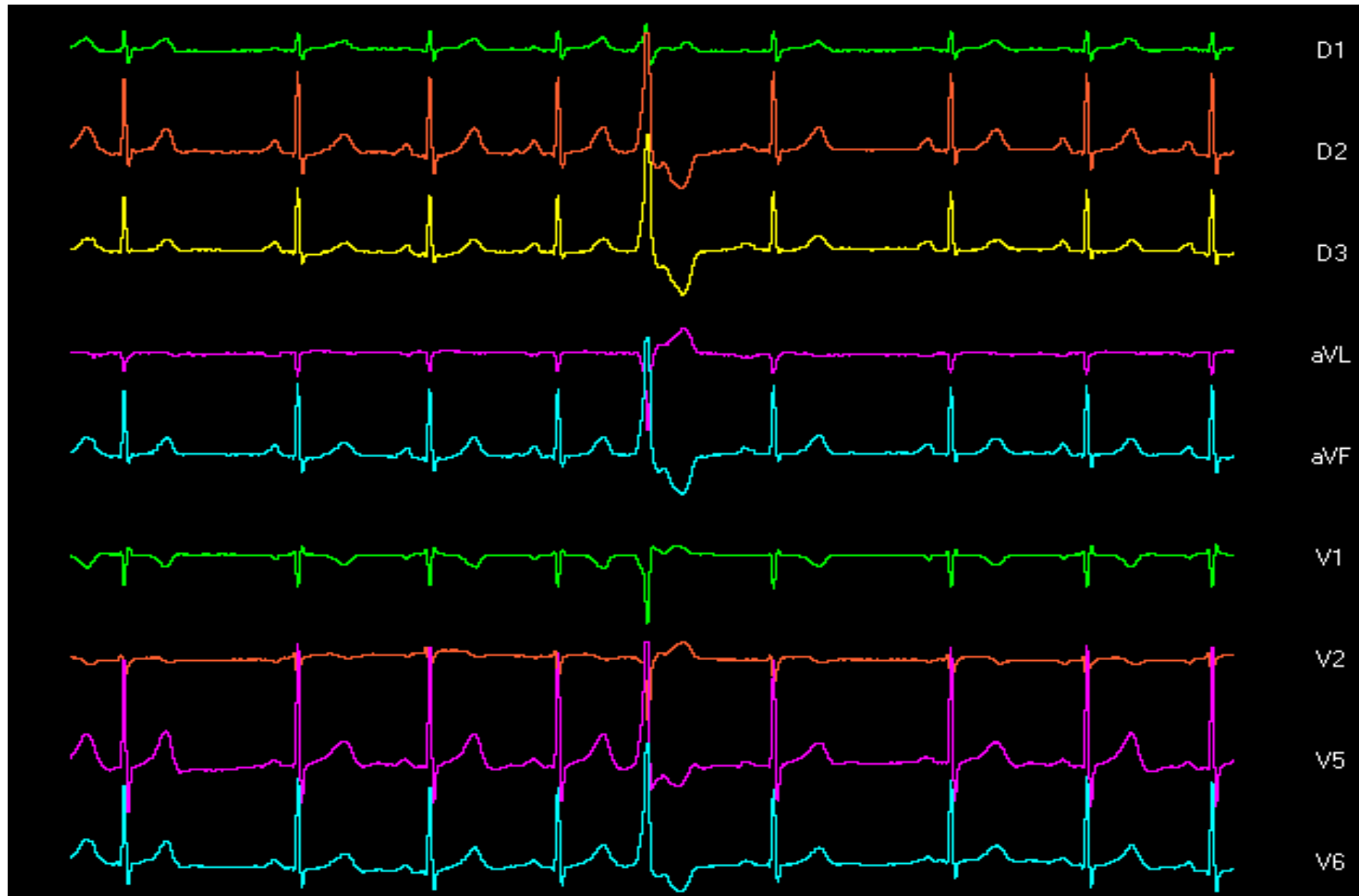


ORIGINAL TASK FORCE CRITERIA		REVISED TASK FORCE CRITERIA
VI. Family History		
Major	Familial disease confirmed at necropsy or surgery	- ARVC/D confirmed in a first-degree relative who meets current task force criteria - ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative - Identification of a pathogenic mutation[†] categorized as associated or probably associated with ARVC/D in the patient under evaluation
Minor	- Family history of premature sudden death (<35 yr of age) due to suspected ARVC/D - Familial history (clinical diagnosis based on present criteria)	- History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current task force criteria - Premature sudden death (<35 yr of age) due to suspected ARVC/D in a first-degree relative - ARVC/D confirmed pathologically or by current task force criteria in second-degree relative

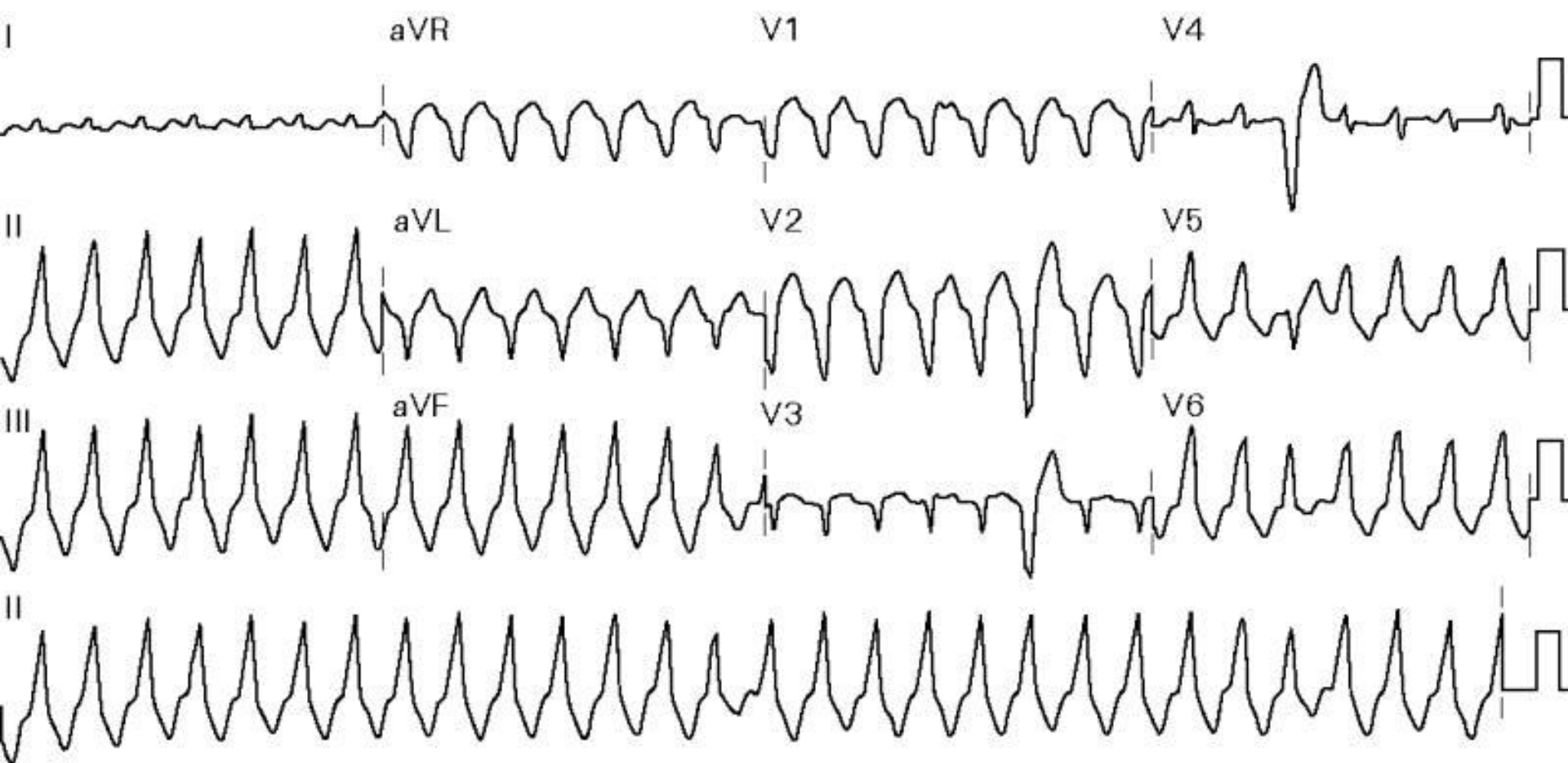


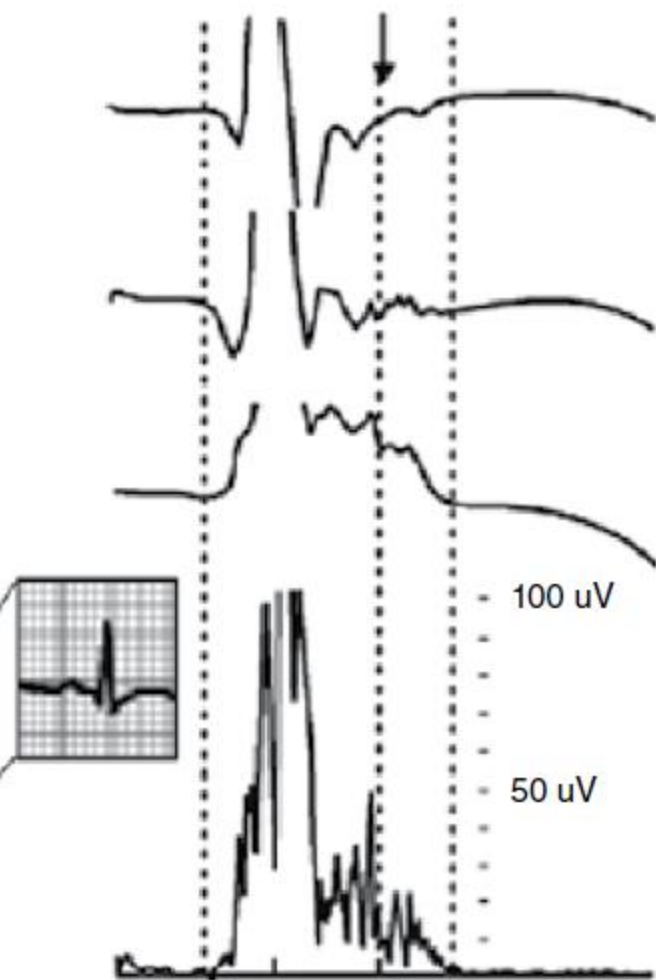


ARRITMIA DE V.D.

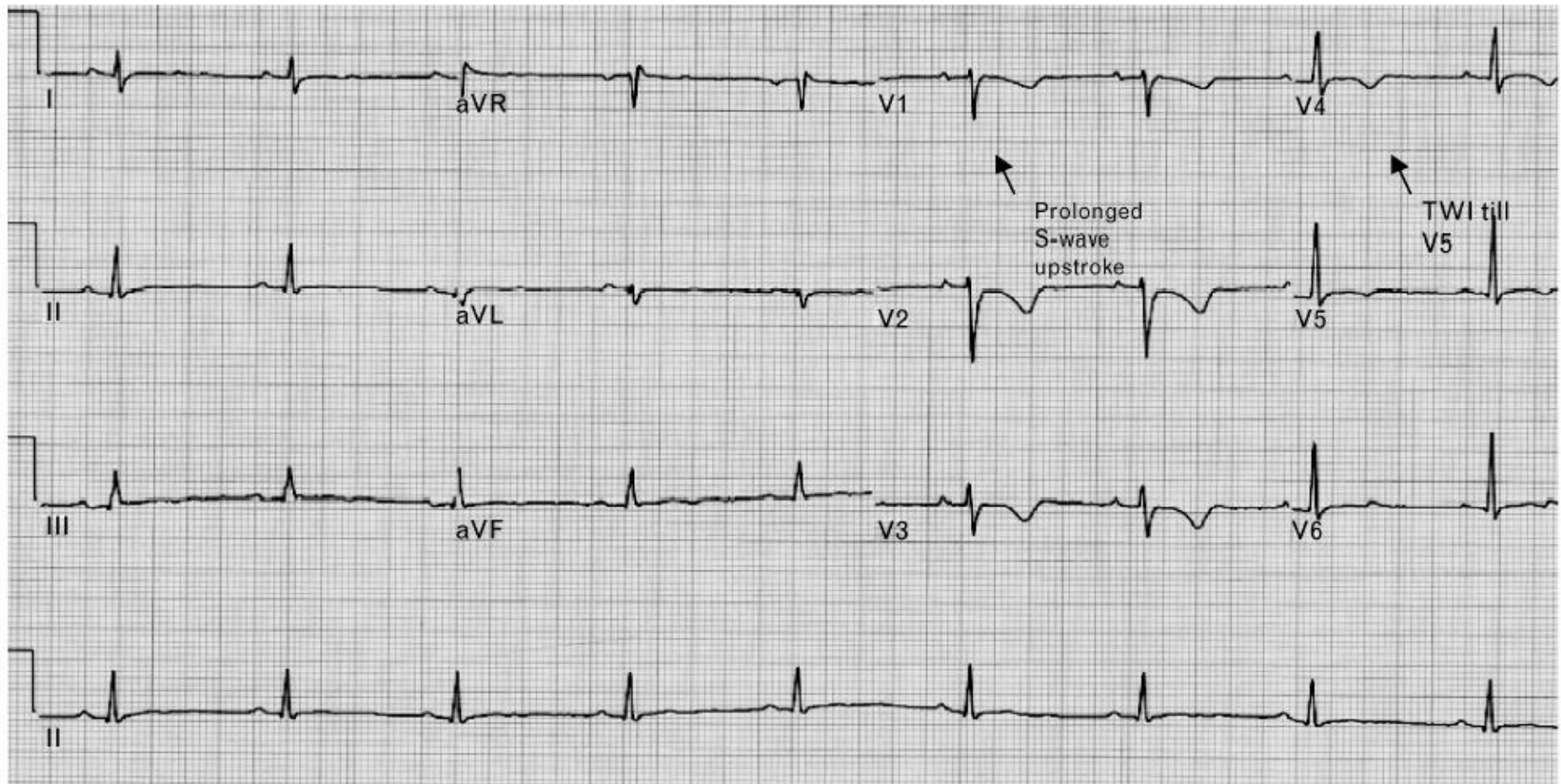
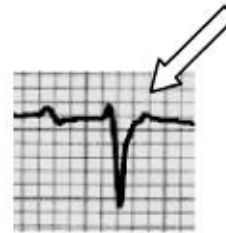


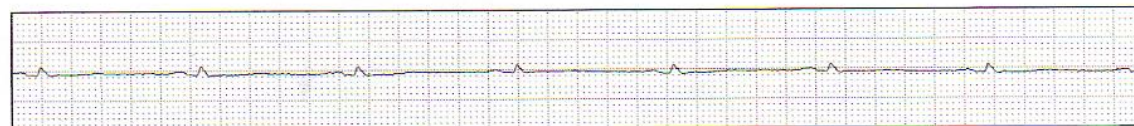
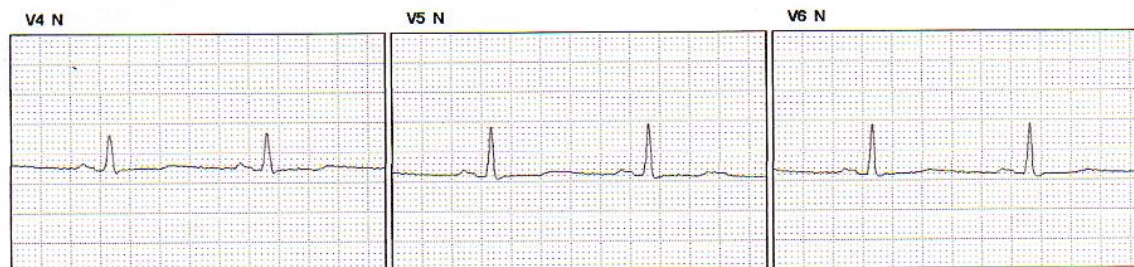
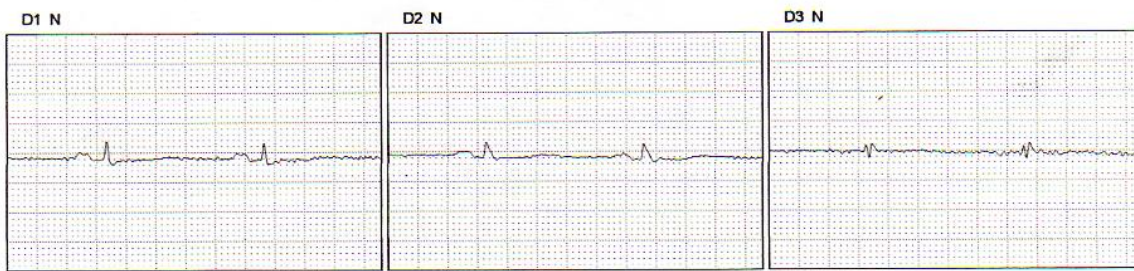
Taquicardia ventricular



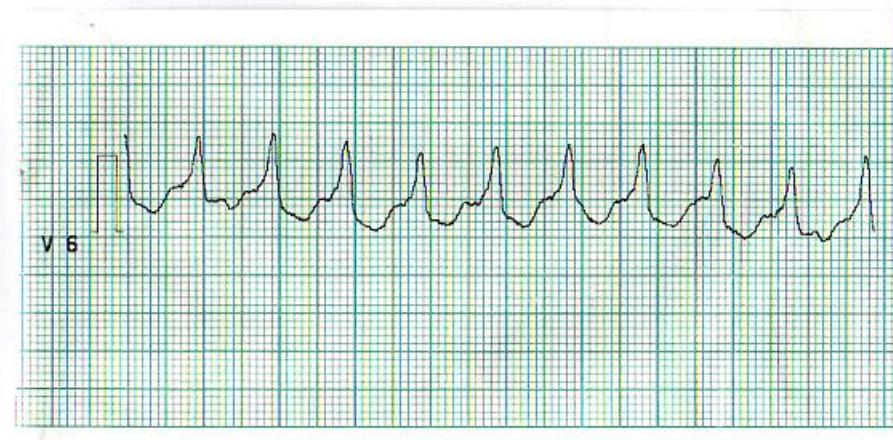
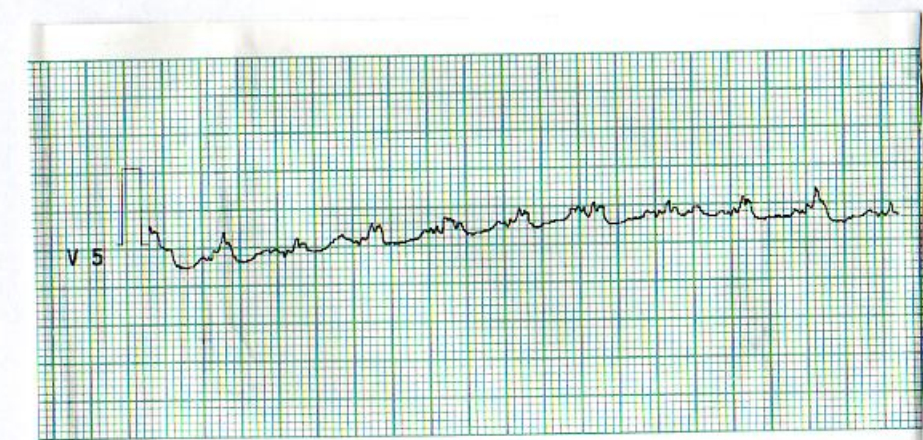


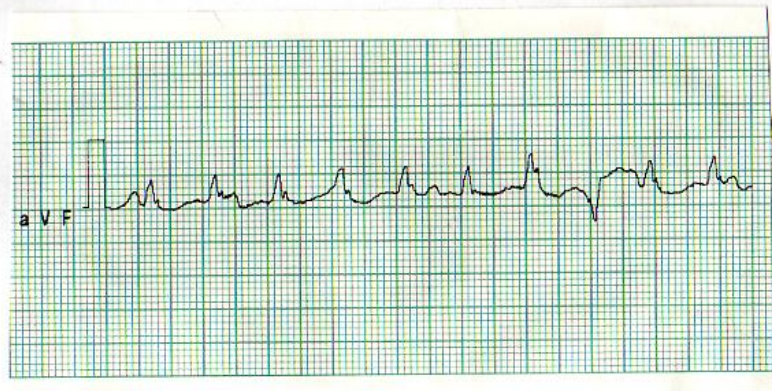
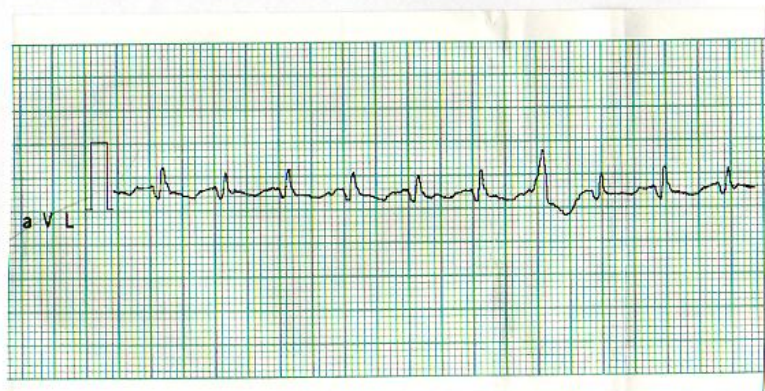
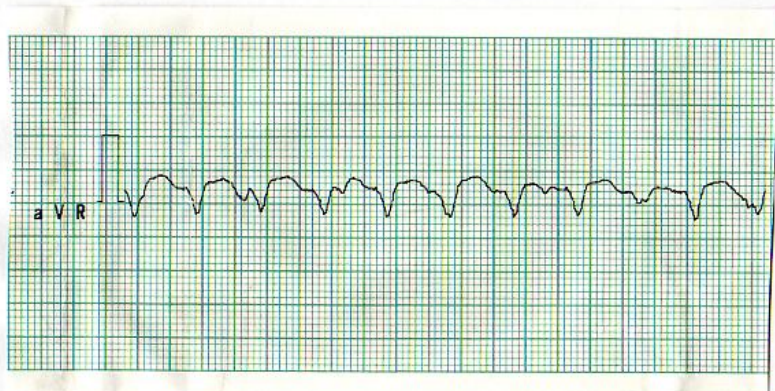
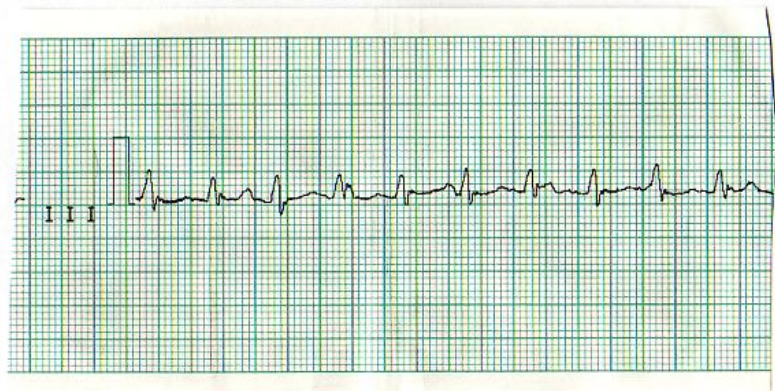
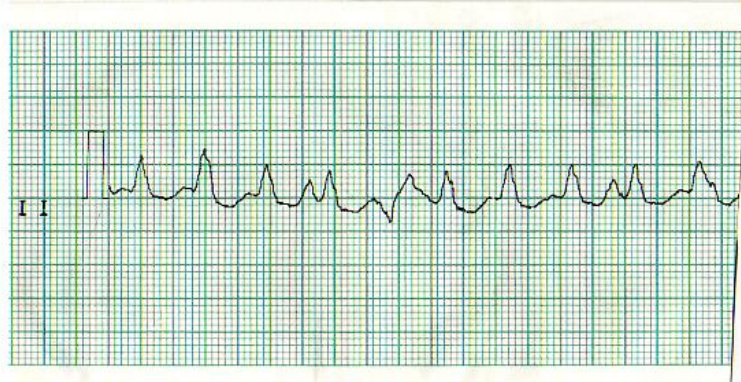
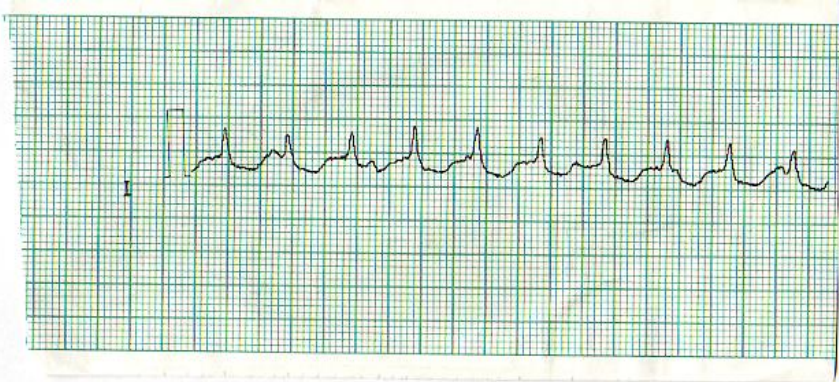
Onda epsilon

