

# **GUIDELINES ON THE DIAGNOSIS AND MANAGEMENT OF PERICARDIAL DISEASES**

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**Full text guidelines: [www.escardio.org](http://www.escardio.org)**

**Pocket Guidelines: Available per request from the ESC**

# **GUIDELINES ON THE DIAGNOSIS AND MANAGEMENT OF PERICARDIAL DISEASES**

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## Guidelines on the Diagnosis and Management of Pericardial Diseases

# MAIN TOPICS

- Acute pericarditis
- Pericardial effusion and cardiac tamponade
- Constrictive pericarditis
- Viral pericarditis
- Bacterial pericarditis
- Pericarditis in renal failure
- Autoreactive pericarditis and pericardial involvement in systemic autoimmune diseases
- The post-cardiac injury syndrome
- Postinfarction pericarditis
- Traumatic pericardial effusion
- Haemopericardium in aortic dissection
- Neoplastic pericarditis
- Pericardial diseases in pregnancy
- Drug- and toxin-related pericardial disease

## **LEVEL OF EVIDENCE**

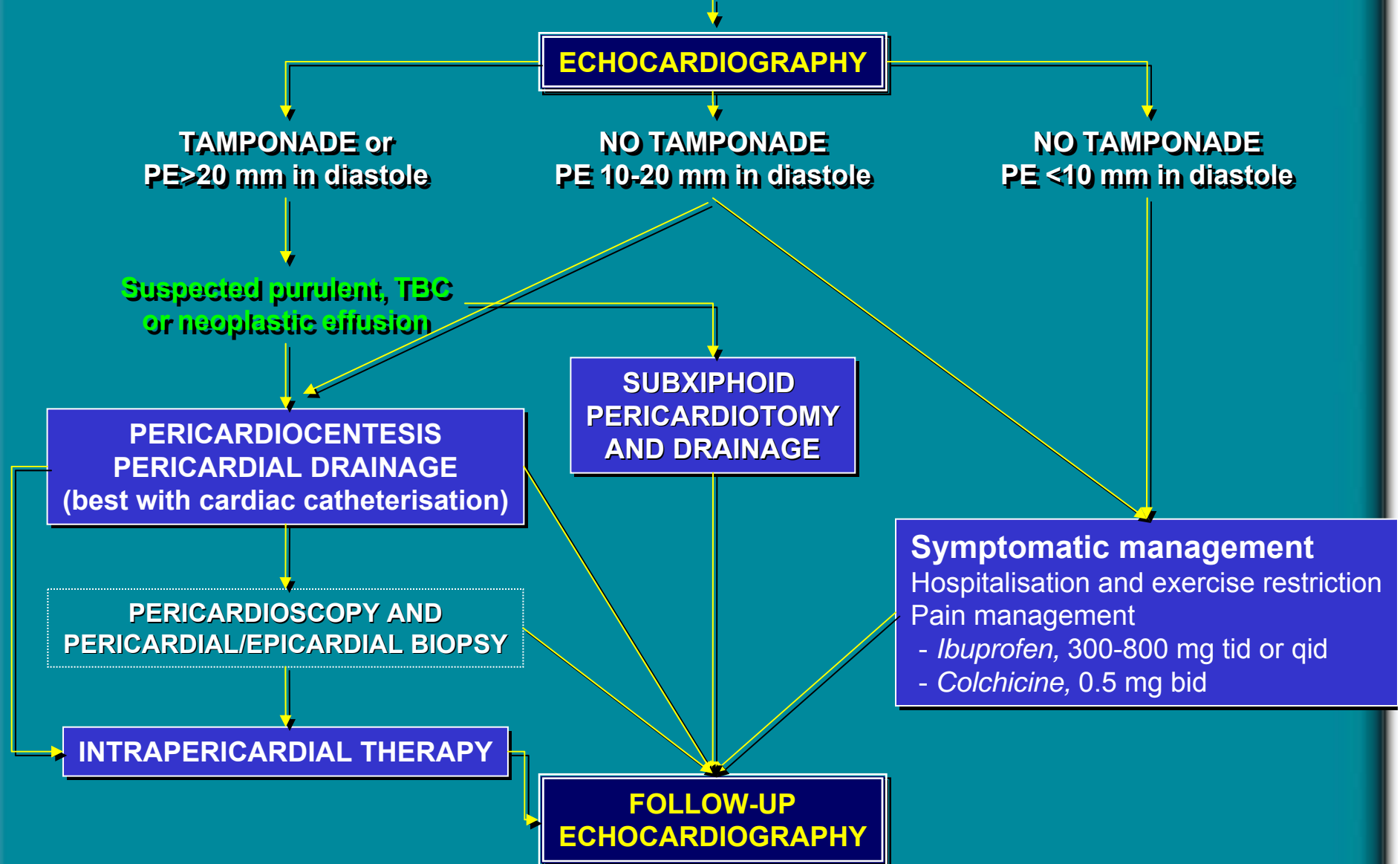
- **Level of evidence A:** Multiple randomised clinical trials or meta-analyses.
- **Level of evidence B:** A single randomised trial or non-randomised studies.
- **Level of evidence C:** Consensus opinion of the experts.

# CLASSES OF RECOMMENDATIONS

- **Class I:** evidence and/or general agreement that a given procedure or treatment is useful and effective.
- **Class II:** conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of a procedure or treatment.
  - **Class IIa:** Weight of evidence/opinion is in favour of usefulness/efficacy.
  - **Class IIb:** Usefulness/efficacy is less well established by evidence/opinion.
- **Class III:** evidence and/or general agreement that the procedure/treatment is not useful/effective and in some cases may be harmful.

# ACUTE PERICARDITIS

# ACUTE PERICARDITIS



# ACUTE PERICARDITIS

## Diagnostic pathway and sequence of performance

**OBLIGATORY (class I, level of evidence B for all procedures):**

<b>Auscultation</b>	Pericardial rub (mono-, bi-, or triphasic)
<b>ECG<sup>a</sup></b>	<p><u>Stage I:</u> anterior &amp; inferior concave ST segment elevation. PR segment deviations opposite to P polarity.</p> <p><u>Early stage II:</u> ST junct. return to baseline, PR deviated.</p> <p><u>Late stage II:</u> T waves progressively flatten and invert</p> <p><u>Stage III:</u> generalised T wave inversions</p> <p><u>Stage IV:</u> ECG returns to prepericarditis state.</p>
<b>Echocardiography</b>	Effusion types B-D (Horowitz) Signs of tamponade
<b>Blood analyses</b>	a) ESR, CRP, LDH, leukocytes (inflammation markers) b) cTnl, CK-MB (markers of myocardial lesion) <sup>b</sup>
<b>Chest x-ray</b>	Ranging from normal to “water bottle” heart shadow. Revealing additional pulmonary/mediastinal pathology.

<sup>a</sup>. Typical lead involvement: I, II, aVL, aVF, and V3-V6.

<sup>b</sup>. Cardiac troponin is detectable in 32.2-49%. An increase beyond 1.5 ng/ml is rare (7.6-22%).

# **ACUTE PERICARDITIS**

## **Diagnostic pathway and sequence of performance**

(level of evidence B for all procedures)

**MANDATORY IN TAMPONADE (class I), OPTIONAL IN LARGE/RECURRENT EFFUSIONS OR IF PREVIOUS TESTS INCONCLUSIVE (class IIa) IN SMALL: EFFUSIONS (class IIb):**

**Pericardiocentesis and drainage**

Pericardial fluid cytology, and cultures, PCRs and histochemistry for determination of infection or neoplasia

**OPTIONAL OR IF PREVIOUS TESTS INCONCLUSIVE (class IIa):**

**CT**

Effusions, peri-, and epicardium

**MRI**

Effusions, peri-, and epicardium

**Pericardioscopy, pericardial biopsy**

Establishing the specific aetiology

# **ACUTE PERICARDITIS**

## **Symptomatic management**

- **Exercise restriction**
- **Hospitalisation to determine the aetiology and observe for tamponade as well as the effect of treatment.**
- **Pain management**

# **ACUTE PERICARDITIS**

## **Pain management**

- NSAIDs are the mainstay (level of evidence B, class I).**
- Ibuprofen is preferred (rare side-effects, favourable impact on the coronary flow, and the large dose range: 300-800 mg every 6-8 h)**
- Aspirin 300-600 mg every 4-6 h**
- Indomethacin should be avoided in elderly patients (flow reduction in the coronary arteries).**
- Gastrointestinal protection must be provided.**

# ACUTE / RECURRENT PERICARDITIS

## Treatment and prevention of recurrences



- Colchicine (0.5 mg bid) added to an NSAID or as monotherapy is well tolerated with fewer side effects than NSAIDs (level of evidence B, class IIa).
- Percutaneous balloon pericardiectomy  
In recurrences resistant to medical treatment (level of evidence B, class IIb).

# **ACUTE / RECURRENT PERICARDITIS**

## **Treatment and prevention of recurrences**

- **Corticosteroids**
  - only in patients with poor general condition or in frequent crises (level of evidence C, class IIa).
  - The recommended regimen is: prednisone 1-1.5 mg/kg, for at least one month.
  - If patients do not respond adequately, azathioprine (75-100 mg/day) or cyclophosphamide can be added.
- **Pericardiectomy** - only in frequent and highly symptomatic recurrences resistant to medical treatment (level of evidence B, class IIa).

# CARDIAC TAMPONADE

# **CARDIAC TAMPONADE**

## **Clinical presentation**

- **Elevated systemic venous pressure**
  - **Jugular venous distension is less notable in hypovolemic patients or in “surgical tamponade”.**
  - **An inspiratory increase or lack of fall of the pressure in the neck veins (Kussmaul’s sign) indicates constriction**
- **Hypotension**
  - **Occasional pts are hypertensive especially if they have pre-existing hypertension**
- **Pulsus paradoxus**
  - **Absent in tamponade complicating atrial septal defect and significant aortic regurgitation.**
- **Tachycardia**
  - **HR may be < 100 b/min in hypothyroidism and in uremic patients**
- **Dyspnoea or tachypnoea with clear lungs**

# CARDIAC TAMPONADE

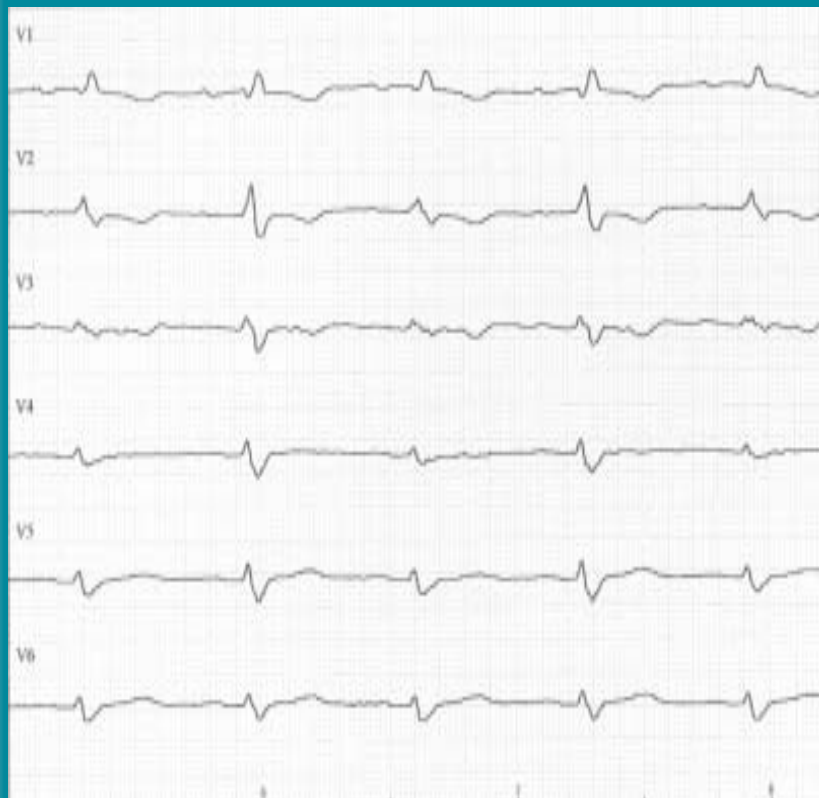
## Precipitating factors



- **Drugs**
  - Cyclosporine
  - Anticoagulants
  - Thrombolytics, etc.
- **Injury**
  - Recent cardiac surgery
  - Indwelling instrumentation
  - Blunt chest trauma
- **Malignancies**
- **Connective tissue disease**
- **Renal failure**
- **Septicaemia**

