Cardiomyopathies

Cardiomyopathies are defined as diseases of the myocardium associated with cardiac dysfunction. Common to all cardiomyopathies is progressive, life-threatening congestive heart failure. New AHA guidelines classify CMs as either primary (confined to heart muscle; genetic, mixed or acquired) or secondary (multiorgan involvement). Predominant primary CMs include Hypertrophic Cardiomyopathy (HCM) and Dilated Cardiomyopathy (DCM). Secondary CMs include inflammatory and infiltrative disorders (among many others) and these often have a restrictive cardiomyopathy (RCM) component.

GENERAL CONSIDERATIONS FOR ALL CARDIOMYOPATHIES:

- Assessment of underlying etiology
- Assessment of cardiac function
- Risk of hemodynamic compromise with anesthesia – recognize hemodynamic goals
- Risk of dysrhythmias – may have pacemaker/AICD
- Risk of thromboembolism – often anticoagulated
- Medications

DILATED CARDIOMYOPATHY:

1. Assessment of underlying etiology (idiopathic, viral, ischemic, alcoholic, peripartum)
2. Assessment of cardiac function (LV or biventricular dilation, ↓contractility, ↓CO, ↑filling pressures)
3. Risk of hemodynamic compromise with anesthesia – recognize hemodynamic goals:
   - Avoid myocardial depression
   - Maintain normovolemia
   - Avoid increased afterload
4. Risk of dysrhythmias – may have pacemaker/AICD
5. Risk of thromboembolism – often anticoagulated
6. Medications (ACE inhibitors, diuretics, digoxin)

RESTRICTIVE CARDIOMYOPATHY:

1. Assessment of underlying etiology (amyloidosis, sarcoidosis, idiopathic)
2. Assessment of cardiac function (biventricular diastolic dysfunction with normal systolic function)
3. Risk of hemodynamic compromise with anesthesia – recognize hemodynamic goals:
   - Maintain preload
   - Maintain normal sinus rhythm
   - Avoid bradycardia (because SV is relatively fixed)
4. Risk of dysrhythmias (A-fib) – may have pacemaker/AICD
5. Risk of thromboembolism – often anticoagulated
6. Medications (diuretics, etc.)

HYPERTROPHIC CARDIOMYOPATHY:

1. Assessment of underlying etiology (inherited, idiopathic)
2. Assessment of cardiac function (syncope, angina, dyspnea on exertion)
3. Risk of hemodynamic compromise with anesthesia – recognize hemodynamic goals:
   - Avoid tachycardia
   - Avoid increased contractility
   - Maintain high-normal preload & afterload
   - Maintain normal sinus rhythm
4. Risk of dysrhythmias (sudden death)– may have pacemaker/AICD
5. Risk of pulmonary embolism (2° to A-fib) – often anticoagulated
6. Medications (beta-blockers, verapamil)

	Dilated	Restrictive	Hypertrophic
General Issues	-Decreased contractile function, CHF, and dysrhythmias -LV +/- global dilation & dysfunction -Impaired contractility -↓CO, ↑filling pressures -Severe dilation may result in afib, MR/TR -Etiologies include ischemic, idiopathic, familial, toxic (EtOH, cocaine, adriamycin), peripartum	-Impaired diastolic filling, CHF, and a rate-dependent CO -Conduction defects -↓ diastolic filling of LV/RV, with preserved systolic function -↑ LV&RV pressures -Similar to constrictive pericarditis except cannot ↑ contractility and LV filling impairment is greater than that of RV	-Inherited autosomal dominant condition -Asymmetric LVH associated with dynamic LVOT obstruction between hypertrophied septum and systolic anterior motion of mitral valve leaflet -Diastolic dysfunction -Risk of ischemia, MR, and fatal dysrhythmias
Hx, Investigations	-Underlying etiology -Symptoms and functional capacity -Medications -Dysrhythmias -Anticoagulation and embolic complications -AICD? -Lytes, coagulation profile -CXR, ECG, Echo -Cath, Holter monitor	-Underlying etiology -Symptoms and functional capacity -Medications -Dysrhythmias -Anticoagulation and embolic complications -AICD? -Lytes, coags -CXR, ECG, Echo, Cath, Holter monitor	-Most asymptomatic -CHF, syncope, angina or cardiac arrest -Functional capacity -Medications (BB/CCBs should be continued throughout peri-op period) -Afib, Anticoagulation? -AICD? -Lytes, coags -CXR, ECG, Echo, Cath, Holter monitor; biopsy (reserved for inconclusive non-invasive testing)
Optimization	-Diuresis, ACEi, nitrates, hydralazine, antidysrhythmics	-Antidysrhythmic -Maintain volume	-Beta blockade/CCB or other rate control -Anxiolysis

