TROUBLES DU RYTHME VENTRICULAIRE

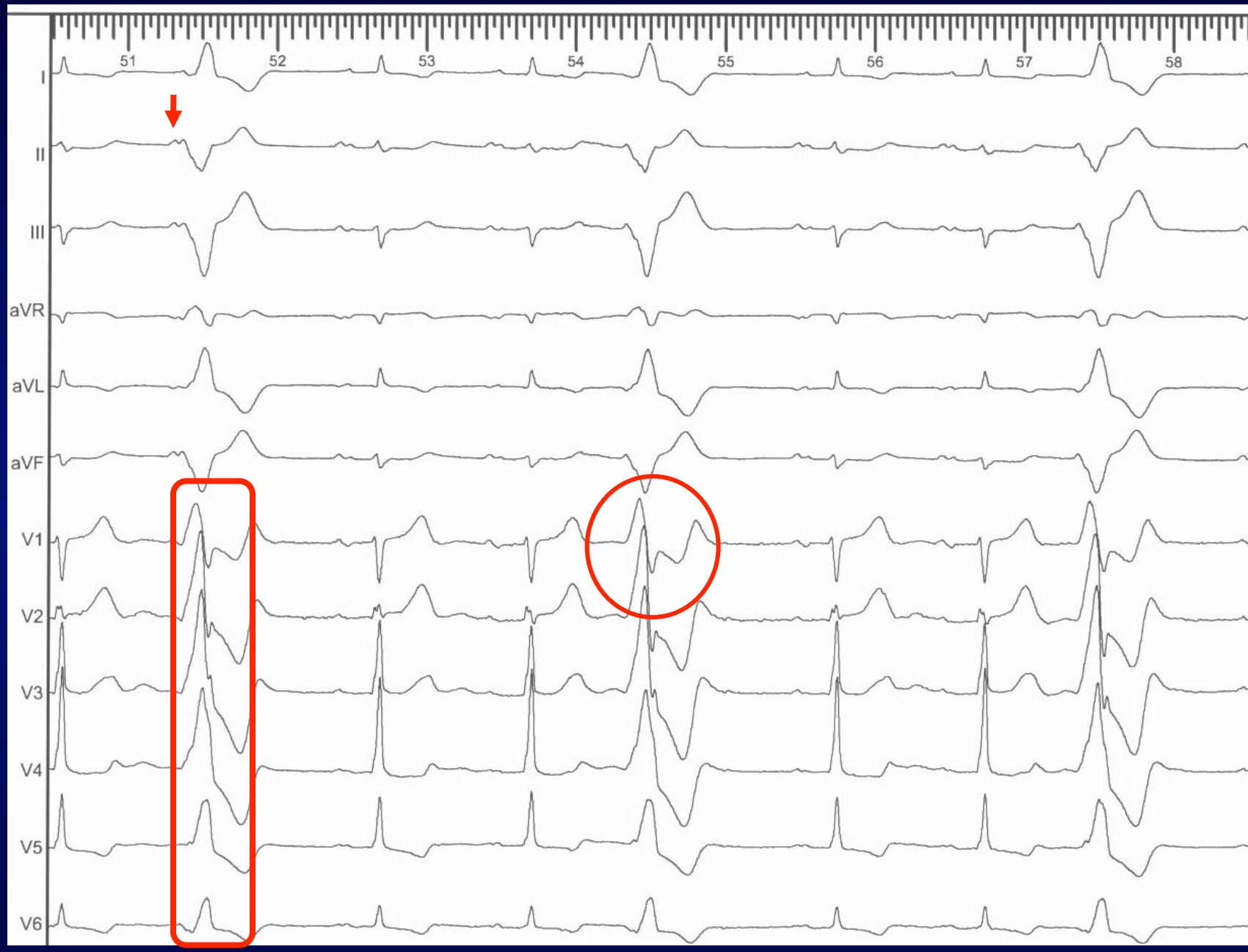
Quelques questions pratiques

Prof Luc DE ROY

CLINIQUES UNIVERSITAIRES UCL
CHU NAMUR
Mont-Godinne

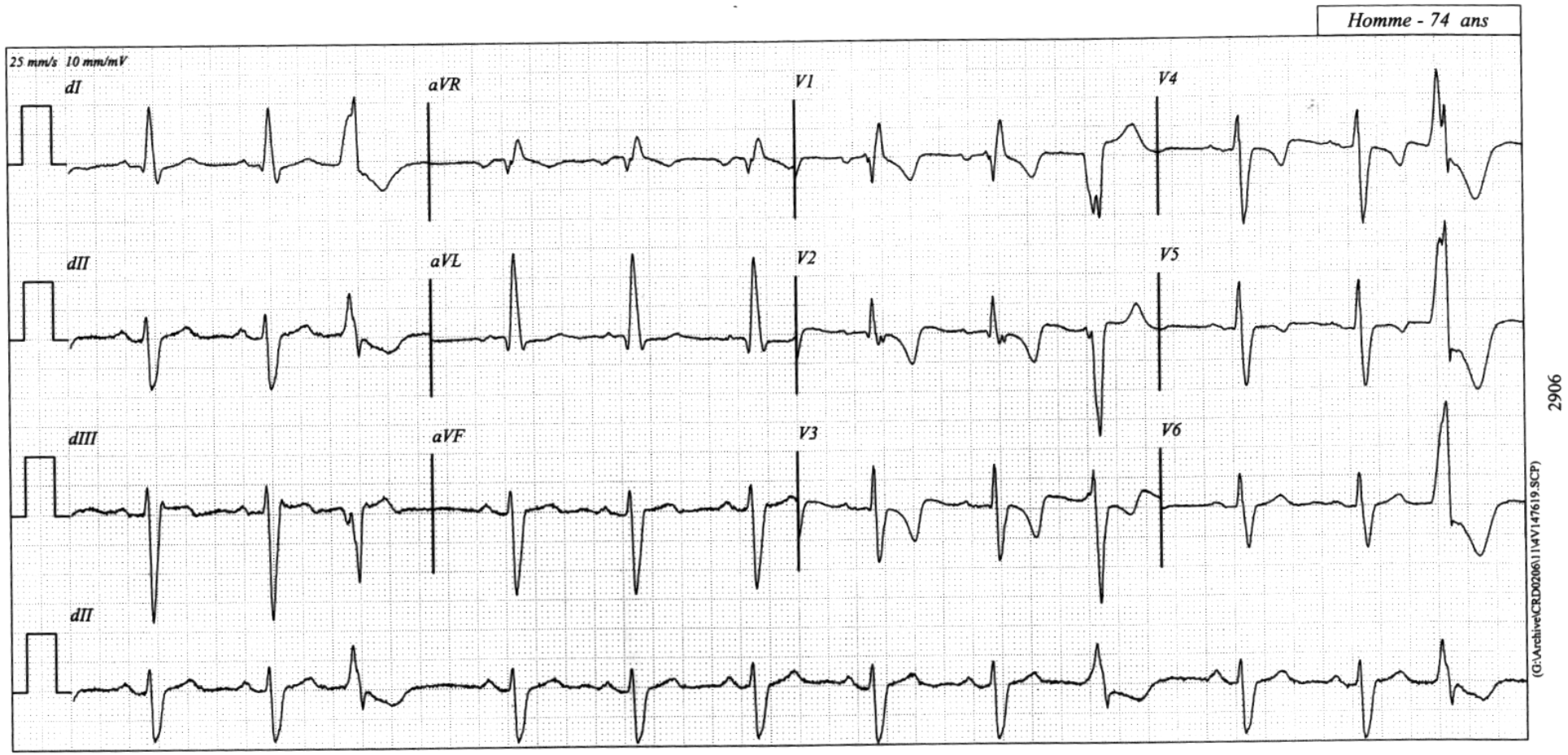
LES ESV

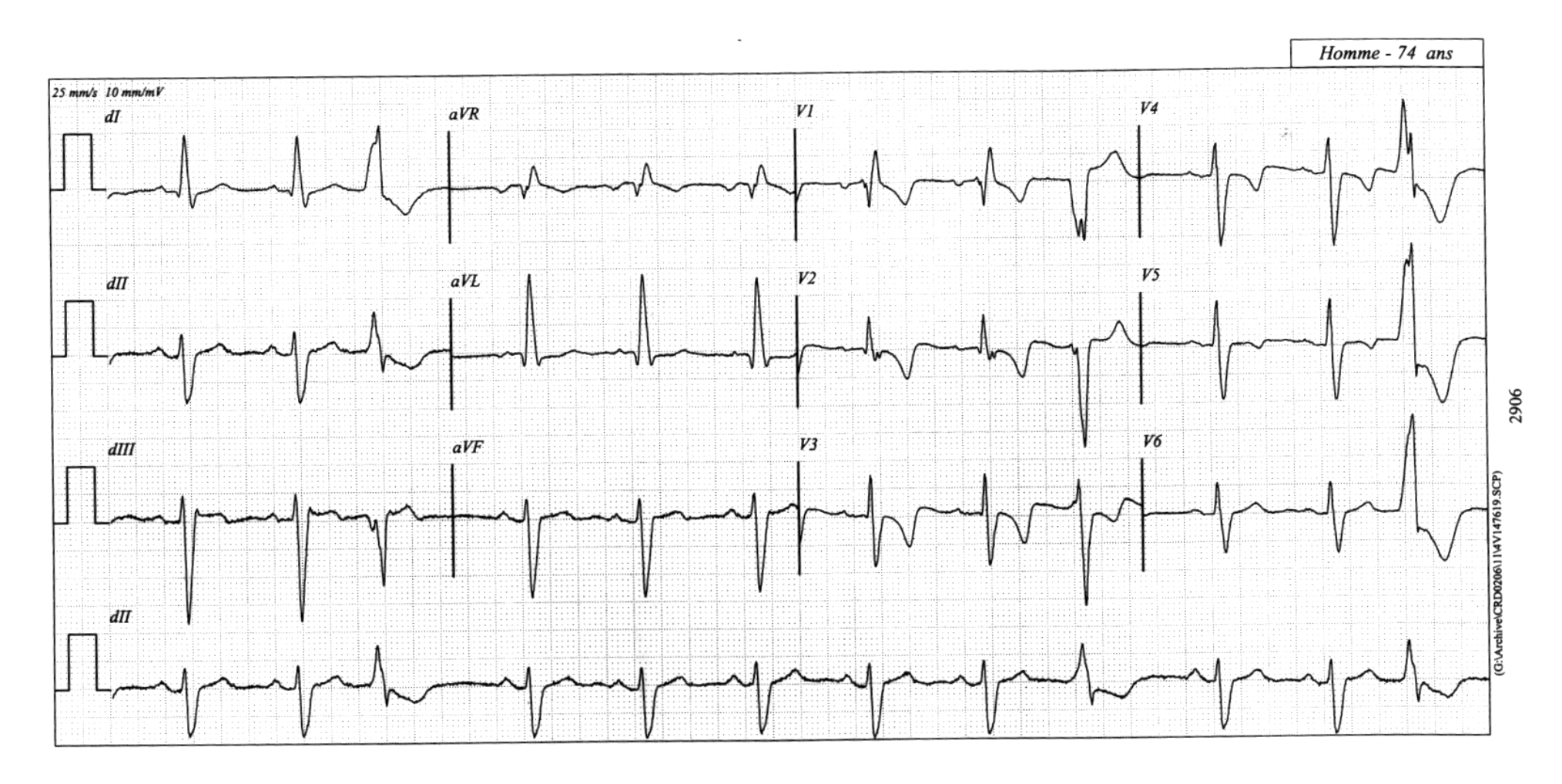
ESV: QRS larges dissociés,
concordance positive,
aspect monophasique V1



ESV: **QRS larges dissociés, concordance positive, aspect monophasique V1**

Présence de Cardiopathie?





En RS

BBD + axe G

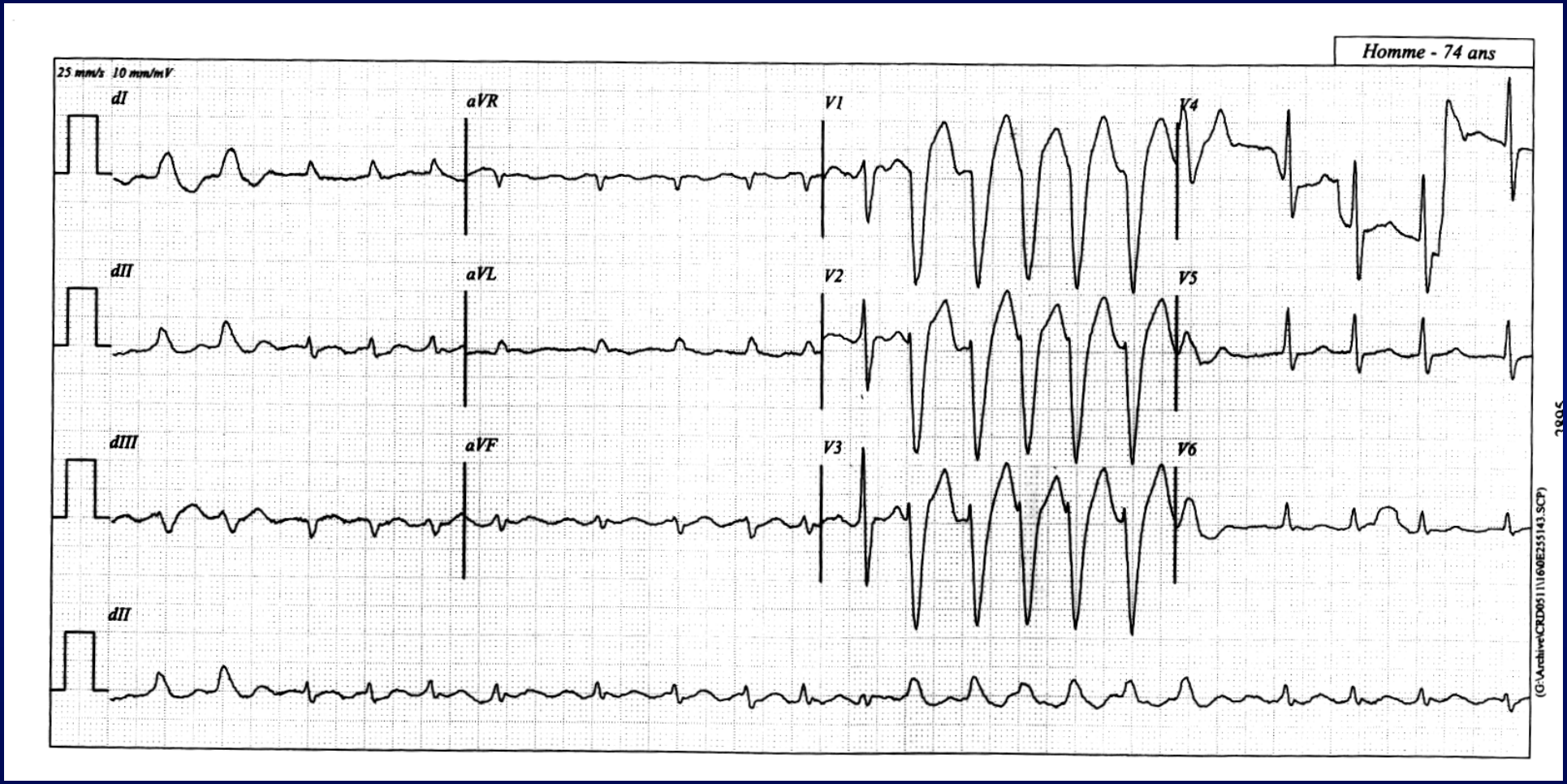
qR en V2

Repolarisation perturbée

ESV: crochetées, atypiques

ESV Polymorphes

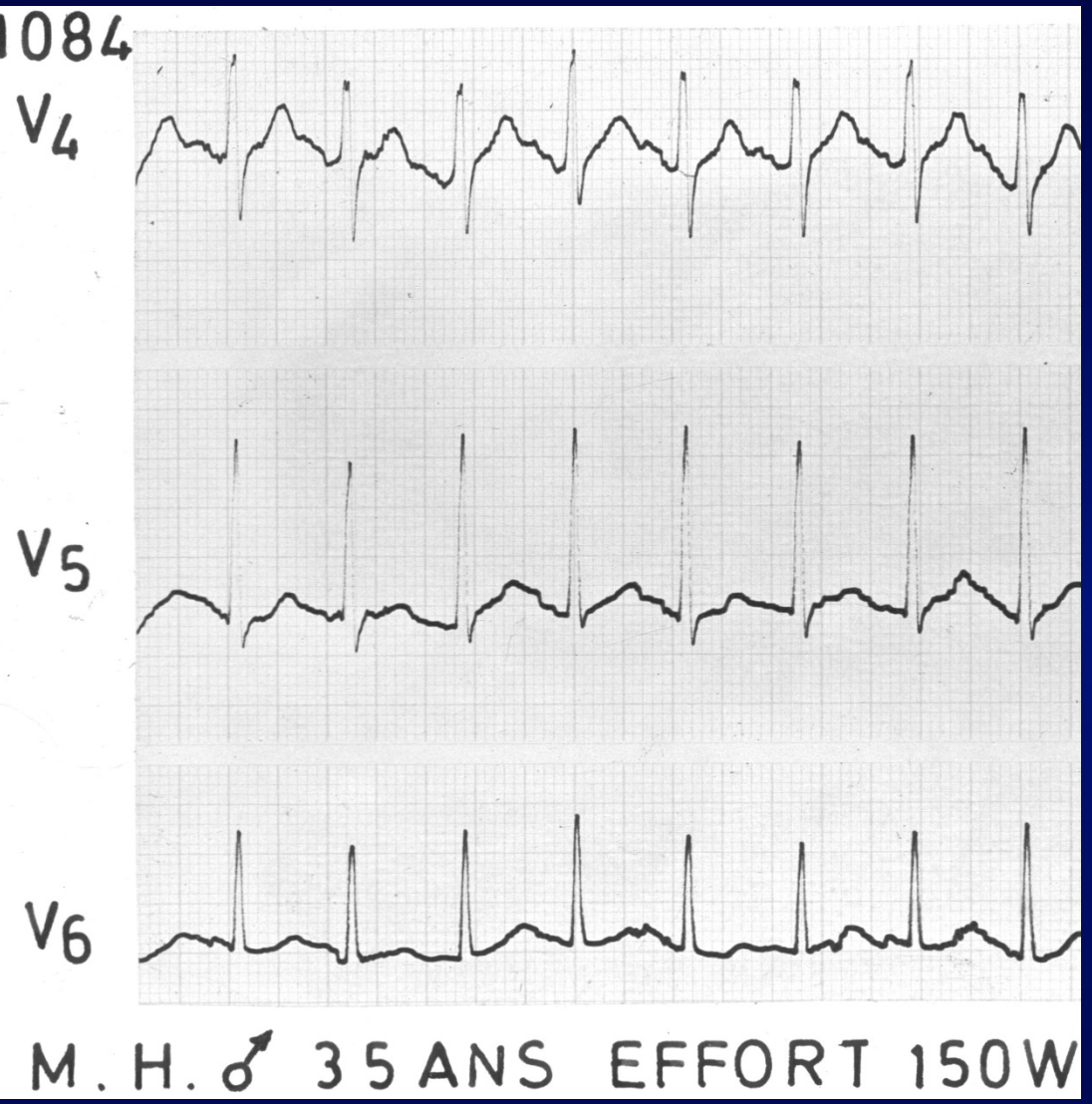
! DD Aberration !

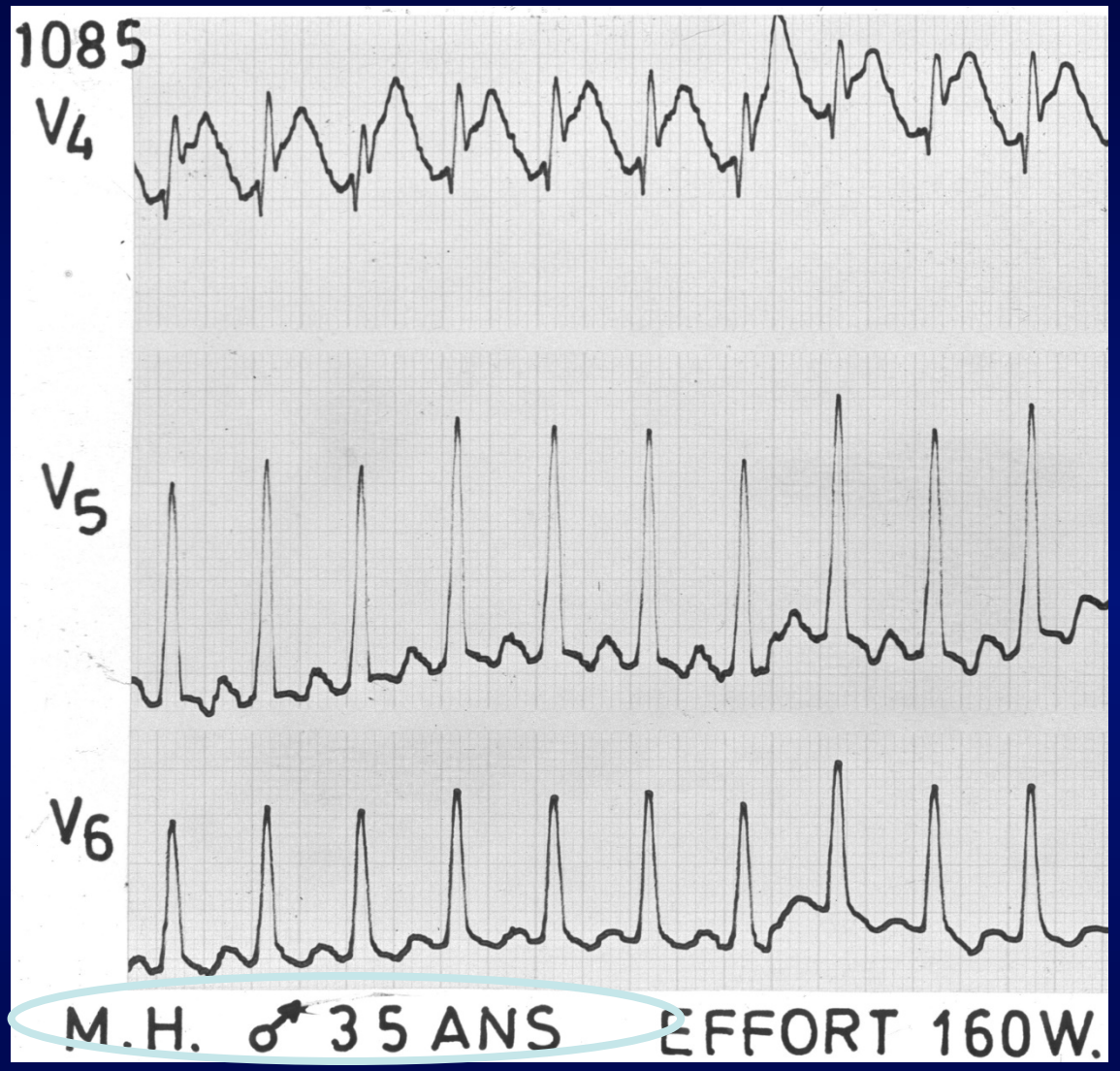


ESV et Test d'Effort

Les ESV à l'effort: signe défavorable?

Les ESV disparaissant à l'effort: signe favorable?



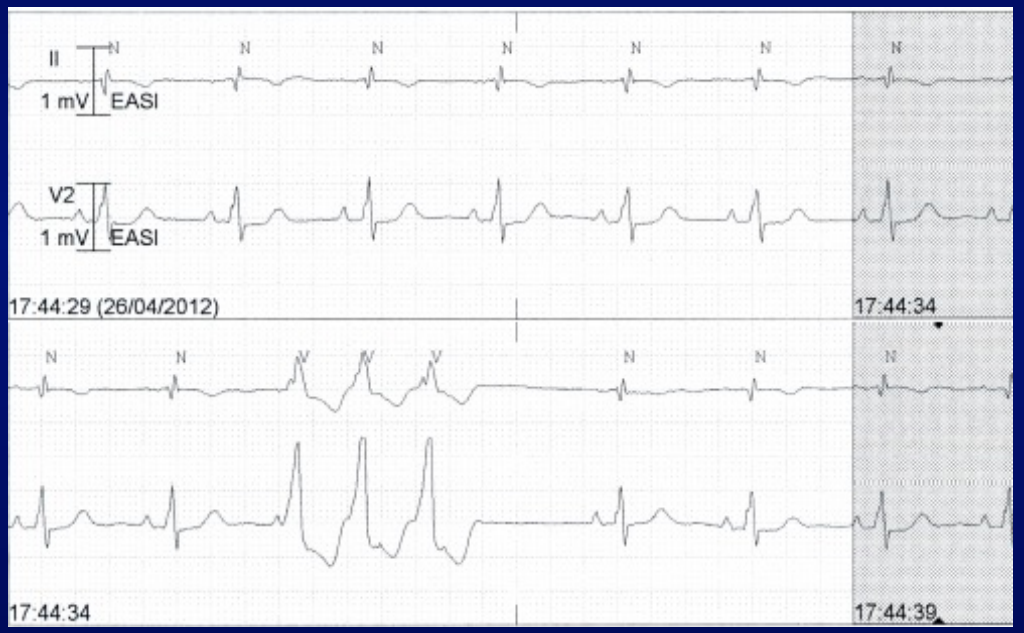


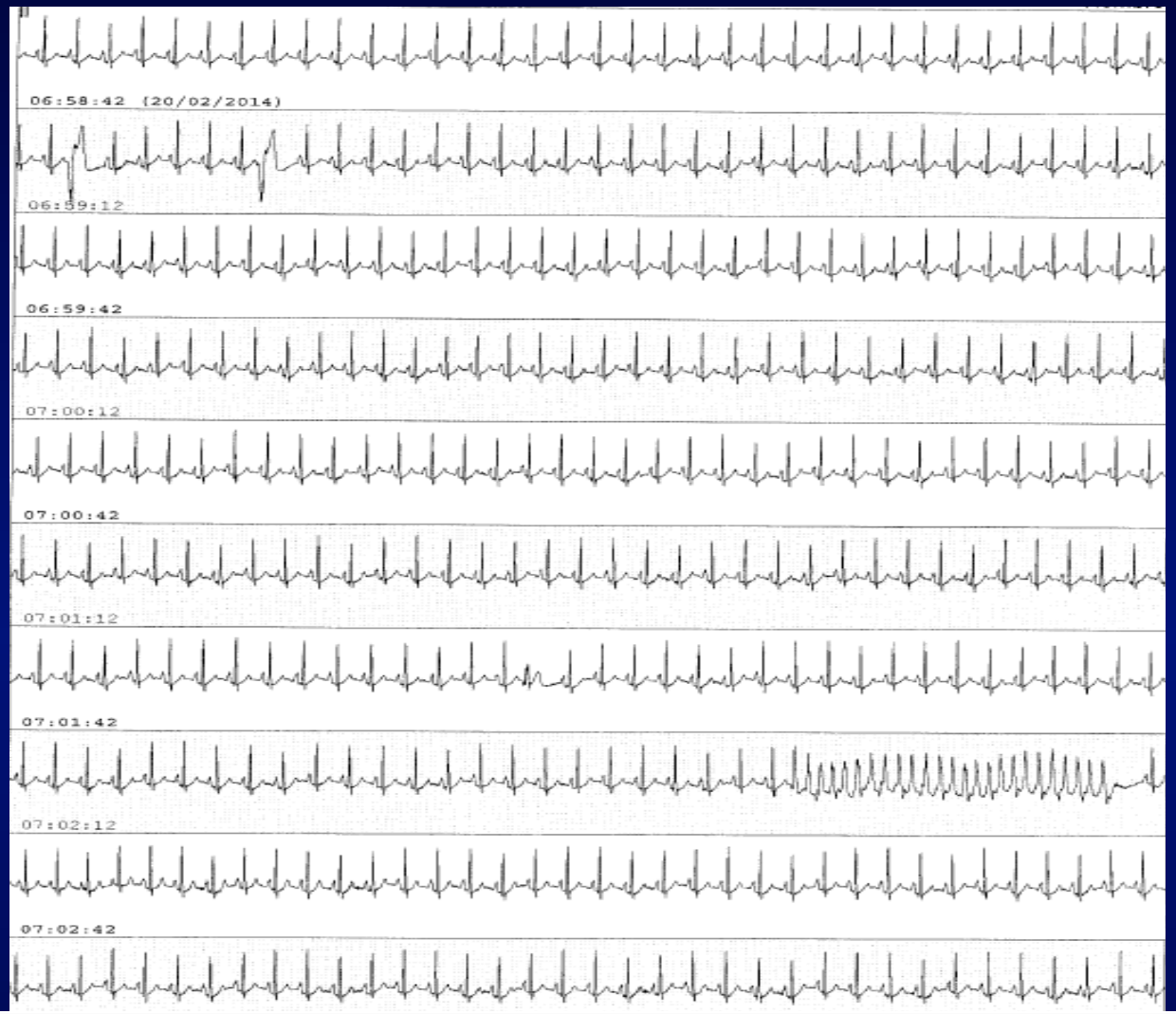
Angor lors de l'Effort

Angor lors de l'Effort

TV non soutenues

≥ 3 ESV
< 30 sec

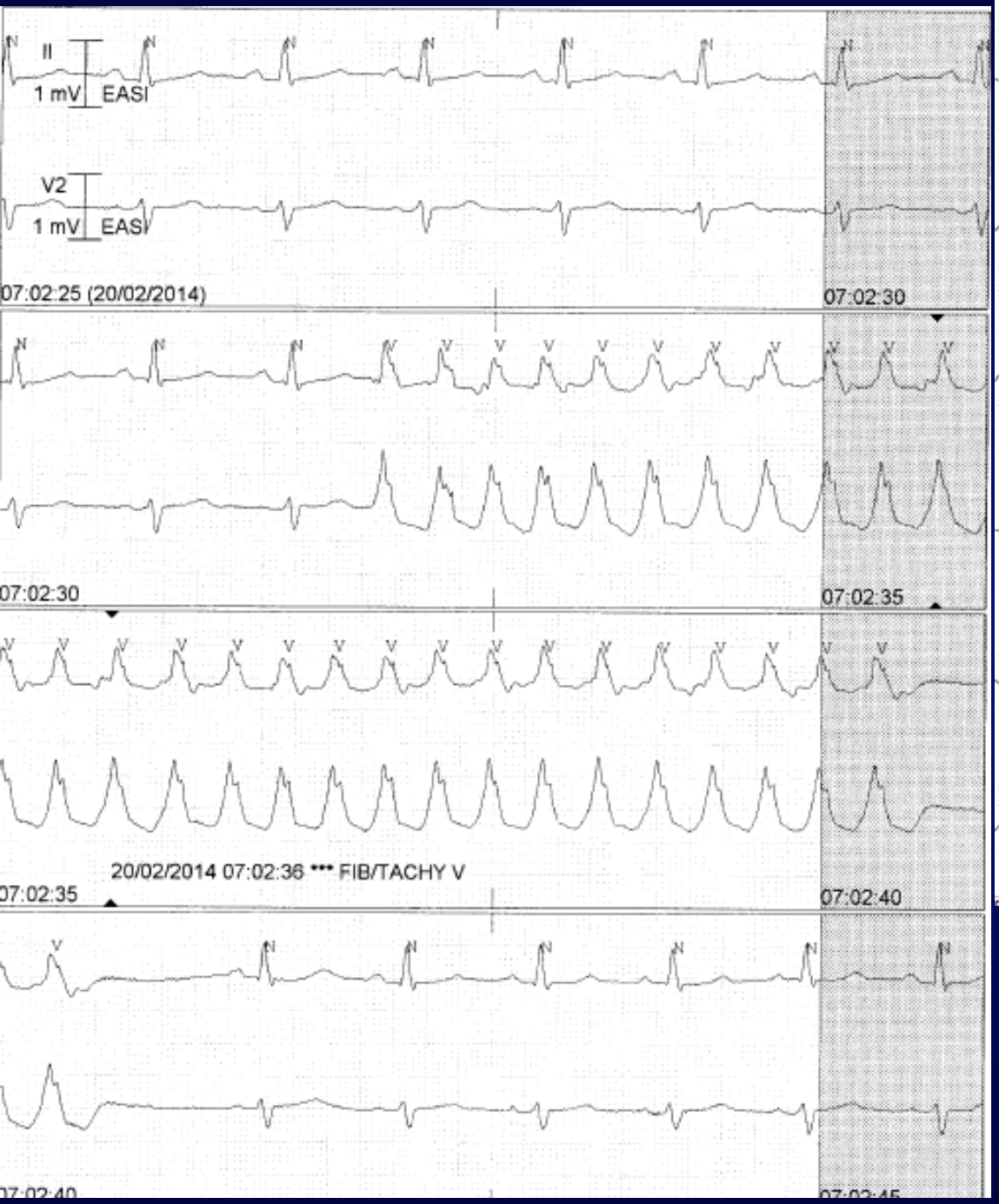


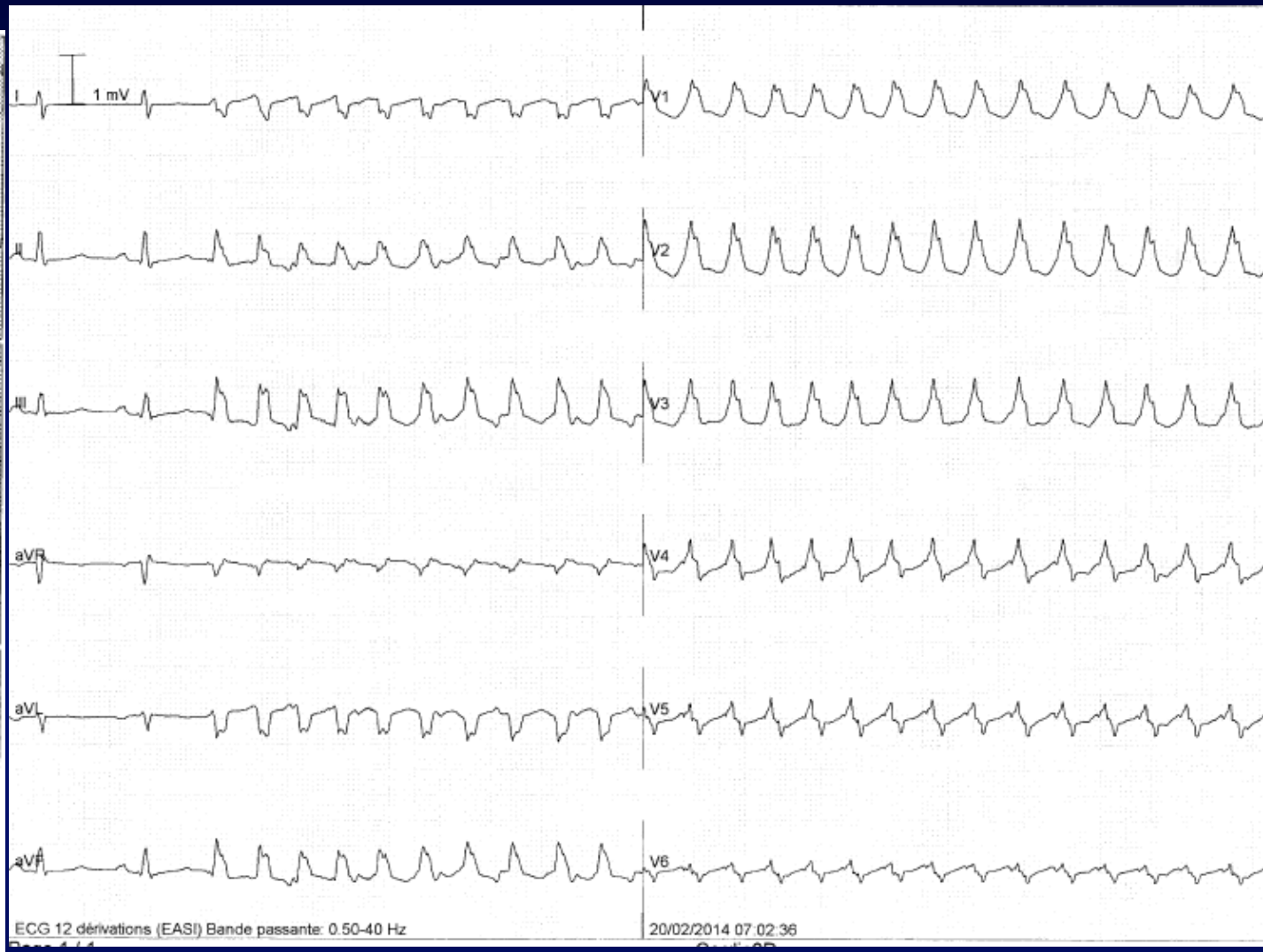


Expert consensus recommendations on non-sustained

(1) Infrequent ventricular ectopic beats, couplets, and triplets without other signs of an underlying SHD or an inherited arrhythmia syndrome should be considered as a normal variant in asymptomatic patients. IIa LOE C

(3) No treatment other than reassurance is needed for patients with neither SHD nor an inherited arrhythmogenic disorder who have asymptomatic or mildly symptomatic PVCs. I LOE C





Pronostic selon la cardiopathie sousjacente

EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias

ESV et Test d'Effort

Les ESV à l'effort: signe défavorable?

- Suspicion de pathologie cardiaque
- Si cardiopathie sousjacente
- Si modification de l'ECG à l'effort

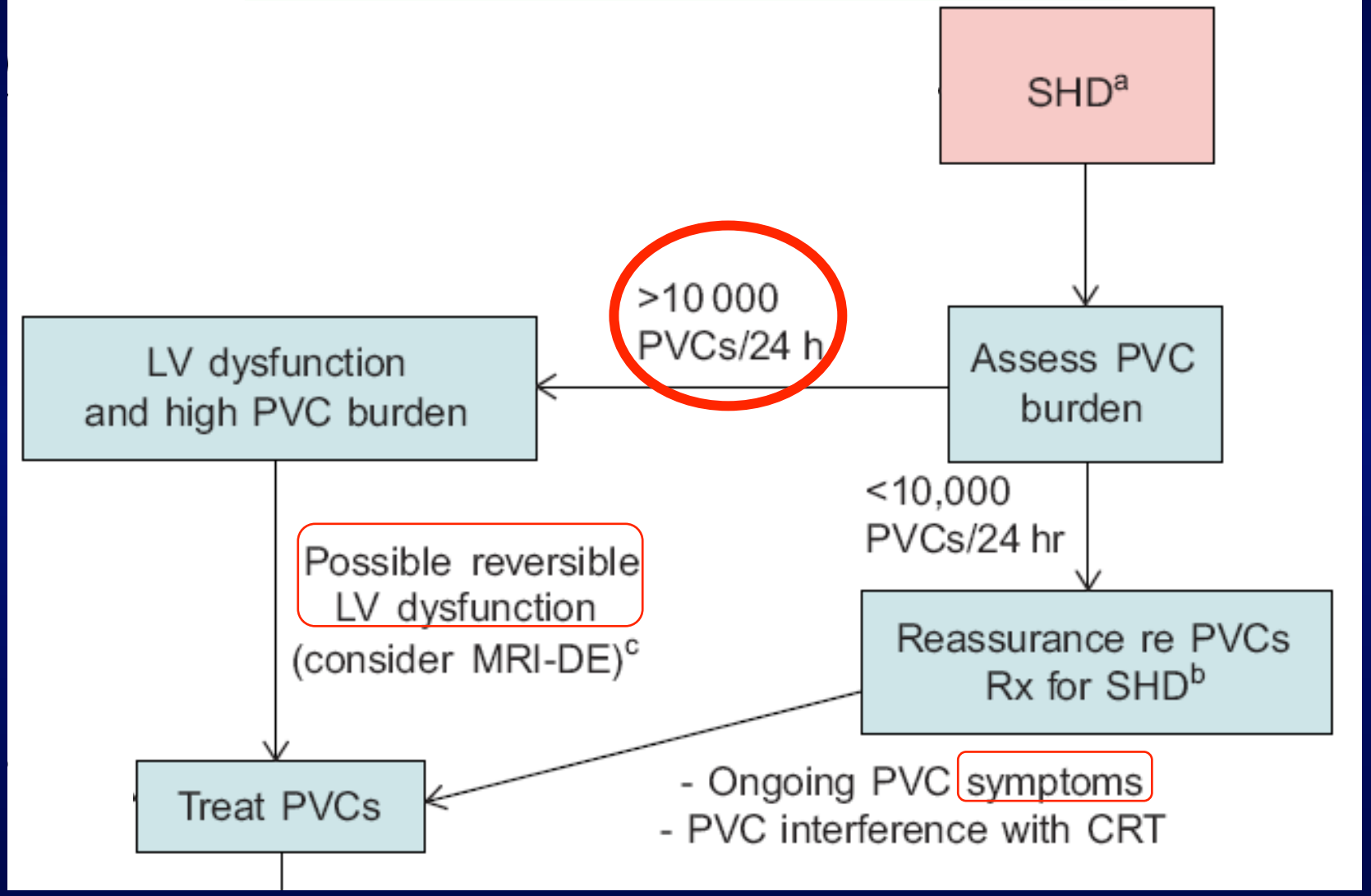
Les ESV disparaissant à l'effort: signe favorable?

