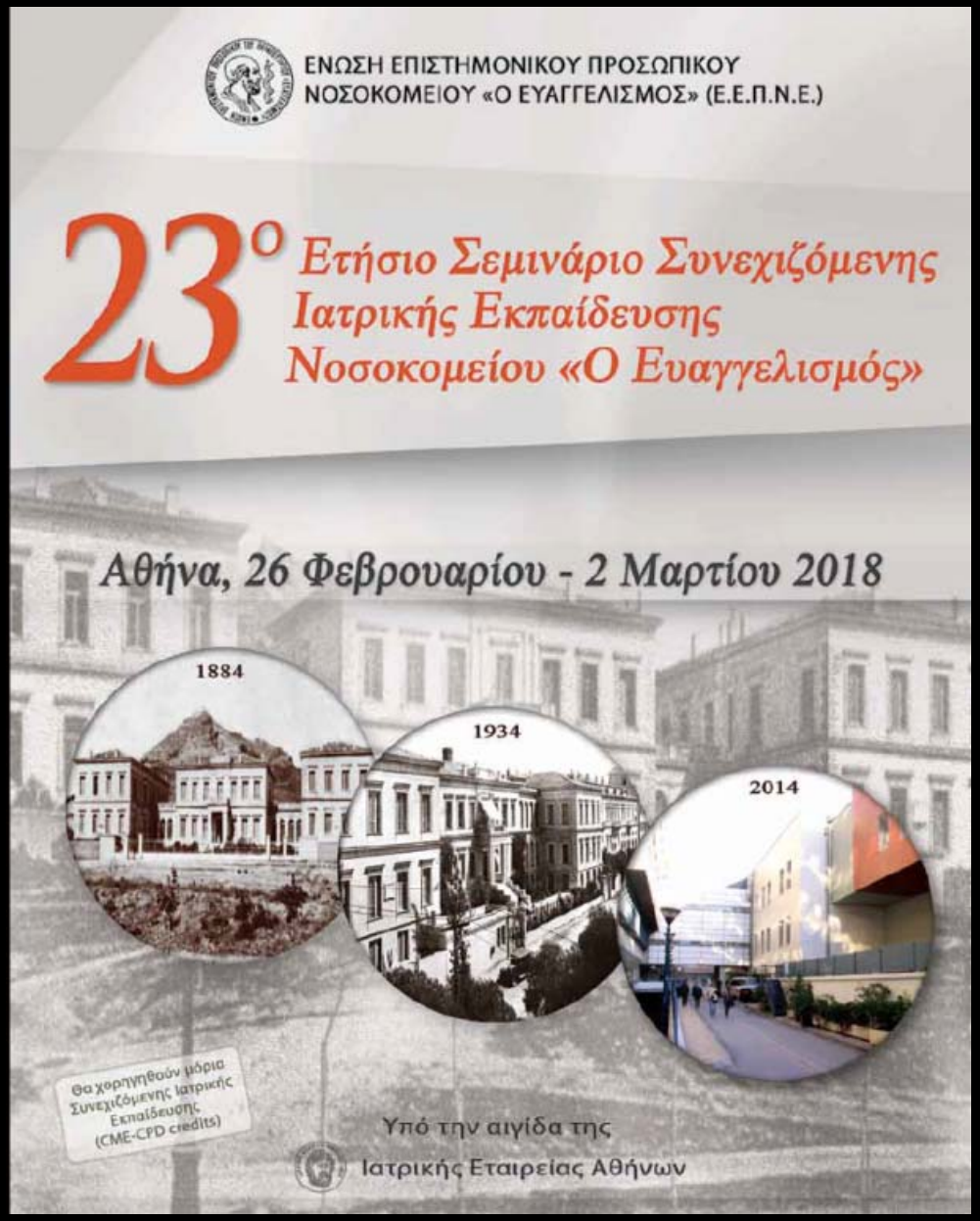


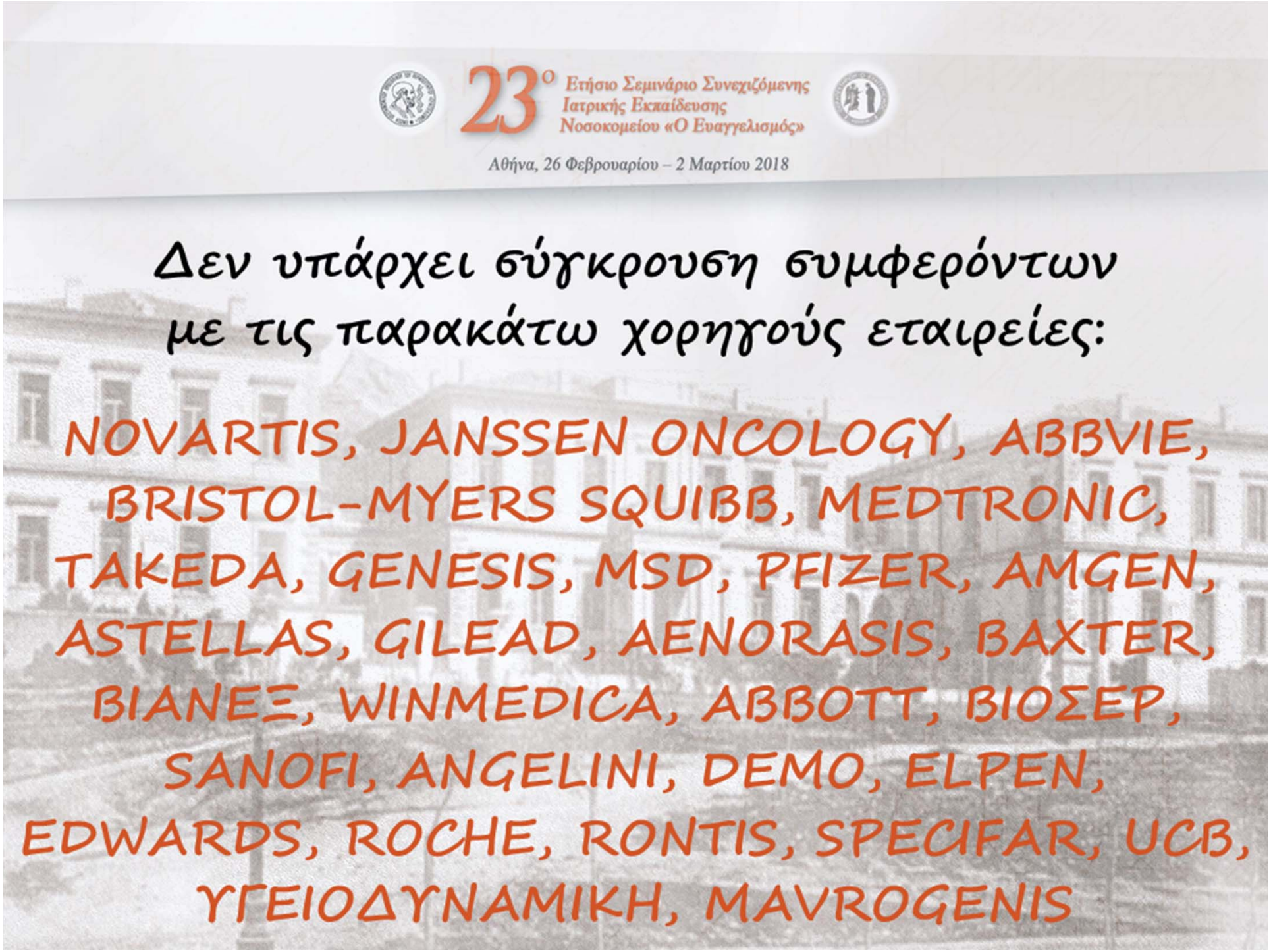

# ΕΚΠΑΙΔΕΥΤΙΚΟ ΣΥΜΠΟΣΙΟ :

Καρδιολογικά ειδικά θέματα
για μη ειδικούς

## *Τι πρέπει να γνωρίζει ο μη καρδιολόγος για τις μυοκαρδιοπάθειες*

ΕΦΗ Ι. ΠΡΑΠΠΑ
Καρδιολόγος

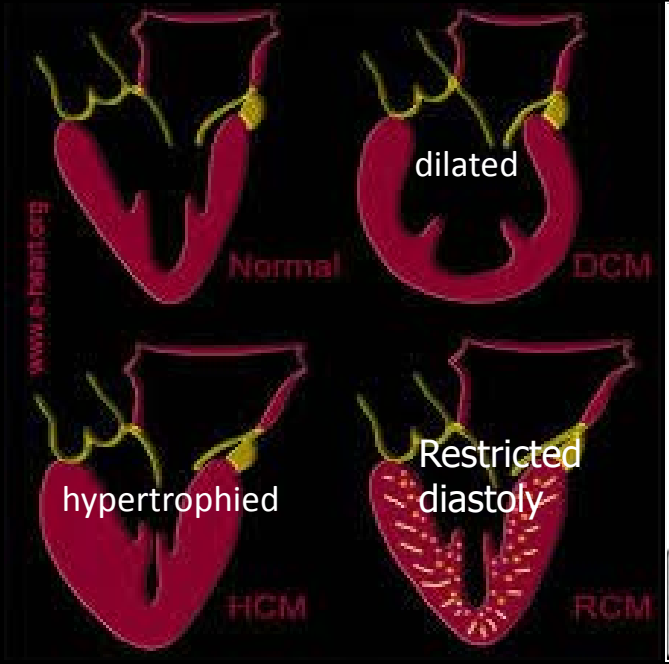
*Β' Καρδιολογική Κλινική, ΠΓΝΑ «Ο ΕΥΑΓΓΕΛΙΣΜΟΣ»*

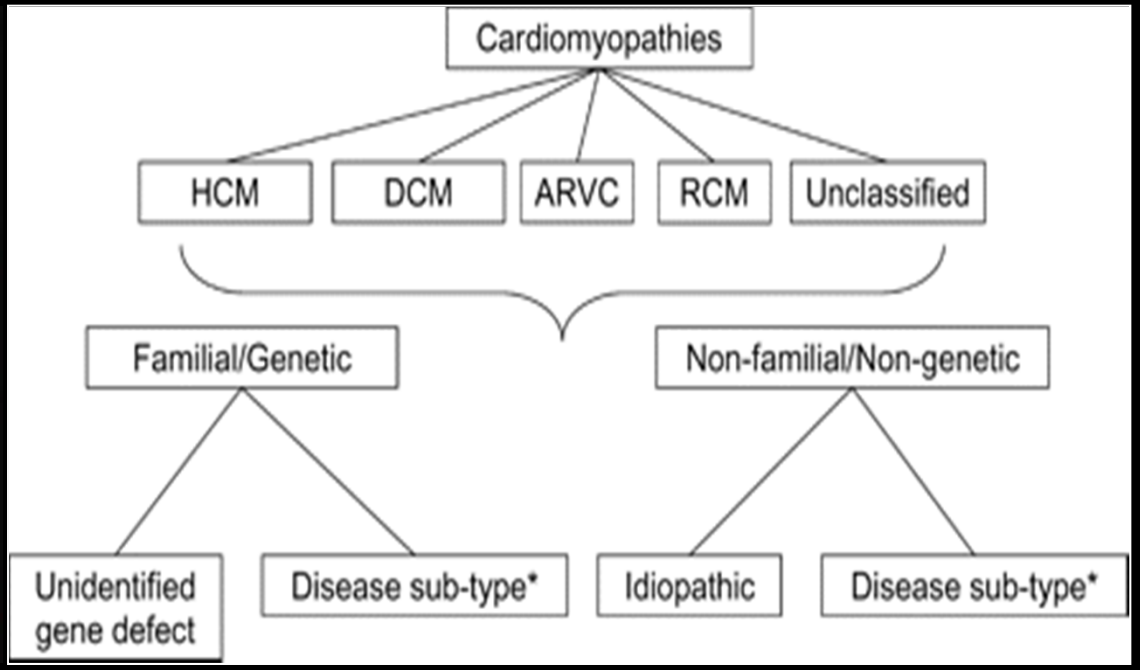

23ο Ετήσιο Σεμινάριο Συνεχιζόμενης Ιατρικής Εκπαίδευσης Νοσοκομείου «Ο Ευαγγελισμός»
Αθήνα, 26 Φεβρουαρίου – 2 Μαρτίου 2018
Δεν υπάρχει σύγκρουση συμφερόντων με τις παρακάτω χορηγούς εταιρείες:
NOVARTIS, JANSSEN ONCOLOGY, ABBVIE, BRISTOL-MYERS SQUIBB, MEDTRONIC, TAKEDA, GENESIS, MSD, PFIZER, AMGEN, ASTELLAS, GILEAD, AENORASIS, BAXTER, BIANEΞ, WINMEDICA, ABBOTT, BIOΣΕΡ, SANOFI, ANGELINI, DEMO, ELPEN, EDWARDS, ROCHE, RONTIS, SPECIFAR, UCB, ΥΓΕΙΟΔΥΝΑΜΙΚΗ, MAVROGENIS

**CLASSIFICATION OF THE CARDIOMYOPATHIES:**
a position statement from the ESC Working Group on myocardial and pericardial diseases

*Eur Heart Journal 2008*

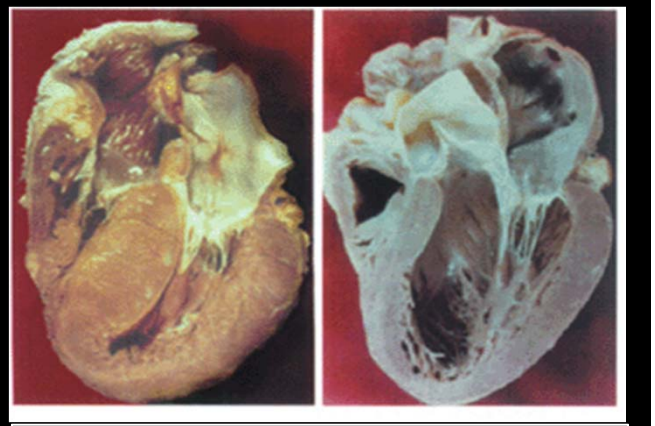
"...we define a cardiomyopathy as:
A myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of CAD, hypertension, valvular disease and CHD sufficient to cause the observed myocardial abnormality"



# ΥΠΕΡΤΡΟΦΙΚΗ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ

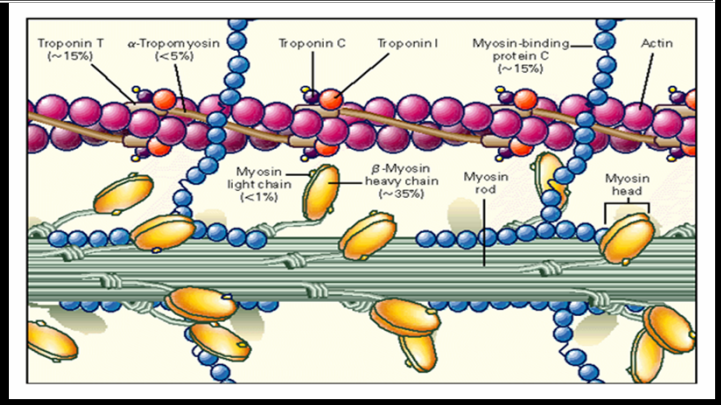
"...HCMs are simply defined by the presence of hypertrophy in the absence of loading conditions (HPT, valve HD) sufficient to cause the observed abnormality."

