The Annual General Pediatric Review & Self Assessment

Nicklaus Children's Hospital

CARDIOLOGY

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Disclosure of Relevant Relationship

Dr. Katz has not had (in the past 24 months) any relevant conflicts of interest or relevant financial relationship with the manufacturers of products or services that will be discussed in this CME activity or in his presentation.

Dr. Katz will support this presentation and clinical recommendations with the "best available evidence" from medical literature.

Dr. Katz does not intend to discuss an unapproved/investigative use of a commercial product/device in this presentation.

Passing the Boards

- High Yield Review
  - Presentations
    - Murmurs
    - CHF
    - Cyanosis
    - Shock
  - Syndromes
  - Scenarios
  - EKGs
  - Arrythmias

Presentation of CHD

- Murmur
- CHF
- Cyanosis
- Shock
- High Risk of Heart Disease
  - Down Syndrome, Trisomy 13, 18, etc..

Murmurs

- Ejection Systolic Murmur
  - Pulmonary stenosis and TOF (LUSB)
  - Aortic stenosis (RUSB)
- Holosystolic Murmurs
  - VSD (LLSB) and Mitral regurgitation (apex)
- Continuous Murmurs
  - PDA (machine-like) (LUSB – left infraclavicular)
  - BT Shunt murmurs (throughout chest)
- Innocent Murmurs
  - PPS (Peripheral pulmonic stenosis) classically heard in infants
    - Ejection systolic (ULSB) radiating to both axilla
  - Still's murmur (classically 3 to 6 year old) (LSB to apex)
    - Vibratory sounding – louder in supine position
  - Venous hum (disappears when the head is turned to the side)
    - Supra and infra clavicular areas

CHF

- Congestive Heart Failure
  - From increased pulmonary blood flow
    - VSD, PDA, TGA, HLHS, Truncus Arteriosus
  - Decrease ventricular function
    - Dilated cardiomyopathy
  - Signs and Symptoms
    - Tachypnea (classic sign is comfortable tachypnea)
    - Tachycardia
    - Poor po intake / tires with feeds (sweats w/ feeds)
    - Poor weight gain

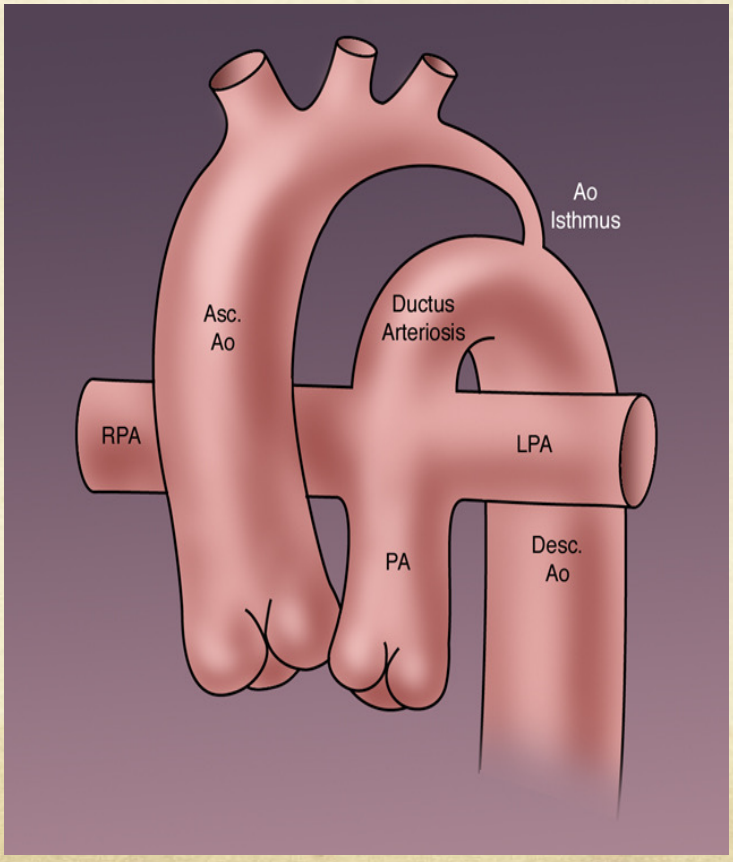
Cyanotic baby work-up

- Increased pulmonary vascular markings (white lungs on CXR)
  - TGA (increased or normal) – comfortable tachypnea
  - TAPVR – respiratory distress
  - HLHS – shock (mottled, acidotic, pulseless)
- Decreased pulmonary vascular markings (Dark lungs on CXR)
  - Tricuspid Atresia – Left superior axis on EKG
  - Pulmonary Atresia
  - Critical Pulmonary Stenosis and TOF – harsh SEM

Shock

- HLHS (Hypoplastic Left Heart Syndrome)
  - Cyanotic (often with sats in high 80's/low 90's)
  - Can present in shock (ductal dependent Systemic BF)
  - Start prostin ASAP
- Critical Coarctation & Critical Aortic Stenosis
  - Can present in shock (ductal dependent Systemic BF)
  - Start prostin ASAP

Critical Coarctation



Prostin

- Start a prostin infusion to keep ductus open
  - If suspect ductal dependent lesion (Echo)
    - Any neonate who presents in shock
    - Any neonate with severe hypoxemia
- Dose
  - To open ductus after closure
    - 0.1 mcg / kg /min
  - To keep ductus open (can use for weeks)
    - 0.02 – 0.05 mcg / kg/ min
- Complications (decreased by lower doses)
  - Apnea, hypotension, fever

Diagnostic Tests

- History & Physical
- Oxygen Saturation
- 4 extremity BPs
- Hyperoxia test
- EKG
- Echo
- (Cardiac Cath, EP Study, Holter)

Treatment

- MURMUR
  - Depends on heart defect
- CHF
  - Lasix, Lasix, Lasix, Digoxin, and good nutrition
- HYPOXEMIA
  - Nothing or if ductal dependent than Prostin infusion
- SHOCK
  - Start Prostin infusion, inotropes and fluid resuscitation

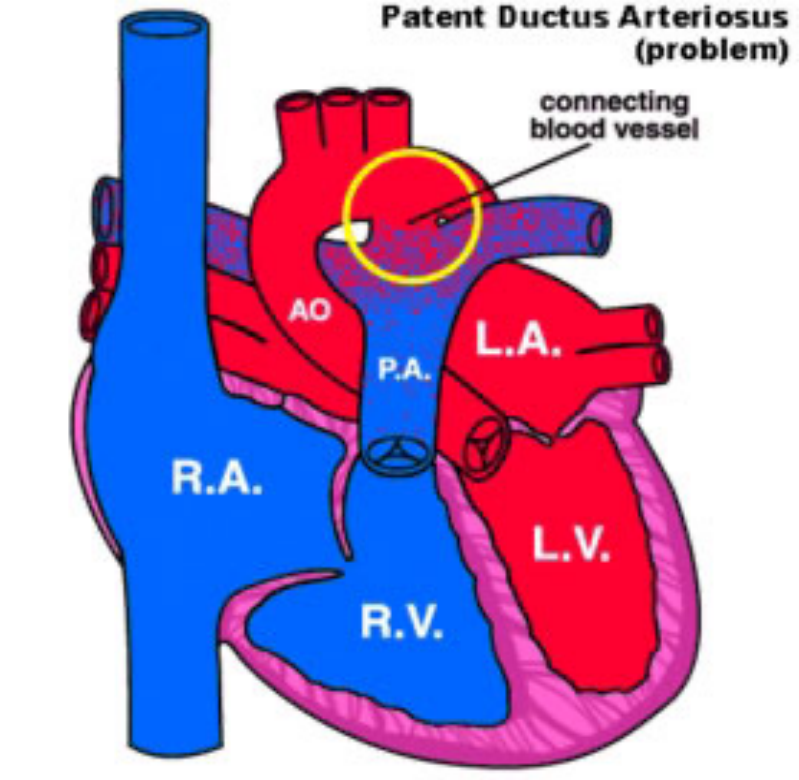
Treatment

- Ultimately most patients with congenital defects require :
  - Surgical correction
    - i.e. VSD closure, TGA repair (ASO), TOF repair
  - Surgical palliation
    - i.e. PA band, Norwood, Glenn, Fontan

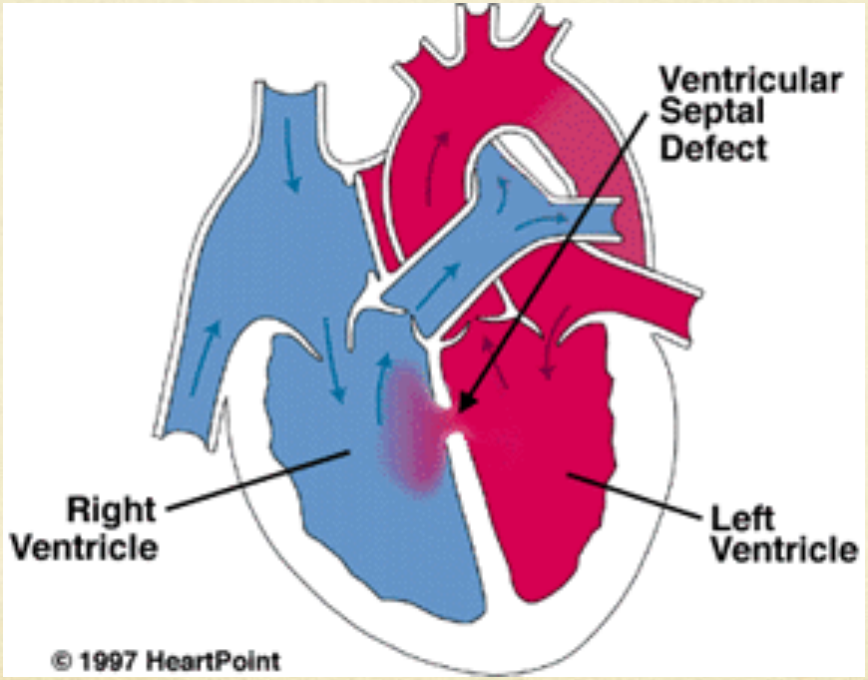
Acyanotic Heart Defects

- PDA
- Coartation of the Aorta
- ASD – soft ESM from relative PS and fixed split S2
- VSD
- AV Canal

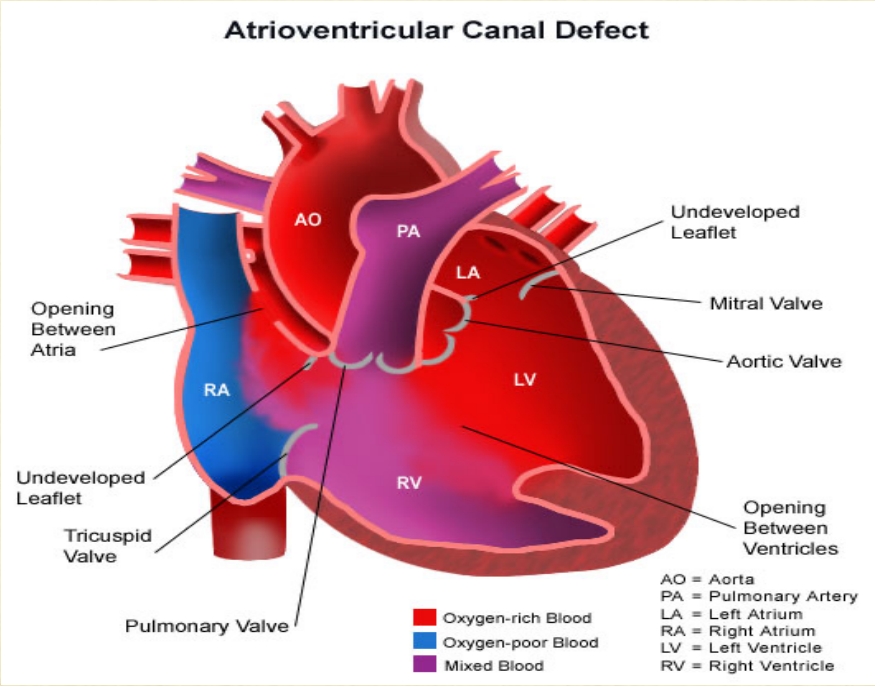
Patent Ductus Arteriosus (PDA)



