Surgery 2

Updates in Hypertrophic Obstructive Cardiomyopathy

Non Surgical Management

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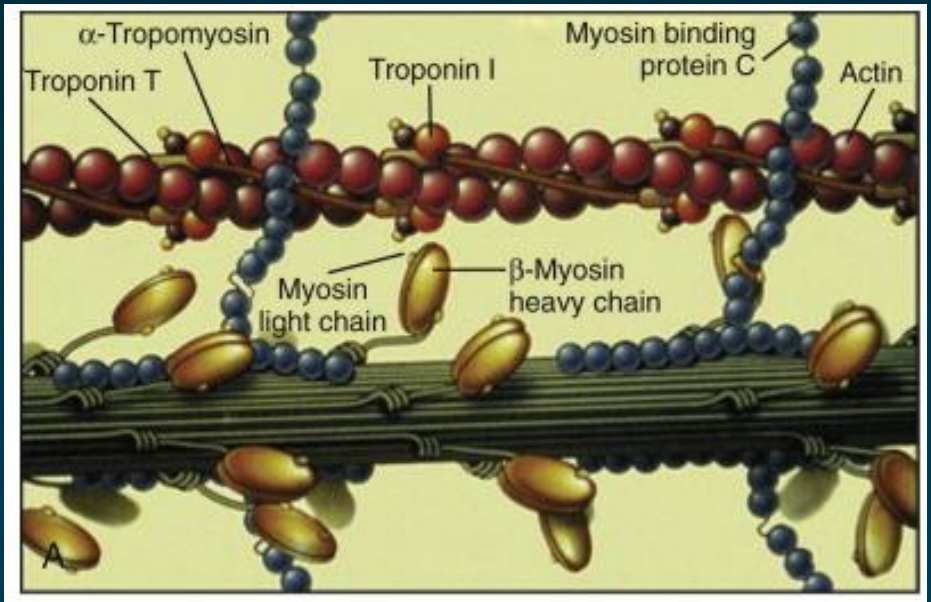
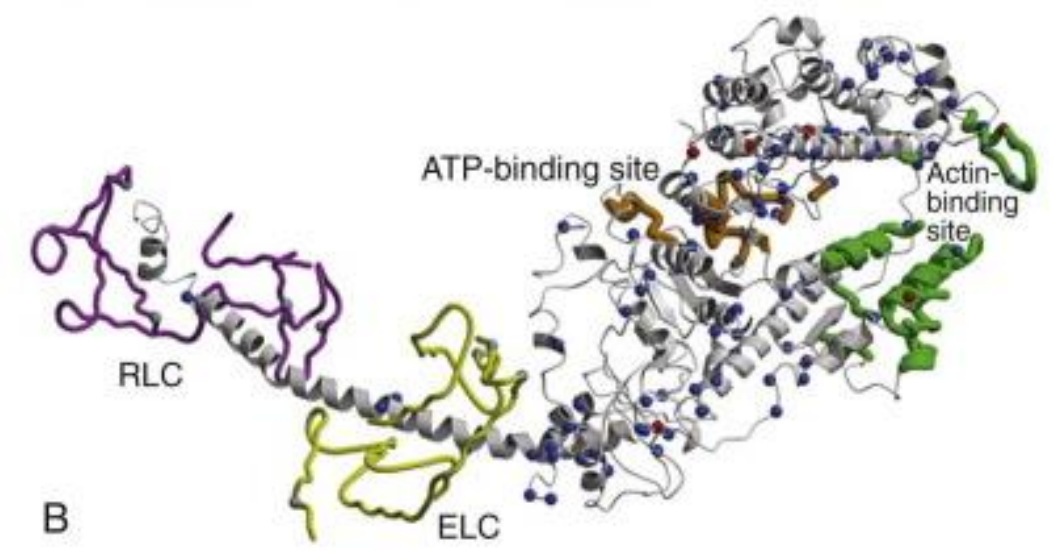
Hypertrophic Cardiomyopathy

- The Presence of increased left ventricular wall (LV) thickness that is not solely explained by abnormal loading conditions (*ESC Guideline 2014*)

Genetic Cardiac Disorder

- Cardiac Sarcomere

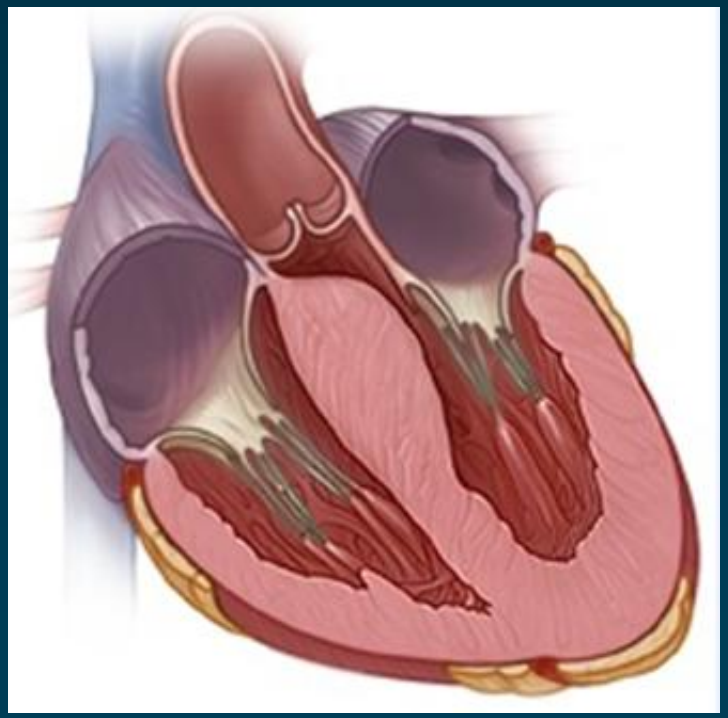




- Autosomal dominant pattern of inheritance (60%)

HCM with or without obstruction

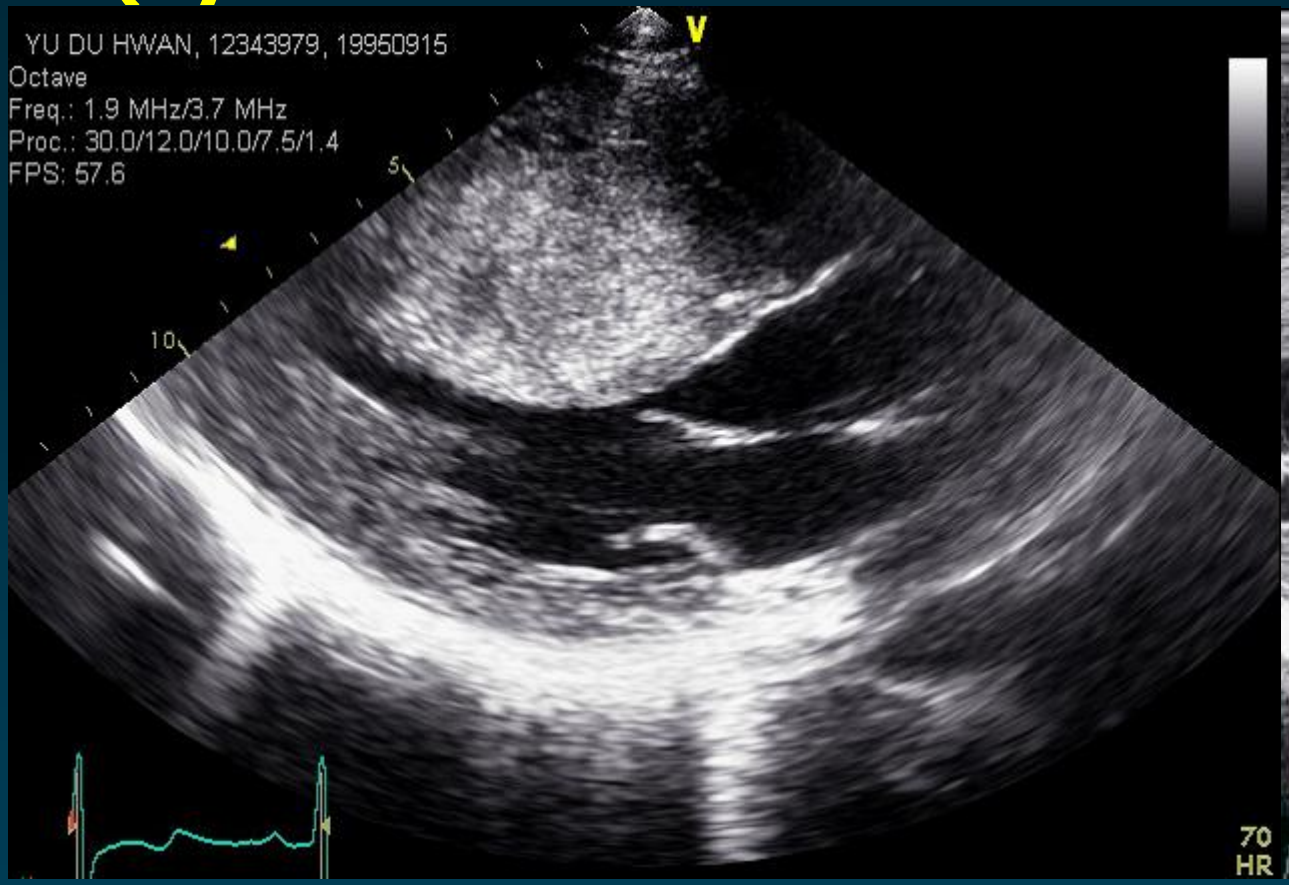
HOCM causes asymmetric hypertrophy of septum → LVOT obstruction