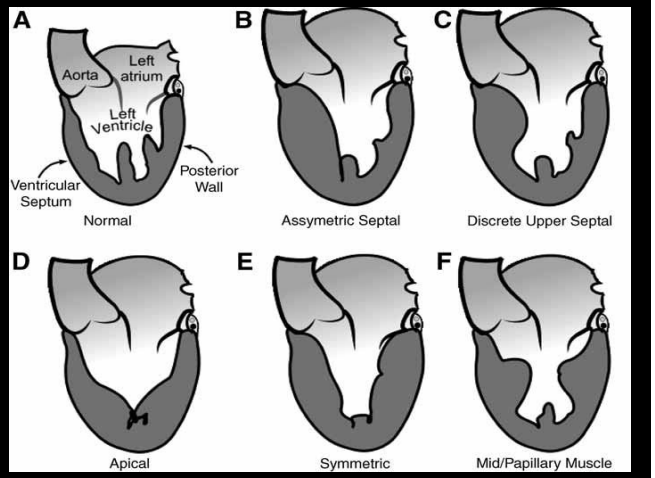


DIAGNOSIS : hypertrophy

Familial (55%) autosomal dominant transmission
Mutations genes encoding proteins of cardiac sarcomere

| | |
|---|---|
| Familial | Familial, unknown gene |
| | Sarcomeric protein mutations |
| | ß myosin heavy chain |
| | Cardiac myosin binding protein C |
| | Cardiac troponin I |
| | Troponin-T |
| | α-tropomyosin |
| | Essential myosin light chain |
| | Regulatory myosin light chain |
| | Cardiac actin |
| | α-myosin heavy chain |
| | Titin |
| | Troponin C |
| | Muscle LIM protein |
| | Glyco... (e.g. Pompe; PRKAG2, Forbes', Danon) |
| | Lysosomal storage diseases (e.g. Anderson–Fabry, Hurler's) |
| | Disorders of fatty acid metabolism |
| | Carnitine deficiency |
| | Phosphorylase ß kinase deficiency |
| | Mitochondrial cytopathies |
| | Syndromic HCM |
| | Noonan's syndrome |
| | LEOPARD syndrome |
| | Friedreich's ataxia |
| | Beckwith–Wiedemann syndrome |
| | Swyer's syndrome |
| | Other |
| | Phospholamban promoter |
| | Familial amyloid |
| Non-familial | Obesity |
| | Infants of diabetic mothers |
| | Athletic training |
| | Amyloid (AL/prealbumin) |



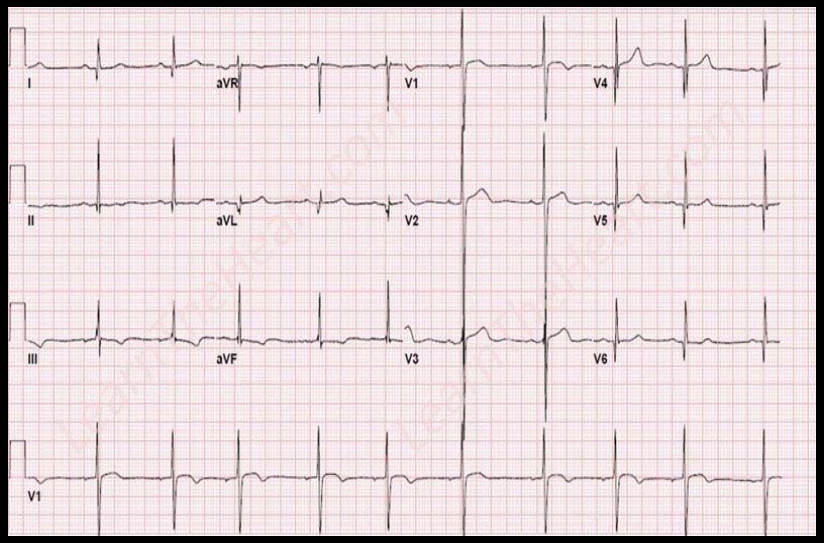

Males ≥ 15mm
Females ≥ 13mm

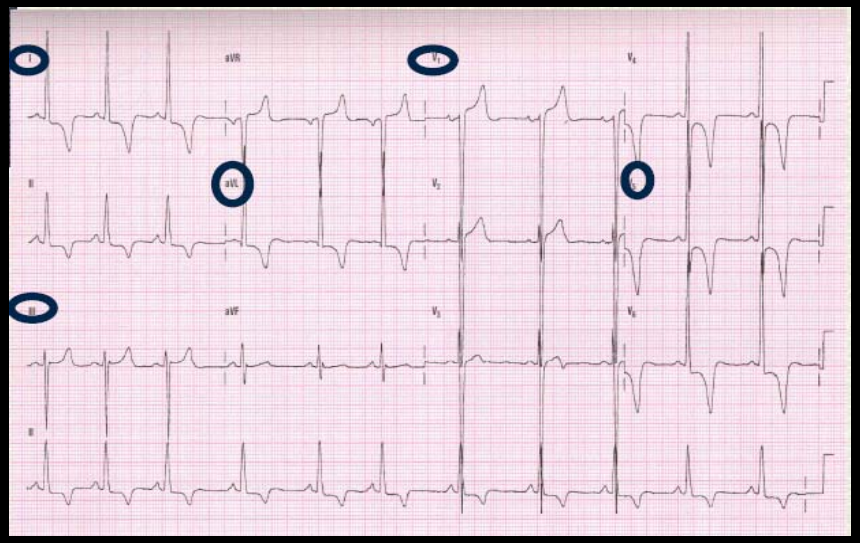
# *ECG always abnormal…*

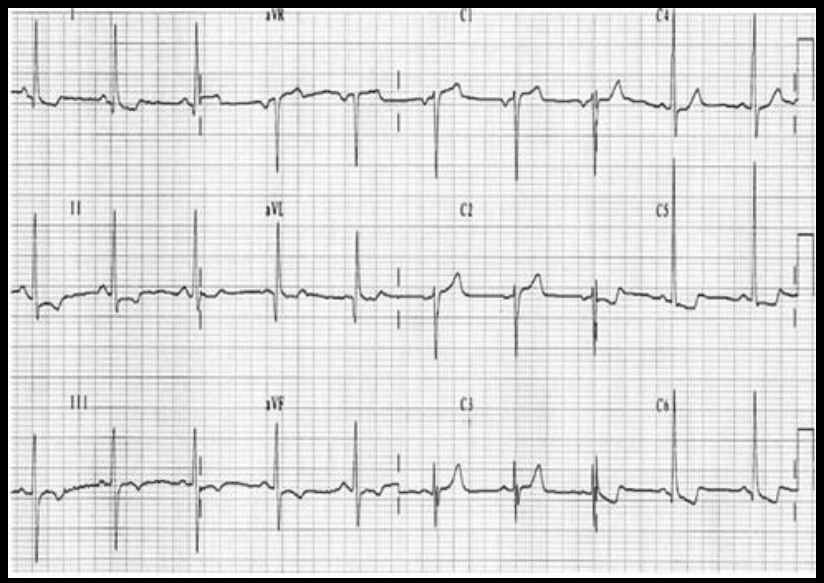
R-wave in AVL >11mm;

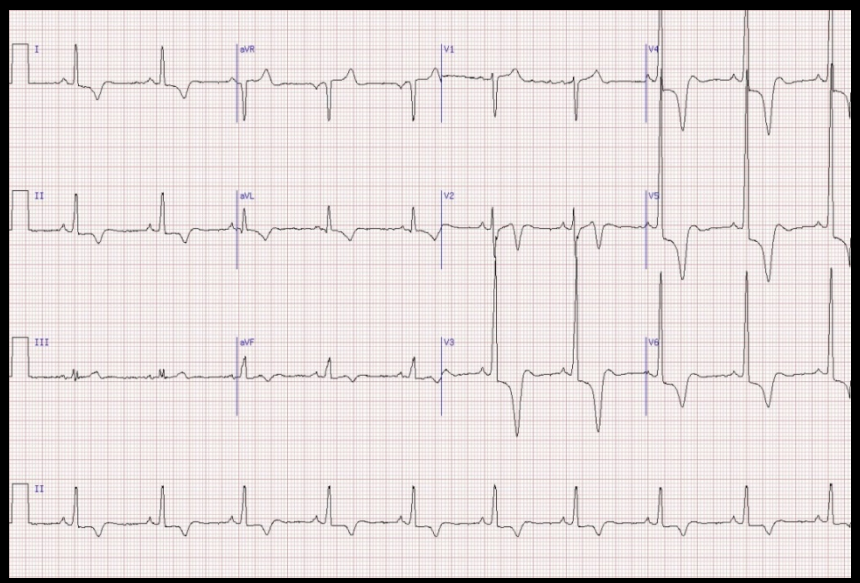
R wave height in Lead I plus the S wave depth in Lead III > 25 mm

*S wave depth in V1 plus the height in V5 that exceeds 35 mm

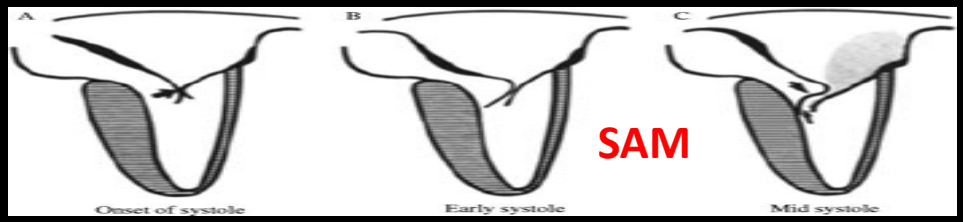


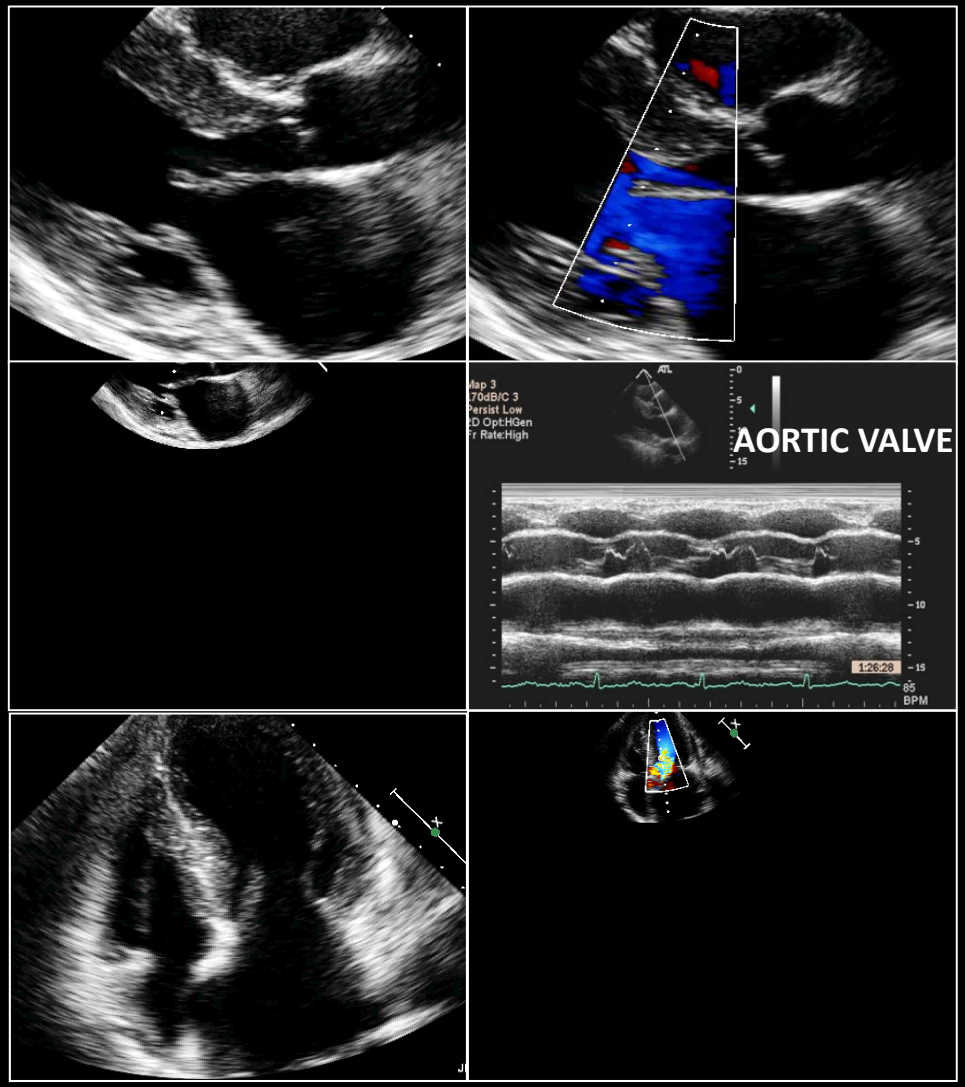







## HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY

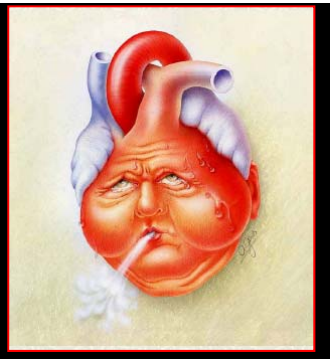



*Dyspnea on exertion*

*Syncope*

*Angina*



***WORSENING.....***

- ✸ contractility (exercise, positive inotropes)
- ✸ heart rate (exercise, fever, ✸ CO)
- ✵ preload (hypovolemia, sepsis, fluid shifts)
- loss of atrial kick (atrial fibrillation, AVB, ventricular arrhythmias)

Maintain normal sinus rhythm

If atrial fibrillation : convert pharmacologically / electrically

Avoid hypotension, vasodilators, dehydration, strenuous exercise, sepsis, chemical withdrawal, shivering, seizures