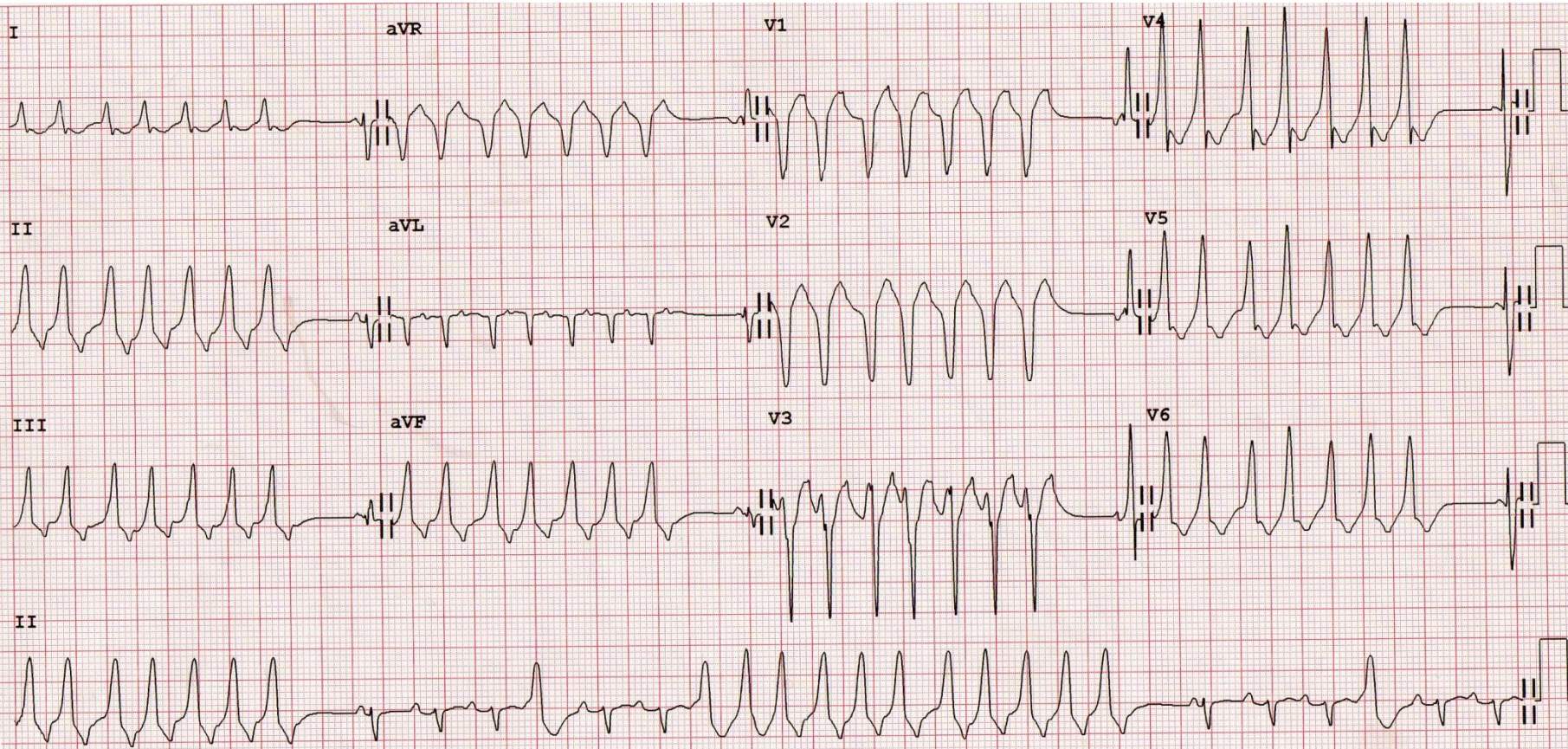




D2 N/2 25 mm/s Filtros: Rede MUSC







Equip:

Veloc: 25 mm/s

Membro: 10 mm/mV Tórax: 10 mm/mV

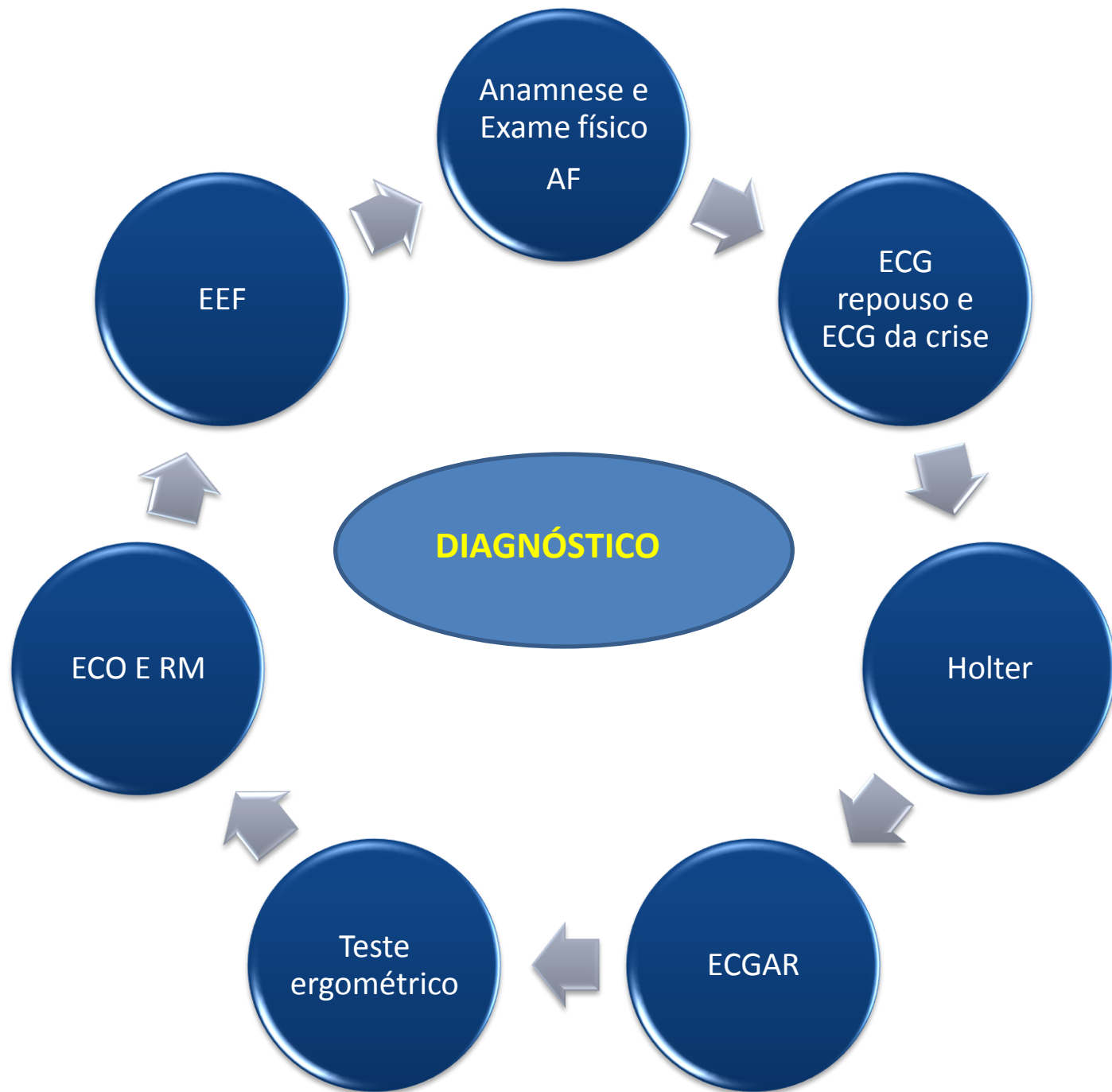
F 60~ 0,5-100 Hz W

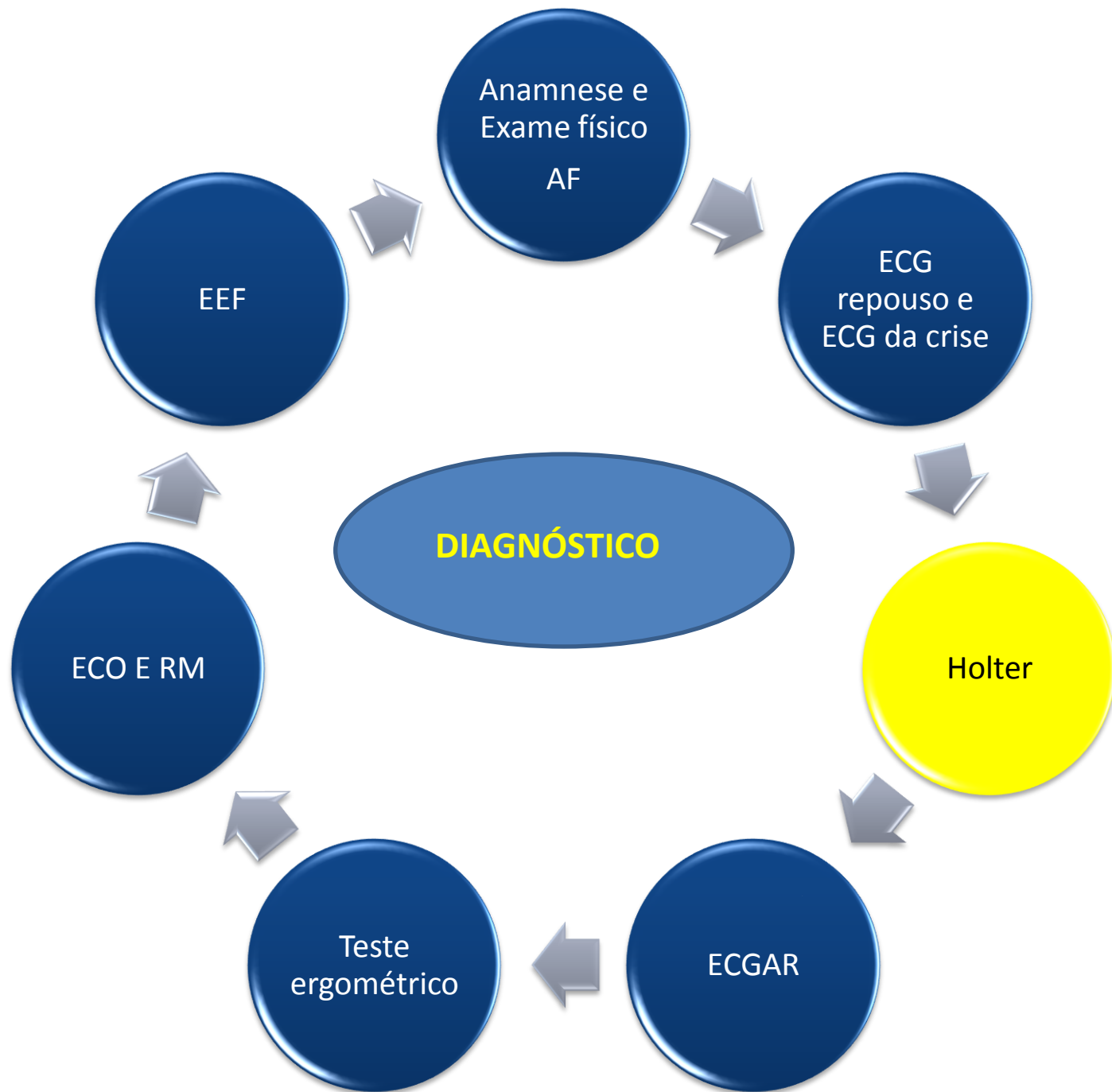
PH080A

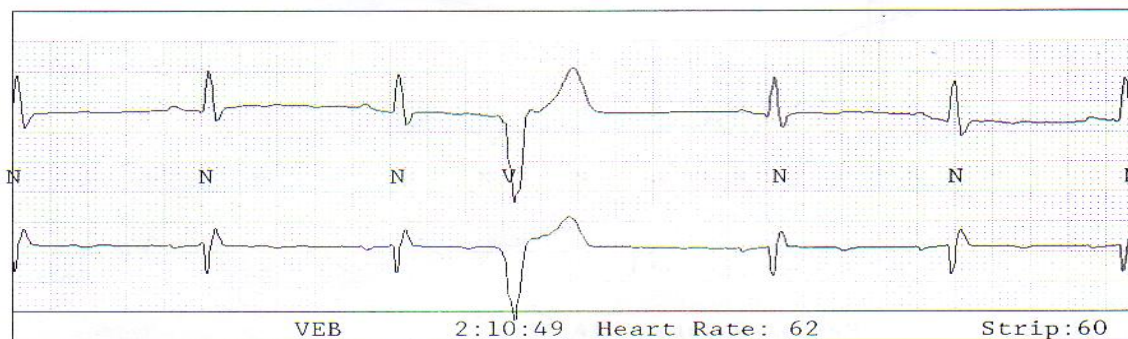
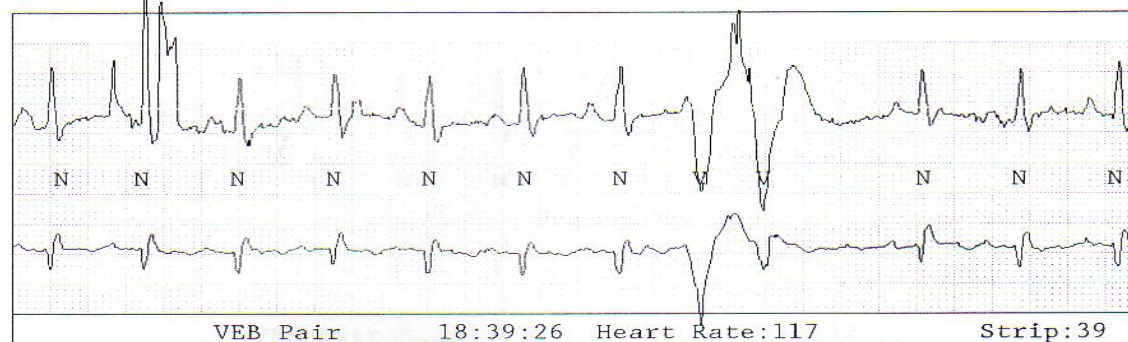
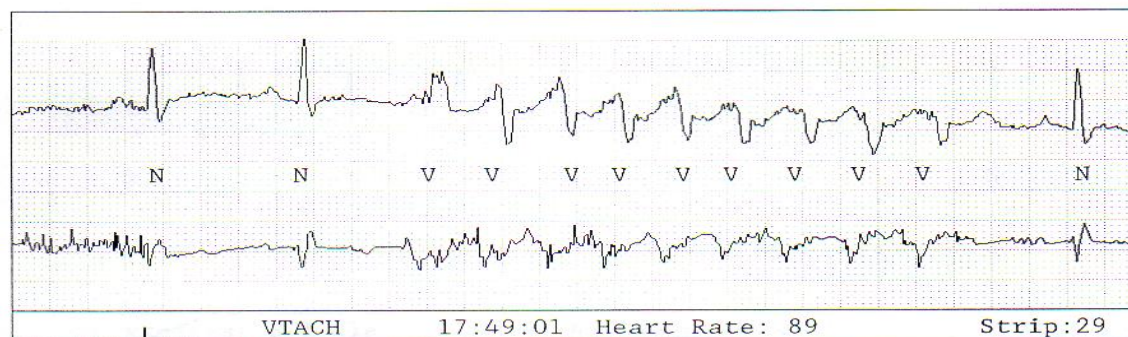
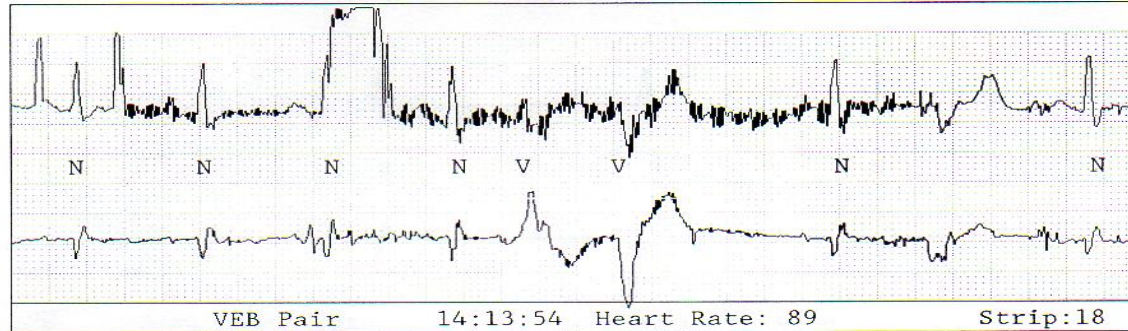
P?

