VENICE 2015 ARRHYTHMIAS

Genetic arrhythmic syndromes

mechanisms and indications for a defibrillator and/or ablation

Venice, oct 17th 2015

October 16 - 18
14th EDITION 2015

NO CONFLICT OF INTEREST TO DECLARE

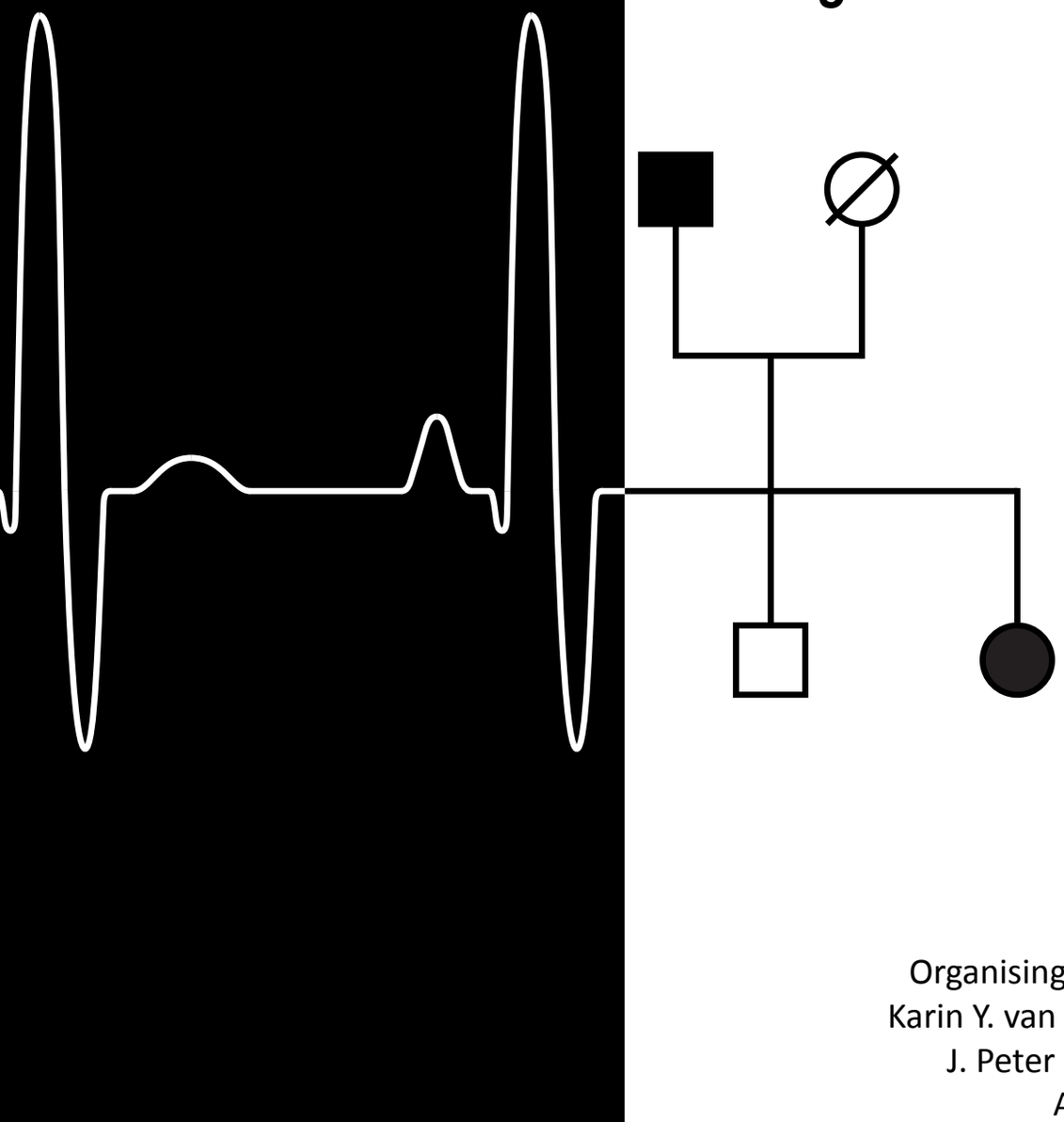
20 years

cardiogenetics in the Netherlands

Amsterdam, the Netherlands

December 4th 2015

For information and registration see www.20yrsCG.nl



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Organising committee:
Karin Y. van Spaendonck
J. Peter van Tintelen
Arthur Wilde

Primary arrhythmia syndromes (2015)

- Long QT syndrome(s)
- Short QT syndrome
- Brugada syndrome
- Catecholamine-induced PMVT/VF
- Short-coupled Torsades de Pointes
- Isolated conduction disorders (AVN, BB)
- Early repolarization syndrome
- Sinus node disease, atrial standstill
- Idiopathic ventricular fibrillation

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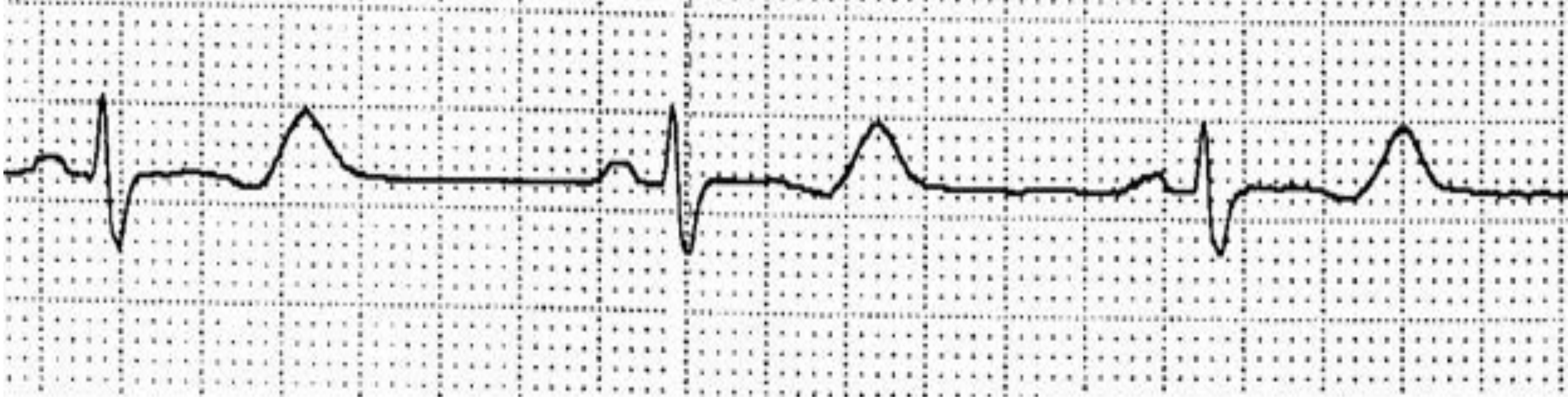
Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Europace 2013, Heart Rhythm 2013, J of Arrhyth 2013

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], Minoru Horie, (APHRS Chairperson)[3], Yongkeun Cho, (APHRS Chairperson)[4], Elijah R. Behr[5], Charles Berul[6], Nico Blom[7*], Josep Brugada[8], Chern-En Chiang[9], Heikki Huikuri[10], Prince Kannankeril[11‡], Andrew Krahn[12], Antoine Leenhardt[13], Arthur Moss[14], Peter J. Schwartz[15], Wataru Shimizu[16], Gordon Tomaselli[17†], Cynthia Tracy[%18]

Document Reviewers: Michael Ackerman (USA), Bernard Belhassen (Israel), N. A. Mark Estes III (USA), Diane Fatkin (Australia), Jonathan Kalman (Australia), Elizabeth Kaufman (USA), Paulus Kirchhof (UK and Germany), Eric Schulze-Bahr (Germany), Christian Wolpert (Germany), Jitendra Vohra (Australia), Marwan Refaat (USA), Susan P. Etheridge (USA), Robert M. Campbell (USA), Edward T. Martin (USA), Swee Chye Quek (Singapore)



Long QT Syndrome(s)

- ♥ Autosomal dominant/autosomal rec.
- ♥ genetically heterogeneous
- ♥ 16 genes ($LQTS_{1-16}$)
- ♥ ≥ 60% genotyped (≥ 90% in families)
- ♥ gene-specific features

LQTS, risk stratification

Risk depends on:

- ♥ **genotype**
- ♥ **phenotype - gender (young: male)**
  - **QTc (≥ 500 ms)**
  - **specific ECG features**

Long QT syndrome, risk stratification

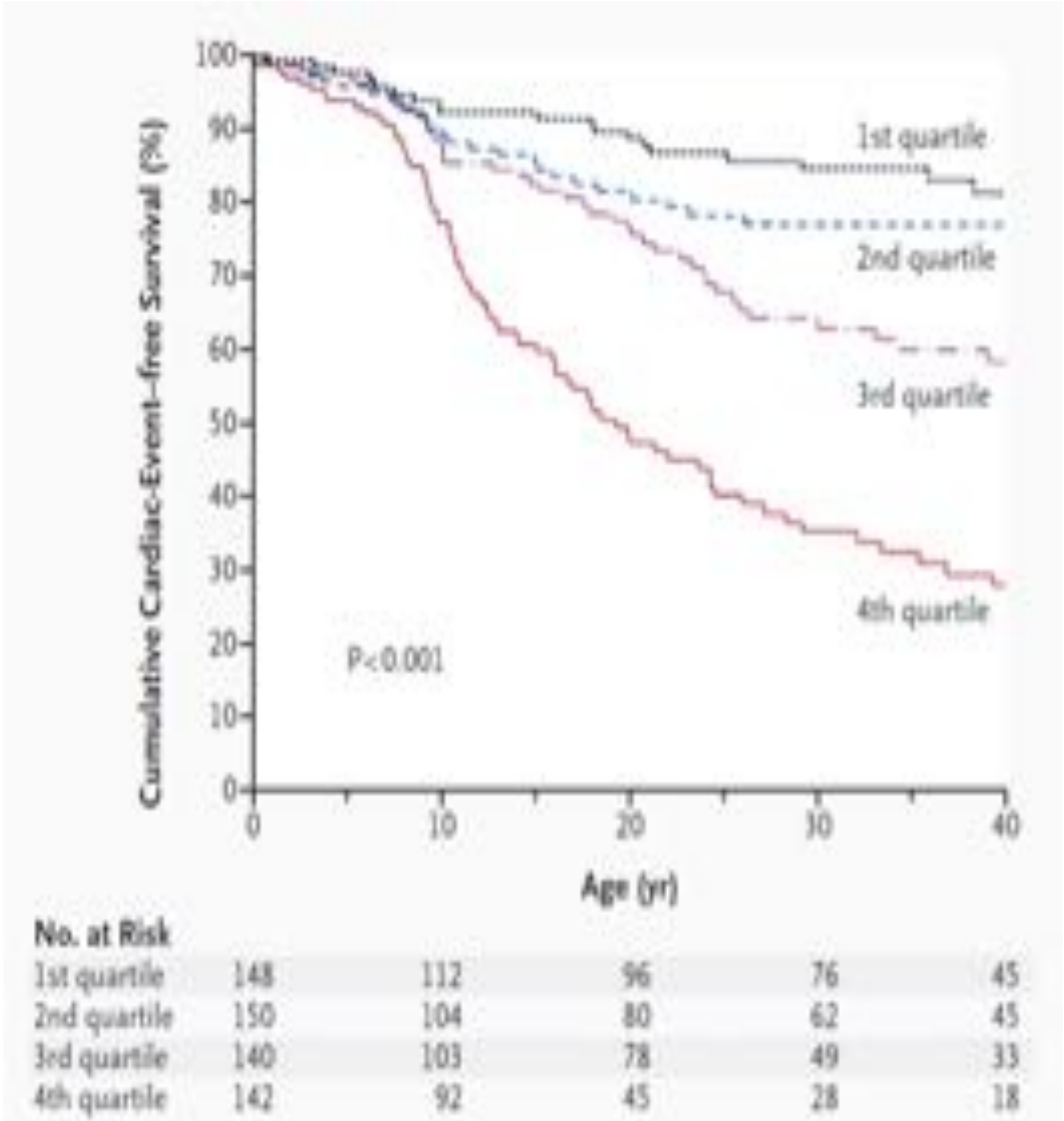
QT_C Quartiles:

1: ≤ 446 ms

2: 447 - 468 ms

3: 469 - 498 ms

4: ≥ 499 ms



NEJM 2003;348:1866-74

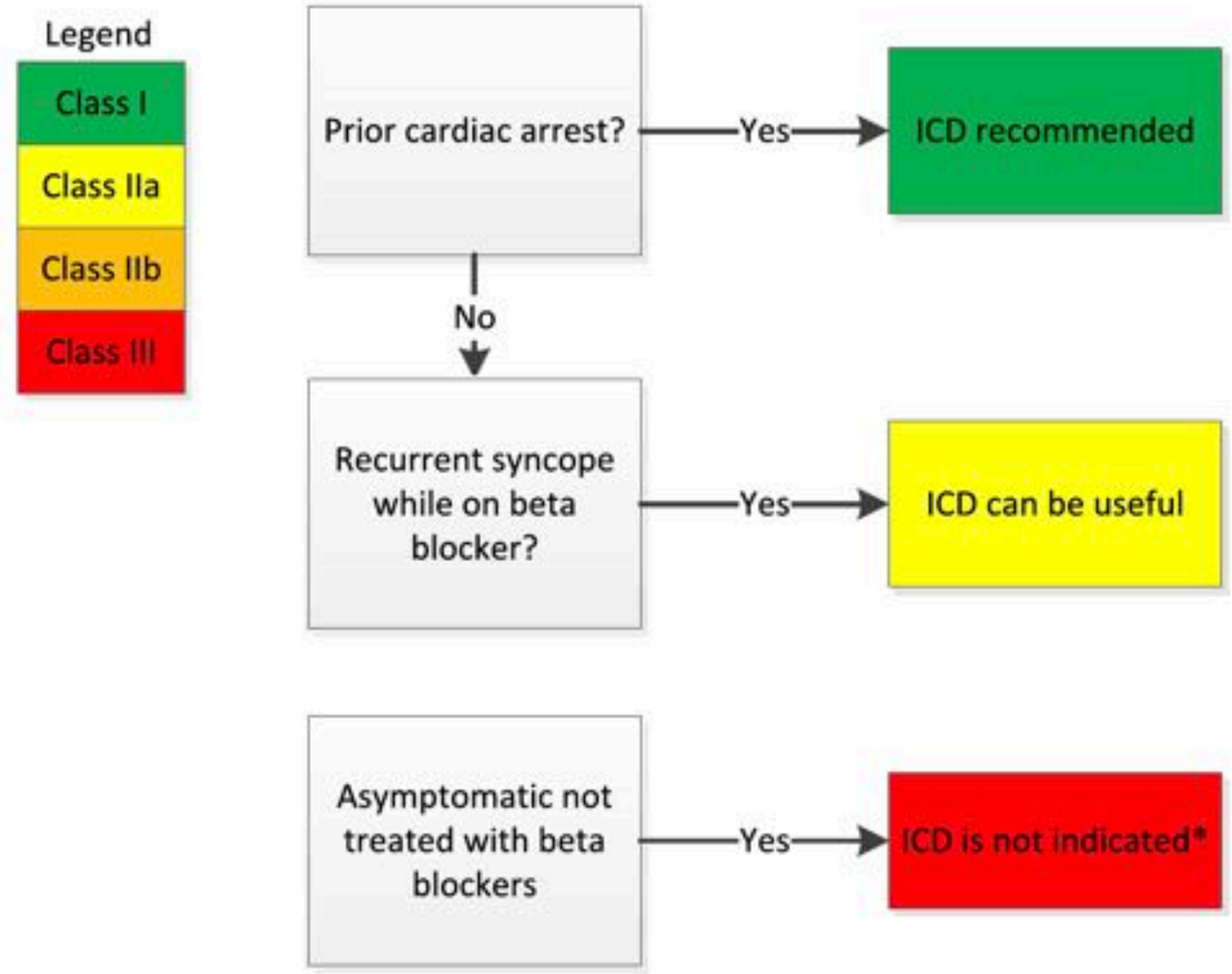
Long QT syndrome

Who are the patients at risk?

