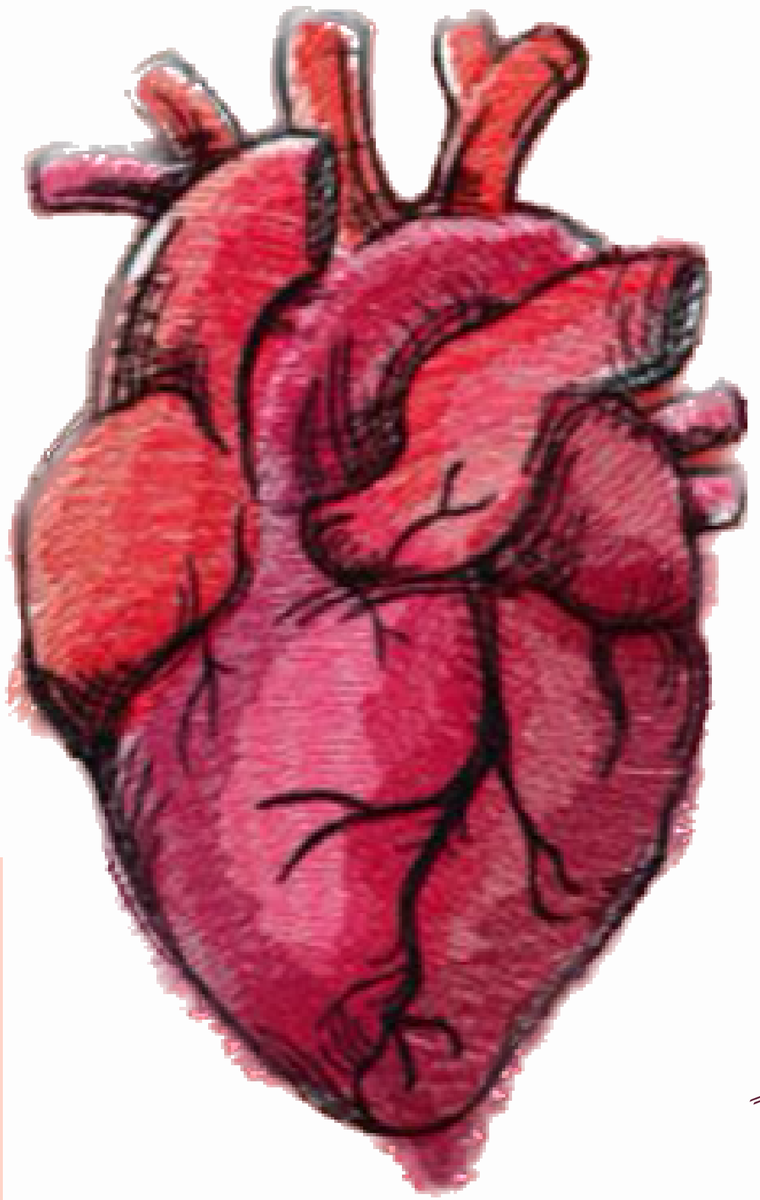


CARDIOVASCULAR SYSTEM

SUBJECT : Pathology

LEC NO. : 5

DONE BY :Hamza alsyouri

وَقُل رَّبِّ زِدْنِي عِلْمًا

SCAN ME!

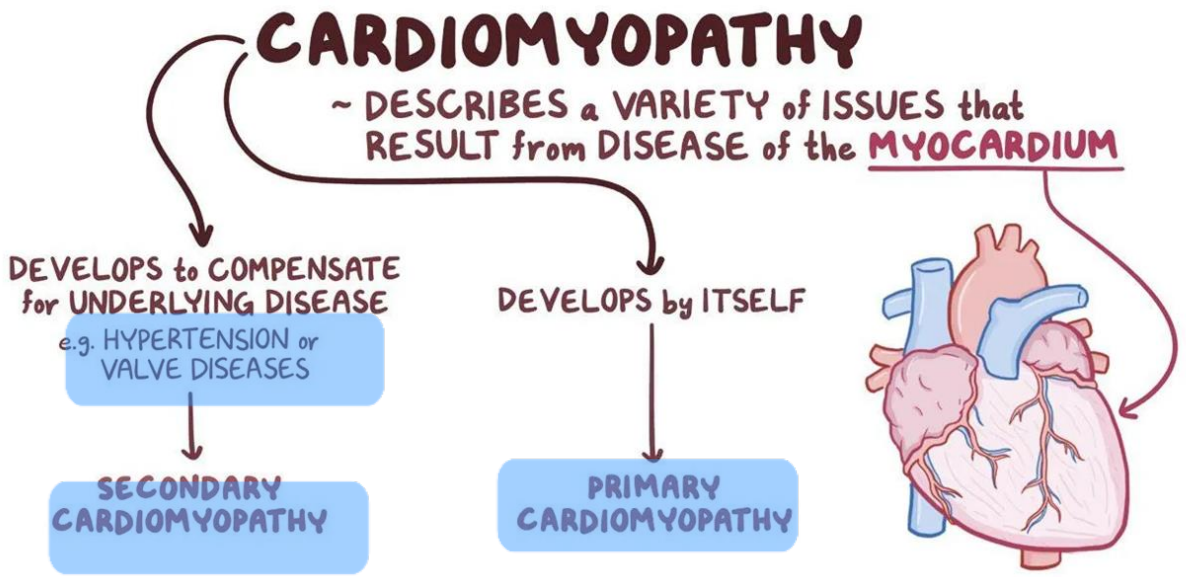
Cardiomyopathy and pericardium

DR. DUA' ABUQUTEISH

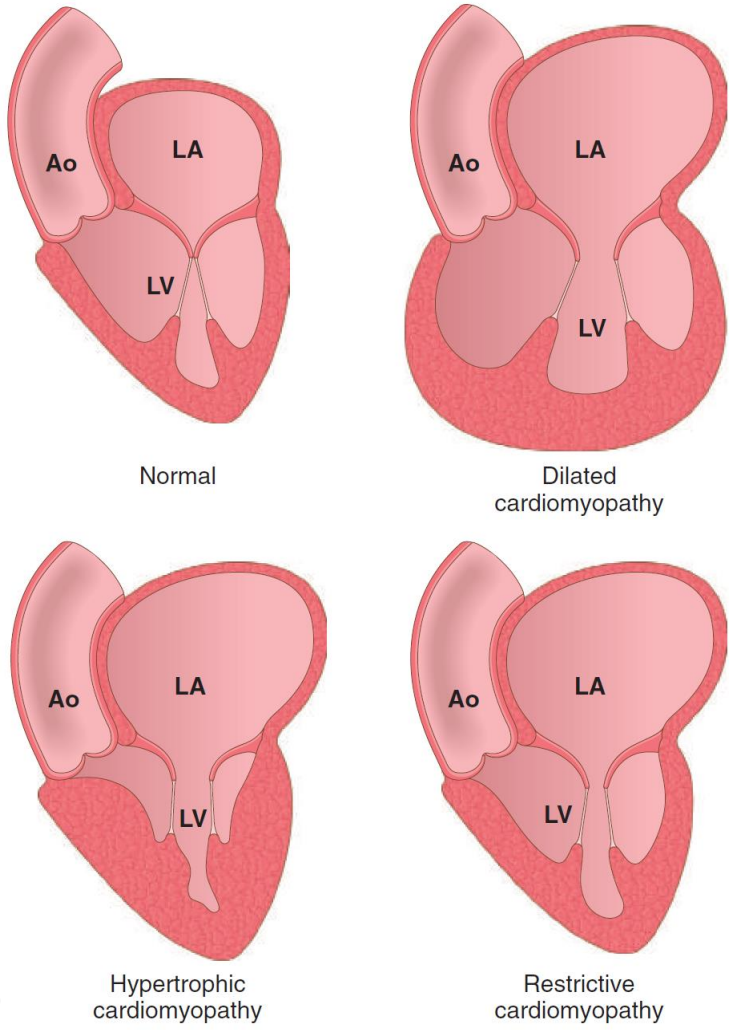
Cardiomyopathy

- **Cardio-myo-pathies** describes cardiac muscle diseases resulting in myocardial dysfunction

يعني فكرة ال secondery ما cardiomyopathy في مشكلة بال muscle نفسها المشكلة جاي من برا ال muscle بس مش هاي محاضرتنا اليوم



اليوم راح نحكي عن ال primary اللي cardiomyopathy بتعني انه المشكله بعضلة القلب نفسها يعني بغياب ال hypertension وغيره لانه المشكله بعضلة القلب



مهم

Fig. 11.23 The three major forms of cardiomyopathy. Dilated cardiomyopathy leads primarily to systolic dysfunction, whereas restrictive and hypertrophic cardiomyopathies result in diastolic dysfunction. Note the changes in atrial and/or ventricular dilation and in ventricular wall thickness. *Ao,* Aorta; *LA,* left atrium; *LV,* left ventricle.

