DOENÇAS DO MIOCÁRDIO E PERICÁRDIO

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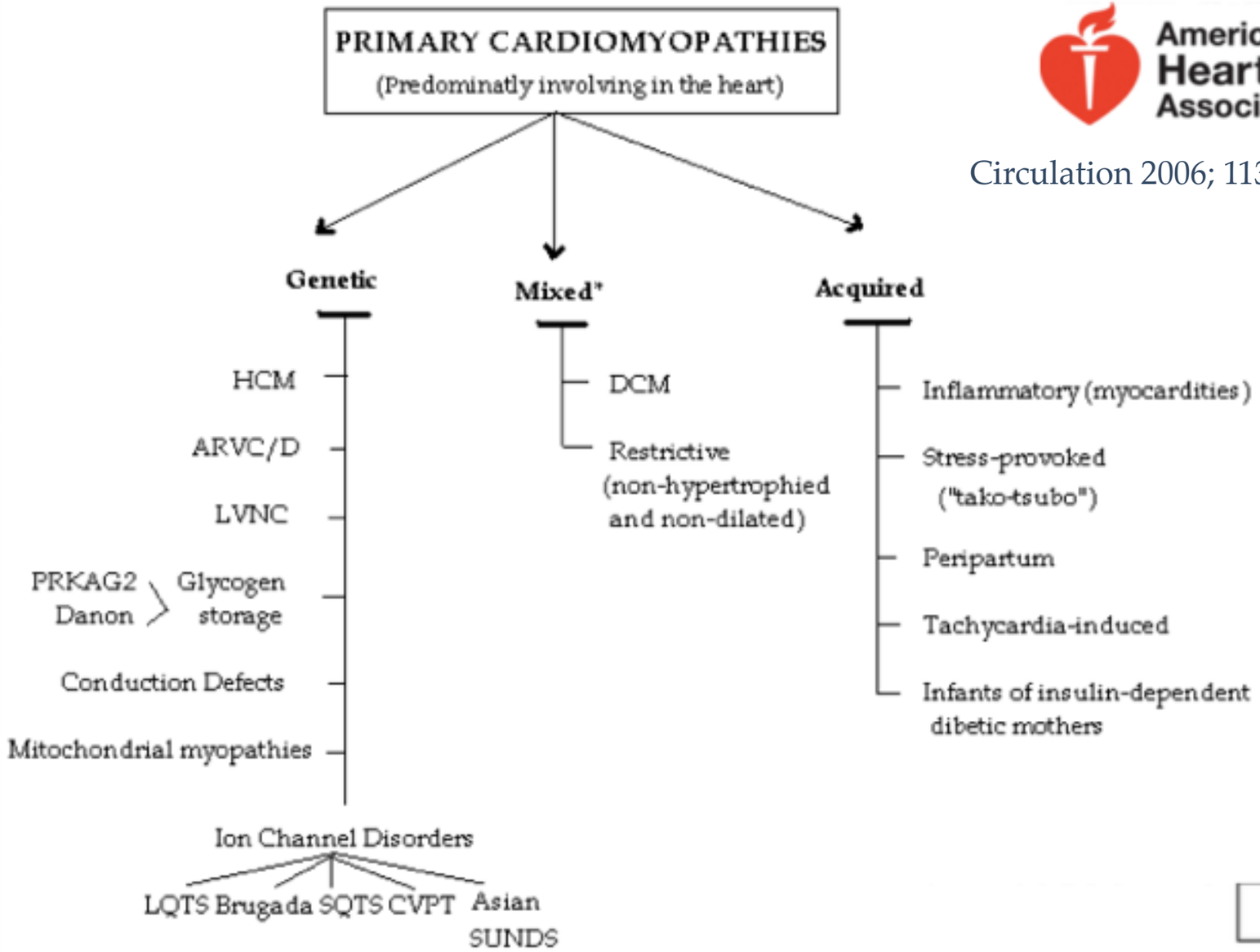
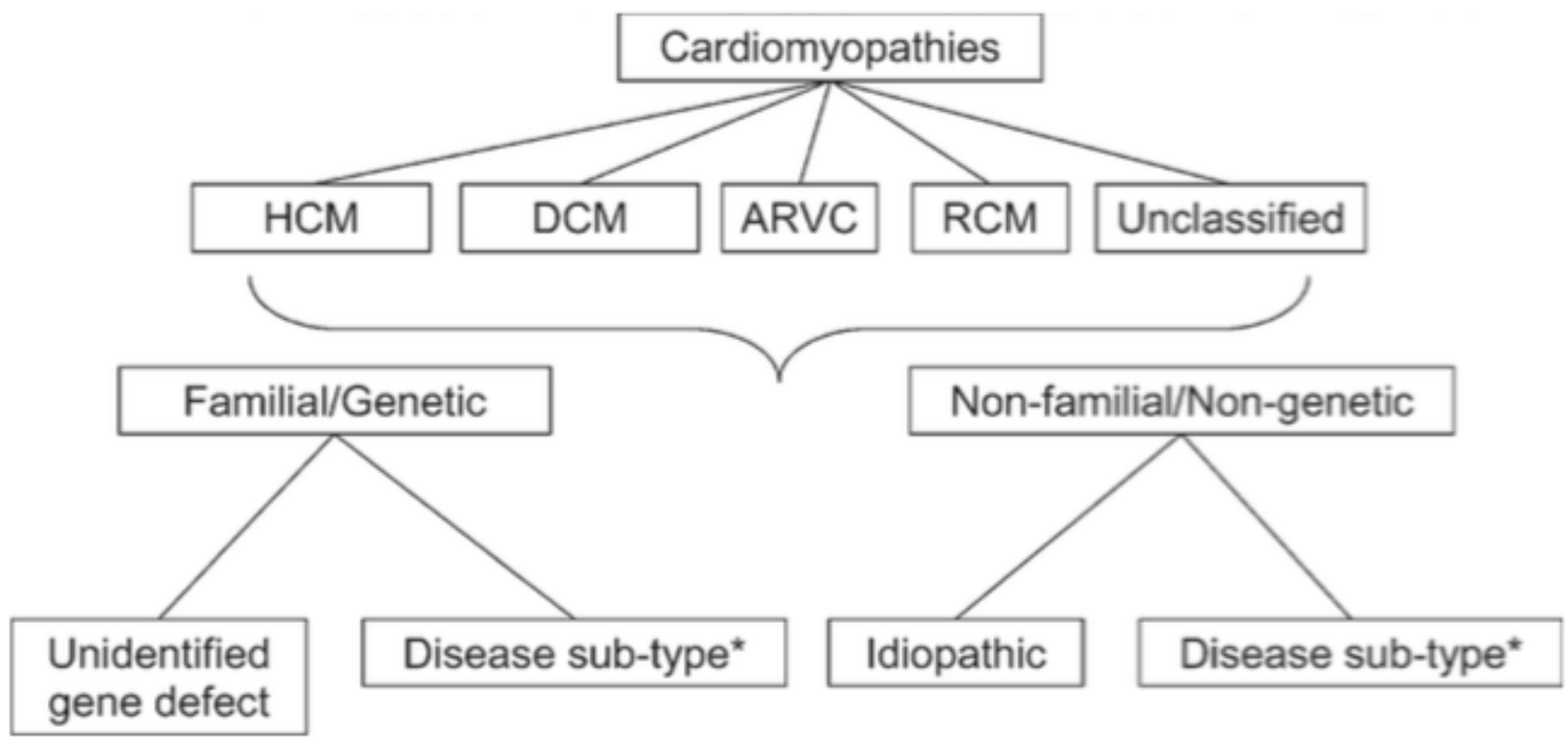
INTRODUÇÃO

