Surgical and Procedural Treatment of HOCM

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Disclosures

Nothing to disclose.

Overview

Diagnosis

Echo

Sudden Death/HCM vs HOCM

Genetics

Alcohol septal ablation

Guidelines

Myomectomy / Surgical Considerations

Papillary muscles

Mitral valve

Myomectomy outcomes

Diagnosis

Hypertrophic cardiomyopathy: Symptoms

- ***May be asymptomatic***
  - ***Found on screening ECG, echocardiogram***
- **Dyspnea**: High LV diastolic pressure because of impaired LV relaxation and increased stiffness. High afterload if LVOT obstruction
- **Angina**: Inadequate coronary arterial blood supply for degree of hypertrophy and increased demand
- **Syncope**: Arrhythmias or hypotension secondary to LVOT obstruction
- **Palpitations**—AF is common
- **Sudden cardiac death**: Ventricular fibrillation[6]

HOCM: Echo findings

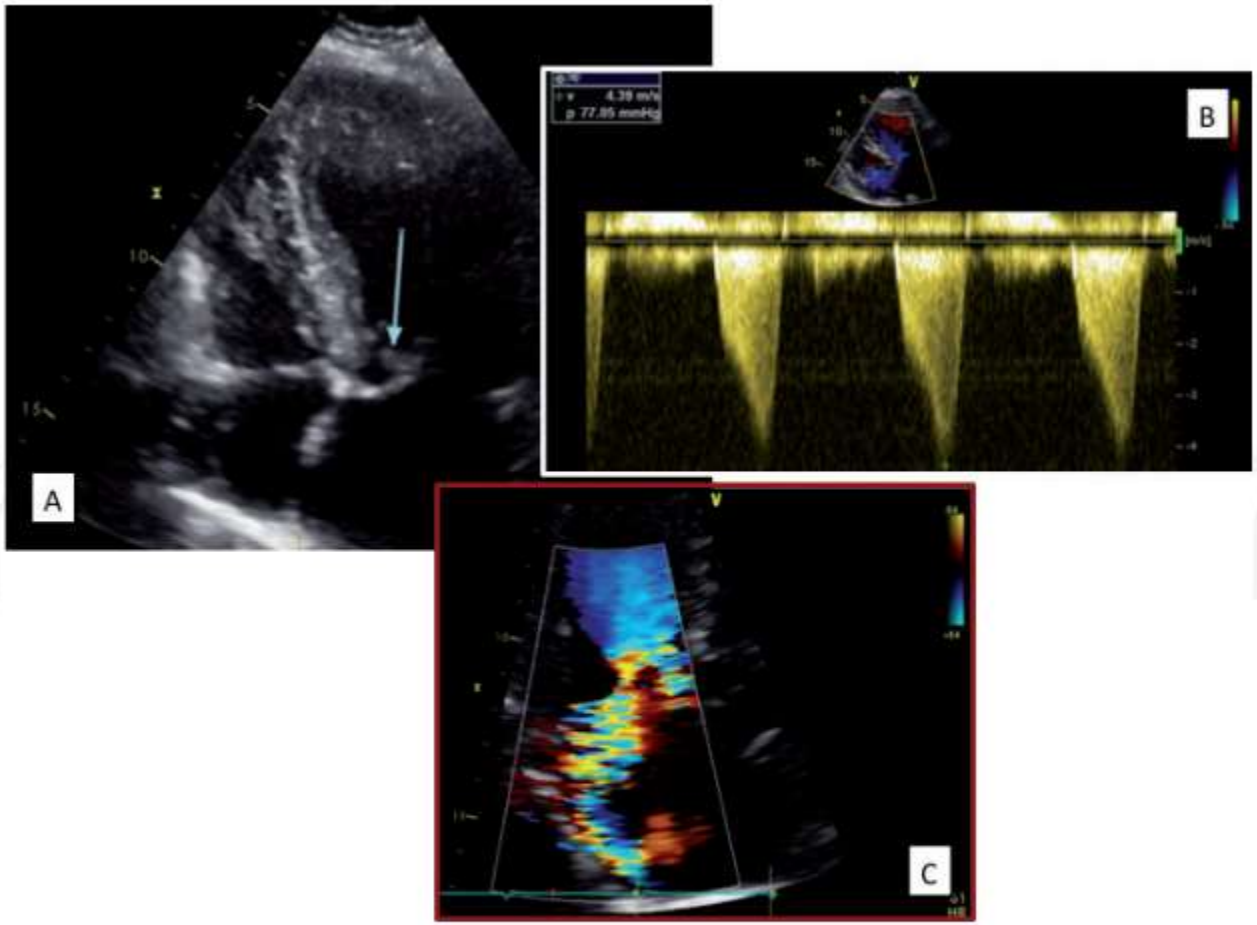
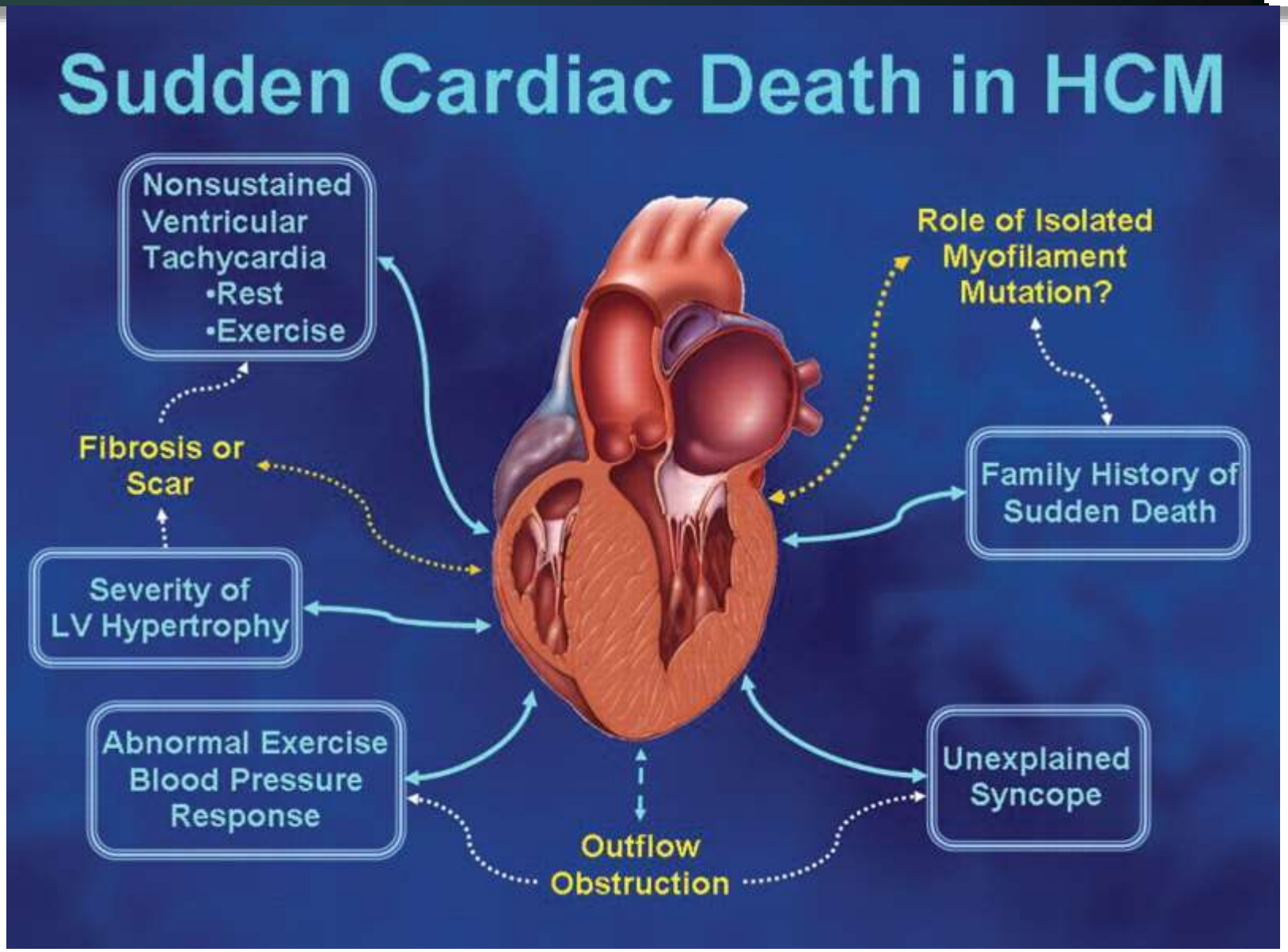


