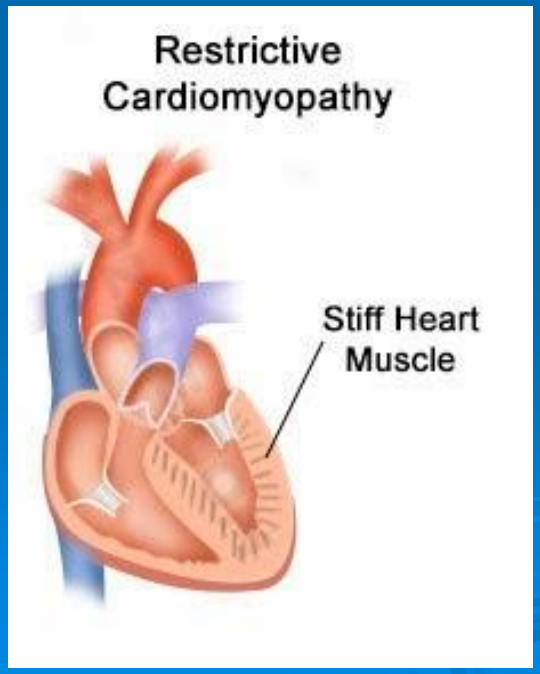
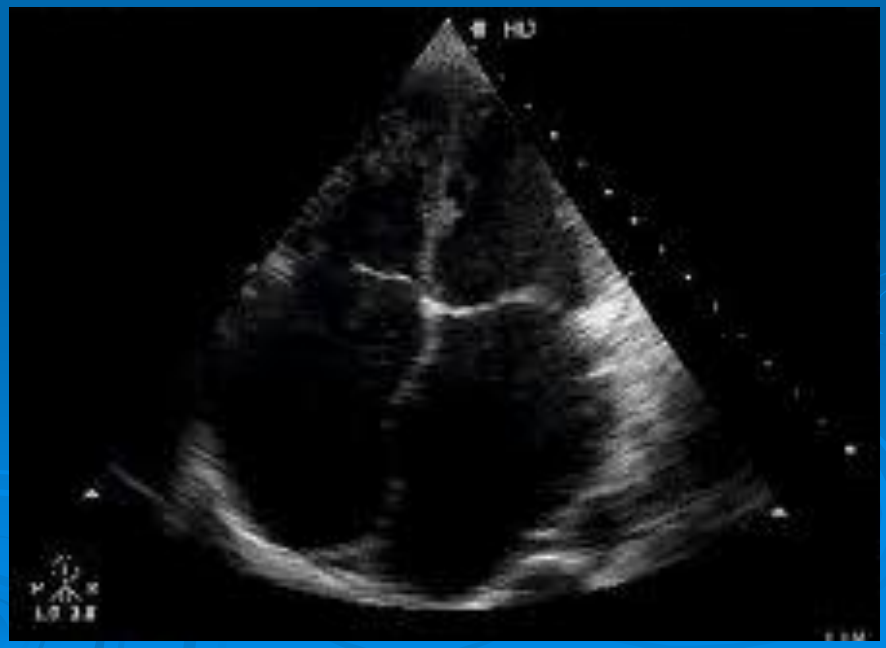
CARDIOMIOPATIA RESTRICTIVA

Cardiomiopatia restrictiva

- Cardiomiopatii foarte rare in arealul nostru geografic
- Forme idiopatice
- In Africa – mai frecventa
  - sub forma endocarditei fibroblastice cu eozinofile





CMR

- Fiziopatologie:
  - Inlocuirea tesutului muscular cu tesut fibros
  - Fibroza endocardica, reduce complianta ventriculara
  - Staza in amonte de ventriculi
  - Se produce dilatare atriala
  - Sunt absente hipertrofia si dilatatia ventriculara
- Sunt interesati ambii ventriculi, predominant VS.
- Scaderea compliantei ventriculare produce un tablou clinic asemanator cu pericardita constrictiva.

Anatomopatologie

- Macroscopic:
  - cordul apare marit de volum, prin dilatarea atriilor
  - ventriculii sunt de volum mic, secundar fibrozei, uni sau bi ventriculare.

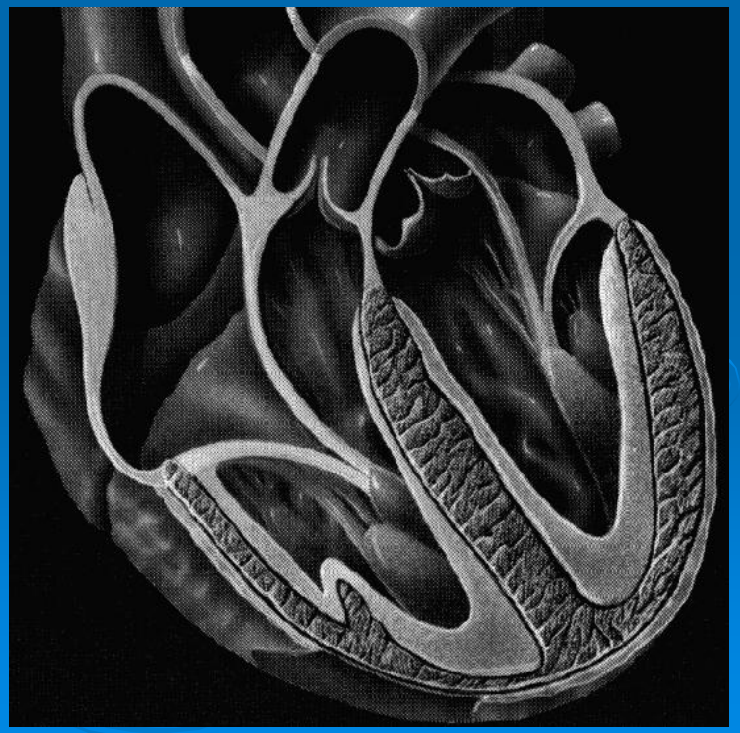
- Endocardul apare alb-sidefiu
- Motilitatea valvelor mitrala si tricuspida este limitata
- Se realizeaza o insuficienta valvulara datorita ingrosarii si retractiei pilierilor si cordajelor.

- Microscopic:
  - se evidentiaza un proces de fibroza, fara leziuni de fibroelastoza endocardica notabila

- Acest aspect a facut sa se considere ca fibroza ar constitui o sechela a unei miocardite necrotice, procesul evoluand in trei etape:
  - miocardita necrotica,
  - stadiul trombotic cu ingrosare endocardica
  - si fibroza cicatriceala.

Etiologie

- Idiopatica - cel mai frecvent
- Parazitara – se insoteste de eozinofilie
- Post iradiere cardiaca - radioterapie