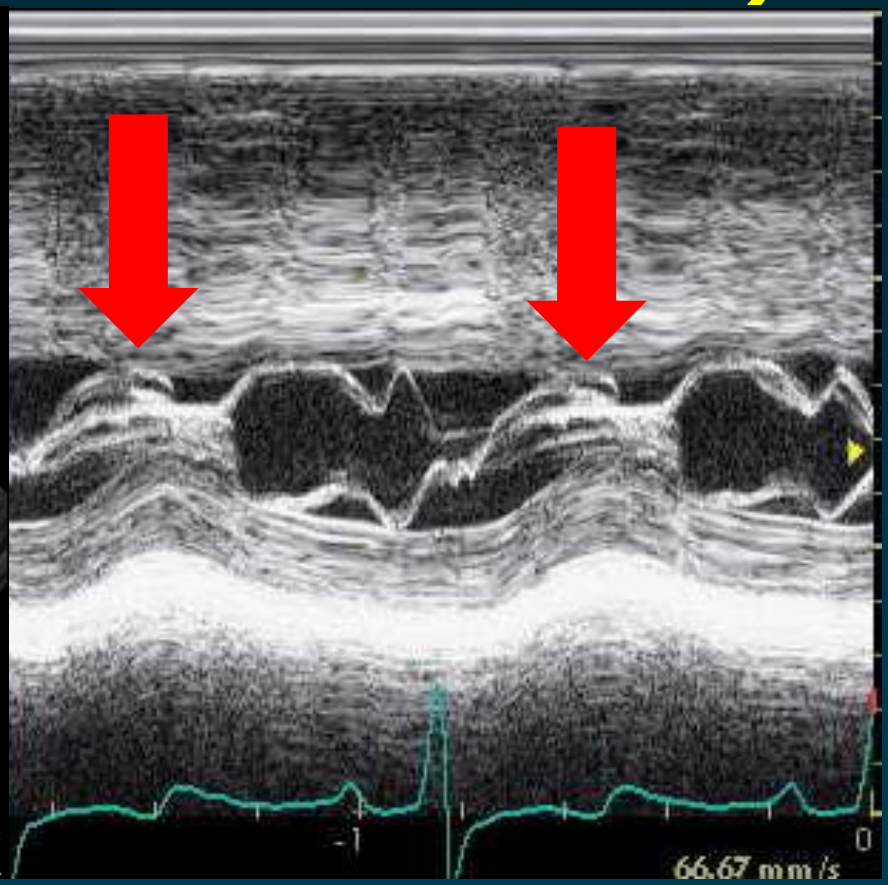


- Formally referred to as
  - Idiopathic hypertrophic subaortic stenosis (IHSS)
  - Hypertrophic obstructive cardiomyopathy (HOCM)

SAM

(systolic anterior motion of ant. mitral leaflet)





anatomic variations in MV

anterior displacement of PM

Flow Drag

(pushing force)

SAM

LVOT obstruction

LVOT obstruction

Factors that Aggravate Condition

↓ **Preload**

: hypovolemia, sepsis

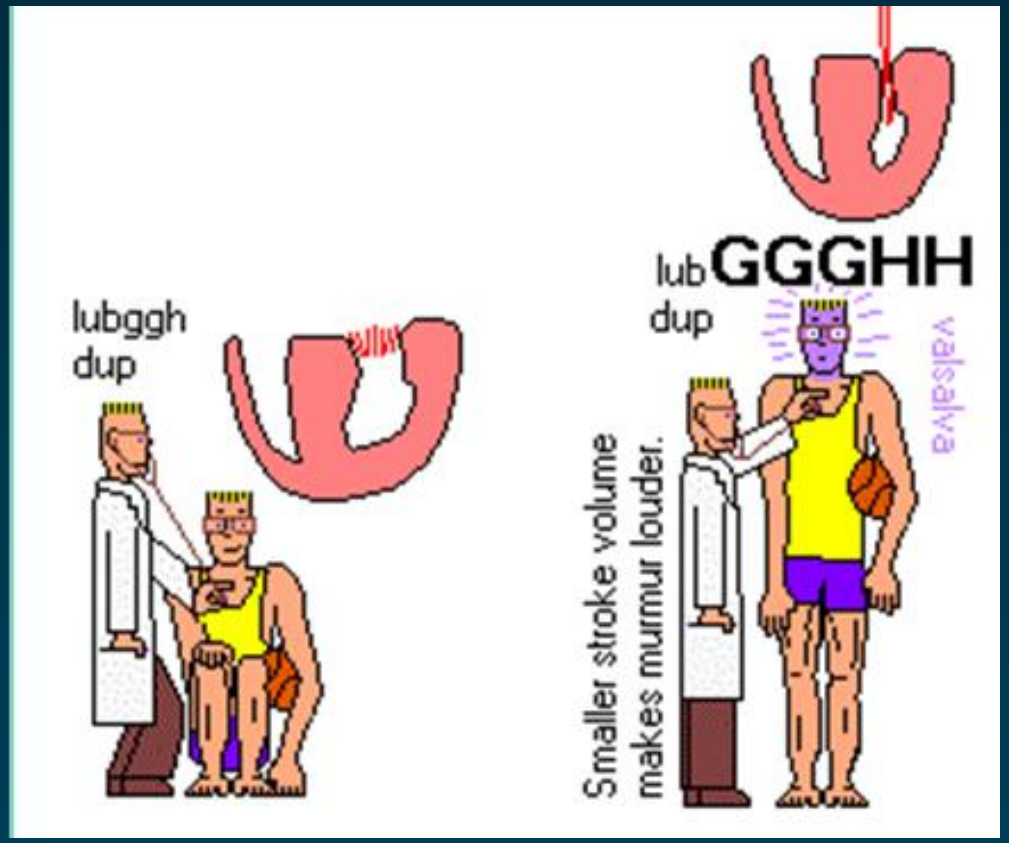
↑ **Contractility**

: exercise, positive inotropes

↑ **Heart rate**

: exercise, fever, ↑ CO

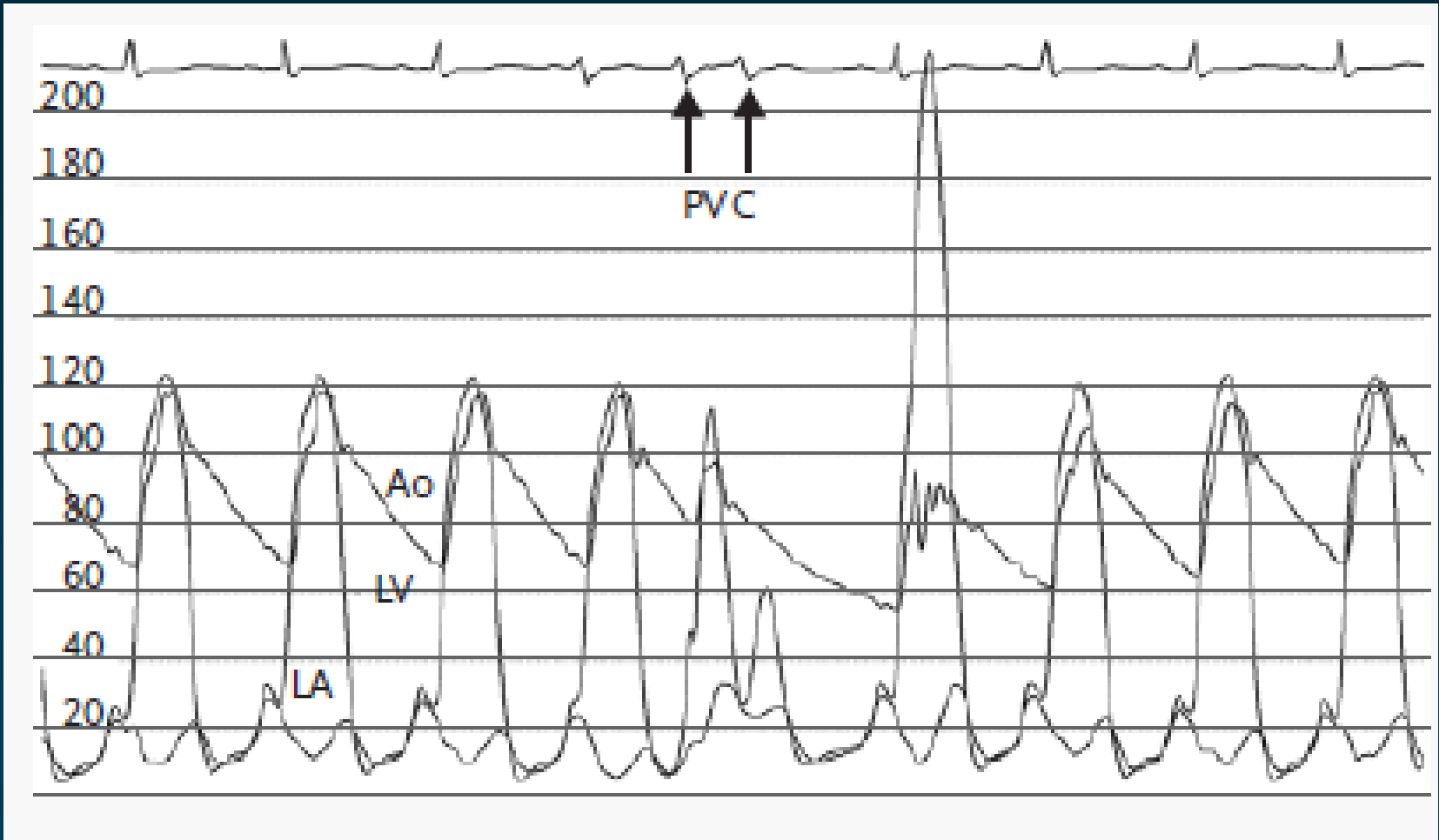
What bedside maneuvers increase intensity of HCM ?