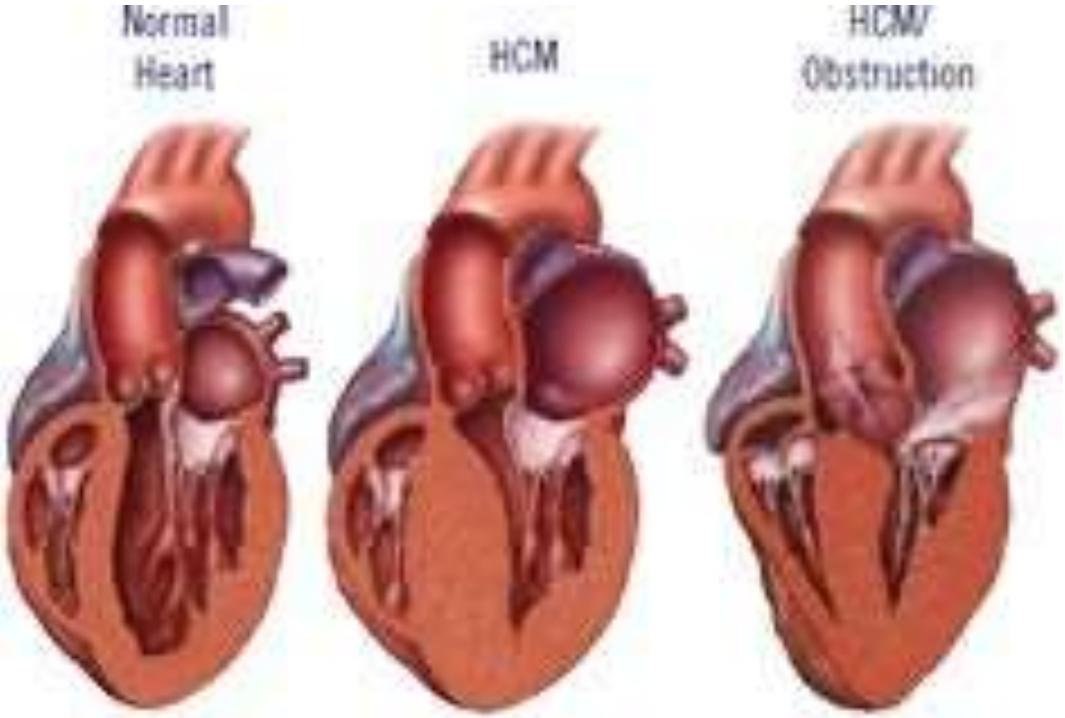
Figure 1.
Transthoracic echocardiography showing LVOT obstruction (A), with a resting LVOT gradient of 77 mmHg (B) and severe MR, due to SAM (C). Figure courtesy of non-invasive cardiology diagnostic laboratory, Cardiology Department, L. Sacco Hospital, Via G.B. Grassi 74, Milano, Italy.


Sudden Cardiac Death in HCM
Nonsustained Ventricular Tachycardia
•Rest
•Exercise
Role of Isolated Myofilament Mutation?
Fibrosis or Scar
Family History of Sudden Death
Severity of LV Hypertrophy
Abnormal Exercise Blood Pressure Response
Unexplained Syncope
Outflow Obstruction

Epidemiology

- Hypertrophic obstructive cardiomyopathy occurs in men and women with equal distribution across all races.
- Females tend to be more symptomatic causing more disability and present at a younger age.
- A 1995 study by Maron et. Al. concluded that it occurs at a rate of 1:500 in the general adult population.
- In the United States alone, there are fewer than 100 deaths per year due to HOCM at a rate of 1:220,000 athletes.
- Most patients present in the second/third decade of life. However, some adults may present between the fourth and sixth decades of life.

HCM vs HOCM

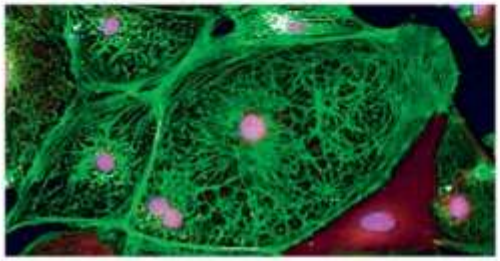


Note the increased left ventricular (LV) muscle wall thickness, decreased LV lumen size, and enlarged left atrium (LA) of the hearts with HCM.

Genetics/Pathophsyiology

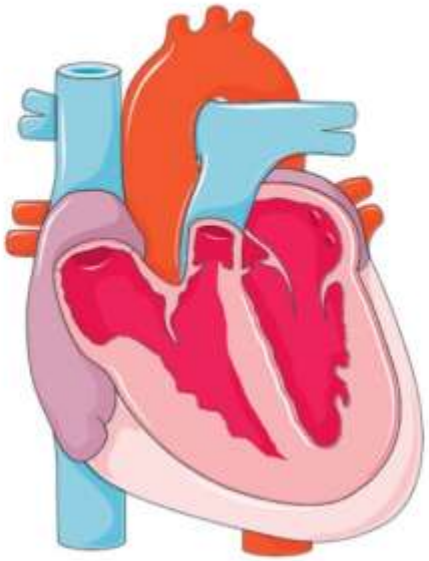
Pathophysiology:

- Increased LV wall thickness without abnormal loading conditions
- Interstitial fibrosis
- Arrhythmias
- Cardiomyocyte hypertrophy and disarray



Disease progression

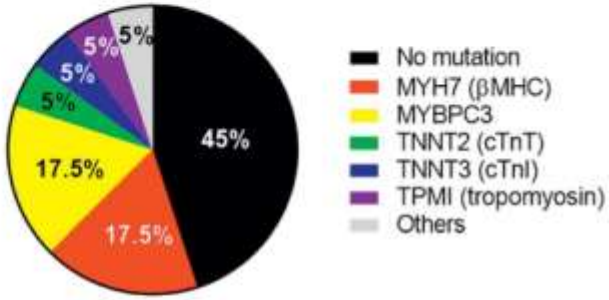
- Compensatory response
  - Hypertrophy
  - Initiation of fetal gene program
  - Metabolic shifts (phosphocreatine→fatty acids →glucose)
  - Fibrosis (interstitial and perivascular)
- End-stage heart failure: energy and functional imbalance



Hypertrophic cardiomyopathy

Genetics:

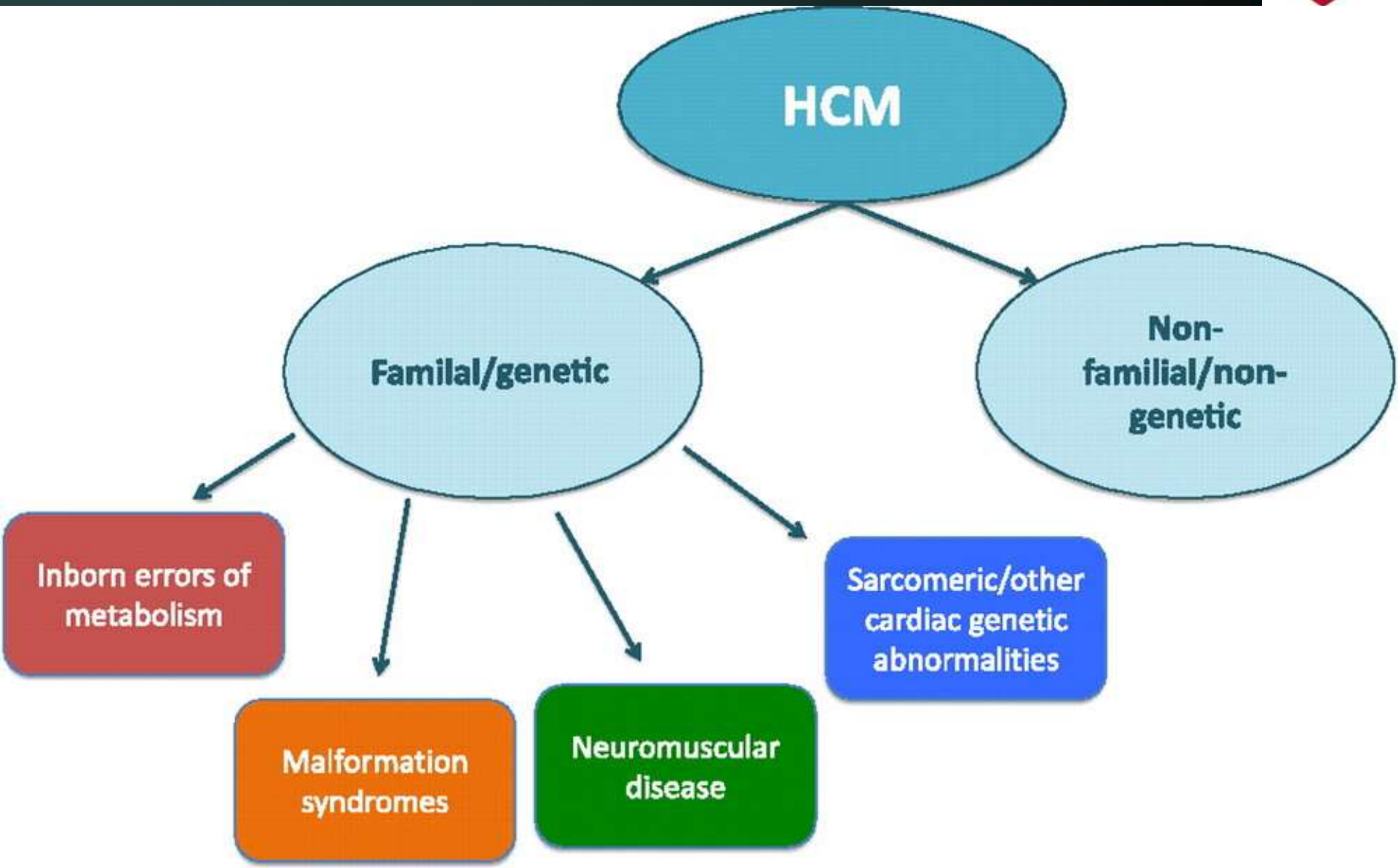
- Approximately 50% of patients have mutations in one or more of >20 sarcomeric genes
- Complex genetic causation (variable penetrance and expressivity):



