Sudden cardiac death

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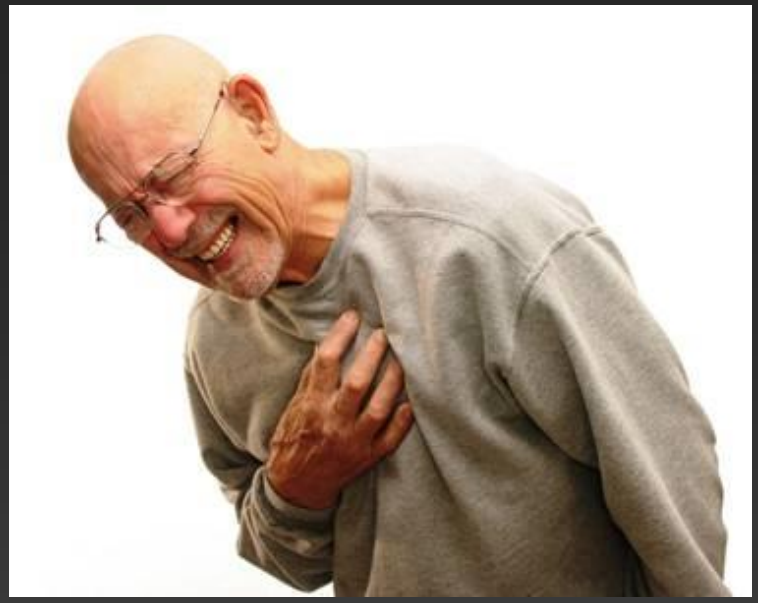
UNIVERSITAT DE BARCELONA

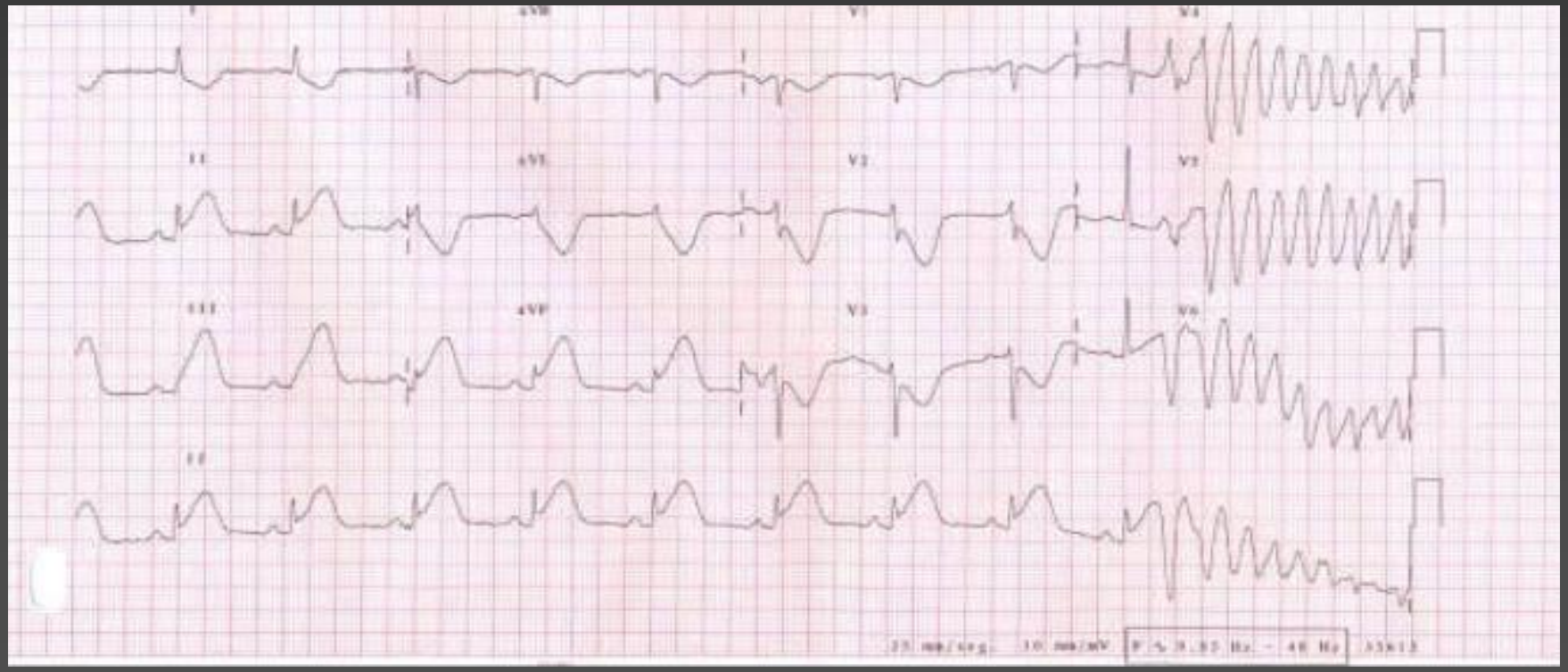
sant Joan de Déu

CLÍNIC BARCELONA

Ischemic:

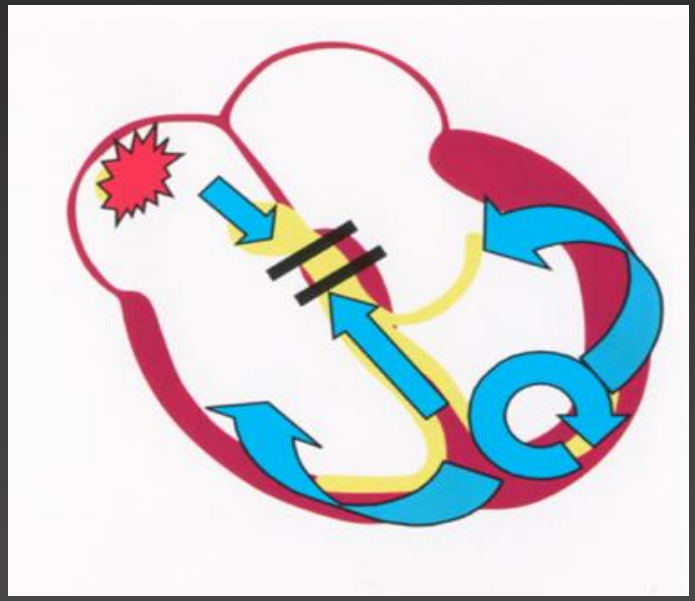
Acute phase: polymorphic VT, VF

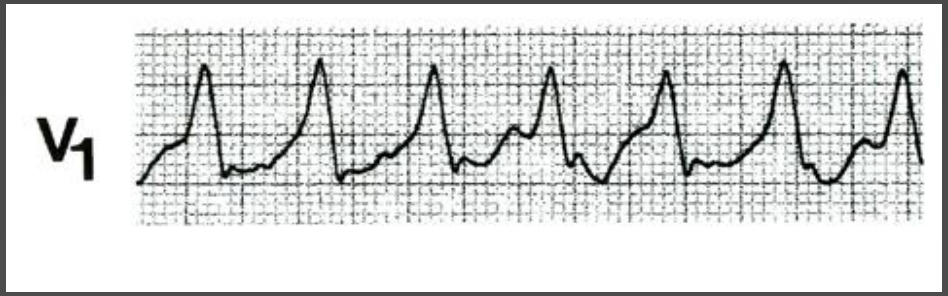


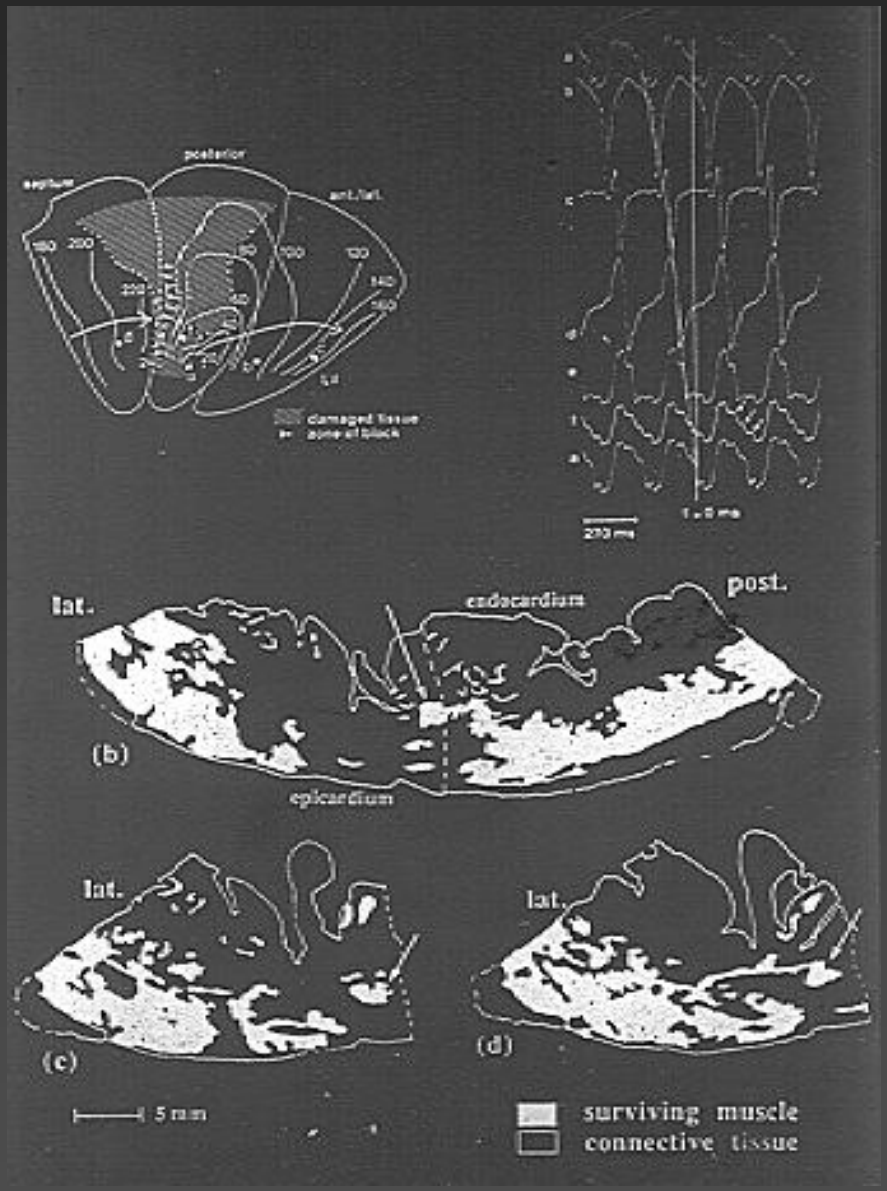


Ischemic:

