

מחלות שריר הלב

והפוקרד



פרופ' אליו די סגני

מכון הלב ביה"ח שיבא תל-השומר

A young man with a flu-like disease

- בן 30 עם חום במשך 5 ימים
- היגיע לחדר מיון בגלל הופעה פתאומית של קוצר נשימה מלווה בהזעה קרה.
- באקג נמצאה ירידה של קטע ST של כ-1 מ"מ ברוב החיבורים.

A young man with a flu-like disease: diagnosis

Echocardiogram:

- LV was enlarged with poor contractility
- Mild -moderate mitral insufficiency
- ACS- acute MI?
- Pulmonary embolism?
- Mitral valve disease?
- Acute myocarditis?

CPK and Troponin I
were elevated

Acute Myocarditis

etiology

Infective

- Viral
 - Coxsackie
 - CMV
 - Influenza
 - HIV
- Bacterial
 - streptococcal
- Spirochetal
 - Lyme
- Fungal
- Parasitic
- Rickettsial

Non- infective

- Toxic
- Chemical
 - daunorubicine
- Immune
 - Giant cell
- Radiation

Acute Myocarditis

clinical manifestation

- Range from asymptomatic to fulminant heart failure
- Fatigue, dyspnea, palpitations, chest pain
- Transient murmur of MR
- ECG: from non specific ST-T changes to arrhythmias
- Laboratory
 - Myocardial markers
- Echocardiogram
 - LV enlargement
 - Diffuse or segmental contraction abnormalities
 - MR
- Endomyocardial biopsy
- Virus culture

Acute Myocarditis

natural history

- Complete recovery
- Dilated cardiomyopathy
 - Long latency
 - Progressive downhill course
- Death

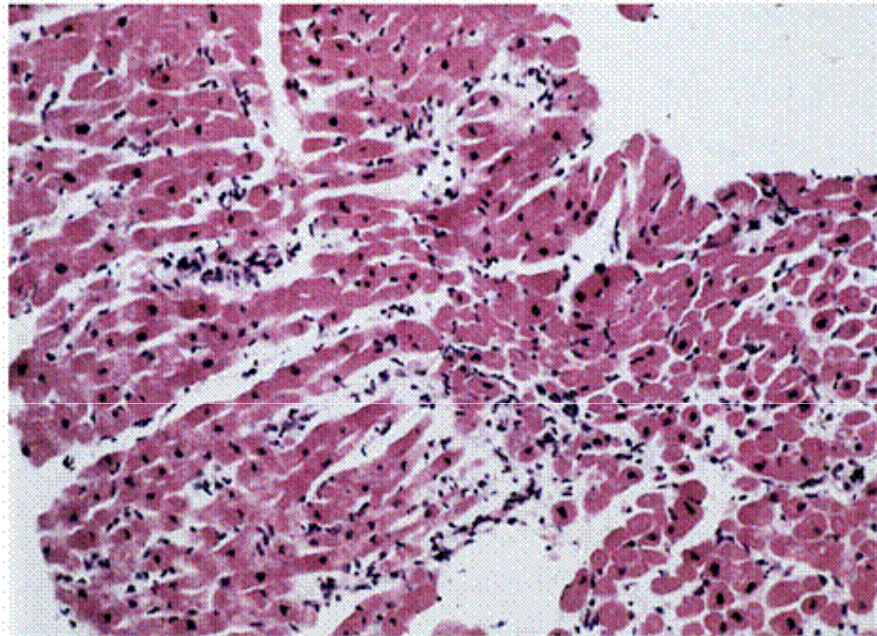


Fig. 2 Endomyocardial biopsy showing mild lymphocytic infiltration and disarrangement of myocytes (haematoxylin and eosin stain) in case 1.

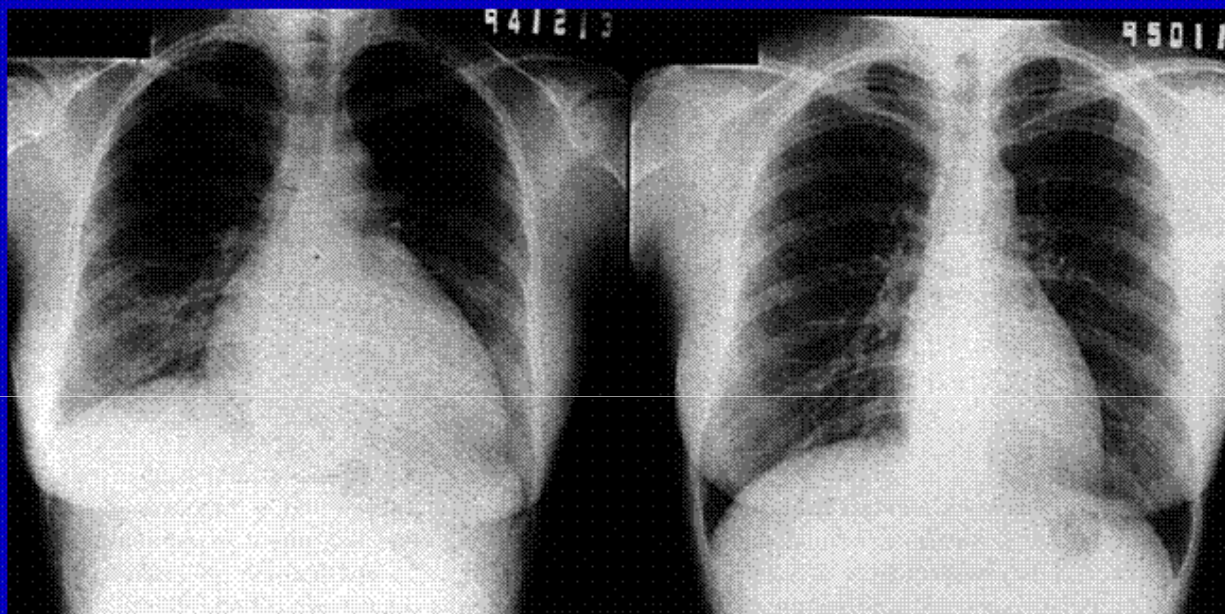
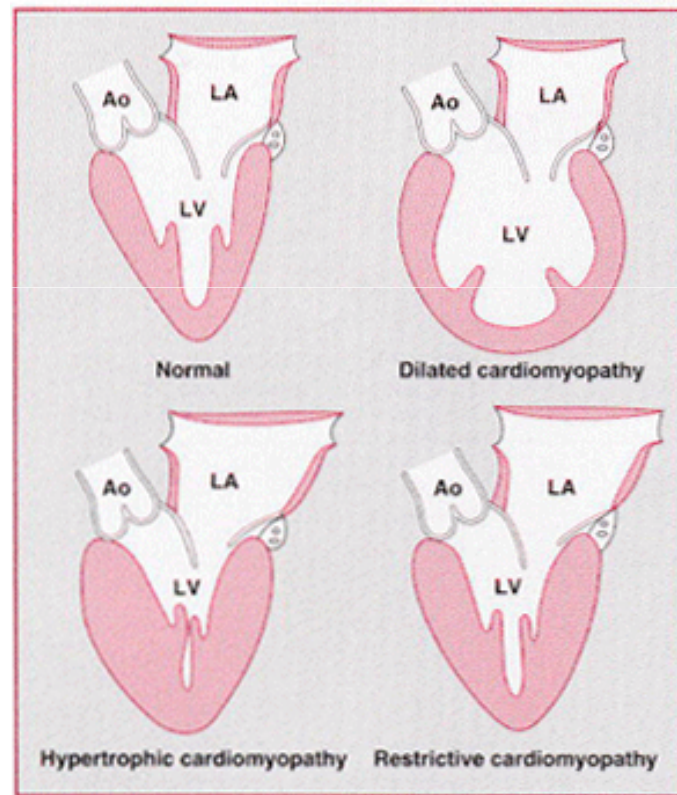


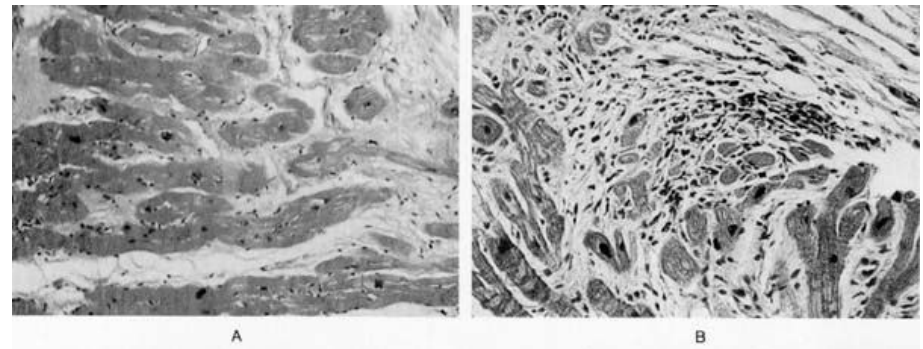
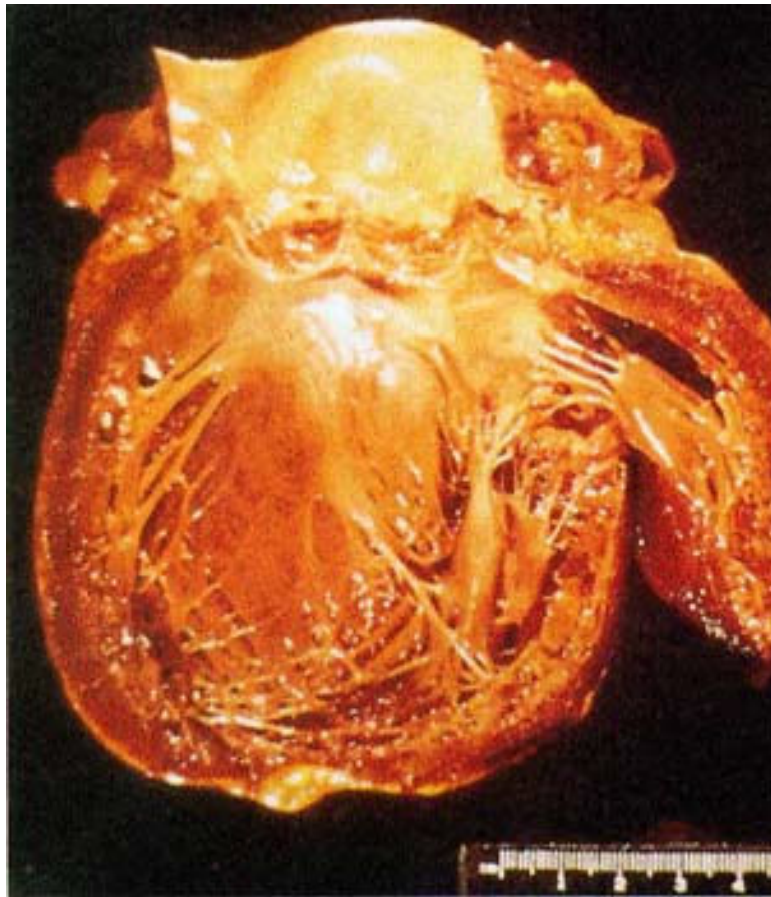
Fig. 3 Chest radiograph on admission (left) and 1 month after admission (right) in case 2.

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Cardiomyopathies classification



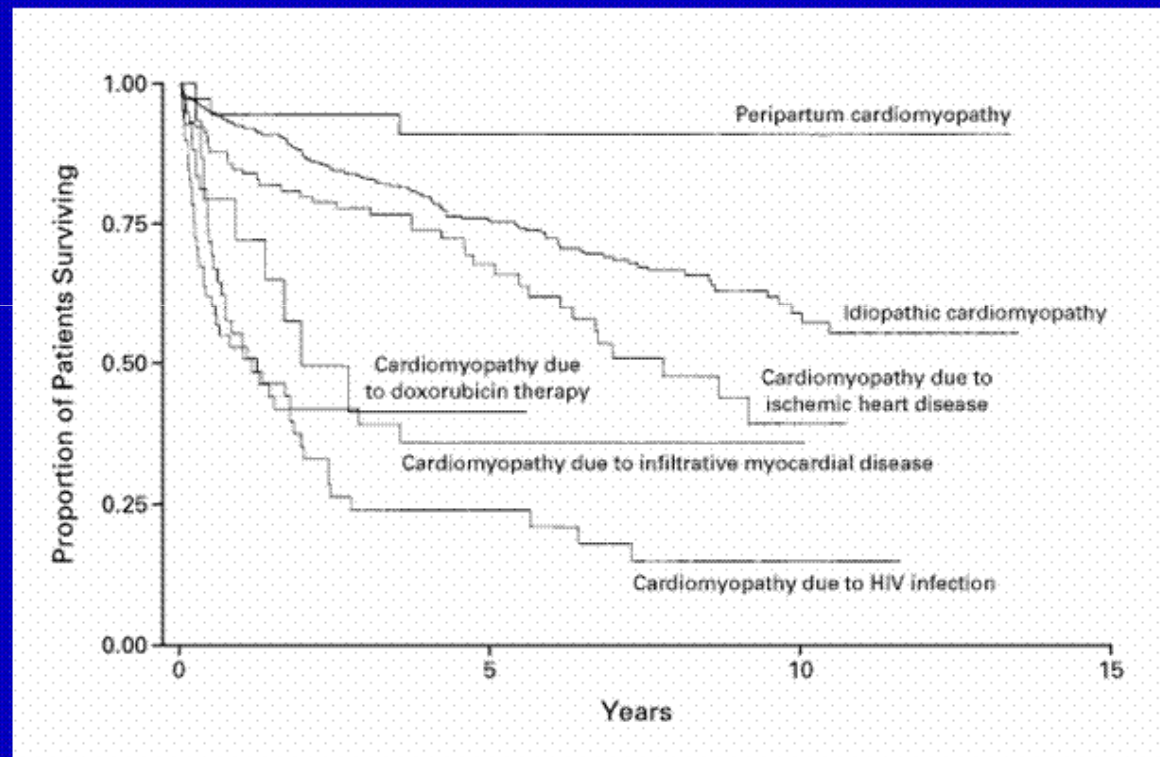
Dilated cardiomyopathy



Dilated cardiomyopathy etiology

- Familial and genetic
- Viral or cytotoxic myocarditis
 - Alcoholic
 - Pharmacological
- Immunological