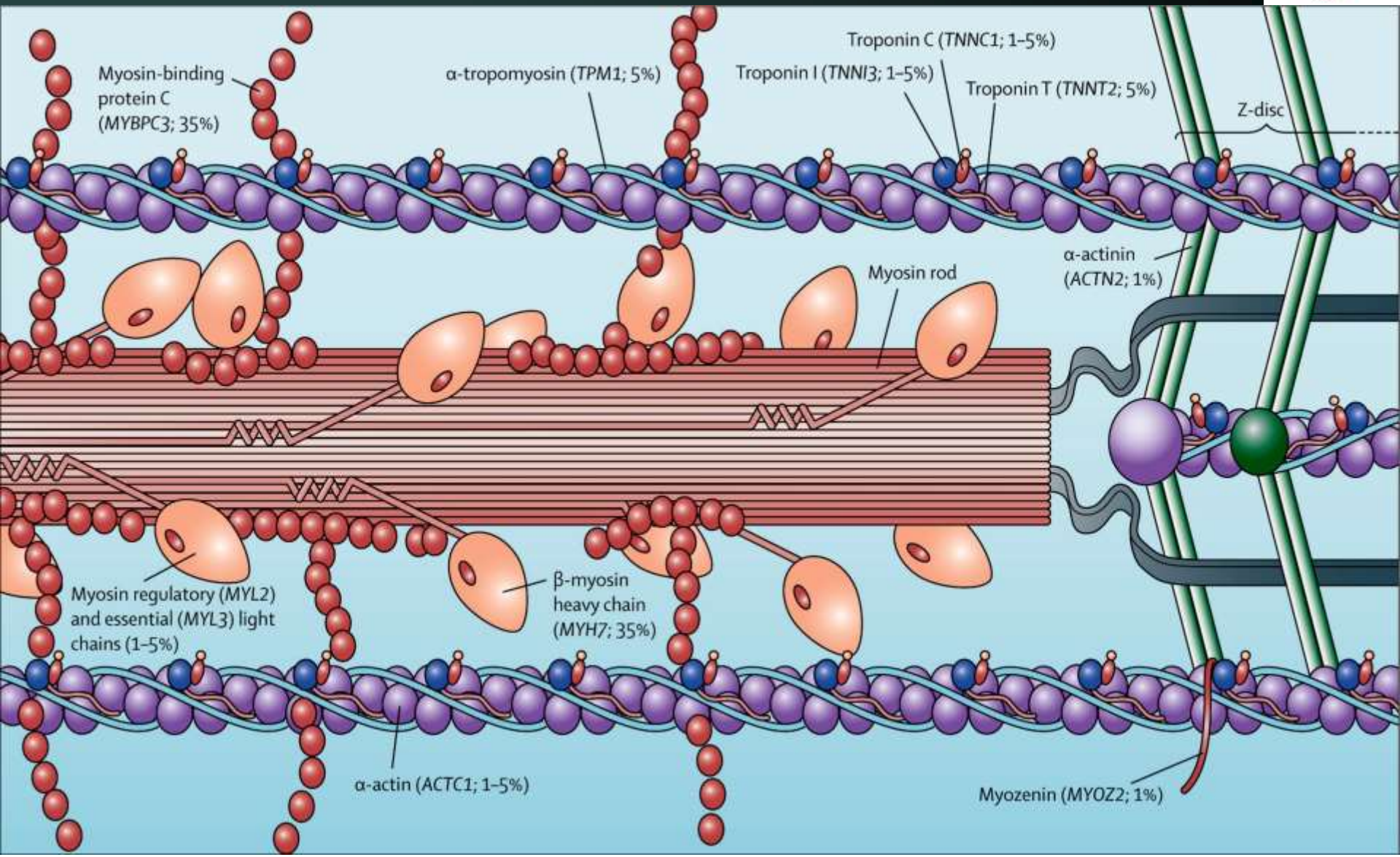
Diagnosis: increased LV wall thickness

- Echocardiography
- Magnetic resonance imaging (e.g., late gadolinium enhancement)
- Nuclear imaging
- Computerized tomography
- Genetic screening

Trends in Molecular Medicine


HCM
Familal/genetic
Non-familial/non-genetic
Inborn errors of metabolism
Malformation syndromes
Neuromuscular disease
Sarcomeric/other cardiac genetic abnormalities

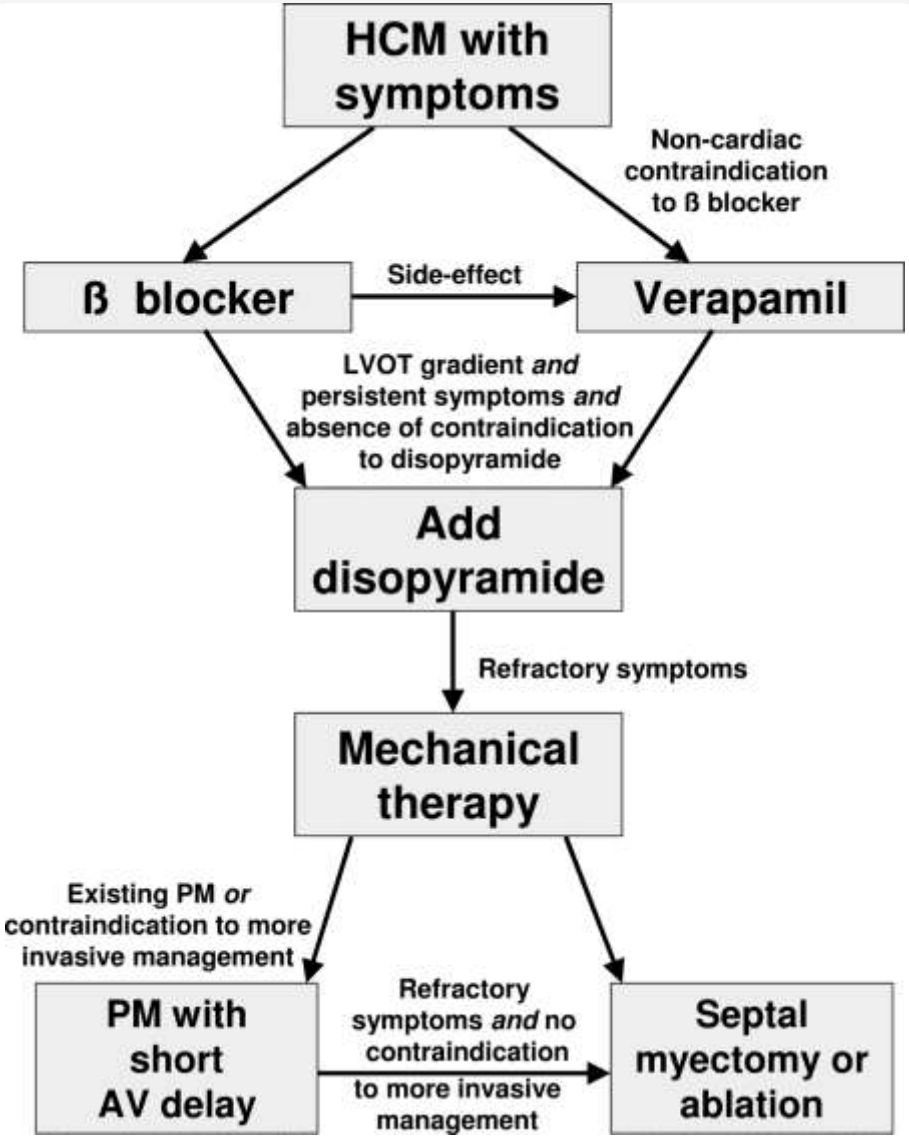
Sarcomeric Proteins



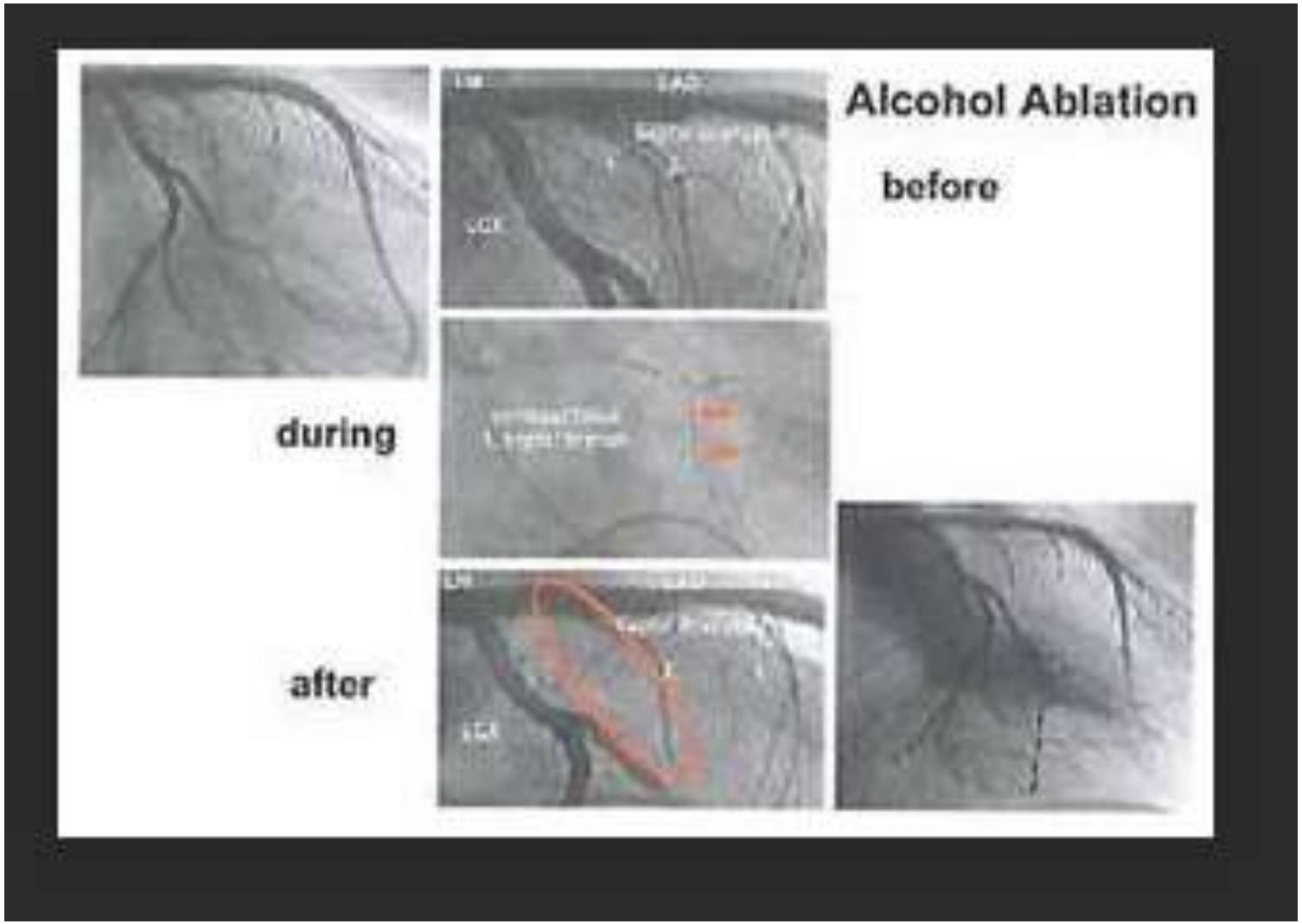
As Surgeons…

We are interested in obstructive forms of HCM (HOCM)

