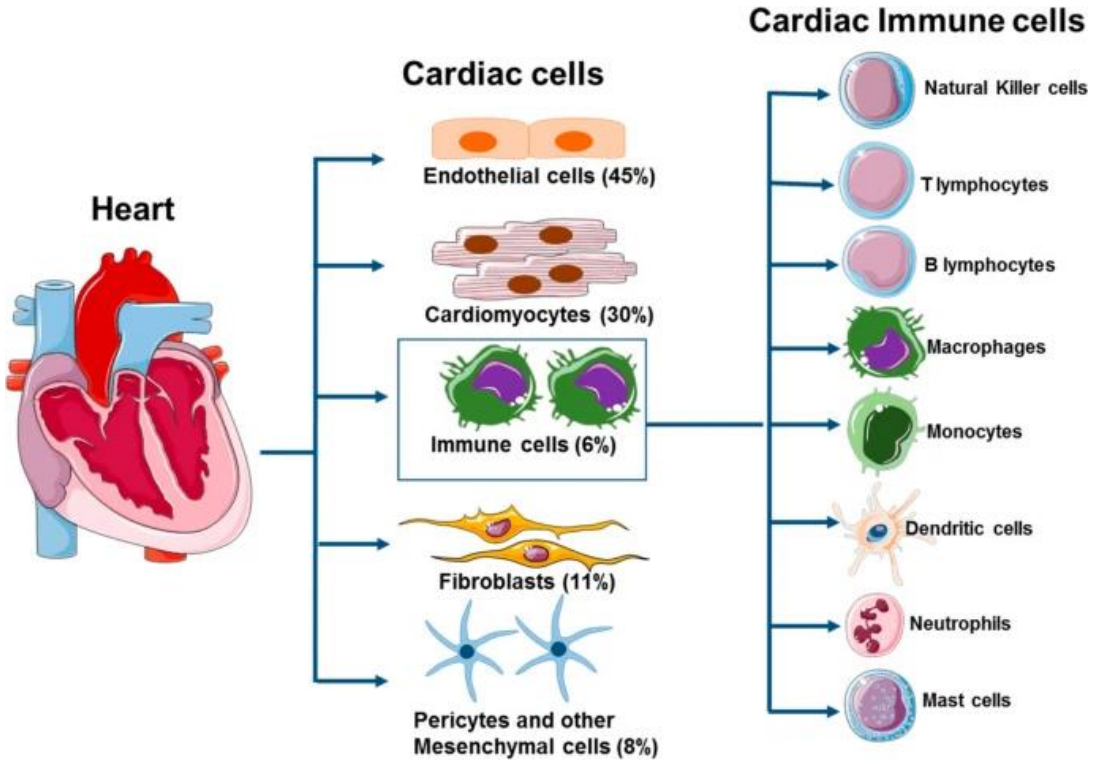
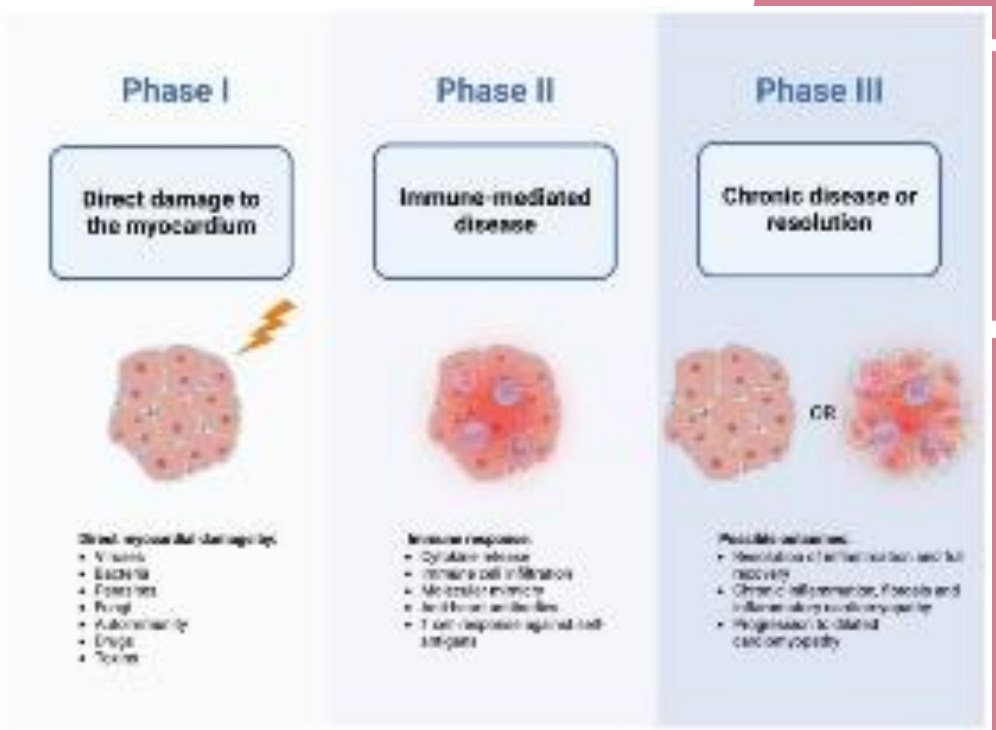
# ESC-guidelines Perikardit/myokardit

**Per Sundbom**

**Höglandssjukhuset Eksjö**

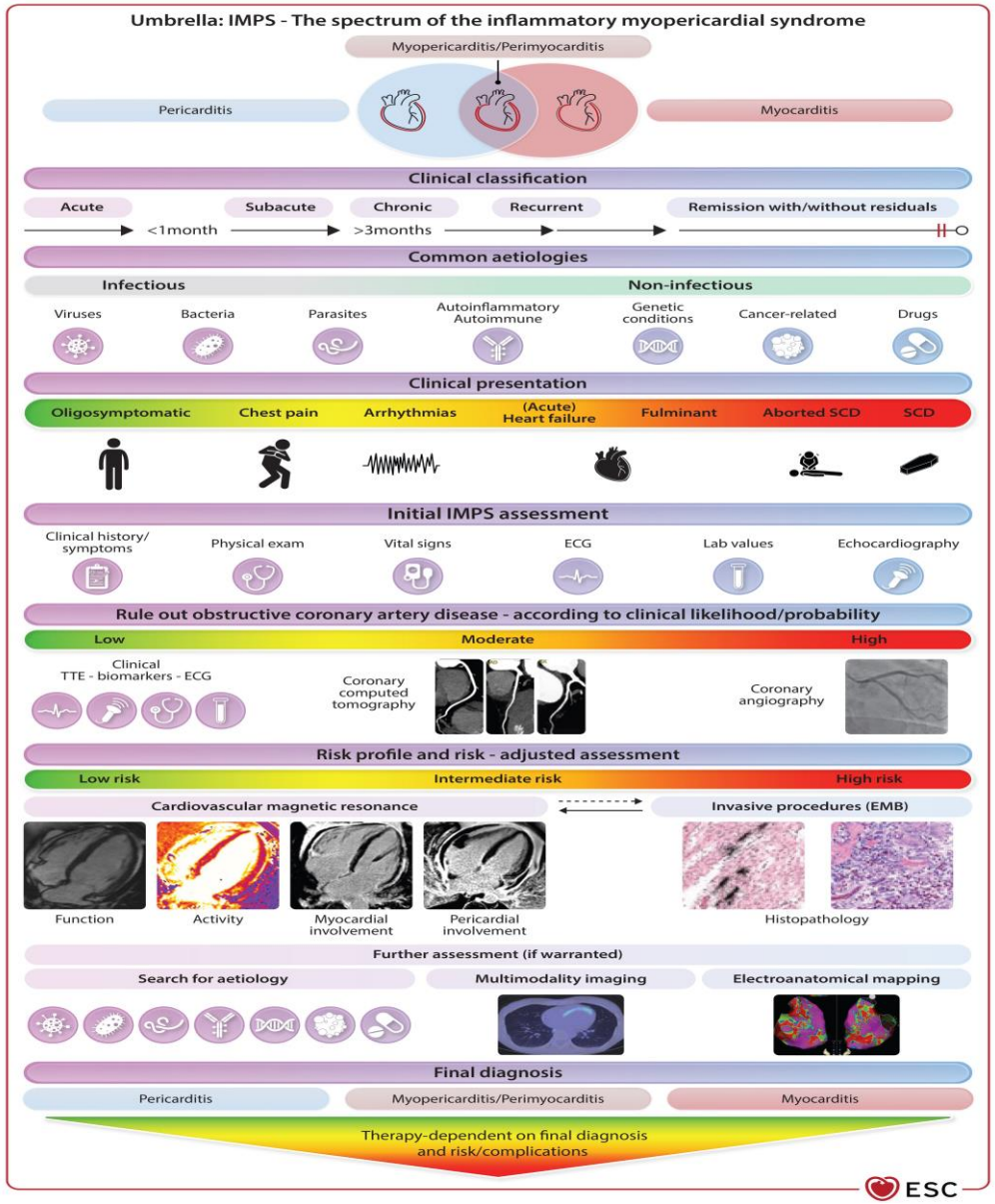
Region Jönköpings län

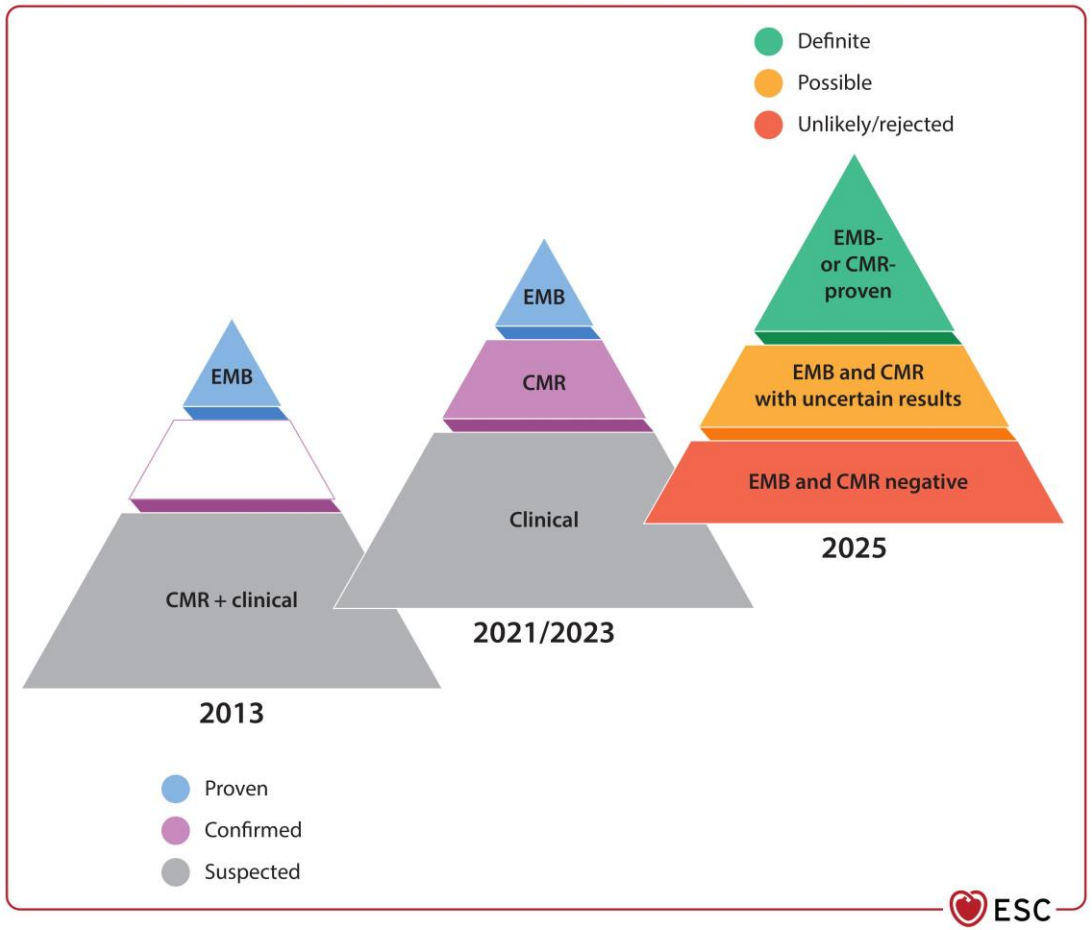



Becker, R.C. Immune checkpoint inhibitors and cardiovascular toxicity: immunology, pathophysiology, diagnosis, and management. *J Thromb Thrombolysis* (2025). https://doi.org/10.1007/s11239-025-03146-7

Brociek E, Tymińska A, Giordani AS, Caforio ALP, Wojnicz R, Grabowski M, Ozierański K. Myocarditis: Etiology, Pathogenesis, and Their Implications in Clinical Practice. Biology (Basel). 2023 Jun 17;12(6):874. doi





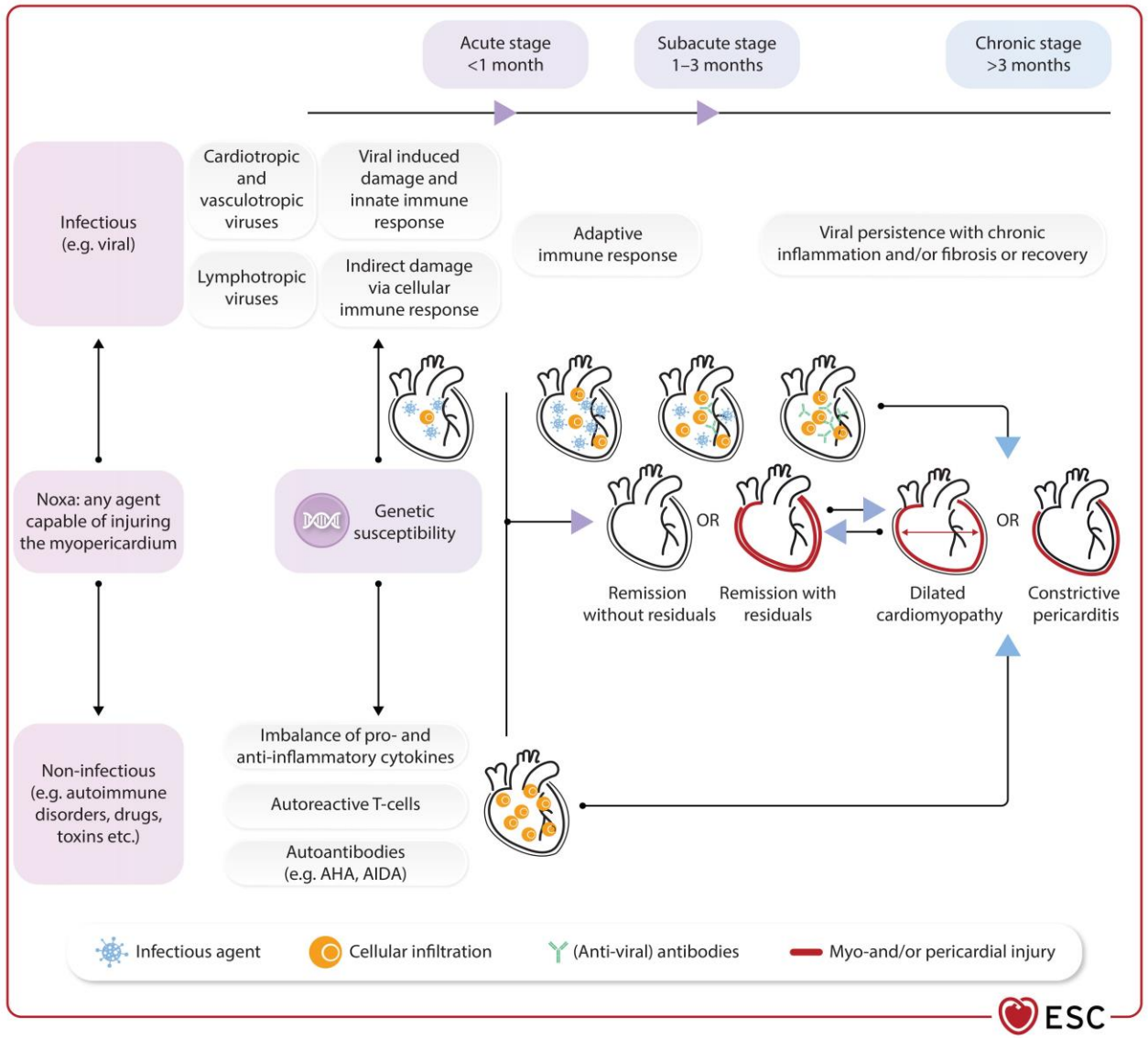

Region Jönköpings län

# Epidemiologi

- Andelen med myokardit ökat från 5% till 13% sedan införande av MRT diagnostik i frånvaro av kranskärlssjukdom och högsensitivt troponin.
- Akut myokardit: 6,3-8,6/100.000 hos unga män.
  4,2-8,7/100.000 hos 35-39åå. Bias?
- Akut perikardit: 3-32/100.000 Bias?
  Återfall hos 20-30% av patienterna inom 18mån.
- 0,2% av de som söker för bröstsmärta på akutmottagningen

