비후성 심근증 (Hypertrophic Cardiomyopathy)

급사 증례

증례

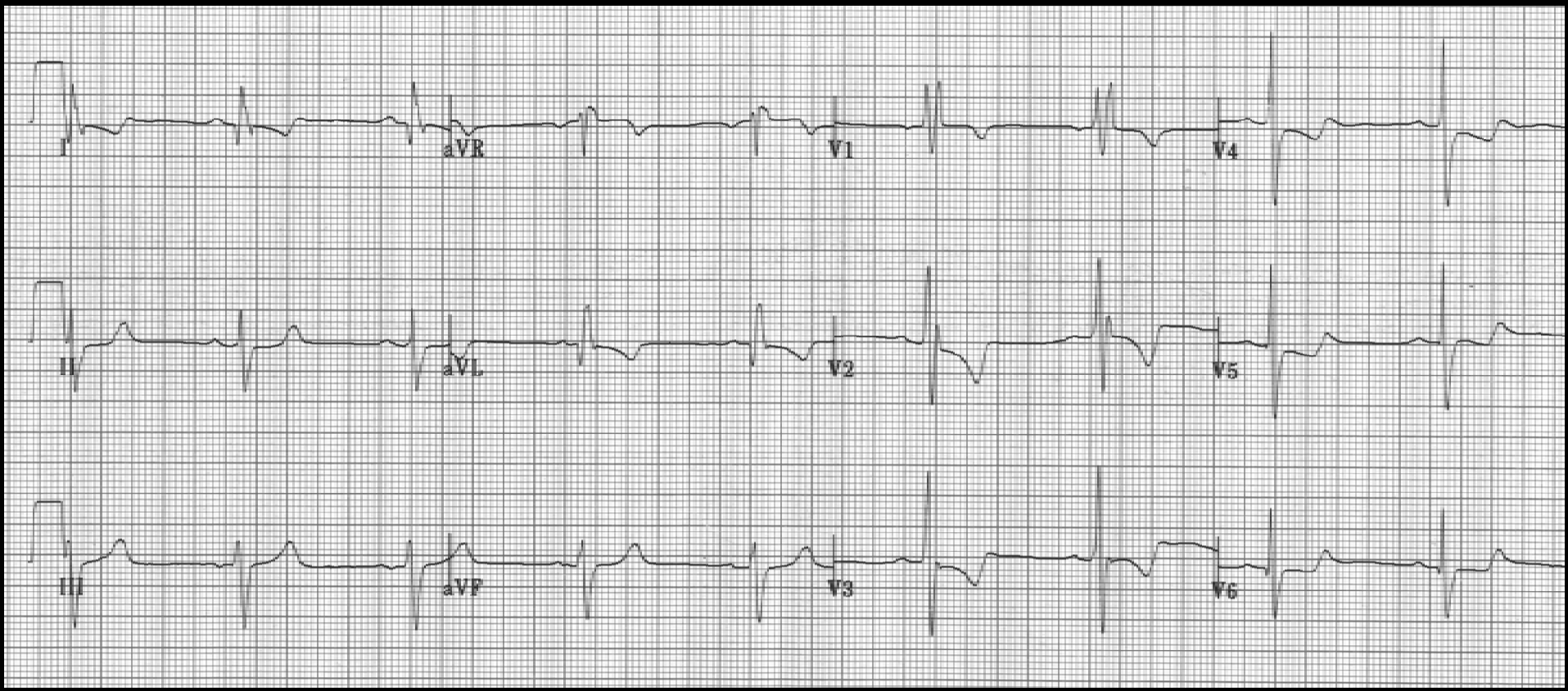
최ㅇ옥 F/45

1988 년 심잡음, 심전도 이상으로 비후성 심근증 진단

2001년 1월 : 맥주마시고 계단 올라가다가 계단에서 잠시 실신

2001년 9월 : 입원

EchoCG : HCM w LVOT Obstruction
(Severe dynamic LVOT obstruction)
Septal wall thickness : 26 mm, PG 118 