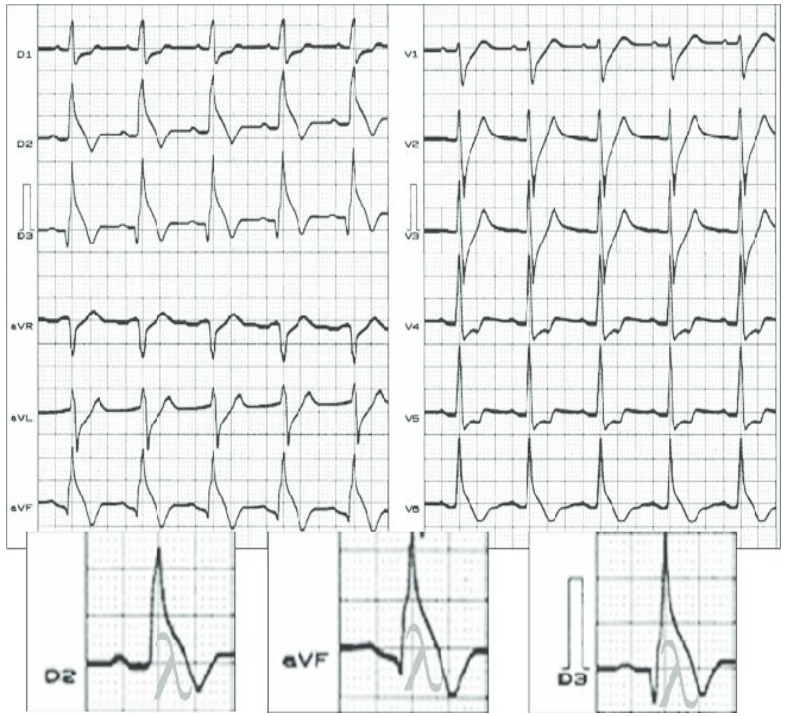
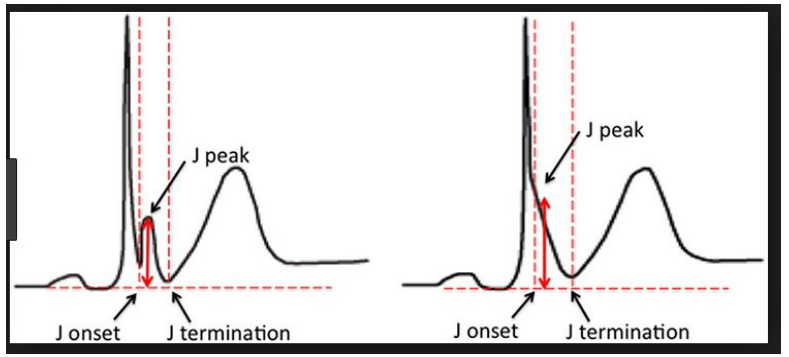
Early repolarization or Inferolateral J wave Context -Diagnostic

Prevalence unclear (Asia >Europe-America) : 7 Men / 1 Woman

Represents up to one third of idiopathic VF (2008)
Lesser now it s clinically taken in account

Mean age ~ 35 yrs at diagnosis :
Symptoms at rest (syncope- sudden death)





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

ER Syndrome

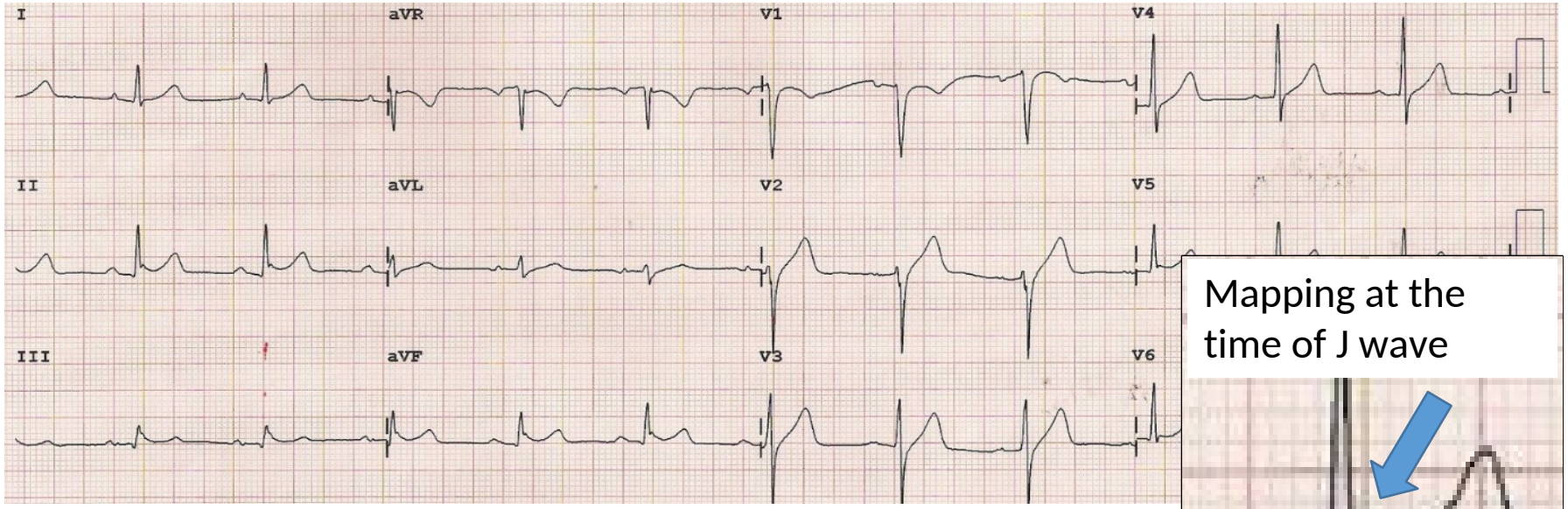
1. ER ***syndrome is diagnosed*** in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG in a patient resuscitated from otherwise unexplained VF/polymorphic VT.
2. ER syndrome ***can be diagnosed*** in a SCD victim with a negative autopsy and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

