John Hancock INSURANCE

Understanding Cardiomyopathy

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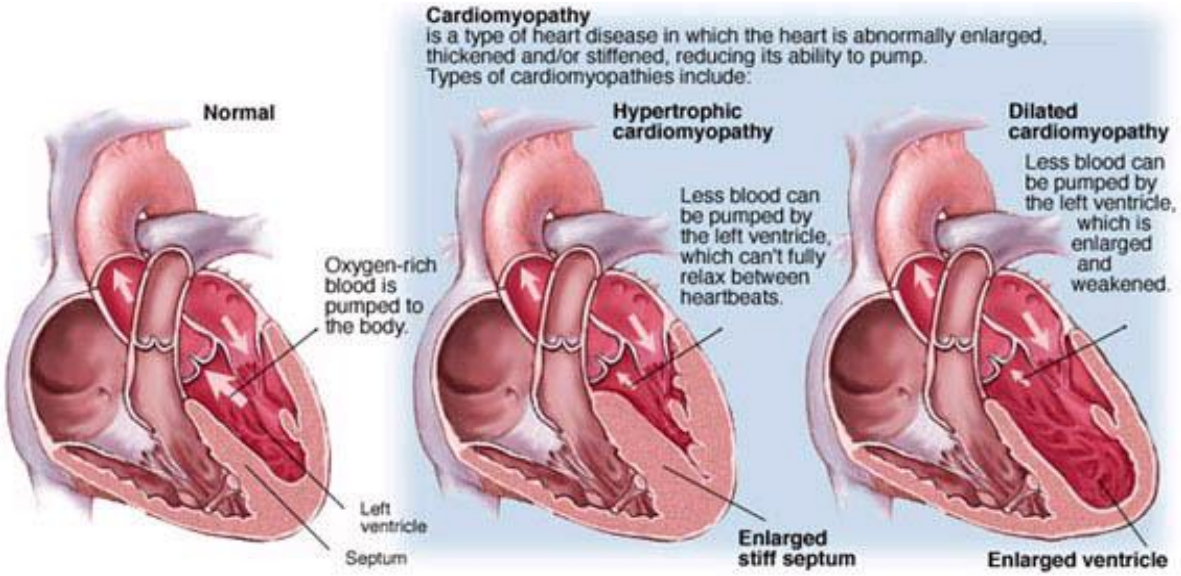
Agenda

- Define Cardiomyopathy
- Cardiomyopathy Functional Classifications:
  - Dilated Cardiomyopathy
  - Hypertrophic Cardiomyopathy
  - Restrictive Cardiomyopathy
  - Arrhythmogenic Right Ventricular Cardiomyopathy
- Underwriting Cardiomyopathy
- Case Studies

Definition

- The term cardiomyopathy is purely descriptive, meaning disease of the heart muscle
- 2006 – AHA defined cardiomyopathies as "a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to a variety of causes that frequently are genetic. Cardiomyopathies either are confined to the heart or are a part of generalized systemic disorders, often leading to cardiovascular death or progressive heart failure-related disability"

Cardiomyopathy



Source: Nursing Review, 2001

Functional Classification

- Dilated (congestive, DCM, IDC)
  - Ventricular dilation, hypokinetic left ventricle, and systolic dysfunction
- Hypertrophic (IHSS, HCM, HOCM, ASH)
  - Inappropriate myocardial hypertrophy, with or without left ventricular obstruction
- Restrictive (infiltrative)
  - Abnormal ventricular filling with diastolic dysfunction
- Arrhthymogenic right ventricular
  - Fibroadipose replacement of right ventricle

