# UNIT 5 CARDIOMYOPATHIES

## Structure



## 5.0 OBJECTIVES

After reading this unit, you will be able to:

- describe the classification, clinical presentation and management of cardiomyopathy; and
- describe causes, clinical feature and management of myocarditis.

## 5.1 INTRODUCTION

Cardiomyopathy is a primary disorder of heart muscle that may cause cardiac dysfunction and is not related to any obvious disease process. It is heart muscle disease of unknown cause or association.

There are many ways in which Cardiomyopathies are classified e.g. WHO classification, Functional classification, Etiologic classification, classification based on endomyocardial Biopsy findings and therapeutic classification. The two commonly used are Functional (Table 5.1, Fig. 5.1) and Pathological (Table 5.2 ). The WHO/International Federation of Cardiology classification is in Table 5.3.

**Table 5.1: Functional Classification of Cardiomyopathies**

| | Dilated | Restrictive | Hypertrophic |
|---|---|---|---|
| A) | **Symptoms** | | |
| | CCF, particularly Lt. sided Syst. and pulm. emboli | Rt. sided failure systemic disease viz. amyloidosis, iron storage disease | Dyspnoea, Angina Fatigue, Synocope palpititons |
| B) | **Physical Exam.** | | |
| | Mod to severe cardiomegaly, $S_3$ $S_4$ A-V valve incompetence particularly mitra | Mild to mod cardiomegaly $S_3$ and $S_4$ A-V Valve regurg, Kussmaul sign + ve (Insp. in venous pressure) | Mild cardiomegaly $S_4$. Brisk carotids SM with Valsava |
| C) | **X-Ray Chest** | | |
| | Pulm. venous Hyp. | Pulm. Venous Hhyp. | LA+ |

D) **ECG**

| | | |
|---|---|---|
| Sinus tachycardia | Low voltage | LVH |
| ST and T Changes | I.V. Conduction defects | ST and T Changes |
| I.V. Conduction | A.V. Conduction defects | Abnormal Q |
| Defect Atrial and V. arrhythmia | | Atrial and V. arrhythmic |

E) Echocardiography

| | | |
|---|---|---|
| LV dilatation | LV thickness | Asymmetric septal hypertrophy |
| LV dysfunction | Small or normal sized | Narrow LV outflow tract |
| Abn diastolic mitral valve notio secondary to abnormal compliance and filling pressures. | LV Cavity Normal syst. Function pericardial effusion. | Systolic anterior motion of mitral valve smallor normal sized LV. |

**Fig. 5.1: Morphologic types of cardiomyopathy**

**Table 5.2: History Classification of Cardiomyopathy**

**Endomyocardial Bispsy Histology Classification of Cardiomyopathy**

I) Inflammatory/immune cardiomyopathy
II) Infectious cardiomyopathy
III) Infiltrative cardiomyopathy
IV) Cardiac tumors
V) Miscellaneous specific cardiomyopathies
VI) Nonspecific abnormalities
VII) No histological abnormality

**Table 5.3: WHO/International Classification of Cardiomyopathy**

| | Category | Definition |
|---|---|---|
| 1) | Dilated (DCM) | EDV ESV EF |
| | (j) Primary | |
| | (ii) Secondary | |
| 2) | Restrictive (RCM) | EDV ESV EF FP |
| | (i) Primary | |
| | (ii) Secondary | |
| 3) | Hypertrophic (HCM) | Septal and Post. wall thickness, |

| | | |
|---|---|---|
| | | Myofibrillar disarray. Mutation in protein Autosomal dominant inheritance |
| 4) | Arrhythmogenic RV Dysplasia (ARVC) | Fibrofatty Replacement of RV myocardium. Autosomal dominant (most and recessive inheritance). |
| 5) | Unclassified | Not meeting criteria for other categories |
| | (i) Primary | features of > one category |
| | (ii) Secondary | |

***Note:*** **ESV** - Endsystolic volume
**EDV** - Enddiastolic volume
**EF** - Ejection fraction
**FP** - Filling Pressure

**Check Your Progress 1**

What is the definition of cardiomyopathy?

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## 5.2 DILATED CARDIOMYOPATHY (DCM)

It is a disease of unknown etiology, affecting myocardium. Its diagnosis is established by presence of left ventricular dilatation and systolic dysfunction in absence of congenital, valvular, hypertensive, coronary or pericardial heart disease. (Fig. 5.2).

**Fig. 5.2: Gross pathology of dilated cardiomyopathy. Prominent ventricular dilatation is apparent**

It's prevalence rate is 0.04 per cent. Although the cause is not definable, more than 75 specific disease of heart muscle can result in a picture of DCM. It represents a final common pathway that is the end result of myocardial damage, produced by a variety of agents.