All the 4 champers dilated

Systoic dysfunction

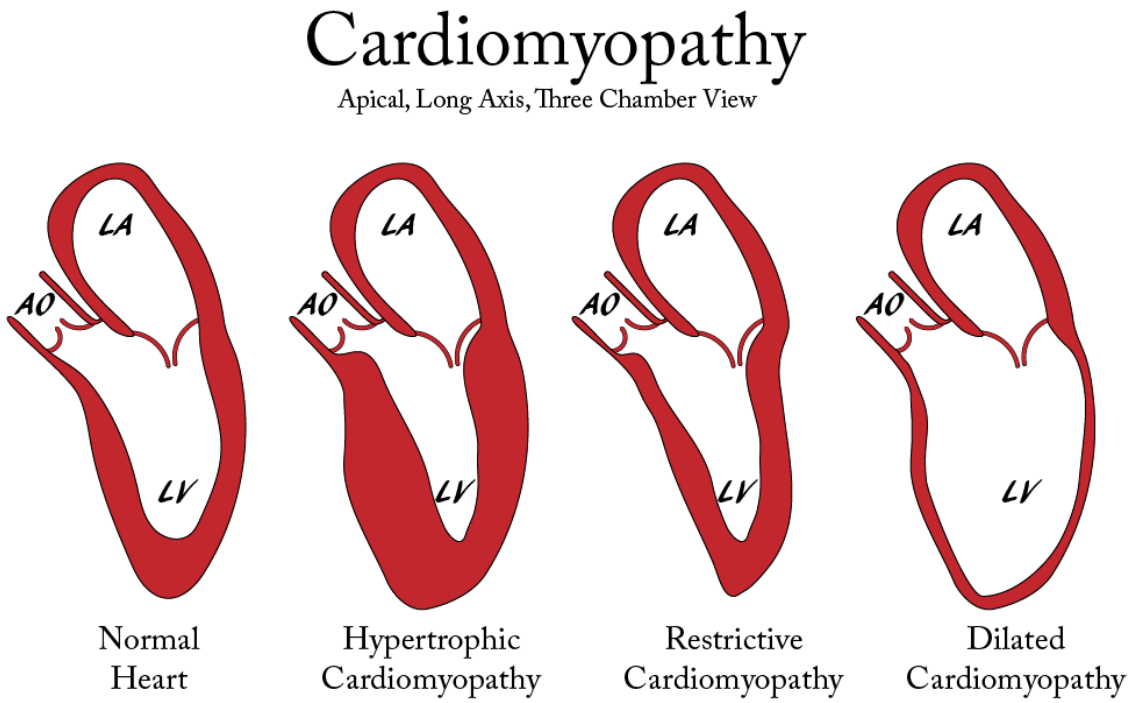
هون ما في عندي اي مساحة بالLV ولانو عندي pressure كثير عالي بالLV فال LA راح يصيرلها dilated لانها بضخ ل LV فيه كثير عالي بالpressure

Diastolic dysfunction

بال restrictive بكون في كمان اشي زي الfibrosis فبطلو بال muscle و compliant و بطلو elastic تبعهم بصير الcontraction فكمان ال pressure بزيد effected فيهم فالLA بتصير dilated

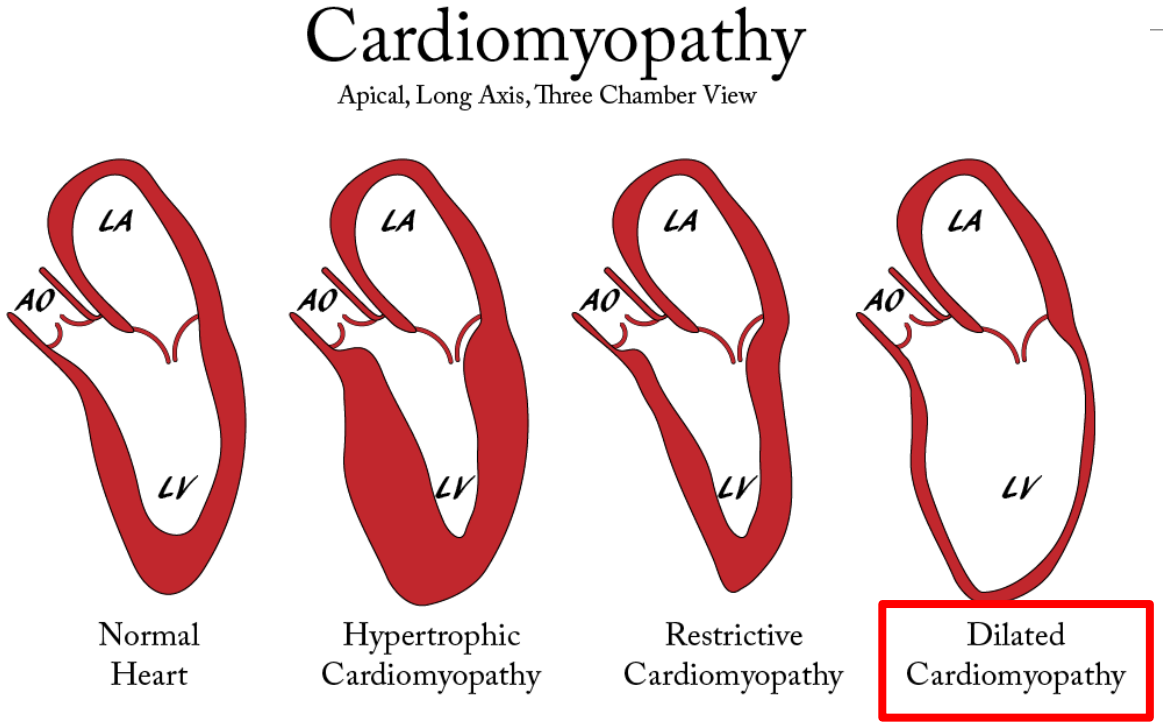
Diastolic dysfunction

Cardiomyopathy



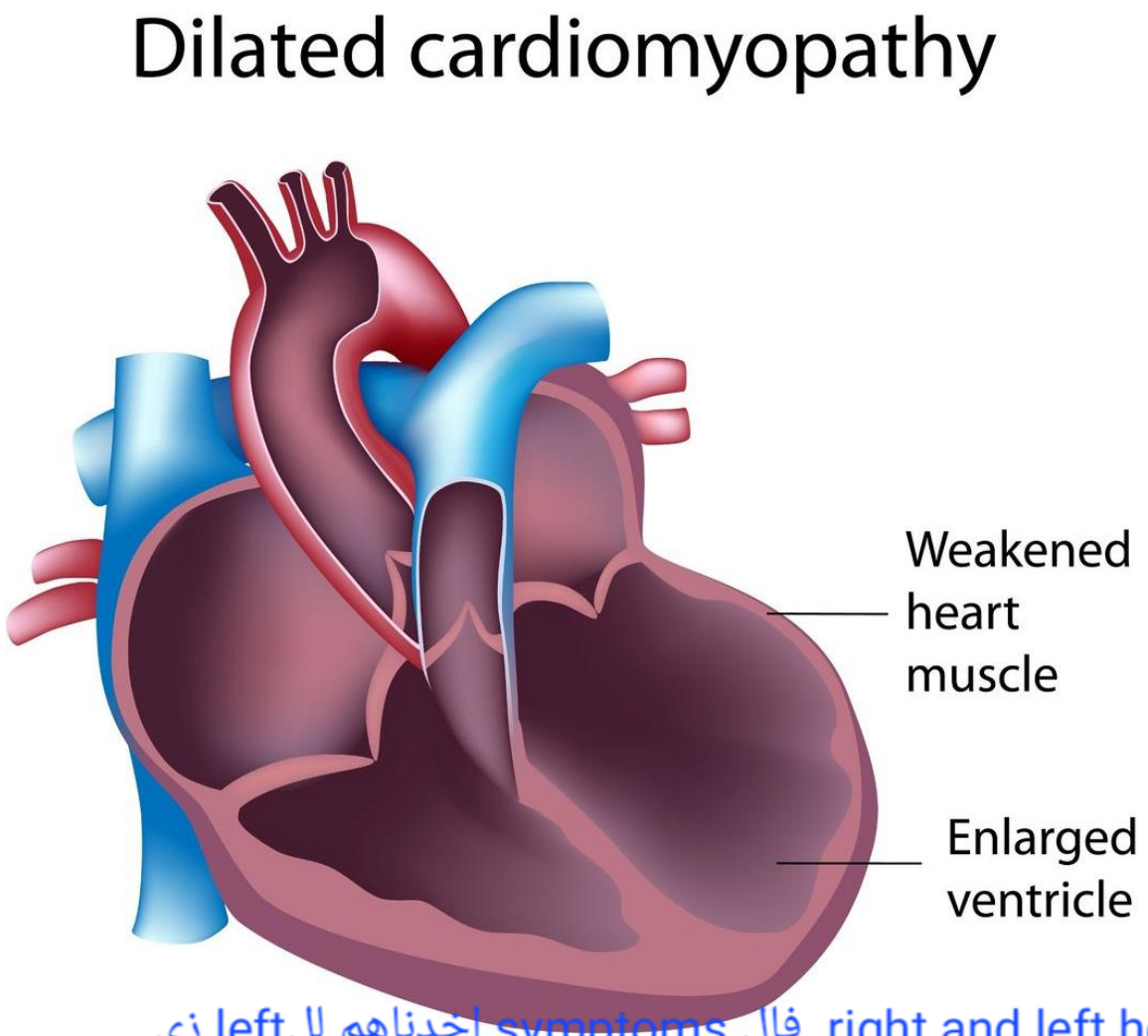
1. Dilated Cardiomyopathy (Most common; approx. 90%)
2. Hypertrophic Cardiomyopathy
3. Restrictive Cardiomyopathy