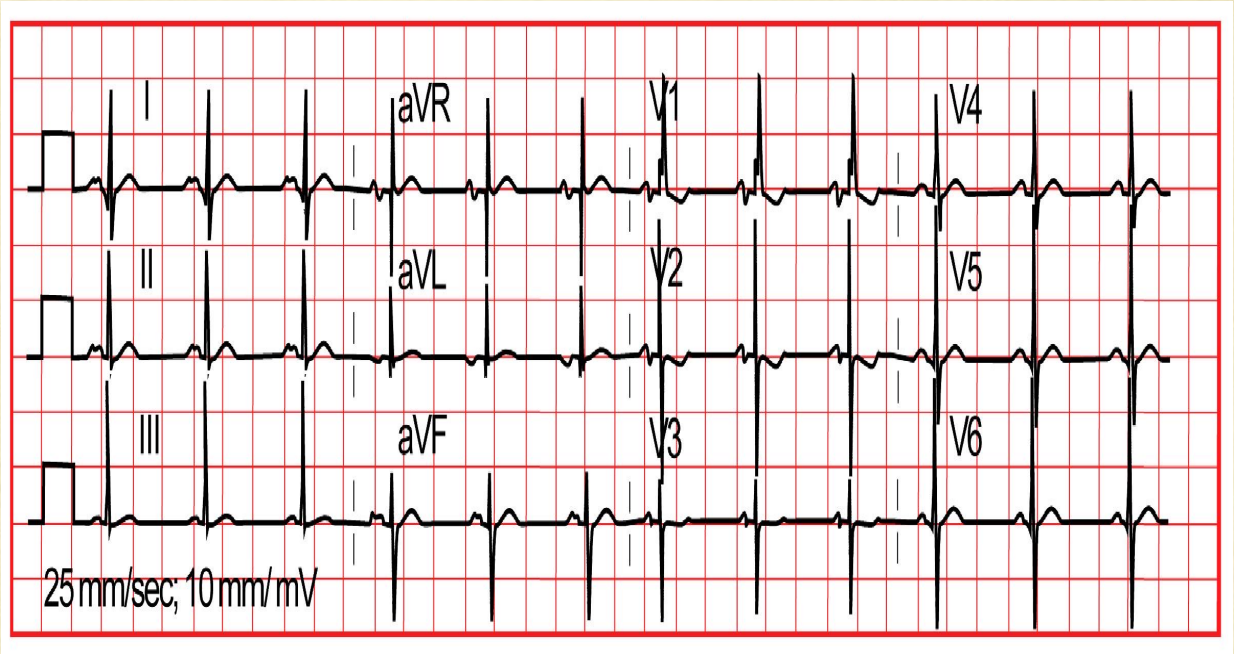
VSD Physiology



AV Canal Defect



Superior Axis



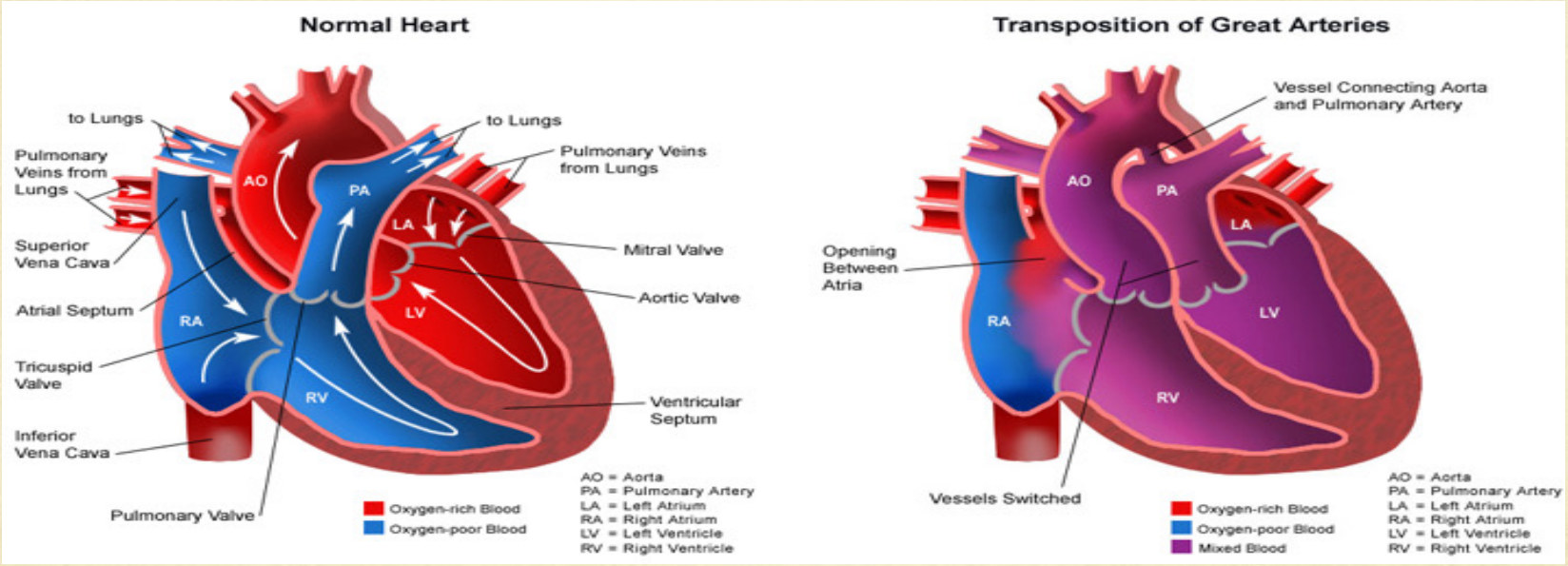
Cyanotic Heart Defects

- Tricuspid Atresia
  - Cyanosis, maybe ductal dependent PBF
- Pulmonary Atresia & Critical pulmonary Stenosis
  - Cyanosis, probable ductal dependent PBF
- TAPVR (Total Anomalous Pulmonary Venous Return)
  - Cyanotic, tachypneic, comfortable
  - Obstructed type (very cyanotic and sick)
    - Surgical emergency

Cyanotic Heart Defects

- TGA (Transposition of the Great Arteries)
  - Cyanosis, tachypneic but comfortable, no murmur
  - Most common cyanotic lesion presenting at birth
    - May need balloon atrial septostomy (if very blue)
    - Surgical repair as neonate

Transposition of The Great Arteries



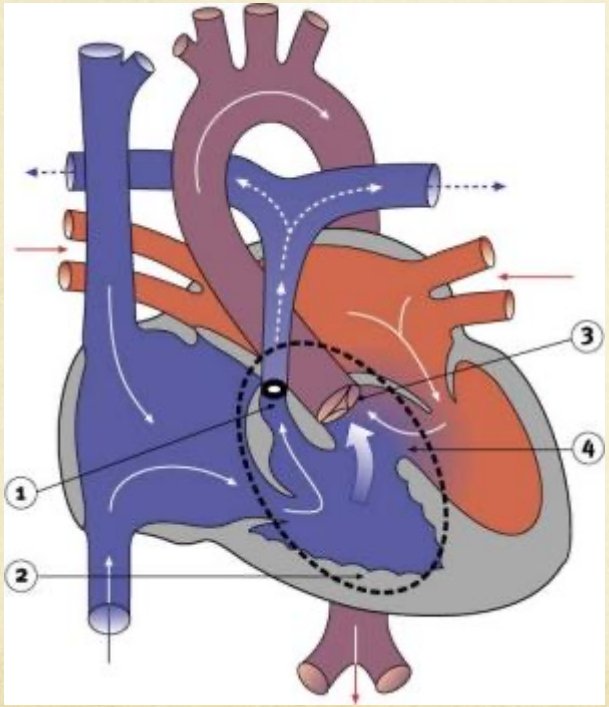
D-TGA (Egg on a string)

Cyanotic Heart Defects

- TOF (Tetrology of Fallot)
  - Ejection Systolic Murmur from birth and cyanosis
  - May present at birth or weeks later
  - Full surgical repair usually at 4 – 6 months old

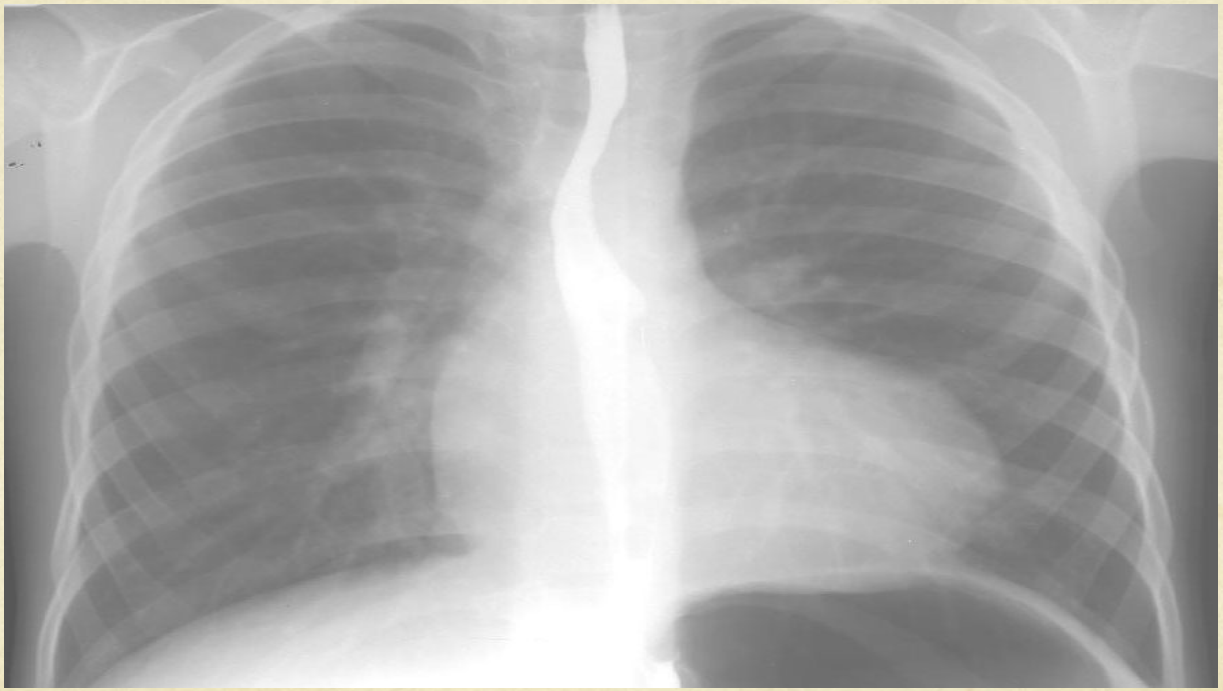
    Sometimes full repair as neonate or just a BT shunt

Tetrology of Fallot



- 1- Pulmonary Stenosis
- 2 - Right Venticular Hypertrophy
- 3 - Overriding Aorta
- 4 - VSD

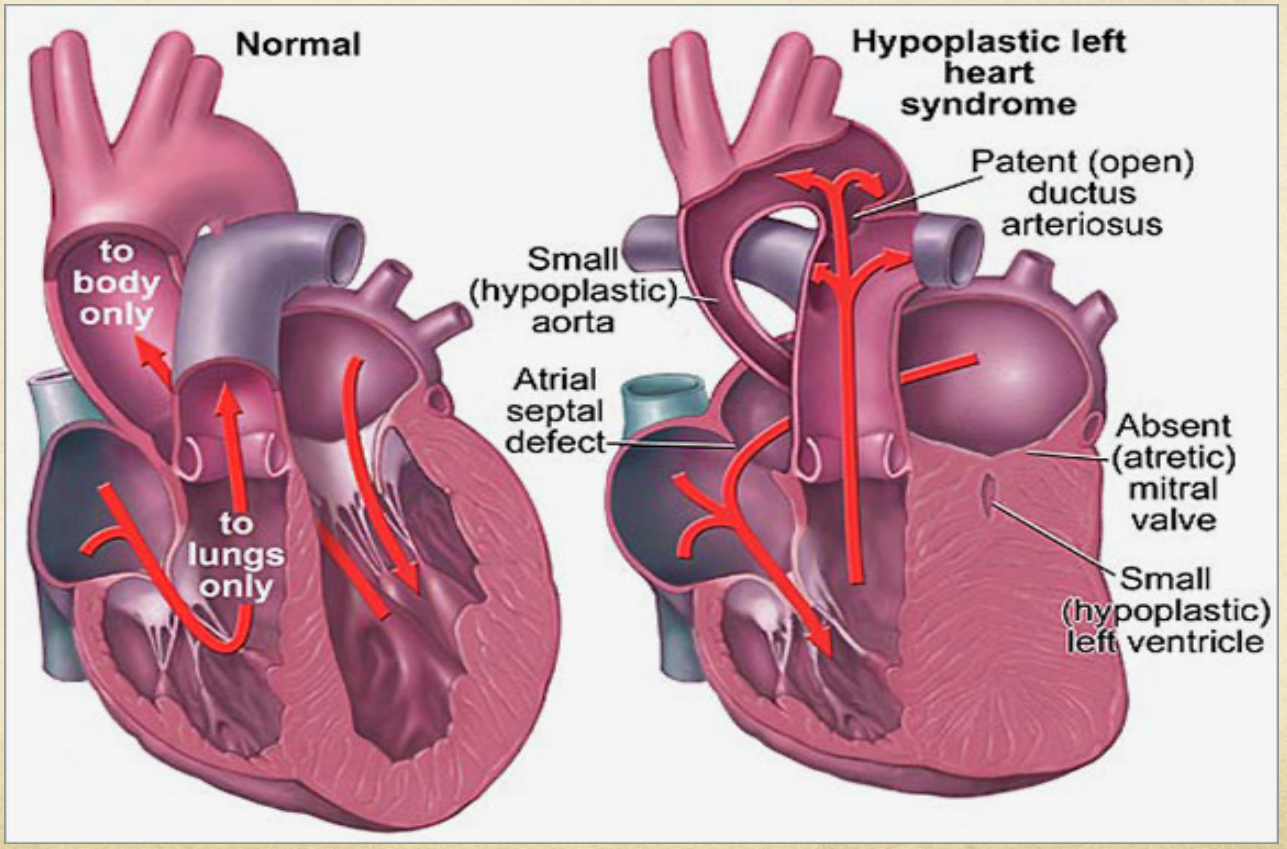
TOF (Boot shaped Heart)