LONG-TERM SURVIVAL IN PTS WITH INITIALLY UNEXPLAINED CARDIOMYOPATHY (Felker et al NEJM 2000;342:1077)

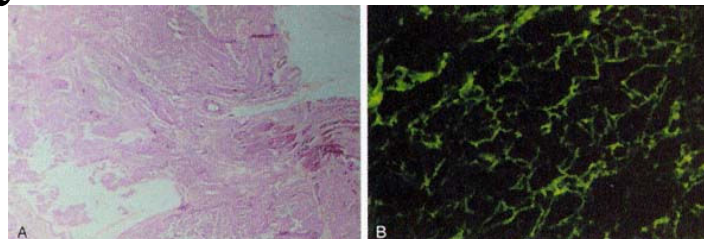
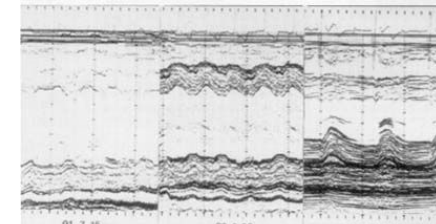
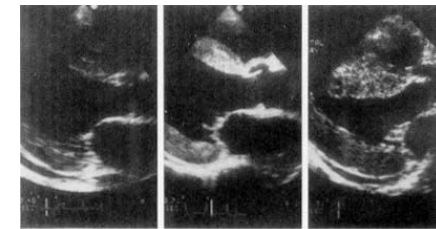
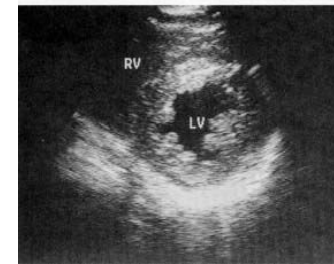
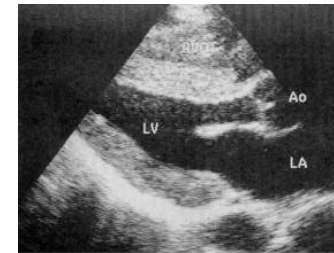


Restrictive cardiomyopathy classification

- Myocardial
 - Non infiltrative
 - scleroderma
 - Infiltrative
 - Amyloid
 - sarcoid
 - Storage disease
 - Hemochromatosis
 - Glycogen storage disease (type II-Pompe` disease)
- Endomyocardial
 - Endomyocardial fibrosis
 - Hypereosinophilic syndrome
 - Radiation

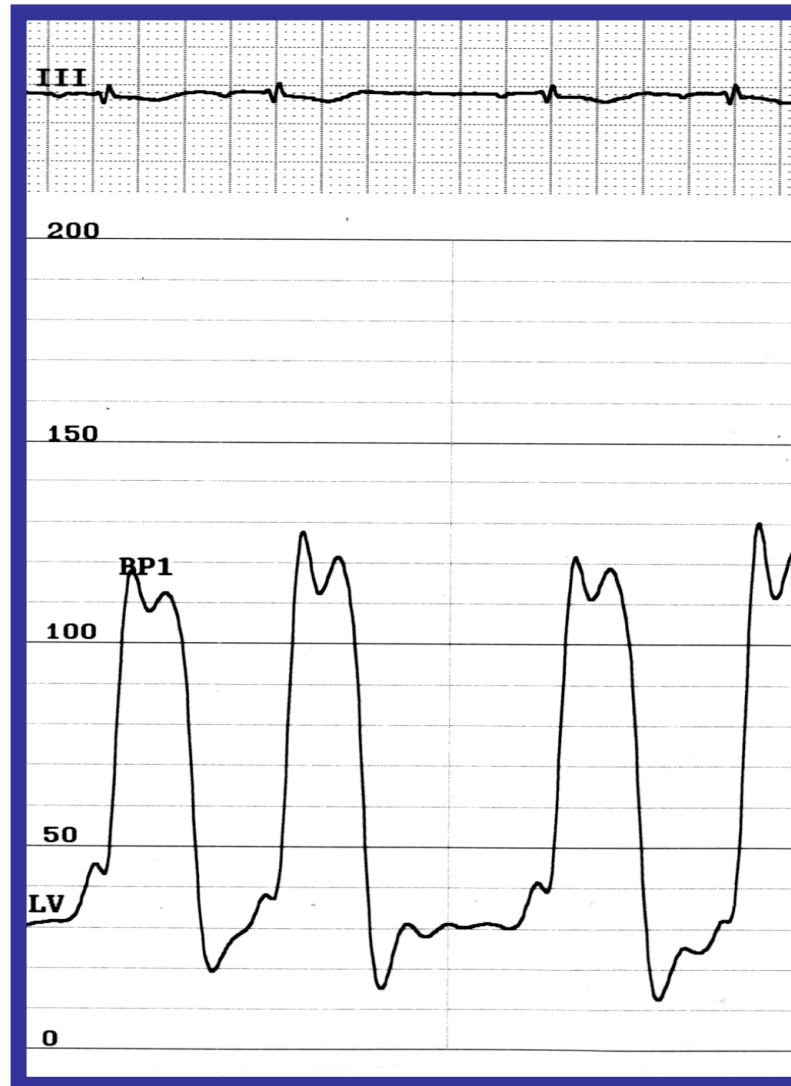
Restrictive cardiomyopathy amyloidosis

- Primary (multiple myeloma)
 - Immunoglobulin light chain AL
- Secondary
 - Non immunoglobulin AA
- Familial
 - transthyretin
- Senile

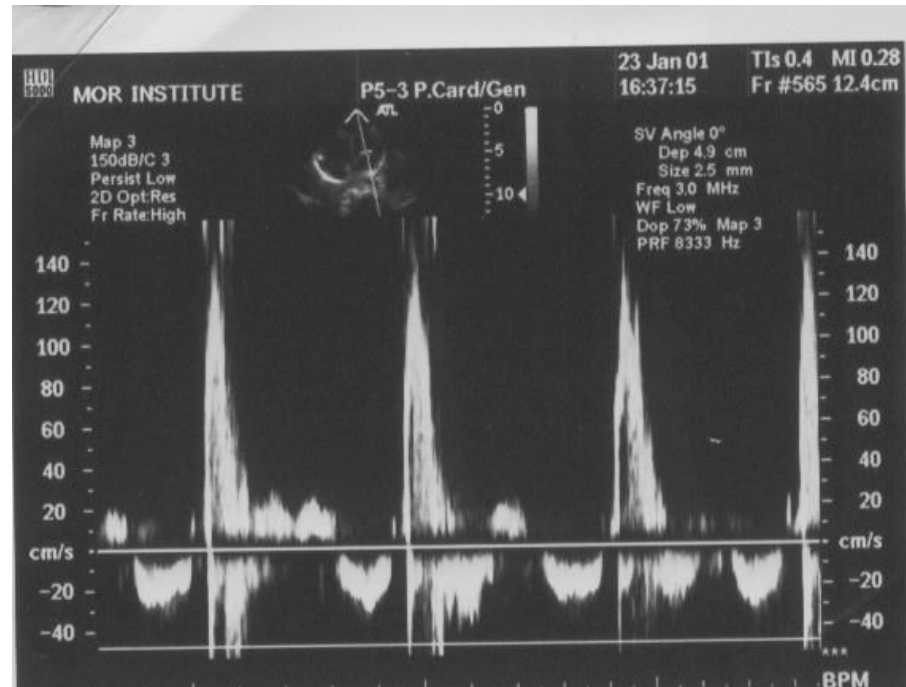
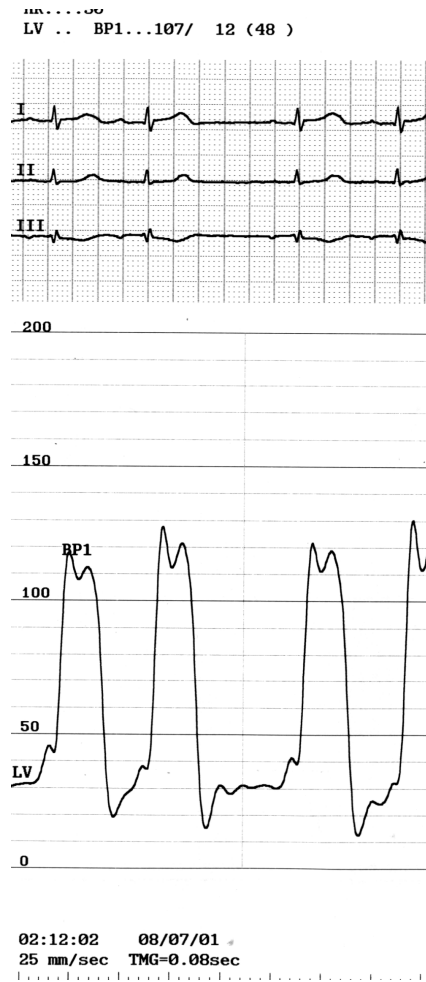


Decreased distensibility

Square root sign



Restrictive pattern

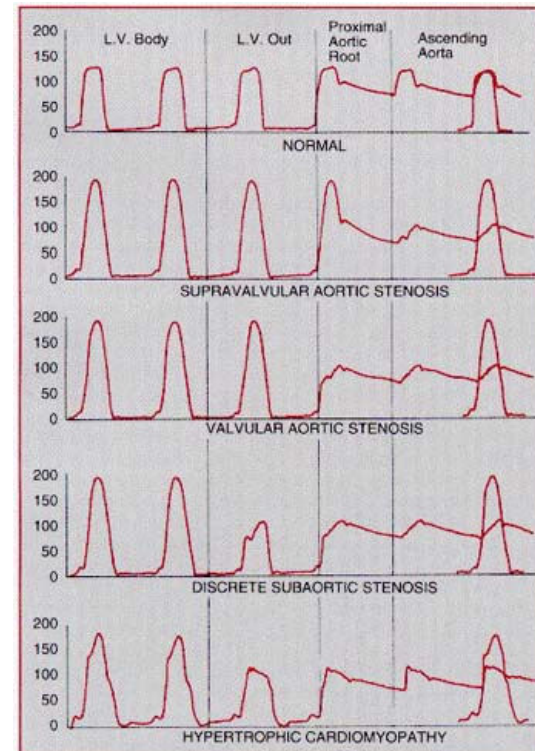
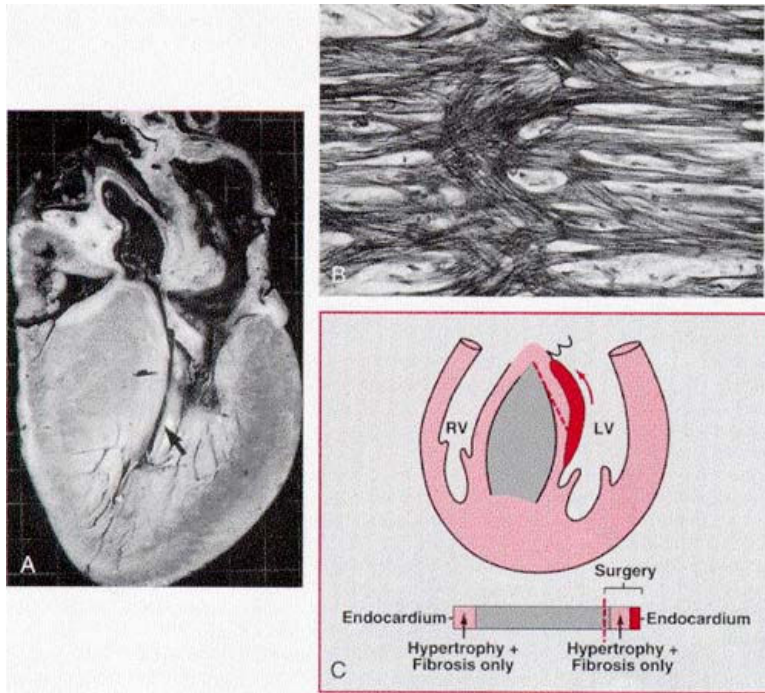


A tragedy in the soccer field

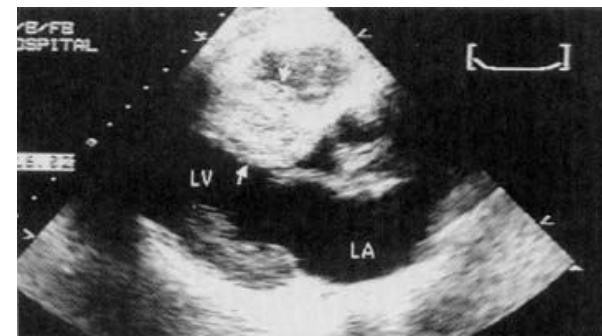
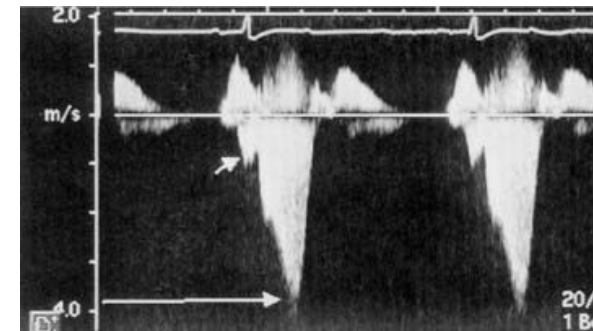
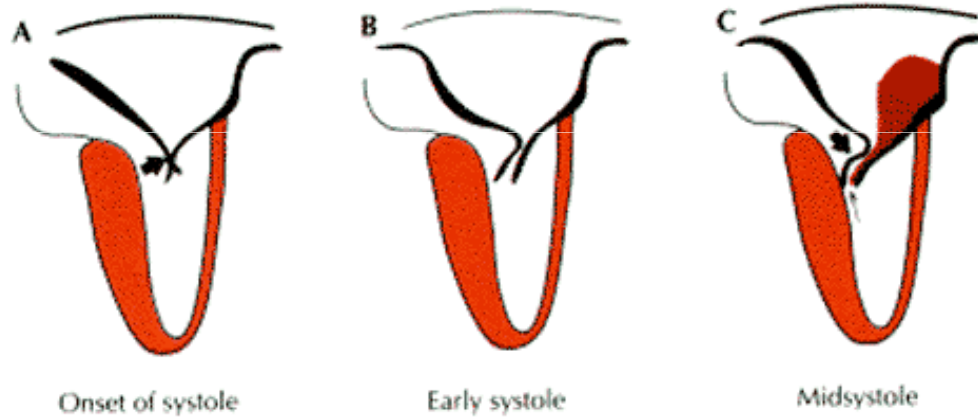
- During a soccer play a 20 year old athlete hit the ball and lost consciousness
- Resuscitation was unsuccessful
- At the autopsy...

