Echocardiography in systemic disease

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Disclosures
No relevant disclosures
Echocardiography in systemic disease

Systemic diseases causing cardiac dysfunction are unusual.

Specific patterns of cardiac dysfunction can provide clues to underlying systemic diseases.

Echocardiography can serve as a marker of disease and prognosis.

Case 1 - AW

71-year-old with HIV who presents with syncope.

After waking to use the restroom, he felt dizzy and nausea and then collapsed. He woke up on the floor with a bloody nose.
12 lead ECG

AP CXR
Thoughts?

- Severe concentric LVH
- Small pericardial effusion
- Moderately reduced LV systolic function
- Severe left atrial enlargement
- Diastolic dysfunction

GLS_Endo_Peak_A4C: -5.6 %
GLS_Endo_Peak_A2C: -6.5 %
GLS_Endo_Peak_A3C: -7.0 %
GLS_Endo_Peak_Avg: -6.4 %
Cardiac amyloidosis

Increasingly recognized.

New therapies for TTR amyloidosis (tafamidis, etc.)

Patients usually present with symptoms of right heart failure.

Syncope is also a common presentation due to frequent bradyarrhythmias or advanced atrioventricular block.
Cardiac amyloidosis - Diagnostic imaging

Strain imaging can be very helpful:
1) Overall reduction in GLS
2) Pattern of relative apical sparing

Sensitivity – 93%; Specificity – 82%

Phelan et al. JASE. 2014;27(8):888

Other modalities

**Cardiac MRI:**
- Diffuse DGE
- Increased ECV
- Inability to “null” myocardium

**Bone tracer scintigraphy:**
- Measures cardiac uptake of 99mTc-labeled pyrophosphate
- Grade 0-3 scale (normal 0)
- Highly specific for ATTR (in absence of monoclonal protein)
Ms. AS

66-year-old woman with ESRD

Restrictive cardiomyopathies

Many etiologies:
- Genetic diseases
- Renal disease
- Hypertrophic cardiomyopathy
- Hypertensive heart disease
- Amyloidosis
- Etc.

Clinical history is key
Strain pattern can be helpful

Case 2 – CS

74-year-old woman with prior breast cancer and recent COVID-19 infection who presents with worsening dyspnea over the past week.

Her dyspnea acutely worsened prompting emergent attention and was accompanied by nausea, vomiting, and chest pressure.
Labs (at presentation):
Troponin-T 0.15 ng/ml
N-terminal proBNP 6862 pg/mL
Acute Pulmonary Embolism

“McConnell’s sign”

Distinct regional pattern of RV dysfunction, with akinesia of the mid-free wall but normal motion at the apex.

COVID-19 – echocardiographic findings

The SARS-CoV-2 has a variable effect throughout the body.

Common cardiac complications include:
- Myocarditis
- Stress-induced cardiomyopathy
- Myocardial infarction
- Right heart failure
COVID-19: Echo findings

Review of 100 hospitalized patients:
RV dilation/dysfunction – 39%
LV diastolic dysfunction – 16%
LV systolic dysfunction – 10%

Patients with elevated troponin – more likely to have abnormalities in right/left ventricular function, pericardial effusions.

Higher rate of mortality.

Szekely et al. Circulation. 2020;142(4):342
Teran et al. JASE. 2020;33(8):1040

Mr. MR

54-year-old man with Hb SC disease
Right heart failure/pulmonary hypertension

Numerous causes – WHO groups

As cardiologists, we often uncover systemic disease.

Echo with RV dysfunction can be the beginning of the evaluation.
Case 3: JR

72-year-old man with HTN, HLD, and T2DM with a history of sudden cardiac arrest.
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72-year-old man with HTN, HLD, and T2DM with a history of sudden cardiac arrest.

Normal coronaries by coronary angiography

History of pulmonary sarcoidosis
Cardiac sarcoidosis

Mid wall enhancement in non-vascular distribution.

Sarcoid can have multiple patterns. The main thing is to keep it in mind.

PET-FDG can be helpful
Sarcoïdose – Granulomes

- 11% du atrium droit
- 73% du septum interventriculaire
- 46% du mur ventriculaire droit
- 7% du atrium gauche
- 96% du mur ventriculaire gauche libre

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**Spectrum of Echocardiographic findings**

<table>
<thead>
<tr>
<th>Normal</th>
<th>Focal aneurysms</th>
<th>Dilated Cardiomyopathy</th>
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Patients with normal CMR are unlikely to have significant cardiac involvement.