# CARDIAC TAMPONADE

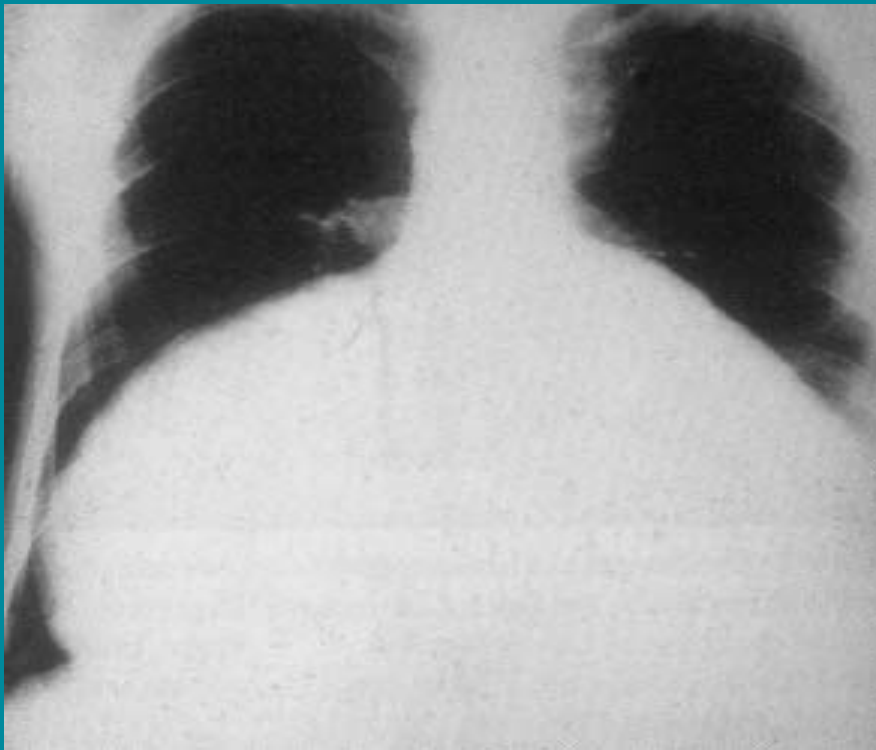
## Electrocardiogram



- Can be normal or
- Non-specifically changed (ST-T wave)
- Electrical alternans (QRS, rarely T)
- Bradycardia (end-stage)
- Electromechanical dissociation (agonal phase)

# CARDIAC TAMPONADE

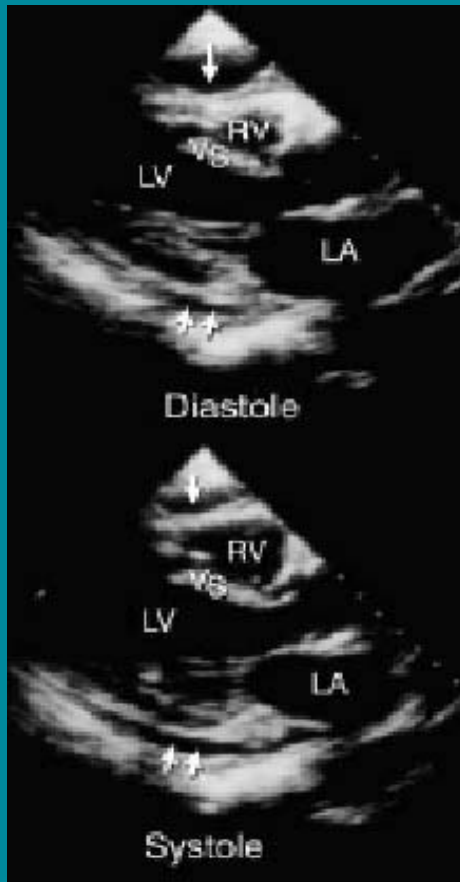
## Chest X-ray



- Enlarged cardiac silhouette with clear lungs

# CARDIAC TAMPONADE

## M-mode/2D echocardiogram

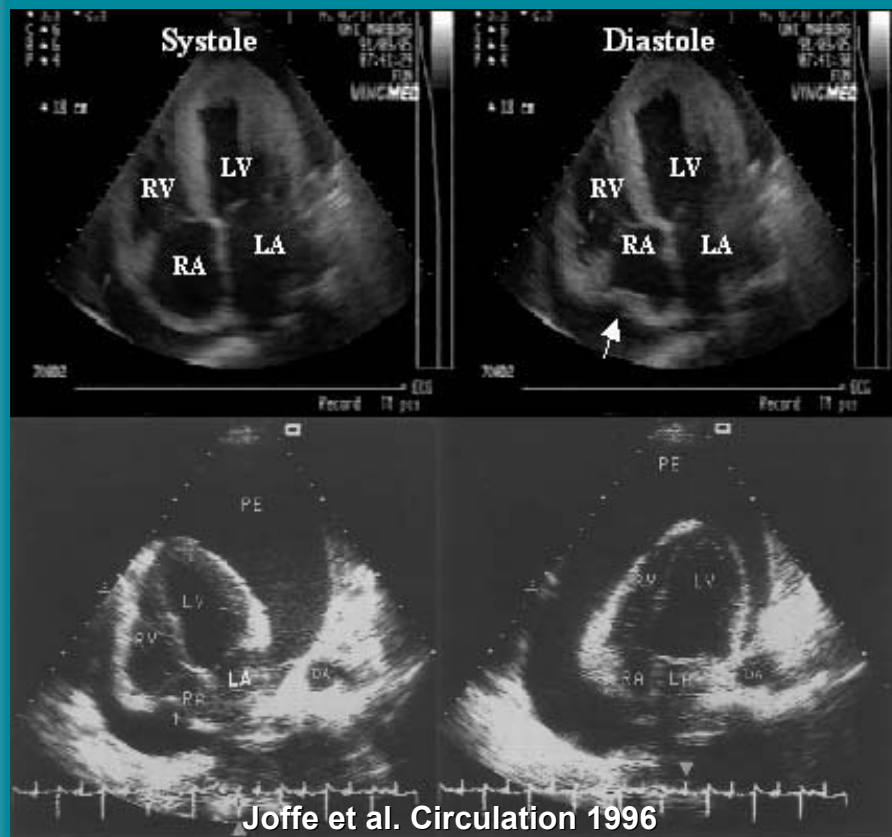


Tsang et al. Herz 2000

- Diastolic collapse of the anterior RV free wall
  - Can be absent in RV hypertrophy or RV infarction.

# CARDIAC TAMPONADE

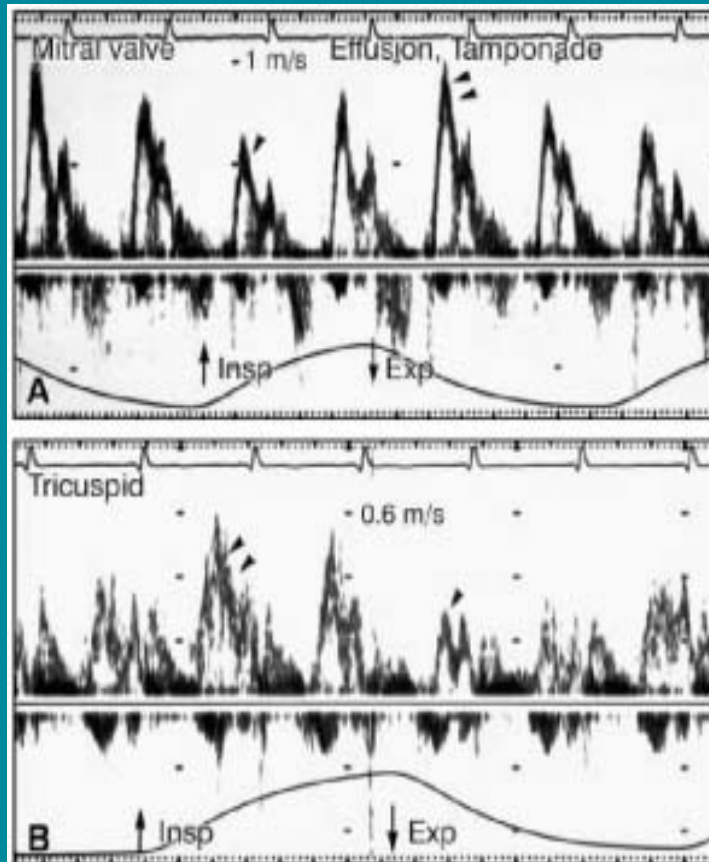
## M-mode/2D echocardiogram



- RA collapse
- LA and very rarely LV collapse
- Increased LV diastolic wall thickness  
"pseudohypertrophy"
- VCI dilatation (no collapse in inspiration)
- "Swinging heart"

# CARDIAC TAMPONADE

## Doppler echocardiography



Oh et al. Mayo Clin Proc 1993

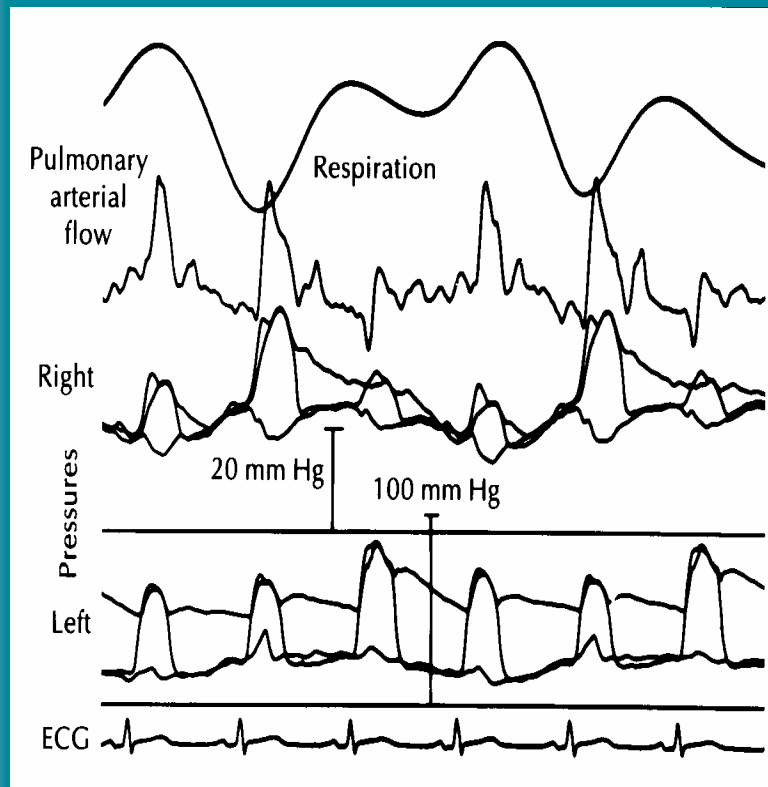
- Tricuspid flow increases and mitral flow decreases during inspiration (reverse in expiration)
- Systolic and diastolic flows are reduced in systemic veins in expiration and reverse flow with atrial contraction is increased.

## M-mode colour Doppler

- Large respiratory fluctuations in mitral/tricuspid flows

# CARDIAC TAMPONADE

## Cardiac catheterisation



Murgo 1982

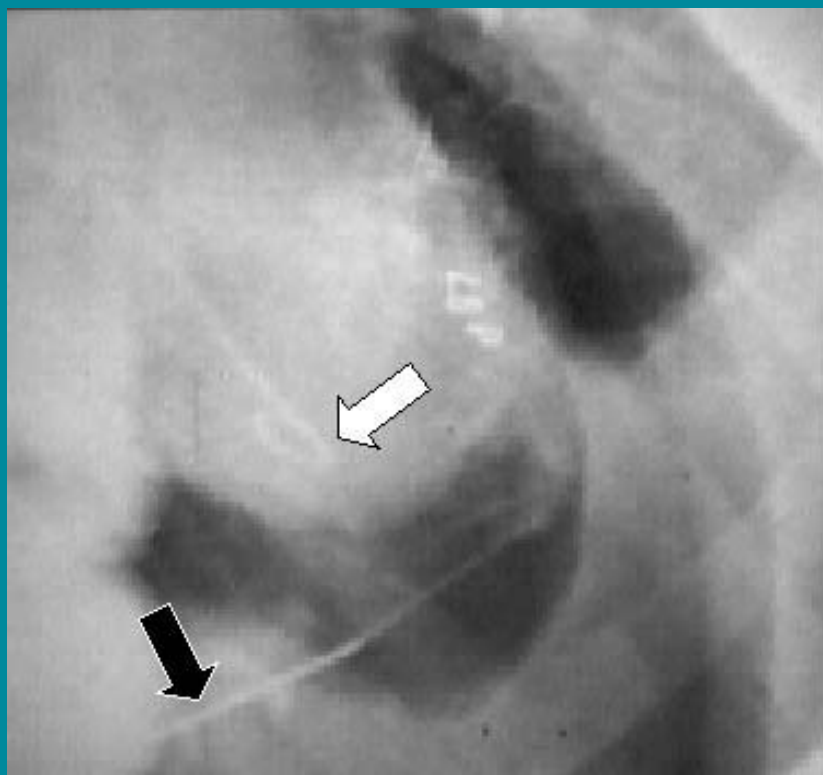
- Confirmation and quantification of the haemodynamic compromise.
- Documenting haemodynamic improvement after pericardiocentesis.
- Detection of the coexisting haemodynamic abnormalities (LV failure, constriction, pulmonary hypertension).
- Detection of associated coronary artery disease or cardiomyopathy.

