Hypertrophic Cardiomyopathy Surgical techniques for obstructive disease

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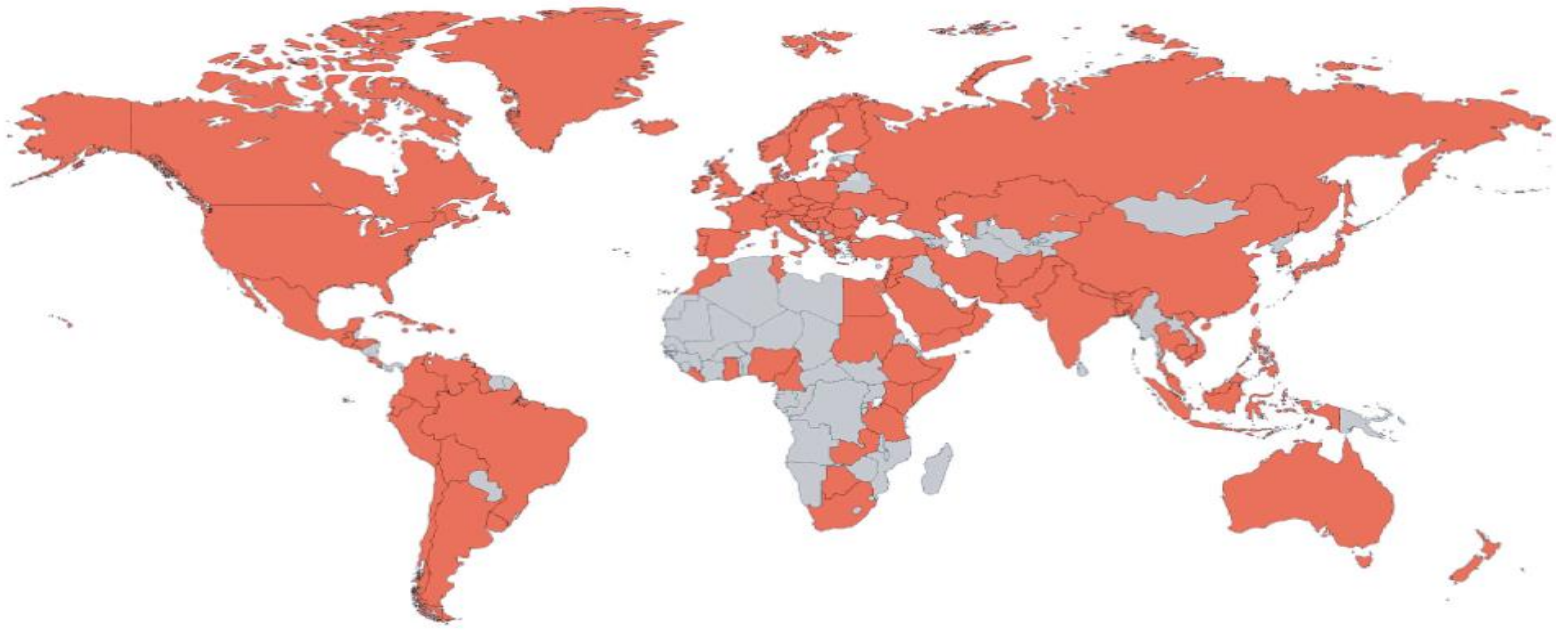
Conflict of interest

NONE

Introduction

- HCM is a relatively common genetic contemporary heart disease
- Diverse and complex phenotypic/genetic expression & clinical course
- Nowadays > highly treatable with effective options that alter natural history > reduced mortality+ normal or extended life expectancy
- In patients with HF symptoms (mainly due to LV outflow tract obstruction), septal myectomy or percutaneous alcohol ablation (in selected patients) can reverse the course of the disease.

Prevalence



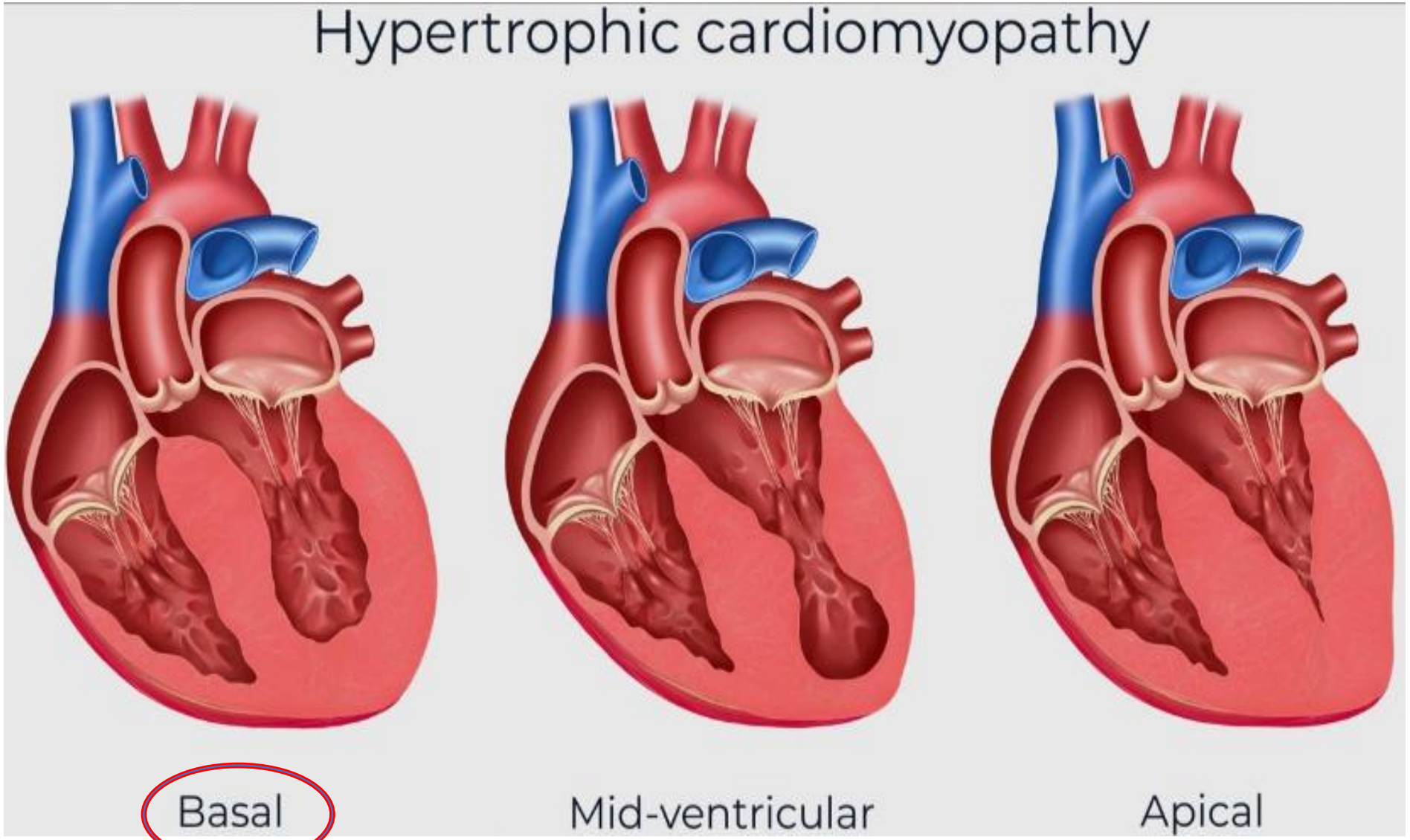
high proportion of unrecognized disease in the general population

Hypertrophic obstructive cardiomyopathy (HOCM)

- **>50%** of symptomatic (Chest pain, exertional dyspnea, presyncope) patients with HCM have **LVOT obstruction**

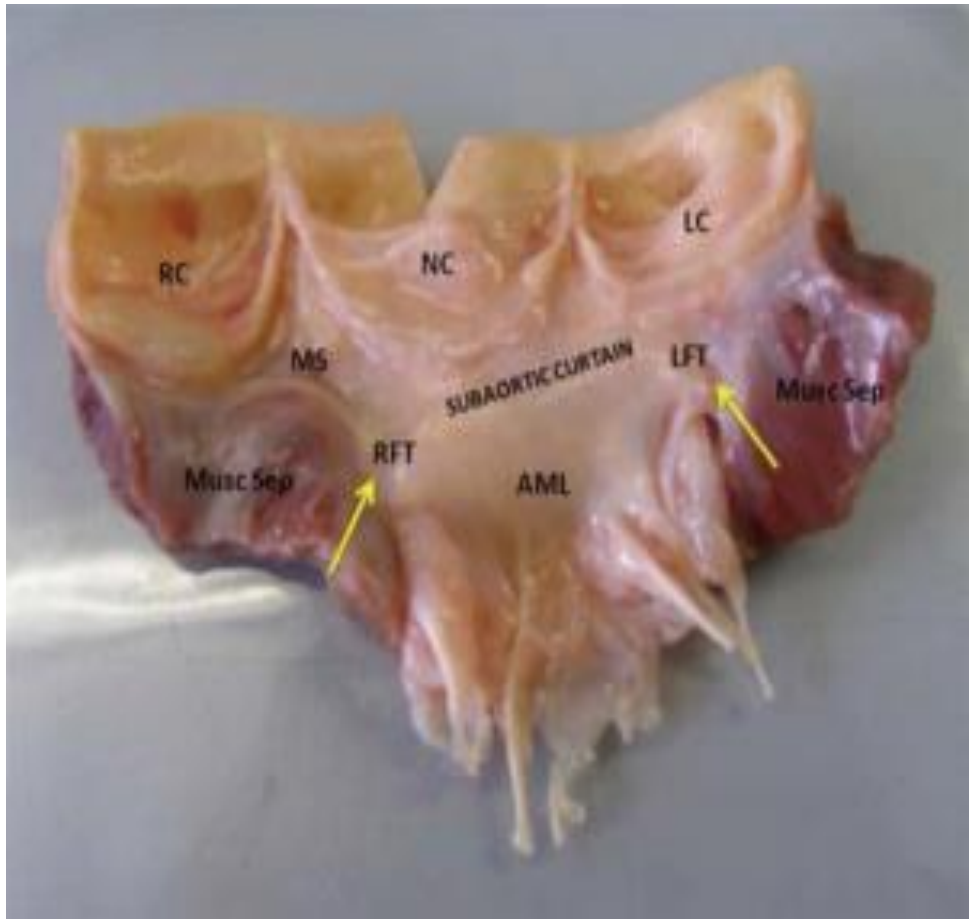
- Various phenotypes of HOCM > clinical manifestation and surgical management.

Phenotypes Variants of hypertrophic cardiomyopathy



Left ventricular outflow track (LVOT)

- Fibrous trigones (RFT & LFT)
- Subaortic curtain
- Anterior mitral leaflet (AML)
- Muscular interventricular septum
- Membranous septum (MS)

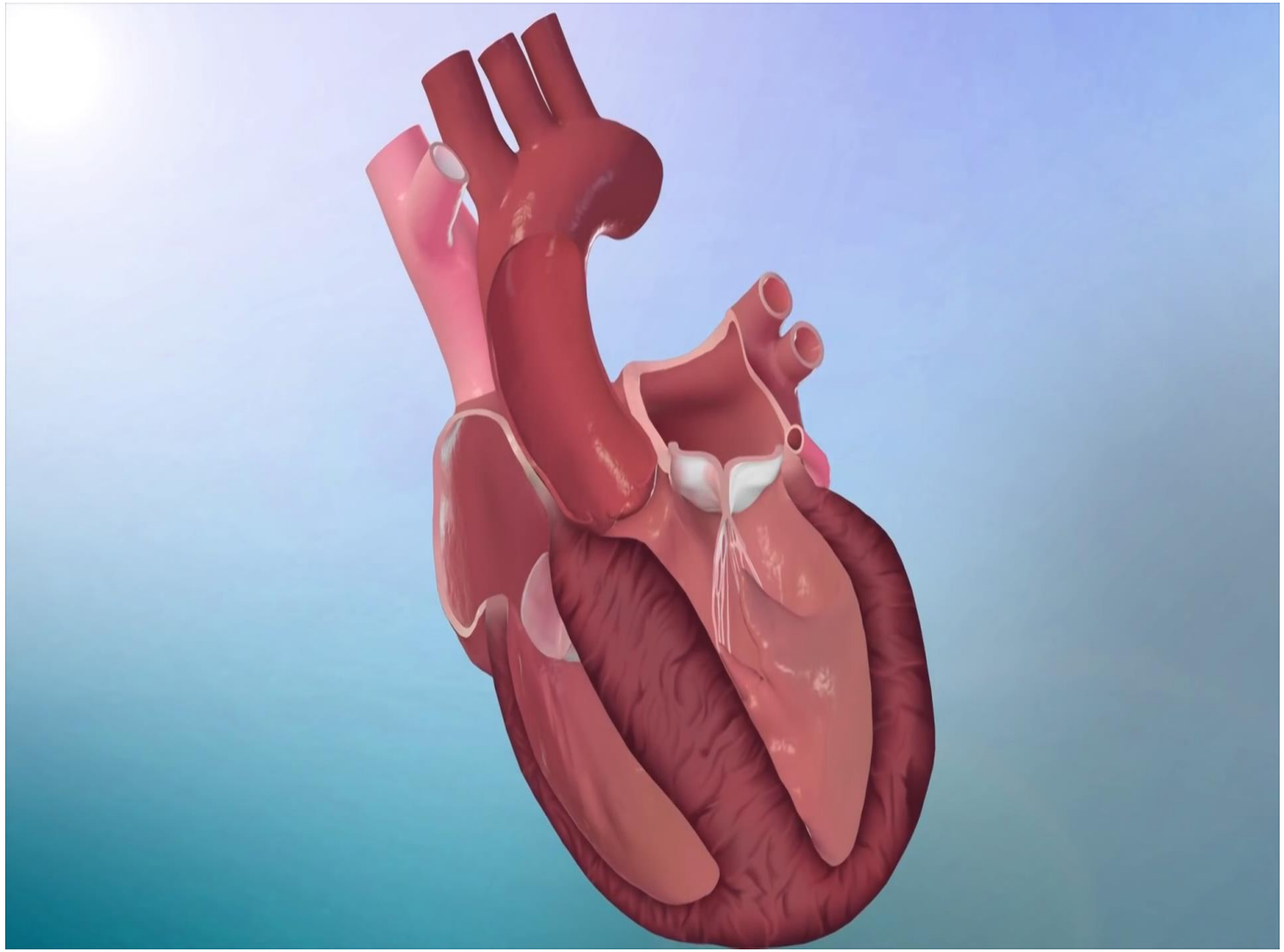


Pathophysiology subaortic obstruction

SAM
Elongated anterior leaflet (+/-)
Mitral regurgitation

Chordae tendinae

Papillary Muscle



Diastolic dysfunction, myocardial ischemia, arrythmia, metabolic & energetic abnormalities