Avoid : digitalis, diuretics, nitrates and vasodilators

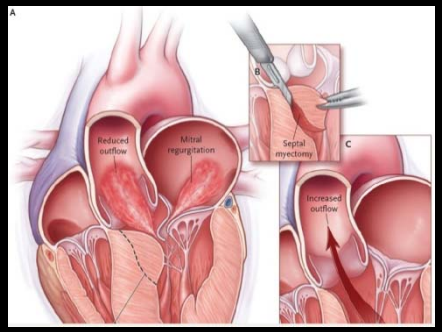
# ΘΕΡΑΠΕΥΤΙΚΟΙ ΣΤΟΧΟΙ

## Β. ΣΥΜΠΤΩΜΑΤΑ

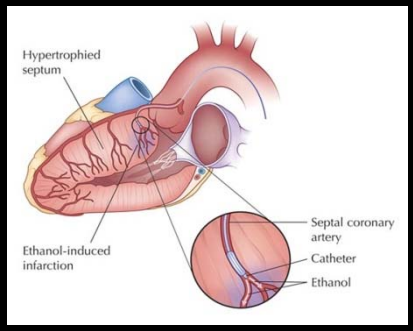
μείωση απόφραξης

- b-blockers
- disopyramide
- pacemaker

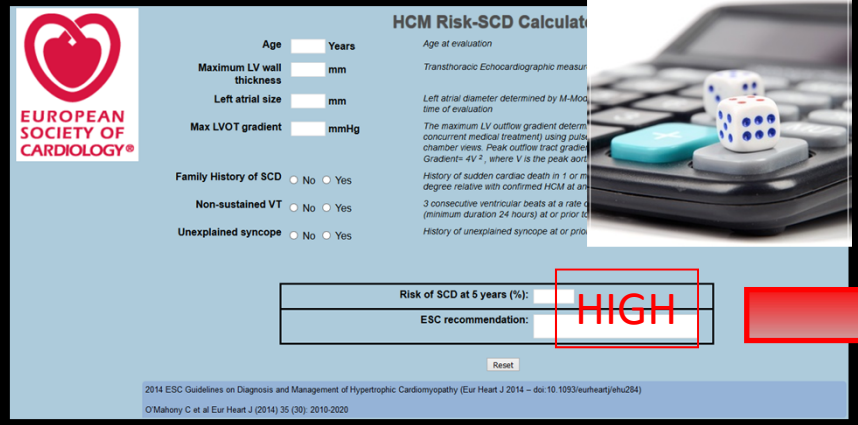
Surgical Septal Myectomy




Alcohol Septal Ablation

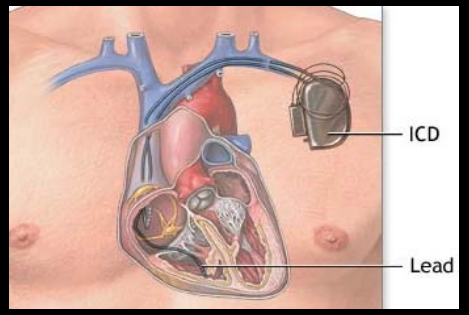



## Γ. ΑΙΦΝΙΔΙΟΣ ΘΑΝΑΤΟΣ






HIGH

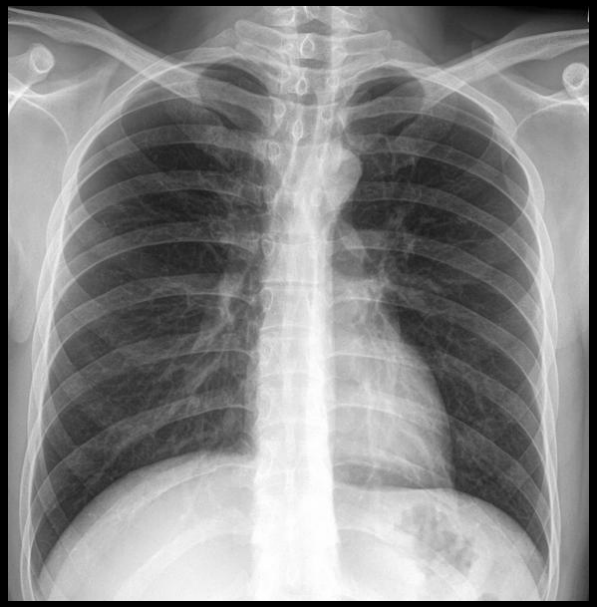


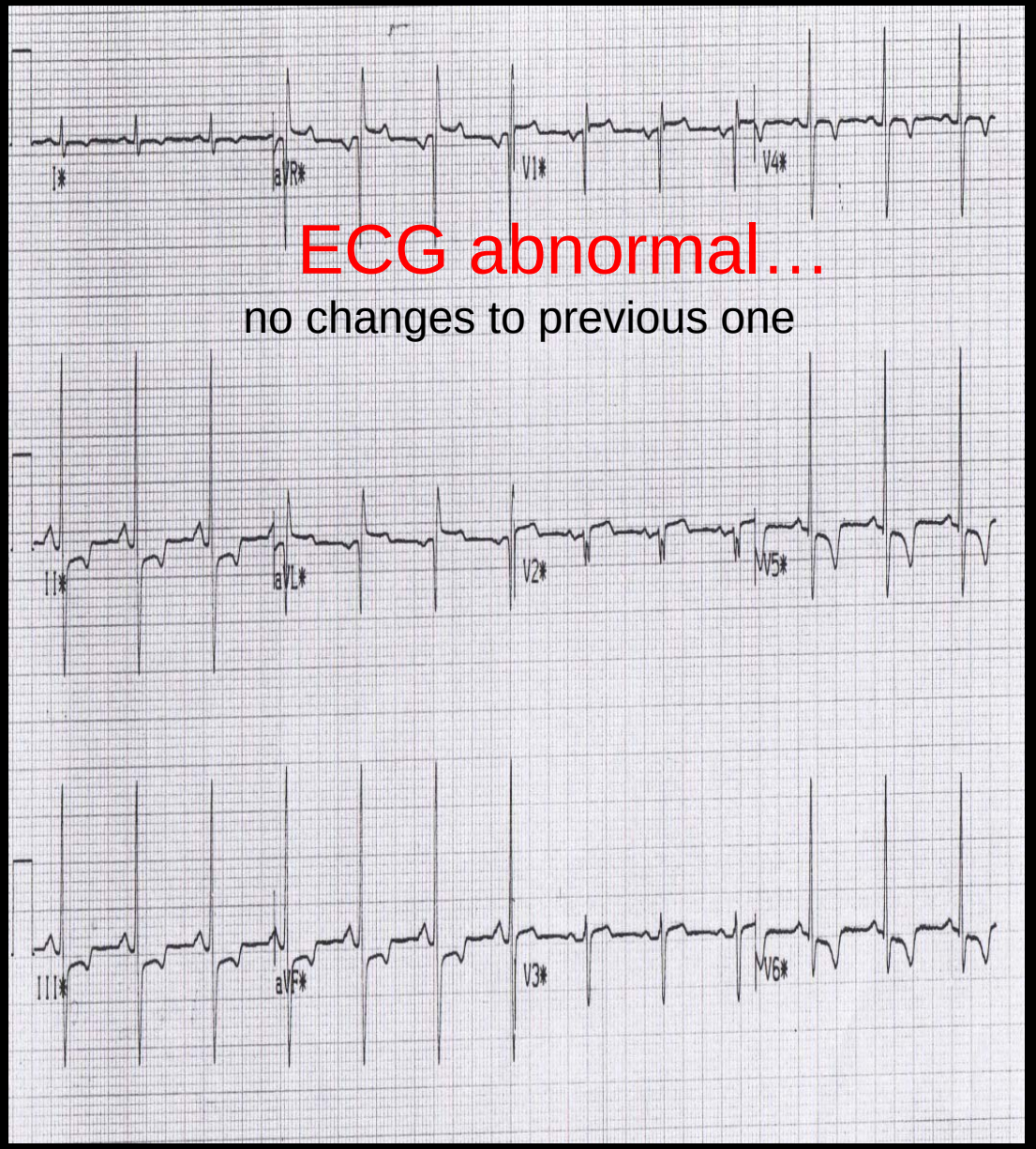

## Δ. ΕΛΕΓΧΟΣ ΟΙΚΟΓΕΝΕΙΑΣ

*patient …*

Male, 55 years old
pre-surgical evaluation
for cholesystectomy

- asymptomatic
- smoker, dyslipidemia
- clinical examination normal
- blood tests normal
  (CPK, TnT, dDimers, proBNP)





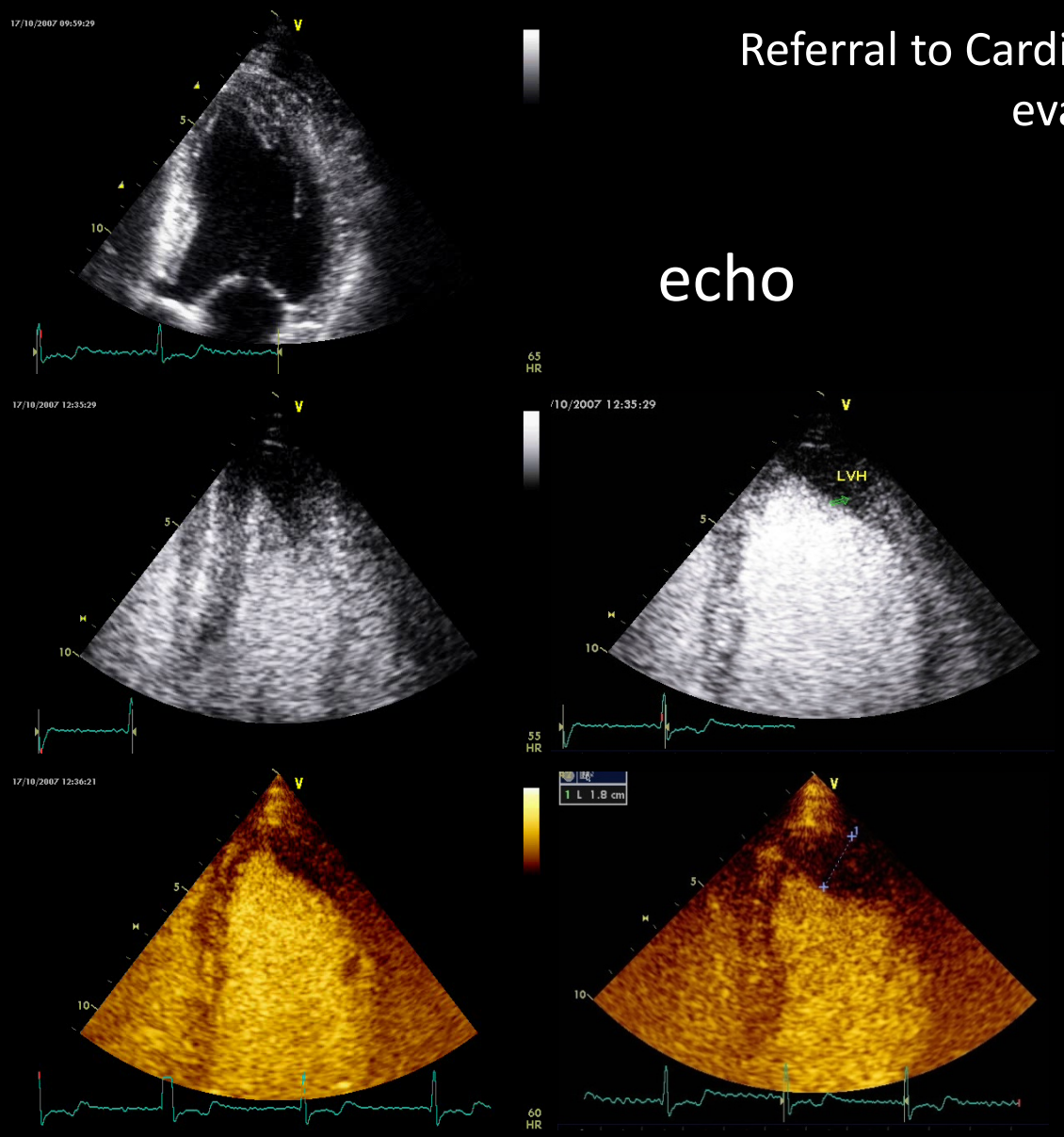

ECG abnormal…

no changes to previous one

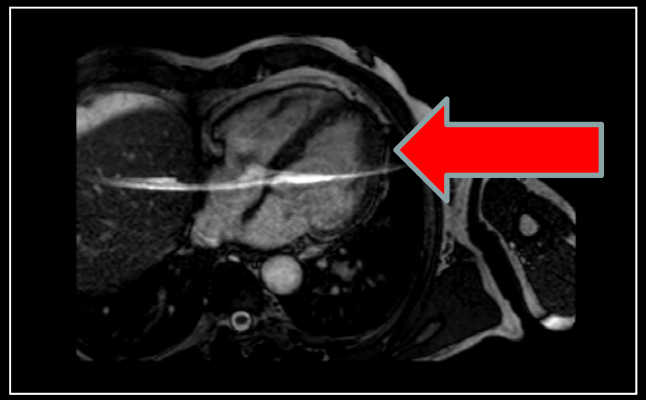
CORONARY ANGIOGRAPHY : normal

Referral to Cardiomyopathy Clinic for further evaluation….

