بسم الله الرحمن الرحيم

CARDIOMYOPATHIES

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The definition of *cardiomyopathies* used by the AHA expert consensus panel in its 2006 document entitled "Contemporary Definition and Classification of the Cardiomyopathies" reads as follows:

> *Cardiomyopathies are a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilation and are due to a variety of causes that frequently are genetic. Cardiomyopathies either are confined to the heart or are part of generalized systemic disorders, often leading to cardiovascular death or progressive heart failure-related disability.*

CLASSIFICATION

1. Primary
(confined to heart muscle)

2. Secondary
(Multi organ disorder)

CLASSIFICATION OF PRIMARY CARDIOMYOPATHIES

TABLE 10.5 Classification of Primary Cardiomyopathies

Genetic	Hypertrophic cardiomyopathy
	Dysrhythmogenic right ventricular cardiomyopathy
	Left ventricular noncompaction
	Glycogen storage disease
	Conduction system disease (Lenègre disease)
	Ion channelopathies: long QT syndrome, Brugada syndrome, short QT syndrome
Mixed	Dilated cardiomyopathy
	Primary restrictive nonhypertrophic cardiomyopathy
Acquired	Myocarditis (inflammatory cardiomyopathy): viral, bacterial, rickettsial, fungal, parasitic (Chagas disease)
	Stress cardiomyopathy
	Peripartum cardiomyopathy

CLASSIFICATION OF SECONDARY CARDIOMYOPATHIES

TABLE 10.6 Classification of Secondary Cardiomyopathies

Infiltrative	Amyloidosis
	Gaucher disease
	Hunter syndrome
Storage	Hemochromatosis
	Glycogen storage disease
	Niemann-Pick disease
Toxic	Drugs: cocaine, alcohol
	Chemotherapy drugs: doxorubicin, daunorubicin, cyclophosphamide
	Heavy metals: lead, mercury
	Radiation therapy
Inflammatory	Sarcoidosis
Endomyocardial	Hypereosinophilic (Löffler) syndrome
	Endomyocardial fibrosis
Endocrine	Diabetes mellitus
	Hyperthyroidism or hypothyroidism
	Pheochromocytoma
	Acromegaly
Neuromuscular	Duchenne-Becker dystrophy
	Neurofibromatosis
	Tuberous sclerosis
Autoimmune	Lupus erythematosus
	Rheumatoid arthritis
	Scleroderma
	Dermatomyositis
	Polyarteritis nodosa

HYPERTROPHIC CARDIOMYOPATHY

1. Prevalence 1 in 500 persons
2. The most common genetic C.V. dis.
3. A.D. trait e´ variable penetrance
4. L.V. hypertrophy in the absence of any other cardiac Dis. (HTN, A.S.)
5. Often hypertrophy confined to septum and anterolateral free wall L.V.
6. In other forms H.C.M can be concentric hypertrophy, or biventricular or only L.V. free wall or apex
7. Histologically: hypertrophied myocardial cells and areas of patchy myocardial scarring

Pathophysiology of H.C.M.