- Influence de la diastole électrique

EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias

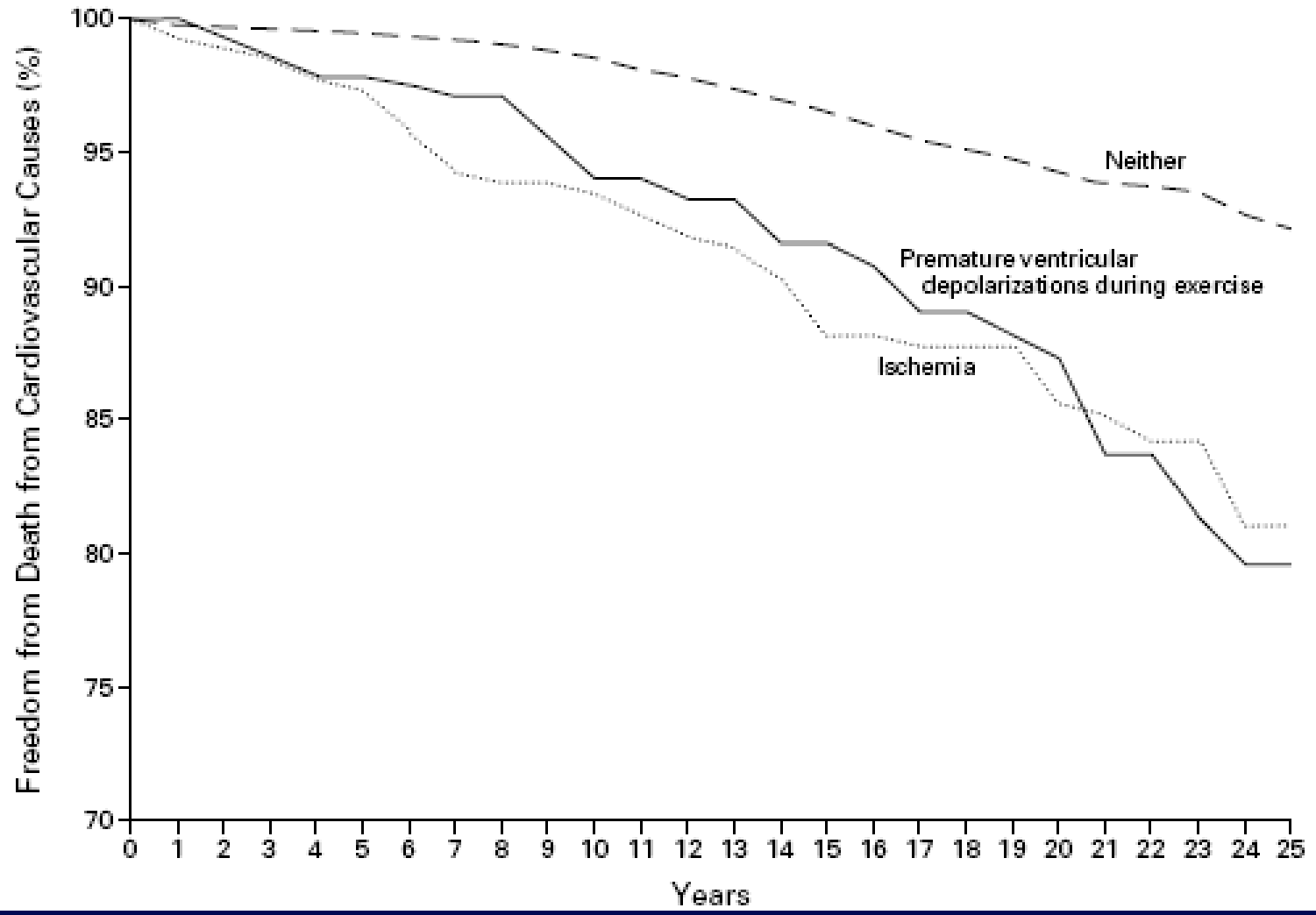
2014

Christian Torp Pedersen (EHRA Chairperson, Denmark), G. Neal Kay (HRS Chairperson, USA),



ESV sans cardiopathie démontrable

Jouven NEJ Med2000



6101 Asymptomatic subjects: **138 Frequent VPB**

Heart. 2012 Sep;98(17):1290-8. doi: 10.1136/heartjnl-2012-302005. Epub 2012 Jul 10.

BMJ Full Text

The prognostic significance of premature ventricular complexes in adults without clinically apparent heart disease: a meta-analysis and systematic review.

Lee V[1], Hemingway H, Harb R, Crake T, Lambiase P.

8 études comparant ESV vs absence d'ESV

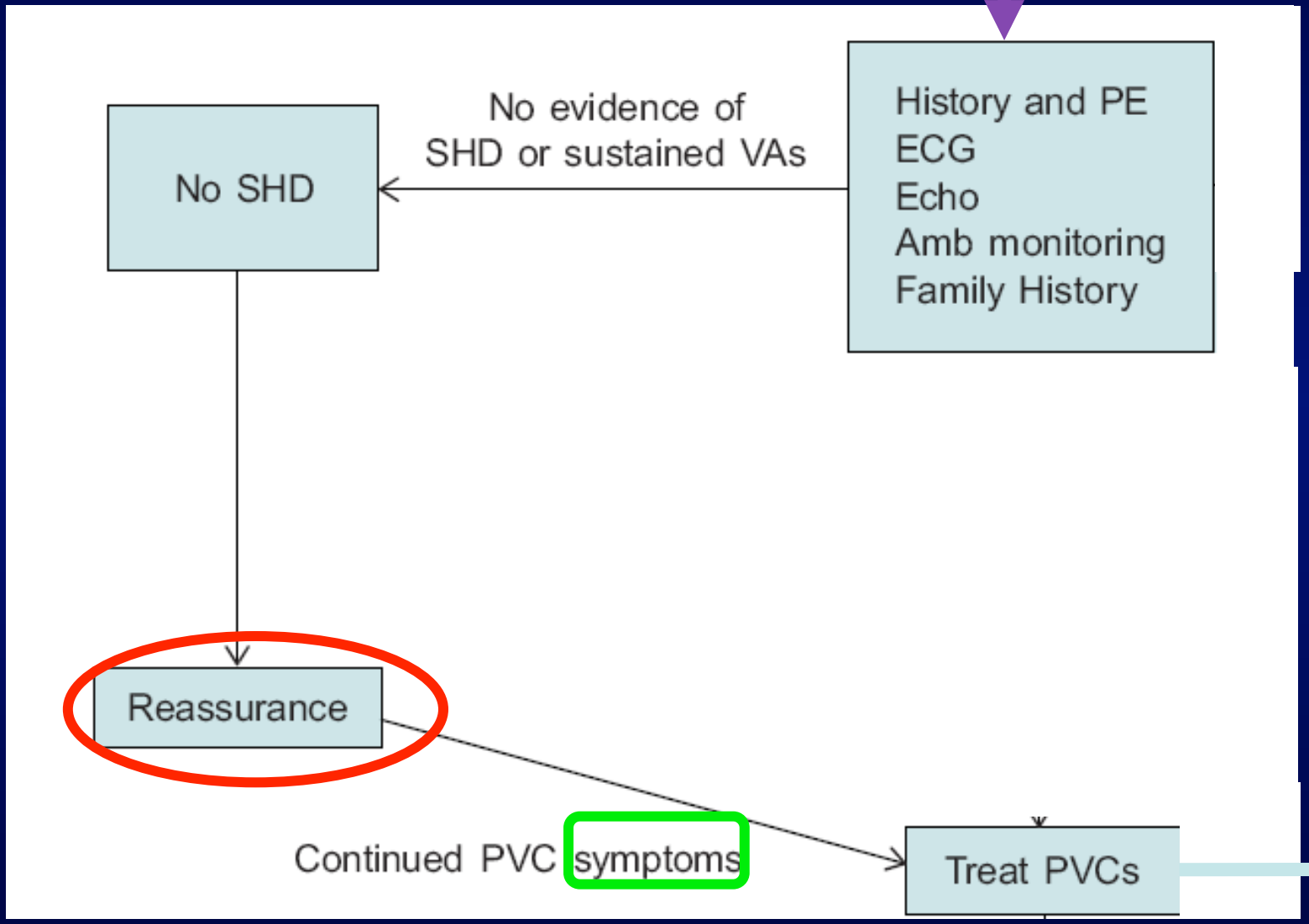
CONCLUSIONS: Most studies on PVC prognosis in 'normal hearts' did not use advanced tests to rule out structural heart disease. Among these patients, PVCs are associated with a worse cardiovascular outcome if patients are older and have higher cardiovascular risk, suggesting that the poor prognosis studies may have inadvertently included patients with occult structural heart disease, the population in which PVCs are known to confer adverse outcomes.

Eléments prédictifs d'événements: Age, Diabète, HTA

ESV sans cardiopathie

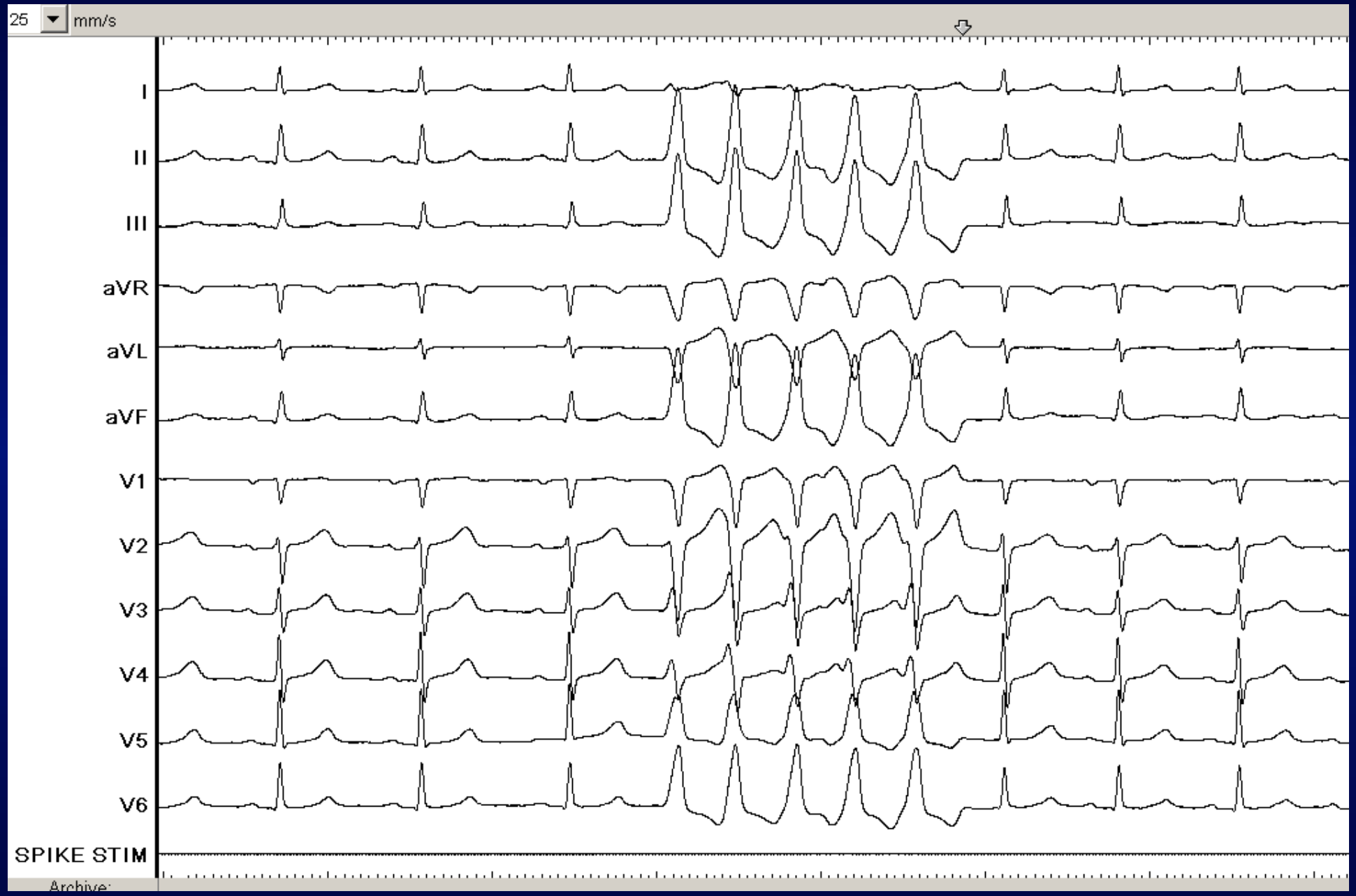
EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias 2014

Christian Torp Pedersen (EHRA Chairperson, Denmark), G. Neal Kay (HRS Chairperson, USA),



1. AA Cl Ic
2. ABL

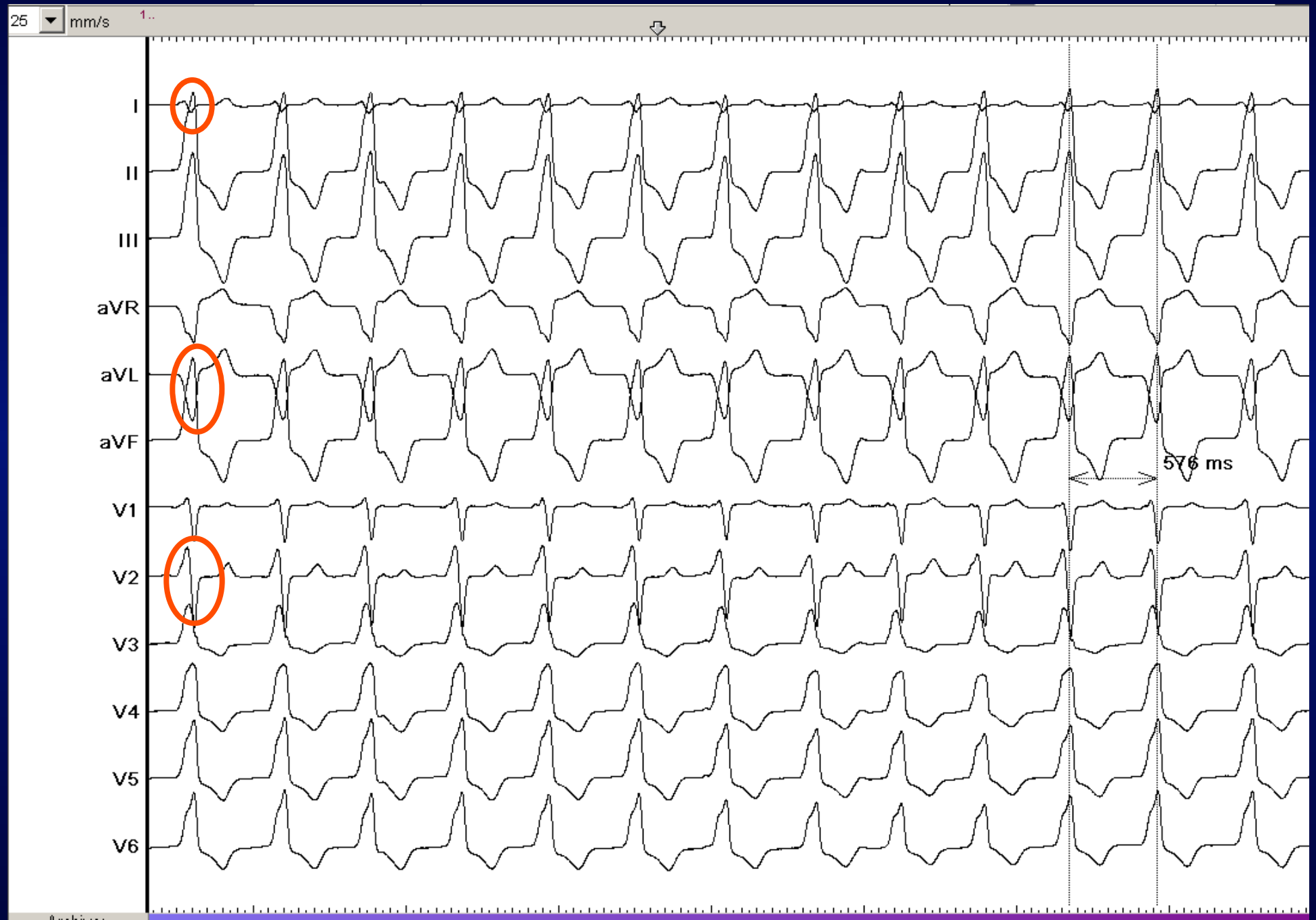
Les Arythmies idiopathiques

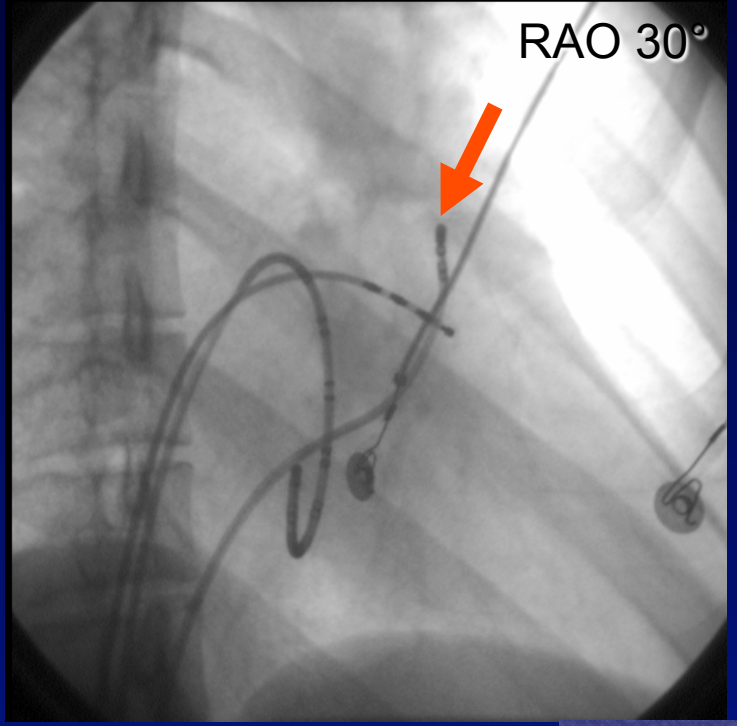


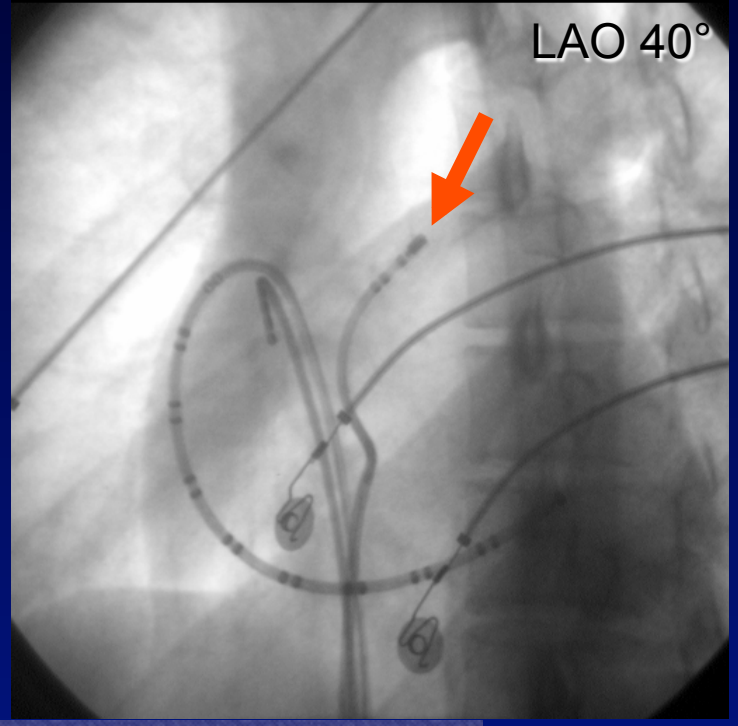
P G

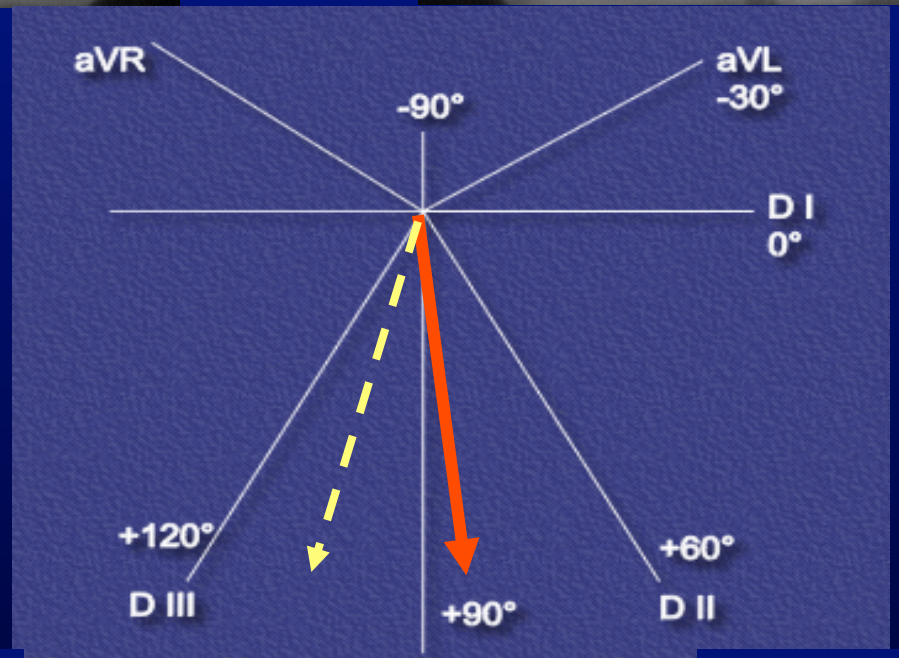
TV 12D

RVOT









QRS neg in I

QRS pos in I

IDIOPATHIC VENTRICULAR ARRHYTHMIAS

A/ LOCATION:

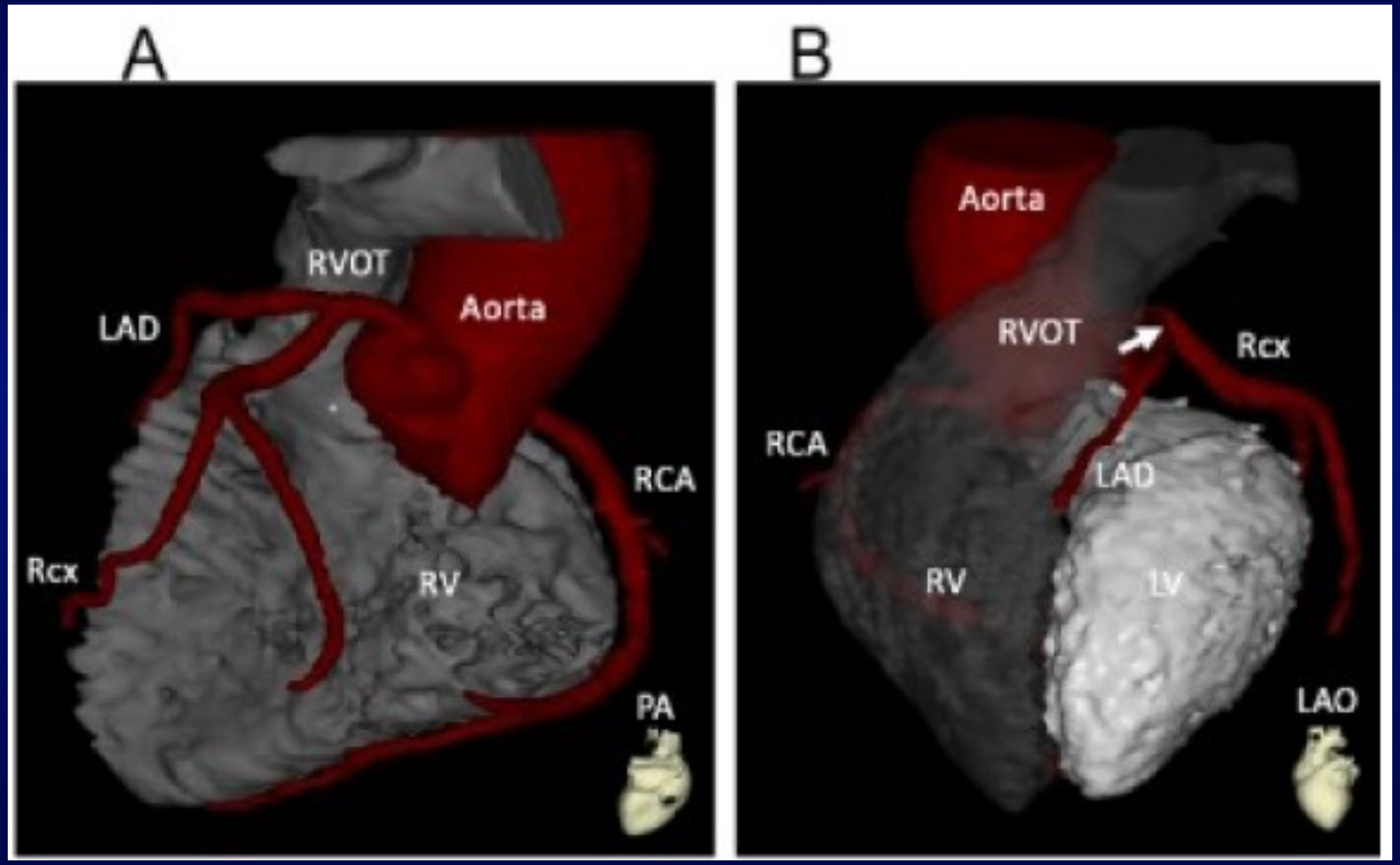
- RVOT
- PULMONARY ARTERY
- LVOT
- AORTIC CUSPS
- MITRAL ANNULUS
- FASCICULAR LV
- EPICARDIAL

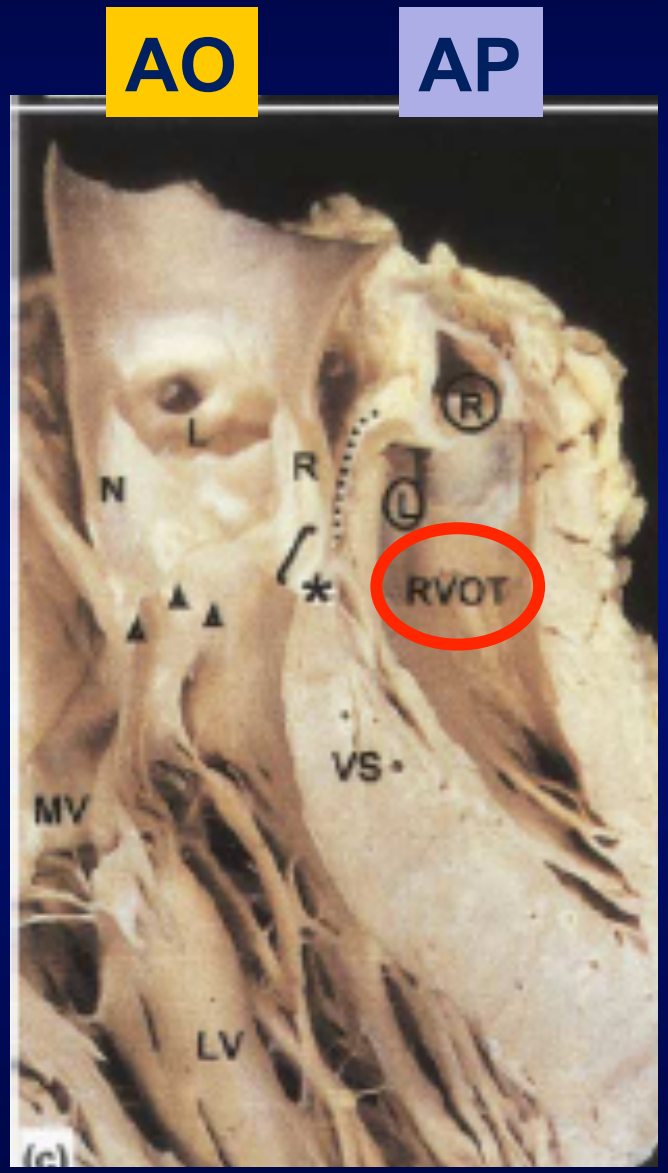
B/ TYPES:

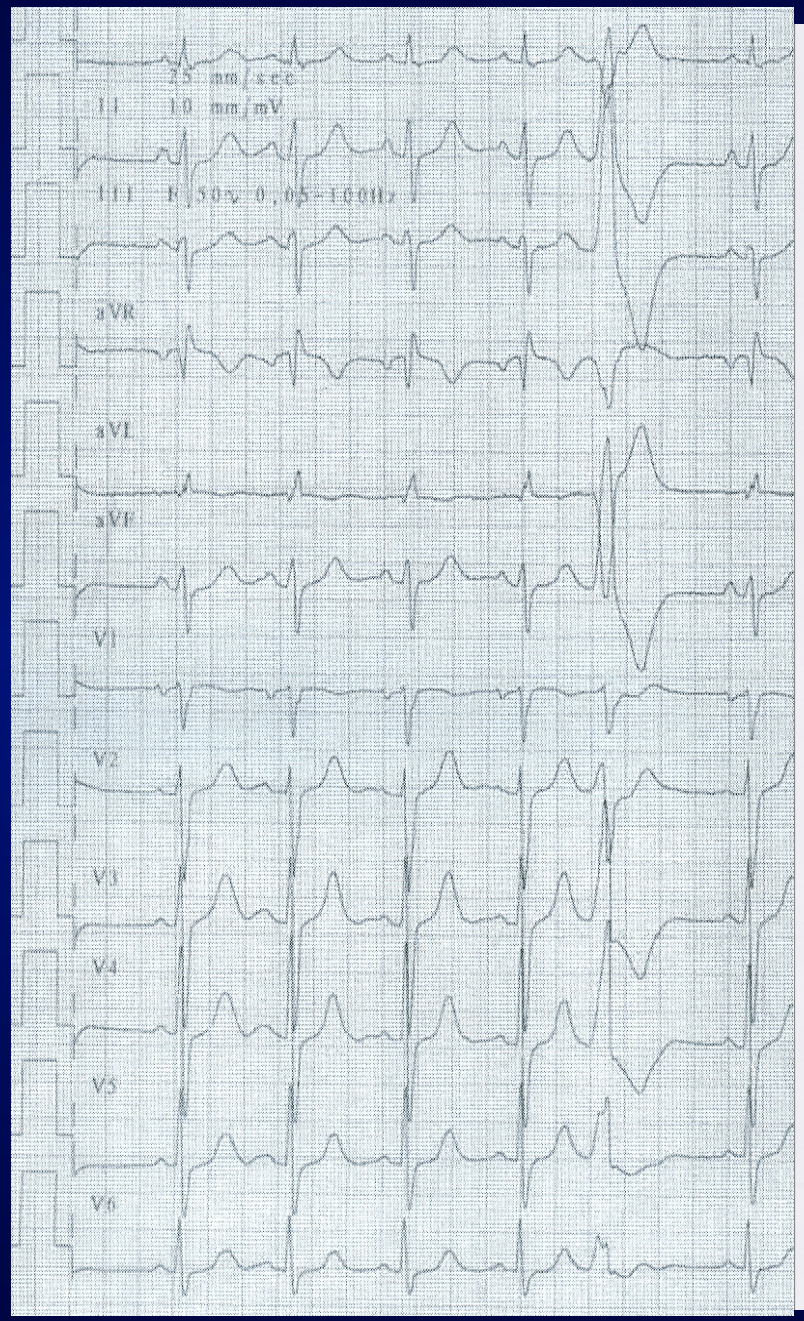
1. BENING MONOMORHIC: long coupling interval
   frequent VPB
2. MALIGNANT POLYMORPHIC: short or intermediate coupling
   rare VPB

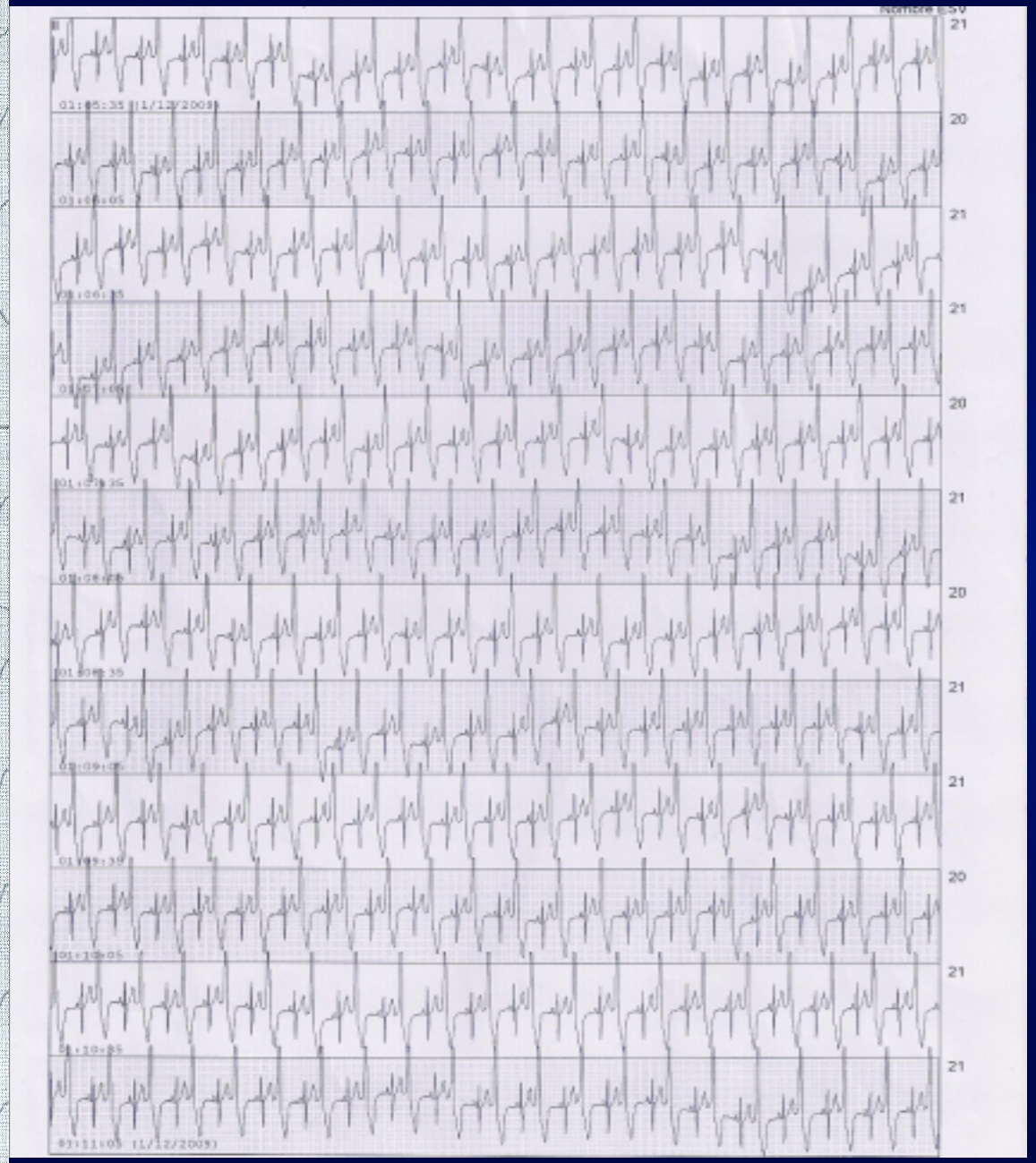
Viskin, Antzelevitch JACC 2005

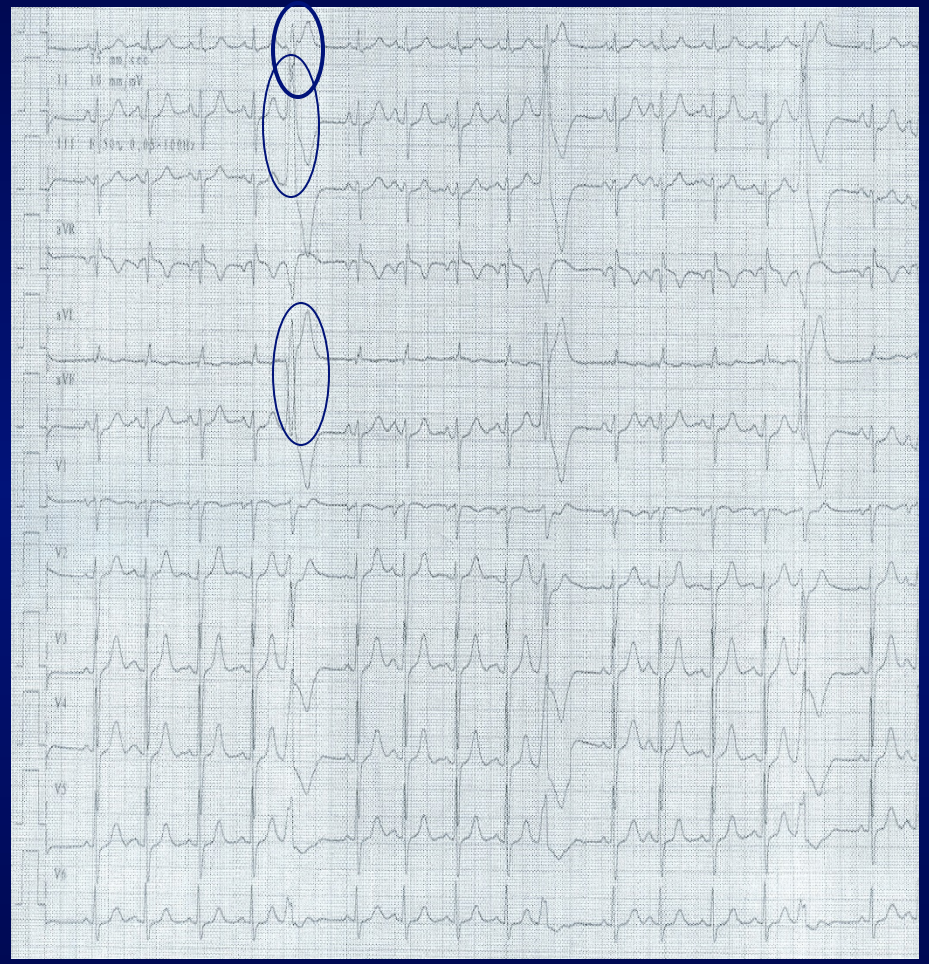
RVOT and LVOT

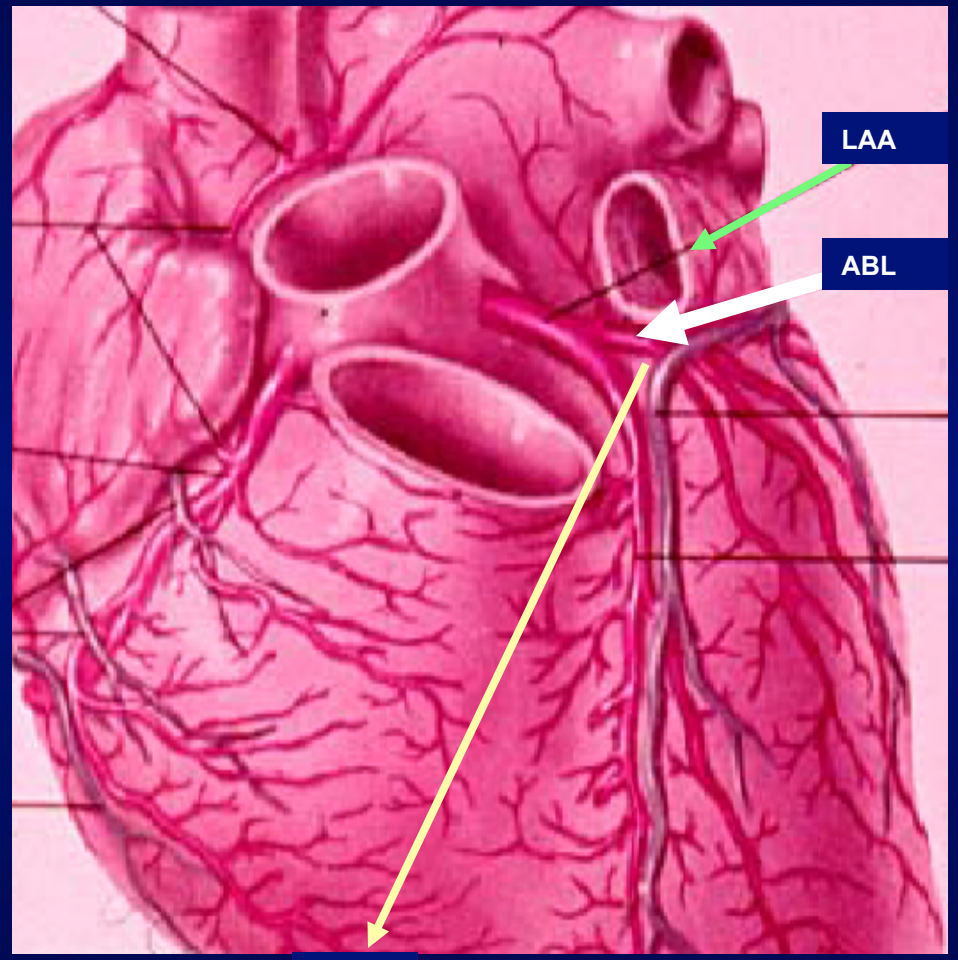


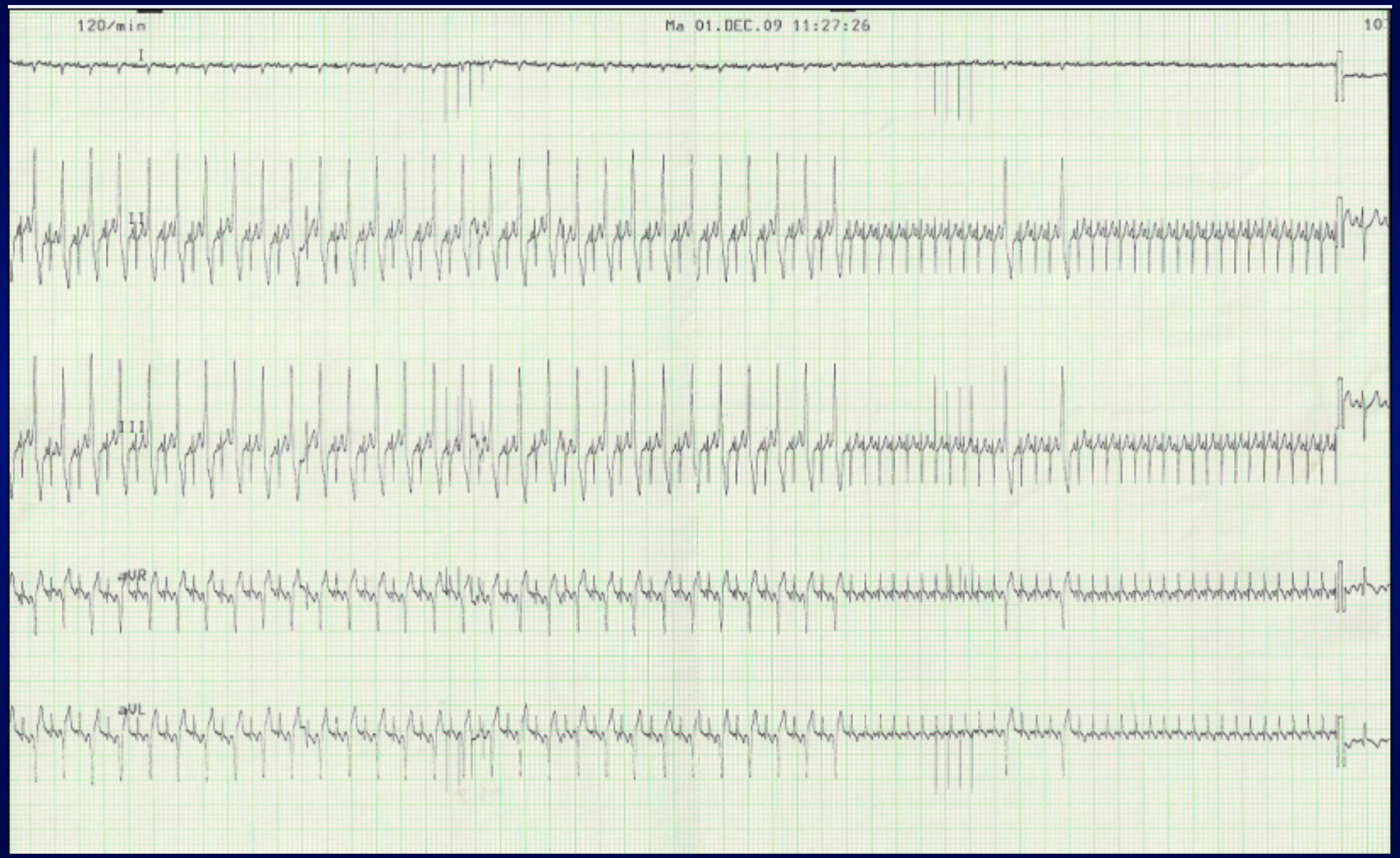



25 mm/sec
I 10 mm/mV
III R 50% 0,05-100Hz
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