Hypertrophic cardiomyopathy

HCM-HOCM



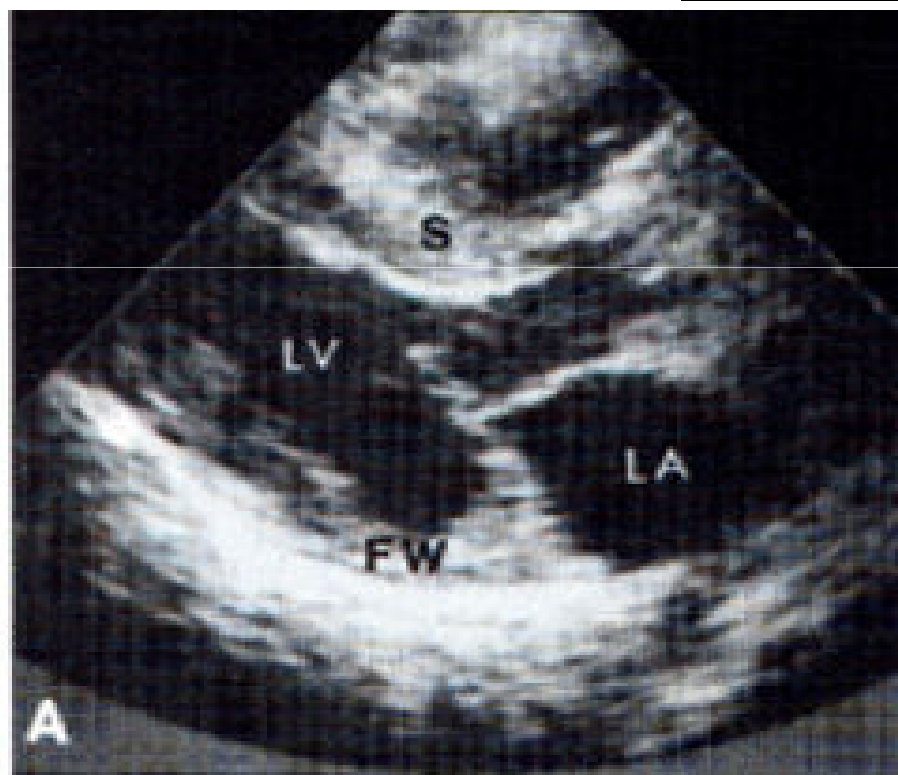
HOCM



HOCM

קרדיומיופטיה היפרטרופית

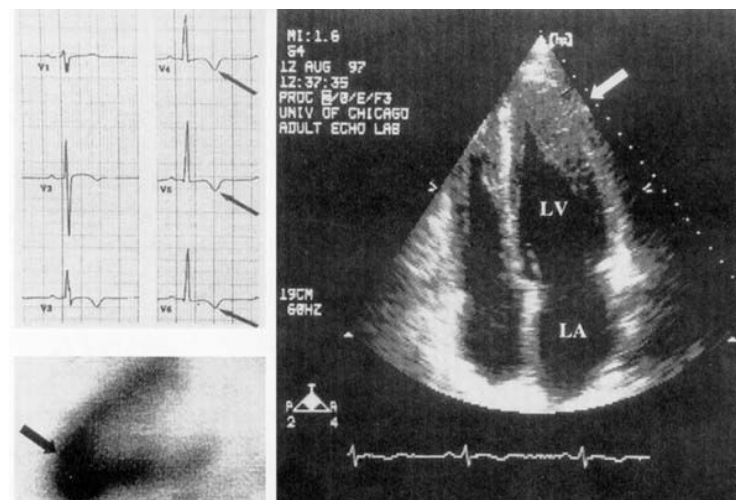
חסימתית



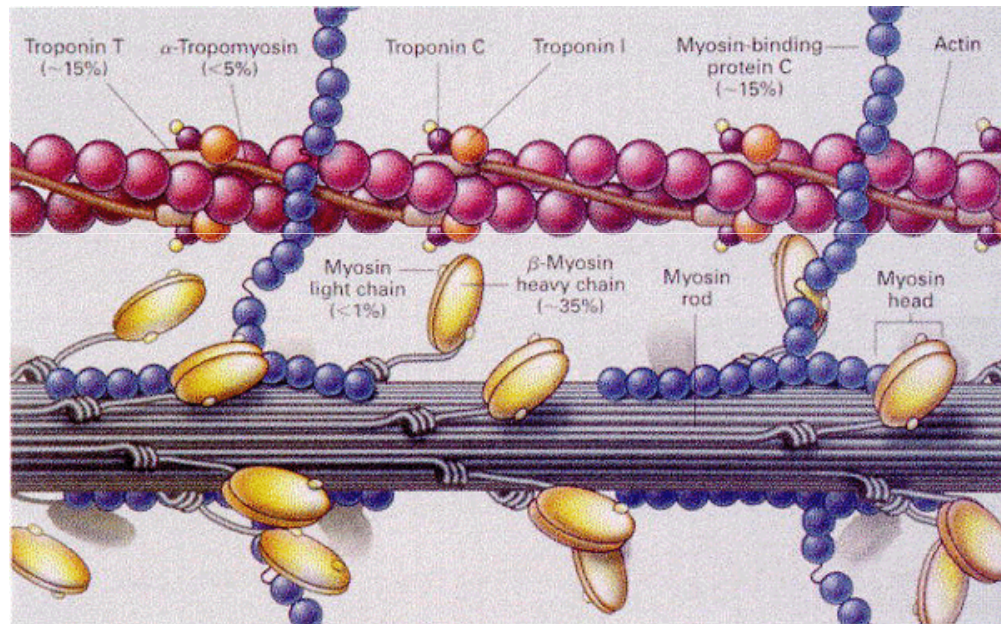
- קול רביעי

- אוושה סיסטולית פליטתית

- אין קליק



HOCM genetics



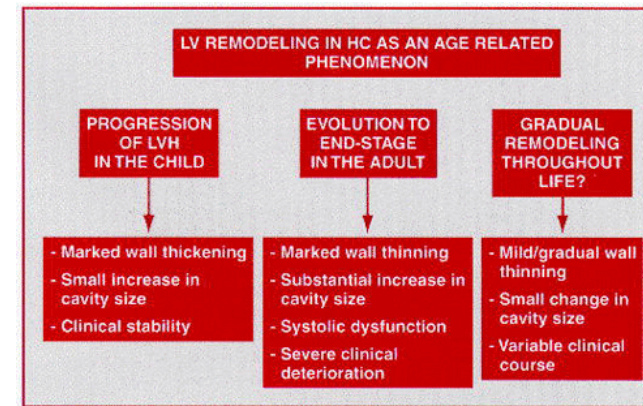
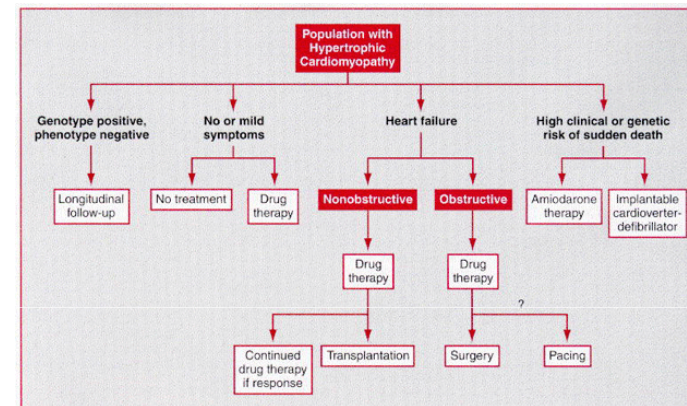
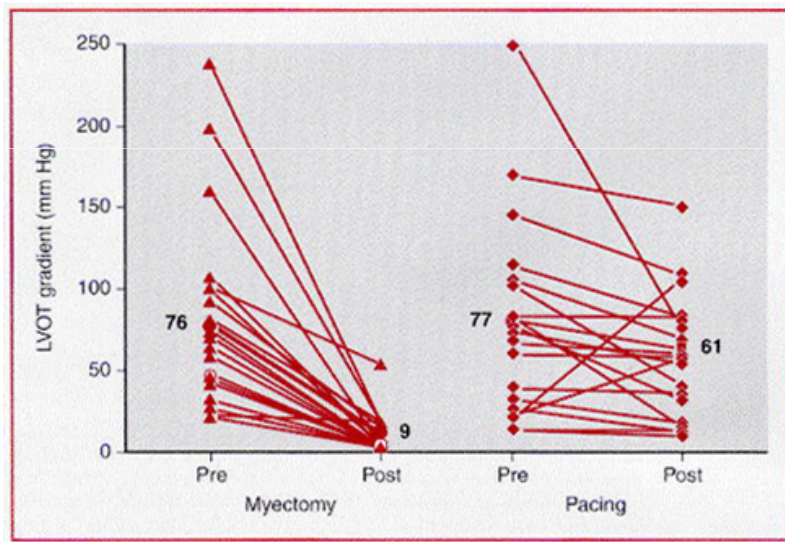
- CMH1
 - cardiomyosine heavy chain
- CMH2
 - cardiac troponin T*
- CMH3
 - α tropomyosin
- CMH4

HCM

sudden death

- Youth
- Family history
- Gene abnormalities associated with SD
- Aborted SD
- Sustained or not sustained VT
- Recurrent syncope in the young
- Bradyarrhythmia

HCM-HOCM



Pericardial disease

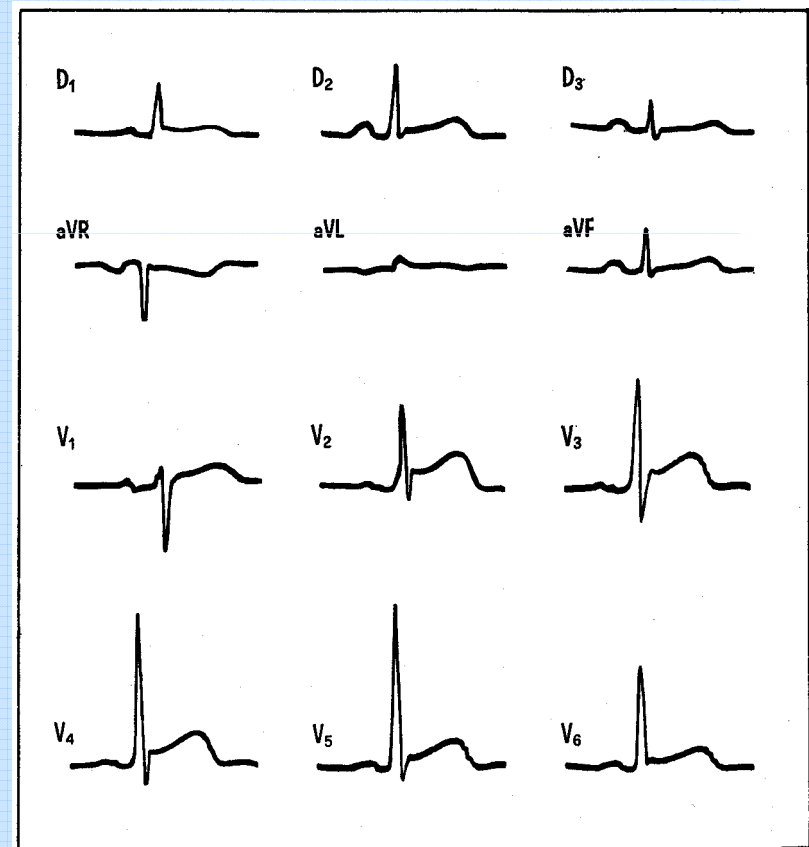
Etiologic classification

- Infectious: from TB to common virus to HIV
- Non-infectious
 - MI
 - Uremia
 - FMF
 - Malignancy
 - Post-radiation
- Hypersensitivity or autoimmunity
 - Collagen disease
 - Dressler syndrome
 - Post-pericardiotomy syndrome

Acute pericarditis - ECG diagnosis:

1. PR and ST changes
2. Time course of repolarization changes

- I
ST elevation
- II
ST Isoelectric T upright
- III
ST Isoelectric T inverted
- IV
ST Isoelectric T upright



Acute pericarditis

Echocardiographic diagnosis

- Edler 1955: first echocardiographic demonstration of pericardial effusion
- No specific echo signs of acute pericarditis
- Possibility of regional wall motion abnormalities
- The value of a negative study in excluding acute MI

Compressive pericardial syndromes

- Pericardial tamponade
- Constrictive pericarditis
- Elastic form of constrictive pericarditis (effusive constrictive)

Pericardial tamponade

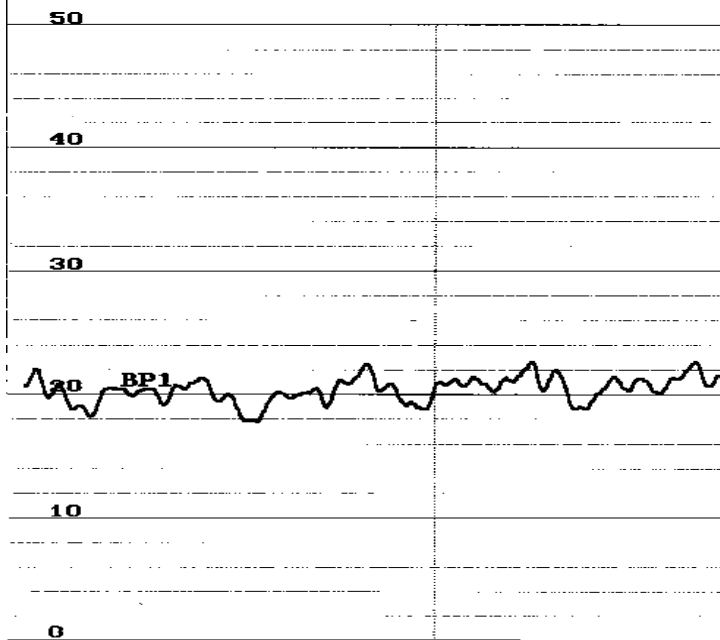
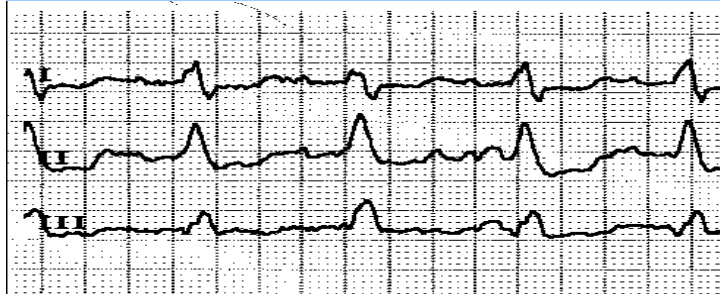
- Accumulation of pericardial fluid causing:
 - Compression of the heart
 - Impaired ventricular diastolic filling
 - Reduction of stroke volume and cardiac output

Pericardial tamponade, a problem of definition: all or none phenomenon or hemodynamic spectrum?