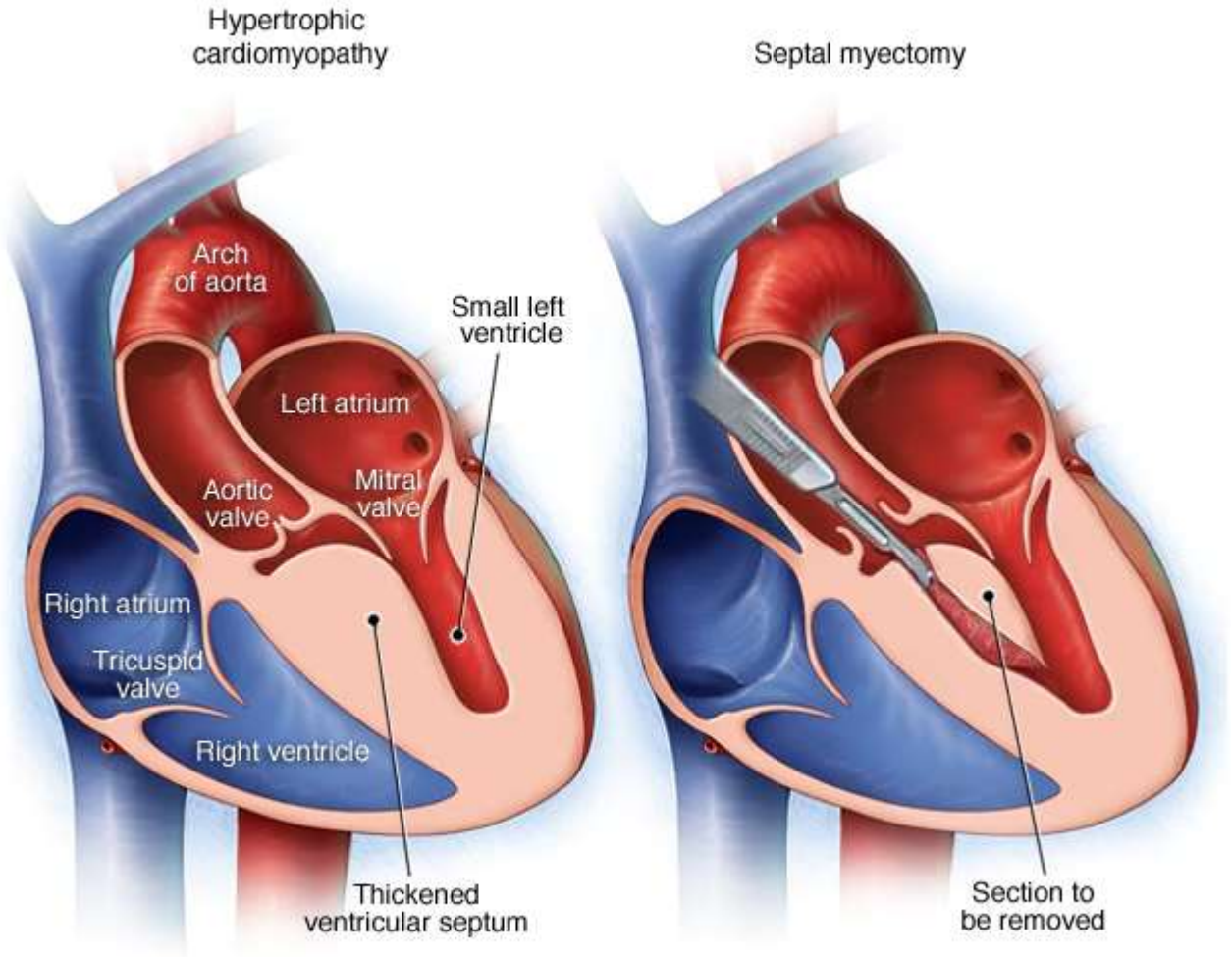
Treatment


HCM with symptoms
Non-cardiac contraindication to ß blocker
ß blocker
Side-effect
Verapamil
LVOT gradient *and* persistent symptoms *and* absence of contraindication to disopyramide
Add disopyramide
Refractory symptoms
Mechanical therapy
Existing PM *or* contraindication to more invasive management
PM with short AV delay
Refractory symptoms *and* no contraindication to more invasive management
Septal myectomy or ablation

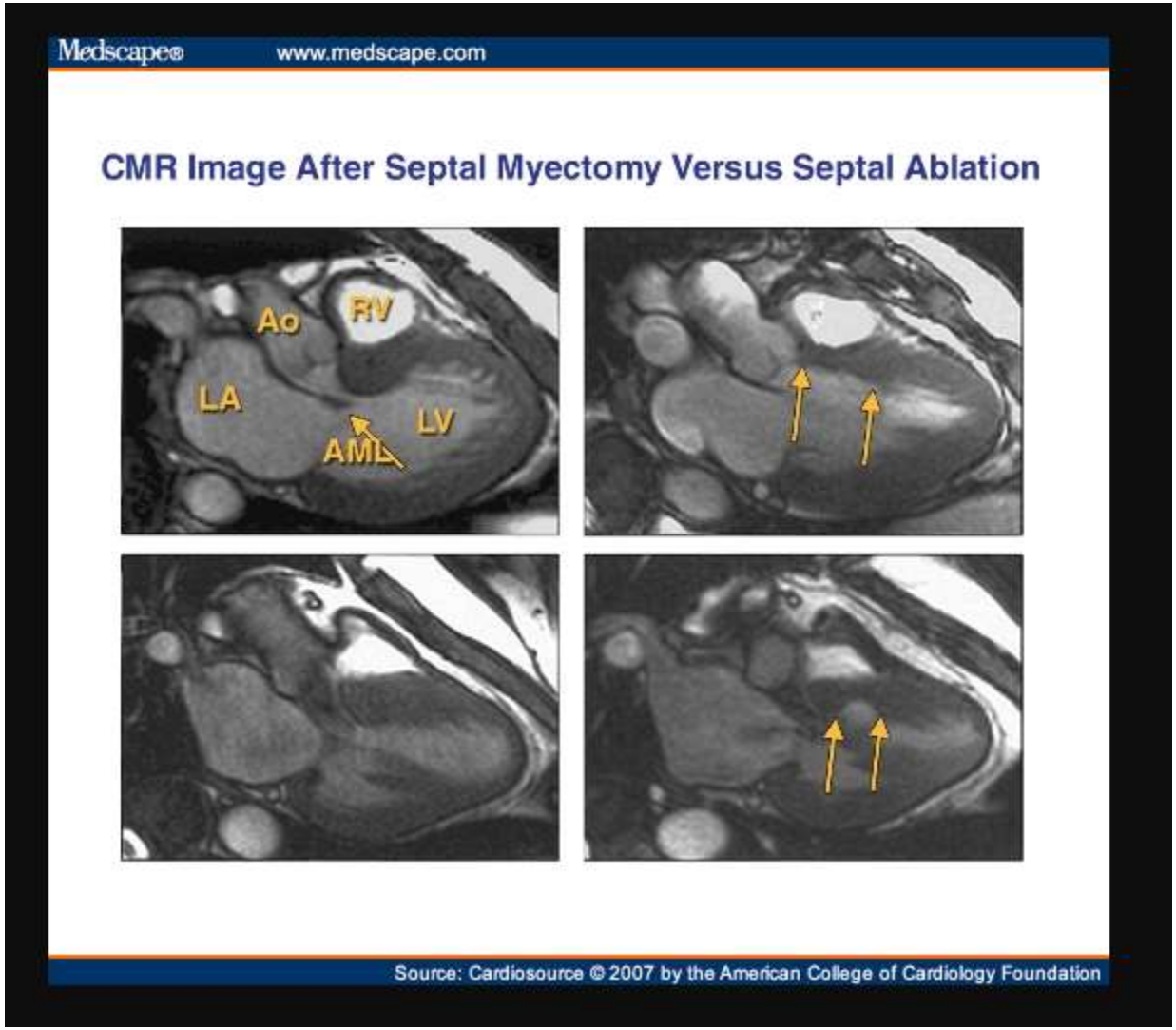
Alcohol Septal Ablation



Myectomy


Hypertrophic cardiomyopathy
Septal myectomy
Arch of aorta
Small left ventricle
Left atrium
Aortic valve
Mitral valve
Right atrium
Tricuspid valve
Right ventricle
Thickened ventricular septum
Section to be removed

Myectomy vs ASAblation



Myectomy vs ASAblation

Key question

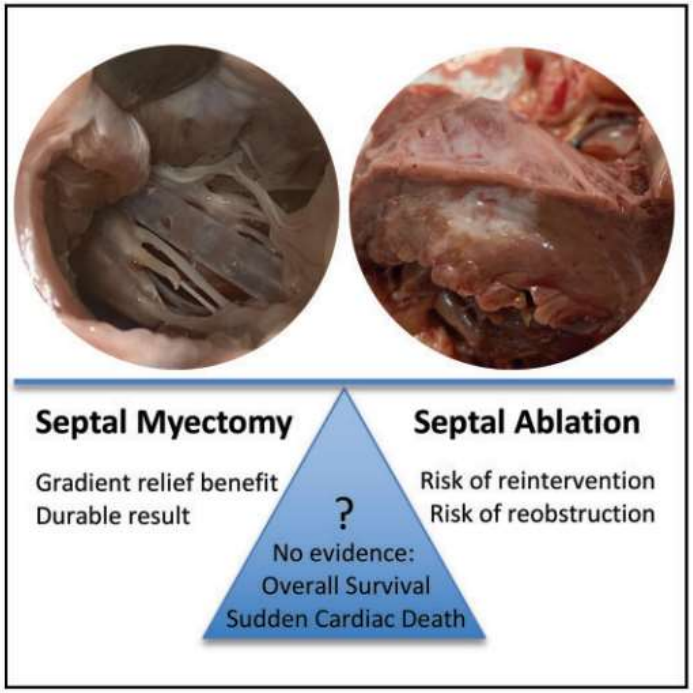
What is the optimal septal reduction therapy for hypertrophic cardiomyopathy: alcohol septal ablation or myectomy?