**Clinical Manifestations**

1) Asymptomatic, when the diagnosis is made by 2D Echocardiography.
2) Enlargement of LV. Apex is shifted down and out. The apex impulse is not forceful. Presystolic gallop ($S_4$) may precede the development of failure. Ventricular gallop ($S_3$) is common during decompensation. Regurgitant murmurs are common. Mitral regurgitation is due to enlargement and abnormal motion of mitral annulus and distortion of the geometry of subvalvar apparatus. Ventricular dilatation plays a lesser role. 2nd sound may show reverse split, if LBBB occurs. They present with symptoms and signs of heart failare, as follows:

   a) ***Pulmonary Congestion***

      Dyspnoea on exertion or at rest, paroxysmal nocturnal dyspnoea. Tachypnoea, pulmonary rales, bronchial spasm and occasional rhonci.

   b) ***Systemic Congestion***

      Distended neck veins, palpable tender liver and edema of legs and at times ascites, and pleural effusion.

   c) ***Fall in Cardiac Output and Poor Peripheral Perfusion***

      Fatigue, weakness, Narrowing of pulse pressure, hypotension, Cold and pale extremities with constricted peripheral veins and delayed capillary refill.

   d) ***Arrhythmias***

      Palpitations, light headedness, syncope etc.

**Electrocardiogarm**

Poor R-wave progression, in precordial leads, or q-waves in anterior leads. Intraventricular conduction defects, esqecially LBBB are common. There is LA enlargement. Sinus tachycardia and atrial/ventricular arrhythmias are common. Complex ventricular arrhythmias, including VT/NSVT may predict sudden death.

**X-Ray Chest**

It shows cardiomegaly with left ventricle (LV) and left atrium (LA) enlargement. There may be signs of pulmonary venous hypertension viz. pulmonary venous redistribution, interstitial edema, alveolar edema and pleural effusion. Systemic venous hypertension may cause dilatation of azygos and Superior Vena Cava (SVC).

**2D Echocardiography**

It shows left sided or 4 chamber enlargement with reduced EF. The wall thickness is usually normal. There is generalized hypokinesis. Occasionally segmental wall abnormalities are present Mitral and tricuspid incompetence is seen on Doppler.

**Coronary Angiography**

It shows normal arteries or insignificant coronary artery disease.

**Complications**

Left and ventricular failure, hypotension. Arrhythmias (both atrial and ventricular) and embolic episodes from LV thrombi. Certain clinical and echo features suggest adverse outcome (Table 5.4).

**Table 5.4: Adverse Outcome in Dilated Cardiomopathy**

| (A) Clinical | (B) 2D |
|---|---|
| • NYHA Class III / IV | Low EF |
| • age, S3 | Marked LV dilatation |
| • Marked intraventricular conduction delay | Low LV Mass |
| • Complex V. arrhythmias | ≥ mod mitral regurgitation |
| • Abnormal signal averaged ECG | Abnormal contractile Reserve |
| • Low exercise peak oxygen consumption | Right Ventricular dilatation or dysfunction. |

**Management**

***Diagnostic Studies***

They are necessary to establish diagnosis, evaluate prognosis and help in medical therapy. They include chest X-Ray, ECG, 2D Echocardiography with Doppler. Coronary angiography is indicated in patients above 40 or when there is history suggestive of ischaemic heart disease. 24 hours ambulatory ECG is necessary when arrhythmias are suspected. Exercise testing is done for assessment of functional capacity. Endocardial biopsy is hardly useful. However, some recent reports have suggested inflammatory etiology in almost 50 per cent with immunohistological methods.

***Medical Therapy***

Many reversible causes of, cardiomyopathy should always be kept in mind (Table 5.5)

i) In asymptomatic stage, preventive therapy with ACE-I/ARBs/β- blockers may help.

ii) If on special diagnostic methods, viral load and/or inflammation is seen, a course of interferon, corticosteroids and azothiaprine may help.

iii) Treatment of cardiac failure viz., ACE-I/ARBs, β-Blockers, digitalis, diuretics are needed. Biventricular Pacing/resynchronization therapy may be useful in patents with wide QRS (> 150 msec), LBBB and LV dysfunction. LV assist devices, and cardiac transplant are other methods but as yet, they are not widely available. Adjunctive therapy includes anticoagulation in subjects with low EF to prevent thromboembolic complications, amiodarone to treat symptomatic arrhythmias, maintaining potassium in high normal range

(4.3–5mg/ml ), to prevent sudden death and frequent visits to clinic for proper adjustment of drug dosages, particularly β- Blockers.

