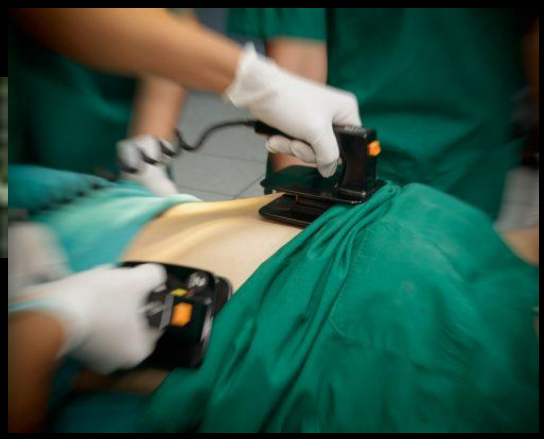
# CLUES TO IMPENDING ELECTRICAL DOOMS!



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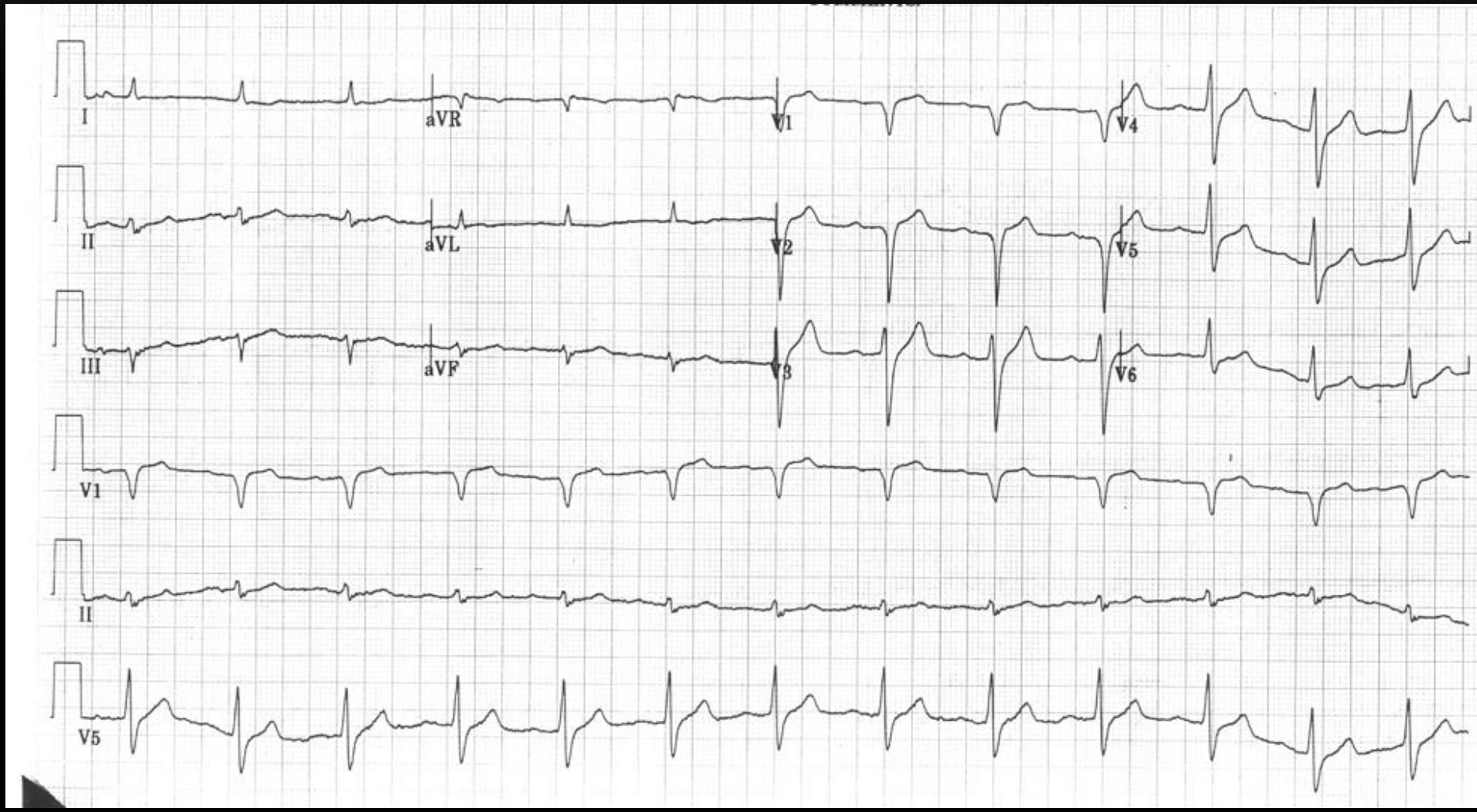
# MR. J.M.

- 65 year old rancher
- DM2 x 10 years, maintained on oral agents
- No diagnosed cardiac disease
- Unheralded syncope while a passenger in a car

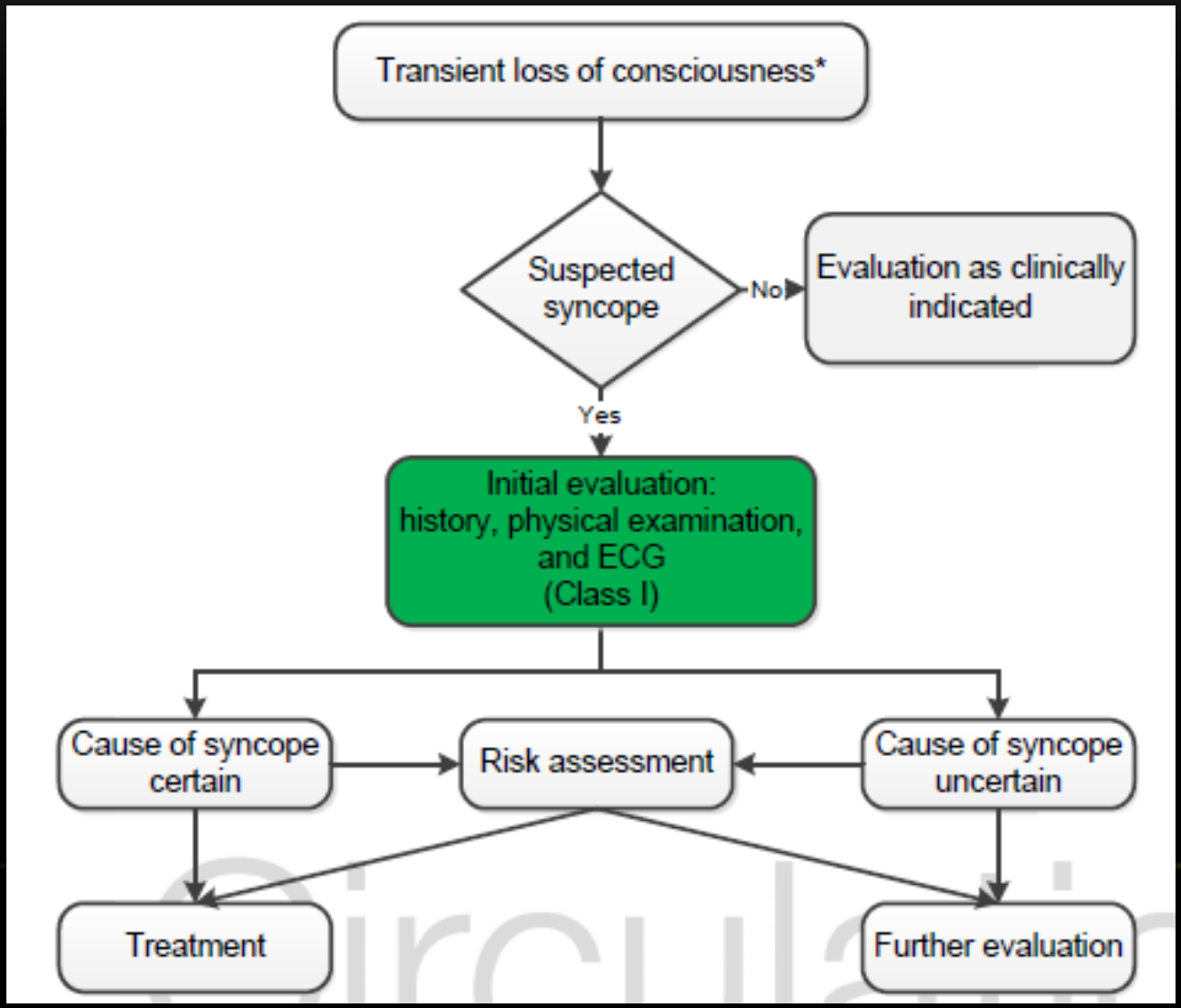
# Resting ECG 1



# Next Step?

1) Admission , monitoring, serial ECG's
2) Out patient assessment ( echo, cath,…)
3) Reassurance, refer in case of recurrent syncope
4) Refer to neurolgist?

