VALVULAR HEART DISEASE

BOARD REVIEW

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Case

- 52 yo male comes to your office for a routine visit and has no concerns.
- PE-mildly hypertensive
  - You find a 2/6 systolic murmur at the right second intercostal space
- What valvular lesion does he have?
- What can you do to confirm your suspicion besides an echo? ☺

Goals and Objectives

- Discuss common valvular lesions
- Examine the clinical context by which valvular lesions could be determined
- Recognize different physical exam maneuvers or findings to assess valvular lesions
- Understand other key laboratory or radiologic studies to assess valvular lesions
- Recognize treatment options for each of the common valvular lesions

PASS YOUR BOARDS!

☺

KEY CONCEPTS

- Understand key historical facts to guide you in understanding valvular conditions
- Use location of the murmur to start your assessment
- Use other physical exam findings to help you narrow your thought process
- Then use other studies to further refine your thoughts

AORTIC STENOSIS

Etiology

- Congenital bicuspid valve
  - 1% of population
  - Male predominance (age: 40-60's)
- Rheumatic heart disease
  - Less common cause in developed nations
- Calcification of normal valve
  - Formerly considered degenerative
  - Now clearly active inflammatory process similar to CAD
    - Lipid accumulation, inflammation, calcification
  - Usually occurs in 60s to 80s
- Congenital Malformation
  - More common cause in young adults

Pathophysiology

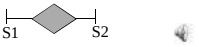
- AS obstruction develops gradually
  - LV hypertrophies to accommodate systolic pressure overload
- Concentric hypertrophy is appropriate and beneficial for adaptation
  - However adaptation causes consequences
    - Reduced coronary blood flow
    - Subendocardial ischemia, contributing to systolic or diastolic heart failure
    - Increased sensitivity to ischemic injury with larger infarcts and higher mortality rates
    - Elderly women at risk of inappropriate degree of hypertrophy leading to higher morbidity and mortality

Symptoms

- Key symptoms: Angina, Syncope, CHF
  - Angina
    - 2^0 to reduced coronary blood flow reserve
    - 50% dead in 5 yrs (w/o AVR)
    - Risk of sudden death: 10%
  - Syncope
    - Usually related to exertion
    - 50% dead in 3 yrs (w/o AVR)
    - Risk of sudden death: 15%
  - CHF
    - 50% dead in 2 yrs (w/o AVR)
    - Risk of sudden death: 25%

Physical Findings

- Diamond shaped, crescendo-decrescendo systolic murmur



  - Loudest in aortic area
  - Radiates to carotids
  - Mild disease: murmurs peaks in early systole
  - Worsening disease: murmur peaks progressively later
  - Can radiate to apex causing confusion with MR (Gallaverdin phenomenon)
- Delay and prolongation of carotid pulse (parvus et tardus)
- S4 with left ventricular hypertrophy
- S3 with left ventricular failure
- Exclusion of severe AS is a normally split S2

Diagnosis

- Normal valve area: 3-4 cm^2
- EKG: LV hypertrophy
- CXR: Usually non-diagnostic
- Echo:
  - Obtain when murmur is >3/6, a single S2 or symptoms due to AS
- Cardiac cath
  - Recommended in pts:
    - Echo with discrepant results
    - Patients at risk for concomitant CAD
    - Patients with chest pain, LV dysfunction
- Exercise Testing
  - Do not perform in symptomatic pts given high risk
  - Okay to perform in asymptomatic pts or in patients with indeterminate sx

Classification of Echo Findings

	Aortic Sclerosis	Mild	Moderate	Severe
Aortic Jet Velocity (m/s)	≤2.5	2.6-2.9	3.0-4.0	>4.0
Mean gradient (mmHg)	...	<20	20-40	>40
Aortic Valve area (cm2)	...	>1.5	1.0-1.5	<1.0
Indexed aortic valve area (cm2/m2)		>0.85	0.60-0.85	<0.6
Velocity ratio		>0.50	0.25-0.50	<0.25

Clinical Management

- Asymptomatic pts
  - With mild stenosis: valve narrowing occurs with a mean gradient increase of 6-7mmHg/yr and a decrease in valve area of 0.1cm2/yr
- Echocardiography
  - Obtain to assess severity
  - Re-evaluation in known AS if changing signs or symptoms
  - During pregnancy if known AS
  - TTE every year for asymptomatic pts with severe AS
  - TTE every 1-2 years for moderate AS
  - TTE every 3-5 years for mild AS
  - Clinical evaluation for all types yearly