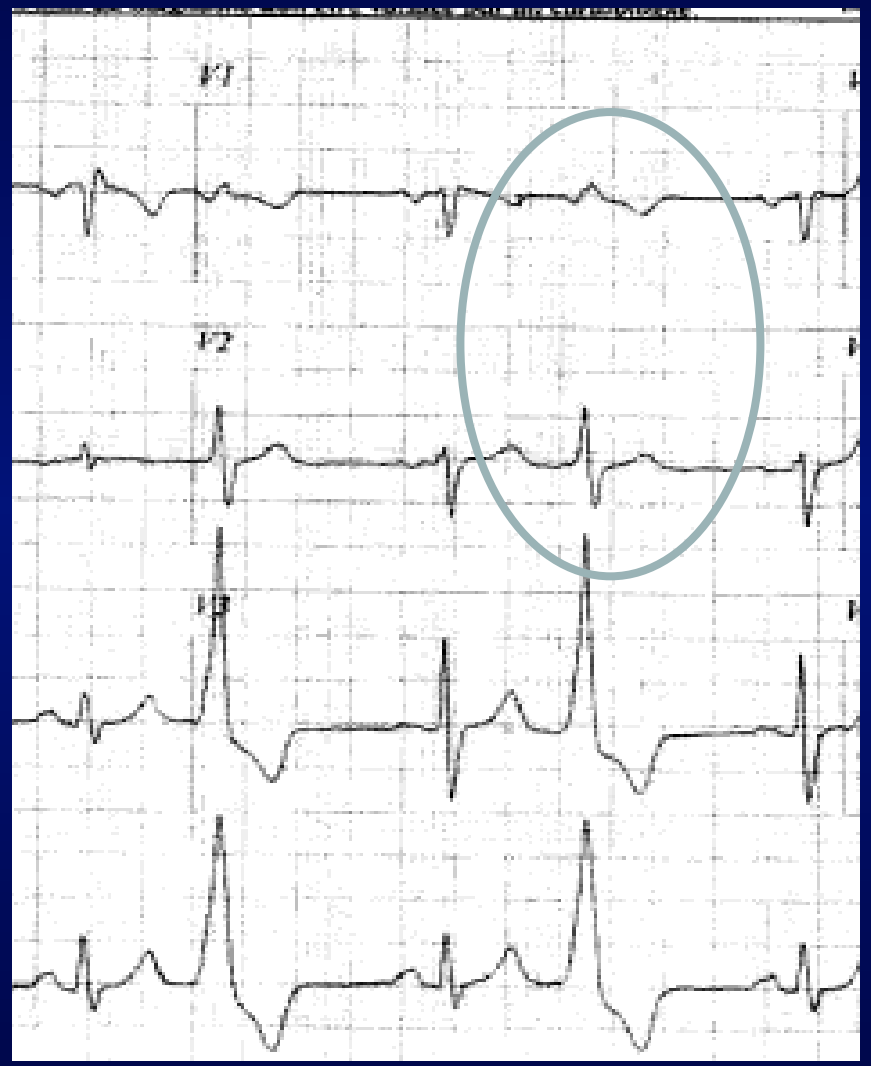


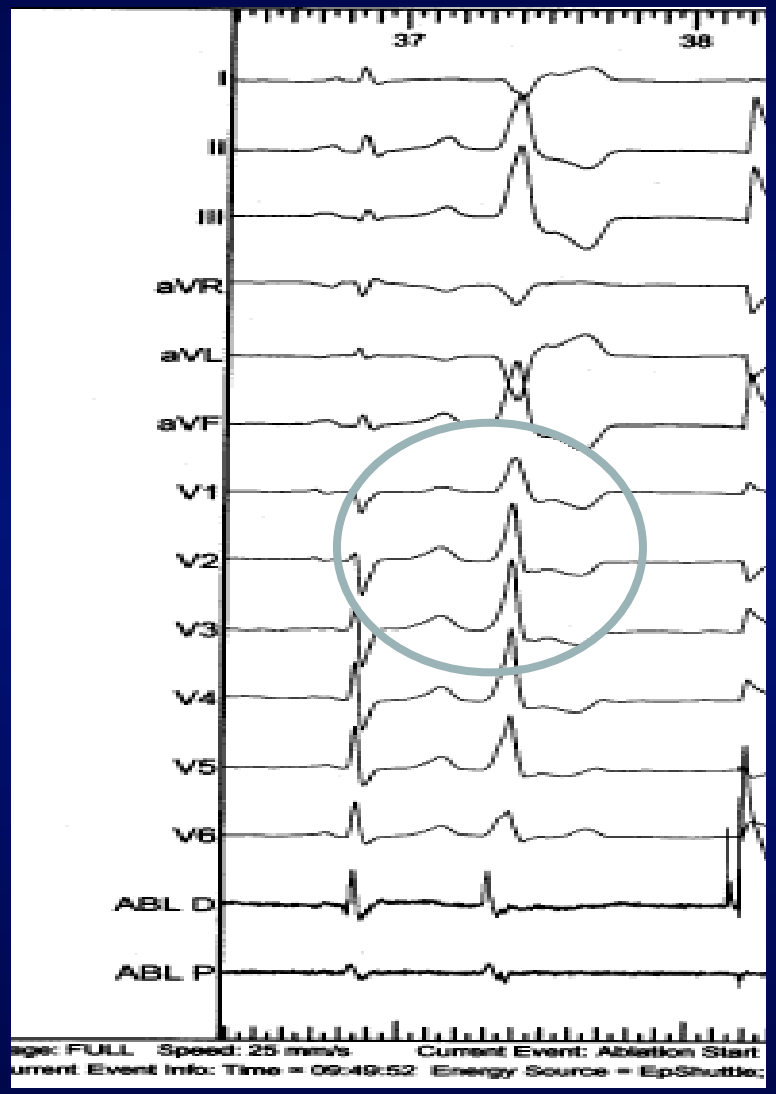



LAA
ABL
QRS
axis

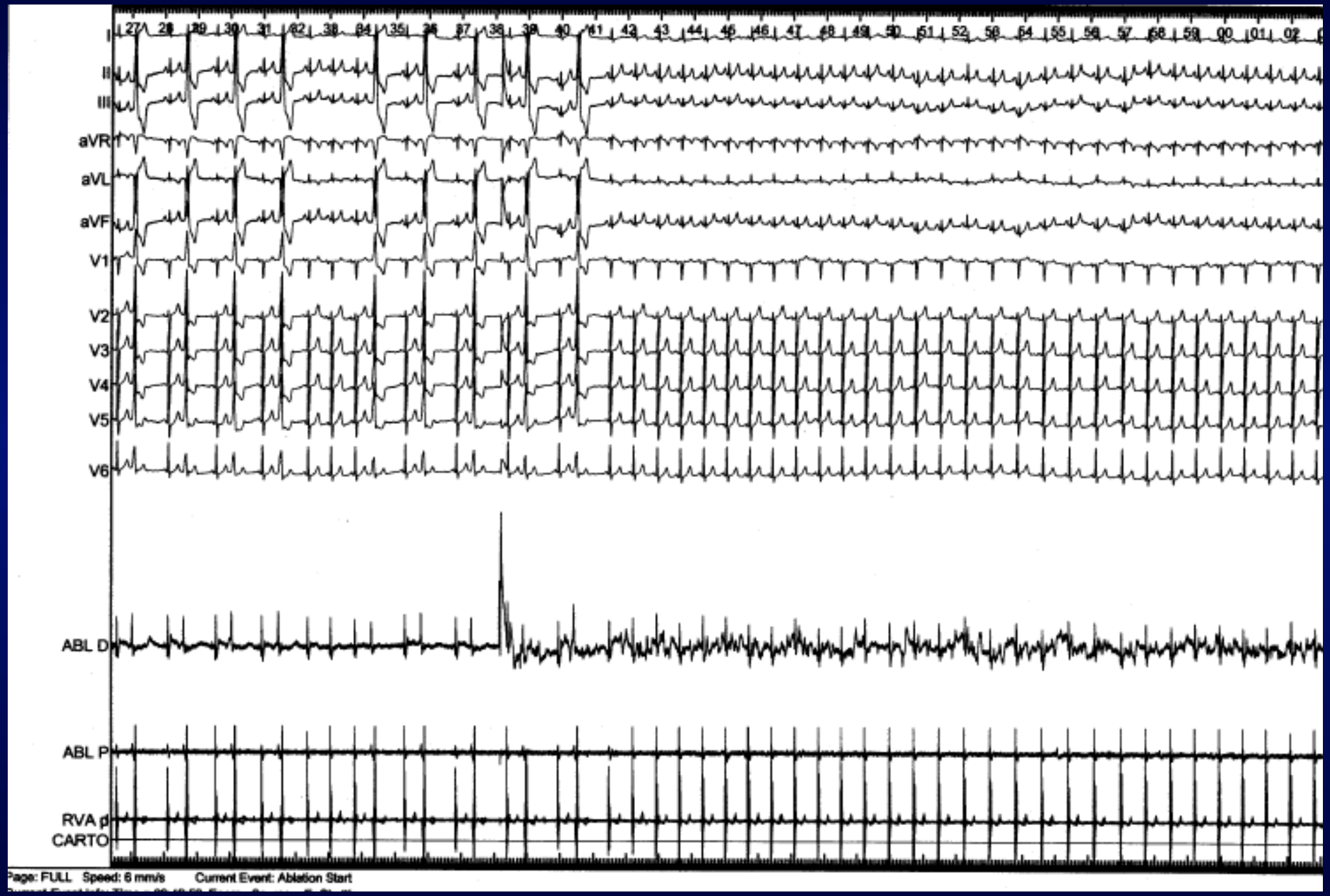

120/min
Ma 01.DEC.09 11:27:26
I
II
III
aVR
aVL

Variabilité de la morphologie des ESV





3-LV (17, 0) Resp
Correlations:
-106 ms
LAT
-33 ms
-100 gs
-106
-90
FTI
300 gs
FOYER DES ESV
CUSPIDE AO GAUCHE
AORTE VUE PAR LE DESSOUS
0.44
CL
LAT
Bi
Imp
714
N/A
N/A
N/A
AP
PA
LAO
RAO
LL
RL
INF
SUP


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
ABL D
ABL P
RVA p
CARTO
Page: FULL Speed: 6 mm/s
Current Event: Ablation Start

CONTEMPORARY REVIEW

Arrhythmias originating from the right ventricular outflow tract: How to distinguish "malignant" from "benign"?

Wataru Shimizu, MD, PhD

Shimuzu Catheter Ablation for Ventricular Fibrillation

Table 1 Comparison of ECG characteristics between malignant RVOT VT, benign RVOT VT, and idiopathic VF

	Malignant RVOT VT	Benign RVOT VT	Idiopathic VF	*P*
CI, ms:				
Haissaguerre et al.	355 ± 30	—	280 ± 26	.01
Viskin et al.	340 ± 30	427 ± 76	300 ± 40	<.001
Noda et al.	409 ± 62	428 ± 65	—	.27
QRS duration, ms:				
Haissaguerre et al.	145 ± 12	—	126 ± 18	.04
Noda et al.	148 ± 8	142 ± 12	—	.03
Cycle length of VT, ms:				
Noda et al.	245 ± 28	328 ± 65	—	<.0001

Shimizu Heart Rhythm 2009

IDIOPATHIC PVC

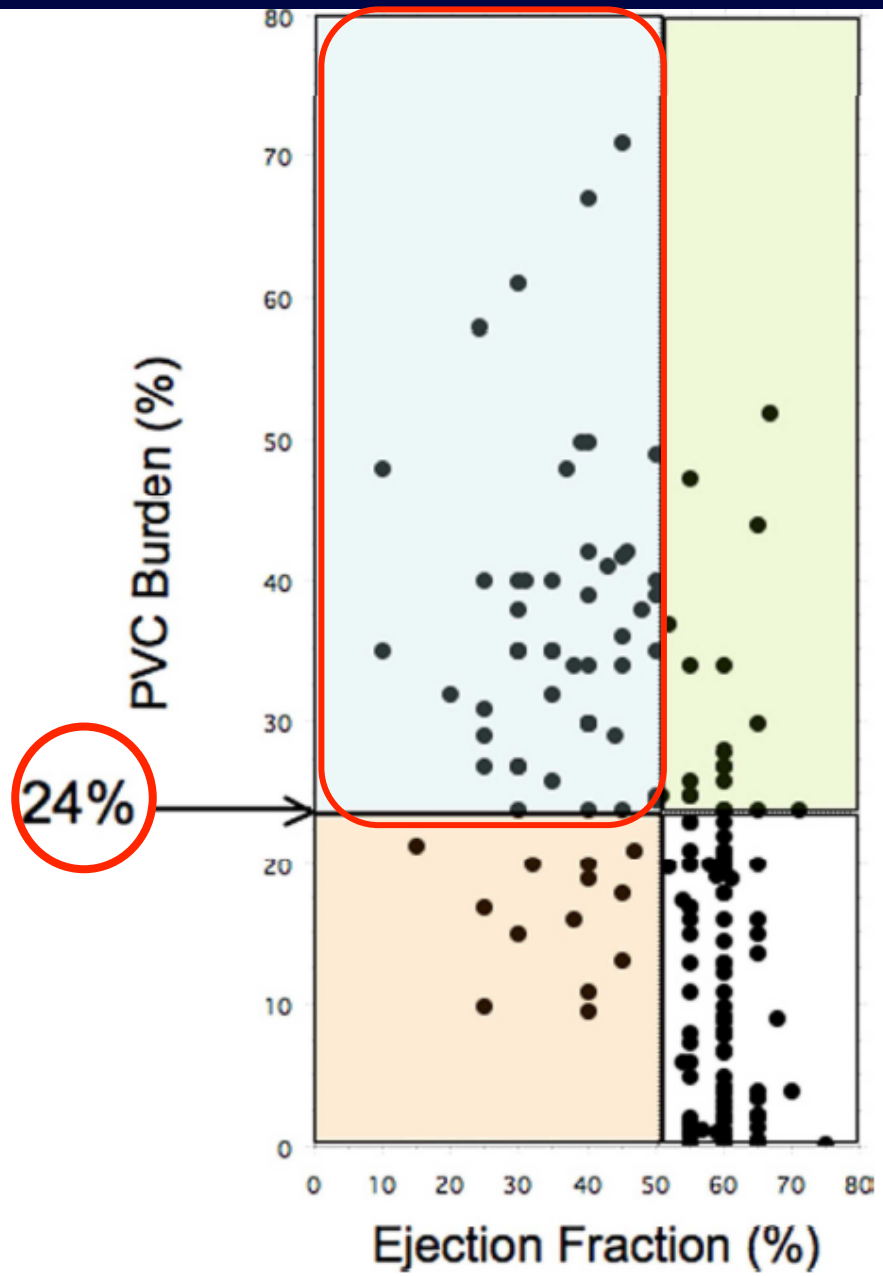


Figure 1 Scattergram indicating the relationship between PVC burden and ejection fraction. PVC = premature ventricular complex.

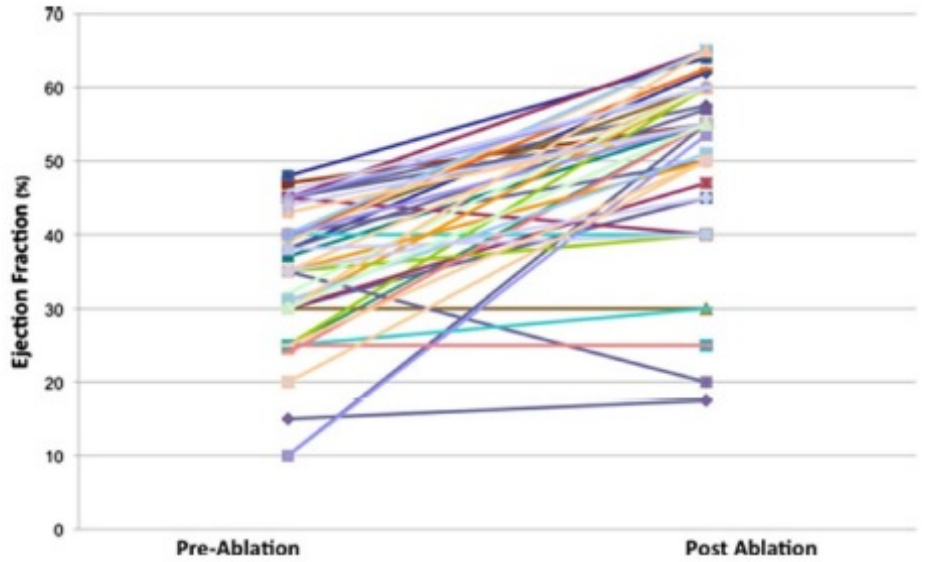


Figure 3 Plot indicating the change of ejection fraction in patients with cardiomyopathy after ablation.

174 PTS

Baman TS et al.: Heart Rhythm, Vol 7 July 2010: 867

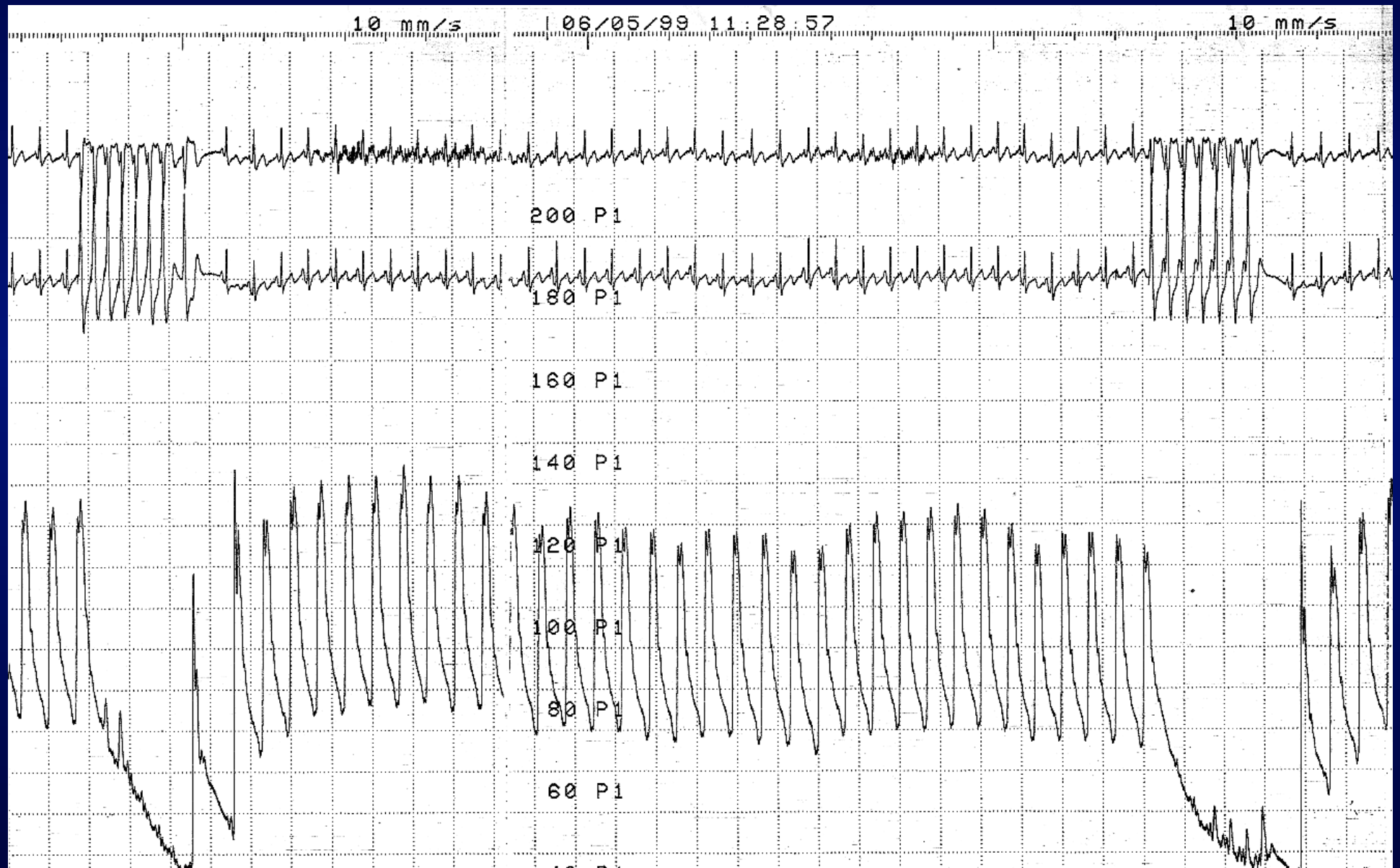
LES TACHYCARDIES VENTRICULAIRES

TV ≥ 3 complexes ventriculaires consécutifs - Soutenue: ≥ 30 s

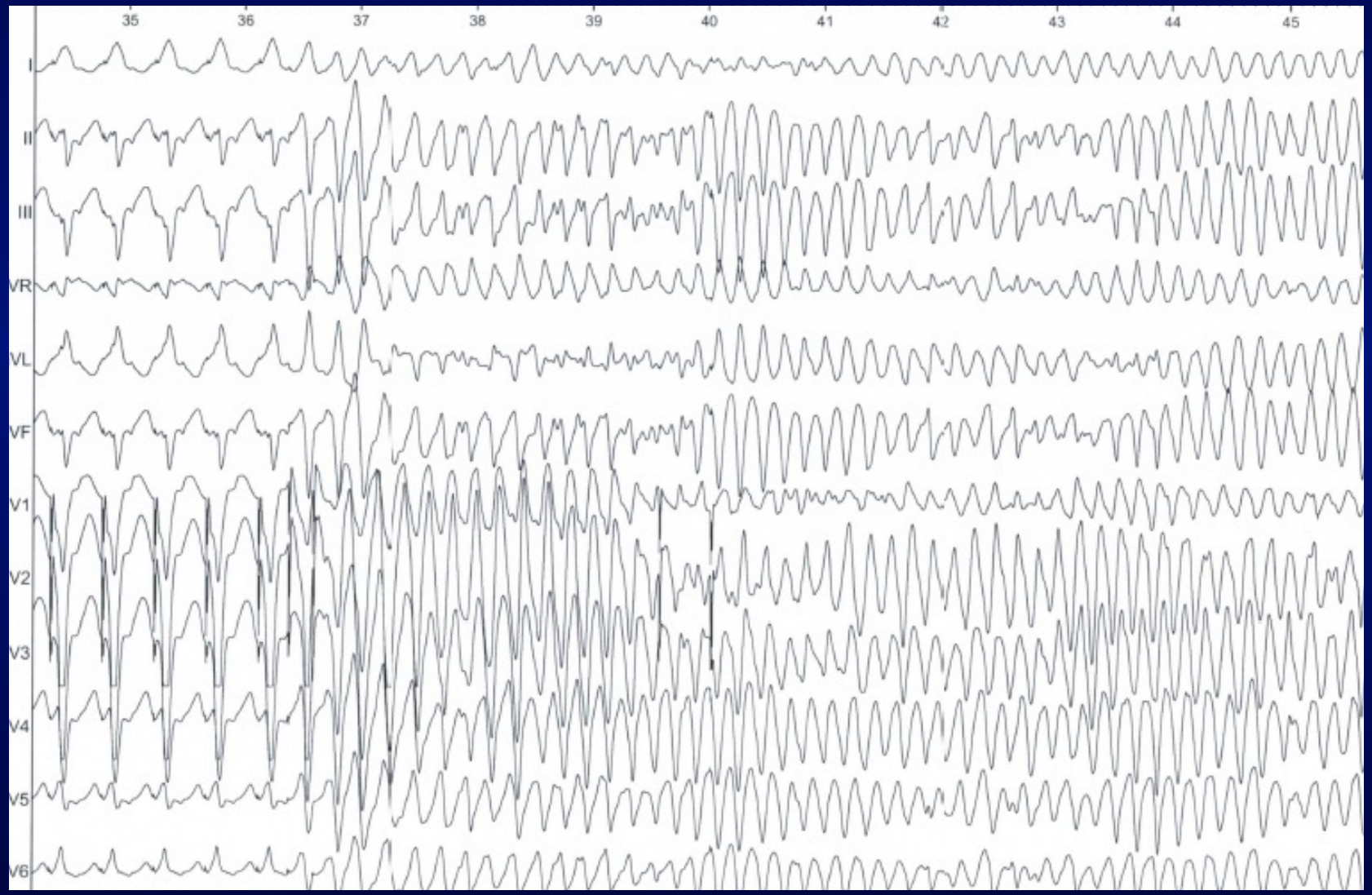
- Non soutenue

1. TV Avec aspect de retard D (BBD): R dominant en V1
2. TV avec aspect de retard G (BBG): négativité dominante en V1
3. TV de morphologie intermédiaire

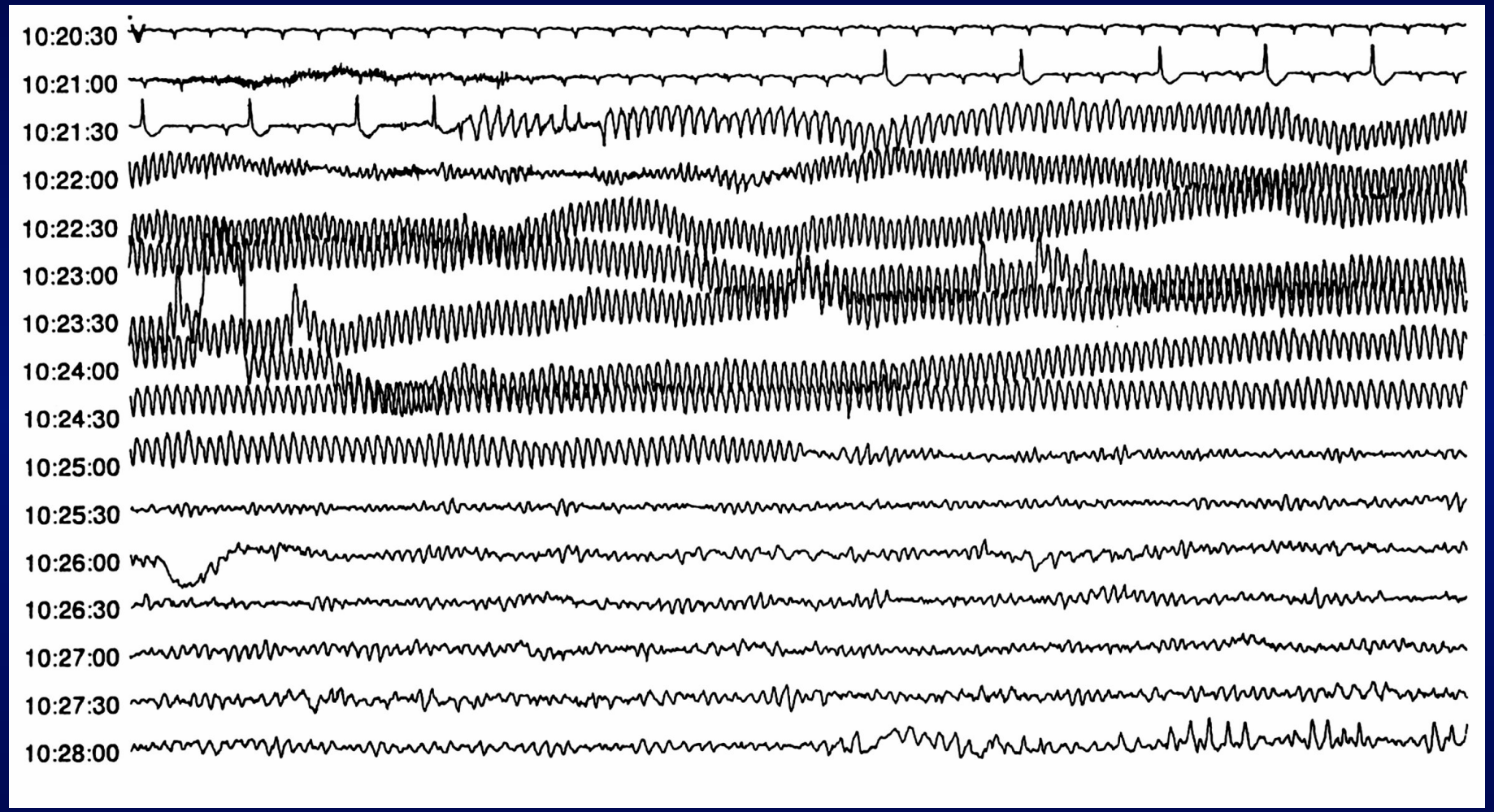
Effet hémodynamique des arythmies ventricualires



TV→ FV



Torsades→ FV


10:20:30
10:21:00
10:21:30
10:22:00
10:22:30
10:23:00
10:23:30
10:24:00
10:24:30
10:25:00
10:25:30
10:26:00
10:26:30
10:27:00
10:27:30
10:28:00

PULSELESS ARREST

Check rhythm
Shockable rhythm?

8 Shockable

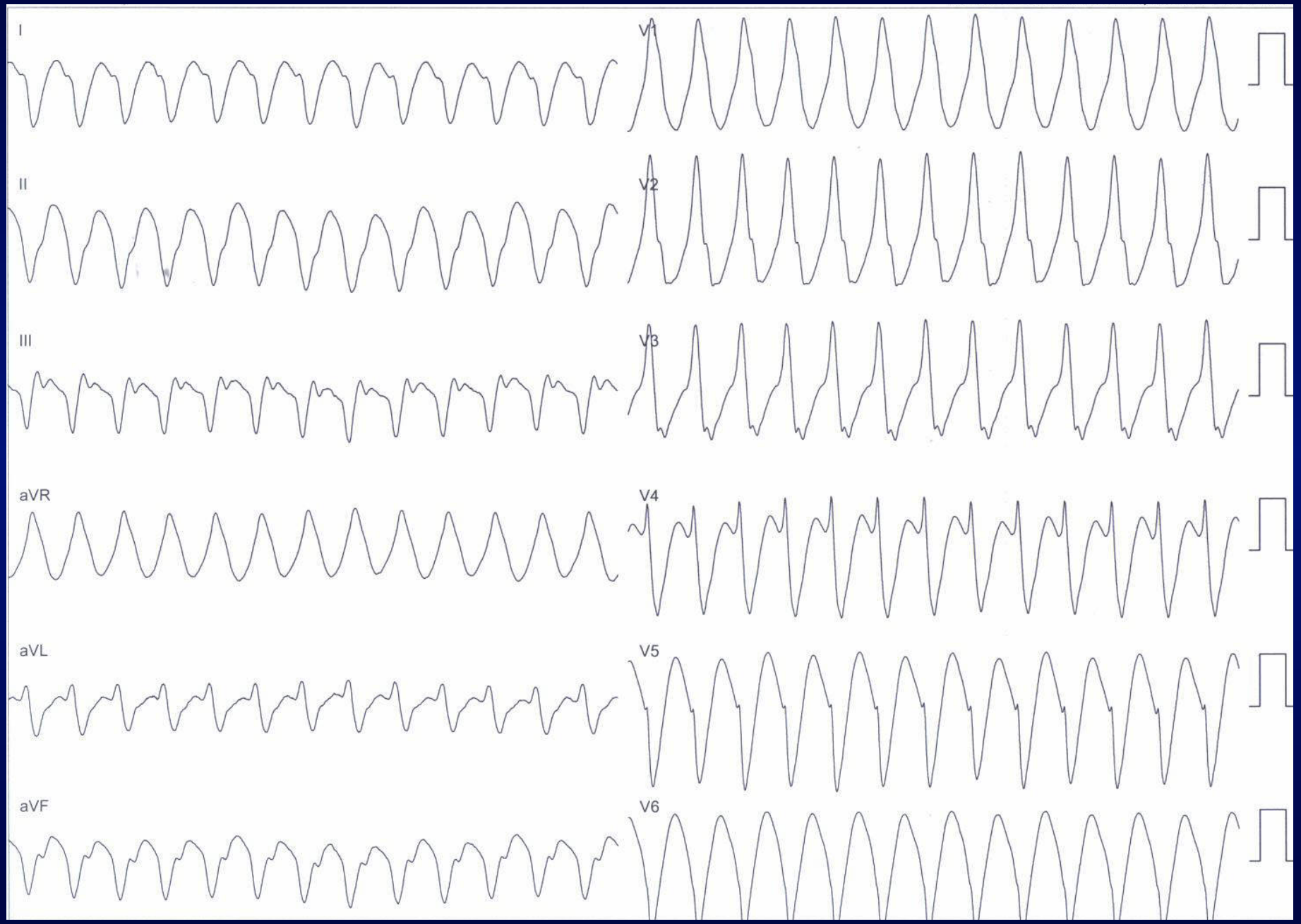
Give 1 shock

- Manual biphasic: device-specific (same or higher dose as first shock)
  Note: if unknown, use 200 J
- AED: device-specific
- Monophasic: 360 J

Resume CPR immediately

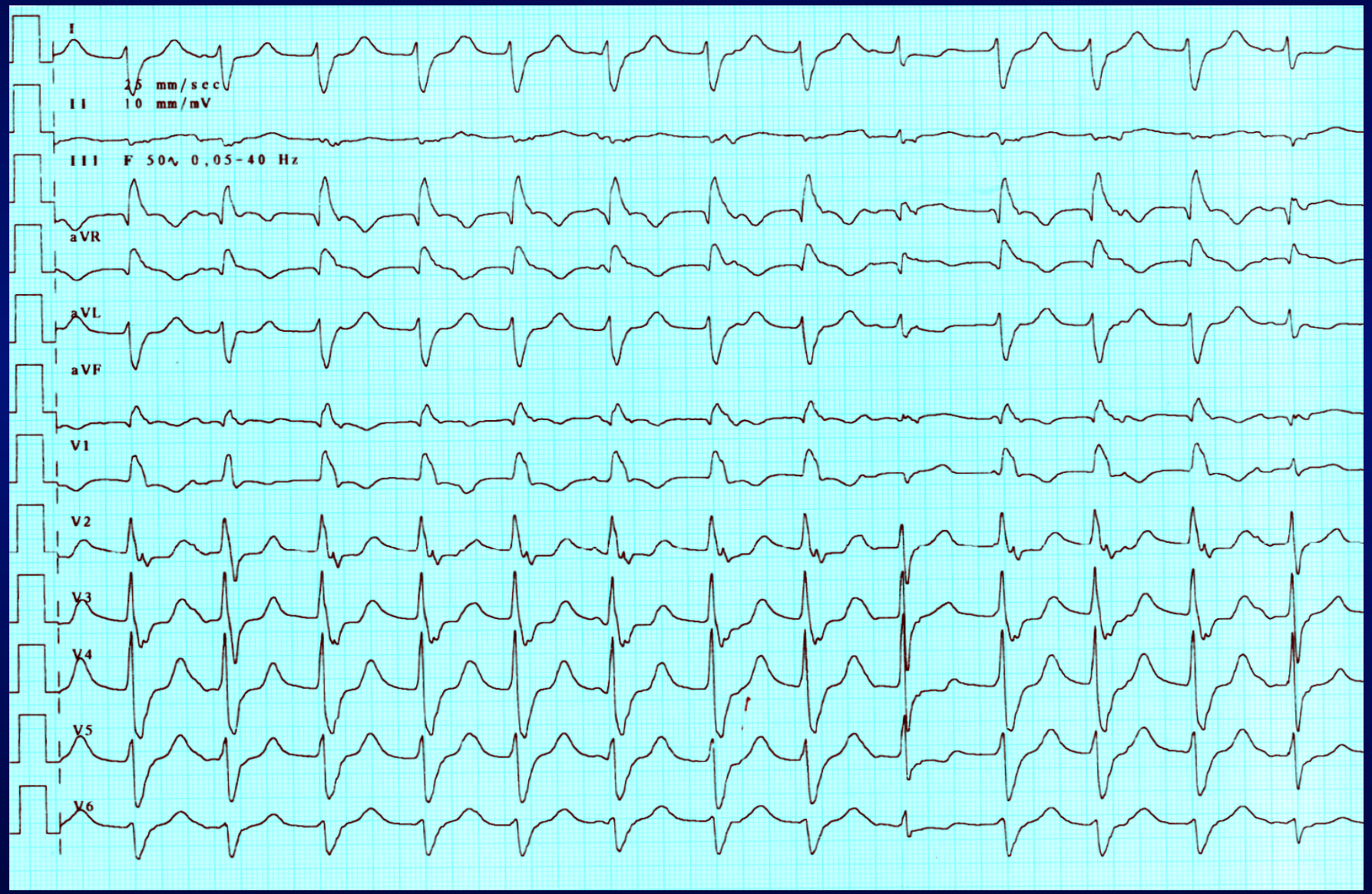
Consider antiarrhythmics: amiodarone (300 mg IV/IO once, then consider additional 150 mg IV/IO once) or **lidocaine** (1 to 1.5 mg/kg first dose then 0.5 to 0.75 mg/kg IV/IO, maximum 3 doses or 3 mg/kg)

Consider **magnesium**, loading dose 1 to 2 g IV/IO for torsades de pointes



QUE FAIRE ?