Key finding(s)

Alcohol septal ablation is associated with reoperation, residual mitral regurgitation and worse gradient relief.

Take-home message

Surgical septal myectomy remains the primary treatment option for symptomatic patients with severe hypertrophic obstructive cardiomyopathy.



Myectomy versus alcohol septal ablation in patients with hypertrophic obstructive cardiomyopathy

Alexander V. Afanasyev [a,*], Alexander V. Bogachev-Prokophiev [a], Maxim G. Kashtanov [b,c], Dmitriy A. Astapov [a], Anton S. Zalesov [a], Sergei A. Budagaev [a], Ravil M. Sharifulin [a], Eduard M. Idov [b,d] and Sergei I. Zheleznev [a]

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Myectomy vs ASAblation

Table 1: Baseline characteristics of septal myectomy and alcohol septal ablation groups before propensity score matching

Covariates before matching	SM (*n* = 345)	ASA (*n* = 150)	Bias (%)	*P*-value
Age (years)	55.0 ± 13.4	50.7 ± 14.6	30.7	0.001
Male, *n* (%)	155 (44.9)	72 (48.0)	-6.1	0.529
PM/ICD, *n* (%)	9 (2.6)	3 (2.0)	4.0	0.686
NYHA class III–IV, *n* (%)	222 (64.3)	71 (47.3)	36.6	<0.001
AF, *n* (%)	67 (19.4)	8 (5.3)	43.7	<0.001
AH, *n* (%)	206 (59.7)	81 (54.0)	12.7	0.193
CAD, *n* (%)	105 (30.4)	20 (13.3)	41.5	<0.001
DM, *n* (%)	33 (9.6)	7 (4.7)	19.1	0.066
MR 2+, *n* (%)	255 (73.9)	36 (24.0)	115.0	<0.001
LVEF (%)	71.1 ± 8.2	70.9 ± 8.0	2.9	0.769
PG (mmHg)	83 (70–96)	57 (38–84)	48.8	<0.001
IVS (mm)	24 (22–27)	22 (19–26)	39.4	<0.001

AF: atrial fibrillation; AH: arterial hypertension; ASA: alcohol septal ablation; CAD: coronary artery disease; DM: diabetes mellitus; ICD: implantable cardioverter-defibrillator; IVS: interventricular septum; LVEF: left ventricular ejection fraction; MR: mitral regurgitation; NYHA: New York Heart Association; PG: instantaneous peak Doppler left ventricular outflow tract pressure gradient; PM: pacemaker; SM: septal myectomy.

Outcomes at 5 years: SM vs ASA

Four and 12 deaths occurred in SM and ASA groups, respectively (NS)

Both procedures improved functional capacity; however, the prevalence of NYHA class III–IV significantly higher in the ASA group 1 year postoperatively [6 (6.4%) vs 0 (0%); P = 0.041].

Cumulative reoperation rates within 5 years were 2.0% (95% CI 0.5–7.6%) and 14.6% (95% CI 8.6–24.1%) in SM and ASA groups, respectively (P = 0.003)

Significantly higher rate of residual LVOT gradient >30 mmHg in the ASA group (5.7% vs 40.4%; P < 0.001).

Surgical Myomectomy vs Alcohol Septal Ablation

ACC/ESC EXPERT CONSENSUS DOCUMENT

American College of Cardiology/ European Society of Cardiology Clinical Expert Consensus Document on Hypertrophic Cardiomyopathy

A Report of the American College of Cardiology Foundation Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines

While alcohol ablation represents an option available to HCM patients and a selective alternative to surgery, it is not at this time regarded as the standard and primary therapeutic strategy for all severely symptomatic patients refractory to maximal medical management with marked obstruction to LV outflow (Table 1). Septal myectomy remains the gold standard for this HCM patient subset (7,11,14,41,232).

Surgical Indications

ACC/AHA and ESC guidelines recommend that septal reduction therapy should be performed only by experienced operators, working as part of a multidisciplinary team expert, in the management of HCM [1, 7]. A SRT should be performed in presence of:

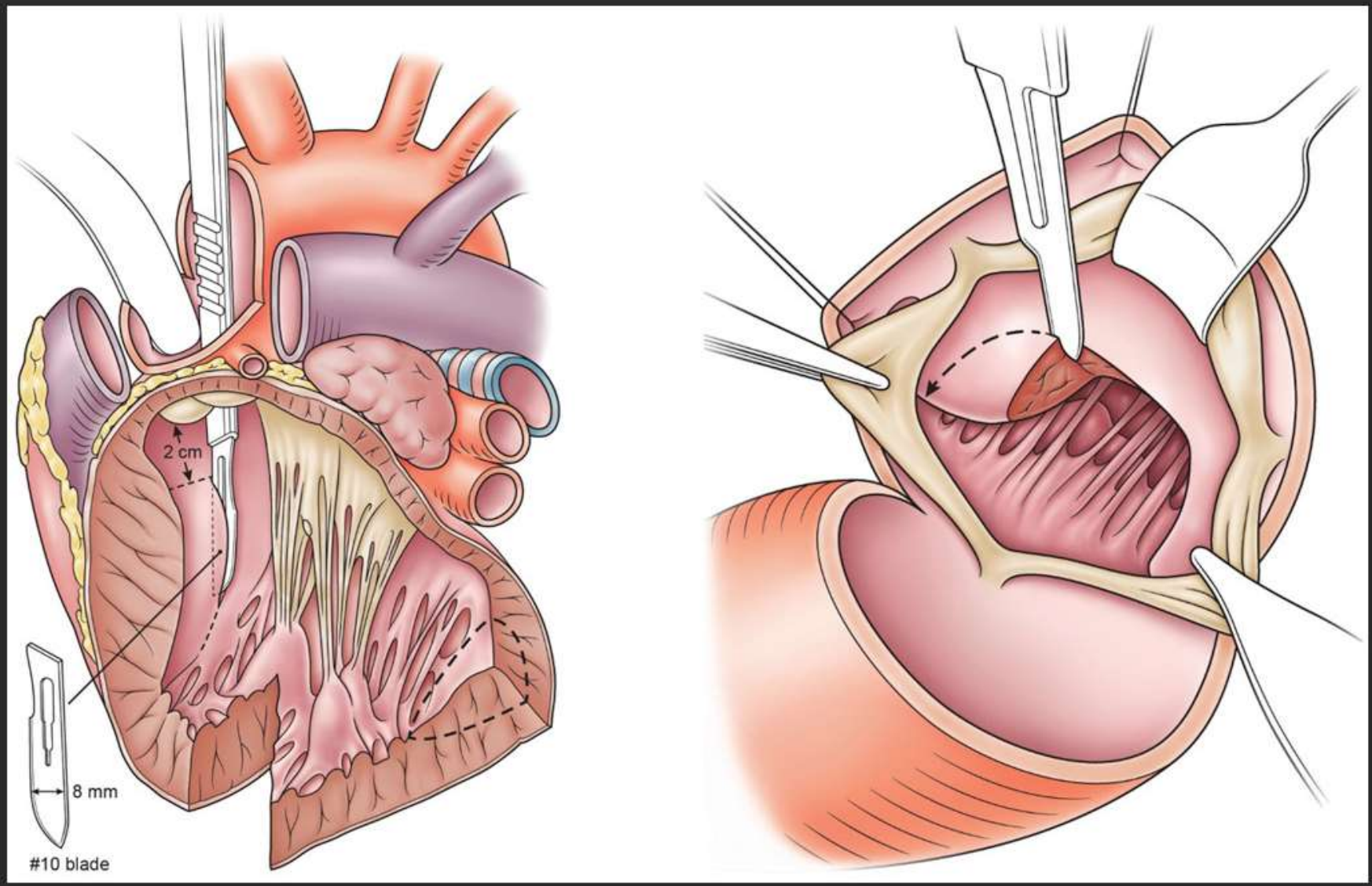
1. Significant LVOT obstruction (resting or maximum provoked LVOT gradient of ≥50 mmHg)