Treatment

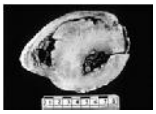
- Aortic Valve Replacement (Class 1 indications)
  - Symptomatic pts with severe AS
  - Severe AS undergoing CABG
  - Severe AS in pts undergoing surgery on the aorta or other valves
  - Severe AS and LV systolic dysfunction
- Antibiotic prophylaxis:
  - For recurrent rheumatic fever in pts with rheumatic AS

Transcatheter Aortic Valve Replacement

- High risk pts with aortic stenosis who are not surgical candidates
- Partner Trial & FRANCE 2 Trial
  - Lower death rate at 30 d and 1 year
  - Major vascular complications significantly higher at 30d
    - Partner trial: 3.8% vs 2.1%; at 2 yrs: 7.7% vs 4.9%
  - More frequent adverse events in surgical group-major bleeding, new onset afib
  - Increased risk of heart block (2 fold)
  - Paravalvular arotic regurgitation

Hypertrophic Cardiomyopathy

Etiology



- Common genetic abnormality
  - 1 in 500 persons
- Autosomal dominant pattern
- Mutations found in 9 genes encoding sarcomeric proteins (over 900 mutations)
  - MYH7 (cardiac β myosin heavy chain), MYBPC3 (cardiac myosin binding protein)
- M:F 2:1
- Family history of "sudden death" or arrhythmia
- Obstruction caused by hypertrophied septum and abnormal anterior motion of the mitral valve

Symptoms

- Present between 20-40 years
  - Occasionally after 50 y.o.
- Symptoms not proportional to degree of hypertrophy
  - Dyspnea on exertion: most common
  - Chest pain
  - Dizziness
  - Syncope: during or after heavy exertion
- Atrial fibrillation
  - Occurs commonly in HCM

Athletes and HCM

- Questions to ask:
  - Have you ever passed out or nearly passed out during exercise?
  - Have you ever passed out or nearly passed out after exercise?
  - Have you ever had discomfort, pain or pressure in your chest during exercise?
  - Does your heart race or skip beats during exercise?
  - Has a doctor ever told you that you have a hear murmur?
  - Has a doctor ever ordered a test for your heart (EKG, echo)?
  - Has anyone in your family died for no apparent reason?
  - Does anyone in your family have a heart problem?
  - Has any family member or relative died of heart problems or of sudden death before age 50?

Physical Exam

- Systolic ejection murmur
  - Less rasping than valvular aortic stenosis
  - Maximal between apex and LLSB
  - NOT well transmitted into the neck
  - Loudest with the following maneuvers:
    - Straining period of valsalva maneuver
    - Standing after squatting
- Bifid carotid pulse
  - 80% of blood volume ejected during the first half of systole
- Audible S4, Palpable A wave

Diagnosis

- Echo
  - Asymmetrical septal hypertrophy
- EKG
  - LVH and increased Q waves

Treatment

- No symptoms-no treatment
- If symptoms present
  - Therapy aimed to improve diastolic filling
    - B adrenergic blocking drugs
    - Verapamil
  - Surgical treatment
    - If refractory severe symptoms (<5% of pts)
    - Myotomy-myectomy or alcohol injection

AORTIC REGURGITATION

Etiology

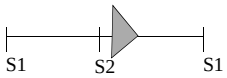
- Any disease conditions which renders the aortic leaflets incompetent
  - Rheumatic heart disease (most common worldwide)
  - Bicuspid valve, calcific degeneration
  - Hypertension
  - Syphilis
  - Connective tissue disorders (Marfan)
  - Arthritic disorders (ankylosing spondylitis, rheumatoid arthritis, Reactive arthritis syndrome)
  - Cystic medial necrosis
  - Aortic dissection
  - Infective endocarditis

Symptoms

- Acute
  - Acute left sided heart failure with shock
  - Pulmonary edema
  - Angina (may occur with flushing)
- Chronic
  - Left sided heart failure symptoms: DOE, orthopnea, fatigue secondary to volume and pressure overload
  - When sx develop mortality increases to 10-20% per year