A. DOENÇAS DO MIOCÁRDIO

Doenças do músculo cardíaco, caracterizadas por anomalias nas dimensões das cavidades cardíacas ou espessura da parede, com/sem disfunção, na ausência de causa óbvia



Circulation 2006; 113: 1807-1816



Eur Heart Journal 2008; 29: 270-276

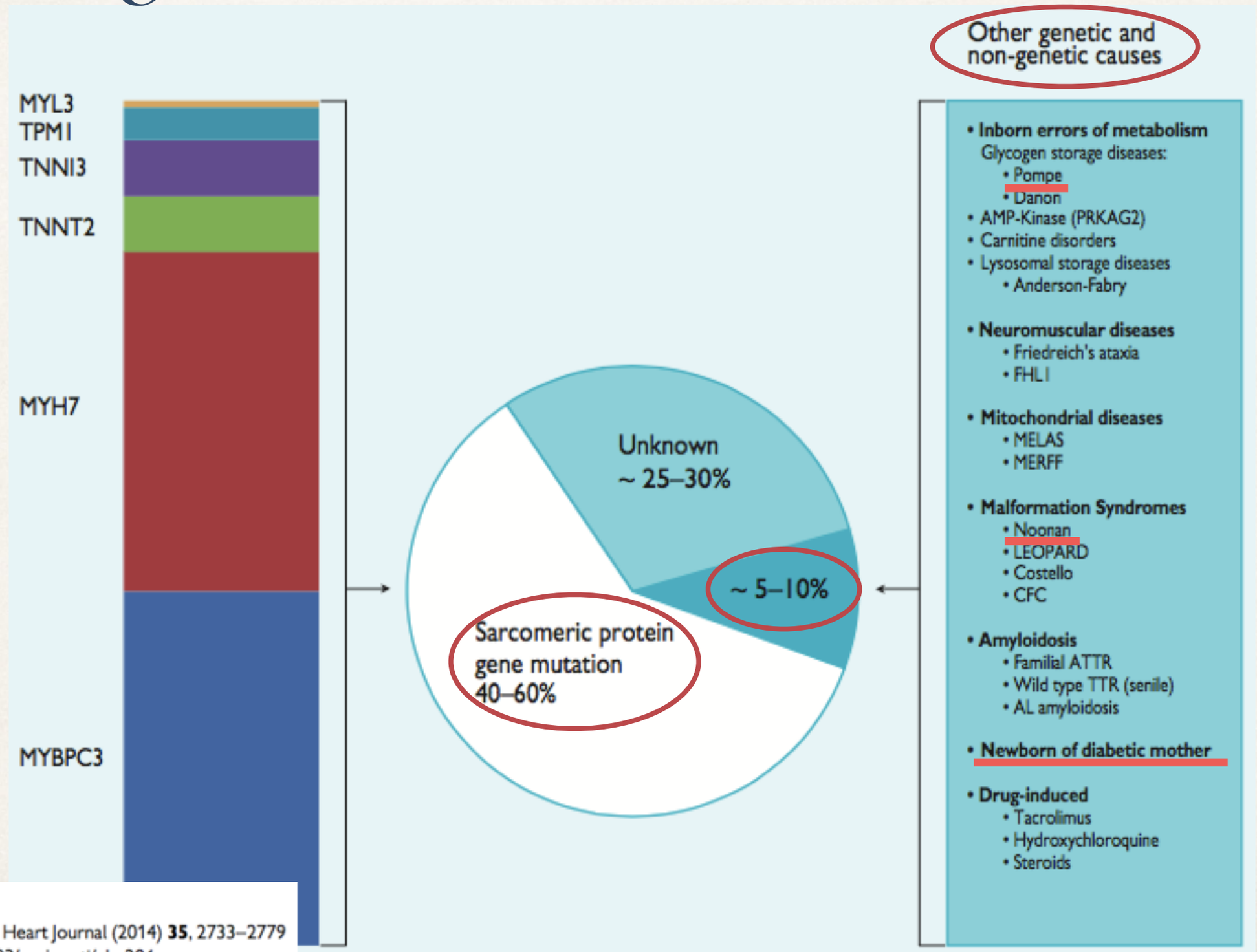
1. Miocardiopatia Hipertrófica

- cardiopatia frequente (0,2% população em geral)
- ++ adultos jovens