**Table 3**
Terminology and stages

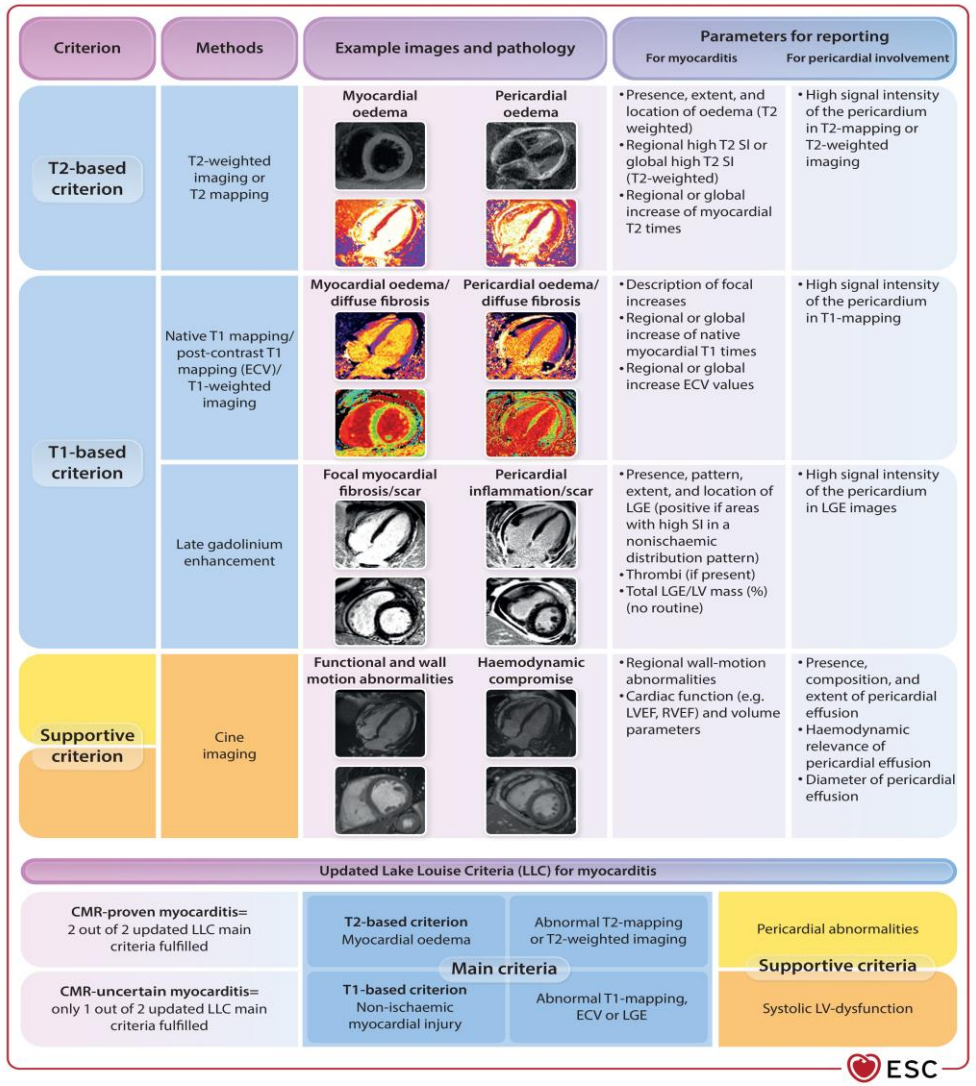
| Terminology | Definition |
|---|---|
| IMPS | Umbrella term for inflammatory myocardial and pericardial syndromes |
| Myopericarditis | Predominant pericarditis[a] |
| Perimyocarditis | Predominant myocarditis[b] |
| Acute myocarditis | Duration of symptoms ≤4 weeks<br>Fulminant if:<br>• Acute onset[28] and haemodynamically unstable patients requiring inotropes or mechanical circulatory support |
| Complicated myocarditis | AM and ≥1 of the following:[28]<br>• LVEF <50% on echocardiogram<br>• Sustained ventricular arrhythmias<br>• Advanced heart block<br>• Heart failure<br>• Cardiogenic shock |
| Acute pericarditis | Duration of symptoms ≤4 weeks |
| Subacute/ongoing myocarditis | Duration of symptoms >4 weeks to ≤3 months |
| Subacute/incessant pericarditis[c] | Duration of symptoms >4 weeks to ≤3 months |
| Chronic myocarditis/pericarditis | Duration of symptoms >3 months |
| Inflammatory cardiomyopathy | Chronic myocarditis in association with cardiac dysfunction and ventricular remodelling with clinical phenotype of hypokinetic, either dilated or non-dilated cardiomyopathy with/without arrhythmogenic substrate |
| Recurrent myocarditis/pericarditis | New symptoms or disease activity after clinical remission |
| Remission without residuals | Regression/absence of symptoms, normalization of ECG, biomarkers, imaging abnormalities (echocardiography and CMR) |
| Remission with residuals | Regression/absence of symptoms, persistence of abnormalities on ECG, biomarkers and/or imaging (functional and/or structural abnormalities in echocardiography or CMR) |






Region Jönköpings län

# Diagnoskriterier baseras på klinisk presentation med stödjande fynd; Myokardit




**IMPS**

If diagnostic criteria for myocarditis and/or pericarditis are fulfilled[a]

| | Myocarditis | Pericarditis |
|---|---|---|
| Definite | Clinical presentation[b] and CMR- or EMB-proven | Clinical presentation[b] with >1 additional criterion |
| Possible | Clinical presentation[b] with at least 1 additional criterion CMR- or EMB-uncertain or not available | Clinical presentation[b] with 1 additional criterion |
| Unlikely/rejected | Only clinical presentation[b] without additional criteria | Only clinical presentation[b] without additional criteria |

Additional criteria beyond clinical presentations[b]

| | Myocarditis | Pericarditis |
|---|---|---|
| Clinical[b] | Non-specific findings | Pericardial rubs |
| ECG[c] | ST-T changes | PR depression, widespread ST-segment elevation |
| Biomarkers | Troponin elevation | C-reactive protein elevation |
| Imaging[d] | Abnormal strain, wall motion, reduced EF<br>Myocardial oedema and/or LGE (CMR findings) | New or worsening pericardial effusion<br>Pericardial oedema and/or LGE (CMR findings) |

Klinisk presentation; bröstsmärta av infarktliknande karaktär, arytmier, hjärtsvikt eller överlevt hjärtstopp

*Eur Heart J*, ehaf192, https://doi.org/10.1093/eurheartj/ehaf192

**Table 5**

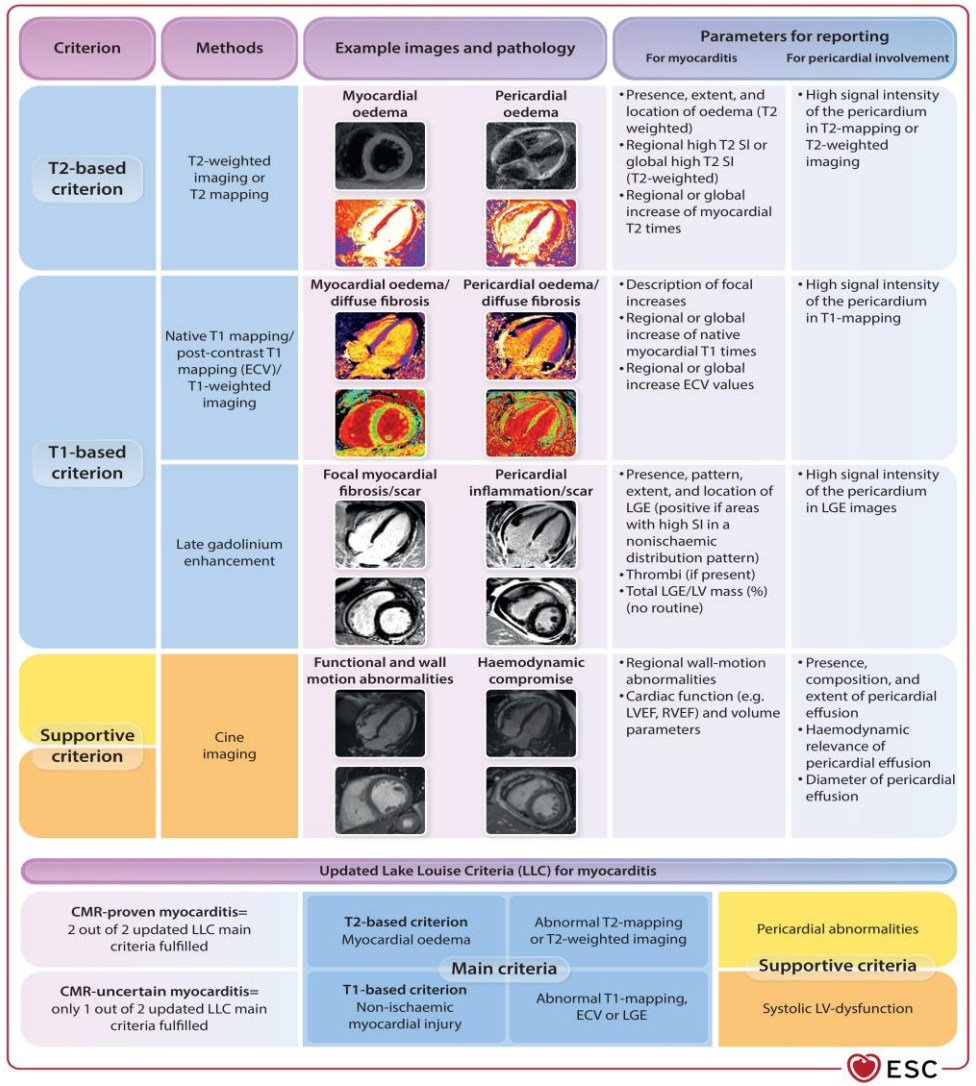
Histopathological criteria for myocarditis

| Term | Predominant inflammatory cells | Myocyte necrosis | Infections PCR positive (viruses, etc.) |
|---|---|---|---|
| **Active lymphocytic myocarditis** | $CD3^+$ T lymphocytes >7/$mm^2$, $CD68^+$ macrophages | yes | yes/no |
| **Persistent lymphocytic myocarditis** | $CD3^+$ T lymphocytes >7/$mm^2$, $CD68^+$ macrophages | yes | yes/no |
| **Resolved lymphocytic myocarditis** | – | no | yes/no |
| **Eosinophilic myocarditis (acute stage)** | Eosinophils, $CD3^+$ T lymphocytes, $CD68^+$ macrophages | yes | yes/no |
| **Giant-cell myocarditis (acute stage)** | Eosinophils, $CD68^+$ giant cells, $CD3^+$ T lymphocytes, $CD68^+$ macrophages | yes | no |
| **Sarcoidosis** | $CD68^+$ giant cells, granuloma, $CD3^+$ T lymphocytes, $CD68^+$ macrophages | yes/no | no |

CD68 används för att lokalisera makrofager och inflammatoriska svar.lokaliserat lysosmalt och i fagocytos

CD3 På cellytan på T-celler och delaktighet i aktivering av T-cellen

# Diagnoskriterier baseras på klinisk presentation med stödjande fynd; Perikardit




| IMPS | | |
|---|---|---|
| If diagnostic criteria for myocarditis and/or pericarditis are fulfilled[a] | | |
| | **Myocarditis** | **Pericarditis** |
| Definite | Clinical presentation[b] and CMR- or EMB-proven | Clinical presentation[b] with >1 additional criterion |
| Possible | Clinical presentation[b] with at least 1 additional criterion CMR- or EMB-uncertain or not available | Clinical presentation[b] with 1 additional criterion |
| Unlikely/rejected | Only clinical presentation[b] without additional criteria | Only clinical presentation[b] without additional criteria |
| Additional criteria beyond clinical presentations[b] | | |
| | **Myocarditis** | **Pericarditis** |
| Clinical[b] | Non-specific findings | Pericardial rubs |
| ECG[c] | ST-T changes | PR depression, widespread ST-segment elevation |
| Biomarkers | Troponin elevation | C-reactive protein elevation |
| Imaging[d] | Abnormal strain, wall motion, reduced EF<br>Myocardial oedema and/or LGE (CMR findings) | New or worsening pericardial effusion<br>Pericardial oedema and/or LGE (CMR findings) |

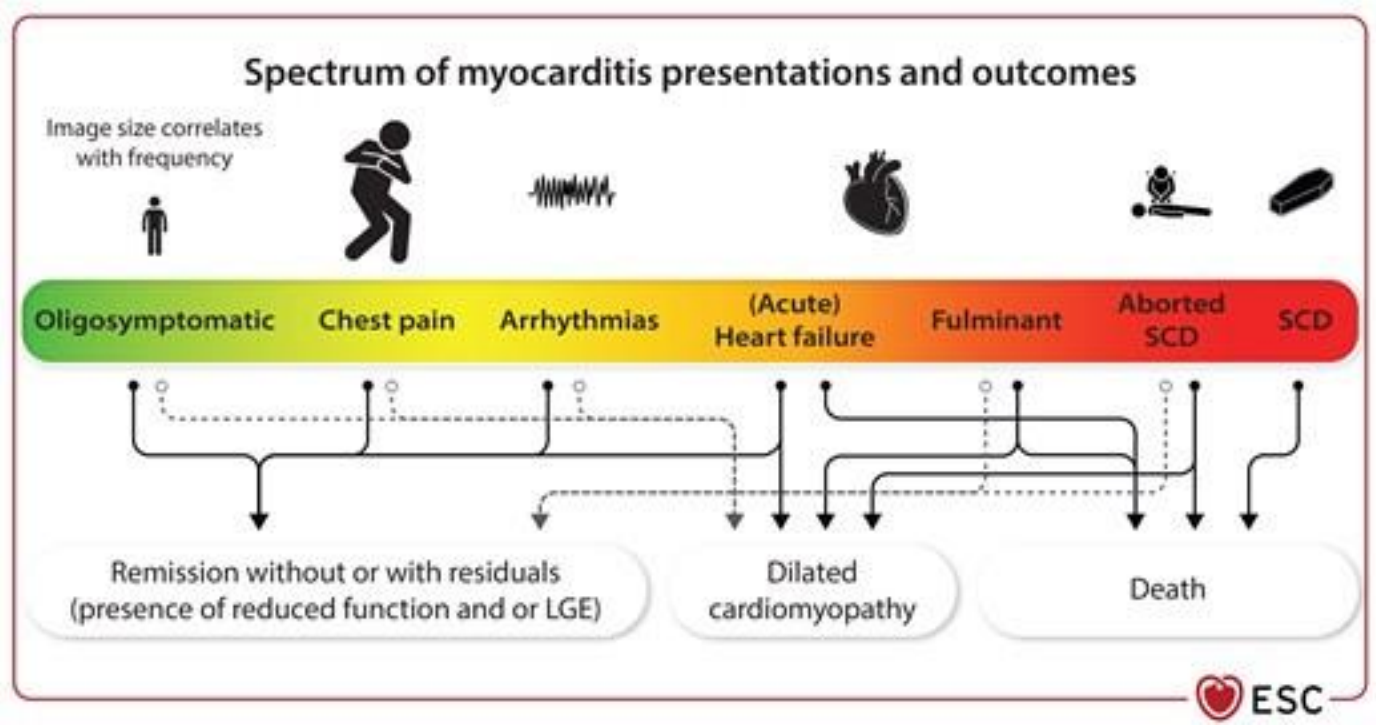
Klinisk presentation; bröstsmärta, oftast skarp och pleuritliknande. Förbättras vid uppesittande och framåtlutad och försämra vid liggande/inandning/sväljning. Andfåddhet. Arytmier är ovanligt men om arytmi, FF vanligast.

