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Disorders of the Cardiovascular System

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Disclosures

Teresa Whited, DNP, APRN, CPNP-PC

- Has no financial relationship with commercial interests
- This presentation contains no reference to unlabeled/unapproved uses of drugs or products

Learning Objectives

Upon completion of this review, the course attendee should be able to:

- Describe the process of history and physical assessment of the cardiovascular system.
- Summarize common diagnostic tests (laboratory and radiology) utilized when evaluating a cardiovascular concern.
- Compare and contrast the pathophysiology, clinical presentation, management, and follow-up of the most common cardiovascular diagnoses seen in primary care.
- Describe education needs related to the most common cardiovascular diagnoses.

Presentation Outline

- Key points in History and Physical exam
- Murmurs
- Classification of heart defects
- Heart Failure
- Acquired Heart Disease

History

- Review of family, maternal, fetal, neonatal, and infant medical history
- Review of known risk factors (maternal, neonatal, and newborn)
 - Genetic syndromes, maternal diabetes, febrile illnesses, drug/alcohol use
- **Deviation from expected G&D** (weight first or significantly less than length, FOC)
- Deviation from expected activity level-syncope, **exercise intolerance**
- Frequent respiratory tract infections or **chronic cough** that will not resolve
- Murmurs, cyanotic episodes
- Hypertension
- Chest pain, palpitations, or **syncope with exertion**
- SOB with exertion (beyond comparable peers)
- **Family history of sudden death, congenital heart disease, cardiomyopathies, or premature MI before age 50 years**

Chromosomal Abnormalities

- Down's Syndrome
 - **AV Canal defects, VSD, TOF**
- Turner's Syndrome
 - **Coarctation of the Aorta**
- Di George's Syndrome
 - Interrupted Aortic Arch
 - **TOF**



Physical Exam

- Vital signs
- General appearance-well or chronically ill?
- Measurement of height and weight and plot on standardized growth chart
- **Oxygen saturation**
- Unusual facial characteristics or other physical stigmata associated with syndromes (Down syndrome, DiGeorge, Marfan, Turner)
- Overall skin color
- **Chest deformities (pectus)**
- Tongue, mucous membranes, clubbing (only with prolonged cyanosis)
- Wheezing, nasal flaring retractions, head bobbing
- Peripheral or periorbital **edema**
- Palpation of the chest, auscultation of heart sounds

Physical Exam

- **Inspection**

- General Appearance
- Color of skin and mucous membranes
- Pulse oximetry
- Clubbing
 - Low arterial oxygen levels > 6 months
 - Angle lost between nail and nail bed and base of nail bed feels soft and spongy
- Respiratory Rate and effort



Cyanosis



Acrocyanosis vs. Cyanosis



Acrocyanosis vs. Cyanosis

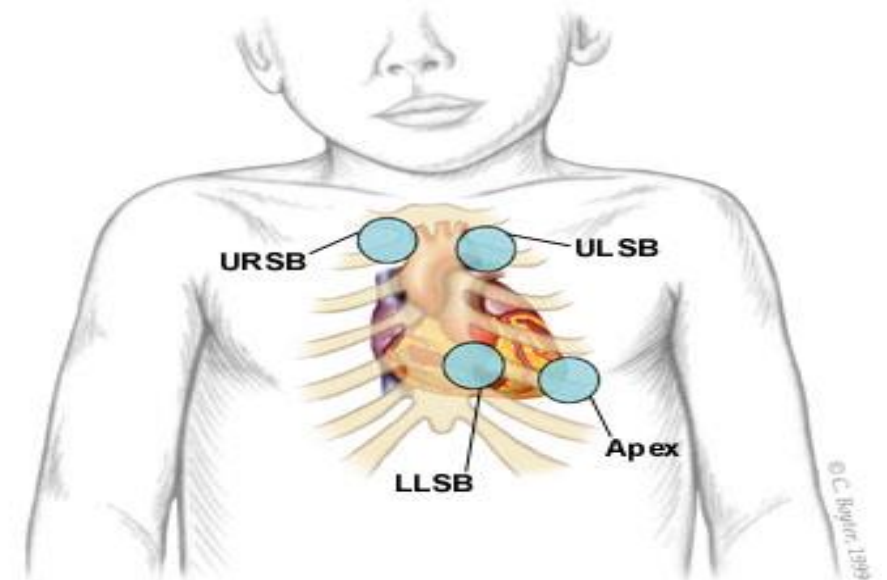


Palpation

- **Pulses, Pulses, Pulses**
 - Equal, diminished or bounding, no-delay
- **Apical Impulse**
 - Location and quality
 - **Cardiomegaly**
- **Precordial Thrill**
 - Fine vibration with palm of hand that correlates with murmur
- Heave/Lift

Auscultation







- Aortic area= 2nd RSB
- Pulmonic area= 2nd LSB
 - Erb's Point/P2= 3rd LSB
- Tricuspid Area = 4th LSB
- Mitral Area = 4-5th MCL/Apex



Heart Sounds

- The presence of **S3** can be present with CHF or high output states in children (Ken-tuck-y)
- The presence of **S4** is almost never normal (Ten-nes-see)
- **A fixed S2 split**
 - ASD
 - Pulmonic Stenosis

Characteristics: Intensity

- Grade 1  • Barely Audible
- Grade II  • Soft, but easily audible in all positions
- Grade III  • Moderately audible, no thrill
- Grade IV  • Louder with palpable thrill
- Grade V  • Stethoscope slightly off chest, thrill
- Grade VI  • Stethoscope off chest

Pitch of murmurs

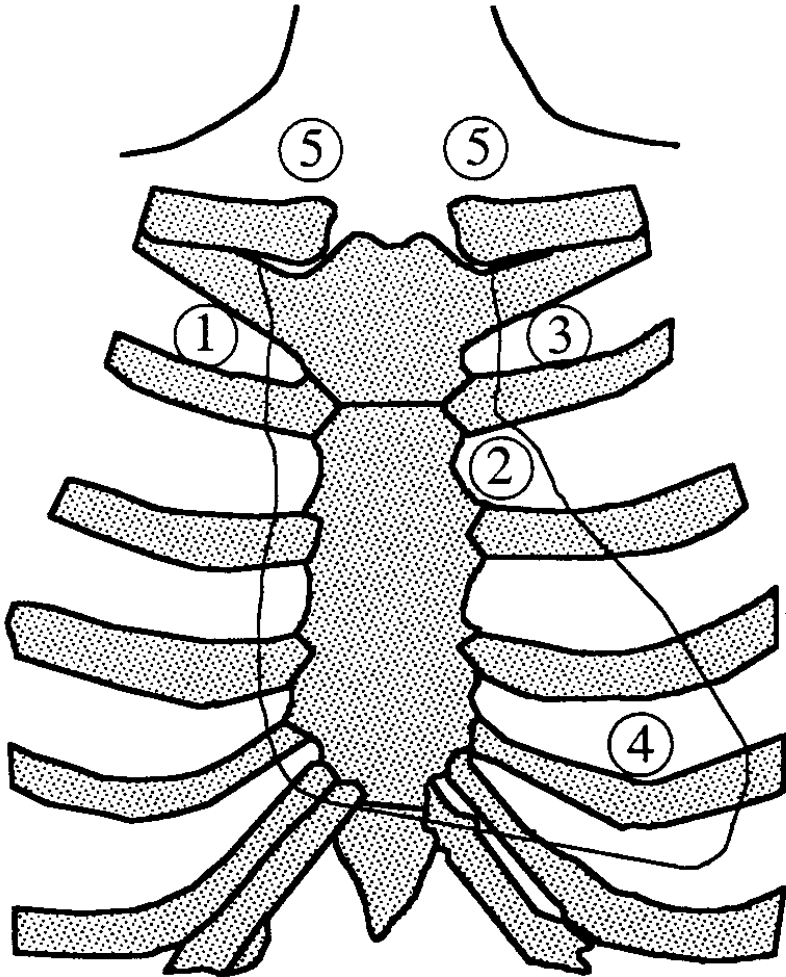
- High vs Low
- Holes vs valves



Innocent Heart Murmurs

- **Systolic**
- **Low pitch, vibratory, twangy**
- Loudest at LLSB or 2nd or 3rd ICS
- Usually soft
- **Changes with positioning**
- Heart loudest lying down & after exercise
- Normal pulses, RR & BP
- **Normal growth and development**