**Table 5.5: Potential Reversible Cause of Dilated Myopathy**

| | |
|---|---|
| **Ischaemic with viable** | **Endocrine** |
| Myocardium | Hyperthyroidism |
| **Valvular with** | Pheochromocytoma |
| Correctable lesion | **Metabolic** |
| **Tachycardia induced** | Hypocalcemia |
| Inflammaory | Hypophosphotemia |
| CMV | Uremia |
| Toxoplasmosis | Carnitine deficiency |
| Mycoplasma | **Nutritional** |
| Lyme | Selenium deficiency |
| **Toxic** | Thiamine deficiency |
| Alcohol | **Infiltrative** |
| Cocaine | Hemachromatosis |
| Cobalt | Sarcoidosis |
| | **Hypersensitivity** |

**Few Specific Dilated Cardiomyopathies**

1) ***Anthracycline Cardiomyopathy***

It is due to anticancer agent doxorubion (Adriamycine), when the cumulative dose exceeds 450 mg/m$^2$ in subjects with normal hearts. In presence of heart disease or other risk factors viz. prior mediastinal radiation or when radiation treatment follows chemotherapy, the cardiomyopathy can occur at lower dosage.

2) ***Postpartum Cardiomyopathy***

It is defined as presentation of LV Systolic dysfunction and heart failure in last trimester of pregnancy or within 6 months of delivery. The treatment is like that of dilated cardiomyopathy. Majority will improve and 50 per cent recover completely. Subsequent pregnancy is absolutely contraindicated even if recovery is complete.

3) ***Alcoholic Cardiomyopathy***

In a case of dilated cardiomyopathy alcohol cardiomyopathy is suspected if there is history of alcohol intake of 100g/day for more than 10 years and when patient presents with high cardiac output.The cause is alcohol toxicity and thiamine ($B_1$) deficiency.

4) ***Cardiomyopathy due to Persistent Tachycardia***

In occasional cases, particularly in children recurrent or incessant episodes of supraventricular or ventricular tachyarrhythymias are actually the cause of and not the result of ventricular dysfunction. Restoration of sinus rhythm or slowing of the heart rate will reverse LV dysfunction. Thus this becomes curative and should not be missed.

5) ***Chagas' Cardiomyopathy***

It is common in south and Central America and is caused by Trypanosoma cruzi.

6) ***Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)*** **( Fig. 5.3)**

It is marked by myocardial cell loss with partial or total replacement of RV muscle by adipose and fibrous tissue. The clinical manifestations are seen in adolescence or early

adulthood, predominantly in males. Physical examination is normal. ECG shows inverted T in right precordial leads. They develop reentrant ventricular tachyarrhythmias of RV origin. LBBB configuration, usually precipitated by exercise. Sudden death is common. Management includes β-blockers, sotalol and amiodarone. Radiofrequency ablation, ICD and cardiac transplant are other options.

**Fig. 5.3: Arrhythmogenic right ventricular dysplasia**

**Check Your Progress 2**

1) What are the echocardiographic features of dilated cardiomyopathy?

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2) What abnormalities of heart sound and murmurs may be present in dilated cardiomyopathy?

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3) What are the some of the complications of dilated cardiomyopathy?

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## 5.3 RESTRICTIVE CARDIOMYOPATHY

It is a systemic or idiopathic disorder of the myocardium with clinical and hemodynamic features of diastolic dysfunction, closely simulating constrictive pericarditis. Restrictive cardiomyopathes could be myocardial or endomyocardial. The former could be noninfiltrative (e.g., idiopathic or familial), infiltrative (e.g., amyloidosis) or storage (e.g., hemochromatosis). The endomyocardial

group could be obliterative (e.g., Endomyocardial Fibrosis) or nonobliterative (e.g., radiation, drugs).

**Clinical Menifestation**

The symptoms are those of pulmonary and systemic congestion viz. Dyspnoea, nocturnal dyspnoea, ankle edema, abdominal discomfort. The findings are those of raised filling pressures on two sides of heart - Raised JVP with prominent X and Y descent. (Y is more prominent than X), enlarged tender liver and ankle edema, and also signs of pulmonary venous congestion viz. S3, rales over both lungs.

**Electrocardiography**

It is always abnormal. LBBB is common but RBBB also can occur. There may be cardiac arrhythmias. Atrial fibrillation is common. Low voltage occurs in cardiac amyloidosis.

**X-Ray Chest**

There are two major features (1) Absence of cardiomegaly (2) signs of pulmonary venous hypertension.

**2D Echocardiography**

The heart is only minimally dilated and there is no thickening of the myocardium in idiopathic causes, but may be thickened in infiltrative disorders. Both atria are enlarged. The filling pattern on Doppler may simulate that of constrictive pericarditis, but early rapid filling is more rapid in some. Prominent E-wave suggests that. It also has severely reduced deceleration time indicative of raised left atrial pressure. LV thickness is usually less than 1.7cm. Ventricular systolic function is normal. LVED volume is not more than 110 ml/m2 and LV end diastolic dimension in not more than 6 cm.

In endomycardial fibrosis, there is diminution of ventricular volumes, frequently associated with complete obliteration of apices of both ventricles.

**Diagnosis**

In a patient, presenting with congestive cardiac failure, absence of obvious cause, normal heart size with enlarged atria and bundle branch blocks on ECG suggest the possibility of restrictive cardiomyopathy. The common differential diagnosis is constrictive pericarditis (Table 5.6).