	-Reverse anticoagulation -Pacemaker clinic re: biventricular pacing or AICD -Endocarditis prophylaxis	-Reverse anticoagulation -Pacemaker clinic re: biventricular pacing or AICD -Endocarditis prophylaxis	-Volume resuscitation -Endocarditis prophylaxis -Pacemaker/AICD considerations -Coagulation status
Anesthetic Goals	-Normal preload -Normal to slightly reduced afterload -Normal to slightly elevated rate -Sinus rhythm if possible -Increase contractility -Caution with neuraxial techniques and positive pressure ventilation -Keep pulmonary hypertension in mind	-Maintain preload -Normal afterload -*Elevated rate* (fixed stroke volume) -*Sinus rhythm* if possible -Increase contractility -Caution with neuraxial techniques and positive pressure ventilation -Keep pulmonary hypertension in mind	-"Splint" the ventricle open with slow rate, sinus rhythm (if possible), high-normal preload & afterload, avoid increased contractility -CHF may worsen with traditional Rx such as diuresis, nitroglycerin, and inotropes -Consider slowing rate with beta blockade instead and use direct α-agonist for hypoTN -Caution with neuraxial techniques and positive pressure ventilation -Keep pulmonary hypertension in mind -Good postop pain control to avoid high catecholamine state
Special Equipment and Monitors	-Pressors, inotropes, antidysrhythmics and defibrillator -IABP or ventricular assist device as appropriate to case -Invasive monitors and TEE guided by symptoms and surgery -Artline in most cases -Postop disposition guided by symptoms and surgery	-Pressors, inotropes, antidysrhythmics and defibrillator -Ventricular assist device as appropriate to case -Invasive monitors and TEE guided by symptoms and surgery -Artline in most cases -Postop disposition guided by symptoms and surgery	-Pressors, inotropes, antidysrhythmics and defibrillator -Phenylephrine (avoid ephedrine/beta-agonism) -Beta blockers -TEE, invasive monitors as appropriate to case -Artline in most cases -Postop disposition as appropriate to severity of symptoms and nature of surgery

