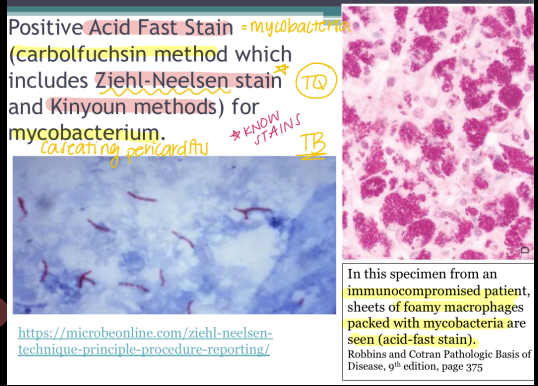


Pericarditis = outer portion

- * immunologic: SLE, scleroderma, Dressler Syndrome, eosinophilic
- * infectious
 - viral: coxackie A & B
 - Bacterial:
 - Fungal:
 - Protozoa:

WBOT * caseous pericarditis
 pt px w/ pleuritic CP, diffuse ST ↑, pain relieved by leaning forward
 → **CONSTRUCTIVE PERICARDITIS**



Pericardial effusion = water bottle heart

ACUTE → cardiac tamponade → **BECK'S TRIAD** + **pulsus paradoxus**: ↓BP during inspiration

- 1 muffled heart sounds
- 2 JVD - blood backing up
- 3 Hypotension & tachycardia

Serous = pericardial effusion of serous fluid

* **viral** infection in **YA**
 - sudden/severe onset pleuritic chest pain relieved by sitting up & leaning forward

* **Diffuse ST elevation**
 + water-bottle heart on Xray

VIRAL = COXACKIE B

Fibrinous/Serofibrinous (MC)

* **Dressler Syndrome**, uremia, radiation, rheumatic fever
 2-6 wks post-MI, pleuritic chest pain, palpitations, arthralgia + Fever, Tach, friction rub
 Tx Dressler syndrome = **IBUPROFEN**
 anti-actin & myosin antibodies causing autoimmune rxn

Purulent/Suppurative

infx by:
 1 extension
 2 pericardial seeding
 3 Lymphatic extension
 4 Direct introduction
 → can progress to **Constrictive pericarditis**

* **gram stain** strep pneumo
 calcification seen in chronic or healed pericarditis
 - ↓C.O.
 - **pulsus paradoxus**
 - **Kussmaul's sign** = ↑JVP w/ inspiration

Myocarditis: non ischemic inflammation (infx & immune mediated)

* **Dallas criteria**
 - inflammatory infiltrate w/ **nerosis** or degeneration

* **Diagnosis based on cell type**

Clinical findings of viral myocarditis

- May be asymptomatic in mild cases
- Clinical history: **viral prodrome** → Angina!
 = recent febrile illness with myalgia followed by angina-like chest pain, dyspnea or palpitations.
- Lab and pathology findings:
 - Elevated troponin levels
 - Increasing viral titers.
 - **Endomyocardial biopsies** yield a diagnosis in 10 to 20% of patients with myocarditis symptoms.
 - PCR may be performed on endomyocardial biopsies to identify possible viral causes of myocarditis

→ **Lymphocytes**: viral infx → coxackie A & B + enterovirus EJ
 * **WBOT**: young pt dies of SCD w/ fib, histology shows lymphocytic infiltrate. = **viral myocarditis**

→ **Neutrophils**: bacterial infx: Lyme, C. diphtheriae, N. meningitidis
 multi-system dz, may need temporary pacemaker

→ **Eosinophils**: parasite: **T. cruzii** Chagas dz
 - chagoma + Romano's sign
 - Reduviid bug
 * Heart & GI affected
 Cardiomyopathy (Dilated)
 - RBBB
 Mega colon