Cardiomyopathy



1. **Dilated Cardiomyopathy (Most common; approx. 90%)**
2. **Hypertrophic Cardiomyopathy**
3. **Restrictive Cardiomyopathy**

Dilated Cardiomyopathy



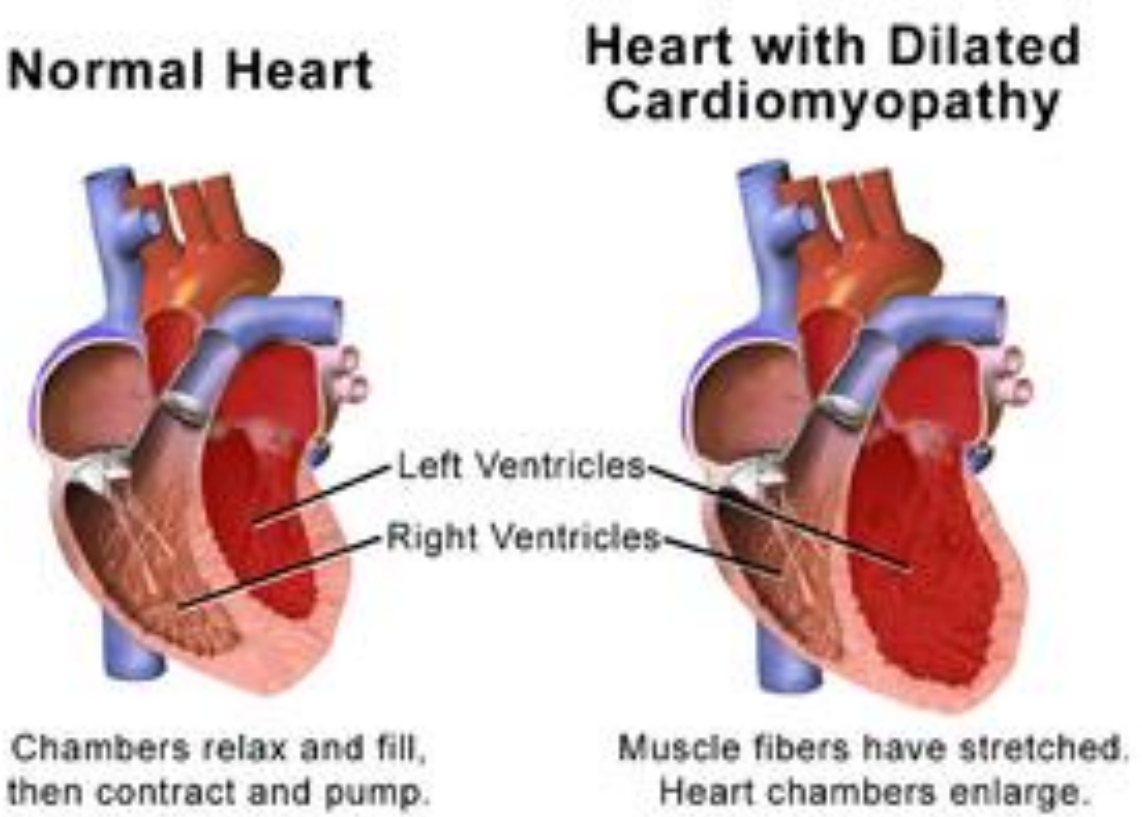
- **Progressive cardiac dilation,** usually with concurrent hypertrophy

Results in systolic dysfunction (ventricles cannot pump), leading to bi-ventricular CHF

- All chambers are dilated

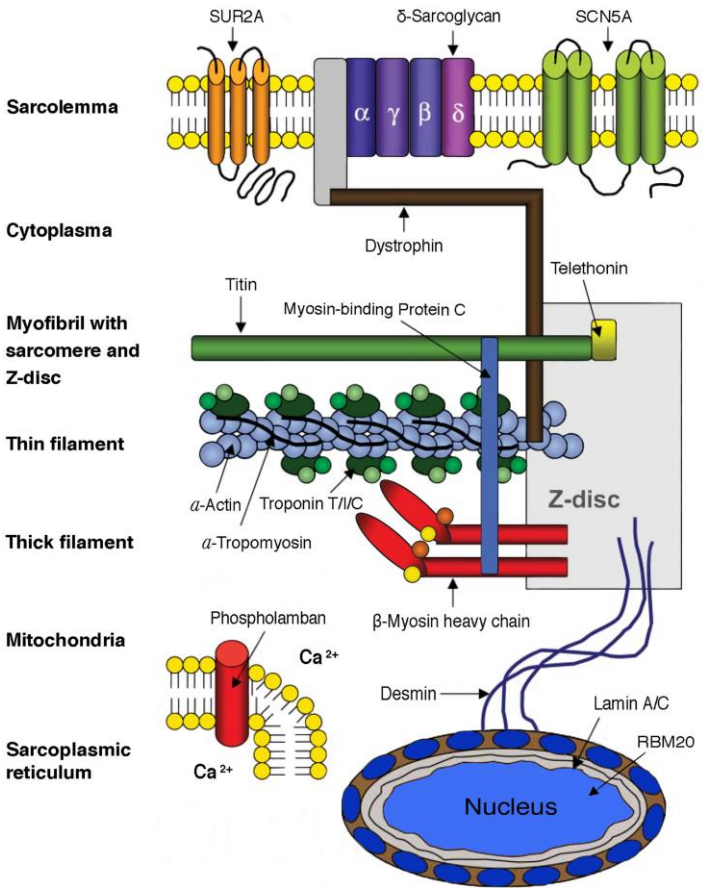
فهذا بأدي الى bi ventricular CHF ال right and left heart failure فال symptoms احدناهم لل left زي
و ال pulmonary edema and cyanosis ل symptoms right زي splenomegaly فهون راح نشوف كل هاي ال
symptoms هون

Dilated Cardiomyopathy DCM



هلا بدنا نحكي عن ال causes
كل ال causes بأثرو على الmuscle نفسها فال
muscle بتبطل contractile
فهلا ليش بصير عندي dilation ؟ المشكلة عندي هون
بال systolic فهلا ال heart لما بدها تضخ blood
للجسمduring systolic ال muscles ما راح
يشتغلو كويس لانه فيهم مشكله بنفسهم هم
يا مشكله genetic يا مشكلة toxic زي ما راح نحكي
هلا
هلا اللي بصير انو ال ejaction fraction هو اللي
بتأثر
فهلا ايش هو الejaction fraction عنا مثلا بال
diastolic لما ال heart يعبي blood بعبي على فرض
100 مل فهاد بنسميه undiastolic volume فقديش
القلب بضخ منه،بضخ تقريبا 50-70%
اذا normal ال ejaction fraction بال heart
50-70% معنا هاد الحكي انو تقريبا 70 مل راح يطلع
وراح يضل عندي 30
فال ejaction fraction عبارة عن قديش طلع من ال
heart (70) على 100
فال ejaction fraction بهدول المرضى بقل لانه

ما عندي systolic contraction كويس
فبدل ما يضخ 50-70% بضخ مثلا 40%
فبصير يطلع 40 مل
فال heart عشان to compensate بتبلش تصير
اكثر dilated
عشان بدل ما تعبي 100 تعبي 200 مثلا
فأذا عبت 200 فلو كانت ال ejaction fraction
40% بتقدر اطلع 80 من ال blood هيك
فهاد هو السبب انه ال heart بتوسع وبصير dilated



Dilated Cardiomyopathy Causes

Most common: no cause (idiopathic)

1. Genetic causes.

- Hereditary in 20% to 50% of cases

- Over 40 genes are known to be mutated

- Autosomal dominant (AD) inheritance: mutations in encoding cytoskeletal proteins, or proteins that link the sarcomere to the cytoskeleton (e.g., α-cardiac actin).

- X-linked: dystrophin gene mutations (Duchenne muscular dystrophy: DMD)

Dilated Cardiomyopathy DCM Causes (cont.)

2. Infections (myocarditis):

- Coxsackievirus B and other enteroviruses, also **Chagas disease** (parasitic)

- Myocarditis; most common cause is coxsackievirus; acutely, it can cause death; in chronic cases, it can cause dilated cardiomyopathy

3. Alcohol or other toxic exposure:

- Alcohol and its metabolites (especially acetaldehyde) have a direct toxic effect on myocardium

4. Medications: Doxorubicin (Adriamycin) and cocaine

5. Thiamine B1 deficiency (Wet Beri Beri)

في نوعين من الBeri Beri اللي هم dry و wet
الwet هو اللي بأدي الى dilated cardiomyopathy

Dilated Cardiomyopathy DCM Causes (cont.)