Kids and Murmurs



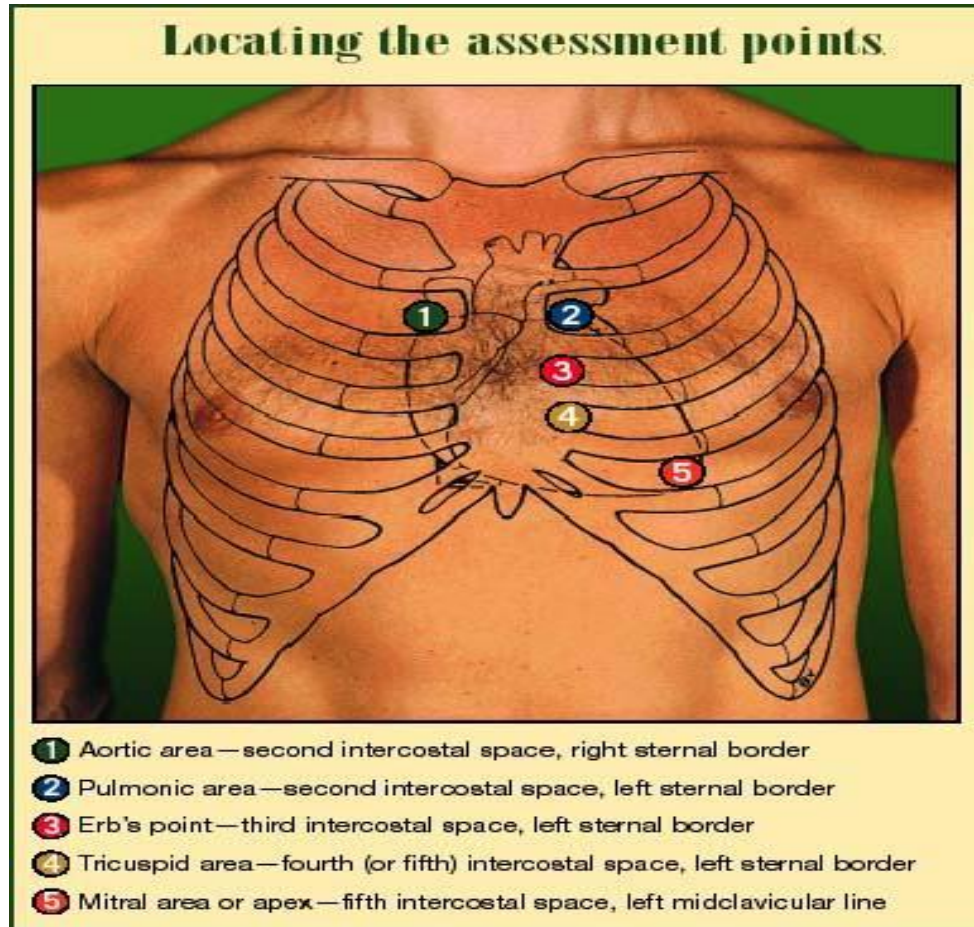
- Normal Murmurs

- 1) Venous hums
- 2) Pulmonary flow – RV to MPA
- 3) MPA to branch PA
- 4) Still's murmur – LV to Ao
- 5) Supraclavicular or carotid bruit

Pathologic Heart Murmurs

- Rough, coarse, harsh
- Loudest at base of heart
- Usually at least a **Grade III**
- **Diastolic**
- **Does not change** with positioning
- Tachycardia, tachypnea
- Failure to thrive/grow

Location of Common Murmurs



1. Aortic stenosis
2. PDA, PS, ASD
3. PS, ASD
4. TS, VSD
5. VSD, MR

Common Tests

- CXR
- ECG
- Echo
- CBC
- Cardiac Cath
- MRI
- Stress testing

Categories of Disorders

- Congenital Heart Disease
 - Acyanotic
 - Obstructive Lesions
 - Cyanotic
- Acquired Heart Disease

Acyanotic Heart Defects vs. Cyanotic Heart Defects

- **Cyanotic Heart Disease**

- Requires immediate attention and evaluation
- May not be detected in the newborn nursery due to the patency of the ductus arteriosus (PDA)

- **Acyanotic Heart Disease**

- Typically presents with s/s of congestive heart failure
- More gradual clinical decompensation
- May not present until after the first 2-8 weeks of age (pulmonary vascular resistance drop)
 - Classic symptoms: tachypnea, tachycardia, hepatomegaly
 - History may include: poor or slow feeding, sweating, color change with feeding, and poor weight gain (FTT)

Question 1

The most common congenital heart defect in children is:

1. Tricuspid atresia
2. Ventricular septal defect
3. Aortic stenosis
4. Pulmonary atresia

The most common congenital heart defect in children is:

1. **Answer: Ventricular septal defect**

Most Common Congenital Heart Defects

- **Ventricular Septal Defect VSD 20%**
- **Atrial Septal Defect ASD 10%**
- **Patent Ductus Arteriosus 8-10%**
- **Tetralogy of Fallot 6-10%**
 - 4 components:
 1. VSD
 2. Pulmonic Stenosis
 3. Right Ventricular Hypertrophy
 4. Overriding Aorta