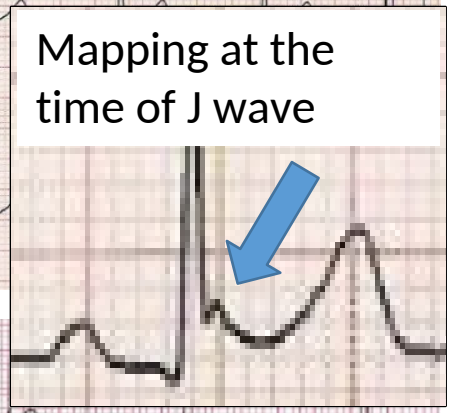
ER Pattern

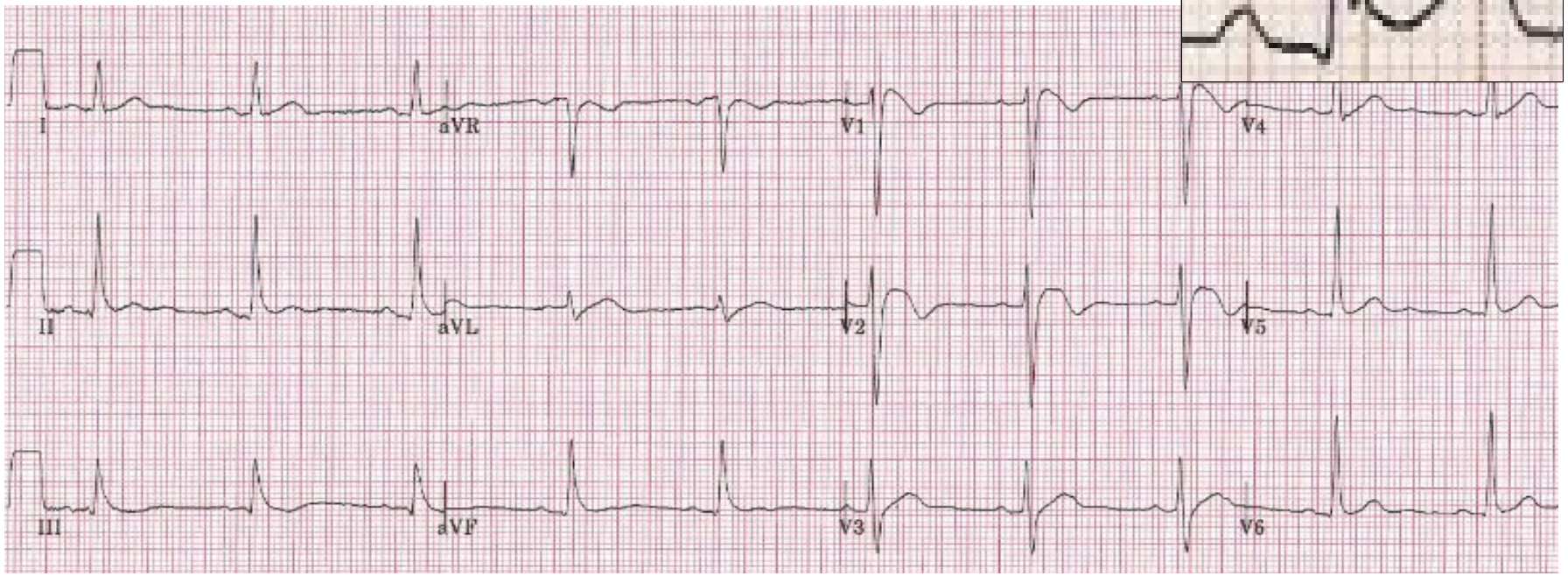
3. ER pattern ***can be diagnosed*** in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.

ER is associated with shorter repolarizatior
(QTc or Tpeak-to-end)

Inferolateral J wave –Early Repolarization







52 pts from Bangkok-Bordeaux -Tsukuba

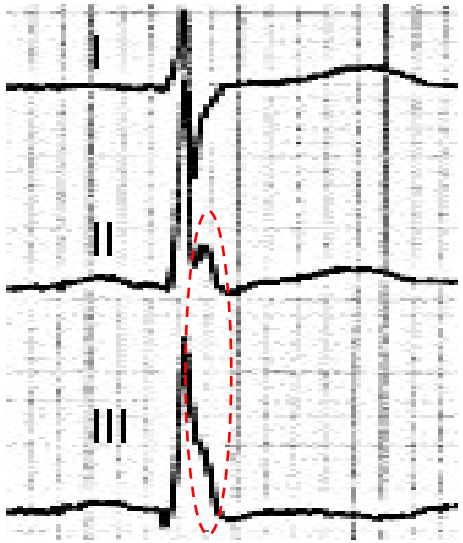
48 men -35±14 years

No Structural Heart Disease

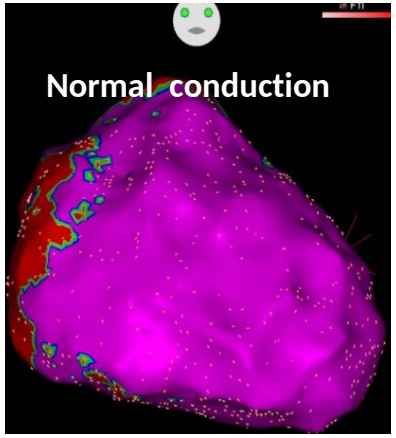
Endocardial+ Epicardial egm mapping ,

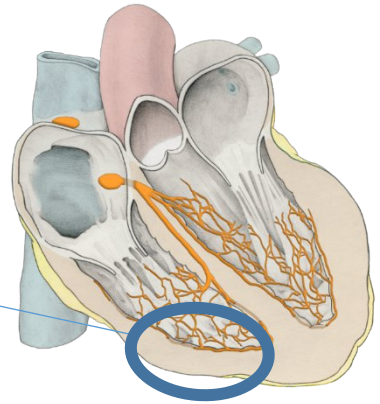
involving the J-wave

Inferolateral J wave has two distinct electrical causes :
True Early repolarization
Late depolarization (delayed conduction)



