HOW TO MANAGE PATIENTS WITH *EARLY REPOLARIZATION* ?

Sami Viskin, M.D.
Tel Aviv Medical Center
Israel.

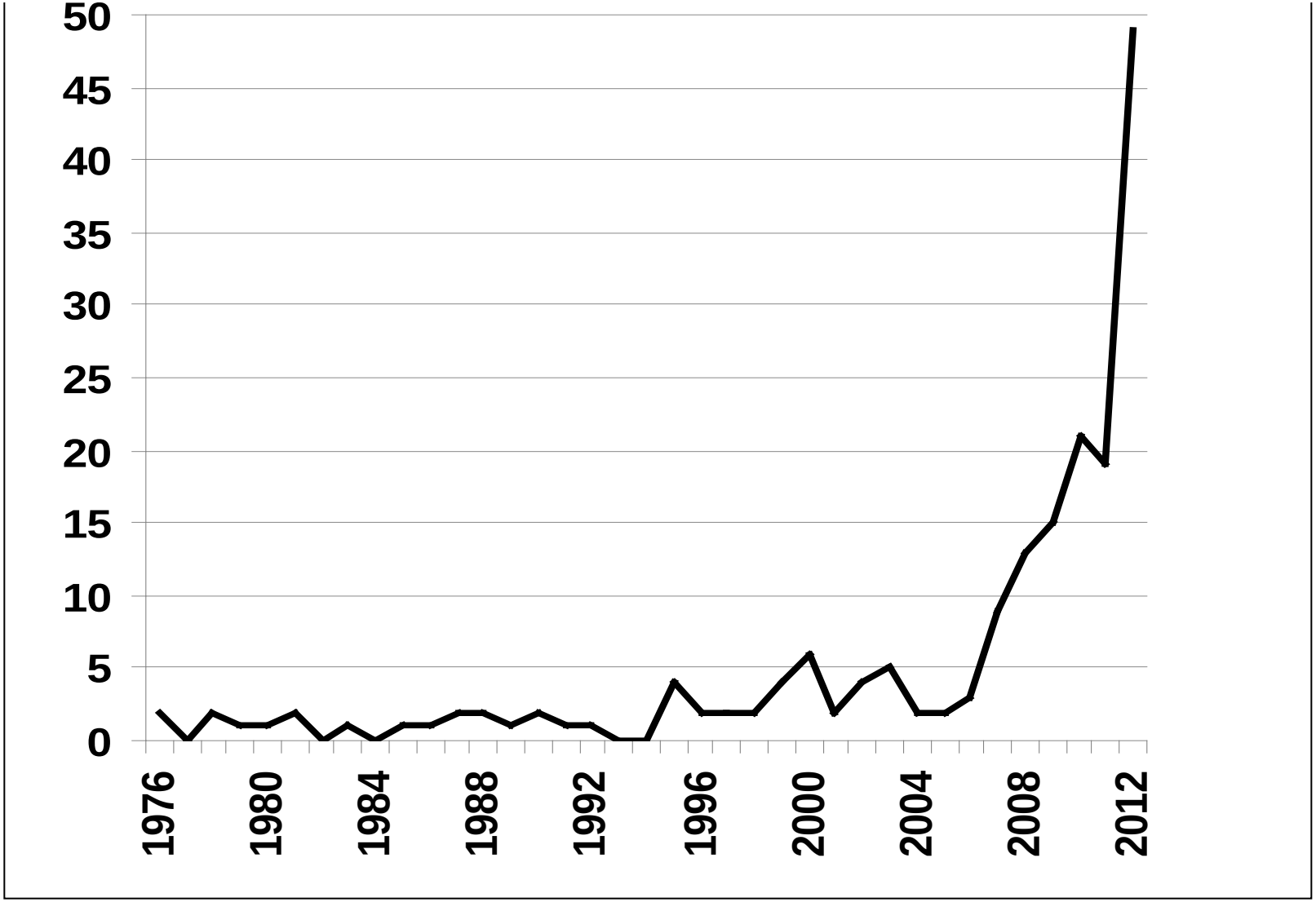
DISCLOSURES:

More people live off J-waves than die from them.

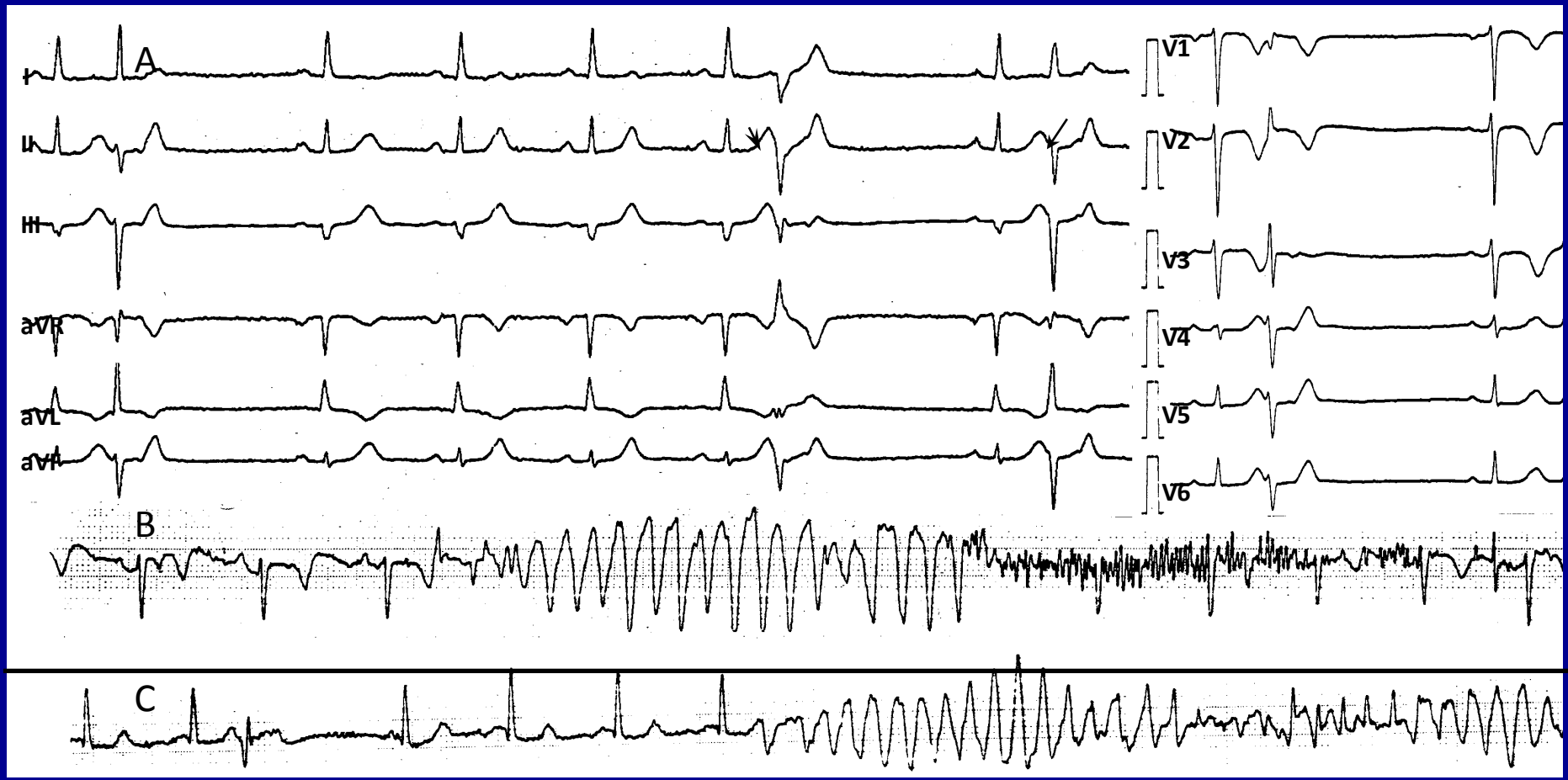
I belong to the first category….

And would like to keep things that way.

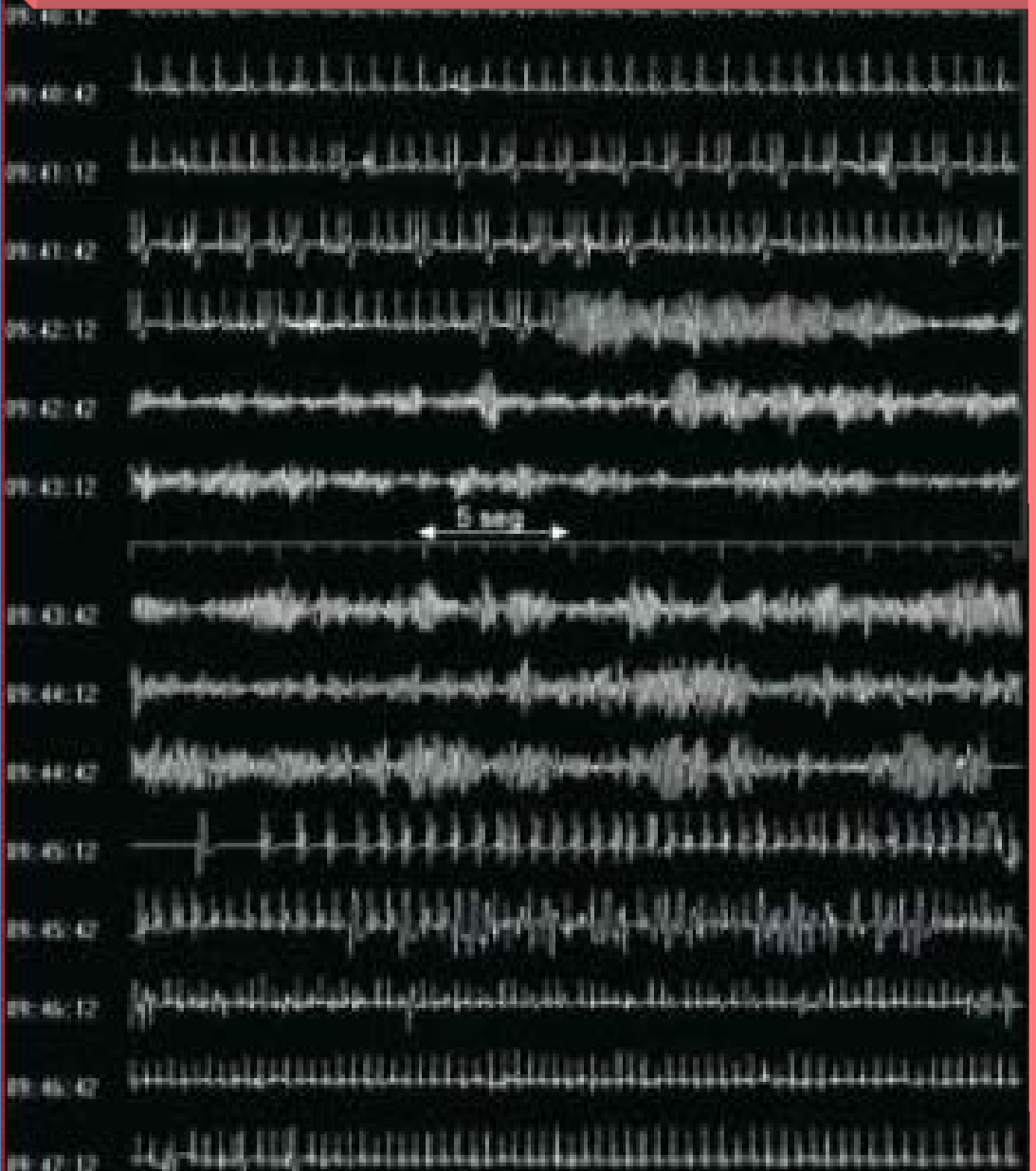
Publications with the terms "early repolarization" or "J-waves" in the title.



