
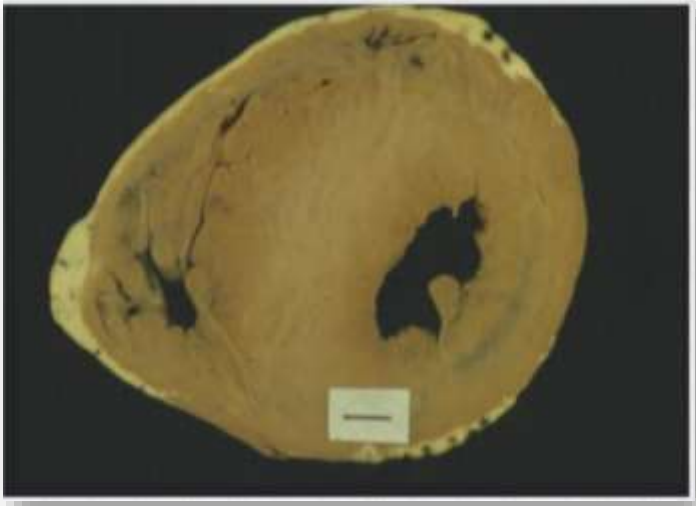


**Surgical or Catheter Based
Intervention Management
of
Hypertrophic Cardiomyopathy**

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2017



Hypertrophic Cardiomyopathy



Historical Perspective

- **HCM was initially described by Teare in 1958:**
 - Found massive hypertrophy of ventricular septum in small cohort of young patients who died suddenly
- **Braunwald 1960s.** was the first to diagnose HCM clinically.
- **Many names for the disease:**
 - Idiopathic hypertrophic sub-aortic stenosis (IHSS).
 - Muscle sub-aortic stenosis.
 - Hypertrophic obstructive cardiomyopathy (HOCM).

Background

- **Prevalence of HCM:** 1:500 to 1:1000 individuals
 - This occurrence is higher than previously thought, suggesting a large number of affected but undiagnosed people
- Men and African-Americans affected by almost 2:1 ratio over women and Caucasians.
- Global disease with most cases reported from USA, Canada, Western Europe, Israel, & Asia.

HCM: Definitions

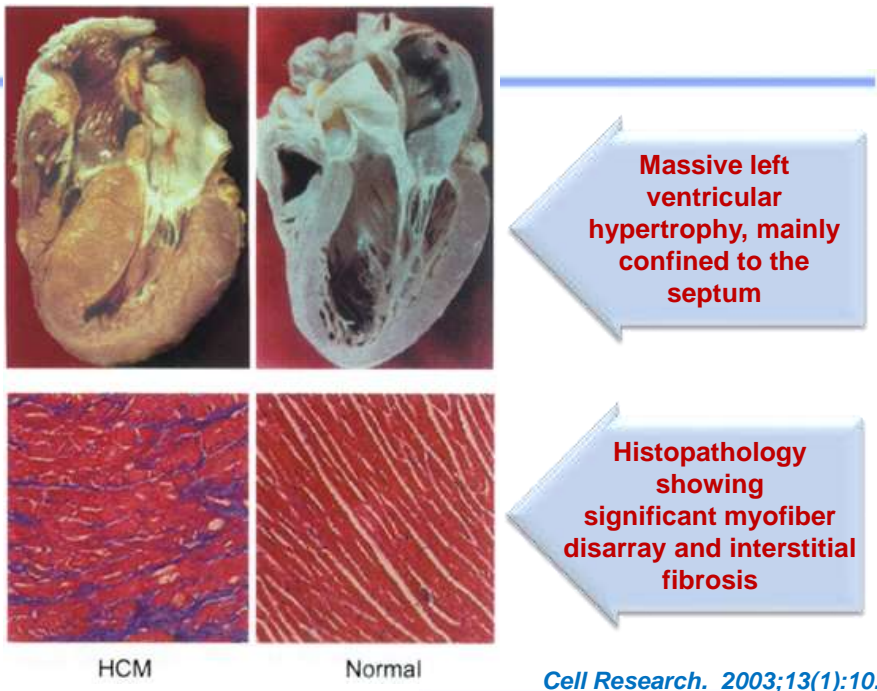
Increased left ventricular wall thickness not solely explained by abnormal loading conditions:

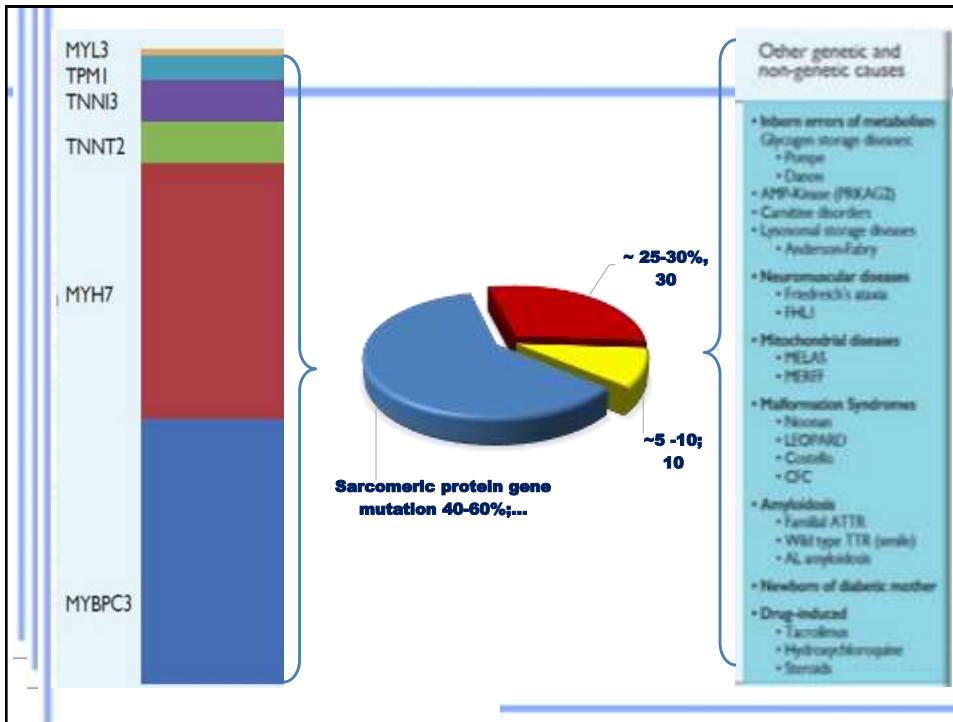
Adults:

- LV wall thickness ≥ 15 mm in one or more LV myocardial segments measured by any imaging technique.


Children:

- LV wall thickness more than two standard deviations above the predicted mean (z-score > 2).





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FASTTRACK CLINICAL RESEARCH

A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy (HCM Risk-SCD)

Constantinos O'Mahony¹, Fatima Jichi², Menelaos Pavlou³, Lorenzo Monserrat³,
 Aristides Anastasakis⁴, Claudio Rapezzi⁵, Elena Biagini⁵, Juan Ramon Gimeno⁴,
 Giuseppe Limongelli⁷, William J. McKenna¹, Rumana Z. Omar^{2,8} and Perry M. Elliott^{1*},
 for the Hypertrophic Cardiomyopathy Outcomes Investigators

O'Mahony, C et al. Eur Heart J. 2013 Oct. 14 [Epub ahead of print]

HCM Risk-SCD: Predictor Variables

Predictor Variable
Age at evaluation.
History of sudden cardiac death in one 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).
Maximal wall thickness: the greatest thickness in the anterior septum, posterior septum, lateral wall, and posterior wall of the LV, measured at the level of the mitral valve, papillary muscles and apex using parasternal short-axis plane using 3-D echocardiography.
Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane.
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 were determined using the modified Bernoulli equation: Gradient = $4V^2$, where V is the peak aortic outflow velocity.
NSVT: ≥ 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.
History of unexplained syncope at or prior to evaluation.

