Arrest 를 유발하는 심전도 판독

삼성서울병원
김주연

HRS/EHRA/APHRS Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes

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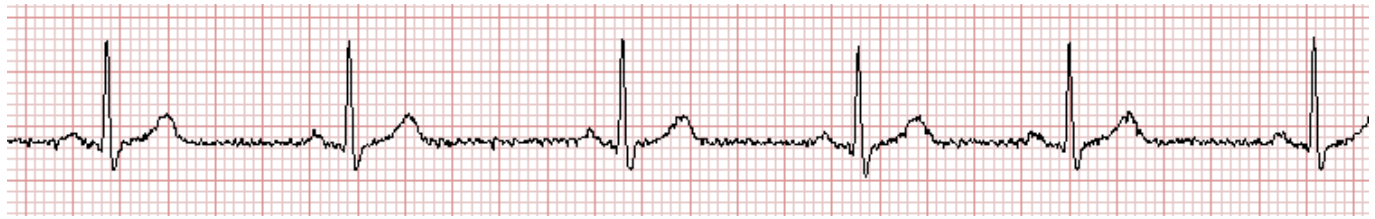
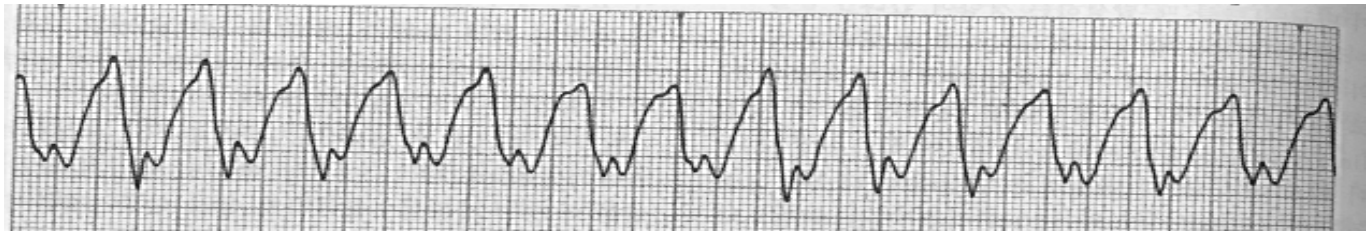
돌연사의 특징

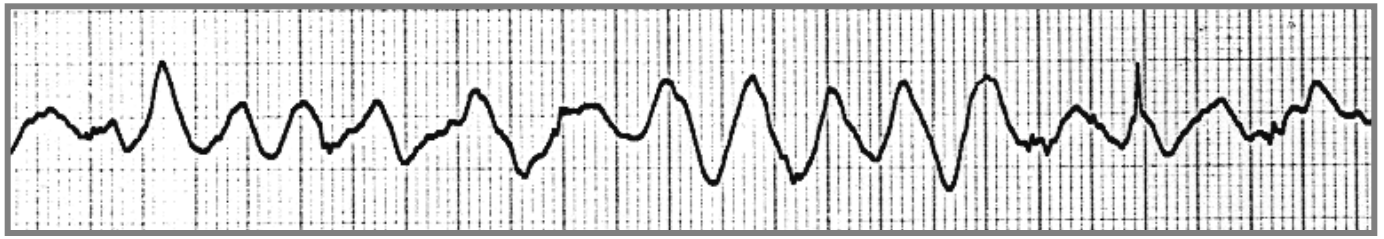
- 남자에게 흔함
- 평소에는 대개 무증상
- 가족력이 있는 경우가 많음
- 격렬하거나 경쟁적인 스포츠에서 보다 흔함

왜 갑자기 죽는가?