**Table 5.6: Effects of Interventions that Increase and Decrease Outflow Gradient and Systolic Murmur in HOCM**

**Management**

There is no satisfactory medical treatment. It remains essentially symptomatic.Some types of endomyocardial fibrosis can be managed surgically.

**Check Your Progress 3**

What happens to the heart size and wall thickening in restrictive cardiomyopathy?

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## 5.4 HYPERTROPHIC CARDIOMYOPATHY

It is a genetic disorder due to mutations in the gene that encodes for β-Cardiac myosin heavy chain (Localised to chromosome 14). It is characterized by inappropriate myocardial hypertrophy in absence of hypertension or aortic stenosis. There is myocardial disarray and interstitial fibrosis (Fig 5.4 a, b). It has asymmetric septal hypertrophy, left ventricular outflow obstruction due to systolic anterior motion of mitral valve, LV diastolic dysfunction, myocardial ischaemia and arrhythmias. LV outflow obstruction occurs only in about ¼th. The prevalence of HCM is about 0.2 per cent.

**Fig. 5.4(a): Myocardial disarray and interstitial fibrosis in HCM**

**Fig. 5.4(b): Interstitial fibrosis in a case of HCM**

## Clinical Menifestation

Patient may be asymptomatic. Chest pain, atypical or typical angina and dyspnoea are usually presenting symptoms in 30-60 per cent in 90 per cent of symptomatic patients. Unexplained syncope (15-25 per cent) and sudden death are known. It presents as cardiac failure in end stage disease.

The clinical types are (a) either symptomatic or asymptomatic. (b) non-obstructive or obstructive (at LV outflow tract or midventricular level (c) type of hypertrophy viz asymmetric septal hypertrophy (in two third), symmetric LV hypertrophy (5 per cent) and apical as in apical cardiomyopathy in Japanese population. (d) Endstage disease. This presents as cardiac failure.

It may rarely be unremarkable.Usually the apex is palpable and will be LV type and may have double apical impulse due to powerful LA contraction.Prominent S4 and systolic murmur at lower sternal border are heard. The murmur is due to labile LV outflow obstruction (Fig 5.5) and it increases with Valsalva manoeuvere, standing up from squatting position, exercise and in postextrasystolic (Fig 5.6) contraction. The SM is harsh and crescendo- decrescendo in configuration. It commences sometime after S1 and is best heard between apex and left sternal border. It radiates to lower sternal border, axilla and base of the heart, but never to carotids. MR murmur may also be present. Brisk carotid pulse and sometimes prominent ‘a’ in JVP are seen.

**Fig. 5.5: Payhophysiology of subaortic anterior motion (SAM) of mitral valve in HOCM**

**Fig. 5.6: Postextrasystolic potentiation of systolic murmur**

## Electrocardiography

The ECG changes usually precede the onset of echocardiographic changes. The abnormalities are seen in QRS, ST Changes and T-Waves. LV voltage is increased. Abnormal Q are seen in 25-50 per cent of patients, usually in inferolateral leads and may mimic myocardial infarction. Early repolarisation and other ST Changes may be seen. Giant negative T-Waves are seen in apical cardiomyopathy. LA is enlarged, Arrhythmias (NSVT 25 per cent, PSVT 35-50 per cent, AF 5 per cent) are seen. In 5 per cent WPW may be seen.

## Echocardiography

It is the most important diagnostic tool. There is asymmetric septal hypertrophy as seen by septal to posterior wall thickness ratio of 1.5 or more. Maximal septal hypertrophy occurs midway between apex and base. Apical hypertrophy, midventricular hypertrophy or RV hypertrophy may be seen. LV thickness can vary from 15 mm to 60 mm. Systolic anterior motion of the mitral valve identifies LV outflow tract obstruction. There is also elongation and enlargement of mitral valve leaflets, leading to abnormal aortic outflow geometry. It contributes to pressure gradient across LV outflow and is also responsible for mitral regurgitation.

There are other echocardiographic findings viz. small LV cavity, partial systolic closure and coarse fluttering of aortic valve (Fig 5.7), reduced septal and exaggerated posterior wall motion.

**Fig. 5.7: M-mode echo shows premature cosure of aortic valve (due to abrupt decline in midsystole) and also prominent $S_4$**

## Diagnosis

Nonobstructive hypertrophic cardiomyopathy needs to be differentiated from silent aortic stenosis, hypertension and physiologic hypertrophy seen in athletes. Elite athletes may demonstrate LV thickness upto 16mm. (normal 12) with ECG abnormalities. However they will not show myocardial disarray or diastolic dysfunction. They will also have an excellent exercise capacity.

The differential diagnosis of SM is shown in Table 5.7. The interventions that increase/decrease outflow gradient and hence SM are shown in Table 5.8. The effects of these interventions on other systolic murmurs are depicted in Table 5.9.