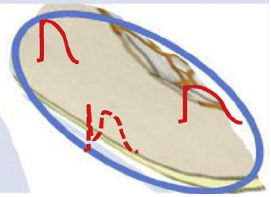
54 patients :
Bangkok- Bordeaux-Tsukuba

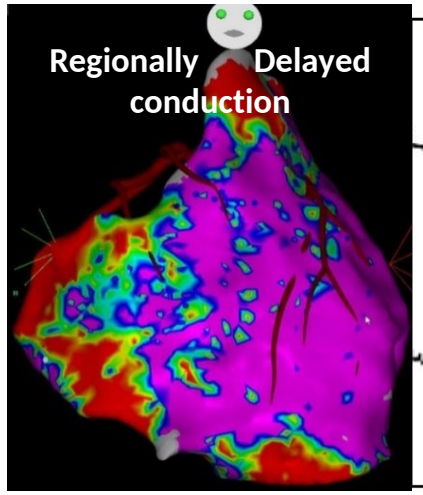


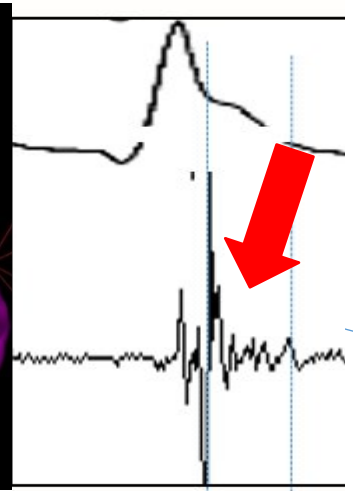


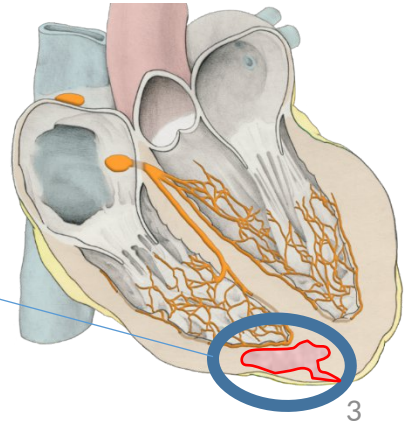
Repolarization Abnormality

Electrical abnormality heterogeneity of repolarization



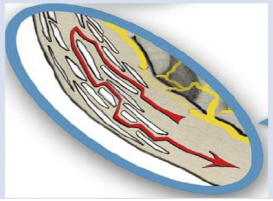






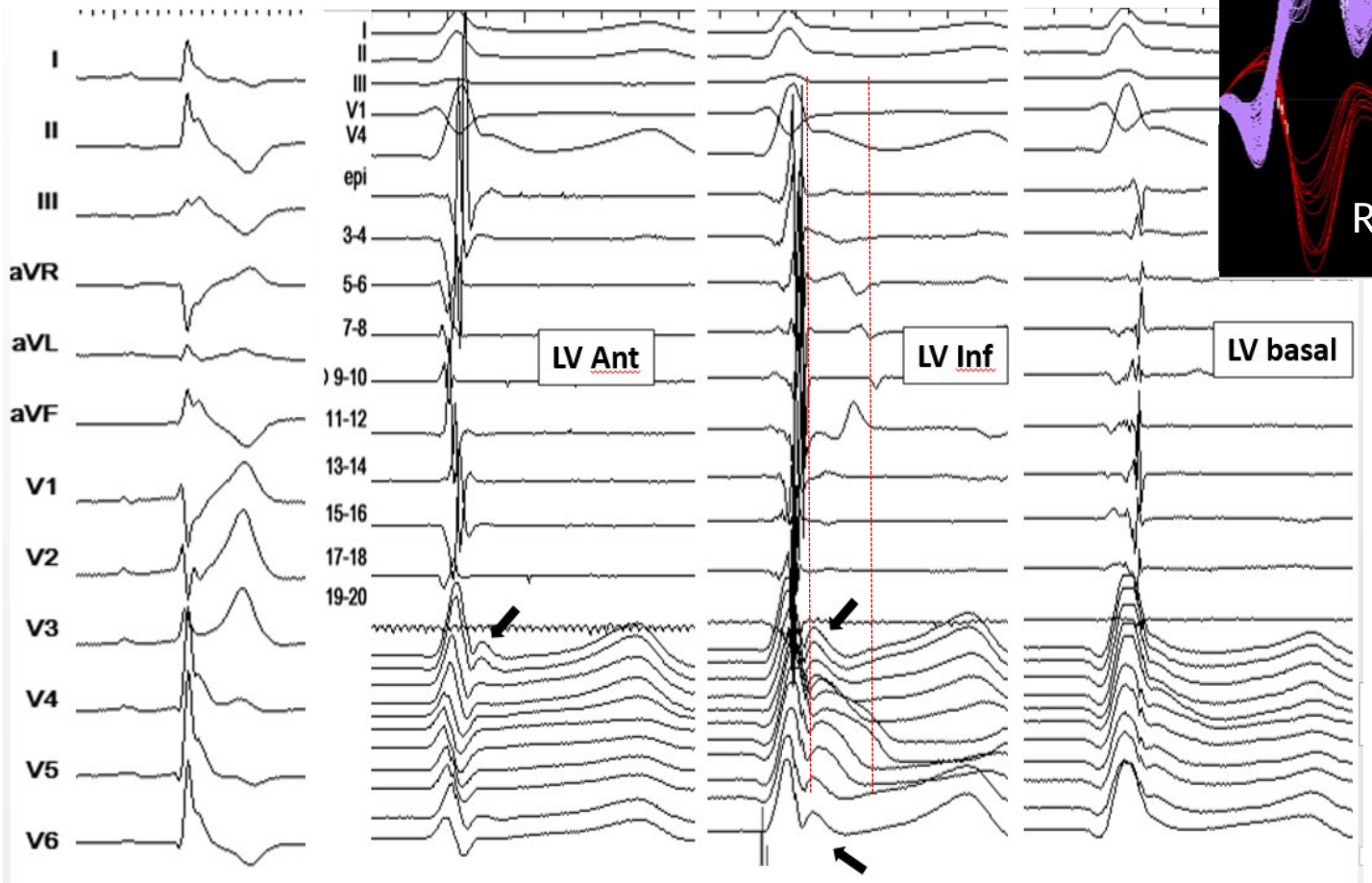
Conduction Abnormality

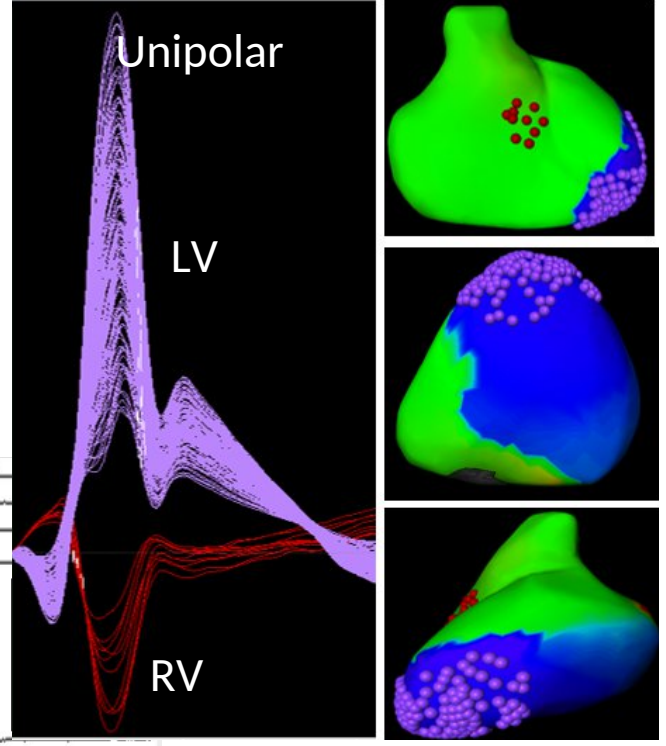
Structural abnormality - Heterogeneity of depolarization