mmHg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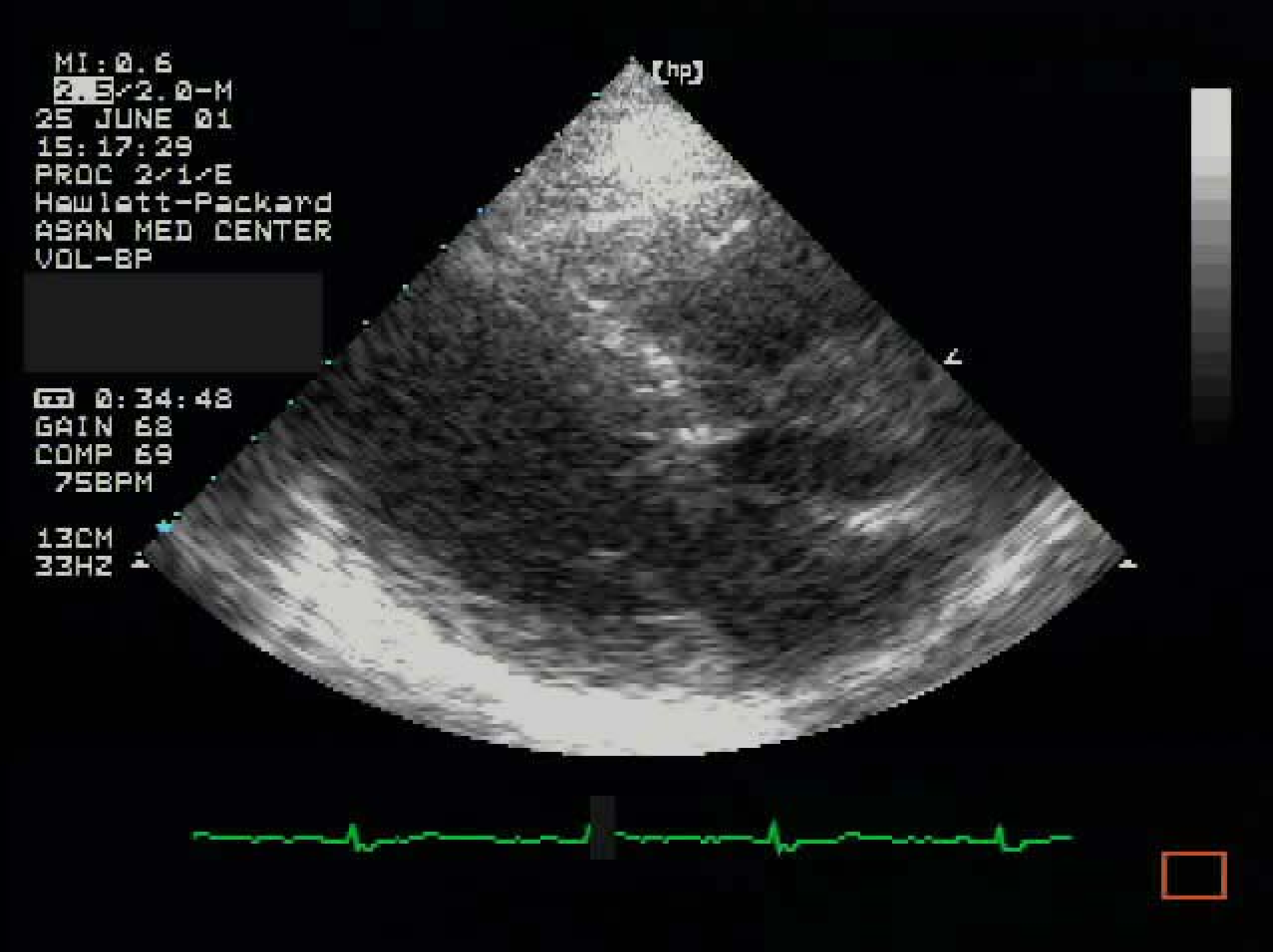
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6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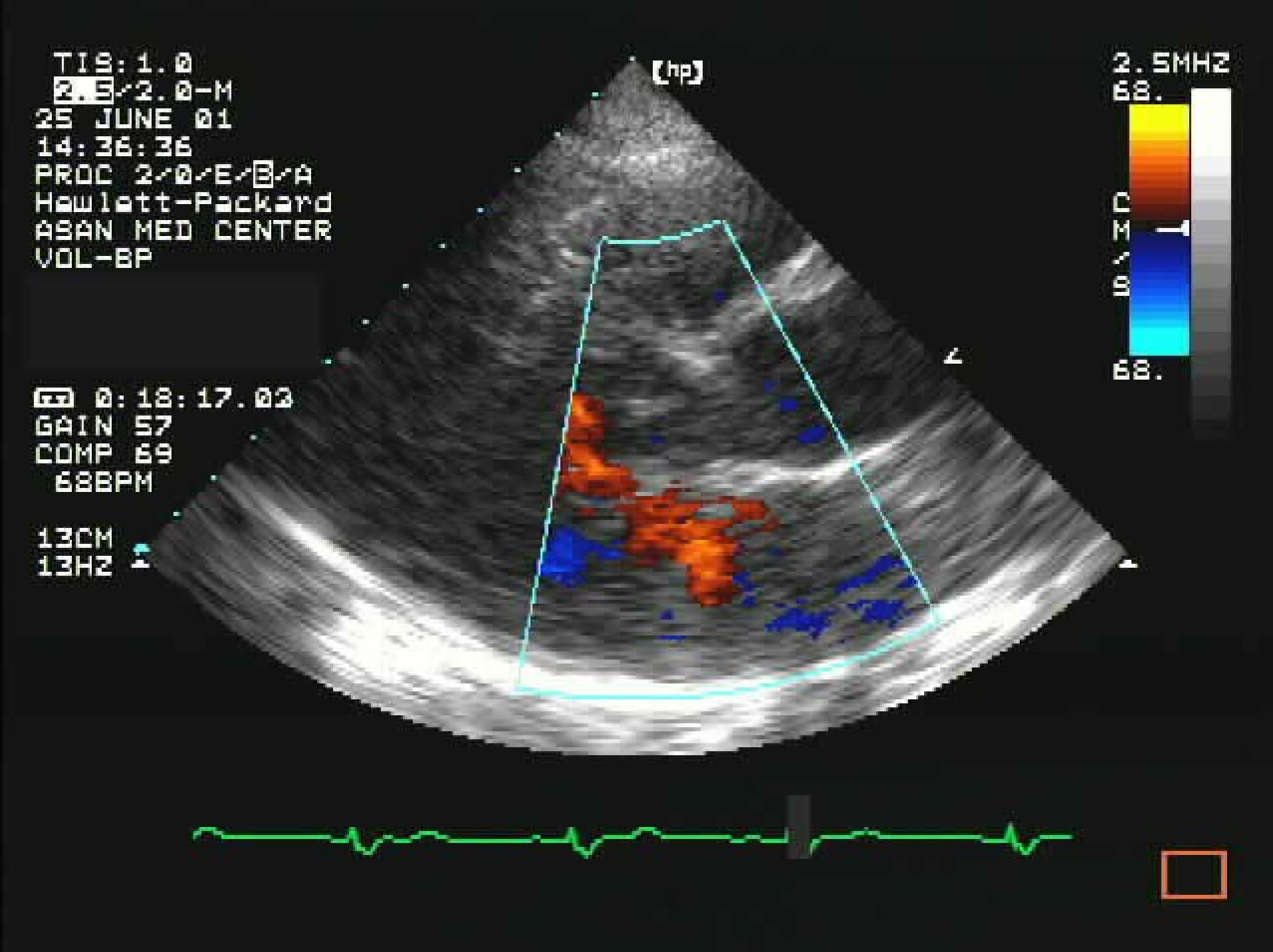
GAIN 68
COMP 69
75BPM
13CM
33HZ
[hp]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Hewlett-Packard