Chronic phase: Monomorphic VT



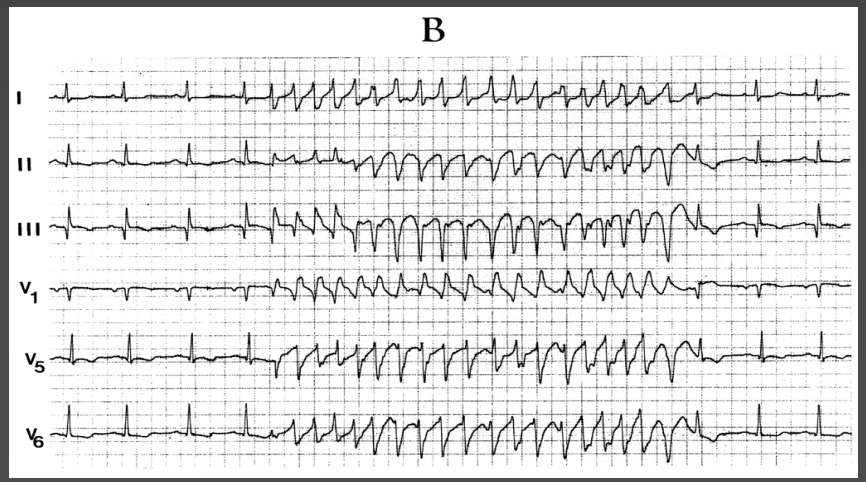


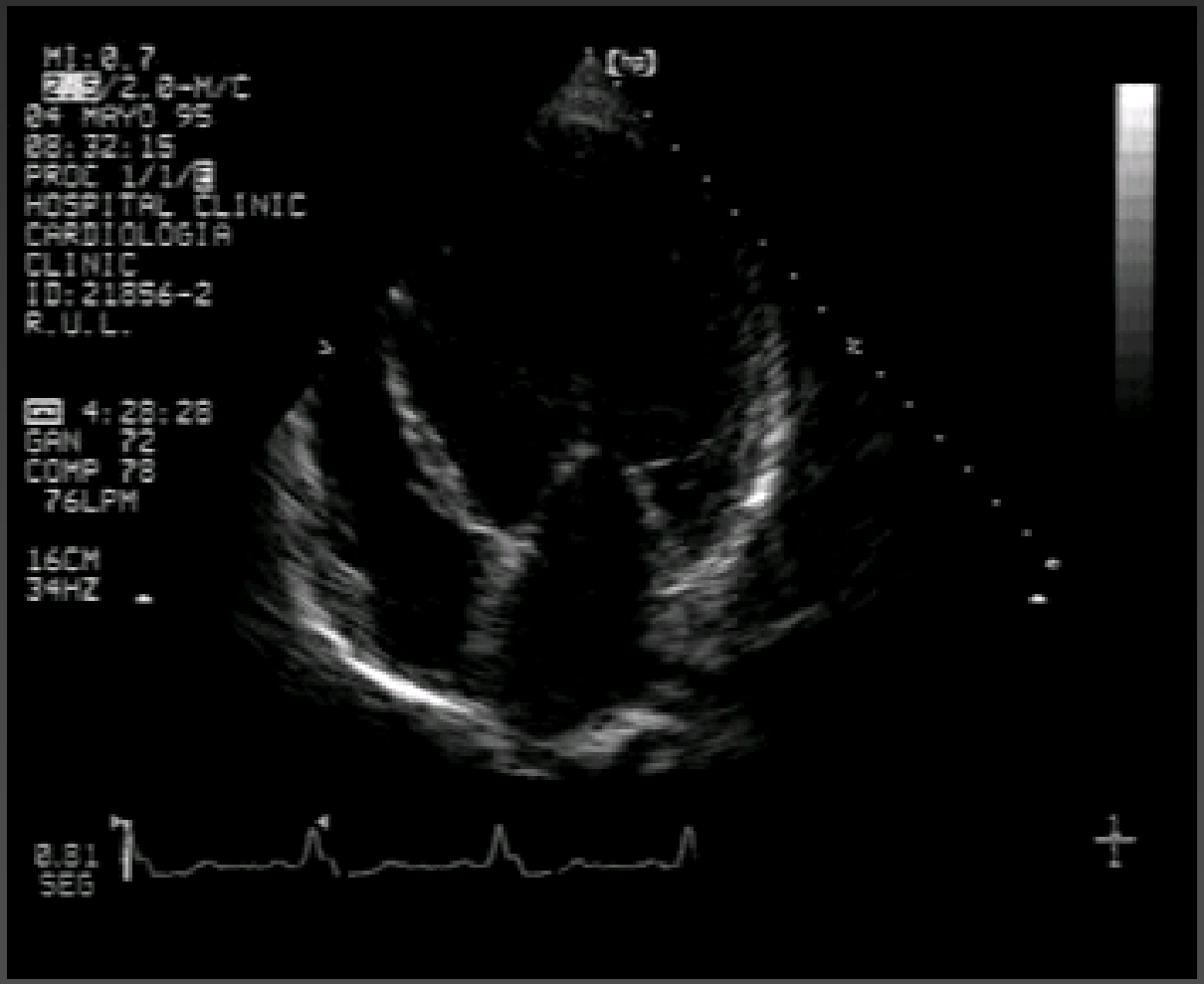


Dilated Myocardiopathy

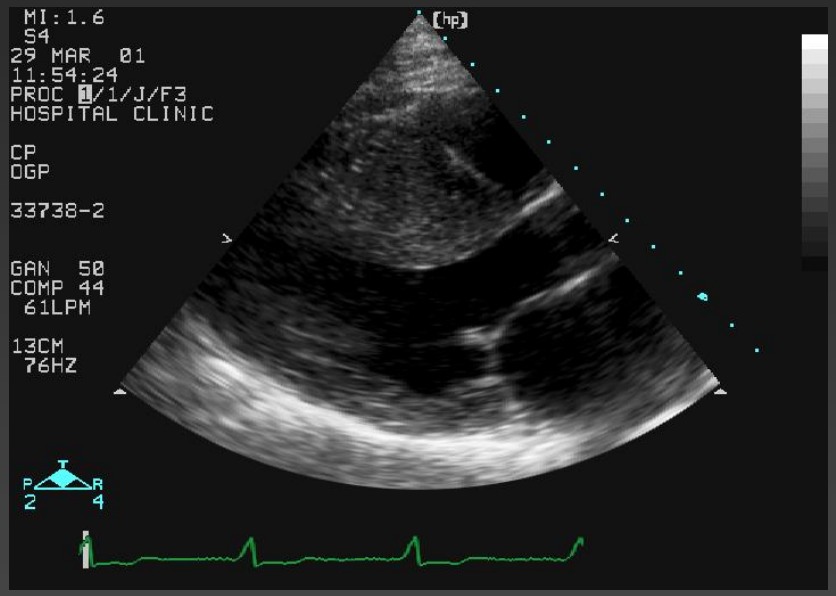
Dilated:
Polymorphic VT, SD

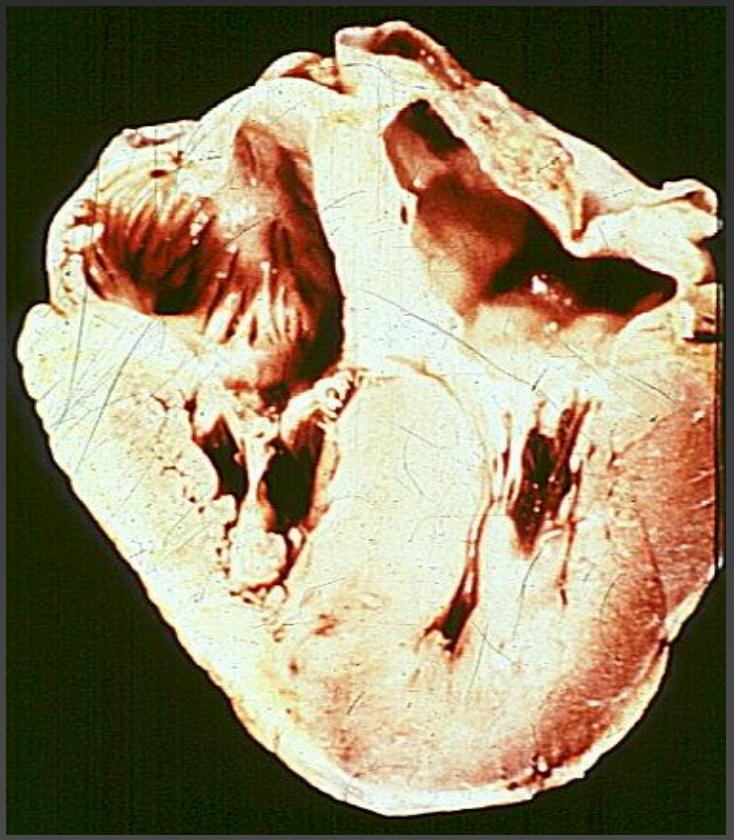
SD: responsibe for 50% of deaths in DCMP

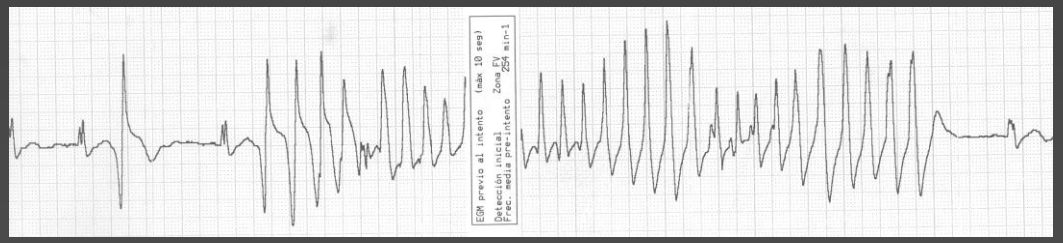




Hypertrophic Cardiomyopathy





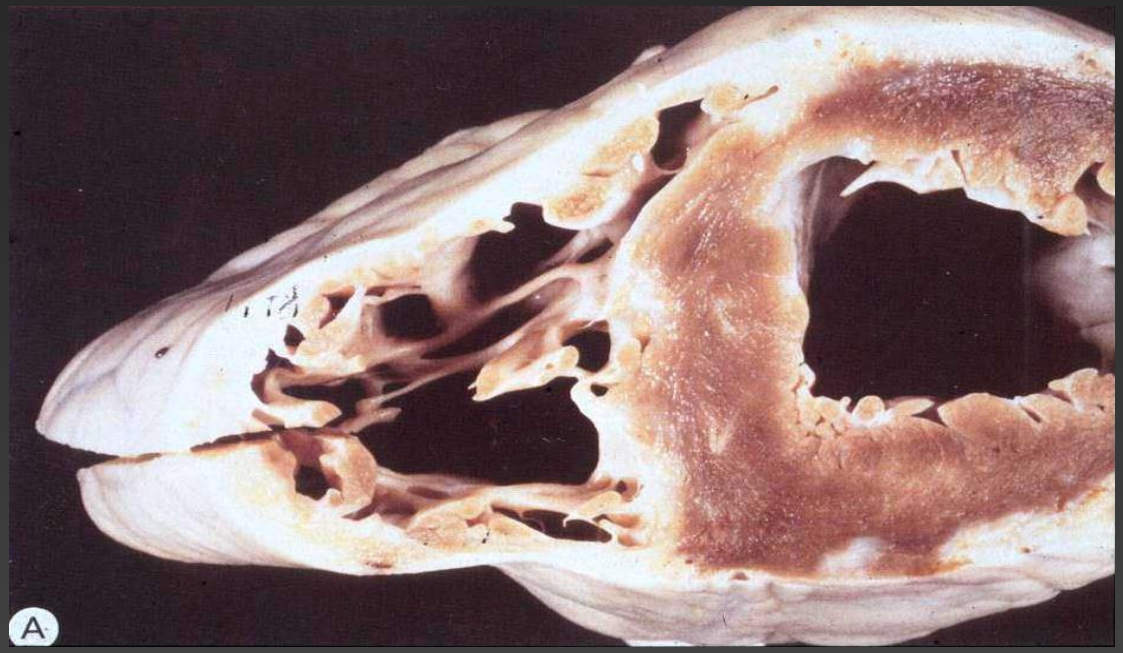


Hypertrophic:

Polymorphic VT, SD

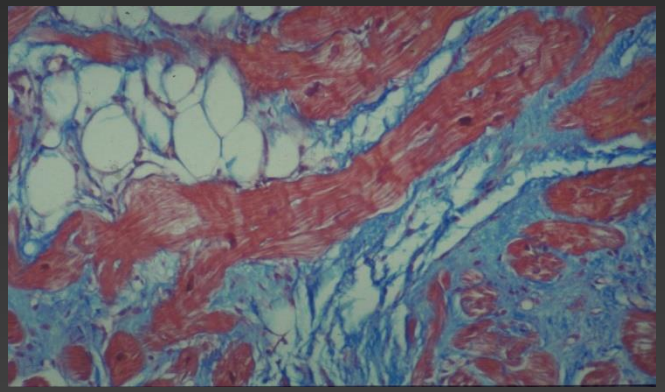
Principal cause of SD in young < 40 years

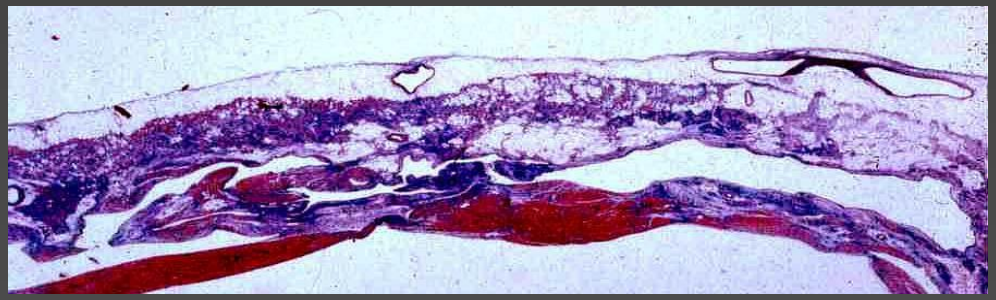
Arrhythmogenic right ventricular cardiomyopathy