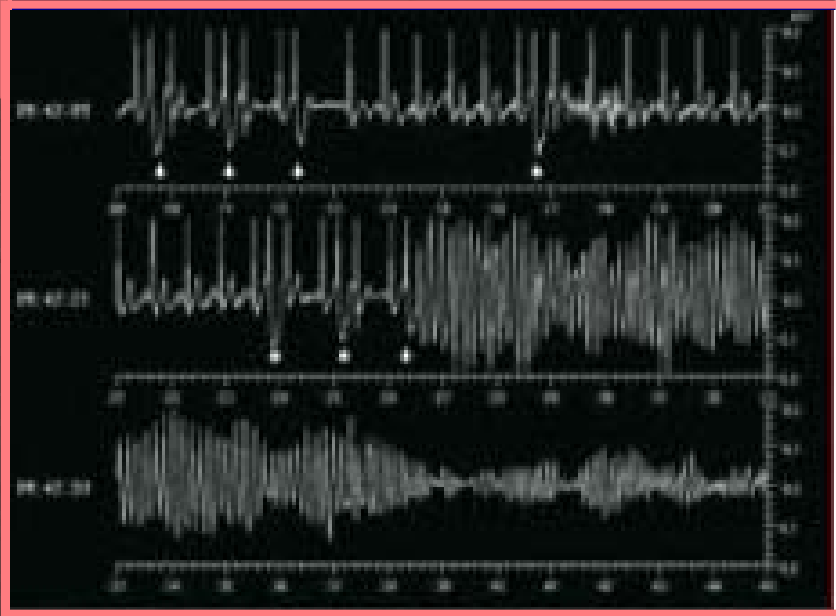
The symptomatic patient

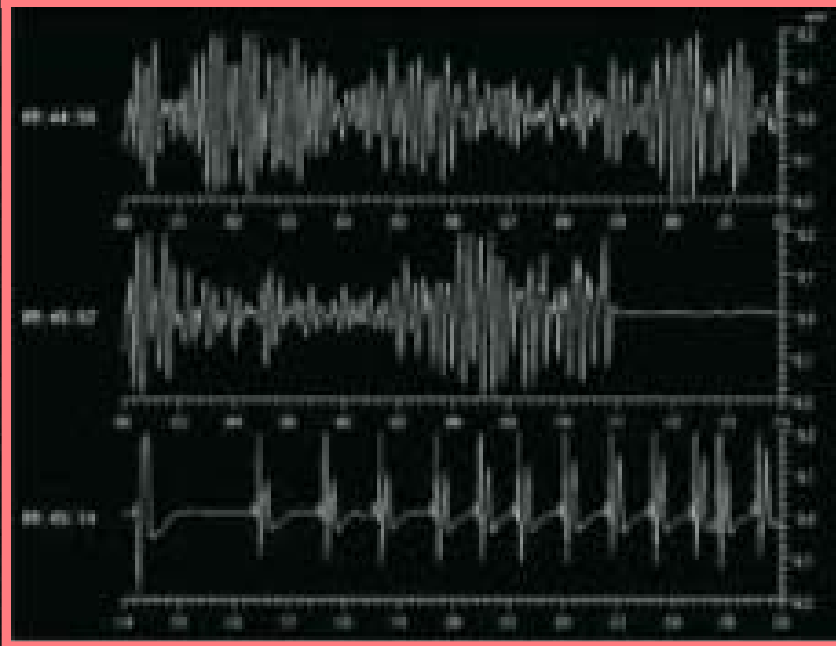


Spontaneous termination of VF after 3 minutes



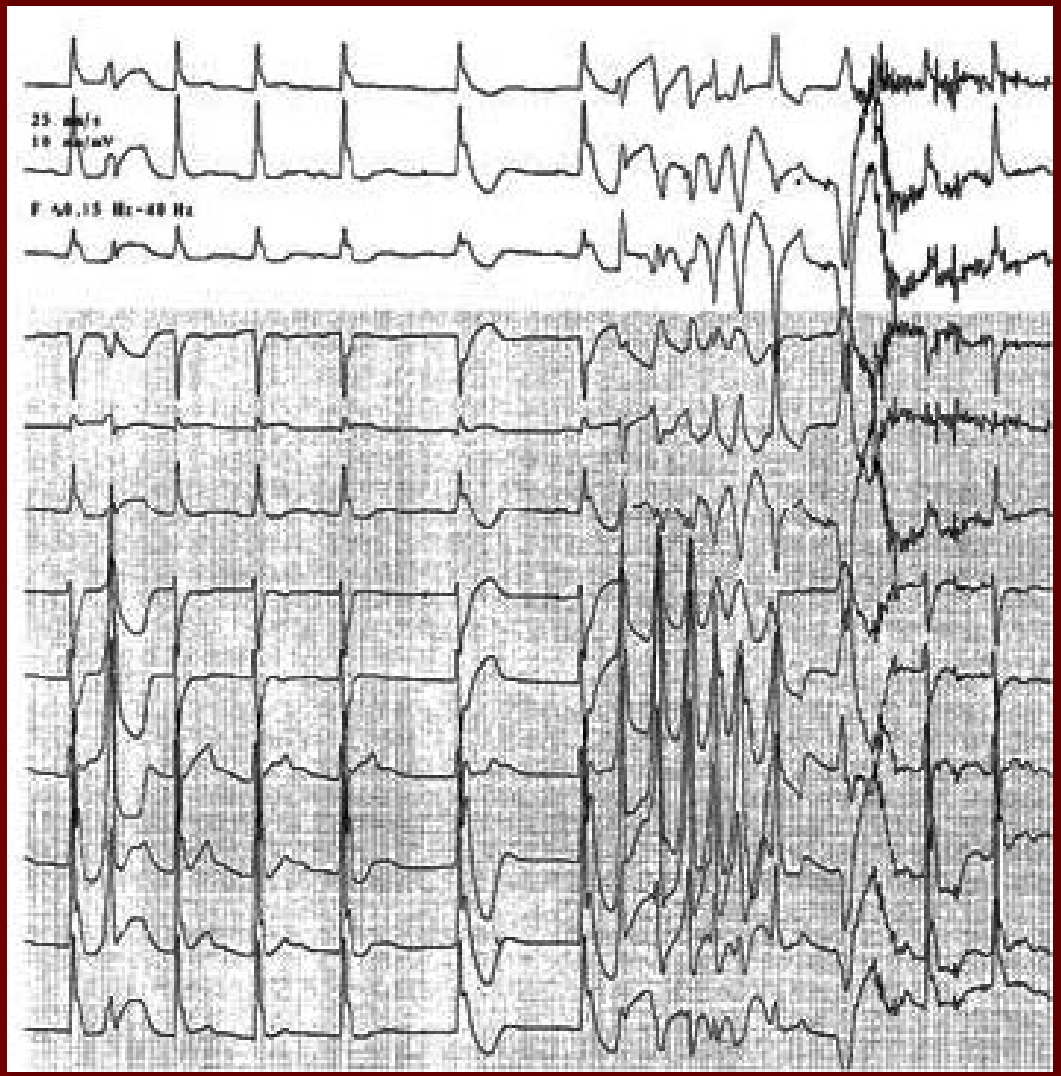
REVISTA ESPAÑOLA DE CARDIOLOGIA

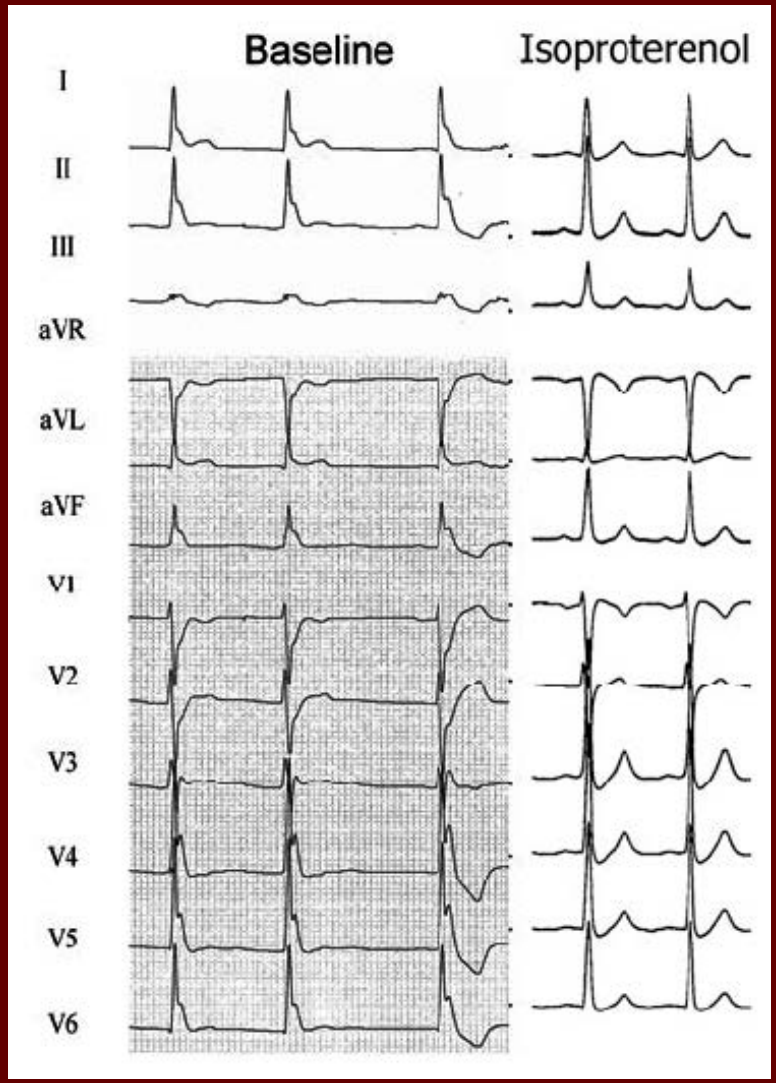




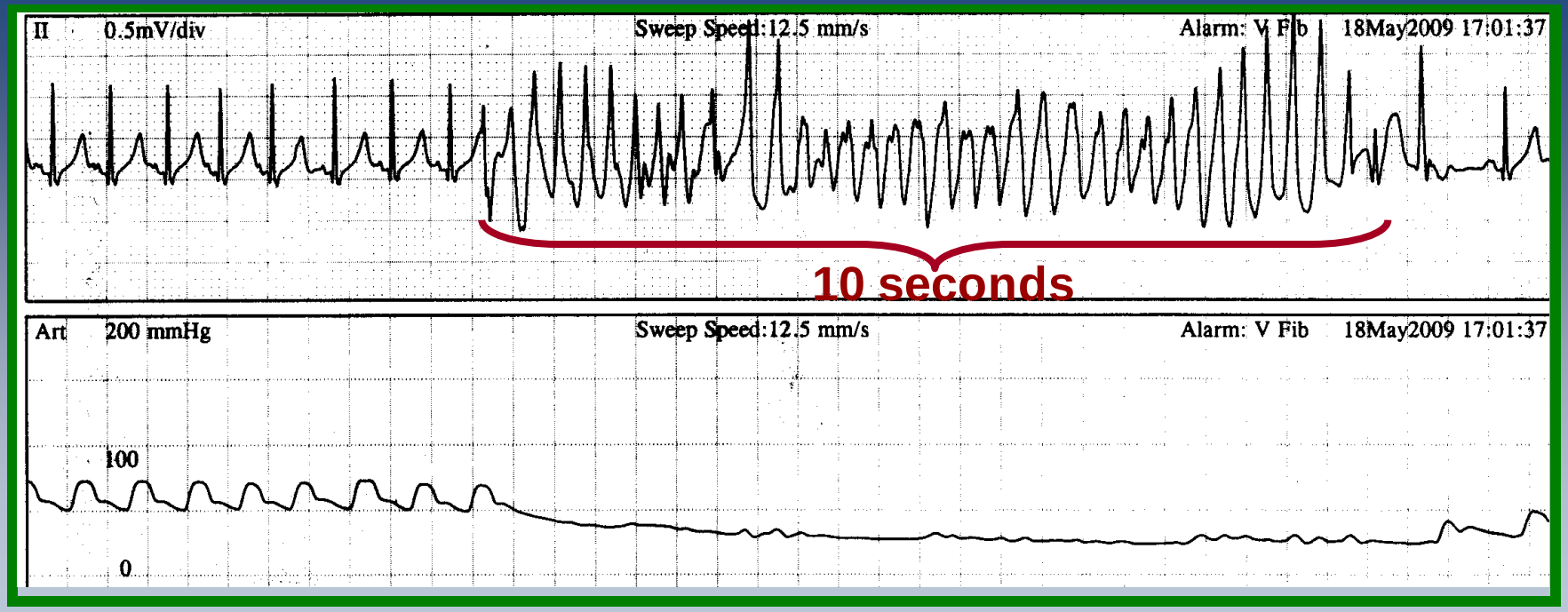
Pedrote A, 2009.

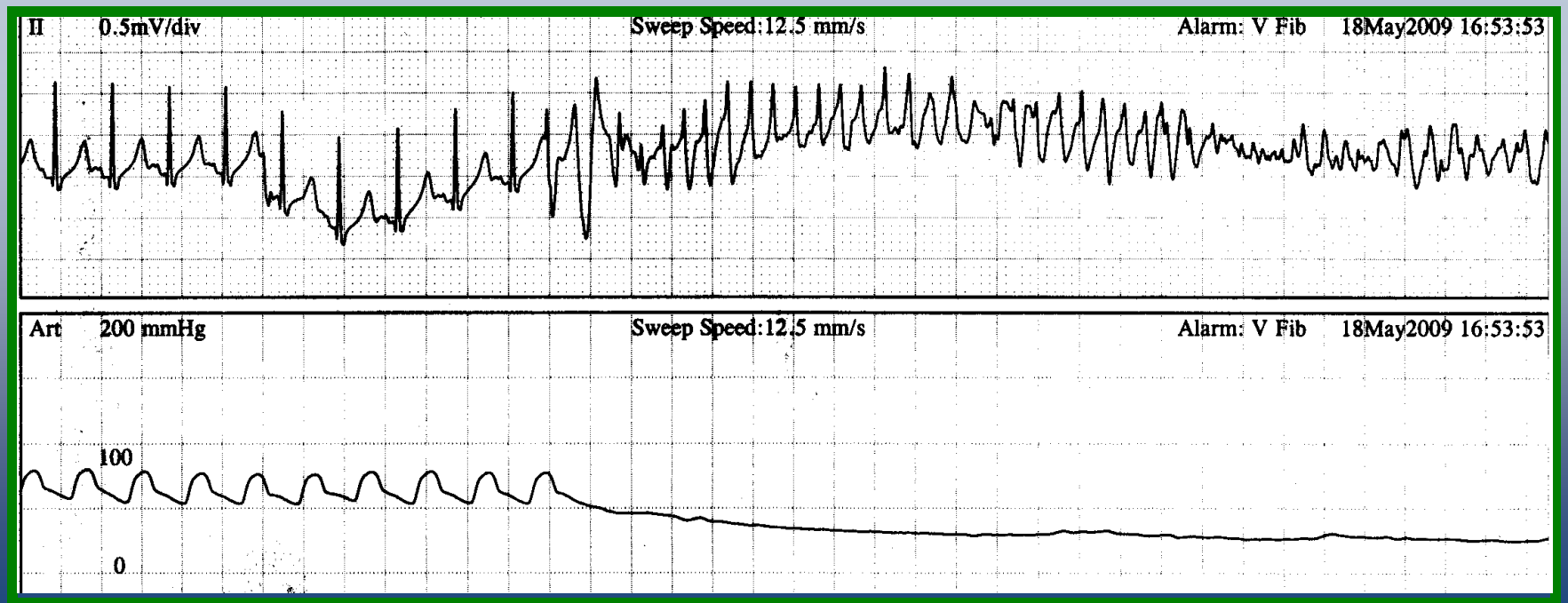
Abolition of J-wave and arrhythmias with isoproterenol



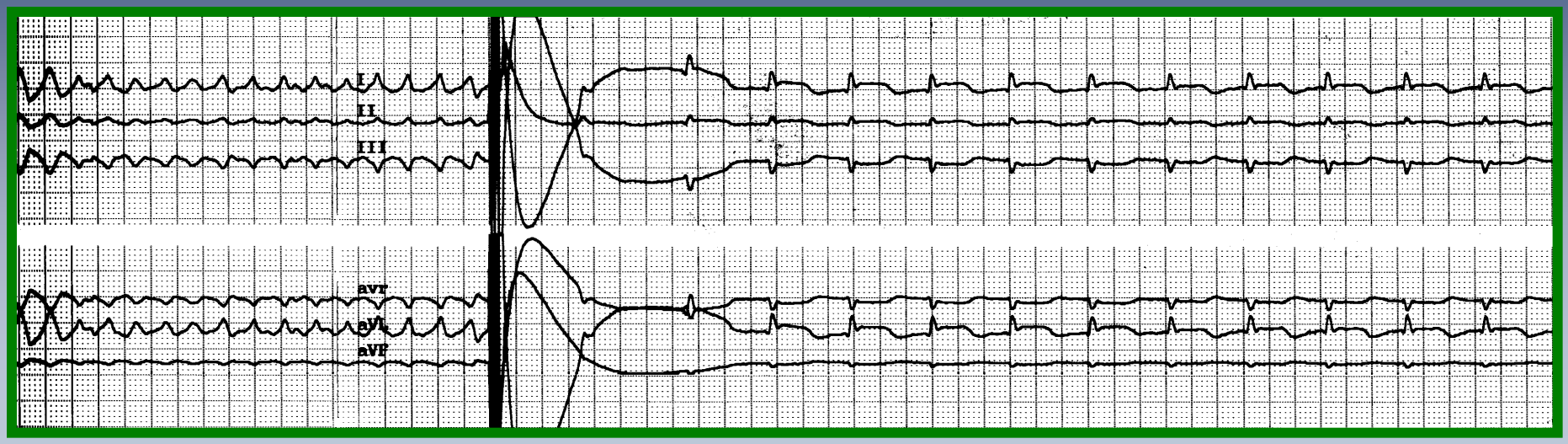


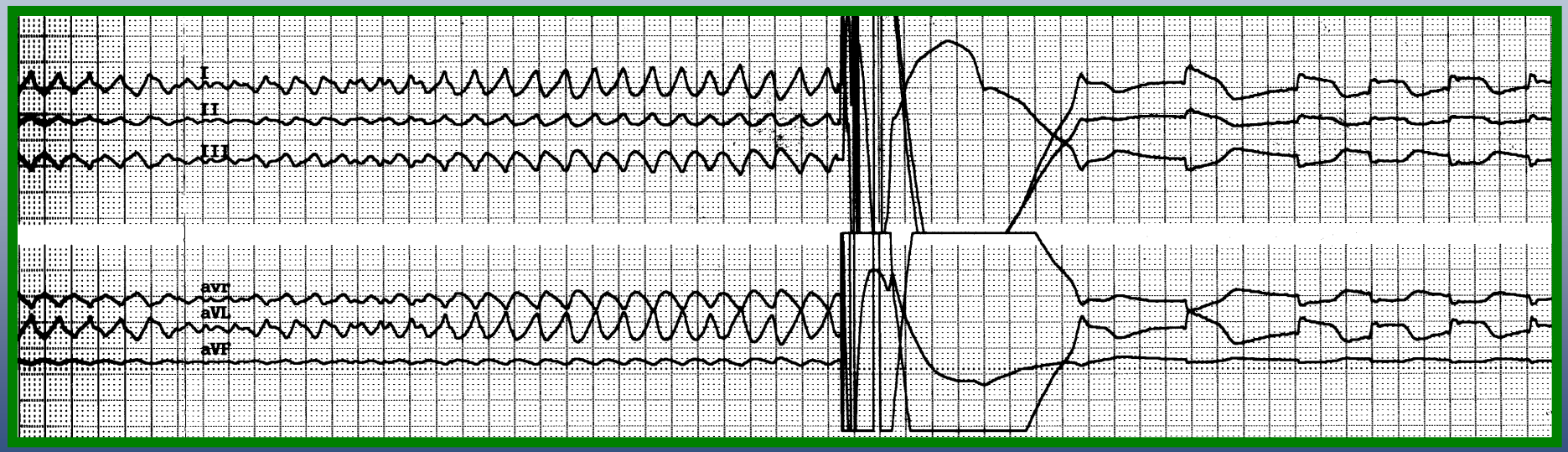
Haissaguerre, JCE *2008*

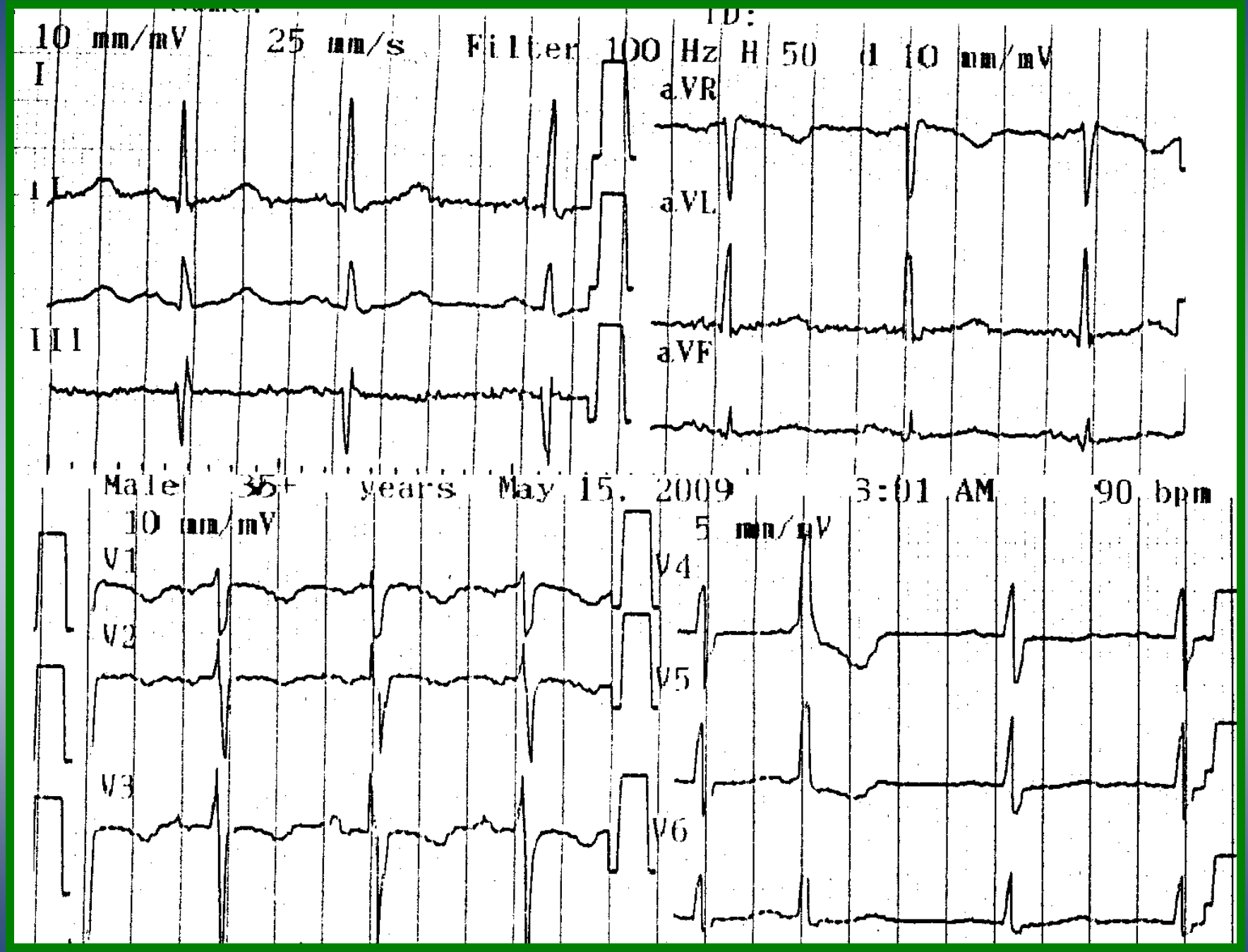

II 0.5mV/div
Sweep Speed:12.5 mm/s
Alarm: V Fib
18May2009 17:01:37
10 seconds
Art 200 mmHg
Sweep Speed:12.5 mm/s
Alarm: V Fib
18May2009 17:01:37
100
0


II 0.5mV/div
Sweep Speed:12.5 mm/s
Alarm: V Fib
18May2009 16:53:53
Art 200 mmHg
Sweep Speed:12.5 mm/s
Alarm: V Fib
18May2009 16:53:53
100
0