# **CARDIAC TAMPONADE**

## **Cardiac catheterisation - haemodynamics**

- **Confirmation and quantification of the haemodynamic compromise:**
  - **RA pressure is elevated (preserved systolic x descent and absent or diminished diastolic y descent).**
  - **Intrapericardial pressure is also elevated and virtually identical to RA pressure (both pressures fall in inspiration).**
  - **RV mid-diastolic pressure elevated and equal to the RA and pericardial pressures (no dip-and-plateau configuration).**
  - **PA diastolic pressure is slightly elevated.**
  - **LV systolic and aortic pressures may be normal or reduced.**

# CARDIAC TAMPONADE



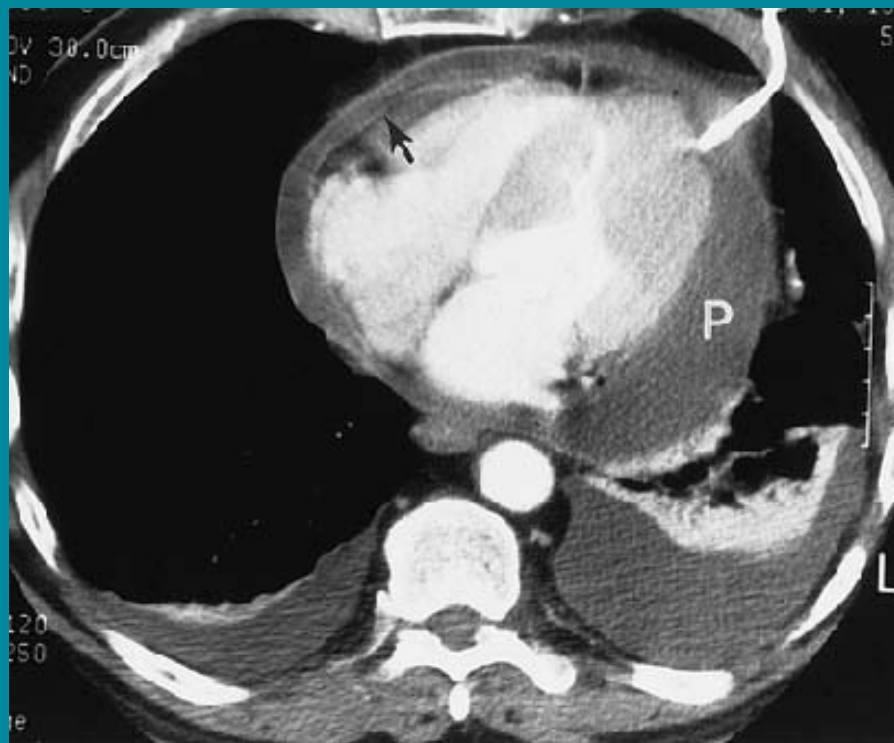
## RV/LV angiography

- **Atrial collapse**
- **Small hyperactive ventricular chambers.**

## Coronary angiography

- **Coronary compression in diastole.**

# CARDIAC TAMPONADE



Chiles et al. Radiographics 2001

## Computer tomography

- **No visualisation of subepicardial fat along both ventricles, which show tube-like configuration and anteriorly drawn atrias**

# PERICARDIOCENTESIS

# **PERICARDIAL EFFUSION / TAMPONADE**

## **Classes of recommendations for pericardiocentesis**

### **Class I**

- Cardiac tamponade
- Effusions >20 mm in echocardiography (diastole)
- Suspected purulent or tuberculous pericardial effusion

### **Class IIa**

- Effusions 10-20 mm in echocardiography in diastole for diagnostic purposes other than purulent pericarditis or tuberculosis (pericardial fluid and tissue analyses, pericardioscopy, and epicardial/pericardial biopsy)
- Suspected neoplastic pericardial effusion

# **PERICARDIAL EFFUSION / TAMPONADE**

## **Classes of recommendations for pericardiocentesis**

### **Class IIb**

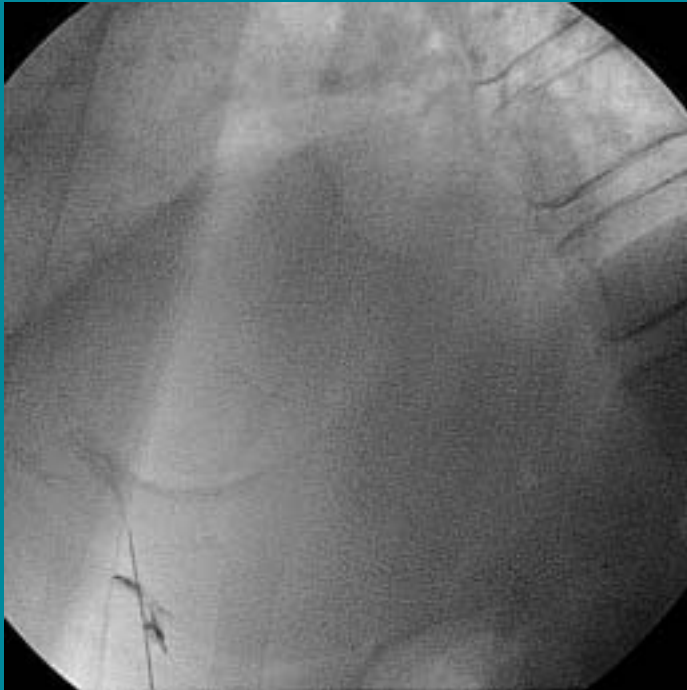
- Effusions <10 mm in echocardiography in diastole for diagnostic purposes other than purulent, neoplastic or tuberculous pericarditis

### **Contraindications (Class III)**

- Aortic dissection
- Relative contraindications include uncorrected coagulopathy, anticoagulant therapy, thrombocytopenia <50000/mm<sup>3</sup>, small, posterior, and loculated effusions.
- If the diagnosis can be made otherwise or the effusions are small and resolving under anti-inflammatory treatment.

# PERICARDIAL EFFUSION / TAMPONADE

## How to perform pericardiocentesis



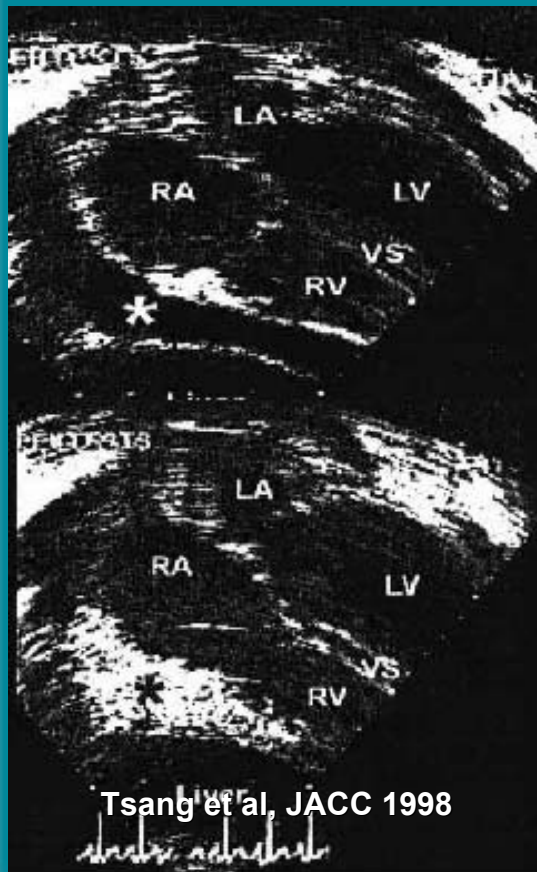
B. Maisch and A.D. Ristic Heart 2003

### Pericardiocentesis guided by fluoroscopy

- Current and reliable echocardiography before the procedure
- Cardiac catheterisation laboratory.
- Local anaesthesia.
- Subxiphoid approach (long needle directed towards the left shoulder at a 30° angle to the skin).

# PERICARDIAL EFFUSION / TAMPONADE

## How to perform pericardiocentesis



### Pericardiocentesis guided by echocardiography

- Bedside, intensive care unit, cardiac cath. lab., or operating theatre.
- Echocardiography should identify the shortest route where the pericardium can be entered intercostally (usually in the sixth or seventh rib space in the anterior axillary line).
- Intercostal arteries should be avoided.

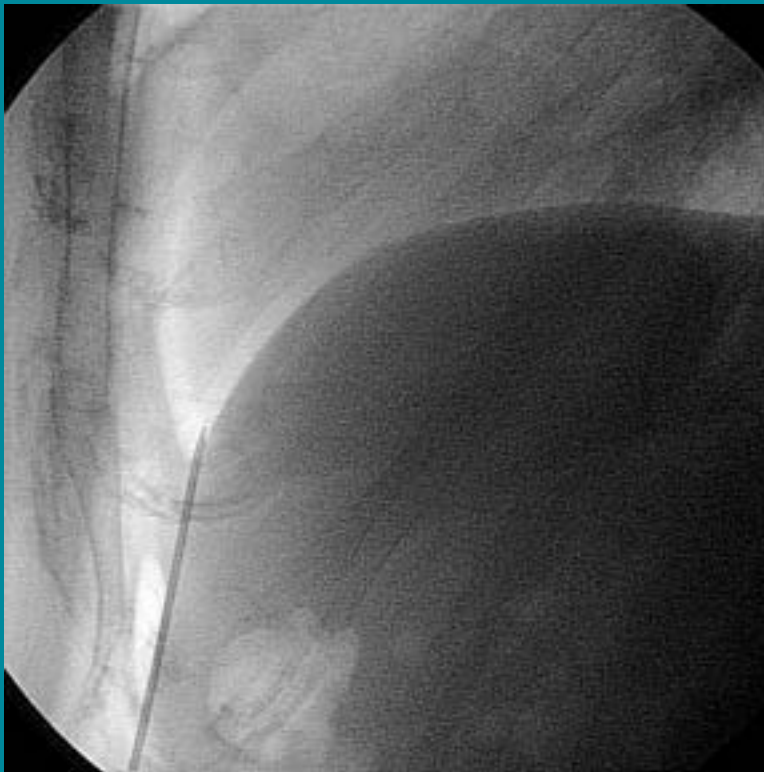
# **PERICARDIAL EFFUSION / TAMPONADE**

## **How to perform pericardiocentesis**

- **Strict aseptic conditions, ECG, and blood pressure monitoring have to be provided.**
- **Direct ECG monitoring from the puncturing needle is not an adequate safeguard.**
- **Right-heart catheterisation can be performed simultaneously, allowing exclusion of constriction.**

# PERICARDIAL EFFUSION / TAMPONADE

## How to perform pericardiocentesis



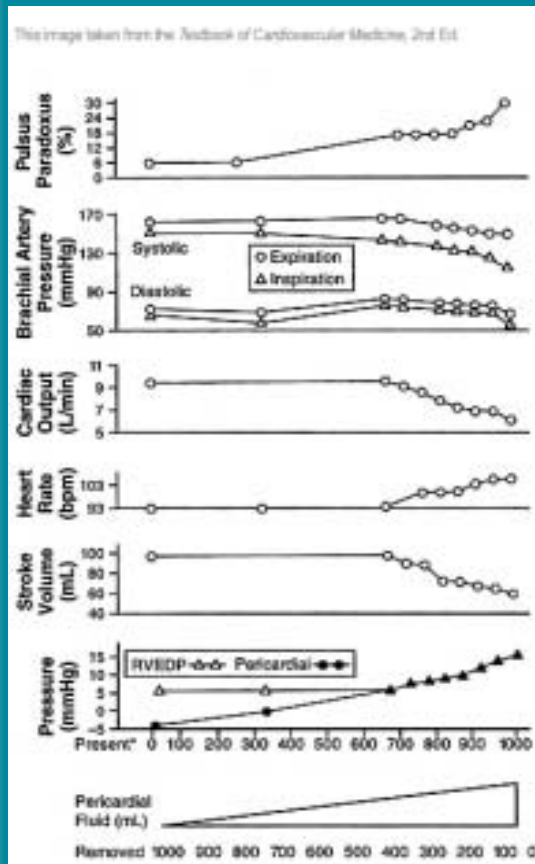
B. Maisch et al. Am J Cardiol 2001

- The needle approaches pericardium slowly.
- Steady manual aspiration is essential.
- Stop the needle as soon as the effusion is aspirated.
- Exchange for soft J-tip guidewire and after dilatation for a multi-holed pigtail catheter.