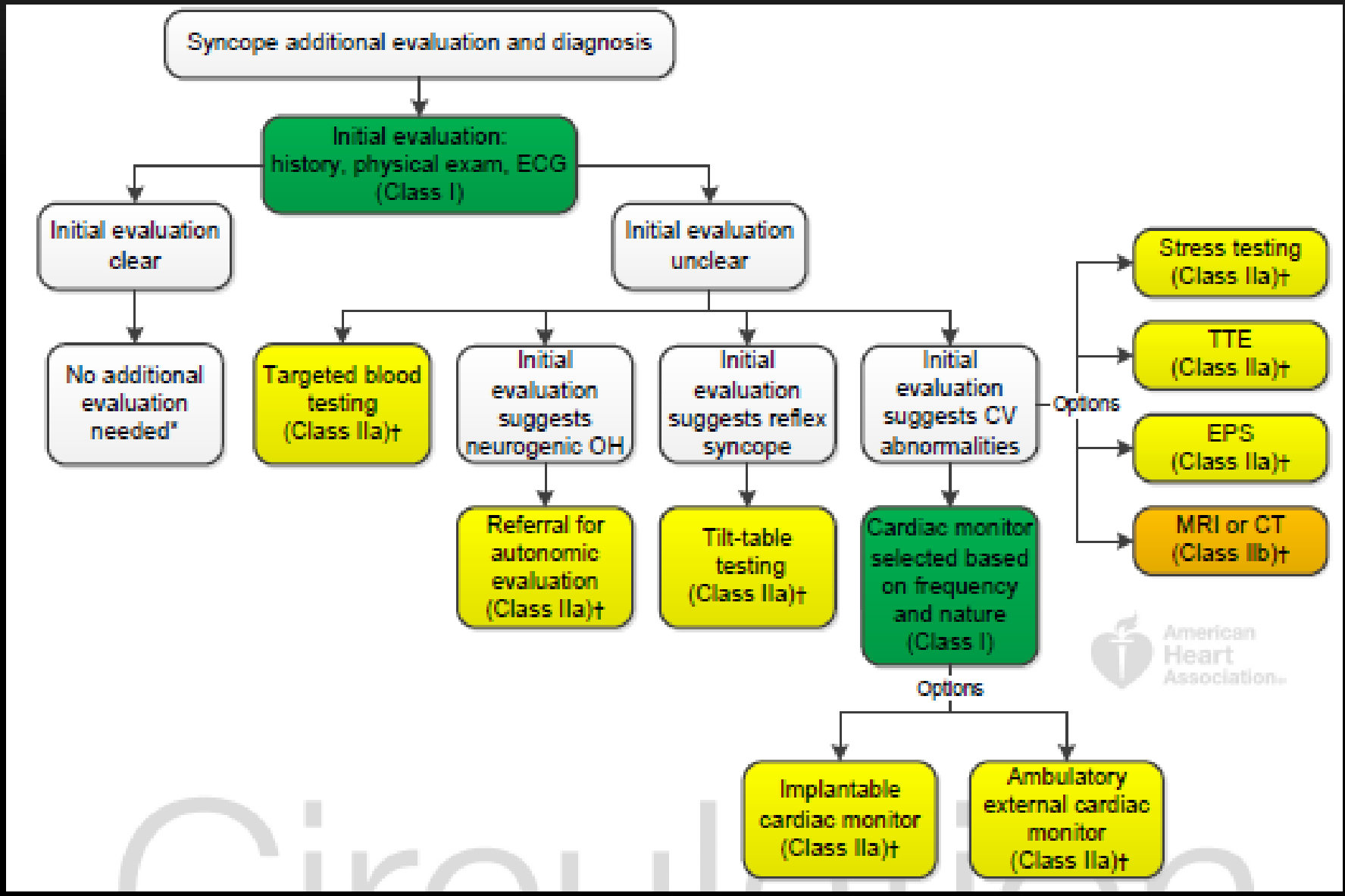
# Initial Evaluation



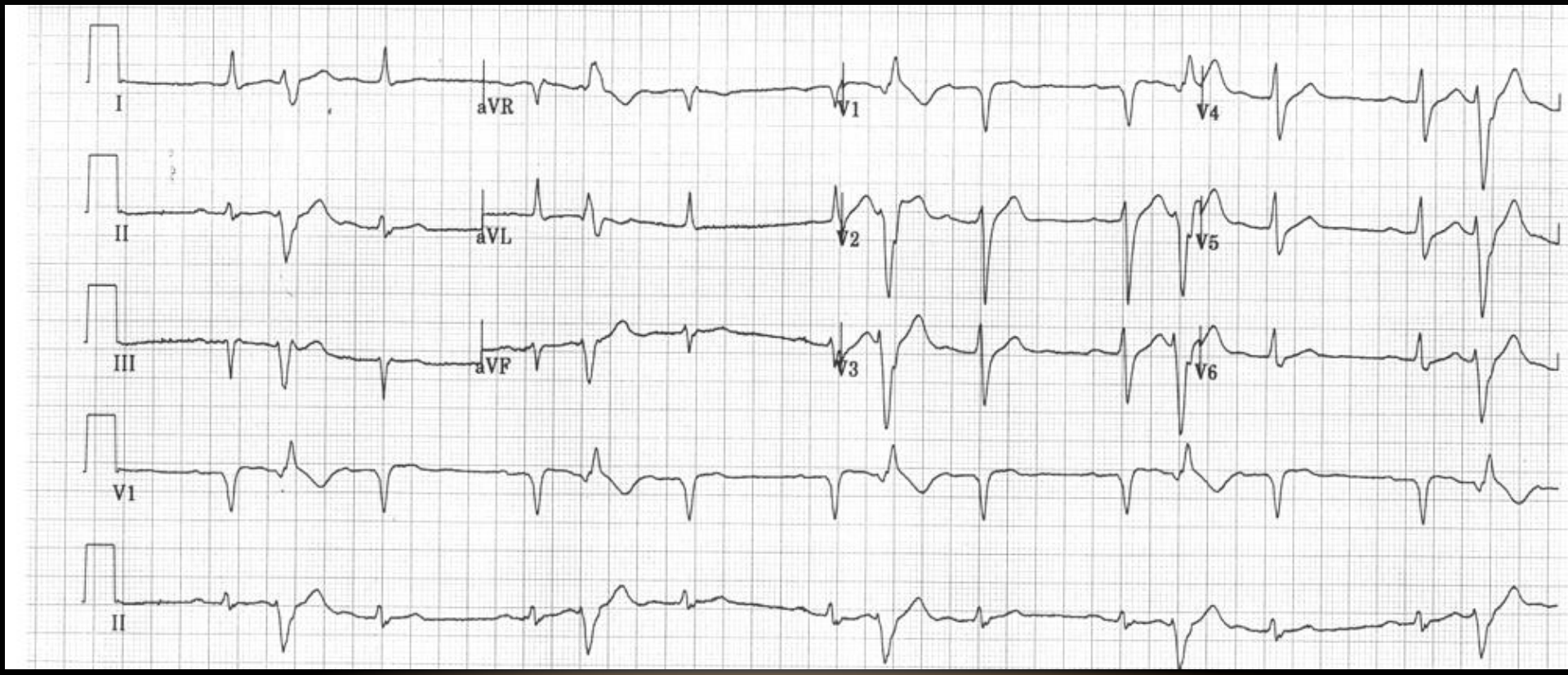
# Historic characteristics Associated with Cardiac & Non-Cardiac Syncope

| More Often Associated With Cardiac Causes of Syncope |
|---|
| • Older age (>60 y) |
| • Male sex |
| • Presence of known ischemic heart disease, structural heart disease, previous arrhythmias, or reduced ventricular function |
| • Brief prodrome, such as palpitations, or sudden loss of consciousness without prodrome |
| • Syncope during exertion |
| • Syncope in the supine position |
| • Low number of syncope episodes (1 or 2) |
| • Abnormal cardiac examination |
| • Family history of inheritable conditions or premature SCD (<50 y of age) |
| • Presence of known congenital heart disease |
| **More Often Associated With Noncardiac Causes of Syncope** |
| • Younger age |
| • No known cardiac disease |
| • Syncope only in the standing position |
| • Positional change from supine or sitting to standing |
| • Presence of prodrome: nausea, vomiting, feeling warmth |
| • Presence of specific triggers: dehydration, pain, distressful stimulus, medical environment |
| • Situational triggers: cough, laugh, micturition, defecation, deglutition |
| • Frequent recurrence and prolonged history of syncope with similar characteristics |

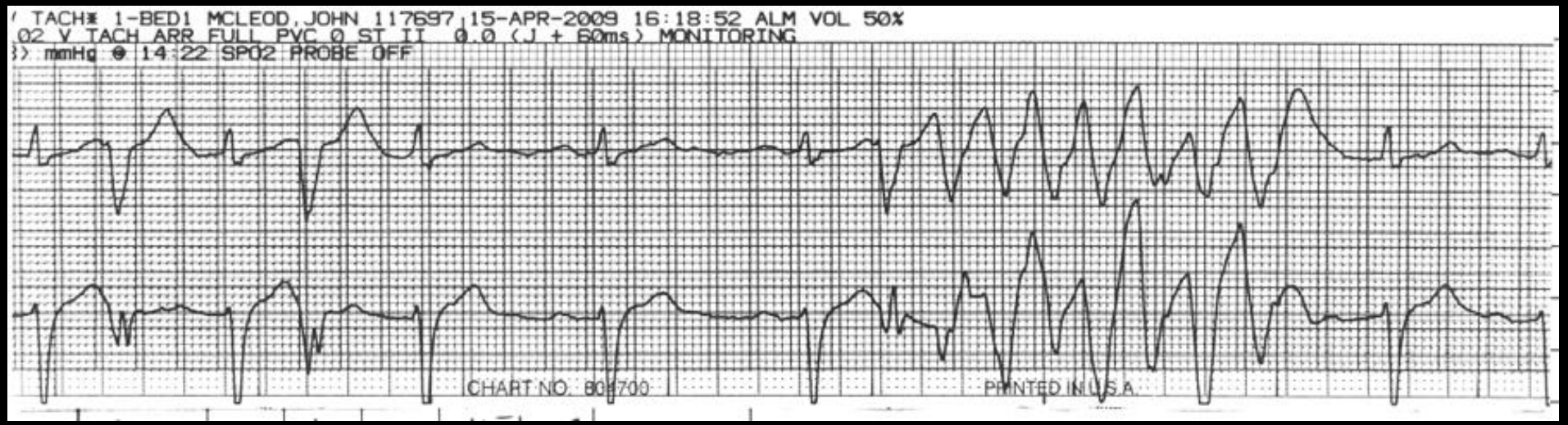
# Additional Evaluation and Diagnosis



# Resting ECG 2



# Rhythm strip



# ICD?

- 1. Yes!  Enough evidence according to NSVT and syncope
- 2. No!  Not indicated with this evidence.