*Eur Heart J*, ehaf192, https://doi.org/10.1093/eurheartj/ehaf192

# Symptom och fynd som skall öka klinisk misstanke om IMPS

| Myocarditis | Pericarditis |
|---|---|
| Recent or concomitant flu-like syndrome or gastroenteritis | Recent or concomitant flu-like syndrome or gastroenteritis |
| Infarct-like chest pain | Pleuritic/infarct-like chest pain |
| Palpitations | Right HF symptoms and signs of constriction |
| HF symptoms | Fever |
| ECG changes[a] | Pericardial rubs |
| Ventricular arrhythmias (isolated, complex) | C-reactive protein elevation |
| Syncope | Pericardial effusion |
| Haemodynamic instability | Pleural effusion |
| Elevated markers of myocardial lesion (hs-Tn, CK-MB elevation) | Polyserositis |
| Elevated markers of HF (NT-proBNP) | CMR imaging with pericardial oedema and/or LGE |
| Abnormal wall motion, increased wall thickness and/or impaired systolic function on imaging | |
| CMR imaging with myocardial oedema and/or LGE | |

# Symptom och fynd som skall öka klinisk misstanke om IMPS



**Viral (main causes)**

*More common*: enteroviruses (including coxsackieviruses), parvovirus B19, human herpesvirus 6, Epstein–Barr virus, cytomegalovirus, herpes simplex virus 1/2, adenoviruses, influenza A and B viruses, coronaviruses, dengue virus

*Less common*: varicella-zoster virus, human immunodeficiency virus, hepatitis C virus, respiratory syncytial virus, mumps virus, measles virus, rubella virus, rabies virus

**Bacterial**

*More common*: *Mycobacterium tuberculosis* (pericarditis), *Coxiella burnetii*, *Borrelia* spp., *Campylobacter jejuni*

*Less common*: *Streptococcus* A, *Streptococcus pneumoniae* (pneumococcus), *Chlamydia pneumoniae*, *Corynebacterium diphtheriae*, *Legionella*, *Neisseria gonorrhoeae* (gonococcus), *Staphylococcus*, *Salmonella*, *Haemophilus influenzae*, *Brucella*, *Mycoplasma pneumoniae*, *Neisseria meningitidis* (meningococcus), *Leptospira* (Weil's disease), *Rickettsia rickettsii* (Rocky Mountain spotted fever)

**Fungal**

*Histoplasma capsulatum*, *Aspergillus* spp., *Blastomyces dermatitidis*, *Candida* spp., *Actinomyces*, *Cryptococcus*, *Nocardia* spp., *Coccidioides*, *Sporothrix*, mucormycosis

**Parasitic/worms**

*Trypanosoma cruzi*, *Toxoplasma gondii*, *Trichinella spiralis*, *Taenia solium*, *Entamoeba histolytica*, *Leishmania*, *Echinococcus*, *Toxocara canis*

**Autoimmune diseases**

Systemic:

Systemic lupus erythematosus, Sjögren's syndrome, rheumatoid arthritis, scleroderma, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, sarcoidosis, inflammatory bowel disease, allergic granulomatosis, Horton disease, Takayasu disease, Behçet syndrome, familial Mediterranean fever, tumour necrosis factor receptor-associated periodic syndrome, Kawasaki's disease, IgG4-related disease, antiphospholipid syndrome

Organ-specific (mainly myocarditis):

Lymphocytic myocarditis, giant-cell myocarditis, eosinophilic myocarditis, isolated cardiac sarcoidosis

**Immune reactions to drugs or vaccines**

Anthracyclines, procainamide, busulfan, hydralazine, methyldopa, isoniazid, phenytoin, immune checkpoint inhibitors (such as pembrolizumab and nivolumab ± ipilimumab), tyrosine kinase inhibitors, doxorubicin, daunorubicin, cytosine arabinoside, cytarabine, 5-fluorouracile, cyclophosphamide, penicillin, ampicillin, cephalosporins, tetracyclines, phenylbutazone, thiazides, p-amino salicylic acid, sulfa drugs, several vaccines, methysergide, mesalazine, clozapine, minoxidil, dantrolene, practolol, streptomycin, thiouracils, streptokinase, bromocriptine, GM-CSF, anti-TNF agents, minocycline and carbamazepine, CAR T-cell therapies

**Specific for pericarditis**

Post-cardiac injury syndromes:

Post pericardiectomy syndrome, post-myocardial infarction, post-traumatic (iatrogenic/non-iatrogenic)

Neoplastic (usually pericardial effusion without pericarditis):

Primary tumours (e.g. pericardial mesothelioma), secondary metastatic tumours

Metabolic:

Uraemia, cholesterol pericarditis

Miscellaneous:

Radiotherapy

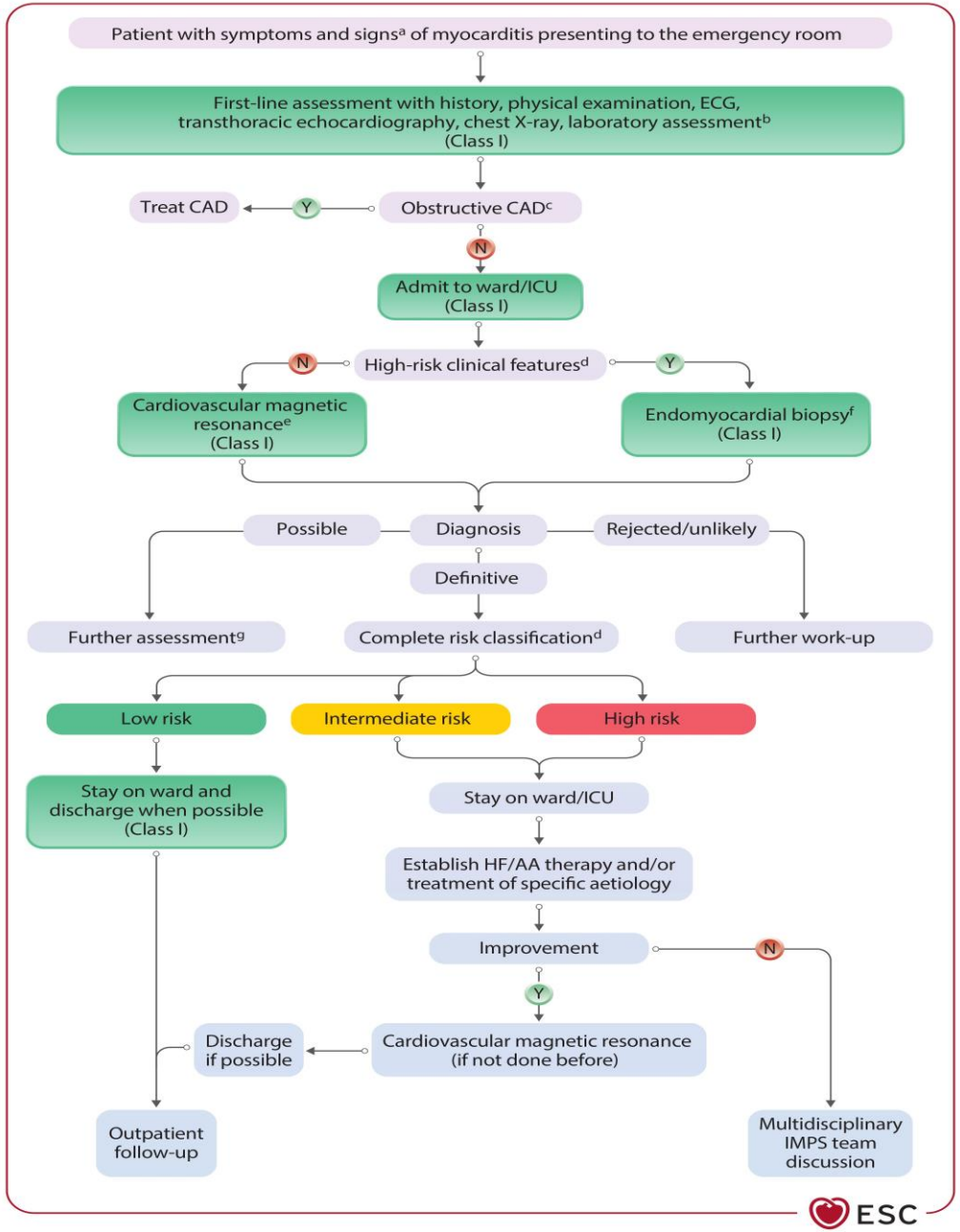
**Specific for myocarditis**

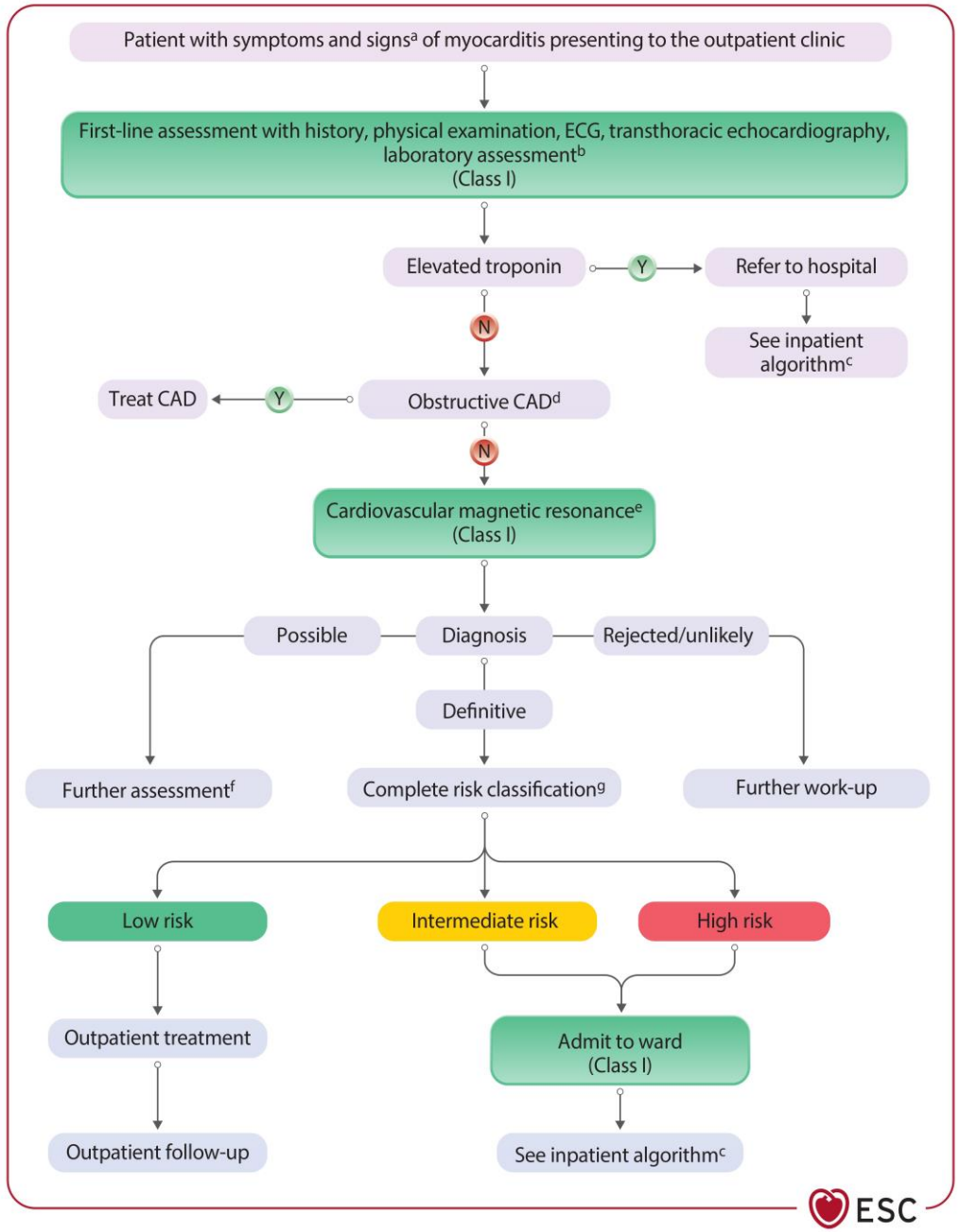
Toxic:

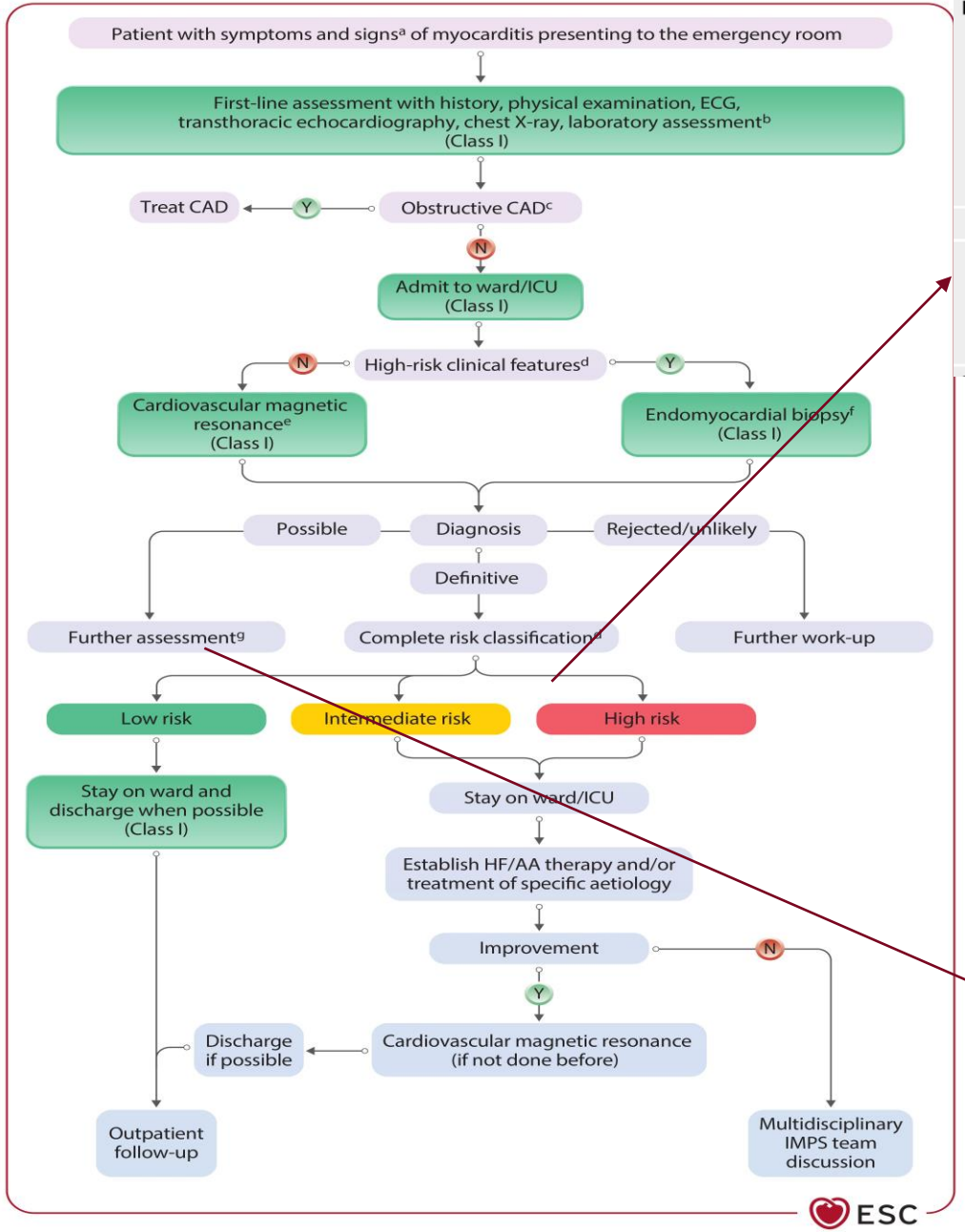
Ethanol, cocaine, amphetamines

Miscellaneous:

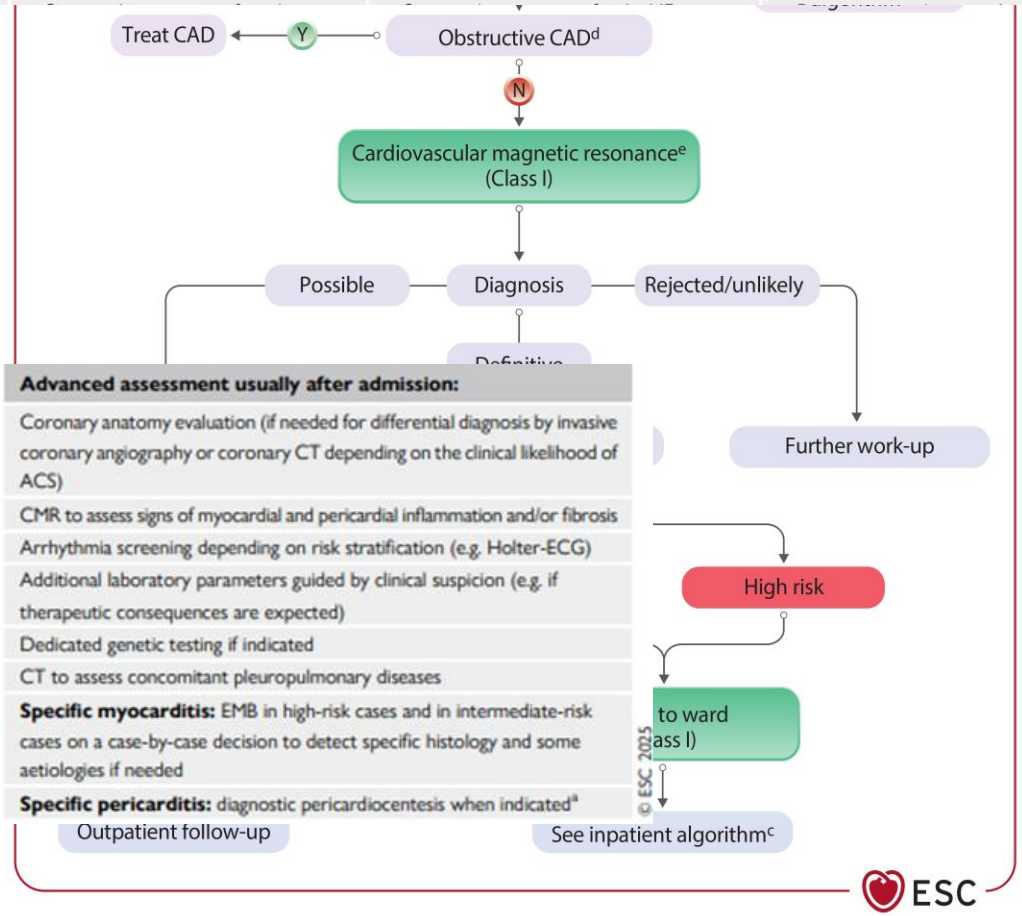
Thyrotoxicosis, arsenic, copper, iron


Patient with symptoms and signs[a] of myocarditis presenting to the emergency room
First-line assessment with history, physical examination, ECG, transthoracic echocardiography, chest X-ray, laboratory assessment[b] (Class I)
Obstructive CAD[c]
Y
Treat CAD
N
Admit to ward/ICU (Class I)
High-risk clinical features[d]
N
Y
Cardiovascular magnetic resonance[e] (Class I)
Endomyocardial biopsy[f] (Class I)
Diagnosis
Possible
Rejected/unlikely
Definitive
Further assessment[g]
Complete risk classification[d]
Further work-up
Low risk
Intermediate risk
High risk
Stay on ward and discharge when possible (Class I)
Stay on ward/ICU
Establish HF/AA therapy and/or treatment of specific aetiology
Improvement
N
Y
Discharge if possible
Cardiovascular magnetic resonance (if not done before)
Outpatient follow-up
Multidisciplinary IMPS team discussion
ESC

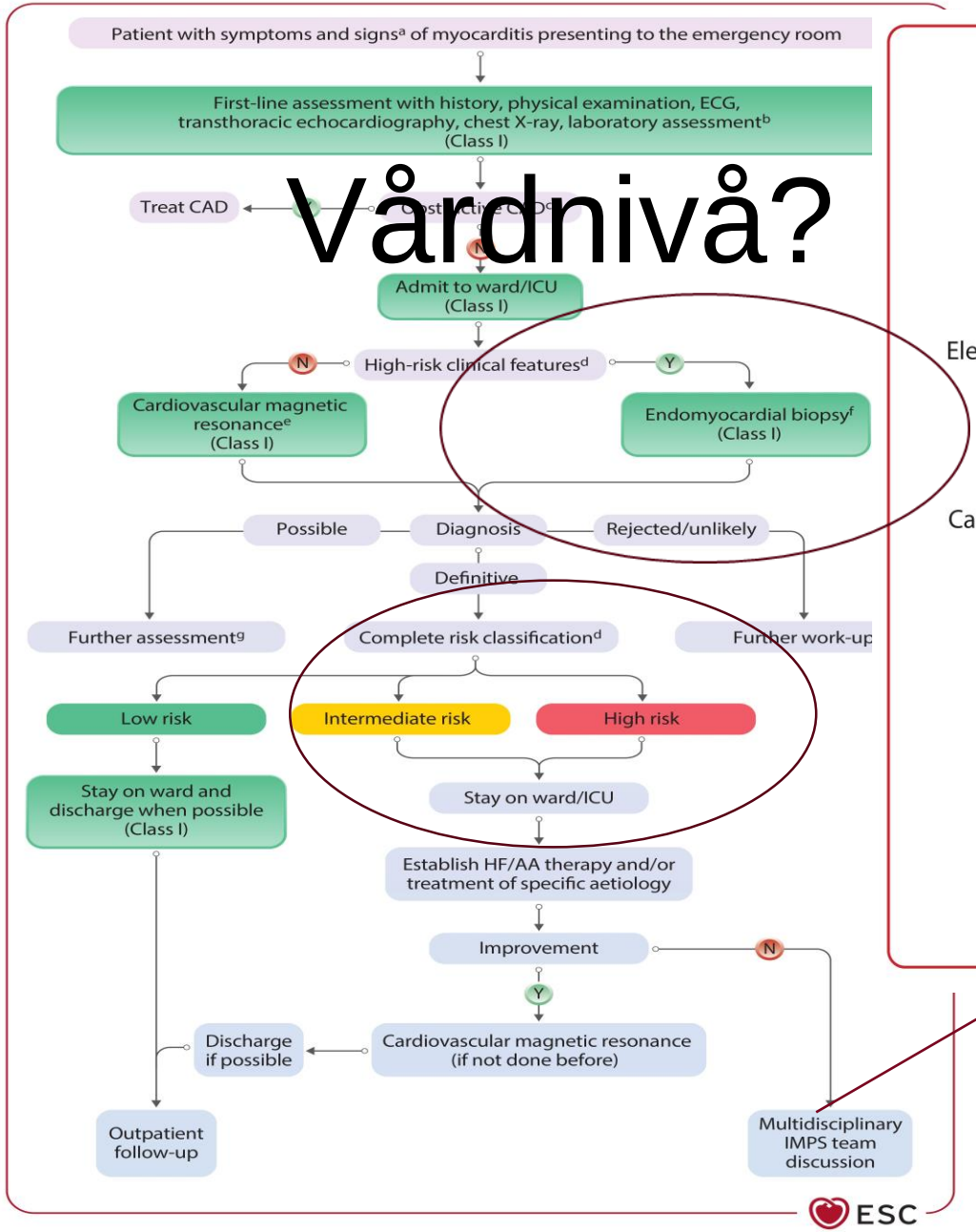

Patient with symptoms and signs[a] of myocarditis presenting to the outpatient clinic
First-line assessment with history, physical examination, ECG, transthoracic echocardiography, laboratory assessment[b] (Class I)
Elevated troponin
Y
Refer to hospital
See inpatient algorithm[c]
N
Treat CAD
Y
Obstructive CAD[d]
N
Cardiovascular magnetic resonance[e] (Class I)
Diagnosis
Possible
Rejected/unlikely
Definitive
Further assessment[f]
Complete risk classification[g]
Further work-up
Low risk
Intermediate risk
High risk
Outpatient treatment
Admit to ward (Class I)
Outpatient follow-up
See inpatient algorithm[c]
ESC

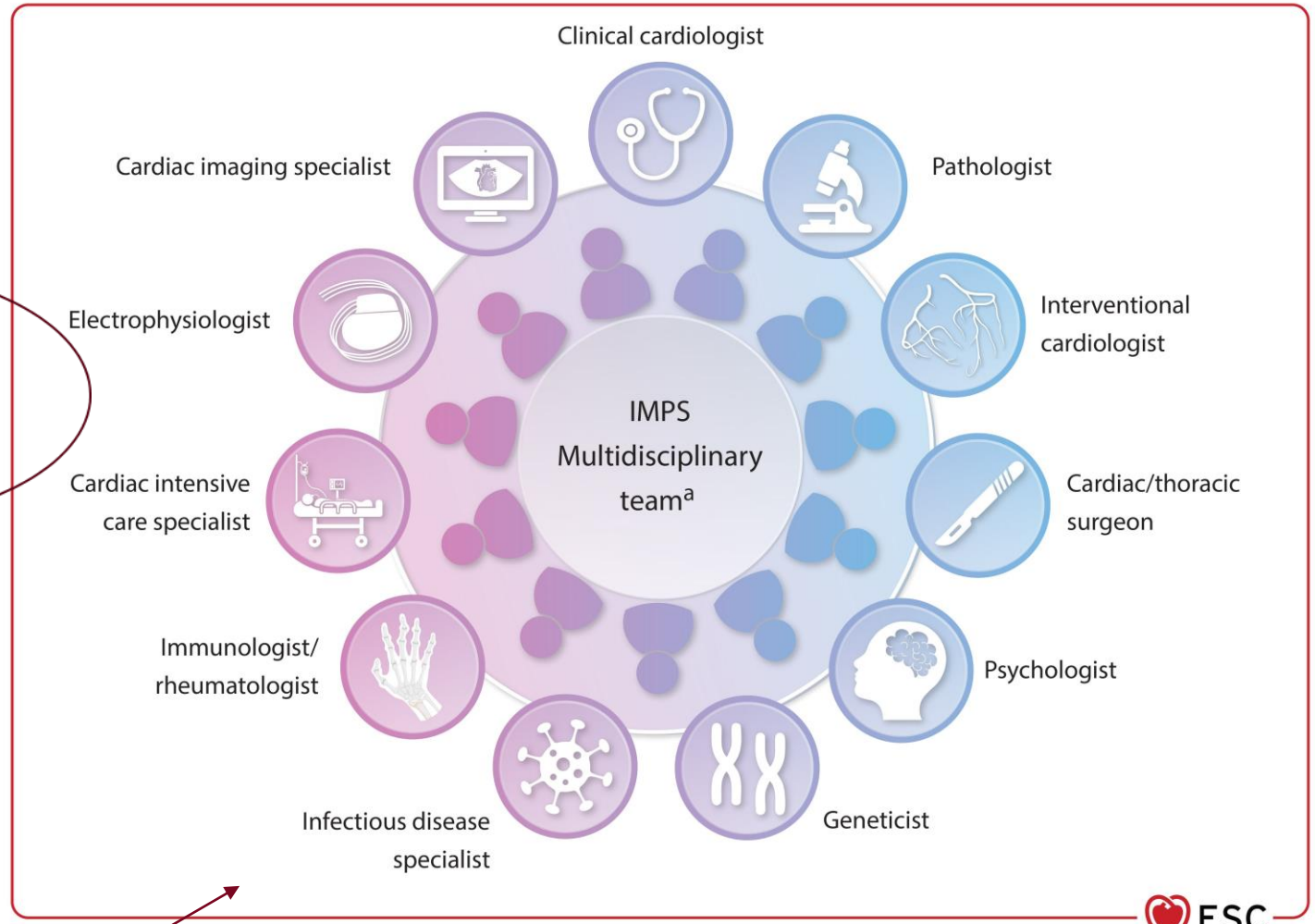



| Risk | High risk | Intermediate risk | Low risk |
|---|---|---|---|
| **Myocarditis** | • Acute HF/cardiogenic shock<br>• Dyspnoea NYHA III–IV refractory to medical therapy<br>• Cardiac arrest/syncope[a]<br>• Ventricular fibrillation/sustained ventricular tachycardia[a]<br>• High-level AV block[a] | • New/progressive dyspnoea<br>• Non-sustained ventricular arrhythmias<br>• Persistent release or relapsing troponin | Stable symptoms or oligosymptomatic |
| | **Imaging criteria:** | **Imaging criteria:** | **Imaging criteria:** |
| | • Newly reduced LVEF (<40%)[a]<br>• Extensive LGE on CMR[a] | • Newly mildly reduced LVEF (41%–49%) and/or WMA<br>• Preserved LVEF (≥50%) and LGE ≥2 segments on CMR | • Preserved LVEF (≥50%) without LGE or limited LGE (<2 segments) on CMR |