Clinic

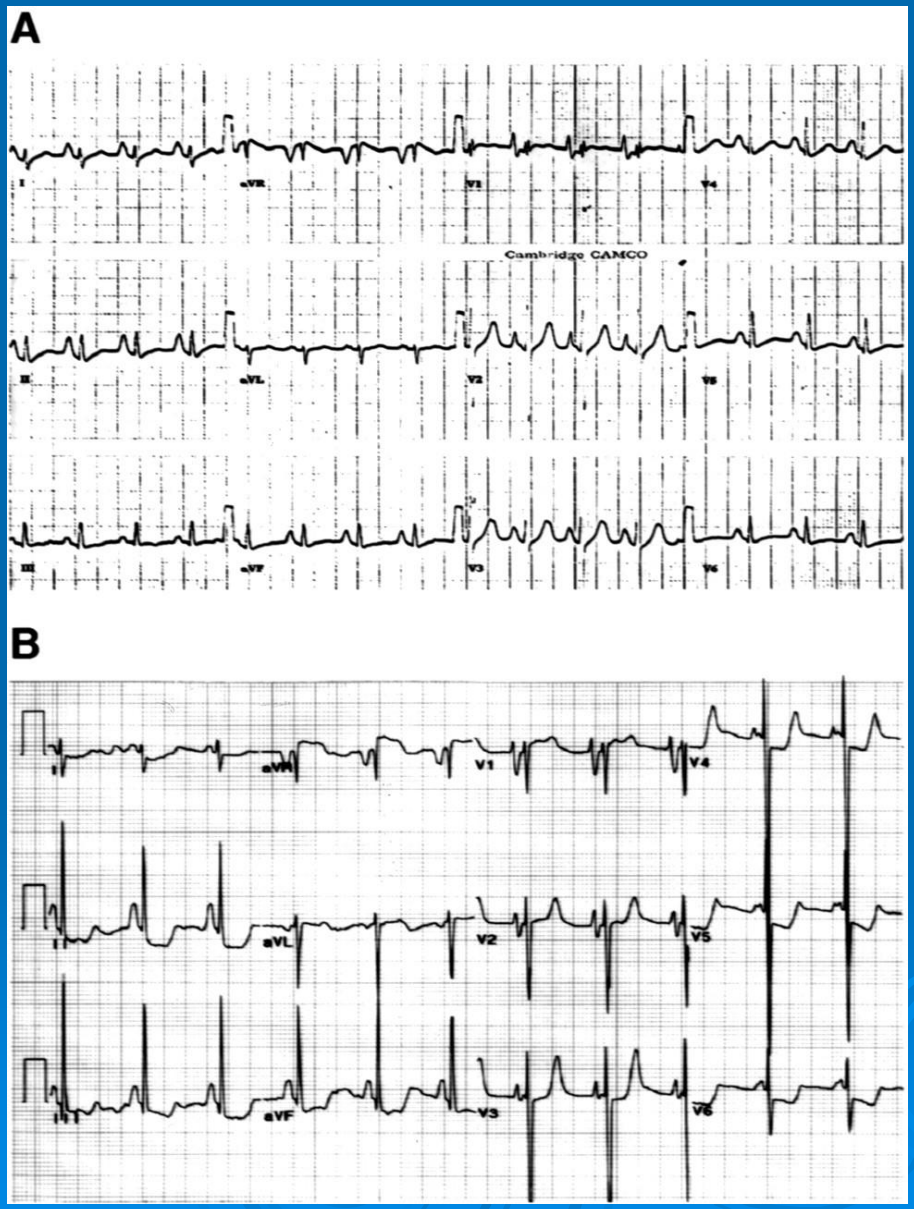
- Diagnostic dificil

- Alterarea starii generale
- Dispnee de efort
- Hepatalgii, hepatomegalie de staza
  - semn cardinal care poate mima o boala hepatica
- Turgescenta jugularelor
- Revarsat pleural, pericardic, ascita
- Edeme ale membrelor inferoare

ECG

- Unda P inalte ascutite
  - tip hipertrofie biatriala
- +/- hipertrofie ventriculara dreapta/stanga
- Uneori tulburari de repolarizare
  - unde T negative in V5 – V6