I. Myocardial hypertrophy

II. Dynamic LVOT obstruction

III. S.A.M. → M.R.

IV. Diastolic dysfunction

V. Myocardial ischemia

VI. Myocardial dysrhythmia (cause of sudden death in youngs)

SYSTOLIC ANTERIOR MOTION

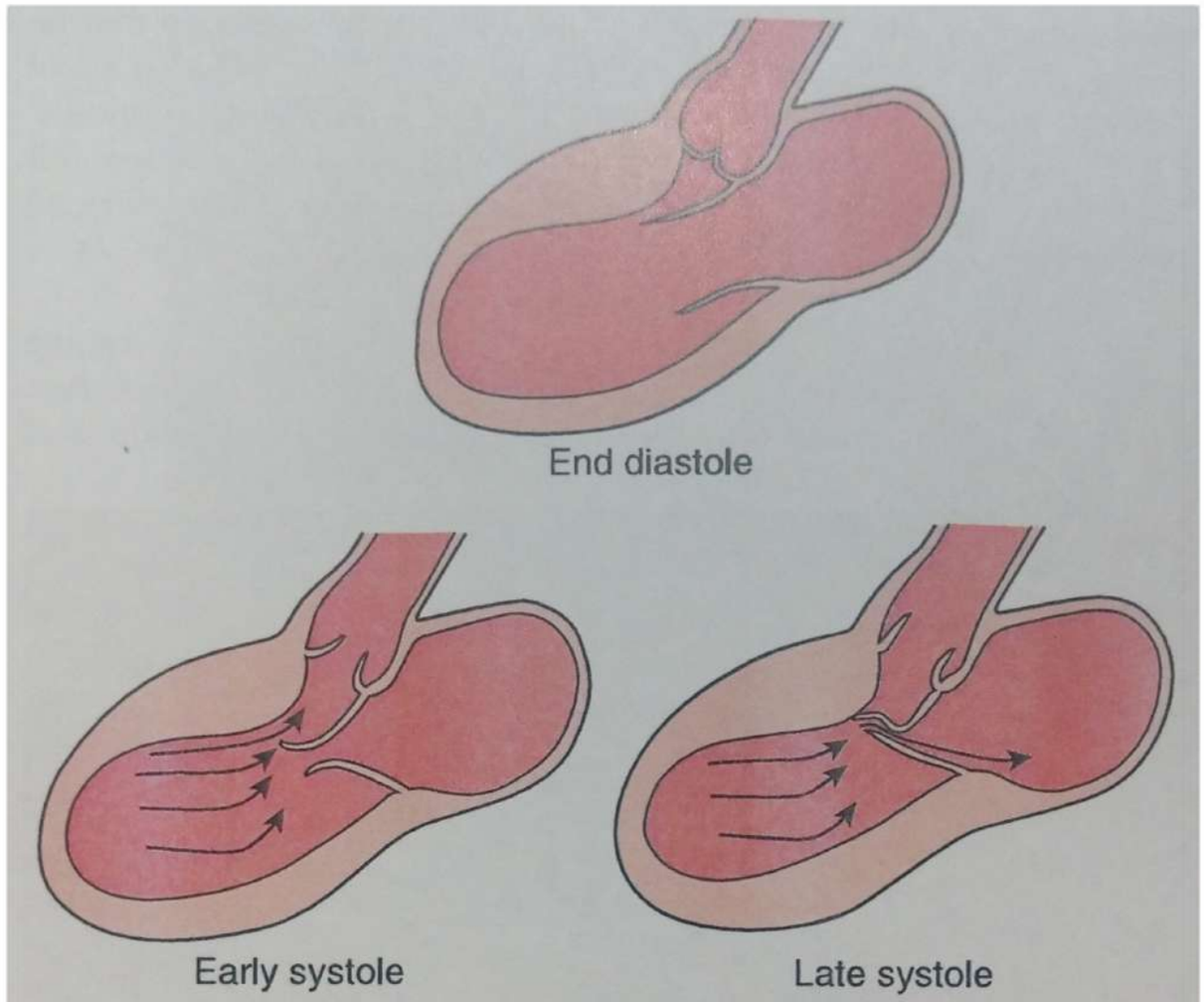

Fig. 24.37 **Mechanisms of left ventricular outflow tract (LVOT) obstruction in hypertrophic cardiomyopathy.** The systolic anterior motion (SAM) of the anterior leaflet of the mitral valve begins early in systole, often during isovolemic left ventricular contraction (not depicted) when Venturi effects are negligible. As the aortic valve opens and the ejection phase of systole proceeds *(lower left and right images)*, the anterior mitral valve leaflet is both pushed and pulled into the LVOT. In addition, the jet of mitral regurgitation associated with SAM is classically directed posterolaterally (see text for details). *(Reproduced with permission from Oliver WC, Mauermann WJ, Nuttall GA. Uncommon cardiac diseases. In: Kaplan JA, Reich DL, Savino JS, eds.* Kaplan's Cardiac Anesthesia: The Echo Era. *6th ed. Philadelphia: Saunders; 675–736; and from Ommen SR, Shah PM, Tajik AJ. Left ventricular outflow tract obstruction in hypertrophic cardiomyopathy: past, present and future.* Heart. *2008;94:1276–1281, 2008.)*

TABLE 10.7 Factors Influencing Left Ventricular Outflow Tract Obstruction in Patients With Hypertrophic Cardiomyopathy

EVENTS THAT INCREASE OUTFLOW OBSTRUCTION

Increased myocardial contractility
- β-Adrenergic stimulation (catecholamines)
- Digitalis

Decreased preload
- Hypovolemia
- Vasodilators
- Tachycardia
- Positive pressure ventilation

