

Constriction versus Restriction

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Objectives

- Recognizing constrictive pericarditis and restrictive cardiomyopathy as a reversible cause of heart failure.
- Understand the pathophysiology of constrictive pericarditis and restrictive cardiomyopathy.
- Echocardiographic findings differentiating between constrictive pericarditis and restrictive cardiomyopathy.
- Invasive hemodynamics findings differentiating between constrictive pericarditis and restrictive cardiomyopathy.

Case 1

A 45-year-old man is evaluated for a 6-month history of progressive dyspnea on exertion and lower-extremity edema. He can now walk only one block before needing to rest. He reports orthostatic dizziness in the last 2 weeks. He was diagnosed 15 years ago with non-Hodgkin lymphoma, which was treated with chest irradiation and chemotherapy and is now in remission. He also has type 2 diabetes mellitus. He takes furosemide (80 mg, 3 times daily), glyburide, and low-dose aspirin.

Physical examination

Afebrile.

Blood pressure of 125/60 mm Hg supine and 100/50 mm Hg standing; pulse is 90/min supine and 110/min standing. Respiration rate is 23/min. BMI is 28.

Presence of jugular venous distention and jugular venous engorgement with inspiration.

CVP of 15 cm H₂O.

Cardiac examination discloses diminished heart sounds and a prominent early diastolic sound but no gallops or murmurs.

Pulmonary auscultation discloses normal breath sounds and no crackles.

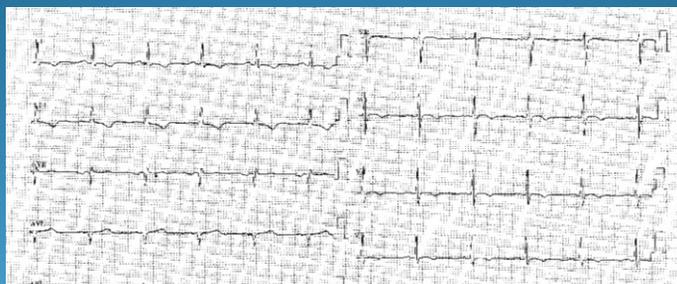
Abdominal examination shows shifting dullness

Lower extremities show 3+ pitting edema to the level of the knees.

Remainder of the physical examination is normal.

LABS and EKG

BUN 40 mg/dL, Cr 2.0 mg/dL, ALT 130 U/L, AST 112 U/L, Albumin 3.0 g/dL, UA negative for protein,



Chest xray



Case 2

70 year old female presented with dyspnea caused by minor stress. Four months ago, the patient presented with signs of dyspnea caused by moderate effort, which 2 month ago progressed to minimum effort and orthopnea. The patient also presented edema on lower limbs. Recurrent admissions for heart failure. She also has a history of hypothyroidism, carpal tunnel syndrome, hypertension and atrial fibrillation. Medications: 80 mg of Furosemide, 50 mg of Spironolactone, 75 mg of Captopril, coumadin, 100 µg of levothyroxin on a daily basis.

Physical examination

Heart rate at 100 bpm and blood pressure 90 / 80 mm Hg, Normal RR

Increased jugular venous pressure,.

Lung examination revealed a decreased vesicular sounds on the right hemithorax middle third and abolition on the lower third of both hemithoraxes.

Cardiac auscultation revealed 4th heart sound, with no murmurs or pericardial friction rub.

Abdomen was painful to percussion in right hypochondrium region. Her liver was palpated at 10 cm from the right costal edge;

There was edema +++/4+ on lower limbs.

Chest radiography revealed a huge bilateral pleural effusion .

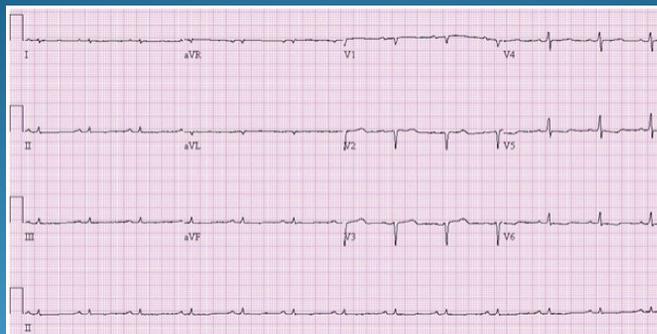
Pertinent LABS

BMP: Creatinine 2.03 m/dL, sodium 133 mEq/L, potassium 3.9 mEq/L, C-reactive protein (CRP) 18.1 mg/L,

LFT: Total bilirubin 3.75 mg/dL, direct bilirubin 2.37 mg/dL, AST 67 U/L, ALT 86 U/L, alkaline phosphatase 266 U/L,

Urinalysis showed urine specific gravity of 1.020, pH 5.5, proteinuria 0.25 g/L, epithelial cells 4,000/mL, leukocytes 2,000/mL, erythrocytes 3,000/mL, and hyaline casts 27,250/mL

EKG



Chest xray



Constrictive pericarditis

Constrictive pericarditis (CP) is a chronic inflammatory process involving the pericardium, which leads to fibrotic thickening, scarring and subsequently calcifications which could be quite extensive, extending into the myocardium

Epidemiology

9% of patients with acute pericarditis for any reason go on to develop constrictive physiology.