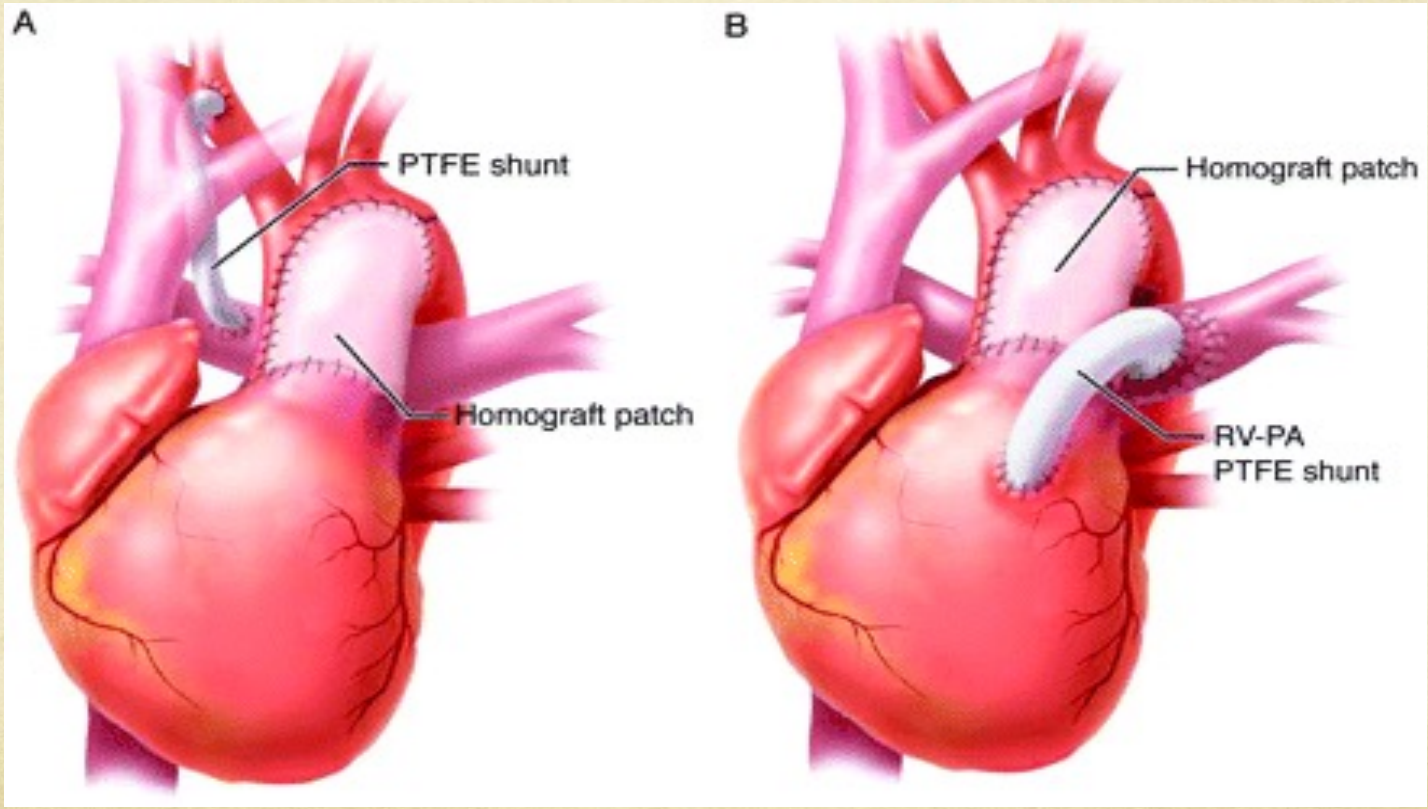
HLHS

- HLHS (Hypoplastic Left Heart Syndrome)
  - Cyanotic (often with sats in high 80's/low 90's)
  - Usually tachypneic
  - Can present in shock (ductal dependent Systemic BF)
  - Start prostin ASAP
  - Presents as neonate
  - 3 stage surgical palliation
    - Norwood – 1st week of life
    - Bidirectional Glenn – 4 months old
    - Fontan – 2 to 4 years old

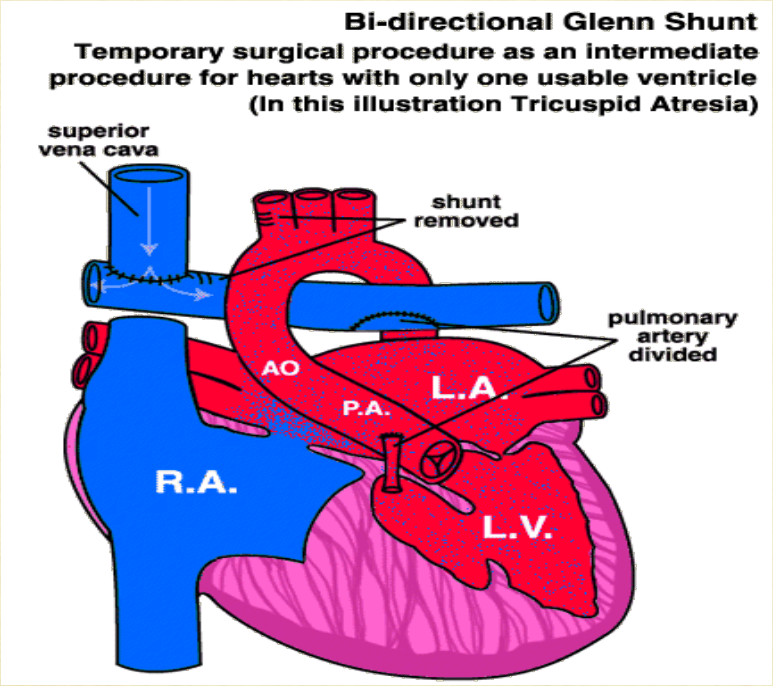
Normal HLHS


Normal
to body only
to lungs only
Hypoplastic left heart syndrome
Patent (open) ductus arteriosus
Small (hypoplastic) aorta
Atrial septal defect
Absent (atretic) mitral valve
Small (hypoplastic) left ventricle

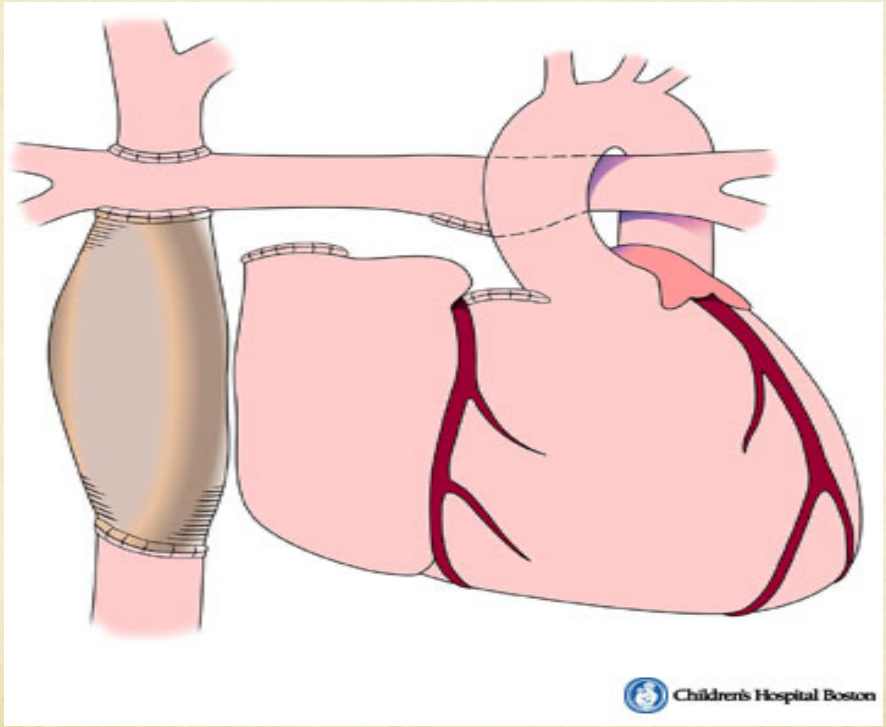
BT shunt vs Sano



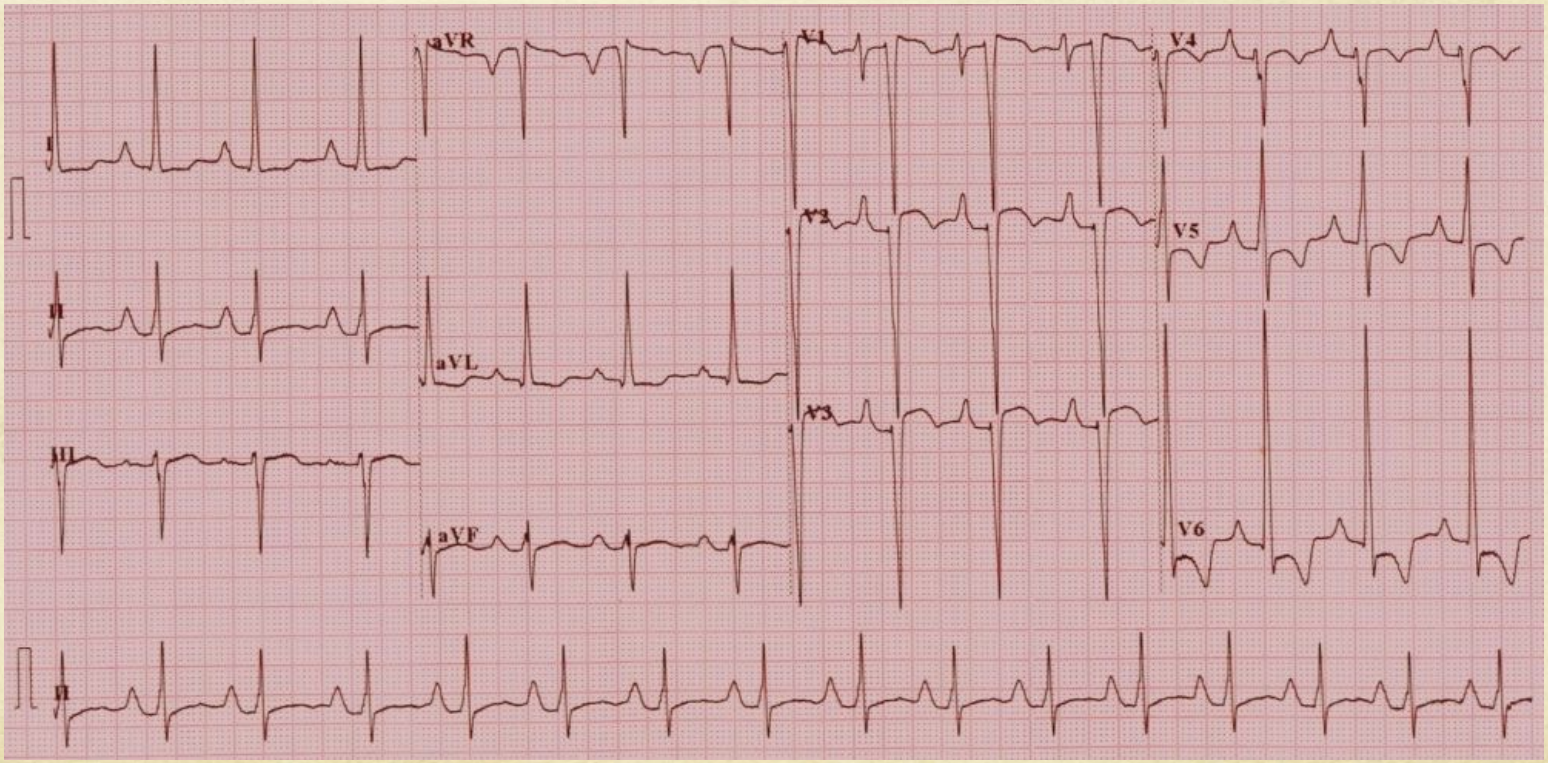
BDG


Bi-directional Glenn Shunt
Temporary surgical procedure as an intermediate procedure for hearts with only one usable ventricle (In this illustration Tricuspid Atresia)
superior vena cava
shunt removed
pulmonary artery divided
AO
P.A.
L.A.
R.A.
L.V.

