Case I: 22 year old man with exertional syncope

Presented to ER after an episode of sudden loss of consciousness immediately after a 3 mile jog. He awoke completely after 3-4 minutes. Denied chest pain, shortness of breath and palpitation.

- FH: positive for SD in a cousin
- PMH: Uncomplicated rotator cuff repair
- PE: HR = 60, BP = 90/60, JVP = 4 cm, S4 gallop, no murmur
Case 1: What is the next best test?

1. Treadmill ETT
2. Event monitor
3. Dobutamine echocardiography
4. Transthoracic echocardiogram
5. Serial troponin I levels
Case 1: Educational Objectives

- Know the causes and recommended evaluation for syncope
- Recognize the symptoms and signs of hypertrophic cardiomyopathy
- Know the risk factors for sudden death in a patient with hypertrophic cardiomyopathy

Cardiac Syncope

**Structural causes**
- Common
  - AS - usually exertional
  - Hypertrophic obstructive cardiomyopathy - often post-exercise or due to arrhythmias
- Less common
  - PE, dissection, tamponade
- Uncommon
  - Pulmonary HTN, atrial myxoma, subclavian steal

**Arrhythmic causes**
- Bradycardia
  - Sinus bradycardia
  - AV block
  - Carotid sinus hypersensitivity
- Tachycardia
  - SVT rare
  - VT
Syncope Evaluation

- Careful history and physical examination crucial in work-up of syncope
  - Reveals probable cause in 50% of patients
  - Vasovagal syncope is strongly suggested by characteristic prodrome and may be confirmed by Tilt table test
- EKG
- Echo indicated with h/o heart disease, abnl PE or EKG or elderly
- Holter or event monitor
- Tilt Table

Exertional syncope often ominous

- Differential diagnosis:
  - Obstruction to left ventricular outflow (aortic stenosis and hypertrophic cardiomyopathy)
  - Ventricular tachycardia
  - Pulmonary hypertension
  - Neurocardiogenic syncope
- Treadmill ETT should be delayed until structural causes excluded
Overall Causes of Syncope

- Cardiac: 18%
- Neurologic: 10%
- Vasovagal: 24%
- Orthostatic: 8%
- Medications: 3%
- Unknown: 37%

Cardiac Structural Causes of Syncope:

- Common:
  - Aortic stenosis:
    - Usually exertional
    - May have associated angina or heart failure.
  - Hypertrophic cardiomyopathy:
    - Affects all ages
    - Obstructive form may manifest in the setting of ↓preload or vasodilation
    - Non-obstructive form due to ventricular arrhythmias
Cardiac Structural Causes of Syncope:

- **Less common:**
  - Pulmonary embolism
  - Aortic dissection
  - Cardiac tamponade
- **Uncommon:**
  - Pulmonary hypertension
  - Atrial myxoma
  - Subclavian steal

Arrhythmic causes of cardiac syncope

- **Bradycardia:**
  - Inappropriate or marked sinus bradycardia:
    - Sick sinus syndrome
    - Medications (B-blocker, calcium channel blocker, etc.)
  - AV block (2nd, 3rd degree):
    - Age-related conduction disease
    - Medications
    - Ischemia
    - Carotid sinus hypersensitivity
- **Tachycardia:**
  - Supraventricular tachycardias: Rare cause of syncope
  - Ventricular tachycardia: Most often due to structural and/or ischemic heart disease.
Hypertrophic Cardiomyopathy

- Autosomal dominant inheritance pattern with variable penetrance
- Usually presents in adolescent or young adult but can present at any age
- Triad of symptoms:
  - Chest pain, Dyspnea and Syncope

HCM: Physical Examination

- Non-obstructive: S4, sustained apical impulse
- Obstructive:
  - Bisferiens carotid pulse ("spike and dome")
  - “Triple” apical impulse
  - Systolic murmur that increases with standing or strain phase of Valsalva and decreases with passive leg raising or squatting, increases post-PVC
- Dynamic:
  - Occasionally may hear murmur only with maneuver
Hypertrophic Cardiomyopathy

- EKG: LVH
- Echocardiogram:
  - Left ventricular hypertrophy
  - Obstruction: associated with systolic anterior motion of the mitral valve, may be provoked with amyl nitrite
- Pharmacologic treatment
  - Beta-blocker
  - Calcium channel blocker
  - Avoid any vasodilator or positive inotropic agent (Dobutamine echocardiography relatively contraindicated)

HCM

- Other Treatment:
  - Avoid high intensity sports
  - Surgical myomectomy or Alcohol septal ablation in obstructive form only and only in pts refractory to med Rx
  - ICD for those at high risk for SD
Case 1: What is the next best test?

1. Treadmill ETT
2. Event monitor
3. Dobutamine echocardiography
4. Transthoracic echocardiogram
5. Serial troponin I levels

Case 2: 76 yo man with paroxysmal atrial fibrillation and HTN

Asymptomatic and presents for routine medical care

- PMH: TURP 4 yrs ago at which time A Fib was dx’d on preop EKG
- Meds: ASA 325 qd, Metoprolol XL 25 qd
- Exercise tolerance: 30 minutes daily on TM, non-smoker
- PE: BP 150/85, HR 70 irreg irreg, JVP nl, 2/6 SEM at base
- EKG: Atrial fibrillation at rate of 65 -70, Borderline LVH
- Echocardiogram: Normal biventricular function, mild left ventricular hypertrophy, moderate biatrial enlargement, aortic valve sclerosis
Case 2: In addition to the addition of HCTZ 12.5 mg, you should:

1. Increase metoprolol XL to 50 qd
2. Start amiodarone 200 mg bid for 4 weeks and admit for cardioversion
3. Start warfarin 5 mg qd and anticoagulate to an INR of 2.0 - 3.0
4. Stop metoprolol and start diltiazem SR 240 qd
5. Continue present medical regimen