Frequency of a diagnosis of constrictive pericarditis is less than 1 in 10,000 hospital admissions.

67 % presented with symptoms of heart failure (HF)

8 % with chest pain

6 % with abdominal symptoms

4 % with atrial arrhythmia

5 % with symptoms of cardiac tamponade

Etiology of Constrictive Pericarditis

Idiopathic or viral — 42 to 49 %

Post cardiac surgery — 11 to 37 %

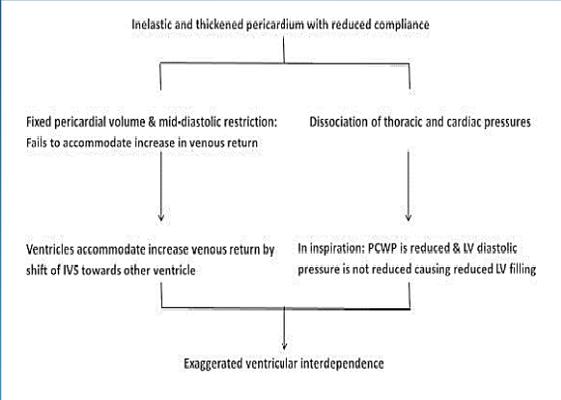
Post radiation therapy — 9 to 31 %

Connective tissue disorder — 3 to 7 %

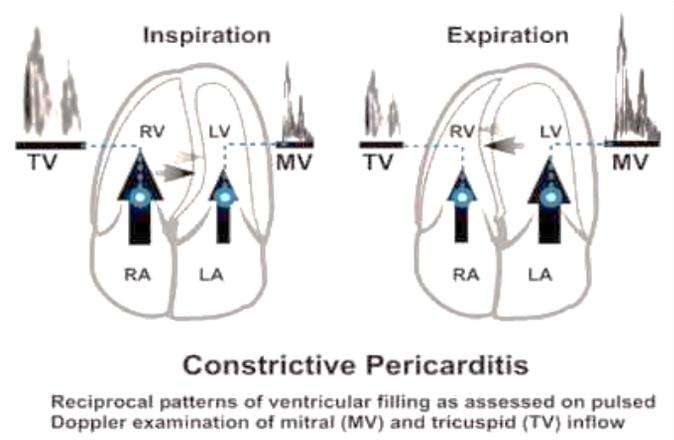
Postinfectious (tuberculous or purulent pericarditis) — 3 to 6 %

Miscellaneous causes (malignancy, trauma, drug-induced, asbestosis, sarcoidosis, uremic pericarditis) — 1 to 10 %

Pathophysiology of constrictive pericarditis



Interventricular dependence



Clinical presentation

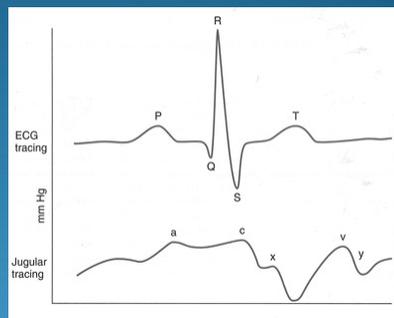
- Symptoms related to fluid overload: predominantly right sided heart failure
 - Dyspnea
 - Distended neck veins
 - Abdominal distension and pain
 - Lower extremity edema
 - Anasarca

- Symptoms related to decrease cardiac output
 - Dyspnea
 - Fatigability/Tiredness
 - Tachycardia

Physical examination

Elevated JVP: deep x and y descent

Kussmaul sign
 Peripheral edema
 Ascites
 Hepatomegaly
 Pleural effusion
 S₃
 Pulsus paradoxus
 Cachexia- late stages



Diagnosis

- High index of clinical suspicion.
- Labs: Non specific....low albumin, elevated liver enzymes, elevated creatinine
- EKG: non specific St T wave changes, low voltage
- Chest xray: pericardial calcification



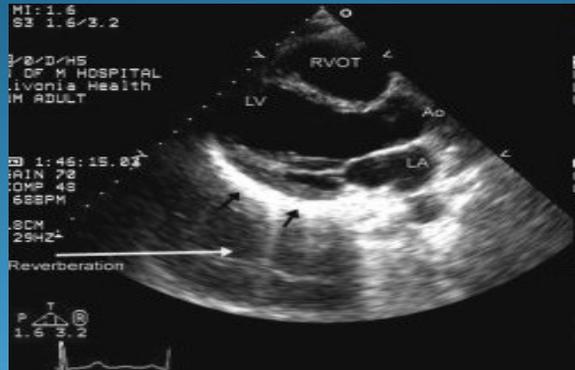
CT cardiac

- Pericardial calcification and thickening (>4 mm)
- Pericardial thickening (>4 mm) seen in 72% and calcification in 25%; Talreja, Circ 2003.
- Normal pericardial thickening does not exclude constrictive pericarditis
- Additional findings: dilated IVC, deformed ventricular contours and angulation of ventricular septum, failure of adjacent structures to pulsate during cardiac cycle.

