Challenging Cases in Cardiomyopathy and Heart Failure

Nowell Fine, MD, FRCPC, SM, FASE, FACC, FHFA
Director – Echocardiography Laboratory
Libin Cardiovascular Institute of Alberta
University of Calgary, Canada

Disclosures

• No relevant disclosures or conflicts
Case 1

• 71 yo M, known CAD with PCI to RCA 6 yrs prior due to episode of unstable angina

• Referred for echo after episode of heart failure following hip replacement surgery

• Systolic murmur noted on physical exam

• Echocardiogram performed
E vel 0.9 m/s, E/A 1.1  
Septal e’ 6 cm/s, E/e’ 15  
Lateral e’ 7 cm/s, E/e’ 13

- Average E/e’ 14
- TR velocity not measurable
- LA vol index 31 mL/m²

Nagueh et al. JASE 2016;29:314

In patients with normal LV EF

- Average E/e’ > 14
- Septal e’ velocity < 7 cm/s or lateral e’ velocity < 10 cm/s
- TR velocity > 2.8 ms
- LA vol index > 31 mL/m²

- ≤ 50% positive
- > 50% positive
- Normal diastolic function
- Indeterminate
- Diastolic Dysfunction

- ¼ criteria
- <50% positive

Apex

Mid cavity

LVOT - PW

LVOT - CW
Report Summary

• Technically difficult study

• Normal LV size and EF
  • Increased septal wall thickness
  • Dynamic apical function with small intra-cavitary gradient

• SAM of the MV, with no significant MR or LVOT obstruction / gradient detected

• Consider cardiac MRI for evaluation of possible HCM

Cardiac MRI

• Asymmetric basal septal hypertrophy (max thickness 17 mm)
• SAM of the mitral valve
• Mid-wall patchy LGE consistent with fibrosis
• Overall findings consistent with septal variant HCM
In Months Following

• Developed worsening exertional dyspnea and angina

• Coronary angiography
  • Severe 3VD

• Left heart cath
  • LVEDP 25 mmHg
  • 50 mmHg gradient between apical and basal LV cavity
  • LVOTO gradient induced with nitrates of 20 mmHg
  • Brockenhrough phenomenon noted

• Referred for CABG +/- myectomy

http://www.clevelandclinicmeded.com

Intra-Operative TEE

![Esmolol – HR 50 BPM](image)

![Isoproterenol – HR 70 BPM](image)

![Low Blood Pressure](image)

![Increased Blood Pressure](image)
Case 1

- CABG x3, no myectomy
- Put on high dose β-blocker, did not tolerate ACE inhibitor
- In months following, dyspnea worsened
- Repeat echo performed

- Resolution of SAM and intra-cavitary gradient
- Otherwise no significant change from previous

- GLS significantly reduced -8%
  - Basal-apical strain gradient
  - Suggestive of cardiac amyloidosis
- RV EMBx confirmed cardiac amyloid infiltration
- Mass spectrometry (Mayo Clinic)
  - Wild-type transthyretin amyloidosis (SSA)
Case Points

• Cardiac amyloidosis is often an under-recognized Dx
  • Especially wild type transthyretin CA

• Classic echo findings may be subtle or absent, and nonspecific

• Strain imaging is very helpful for improving diagnostic accuracy

Echo Evaluation of Cardiac Amyloidosis

• Increased LV wall thickness
  • wt ATTR > mutant ATTR > AL

• Early preserved LVEF, late reduced LVEF

• Diastolic dysfunction
  • +/- Restrictive physiology

• Thickened cardiac valves

• RVH, RV systolic dysfunction

• Increased myocardial reflectivity

• Biatrial enlargement
  • Increased interatrial septum thickness

• Small pericardial effusion

• Strain imaging
  • Impaired longitudinal strain
  • Basal-apical gradient
Summary