- aumento da espessura da parede ou da massa
    - hipertrofia assimétrica padrão mais comum
- causa mais frequente de MSC nos jovens

Disfunção diastólica

M. Hipertrófica

Etiologia



European Heart Journal (2014) **35**, 2733–2779
doi:10.1093/eurheartj/ehu284

Achados Clínicos

Clínica:

- cansaço
- palpitações
- dor anginosa
- tonturas, sincope
- HF + MCH (30-60%)

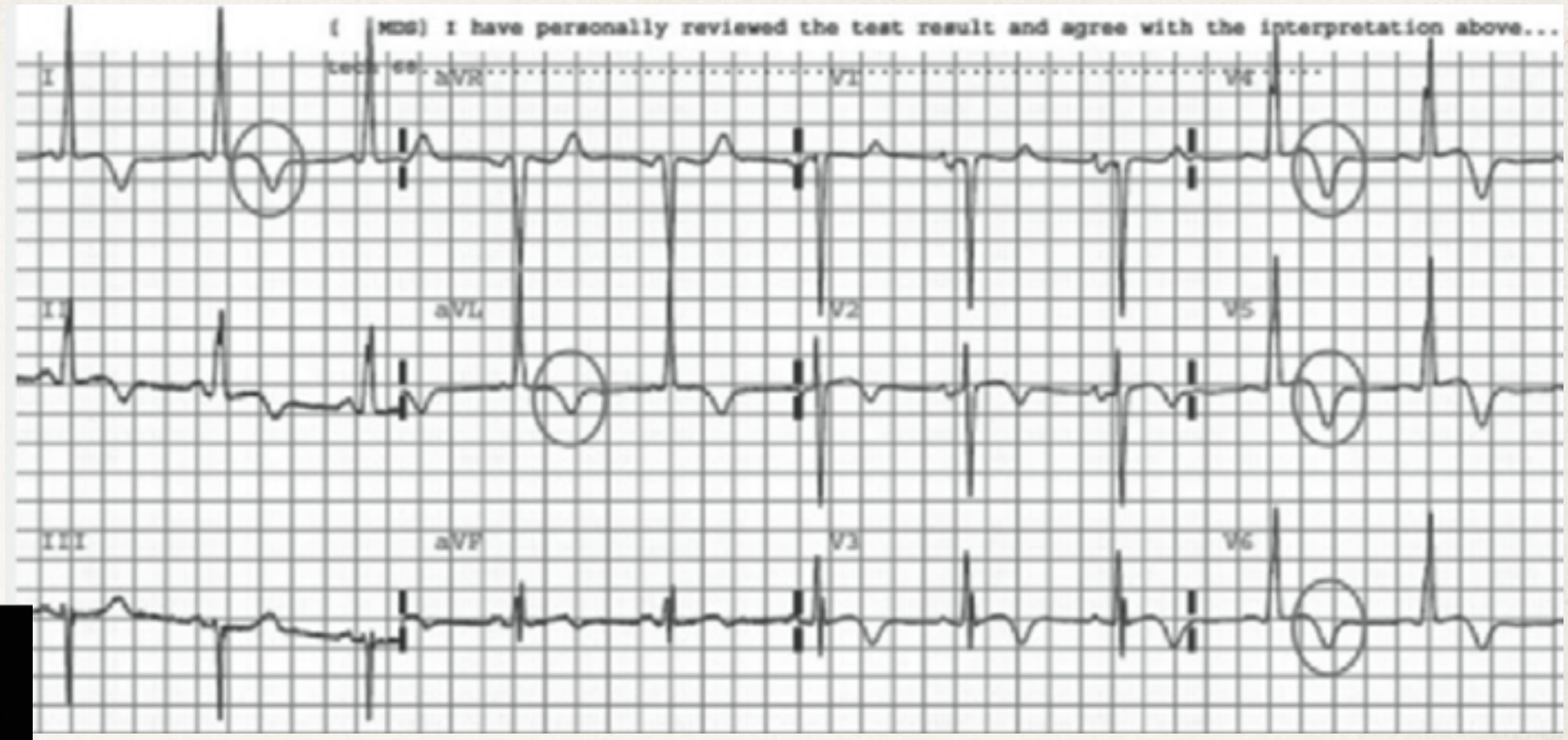
Exame Físico:

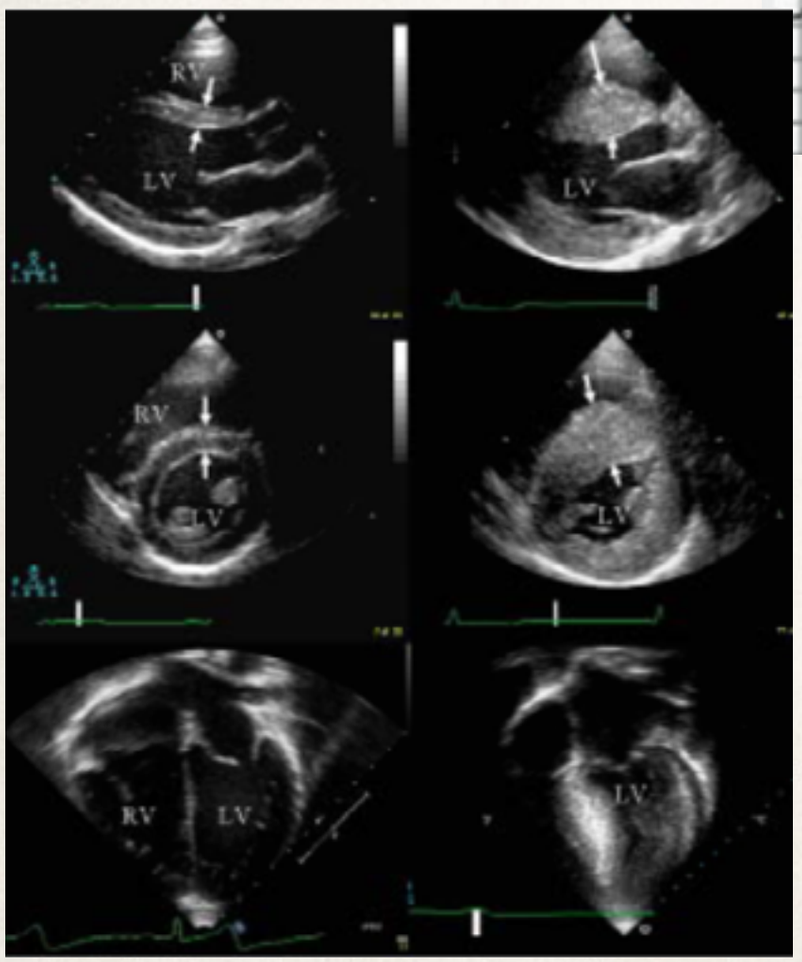
- sopro sistólico BEE inferior / apex

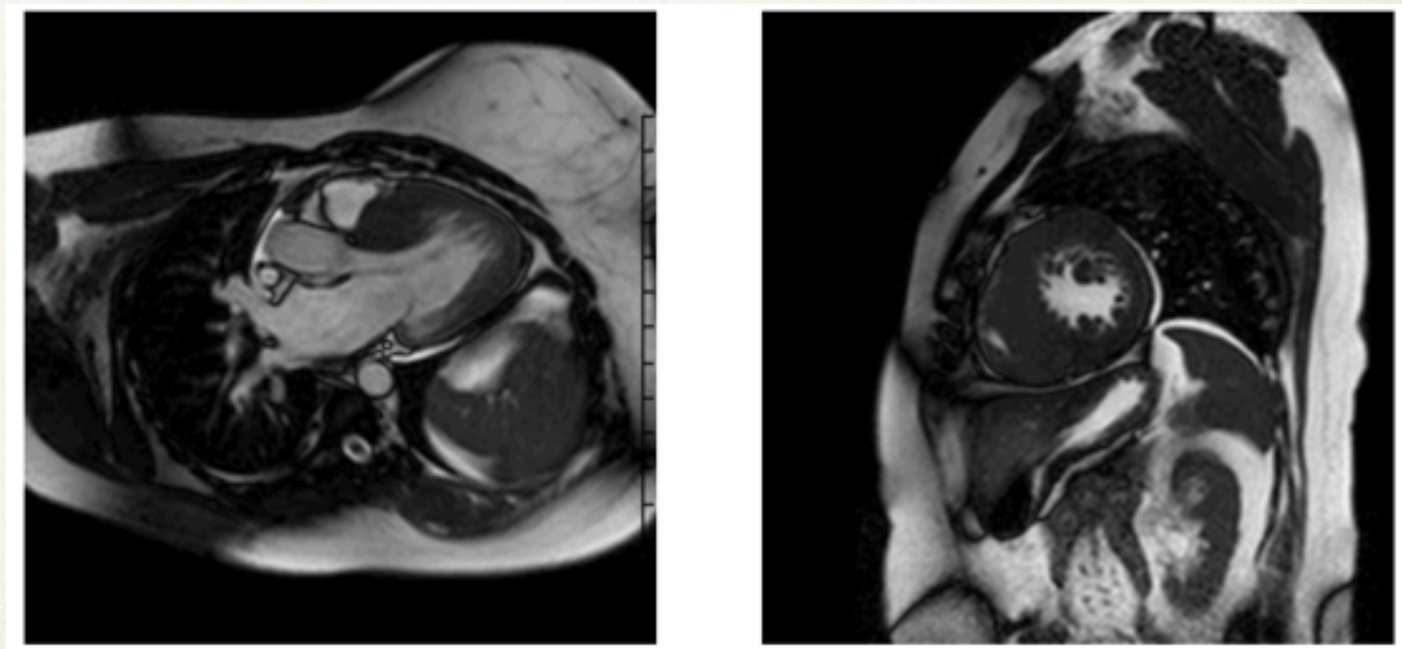
Agravamento clínico nas situações de < volume sistólico