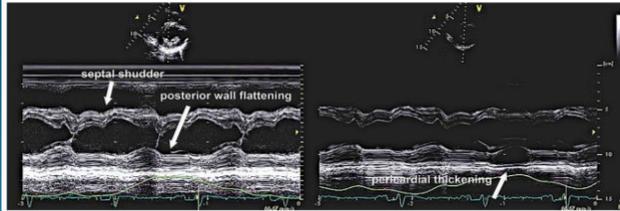


Echocardiography

Pericardial calcification in echo

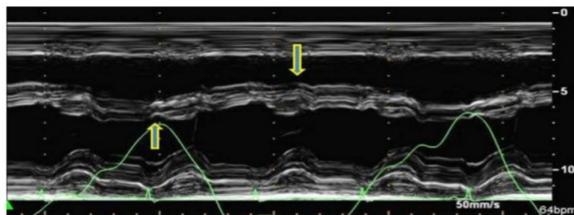


M Mode

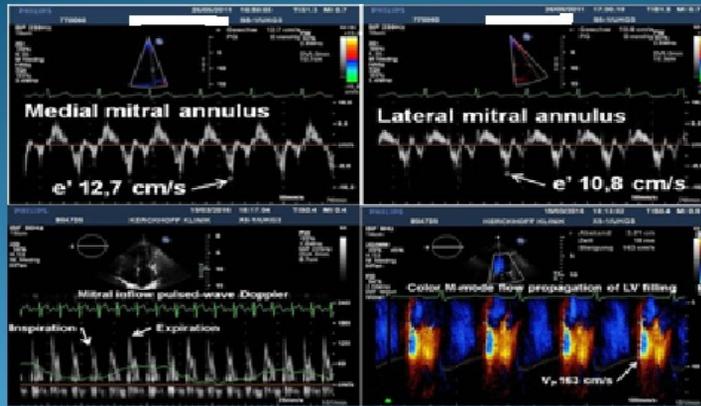


CP: M-mode of parasternal short-axis view. Several M-mode features of CP are seen, including diastolic flattening of the posterior wall, pericardial calcification (better seen in the right panel with a reduced 2D gain and fundamental imaging), a septal diastolic shudder consistent with the septal bounce seen by 2D imaging, and respiratory variation in LV cavity size.

Ventricular interdependence

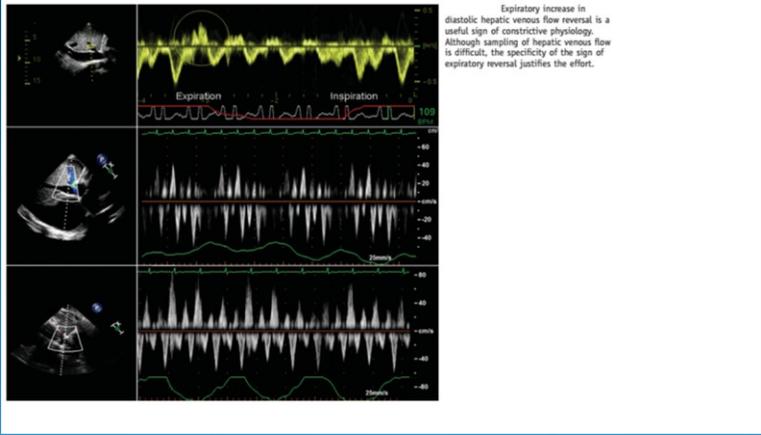


M-mode echocardiogram of a typical CP patient with the ventricular septum moving with respiration toward the left ventricle with inspiration (upward arrow) and toward the right ventricle with expiration (downward arrow). A simultaneous respirometric recording is shown at the bottom of the figure.

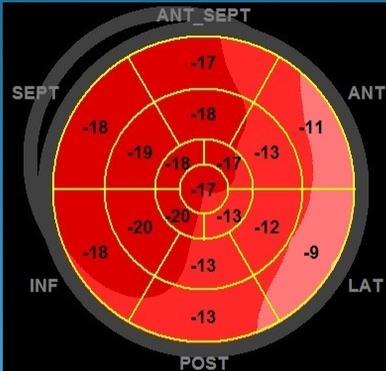


Tissue Doppler Characteristics of CP

- Annulus Reversus: Lateral $e' <$ Medial e'
- Annulus Paradoxus: Inverse correlation between E/E' and PCWP