Atrial fibrillation, Stroke Risk and Rate Control

- The stroke rate in non-rheumatic AF is about 5%/year
- Risk factors based on CHADS2 > 2
  - CHF (MI) - 1
  - HTN - 1
  - Age ≥ 75 yrs - 1
  - Diabetes - 1
  - Secondary protection for previous history of stroke, TIA (systemic embolization) - 2
- Rate control defined in AFFIRM study as resting rate of 80 bpm or 110 bpm after 6 minute walk (Thus no need to increase metoprolol or change to diltiazem)
- Embolic risk similar in rate control vs. rhythm control arms of AFFIRM
Agents used for Rate Control in Atrial Fibrillation

Initiate Rx with beta-blocker, Diltiazem or verapamil, Digoxin or amiodarone (if CHF present)

Inadequate Rate Control or Exercise intolerance

Add second agent

Adequate Rate Control with good exercise tolerance

Inadequate Rate Control or Exercise intolerance

Consider AV node ablation + VVIR pacing

Management of atrial fibrillation in the elderly

- Recent data from the AFFIRM trial suggests that maintenance of NSR does not improve QOL or increase life expectancy in patients > 65 yrs of age
- Rate control should be achieved
- Anticoagulation with Warfarin is indicated in patients with PAF as well as those in permanent atrial fibrillation.
- ASA alone is considered adequate only for patients < 75 yrs of age with "lone" atrial fibrillation (i.e. no structural heart disease or HTN)
- ACTIVE Trial, NEJM 5/2009: “In patients with atrial fibrillation for whom vitamin K−antagonist therapy was unsuitable, the addition of clopidogrel to aspirin reduced the risk of major vascular events (6.8% vs. 7.6%/year; stroke 2.4% vs. 3.3%) and increased the risk of major hemorrhage (2.0% vs. 1.3%)”
- Future guidelines??
Additional caveats re: Atrial fibrillation

- Unstable patient with atrial fibrillation
  - Electrical cardioversion is indicated
- Anticoagulation indicated prior to elective cardioversion (electrical or pharmacological) for AF > 48 hours
  (Amiodarone should not be started prior to Anticoagulation)
- Atrial fibrillation in setting of WPW
  - Recognize
    - Broad bizarre QRS complexes varying in width
    - Irregularly irregular rhythm
  - Avoid AV nodal blocking agents
  - Treatment
    - Cardioversion if unstable
    - IV Procainamide

Atrial flutter

- Anticoagulation recommendations are similar
- High success rate for ablation in typical flutter
- Treatment algorithm otherwise similar
Case 2: In addition to the addition of HCTZ 12.5 mg, you should:

1. Increase metoprolol XL to 50 qd
2. Start amiodarone 200 mg bid for 4 weeks and admit for cardioversion
3. Start warfarin 5 mg qd and anticoagulate to an INR of 2.0 - 3.0
4. Stop metoprolol and start diltiazem SR 240 qd
5. Continue present medical regimen

Case 3: 71 year old woman with history of CHF having episodic lightheadedness

Patient has had well-compensated CHF for 1 year after a hospital admission with pulmonary edema when she ruled out for MI. She has had no further admissions. Two weeks ago, she developed episodes of lightheadedness unrelated to exertion or standing. No frank syncope and no palpitation

- **PMH:** Hypercholesterolemia, HTN
- **Medications:**
  - Lisinopril 10 mg qd
  - Furosemide 20 mg qd
  - Digoxin 0.25 mg qd
  - KCL 20 meq qd
  - Metoprolol XL 25 mg qd
Case 3

- Physical examination:
  - BP 106/60 lying, 104/64 standing
  - HR 56 irreg
  - JVP estimated at 6 cm
  - Lungs clear
  - CV exam: non-displaced PMI, intermittent S4
  - No edema

- Echocardiogram:
  - LV size normal, ejection fraction 47% without segmental wall motion abnormalities

- Labs: BUN 24, creatinine 1.3, digoxin level 1.9

Case 3: ECG
Case 3: The next step in her management should be:

1. Decrease the dose of lisinopril to 5 mg qd
2. Discontinue metoprolol XL
3. Discontinue digoxin
4. Decrease the dose of furosemide to 20 mg qod.
5. Admit to the hospital for urgent permanent pacemaker placement

Case 3: Educational Objectives

- Recognize types of second-degree AV block
- Recognize digoxin toxicity
- Know indications and target level for digoxin in CHF
Discussion:

- Mobitz I 2° AV block may be seen with digoxin excess.
- Mild renal insufficiency increases her risk for digoxin toxicity.
- Digoxin not clearly indicated in this asymptomatic patient with only mildly reduced ejection fraction.
- Her blood pressure is adequately controlled and it is unlikely that metoprolol is causing the Wenckebach conduction so it should be continued at the current dose. ACE inhibitor should be continued.
- Given her prior hx of pulmonary edema, furosemide should be maintained.

Second-degree Atrioventricular Block

- Type I or Wenckebach
  - Progressive PR prolongation before the blocked beat
  - QRS usually narrow
  - Usually supra-His ie. AV nodal
  - Responds to atropine
- Type II
  - No progressive PR prolongation
  - QRS usually wide
  - Usually intra- or infra-His
  - Usually no response to atropine
  - May have paradoxical slowing
Indications for Permanent Pacemaker

- Reversible causes should be excluded
- If culprit medication cannot be discontinued (eg. Beta blocker), pacemaker may be considered
- **Symptomatic** bradycardia underlies the majority of indications
- In **Asymptomatic** patients, pacing may be indicated for:
  - Pauses > 3 seconds during waking hours
  - Heart rates < 40 bpm during waking hours
  - Second degree type II AV block (class II indication)
- Other Class I indications:
  - Third-degree AV block and advanced second degree AV block

Case 3: The next step in her management should be:

1. Decrease the dose of lisinopril to 5 mg qd
2. Discontinue metoprolol XL
3. **Discontinue digoxin**
4. Decrease the dose of furosemide to 20 mg qod
5. Admit to the hospital for urgent permanent pacemaker placement
Case 4: 22 yo woman referred for evaluation of murmur in 24th week of gestation

Mild fatigue and dyspnea during 1st trimester, now improved. However, she experiences an occasional "skipped beat", similar to syx she had before she conceived. No syncope, presyncope or chest pain.

