HYPERTROPIC CARDIOMYOPATHY

By
Dr RAJESH MD General Medicine
Postgraduate 2nd year
HCM

- HCM was described as the presence of left ventricular hypertrophy but non dilated left ventricle in the absence of obvious cause (htn, valvular disease, etc) often involving the interventricular septum.
HISTORY

• The pathology of HCM was first described by a French pathologist back in mid 19th century.

• Simultaneous reports by Brock and Teare in England back in the 1950’s brought the subject to modern attention.

• Names commonly in use are idiopathic hypertropic subaortic stenosis, Hypertropic obstructive cardiomyopathy, muscular subaortic stenosis (IHSS, HOCM, MSS) have been largely abandoned.

• Hence, the preferred and generally accepted name for this condition is now hypertrophic cardiomyopathy (HCM).
Genetic basis

- HCM was initially thought to be idiopathic in origin, but now it is known to be due to mutations in genes encoding proteins in the contractile apparatus.
- HCM is transmitted by autosomal dominant inheritance (few cases caused by mutations in the mitochondrial genome)
- 2/3 of patients have family hx, the rest have sporadic mutations (de novo)
- Genetic analysis for diagnosis is not available routinely
- In future likely to be the Gold Standard for diagnosis
<table>
<thead>
<tr>
<th>Gene</th>
<th>Symbol</th>
<th>Locus</th>
<th>Frequency</th>
<th>Mutations</th>
</tr>
</thead>
<tbody>
<tr>
<td>β-Myosin heavy chain</td>
<td>MYH7</td>
<td>14q12</td>
<td>~ 30%</td>
<td>67, mostly missense, a few non-sense, and 3 deletion mutations</td>
</tr>
<tr>
<td>Myosin binding protein-C</td>
<td>MYBPC3</td>
<td>11p11.2</td>
<td>~ 20%</td>
<td>29; 11 missense/nonsense, 10 splice, 8 deletion/insertion mutations</td>
</tr>
<tr>
<td>Cardiac troponin T</td>
<td>TNNT2</td>
<td>1q32</td>
<td>~ 20%</td>
<td>14: 12 missense, 1 splice, and 1 deletion mutations</td>
</tr>
<tr>
<td>α-Tropomyosin</td>
<td>TPM1</td>
<td>15q22.1</td>
<td>~ 5%</td>
<td>4 missense mutations</td>
</tr>
<tr>
<td>Cardiac troponin I</td>
<td>TNNT2</td>
<td>19p13.2</td>
<td>~ 5%</td>
<td>7 missense and 1 deletion mutations</td>
</tr>
<tr>
<td>Myosin light chain, essential</td>
<td>MYL3</td>
<td>3p21.3-p21.2</td>
<td>&lt;5%</td>
<td>2 missense mutations</td>
</tr>
<tr>
<td>Myosin light chain, regulatory</td>
<td>MYL3</td>
<td>3p21.3-p21.2</td>
<td>&lt;5%</td>
<td>7 missense and 1 truncation mutations</td>
</tr>
<tr>
<td>Cardiac α-actin</td>
<td>ACTC</td>
<td>11q</td>
<td>&lt;5%</td>
<td>2 missense mutations</td>
</tr>
<tr>
<td>K voltage-gated channel</td>
<td>KCNQ4</td>
<td>1p34</td>
<td>Rare</td>
<td>1 deletion mutation in conjunction with deafness</td>
</tr>
<tr>
<td>Titin</td>
<td>TTN</td>
<td>2q24.1</td>
<td>&lt;5%</td>
<td>1 missense mutation</td>
</tr>
<tr>
<td>Protein kinase A, γ-subunit</td>
<td>PRKAG2</td>
<td>7q22-q31.1</td>
<td>?</td>
<td>1 point mutation in conjunction with WPW</td>
</tr>
<tr>
<td>α-Myosin heavy chain</td>
<td>MYH6</td>
<td>14q</td>
<td>Rare</td>
<td>1 missense and 1 rearrangement mutations</td>
</tr>
<tr>
<td>Mitochondrial DNA</td>
<td>MTTI</td>
<td>Mitochondrial</td>
<td>Rare</td>
<td>tRNA isoleucine and tRNA glycine</td>
</tr>
</tbody>
</table>

• J Mol Cell Cardiol 33, 655-670 (2001)
INCIDENCE AND PREVALENCE

• HCM is now estimated to have a prevalence of 1 in 500.