PHILIPS

REORDER M3707A

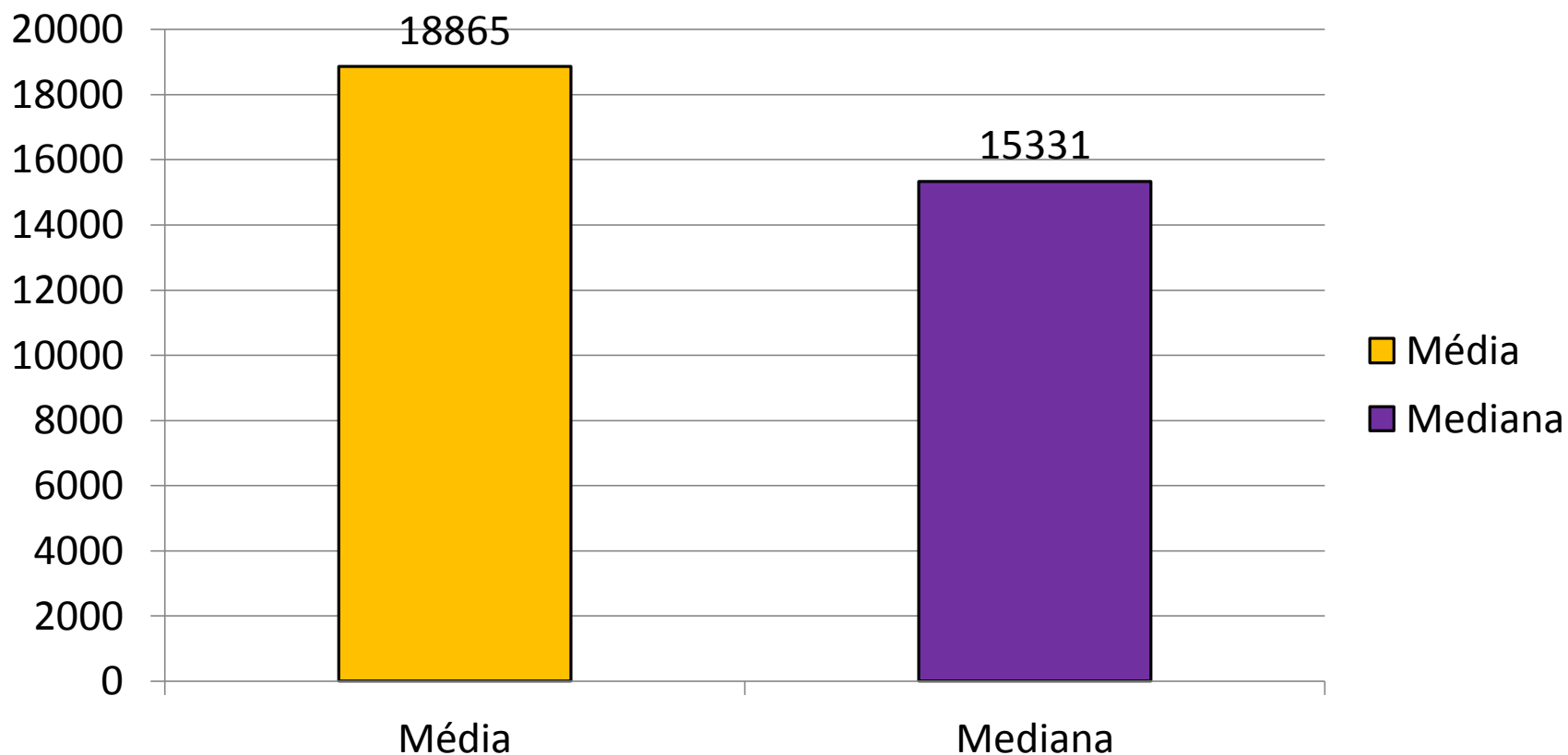






Densidade das ectopias ventriculares

Número de Ectopias em 24h



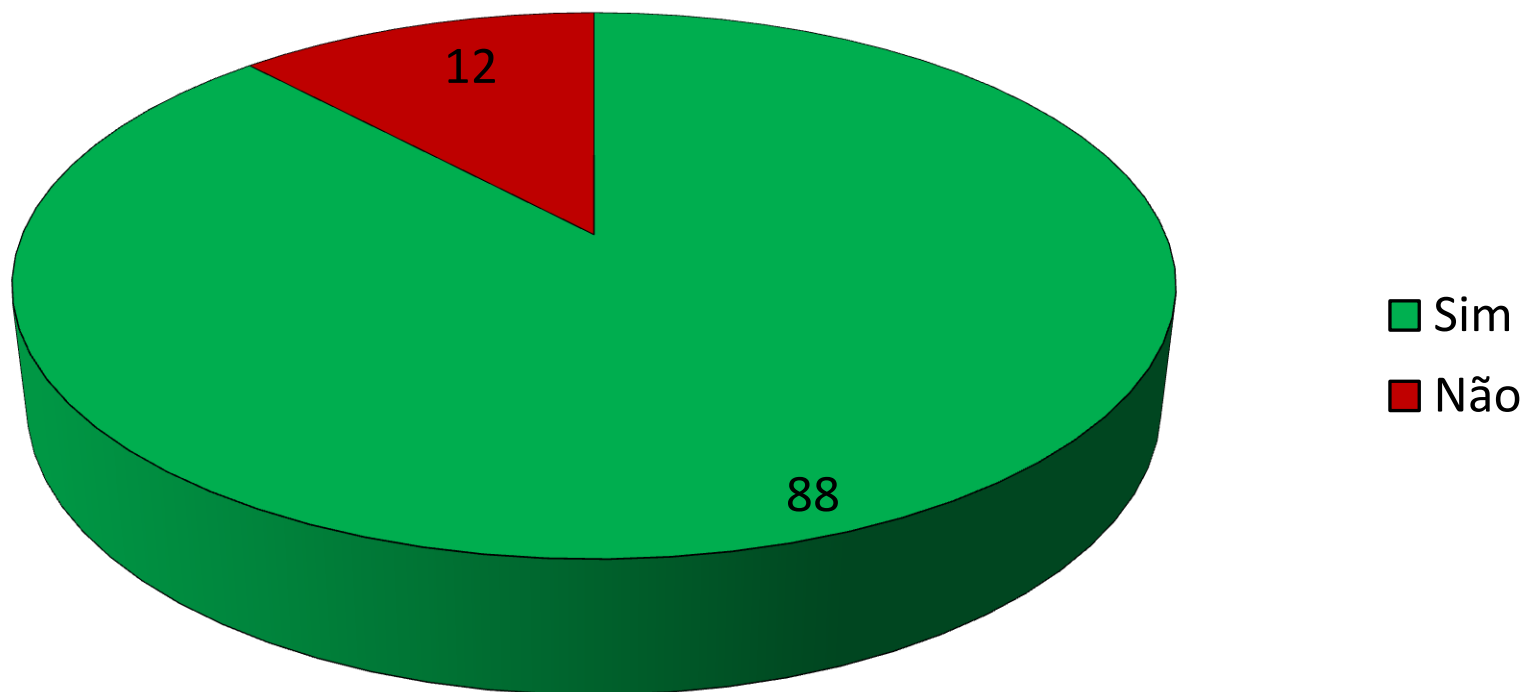
Resposta ao Teste ergométrico

- 100% reduziram ou suprimiram por completo as ectopias no pico do esforço



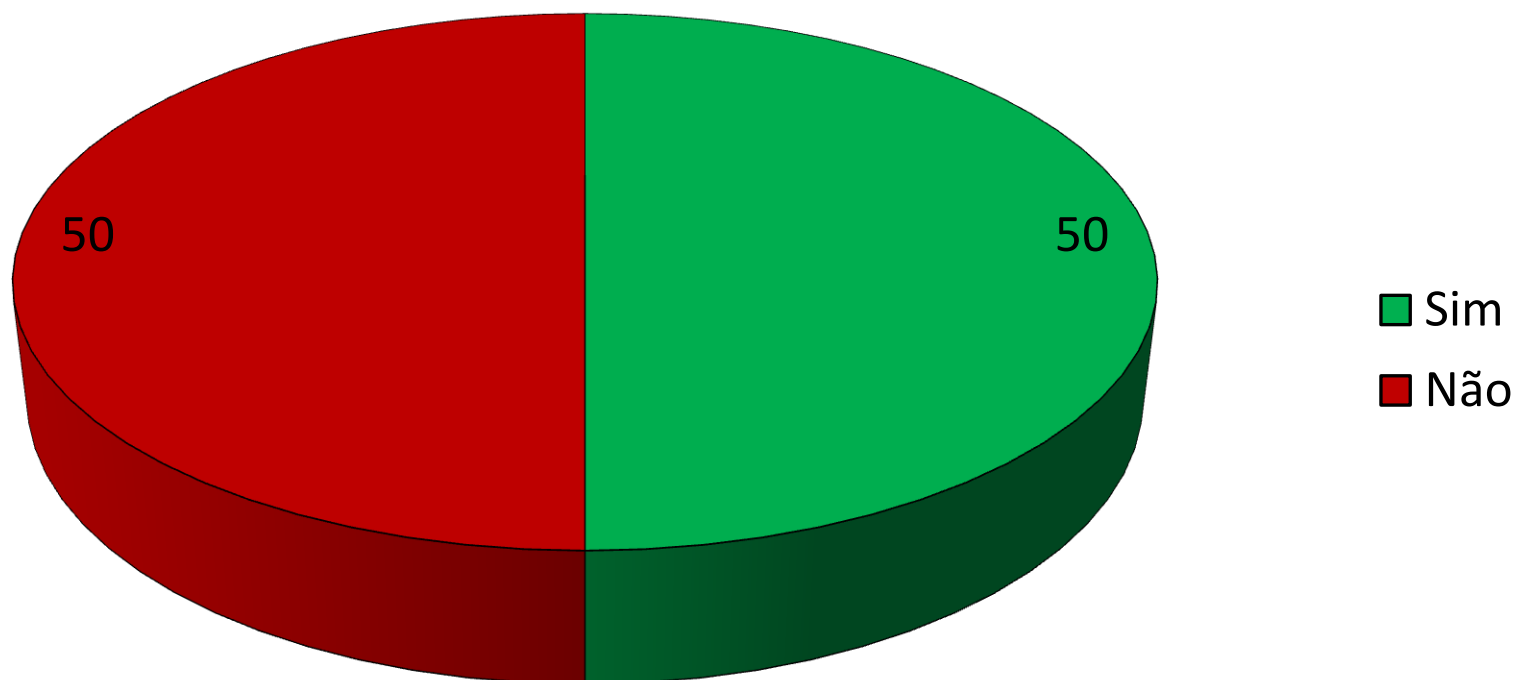
Evolução das ectopias

Redução para menos de 10 ectopias por hora %



Evolução

Remissão espontânea ao longo do seguimento %



Resolução espontânea de EV

Relatório de Holter

1 - Dados do Exame

Nº do Exame 1435	Data do Exame: 03/12/2015 09:01 Protocolo: Holter de 3 canais	Código: 5A0-06882 Convênio: Particular
----------------------------	--	---

2 - Dados do

Nome: [REDACTED]	Idade: 12
Sexo: M	Fumante:
Altura: 1,55	Peso: 42

3 - Médico Solicitante

Nome: Drº. Rogério Braga	Tel.:
Clínica: -	Fax:

4 - Resumo Estatístico

Totais:	Frequência Cardíaca:
Duração (h): 23:42	Mín: 47 bpm às 01:38:25
Nº Total de QRS's: 107.313	Média: 77 bpm
Ectópicos Ventriculares: 14.983 (14%)	Máx: 136 bpm às 08:05:41
Ectópicos Supraventriculares: 139 (<1%)	F.C. >= 120 bpm durante 00:02:23 h
Artefatos (%): 8	F.C. <= 50 bpm durante 01:00:31 h

Arritmias Ventriculares:

- 14.981 Isoladas, das quais
- 584 em 154 episódios de Bigeminismo
- 1 Episódios em Pares
- 0 Taquicardias

Arritmias Supraventriculares:

- 139 Isoladas
- 0 Pareadas
- 0 Taquicardias

Pausas

0 Pausas (>= 2,0 s.)

Depressão do ST

- C1: 0 episódios
- C2: 0 episódios
- C3: 0 episódios

Elevação do ST

- C1: 0 episódios
- C2: 0 episódios
- C3: 0 episódios

Relatório de Holter

1 - Dados do Exame

Nº do Exame 1987	Data do Exame: 10/06/2016 10:03 Protocolo: Holter de 3 canais	Código: 5A0-07491 Convênio: Amil
----------------------------	--	-------------------------------------

2 - Dados do

Nome: [REDACTED]	Idade: 13
Sexo: M	Fumante:
Altura: 1,60	Peso: 45,0

3 - Médico Solicitante

Nome: Drº. Rogério Braga	Tel.:
Clínica: -	Fax:

4 - Resumo Estatístico

Totais:	Frequência Cardíaca:
Duração (h): 23:09	Mín: 46 bpm às 03:56:47
Nº Total de QRS's: 110.906	Média: 82 bpm
Ectópicos Ventriculares: 19 (<1%)	Máx: 147 bpm às 16:05:18
Ectópicos Supraventriculares: 135 (<1%)	F.C. >= 120 bpm durante 00:19:39 h
Artefatos (%): 11	F.C. <= 50 bpm durante 01:13:16 h

Arritmias Ventriculares:

- 19 Isoladas, das quais
- 0 em 0 episódios de Bigeminismo
- 0 Episódios em Pares
- 0 Taquicardias

Arritmias Supraventriculares:

- 135 Isoladas
- 0 Pareadas
- 0 Taquicardias

Pausas

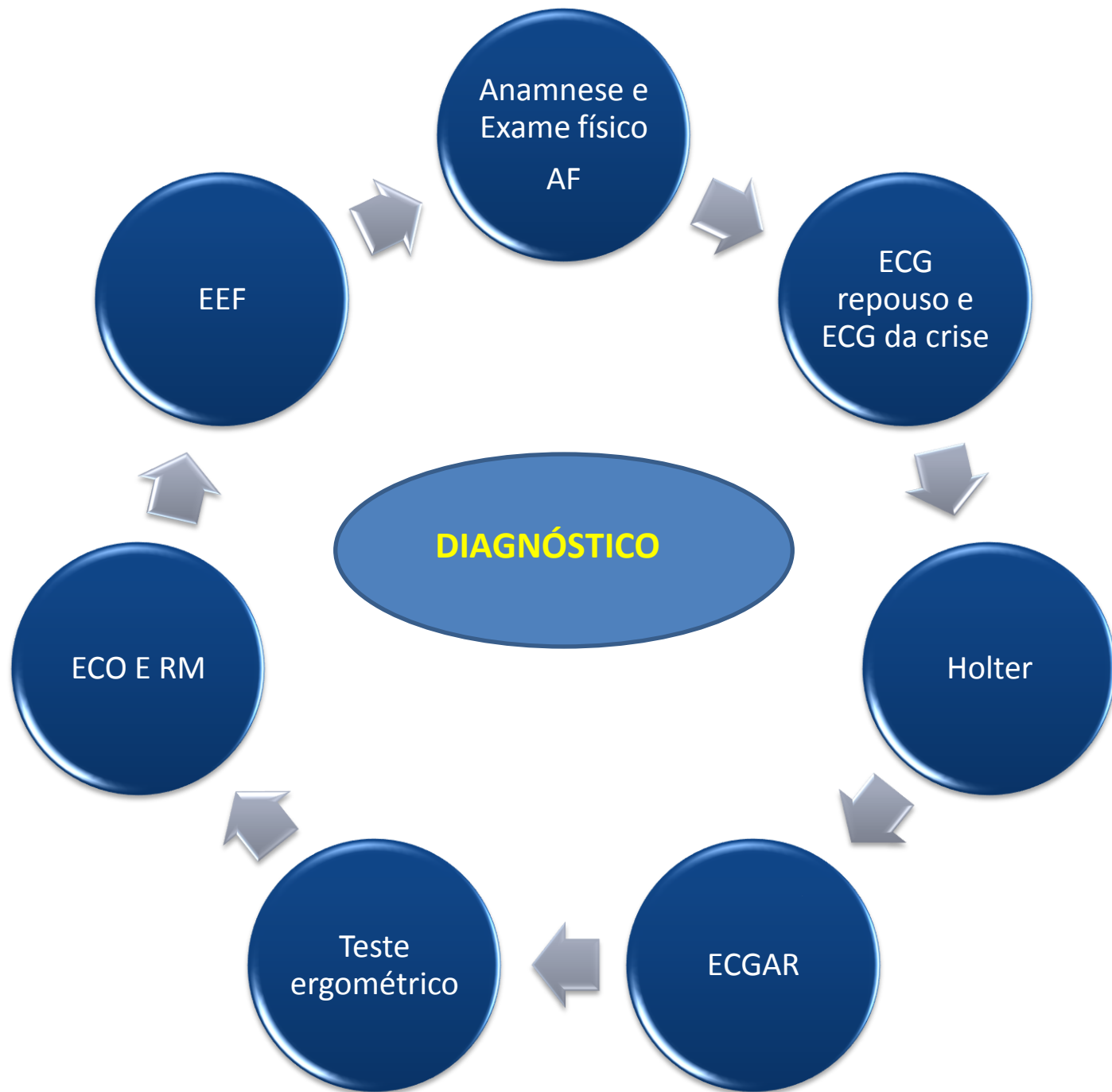
0 Pausas (>= 2,0 s.)

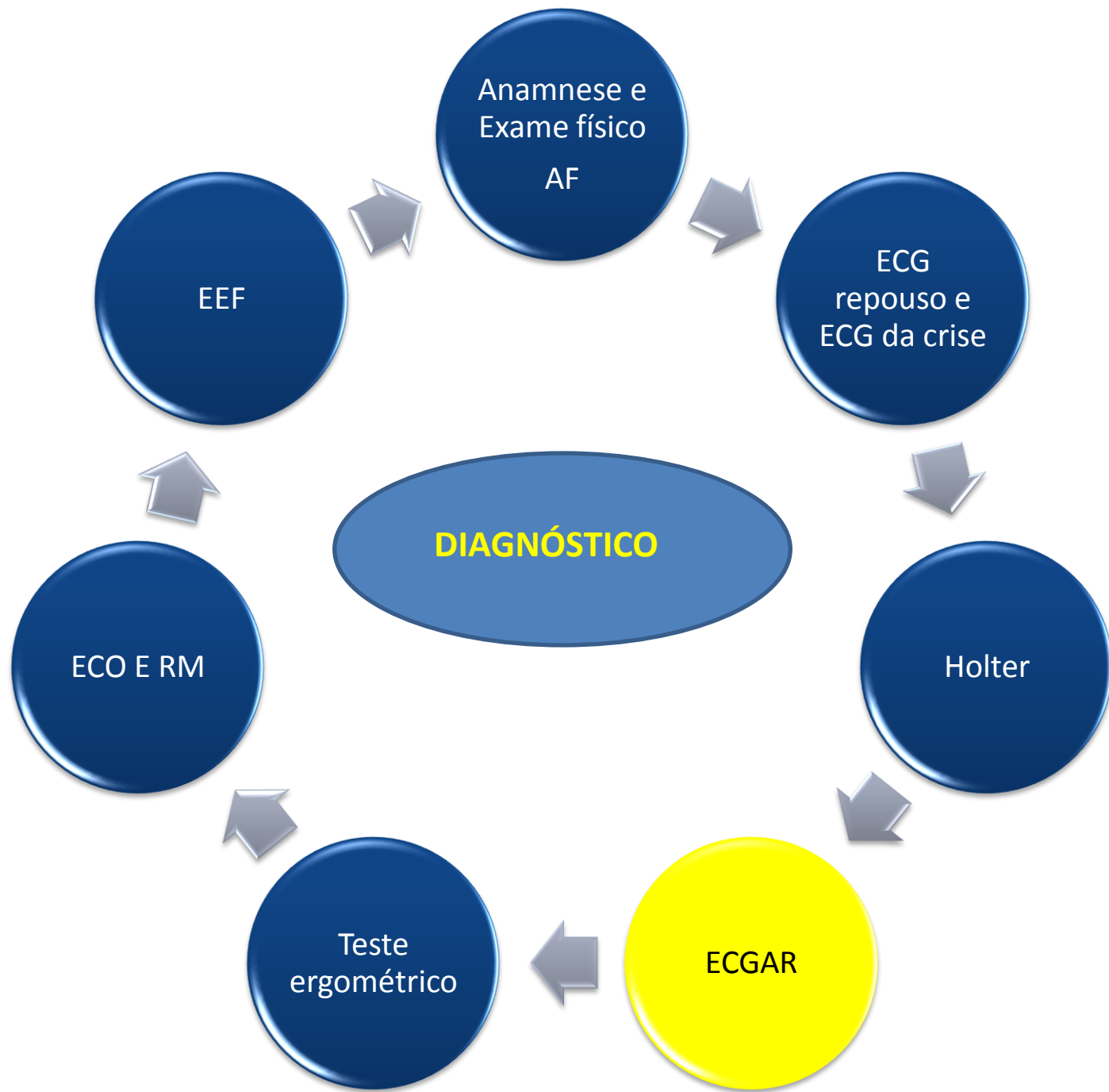
Depressão do ST

- C1: 0 episódios
- C2: 0 episódios
- C3: 0 episódios

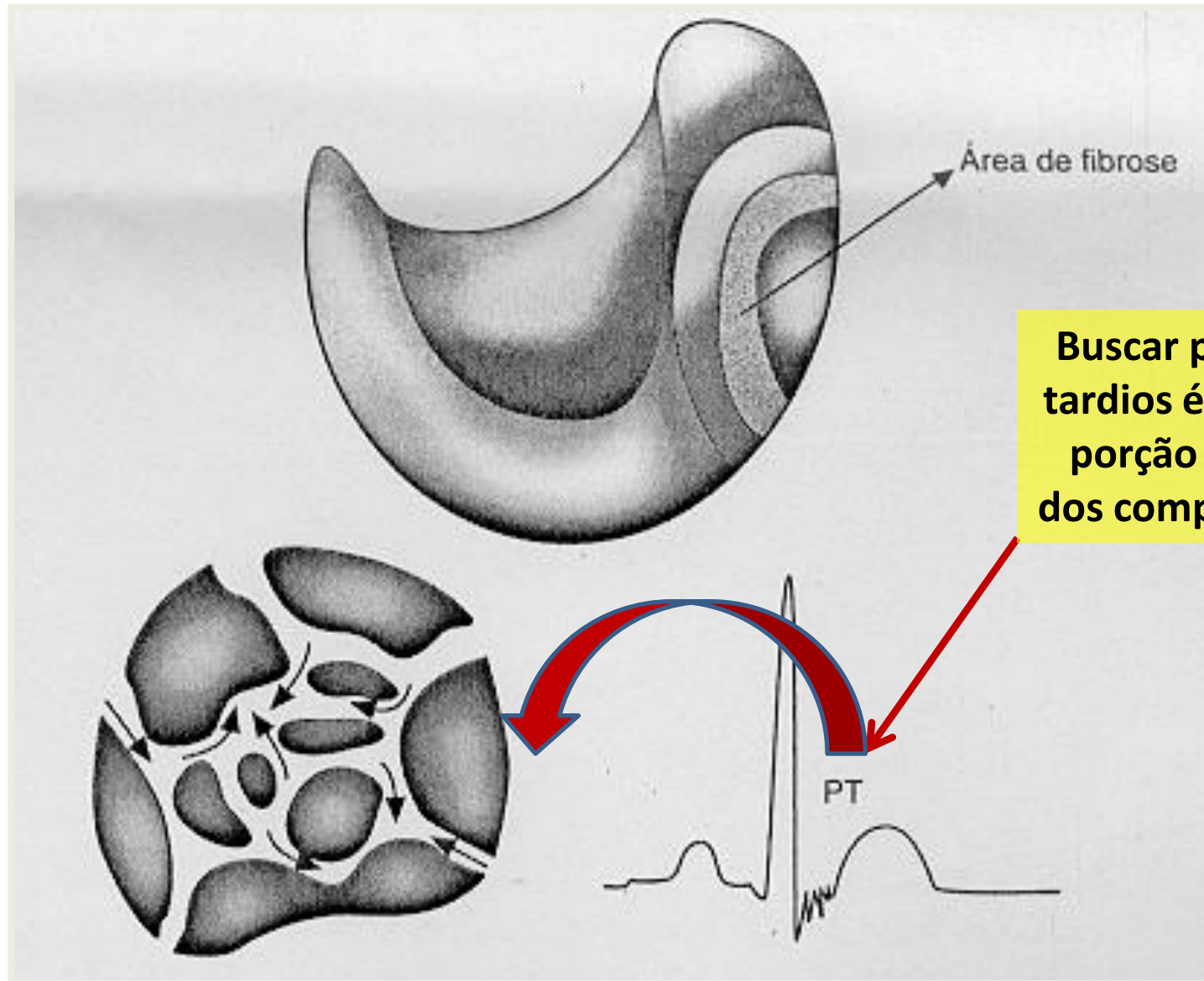
Elevação do ST

- C1: 0 episódios
- C2: 0 episódios
- C3: 0 episódios





Áreas de Fibrose Entremeadas com Tecido Sadio



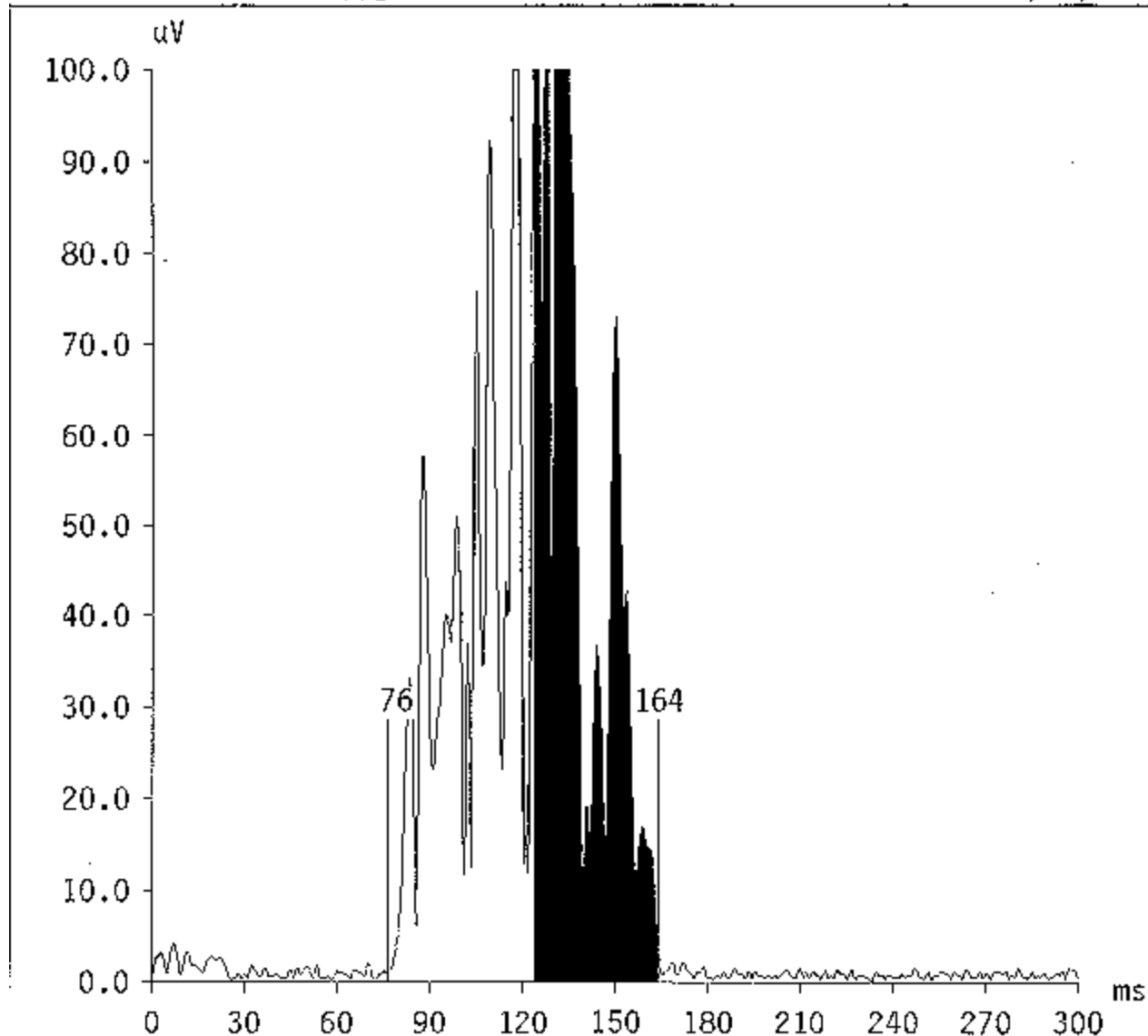
Buscar potenciais tardios é analisar a porção terminal dos complexos QRS