Source: my.clevelandclinic.org/disorders/Cardiomyopathy

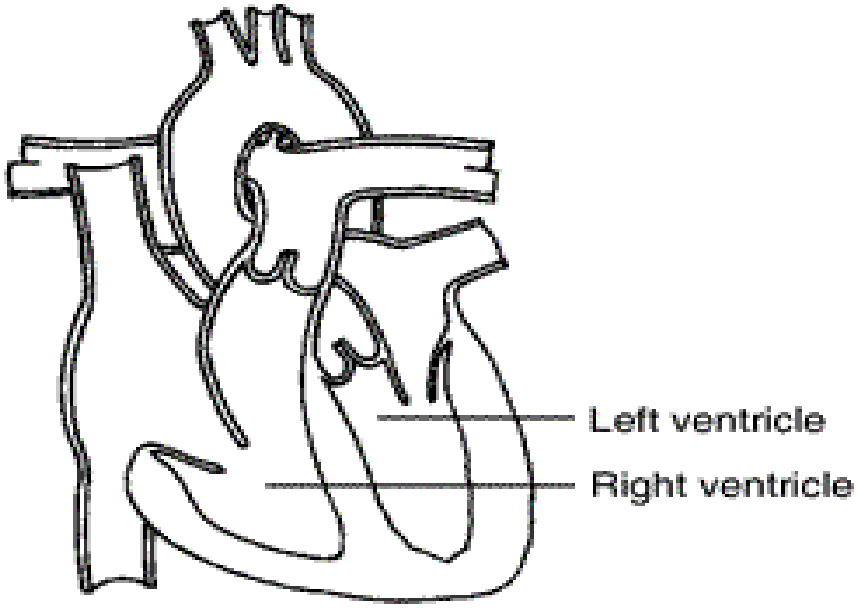
Dilated Cardiomyopathy

Dilated Cardiomyopathy (DCM) – Defined

- Primary (idiopathic) is a disease of unknown etiology that principally affects the myocardium leading to LV dilation and systolic dysfunction
- Secondary causes include ischemia, alcoholic, peripartum, post-infectious, viral
- Most common of the cardiomyopathies

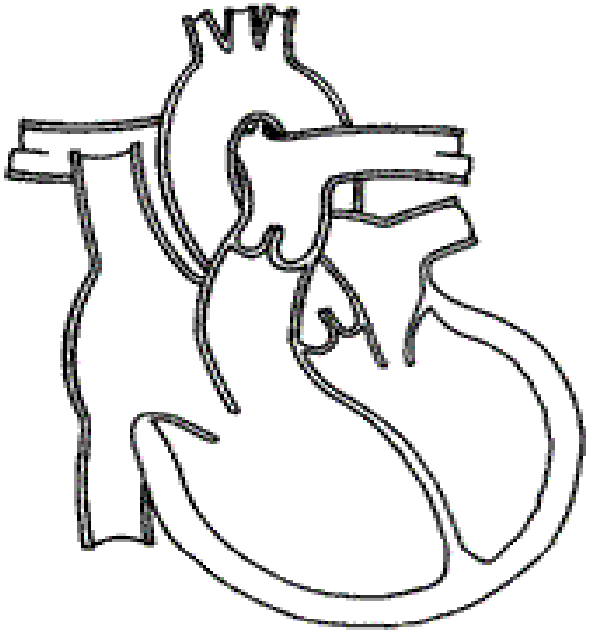
Schematic of DCM

Normal Heart



Heart chambers relax and fill, then contract and pump.