Constrictive Pericarditis

- A chronic or healed pericarditis
- Heart is encased in a dense, fibrous or fibrocalcific (cap) that may be up to 1.0 cm thick with obliteration of pericardial space and may resemble a plastic mold (concreto cordis)
- Limits diastolic expansion and cardiac output ↓C.O.
- Cardiac hypertrophy and dilation cannot occur
- Cardiac output may be reduced at rest and the heart may not be able to increase cardiac output with increased demand
- **Physical findings**: Most common finding is elevated jugular venous pressure (JVP) - backed up blood
- Less common findings are:
 - Pulsus paradoxus
 - **Kussmaul's sign**: a paradoxical increase in jugular venous pressure with inspiration (normally declines with inspiration)
 - This sign is NOT usually seen in cardiac tamponade
 - Pericardial knock heart sound heard earlier than an S3
 - Peripheral edema, ascites, pleural effusion, pulsatile hepatomegaly, cachexia

| | PERICARDITIS | MYOCARDITIS |
|-----------|--|--|
| CLINICAL | Sharp or pleuritic chest pain, worse when supine Pain radiating to left trapezius ridge Dyspnea | Flu-like (myalgias, fatigue, fever) Dyspnea, new CHF Pediatrics - grunting, retractions |
| ETIOLOGY | Viruses & Idiopathic Most Common Lupus/Sarcoid Radiation Drugs (procainamide, hydralazine) | Infectious (Influenza, Lyme, Chagas, etc) |
| DIAGNOSIS | Clinical EKG Stages: Beck's Triad? (1) PR depression with ST elevation (if ST and no ST depression in aVL suggests pericarditis) (2) Normalization (3) T wave inversion Pericardial effusion on ultrasound Troponin may be elevated | Difficult EKG - Sinus tachy, dysrhythmia, Troponin may be elevated ECHO may show hypokinesis Gold std: biopsy |
| TREATMENT | Ibuprofen 400-800 mg q6-8 hrs x 2 weeks Colchicine 0.6 mg BID (prevents recurrence) *Note: in US formulation is 0.6mg tablets Admit for high risk: large effusion, T2-3 C, signs of myopericarditis | Admit to monitored setting Supportive (may need LVAD, ECMO) Extracorporeal membrane oxygenation |

Immune-mediated myocarditis → Lupus, drug hypersensitivity

Endocarditis

* Infective endocarditis: valves!
Sxs: Fever, arrhythmia, anemia

- Acute = toxic presentation ^{- treat empirically}
= Staph. Aureus

- Subacute = mild toxicity
= Strep viridans / enterococcus

* Septicemia: FEVER
- alt. mental status

IE: Bacterial etiologies

Etiology:

- **Native valves**
 - Oral Streptococcus #1 (Enterococcus) strep viridans
 - *S. aureus*, Coagulase negative *Staphylococcus*, Gram Negative rods (HACEK)
 - **Prosthetic valves (early)**
 - Coagulase negative *Staphylococcus* #1 (*S. epidermidis*)
 - *S. aureus*, HACEK, Oral *Streptococcus* and *Enterococcus*, *Candida*
 - **Prosthetic valve (late)**
 - Oral Streptococcus #1 (Enterococcus)
 - Coagulase negative *Staphylococcus*, *S. aureus*, HACEK, *Candida*
 - **IV drug user**
 - *Staphylococcus aureus* #1
 - Oral *Streptococcus* and *Enterococcus*, Gram Negative rods (HACEK), *Candida*, Coagulase negative *Staphylococcus*
 - ❖ **HACEK Group Infections**: Group of gram-negative fastidious rods
 - *Haemophilus aphrophilus*, *Actinobacillus actinomycetemcomitans*, *Cardiobacterium hominis*, *Eikenella corrodens*, *Kingella kingae*
- Most common cause of gram-negative endocarditis in non-IV drug users

LO3 Congenital Heart Dz of Adults

* Greatest env. risk factor = MATERNAL DIABETES → ESP Transposition of Great Vessels

* Congenital aortic stenosis = BICUSPID VALVE fused @ raphe

- younger pt w/ exertional syncope, SOB, chest pain
- crescendo-decrescendo systolic murmur

* COMPLICATIONS:

* aortic dissection/dilation ** WBOT & BERRY ANEURYSMS

- bicuspid often involved w/ coarctation → think TURNERS
- LV Hypertrophy

DING
DING
DING

* Coarctation of Aorta:

- young pt w/ HTN, diff. BP UE vs LE + "3-sign on xray" + Rib Notching

* males & TURNERS SYNDROME

* COEXISTS w/:

- * WBOT [* saccular / berry aneurysms @ circle of willis
- * bicuspid aorta
- * congenital mitral valve dz

* Pulmonic stenosis

- Noonan's or part of TDF
- crescendo decrescendo systolic murmur @ left 2nd intercostal
- px w/ exertional fatigue, dyspnea, CP, syncope
- RIGHT V Hypertrophy - pulmonary edema?