O'Mahony, C et al. *Eur Heart J.* 2014 Aug. 7; 35 (30):2010-20

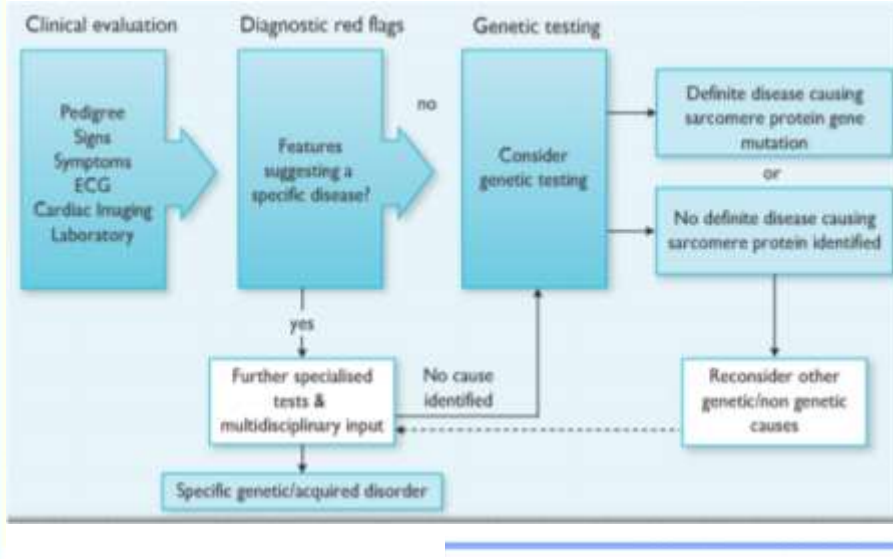
HCM Risk-SCD model for predicting 5 year risk

$$\text{Probability}_{\text{SCD at 5 years}} = 1 - 0.998^{\text{exp}(\text{prognostic index})}$$

where Prognostic index = $[0.15939858 \times \text{maximal wall thickness (mm)}] - [0.00294271 \times \text{maximal wall thickness}^2 \text{ (mm}^2)] + [0.0259082 \times \text{left atrial diameter (mm)}] + [0.00446131 \times \text{maximal (rest/Valsalva) left ventricular outflow tract gradient (mmHg)}] + [0.4583082 \times \text{family history SCD}] + [0.82639195 \times \text{NSVT}] + [0.71650361 \times \text{unexplained syncope}] - [0.01799934 \times \text{age at clinical evaluation (years)}]$.

O'Mahony, C et al. *Eur Heart J.* 2014 Aug. 7; 35 (30):2010-20

General Approach to The Diagnosis of Hypertrophic Cardiomyopathy



Assessment of Symptoms

Chest pain
Dyspnoea
Fatigue
Palpitations

LVOT obstruction

Arrhythmia

LV Diastolic failure

LV Systolic failure

Valve Disease

Microvascular
 dysfunction

Syncope

LVOT obstruction

Arrhythmia

Abnormal Vascular
 Function

Treatment strategies in HCM are three-fold:

- 1) Medical therapy in patients with mild to moderate symptoms,
- 2) **Reduction of the myocardial septum by surgery or alcohol ablation in patients with severe symptoms or significant outflow tract obstruction, and**
- 3) Implantation of an automatic cardioverter-defibrillator in the presence of non-sustained ventricular tachyarrhythmias.

Surgical Myectomy

1. Drug-refractory heart failure symptoms.
2. NYHA Classes III and IV.
3. **LV outflow obstruction:**
 - Rest - gradient ≥ 30 mm Hg.
 - Physiologic exercise - gradient ≥ 50 mm Hg.