echo



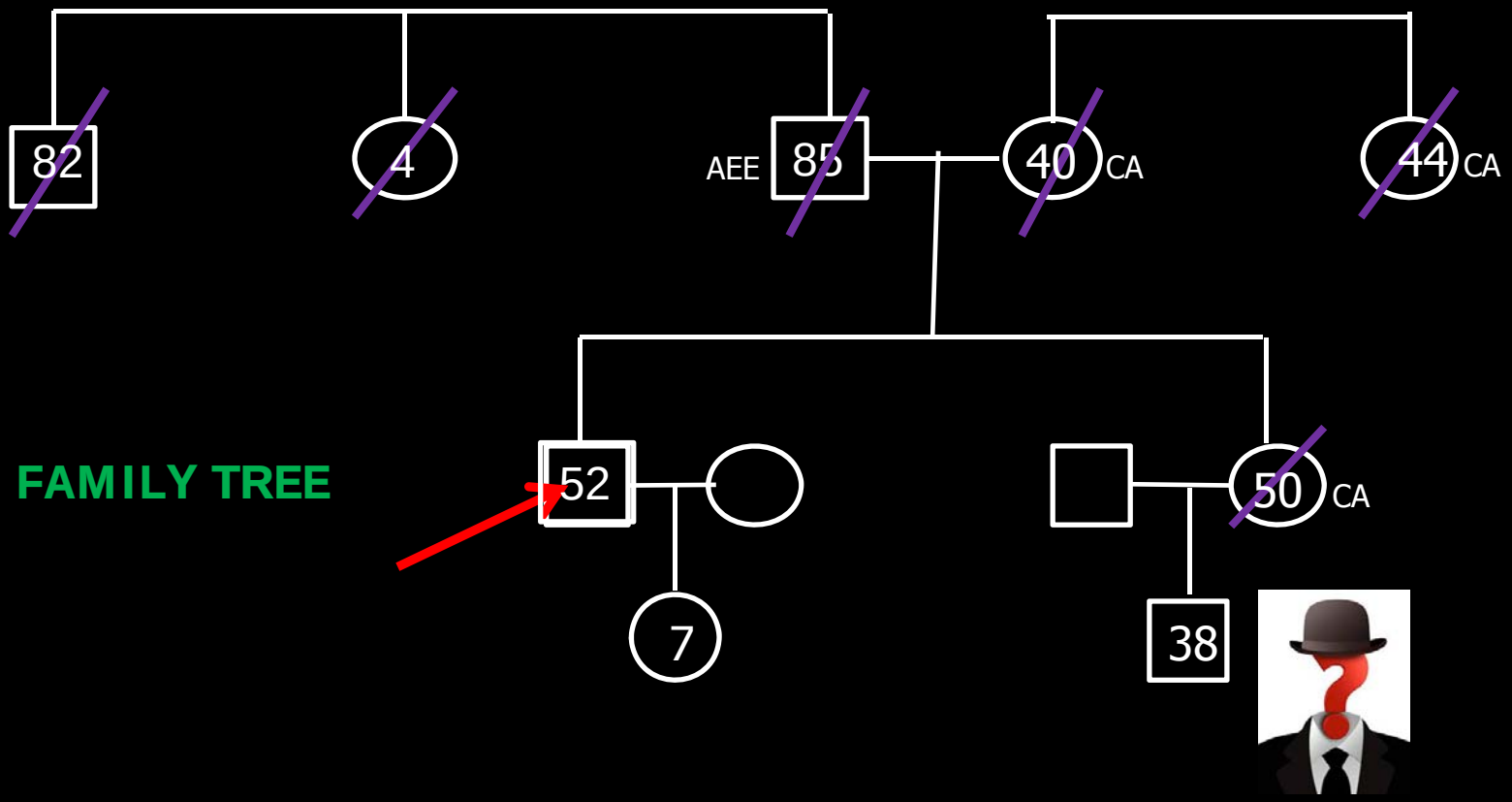

MRI



Apical HCM

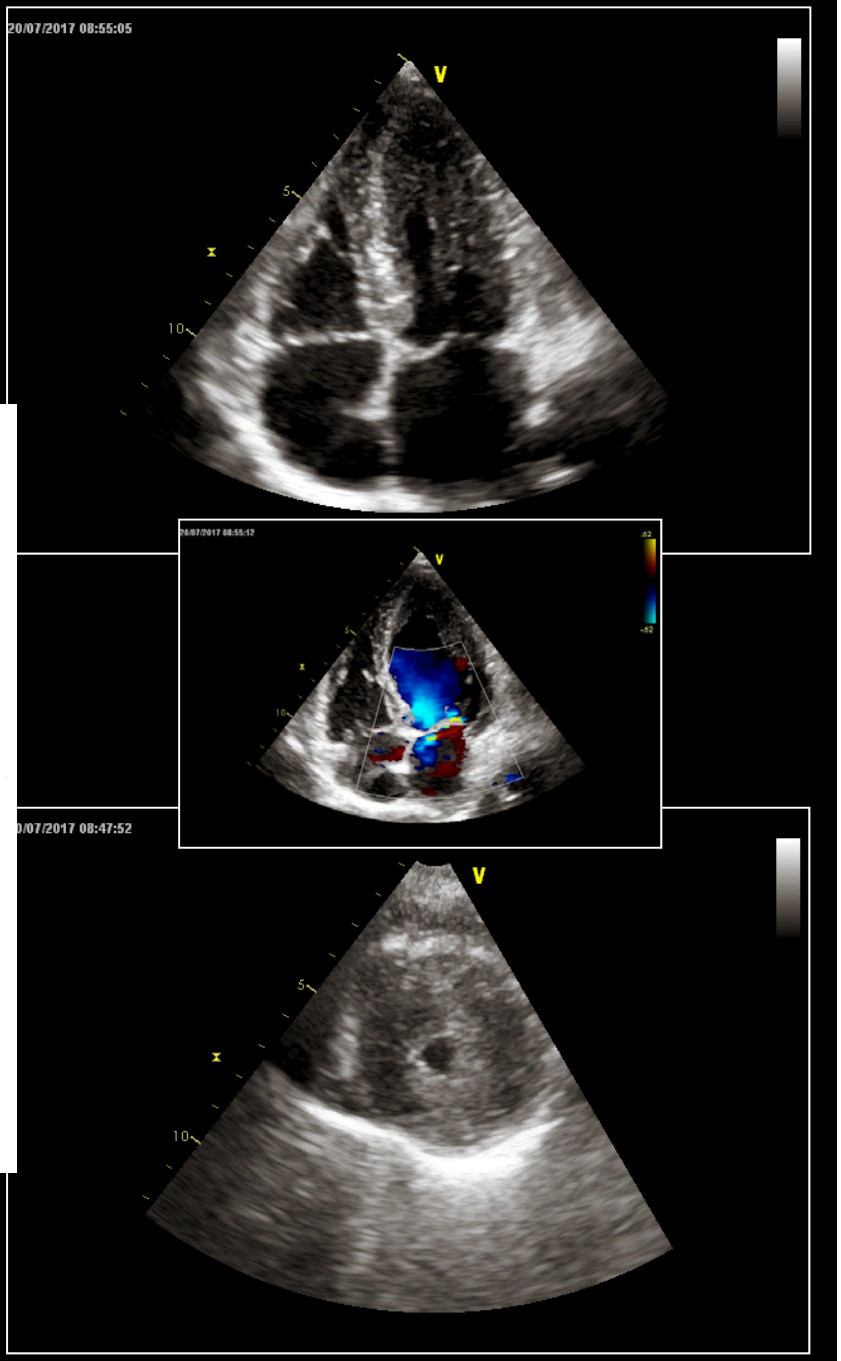
# *MANAGEMENT*

- no medication
- SD risk stratification  low (Holter, stress test…)
- 1st degree relatives screening… (nephew)



# *the nephew !!!*

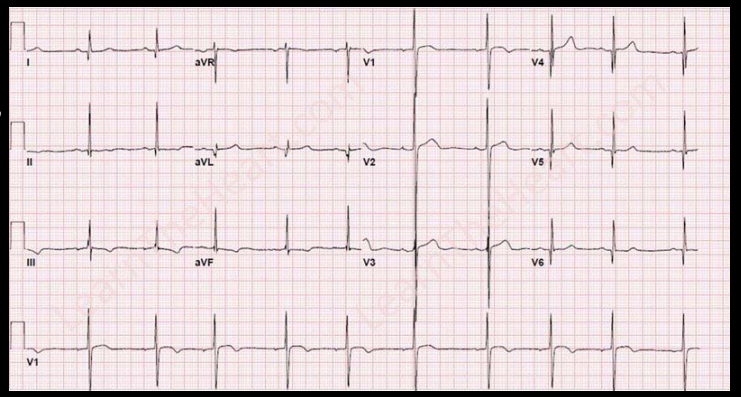

ID:
Name:
Sex:Male
Birth Date:
Years
Medication:
cm
kg
mmHg
Symptoms:
History:
26-Sep-2017 12:31
10 mm/mV 25 mm/s Filter: H50 d 35 Hz
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6


20/07/2017 08:55:05
V
5
x
10
20/07/2017 08:55:12
V
20/07/2017 08:47:52
V
5
x
10

# HCM “take home message”

A. Ανεξήγητα παθολογικό ΗΚΓ ή ανεξήγητη υπερτροφία, ειδικά σε συνδυασμό με:
   - οικογενειακό ιστορικό αιφνιδίου θανάτου
   - αρρυθμίες, συγκοπή

   θέτει την υποψία υπερτροφικής μυοκαρδιοπάθειας.



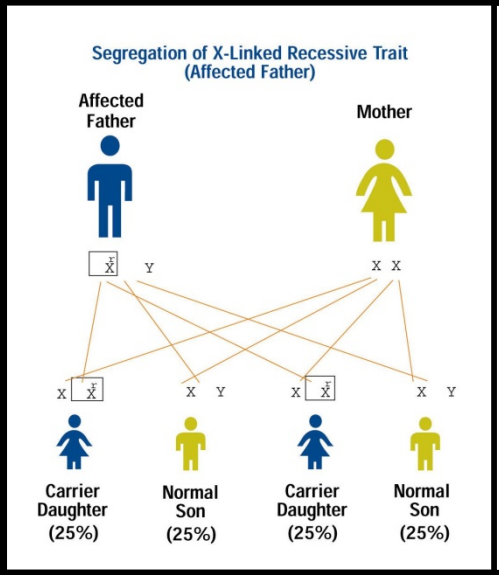

B. Ο έλεγχος της οικογένειας επιβεβλημένος.
   Μπορεί να εντοπιστούν «αμέριμνοι» ασθενείς που κινδυνεύουν όμως από αιφνίδιο θάνατο…

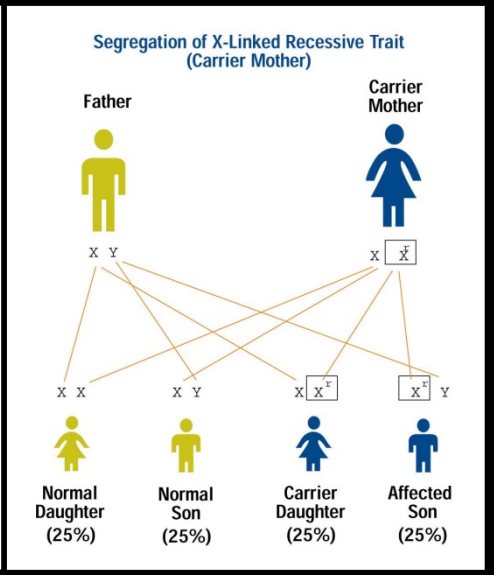

***Η έγκαιρη διάγνωση σώζει ζωές…***

| | HCM |
|---|---|
| Familial | Familial, unknown gene |
| | Sarcomeric protein mutations: |
| | ß myosin heavy chain |
| | Cardiac myosin binding protein C |
| | Cardiac troponin I |
| | Troponin-T |
| | α-tropomyosin |
| | Essential myosin light chain |
| | Regulatory myosin light chain |
| | Cardiac actin |
| | α-myosin heavy chain |
| | Titin |
| | Troponin C |
| | Muscle LIM protein |
| | Glycogen storage disease (e.g. Pompe, PRKAG2, Forbes', Danon) |
| | Lysosomal storage diseases (e.g. Anderson–Fabry, Hurler's) |
| | Disorders of fatty acid metabolism |
| | Carnitine deficiency |
| | Phosphorylase B kinase deficiency |
| | Mitochondrial cytopathies |
| | Syndromic HCM |
| | Noonan's syndrome |
| | LEOPARD syndrome |
| | Friedreich's ataxia |
| | Beckwith–Wiedemann syndrome |
| | Swyer's syndrome |
| | Other |
| | Phospholamban promoter |
| | Familial amyloid |
| Non-familial | Obesity |
| | Infants of diabetic mothers |
| | Athletic training |
| | Amyloid (AL/prealbumin) |

# FABRY ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ

Υπολειπόμενη φυλοσύνδετη κληρονομικότητα
X-linked recessive



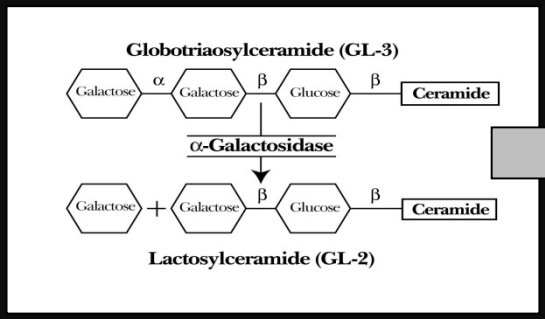

Άντρας πάσχων, γυναίκα φορέας

Πατέρας: πάντα υγιείς γιούς και φορείς κόρες

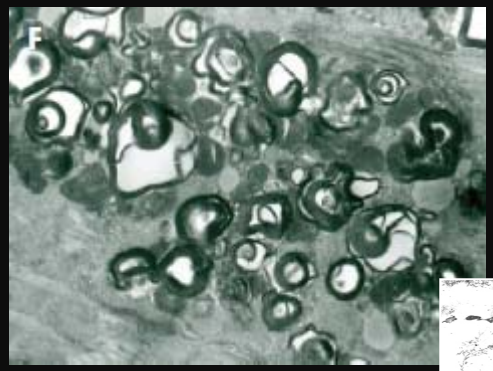
Μητέρα: από 25% όλες οι εκδοχές

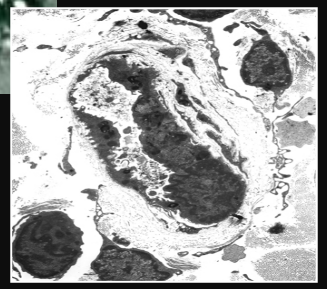
# *FABRY DISEASE*

familial storage disease (metabolic cardiomyopathy)
caused by a deficiency of the lysosomal enzyme A-galactosidase A (a-Gal)




substrate accumulation



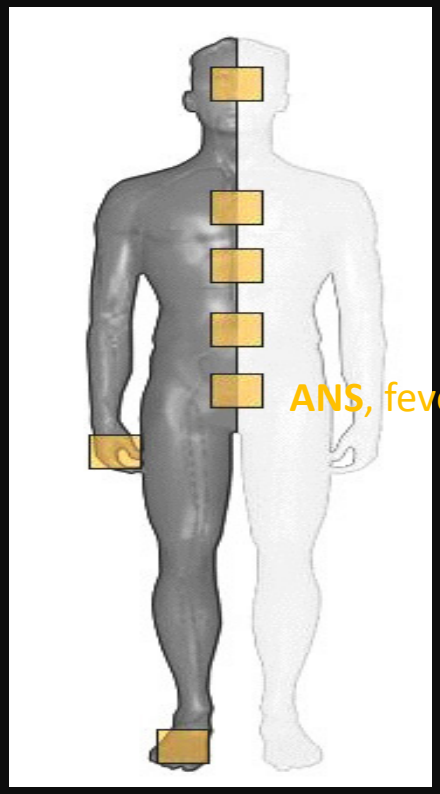


"vacuolization" in lysosomes

Stroke, ear, eye, CNS

heart

renal



ANS, fever, hypohidrosis, heat intolerance

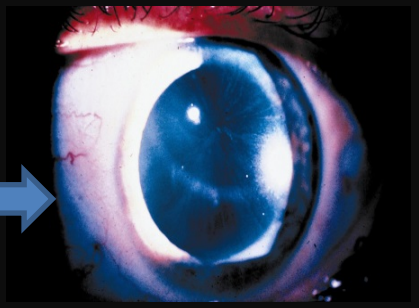
angiokeratoma

acroparesthesia

Prevalence
4-12% HCM

## CHILDHOOD

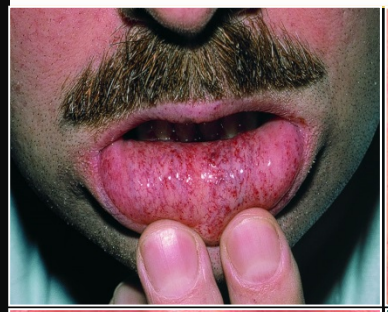
episodic pain crises, acroparesthesia
hypohidrosis
corneal and lenticular opacities
recurrent fever
heat and cold intolerance

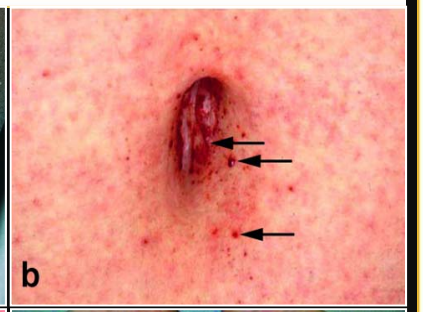


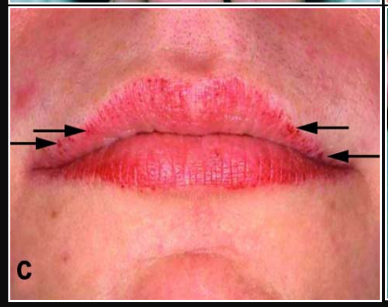
Θολερότης φακού και κερατοειδούς, με φυσιολογική όραση
«Fabry cataract»
•Split-lamp

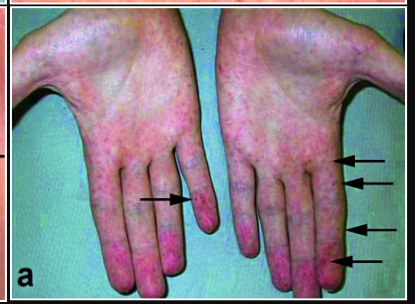
## ADOLESCENCE

angiokeratomas
fatigue
episodic pain crises, acroparesthesia
hypohidrosis
corneal and lenticular opacities
recurrent fever
heat and cold intolerance