- 비후성 심근증, 브루가다 증후군, 심근경색, 모두 결국에는 심실세동 혹은 심실빈맥이 발생하여 사망

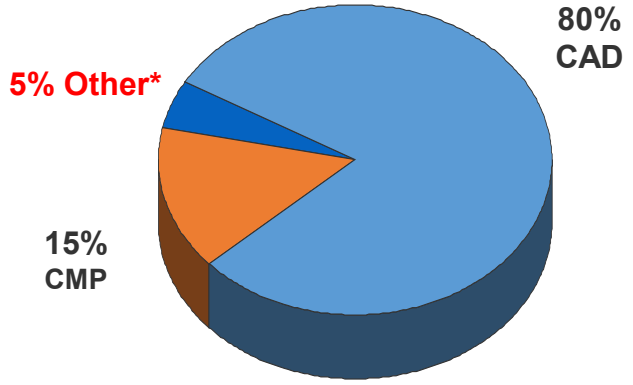


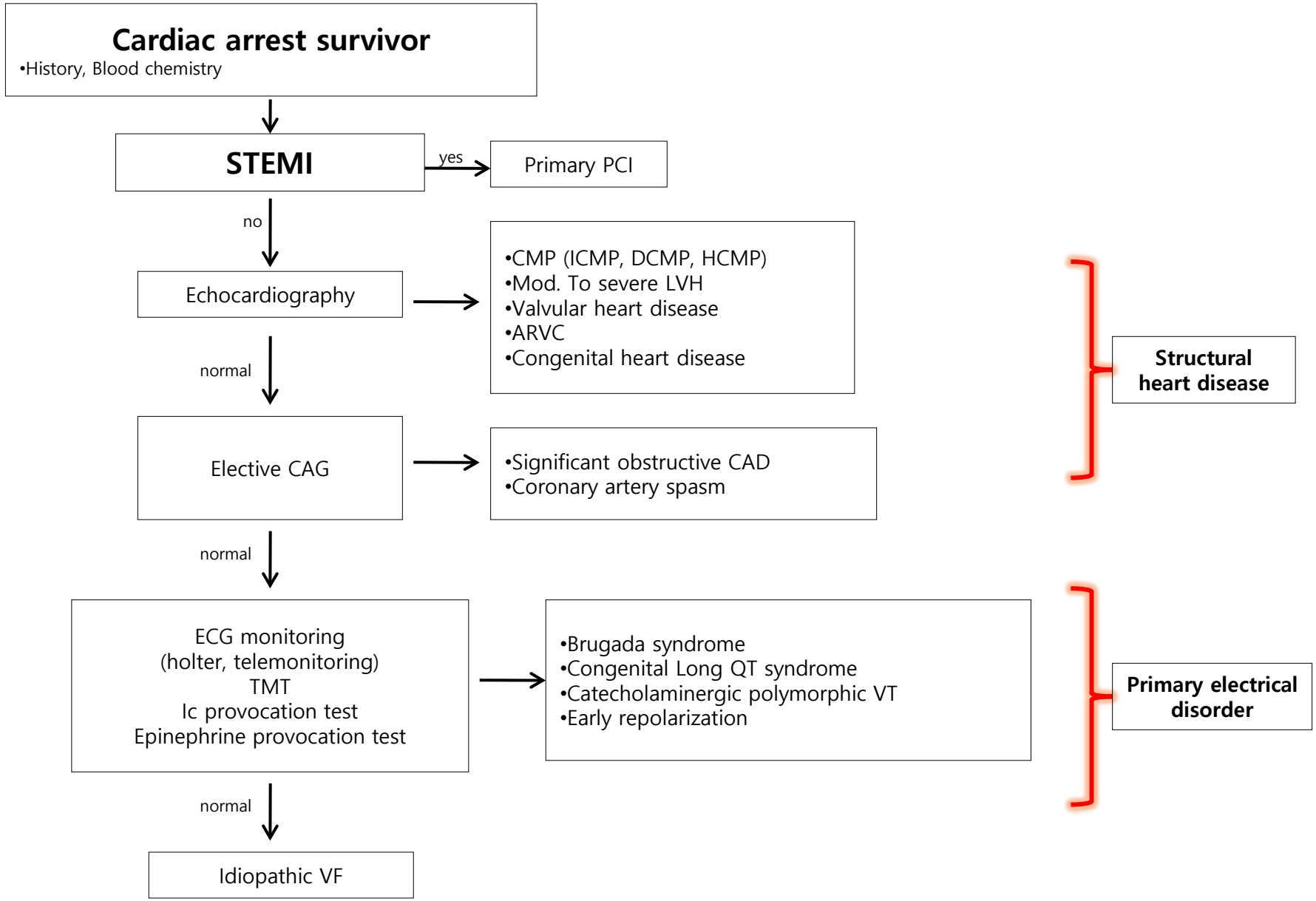




Aborted sudden cardiac death

- Structural heart disease : DCMP, ICMP, HCMP, ARVC
- Inherited primary arrhythmia syndrome
  - LQTS
  - BrS
  - Catecholaminergic polymorphic VT
  - Short QT syndrome
  - Early repolarization
  - Idiopathic VF




Cardiac arrest survivor
•History, Blood chemistry
STEMI
yes
Primary PCI
no
Echocardiography
•CMP (ICMP, DCMP, HCMP)
•Mod. To severe LVH
•Valvular heart disease
•ARVC
•Congenital heart disease
normal
Elective CAG
•Significant obstructive CAD
•Coronary artery spasm
normal
ECG monitoring
(holter, telemonitoring)
TMT
Ic provocation test
Epinephrine provocation test
•Brugada syndrome
•Congenital Long QT syndrome
•Catecholaminergic polymorphic VT
•Early repolarization
normal
Idiopathic VF
Structural heart disease
Primary electrical disorder