Transaortic resection of muscle from the proximal to midseptal region. (Up to PM)

Operative mortality < 1 percent.

Maintain long-lasting improvement in symptoms and exercise capacity.

Mortality may be improved after septal myectomy.

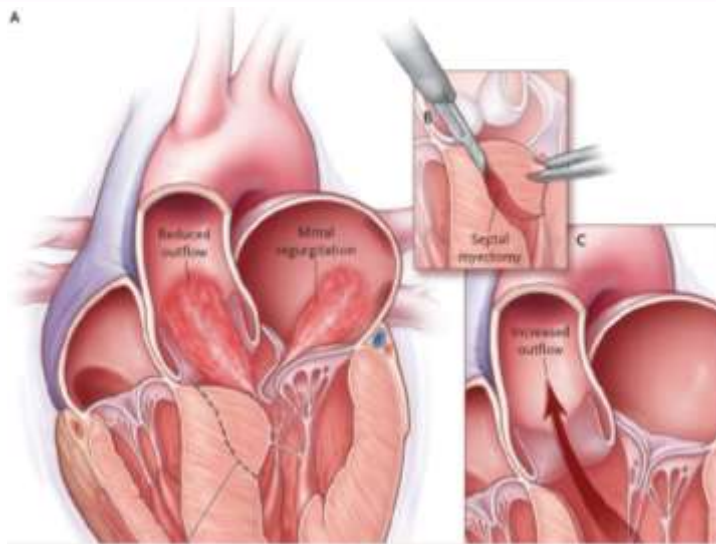
Septal Myectomy

- In symptomatic patients despite medical Rx.
- With gradient > 50 mmHg at rest or provocation.
- After aortic cross-clamping and aortotomy, bar of myocardium is excised from proximal septum.
- Complications include total AV block and VSD.

The surgical treatment of HOCM has been reserved for patients with comorbidities such as organic mitral regurgitation or severe coronary artery disease requiring coronary bypass grafting, although simultaneous treatment of coronary artery disease by percutaneous interventions with stent implantation and alcohol ablation of the septum has been performed in patients with HCM.

However, dilation of the left ventricle after surgical myectomy and the occurrence of aortic regurgitation after transvalvular reduction of the myocardium have been reported as potential complication of this procedure.

Surgical Septal Myectomy



Nishimura RA et al. NEJM. 2004. 350(13):1320.

1- Surgery

Septal myotomy/myectomy:

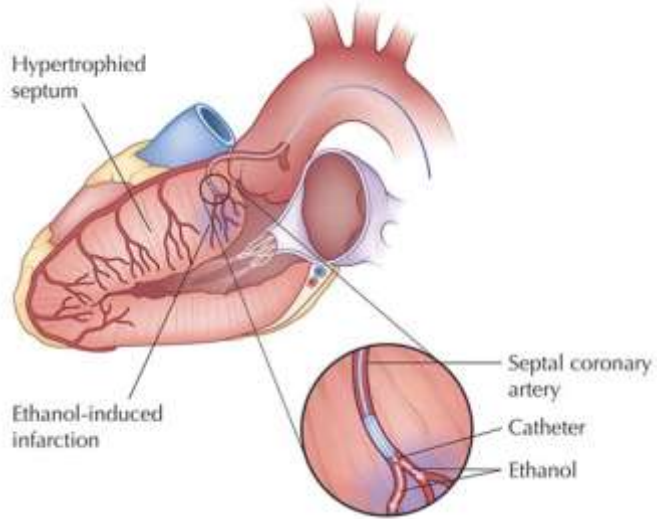
- **Patients < 40 years:** mortality < 1 %
- **Patients > 65 years:** mortality 10-15 %
- Survival better than medically treated patients
- **Should be considered in:** resting gradient > 50 mmHg, or refractory to medical Rx.
- Young patients, particularly those with severe disease
- Additional structural abnormalities affecting the mitral valve or coronary arteries.
- **Complication (rare):** Aortic incompetence

Myotomy / Myectomy

Results of Myotomy/Myectomy at the Mayo Clinic for Hypertrophic Cardiomyopathy

- **Mortality < 5% overall:**
 - Patients younger than 40 years, < 1%.
 - Higher in older patients with other disease.
- Substantial decrease in gradient.
- Substantial decrease in mitral regurgitation.

