Syncope in Structural Heart Disease

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심장내과 진 은 선

ICD 보험 인정 기준

- 가. 일시적이거나 가역적인 원인에 의한 것이 아닌 심실세동이나 심실 빈맥에 의한 심정지
- 나. 기질적 심질환이 있는 자발성 지속성 심실빈맥환자

ICD 보험 인정 기준

- 사. 비후성 심근병증 환자로서 아래의 ① ~ ⑤ 중 두 가지이상에 해당되는 경우

  (1) 실신의 증상

  (2) 급사의 가족력

  (3) 좌심실중격의 과도한 비후(>30mm)

  (4) 24시간 활동 중 심전도에서 나타난 비지속성 심실빈맥

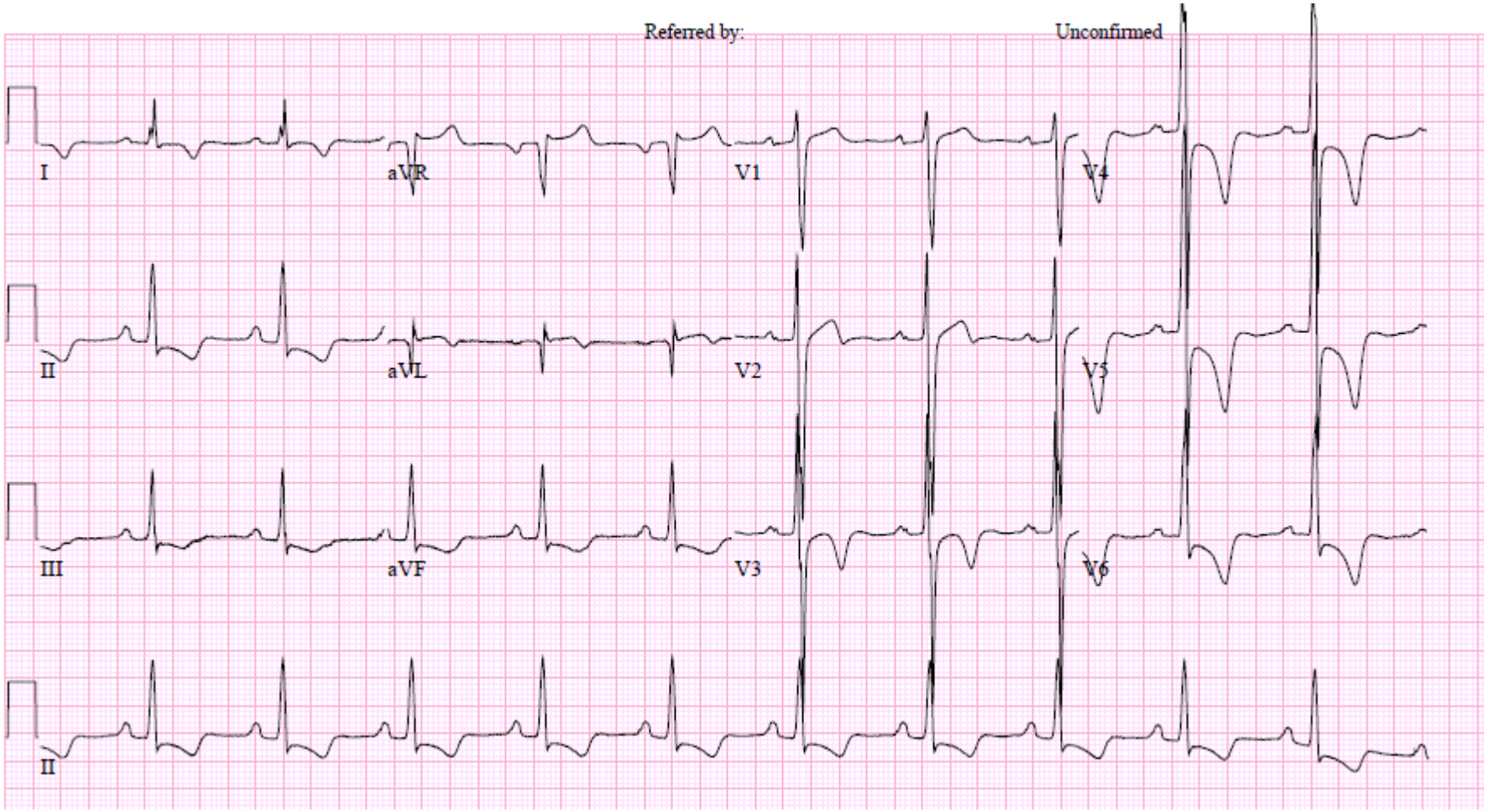
  (5) 운동부하검사 상 이상 혈압증가 반응이 없는 경우(충분한 운동부하에도 혈압상승이 < 20mmHg 인 경우)

CASE 1

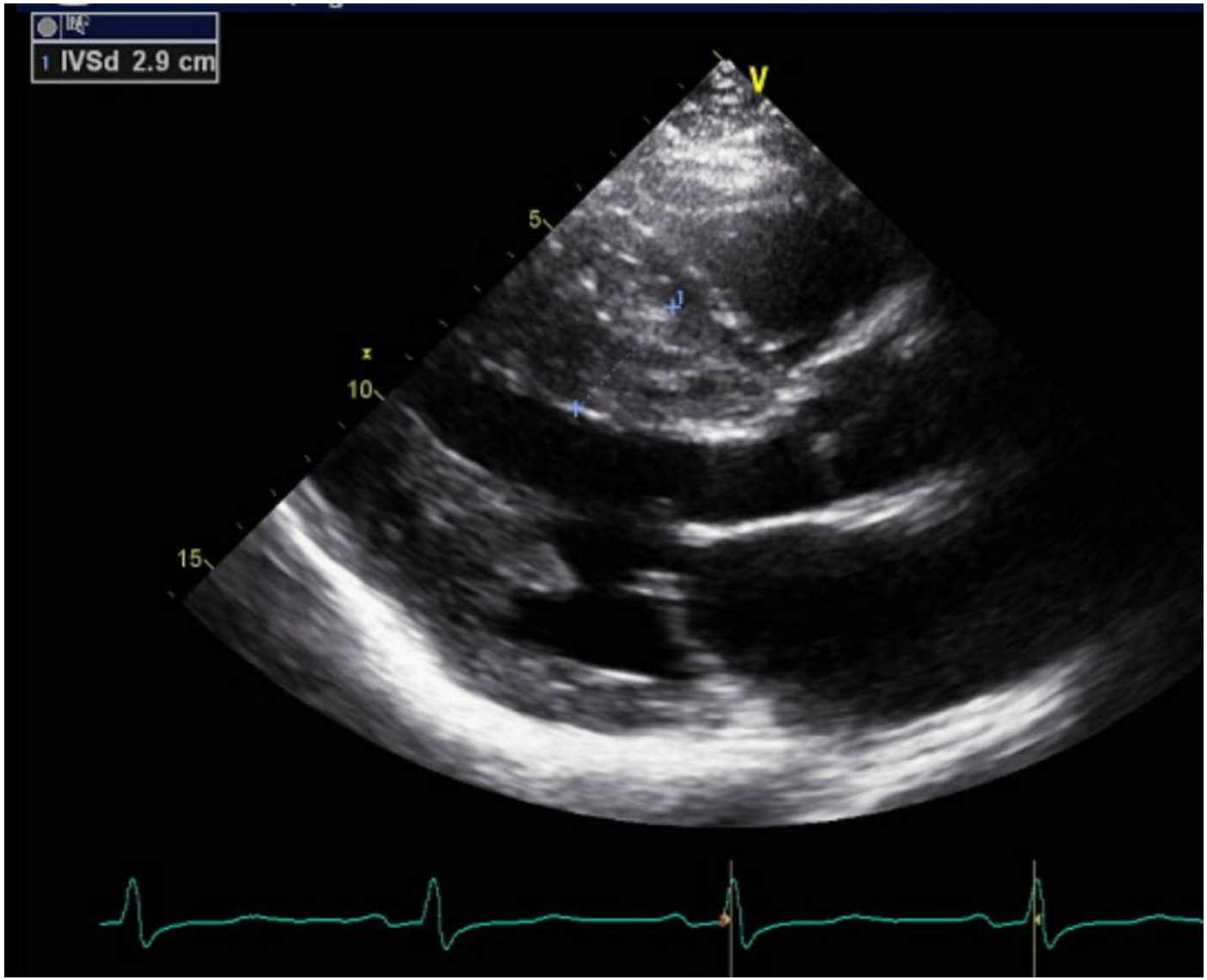
- M/53
- Near syncope
- “가끔 뒤통수가 조였다가 풀리면서 어지럽다” “쓰러질 것 같다”
- 신경과 방문, brain MRI 정상
- 매일 음주