Physical Findings

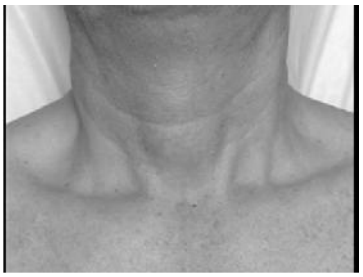
- High pitched, blowing, decrescendo diastolic (early) murmur at LSB in expiration (sitting forward)
- Wide pulse pressure (e.g. 140/40)



Physical Findings

- Other findings with chronic AI:
  - Head bobbing (De Musset's)
  - Pistol shot sounds over the femoral arteries (Duroziez)
  - Pulsations in the nail bed (Quincke's)
  - Water hammer pulse (Corrigan)
  - Retinal pulsations
  - Increase in femoral systolic pressure >40mmHg over brachial artery

Other Physical Exam



Testing

- Asymptomatic pts with mild AR, little to no LV dilatation, normal LV systolic function
  - Seen on yearly basis but echo not necessary every year (every 2-3 yrs)
- Asymptomatic pts, severe AR, significant LV dilatation, normal systolic function
  - H&P every 6 months, echo every 6-12 months
- Advanced LV dilatation (EDV>70mm)
  - Risk of developing symptoms 10-20% per year
  - Echo every 4-6 months

Treatment

- Asymptomatic patients with normal left ventricular function (but LV dilation)
  - Afterload reduction is recommended
    - Nifedipine, Hydralazine, ACE-i
    - 2005 NJEM article
      - 95 pts with severe aortic regurgitation
        - Treated with nifidepine (20 mg BID or enalapril (20 mg QD) or no treatment
        - Rate of aortic valve replacement the same in all groups
      - Criticisms of the study
        - Small number of pts, inadequate control of BP
      - Final conclusion: the current doses recommended by an earlier study do not delay the progress of disease requiring valve replacement

Treatment

- Class I indication for surgery
  - Symptomatic patients with severe AR
  - Asymptomatic pts with chronic severe AR and LV systolic dysfunction (EF<50%)
  - Pt with chronic severe AR undergoing CABG
- Acute Aortic Regurgitation
  - Any signs or symptoms even if mild dictates surgery
  - Medical mortality as high as 75%

MITRAL STENOSIS

ETIOLOGY

- F:M 2:1
  - Develops in 40s and 50s
- Generally rheumatic in nature; rarely is congenital
- Pure or predominant MS occurs in approximately 40% of all patients with rheumatic heart disease

NATURAL HISTORY

- Continuous, progressive, lifelong disease, usually consisting of a slow, stable course in the early years
- Progressive acceleration in the later years
- 20-40 years from the occurrence of rheumatic fever to onset of symptoms
- Almost another 10 years before symptoms become disabling
- 0-15% 10 year survival once significant/limiting symptoms develop

NATURAL HISTORY

- Mortality
  - Progressive heart failure 60-70% of pts
  - Systemic embolism 10-30% of pts
  - Pulmonary embolism 10%
  - Infection in 1-5% of pts

PATHOPHYSIOLOGY

- Normal valve orifice: 4-5 cm^2
- Narrowing of valve area before producing symptoms: <2.5 cm^2
- Valve area >1.5 cm^2 usually does not produce symptoms at rest
- Elevated left atrial pressure raises pulmonary venous pressure causing dyspnea

SYMPTOMS AND COMPLICATIONS

- Elevations of pulmonary capillary pressure:
  - extreme exertion, excitement, fever, severe anemia, paroxysmal tachycardia, sexual intercourse, pregnancy, and thyrotoxicosis
    - Orthopnea, PND, and pulmonary edema
    - If RV failure: ascites, edema
    - Hemoptysis: common in MS but not in other causes of left atrial hypertension
      - High left atrial pressure ruptures small bronchial veins

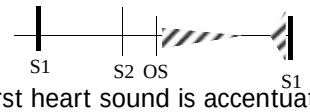
SYMPTOMS AND COMPLICATIONS

- Atrial arrhythmias
  - Premature contractions, paroxysmal tachycardia, flutter and fibrillation
    - Occur with increasing frequency in moderately severe MS existing for several years
  - Atrial fibrillation develops in 30% to 40% of patients with symptomatic MS
    - Risk of arterial embolization (10-30%), especially stroke, is significantly increased in patients with atrial fibrillation