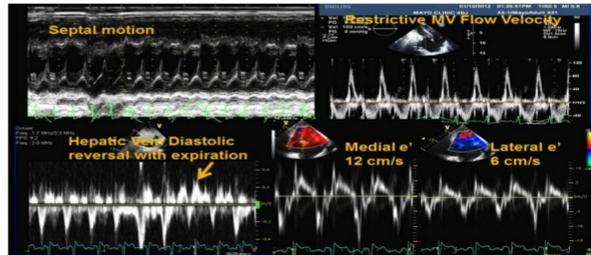


Strain imaging



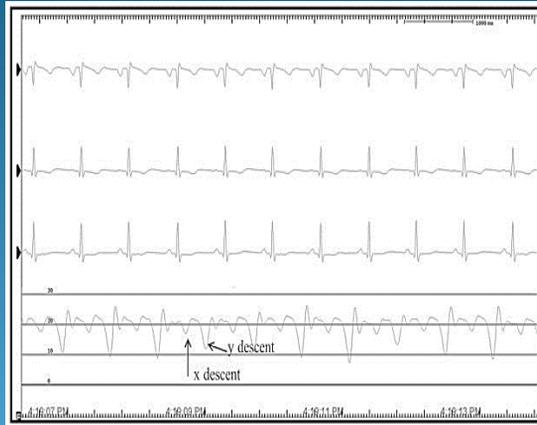
87% sensitive and 91% specific for constrictive pericarditis

CP Echo Criteria – ACC 2015 (Jae K Oh)



Invasive hemodynamics

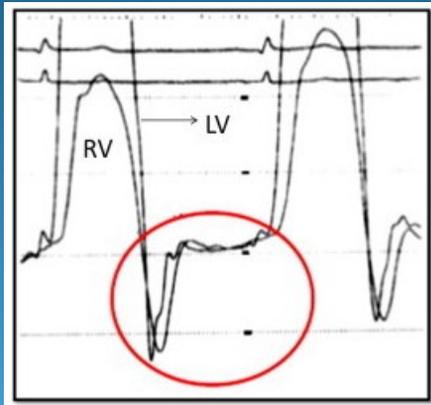
RA pressure



Simultaneous LV and RV pressure



Dip & plateau pattern or square root sign



Treatment

- Initial trial of NSAIDs and colchicine in early phase. Definitive treatment is surgical
- Earlier the better
- Complications
 - excessive bleeding
 - Atrial and ventricular arrhythmias
 - Ventricular wall ruptures.
- Published surgical mortality 5-15%.
- Perioperative mortality rate 6.1%.

Survival postcardiotomy

- Depends on the underlying cause.
- Idiopathic with best prognosis (88% survival at 7 yrs),
- Constriction due to cardiac surgery (66% at 7 years).
- Worst prognosis in post radiation constrictive pericarditis (27% survival at 7 years).
- Predictors of poor outcomes
 - Prior Radiation
 - Renal dysfunction
 - Pulmonary hypertension
 - Systolic heart failure
 - Hyponatremia
 - Advanced age.

Restrictive Cardiomyopathy

Restrictive cardiomyopathy is a myocardial disorder that usually results from increased myocardial stiffness that leads to impaired ventricular filling.

Types of restrictive cardiomyopathy

Myocardial

Non-Infiltrative

- Idiopathic
- Familial
- Hypertrophic
- Scleroderma

- Infiltrative
- Amyloid
- Sarcoid
- Gaucher's

Storage Disease

- Hemochromatosis
- Glycogen storage disease
- Fabry's

Endomyocardial

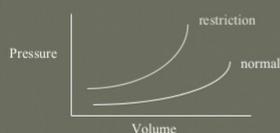
- Endomyocardial Fibrosis
- Hypereosinophilic syndrome
- Carcinoid
- Metastatic Malignancy
- Radiation
- Chemotherapy toxicity
- Drugs: serotonin, methysergide, ergotamine, mercurial agents, busulfan