- No FHx of SCD

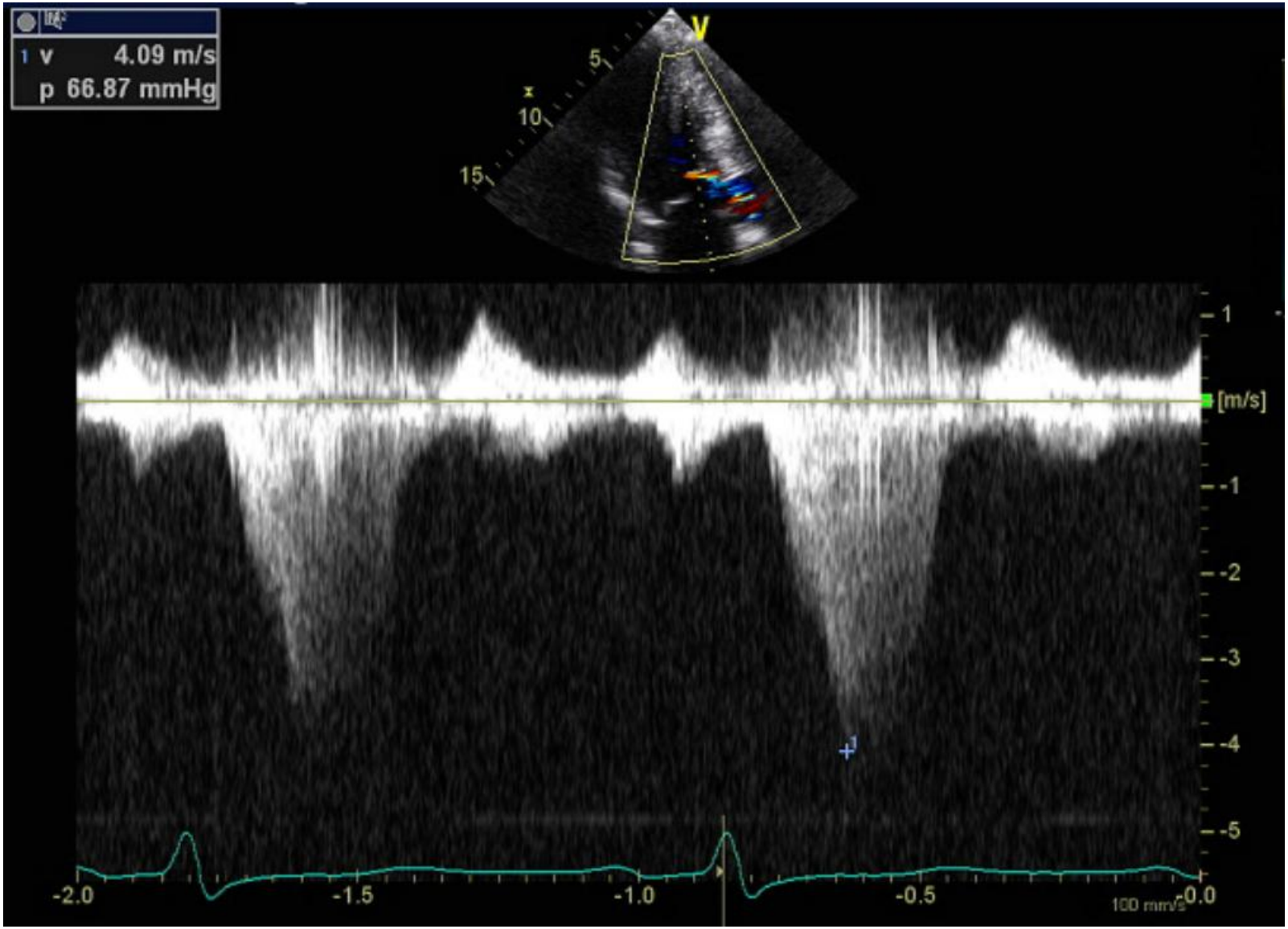
CASE 1 - ECG


Referred by:
Unconfirmed
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II

CASE 1 - Echocardiography

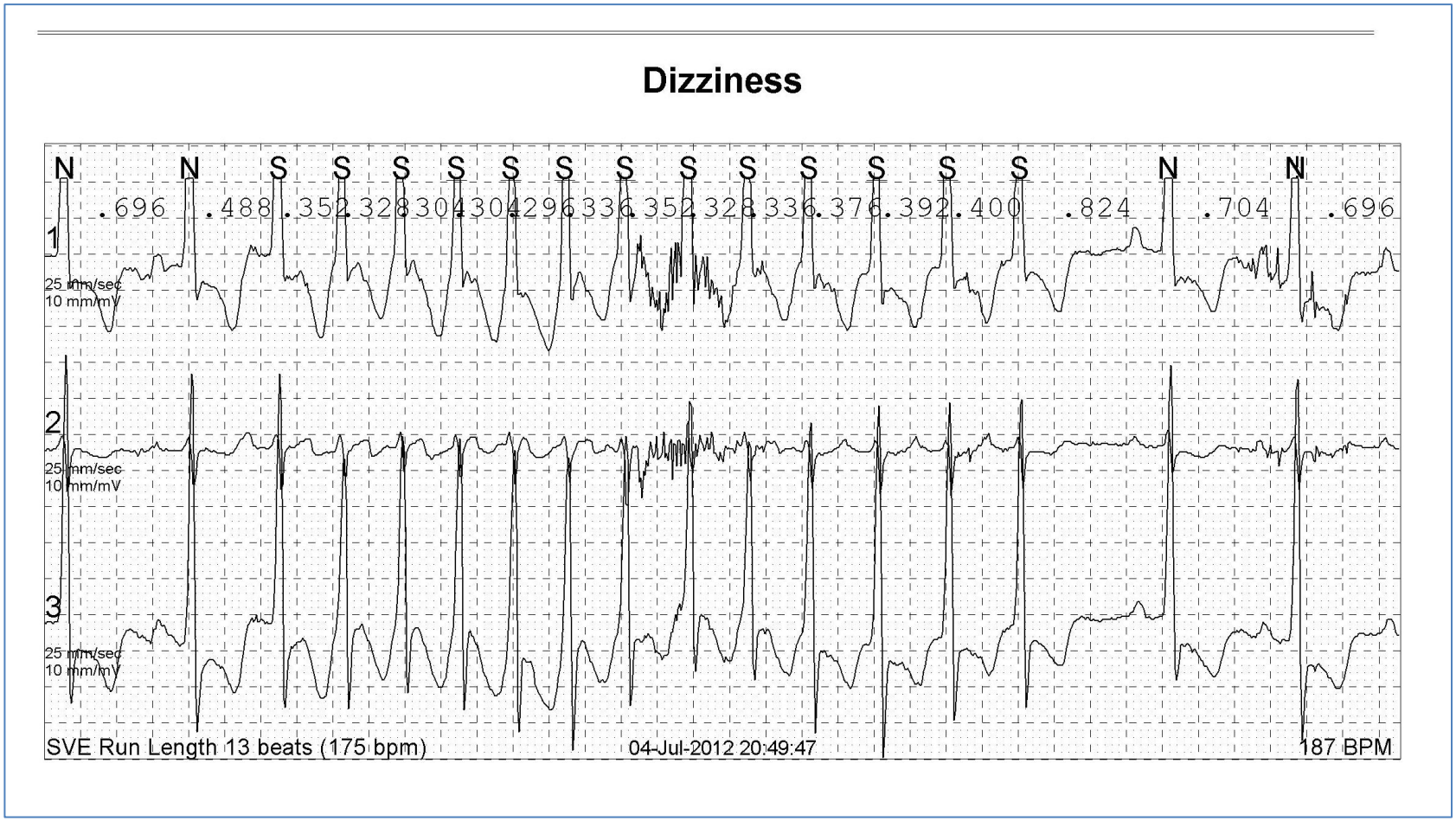


CASE 1 - Echocardiography



CASE 1 - Holter

- Several A tachy episodes, No VT



- TMT; Stage 2, SBP 110 → 125mmHg

CASE 1 – Initial Presentation

- Near syncope
- No Family Hx of SCD
- Septum 29mm
- Short atrial tachyarrhythmia in Holter
- Abnormal BP response?
- LVOT pressure gradient 66 mmHg (Max 77mmHg)

→ **Need for EP Study? NO**

Role of EP Study in HCM

Recommendations on electrophysiologic testing

Recommendations	Class[a]	Level[b]	Ref.[c]