ECG

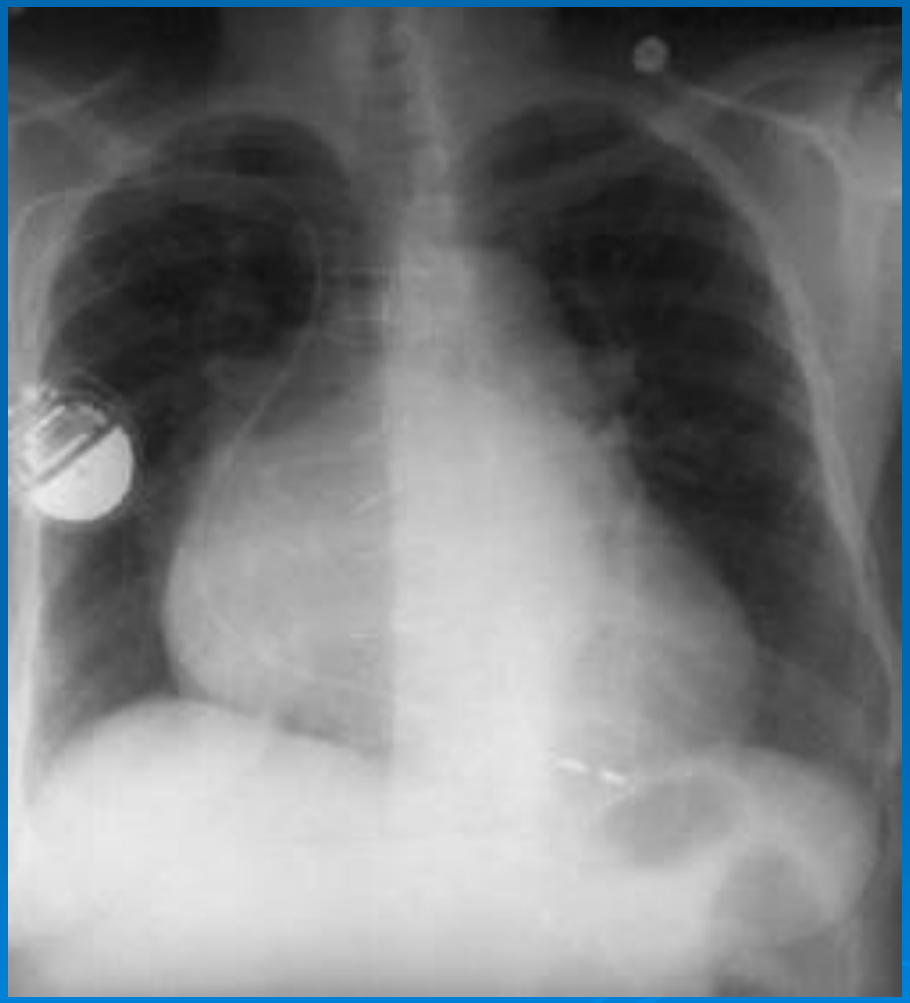


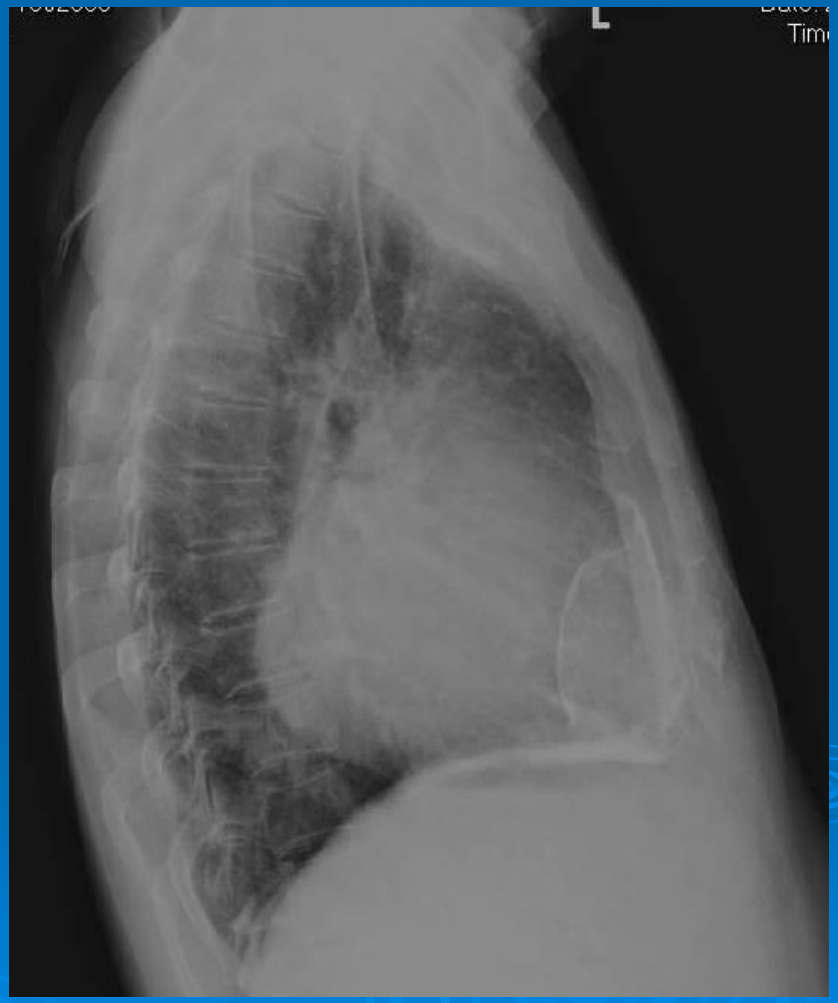
Rx toracic

- Cord cu silueta normala sau cardiomegalie prin dilatarea atriala

- Staza venoasa pulmonara

Rx cardiopulmonar

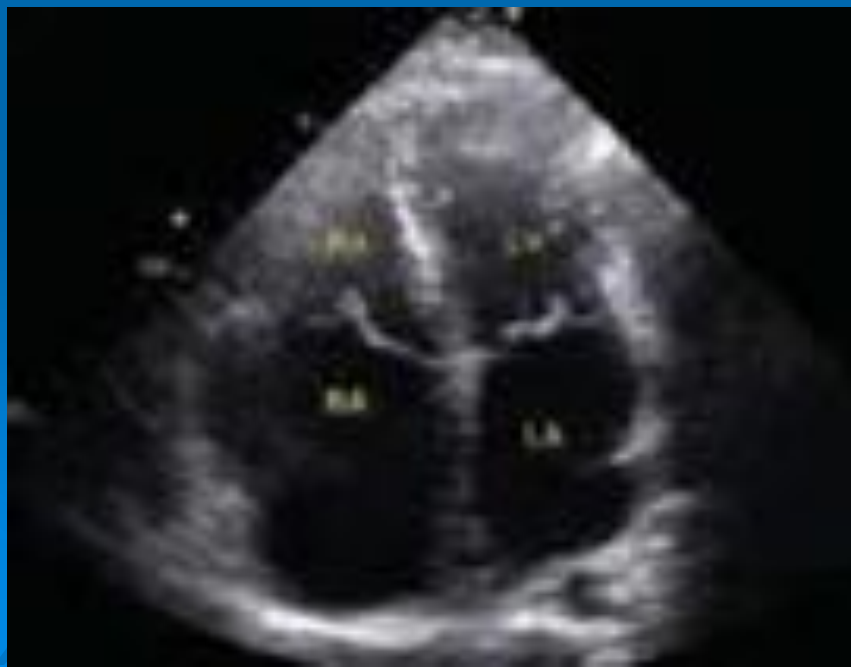




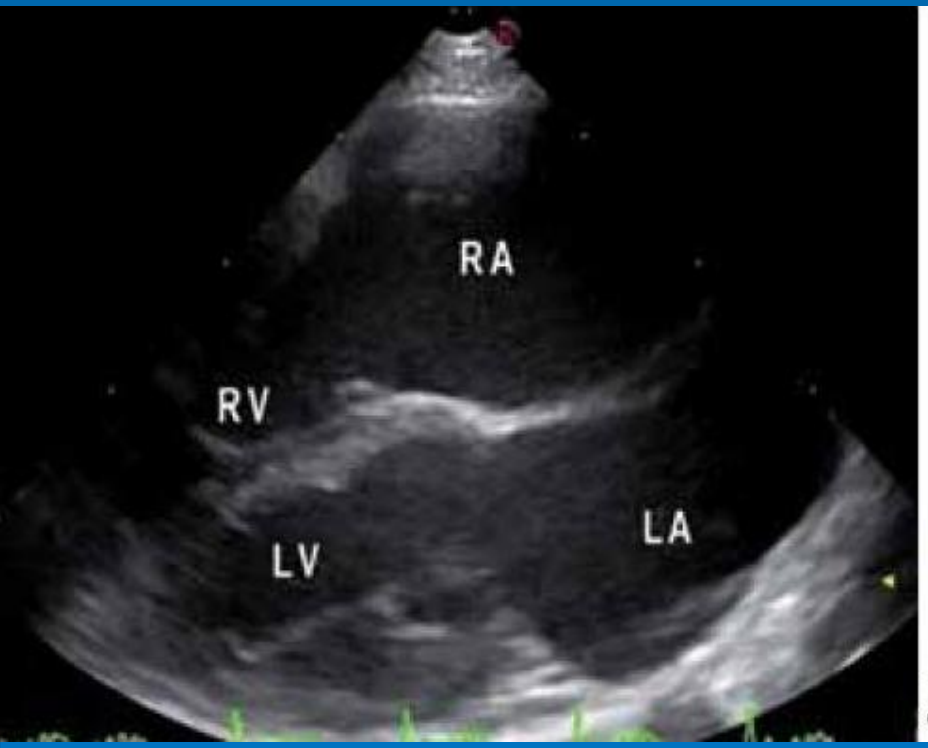
Dilatare de AD si AS, dublu contur, proiectie pana la coloana

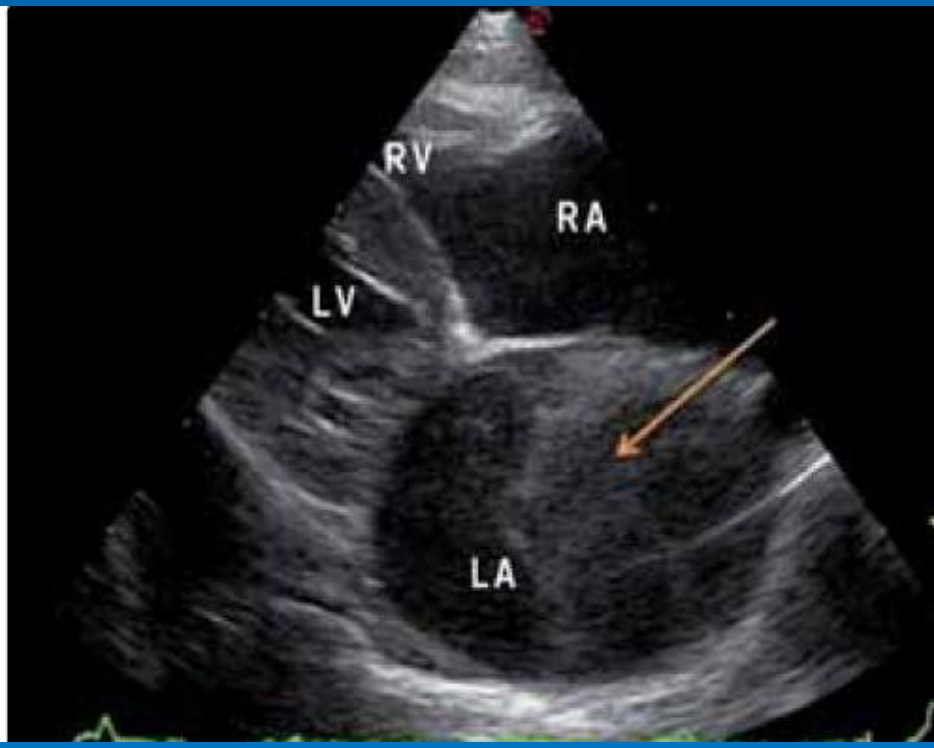
Echocardiografia

- Semne de restrictie a functiei ventriculare
- Disfunctie diastolica ventriculara
- Foarte dificil de diferentiat de pericardita constrictiva
- Flux diastolic mitral : unda E >> A
- Ingrosarea endocardului
- Dilatarea marcata a atriilor



