A GUIDE TO PEDIATRICS MINI-OSCE

DISCLAIMER: this file is by no means a replacement to the material.

*note that the file only contains high-yield pediatrics' topics for end-of-rotation exam purposes.

PREPARED BY LAYLA RAFEH / LAST UPDATE SEPTEMBER-2021

- Breast/human milk: more lactoalbumin(whey), more lactoferrin, more vitamin (a,c,e), more igA, lower iron content, lower casein content,same caloric fat and carbs content.
- Highest caloric content in human milk is fat.
- Cow and human milk are not equal in calcium,phosphorus and vitamin d content.
- Between whey and casein, whey is the "faster" protein supplement because its amino acids are absorbed quickly by your body. While casein is the "slower" supplement — since it's digested more slowly
- Human milk has more whey:casein ratio than cows formula
- TPN complications: hypo or hyperglycemia, low copper levels, cholestatsis.
- Fresh cow’s milk introduction-12 months.
- Although iron and calcium are present in smaller quantities in human milk, they’re absorbed better by the infant.
- Low fat milk introduction at 24 months.
- Protein is the antigenic part of cow’s milk(whey).
- Human milk increases the risk of vitamin d deficiency and iron deficiency anemia.(but iron and calcium are absorbed better).
- Breast milk does NOT have more sodium than formula milk.
- Pts with galactosemia-lactose free formula.
- Patients with cow milk allergy place on hydrolysed formula/cross reactivity with soy milk.

Comparison of Breast Milk to Cow Milk

Component	Human Milk	Cow Milk
Water/solids	Same	Same
Calories	20 cal/oz	20 cal/oz
Protein	1–1.5% (whey dominant)	3.3% (casein dominant)
Carbohydrate	6.5–7% lactose	4.5% lactose
Fat	high in LCFAs	high in MCFAs
Minerals	Iron better absorbed	Low iron and copper
Vitamins	Diet dependent, low in K	Low in C, D
Digestibility	Faster emptying	Same after 45 days
Renal solute load	**Low** (aids in renal function)	Higher

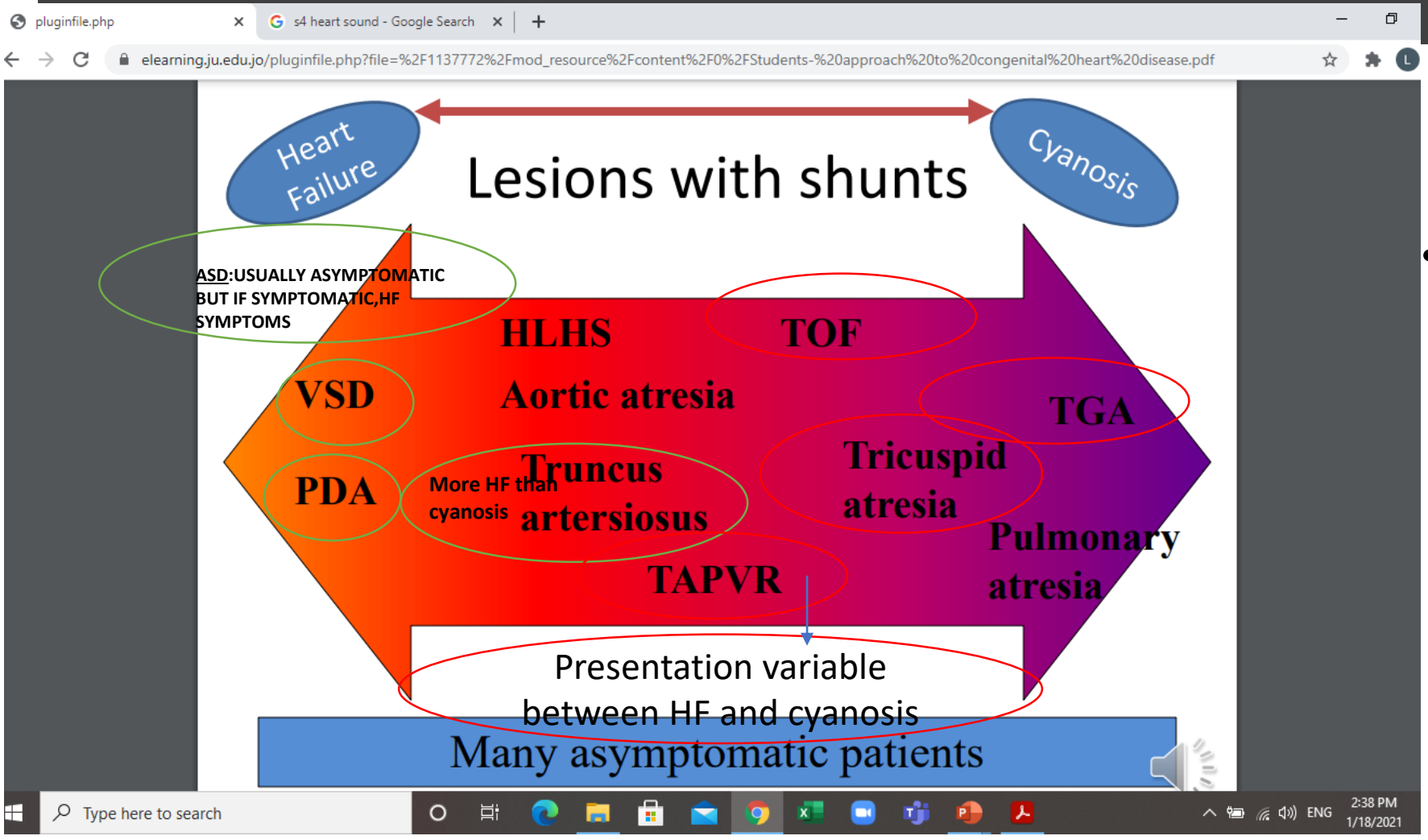
Cardiology

General notes

- Cardiac output=SV(preload,afterload,contractility)*HR(ANS)
- SV=LV end diastolic volume-LV end systolic volume
- Ejection fraction=SV/LVEDV *100%(percentage of how much blood is ejected with each contraction)
- Principle differences in fetal circulation compared to post-natal circulation: PFO (from RA to LA) – PDA (from PA to AO) – Ductus venosus (from UV to IVC)