Invasive electrophysiological study is recommended in patients with documented persistent or recurrent supraventricular tachycardia (atrial flutter, atrial tachycardia, atrioventricular nodal re-entry tachycardia, accessory atrioventricular pathway mediated tachycardias) and in patients with ventricular pre-excitation, in order to identify and treat an ablatable substrate.	I	C	249,254 255
Invasive electrophysiological study may be considered in selected patients with documented,			

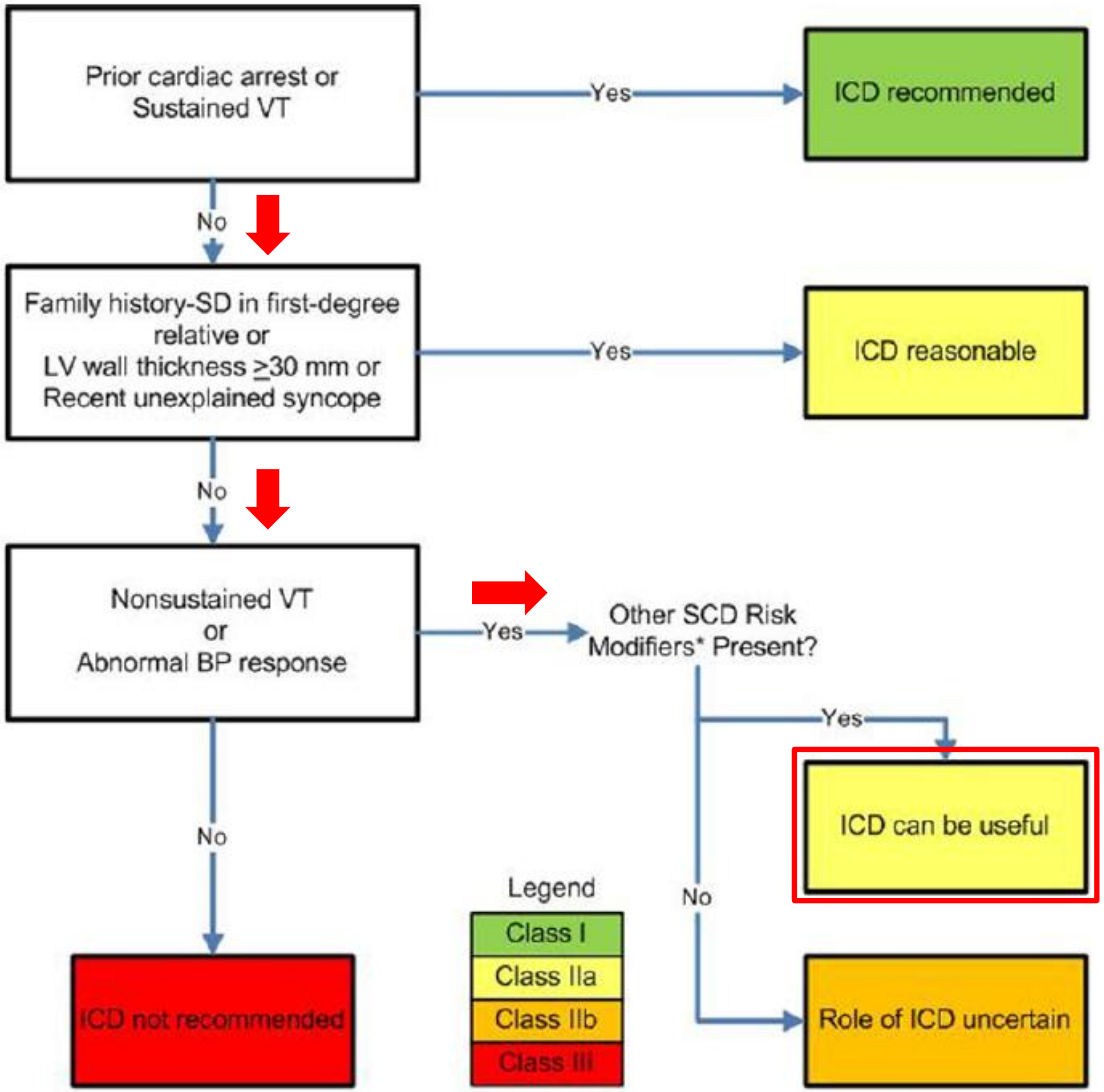
Invasive electrophysiological study with programmed ventricular stimulation is not recommended for sudden cardiac death risk stratification.

