Surgical Management of Hypertrophic Obstructive Cardiomyopathy

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**Hypertrophic cardiomyopathy**

Autosomal-dominant inherited disease characterized by:
- increased cardiac mass,
- arrhythmias,
- sudden death.

caused by mutations in any 1 of 10 encoding proteins of cardiac sarcomere

i.e., primary sarcomere disorder.
marked increase in thickness of walls of heart in patient with HCM.
Nomenclature

- HCM known by a confusing array of names such as:
  - Hypertrophic obstructive cardiomyopathy [HOCM]
  - Idiopathic hypertrophic subaortic stenosis [IHSS]
  - Asymmetric septal hypertrophy

_Hypertrophic cardiomyopathy_ is the preferred name because it describes overall disease spectrum without introducing misleading inferences such that LVOT obstruction is invariable feature.

- HCM is predominantly a non-obstructive disease (75% of patients do not have a sizable resting outflow tract gradient).
Pathophysiology

Triad:
- ↓ ventricular compliance in diastole
- ↑ contractility of left ventricle in systole
- presence of outflow obstruction

Diastolic dysfunction may result in reduction in coronary blood flow ± angina

Gradient in the ventricular cavity is variable depending on pre and after load and contractility
Pathophysiology

- Mechanism of obstruction is
  - hypertrophied septum
  - SAM
- Mitral valve
  - AML 1 cm longer
  - PML 0.5 cm longer
  - Coaptation point is 1 cm from the tip of AML
- Sequence: eject-obstruct-leak
Other phenotypic manifestations

Several phenotypic manifestations of HCM do not primarily involve sarcomeric proteins, including:

- myocardial bridges
- mitral valve malformations (such as elongated leaflets or direct papillary muscle insertion into mitral valve).
Myocardial bridging in a patient with hypertrophic cardiomyopathy.

LAD during diastole (A) & systole (B).

Bridging of middle LAD (black arrows on B) is evident during systole associated with complete systolic obliteration of 2nd septal perforator artery, which is visible only during diastole. There also was mild systolic compression of distal LAD.
R. Levine 1995
Systolic anterior motion (SAM) of MV leaflets

- Subaortic obstruction is exaggerated by SAM of MV leaflets & mid-systolic contact with ventricular septum.

- SAM is also responsible for concomitant MR:
  - usually mild-to-moderate
  - due to incomplete leaflet apposition
    {typically directed posteriorly into LA}
HCM in systole.
Mitral leaflet is distorted toward septum (SAM, *black arrow*) resulting in LVOT obstruction and posteriorly directed MR.
principal pathways of disease progression in hypertrophic cardiomyopathy (HCM)

Treatment options

- Medical (drug) therapy.
- Chronic dual-chamber pacing
- Alcohol septal ablation technique
- Surgical therapy:
  1. Morrow procedure & its modifications
  2. Septal myomectomy through ventriculotomy
  3. MV surgery
  4. Modified KONO
  5. Lt ventricle to aortic conduit
- Heart Transplantation.
General indications

Factors involved in the decision of mode of therapy:

- Availability of experienced surgeon to do myotomy-myectomy
- Concomitant medical conditions
- Advanced age
- Prior cardiac surgery
- Availability of experienced interventional cardiologist or EP
Doctor Andrew Morrow, a pioneer of myectomy wrote

"the incisions are made quite close to the seat of the soul"
ventricular septal myotomy-myectomy (Morrow procedure)

Surgery for obstructive HCM, evolved over past 4 decades, :

- from ventricular septal myotomy (i.e., without muscular resection),

  to classic Morrow myectomy

  to Extended septal myomectomy

Morrow’s modification of initial procedures, the wider more extensive trough myectomy, remained the standard operation.

Rectangular trough is created by:
- first making 2 parallel longitudinal incisions in basal septum.
- Incisions extended distally & then transversely connected proximally below aortic valve & distally just beyond level of mitral-septal contact & subaortic obstruction
The trough myectomy of Morrow

Myotomy in hypertrophied septum extended toward lumen

Retractor protects anterior leaflet mitral valve and papillary muscle

Thickened endocardium and mitral leaflet

A (left), Classic left ventricular septal myectomy for hypertrophic obstructive cardiomyopathy.
B (right), Extended left ventricular septal myectomy for anomalous papillary muscle with direct insertion into anterior mitral leaflet and also fusion to the septum.
Perioperative results of myectomy

Operative mortality = 0% to 2%
performed at experienced surgical centers in absence of associated conditions

Postoperative morbidity:
- permanent pacemaker for HB 0% to 10%
- VSD = 0% to 2%
Age and gender matched pop.

Clinical and Echocardiographic Determinants of Long-Term Survival After Surgical Myectomy in Obstructive Hypertrophic Cardiomyopathy

Woo, Anna MD, SM; Williams, William G. MD; Choi, Richard MD; Wigle, E Douglas MD; Rozenblyum, Evelyn; Fedwick, Katie; Siu, Samuel MD, SM; Ralph-Edwards, Anthony MD; Rakowski, Harry MD

Circulation
Volume 111(16), 26 April 2005, pp 2033-2041

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[Original Articles: Cardiovascular Surgery]

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Long-Term Effects of Surgical Septal Myectomy on Survival in Patients With Obstructive Hypertrophic Cardiomyopathy

Total and HCM-related mortality compared in 3 subgroups comprised of 1,337 consecutive HCM patients evaluated from 1983 to 2001:
1) surgical myectomy (n = 289);
2) LVOT obstruction without operation (n = 228);
3) non-obstructive without operation (n = 820).