Findings Specific for Common Cardiac Defects

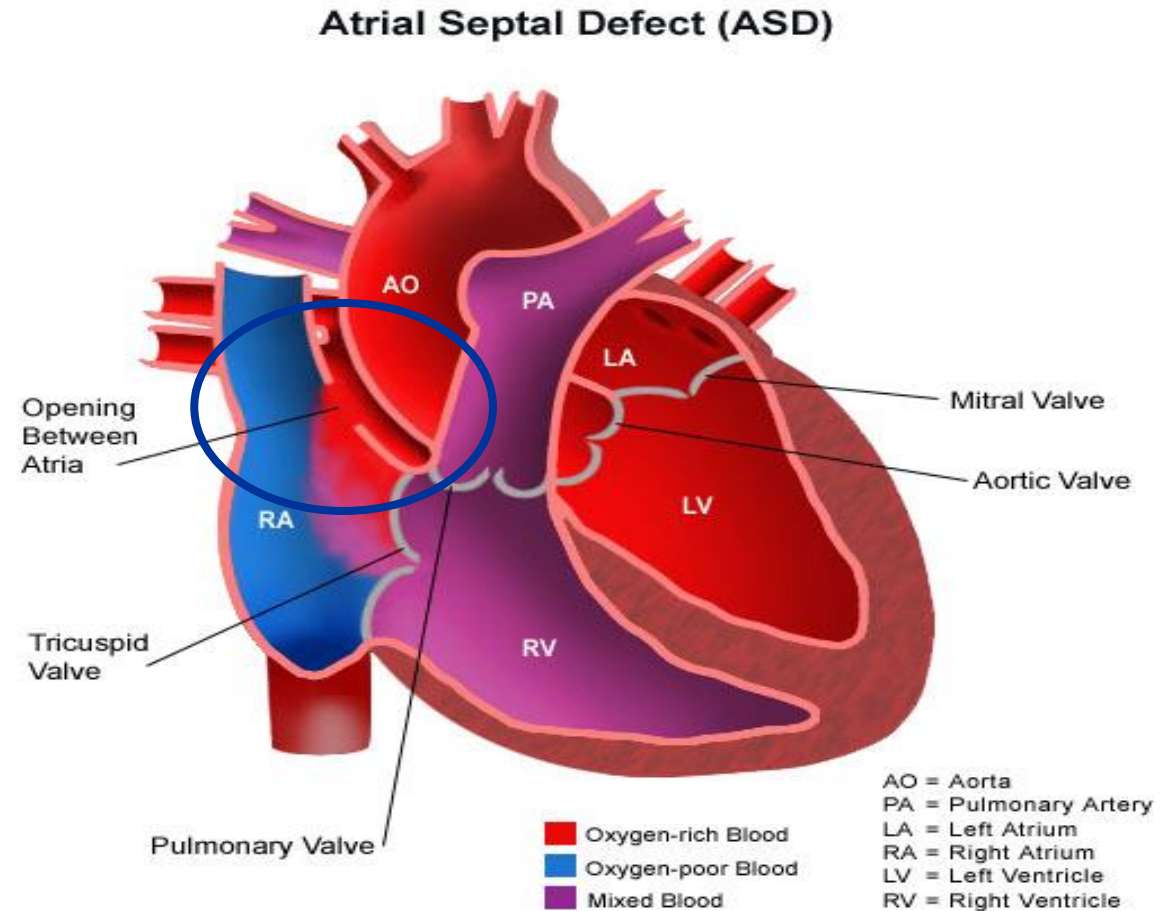
- **ASD:** Fixed S2 split
- **Coarctation of the Aorta**
 - High BP in the right upper extremity
 - Low BP in the lower extremities
 - Bounding pulses in the right upper extremity
 - Weak, absent or delayed pulses in the lower extremities

Acyanotic Lesions

- Left to Right Shunting
 - Atrial Septal Defect
 - Ventricular Septal Defect
 - Patent Ductus Arteriosus

Atrial Septal Defect

- An abnormal hole between the two atria of the heart
 - Three types
 - Most common Secundum
 - Often closed in cath lab
 - If inadequate rims or locations-surgery
- **Asymptomatic**
- Blood flows through this hole from greater pressure to lower pressure

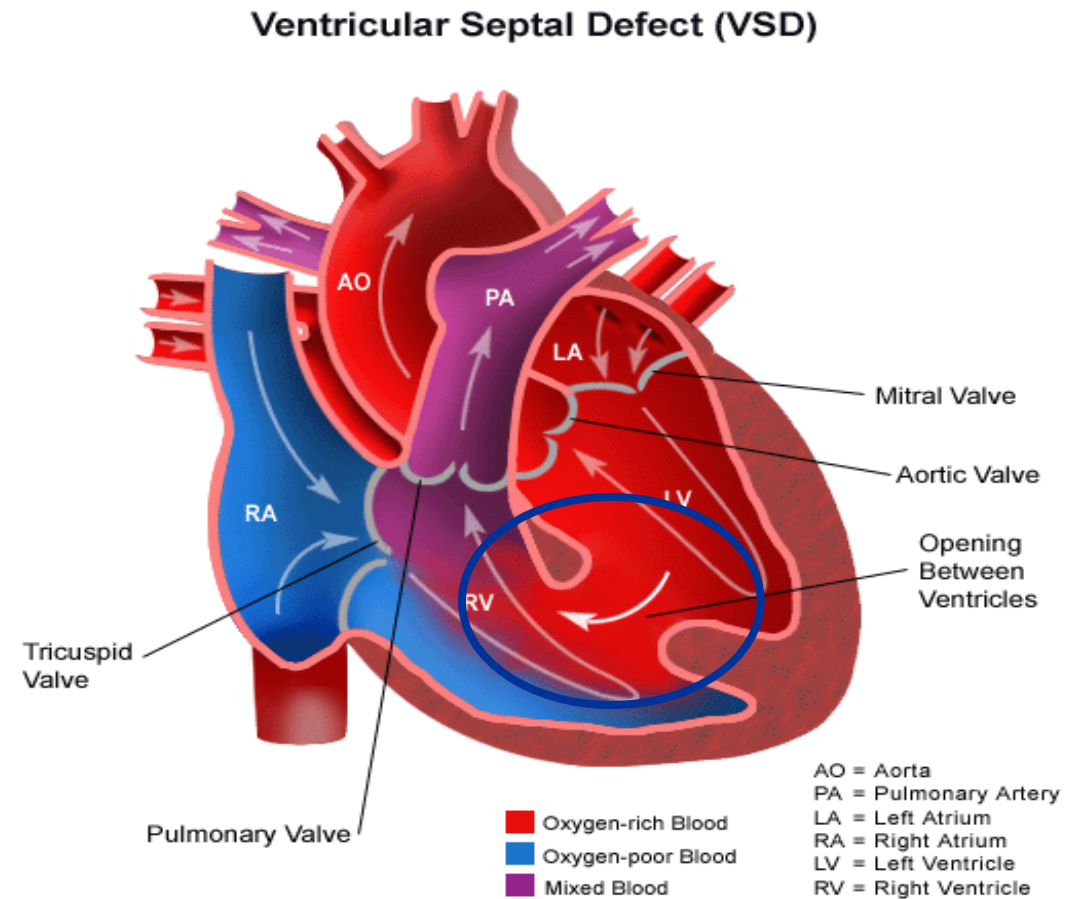


ASD Murmur

- Systolic ejection heart best at **LUSB-if present**
- Results from increased blood flow through the PA
- **Fixed S2 split**

Ventricular Septal Defect

- An abnormal hole between the ventricles (bottom chambers of the heart)
- Multiple types: Most common
 - Muscular
 - Perimembranous
- Size dictates symptoms
- Blood flows through this hole from greater pressure to lower pressure