2. Angina, Dyspnea (NYHA functional class III-IV), Syncope

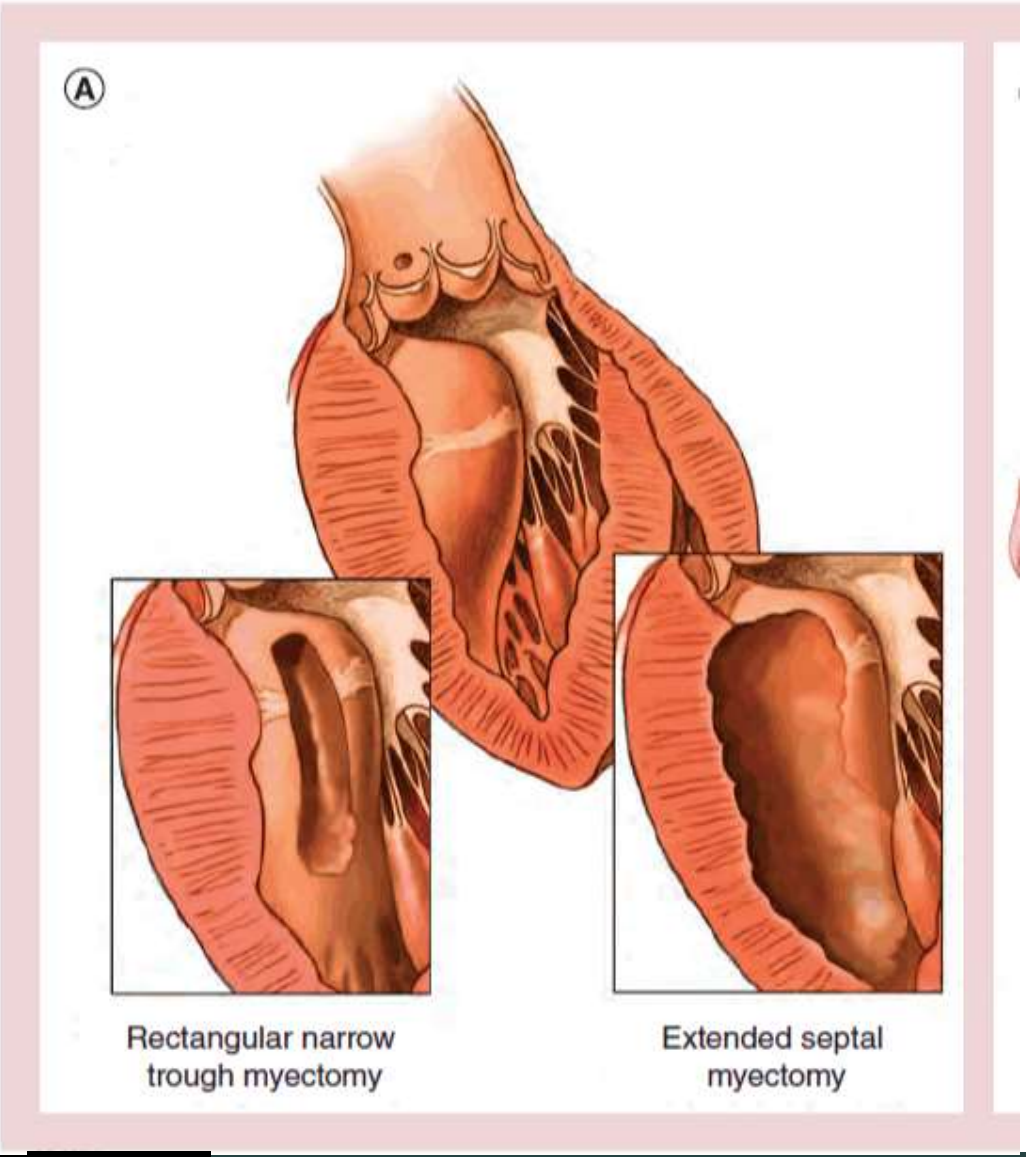
3. Persistent symptoms on maximal medical therapy.

4. The current indications have expanded recently to include symptomatic patients with low resting outflow gradients and latent obstruction.