Decreased afterload
- Hypotension
- Vasodilators

EVENTS THAT DECREASE OUTFLOW OBSTRUCTION

Decreased myocardial contractility
- β-Adrenergic blockade
- Volatile anesthetics
- Calcium entry blockers

Increased preload
- Hypervolemia
- Bradycardia

Increased afterload
- Hypertension
- α-Adrenergic stimulation

Based on obstruction pattern

1. Non obstructive< P.P. gradient <30 mmHg
2. Obstructive > 30 mmHg
3. Latent (exercise - induced press. gradient > 3ommHg)

Signs and Symptoms

1. Asymptomatic to → sudden death
2. Angina pectoris - fatigue - tachy dysrhythmias - syncope (aborted S.D.) - heart failure
3. Lying down often relives angina pectoris of H.C.M. (enlargement L.V. size)
4. In physical exam → double apical impulse - gallop rhythm, murmur and thrill
5. valsalva manuver → ↑LOVT obs.↑ → systolic ↑murmur
   → MR ↑
6. TNG and standing → syst. murmur ↑
7. Sudden death → is a major complication in HCM
8. Severity of hypertrophy ↑ → risk of S.D. ↑
9. death occurs in 10-30 yrs

Diagnosis

1. EKG shows L.V. hypertrophy
2. High QRS voltage - ST - T alteration
3. Q wave similar to old MI
4. Left Atrial enlargement
5. In echo : Myocardial hypertrophy - assess S.A.M. - revealing LVOT Obs. and MR (by turbulent out flow) - measuring , gradient LVOT Obs. - Evaluating Dias. function
6. Catheterization : direct measurement LVEDP and LOVT and aorta pressure
7. Definitive diagnosis of HCM is by endomyocardial biopsy and DNA analysis

Treatment

1. diverse features make it impossible to define precise guid line for therapy
2. Sudden death is a major problem
3. pharmacologic therapy : diastolic filling ↑ , LOVT obs. and myocardial ischemia ↓
4. Surgical therapy in 5% patients: removal of hypertrophy cause LVOT obs.↓ and severe↓ symptoms unresponsive to medical therapy