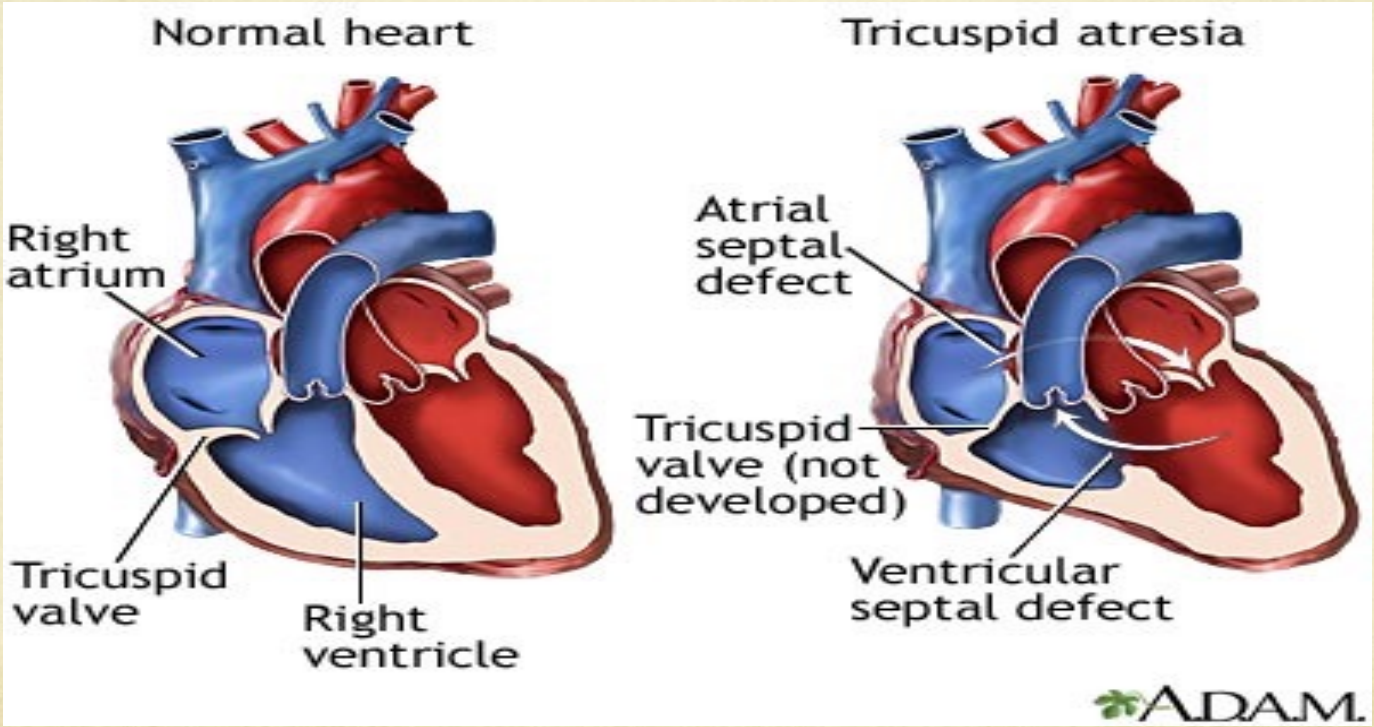
Extracadiac fontan



Superior Axis (blue with a murmur)



Tricuspid Atresia



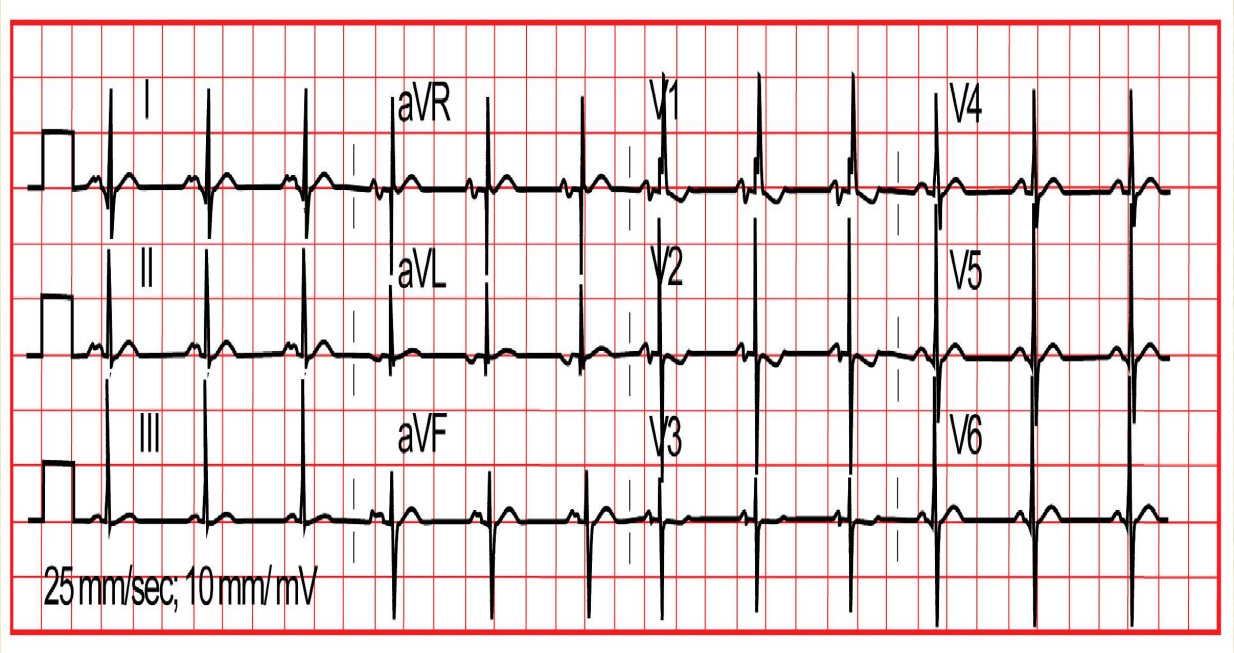
Down Syndrome

(Atrioventricular Septal Defect)

Syndromes

- Down Syndrome
  - VSD, Atrioventricular septal defect
  - Pink, holosystolic murmur
  - Superior axis on EKG (negative in lead AVF)

Superior Axis (Pink with murmur)



Williams Syndrome

Supravalvar Aortic Stenosis

Syndromes

- William Syndrome
  - Supravalvar Aortic Stenosis
  - Pink, ejection systolic murmur
  - Autosomal Dominant

Turner's Syndrome

Coarctation of the Aorta and Aortic Stenosis

DiGeorge Syndrome (VCF/22q1.1)

TOF/IAA/Truncus (conotruncal defects)

Charge Syndrome

Noonan's Syndrome

HCM, PS, ASD

Syndromes

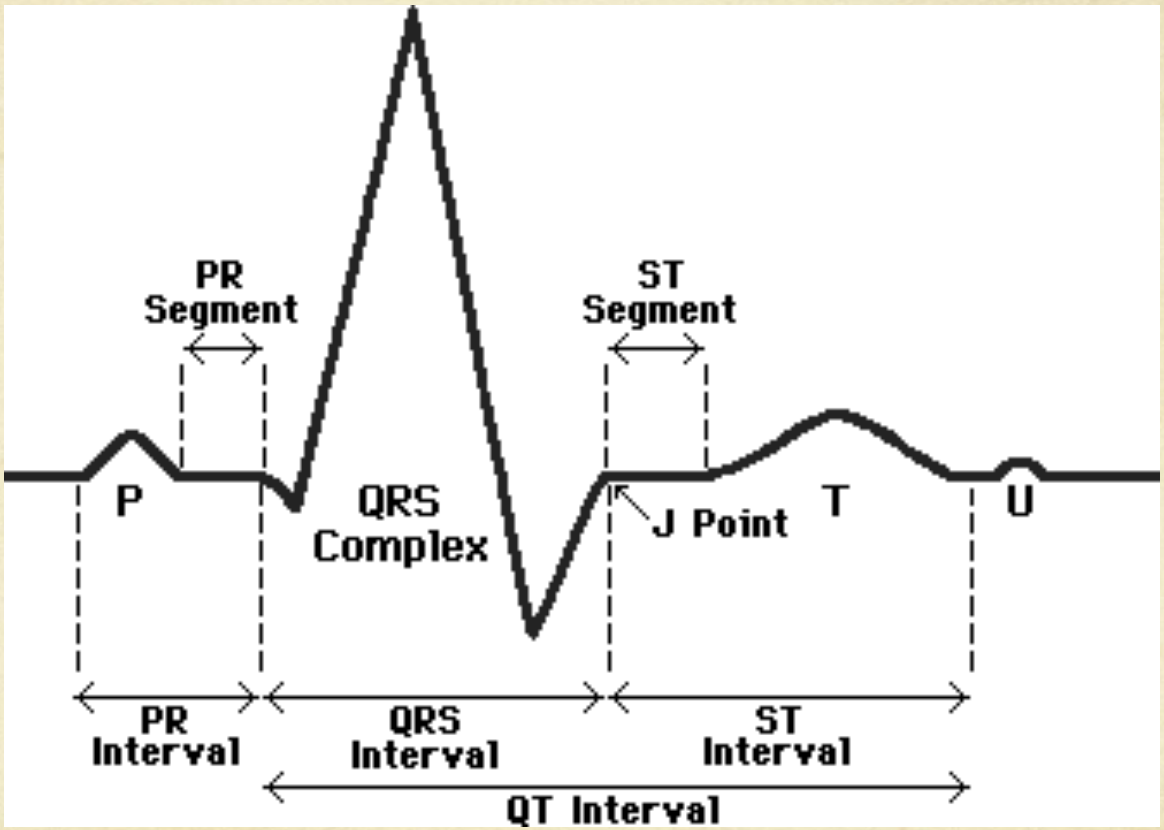
- Turner's Syndrome – Coarctation of the Aorta, AS
- DiGeorge Syndrome/22q1.1 del - TOF, IAA, Truncus
- CHARGE Syndrome – Wide variety
- Noonan Syndrome – HCM, PS, ASD (Autosomal Dominant)

Other Genetic diseases

- Tuberous Sclerosis – cardiac rhabdomyomas
- Marfans – aortic root aneurysm, MVP (AD)
- Romano-Ward – LQTS (AD)
- Jervell & Lange-Nielsen – LQTS & Deafness (AR)

Scenarios

- LQTS (Long QT syndrome)
  - QTc > 460ms (QT / square root or previous RR)
  - Alarm clock, swimming pool
  - Common treatment is Beta blocker
  - May need automatic internal cardiac defibrillator (AICD)
    - Refer to pediatric cardiology
- Maternal Diabetes
  - Neonate with hypertrophic cardiomypathy
  - Usually gets better with time
- Fetal Echo
  - Can miss VSDs, Coarctaion of the Aorta


PR
Segment
ST
Segment
P
QRS
Complex
J Point
T
U
PR
Interval
QRS
Interval
ST
Interval
QT Interval