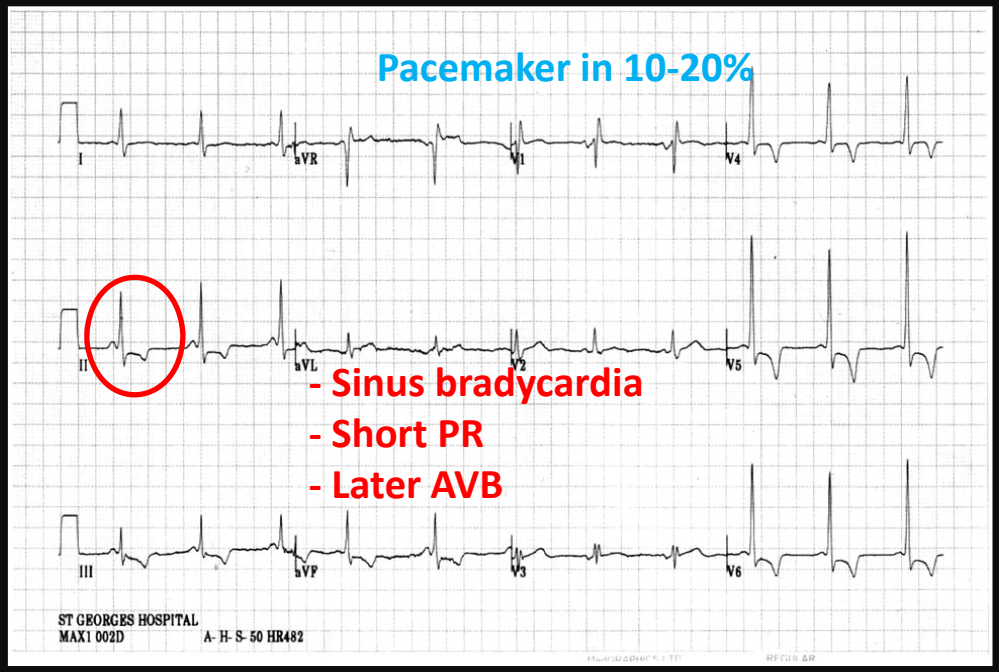




## ADULTHOOD

*Life expectancy: 40yrs*

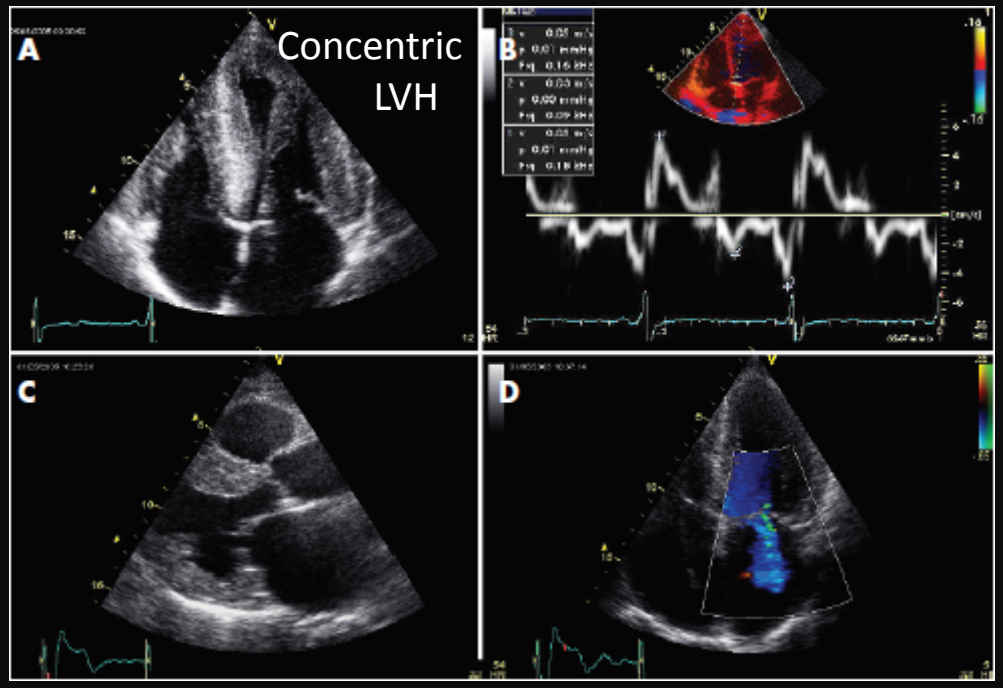
renal dysfunction
neurological complications
cerebrovascular disease
cardiac dysfunction
hearing loss and tinnitus
angiokeratomas
fatigue
episodic pain crises, acroparesthesia
hypohidrosis
corneal and lenticular opacities
recurrent fever
heat and cold intolerance

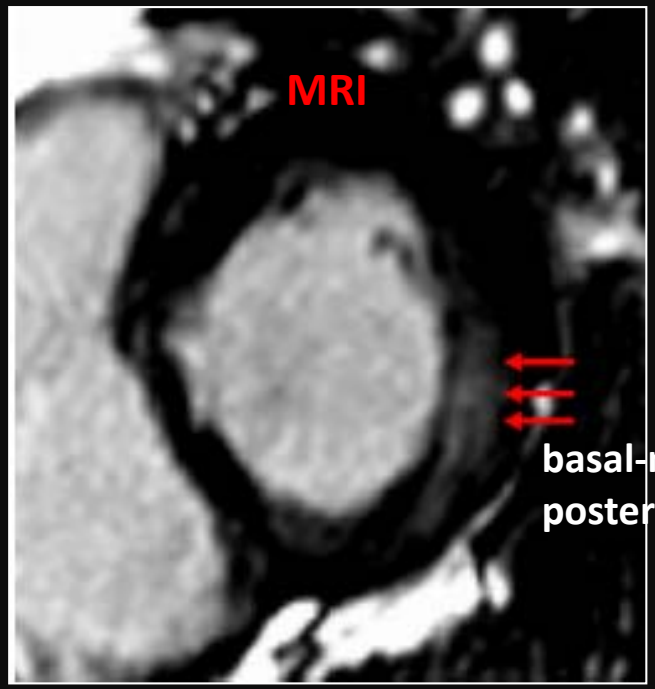
*Delay in diagnosis : 14 years!!!*




*DIAGNOSIS*

- αGAL activity
- biopsy
- Genetic analysis



## Therapy of the Fabry cardiomyopathy

*Enzyme replacement therapy*

Enzyme replacement therapy allows a causal treatment of Fabry disease. Intravenous infusion with recombinant alpha-galactosidase A replaces the missing enzyme and catabolizes the lipid deposits [9,10]. The ERT infusion has to be given intravenously every two weeks throughout life. Therefore, it is essential to carefully establish the diagnosis and define the indication for ERT. ERT

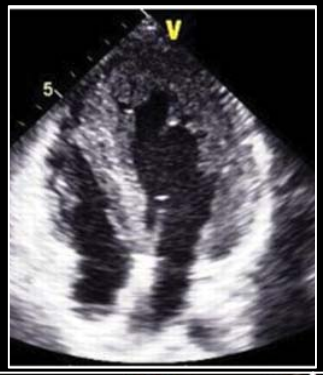
Distinguish FABRY from HCM is crusial!

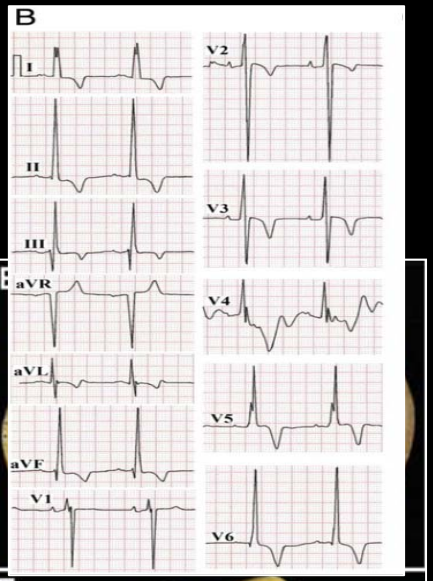
# *DANON DISEASE*

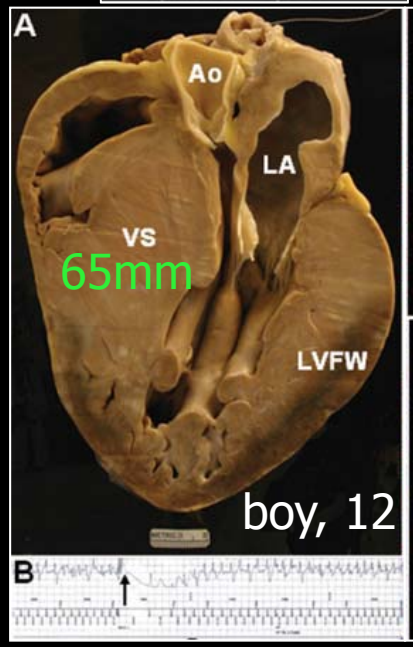
X-linked dominant lysosomal disease,
due to a primary deficiency of
lysosome-associated membrane protein 2 caused
by several mutations
in the LAMP2 gene
Phenotype:
cardiomyopathy (88%, HCM), skeletal myopathy
mental retardation (70%).

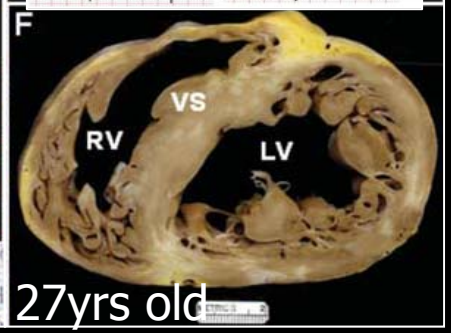
Danon is characterized by early morbidity
and limited life expectancy ,
with survival beyond 25yrs uncommon!

The DANON REGISTRY reports the following
disease landmarks on average:
First symptoms 12yrs, rapid deterioration (6mo),
transplant 18yrs, death without transplant 19yrs...






There is no specific treatment for Danon disease, except heart transplantation!
ICD fails to terminate lethal ventricular tachyarrhythmias in most Danon pts.

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

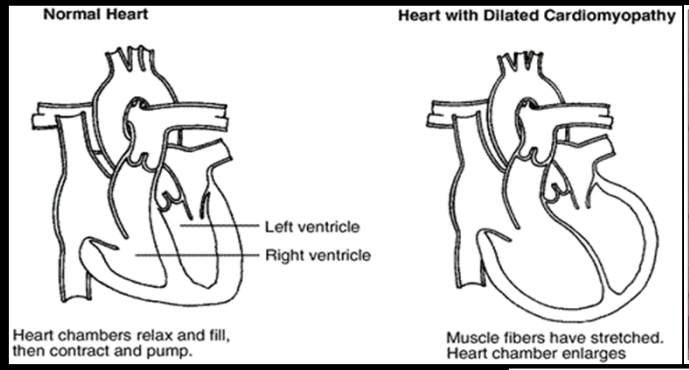
Myocarditis (infective/toxic/immune)
Kawasaki disease
Eosinophilic (Churg Strauss syndrome)
Viral persistence
Drugs
Pregnancy
Endocrine
Nutritional — thiamine, carnitine, selenium, hypophosphataemia, hypocalcaemia
Alcohol
Tachycardiomyopathy

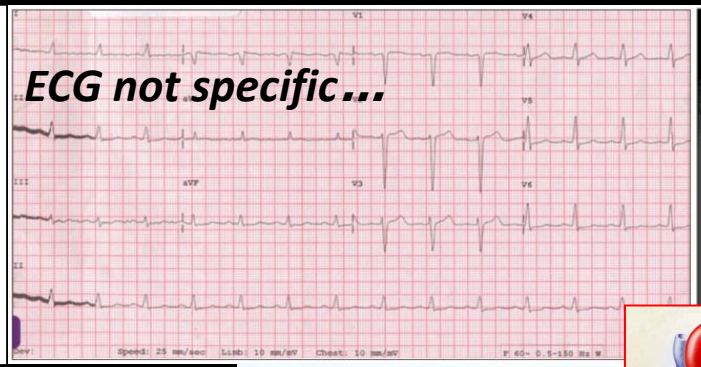
# ΔΙΑΤΑΤΙΚΗ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ

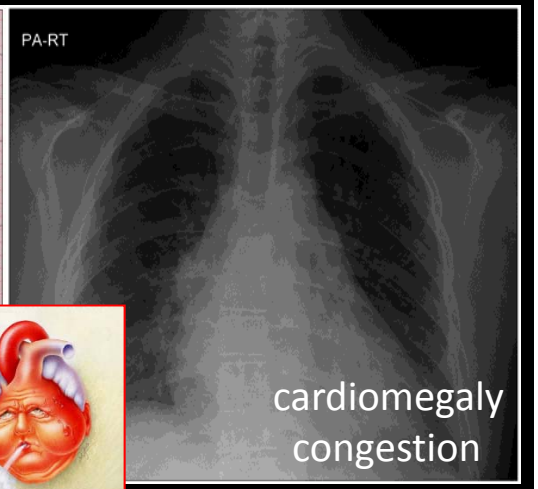
"DCM is defined by the presence of LV dilatation and LV systolic dysfunction in the absence of abnormal loading conditions (hypertension, valve disease) or CAD sufficient to cause global systolic impairment."