Preload (rapid standing, valsalva) ↓

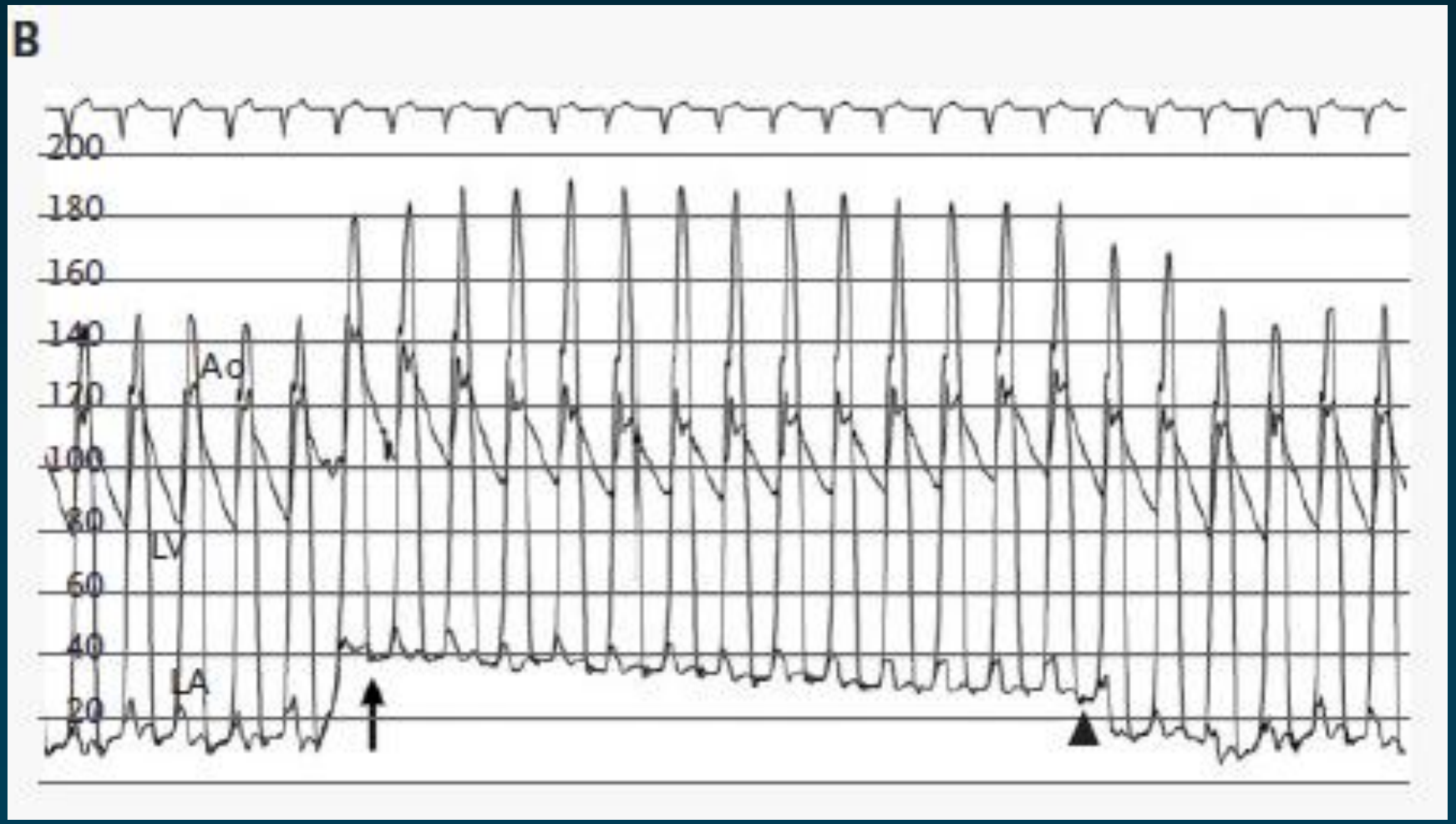
→ Less blood in heart allows septum cause LVOT obstruction

Brockenbrough phenomenon



NEJM 2004

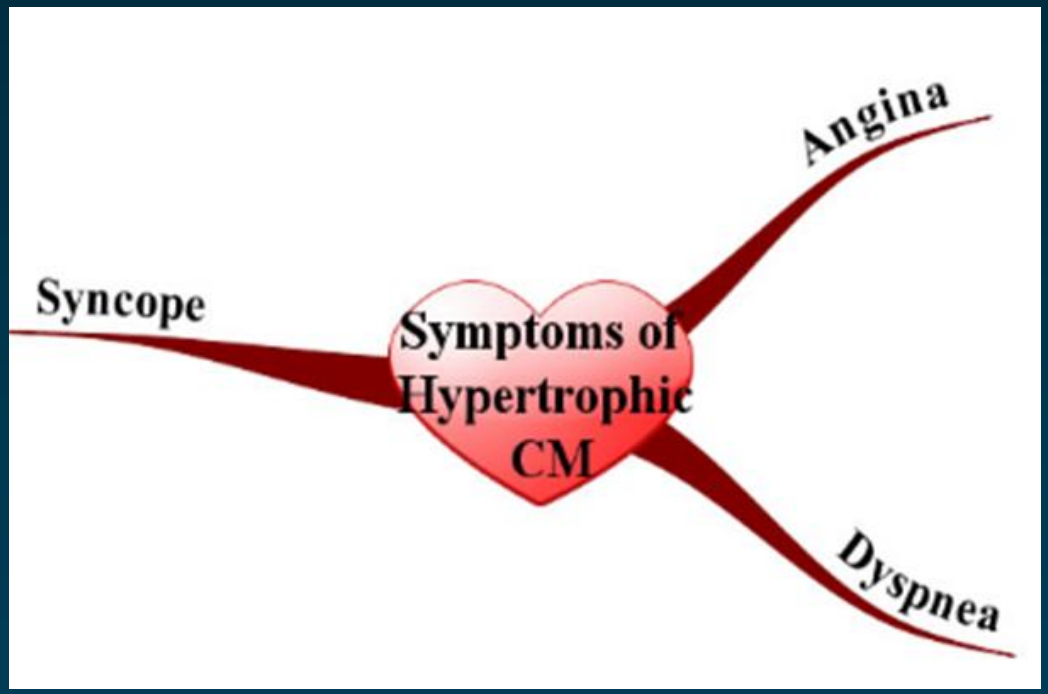
Valsalva Maneuver



Strain Phase **Release**

Symptom & Sign of HOCM

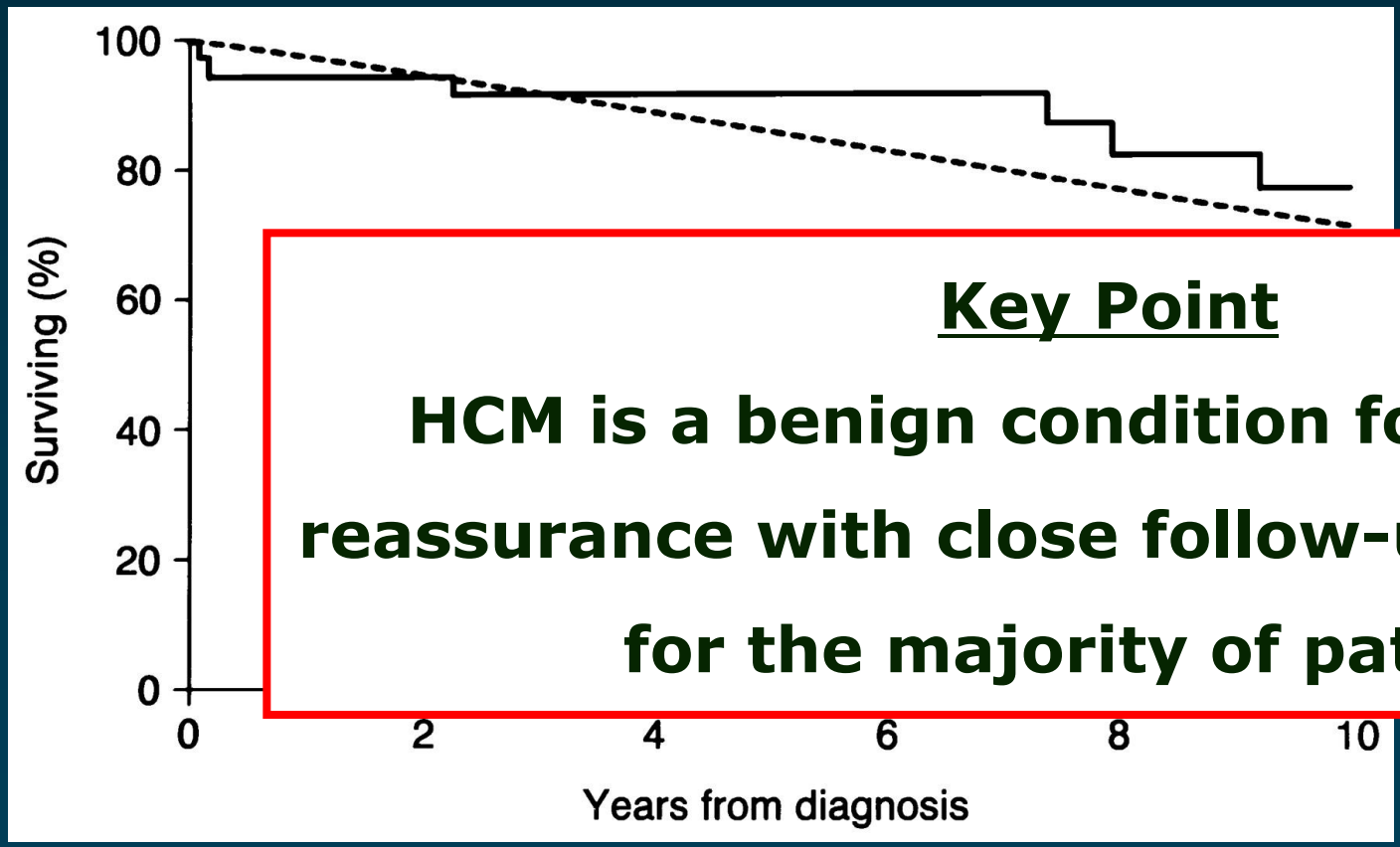
- Syncope, dyspnea, chest pain that worsens with exercise or dehydration



- Systolic murmur at left sternal border
- S4 gallop
- Sudden cardiac death due to ventricular fibrillation

HCM in the "General Population"

Natural History of HCM
Rochester Epidemiology Project : 1976 ~ 1990



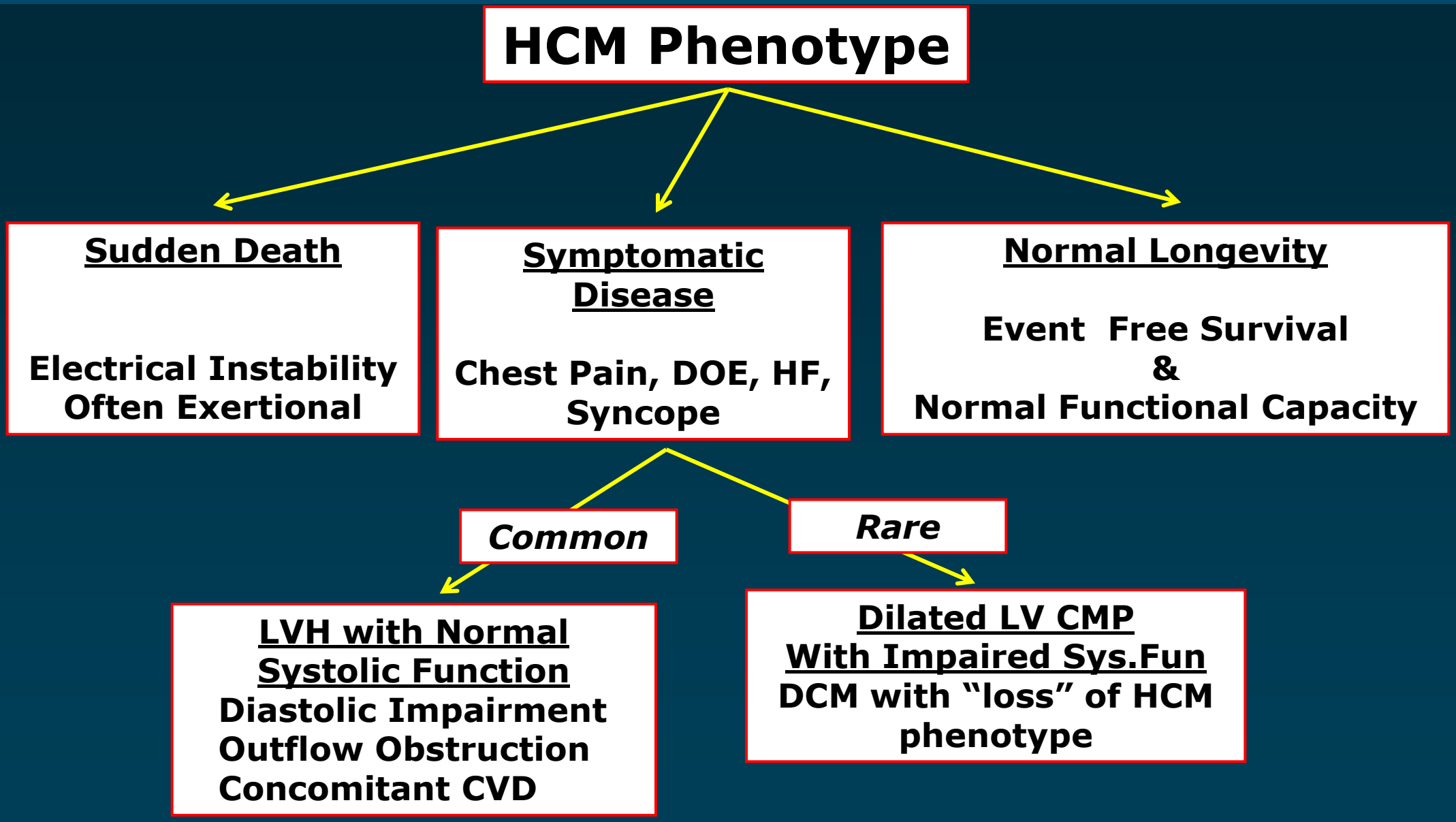
- **solid line : HCM pt**
- **dotted line : age-**