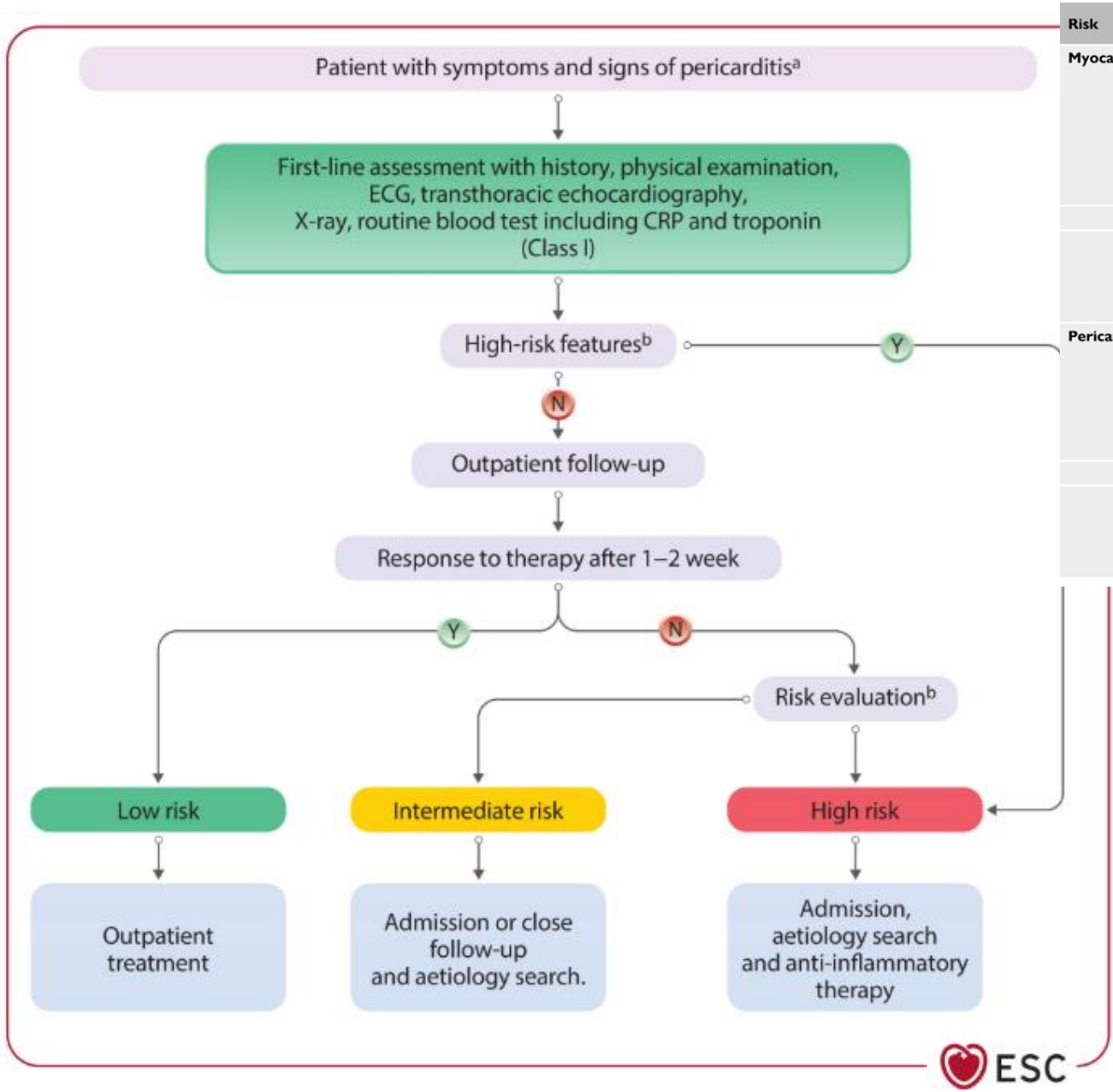


# Vårdnivå?

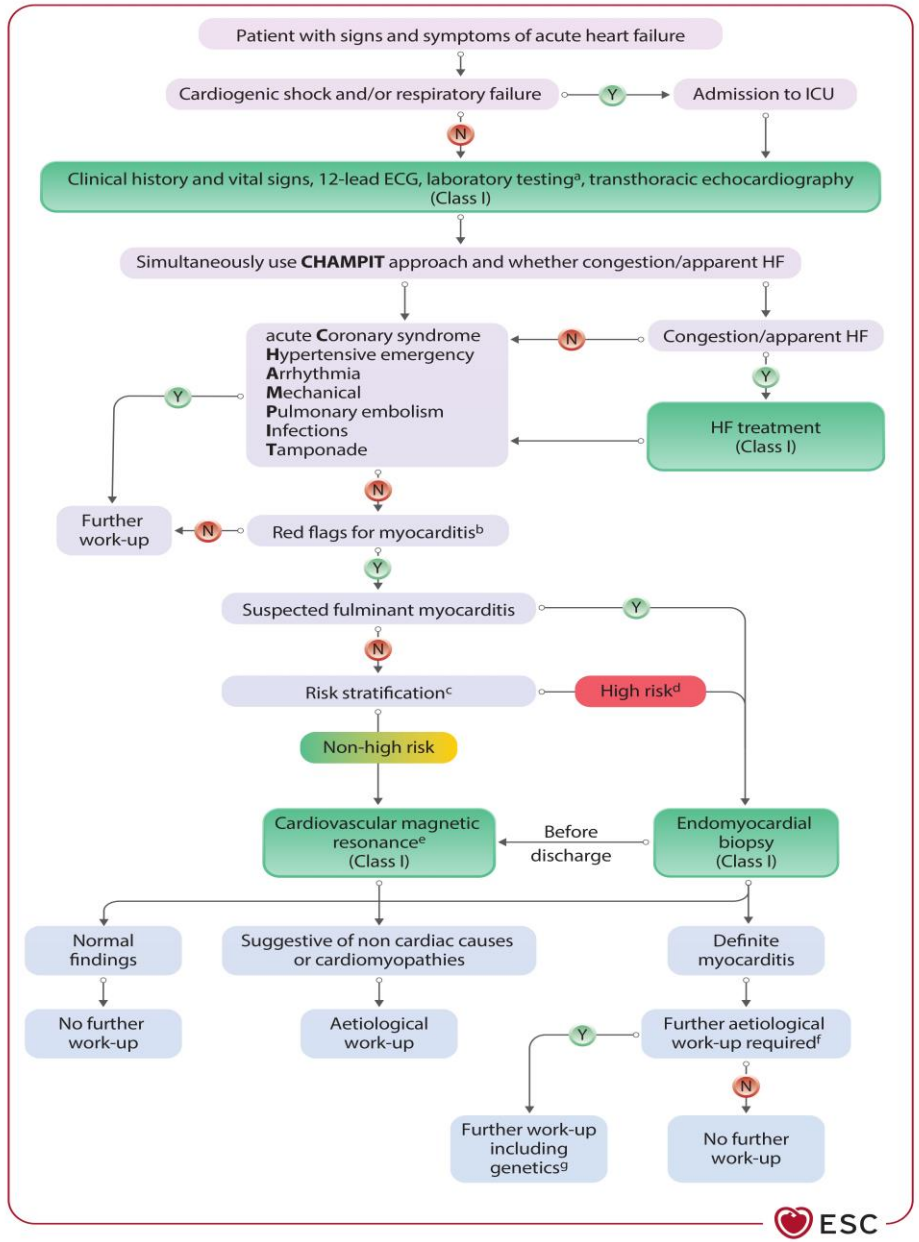

Patient with symptoms and signs[a] of myocarditis presenting to the emergency room
First-line assessment with history, physical examination, ECG, transthoracic echocardiography, chest X-ray, laboratory assessment[b] (Class I)
Treat CAD
Admit to ward/ICU (Class I)
N
Y
High-risk clinical features[d]
Cardiovascular magnetic resonance[e] (Class I)
Endomyocardial biopsy[f] (Class I)
Possible
Diagnosis
Rejected/unlikely
Definitive
Further assessment[g]
Complete risk classification[d]
Further work-up
Low risk
Intermediate risk
High risk
Stay on ward and discharge when possible (Class I)
Stay on ward/ICU
Establish HF/AA therapy and/or treatment of specific aetiology
Improvement
Discharge if possible
Cardiovascular magnetic resonance (if not done before)
Outpatient follow-up
Multidisciplinary IMPS team discussion
ESC

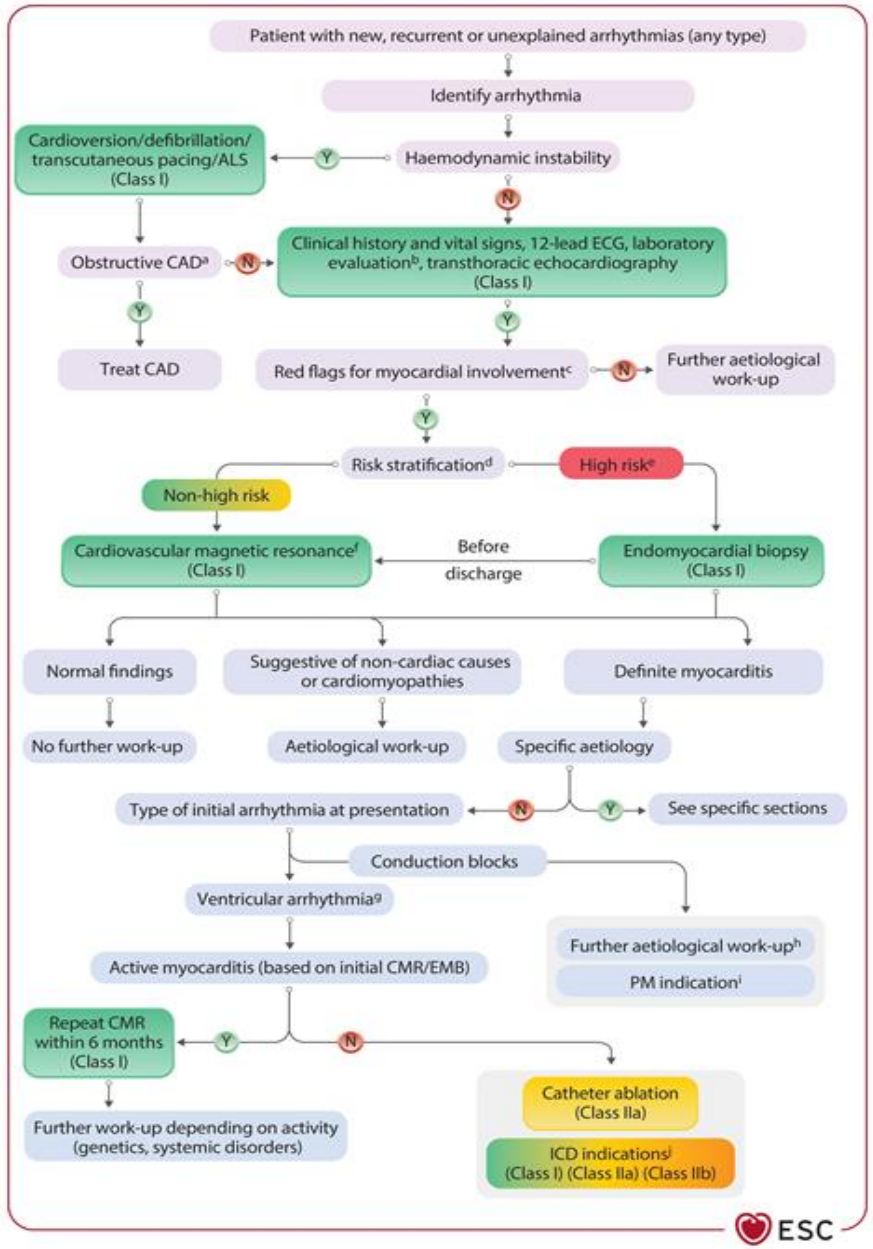

Clinical cardiologist
Cardiac imaging specialist
Pathologist
Electrophysiologist
Interventional cardiologist
IMPS Multidisciplinary team[a]
Cardiac intensive care specialist
Cardiac/thoracic surgeon
Immunologist/ rheumatologist
Psychologist
Infectious disease specialist
Geneticist
ESC


Outpatient treatment
Outpatient follow-up
Admit to ward (Class I)
See inpatient algorithm[c]
ESC



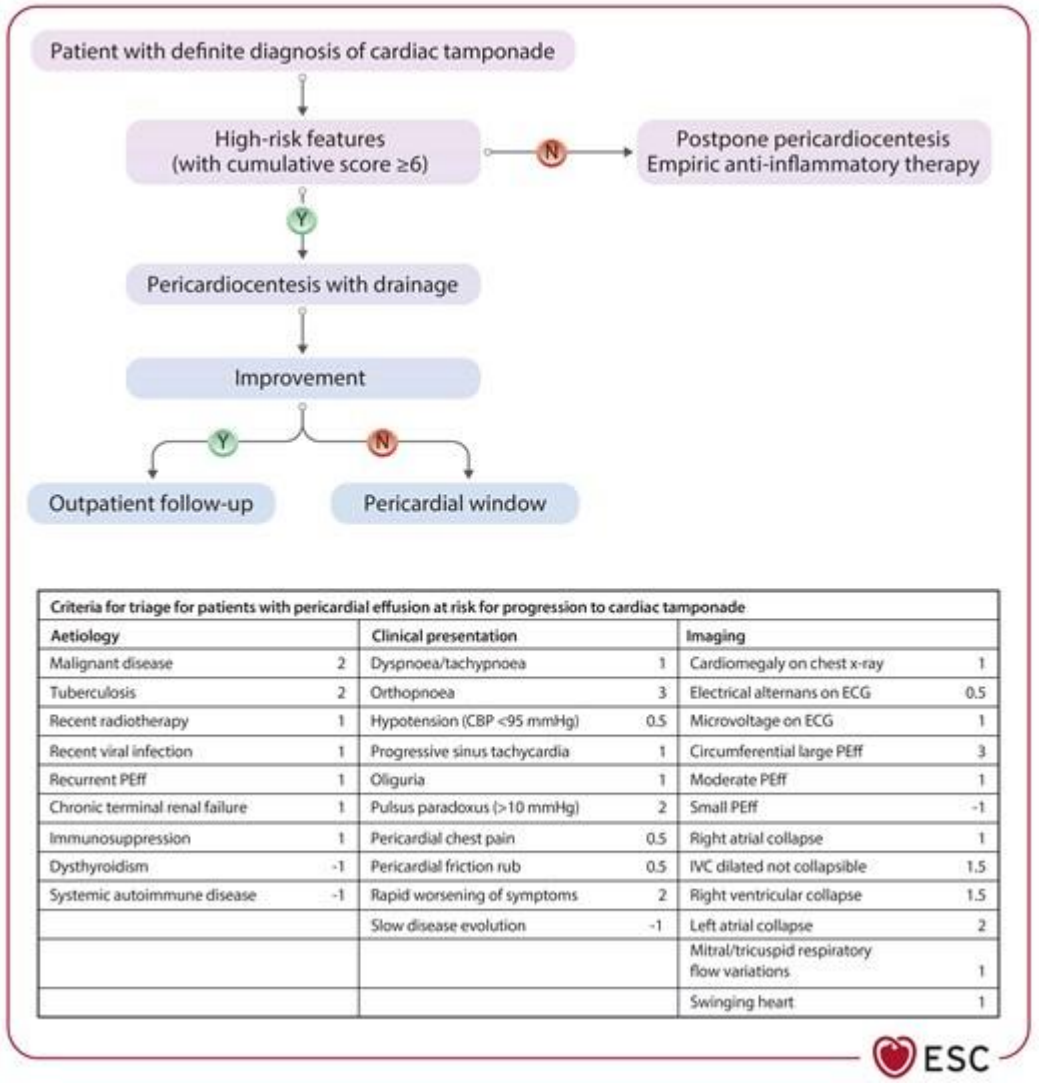

| Risk | High risk | Intermediate risk | Low risk |
|---|---|---|---|
| **Myocarditis** | • Acute HF/cardiogenic shock<br>• Dyspnoea NYHA III–IV refractory to medical therapy<br>• Cardiac arrest/syncope[a]<br>• Ventricular fibrillation/sustained ventricular tachycardia[a]<br>• High-level AV block[a] | • New/progressive dyspnoea<br>• Non-sustained ventricular arrhythmias<br>• Persistent release or relapsing troponin | Stable symptoms or oligosymptomatic |
| | **Imaging criteria:** | **Imaging criteria:** | **Imaging criteria:** |
| | • Newly reduced LVEF (<40%)[a]<br>• Extensive LGE on CMR[a] | • Newly mildly reduced LVEF (41%–49%) and/or WMA<br>• Preserved LVEF (≥50%) and LGE ≥2 segments on CMR | • Preserved LVEF (≥50%) without LGE or limited LGE (<2 segments) on CMR |
| **Pericarditis** | • Signs and symptoms of cardiac tamponade<br>• Fever (temperature >38°C)<br>• Effusive–constrictive pericarditis<br>• Failure of NSAID therapy<br>• Incessant pericarditis | • Signs and symptoms of right HF | • Response to adequate therapy within 1–2 weeks |
| | **Imaging criteria:** | **Imaging criteria:** | **Imaging criteria:** |
| | • Large PEff (>20 mm end-diastole)<br>• Cardiac tamponade<br>• Extensive pericardial LGE on CMR | • Moderate–large PEff (10–20 mm end-diastole)<br>• Constrictive physiology regardless of the size of the effusion | • Absence or mild PEff<br>• Absence of pericardial LGE on CMR |


Patient with signs and symptoms of acute heart failure
Cardiogenic shock and/or respiratory failure
Y
Admission to ICU
N
Clinical history and vital signs, 12-lead ECG, laboratory testing[a], transthoracic echocardiography (Class I)
Simultaneously use **CHAMPIT** approach and whether congestion/apparent HF
acute **C**oronary syndrome
**H**ypertensive emergency
**A**rrhythmia
**M**echanical
**P**ulmonary embolism
**I**nfections
**T**amponade
N
Congestion/apparent HF
Y
HF treatment (Class I)
Y
N
Further work-up
N
Red flags for myocarditis[b]
Y
Suspected fulminant myocarditis
Y
N
Risk stratification[c]
High risk[d]
Non-high risk
Cardiovascular magnetic resonance[e] (Class I)
Before discharge
Endomyocardial biopsy (Class I)
Normal findings
Suggestive of non cardiac causes or cardiomyopathies
Definite myocarditis
No further work-up
Aetiological work-up
Y
Further aetiological work-up required[f]
N
Further work-up including genetics[g]
No further work-up
ESC


Patient with new, recurrent or unexplained arrhythmias (any type)
Identify arrhythmia
Cardioversion/defibrillation/ transcutaneous pacing/ALS (Class I)
Y
Haemodynamic instability
N
Obstructive CAD[a]
N
Clinical history and vital signs, 12-lead ECG, laboratory evaluation[b], transthoracic echocardiography (Class I)
Y
Y
Treat CAD
Red flags for myocardial involvement[c]
N
Further aetiological work-up
Y
Risk stratification[d]
High risk[e]
Non-high risk
Cardiovascular magnetic resonance[f] (Class I)
Before discharge
Endomyocardial biopsy (Class I)
Normal findings
Suggestive of non-cardiac causes or cardiomyopathies
Definite myocarditis
No further work-up
Aetiological work-up
Specific aetiology
Type of initial arrhythmia at presentation
N
Y
See specific sections
Conduction blocks
Ventricular arrhythmia[g]
Active myocarditis (based on initial CMR/EMB)
Further aetiological work-up[h]
PM indication[i]
Repeat CMR within 6 months (Class I)
Y
N
Catheter ablation (Class IIa)
ICD indications[j] (Class I) (Class IIa) (Class IIb)
Further work-up depending on activity (genetics, systemic disorders)
ESC

# Perikardit

- Positionell bröstsmärta, typiska besvär. Hos hälften PEX.
- RHF-symptom vid konstriktion.
- Torr perikardit:50%
- ”blöt” 50-60% PEX. Måttliga till stora mängder bör ge misstanke om icke-ideopatisk etiologi
- Blöt-kontriktiv: Stelt perikard med tillkomst av PEX. RA-tryck sjunker mer än 50% eller närmre 0mmHg vid perikardiocentes
- Kontriktiv perikardit och tamponad är allvarligaste komplikationerna.