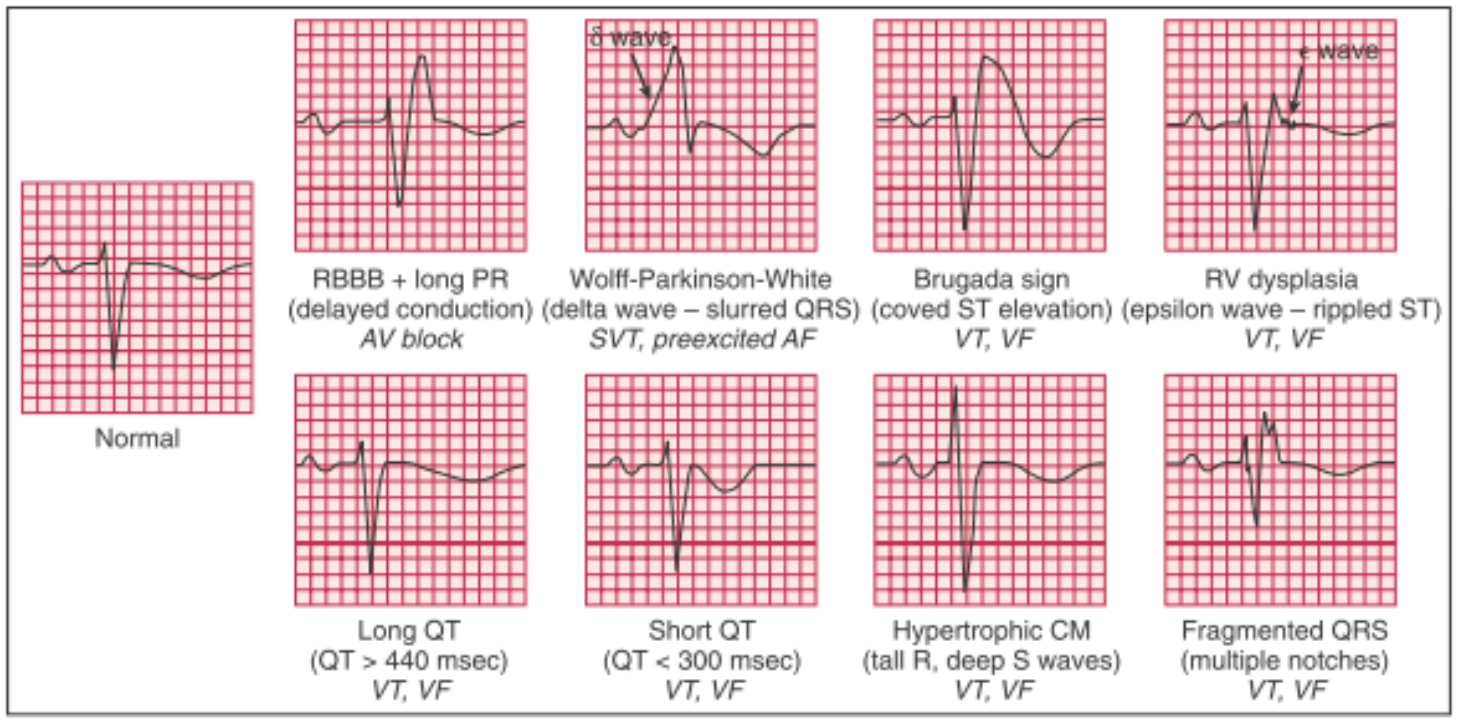


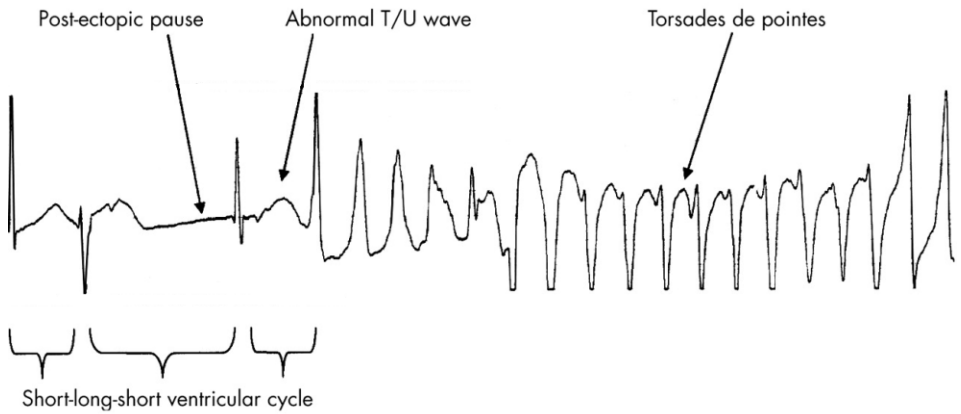
FIGURE 36-4 Electrocardiographic abnormalities in resting rhythm that suggest arrhythmia potential. Lead V_1 is shown in each example; a normal complex is shown at left for reference. AF = atrial fibrillation; AV = atrioventricular; CM = cardiomyopathy; RBBB = right bundle branch block; SVT = supraventricular tachycardia; VF = ventricular fibrillation; VT = ventricular tachycardia.

Long QT syndrome

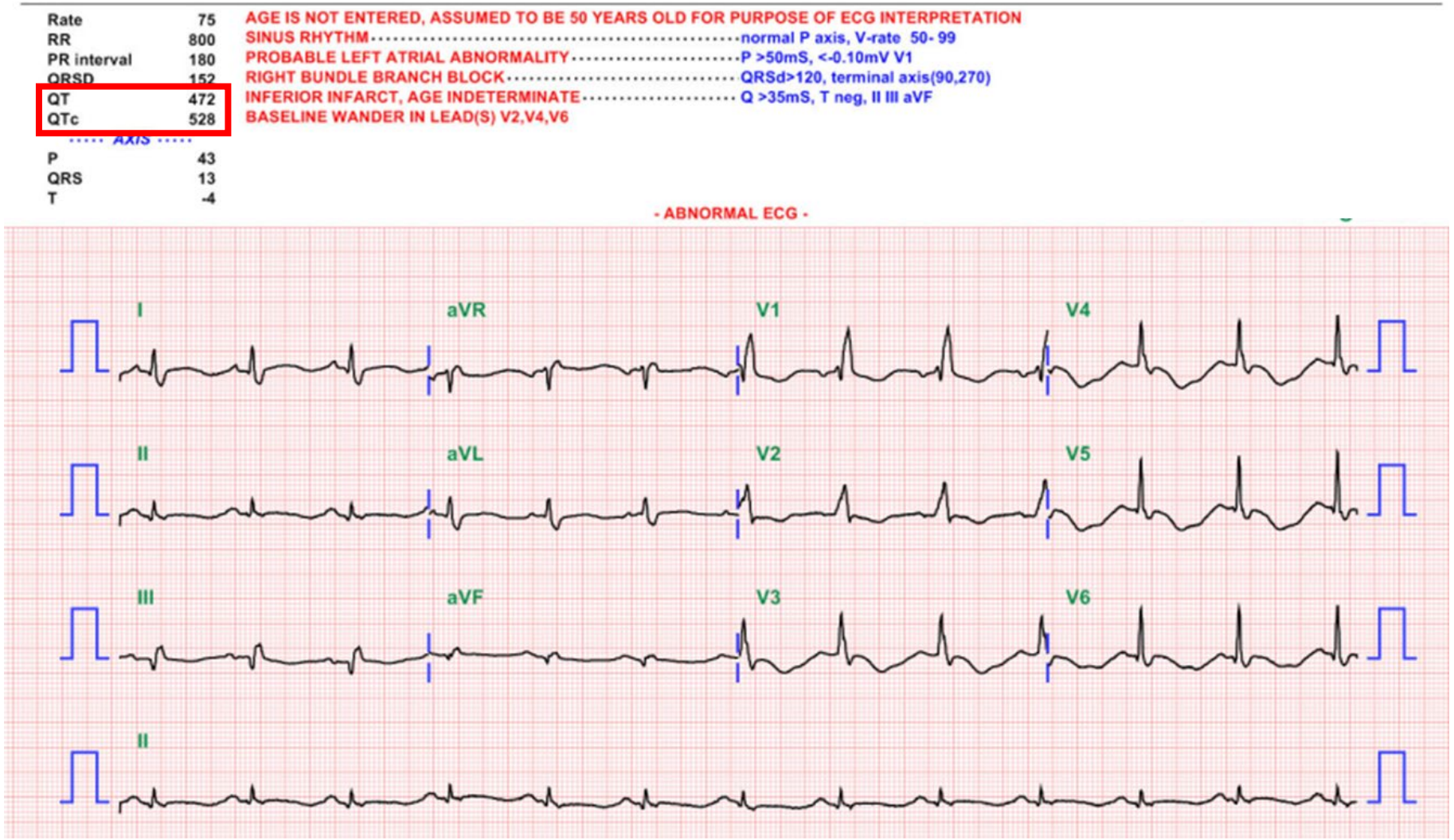
Long QT syndrome

- Congenital
- Acquired : drug, hypokalemia...