Echocardiografia

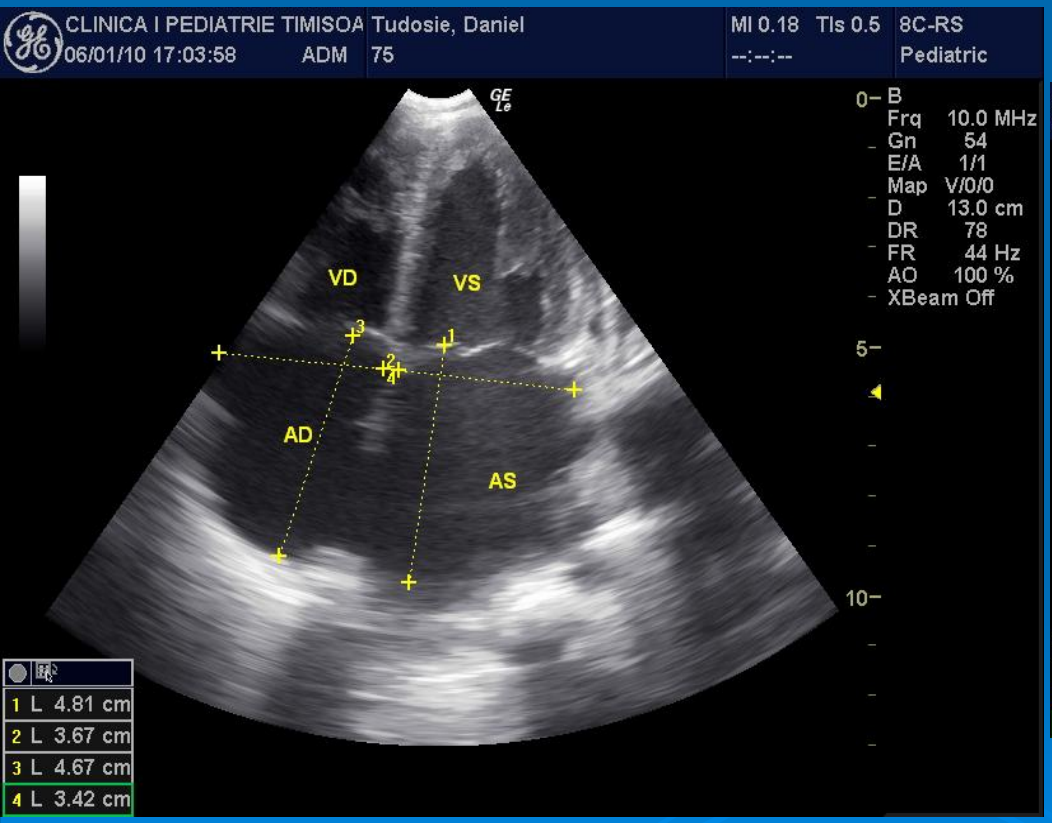




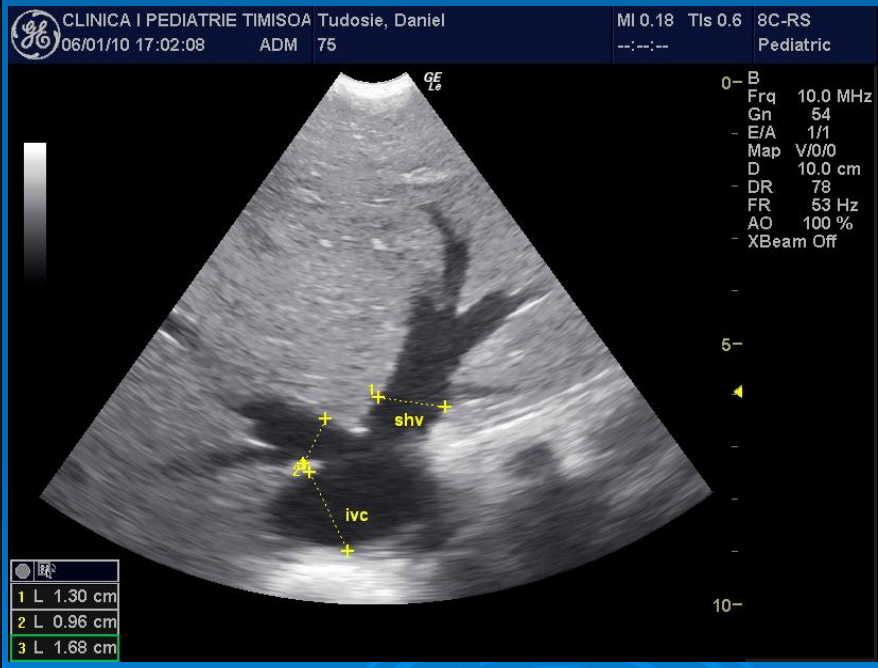
Dilatare de atrii importanta

Contrast spontan in AS

Echocardiografie

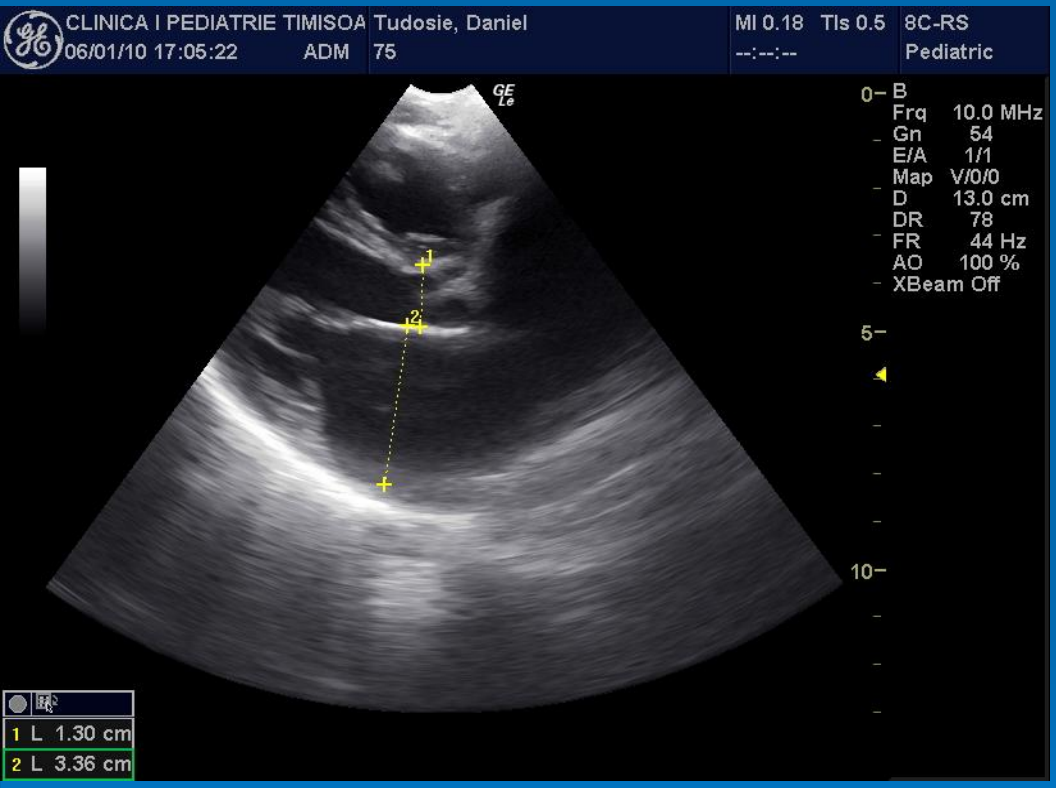


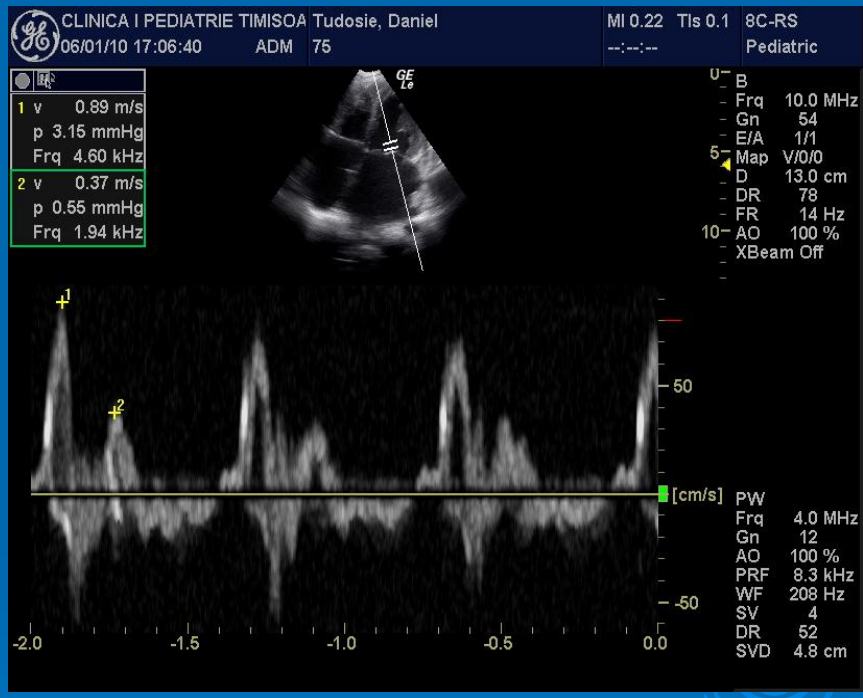
Dilatare de atrii



Dilatare VCI si vase hepatice

Echocardiografie

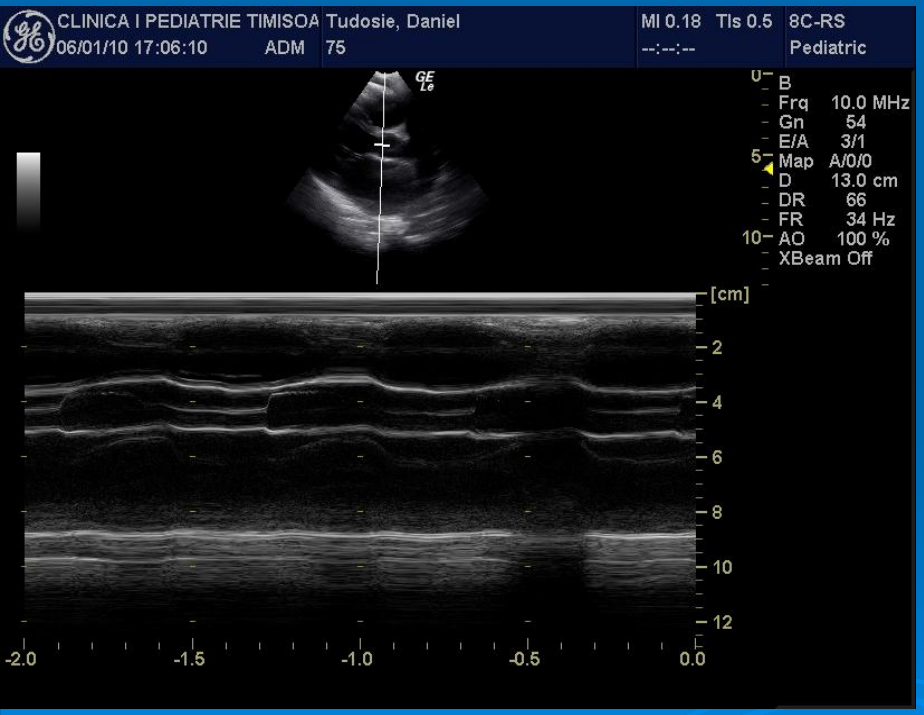


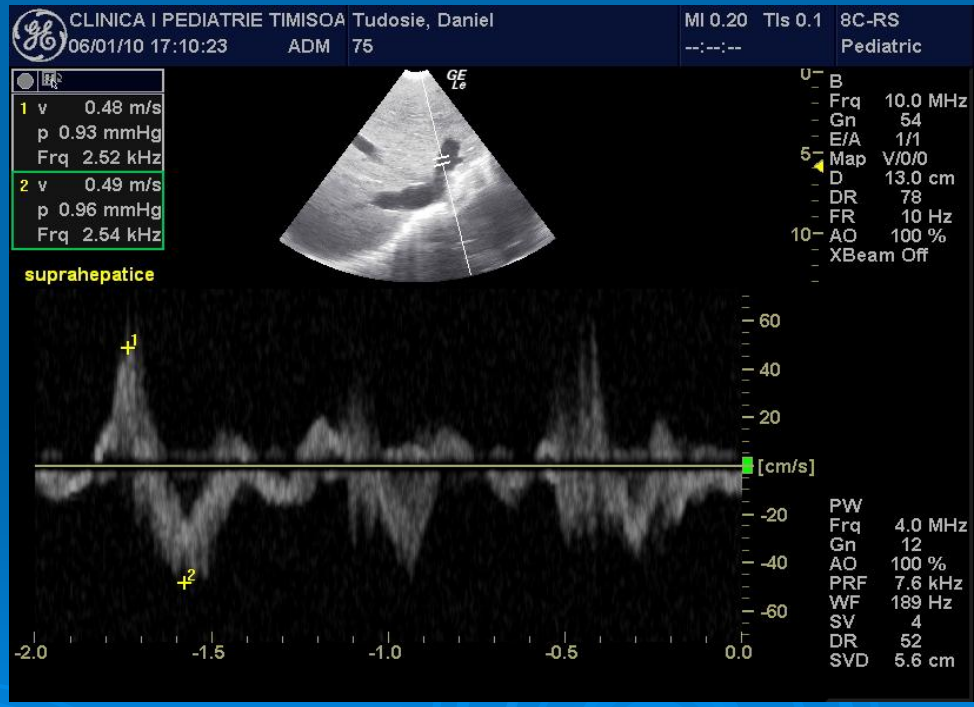


Dilatare de AS

E mult mai mare decat A