Rheumatic Fever

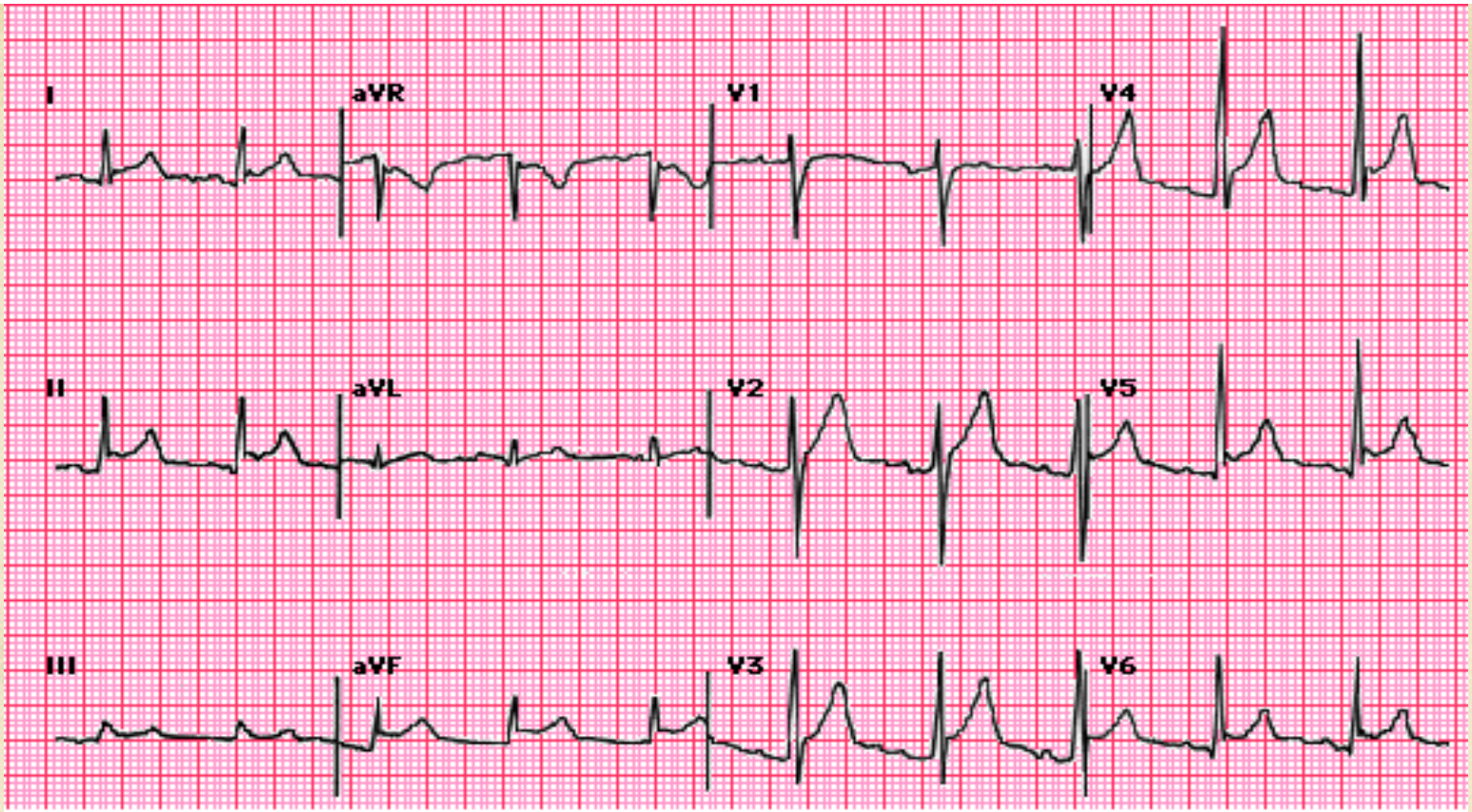
- History of recent group A strep pharygitis
  - Polyarthritis
  - New Murmur (MR, AR)
  - Sydenham's chorea
  - Erythema marginatum
  - Subcutaneous nodules
- Treatment
  - Aspirin for arthritis relief and Penicillin course
  - Penicillin for prophalaxis to prevent repeat infection

Kawasaki Disease

- Need fever x 5 days or more and 4 out of 5 criteria below:
  - Eyes - Non-exudative conjunctivitis
  - Lips /mouth – erythema, cracked lips or strawberry tongue
  - Neck – unilateral cervical lymphadenopathy
  - Hands and feet – edema/erythema – eventually peeling
  - skin
- Treat with ASA and IVIG
  - Decreases risk of coronary artery anerysms
  - Echo to look at coronaries and ventricular function
    - Initial echo for baseline and rpt in 2 – 3 wks

Chest Pain

- Benign if:
  - Reproducible to palpation
  - Worse with deep breathing
- Red flags
  - happens with exercise!!!!
  - If yes - cardiology referral and not allowed to exercise
- If worse when lying down think pericarditis
  - Friction rub and possible history of recent URI



Pericarditis Electrocardiogram in acute pericarditis showing diffuse upsloping ST segment elevations seen best here in leads II, III, aVF, and V3 to V6. There is also subtle PR segment deviation (positive in aVR, negative in most other leads). ST segment elevation is due to a ventricular current of injury associated with epicardial inflammation; similarly, the PR segment changes are due to an atrial current of injury which, in pericarditis, typically displaces the PR segment upright in lead aVR and downward in most other leads. Courtesy of Ary Goldberger, MD.

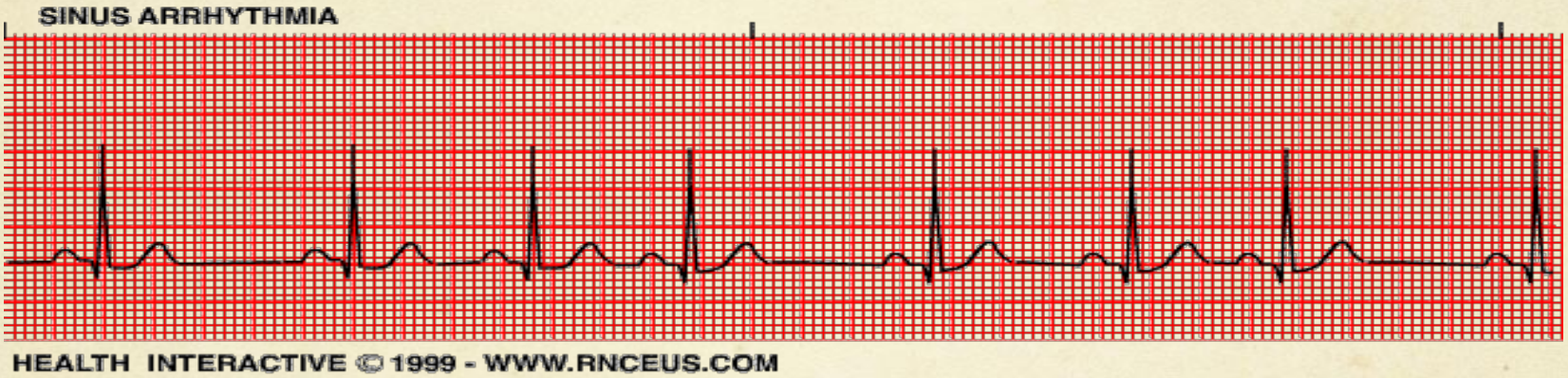
Syncope

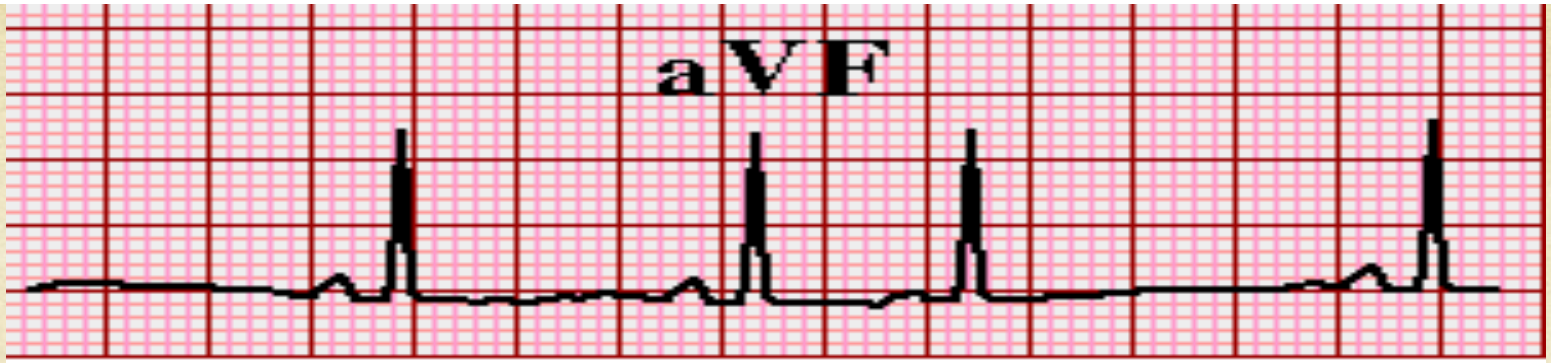
- Vasovagal Syncope (benign)
  - Caused by pain, fear, standing too long
  - Treat with increasing salt and fluids in diet
- Syncope (malignant)
  - Happens with exercise
    - Coronary abnormality
    - Hypertrophic cardiomyopathy
  - Fast onset (arrhythmia) – patient usually falls and gets hurt
  - Long QT syndrome
  - Refer for cardiology consult (no exercise until evaluation)

EKGs

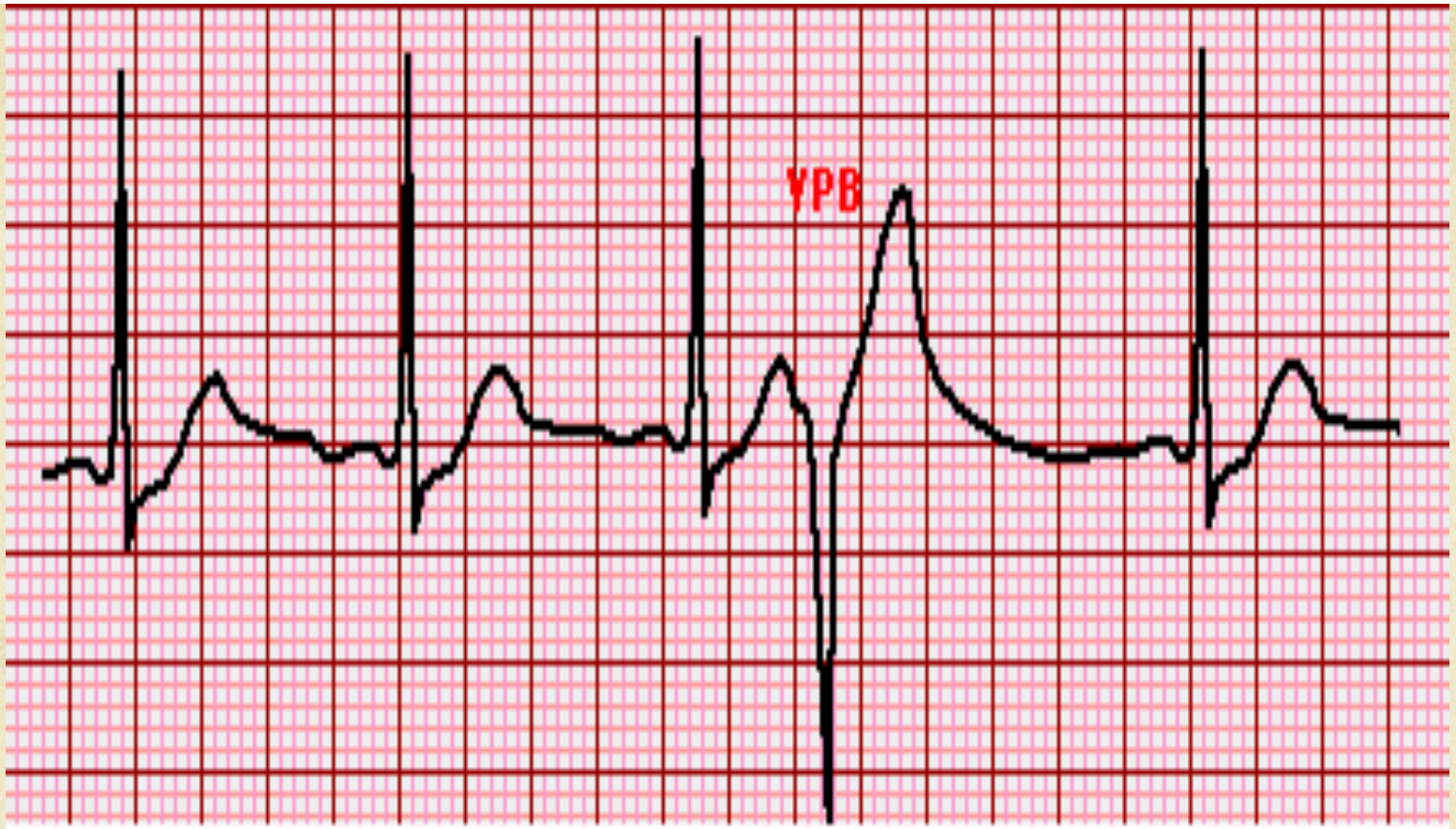
- Sinus arrhythmia – due to respiratory variation
- Premature atrial contraction (PAC or APB)
- Premature ventricular contraction (PVC or VPB)