S1	CLOSURE OF THE AV VALVES	Best heard at the apex(mitral area),lower left sternal angle(tricuspid area)
S2	CLOSURE OF THE SEMILUNAR VALVES	Best heard at the apex,splitting during inspiration(A2P2) best heard at the left upper sternal angle(pulmonic area)
S3(during early diastole)	JUST AFTER S2,RAPID FILLING OF A COMPLIANT VENTRICLES,VOLUME OVERLOAD, normal finding in children, pregnant females and well-trained athletes,pathologically heard with acute left heart failure.	Best heard at the point of maximal impulse which is the apex normally with the bell in left lateral decubitus position.
S4(during late diastole)	JUST BEFORE S1,ALWAYS PATHOLOGICAL,atrial gallop, atrial contraction forces blood through the atrioventricular valves, a **S4** is produced by the blood striking the left ventricle,pathologically heard **with stiff ventricles**.	Best heard at the point of maximal impulse which is the apex normally with the bell in left lateral decubitus position

Congenital heart diseases



- Heart failure symptoms
  - Respiratory symptoms(dyspnea,rapid breathing,grunting)
  - Diaphoresis
  - Poor feeding
  - Poor weight gain or FTT
  - Exercise intolerance

- Plethoric lung field: ASD(if large enough to cause HF),VSD,PDA, TGA(the aorta is pumping into the lungs), Truncus arteriosus(also the common trunk is pumping more blood into the pulmonary artery)
- Oligemic lung fields: tricuspid atresia(no connection between RA & RV), TOF, ebstein anomaly(dysplasia of the tricuspid valve), nonaan syndrome.
- TGA – ASD or PFO to survive, if PDA is found its better, indication for keeping it patent by IV PG-E1.
- Truncus arteriosus born with a VSD
- Tricuspid atresia born with a PFO, they require either a VSD or PDA for mixing up blood/surviving,the smaller the more the cyanosis.
- TAPVR, infracardiac present more earlier more possible for an obstruction, presentation is variable between HF and cyanosis, they require PFO to survive, xray findings dilated right heart border and dilated SVC, Dilated left heart border and dilated anomalous vessels.

Lesions of CHD;

- 1.septal;ASD,VSD,PDA
- 2.valvular;stenosis or atresia(complete absence)
- 3.abnormal connections
- 4.a combination of the above
- Lesions are either non-cyanotic;left to right shunting;PDA,ASD,VSD or cyanotic right to left the 5 Ts;tetralogy of fallot,transposition of great vessels,truncus arteriosus,tricuspid atresia,total anomalous pulmonary venous return.

Normal CXR in a neonate:

Look for:

- Heart size
- Heart shape
- Mediastinal width, thymus
- Lung fields (plethoric or oligemic indicating increased or decreased blood flow)

Atrial septal defect

- Atrial septal defect Often **asymptomatic** ; heart failure in large defect, along with the relevant HF symptoms mentioned before.(history)
- P/E: poor growth,heart failure signs;(heart failure signs displaced heart beat due to cardiomegaly,edema and hepatomegaly,delayed capillary refill and weak peripheral pulses),auscaltatory findings; Atrial septal defect Murmur midsystolic **murmur** , S2 splitting(fixed),best heard at the pulmonic area(left upper sternal area)
- What causes fixed splitting of the S2 is the ASD.
- Chest xray findings(related to heart failure)
    1. Cardiomegaly(increased cardiothoracic ratio,abnormal>60%)
    2. Increased pulmonary vascular markings(hazy lung fields)-plethoric lung fields

Ventricular septal defect

- Ventricular septal defect Heart failure, poor growth and failure to thrive.(history)
- P/E; heart failure signs as mentioned earlier, poor growth, auscaltatory findings Ventricular septal defect Murmur"pansystolic".
- X-ray findings in VSD is findings of HF.
- The smaller the defect the more intense(louder) the sound of the murmur.
- Murmur grading;(2/6: soft, 3/6 loud with no thrill, 4-6/6 loud with thrill)
- At risk of infective endocarditis,
- Pts at risk of infective endocarditis: VSD, coarctation of aorta and PDA.

Patent ductus arteriosus

- Patent ductus arteriosus**;heart failure symptoms** Fast or increased work of breathing, **respiratory infections** , easy fatigability**, poor growth**, asymptomatic if small PDA.(history)
- P/E: Patent ductus arteriosus **Continuous "machine-like"Murmur**;heard everywhere along the heart, heart failure signs mentioned earlier, endocarditis,**wide pulse pressure(sys-dia)**, tachycardia, and **bounding peripheral pulses** due to increased cardiac output.
- A **widened pulse pressure** (> 30mmHg) occurs both because of a mild increase in systolic blood **pressure** to overcome the decrease in distal blood flow due to run-off through the **PDA** during diastole, in addition to a lower diastolic blood **pressure** from the run-off.