Surgery

Surgical Indications

- **Clinical**: symptomatic severe dyspnea or chest pain (NYHA III or IV), or syncope attribuable to LVOTO interfering with everyday activity or QOL despite OMT

- **Hemodynamic**: dynamic LVOT gradient at rest or with physiologic provocation (peak Gradient ≥50 mmHg + septal LVH+SAM)

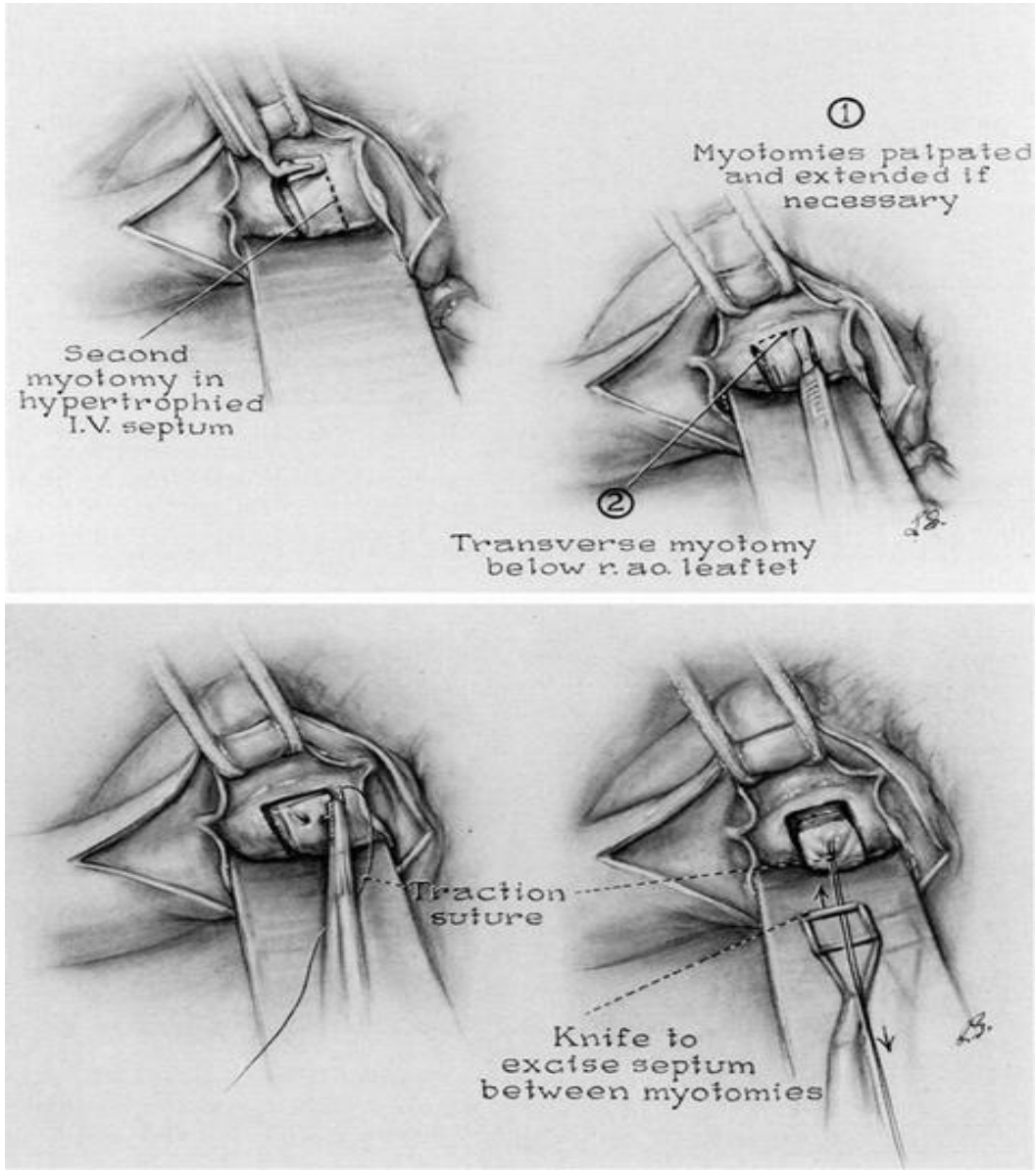
- **Anatomic**: targeted anterior septal thickness

Historical perspective

- Russel Brock (1957): description
- Clealand technique (1958) : very limited septal myectomy
- Morrow technique (1961) : limited Septal myectomy
- Messmer technique (1994): extended Septal myectomy