Pathophysiology

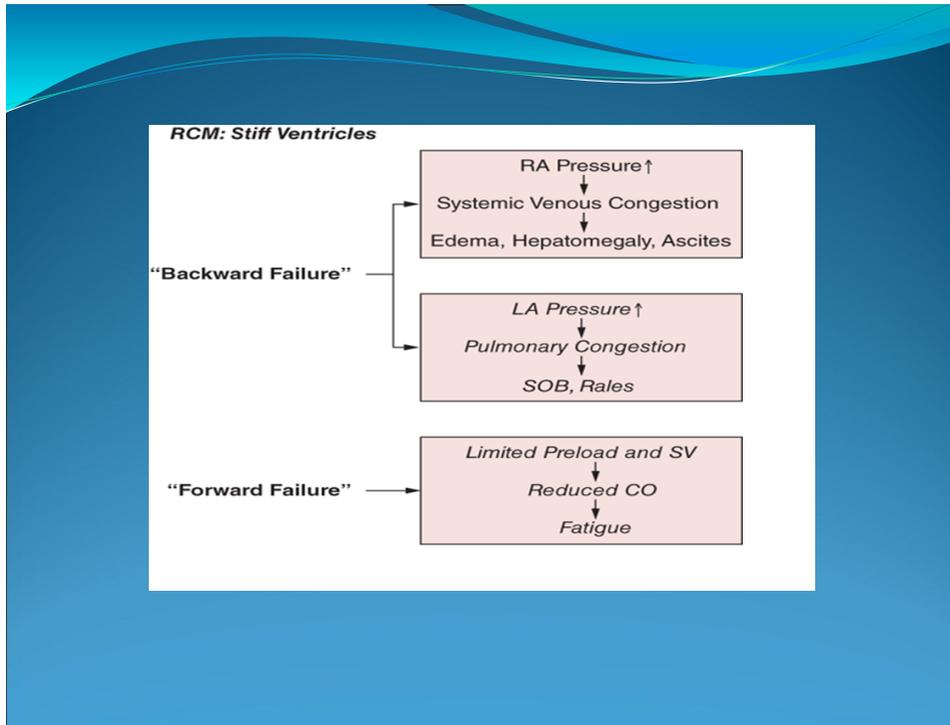
Restrictive Cardiomyopathy

Characterized by:

- * impaired ventricular filling due to an abnormally stiff (rigid) ventricle
- * normal systolic function (early on in disease)
- * intraventricular pressure rises precipitously with small increases in volume



Causes : infiltration of myocardium by abnormal substance
fibrosis or scarring of endocardium



Symptoms

- Volume Overload
 - Fatigue
 - Dyspnea
 - Orthopnea
 - Nocturnal dyspnea
- Arrhythmia : palpitations, syncope, exercise intolerance
- Reduced Cardiac Output
 - Exercise intolerance
 - Cognitive difficulties
 - Angina, Dyspnea, Syncope-on exertion
 - Sudden cardiac death

Restrictive Cardiomyopathy Findings

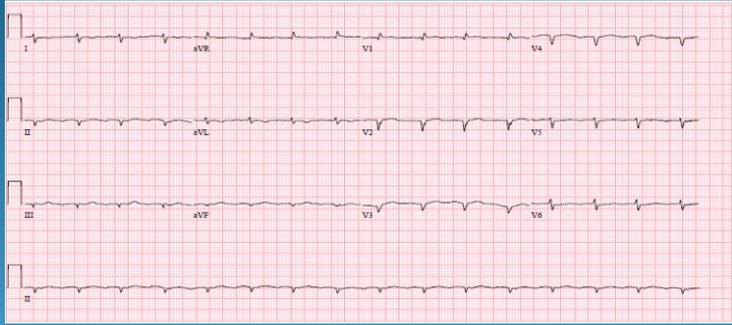
- Pulse-Tachycardia, Bradycardia, Irregular, Weak peripheral pulse
- BP-Low
- Jugular venous pressure- Raised
- S₃ and/or S₄
- Pulmonary Crackles
- Inspiratory increase in venous pressure (Kussmaul's sign)
- Findings of Rt. Heart Failure may predominate i.e edema, hepatomegaly

Diagnosis modalities

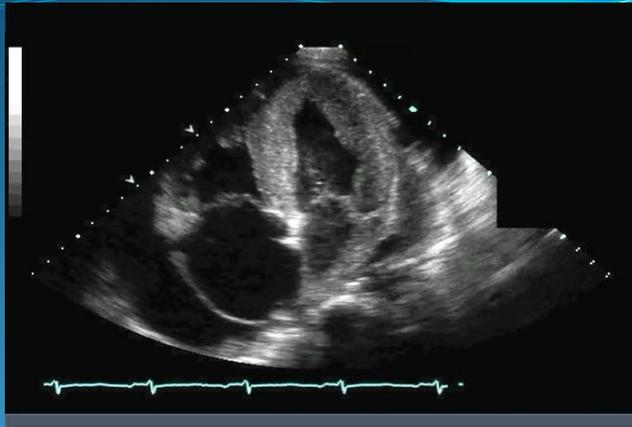
➤ Labs

- Hypereosinophilic syndrome, endomyocardial fibrosis: Eosinophils count
- Amyloid: paraproteinemia, elevated liver enzymes
- Sarcoidosis: Evidence of sarcoidosis in lung
- Hemochromatosis: elevated Ferritin level