مع الحمل

5. Peripartum cardiomyopathy:

- Occurs late in gestation or several weeks to months postpartum.
- Pathogenesis is multifactorial
- Approximately half of these patient spontaneously recover normal function

6. Iron overload in the heart (hemochromatosis)

ممكن تعملي شغلتين heart بال iron overload اذا صار عندي hematochromatosis ال
زي اللي بنحكي عنه هلا يا بتعمل اشي راح ناخده dilated cardiomyopathy يا بتعمل
restrictive cardiomyopathy بثالث نوع اللي هو ال

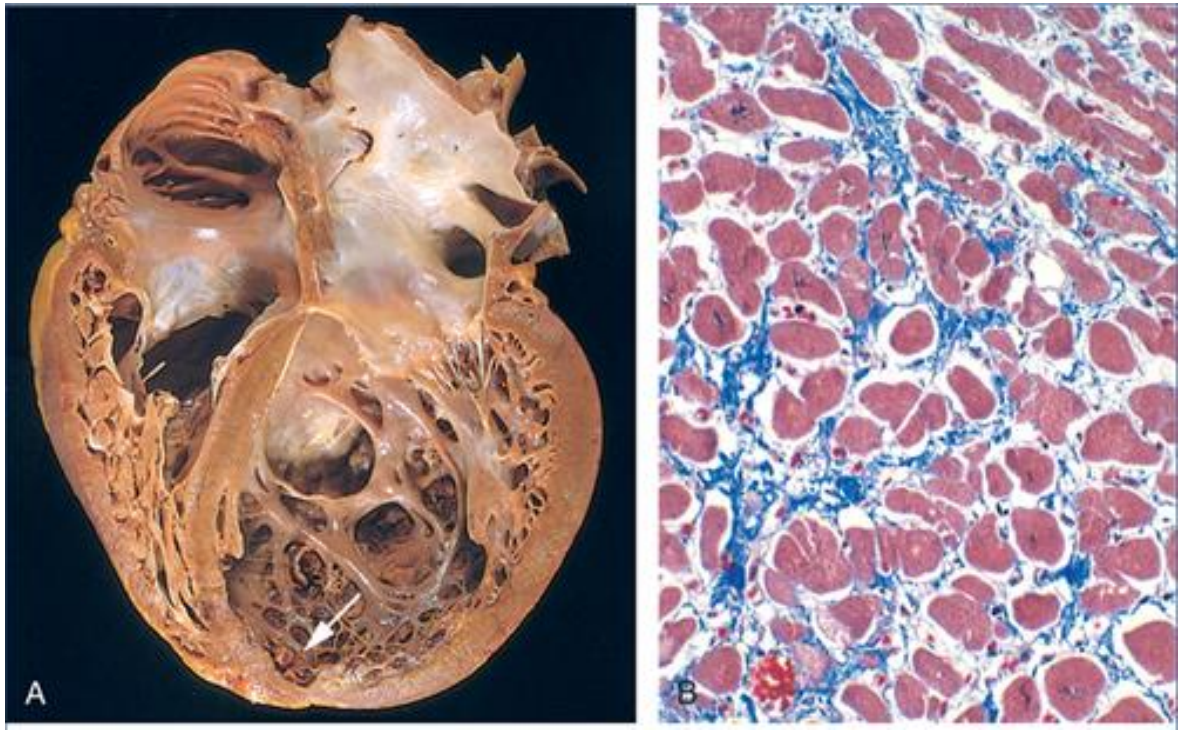
Dilated Cardiomyopathy DCM Morphology

- The heart is enlarged (up to 2-3 times the normal weight) , **with dilation of all chambers**
- **Mural thrombi** are often present and may be a source of thromboemboli.

بتصير لانه معظم الدم اللي بيدخل ما بيطلع فبيصير stasis

Histologic abnormalities are nonspecific:

- Myocytes exhibit **hypertrophy** with enlarged nuclei.
- Variable interstitial fibrosis



Dilated Cardiomyopathy Morphology

A. **Four-chamber dilatation & hypertrophy**, with a small mural thrombus (arrow) in the apex of the LV. There was no coronary artery disease.

B. **Typical myocyte hypertrophy & interstitial fibrosis** (Masson trichrome stain collagen blue).

Dilated Cardiomyopathy DCM
Clinical features

- DCM occurs at any age, but most commonly between 20-50 years.
- Patients present with slowly progressive bi-CHF, including dyspnea, easy fatigability.
- The fundamental defect in DCM is ineffective contraction.
- In end-stage DCM :the **cardiac ejection fraction** typically is **less than 25%** (normal 50% to 65%)

مهم انه نضل متذكرين انه systolic dysfunction

Dilated Cardiomyopathy DCM
Clinical features

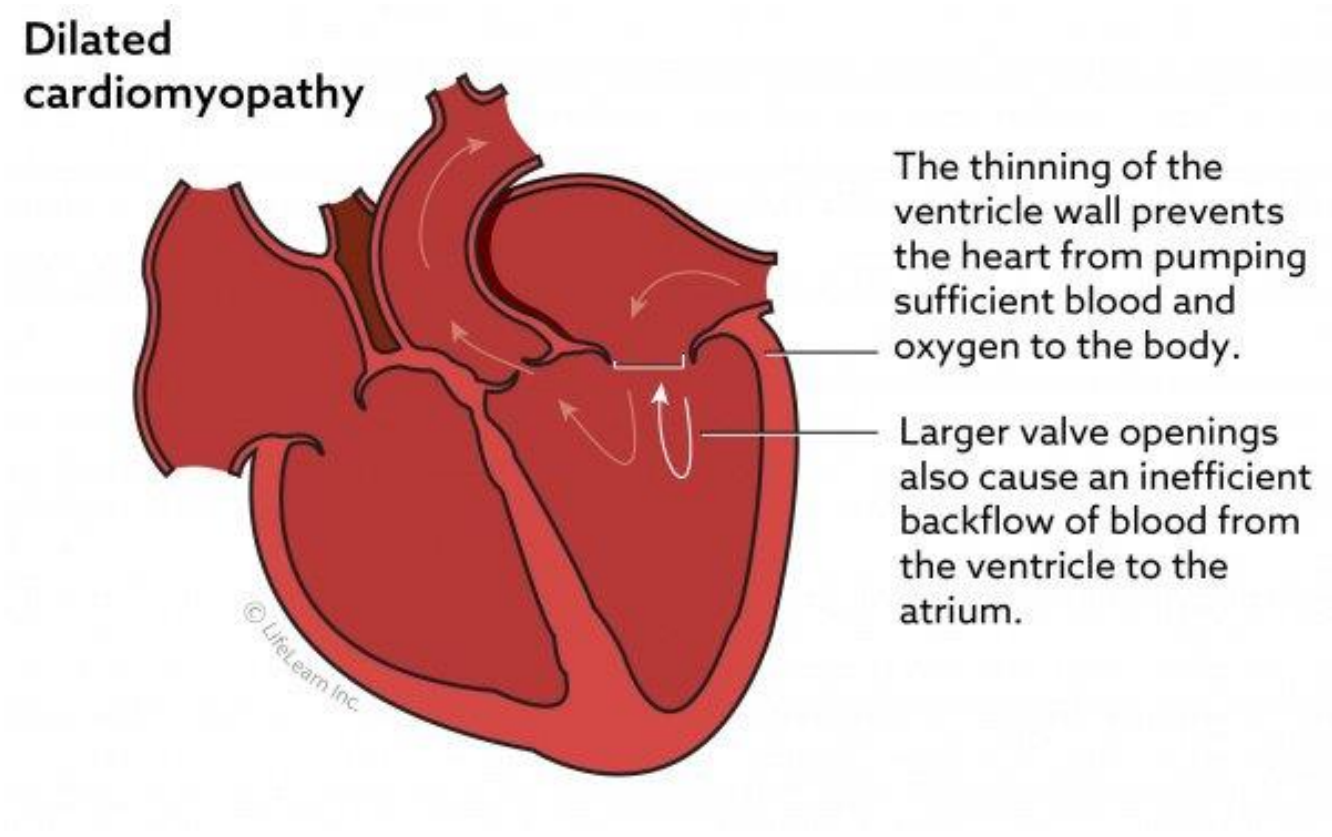
Complications:

- Mitral and tricuspid regurgitation لانهم بتوسعو مع توسع ال heart فال cusps بيعدو عن بعض
- Arrhythmia (heart's conduction system is stretched up)
  لانه مليان صار عندي fibrosis