**Table 5.7: Differential Diagnosiso of Left Nautricular Outflow Obstruction Murmurs**

| Type of stenosis | Predominant site | Aortic ejection | A2 | a.1 murmur | Arterial pulse |
|---|---|---|---|---|---|
| Acquired A.S. | 2nd Rt. sternal border to neck; at apex in aged. | Uncommon | or Ab | Common | Delayed, upstroke, anacrotic notch +, small amplitude |
| HOCM | 4th lt. sternal border to apex.+ murmur, a little after S1., Never Conducted to carotids, m.i. murmur at apex | Rare | N or | V. Rare | Brisk upstroke or Bisferiens |
| Congenital valvular | 2nd right sternal border to neck (along sternal border in infants) | Very common decreases with age | N or | May be in adults | As in acquired a.s. |
| Congenital subvalvar | Discrete like valvular Tunnel Lt. sternal border | Rare | N, or ab | Almost all | Do |

| | | | | | |
|---|---|---|---|---|---|
| Congenital supravalvar | Rt. sternal border to neck, occ. greater in neck. | Rare | N or arm pulse | Uncommon | Rapid upstroke in Rt carotid, delayed in 1t. carotid. Rt. pressure > Lt arm P.P. |

**Table 5.8: Effect of Interventions in Systolic Murmurs**

## Complications

**Table 5.9: Mechanisms of Ischaemia in HOCM**

| | | | |
|---|---|---|---|
| 1 | Myocardial $O_2$ Demand | 1 | Small vessel Disease |
| 2 | Diastolic Dysfunction | 2 | Abnormal vascular response |
| 3 | Myocyte Disarray | 3 | Mocardial Bridges |
| 4 | LV outflow obstruction | 4 | Coronary vascular Resistance |
| 5 | Arrhythmias | 5 | - |

## Complication

The clinical course is variable. The annual mortality is closer to 1 per cent. The risk of sudden death is higher in children and young adults and is close to 6 per cent. Development of atrial fibrillation will increase symptoms suddenly. The complications thus are: Progressive increase in hypertrophy, development of outflow obstruction, ischaemia arrhythmias and sudden death (Table 5.10, 5.11) and endstage disease with progressive dilation and CCF (5-10 per cent) (Fig. 5.8).

**Table 5.10: Adverse outcome in HOCM**

| | |
|---|---|
| 1 | H/O sudden death/syncope |
| 2 | H/O sudden death in family members |
| 3 | Severe LVH > 33 mm |
| 4 | Extent of Myocardial disarray |
| 5 | Extent of Interstitial Fibrosis |
| 6 | Early onset of disease |

7 Myocardial Ischaemia on perfusion tomography

8 Abnormal B.P. response to exercise (Failure to rise or fall of B.P.)

9 LV outflow obstruction more than 30 mmHg at rest.

10 VT/NSVT on Holter

11 “Malignant” causal mutations

12 “Malignant” modifier genes

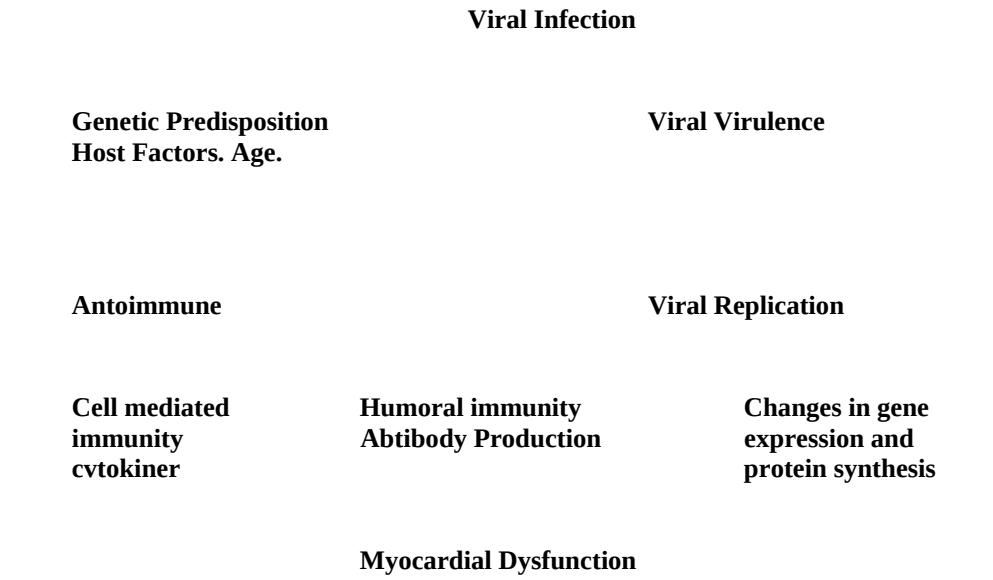
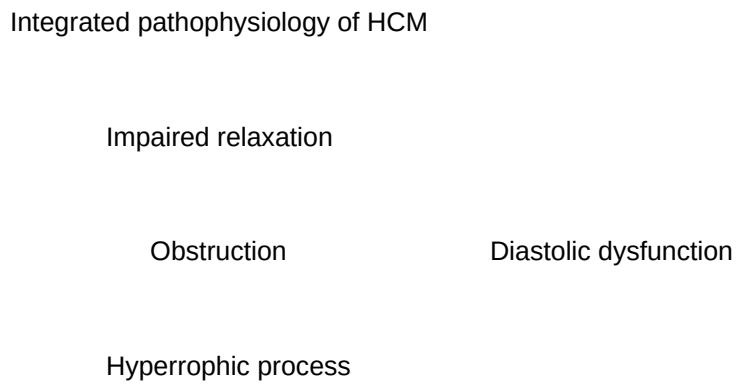



