Hypertrophic Cardiomyopathy
Anatomy, Hemodynamics, and Prognosis

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29 yo Man with Class III DOE
IV/VI Murmur Despite Triple Rx

Severe HCM with SAM

Stage 2 DD

105 mmHg
29 Year Old Man with HCM
100 mmHg Resting LVOT Gradient

Pre-myectomy

Post-myectomy
29 Year Old Man with HCM

*What Got Cut Out.....*
One Year Later

Class 1-2 Symptoms

No further SAM, LVOT open, no MR

LVOT gradient 9 mmHg post amyl
3D LVOT Area in 11 HOCM Patients Pre- and Post Myectomy

$A_{LVOT}$ significantly increased after myectomy

Qin et al. Am J Cardiol 2004; 94: 964-6
Hypertrophic Cardiomyopathy

• **Subtypes**
  – Apical hypertrophy (Yamaguchi’s cardiomyopathy)
  – Upper septal hypertrophy
  – Displaced/abnormal papillary muscles

• **Phenotypic manifestations**
  – LVOT obstruction (resting or inducible)
  – Mitral regurgitation
  – Ventricular fibrillation
  – Diastolic dysfunction

• **Therapeutic interventions**
  – Negative inotropes
  – RV pacing
  – Defibrillator implantation
  – Surgical myectomy
  – Alcohol septal ablation
  – Papillary muscle realignment
Apical Hypertrophy
Yamaguchi’s Syndrome

Apical > basal hypertrophy
Apical Hypertrophy
Yamaguchi’s Syndrome

Exaggerated coronary flow
Upper Septal Hypertrophy

Systolic anterior motion of the mitral valve (SAM) with LVOT obstruction
18yo Gene+ Man with LVOT Obstruction

Septum 1.1 cm
Anteriorly displaced papillary muscles

LVOT gradient
Rest 6 mmHg
Amyl 115 mmHg
7.9 METs 118 mmHg

Kwon et al. JASE 2009; 22: e5-e6
Abnormal papillary muscle morphology is independently associated with increased left ventricular outflow tract obstruction in hypertrophic cardiomyopathy

Kwon et al. Heart 2008; 94: 1295-1301
Resting LVOT Gradient Mainly Depends on Papillary Muscle Morphology

Kwon et al. Heart 2008; 94: 1295-1301
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Hypertrophic Cardiomyopathy

32 yo Woman with Progressive DOE

7 m/sec 196 mmHg
Predictor of LVOT Gradient in HCM

3D Assessment of LVOT Area

y = 115x^{-1.7}

r = 0.92

n=15

J Am Coll Cardiol 2002; 39:308-14
Both Age and LVOT Gradient Predict Progression to CHF (NYHA Class III or IV)

No Progression to NYHA Class III or IV and Death from Heart Failure or Stroke (%)

<table>
<thead>
<tr>
<th>Years after Gradient Measurement</th>
<th>No obstruction, &lt;40 yr of age</th>
<th>Obstruction, &lt;40 yr of age</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>100</td>
<td>98</td>
</tr>
<tr>
<td>2</td>
<td>88</td>
<td>85</td>
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<tr>
<td>4</td>
<td>76</td>
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<td>6</td>
<td>64</td>
<td>60</td>
</tr>
<tr>
<td>8</td>
<td>52</td>
<td>48</td>
</tr>
<tr>
<td>10</td>
<td>40</td>
<td>36</td>
</tr>
</tbody>
</table>

Overall P<0.001

No. at Risk

| No obstruction, <40 yr of age | 349  251  206  146  103  80 |
| No obstruction, 40 yr of age  | 421  306  258  188  128  108 |
| Obstruction, <40 yr of age    | 106  70   52   37   21   15  |
| Obstruction, 40 yr of age     | 118  74   51   29   18   10  |

Maron et al. NEJM 2003; 348;295-303
Hypertrophic Cardiomyopathy

*Induction of LVOT Obstruction*

- **Vasodilator**
  - *Amyl nitrite, nitroprusside*
- **Exercise**
  - *Most physiologic but challenging to get maximum*
- **Inotropic stimulation**
  - *Isoproterenol, dobutamine*
  - *Easiest stressor in the OR*
- **Post-extrasystolic beat augmentation**
  - *Best in cath lab*
- **To fully exclude inducible obstruction requires both amyl and exercise**
33 yo man with exertional dyspnea and LVOT murmur
Resting SAM present but gradient only 16 mmHg, 21 mmHg post-amyl
33 yo man with exertional dyspnea and LVOT murmur

Developed marked dyspnea at 6 METs of exercise, now with 119 mmHg LVOT gradient

Provocation of Gradient

57 patients

• 18% only with amyl
• 11% only with exercise
• 26% with both
• 46% with neither

Marwick et al. AJC 1995; 75: 805-9
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Mitral regurgitation in HCM occurs when SAM pulls the anterior leaflet away from the posterior one.

At rest, mild SAM, 1+ MR

Post exercise, severe SAM, 4+ MR
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This Can Really Ruin your Day......

Ventricular fibrillation 3 days following alcohol septal ablation
Hypertrophic Cardiomyopathy

Risk Factors for Sudden Death

- Prior history of VT/VF
- Family history of sudden death
- History of syncope
- Septal thickness > 30 mm
- cTnT genotype
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Septal LV Hypertrophy

- LVOT obstruction
- Mitral regurgitation
- Hyperdynamic systolic function
- Secondary LVH
- Impaired diastolic relaxation
- Stiff ventricle
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Severe Mitral and Aortic Regurgitation

Volume Overload Causing LV Dilation

Underwent successful AVR and MV repair

Smooth post-op course but dyspnea after several months, with loud murmur noted........
LVOT Obstruction After AVR/MVr
62 Year-Old Man

Pre-op
LV: 7.3/4.0 cm
Walls: 1.7/1.4 cm

8 months post-op
LV: 4.0/2.6 cm
Walls: 2.8/1.9 cm

Marked reverse remodeling
β-Blocker

β- + Ca^{++} + Norpace

8.5 m/sec = 289 mmHg
(Ohio indoor record...)