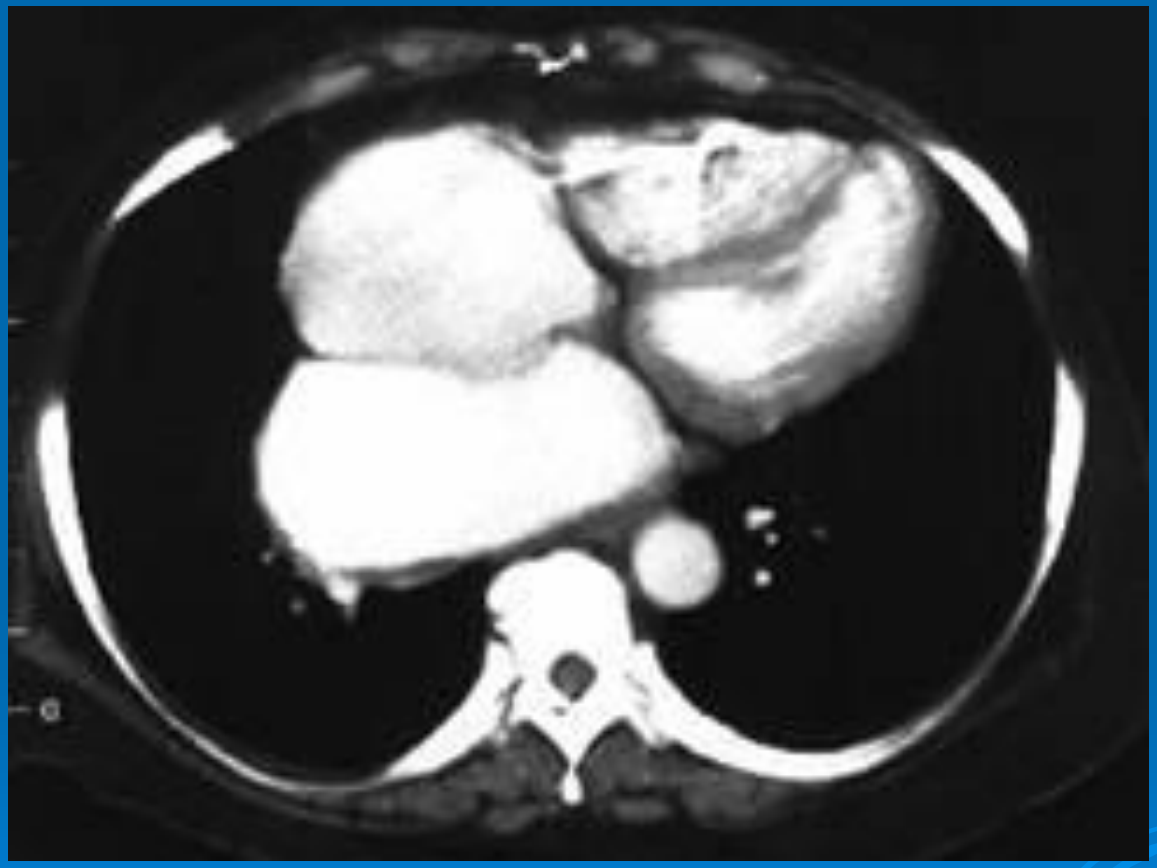
Echocardiografie





Dilatare de AS si vene hepatice

RMN Cardiac



- Dilatare biatriala
- RMN sau CT cardiac –diferentiaza diagnosticul de o pericardita constrictiva, prin evidentierea unui pericard de grosime normala < 1mm

Evolutie

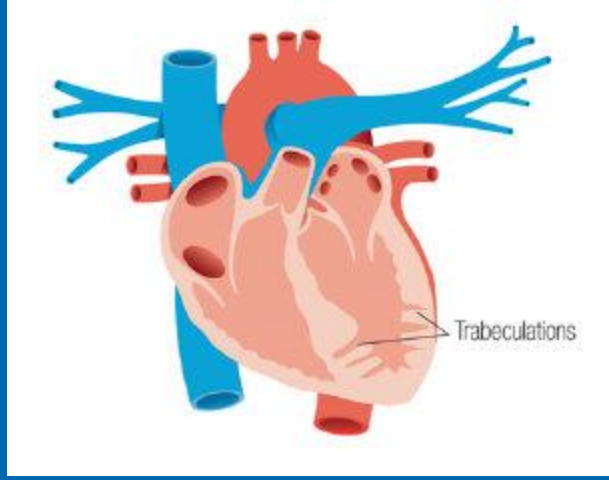
- Spre agravare si deces
- Tratament:
  - diuretic si al insuficientei cardiace – temporar eficient
- Transplantul cardiac - singurul eficient!