PREDICTOR Version 6.1

SAECG ANALYSIS

Date: 05/06/02

VECTOR



HAS.004

1/14/02 10:45a

Proto: BASIC

Type: BiSpec

Hi freq: 40

Total QRS

Dur: 87.5

RMS: 61.32

INT: 4.29

Terminal QRS

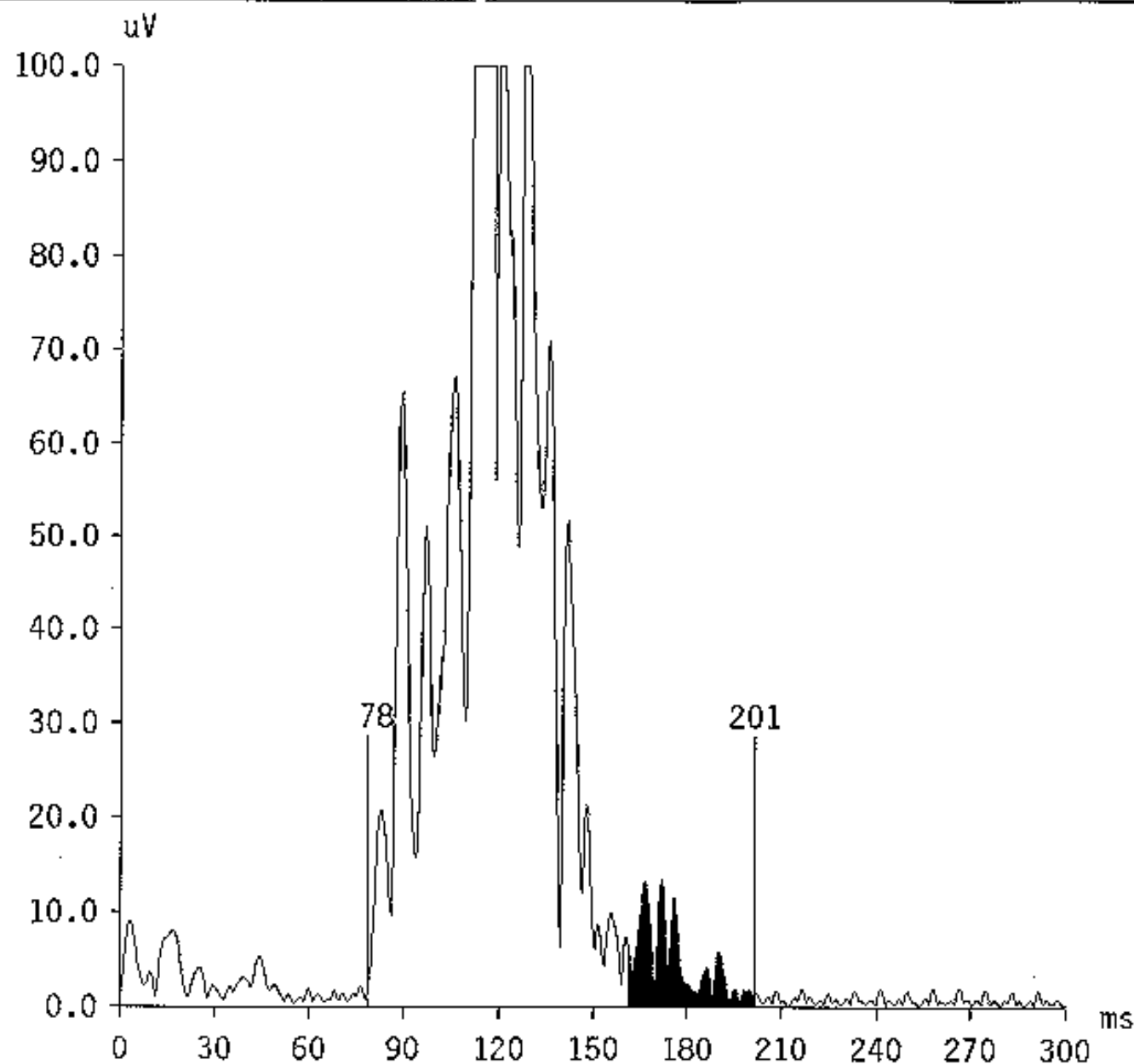
RMS: 73.93

MN: 57.34

LAS: 10.0

iQRSD: 87.5

Scale: 10.0



Proto: BASIC

Type: BiSpec

Hi freq: 40

Total QRS

Dur: 123.0

RMS: 73.19

INT: 5.09

Terminal QRS

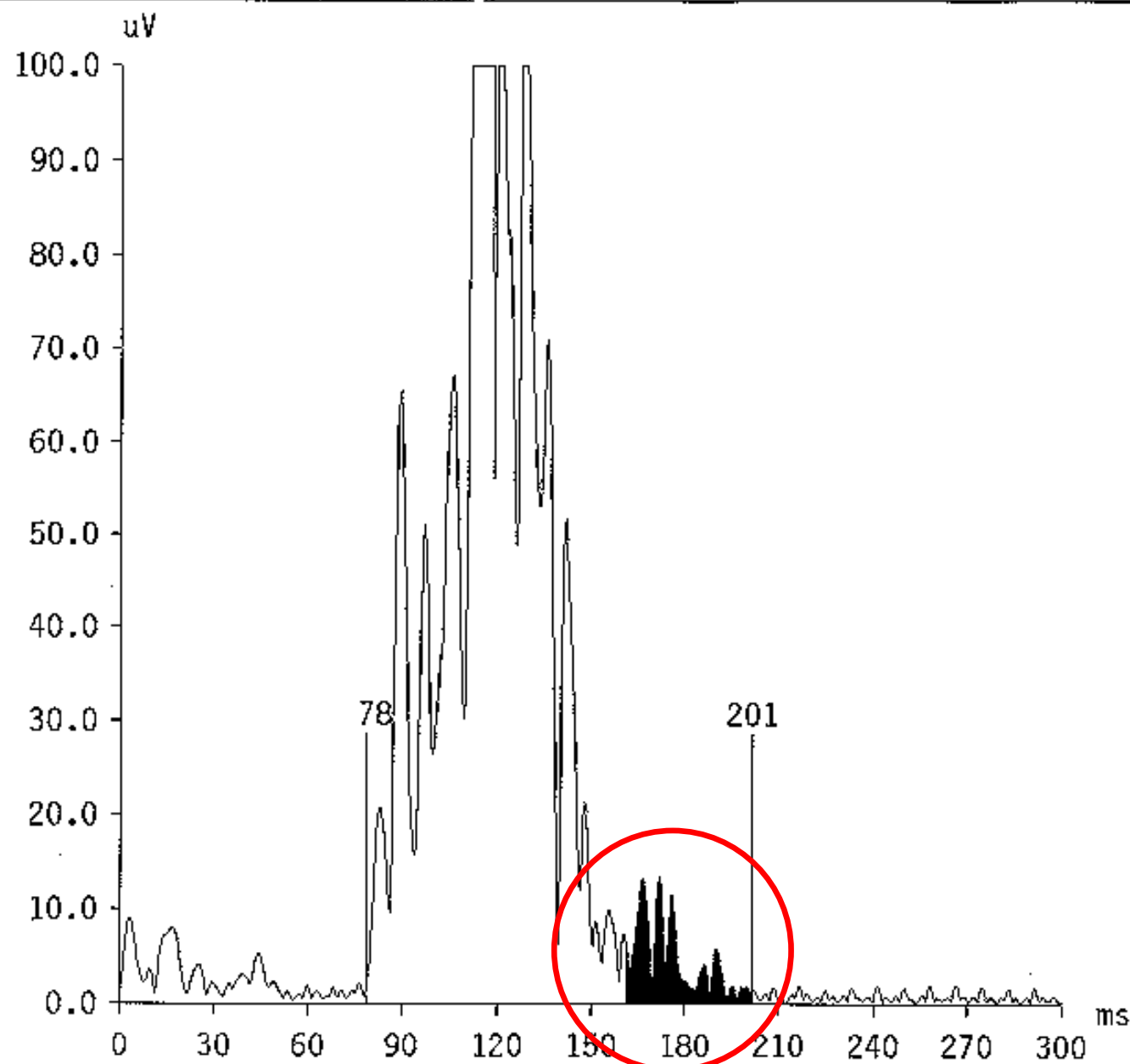
RMS: 5.30

MN: 4.22

LAS: 58.0

iQRSD: 123.0

Scale: 10.0



Proto: BASIC

Type: BiSpec

Hi freq: 40

Total QRS

Dur: 123.0

RMS: 73.19

INT: 5.09

Terminal QRS

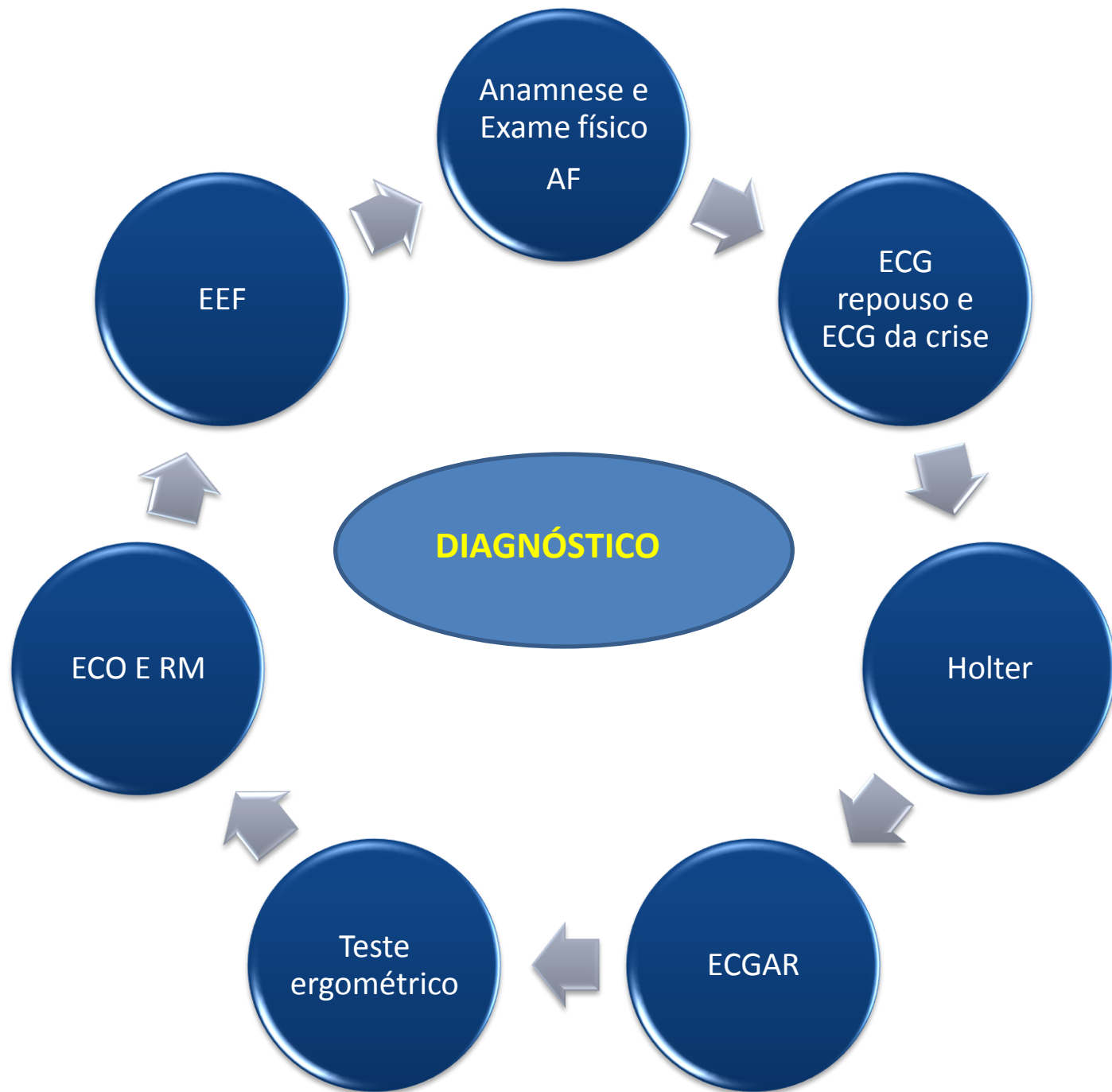
RMS: 5.30

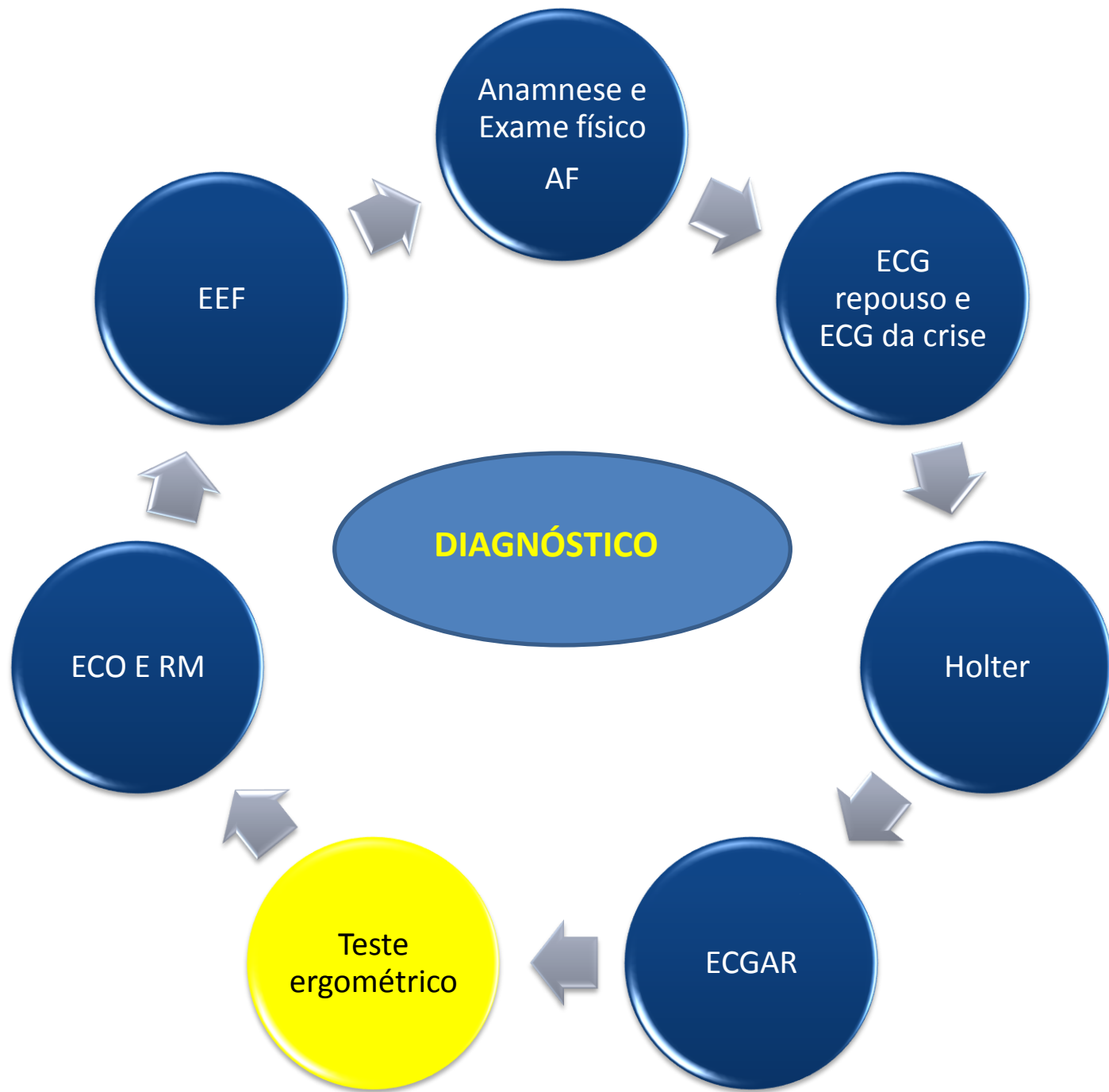
MN: 4.22

LAS: 58.0

iQRSD: 123.0

Scale: 10.0

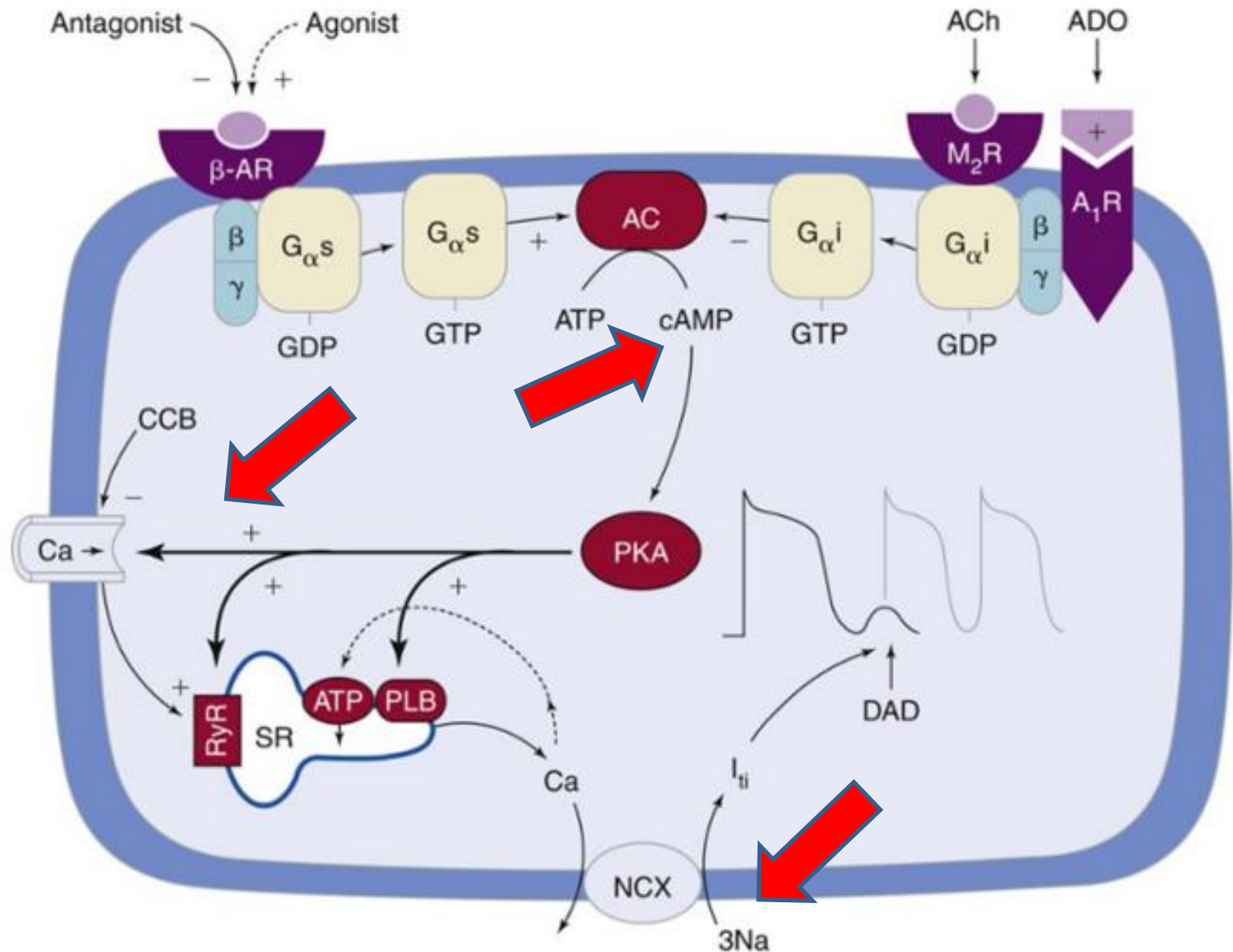








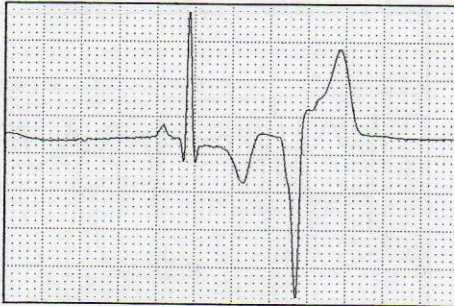
Mecanismo iônico da atividade deflagrada