III

- NOT for SCD risk
- For SVT
- For VT ablation (Class IIa)

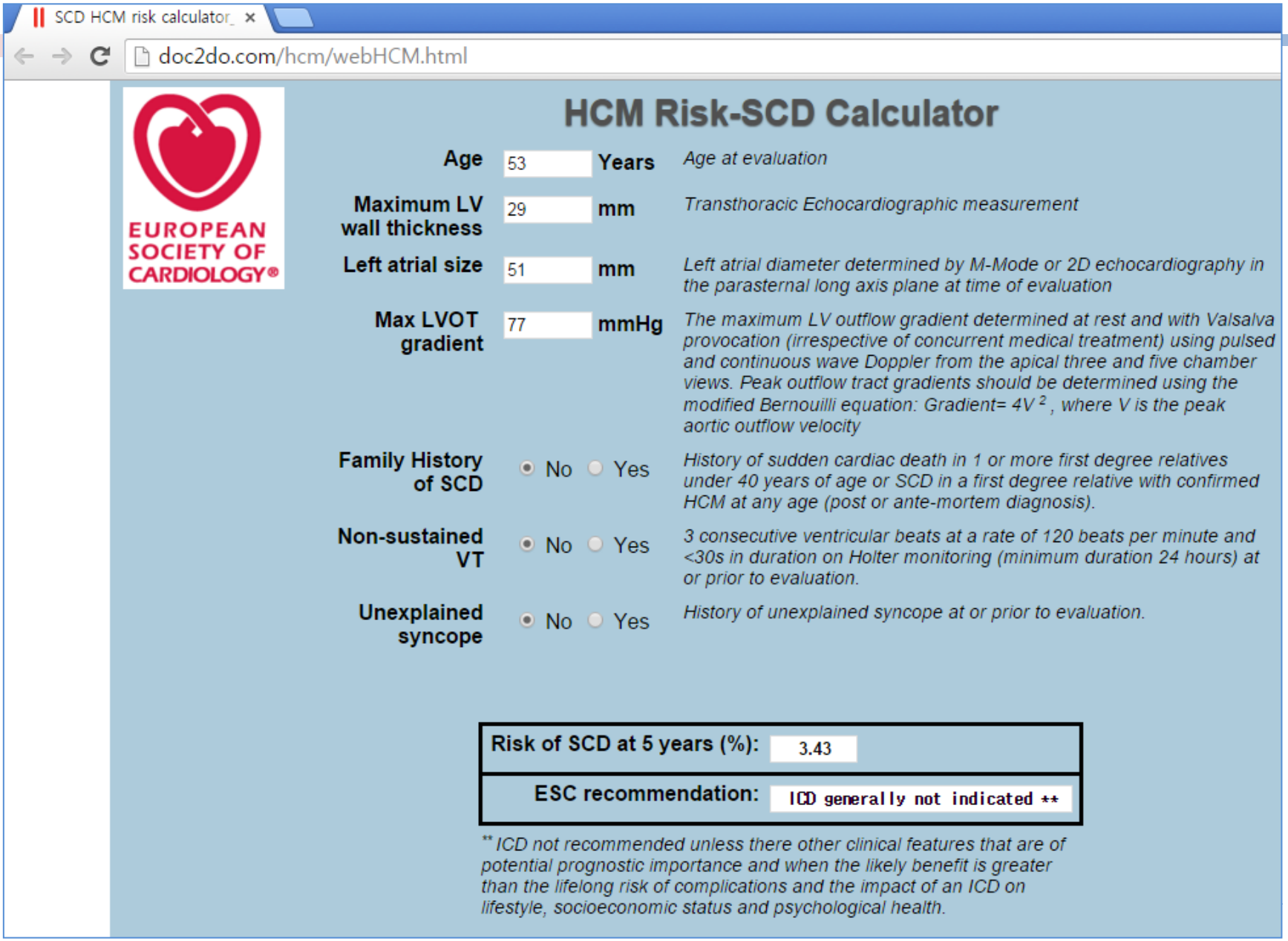
ESC guidelines for HCM 2014

Need for ICD?



ACCF/AHA guidelines for HCM 2011

CASE 1 – SCD Risk score



2014

Clinical Course

- Amiodarone po 200mg → 100mg
  ; "아주 편안하다"

- F/U Holter

General

75092	QRS complexes
238	Ventricular beats (< 1%)
31	Supraventricular beats (< 1%)
< 1	% of total time classified as noise

Ventriculars (V, F, E, I)

236	Isolated
1	Couplets
0	Bigeminal cycles
0	Runs totaling 0 beats

Heart Rates

43	Minimum at 05:28:13 11-Feb
62	Average
114	Maximum at 10:06:47 11-Feb
453	Beats in tachycardia (>100 bpm), < 1% total
24310	Beats in bradycardia (<60 bpm), 32% total
1.52	Seconds Max R-R at 07:11:47 11-Feb

Supraventriculars (S, J, A)

24	Isolated
1	Couplets
0	Bigeminal cycles
1	Runs totaling 5 beats
5	Beats longest run 124 bpm 22:15:08 10-Feb
5	Beats fastest run 124 bpm 22:15:08 10-Feb