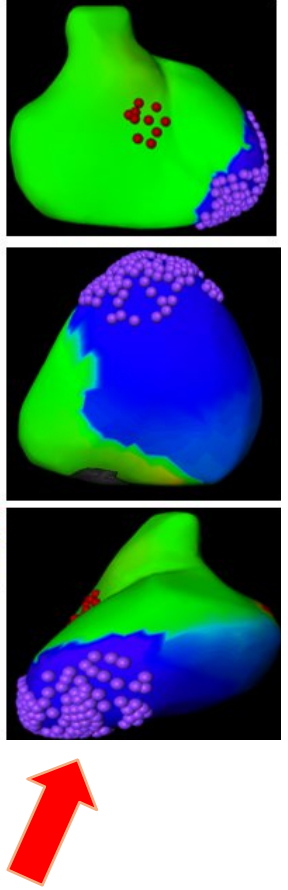


M 23 yrs - J wave caused by early repolarization

12 lead ECG

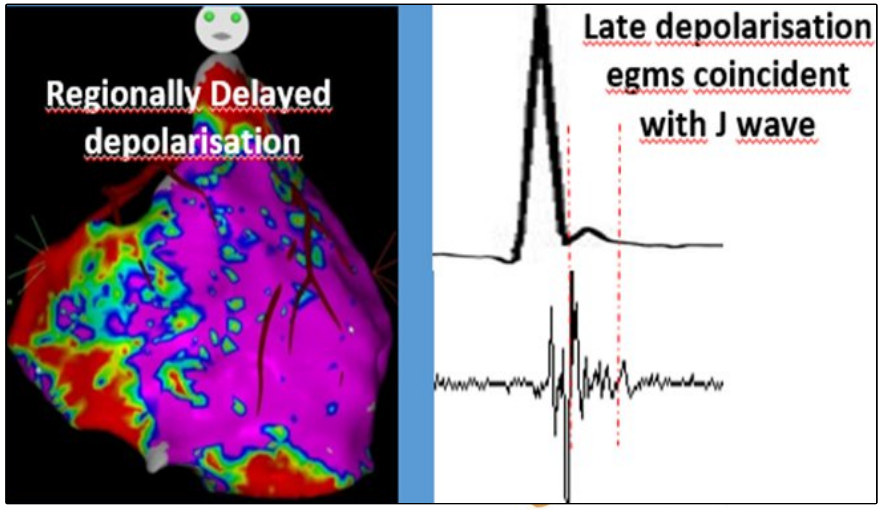






Extent of Epicardial Early repolarisation potentials

J-wave phenotype can be also caused by delayed conduction located at any inferior region in RV or LV

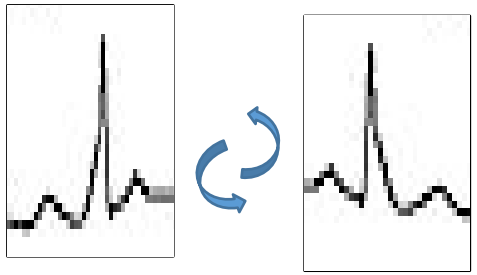


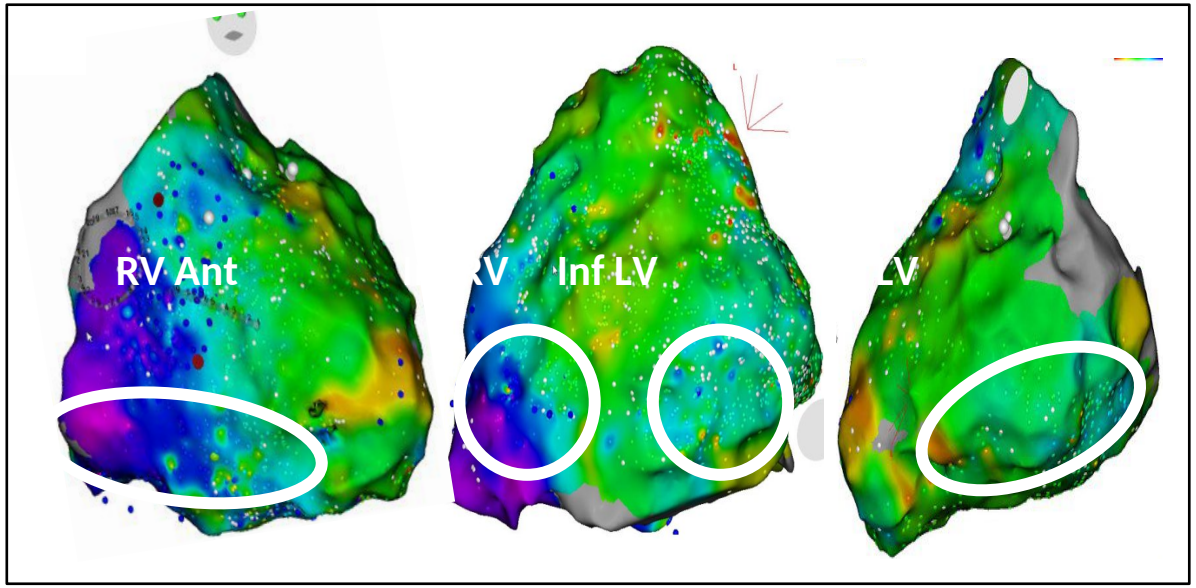
Areas of terminal activation are **mostly in epicardial** layers:

RV inferior or anteroinferior

LV inferior or inferolateral

Like a 'reversed' delta wave

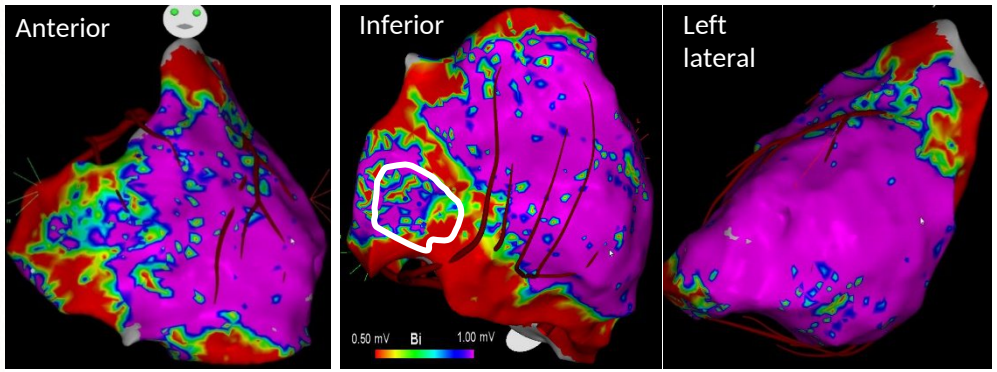




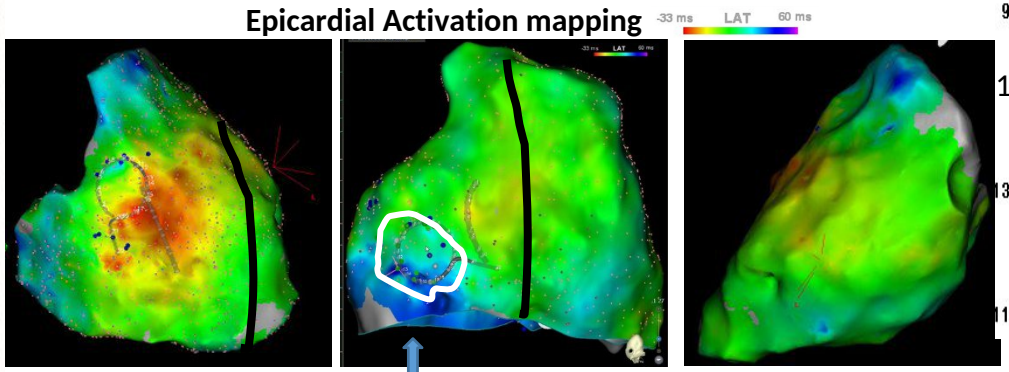
M 19yrs- J wave caused by abnormal depolarisation