Sinus Arrythmia





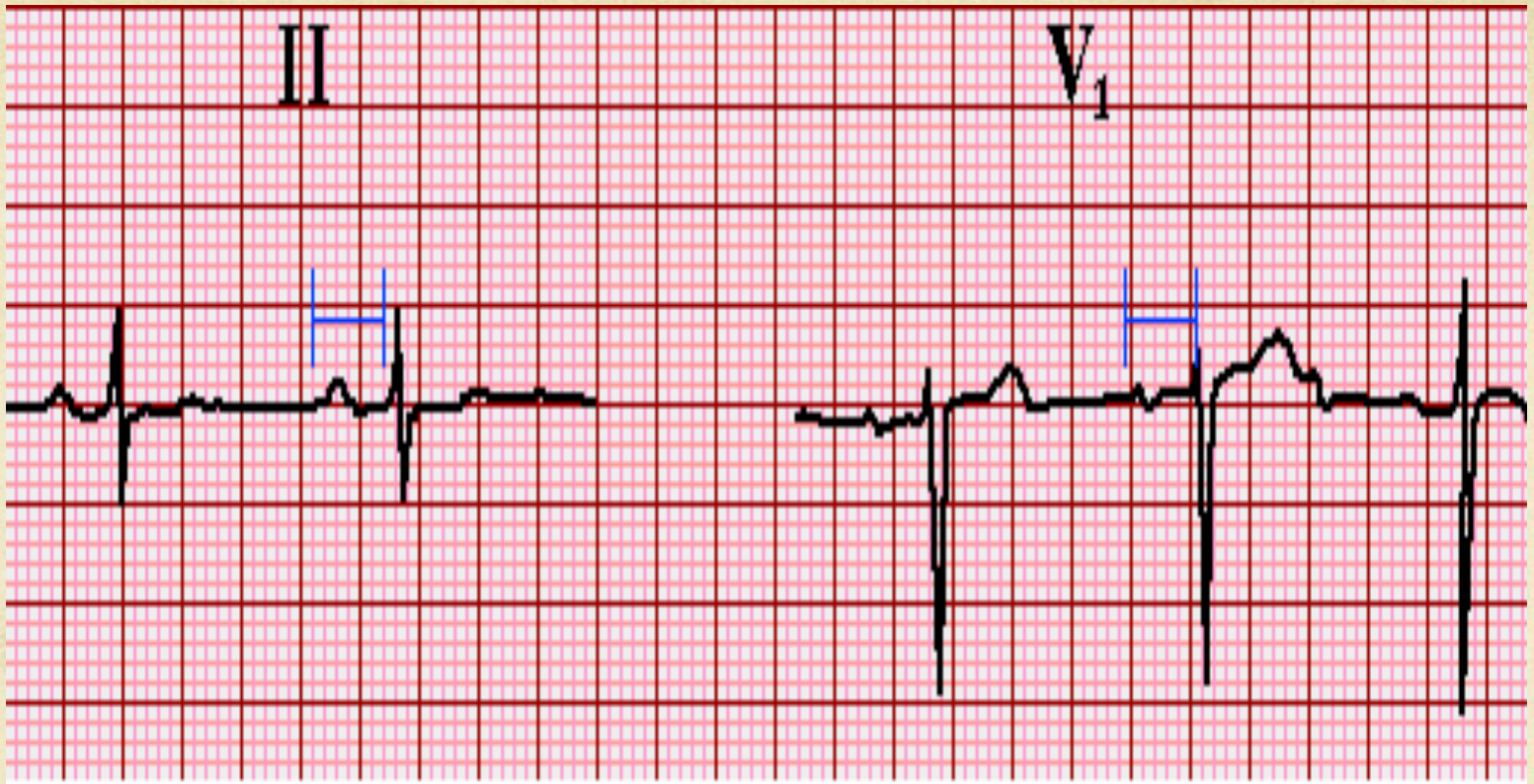
Atrial premature beats The third beat is an atrial premature beat that is normally conducted. The interval between the second sinus beat and the ectopic beat is shorter than the interval between the first two sinus beats. The P wave morphology differs from that of sinus rhythm. Since the RR cycle length is shorter, there is a decrease in the rate of conduction of the ectopic beat through the atrioventricular node. The PR interval is therefore longer than that of the sinus beat. Activation of the ventricular myocardium occurs in a normal fashion; as a result, the QRS complex is unchanged from that of sinus rhythm.



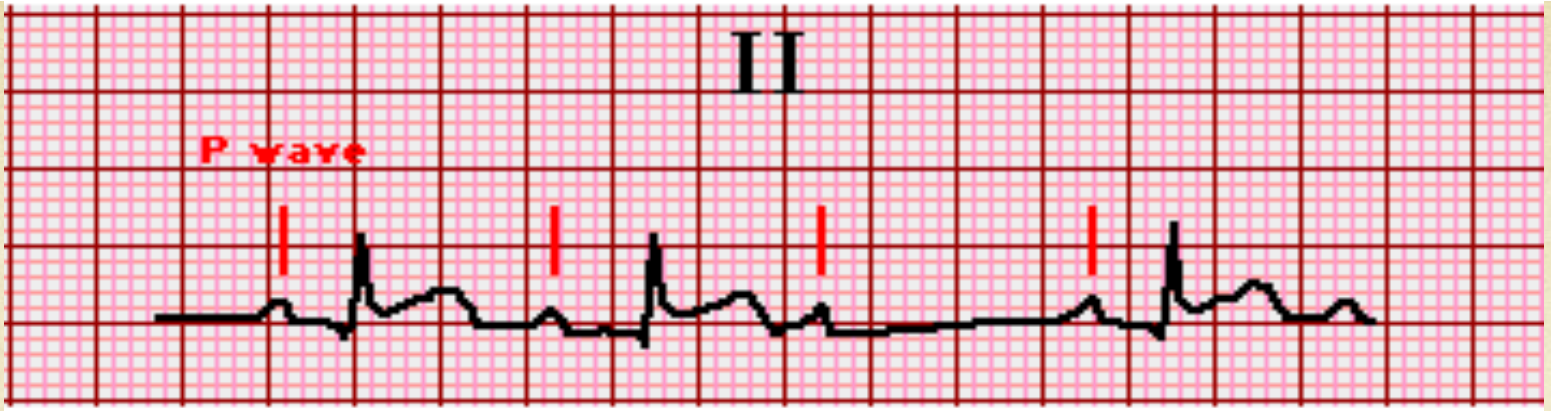
Ventricular premature beat The fourth beat is a ventricular premature beat (VPB). It has a wide, bizarre morphology, with a duration >0.16 seconds.

EKGs

- AV Block (1st 2nd and 3rd degree)

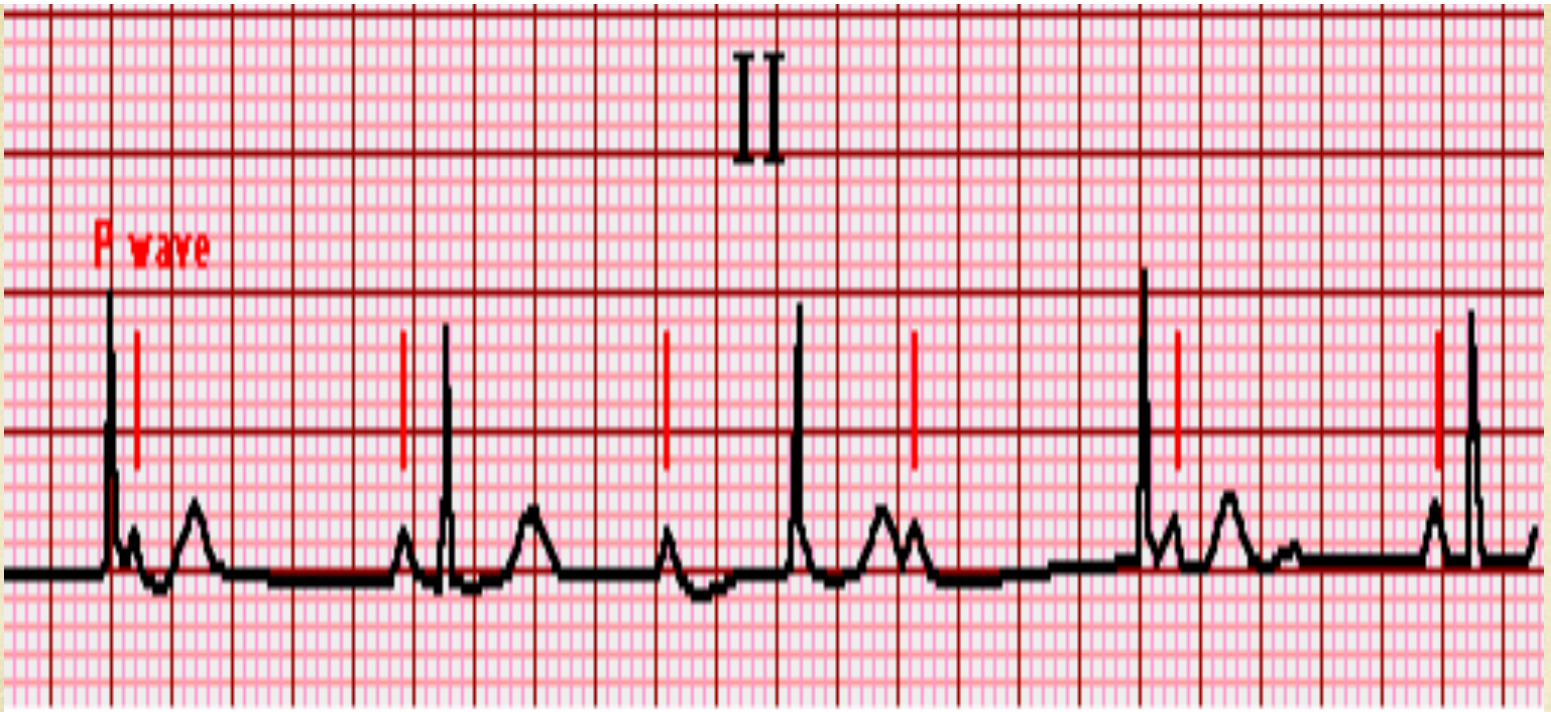


First degree atrioventricular (AV) block First degree AV block is caused by a prolongation or delay in impulse conduction through the AV node. It is defined as a PR interval >0.20 seconds. In this case the PR interval (blue lines) is approximately 0.22 seconds.



Mobitz type I second degree atrioventricular (AV) block Several characteristic features of Wenckebach second degree AV block are noted on this ECG (P waves are marked by arrows): 1) The PR interval after the second P wave is longer than the preceeding PR interval; the third P wave is not conducted at all. 2) The PP intervals are constant; however, the RR interval surrounding the completely blocked P wave is longer than the preceeding normal RR interval. 3) The fourth P wave (after the blocked beat) is conducted normally; this PR interval is shorter than the PR interval that immediately preceeded the pause.

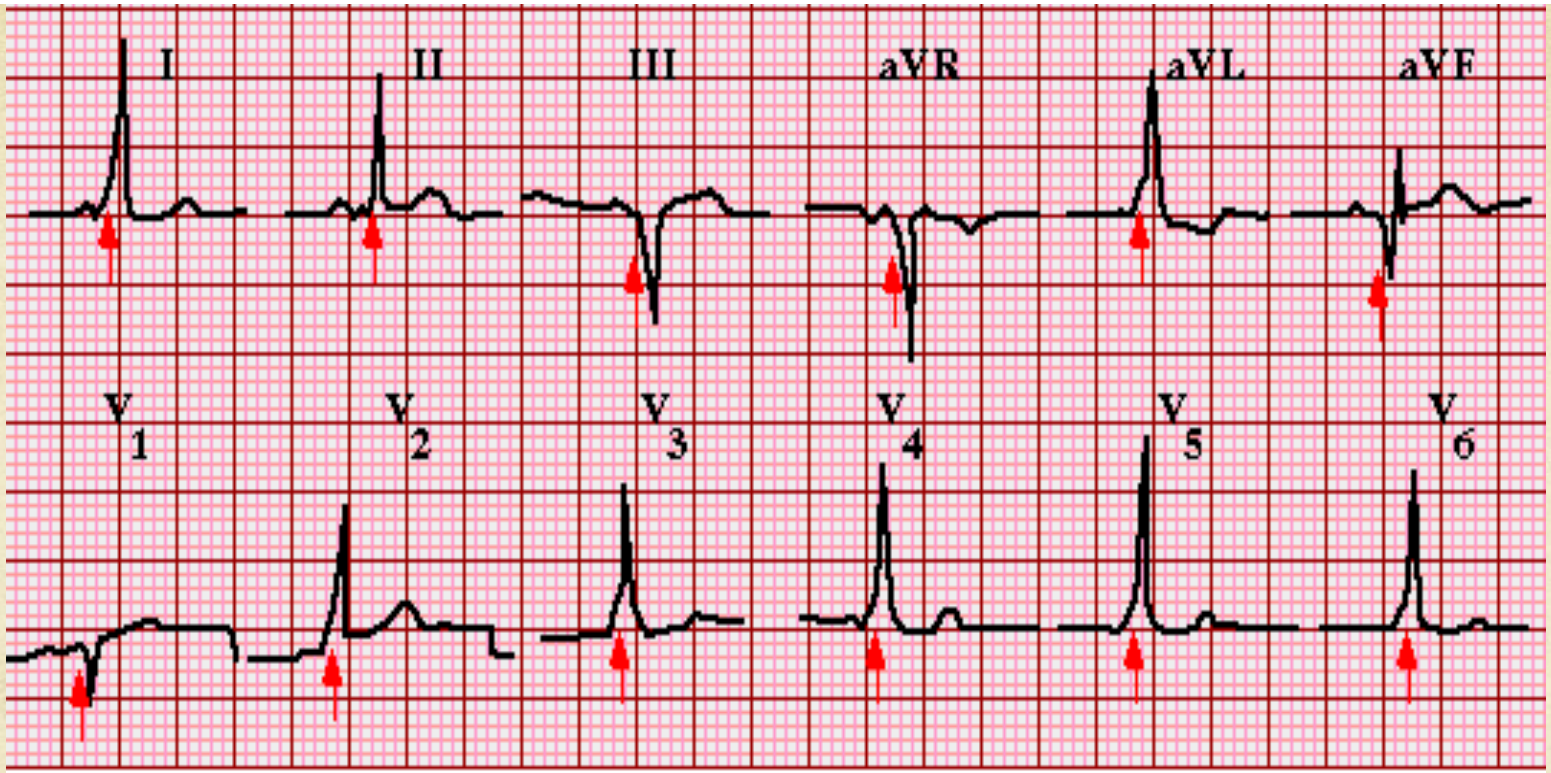
Mobitz type II second degree atrioventricular (AV) block The third and sixth P waves are not conducted through the AV node (there is no associated QRS complex). The PR interval is constant prior to and after the non-conducted beats.



