Management of HOCM: Non-Surgical Options

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Case

- 69 yo with HOCM
- Class III DOE – cannot walk more than 1 block (Stage C HF)
- Medications
  - Apixaban 5 bid
  - Metoprolol tartrate 50 qd
  - Verapamil ER 120 qd
- Exam: III/VI SEM that increases with Valsalva
- PH:
  - Stroke due to AF 2015, residual tremor, writing difficulty
- Echo
Echo

- Dynamic gradient of 62 mmHg at rest which increases to 116 mmHg with Valsalva maneuver
- Severe SAM
- Mod-severe posteriorly directed eccentric MR
What would you recommend to this 69 yo with HOCM and MR?

1. Septal myomectomy
2. Septal myomectomy and MV repair (Alfieri)
3. Septal myomectomy and MV replacement
4. Alcohol septal ablation
5. Alcohol septal ablation and MitraClip
6. Further medical management
7. Permanent Pacemaker
Success with Alcohol Septal Ablation is All About Choosing the Right Patient

- HCM with symptoms
  - Non-cardiac contraindication to β blocker
    - Side-effect
    - 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
Mavacamten

- Oral small molecule inhibitor of cardiac myosin ATPase
- Produces dose-dependent decrease in contractility in Pioneer-HCM (phase 2):

Global Phase 3 Trial Currently Being Planned (EXPLORER-HCM)
Characterization of a Patient with HOCM Must Combine Echo and Cath (hemodynamic and anatomic) Data

- Echo
  - Systolic anterior motion of the anterior mitral leaflet
  - Ventricular dimensions, wall thicknesses, contractile state
  - Outflow gradient location
  - Diastolic function
  - Outflow tract dimensions and geometry
  - Mitral valve anatomy, regurgitation severity and pattern
Catheterization Technique in HCOM

- Simultaneous LV-sheath pressure
- End-hole catheter for LV pressure
- Resting gradient
- Provoke gradient
  - PVC
  - Nitrates
  - Inotropic stimulation
- Beware catheter entrapment artifact
- Assess coronary and septal anatomy
Pressure Waveforms - Fixed Obstruction vs HOCM

**HOCM**
- **Peak gradient in late systole**
- **Spike and dome**
- **Rapid early upstroke**
- **Late secondary peak**

**AS**
- **Peak gradient in early systole**
- **Slow upstroke**
- **Late single peak**
Determine the Location of LVOT Obstruction

- VALVULAR
- CLASSIC HOCM
- MID VENTRICULAR
Intrinsic MV pathology is frequent in HOCM

Degenerative with thin leaflets, excessive mobility, torn chords to P2

Myxomatous with thick, redundant leaflets

Large anteromedial pap muscle inserting in A1

Restrictive chords restricting and tenting ant leaflet

Restrictive leaflet with MAC

Long leaflets

*Kaple et al, Ann Thorac Surg 2008;85:1527*
Importance of MR in HOCM (Yu et al, JACC 2000;36:2219)

• Direction of MR jet can be clue to mechanism: posterior is typical of SAM-related MR
• Severity of MR correlates with LVOT gradient in patients without independent MV disease
• In these patients, septal myectomy reduced the severity in direct proportion to the reduction in gradient

Typical post jet  Ant jet due to post prolapse  Central jet due to mitral stenosis
Case 3 Continued

- Over ~10 mos, she underwent up titration of her Metoprolol from 50 to 175 bid, remaining on verap
- No improvement in her symptoms
- Echo showed similar gradients (148 mmHg at rest, 183 with Valsalva) and worsening of her MR to mod-severe

- Cardiac surgical and interventional cardiology consultations were obtained and she opted to proceed with cath, possible alcohol septal ablation as a first step.
CLASS IIb

1. Alcohol septal ablation, when performed in experienced centers, may be considered as an alternative to surgical myectomy for eligible adult patients with HCM with severe drug-refractory symptoms and LVOT obstruction when, after a balanced and thorough discussion, the patient expresses a preference for septal ablation (153,273,278,280,281). (Level of Evidence: B)

CLASS III: HARM

5. Alcohol septal ablation should not be done in patients with HCM who are less than 21 years of age and is discouraged in adults less than 40 years of age if myectomy is a viable option. (Level of Evidence: C)
Surgery vs Alcohol Ablation

Advantages of Alcohol Ablation:
- Shorter hospital stay and recovery time
- Less expensive
- Doesn’t preclude surgery

Advantages of Surgical Myectomy:
- Lower incidence of CHB requiring PPM
- Proven long-term (>20yr) efficacy (data n/a for alcohol ablation)
- No risk of coronary dissection and minimal risk of myocardial damage remote from the septum
- Ability to tx concomitant problems
- More immediate and complete relief of obstruction
Larger Meta-analysis (Leonardi et al, CCI 2010;3:97-104)

- 19 studies (4000 pts)
- ASA pts older, less LVH, shorter f/u

<table>
<thead>
<tr>
<th></th>
<th>ASA (%)</th>
<th>Myomectomy</th>
<th>95% CI</th>
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<tbody>
<tr>
<td><strong>All-Cause Mortality</strong></td>
<td>2.1</td>
<td>1.8</td>
<td>NS</td>
</tr>
<tr>
<td><strong>Adjusted</strong></td>
<td>0.28 (0.16-0.46) favoring ASA</td>
<td>0.32 (0.11-0.97) favoring ASA</td>
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• 1197 pts
• 369 young (mean age 42)
• Young c/w older patients had:
  • Lower 30 day mortality
  • Lower PPM rates
  • Excellent long-term survival

Liebregts et al, JACC Intv 2017;10:1134
Fifer, JACC Intv 2017;10:1144
Superior septal instrumented, agitated saline injection followed by 1.5 cc alcohol over 2 mins with transient CHB
Recurrent transient 2nd degree AVB on day 2 prompted placement of a dual chamber PPM, discharged on day #4
1 Month Follow up: NYHA Class II, feeling better, MR resolved, SR/not pacing!