**Criteria for triage for patients with pericardial effusion at risk for progression to cardiac tamponade**

| Aetiology | | Clinical presentation | | Imaging | |
|---|---|---|---|---|---|
| Malignant disease | 2 | Dyspnoea/tachypnoea | 1 | Cardiomegaly on chest x-ray | 1 |
| Tuberculosis | 2 | Orthopnoea | 3 | Electrical alternans on ECG | 0.5 |
| Recent radiotherapy | 1 | Hypotension (CBP <95 mmHg) | 0.5 | Microvoltage on ECG | 1 |
| Recent viral infection | 1 | Progressive sinus tachycardia | 1 | Circumferential large PEff | 3 |
| Recurrent PEff | 1 | Oliguria | 1 | Moderate PEff | 1 |
| Chronic terminal renal failure | 1 | Pulsus paradoxus (>10 mmHg) | 2 | Small PEff | -1 |
| Immunosuppression | 1 | Pericardial chest pain | 0.5 | Right atrial collapse | 1 |
| Dysthyroidism | -1 | Pericardial friction rub | 0.5 | IVC dilated not collapsible | 1.5 |
| Systemic autoimmune disease | -1 | Rapid worsening of symptoms | 2 | Right ventricular collapse | 1.5 |
| | | Slow disease evolution | -1 | Left atrial collapse | 2 |
| | | | | Mitral/tricuspid respiratory flow variations | 1 |
| | | | | Swinging heart | 1 |

ESC

**Table 8**

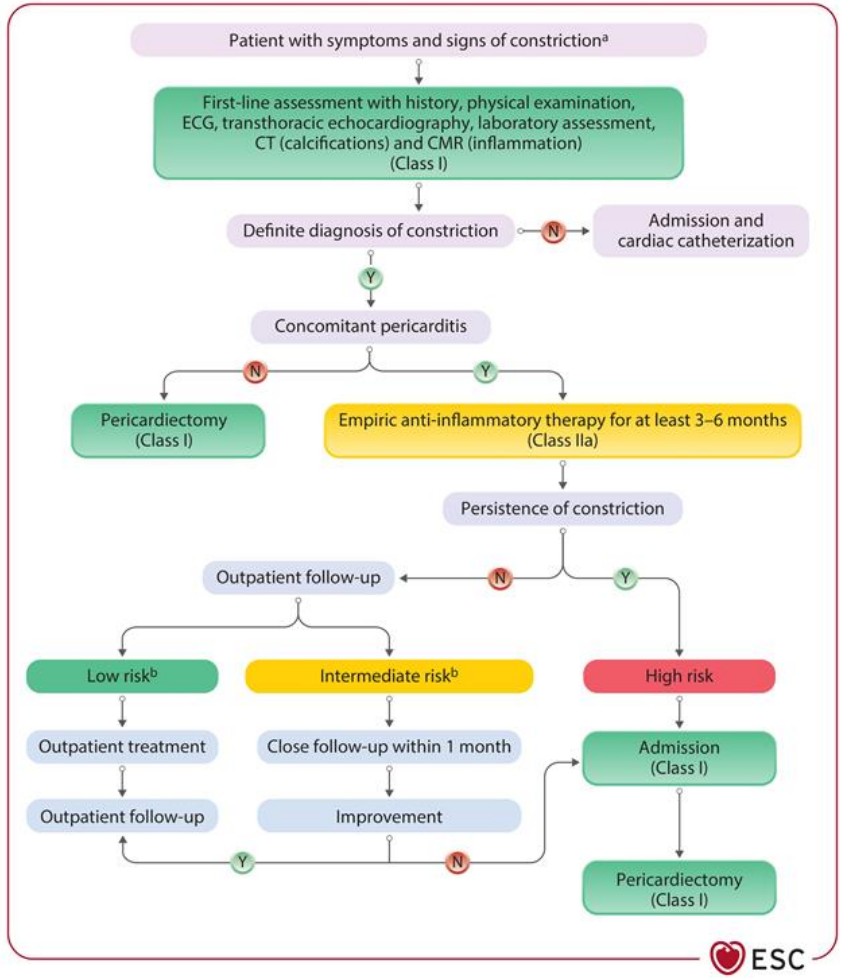
Causes of cardiac tamponade

| |
|---|
| Common causes (in order of relative frequency):<br>1. Neoplasm/malignancy<br>2. Iatrogenic/trauma<br>3. Pericarditis<br>4. Tuberculosis (most common in developing countries) |
| Less common causes (in order of relative frequency):<br>1. Collagen vascular diseases (systemic lupus erythematosus, rheumatoid arthritis, scleroderma)<br>2. Pericardial injury syndrome<br>3. Acute myocardial infarction<br>4. Aortic dissection<br>5. Uraemia<br>6. Bacterial infection<br>7. Pneumopericardium |

# Konstriktiv perikardit

Kroniskt tillstånd pga kalcifierat fibrotiskt perikardium som ger nedsatt diastolisk fyllnad.

Symptom som högersidig hjärtsvikt, dyspné, ascites, bensvullnad, ortopne, trötthet. Ofta väsentligen normala nivåer av NT-pro BNP.

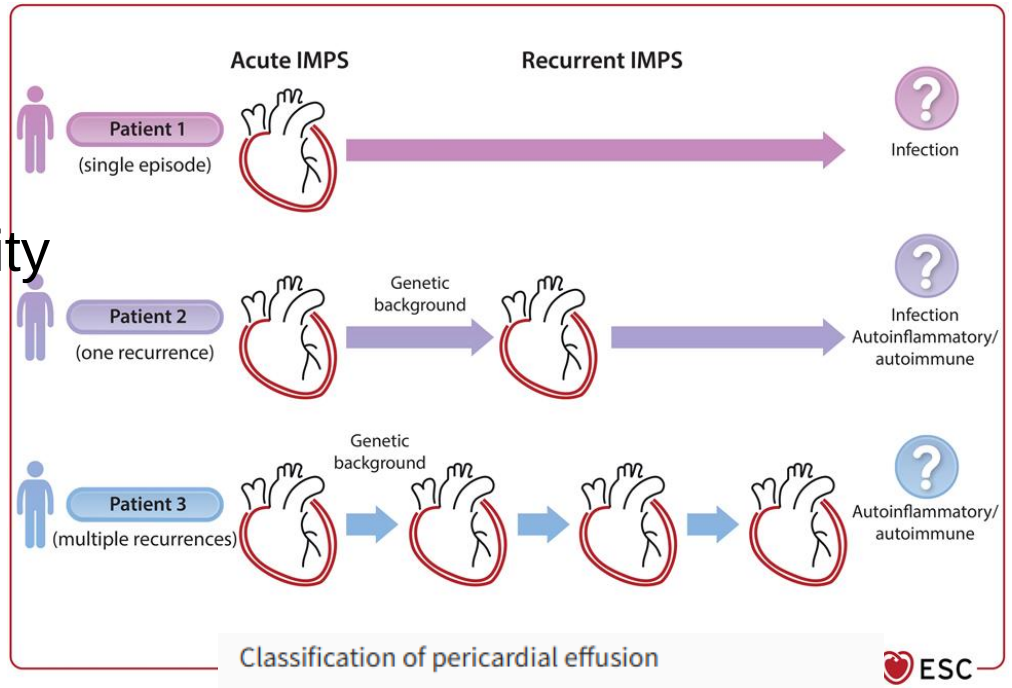



Generellt perikardit:
Fenotyp? Inflammatorisk? Icke-inflammatorisk?
Vad driver perikarditen?CRP-negativa oftast svårare att behandla. Interferon-drivet?

# Handläggning av IMPS

- Anamnes, status, riskvärdering, EKG, Multimodality imaging. Virusserologi är ej att rekommendera förutom HIV/hep C. Bakteriell analys utifrån symptom/sjukhistoria.
- Lungrtg
- UKG/EKO är första imagingmetoden inkl strain
- Troponin, CRP, SR, blodstatus inkl diff+elstatus, NT-pro BNP, TSH, leverprover (ESC basala utredninsgprover initialt.)

Avancerade metoder: Kranskärlsvärdering (CT? Angio), MRT, specifik provtagning, Biopsi! Gentest hos vissa individer, tidigare ICD-implantation? CT? IL1? TNF? IL6? Autoantikroppar? PET-CT?




Classification of pericardial effusion

ESC

| | |
|---|---|
| Onset | Acute (≤4 weeks) |
| | Subacute (>4 weeks to ≤3 months) |
| | Chronic (>3 months) |
| Size[a] | Mild: <10 mm |
| | Moderate: 10–20 mm |
| | Large: >20 mm |
| Distribution | Circumferential/loculated |
| Composition | Transudate/exudate |

[a]Maximal end-diastolic diameter.

# MR, MR, MR, MR, CT, PET

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| **Myocarditis** | | |
| CMR is recommended in patients with suspected myocarditis to reach a clinical diagnosis and to determine the cause of acute myocardial injury, including assessment of oedema, ischaemia, and necrosis/fibrosis/scarring.[115,164,169–183] | I | B |
| CMR is recommended for follow-up at least within the first 6 months in patients with myocarditis to identify a healed or ongoing process, for risk stratification and personalized therapy, and to enable a return to exercise.[10,62,184–186] | I | C |
| **Pericarditis** | | |
| CMR is recommended in patients with suspected pericarditis when a diagnosis cannot be made using clinical criteria to assess evidence of pericardial thickening, oedema, LGE, and to assess the persistence of disease during follow-up in selected cases.[110,129,165,187–194] | I | B |



CMR, cardiovascular magnetic resonance; LGE, late gadolinium enhancement.
[a]Class of recommendation.
[b]Level of evidence.

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| CT is recommended to evaluate pericardial thickness, calcifications, masses, and loculated pericardial effusions, as well as concomitant pleuropulmonary diseases and chest abnormalities.[193,197,199,200] | I | C |



| Recommendations | Class[a] | Level[b] |
|---|---|---|
| Carb-free $^{18}$F-FDG-PET or $^{18}$F-FDG-PET/CT should be considered for the diagnostic work-up in patients with suspected myocarditis and/or pericarditis in whom echocardiography and CMR are inconclusive for the clinical diagnosis.[201,206] | IIa | C |

# MR: Lämnar hela MR till Jan…

Kostnadseffektivitet

Undanträngning?

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| **Myocarditis** | | |
| CMR is recommended in patients with suspected myocarditis to reach a clinical diagnosis and to determine the cause of acute myocardial injury, including assessment of oedema, ischaemia, and necrosis/fibrosis/scarring.[115,164,169–183] | I | B |
| CMR is recommended for follow-up at least within the first 6 months in patients with myocarditis to identify a healed or ongoing process, for risk stratification and personalized therapy, and to enable a return to exercise.[10,62,184–186] | I | C |
| **Pericarditis** | | |
| CMR is recommended in patients with suspected pericarditis when a diagnosis cannot be made using clinical criteria to assess evidence of pericardial thickening, oedema, LGE, and to assess the persistence of disease during follow-up in selected cases.[110,129,165,187–194] | I | B |

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CMR, cardiovascular magnetic resonance; LGE, late gadolinium enhancement.
[a]Class of recommendation.
[b]Level of evidence.

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| CT is recommended to evaluate pericardial thickness, calcifications, masses, and loculated pericardial effusions, as well as concomitant pleuropulmonary diseases and chest abnormalities.[193,197,199,200] | I | C |

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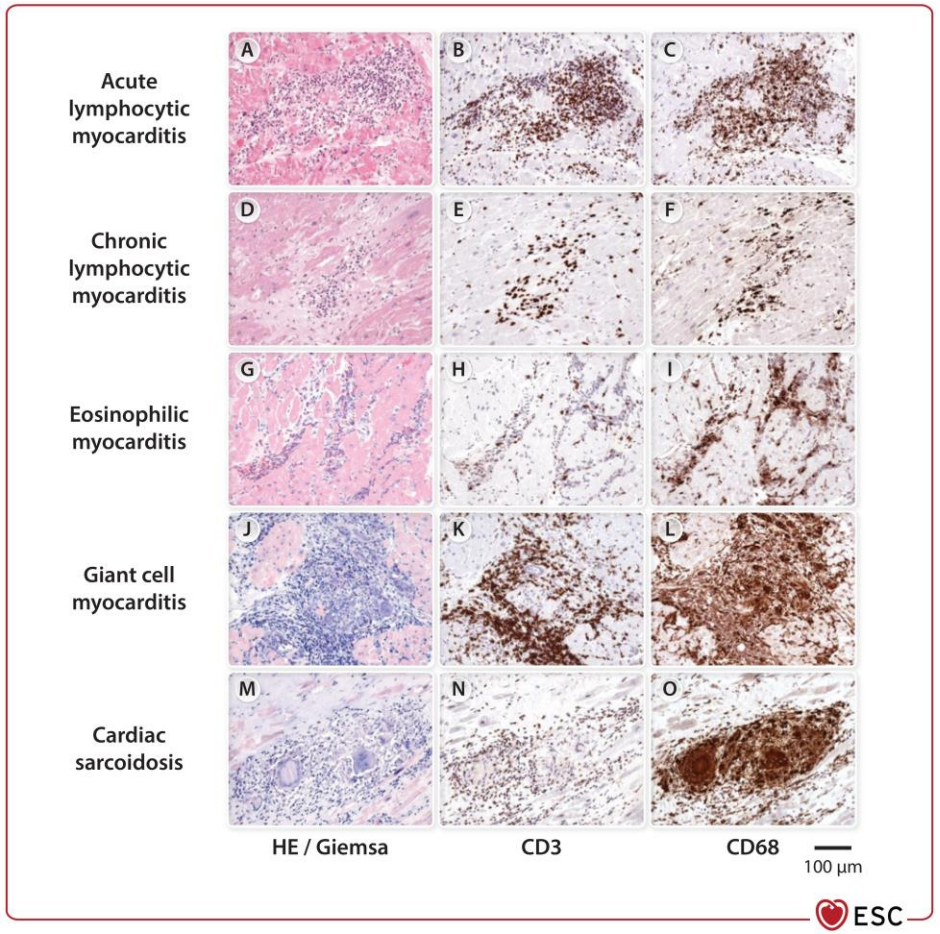
Kloka kliniska val?

Svår, tidskrävande undersökning för patient

Psykiskt belastande för patient

Leder undersökningarna till någon åtgärd?