- PMH: Juvenile rheumatoid arthritis - in remission
- PE: BP 100/55, HR - 82
  - JVP - 7 cm
  - Carotids - brisk
  - Lungs - clear
  - Cardiac - PMI 1cm displaced, nl heart sounds, 3/6 early peaking systolic murmur heard throughout precordium, no diastolic murmur, S3
  - Extremities - trace edema
- EKG - nl

The next step in your evaluation is:

1. Echocardiogram
2. Holter monitor
3. Event monitor
4. Reassurance
Case 4: Educational objectives

- Know the hemodynamic changes associated with pregnancy.
- Know the normal physical findings associated with pregnancy.
- Know the indications for non-invasive testing during pregnancy.

Hemodynamic changes associated with pregnancy:

- Increased blood volume with relative increase in plasma volume (reduced hematocrit)
- Increased stroke volume with associated increase in cardiac output until 28th week, followed by an increase in heart rate to maintain high cardiac output
- Mildly increased central venous pressure
- Decreased systemic vascular resistance
- Decreased or unchanged pulmonary vascular resistance
Normal Physical Examination in Pregnancy

- Carotids brisk
- JVP may be slightly increased with normal contour
- Apical impulse displaced and hyperdynamic
- 2-3/6 early peaking systolic flow murmur at base
- S3 in younger women

Indications for Echocardiogram in Pregnancy

- Pre-existing heart disease
- Progressive dyspnea
- Systolic murmur
  - > 3/6 intensity (ie. thrill present)
  - Late peaking
  - Holosystolic murmur
- Diastolic murmur always abnormal
  - Usually AI or MS
Case 4:
The next step in your evaluation is:

1. Echocardiogram
2. Holter monitor
3. Event monitor
4. Reassurance

Case 5: 46 yo woman with palpitations and systolic murmur

Onset of intermittent palpitations that are sporadic and unassociated with exertion. Previously healthy but admits to cough and mild dyspnea during her work-outs

- PMH: Several episodes of pneumonia during childhood. Now with a bout of bronchitis every winter
- PE: BP 120/70, HR 70 with occasional extrasystole, nl JVP, Carotids nl, S1 nl, S2 widely and fixed split, 3/6 systolic ejection murmur at base, S3 at lower left sternal border.
- EKG: shown
- Echocardiogram shows dilated right ventricle, right atrium, pulmonary artery pressure of 45 mmHg.
The next most appropriate test is:

1. Helical CT scan
2. Cardiac MRI
3. Cardiac catheterization
4. Contrast echocardiogram
5. Radionuclide ventriculography
Case 5: Educational objectives

- Recognize the characteristics of murmurs associated with congenital heart disease

ASD

- Presentation:
  - 2nd most common congenital heart lesion in adults after bicuspid aortic valve
  - No classic symptom presentation
  - Recurrent pulmonary infections are common
  - May present with paradoxical embolism
- Physical examination:
  - Systolic murmur
  - Fixed split S2 is pathognomic
- EKG:
  - IRBBB and RAD
  - Ostium primum defects associated with LAD
- CXR: RV enlargement with dilated PA and pulmonary plethora
• Helical CT to exclude a pulmonary embolus.
  • PE: loud P2
  • EKG: sinus tachycardia and possibly S1, Q3, T3 pattern.

• Cardiac MRI would likely reveal the atrial septal defect, but is not necessary as surface and transesophageal echocardiography are generally diagnostic.

• Radionuclide ventriculography with 1st pass imaging can be used to quantify the magnitude of a left to right shunt, but will not delineate the structural abnormality.

• If cardiac catheterization is performed, it should be done in conjunction with percutaneous ASD closure when feasible.

Features of systolic murmurs associated with congenital heart disease:

• Aortic stenosis: loudest in 2nd RICS, preceded by systolic ejection click, S4 common, ejection quality, EKG - LVH
• Pulmonic stenosis: Loudest in the 2nd LICS increases w/ inspiration, preceded by ejection click, ejection quality EKG - RVH
• Atrial septal defect: Loudest in the 2nd LICS, wide fixed split S2, diastolic rumble over tricuspid region, R sided S3 EKG - right axis deviation, IRBBB
• Ventricular septal defect: Holosystolic heard best over lower left sternal border radiating to right, displaced apical impulse, diastolic rumble over mitral region, L sided S3, EKG - LVH
• Patent ductus arteriosus: continuous “machinery” murmur loudest in systole, displaced apical impulse, L sided S3
Coarctation of the Aorta

- Upper extremity hypertension
- Murmur over the back
- Decreased and delayed femoral pulses
- CXR - rib notching
- EKG - LVH

Eisenmenger’s Syndrome

- Can occur with ASD, VSD or PDA
- Pulmonary vascular disease with reversal of shunt
- PE: Clubbing and cyanosis on exam, murmur may be inaudible or replaced by murmur of PI or TR
- Labs: High HCT
- EKG: RVH, RAE
- Management:
  - Pregnancy contraindicated
  - Closing defect contraindicated
  - Don’t overphlebotomize
The next most appropriate test is:

1. Helical CT scan
2. Cardiac MRI
3. Cardiac catheterization
4. Contrast echocardiogram
5. Radionuclide ventriculography

Case 6: 67 yo man intubated in ER for respiratory distress

- CC: 2 wk H/O progressive dyspnea and wt gain
- PMH: Murmur for more than 20 years, prescribed Abx prophylaxis
- PE: HR = 102, BP = 80/60, JVP nl, Carotids 1+ bilat LV heave, single S2, 1/6 SEM, Summation gallop
- CXR: pulm edema, cardiomegaly
- EKG: shown
Case 6: The most likely diagnosis is:

1. Acute mitral regurgitation due to ruptured chordae tendinae
2. Critical aortic stenosis
3. Ventricular septal defect associated with myocardial infarction
4. Acute mitral regurgitation due to ruptured papillary muscle
5. Ischemic papillary muscle dysfunction
Case 6: Educational Objectives

