Echocardiography in Systemic Diseases

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DISCLOSURE

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Off Label Usage
None
Echo in Systemic Diseases

• Systemic diseases with secondary cardiac involvement are uncommon

But

• Echo can identify unique, characteristic features and echo may be the first clue to the underlying systemic illness

Cardiac Involvement in Systemic Diseases

• Autoimmune
• Endocrine
• Collagen Vascular Diseases
• Malignancy
• Amyloid/Infiltrative Diseases
• Radiation Induced Heart Disease
• Drug Induced Valvulopathy
Case

• 27 y/o female who presents with dyspnea, chest pain, and fatigue
  – NYHA class III
• Abnormal nuclear perfusion stress test led to coronary arteriography
  – Normal coronaries but LV gram suggestive of “Hypertrophic CM” (EF 75%)
• Elevated Sedimentation Rate
• Referred to Mayo Clinic → Echo performed
Apical 4 Chamber Views

Diastolic Function

- MV Dec. Time = 105 msec
- MV Emax = 1.1 m/sec
  - e' = 0.04 m/sec
  - E/e' 28
What is the Diagnosis?

1. Hypertrophic Cardiomyopathy (Apical Variant)
2. Amyloidosis
3. Eosinophilic Endomyocardial Disease
4. LV Noncompaction
5. LV Myxoma

RV Biopsy (H&E Stain)
Hypereosinophilic Syndrome
Cardiac Manifestations

• Persistent increase in eosinophil count eosinophil count > 1500 cells/mm3
• CHF (dyspnea)
  – Restrictive Cardiomyopathy
  – Mitral regurgitation
• Systemic embolization

Eosinophilic Heart Disease
4 Stages:
1) Acute inflammatory myocarditis
2) Eosinophil rich thrombus deposition
   - Mediated by injured endothelium
3) Endocardial thickening
   - Valve involvement
4) Fibrosis

Hypereosinophilic Syndrome (HES)
Cardiac Involvement: 40-60% of patients

LV > RV inflow apical thrombo-obliteration, endocardial thickening

2-D Echo & Doppler Findings

Restrictive diastolic dysfunction

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

Natural History Hypereosinophilic Syndrome

Myocarditis → Thrombus → Fibrosis

Image courtesy of Leslie Elvert RDCS
Basal LV Fibrosis with Mitral Posterior Leaflet Tethering

Eosinophilic Heart Disease
Contrast Helpful
Hypereosinophilic Syndrome

Treatment

- Medical therapy
  - Corticosteroids
  - Hydroxyurea
  - Interferon
  - CHF Meds
- Surgical Therapy
  - Palliative

Echo Differential Diagnosis

- Apical hypertrophic CM
- LV Noncompaction
- LV tumor
  - Myxoma
  - Papillary fibroelastoma
- Ischemic LV dysfunction with apical thrombus

Differential Diagnosis: LV Myxoma
Differential Diagnosis: Apical HCM

Our Case:
TTE after 2 months of anticoagulation and 1 month of prednisone therapy
Patient with CREST Syndrome: Dyspnea and Edema

RVSP: 75 mmHg

Scleroderma and Pulmonary HTN

- PH present in 8-12% of scleroderma patients
  - Higher risk in CREST patients
- Accounts for 30% of deaths
- Screening for PH recommended
- RV dysfunction, cardiac index and pericardial effusion are markers of poor prognosis in PH
33 Year Old Female → Multiple Strokes

- ANA positive and Antiphospholipid antibodies present
- Libman-Sacks endocarditis

Systemic Lupus Erythematosus Cardiac Involvement

- Pericarditis (fluid ANA+)
  - 50-60% of cases have pericardial effusions
- Lupus anticoagulant
- Anticardiolipin or Antiphospholipid Abs
- Myocarditis
- Coronary arteritis
- Libman-Sacks (Marantic) vegetations
18 y.o. Female with Occipital Stroke

TEE

- Lupus anticoagulant + antiphospholipid antibodies present
- Libman-Sacks endocarditis

Not only the mitral valve!
Antiphospholipid Syndrome Diagnosis confirmed at surgery

- IgG and IgM Antiphospholipid antibody
- Importance of recognition
  - Unlikely repair
  - Choice of prosthesis??
  - Anticoagulation??