# Guidelines on the Diagnosis and Management of Pericardial Diseases

## PERICARDIAL EFFUSION / TAMPONADE

### How to perform pericardiocentesis



- Drain the fluid in  $<1$  l steps to avoid the acute right-ventricular dilatation.
- Perform prolonged pericardial drainage (several days) until  $<25$  ml/day.

# PERICARDIAL EFFUSION ANALYSES

Should be ordered according to the clinical presentation

## CLASS I

- **Suspected malignant effusion:** CYTOLOGY.
- **Suspected tuberculous effusion:** ACID-FAST BACILLI STAINING, mycobacterium CULTURE (preferably with radiometric growth detection e.g., BACTEC-460), adenosine deaminase, IFN-gamma, pericardial lysozyme, PCR analyses
- **Suspected bacterial infection:** at least three cultures of pericardial fluid for aerobes and anaerobes as well as three blood cultures. Positive cultures should be followed by sensitivity tests for antibiotics.

# PERICARDIAL EFFUSION ANALYSES

Should be ordered according to the clinical presentation

## CLASS IIa

- **Viral vs. autoreactive pericarditis:** PCR analyses for cardiotropic viruses.
- **Suspected neoplastic pericarditis:** Tumour markers (CEA, AFP, CA 125, CA 72-4, CA 15-3, CA 19-9, CD-30, CD-25...).
- **Benign reactive mesothelial cells vs. adenocarcinoma:** Combination of epithelial membrane antigen, CEA, and vimentin immunocytochemical staining.

# PERICARDIAL EFFUSION ANALYSES

Should be ordered according to the clinical presentation

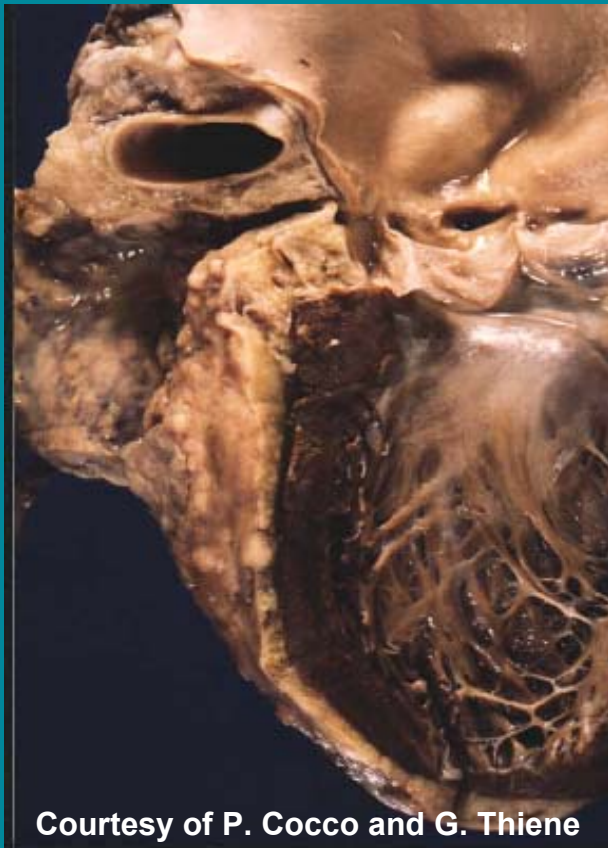
## CLASS IIb

- **Exudate vs. transudate:**
  - Pericardial fluid specific gravity ( $>1015$ )
  - Protein level ( $>3.0$  g/dl; fluid/serum ratio  $>0.5$ )
  - LDH ( $>200$ mg/dL; serum/fluid  $>0.6$ ), and
  - Glucose (exudates vs. transudates =  $77.9 \pm 41.9$  vs.  $96.1 \pm 50.7$  mg/dl)

# CONSTRICTIVE PERICARDITIS

# **CONSTRUCTIVE PERICARDITIS**

## **Clinical presentation**



Courtesy of P. Cocco and G. Thiene

- **Severe chronic systemic venous congestion**
- **Jugular venous distension**
- **Hypotension**
- **Low pulse pressure**
- **Abdominal distension**
- **Oedema**
- **Muscle wasting**

# CONSTRUCTIVE PERICARDITIS

## Thickening of the pericardium

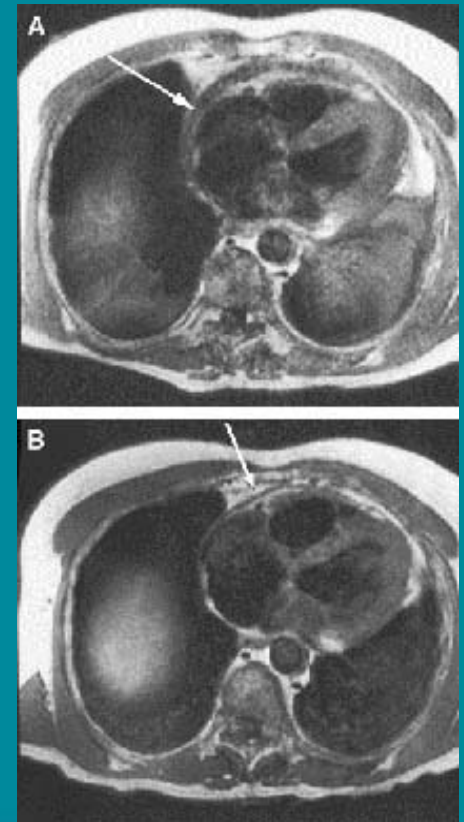
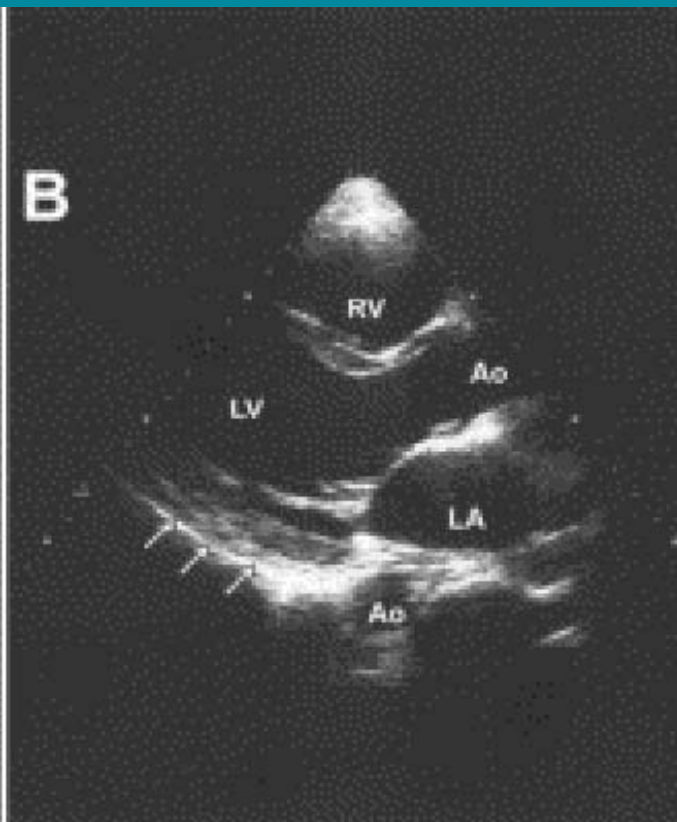
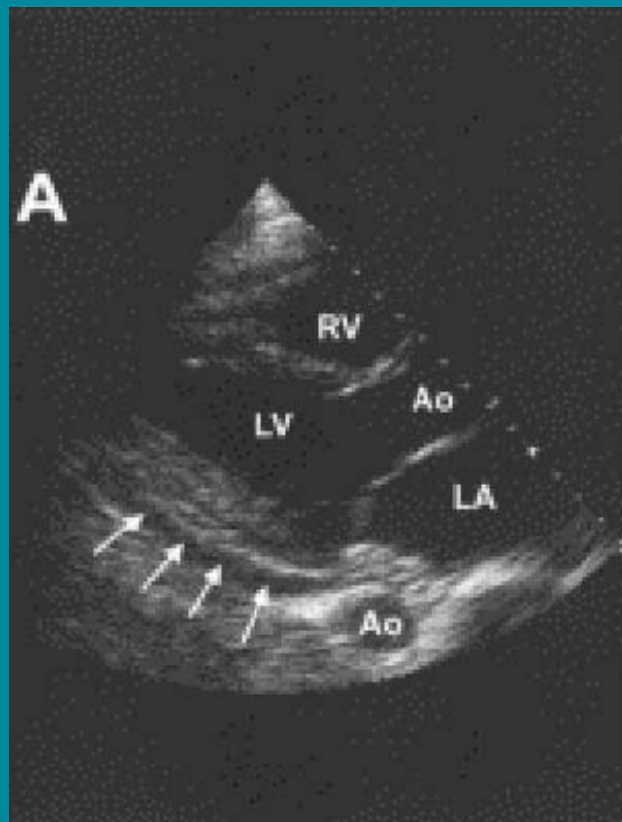


- Not always equal to constrictive physiology
- May also be absent in proven constriction (18% of 143 surgically proven cases, Talreja et al. Circulation 2003).

# CONSTRUCTIVE PERICARDITIS

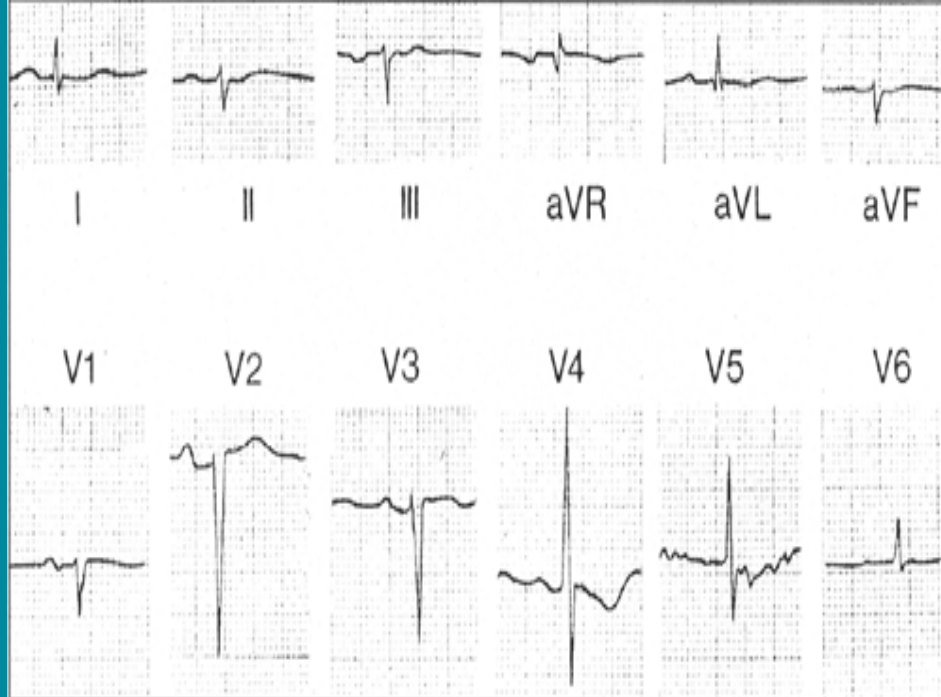
## Transient forms

Haley et al. J Am Coll Cardiol 2004



# CONSTRICTIVE PERICARDITIS

## Electrocardiogram



**Can be normal or:**

- Low QRS voltage
- Generalized T-wave inversion/flattening
- LA abnormalities
- Atrial fibrillation
- AV block
- Intraventricular conduction defects
- Pseudoinfarction pattern (rarely)

# CONSTRICTIVE PERICARDITIS

## Chest X-ray



Ling et al. Ann Intern Med 2000

- Pericardial calcifications
- Pleural effusions

# CONSTRICTIVE PERICARDITIS

## M-mode/2D echocardiogram



Oxantenko et al, J Clin Gastroenterol 2002

- Pericardial thickening and calcifications
- Indirect signs of constriction

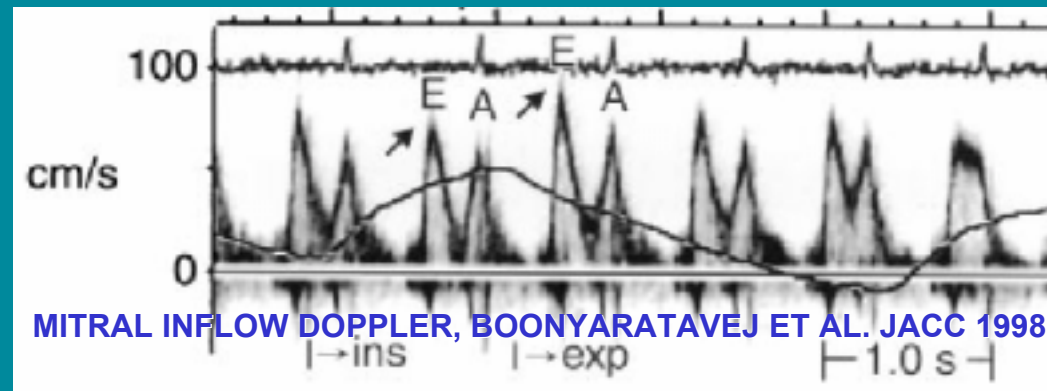
# CONSTRICTIVE PERICARDITIS

## Indirect echo signs of constriction

- RA & LA enlargement with normal ventricles, and systolic function.
- Early pathological outward and inward movement of the interventricular septum (“dip-plateau phenomenon”)
- Fluttering waves at the LV posterior wall
- LV diameter is not increasing after the early rapid filling phase.
- VCI and the hepatic veins are dilated with restricted respiratory fluctuations.