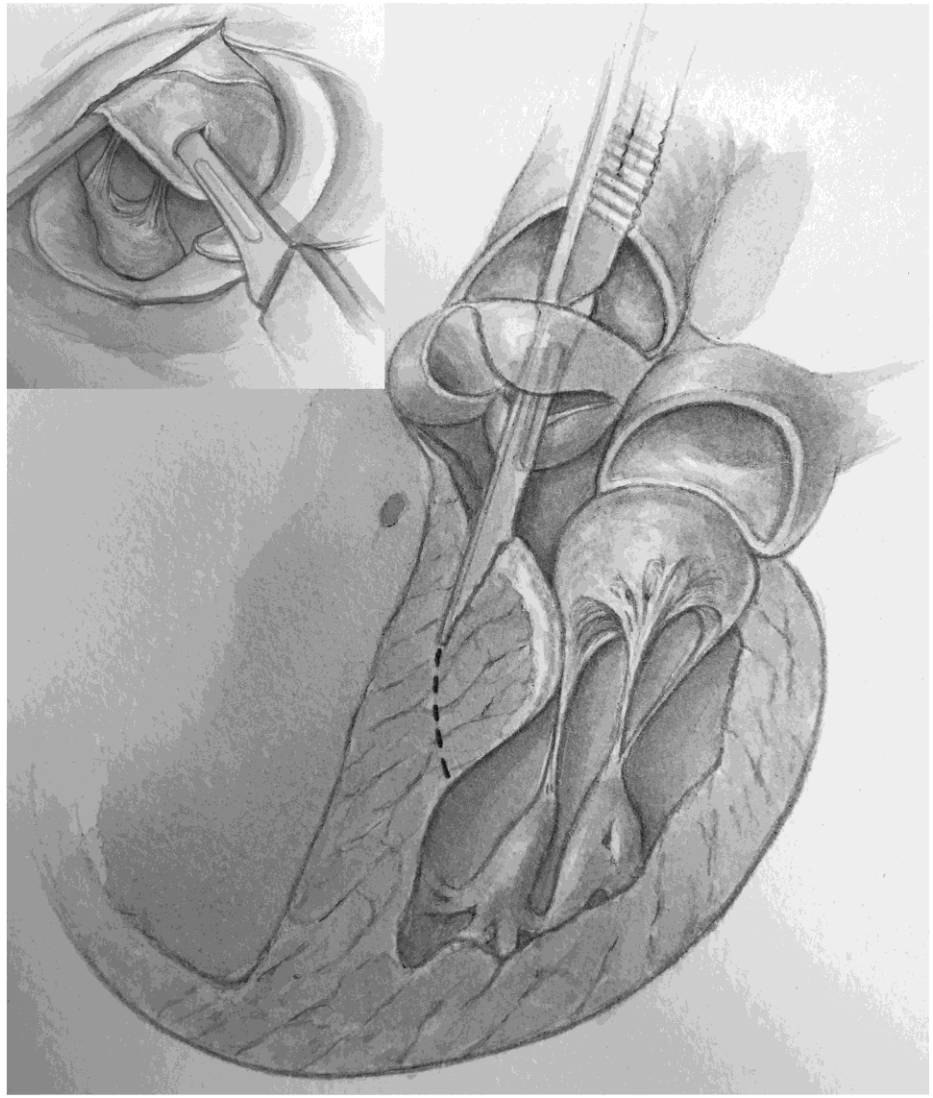
Morrow

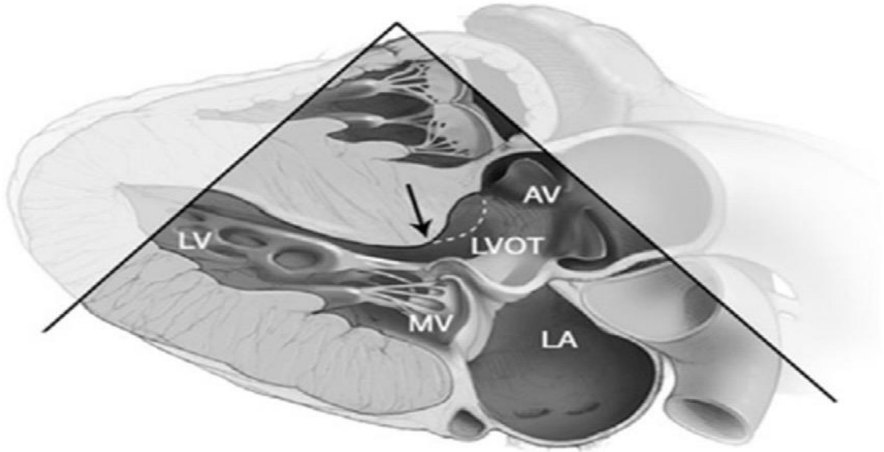
Poor exposure
Limited resection



Messmer

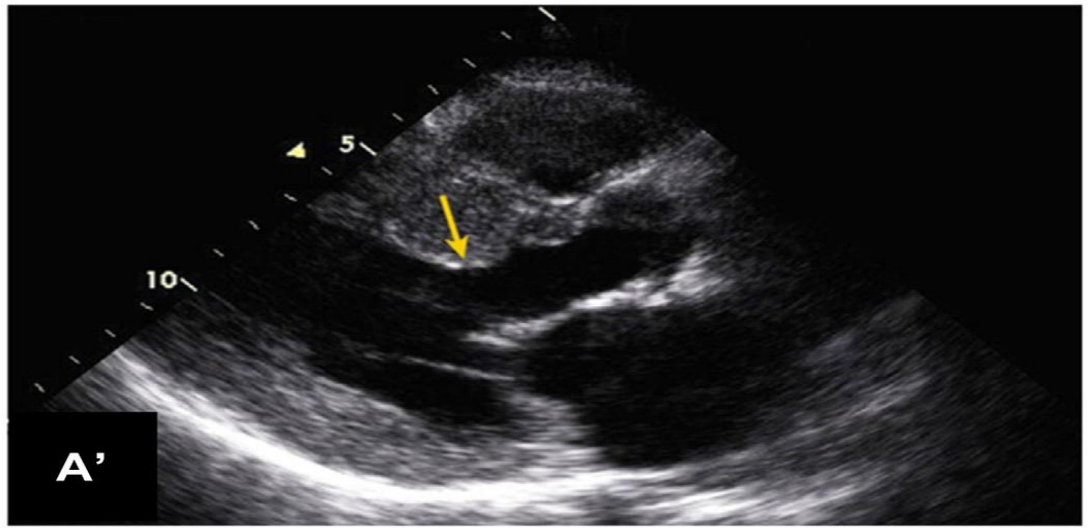
Better exposure
Additional strategies

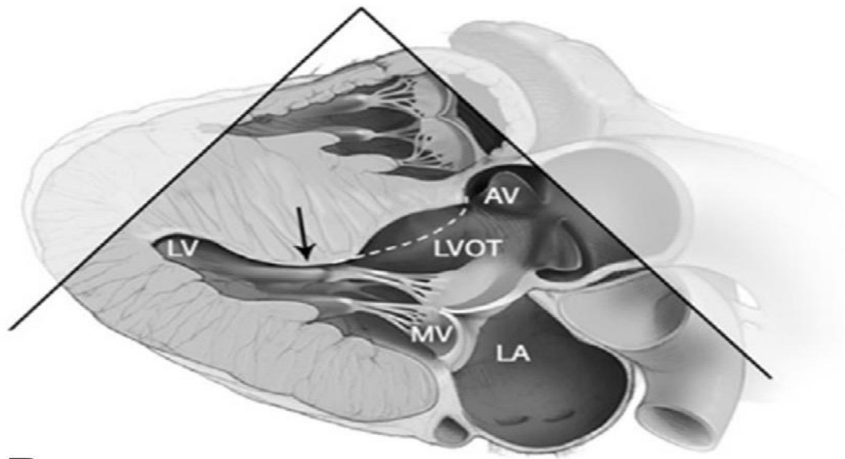




A

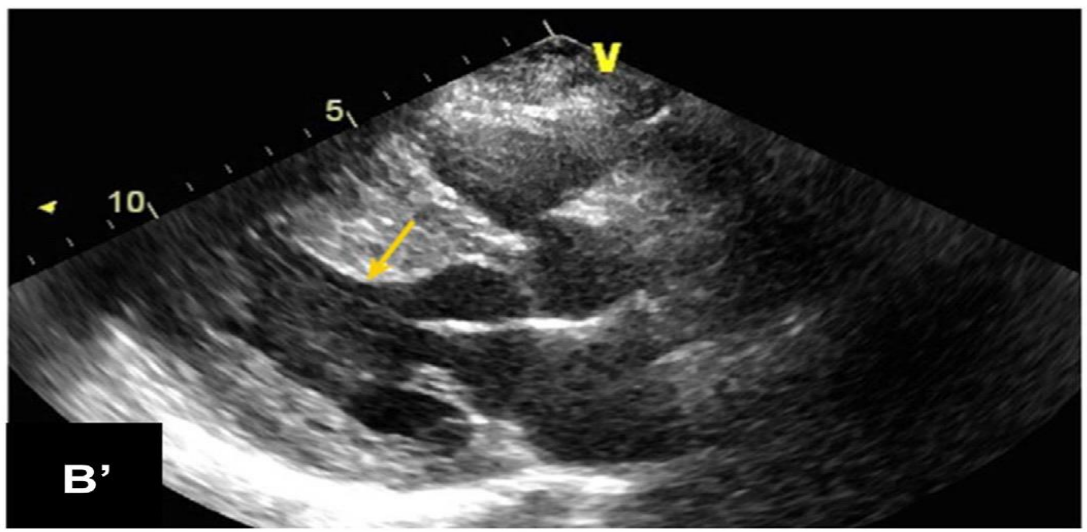
Subaortic residual hypertrophy





B

Unrecognized midventricular hypertrophy



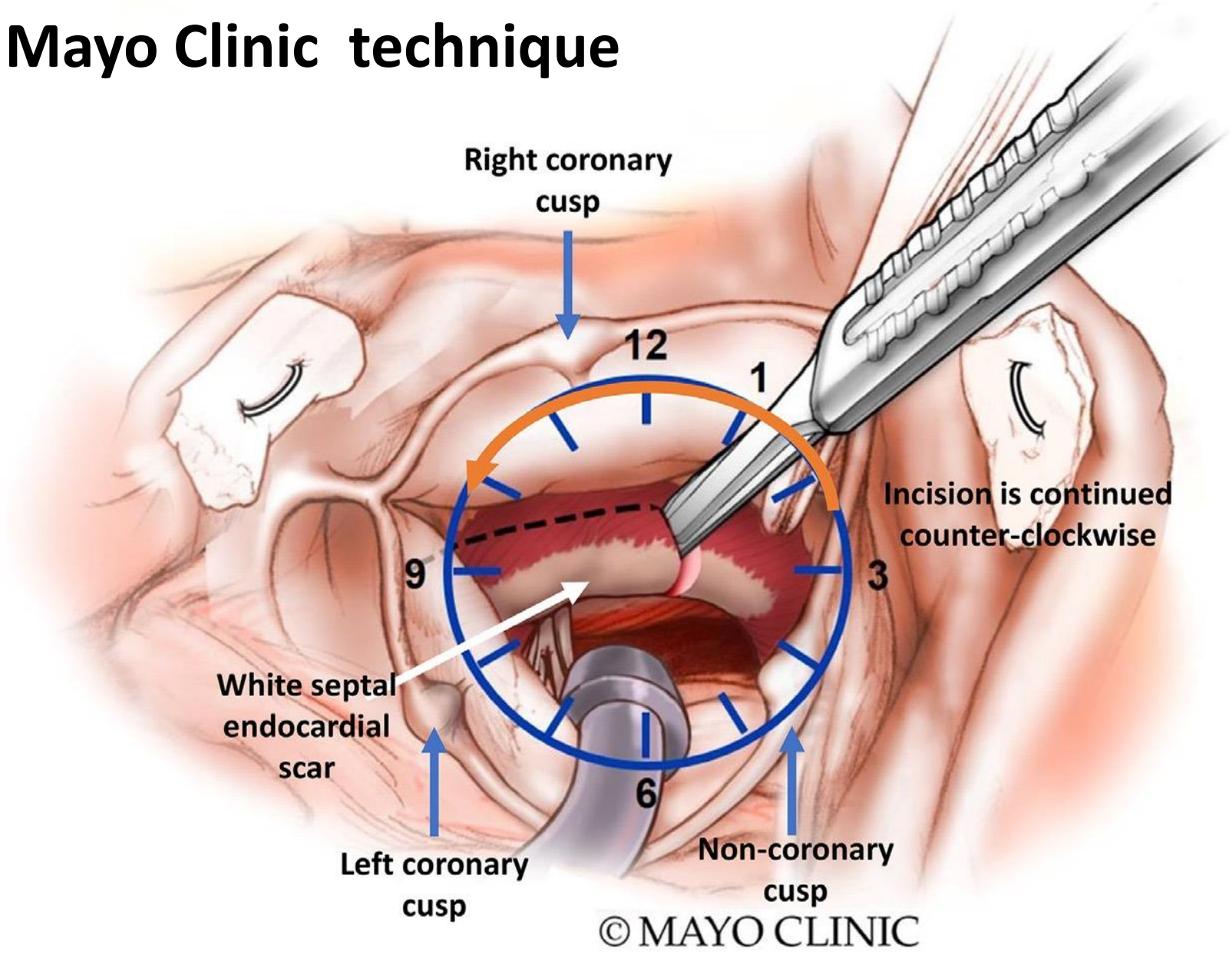
Preoperative assessment of potential myectomy candidates

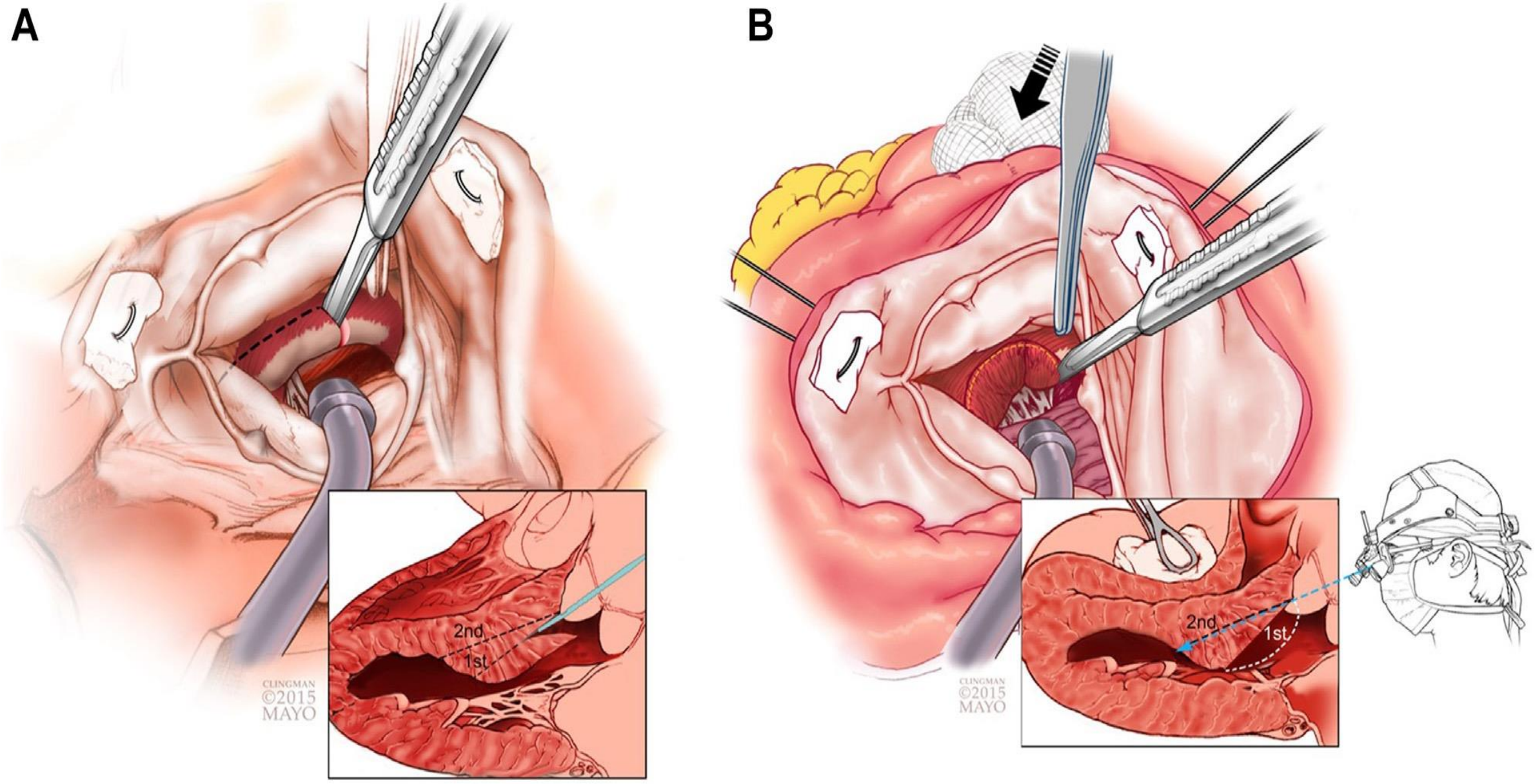
Modality	Findings
Echocardiography	• Wall thickness • SAM • LVOTO • LA dimensions • Systolic and diastolic function & LV cavity size • Rule out others etiologies
Cardiac magnetic resonance	• Pattern and degree of hypertrophy Fibrosis with LGE
Cardiac CT	• Myocardial bridges
Cardiac catheterization	• LVOTO • Myocardial bridges

Surgical Challenges

- **Broad morphological spectrum** (septal thickness mild to massive) associated with diverse **abnormalities of mitral valve apparatus**.
- Basal anterior **septum** (in continuity with the anterior free wall) the most common location for LVH. But LVH can be limited and focal in some patients
- Compression of coronary arteries by **myocardial bridges** (diastolic FFR)

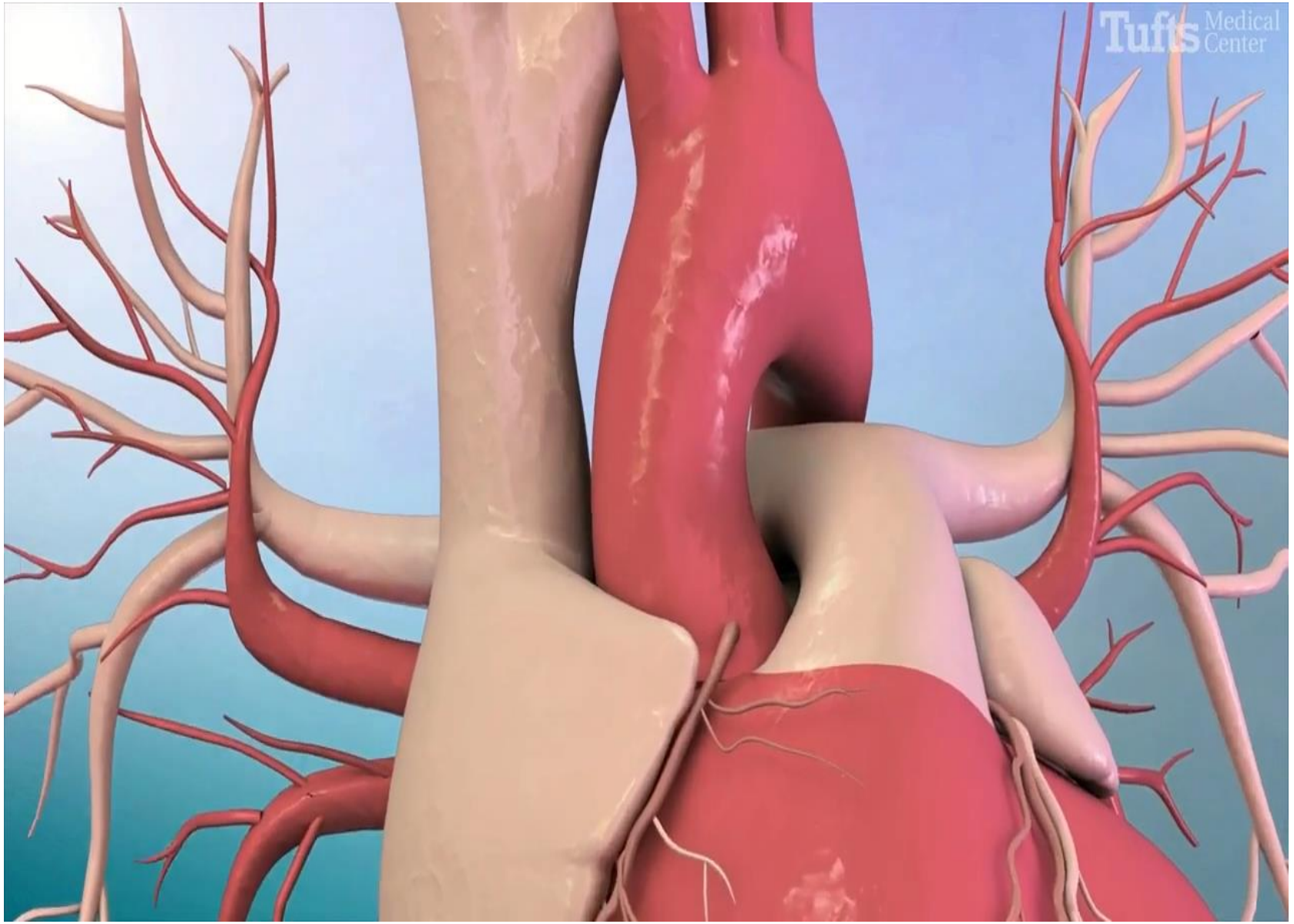
The Mayo Clinic technique



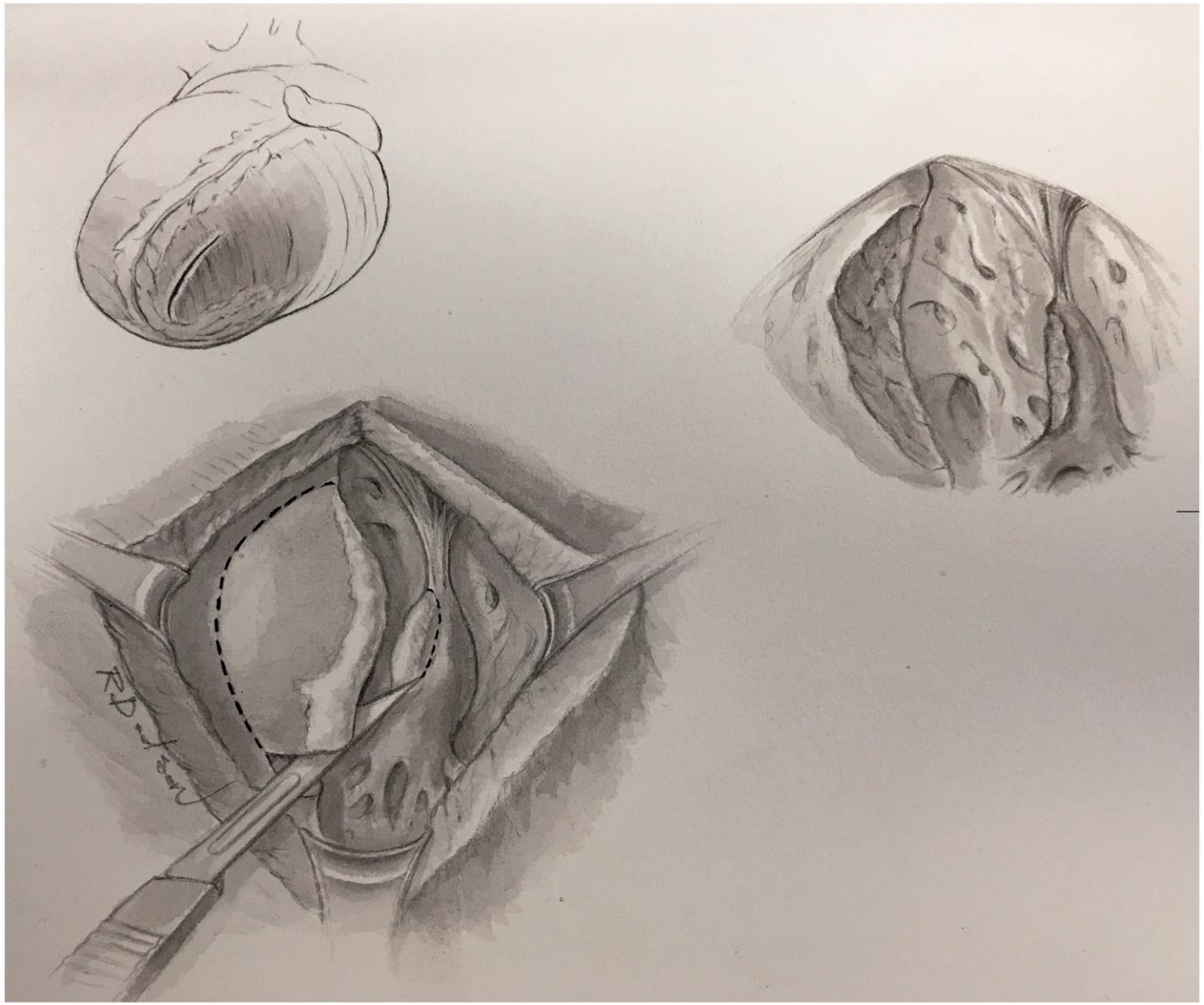


3 to 12 g of muscle

Transaortic extended septal myectomy



Apical Myectomy for Symptomatic patients with apical HCM and small LV end-diastolic volume