# Biopsi; för högrisk och i vissa fall intermediär-risk




- Oftast i RV-septum
- Immunohistokemi ökar sannolikhet för fynd
- Möjlighet för virusdiagnostik
- Styr behandling
- Behov av snabbt svar
- Viss risk, skall göras av van personal
- Elektroanatomisk mapping kan förbättra biopsiresultat och kan hjälpa till att differentiera ARVC vs Sarkoidos

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| EMB[c] is recommended in patients with high-risk myocarditis[d], and/or haemodynamic instability, and/or in patients with intermediate-risk myocarditis not responding to conventional therapy in order to detect a specific histologic subtype and to assess the presence of viral genome for treatment.[34,63,73,131] | I | C |

# Mål med behandling

- Minska symptom och minska komplikationsrisk
- Behandla infektion
- Behandla inflammation
- Behandla immuno-medierade processer
- Behandla hjärtsvikt
- Behandla arytmier
- Behandla konstriktion

- Restriktion av fysisk aktivitet. Minst 1mån eller till normalisering av inflammation.
- Frånvaro av symptom, normaliserade prover, normaliserat EKG, frånvaro av perikardexsudat och frånvaro av inflammation på MR. Gradvis återgång och eventuellt arbetsprov och holter.
- Okomplicerad myokardit kan behandlas med NSAID eller ASA. Små studier visar ingen skada. Kolkicin är säkert vid myoperikardit.
- Viral behandling vid tydlig etiologi.
- Kortison vid inflammatorisk sjukdom

# Farmakologisk behandling

**Table 12**

Therapy for specific forms of myocarditis

| **Lymphocytic myocarditis (virus-negative)** | |
|---|---|
| 1st line therapy | Non-severe: prednisone 1 mg/kg/day p.o. then tapered<br>Severe: i.v. methylprednisolone 7–14 mg/kg/day for 3 days, then 1 mg/kg/day p.o. |
| 2nd line therapy | Oral corticosteroids + azathioprine[a] or mycophenolate mofetil[b], cyclosporine[c], methotrexate[d] |
| 3rd line therapy | IVIG[e] or plasmapheresis[f] |

| **Eosinophilic myocarditis** | |
|---|---|
| 1st line therapy | Same as lymphocytic myocarditis + Treat EM-associated condition if identified |
| 2nd line therapy | Same as lymphocytic myocarditis + Treat EM-associated condition if identified |
| 3rd line therapy | – |

| **Giant-cell myocarditis** | |
|---|---|
| 1st line therapy | Non-severe: prednisone 1 mg/kg/day p.o. then tapered<br>Severe: i.v. methylprednisolone 7–14 mg/kg/day for 3 days, then 1 mg/kg/day p.o. + immunosuppressive (azathioprine[a] or mycophenolate mofetil[b], cyclosporine[c]) |
| 2nd line therapy | Antithymocyte Globulin (ATG)[g] cyclophosphamide[h], rituximab[i] |
| 3rd line therapy | – |

| **Cardiac sarcoidosis** | |
|---|---|
| 1st line therapy | Non-severe: prednisone 1 mg/kg/day p.o., tapering from 40–60 mg daily<br>Severe: i.v. methylprednisolone 7–14 mg/kg/day for 3 days, then 1 mg/kg/day p.o. |
| 2nd line therapy | Methotrexate[d] (1st choice), or azathioprine[a] mycophenolate mofetil[b], cyclophosphamide[h] |
| 3rd line therapy | Infliximab[j] or adalimumab[k], rituximab[i] |

| **Lyme carditis** | |
|---|---|
| 1st line therapy | (a) Oral antibiotics (mild cases):<br>– Doxycycline 100 mg b.i.d. (14–21 days)<br>– Amoxicillin 500 mg t.i.d. (14–21 days)<br>– Cefuroxime axetil 500 mg b.i.d. (14–21 days)<br>(b) i.v. antibiotics (severe cases):<br>– Ceftriaxone 2 g/day (14–21 days) |
| 2nd line therapy | i.v. antibiotics:<br>Cefotaxime (2 g q8h × 14–21 days) or Penicillin G (18–24 MU/day i.v. q4h × 14–21 day) |
| 3rd line therapy | – |

| **Chagas disease** | |
|---|---|
| 1st line therapy | Benznidazole 5–7 mg/kg/day in 2 doses for 60 days<br>Nifurtimox 8–10 mg/kg/day in 3 doses for 60–90 days |
| 2nd line therapy | – |
| 3rd line therapy | – |

| **ICI-induced myocarditis** | |
|---|---|
| 1st line therapy | Withdraw ICI, reassess<br>Non-severe: methylprednisolone 500–1000 mg/day × 3 days, then taper with oral prednisone<br>Severe: i.v. methylprednisolone 7–14 mg/kg/day × 3 days, then 1 mg/kg/day |
| 2nd line therapy | If no response in 24–48 h: mycophenolate mofetil[b], ATG[g] abatacept[l], alemtuzumab[m] |
| 3rd line therapy | Infliximab[j] or adalimumab[k], rituximab[i] |

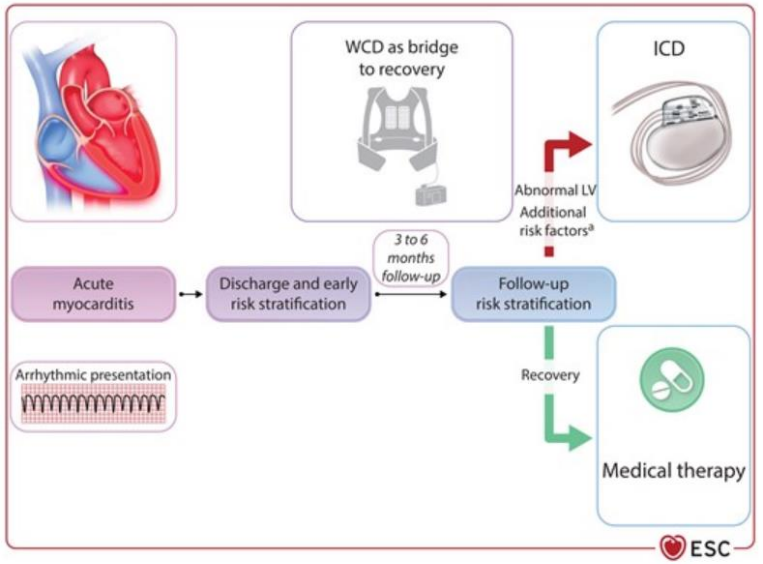
| Recommendations | Class[a] | Level[b] |
|---|---|---|
| **Management of symptoms** | | |
| NSAIDs (together with proton pump inhibition) should be considered in patients with associated symptoms of pericarditis to reduce symptoms. | IIa | C |
| Colchicine should be considered in patients with myopericarditis to reduce recurrences.[263] | IIa | B |
| **Management of heart failure** | | |
| Adherence to the ESC HF guidelines is recommended in cases of myocarditis with LV systolic dysfunction and/or HF to reduce symptoms and to improve LV function.[12] | I | C |
| HF therapy should be considered in patients with myocarditis and LV systolic dysfunction for at least 6 months upon complete LV functional recovery to stabilize LV function. | IIa | C |
| **Management of arrhythmias** | | |
| β-Blockers, with a continuation for at least 6 months, should be considered in patients with acute myocarditis, especially those with troponin elevation, to control symptoms and prevent arrhythmias. | IIa | C |
| Anti-arrhythmic treatment should be considered in post-myocarditis patients with recurrent, symptomatic VT to reduce arrhythmic burden.[58] | IIa | C |
| **Immunosuppressive therapy** | | |
| Corticosteroids should be considered in patients with fulminant, non-infectious forms of myocarditis to stabilize the patients. | IIa | C |
| Corticosteroids may be considered in patients with acute myocarditis with impaired LVEF if refractory to standard HF therapy to stabilize patients. | IIb | C |
| Routine use of immunosuppressive therapy is not recommended in acute myocarditis with preserved LV function because no outcome benefit has been shown. | III | C |

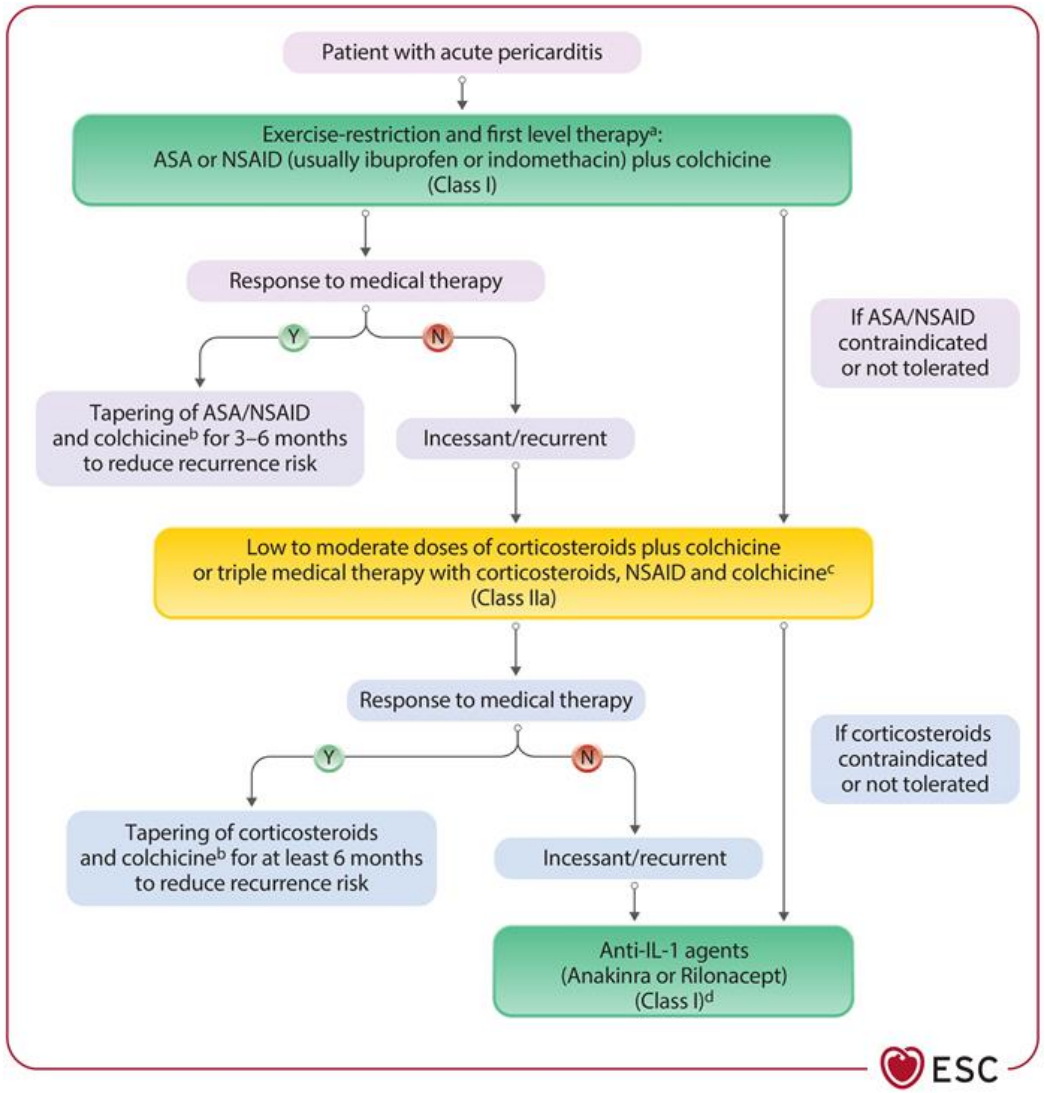
Majoriteten ca 50% återhämtar sig spontant vid akut myokardit
10-25% har en progressiv VK-dysfunktion
IL-5 antagonist?

Återkommande/kronisk myokardit; leta annan genes

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| **Pacing in myocarditis** | | |
| Temporary transvenous external pacing should be considered in patients with acute myocarditis and high-degree conduction disorders as a bridge to recovery.[86,317,335] | IIa | C |
| **WCD in myocarditis** | | |
| A WCD should be considered for 3–6 months in patients with sustained ventricular arrhythmia during the acute phase of myocarditis as a bridge to recovery.[323,325,327–330] | IIa | C |
| **Ablation in myocarditis** | | |
| Catheter ablation, performed in specialized centres, should be considered in post-myocarditis patients with recurrent symptomatic SMVT or ICD shocks in whom AAD are ineffective, not tolerated, or not desired.[58] | IIa | C |
| **ICD in myocarditis** | | |
| ***Secondary prevention*** | | |
| ICD implantation is recommended in patients with non-active[c] myocarditis and haemodynamically not-tolerated sustained VT to prevent SCD.[78,79,322,336] | I | C |
| ICD implantation should be considered in patients with non-active[c] myocarditis and haemodynamically tolerated sustained VT to prevent SCD.[78,79,322,336] | IIa | C |
| ICD implantation may be considered in patients with acute myocarditis and sustained VA (VT/VF) in the acute phase to prevent SCD.[71,79,89,222,323–325] | IIb | C |
| ***Primary prevention*** | | |
| ICD implantation may be considered in patients with myocarditis after the acute phase (3–6 months) and persistent risk factors for VA[d] to prevent SCD.[89,332–334,336.] | IIb | C |

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| A timely and dedicated Shock Team discussion is recommended in patients with myocarditis in the presence of haemodynamic compromise, to decide on the need for escalation to MCS and to determine a long-term management plan. | I | C |
| Temporary MCS[c] should be considered in patients with myocarditis and cardiogenic shock or acute decompensation in chronic myocarditis to stabilize the patients. | IIa | C |






| Therapy | Dosing | Duration[a] | Tapering[a] |
|---|---|---|---|
| Aspirin[b] | 750–1000 mg 3 times daily | 1–2 weeks | Decrease by 250 mg every 1–2 weeks |
| Ibuprofen[b] | 600–800 mg 3 times daily | 1–2 weeks | Decrease by 200 mg every 1–2 weeks |
| Indomethacin | 25–50 mg 3 times daily | 1–2 weeks | Decrease by 25 mg every 1–2 weeks |
| Colchicine[b] | 0.5 mg once daily (<70 kg or severe renal impairment) or 0.5 mg twice daily | 3–6 months | Not required |
| Prednisone | 0.2–0.5 mg/kg/day | 2–4 weeks | Several months |
| Treatment for recurrences only: | | | |
| Azathioprine | Starting with 1 mg/kg/day then gradually increased to 2–3 mg/kg/day | Several months | Several months |
| IVIG | 400–500 mg/kg i.v. daily for 5 days | 5 days | Not required |
| Anakinra | 1–2 mg/kg/day up to 100 mg/day in adults | At least 6 months/ >12 months | Needed (at least 3–6 months)/ unknown |
| Rilonacept[c] | 320 mg once daily followed by 160 mg weekly | | |

# Prognos

- VK-funktion vid insjuknandet
- Kön, kvinnor verkar ha bättre prognos
- Ung ålder och tidigare myokardit medför en sämre prognos för återfall.
- Fulminant myokardit och tydlig autoimmunitet, pos ANA har ökad risk för död och Htx
- Etiologi, Sarkoidos, jättecellsmyokardis och eosinofil myokardit har sämre prognos. QRS-bredd över 120ms.
- MR-fynd med LGE
- Ett fåtal kan utveckla dilaterad karidomyopti )50-94% normaliseras i VK-funktion
- Vissa specifika virus, HHV6, Parvovirus B19.
- Fulminant myokardit har 60dagars mortalitet på 24%

# Prognos fortsatt

- Återhämtning från dagar till månader. Okomplicerade patienter kan skrivas ut när enzymer närmar sig normalisering.
- Uppföljning är rekommenderat hos alla med myokardit
- Imaging; Rekommenderat hos alla; MR efter 6mån. Om första är onormal, ytterligare uppföljande MR

| | | Within 1 month | Within 3–6 months | 12 months | >1 year and long-term FU[a] |
|---|---|---|---|---|---|
| Clinical evaluation and ECG | Myocarditis | X | X | X | X |
| | Pericarditis | X | X | X | X |
| Biomarkers (TnI, C-reactive protein) | Myocarditis | X | X | (X) | (X) |
| | Pericarditis | X | X | (X) | (X) |
| Rhythm (stress and/or Holter-ECG) | Myocarditis | – | X | (X) | (X) |
| | Pericarditis | – | – | – | – |
| Imaging myocarditis | TTE | X[b] | | X[c] | X[c] |
| | CMR | X[b] | | X[c] | X[c] |
| Imaging pericarditis | TTE | X[b] | | X[c] | X |
| | CMR | (X)[b] | | (X)[d] | (X)[d] |

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| Follow-up with clinical assessment, biomarkers[c], ECG, exercise test, Holter-ECG monitoring, echocardiography, and CMR at least within 6 months after the index hospitalization is recommended in all patients with myocarditis to identify a potential progression or new risk factors.[62] | I | C |
| Long-term follow-up is recommended for patients with complicated myocarditis[d] to identify a potential progression or new complications.[28,74] | I | C |
| Long-term follow-up is recommended for patients with incessant or recurrent pericarditis to identify a potential progression and new complications.[104,347] | I | C |

# Inflammatorisk IMPS

- Dr. Lucchetti guidar er igenom detta imorgon. (hoppas jag)
- De med inflammatorisk kardiomyopati behandlas i enlighet med svikt-guidelines.