• It affects people of all ages.

• Detectable cardiovascular abnormalities usually develop during periods of rapid somatic growth.

• HCM has been described in infants and young children, but data in these age groups is limited.
Mechanism of Hypertrophy

• Not fully understood
• Likely due to:
  • Alteration of protein structure that changes delicate interactions
  • Change in the sensitivity to regulators like calcium or ATP
  • Impaired energy metabolism
  • Decreased velocity of myocyte contraction
Macroscopic Anatomy

• Myocardial mass is increased
• Septal hypertrophy (most common)
• Small ventricular cavity
• Atria are dilated & hypertrophied
• Localised, mild hypertrophy may be seen in 30%
• Prominent hypertrophy is less common in infants & spurt of hypertrophy develops during adolescence
• Development of morphologic features after age of 18 years is unusual
• Atria are dilated, often hypertrophied (due to high resistance to filling due to diastolic dysfunction and Atrioventricular valve leak)
Mitral Valve Apparatus Abnormalities

- Seen in 2/3rd of patents
- Elongation of the mitral leaflets
- Anterior displacement of papillary muscle
- Very short chordae tendineae
- Direct insertion of the papillary muscle onto the mitral leaflets or septum
Mitral Valve Apparatus Abnormalities
Microscopic Anatomy

- Myocyte disarray
- Myofibrillar disarray
- Abnormal intramural coronary arteries
Myocyte Disarray

- Loss of normal parallel arrangement of myocytes
- Abnormal intercellular connections
- Abnormal whorled appearance of myofiber arrangement
- Variation in length and diameter of individual myocytes
ROBBINS PATHOLOGIC BASIS OF DISEASE.
Cotran, Kumar, Robbins. 5th edition.
Abnormal Intramural Coronary Arteries

- Abnormal intramural coronary arteries with thickened walls (composed of increased intimal and medial components) and narrowed lumen are present in 80% of patients at necropsy, most frequently within or close to areas of replacement fibrosis.

- This microvascular small-vessel disease is responsible for clinically silent myocardial ischemia and myocyte death, leading to a repair process in the form of replacement (often transmural) with fibrosis.
Abnormal conduction

• The disorganized cellular architecture and replacement fibrosis evident in HCM impair transmission of electrophysiologic impulses and predispose to disordered patterns and increased dispersion of electrical depolarization and repolarization
• This serving as an electrically unstable substrate and nidus for reentry ventricular tachyarrhythmias and sudden death.
Pathophysiology

- **Diastolic function** (Hallmark)
- LVOT obstruction
- Mitral regurgitation
- Myocardial ischemia
- Autonomic Dysfunction
- Systolic function
Diastolic Dysfunction

- Reduced ventricular compliance in HCM is due to loss of elastic properties of the LV chamber due to
  - hypertrophy
  - scarring
  - Interstitial fibrosis
  - disorganized cellular architecture.
- Diastolic dysfunction is likely to be the fundamental mechanism by which heart failure occurs in nonobstructive HCM with preserved LV systolic function.
- In small subset of pts biatrial enlargement pulmonary HTN, and severe Right heart failure
Systolic anterior motion of mitral valve

- Nearly always associated with systolic anterior motion (SAM) of AML, less commonly PML
- **Mechanism of SAM:**
  - MV is pulled against the septum by the contraction of abnormally oriented papillary muscles and elongated leaflets
  - Mitral valve is pushed against the septum because of the abnormal position of LVOT
  - **Venturi Effect:** Mitral valve is drawn towards ventricular septum because of lower pressure that occur as blood is ejected at a high velocity through narrowed LVOT
  - **Drag Effect:** Hyrodynamic pushing force of flow on leaflets
  - In 15%, one or both papillary muscle insert anomalously directly into AML causing mid cavity obstruction
Mitral Regurgitation

• More frequent in obstructive type
• Directly related to the severity of SAM
• In 80% of HCM, MR is due to SAM & the jet mild to moderate and is directed posteriorly
• In 20% of pts, related to intrinsic abnormality of MV and jet is directed anteriorly
# Myocardial Ischemia