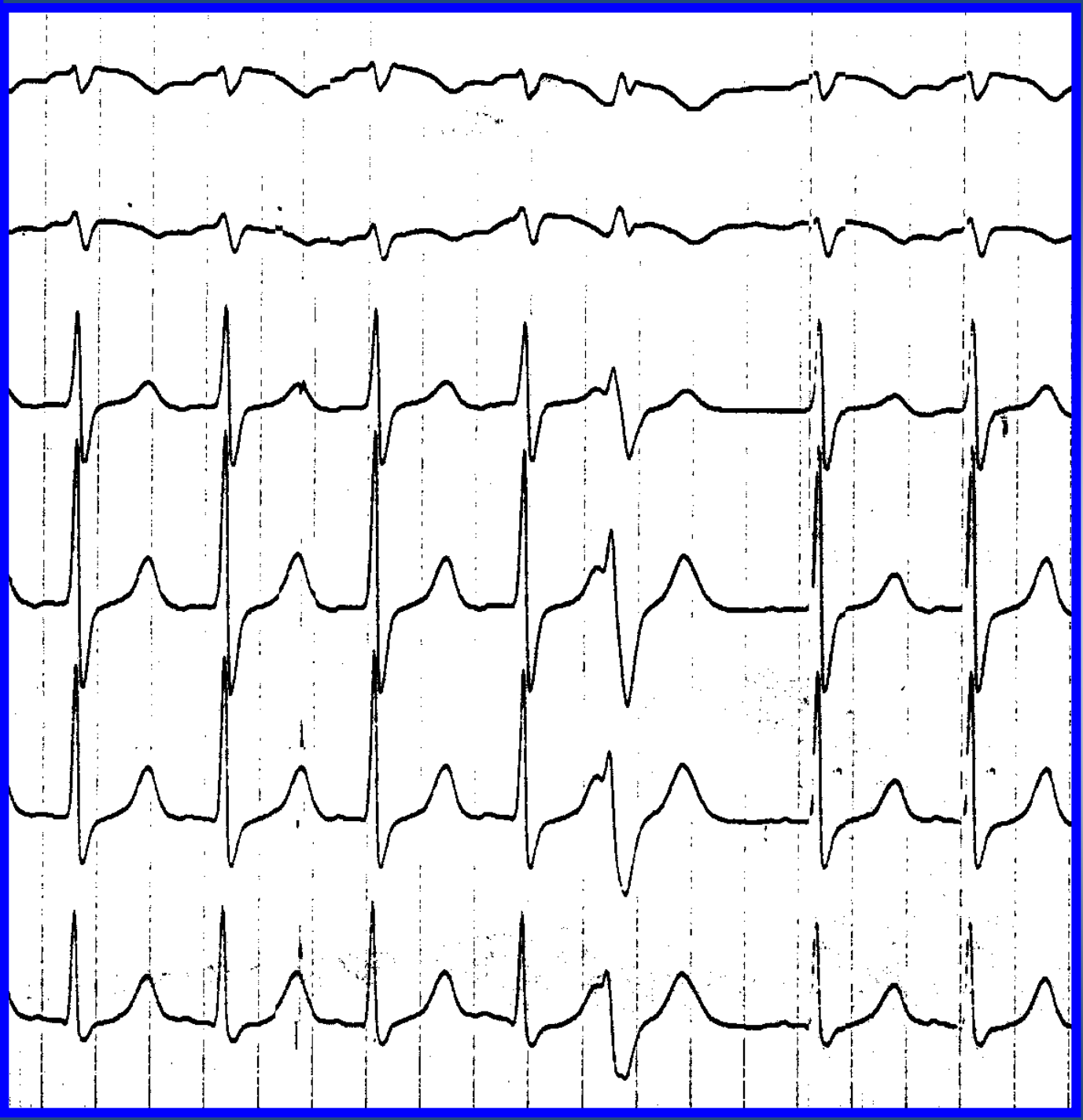
VF storm: 28 VF episodes in 3 hours






10 mm/mV
25 mm/s
Filter 100 Hz H 50
10 mm/mV
I
II
III
aVR
aVL
aVF
Male 35+ years May 15, 2009 3:01 AM 90 bpm
10 mm/mV
5 mm/mV
V1
V2
V3
V4
V5
V6

One beat,
one diagnosis.



VF despite ventricular pacing at 125 beats/min.

Moe, *Am Heart J* 1949

MORGAGNI-ADAMS-STOKES ATTACKS CAUSED BY TRANSIENT RECURRENT VENTRICULAR FIBRILLATION IN A PATIENT WITHOUT APPARENT ORGANIC HEART DISEASE

A Case Report

Torjus Moe, M.D.

VENTRICULAR fibrilla
sudden death in man

Kontny, *J Intern Med* 1990

Case report

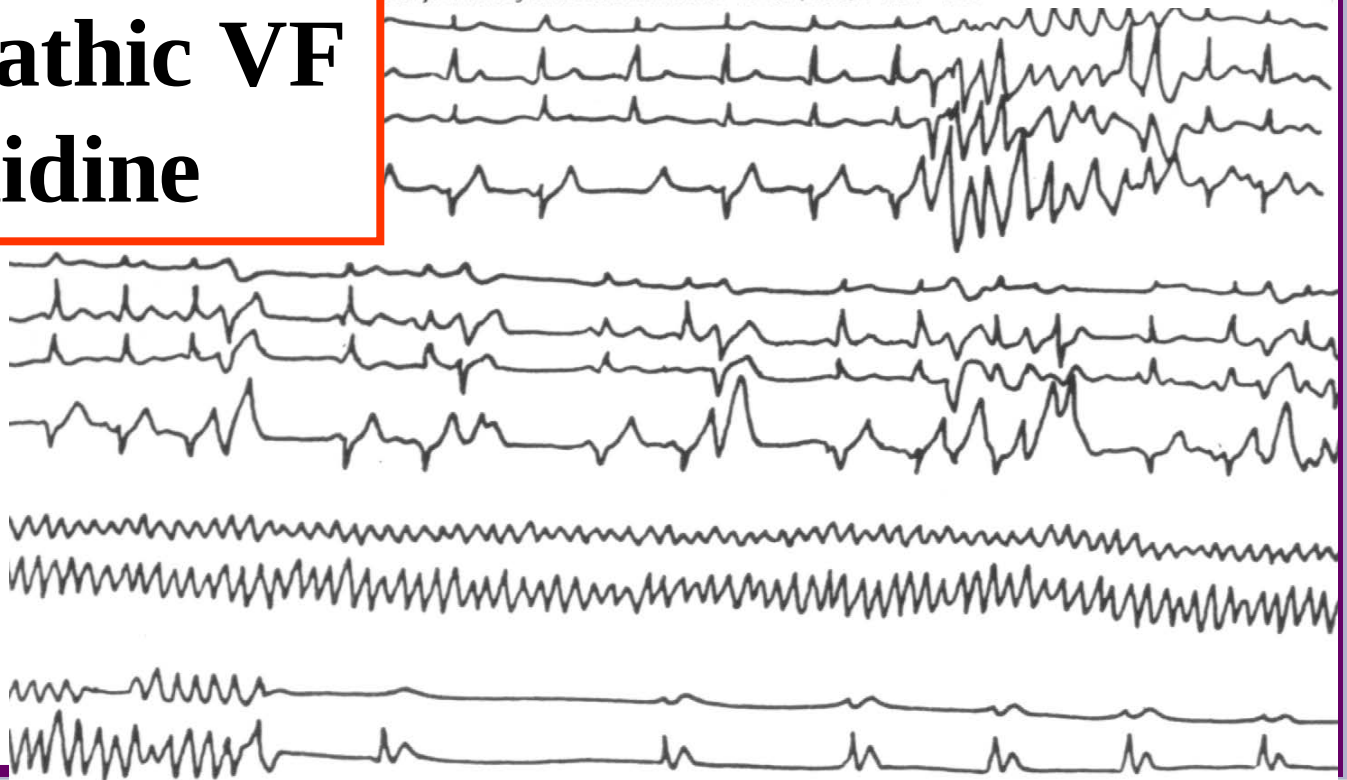
Self-terminating idiopathic ventricular fibrillation presenting as syncope: a 40-year follow-up report

F. KONTNY & J. DALE

the Medical Department, Aker Hospital, Oslo, Norway

tract. Kontny F., Dale J (Medical Department, Aker Hospital, Oslo, Norway). Self-inating idiopathic ventricular fibrillation presenting as syncope: a 40-year follow-up rt. *Journal of Internal Medicine* 1990; **227**: 211–13.

40 years of follow-up of idiopathic VF on quinidine



Reprinted from AMERICAN HEART JOURNAL St. Louis
Vol. 120, No. 3, pp. 661-671, September, 1990, (Printed in the U.S.A.)
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Idiopathic ventricular fibrillation

Sami Viskin, MD,[a] and Bernard Belhassen, MD. *Tel-Aviv, Israel*

Reprinted from AMERICAN HEART JOURNAL St. Louis
Vol. 120, No. 3, pp. 661-671, September, 1990, (Printed in the U.S.A.)
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Am Heart J 1990

Idiopathic ventricular fibrillation

Sami Viskin, MD,[a] and Bernard Belhassen, MD. *Tel-Aviv, Israel*

Ventricular fibrillation (VF) is the most common arrhythmia documented at the time of sudden cardiac death.[1-3] The major underlying etiology is by far atherosclerotic coronary artery disease; however, many other causes, such as valvular heart disease, congenital cardiac anomalies, accessory pathways, and idiopathic or secondary cardiomyopathies, have also been identified.[4-6] VF without obvious cardiac pathology also occurs in the long QT syndrome.[7,8] Recently, VF has been documented as the cause of death in the "syndrome of nocturnal sudden death in men from Southeast Asia."[9]

The list of potential causes for sudden cardiac death continues to be expanded in detail.[10-12] Yet notwithstanding the difficulty of defining sudden cardiac death,[5] no underlying etiology is found to 8% of patients dying suddenly[13] survivors of out-of-hospital cardiac arrest[14] extensive evaluation. [illegible] crease in selected [illegible] forensic [illegible] old) [illegible]

[illegible] literature on this [illegible] is the first systematic review of idiopathic VF, a disorder mentioned only briefly or totally ignored[5, 30, 31] in textbooks and reviews.

METHODOLOGY

cardiac or noncardiac [illegible] citation or whose [illegible] Positive identification [illegible] by a paramedic or [illegible] diac arrest" or "ventricular [illegible] when these episodes [illegible] mination, was a requirement.

Patients were excluded [illegible] reported in any of the [illegible] diac physical examination [illegible] agnostic of acute myocardial [illegible] of any recognized metabolic cause for arrhythmogenic sudden death; (3) resting electrocardiogram and exercise test (except for arrhythmias); (4) chest roentgenogram (if relevant to cardiac pathology); [illegible] phy; (6) radionuclide ventriculography; [illegible] catheterization and coronary angiography; [illegible] electrophysiologic studies (if evidence of [illegible] over an accessory pathway [illegible] considered minimal for inclusion. [illegible] sports consisted of clinical and laboratory assessment and 12-lead electrocardiogram in addition to (1) echocardiography and cardiac catheterization with coronary angiography or (2) postmortem examination. All patients included, however, had additional tests (Table I). Altogether, only four patients without coronary angiographic evaluation were nevertheless included: one who died 1 year after VF diagnosis and had a normal postmortem examination[32]; two patients (both younger than 25 years and with negative exercise stress test results) for whom coronary angiography was not considered clinically indicated[33, 34]; and one of our own patients [illegible] remained [illegible] in response [illegible] 5-year [illegible] patients for [illegible] reasonably [illegible] with this [illegible] criteria [illegible] syndrome" [illegible] (1) [illegible] to be [illegible] in any [illegible] provocation

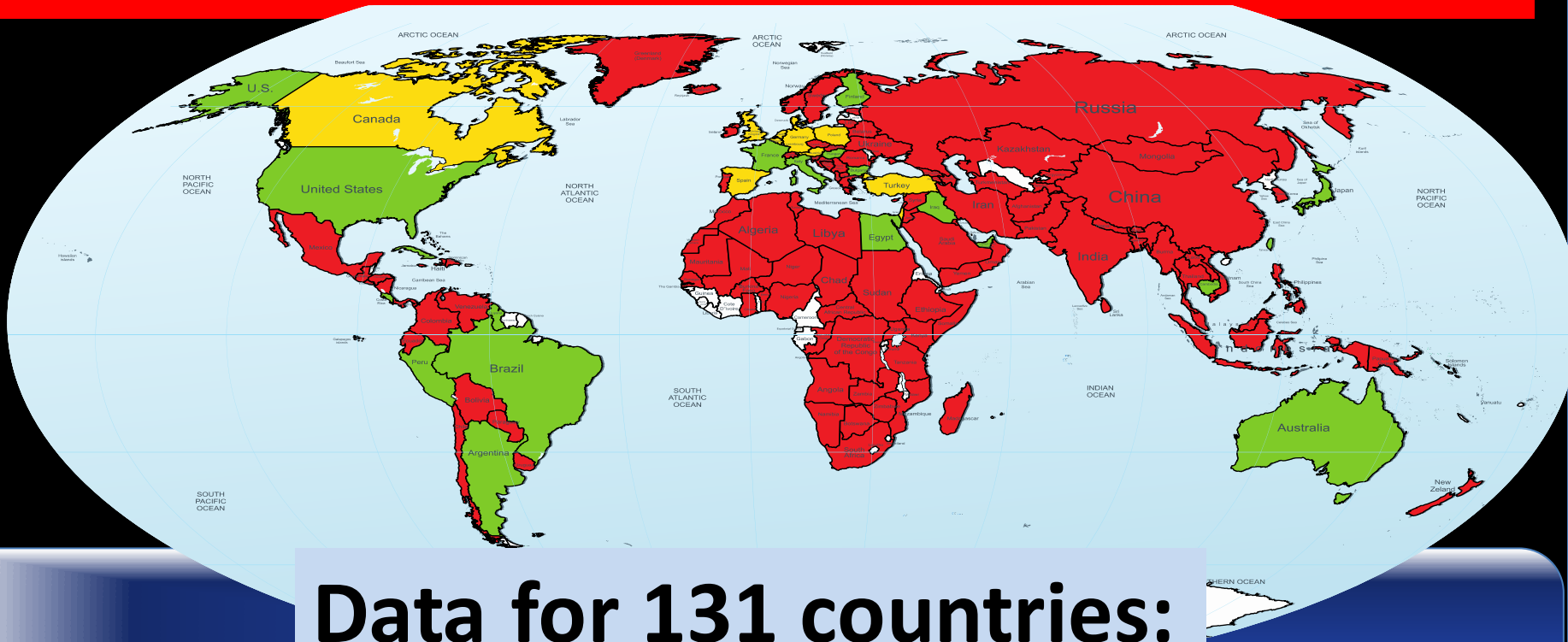
VF-storm !!

Excellent response to quinidine

Males ≈ females.
Age 20 – 65 *(35-45)*.

Syncope/cardiac arrest:
At rest while awake
Not related to effort

Quinidine is inaccessible in many countries



Data for 131 countries:

Yes = 14%, **No = 76%** **With restrictions = 10%**

Viskin, *Rejected from N Engl J Med* 2012.

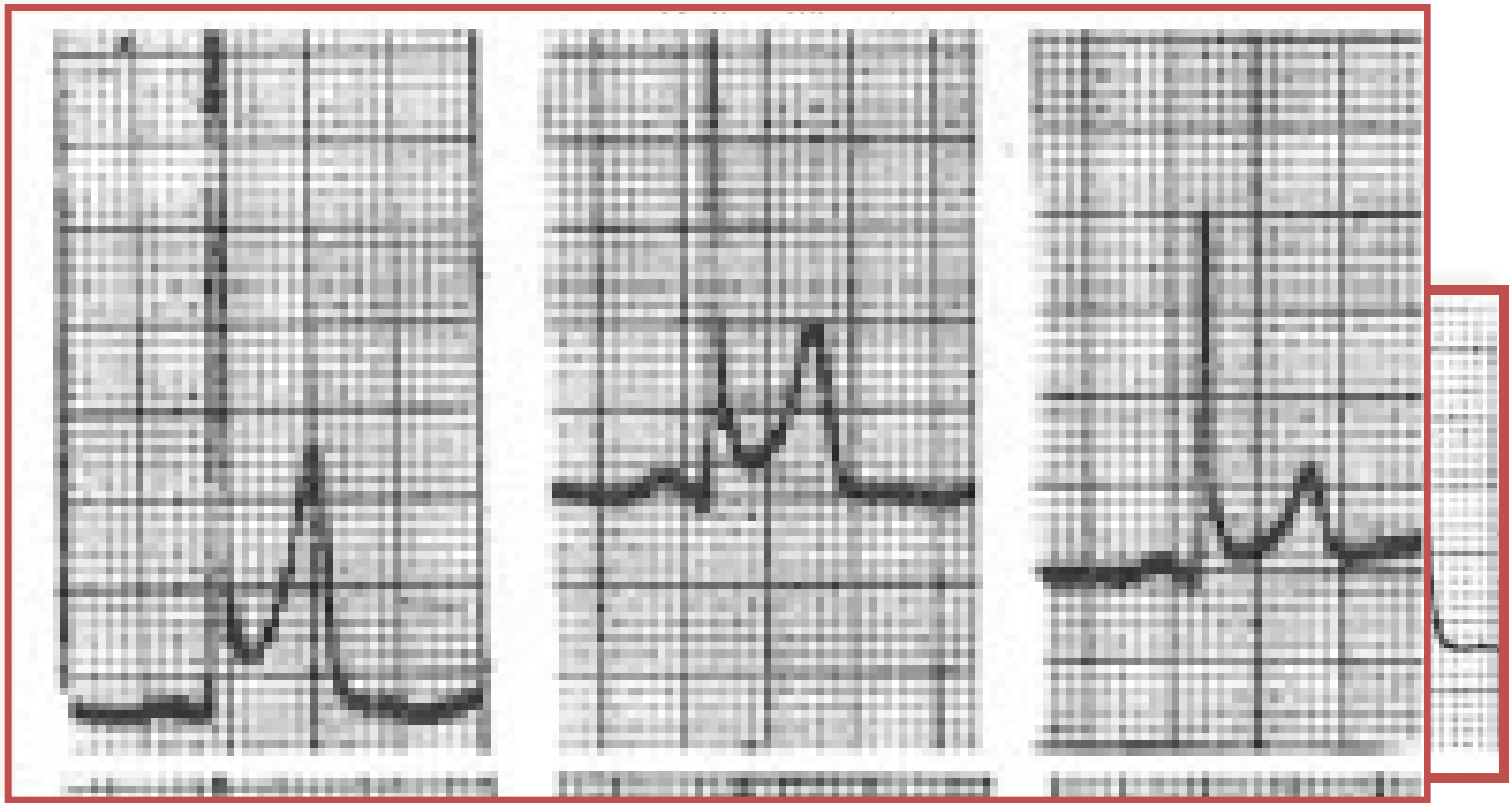
How to manage the patient with ASYMPTOMATIC early repolarization

Early repolarization: a package deal…

American Journal of Cardiology, 1961

The Normal RS-T Segment Elevation Variant*

RICHARD H. WASSERBURGER, M.D. *and* WILLIAM J. ALT,† M.D. WITH THE TECHNICAL ASSISTANCE OF CAROLINE J. LLOYD



The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE ARCHIVE

Differential Characteristics of the Electrocardiogram in Early Repolarization and Acute Pericarditis

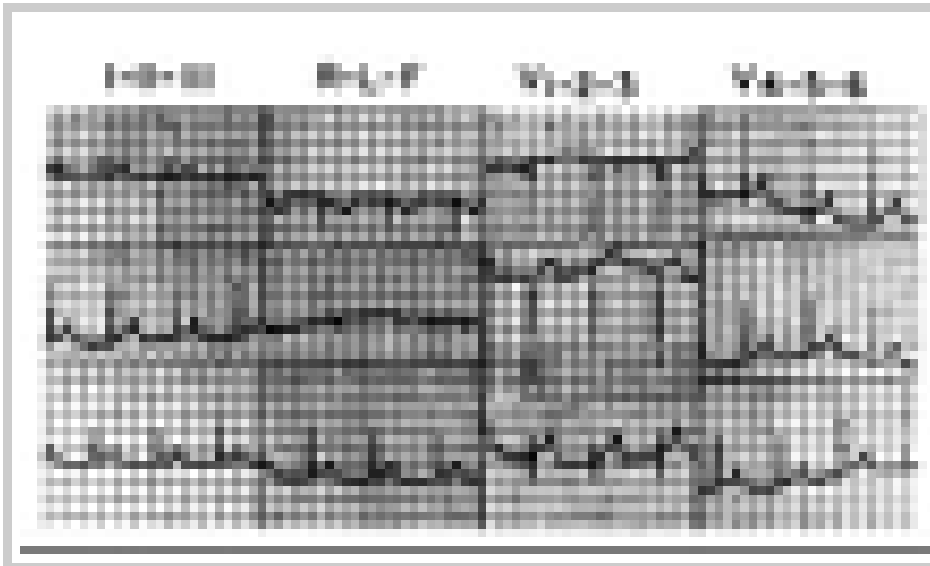
David H. Spodick, M.D.