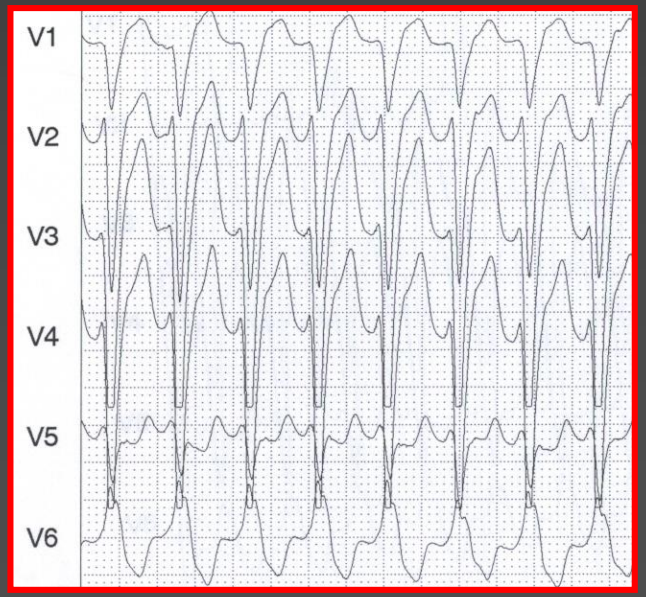


ARVC:

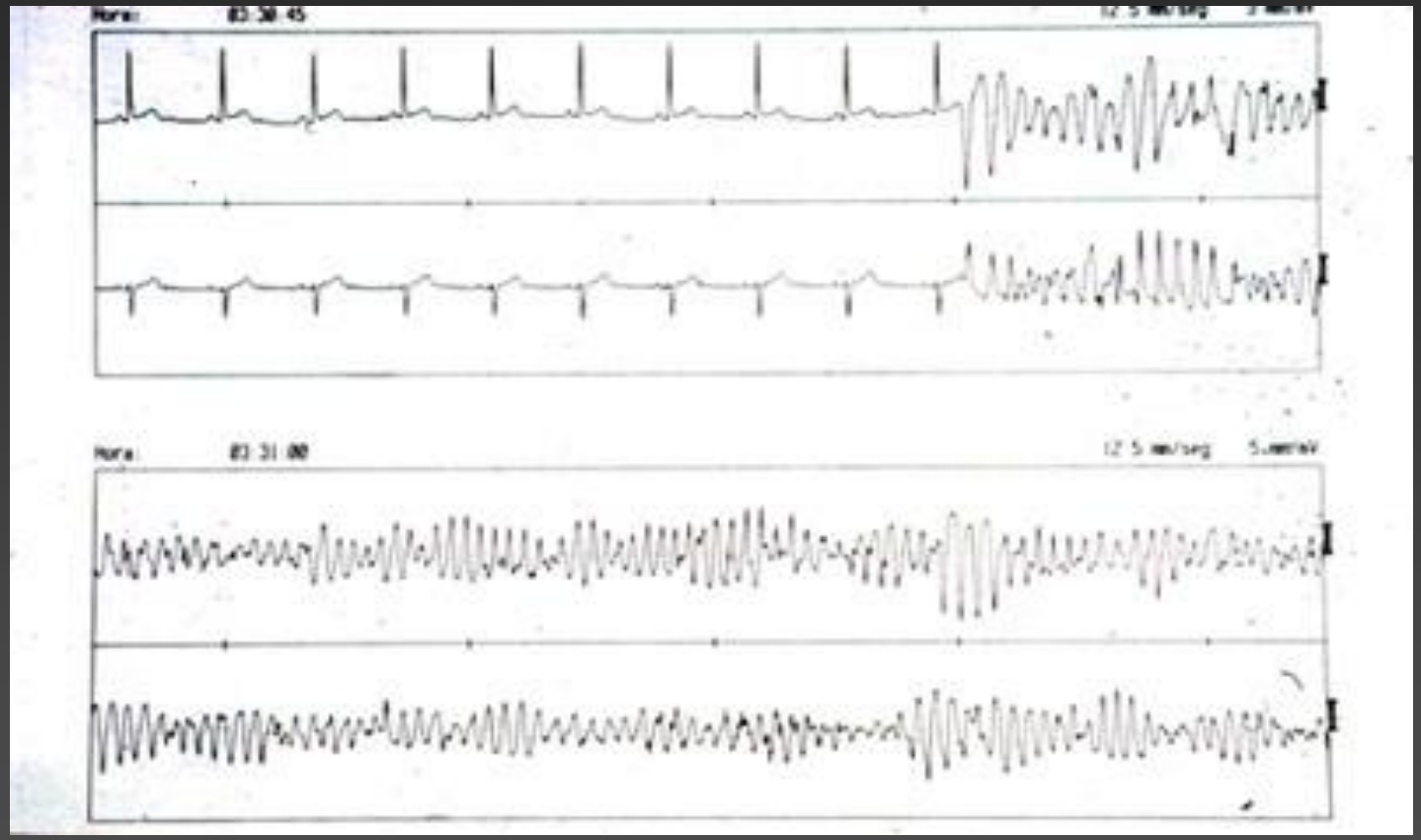
Monomorphic VT, pleomorphic VT

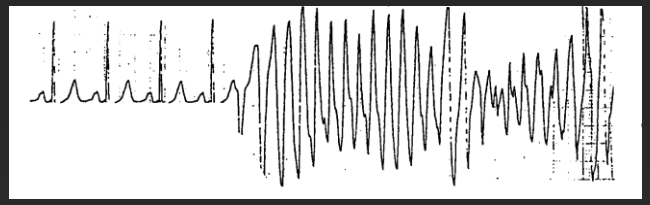






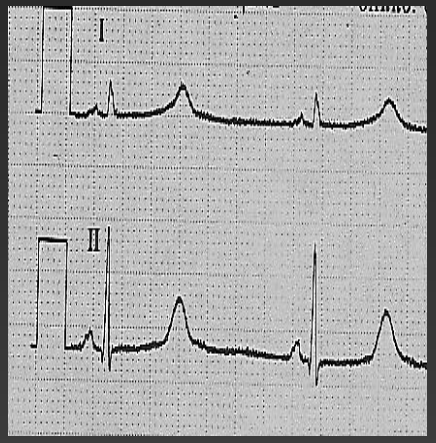
Dx- fibrofatty replacement of the right ventricle

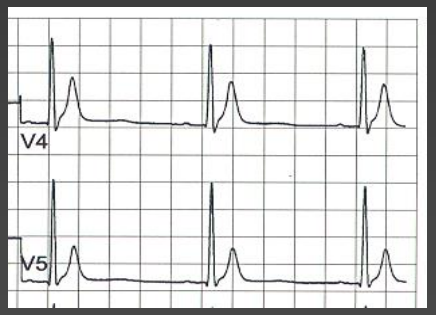


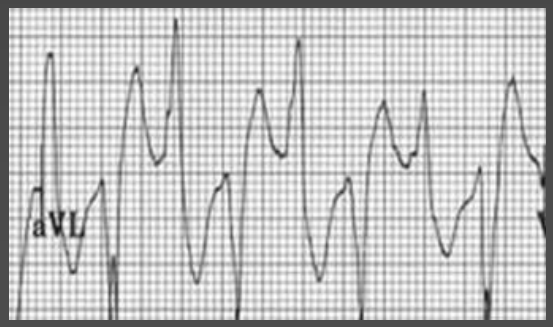


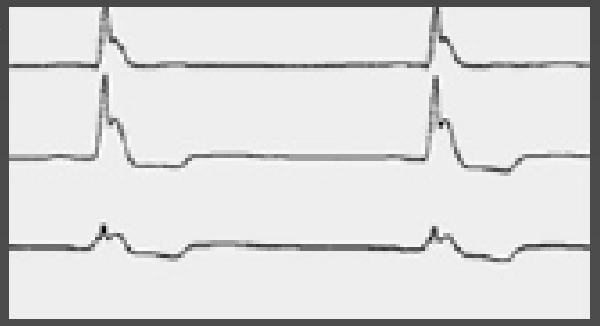
Channelopathies

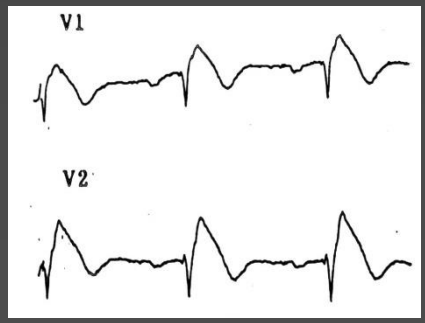
- Long QT Syndrome
- Short QT Syndrome
- Brugada Syndrome
- CPVT
- Malignant early repolarization