**ARRITMIAS POR REENTRADA
HABITUALMENTE PIORAM NO PICO
DO ESFORÇO**

PRÉ ESFORÇO

MC5m N



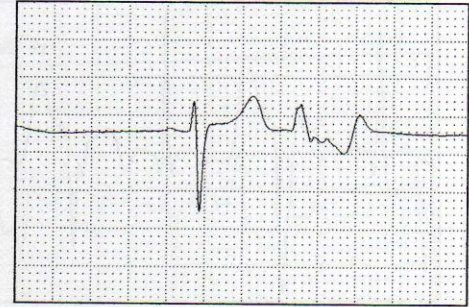
076 BPM P=105/080 00:00 EM PÉ

D2 N



25 mm/s REDE MUSC

V2 N



MC5m N



039 BPM 00:00 APNÉIA INS 25 mm/s REDE MUSC

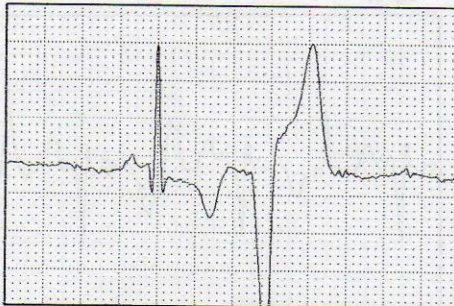
D2 N



V2 N

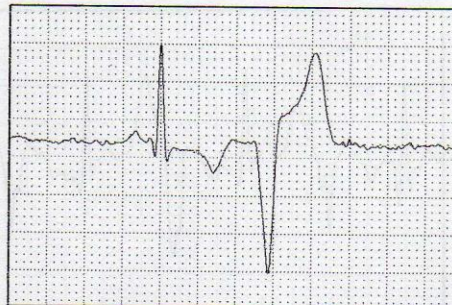


MC5m N

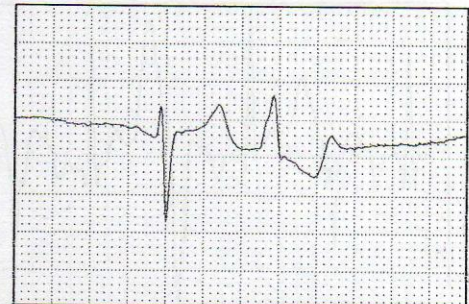


067 BPM 00:00 HIPERPNÉIA 25 mm/s REDE MUSC

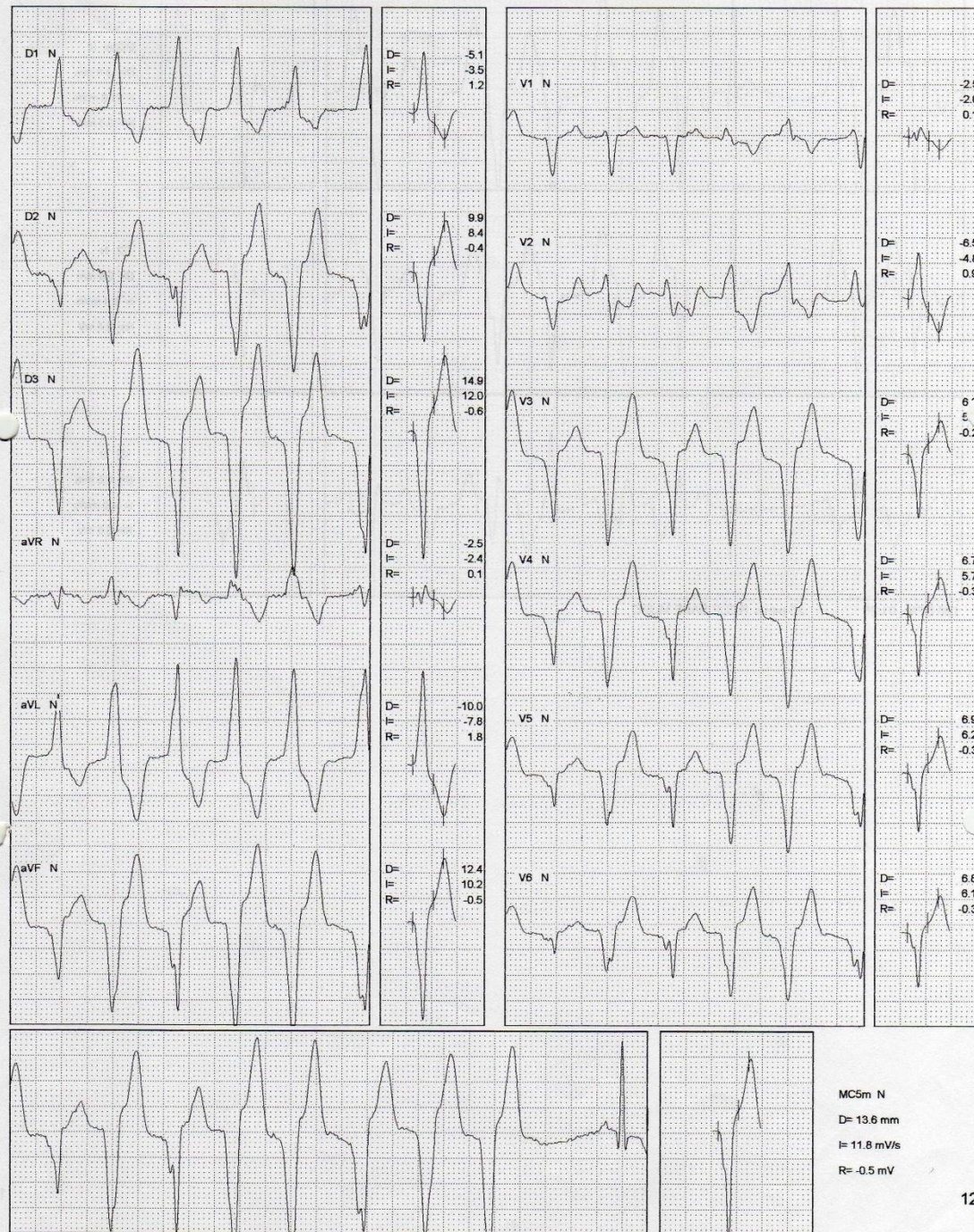
D2 N

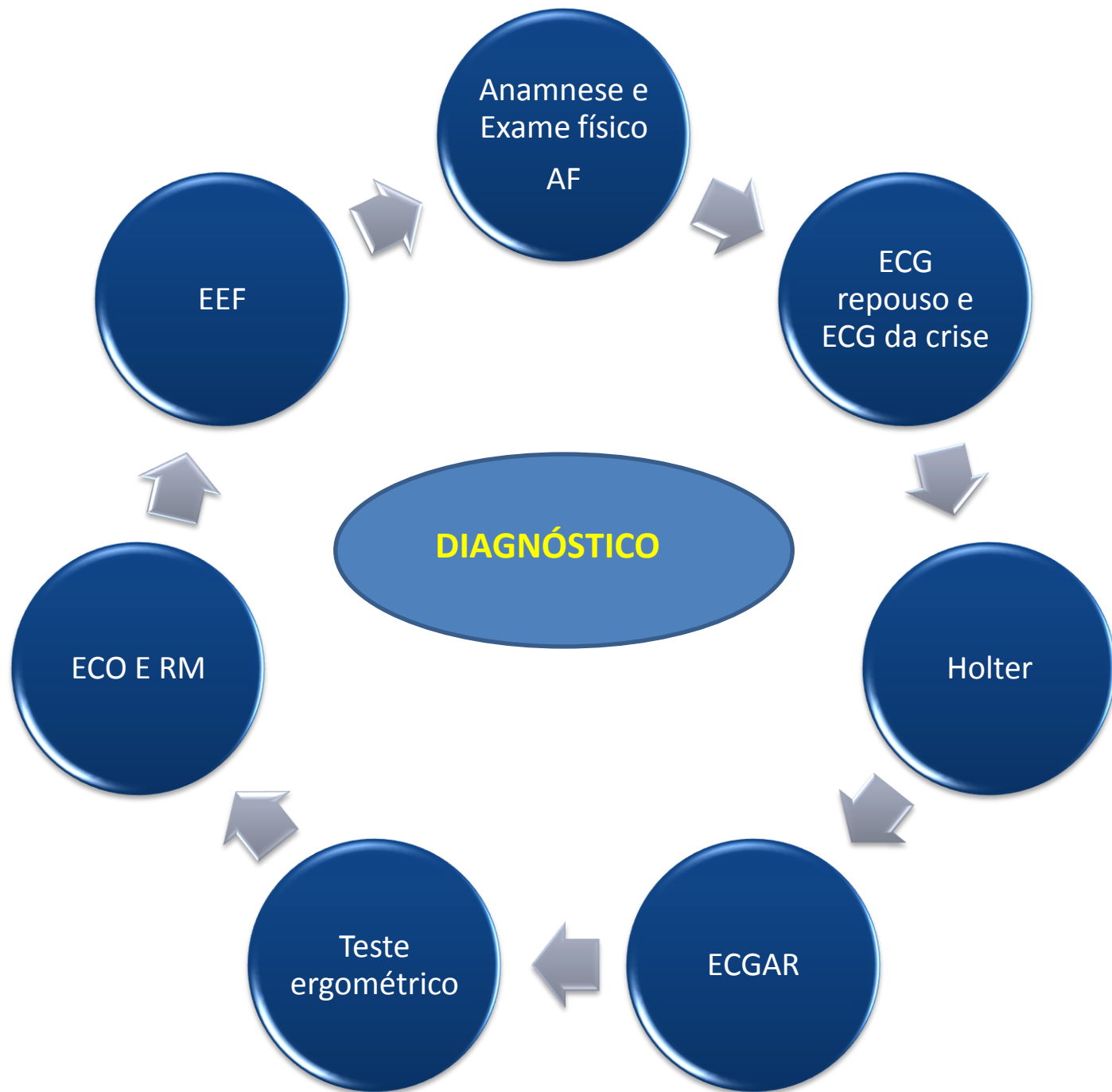


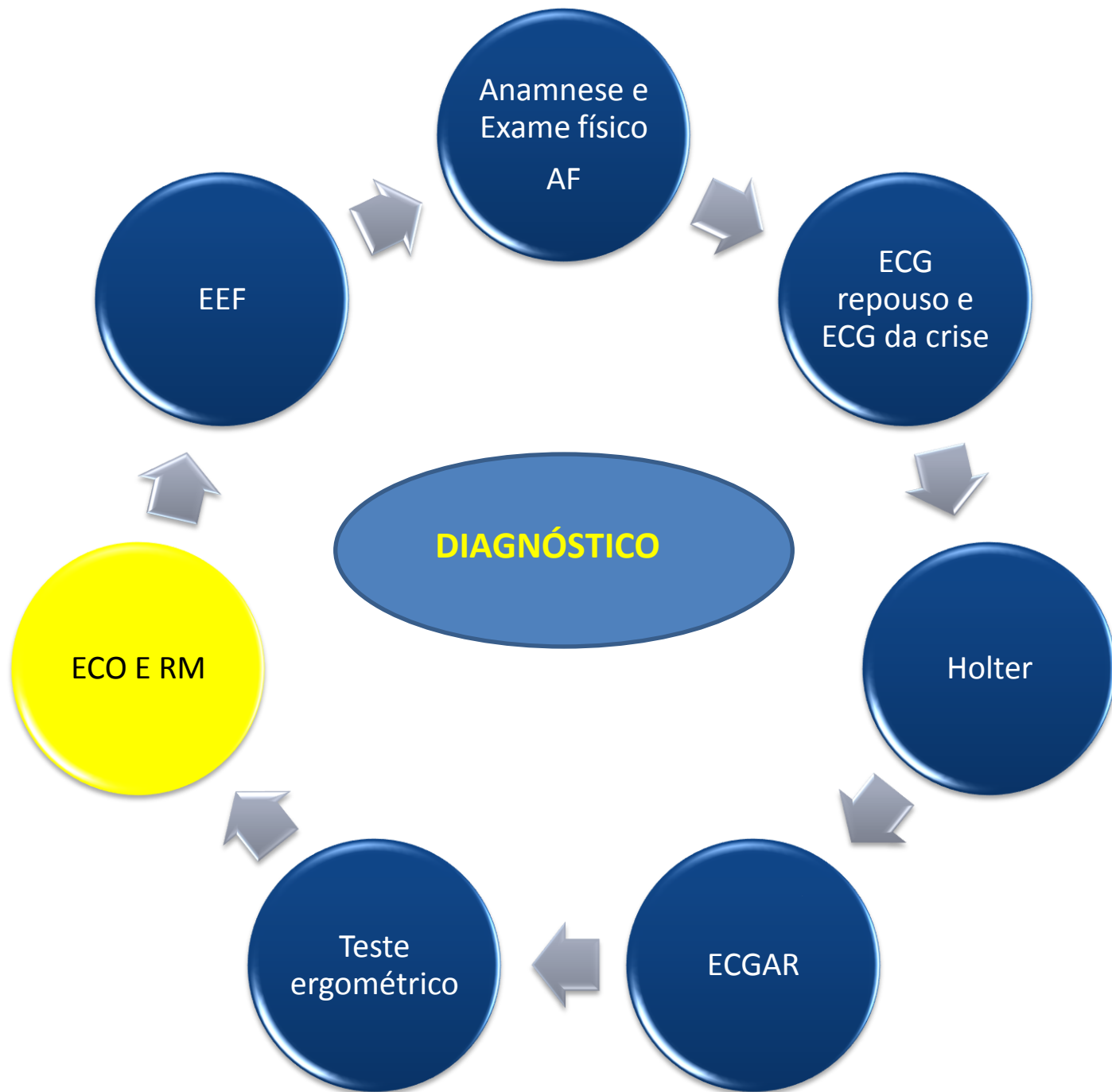
V2 N

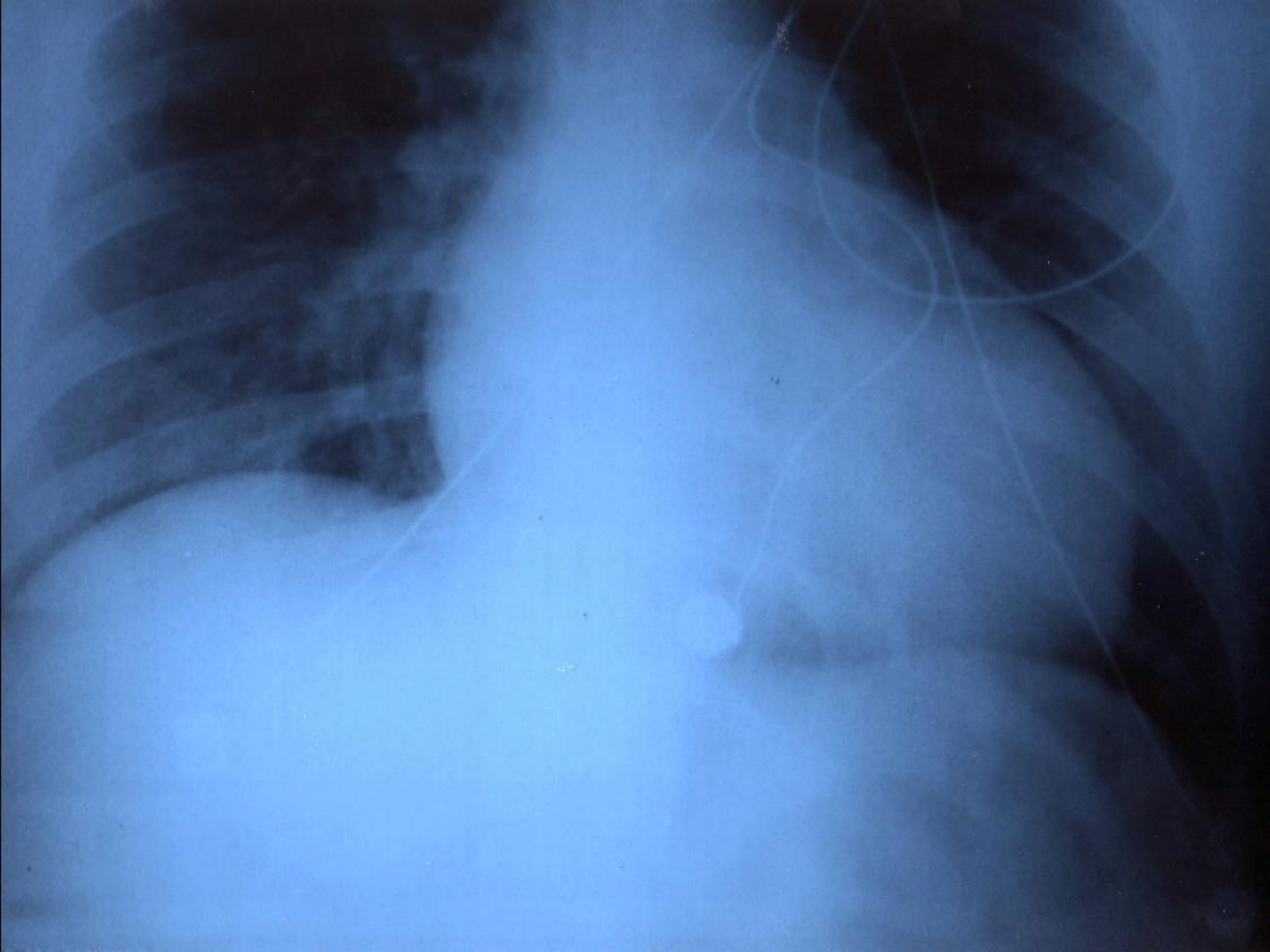












A/2535AA-C CENTER: 18/Jan/05
 2.5/2.0MHz ID: SIMONE # 015148 08:21
 20028E085F

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
 o OFF

HR bpm

FROZEN

MENU TRACKBALL COMPUT ☐ LOOP ☐ TEXT ☐ SAVE ☐ RECALL ☐
 Press <ENTER> or <ESC> when ready



A/2535AA-C CENTER: 18/Jan/05
 2.5/2.0MHz ID: SIMONE # 015148 08:23
 20028F07CF

P MAX %
 G 6
 R 0 %
 Focus 7
 D 21 cm
 PRE 4
 PST 1

PRF 3.0KHz

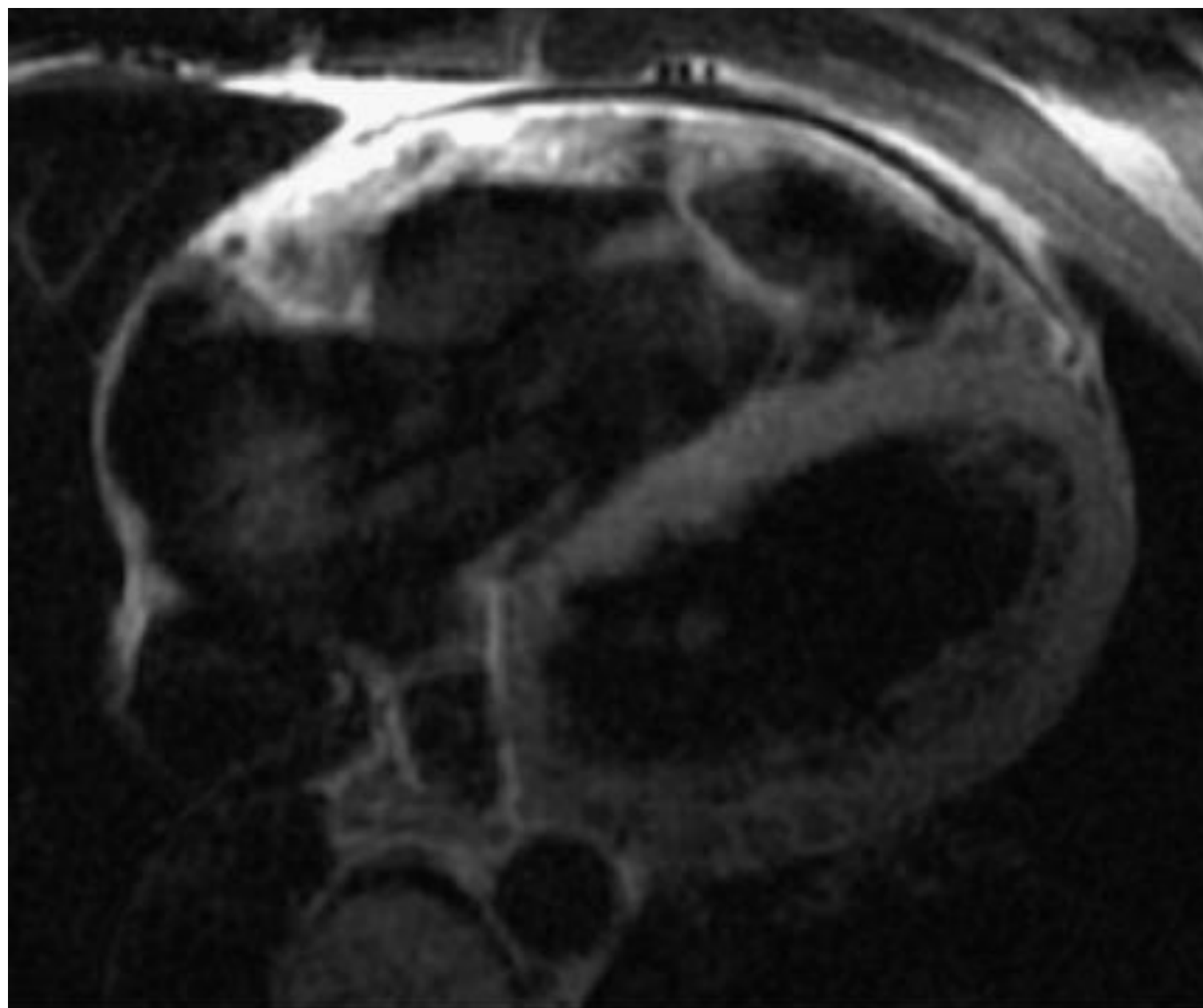
SD 133 mm
 SS 8 mm
 WF 200 Hz
 o OFF

HR bpm

FROZEN

MENU TRACKBALL COMPUT ☐ LOOP ☐ TEXT ☐ SAVE ☐ RECALL ☐
 Press <ENTER> or <ESC> when ready

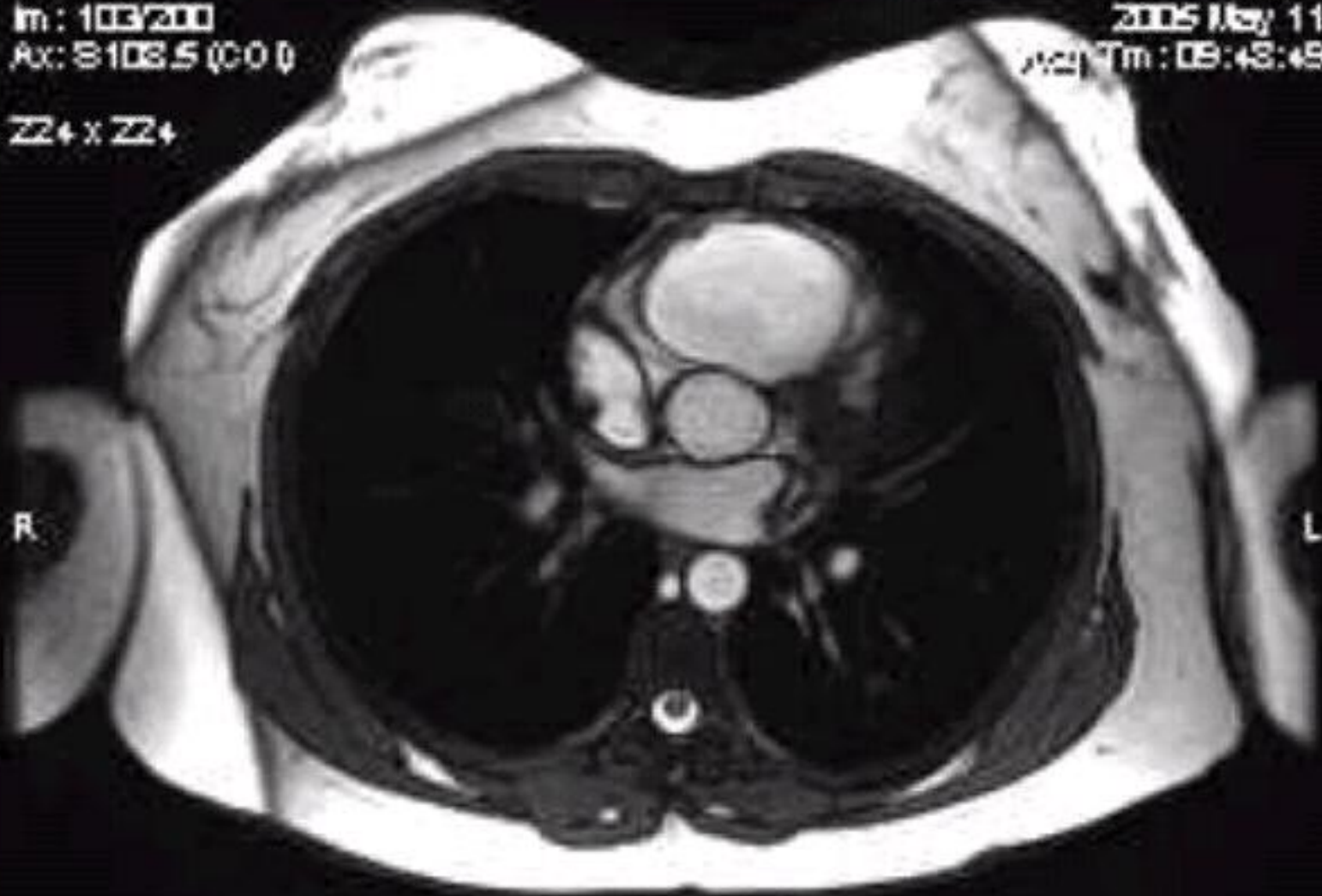




Cine RM - 4 Câmaras 6

1.5T GEH50W
Ex: 12517
CINE DIR
Se: 29
Im: 103/200
Ac: S103.5 (C00)
Z2+ x Z2+

R MED IMAGEM B PORTUGUESA
SNOHEM, PULINI
0357 F 2053062
Acq:
2005 May 11
7:21 Tm: 09:48:49



CINE DIR
Se: 35
Im: 53/200
Ax: 566.5 (C-0)