- Recognize the acute presentation of critical aortic stenosis
- Know structural causes of pulmonary edema

Differential diagnosis of pulmonary edema

- Elevation of pulmonary capillary pressure in the presence of severe systolic or diastolic dysfunction or left sided valvular disease.
- Etiologies:
  - Ischemia
  - Structural defects
  - Systolic dysfunction
  - Diastolic dysfunction
  - Volume overload (also, remember renal artery stenosis!)
Structural Causes of Pulmonary Edema

- Aortic stenosis - usually chronic, rarely presenting acutely
- Acute aortic regurgitation - endocarditis, aortic dissection
- Acute mitral regurgitation
  - Ruptured chordae tendinae
  - Papillary muscle rupture
  - Ischemic left ventricular dysfunction
- Mitral stenosis - usually associated with pulmonary edema in presence of:
  - Rapid atrial fibrillation
  - Pregnancy
- Acquired ventricular septal defect
  - Traumatic
  - Infarct-associated

Case 6: Discussion

- The most likely diagnosis in this patient is aortic stenosis based on the presence of a single S2, the presence of LVH on EKG and the chronicity of the murmur.

- Patients with low output may have a soft systolic murmur in the presence of severe AS.

- Most likely etiology is a bicuspid aortic valve given that murmur has been recognized for over 20 years.

- Management should include emergency echocardiogram, catheterization to r/o CAD and emergent surgery.
Case 6: The most likely diagnosis is:

1. Acute mitral regurgitation due to ruptured chordae tendinae
2. Critical aortic stenosis
3. Ventricular septal defect associated with myocardial infarction
4. Acute mitral regurgitation due to ruptured papillary muscle
5. Ischemic papillary muscle dysfunction

- Acute mitral regurgitation due to ruptured chordae
  - Loud P2
  - Apical murmur (may be early, mid or holosystolic)
  - Hyperdynamic precordium

- Infarct-related VSD or papillary muscle rupture
  - Timing: within 72 hours of a STEMI infarction, more often IMI
  - EKG: STEMI MI
  - VSD: Harsh holosystolic murmur usually at LLSB, thrill
  - Papillary muscle rupture: murmur variable in timing, no thrill

- Ischemic papillary muscle dysfunction
  - EKG: Ischemic ST segment changes
Aortic Stenosis

- **Etiology**
  - Congenital (bicuspid, unicuspid)
  - Rheumatic (mixed AS/AR)
  - Calcific/Degenerative (risk factors – age, male, current smoking, HTN, high LDL and Lp(a), renal insufficiency, hypercalcemia)

AS is present in ~25% of adults over 65 years of age

Not to be confused with: HCM; Subvalvular AS; Supravalvular AS

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Calcific Bicuspid Aortic Valve

[Image: Courtesy Dr. Phil Ursell, UCSF]
Aortic Stenosis

• **Pathophysiology**
  - Pressure overload hypertrophy
  - Elevated LV systolic pressure; increased LV ejection time and ↑ LV EDP
  - Decrease compliance of the hypertrophied LV
  - ↑ myocardial O$_2$ consumption and decreased O$_2$ supply produce myocardial ischemia
  - Myocardial dysfunction

Aortic Stenosis

• **Classification of Severity**
  - **Mild** (area 1.5 cm$^2$, mean gradient <25 mmHg, jet velocity <3.0 m/sec)
  - **Moderate** (area 1.0-1.5 cm$^2$, mean gradient 25-40 mmHg, jet velocity 3.0-4.0 m/sec)
  - **Severe** (area <1.0 cm$^2$, mean gradient >40 mmHg, jet velocity >4.0 m/sec)

*NOTE: Do not rely on single number; Account for BSA; Symptoms predominate*
Aortic Stenosis

• Natural History
  – Prolonged latent period during which morbidity and mortality very low
  – Marked individual variability
  – Average rate of progression is:
    • Increase in jet velocity of 0.3 m/sec/year
    • Increase in mean pressure of 7 mmHg/year
    • ↓ AVA 0.1 cm²/year

Management of Asymptomatic Aortic Stenosis

– Risk of sudden cardiac death is low <0.5%/year
– Patients need frequent monitoring for development of symptoms and progressive disease:
  • Physical exam
  • Echo (every year for severe AS; 1-2 yrs for moderate AS and 3-5 yrs for mild AS)
  • Selective ETT in asymptomatic patients (BP response, exercise induce symptoms)
  • Antibiotic prophylaxis not recommended
  • ? Statins
  • Patients with moderate-severe AS should avoid competitive sports
Aortic Stenosis

- **Echocardiography**
  - LV wall thickness
  - LV size
  - LV function
  - Outflow tract area (pay attention to technical detail)
  - Continuity equation valve area
  - Presence of other associated valvular disease

Aortic Stenosis

- **Indication for cardiac catheterization**

  **Class I Indications:**
  - Coronary angiography before AVR
  - In symptomatic patients when non-invasive studies are inconclusive or discrepant
Aortic Stenosis

Low-Flow/Low-Gradient AS

– Patients with severe AS and low cardiac output often present with low AV pressure gradient (have increased operative mortality)

– Can be difficult to distinguish:
  • Severe end-stage AS
  
  VS.