Case 4: AG

47-year-old with Hb SS disease presenting with lower extremity edema, abdominal fullness, and orthopnea.

History of multiple blood transfusions.
Iron overload/Hemochromatosis

Iron overload results in an infiltrative cardiomyopathy

Iron overload typically causes restrictive or dilated cardiomyopathy.

Hypertrophy is uncommon.

Hemochromatosis

- Diagnosis is critical, since reversible
  - Males 9:1
  - 2-3/1000 population
  - Ferritin usually > 500, transferrin > 50%
- Normal wall thickness
- Arrhythmias, conduction abnormalities

Intracellular iron – directly toxic to myocytes

Courtesy of William Edwards, MD
Iron overload

Idiopathic cardiomyopathy

Patients at risk for IOC

Transthoracic echocardiography with full diastolic function assessment and mitral annular tissue velocities

Abnormal LV diastolic function
Or decreased peak systolic tissue velocity

Yes

No

Cardiac MRI with T2*

Repeat transthoracic echocardiography every 1-2 years

Blue= reversible, yellow= usually not reversible; Modified from AGA clinical teaching project
Diagnosing iron overload

Iron overload is a unique circumstance when T2* times are decreased by MRI.

Iron overload results in an infiltrative cardiomyopathy. CMR can help resolve which organs are involved.

Gujja et al. JACC. 2010 Sep, 56 (13) 1001–1012
Case 5: AP

35-year-old-woman with worsening dyspnea on exertion.

Pruritic rash present on the upper arms.

Elevated ESR.

Laminar thrombus in the left ventricular apex.
Hypereosinophilic syndrome (HES)

HES is defined as unexplained eosinophilia >1500 cells/mL for >6 months

AND

Single/multiple organ system dysfunction

3-year mortality is 75% in untreated patients.


Echo contrast helps identify thrombus
Hypereosinophilic syndrome (HES)

LV > RV inflow apical thrombo-obliteration, endocardial thickening

Restrictive diastolic dysfunction

Subvalvular thrombosis, leaflet entrapment MV > TV Leaflets; MR&TR

Hypereosinophilic syndrome (HES)

Myocarditis → Thrombus → Fibrosis

Courtesy of Sunil Mankad, MD
HES - presentation

Dermatologic (e.g., rash) – 37%
Cardiac – 5%

Commonly present with pruritic, erythematous nodules and plaques.

Can resemble hives.

Case 6: RL

46-year-old woman presenting with left-sided weakness and discovered to have a right MCA stroke.

History of multiple VTEs.
Family history of TTP.
Case 6: RL

46-year-old woman presenting with left-sided weakness and discovered to have a right MCA stroke.

History of multiple VTEs.
Family history of TTP.

Blood cultures negative x3
Procalcitonin low
Nonbacterial thrombotic endocarditis

Multiple names:
- Nonbacterial thrombotic endocarditis (NBTE)
- Marantic endocarditis
- Libman-Sacks endocarditis
- Verrucous endocarditis

NBTE are typically asymptomatic until embolization occurs.

Rare condition.

Postmortem rate in autopsy series 0.9-1.6% of cases

Rev Esp Cardiol. 2007 May;60(5):493-500.

Nonbacterial thrombotic endocarditis

Deposition of sterile platelet thrombi on heart valves (commonly aortic and mitral).

Usually affects undamaged valves.

Associations:
- Advanced malignancy: ~80% of cases
- Systemic lupus erythematosus: ~10% of cases
- Others: antiphospholipid syndrome, rheumatoid arthritis, sepsis, burns, etc.
Case 7: CS

56-year-old woman with positional chest discomfort, shortness of breath, and palpitations.

History of alopecia.
Case 7: CS

56-year-old woman with positional chest discomfort, shortness of breath, and palpitations.
History of alopecia.

Laboratory evaluation:
(+) ANA 1:1280 nuclear and speckled
(+) dsDNA antibodies
(+) Smith
(+) Sm/RNP
(+) SSA

Systemic lupus erythematous

SLE can affect any part of the heart.

Coronary artery disease is more common in patients with lupus.

Congenital heart block occurs frequently as a result of maternal anti-Ro/SS-A antibodies.

Cardiac manifestations include:
Myocarditis
Pericarditis (effusion is often ANA positive)
Valvular disease
Thrombosis/NBTE
Cardiac conduction defects.

Case 8:

44-year-old woman who presents with shortness of breath, tremors, palpitations, and lower extremity edema.