Κριτήρια MESTRONI (EHJ 1999)
LVEDD>117% predicted *
EF<45%, FS<25%

Henry equation
$EDD = 45.3 \times BSA^{1/3} - 0.03 \times age - 7.2$

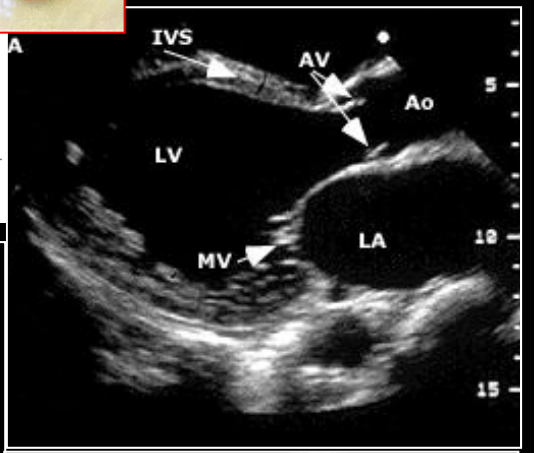


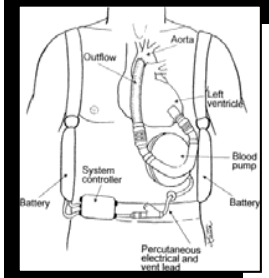




- chronic fatigue ; weakness
- orthopnea ; paroxysmal nocturnal dyspnea (PND)
- cough ; chest pain
- weight gain
- palpitations
- dizziness ; syncope
- impotence
- insomnia






## ΘΕΡΑΠΕΥΤΙΚΟΙ ΣΤΟΧΟΙ

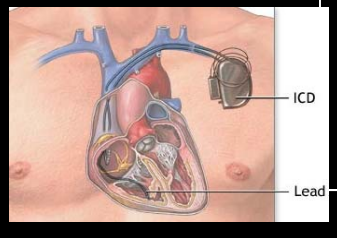
Α. ΑΝΑΔΕΙΞΗ ΥΠΟΚΕΙΜΕΝΗΣ ΝΟΣΟΥ

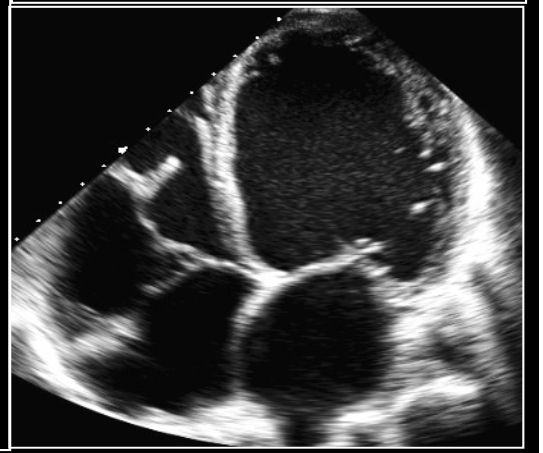
Β. ΣΥΜΠΤΩΜΑΤΑ (καρδιακή ανεπάρκεια)

- α. φάρμακα (διουρητικά, aMEA…)
- β. Αμφικοιλιακός βηματοδότης
- γ. LVAD (LV assist device)

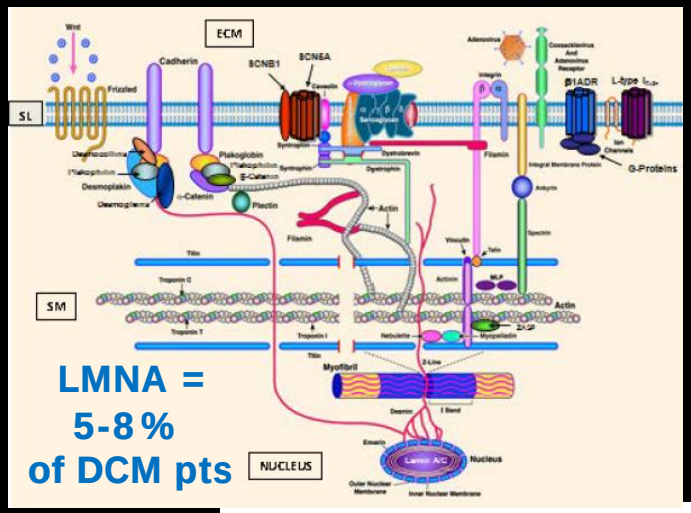
Γ. ΑΙΦΝΙΔΙΟΣ ΘΑΝΑΤΟΣ

- α. εμφυτεύσιμος απινιδωτής ICD

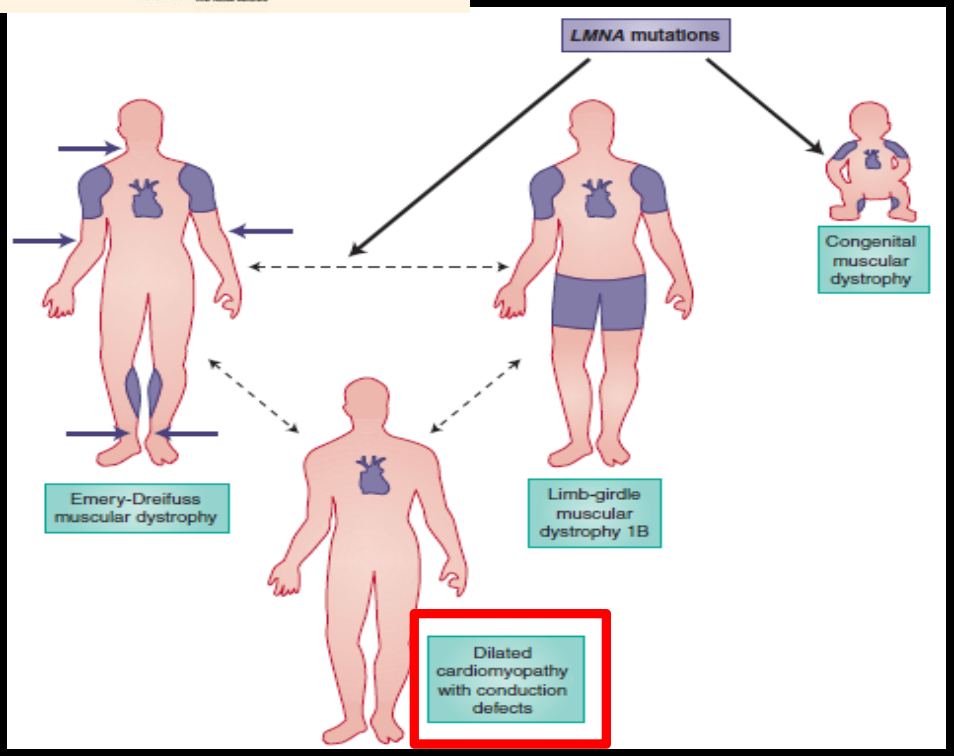


# *LAMINOPATHIES*




The LMNA gene encodes nuclear lamin A and C, intermediate filament proteins that are components of the nuclear lamina.




Lu. LMNA. Disease Models and Mechanisms 2011

## Box 1. Clinical entities caused by *LMNA* mutations

Mutations in the single *LMNA* gene cause several defined clinical entities that can be grouped primarily into those with phenotypes selectively involving striated muscle, adipose tissue (lipodystrophy syndromes), peripheral nerve (peripheral neuropathy) or multiple systems with features of accelerated aging (progerias).

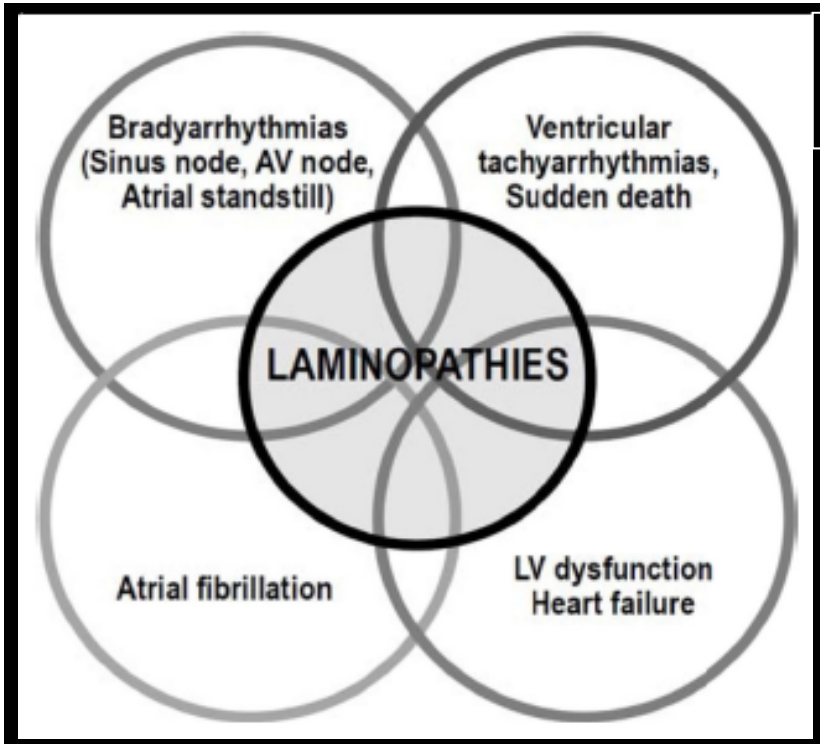
**Diseases of striated muscle (see also Fig. 1)**
- Autosomal Emery-Dreifuss muscular dystrophy
- Cardiomyopathy dilated 1A
- Limb-girdle muscular dystrophy type 1B
- Congenital muscular dystrophy

**Lipodystrophy syndromes**
- Dunnigan-type familial partial lipodystrophy
- Lipoatrophy with diabetes and other features of insulin resistance
- Insulin resistance without lipoatrophy
- Mandibuloacral dysplasia[a]

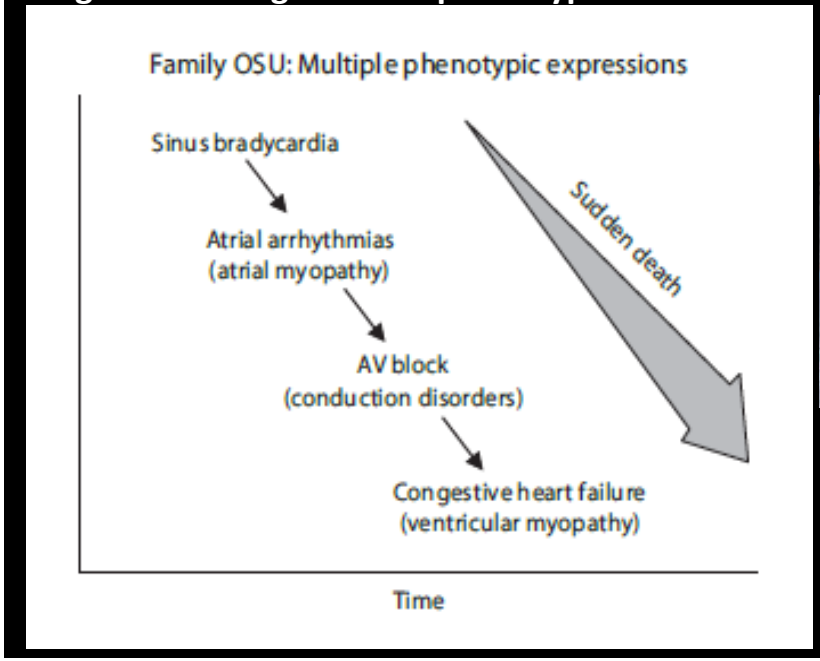
**Peripheral neuropathy**
- Charcot-Marie-Tooth disorder type 2B1

**Accelerated aging disorders (progerias)**
- Hutchinson-Gilford progeria syndrome
- Atypical Werner syndrome
- Restrictive dermopathy
- Variant progeroid disorders
- Mandibuloacral dysplasia[a]

[a]Diseases with features of both lipodystrophy and progeria




**Progression of age-related phenotypes in LMNA**



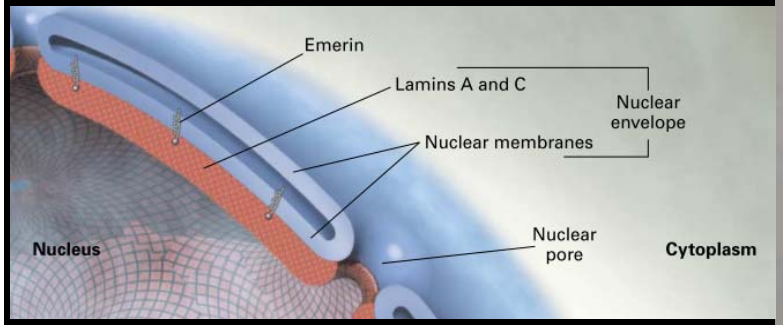

## CARDIAC INVOLVEMENT

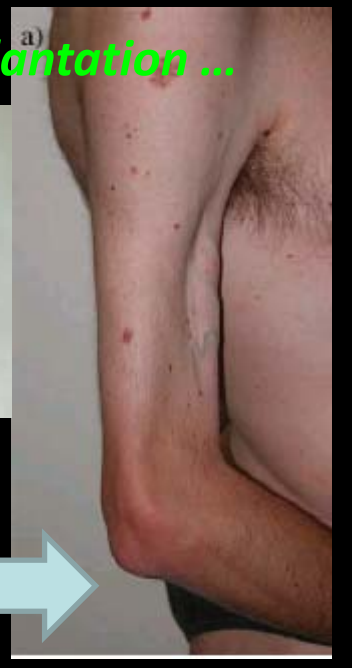
: DCM ± arryhthmias

Patients with LMNA may present a wide range of arrhythmic disturbances,
either bradyarrhytmias
(conduction disturbances and AV-blocks, sinus node dysfunction, atrial standstill)
or tachyarrhythmias (AF, VT, VF),
in variable combinations, and with frequent association with LV dysfunction and HF.

Severity of arrhythmias is usually not related to the presence and degree of neuromuscular impairment.

***early ICD implantation …***




Emery Dreifuss muscular dystrophy:
wasting of humeral muscle
+ elbow contracture
AVB, later DCM

*specific follow-up…*

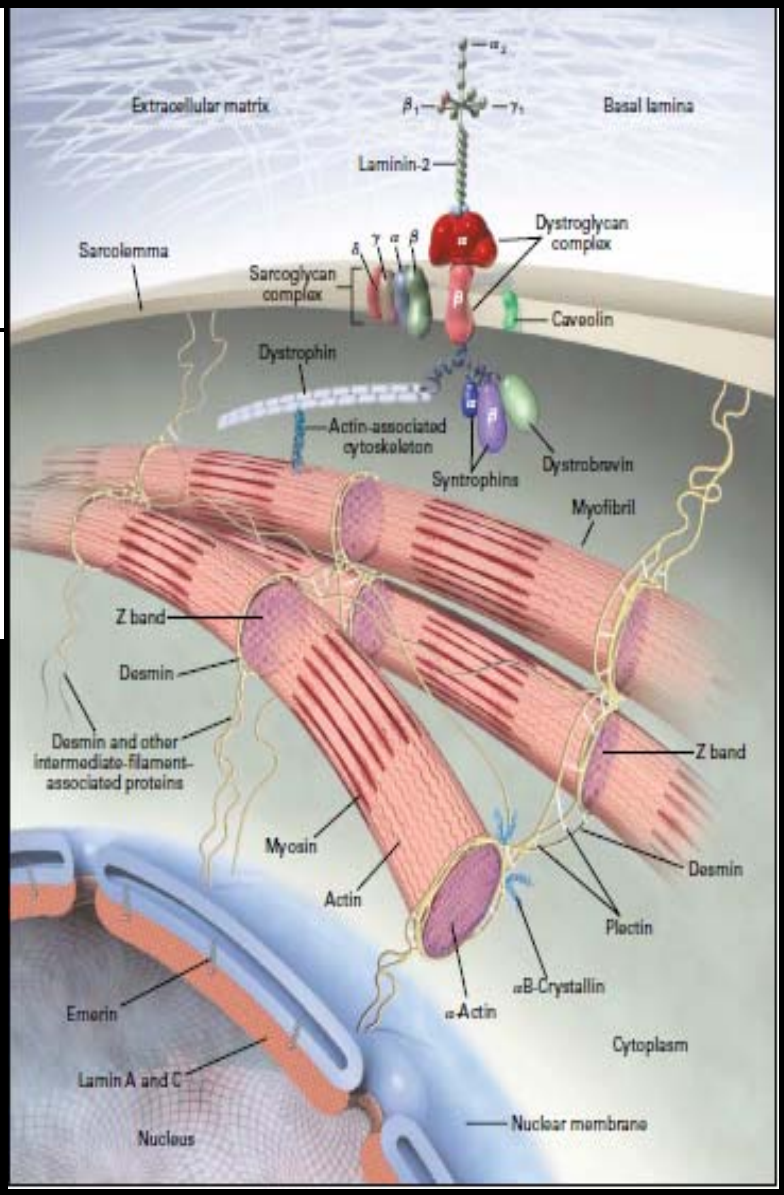
# DESMIN myopathy/cardiomyopathy

The New England Journal of Medicine

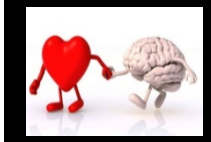
DESMIN MYOPATHY, A SKELETAL MYOPATHY WITH CARDIOMYOPATHY CAUSED BY MUTATIONS IN THE DESMIN GENE

MARINOS C. DALAKAS, M.D., KYE-YOON PARK, PH.D., CRISTINA SEMINO-MORA, M.D., PH.D., HEE SUK LEE, M.D., KUMARASWAMY SIVAKUMAR, M.D., AND LEV G. GOLDFARB, M.D.