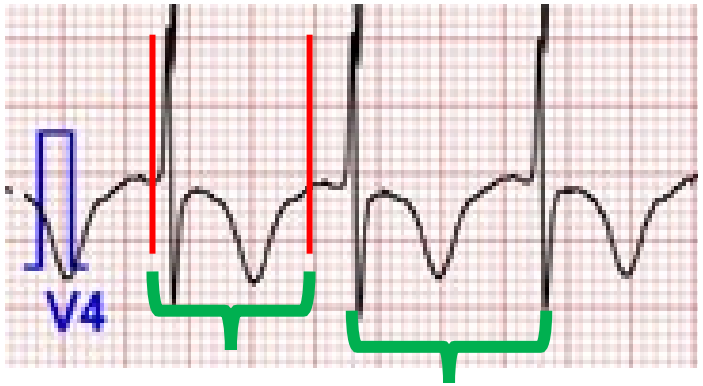
- Torsades de pointes



LQTS

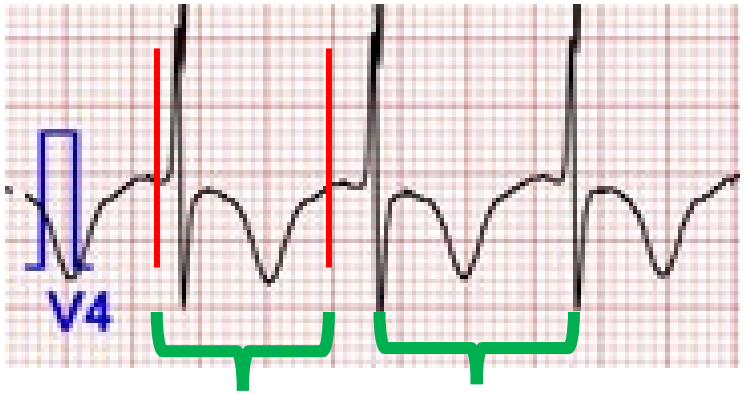

Rate 75
RR 800
PR interval 180
QRSD 152
QT 472
QTc 528
AXIS
P 43
QRS 13
T -4
AGE IS NOT ENTERED, ASSUMED TO BE 50 YEARS OLD FOR PURPOSE OF ECG INTERPRETATION
SINUS RHYTHM normal P axis, V-rate 50- 99
PROBABLE LEFT ATRIAL ABNORMALITY P >50mS, <-0.10mV V1
RIGHT BUNDLE BRANCH BLOCK QRSd>120, terminal axis(90,270)
INFERIOR INFARCT, AGE INDETERMINATE Q >35mS, T neg, II III aVF
BASELINE WANDER IN LEAD(S) V2,V4,V6
- ABNORMAL ECG -
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II

QT interval



$$QTc = \frac{QT}{\sqrt{RR}}$$

- QT = 40ms x 13 = 520ms
- QT 는 HR 이 빨라지면 짧아진다!
- QTc = 520 / √0.6 = 620ms



QT > RR / 2

Long QT Syndrome (LQTS)

- Congenital
  - Various gene mutations → ventricular APD ↑
  - The most common gene mutations of LQTS
    - *KCNQ1* (LQT1), *KCNH2* (LQT2), and *SCN5A* (LQT3)
- Acquired (Drugs, electrolytes imbalances)

Drug-induced QT Prolongation

Table 1. Drugs That May Cause Torsade de Pointes.*

Drugs commonly involved

Disopyramide
Dofetilide
Ibutilide
Procainamide
Quinidine
Sotalol
Bepridil

Other drugs†

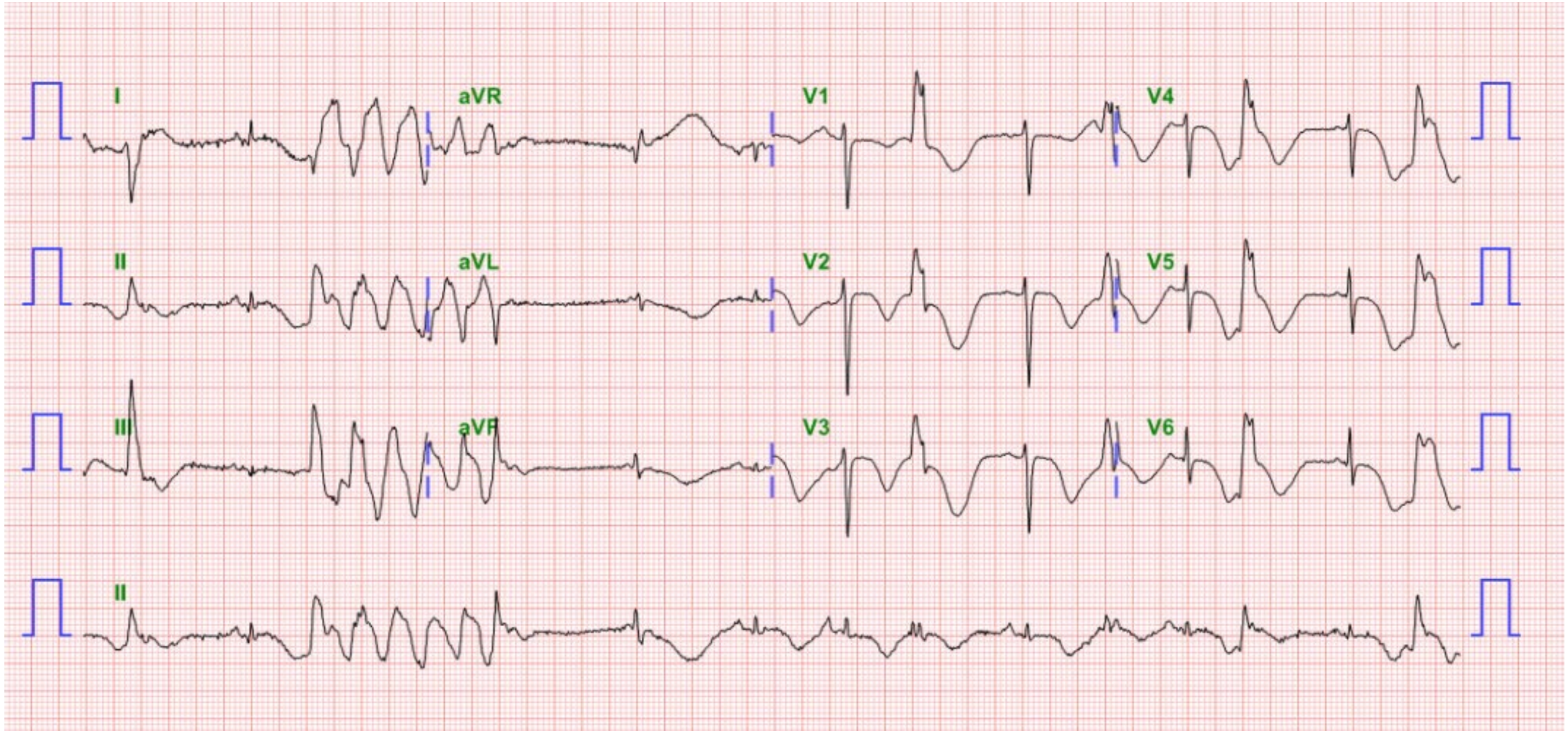
Amiodarone
Arsenic trioxide
Cisapride
Calcium-channel blockers: lidoflazine (not marketed in the United States)
Antiinfective agents: clarithromycin, erythromycin, halofantrine, pentamidine, sparfloxacin
Antiemetic agents: domperidone, droperidol
Antipsychotic agents: chlorpromazine, haloperidol, mesoridazine, thioridazine, pimozide
Methadone

Table 2. Risk Factors for Drug-Induced Torsade de Pointes.*

Female sex[10]
Hypokalemia[11,12]
Bradycardia[11,12]
Recent conversion from atrial fibrillation, especially with a QT-prolonging drug[13,14]
Congestive heart failure[15]
Digitalis therapy[16]
High drug concentrations (with the exception of quinidine)
Rapid rate of intravenous infusion with a QT-prolonging drug[17]
Base-line QT prolongation[16]
Subclinical long-QT syndrome[18,19]
Ion-channel polymorphisms[20-22]
Severe hypomagnesemia

N Engl J Med 2004;350:1013-22.