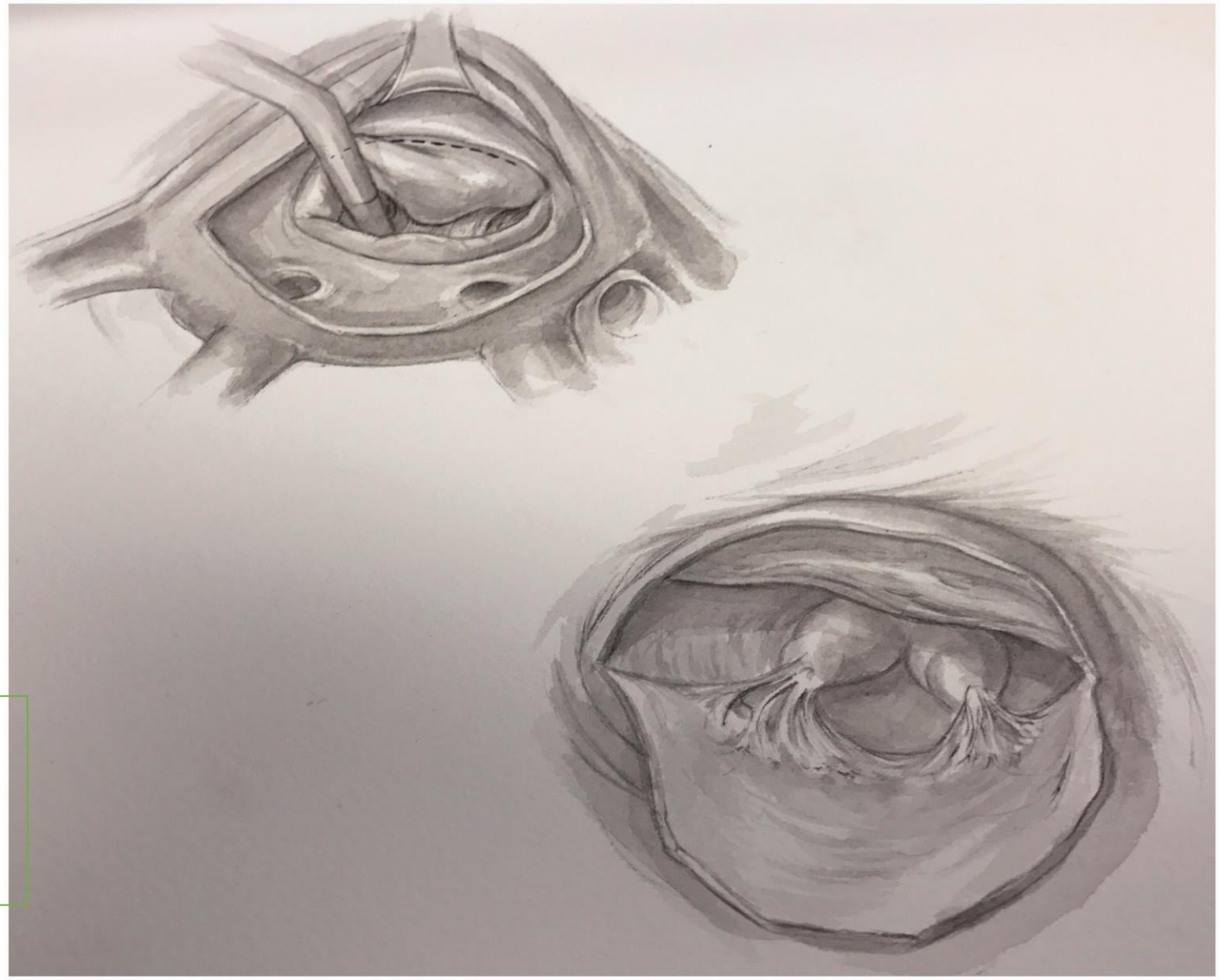


Transatrial transmitral approach

- Sternotomy
- Robotic
- Thoracotomy

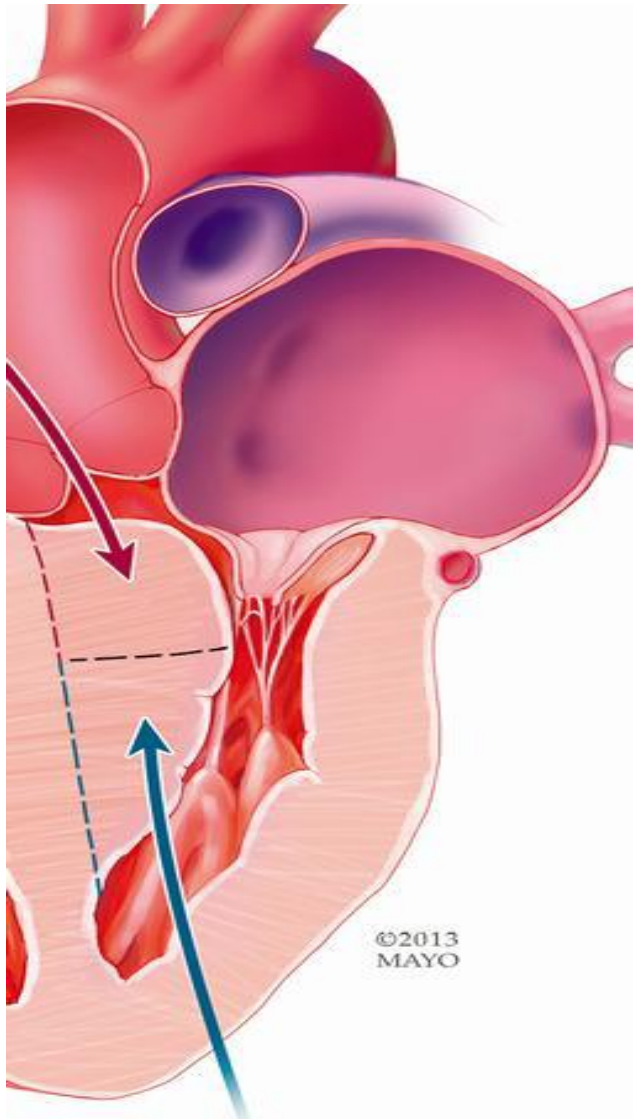
- Limited resection
- Detachment of AML

- Infants & small children
- Association with degenerative
- MV disease



Double approach

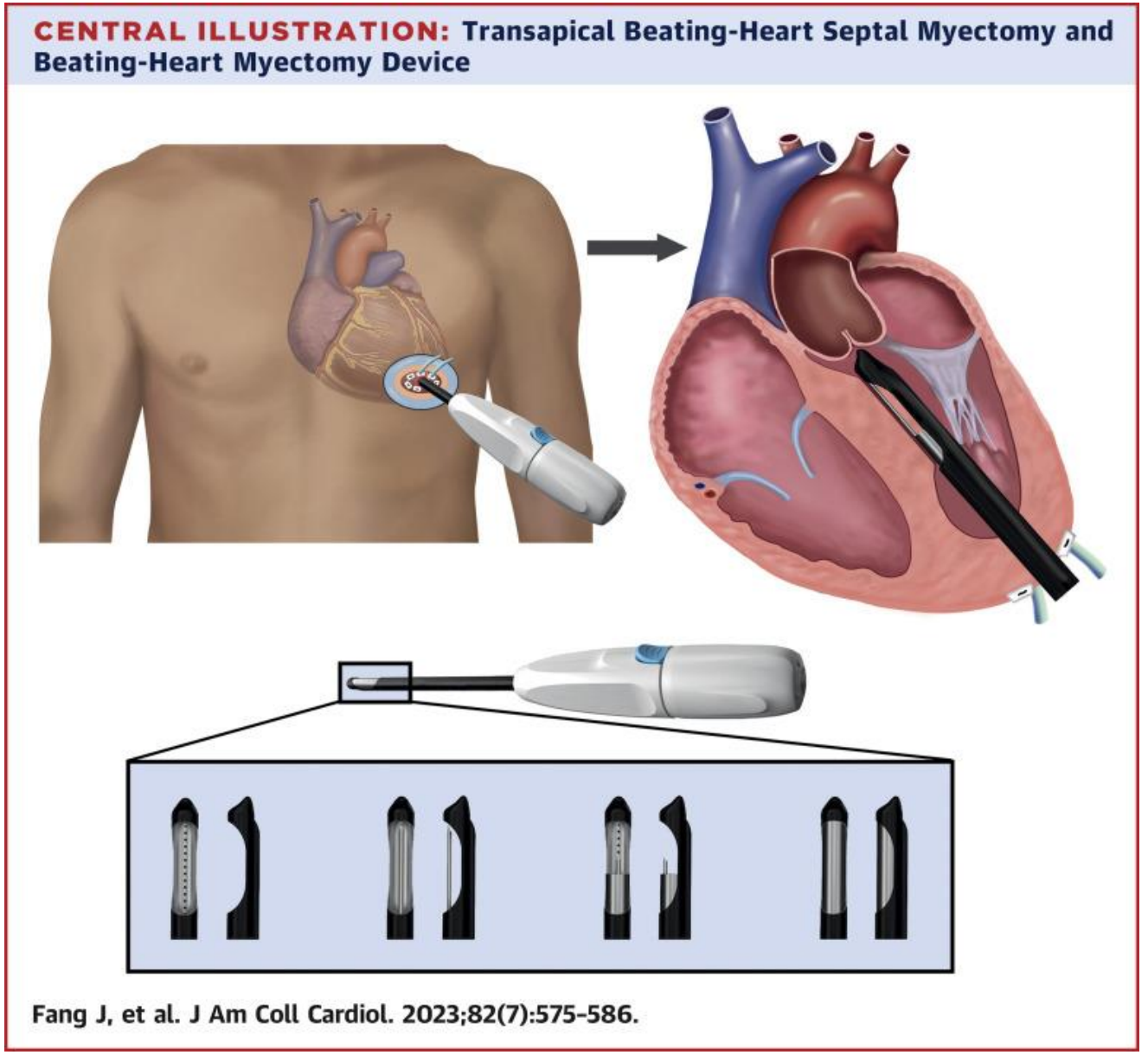
Transaortic & transapical



ALTERNATIVES APPROACHES

Transapical beating heart under TEE

Faster recovery

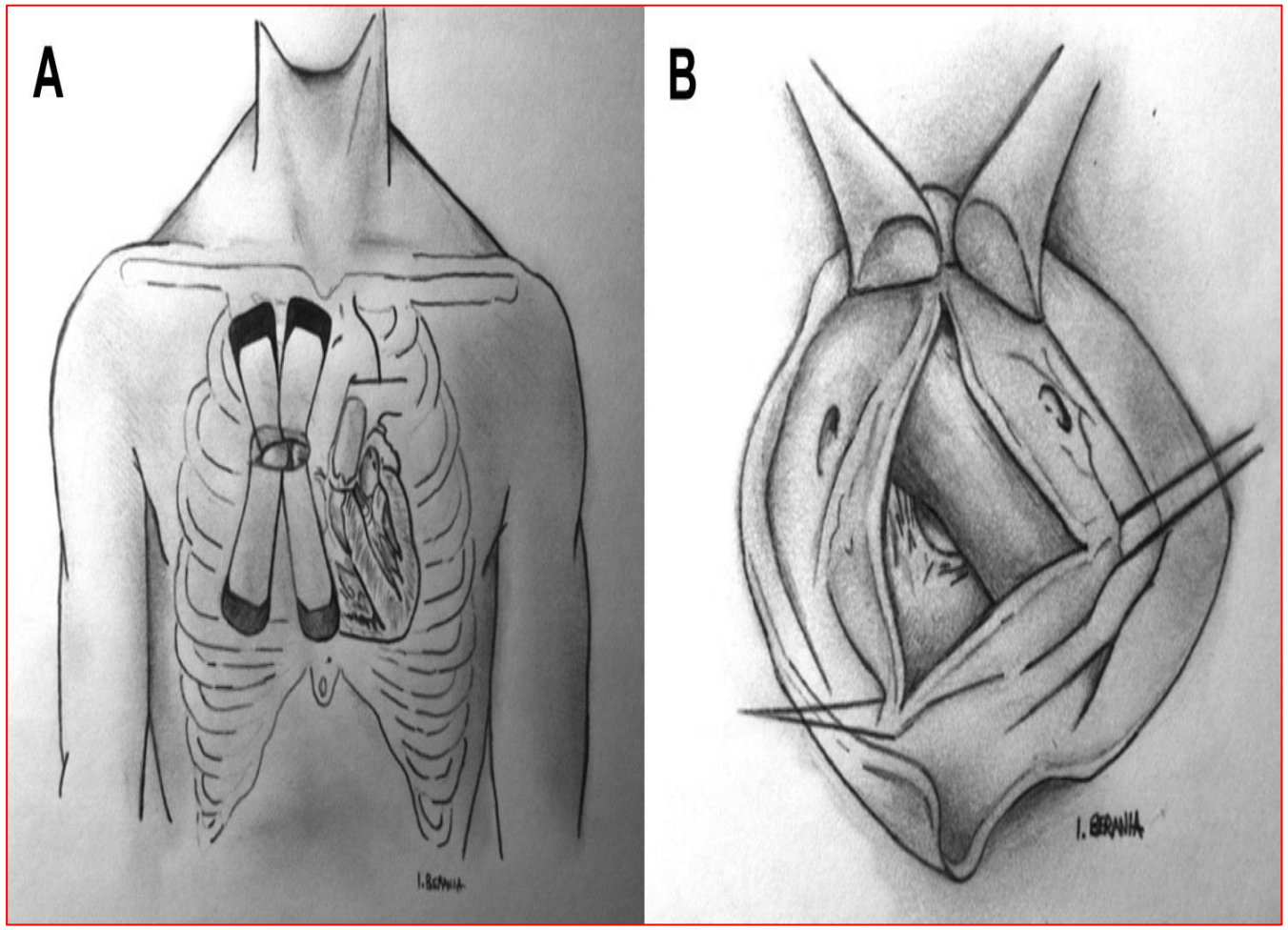


ALTERNATIVES APPROACHES

Minithoracotomy

- Subaortic phenotype
- Limited exposure

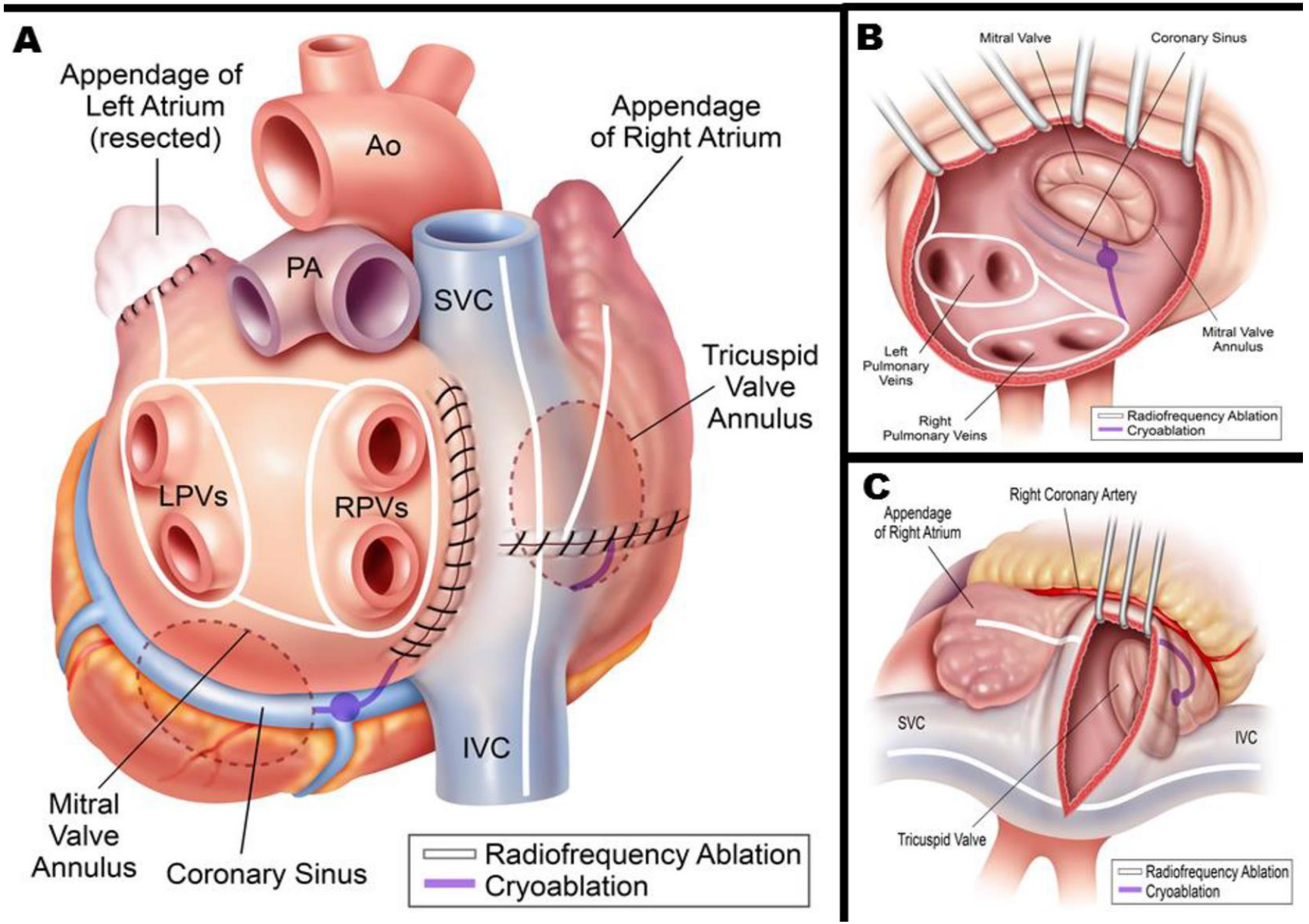
Faster recovery



Mazine et al. Canadian journal of cardiology 2016

Adjuncts to septal myectomy

Arrythmia surgery : Cox-Maze



Concomitant mitral valve disease

Around **7%** of patients with obstructive HCM undergoing myectomy have **intrinsic MV disease**

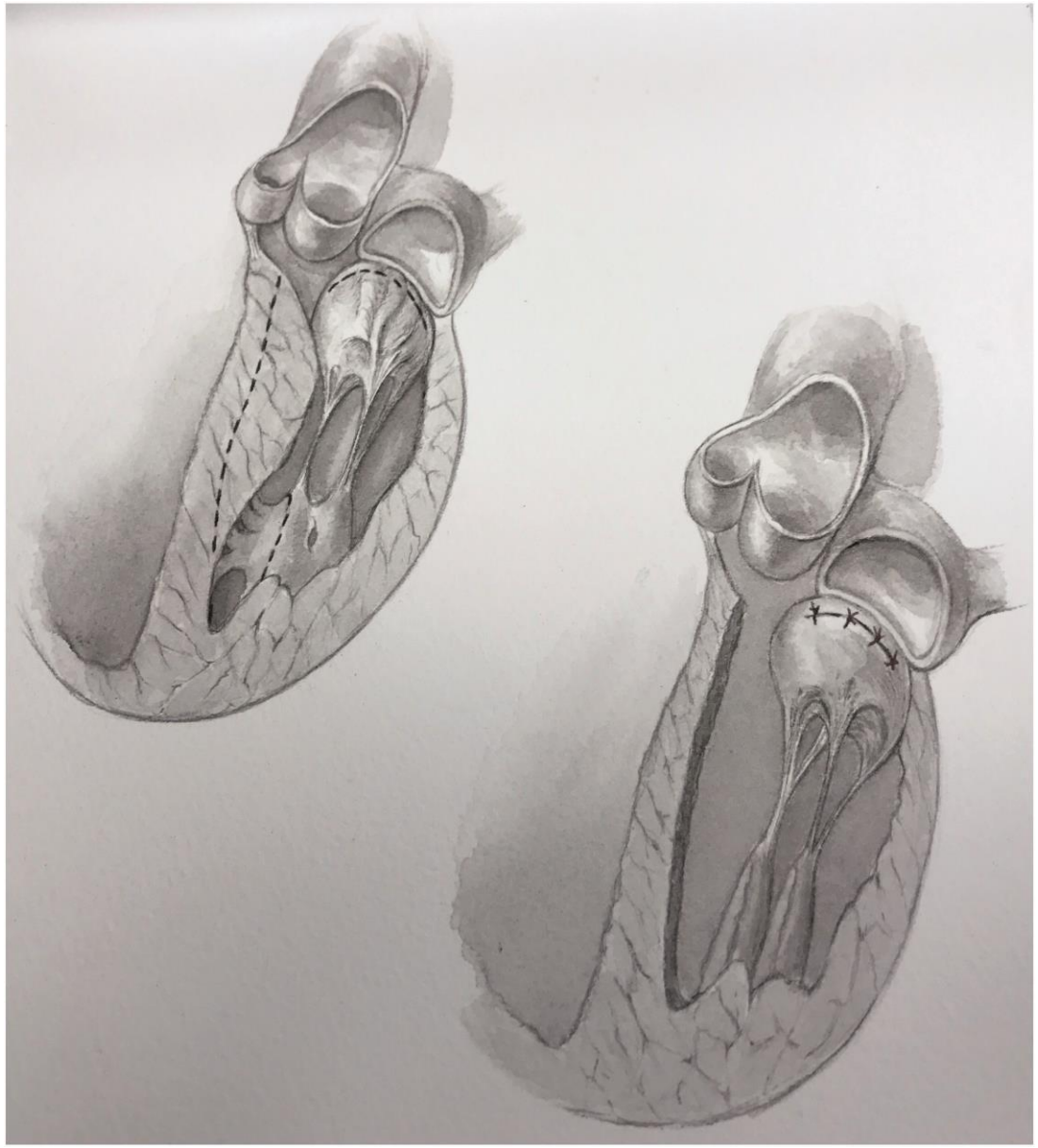