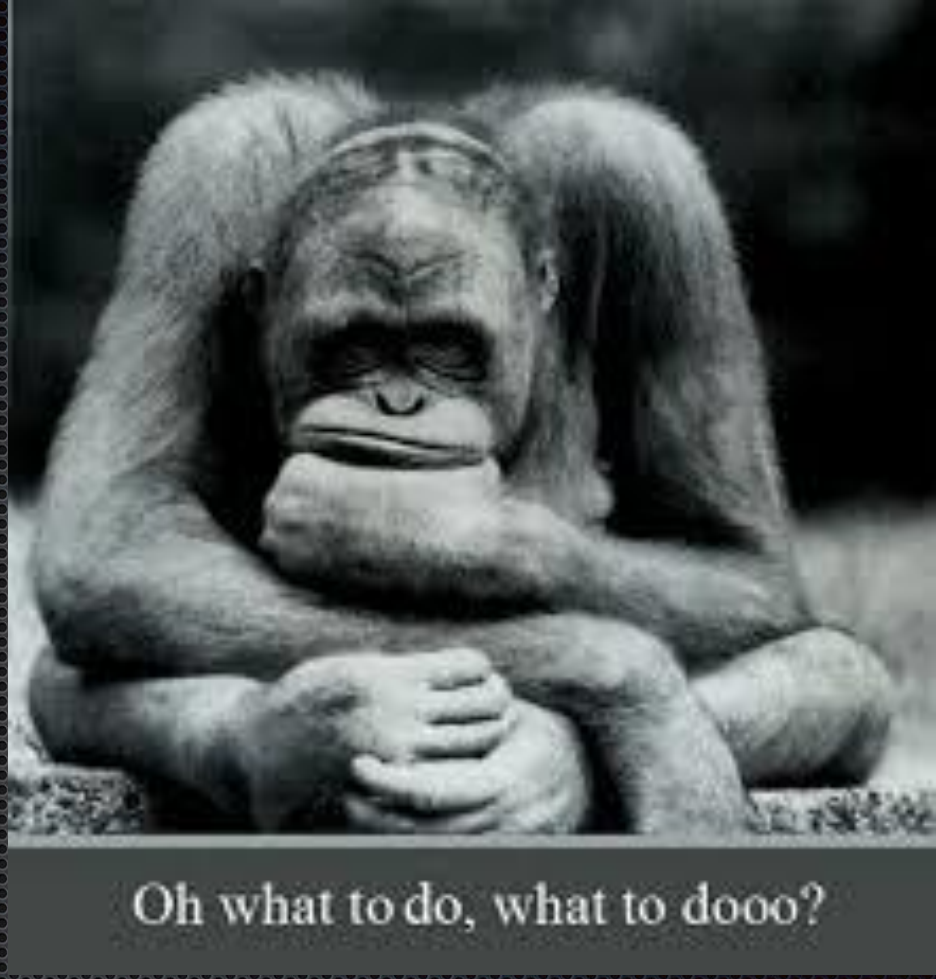
# Further info?

- No other symptoms recently
- 1 prior syncopal event - drinking
- ? other pre-syncopal events
- no family history of SCD

# Hospital course

- frequent PVCs, and short runs of polymorphic VT
  - mostly suppressed with BB
- cardiac cath: normal LV function, normal coronaries
- CMR:
  - no evidence of ARVC
  - mild global LV/RV dilation, EF 0.53
  - no scar/edema

- 1. Reassurance and discharge
- 2. Sent home with additional work up as outpatient
- 3. Not out of the woods yet?



# Some Facts

- High prevalence of Syncope

  40% will experience syncope at least once in a lifetime, 1-6% of hospital admissions

Most common cause: Neurally mediated syncope

- Cardiac cause can mean sudden death

*Syncope - The same as sudden death except that you wake up* *Engel GL. *Ann Intern Med*. 1978;89:403-412

# Some other Facts

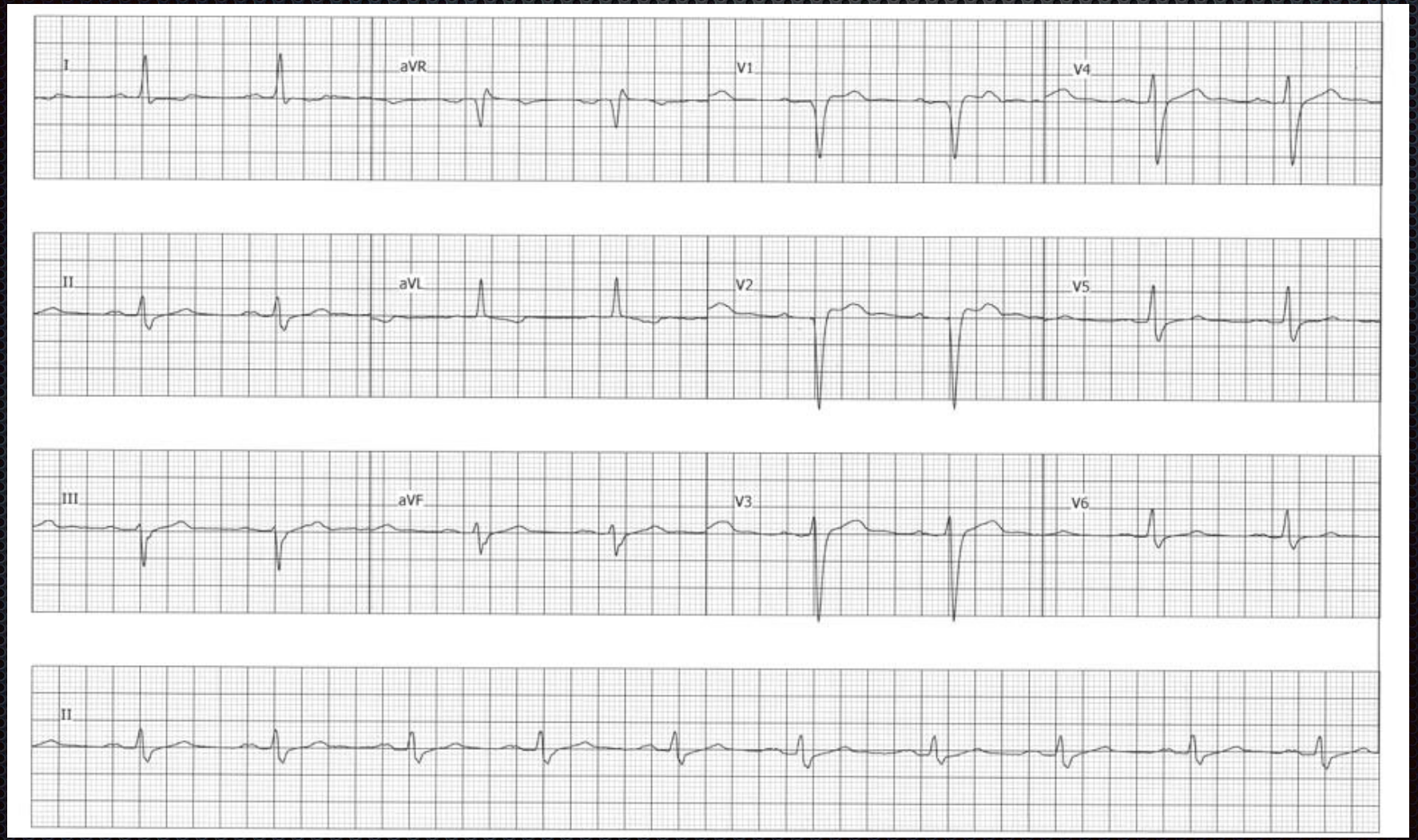
- No structural heart abnormalities are detectable in 5 to 8 percent of SCDs

- Long QT syndrome,

  Catecolaminergic Polymorphic VT (CPVT)

  J wave syndromes including Brugada syndrome

# Think again?



# And another fact

- Among affected patients with congenital arrhythmic disorders, resting ECG is frequently borderline, intermittently normal or frankly normal
- Genetic testing is often neither sensitive nor specific
- So, the diagnosis requires a high level of suspicion