- Hemodynamic tamponade
- Echocardiographic tamponade
- Clinical tamponade

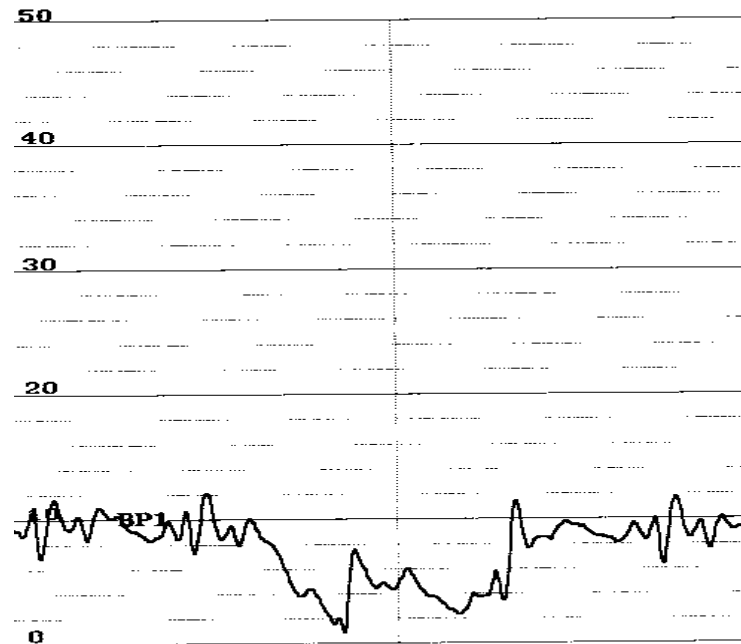
Intra-pericardial pressure

tamponade



12:45:06 09/13/01
25 mm/sec TMG=0.08sec

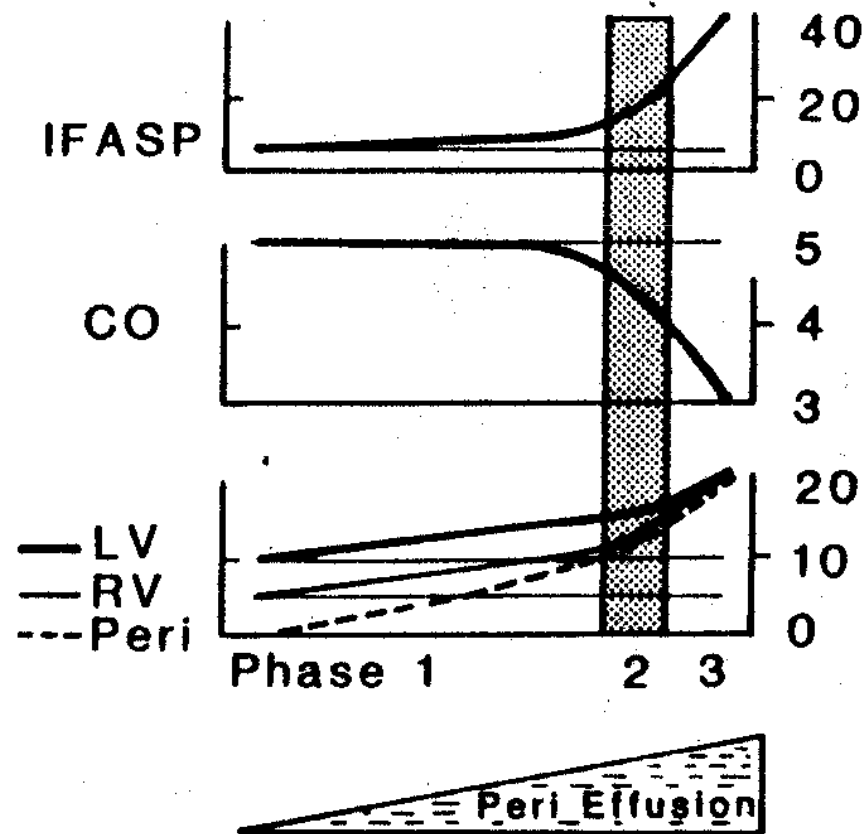
post pericardiocentesis



13:03:56 09/13/01
25 mm/sec TMG=0.08sec

Spectrum of hemodynamic changes in cardiac tamponade

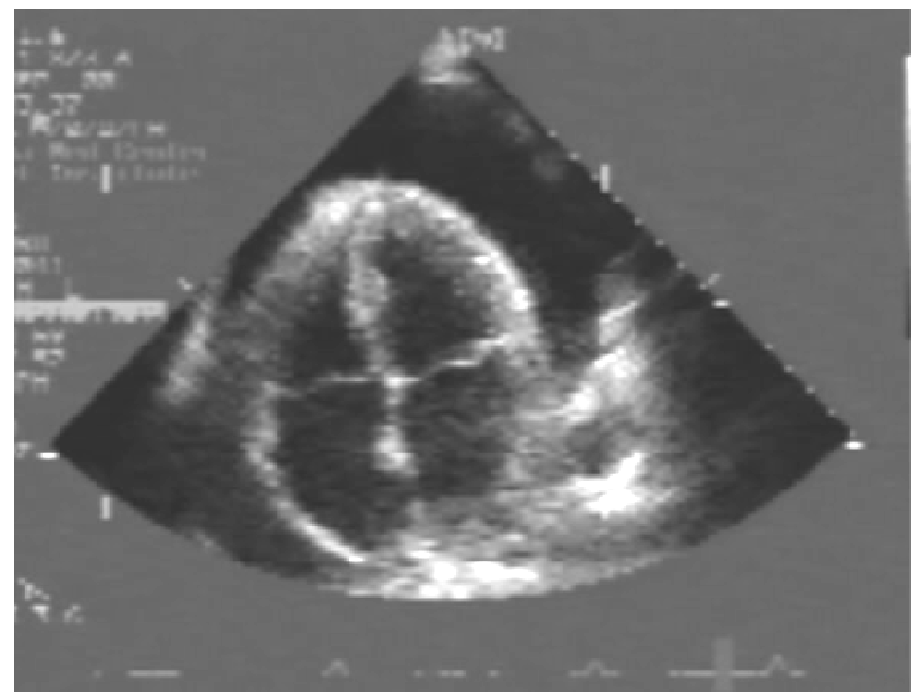
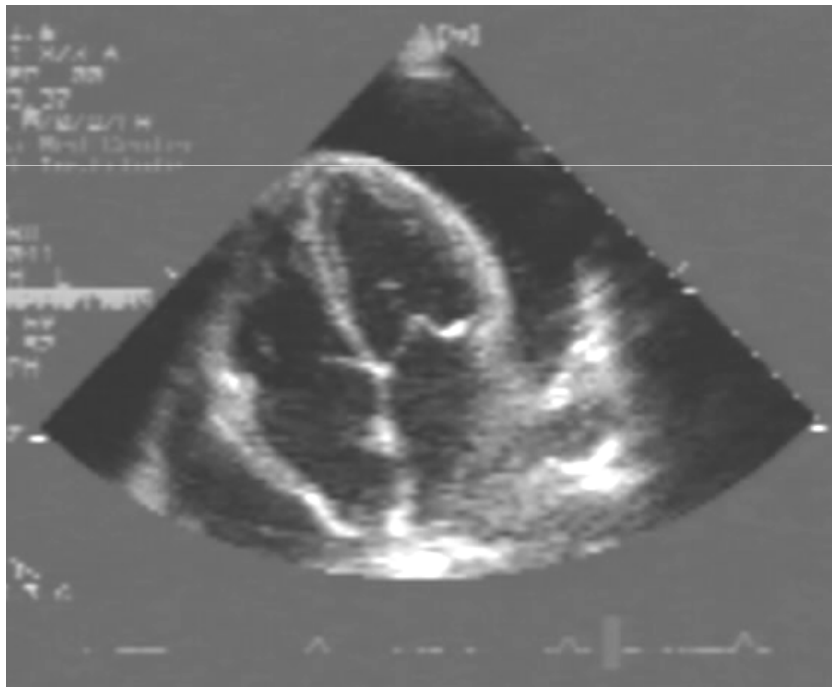
Reddy PS et al. Am J Cardiol 1990;66:1487-91



Pericardial tamponade, a problem of definition: all or none phenomenon or hemodynamic spectrum?

- Hemodynamic tamponade
- Echocardiographic tamponade
- Clinical tamponade

Echocardiography in cardiac tamponade:
Atrial diastolic collapse and
atrio-ventricular interdependance