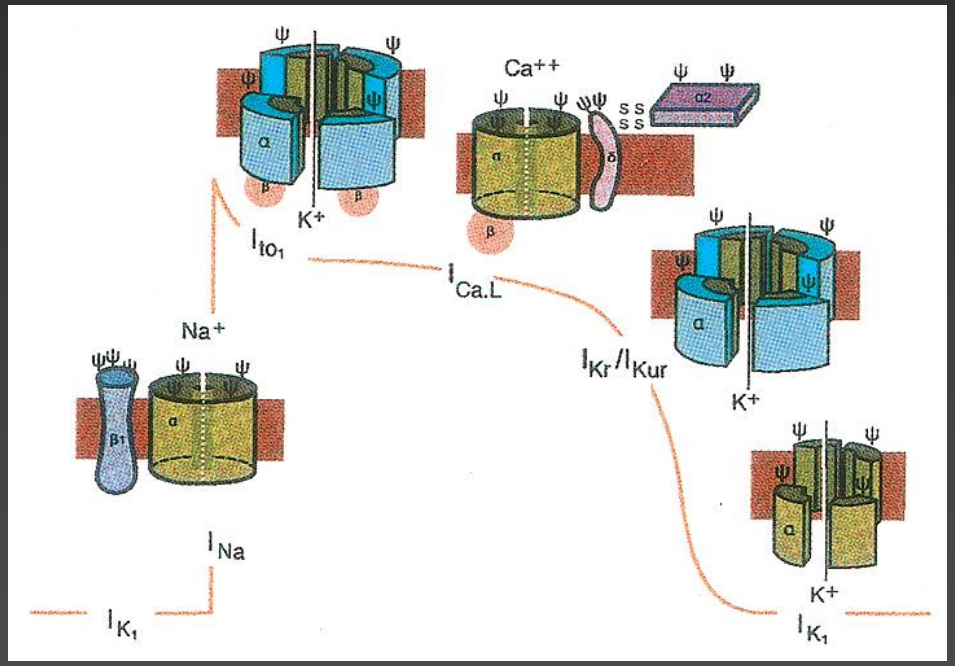








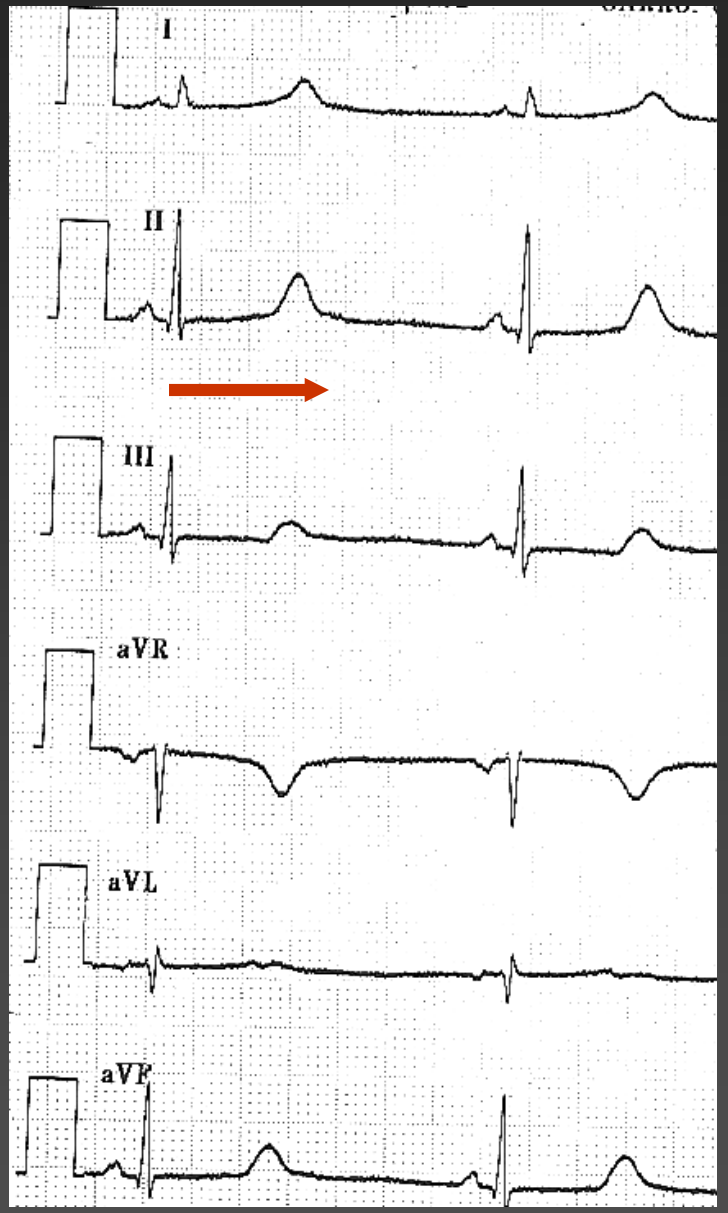
IONIC CHANNELS
Familial Arrhythmias

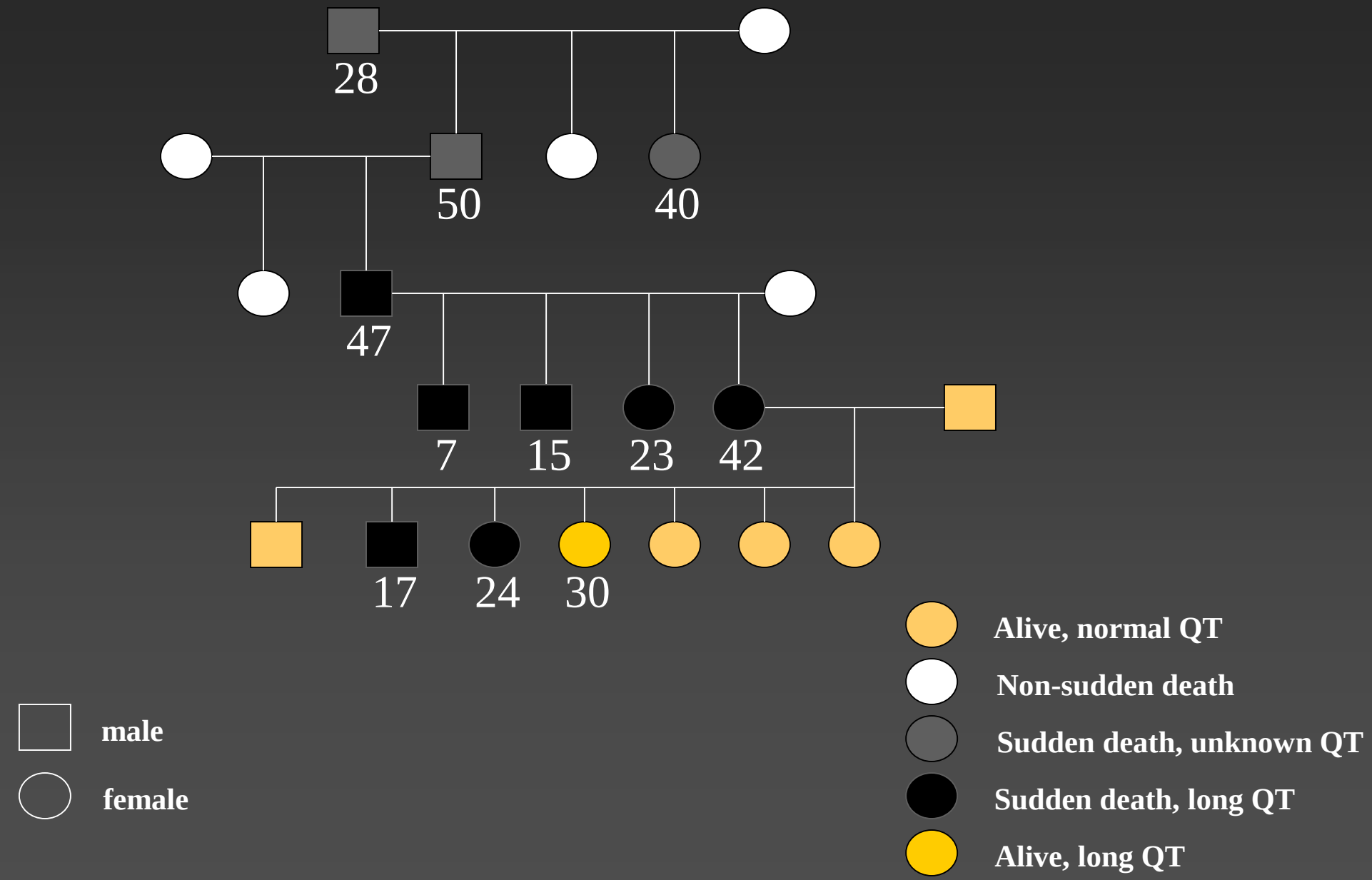


Chien K. Molecular Biology of Cardiovascular disease

QT=640 msec

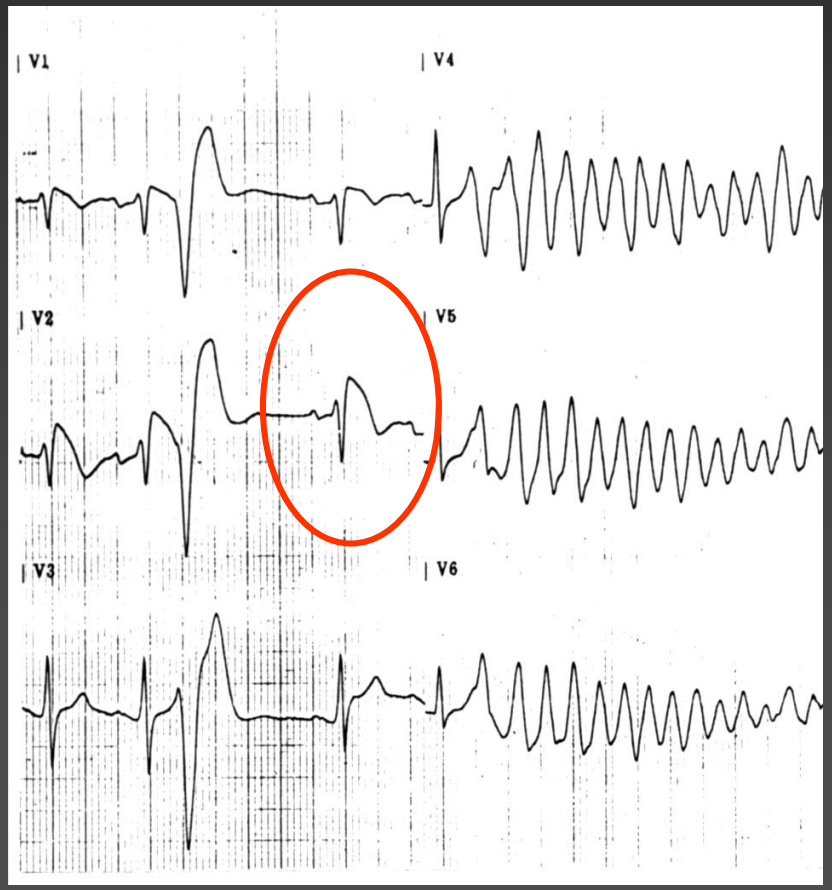
QTc=584 msec

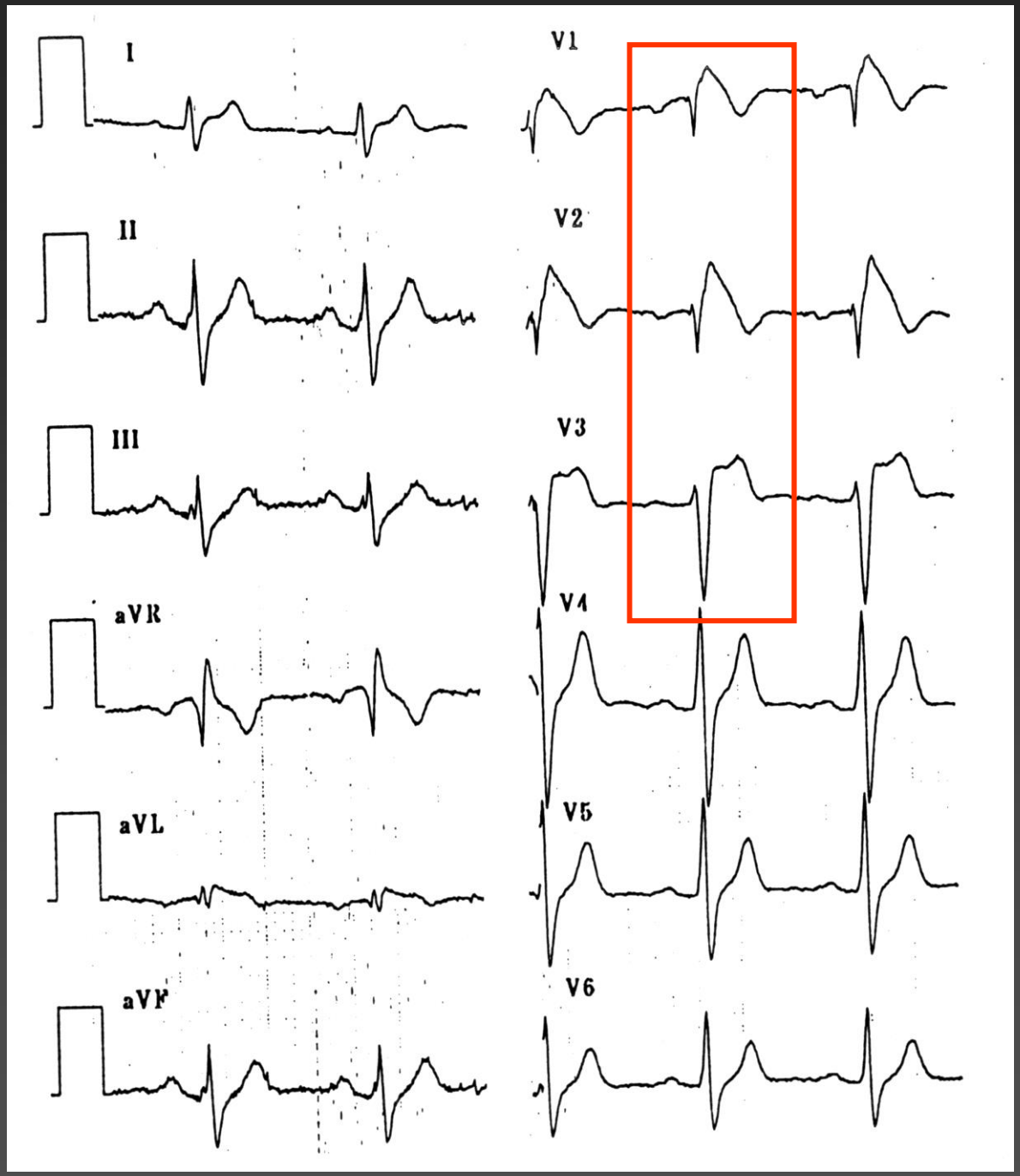



28
50
40
47
7
15
23
42
17
24
30
male
female
Alive, normal QT
Non-sudden death
Sudden death, unknown QT
Sudden death, long QT
Alive, long QT