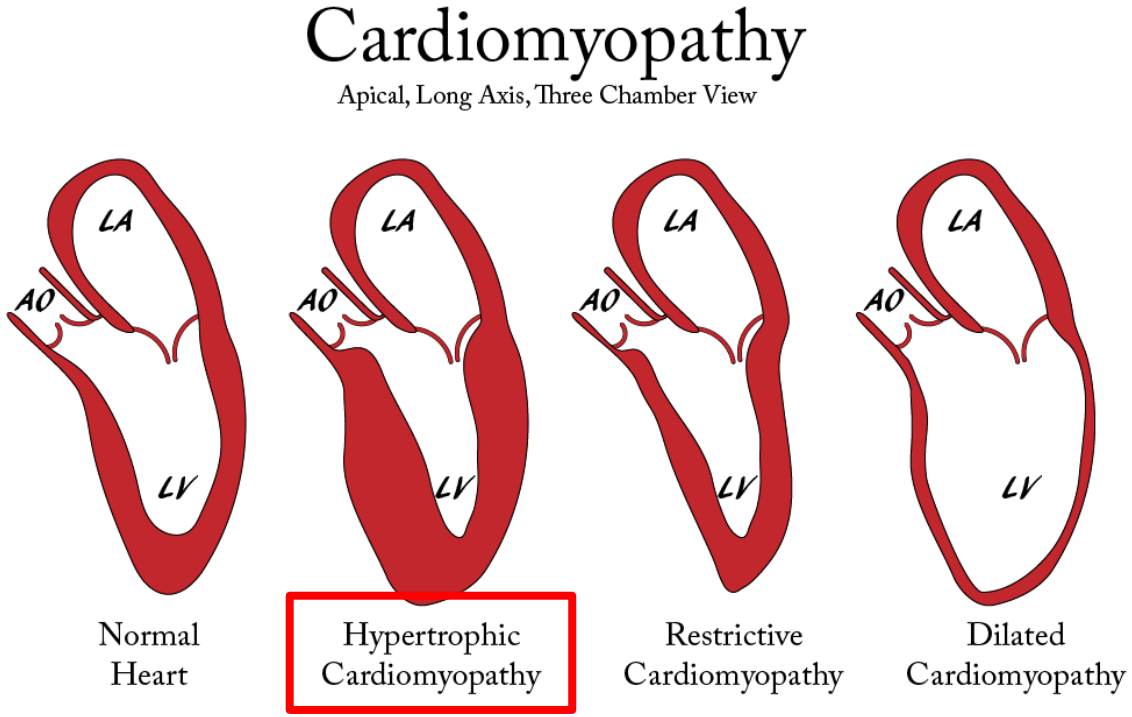
- Half of the patients die within 2 years, and only 25% survive longer than 5 years
- Death usually is due to progressive cardiac failure or arrhythmia.
- **Cardiac transplantation is the only definitive treatment.**

Dilated Cardiomyopathy

هاي الصورة بس عشان تشوف ال mitral وال tricuspid regurgitation



Cardiomyopathy



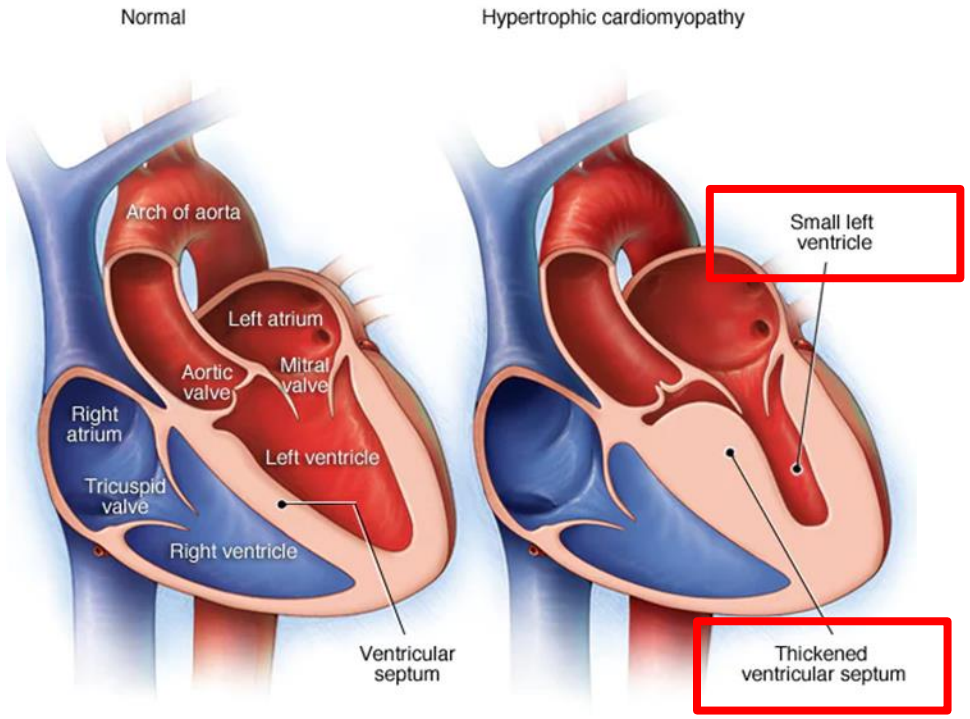
1. Dilated Cardiomyopathy (Most common; approx. 90%)
2. Hypertrophic Cardiomyopathy
3. Restrictive Cardiomyopathy

Hypertrophic Cardiomyopathy(HCM)
Also called Hypertrophic obstructive cardiomyopathy (HOCM)

HCM is characterized by:

1. **Massive LV & IVS myocardial hypertrophy**
2. **Defective diastolic filling**
3. **Ventricular outflow obstruction**

- ❑ Heart is thick-walled, heavy, and **hypercontractile**
- ❑ Systolic function usually is preserved , <u>but the myocardium does not relax</u>



ال defective diastolic filling : هلا انا عندي هون
diastolic dysfunction فلو تتطلع على الصورة
بتشوف انو ال champer صارت كثير صغيرة من كثر
ال hyperatrophy مقارنة بال heart normal فلما
يجي يعبي دم بال diastole اشي طبيعي انو
heart musles they stretch outال عشان تعبي
ventricle ال (HOCM) ولكن في حالة ال blood
ejection fraction صغير فممكن يعبي 50 وال
كويسة تساوي 50 ولكن راح يطلع فقط 25 % من الدم
لانه المشكله بقديش عبى blood

Ventricular outflow obstruction :ال
obstruction بتصير
لانه ال blood بيمشي بمنطقة ديقة بشكل سريع وال
mitral valve كثير قليل فممكن ال pressure
obstruction فبيتصير suptum تندفش وتلزق بال
aorta اللي بيدخل لل blood لل

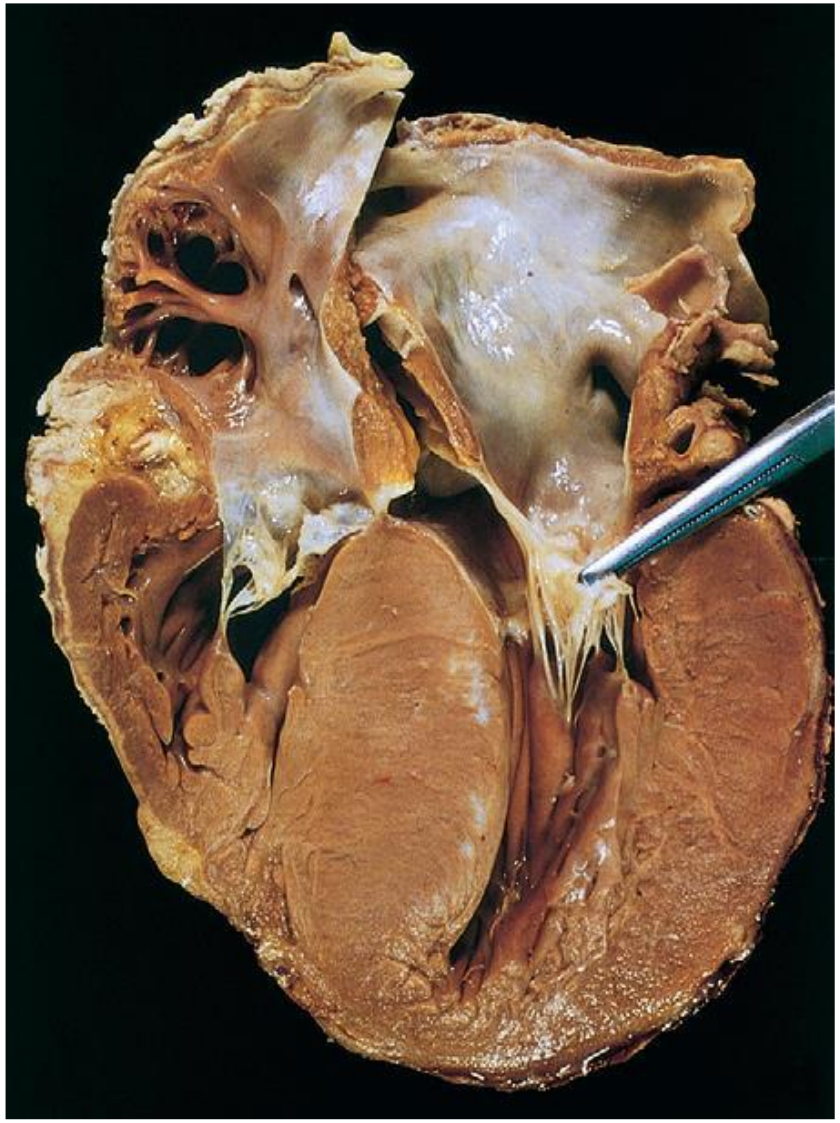
Hypertrophic Cardiomyopathy(HCM) Causes

Most common cause is hereditary and is due to autosomal dominant mutations in sarcomere proteins

Disorder of sarcomeric proteins:

- **β-myosin heavy chain is most frequently affected**
- **Myosin-binding protein C and troponin T.**

Mutations in these three genes account for 70% to 80% of all cases of HCM.