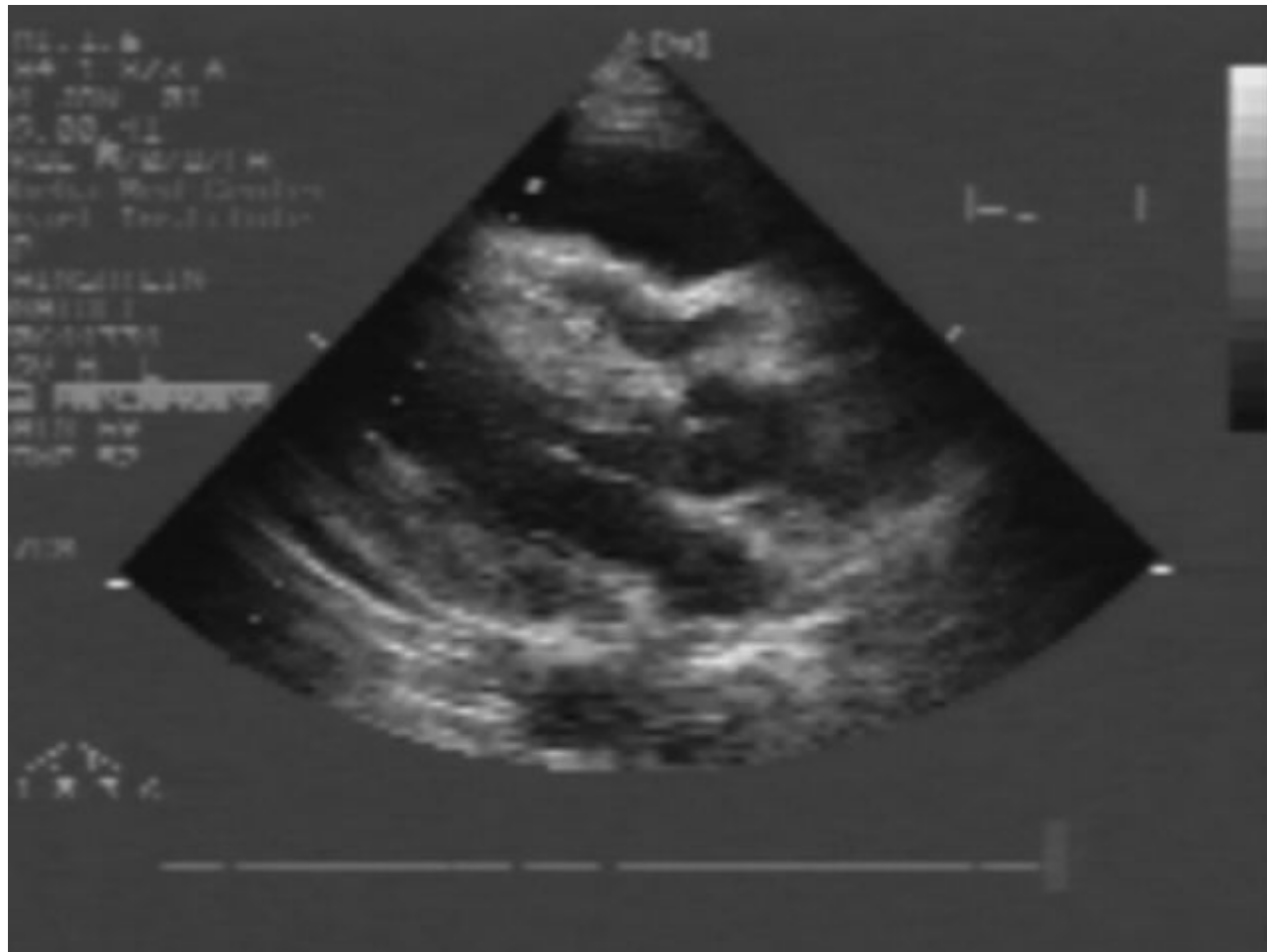


Atrio-ventricular interdependance



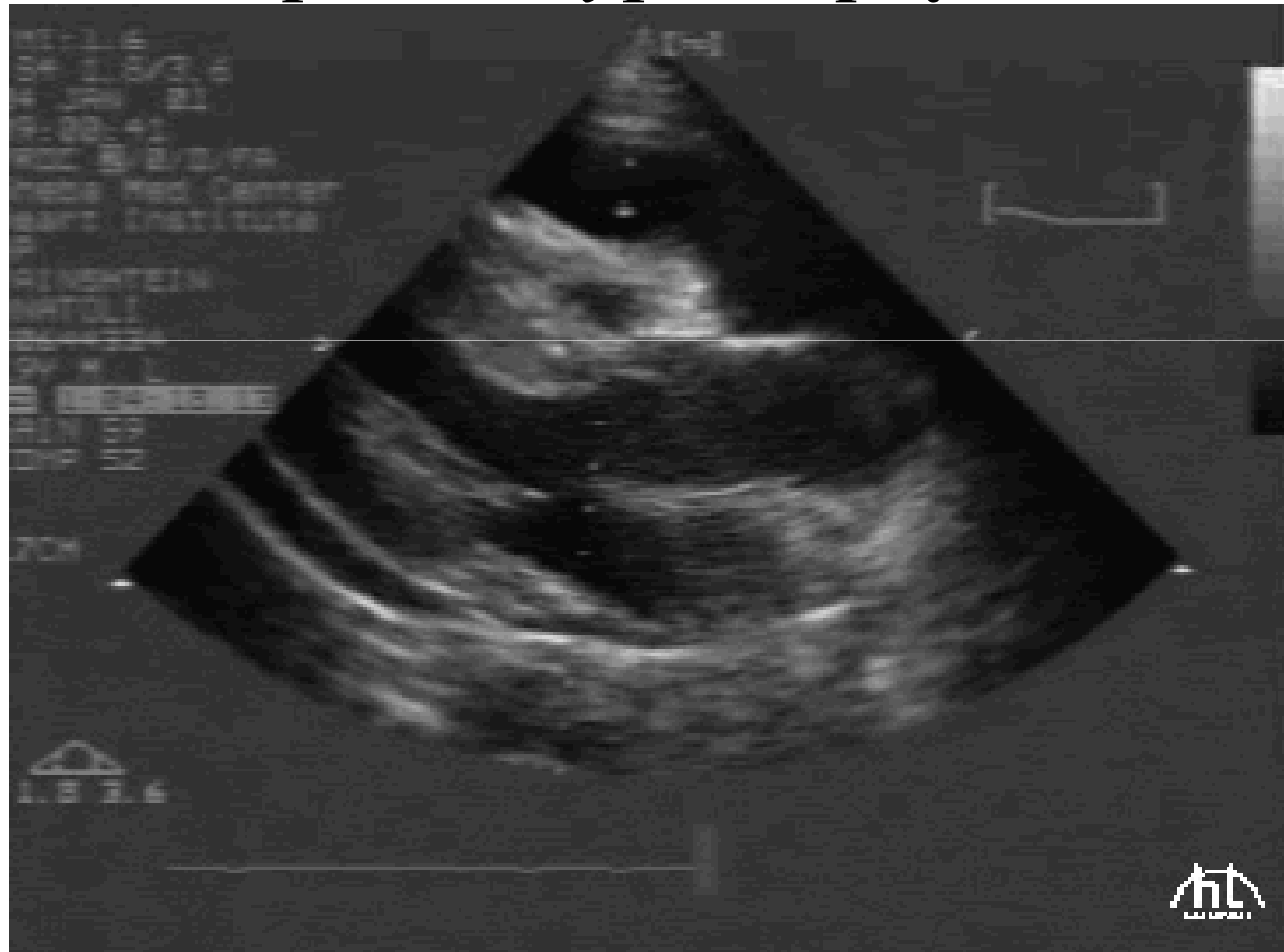
Echocardiography in cardiac tamponade

Right ventricular diastolic collapse and pseudohypertrophy

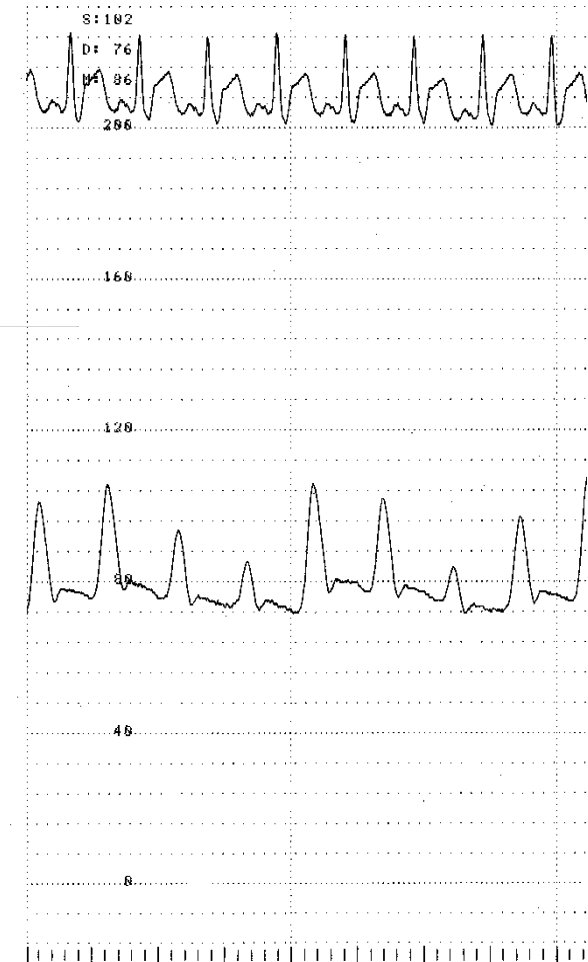
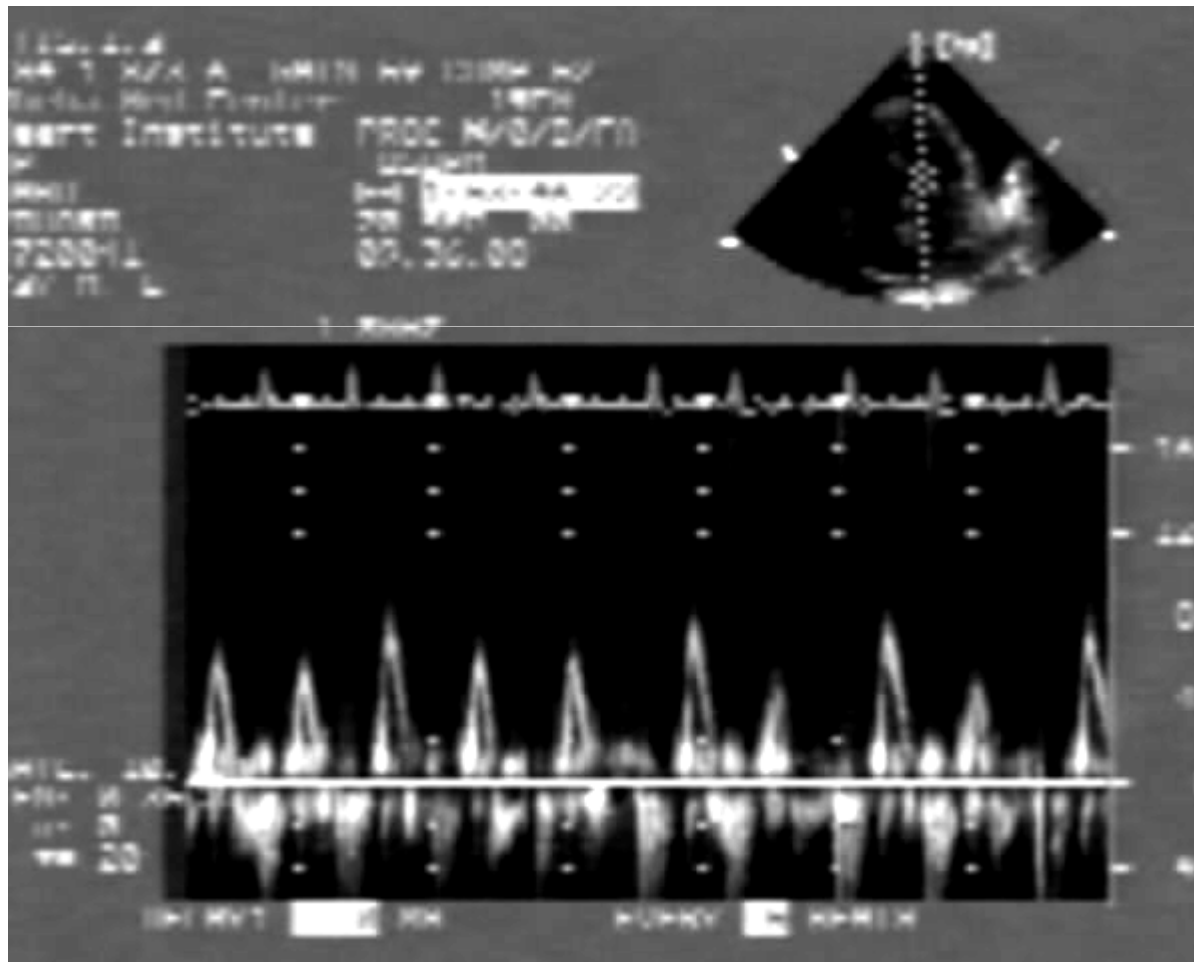


Echocardiography in cardiac tamponade

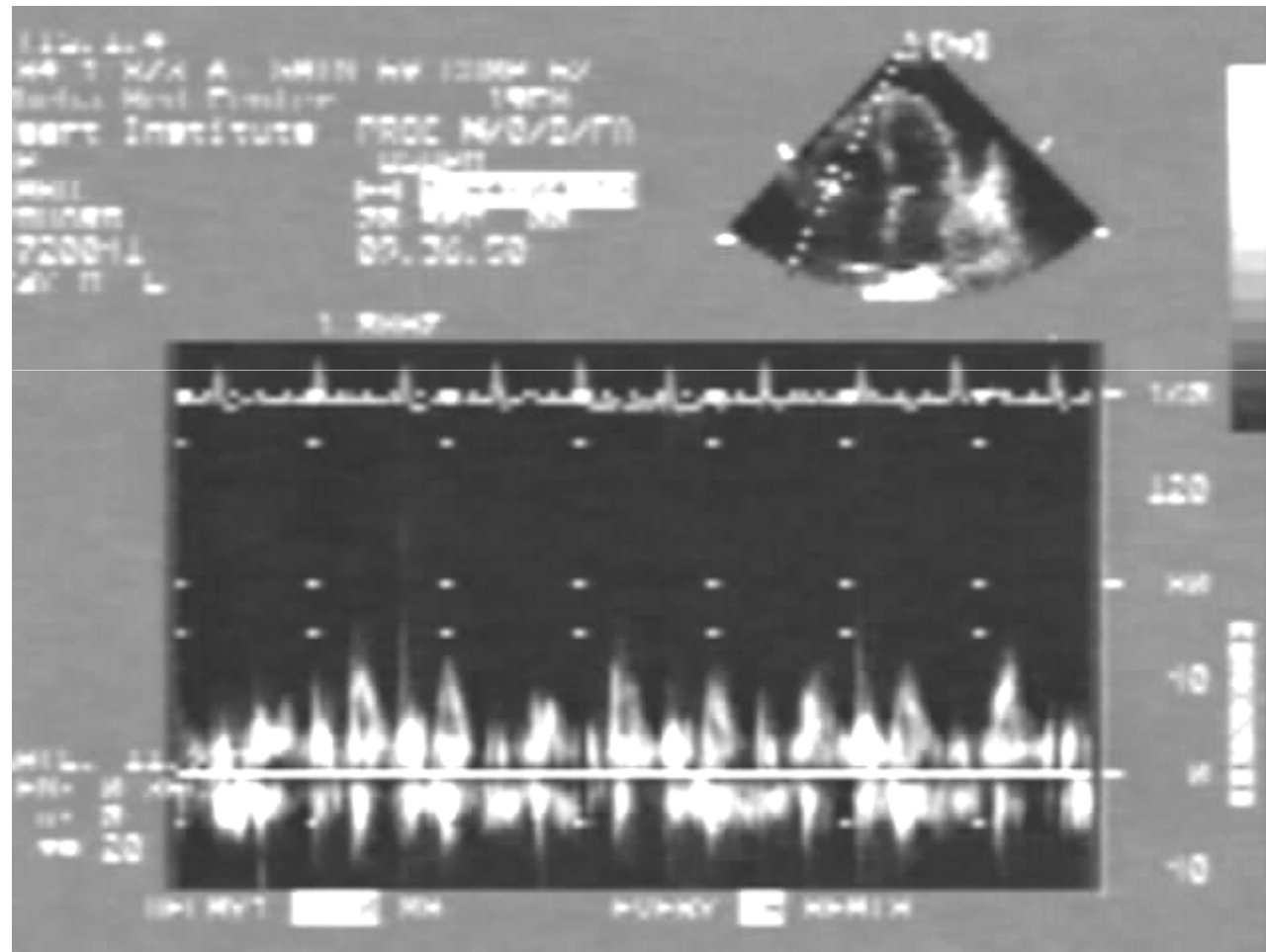
Right ventricular diastolic collapse and pseudohypertrophy



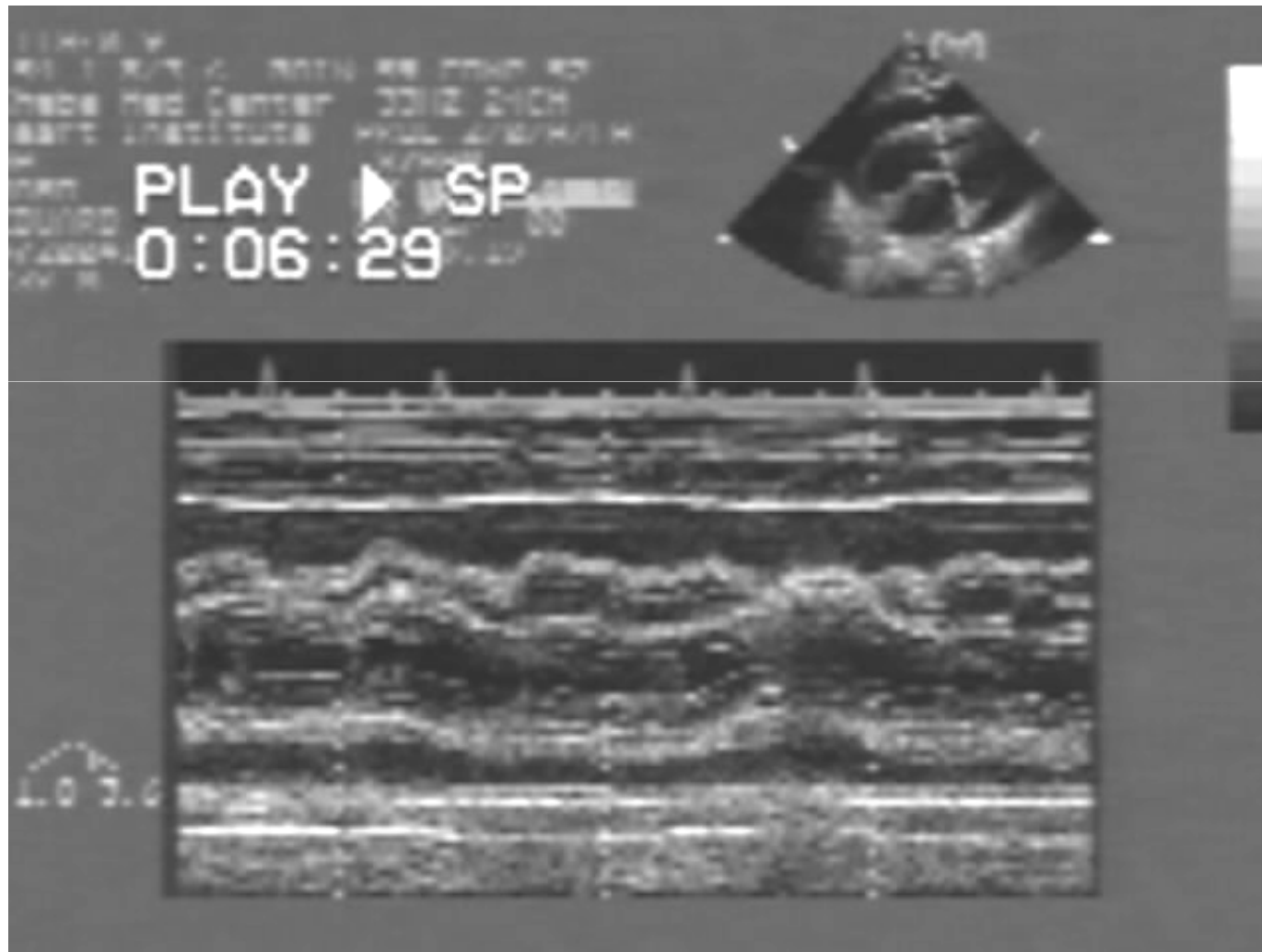
Respiratory variation in mitral Doppler flow



Tricuspid respiratory changes in cardiac tamponade



Ventricular inter-dependance



Pericardial tamponade, a problem of definition: all or none phenomenon or hemodynamic spectrum?