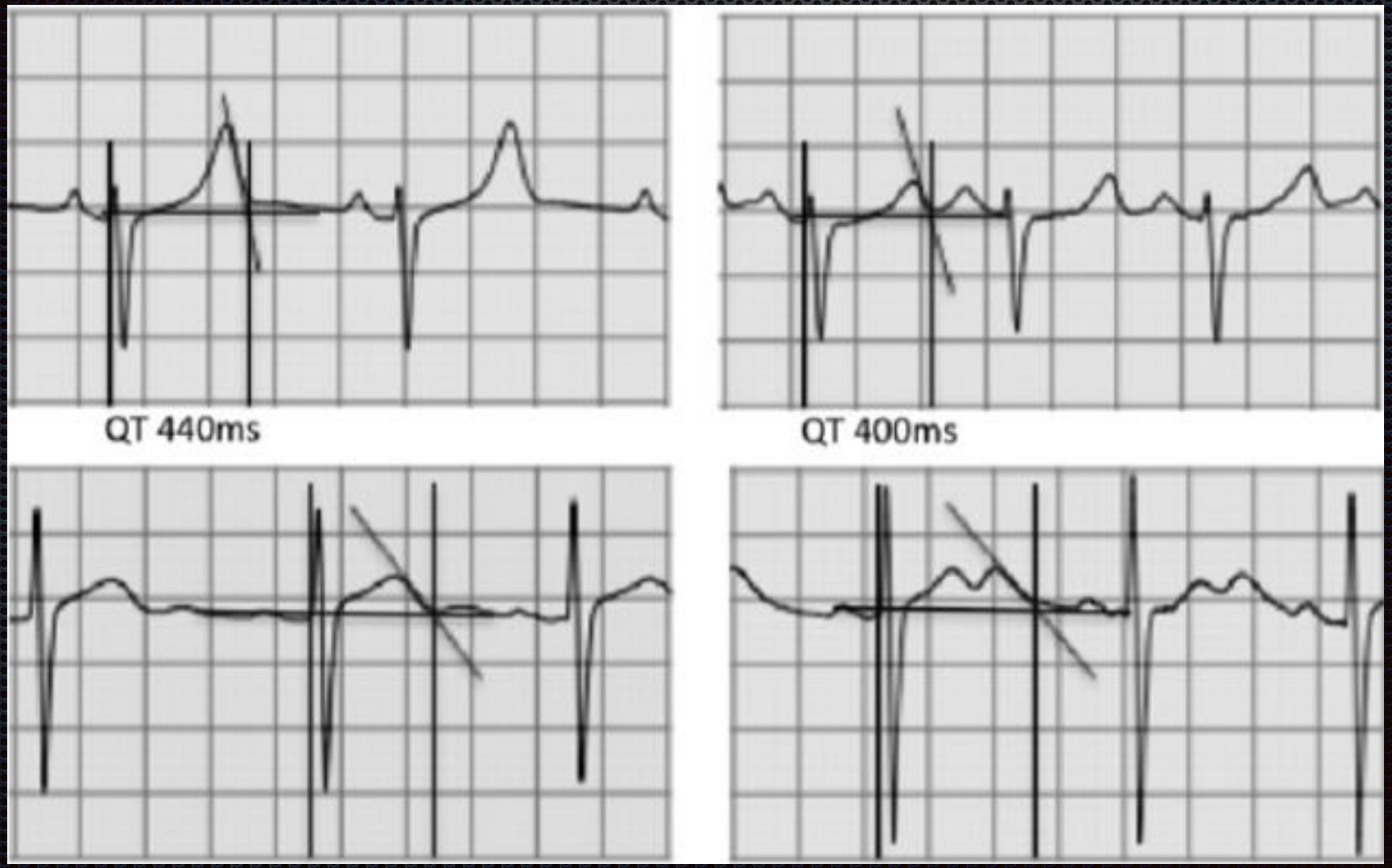
# Provocative testing

- Based on our knowledge about the pathophysiology of the disorder, we can provide the conditions to produce the classic ECG response

# Long QT syndrome

- 1. Don’t rely on ECG machine measurements!
- 2. Longest QT intervals are generally on precordial leads, standard leads are?
- QT/RR in normal people is never greater than?
- Bazett formula is still the most widely used to measure corrected QT. May I rely on machine?
- 2. U wave should not be included in the measurement
- 3. In atrial fibrillation or sinus arrhythmia?

# Maximum slope technique



# Dilemma

- Up to 50% of patients with LQTS display a non-diagnostic QTc (<460 mse)
- Sensitivity of genetic testing is 75%

# Take advantage of ion channels

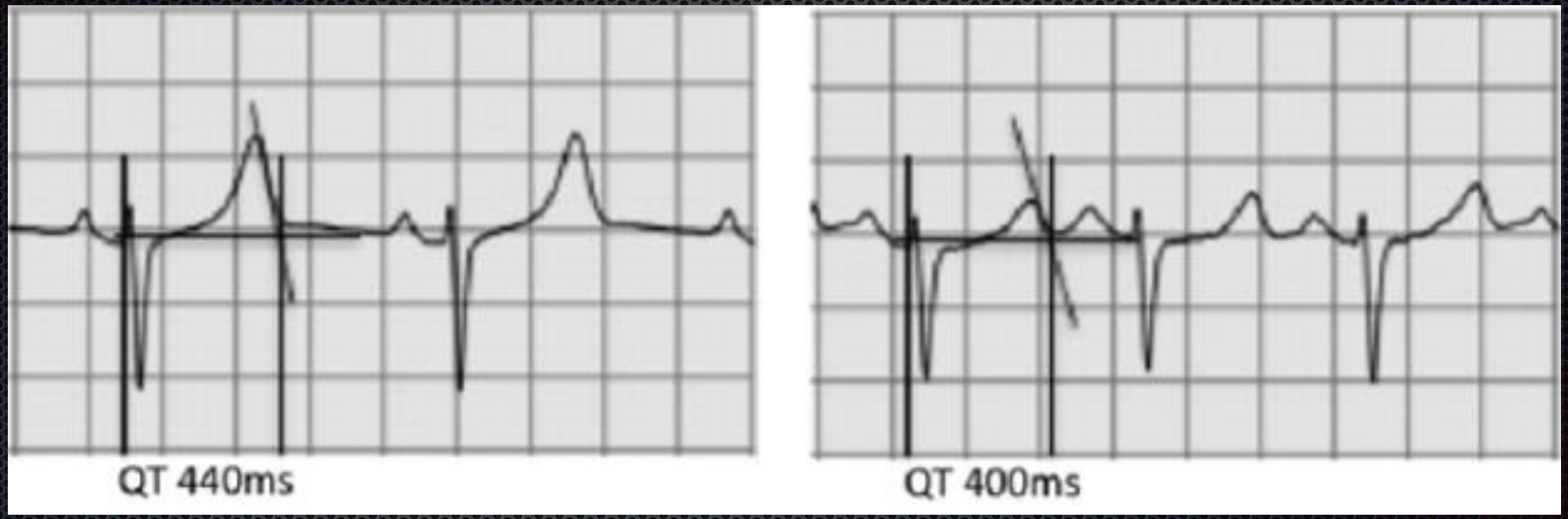
- In normal individuals, the K channels improve conduction with cathecolamines resulting in shortened QT and QTc
- In LQTS types I and II (but not LQT type 3), the impaired K channel does not respond properly to catecholamines

What is the implication?

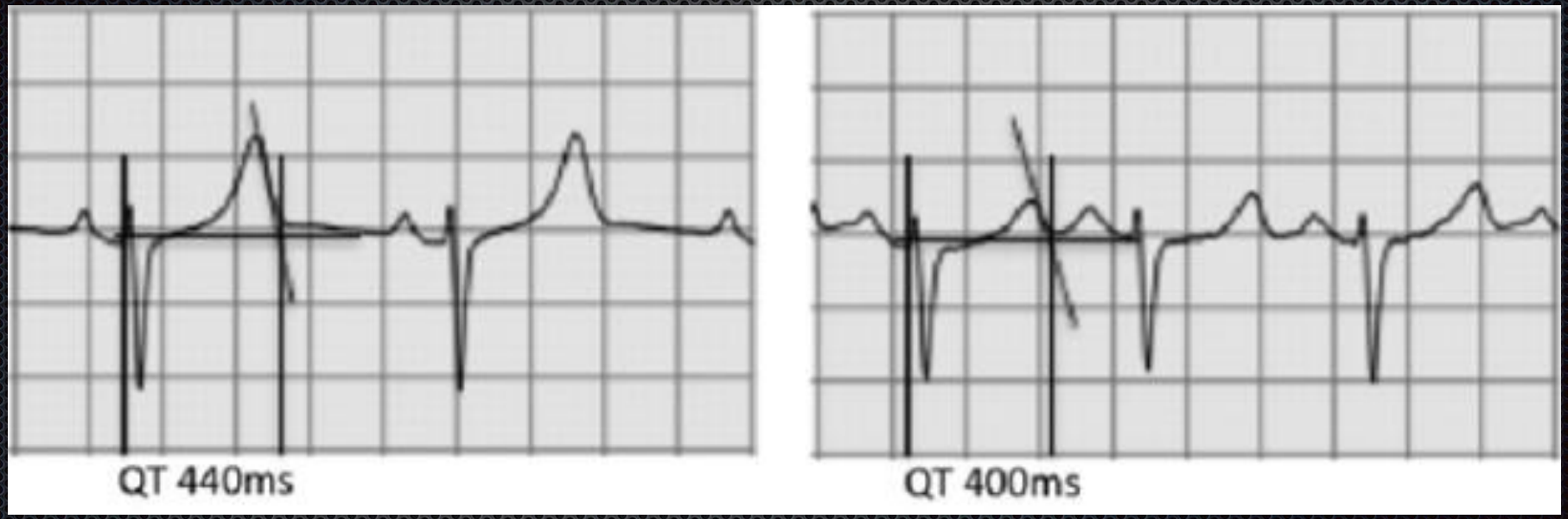
# Paradoxical QT prolongation

- 1. With brisk standing
- 2. During exercise
- 3. Epinephrine infusion provocation test