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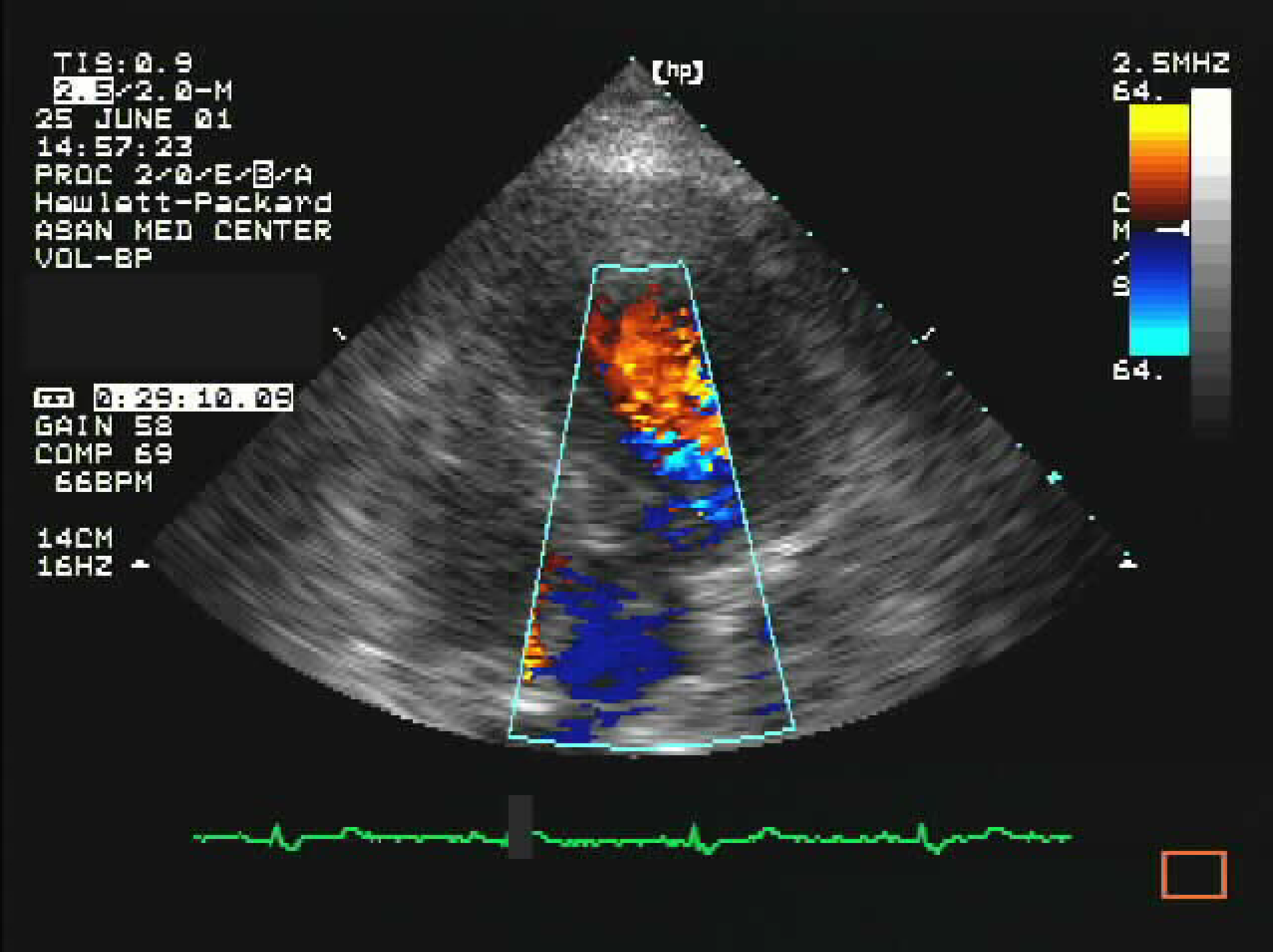
COMP 69
68BPM
13CM
13HZ
[hp]
2.5MHZ
68.
68.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Hewlett-Packard
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VOL-BP
0:29:10.05
GAIN 58
COMP 69
66BPM
14CM
16HZ
[hp]
2.5MHZ
64.
CM/S
64.