Recuperarea

- Necesita timp.
- Pana la transplantul de cord efortul fizic este limitat, spunem ca poate fi in limita tolerantei fizice.
- Copilul trebuie ajutat sa miste toate grupele musculare deoarece poate avea tendinta sa stea mai mult la pat in fazele pretransplant.
- Trebuie stimulat la jocuri usoare.
- Dupa transplant, recuperarea este lenta si progresiva.

CARDIOMIOPATIA NONCOMPACTANTA de VS

Definitie



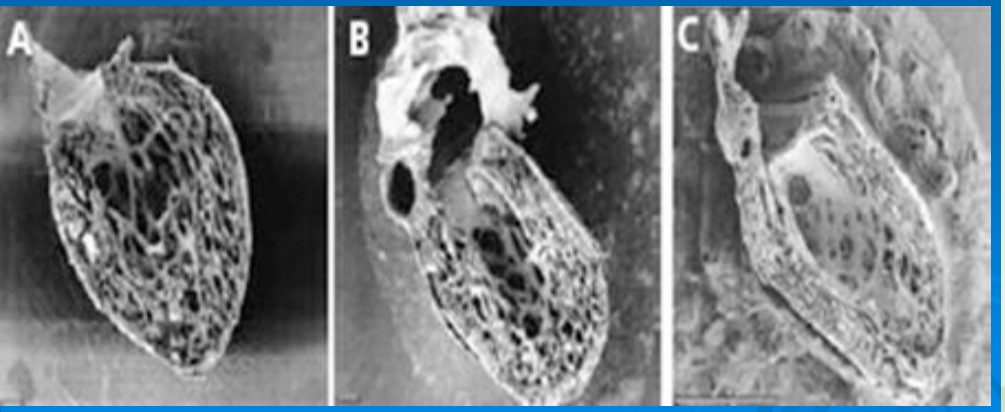
- Denumita si:
  - Cardiomiopatie spongiforma
- Cardiomiopatie congenitala rara
  - AHA o considera cardiomiopatie genetica

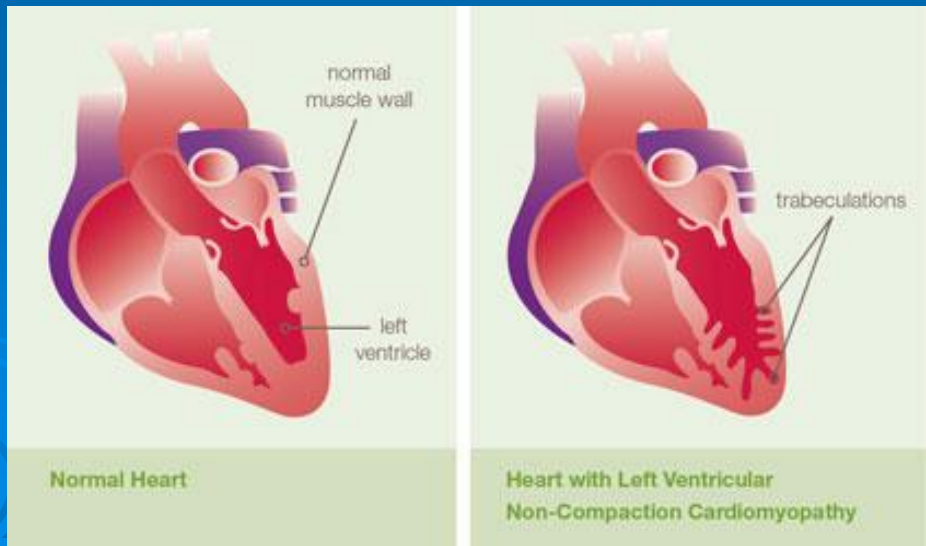
- Defect de dezvoltare miocardica din timpul embriogenezei:
  - Structura spongiforma embriogenetica nu sufera procesul de compactare si ramane spongioasa
  - Afecteaza in general VS – noncompactare de VS

Anatomopatologic

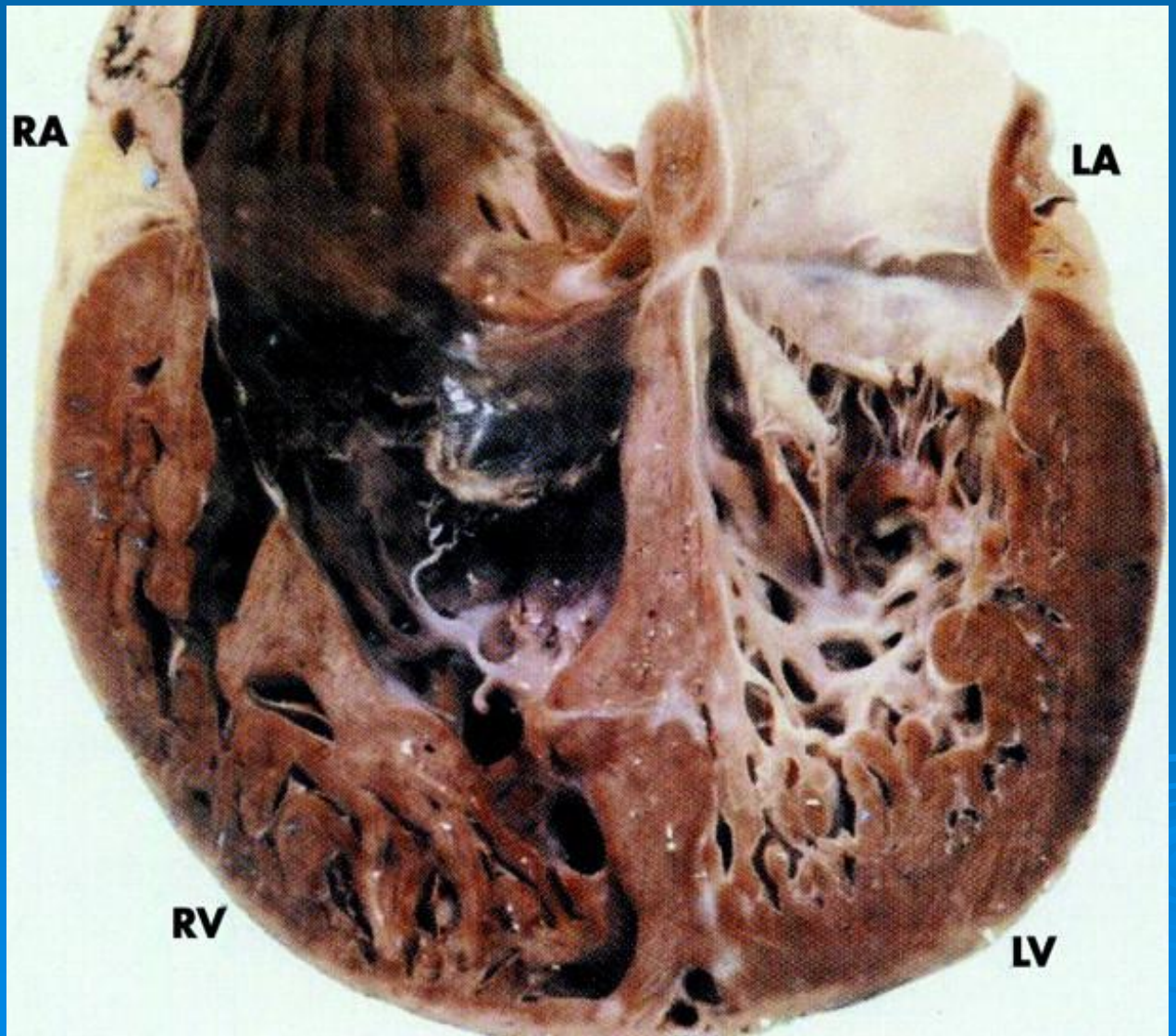
- Trabecule proeminente in “deget de manusa”
- Benzi trabeculare de peste 2 mm grosime
- Recese intertrabeculare

- Zona trabeculara trebuie sa fie dublul zonei compactate pt Dg.