# Before and after Epi



# Before and after Epi

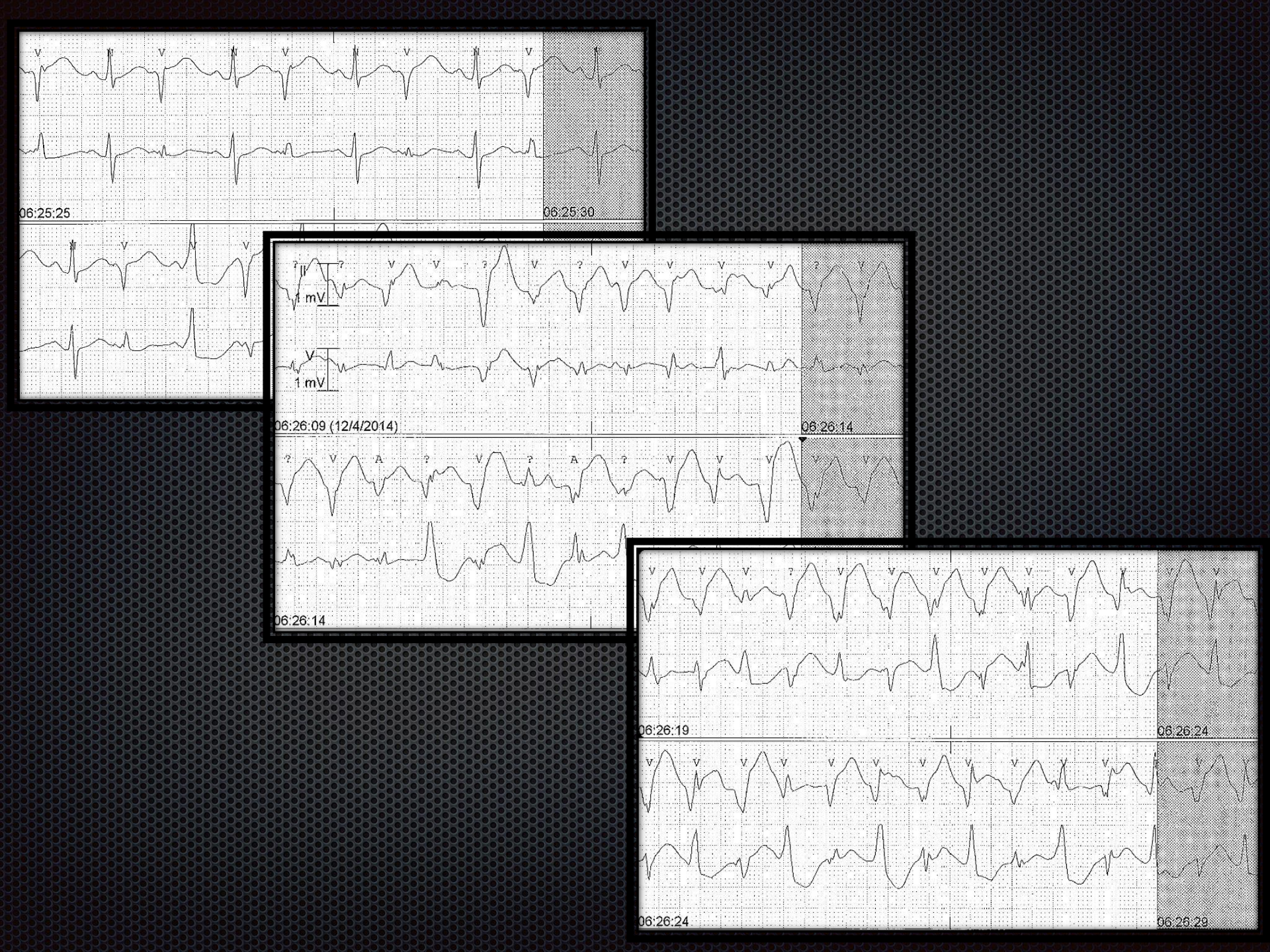



- QTc: 475 msec
- QTc: 550 msec

- Mr. J. M. had a normal QT response to Epi challenge test

# Catecholaminergic Polymorphic VT (CPVT)

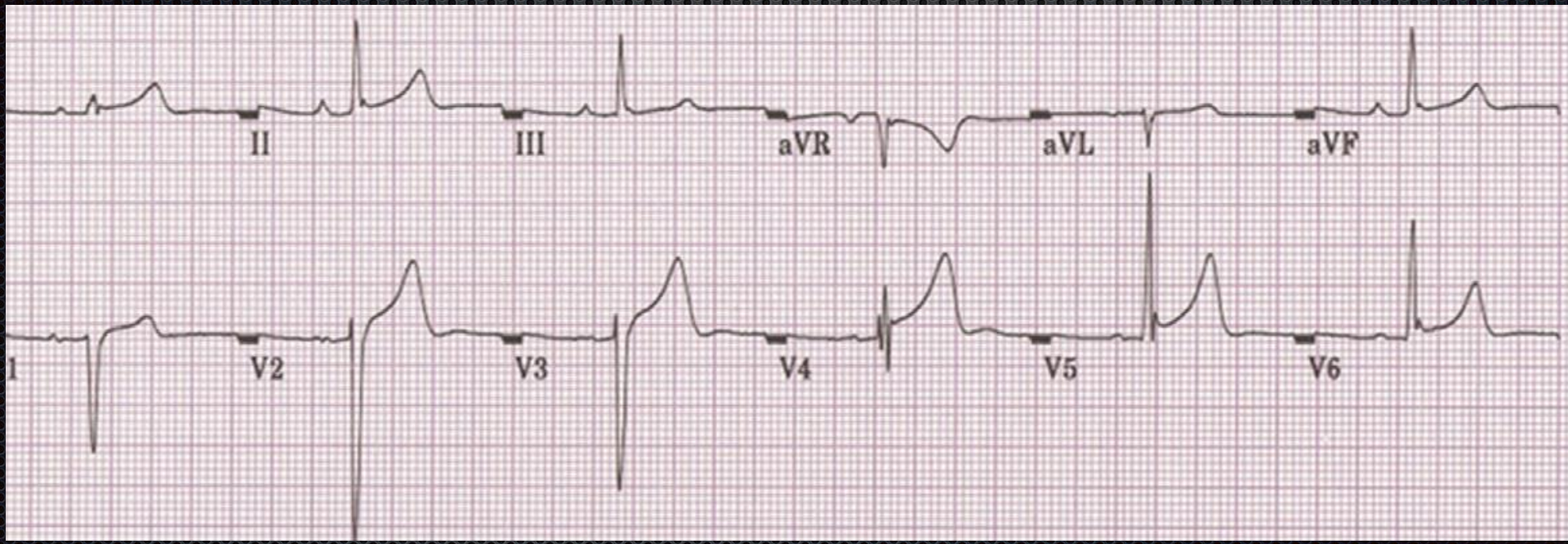
- Normal resting ECG
- Genetic malfunction of calcium channel resulting in bidirectional VT with catecholamine release particularly with exercise


06:25:25
06:25:30
1 mV
1 mV
06:26:09 (12/4/2014)
06:26:14
06:26:14
06:26:19
06:26:24
06:26:24
06:26:29

- Mr. J.M. did an exercise test with no VT.

Anything else should be observed during test?

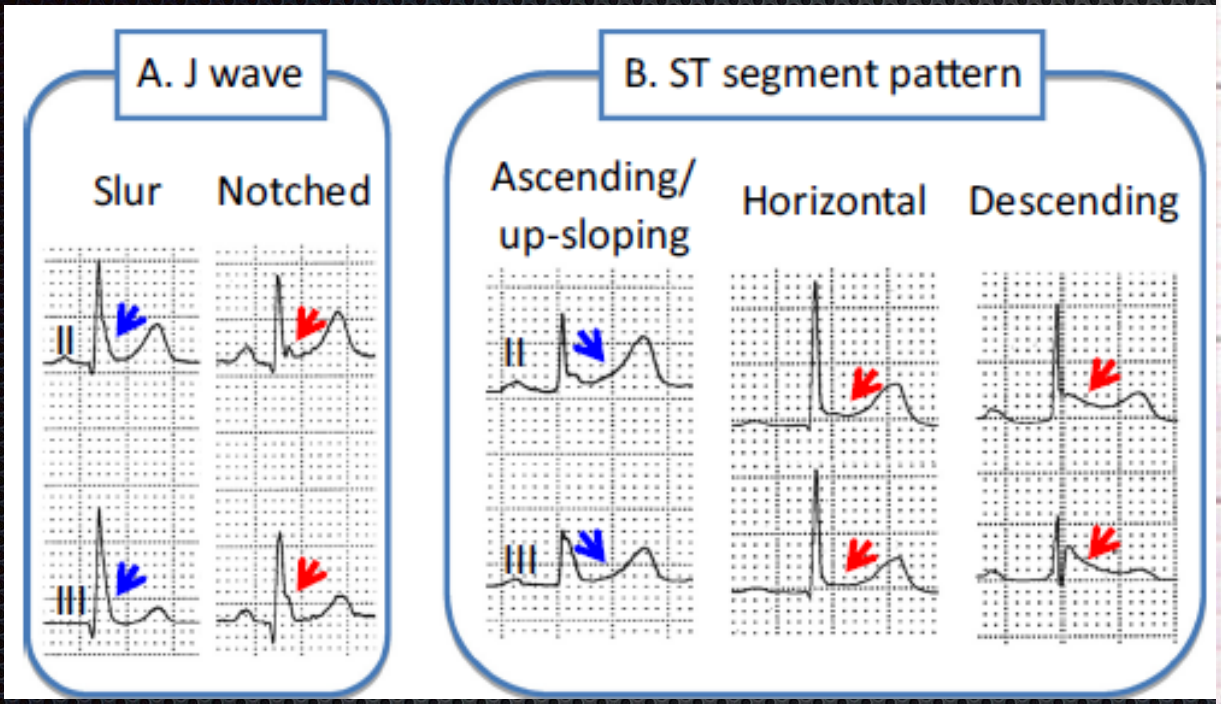
# Is this ECG normal?