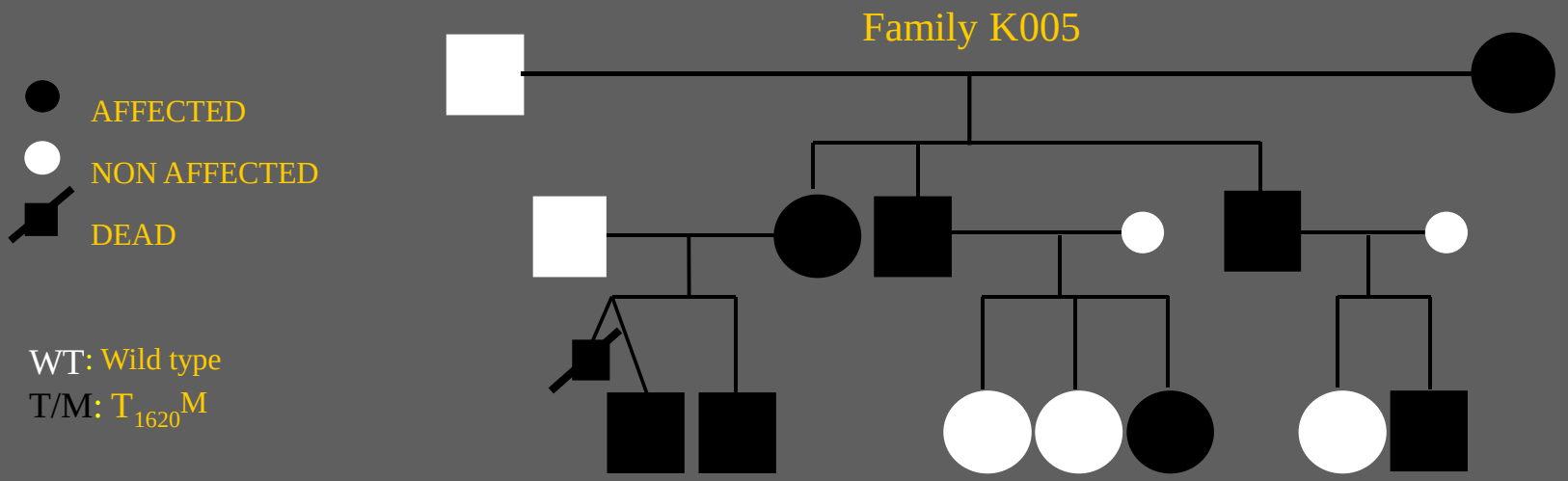
ECG in Brugada syndrome

- prolonged PR
- RBBB
- ST sement elevation
- negative T waves



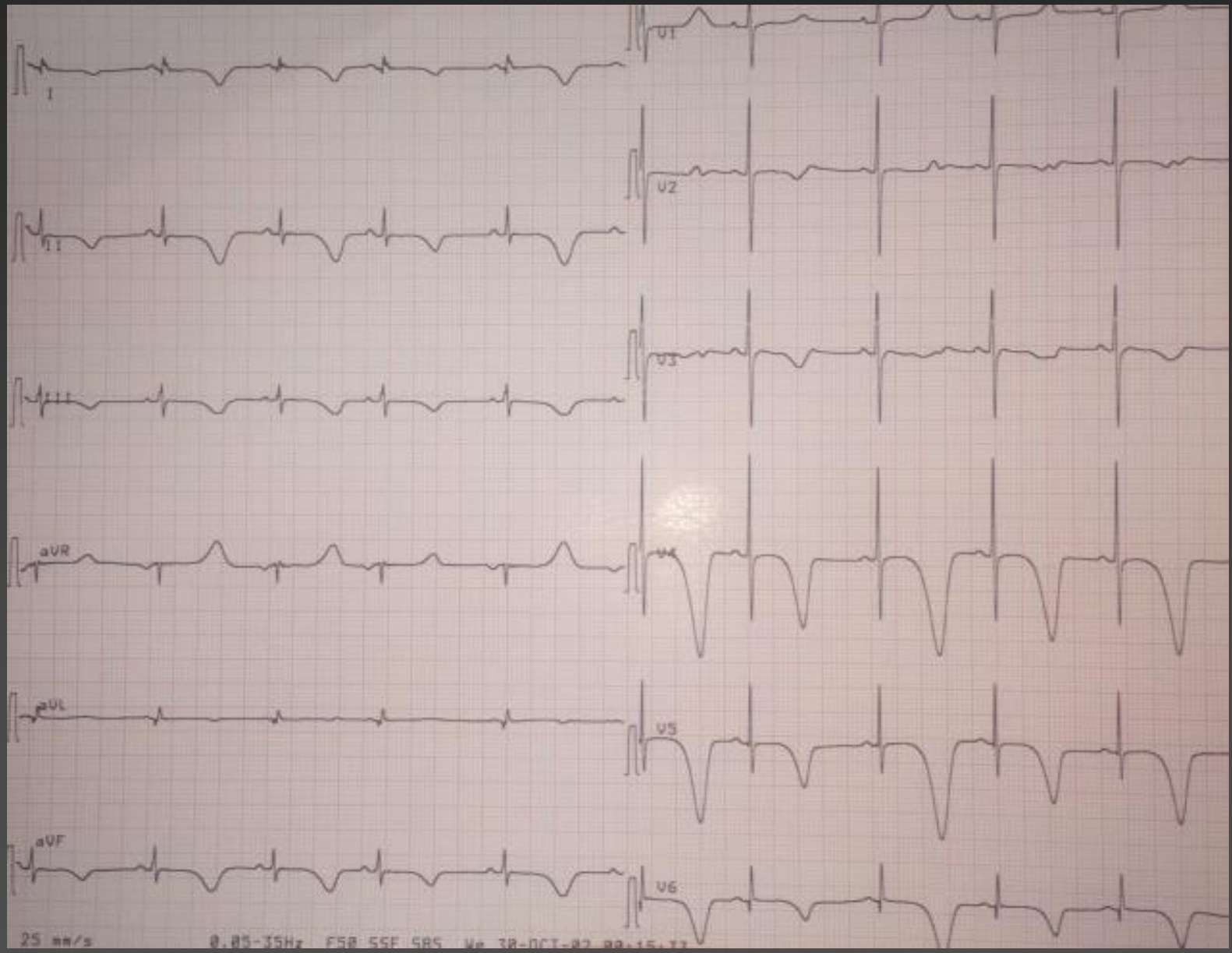


Phenotype-Genotype correlation

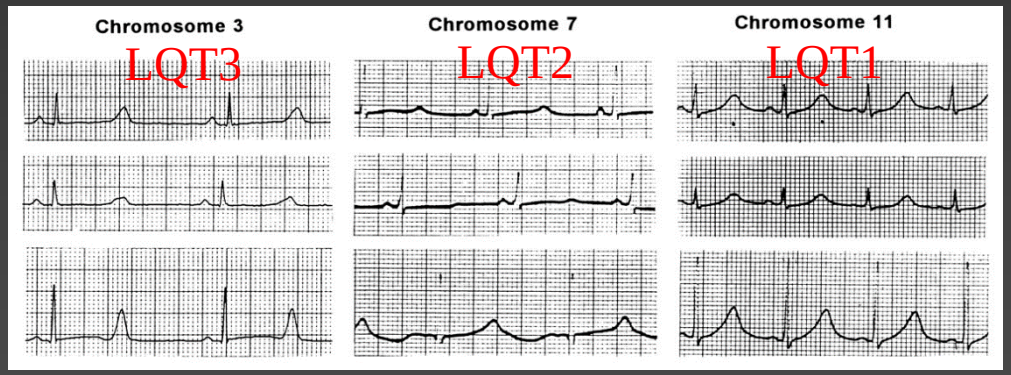


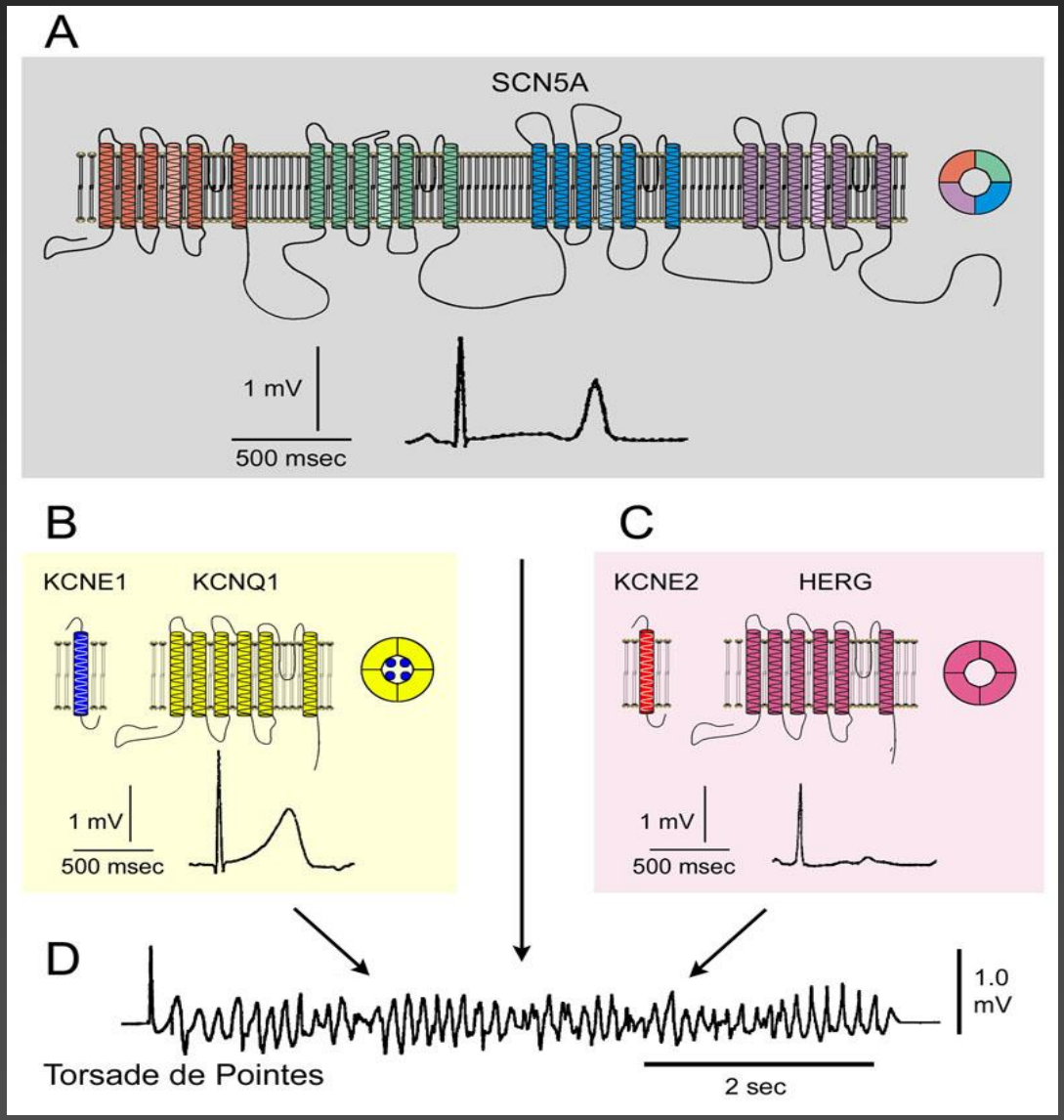
ECG AT BASELINE	-	-	-	+	-	-	-	-	-	-	-	-	+
ECG WITH AJMALINE	-	-	+		+	+	-	-	+	N/A	-	N/A	
MUTATION ANALYSIS	WT	WT	T/M	T/M	T/M	T/M	WT	WT	T/M	T/M	WT	T/M	T/M