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(Severe dynamic LVOT obstruction)
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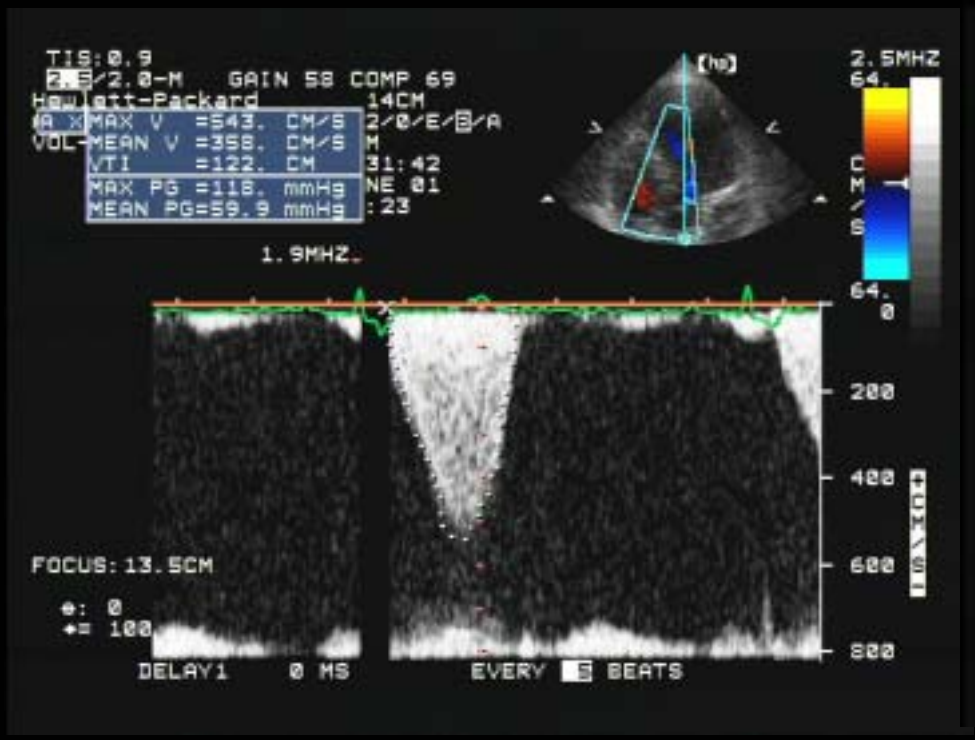
Normal CAG, PG =200 mmHg(postextrasystolic), 110 mmHg (resting)
MCE상 midseptal portion에 국한된 septal branch
(alcohol ablation, not feasible)