TV bien supportée



QUE FAIRE ?

G R

Cardioversion or defibrillation and acute treatment of sustained ventricular arrhythmias

Recommendations	Class[a]	Level[b]	Ref.[c]
Direct current cardioversion is recommended for patients presenting with sustained VT and haemodynamic instability.	I	C	180

Si bien tolérée: **CEE !**

Procainamide
Flecainide
Ajmaline ?

Si Insuff card/ ischémie:

amiodarone
lidocaine?

BRUGADA

BRUGADA et GENETIQUE

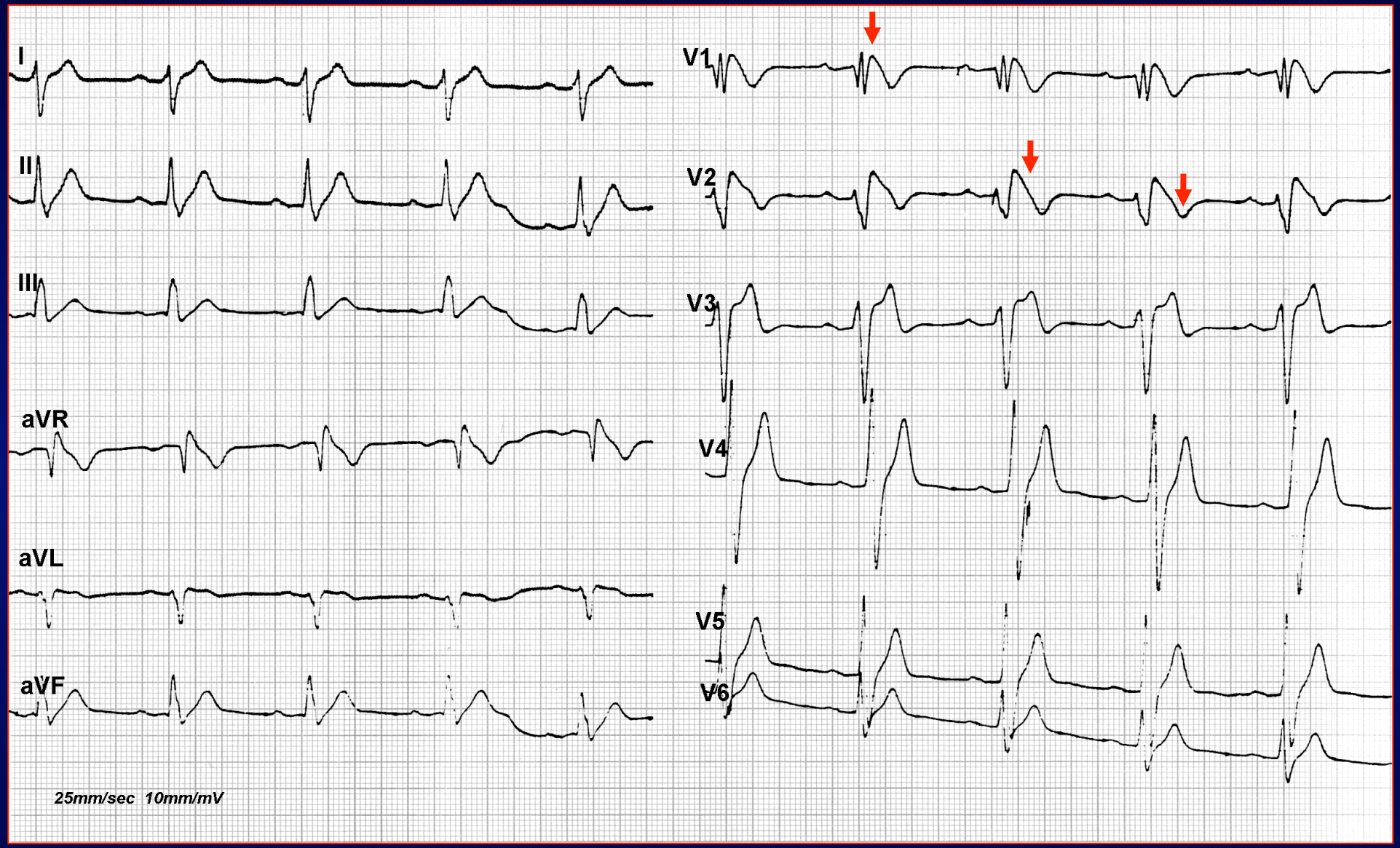
Table 4. Genes Involved in Inherited Arrhythmias

Phenotype	Variant	Gene	Protein	Functional Defect
Brugada syndrome	BrS1	*SCN5A*	Cardiac sodium channel α subunit (Nav1.5)	Loss of function, reduced Na^+ current
	BrS2	*GPD1-L*	Glycerol-6-phosphate-dehydrogenase	Loss of function, reduced Na^+ current
	BrS3	*CACNA1c*	L-type calcium channel α subunit (Cav1.2)	Loss of function, reduced Ca^{2+} current
	BrS4	*CACNB2*	L-type calcium channel β_2 subunit	Loss of function, reduced Ca^{2+} current
	BrS5	*SCN1B*	Cardiac sodium channel β_1 subunit	Loss of function, reduced Na^+ current
	BrS6	*KCNE3*	Transient outward current β subunit	Gain of function, increased K^+ I_{to} current
	BrS7	*SCN3B*	Cardiac sodium channel β_3 subunit	Loss of function, reduced Na^+ current
	BrS8	*MOG1*	Nucleocytoplasmic and microtubule transport	Loss of function, reduced Na^+ current
	BrS9	*KCNE5*	Transient outward current β subunit	Gain of function, increased K^+ I_{to} current
	BrS10	*KCND3*	I_{to} potassium channel (Kv4.3)	Gain of function, increased K^+ I_{to} current

LE BRUGADA

NE PAS CONFONDRE

- **ECG de type BRUGADA (Brugada Pattern)**
- **SYNDROME de BRUGADA**
- **PHENOCOPIES**



1/ 1000 subjects

Wilde AA et al. Eur Heart J 2002; 23:1648-54

Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Silvia G. Priori, (HRS Chairperson)[1], Arthur A. Wilde, (EHRA Chairperson)[2],

BRUGADA : DIAGNOSTIC

Expert Consensus Recommendations on ***BrS Diagnosis***

1. BrS ***is diagnosed*** in patients with ST-segment elevation with **type I** morphology ≥ 2 mm in ≥ 1 lead among the right precordial leads V_1,V_2 positioned in the 2nd, 3rd, or 4th intercostal space occurring either spontaneously *or* after provocative drug test with intravenous administration of Class I antiarrhythmic drugs.
2. BrS ***is diagnosed*** in patients with type 2 or type 3 ST- segment elevation in ≥ 1 lead among the right precordial leads V_1,V_2 positioned in the 2nd, 3rd, or 4th intercostal space when a provocative drug test with intravenous administration of Class I antiarrhythmic drugs induces a **type I** ECG morphology.

Sudden Cardiac Death and Genetic Ion Channelopathies

Long QT, Brugada, Short QT, Catecholaminergic Polymorphic Ventricular Tachycardia, and Idiopathic Ventricular Fibrillation

Carlo Napolitano, MD, PhD; Raffaella Bloise, MD; Nicola Monteforte, MD; Silvia G. Priori, MD, PhD

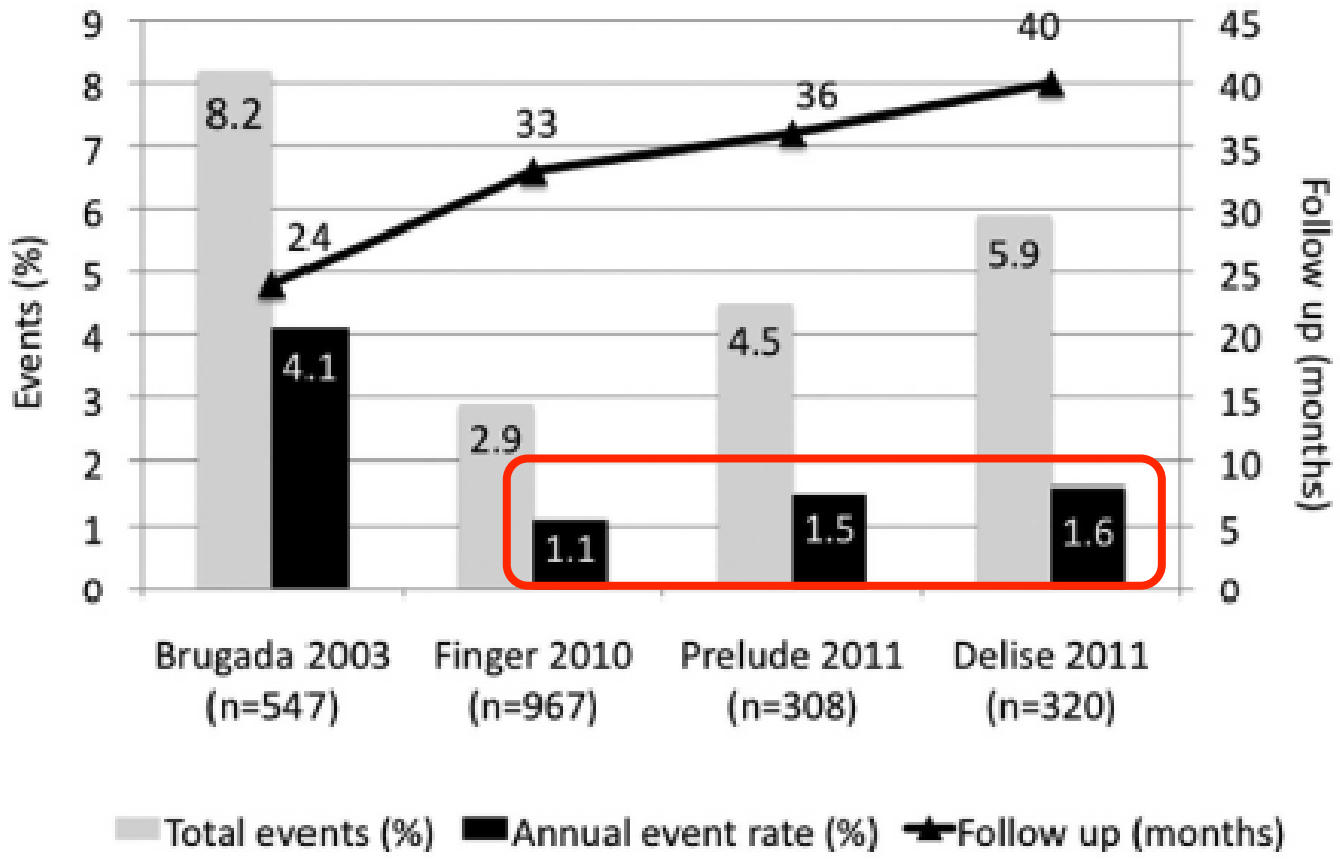
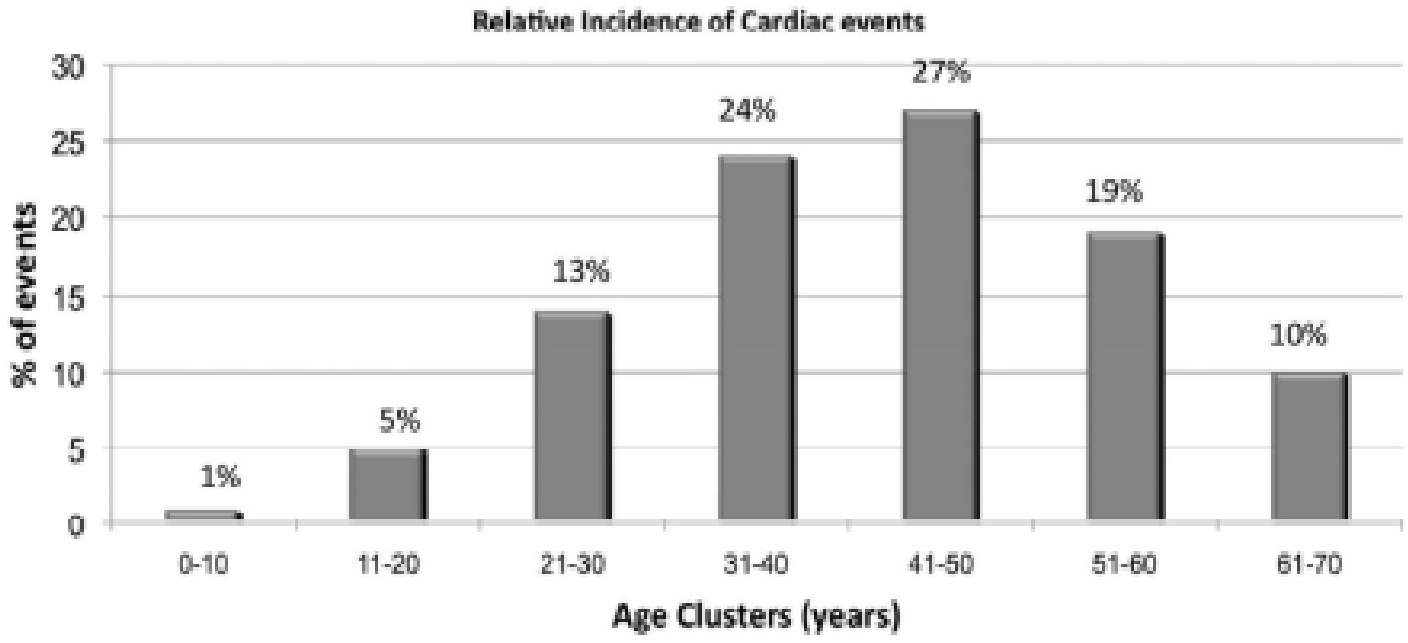


Figure 3. Absolute and annual event rate in the major Brugada syndrome registries.

Napolitano 2012

Symptoms by age cluster in Brugada syndrome



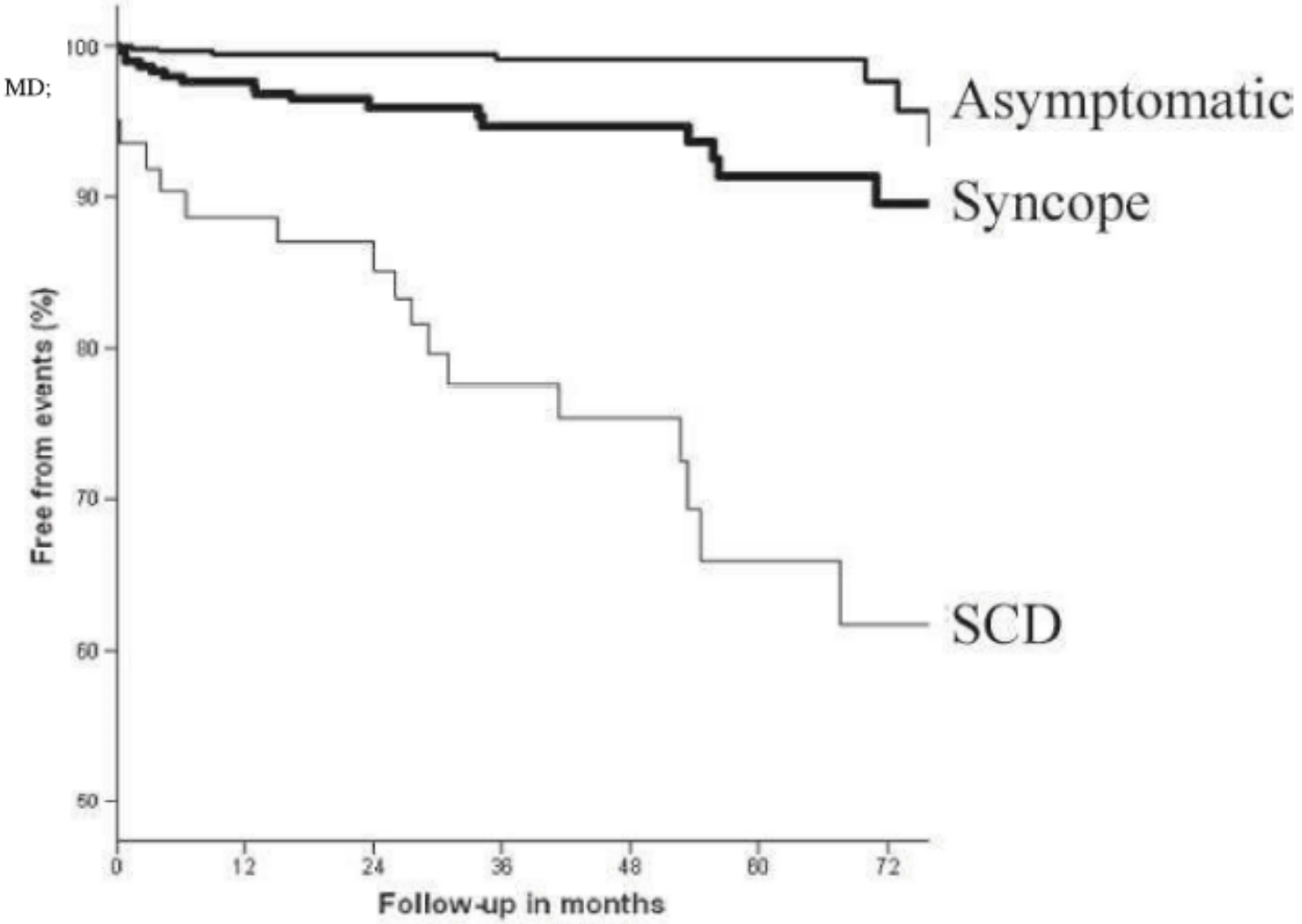
N = 1057 patients - 269 events

Figure 4. Relative percentage of symptomatic Brugada syndrome patients by age clusters showing a peak of incidence in the third and fourth decades of life (data from the Pavia Brugada syndrome registry).

Long-Term Prognosis of Patients Diagnosed With Brugada Syndrome

Results From the FINGER Brugada Syndrome Registry

V. Probst, MD, PhD*; C. Veltmann, MD*; L. Eckardt, MD*; P.G. Meregalli, MD*; F. Gaita, MD;



	0	12	24	36	48	60	72
group A	62	54	47	36	29	18	15
group B	313	244	192	148	99	73	49
group C	654	505	379	275	195	109	54

Long-Term Prognosis of Patients Diagnosed With Brugada Syndrome

Results From the FINGER Brugada Syndrome Registry

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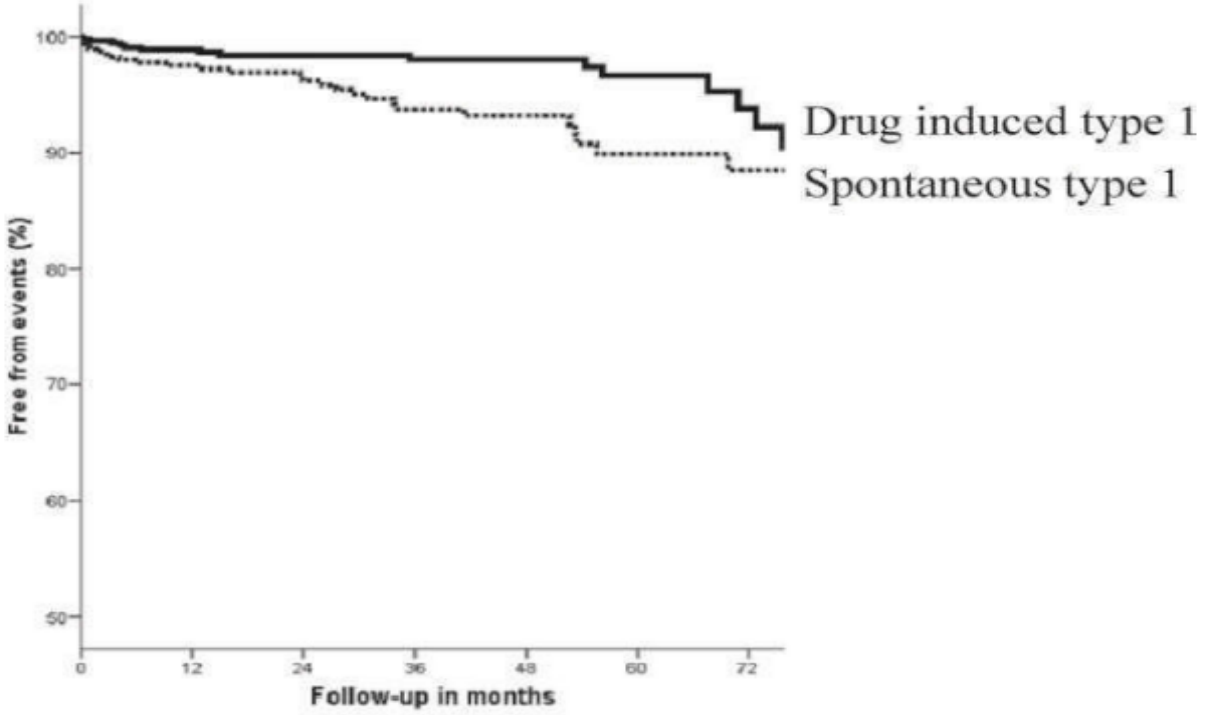
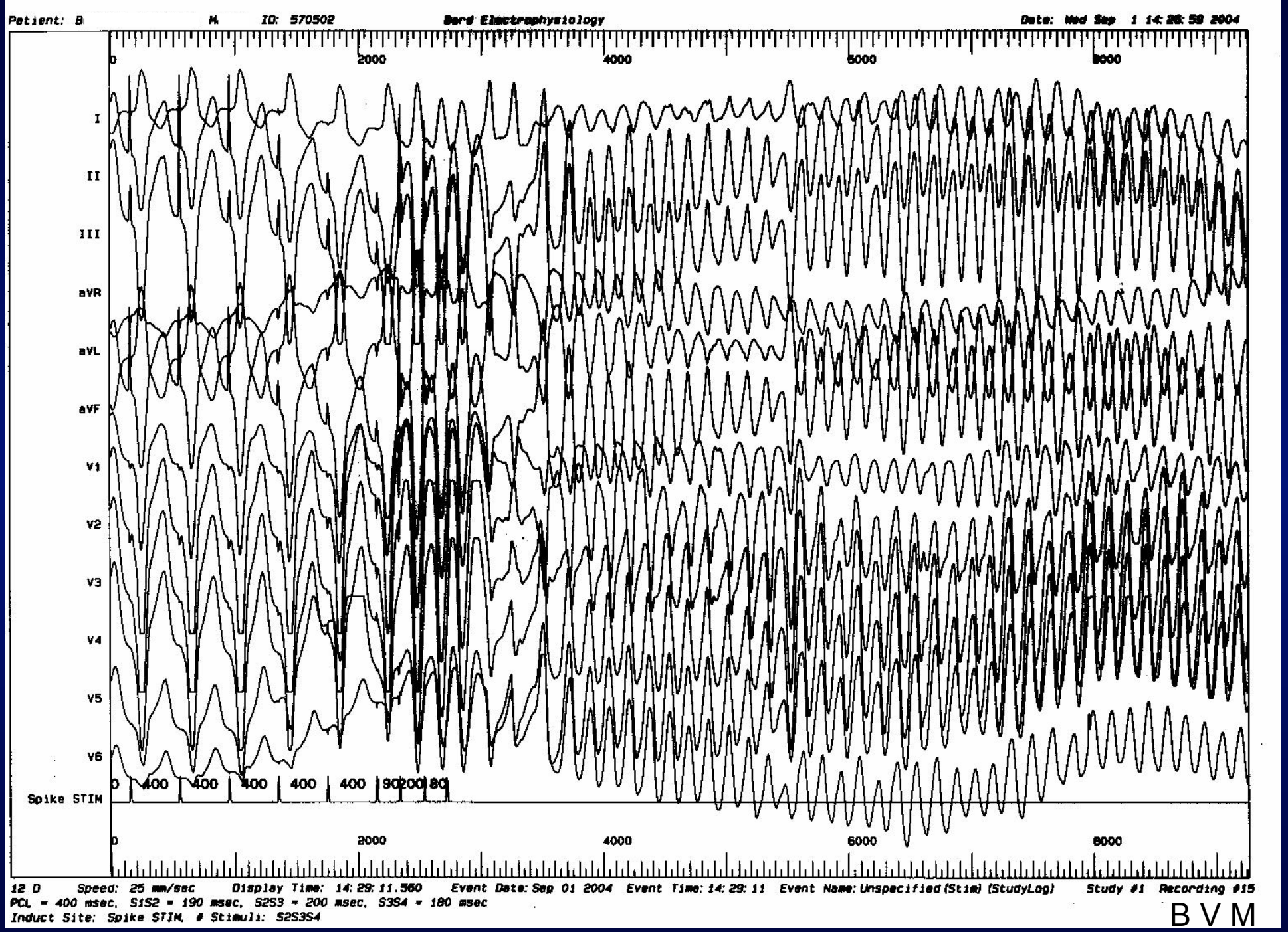


	0	12	24	36	48	60	72
type1	468	350	269	200	135	88	58
no type 1	561	453	349	259	188	112	60

Figure 4. Kaplan-Meier curves of arrhythmic events during follow-up depending on the ECG pattern. Patients displaying a spontaneous type 1 ECG (n=468) had a shorter time to first arrhythmic event than patients in whom the type 1 ECG was induced after challenge with sodium channel blockers (n=561); $P=0.01$.

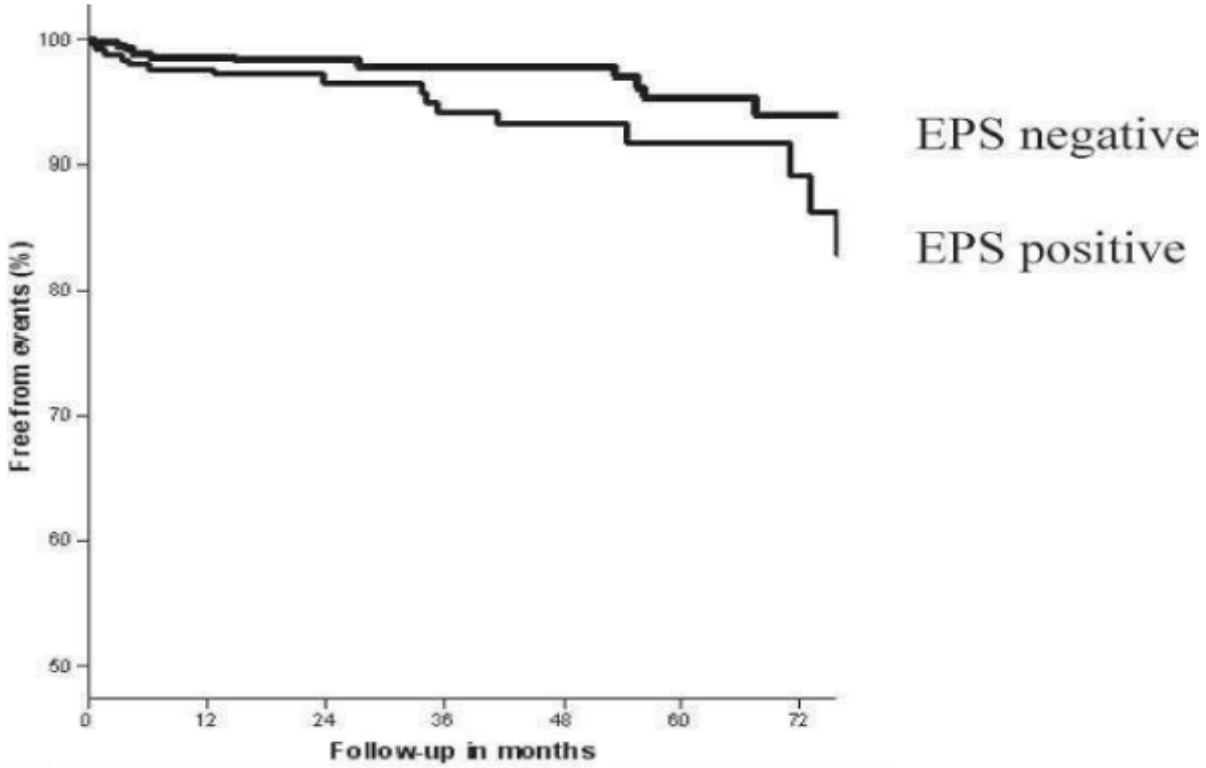
2010


Patient: B M ID: 570502
Bard Electrophysiology
Date: Wed Sep 1 14:28:59 2004
0
2000
4000
6000
8000
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
Spike STIM
0 400 400 400 400 400 90 200 80
12 D Speed: 25 mm/sec Display Time: 14:29:11.560 Event Date: Sep 01 2004 Event Time: 14:29:11 Event Name: Unspecified (Stim) (StudyLog) Study #1 Recording #15
PCL = 400 msec, S1S2 = 190 msec, S2S3 = 200 msec, S3S4 = 180 msec
Induct Site: Spike STIM, # Stimuli: S2S3S4
BVM

Long-Term Prognosis of Patients Diagnosed With Brugada Syndrome

Results From the FINGER Brugada Syndrome Registry

V. Probst, MD, PhD*; C. Veltmann, MD*; L. Eckardt, MD*; P.G. Meregalli, MD*; F. Gaita, MD;



	0	12	24	36	48	60	72
negative	376	301	237	187	136	94	59
positive	262	212	161	113	81	52	34

Figure 5. Kaplan–Meier curves of arrhythmic events during follow-up depending on the results of the EPS. Patients in whom EPS induced ventricular fibrillation (n=262) had a shorter time to the first arrhythmic event than those with a negative EPS (n=376); P=0.05.

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Heart Rhythm Disorder

Risk Stratification in Brugada Syndrome

Results of the PRELUDE (PRogrammed ELectrical stimUlation preDictive valuE) Registry

Silvia G. Priori, MD, PHD,*†‡ Maurizio Gasparini, MD,§ Carlo Napolitano, MD, PHD,*‡

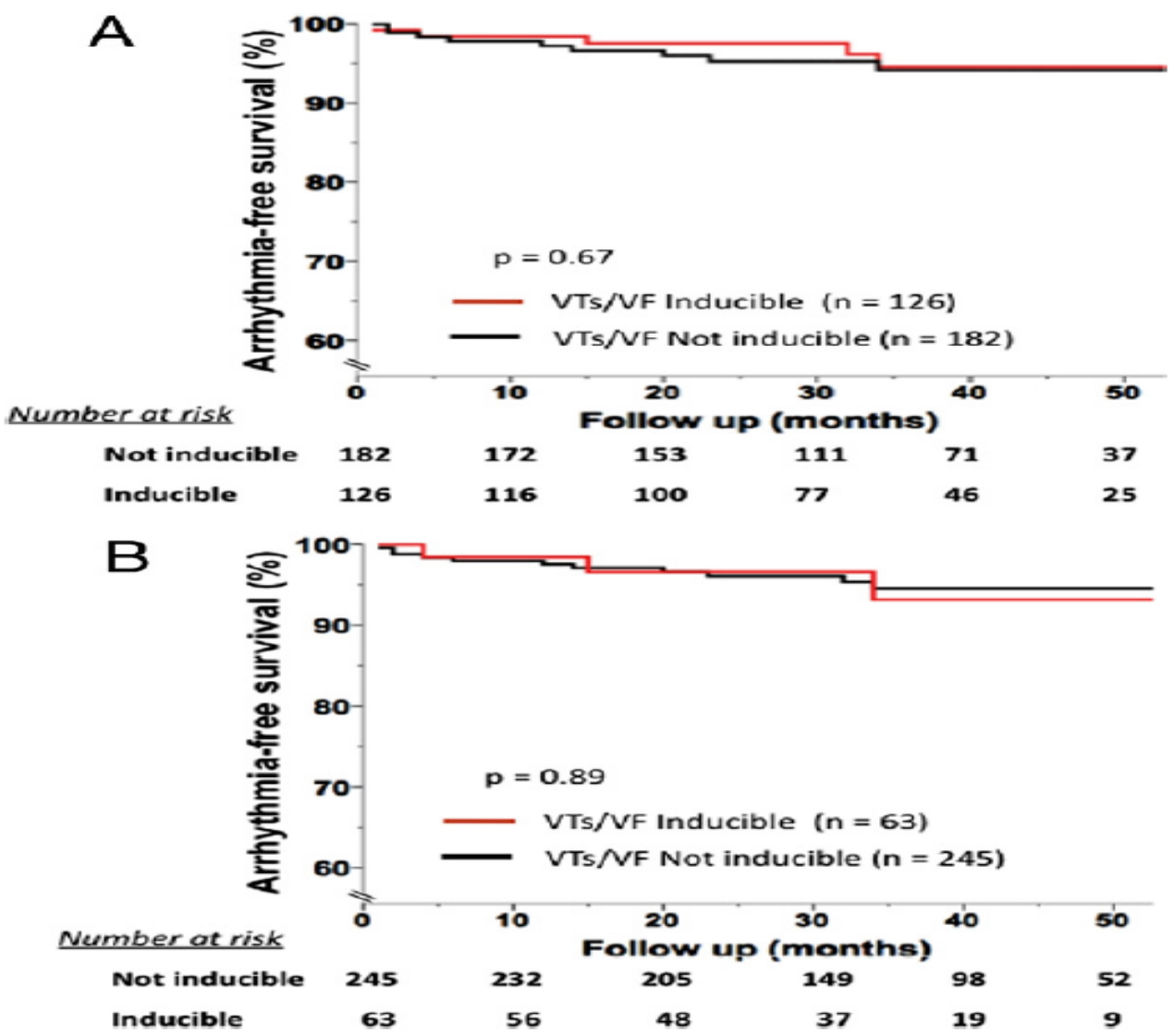

Figure 3 Survival According to VTs/VF Inducibility

Kaplan-Meier survivorship analysis of arrhythmic event-free survival according to the presence/absence of sustained ventricular tachycardia/ventricular fibrillation (VTs/VF) inducibility in the entire PRELUDE cohort **(A)** and in the subgroup of patients inducible with 1 or 2 extrastimuli **(B)**.

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Results of the PRELUDE (PRogrammed ELectrical stimUlation preDictive valuE) Registry

Silvia G. Priori, MD, PHD,*†‡ Maurizio Gasparini, MD,§ Carlo Napolitano, MD, PHD,*‡

Spontaneous type I Syncope

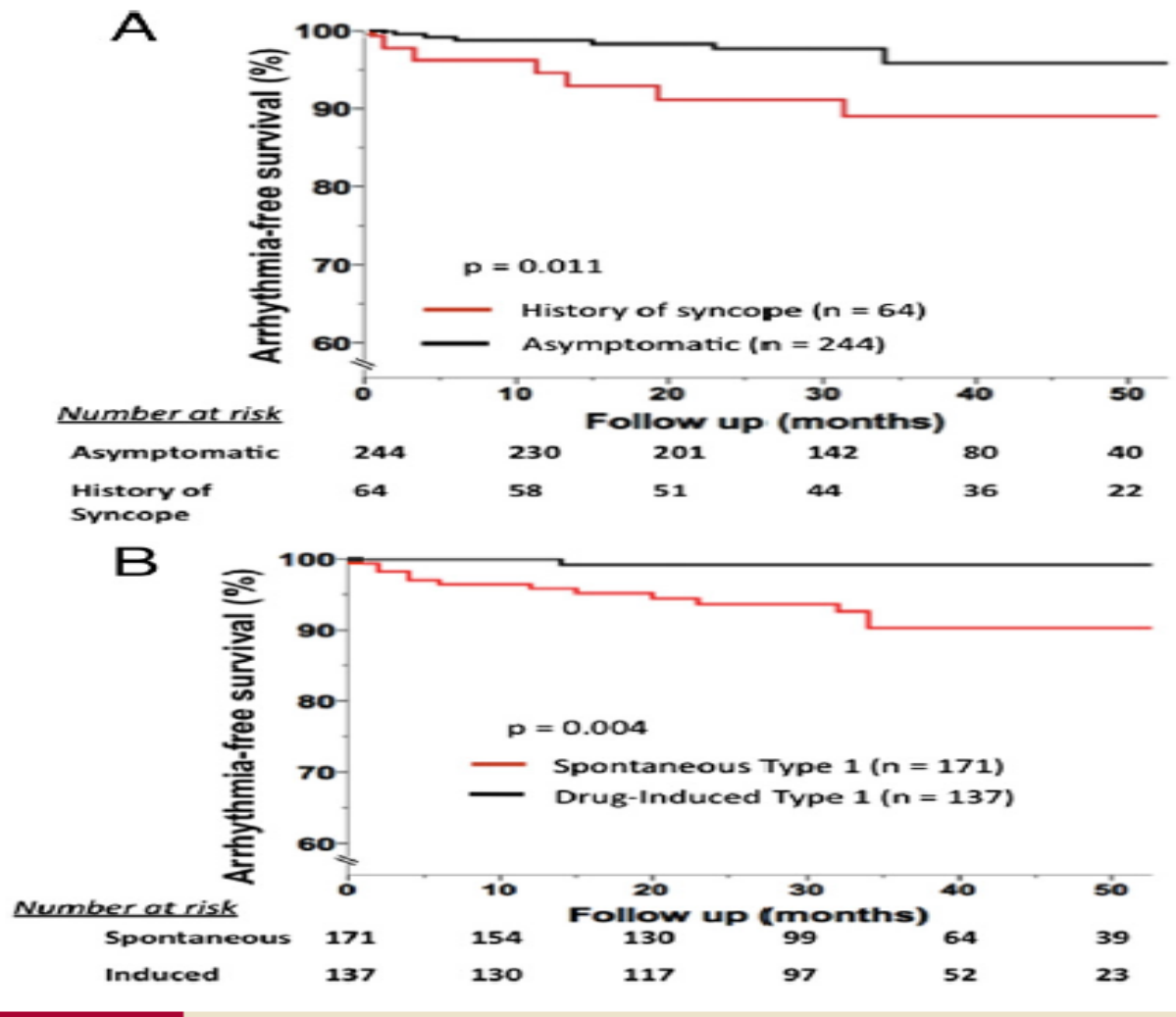

Figure 4 Survival According to Syncope Before Enrollment and ECG Presentation

Kaplan-Meier survivorship analysis of arrhythmic event-free survival according to (A) history of syncope before enrollment and (B) presence or absence of spontaneous type 1 electrocardiogram (ECG).

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Heart Rhythm Disorders

Risk Stratification in Brugada Syndrome

Results of the PRELUDE (PRogrammed ELectrical stimUlation preDictive valuE) Registry

Silvia G. Priori, MD, PHD,*†‡ Maurizio Gasparini, MD,§ Carlo Napolitano, MD, PHD,*‡
Paolo Della Bella, MD,‖ Andrea Ghidini Ottonelli, MD,¶ Biagio Sassone, MD,#
Umberto Giordano, MD,** Carlo Pappone, MD,†† Giosuè Mascioli, MD,‡‡ Guido Rossetti, MD,§§
Roberto De Nardis MD,‖‖ Mario Colombo, MS¶¶

Pavia, Rozzano, Milano, Lido di Camaiore, Bentivoglio, Palermo, Ravenna, Bergamo, Cuneo, and Vicenza, Italy; and New York, New York

At present however, there is no robust algorithm to link BrS genotype with either a specific phenotype or a distinctive risk profile. As a consequence, genetic testing cannot guide therapeutic decisions.