Labs:
TSH <0.01 uIU/mL
Free T4 >7.77 ng/dL
TPO >900 IU/mL
Case 8:

44-year-old woman who presents with shortness of breath, palpitations, and lower extremity edema.

Treated for thyroid storm with methimazole, beta-blockers, and anticoagulation.

Spontaneously converted to sinus rhythm.
Estimated RVSP 62 mmHg
Estimated mean PA 34 mmHg

\[ PA_{\text{mean}} = 0.6 \times PA_{\text{SP}} + 2 \text{ mm Hg} \]

Case 8:

44-year-old woman who presents with shortness of breath, palpitations, and lower extremity edema.

Eventually, underwent thyroidectomy.

1 year follow-up for surveillance of valvular heart disease
Estimated RVSP 24 mmHg
Estimated mean PA 16 mmHg

PA\textsubscript{mean} = 0.6 \times PA\textsubscript{SP} + 2 \text{ mm Hg}

Graves’ disease

Atrial fibrillation is common
- Seen in 10% of all patients with hyperthyroidism
- More common in thyroid storm

Congestive heart failure is also common
- 94% patient have co-existing atrial fibrillation
- ~50% will have systolic dysfunction – thought to be tachycardia-mediated cardiomyopathy

Pulmonary hypertension increasingly described.

Reversible Pulmonary Hypertension, Tricuspid Regurgitation and Right-sided Heart Failure Associated With Hyperthyroidism: Case Report and Review of the Literature

Lozano, Hector F. MD, Sharma, Charu N. MD, FACCI  
Author Information 
Cardiology in Review: November-December 2004 - Volume 12 - Issue 6 - p 299-303
doi: 10.1097/01.lcr.0000137529.03186.e3

Cardiovascular abnormalities in hyperthyroidism: a prospective Doppler echocardiographic study

Jordi Mercè, Sara Ferràs, Carmina Oltra, Esther Sanz, Joan Vendrell, Immaculada Simón, Mercè Campsbi, Alfredo Bardají, Cristóbal Rídao

Conclusion: In patients with hyperthyroidism, there is a high prevalence of pulmonary hypertension and atrioventricular valve regurgitation. These abnormalities usually correct after treatment for hyperthyroidism.

Case 9:

70-year-old woman with history of Graves’ disease s/p remote radioactive iodine ablation referred for abnormal finding on echocardiogram.

Recently admitted for complicated UTI with E. coli bacteremia.
Hypothyroidism

Pericardial effusion frequently seen (25% patients)
  ◦ Rarely compromises ventricular function

Severe hypothyroidism results in reduced cardiac output.

Hypertension present in 20-40% patients
  ◦ Particularly diastolic hypertension

Gumieniak O. J Clin Endocrinol Metab. 2004;89(7):3455.
Case 10:

72-year-old man with known metastatic neuroendocrine tumor being evaluated for worsening abdominal distention and lower extremity edema.
Carcinoid disease

Rare neuroendocrine tumors – 5/100,000

Known to release vasoactive substances with paracrine and endocrine effects
- Serotonin (5-hydroxytryptamine [5-HT])
- Tachykinins
- Prostaglandins
- Histamine
- Kallikrein

Carcinoid heart disease (paraneoplastic process) occurs in approximately 20% of all carcinoid patients
Pathophysiology

5-HT induced plaque formation on the valve cusps and subvalvular apparatus.
- Myofibroblasts
- Smooth muscle cells
- Extracellular material including collagen.

The valve cusps thicken, stiffen, and retract leading to combined regurgitation and stenosis.

Images courtesy of Muhamed Saric, MD.

Carcinoid – tricuspid disease

Dilated tricuspid annulus

Diffusely thickened, retracted leaflets.

Can often result in “torrential TR” due to coaption gap.

Baron et al. JACC Img. 2021.
Carcinoid – pulmonic disease

Like tricuspid valve, the pulmonic valve becomes thickened and retracted.

Reduced leaflet mobility.

Results in pulmonic insufficiency and stenosis.

Baron et al. JACC Img. 2021.
Images Courtesy of Sunil Mankad, MD

Carcinoid heart disease

Treatment of tumor usually does not lead to regression of valvular disease.

Surgical treatment often necessary.

Left-sided valves involved in ~10% cases (shunt or bronchial lesions present).

Baron et al. JACC Img. 2021.
In review

Specific patterns of cardiac dysfunction can provide clues to underlying systemic diseases.

Echocardiography can serve as a marker of disease and prognosis.

Echo often a clue to look further!

Thank you!

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