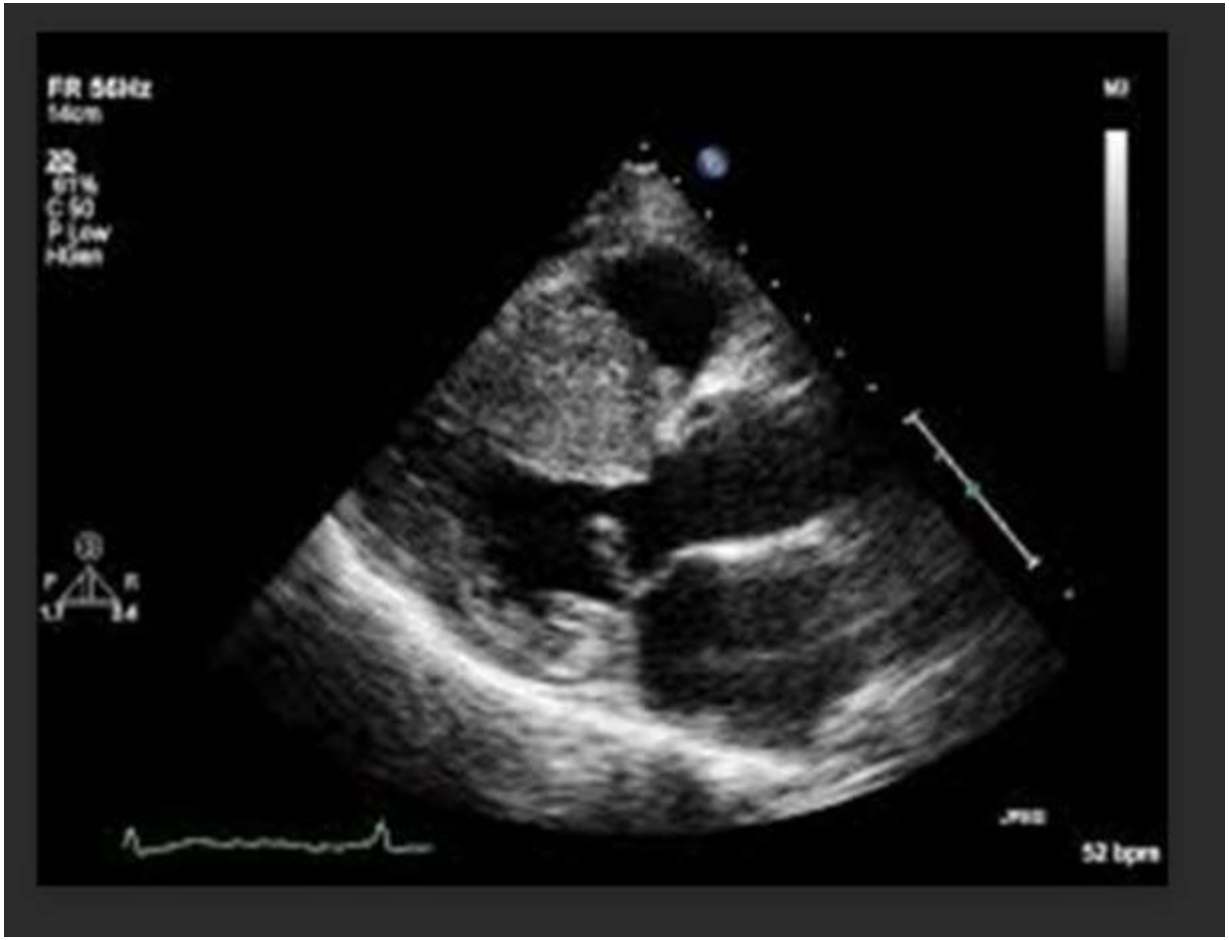
Myectomy



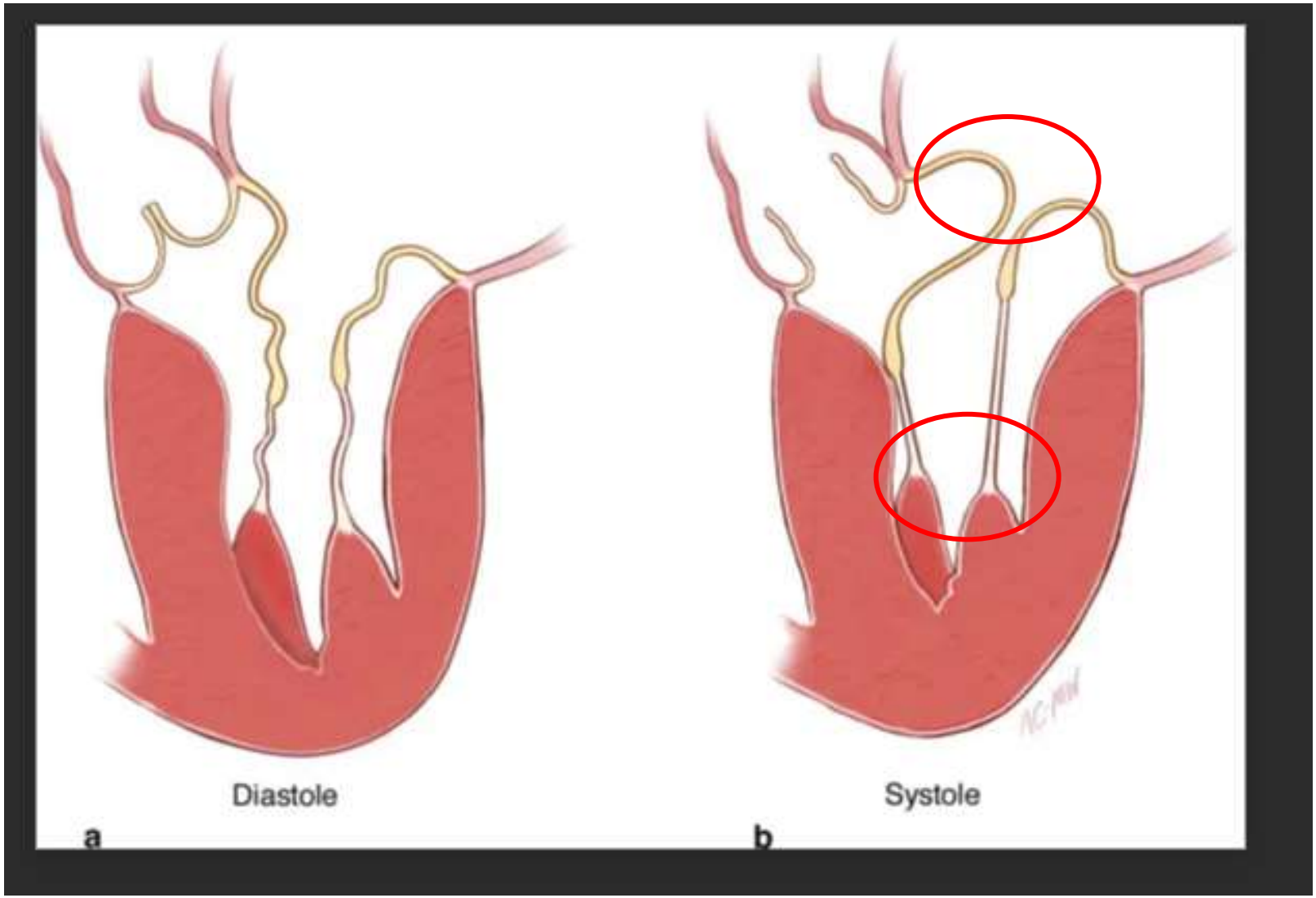
Extended Myectomy



SAM/Mitral Valve

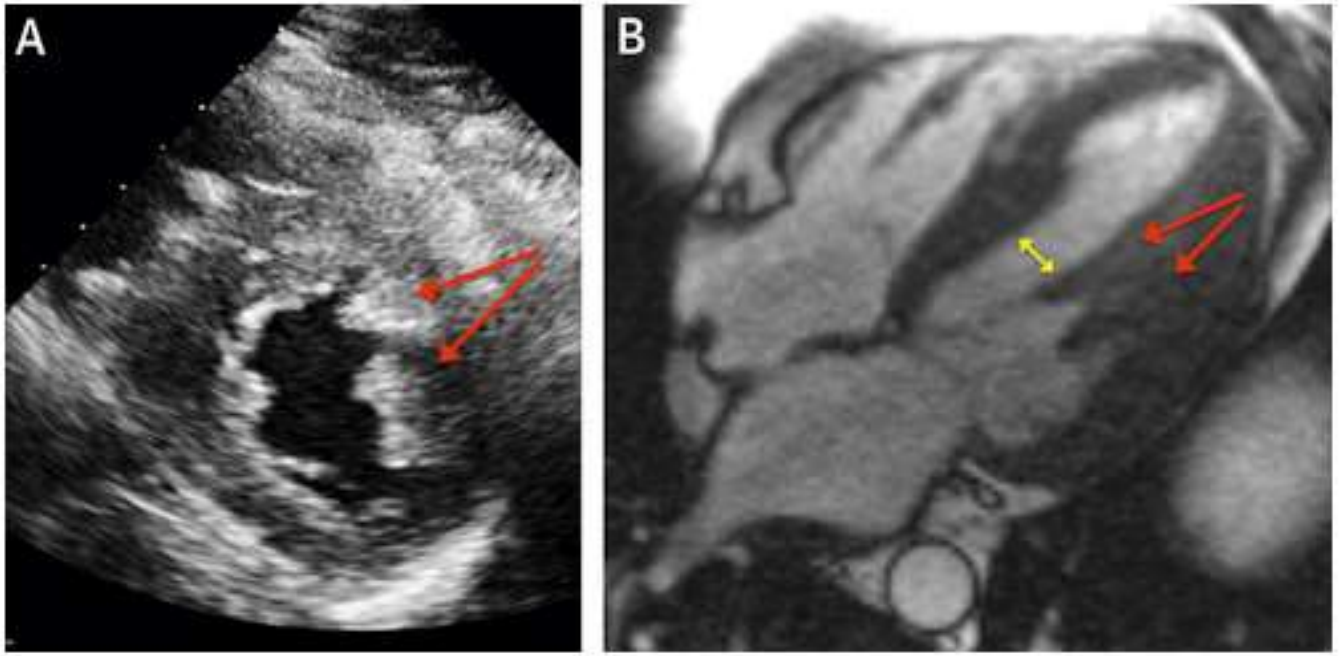


MV repair

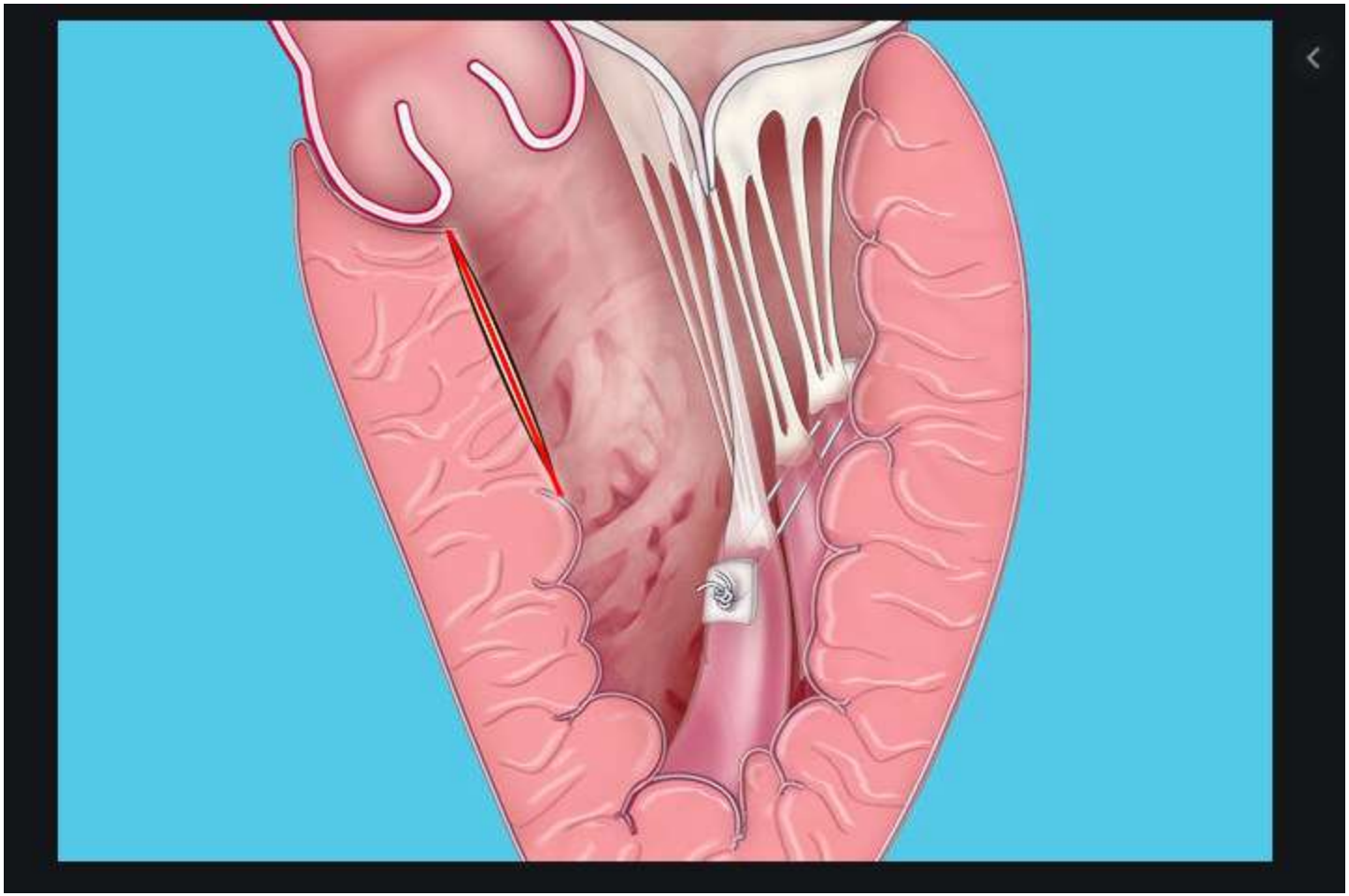


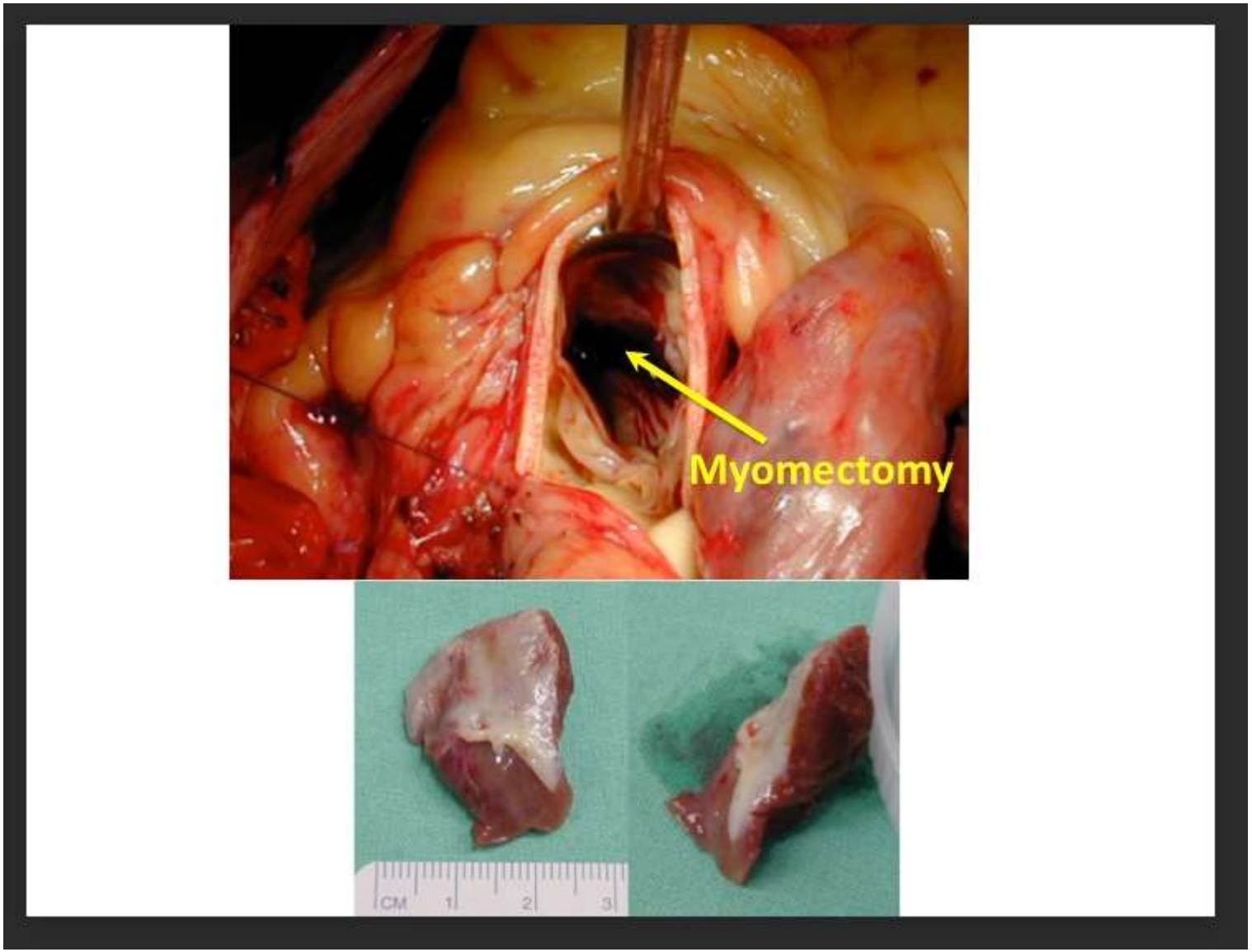
Long Anterior Leaflet

Abnormal Papillary Muscles

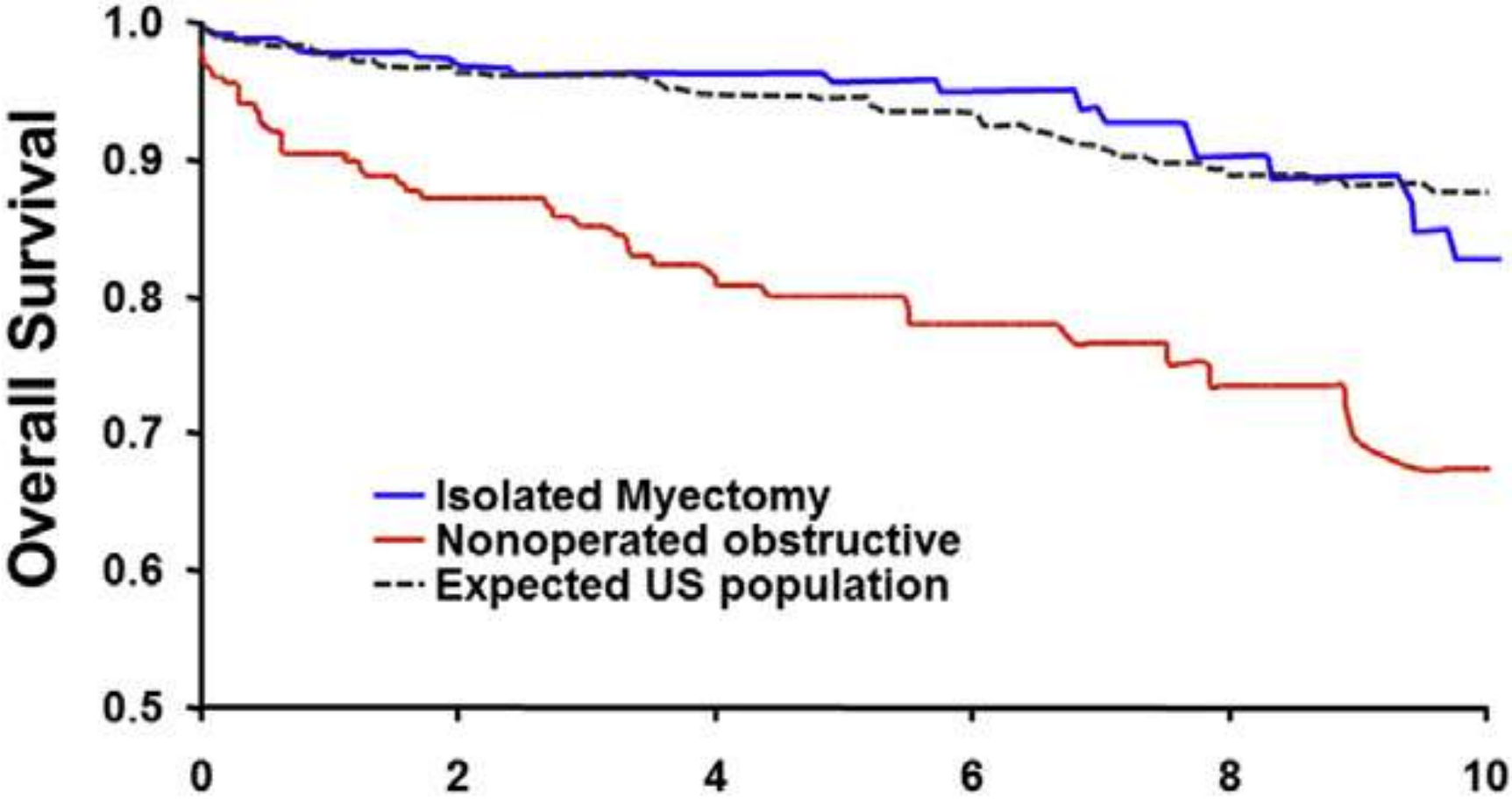