Heart with Dilated Cardiomyopathy



Muscle fibers have stretched. Heart chamber enlarges

Source: Medslides.com

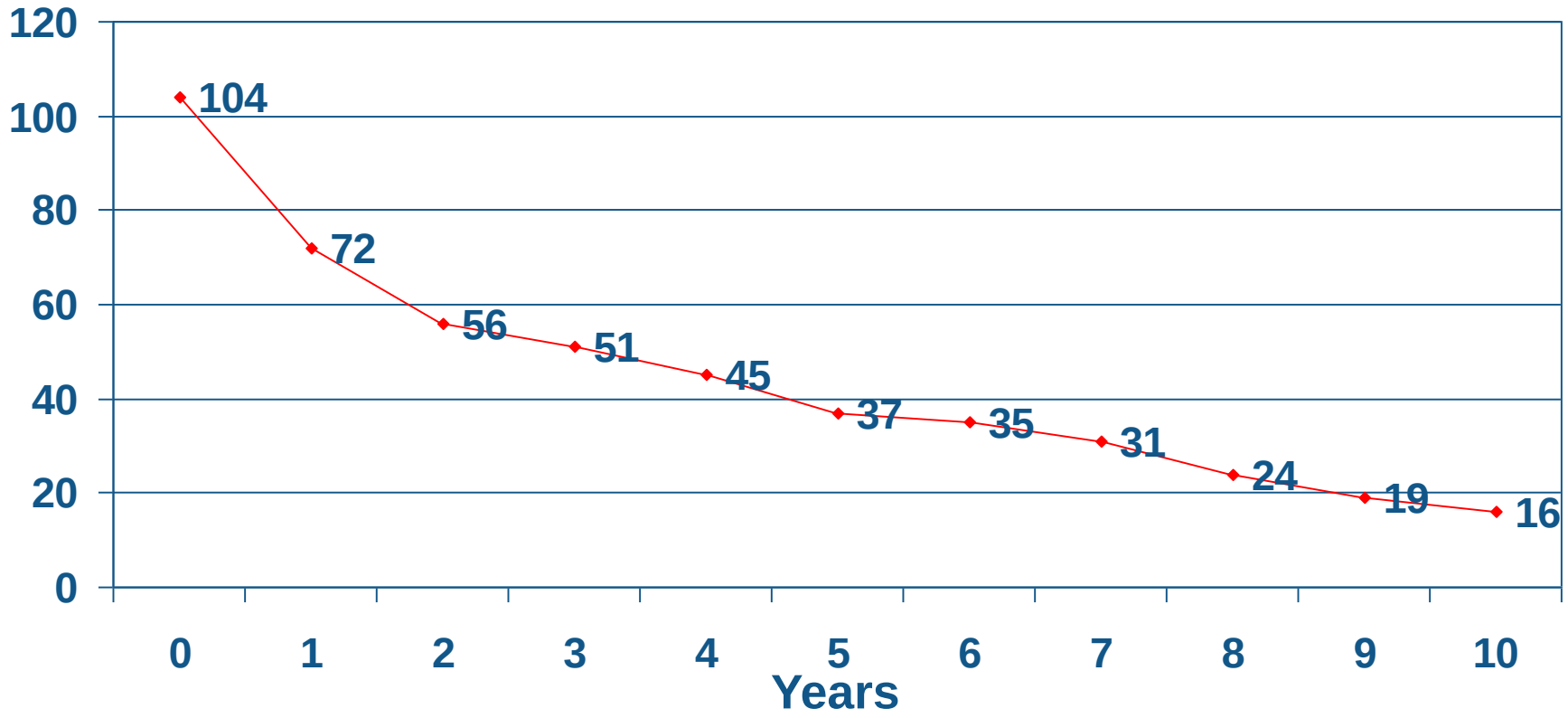
DCM – Incidence and Prognosis

- Prevalence is 36 per 100,000
- Third most common cause of heart failure
- Most frequent cause of heart transplantation
- DCM accounts for approximately 10,000 deaths and 46,000 hospitalizations per year in the U.S.
- Complete recovery is rare

Source: Up to Date – "Definition and classification of the cardiomyopathies" Leslie T Cooper, Jr, MD last updated 4/2/12 – www.uptodate.com/contents/definition-and-classification-of-the-cardiomyopathies

Idiopathic Dilated Cardiomyopathy

- Observed survival of 104 patients



Source: Am J Cardiol 1981; 47:525

Clinical Manifestations

- Highest incidence in middle age
  - Blacks 2x more frequent than whites
  - Men 3x more frequent than women
- Symptoms may be gradual in onset
- Acute presentation
  - Misdiagnosed as viral URI in young adults
  - Uncommon to find specific myocardial disease on endomyocardial biopsy

Source: UpToDate – "Evaluation of the patient with heart failure or cardiomyopathy" Wilson S Colucci, MD, last updated 6/7/13 – www.uptodate.com/contents/evaluation-of-the-patient-with-heart-failure-or-cardiomyopathy

Clinical Manifestations (cont'd)

- Symptoms/signs of heart failure
  - Pulmonary congestion (left heart failure) dyspnea (rest, exertional, nocturnal), orthopnea
  - Systemic congestion (right heart failure), edema, nausea, abdominal pain, nocturia
  - Low cardiac output
  - Hypotension, tachycardia, tachypnea
  - Fatigue and weakness
- Arrhythmia
  - Atrial fibrillation, conduction delays, complex PVC's, sudden death

Diagnostics

- CXR (enlarged heart, CHF)
- Electrocardiogram (tachycardia, A-V block, LBBB, NS S-T's, PVC's)
- 24-hour Holter monitor
  - If lightheadedness, palpitation, syncope
- Two-dimensional echocardiogram (left ventricular dilation, global hypokinesis, low EF)
- Myocardial biopsy, rare
- Cardiac catheterization (R/O CAD)
  - if age >40, ischemic history, high risk profile, abnormal ECG

DCM – Treatment

- Limit activity based on functional status
- Salt restriction
- Fluid restriction
- Initiate medical therapy
  - ACE inhibitors, diuretics – slow progression and improve survival
  - Digoxin, Coreg – slow progression and improve survival
  - Hydralazine/nitrate combination – correct fluid overload
  - Anticoagulation prn (EF <30%, hx of embolic events)
  - Implantable defibrillators

DCM – Treatment (cont'd)

- Cardiac transplantation
  - This disorder is the most common indication for cardiac transplantation
  - Survival after transplant is
    - 80% one year
    - 70% 5 years

- Left Ventricular Reduction Procedures
  - LV reshaping procedures

Source: UpToDate – "Diagnosis and management of ischemic cardiomyopathy" James C Fang MD, Sary Aranki MD, last updated 6/10/13 – www.uptodate.com/contents/diagnosis-and-management-of-ischemic-cardiomyopathy