- ♥ Aborted sudden death
- ♥ Syncope
- ♥ Patients with long QTc intervals (>500ms)
- ♥ Torsades de Pointes, T-wave alternans
- ♥ Specific mutations (compound mutations)
- ♥ congenital deafness (JLN)

Class	ICD Recommendations
Class I	ICD implantation **is recommended** for patients with a diagnosis of LQTS who are survivors of a cardiac arrest
Class IIa	IICD implantation **can be useful** in patients with a diagnosis of LQTS who experience recurrent syncopal events while on beta-blocker therapy.
Class III	Except under special circumstances, ICD implantation **is <u>not</u> indicated** in asymptomatic LQTS patients who have not been tried on beta-blocker therapy

Family history is NOT a risk factor

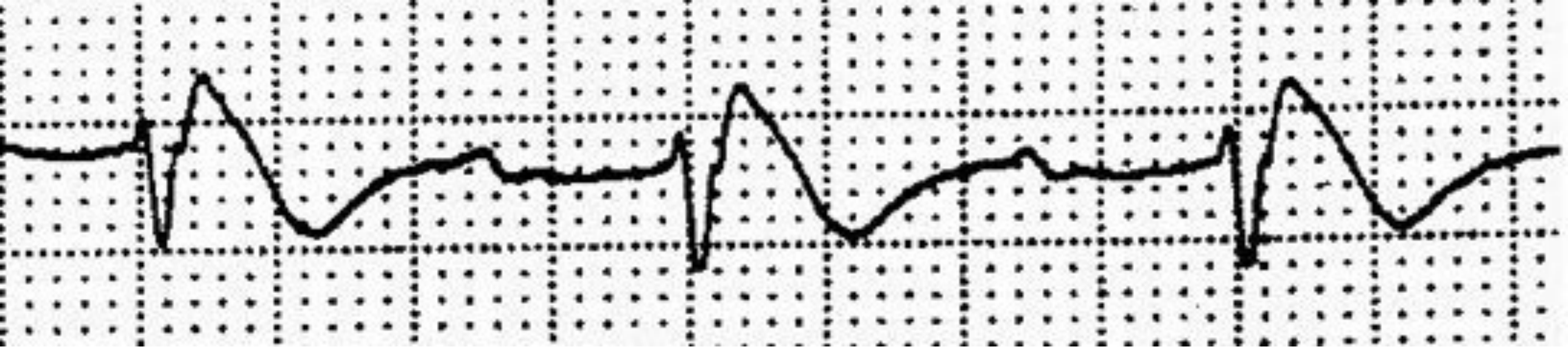


*Except under special circumstances, ICD implantation is not indicated in asymptomatic patients who have not been tried on beta-blocker therapy

Long QT syndrome, asymptomatic pt

When should an ICD be considered?

- ♥ JLNS patient with a long QTc (>500msec)
- ♥ LQT2 pt with QTc > 550
- ♥ LQT3 pt with QTc > 500
- ♥ Torsades de Pointes, T-wave alternans
- ♥ rarely LQT1!
- ♥ Family history of (a)SCD is not a riskfactor

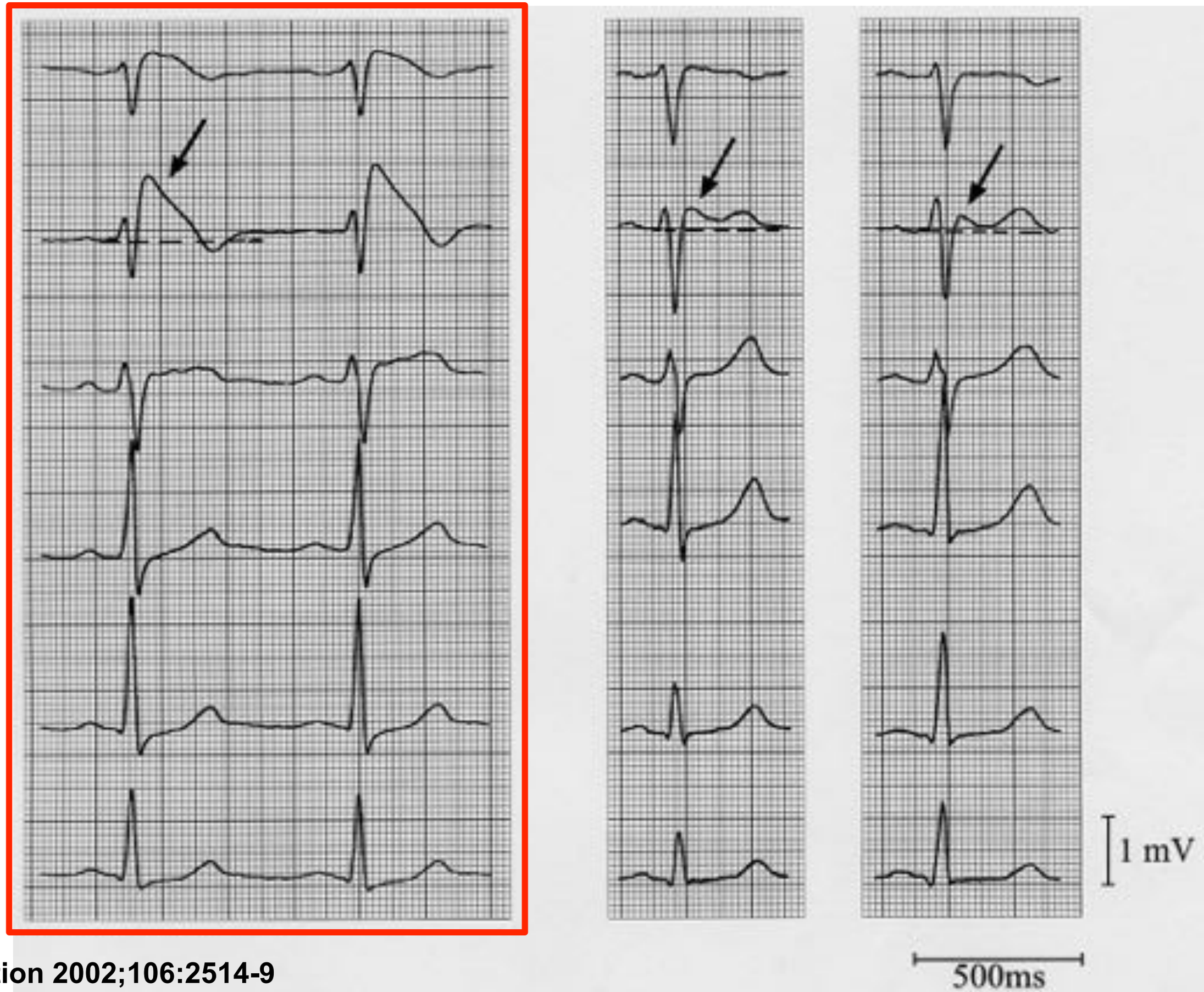


Brugada syndrome

- Monogenetic disease? Oligogenetic!
- ≥18 genes involved
- Type 1 ECG (± drugs)
- documented VF or self terminating PMVT
- Family history of SCD < 45 y.
- 40 years of age, male

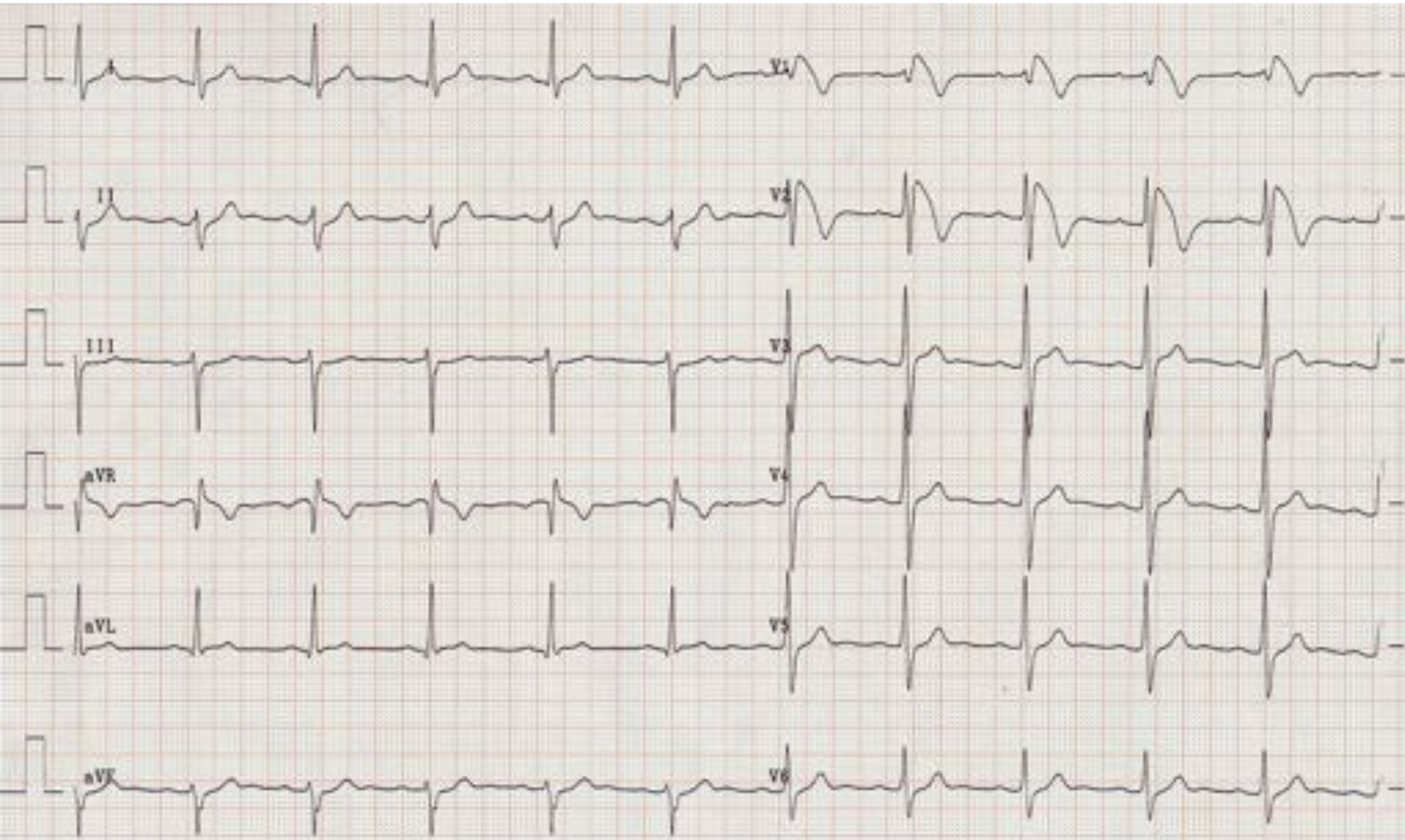
Type 1

Type 2 & 3



Circulation 2002;106:2514-9

Male 39 years



Brugada Syndrome, risk stratification

Asymptomatic patients

♥ spontaneous variation	++
♥ fragmented QRS	+
♥ Genotype (SCN5a or not)	-
♥ ECG variables (HV-interval)	-
♥ EPS inducibility	± ?*

*: mild protocol

Brugada Syndrome, risk stratification

Symptomatic patients

- ♥ documented arrhythmias/VF ++
- ♥ (presumed) arrhyth. syncope ++

Brugada Syndrome

Class	ICD Recommendations
Class I	ICD implantation **is recommended** in patients with a diagnosis of BrS who: • Are survivors of a cardiac arrest, and/or • Have documented spontaneous sustained VT with or without syncope.

Brugada Syndrome

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Class IIa	ICD implantation **can be useful** in patients with a spontaneous diagnostic Type I ECG who have a history of syncope judged to be likely caused by ventricular arrhythmias.

Brugada Syndrome

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Class IIb	ICD implantation **may be considered** in patients with a diagnosis of BrS who develop VF during programmed electrical stimulation (inducible patients).

Brugada Syndrome

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Class IIb	ICD implantation **may be considered** in patients with a diagnosis of BrS who develop VF during programmed electrical stimulation (inducible patients).
Class III	ICD Implantation **is not indicated** in asymptomatic BrS patients with a drug induced type 1 ECG and on the basis of a family history of SCD alone.