	Hypertrophic	**Dilated**	**Restrictive**
Incidence	-1/500, but majority asymptomatic	-More common	-Less common
Etiology	-AD, but many cases idiopathic -Mutation in sarcomeric contractile protein genes	-Idiopathic, familial, toxic (adriamycin), ETOH, peripartum	-Idiopathic, *amyloidosis*, sarcoidosis
Pathophysiology	-LV (and/or right RV) hypertrophy, usually asymmetric & involves the interventricular septum and anterolateral free wall -Dynamic (Systolic) LVOT obstruction caused by systolic anterior motion of the anterior leaflet of the mitral valve against hypertrophied septum -Obstruction increased by ↓ LVESV: ↑contractility, ↓afterload, ↓ preload -Diastolic dysfunction due decreased LV compliance -Prone to ischemia due to hypertrophied ventricle (multifactorial) -At risk for ventricular arrhythmias/sudden death	-LV +/- RV dilation -Impaired contractility -↓CO, ↑filling pressures → CHF -Severe dilation may result in MR/TR	-↓ diastolic filling of LV/RV, systolic function preserved -↑ LV&RV pressures → CHF -Similar to constrictive pericarditis except: 1) cannot ↑ contractility 2) LV filling impairment > RV
History	-Dyspnea (most common), presyncope, syncope, palpitations; angina relieved by recumbent position is pathognomic -Arrhythmias, sudden death	-CHF, chest pain on exertion, arrhythmias, embolism, sudden death	-CHF; angina (amyloid), arrhythmias, thromboembolic disease
Physical	-Palpable S4, prominent apical impulse -Harsh crescendo-decrescendo systolic murmur b/w apex & left sternal border – radiates to axilla +/- MR; murmur ↑ with standing/valsalva (↓preload)	-CHF, diffuse/displaced, apex, murmur of MR/TR	-CHF, Rapid x&y descents, Kussmaul's sign (JVP fails to fall during inspiration), S3 more common than S4
Investigations			
-CXR	-LV/LA enlargement	-Cardiomegaly	-No cardiomegaly
-ECG	-LVH, Q waves, ST/T changes, ventricular arrhythmias	-LVH, ST/T changes, BBBs, PVCs, AF	-BBBs, low voltage
-ECHO	-Asymmetric septal hypertrophy, SAM of MV, MR, LVOT gradient	-LV dilatation/↓EF	-↓ EF, may see ↑wall thickness (speckled = amyloid)
-CATH	-Provokable LVOT obstruction, MR	-↑ LVEDP	-LVEDP usually >5mmHg > RVEDP
-Other	-Consider endomyocardial biopsy		
-Blood	-CBC (anemia), Lytes (diuretics), Cr (hypoperfusion), INR/PTT (anticoagulation)		
-Medical Tx	-B-blockers (1st line), CCB, Disopyramide -Dual-chamber pacing another option	-ACEi, hydralazine, nitrates, antiarrhythmics -Anticoagulation -AICD	-Diuretics, antiarrhythmics -Anticoagulation -AICD/pacemaker prn
-Surgical Tx	-Septal myomectomy, ETOH ablation of septal arteries	-Transplant	-Transplant
Anesthetic Management			
Optimization	-Cardiology consult/ AICD management -Anxiolytic (avoid tachycardia) -Antibiotic prophylaxis (moderate risk category)	-Cardiology consult -AICD management -Consider anxiolytic	
Anesthetic Options	-Caution with spinal/epidural (↓ afterload/preload may worsen obstruction); many case reports of success in OB	-Neuraxial a good option (but patients often anticoagulated)	
OR setup	-Defibrillator/external pacing pads; preinduction arterial line; consider CVP, PAC, TEE		
GOALS			
Contractility	-Mild decrease with β blockade/inhalational agents	-Maintain	-Maintain
Rate& Rhythm	-Slow SR	-NSR/normal rate	-*SR crucial*/ avoid bradycardia (SV fixed)
Afterload	-Maintain	-Low-normal	-maintain
Preload	-High-normal	-normal	-High-normal
Treatment of Hypotension	-Volume first, then Vasopressor (phenylephrine), finally consider decreasing contractility (volatiles, beta-blockers)	-Inotrope, judicious use of volume, caution with vasopressor	-Inotrope, be very careful with volume
	→ IABP, LVAD, ECMO (last resorts)		

Induction Technique	-IV or IH safe choices (avoid ketamine – tachycardia) minimize sympathetic response to laryngoscopy (B-blocker, spray cords, deep anesthesia)	-cardiac (++ opioids beneficial)	-cardiac, maintain high-normal heart rate
Maintenance	-mild myocardial depression, maintain IV volume and SVR	-minimize myocardial depression	-avoid bradycardia, maintain IV volume

