



### CARDIOVASCULAR SYSTEM

SUBJECT : \_\_\_\_ Pathology LEC NO. : \_\_\_\_\_ 5

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#### **Cardiomyopathy and pericardium**

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# Cardiomyopathy

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



#### 🖀 We are looking to a cross section from left side 😁



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.

ventricl .....so the blood will acummulate in left atrium leading to left atrial dialation.

Ø Diastolic dysfunction

Hypertrophy mainly in the A left ventricle and venticular septum. Diastolic dysfunction



### Cardiomyopathy



- 2. Hypertrophic Cardiomyopathy
- **3.** Restrictive Cardiomyopathy



# Cardiomyopathy



- 1. Dilated Cardiomyopathy (Most common; approx. 90%)
- 2. Hypertrophic Cardiomyopathy
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#### **Dilated cardiomyopathy**



# Dilated Cardiomyopathy

عضلة القلب اللي فيها المشكله بتبطل contactile The myocardiam Can be strethched and filled with blood as normal durring diastol ,but the problem appears durring systole when the heart need to contarct the EF is affected . (normal EF = 70%......here EF is reduced almost =30%) وعني ال SV درح تقل .....بروح القلب بيتوسع (بيعمل. Dilation ) عشان اشاء ال diastol يعبي اكتر عشان يطلع اكتر.

ورب المسلمانية بالبرية. vv و كميه الدم اللي بتضلها بالقلب بعد ما يطلع ال↑. EF . ( EF).

Progressive cardiac dilation, usually with concurrent hypertrophy

Results in systolic dysfunction (ventricles cannot pump), leading to bi-ventricular CHF <sup>? dilation</sup> في fibrosis btwn myocardial Fibers

So the fibers won't be attached to each other ( the ventricle can not pump) Leading to bilateral congestive HF

■ All chambers are dilated → Like ballon

### **Dilated Cardiomyopathy DCM**





# 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.,  $\alpha$ -cardiac actin).

- X-linked: dystrophin gene mutations (Duchenne muscular dystrophy: DMD) -> They have problems

# Dilated Cardiomyopathy DCM Causes (cont.)



- **3.** Alcohol or other toxic exposure:
- Alcohol and its metabolites (especially acetaldehyde) have a direct toxic effect on myocardium



5. Thiamine B1 deficiency (Wet Beri Beri)



# Dilated Cardiomyopathy DCM Causes (cont.)

#### is,

#### **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)

اخدناه بال HLS اخدناه بال Patients with thalasemia need frequent

# 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.
stasis معظم الدم اللي بيفوت ما بيطلع فبيصير stasis معظم الدم اللي المعالية فيسيط فبيصير So mural thrombi formatio

Histologic abnormalities are nonspecific:

> Myocytes exhibit **hypertrophy** with enlarged nuclei.

Variable interstitial fibrosis



Hypertrophi myocyte

#### Small mural thrombi

#### Dilated Cardiomyopathy Morphology

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

B. **Typical myocyte hypertrophy & interstitial fibrosis** (Masson trichrome stain collagen blue). conductivity btwn myocyte معيق ال fibrosis ( arrythmia يصير arrythmia

#### 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%)



### Dilated Cardiomyopathy DCM Clinical features

**Complications:** 

Mitral and tricuspid regurgitation

Arrhythmia (heart's conduction system is stretched up)

Half of the patients die within 2 years, and only 25% survive longer than 5 years

بيبعدوال cusps عن بعض

Death usually is due to progressive cardiac failure or arrhythmia.

Cardiac transplantation is the only definitive treatment.

The drugs only reduce





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النوع الثاني من Cardiomyopathy وهون المشكله بتكون بال

اول اشى لازم نعرف معلومات عن هذا نوع 1-effect left side only (massive hypertrophy in LV Right side ----> normal 2-hypertrophy in ventricular suptum suptum ج تعون ملزقه بال suptum عنائل عون ملزقه بال suptum معن العامي معن ما يحدث contraction معن تسكر outFlow 4-chamber size very small بتالی عندی مشکله فی diastolic filling و هذا يؤدي الى انخفاض stroke volume will decrease 5-hypertrophic muscle بدها كثير blood والدم المتوفر قليل بتالي ممكن يصير عنده ischemia

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



### Hypertrophic Cardiomyopathy(HCM) Causes

Most common cause is <u>hereditary and is due to autosomal dominant</u> mutations in sarcomere proteins

المرض بصير بسبب mutations في اي واحد من هدول proteins 3

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



هاى المنطقة غير كافية ابدا انه يتعبى الدم فيها، لهيك بنشوف diastolic dysfunction واحنا بنعرف انه المسؤول عن تعبية الـ ventricle هو الـ atrium، ولائه ما في تعباية منيحة للدم بصير يتراكم الدم ....pulmonary edema, shortness of breath, etc... منيحة للدم بصير فبصير عنده lung وبعدها يرجع للـ atrium بالـ

**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
- كل خليه رايحه بأتجاه شكل و Haphazard myocyte disarray
- Interstitial fibrosis . ך



هون برضه بنلاحظ انه الـ ejection fraction طبيعية بس الـ CO يكون منخفض، ليش ؟

لانه اصلا كمية الدم اللي بتفوت عليه واللي بضخها قليلة بدي اعطيكوا ارقام وهمية، مثلا EDV هو 50 والـ ESV هو 20 هون فعلا الـ EF طبيعية ولكن كمية الدم قليلة



دائماً بس بكون في fibrosrs بكون عندي arithmetic

### 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.

القمةالوحيده يلي وصلتها هي قمه القرف

# Hypertrophic Cardiomyopathy(HCM) Alc Largery -> Cut part Filling 9. (1) July 2 Lasses 1- B or Cat channels blacker 2- Surgary -> Cut part From suptom

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.

# Cardiomyopathy



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**B.** Restrictive Cardiomyopathy

<u>جندعے شفلات بتجمع بالفلب Fibrosis</u> بندعے بند کے جند کر بند کر بم بند کر بند کر بند بند کر بند کر بم بند کر بند کر بند کر بند ک



#### **Restrictive Cardiomyopathy**

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

> normal

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

### Restrictive Cardiomyopathy Causes





#### Morphology Normal **Restrictive Cardiomyopathy** Ascending aorta Enlarged $\checkmark$ during diastole of ventricle $\rightarrow$ atrial try to contract to fill ventricle - atrium Aortic valve and when the ventricle is stiff, the pressure in ventricle is high →hypertrophy in atrium □ The ventricles are of approximately atrium normal size or only slightly enlarged Mitral valve ventricle Cavities are not dilated, and the م حيدتنا رها myocardium is **firm**. Endomyocardial biopsy often can Interventricular Rigid ventricular - septum walls reveal a specific etiologic disorder.

R. atrium

Tricuspid valve

R. ventricle

**Restrictive Cardiomyopathy** 



#### Restrictive Cardiomyopathy

### Restrictive Cardiomyopathy Endomyocardial fibrosis

- Endomyocardial fibroelastosis : Fibrosis + elestic = dense Fibrosis
- 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.

دود ہ

Linked to nutritional deficiencies and/or inflammation related to <u>helminthic</u> infections



هدول فيديو هات بيساعدوكم .....

#### https://youtu.be/2RvZQDj2ZNg?si=Z\_5tDJ9XbT300fJL

#### https://youtu.be/dMPUnvPgTVk?si=mlB8kPLLdbOsSDyM