POTASSIUM CHANNELS

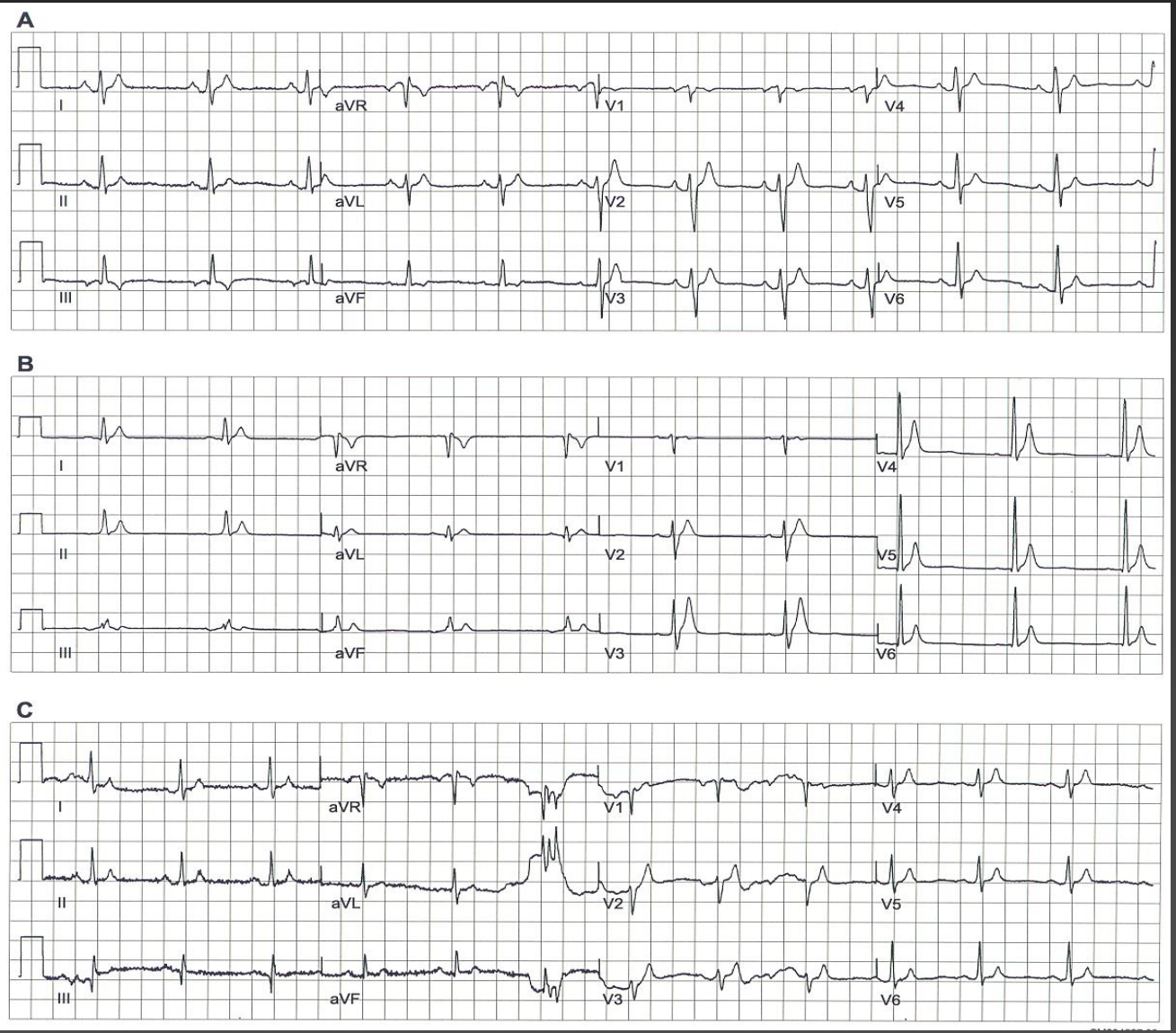


genotype – phenotype correlation





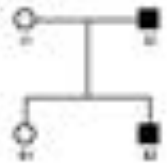
Short QT syndrome

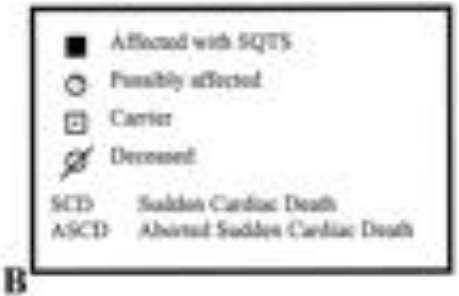


Gussak I, Brugada P, Brugada J, Cardiology 2000

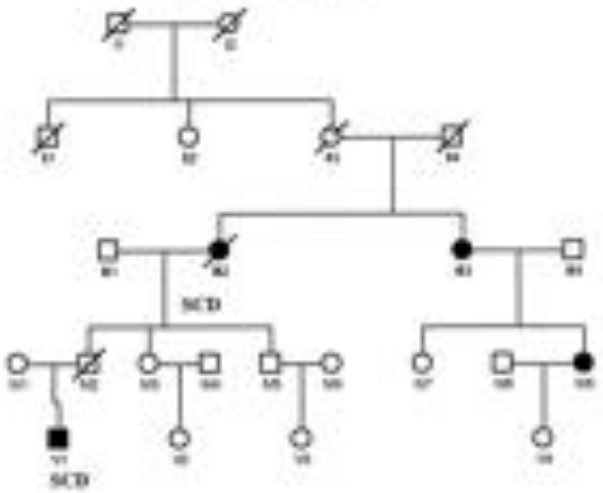
A

FAMILY 30-339

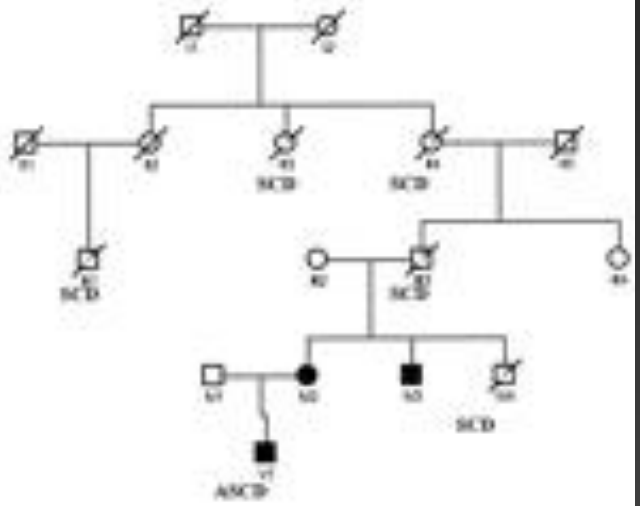




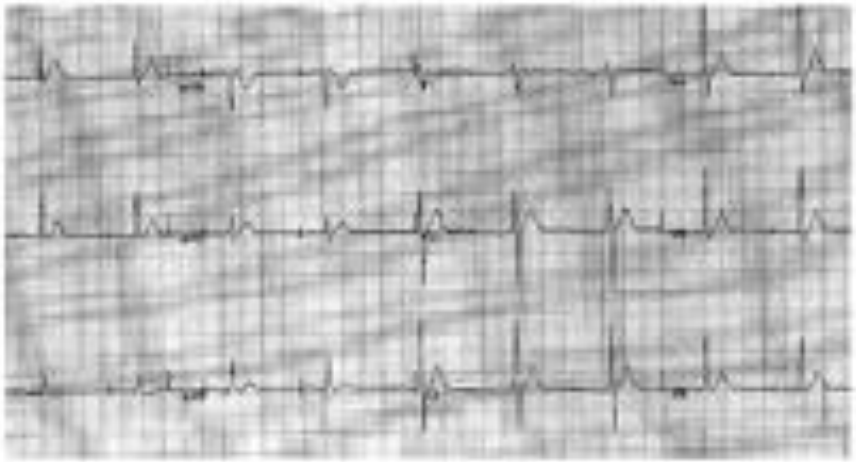
FAMILY 30-371



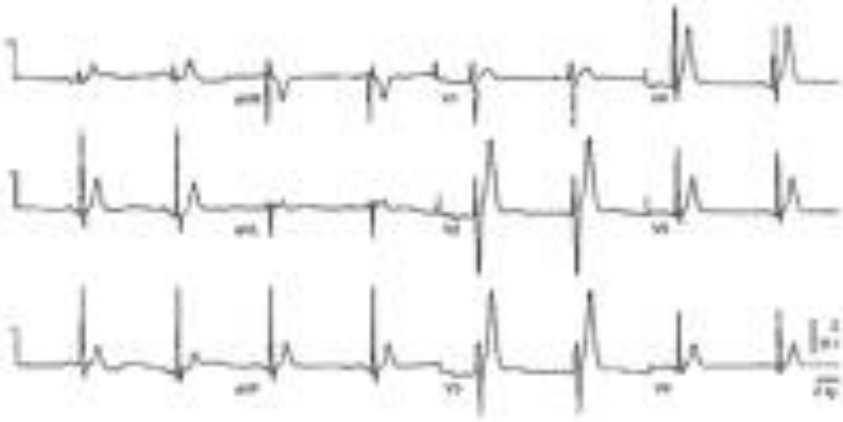
FAMILY 30-335



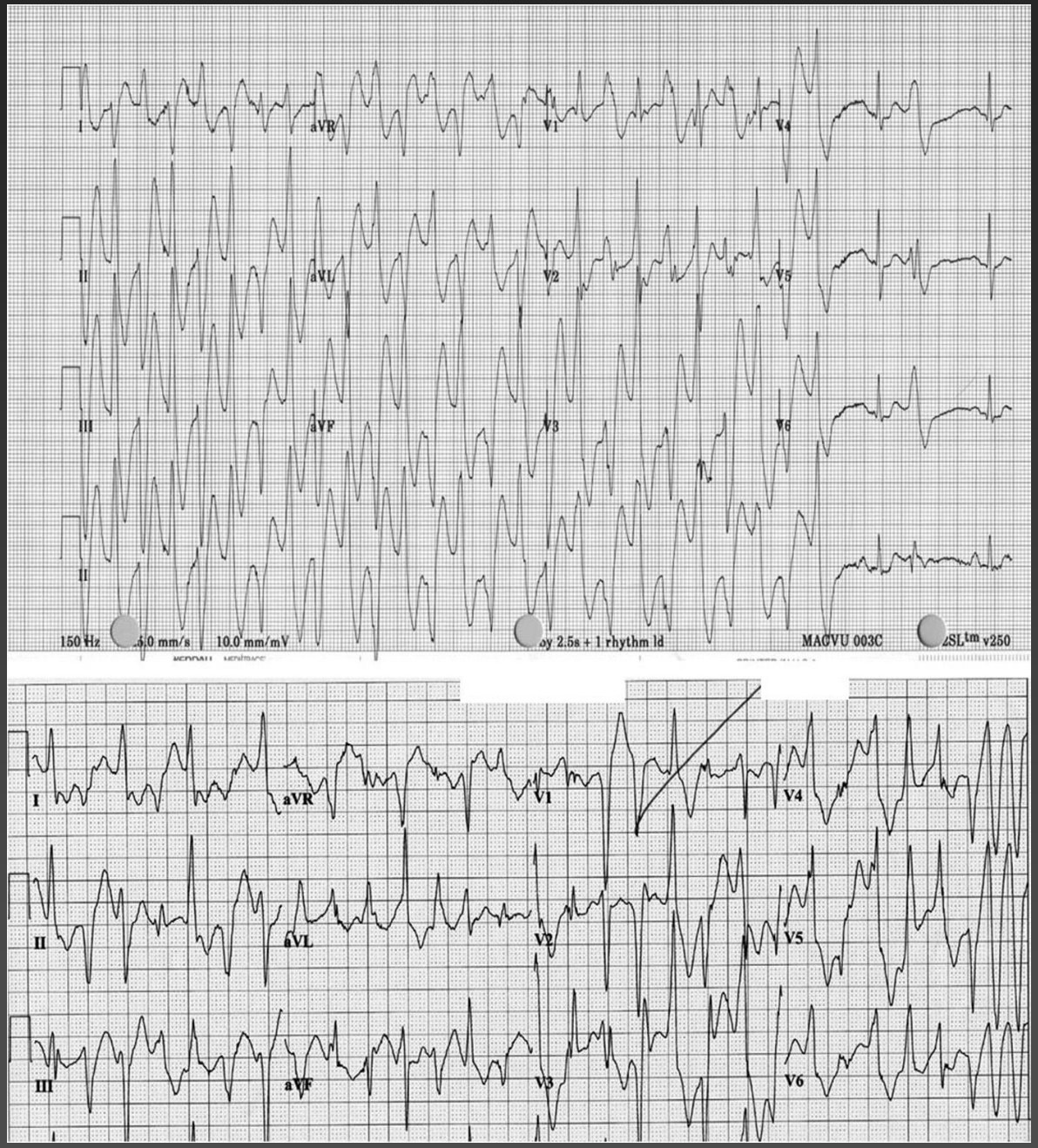
B



FAMILY 30-339, PATIENT I:2
QTc 288 msec



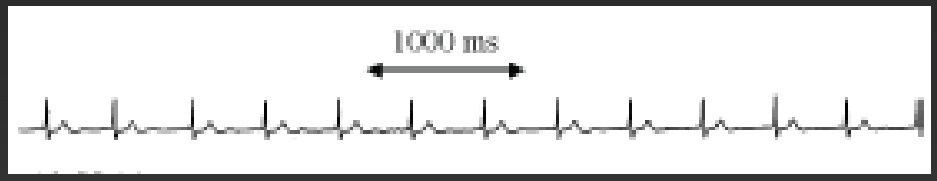
FAMILY 30-339: PATIENT II:2
QTc 293 msec


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
150 Hz
10.0 mm/mV
MACVU 003C
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6

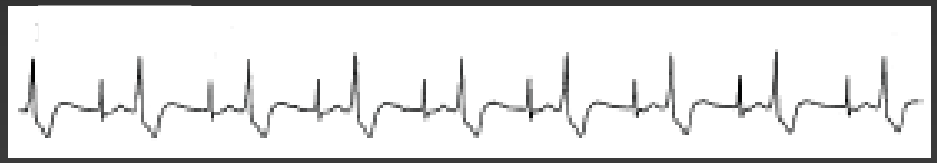
Familial Cathecholaminergic Ventricular Tachycardia