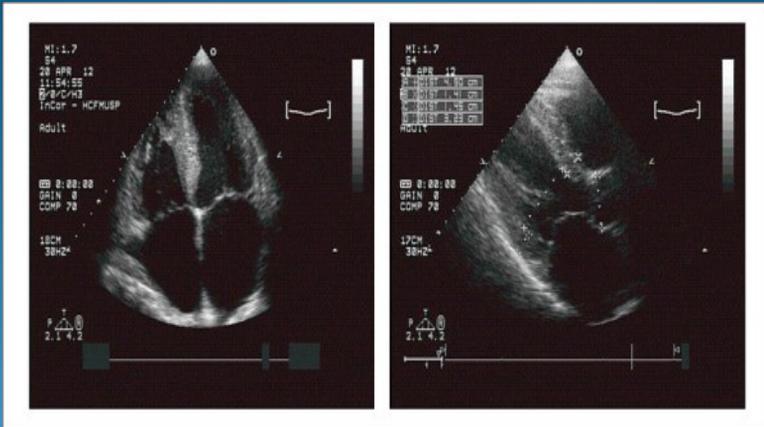
EKG

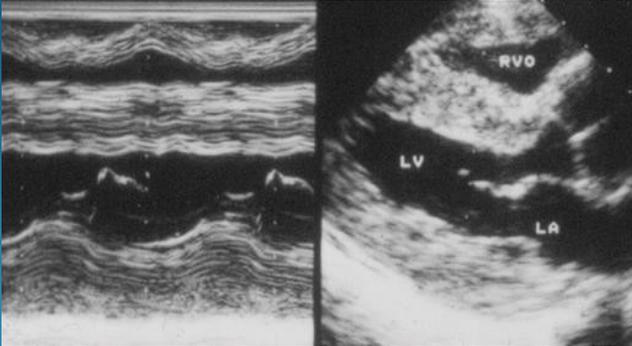


Echocardiography

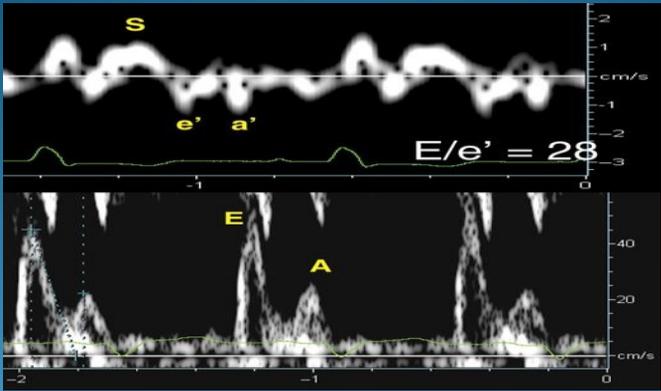


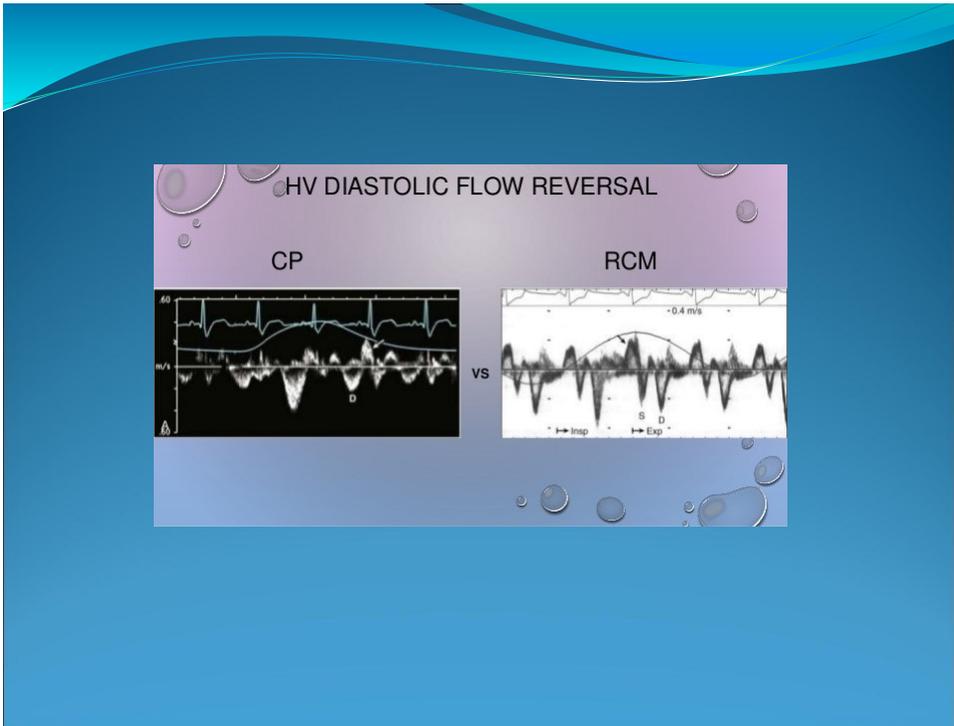
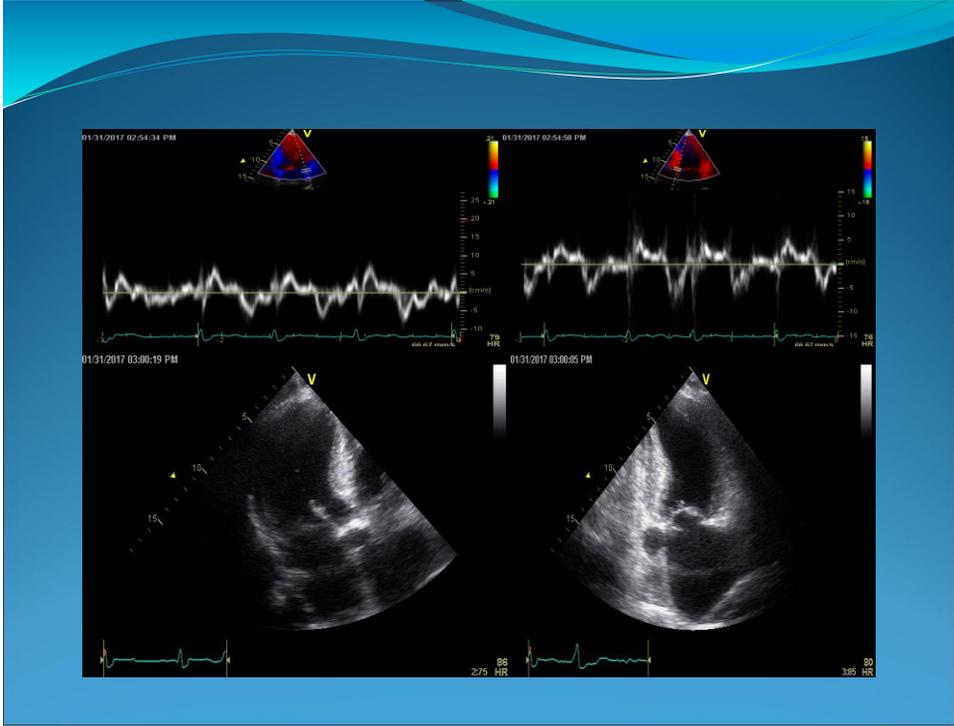
Severely dilated atrias and small ventricles