- Hemodynamic tamponade
- Echocardiographic tamponade
- Clinical tamponade

Clinical tamponade:

Acute tamponade

- Iatrogenic
 - Catheterization diagnostic and therapeutic: 9.6% of all pericardiocentesis
 - Pacemaker
- Trauma
- Dissecting aneurysm

- Beck's triad:
 - Increased venous pressure
 - Hypotension
 - A quiet heart
- Tachycardia
- Tachypnea
- Stupor-agitation
- Orthopnea

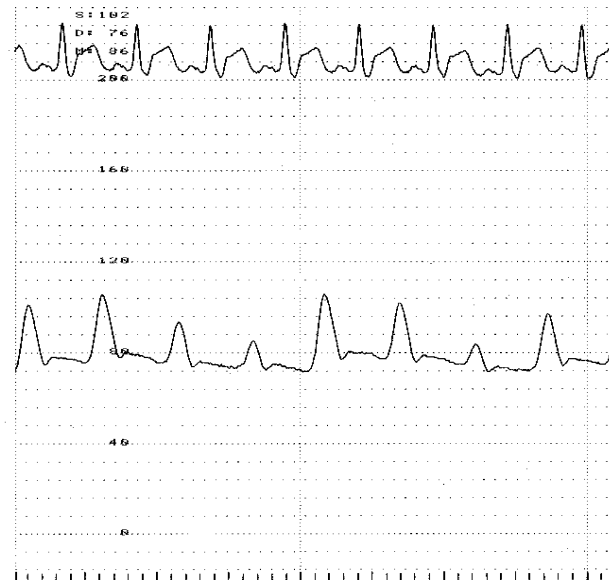
Clinical tamponade: subacute and chronic cases

- Compensated

- High venous pressure
- Pulsus paradoxus
- **Normal or high BP**

- De-compensated

- High venous pressure
- Pulsus paradoxus
- **Low blood pressure**



Elevated arterial blood pressure in cardiac tamponade

Brown J et al N Engl J Med 199;327:463-6

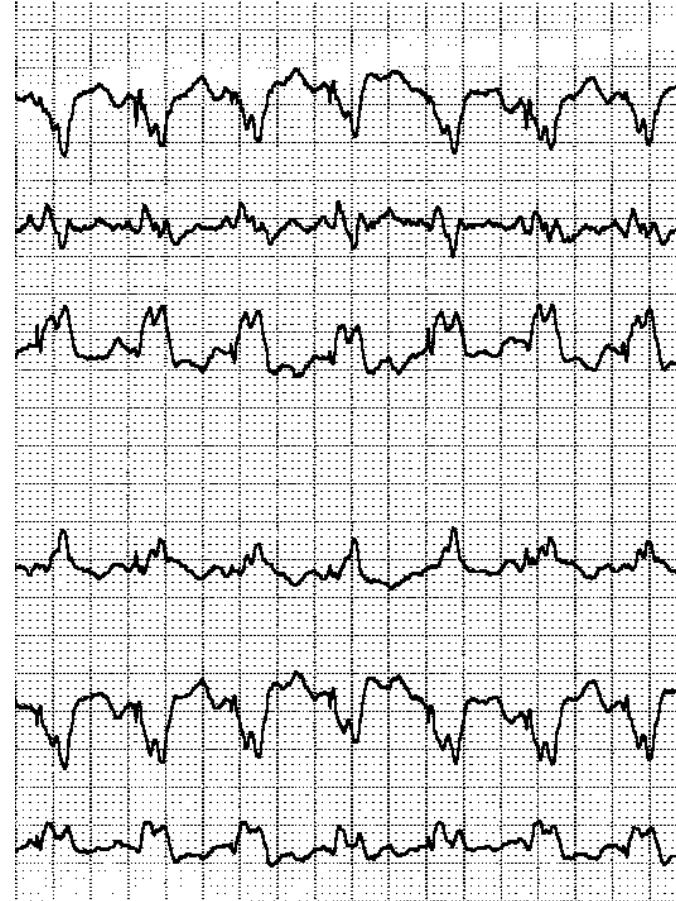
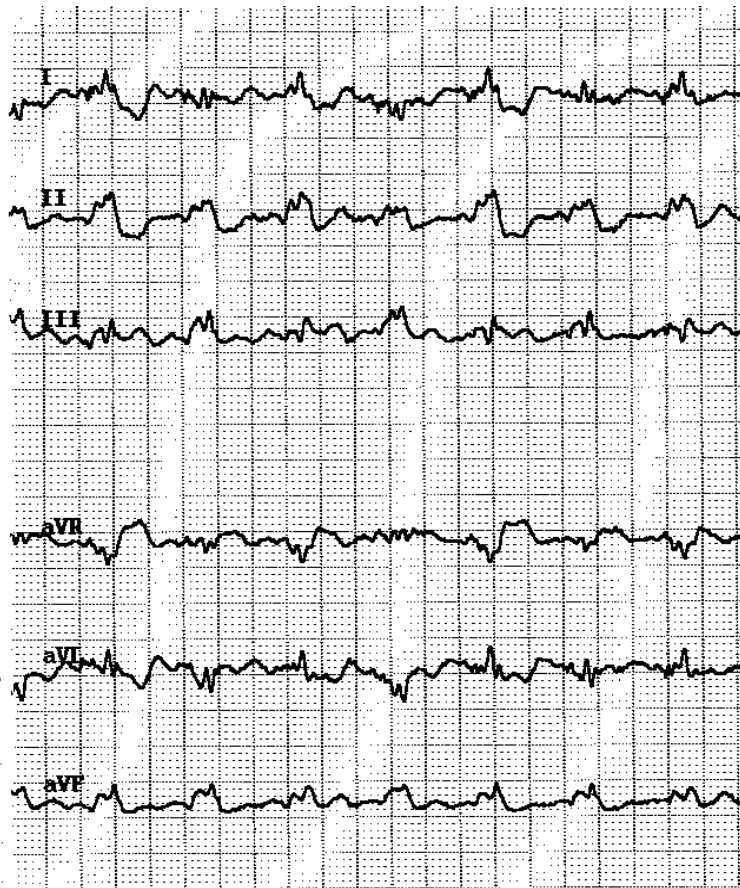
- 18 consecutive patients with cardiac tamponade
- All with equal right atrial and pericardial pressure
- In 6 patients with an history of hypertension BP ranged from 150/100 to 210/130
- After pericardial tap BP decreased from 176/113 to 139/83 and CO increased
- In 12 patients with normal or low BP, pericardial tap increased BP and CO

Treatment of cardiac tamponade

- When
- How
 - Pericardiocentesis
 - Percutaneous balloon pericardiostomy
 - Thoracoscopic pericardial window
 - Surgical window

Cardiac tamponade

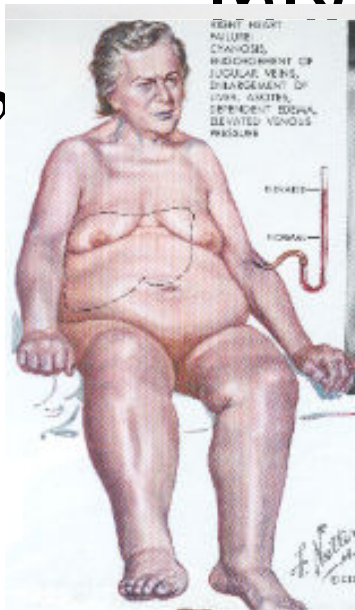
effect of pericardiocentesis on ECG



A puzzling right heart failure

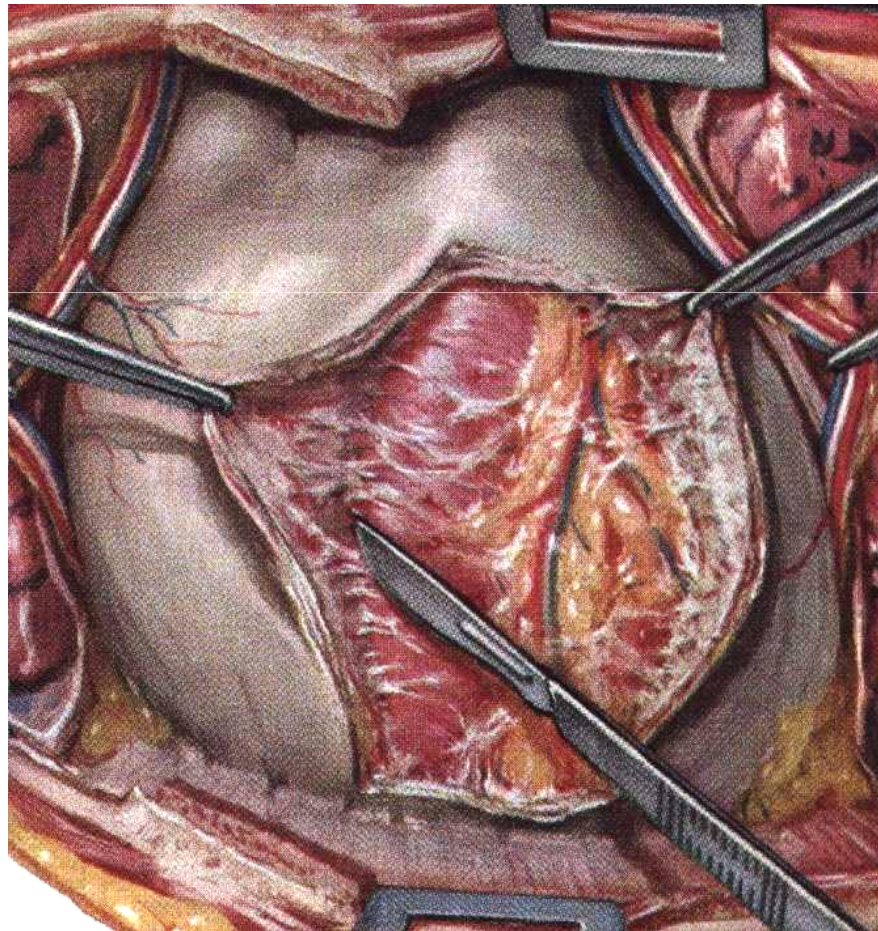
- חולה בת 50 סובלת מזה שנה מאיספיקת לב ימנית

- בבדיקת הלב לא היו ממצעים שהתאימו למחלת של המסתמים או למחלה של לי



- מה האבחנות האפשריות?
- איזה בדיקות עזר מומלצות?