Key Point

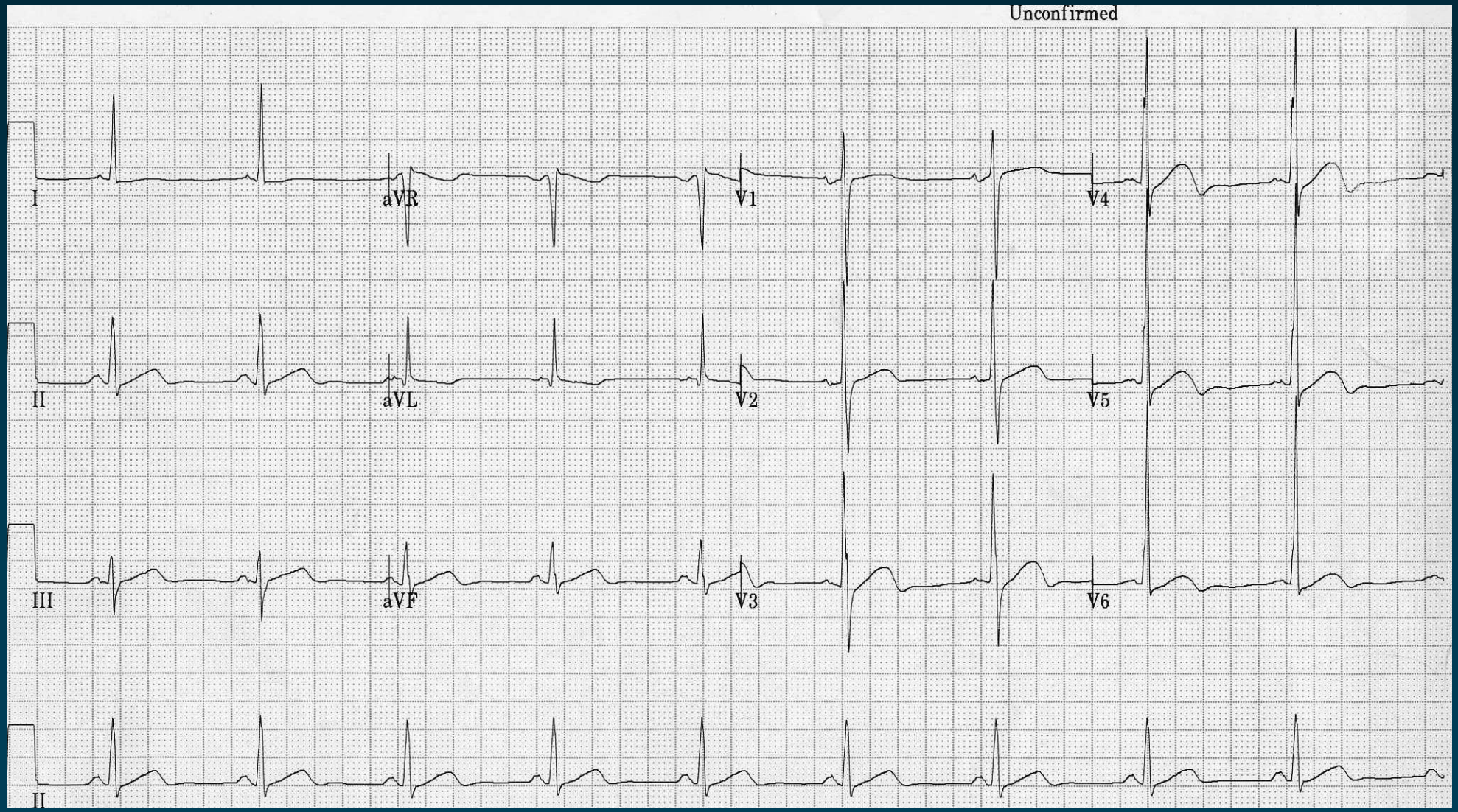
HCM is a benign condition for most and reassurance with close follow-up is sufficient for the majority of patients

Cannan CR.
Circulation 1995

HCM Disease Trajectories



Female, 65 yr (2004.5)