*Conclusions* Mutations in the desmin gene affecting intermediate filaments cause a distinct myopathy that is often associated with cardiomyopathy and is termed "desmin myopathy." The mutant desmin interferes with the normal assembly of intermediate filaments, resulting in fragility of the myofibrils and severe dysfunction of skeletal and cardiac muscles.
(N Engl J Med 2000;342:770-80.)




Desminopathies
are heterogeneous group of
severe, dominantly inherited myopathies,
often accompanied by cardiomyopathy,
with syncope or sudden death
due to conduction defects.
Onset and progression are variable.



«Ενημερωμένος» νευρολόγος ζητά
καρδιολογικό follow-up!

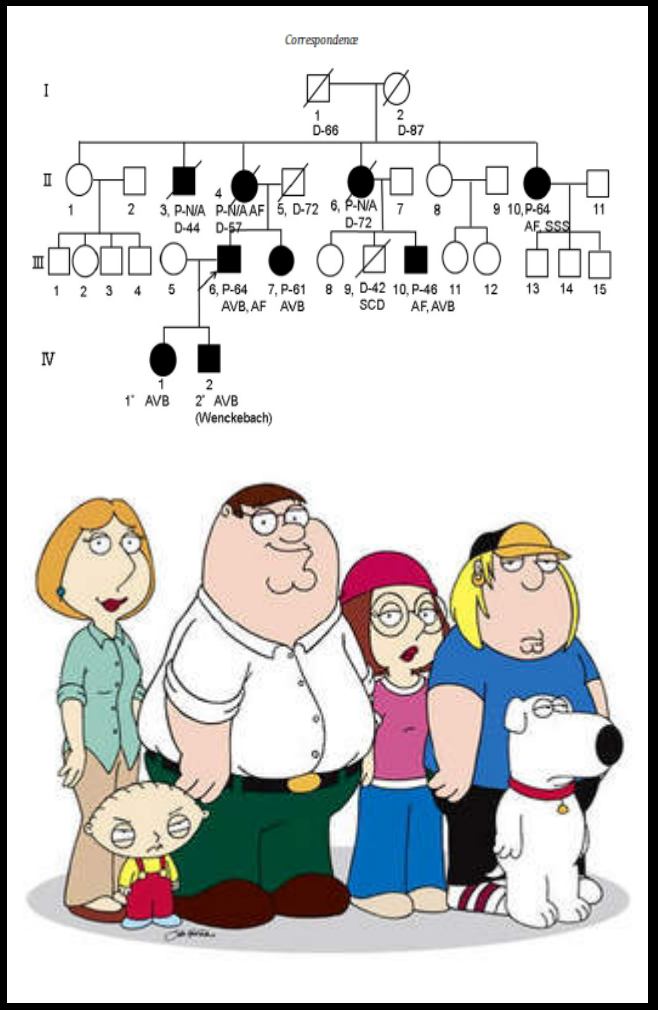
# DCM “take home message”

Ο όρος διατατική μυοκαρδιοπάθεια
περικλείει ένα φάσμα ετερογενών παθήσεων,
με διαφορετική αντιμετώπιση και ποικίλη πρόγνωση.

Πολύ συχνά, μη-καρδιακά συμπτώματα/σημεία
μπορούν να οδηγήσουν στη σωστή διάγνωση
(βλέπε laminopathies, desminopathies).
Στις μέρες μας, άνθρωποι θεραπεύονται από πολλούς
γιατρούς για πολλές παθήσεις, ενώ στην πραγματικότητα
πρόκειται για πολλές εκφάνσεις της ίδιας νόσου.

Η μελέτη του «οικογενειακού δέντρου» (pedigree)
και η συστηματική παρακολούθηση (long-term follow-up)
φαίνεται να είναι η λύση στο πρόβλημα…

Χαρίσιος Μπουντούλας. Cardiology 2011



# ΠΕΡΙΟΡΙΣΤΙΚΗ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ

RCM are defined as restrictive ventricular physiology (increased stiffness) in the presence of normal or reduced diastolic and systolic volumes.
Historically, systolic function is preserved

- Idiopathic
- Familial
- Result from various systemic disorders
  (amyloidosis, sarcoidosis, carcinoid, scleroderma, anthracycline)

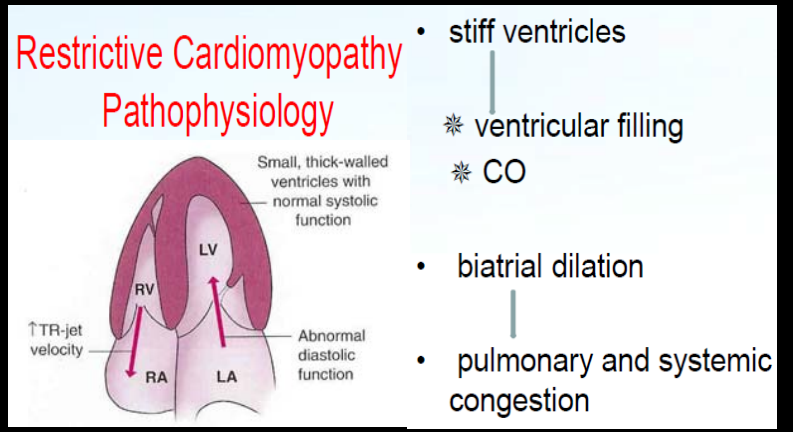


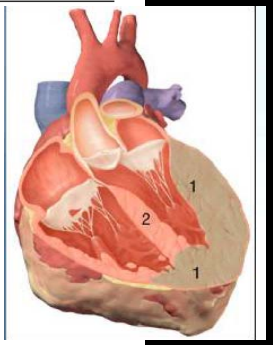

**TABLE 4.** CAUSES OF RESTRICTIVE CARDIOMYOPATHY.

**Myocardial**
Noninfiltrative disorders
- Idiopathic disease
- Familial disease
- Hypertrophy
- Scleroderma
- Diabetes mellitus
- Pseudoxanthoma elasticum

Infiltrative disorders
- Amyloidosis
- Sarcoidosis
- Gaucher's disease
- Hurler's syndrome
- Fatty infiltration

Storage disorders
- Hemochromatosis
- Fabry's disease
- Glycogen storage disease

**Endomyocardial**
Endomyocardial fibrosis
Hypereosinophilic (Löffler's) syndrome
Carcinoid syndrome
Metastatic cancer
Exposure to radiation
Toxins
Anthracycline (doxorubicin or daunorubicin)
Serotonin
Methysergide
Ergotamine
Mercurial agents
Busulfan

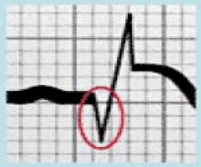



## CLINICAL PRESENTATION

- **Signs of pulmonary and systemic congestion in absence of cardiomegaly**
  - Dyspnea
  - PND, orthopnea
  - Peripheral edema
  - Palpitations
  - Fatigue, weakness, exercise intolerance
- **Thromboembolic complications** (up to 1/3 with idiopathic RCM)
- **Cardiac conduction disturbances**
  - Amyloid , sarcoid, hemochromatosis
  - AF common in IRCM & amyloidosis
- **Advanced Stage**
  - Marked elevation in CVP
    - Hepatosplenomegaly, ascites, anasarca

# Restrictive Cardiomyopathy Diagnosis

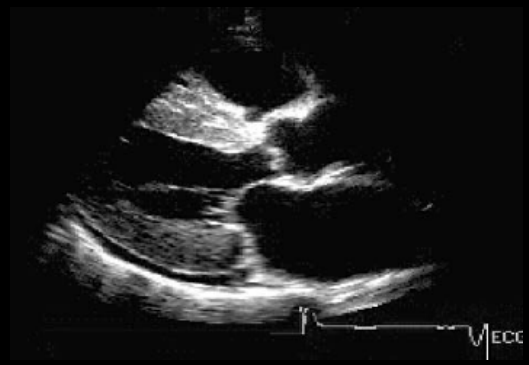
**infarction!**

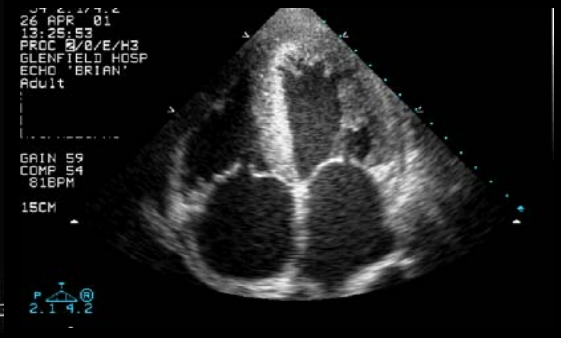


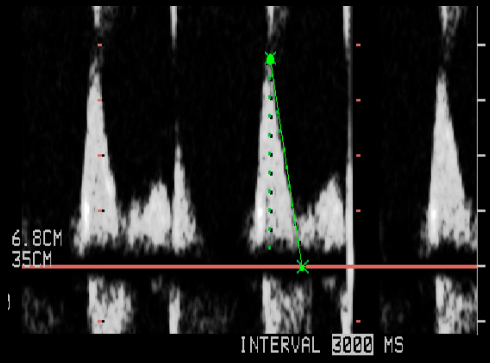
Q wave

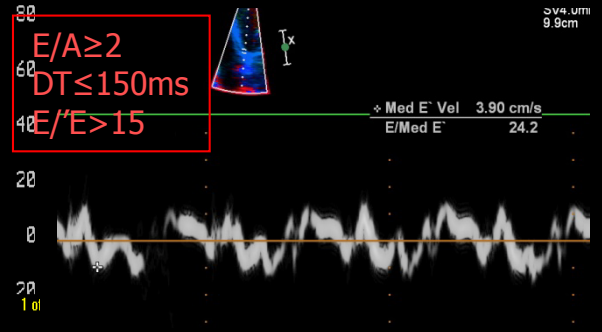
- low voltage QRS
- sinus tachycardia, atrial fibrillation, sinus bradycardia if SA node infiltrated
- complex ventricular arrhythmias : are poor prognostic sign
- Q waves : pseudo infarct from fibrosis
- BBB, AVB

*RESTRICTIVE DIASTOLIC DYSFUNCTION*






# Restrictive Cardiomyopathy : Management

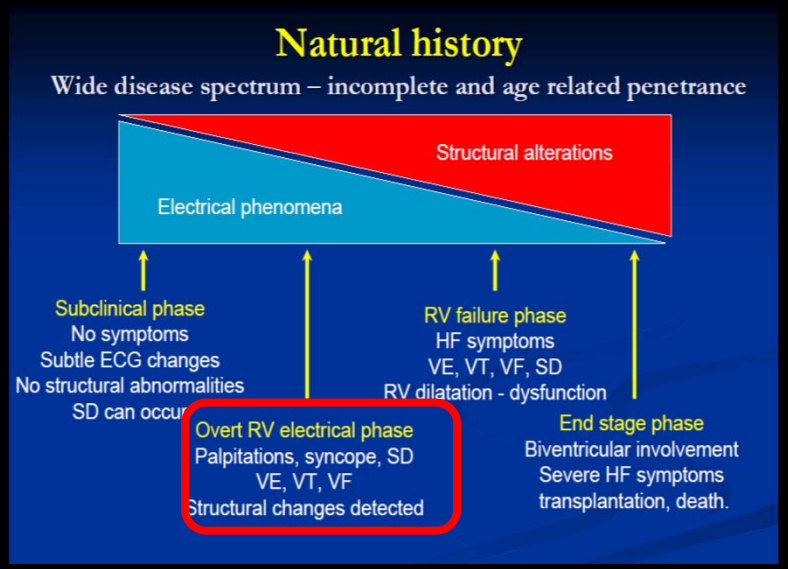
- Pharmacological support :
  - Mild diuretic therapy : prevent excessive volume depletion to prevent syncope from ❊ SV secondary to ❊ ventricular filling
  - Vasodilator : NTG, ACE inhibitors
  - No digoxin : prone to digitalis induced arrhythmias and heart block
  - No calcium channel blockers : predisposes to hypotension due to amyloidosis

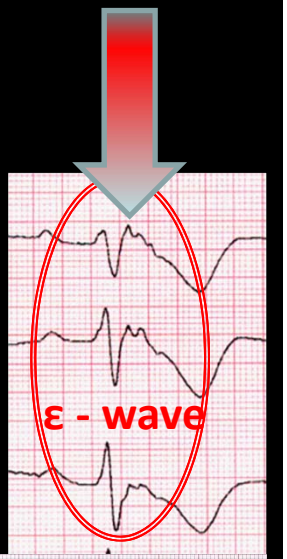
- Restrict sodium intake
- Hemochromatosis CM
  - repeated phlebotomy to reduce iron deposition in the heart
- Sarcoidosis may respond to corticosteroids
- Eosinophilic CM
  - corticosteroids and cytotoxic drugs

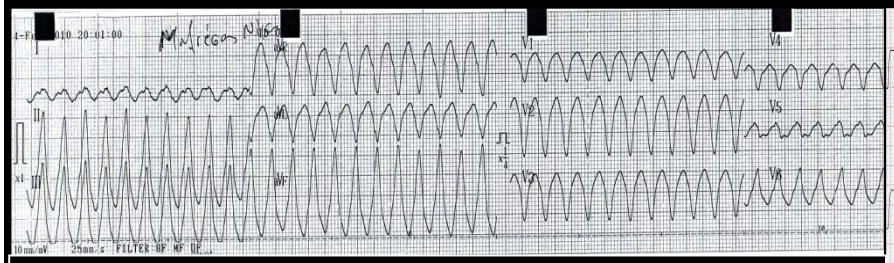
# ΑΡΡΥΘΜΙΟΓΟΝΟΣ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ ΔΕΞΙΑΣ ΚΟΙΛΙΑΣ

is a heart muscle disease,
often familial, characterized by
structural and functional
abnormalities of the right ventricle,
due to RV myocardial
atrophy and fibro-fatty replacement.

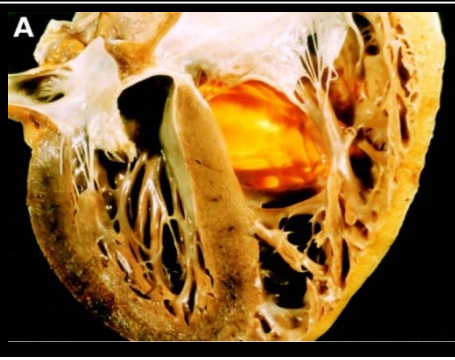
Presents clinically with ventricular
arrhythmias of RV origin.