PATHOPHYSIOLOGY

- **Classification (old classification system, most common types)**
  - Dilated cardiomyopathy
    - Idiopathic - 50%
    - Myocarditis - 9%
    - Ischemic heart disease - 7%
    - Infiltrative disease - 5%
    - Peripartum cardiomyopathy - 4%
    - Hypertension - 4%
    - HIV infection - 4%
    - Connective tissue disease - 3%
    - Substance abuse - 3%
    - Doxorubicin - 1%
    - Other - 10%
  - Hypertrophic cardiomyopathy
    - Hypertrophic obstructive cardiomyopathy (HOCM) is a subset (less than 25%)
    - Hypertensive cardiomyopathy
    - Valvular cardiomyopathy (e.g. AS)
  - Restrictive cardiomyopathy
    - A non-dilated ventricle with normal wall thickness
    - Ventricular walls that are rigid, resulting in severe diastolic dysfunction and restrictive filling that produces elevated filling pressures and dilated atria
    - Normal left ventricular systolic function in the vast majority of patients
  - Arrhythmogenic right ventricular cardiomyopathy
    - characterized by progressive fibrofatty replacement of RV myocardium, with relative sparing of the septum, and potential for LV involvement in later stages
    - familial and progressive with a predominance in young males
    - AD inheritance with incomplete penetrance
    - should be suspected in patients with multiple ectopic beats or tachycardia of right ventricular origin
    - cause of sudden death in previously symptom-free young people, often during strenuous exercise.
  - Unclassified cardiomyopathies
- **Secondary Classification**
  - Can also classify cardiomyopathies based on specific etiologies:
    - Ischemic cardiomyopathy
    - Valvular cardiomyopathy
    - Hypertensive cardiomyopathy
    - Inflammatory cardiomyopathy
    - Metabolic cardiomyopathy
    - General systemic disease (eg CTD, infiltrative diseases)
    - Muscular dystrophies and neuromuscular disorders
    - Drugs

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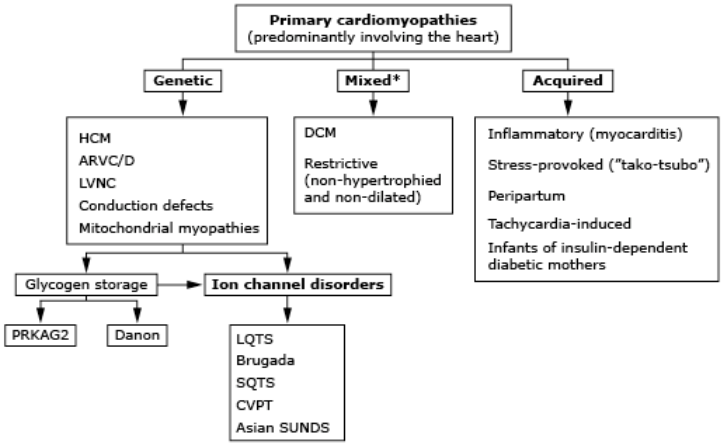
- UpToDate

AHA primary cardiomyopathies



Primary cardiomyopathies in which the clinically relevant disease processes solely or predominantly involve the myocardium. The conditions have segregated according to their genetic or nongenetic etiologies.
* Predominantly nongenetic; familial disease with a genetic origin has been reported in a minority of cases.
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Etiologic classification of cardiomyopathy-I

Infectious
Bacterial
Diptheria*
Tuberculosis*
Typhoid fever*
Rheumatic fever*
Scarlet fever*
Meningococcal*
Pneumococcal
Gonococcal
Brucellosis
Tetanus
Melidosis
Tularemia
Pertussis
Spirochetal
Syphilis
Leptospirosis*
Lyme disease*
Rickettsial
Typhus
Rocky mountain spotted fever*
Q fever
Viral
Poliomyelitis*
Influenza*
Mumps*
Rubella*
Rubeola*
Variola*
Varicella*
Epstein-Barr*

Viral (cont'd)
Coxsackievirus*
Echovirus*
Cytomegalovirus*
Hepatitis*
Rabies*
Mycoplasma*
Psittacosis*
Herpes
Encephalitis
Arboviruses*
Mycotic
Actinomycosis
Blastomycosis
Moniliasis
Aspergilliosis
Histoplasmosis*
Coccidiomycosis
Cryptococcosis*
Candidiasis
Protozoal
South American
trypanosomiasis*
African trypanosomiasis*
Toxoplasmosis*
Malaria
Amebiasis
Leishmaniasis
Balantidiasis
Sarcosporidiosis
Helminthic
Trichiniasis*
Echinococcosis