**Fig. 5.11: Development of myocardial dysfunction in viral infection**



| | | |
|---|---|---|
| Heart block | Malignant family history (molecular genetic defect) | Neurocardiogenic mechanisms |

**Fig. 5.8: Integrated pathophysiology of HOCM**

Marked LVH

Ischemia

Cell Death

Replacement Fibrosis

Cavity enlargement Wall thining

LV Remodelling

Systolic dysfunction

Progressive CHF

**Fig. 5.9: Progression to endstage phase in HOCM**

## Management

It is directed towards amelioration of symptoms, prevention of complications including sudden death. Risk assessment stratification should be done in all and at various stages. (Fig. 5.10).

a) ***Asymptomatic/Mildly Symptomatic Patients***

The treatment is controversial. Some give them betablockers/or calcium channel blockers like verapamil, in the hope of preventing progression of the disease. However there is no evidence for the same. Amiodarone is given, if Holter shows episodes of nonsustained ventricular tachycardia.

b) ***Moderate/Severe Symptoms***

i) *Medical Management*

β-blocking drugs or verapamil are useful. Beta-blocker blunt heart's chronotropic response, thus limiting oxygen demand and improving diastolic dysfunction. 1/3 - 2/3 rd of patients have symptomatic improvement. Ca channel blockers are useful because both the hyper contractile systolic function and abnormalities of diastolic filling may be related to abnormal Ca kinetics. They block inward transport of Ca across myocardial cell. Verapamil improves exercise performance better than β-blockers. In non-obstructive patients,usually verapamil is preferred because of its greater effect on diastolic dysfunction. When either of them is not effective, a trial of disopyramide may be useful. Disopyramide is useful as it alters Ca kinetics. It is particularly useful with beta-blocker in reducing outflow gradient.

ii) *Interventional Treatment*

This is usually reserved for severely symptomatic patients with obstruction who do not respond to medical treatment. It is performed to relieve the sub aortic obstruction and normalize markedly increased LV systolic pressure. The indications are:

— refractory to standard medical treatment and

— gradient of at least 50mmHg across LV outflow tract, at rest or on provocation.

**DDD Pacing**

Although true benefit is uncertain, it is used as an appropriate medical therapy in persistently symptomatic patients, viz.

i) in whom there is an independent need for permanent pacing

ii) Severe bradycardia due to b-Blockers

iii) contraindications to surgery/septal ablation.

**Septal Ablation (TASH/PTSMA)**

This is a nonsurgical interventional treatment. The septal myocardium supplied by 1st septal branch of left anterior descending artery is destroyed by alcohol, by percutaneous technique. It creates limited myocardial infarction and reduces LV outflow obstruction. It reduces mitral incompetence and improves relaxation. Improvement occurs in almost 90 per cent.

However there is some concern about 2 complications viz. Heart block which needs pacing in a few patients. There is a theoretical concern that myocardial scar that follows infarction might lead to ventricular arrhythmias. It is therefore a preferred option in patients who already have a pacemaker.

ICD in place or where surgery is contraindicated because of other medical conditions. It is however being widely used in select centres.

**ICD Implantation**

It is used as a method of secondary prevention for cardiac arrest or history of sustained hemodynamically unstable ventricular tachycardia. For primary prevention, it is used with history of sudden death in family with history of sustained ventricular tachycardia.

**Surgical**

Septal Myotomy/Myectomy (Fig 5.8) it is widely used. About 5 gms. of hypertrophied septum are removed using a tranaortic approach (Morrow procedure). It gives sysmptomatic relief in almost 70-90 per cent of patients. It should be preferred in younger patients and in whom septal ablation has failed. Occasionally mitral valve replacement or suture plication of anterior mitral leaflet are also employed.

iii) *Endstage Disease*

The treatment is of cardiac failure, including cardiac transplant.

**Check Your Progress 4**

1) What are some of the features of hypertrophic cardiomyopathy?

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2) What are the features of hypertrophic obstructive cardiomyopathy on cardiac examiniation?

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3) Whar are some of the ECG changes in hypertrophic cardiomyopathy?

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4) What are some of the echocardiographic features of hypertrophic obstructive cardiomyopathy?