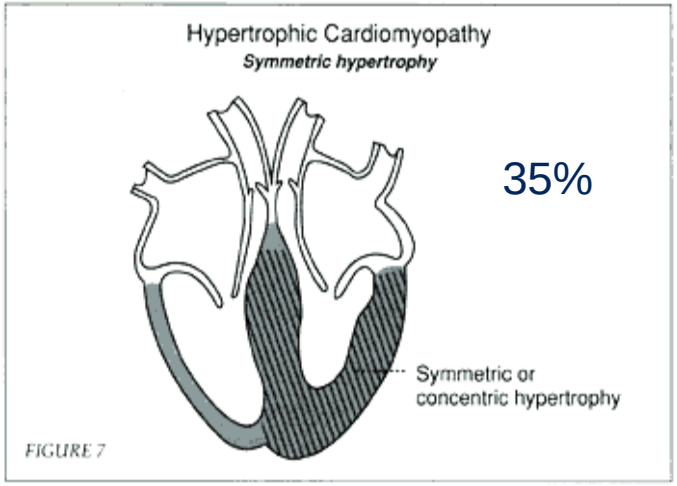
Hypertrophic Cardiomyopathy

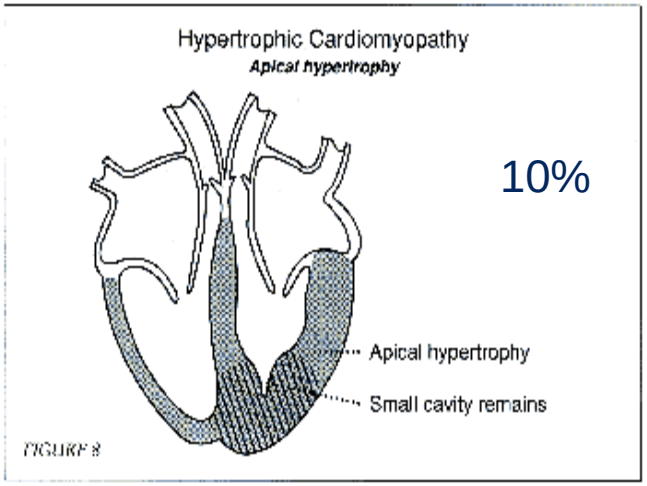
Hypertrophic Cardiomyopathy (HCM) – Defined

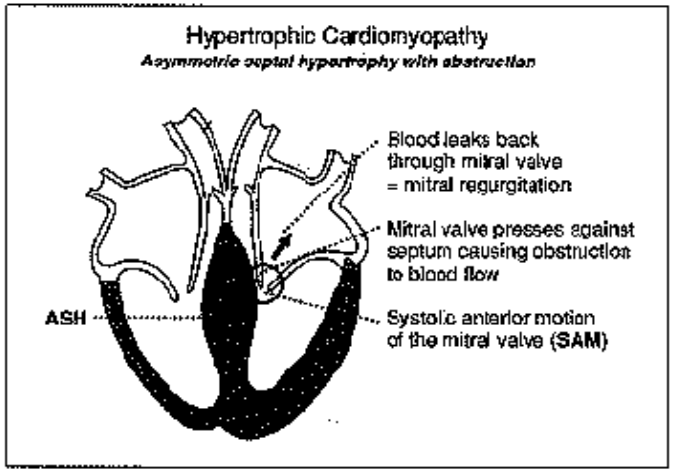
- First described in 1869 and accepted as a clinical entity in the 1950's
- Prevalence 1:500 (0.2%)
- Genetic disease characterized by hypertrophy of the left ventricle with marked variable clinical manifestations morphologic and hemodynamic abnormalities
- Small LV cavity, septal hypertrophy which can be asymmetric (ASH), systolic anterior motion of the mitral valve leaflet (SAM), +/- obstruction of left ventricular outflow with low stroke volume, but elevated EF

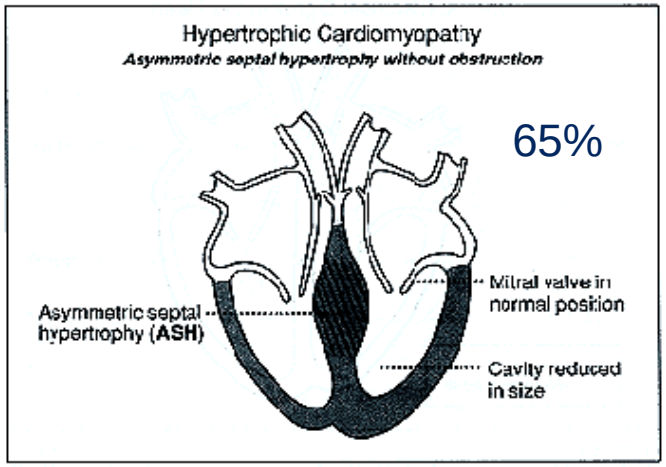
Source: UpToDate – "Natural history of hypertrophic cardiomyopathy" Martin S Maron MD, Perry M Elliott, MD, last updated 8/14/13 – www.uptodate.com/contents/natural-history-of-hypertrophic-cardiomyopathy