Macroscopic

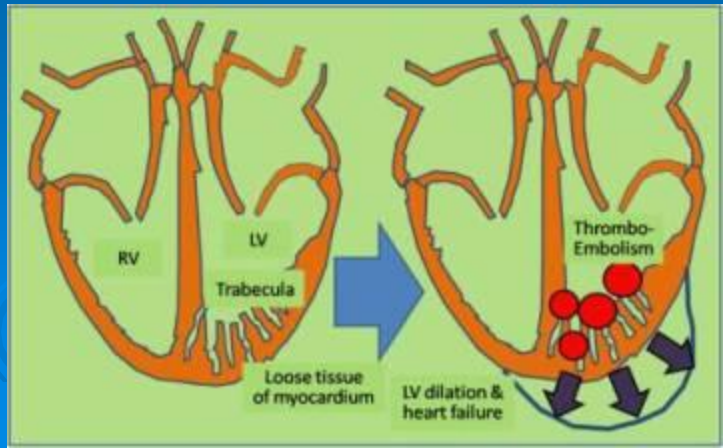


Epidemiologie

- Foarte rara:
  - 0.12 la 100.000 ???

- Se confunda cu:
  - CMR, CMD, IC, CM iskemica

- Produce disfunctie diastolica si sistolica
- Apar evenimente trombotice



Asocieri

- Cardiace
  - ALCAPA
  - Atrezie de pulmonara
  - Stenoze
  - Obstructii ventriculare stg/dr
  - VS hipoplazic
  - Insuficienta mitrala
- Neuromusculare
  - Distrofie musculara Becker
  - Miopatie mitocondriala
  - Polineuropatie si miopatie metabolica
- Genetice
  - Distrofie musculara Emery-Dreifuss
  - Cardiomiopatie miotubulara
  - Sindrom Barth

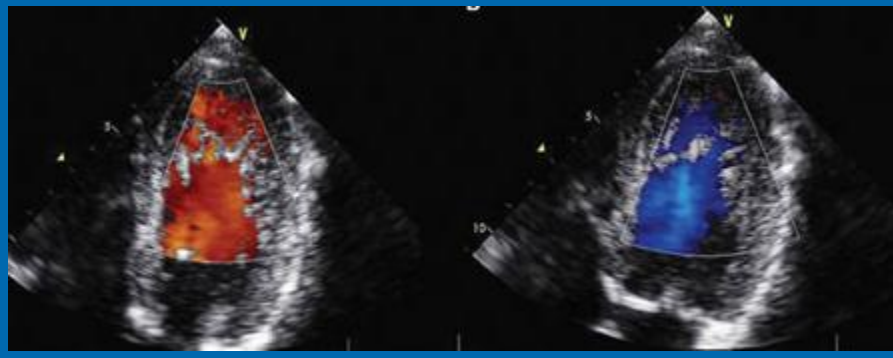
Clinic

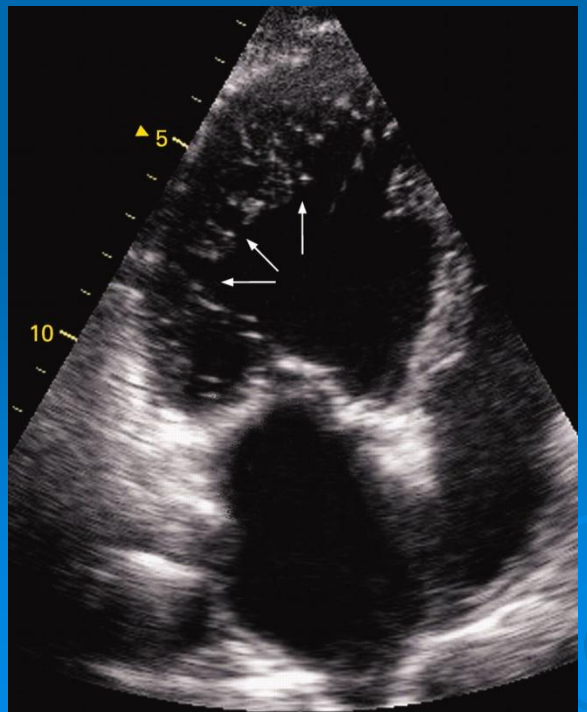
- Variaza de la asimptomatic la simptomatic
  - Astenie
  - Dispnee
  - Intoleranta fizica

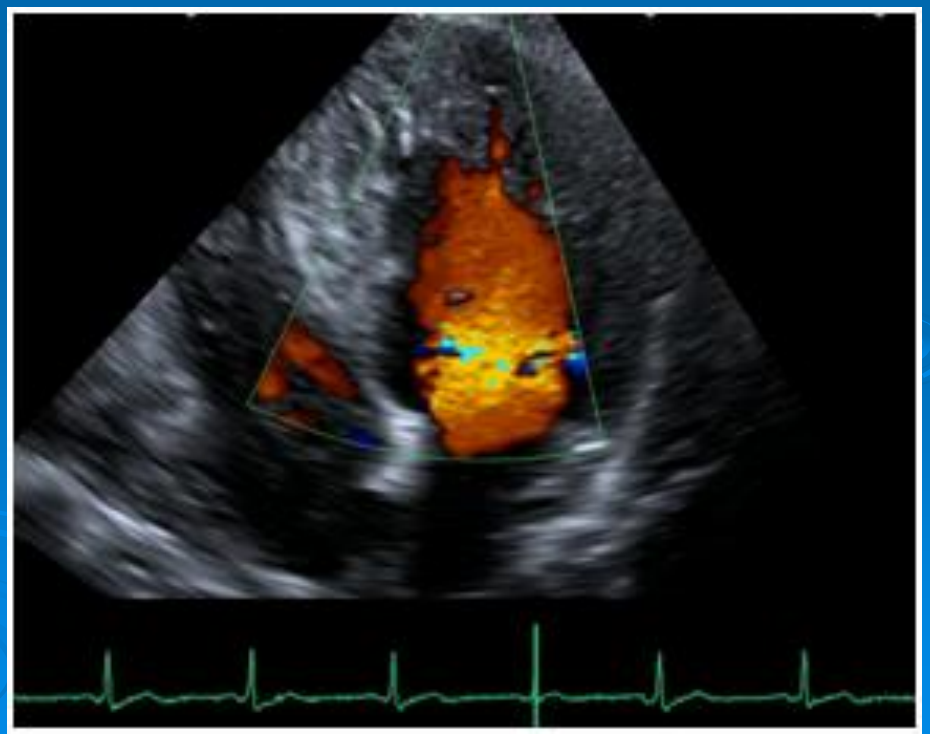
- Evolutia este in timp:
  - Disfunctie diastolica de VS
  - Disfunctie sistolica de VS
  - Insuficienta cardiaca
  - Aritmii
  - Moarte subita prin tromboembolism