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5) What are some of the drugs that can be used in the management of hypertrophic cardiomyopathy?

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## 5.5 MYOCARDITIS

Myocarditis is defined as inflammation of the myocardium.

The most common cause is Coxsackie B virus infection. But it can also be due to other viruses (e.g. HIV, cytomegalo virus), bacterial, protozoal, metazoal and fungal infections. Rarely it could be due to hypersensitivity, or a toxic agent.

Few other specific causes are:

i) Rheumatic Carditis. (See section on rheumatic fever)

ii) Chagas Disease. It is seen in central and south America and is due to infection with trypanosome cruzi.

iii) Toxoplasmosis and cytomegalo virus myocarditis are usually seen in cardiac transplant recipients.

iv) Lyme disease (due to spirochete), eosinophilic myocarditis, giant cell myocarditis are other entities.

**Pathology**

Infection usually viral is responsible for cardiac injury. The infective agents induce adverse immunologic responses that persist despite eradication of the infective agent. (Fig. 5.11).

The heart usually shows foci of active inflammation with necrosis. In toxoplasmosis, toxoplasma cysts are seen in myocytes. In eosinophilic and giant cell myocarditis, the predominant cells infiltrating myocardial interstitium are eosinophils and giant cells respectively.

**Clinical Menifestation**

There is H/O antecedent viral syndromes with flu like symptoms Patient may be totally asymptomatic and may have no signs.

The clinical symptoms and signs are due to:

i) depressed LV function (both systolic and diastolic) viz. dyspnoea, cardiac failure and S3.

ii) arrhythmias-usually ventricular viz. palpitations. Syncope and sudden death is known to occur.

iii) Heart blocks, may occur. Sometimes patient may present as dilated cordiomyopathy with failure. Patients with peripartum cardiomyopathy have high frequency of myocarditis. The severity of myocarditis is judged by degree of LV dysfunction, presence of cardiac failure, arrhythmias and heart block. Clinically, a classification of primary myocarditis is proposed viz. Fulminant, subacute, chronic active and chronic persistent (Table 5.11).

**Table 5.11: Pathologic Classification of Myocarditis**

| | | Fulminant | Subacute | Chronic | Chronic |
|---|---|---|---|---|---|
| 1 | Onset | Distinct | Indistinct | Indistinct | Indistinct |
| 2 | LV | Severe dysfunction | Moderate | Moderate | Nil |
| 3 | Biopsy | Multiple Foci | Active or Borderline | Active or Borderline | Active or Borderline |
| 4 | Clinical History | Recovery/ Death | Incomplete dilated cardiomyopathy | Restrictive CM | Normal |
| 5 | Histological outcome | Complete Resoluation | Complete Resolution | Giant cell fibrosis | Ongoing |

Blood studies are nonspecific. There may be rise in ESR, CPK-MB. There may be abnormalities in T and B-lymphocyte counts CD 4/ CD8 ratios may be abnormal. Elevated IgM antibody titer to enterovirus may suggest previous infection.

**X-Ray Chest**

May show cardiomegaly and signs of pulmonary venous congestion.

**Electrocardiography**

Sinus tachycardia, diffuse ST.T. changes and prolonged QTc suggest myocarditis in setting of viral infection. Occasionally pattern of myocardial infarction is seen. LBBB and complete heart block can occur, but they are usually transient both supraventricular and ventricular arrhythmias are seen. The former occurs usually in presence of cardiac failure. The latter at times may be the

only manifestation of myocarditis. Holter shows episodes of unsustained ventricular tachycardia in almost 25 per cent of cases even with mild symptoms.

**Echocardiography**

It will usually show LV systolic dysfunction. Rarely segmental wall abnormalities may occur. LV size is normal or only slightly dilated Ventricular thrombi are seen in 15 per cent. Cardiac catheterization and hemodynamic studies are rarely done. The important diagnostic tool is endomyocardial biopsy. The diagnosis is based on Dallas criteria which define active myocarditis, as presence of inflammatory infiltrate and areas of necrosis (not typical of ischaemic changes). Imaging with radioisotopes which are inflammation avid e.g. gallium 67 or iridium III-antimyosin monoclonal antibody and MRI imaging are promising tools for the future.

**Diagnosis**

The diagnosis of myocarditis should be suspected under following circumstances.

1) In the setting of antecedent viral infection, diffuse ST.T Changes on EKG and/or presence of LV dysfunction on clinical examination or echo or cardiac failure.
2) In cardiac failure of no obvious etiology, if heart size is normal or only slightly dilated, in the young population.
3) Peripartum cardiomyopathy. At least 20 per cent are due to myocarditis.
4) Ventricular tachyarrhythmia in absence of any obvious cause. This needs to be confirmed by endomyocardial biopsy.
5) Sudden death in absence of any other cause.