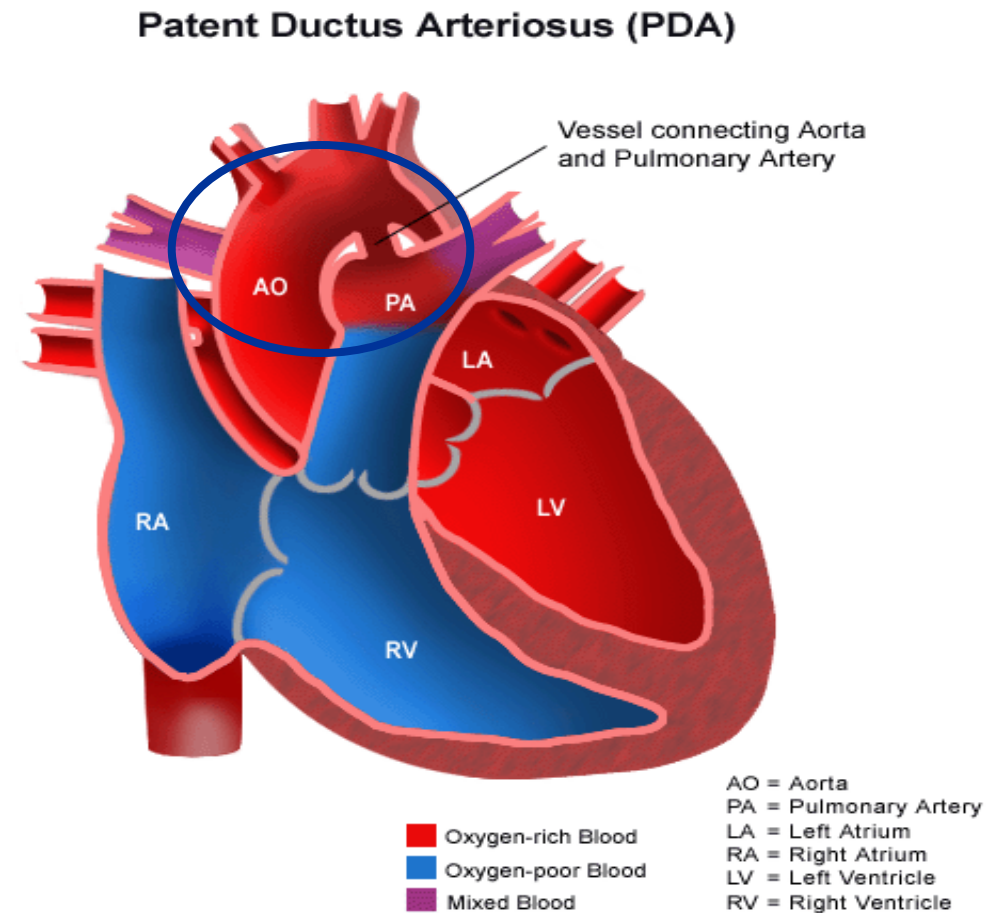


VSD Murmur

- Heard over **LLSB** and **mitral region**
- **Holosystolic** murmur
- Size dictates intensity and pitch
- 38% of pathologic murmurs

Patent Ductus Arteriosus

- Persistent connection between Pulmonary artery and Aorta
- More common in **premature infants/abnormal birth**
- Size dictates symptoms



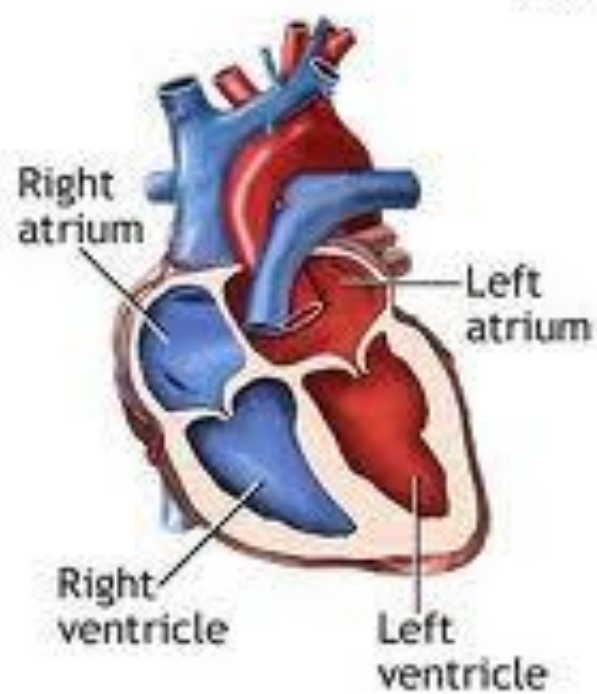
PDA Murmur

- **Continuous murmur** with systolic and diastolic qualities
- Heard over LUSB
- **Can be systolic in nature in first 1-2 days**
 - Pressures in lungs and body almost equal
- Thrill (+/-), continuous and machinery like
- 4% of pathological murmurs

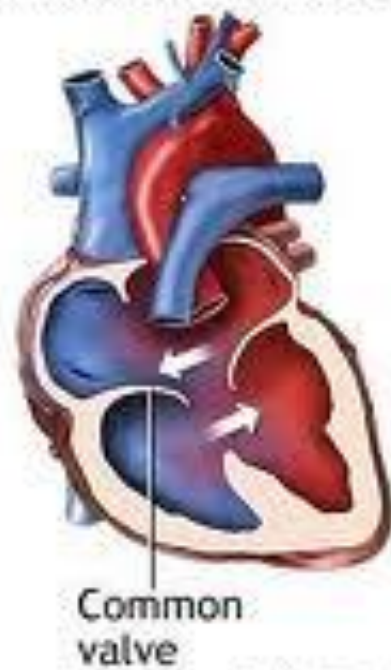
AV Canal

- AVSD, Endocardial cushion defect
- Often associated with Down's
 - 50% of Down's have heart disease
 - 50% of which is AV canal

Normal heart



Atrioventricular canal
(Endocardial cushion defect)

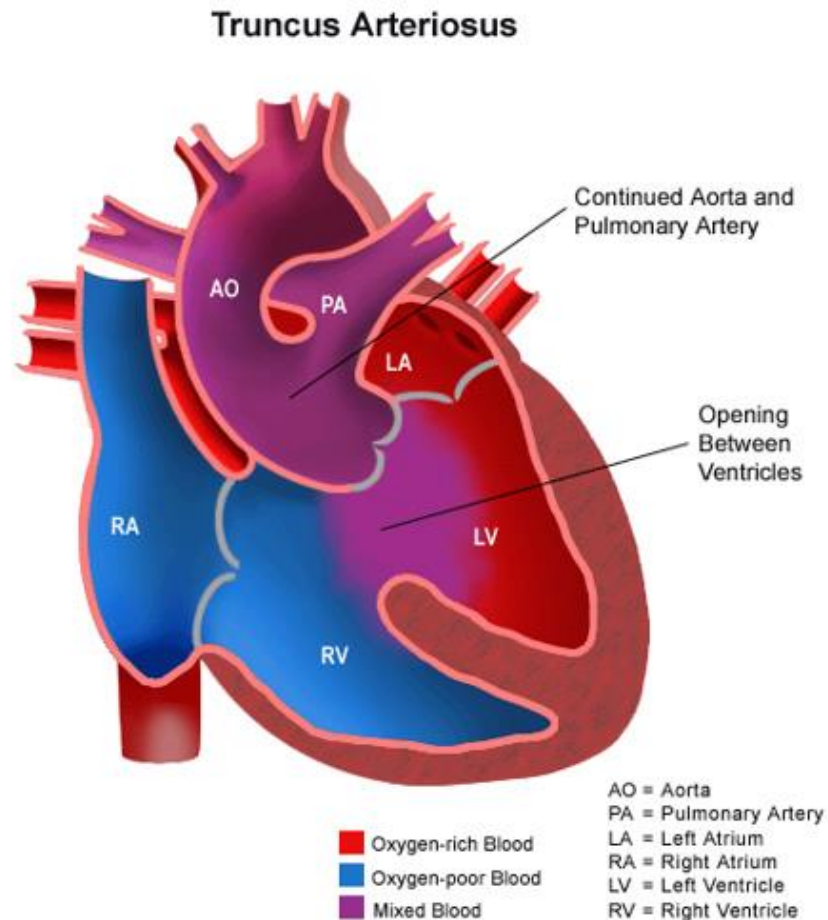


ADAM

AV Canal

- **Heart failure** develops
 - Early
 - Or not at all
- Usually repaired once failure develops
 - 3-6 months
 - **Earlier in Down's** given risk of pulmonary hypertension
- Murmur-**Diastolic rumble** or mitral valve murmur

Truncus Arteriosus



- **Single artery comes from heart giving origin to coronaries, pulmonary arteries, and aortic arch (Lev 1942)**
- Without treatment 80% die during infancy
- 1-4% of patients with congenital heart disease

Congestive Heart Failure

- Definition
 - **Heart is unable to pump enough blood to the body to meet its needs**
 - Poor term – actually working overtime!!
- Signs and symptoms
 - **Sweating, poor feedings, edema, increased sleeping, poor activity, Failure to Thrive**

Congestive Heart Failure

- **Volume overload most common cause** in infants: VSD, PDA, AV canal.
- Treatment:
 - Digoxin
 - Lasix
 - Aldactone
 - Enalapril/Captopril

Question 2

An otherwise healthy child with an acyanotic heart defect may have:

1. Low oxygen saturations
2. Right to left shunting
3. Left to right shunting
4. Clubbing of the digits

An otherwise healthy child with an acyanotic heart defect may have:

1. Answer: Left to right shunting

Obstructive Lesions

- These are lesions that **high pitch is bad** to hear
- The more obstruction, the sicker the child
- **Can present in shock** with severely obstructed lesions.