- myxomatous with prolapse/ruptured CT > MV Repair
- mixed stenosis/regurgitation > MV Replacement
- isolated MV replacement is not recommended
- Mitraclip? Inconsistent results

Concomitant mitral valve disease

Remodeling or repair of the Mitral valve apparatus and submitral structures :

- plication or pericardial patch to shorten an elongated AML ??
- resection of residual leaflet tissue ??
- Cutting of secondary fibrotic CT, accessory muscular and fibrous structures connecting PM to septum or free wall.

Myectomy and AML shortening



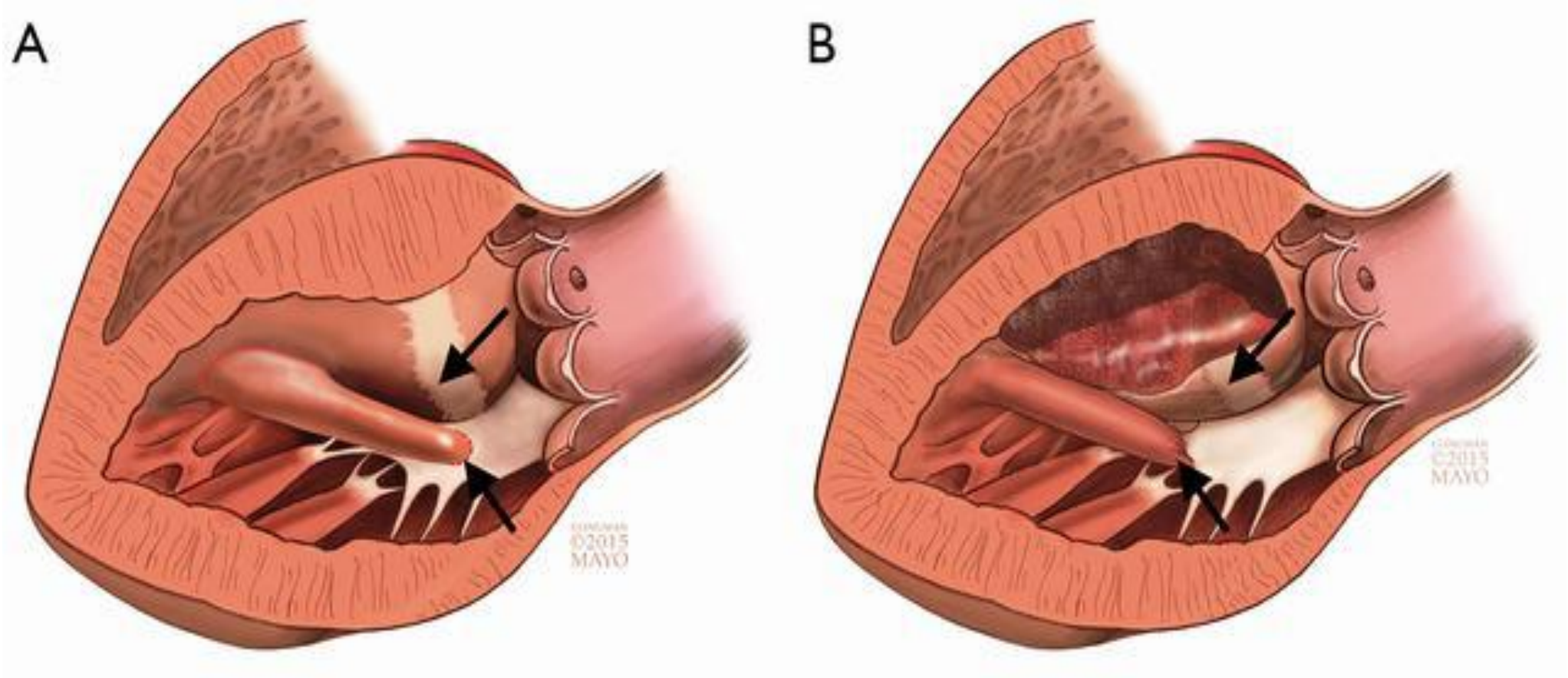
CENTRAL MESSAGE

MV leaflet lengths are increased in patients with obstructive HCM, but leaflet length is not associated with LVOT gradients before or after septal myectomy.

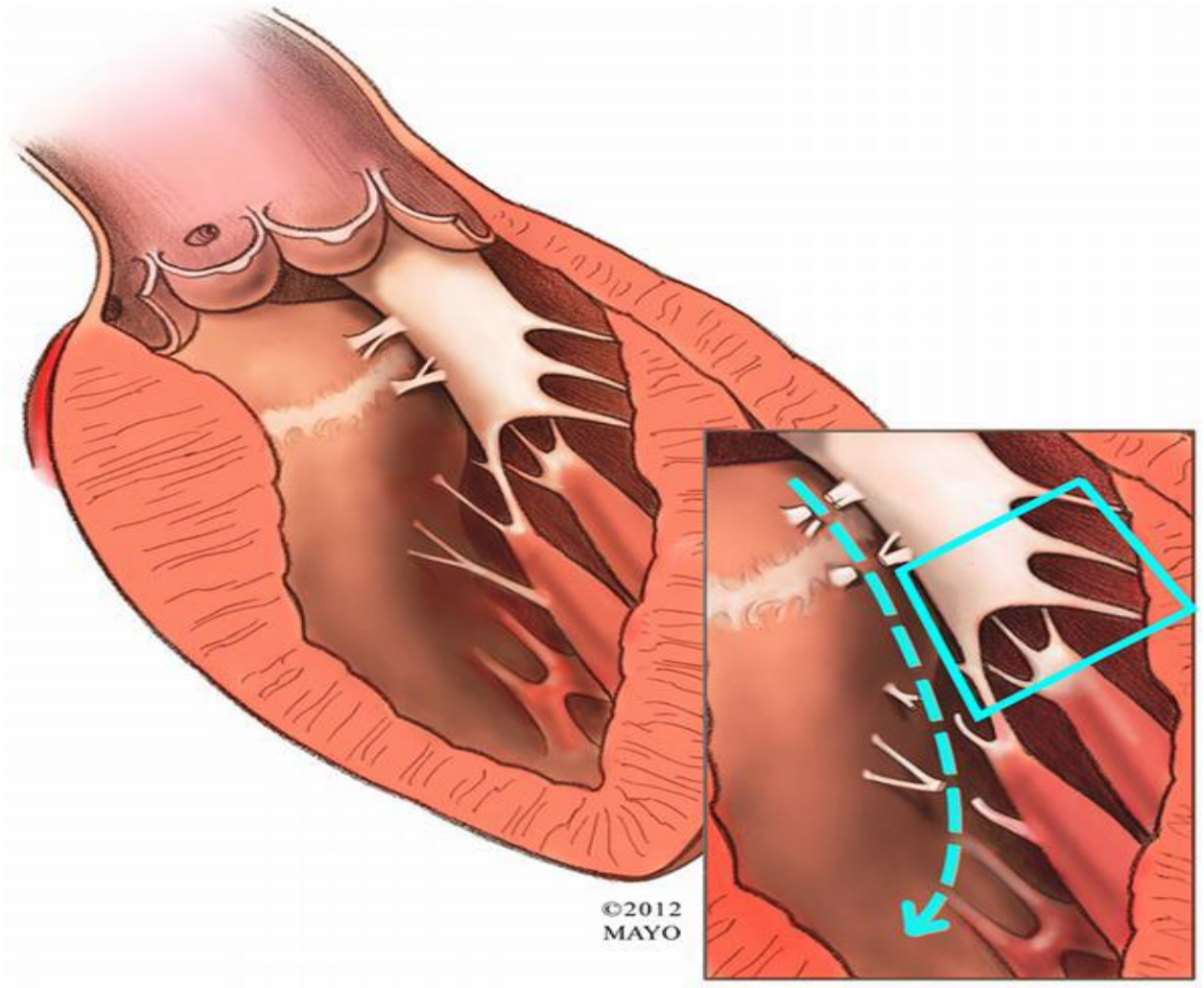
Additional MV procedures are unnecessary unless intrinsic MV disease is present (+/- 7% patients)