Echo Evaluation of Cardiac Amyloidosis

- Difficult diagnosis, multiple echo findings of variable specificity
- Strain imaging improves diagnostic accuracy\(^1\)
  - Consider in patients with unexplained HF and normal EF
- Clinical context
  - AL amyloidosis - plasma cell dyscrasia (MM), neuropathy
  - Wild type-ATTR – Caucasian males, >50 yrs
  - Mutant-ATTR – family history
  - Consider other testing – CMR, Tc99m-Pyrophosphate, biopsy
- Can present with intra-cavitary gradient and/or LVOT obstruction\(^2\)

\(^1\) Phelan et al. Heart 2012;98:1442
\(^2\) Philippakis et al. Circ 2012;125:1821

Case 2

- 67 yo F, DM II, hypertension, hyperlipidemia, ex-smoker, obesity (BMI 39 kg/m\(^2\))
- Referred with worsening dyspnea on exertion, NYHA class III symptoms
- Physical exam (challenging)
  - No JVD, S3, pulmonary rales or peripheral edema
- Referred for echo and coronary angiography
  - Angiogram – nonobstructive CAD (<50% lesions)
Normal LV and RV size, wall thickness and function

LA volume index 36 mL/m²  TR velocity 2.1 m/s

Mitral E velocity 0.9 m/s  E/A ratio 1.0

Septal e’ 6 cm/s, E/e’ 15

Lateral e’ 10 cm/s, E/e’ 9, Average 13

Diastolic function = Indeterminate
Strain imaging not performed

Case 2

• Additional work-up – CXR, PFTs unremarkable

• Symptoms progressed
  • Referred for Exercise Echo for systolic and diastolic function assessment

• Exercise – Bruce supine bicycle protocol
  • 4:18 mins:secs, METs 4.1, stopped due to dyspnea
  • Normal BP response to exercise
  • Blunted HR response to exercise

• Echo findings at with exercise
Exercise Echo – Systolic Function

Baseline

Peak Exercise

Exercise Echo – Diastolic Function

• Conclusion
  • Preserved systolic function at rest and with exercise, no regional wall motion abnormalities
  • Normal diastolic function at rest with evidence of diastolic dysfunction with exercise
  • Consider diagnosis of HFpEF if other causes of diastolic heart failure are excluded. Clinical correlation required
Case Points

• **HFpEF** is common (half of all HF\(^1\)) but remains a **challenging** disease to assess clinically and by echo

• **HFpEF** patients may have **unremarkable** 2D and hemodynamic profiles by echo **at rest**

• **Diastolic exercise stress testing** indicated when resting evaluation fails to explain HF symptoms\(^2\)
  - Impaired relaxation, normal estimated mean LAP, indeterminate diastolic function

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HF with Preserved EF

• Normal response to exercise
  - Increase cardiac output, preservation of filling pressures due to **augmented myocardial relaxation** causing enhanced ventricular diastolic ‘**suction**’
  - Increased mitral early diastolic E velocity and annular e’ velocity for preserved E/e’ ratio

• HF with Preserved EF
  - Unable to augment myocardial relaxation resulting in **increased filling pressures** with variable cardiac output response
  - Unchanged or decreased mitral E with reduced e’ velocities, resulting in increased E/e’

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1. Borlaug et al. EHI 2011;32:670
2. Nagueh et al. JASE 2016;29:314

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Oh et al. Circ CV Img 2011;6:444
Diastolic Exercise Echo Testing

• Supine bike easier than treadmill, DSE not indicated

• Compare rest and exercise mitral E, annular e’ and peak TR velocities

• Often have fusion of E and A velocities at peak exercise
  • Measure in recovery when heart rate slows, get TR velocity first

• When primary indication of stress echo is CAD evaluation, prioritize LV wall motion assessment

Nagueh et al. JASE 2016;29:314

Diastolic Exercise Echo Testing

• Positive test criteria, all of the following
  1. Average E/e’ >14, OR septal E/e’ >15
  2. Septal e’ velocity <7 cm/s
  3. TR velocity >2.8 m/s

• Isolated TR velocity elevation with exercise may be secondary to increasing pulmonary blood flow

• If indeterminate, consider invasive exercise hemodynamic assessment if available

Nagueh et al. JASE 2016;29:314
Cases in Cardiomyopathy and Heart Failure – Normal EF

• Thank you

• nmfine@ucalgary.ca