Valve Disease
Board Review Questions

Dennis A. Tighe, MD, FASE
University of Massachusetts Medical School
Worcester, MA
Case 1
History

• A 61 year-old man
  – Presents to hospital with worsening shortness of breath, back pain, and a 20-pound weight loss over the past 6-months.
  – Two weeks prior to presentation he developed orthopnea.
  – As an outpatient, an oral antibiotic was prescribed for presumed pneumonia.
  – Transferred from an OSH for further care.
History/Data

- **PMH**
  - HTN
  - Dyslipidemia
  - CAD
  - Type B aortic dissection 1996

- **SH**
  - Manual laborer
  - Non-smoker
  - No EtOH
  - No illicit drug use

- **Exam**
  - Labored breathing (50% FM); HR 80/min, regular
  - JVD
  - Bilateral rales
  - HSM apex, diastolic decresendo murmur LLSB
  - LE edema

- **Labs**
  - WBC 14, Hgb 9.2
  - SR 1° AVD, IRBBB, LAE
  - Pulm edema, b/l effusions
Based on the history and TEE images, which one of these conditions most likely explains the mitral valve findings?

- A. Myxomatous degeneration
- B. Congenital diverticulum
- C. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa
- D. Endocarditis involving the aortic valve
- E. Blood cyst of the mitral valve
Sub-aortic Complications of IE

<table>
<thead>
<tr>
<th></th>
<th>Mitral valve aneurysm</th>
<th>Mitral valve diverticulum</th>
<th>Mitral valve dissection</th>
<th>Mitral valve prolapse</th>
<th>Cardiac tumors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appearance</td>
<td>Saccular with distinct mouth and neck</td>
<td>Saccular with distinct mouth and neck</td>
<td>Saccular. Absence of distinct neck and mouth</td>
<td>Myxomatous thickening with redundant tissue.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Absence of distinct neck and mouth</td>
<td>Typically solid. Rare cystic changes. Absence of distinct neck and mouth</td>
</tr>
<tr>
<td>Location and shape</td>
<td>Mouth facing left ventricle, aneurysmal</td>
<td>Mouth facing left atrium with diverticulum</td>
<td>Double layer pouch. May resemble flail leaflet</td>
<td>Bowing of mitral leaflet may approximate a</td>
<td></td>
</tr>
<tr>
<td></td>
<td>sac bulging into left atrium</td>
<td>bulging into left ventricle</td>
<td></td>
<td>semicircle</td>
<td>Shape variable. Myxomas rarely found attached to valves</td>
</tr>
<tr>
<td>Changes in appearance</td>
<td>Systolic expansion into left atrium, ↓</td>
<td>Systolic expansion</td>
<td>Absence of systolic expansion</td>
<td>Leaflet tip bulging into left atrium with</td>
<td></td>
</tr>
<tr>
<td>during the cardiac cycle</td>
<td>size or collapse in diastole</td>
<td></td>
<td></td>
<td>systole</td>
<td>Absence of significant changes</td>
</tr>
<tr>
<td>Doppler findings</td>
<td>Color flow swirling in sac. Perforation</td>
<td>Color flow swirling in sac. Perforation</td>
<td>Color flow swirling (?). May be associated</td>
<td>Posteriorly directed mitral regurgitant jet</td>
<td></td>
</tr>
<tr>
<td></td>
<td>of sac. Perforation of sac may mimic</td>
<td>not reported</td>
<td>with mitral regurgitation</td>
<td>may be demonstrated</td>
<td>Absence of color flow swirling in cases</td>
</tr>
<tr>
<td></td>
<td>mitral regurgitation</td>
<td></td>
<td></td>
<td>of cystic changes</td>
<td>of cystic changes</td>
</tr>
</tbody>
</table>
Case 2

• An 84-year old woman with Stage IV chronic kidney disease and systemic hypertension presents to an outside hospital with worsening shortness of breath.
  – Physical examination and chest radiography were consistent with pulmonary edema
    • Diuretics were given
  – Transthoracic echocardiography was performed
Which of the following entities constitutes the most likely etiology for the finding shown?

• A. Left atrial myxoma
• B. Intracavitary thrombus
• C. Infective endocarditis
• D. Caseous calcification
• E. Papillary fibroelastoma
Caseous Calcification of the Mitral Annulus

- Relatively rare
  - Estimated prevalence of 0.07%
- Annular-based mass with echoluscencies
  - Putty-like admixture of fatty acids, cholesterol, and calcium
    - “Toothpaste” tumor
  - Rounded
  - Smooth borders
- Posterior location
- Associated conditions
  - Elderly
  - HTN
  - Women
- Natural history appears benign
  - Some cases may regress spontaneously
- Differential diagnosis
  - Abscess
  - Tumors
  - Thrombus
Case 3
A 54 year-old woman with hypothyroidism presents with worsening of shortness of breath.

- Systolic and diastolic murmurs are auscultated
- Transthoracic echocardiography is requested for further evaluation
Q1. Echocardiography confirms the presence of aortic stenosis (orifice area 0.6 cm$^2$) and identifies the presence of moderate aortic regurgitation. Mitral valve thickening is also described. The most likely etiology accounting for the observed valvular abnormalities is:

- A. Age-related degenerative valve disease
- B. Rheumatic heart disease
- C. Annular calcific disease
- D. Carcinoid heart disease
- E. Radiation-associated valve disease
Q2. Which of the following conditions would be an expected complication resulting from the disease process causing the observed left-sided valvular abnormalities?