Hypertrophic Cardiomyopathy(HCM) Morphology

- Massive myocardial hypertrophy without ventricular dilation.
- Disproportionate thickening of the ventricular **septum** relative to the left ventricle free wall (**Asymmetric septal hypertrophy**)
- On longitudinal sectioning, the ventricular cavity loses its usual round-to-ovoid shape and is compressed into a “banana-like” configuration.

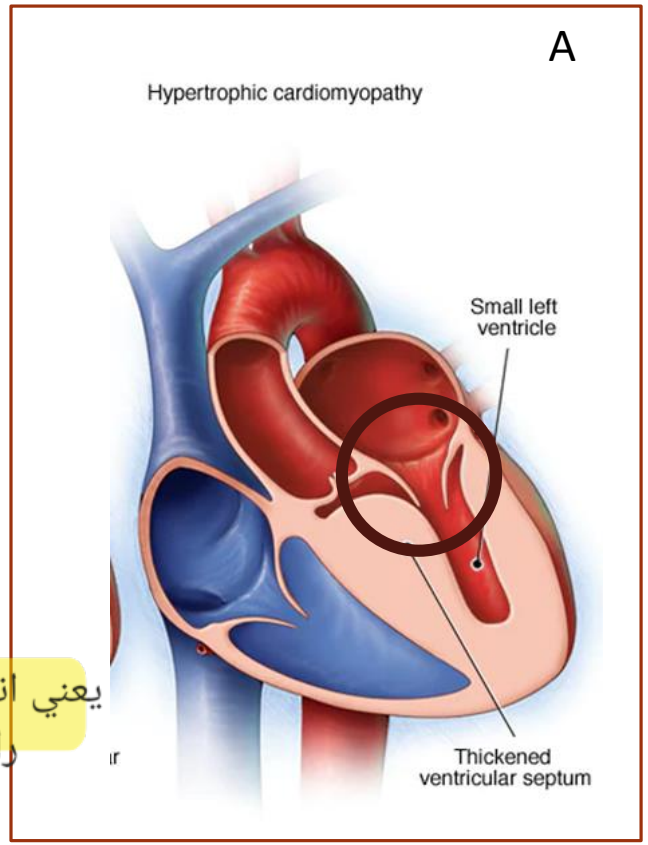
Hypertrophic Cardiomyopathy(HCM) Morphology

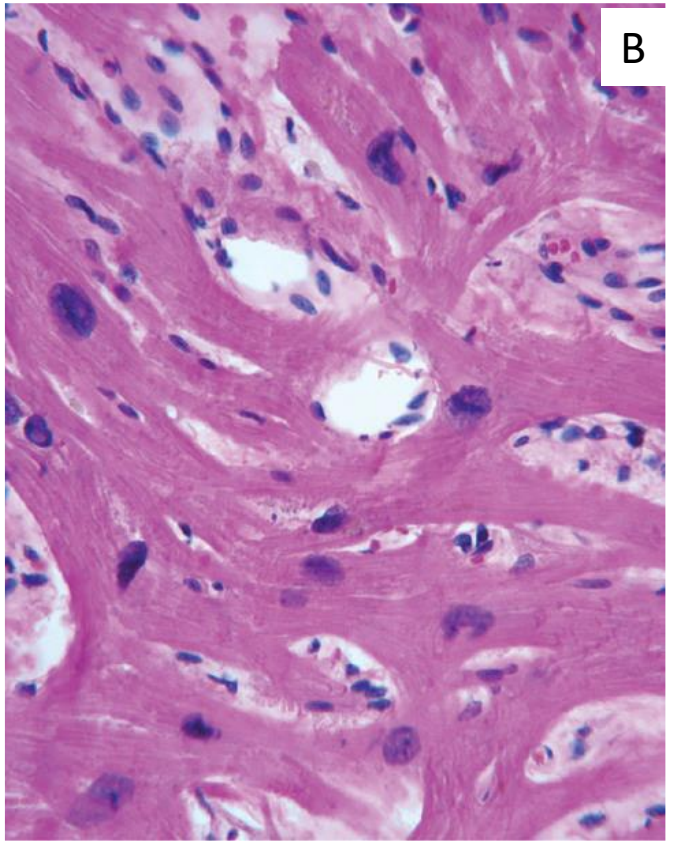
A. Contact of the anterior mitral leaflet with the septum during ventricular systole, correlate with functional **left ventricular outflow tract obstruction.**
Venturi effect

B. Histologic features:

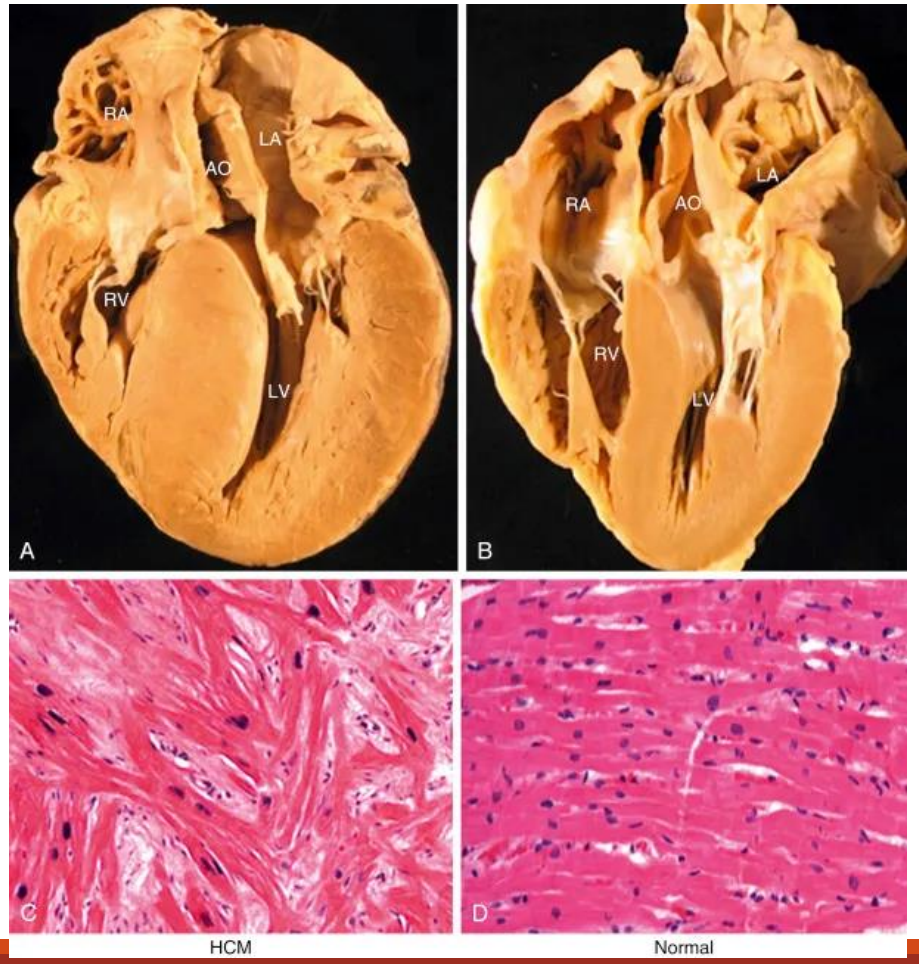
- **Marked myocyte hypertrophy**
- **Haphazard myocyte disarray**
- **Interstitial fibrosis .**

يعني انه كل عضلة رايحة باتجاه





Hypertrophic Cardiomyopathy(HCM) Morphology



Hypertrophic Cardiomyopathy(HCM) Clinical features

- Can present at any age but it typically manifests during the post pubertal growth spurt.
- Massively hypertrophied left ventricle that paradoxically provides a markedly reduced stroke volume due to impaired diastolic filling and overall smaller chamber size.
- Patients can present with syncope during exercise
- Reduced cardiac output exertional dyspnea, with a harsh systolic ejection murmur.

Hypertrophic Cardiomyopathy(HCM) Clinical features

- Myocardial ischemia, even in the absence of concomitant CAD, due to combination of massive hypertrophy, high left ventricular pressures, and compromised intramural arteries.

