MYOCARDIAL DISEASES

MYOCARDITIS
Disease of the myocardium - Inflammatory Infiltrate
Damage to myocytes
focal/diffuse
Unknown, asymptomatic, resolves in time.

Causes of myocarditis

Idiopathic - Giant cell Myocarditis
Infectious - Viral
- Rickettsial
- Bacterial
- Fungal
- Parasitic
Non - infectious: - Hypersensitivity/ Imm - med
- Drugs & Toxins
- Radiation
- Others
Clinical Features

- **Asymptomatic** - fulminant fatal course
  - fever, dyspnea, fatigue, palpitation, CHF
- Enzyme & EKG changes are seen

Morphology

**Gross:** normal / Dilatation of ventricles
- May mimic MI

**Micro:**
- Infiltrates Ly, Mo, Neu & Eos
- Granuloma & giant cells
  - Interstitial Edema
  - Myofiber Necrosis
DEFINITIVE DIAGNOSIS

Endomyocardial biopsy & Histology

Recombinant DNA Tech. -Viral genome

CARDIOMYOPATHY (C.M.P.)

Disease of the Heart Muscle (WHO/IFOC)

Heart Muscle Diseases of unknown cause excluding those due to Cardiac Valves, Coronary Artery Disease, Cong. H. D. Pericardial Disease, etc.

• Non inflammatory, non ischemic, non hypertensive cardiac muscle disease
**Primary/Idiopathic Cardiomyopathy**

Profound and Fatal C. H. F

**Three categories of CMP:** based on Structure & Patho-physiology

- Dilated - most common
- Hypertrophic
- Restrictive - least common

New entity: Arrhythmogenic RV CMP/dysplasia

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**DILATED CARDIOMYOPATHY (DCM)**

- Gradual development of cardiac failure associated with four-chamber hypertrophy and dilatation of heart of unknown cause.
Aetio pathogenesis

Auto Dom
Genetic factors → Auto Rec.
(30-40%)
X-linked

Suspected underlying mechanisms
Alcohol/toxic agent (Adriamycin)
Inflammatory - Viral/Non-viral myocarditis
Pregnancy associated
Nutritional deficiency - Beri Beri
Immunologic
Mutations: sarcomere, Cytoskeleton, Nuclear env, Mito

MORPHOLOGY

Gross: Heart - Heavy - 1 1/2 - 2 N Weight

Large and flabby. All chambers are dilated
Dilatation - Ventricular wall - Thin of Thick
Mural Thrombi Thromboembolism
M. V. R. - Valve ring dilatation
Coronaries - N or obstruction healed ischaemic lesions
**MORPHOLOGY**

**Micro:** Variation in myocyte size
- Hypertrophic, or attenuated
  - Nuclei - large, Interstitial & Endocardial
- Fibrosis

**Final Abnormality:**
Flabby hypocontracting heart with ___systolic failure
Clinical Features & Treatment

- Any age (20 - 60 yr.), Dyspnoea, chest pain, fatigue, palpitation, features of CHF - progressive and unremitting

Rx: Cardiac transplantation
HYPERTROPHIC CARDIOMYOPATHY  
(HCM)

Idiopathic Hypertrophic Subaortic  
Stenosis/Hypertrophic obstructive CMP  
Ventricular muscle thickening  
Irregular hypertrophy  
Myocyte disarray

Important Genes responsible for this familial  
disease  
Genes are:  
Chr. 14 - β-heavy chain myosin  
Chr. 1 - Troponin - T  
Chr. 15 - α - Tropomyosin

Responsible for abnormal myofibril formation  
and alignment
MORPHOLOGY

- Massive myocardial hypertrophy - \( \text{\( \text{\textup{\texttimes}} \) wt.} \)
- Asymmetric septal hypertrophy - classic
- Ventricular cavity - ' Banana Shaped ' 
- Anterior mitral valve leaflet thickened
- L. V. outflow narrowed, endocardial thickening
MORPHOLOGY

Micro:
Extensive myocyte hypertrophy
Disarray of bundles, myocytes and myofibres
Interstitial fibrosis
Small vessel disease

Functional Abnormality: - Heavy muscular hypercontracting heart with diastolic dysfunction
CLINICAL FEATURES

• Palpitation, dyspnoea, cardiac murmur, and sudden death in young males

TREATMENT : Septal surgery
RESTRICTIVE CARDIOMYOPATHY

Diastolic relaxation and L. V. chamber filling is impeded

Morphology: Gross:
Ventricles - N or slightly enlarged, cavity N
Myocardium – Firm, Biatrial dilatation

Micro: Patchy/diffuse Interstitial fibrosis

Functional Abnormality:
Diastolic function impaired, systolic - N or impaired. Variants/subtypes:-

ENDOMYOCARDIAL FIBROSION

Unknown cause
Children, young adults - Africa and Tropics

- Fibrosis of ventricular endo/ Subendo, Apex & Inflow tract
  Reduced compliance of chamber
  Mural thrombi
Lefflers Endomyocarditis

- Endomyocardial fibrosis & mural thrombi
  Eosinophilic infiltration - release toxic products - Major Basic Protein
  Myocardial damage
  Rx Endomyocardial stripping
Endocardial fibroelastosis

uncommon, unknown cause
Age: 1-2 years
Focal/diffuse cartilage like fibroelastic thickening
Assoc. with Cong. Cardiac anomalies
- CCF

LAB DIAGNOSIS

X-ray, ECG, ECHO, Cardiac Cath & Endomyocardial Biopsy
Endomyocardial Biopsy:
Bioptome → (R) Internal Jugular Vein (R) → Heart → R. V. Septum - Snip (1-3mm)
Study → Virology
  E.M.
  Imm. Histo.
  Biochemical
  Molecular
COMPLICATIONS OF CMP

Heart failure

Cardiac arrhythmia’s

Atrial

Ventricular

Mural thrombi and embolism

Endocarditis and sudden death - HCM

SUMMARY

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<th>RCM</th>
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<tr>
<td>L.V. Cavity</td>
<td>Dilated</td>
<td>Small</td>
<td>Normal</td>
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<tr>
<td>L.V. Wall Thickness</td>
<td>N, ↓</td>
<td>↑</td>
<td>N or ↑</td>
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<tr>
<td>Systolic Function</td>
<td>↓</td>
<td>↑ or N</td>
<td>N or ↓</td>
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<td>Diastolic Function</td>
<td>N</td>
<td>↓</td>
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<tr>
<td>Septal Asymmetry</td>
<td>-</td>
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