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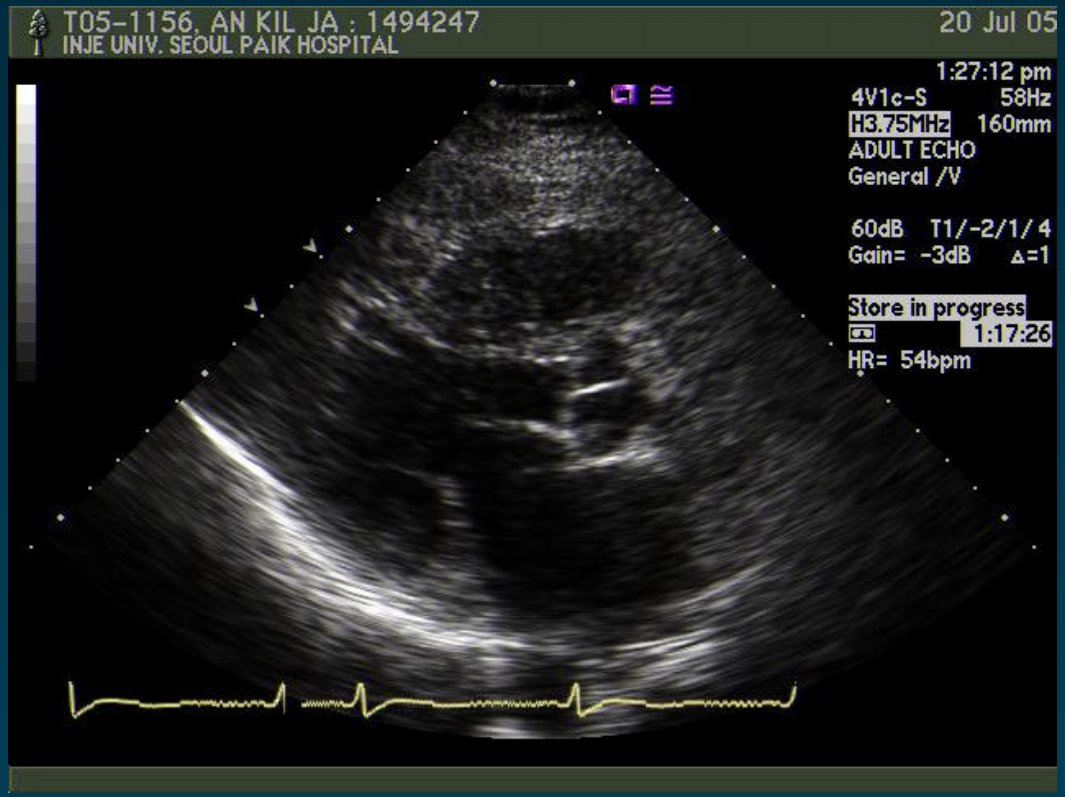
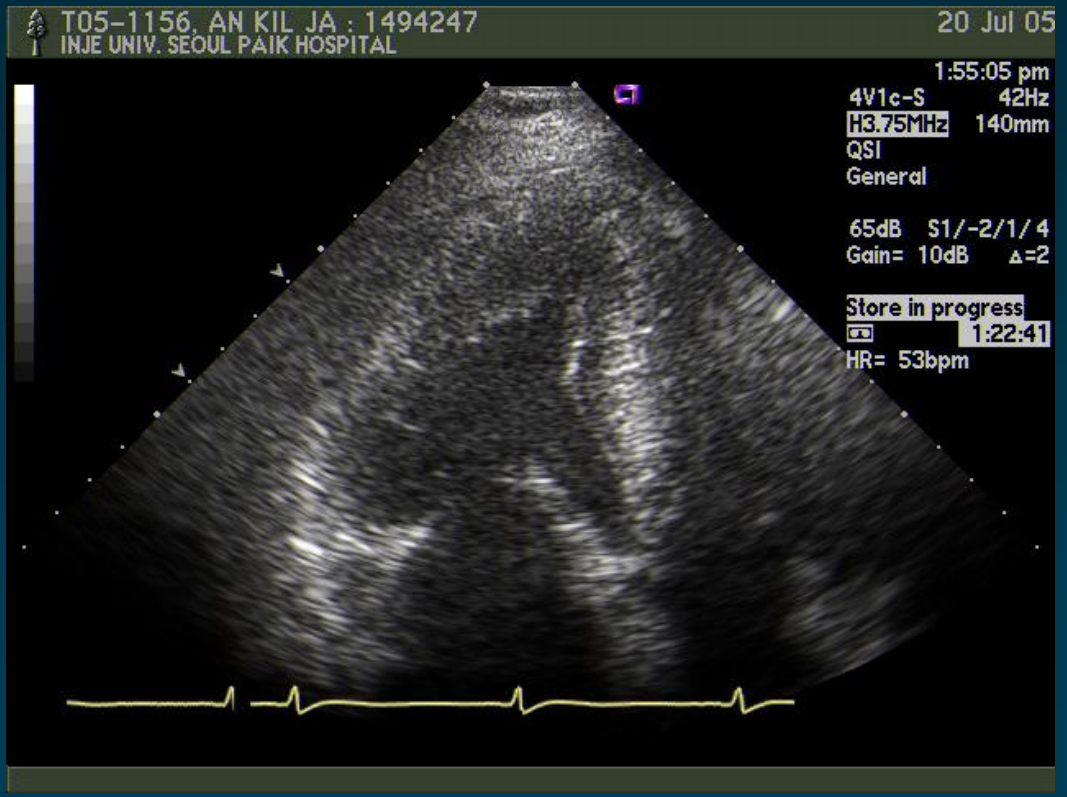
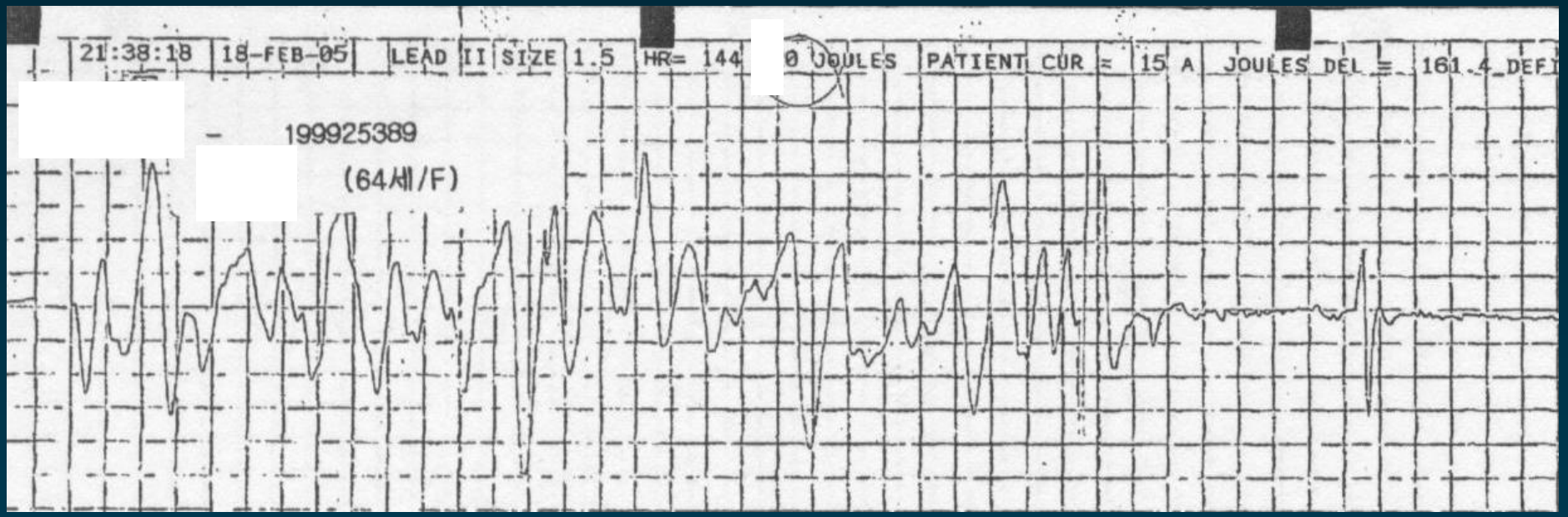
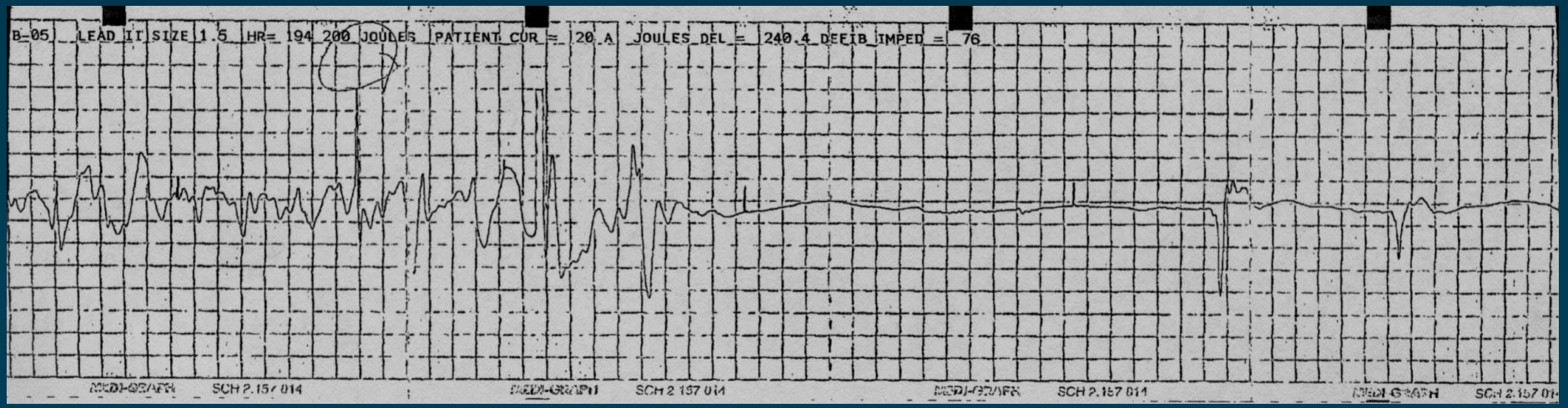




ER, 2005.2


21:38:18 18-FEB-05 LEAD II SIZE 1.5 HR= 144 0 JOULES PATIENT CUR = 15 A JOULES DEL = 161.4 DEFI
199925389
(64M/F)


B-05 LEAD II SIZE 1.5 HR= 194 200 JOULES PATIENT CUR = 20 A JOULES DEL = 240.4 DEFIB IMPED = 76
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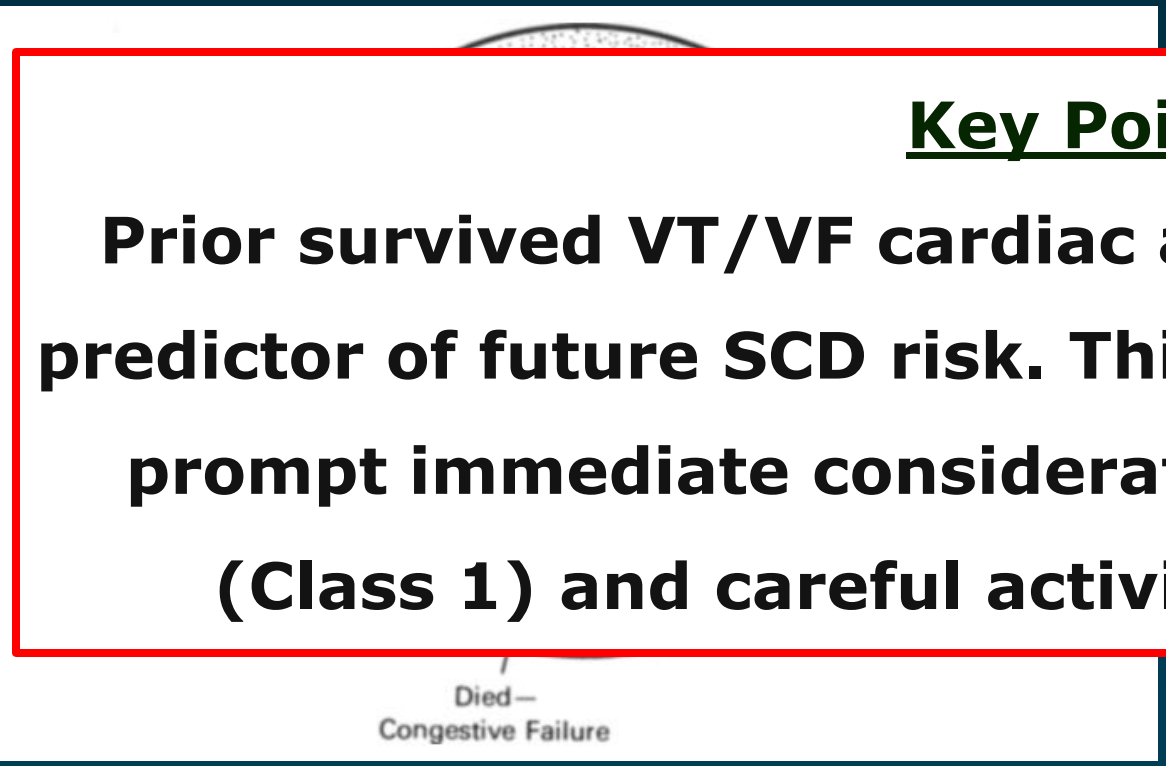
1 Prior SCD

Long-Term Outcome of Patients with HCM Successfully Resuscitated after Cardiac Arrest