Study: 20050002
Acq: 2005 May 11
Acq Tm: 09:48:49

224 x 224

R

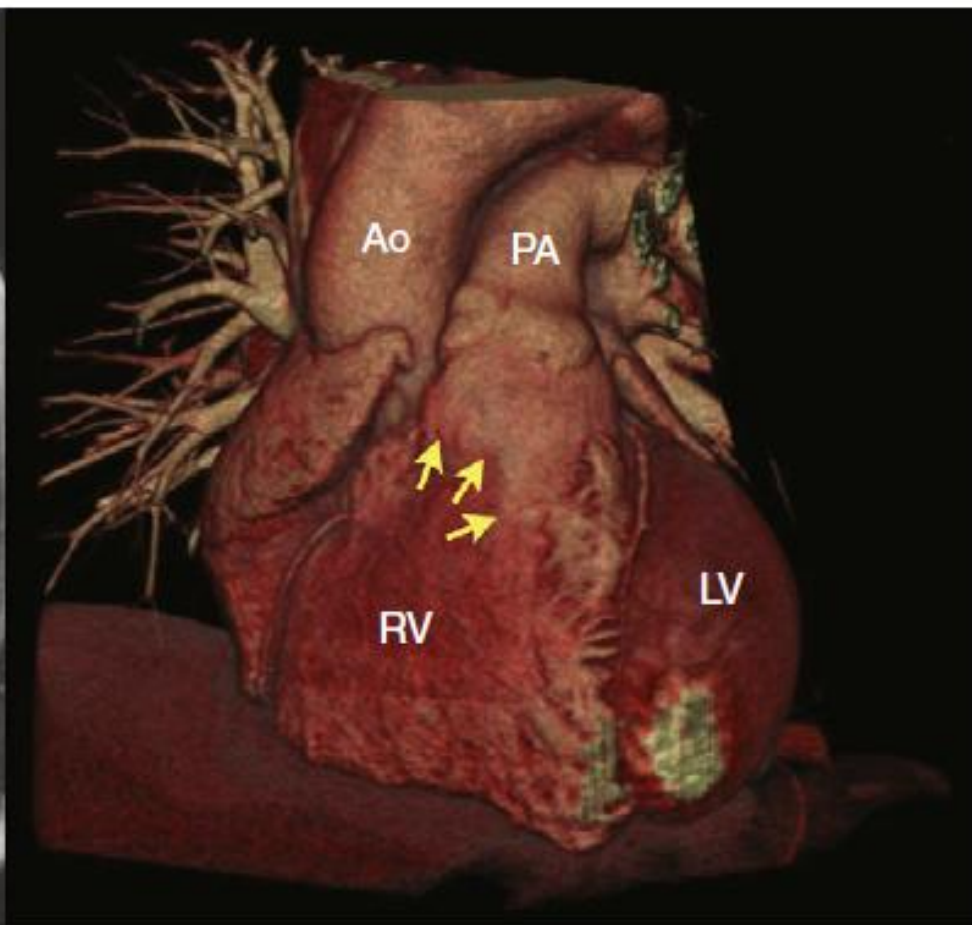
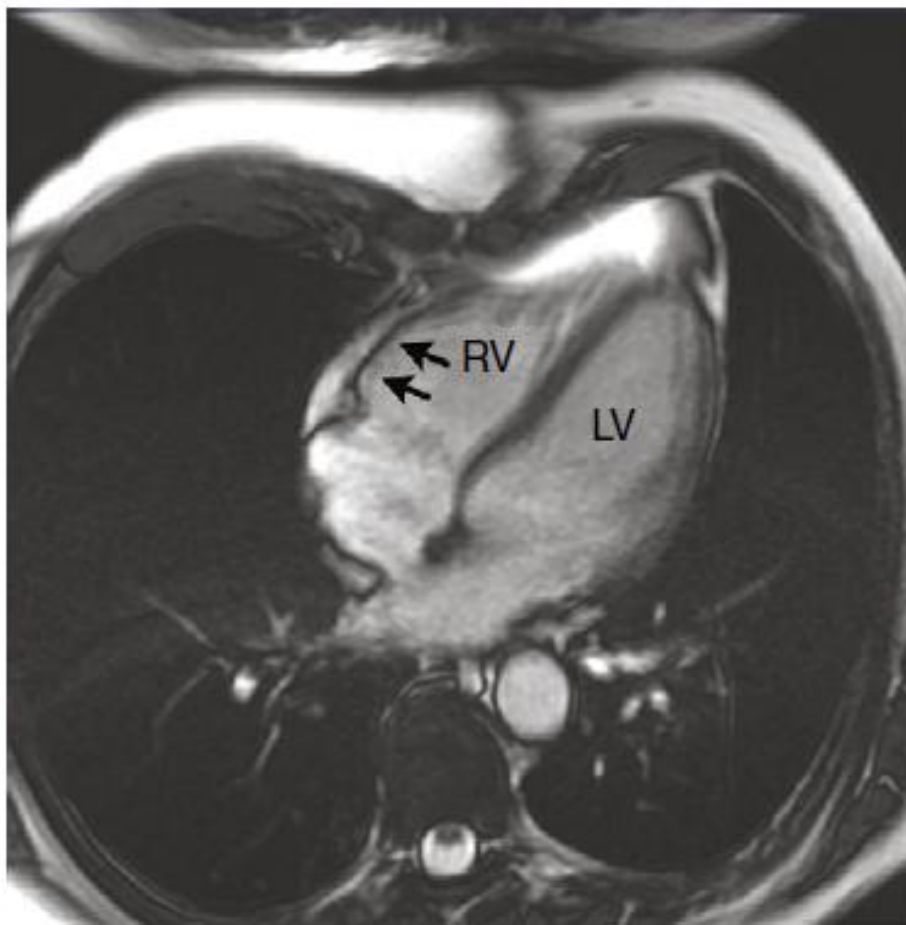
L

ET: 0
TR: 3.3
TE: 1.5
SCARDIAC
SDINW5Dsp
W: 1262 L: 584

P

DFOV: 42.0 x 42.0cm





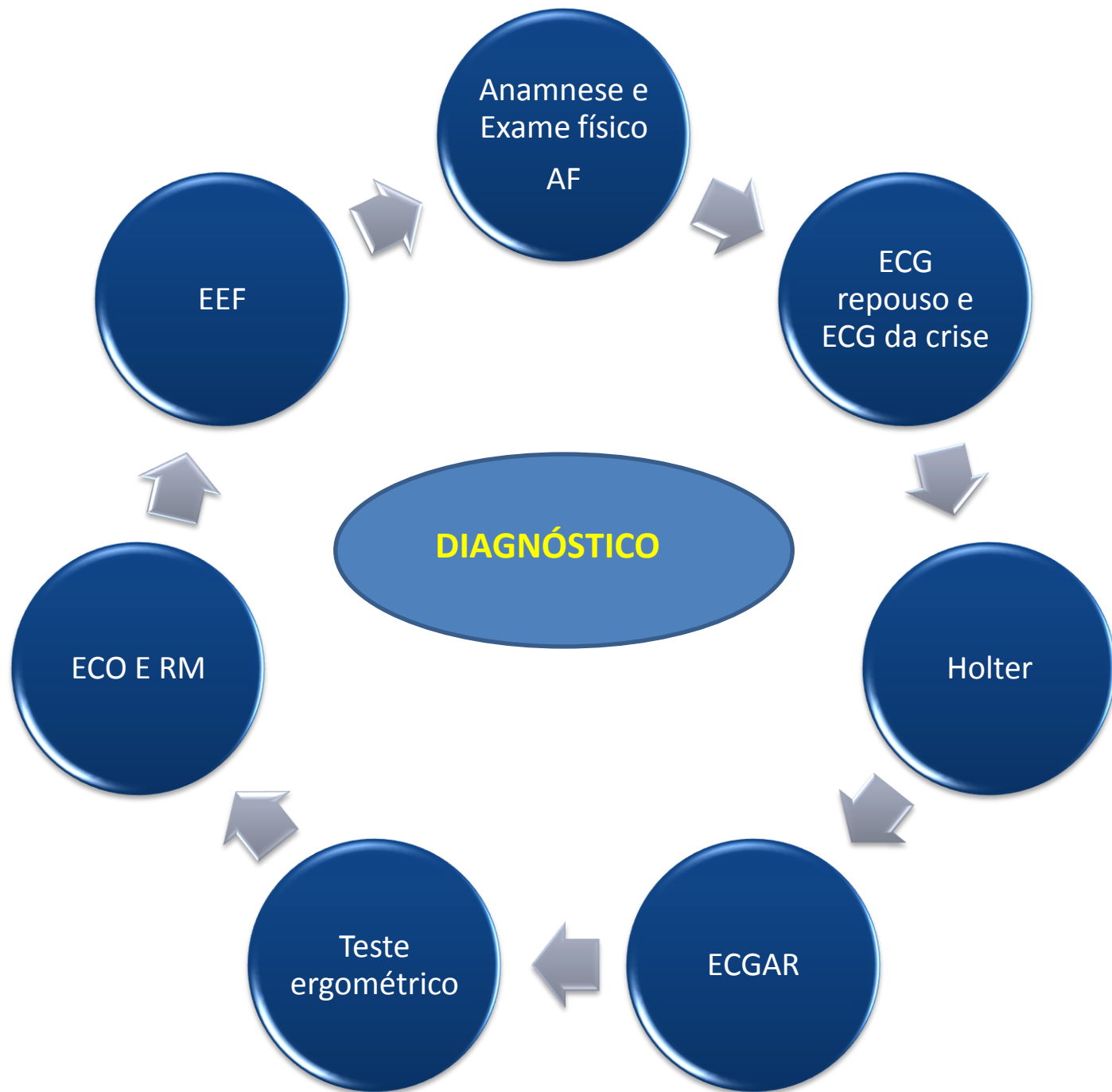




Table 13.1 Task Force criteria for diagnosis of ARVC^a

GROUP	Major	Minor
1. Global and/or regional dysfunction and structural alterations	Severe dilatation and reduction of right ventricular ejection fraction with no (or only mild) left ventricular involvement Localized right ventricular aneurysms (akinetic or dyskinetic areas with diastolic bulgings) Severe segmental dilatation of the right ventricle	Mild global right ventricular dilatation and or ejection fraction reduction with normal left ventricle Mild segmental dilatation of the right ventricle Regional right ventricular hypokinesia
2. Tissue characteristics of walls	Fibrofatty replacement of myocardium on endomyocardial biopsy	
3. ECG depolarization/conduction abnormalities	Epsilon waves or localized prolongation (≥ 110 ms) of the QRS complex in the right precordial leads (V1–V3)	Late potentials seen on signal averaged electrocardiography
4. ECG repolarization abnormalities		Inverted T waves in right precordial leads (V2 and V3) in people > 12 years and in the absence of right bundle branch block
5. Arrhythmias		Sustained or nonsustained left bundle branch block type ventricular tachycardia documented on the electrocardiography, Holter monitoring, or during exercise testing Frequent ventricular extrasystoles (more than 1,000/24 h on Holter monitoring)
6. Family history	Familial disease confirmed at necropsy or surgery	Family history of premature sudden death (<35 years) due to suspected ARVC/D Family history (clinical diagnosis based on present criteria)

^aDiagnosis of ARVC requires the presence of at least two major criteria or one major and two minor criteria or four minor criteria.

LOCUS NAME	CAUSATIVE GENE	MODE OF INHERITANCE
ARVD-1	Transforming growth factor- β_3	Autosomal dominant
ARVD-2	Cardiac ryanodine receptor (RYR2)	Autosomal dominant
ARVD-3	Unknown	Autosomal dominant
ARVD-4	Unknown	Autosomal dominant
ARVD-5	Transmembrane protein-43	Autosomal dominant
ARVD-6	Unknown	Autosomal dominant
ARVD-7	Unknown	Autosomal dominant
ARVD-8	Desmoplakin (DSP)	Autosomal dominant
ARVD-9	Plakophilin-2 (PKP2)	Autosomal dominant
ARVD-10	Desmoglein-2	Autosomal dominant
ARVD-11	Desmocollin-2	Autosomal dominant
ARVD-12	Plakoglobin (JUP)	Autosomal dominant
Naxos disease	Plakoglobin (JUP)	Autosomal recessive
Carvajal syndrome	Desmoplakin (DSP)	Autosomal recessive

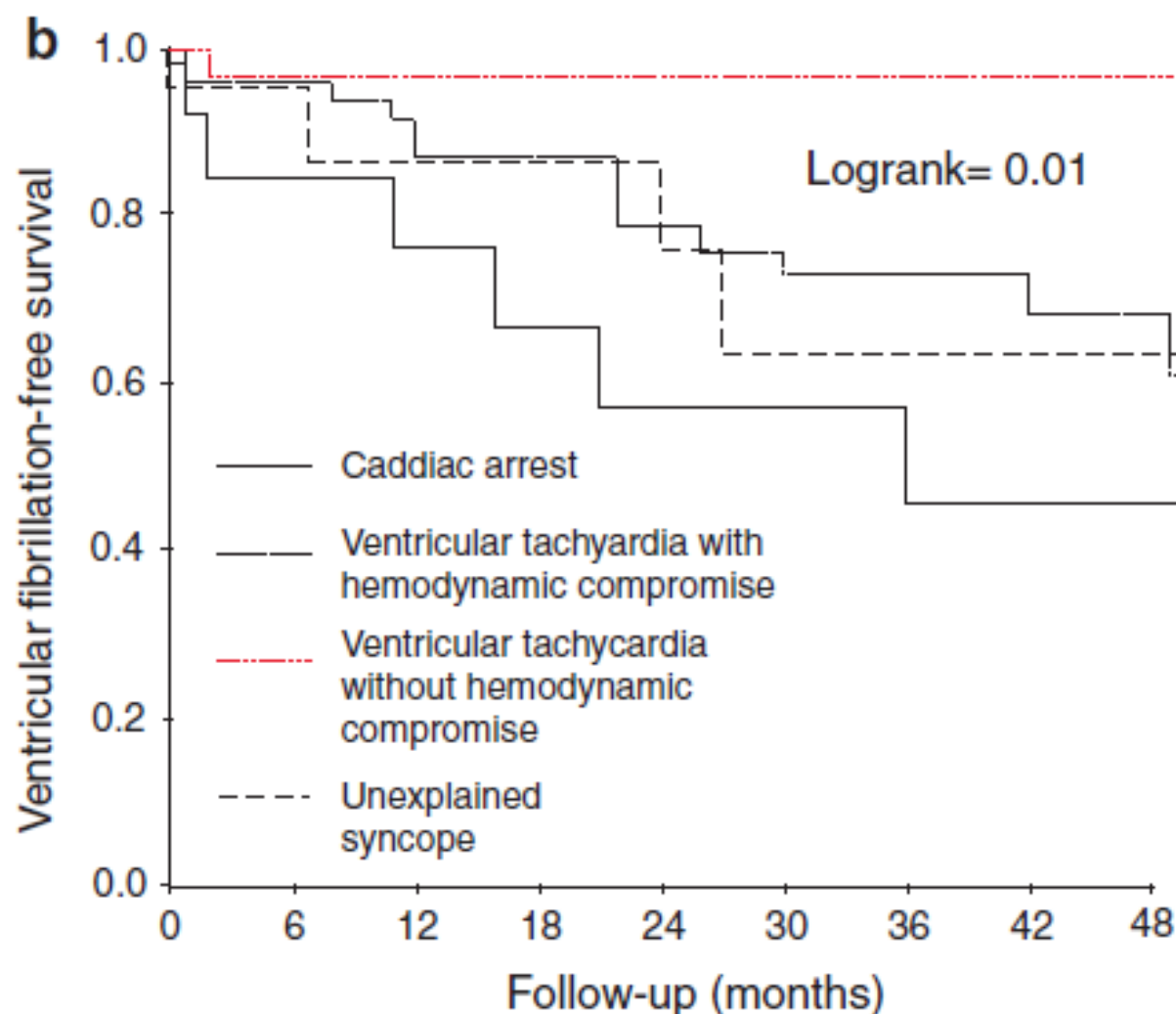
ORIGINAL TASK FORCE CRITERIA		REVISED TASK FORCE CRITERIA
I. Global or Regional Dysfunction and Structural Alterations*		
Major	- Severe dilation and reduction of RV ejection fraction with no (or only mild) LV impairment - Localized RV aneurysms (akinetic or dyskinetic areas with diastolic bulging) - Severe segmental dilation of the RV	By 2-D echo: - Regional RV akinesia, dyskinesia, or aneurysm <i>and</i> one of the following (end diastole): - PLAX RVOT ≥ 32 mm (corrected for body size [PLAX/BSA] ≥ 19 mm/m²) - PSAX RVOT ≥ 36 mm (corrected for body size [PSAX/BSA] ≥ 21 mm/m²) - or fractional area change $\leq 33\%$ By MRI: - Regional RV akinesia or dyskinesia or dyssynchronous RV contraction <i>and</i> one of the following: - Ratio of RV end-diastolic volume to BSA ≥ 110 mL/m² (men) or ≥ 100 mL/m² (women) - or RV ejection fraction $\leq 40\%$ By RV angiography: - Regional RV akinesia, dyskinesia, or aneurysm
Minor	- Mild global RV dilation and/or ejection fraction reduction with normal LV - Mild segmental dilation of the RV - Regional RV hypokinesia	By 2-D echo: - Regional RV akinesia or dyskinesia <i>and</i> one of the following (end diastole): - PLAX RVOT ≥ 29 to < 32 mm (corrected for body size [PLAX/BSA] ≥ 16 to < 19 mm/m²) - PSAX RVOT ≥ 32 to < 36 mm (corrected for body size [PSAX/BSA] ≥ 18 to < 21 mm/m²) - or fractional area change $> 33\%$ to $\leq 40\%$ By MRI: - Regional RV akinesia or dyskinesia or dyssynchronous RV contraction <i>and</i> one of the following: - Ratio of RV end-diastolic volume to BSA ≥ 100 to < 110 mL/m² (men) or ≥ 90 to < 100 mL/m² (women) - or RV ejection fraction $> 40\%$ to $\leq 45\%$
II. Tissue Characterization of Wall		
Major	Fibrofatty replacement of myocardium on endomyocardial biopsy	Residual myocytes $< 60\%$ by morphometric analysis (or $< 50\%$ if estimated) with fibrous replacement of the RV free wall myocardium in at least one sample, with or without fatty replacement of tissue on endomyocardial biopsy
Minor		Residual myocytes $< 60\%$ by morphometric analysis (or $< 50\%$ if estimated) with fibrous replacement of the RV free wall myocardium in at least one sample, with or without fatty replacement of tissue on endomyocardial biopsy
III. Repolarization Abnormalities		
Major		Inverted T waves in right precordial leads (V₁, V₂, and V₃) or beyond in individuals > 14 yr of age (in the absence of complete right bundle branch block QRS ≥ 120 msec)
Minor	Inverted T waves in right precordial leads (V₂ and V₃) (people age > 12 yr, in absence of right bundle branch block)	- Inverted T waves in leads V₁ and V₂ in individuals > 14 yr of age (in the absence of complete right bundle branch block) or in V₄, V₅, or V₆ - Inverted T waves in leads V₁, V₂, V₃, and V₄ in individuals > 14 yr of age in the presence of complete right bundle branch block
IV. Depolarization/Conduction Abnormalities		
Major	Epsilon waves or localized prolongation (> 110 msec) of the QRS complex in right precordial leads (V₁ to V₃)	Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V₁ to V₃)
Minor	Late potentials (SAECG)	- Late potentials by SAECG in at least one of three parameters in the absence of a QRS duration ≥ 110 msec on the standard ECG - Filtered QRS duration (fQRS) ≥ 114 msec - Duration of terminal QRS < 40 μV (low-amplitude signal duration) ≥ 38 msec - Root-mean-square voltage of terminal 40 msec ≥ 20 μV - Terminal activation duration of QRS ≥ 55 msec measured from the nadir of the S wave to the end of the QRS, including R', in V₁, V₂, or V₃, in the absence of complete right bundle branch block
V. Arrhythmias		
Major		Nonsustained or sustained ventricular tachycardia of left bundle branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)
Minor	- Left bundle branch block-type ventricular tachycardia (sustained and nonsustained) (ECG, Holter, exercise) - Frequent ventricular extrasystoles (> 1000 per 24 hr) (Holter)	- Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle branch block morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL) or of unknown axis > 500 ventricular extrasystoles per 24 hr (Holter)

Continued

Critérios diagnóstico DAVD

Table 9.3 ARVC: summary of revised Task Force Criteria (Marcus *et al.*, 2010).

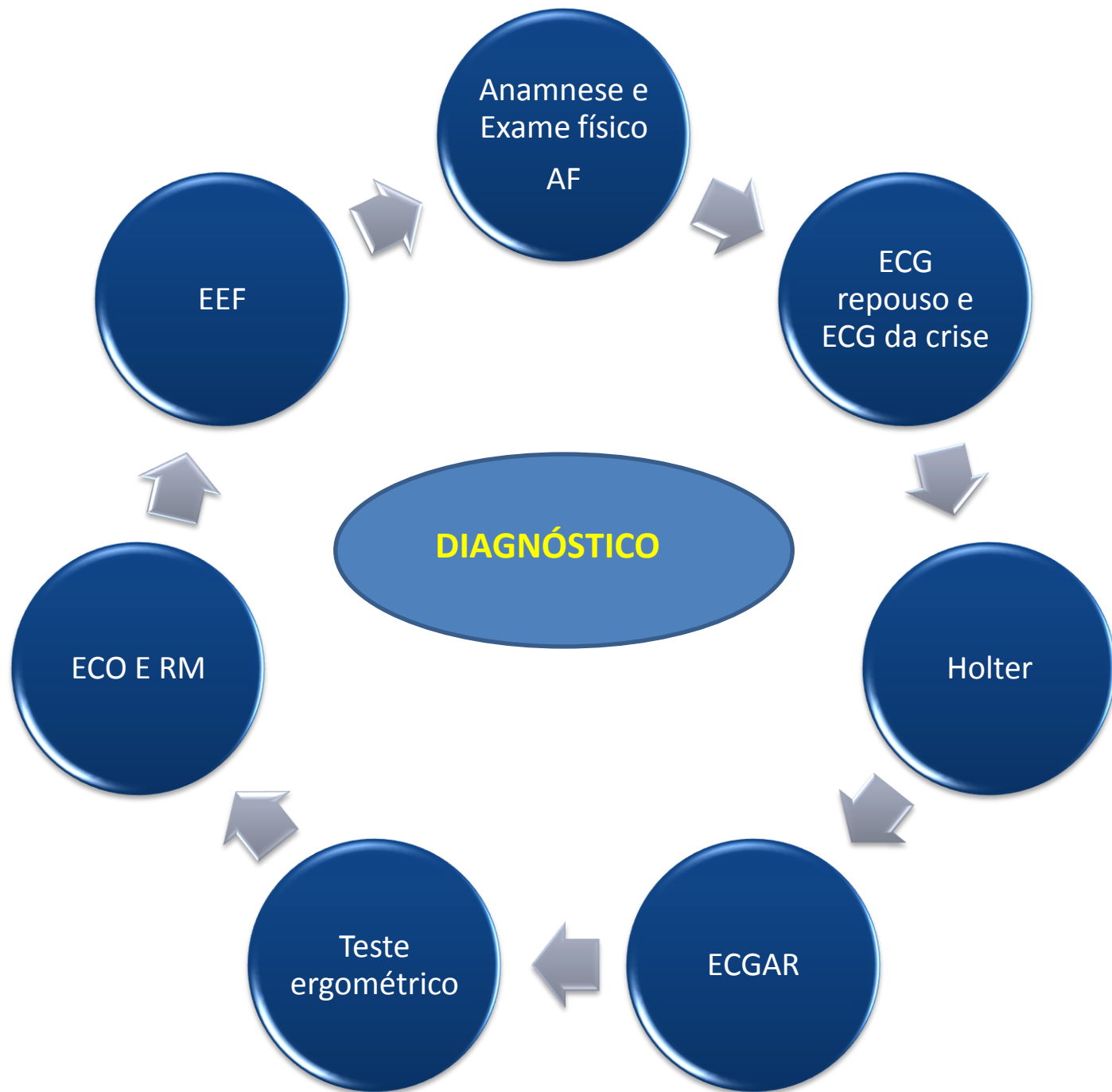
I. Global or regional dysfunction And structural alterations	● By 2D echo, MRI or RV angiography: A Regional RV akinesia, dyskinesia or aneurysm ● By MRI: ↓RV ejection fraction (<40% major, 40–45% minor dysfunction)
II. Repolarization abnormalities	● Major: Inverted T wave (V1–V3) or beyond in individuals >14 y. No RBBB (Figure 9.7A) ● Minor: Inverted T wave only in V1–V2 in individuals >14 y. No RBBB Inverted T waves in V1–V4 individuals >14 y, in presence of RBBB (Figure 9.6)
III. Depolarization abnormalities	● Epsilon wave in V1–V3 (Figure 9.8) ● Localized prolongation of QRS in V1–V3 (>110 ms) ● Late potentials. In the absence QRS ≥110 ms
IV. Arrhythmias	● Non-sustained or sustained VT (LBBB morphology superior axis) Major criteria (Figure 9.7) ● Non-sustained or sustained VT (LBBB morphology inferior axis). Minor criteria ● >500 PVC per 24 h. (Holter) Minor criteria
V. Family history	● History of ARVC/D in a first-degree relative ● Premature sudden death (<35 years of age) due to suspected ARVC/D in a first-degree relative ● ARVC/D confirmed pathologically or by current Task Force Criteria in second-degree relative



Corrado D, Leoni L, Link MS, et al. Implantable cardioverter-defibrillator therapy for prevention of sudden death in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia. *Circulation*. 2003;108:3084–3091