Carvahlo et al.; JTCVS 2021

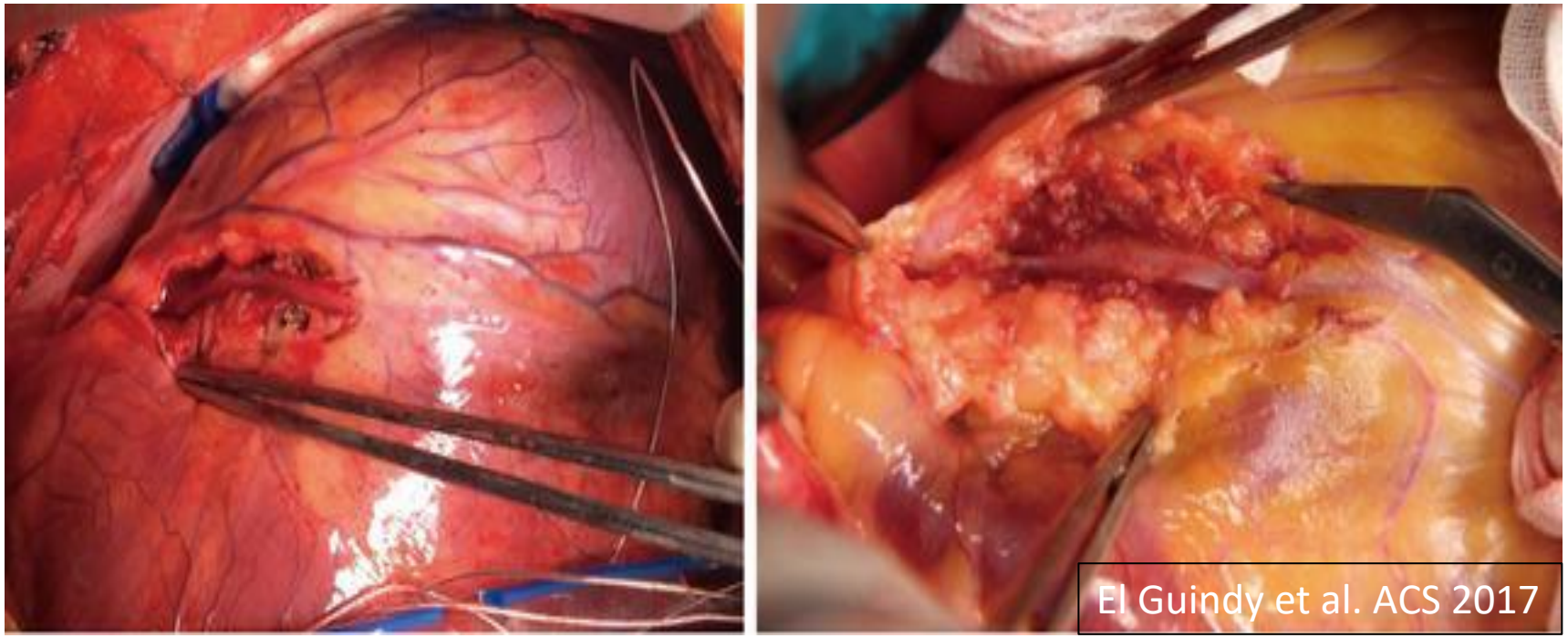
Papillary Muscle abnormal insertion on the Mitral valve lealfet



Fibrous attachment between AML and Septum

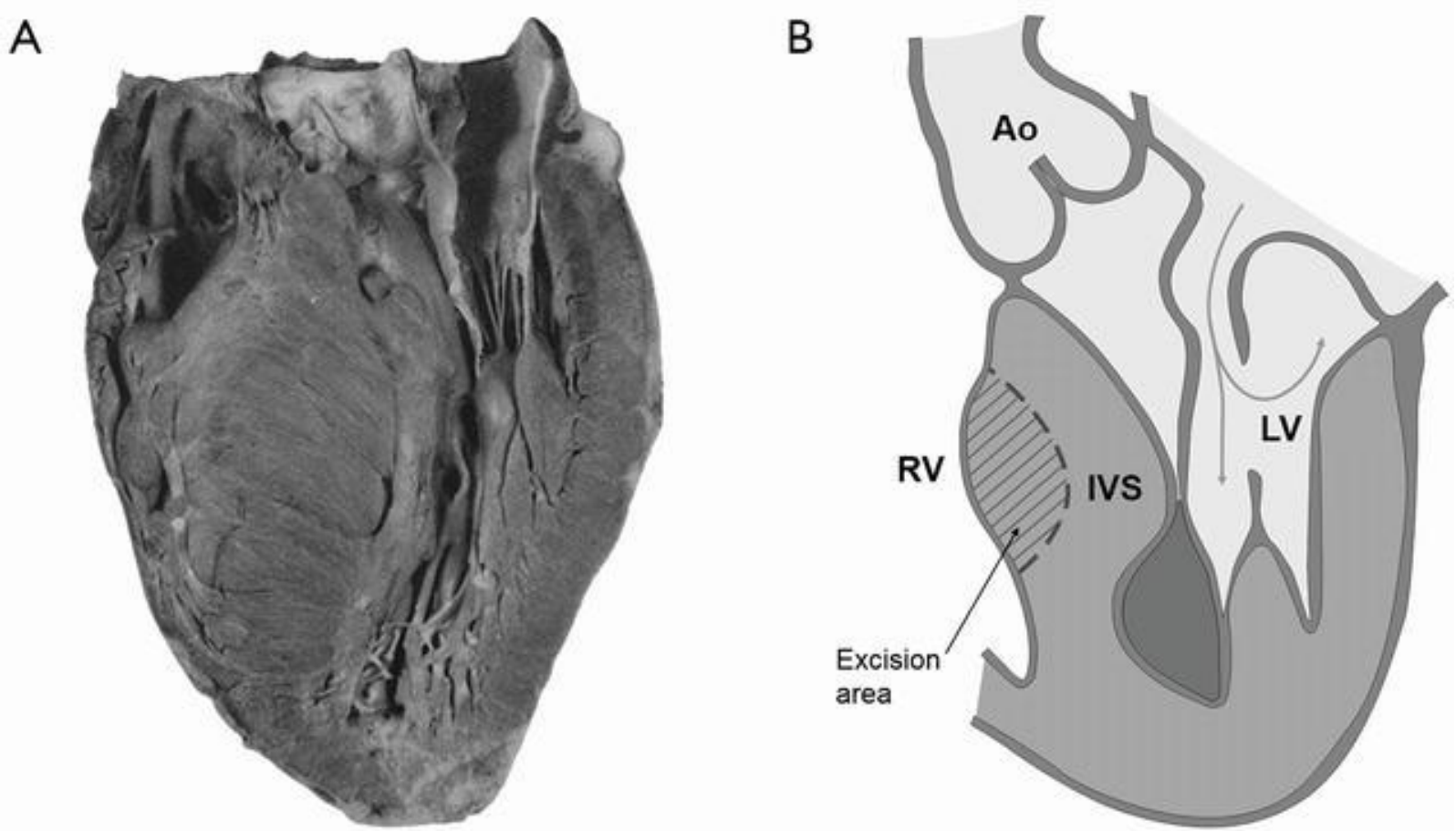


Myocardial Bridges

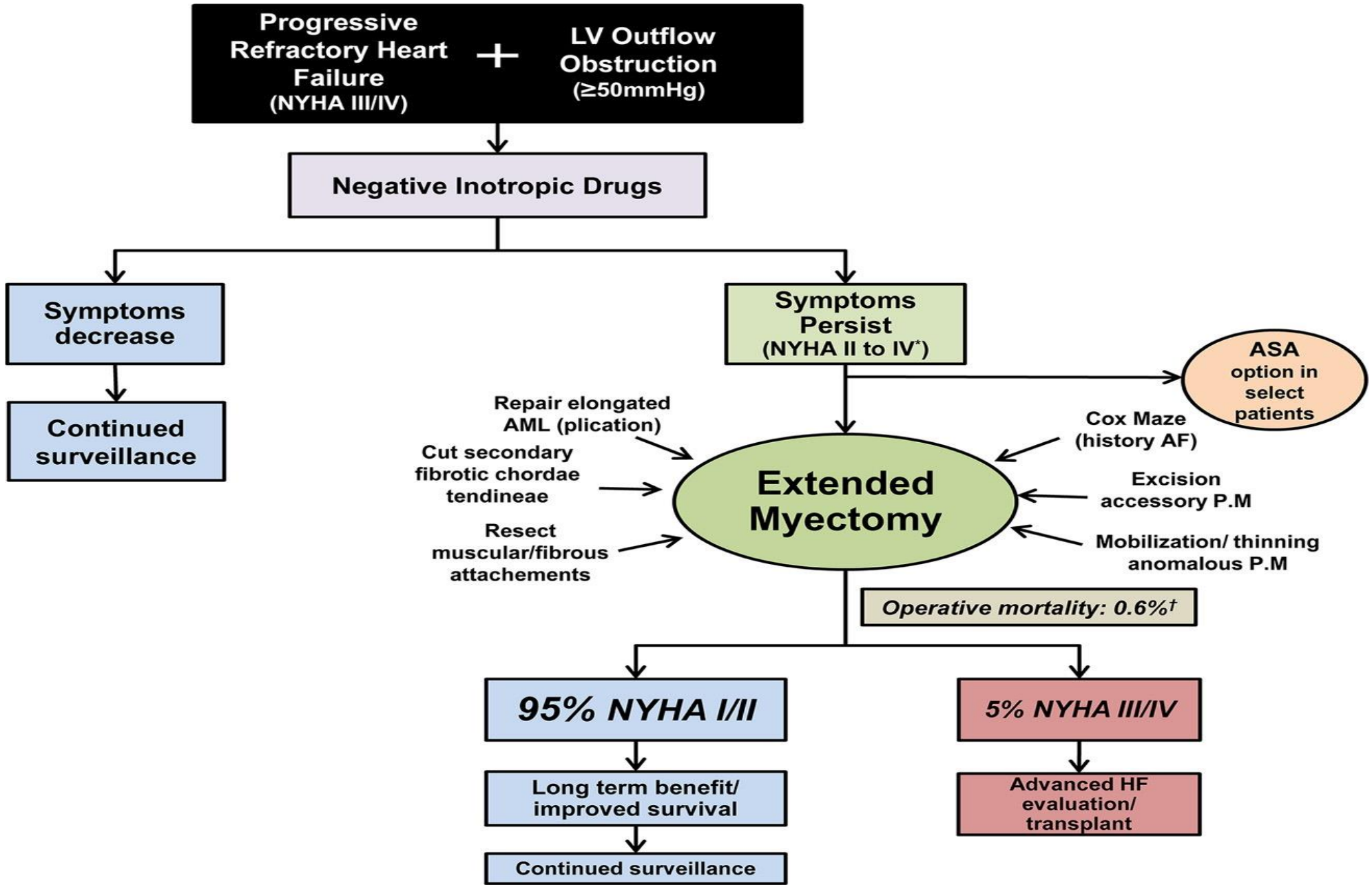


Detunneling of the LAD

Right ventricular HOCM



RESULTS


Progressive Refractory Heart Failure (NYHA III/IV)
+
LV Outflow Obstruction (≥50mmHg)
Negative Inotropic Drugs
Symptoms decrease
Continued surveillance
Symptoms Persist (NYHA II to IV*)
ASA option in select patients
Repair elongated AML (plication)
Cut secondary fibrotic chordae tendineae
Resect muscular/fibrous attachements
Extended Myectomy
Cox Maze (history AF)
Excision accessory P.M
Mobilization/ thinning anomalous P.M
Operative mortality: 0.6%†
95% NYHA I/II
Long term benefit/ improved survival
Continued surveillance
5% NYHA III/IV
Advanced HF evaluation/ transplant