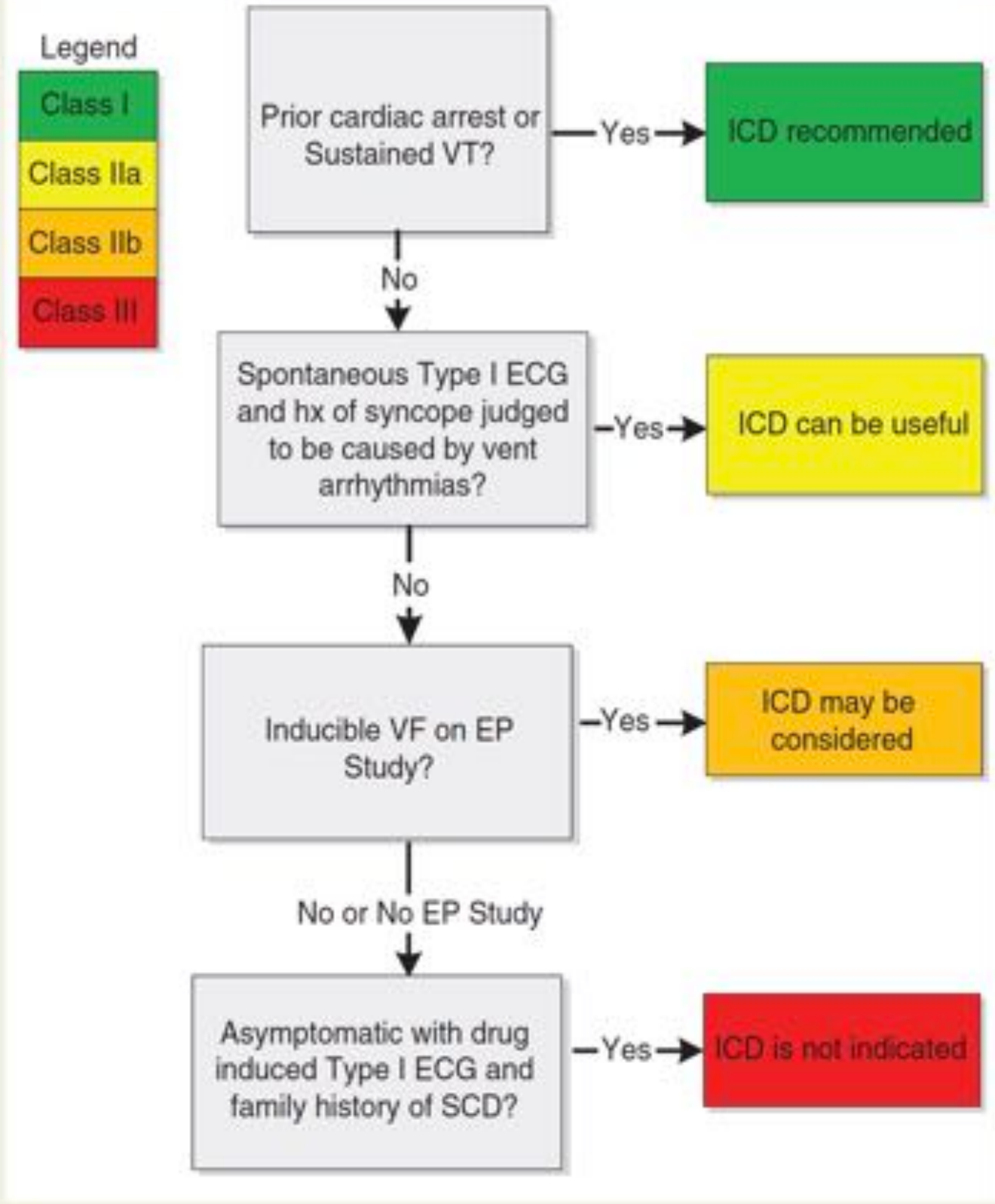
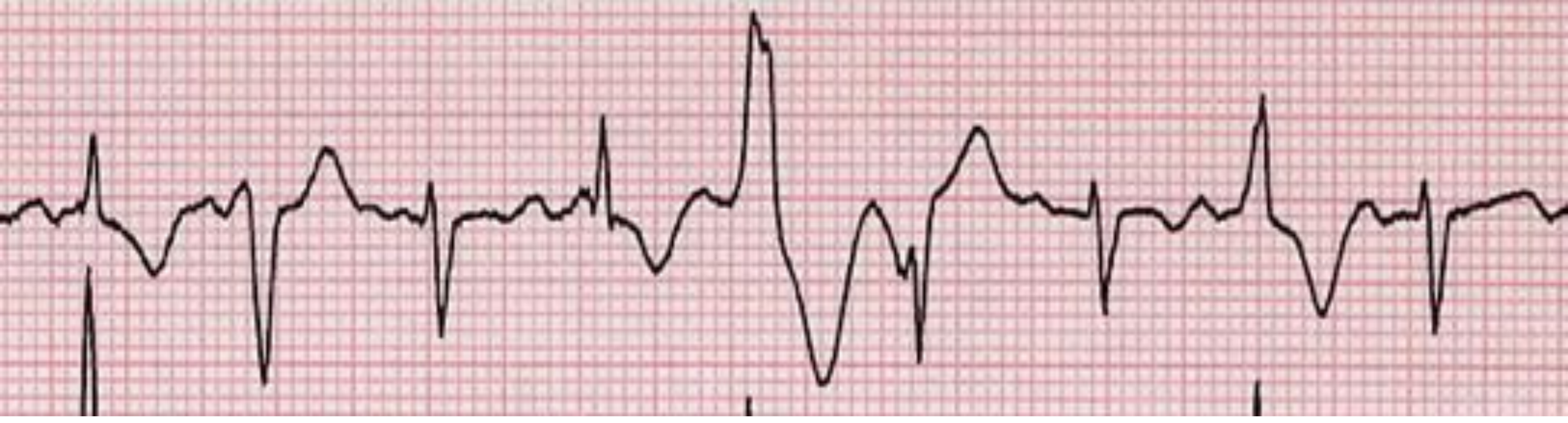


Figure 2 Consensus recommendations for ICDs in patients diagnosed with Brugada syndrome.

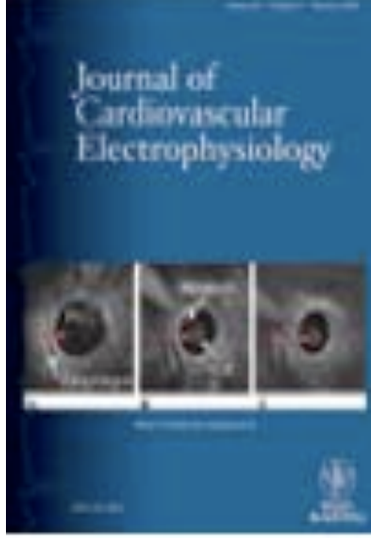


Catecholamine-induced PMVT/VF

- Autosomal dominant
- genetic heterogeneous (5 genes, 1 locus)
- complaints during exercise, emotion, etc
- can start at young age
- baseline ECG = normal!

Expert Consensus Recommendations on CPVT Therapeutic Interventions

Class I	1. The following lifestyle changes ***are recommended*** in all patients with diagnosis of CPVT: a) Limit/ avoid competitive sports; b) Limit/avoid strenuous exercise; c) Limit exposure to stressful environments. 2. Beta-blockers ***are recommended*** in all symptomatic patients with a diagnosis of CPVT.
	3. ICD implantation ***is recommended*** in patients with a diagnosis of CPVT who experience cardiac arrest, recurrent syncope or polymorphic/ bidirectional VT despite optimal medical management, and/or LCSD.
Class IIa	4. Flecainide ***can be a useful*** addition to beta- blockers in patients with a diagnosis of CPVT who experience recurrent syncope or polymorphic/ bidirectional VT while on beta-blockers. 5. Beta-blockers ***can be useful*** in carriers of a pathogenic CPVT mutation without clinical manifestations of CPVT (concealed mutation-positive patients).
Class IIb	6. LCSD ***may be considered*** in patients with a diagnosis of CPVT who experience recurrent syncope or polymorphic/bidirectional VT/ several appropriate ICD shocks while on beta-blockers and in patients who are intolerant or with contraindication to beta-blockers.
Class III	7. ICD as a standalone therapy ***is not indicated*** in an asymptomatic patient with a diagnosis of CPVT.
	8. Programmed Electrical Stimulation ***is not indicated*** in CPVT patients.



Sudden Death in a Young Man with Catecholaminergic Polymorphic Ventricular Tachycardia and Paroxysmal Atrial Fibrillation

STEPHEN PIZZALE, B.H.Sc., B.S.c.N.,* MICHAEL H. GOLLOB, M.D.,* ROBERT GOW, M.D.,† and DAVID H. BIRNIE, M.B. ChB., M.D.*

From the *University of Ottawa Heart Institute, Ottawa, Ontario, Canada; and †Children's Hospital of Eastern Ontario, Ottawa, Ontario, Canada

Sudden Death in Patient with CPVT and PAF. Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a familial condition that presents with exercise-induced syncope or sudden death in children or young adults. In most cases the disease is caused by a mutation in the cardiac ryanodine receptor (RyR2) gene. Current evidence suggests that primary therapy for CPVT is beta blockade and implantable cardioverter defibrillator (ICD) placement. There is a recent report of a patient with CPVT who died despite appropriate ICD therapies, and we report a similar case. Our patient died after probably initially receiving inappropriate ICD shocks for atrial fibrillation. We recommend that utmost efforts should be made to prevent shocks including repeated exercise testing to confirm suppression of PVT. *(J Cardiovasc Electrophysiol, Vol. 19, pp. 1319-1321, December 2008)*

This starts with not implanting an ICD!!

IVF - short coupled TdP, ICD

Expert Consensus Recommendations on ***IVF Therapeutic Interventions***

Class I	1. ICD implantation ***is recommended*** in patients with a diagnosis of IVF.
Class IIb	2. Antiarrhythmic therapy with quinidine, programmed electrical stimulation guided or empirical, ***may be considered*** in patients with a diagnosis of IVF in conjunction with ICD implantation or when ICD implantation is contraindicated or refused.
	3. Ablation of Purkinje potentials ***may be considered*** in patients with a diagnosis of IVF presenting with uniform morphology premature ventricular contractions in conjunction with ICD implantation or when ICD implantation is contraindicated or refused.
	4. If a first-degree relative of an IVF victim presents with unexplained syncope and no identifiable phenotype following thorough investigation, then after careful counseling an ICD implant ***may be considered***.

Primary arrhythmia syndromes (2015)

- ♥ Long QT syndrome(s)
- ♥ Short QT syndrome
- ♥ Brugada syndrome
- ♥ Catecholamine-induced PMVT/VF

Ablation?

- ♥ Short-coupled Torsades de Pointes
- ♥ Isolated conduction disorders (AVN, BB)
- ♥ Atrial fibrillation
- ♥ Sinus node disease, atrial standstill
- ♥ Idiopathic ventricular fibrillation

Primary arrhythmia syndromes (2015)

	Ablation
♥ Long QT syndrome(s)	±
♥ Short QT syndrome	-
♥ Brugada syndrome	+
♥ Catecholamine-induced PMVT/VF	±
♥ Short-coupled Torsades de Pointes	+
♥ Isolated conduction disorders (AVN, BB)	-
♥ Atrial fibrillation	+
♥ Sinus node disease, atrial standstill	-
♥ Idiopathic ventricular fibrillation	+

Mapping and Ablation of Ventricular Fibrillation Associated With Long-QT and Brugada Syndromes

Michel Haïssaguerre, MD; Fabrice Extramiana, MD; Mélèze Hocini, MD; Bruno Cauchemez, MD; Pierre Jaïs, MD; Jose Angel Cabrera, MD; Geronimo Farre, MD; Antoine Leenhardt, MD; Prashanthan Sanders, MBBS; Christophe Scavée, MD; Li-Fern Hsu, MBBS; Rukshen Weerasooriya, MBBS; Dipen C. Shah, MD; Robert Frank, MD; Philippe Maury, MD; Marc Delay, MD; Stéphane Garrigue, MD; Jacques Clémenty, MD

Background—The long-QT and Brugada syndromes are important substrates of malignant ventricular arrhythmia. The feasibility of mapping and ablation of ventricular arrhythmias in these conditions has not been reported.

Methods and Results—Seven patients (4 men; age, 38±7 years; 4 with long-QT and 3 with Brugada syndrome) with episodes of ventricular fibrillation or polymorphic ventricular tachycardia and frequent isolated or repetitive premature beats were studied. These premature beats were observed to trigger ventricular arrhythmias and were localized by mapping the earliest endocardial activity. In 4 patients, premature beats originated from the peripheral right (1 Brugada) or left (3 long-QT) Purkinje conducting system and were associated with variable Purkinje-to-muscle conduction times (30 to 110 ms). In the remaining 3 patients, premature beats originated from the right ventricular outflow tract, being 25 to 40 ms ahead of the QRS. The accuracy of mapping was confirmed by acute elimination of premature beats after 12±6 minutes of radiofrequency applications. During a follow-up of 17±17 months using ambulatory monitoring and defibrillator memory interrogation, no patients had recurrence of symptomatic ventricular arrhythmia but 1 had persistent premature beats.

Conclusion—Triggers from the Purkinje arborization or the right ventricular outflow tract have a crucial role in initiating ventricular fibrillation associated with the long-QT and Brugada syndromes. These can be eliminated by focal radiofrequency ablation. (***Circulation*. 2003;108:925-928.**)

Long QT syndrome

Ablation experience

- 4 patients
- 1 patient RVOT ectopy,
- 1 patient post fascicle, 2 P-fiber activity LV
- ectopy as the target
- FU 17+7 months: arrhythmia free

Brugada Syndrome, ablation

Methods

- ♥ Ectopy as the target
- ♥ Substrate as the target

Brugada Syndrome

Class	Catheter Ablation Recommendation
Class IIb	Catheter ablation ***may be considered*** in patients with a diagnosis of BrS and history of arrhythmic storms or repeated appropriate ICD shocks.

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

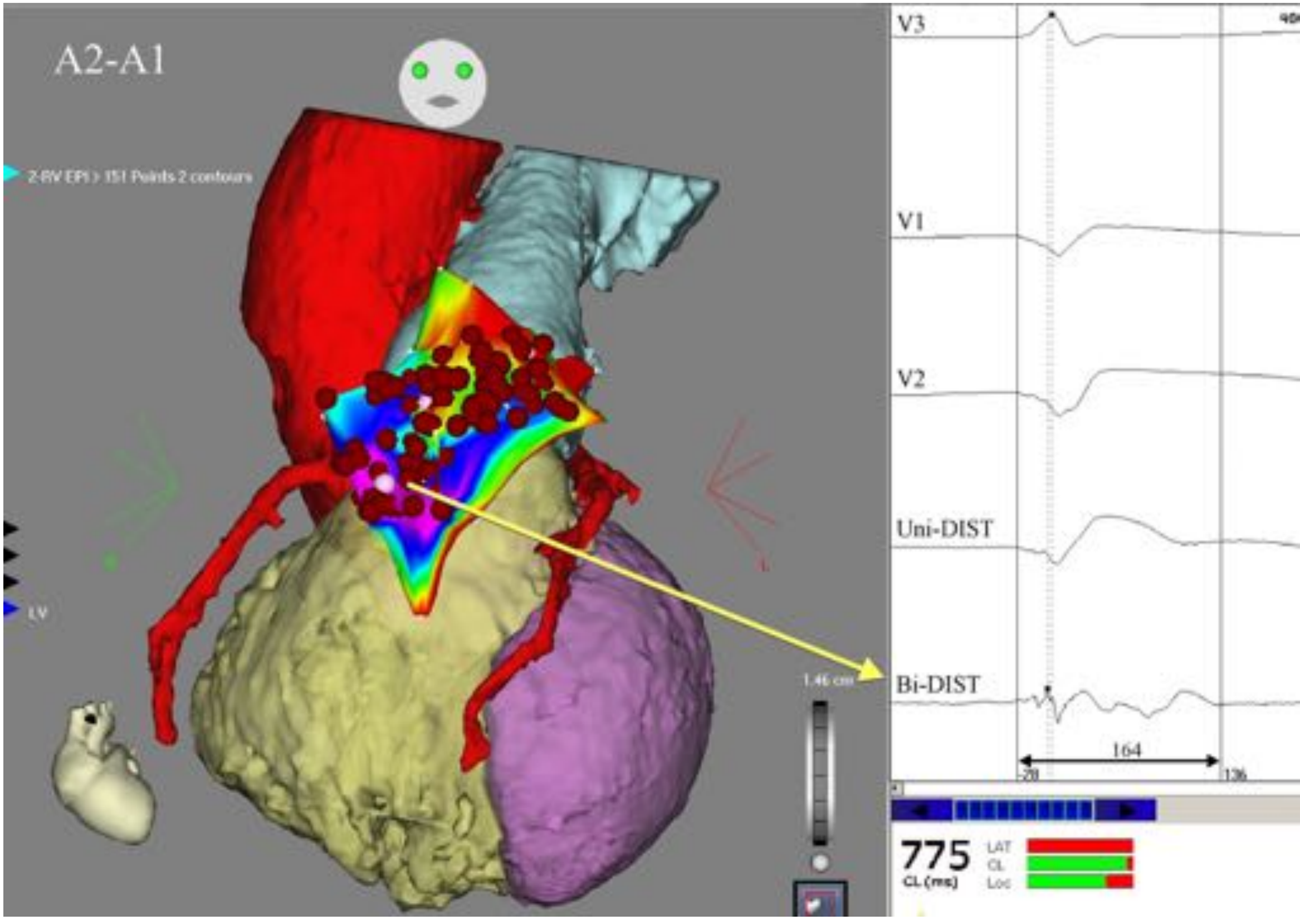
American Heart Association.
Learn and Live.