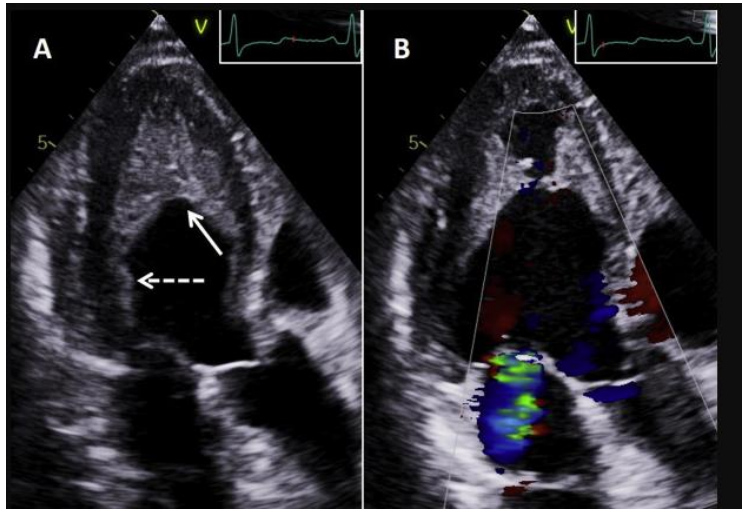
# Lymfocytär myokardit

- Inflammatorisk genes, kan bero på virus, bakterier.
- Cytotoxiska virus-cytolys och efterföljande immunaktivering/infiltration
- Indirekt aktivering genom aktivering av immunsystemet, eventuellt molecular mimicry?

- Åtgärd enl tidigare; varierande

# Eosinofil myokardit

- Ovanlig form.
- Bakomliggande orsak? Eosinofil granulomatös polyangit? Hypersensitivitet? Allergi? Parasitinfektion?Malignitet?
- Mortalitet på 22% på sjukhus.
- Varierande presentation, allt från fulminant myokardit loeffler kardiomyopati (progressiv fibros, hjärtförstoring, Infyllnad i apex). Oftast perifer eosinofili men 25% har normala eosinofil
- Biopsi. CMR med typiska eosinofila symptom
- Behandling; Kortison IV. IL-5 antagonist? Antikoagulation?



# Jättecellsmyokardit

- Sällsynt med dålig prognos. 85% har dött eller transplanterats efter 3år.
- Drabbar unga och medelålders utan könsskillnad.
- Icke infektiös myosin-inducerad autoimmun form. Hos 20% ses association med annan autoimmunitet.
- Symptom; Sedvanliga myokardit-symptom med snabb progress. Försämrad heomdynamik och arytmier. Bradyarytmier, retledningsrubbningar
- Biopsi krävs
- Behandling; Immunosuppression 2-3 olika varianter.
- Transplantation

# Sarkoidos

- Henning drar en fördjupning inom detta område

# Borreliakardit/ Chagas

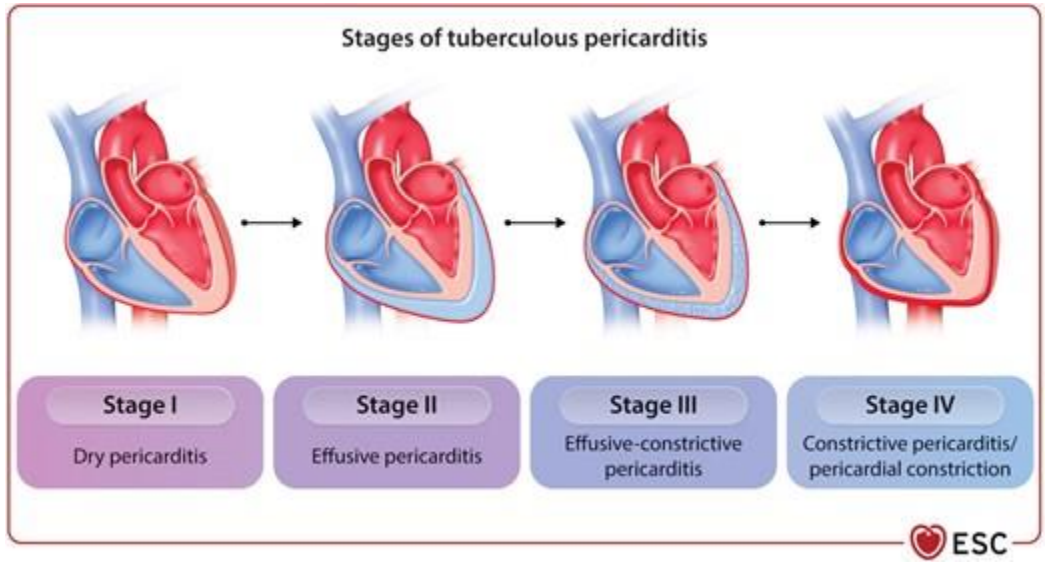
- Akut myokardit kan förekomma.
- Ofta i kombination med varierande grad av AV-blockering AVBIII hos ca 25% av fallen.
- Långa PQ-intervall.
- AV-block försvinner ofta efter 1-2v efter antibiotikabehandling.
- Chagas är pga parasit, migration medför att det numer uppträder fall i Europa.
- Ofta milda symptom. Taky, brady-arytmier. Dilaterade ventriklar. Behov av hög misstanke. PCR-testning och serologi
- Hög mortalitet. Behandling tidigt kan ge bättre prognos. Arytmier

# ICI-inducerad myokardit

- Cancerbehandling som är effektiv.
- Myokardit allvarligaste komplikationen, drabbar ca 1% av behandlingspopulationen.
- Majoriteten drabbas under de första 30 dagarna efter initerad behandling. Hög mortalitet.
- Oftare män med komorbiditet.
- Arytmier. Retledningsgrubbningar, Hjärtsvikt. Kombinerat med perifer myosit.
- Tidig diagnos är viktigt, Imaging, eventuell biopsi.
- Sätt ut behandling, kortison, dock 50% svarar ej på behandlingen. Oftast behov av flera olika behandlingsstrategier med andra immunhämmare.

# Tuberkulös Perikardit

- Mycket vanligt i världen, ovanligt i Sverige.
- Oftast akut perikarditinsjuknande med efterföljande exsudat.
- Genomgår 4 stadier
- Behov av analys av perikardvätska
- 50% går till konstriktion



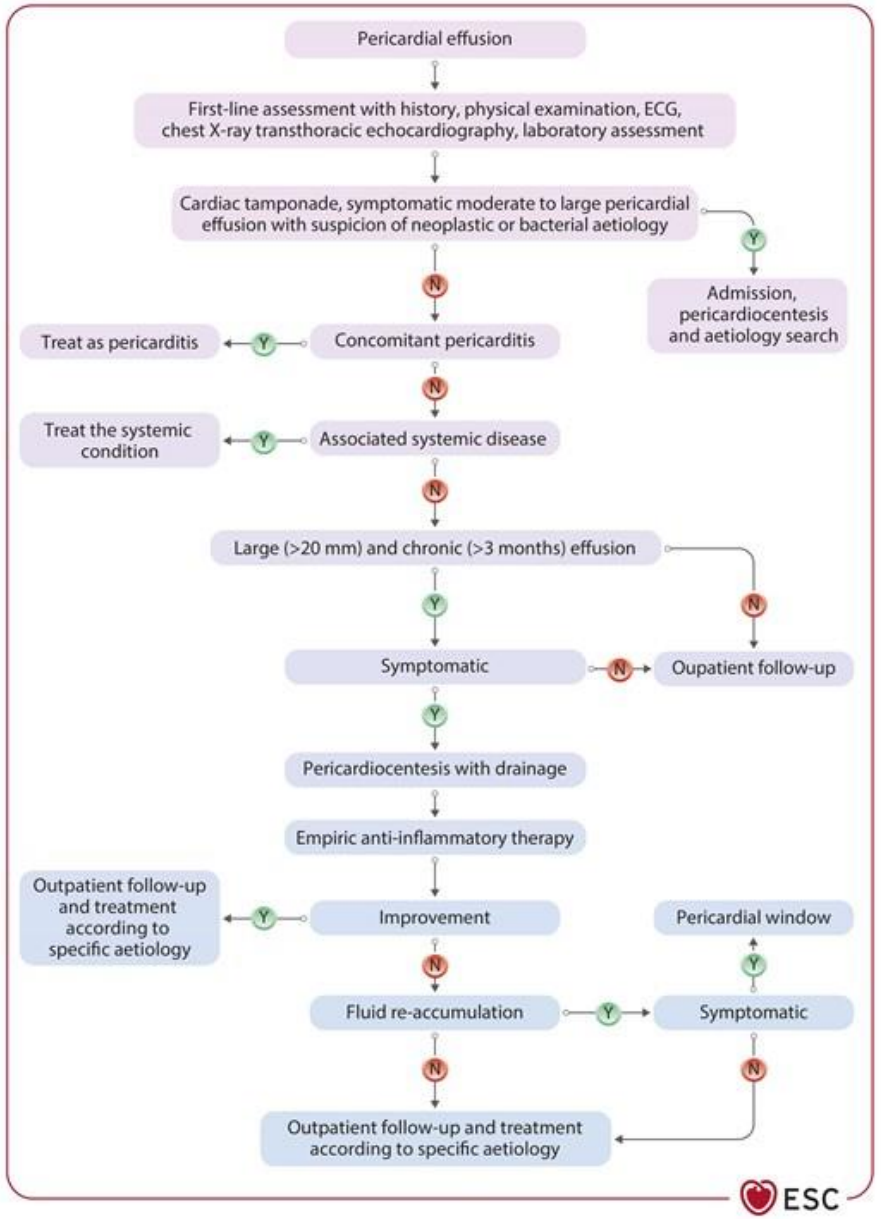
# Post-cardiac injury perikardit

- Vanligare och vanligare sekundärt till fler ingrepp.
- Innefattar Dresslers och postkardiotomi-syndrom
- Ca 10% får perikardit efter förmaksflimmer-ablation
- Symptom; feber perikardiell/pleuritisk smärta, perikardiella eller pleuritska gnindningsljud, Perikardvätska och pleuravätska. CRP-stegring
- Behandling: Kolkicin kan användas profylaktiskt. Post-AMI rimligt med ASA.
- IL-1 Antagonist?
- NSAID fungerar inte på post-kardiotomi

| | | |
|---|---|---|
| Anti-inflammatory therapy is recommended in patients with PCIS to hasten symptom remission and reduce recurrences.[292,600] | I | B |
| IL-1 antagonists are recommended in patients with refractory PCIS to prevent recurrences and progression to constriction.[604] | I | B |
| High-dose aspirin is recommended as the first-choice anti-inflammatory therapy for post-myocardial infarction pericarditis and in patients being already on antiplatelet therapy. | I | C |
| Colchicine, started 48 to 72 h before cardiac surgery, should be considered for 1 month in patients after cardiac surgery for the prevention of PCIS if there are no contraindications and if it is tolerated.[594,596] | IIa | A |
| Careful follow-up should be considered in patients with PCIS to exclude possible evolution towards constrictive pericarditis.[347] | IIa | C |

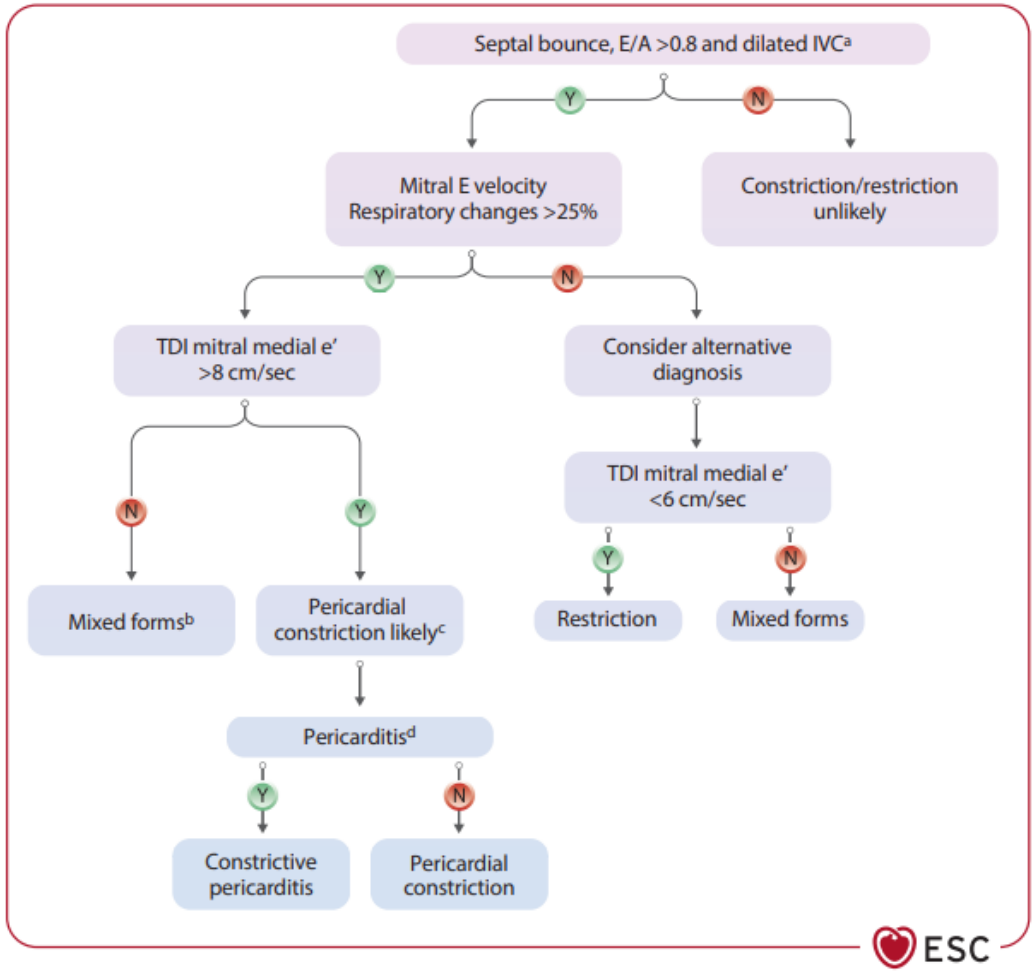
# Pex av annan genes

- Kan bero på flera olika orsaker. Inflammation? Lymfavflöde? Minskad reabsorption pga ökat CVP, pulmonell hypertension. Malignitet? Ideopatisk
- Ideopatisk 50%, cancer 10-30%, Iatrogena orsaker 15-20%, Bindvävssjukdomar 5-15%. Endemiska områden, TB 60%.
- Tamponad; kräver tappning



# Konstriktiv perikardit/konstriktion

- Relativt ovanligt
- Låg risk hos de med viral och ideopatisk perikardit men i västvärlden ändock vanligaste.
- Symptom i form av högersidiga sviktsymptom med bibevarad funktion på UKG. Ibland arytmi
- Varierande tid från perikardiella inflammation till konstriktion.
- Ibland utan bakomliggande perikardit
- Ofta lång tid till diagnos
- Mutimodal imaging, hjärtkateterisering



# Sammanfattning av guideline

- Det vanliga är det vanliga
- Vid hemodynamisk påverkan, arytmier etc. ta kontakt med högre vårdnivå för mer specifik diagnosticering. Biopsi? MCS?
- Följ upp patienterna (anpassa uppföljningen utifrån risken)
- På det som inte stämmer på det vanliga, gör det svåra roligt och driv diagnostiken. Ta hjälp av kollega, fördjupa anamnesen, hud, mun, händer, fötter etc.
- Tänk konstriktion vid besvär som inte passar in