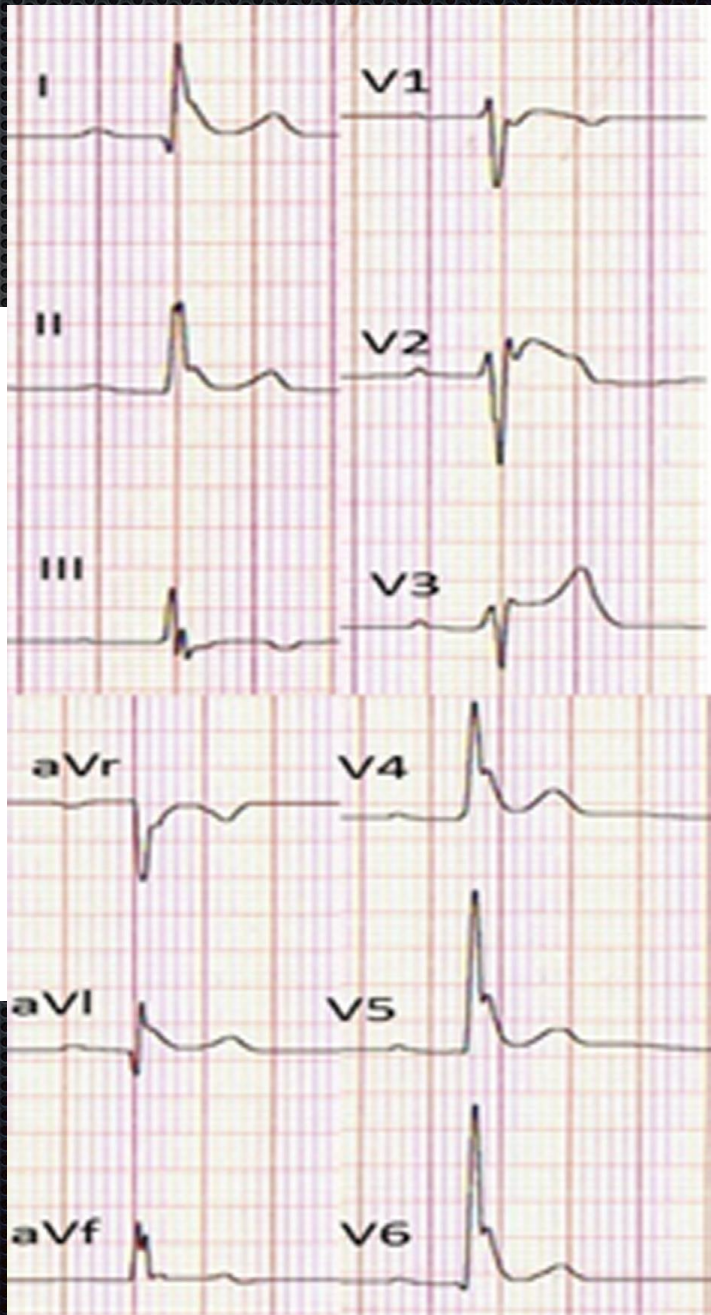


# Early Repoplariztion Syndrome

- Previously known to be benign
- Commonly seen in young athletes
- Needs attention in case of syncope and particularly when there is family history of SCD
- Highest risk with J point elevation in both precordial and inferior leads

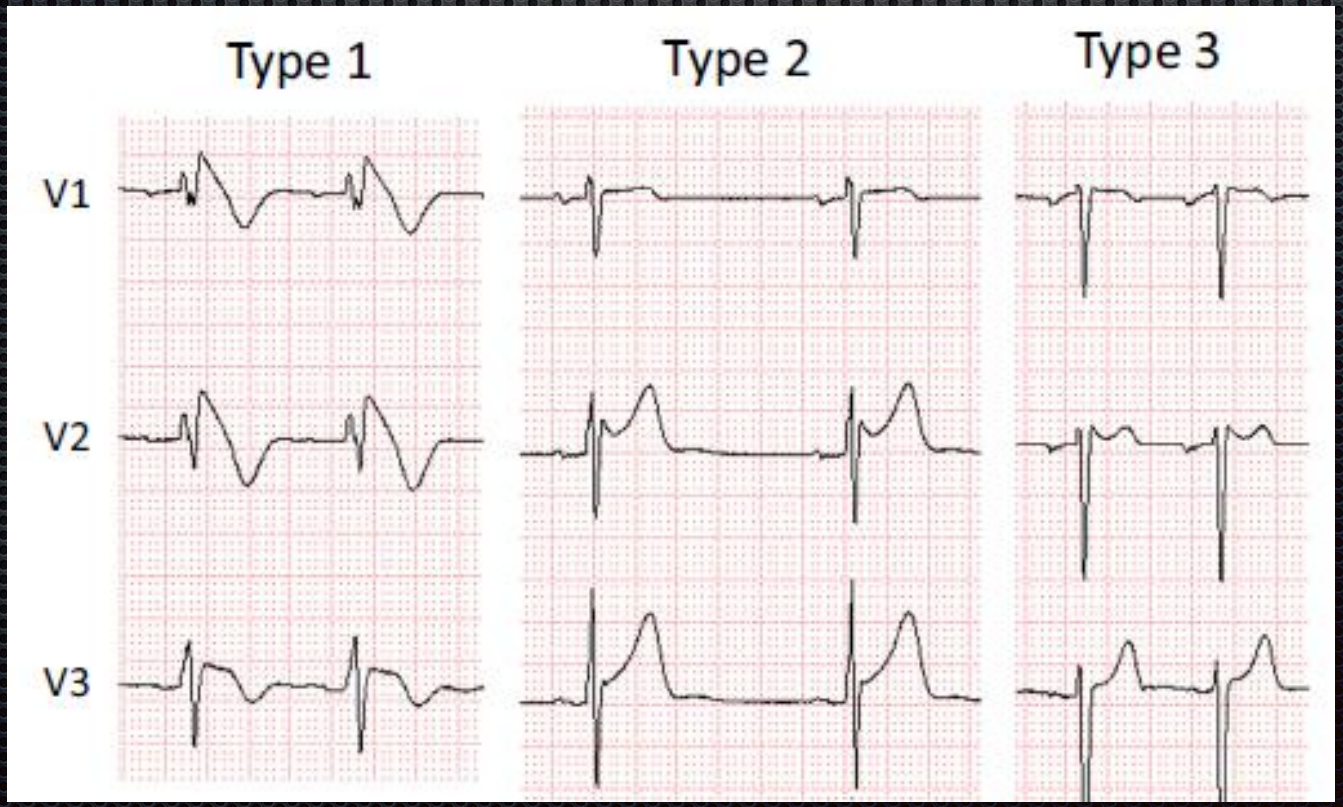
# Don’t ignore unusual JT segment



# Brugada Syndrome

- Autosomal dominant heterogeneity of the diastolic activation in epicardium vs endocardium of the right ventricle