RESULTS

Table 1
Tabulated data for surgical myectomy at 10 consortium HCM centers

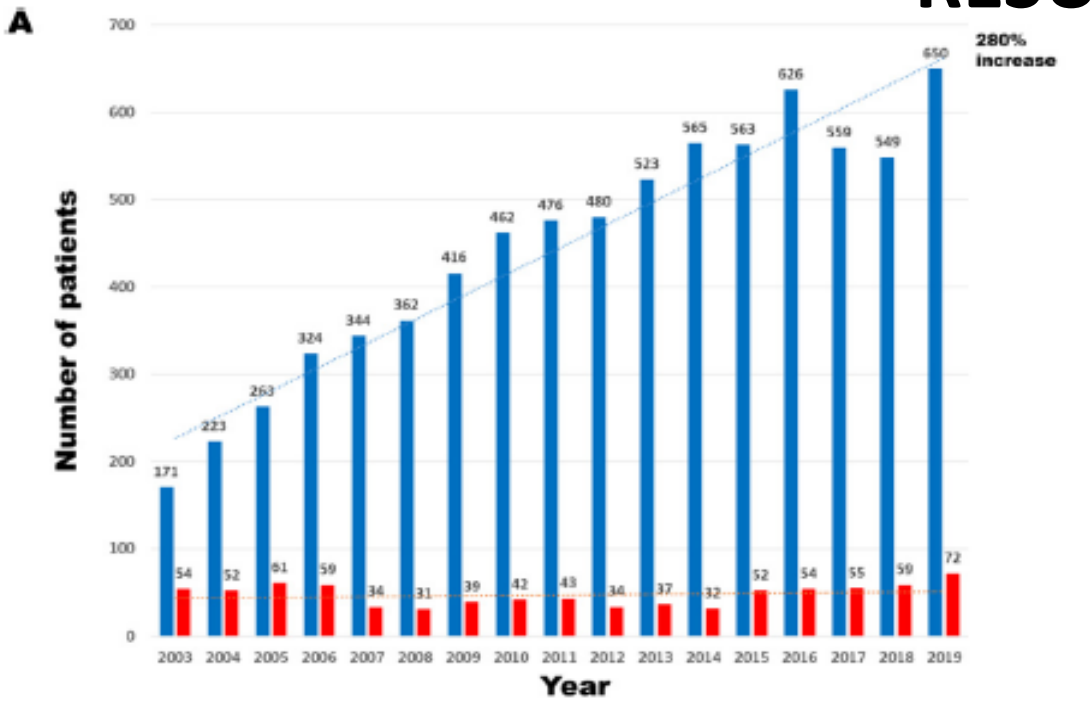
Variable	Total number myectomy	Age at myectomy (years)	Male	VSD	PPM†	MVR as sole operative strategy	Operative (30-day) mortality, n	Operative mortality
Clevland Clinic	2851	56 ± 14	53%	0.1%	4.1%	153*	21	0.7%
Mayo Clinic	2782	57 ± 16	54%	0.1%	6.6%	0	14	0.5%
Fuwai	2220	47 ± 15	60%	0.5%	0.9%	0	11	0.5%
Tufts	825	54 ± 15	54%	0.7%	6.0%	0	5	0.6%
Toronto	740	55 ± 14	61%	0.3%	6.6%	14	7	0.9%
Monza/Bergamo	665	53 ± 16	55%	0.2%	3.2%	0	3	0.4%
NYU	515	57 ± 13	51%	0.6%	2.7%	3	4	0.8%
UCLA	171	41 ± 29	50%	0	8.8%	24	1	0.6%
Barcelona	144	61 ± 14	45%	1%	4.8%	8	1	0.6%
Sydney	60	52 ± 18	46%	5%	15.0%	0	0	0
Total	10,973	54	55%	0.3%	4%	202 (1.8%)	67	0.6%

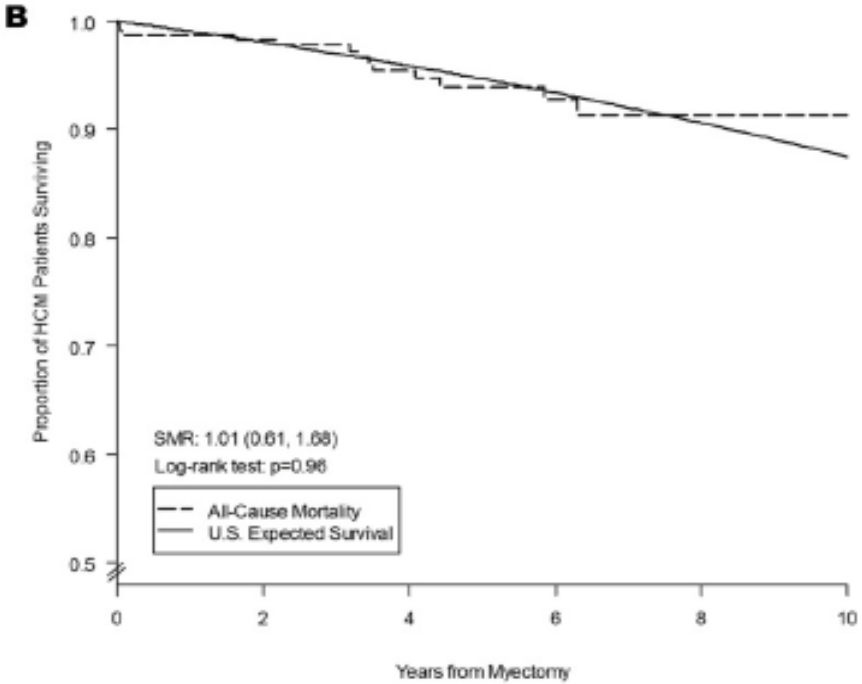
Abbreviations: MVR = mitral valve replacement; NYU: New York University; PPM= permanent pacemaker; UCLA: University of California Los Angeles; VSD: (iatrogenic) ventricular septal defect; Y: years

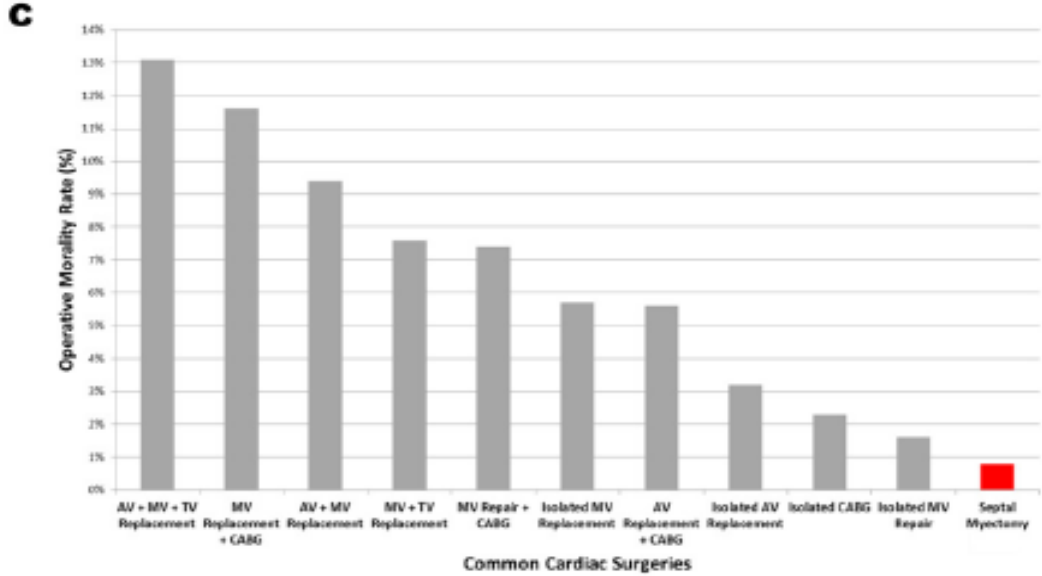
* predominantly in patients with mild anterior septal thickness (<18 mm) and LV outflow tract obstruction, usually after mitral valve repair unsuccessful in satisfactorily relieving outflow gradient with or without papillary muscle reorientation.

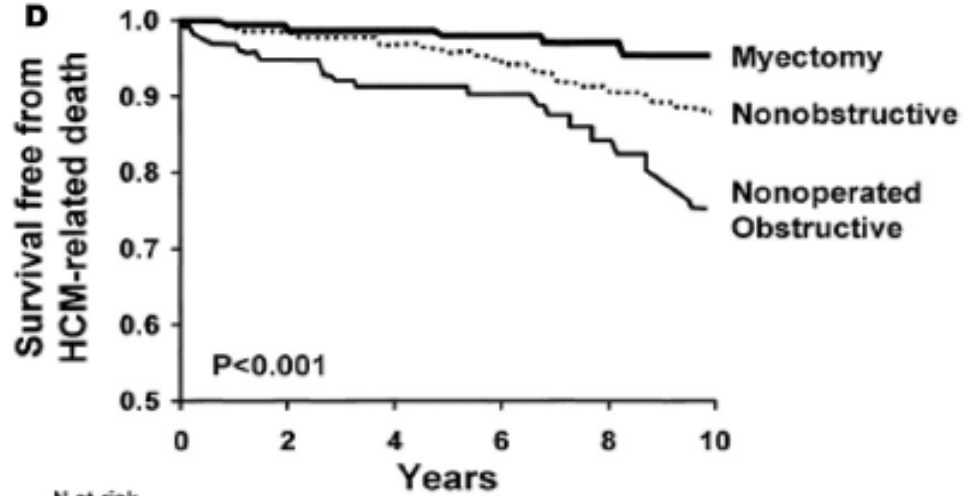
† can include patients with preoperative conduction disease.

RESULTS



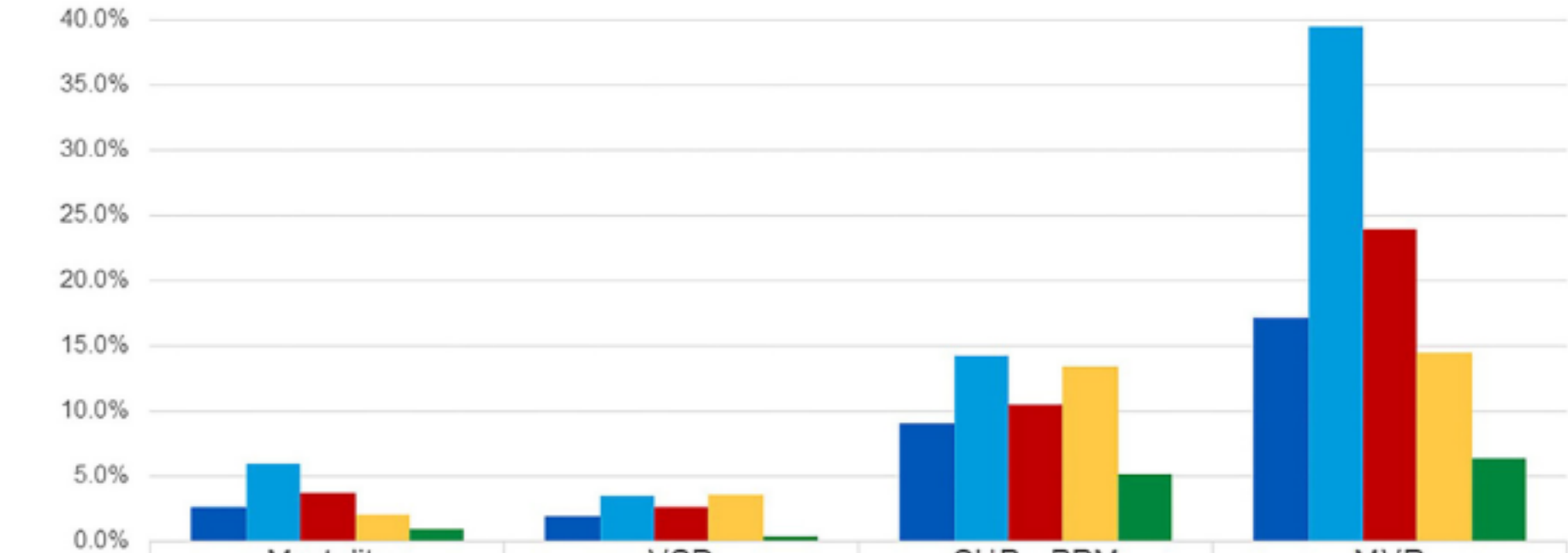






N at risk						
Myectomy	289	249	179	108	66	39
Nonobstructive	820	587	490	355	244	201
Nonoperated obstructive	228	146	106	69	42	28

RESULTS



	Mortality	VSD	CHB - PPM	MVR
Overall	2.6%	1.9%	9.0%	17.1%
<1	5.9%	3.5%	14.2%	39.5%
1-5	3.7%	2.6%	10.5%	23.9%
5-10	2.0%	3.6%	13.3%	14.5%
>10	0.9%	0.4%	5.1%	6.3%

National survey of Septal myectomy by yearly institutional volumes
(Holst et al from the STS database)

Alcohol Septal ablation (ASA)

HOCM: alcohol septal ablation

- High volume experienced centers
- Rapid recovery
- Morbi-Mortality similar to surgery
- less uniform and slower reduction in gradient
- high rate of complete heart block 10-15%
- inconsistent results (thickness & MV apparatus)
- Ventricular tachyarrythmias in susceptible patients

	Extended Septal Myectomy	Alcohol Septal Ablation
Pre-op evaluation	• Marked Septal hypertophy > 30 mm • Marked LVOT obstruction ≥ 50mmHg • Anatomic abnormalities involving the MV apparatus and PM	• Older (>60 yo) patients • Poor surgical candidates with multiple co-morbidities • Adequate distribution of septal perforator arteries
Procedures	• Open heart myocardial excision +/- release of abnormal septal connections, MV annuloplasty	• Percutaneous ethanol injection through septal perforator arteries for myocardial scar formation
Limitations	• Risk of RBBB, Iatrogenic VSD • Experienced centers • Follow-up	• Higher risk of arrhythmias and SCD when compared to surgery • Higher rates of symptom recurrence and re-operation
Advantages	• Long-term LVOT gradient reduction • Low re-operation rates • Low short and long term mortality rates at experienced centers	• Widely available procedure • Short recovery period

Solomon et al. Seminar Th 2018

Conclusions

- Exended septal myectomy is a **safe and effective** therapy for HOCM
- Best outcomes are observed at **centers with interdisciplinary expertise (≥ 10 procedures/y)**
- **Evolution of operatives techniques** (better understanding of pathophysiology) > combined approach, minimally invasive approaches, …..