RVOT

- Stenosis
 - Supravalvar
 - Valvar
 - Subvalvar
 - Can be multi-level
 - Valvar can be dealt with in cath lab usually
 - Supra and sub need surgical repair

Pulmonic Stenosis

- Deep inspiration will increase the intensity of the murmur
- **Systolic ejection** murmur
- Heard over **LUSB** and base
- **Radiates to lungs**
- Harsh quality
- Widening of S2
- 13% of pathologic murmurs

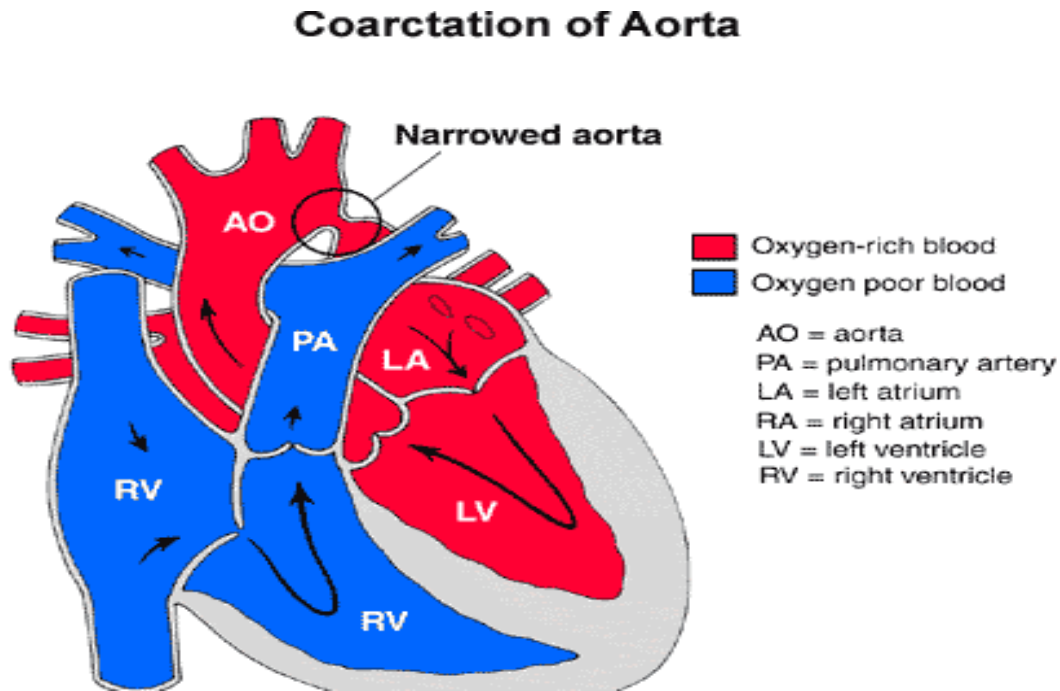
LVOT

- Stenosis
 - Supravalvar
 - Valvar
 - Subvalvar
 - Can be multi-level
 - Valvar can be dealt with in cath lab usually
 - Supra and sub need surgical repair

Aortic Stenosis

- **Midsystolic ejection** murmur
- Heard over **RUSB** and base
- **Radiates to the neck or back**
- Harsh quality
- S2 may be silent

Coarctation of the Aorta



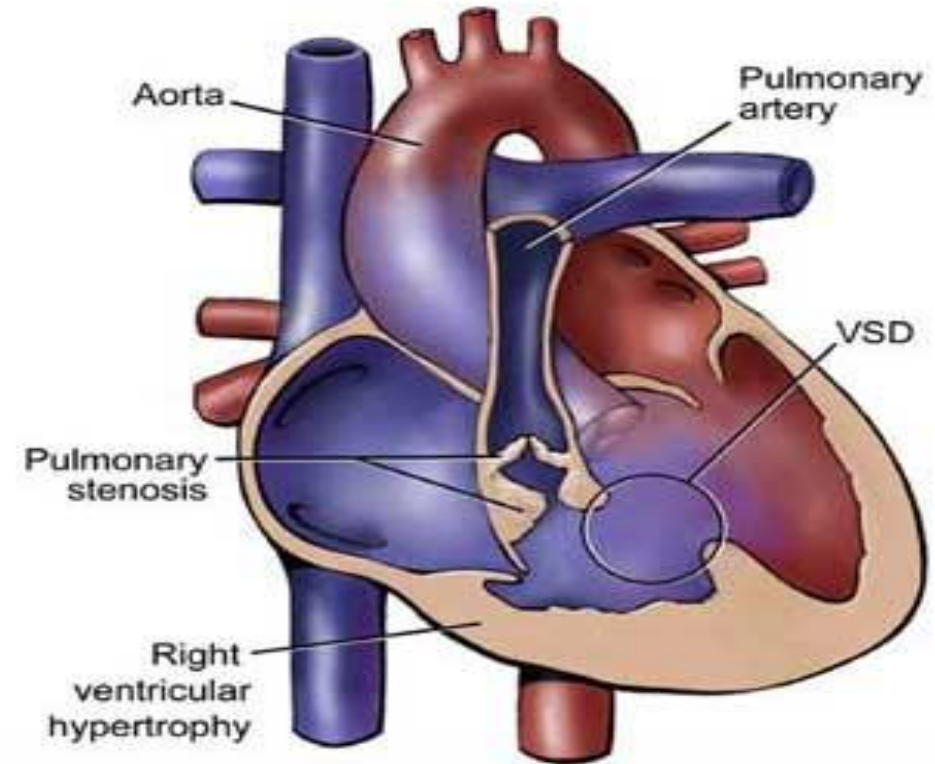
- Pulses/Shock
- Older children: Headaches, fatigue
- **High BP and bounding pulses in UE, Low BP in LE**
- Usually occurs between **right subclavian and left subclavian**
- Systolic ejection murmur

Cyanotic Lesions

- Right to Left shunting
 - Tetralogy of Fallot
 - Transposition of the Great Vessels/Arteries
 - Pulmonary Atresia
 - TAPVR
 - Single Ventricles

Tetralogy of Fallot

- **Combination of 4 defects:**
 - VSD
 - Pulmonary Stenosis
 - Right Ventricular Hypertrophy
 - Overriding Aorta – the aorta lies over the VSD (R & L ventricles)
- **Cyanosis occurs when blood flows from the right heart to the left heart and then out into the body**



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“TET Spell”

Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.



“Tet spell”

 ADAM.

