Dilated Cardiomyopathy (D-CM) 95%

- Mechanism
  - Insult → Eccentric Hypertrophy (dilation>thickening) → Systolic Dysfunction
- Etiology (NB you must rule out ischemic induced D-CM by always doing coronary angiography)
  - **Idiopathic** (25%) likely undiagnosed viral, familial, alcoholic, etc
  - **Familial** (25%) mutation in genes that code for cytoskeletal, nuclear, and filament proteins
  - Secondary Heart Problem (25%)
    - "Burn Out" **SVT**
    - "Burn Out" **HTN**
    - Chronic Uncontrolled **AI/PI**
  - Metabolic (rare)
    - **Peripartum**: 3mo prepartum to 6mo postpartum, RF: old AA, 50% will improve, overall risk is <0.1%, 20% maternal mortality, increased risk with multiparity, age, etc., mothers should be advised to have no more children b/c they are at increased risk with subsequent pregnancies
    - Endocrine: **HypoTh**, Pheo, Acromegaly
    - Vitamin: **LATE Hemochromatosis, Selenium Deficiency, Thiamine Deficiency aka "Wet Beriberi"**
    - Electrolyte: **Hypophosphatemia**
  - Infections (rare) (refer to myocarditis/pericarditis but below are some unique ones)
    - Bacterial
      - **Lyme** Disease w/ *Borrelia burgdorferi*
    - Parasite
      - **Chagaz** Disease w/ *Trypanosomi cruzi*
    - Viral
      - **HIV** Pts (some controversy as to whether 2/2 HIV itself, HAART or some other infection but seen in 8% of HIV pts)
  - Toxins (rare)
    - **EtOH**
    - **Radiation**
    - **Glue Sniffing**
    - **Anthracyclines/Mitoxantrone/Trastuzumab**: generate free radicals from iron which particularly damage the heart b/c the heart is not able to detoxify the radicals b/c it has low levels of catalase, dexrazoxane removes iron from anthracycline-iron complex preventing free-radical formation and thus is given to pts receiving anthracycline chemotherapy
    - **Sympathomimetics**: Cocaine, Amphetamines (causing direct toxic injury along w/ vascoconstriction)
    - Antimicrobials: Alpha-Interferon, Penicillin, Sulfa-containing drugs, Tetracycline, TB meds, HAART
    - Heavy Metals: Lead, Arsenic, Cobalt
  - Autoimmune (rare)
    - Rheumatologic Dz: Scleroderma, RA, SLE, Polymyositis
    - Infiltrative: Amyloidosis, Sarcoidosis
  - Other (rare)
    - "**Takasubo**" aka Broken Heart Syndrome, pt has recently undergone stress either physiologic (many poorly controlled diseases, etc) or psychiatric (death in family, etc), results in apical dilation/ballooning, mimics an MI w/ pain, ST-T changes and troponin elevation, completely resolves in a few weeks
    - Hypereosinophilia
    - OSA
- S/S
  - L/R Systolic CHF
  - MR/TR b/c of annular dilation
  - Thromboembolism b/c of stagnant blood in big ventricles
  - Arrhythmias
  - CP despite clean coronaries
- Dx
  - EKG: decreased voltage 2/2 decreased mass, conduction abnormalities
  - CXR: CM and signs of CHF
  - Echo: decreased EF, ventricular dilation, ventricular thinning, regional/global hypo/akinesis, mural thrombi
  - Coronary Angiography: always rule out ischemia/infarction as a cause
  - Endomyocardial Bx: sometimes helpful if you suspect systemic disease
- Tx (refer to CHF notes)

Hypertrophic Cardiomyopathy (H-CM) 4%

- Mechanism
  - Genetics → Different Types of Ventricular Hypertrophy →
    - 1° Concentric Hypertrophy (thickening>dilation) → Diastolic Dysfunction