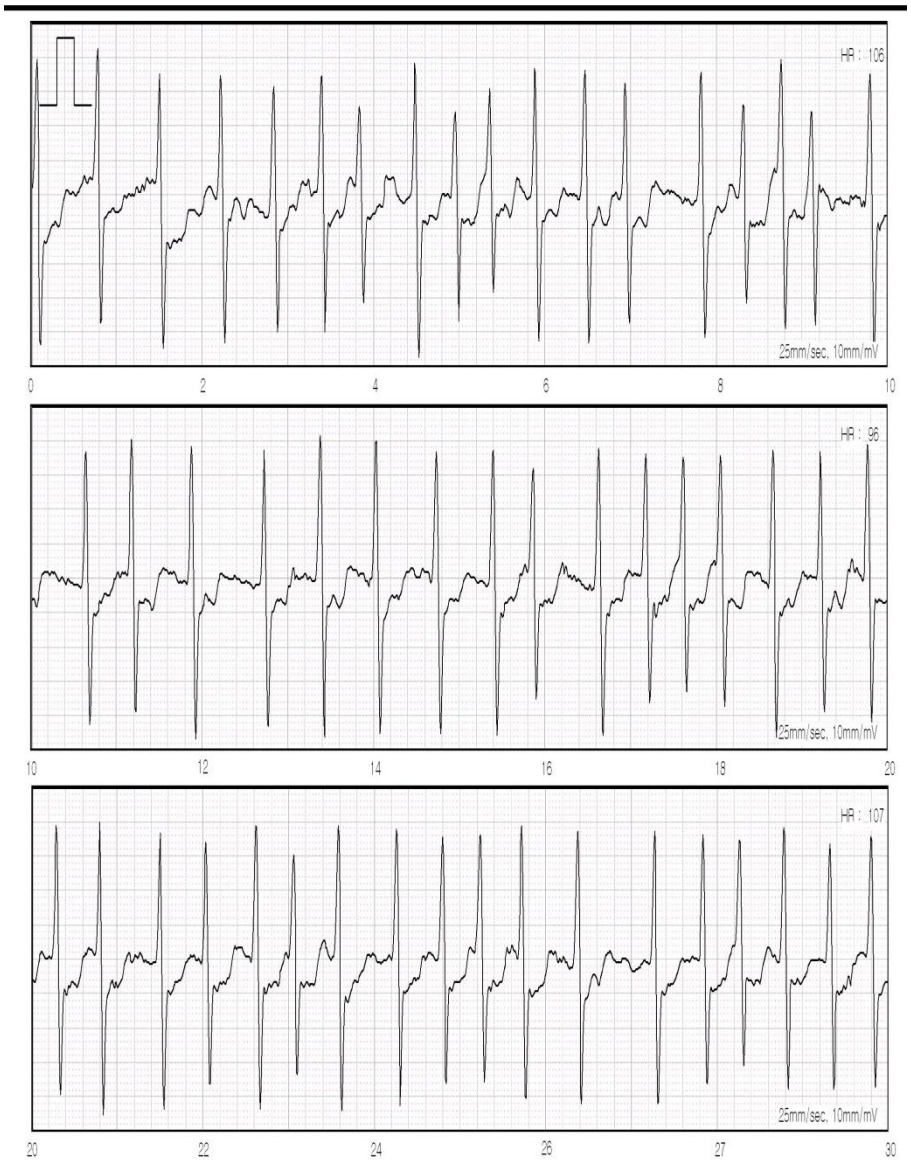
CASE 1 – Clinical Course

- 5개월 후
  ; “음주 조금하고 밤에 자다가 가슴 답답,
  심장이 이상하게 뛰는 일이 있었다”
  “의식 소실, 어지러움 없다”

  → Event recorder 가지고 귀가

- 2주 뒤 cardiac arrest, Successful resuscitation

CASE 1 – Event Recorder



CASE 1 - Debate

- AF with RVR + dynamic LVOT obstruction
  - Precipitates cardiac arrest?

- Severity of LVOT pressure gradient
  - Significance for SCD risk estimation

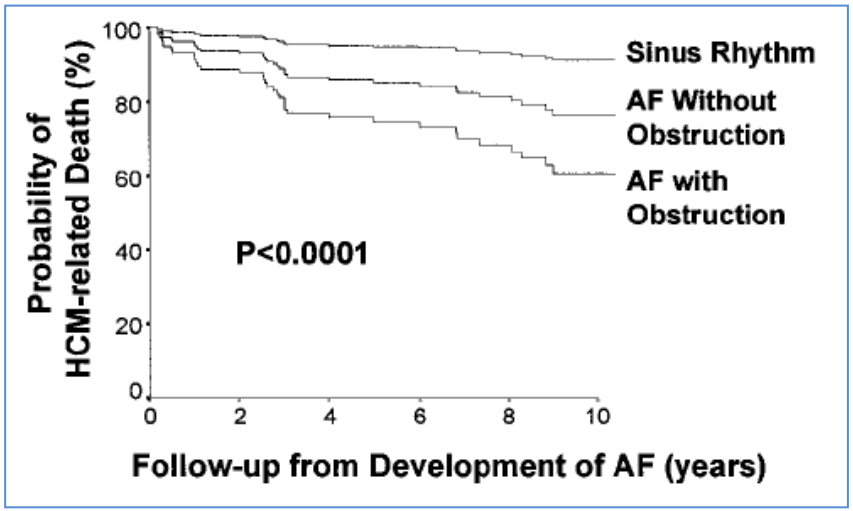
- Was he a high-risk patient of SCD?

AF in HCM patients

Impact of Atrial Fibrillation on the Clinical Course of Hypertrophic Cardiomyopathy

Iacopo Olivotto, MD; Franco Cecchi, MD; Susan A. Casey, RN; Alberto Dolara, MD; Jay H. Traverse, MD; Barry J. Maron, MD

- US cohort, 480 HCM pts, age 45±20 years, F/U for 9.1±6.4 yrs
- AF occurred in 22% (2%/yr)



Impact of AF on HCM-Related Mortality

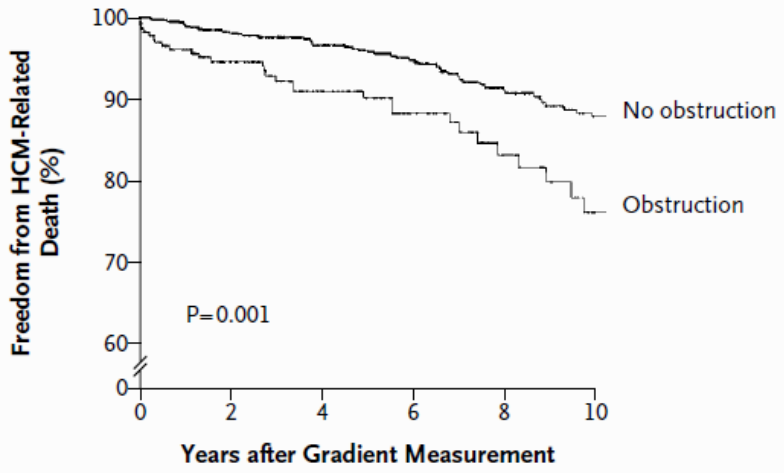
The present study identifies AF as a key determinant of HCM-related mortality and limiting symptoms. Development of AF may indeed represent a clinical turning point, often dominating the clinical picture and decisively influencing long-term outcome. Over a 9-year follow-up, patients with AF showed an ≈4-fold increase in the risk of HCM-related death compared with matched control subjects in sinus rhythm, reflecting significant increases in heart failure and stroke-related mortality. Conversely, we found no relation between clinically evident AF and the occurrence of sudden unexpected death. Therefore, although a causal link between AF and potentially lethal ventricular arrhythmias has been suggested in individual patient reports,[7,22] our data do not support AF as a consistent trigger of sudden death in HCM.

Circulation 2001;104:2517

LVOT pressure gradient

ORIGINAL ARTICLE

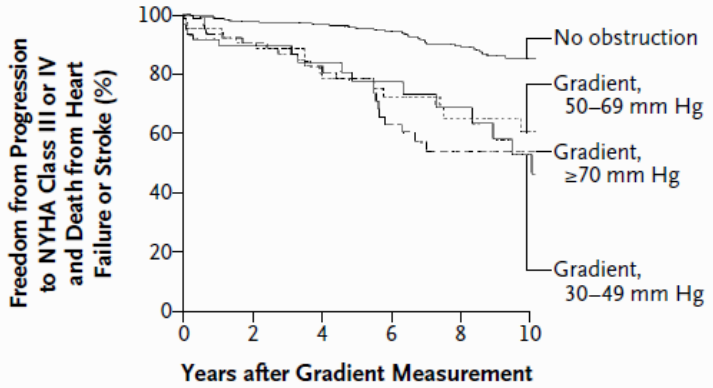
Effect of Left Ventricular Outflow Tract Obstruction on Clinical Outcome in Hypertrophic Cardiomyopathy



No. at Risk

No obstruction	828	594	495	360	247	201
Obstruction	273	178	130	84	54	35

Figure 1. Probability of Hypertrophic Cardiomyopathy (HCM)–Related Death among 273 Patients with a Left Ventricular Outflow Gradient of at Least 30 mm Hg under Basal Conditions and 828 Patients without Obstruction at Entry.