Alcohol Septal Ablation



Braunwald. *Atlas of Heart Diseases: Cardiomyopathies, Myocarditis, and Pericardial Disease. 1998.*

Alcohol Septal Ablation



Source: Foster V, O'Rourke RA, Walsh RA, Royle-Wilson P. *Hurst's The Heart, 12th Edition* <http://www.accessmedicine.com>

Before



Source: Foster V, O'Rourke RA, Walsh RA, Royle-Wilson P. *Hurst's The Heart, 12th Edition* <http://www.accessmedicine.com>

After

Alcohol Septal Ablation

- Outflow tract gradient is reduced from a mean of 60 to 70 mm of mercury often to < 20 mm of mercury.
- 80–85 % symptomatic improvement.
- **Complications:**
 - Complete heart block < 10 %.
 - Coronary dissections.
 - Large myocardial infarctions.
 - Ventricular septal defects.
 - Myocardial perforations.
 - Ventricular fibrillation.

Alcohol Septal Ablation

- Successful short-term outcomes
 - LVOT gradient reduced from a mean of 60-70. mmHg to < 20 mmHg.
 - Symptomatic improvements, increased exercise tolerance.
- Long-term data not available yet.
- **Complications:**
 - Complete heart block.
 - Large myocardial infarctions.
- No randomized efficacy trials yet for alcohol septal ablation vs. surgical myectomy.

Catheter-based treatment for HOCM has received much attention and has become the treatment of choice.

Surgery is an effective treatment strategy but is associated with considerably higher perioperative morbidity.

Surgical myectomy was the treatment of choice for the past 40 years, whereas alcohol ablation of the septum is the treatment of choice for patients with HOCM in the 21st century.

Outcome of Alcohol Septal Ablation for Obstructive Hypertrophic Cardiomyopathy

Paul Sorajja, MD; Uma Valeti, MD; Rick A. Nishimura, MD; Steve R. Ommen, MD; Charanjit S. Rihal, MD; Bernard J. Gersh, MB, ChB, DPhil; David O. Hodge, MS; Hartzell V. Schaff, MD; David R. Holmes, Jr, MD

Table 4. Thirty-Day Clinical Events

New permanent pacemaker dependency, n (%)	28 (20.4)
Cardiac tamponade, n (%)	5 (3.5)
Urgent or emergent cardiac surgery, n (%)	2 (1.3)
Stroke, n (%)	1 (0.7)
Sustained ventricular tachycardia, n (%)	1 (0.7)
Resuscitated sudden cardiac arrest, n (%)	2 (1.3)
Death resulting from progressive heart failure, n (%)	1 (0.7)
Sudden death, n (%)	1 (0.7)
All deaths, n (%)	2 (1.4)

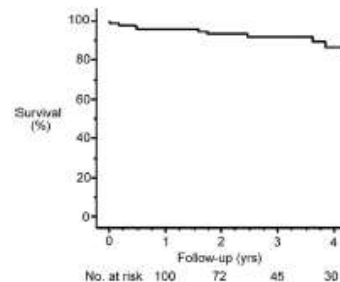


Figure 3. Overall survival after septal ablation.

Circulation. 2008; 118(2): 131-9.

Updated Meta-Analysis of Septal Alcohol Ablation Versus Myectomy for Hypertrophic Cardiomyopathy

Shikhar Agarwal, MD, MPH,* E. Murat Tuzcu, MD,† Milind Y. Desai, MD,†
Nicholas Smedira, MD,‡ Harry M. Lever, MD,† Bruce W. Lytle, MD,‡ Samir R. Kapadia, MD,†
Cleveland, Ohio

Conclusion

SA does seem to show promise in treatment of HOCM owing to *similar mortality rates as well as functional status* compared with SM; however, the caveat is *increased conduction abnormalities and a higher post-intervention LVOTG*. The choice of treatment strategy should be made after a thorough discussion of the procedures with the individual patient.