* ATRIAL SEPTAL DEFECT mc- ostium secundum Trisomy 21 - ostium primum

* FIXED SPLIT S2 in young adult w/ new exertional fatigue

- complications:

- irreversible pulmonary HTN
- paradoxical embolism → pt may present w/ DVT that became stroke
- atrial arrhythmias → px w/ AFib

Repair
that
shit!!

* VENTRICULAR SEPTAL DEFECT = MOST COMMON CONGENITAL BIRTH DEFECT

- L → R shunt can produce irreversible pulm HTN
- small, moderate or large

→ EISENMENGER'S COMPLEX WBOT

L → R shunt → pulm HTN

↑ R pressure reverses shunt → cyanosis

* PDA = CONTINUOUS MACHINE-LIKE MURMUR

- also causes eisenmenger syndrome

* TO CLOSE → indomethacin / ibuprofen // TO OPEN: PGE2

* Ebstein's Anomaly = atrialization of RV

* LITHIUM use in first trimester

→ SVT & WPW + exercise induced cyanosis ** WBOT

* Congenitally corrected Transposition of GV = NOT CYANOTIC aka can px in adults w/o detection

* Tetralogy of Fallot = DIGEOGE SYNDROME

22q11.2 microdeletion

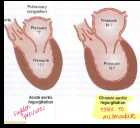
* Valvular Heart Dz

GROSS/ETIO

SYMPTOMS

DIAGNOSIS

AORTIC REGURG



CHRONIC: volume overload, thick LV = dilation to ↑ SV
- caused by aortic root dilation & leaflet abnormalities

ACUTE: commonly caused by
* AORTIC DISSECTION
* INFECTIVE ENDOCARDITIS
→ SURGICAL EMERGENCY

CHRONIC: Diastolic blowing murmur
* widened pulse pressure
⊕ Quinke pulse - finger pulsations

if symptomatic or LVEF < 50%
= valve REPLACEMENT
WBOT

ACUTE: short diastolic blowing murmur
→ Both require surgery

AORTIC STENOSIS

* Senile - NO FUSION
* WBOT just calcification

* RHD - only rheumatic if fusion AND Mitral valve involvement

Angina, syncope, SOB

* Systolic ejection murmur
RADIATING TO NECK
+ S4

* BICUSPID AORTA
- calcifications, endarteritis
- Ejection click
- AORTIC ANEURYSM
↓
GET AN ECHO
WBOT

Tx
→ if symptomatic or LVEF < 50%
→ valve replacement

MITRAL REGURG

* MVP - Marfan!
* ischemic papillary muscle after MI

Dyspnea, orthopnea, & PND

CHRONIC Holosystolic apical murmur radiating to axilla maybe S3 w/ CHF

follow before they get symptomatic

ACUTE = EMERGENCY
* Hypotension, HF, SHOCK
* Holosystolic murmur radiates to BACK
→ Require surgical intervention

MITRAL STENOSIS

ALL CASES CAUSED BY RHEUMATIC HEART DZ
p+ px in 40s-50s
* fish mouth/button hole
- LA dilates
- pulm pressure increases
→ pulm. HTN

Pulmonary HTN → hemoptysis
Hoarseness - LA presses on Rec. L. = Ortner's syndrome

Opening SNAP followed by low pitched early diastolic rumble

* Mitral Facies

* surgical intervention based on Diameter symptoms @ any point = valvotomy open valve up w/ balloon (Not a replacement)

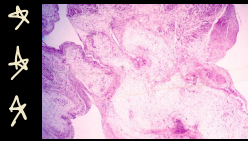
MITRAL VALVE PROLAPSE

* HOODING of leaflets w/ elongated chordae tendinae
ON HISTO: myxomatous increase of proteoglycans