Variants of HCM



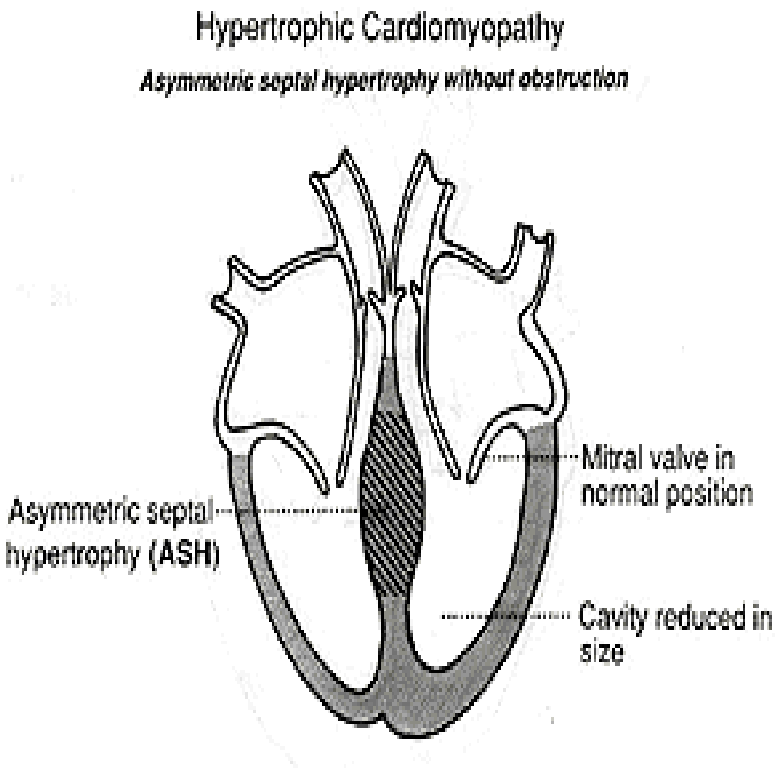






Souce: www.kanter.com/hcm

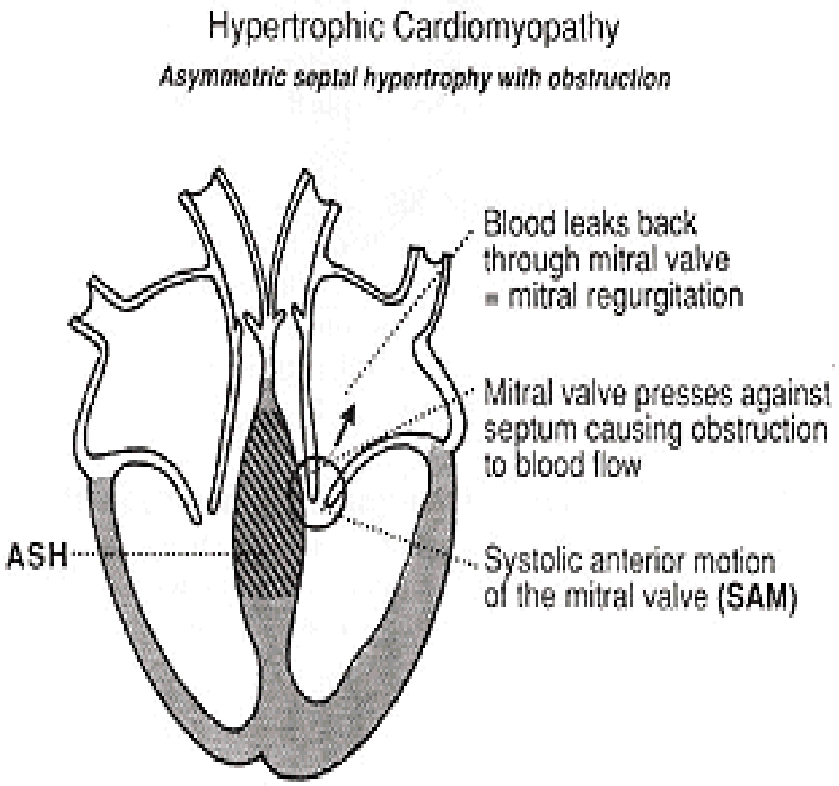
HCM – ASH Without Obstruction



- The major abnormality of the heart in HCM is an excessive thickening of the muscle. Thickening usually begins during early adolescence and stops when growth has finished. It is uncommon for thickening to progress after this age
- The left ventricle is almost always affected, and in some patients the muscle of the right ventricle also thickens
- Hypertrophy is usually greatest in the septum. The muscle thickening in this region may be sufficient to narrow the outflow tract. This thickening is associated with obstruction to the flow of blood out of the heart into the aorta