TOF:Surgery

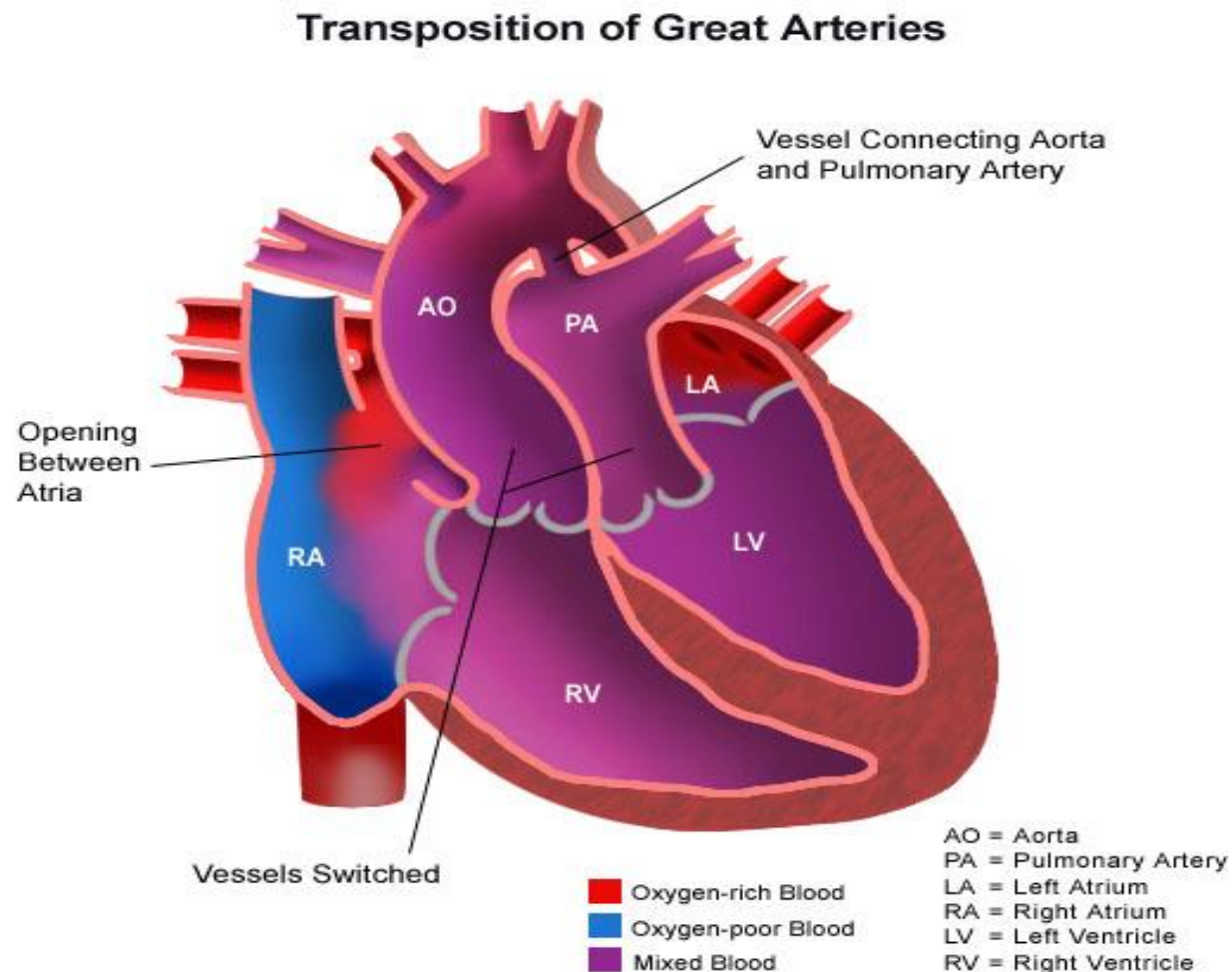
- First palliative surgery: BT (Blalock-Taussig) shunt (1945)
- Complete repair-performed CHO @ 3-4 m/o, other centers range from neonatal repair to 3-4 years old
- **Close VSD, open RVOT**

TOF: Long-term

- 5-10% Reoperation rate after complete repair
- Long-term issues
 - **Rhythm** – including ventricular
 - **RV failure**
 - May need **pulmonary valve replacement**

Transposition of Great Arteries

- Major arteries are switched
 - Right ventricle to Aorta
 - Left ventricle to Pulmonary artery
- **Parallel circuits**



TGA: Presentation

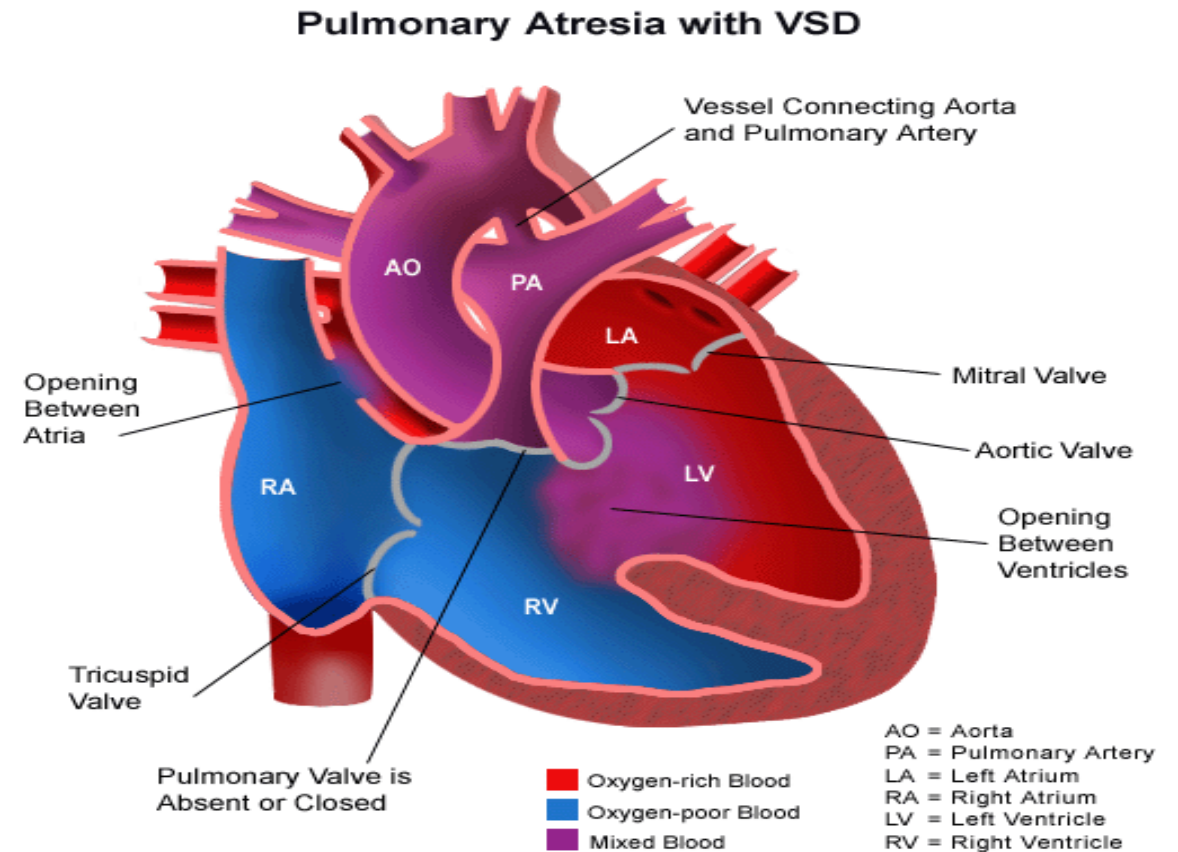
- Defect is incompatible with life unless there is an adequate communication between the two circuits. I.E **ASD, VSD, PDA**
- Notably cyanotic from birth. May progress as PDA closes
 - **Requires Prostaglandins**
 - **Early surgery**