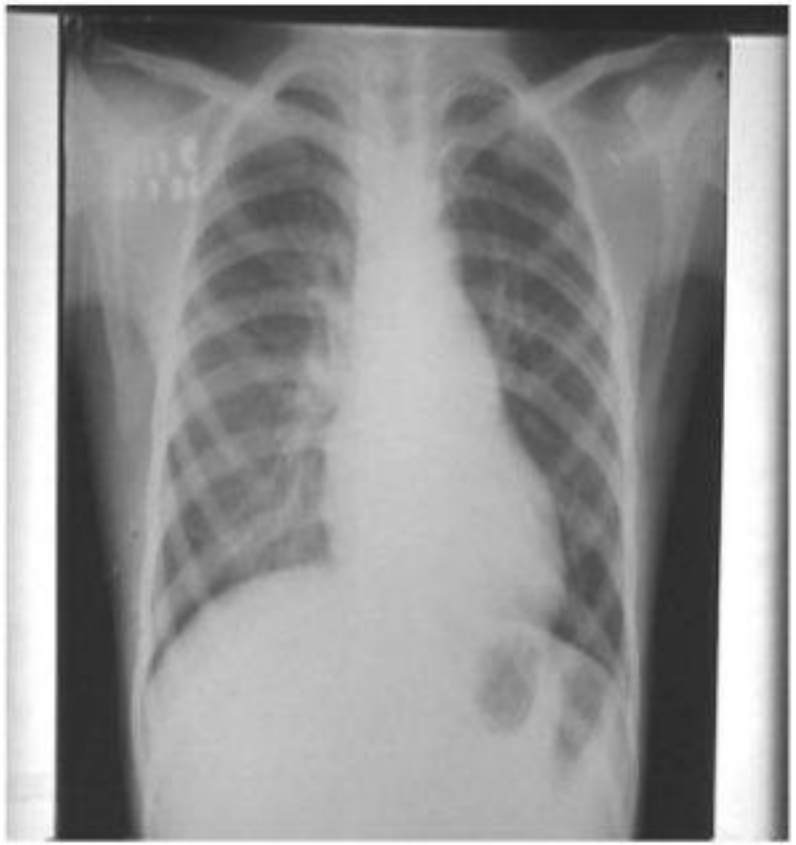
- All children of any age with congenital heart disease (except secundum ASD), including neonates,are at risk of infective endocarditis. The risk is highest when there is a turbulent jet of blood, as with a VSD, coarctation of the aorta and PDA or if prosthetic material has been inserted at surgery. It may be difficult to diagnose, but should be suspected in any child or adult with a sustained fever, malaise, raised ESR, unexplained anemia or haematuria.

Chest xray findings

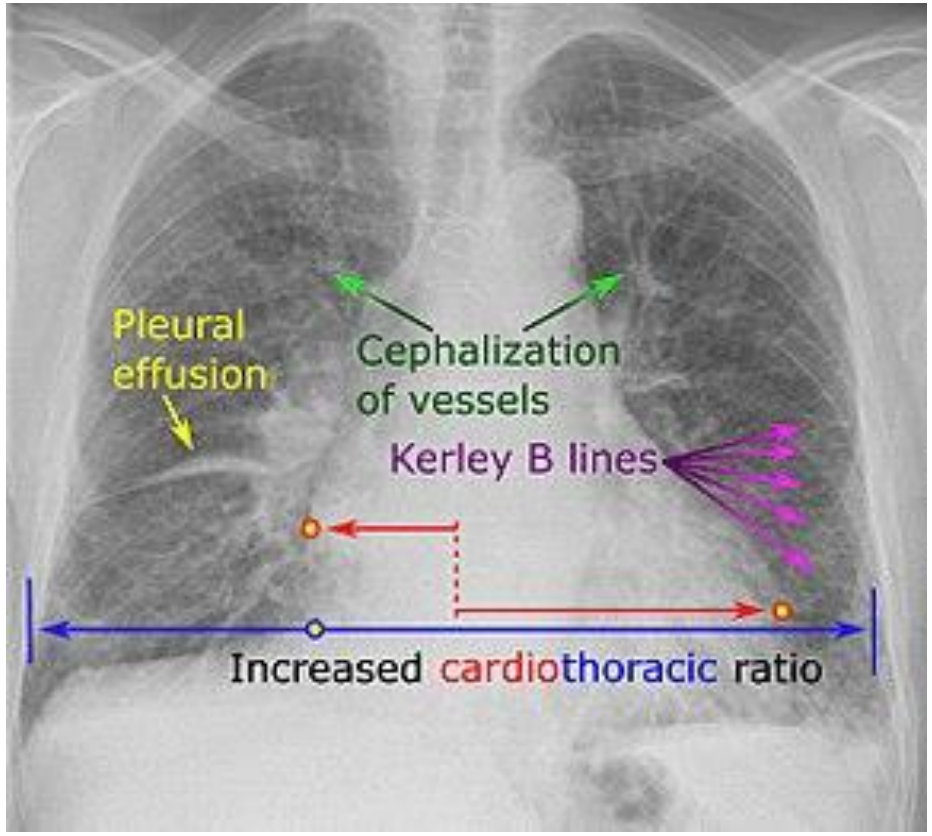
the angle between the pulmonary artery and the aorta is filled in contrast to TOF where there is concavity of the pulmonary artery

PATENT DUCTUS ARTERIOSUS

- Enlargement of the left heart chambers.
- Enlargement of the ascending aorta or aortic arch.
- Pulmonary plethora with enlarged central and peripheral artery.
- Filling up of the angle between the aortic arch and the pulmonary artery : Most specific sign.
- Possible PDA calcification in adults.

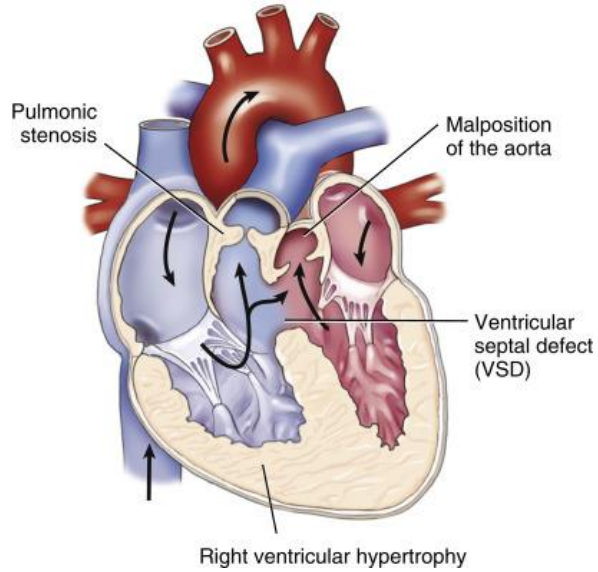


- Xray findings in a child with CHD causing HF:
    1. Cardiomegaly or increased cardiothoracic ratio>60%
    2. Plethoric lung field or congestion of pulmonary vasculatre (kerley B lines)
    3. Pleural effusion or obliteration of the costophrenic angles coulde be found.