Third degree atrioventricular block The P waves are completely dissociated from the QRS complexes. The QRS complexes are narrow, indicating a junctional escape rhythm. The atrial and ventricular rates are stable; the former is faster than the latter.

EKGs

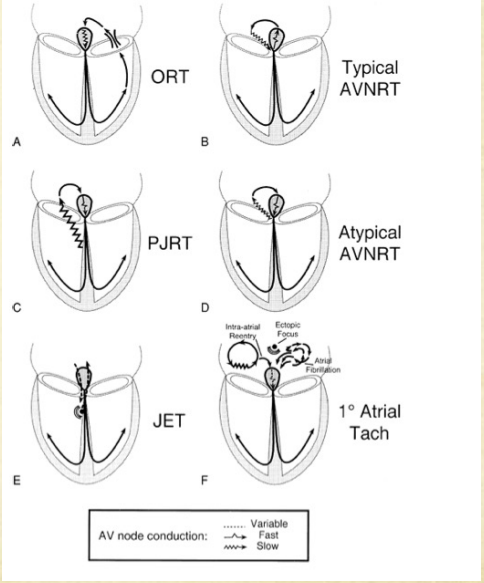
- WPW
  - Delta wave (pre-excitation)
  - Short PR
  - SVT
  - Treat Beta-blocker not with Digoxin



Wolff-Parkinson-White syndrome The two main electrocardiographic features of Wolff-Parkinson-White syndrome (WPW) include a short PR interval (<0.12 seconds) and a delta wave (red arrows). The QRS complex is wide (>0.12 seconds) and represents a fusion beat; the initial portion (delta wave) results from slow ventricular activation via the accessory pathway, while the termination of ventricular activation is via the normal conduction system leading to a fairly normal terminal portion of the QRS.

Atrial Tachyarrythmias

- Re-entrant Tacycardia
  - Fast on / Fast off
    - Abrupt onset / termination
  - AVRT, AVNRT, AFlutter
- Ectopic Tachcardias
  - Warm up and cool off
    - Gradually gets faster
    - Gradually gets slower
  - JET, EAT, Sinus Tach



Supra Ventricular Tachycardia

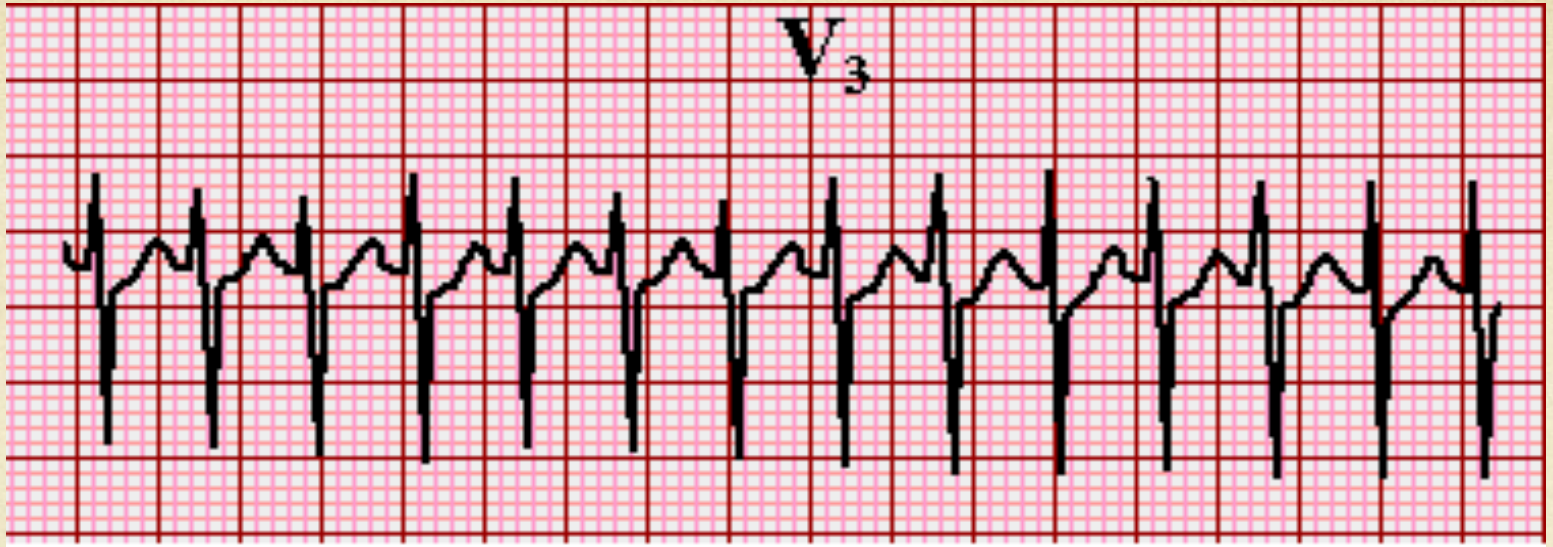
Reentrant tachycardia

- Narrow complex, fast (>200), not variable
- Abruptly starts and abruptly stops
- Treatment
  - Vagal maneuver – ice, temperature, PIV
  - Adenosine IV given as a **push** – very fast!!!
    - Very short half-life, so must push fast
    - Preferable in upper extremity
    - Can cause VFib

SVT: Presentation

- Infants
  - Poor feeding
  - Pallor
  - S+S of CHF
- Older Children
  - Palpitations
  - Chest pain/discomfort

SVT HR > 200 in children

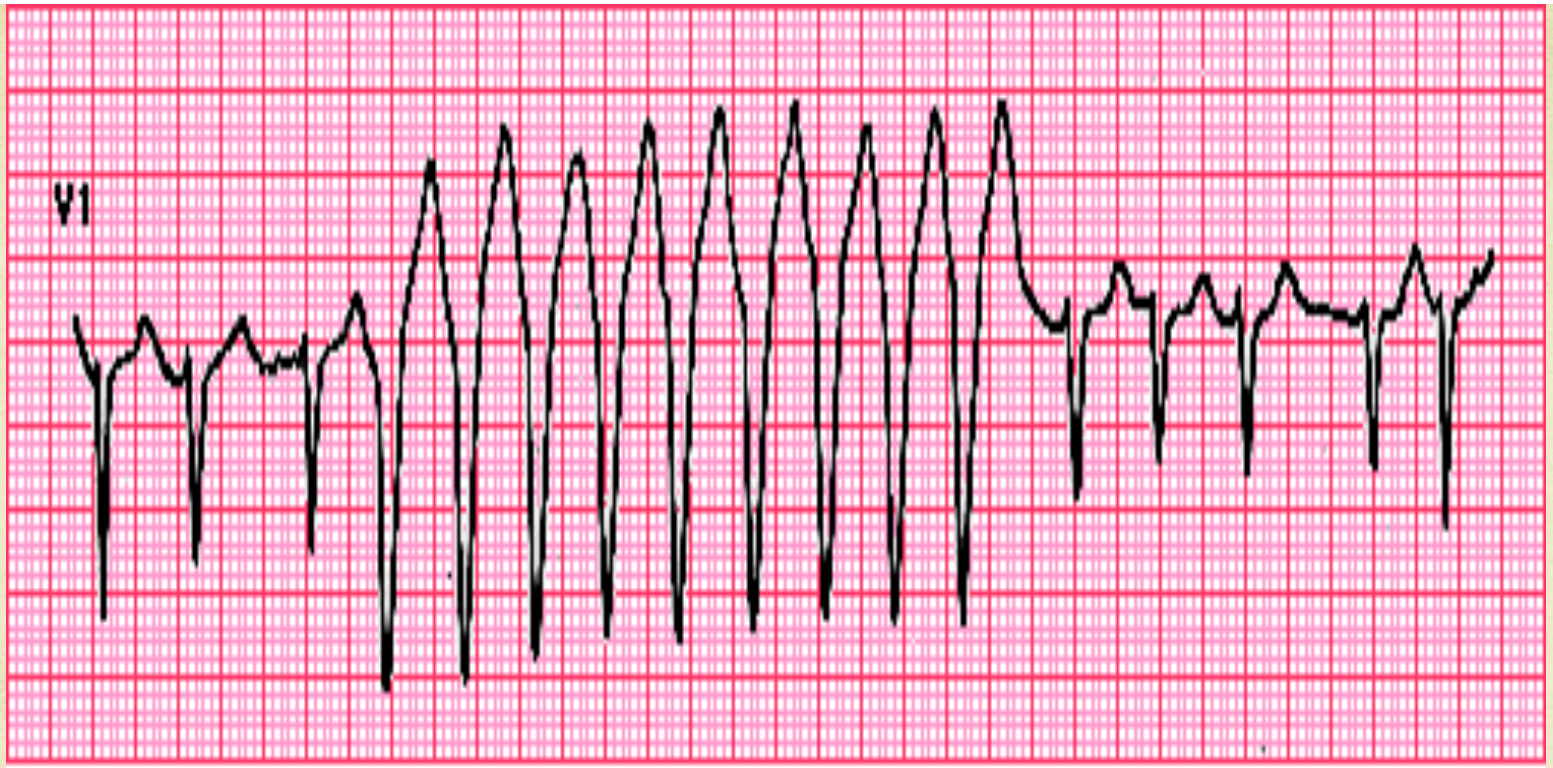


AV nodal reentrant tachycardia Junctional tachycardia or atrioventricular nodal reentrant tachycardia is a supraventricular tachyarrhythmia that originates within the atrioventricular node. The rate is generally between 140 to 220 beats per minute and there is usually a 1:1 atrial-ventricular association; as a result, every QRS complex has an associated P wave. In this case, atrial and ventricular activation are simultaneous; as a result, the P wave is superimposed upon the QRS complex

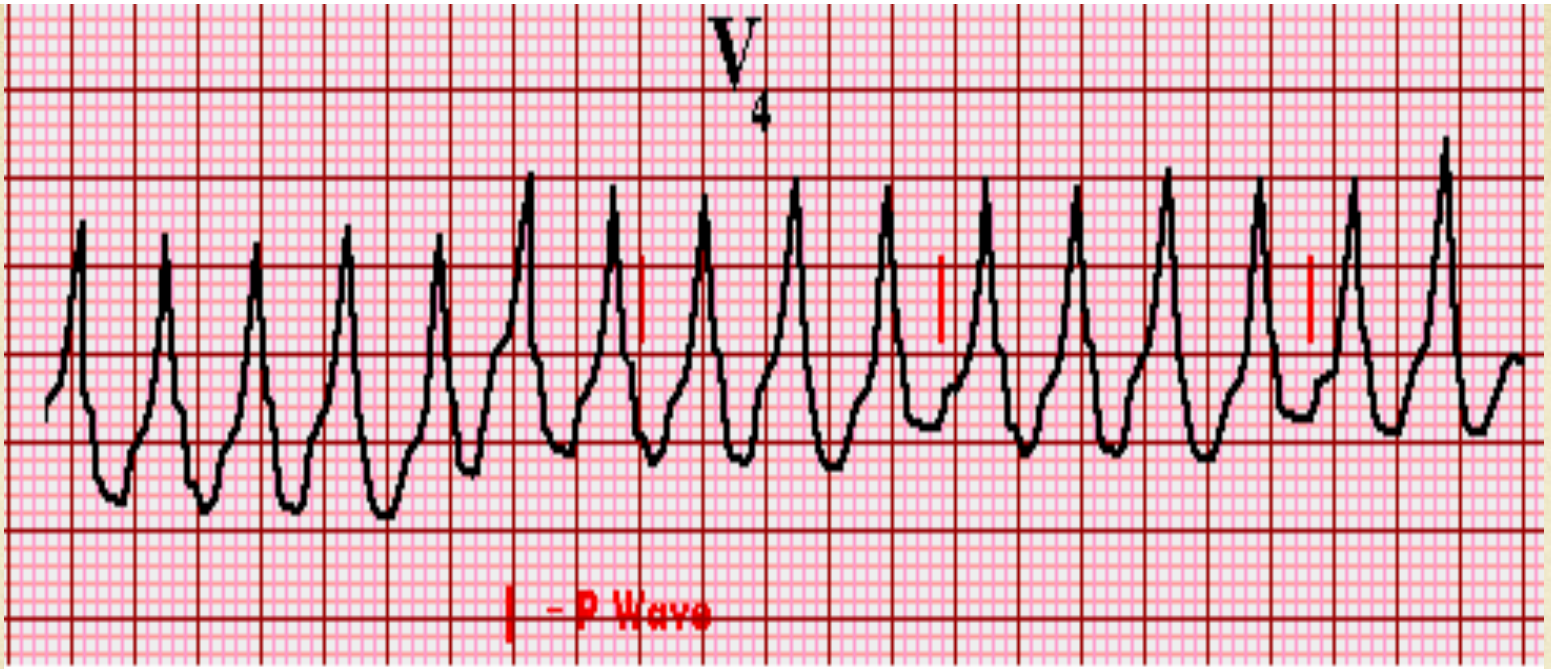
Ventricular Arrythmias

- Pulseless Rythyms
  - Too fast to fill the heart and maintain a BP
  - Ventricular Tachycardia
  - Ventricular Fibrillation