Cecchi et al. JACC 1989

Survival after Cardiac Arrest or Sustained VT in Patients with HCM

Elliott et al. JACC 1999

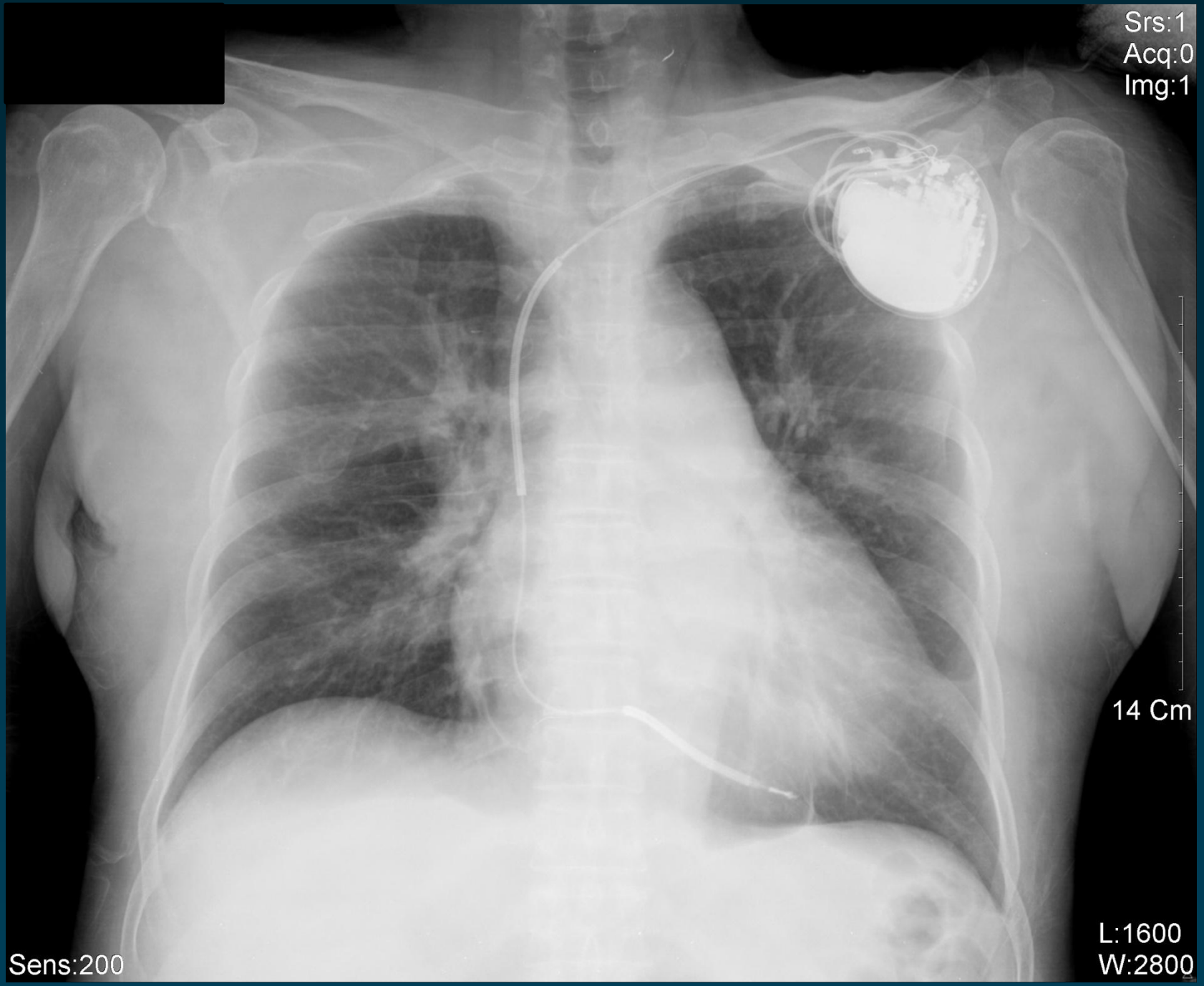


Key Point

Prior survived VT/VF cardiac arrest is our most potent predictor of future SCD risk. This historical feature should prompt immediate consideration of ICD implantation (Class 1) and careful activity recommendations

N = 33, Age =32 y, F/U = 7 yr

N = 16, Age =19 y, F/U = ~6 yr


Srs:1
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14 Cm
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W:2800

Secondary Prevention

- **Cardiac arrest due to VT or VF**
- **Spontaneous sustained VT causing syncope or hemodynamic comprise**

Life expectancy >1 year

ICD recommended

Primary Prevention
Prognostic Factors in HCM

The Big 5 (7) for SCD Risk

- Prior SCD
- Sustained VT
- Familial SD (<40 y)
- Unexplained syncope (>2/y)
- NSVT (Holter)
- Exercise hemodymics
- LV wall thickness >30 mm

- LVOT obstruction
- Myocardial fibrosis
- Exercise habits
- High-risk genetics
- Age of present
- Diastolic function
- LA dilation
- Concomitant CV Disease (CAD/AF)

Primary Prevention

Risk Factor
Age
Exercise blood pressure respon

The HCM Risk-SCD formula is as fo

$$\text{Probability}_{\text{SCD at 5 years}} = 1 - 0.998^{\text{}}$$

where Prognostic index = [0.1593985 (mm)] − [0.00294271 x maximal [0.0259082 x left atrial diameter (mm (rest/Valsalva) left ventricular outflow [0.4583082 x family history SCD] − [0.71650361 x unexplained syncope] − evaluation (years)].

HCM Risk-SCD Calculator

Age	**Years**	*Age at evaluation*
Maximum LV wall thickness	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	**mmHg**	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	○ No ○ Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	○ No ○ Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	○ No ○ Yes	*History of unexplained syncope at or prior to evaluation.*

Risk of SCD at 5 years (%):

ESC Guideline 2014

Primary Prevention

- History
- 2D/Doppler echo
- 48h ambulatory ECG

HCM Risk-SCD Variable

- Age
- Unexplained syncope
- Maximum LV wall thickness
- NSVT
- Family history of sudden cardiac death
- LVOT gradient
- LA diameter

HCM Risk – SCD Score

LOW RISK 5-year risk <4%	Intermediate RISK 5-year risk ≥4% ~ <6%	HIGH RISK 5-year risk ≥6%
ICD generally not indicated	*ICD may be considered*	*ICD should be considered*

Tx of LVOT Obstruction (general measures i)

- dehydration, excess alcohol & weight loss
- Arterial and venous dilators (nitrates and phosphodiesterase type 5 inhibitors)
- Digoxin is not recommended

Case, NEJM 1999;341(9):700

- 54-year-old man with HCM
- Echo: no LVOT gradient (rest),
  mild LVOT obstruction (inhalation of amyl nitrite)
- Tx) 40 mg of verapamil bid

Erectile dysfunction
; 2 hours after taking sidenafil and came to the ER

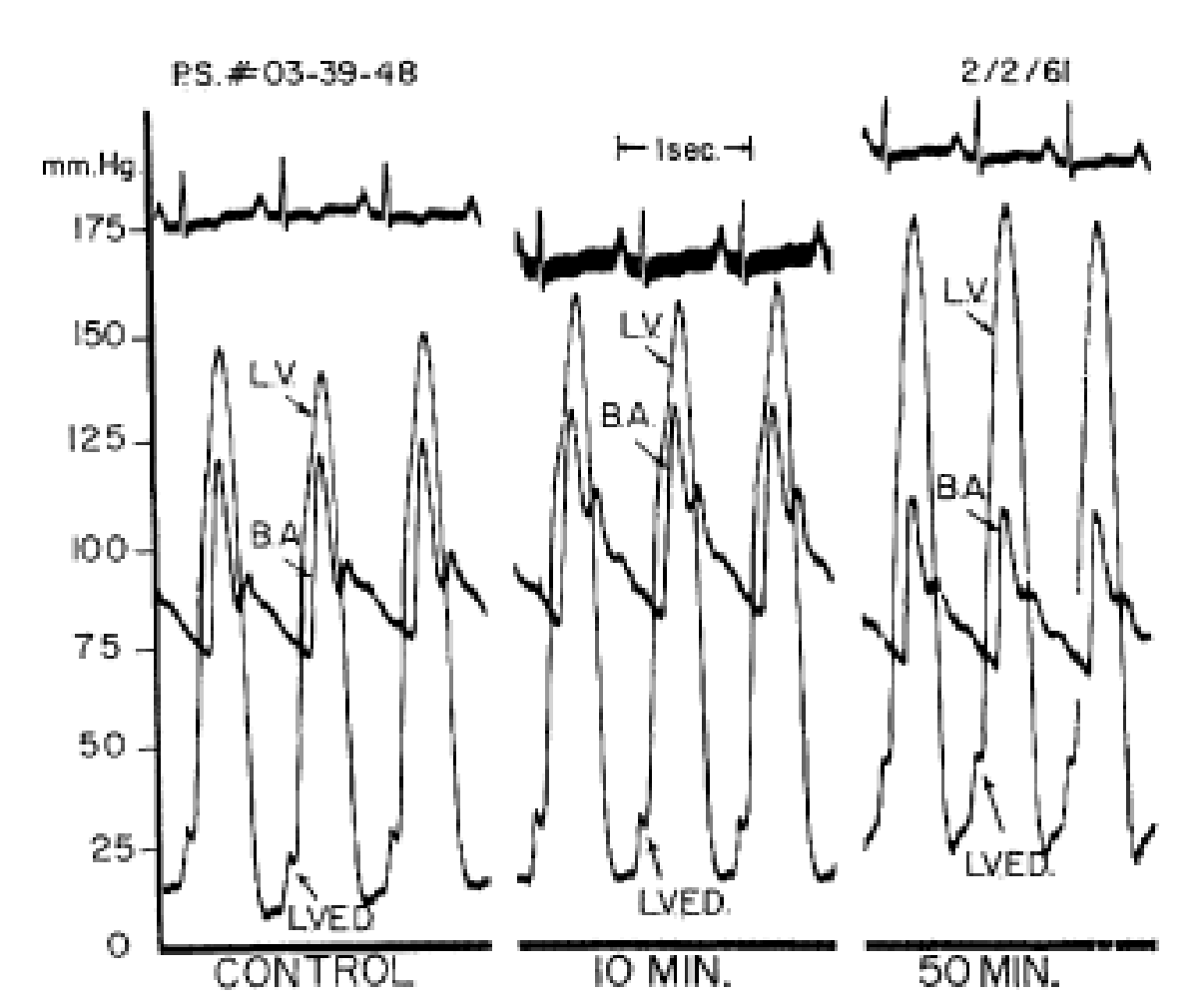
N Engl J Med. 1999 Aug 26;341(9):700-1.

Case, NEJM 1999;341(9):700

	Before Sidenafil	After Sidenafil
LVDd (mm)	47	39
LVSd (mm)	26	20
EF (%)	75	90
Subaortic gradient at rest (mmHg)	16	52
MR (grade)	1/4	2/4

N Engl J Med. 1999 Aug 26;341(9):700-1.

