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
    - 2x 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 (Gallavardin 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 cm²
- 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 unrevealed 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

<table>
<thead>
<tr>
<th>Aortic Jet Velocity (m/s)</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
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</thead>
<tbody>
<tr>
<td>≤2.3</td>
<td>2.4-2.9</td>
<td>3.0-4.0</td>
<td>&gt;4.0</td>
</tr>
</tbody>
</table>

| 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) | ≤0.60 | 0.60-0.85 | >0.85 |

| Velocity ratio           | ≤0.25 | 0.25-0.50 | >0.50 |

| S1 | S2 |
Clinical Management

- Asymptomatic pts
  - With mild stenosis: valve narrowing occurs with a mean gradient increase of 6-7 mmHg/yr and a decrease in valve area of 0.1 cm²/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 aortic 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

- 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

Testing

- Asymptomatic patients with normal left ventricular function (but LV dilation)
  - Afterload reduction is recommended
    - Nifedipine, Hydralazine, ACE-inhibitor
    - 2005 N Engl J Med article
      - 95 pts with severe aortic regurgitation
        - Treated with nifedipine (20 mg BID or enalapril (20 mg OD) 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
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 cm²
- Narrowing of valve area before producing symptoms: <2.5 cm²
- Valve area >1.5 cm² 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

- 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

- 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
- Hemoptysis and systemic 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
    - 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%
  - NJ EM 2005

- Effective regurgitant orifice independently predicted survival

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-moderate 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 30 years
Symptoms

- Many have chest pain which is difficult to evaluate.
  - Often substernal, prolonged, poorly related to exertion, and rarely resembles typical angina
- Arrhythmias-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.
  \[ \text{s1} \quad \text{c} \quad \text{s2} \]
- 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

\[ S_1 \ 
\text{C} \ 
S_2 \]

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:**

<table>
<thead>
<tr>
<th>Systolic Murmurs</th>
<th>Diastolic Murmurs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic stenosis</td>
<td></td>
</tr>
<tr>
<td>Mitral insufficiency</td>
<td></td>
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<tr>
<td>Mitral valve prolapse</td>
<td></td>
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<tr>
<td>Tricuspid insufficiency</td>
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S1  S2  S1