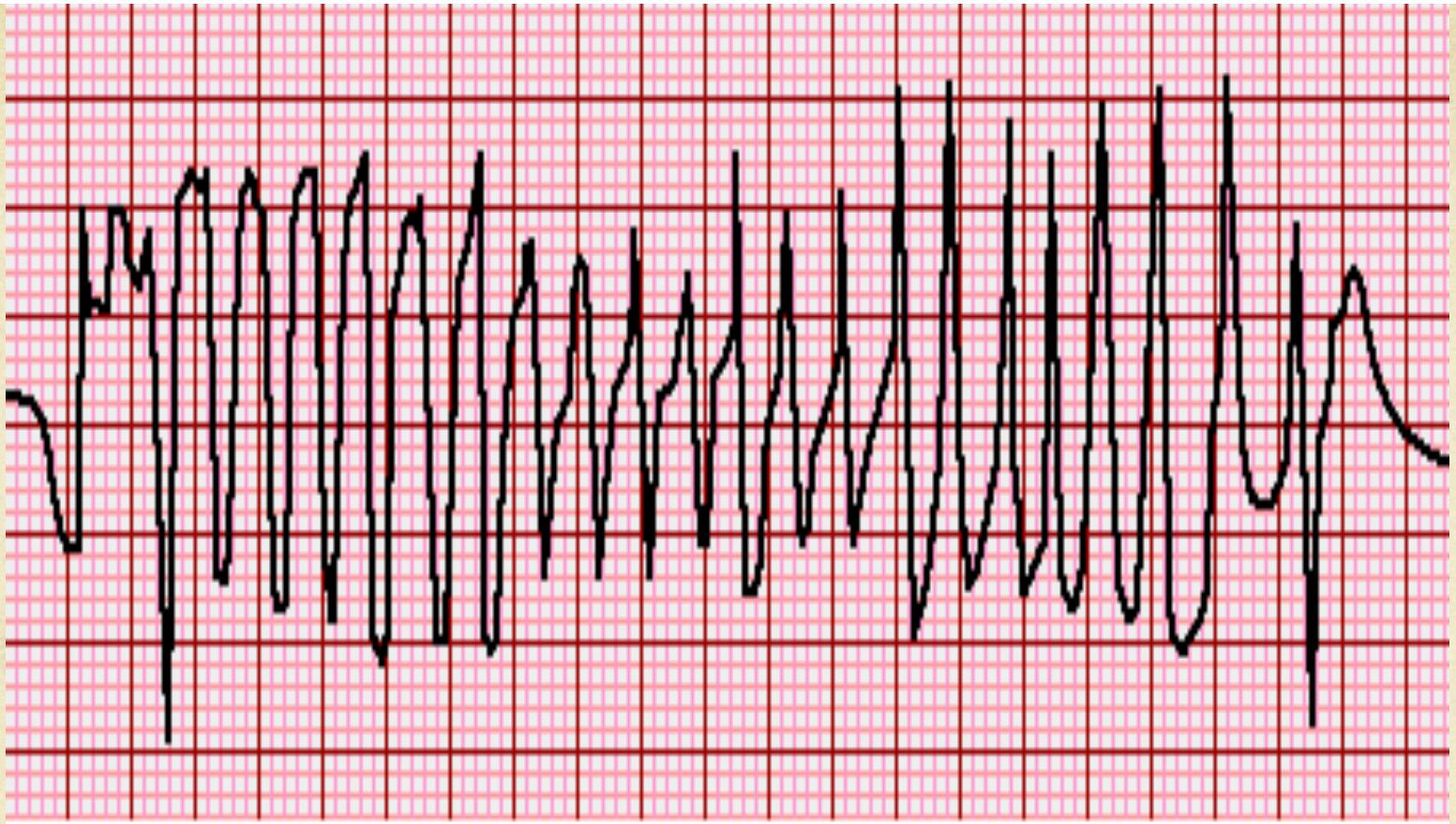
- Treatment
  - Asynchronous Defibrillation



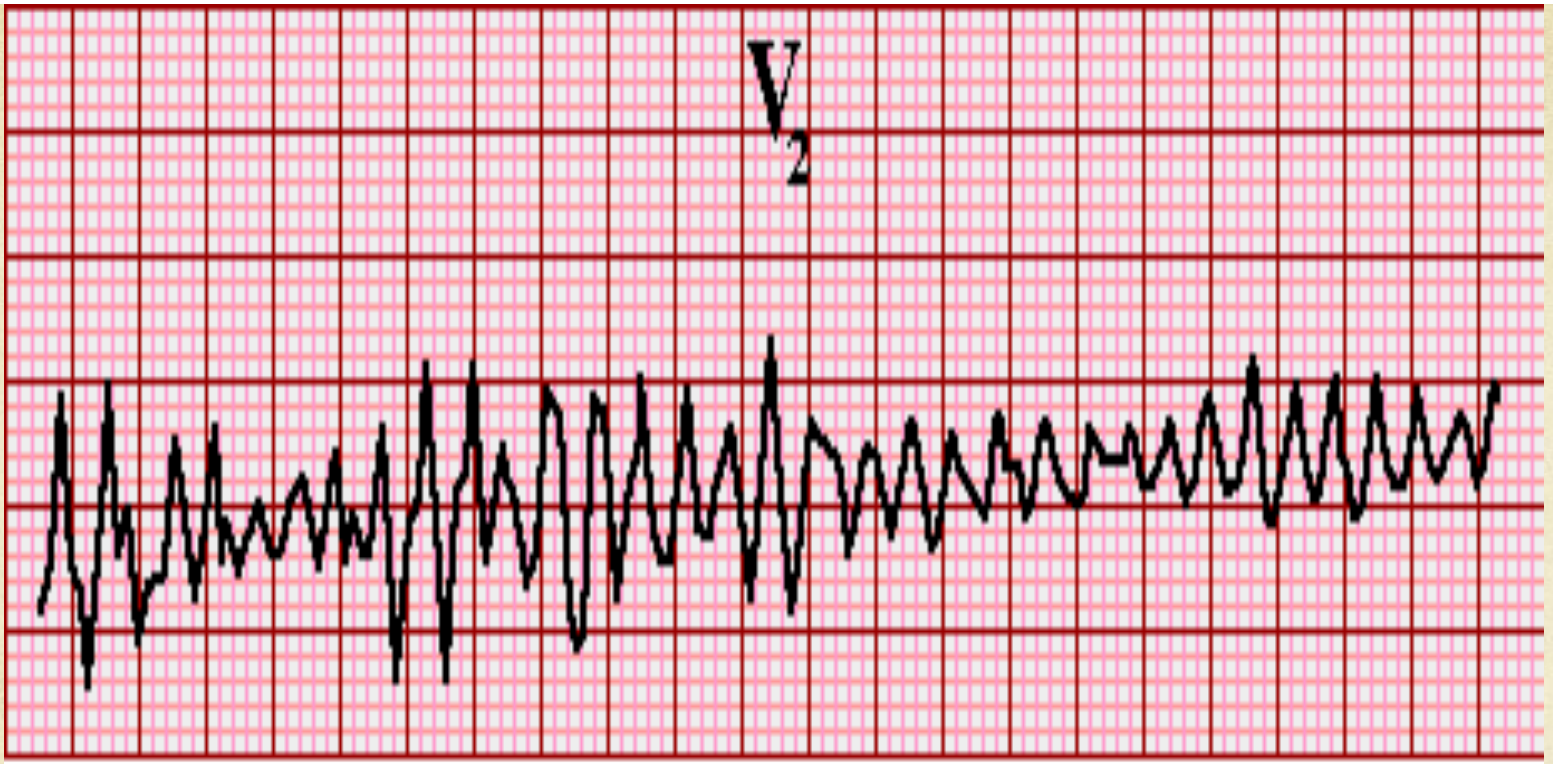
Nonsustained ventricular tachycardia Nonsustained ventricular tachycardia in a patient with underlying atrial fibrillation. The ventricular arrhythmia consists of nine beats at an approximate rate of 170 beats/min. Courtesy of Ary Goldberger, MD.



Monomorphic ventricular tachycardia Three or more successive ventricular beats are defined as ventricular tachycardia (VT). This VT is monomorphic since all of the QRS complexes have an identical appearance. Although the P waves are not distinct, they can be seen altering the QRS complex and ST-T waves in an irregular fashion, indicating the absence of a relationship between the P waves and the QRS complexes, i.e. AV dissociation is present.



Polymorphic ventricular tachycardia The QRS complexes have markedly differing morphologies due to changes in the direction (vector) of myocardial activation.



Ventricular fibrillation There is a complete absence of properly formed QRS complexes and no obvious P waves. A recent onset (eg, within minutes) of the arrhythmia is suggested by the coarse morphology of the fibrillatory waves.

Antiarrhythmic drug classification

- Class I – Na channel blocker
  - Class IA: Quinidine, Procainamide
  - Class IB: **Lidocaine**
  - Class IC: Propafenone, Flecainide
- Class II-Beta-blockers (propranolol, esmolol)
- Class III-K channel blocker (**Amiodarone**)
  - Adverse reactions of Amiodarone
    - Hypotension, Torsade, QT prolongation, Severe bradycardia, AV block
    - Hypo/hyperthyroidism, pulmonary toxicity, blue-grey skin discoloration
- Class IV-Ca channel blocker (Verapamil, Diltiazem)
  - Do not use if under one year old

Some Tips

- Stay away from answers with **Always** and **Never**
- Referring patient is usually not a good answer
- Start with the most basic test or study
- They love to give you normal patients that just need reassurance (always consider this as an option)

Thank you

- Good Luck
- Any questions contact me at jason.katz@mch.com

Question #1

- 13 year male seen in your office for a history of chest pain. He explains the chest pain is located in the left side of his chest, no radiation and resolves spontaneously. It is worsens with deep breathing and is reproducible on your exam. He also explains his grandfather had a myocardial infarction at the age of 62. Of the following, the MOST appropriate approach for this patient at this time is
- A. Cardiac catheterization
- B. Reassurance with clinical follow-up
- C. Echocardiography
- D. Electrocardiogram

Question #2

- Newborn baby born full term via NSVD who at 12 hours of age is noted to be mottled and tachypneic. Upon arrival in the NICU, his arterial blood gas shows a bicarbonate of 12 and a po2 of 42. Of the following, the best management plan is
- A. Order an echocardiogram
- B. Start a prostin infusion
- C. Give a 10cc/kg bolus of normal saline
- D. Start an infusion of epinephrine

Question #3

- A two month old is seen in your office for a well baby visit. You note he has a loud holosystolic murmur heard best at the left midsternal border. His mom states he is now taking longer to finish his feeds and seems to be sweating and breathing harder with his feeds. He has gained no weight over the past month. Of the following, the best management plan is
- A. Start propranolol
- B. Admit to the hospital for immediate surgery
- C. Reassure mother that baby is OK
- D. Start furosemide

Question #4

- A six month old male seen in the ER for a history of poor feeding for the past 24 hours. The baby appears to be tachypneic and pale. Once the baby is placed on a telemetry monitor it is noted the baby's heart rate is 240bpm. The blood pressure and oxgen saturation are normal. Of the following, the best management plan is
- A. Start furosemide
- B. Synchronized cardioversion
- C. Place an IV and give adenosine rapidly
- D. Start propranolol

Question #5

- A 14 year old girl is found unconscious in a swimming pool after diving into the pool. She is taken out and found to be in pulseless ventricular fibrillation. She is shocked back into normal sinus rhythm and is brought to the hospital in good condition. Of the following, the appropriate diagnostic study to start with is
- A. Echocardiogram
- B. Electrocardiogram
- C. Chest x-ray
- D. Cardiac caheterization