2001년 9월 20일 : LV septal myectomy



PG 118 mmHg

PG 11 mmHg

증례

2003년 12월 주방에서 가스 불 보다가 1-2분간 실신

2004년 3월 입원

syncope wu

TMT : BP drop during recovery

EPS : HV 65 msec

inducible pVT by RVP 250 ms - collapse with BP 50mmHg

Nonsustained pVT by dVEST

ICD on Mar 5th

2004년 10월

아침에 7시 수영 중 턴 하다가 쇽 느낌

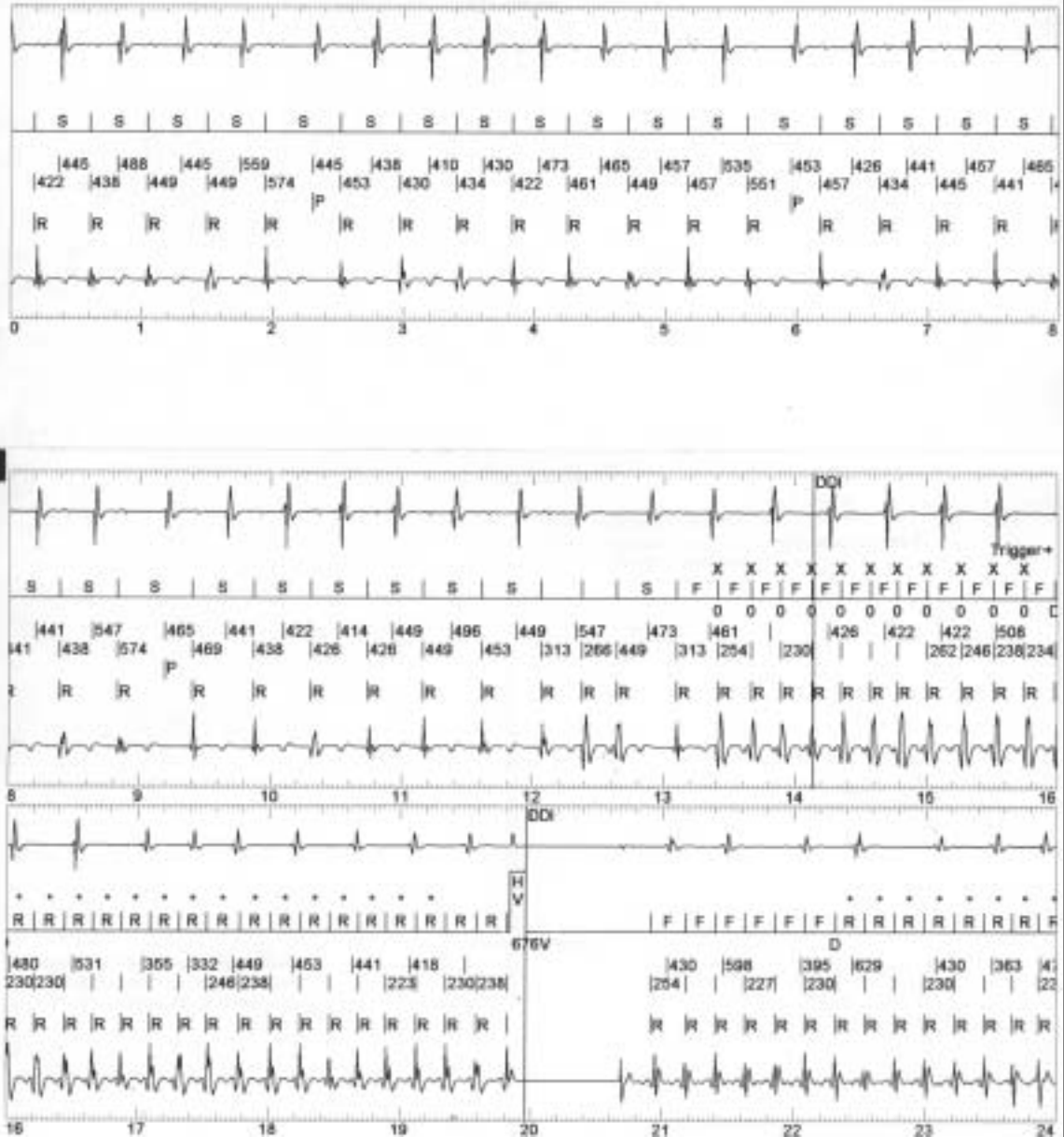
Stored EGM Report (Archive)

Patient: ChoiSoon-Ok
EGM 1: (0-24 sec)

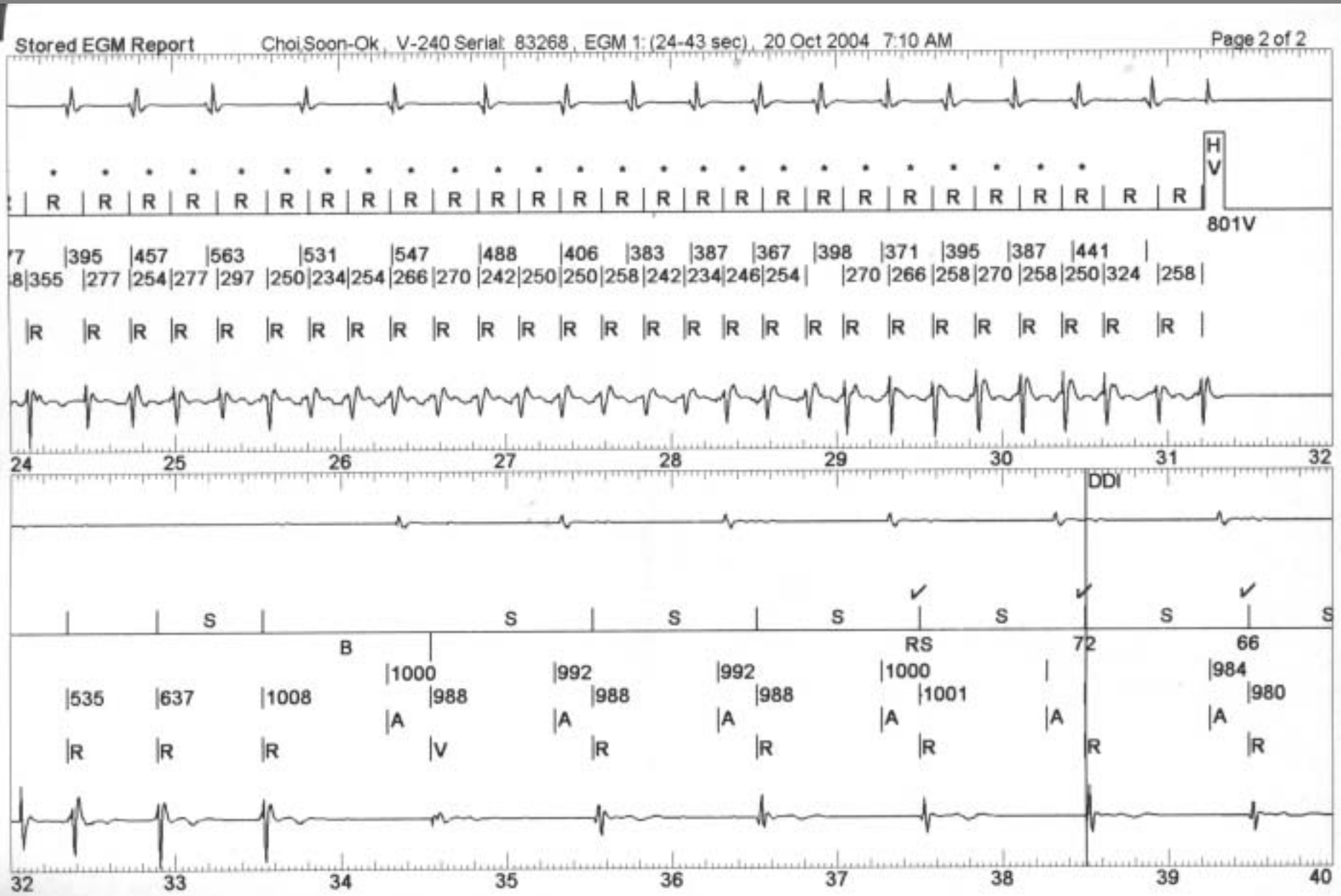
Atlas™ DR Model V-240, Serial: 83268
Summary : Fib Detection