Constrictive pericarditis

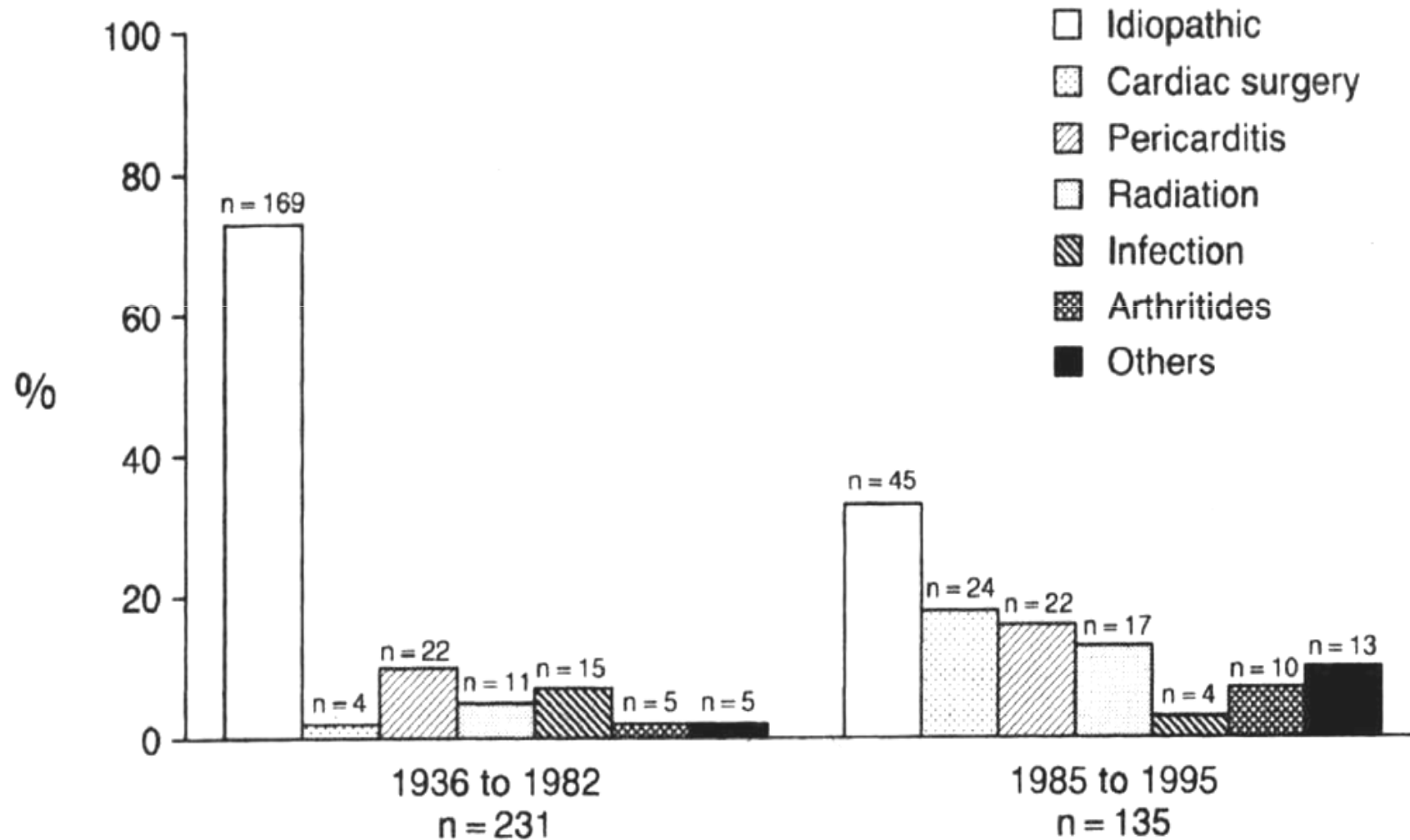


Constrictive pericarditis

- Compression of the heart
- Impaired diastolic filling
- *Increased filling pressure*

Constrictive pericarditis at Mayo Clinic

Ling et al. *Circulation* 1999



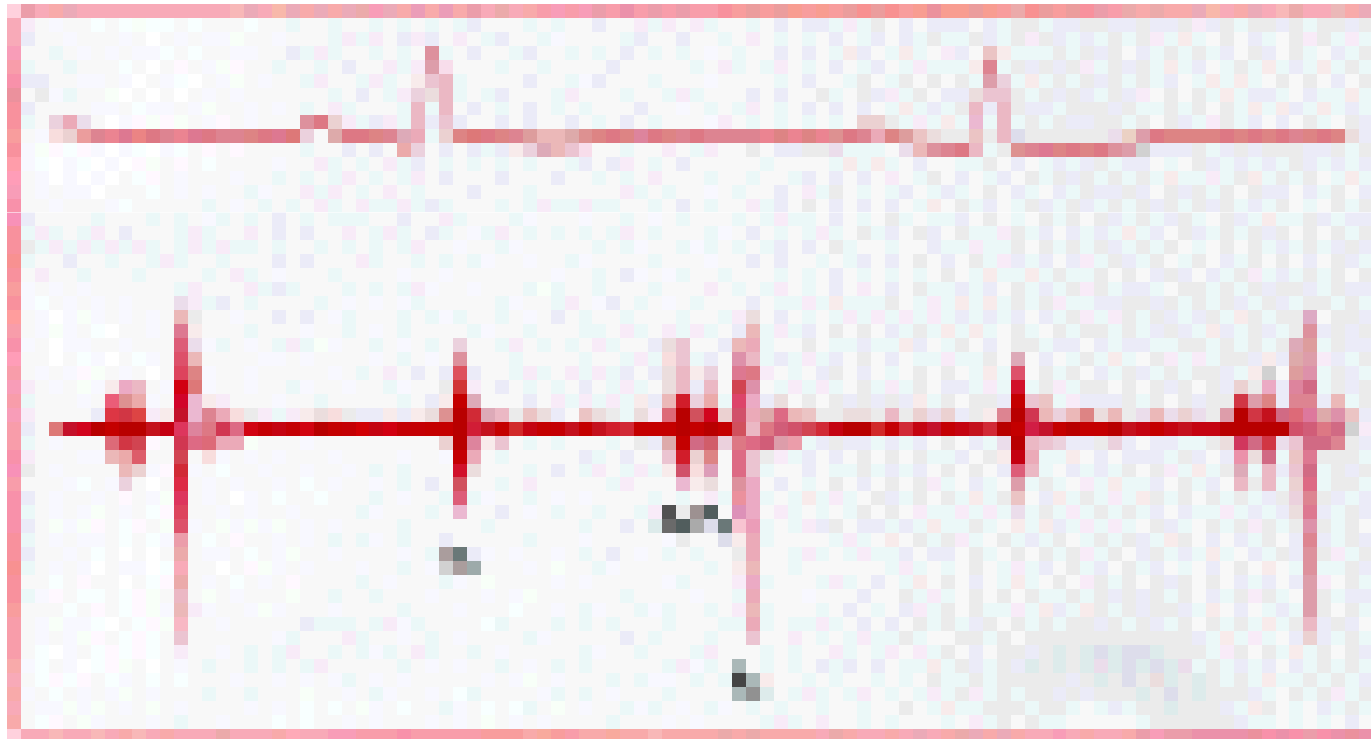
Constrictive pericarditis: diagnosis

- Clinical signs of right heart failure
- Chest X ray
- CT
- MRI
- Echo-Doppler
- Cardiac catheterization