Diagnóstico diferencial de Displasia arritmogênica do VD

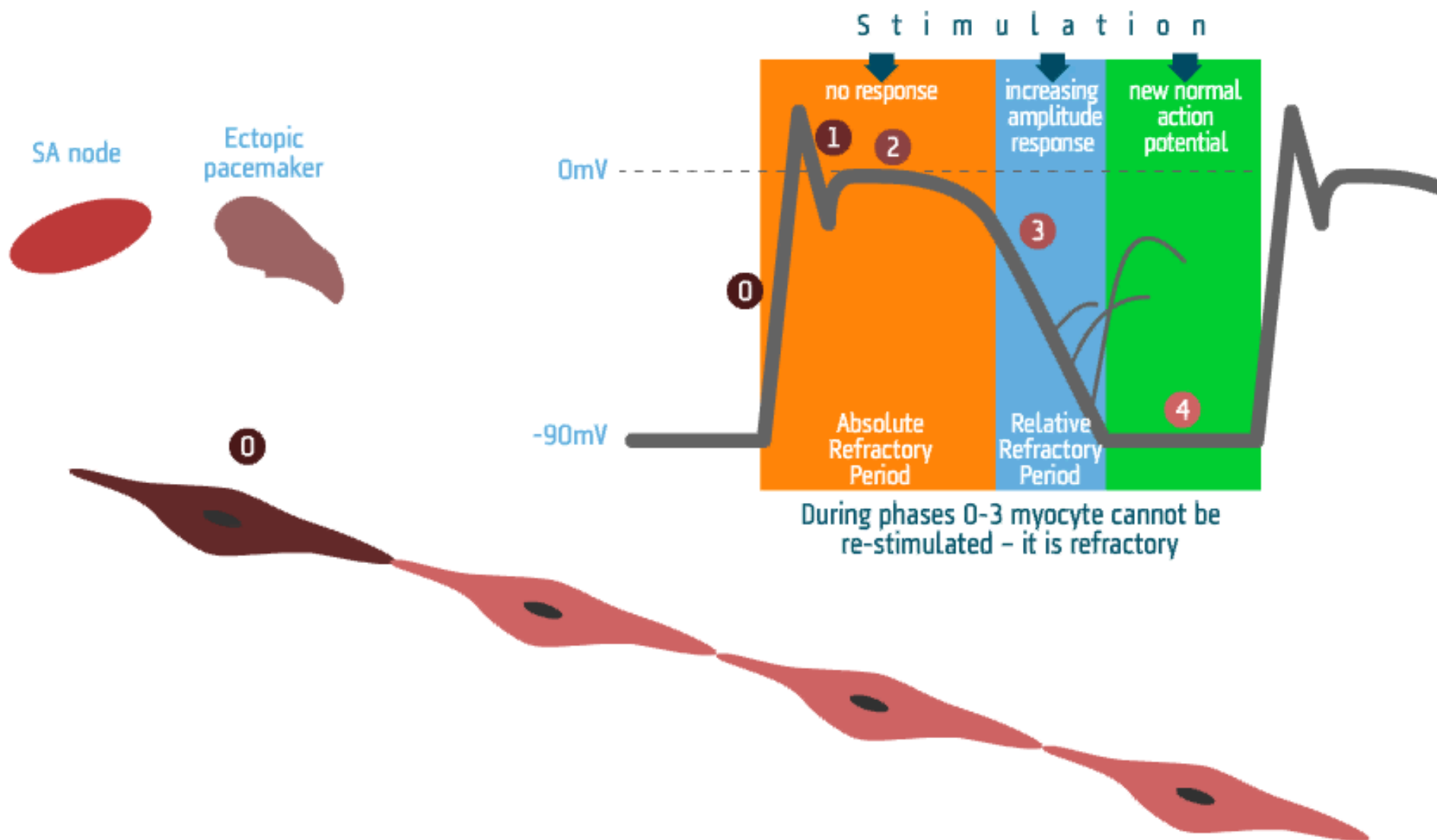
	TV idiopática de VD		DAVD
História Familiar de MS	Não		•Frequentemente sim
Arritmias	EV, TVNS, TVS Em repouso ou exercício		•idem
Morfologia onda T	T normais		•T negativas V1 a V3
Duração do QRS	Menor que 110 ms		•Maior que 110 ms
Onda Epsilon	ausente		•Presente em 30%
Nadir S final QRS > 55 ms	Raramente 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



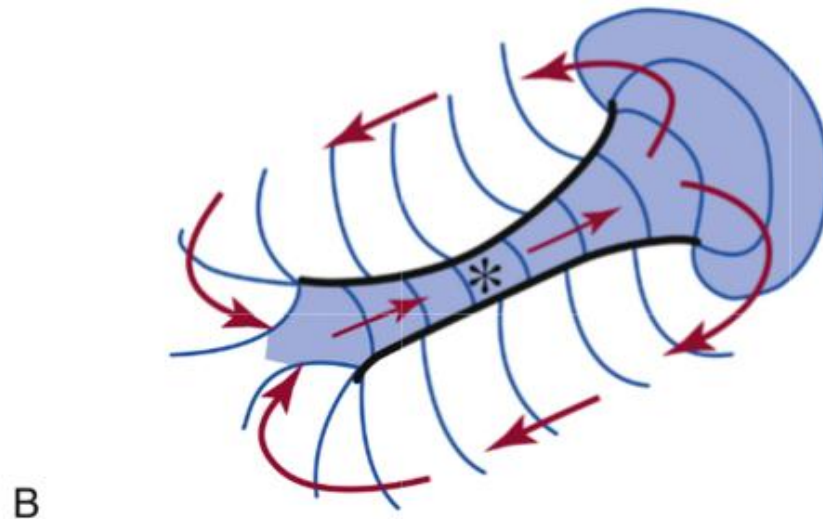
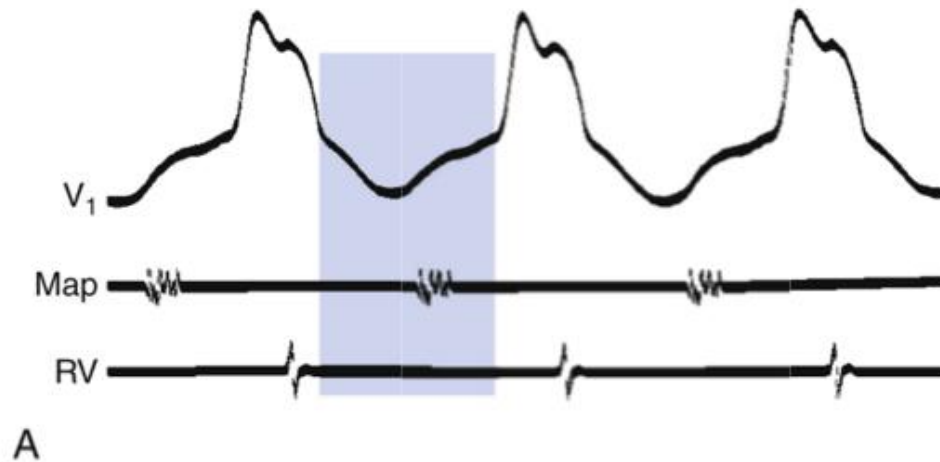


Diagnóstico diferencial de Displasia arritmogênica do VD

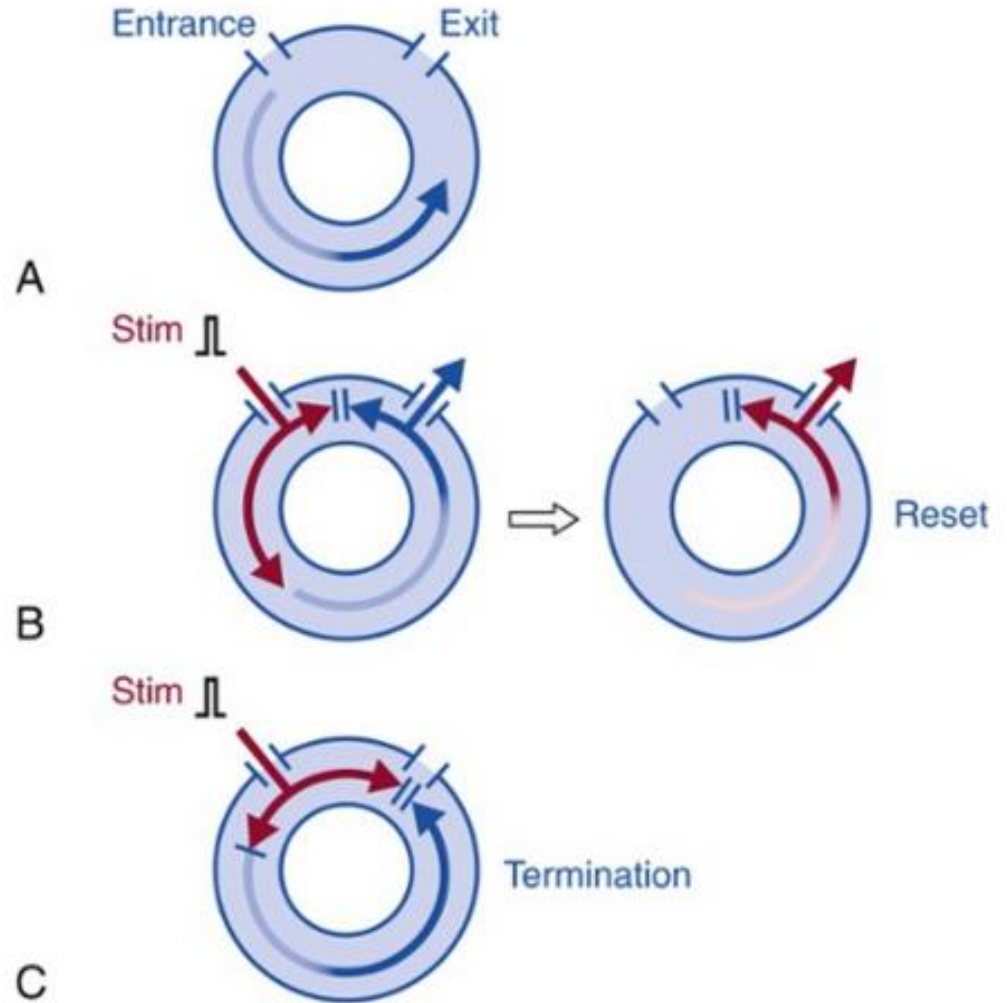
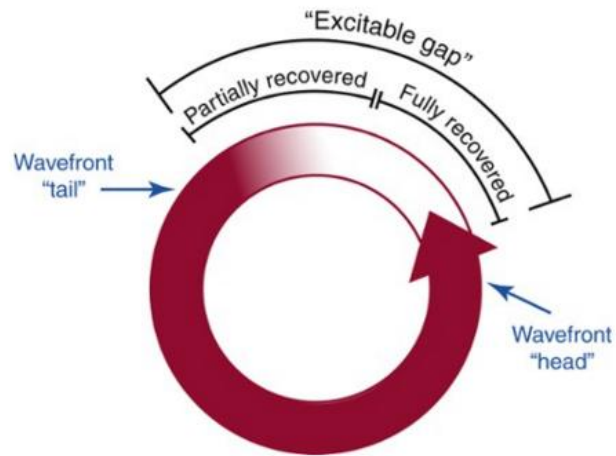
	TV idiopática de VD	DAVD
Resposta à terapia	Manobra vagal, adenosina Beta bloq, verapamil	•Sotalol, amiodarona com ou sem betabloqueadores
Ablação por cateter	Geralmente curativa	•Raramente curativa mas pode modificar o substrato e permitir melhor ação das drogas •Diferentes morfologias tendem a ocorrer



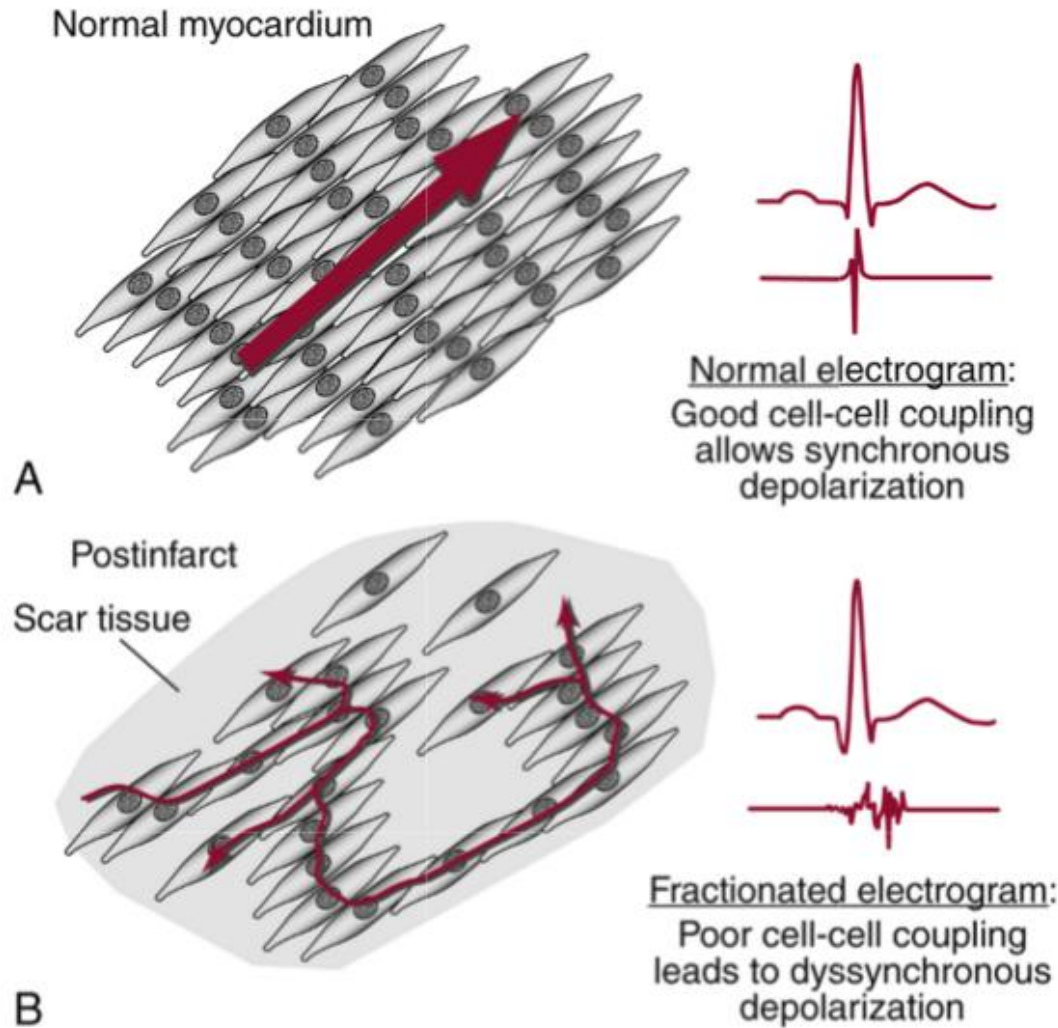
Reentrada e o istmo do circuito



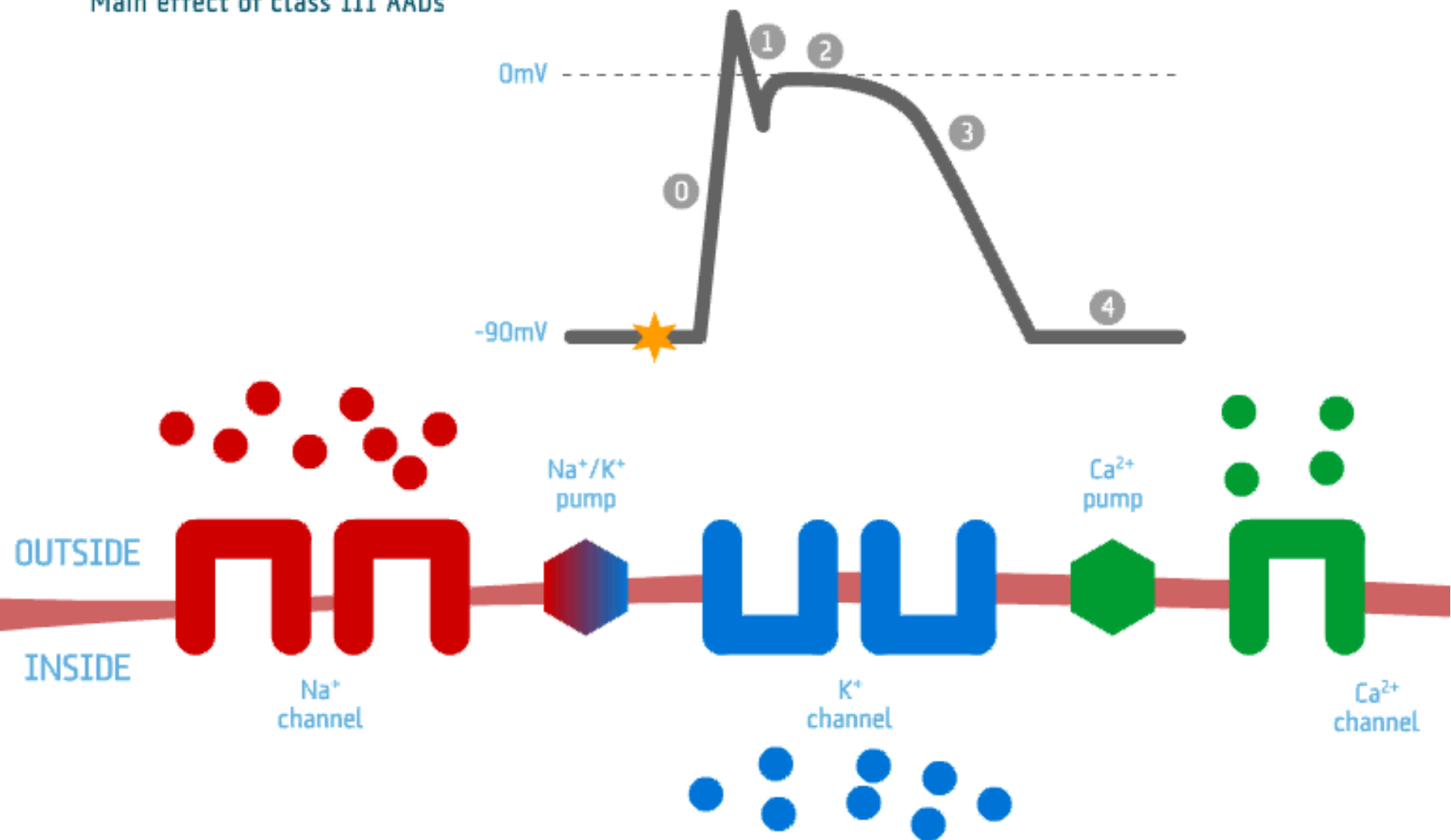
Importância para encarrilhamento e estimulação