  • Mild to moderate AS with severe contractile dysfunction

Dobutamine pharmacological stress

• Echo
• Cath

⇒ Severe AS: if increase in SV (↑ CO), ↑ gradient, FIXED AVA

⇒ "Pump issue:" no increase or ↑ gradient, ↑ CO, ↑ AVA (by >0.2 cm²)
Aortic Stenosis

Indications for AVR

Class I Indications:
- Symptoms (dyspnea, angina, syncope)
- Severe AS in patients undergoing CABG
- Severe AS in patients undergoing surgery of aorta or other valves
- Severe AS for patients with LV systolic function < 50%
Aortic Stenosis

Indications for AVR

Class IIa Indications:
- Moderate AS in patients undergoing CABG or other cardiac surgery

Class IIb Indications:
- May be considered for asymptomatic patients with severe AS and abnormal response to stress test
- May be considered for asymptomatic patients with severe AS if there is a high likelihood of rapid progression
- May be considered in patients undergoing CABG with mild AS when there is evidence that progression may be rapid (e.g. moderate to severe valve calcification)
- May be considered for asymptomatic patients with AVA < 0.6 cm² when operative mortality is ≤ 1%

Survival After AVR for AS

Connolly HM et al Circulation 1997
Aortic Stenosis

Aortic Balloon Valvuloplasty (high rate of restenosis in 6-12 months)

Class IIb Indications:
- Reasonable as a bridge to surgery in unstable patients
- Palliation in patients with co-morbidities

Aortic Stenosis

Percutaneous AV Replacement
- Experimental
- Rapid progress being made
Aortic Regurgitation

- **Etiology**
  - Valvular
  - Root

Aortic insufficiency

- **Etiology**:
  - Valvular: rheumatic, bicuspid, endocarditis
  - Aortic root disease: Marfan’s, dissection, syphilis, ankylosing spondylitis, etc
- **History**: Acute (shock) or Chronic (dyspnea, CHF)
- **PE**: Wide pulse pressure, soft S1, soft or absent A2, diastolic blowing murmur (**Acute AI**: short murmur, narrow pulse pressure)
- **Tx**:
  - Chronic: vasodilators, valve replacement if syx or asyx with LV dilatation
  - Acute: Surgery, IV vasodilators, No IABP
Aortic Valve Endocarditis

Courtesy Dr. Phil Ursell, UCSF

Aortic Regurgitation

Pathophysiology

**ACUTE**
- sudden large volume load
- ↑ LVEDP; ↑ Left atrial pressure
- ↓ stroke volume; ↑ HR; narrow pulse pressure
- Soft murmur, soft S1
- Pulmonary edema; cardiogenic shock

**CHRONIC**
- ↑ LVEDP, ↑ LV compliance
- eccentric and concentric hypertrophy
- ↑ afterload
- Prognostic value of LV EF and LV ESD
Chronic Aortic Regurgitation

Natural History of **Asymptomatic AR with Normal LV function:**

- Symptoms/LV dysfunction 4.3%/year
- Sudden death 0.2%/year
- LV dysfunction (no syx’s) 1.2%/year

Chronic Aortic Regurgitation

Natural History of **Asymptomatic AR with Depressed LV function:**

- Symptoms >25%/year (majority will need AVR in 2-3 years)
Chronic Aortic Regurgitation

Natural History of Symptomatic AR:

- No large-scale trials in the modern era as onset of angina and/or dyspnea are indications for valve surgery
- However, data from pre-surgical era suggests a poor outcome without surgery
  - Mortality >10%/year with angina
  - Mortality >20%/year with heart failure

Chronic Aortic Regurgitation

Symptomatic and LV dysfunction:

<table>
<thead>
<tr>
<th>LVEDD</th>
<th>≥70 mm</th>
<th>10% mortality/year</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>&lt;70 mm</td>
<td>12% mortality/year</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>LVESD</th>
<th>≥50 mm</th>
<th>19% mortality/year</th>
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</thead>
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<tr>
<td></td>
<td>40-49 mm</td>
<td>6% mortality/year</td>
</tr>
<tr>
<td></td>
<td>&lt; 40 mm</td>
<td>~0% mortality/year</td>
</tr>
</tbody>
</table>
Chronic Aortic Regurgitation

Diagnosis/Assessment:

- Physical examination
- Echocardiography
- EKG
- CXR
- Cardiac Cath
- CT/MRI
- Radionuclide angiography

Medical Therapy in Chronic AR

- Endocarditis prophylaxis not recommended

- Vasodilator therapy is indicated in patients with severe AR who have symptoms or LV dysfunction when surgery is not recommended because of additional cardiac or noncardiac factors (Class I)

- Vasodilator therapy is reasonable for short-term to improve hemodynamics in patients with AR, depressed LV EF and CHF before AVR (Class IIa)

- Vasodilator therapy reasonable for long-term in asymptomatic patients with severe AR who have LV dilatation but normal systolic function (Class IIb)
Indications for Surgery in Chronic Aortic Regurgitation

Class I Indication:
- Symptoms irrespective of LV function
- Asymptomatic patients with severe AR and LV dysfunction (EF ≤ 50%)
- Need for other surgery such as CABG or root surgery with severe AR

Pre-op Functional Class Predicts Outcome after AVR for Chronic AR

Klodas E et al JACC 1997
**Indications for Surgery in Chronic Aortic Regurgitation**

**Class Ila Indication:**
- AVR is reasonable for asymptomatic patients with severe AR with normal LV systolic function, but with severe LV dilatation (EDD > 75 mm or ESD > 55 mm)

**Class Ilb Indication:**
- Moderate AR in patients with moderate AR while undergoing surgery on the ascending aorta or CABG
- AVR may be considered for asymptomatic patients with severe AR and normal LV function when there is LV EDD > 70mm or ESD > 50 mm, when there is evidence of progressive LV dilatation, declining exercise tolerance, or abnormal hemodynamic responses to exercise

**Case 7: 40 yo woman w/ hx of mantle XRT for Hodgkin’s disease at age 24**

- **CC:** increasing abdominal girth and edema
- **PMH:**
  - Also received chemotherapy for Hodgkin’s Disease: adriamycin, bleomycin, vinblastine, dacarbazine.
  - MVA 5 yrs earlier: required Tx 2u PRBC’s
- **PE:** HR = 98 reg, BP = 100/70
  - Dullness to percussion and ↓ breath sounds at R base
  - Jugular venous pressure elevated
  - PMI not palpable, cardiac border displaced
  - Distant heart sounds, no Murmurs, mid diastolic sound
  - Peripheral edema
- **CXR:** normal cardiac silhouette, right pleural effusion
Case 7: The most likely diagnosis is:

1. Effusive pericarditis
2. Constrictive pericarditis
3. Cirrhosis associated with hepatitis C
4. Dilated cardiomyopathy
5. Restrictive cardiomyopathy

Cardiac complications of radiation therapy:

- Risk of heart disease approximately 4% at an average f/u of 10 yrs in pts with mantle radiation for HD
- Spectrum of heart disease
  - Chronic pericardial effusion or constriction
  - Coronary artery disease
  - Valvular disease
  - Restrictive cardiomyopathy
  - Conduction defects
Case 7: Discussion

- **Features c/w constrictive pericarditis**
  - Insidious onset of ascites and edema.
  - Physical exam:
    - Pulsus paradoxus uncommon
    - ↑ JVP with prominent y descent
    - Positive Kussmaul’s sign
    - Distant heart sounds
    - Early diastolic sound c/w pericardial knock.
- **CXR - pericardial Ca++ in 25%**
- **Echo evidence of hemodynamic constriction**
- **MRI or CT to demonstrate pericardial thickening**
- **Cath recommended to exclude restrictive cardiomyopathy and coronary artery involvement**

- **Restrictive cardiomyopathy** often coexists with constrictive pericarditis.
- **Effusive pericarditis** is more likely to be an early complication of radiation and is likely to be associated with cardiomegaly on chest x-ray.
- **Liver disease** : JVP usually normal
- **DCM**: can complicate Adriamycin treatment, expect cardiomegaly, ascites uncommon.
## Constriction vs. Tamponade vs. Restriction

<table>
<thead>
<tr>
<th></th>
<th>Tamponade</th>
<th>Constriction</th>
<th>Restrictive Cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pulsus Paradoxus</strong></td>
<td>90%</td>
<td>10%</td>
<td>Absent</td>
</tr>
<tr>
<td><strong>JVP</strong></td>
<td>↑↑, Loss of y descent</td>
<td>↑↑, Steep y descent</td>
<td>↑↑, Steep y descent</td>
</tr>
<tr>
<td><strong>Kussmaul’s</strong></td>
<td>10%</td>
<td>90%</td>
<td>Common</td>
</tr>
<tr>
<td><strong>CXR</strong></td>
<td>↑↑ CT ratio “waterbottle”</td>
<td>Mild ↑↑ CT ratio Ca++ 25%</td>
<td>Mild ↑↑ CT ratio</td>
</tr>
<tr>
<td><strong>Echo</strong></td>
<td>Effusion, RA/RV collapse, resp var MV and TV flow, dil IVC</td>
<td>Adhesion, small ventricles, resp var MV and TV flow, dil IVC</td>
<td>Thick walls, preserved systolic fxn, abnl diastolic fxn</td>
</tr>
<tr>
<td><strong>CT/MRI</strong></td>
<td>N/A</td>
<td>Pericardial thickening</td>
<td>Normal pericardial thickness</td>
</tr>
</tbody>
</table>

## Case 7: The most likely diagnosis is:

1. **Effusive pericarditis**
2. **Constrictive pericarditis**
3. **Cirrhosis associated with hepatitis C**
4. **Dilated cardiomyopathy**
5. **Restrictive cardiomyopathy**
Case 8: 58 yo Mexican woman with progressive dyspnea presents to ER

- CC: Increasing exertional shortness of breath for several months. 2 pillow orthopnea, occasional palpitation.
- PMH: 2 uncomplicated pregnancies at age 18, 22
- Medications: none
- PE: BP = 95/65, HR = 110 irreg irreg, afebrile
  - JVP 10 cm, carotids - 1+ bilaterally
  - Lungs: crackles 1/2 way up bilaterally
  - Cor: sustained right parasternal impulse, variable and loud S1, physiologically split S2 with accentuated P2, Grade 2/6 diastolic murmur, Grade 3/6 holosystolic murmur at R lower sternal border

Case 8, continued

- EKG - atrial fibrillation, QRS axis + 90, RVH
- Echocardiogram:
  - Mitral stenosis with a mean gradient of 12 mmHg, MVA = 0.8 cm²
  - Moderate to severe TR
  - PA systolic pressure estimated at 70 mmHg.
  - Dilated RV, normal LV
  - Severe biatrial enlargement
Case 8: The patient is admitted. The next step in her management should be:

1. Furosemide 20 mg IV and digoxin 0.5 mg IV
2. Furosemide 20 mg IV and metoprolol 5 mg IV
3. Urgent cardioversion
4. Urgent percutaneous balloon valvuloplasty
5. Helical CT scan to rule out pulmonary embolism

Mitral stenosis:

- **Etiology:**
  - 75% Post-inflammatory (ie. rheumatic)
    - Only 50-70% have clear h/o RF
  - Other etiologies < 10%:
    - Mitral annular calcification
    - SLE or RA
    - Carcinoid
    - Methysergide ingestion
    - Congenital

- **Pathology - important for treatment**
  - Commisural fusion
  - Thickening, fibrosis and calcification of the leaflets
  - Thickening, fusion and foreshortening of the chordae tendinae
• Symptoms: Exertional dyspnea, hemoptysis, edema
• Physical examination:
  • RV lift
  • Loud S1, P2 increased in presence of PHT
  • Opening snap in early diastole
  • Diastolic rumble at apex
  • Systolic murmur in presence of associated MR
• CXR: LAE, RV enlargement
• EKG: Atrial fibrillation common, LAE, vertical axis or RAD
• Echo: Diagnostic

Mitral Stenosis:
Indications for treatment

• Symptoms of dyspnea, exercise intolerance
• Hemoptysis
• Pulmonary hypertension
• Evidence of right heart failure
• Atrial fibrillation ± embolism

MVA < 1.5 cm²
Mitral Stenosis Severity

<table>
<thead>
<tr>
<th></th>
<th>MILD</th>
<th>MODERATE</th>
<th>SEVERE</th>
</tr>
</thead>
<tbody>
<tr>
<td>MEAN GRADIENT (mmHg)</td>
<td>5 - 8</td>
<td>8 - 12</td>
<td>&gt; 12</td>
</tr>
<tr>
<td>MITRAL VALVE AREA (cm²)</td>
<td>&gt; 1.5</td>
<td>1.0 - 1.5</td>
<td>&lt; 1.0</td>
</tr>
</tbody>
</table>