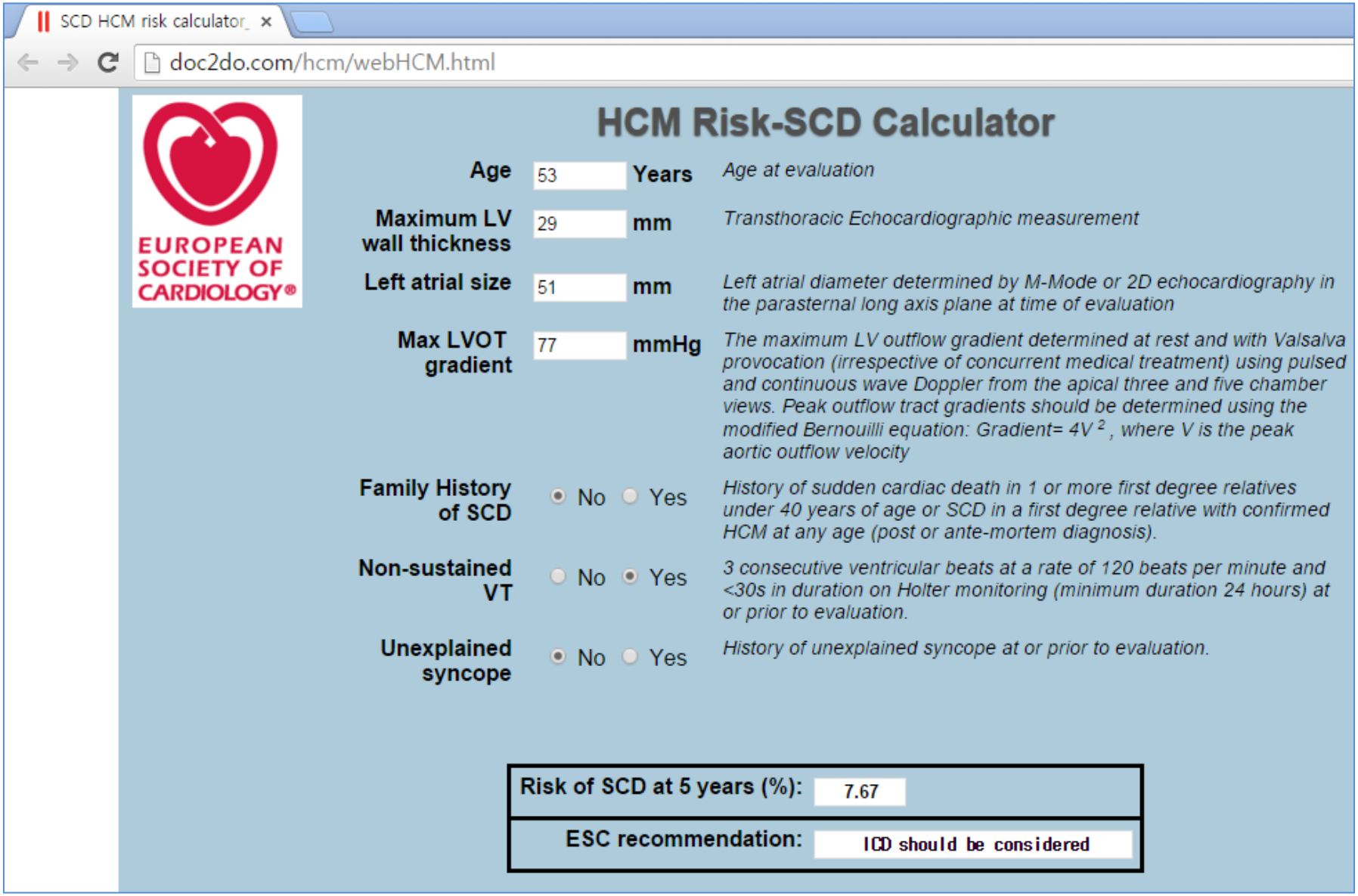
No. at Risk

No obstruction,	770	557	464	334	231	188
Gradient, 30–49 mm Hg	62	38	28	18	12	8
Gradient, 50–69 mm Hg	73	50	37	24	16	10
Gradient, ≥70 mm Hg	89	56	38	24	11	7

Figure 5. Relation of the Magnitude of Left Ventricular Outflow Tract Gradient or the Absence of a Gradient to the Probability of Progression to Severe Heart Failure (NYHA Class III or IV) or Death from Heart Failure or Stroke.

P<0.001 for the comparison of the group without obstruction with each subgroup with obstruction; P>0.30 for each comparison among the subgroups with obstruction. Patients who were already in NYHA class III or IV at entry were excluded from the analysis.

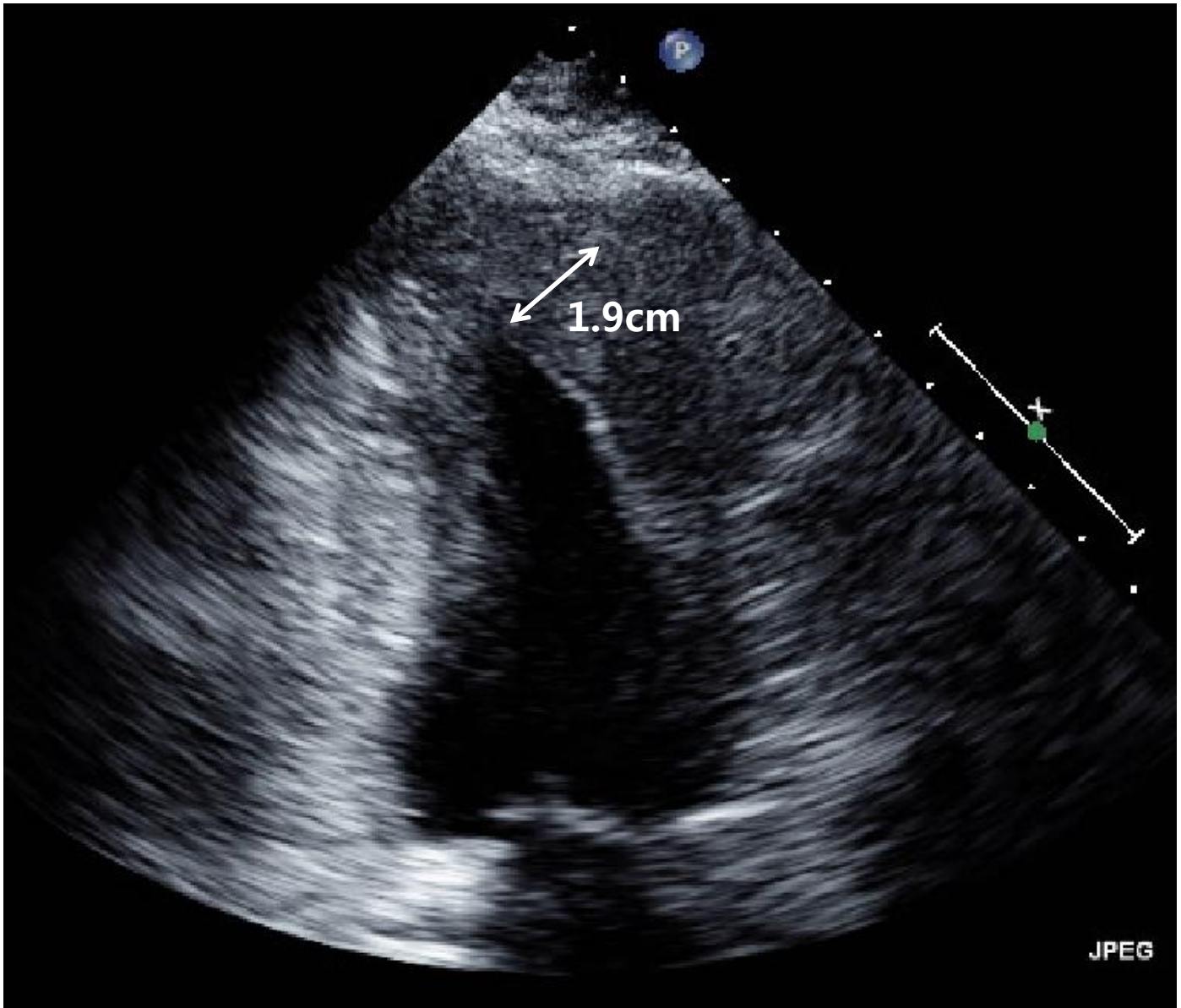
CASE 1- If NVST(+)