# CONSTRICTIVE PERICARDITIS

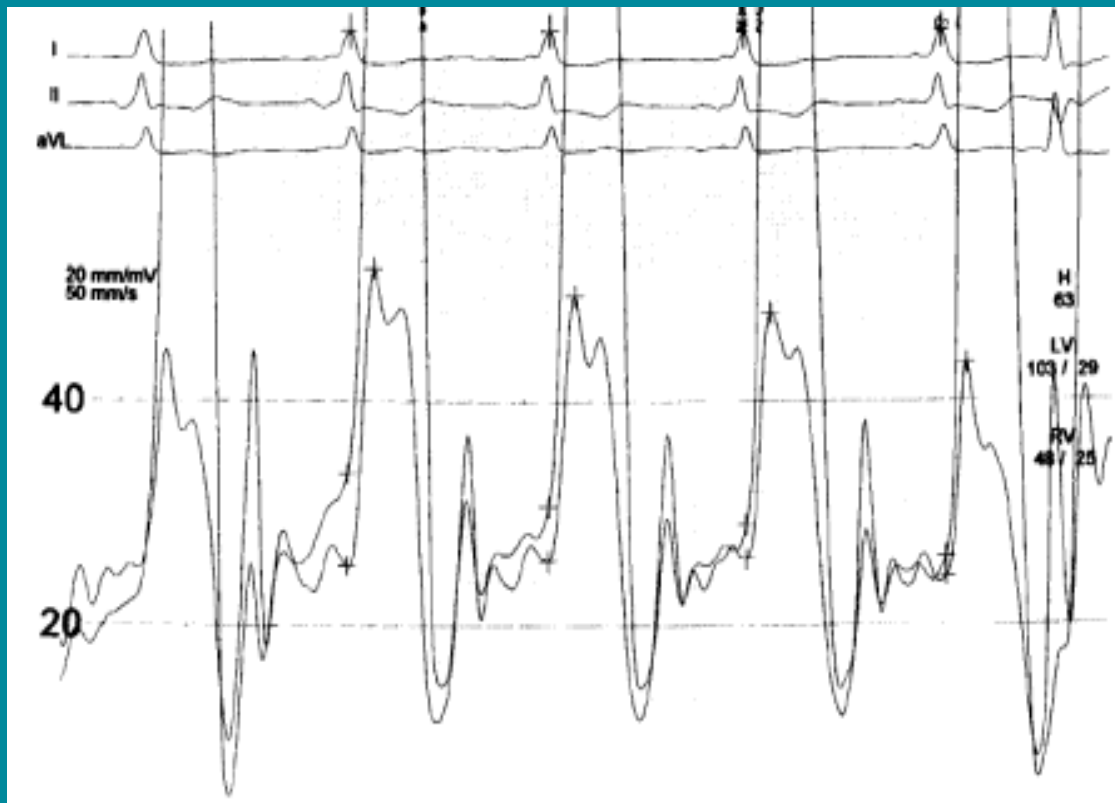
## Doppler echocardiography



- Restricted filling of both ventricles with respiratory variation (>25% over the AV-valves).
  - In mixed constriction-restriction and increased atrial pressures respiratory changes are <25%.
  - In atrial fibrillation flow velocity pattern is inconclusive, but hepatic diastolic vein flow reversal in expirium is observed.
- Provocation test with head-up tilting or sitting position with decrease of preload may unmask the constrictive pericarditis.

# CONSTRUCTIVE PERICARDITIS

## Cardiac catheterisation



Myers and Spodick. Am Heart J 1999

- “Dip and plateau” or “square root” sign in the RV and/or LV pressure curve.
- Equalisation of LV/RV end-diastolic pressures.
- In occult constriction rapid infusion of 1-2 l of normal saline may reveal the diagnosis.

# **CONSTRICTIVE PERICARDITIS**

## **RV/LV angiography**

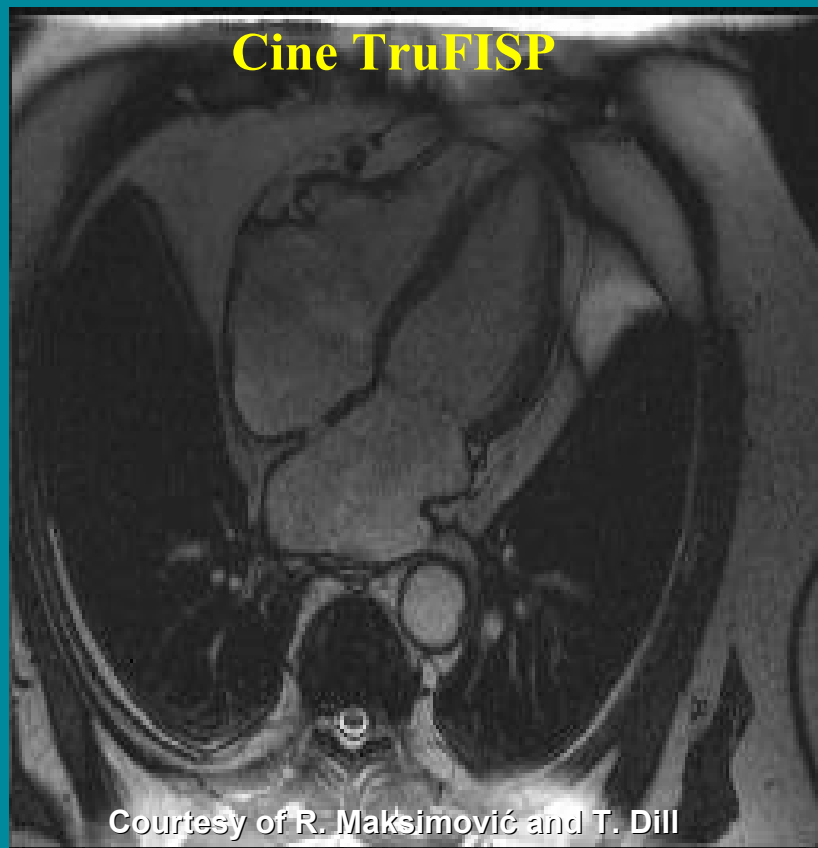
- **The reduction of RV & LV size and increase of RA & LA size**
- **Dip-plateau - rapid early filling with stop of further enlargement in diastole**

## **Coronary angiography**

- **Indicated in all patients over 35 years and in pts with a history of mediastinal irradiation, regardless of the age.**

# CONSTRICTIVE PERICARDITIS

## CT / MRI



- Thickened and/or calcified pericardium
- Tube-like configuration of one or both ventricles
- Enlargement of one or both atria
- Narrowing of one or both atrio-ventricular grooves
- Congestion of the caval veins

**Guidelines on the Diagnosis and Management of Pericardial Diseases**

**CONSTRICTIVE PERICARDITIS VS. RESTRICTIVE CARDIOMYOPATHY**

<b>METHOD</b>	<b>RESTRICTIVE CARDIOMYOPATHY</b>	<b>CONSTRICTIVE PERICARDITIS</b>
<b>Physical findings</b>	Kussmaul's sign $\pm$ , apical impulse +++ S <sub>3</sub> (advanced), S <sub>4</sub> (early disease), regurgitant murmurs ++	Kussmaul's sign +, apical impulse - pericardial knock+, regurgitant murmurs -
<b>ECG</b>	Low voltage, pseudoinfarction, left-axis deviation, AF, conduction disturbances	Low voltage (<50%)
<b>Chest radiography</b>	No calcifications	Calcifications may be present (low diagnostic accuracy)

**Guidelines on the Diagnosis and Management of Pericardial Diseases**

**CONSTRICTIVE PERICARDITIS VS. RESTRICTIVE CARDIOMYOPATHY**

<b>METHOD</b>	<b>RESTRICTIVE CARDIOMYOPATHY</b>	<b>CONSTRICTIVE PERICARDITIS</b>
<b>2D- Echocardiography</b>	Small LV cavity with large atria Increased wall thickness sometimes present (especially thickened interatrial septum in amyloidosis) Thickened valves and granular sparkling (amyloidosis)	Normal wall thickness Pericardial thickening, prominent early diastolic filling with abrupt displacement of IVS
<b>Tissue Doppler Echocardiography</b>	Peak early velocity of longitudinal expansion (peak Ea) of $\geq 8.0$ cm/s (89% sensitivity and 100% specificity)	Negative

**Guidelines on the Diagnosis and Management of Pericardial Diseases**

**CONSTRICTIVE PERICARDITIS VS. RESTRICTIVE CARDIOMYOPATHY**

<b>DOPPLER STUDIES</b>	<b>RESTRICTIVE CARDIOMYOPATHY</b>	<b>CONSTRICTIVE PERICARDITIS</b>
<b>Mitral inflow</b>	No respiration variation of mitral inflow E wave velocity, IVRT E/A ratio $\geq 2$ , short DT, diastolic regurgitation	INSPIRATION: decreased inflow E wave velocity, prolonged IVRT EXPIRATION: opposite changes, short DT, diastolic regurgitation
<b>Pulmonary vein</b>	Blunted S/D ratio (0.5), prominent and prolonged AR No respiration variation, D wave	S/D ratio = 1, INSPIRATION: decreased PV S and D waves EXPIRATION: opposite changes
<b>Tricuspid inflow</b>	Mild respiration variation of tricuspid inflow E wave velocity, E/A ratio $\geq 2$ , TR peak velocity, no significant respiration change	INSPIRATION: increased tricuspid inflow E wave velocity, increased TR peak velocity, EXPIRATION: opposite