(*J Am Coll Cardiol* 2010;55:823–34)

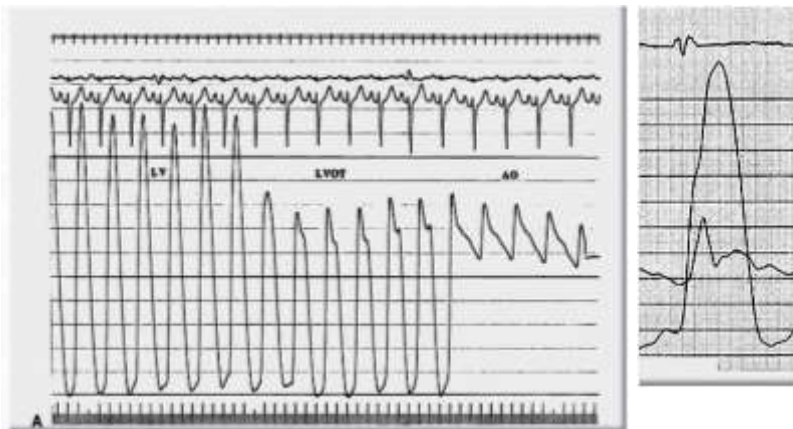
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DDD Pacing

- Objective measurements of exercise capacity did not differ significantly.
- Overall decrease in outflow tract gradient (25 to 40 percent of baseline).
- Role of dual-chamber pacing - patients at high risk for other therapeutic modalities.
- **Candidates for dual-chamber pacing:**
 - Significant bradycardia in which pacing may allow an increased dosage of medication.
 - Patients who need ICD as a primary treatment.

LV Outflow Tract Obstruction



Elliott & McKenna: Textbook of Cardiology (Topol ed.)

Dual-Chamber Pacing

- Proposed benefit: pacing the RV apex will decrease the outflow tract gradient.
- Several RCTs have found that the improvement in subjective measures provided by dual-chamber pacing is likely a placebo effect.
- Objective measures such as exercise capacity and oxygen consumption are not improved.
- No correlation has been found between pacing and reduction of LVOT gradient.

Efficacy of Therapeutic Strategies

Table 3. Comparative Features of Septal-Reduction Therapies.

Therapy	Mortality	Residual Gradient	Effectiveness	Follow-up	Complications	Time to Resolution of Gradient	
	%	mm Hg	% of Patients	yr	Type	% of Patients	
Dual-chamber pacing	<1	<40	10–40	10	Infection or perforation	<2	4 wk
Septal myectomy*	<2–3	<10	>90	>10	Complete heart block Ventricular septal defect Aortic regurgitation	<1 <1 <1	Immediate
Septal ablation†	<2–3	<20	70–80	<5	Complete heart block Ventricular septal defect Large myocardial infarction	10–40 Unknown Unknown	8–12 wk

* Surgical septal myectomy is the only intervention that can treat concomitant problems, such as multivessel coronary disease, intrinsic mitral-valve disease, midventricular obstruction, and fixed subaortic obstruction.

† The true rates of death and complications may be underestimated, since complications may occur at a higher frequency in the inexperienced centers and may be underreported.

Nishimura et al. *NEJM*. 2004. 350 (13):1323.

It is clear that answering the question whether alcohol ablation or surgical myectomy is the treatment of choice would require a randomized trial comparing these two treatment.

However, for the invasive cardiologist who is doing alcohol ablation, it is clear that the percutaneous approach is much easier and is successful in many patients who would be hard to enroll in a randomized trial. Thus, it appears obvious to us that alcohol ablation is the treatment of choice for the new century.