<table>
<thead>
<tr>
<th>TABLE 59–5</th>
<th>Possible Mechanisms for Ischemia in Hypertrophic Cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Increased Myocardial Oxygen Demand</td>
<td>Reduced Myocardial Perfusion</td>
</tr>
<tr>
<td>Myocardial hypertrophy</td>
<td>Small vessel disease</td>
</tr>
<tr>
<td>Diastolic dysfunction</td>
<td>Abnormal vascular responses</td>
</tr>
<tr>
<td>Myocyte disarray</td>
<td>Myocardial bridges</td>
</tr>
<tr>
<td>Left ventricular outflow obstruction</td>
<td>Increased coronary vascular resistance</td>
</tr>
<tr>
<td>Arrhythmias</td>
<td></td>
</tr>
</tbody>
</table>

Autonomic Dysfunction

- Nearly 25%, will have an abnormal blood pressure response to exercise as defined by either:
  - Failure of SBP to rise greater than 20 mm Hg or a fall in SBP
  - This is due to systemic vasodilation during exercise and occurs in spite of an appropriate rise in cardiac output
  - Abnormal BP response and autonomic tone are associated with a poorer prognosis
Clinical manifestations of HCM

SYMPTOMS IN HCM

Diagnosis 3rd/4th decade
Rarely in infants ---Heart failure
Majority are mildly symptomatic
- Dyspnea (90%)
- Angina (75%)
- Presyncope
- Syncope
- PND, palpitation rare
- Sudden Death
- Asymptomatic

Congestive heart failure is rarely seen in HCM pts. In normal sinus rhythm, but may be seen with severe LVOT obstruction or systolic and/or diastolic dysfunction and is common in the presence of AF
Physical Findings

With outflow obstruction
Arterial pulses rapid rise - with bisferiens contour
Double or triple apical impulses may be palpable
  • Outward systolic thrust - ventricular contraction
  • Presystolic accentuated atrial contraction.
Medium-pitch mid systole at the lower left sternal border and apex
Loud murmurs > 3/6 - LV outflow gradients >30 mm Hg

Without subaortic gradients
Subtle - with no or soft systolic murmur
Forceful apical impulse
<table>
<thead>
<tr>
<th>Intervention</th>
<th>Contractility</th>
<th>Preload</th>
<th>Afterload</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Increase in Gradient and Murmur</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valsalva maneuver (during strain)</td>
<td>—</td>
<td>↓</td>
<td>↓</td>
</tr>
<tr>
<td>Standing</td>
<td>—</td>
<td>↓</td>
<td>—</td>
</tr>
<tr>
<td>Postextrasystole</td>
<td>↑</td>
<td>↑</td>
<td>—</td>
</tr>
<tr>
<td>Isoproterenol</td>
<td>↑</td>
<td>↓</td>
<td>↓</td>
</tr>
<tr>
<td>Digitalis</td>
<td>↑</td>
<td>↓</td>
<td>—</td>
</tr>
<tr>
<td>Amyl nitrite</td>
<td>—</td>
<td>then ↑</td>
<td>↓ then ↑</td>
</tr>
<tr>
<td>Nitroglycerin</td>
<td>—</td>
<td>↓</td>
<td>↓</td>
</tr>
<tr>
<td>Exercise</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Tachycardia</td>
<td>↑</td>
<td>↓</td>
<td>—</td>
</tr>
<tr>
<td>Hypovolemia</td>
<td>↑</td>
<td>↓</td>
<td>↓</td>
</tr>
<tr>
<td><strong>Decrease in Gradient and Murmur</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Müller maneuver</td>
<td>—</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Valsalva overshoot</td>
<td>—</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Squatting</td>
<td>—</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Alpha-adrenoceptor stimulation</td>
<td>—</td>
<td></td>
<td>↑</td>
</tr>
<tr>
<td>(phenylephrine)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Beta-adrenoceptor blockade</td>
<td>↓</td>
<td>↑</td>
<td>—</td>
</tr>
<tr>
<td>General anesthesia</td>
<td>↓</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Isometric handgrip</td>
<td>—</td>
<td></td>
<td>↑</td>
</tr>
</tbody>
</table>

↑ = increase; ↓ = decrease; — = no major change.
DIAGNOSTIC TOOLS

- ECG. It is abnormal in 75 to 95% of patients, usually LVH strain pattern. Abnormal Q waves may reflect septal hypertrophy. Negative T waves in V3-V5 (apical HCM)
- ECHO
- CXR
- MRI
ECG

Abnormal - >90% of pts & >75% of asymptomatic relatives
  • Increased voltages consistent with LV hypertrophy
  • ST-T changes - marked T wave inversion in the lateral precordial leads
  • Left atrial enlargement
  • Deep and narrow Q waves
  • Diminished R waves in the lateral precordial leads.