Mean follow-up duration was 6 ± 6 years.

Ommen, S. R  J Am Coll Cardiol, 2005; 46:470-476,
Operative deaths = 0.8%

1-, 5-, and 10-year overall survival after myectomy 98%, 96%, and 83%, respectively,

this overall survival did not differ from that of general U.S. population matched for age and gender (p = 0.2) nor from patients with nonobstructive HCM (p = 0.8).

Multivariate analysis showed:
myectomy to have a strong, independent association with survival (hazard ratio 0.43; p < 0.001).

Ommen, S. R  J Am Coll Cardiol, 2005; 46:470-476,
Survival free from all-cause mortality after surgical myectomy for OHCM (n = 289) compared with age- & gender-matched general U.S. white population.

Survival free from all-cause mortality in 3 HCM patient subgroups: surgical myectomy (n = 289), nonoperated with obstruction (n = 228), and nonobstructive (n = 820)

Survival free from sudden cardiac death among patients in 3 HCM subgroups: surgical myectomy (n = 289), nonoperated with obstruction (n = 228), and nonobstructive (n = 820)

Survival free from HCM-related death among patients in 3 HCM subgroups: surgical myectomy (n = 289), nonoperated with obstruction (n = 228), and nonobstructive (n = 820)

Mechanisms for Recurrent Left Ventricular Outflow Tract Obstruction After Septal Myectomy for Obstructive Hypertrophic Cardiomyopathy

- 610 septal myectomies performed from 1975 to July 2003.

- 13 of these were repeat myectomies after classic myectomies performed (6 at Mayo) (7 elsewhere).

- Interval between initial myectomy & repeat myectomy ranged from 13 months to 11 years (mean, 5.0 ± 3.7 years).

Mechanisms for Recurrent Left Ventricular Outflow Tract Obstruction After Septal Myectomy for Obstructive Hypertrophic Cardiomyopathy

Mechanisms for obstruction included:
- limited myectomy at initial myectomy \( (n = 11), \)
- septal hypertrophy at midventricular level \( (n = 8), \)
- anomalous papillary muscles \( (n = 3). \)

- no early deaths.
- No MVR performed
- Mean follow-up = 5.8 ± 5.8 years:
  - one late death.
  - All surviving patients were:
    - free from recurrence of outflow tract obstruction
    - NYHA functional class I or II.
Independent abnormalities intrinsic to mitral valve (e.g., myxomatous degeneration, mitral leaflet fibrosis, or anomalous papillary muscle insertion) should be suspected:

- when MR jet is directed centrally
- or anteriorly into LA,
- or if multiple jets are present.
Mitral Valve Replacement

Indications

1- unfavorable septal morphology.

2- intrinsic mitral valve disease accounting for severe MR (such as myxomatous degeneration).

3- mid-cavity obstruction due to anomalous insertion of papillary muscle into AML;

4- mild septal hypertrophy, which suggests that:
   - muscular resection would be associated with high risk of septal perforation
   - or inadequate hemodynamic result.
MV repair

- AML extension
- Resection-Plication-Release (RPR) Repair
- AML plication (horizontal, longitudinal)
- Edge to edge repair
Patch extension of AML
Patch extension of AML
Longitudinal plication of AML

Horizontal plication of AML to reduce leaflet length & leaflet/chordal slack

Effects of dual-chamber pacing in HCM to be less favorable and possibly harmful?

Recent and more carefully controlled investigations have found:

- In a randomized, double-blind, crossover study, average decrease in outflow gradient was small (only about 25 percent).

- Subjective symptomatic improvement reported with similar frequency by patients after two to three months of pacing and after the same period without pacing.

- Moreover, two other studies shown that a decrease in outflow gradient produced by temporary AV sequential pacing may be associated with detrimental effects on ventricular filling and COP.
Alcohol ablation or surgery??

- In contrast to that for surgery, postprocedural follow-up for alcohol septal ablation is relatively brief (about 3-5 years compared with 40 years for myotomy-myectomy).

- Recent comparative analysis showed surgery to be superior to ablation in reducing resting and provocable gradients.
HOCM: comparison of outcome after myectomy and alcohol ablation

- Alcohol ablation = 60
- Surgical myectomy = 95

Treatment summary

1. Avoidance of dehydration, alcohol, & isometric or anaerobic exercise.
2. Pharmacologic therapy with maximally tolerated doses of 1 or more negative inotropic agents.
3. Direct relief of obstruction through septal debulking should be considered.
4. For isolated basal septal hypertrophy, either surgical myectomy (±MVR, MV repair) or catheter-based septal ablation can be used.
5. Dual-chamber pacing can be considered in patients with severe comorbidities or other extenuating circumstances that unacceptably increase risk of other procedures.
THANK YOU