Stress test

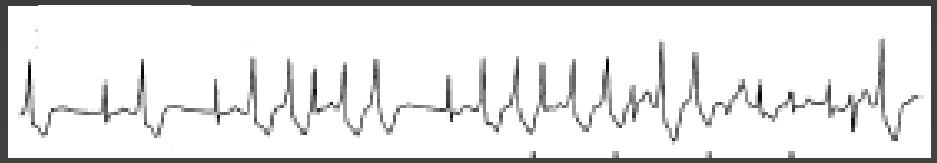
basal

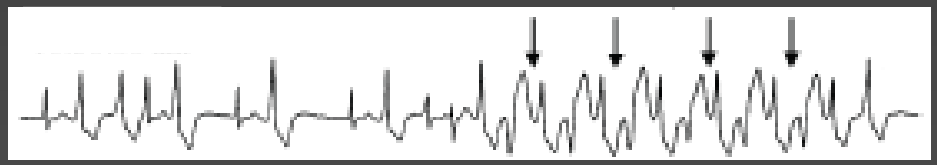


Begining

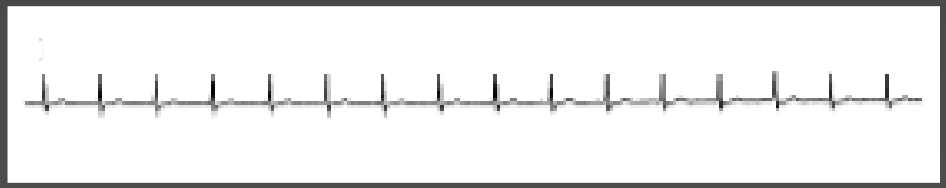


Progression





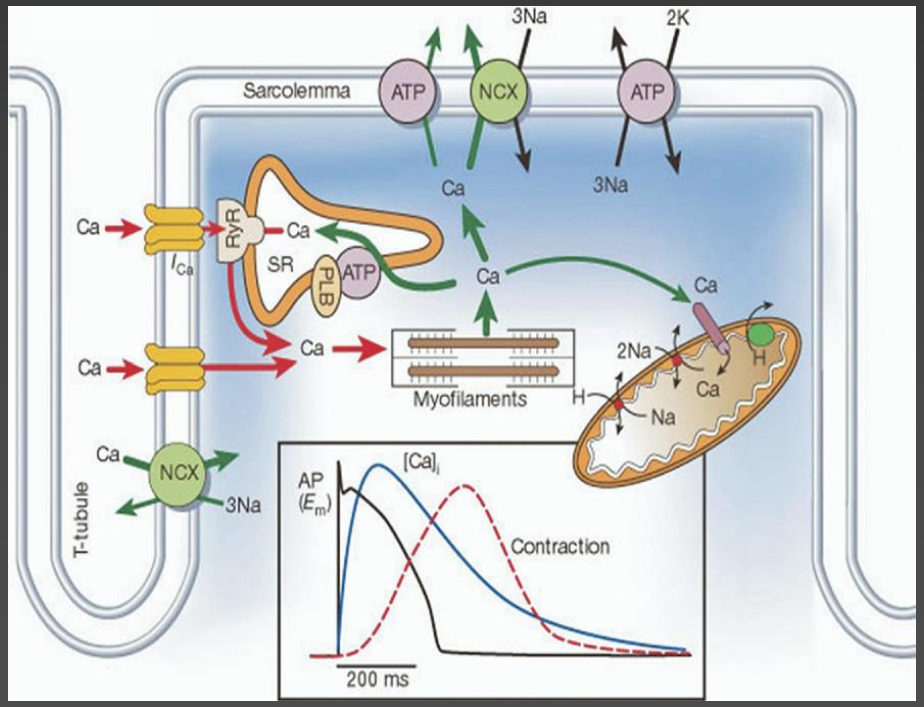
Recovery

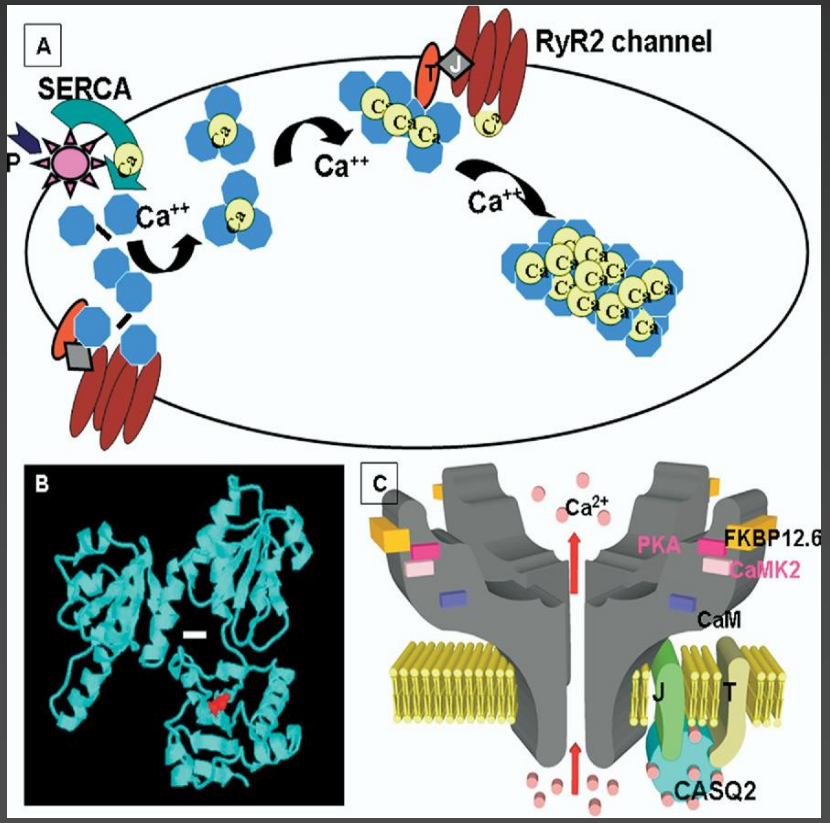


Ngai-Shing Mok et al. Chinese Medical Journal 2006;119(24):2129-2133

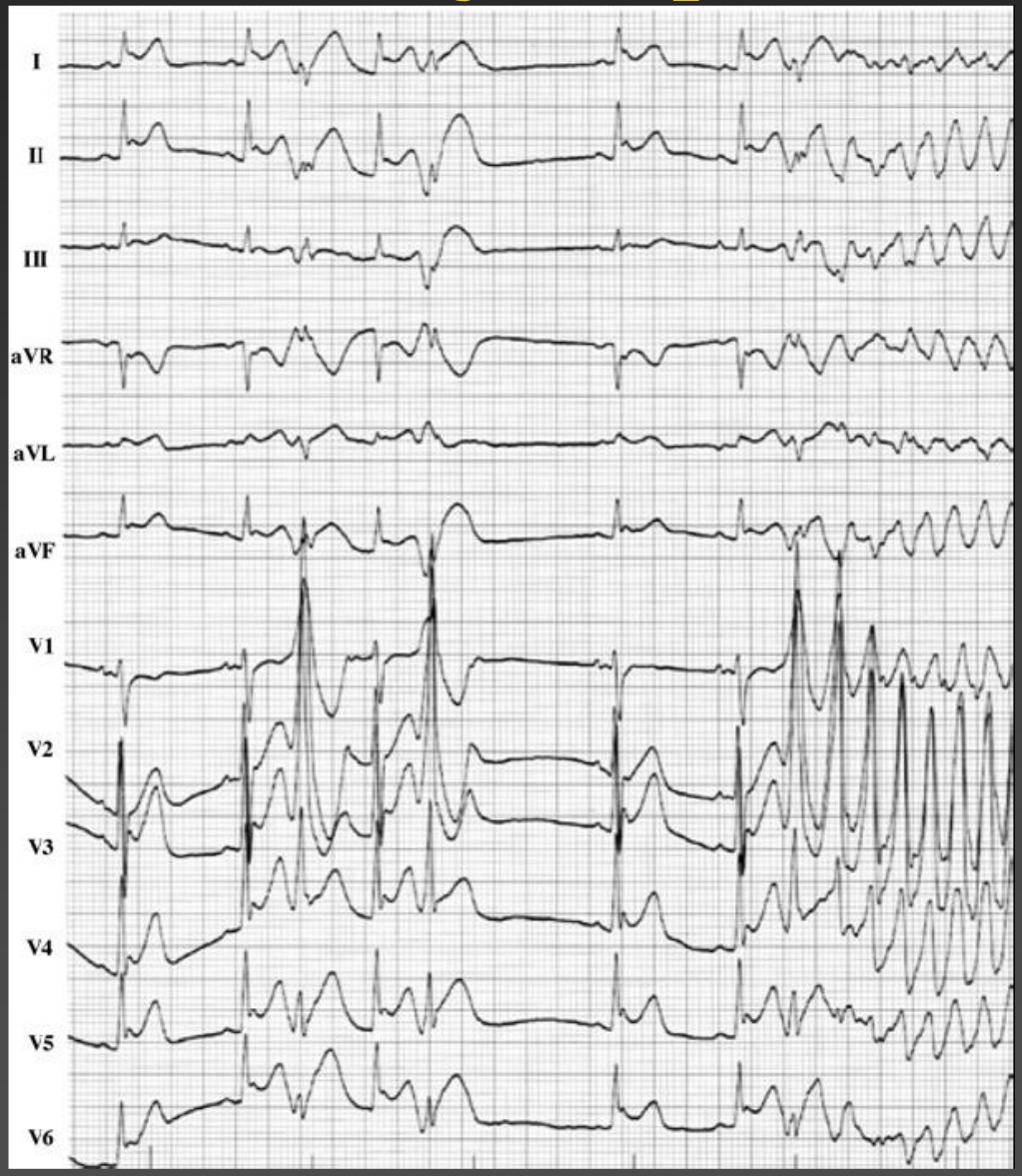
Familial Polymorphyc VT

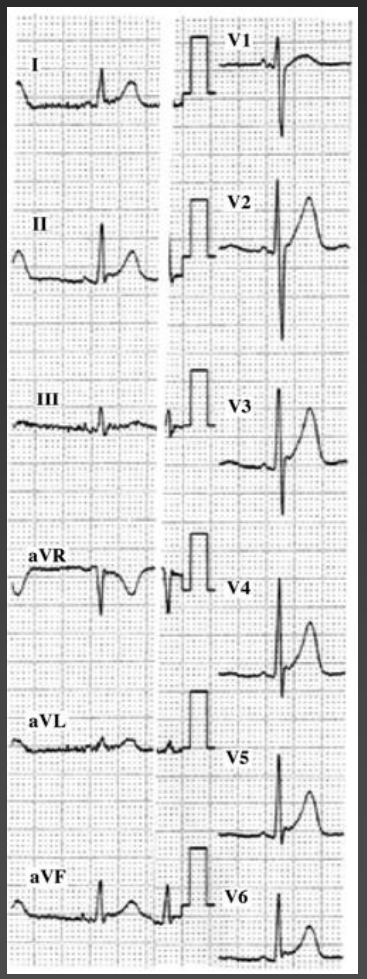
- **Rianodine (RyR2) mutatios responsible for 50% and calsequestrine mutations responsible for 2% of cases. Both genes are responsible for cardiac cellular calcium hemostasis**