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Courtesy of W Edwards MD
39 year old male with diarrhea, flushing and weight loss

Carcinoid Syndrome
Carcinoid: Echo Features

Tricuspid valve
- Thickened leaflets
- Retracted leaflets
- Fixed semi-open position

Pulmonary valve
- Thickened cusps
- Retracted and rigid

Connolly HM. Curr Cardiol Rep. 2006

Severe (Torrential) Tricuspid Regurgitation
Systolic RV → RA pressure equalization

TR CW Doppler

RV
RA

Courtesy of Dr. WK Freeman
Pulmonary Valve Involvement

Adapted from Mayo Image Data Base, William Edwards, MD
Carcinoid Heart Disease

- Carcinoid tumors: 1-2/100,000
- Carcinoid syndrome in 20-30%
- Deposition of a matrix-like material on the valves and endocardium of the right side of the heart
- Treatment of tumor does not cause regression of valve disease

Connolly HM. Curr Cardiol Rep. 2006

Carcinoid Syndrome: 3D TTE

Courtesy of Denisa Muraru, MD, PhD
Padua, Italy

Eur Heart J Cardiovasc Imaging 2012
Carcinoid Heart Disease

Over 50% of patients with Carcinoid Syndrome develop cardiac involvement

Flushing  ➡️  Vasoactive Substances 5-HIAA ➡️  Diarrhea

Wheezing

Courtesy of Dr. Heidi Connolly
Outcome of Cardiac Surgery for Carcinoid Heart Disease

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Objectives. The hypothesis was that cardiac surgery for symptomatic carcinoid heart disease in conjunction with adjuvant therapy could improve the long-term survival of patients with carcinoid heart disease.

Background. Patients with carcinoid heart disease have a distressing prognosis, most die of progressive right heart failure within 1 year after onset of symptoms. Improved therapies for the systemic manifestations of the carcinoid syndrome have resulted in symptomatic improvement and prolonged survival in patients without heart disease.

Methods. Twenty-six patients with symptomatic carcinoid heart disease underwent valvular surgery. Preoperative clinical, laboratory, Doppler echocardiographic and hemodynamic factors were evaluated. The survival of the surgical group was compared with that of a control group of 40 medically treated patients.

Results. There were nine perioperative deaths (35%), primarily from postoperative bleeding and right ventricular failure. Of the 17 surgical survivors, 8 were alive at a mean of 28 months of follow-up. The postoperative functional class of the eight surviving patients was substantially improved. Late deaths were primarily due to hepatic dysfunction caused by metastatic disease. The only predictors of operative mortality (p = 0.03) was low voltage on preoperative electrocardiography (limb lead voltage ≤5 mm). Predictors of late survival included a lower preoperative somato- statin requirement and a lower preoperative urinary 5-hydroxy indoleacetic acid level. There was a trend toward increased survival for the surgical group compared with the control group.

Conclusions. Because new therapies have improved survival in patients with the malignant carcinoid syndrome, cardiac involvement has become a major cause of morbidity and mortality. Valvular surgery is the only definitive treatment. Although cardiac surgery carries a high perioperative mortality, marked symptomatic improvement occurs in survivors. Surgical intervention should therefore be considered when cardiac symptoms become severe.