Usually asymptomatic

* Mid-systolic click

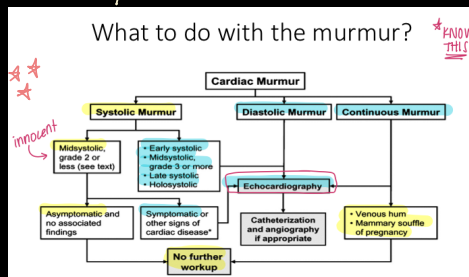
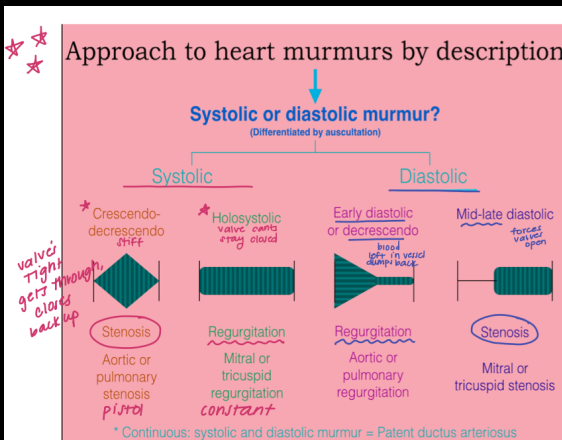
* watch out for Marfan's & Ehlers Danlos



WBOT

* indications for ECHO:

- Diastolic murmur
- grade 3+
- symptomatic **WBOT**
- systolic murmur + click



WBOT

INFECTIVE ENDOCARDITIS

- strep. viridans - previously damaged native valves
 - gram ⊕ cat ⊖ optochin resistant diplococci
- S. aureus - IV drug users & nosocomial
 - gram ⊕ cat ⊕ coag ⊕ β hemolytic cocci
- S. epidermidis - prosthetic valves
 - gram ⊕ coag ⊖ cat ⊕ cocci
- HACEK group - commensals of oral cavity
 - may be culture NEGATIVE

WBOT

Osler nodes = immunologic vasculitis
•• painful

Janeway lesions = hemorrhagic microabscess due to septic emboli

Haemophilus aphrophilus (now called Aggregatibacter aphrophilus) and Aggregatibacter paraphrophilus, Actinobacillus actinomycetemcomitans (subsequently called Aggregatibacter actinomycetemcomitans), Cardiobacterium hominis, Eikenella corrodens and Kingella kingae
• gram-negative bacilli that are commensals in the oral cavity.
• may be culture negative because fastidious, slow-growing

Dukes Criteria - Dx

- Janeway, Osler nodes, Roth spots

Gross Morphology - BULKY vegetations can cause **EMBOLI**

WBOT

septic infarcts @ brain/kidney
mycotic aneurysms

→ **STROKE**

Histology - lots of NØ, fibrin, bacterial colonies

NON-INFECTIVE ENDOCARDITIS

WBOT

pt has pancreatic cancer

NBTE: @ line of closure of mitral valve loosely attached vegetations
ASSOCIATED w/ hypercoagulable states → mucinous adenocarcinoma

Non Bacterial Thrombotic Emboli

Libman-Sacks vegetations on both sides of leaflets

+ fibrinoid necrosis
SLE - rarely causes problems

→ Antiphospholipid syndrome may cause NBTE

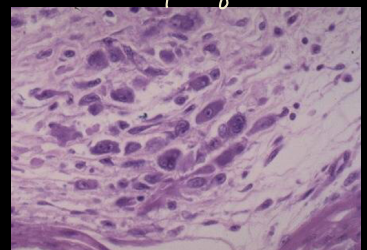
RHEUMATIC HEART DISEASE due to M-protein molecular mimicry

Fibrinoid necrosis
McCallum's plaques

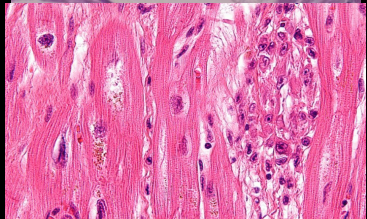
ACUTE = **Mitral regurg**; friction rub, tach, arrhythmias, HF

ASCHOFF BODIES = activated MØ

Anitschkow cells = caterpillar cells
- made of nuclear chromatin



CHRONIC = **Mitral stenosis**; YEARS / DECADES → **FISH MOUTH** fused chordae
+ Aortic stenosis → **FUSION**



* For Tachydysrhythmias:

NEVER use CCB to tx:

- Infants
- anyone w/ WPW
- children w/ CHF
- children on Beta Blockers

→ hypotension & cardiac arrest

* Girl got up to brush hair & fell over = situational syncope (Vagal tone)

RED FLAGS FOR SYNCOPE

shouldn't play sports, refer to cardiologist

- Recurrent, atypical or unexplained episodes.
- Syncope with exercise !! *During sports!*
- Syncope with palpitations or chest pain
- History of cardiac abnormalities *repaired*
- Abnormal cardiac physical exam or ECG
- Neurologic deficits *New murmurs*
- History or family history of neurologic disorders. *Romberg*
any hx of unexplained death <50

L13 Ped CV disorders

WBOT

Kawasaki

Tx: IVIG & Aspirin

There are three distinct phases of Kawasaki disease:

WBOT

1. acute febrile phase day 1-11
2. subacute phase days 11-21
3. convalescent phase days 21-60

CHRONIC up to 2 mos → cardiac complications

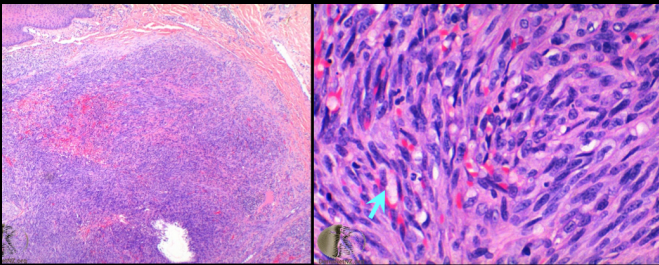
→ rash, conjunctivitis, lymphadenopathy, strawberry tongue

→ peeling hands & feet → *greatest risk for sudden death*

There is also a chronic phase for children developing cardiac complications.

→ presence of coronary aneurysms

Kaposi sarcoma CD31 & 34 ⊕



- Vessels lined by thin endothelial cells dissecting stroma
- Spindle cell proliferation with slitlike spaces containing RBC
- Admixed with a variable chronic inflammatory infiltrate
- Positive for both endothelial (CD31, CD34) and smooth muscle markers (smooth muscle actin)

spindle cells!
* TQ
**

* LIVER PROBLEMS / HYPERESTRINISM

Spider telangiectasia

- Radial, pulsatile array of dilated subcutaneous arteries or arterioles around a central core
- Blanches with pressure applied to its center, usually on face, neck, upper chest of pregnant women and patients with cirrhosis, may be associated with hyperestrinism

Epithelioid Hemangioendothelioma:

Clinical and pathologic findings

- Gross: Reddish-brown lobulated masses, well demarcated with irregular scalloped borders and a red hemorrhagic appearance
- Microscopically: Anastomosing cords, solid nests, or short strands of round to slightly spindled eosinophilic neoplastic endothelial cells embedded in a chondroid-like or hyalinized stroma

Glomus Tumor = PAINFUL smooth muscle & CD34 ⊕

- Blue or red blanchable papules or nodules in deep dermis or subcutis
- Round and regular tumor cells with uniform circular nuclei
- Positive for smooth muscle actin, may be positive for CD34

myoleioma - marker for smooth muscle!!

small, regular nuclei

PAINFUL

<http://emedicine.medscape.com/article/1083405-overview>
<http://webpathology.com/case.asp?case=480>

CHECK FOR VHL ↓

Hemangioblastoma Benign

- Benign, predominantly involving central nervous system
- 1-2% of intracranial tumors
- Often in cerebellum; also spinal cord, meninges
- Slow growing and indolent
- Symptoms due to mass effect and peritumoral edema
- Either part of von Hippel-Lindau disease or sporadic (often with somatic mutation of VHL gene)
- Loss of VHL promotes increased production of vascular endothelial growth factor and erythropoietin (VEGF)
- May be associated with loss of unknown tumor suppressor gene at 22q13

*** do genetic studies (like clear cell renal carcinoma)



Pathological findings

- Reddish neoplastic growth
- Proliferation of capillaries with variable sized, closely packed, thin walled vessels
- Atypical stromal cells with hyperchromatic nuclei apoptosis seen

DILATED VEINELS - IRREGULAR

cerebellum but can be seen anywhere

* pay attn to genetic study VHL *