SYMPTOMS AND COMPLICATIONS

- Recurrent pulmonary emboli
  - sometimes with infarction
  - important cause of morbidity and mortality late in MS
- Pulmonary infections
  - bronchitis, bronchopneumonia, lobar pneumonia
- Chest pain
  - 10% of pts with severe MS

PHYSICAL DIAGNOSIS



- First heart sound is accentuated and snapping
- Opening snap after aortic valve closure
- Low pitch diastolic rumble at the apex
- Presystolic murmur

Clinical Management

- Afib
  - Digoxin, BB
- Anticoagulation
  - Indicated in MS with afib
  - MS with prior embolic event
  - MS and left atrial thrombus

Clinical Management

- Asymptomatic patients
  - If normal sinus rhythm, no therapy
  - Symptoms of mild dyspnea and orthopnea: treat with diuretics
  - Limit exercise to symptoms
- Symptomatic pts
  - mechanical correction for increased symptoms or pulmonary hypertension
    - Mitral valve valvotomy
    - Mitral valve replacement or open commissurotomy (if unfavorable valve anatomy)

MITRAL REGURGITATION

ETIOLOGY

- Chronic rheumatic heart disease is the cause in about 1/3rd of patients
  - Rheumatic MR occurs more frequently in males
- Degenerative forms more common in US
- Most common in the US is MVP
- Functional MR occurs with any cause of mitral annulus enlargement or myocardial infarction
- Acute MR may occur secondary to infective endocarditis, acute myocardial infarction, or trauma

SYMPTOMS

- Fatigue, exertional dyspnea, and orthopnea are most prominent with chronic, severe MR
- Hemopytsis and system embolization occur less frequently in MR

Physical Findings

- Usually apical holosystolic murmur with radiation to the axillae

- Increases with expiration, squatting, and hand grip
- S3 may be a finding of severe MR

EVALUATION

- EKG: left ventricular hypertrophy
- CXR: left atrial enlargement
- Echo: extent of left atrial and ventricular enlargement

Evaluation

- Echo
  - Determining cause of MR
  - Assessment of LV function
  - Change in signs or symptoms
  - Baseline determination of LV size and function

Follow Up

- Asymptomatic pt with severe MR
  - Follow up every 6-12 months
- LV enlargement, pulmonary HTN or LV dysfunction
  - No participation in competitive sports

Treatment

- Medical
  - Restrict physical activities that produce dyspnea and fatigue
  - Reduce sodium intake and induce sodium excretion with diuretics
  - Vasodilators, BB, digitalis, and ACE-I are used
- Surgical: Symptomatic MR
  - Pts with severe MR and severe limitations despite optimal medical management

Treatment

- Surgery
  - Asymptomatic MR
    - NJEM 2005
      - Effective regurgitant orifice independently predicted survival
        - 40mm2 orifice: 5x risk of death from cardiac causes and cardiac events
      - 5 yr death rate of any cause-22%
      - 5 yr adverse event (death from cardiac causes, heart failure, new afib)-33%

Treatment

- Medical Therapy
  - No generally accepted therapy
  - If LV dysfunction, ACEi, BB and pacing have been shown to reduce severity of MR
- Surgery (class I indication)
  - Symptomatic pts with acute, severe MR
  - Chronic MR with functional class II, III or IV symptoms but no LV dysfunction
  - Asymptomatic pts with severe MR and mild-mod LV dysfunction

MITRAL VALVE PROLAPSE

Etiology

- Most common form of valvular heart disease- 2-6% of the population
- MVP has become the most common cause of isolated severe MR
- Cause of MVP is unknown in a majority of pts
- More common in females between ages of 14 and 30years

Symptoms

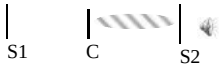
- Many have chest pain which is difficult to evaluate.
  - Often substernal, prolonged, poorly related to exertion, and rarely resembles typical angina
- Arrythmias-PVCs, PSVT, VT-have been reported and may cause palpitations, lightheadness, and syncope

Symptoms cont'd

- Transient cerebral ischemic attacks have been reported.
- Infective endocarditis may occur in patients with MR associated with MVP
- Sudden death is a rare complication

Physical Findings

- Most important finding is the mid or late systolic click.