(J Am Coll Cardiol 1995;25:418–4)

Carcinoid Tumor: Liver Metastases
58 yo woman with weight loss, tremor and HR of 125

Hyperthyroidism

- Atrial fibrillation
  - difficult to rate control
- Decreased Peripheral resistance
  - hypotension
- Exacerbation of underlying CAD
  - increased myocardial O2 demand
- Tachycardia induced cardiomyopathy
Tachycardia Mediated Cardiomyopathy

- 25% of patients with LV dysfunction & AF will have improved EF with rate control
- Usually *unaware* of rhythm
- Resting heart rate - poor indicator of overall rate control
- Consider in all pts with AF & LV dysfunction


2 Years after Cardioversion and Treatment of Hyperthyroidsin
Hypothyroidism:
Large Pericardial Effusion

43 year old man
43 year old man with amyloidosis

Primary Hyperoxaluria

- Rare metabolic disorder with autosomal recessive inheritance
- PHO type 1 (0.11 - 0.26 per 100,000 live births)
- Enzymatic defect resulting in enhanced conversion of glyoxalate to poorly soluble oxalate which is excreted in the urine
Hydroxychloroquine-induced Cardiotoxicity

Infiltrative Cardiovascular Diseases
Cardiomyopathies That Look Alike
James B. Sneed, MD, and Camilo-Vergara, MD
Kathleen Al-Mansour

Infiltrative cardiomyopathies are characterized by the deposition of abnormal substances that cause the myocardial wall to become progressively rigid, thereby impairing ventricular filling and decreasing cardiac chamber excursion. This decrease in ventricular excursion is reflected in a reduction of ejection fraction and an increase in end-diastolic volume (Table 1). Simultaneously, the diagnosis of infiltrative cardiomyopathy is confirmed on echocardiography, magnetic resonance imaging, and computed tomography. The severity of cardiac dysfunction is best characterized by depressed left ventricular ejection fraction, measured by echocardiography. Echocardiographic indices of cardiac function include stroke volume index (SVI), cardiac output index (COI), and left ventricular ejection fraction (LVEF).
A 60 year old male farmer is referred for evaluation of dyspnea

- NYHA Class III symptoms
- PMH: Type 2 DM
- Abnormal LFT’s
- Physical Exam:
  - 110/70 mmHg, HR 70 BPM
  - S3 gallop
  - Bronze skin

### EKG

Normal sinus rhythm
Premature atrial complexes
Left bundle branch block
Replacement of ST-T abnormalities
When compared with EKG of 16-AUG-2007 16:16, premature atrial complexes are now present and T waves have changed
CXR

Apical Images
Coronary Angiography

60 year old male farmer with Type 2 DM, bronze skin, and abnormal LFT’s

What is the most likely diagnosis?

a. Cardiac hemochromatosis
b. Cardiac amyloidosis
c. Cardiac sarcoidosis
d. Fabry’s Disease
e. Carcinoid syndrome
The evaluation of the T2* relaxation time is an excellent noninvasive correlate of myocardial iron deposition and is a useful technique to follow response to iron-chelation therapy. Myocardial T2* has been shown to have no relation to serum ferritin and liver iron overload. T2* relaxation time predicts CHF and Arrhythmias.

This patient had a T2* relaxation time of 20ms that suggests hemochromatosis.

Circulation 2009;120:1961-8
Hemochromatosis

• ↑ total body iron – intracellular deposits in heart, liver, pituitary, pancreas, gonads, skin
• Think of this when DCM seen in setting of hepatic dysfunction; diabetes, tanned skin
• Diagnosis is critical, since reversible
  – Males 9:1
  – 2-3/1000 population
  – Ferritin usually > 500, transferrin > 50%
• Normal wall thickness
• Arrhythmias, conduction abnormalities

26 year old with Hemochromatosis
Heomochromatosis: Take Home Points

- The Iron Heart is a weak heart...
- Hemochromatosis may be a cause of idiopathic dilated cardiomyopathy
  - Reversible with treatment
- Cardiac MRI (T2 relaxation time) is important in helping to establish diagnosis and monitoring treatment effects
Conclusions:
Systemic Diseases and the Echo Boards

• Carcinoid Syndrome
• Hypereosinophilic endomyocardial disease
• Sarcoidosis
• Systemic Lupus Erythematosus
• Scleroderma/Crest: Pulm Hypertension
• Amyloidosis
• Hyper or Hypothyroidism
• Radiation Heart Disease
• Drug Induced Valve Disease
• Hemochromatosis

Thank You!
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