Tetralogy of fallot

1. RV outlet obstruction or narrow pulmonary valve(stenosis)
2. RV hypertrophy,both ventricles pump at the same pressure
3. VSD
4. Over-riding aorta(arises from both the RV AND LV)



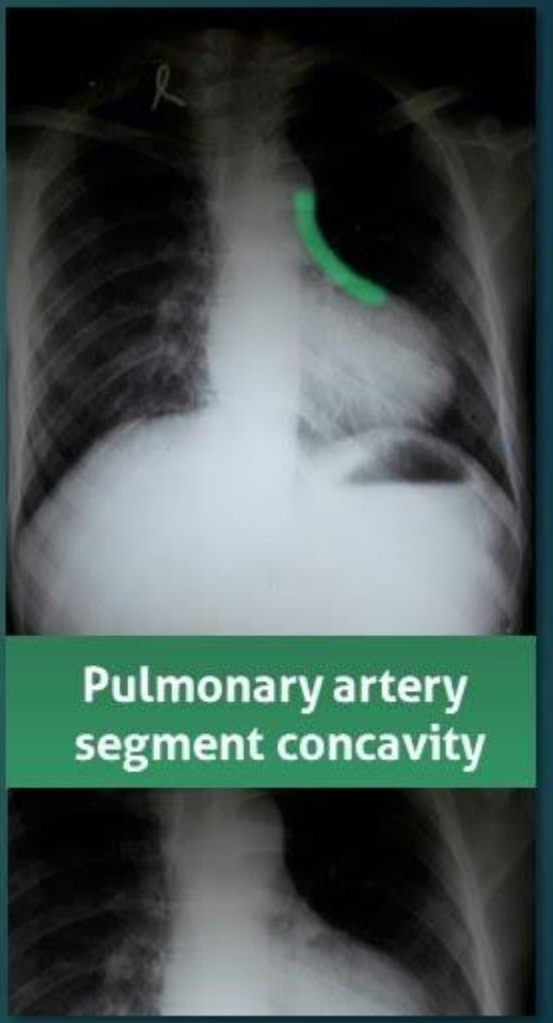
- The most common CHD,the amount of right to left shunting depends on the amount of pulmonary stenosis,variable degrees of cyanosis
- See next slide for chest xray findings(oligemic lung field because of decreased BF to the lungs because of pulm. Stenosis or atresia).
  - P/E; Ejection systolic murmur.Thrills.Single S2.Cyanosis.
  - Complications;clubbing,FTT(all CHD)

Tetralogy of Fallot: Boot-Shaped Heart

Apex rotated superiorly

Oligemia

Enlarged aorta



Pulmonary artery segment concavity

Transposition of great arteries(TGA)

- SEVERE CYANOSIS EARLY AFTER BIRTH.
- Aorta originating from the right ventricle, and pulmonary artery originating from the left ventricle.
- They require ASD or PFO for mixing

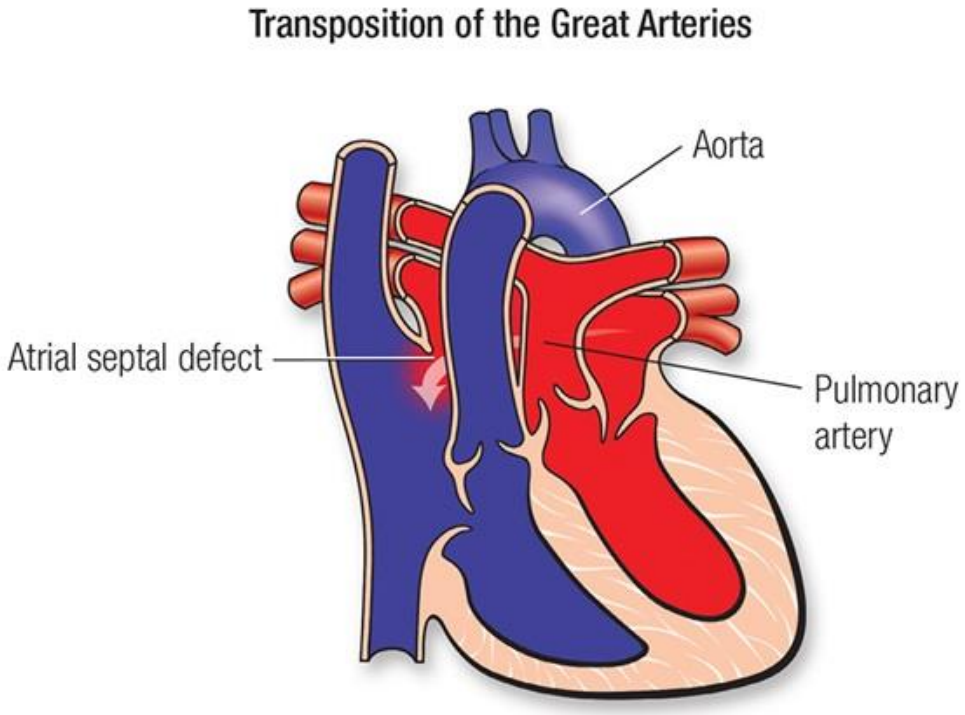
Some blood to survive.