Medical Therapy

1. B-Blockers (HR ↓ dias ↑ time - myocardial ↓ requirement - sympathetic activity ↓ LOVT ↓ obs.

2. Ca channel Blockers (Verapamil - Diltiazem) → Vent ↑ filling , MYO. ↓ Ischemia

3. Diuretics

4. Amiodarone (esp. in sudden death risk)

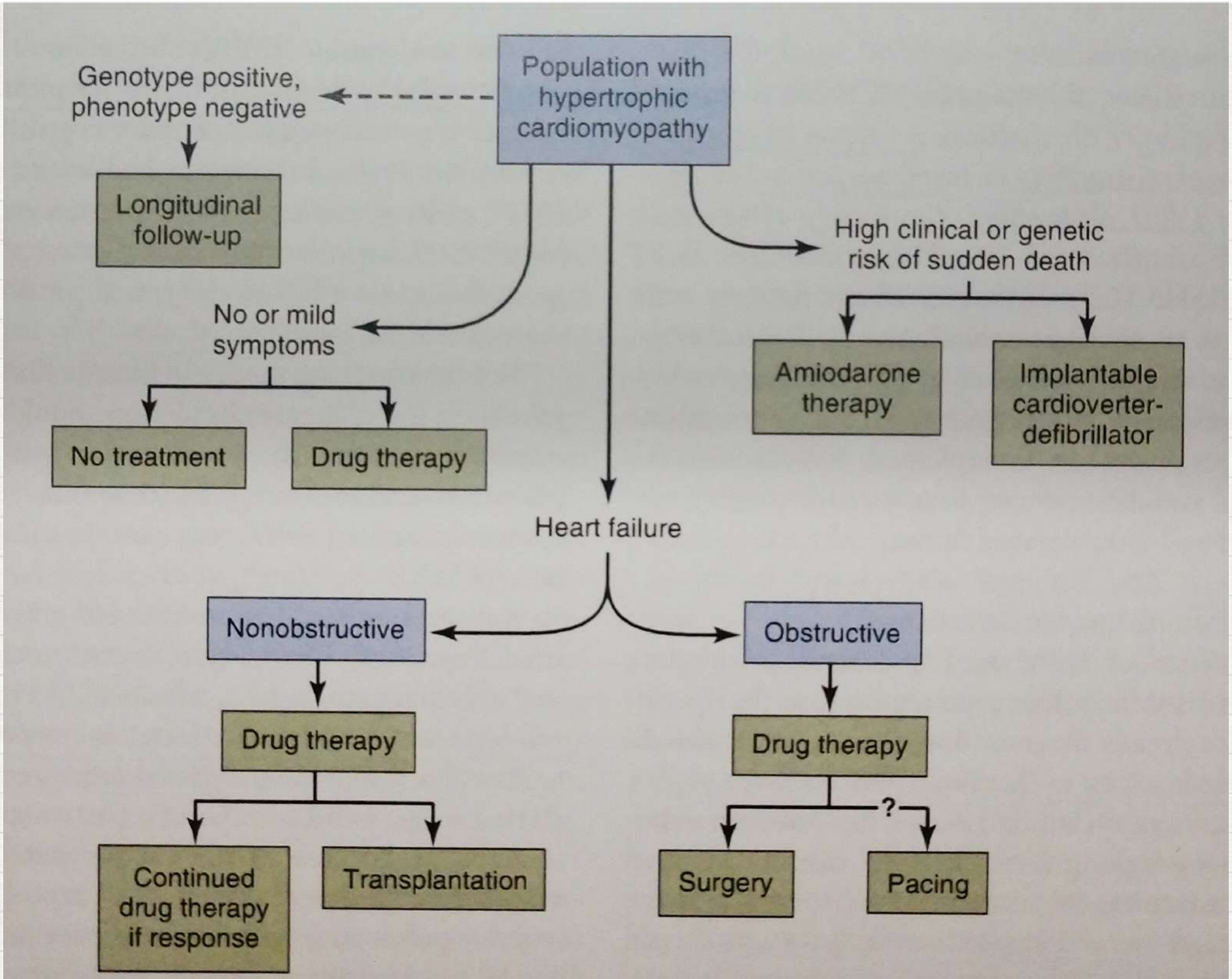

FIG. 10.9 Clinical presentations of hypertrophic cardiomyopathy and corresponding treatment strategies. (Adapted from Spirito P, Seidman CE, McKenna WJ, et al. The management of hypertrophic cardiomyopathy. *N Engl J Med*. 1997;336:775-785. Copyright 1997 Massachusetts Medical Society.)

Surgical therapy

1. In patient who have both LVOT gradient > 50mmHg and gross CHF despite medical therapy are select for surgery

2. Septal myomectomy or alcohol injection into septal perforator arteries → septum necrosis → LOVT obs.

3. If patient remain symptomatic → prosthetic mitral valve replacement → counteract S.A.M.

Prognosis

1. overal mortality (annually) is 1%
2. In history of S.D. and malignant - vent. dysrhythmia → 5%

Management of Anesthesia in H.C.M.

1. Main aim is LVOT Ob ↓
2. Contractility ↓ preload ↑ and after load ↑ → LOVT OB ↓
3. Sympathetic stimulation , hypovolemia and vasodilation → worsen LVOT Obs.
4. Unrecognized HCM may manifest intraoperatively as hypotension (unexplained) or systolic murmur (in response to hemorrhagie or vasodilation)

Key Points in H.C.M. Operations

1. Update EKG and Echo evaluation before elective surgeries
2. Patient with I.C.D. : ICD must be turned off
3. Anxiolysis should be administered (Relief sympathetic out flow)
4. Volume expansion is useful in LVOt ob ↓ and P.P.V.
5. Anesthesiologist be aware of risks any regional or general anesthesia can be selected
6. Before intubation å ß blocker or a volatile agent can be used for blunting sympathetic N.S.
7. PPV (with TV ↓ - RR ↑) and avoidance of PEEP
8. In laparoscopy → insufflation must be slow
9. Anesthetic agent with mild myocardial depression and minimal effects on preload and after load such as a volatile in mod. dose
10. hypotension should be treated by an å adrenergic agonist (phenylephrine)