Long QT and Torsades de Pointes (drug induced)



Torsades de Pointes (TdP)

- "Twisting of the points"
- VT characterized by QRS complexes of changing amplitude (polymorphic) that appear to twist around the isoelectric line and occur at 200-250 bpm
- Causes
  - Congenital/Acquired long QT syndrome
  - Class I_A, III AAD
  - Hypomagnesemia, Hypokalemia
  - Heart block

Torsade de pointes

- Not unusual (especially, post-operation setting)
  - poor diet, hypokalemia, numerous drug, amiodarone…

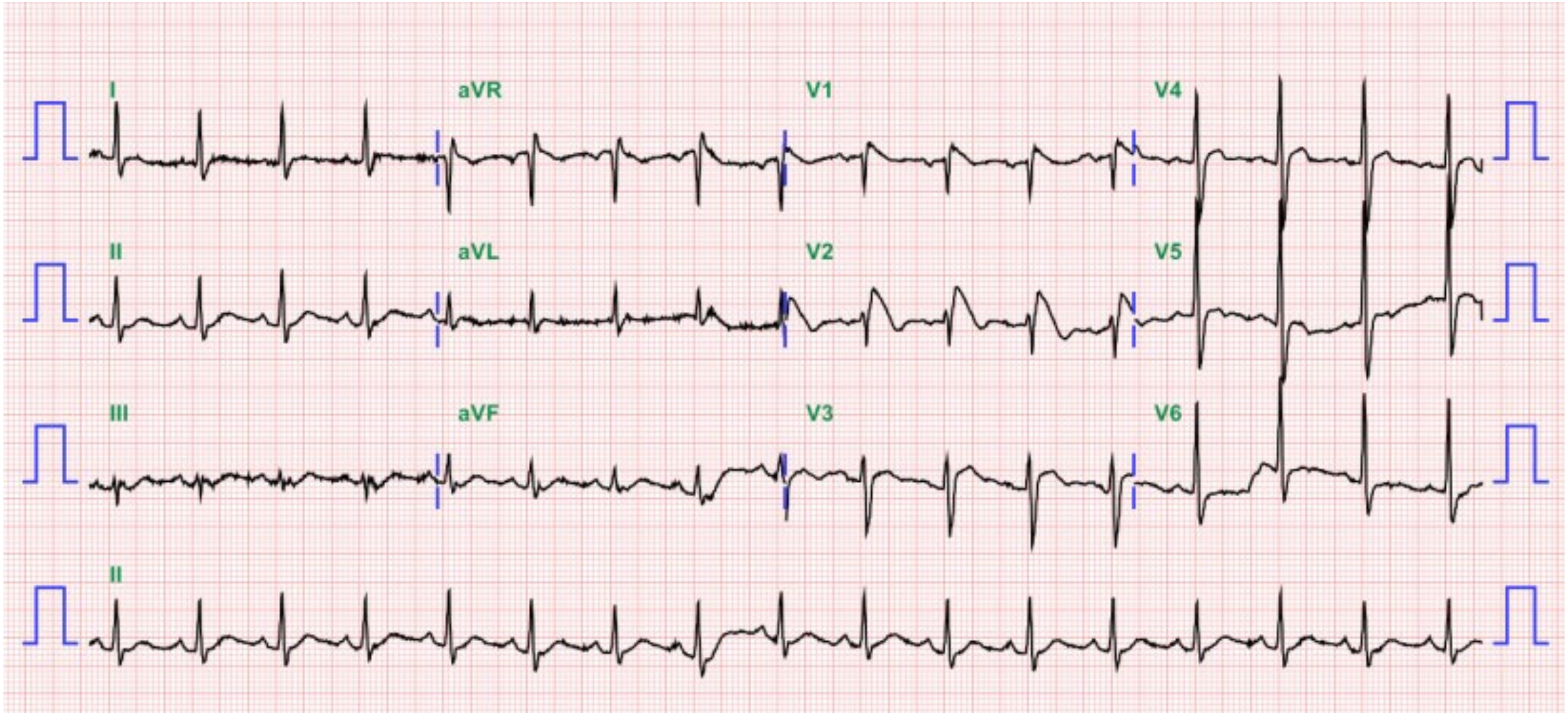
- **Hypokalemia**
  => QT prolongation

- Treatment
  - K^+ correction, iv magnesium (6~10g/day) regardless of level
  - Isoproterenol or temporary pacing

- The only contraindication of **amiodarone** !
  (short-term)

Brugada syndrome

BS



Diagnosis

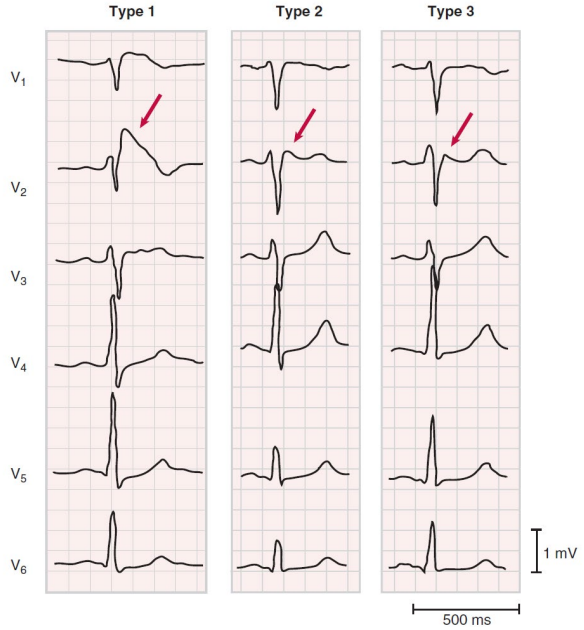
- 2, 3 또는 4번째 흉골간부위에서 V1 또는 V2 중 1개 이상에서 ST-분절 상승이 type 1 morphology ≥2 mm이 자발적 또는 Sodium channel blocker (ajmaline, flecainide, procainamide, pilsicainide) 의한 유발 검사에서 보이는 경우 진단