Thickened left ventricle walls and valves

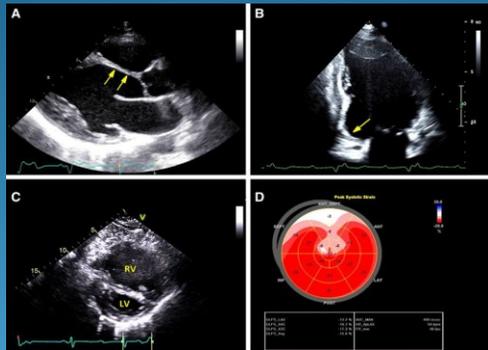




Strain imaging



Sarcoidosis



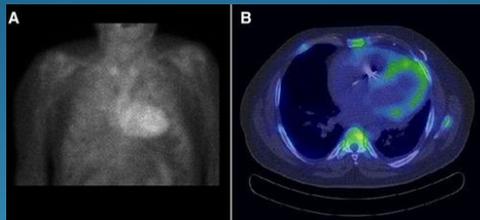
Other imaging modalities

- FDG-PET scan for sarcoidosis
- Tc-PYP scan for amyloidosis

Tc 99 Pyrophosphate scan

Eli muchtar, et al, Restrictive cardiomyopathy, Circ Research 2017

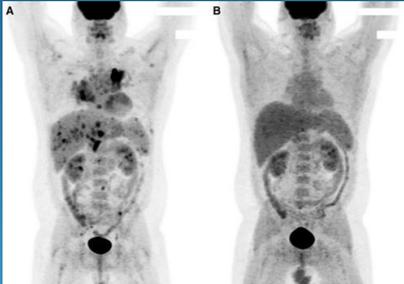
Specificity of 86% in ATTR Amyloidsis



FDG-PET scan

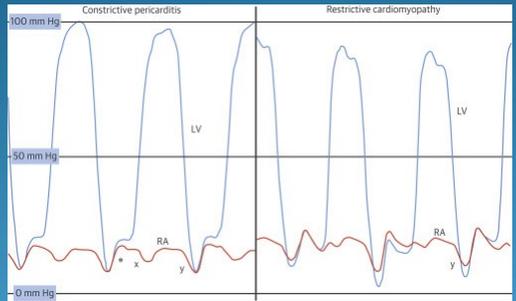
Eli muchtar, et al, Restrictive cardiomyopathy, Circ Research 2017