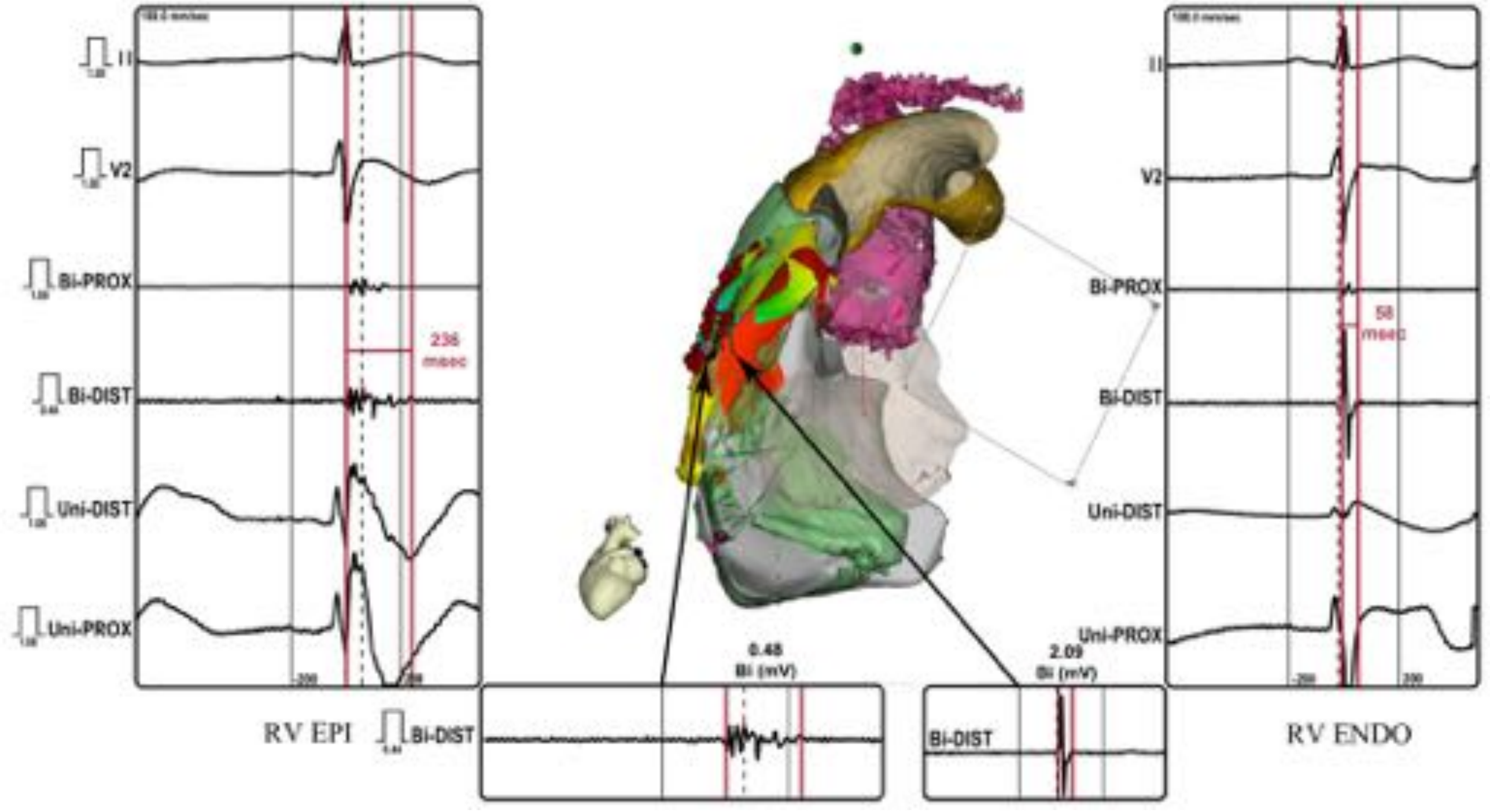
Prevention of Ventricular Fibrillation Episodes in Brugada Syndrome by Catheter Ablation Over the Anterior Right Ventricular Outflow Tract Epicardium

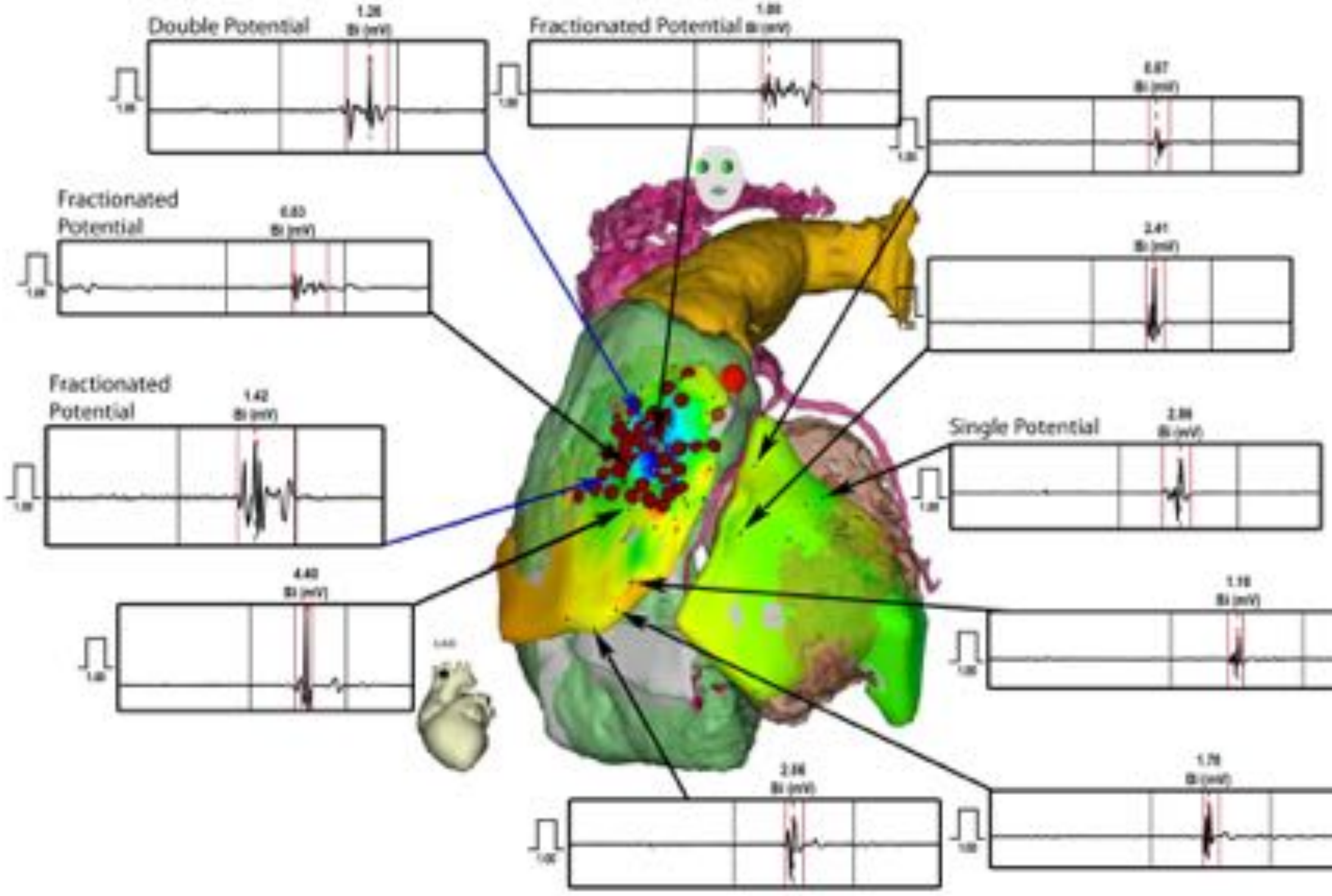
Koonlawee Nademanee, Gumpanart Veerakul, Pakorn Chandananattha, Lertlak Chaothawee, Aekarach Ariyachaipanich, Kriengkrai Jirasirirojanakorn, Khanchit Likittanasombat, Kiertijai Bhuripanyo and Tachapong Ngarmukos

Circulation published online Mar 14, 2011:

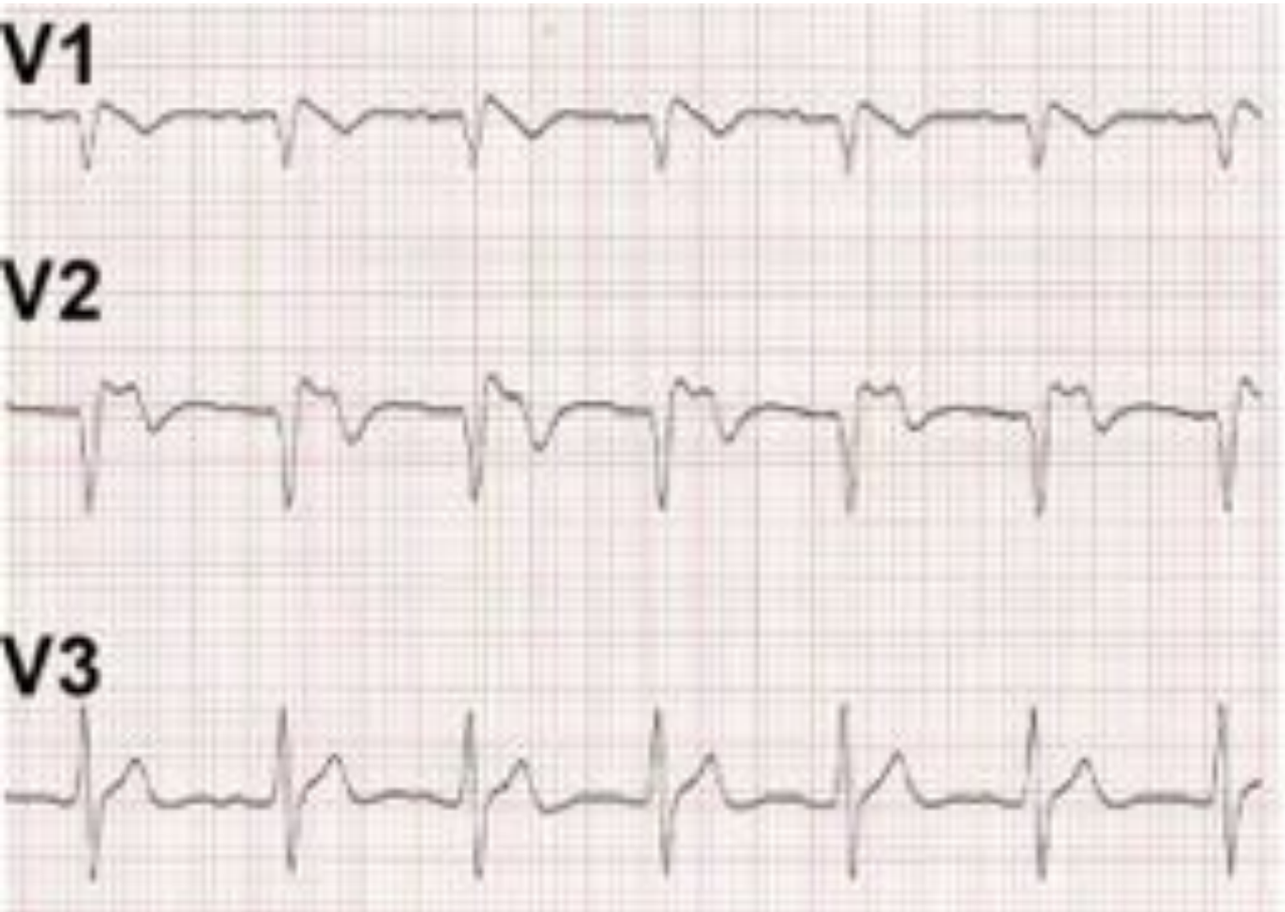
Circulation. 2011;12:1270-9


A2-A1
2-RV EP1 > 151 Points 2 contours
LV
1.46 cm
V3
V1
V2
Uni-DIST
Bi-DIST
164
-20
136
775
CL (ms)
LAT
CL
Loc