N Engl J Med 1976; 295:523-526

September 1976

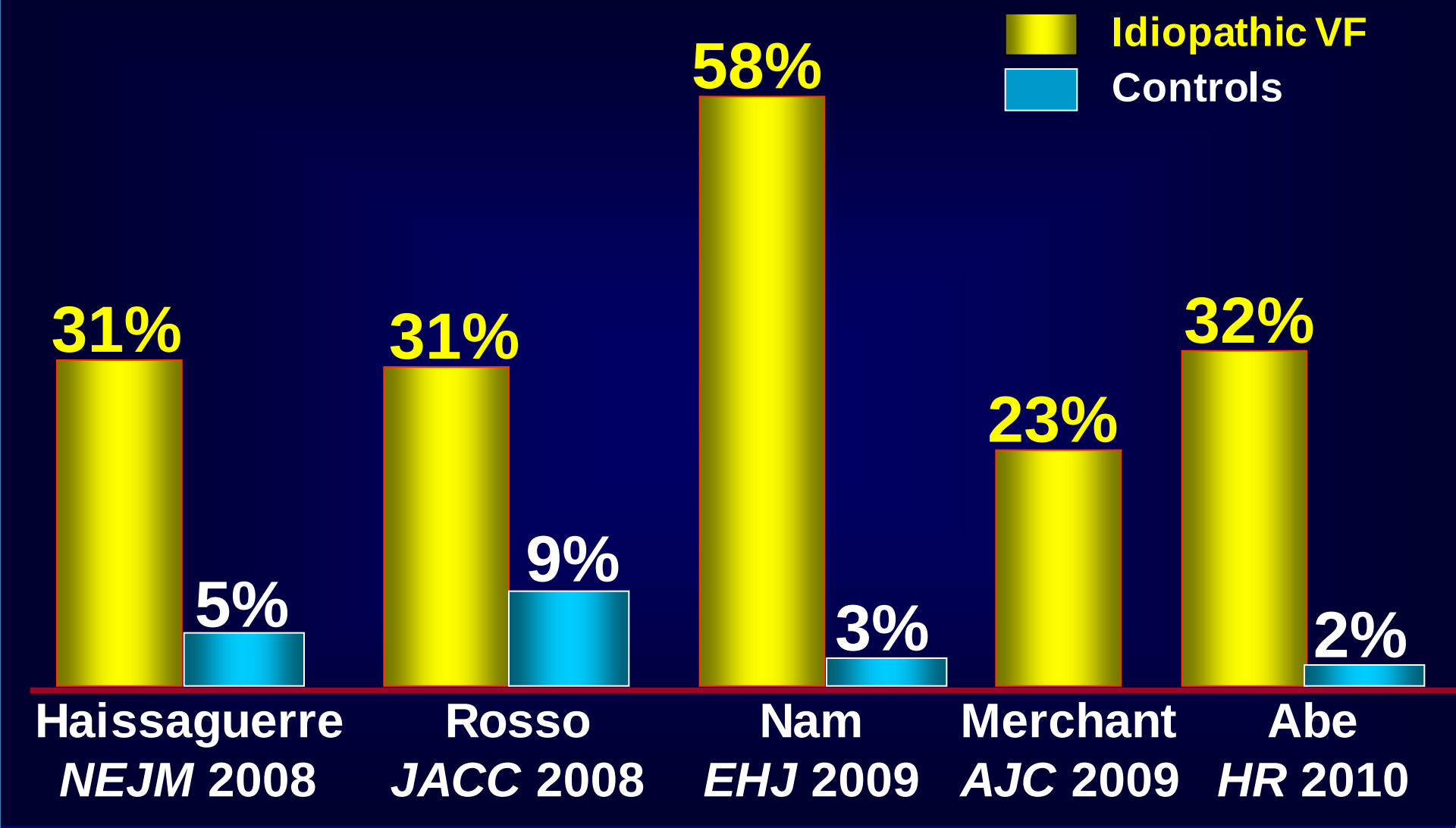
Abstract

Electrocardiographic mimicry necessitates differen
acute pericarditis and early repolarization. Among
lead RS-T deviations occurred in all 48 with perica
with early repolarization. RS-T vectors (Â RS-T) in
to be horizontal (25 patients) and left of the T vecto
patients); in early repolarization, Â RS-T was vertic
and right of Â T (20 subjects); P≤0.01. RS-T depre
was more common in pericarditis (14 vs. two); isoe
Lead V_6 was more common in early repolarization
P≤0.01. PR segment deviations occurred in both li
leads in pericarditis; in early repolarization they we
either lead group. Thus, R-ST deviations in both lin
leads with horizontal Â RS-T to left of Â T and RS-
Lead V_1 favor pericarditis; vertical Â RS-T and isoe
Lead V_6 favor early repolarization. (N Engl J Med 2



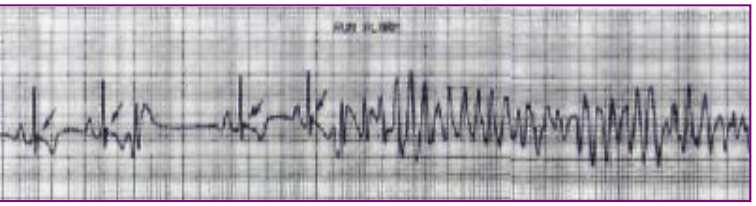
THINGS ARE NOT ALWAYS WHAT THEY SEEM

Prevalence of Early Repolarization in Patients with Idiopathic VF and controls.



Ventricular Fibrillation in a Patient with Prominent J (Osborn) Waves and ST Segment Elevation in the Inferior Electrocardiographic Leads: A Brugada Syndrome Variant?

HARI KALLA, M.D., GAN-XIN YAN, M.D., PH.D., and ROGER MARINCHAK, M.D.
From Lankenau Hospital, Wynnewood, Pennsylvania

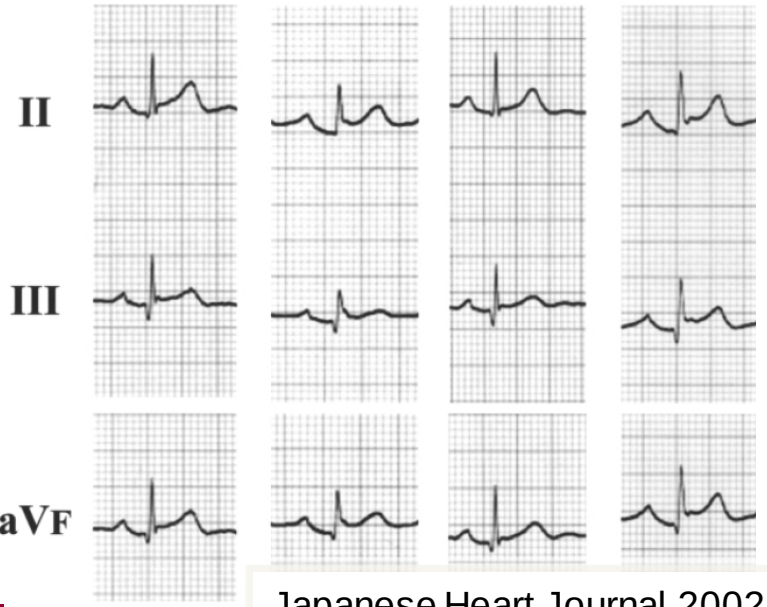


J Cardiovasc Electrophysiol 2000

J Wave and ST Segment Elevation in the Inferior Leads

A Latent Type of Variant Brugada Syndrome ?

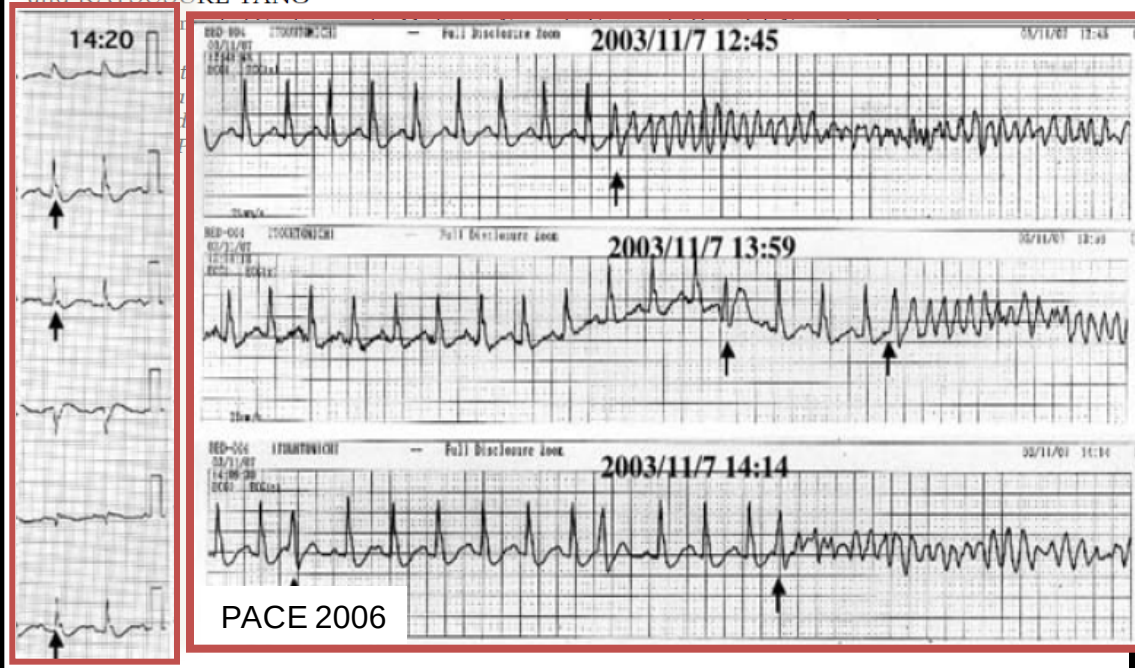
Makoto SAHARA,[1] MD, Kouichi SAGARA,[1] MD, Takeshi YAMASHITA,[1] MD, Tsuyoshi ABE,[1] MD, Hajime KIRIGAYA,[1] MD, Misao NAKADA,[1] MD, Hiroyuki IINUMA,[1] MD, Long-Tai FU,[1] MD, *and* Hiroshi WATANABE,[1] MD



Japanese Heart Journal 2002

Ventricular Fibrillation in a Patient with Prominent J Wave in the Inferior and Lateral Electrocardiographic Leads After Gastrostomy

NORIHIRO KOMIYA, RYO IMANISHI, HIROAKI KAWANO, RIYAKO SHIBATA, MANABU MORIYA, SATOKI FUKAE, YOSHIYUKI DOI, KOJIRO NAKAO, SHINJI SETO, and KATSUSUKE YANO

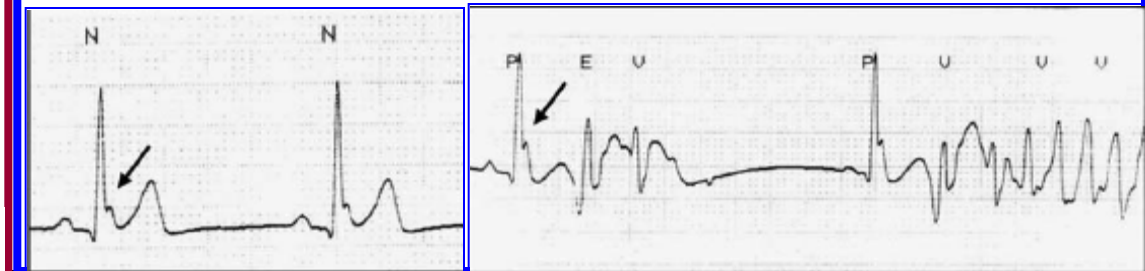


PACE 2006

Characterization of J wave in a patient with idiopathic ventricular fibrillation

HeartRhythm 2006

Tetsuji Shinohara, MD,* Naohiko Takahashi, MD,* Tetsunori Saikawa, MD,† Hironobu Yoshimatsu, MD*



Experimental Hypothermia: Respiratory and Blood pH Changes in Relation to Cardiac Function

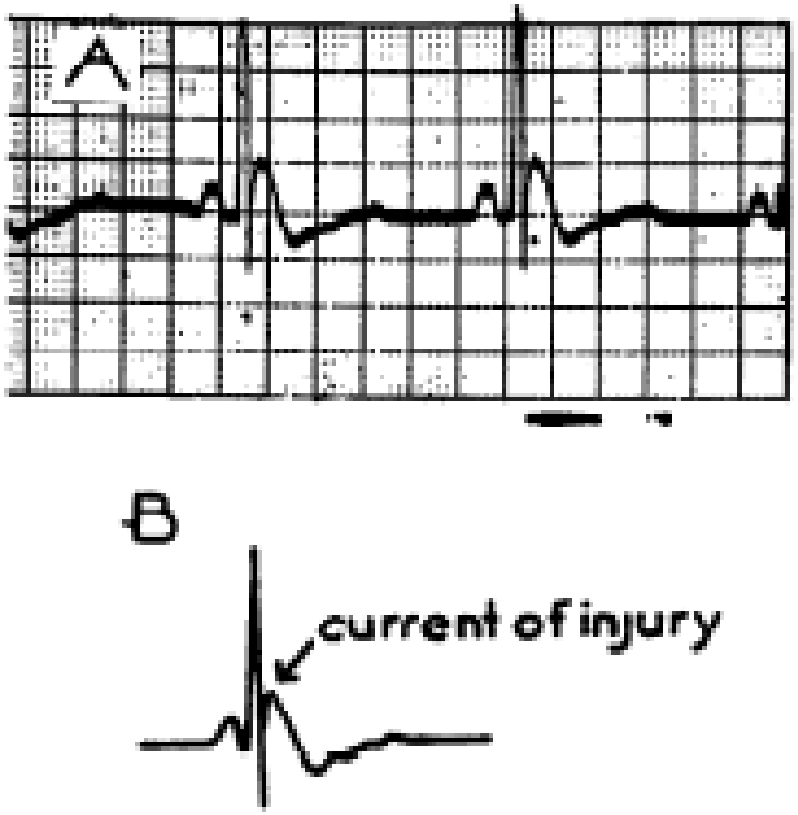
JOHN J. OSBORN[1]

From the Department of Pediatrics, New York University College of Medicine, New York City

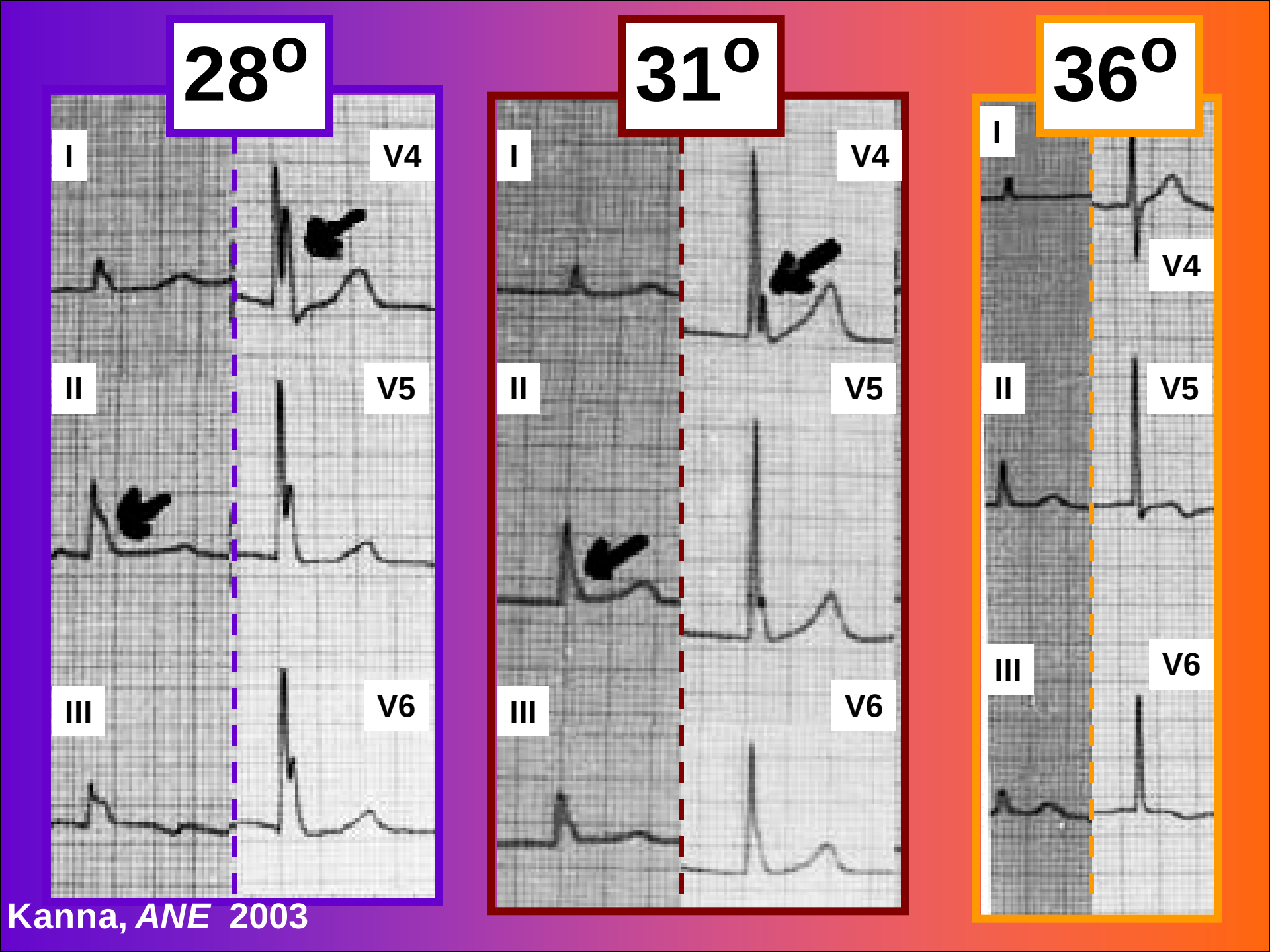
TEMPORARY whole-body hypothermia in theory offers an ideal way of greatly reducing metabolism, and seems to hold great promise in clinical surgery. Yet, although the reptile or the hibernating mammal can withstand very low body temperatures without distress, body temperatures much below 28°C produce severe and often fatal physiological stress in the non-hibernating mammal.