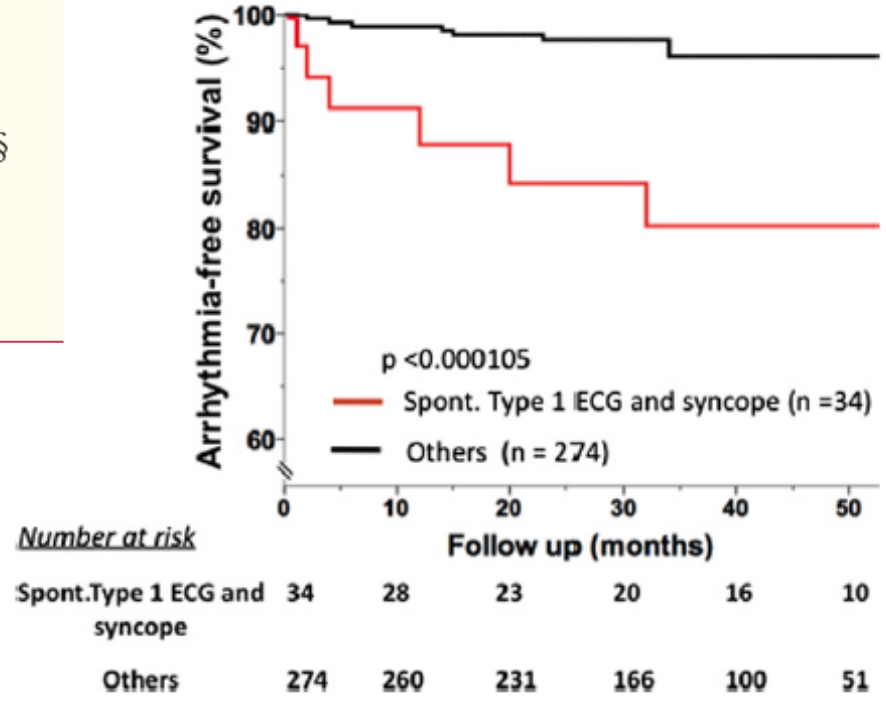


Figure 6 Survival According to Spontaneous Type 1 ECG and History of Syncope

Kaplan-Meier survivorship analysis of arrhythmic event-free survival according to the presence of spontaneous (Spont.) type 1 electrocardiogram (ECG) and history of syncope.

EUROPEAN SOCIETY OF CARDIOLOGY®
Europace (2011) **13**, 1077–1109
doi:10.1093/europace/eur245

HRS/EHRA EXPERT CONSENSUS STATEMENT

HRS/EHRA Expert Consensus Statement on the State of Genetic Testing for the Channelopathies and Cardiomyopathies

This document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA)

Michael J. Ackerman, MD, PhD[1], Silvia G. Priori, MD, PhD[2], Stephan Willems, MD,

III. State of genetic testing for Brugada syndrome (BrS)

Expert Consensus Recommendations

(1) Comprehensive or BrS1 (*SCN5A*) targeted BrS genetic testing **can be useful** for any patient in whom a cardiologist has established a clinical index of suspicion for BrS based on examination of the patient's clinical history, family history, and expressed electrocardiographic (resting 12-lead ECGs and/or provocative drug challenge testing) phenotype.

(2) Genetic testing **is not indicated** in the setting of an isolated type 2 or type 3 Brugada ECG pattern.

(3) Mutation-specific genetic testing **is recommended** for family members and appropriate relatives following the identification of the BrS-causative mutation in an index case.

Table 1

Drugs to be avoided by Brugada syndrome patients

Drug category	Drug (generic)	Recommendation
Antiarrhythmic drugs	Ajmaline[26-29]	Class I
	Flecainide[30-34]	Class I
	Pilsicainide[35-38]	Class I
	Procainamide[17,26,39,40]	Class I
	Propafenone[41-45]	Class IIa
Psychotropic drugs	Amitriptyline[46-49]	Class IIa
	Clomipramine[50,51]	Class IIa
	Desipramine[52-55]	Class IIa
	Lithium[52,56]	Class IIa
	Loxapine[47,57]	Class IIa
	Nortriptyline[55,58,59]	Class IIa
	Trifluoperazine[47,60]	Class IIa

Table 2

Drugs preferably avoided by Brugada syndrome patients

Drug category	Drug (generic)	Recommendation
Antiarrhythmic drugs	Amiodarone[77-79]	Class IIb
	Cibenzoline[80-82]	Class IIb
	Disopyramide[14,17,83-85]	Class IIb
	Lidocaine[17,86*]	Class IIb
	Propranolol[17,18,70,87,88]	Class IIb
	Verapamil[17,89,90]	Class IIb
Psychotropic drugs	Carbamazepine[91,92]	Class IIb
	Cyamemazine[47,93]	Class IIb
	Doxepin[48,94]	Class IIb
	Fluoxetine[47,51]	Class IIb
	Imipramine[95]	Class IIb
	Maprotiline[46,96]	Class IIb

BrugadaDrugs.org

Safe drug use and the Brugada syndrome

Table 3

Potential antiarrhythmic drugs in Brugada syndrome patients

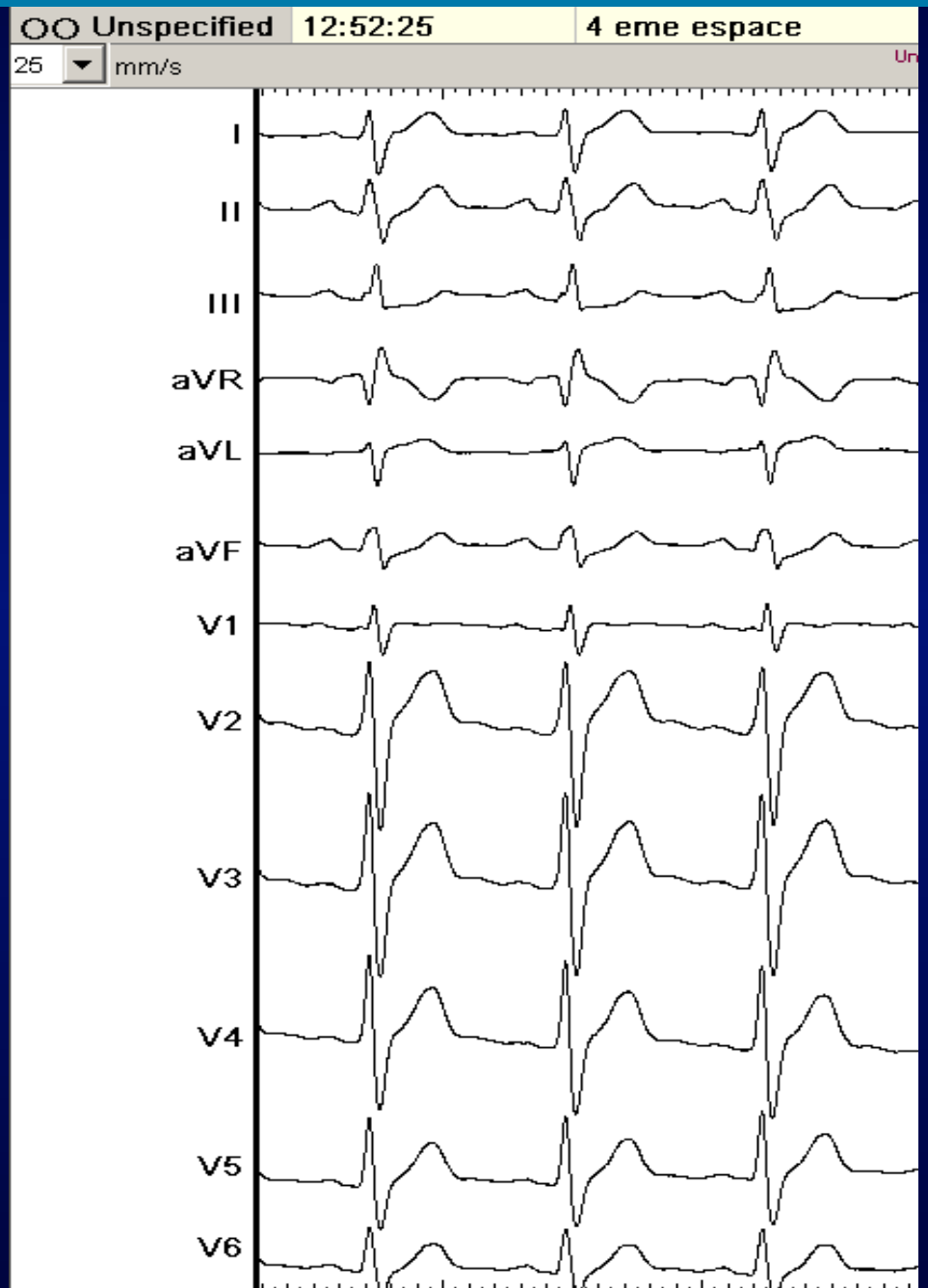
Drug category	Drug (generic)	Recommendation
Antiarrhythmic drugs	Isoproterenol / Isoprenaline[15,17,113,114*]	Class I
	Orciprenaline[115]	Class IIa
	Quinidine[8-10,15,116,117†]	Class I
Other substances	Cilostazol[118-120]	Class IIb

Recommendation: Class I: convincing evidence/opinion; Class IIa: evidence/opinion less clear; Class IIb: conflicting evidence/opinion.

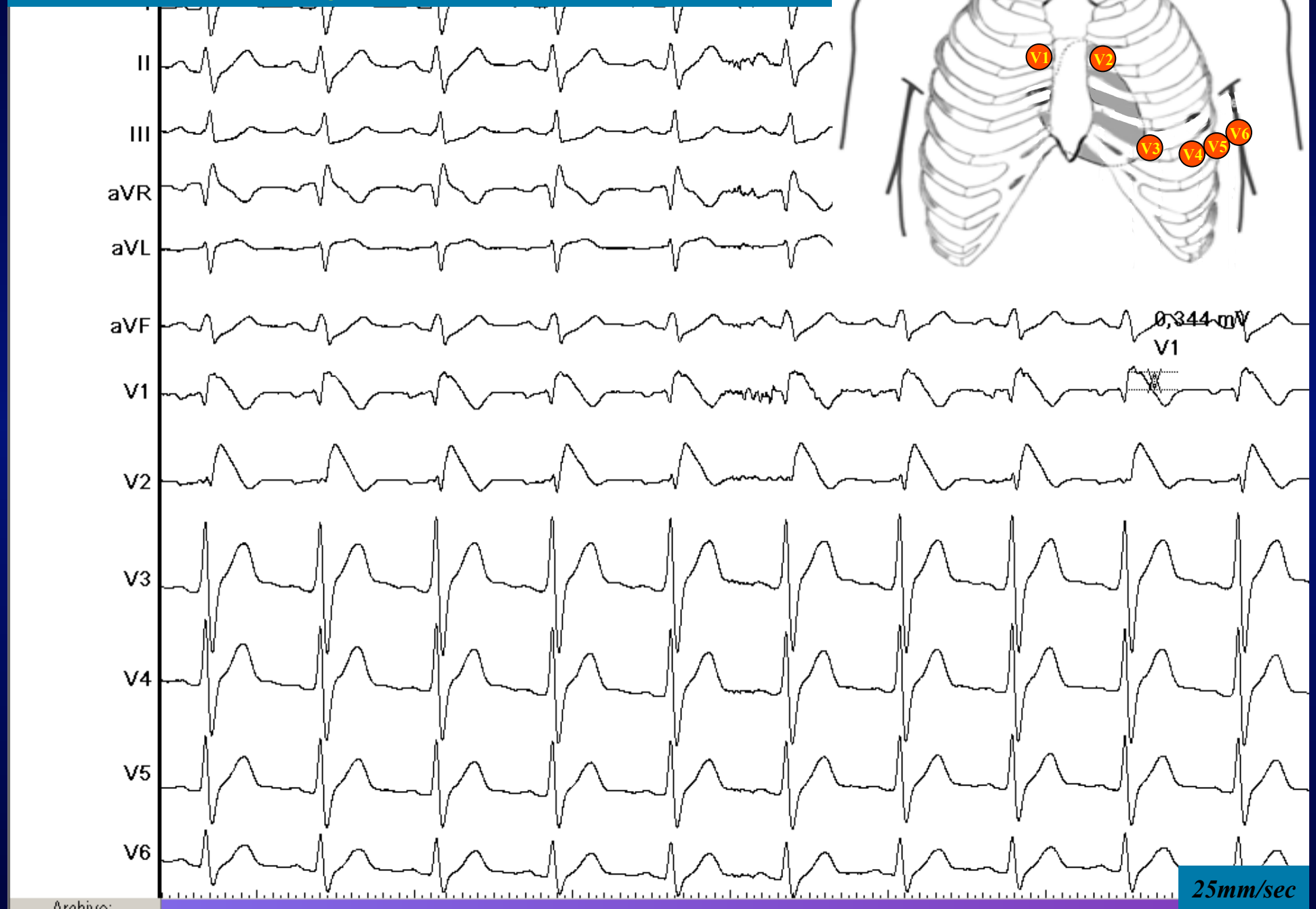
CASE R. V.

- ♂ 38 years
- History : - 2002-Atypical thoracic pain
  Coronary angiogram : normal (U.S.A.)
  ECG : atypical RBBB
  EPS : Procaïnamide : no modification
- PVS : no ventricular vulnerability.
  - 2006-Ajmaline (75 mg IV)

Post Ajmaline



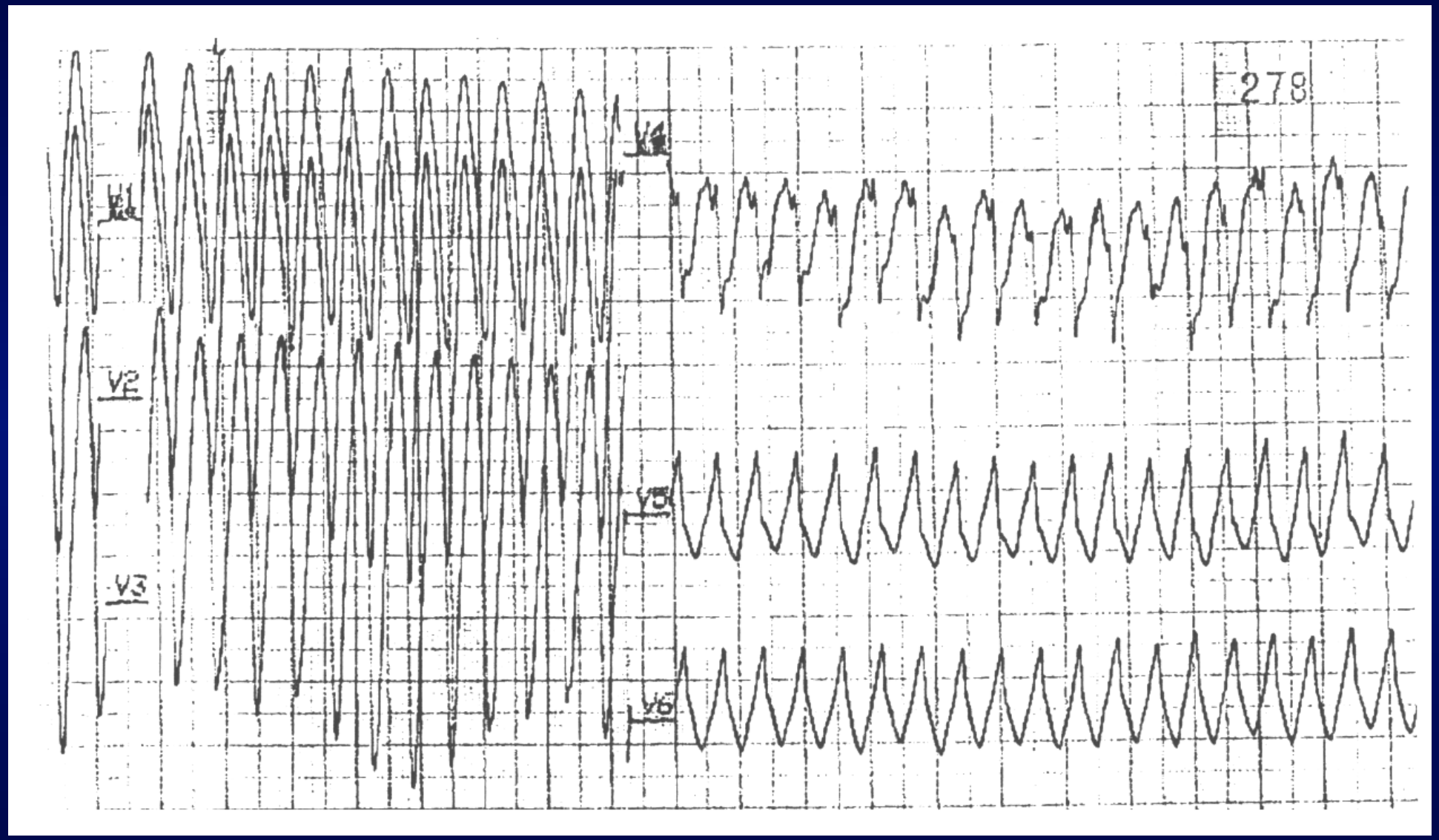
Post Ajmaline 2e EIC



Post Aimaline

4th intercostal space 3th intercostal space 2th intercostal space

CAS CLINIQUE



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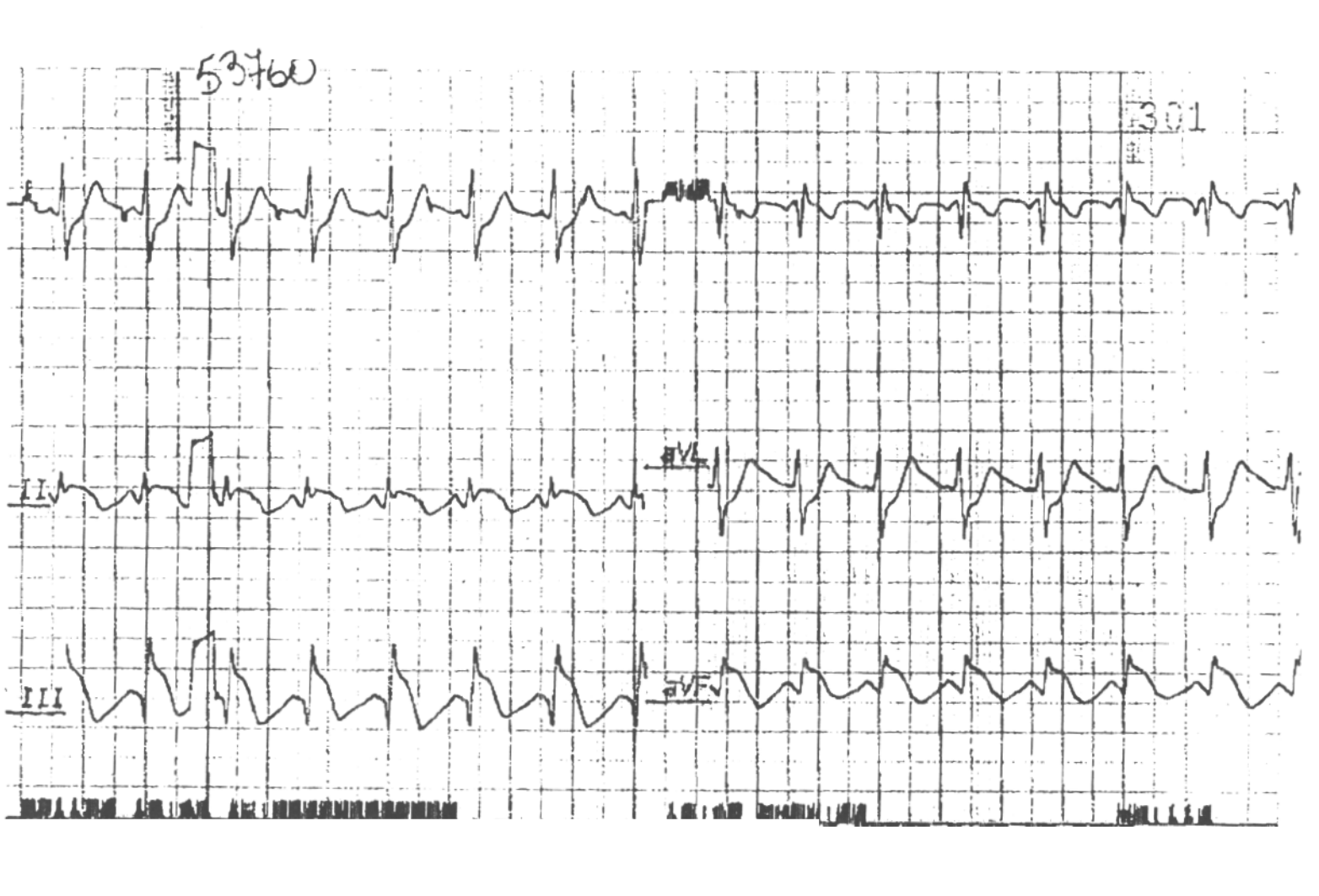
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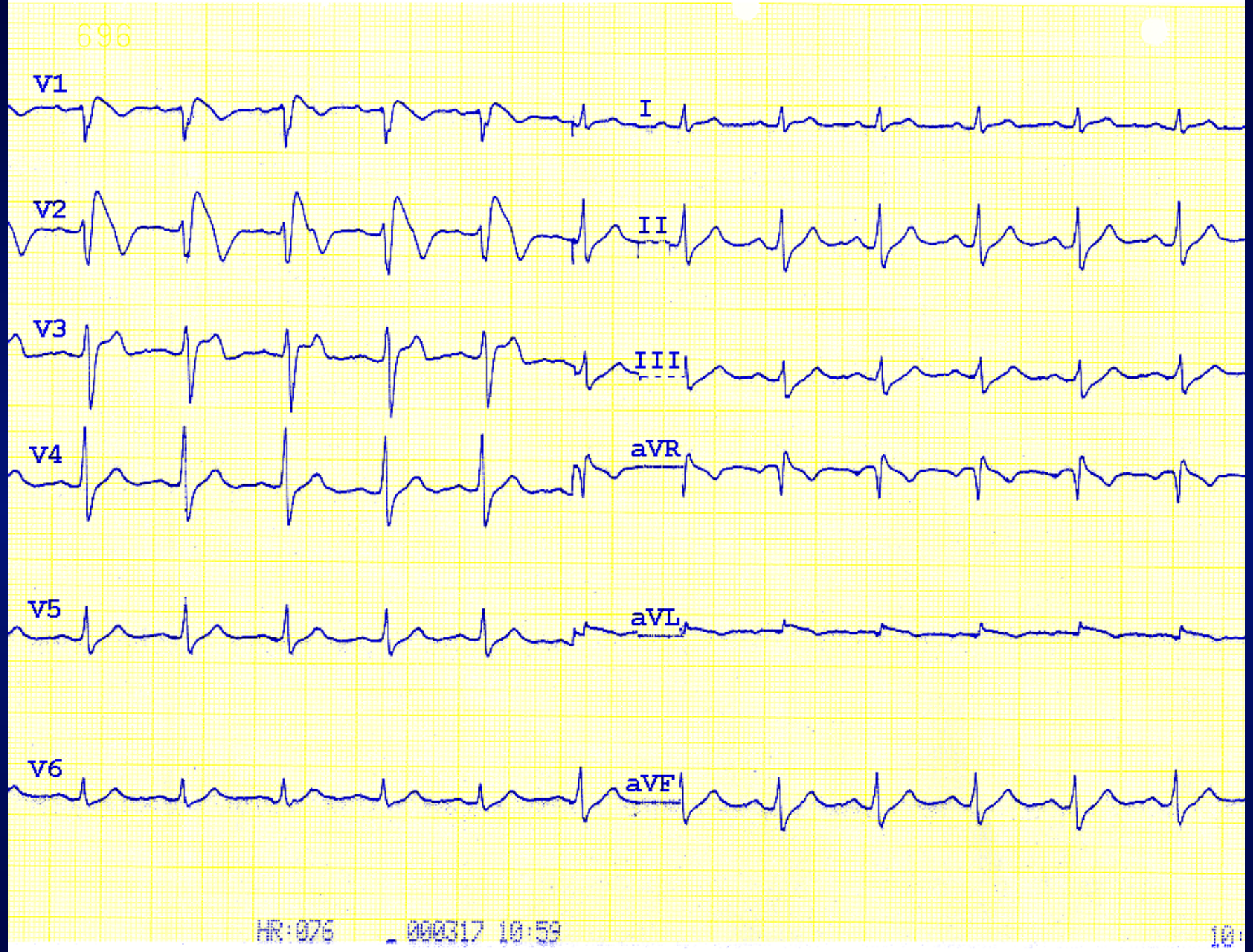
301
II
aVL
III
aVF



C E 580306 (Mère)

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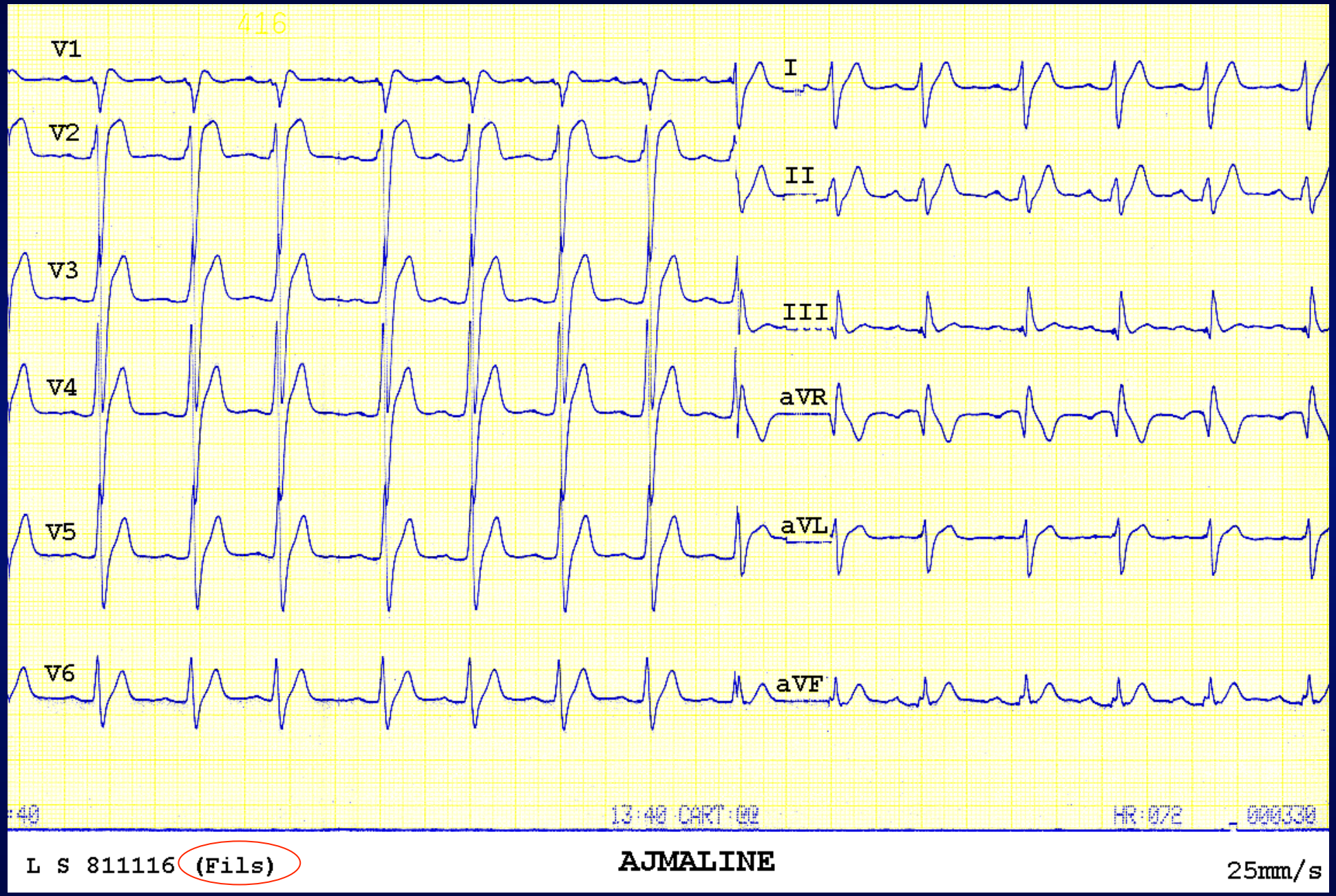
416
V1
V2
V3
V4
V5
V6
I
II
III
aVR
aVL
aVF
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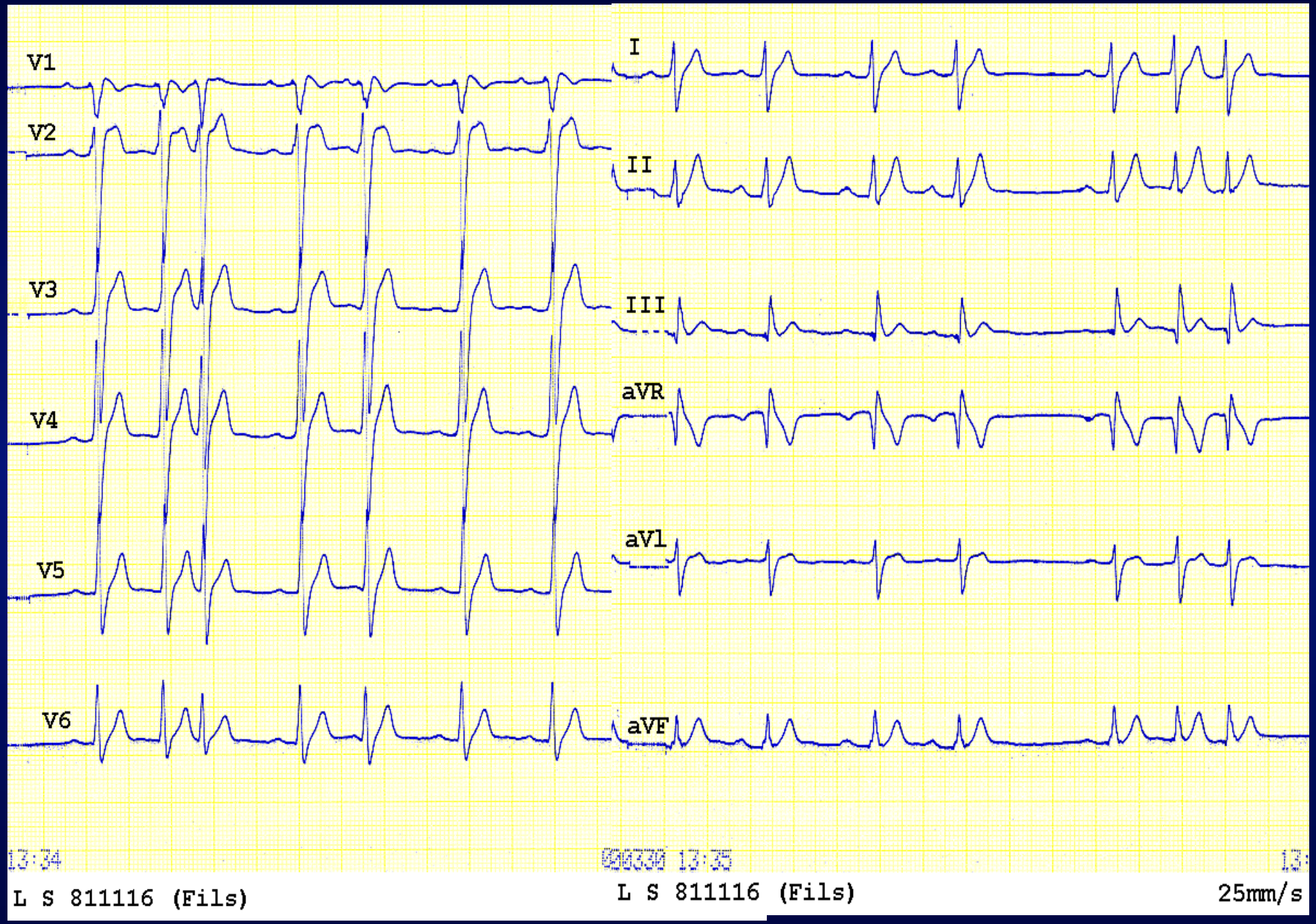
V1
V2
V3
V4
V5
V6
I
II
III
aVR
aVl
aVF
L S 811116 (Fils)
L S 811116 (Fils)
25mm/s

422
I
II
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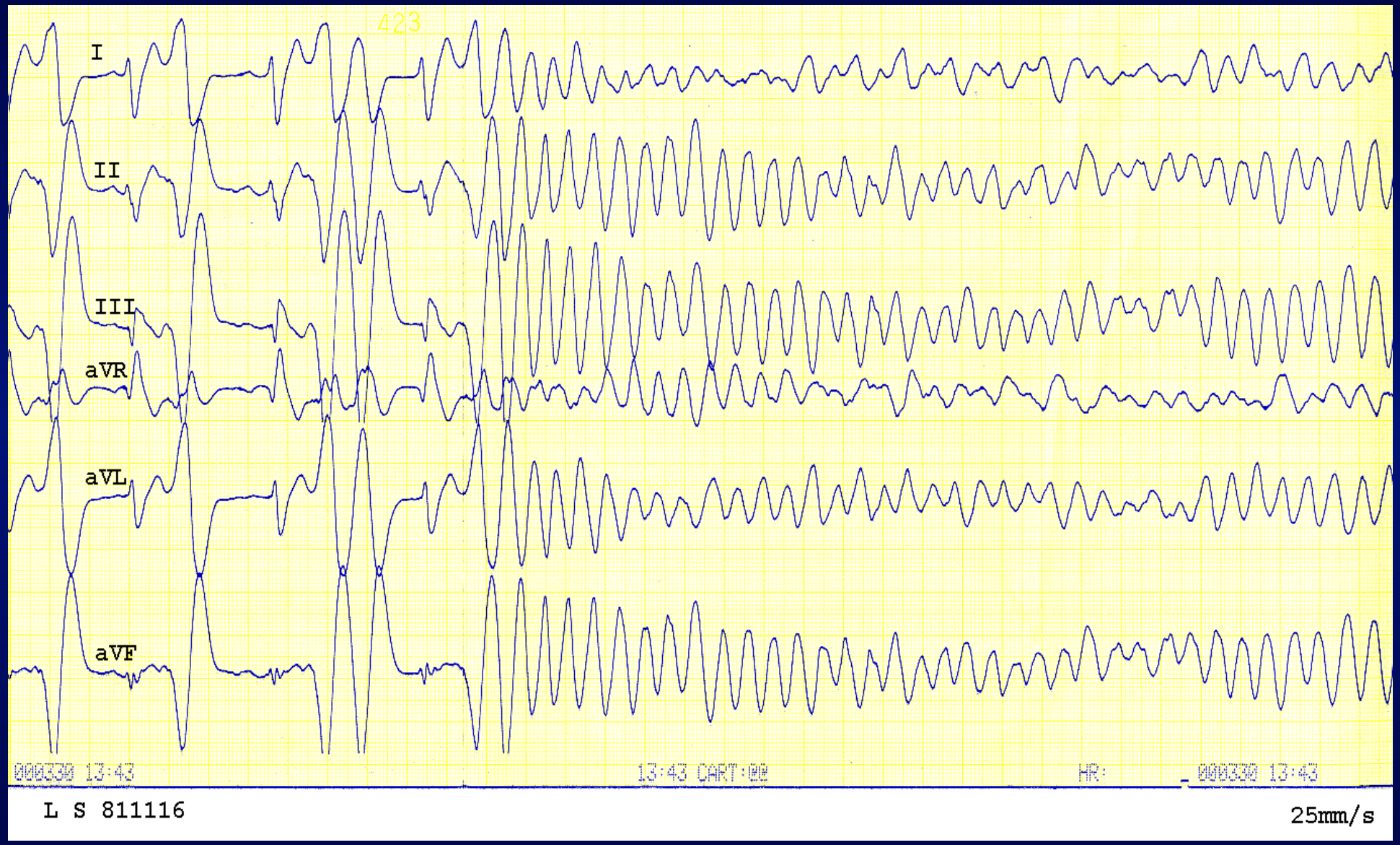
25mm/s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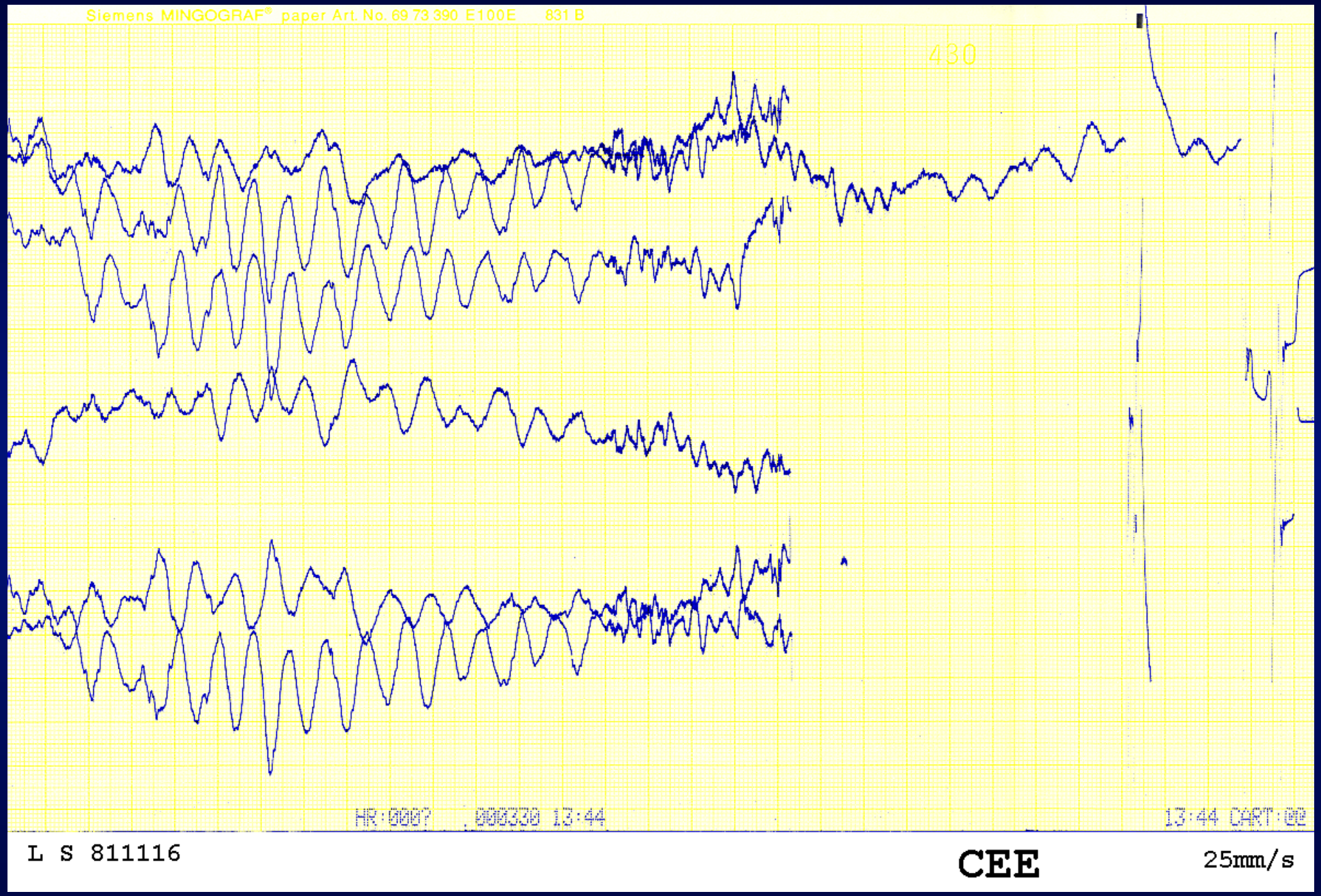
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L S 811116
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25mm/s

Analyse:

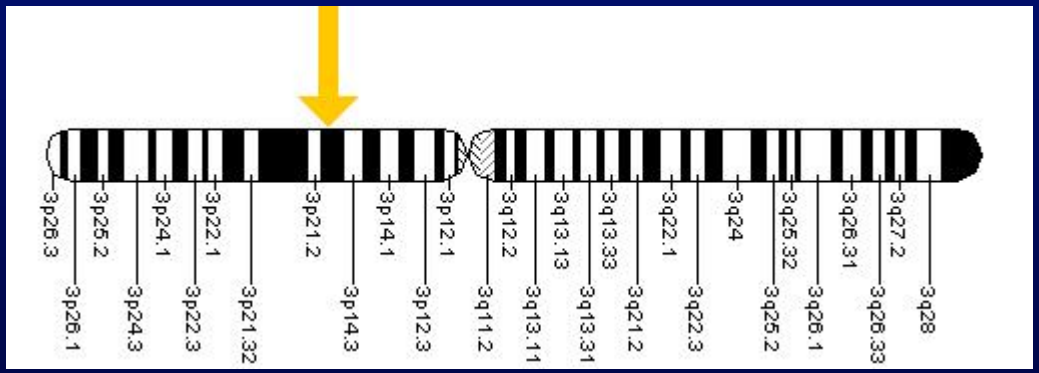
Recherche de mutation ponctuelle du gène *SCN5A* par amplification PCR de l'entièreté de la région codante et des jonctions exon-intron suivie du séquençage direct:

→ Présence à l'état hétérozygote d'une transition de la cytosine située en position -3 du site accepteur d'épissage de l'intron 9 par une adénine:

c.1253-3C>A.