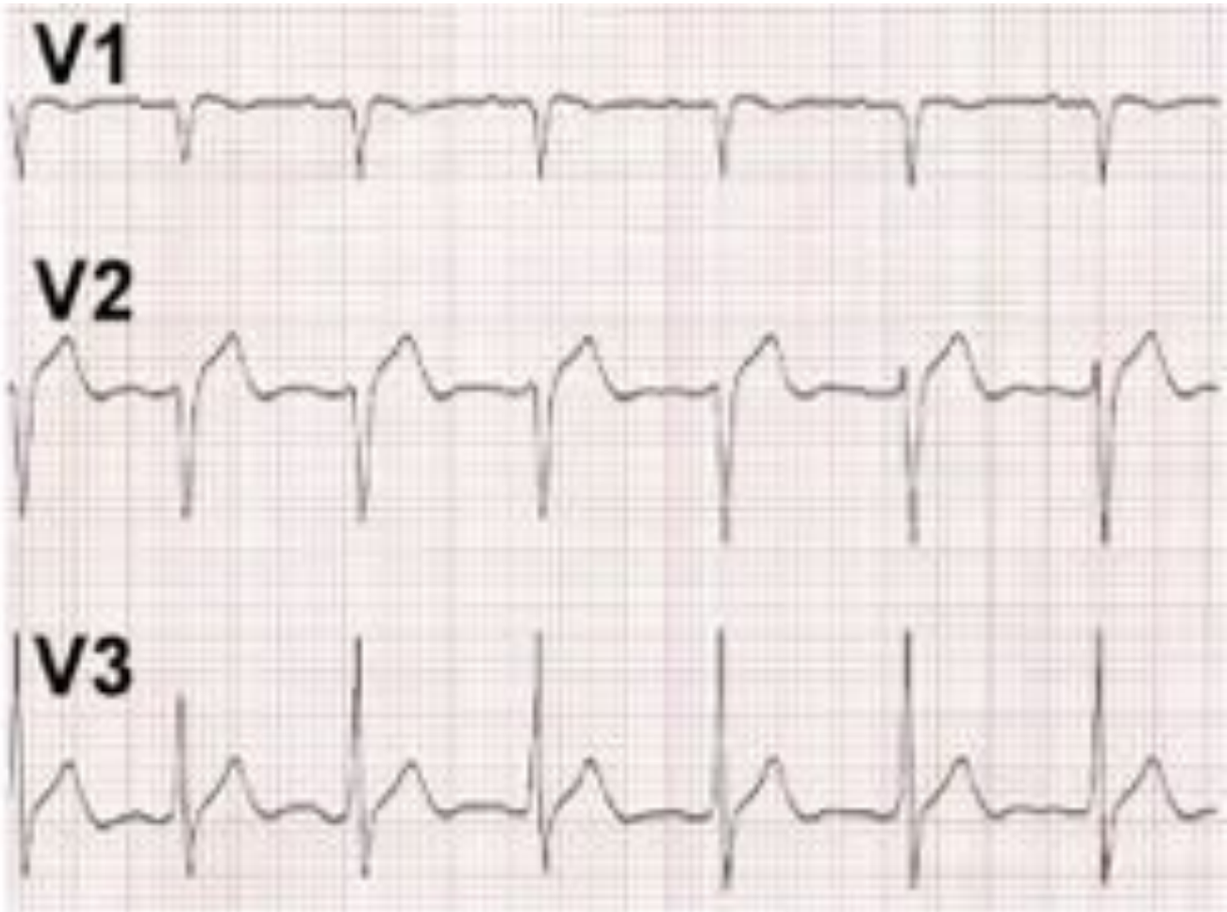

II
V2
Bi-PROX
236
msec
Bi-DIST
Uni-DIST
Uni-PROX
-200
200
0.48
Bi (mV)
RV EPI
Bi-DIST
2.09
Bi (mV)
Bi-DIST
II
V2
Bi-PROX
58
msec
Bi-DIST
Uni-DIST
Uni-PROX
-200
200
RV ENDO

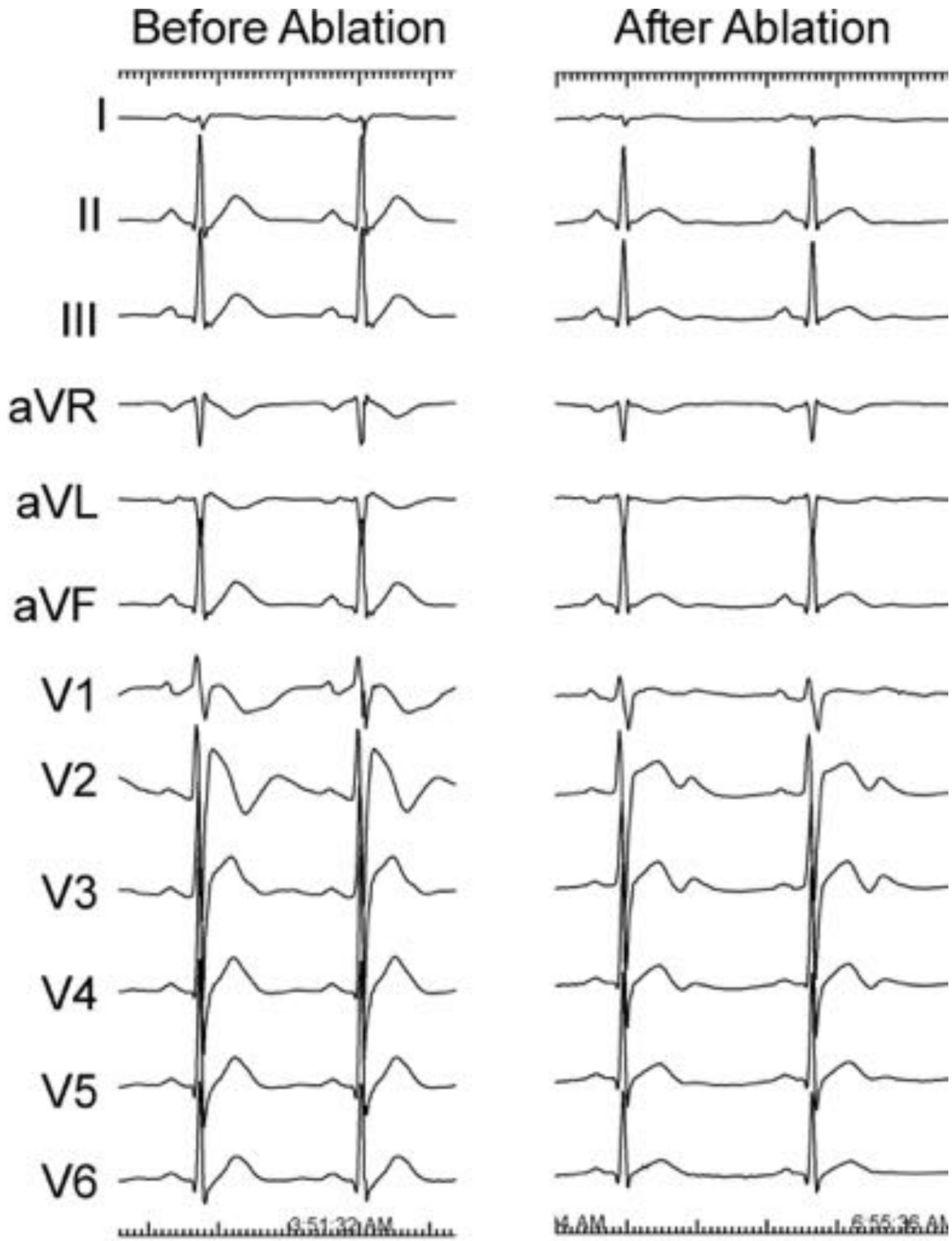

Double Potential
1.26
Bi (mV)
Fractionated Potential
1.88
Bi (mV)
6.87
Bi (mV)
Fractionated Potential
6.83
Bi (mV)
2.41
Bi (mV)
Fractionated Potential
1.42
Bi (mV)
Single Potential
2.88
Bi (mV)
4.40
Bi (mV)
1.18
Bi (mV)
2.16
Bi (mV)
1.76
Bi (mV)

Before ablation

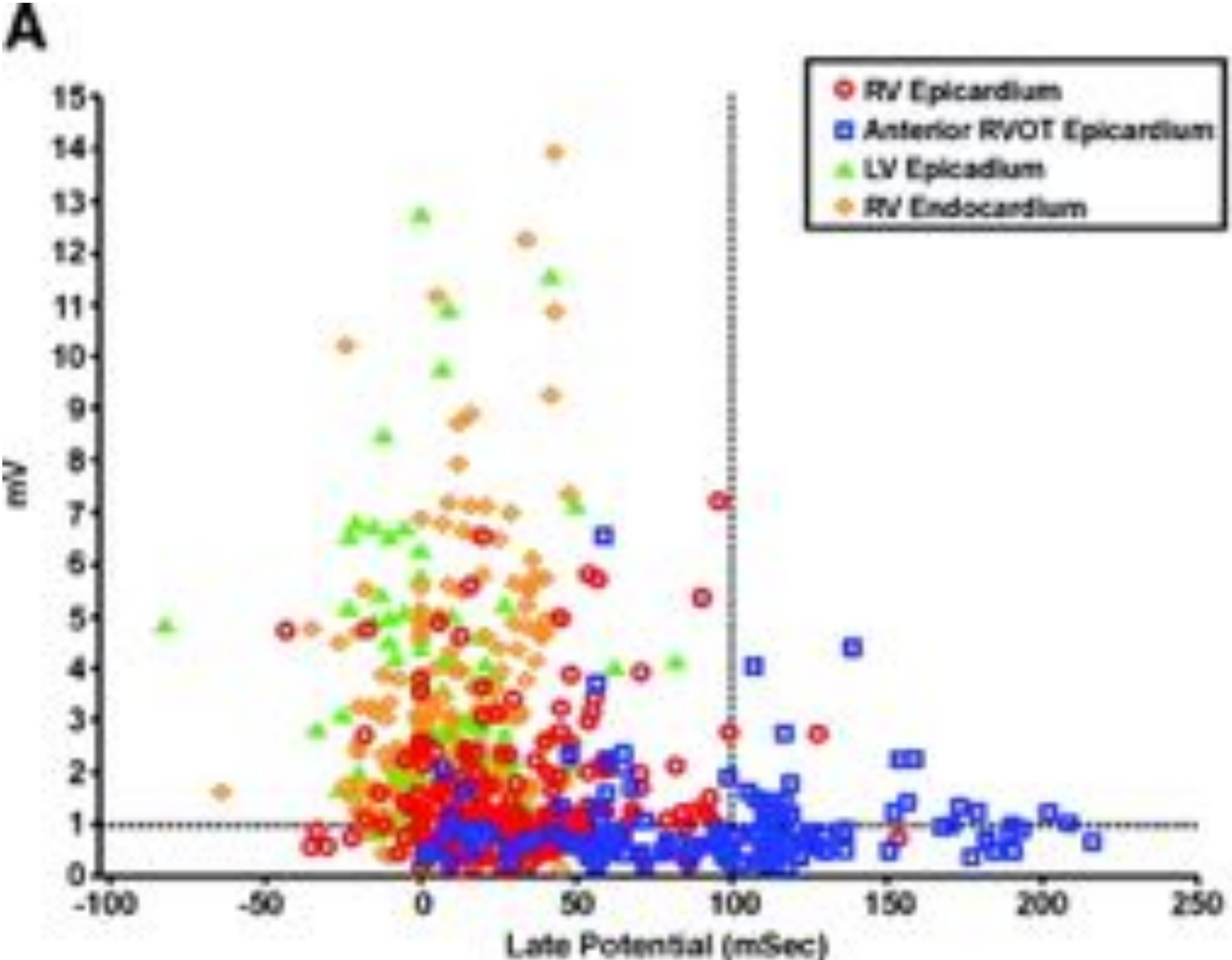


1 month post ablation




Before Ablation
After Ablation
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

A


RV Epicardium
Anterior RVOT Epicardium
LV Epicadium
RV Endocardium
mV
15
14
13
12
11
10
9
8
7
6
5
4
3
2
1
0
-100
-50
0
50
100
150
200
250
Late Potential (mSec)

B

RV Epicardium
Anterior RVOT Epicardium
LV Epicadium
RV Endocardium

Electrogram Duration (mSec)

300
250
200
150
100
50
0

-100
-50
0
50
100
150
200
250

Late Potential (mSec)

BrS, epicardial approach

- **Only at the epicardium of the RVOT area (anterior) one finds**
  - **late to very late potentials**
  - **low voltage signals**

Nademanee e.a. Circulation 2011

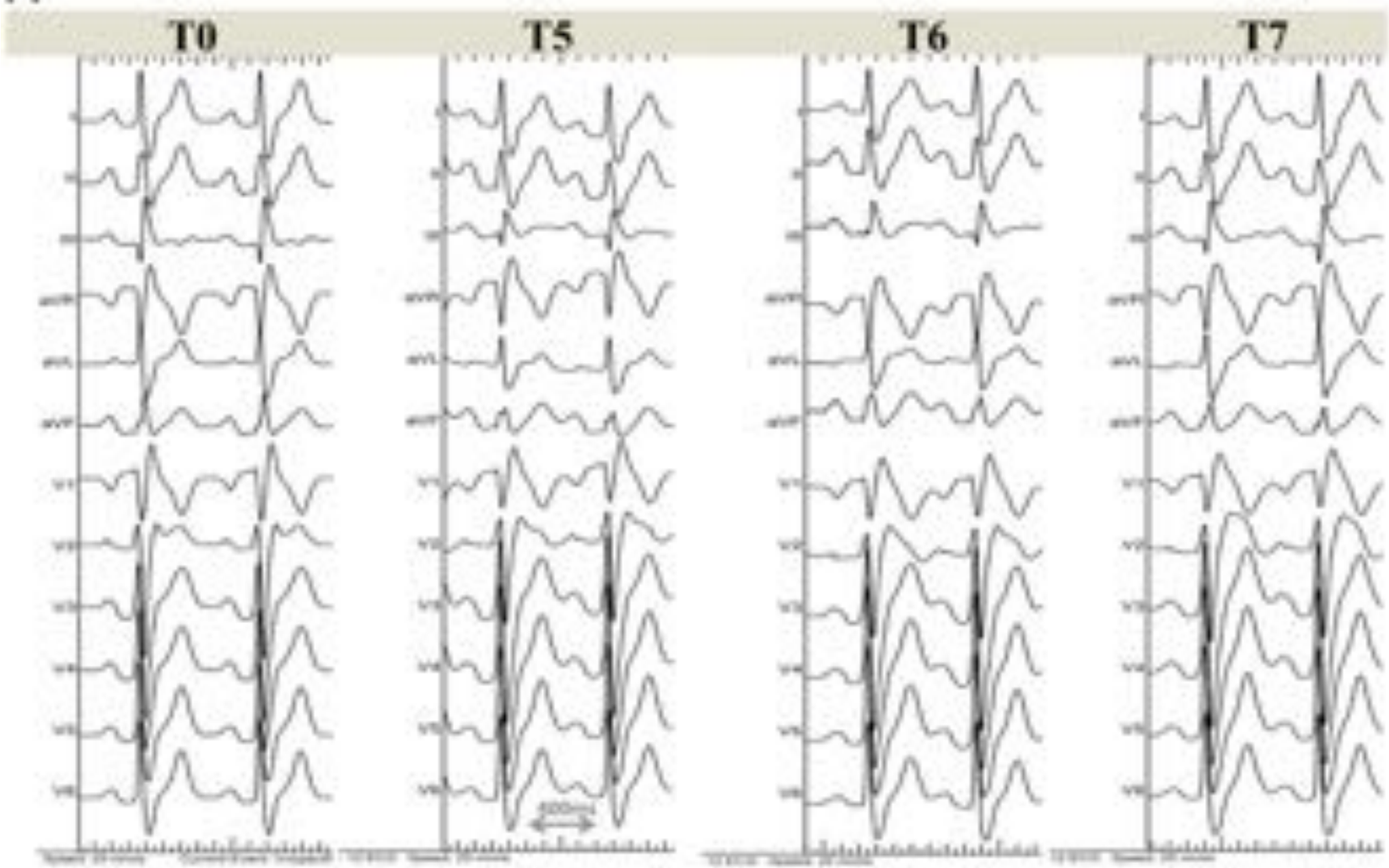
2013

IMAGE

Insight into the mechanism of Brugada syndrome: Epicardial substrate and modification during ajmaline testing