M. Hipertrófica

ECD







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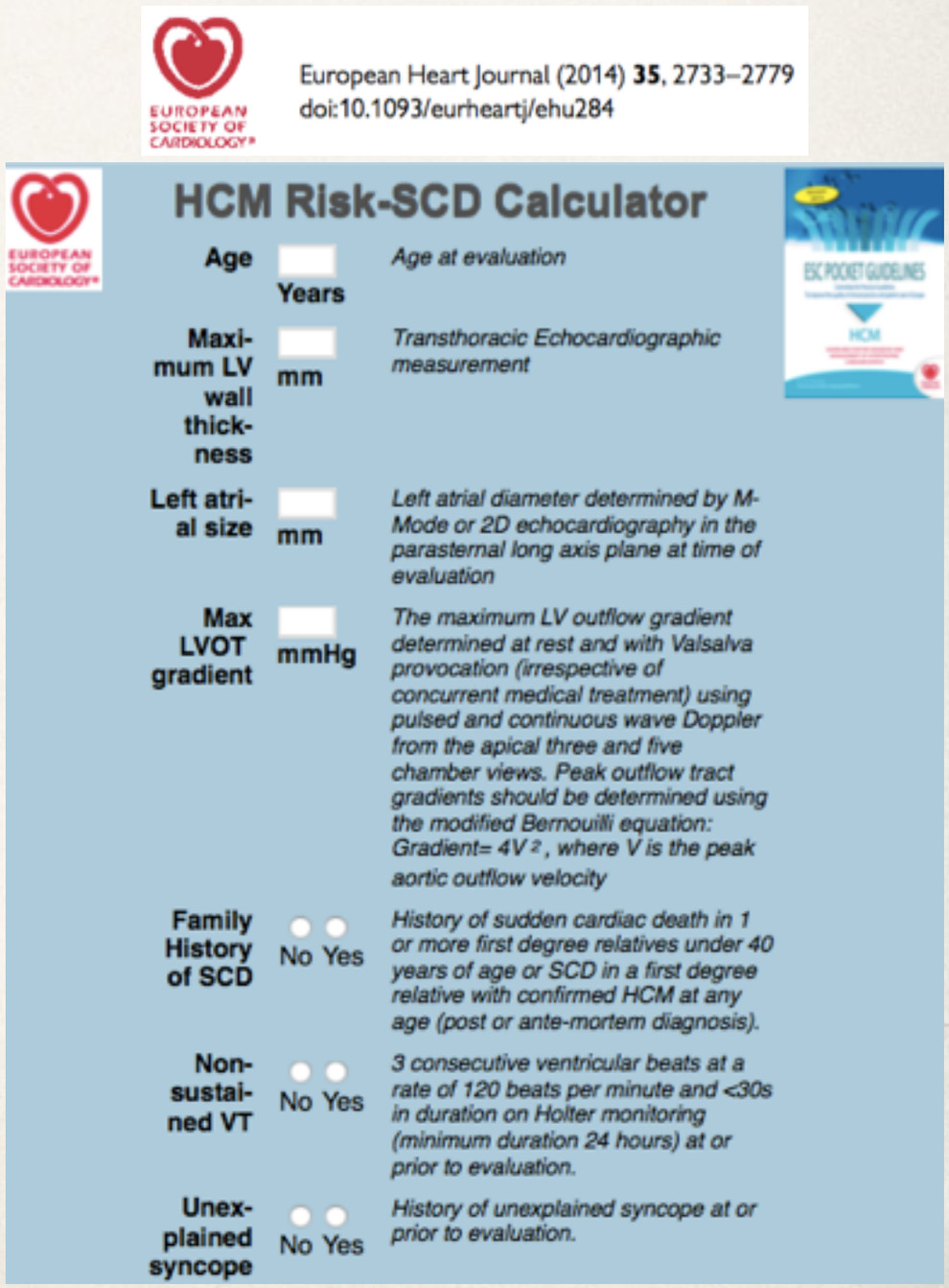
Terapêutica

- Evicção de desporto federado
- Beta bloqueantes
- Antagonistas canais de Ca++

  evitar vasodilatadores e diuréticos
- CDI
- Miectomia Morrows
- Ablação percutânea com álcool



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HCM Risk-SCD Calculator

Age	Years	*Age at evaluation*
Maximum LV wall thickness	mm	*Transthoracic Echocardiographic measurement*
Left atrial size	mm	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	mmHg	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	No Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	No Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	No Yes	*History of unexplained syncope at or prior to evaluation.*

2. Miocardiopatia Dilatada

- miocardiopatia mais frequente
- dilatação das 4 cavidades, com disfunção ventricular
- deterioração clínica progressiva
  - ICC
  - disritmias ventriculares
  - eventos embólicos