Thank you for your attention

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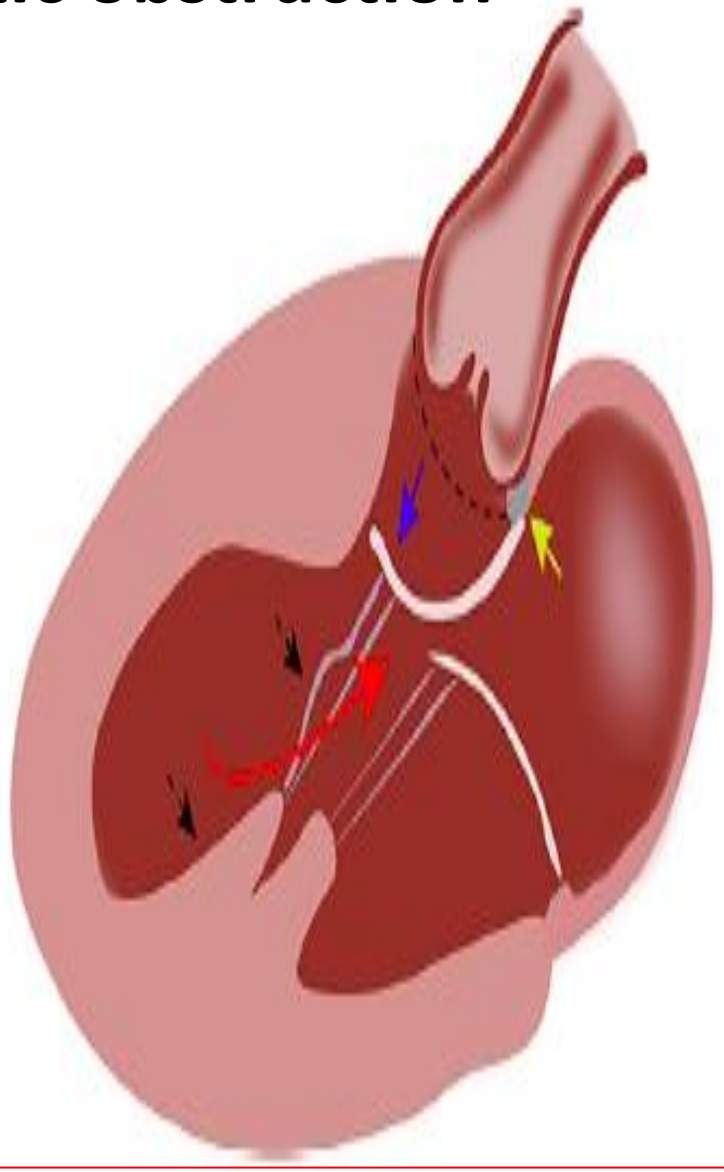
Pathophysiology subaortic obstruction

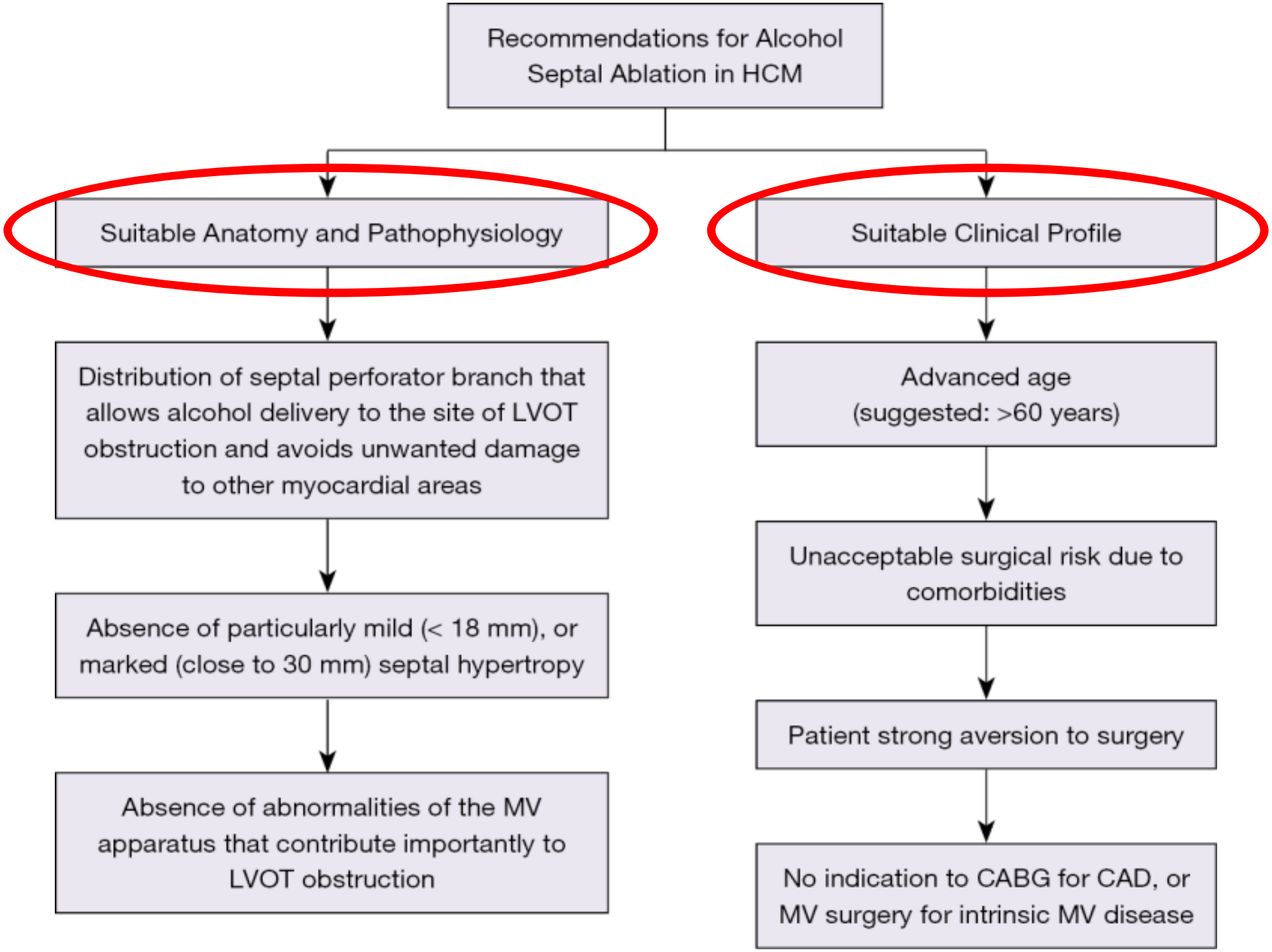
SAM
Elongated anterior leaflet (+/-)
Mitral regurgitation

Chordae tendinae

Papillary Muscle



Diastolic dysfunction, myocardial ischemia, arrythmia, metabolic & energetic abnormalities


Recommendations for Alcohol Septal Ablation in HCM
Suitable Anatomy and Pathophysiology
Distribution of septal perforator branch that allows alcohol delivery to the site of LVOT obstruction and avoids unwanted damage to other myocardial areas
Absence of particularly mild (< 18 mm), or marked (close to 30 mm) septal hypertropy
Absence of abnormalities of the MV apparatus that contribute importantly to LVOT obstruction
Suitable Clinical Profile
Advanced age (suggested: >60 years)
Unacceptable surgical risk due to comorbidities
Patient strong aversion to surgery
No indication to CABG for CAD, or MV surgery for intrinsic MV disease

HOCM: alcohol septal ablation

- **Anatomy**: suitable septal perforator coronary anatomy and absence of structural abnormalities of mitral apparatus

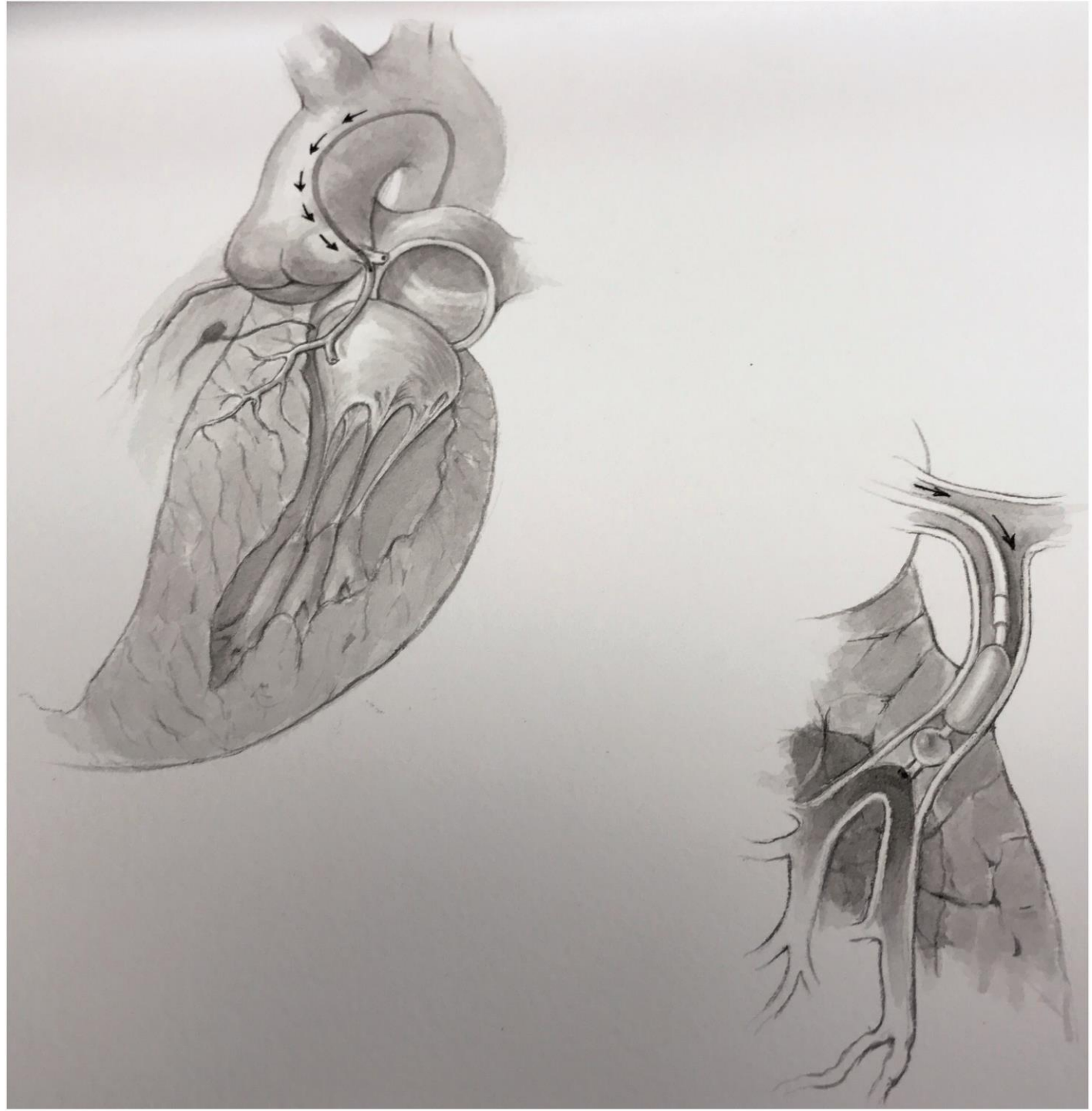
- **Clinical profile**: advanced age
  important comorbidities
  aversion to open-heart surgery

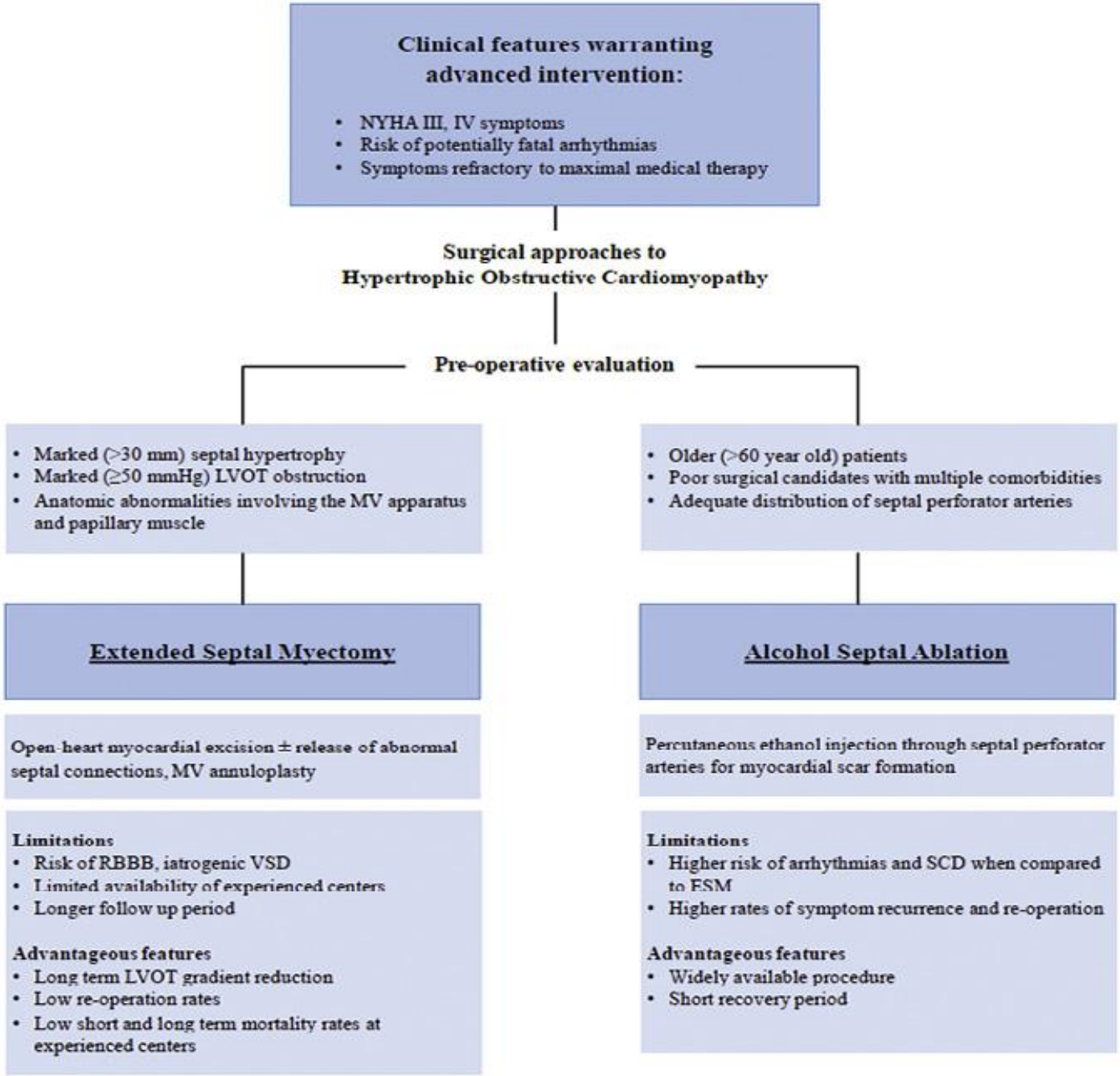
- **Contraindications**: children and young adults, other cardiac abnormalities requiring operation (valve, coronary, subaortic membrane, etc….).

MERCI POUR VOTRE ATTENTION

HOCM: Pharmacological strategies

- Most of the time: objective is palliation, mitigation and control of HF symptoms.
- Established drugs: Negative inotropic drugs (BB, verapamil, disopyramide)
- Newer drugs: Mavacamten, genetic therapy?




Clinical features warranting advanced intervention:
• NYHA III, IV symptoms
• Risk of potentially fatal arrhythmias
• Symptoms refractory to maximal medical therapy
Surgical approaches to Hypertrophic Obstructive Cardiomyopathy
Pre-operative evaluation
• Marked (>30 mm) septal hypertrophy
• Marked (≥50 mmHg) LVOT obstruction
• Anatomic abnormalities involving the MV apparatus and papillary muscle
• Older (>60 year old) patients
• Poor surgical candidates with multiple comorbidities
• Adequate distribution of septal perforator arteries
Extended Septal Myectomy
Alcohol Septal Ablation
Open-heart myocardial excision ± release of abnormal septal connections, MV annuloplasty
Percutaneous ethanol injection through septal perforator arteries for myocardial scar formation
Limitations
• Risk of RBBB, iatrogenic VSD
• Limited availability of experienced centers
• Longer follow up period
Advantageous features
• Long term LVOT gradient reduction
• Low re-operation rates
• Low short and long term mortality rates at experienced centers
Limitations
• Higher risk of arrhythmias and SCD when compared to ESM
• Higher rates of symptom recurrence and re-operation
Advantageous features
• Widely available procedure
• Short recovery period