

Cardiomyopathies

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cardiomyopathies

- **Definition:** A group of cardiac muscle disease , of unknown etiology.
- The heart muscle, become enlarged - DCM.
- Thick – HOCM
- Rigid – Restrictive CMP .
- Replaced by scar(fat) tissue – ARVD.

CLASSIFICATION

- 1. dilated cmp (dcm).
- 2. hyperthrophic cmp (hocm).
- 3. restrictive cmp.
- 4. arrhythmogenic rv dysplasia (arvd).
- cmp can be congenital or acquired.
- cmp can be primary or 2dry.

COMMON FEATURES

- I. clinical manifestations: 4 syndromes
- 1. CHF
- 2. Arrhythmia.
- 3. Thromboembolic complications.
- 4. Valvular problems.
- II. PROGNOSIS : causes of death.
- 1. 80 – 85 % - arrhythmia.
- 2. 10 – 15 % - thromboembolic complications.
- 3. 5 – 10 % - CHF (cardiac cachexia).
- III. TREATMENT (as CHF).
- 1. Lifestyle changes.
- 2. drugs .
- 3. Surgery.
- 4. Implanted devices to correct arrhythmia and improve contractility.

.DCM (DILATED CMP)

- Etiology : primary – unknown cause: congenital or acquired at any age).
- 2dry – any cardiac disease.
- 1.IHD – ischemic CMP.
- 2.HTN – hypertensive CMP.
- 3. VALVULAR HD – valvular CMP.
- 4.post myocarditis.
- 5.metabolic causes (DM, thyroid diseases).
- 6. peripartium.
- 7. Alcohol .
- 8. toxic – cobalt.
- 9. drugs – cocaine, amphetamines, doxorubicin.
- 10. tachycardia induced CMP.

.SYMPTOMS AND SIGNES

- 1.CHF: dyspnea, low cardiac output symptoms.
- 2. Arrhythmia : palpitation.
- SIGNS: LL oedema, ascitis, anasarka,raised jvp(congested,pulsating neck veins), hepatomegaly (enlarged tender liver).
- Precordial examination :
 - 1. apex beat shifted outward, downward.
 - 2. heart sounds muffled , and gallop rhythm (3d HS).
 - 3.MR , TR murmurs.

DIAGNOSIS

- I. ECG : LVH, ARRHYTHMIA (PVCs, AF) .
- II. CXR : cardiomegaly, signs of lung congestion.
- III. ECHODOPPLER (diagnostic).
 - 1. dilated all chambers.
 - 2. global hypokinesia.
 - 3. low EF.
 - 4. MR , TR.
 - 5. Thin wall (excentric LVH).
- OTHERS :
 - 1. Stress (dobutamine Echo).
 - 2. coronary angiography : to r/o ischemic cause.
 - 3. Isotope scanning : to asses viability of myocardium.

TREATMENT OF DCM

- i. lifestyle :
 - 1. healthy diet : fruits, vegetables, fish, white meats, fat free or low fat milk and milk products , salt restriction.
 - 2. smoking cessation.
 - 3. weight reduction.
 - 4. enough sleep and rest.
 - 5. reducing stress.
 - 6. treating dm , htn.
- ii. drug treatment: optimal medical therapy (omt) as chf.
 - 1. diuretics: loop d (frusemide), k sparing d (spironolactone).
 - 2. ace inhibitors or arb.
 - 3. b blockers(bisoprolol, metoprolol, carvedelol, nibovelol).
- others : digoxin. warfarin – if af present.
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TREATMENT OF DCM

- III. DEVICES:
- ICD (implantable cardiovertor defibrillator).
- CRT (cardiac resynchronization therapy).
- LVAD (left ventricular assist device)'
- Surgery : heart transplantation .
- Used in end stage (refractory) HF, the condition is so severe , that all treatments , other than transplant , have failed.

HYPERTROPHIC CMP (HCM , HOCM)

- IN this type there is asymmetrical LVH, mainly septum, which can lead to LV obstruction (HOCM). Very common and can affect people of any age, about 1 of every 500.
- This is the most common cause of sudden cardiac death in young people, including athletes.
- ETIOLOGY : primary – autosomal dominant .
- 2dry – association with:
 - 1. pheochromocytoma.
 - 2. acoustic neuroma.
 - 3. neurofibromatosis.
 - 4. Dushene myopathy.

SYMPTOMS AND SIGNS

- 1. dyspnea.
 - 2. palpitation.
 - 3. thromboembolic complications.
 - 4. dizziness and syncope.
 - 5. chest pain.
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- PRECORDIAL EXAMINATION as AOS.
 - 1. apex beat shifted downward, forceful, localized, heave sustained.
 - 2. 1st HS loud.
 - 3. ejection systolic murmur over 1st AO area , without radiation to carotids.
 - 4. MR murmur.

SYMPTOMS AND SIGNS

- ECG: Severe -ve T wave seen in $V_4 > V_5$ & V_6 due to symmetrical strain pattern hypertrophy.
- Echo: 1- Asymmetrical LVH
- 2- Pressure Grading Changes
- 3- MR
- 4- EF $> 80\%$
- 5- Systolic anterior motion (SAM) with Asymmetrical septal hypertrophy (ASH)

Treatment

- For Diastolic Dysfunction:
- B Blockers (propranolol) or Ca channel blockers for eg (Verapamil)
- Note: Don't Give Vasodilators for patients with HOCM & AS as it would increase the pressure gradient between the chambers.

Treatment

- For Arrhythmia:
- Amiodarone or pacemaker.
- Surgery: Resection of the septum.
- Myotomy: complicated by AV block
- Myomectomy.
- Non Surgical : Non surgical septal ablation by alcohol infusion in septal perforators of LAD.

Indications for ICD

- Family history of Sudden cardiac death.
- Family history of HOCM.
- VT or VF sustained on ECG.
- Marked increase in LV thickness > 25 mm.
- Systolic blood pressure not increased by exercise

Restrictive CMP

- Causes: 1. Primary
- 2. Secondary:
- A- Sarcoidosis.
- B- Amyloidosis.
- C- Loffler hypereosinophilic syndrome.
- D- Carcinoid syndrome.

Presentation

- Refractory HF.
- Ventricular Filling Impaired due to Ventricular Stiffness so increased Atrial pressure and atrial hypertrophy, Dilatation and later AF may develop.
- Restrictive pattern In doppler:
 - 1- 3- 3.5 ration
 - 2- MR, TR
 - 3- Bilateral Atrial Enlargement

Treatment

- Restrictive CMP is an indication for heart transplantation.

Arrhythmogenic RV dysplasia

- Defined by fatty or fibrous tissue replace the RV myocardium which may lead to Arrhythmias due to abnormal conduction.
- Clinically:
 - 1- Arrhythmias.
 - 2- Sudden Cardiac Death.
 - 3- Rt Sided HF.

- **ECG findings**

- Epsilon Wave: U wave Before T wave

- **Treatment**

- ICD for Arrhythmias.
- Medical treatment of HF.
- Radiofrequency Ablation.

Note

- The most common causes of death at rest are:
- 1- Restrictive CMP.
- 2- Prolonged QT interval.

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