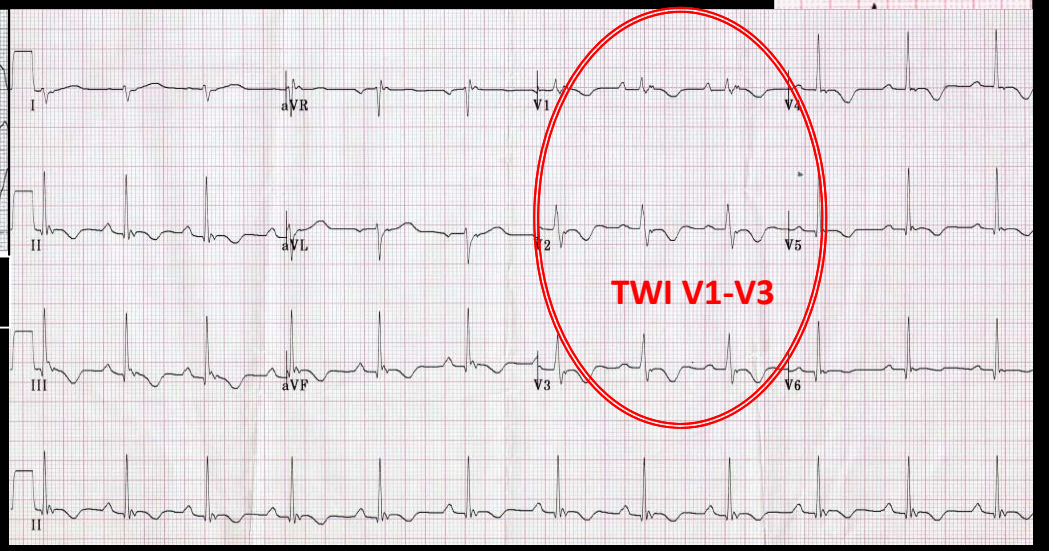


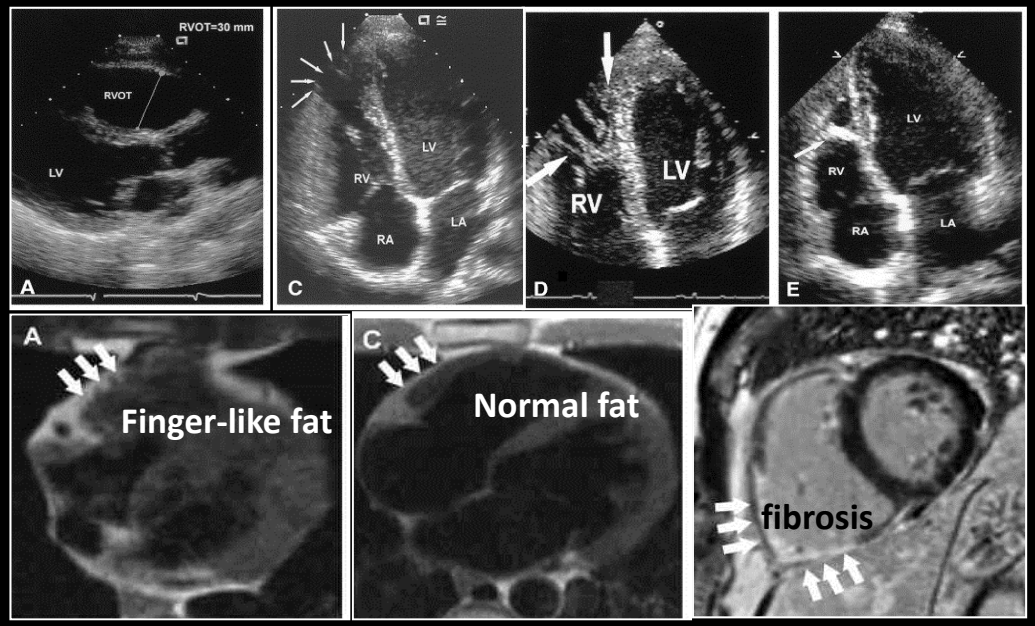



***VT morphology : LBBB – inferior axis***



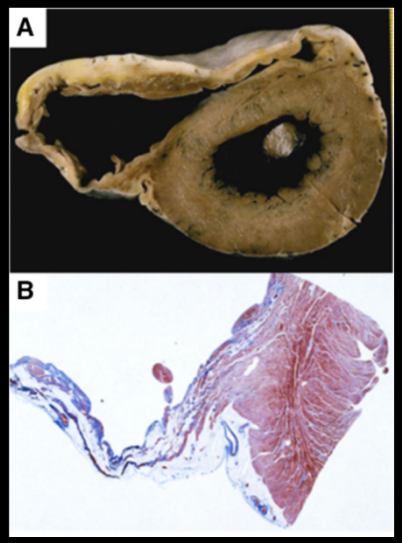



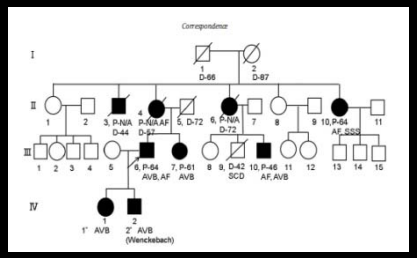



Although MRI may be regarded as the ideal technique for the evaluation of RV (morphology, function and tissue characterization in a single investigation), the use of MRI alone is not the "gold standard" for ARVC diagnosis.

Rather, the TASK FORCE CRITERIA prescribe the use of multiple diagnostic tests.

# ΔΙΑΓΝΩΣΗ ΑΡΡΥΘΜΙΟΓΟΝΟΥ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑΣ



SD family history
ECG – Holter – SA ECG
Echo – MRI
Cardiac biopsy

*CIRCULATION 2010*

**Special Report**

## Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

### Proposed Modification of the Task Force Criteria

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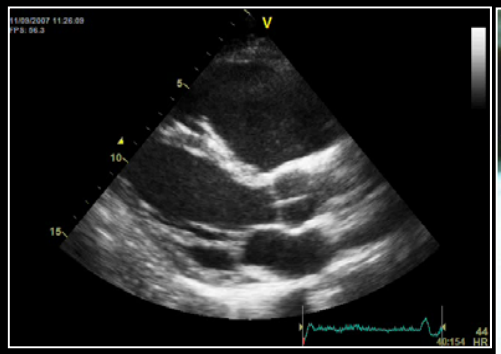
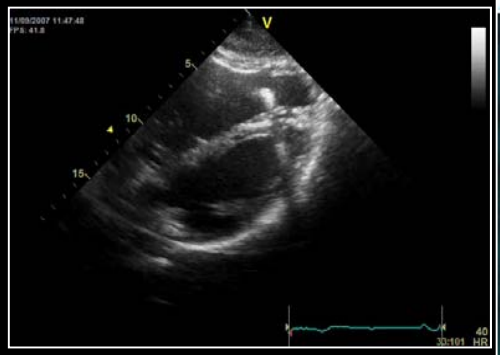
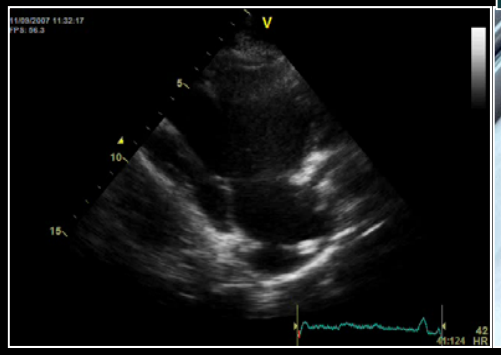
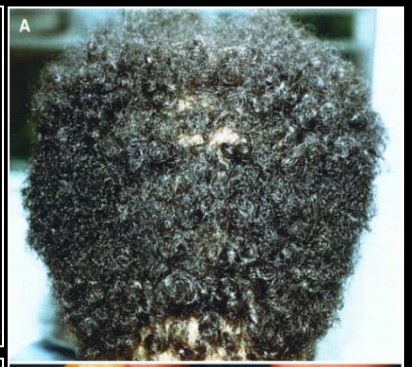
# ΑΡΡΥΘΜΙΟΓΟΝΟΣ ΜΥΟΚΑΡΔΙΟΠΑΘΕΙΑ – *νόσος NAXOS*

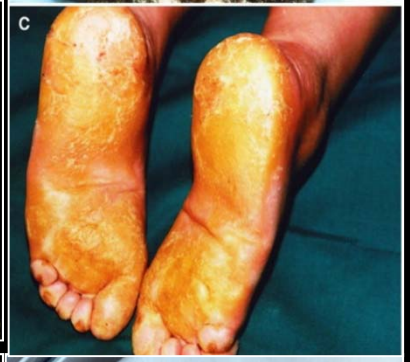
Autosomal Recessive ARVC - Plakoglobin gene

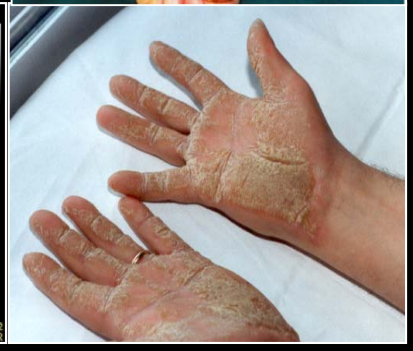








cardiomyopathy
+
wooly hair
+
palmoplantar keratoderma

*Br Heart J* 1986;**56**:321–6

## Cardiac abnormalities in familial palmoplantar keratosis

N PROTONOTARIOS, A TSATSOPOULOU, P PATSOURAKOS, D ALEXOPOULOS, P GEZERLIS, S SIMITSIS, G SCAMPARDONIS

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Gene for ARVC With Diffuse Nonepidermolytic Palmoplantar Keratoderma and Woolly Hair (Naxos Disease) Maps to 17q21

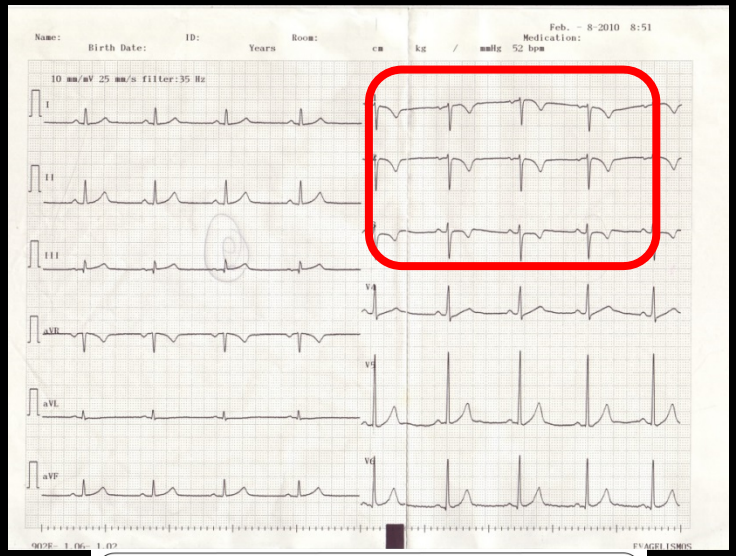
A. Coonar, N. Protonotarios, A.Tsatsopoulou;
CIRCULATION 1998

# ARVC “take home message”

Πάθηση ιδιαίτερα αρρυθμιογενής. Πρωτίστως ηλεκτρική, δευτερευόντως δομική.

ΠΡΟΣΟΧΗ

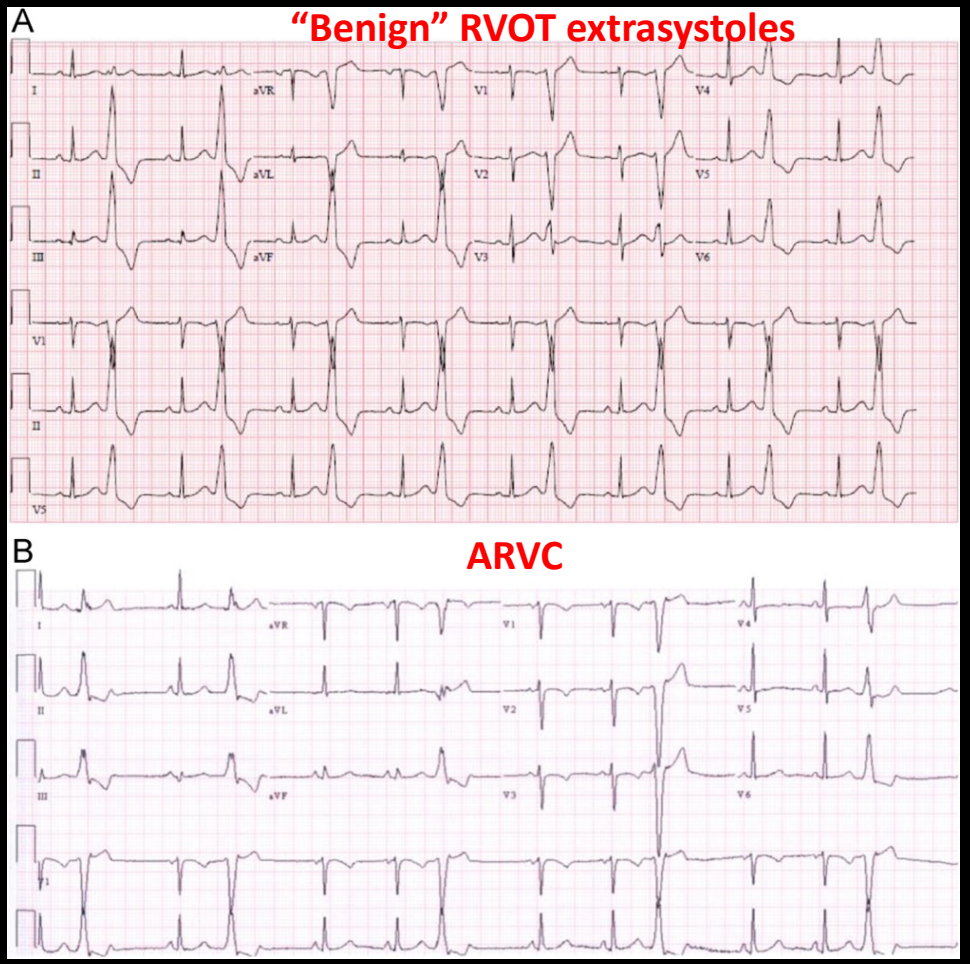
στα «αρνητικά Τ» στις V1-V3, σε ανθρώπους με αρρυθμίες ή ιστορικό αιφνιδίου θανάτου



Repolarization Abnormalities

- Repolarization abnormalities are ***early and sensitive*** markers of disease expression in ARVC/D
- T-wave inversion in V1, V2, and V3 and beyond in individuals >14 years of age who are otherwise healthy is observed in only 4% of healthy women and 1% of men. Therefore, it is ***reasonably specific*** in this population and considered a major diagnostic abnormality in ARVC/D

**Marcus FI. Prevalence of T-wave inversion beyond V1 in young normal individuals and usefulness for the diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia. Am J Cardiol. 2005; 95: 1070–1071.**



*Dealing with cardiomyopathies is like "acting as Hercule Poirot"…*

H. Boudoulas et al. Cardiology 2011

Συχνά απαιτούνται ειδικές γνώσεις και συνεργασία διαφόρων ειδικοτήτων (καρδιολόγων, γενετιστών, ψυχολόγων, νευρολόγων…)

ασθενείς

ΚΑΙ οι οικογένειές τους

Ιδανική Δομή : Ιατρεία Μυοκαρδιοπαθειών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# CARDIOMYOPATHIES "take home message"

Η κληρονομούμενη μυοκαρδιοπάθεια είναι νόσος της οικογένειας κι όχι του ατόμου...

Ο παππούς με υπερτροφική μυοκαρδιοπάθεια έχει
πολύ μικρότερο κίνδυνο αιφνιδίου θανάτου
από τον «αμέριμνο» μεν, πάσχοντα δε, εγγονό του...

Η έγκαιρη διάγνωση λοιπόν στον εγγονό, είναι εξαιρετικής σημασίας!

Γιατί....

Άμα γλυτώσει το παιδί,
ΥΠΑΡΧΕΙ ΕΛΠΙΔΑ...