Normal ECG - 5% of pts
  • Less severe phenotype and favorable course
  • Not predictive of future sudden death

Increased voltages
  • Weakly correlated with the magnitude of LV hypertrophy
  • Do not distinguish the obstructive and nonobstructive forms
Fig. 5. ECG of patient with apical hypertrophic cardiomyopathy variant with deeply inverted T waves in chest leads V₂-V₆ and limb leads II, III, and aVL.
ECG abnormalities

- Supraventricular tachycardia (46 percent)
- Premature ventricular contractions (43 percent)
- Nonsustained ventricular tachycardia (26 percent)
- Atrial fibrillation (25 to 30 percent)
- Preexcitation has also been associated with HCM
Mechanisms for cardiac arrest

- Observations have recorded several mechanisms for the generation of ventricular arrhythmias.
  - Paroxysmal AF
  - Sinus tachy with abnormal vascular responses/ischemia
  - Sustained monomorphic VT
  - Rapid AV conduction via accessory path
  - AV block
Echocardiography

• Diffuse hypertrophy of the ventricular septum and anterolateral free wall (70% to 75%)
• Basal septal hypertrophy (10% to 15%)
• Concentric hypertrophy (5%)
• Apical hypertrophy (<5%)
• Hypertrophy of the lateral wall (1% to 2%).
• Mitral annulus velocity, Ea - status of myocardial relaxation - reduced in most patients with HCM
HETEROGENEITY IN THE PATTERN AND EXTENT OF LEFT VENTRICULAR WALL THICKENING IN HCM
Mimicking Hypertrophic Cardiomyopathy

- Chronic hypertension
- RV hypertrophy
- Cardiac amyloidosis
- Athlete's heart
- Pheochromocytoma
- Long-term hemodialysis
- Fabry disease
- Friedreich ataxia.

Apical hypertrophy - apical cavity obliteration caused by hypereosinophilic syndrome or noncompaction.
# Athlete's Heart Vs Hypertrophic Cardiomyopathy

<table>
<thead>
<tr>
<th>HCM</th>
<th>Athletic heart</th>
</tr>
</thead>
<tbody>
<tr>
<td>Can be asymmetric</td>
<td>Concentric &amp; regresses</td>
</tr>
<tr>
<td>Wall thickness: &gt; 15 mm</td>
<td>&lt; 15 mm</td>
</tr>
<tr>
<td>LA: &gt; 40 mm</td>
<td>&lt; 40 mm</td>
</tr>
<tr>
<td>LVEDD: &lt; 45 mm</td>
<td>&gt; 45 mm</td>
</tr>
<tr>
<td>Diastolic function: always abnormal</td>
<td>Normal</td>
</tr>
</tbody>
</table>
Magnetic resonance imaging

- Useful in patients where Echo is inadequate
- Can distinguish hypertrophy VS infiltration
- Gadolinium enhanced MRI gives clear picture of scarring and disarray
- Helps in risk stratification: patients with high risk of death have hyper enhancement
General guidelines

- Screening all first-degree relatives is recommended by ECHO
- <12 year old optional
- 12 to 21 years every 12-18 months
- >18-21 years every 5 years
- Children & participating in competitive athletics Every 12 to 18 months
- Adults no competitive athletics - every 5 years
- Counseled against engaging in competitive athletics
- Maintain hydration
Treatment

Hypertrophic cardiomyopathy

Treatment

• Reduce symptoms due to haemodynamic abnormalities
  • LV diastolic dysfunction: B blocker/verapamil
  • LVOT obstruction: myomectomy (surgical or interventional), pacing, alcohol septal ablation
  • LV systolic dysfunction – treated like CCF

• Reduce angina
  • B blocker/verapamil
  • Correction of myocardial bridging
  • Treat coronary atheroma if present