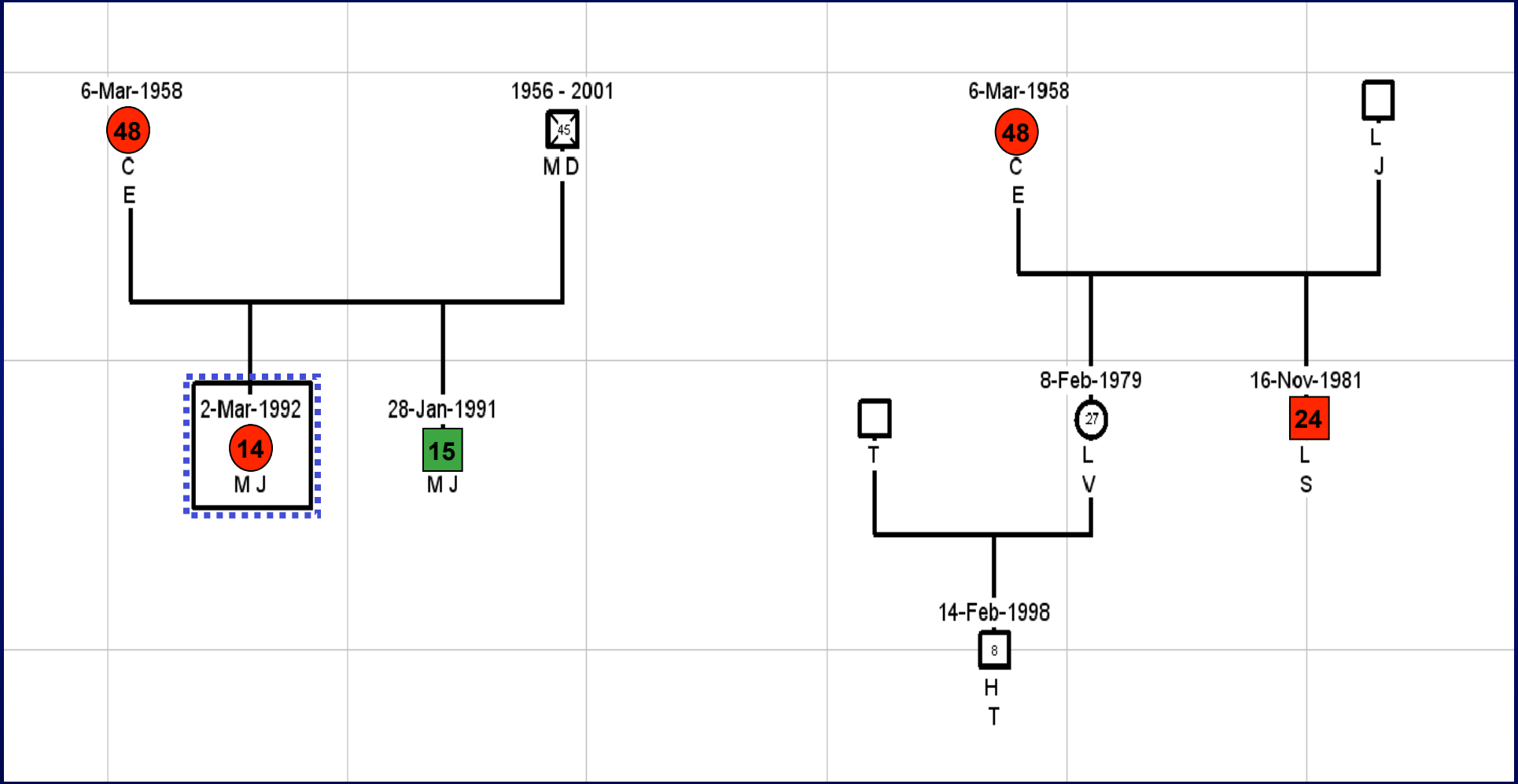
Cette mutation est théoriquement considérée **comme une mutation affectant l'épissage de l'exon 10** et donc responsable de sa non-transcription dans l'ARNm (skipping de l'exon 10). La délétion de l'exon 10 respecterait le cadre de lecture et entraînerait la perte du 6° domaine transmembraire au niveau protéique (acides aminés 385-450).

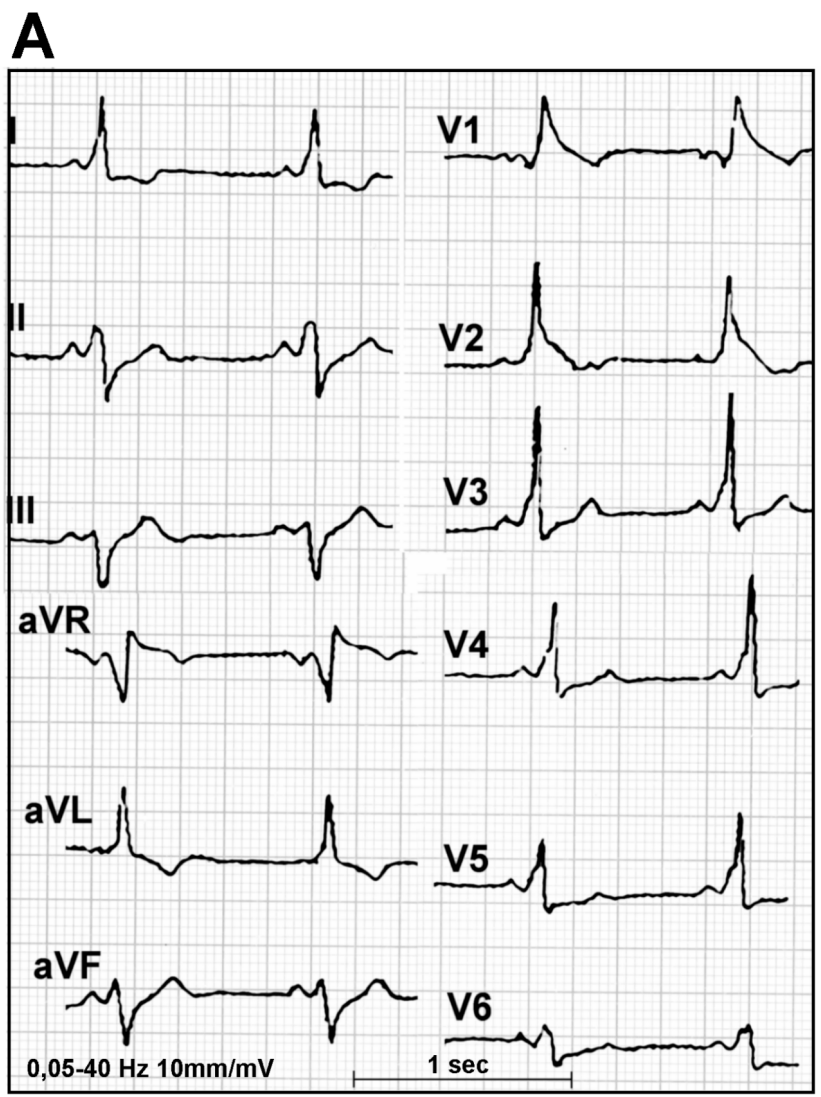
Gène SCN5A: mutation c.1253-3C>A

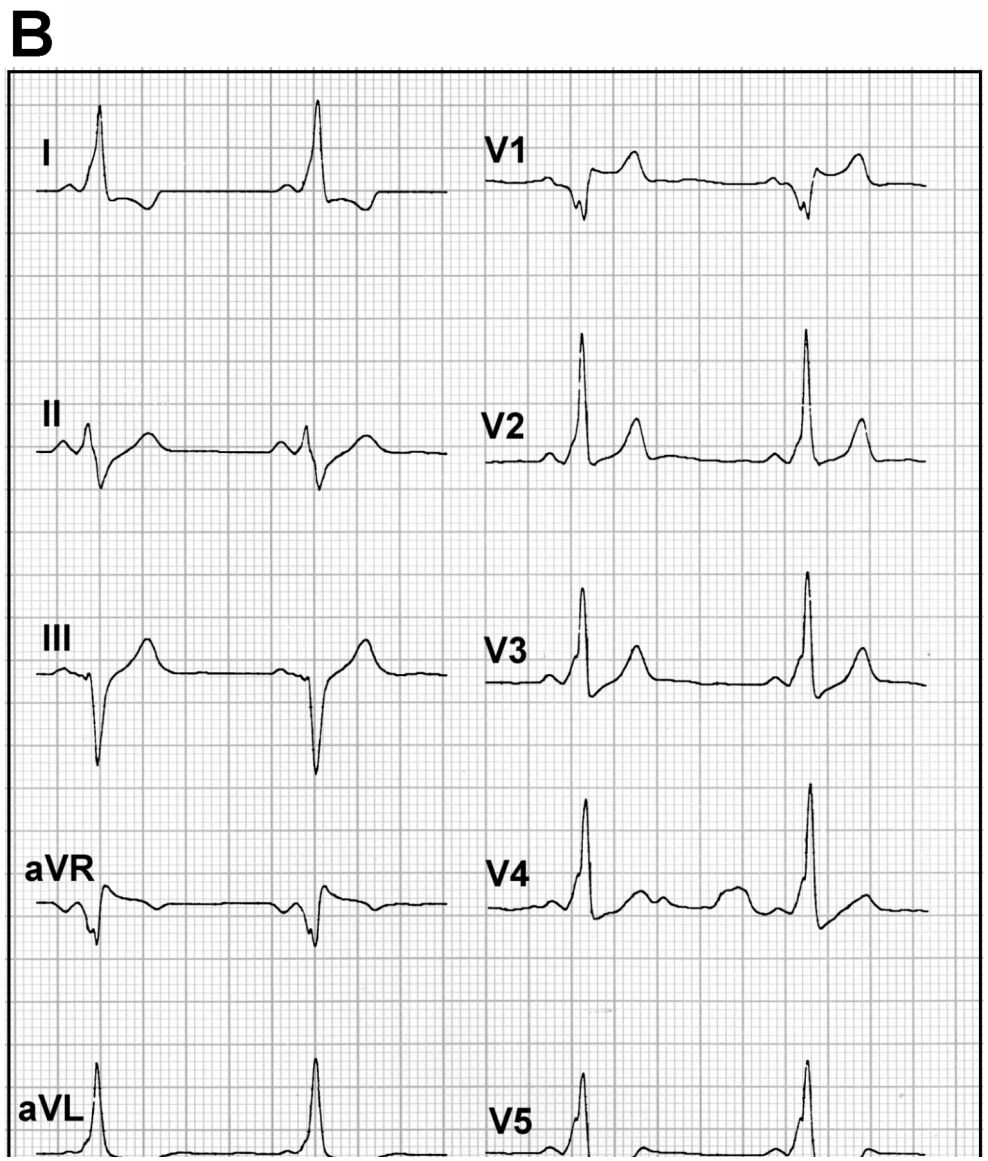


CHROMOSOME 3

Cas Clinique







J Interv Card Electrophysiol. 2007 Nov;20(1-2):39-41. Epub 2007 Oct 18.

Variable morphologies of preexcitation due to a concomitant Brugada pattern.

De Roy L[1], Popescu E, Floria M, Blommaert D, Deceuninck O, Collet B, Dormal F, Alsteen G, de Olival JR.

Tracés à des moments différents

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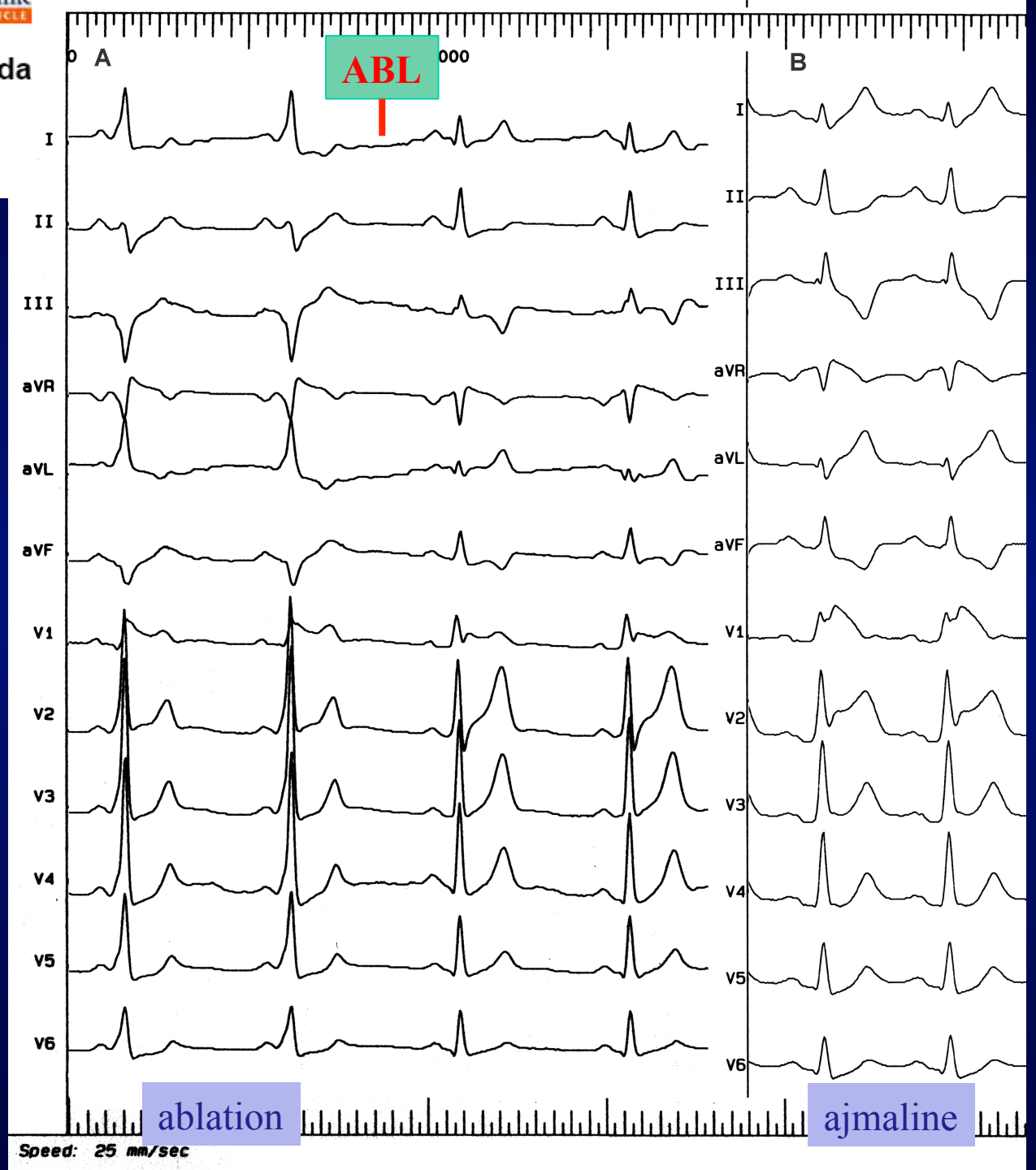
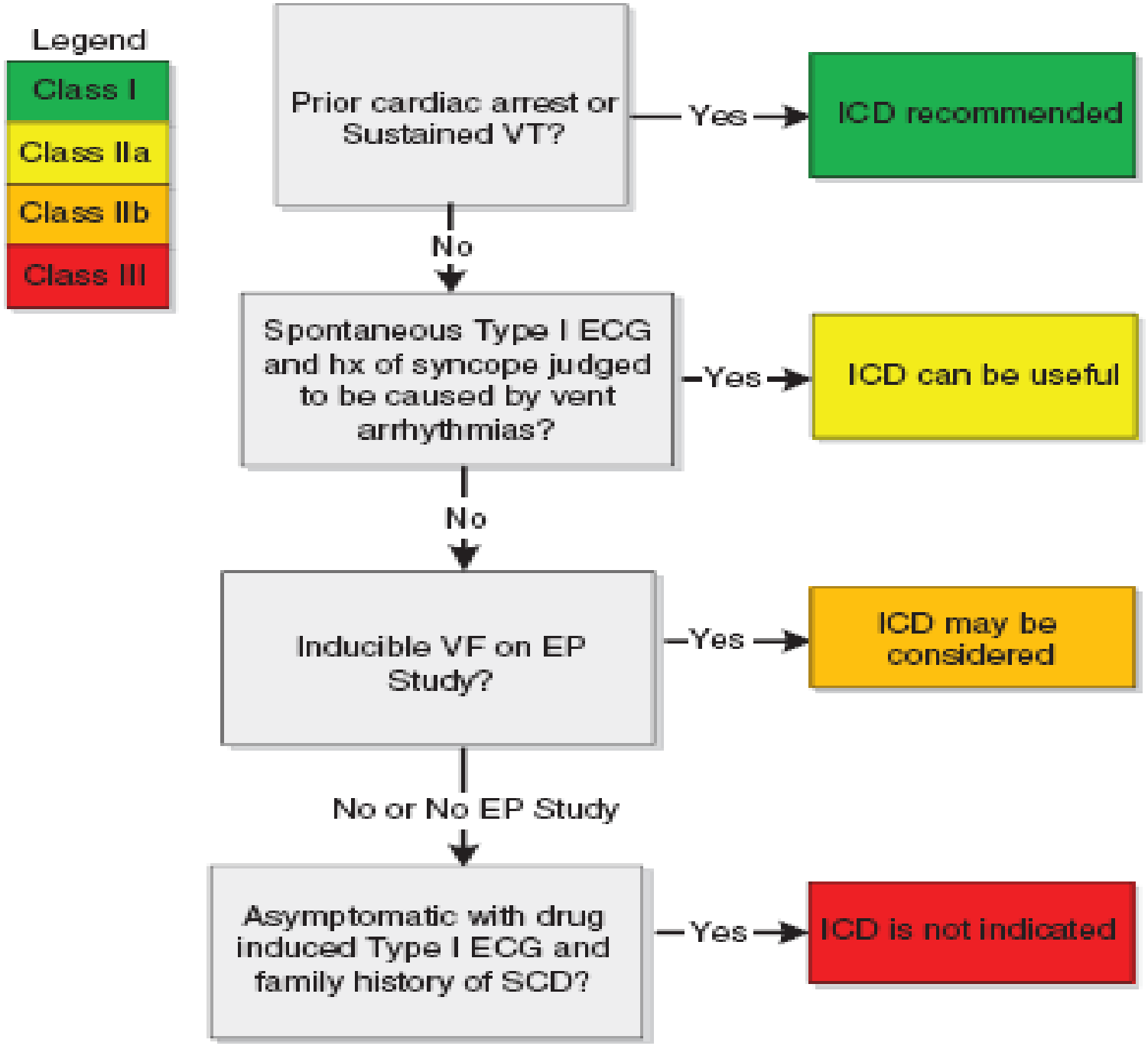
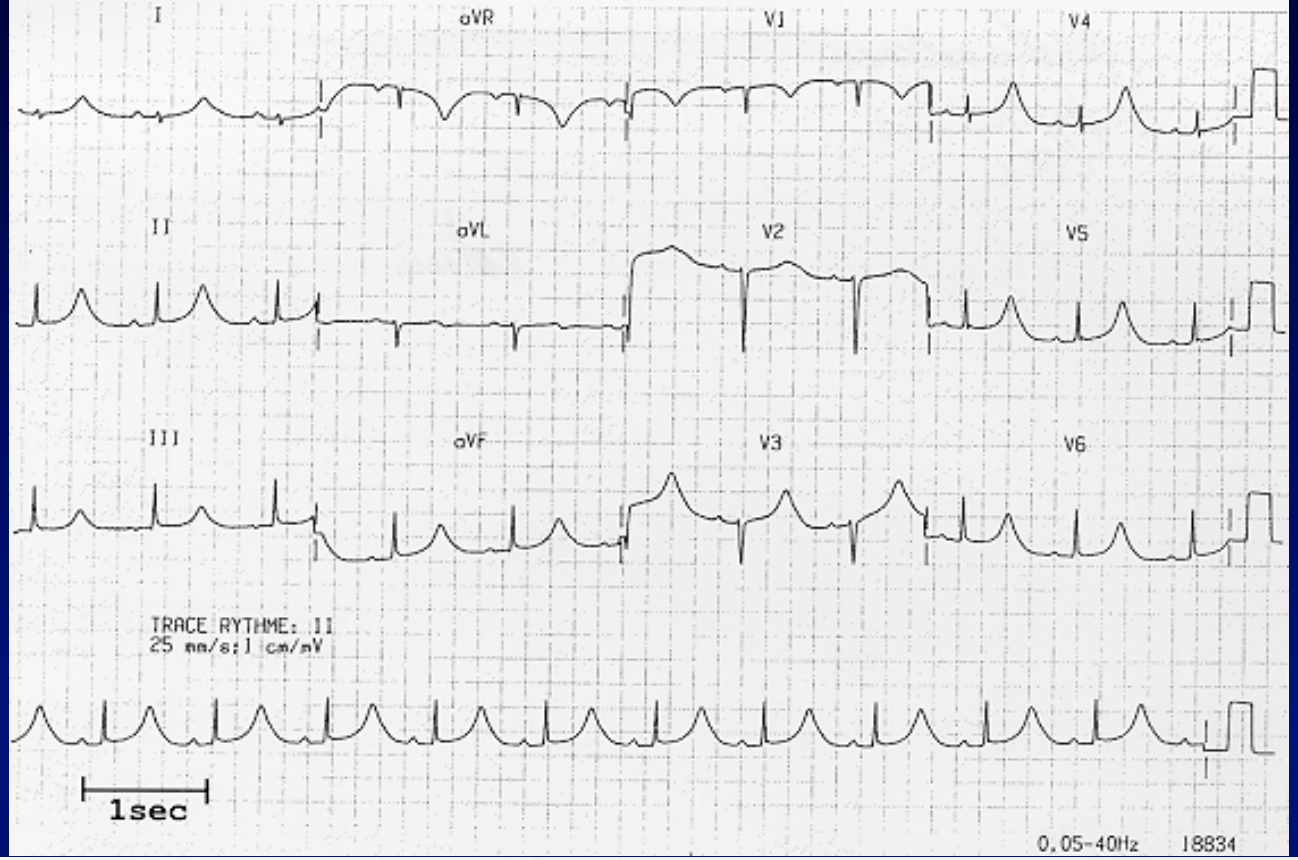


Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Silvia G. Priori, (HRS Chairperson)[1], Arthur A. Wilde, (EHRA Chairperson)[2],



LQT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aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
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1sec
0.05-40Hz
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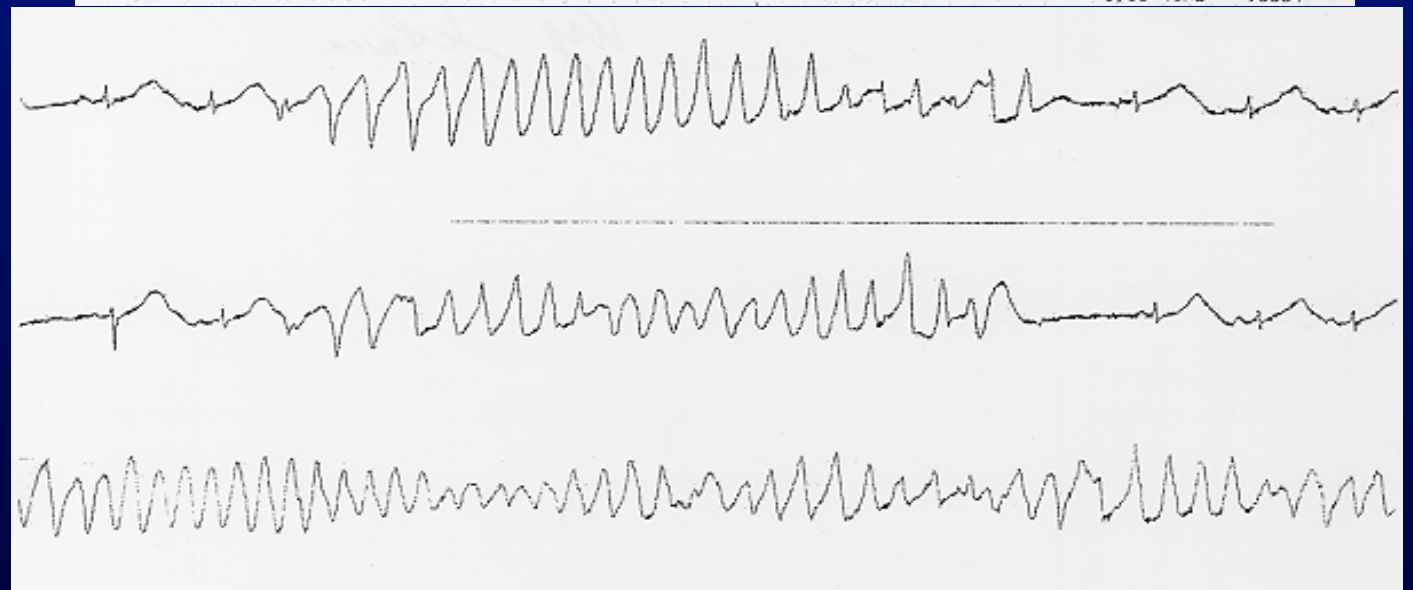

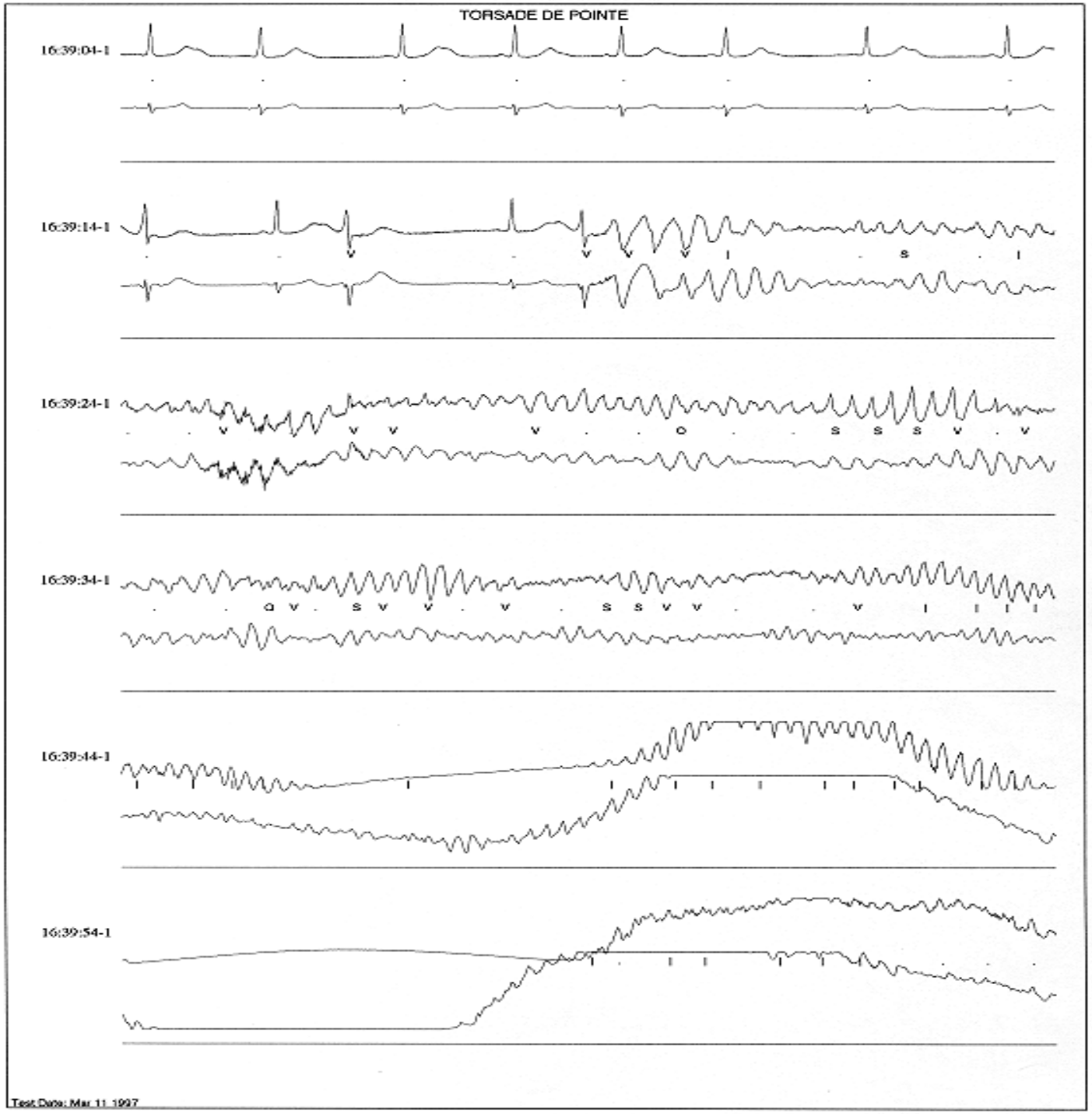
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Test Date: Mar 11 1997

Genetics of Long-QT Syndrome

Genetics of the Long-QT Syndrome

	LQT1	LQT2	LQT3	LQT4	LQT5
Chromosome locus	11p15.5	7q35-36	3p21-24	4q25-27	21q22.1
Gene mutation/protein	KvLQT1	HERG	SCN5A	?	KCNE1*
Current/channel (subunit)	I_{Ks} (α)†	I_{Kr}	I_{Na}	?	I_{Ks} (β)†
Channel function	Reduced function	Reduced function	Gain of function	?	Reduced function
Action potential	Delayed phase 3	Delayed phase 3	Prolonged phase 2	?	Delayed phase 3
Clinical syndrome					
Heterozygous mutation	R-W	R-W	R-W	R-W	R-W
Homozygous mutation	J-L-N	NR	NR	NR	J-L-N

R-W indicates Romano-Ward syndrome; J-L-N, Jervell and Lange-Nielsen syndrome with deafness; and NR, not reported.

*KCNE1 is also referred to as minK.

†The KvLQT1 gene encodes the α-subunit protein of the I_{Ks} channel, with the KCNE1 gene encoding the β-subunit protein of this same channel. The α- and β-subunit proteins combine together to form a cardiac potassium channel expressing the I_{Ks} current.

Zipes and Wellens
1998

Table 2. Genes Involved in the Long-QT Syndrome

Variant	Gene	Protein	Effect of Mutations
LQT1	*KCNQ1*	KvLQT1	Reduced I_{Ks}
LQT2	*KCNH2*	HERG	Reduced I_{Kr}
LQT3	*SCN5A*	Nav1.5	Increased I_{Na}
LQT4	*ANK2*	Ankyrin B	Reduced membrane expression of Na^+ and Ca^{2+} channels
LQT5	*KCNE1*	MinK	Reduced I_{Ks}
LQT6	*KCNE2*	MiRP	Reduced I_{Kr}
LQT7, Andersen syndrome	*KCNJ2*	Kir2.1	Reduced outward I_{K1}
LQT8, Timothy syndrome	*CACNA1c*	Cav1.2	Increased I_{Ca}
LQT9	*CAV3*	Cardiac caveolin gene	Increased I_{Na} resulting from altered gating kinetic
LQT10	*SCN4B*	Sodium channel β_4 subunit	Reduced subunit expression causing increased I_{Na}
LQT11	*AKAP9*	Yotiao	Impaired I_{Ks} activation by catecholamines
LQT12	*SNTA1*	Syntrophin	Reduced NaV1.5 nitrosylation and increased current
LQT13	*KCNJ5*	Kir3.4/GIRK4	Reduced I_{KAch}, acetylcholine-dependent potassium current

Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Silvia G. Priori, (HRS Chairperson)[1], Arthur A. Wilde, (EHRA Chairperson)[2],

Expert Consensus Recommendations on ***LQTS Diagnosis***

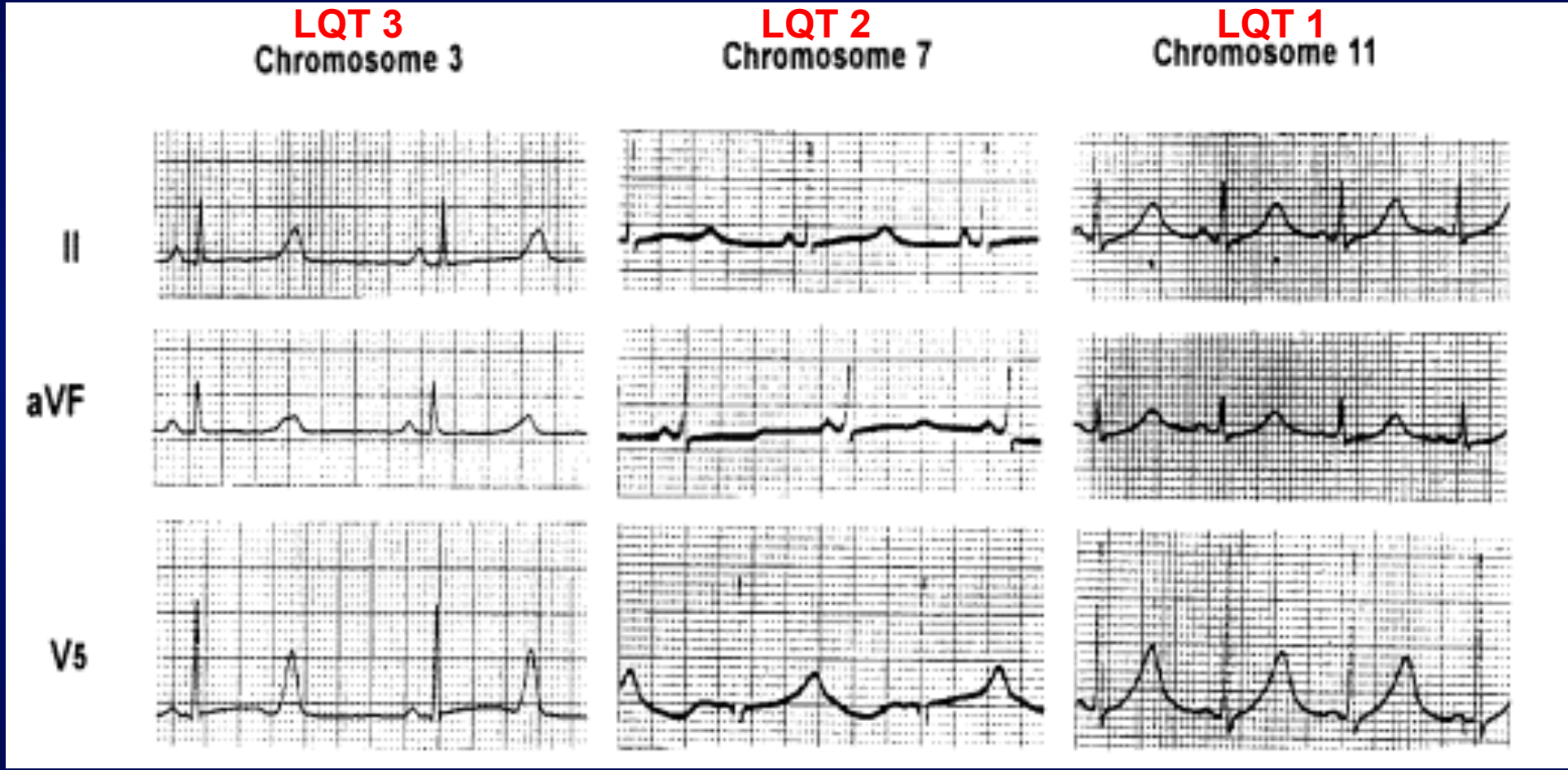
1. LQTS is diagnosed:
   a. In the presence of an LQTS risk score ≥ 3.5 in the absence of a secondary cause for QT prolongation *and/or*
   b. In the presence of an unequivocally pathogenic mutation in one of the LQTS genes *or*
   c. In the presence of a corrected QT interval for heart rate using Bazett's formula (QTc) ≥500 ms in repeated 12- lead electrocardiogram (ECG) and in the absence of a secondary cause for QT prolongation.
2. LQTS can be diagnosed in the presence of a QTc between 480 and 499 ms in repeated 12-lead ECGs in a patient with unexplained syncope in the absence of a secondary cause for QT prolongation and in the absence of a pathogenic mutation.

Table 2. 1993–2011 LQTS Diagnostic Criteria

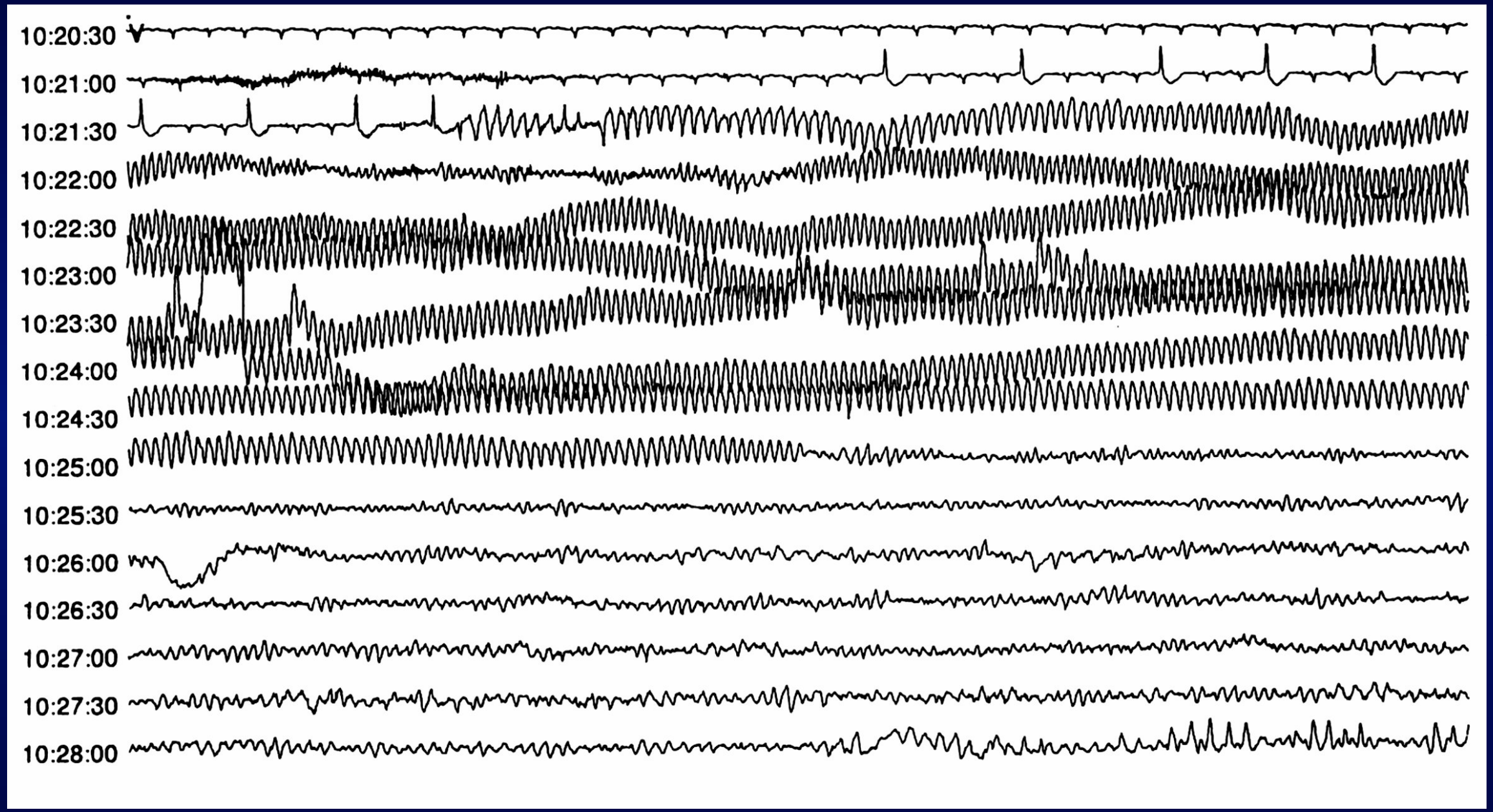
	Points
Electrocardiographic findings #	
A QTc^	
≥480 ms	3
460–479 ms	2
450–459 ms (in males)	1
B QTc^ 4th minute of recovery from exercise stress test ≥480 ms	1
C Torsade de pointes*	2
D T wave alternans	1
E Notched T wave in 3 leads	1
F Low heart rate for age@	0.5
Clinical history	
A Syncope*	
With stress	2
Without stress	1
B Congenital deafness	0.5
Family history	
A Family members with definite LQTS$	1
B Unexplained sudden cardiac death below age 30 among immediate family members$	0.5

Schwartz Circulation 2011

T wave patterns in LQT1, LQT2, and LQT3 Carriers.