Brugada Syndrome; ECG Features

TABLE 1. ST-Segment Abnormalities in Leads V_1 to V_3

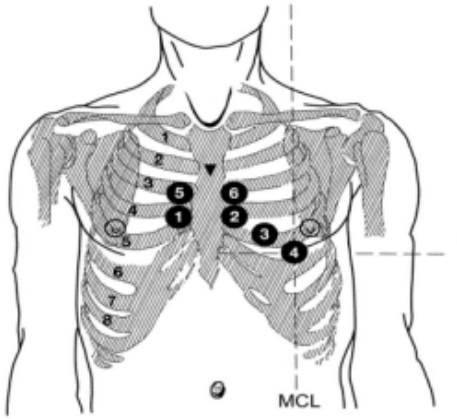
	Type 1	Type 2	Type 3
J wave amplitude	≥2 mm	≥2 mm	≥2 mm
T wave	negative	positive or biphasic	positive
ST-T configuration	coved type	saddleback	saddleback
ST segment (terminal portion)	gradually descending	elevated ≥1 mm	elevated <1 mm

1 mm=0.1 mV. The terminal portion of the ST segment refers to the latter half of the ST segment.



- RBBB pattern and ST elevation in the anterior precordial leads, without structural heart disease
- Signature findings on the ECG can be transient. (Dynamic ECG changes)
- V1 and V2 are placed in a standard or a superior position (up to the 2nd intercostal space).

Flecainide test protocol



- 정상 sinus rhythm 일때 supine position
- Flecainide 2mg/kg – 10min infusion
- VF 발생 가능하므로 응급카트, DC기 준비
- 12lead ECG – 검사전/infusion 동안 2분간격/15분후/30분후

치료

- 증상 없으면 관찰
- ASCD or sustained ventricular arrhythmia = ICD

Catecholaminergic polymorphic VT

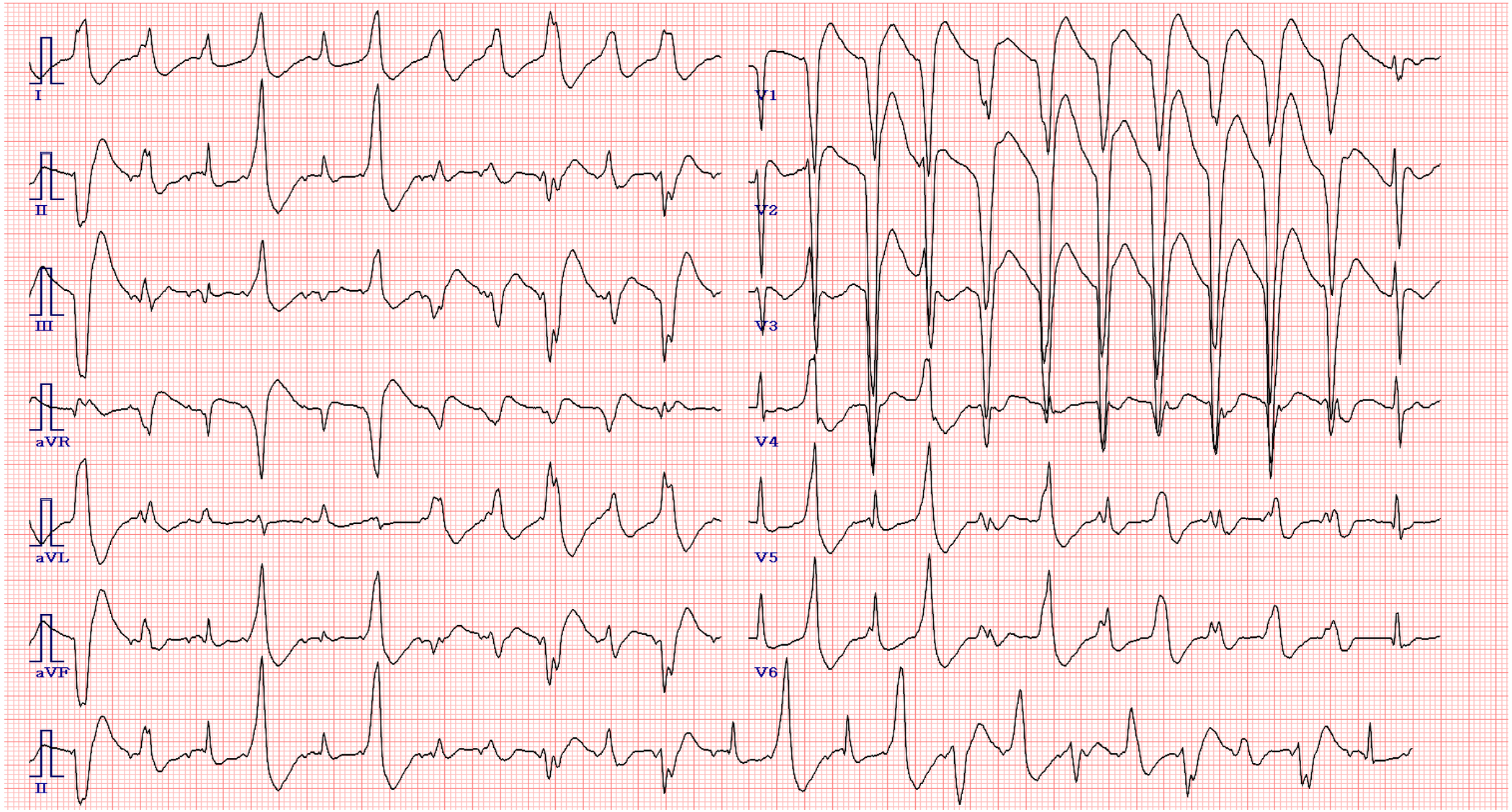
Catecholaminergic polymorphic VT

- CPVT is diagnosed in the presence of a structurally normal heart, normal ECG and exercise- or emotion-induced bidirectional or polymorphic VT.
- CPVT is diagnosed in patients who are carriers of a pathogenic mutation(s) in the genes RyR2 or CASQ2.

4. Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) *Expert Consensus Recommendations on CPVT Diagnosis*

1. CPVT ***is diagnosed*** in the presence of a structurally normal heart, normal ECG, and unexplained exercise or catecholamine-induced bidirectional VT or polymorphic ventricular premature beats or VT in an individual <40 years of age.
2. CPVT ***is diagnosed*** in patients (index case or family member) who have a pathogenic mutation.
3. CPVT ***is diagnosed*** in family members of a CPVT index case with a normal heart who manifest exercise-induced premature ventricular contractions (PVCs) or bidirectional/polymorphic VT.
4. CPVT ***can be diagnosed*** in the presence of a structurally normal heart and coronary arteries, normal ECG, and unexplained exercise or catecholamine-induced bidirectional VT or polymorphic ventricular premature beats or VT in an individual >40 years of age.

18/M, syncope



치료

- Avoidance of competitive sports, strenuous exercise and stressful environments.

- Beta-blockers are recommended in all patients

- ICD implantation in addition to beta-blockers with or without flecainide is recommended in patients with a diagnosis of CPVT who experience cardiac arrest, recurrent syncope or polymorphic/bidirectional VT despite optimal therapy.

Early repolarization

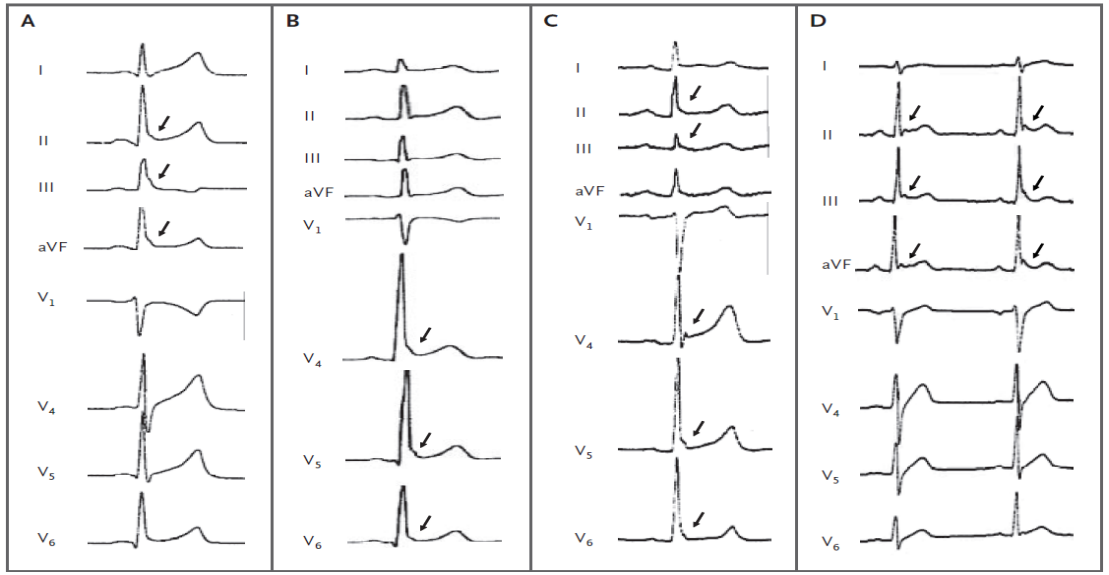
J wave syndrome

- Early repolarization syndrome
- Brugada syndrome
  - RV variant of hereditary J-wave syndromes
  - The region most affected is the ant RVOT
  - J point and ST elevation in the right precordial leads

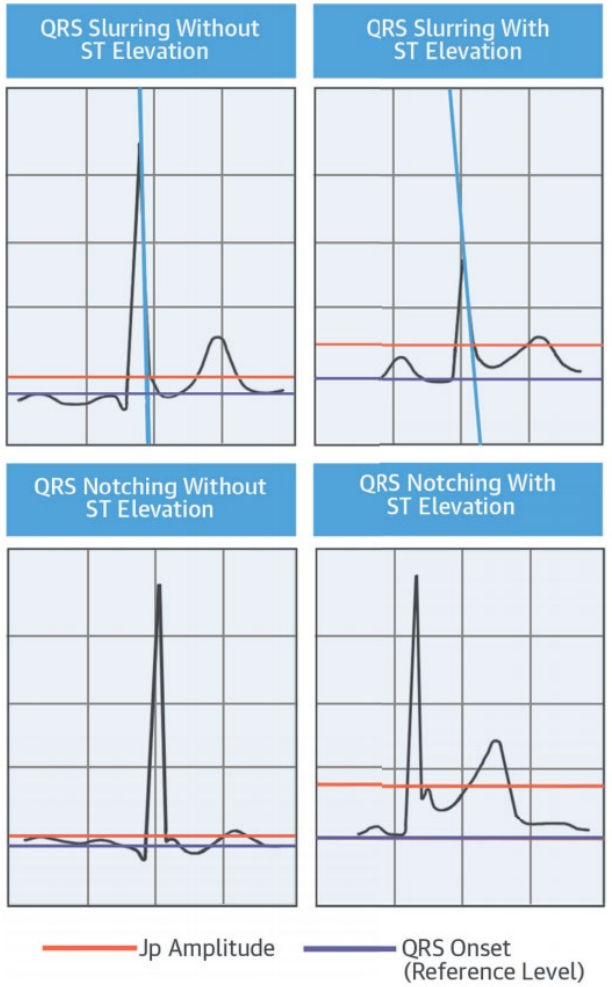
- Vulnerability to polymorphic VT and VF leading to sudden cardiac death

Early repolarization

- Definition
  - J point elevation (QRS slurring or notching)
  - ≥ 0.1 mV, ≥ 2 contiguous inferior and/or lateral leads



Haissaguerre et al, *NEJM* 2008



Macfarlane, P.W. et al. J Am Coll Cardiol. 2015; 66(4):470-7.