Helminthic (cont'd)
Schistosomiasis*
Ascariasis
Heterophydiasis
Filariasis
Paragonimiasis
Strongyloidiasis
Cysticercosis
Visceral larva migrans
Toxins and drugs
Adriamycin*
Amphetamine*
Antimony
Arsenic*
Carbon monoxide
Carbon tetrachloride
Catecholamines*
Cobalt*
Cocaine*
Cyclophosphamide
Emetine
Ethyl alcohol*
Lithium
Lead
Methysergide
Phenothiazine drugs
Phosphorus*
Tricyclic antidepressants
Zidovudine*
Radiation*•

* Conditions that may manifest clinically as dilated cardiomyopathy.
¥ Conditions that may manifest clinically as restrictive cardiomyopathy by daggers.
? Conditions that may manifest clinically as hypertrophic cardiomyopathy (These designations are neither obligatory nor exclusive).
Adapted with permission from Abelmann, WH. Introduction to Atlas of Heart Diseases, Vol. II: Cardiomyopathies, Myocarditis and Pericardial disease, Abelmann, WH (Ed), Current Medicine, Philadelphia, 1995, p. 1.

Etiologic classification of cardiomyopathy-II

Genetic
Genetic hypertrophic cardiomyopathyΔ•
Genetic dilated cardiomyopathy*
Metabolic
Endocrine
Acromegaly*•Δ
Thyrotoxicosis*
Hypothyroidism*•Δ
Pheochromocytoma*•Δ
Diabetes mellitus
Familial storage diseases
Glycogen storage diseases*Δ
Refsum disease
Niemann-Pick disease
Hand-Schuller-Christian disease
Fabry's disease*Δ
Gangliosiderosis
Gaucher's disease*•
Sandhoff's disease*
Mucopolysaccharidosis*
Hunter's syndrome
Hurler's syndrome
Nutritional
Beriberi*
Kwashiokor*
Pellagra
Selenium deficiency
(Keshan's disease)*
Other
Hypokalemia*
Carnitine deficiency*
Uremia*

Hematologic/oncologic
Hematologic disorders
Leukemia*
Myeloma
Sickle cell anemia*
Anemia*
Henoch-Schonlein purpura*
Neoplastic diseases
Primary neoplasms•
Metastatic neoplasms•
Deposits
Hemochromatosis*•
Oxalosis
Ochronosis
Amyloid disease•
Heredofamilial neurologic and neuromuscular diseases
Progressive muscular dystrophy (Duchenne)*
Limb-girdle muscular dystrophy (Erb)*
Fascioscapulohumeral dystrophy (Landouzy-Dejerine)
Humeroperoneal ataxia
Friedreich's ataxia Δ
Myotonia atrophica (Steinert)*
Myasthenia gravis
Chronic progressive external opthmoplegia (Kearns-Savre)
Familial centronuclear myopathy
Juvenile progressive spinal muscular atrophy (Kugelberg-Welander)
NeurofibromatosisΔ

Endomyocardial diseases
Endomyocardial fibrosis*•
Hypereosinophilic heart disease
(Loffler's)•
Endocardial fibroelastosis*•
Inflammatory
Connective tissue diseases
Rheumatoid heart disease*
Ankylosing spondylitis
Systemic lupus erythematosus*
Scleroderma*•
Dermatomyositis*
Periarteritis nodosa
Granulomatous
Sarcoid*
Wegener's granulomatosis*
Granulomatous myocarditis*
Other inflammation
Giant cell myocarditis*
Hypersensitivity myocarditis*

* Conditions that may manifest clinically as dilated cardiomyopathy.
Y Conditions that may manifest clinically as restrictive cardiomyopathy by daggers.
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