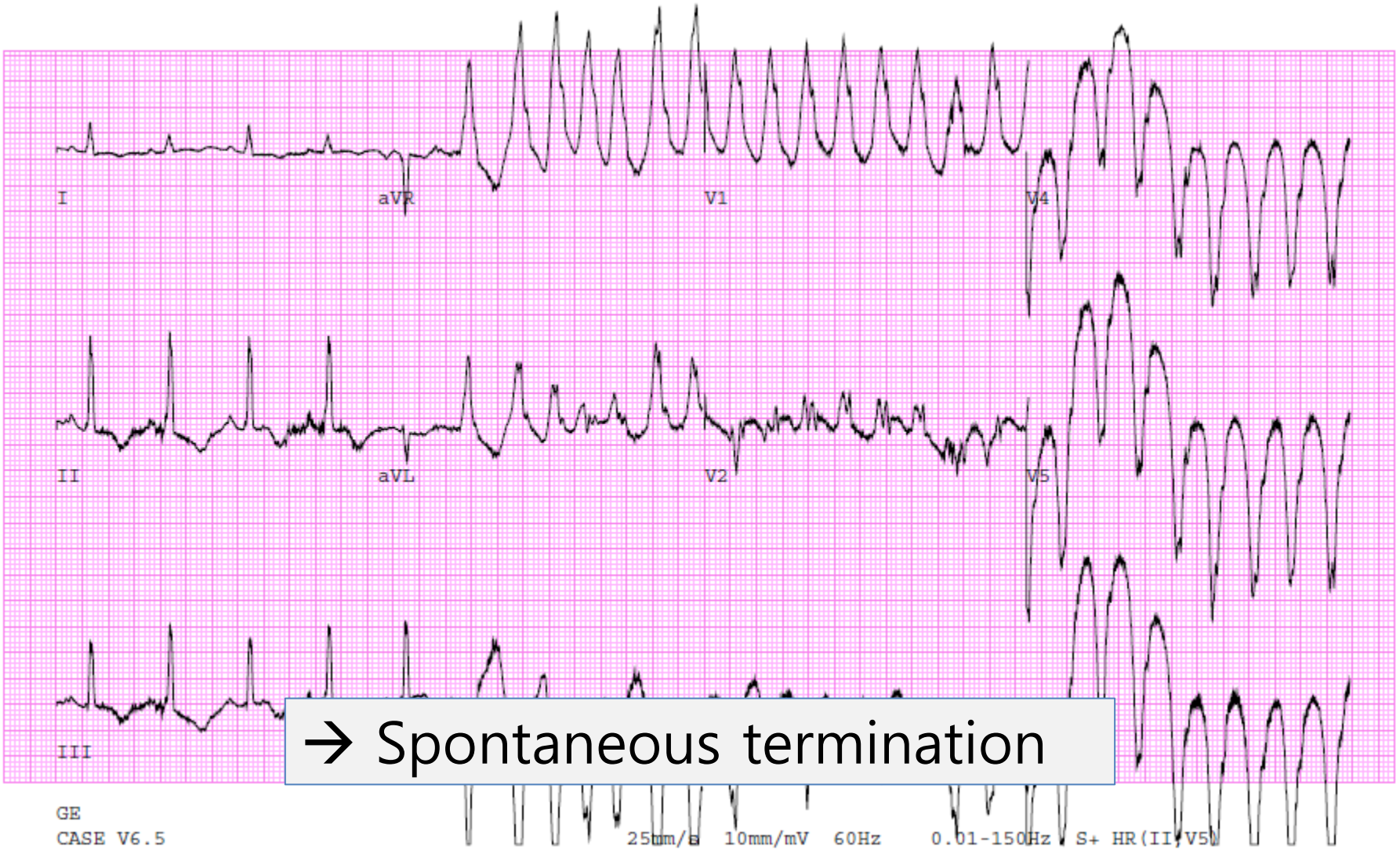
CASE 2

- M/59
- 4-5년 전 앉았다가 일어설 때 눈앞이 깜깜
- “화낼 때 가슴 두근거릴 때가 있었다”
- Treadmill test 중 VT 발생, “답답하다”