Source: www.kanter.com/hcm

HCM – ASH With Obstruction



- Asymmetric septal hypertrophy with obstruction to the outflow of blood from the heart may occur. The mitral valve touches the septum, blocking the outflow tract. Some blood is leaking back through the mitral valve causing mitral regurgitation

Source: www.kanter.com/hcm

Pathophysiology of HCM

- Dynamic LV outflow tract (LVOT) obstruction
  - Outflow tract gradient (>30 mm Hg), considered severe if >50 mm Hg (occurs in 25-30% of cases leading to name hypertrophic obstructive cardiomyopathy)
- Diastolic dysfunction
  - Impaired diastolic filling, ↑ filling pressure
- Myocardial ischemia
- Mitral regurgitation
- Arrhythmias

Left Ventricular Outflow Tract Gradient

- Approximately 20-25% of patients with HCM have a dynamic systolic pressure gradient in the left ventricular outflow tract caused by contact between the mitral valve leaflet(s) and the interventricular septum under resting conditions
- Outflow tract gradient in excess of 30 mmHg is an important cause of symptoms and is an independent predictor of poor prognosis
- Magnitude of the LVOT gradient relates to symptoms and exercise limitations which provides rationale for treatments aimed at gradient reduction

Source: UpToDate – "Types and pathophysiology of obstructive hypertrophic cardiomyopathy" William J McKenna, MD, last updated 03/01/12 – www.uptodate.com/contents/types-and-pathophysiology-of-obstructive-hypertrophic-cardiomyopathy

Clinical Manifestations

- Asymptomatic
  - Echocardiographic finding only
- Symptomatic
  - Dyspnea
  - Angina pectoris
  - Fatigue, pre-syncope, syncope,↑ risk of SCD
  - Palpitation, PND, CHF, dizziness
  - Atrial fibrillation, thromboembolism

EKG Findings

- Abnormal in 85-90% of cases
- LVH, Strain pattern
- Abnormal ST-T's, giant T wave inversions
- Abnormal Q's
- Bundle Branch Block
- Left atrial enlargement
- Ventricular arrhythmias

Source: UpToDate – "Clinical manifestations, diagnosis and evaluation of hypertrophic cardiomyopathy" Martin S Maron, MD, Perry M Elliott, MD, last updated 12/13/12 – www.uptodate.com/contents/clinical-manifestations-diagnosis-and-evaluation-of-hypertrophic-cardiomyopathy

Echocardiogram

- Left ventricular hypertrophy >1.3 cm (usually >1.5 cm)
- Septal to posterior wall ratio >1.3:1
- Mitral regurgitation
- Systolic anterior motion of the mitral valve (SAM)
- Premature midsystolic closure of the aortic valve
- Asymmetric septal hypertrophy (ASH)
- Diastolic dysfunction
- Left ventricular outflow tract obstruction

Echocardiogram (cont'd)

- LVH usually develops between 5-15 years of age in HCM
- A normal ECHO in a young child does not R/O the diagnosis
- Serial ECHOs are recommended up to the age of 20 where there is a family history of HCM

Source: UpToDate – "Clinical manifestations, diagnosis and evaluation of hypertrophic cardiomyopathy" Martin S Maron, MD, Perry M Elliott, MD, last updated 12/13/12 – www.uptodate.com/contents/clinical-manifestations-diagnosis-and-evaluation-of-hypertrophic-cardiomyopathy

Natural History & Clinical Course

- Clinical presentation from infancy to old age
- Variable clinical course 25% of cohort achieve normal longevity
- Annual mortality 3% in referral centers probably closer to 1% for all patients
- Course may be punctuated by adverse clinical events: sudden cardiac death, embolic stroke, and consequences of heart failure
- Sustained V-Tach and V-Fib: most likely mechanism of syncope/ sudden death

Source: UpToDate – "Natural history of hypertrophic cardiomyopathy" Martin S Maron MD, Perry M Elliott, MD, last updated 8/14/13 – www.uptodate.com/contents/natural-history-of-hypertrophic-cardiomyopathy

Natural History & Clinical Course (cont'd)