**Prognosis**

1) Vast majority may remain asymptomatic at onset and have no residual evidence of LV dysfunction.
2) About 1/3rd of patients recover completely.
3) Severe forms of myocarditis may lead to dilated cardiomyopathy. However the frequency of progression is unknown. Sudden death is a rare event.

**Management**

***Specific Therapy***

A number of uncontrolled and nonrandomized studies suggested usefulness of immunosuppressant agents viz. prednisolone and azathioprine or more aggressively with interferon and anti-CD3. They remain unproven on multicentric, randomized trials. Similarly immune modulatory therapy with a single infusion of high dose immunoglobulin (29mg/kg) has shown no benefit.

***Symptomatic Therapy***

This includes treatment of LV dysfunction and cardiac failure. Ventricular arrhythmias need to be treated with amiodarone and complete heart block with temporary pacing. Permanent pacemaker, AICD implantation or cardiac transplant are not indicated in acute phase. All need anticoagulant therapy in view ventricular thrombi.

## 5.6 LET US SUM UP

Cardiomyopathy is a primary disorder associated with unknown cause of heart muscle that there are three types of cardiomyopathy i.e. dilated cardiomyopathy, restrictive cardiomyopathy and hypertrophic cardiomyopathy.

Management of dilated cardiomyopathy is satisfactory if it is started in the asymptomatic stage and if the causes of the cardiomyopathy is reversible cause.

Treatment of restrictive cardiomyopathy is not satisfactory. Nature of the treatment and treatment of hypertrophic cardiomyopathy is controversial. Myocardities is mainly cause by coxackie birus and about one third of the patient recover completely.

## 5.7 ANSWERS TO CHECK YOUR PROGRESS

**Check Your Progress 1**

Cardiomyopathy is a primary disorder of heart muscle that may cause cardiac dysfunction and is not related to any obvious disease process. It is heart muscle disease of unknown cause or association.

**Check Your Progress 2**

1) The ventricles and atria are dilated without much hypertrophy and the ejection fraction is depressed.

2) A 3rd heart sound is common with accentuation of the pulmonary component of the 2nd sound from pulmonary hypertension.The presence of LBBB or RBBB can alter the second sound.

   If mitral and tricuspid regurgitation are present then the corresponding murmurs may be heard.

3) Left and biventricular failure, hypotension. Arrhythmias (both atrial and ventricular) and embolic episodes from LV thrombi

**Check Your Progress 3**

The heart is only minimally dilated and there is no thickening of the myocardium in idiopathic causes, but may be thickened in infiltrative disorders. Both atria are enlarged.

**Check Your Progress 4**

1) It is a genetic disorder due to mutations in the gene that encodes for β-Cardiac myosin heavy chain (Localised to chromosome 14). It is characterized by inappropriate and often massive myocardial hypertrophy There is myocardial disarray and interstitial fibrosis. It has asymmetric septal hypertrophy, left ventricular outflow obstruction due to systolic anterior motion of mitral valve, LV diastolic dysfunction, myocardial ischaemia and arrhythmias. LV outflow obstruction occurs only in about ¼th.

2) Usually the apex is palpable and will be LV type and may have double apical impulse due to powerful LA contraction. Prominent S4 and systolic murmur at lower sternal border are heard. The murmur is due to labile LV outflow obstruction and it increases with Valsalva manoeuvere, standing up from squatting position, exercise and in postextrasystolic contraction. The SM is harsh and crescendo-decrescendo in configuration. It commences sometime after S1 and is best heard between apex and left sternal border. It radiates to lower sternal border, axilla and base of the heart, but never to carotids. MR murmur may also be present. Brisk carotid pulse and sometimes prominent ‘a’ in JVP are seen.

3) The abnormalities are seen in QRS, ST Changes and T-Waves. LV voltage is increased. Abnormal Q are seen in 25-50 per cent of patients, usually in inferolateral leads and may mimic myocardial infarction. Early repolarisation and other ST changes may be seen. Giant negative T-waves are seen in apical cardiomyopathy. LA is enlarged, Arrhythmias (NSVT 25 per cent, PSVT 35-50 per cent, af 5 per cent) are seen. In 5 per cent WPW may be seen.

4) It is the most important diagnostic tool. There is asymmetric septal hypertrophy as seen by septal to posterior wall thickness ratio of 1.5 or more. Maximal septal hypertrophy occurs midway between apex and base. Apical hypertrophy, midventricular hypertrophy or RV hypertrophy may be seen. LV thickness can vary from 15mm to 60 mm. Systolic anterior motion of the mitral valve identifies LV outflow tract obstruction. There is also elongation and enlargement of mitral valve leaflets, leading to abnormal aortic outflow geometry. It contributes to pressure gradient across LV outflow and is also responsible for mitral regurgitation. There are other echocardiographic findings viz. small LV cavity, partial systolic closure and coarse fluttering of aortic valve reduced septal and exaggerated posterior wall motion.

5) Beta blockers

Calcium channel blockers like Verapamil

Disopyramide.