Key Points in H.C.M. Operations

11. Ephedrine - Dopamine - Dobutamine are contraindicated (because LVOT↑ obs)
12. Prompt replacement of blood loss and careful titration of IV fluids is important for maintaining preload and blood pressure
13. Because of Dias. dysfunction aggressive fluid administration → pul. edema
14. Vasodilators not to use to lower BPB → because LVOT obs↑
15. Maintenance of normal sinus rhythm is very important
16. Intraoperative PSVT need immediate pharmacologic or electrical cardioversion
17. Cardioverter - defibrillator must be available
18. Metoprolol and Esmolol → slow persitently HR

HCM in Parturient patients

1. Inspite of S.V.R. ↓ and V.R. ↓ (Aortocaval compression)
2. Labor pain , bearing down (valsalva M.) → LVOT ↑ obs. (Cathecholamine release)
3. Regional Anesthesia is safe if euvolemia or slight hypervolemia administered
4. If hypotension → fluid therapy → phenylephrine
5. Oxytocin must be administered carefully (vasodilation and compensatory tachycardia) and abrupt inflow of large fluid into central circulation (uterine contraction)

HCM in Parturient patients

6. Pulmonary edema may occur after delivery treatment include → phenylephrine - esmolol → LVOT ↑ obs.
7. Diuretics and nitrates cannot be used to treat P.E. → hypovolemia , SVR↓ → LVOT ↓ obs.
8. HCM parturients must be carefully monitored in ICU and recovery room and any factor that stimulate sympathetic nervous system (pain - shivering - anxiety - hypoxia , hypercarbia) must be eliminated.
9. Maintenance of euvolemia and prompt that treatment of hypotension are crucial.

Dilated cardiomyopathy (D.C.M.)

I. Is a primary myocardial Dis. (LV or biventricular dilation - sys. dysfunction and normal ventricular thickness)

II. Ethiology unknown (genetic or infection)

III. Many type of secondary C.M.P. are dilated (alcohol abuse - cocaine- peripartum , pheochromocytoma , infectious such as HIV , uncontrolled tachycardia , duchenne , HTN , CAD, valvular H.D. , thyroid Dis. , chemotherapeutic drugs - radiation)

IV. Is the most common type C.M.P. and th 3th most common cause of H.F. and the most common indication for cardia transplantation

Signs and Symptoms of D.C.M.

1. Initial manifestation is usually H.F.

2. Chest pain in some patients

3. functional M.R. or T.R. may occur

4. VT , PSVT, conduction abnormality and sudden death are common

5. Systemic embolization from thrombi in hypokinetic cardiac chamber is common

Diagnosis of D.C.M.

1. ST-T , abnormalities and LBBB , PVC , AF
2. CXR : 4 chambers enlargement , LV dilation is principle feature
3. Echo : Dilation of 4 CHAMBERS es. L.V. - global hypokinesia - regional wall motion abnormalities may be seen - mural thrombi - valvular regurgitation (annular dilation)
4. Other causes should be ruled out
5. Right heart catheterization : PCWP ↑ , SVR ↑ , Co ↓
6. Endomyocardial biopsy is not recommended

Treatment of D.C.M.

1. Treatment includes general supportive measures such as (adequate rest - weight control - low Na diet - fluid restriction - abstinence from tobacco and alcohol - decrease physical activity during decompensation)
2. Cardiac rehabilitation if possible
3. Treatment similar to C.H.F.
4. Embolic ↑ events because hypo contractile cardiac chambers
5. Anticoagulation is needed (warfarin - antixa)
6. V.T. is common → if medical treatment failed → I.C.D.
7. Heart transplant (main indication in adult - children) is beneficial if max. medical treatment not respond in active under 60 yrs patients.

Prognosis of D.C.M.

1. 5 yrs survival in symptomatic D.C.M. is 50%

2. If biventricular → prognosis is even worse

3. Some of hemodynamic abnormalities predict poor prognosis :
EF<25% - PCWP> 20 - CI < 2.5 lit/min/m - syst. hypotension - pul. hypertension - CVP ↑

4. If complete abstinence from alcohol is maintained → alcohol D.C.M. is largely reversed

Management of anesthesia in D.C.M.

1. Similar to C.H.F.

2. Regional anesthesia may be an alternative but anticoagulant therapy may limit this option

Apical ballooning syndrome

1. Stress - include C.M.P. (broken heart synd.) or (takotsubo C.M.P.)
2. Characterized by temporary apical hypokinesia with Ischemic EKG changes (unobstructed coronary A.)
3. Rest of heart is normal
4. The most common symptoms : chest pain - dyspnea
5. The main factor is stress (physical or emotional)
6. Women > men are effected
7. Pheochromocytoma and myocarditis must be ruled out
8. High catecholamine state (isotopes should be avoided)
9. ß-Blocker and Ca-channel blockers and IABP is useful
10. Prognosis is good (in 2 month)

Permpartum C.M.P.