## Guidelines on the Diagnosis and Management of Pericardial Diseases

# CONSTRICTIVE PERICARDITIS VS. RESTRICTIVE CARDIOMYOPATHY

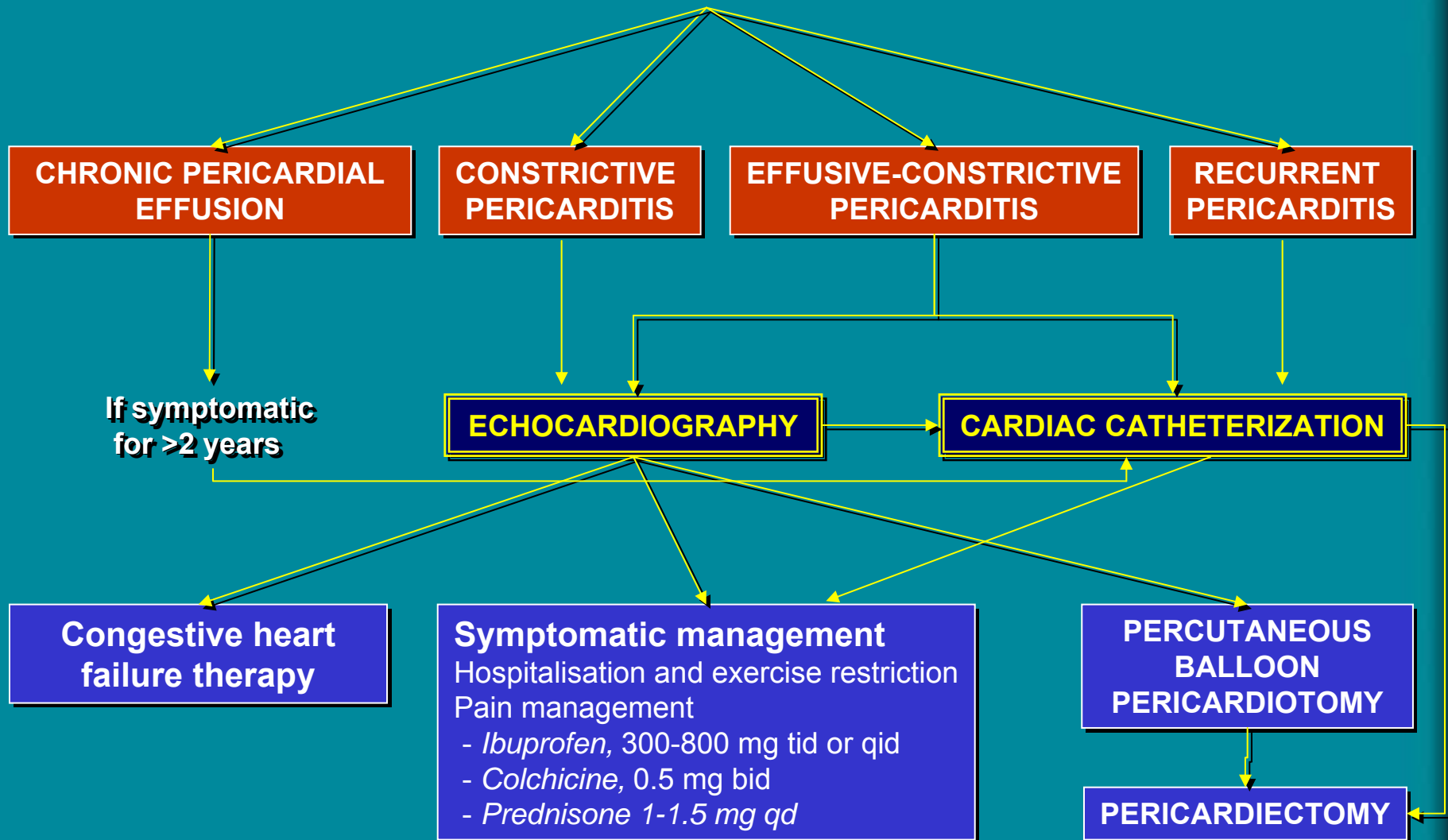
DOPPLER STUDIES	RESTRICTIVE CARDIOMYOPATHY	CONSTRICTIVE PERICARDITIS
<b>Hepatic veins</b>	Blunted S/D ratio, increased inspiratory reversals	INSPIRATION: minimally increased HV, S and D  EXPIRATION: opposite changes
<b>Inferior vena cava</b>	Plethoric	Plethoric
<b>Colour M-mode</b>	Slow flow propagation	Rapid flow propagation ( $\geq 100$ cm/s)
<b>Mitral annular motion</b>	Low-velocity early filling ( $< 8$ cm/s)	High-velocity early filling ( $\geq 8$ cm/s)

**Guidelines on the Diagnosis and Management of Pericardial Diseases**

**CONSTRICTIVE PERICARDITIS VS.  
RESTRICTIVE CARDIOMYOPATHY**

<b>METHOD</b>	<b>RESTRICTIVE CARDIOMYOPATHY</b>	<b>CONSTRICTIVE PERICARDITIS</b>
<b>Cardiac catheterisation</b>	Dip and plateau LVEDP often >5 mmHg greater than RVEDP, but may be identical, RV systolic pressure >50 mmHg RVEDP<1/3 RVSP	Dip and plateau, RVEDP and LVEDP usually equal, INSPIRATION: Increase in RV systolic pressure Decrease in LV systolic pressure, with EXPIRATION, opposite changes
<b>EMB</b>	May reveal specific cause of restrictive cardiomyopathy	May be normal or show nonspecific hypertrophy or fibrosis
<b>CT/MRI</b>	Pericardium usually normal	Pericardium thickened or calcified.

## CLINICAL SUSPICION FOR:



# **CONSTRICTIVE PERICARDITIS**

## **Pericardiectomy**

- **The only treatment for permanent constriction.**
  - **Antero-lateral thoracotomy**
  - **Median sternotomy (faster access to the aorta and right atrium for extracorporeal circulation).**
- **Primary installation of cardiopulmonary bypass is not recommended (diffuse bleeding following systemic heparinisation).**
- **Areas of strong calcification or dense scarring may be left as islands to avoid major bleeding.**

# **PERICARDIECTOMY FOR CONSTRICTIVE PERICARDITIS**

## **Major complications**

- Acute perioperative cardiac insufficiency (should be treated by fluid substitution and catecholamines, high doses of digitalis, and intraaortic balloon pump in most severe cases).
- Ventricular wall rupture.
- Mortality (in properly selected cases 6-12%, but >40% in unselected patients).

## **Long term results**

- If indication for surgery was established early, long-term survival after pericardiectomy corresponds to that of the general population

# Guidelines on the Diagnosis and Management of Pericardial Diseases

## PERICARDIECTOMY FOR VARIOUS PATHOANATOMICAL FORMS OF CONSTRICTION

- Exclusion of patients with extensive myocardial fibrosis and/or atrophy significantly reduces the mortality rate



Annular form



Left-sided form



Right sided form



Global form



Global form with  
MYOCARDIAL ATROPHY



Global form with  
MYOCARDIAL FIBROSIS

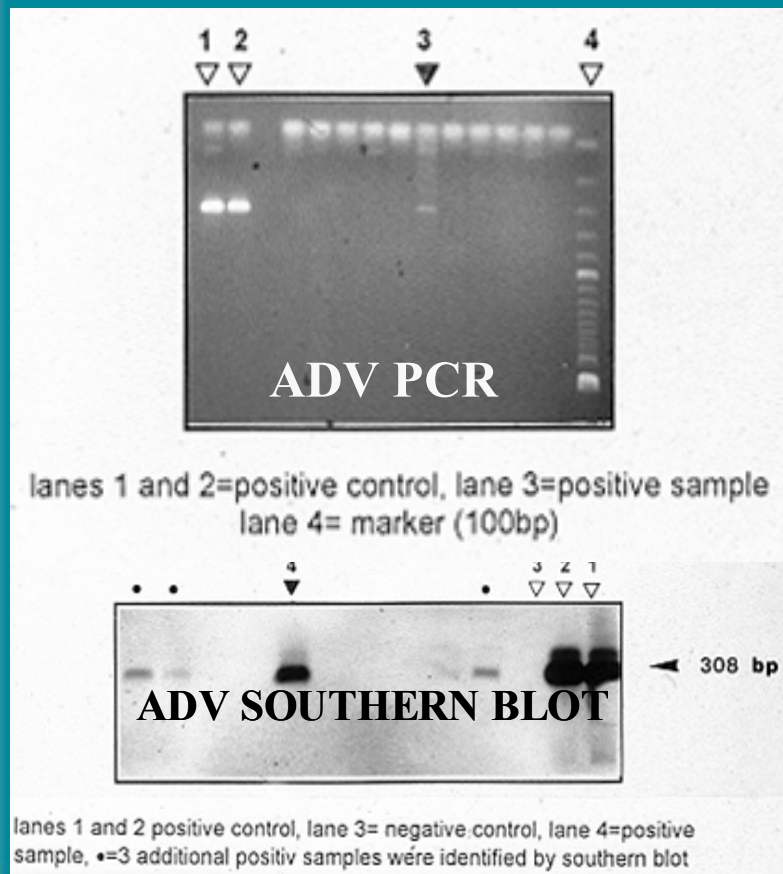
**Very high mortality in pericardiectomy**

Rienmüller et al. J Thorac Imaging 1993

# VIRAL PERICARDITIS

# VIRAL PERICARDITIS

## Diagnosis

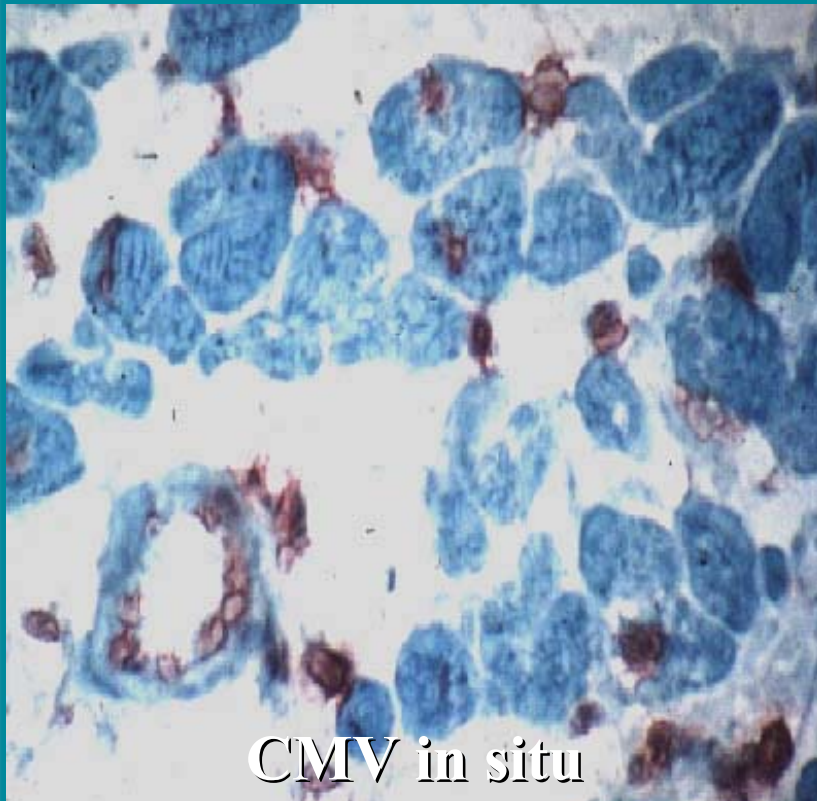


Courtesy of S. Pankuweit

- Not possible without the evaluation of pericardial effusion and/or pericardial/epicardial tissue, preferably by PCR or in-situ hybridisation (level of evidence B, class IIa).
- A four-fold rise in serum antibody levels (two samples within 3-4 weeks) is suggestive but not diagnostic for viral pericarditis (level of evidence B, class IIb).

# VIRAL PERICARDITIS

## Management



CMV in situ

B. Maisch et al. Clin Cardiol 1999

- In most cases the disease is self-limiting and no specific treatment is necessary.
- Symptomatic treatment for chest pain, and eventual rhythm disorders and congestive heart failure is indicated.

# VIRAL PERICARDITIS

## Management

- In patients with chronic or recurrent symptomatic pericardial effusion and confirmed viral infection the following specific treatment is under investigation:
  - **CMV pericarditis**: hyperimmunoglobulin – one daily 4ml/kg on day 0, 4, and 8; 2 ml/kg on day 12 and 16;
  - **Coxsackie B pericarditis**: Interferon alpha or beta 2,5 million. IU/m<sup>2</sup> surface area s.c. 3 x per week;
  - **Adenovirus and parvovirus B19 pericarditis**: immunoglobulin treatment: 10 g intravenously at day 1 and 3 for 6-8 hours.

# **PERICARDITIS IN AIDS**

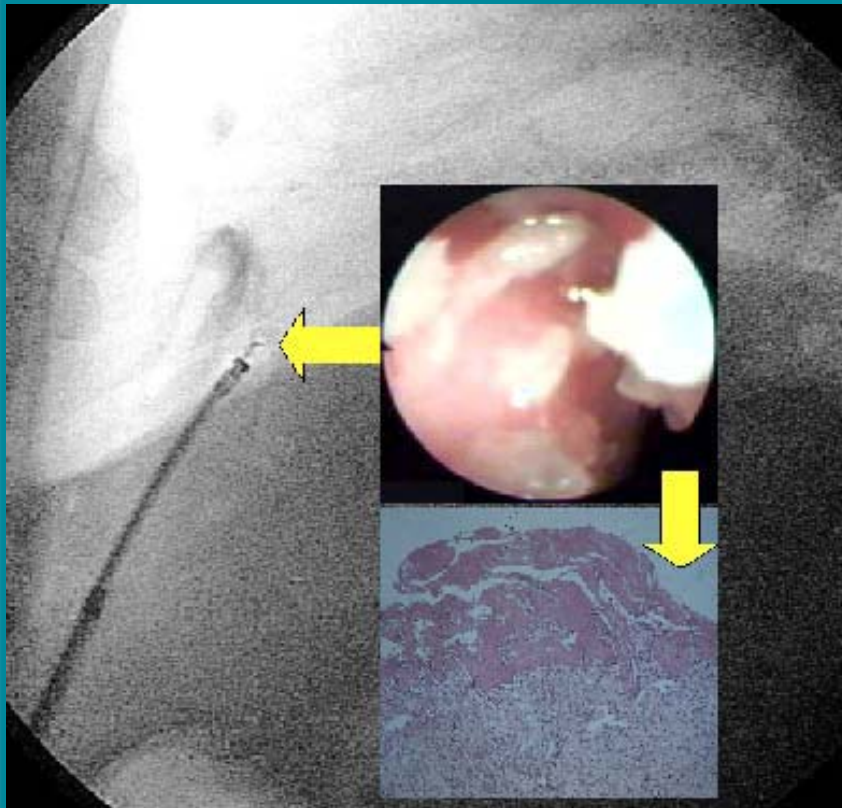
## **Management**

- **Symptomatic treatment**
- **Pericardiocentesis In large effusions/tamponade**
- **Standard (prolonged) anti-tuberculous regimens for TBC pericarditis in AIDS**
- **Use of rifampicin is precluded (if pts are treated with protease inhibitors or non-nucleoside reverse transcriptase inhibitors).**
- **Corticoid therapy as an adjunct to tuberculostatic treatment is allowed (level of evidence A, class I).**

# BACTERIAL PERICARDITIS

# **BACTERIAL PERICARDITIS**

## **Diagnosis**



- Pericardiocentesis must be promptly performed.
- Pericardial fluid should undergo Gram, acid-fast, and fungal staining, followed by cultures for aerobes, anaerobes, and *M. tuberculosis* (preferably with radiometric growth detection).
- Drug sensitivity testing is essential for treatment selection.