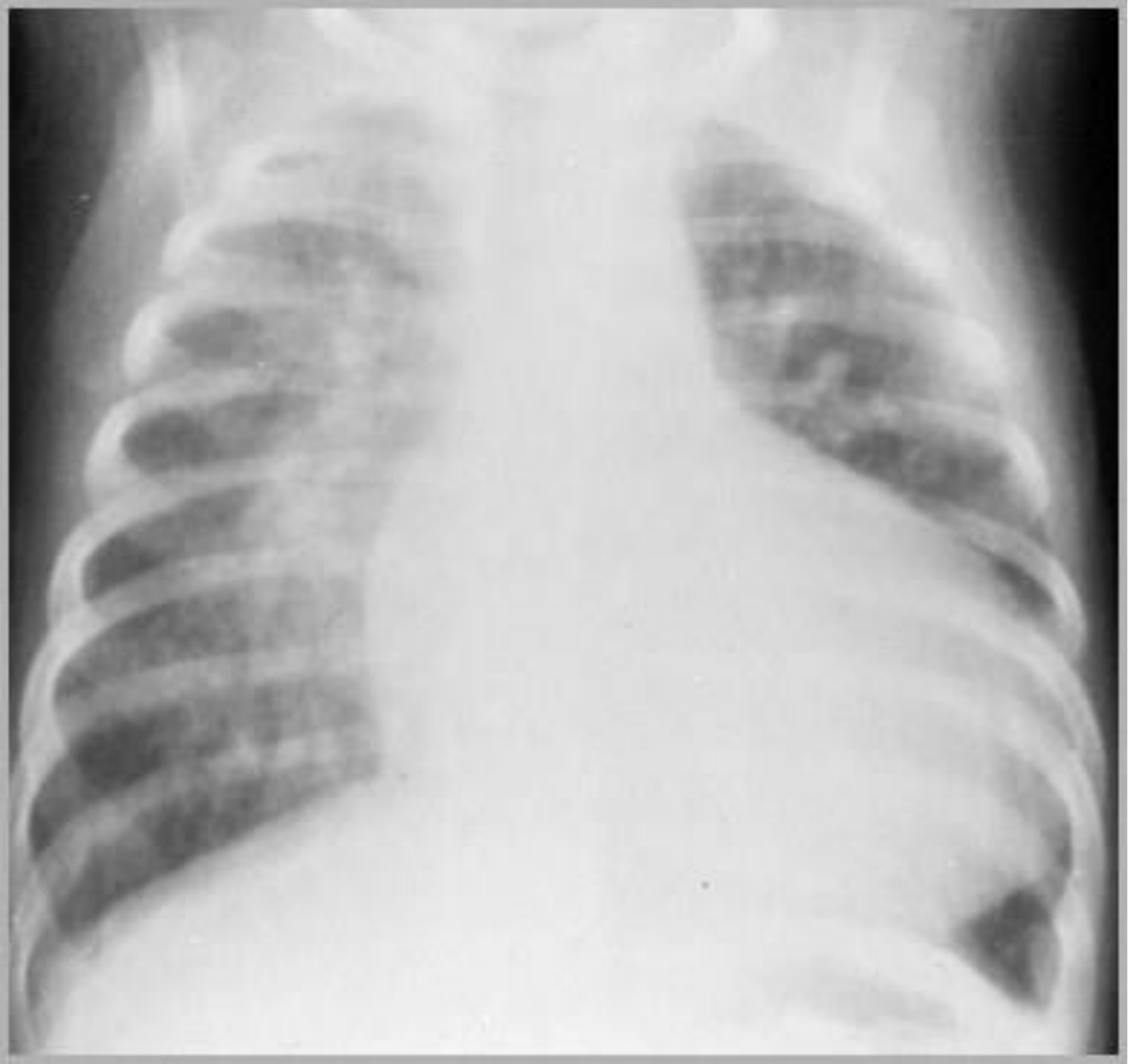
- If theres PDA a better condition

(another source for mixing blood)

- One of the indications to keep the

PDA open is TGA by giving IV PG-E1.



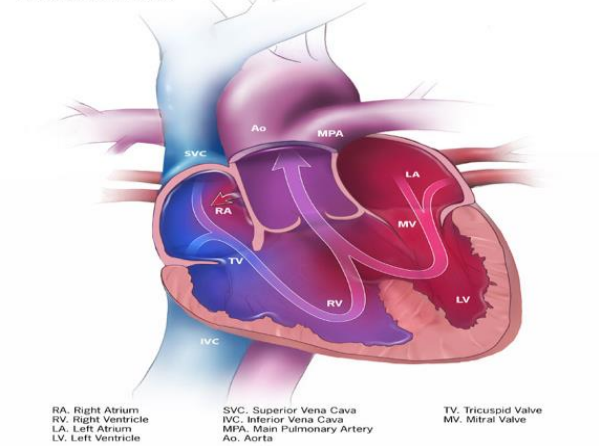


You obtained an X- ray for this baby What are the findings?(egg-on-string)

- Cardiomegally
- Abnormal heart shape (like an egg on its side)
- Narrow mediastinum(string in contrast to TOF where there is enlarged aorta)
- Plethoric lung fields in contrast to TOF(the aorta is pumping blood to the lungs)

These findings are suggestive of TGA

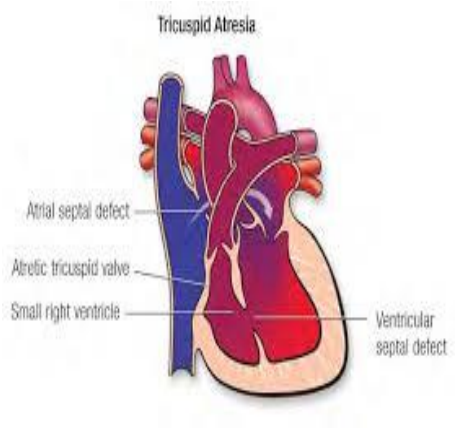
Truncus arteriosus(RARE)

