

Cardiac diseases due to :
intrinsic myocardial dysfunction



Cardiomyopathies

Dilated cardiomyopathy (DCM)
(90% of cases)



Hypertrophic cardiomyopathy (HCM)

Restrictive cardiomyopathy(RCM).



مورثي

- flabby, poorly contractile
- Systolic dysfunction
- Low ejection fraction <40%

- thick-walled, heavy, and hypercontractile
- diastolic dysfunction
- normal to high ejection fraction

Def:

Dilated cardiomyopathy (DCM) is characterized by a **poorly contracting** dilated left ventricle with a normal or **reduced left ventricular wall thickness**

Dilated Cardiomyopathy

more frequently in **men** 🧑
between ages **20 and 60**

valvular and vascular lesions (e.g., atherosclerotic coronary artery disease) that can cause cardiac dilation secondarily are **absent**.

most common
cause of congestive
cardiac failure 😓

Dilated Cardiomyopathy

Pathogenesis

Genetic

X-Linked

- mutation in **dystrophin**

Autosomal dominant

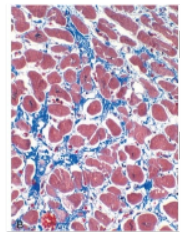
- mutation in **cytoskeleton proteins**

Acquired

- Infections: e.g. coxsackievirus, adenovirus.
- Nutritional deficiency: carnitine selenium deficiencies
- Cardiotoxins: e.g. Adriamycin: a chemotherapeutic drug.
- Puerperium "post partum"
- Alcohol :
 - ☒ direct toxic effect on the myocardium.
 - ☒ thiamine deficiency

Histology

- Myocytes exhibit hypertrophy with enlarged nuclei.
- interstitial and endocardial **fibrosis**,



Clinical features

The fundamental defect in DCM is ineffective contraction.

- slowly progressive CHF, including **dyspnea**, easy **fatigability**, and **poor exertional capacity**.
- Secondary **mitral regurgitation**.
- abnormal cardiac **rhythms**
- **embolism** from intracardiac (mural) thrombi.

Treatment

Cardiac transplantation

2. Hypertrophic Cardiomyopathy

defined by the presence of increased left ventricular (LV) wall thickness (in a non dilated chamber) that is not explained by abnormal loading conditions

- associated with defective diastolic filling, and ventricular outflow obstruction. [myocardium does not relax and therefore exhibits primary diastolic dysfunction]
- Systolic function usually is preserved

2. Hypertrophic Cardiomyopathy

Clinical Features

Pathogenesis

□ Autosomal dominant mutations affect contractile apparatus

- 1. β myosin Heavy chain
- 2. Myosin binding protein C
- 3. Troponin T

• manifests during the postpubertal growth spurt

• smaller size chamber → impaired Diastolic filling

↓ stroke volume

↓ cardiac output

↑ Pulmonary Venous Pressure

• high LV pressure

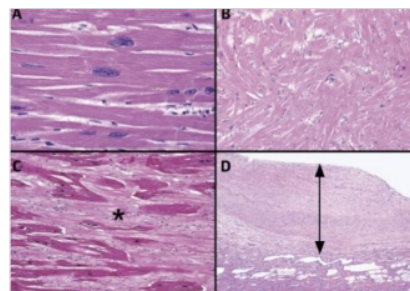
↓ Compromised intramural arteries

↓ Angina

• Atrial & Ventricular Fibrillation

Histology

- A) Myocyte hypertrophy.
- (B) myocyte disarray.
- (C) interstitial (pericellular-type) fibrosis (asterisk).
- (D) endocardial fibrosis (double-headed arrow).



one third of cases of sudden cardiac death in athletes younger than 35 years of age, the underlying cause is HCM.