Disfunção sistólica

M. Hipertrófica

M. Dilatada

Etiologia

DCM

- Familial, unknown gene
- Sarcomeric protein mutations (see HCM)
- Z-band
  - Muscle LIM protein
  - TCAP
- Cytoskeletal genes
  - Dystrophin
  - Desmin
  - Metavinculin
  - Sarcoglycan complex
  - CRYAB
  - Epicardin
- Nuclear membrane
  - Lamin A/C
  - Emerin
- Mildly dilated CM
- Intercalated disc protein mutations (see ARVC)
- Mitochondrial cytopathy

- Myocarditis (infective/toxic/immune)
- Kawasaki disease
- Eosinophilic (Churg Strauss syndrome)
- Viral persistence
- Drugs
- Pregnancy
- Endocrine
- Nutritional — thiamine, carnitine, selenium, hypophosphataemia, hypocalcaemia
- Alcohol
- Tachycardiomyopathy

European Heart Journal (2008) **29**, 270–276
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Achados Clínicos

Clínica:

- intercorrência vírica recente
- anorexia
- cansaço
- ortopneia, dispneia paroxística

Exame Físico:

- taquicardia
- crepitações
- ingurgitamento jugular
- hepatomegália
- sopro holossistólico apex

M. Hipertrófica

M. Dilatada

ECD

Analiticamente:

- Hemograma + VS
- Bioquímica
- Gasometria venosa
- Enzimologia Cardíaca
- pro BNP
- ….

Terapêutica

- Diuréticos
- Vasodilatadores
- Inotrópicos
- Beta Bloqueantes
- Antiplaquetários, ACO
- Terapêutica doença base
- Transplante cardíaco

3. Miocardite

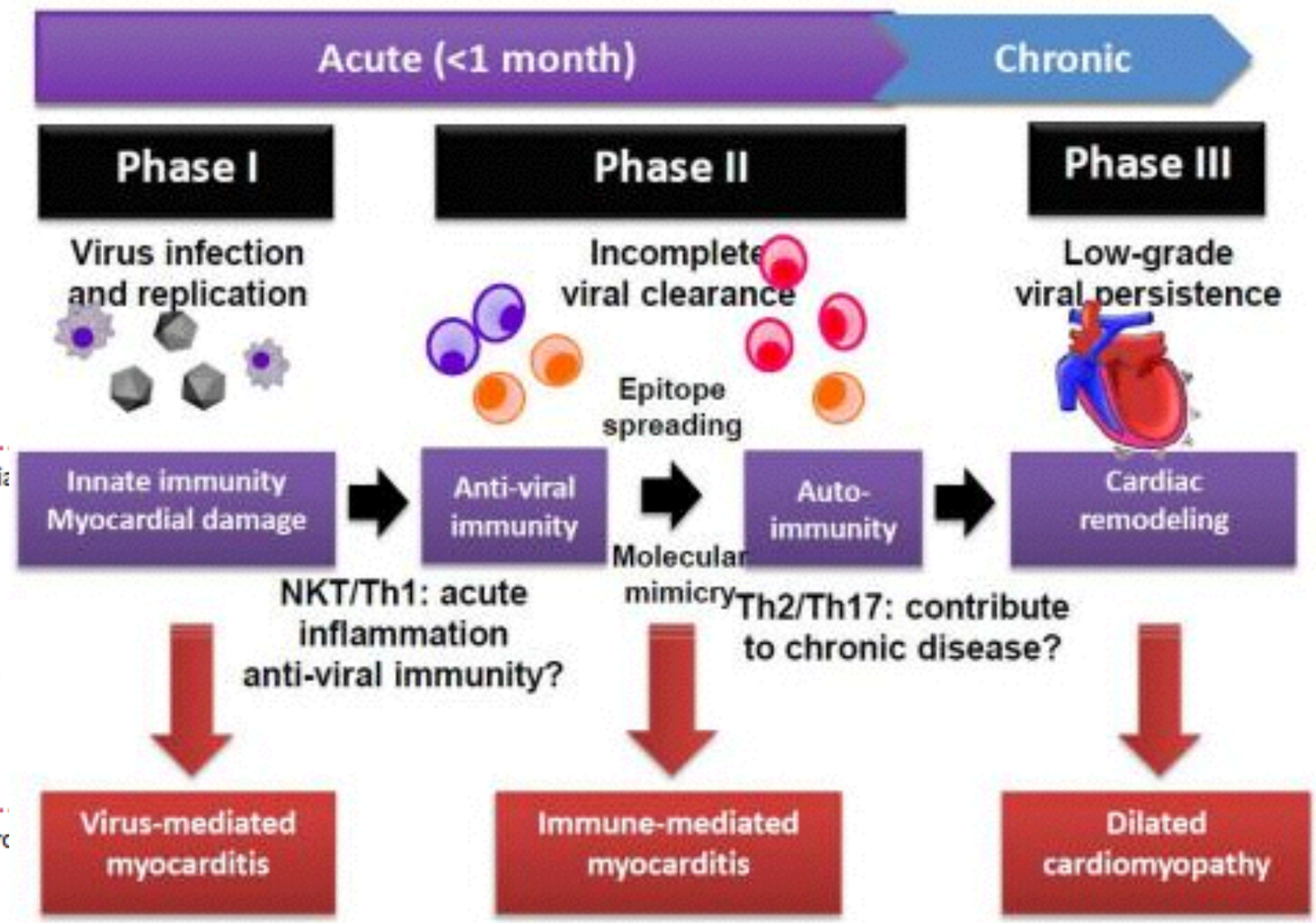
- doença com prevalência desconhecida (aguda << subaguda)
- infiltração inflamatória do miocárdio, com necrose dos miócitos
- recuperação em grau variável
  - ICC
  - substracto para arritmias ventriculares