Hemodynamic effects of digitalization (ouabain, iv)



- LVEDP ↑
- Mean LAP ↑
- CO ↓ →
- Systolic PG between LV and brachial artery ↑

Brunwakd E, Circulation. 1962

Tx of LVOT Obstruction (general measures ii)

- Restoration of sinus rhythm or appropriate rate control should be considered before considering invasive therapies in patients with new-onset or poorly controlled AF

LVOT obstruction
Medical Treatment

- ↓ outflow tract obstruction

- Negatively inotropic drugs
  - beta-blocker
  - calcium antagonists of verapamil type
  - disopyramid
  - cibenzoline

Beta-blockers

- Drug of first choice
- ↓ LVOT obstruction (mild effect)
- Special effect in latent obstruction (low resting and high provocable gradients)

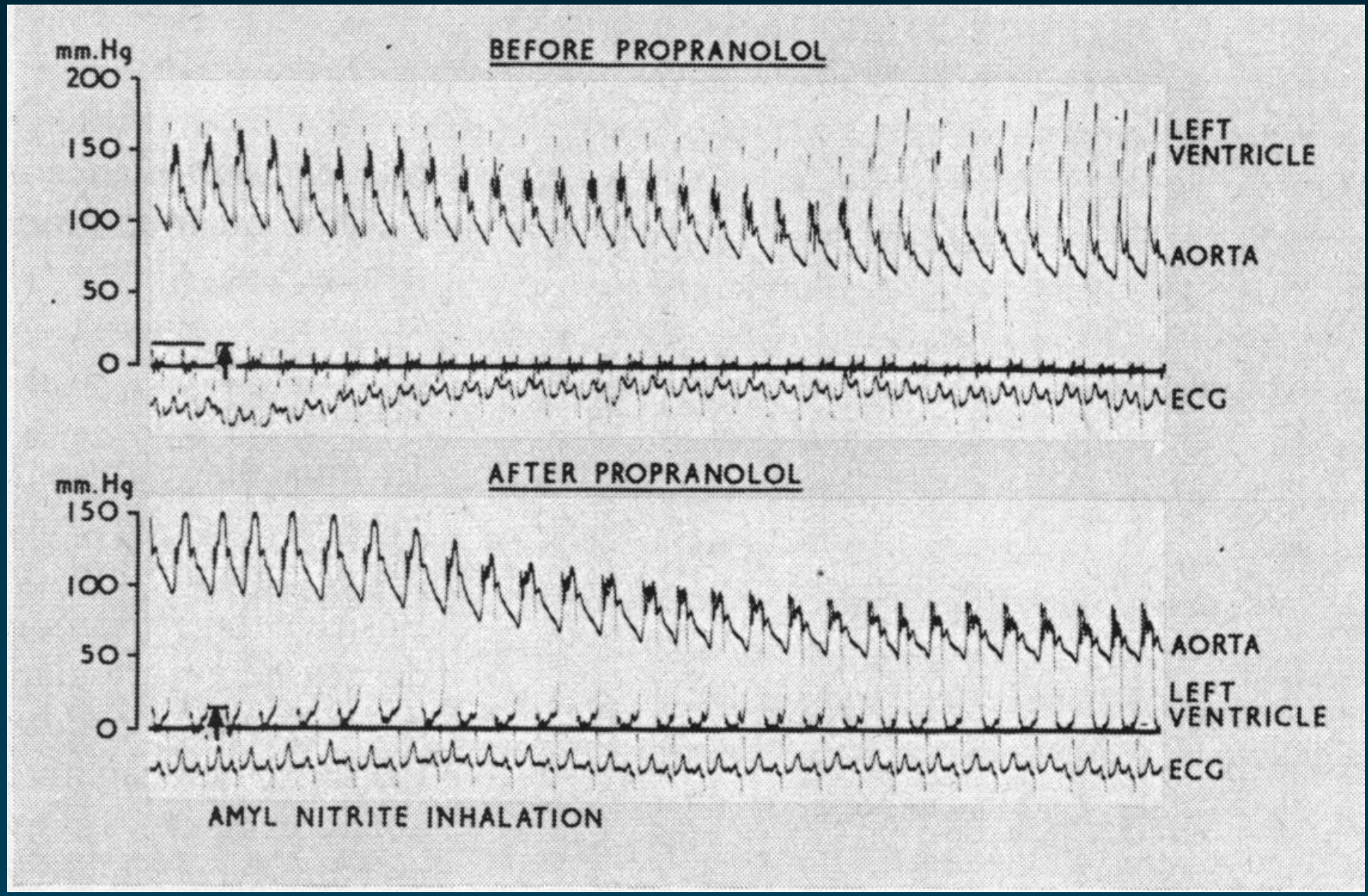
Drug	Drug Actions*			Dose			Side Effects
	Decrease Resting Gradient	Decrease Exercise Gradient	Improve Diastolic Function	Initial	Maximal	End Point of Adjustment	
Beta-blockers (e.g., atenolol, propranolol, and metoprolol)	+	+++	+	25 mg twice daily	600 mg daily	Resting heart rate <60–70 beats/min	Bradycardia, hypotension, fatigue, bronchospasm

Propranol therapy in HOCM

- Adelman AG (Toronto Unv., Canada)
- British Heart J 1970; 32: 804

- 21 patients
- Oral prorpranolol ;

  10 mg qid $\rightarrow$ 300 mg/day
- Follow-up ; 6 - 34 months



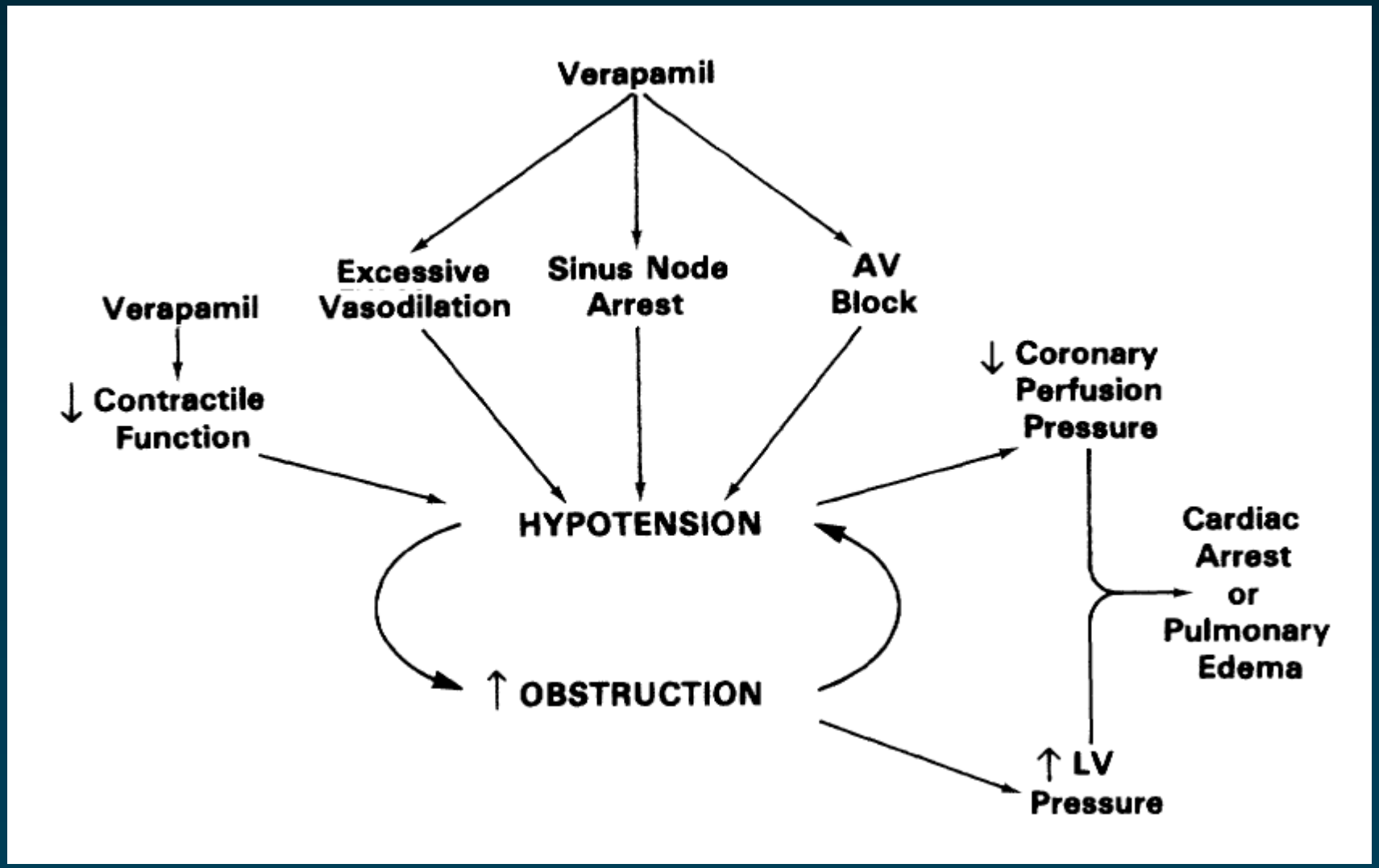
Adelman AG, British Heart J 1970; 32: 804

Calcium blockers of Verapamil type

- **Drug of first choice in Germany**
- **Outflow tract gradient reduction >50% at rest and provocation**
- **Improve diastolic function**

Drug	Drug Actions*			Dose			Side Effects
	Decrease Resting Gradient	Decrease Exercise Gradient	Improve Diastolic Function	Initial	Maximal	End Point of Adjustment	
Calcium blockers (e.g., verapamil)	+	+++	++	240 mg daily (long-acting formulation)	480 mg daily	Resting heart rate <60–70 beats/min	Bradycardia, hypotension, constipation

Calcium blockers of Verapamil type



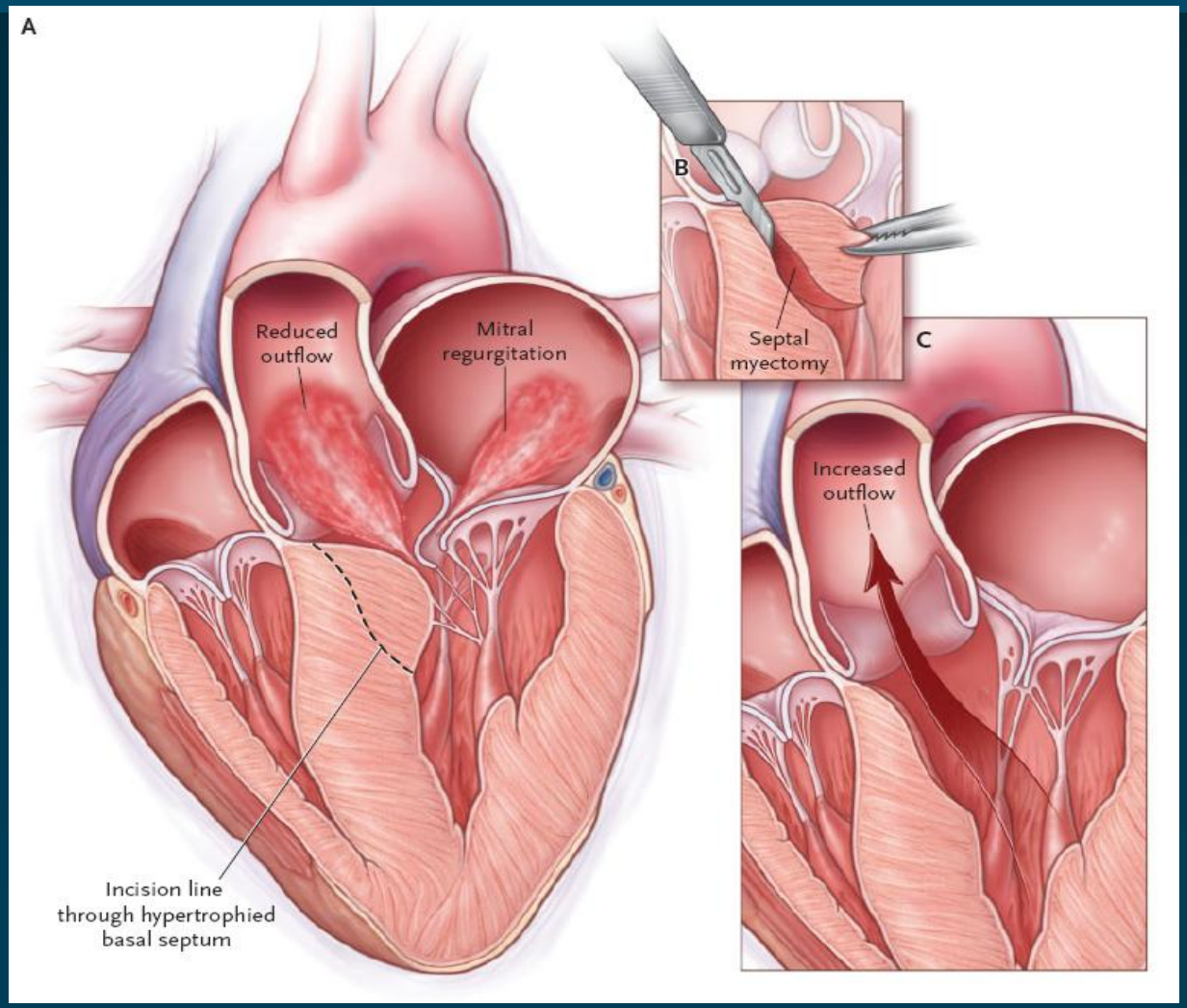
Invasive treatment of LVOT obstruction (1)