In the course of a series of studies of the physiology of experimental hypothermia in the dog, we have observed profound changes in the auto-regulation of respiration and of blood pH. These changes appear to be im-

volume was measured directly in several ways, eventu-

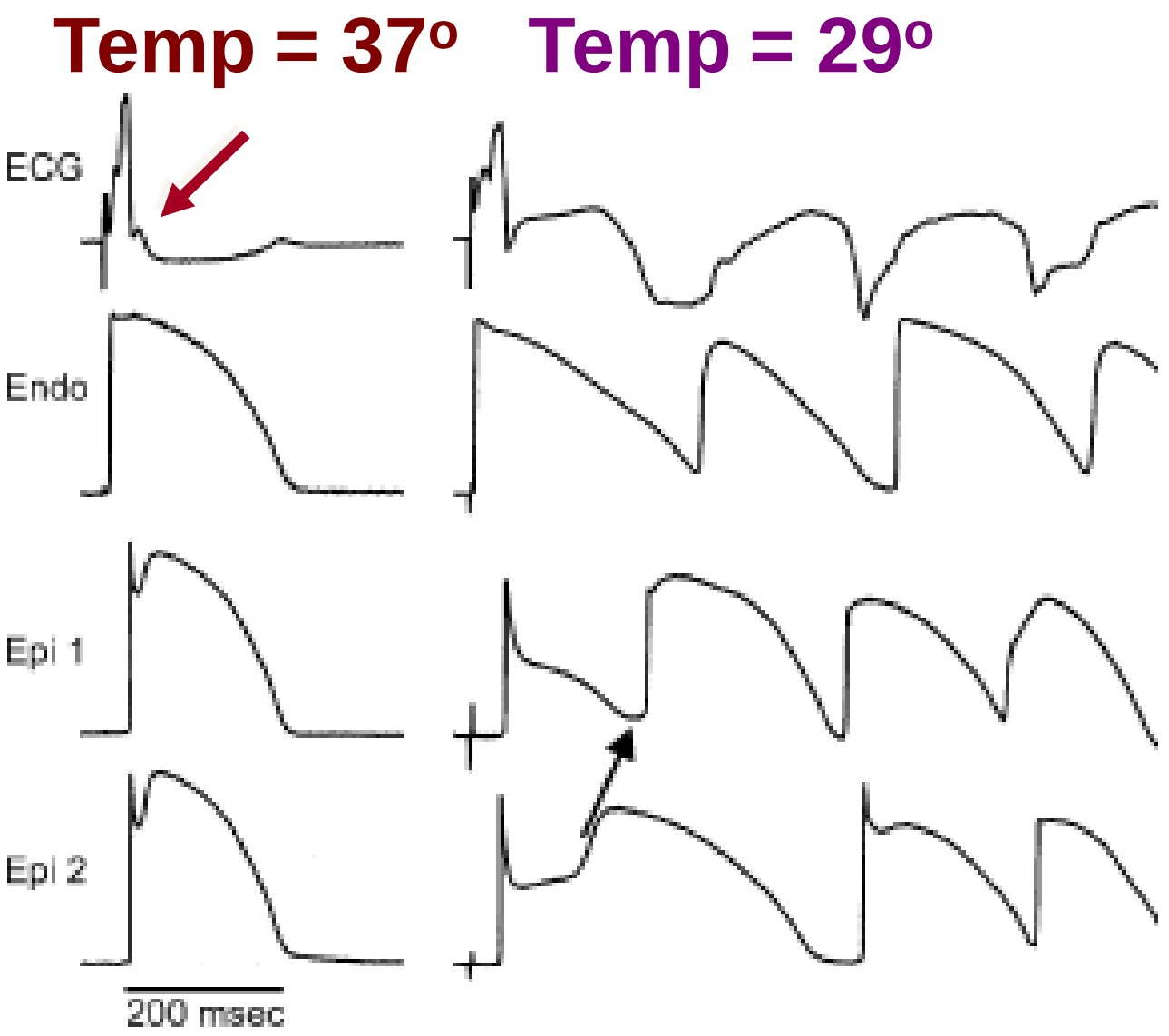


Am J Physiol 1953



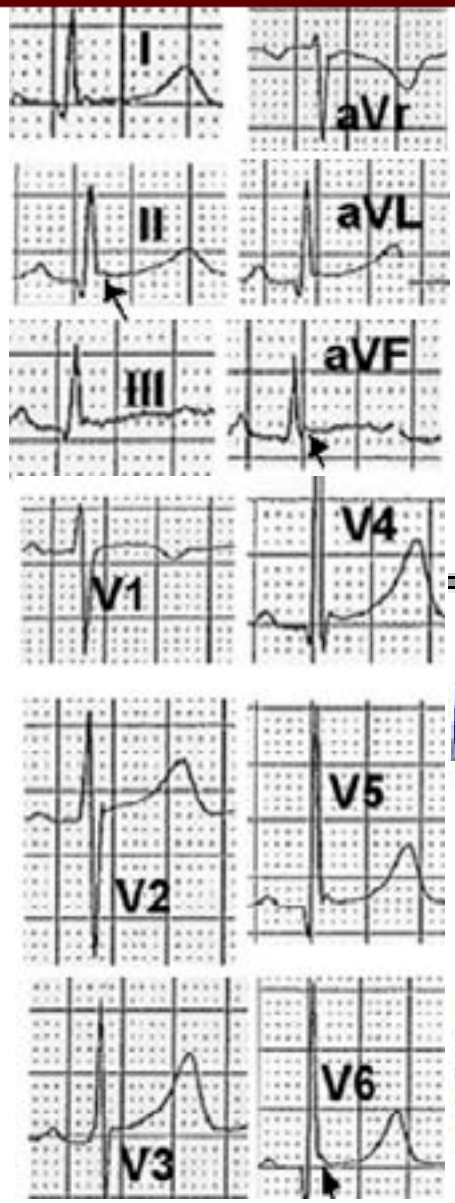
Kanna, *ANE* 2003

Phase 2 reentry by cooling

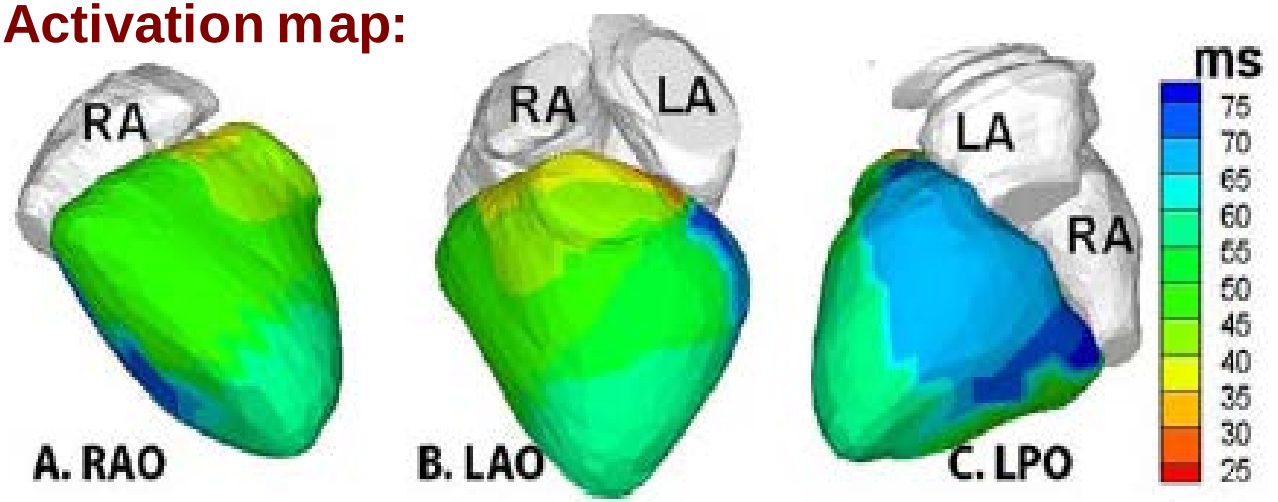


Fish, Antzelevitch, *JCE 2004*

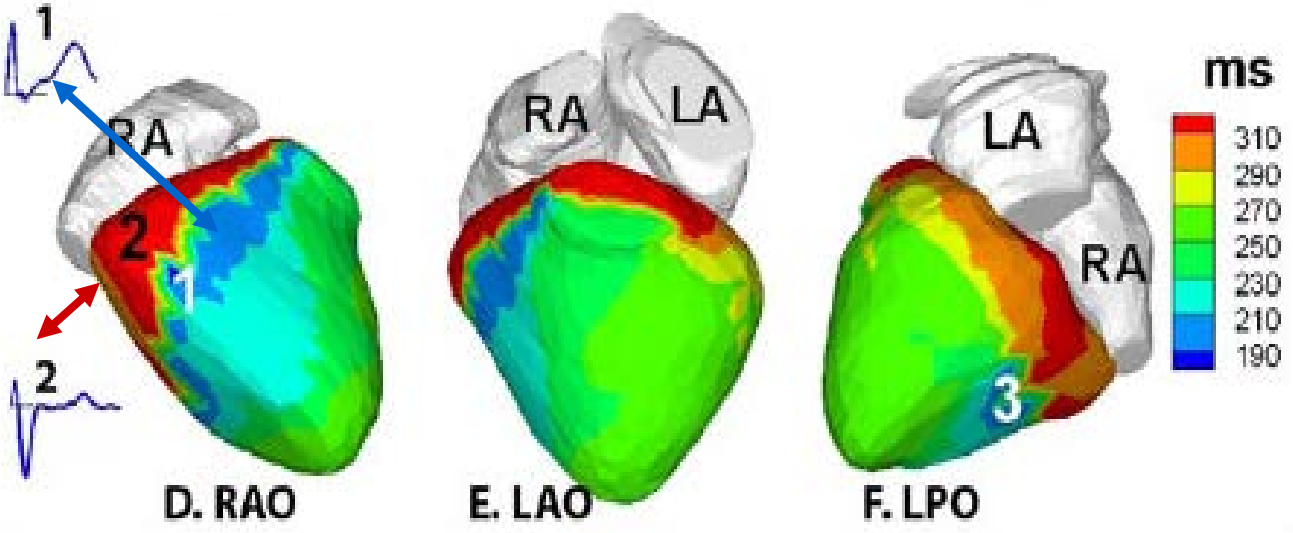
Non-invasive Electrocardiographic Imaging of Idiopathic VF



Activation map:



Activation Recovery Interval (ARI):



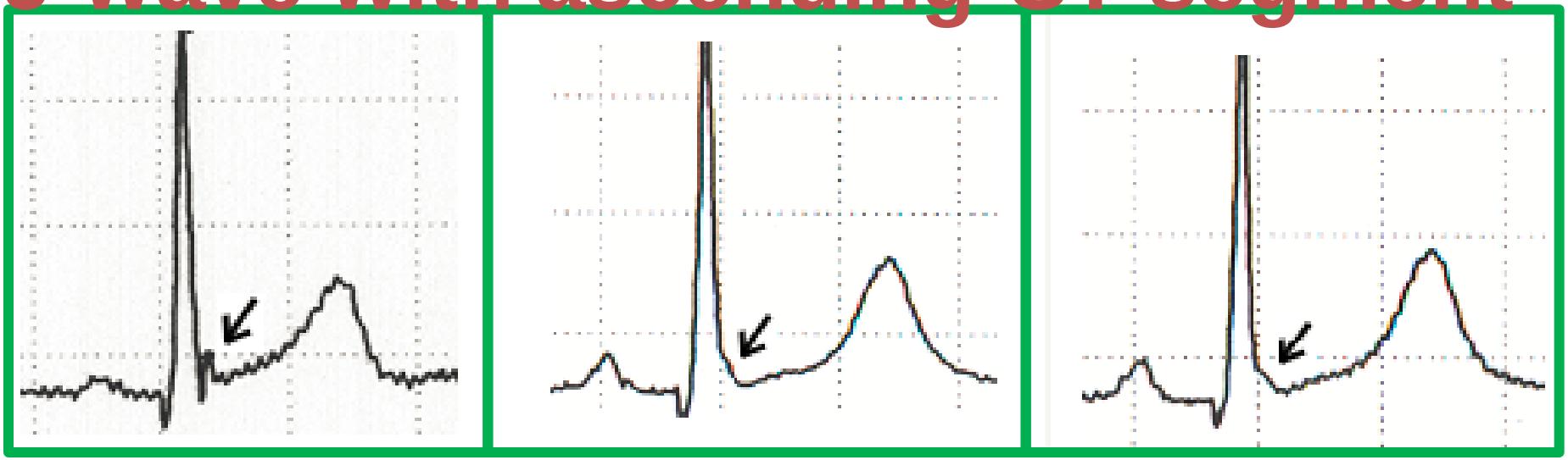
Ghosh, Heart*Rhythm* 2010

Early Repolarization: Electrocardiographic Phenotypes Associated with Favorable Long-Term Outcome

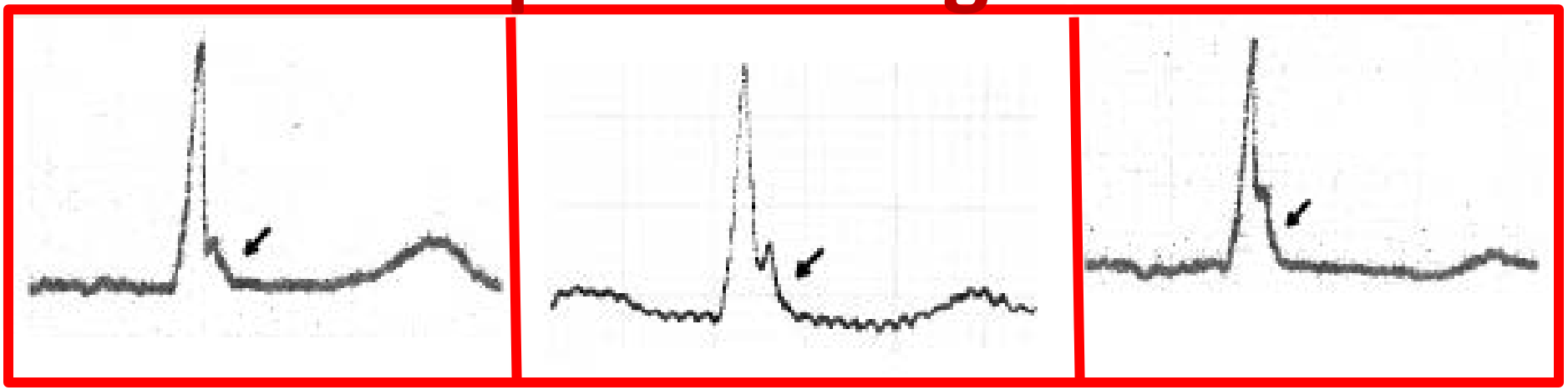
Jani T Tikkanen, Juhani M Junttila, Olli Anttonen, Aapo L Aro, Samuli Luttinen, Tuomas Kerola, Solomon J Sager, Harri A Rissanen, Robert J Myerburg, Antti Reunanen, and Heikki V Huikuri

CIRCULATIONAHA/2010/014068 [R1]