Episode Date/Time: 20 Oct 2004 7:10 AM
Template Date/Time: 8 Oct 2004 7:57 PM

Position 1: A Sense/Pace (4.3 mV/cm, ±3.0 mV)
Position 2: V Sense/Pace (12.5 mV/cm, ±8.9 mV)
Sweep Speed: 25.0 mm/sec



ICD interrogation상 fast VT (CL 250 msec)



Hypertrophic Cardiomyopathy : overview

HCM : the most common cause of sudden cardiac death in young people (BM, Heart 2003)

Approx 60-70 % of all pt die suddenly (Otto Hess JACC 2003)

Over 60 % of cases of SCD die during or immediately after mild to moderate exertion (Heart 2002, WM)

SCD annual incidence : 2-4 % in tertiary referral center, 1% in regional population (WM, 1999, JACC)

Risk stratification : difficult due to relatively low prevalence and striking heterogeneity in clinical expression /outcome (BM, Heart 2003)

Risk stratification : Secondary prevention

1. Cardiac arrest (ventricular fibrillation)

2. Spontaneous sustained ventricular tachycardia

Risk stratification Primary prevention

1. Familial sudden HCM-related death
   (particularly in a first degree relative and/or multiple in occurrence)

2. Syncope
   (one or more episode, and particularly if recurrent, exertional, or in the young)

3. Nonsustained ventricular tachycardia (NVST) on Holter ECG
   (frequent, repetitive,or prolonged; arbitrarily defined: ≥3 bursts of NSVT at ≥120 bpm on ≥2 Holters within 6 months, or any runs ≥10 beats)

4. Abnormal blood pressure response with exercise
   (a fall or sustained failure to rise ≥20 mm Hg during exercise or recovery, in patients <50 years of age)