Sn 89% and Sp 78%

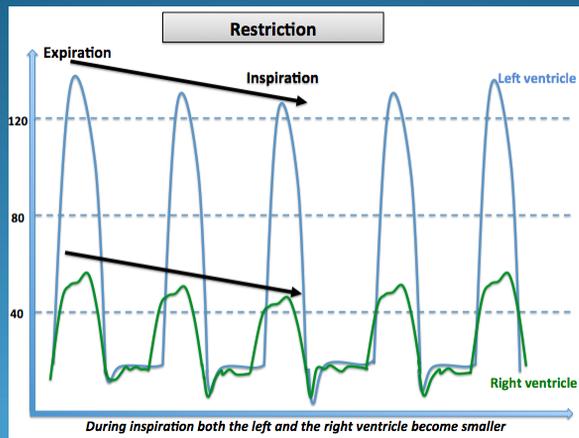


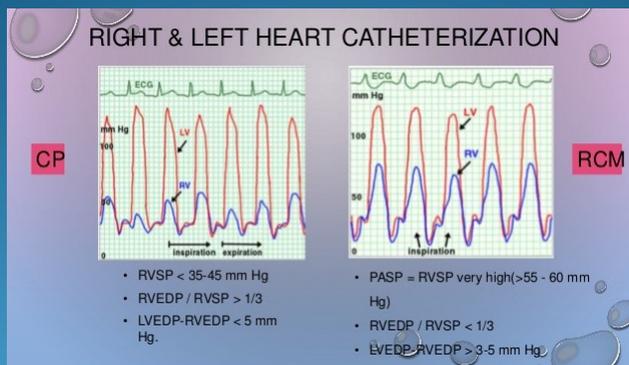
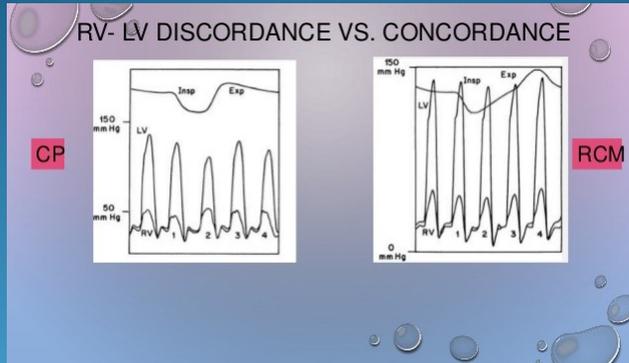
Invasive hemodynamics

Right atrial tracing



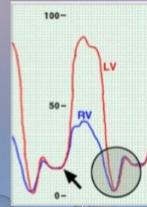
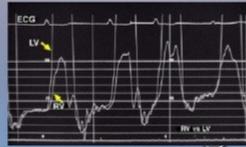
Geske et al. JACC 2016





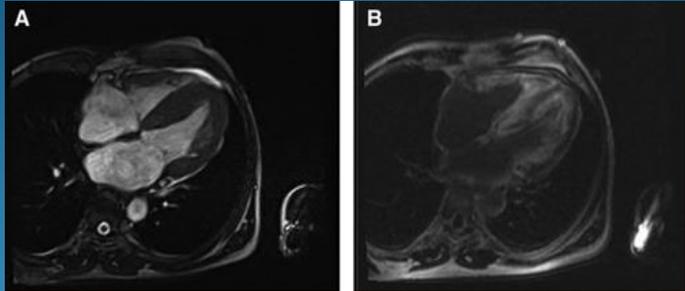
RIGHT & LEFT HEART CATHETERIZATION

- Dip and plateau pattern in diastolic waveform (square root sign)
 - Constrictive pericarditis
 - Restrictive cardiomyopathy
 - RV ischemia



Cardiac MRI

- LA/RA volume >1.32 differentiate between CP and RC.
- 92% specific for pericardial thickening >4 mm.
- Extent of pericardial inflammation in CP.
- Extent of myocardial involvement in CP.
- Pattern of myocardial enhancement in RC.



A: early gadolinium phase and B: late gadolinium enhancement.

Eli Muchtar. Circulation Research. Restrictive Cardiomyopathy. 2017 AHA

Differentiation between constrictive pericarditis and restrictive cardiomyopathy
Geske et al JACC 2016

	Constrictive pericarditis	Restrictive cardiomyopathy
Patient history	<ul style="list-style-type: none"> Cardiac procedures Radiation Connective tissue disease Infection (TB) 	<ul style="list-style-type: none"> Genetic disease Radiation Connective tissue disease Infection (eosinophilic disease) Amyloidosis
Jugular venous pressure (JVP)	<ul style="list-style-type: none"> ↑y ↑x Kussmaul's sign 	<ul style="list-style-type: none"> ↑y ↓x Kussmaul's sign
Lab/ECG/X-ray	Nonspecific	Nonspecific
Advanced cardiac imaging	<ul style="list-style-type: none"> ↑ Pericardial thickness Pericardial calcification Pericardial effusion ↑ Imaging correlates of ventricular interdependence Pericardial inflammation 	<ul style="list-style-type: none"> ↑ Wall thickness Biatrial enlargement Systolic and diastolic dysfunction Pulmonary hypertension Tissue characterization abnormalities
Invasive hemodynamics	Discordant respirophasic ventricular pressure changes	Concordant respirophasic ventricular pressure changes

Teaching points

- High index of suspicion for constrictive pericarditis and restrictive cardiomyopathy in patient with predominantly right side heart failure.
- Always think of CP in patients with previous history of pericarditis, radiation and open heart surgery and presenting with signs of heart failure.
- Low voltage QRS in EKG with LVH in echocardiography: think of RC.
- Septal or lateral E' <8 excluded CP unless concomitant myocardial disease.
- Be careful in interpreting hemodynamics in patients suspecting CP with normal or low filling pressure.
- Ventricular interdependence is the hall mark of constrictive pericarditis

Thank you