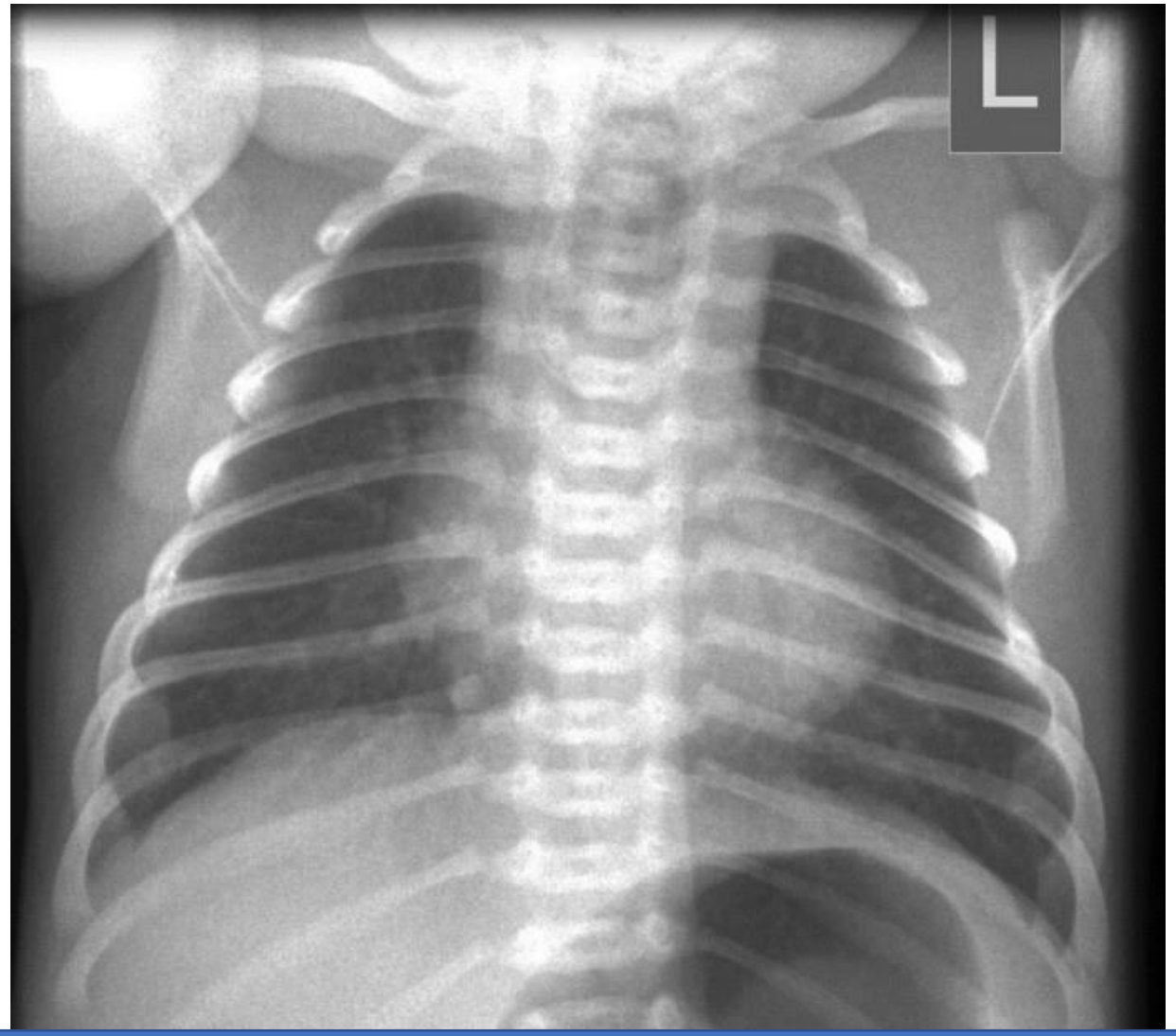


- The presence of a common trunk that supply the systemic, pulmonary and coronary circulation.
- Almost always associated with a VSD.
- The flow to each vessel depends on the resistance, usually the resistance in the PA is less, so more flow goes to the pulmonary circulation causing pulmonary congestion causing more blood return to the LA&LV causing hypertrophy and cardiomegaly.
- This patients with truncus arteriosus present with HF symptoms and less cyanotic symptoms.
- Xray findings include those of HF.

Tricuspid atresia

- Complete atresia,no connection between the RA&RV.
- Born with PFO.
- To survive they need to have pulmonary blood flow thru either PDA or VSD.
- Degree of cyanosis depend on size of VSD or PDA,the smaller the less blood supply the more the cyanosis.



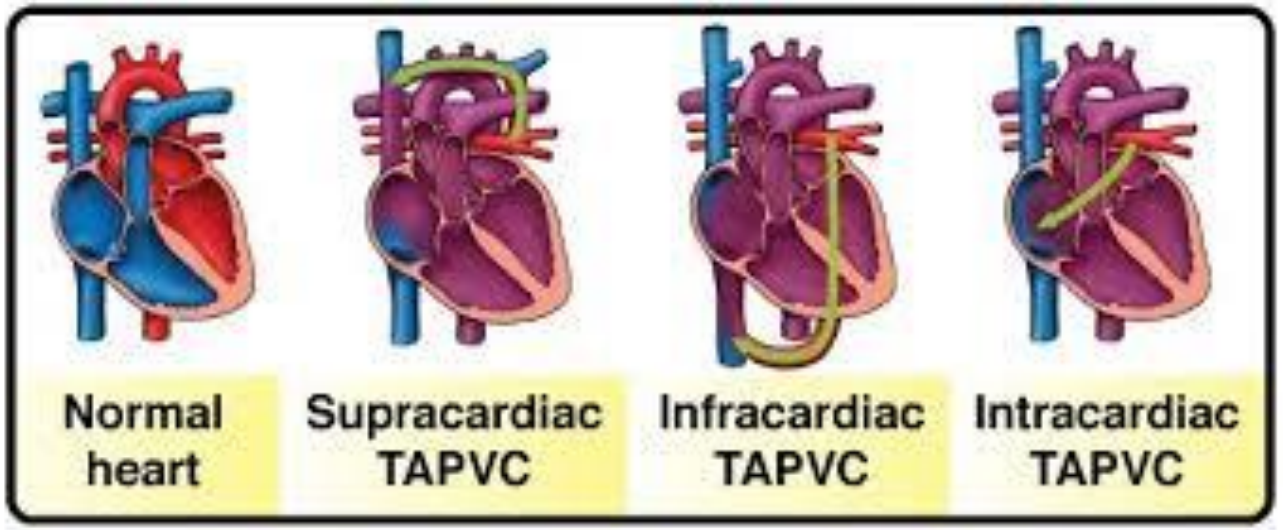


Dark lung fields, indicates decreased pulmonary blood flow
Examples (TOF with severe PS, Pulmonary atresia)