1. A rare form of D.C.M. (3th trimester until 5months after delivery)
2. It occur in women with no history of heart dis.
3. Incidence is 1 to 3000-4000 parturients (blacks ↑)
4. May be related to diet and lifestyle
5. Risk factors : HTN - obesity - prior toxin exposure (cocaine) - multiparity - age>30 yrs - multifetal pregnancy - pre eclampsia - long-term oral tocolytic therapy - african american
6. Viral myocarditis - abnormal immune response to pregnancy and maladaptive response to the hemodynamic stress of pregnancy

Signs and symptoms of peripatum D.C.M.

1. Similar to CHF

2. Dyspnea - fatigue - peripheral edema

3. There are no specific criteria

4. Amniotic fluid or pul. emboli those mimic CHF should be excluded

Diagnosis of peripartum D.C.M.

1. Based on three criteria:
    - A. Development of H.F. in surrounding delivery
    - B. Absence of another explainable cause of H.F.
    - C. L.V. sys dysfunction e EF< 45%
2. EKG - BNP level - CXR - Echocardiography - cardiac MRI - cardiac catheterization and endomyocardial biopsy can assist

Treatment of parturient D.C.M.

1. Goal is to alleviate the symptoms of H.F.
2. Diuretics and vasodilators can be used
3. ACE inhibitors are teratogenic , Hydralazine and nitrates are recommended
4. IV IG may have a beneficial effect
5. Thromboemboli is common and anticoagulation is often recommended
6. Mechanical circulatory support or transplant if medical therapy failed

Prognosis in parturient D.C.M.

1. Mortality rate ranges from 25% - 50%
2. Mortality rate ↑ in african american
3. Most death occur within 3 months of delivery
4. Death is usually a result of progression of H.F. - sudden death related to dysrhythmias or thromboembolic events
5. Prognosis depend on : The degree of normalization of LV size and function within 6 months of delivery

Management of anesthesia in parturient D.C.M.

1. Require a) assessment of cardiac status and b) careful planning of analgesia and or anesthesia required for delivery

2. Regional anesthesia may provide a desirable afterload reduction

Secondary CMP with restrictive Physiology

1. Is due to systemic Dis → myocardial infiltration → severe dias. dysfunction
2. Amyloidosis is the most common cause
3. Hemochromatosis , sarcoidosis and carcinoid may → similar C.M.P.
4. H.F. witout cardiomegaly and without sys. dysfunction
5. Deposition of abnormal substances → stiffness of myocardium
6. Must be differentiated from constrictive pericarditis

Signs and symptoms RCMP

1. Biventricular failure symptoms and signs may present

2. Amyloid CMP often present with thromboembolic events

3. Atrial fibrillation is common - Arrhythmia also occur → progression involvement conduction system → heart block or vent. arrhythmias → sudden death

Diagnosis

1. EKG: conduction abnormalities
2. CXR: PUL. congestion and/or pleural effusion, but cardiomegaly is absent
3. Lab. tests should be used for diagnosis of systemic dis.
4. Echo: diastolic dysfunction - normal systolic function - enlargement of atrium but normal ventricle
5. Endomyocardila biopsy can help the cause of infiltrative C.M.P.

Treatment Of Resrictive C.M.P.

1. Symptomatic treatment similar to dias. heart failure
2. Administration of diuretics to treat pul, and sys, congestion
3. Maintenance of artrial Kick (normal sinus rhythm)
4. Bradycardia → acute H.F. (because S.V. is fix)
5. Cardiac block or bradycardia → need to pacemaker
6. In sarcoidosis → VT ↑ → ICD is needed
7. Anticoagulation is needed in low C.O. ↓ and A.F.
8. Cardiac transplantation is not a choice

PROGNOSIS AND TREATMENT IN RESTRICTIVE C.M.P.

1. Prognosis is very poor
2. Management of anesthesia similar to tamponade
3. Because S.V. is fixed → maintaining sinus normal rhythm + avoid bradycardia
4. Maintenance of venous return and intravascular fluid volume are necessary for an acceptable C.O.

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THE END

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