- Risk of SCD higher in children, may be as high as 6% per year, majority have progressive hypertrophy
- Accounts for 36% of deaths in athletes <35 years
- Clinical deterioration usually is slow
- Poor prognosis in males, young age of onset, family history of SCD, history of syncope, exercise induced hypotension (worst)
- Progression to DCM occurs in 10-15%

Source: UpToDate – "Natural history of hypertrophic cardiomyopathy" Martin S Maron MD, Perry M Elliott, MD, last updated 8/14/13 – www.uptodate.com/contents/natural-history-of-hypertrophic-cardiomyopathy

Risk Factors For SCD

- Young age (<35 years)
- “Malignant” family history of sudden death
- Aborted sudden cardiac death
- Sustained VT or SVT
- Non-sustained VT on Holter monitoring
- Atrial fibrillation
- Dilated left ventricle
- NYHA classes III or IV

Risk Factors For SCD (cont'd)

- Syncope
- Severe hypertrophy (>3.0 cm)
- Abnormal BP response to exercise
- Coronary artery disease
- Strenuous exercise or work

Recommendations for Athletic Activity

- Low-risk, older patients (>30 years) may participate in athletic activity if all of the following are absent:
  - Ventricular tachycardia on Holter monitoring
  - Family history of sudden death due to HCM
  - History of syncope
  - Severe hemodynamic abnormalities, gradient ≥50 mmHg
  - Exercise induced hypotension
  - Moderate or severe mitral regurgitation
  - Enlarged left atrium (≥5.0 cm)
  - Paroxysmal atrial fibrillation
  - Abnormal myocardial perfusion

Management of HCM

- Beta-adrenergic blockers (Atenolol, Toprol, Tenormin etc)
- Calcium channel blockers (Norvasc, Cardizem, etc)
- Anti-arrhythmics (Amiodarone, Norpace)
- Pacemakers (ICD)
- Myomectomy (resection of septum)
- Alcohol septal ablation (controlled MI through septal perforator perfusing basal septum) → wall thinning → decreases in LVOTO
- Transplantation

HCM vs. Athletic Heart

- HCM
  - Can be asymmetric
  - Wall thickness: > 15 mm
  - LA: >40 mm
  - LVEDD: <45 mm
  - Diastolic function: always abnormal
- Athletic Heart
  - Concentric & regresses
  - <15 mm
  - <40 mm
  - >45 mm
  - Normal
  - Occurs in about 2% of elite athletes: typical sports, rowing, cycling, canoeing
  - Former athletes & weekend warriors do NOT develop athletic heart
  - Elite female athletes do NOT develop athletic heart

Hypertensive HCM of the Elderly

- Characteristics
  - Modest concentric LV hypertrophy (<22 mm)
  - Small LV cavity size
  - Associated hypertension
  - Ventricular morphology greatly distorted with reduced outflow tract
  - Sigmoid septum and “grandma SAM” echocardiographic finding only

Restrictive Cardiomyopathy

Restrictive Cardiomyopathies

- Hallmark – abnormal diastolic function
- Rigid ventricular wall with impaired ventricular filling
- Bear some functional resemblance to constrictive pericarditis
- Importance lies in its differentiation from operable constrictive pericarditis
- Much less common then DCM or HCM outside the tropics, but frequent cause of death in Africa, India, South and Central America and Asia primarily because of the high incidence of endomyocardial fibrosis in those regions

Classification

- Idiopathic
- Myocardial
  - Noninfiltrative
    - Idopathic
    - Scleroderma
  - Infiltrative
    - Amyloid
    - Sarcoid
    - Gaucher disease
    - Hurler disease
  - Storage disease
    - Hemochromatosis
    - Fabry disease
    - Glycogen storage
- Endomyocardial
  - Endomyocardial fibrosis
  - Hyperesinophilic synd
  - Carcinoid
  - Metastatic malignancies
  - Radiation, anthracycline

Clinical Manifestations

- Symptoms of right and left heart failure
- Jugular venous pulse
- Echo Doppler
  - Abnormal mitral inflow pattern
  - Prominent E wave (rapid diastolic filling)
  - Reduced deceleration time (↑ LA pressure)

Restriction vs. Constriction

- History can provide important clues
  - Constrictive pericarditis
    - History of TB, trauma, pericarditis, collagen vascular disorders
  - Restrictive cardiomyopathy
    - Amyloidosis, hemochromatosis
  - Mixed
    - Mediastinal radiation, cardiac surgery

Treatment

- No satisfactory medical therapy
- Drug therapy must be used with caution
  - Diuretics for extremely high filling pressures
  - Vasodilators may decrease filling pressure
  - (?) Calcium channel blockers to improve diastolic compliance
  - Digitalis and other inotropic agents are not indicated

Arrhythmogenic Right Ventricular Cardiomyopathy

Arrhythmogenic RV Cardiomyopathy

- Characterized by fibroadipose replacement of segments of the free wall of the right ventricle
- Familial and progressive
- Predominately found in young adults
- Cause of young adult sudden death
- ICD implantation in ALL patients who are symptomatic with arrhythmias
- ICD implantation vs. anti-arrhythmic meds in asymptomatic patients?
- Prognosis?