• Reduce symptoms due to rhythm abnormalities
  • drugs to prevent VT or AF
    • B blocker, Amidarone
MEDICAL TREATMENT

• Empirical & highly variable

  Beta blockers

• Slowing heart rate
• Reducing force of LV contraction
• Augmenting ventricular filling and relaxation
• Decreasing myocardial oxygen consumption
• Long-acting preparations - propranolol, atenolol, metoprolol or nadolol
• Propranolol 200-400 mg/d(large doses)
• Selective β- Blocker lose selectivity at high doses
• Target resting heart rate - 60 beats/min
• May require up to 400 mg equivalent of metoprolol
Verapamil

- Dose 240-320 mg/dl
- Improves symptoms and exercise capacity (patients without marked obstruction to LV outflow)
- Beneficial effect on ventricular relaxation and filling
- Better angina control than BB
- Hemodynamic deterioration with CCB agents - lowering of the afterload in the presence of severe outflow tract gradients and high diastolic filling pressures
Disopyramide

- Negative inotropic effect decreases the gradient and improve symptoms.
- Concomitant beta blockade may be important to prevent rapid atrioventricular node conduction
- Dose 300 and 600 mg/d
- The corrected QT interval must be monitored
- Diuretic agents may be judiciously administered
- Either beta blockers or verapamil initially
- No advantage by combinations of BB & CCB
- Disopyramide may be combined with BB or CCB
Sudden Death & Risk stratification

- Primary ventricular tachycardia and ventricular fibrillation
- Adolescents and young adults <30 to 35 years of age
- Most common cause of Athletic field deaths
- Death most commonly occur at rest
**High Risk**

Secondary prevention
1. Prior cardiac arrest
2. Sustained ventricular tachycardia

Primary prevention
one or more of the following
1. Family history of one or more premature HCM-related deaths, particularly if sudden and multiple
2. Unexplained syncope, especially if recent and in the young
3. Hypotensive or attenuated blood pressure response to exercise
4. Multiple, repetitive (or prolonged) NSVT on Holter
5. Massive LVH (wall thickness, $\geq 30$ mm), particularly in young patients
<table>
<thead>
<tr>
<th>Major Risk Factor</th>
<th>Screening Technique</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of cardiac arrest or spontaneous sustained ventricular tachycardia</td>
<td>History</td>
</tr>
<tr>
<td>Syncope</td>
<td>Usually with or after exertion</td>
</tr>
<tr>
<td>Family history of sudden cardiac death</td>
<td>Or possibly with a documented gene mutation associated with high risk</td>
</tr>
<tr>
<td>Spontaneous nonsustained ventricular tachycardia</td>
<td>&gt;3 beats at rate &gt;120</td>
</tr>
<tr>
<td>LV thickness &gt;30 mm</td>
<td>Present in about 10% of patients, but many sudden deaths occur with wall thickness &lt;30 mm</td>
</tr>
<tr>
<td>Abnormal blood pressure response to exercise</td>
<td>Systolic blood pressure fall or failure to increase at peak exercise</td>
</tr>
</tbody>
</table>
SUDDEN DEATH PREVENTION

• There is little evidence that pharmacological strategies and rhythm modulating drugs reduce risk for sudden death.
• ICD may be the most effective treatment modality for the high-risk patient, with the potential to alter natural history.
• ICD should be considered for patients with prior cardiac arrest or ventricular arrhythmias.
• Another author recommends that patients with 2 or more risk factor should have ICD and/or amiodarone therapy. 3% risk of sudden death per year.
TREATMENT OF ATRIAL FIBRILLATION

- Electrical or pharmacological cardioversion.
- Amiodarone is effective for reducing AF recurrences.
- Bblocker and verapamil for rate control.
- A-V node ablation and PM placement.
- Anticoagulation.
INDICATION FOR SURGERY

• Small group of patients with heart failure and outflow gradient of 50 mm HG or more.
• Usually patients have heart failure symptoms refractory to medical therapy.
• Patients with provicable obstruction sometimes are referred for surgery.
• Marked functional disability (NYHA Classes III and IV)
Septal myectomy