- May be followed by high pitched late systolic crescendo-decrescendo murmur, occasionally "whooping" or "honking" at the apex

Physical Findings

- Standing and valsalva maneuver move the click closer to the first sound

CLINICAL COURSE

- Most with benign course
- Approx 10% of pts (with thickened leaflets)
  - Infective endocarditis
  - Stroke
  - Progression to severe MR (men 2x more)
  - Sudden death

Management

- ASA for symptomatic pts with MVP who experience TIA
- MVP with afib: coumadin if age >65 or with HTN, MR murmur or heart failure
- MVP with afib: asa if age <65 and no history of above
- MVP, h/o stroke: warfarin recommended for pts with MR, afib, LA thrombus

Treatment

- Reassurance of the asymptomatic patient without severe MR or arrhythmias
- Infective endocarditis prophylaxis in patients with a systolic murmur
- Beta blocker treatment for atypical chest pain

TRICUSPID STENOSIS

Etiology, Symptoms, Physical Findings

- Rheumatic fever is the most common cause
- Rare conditions such as carcinoid tumor, right atrial myxoma
- Symptoms of fatigue and dyspnea
- Diastolic murmur heard best along the LLSB

TRICUSPID REGURGITATION

ETIOLOGY

- Most commonly secondary to hemodynamic load on the right ventricle
- Primary valve abnormality
  - Infective endocarditis
  - Carcinoid syndrome
  - Rheumatic involvement of tricuspid valve
- Secondary valve abnormality
  - Pulmonary hypertension
  - Left ventricular dysfunction

SYMPTOMS

- Right sided failure
  - High jugular venous pressure
  - Ascites
  - Lower extremity edema

DIAGNOSIS

- Increased JVP
- Hepatic enlargement/liver pulsation
- RV enlargement (parasternal lift)
- Holosystolic murmur
  - Heard best left lower sternal border
  - Increases with inspiration

TREATMENT

- Aimed at underlying cause
  - LV dysfunction
  - RV dysfunction
    - Pulmonary hypertension
- Rarely ever do surgery

INFECTIVE ENDOCARDITIS AND COMPLICATIONS

Native Valve Endocarditis

- Symptoms begin within 2 weeks of the inciting bacteremia
- Acute endocarditis is caused by S. aureus (50-70%)
- In the non-IVDU, the aortic valve is involved

Systemic manifestations

- Fever, drenching night sweats, arthralgias, myalgias, and weight loss
- Murmurs, valve ring abscess, myocardial infarction, myocardial abscess, diffuse myocarditis are all complications
- CHF is the most common complication, occurring more with left sided endocarditis

Extracardiac Manifestations

- Embolic events
  - Infarction of the lungs in right sided lesions
  - Infarction of brain, spleen, or kidneys in left sided endocarditis
- Immunologic manifestations:
  - Roth spots, glomerulonephritis, meningitis, petechiae, splinter hemorrhages, Osler's nodes, Janeway lesions

IVDU Endocarditis

General

- Murmurs and heart failure usually absent
- Septic pulmonary complications occur in 75% of these patients
- S. aureus is the usual pathogen

Treatment

- S. aureus
  - Nafcillin, Oxacillin with or without Gentamycin
  - If PCN allergy: Cefazolin with or without Gentamycin or Vancomycin
  - If MRSA: Vancomycin

Physical Exam Maneuvers

P.E. Maneuvers

- Respiration
  - Inspiration: increases right sided murmurs
  - Expiration: increases left sided murmurs
- Valsalva
  - All murmurs decrease except HCM and MVP which increase in intensity
- Standing
  - All murmurs decrease except HCM and MVP which increase in intensity

P.E. Maneuvers

- Squatting/Passive Leg Raise
  - All murmurs increase in intensity except HCM and MVP which decrease in intensity
- Hand grip exercise
  - Murmurs of PS, MS, MR, VSD, and AR increase
  - HCM often decreases with near max hand grip

P.E. Maneuvers

- Postventricular premature beat/afib
  - Systolic murmurs due to AV valve regurgitation do not change, diminish or become shorter (MVP)
  - Murmurs due to stenosis increase in intensity following a premature beat
- Transient arterial occlusion
  - MR, VSD, and AR only increase in intensity

Common Valvular Heart Diseases:

Systolic Murmurs
- Aortic stenosis
- Mitral insufficiency
- Mitral valve prolapse
- Tricuspid insufficiency

Diastolic Murmurs
- Aortic insufficiency
- Mitral stenosis