Hypertrophic Cardiomyopathy(HCM) Clinical features

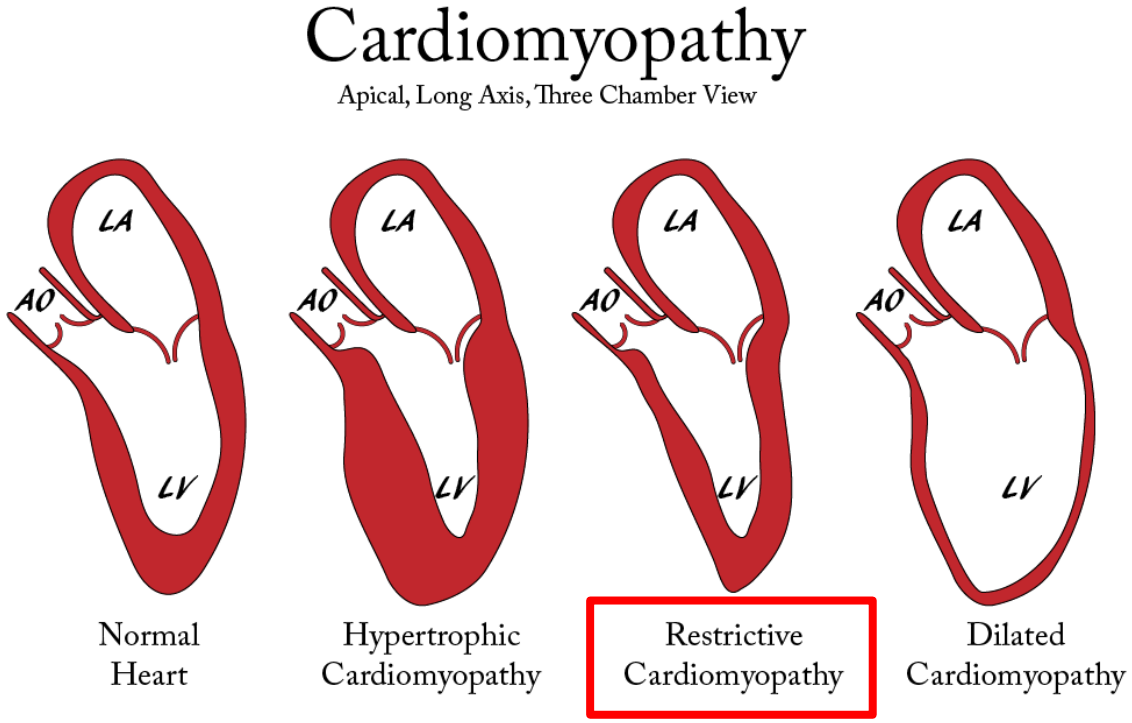
Major clinical problems:

- Arrythmias: such as atrial and ventricular fibrillations
- CHF
- Sudden death: **HCM is an important cause of sudden cardiac death,** one third of sudden cardiac death in athletes under the age of 35
- Most patients are improved by therapy that promotes ventricular relaxation
- Partial surgical excision of septal muscle also can relieve the outflow tract obstruction.

لما يجو يشخصو المرضى في عنا ultrasound to the heart اسمه eco cardiogram وهو من اهم ال diagnostic modalities لكل هدول ال cardiomyopathies

بنعالجهم عن طريق انه بنحاول نساعد ال heart to relax during diastole فممكن نعطيهم ca+2 or B blockers و بنبتعد تماما عن الركض وعن اي excertion of demand وكمان في surgical excision وبشيلو جزء من ال septal muscle

Cardiomyopathy



1. **Dilated Cardiomyopathy (Most common; approx. 90%)**
2. **Hypertrophic Cardiomyopathy**
3. **Restrictive Cardiomyopathy**

Restrictive Cardiomyopathy

- **Primary decrease in ventricular compliance**, resulting in impaired ventricular filling during diastole

- The contractile (systolic) function of the left ventricle usually is unaffected.

Restrictive Cardiomyopathy Causes

Idiopathic or associated with systemic diseases:

Systemic diseases:

مهم تعرفو ال causes

- ❑ Amyloidosis
- ❑ Sarcoidosis
- ❑ Radiation fibrosis
- ❑ Hemochromatosis

LASHER

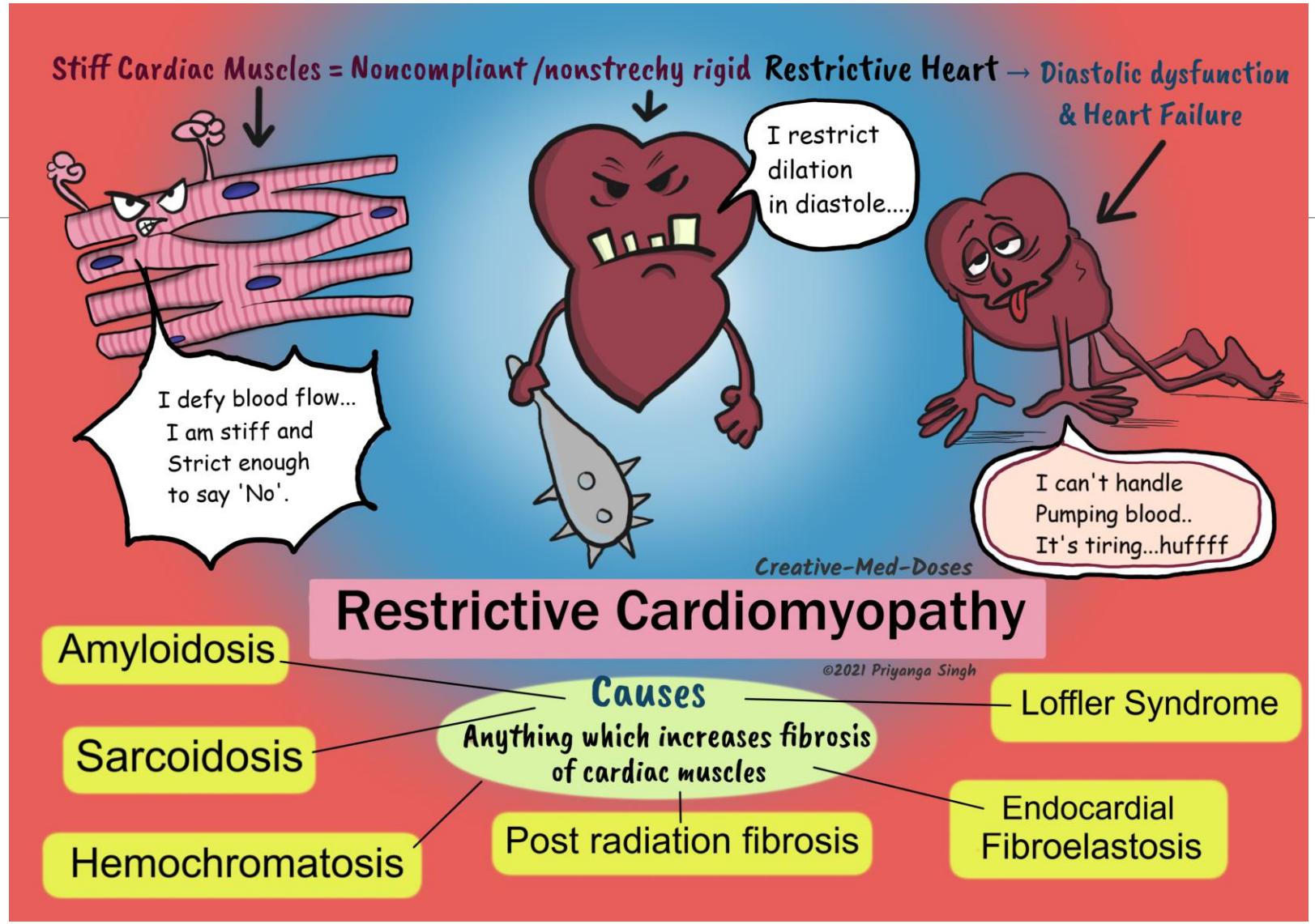
اختصار عشان نحفظهم

- ❑ Endocardial fibroelastosis (in kids) - there's fibrosis and elastosis in endocardium
- ❑ Loeffler syndrome - eosinophilic inflammation of endocardium and myocardium