5. Extreme LV hypertrophy
   (maximum LV thickness ≥30 mm from echocardiogram)

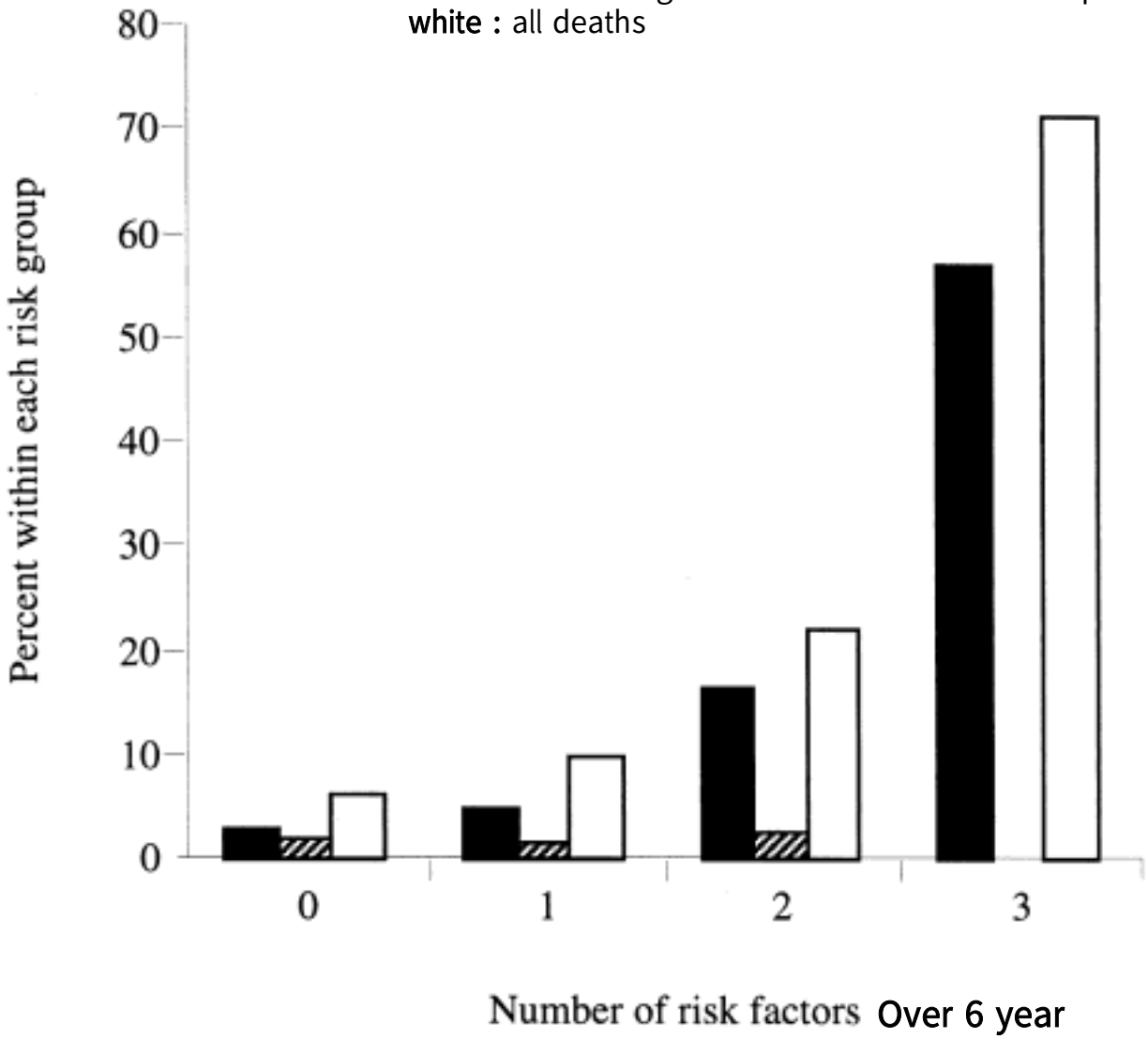
Other tests

HRV : may be useful (BM, Heart 2003)

SAECG, QT dispersion, EPS : not proven to be useful for the risk stratification

Severity of outflow obstruction, degree of functional limitation, cardiac symptoms in general did not correlated with the risk of SCD.

black bars sudden death
hatched bars congesitve cardiac failure or transplant
white : all deaths




3 Risk Factors (2%)
2 Risk Factors (10%)
45%
1 Risk Factor
(33%)
Zero Risk Factors
(55%)

Role of amiodarone

There is little evidence that prophylactic pharmacological strategies and rhythm modulating drugs effectively reduce risk for SCD. Furthermore, because of its potential toxicity, amiodarone is unlikely to be tolerated throughout the long risk periods characteristic of young HCM patients. BM JAMA 2002, 1315

BB, Verapamil, Class IA AAD (quinidine, procainamide), Amiodarone
: has not been shown to mitigate SCD risk,
cannot be tolerated over the long periods of risk incurred by young HCM pts

Role of ICD

- Aborted potentially lethal ventricular tachycarrhythmias in almost 25% of the patients over only a three year observation period. (NEJM, 2000, BM)

- ICD discharge 11 % / year for secondary prevention,
5 %/ year for primary prevention.

- Of note, ICDs often remained dormant for prolonged periods of time (up to 9 year) before discharge. (BM, 2000, NEJM)

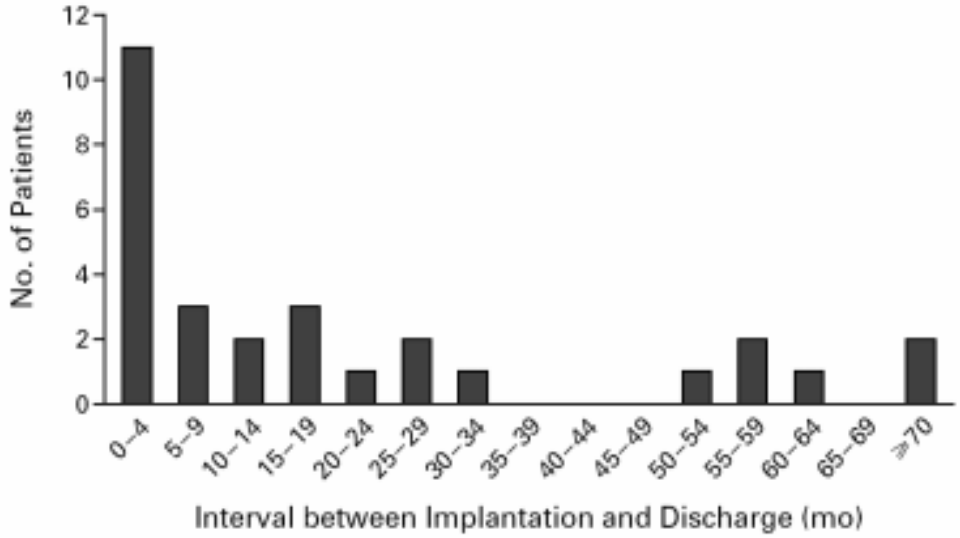


Figure 3. Interval between Implantation of the Defibrillator and the First Appropriate Discharge in 29 Patients.