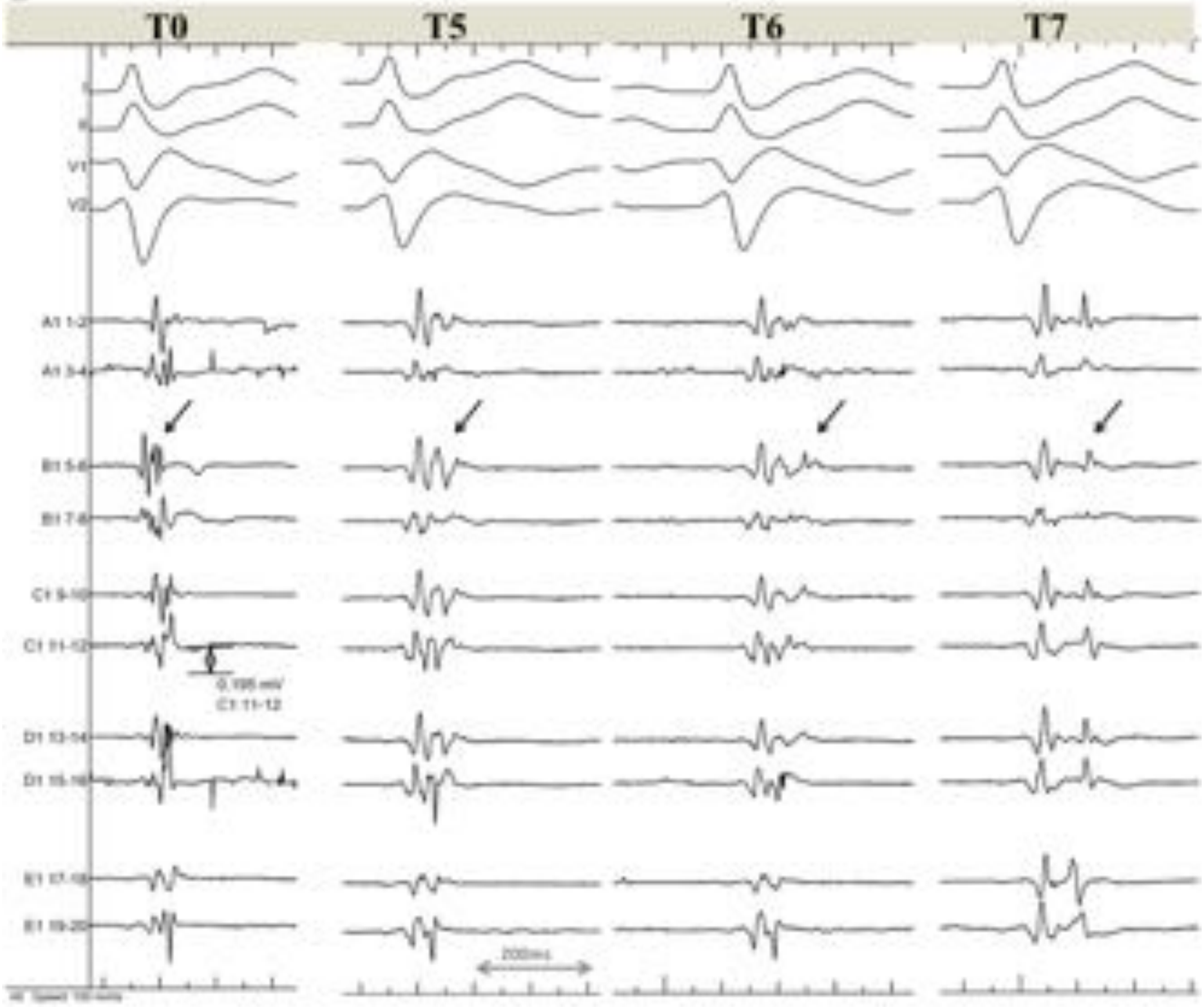
Frédéric Sacher, MD, Laurence Jesel, MD, Pierre Jais, MD, Michel Haïssaguerre, MD

From the Bordeaux University Hospital and LIRYC, L'Institut de rythmologie et modélisation cardiaque, Université de Bordeaux, Bordeaux-Pessac, France.

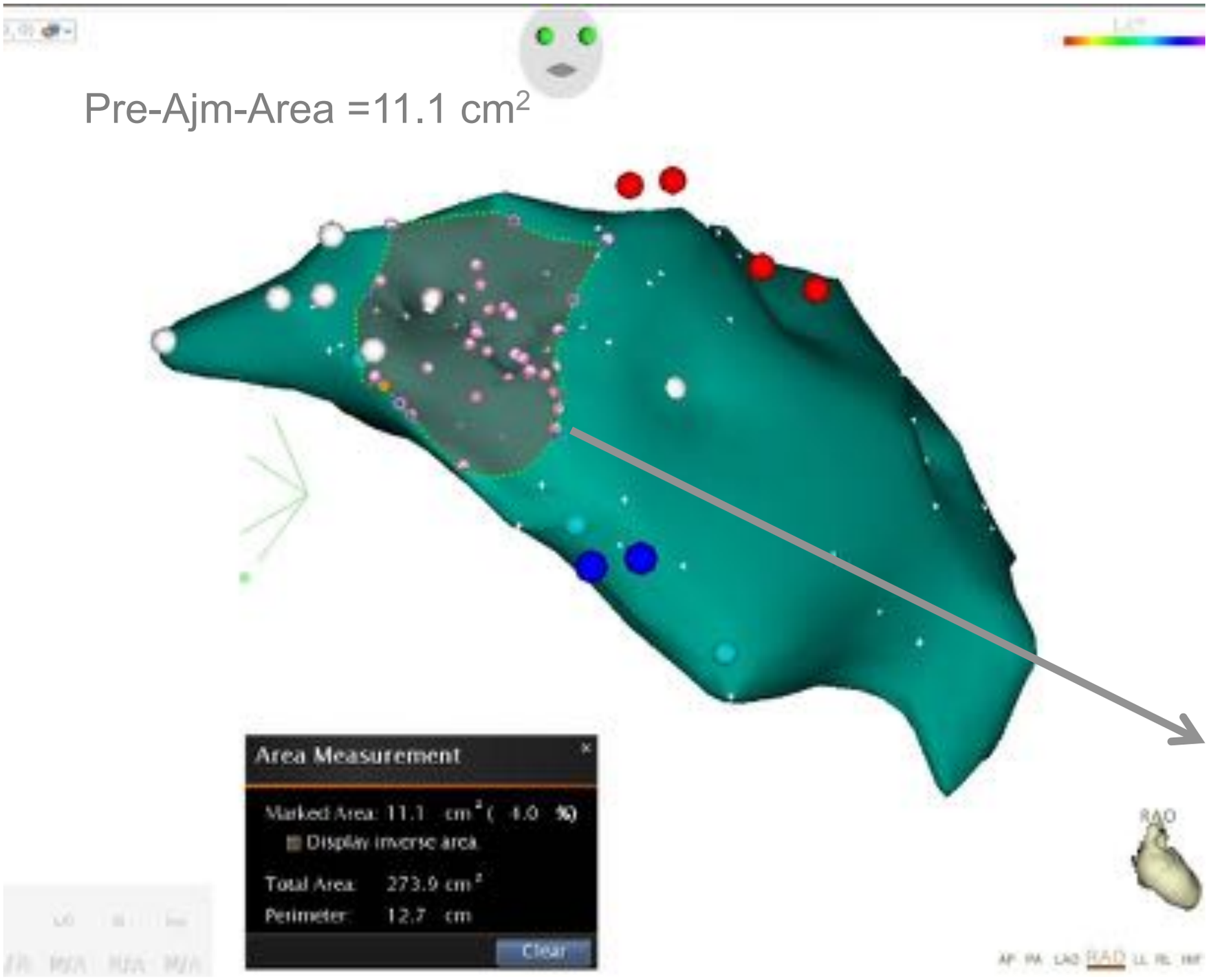


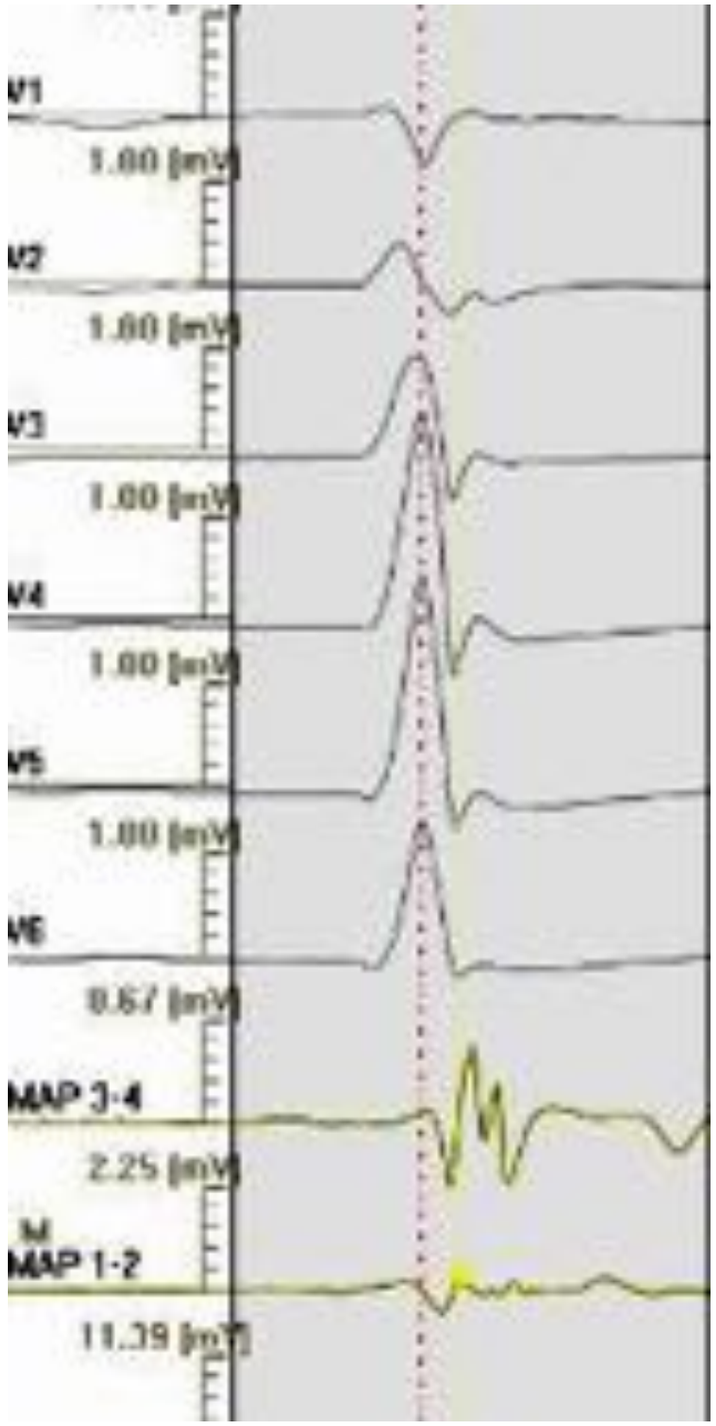
Ajmaline

B

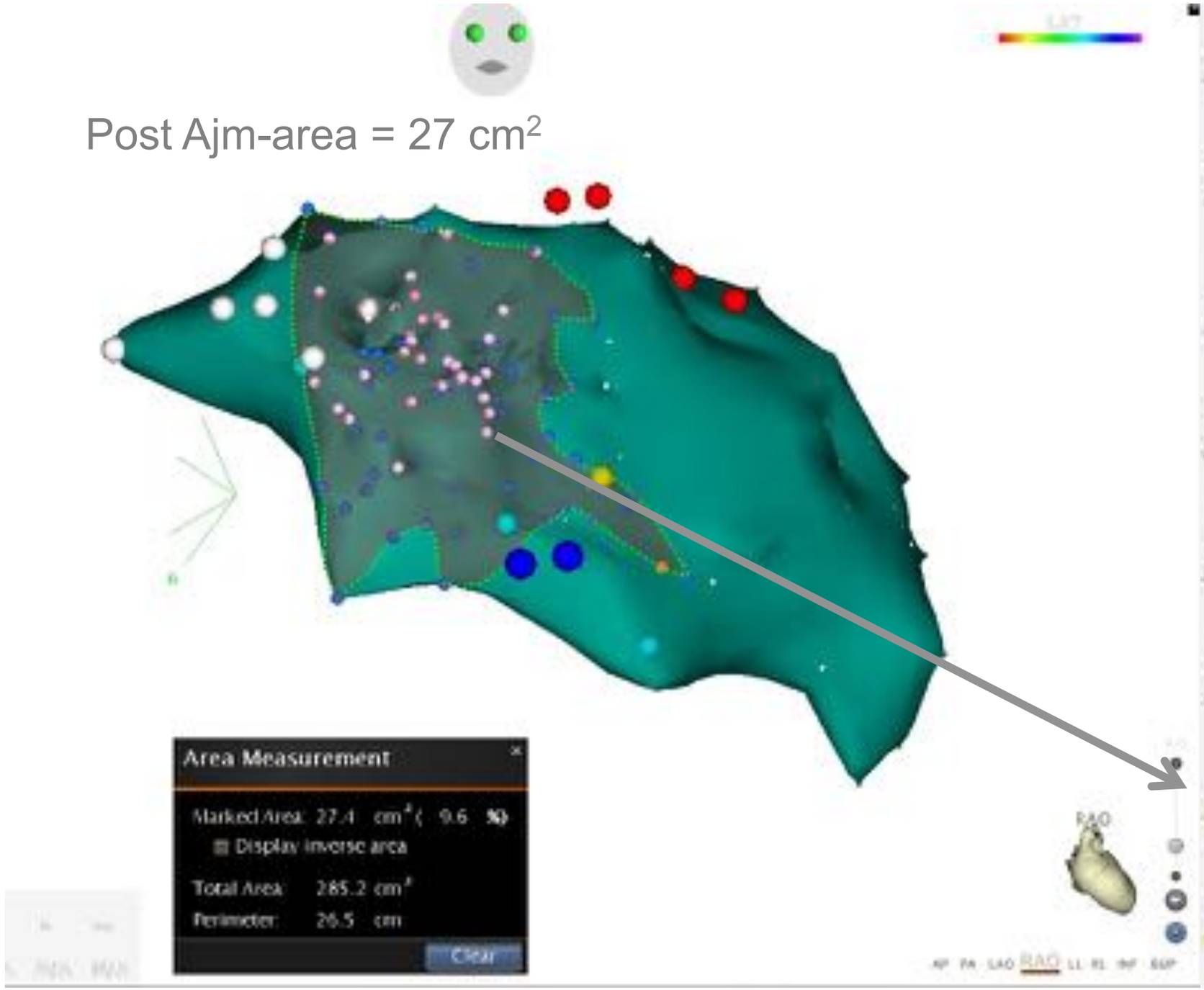

T0
T5
T6
T7
I
II
V1
V2
A1 1-2
A1 3-4
B1 5-6
B1 7-8
C1 9-10
C1 11-12
0.195 mV
C1 11-12
D1 13-14
D1 15-16
E1 17-18
E1 19-20
200ms