Repair papillary muscle abnormalities




Myomectomy
CM
1
2
3

Outcomes after Surgical Resection



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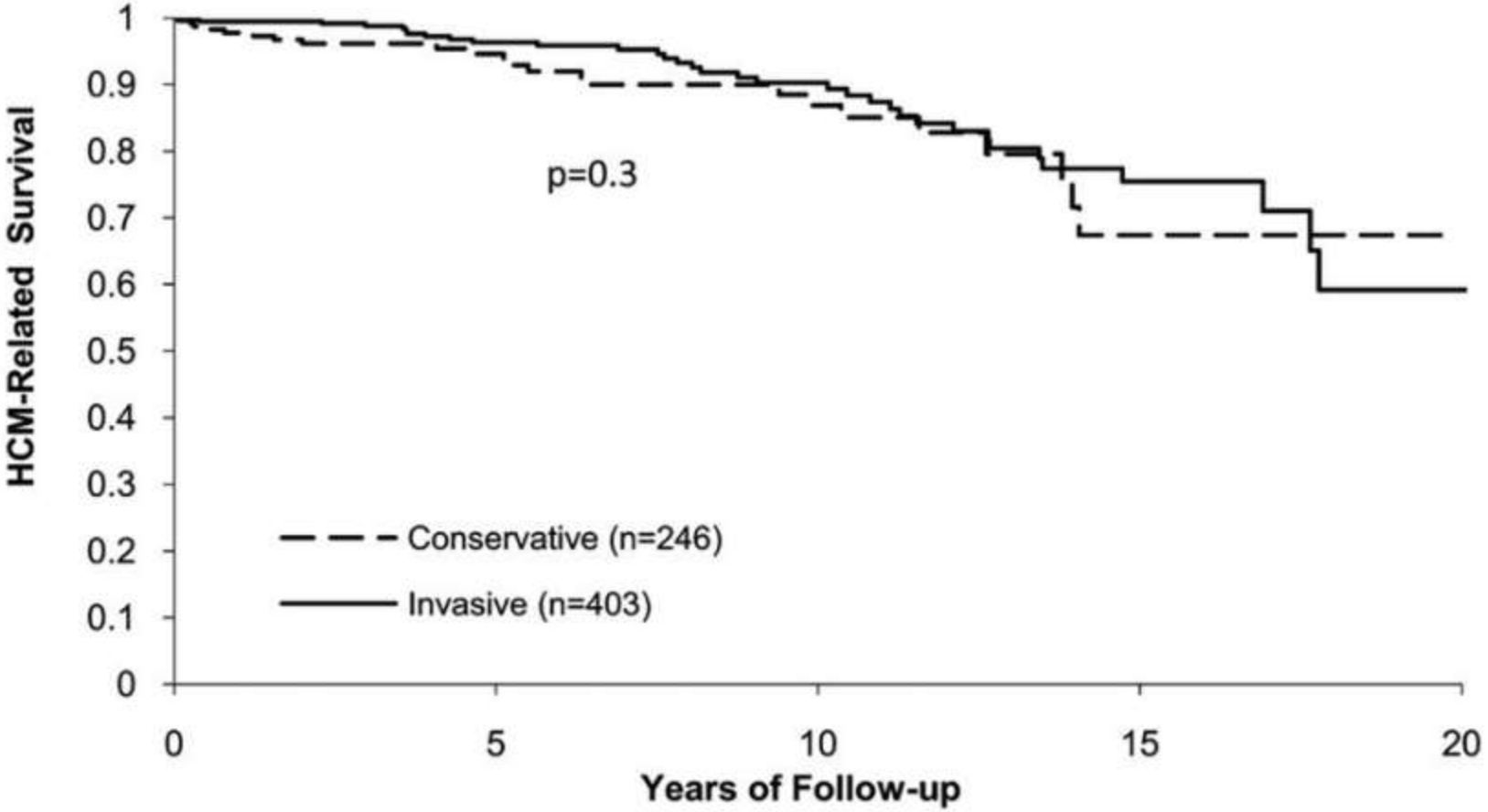
Staged Medical—Surgical Therapy



Summary

Myectomy superior to ASA?

Must address abnormal papillary muscles and mitral valve

Extended myectomy

Long term outcomes excellent despite concern about sudden death