Mitral Stenosis: Medical Treatment

- Diuretic + Na restriction
- Beta-blocker
- Warfarin if AF present or H/O embolus, Heparin should be considered acutely
- Secondary prevention for rheumatic fever
  - Up to age 25 or 10 yrs after last episode
  - Older patients with continued exposure to Strep A pharyngitis
  - Pen VK 250 bid or Benzathine PCN G 1.2 m units IM monthly
Mitral stenosis: Treatment

- Percutaneous Balloon Valvuloplasty
  - Anatomy suitable
  - Thrombus excluded
  - No more than mild mitral regurgitation
- Surgical
  - Open commissurotomy vs MVR

Case 8: The patient is admitted. The next step in her management should be:

1. Furosemide 20 mg IV and digoxin 0.5 mg IV
2. Furosemide 20 mg IV and metoprolol 5 mg IV
3. Urgent cardioversion
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5. Helical CT scan to rule out pulmonary embolism
Mitral regurgitation

- **Etiology:**
  - Primary valvular: myxomatous, rheumatic, endocarditis, MVP
  - Functional: cardiomyopathy, ischemia

- **Presentation**
  - Acute: pulmonary edema
  - Chronic: dyspnea, CHF

- **PE:** soft S1, holosystolic murmur, S3, displaced apical impulse (MVP - mid-systolic click, late systolic murmur), loud P2 if pulmonary HTN

- **CXR, EKG - LVH**
- **Echo diagnostic**
- **Management:**
  - ACE for LV dysfunction, HTN
  - Surgery for syx and LV dilatation with LVESD > 45 cm
  - Repair preferred over replacement

---

Mitral Regurgitation

- Common valvular disorder that can arise from abnormalities of any part of the mitral valve apparatus (leaflets; annulus; chordae and papillary muscles)
- More frequently diagnosed due to the widespread use of echocardiography
- Mostly mild in severity
- Onset of symptoms often insidious, unless acute MR
Mitral Regurgitation: Etiologies

**Primary Valvular Disease**
- Myxomatous Degeneration
  - Prolapse
  - Ruptured Chordae Tendinae
- Post-inflammatory “Rheumatic”
- Endocarditis
- Papillary muscle rupture
- Congenital “cleft”
- Trauma
- Ischemia
- Tumor

**Functional or Secondary**
- Cardiomyopathy - dilated or hypertrophic
- Ischemic
- HCM

Mitral Valve Endocarditis

Courtesy Dr. Phil Ursell, UCSF
Physiology of Mitral Regurgitation Depends on Acuity vs. Chronicity

- Left ventricular volume overload
- LA enlargement
- Eccentric hypertrophy
- LVEF normal to hyperdynamic
- Pulmonary venous hypertension

Clinical Manifestations of Chronic Mitral Regurgitation

- Symptoms related to the severity of regurgitation, rate of progression, pulmonary artery pressure, and associated cardiac disease

- Most cases, patients can remain asymptomatic for many years

- When symptoms occur, they reflect a decrease in forward and effective cardiac output – weakness, fatigue, exercise intolerance, shortness of breath, congestive heart failure, hemoptysis, right sided heart failure, endocarditis, thromboembolism
Issues for Timing of MV Surgery in Patients with Mitral Regurgitation

- Patients remain asymptomatic until late in the course of the disease
- Afterload reduction has no proven role in delaying disease progression
- Ejection fraction tends to overestimate LV function in patients with MR
- Prosthetic mitral valves are never ideal
- MV repair and chordal sparing operations have changed our practice to recommend earlier surgery
- Percutaneous techniques are being developed

Guidelines for Surgical Timing

**Class I**

- Acute symptomatic MR in which repair is likely
- NYHA II, III, IV symptoms with normal LV function
- Asymptomatic patients with chronic severe MR and mild or moderate LV dysfunction
  - EF 30-60% and/or LV ES dimension ≥ 40 mm
Guidelines for Surgical Timing

Class IIa

• Asymptomatic patients with preserved LV function and new onset Atrial fibrillation
• Asymptomatic patients with preserved LV function and Pulmonary HTN (PASP >50 mmHg at rest; >60 mmHg with exercise)
• Asymptomatic patients with LVEF <60% and LV end systolic Ds > 45 mm in whom likelihood of successful repair without residual MR is greater than 90%
• Patients with severe LV dysfunction in whom repair or chordal sparing is highly likely

Repair vs. Replacement of the Mitral Valve?

• The choice of procedure depends, at least in part, upon the cause of the MR, the anatomy of the mitral valve, and the degree of left ventricular dysfunction

• Mitral repair procedures are increasingly being used with potential benefits:
  - It preserves the functional components of the native valve and is associated with better postoperative ventricular function
  - It avoids the complications of prosthetic heart valve
  - Outcomes are excellent (in-hospital mortality ~1% and the survival rate after 8 years ~ 90%).
Repair vs. Replacement: Long-term results

Mohty D et al. Circ 2001

Percutaneous approaches to MV repair

- Annuloplasty device via coronary sinus
- Percutaneous edge-to-edge repair
  - Suture
  - Clip
Percutaneous Annuloplasty

Edge-to-Edge Surgical Repair
Catheter-based MV Repair
Endovascular Approach

Off-pump Edge-to-Edge Mitral Valve Technique
Using a Mechanical Clip in a Chronic Model

Clip repair in porcine heart (6 months post repair)

Fann JI; St. Goar FG; Komtebedde J; Oz MC; Block PC; Foster E; Feldman T;
Case 9: 42 yo woman with metastatic breast cancer presents with syncope