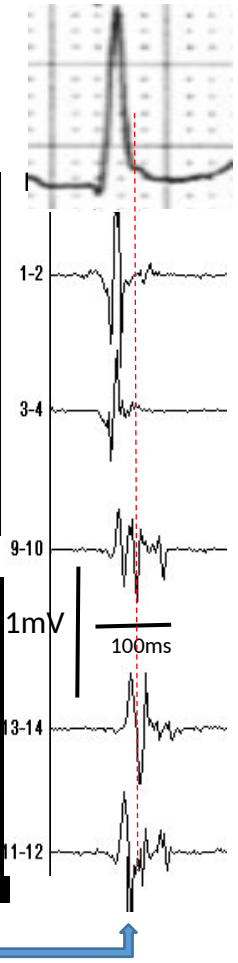
VF while taking a shower

Epicardial Voltage mapping



Epicardial Activation mapping

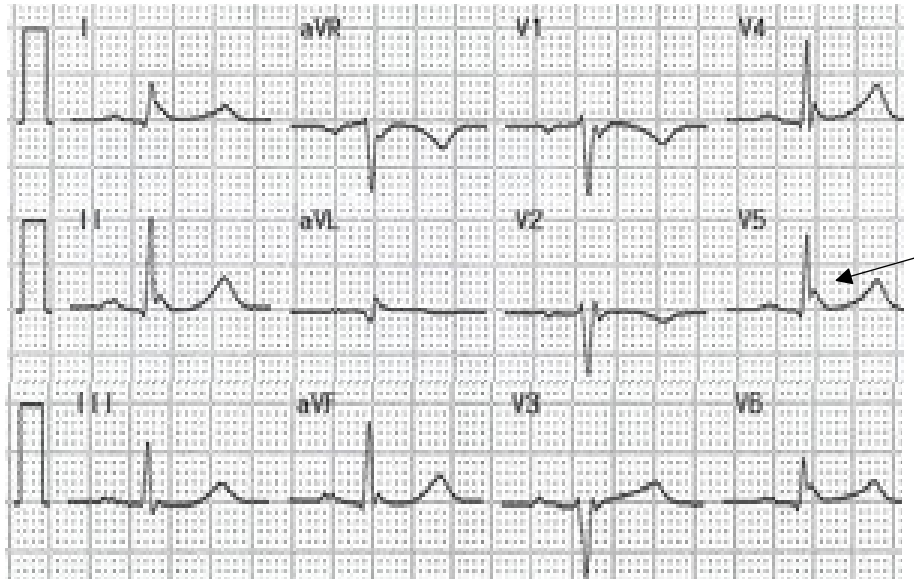




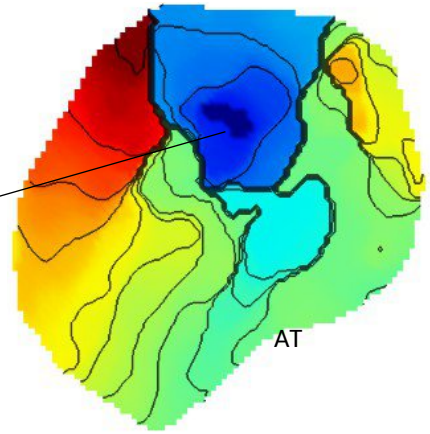
An island of delayed conduction with long fractionated electrograms
(associated here with secondary repolarization disturbances ST+)

J wave – optical mapping of 4 *ex-vivo* human hearts confirms the two main mechanisms under the same phenotype