Echocardiac

- Noncompactare VS
- Trabeculatii
- Recese intertrabeculare

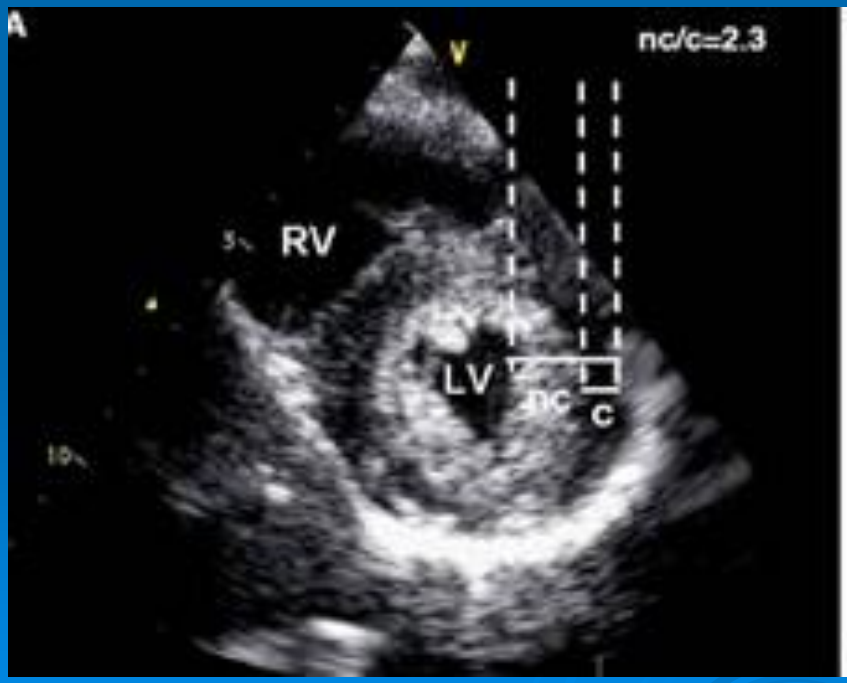


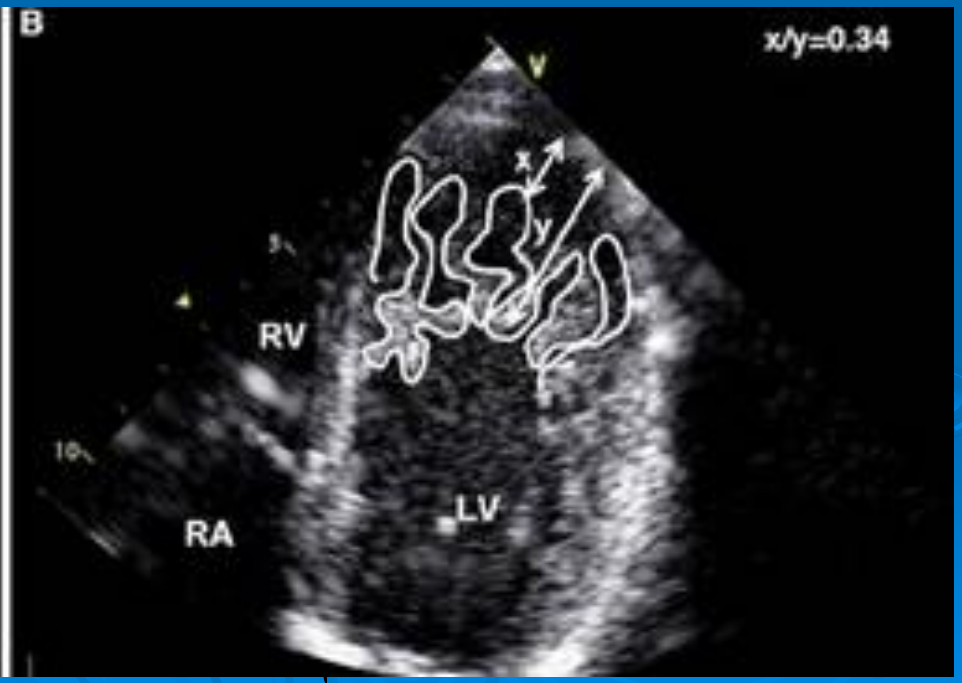




Echocardiac

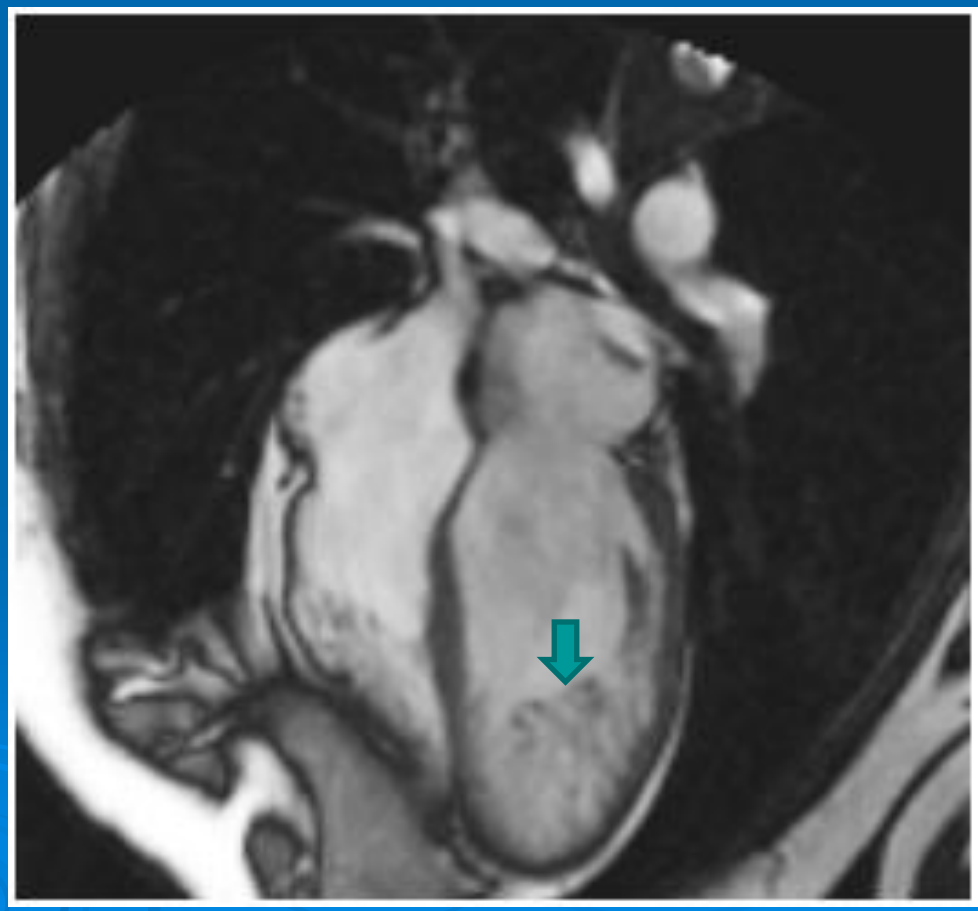
- Criterii Petersen
  - NonCompactare/Compactare 2.3





RMN

- Noncompactare VS
- Trabeculatii
- Recese intertrabeculare



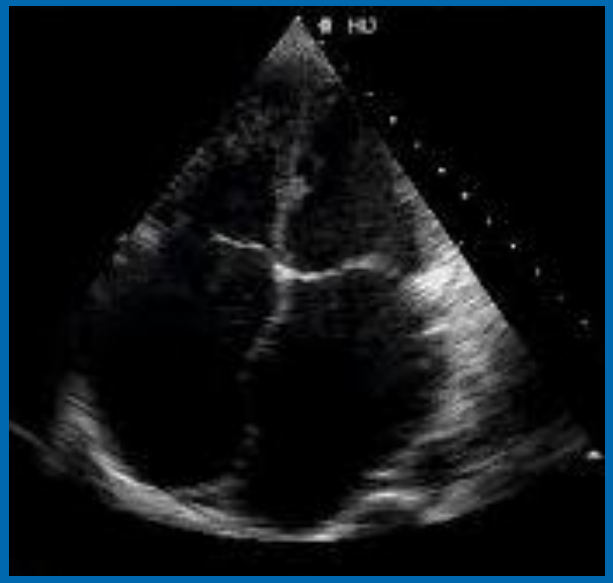
Tratament

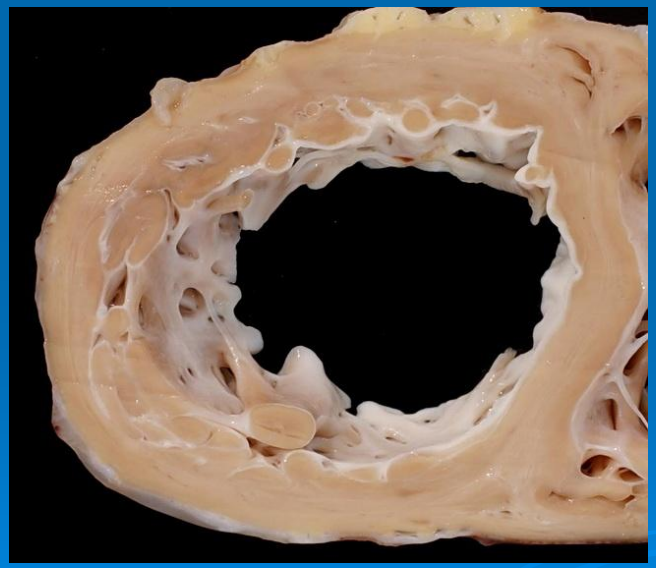
- Betablocanti
- Inhibitori ACE
- Aspirina – antiagregant
- Antiaritmice in aritmii +/- pacemaker

Prognostic

- Necunoscut, boala fiind relativ noua

- Evolutie:
  - lent progresiva spre insuficienta cardiaca
  - tromboembolism si deces





Va multumesc!