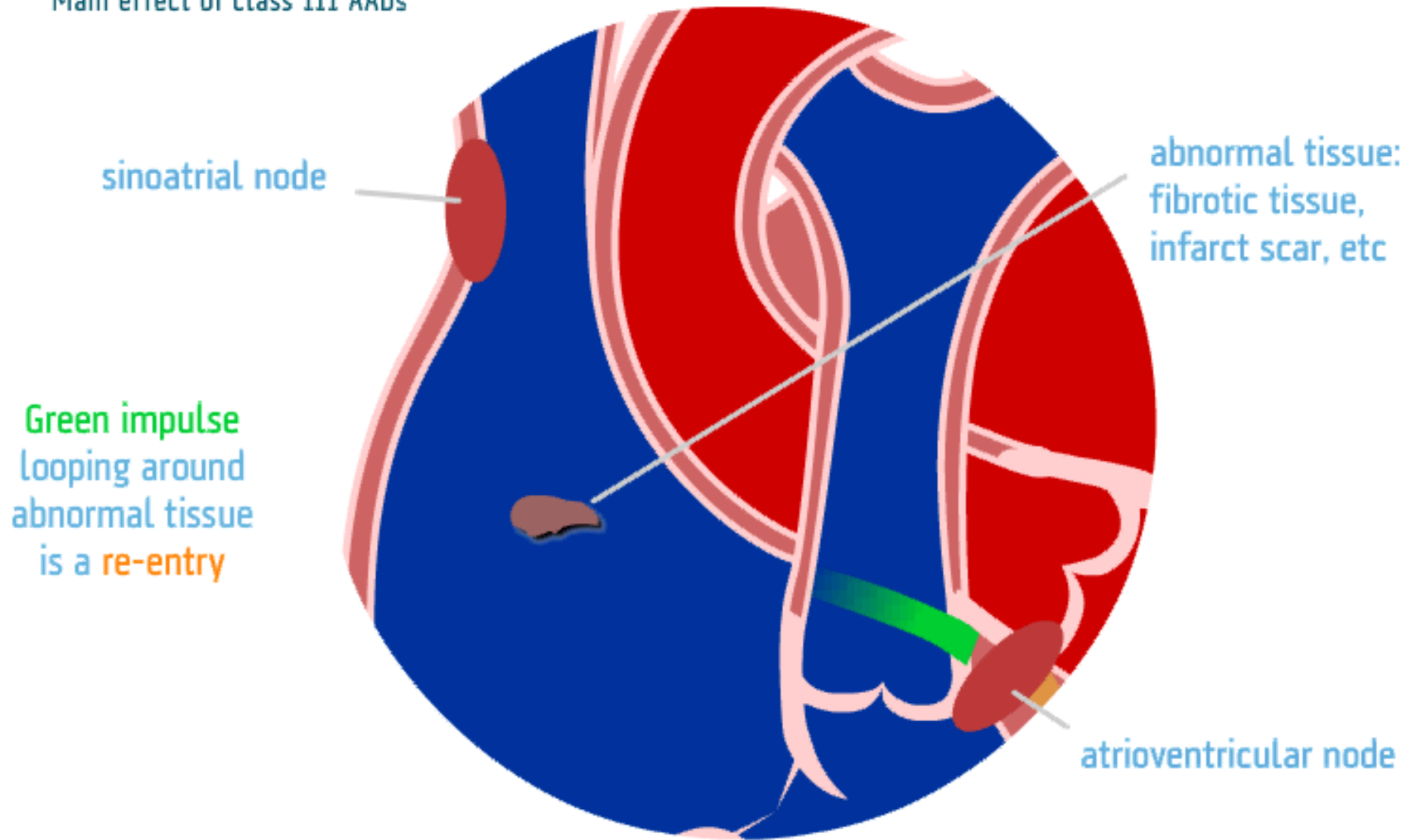
Cicatrices e reentrada



Main effect of class III AADs



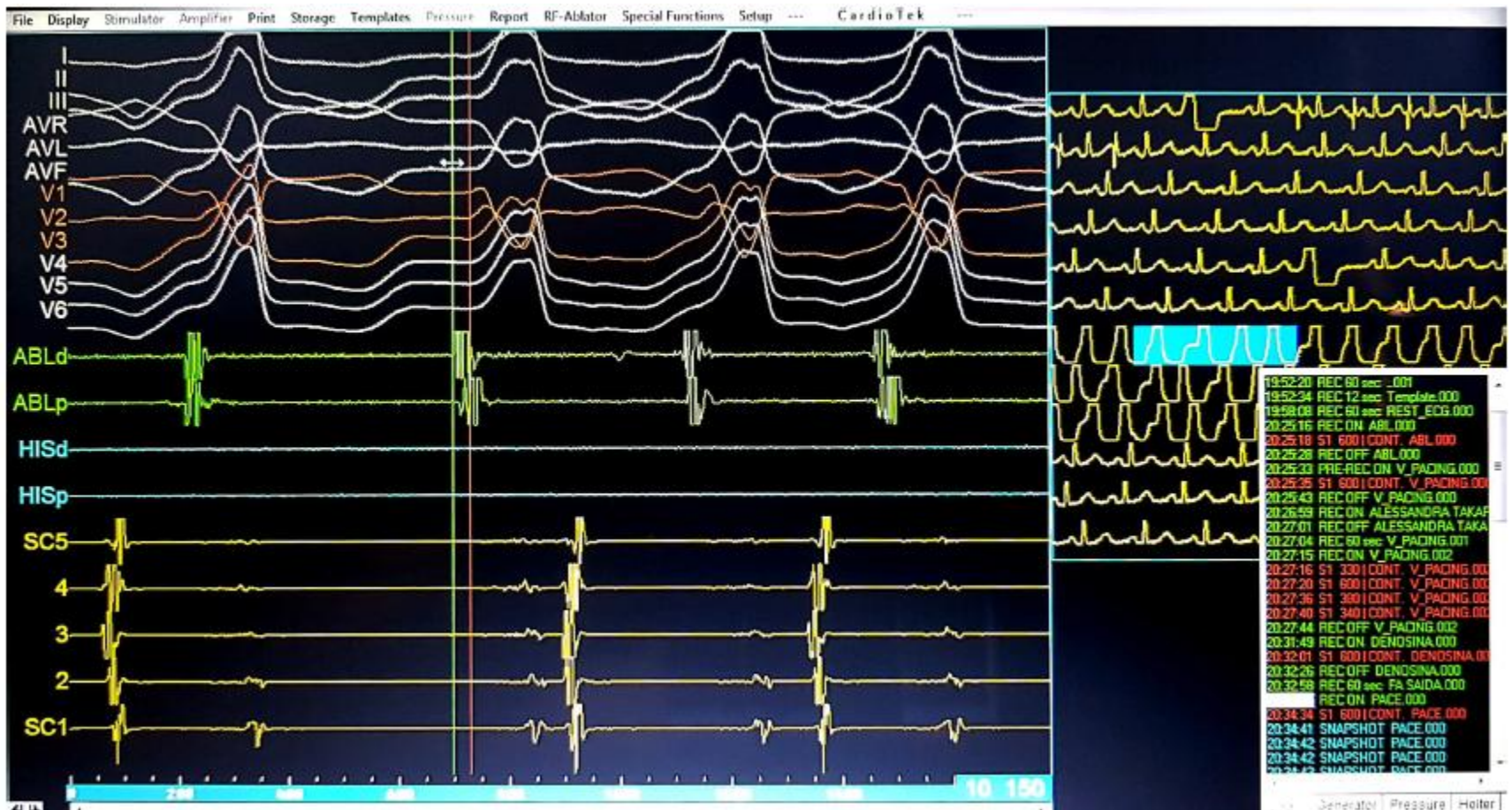
Main effect of class III AADs



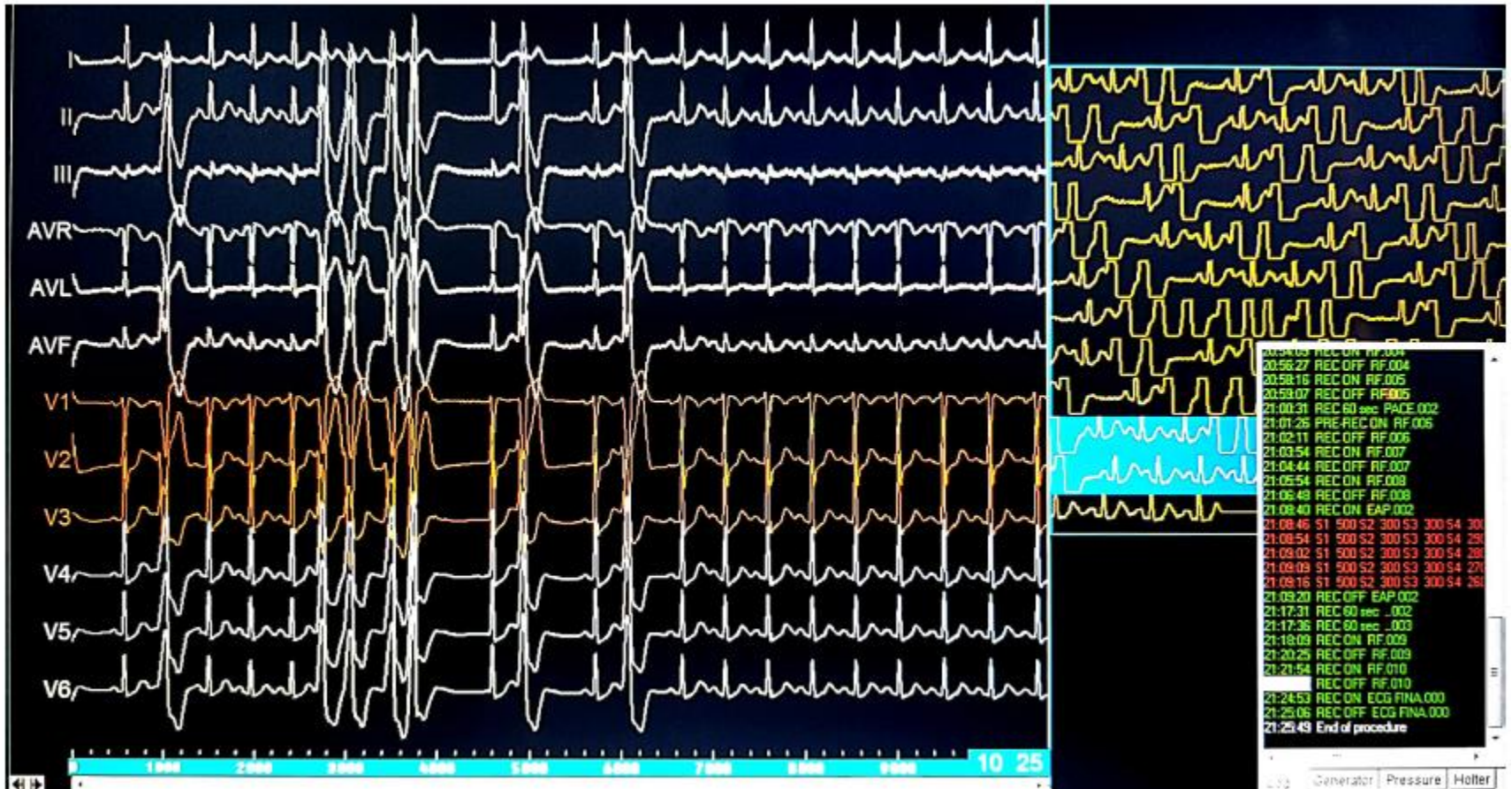




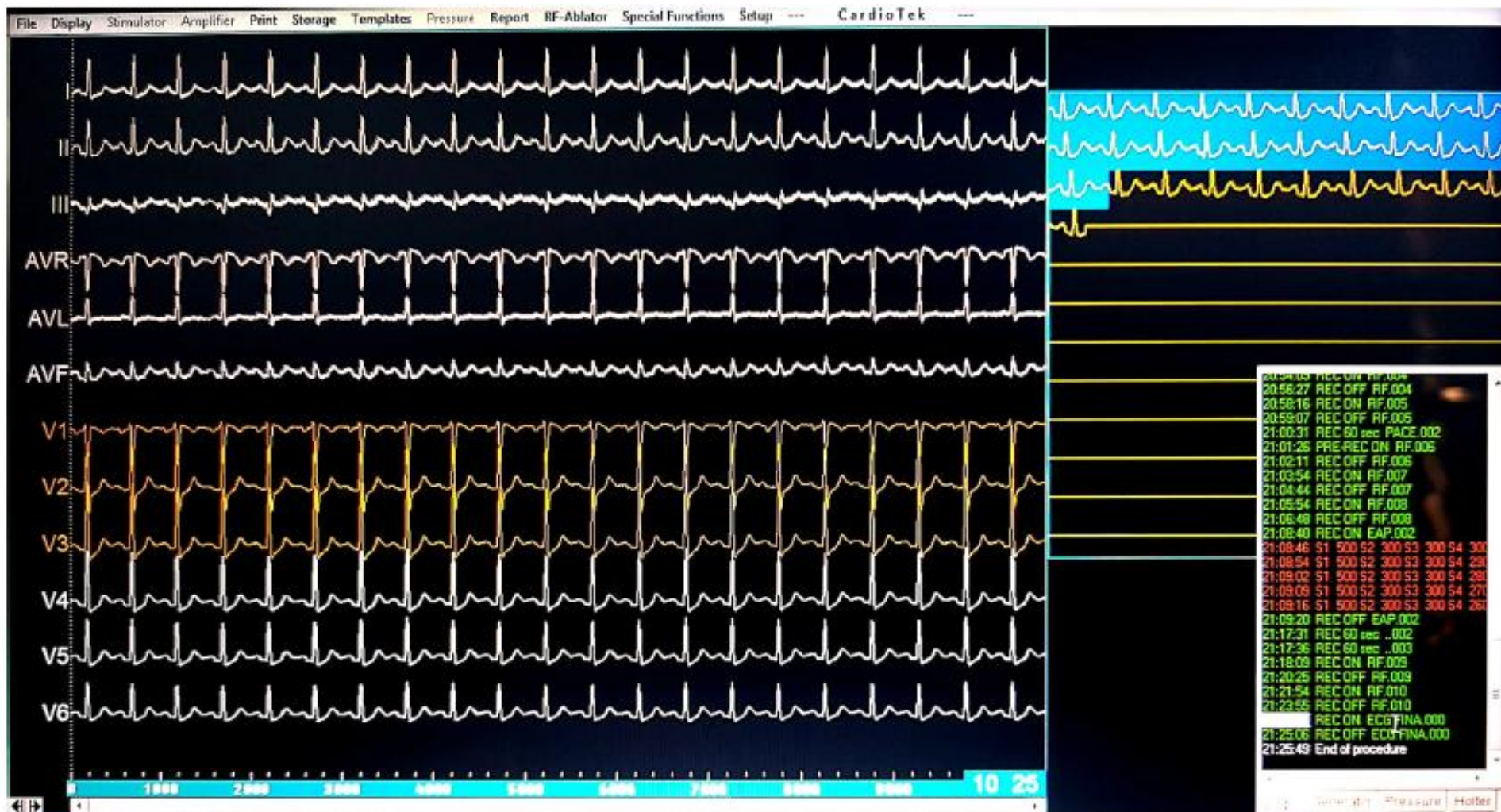
Avaliação da precocidade no ablator

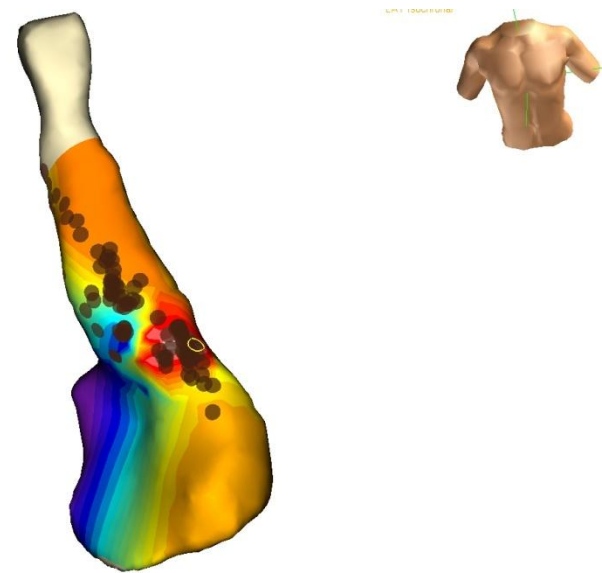
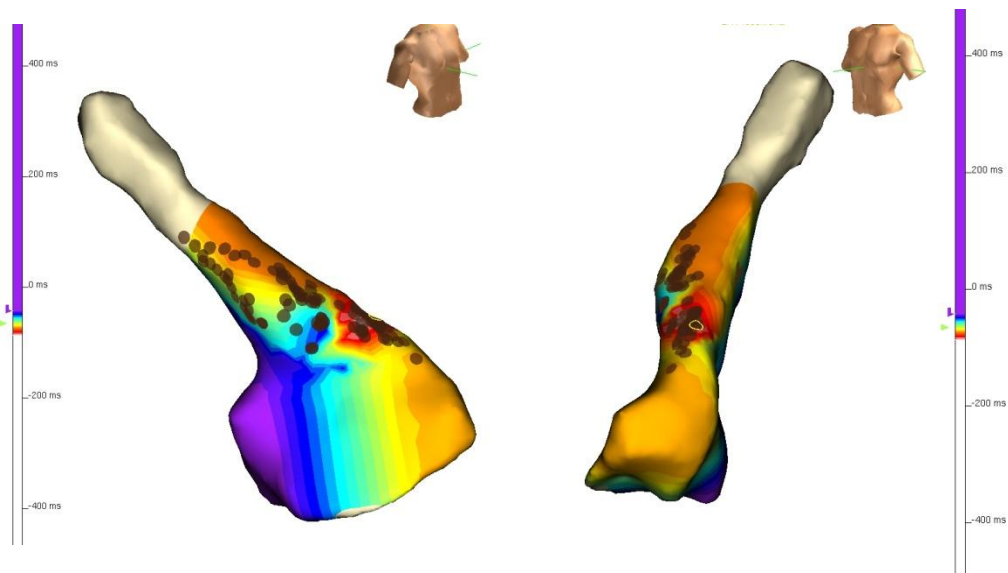
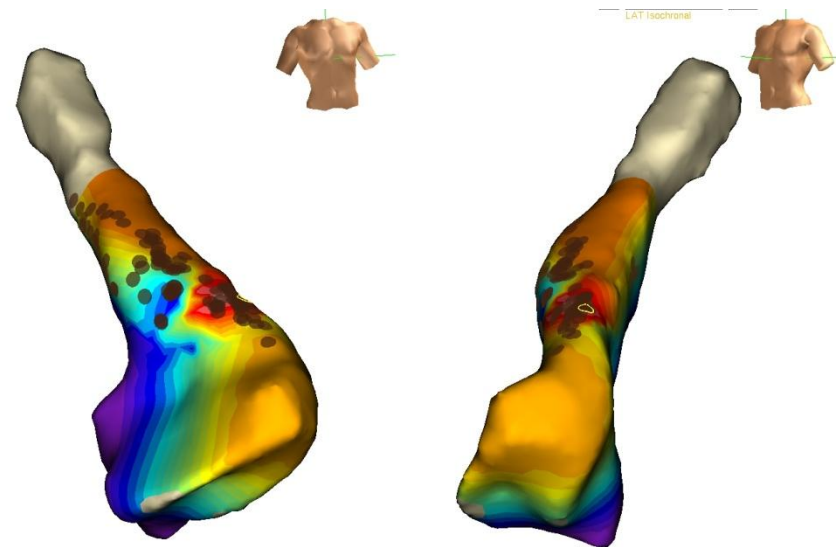
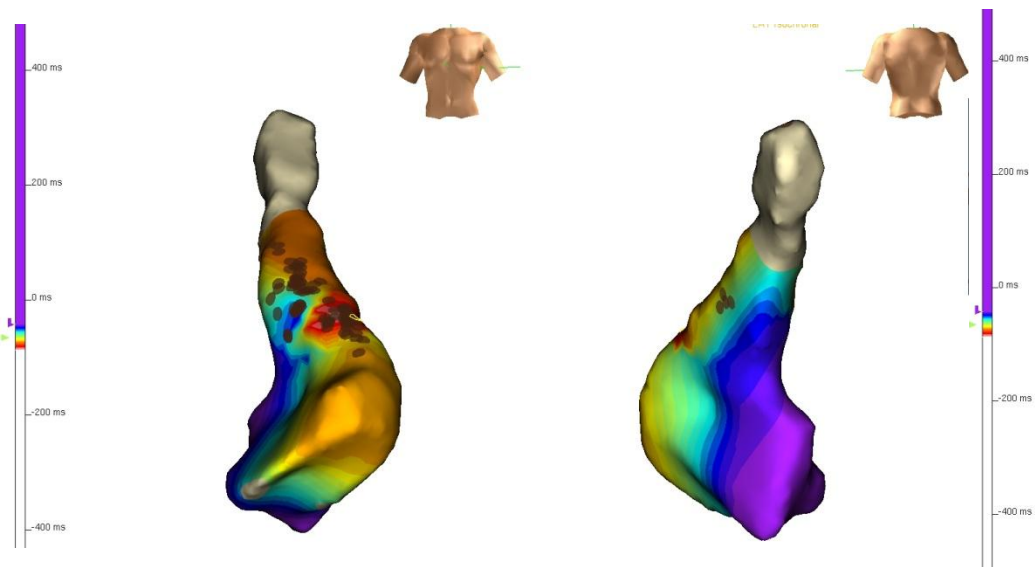


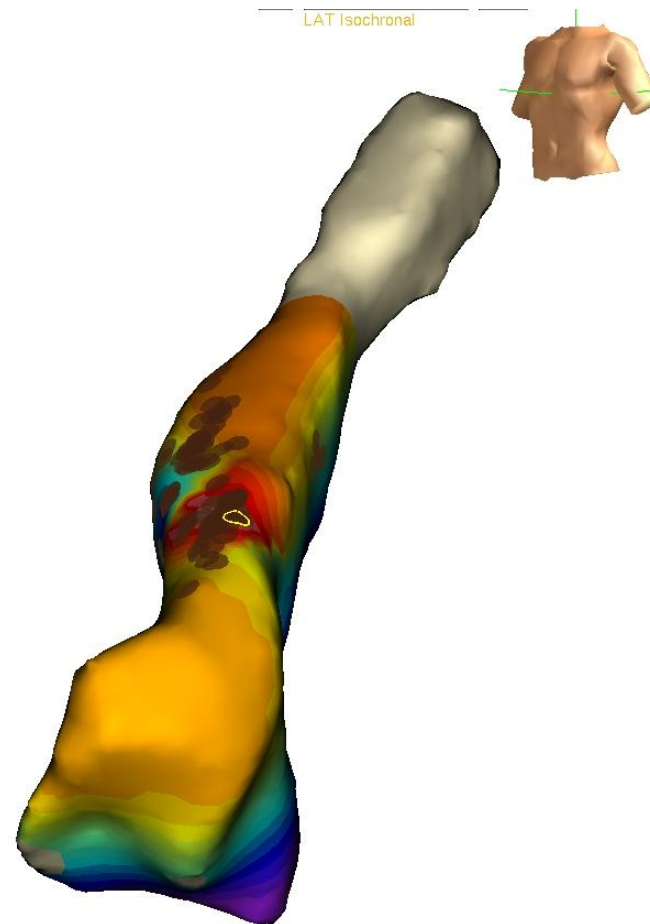
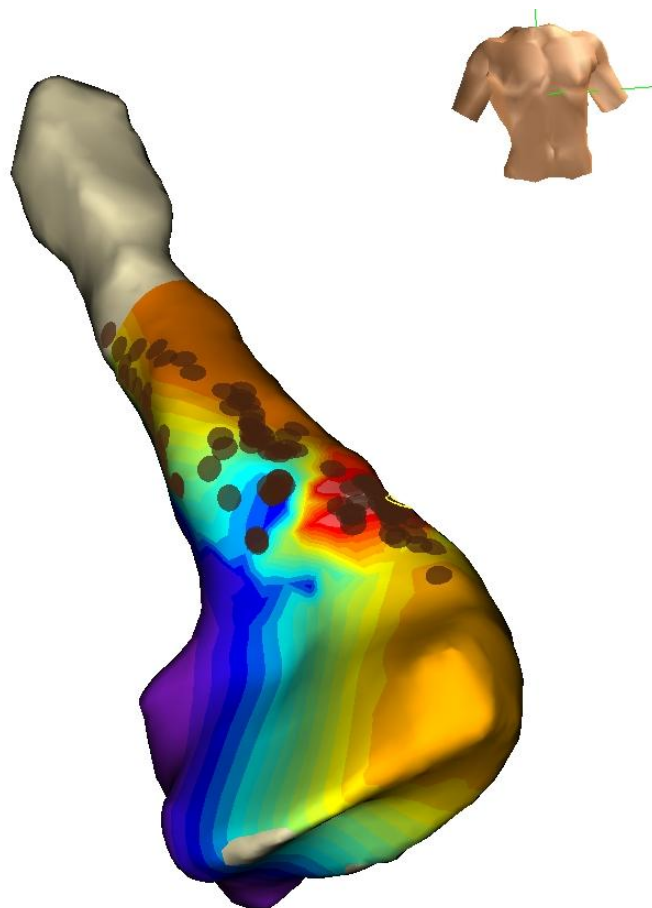
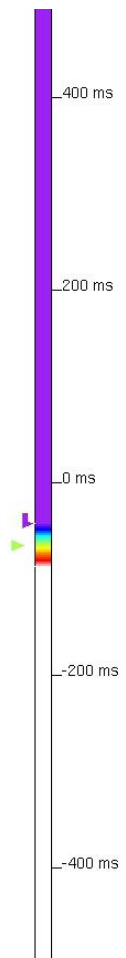
Aceleração do foco durante a aplicação



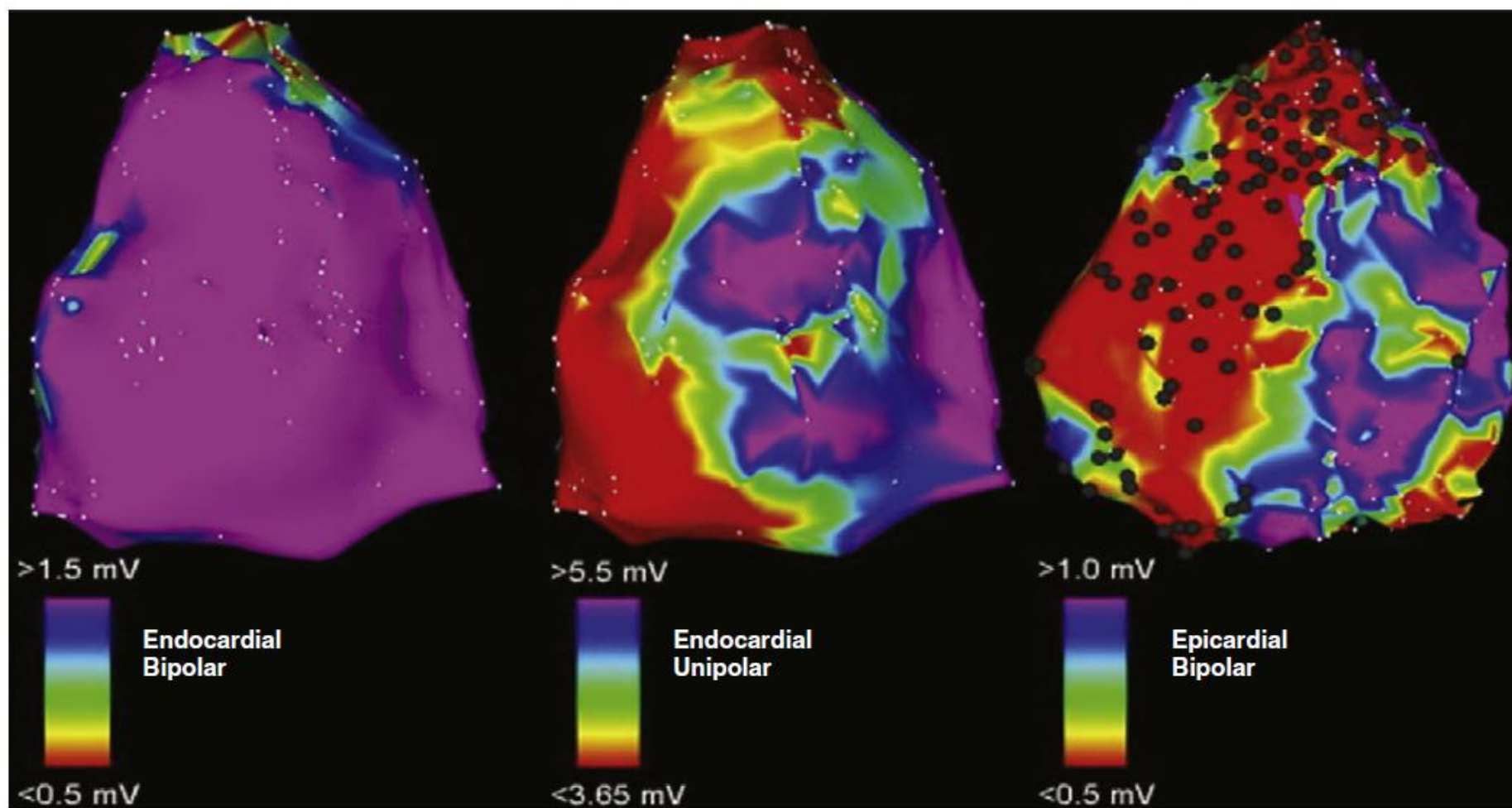
Final do procedimento







LAT Isochronal



Estratificação de risco Displasia arritmogênica do ventrículo direito

	I	Ila	Ilb
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	-	-



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

Obrigado!!!

rogerio.andalaft@einstein.br

Download em

www.arrytmiaonline.com.br