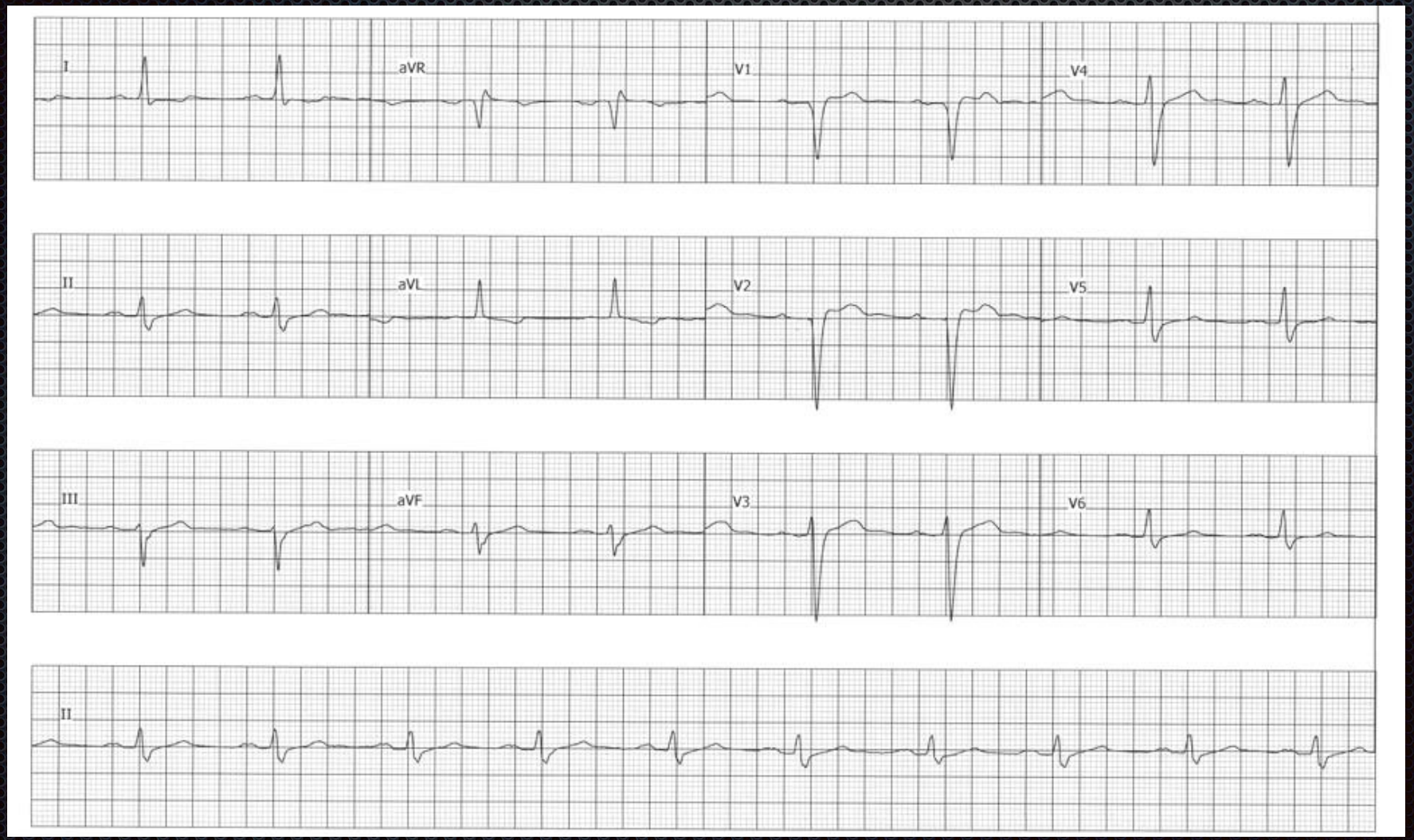
# Brugada Syndrome

- The prevalence of fluctuations between diagnostic and non-diagnostic ECG is high.
- Standard genetic testing is not universally feasible and is not sensitive

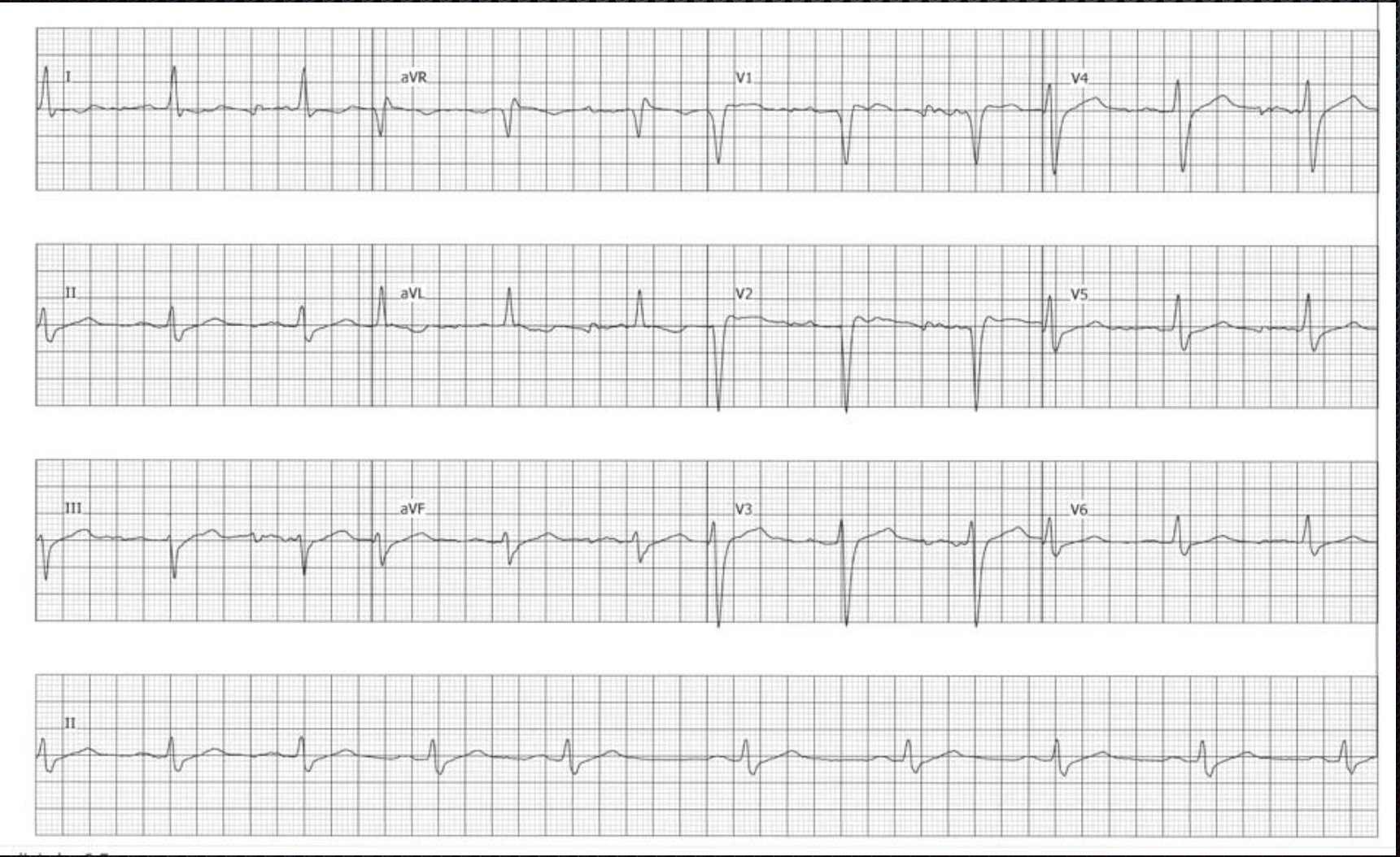
# Provocative test

- Should block the impaired Na channel
- Procainamide is usually utilizes to uncover the characteristic ECG pattern