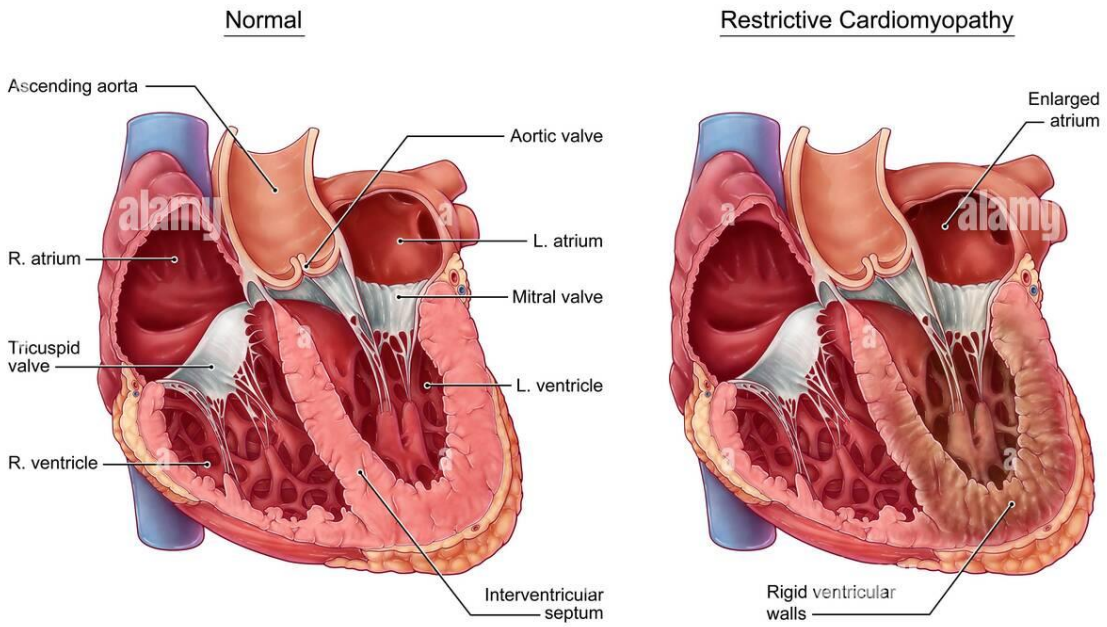
Amyloidosis:عن عبارة هي systemic disease misfolded protein فهاي ال protein ببلش يصير الها aggregate بال tissue فبتخلي النسيج thick وما بصيرلها dilation

Sacoidosis:هو systemic disease mainly the manifestation in the RS : granuloma ولكن ممكن برضو هاي ال granuloma تصيب القلب وتعمل fibrosis

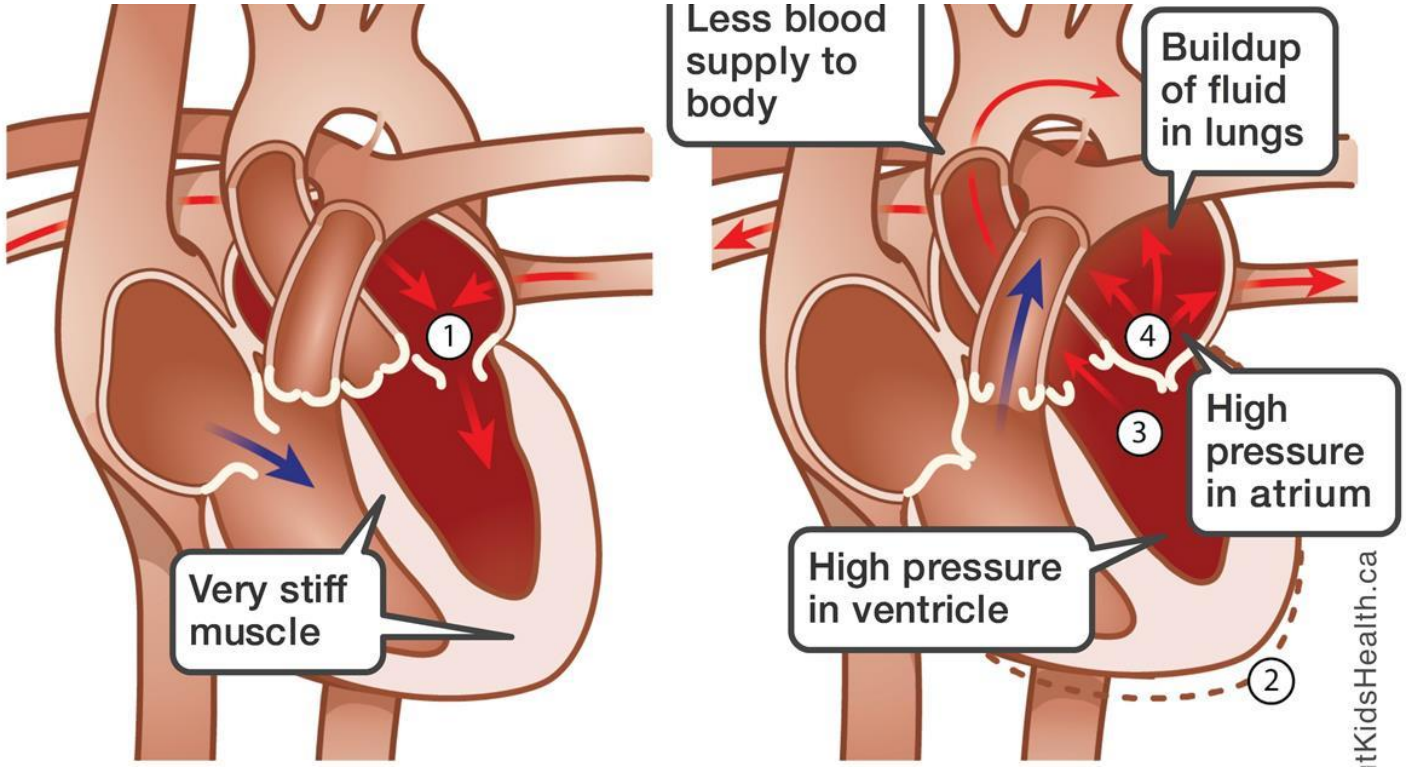
Radiation fibrosis: يعني لو واحد عنده cancer in the lung or around the heart واخد اشعاع كدواء complication فممكن تكون انو fibrosis in the heart انو يصير عندو


Stiff Cardiac Muscles = Noncompliant /nonstrechy rigid Restrictive Heart → Diastolic dysfunction & Heart Failure
I defy blood flow... I am stiff and Strict enough to say 'No'.
I restrict dilation in diastole....
I can't handle Pumping blood.. It's tiring...huffff
Creative-Med-Doses
Restrictive Cardiomyopathy
©2021 Priyanga Singh
Causes
Anything which increases fibrosis of cardiac muscles
Amyloidosis
Sarcoidosis
Hemochromatosis
Post radiation fibrosis
Loffler Syndrome
Endocardial Fibroelastosis

Restrictive Cardiomyopathy Morphology



- The ventricles are of approximately normal size or only slightly enlarged
- Cavities are not dilated, and the myocardium is **firm**.
- Endomyocardial biopsy often can reveal a specific etiologic disorder.



Restrictive Cardiomyopathy

Restrictive Cardiomyopathy
Endomyocardial fibrosis

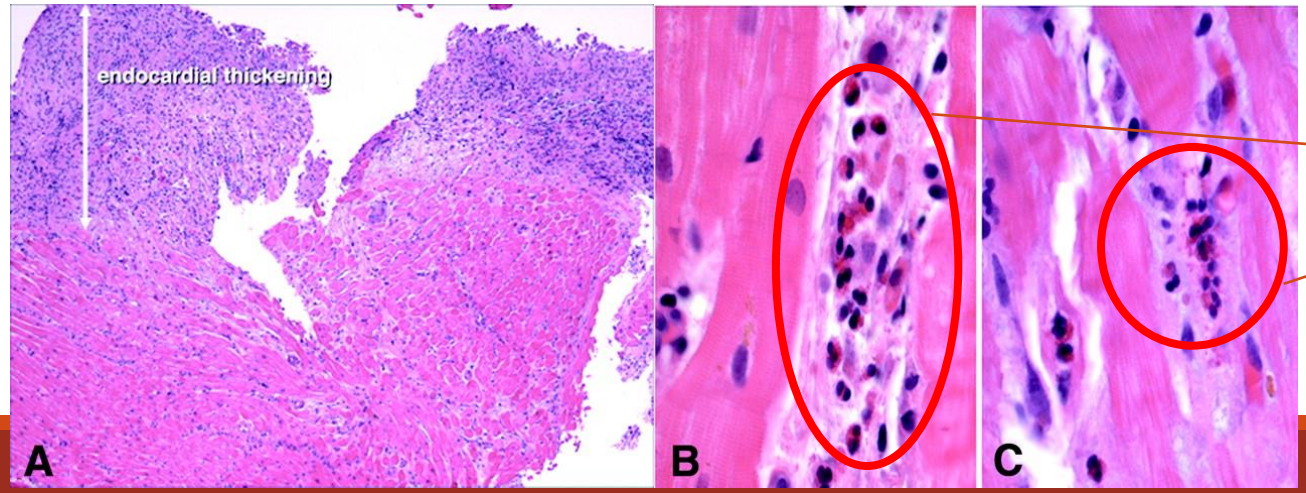
Endomyocardial fibroelastosis :

- Most common form of restrictive cardiomyopathy.
- Children and young adults in Africa and other tropical areas
- Dense diffuse fibrosis of the ventricular endocardium and subendocardium.
- Linked to nutritional deficiencies and/or inflammation related to helminthic infections

Restrictive Cardiomyopathy
Loeffler endo myocarditis

Loeffler end myocarditis:

- Endo-myocardial fibrosis, but without geographic predilection.
- Histologic examination: peripheral hyper eosinophilia and eosinophilic tissue infiltrates



Collection of interstitial eosinophils