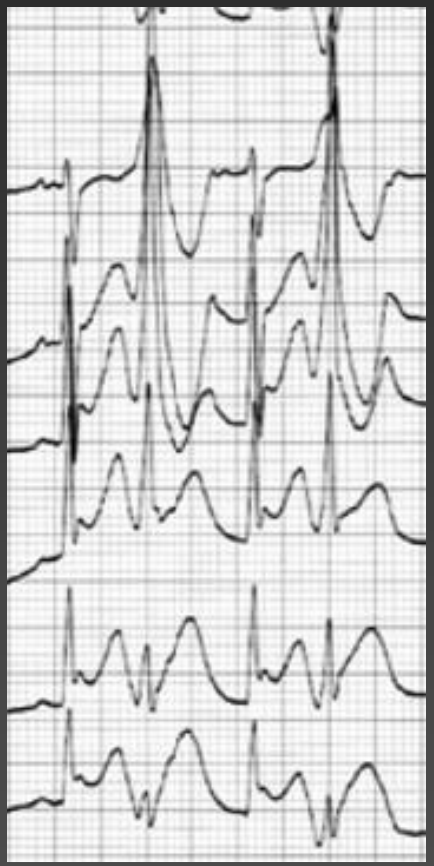


Early Repolarization and VF

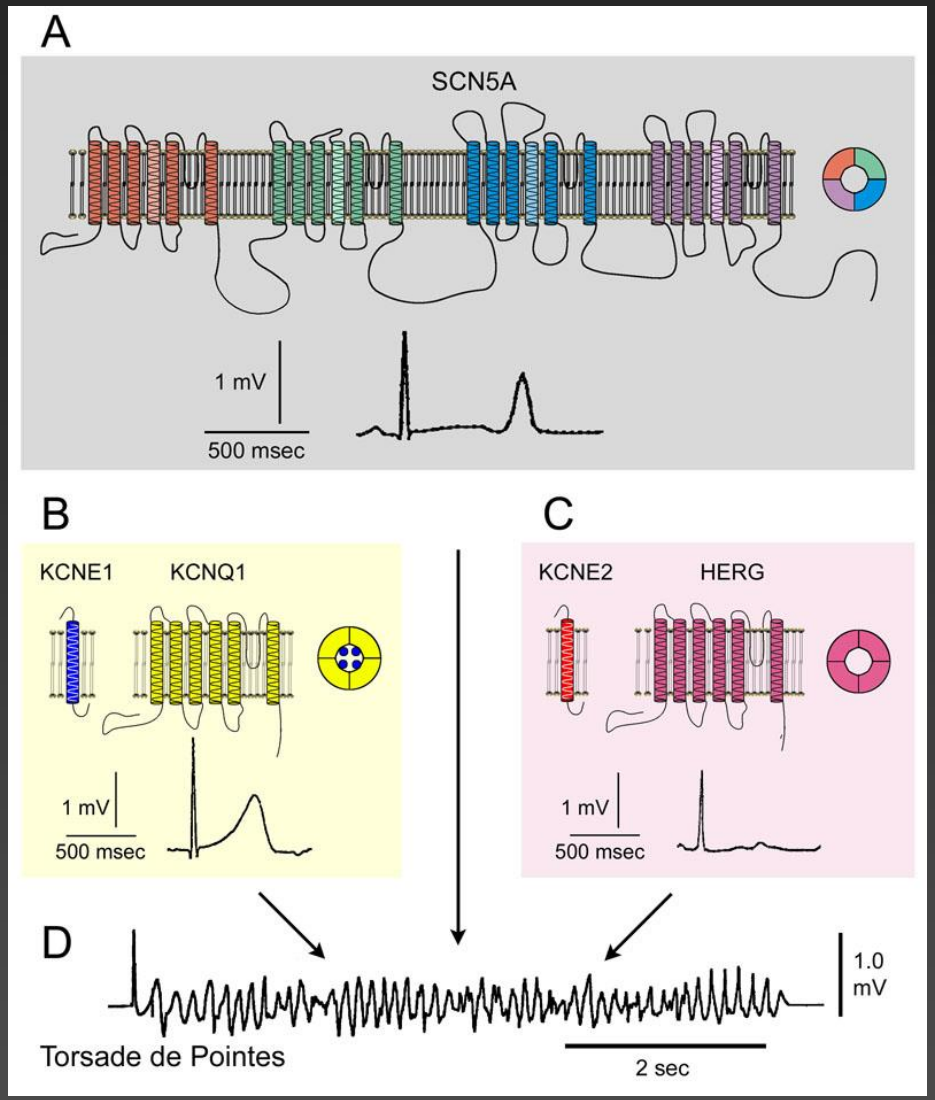


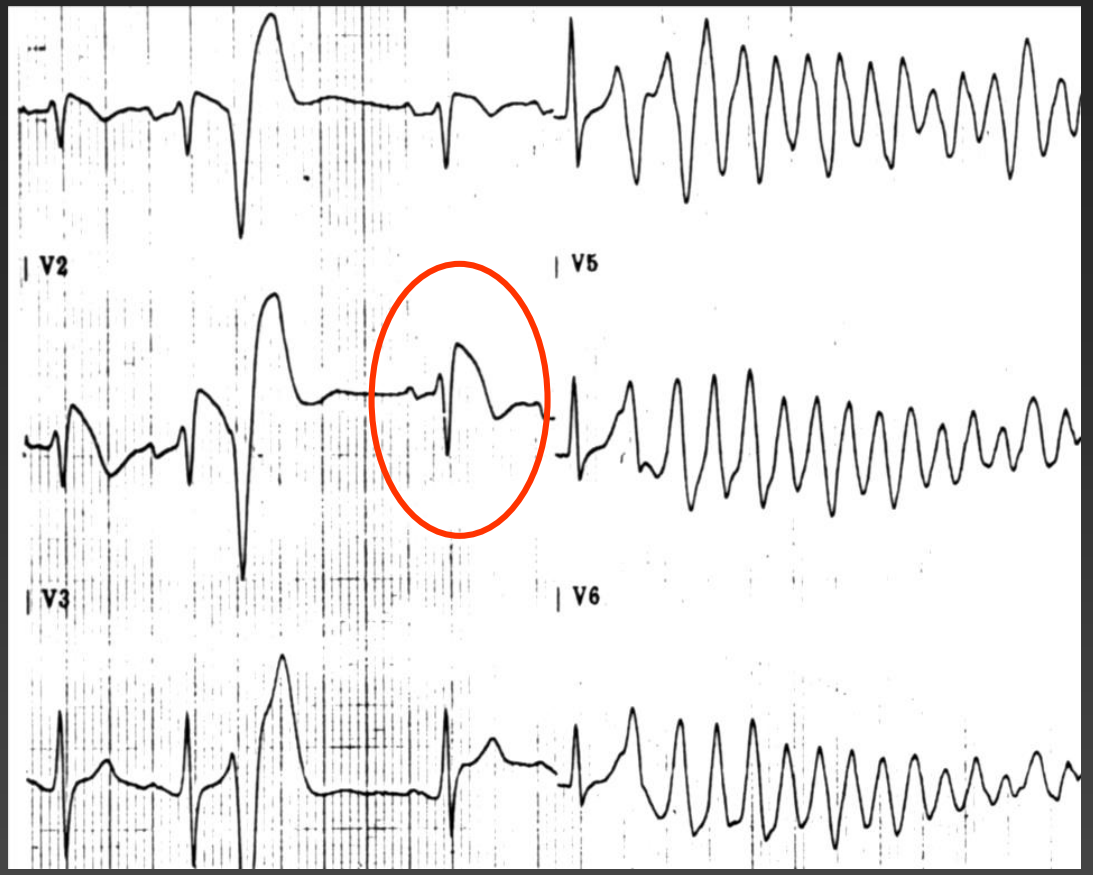


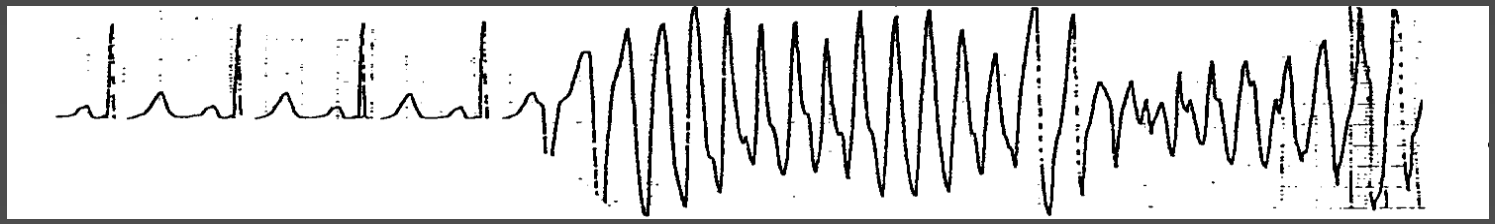
Epicardial



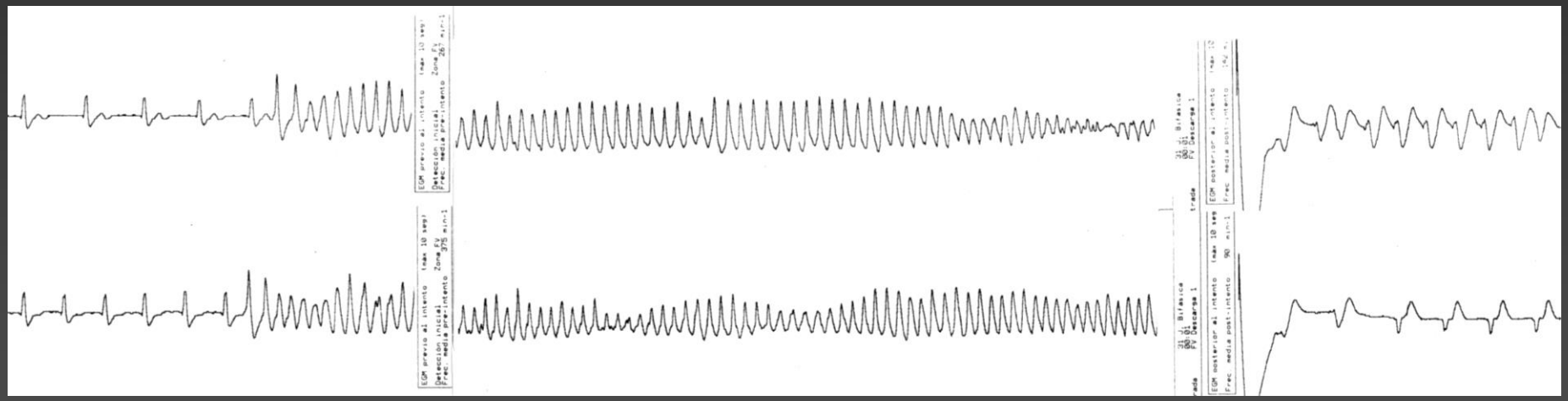
Haissaguerre M, et al. JACC 2009


A
SCN5A
1 mV
500 msec
B
KCNE1
KCNQ1
1 mV
500 msec
C
KCNE2
HERG
1 mV
500 msec
D
1.0 mV
2 sec
Torsade de Pointes

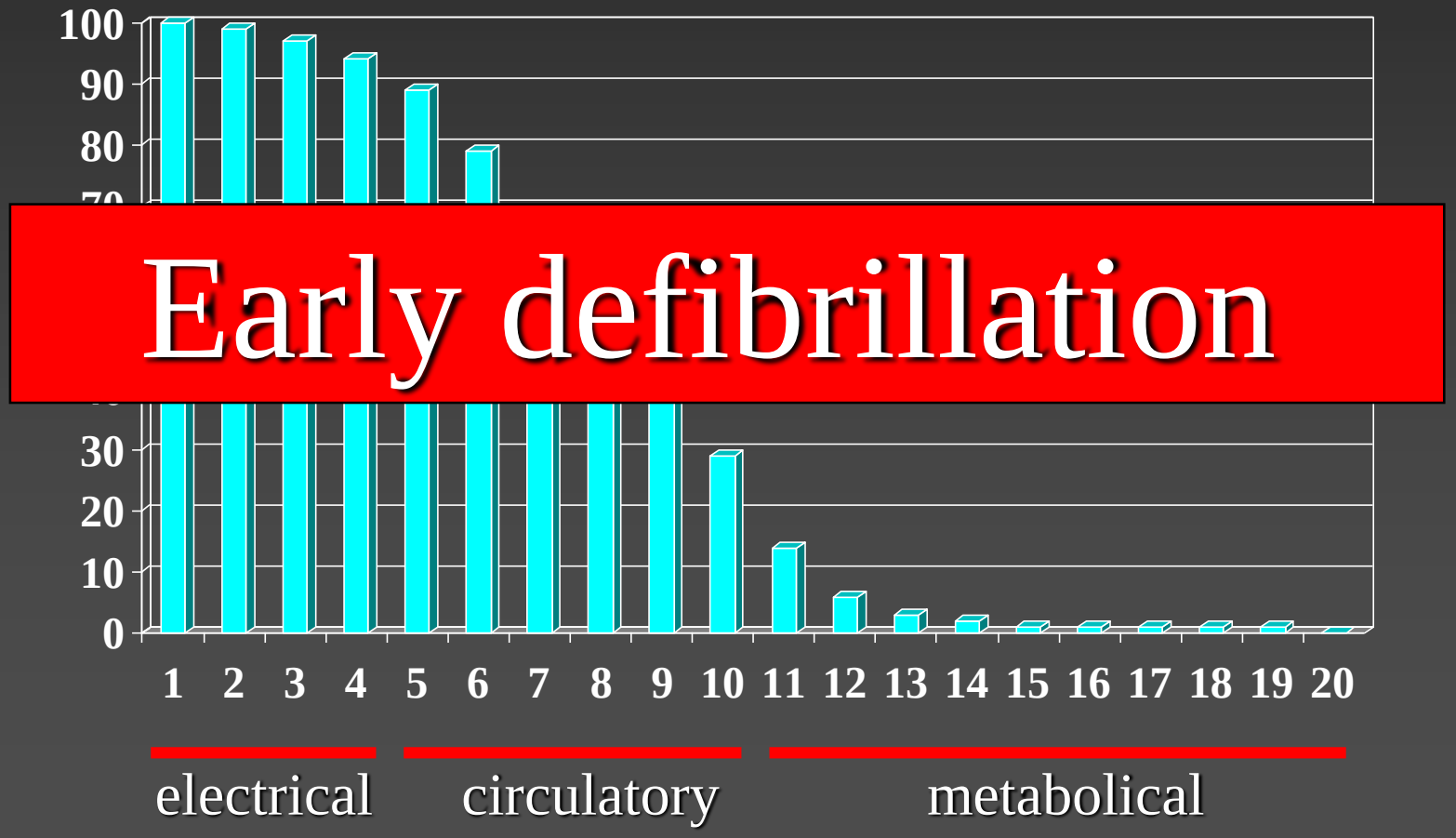

V2
V5
V3
V6



Defibrillator therapy



Survival depending on time to defibrillation



Public access defibrillation

Syncope and sudden death in patients without demonstrable structural heart disease

- Genetic studies have identified a new category of dieases responsible for syncope and sudden death in patients without demonstrable structural heart disease: channelopathies.
- ECG allows identification of most patients at risk