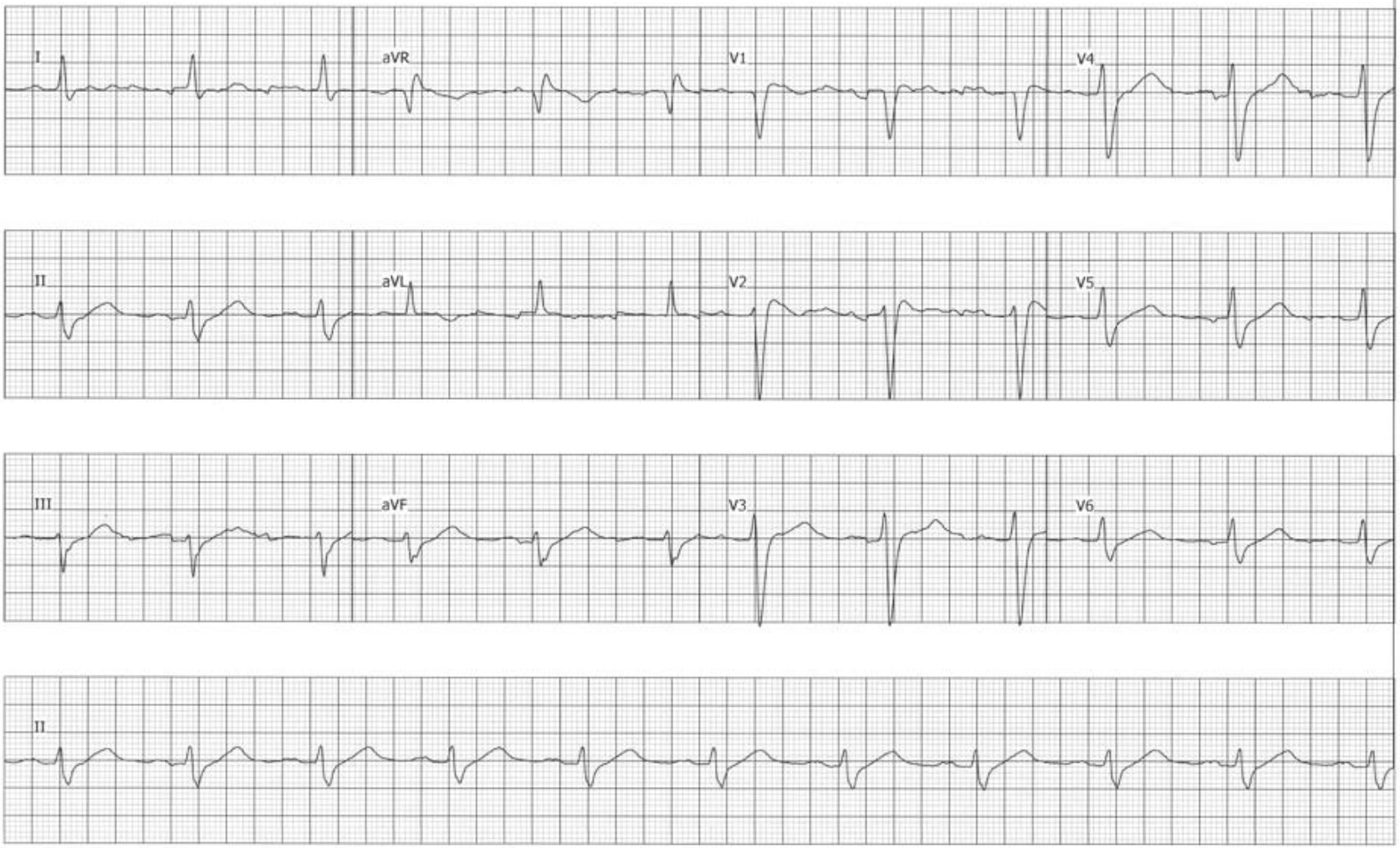
# Procainamide - baseline



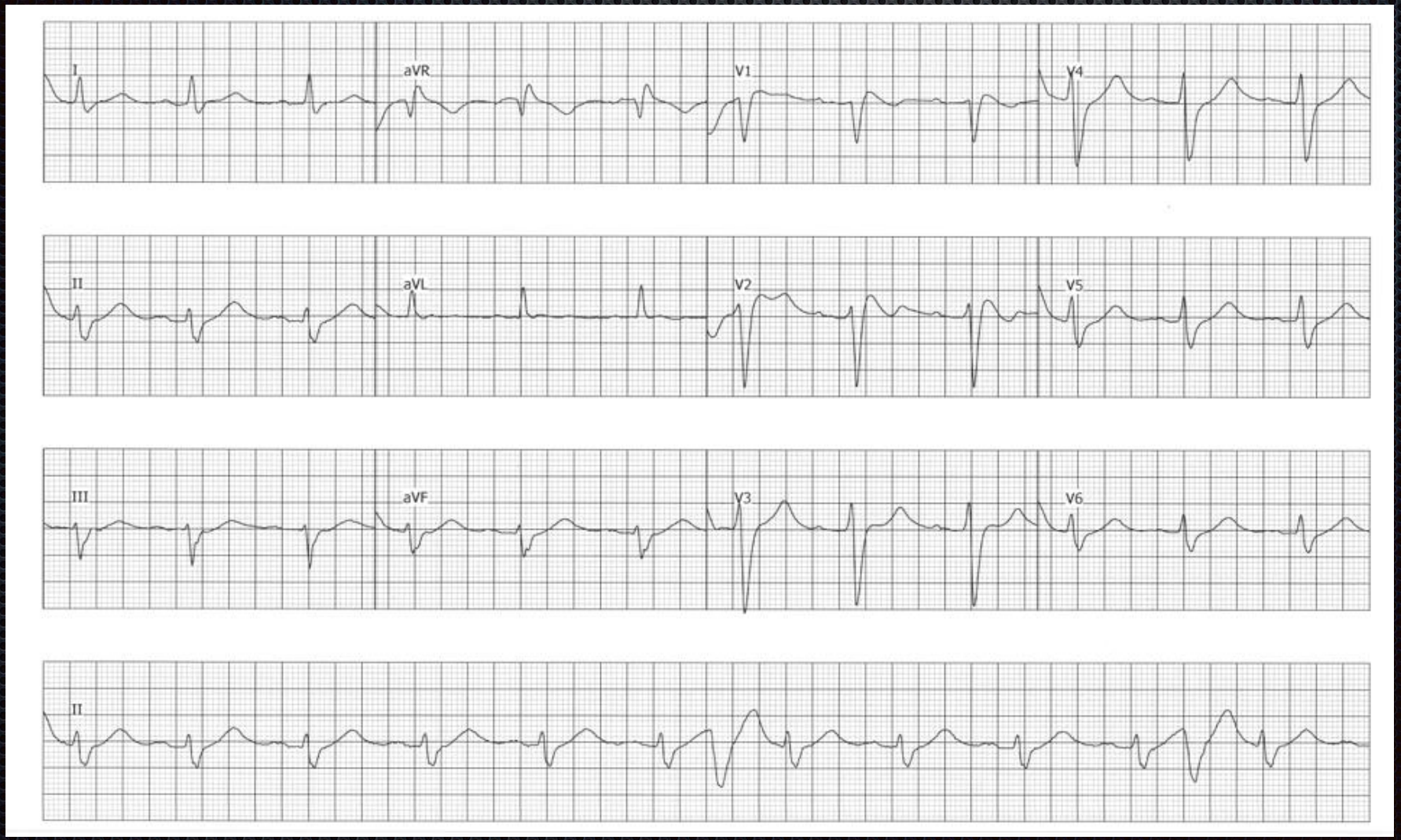
# Procainamide - 5 minutes



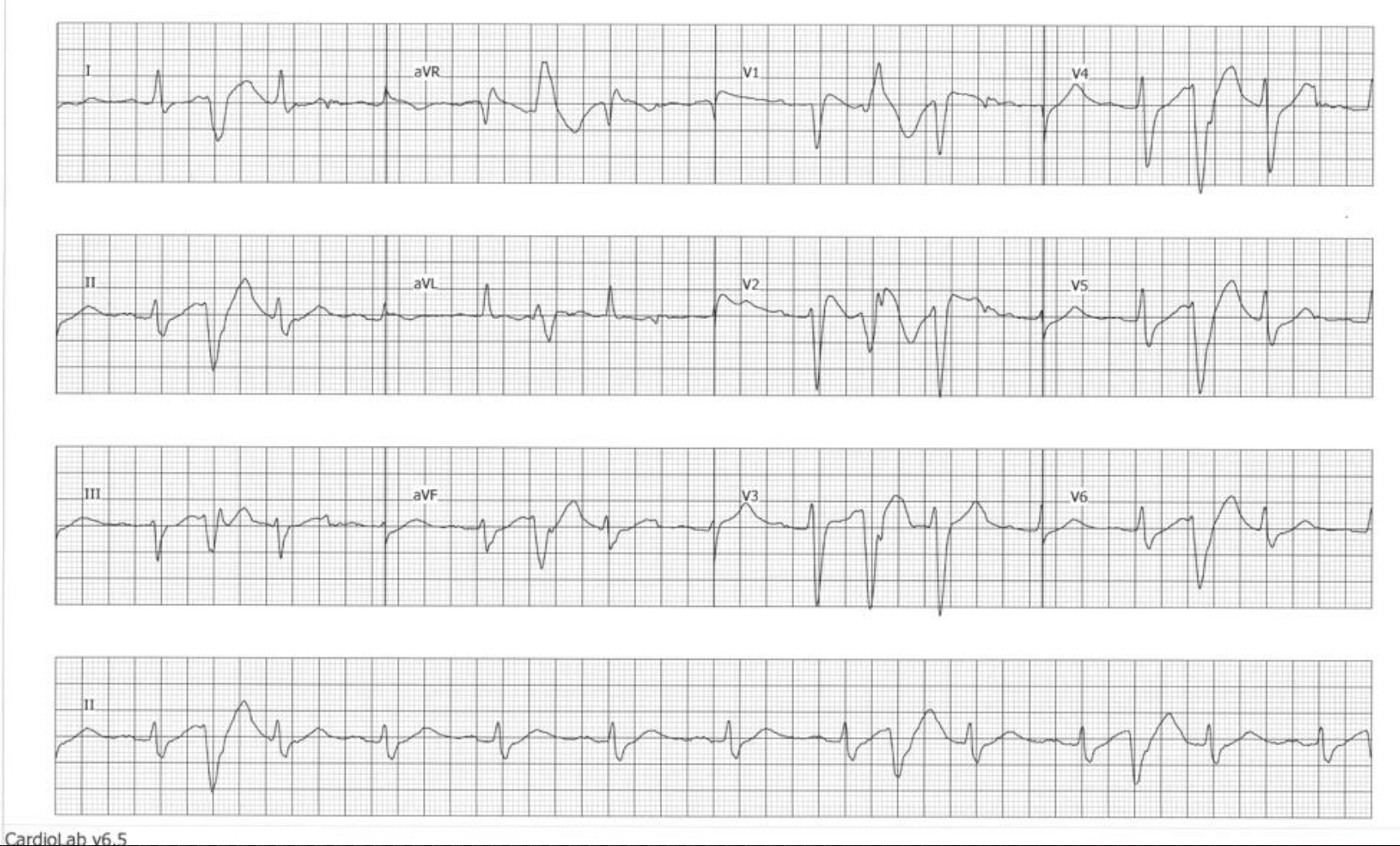
# Procainamide - 8 minutes



# Procainamide - 10 minutes



# Procainamide washout



Mr. J.M. received an ICD
Anything further?

# TAKE HOME

- 1. A good history taking is a cruicial part of assessment of a patient with syncope; *Look for red flags*
- 2. Arrhythmogenic disorders with normal heart structure are eerily missed even by cardiologists. Take an expert opinion if there is alittle doubt!
- A normal ECG may fool a physician. The next syncopal episode may be the doom!