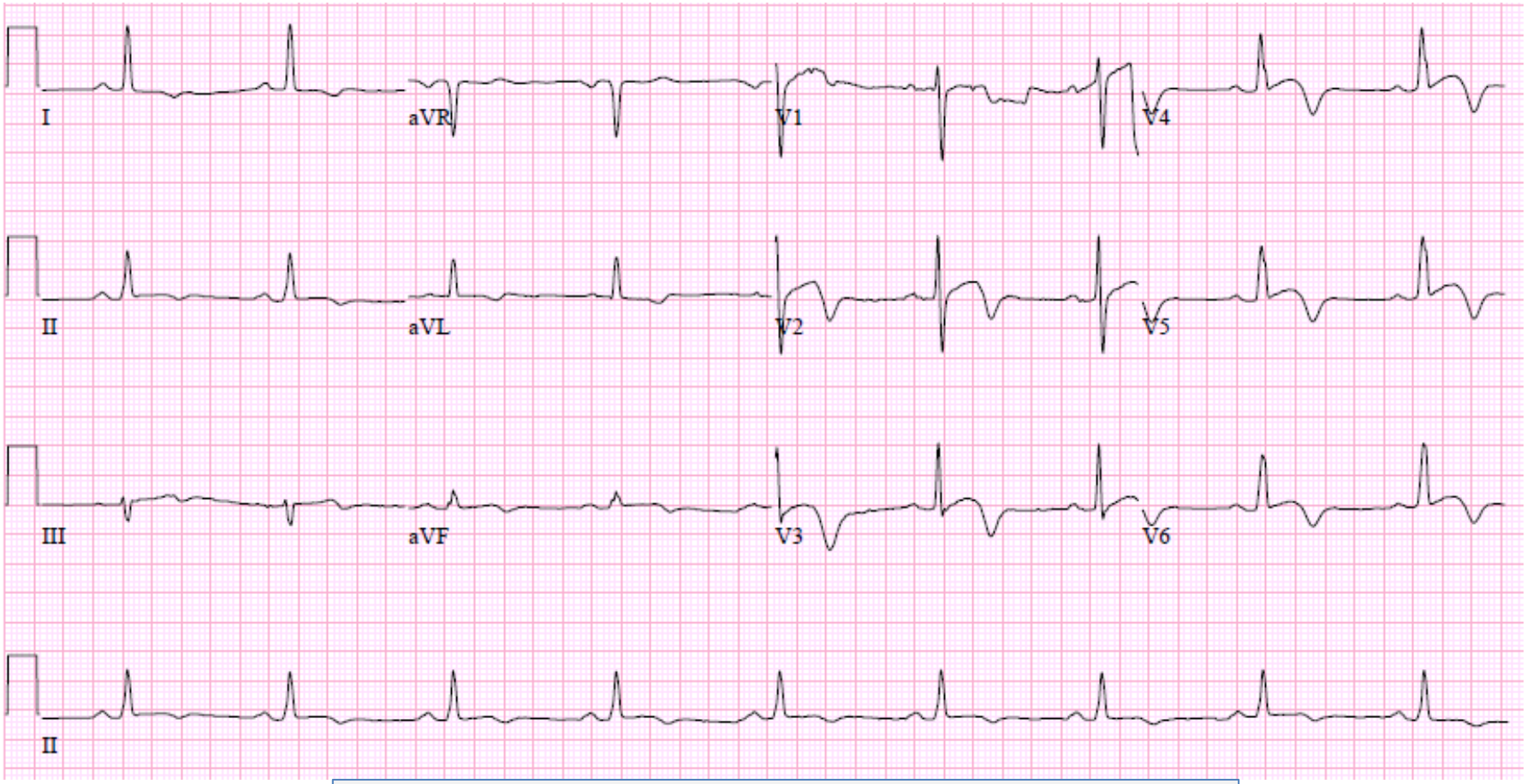
CASE 2



CASE 2 - TMT

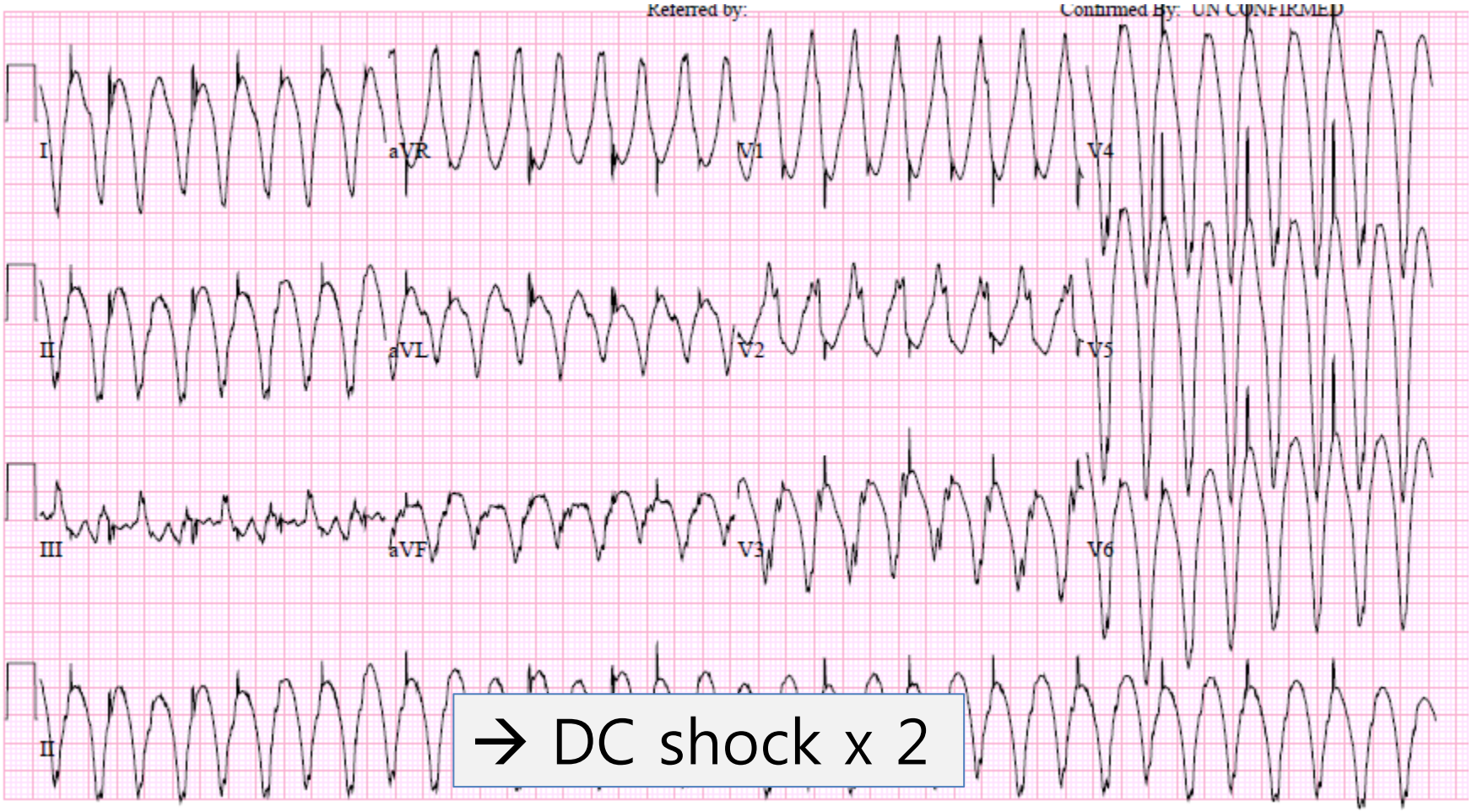

I
aVR
V1
V4
II
aVL
V2
V5
III
→ Spontaneous termination
GE
CASE V6.5
25mm/s 10mm/mV 60Hz 0.01-150Hz S+ HR(II,V5)

CASE 2 - ECG



→ Medical Treatment & F/U

CASE 2 – 4m later, In Anger



가슴이 답답하고 두근거림, SBP 100mmHg

CASE 2 – Clinical Course

- ICD implantation
- Propranolol, verapamil po → stopped
- No VT during F/U

- 2 years later, ICD pocket site skin erosion & infection → Device Extraction

Apical HCM

Long-Term Outcome in Patients With Apical Hypertrophic Cardiomyopathy

Maria J. Eriksson, MD, PHD, Brian Sonnenberg, MD, Anna Woo, MD, FACC, Paul Rakowski, Thomas G. Parker, MD, FACC, E. Douglas Wigle, MD, FACC, Harry Rakowski, MD, FACC

- 105patients aged 41.4±14.5 years
- F/U for 13.6±8.3 years
- AF 12%, MI 10%, VF in one case
- CV mortality 1.9%, annual CV mortality 0.1%
- Predictors of CV morbidity; age at presentation <41 yrs, LA enlargement, NYHA class ≥II at baseline

J Am Coll Cardiol 2002;39:638

Apical HCM

- Milder clinical course
- Annual mortality; HCM 6% vs apical HCM ≤1%
- Can cause VT/VF, cardiac arrest

- SCD risk equal to typical HCM? Possibly, NO
- Further study is needed to set up the guidelines

Messages

- In HCM patients, SCD risk estimation is always important
- Try to collect enough data
- Calculated SCD risk score is useful

- Apical HCM
  - A variant of HCM
  - Common in Asia
  - Mild clinical course
  - SCD risk estimation?

Thank you
for your attention