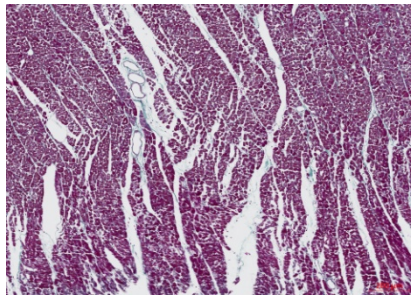
F 64a



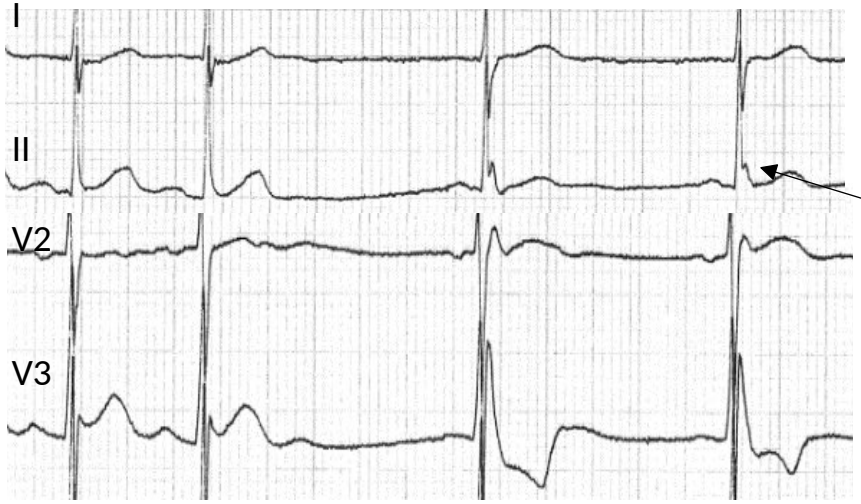
Delayed Activation of basal LV



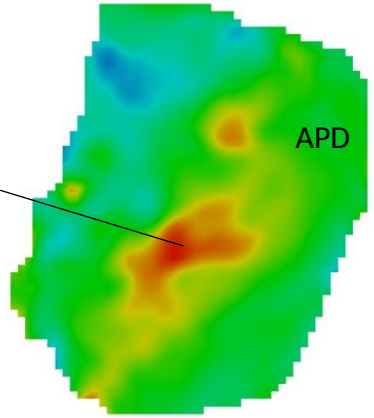
High collagen content



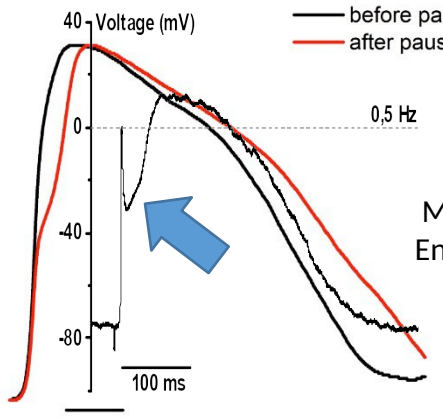
F 15a



Early repolarization LV endocardium



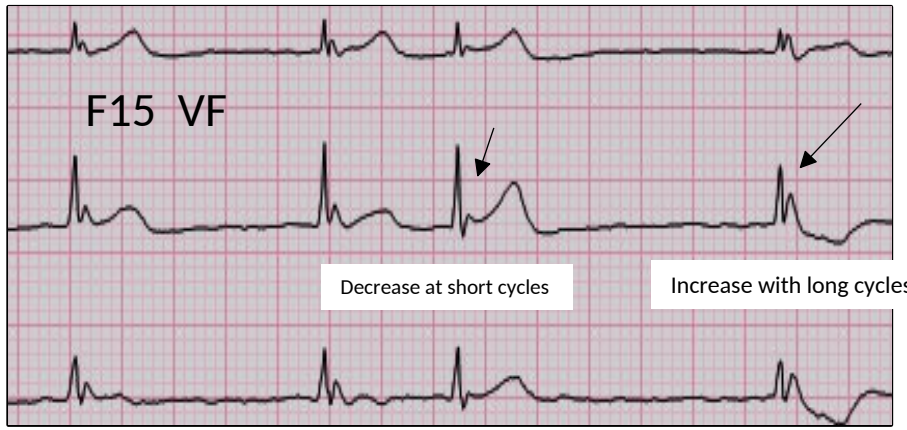
Bernus et al HRS 2019



Microelectrode Endo-LV

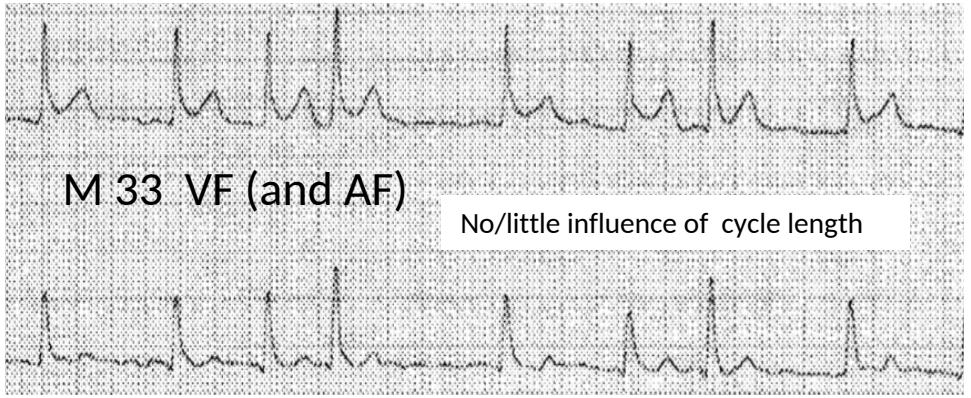
Electrocardiographic distinction of Inferolateral J waves

True Early repolarization



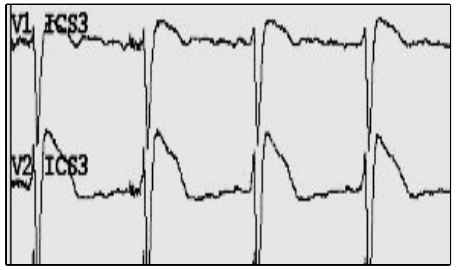
Influence of cycle length /rate

Late depolarization (delayed conduction)

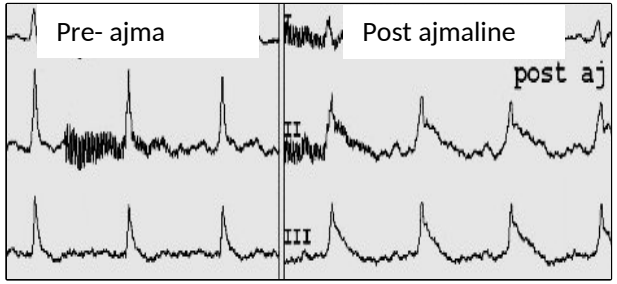


Sodium-Blocker infusion

Brugada sd (ST elevation in V1-V2) in a subset



'Inferior ST elevation in a subset

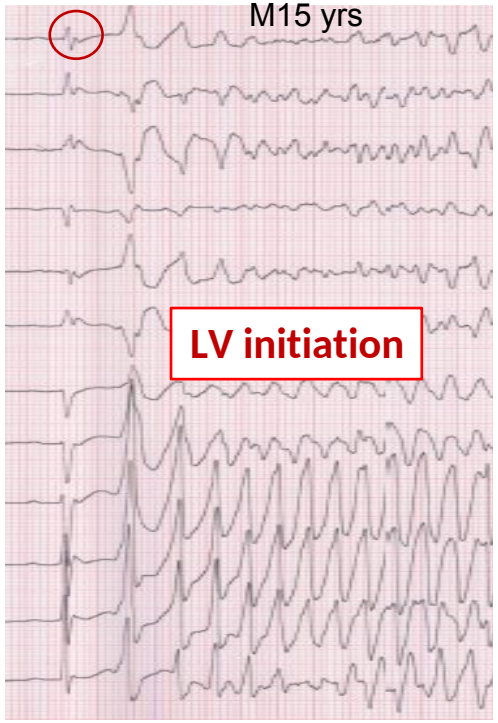


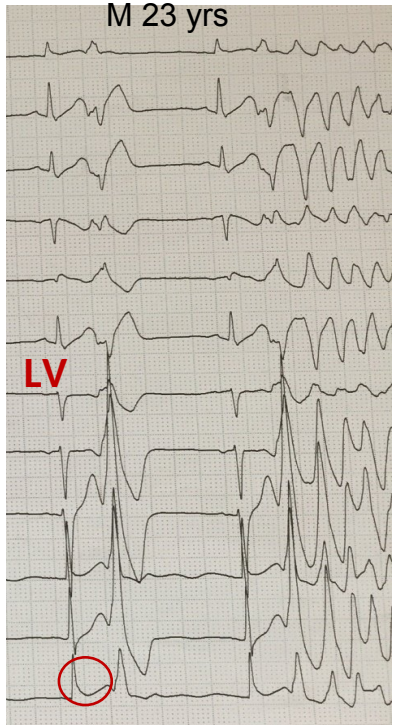
Thailand 22 patients (Pr Nademanee) : BrugadaSd in 18/22 Inferior ST in 6

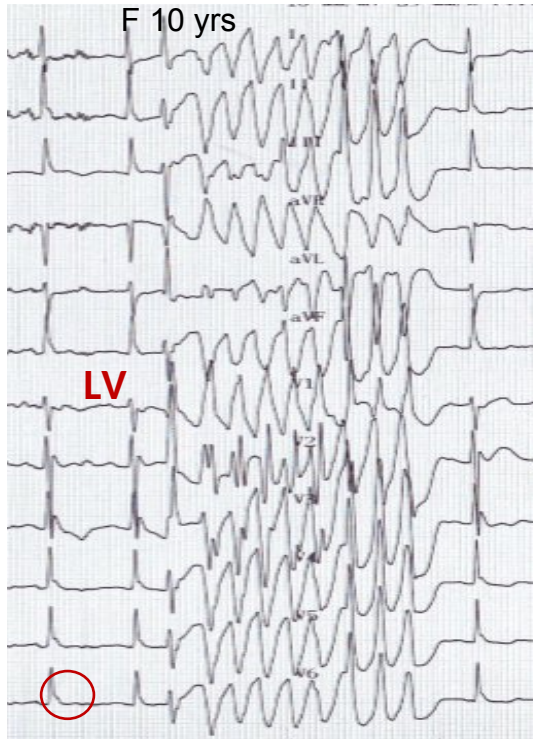
(Most had SCN5A mutation)