- S/P bone marrow transplant 1 year ago with recently discovered new hepatic metastases presents with dyspnea and worsening fatigue for 1 week with poor oral intake, syncope on rising from bed on the morning of admission.
- PMH:
  - Left sided lumpectomy with XRT 2 yrs PTA
  - Adjuvant chemoRx with adriamycin, cyclophosphamide, 5-FU
- PE: HR 97, BP 95/70, RR 30, afebrile
  - JVP 9 cm, Lungs clear, Cardiac distant but audible heart sounds, no edema
- CXR: large heart, clear lung fields
Case 9: The most likely diagnosis is:

1. Ventricular tachycardia in the setting of doxorubicin-induced cardiac toxicity
2. Cardiac tamponade due to neoplastic pericardial involvement
3. Orthostatic hypotension due to dehydration
4. Acute inferior myocardial infarction with right ventricular extension
5. Pulmonary embolism

EKG: Pulsus Alternans
Discussion:

- Significant risk factors for cardiomyopathy include associated XRT and bone marrow transplant.
- Labs suggest dehydration which may have contributed to her syncope, but ↑ JVP suggests that this is not the sole mechanism for syncope.
- CAD may be a late manifestation of mediastinal XRT. IMI with RV involvement can mimic tamponade. However, the EKG is not consistent with IMI.
- Pulmonary embolism should be considered in all cancer patients as they may be hypercoagulable. Again the clinical presentation of syncope and the elevated JVP are consistent with pulmonary embolism.

Neoplastic Pericardial Disease

- Most common secondary involvement in lung, breast CA and lymphomas
- Presentation may be subacute with dyspnea, fatigue, chest fullness or edema, syncope may occur
- PE: pulsus paradoxus ± hypotension, JVD with loss of y descent, distant heart sounds
- EKG:
  - Sinus tachycardia
  - electrical alternans
  - Low voltage (60% sensitive, low specificity)
Cardiotoxicity with Chemo-Rx agents I

• Most common offending agents:
  - Doxorubicin, epirubicin, daunorubicine, idarubicine, mitoxantrone

• Clinical manifestations depend on timing
  - Acute - arrhythmias, myopericarditis usually self-limited
  - Early - congestive heart failure
  - Late - congestive heart failure

Cardiotoxicity with Chemo-Rx agents II

• Risk factors:
  - Cumulative dose > 550 mg/m2 of doxorubicin
  - XRT
  - Age extremes
  - Other cardiovascular disease
  - Serial monitoring - functional studies, biopsies
Case 9: The most likely diagnosis is:

1. Ventricular tachycardia in the setting of doxorubicin-induced cardiac toxicity
2. Cardiac tamponade due to neoplastic pericardial involvement
3. Orthostatic hypotension due to dehydration
4. Acute inferior myocardial infarction with right ventricular extension
5. Pulmonary embolism

Case 10: 72 year old woman with myalgias, fever and chills

- 6 months prior to admission, she had porcine AVR for aortic stenosis complicated by atrial fibrillation on day 5 post-op. Initially recovered well but had progressive fatigue and myalgias for 2 weeks and began to notice nightly fever and diaphoresis 3 days PTA. In addition, she complained of chest pain on the day of admission
- PMH: hypercholesterolemia - dx’d during hospitalization
- Meds:
  - Simvastatin 40 mg qd
  - ASA 325 mg qd
  - Amiodarone 200 mg qd
  - Digoxin 0.125 mg qd
- PE: BP 135/55, HR 92 regular, T - 38°C, HEENT negative, Lungs - bibasilar rales, Cor - apically displaced PMI, S1 nl, S2 diminished, 2/6 systolic ejection quality murmur, 1/6 early diastolic murmur along L sternal border.
Case 10: Three sets of blood cultures are drawn, the next test should be:

1. CPK level
2. Troponin I
3. Transesophageal echocardiogram
4. Helical CT of thoracic aorta
5. Digoxin level
Discussion:

- Probable prosthetic valve endocarditis:
  - TEE indicated because of higher sensitivity for vegetations compared to TTE
  - Perivalvular abscess suspected because of 1st degree AV block
  - Chest pain possible due to associated pericarditis
- CPK would suggest statin-induced myositis
  - Unlikely in the presence of fever and chills
- Troponin not indicated in absence of evidence for myocardial ischemia
- Helical CT of the aorta not indicated
  - Chest pain not consistent with dissection.
- Digoxin may cause 1st degree AV block but the clinical scenario is more worrisome for perivalvular abscess.

Prosthetic Valve Endocarditis

- Classified according to timing of infection
  - Early (< 2 months after implantation)
  - Mid (2 - 12 months)
  - Late (> 12 months)
PVE: Mechanism and microbiology

- Early infections more virulent
  - Greater involvement of perivalvular tissue with abscess formation, fistulae, etc (lack of endothelialization)
  - Mechanical valves = bioprosthetic valves
  - Coagulase negative staph and staph aureus account for > 50%
- Late infections similar to Native valve endocarditis
  - Abscess and perivalvular extension less common except in S aureus
  - Bioprosthetic valves > mechanical valves because of degenerative changes associated with bioprosthesis
  - Organisms similar to Native valve endocarditis with Strep and S aureus accounting for > 50%

Indications for surgery in PV endocarditis

- Class I
  - Early (≤ 2 mo)
  - CHF due to prosthetic valve dysfunction
  - Fungal endocarditis
  - Staph endocarditis not responding to antibiotics
  - Evidence of perivalvular leak, annular or aortic abscess, sinus or aortic true of false aneurysm, fistula formation or new-onset conduction disturbance
  - Gram negative or other organisms with poor response to antibiotics
- Class Ila
  - Persistent bacteremia on Rx
  - Recurrent embolism on Rx
  - Vegetation on or near the prosthesis
Other Prosthetic Valve Complications:

- Valve failure: bioprosthetic > mechanical, usually after 10 yrs
- Thrombosis
  - Inadequate anticoagulation
  - Acute or subacute CHF
  - Valve clicks muffled
  - When small - heparin, when large - thrombolysis or replacement
- Emboli
- Hemolysis - usually associated with perivalvular leak