Disfunção sistólica

Etiologia

Table I Causes of myocarditis/inflammatory cardiomyopathy

1. Infectious myocarditis
   - Bacterial — nophilus influenzae,
   - Spirochaetal
   - Fungal — rix
   - Protozoal
   - Parasitic
   - Rickettsial
   - Viral — irus, mumps virus, a fever virus, rabies; ster virus, herpes



2. Immune-media
   - Allergens — utazone,
   - Alloantigens
   - Autoantigens — Churg-Strauss; dependent diabetes
3. Toxic myocard
   - Drugs — e, interleukin-2,
   - Heavy metals — Copper, iron, lead (rare, more commonly cause intramyocyte accumulation)
   - Miscellaneous — Scorpion sting, snake, and spider bites, bee and wasp stings, carbon monoxide, inhalants, phosphorus, arsenic, sodium azide
   - Hormones — Phaeochromocytoma, vitamins: beri–beri
   - Physical agents — Radiation, electric shock

Future Virol (2012)

European Heart Journal (2013) **34**, 2636–2648
doi:10.1093/eurheartj/eht210

Achados Clínicos

Clínica:

- intercorrência virica recente
- anorexia
- dor pré cordial
- palpitações / sincope
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Exame Físico:

- taquicardia
- crepitações
- ingurgitamento jugular
- hepatomegália
- sopro holossistólico apex
- choque cardiogénico

ECD

Analiticamente:

- Hemograma
- Bioquímica
- Marcadores da inflamação
- Lactato
- Enzimologia Cardíaca
- pro BNP
- …

Terapêutica

- Repouso durante fase activa

- Diuréticos
- IECA / ARA
- Inotrópicos
- Beta Bloqueantes

- Imunomodeladores
- Imunossupressores

- VMI
- ECMO / Assistência Ventricular

5. Displasia Arritmogénica VD

- doença rara
- subtituição de miocárdio por tecido fibroadiposo (triângulo da displasia)
  - substracto arritmogénico
- causa importante de MSC em atletas

Etiologia

ARVC

- Familial, unknown gene
- Intercalated disc protein mutations
  - Plakoglobin
  - Desmoplakin
  - Plakophilin 2
  - Desmoglein 2
  - Desmocollin 2
- Cardiac ryanodine receptor (RyR2)
- Transforming growth factor-β3 (TGFβ3)

Inflammation?

Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

Proposed Modification of the Task Force Criteria

	Major Criteria	Minor Criteria
Family history	Familial disease confirmed at necroscopy or surgery	Familial history of premature sudden death (35 years of age) due to suspected ARVD. Family history (clinical diagnosis based on present criteria)
ECG abnormalities	Epsilon waves or localized prolongation (<110 ms) of QRS complex in right precordial leads (V1-V3)	Late potential on signal averaged ECG. Inverted T waves in right precordial leads (V2-V3) in subjects >12 years of age and in the absence of right bundle branch block
Arrhythmias	Sustained or nonsustained LBBB-like ventricular tachycardia documented on ECG or Holter monitoring or during exercise testing. Frequent ventricular extrasystoles (1000/24 hj on Holter	
Global or Regional dysfunction and structural abnormalities	Severe dilatation and reduction of RV ejection fraction with no or mild LV involvement. Localized RV aneurysms (akinetic or dyskinetic areas with diastolic bulgings). Severe segmental dilatation of RV.	Mild global RV dilatation or ejection fraction reduction with normal LV. Mild segmental dilatation of RV. Regional RV hypokinesia
Tissue characteristics of walls	Fibro-fatty replacement of myocardium on endomyocardial biopsy	

Circulation 2010; 121: 1533-1541

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European Heart Journal (2008) **29**, 270–276
doi:10.1093/eurheartj/ehm342

Achados Clínicos

Clínica:

- palpitações
- síncope
- Morte súbita
- HF + MSC

Exame Físico:

- frequentemente N

ECD

Terapêutica

- Evicção de desportos federados
- Anti-arrítmicos
- CDI

4. Miocardiopatia Restritiva

- causa rara (< 5%)
- dilatação severa biauricular, com dimensões ventriculares normais

Disfunção diastólica

Etiologia

RCM

- Familial, unknown gene
- Sarcomeric protein mutations
  - Troponin I (RCM +/− HCM)
  - Essential light chain of myosin
- Familial amyloidosis
  - Transthyretin (RCM + neuropathy)
  - Apolipoprotein (RCM + nephropathy)
- Desminopathy
- Pseuxanthoma elasticum
- Haemochromatosis
- Anderson–Fabry disease
- Glycogen storage disease

- Amyloid (AL/prealbumin)
- Scleroderma
- Endomyocardial fibrosis
  - Hypereosinophilic syndrome
  - Idiopathic
  - Chromosomal cause
  - Drugs (serotonin, methysergide, ergotamine, mercurial agents, busulfan)
- Carcinoid heart disease
- Metastatic cancers
- Radiation
- Drugs (anthracyclines)

European Heart Journal (2008) **29**, 270–276
doi:10.1093/eurheartj/ehm342

Achados Clínicos

Clínica:

- intolerância ao exercício
- dispneia esforço
- dor torácica

Exame Físico:

- distensão jugular
- hepatomegália
- sopro holossistólico apex

ECD

Terapêutica

- Diuréticos
- IECA / ARA
- Antiplaquetérios / ACO
- Corticódes e imunossupressores
- Transplante cardíaco

B. DOENÇAS DO PERICÁRDIO

- Pericardite
  - aguda
  - sub-aguda
  - crónica
  - recorrente
- Derrame pericárdico
- Tamponamento cardíaco
- Pericardite constritiva
- Massas pericárdicas

Pericardite Aguda

- doença do pericárdio mais frequente
- ++ homens 16-65A
- inflamação das superfícies pleurais
  - associação com derrame pericárdio
- prognóstico favorável
  - risco de evolução para pericardite constritiva

Etiologia

A. Infectious causes:
Viral (common): Enteroviruses (coxsackieviruses, echoviruses), herpesviruses (EBV, CMV, HHV-6), adenoviruses, parvovirus B19 (possible overlap with aetiologic viral agents of myocarditis).
Bacterial: *Mycobacterium tuberculosis* (common, other bacterial rare), *Coxiella burnetii*, *Borrelia burgdorferi*, rarely: *Pneumococcus* spp, *Meningococcus* spp, *Gonococcus* spp, *Streptococcus* spp, *Staphylococcus* spp, *Haemophilus* spp, *Chlamydia* spp, *Mycoplasma* spp. *Legionella* spp, *Leptospira* spp, *Listeria* spp, *Providencia stuartii*.
Fungal (very rare): *Histoplasma* spp (more likely in immunocompetent patients), *Aspergillus* spp, *Blastomyces* spp, *Candida* spp (more likely in immunocompromised host).
Parasitic (very rare): *Echinococcus* spp, *Toxoplasma* spp

B. Non-infectious causes:
Autoimmune (common): Systemic autoimmune and auto-inflammatory diseases (systemic lupus erythematosus, Sjögren syndrome, rheumatoid arthritis, scleroderma), systemic vasculitides (i.e. eosinophilic granulomatosis with polyangiitis or allergic granulomatosis, previously named Churg-Strauss syndrome, Horton disease, Takayasu disease, Behçet syndrome), sarcoidosis, familial Mediterranean fever, inflammatory bowel diseases, Still disease.
Neoplastic: Primary tumours (rare, above all pericardial mesothelioma). Secondary metastatic tumours (common, above all lung and breast cancer, lymphoma).
Metabolic: Uraemia, myxoedema, anorexia nervosa, other rare.
Traumatic and Iatrogenic: Early onset (rare): • Direct injury (penetrating thoracic injury, oesophageal perforation). • Indirect injury (non-penetrating thoracic injury, radiation injury). Delayed onset: Pericardial injury syndromes (common) such as postmyocardial infarction syndrome, postpericardiotomy syndrome, posttraumatic, including forms after iatrogenic trauma (e.g. coronary percutaneous intervention, pacemaker lead insertion and radiofrequency ablation).
Drug-related (rare): Lupus-like syndrome (procainamide, hydralazine, methyldopa, isoniazid, phenytoin); antineoplastic drugs (often associated with a cardiomyopathy, may cause a pericardiopathy): doxorubicin, daunorubicin, cytosine arabinoside, 5-fluorouracil, cyclophosphamide; penicillins as hypersensitivity pericarditis with eosinophilia; amiodarone, methysergide, mesalazine, clozapine, minoxidil, dantrolene, practolol, phenylbutazone, thiazides, streptomycin, thiouracils, streptokinase, p-aminosalicylic acid, sulfa-drugs, cyclosporine, bromocriptine, several vaccines, GM-CSF, anti-TNF agents.
Other (common): Amyloidosis, aortic dissection, pulmonary arterial hypertension and chronic heart failure.
Other (uncommon): congenital partial and complete absence of the pericardium.

European Heart Journal
doi:10.1093/eurheartj/ehv318

Achados Clínicos

Clínica:

- intercorrência infecciosa recente
- dor pré-cordial
- irradiação interescapular e ombro esquerdo
- agravamento com inspiração profunda e decúbito dorsal
- posição anti-álgica: flexão anterior do tronco

Exame Físico:

- atrito pericárdico

Factores de mau prognóstico:

- febre alta
- evolução sub aguda
- derrame pericárdico volumoso
- ausência de resposta aos AINEs
- miopericardite

ECD

Analiticamente:

- Hemograma
- Bioquímica
- Marcadores inflamatórios
- Enzimologia Cardíaca
- Investigação adicional se FR

Terapêutica

- repouso durante fase activa
- anti-inflamatórios
  - AAS 80-100mg/kg/dia, 6/6h (máx 4gr/dia), 1-2S
  - Ibuprofeno 20mg/kg/dose, 8/8h (máx 600mg 8/8h), 1-2S
  - Colchicina 0,5mg, 1id se <70Kg e 2id se >70Kg, 3M
  - Prednisolona 0,2-0,5mg/kg/dia
- pericardiocentese

CONCLUSÕES

- As doenças do miocárdio são um grupo heterogéneo de doenças
  - MCH - risco de MSC
  - MCD - evolução progressiva para ICC
  - Miocardite - evolução possível para MCD / risco MSC
- As doenças do pericárdio são relativamente frequentes na práctica clínica
  - Pericardite vírica com prognóstico frequentemente favorável

Fundamental investigação etiológica nas formas graves

Rastreio familiar