Zareba et al. IJBEM 2003 Vol.5, No.1: 46-9


10:20:30
10:21:00
10:21:30
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10:22:30
10:23:00
10:23:30
10:24:00
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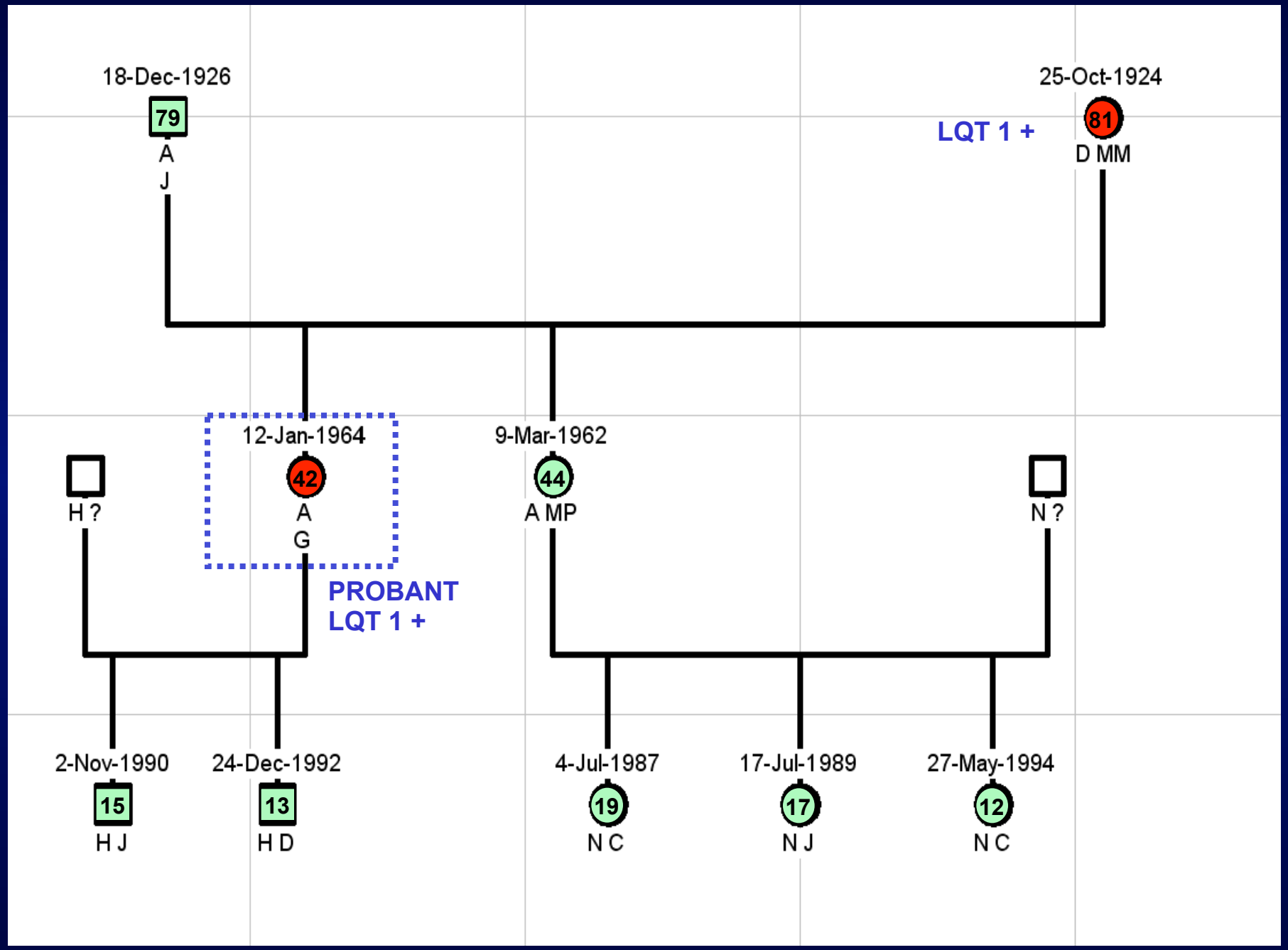
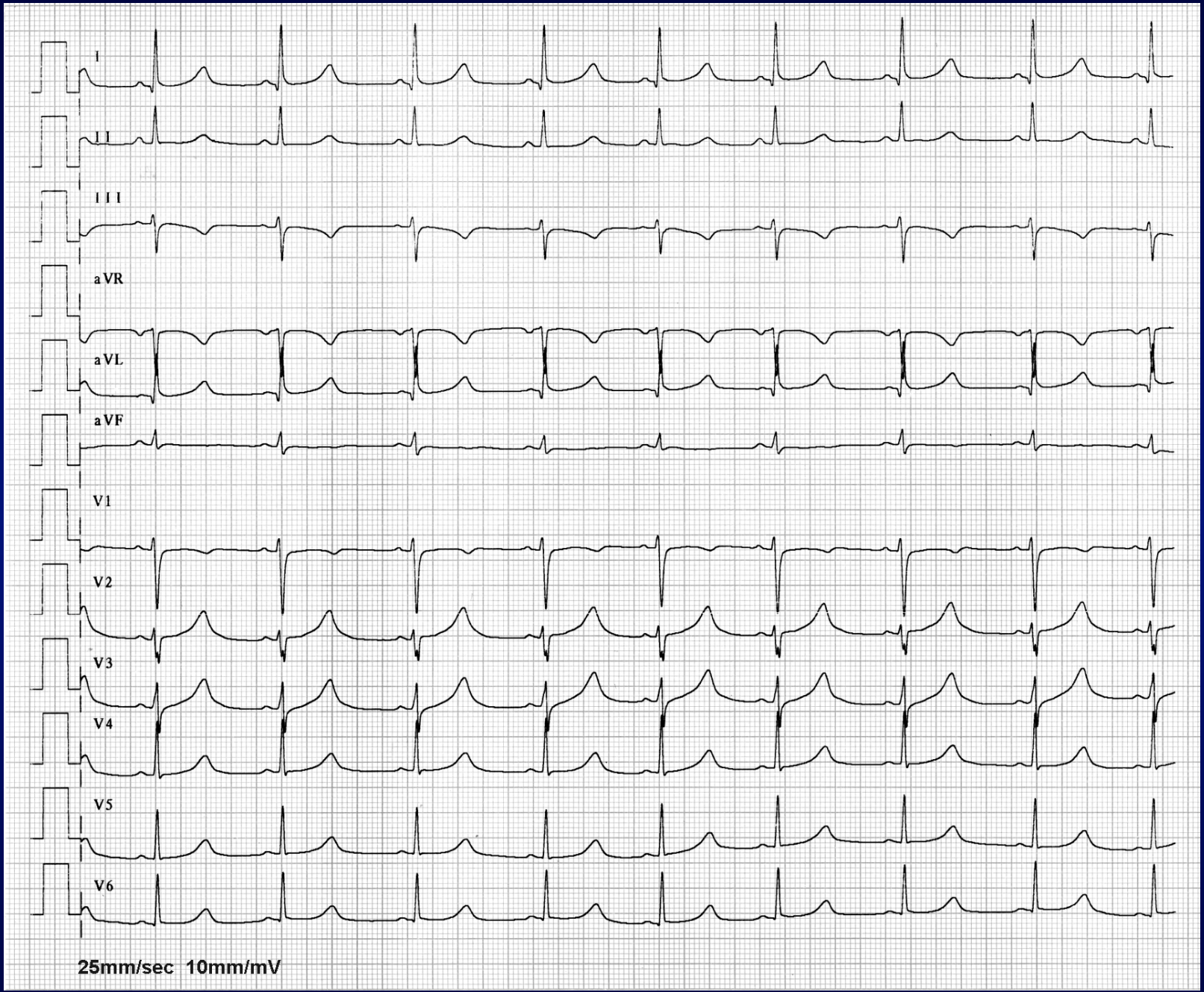
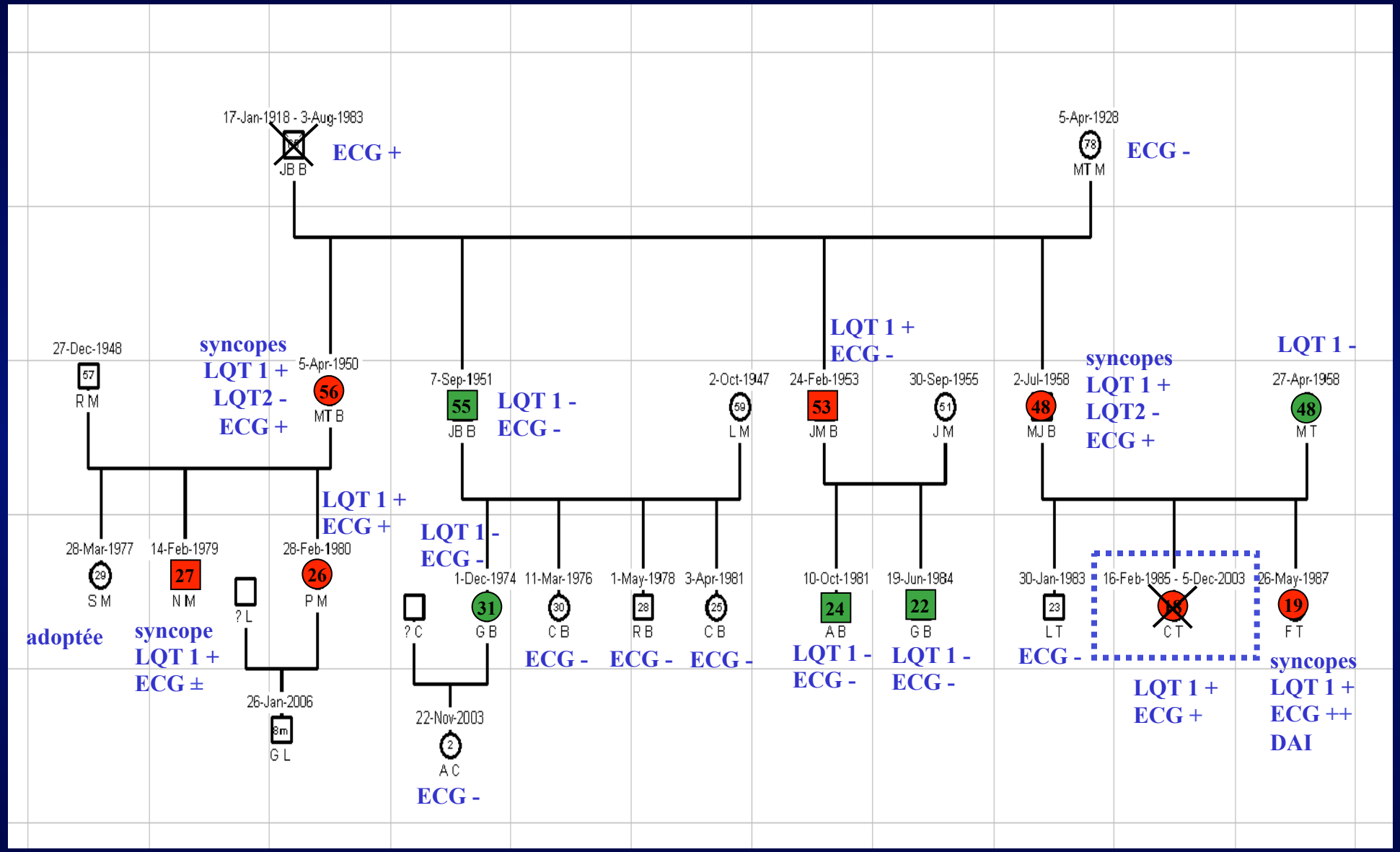
10:28:00


18-Dec-1926
79
A
J
25-Oct-1924
81
LQT 1 +
D MM
12-Jan-1964
42
A
G
9-Mar-1962
44
A MP
H ?
N ?
PROBANT
LQT 1 +
2-Nov-1990
15
H J
24-Dec-1992
13
H D
4-Jul-1987
19
N C
17-Jul-1989
17
N J
27-May-1994
12
N C


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
25mm/sec 10mm/mV


17-Jan-1918 - 3-Aug-1983
JB B
ECG +
5-Apr-1928
78
MT M
ECG -
27-Dec-1948
57
R M
syncopes
LQT 1 +
LQT2 -
ECG +
5-Apr-1950
56
MT B
7-Sep-1951
55
JB B
LQT 1 -
ECG -
2-Oct-1947
59
L M
24-Feb-1953
53
JM B
LQT 1 +
ECG -
30-Sep-1955
51
J M
2-Jul-1958
48
MJ B
syncopes
LQT 1 +
LQT2 -
ECG +
27-Apr-1958
48
M T
LQT 1 -
LQT 1 +
ECG +
28-Mar-1977
29
S M
adoptée
14-Feb-1979
27
N M
syncope
LQT 1 +
ECG ±
? L
28-Feb-1980
26
P M
26-Jan-2006
8m
G L
LQT 1 -
ECG -
1-Dec-1974
31
G B
11-Mar-1976
30
C B
ECG -
1-May-1978
28
R B
ECG -
3-Apr-1981
25
C B
ECG -
? C
22-Nov-2003
2
A C
ECG -
10-Oct-1981
24
A B
LQT 1 -
ECG -
19-Jun-1984
22
G B
LQT 1 -
ECG -
30-Jan-1983
23
L T
ECG -
16-Feb-1985 - 5-Dec-2003
C T
LQT 1 +
ECG +
26-May-1987
19
F T
syncopes
LQT 1 +
ECG ++
DAI

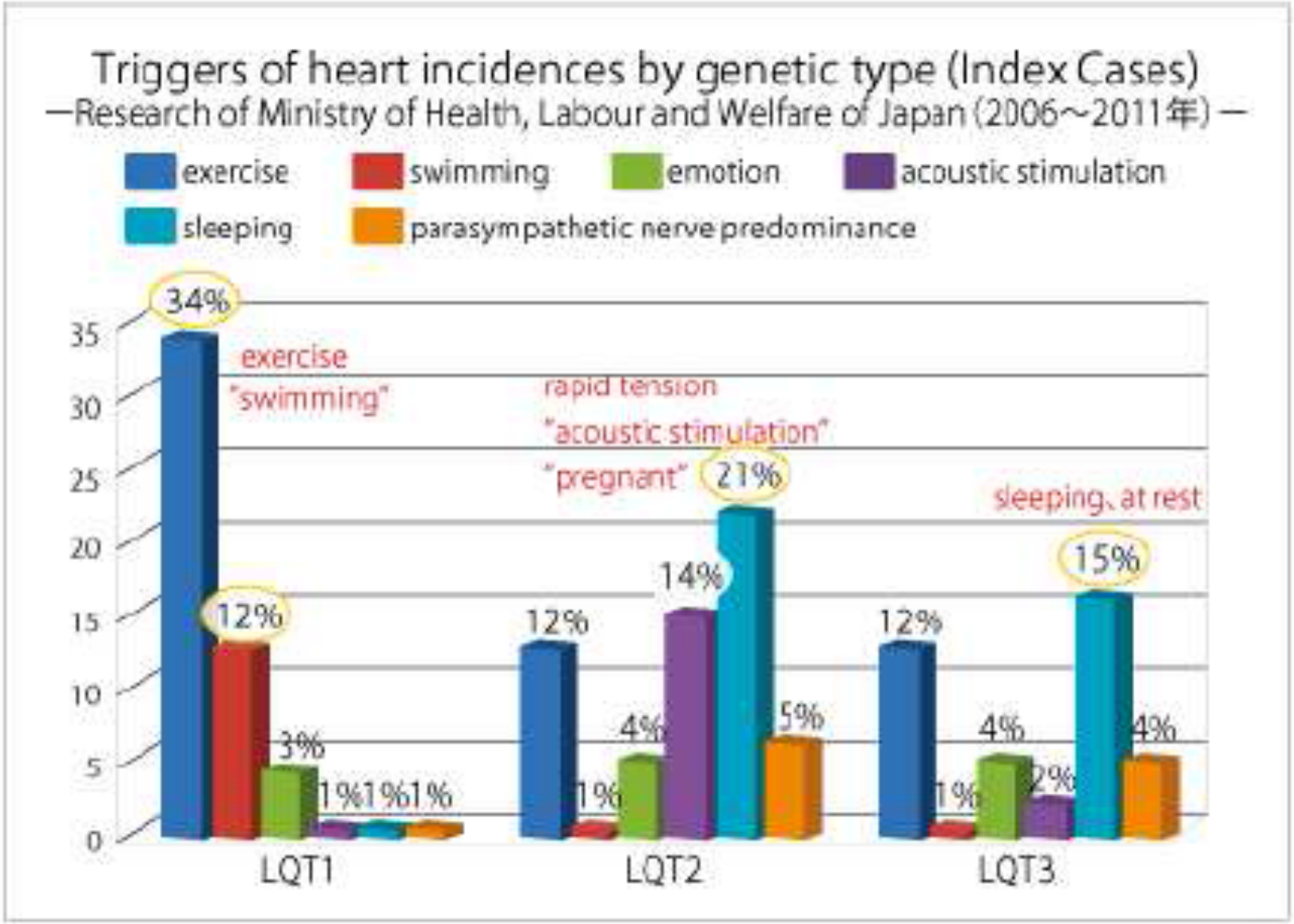


Figure 2 : Triggers for heart incidences by gene

Shimizu

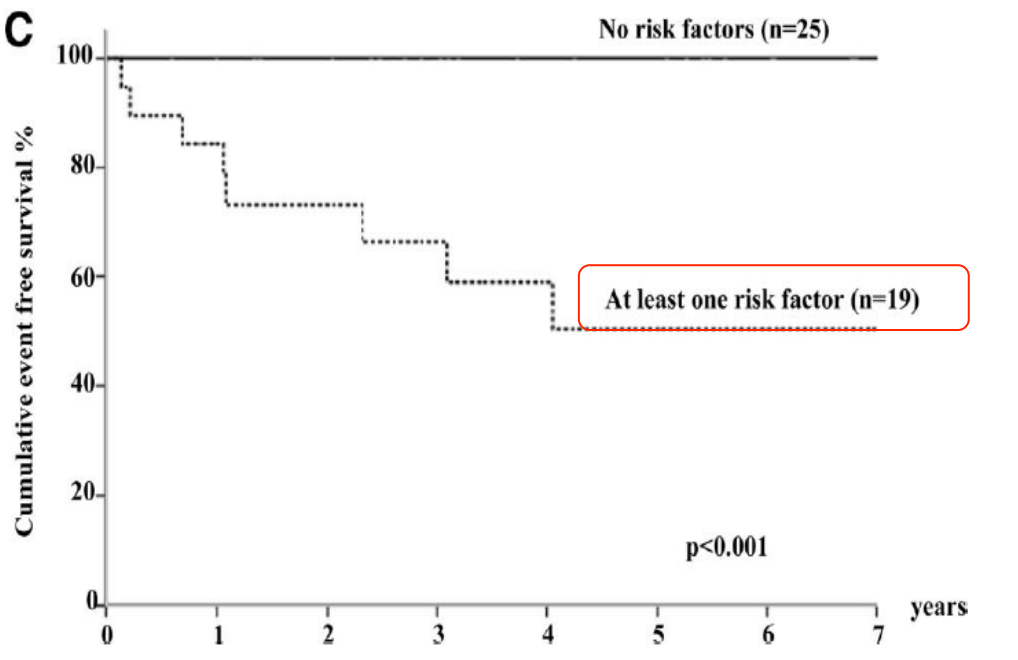
Who Are the Long-QT Syndrome Patients Who Receive an Implantable Cardioverter-Defibrillator and What Happens to Them? : Data From the European Long-QT Syndrome Implantable Cardioverter-Defibrillator (LQTS ICD) Registry

Peter J. Schwartz, Carla Spazzolini, Silvia G. Priori, Lia Crotti, Alessandro Vicentini, Maurizio Landolina, Maurizio Gasparini, Arthur A.M. Wilde, Reinoud E. Knops, Isabelle Denjoy, Lauri Toivonen, Gerold Mönnig, Majid Al-Fayyadh, Luc Jordaens, Martin Borggrefe, Christina Holmgren, Pedro Brugada, Luc De Roy, Stefan H.

Table 4. Multivariate Risk Predictors of Appropriate Shock During Follow-Up in Patients With an ICD

Clinical Variables	Hazard Ratio (95% CI)	*P*
Prior ACA	1.81 (1.09–3.0)	0.023
Events on therapy	1.81 (1.08–3.0)	0.025
Age at implantation <20 y	2.3 (1.38–3.8)	0.001
QTc*	1.41 (1.03–1.92)	0.03

*This 3-level (≤500, >500 to ≤550, and >550 milliseconds) factor was treated as a continuous variable, its linear effect having been evaluated.



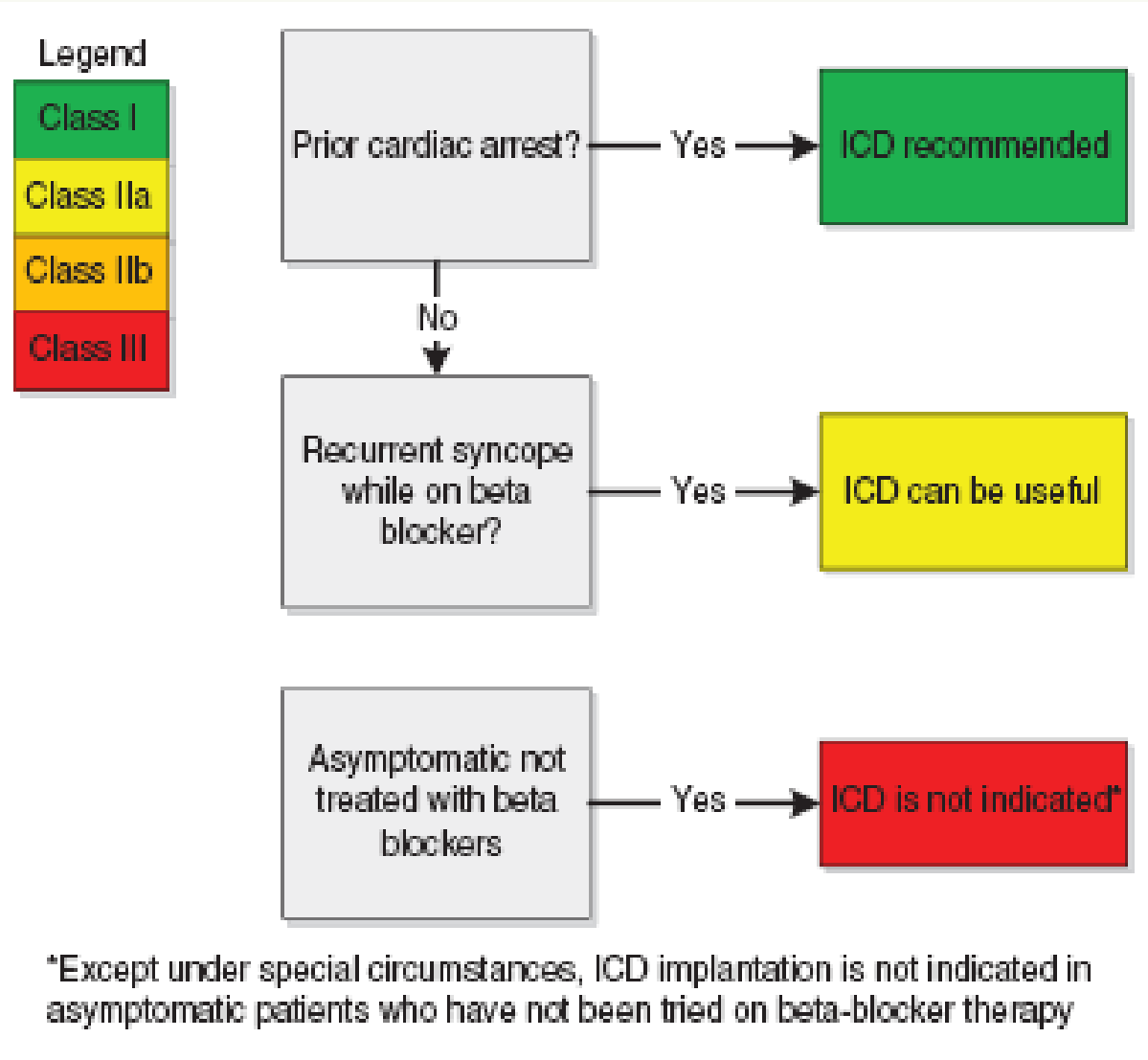
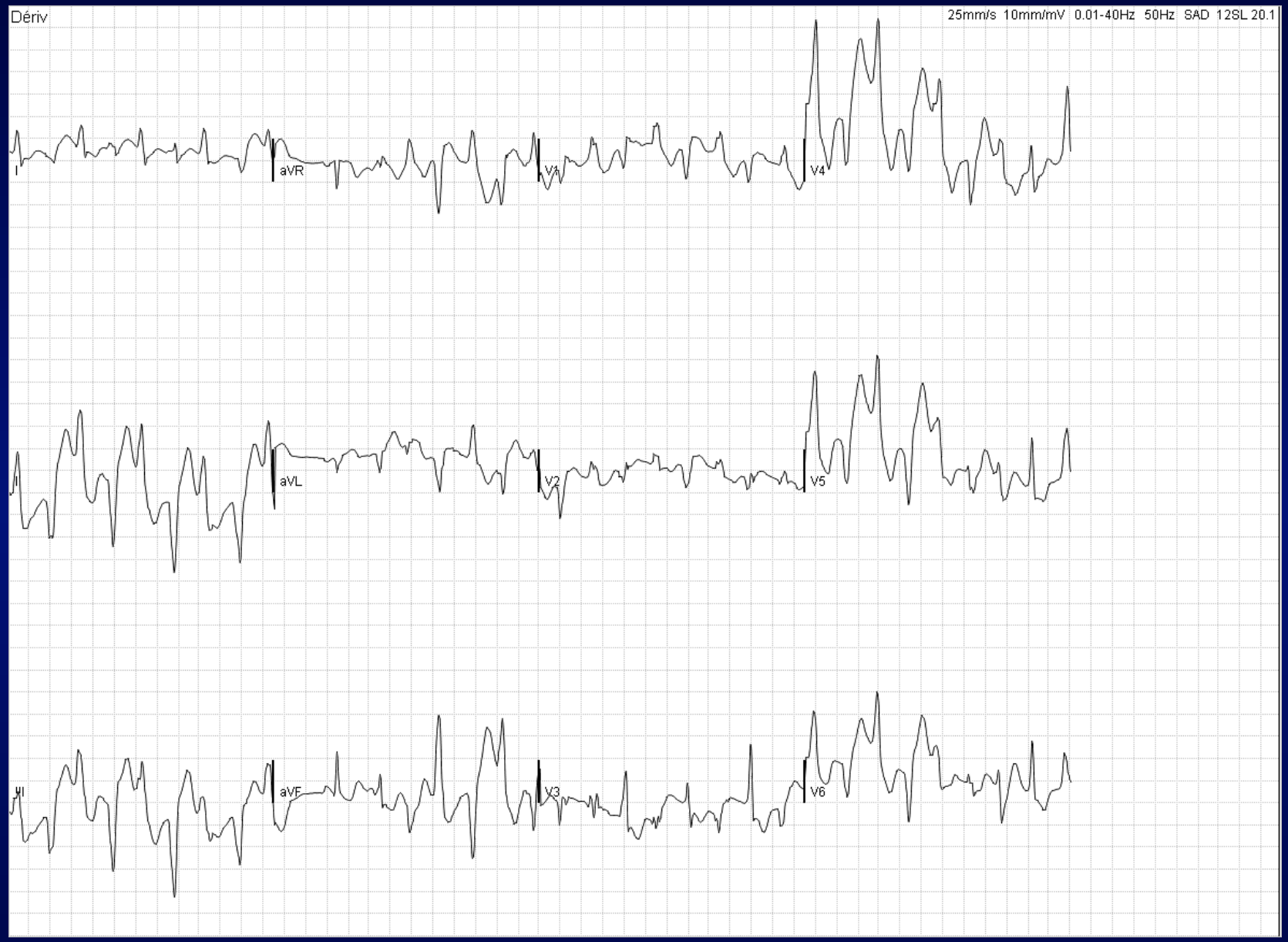


Figure 1 Consensus recommendations for ICDs in patients diagnosed with long QT syndrome.



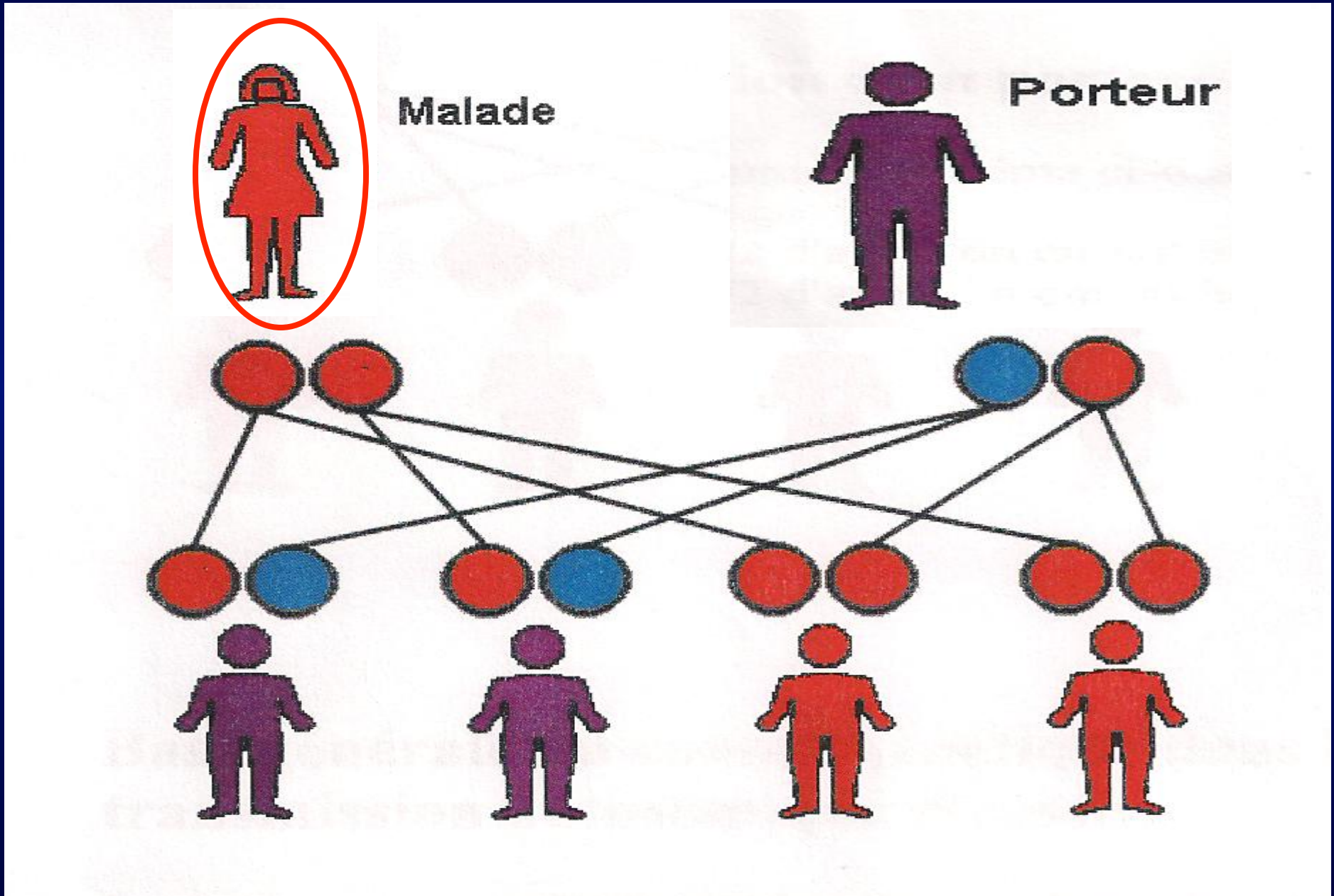
Malaise au cabinet du cardiologue

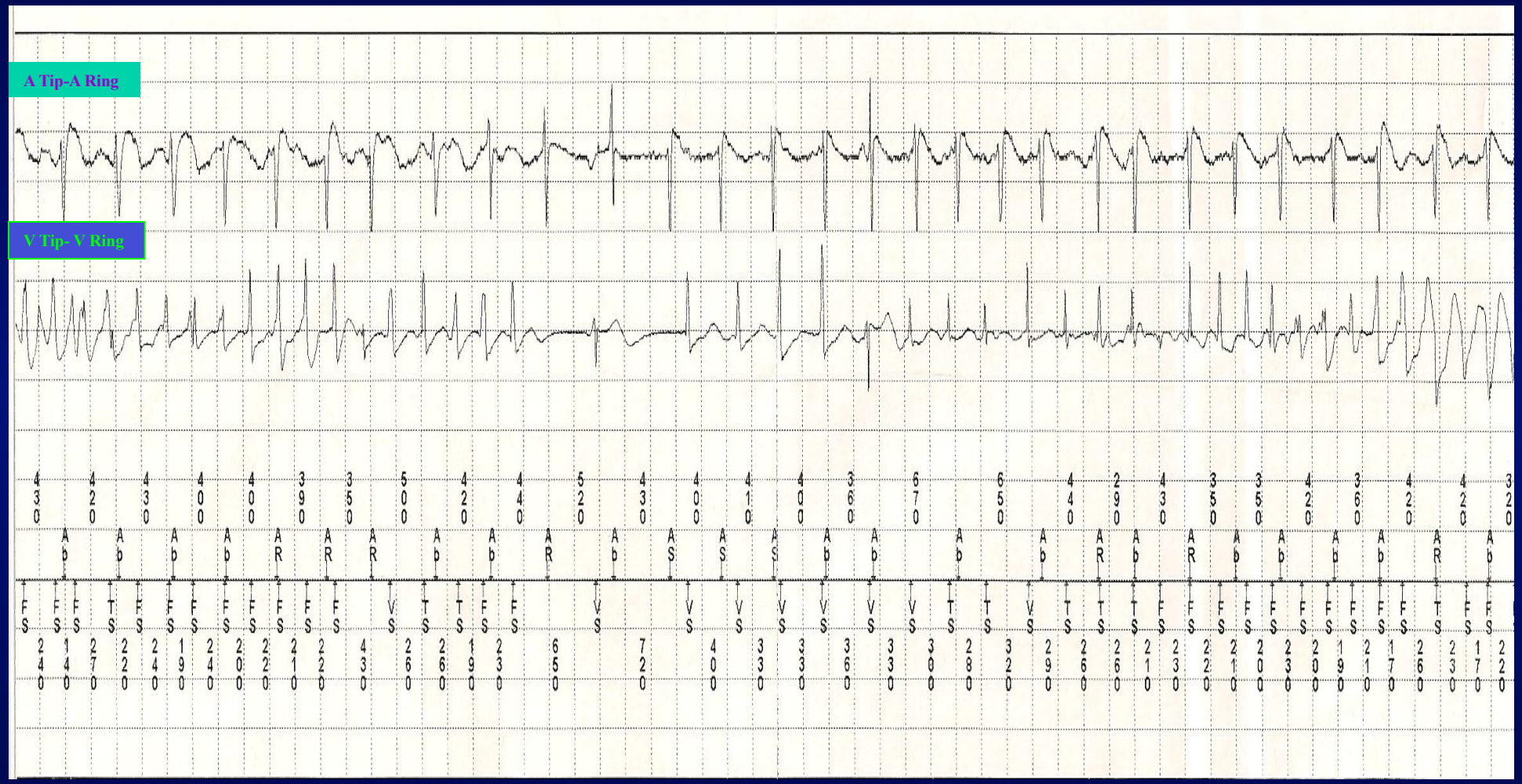
CPVT: Base génétique

- RYR2: (Ryanodine Receptors): 63 %
- CASQ2: (Calsequestrine): 8 %

NB: - Porteurs silencieux (Silent cariers)
- Phenocopies
- mutations RYR2 associés à d'autres pathologies (VD)

Pathologie héréditaire autosomique récessive




A Tip-A Ring
V Tip- V Ring

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Sudden Cardiac Arrest Associated with Early Repolarization

Michel Haïssaguerre, M.D., Nicolas Derval, M.D., Frederic Sacher, M.D., Laurence Jesel, M.D., Isabel Deisenhofer, M.D., Luc de Roy, M.D., Jean-Luc Pasquié, M.D., Ph.D., Akihiko Nogami, M.D., Dominique Babuty, M.D., Sinikka Yli-Mayry, M.D., Christian De Chillou, M.D., Patrice Scanu, M.D., Philippe Mabo, M.D., Seiichiro Matsuo, M.D., Vincent Probst, M.D., Ph.D., Solena Le Scouarnec, Ph.D., Pascal Defaye, M.D., Juerg Schlaepfer, M.D., Thomas Rostock, M.D., Dominique Lacroix, M.D., Dominique Lamaison, M.D., Thomas Lavergne, M.D., Yoshifusa Aizawa, M.D., Anders Englund, M.D., Frederic Anselme, M.D., Mark O'Neill, M.D., Meleze Hocini, M.D., Kang Teng Lim, M.B., B.S., Sebastien Knecht, M.D., George D. Veenhuyzen, M.D., Pierre Bordachar, M.D., Michel Chauvin, M.D., Pierre Jais, M.D., Gaelle Coureau, Ph.D., Genevieve Chene, Ph.D., George J. Klein, M.D., and Jacques Clémenty, M.D.

6. Early Repolarization (ER)

Expert Consensus Recommendations on ***ER Diagnosis***

1. ER ***syndrome is diagnosed*** in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG in a patient resuscitated from otherwise unexplained VF/polymorphic VT.
2. ER syndrome ***can be diagnosed*** in a SCD victim with a negative autopsy and medical chart review, with a previous ECG demonstrating J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.
3. ER pattern ***can be diagnosed*** in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.