# TUBERCULOUS PERICARDITIS

## Diagnosis

- PCR analyses
- Adenosine deaminase  
>40 IU/L
- Interferon-gamma  
>200 pg/L
- Pericardial lysozyme  
>6.5 microg/dL

Cost-effective  
only if the pre-  
test probability  
is high  
(populations  
with high  
incidence of  
tuberculosis).

# **BACTERIAL PERICARDITIS**

## **Management**

- **Urgent pericardial drainage**
- **Intravenous antibiotic therapy (e.g. vancomycin 1 g bid, ceftriaxone 1-2 g bid, and ciprofloxacin 400 mg/day (MIC and MBC need to be considered))**
- **Irrigation with urokinase or streptokinase, using large catheters, may liquefy the purulent exudate**
- **Open surgical drainage is preferable.**

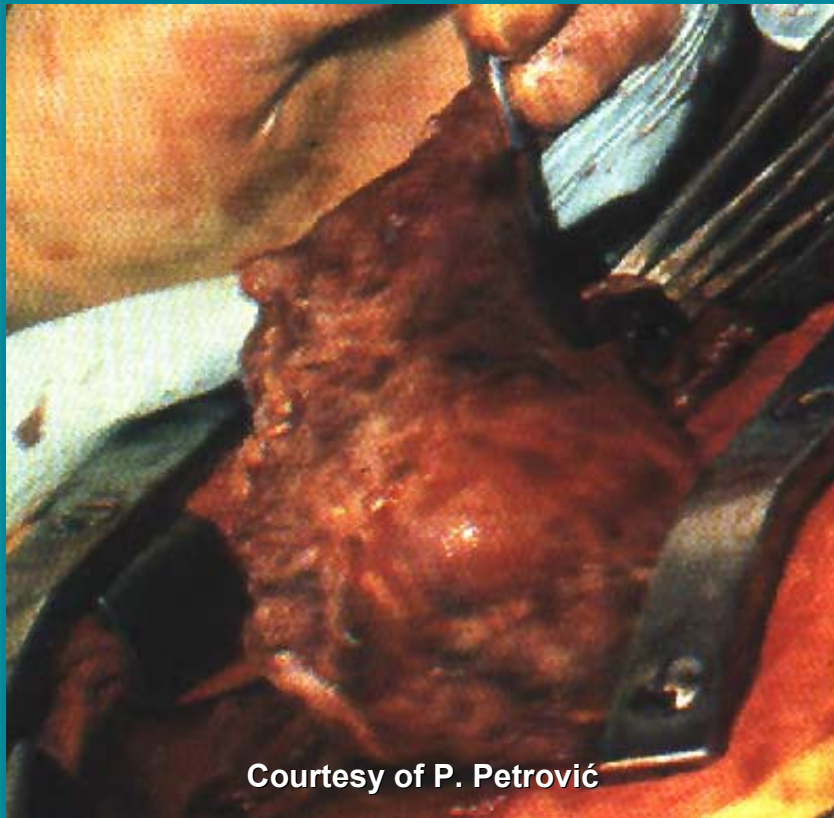
# TUBERCULOUS PERICARDITIS

## Management

- Respiratory isolation in active laryngeal or lung TBC.
- The initial treatment:
  - Isoniazid 300 mg/day
  - Rifampicin 600 mg/day
  - Pyrazinamide 15-30 mg/kg/day
  - Ethambutol 15-25 mg/kg/day.
  - Prednisone (1-2 mg/kg/day) may be given simultaneously with antituberculous therapy for 5-7 days and progressively reduced to discontinuation in 6-8 weeks.
- After two months most patients can be switched to two-drug regimen (isoniazid and rifampicin) for the total of 6 months.

# **TUBERCULOUS PERICARDITIS**

## **Pericardiectomy**



Courtesy of P. Petrović

- **Recurrent effusions**
- **Constriction (continued elevation of central venous pressure after 4-6 weeks of antituberculous and corticosteroid therapy).**

# PERICARDITIS IN RENAL FAILURE

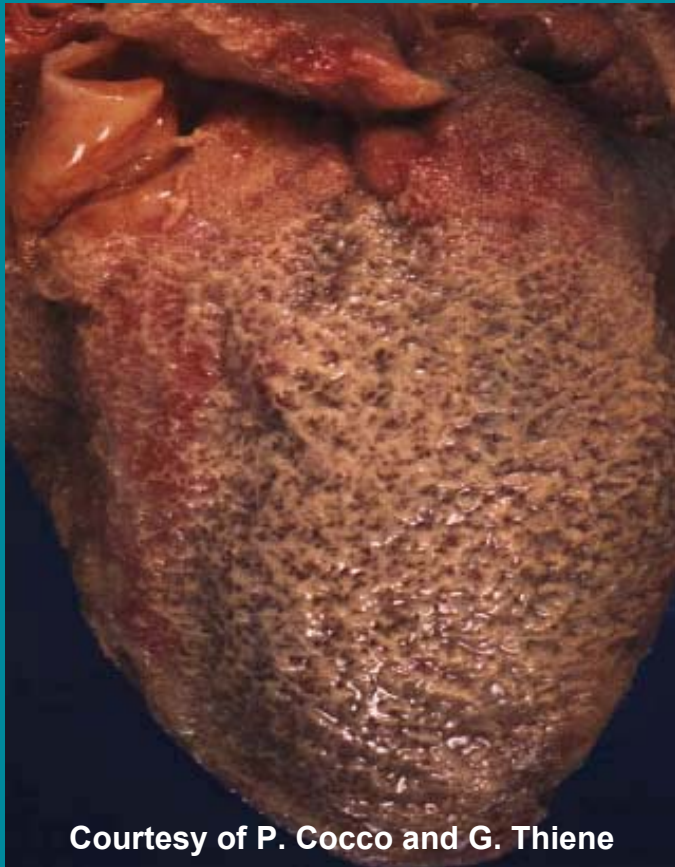
# **PERICARDITIS IN RENAL FAILURE**

## **Diagnosis**

- Chest pain, pericardial friction rub and pericardial effusion in a patient with advanced renal failure.
- Patients on maintenance chronic haemodialysis or peritoneal dialysis can also be affected.
- Heart rate may remain slow (60–80 beats/min) during tamponade, despite fever and hypotension (uremic autonomic impairment).
- No ST/T elevations in ECG due to the lack of the myocardial inflammation.

# **PERICARDITIS IN RENAL FAILURE**

## **Management**



Courtesy of P. Cocco and G. Thiene

- Frequent (heparin-free) haemodialyses.
- Peritoneal dialysis (no heparinisation), may be therapeutic in pericarditis resistant to haemodialysis, or if heparin-free haemodialysis cannot be performed.
- NSAIDs and systemic corticosteroids have limited success when intensive dialysis is ineffective.

# **PERICARDITIS IN RENAL FAILURE**

## **Management**

- **Cardiac tamponade and large chronic effusions resistant to dialysis must be treated with pericardiocentesis (level of evidence B, class IIa).**
- **Large, non-resolving symptomatic effusions may be treated with intrapericardial instillation of corticosteroids (triamcinolone hexacetonide 50 mg every 6 h for 2-3 days).**
- **Pericardiectomy is indicated only in refractory, severely symptomatic patients.**

# PERICARDITIS AND AUTOIMMUNITY

# AUTOREACTIVE PERICARDITIS

## Diagnosis I

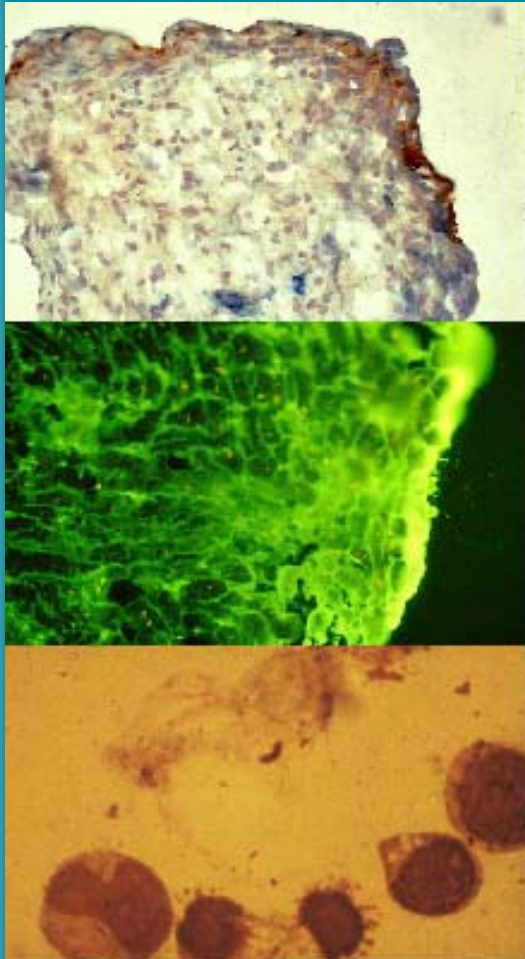
- Pericardial effusion (PE) with  $>5000/\text{mm}^3$  lymphocytes and mononuclear cells (autoreactive lymphocytic), or antibodies against heart muscle tissue (autoreactive antibody-mediated).
- Inflammation in epicardial/endomyocardial biopsies by  $\geq 14$  cells/ $\text{mm}^2$ .
- Exclusion of active viral infection both in PE and endomyocardial/epimyocardial biopsies.
  - No virus isolation.
  - No IgM-titer against cardiotropic viruses in PE.
  - Negative PCRs for major cardiotropic viruses.



B. Maisch et al. Eur Heart J 2002

# AUTOREACTIVE PERICARDITIS

## Diagnosis II



- TBC, *B. burgdorferi*, *C. pneumoniae*, and other bacterial infection excluded by PCR and/or cultures
- Neoplastic infiltration absent in pericardial effusion and biopsy samples
- Exclusion of systemic, metabolic disorders, and renal failure

B. Maisch et al. Eur Heart J 2002

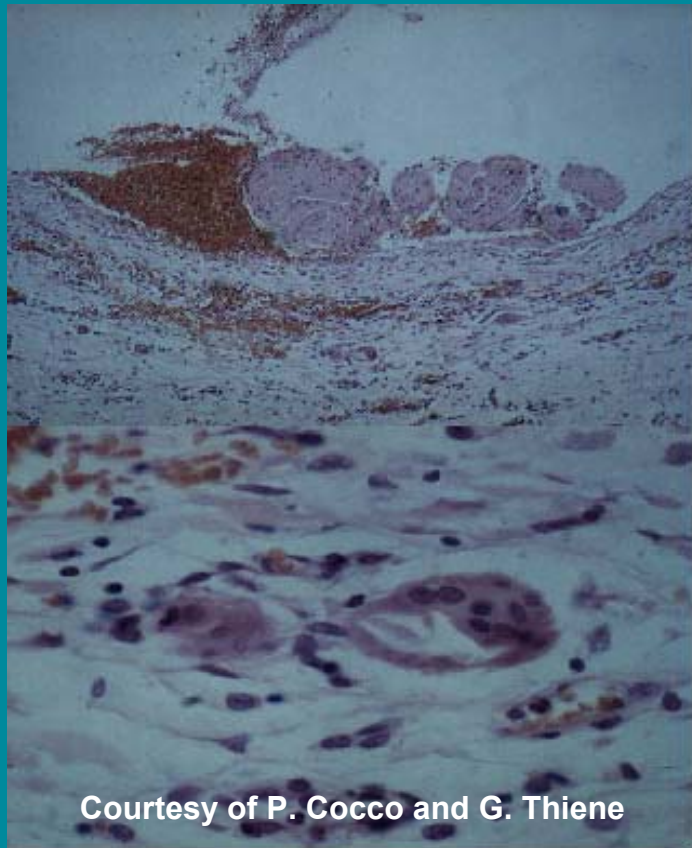
**Guidelines on the Diagnosis and Management of Pericardial Diseases**  
**AUTOREACTIVE PERICARDITIS AND PERICARDIAL INVOLVEMENT IN SYSTEMIC AUTOIMMUNE DISEASES**

## **Management**

- Intrapericardial treatment with triamcinolone plus colchicine per os 0.5 mg bid for six months is highly efficient with rare side effects (level of evidence B, class IIa).
- In systemic autoimmune diseases intensified treatment of the underlying disease and symptomatic management are indicated (evidence level B, class I).
- For tapering of prednisone, ibuprofen or colchicine should be introduced early.