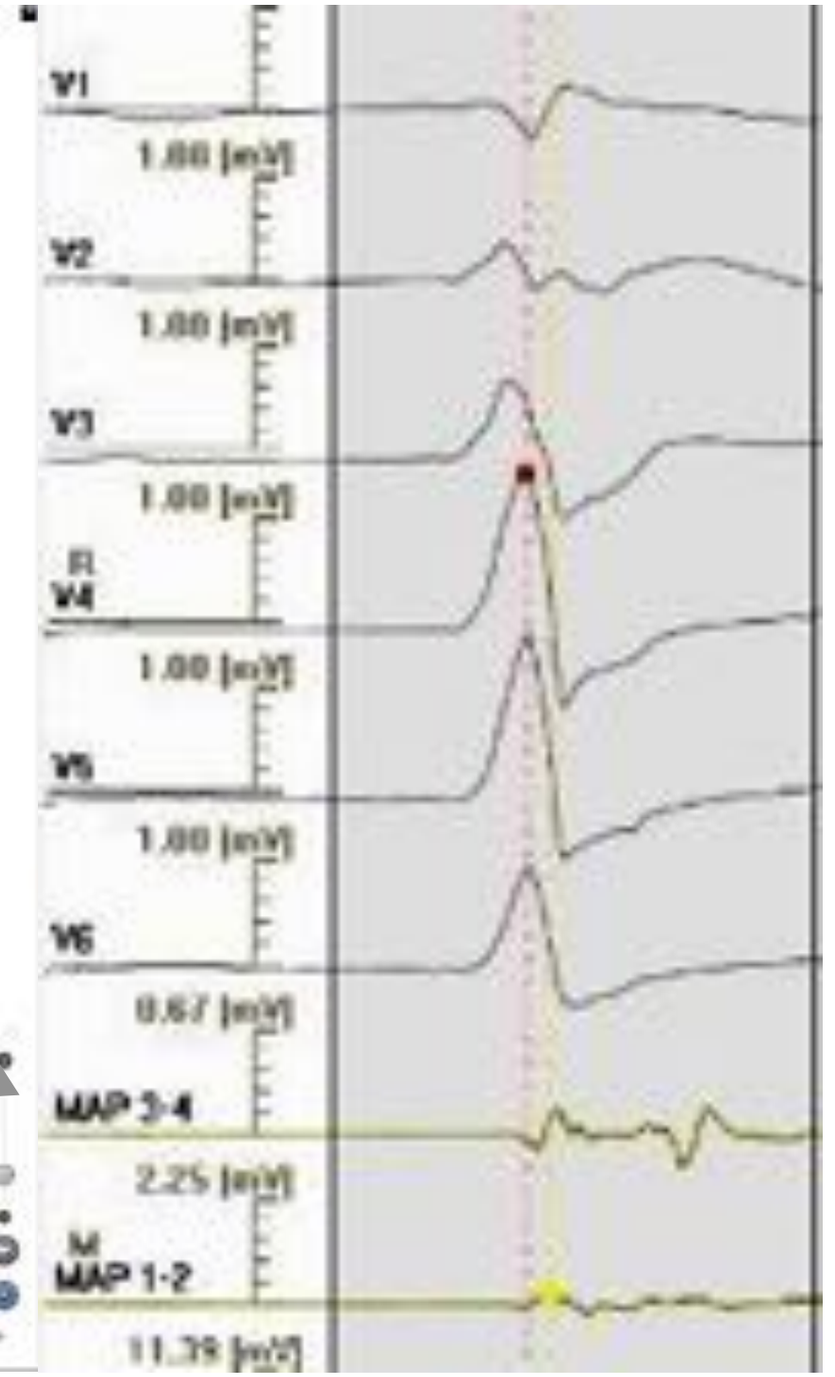
Pre-Ajm-Area =11.1 cm^2





Post Ajm-area = 27 cm^2





Brugada Syndrome Phenotype Elimination by Epicardial Substrate Ablation

Running title: *Brugada et al.; Brugada Syndrome Phenotype Elimination*

Josep Brugada, MD[1]*; Carlo Pappone, MD, PhD[2]*; Antonio Berruezo, MD, PhD[1];

Gabriele Vicedomini, MD[2]; Francesco Manguso, MD, PhD[2]; Giuseppe Ciconte, MD[2];

Luigi Giannelli, MD[2]; Vincenzo Santinelli, MD[2]

[1]Arrhythmia Section, Cardiology Department, Thorax Institute, Hospital Clinic and IDIBAPS (Institut d'Investigació Agustí Pi i Sunyer), Barcelona, Catalonia, Spain; [2]Arrhythmology Departments, Maria Cecilia Hospital, Cotignola and Policlinico San Donato, University of Milan, Milan, Italy

*contributed equally as first authors

Circ Arrhythmia and Electrophysiology 2015, in press

CPVT, ablation

Methods

- ♥ Ectopy as the target

Circulation
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American Heart Association.

Successful Catheter Ablation of Bidirectional Ventricular Premature Contractions Triggering Ventricular Fibrillation in Catecholaminergic Polymorphic Ventricular Tachycardia With *RyR2* Mutation

Takashi Kaneshiro, Yoshihisa Naruse, Akihiko Nogami, Hiroshi Tada, Kentaro Yoshida, Yukio Sekiguchi, Nobuyuki Murakoshi, Yoshiaki Kato, Hitoshi Horigome, Mihoko Kawamura, Minoru Horie and Kazutaka Aonuma

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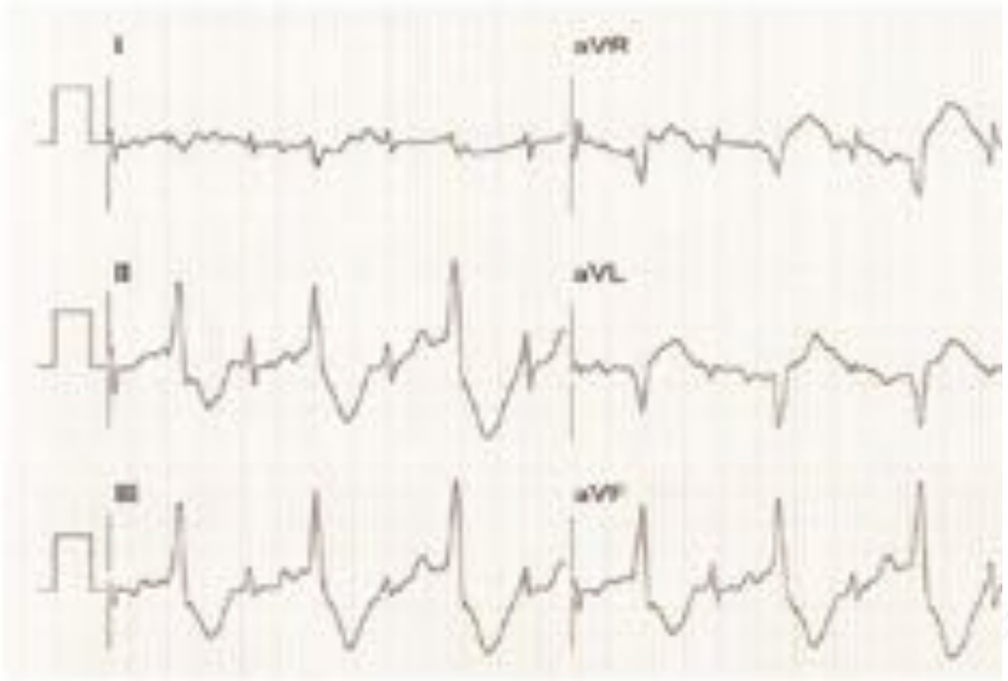
doi: 10.1161/CIRCEP.111.966549

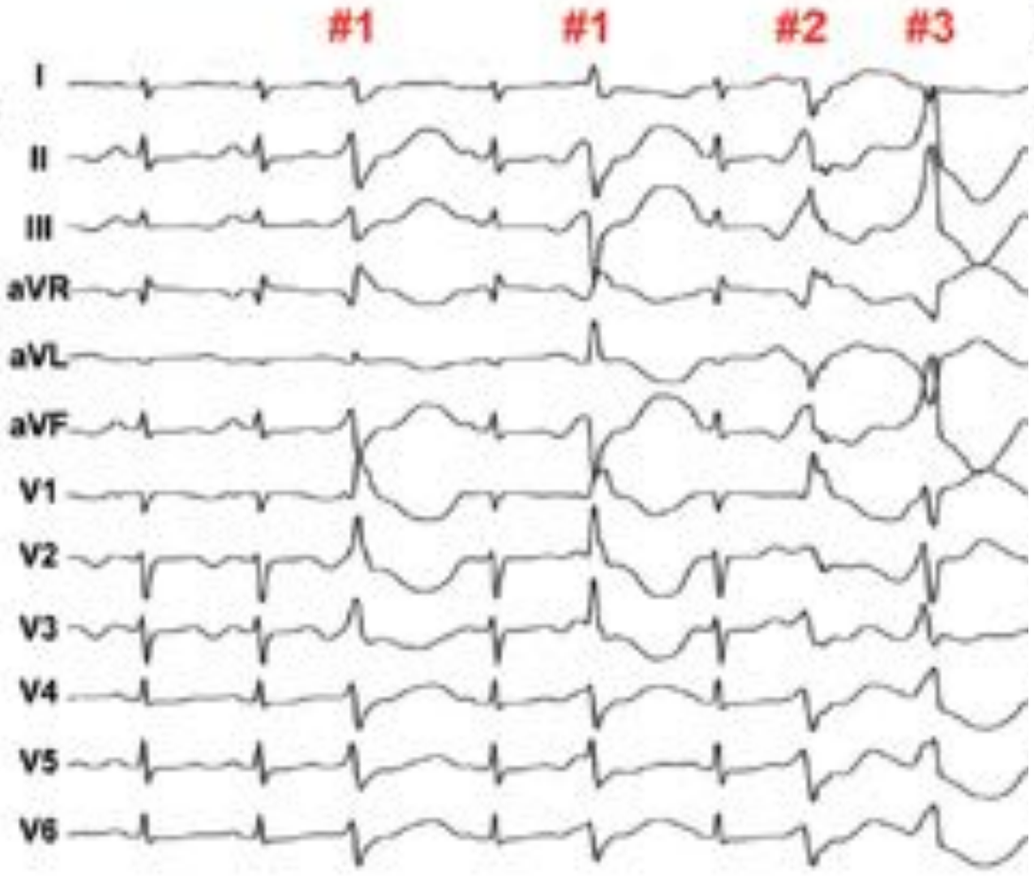
One symptomatic RyR2+ patient, only betablocker, VES targeted approach