- Transaortic ventricular septal myectomy (Morrow procedure) involves resection of a small portion of muscle (usually 3 to 10 g) from the basal septum.
- Operative mortality has steadily decreased and is now <1% during the last 15 years at the most experienced myectomy centers.
- Myectomy also beneficially alters the clinical course of HCM;
- surgical patients achieve long-term survival equivalent to that expected in the general population and superior to that of nonsurgical HCM patients with outflow obstruction.
Septal myectomy

- Patients < 40 years: mortality < 1%
- Patients > 65 years: mortality 10-15%
- Survival better than medically treated patients
- At present, surgical myectomy is not recommended for asymptomatic (or mildly symptomatic) patients,
- Conclusive evidence is lacking that prophylactic relief of obstruction is of benefit long term.
- Complication (rare): Aortic incompetence
ALCOHOL SEPTAL ABLATION

- Described in 1995 by Sigwart.

- Percutaneous alcohol septal ablation involves introduction of 1 to 3 mL of 95% alcohol into a major septal perforator coronary artery to create necrosis and a permanent transmural myocardial infarction in the proximal ventricular septum.

- This scar leads to progressive thinning and restricted septal excursion, outflow tract enlargement, and reduction in LV outflow tract gradient and mitral regurgitation in most patients.
**Ethanol ablation in HCM**  Representation of ethanol ablation via a septal coronary artery in hypertrophic cardiomyopathy (HCM). A section of hypertrophied left ventricle is shown, while the inset demonstrates a balloon occluding a septal coronary artery and ethanol-induced infarction. (Reproduced with permission from: Braunwald, E. Hypertrophic cardiomyopathy--the benefits of a multidisciplinary approach. N Engl J Med 2002; 347:1306. Copyright © 2002 Massachusetts Medical Society.)
• An American College of Cardiology/European Society of Cardiology expert consensus panel recommended surgical myectomy as the preferred and primary management option for patients with severe refractory symptoms and marked outflow obstruction.

• Alcohol ablation is regarded as an alternative treatment strategy for patients not considered optimal operative candidates (e.g., advanced age, significant comorbidity and increased operative risk, or strongly adverse to surgery personally).
COMPLICATIONS OF SEPTAL ABLATION

- 2 % MORTALITY
- Bundle branch block up to 60%.
- Complete AV block with pacemaker implantation 0-40%.
- Early ventricular arrhythmias causes sudden death
- On the basis of this consideration, some practitioners have prudently implanted ICDs prophylactically in selected patients with alcohol septal ablation
DUAL CHAMBER PACING

• Permanent dual chamber was promoted as an alternative to myectomy for obstructive HCM patients with refractory heart failure symptoms.
• Reduction in subaortic gradient may result from pacing in some patients, this benefit is inconsistent, particularly compared with that achieved by myectomy or alcohol ablation.
• The overall role for pacing in HCM has become particularly limited during the last decade.
HCM AND PREGNANCY

• Thaman R, et al. studied 127 patients referred to St. George’s Hospital in UK.
• Study concluded that most patients with HCM tolerate pregnancy well.

Heart 2003; 89: 752-756.
“Burned-Out” HCM

• About 5-10% of pts eventually develop a burned out phase characterized by:
  • Marked myocardial thinning
  • Ventricular dilation
  • Systolic dysfunction
  • Progressive CCF
  • Resembles DCM
FIGURE 59–12  Hypothetical model for the pathogenesis of the end-stage phase of hypertrophic cardiomyopathy. Asterisk designates the following possibilities: (1) enhanced myocardial oxygen requirements and reduced myocardial capillary density relative to marked left ventricular (LV) hypertrophy (LVH) and (2) increased diastolic wall tension and coronary vascular resistance resulting from abnormal LV relaxation and impaired filling. CHF = congestive heart failure. (Modified from Maron BJ, Spirito P: Implications of left ventricular remodeling in hypertrophic cardiomyopathy. Am J Cardiol 81:1339-1344, 1998.)
Prognosis

• The general prognosis for hypertrophic cardiomyopathy is good.
• Survival is comparable to an age-matched population without cardiomyopathy.
• The sudden death risk is less than 1 % per year
• 1 in 20 patients will progress to overt systolic dysfunction with a reduced ejection fraction with or without dilated remodeling ("burned out" or end-stage hypertrophic cardiomyopathy)
• These patients suffer from low cardiac output and have a high risk of death from progressive heart failure and sudden death unless they undergo cardiac transplantation
THANK YOU