J-wave with ascending ST-segment

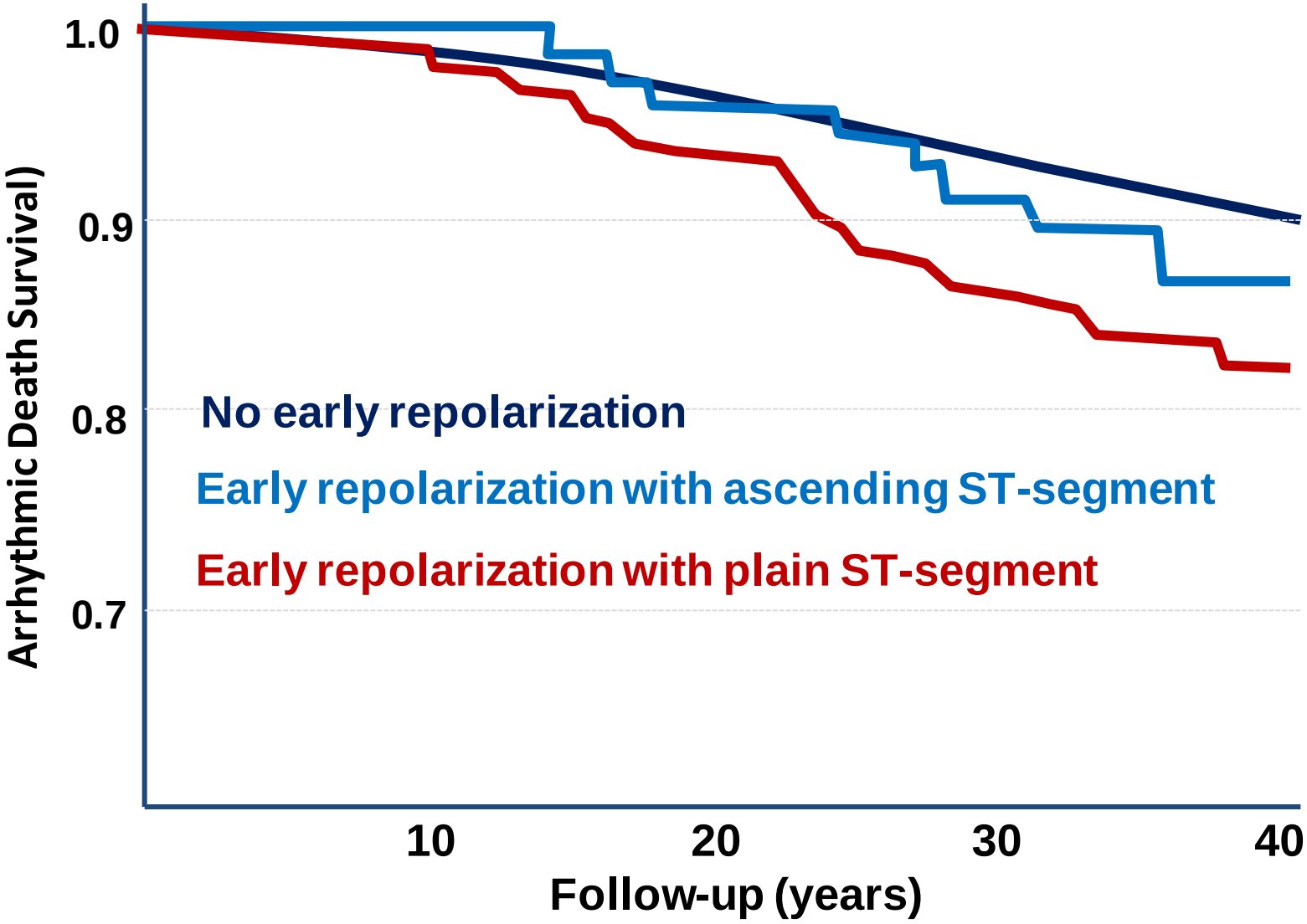


J-wave with plain ST-segment



Only the “plain-type” ST –segment predicts long-term arrhythmic risk in patients with early repolarization.

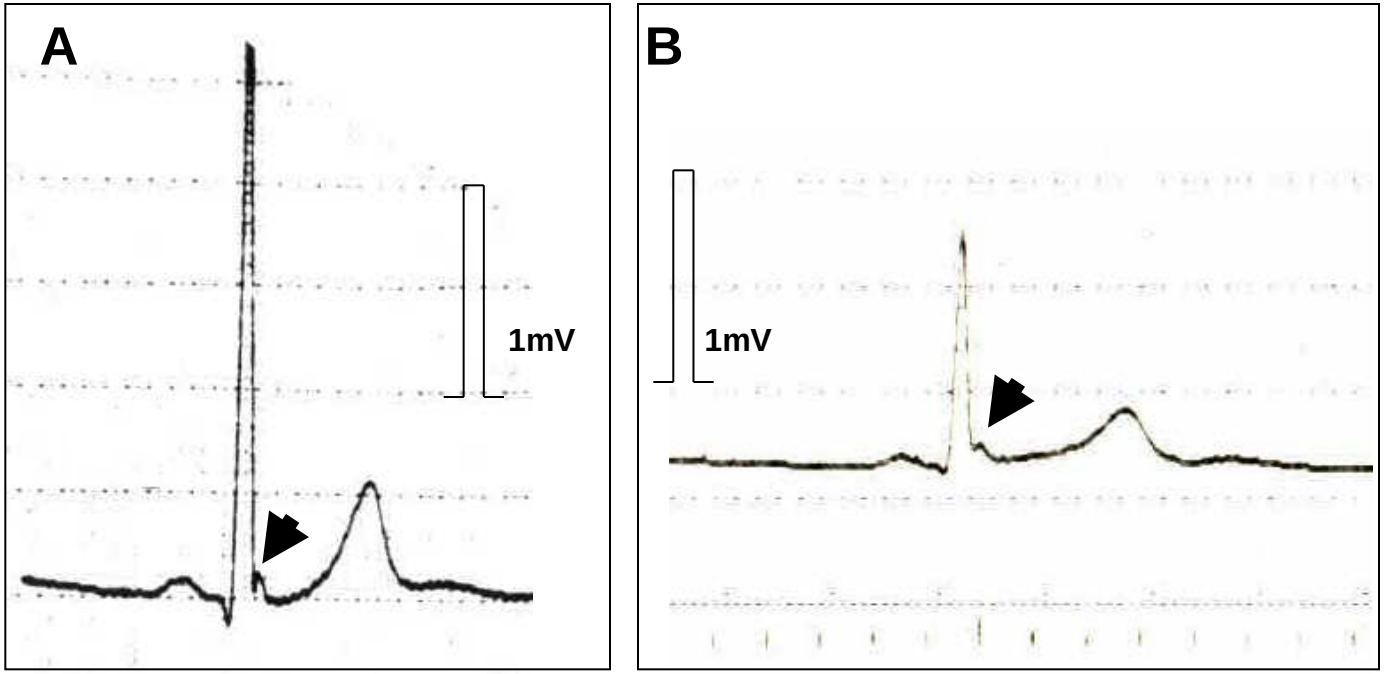
Tikkanen, *Circulation* 2011 (in press).



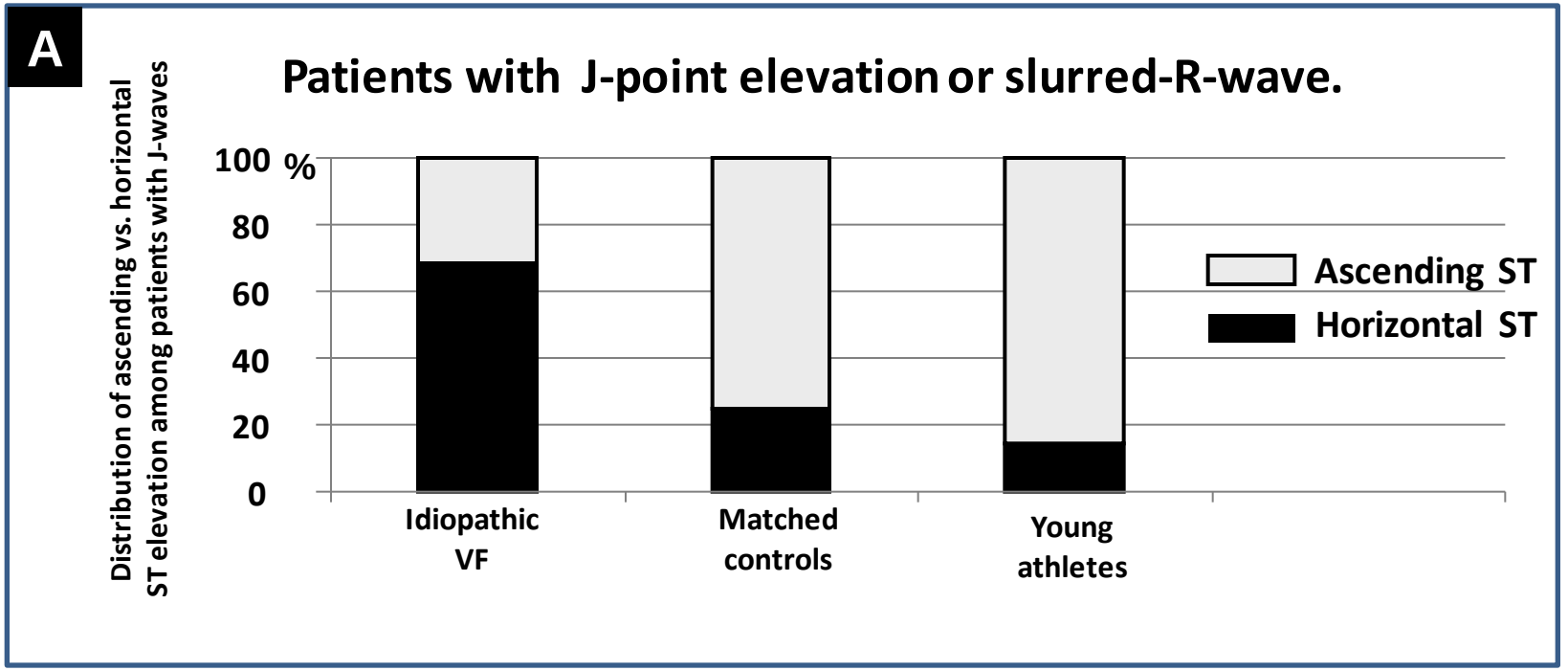
J-Point Elevation in Survivors of Primary Ventricular Fibrillation and Matched Control Subjects

Incidence and Clinical Significance

Distinguishing "benign" from "malignant early repolarization:" The value of ST-segment morphology.



Rosso, Viskin. Heart*Rhythm* (in press)


A
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

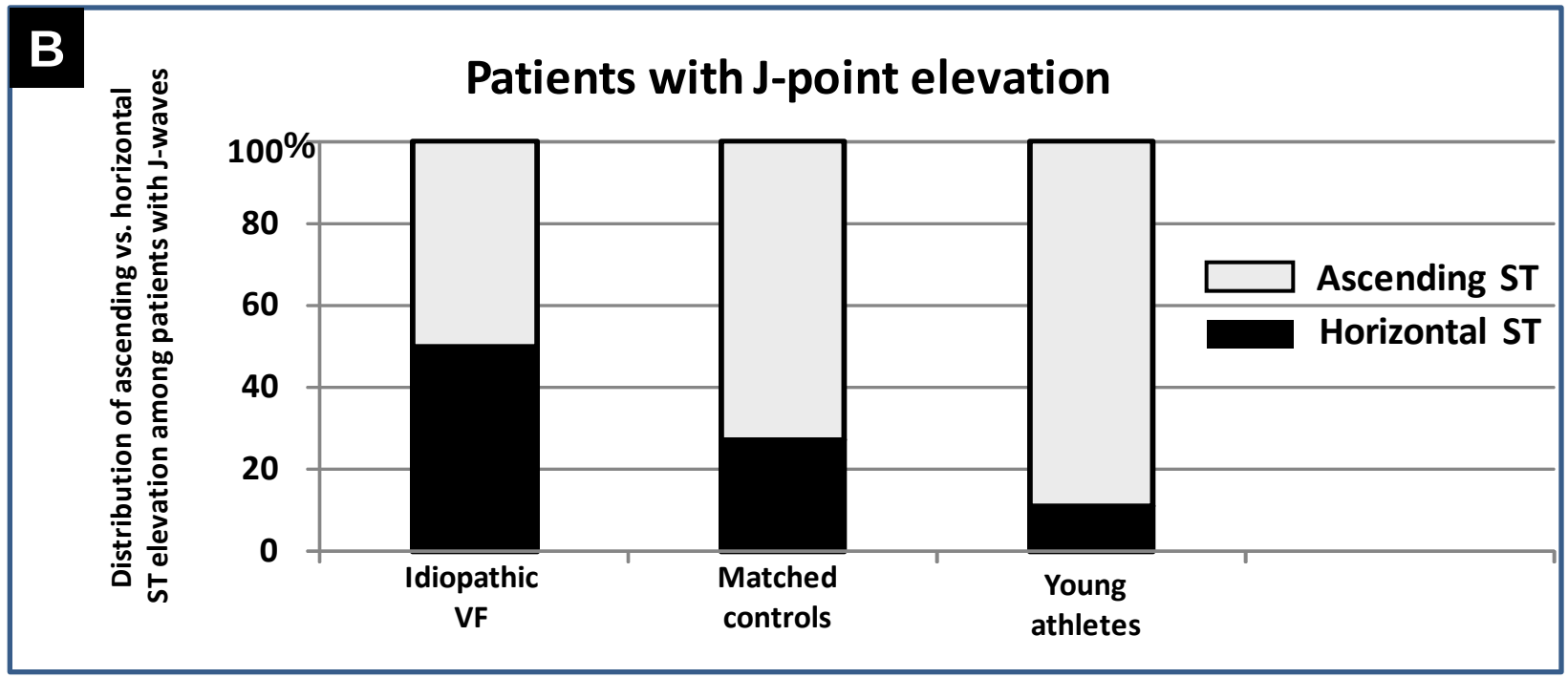
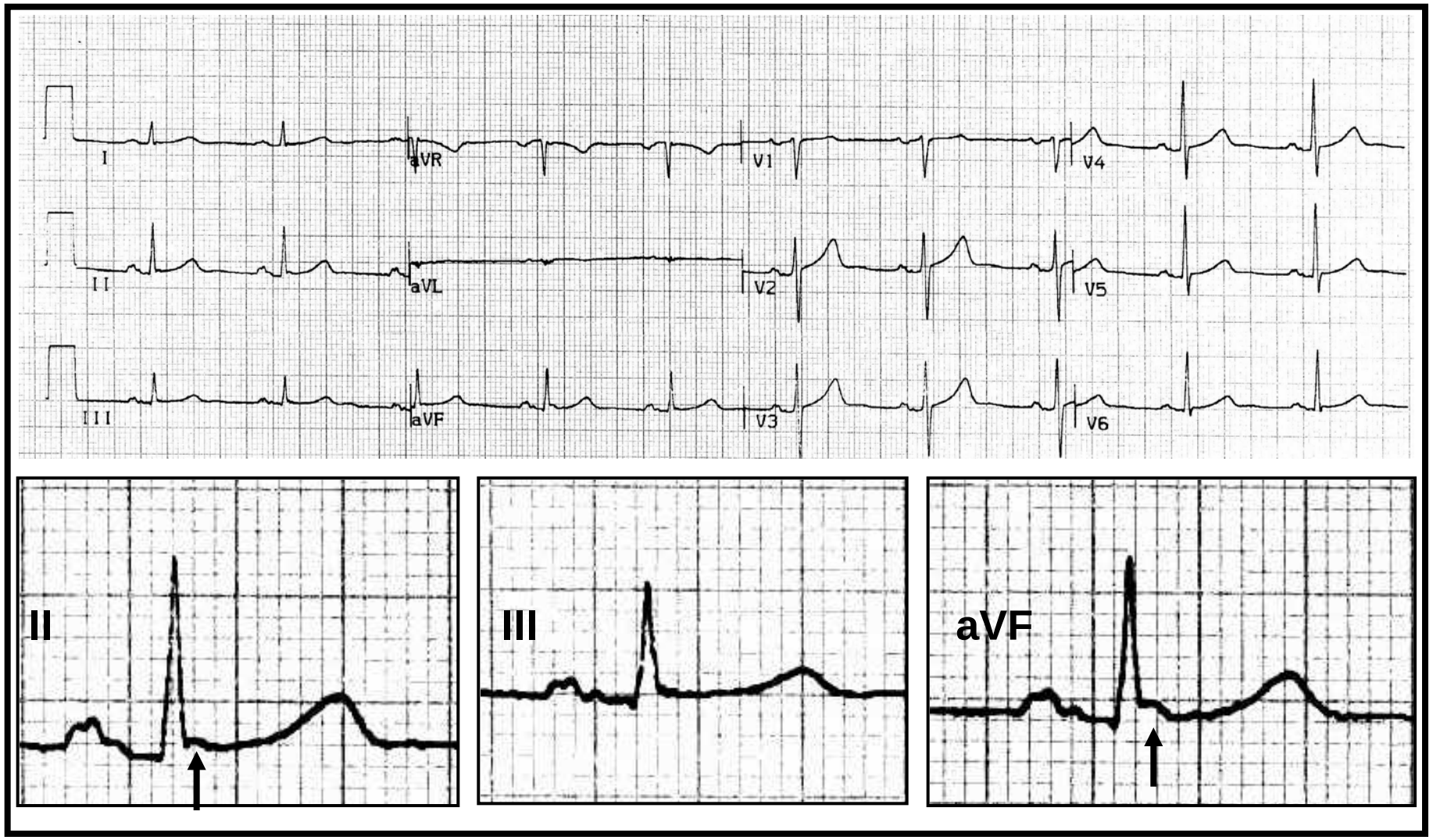

B
Patients with J-point elevation
Distribution of ascending vs. horizontal ST elevation among patients with J-waves
100%
80
60
40
20
0
Idiopathic VF
Matched controls
Young athletes
Ascending ST
Horizontal ST

Figure 2.



Rosso (Viskin), Heart*Rhythm* 2011 (in press).

