Board Review
Aortic Valve, HCM, Systemic Disease

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Case 1
History

- A 54 year-old woman with hypothyroidism presents to her PCP 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 cm²) and identifies the presence of moderate aortic regurgitation. Mitral valve thickening is also observed. Which of the following constitutes the most likely etiology for the valvular abnormalities:

- 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 these 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 ≥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 2
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 3

History

• A 56-year old woman with shortness of breath and the finding of right heart volume overload on outside echocardiography due an ostium secundum ASD is referred for percutaneous closure. RHC revealed MAP 29 mm Hg and PVR <2 WU. Non-obstructive CAD was found.

• TEE performed at the time of the procedure confirms the presence of the ASD and documents adequate surrounding rims and appropriate PV drainage. However an incidental finding is made.
Q1. Based on this finding which of the following courses of action should you recommend?

- A. Inform your interventional colleague to cease the procedure immediately and discuss the finding with the patient and her family.
- B. Obtain an urgent CT surgery consult.
- C. Provide additional antibiotic coverage for oral cavity organisms given a high-risk of infective endocarditis.
- D. Continue with the planned procedure and discuss the incidental finding and its implications with the referring cardiologist.
Q2. Which of the following statements concerning this valvular abnormality is correct?

- A. Aortic stenosis is the predominant hemodynamic abnormality.
- B. Aortic regurgitation is the predominant hemodynamic abnormality.
- C. Long-term survival with this condition is poor.
- D. Aortic dilatation is not commonly found in association with this lesion.
- E. Aortic dissection is strongly associated with this valvular abnormality.
History

• Echo number 25 of a long day.
• A 22 year-old man previously cared for in pediatric cardiology clinics is referred by his new cardiologist for echocardiography to evaluate mitral regurgitation and LV function.
• He has seen multiple subspecialists over the years. The patient has ESRD and was recently initiated on HD. He has poorly-controlled HTN and his CBC/diff is distinctly abnormal.
LAVI = 39 mL/m²
GLS = -12%
RV-RA grad
Pulm vein
GLS = -12%
LAVI = 39 mL/m²
Q1. Based on the available data, how would you best characterize his left heart function?

- A. Normal LVEF, elevated mean LAP, elevated LVeDP
- B. Normal LVEF, elevated mean LAP, normal LVeDP
- C. Normal systolic function with normal filling pressures.
- D. Normal systolic function with elevated filling pressures.
Q2. Which of the following conditions is the most likely etiology for the observed echo findings?

- A. Amyloidosis
- B. Apical hypertrophic cardiomyopathy
- C. Hypertensive heart disease
- D. Hypereosinophilic syndrome
- E. Rheumatic heart disease

Lab Studies

- BMP
  - Na 139; K 3.7; Cl 100; Co2 29; BUN 15; Cr 6.4; eGFR 11.
- CBC/diff
  - WBC 12.7; Hgb 8.5; Hct 25.7.
  - PMN 18%; Lymph 13%; Eos 64%; Baso 2%.
Hypereosinophilic Syndrome

Table 3  Stages of cardiac pathology in HES

<table>
<thead>
<tr>
<th>Stages of cardiac involvement in HES</th>
<th>Characterisation of stage</th>
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<tbody>
<tr>
<td>Acute necrosis</td>
<td>Eosinophilic myocarditis with eosinophilic and lymphocytic infiltration. Myocardial necrosis and apoptosis with rare microembolic phenomena. Typically no other cardiac symptoms. Rarely can exhibit a fulminant course.</td>
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<tr>
<td>Thrombotic stage</td>
<td>Thrombus along damaged endocardium. Thrombi within apices of one of both ventricles, and can encroach on base of the heart into the subvalvular regions. Embolic phenomena can occur.</td>
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<tr>
<td>Fibrotic stage</td>
<td>Thrombi are replaced by fibrosis. Scarring occurs. Restrictive cardiomyopathy with signs and symptoms of left or right sided heart failure. Fibrosis that occurs at the base of the heart can lead to valvular regurgitation.</td>
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HES, hypereosinophilic syndrome.


As recommended by the 2014 AHA/ACC Valvular Heart Disease Guideline, which of the following statements regarding follow-up of prosthetic heart valves by echocardiography is true?

• A. Annual TTE is reasonable starting at 5 years following mechanical valve replacement.
• B. An initial TEE should be performed routinely to assess valve hemodynamics within 2 months of implantation.
• C. Change in clinical status should prompt early echocardiography.
• D. Annual TTE is reasonable starting at 5 years following bioprosthetic valve replacement.

Which of the following statements regarding the obstructed/thrombosed prosthetic heart valve is correct?

• 1. A PHT > 130 msec is the single best indicator of prosthetic mitral obstruction.
• 2. Taking into account heart rate is not necessary when assessing trans-mitral gradients.
• 3. Pannus in-growth is more common in the aortic position than with mitral PHVs.
• 4. A peak velocity ≥2.5 m/sec suggests significant aortic PHV stenosis.
• 5. Randomized, controlled trials have demonstrated that bolus infusion of rt-PA is the fibrinolytic regimen of choice.
Which of the following statements concerning prosthetic heart valve regurgitation is correct?

• A. Pseudo-regurgitation is an issue most often encountered during performance of TEE.
• B. Any degree of regurgitation indicates dysfunction of a mechanical valve.
• C. Structural valve deterioration is an uncommon cause of pathological regurgitation.
• D. Mitral bioprostheses are less prone to suffer structural valve deterioration than are aortic bioprostheses.
• E. Annular dehiscence most often is a consequence of infective endocarditis.