Spontaneous VF documented in 30 patients (multicenter)

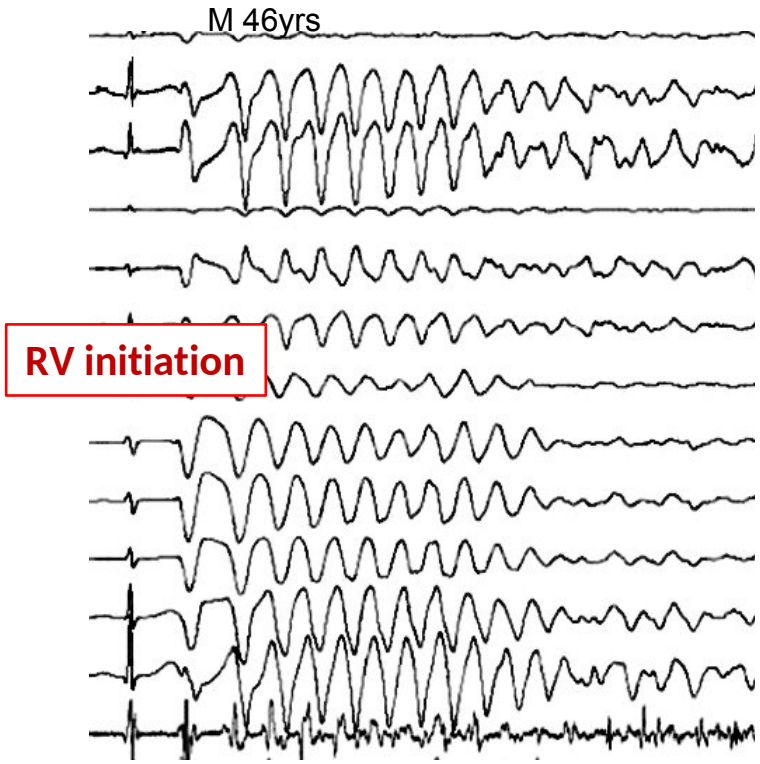
Triggers from LV when inferolateral Jw (I aVL V5 V6)







Triggers from RV when inferior Jw (only II III aVF)



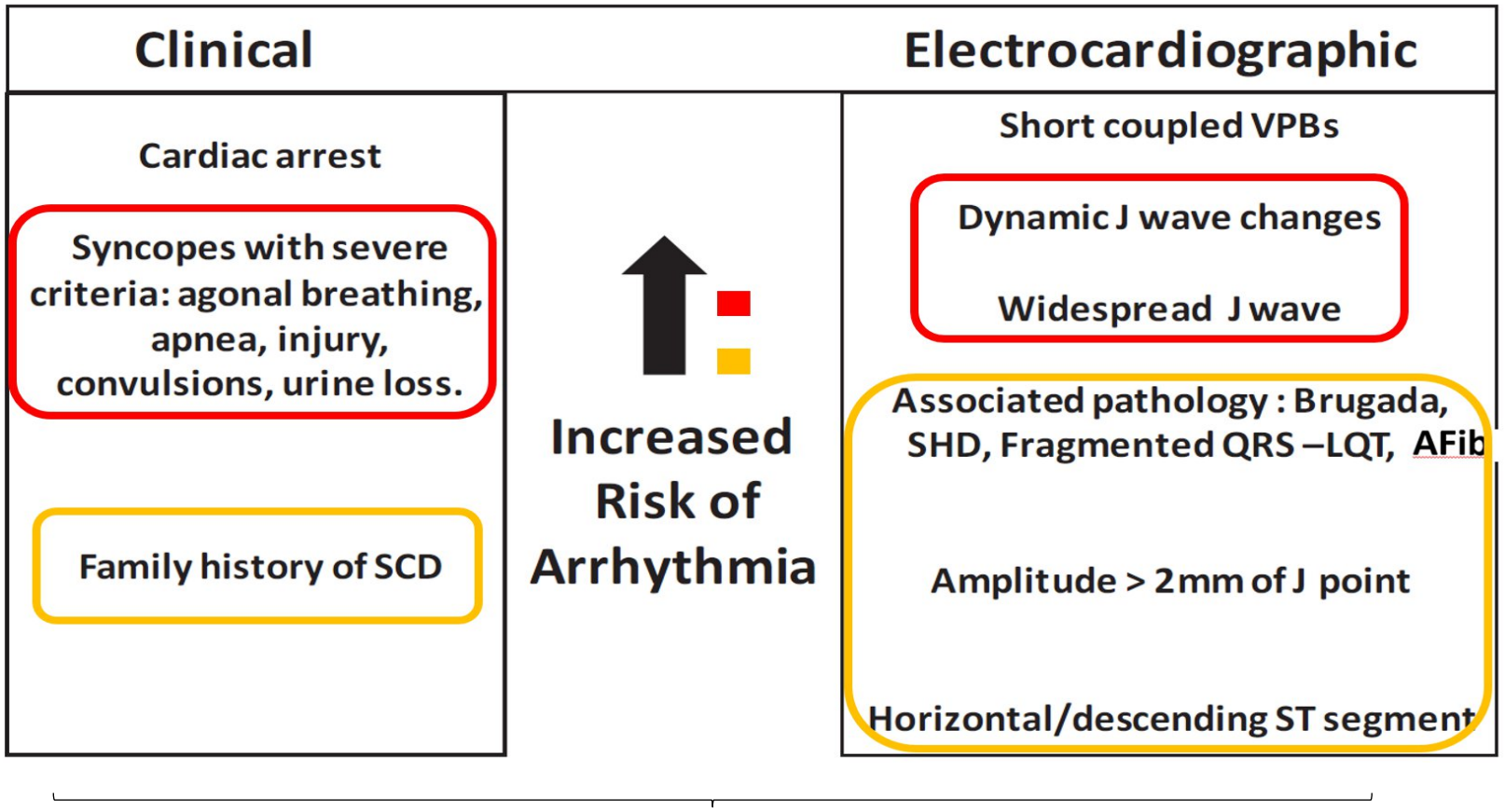
VF is much more rapid in true early repolarization

VF cycle length (10 sec)	Early repolarization N=6		Abnormal conduction N=10	
Right Ventricle	151 ±6 ms	400bpm	177 ±4 ms	345bpm
Left Ventricle	146 ±5 ms		173±2 ms	

Slower VF on ICD monitoring may be used to indicate the type of underlying substrate

Inferolateral J waves : which patients at risk ?
Patients with Syncope

No drug test is available **EPS: VF inducibility is not predictive** **Genetic analysis :little utility**



LOW risk: Clinical FUp

INTERMEDIATE: Loop Recorder

HIGH risk: Quinidine – ICD

Inferolateral J waves : Outcome of patients with syncope

Syncope clinically considered as severe , implanted with a device (Kamakura T et al HR journal 2022)

Prospective registry :143 patients (124 M,34yrs) with ER and syncope (2009-14)

Initial examination >> 13 ICD and 84 loop recorders (97 pts, 68%); one pt* refused device

FUp 97 pts for 5.5 years: 16 Arrhythmias with syncope (5 VF , 10 Bradycardias, and 1 SVT)

16 pts with syncope and normal ECG (reflex syncope)

One SCD*

In brief : 24% of causes identified on device and 6% VF in 5 yr (1%/yr) .