- LVOT gradient ≥50 mmHg
- Moderate ~ severe Sx (NYHA III-IV)
- Recurrent exertional syncope in spite of maximally tolerated drug therapy

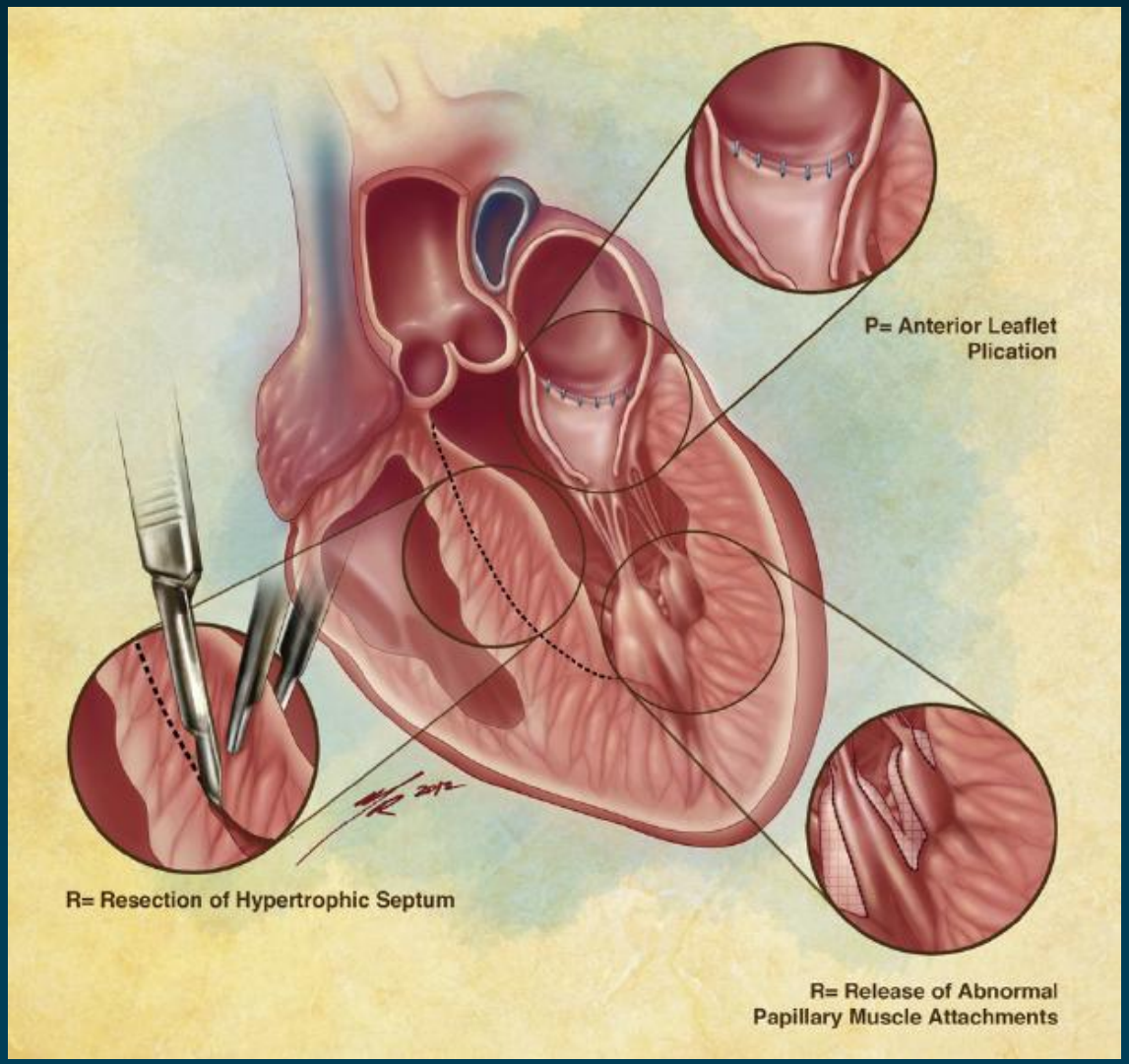
Invasive treatment of LVOT obstruction (2)

- Mild Sx + Resting or maximum provoked gradient of ≥50 mmHg (exercise or Valsalva)
- Moderate-to-severe SAM related MR
- AF
- Moderate-to-severe LA dilation

Invasive Tx
Surgical Septal Myectomy

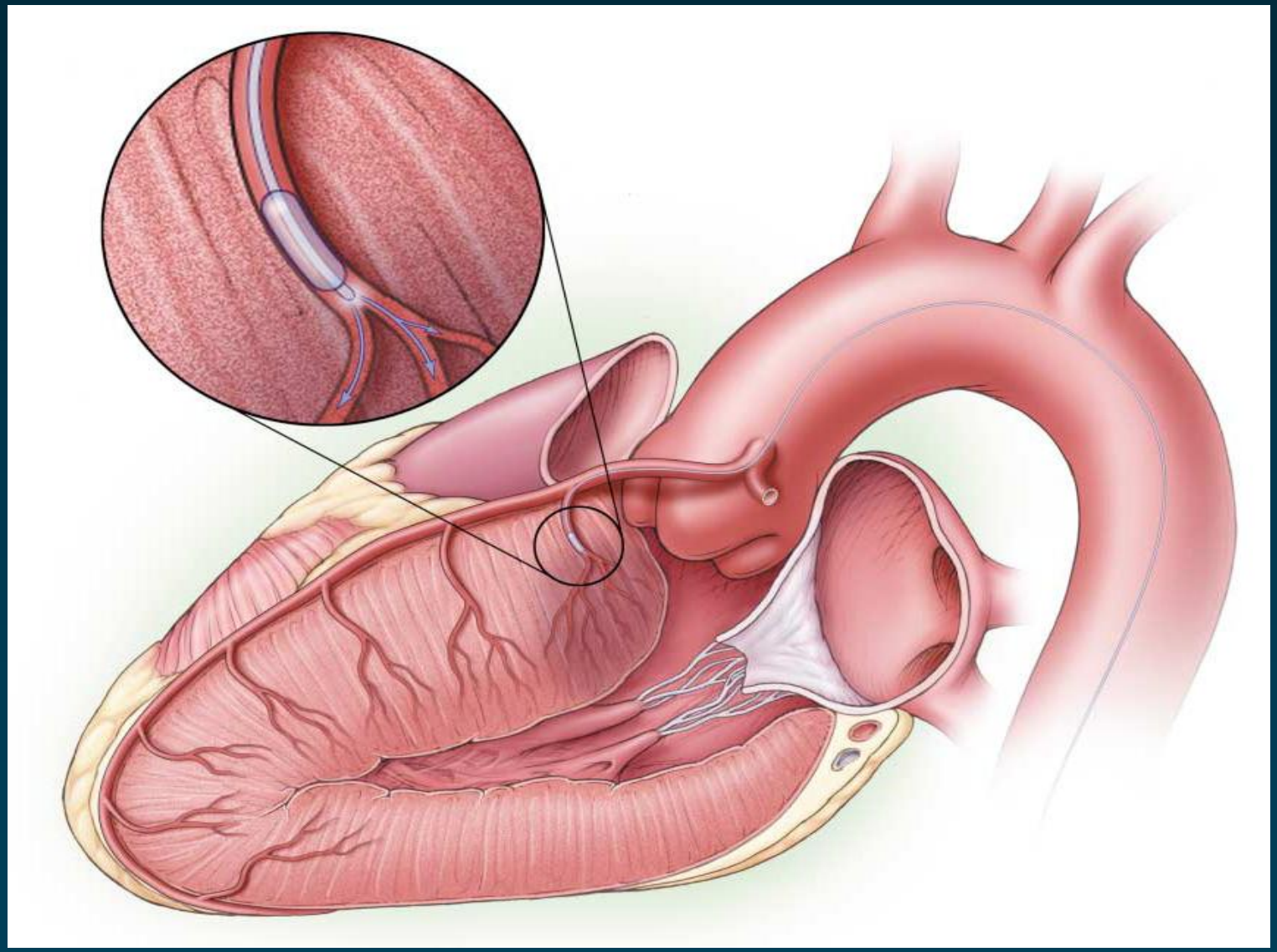


Resection-Plication-Release (RPR) Repair for HCM



Swistel DG.
Prog Cardiovasc Dis
2012

Invasive Tx :
Alcohol-induced septal infarction



Septal Myectomy

- Age <55 years (and children)
- Obstruction due at least in part to anomalies of submitral apparatus
- Intrinsic MV disease (severe MR)
- Acute gradient reduction required
- Presence of coexisting disease: CAD, fixed AS, AF (for MAZE)
- Particularly high gradients and extreme LVH
- Coronary anatomy not amenable to ablation
- Previous alcohol ablation unsuccessful

Alcohol Septal Ablation

- Age ≥55 years
- Unfavorable surgical candidate with significant co-morbidity
- Patient unwillingness to undergo surgery
- No access to surgical center

Dual Chamber Pacing

- Not candidate for either myectomy or alcohol ablation

LVOT Obstruction
Non Surgical Management

- General managements
- Drug therapy
- Invasive treatment

Conclusion

- Tx of arrhythmias
- Reduce the extent of the LVOT obstruction
- Improve functional disability
- Improve diastolic filling
- Prophylaxis of endocarditis


경청해 주셔서
감사합니다.