TGA: Surgery

- Arterial Switch Operation: **Jatene** 1975
 - Must move coronaries
- Now performed regularly with excellent results (mortality < 2-3%), long term looks great

Single Ventricle Lesions

- Types of lesions:
 - HLHS
 - Pulmonary Atresia
 - Unbalanced AV canal
 - Tricuspid Atresia
 - Others
- Management:
 - Prostaglandins at birth
 - Staged palliative procedure
 - Norwood, Glenn and Fontan
 - Long term may require transplant



Question 3

The mother of a 4-month-old infant reports that he turned "blue" and seemed to have fast, labored breathing seemed to improve. On physical examination, the mucous membranes of the lips and mouth appear mildly cyanotic. A systolic murmur is heard best at the left sternal border. Vital signs are normal with normal peripheral pulses. There is no hepatomegaly. A likely diagnosis is:

1. Congestive heart failure
2. Apnea
3. Coarctation of the aorta
4. Cyanotic spell related to tetralogy of fallot

The mother of a 4-month-old infant reports that he turned “blue” and seemed to have fast, labored breathing seemed to improve. On physical examination, the mucous membranes of the lips and mouth appear mildly cyanotic. A systolic murmur is heard best at the left sternal border. Vital signs are normal with normal peripheral pulses. There is no hepatomegaly. A likely diagnosis is:

1. Answer: Cyanotic spell related to tetralogy of fallot

Question 4

Which of the following is true regarding innocent murmurs?

1. The murmur is holosystolic
2. Prompt referral to a cardiologist is indicated
3. A precordial thrill is present
4. The murmur is low intensity, grade 1-3

Which of the following is true regarding innocent murmurs?

1. The murmur is low intensity, grade 1-3

Physical Exam Findings with Right Sided Heart Failure (Systemic Congestion)

- **Periorbital/ facial edema**
- **Hepatomegaly**
- **Sudden weight gain**
- **Dependent edema and ascites**
- **Distended neck veins (rare in children)**

Physical Exam Findings with Left Sided Heart Failure (Pulmonary Congestion)

- **Tachypnea**
- **Increased respiratory effort**
- **Grunting and nasal flaring**
- **Retractions**
- **Crackles**

Management for CHD

- Refer to a pediatric cardiologist
- Provide good nutrition: small, frequent feeding for infants
 - **Increase calories-24kcal or greater**
 - 120-150kcal/kg/day requirements
- Monitor growth and development
- Prevent disease with routine immunization
 - **Possible RSV prophylaxis**

Management for CHD

- Educate child and family
 - Prevent dehydration, especially with children with cyanotic heart defects
 - Management of cyanotic spells
 - Prevention of respiratory infections

Infectious Endocarditis (IE) prophylaxis recommendations...

- In 2008 the American Heart Association made major changes to recommendations
- NO longer recommended for GU or GI tract procedures!!!
- Antibiotic prophylaxis still recommended for dental, respiratory, and children with infected skin or skin structures or MS tissue...

SBE Prophylaxis

- SBE prophylaxis, including dental cleaning
 - **Amoxicillin one hour prior to the procedure**
- Cyanotic congenital heart disease
 - **Has not been fully repaired**, including children who have had a surgical shunts and conduits.
- A congenital heart defect completely repaired with **prosthetic material or a device**
 - for the **first six months** after the repair procedure.
- Repaired congenital heart disease with residual defects
 - **persisting leaks** or abnormal flow at or adjacent to a prosthetic patch or prosthetic device.

Acquired Heart Disease

Bacterial (Infectious) Endocarditis (SBE)

- *Infection of the valves and inner lining of the heart
 - *Usually 2ndary to bacteremia
- *occurs in children with CHD, vascular abnormalities, prosthetic valves, recent cardiac surgery
- *Most common causative agent: **Streptococcus viridans**

Bacterial Endocarditis (cont.)

- Clinical Manifestations
 - **Unexplained fever**
 - Anorexia
 - Malaise
 - Weight loss
 - **Splinter hemorrhages**
 - **Petechia**

Bacterial Endocarditis (cont.)

- Treatment
 - Immediate use of high dose **antibiotics, long-term (2-8 weeks)**
 - **Potential surgery** to remove infectious material/rebuild areas of damage
- Prevention
 - Prophylactic antibiotic administration before procedures that risk entry of organisms to child

Acute Rheumatic Fever (ARF)

- An **inflammatory connective tissue** disease that affects the heart, joints, CNS, & subcutaneous tissue
- Peak age 5-15 yrs
- Usually the result of **Group A Beta Hemolytic Strep** Infection
- Risk of ARF ↑ with ↑ GABHS infections

Criteria for Diagnosis of ARF

- The presence of **2 major or 1 major and 2 minor criteria**
- **(The Jones Criteria)**



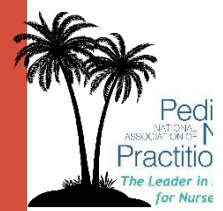
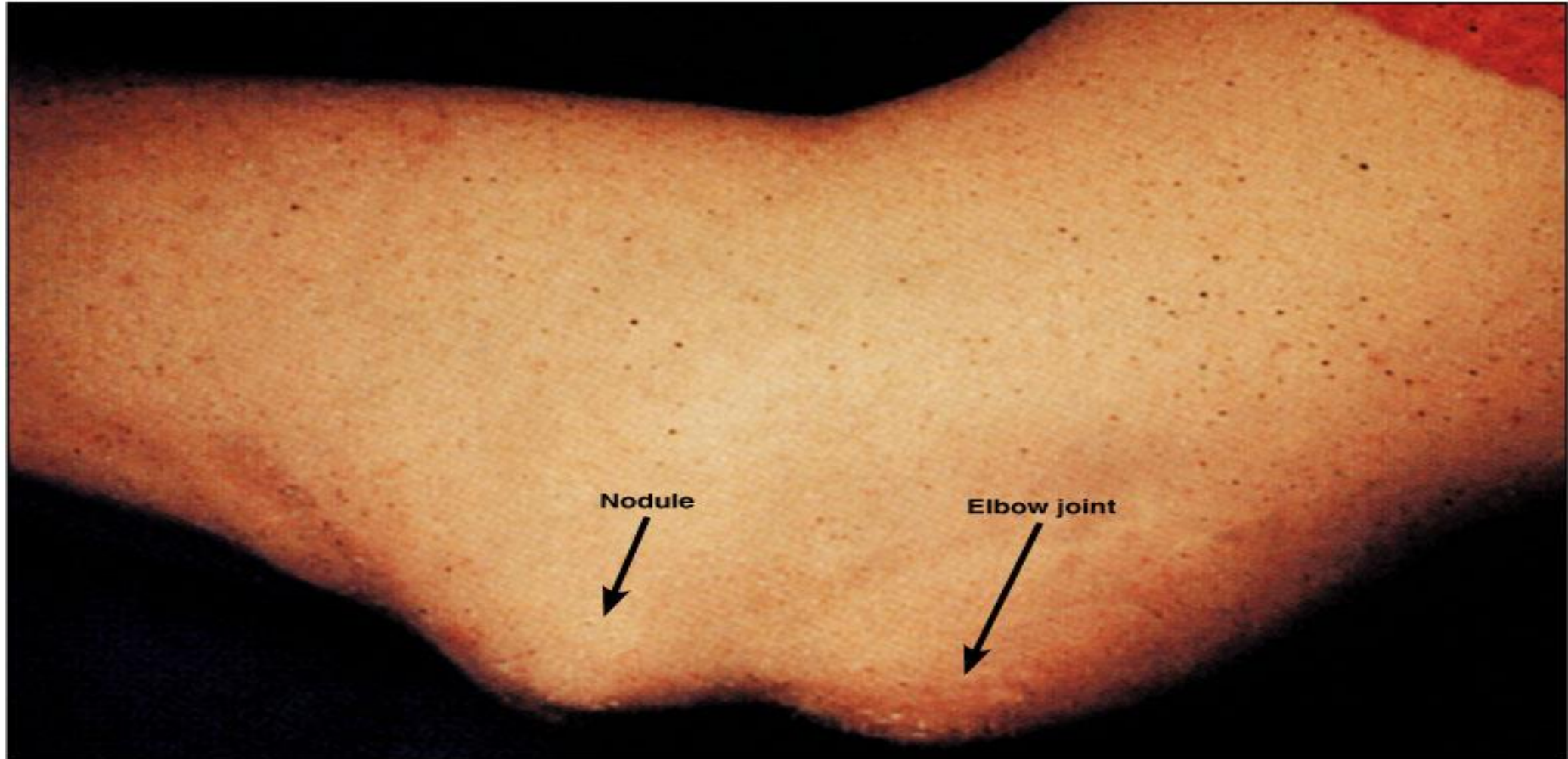
Major Criteria - JONES