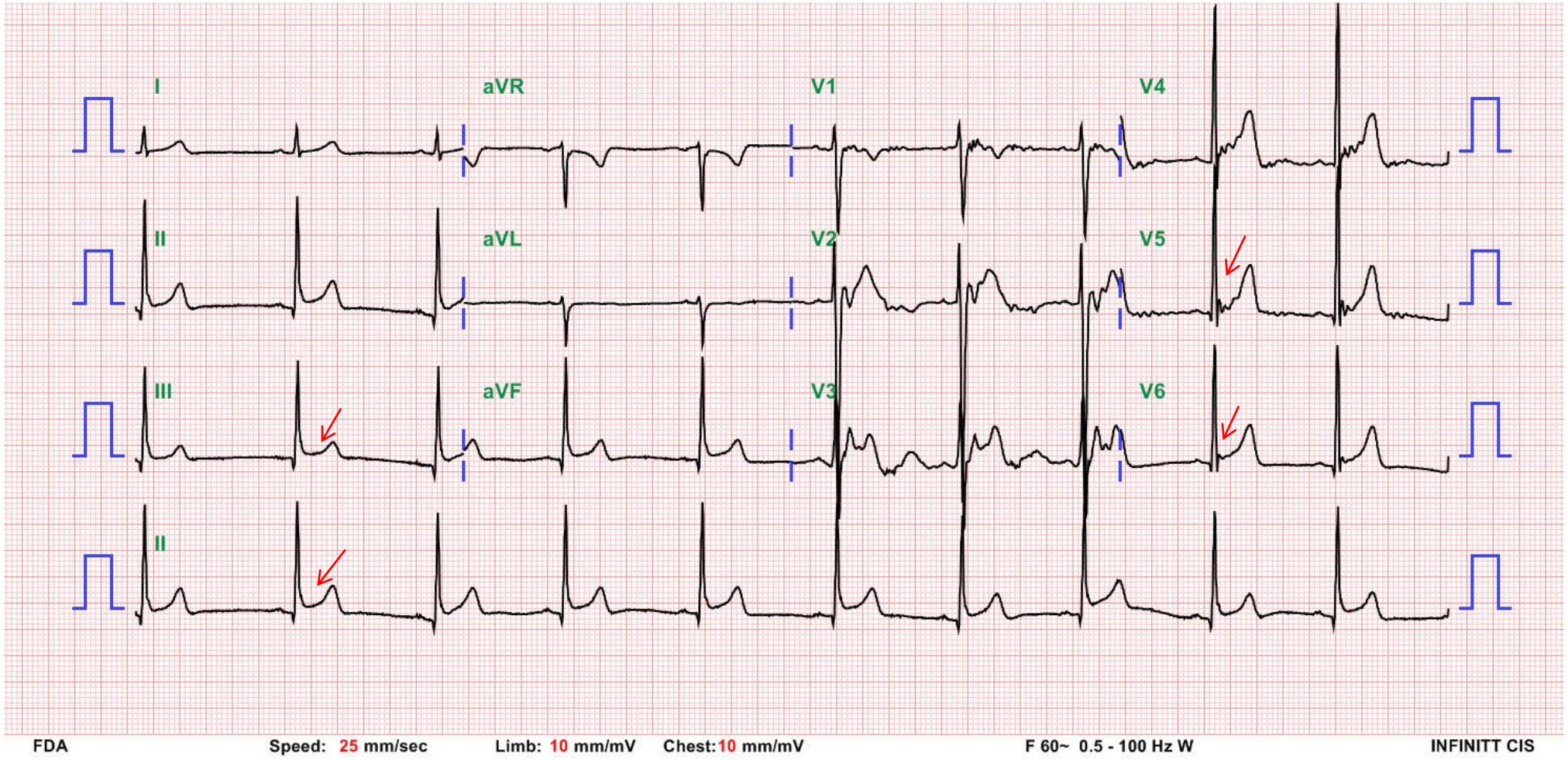
J wave syndrome

- The highest incidence of VF or SCD in the third decade of life (testosterone levels peak in males)
- ST elevation is accentuated during bradycardia or after pauses
- VF occurs during sleep

6. Early Repolarization (ER) *Expert Consensus Recommendations on* ***Early Repolarization Diagnosis***

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 an 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
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

M/18, ECG abnormality



Treatment

- ICD in patients with aborted SCD

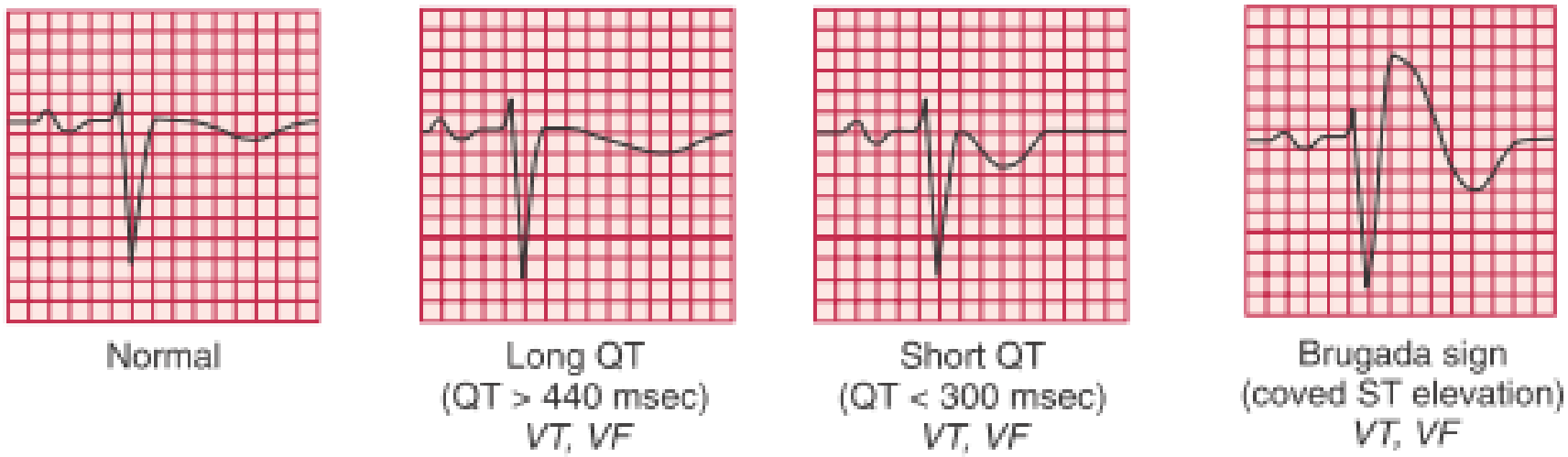
- Pharmacologic approach
  - Effective for electrical storm : isoproterenol, cilostazol
  - Effective general therapy : Quinidine

However,

- Early repolarization is very popular
  - Up to 40% in general population
  - Up to 90% in high-performance athletes

- *How we advise a patient with early repolarization ?*

Summary


Normal
Long QT
(QT > 440 msec)
VT, VF
Short QT
(QT < 300 msec)
VT, VF
Brugada sign
(coved ST elevation)
VT, VF