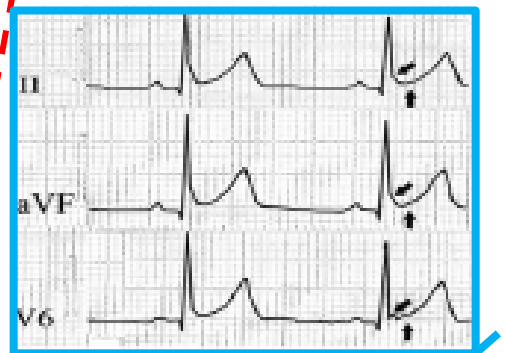
Original Articles

J Wave, QRS Slurring, and ST Elevation in Athletes With Cardiac Arrest in the Absence of Heart Disease

Cardiac arrest

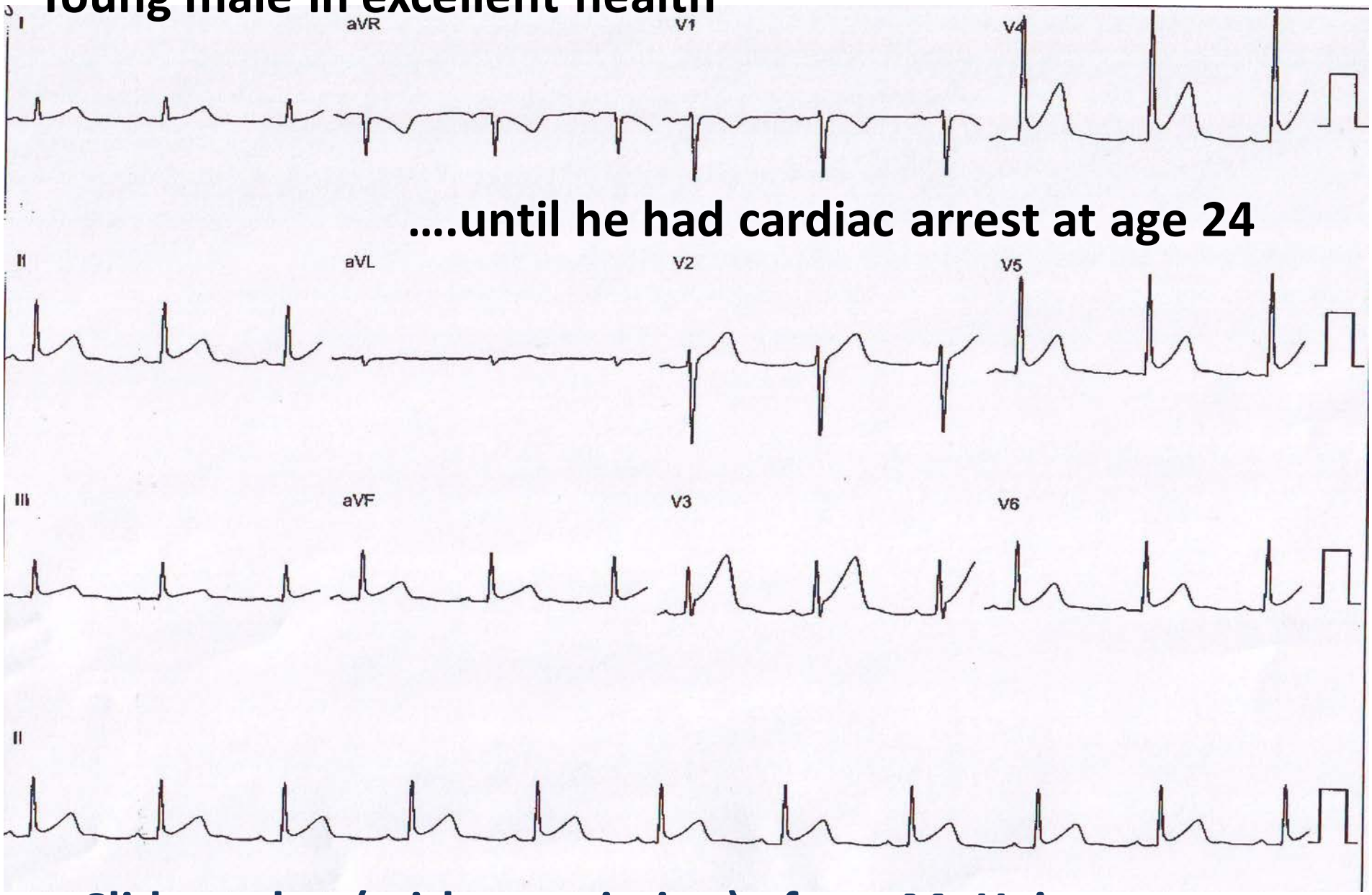
Healthy athlete

slurring du
Conclusions
than in co
malignant



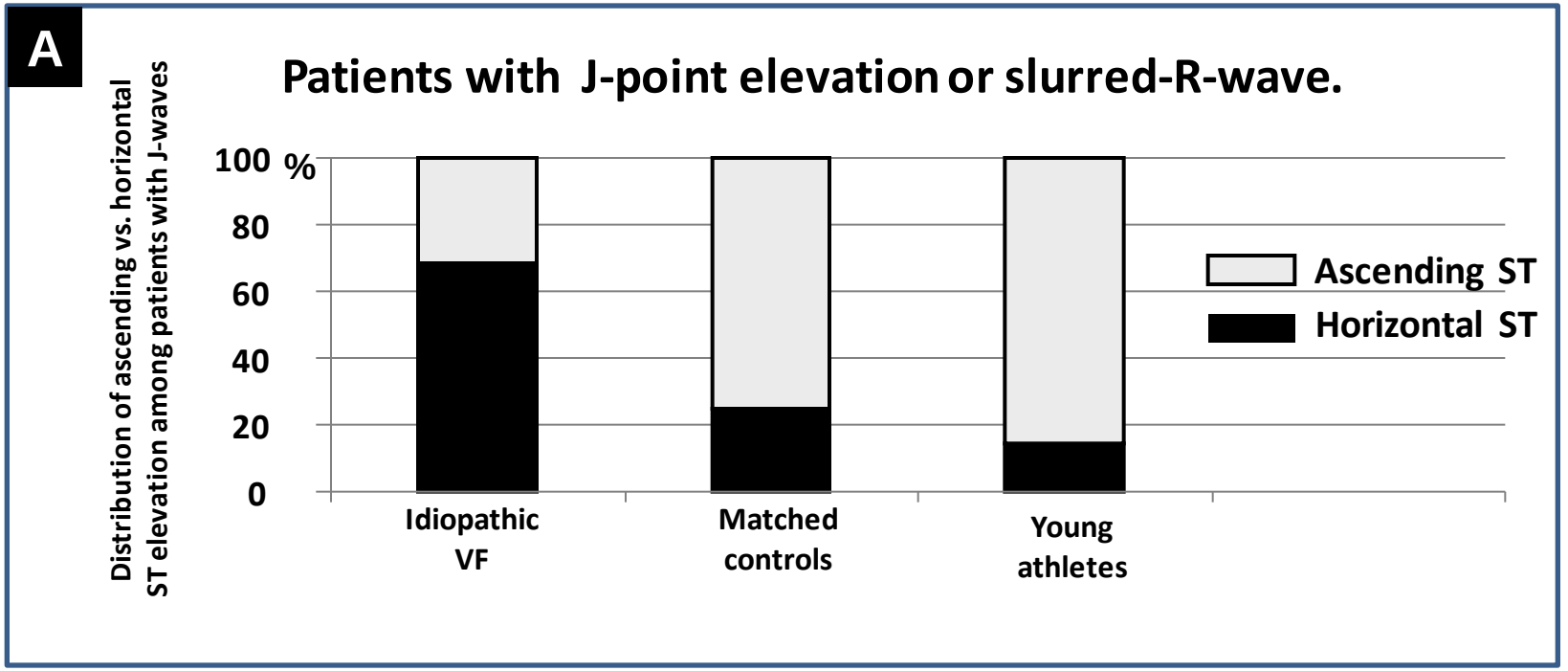
t/sudden death
k for recurrent

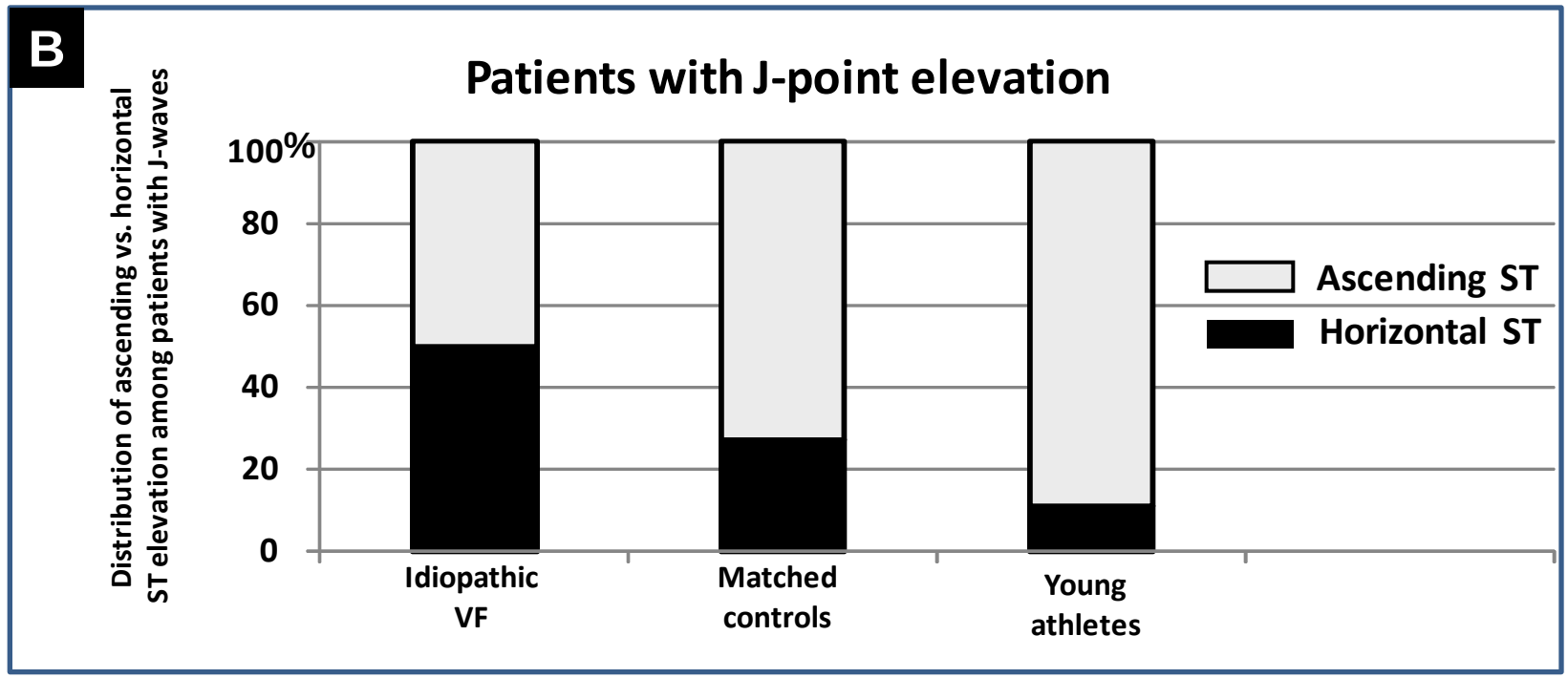
Young male in excellent health



….until he had cardiac arrest at age 24

Slide stolen (with permission) from M. Haissaguerre.


A
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

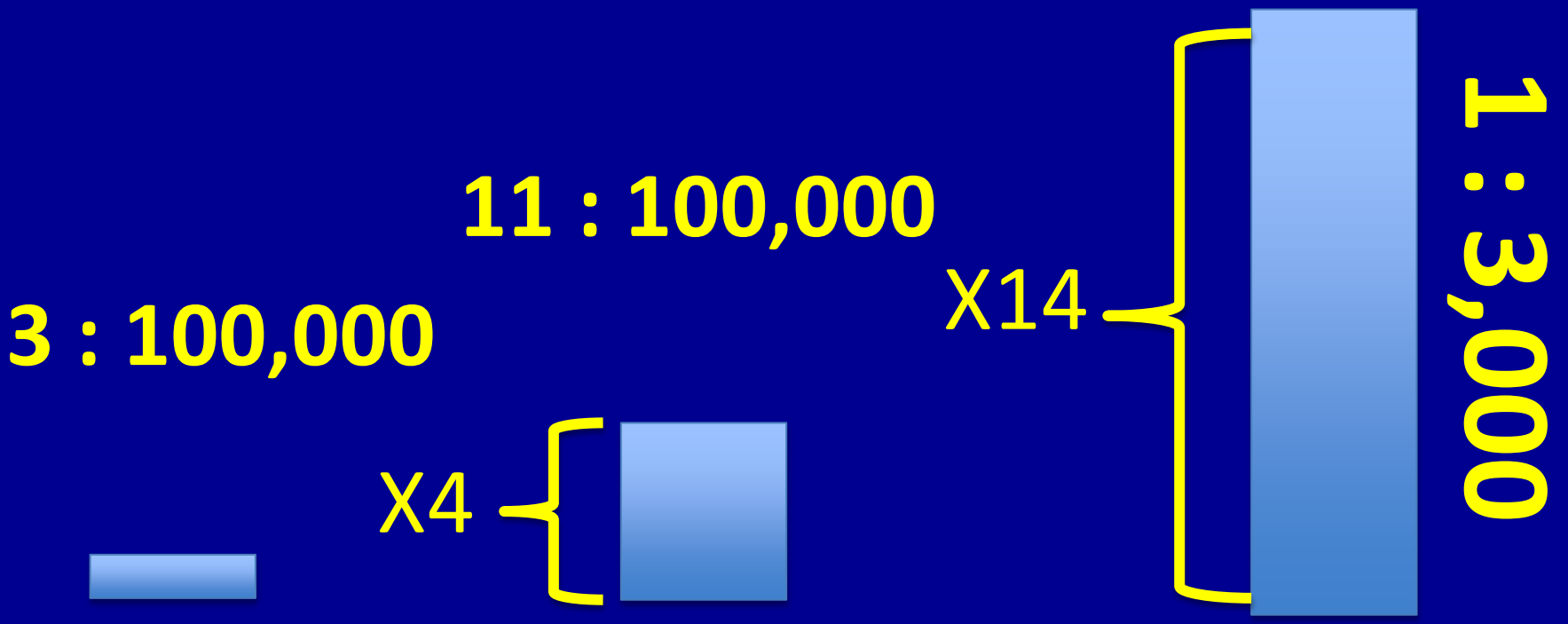

B
Patients with J-point elevation
Distribution of ascending vs. horizontal ST elevation among patients with J-waves
100%
80
60
40
20
0
Idiopathic VF
Matched controls
Young athletes
Ascending ST
Horizontal ST

Risk of dropping dead
(asymptomatic young individuals).