- A. Flushing
- B. Constrictive pericarditis
- C. Coronary artery spasm
- D. Hypertrophic cardiomyopathy
- E. Cardioembolic stroke
Radiation-Associated Valve Disease

- Frequent complication
- Regurgitant lesions > Stenotic lesions
  - Left sided > right sided
- Risk greater with $\geq 30$ Gy
- Women > men
- Suggestive echocardiographic appearance
  - Calcification and thickening of aortic-mitral curtain
  - Anterior changes more profound than posterior (vs MAC)
  - No leaflet doming/commissural involvement (vs RHD)
  - Aortic root calcification increases the likelihood
- Progressive
- Periodic screening required
Radiation Therapy

- Cardiovascular complications
  - Coronary artery disease
  - Cardiomyopathy
    - Restrictive or dilated
  - Pericardial effusion
  - Constrictive pericarditis
  - Conduction system/arrhythmias
  - Valvular heart disease
  - Carotid artery disease
Case 4
Which of the following values best estimates the mitral orifice area?

- A. 0.40 cm$^2$
- B. 0.75 cm$^2$
- C. 1.0 cm$^2$
- D. 1.4 cm$^2$
- E. 2.6 cm$^2$
Choice Explanations

• D. 1.4 cm$^2$.

• This continuous wave spectral profile of the mitral valve shows increased transvalvular velocities and a prolonged deceleration time (measured).
  – Given the known deceleration time, the relationship between deceleration time (DT) and mitral pressure half-time (PHT) is:
    \[
    \text{PHT (in msec)} = 0.29 \times \text{DT}.
    \]
  – Once the PHT is known, the Hatle formula (MVA (in cm$^2$) = $220$/PHT) can be used to estimate the mitral orifice area.
    • In this case, the PHT $= 163$ msec.

• Alternatively, the formula MVA $= 759$/DT can be utilized.
Case 5
Based on this M-mode tracing, which of the following findings is *unlikely* to be present?

- A. Restrictive mitral inflow pattern
- B. Soft S1
- C. Diastolic mitral regurgitation
- D. Premature closure of the aortic valve
- E. Brief diastolic murmur
Choice Explanations

- D. Premature closure of the aortic valve. *This is the correct answer.*
  - This M-mode tracing displays premature closure of the mitral valve along with high frequency diastolic fluttering of the anterior mitral leaflet (and the interventricular septum). This constellation of findings occurs when acute, severe aortic regurgitation is present.
  - This answer is *false* because the aortic valve is incompetent. With the rapid rise in LV diastolic pressure characteristic of this lesion, premature opening of the aortic valve may be observed.

- A. Restrictive mitral inflow pattern. This answer is true due to the rapid increase in LV diastolic pressure characteristic of acute severe AR.

- B. Soft S1. This answer is true because the rapidly rising LV diastolic pressure leads to premature closure of the mitral valve.

- C. Diastolic mitral regurgitation. This answer is true. Rapid increases in LV diastolic pressure can lead to transient reversal of the LA-LV pressure gradient in diastole and the occurrence of (low velocity) diastolic mitral regurgitation.

- E. Brief diastolic murmur. This answer is true. The regurgitant murmur is brief in duration because the aortic diastolic pressure rapidly equilibrates with that of the LV.
Case 6
Two Patients with Mitral Regurgitation

A.

B.
When comparing patients with MR depicted in panels A and B, which of the following statements is true?

A. The ERO area by PISA is consistently smaller among patients depicted in panel A versus B.
B. The peak mitral inflow velocity is consistently lower among patients in panel B versus A.
C. Clinical outcomes are often better for patients depicted in panel A versus B.
D. Color jet area is often smaller among patients depicted in panel A compared to B.
Case 7
History

- A 54 year-old man presents with complaints of exertional dyspnea and abdominal bloating. He denies chest pain, PND/orthopnea, weight gain or the presence of edema
  - Physical examination reveals distended neck veins, a soft early systolic murmur and an early-diastolic rumble audible at the left parasternal border
  - Echocardiography is requested to evaluate cardiac function
Based on the clinical presentation and imaging findings which of the following statements is correct?

• A. Prominent diastolic flow reversals in the hepatic vein spectral profile are expected
• B. Pulmonary artery systolic pressure can be estimated accurately
• C. Mild-to-moderate tricuspid regurgitation is present
• D. Spatial extent of the color-flow disturbance underestimates disease severity
• E. Tricuspid stenosis is the predominant valvular abnormality
<table>
<thead>
<tr>
<th>Parameter</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid valve</td>
<td>Usually normal</td>
<td>Normal or abnormal</td>
<td>Abnormal/Flail leaflet/Poor coaptation</td>
</tr>
<tr>
<td>RV/RA/IVC size</td>
<td>Normal*</td>
<td>Normal or dilated</td>
<td>Usually dilated**</td>
</tr>
<tr>
<td>Jet area-central jets (cm²)§</td>
<td>&lt; 5</td>
<td>5-10</td>
<td>&gt; 10</td>
</tr>
<tr>
<td>VC width (cm)♭</td>
<td>Not defined</td>
<td>Not defined, but &lt; 0.7</td>
<td>&gt; 0.7</td>
</tr>
<tr>
<td>PISA radius (cm)♭</td>
<td>≤ 0.5</td>
<td>0.6-0.9</td>
<td>&gt; 0.9</td>
</tr>
<tr>
<td>Jet density and contour–CW</td>
<td>Soft and parabolic</td>
<td>Dense, variable contour</td>
<td>Dense, triangular with early peaking</td>
</tr>
<tr>
<td>Hepatic vein flow†</td>
<td>Systolic dominance</td>
<td>Systolic blunting</td>
<td>Systolic reversal</td>
</tr>
</tbody>
</table>