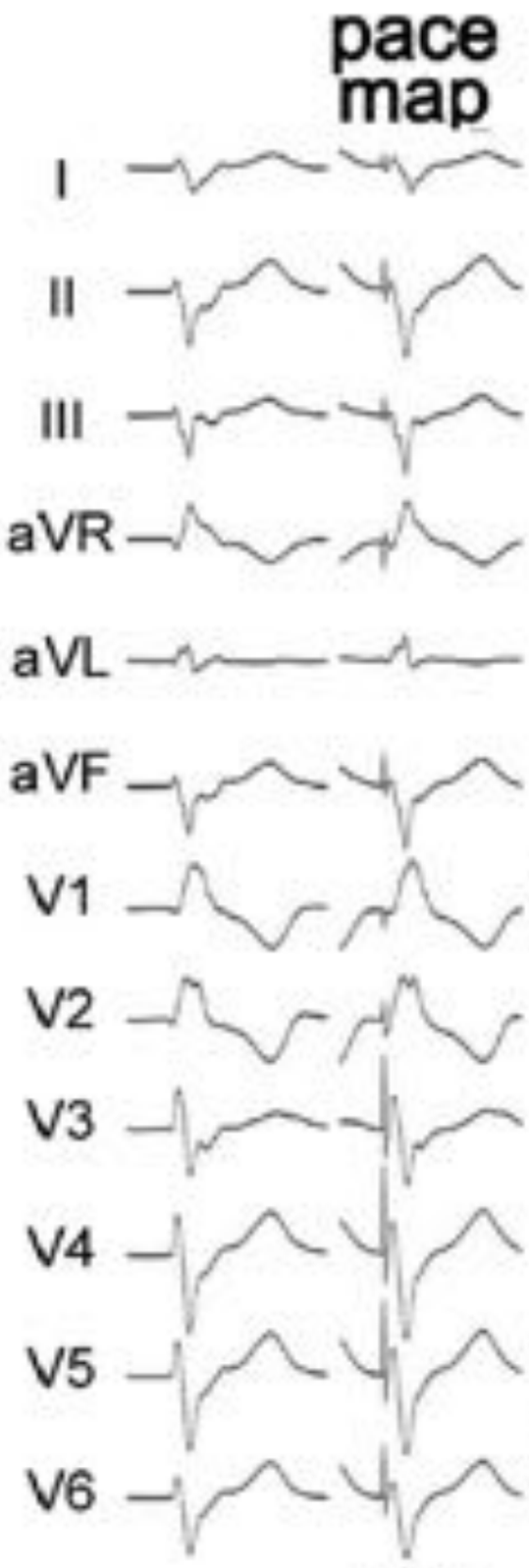
VPC #1 : LV inferior septum origin

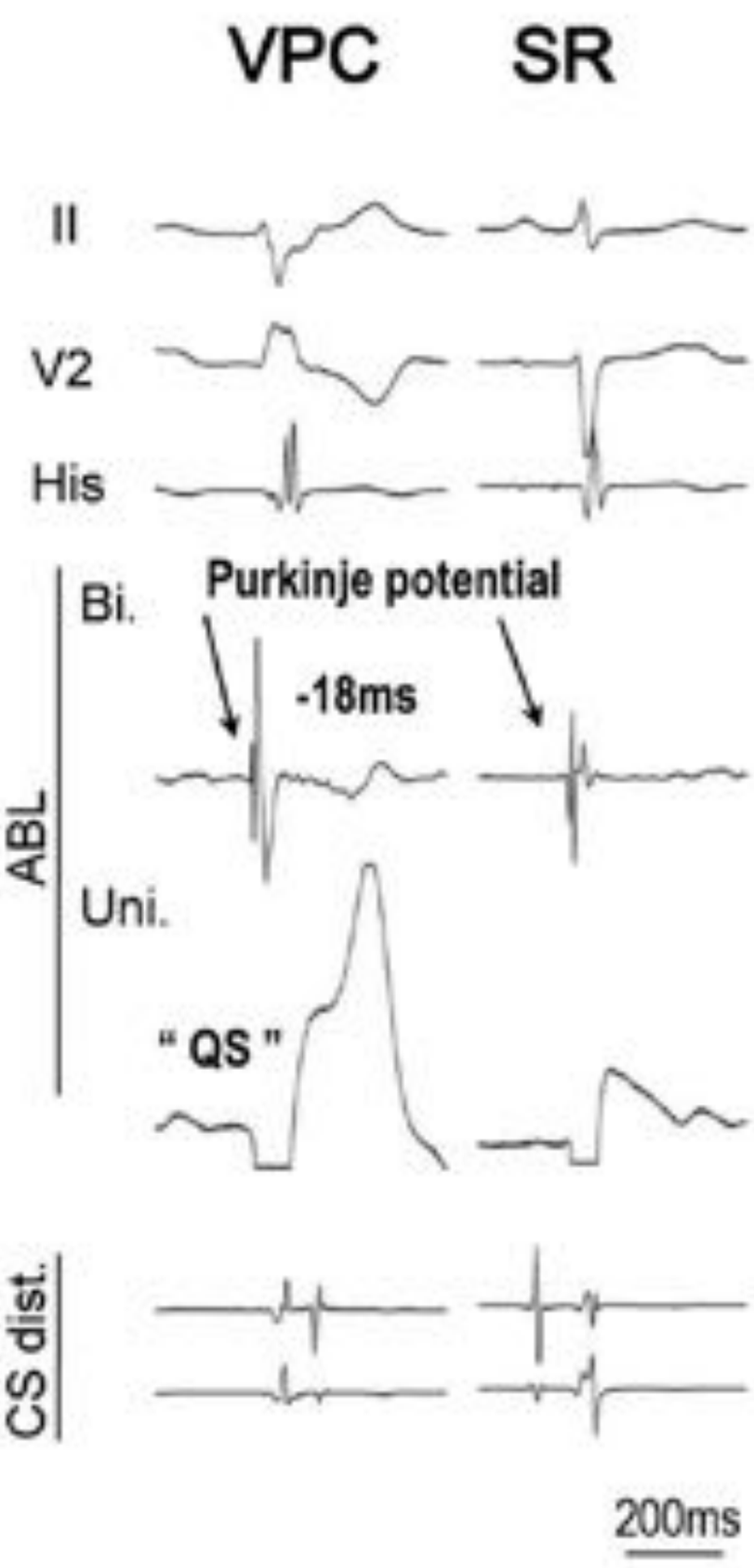
A



B







IVF, short coupled TdP, ablation

Methods

- Ectopy as the target

Mapping and Ablation of Idiopathic Ventricular Fibrillation

Michel Haïssaguerre, MD; Morio Shoda, MD; Pierre Jaïs, MD; Akihiko Nogami, MD; Dipen C. Shah, MD; Josef Kautzner, MD; Thomas Arentz, MD; Dietrich Kalushe, MD; Dominique Lamaison, MD; Mike Griffith, MD; Fernando Cruz, MD; Angelo de Paola, MD; Fiorenzo Gaïta, MD; Mélèze Hocini, MD; Stéphane Garrigue, MD; Laurent Macle, MD; Rukshen Weerasooriya, MD; Jacques Clémenty, MD

Background—Ventricular fibrillation is the main mechanism of sudden cardiac death. The feasibility of eliminating recurrent episodes by catheter ablation has not been reported.

Methods and Results—Twenty-seven patients without known heart disease (13 men, 14 women, 41±14 years of age) were studied after being resuscitated from recurrent (10±12) episodes of primary idiopathic ventricular fibrillation; 23 had received a defibrillator. The first initiating beat of ventricular fibrillation had an identical electrocardiographic morphology and coupling interval (297±41 ms) to preceding isolated premature beats typically noted in the aftermath of resuscitation. These triggers were localized by mapping the earliest electrical activity and ablated by local radiofrequency delivery. Outcome was assessed by Holter and defibrillator memory interrogation. Premature beats were elicited from the Purkinje conducting system in 23 patients: from the left ventricular septum in 10, from the anterior right ventricle in 9, and from both in 4. The interval from the Purkinje potential to the following myocardial activation varied from 10 to 150 ms during premature beat but was 11±5 ms during sinus rhythm, indicating location at peripheral Purkinje arborization. The premature beats originated from the right ventricular outflow tract muscle in 4 patients. The accuracy of mapping was confirmed by acute elimination of premature beats during local radiofrequency delivery. During a follow-up of 24±28 months, 24 patients (89%) had no recurrence of ventricular fibrillation without drug.

Conclusions—Primary idiopathic ventricular fibrillation is a syndrome characterized by dominant triggers from the distal Purkinje system. These sources can be eliminated by focal energy delivery. (***Circulation*. 2002;106:962-967.**)

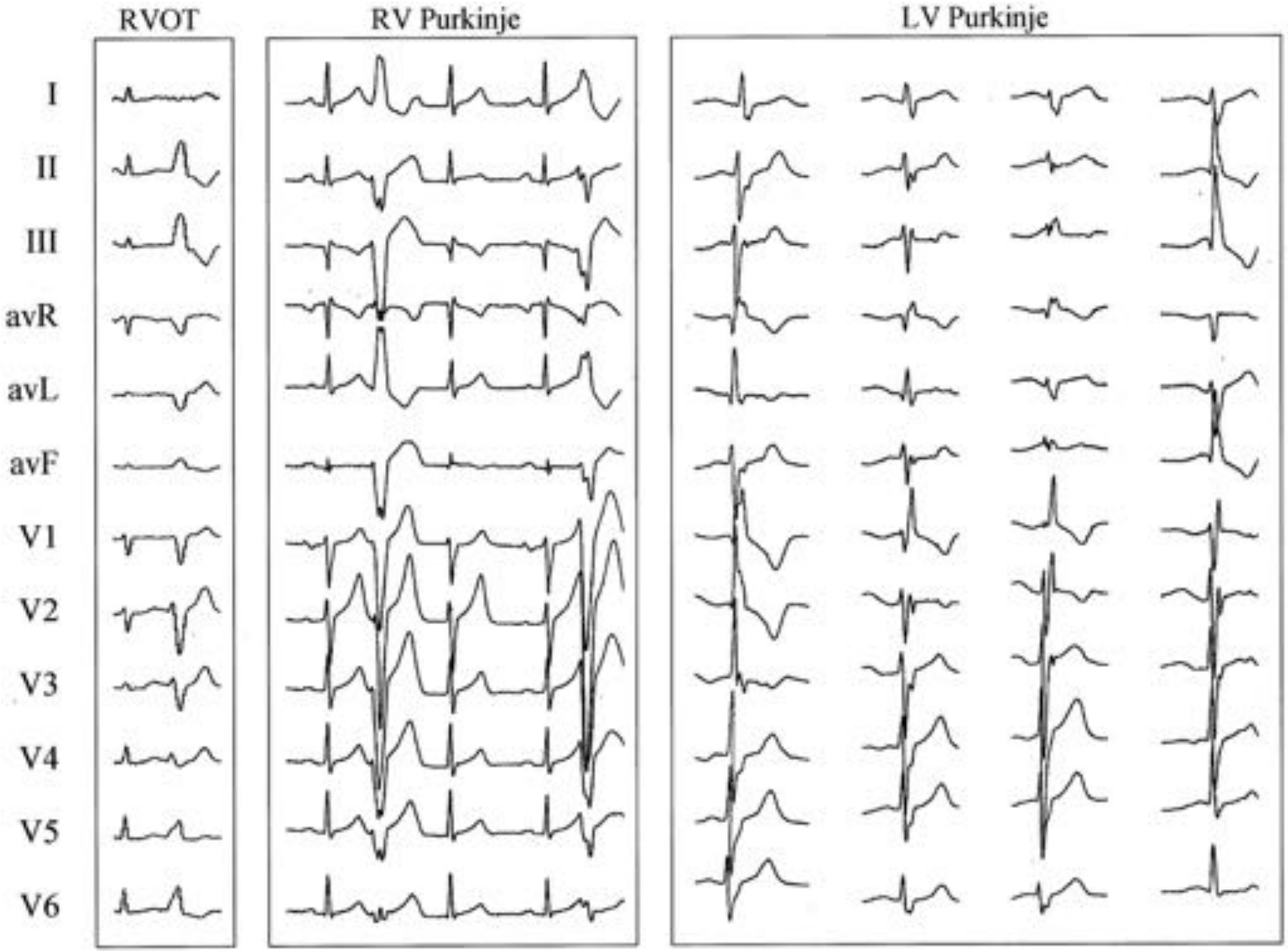
Key Words: ablation ■ death, sudden ■ heart arrest ■ fibrillation ■ mapping

IVF, short coupled TdP, ablation

Methods and Results

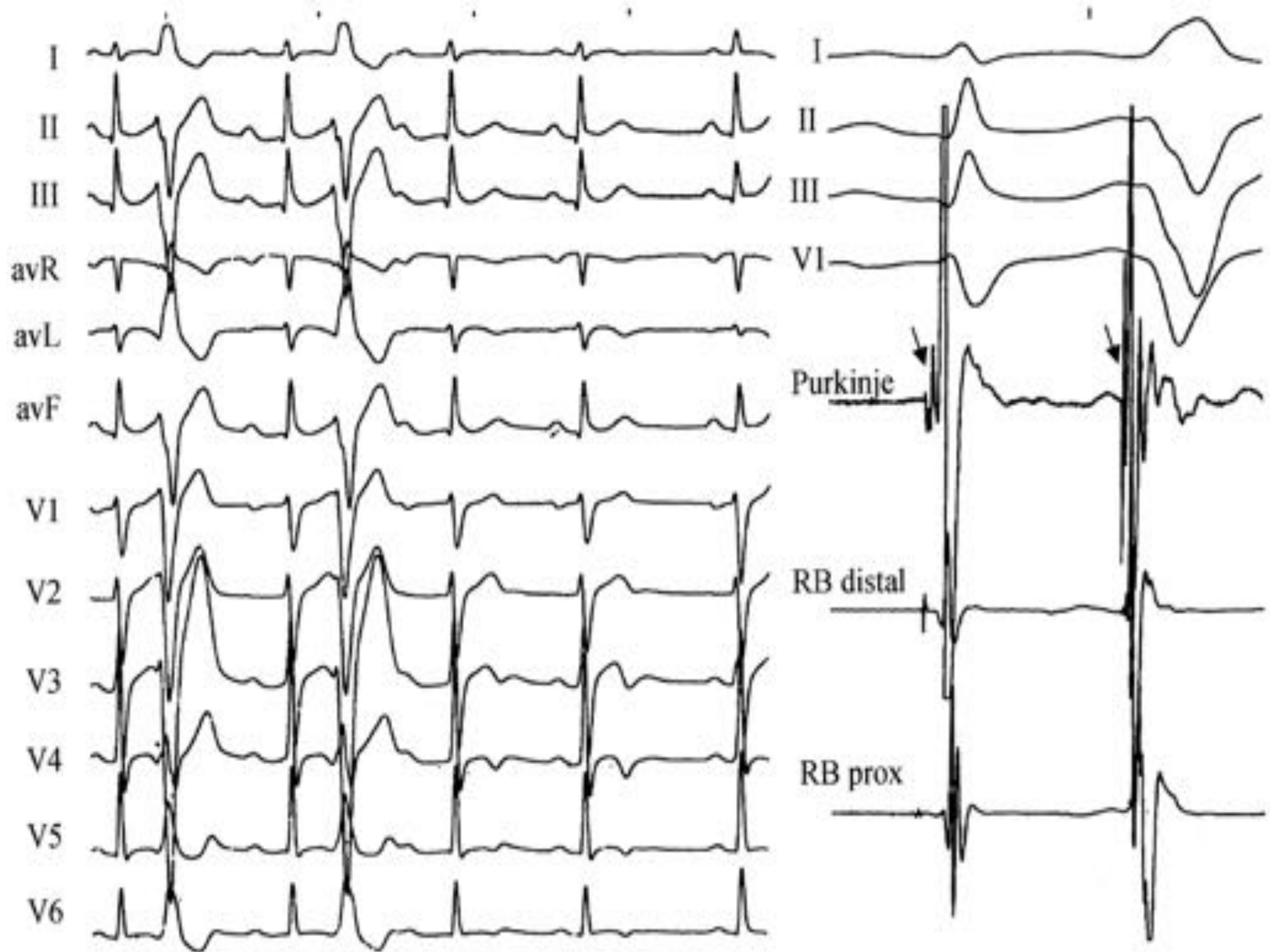
- 23/27 pts ectopy from LV/RV Purkinje
- 4 pts from the RVOT area
- ectopy targeted ablation
- FU 24±28 mths, drug free
- 89% arrhythmia free survival

Electrocardiographic morphology of premature beats



Haissaguerre, M. et al. Circulation 2002;106:962-967

Circulation


I
II
III
avR
avL
avF
V1
V2
V3
V4
V5
V6
I
II
III
VI
Purkinje
RB distal
RB prox

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Long-Term Follow-Up of Idiopathic Ventricular Fibrillation Ablation

A Multicenter Study

Sébastien Knecht, MD,*‡‡ Frédéric Sacher, MD,* Matthew Wright, MBBS, PHD,* Mélèze Hocini, MD,* Akihiko Nogami, MD,† Thomas Arentz, MD,‡ Bertrand Petit, MD,§ Robert Franck, MD,‖ Christian De Chillou, MD,¶ Dominique Lamaison, MD,# Jéronimo Farré, MD,** Thomas Lavergne, MD,†† Thierry Verbeet, MD,‡‡ Isabelle Nault, MD,* Seiichiro Matsuo, MD,* Lionel Leroux, MD,* Rukshen Weerasooriya, MD,* Bruno Cauchemez, MD,§§ Nicolas Lellouche, MD,* Nicolas Derval, MD,* Sanjiv M. Narayan, MD, PHD,* Pierre Jaïs, MD,* Jacques Clementy, MD,* Michel Haïssaguerre, MD*

Bordeaux, Saint Pierre, Paris, Nancy, and Clermond-Ferrand, France; Yokohama, Japan; Bad Krozingen, Germany; Madrid, Spain; and Brussels, Belgium

IVF, short coupled TdP, ablation

Methods and Results

- 38 pts, multicentre study
- ectopy from LV (16) / RV (14) Purkinje
- ectopy targeted ablation
- FU median 63 months
- 7/18 (18%), recurrent VF after 4 mnths
- 5/7 arrhythmia free after re ablation.

Conclusions

ICD's and ablation options

- ♥ are very much disease dependent
- ♥ ICD's should be carefully chosen
- ♥ Ablation options are available
- ♥ in particular in IVF, BrS

Thank you

1. BrS ***is diagnosed*** in patients with ST segment elevation with type 1 morphology ≥ 2 mm in ≥ 1 lead among the right precordial leads V1,V2, 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 V1,V2 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 1 ECG morphology

Class	Therapeutic Intervention Recommendations
Class I	1. ICD implantation **is recommended** in symptomatic patients with a diagnosis of SQTS who a) Are survivors of a cardiac arrest and/or b) Have documented spontaneous sustained VT with or without syncope

Class	Therapeutic Intervention Recommendations
Class I	1. ICD implantation **is recommended** in symptomatic patients with a diagnosis of SQTS who a) Are survivors of a cardiac arrest and/or b) Have documented spontaneous sustained VT with or without syncope
Class IIb	2. ICD implantation **may be considered** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD. 3. Quinidine **may be considered** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD. 4. Sotalol **may be considered** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD

Indications for ICDs in Patients Diagnosed with Short QT