Underwriting Cardiomyopathy

Underwriting Tips

- Diagnosis?
  - When diagnosed?
  - Cardiologist?
  - Family Hx
    - SCD
- Serial echo findings?
  - EF
- Treatment?
- Any other co-morbidities?
  - CAD
  - CVD
  - COPD

Unfavorable Cases of Cardiomyopathy

- Young age (<30 years)
- Family history of sudden death
- Aborted sudden death
- History of sustained SVT, AF, VT, 2’ or 3’ heart block
- Syncopal attacks
- Pacer and/or implanted defibrillator
- Prohibited from participating in any exercise
- Enlarged heart (CT >55%)
- EF <40% and/or generalized mod/severe hypokinesis
- CHF
- Substantial septal hypertrophy (> 2cm)

Case Studies

Quick Quote

- 67-year-old male
- History of hypertension, diabetes and hyperlipidemia
- Normal height/weight
- Blood pressure readings
  - 5-08 160/100
  - 8-08 150/108
  - 10-08 122/88
  - 12-08 136/98

Quick Quote (cont'd)

- 2004 and 2006 resting EKGs with T wave inversions in lateral leads
- 2006 stress test, exercised 7:30 minutes, flat ST depression at peak, borderline for ischemia
- 2008 echocardiogram
  - LVH (IVS 16mm, PW 15mm)
  - Left atrial enlargement (42mm)
  - Mild MR with SAM (systolic anterior motion)
  - Mild to moderate E:A reversal
  - Mild aortic insufficiency
  - No wall motion abnormalities, EF 55%

- Opinion?

Informal

- 81-year-old female
- History of hypertension, palpations/PAF
- Normal height/weight
- PAF history noted in 2004 with no formal evaluation at that time
- No indication of any PAF since 2006
- 10/09 Holter monitor with several non-sustained runs of SVT, rare pac, rare pvc's

Informal (cont'd)

- 10/09 Echo findings:

A/O Root Diameter (Diastole): 2.9 cm (Normal: 2.0 - 3.8 cm)
LA Dimension (Systole): 4.4 cm (Normal: 1.9 - 4.0 cm)
Right Ventricular Dimension (Diastole): 2.6 cm (Normal: 2.9 cm max)
Septal Thickness (Diastole): 2.0 cm (Normal: 0.7 - 1.1 cm)
Left Ventricular Size (Diastole): 3.1 cm (Normal: 3.5 - 5.6 cm)
Posterior Wall Thickness (Diastole): 1.1 cm (Normal: 0.7 - 1.1 cm)
Left Ventricular Size (Systole): 2.0 cm (Normal: 2.8 - 4.0 cm)

Aortic valve area: 2.3 cm/sq (mild 1.1-1.9 cm/sq moderate 0.75-1.1 cm/sq severe <0.75 cm/sq)
V1 PG: 101 cm/sec V2 PG: 142 cm/sec
Mitra Valve P1/2 time: 92 m/sec (mild 90-150 m/sec moderate 150-210 m/sec severe >220 m/sec)
Mitral valve area: 2.3 cm/sq (mild 1.1-1.9 cm/sq moderate 1.0-1.5 cm/sq severe <0.75 cm/sq)
Estimated Systolic Pulmonary Artery Pressure: 35 mmHg

Estimated Ejection Fraction: 65 % (Normal: 50 - 75%)

CONCLUSIONS:
1) The patient was in a normal sinus rhythm throughout study.
2) The left ventricle reveals left ventricular hypertrophy
3) The right ventricle is of normal size, wall thickness, and wall motion.
4) There is left atrial enlargement
5) The mitral valve is sclerotic and has mitral annular calcification
6) Analysis of the mitral inflow velocity reveals diastolic dysfunction
7) The aortic valve is sclerotic
8) Doppler interrogation reveals a normal aortic outflow velocity
9) The pulmonic valve is normal The tricuspid valve has 1+ regurgitation
10) Analysis of the tricuspid regurgitant jet reveals no pulmonary hypertension.
11) There is no pericardial thickening or effusion noted

RECOMMENDATIONS:

- Opinion?

QUESTIONS?