- **Migratory polyarthritits (Joints)**
- **Carditis (significant new murmur)**
- **Subcutaneous Nodules**
- **Erythema marginatum**
- **Syndenham's Chorea**

Erythema Marginatum



Subcutaneous Nodule



Minor Criteria

- Fever
- Arthralgia
- Previous rheumatic fever
- Elevated acute phase reactants
 - Increased ESR or C-reactive protein
- Prolonged PR interval

Management of ARF

- Refer to a pediatric cardiologist
- Treatment of GABHS:
 - Penicillin VK or Erythromycin or
 - Benzathine penicillin G IM**
- Anti-inflammatory agents
 - ASA**
 - Prednisone
- Daily prophylaxis
- Educate child and family
- Administer routine immunizations

Kawasaki Disease

- **Acute systemic vasculitis** of unknown cause
- 80% of cases in children under **5 years of age**
- 3 phases
- **Cardiac involvement** is most serious complication and risks include the potential for MI due to **coronary narrowing/occlusion** (due to inflammation)
- S/S: **strawberry tongue, pink eyes (conjunctivitis but without discharge), trunk rash, dry/cracked lips, redness, swelling, flaking of palms and soles of feet**
- Treatment: **Aspirin, IVIG therapy**

Kawasaki Disease



Hypertension (HTN)

- An average systolic and/or diastolic **BP** > **95%** for age and sex on at least 3 occasions over 1-2 weeks
- Use NHLBI guidelines
- PreHTN: average SBP or **DBP** \geq **90th** percentile but < **95th** percentile

Causes of Primary HTN

- Genetics
- Obesity
- Stress
- ↑ Sodium intake

Causes of Secondary HTN

- Renal disorders
- Vascular disorders
- Endocrine disorders
- Other (corticosteroids, oral contraceptives, amphetamines, \square ICP, psychotropic medications)

***The younger the child and the higher the BP, the greater the probability of renal pathology**

Thorough History with Emphasis On:

- Family health history (early cardiac disease)
- Past medical history
- Nutrition
- Medications
- Drug and caffeine usage
- ROS

Thorough Physical Exam with Emphasis On:

- **Three limb blood pressures/
-3 positions**
- Height and weight
- Skin
- Eyes
- Thyroid
- CV
- Respiratory
- Abdomen

Diagnostic Workup

Urinalysis

Rule out infection, hematuria, proteinuria

Protein/Cr Ratio

Kidney function

Renal Ultrasound

Rule out renal scarring, congenital renal anomalies

EKG

Cardiomegaly

CBC with differential

Rule out anemia, consistent with chronic renal disease

Electrolytes, BUN, Cr

Rule out renal disease, pyelonephritis

Management of Primary HTN

- Non-pharmacologic management

- Weight loss, exercise**, low fat, low cholesterol diet, ↓ sodium intake, avoid alcohol, caffeine and OCPs, stress reduction techniques

- Pharmacologic management

- Obtain baseline **ECHO, ACE inhibitors**, calcium channel blockers, diuretics, beta adrenergic blockers, vasodilators

Medication therapy

- Choice often dictated by underlying or concurrent medical condition or undesired side effects
- **Max out the dose of monotherapy** before adding a 2nd agent

Goals of therapy

- In uncomplicated primary HTN
 - < 95th percentile
- **In patients with diabetes, and/or evidence of end organ damage**
 - < 90th percentile
- **Chronic kidney disease patients**
 - 50th percentile

Management of Secondary HTN

Refer to appropriate specialty

Myocarditis & Cardiomyopathy

- Rare inflammatory illness of the muscular walls of the heart
- Etiology: often viral
- H&P: history of **fever/viral illness**, lethargy, low-grade fever, decreased appetite, exercise intolerance, **palpitations**
- S/S: pallor, mild cyanosis, cool and mottled, poor perfusion, tachypnea, grunting, tachycardia, gallop rhythm, **muffled heart sounds**, weak pulses, **hepatomegaly**

Question 5

A MOST concerning cardiac finding in a newborn or infant assessment is:

1. A blood pressure of 70/40
2. Decreased femoral pulses
3. A regular heart rate of 100 while sleeping
4. A soft, grade III/VI systolic murmur

A MOST concerning cardiac finding in a newborn or infant assessment is:

1. Answer: Decreased femoral pulses

Question 6

A 12-year-old girl seen at a routine visit has a blood pressure of 140/90. She denies any symptoms. The initial management would include:

1. Intravenous pyelogram
2. Return for two repeat blood pressure measurements
3. No follow-up needed-blood pressure probably related to anxiety
4. Diuretic therapy

A 12-year-old girl seen at a routine visit has a blood pressure of 140/90. She denies any symptoms. The initial management would include:

1. Answer: Return for two repeat blood pressure measurements

Question 7

The NP is assessing a neonate for congestive heart failure. The NP would expect to find:

1. Poor feeding and tachypnea
2. Jaundice and the liver at 1 cm below right costal margin on palpation
3. Pitting ankle edema and lethargy
4. Bradycardia and sweating

The NP is assessing a neonate for congestive heart failure. The NP would expect to find:

1. Answer: Poor feeding and tachypnea

Question 8

The rash associated with Acute Rheumatic Fever is:

1. Pink papular with pale center on face and trunk that is pruritic
2. Macular erythematous with pale center on the trunk and extremities
3. Nodular near the extensors
4. Non-pruritic vesicular on trunk

The rash associated with Acute Rheumatic Fever is:

1. Answer: Macular erythematous with pale center on the trunk and extremities

Question 9

Blood pressure should be measured at well child visits, beginning at age:

1. 2 years
2. 3 years
3. 4 years
4. 5 years

Blood pressure should be measured at well child visits, beginning at age:

1. Answer: 3 years

Question 10

The initial presentation of acute rheumatic fever is preceded by:

1. A viral illness
2. A group A streptococcal infection
3. Exposure to mites
4. Exposure to chicken pox

The initial presentation of acute rheumatic fever is preceded by:

1. Answer: A group A streptococcal infection

Question 11

Which of the following is a common cause of acquired coronary artery disease during childhood?

1. Rheumatic fever
2. Hypertension
3. Systemic lupus erythematosus
4. Kawasaki disease

Which of the following is a common cause of acquired coronary artery disease during childhood?

Answer: Kawasaki disease