        - 2° Asymmetric Septal Hypertrophy (outflow tract obstruction) → Systolic Dysfunction aka Hypertrophic Obstructive CM ("HOCM")
        - 3° many others
- Etiology (NB you must rule out HTN/AS thru history but in pts with both H-CM and HTN/AS the hypertrophy of H-CM is out of proportion to the hemodynamic load just due to HTN/AS)
    - Idiopathic (50%) spontaneous mutation
    - Familial (50%) AD mutation in genes that code for sarcomere proteins
        - more common in AA w/ three age peaks depending on which protein affected: 10s(M>F), 40s(M=F), 60s(F>M)
        - Costello Syndrome: H-CM, Mental/Growth Retardation, Craniofacial Distortions, Acanthosis Nigricans, Veruccous Papillomata of the Nose, Hyperextensibility of Digits, Soft Skin w/ Excess Wrinkling on Dorsum of Hands and Deep Creases on Palms/Soles
        - Dr. Grayburn/Anwar/Kowal are doing a study looking at potential genes
- S/S (depends on the type of hypertrophy the pt has but overall 70% are asymptomatic at diagnosis)
    - 1° Concentric
        - L/R Diastolic CHF
        - CP b/c (1) there is increased mass which compresses micro-coronaries and (2) the normal macro-coronaries are unable to meet the demands of increased mass (NB pts have clean epicardial coronaries)
        - Sudden Death from ventricular arrhythmias
    - 2° Asymmetric Septal aka "HOCM"
        - There is a subaortic outflow obstruction due to the septal hypertrophy but also due to the Systolic Anterior Motion "SAM" effect on the anterior leaflet of the MV (as blood shoots around the septum a venturi effect occurs which decreases pressure pulling mitral leaflet toward septum blocking flow mid systole) resulting in a unique systolic ejection murmur and a normal mitral regurgitation murmur b/c of incomplete coaptation of leaflets, these murmurs change with certain maneuvers to differentiate b/t AS and true MR (refer). This obstruction results in Sx similar to AS w/ angina, syncope and systolic CHF
        - Asymptomatic with just Sudden Death esp in 15-35yo athletes engaging in sports (NB primary cause of SCD in this population is trauma but the next most common is HOCM →Coronary Anomalies → Cardiac Hypertrophy → Ruptured Aorta → Intramyocardial Course of LAD → AS)
        - Bisferiens "Spike & Dome" Carotid Pulse (rapid upstroke 2/2 increased contractility 2/2 hypertrophy but at about midsystole the "SAM" effect occurs which results in partial occlusion and thus a dip in mid-upstroke resulting in a bisferious) and prominent a wave in jugular venous wave
- Dx
    - FHx
    - EKG: ischemic STT changes, L/RVH, L/RAE, conduction abnormalities
    - CXR: CM and signs of CHF
    - Echo: determine type of hypertrophy (septum/post wall thickening >1.3 or septum >15mm), "SAM" effect
    - Coronary Angiography: outflow gradients, "Brockenbrough Sign" (decrease in pulse pressure post extrasystole which in AS actually increases)
    - Endomycoardial Bx: fiber hypertrophy AND disarray (unlike the hypertrophy seen in HTN/AS/PS in which there is only hypertrophy NO disarray)
- Tx
    - Asymptomatic
        - Avoid Extreme Strenuous Activity & Dehydration
    - Symptomatic
        - Medical Therapy
            - Negative Chronotropes
            - Disopyramide (decreases outflow gradient, improve symptoms and improve exercise tolerance)
            - IE Prophylaxis
            - ICD if + RFs (h/o NSVT or VT/VFib, +FHx of SCD, syncopal episodes, hypotension w/ exercise, LV wall >30mm, gradient >30mm of Hg, high risk mutations)
        - Surgical/Percutaneous Intervention
            - Concentric Hypertrophy (outflow gradient <50mm of Hg) then…
                - Transplant
            - Asymmetric Septal Hypertrophy (outflow gradient >50mm of Hg) then…
                - 1° Surgical Myomectomy
                - 2° Percutaneous Catheter EtOH Septal Ablation of first septal perforator of LAD resulting in a controlled MI that then thins out the septum (during the procedure there is a high r/o AV block, coronary dissection, anterior MI vs after the procedure the resulting scar can become a nidus for tachyarrthmias)
    - Screening: if +FHx pt should undergo echo Q1yr

Restrictive Cardiomyopathy (R-CM) 1%

- Mechanism
    - Insult → Fibrosis → Diastolic>Systolic Dysfunction
- Etiology (NB you must rule out Restrictive Pericarditis with various studies (refer) esp b/c pericarditis is treatable while R-CM is not)

- **Myocardial**
  - NON-Infiltrative Disease
    - Idiopathic
      - Type I additional skeletal myopathy
      - Type II NO skeletal myopathy but associated w/ Noonan's Syndrome
    - **Diabetes**
  - Infiltrative Disease
    - Other: **Amyloidosis** and **Sarcoidosis**
    - Metabolic: **EARLY Hemochromatosis**
    - **Autoimmune**: Scleroderma, Poly/Dermatomyositis
    - **Storage Disease**
- **Endomycardial**
  - **Hypereosinophilia**: Loeffler's Syndrome, Endomyocardial Fibrosis Syndrome, etc
  - **Toxins**: Radiation, Anthracyclines, Bulsulfan, etc
  - **Serotonins**: Carcinoid Syndrome, Serotonin Drugs, etc
  - **Metastatic Cancer**
  - **Post Open Heart Surgery**

- S/S
  - R/L Systolic/Diastolic CHF (but importantly it usually affects the **R** heart more)
  - **Conduction Abnormalities** are particularly common
- Dx
  - EKG: low voltages, various conduction abnormalities NB LVH on TTE but low voltage on EKG
  - CXR: nl heart but signs of CHF
  - Echo: (normal ventricles but enlarged atria looking like "Mickey Mouse") NB "bright sparkled" appearance of myocardium suggesting amyloidosis
  - MRI (refer to pericarditis section)
  - Cath (refer to pericarditis section)
  - Endomyocardial Bx (sometimes helpful if you suspect infiltrative disease)
- Tx
  - Remove/Treat Offending Agent
  - CHF Tx (refer)
  - Anticoagulation
  - In the end no true Tx with very high mortality
  - do not use digoxin b/c pro-arrhythmic
  - Transplant