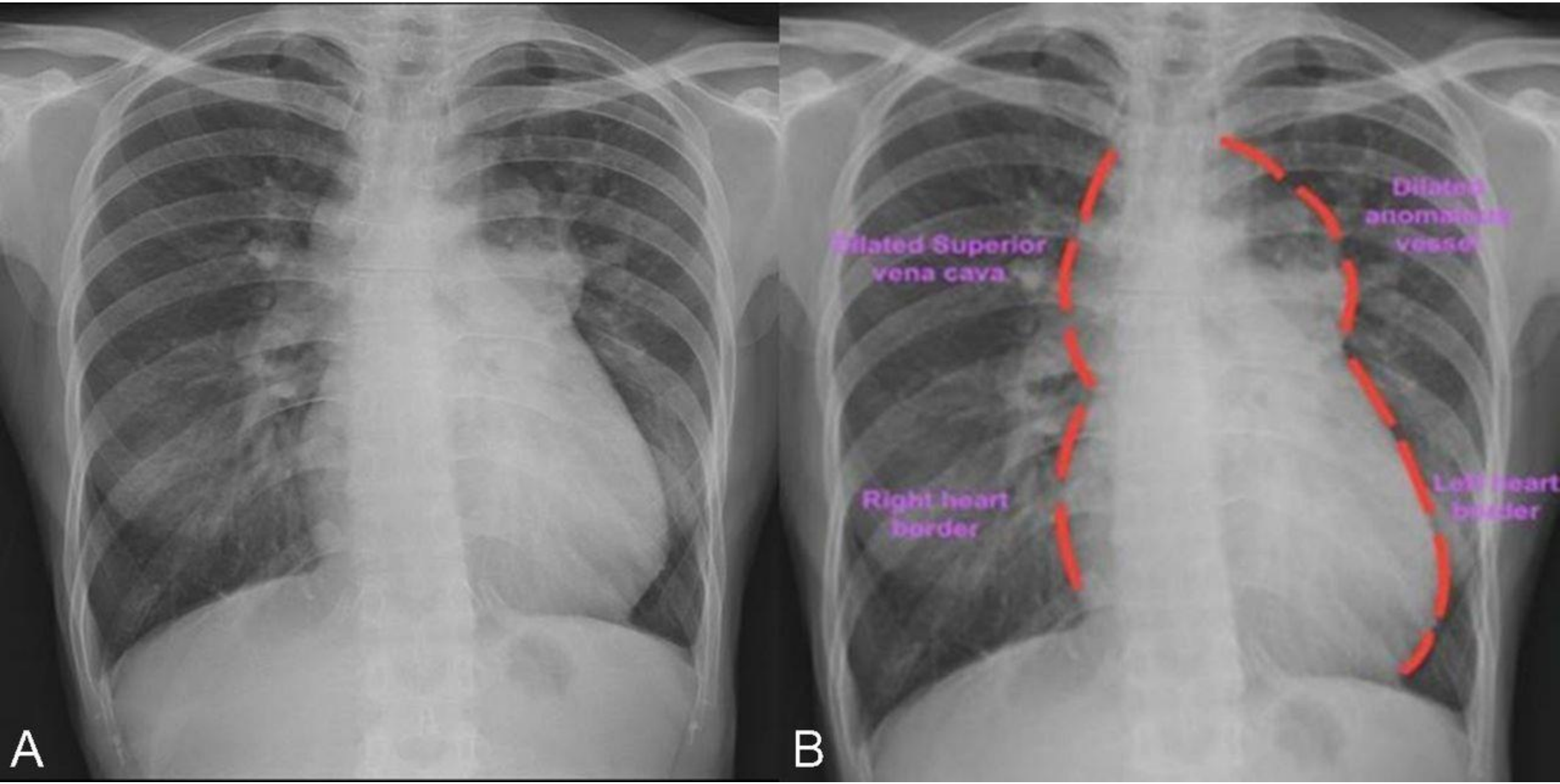
- Cardiomegaly,wide mediastinum plethoric lung field goes with decreased pulmonary supply either due to pulmonary atresia or TOF with severe Pulmonary stenosis

Total anomalous pulmonary venous return(TAPVR)

- They are two types supradiaphragmatic and infradiaphragmatic.
- Supradiaphragmatic;the pulmonary veins drain into the SVC, the infra diaphragmatic the pulmonary veins drain into the IVC,intracardiac to the RA, results in mixing of blood.
- To survive they need a PFO for systemic blood flow

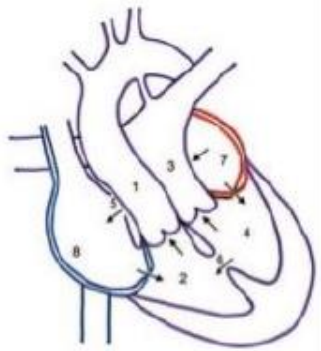


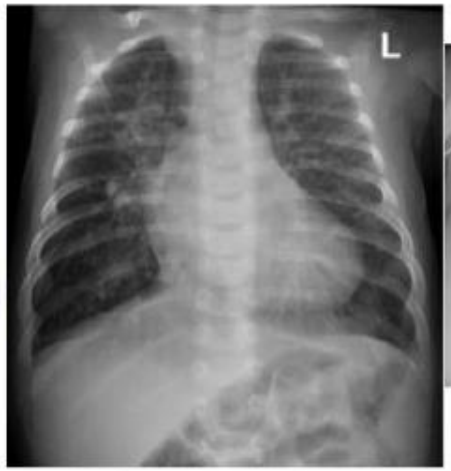
- Presentation is variable between over circulation symptoms because of increased preload,cyanosis.
- The infradiaphragmatic type presents earlier because typically will have obstruction in the pathway.
- Extra note;obstruction;pressure overload,downstream hypertrophy
- Regurge;volume overload,downstream and receiving chamber hypertrophy.