Someone like me

With J-Waves

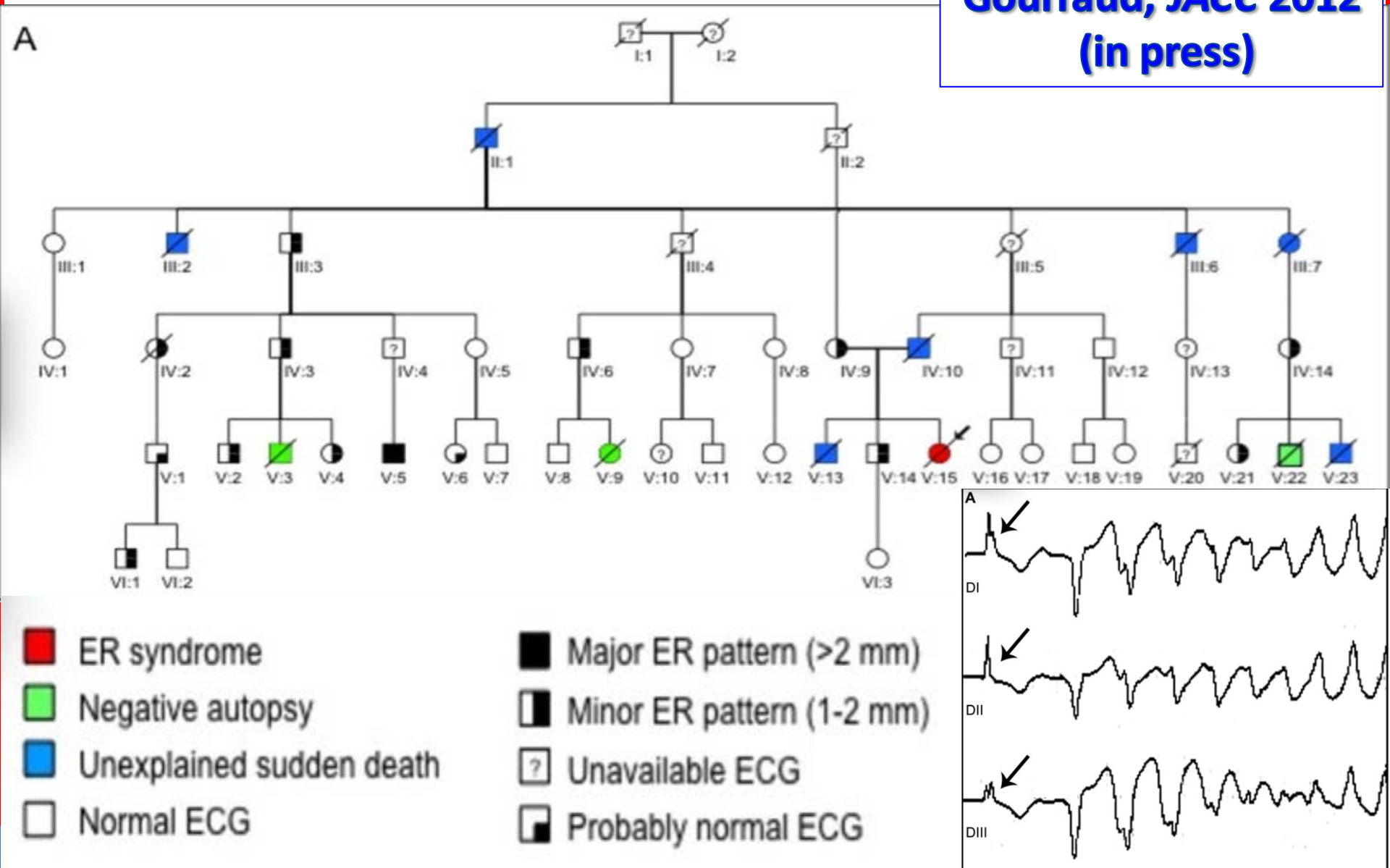
With J-waves and malignant ST



Rosso, Viskin, *Heart*Rhythm *2012*

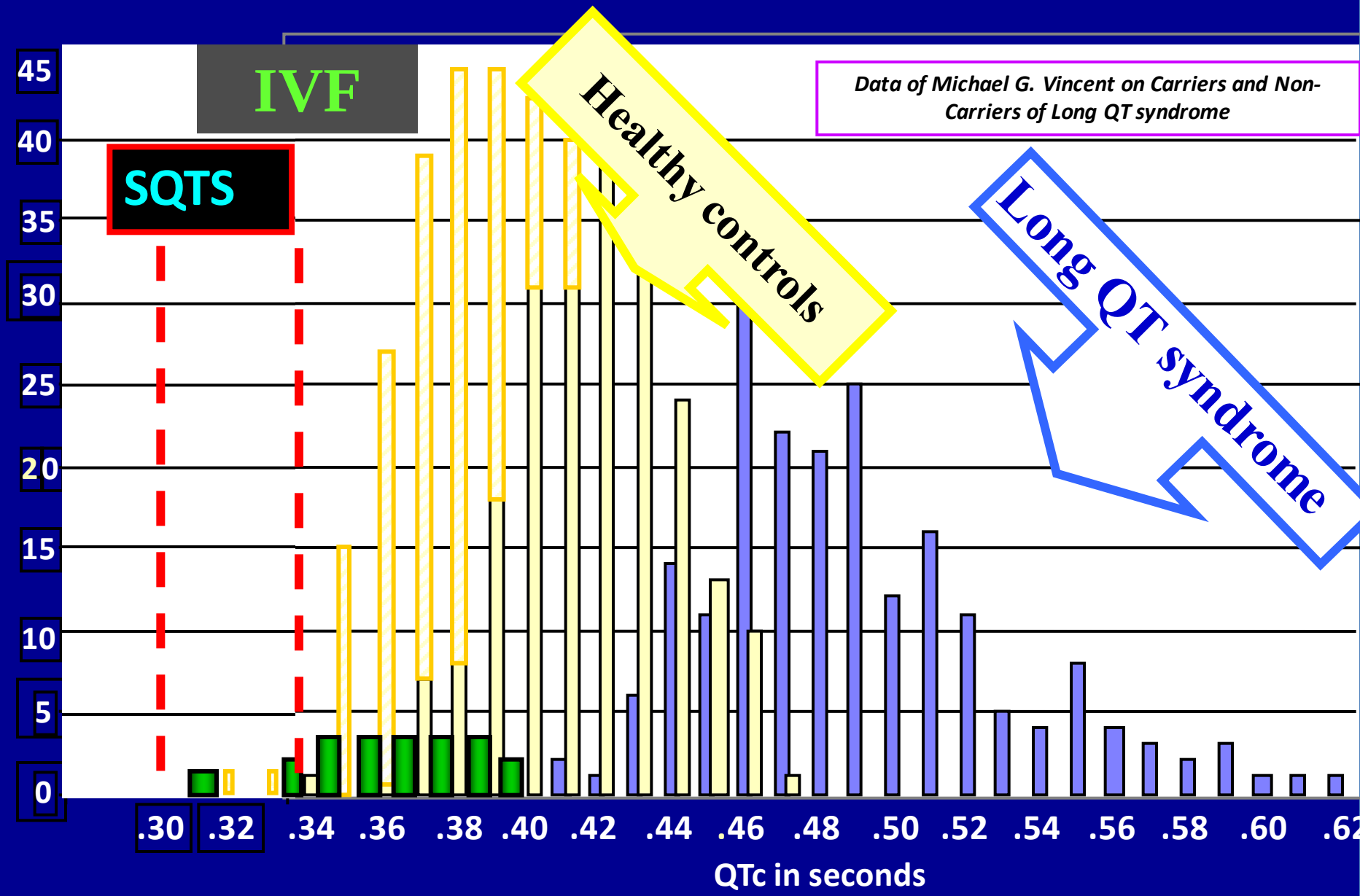
Identification of large families with early repolarization syndrome

Gourraud, *JACC* 2012 (in press)



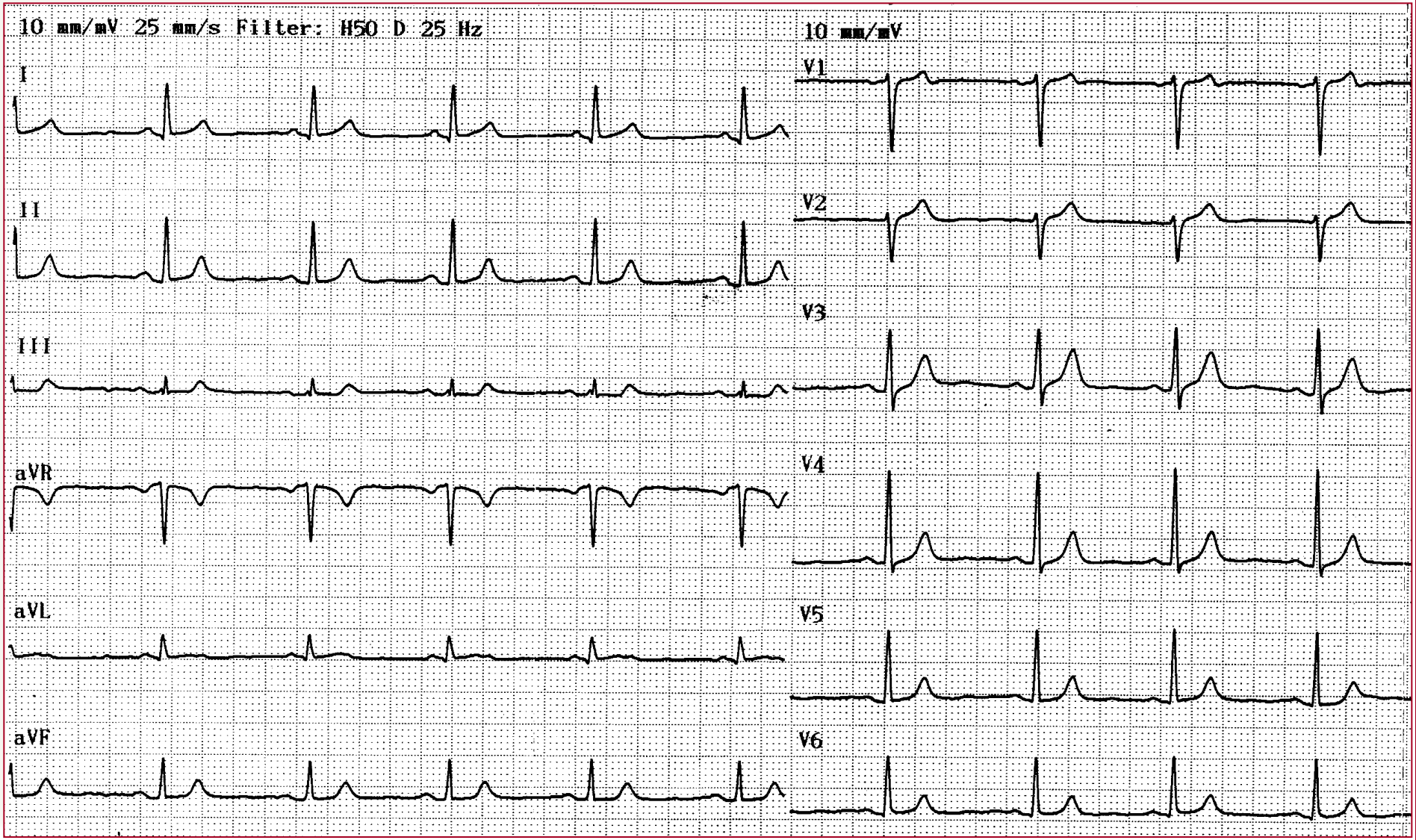

It's the same,
but different

Idiopathic VF: A short QT syndrome with not-so-short QT interval.



Viskin, Heart*Rhythm* 2004

Female, age 35. Syncope during a phone conversation

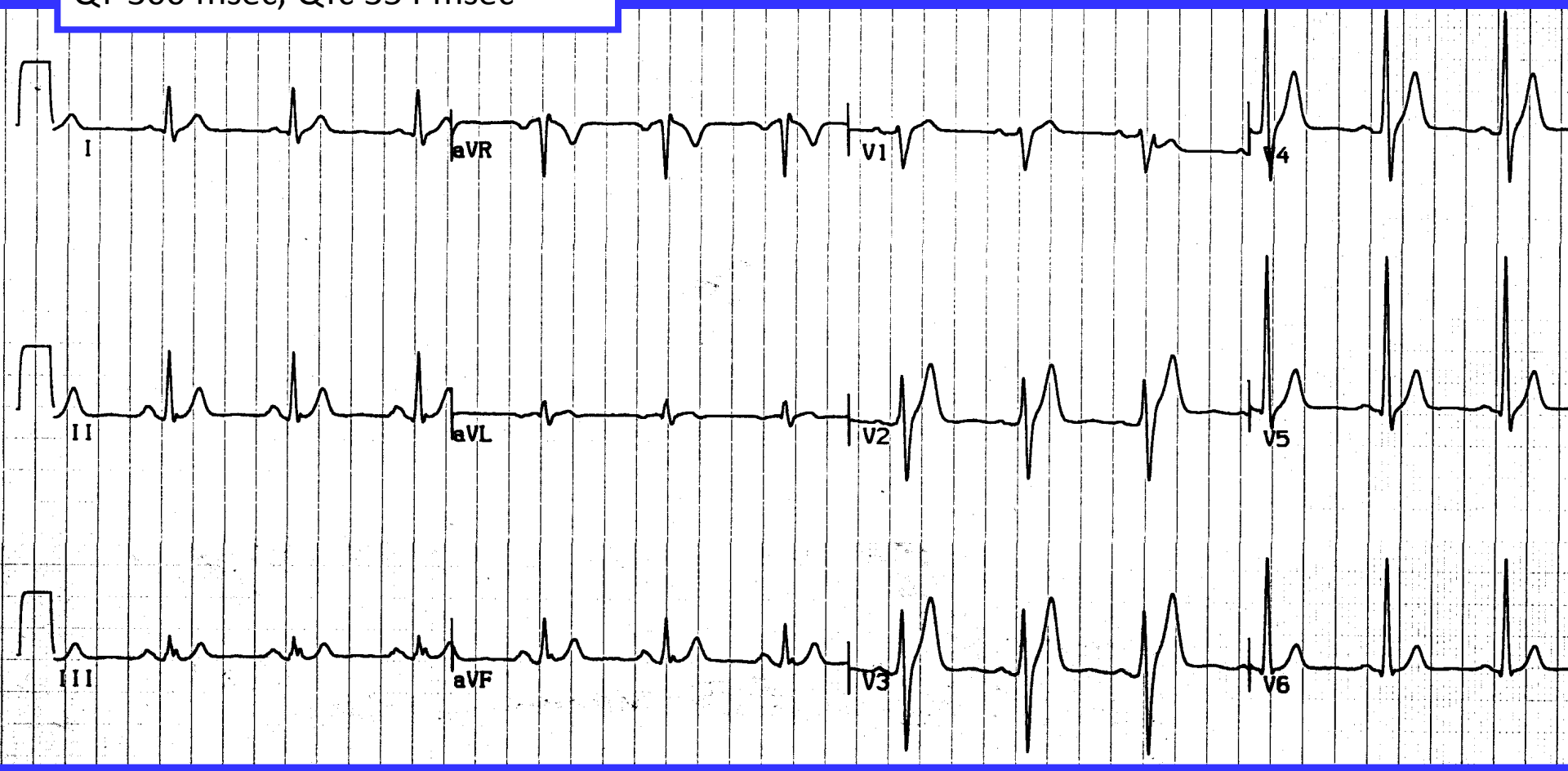



I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
N
N
N
V
V
V
V
Sensitivity: 10 mm/mV
Paper speed: 25 mm/sec
An ambulatory 12-lead ECG obtained with torso-located limb electrodes is not equivalent to a conventional diagnostic ECG.

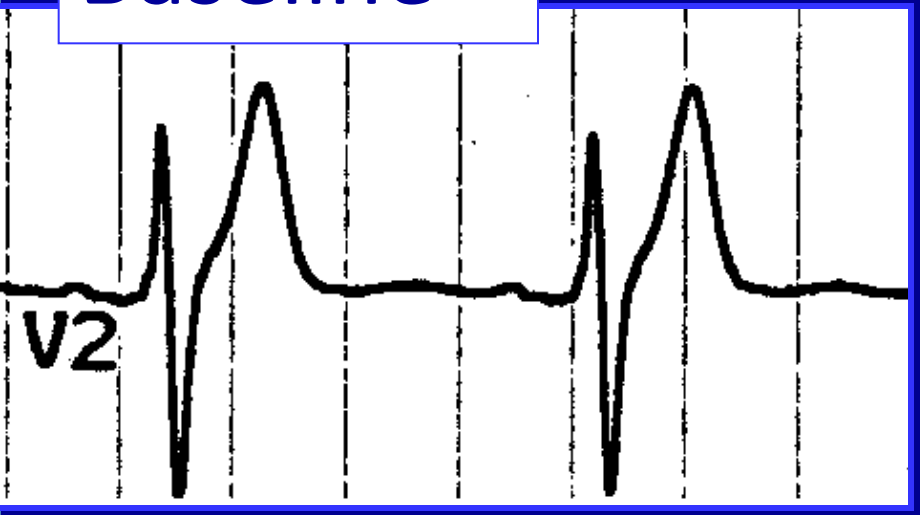

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
N
N
V
V
V
N
Sensitivity: 10 mm/mV
Paper speed: 25 mm/sec
An ambulatory 12-lead ECG obtained with torso-located limb electrodes is not equivalent to a conventional diagnostic ECG.

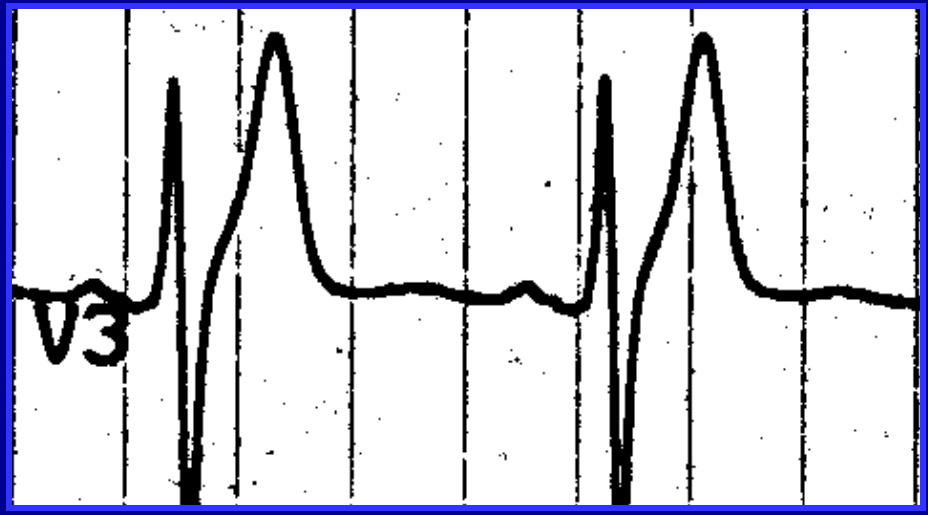
Her asymptomatic brother: 34 years old.

QT 300 msec, QTc 354 msec

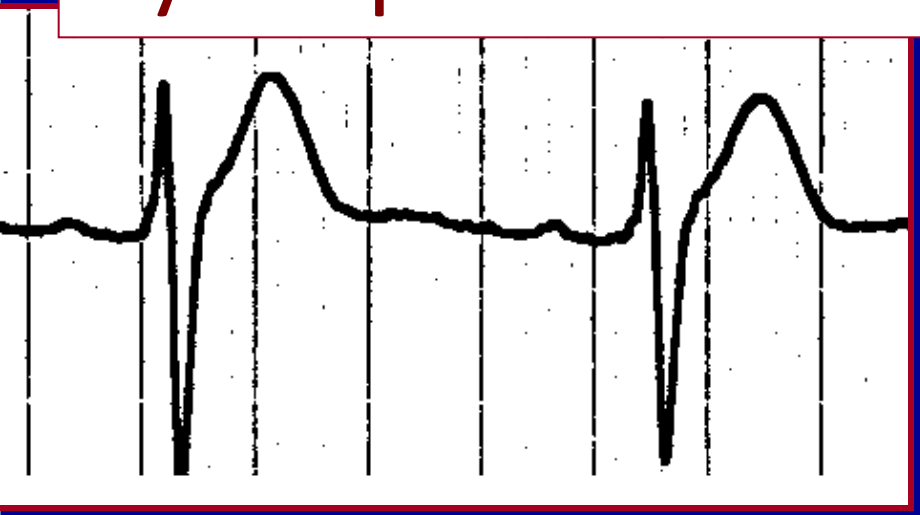


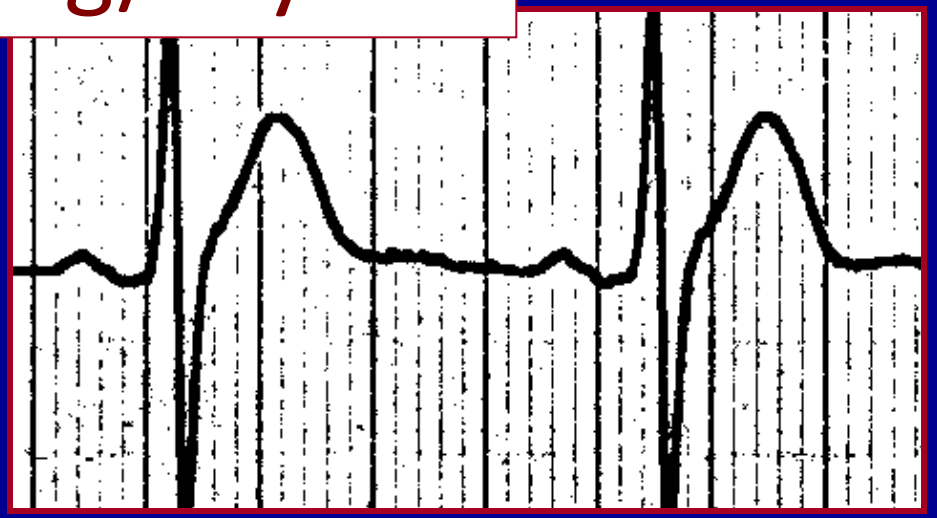
Baseline





Hydroquinidine 900 mg/day





QT = 360 msec, QTc = 390 msec.

“There are known knowns;
these are things we know that we know.
There are known unknowns;
these are things we no know we don't know.
But there are also unknown unknowns – these are things we do not even know we don't know.”

Donald Rumsfled
U.S. Defense Secretary