Conclusion: VAs occurred in 5-6% of patients with an ER pattern and syncope in 5 yrs . Same pts may have arrhythmic and reflex syncope.

Most specific sign : Dynamicity of J wave (here with bradycardia)

Emina.. 15 yrs

Inferolateral J waves : which patients at risk ?
What to do in asymptomatic patients

CURRENTLY NOTHING ! AS THE REAL RISK IS LOW

EP Study is useless for risk stratification
Possibly in the future , insights from specific pharmacological test or repolarization maps or genetic screening

In individual situations (large and dynamic J wave + family history of SCD) , Quinidine as an option

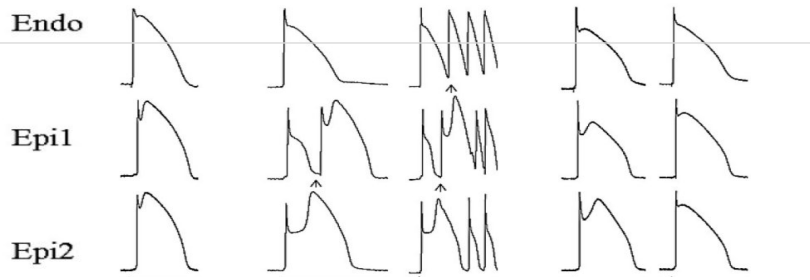
Inferolateral J waves : Treatments in Symptomatic patients

Early repolarization

VF storm/emergency : Isoproterenol Quinidine

AAD treatment : Quinidine

Catheter Ablation of VF triggers (Purkinje)



Baseline Ito Agonist + Ca block Normalize on Ito Blocker

e robinier faux acacia - (Mrs. Gemstone / flickr.com)

Acacetin suppresses the electrocardiographic and arrhythmic manifestations of the J wave syndromes

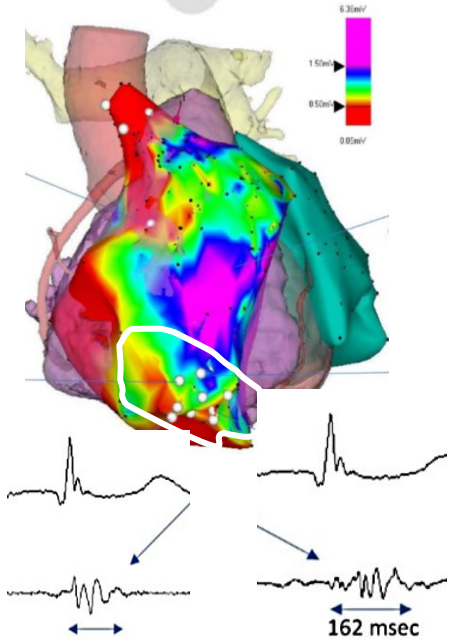
José M. Di Diego[1†*], Bence Patocskai[2‡], Hector Barajas-Martinez[1‡], Virág Borbáth[3], Michael J. Ackerman[4], Alexander Burashnikov[1,5], Jérôme Clatot[1], Gui-Rong Li[6], Victoria M. Robinson[1,7], Dan Hu[8], Charles Antzelevitch[1,5,9*]

Abnormal delayed Conduction

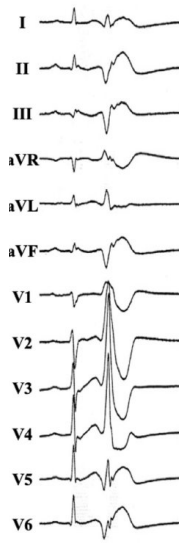
AAD treatment : Quinidine ? Other AADs ?

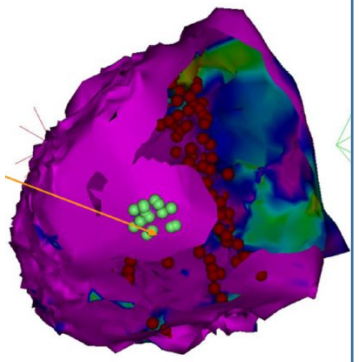
Catheter Ablation of VF triggers (Myocardial) and Substrate (Mostly Epicardial) : Both share the same region

Inferior RV Epicardium



Inferior Lateral LV Epicardium





TAKE HOME MESSAGES

1 Two substrates of Inferior J-waves (Repol or Depolarization): discernible on ECG

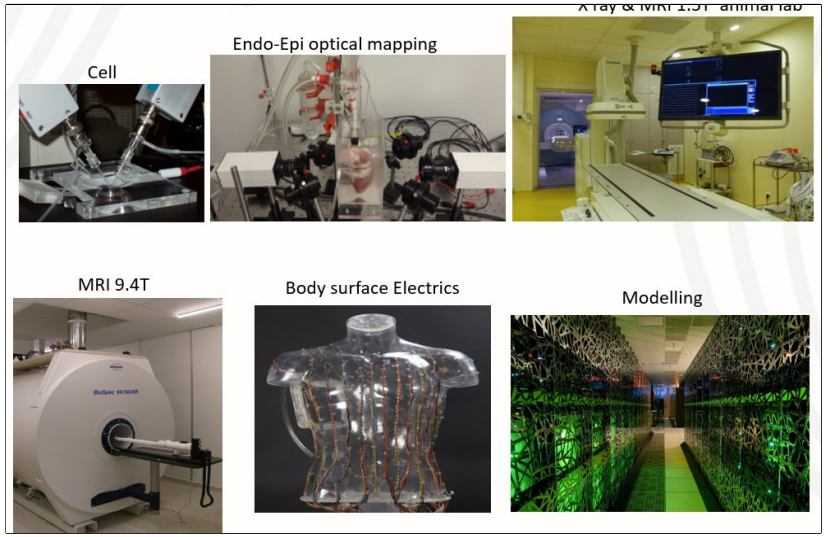
2 The presence of inferior J wave –with underlying heart disease- increases the arrhythmic risk

3 The risk assessment of ERS/J-wave remains difficult – but ILR are helpful in the cases of unexplained syncope

4 Treatments (Quinidine, Ablation) are effective

Members from 18 countries : Cardiac electrophysiology-Imaging-Signal processing-Modeling …

LIRYC : Cardiac Electrophysiology & Modeling Institute



OPEN Positions for SIGNAL processing engineering in sudden cardiac death