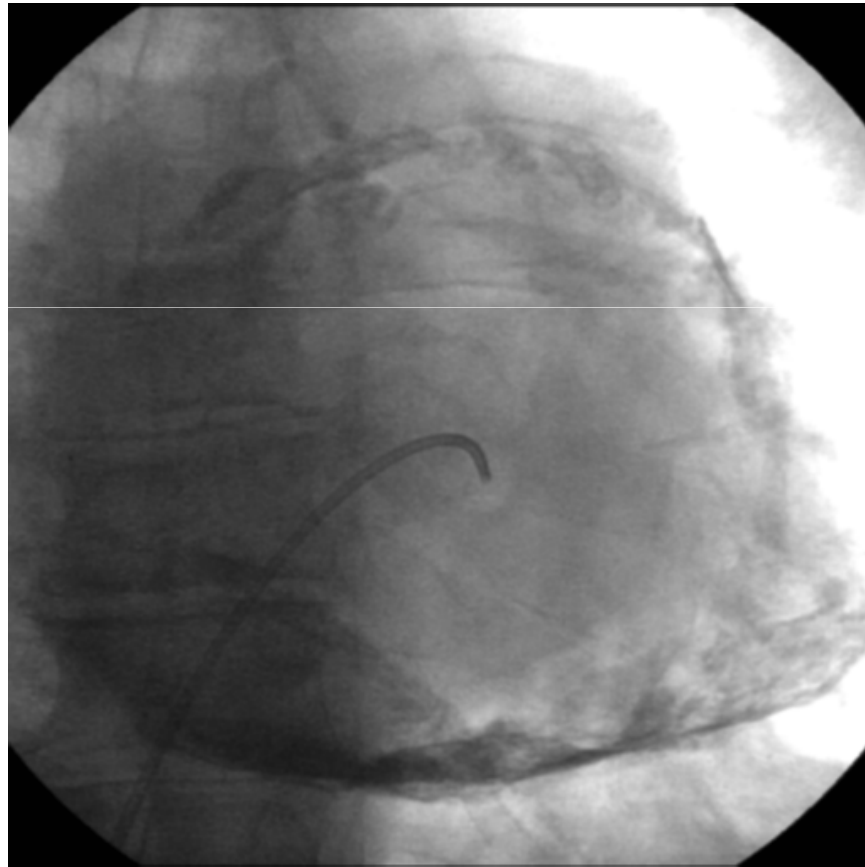
Kussmaul's sign



Pericardial honk in constrictive pericarditis

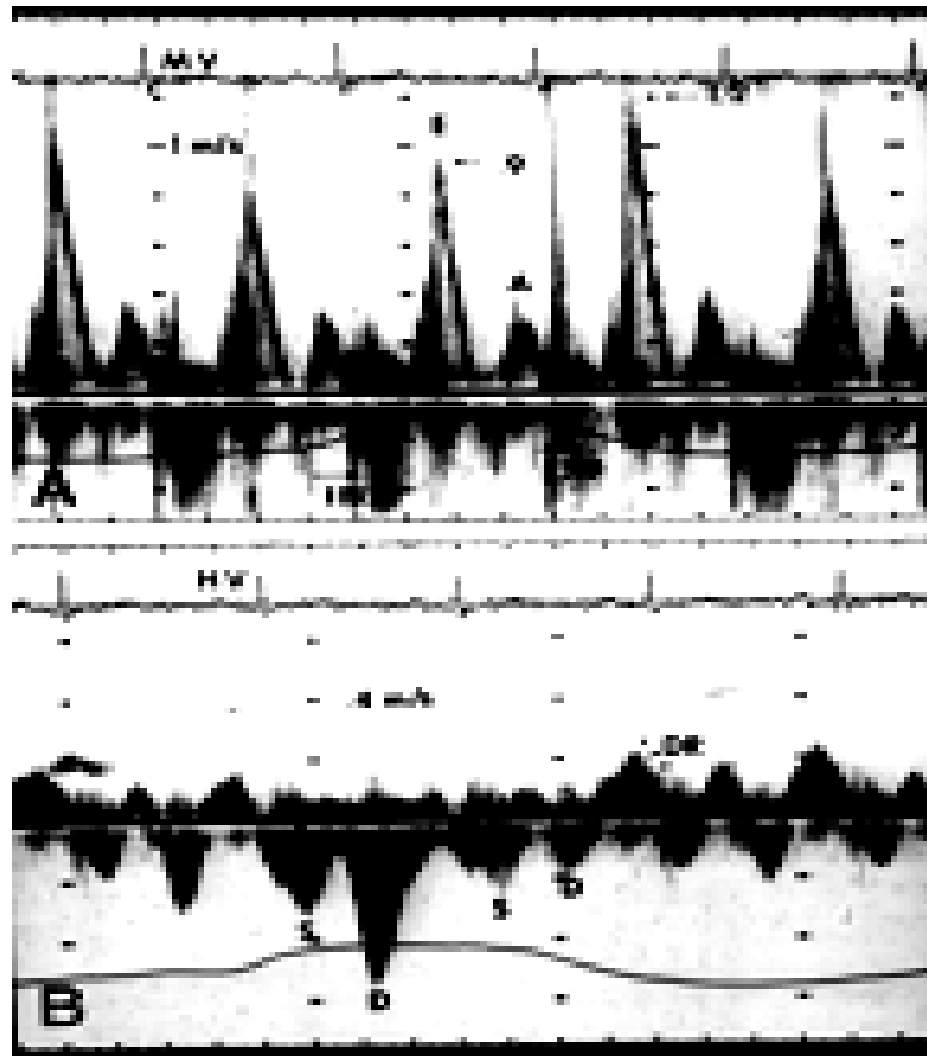


Calcified pericardium in constrictive pericarditis

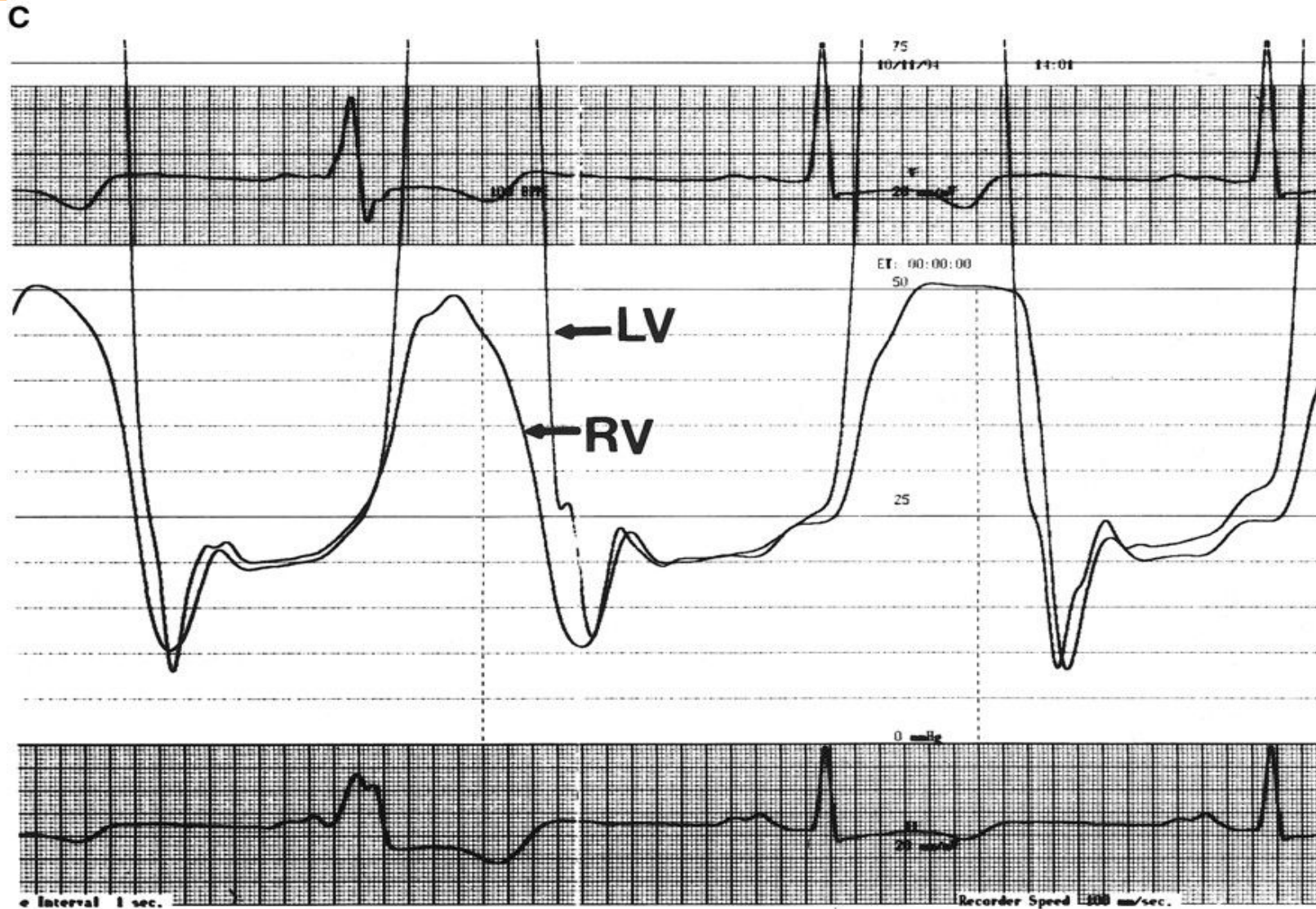


Constrictive pericarditis

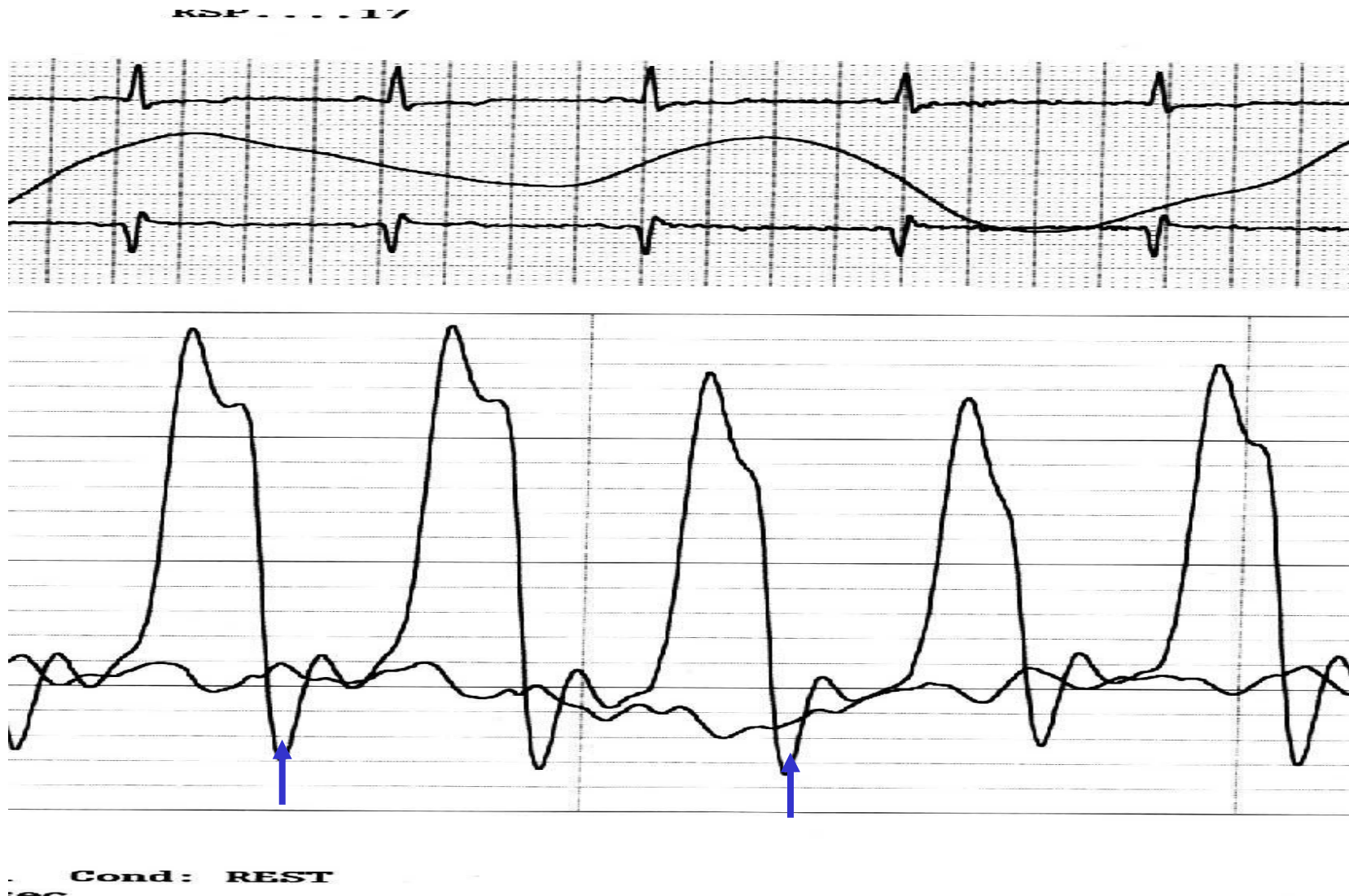
Pulsed Doppler



Constrictive pericarditis classic signs

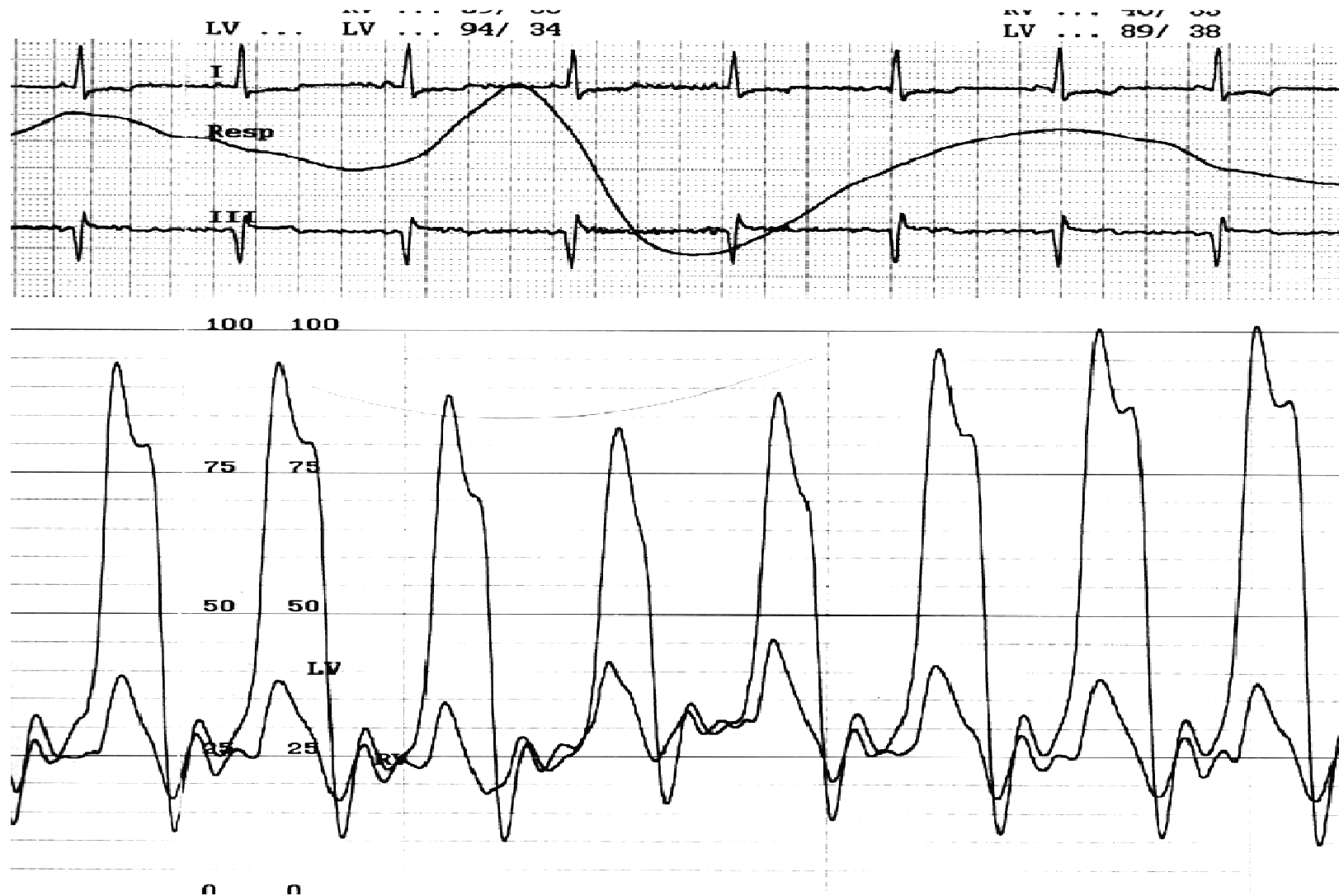


Dissociation of intrathoracic and intracardiac pressures



Constrictive pericarditis

Ventricular interdependence



Constrictive pericarditis

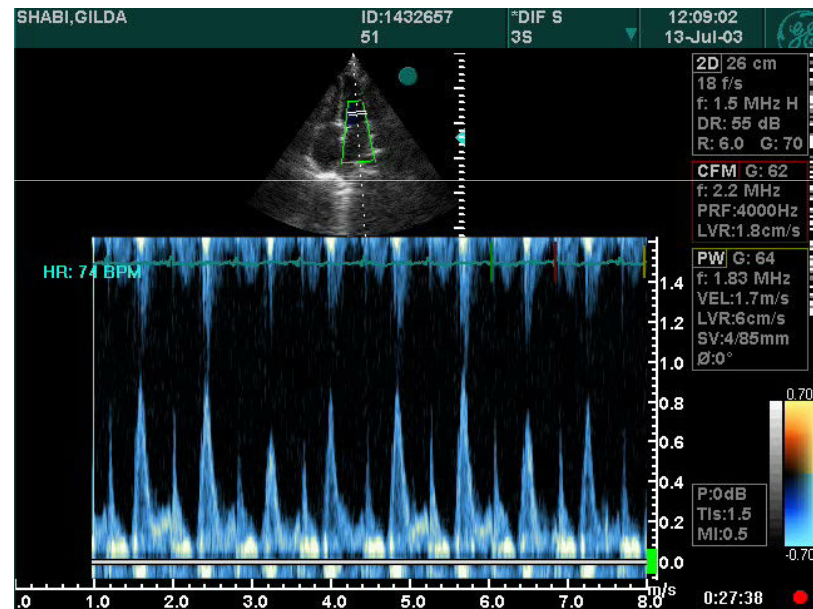
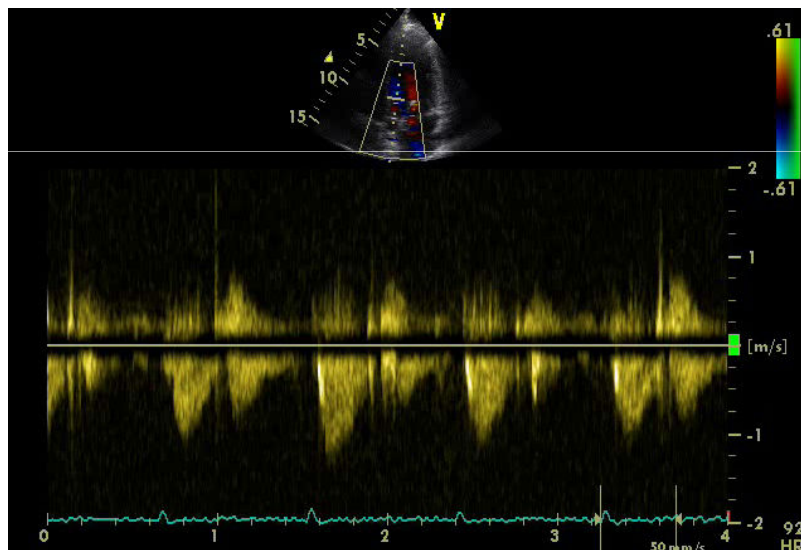
Dynamic respiratory changes

Criteria	Sensitivity	Specificity	PPV
• PCWP/LV	93	81	78
• LV/RV interdep.	100	95	94

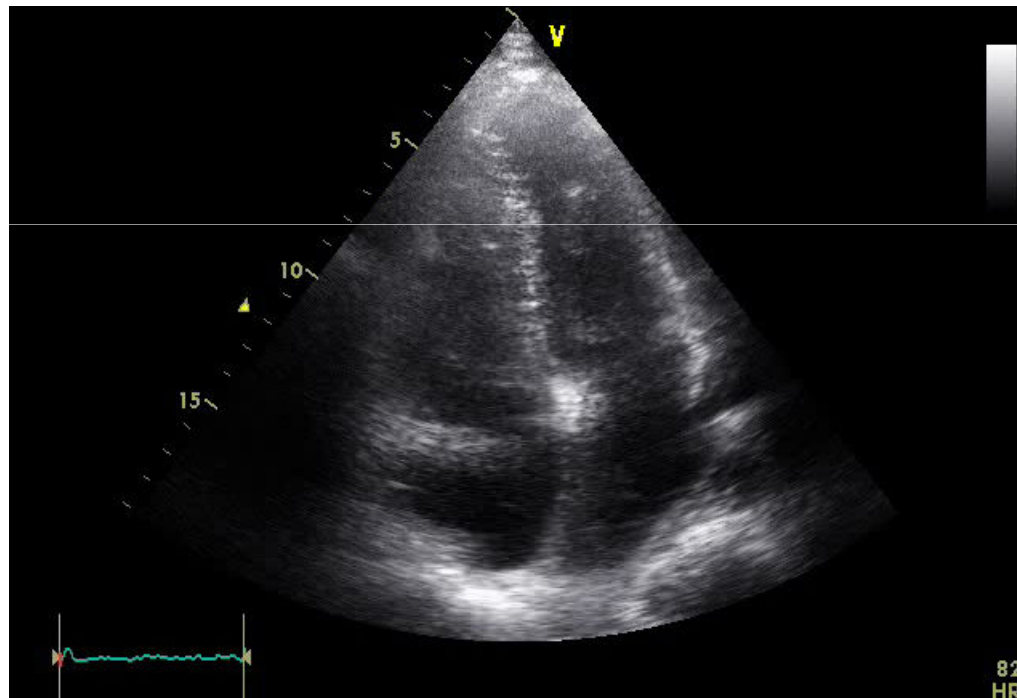
Hurrel et al. *Circulation* 1996

CP: 15 pts R: 21 pts

A puzzling right heart failure



A puzzling right heart failure



Elastic form of constrictive pericarditis

- **Effusive constrictive pericarditis:**
persistence of compressive hemodynamics after fluid removal
- **Subacute constrictive pericarditis**
- **Transient constrictive pericarditis**
 - Spontaneous remission (Soler-Soler, 1986)
 - Steroid treatment