□ Restrictive cardiomyopathy

- ↓ ventricular compliance
- Impaired ventricular filling during Diastole
- causes :
 - Δ Radiation Fibrosis
 - Δ Amyloidosis
 - Δ Sarcoidosis

⇒ Fibrosis is caused by :

- nutritional
- helminthic infection



Myocarditis

⊙ ***Inflammatory*** disease of the myocardium

⊙ ***Causes are :***


1. ***Infectious causes*** → coxsackie viruses A and B , enteroviruses, Cytomegalovirus (CMV), ***HIV***

2. ***Non infectious*** → ***SLE , drug hypersensitivity***



Cardiac Tumors

تتضمن

- ▶ cardiac tumors are rare. Cardiac tumors comprise primary and secondary metastatic tumors.
- ▶ Primary tumors:
- ▶ **Primary** cardiac tumors are **uncommon**; and usually **benign**.
- ▶ In descending order of frequency, the most common tumors are:
 - **Myxomas** (most common). 
 - **Fibromas**.
 - **Lipomas**.
 - **papillary fibroelastomas**.
 - **Rhabdomyomas** (most frequent in infants and children, they often regress spontaneously).
↳ *sk. muscles*
- ▶ **Angiosarcomas** constitute the **most common primary malignant tumor** of the heart.



Myxoma

- **Most common** primary benign tumors of the heart
- usually **single** in **sporadic** forms
- mainly located in the **left atrium**.

clinical manifestations:

- sudden death, usually due to **mitral valve obstruction**.
- valvular “ball-valve” obstruction
- embolization.
- fever and malaise.

 **diagnostic** modality of choice : ■ **Echocardiography**

 **surgical resection** is almost uniformly **curative**.

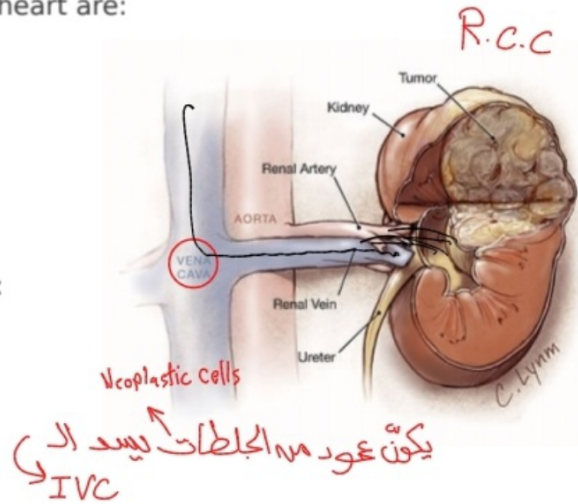
[mets] Secondary cardiac tumors

▶ The most frequent metastatic tumors involving the heart are:

- carcinomas of the lung. ↘
- Carcinoma of the breast. ↘
- melanomas. [skin]
- leukemia's and lymphomas.

▶ Metastases can reach the heart and pericardium by:

- lymphatic extension.
- hematogenous seeding
- direct contiguous extension. → Lung - breast
- venous extension



Carcinoid Heart Disease

- ▶ The carcinoid syndrome results from **bioactive compounds such as serotonin** released by **carcinoid tumors** (tumor arising from Neuroendocrine cells).
- ▶ Cardiac lesions typically do not occur until there is a **massive hepatic metastasis** since the liver normally inactivates circulating mediators before they can affect the heart.
- ▶ Classically, **endocardium** and **valves of the right heart** are primarily affected since they are the first cardiac tissues bathed by the mediators.

↳ Not Left side

▶ The mediators elaborated by carcinoid tumors include **serotonin** (5-hydroxytryptamine), **kallikrein**, **bradykinin**, **histamine**, **prostaglandins**, and **tachykinins**.

▶ systemic manifestations include :

- Flushing.
- diarrhea.
- Dermatitis.
- bronchoconstriction.

"لا يوجد في
هذه الحياة
كلمة أكثر رافة
ورحمة من كلمة
-يارب- ونبرة
الالتجاء
المصاحبة لها..
أن تترك العالم
كله، وتبتعد عن
الأهل والصحب،
وتنادي -يارب-،
بيقين أنه
يسمعك ويفهم
كل ما تخبئه وراء
هذه الكلمة..
الحمد لله
الرؤوف الرحيم."

يارب