ER

Prévalence: 1-13% population générale

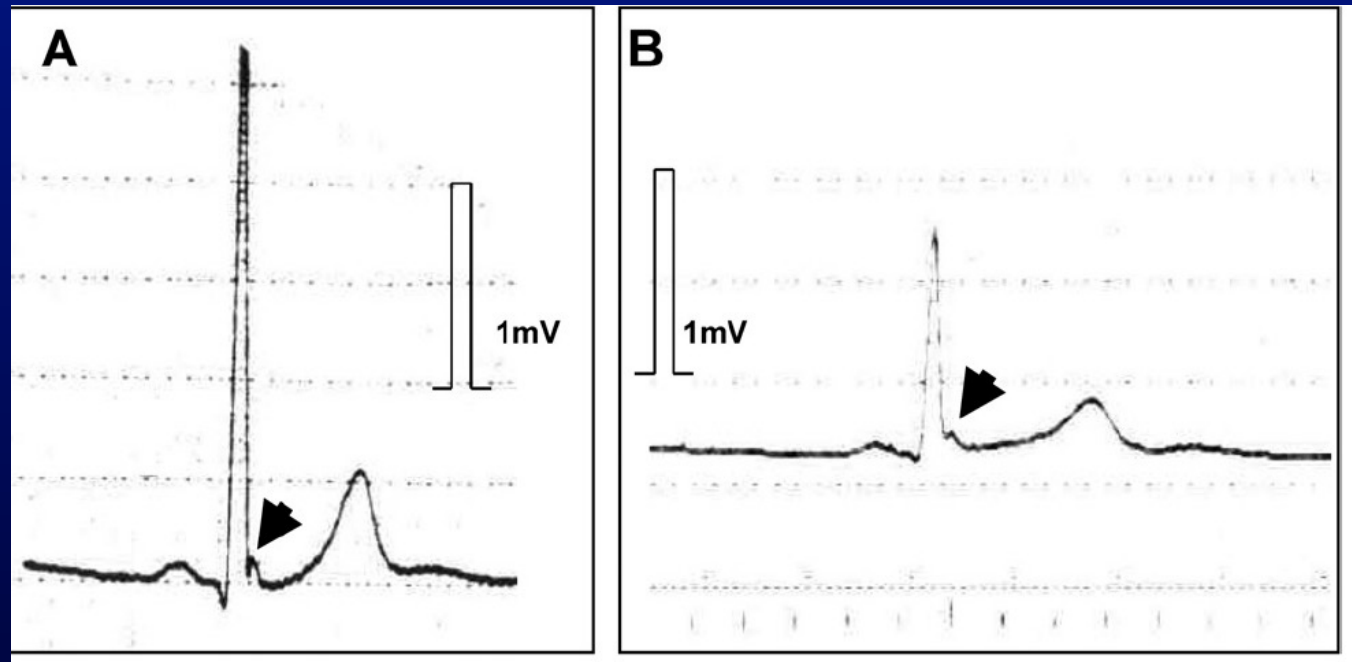
15-70% en cas de FV Idiopathique

>70 % hommes

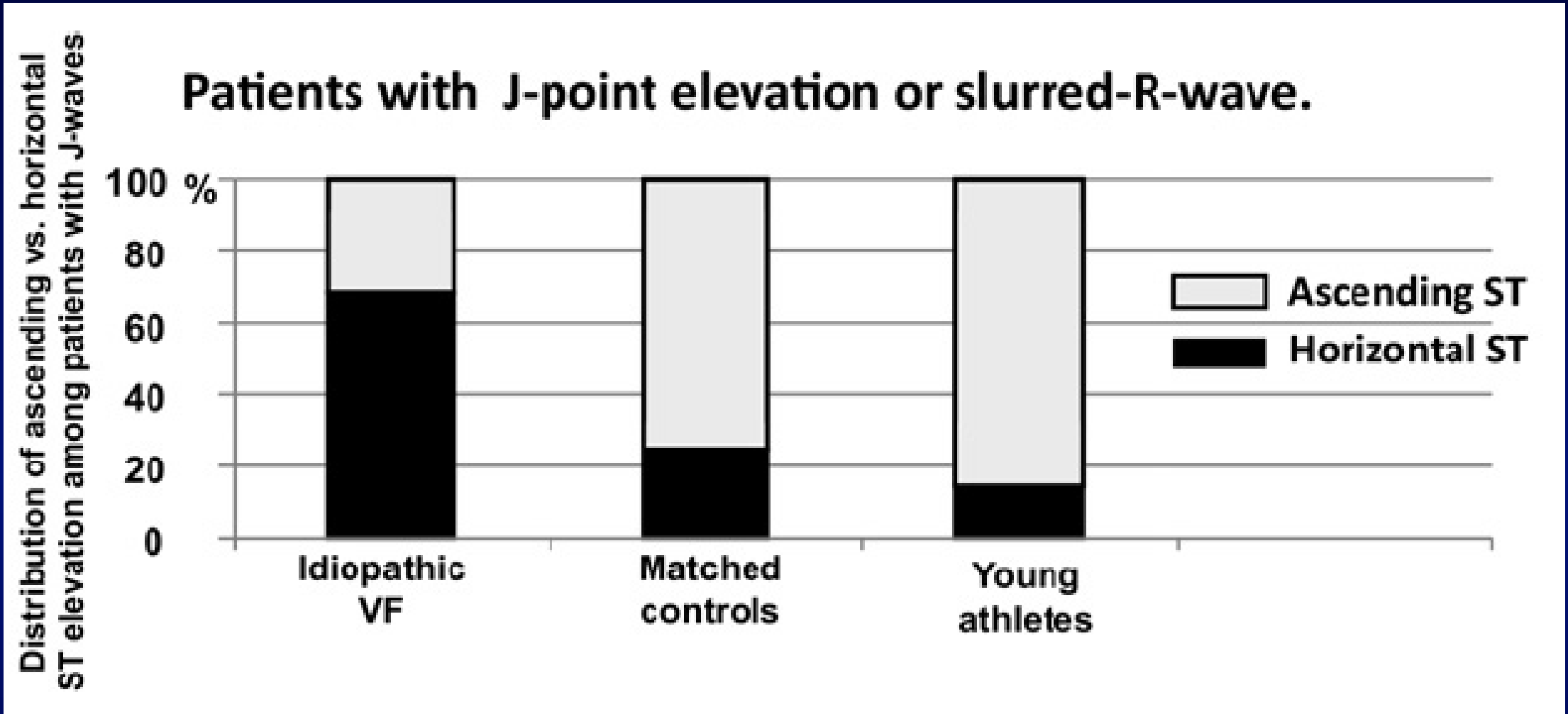
Plus fréquent enfants /afro-americains/ SE asiatique

Risk stratification

1. point J ≥ 0,2 mV
2. ST horizontal ou descendant
3. Etendue plus grande
4. Point J plus marqué en amplitude



Rapidly ascending (**A**) and horizontal (**B**) ST segment in the leads deploying J waves (J waves marked with arrowhead


Patients with J-point elevation or slurred-R-wave.
Distribution of ascending vs. horizontal ST elevation among patients with J-waves
100 %
80
60
40
20
0
Idiopathic VF
Matched controls
Young athletes
Ascending ST
Horizontal ST

ROSSO 2012

Expert Consensus Recommendations on ***ER Therapeutic Interventions***

Class I	1.	ICD implantation ***is recommended*** in patients with a diagnosis of ER syndrome who have survived a cardiac arrest.
Class IIa	2.	Isoproterenol infusion ***can be useful*** in suppressing electrical storms in patients with a diagnosis of ER syndrome.
	3.	Quinidine in addition to an ICD ***can be useful*** for secondary prevention of VF in patients with a diagnosis of ER syndrome.
Class IIb	4.	ICD implantation ***may be considered*** in symptomatic family members of ER syndrome patients with a history of syncope in the presence of ST-segment elevation > 1 mm in two or more inferior or lateral leads.
	5.	ICD implantation ***may be considered*** in asymptomatic individuals who demonstrate a high-risk ER ECG pattern (high J-wave amplitude, horizontal/descending ST-segment) in the presence of a strong family history of juvenile unexplained sudden death with or without a pathogenic mutation
Class III	6.	ICD implantation ***is not recommended*** in asymptomatic patients with an isolated ER ECG pattern.

European Heart Journal (2010) **31**, 243–259
doi:10.1093/eurheartj/ehp473

ESC REPORT

Recommendations for interpretation of 12-lead electrocardiogram in the athlete

Table 1 Classification of abnormalities of the athlete's electrocardiogram

Group 1: common and training-related ECG changes	Group 2: uncommon and training-unrelated ECG changes
Sinus bradycardia	T-wave inversion
First-degree AV block	ST-segment depression
Incomplete RBBB	Pathological Q-waves
Early repolarization	Left atrial enlargement
Isolated QRS voltage criteria for left ventricular hypertrophy	Left-axis deviation/left anterior hemiblock
	Right-axis deviation/left posterior hemiblock
	Right ventricular hypertrophy
	Ventricular pre-excitation
	Complete LBBB or RBBB
	Long- or short-QT interval
	Brugada-like early repolarization

RBBB, right bundle branch block; LBBB, left bundle branch block.

Meta-analyse 9 études de Repolarisation précoce

Table 1 Characteristics of the Included Studies

First Author (Year) (Ref. #)	Location	Men (%)	Mean Age (yrs)	Follow-Up (yrs)	Ratio of ERP (%)	Number of Events	Total Number of Subjects/ Person-Years	End Point
Cohort studies								
Sinner et al. (2010) (9)	Germany	48.9	52 ± 10	18.9	13.1	2,251	6,213/323,076	Cardiac death, all-cause death
Tikkanen et al. (2009) (8)	Finland	52.4	44 ± 8	30 ± 11	5.8	8,897	10,864/325,920	Cardiac death, all-cause death, arrhythmia death
Haruta et al. (2011) (7)	Japan	43.7	47 ± 15	24 ± 15	23.9	3,515	5,976/143,424	All-cause death, cardiac death, arrhythmia death
Olson et al. (2011) (11)	United States	44.3	54	17 ± 4	12.3	3,555	15,141/257,397	Arrhythmia death, all-cause death,
Uberoi et al. (2011) (6)	United States	87	55 ± 15	7.6 ± 3.8	3.2	1,995	29,281/222,536	Cardiac death
Klatsky et al. (2003) (5)	United States	44	42.5	14	0.9	11,075	73,088/102,3232	All-cause death, cardiac death
Rollin et al. (2012) (12)	France	1.6	49.8 ± 8.6	14.2 ± 2.0	13.2	77	157/16,254	All-cause death, cardiac death
Case-control studies								
Haissaguerre et al. (2008) (1)	Multinational	63.6	36 ± 12	5.1 ± 4.2	31.0	796	206/1,047	Recurrent VF
Rosso et al. (2008) (4)	United States	71	38	3	31.1	45	169/NA	NA

Wu SH 2012 JACC

Table 3 Incidence Rates and Risks for Arrhythmia Death, Cardiac Death, and All-Cause Death in Subjects With ERP During Follow-Up

	Events per Person-Years			
	ERP Positive	ERP Negative	RR (95% CI)	p Value
Primary end point				
All-cause death	1,899/111,331	20,503/1,771,583	1.06 (0.85–1.31)	0.62
Cardiac death	387/80,388	9,570/1,747,749	0.78 (0.27–2.21)	0.63
Arrhythmia death	142/84,346	966/640,004	1.70 (1.19–2.42)	0.003
Subgroup study: J-point elevation				
≥0.1 mV in inferior leads	90/22,170	—	1.72 (1.39–2.13)	<0.00001
≥0.2 mV in inferior leads	16/2,196	—	3.06 (1.87–4.99)	<0.00001
≥0.1 mV in lateral leads	31/16,106	—	0.8 (0.56–1.14)	0.22
Configuration				
Notching	38/25,852	—	1.54 (1.11–2.15)	0.01
Slurring	31/12,483	—	1.36 (0.95–1.94)	0.09

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Prevalence and Characteristics of Early Repolarization in the CASPER Registry

Cardiac Arrest Survivors With Preserved Ejection Fraction Registry

Nicolas Derval, MD,* Christopher S. Simpson, MD,# David H. Birnie, MD,¶ Jeffrey S. Healey, MD,‡

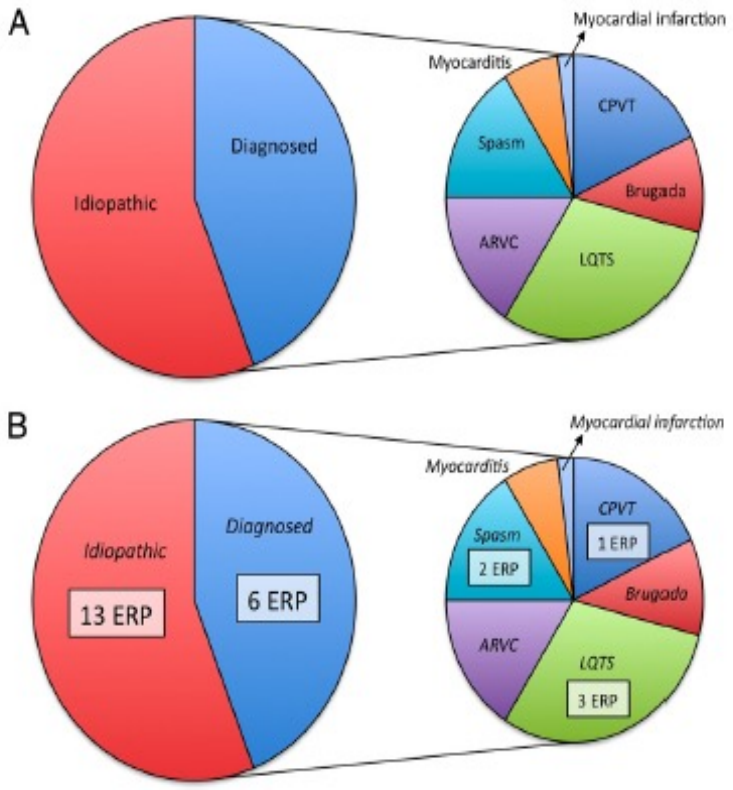


Figure 3 Distribution of Diagnoses and Early Repolarization in the Patient Cohort

(A) Diagnostic yield in 100 patients with unexplained cardiac arrest. The proportion of subclinical primary electrical disease is depicted in the pie chart to the right. (B) Distribution of ERP in IVF patients (n = 56) and in diagnosed VF patients (n = 44). ARVC = arrhythmogenic right ventricular cardiomyopathy; CPVT = catecholaminergic polymorphic ventricular tachycardia; LQTS = long QT syndrome; Spasm = coronary spasm; VF = ventricular tachycardia; other abbreviations as in Figure 1.

LE SPORT

LES SPORTIFS

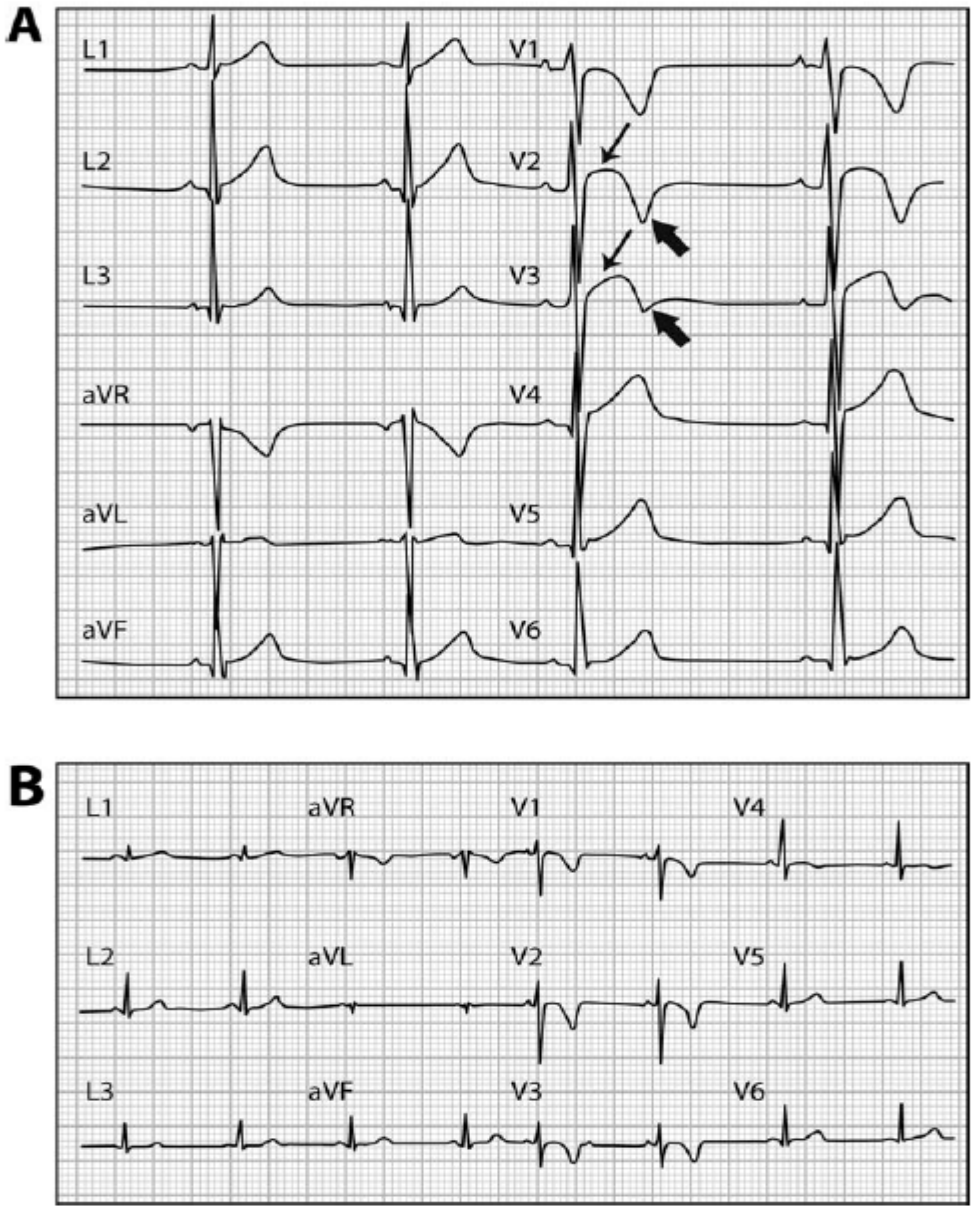


Figure 3 (*A*) Early repolarization pattern in a healthy black athlete characterized by right precordial T-wave inversion (arrowhead) preceded by ST-segment elevation (arrow). (*B*) Right precordial T-wave inversion in a patient with ARVC. Note that unlike early repolarization, in the ARVC the right precordial leads do not demonstrate any elevation of the ST-segment.

Athletes

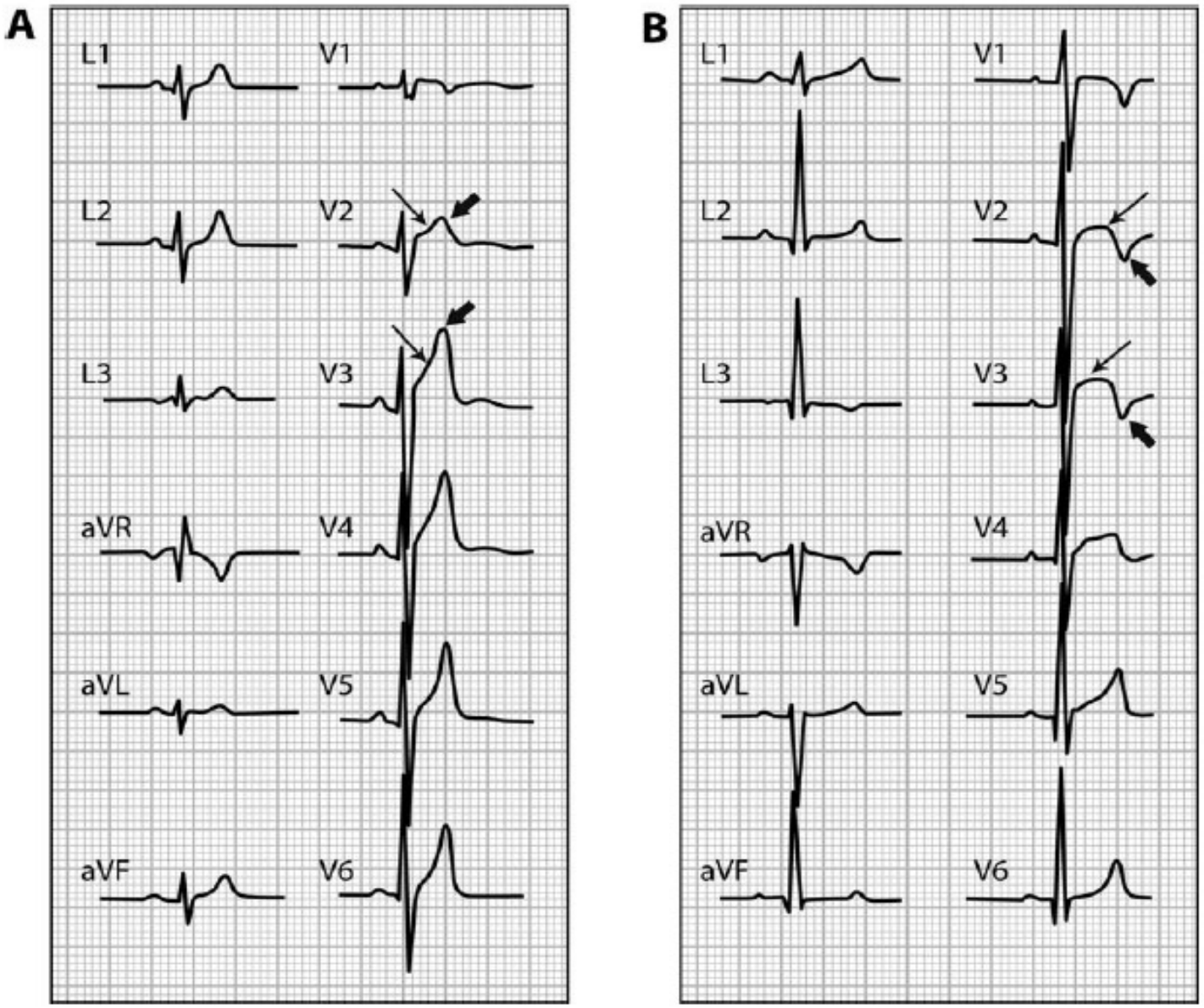


Figure 2 Different patterns of precordial early repolarization in two healthy athletes. (*A*) ST-segment elevation with upward concavity (arrows), followed by a positive T-wave (arrowheads). (*B*) ST-segment elevation with upward convexity (arrows), followed by a negative T-wave (arrowheads).

Benign clinical significance of J-wave pattern (early repolarization) in highly trained athletes

Filippo M. Quattrini, MD, PhD,* Antonio Pelliccia, MD,* Riccardo Assorgi, MD,* Fernando M. DiPaolo, MD,* Maria Rosaria Squeo, MD,* Franco Culasso, PhD,† Vincenzo Castelli, MD,‡ Mark S. Link, MD,§ Barry J. Maron, MD¶

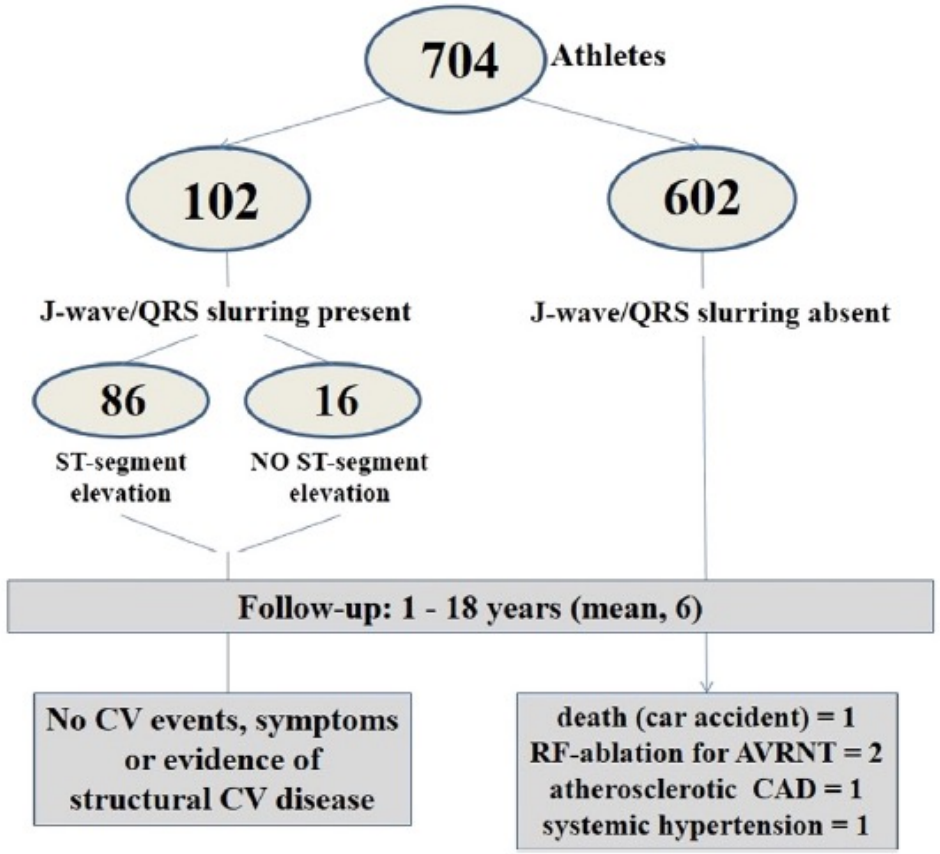
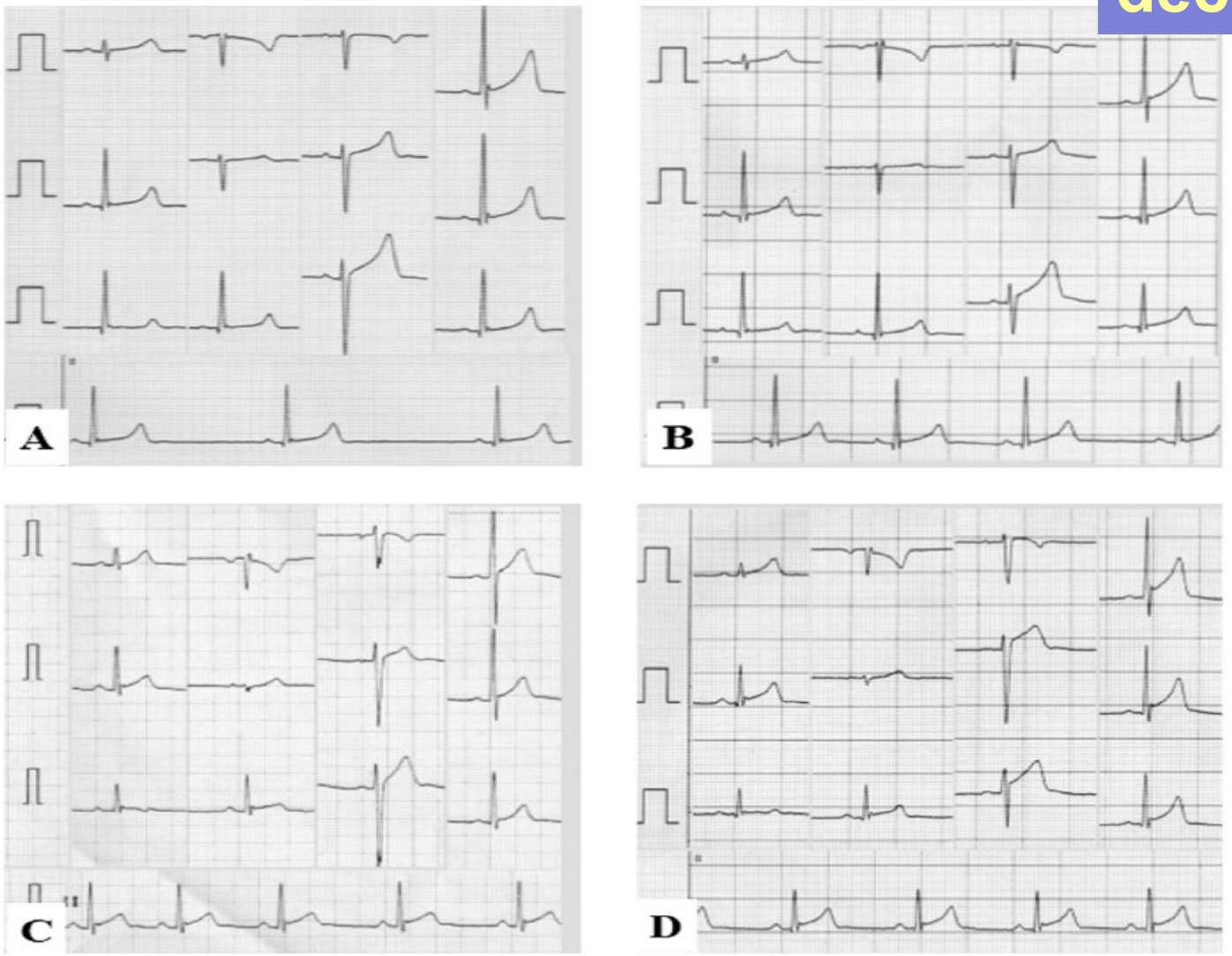


Figure 3 Flow chart showing the distribution of the athlete population according to the ECG presence of J wave/QRS slurring and/or ST-segment elevation, and clinical outcome over a 5-year follow-up period. AVNRT = atrioventricular reentrant tachycardia; CAD = coronary artery disease; CV = cardiovascular; RF = radiofrequency.

déconditionnement



reprise

Figure 2 Twelve-lead ECGs showing dynamic changes of J wave in athletes according to intensity of training. A 27-year-old middle-distance runner at the time of peak training **(A)** and after detraining **(B)**. A 20-year old rower at the time of low-intensity training **(C)** and at peak training **(D)**. **A:** Peak training. The J wave/QRS slurring pattern is evident in leads II, III, aVF, and V_4–V_6. Maximum J-wave amplitude is 0.2 mV in lead V_4. Note the low heart rate (37 bpm), increased R/S-wave voltages in precordial leads (maximum Sokolow-Lyon score = 39 mm), and ST-segment elevation in leads V_2–V_4. **B:** Low-intensity training. In the same athlete, the J wave became less evident in leads V_4–V_6 , and the amplitude reduced to 0.1 mV. Note the increased heart rate (55 bpm) and reduced R/S-wave voltages (maximum Sokolow-Lyon score = 35 mm). **C:** Low-intensity training. Only a small J-point deflection is evident in leads II, III, and aVF. The heart rate is 68 bpm. The Sokolow-Lyon score is 37 mm. **D:** Peak training. The J wave became evident in leads V_4–V_6, and the amplitude exceeds 0.4 mV (in lead V_4). Note the reduced heart rate (60 bpm) and increased ST-segment elevation in leads V_2–V_6 and II, III, and aVF. The Sokolow-Lyon score is 43 mm.

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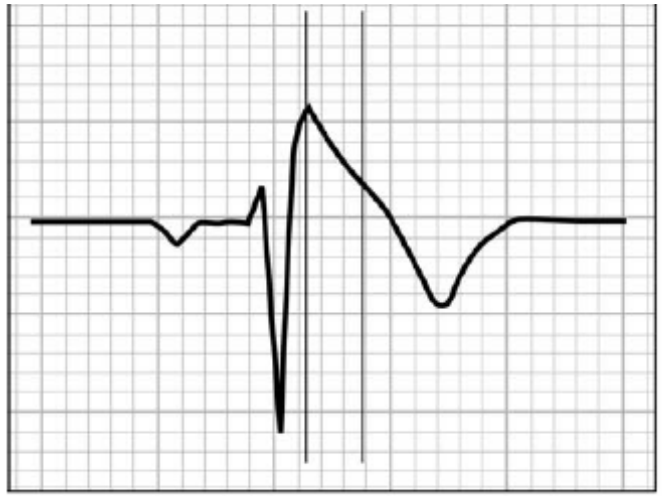
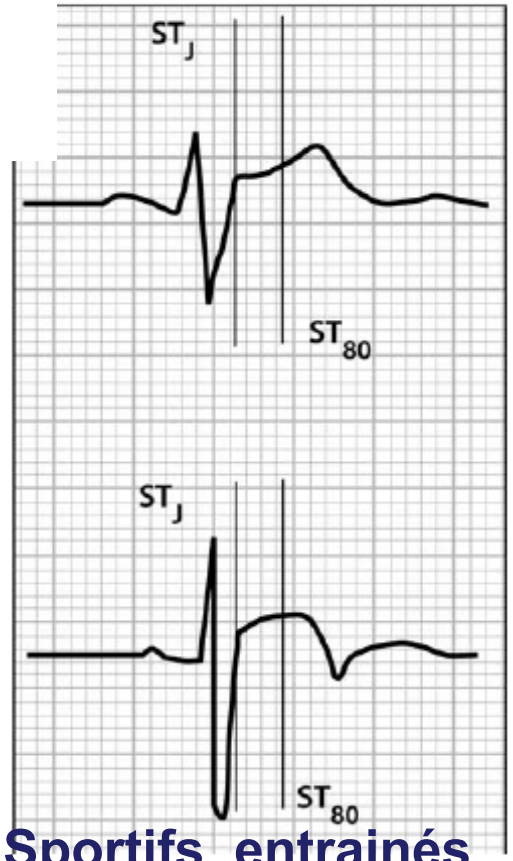
ESC REPORT

Recommendations for interpretation of 12-lead electrocardiogram in the athlete



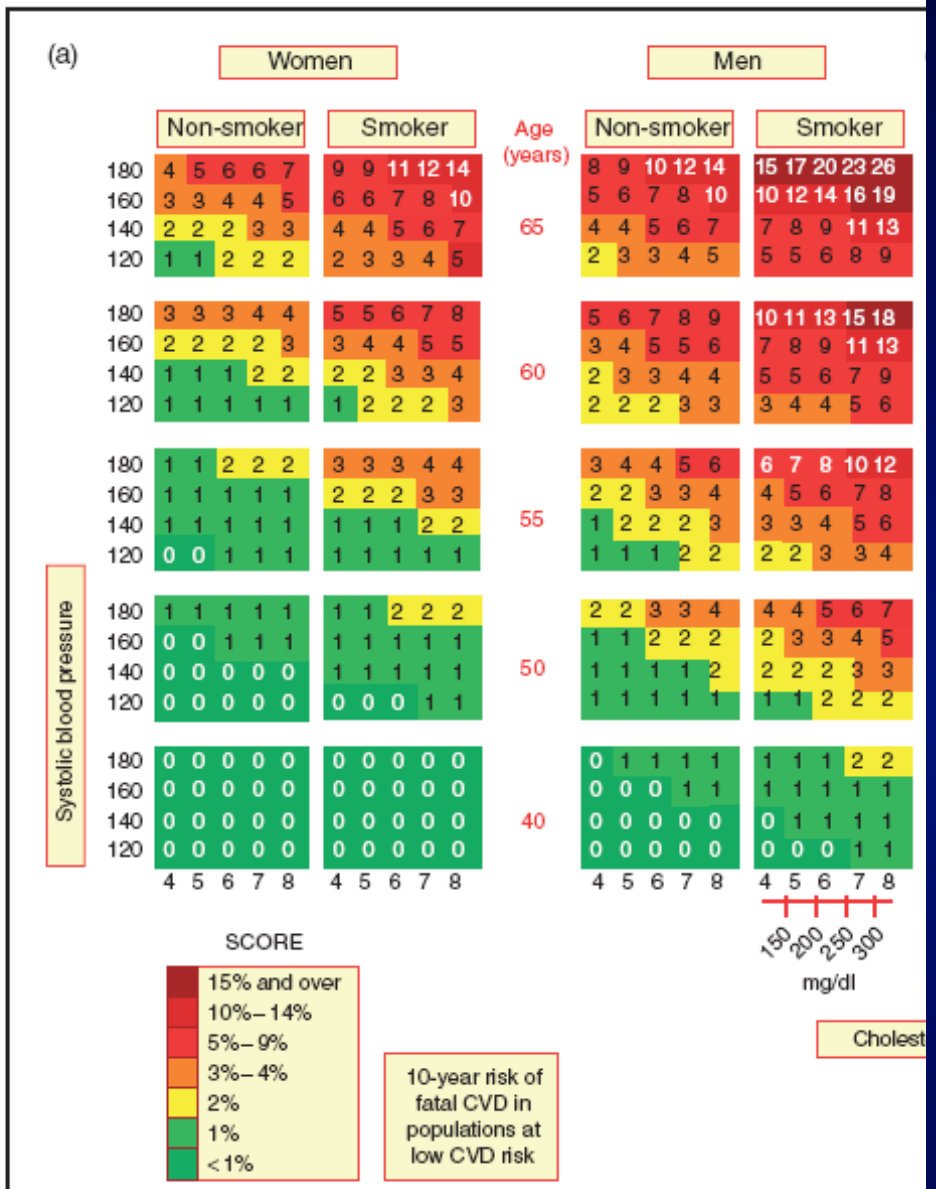
Brugada



Sportifs entrainés

Figure 8 Differential diagnosis between representative right precordial ECG patterns from (*A*) a Brugada patient and (*B*) two trained athletes. Vertical lines mark the J-point (ST_J) and the point 80 ms after the J-point (ST_{80}) where the amplitudes of ST-segment elevation are calculated. 'Coved' type ST-segment elevation in the patient with Brugada syndrome is characterized by a 'downsloping' elevated ST-segment with a ST_J/ST_{80} ratio of 1.9. Right precordial early repolarization patterns in both athletes show an 'upsloping' ST-segment elevation with ST_J/ST_{80} ratio <1; 0.7 for the 'concave' toward the top (*B*, top) and 0.68 for the 'convex' toward the top (*B*, bottom) ST-segment elevation. Modified from Corrado *et al.*[62]

Fin Provisoire



SCORE risk in low (a) or high-risk (b) countries. CVD, cardiovascular di

Figure 1. SCORE risk in low (panel a) or high-risk (panel b) coun

LOW RISK

< 5 % 10 YEAR RISK

NO Diabetes

No family history for CAD

BMI < 28

Activité physique	MET
Activités physiques d'intensité légère	**< 3**
Dormir	0,9
Regarder la télévision	1,0
Écrire à la main ou à l'ordinateur	1,8
Marche à 2,7 km/h, sans pente	2,3
Marche à 4 km/h	2,9
Activités physiques d'intensité modérée	**3 à 6**
Vélo stationnaire, 50 W, effort très léger	3,0
Marche à 4,8 km/h	3,3
Exercices à la maison (général), effort léger ou modéré	3,5
Marche à 5,4 km/h	3,6
Vélo de plaisance, <16 km/h	4,0
Vélo stationnaire, 100 W, effort léger	5,5
Activités physiques intenses	**> 6**
Course à pied, général	7
Pompes, redressements assis, effort élevé	8
Course à pied, sur place	8
Saut à la corde	10
Course à pied, >17,5 km/h	18

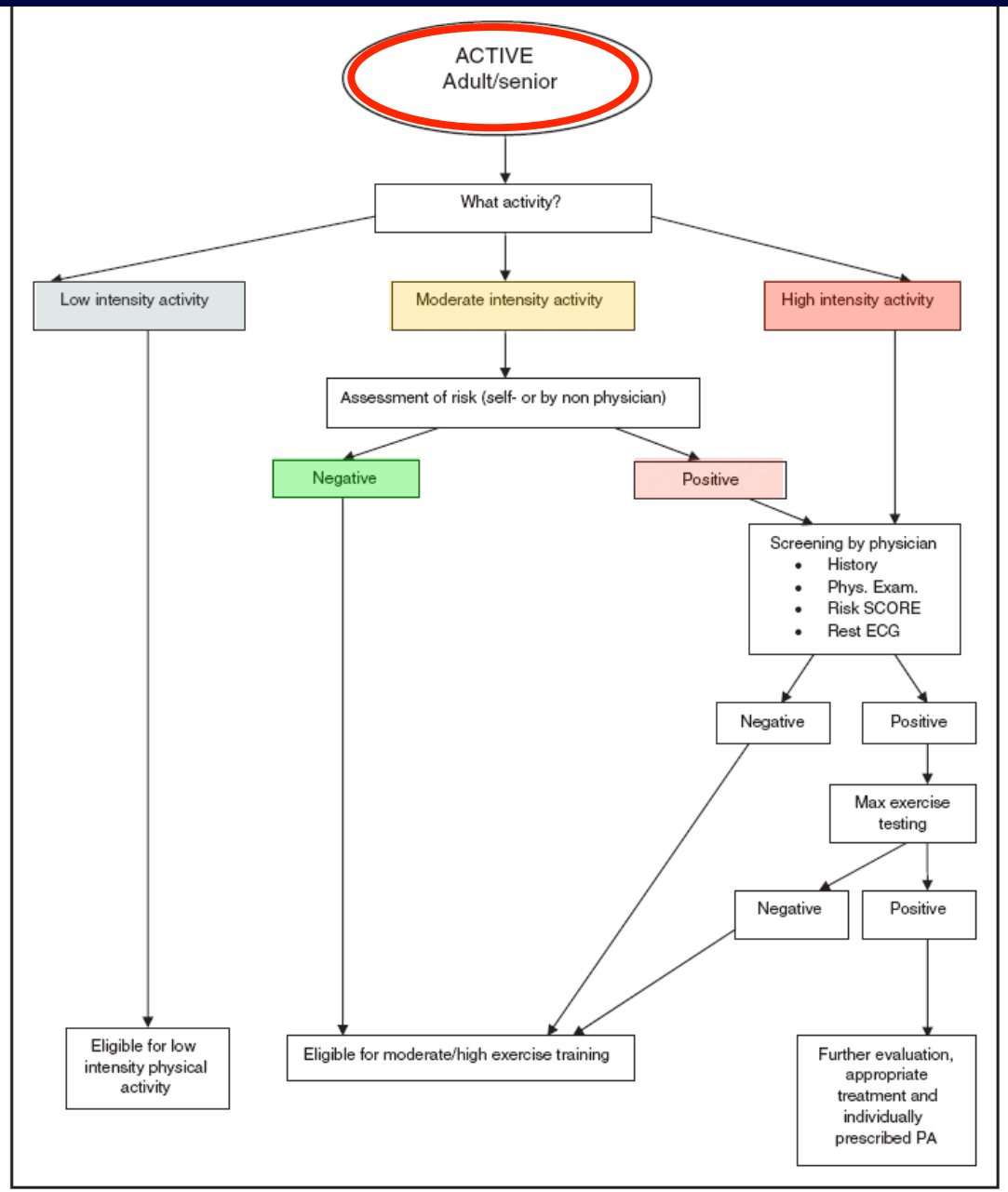


Figure 3. Proposed pre-participation evaluation protocol for asymptomatic active adult/senior individuals. PA, physical activity.

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EUROPEAN SOCIETY OF CARDIOLOGY®

ACC/AHA/ESC Guidelines

ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

13.1. Athletes

Recommendations
Class I

(1) Preparticipation history and physical examination, including family history of premature or SCD and specific evidence of cardiovascular diseases such as cardiomyopathies and ion channel abnormalities, is recommended in athletes. *(Level of Evidence: C)*