43 mmHg
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# Impact of DDD Pacing on Functional Status

<table>
<thead>
<tr>
<th>NYHA-FC</th>
<th>Baseline</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>0</td>
<td>21</td>
</tr>
<tr>
<td>II</td>
<td>0</td>
<td>17</td>
</tr>
<tr>
<td>III</td>
<td>29</td>
<td>6</td>
</tr>
<tr>
<td>IV</td>
<td>15</td>
<td>0</td>
</tr>
</tbody>
</table>

Fananapazir, Circulation 1992; 85: 2149
Impact of RV Pacing on Invasive Parameters of Systolic and Diastolic Function

- 29 patients studied in cath lab
- Millar catheter studies with and without RV pacing
- With pacing, LVOT gradient fell only 73 to 61 mmHg
- Significant slowing in relaxation and rise in LA pressure

Nishimura, JACC 1996; 28: 1226

- 21 patients randomized to RV pacing in cross-over study
- Gradient fell 76 to 55 mmHg with pacing
- No difference in Quality of Life score or exercise capacity
- RV pacing not routinely recommended

Nishimura, JACC 1996; 28: 1226
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Defibrillators in HCM

• 128 pts. (69 < 41yo, 69% male) mean followup 3.1yrs
  – 43 post arrest, 85 for syncope, family history, septum>30 mmHg

• 29 pts with 1 or more appropriate discharge
  – 19/43 post-arrest, 10/85 prophylactic implants (5 syncope, 2 with thick septums, 2 with VT on monitor, 1 with family history)

• 25/29 patients were at rest, 4/29 with exercise

• Discharge rate
  – Post-arrest: 11%/year
  – Prophylactic: 5%/year

Maron, NEJM 2000; 342: 365-73
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Myectomy for HCM

- Treatment gold standard
- >40 years experience
- Symptom/gradient improvement
  - (immediate, consistent, permanent)
- Valve and CAD managed
- Indicated for Class III-IV patients with resting or inducible gradients > 50 mmHg
- But mortality has been reported as high as 16% in elderly patients
CCF Isolated Myectomy Experience
1994 - 2005

- 325 pts, 50 ±14 years, 53% male
- Mean IVS 23±5 mm, LVOT gradient 68±43 mmHg
- Results:
  - In hospital mortality: 0/325
  - 22/325 required pacers
  - 10 needed HCM-related reoperations
  - Survival at 3.6±2.8 years was 90%, equivalent to general population

Smedira NG et al. ATS 2008; 85: 127-33
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Septal Ablation for HCM

- Sigwart reported 3 pts, 1995
- Alcohol injected into 1st septal perforator to create a “localized myocardial infarction”
- Advantages of nonsurgical procedure
  - Faster recovery
  - Less pain
  - Less complications??
  - Quick return to daily lifestyle
Indications and Anatomic Requirements for PTMSA

• **Indications**
  – Class III or IV patients
  – Resting or inducible gradient >50 mmHg

• **Anatomic requirements**
  – Septum >18mm
  – No intrinsic mitral valve disease
  – Relatively normal coronary arteries
  – Septal perforator of sufficient size and proper location
Alcohol Septal Ablation
79 yo Man, FEV1 0.8L, s/p CVA
Alcohol Septal Ablation
79 yo Man, FEV1 0.8L, s/p CVA

18 month f/u
CCF Alcohol Septal Ablation Experience
1994 - 2005

- 55 pts, 63±13 years, 67% female, 8±1 year f/u

Results:
- Mortality: 0@48h, 2@1y, 7@5y, 13@10y, age only predictor
- Max gradient: 104±35 mmHg ⇒ 49±28 mmHg
- Septum: 24±4 mm ⇒ 18±6 mm
- Minnesota Living w/ HF: 63 ⇒ 25

Kwon DH et al. JACCint 2008; 1: 432-8
Myectomy vs Alcohol Septal Ablation

Updated Meta-Analysis

- 8 observational studies, 380 ASA/326 myectomy

**Results:**
- No difference in short- or long-term mortality
- No difference in functional class or improvement
- Greater risk of pacer with ASA (OR 2.6)
- Residual gradient higher in ASA

Kwon DH et al. JACC 2010; 55: 823-34
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Papillary muscle realignment for symptomatic left ventricular outflow tract obstruction

Roosevelt Bryant, III, MD, and Nicholas G. Smedira, MD, Cleveland, Ohio

Pledgetted mattress sutures direct PM away from LVOT

Post-op result, no LVOT obstruction, even with amyl

Kwon et al. JASE 2009; 22: e5-e6

Bryant et al. JTCVS 2008; 135: 223-4
Papillary Muscle Realignment

Initial Results

• **204 consecutive patients**
  – 143 myectomy, 39 myectomy + MV surgery, 22 papillary muscle realignment ± myectomy

• **Results:**
  – No hospital or 30 day deaths
  – No difference in initial or predischarge gradients
  – 21/22 pap muscle realignment pts became asymptomatic (1 MVR for persistent gradient)

Kwon DH et al. JTCVS 2010; 140: 317-24
Thanks!