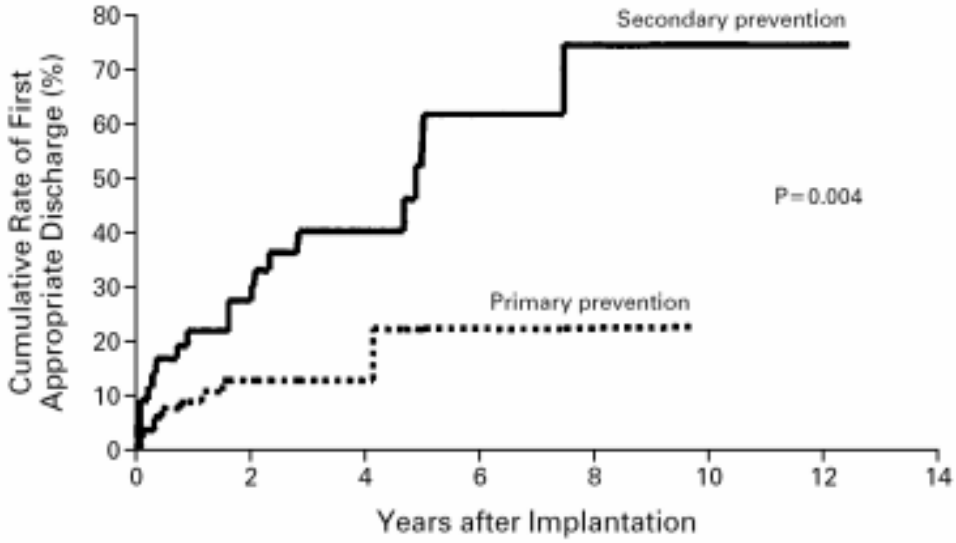


Figure 4. Estimated Cumulative Rates of First Appropriate Discharges, Calculated Separately for the 85 Patients with Defibrillators for Primary Prevention and the 43 Patients with Defibrillators for Secondary Prevention.

Risk stratification Primary prevention

1. Familial sudden HCM-related death
2. Syncope
3. Nonsustained ventricular tachycardia (NVST) on Holter ECG
4. Abnormal blood pressure response with exercise
5. Extreme LV hypertrophy

Hypertrophic Cardiomyopathy : Treatment

Outflow tract obstruction
Beta blocker
Verapamil
Dual chamber pacing
Alcohol ablation
Septal myectomy

Ventricular tachyarrhythmias
Amiodarone
ICD

Syncope

One or more episodes of unexplained LOC within 12 month (WM)

One or more episodes of syncope, particularly if recurrent, exertional or in the young (BM, Cir 2872)

Insensitive because most of patients with SCD have no prior history of syncope

Family history of SCD

Two or more SCD in the first degree relatives < 40 yo.(WM)

* statistical interaction btw FHSD and syncope (WM, 2000 JACC)
* Definition of Syncope and FHSD is arbitrary (BM, Lancet 2001, 357;1975)

FH of SCD, particularly in a first degree relative a/o multiple in occurrence (BM, Cir 2872)

Echocardiogram

Max LV wall thinkness LVWT >30mm

Young patients with extreme hypertrophy (maximal wall thickness, > 30 mm), although they usually have few or no symptoms, appear to be at substantial long-term risk. Such patients, in consideration of their youth and the potential for a near-normal life expectancy if sudden death is prevented, should be informed about the lifesaving protection afforded by the implantable cardioverter .defibrillator. (BM, NEJM)

The severity of wall thickness in isolation has insufficient predictive accuracy to guide decisions regarding prophylactic Tx.

All the individuals with LVWT >3 who died suddenly had additional risk factors, while those without other risk factors all survived.

74-82% of subgroup who died suddenly had hypertrophy of < 30 mm .

Holter monitoring

NSVT (frequent, repetitive, or prolonged)
: arbitrarily defined, ≥3 bursts at ≥120 bpm on ≥2 Holters within 6 months, or any runs ≥10 beats) (BM Cir 2872)

The association between NSVT and sudden death risk is particularly strong in patients ≤ 30 years of age
(when NSVT occurs in isolation in young patients, it may justify prophylactic therapy to prevent SCD)
(WM, JACC 2003)

BP response to exercise testing

Age <40 yo,
flat BPR, < 25 mmHg, hypotensive BPR, >15 mmHg (WM, 2000 JACC)
Abnormal BPR was of prognostic significance only in pt younger than 40 years of age. (WM, 2000 JACC)

flat, BP rise < 20 mmHg above resting,
hypotensive BPR, a continuous fall throughout exercise of > 20 mmHg or an initial increase with a subseq fall of >20 mmHg comp with peak pressure. (WM,1999 JACC)

Abnormal BP response with exercise
a fall or sustained failure to rise ≥20 mmHg during exercise or recovery, in pts ≤50 years of age (BM, Cir 2872)