# POSTPERICARDIOTOMY SYNDROME

## Diagnosis



Courtesy of P. Cocco and G. Thiene

- Chest pain
- Pericardial friction rub
- ECG changes
- PE within days to months after cardiac, pericardial injury or both.

# **POSTPERICARDIOTOMY SYNDROME**

## **Management**

- Symptomatic treatment as in acute pericarditis
- In refractory forms long term (3-6 months) oral corticoids or preferably intrapericardial instillation of triamcinolone (300 mg/m<sup>2</sup>)
- Redo surgery or pericardiectomy is rarely needed.
- Primary prevention with short-term perioperative steroid treatment or colchicine is under investigation.
- Warfarin administration in patients with early postoperative PE imposes greatest risk.

# **POSTINFARCTION PERICARDITIS**

**(Pericarditis epistenocardica and Dressler's syndrome)**

## **Diagnosis**

- **Detection of PE after acute myocardial infarction**
- **ECG changes are often overshadowed by myocardial infarction changes.**
- **Postinfarction PE >10 mm is most frequently associated with haemopericardium, and two thirds of these pts may develop tamponade/ free wall cardiac rupture.**

## POSTINFARCTION PERICARDITIS

(Pericarditis epistenocardica and Dressler's syndrome)

### Management I

- **Hospitalisation** to observe for tamponade, differential diagnosis, and adjustments of treatment is needed.
- **Ibuprofen**, which increases coronary flow, is the agent of choice.
- **Aspirin**, up to 650 mg every 4 hours for 2 to 5 days has also been successfully applied (other NSAIDs risk thinning the infarction zone).

# **POSTINFARCTION PERICARDITIS**

**(Pericarditis epistenocardica and Dressler's syndrome)**

## **Management II**

- **Corticosteroid therapy should be used for refractory symptoms only (potential delay in myocardial infarction healing).**
- **In cardiac rupture, urgent surgical treatment is life saving.**
- **If the immediate surgery is not possible pericardiocentesis and intrapericardial fibrin-glue instillation could be an alternative.**

# PERICARDIAL EFFUSION IN TRAUMA AND AORTIC DISSECTION

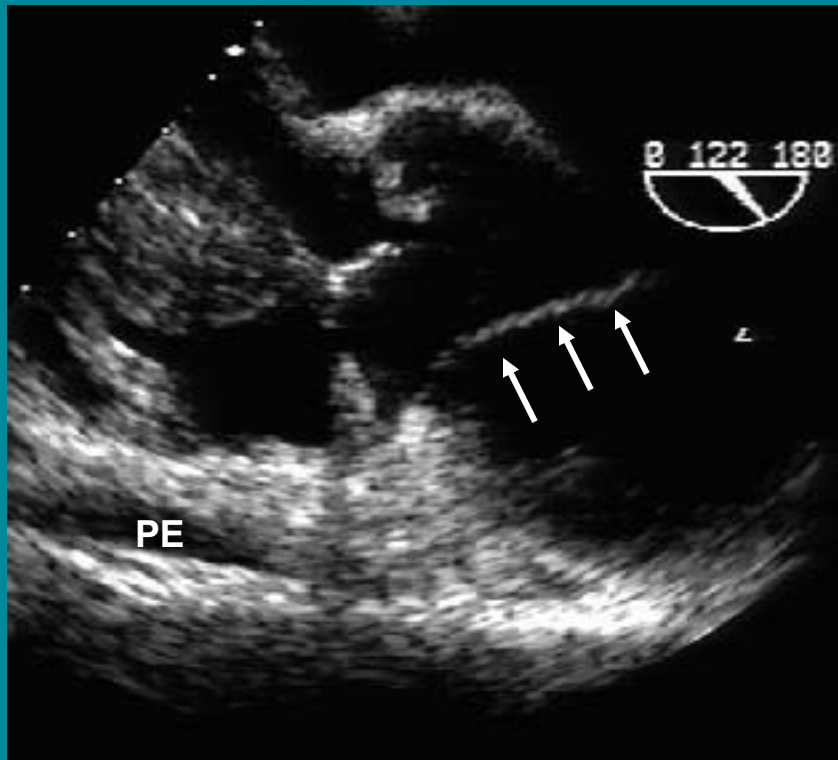
# **TRAUMATIC PERICARDIAL EFFUSION**

## **Management**

- **Urgent echocardiography, if available TEE**
- **Rescue pericardiocentesis**
- **Autotransfusion**
- **Urgent thoracotomy and surgical repair**

# HAEMOPERICARDIUM IN AORTIC DISSECTION

## Diagnosis



- Echocardiography (both TTE and TEE)
- CT or MRI in complex cases
- Angiography (only in stable patients)

# **HAEMOPERICARDIUM IN AORTIC DISSECTION**

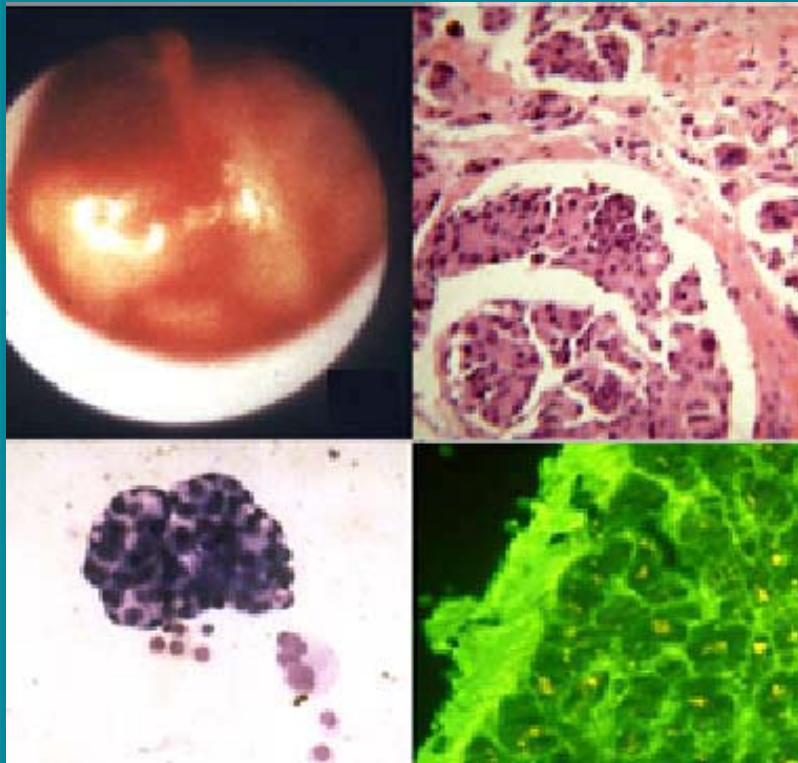
## **Management**

- Pericardiocentesis is contraindicated (risk of intensified bleeding and extension of the dissection).
- Surgery should be performed immediately (evidence level B, class I).

# NEOPLASTIC PERICARDITIS

# NEOPLASTIC PERICARDITIS

## Diagnosis



- Confirmation of the malignant infiltration within the pericardium (cytology, histology, tumour markers).
- In up to 2/3 of pts with documented malignancy PE is caused by non-malignant diseases, e.g. radiation pericarditis, or opportunistic infections.

B. Maisch et al. Eur Heart J 2002

# NEOPLASTIC PERICARDITIS

## Management I

- Systemic antineoplastic therapy whenever possible (prevents recurrences in up to 67% of cases)
- Pericardiocentesis to relieve symptoms and establish diagnosis (level of evidence B, class IIa)
- Intrapericardial instillation of a cytostatic/sclerosing agent (level of evidence B, class IIa).
  - Cisplatin (single instillation of 30 mg/m<sup>2</sup>) is preferred for pericardial metastases of the lung cancer and
  - Intrapericardial instillation of thiotepa (15 mg on days 1,3, and 5) for breast cancer.

# **NEOPLASTIC PERICARDITIS**

## **Management II**

- **Prolonged pericardial drainage is recommended, in all pts with large effusions because of the high recurrence rates (40-70%)(level of evidence B, class I).**
- **In resistant cases percutaneous balloon pericardiectomy or rarely pericardiectomy may be indicated.**
- **Radiation therapy is very effective in controlling malignant effusion due to radiosensitive tumours (e.g. lymphomas and leukemias), may however, cause myocarditis and pericarditis by itself.**

# PERICARDIAL DISEASES IN PREGNANCY

# **PERICARDIAL DISEASES IN PREGNANCY**

## **Diagnosis**

- **Many pregnant women develop a minimal to moderate clinically silent hydropericardium by the third trimester. Cardiac compression is rare.**
- **ECG changes of acute pericarditis in pregnancy should be distinguished from the slight ST-segment depressions and T-wave changes seen in normal pregnancy.**
- **Occult constriction becomes manifest in pregnancy due to the increased blood volume.**

# PERICARDIAL DISEASES IN PREGNANCY

## Management

- Most pericardial disorders are managed as in nonpregnant pts.
- Caution is necessary while high-dose aspirin may prematurely close the ductus arteriosus.
- Colchicine is contraindicated in pregnancy.
- Pericardiotomy and pericardiectomy can be safely performed if necessary and do not impose a risk for subsequent pregnancies.

# **FOETAL PERICARDIAL EFFUSION**



- Foetal pericardial fluid can be detected by echocardiography after 20 weeks' gestation (normally  $\leq 2$  mm).
- More fluid should raise questions of hydrops foetalis, Rh disease, hypoalbuminemia, and immunopathy or maternally transmitted mycoplasmal or other infections, and neoplasia.

# DRUG- AND TOXIN- RELATED PERICARDIAL DISEASE

# DRUG- AND TOXIN-RELATED PERICARDIAL DISEASE

## A. Drug-induced lupus erythematosus

- Procainamide
- Tocainide
- Hydralazine
- Methyldopa
- Mesalazine
- Reserpine
- Isoniazid
- Hydantoins

## B. Hypersensitivity

- Penicillins
- Tryptophan
- Cromolyn sodium

## C. Idiosyncratic reaction or hypersensitivity

- Methysergide
- Minoxidil
- Practolol
- Bromocriptine

# DRUG- AND TOXIN-RELATED PERICARDIAL DISEASE (continued)

## C. Idiosyncratic reaction or hypersensitivity

- Psicofuranine
- Polymer fume inhalation (Teflon)
- Cytarabine
- Phenylbutazone
- Amiodarone
- Streptokinase
- p-Aminosalicylic acid
- Thiazides
- Streptomycin
- Thiouracils
- 5-Fluorouracil
- Sulfa drugs
- Cyclophosphamide
- Cyclosporine
- Mesalazine
- Vaccines (Smallpox, Yellow fever)
- GM-CSF

# DRUG- AND TOXIN-RELATED PERICARDIAL DISEASE (continued)

## D. Anthracyclines

- Doxorubicin
- Daunorubicin

## E. Serum sickness

- Foreign antisera (e.g., antitetanus)
- Blood products

## F. Venom

- Scorpion fish sting

## G. Foreign-substance reactions

- Talc (Mg silicate)
- Silicones
- Tetracycline/other sclerosants
- Asbestos
- Iron in  $\beta$ -thalassemia

## H. Haemopericardium

- Anticoagulants
- Thrombolytic agents

# DRUG- AND TOXIN-RELATED PERICARDIAL DISEASE (continued)

- I. Polymer fume fever – Inhalation of the burning fumes of polytetrafluoroethylene (Teflon)