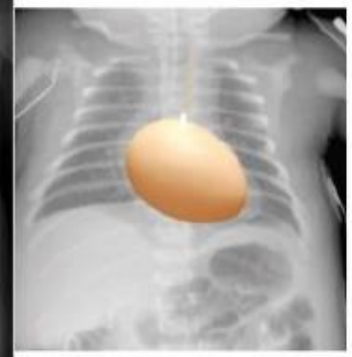

A
B
Dilated Superior vena cava
Right heart border

Transposition of great vessels

EGG ON STRING SIGN

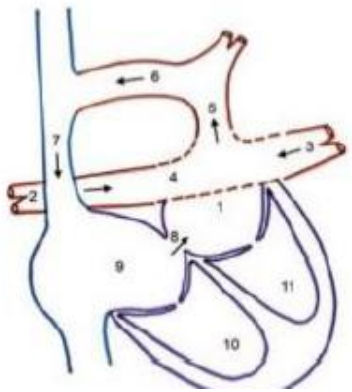


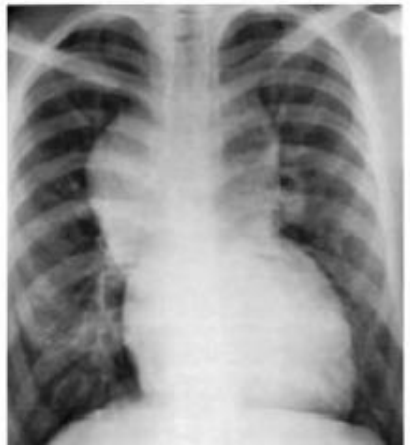


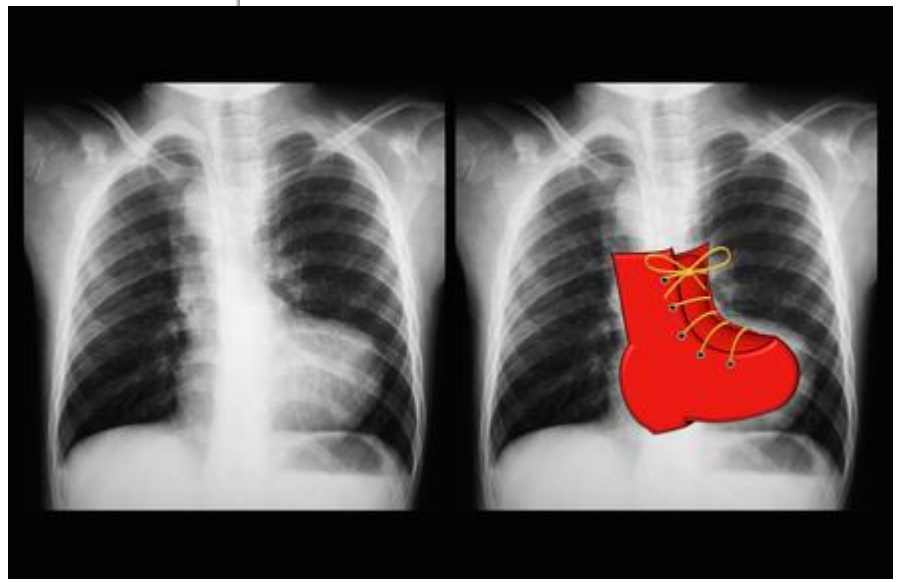


Total Anomalous Pulmonary Venous Return

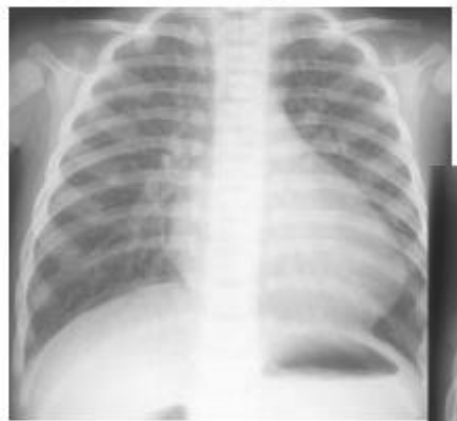
SNOWMAN SIGN



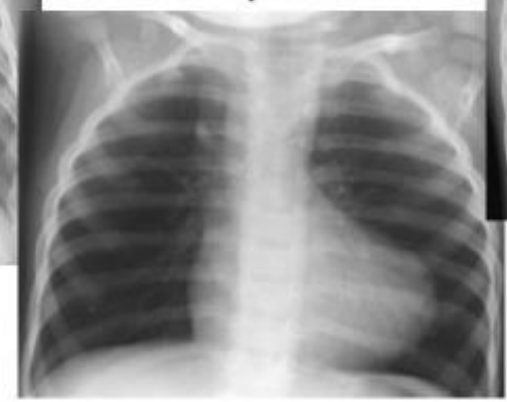




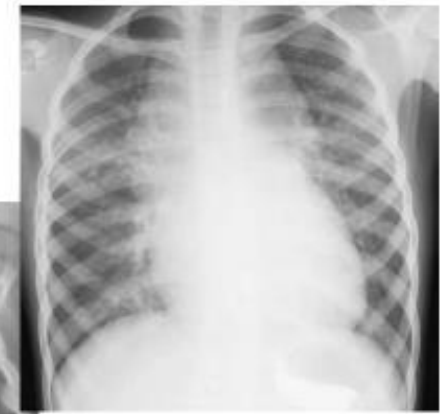
Egg on a string = TGA



Boot shaped = ToF



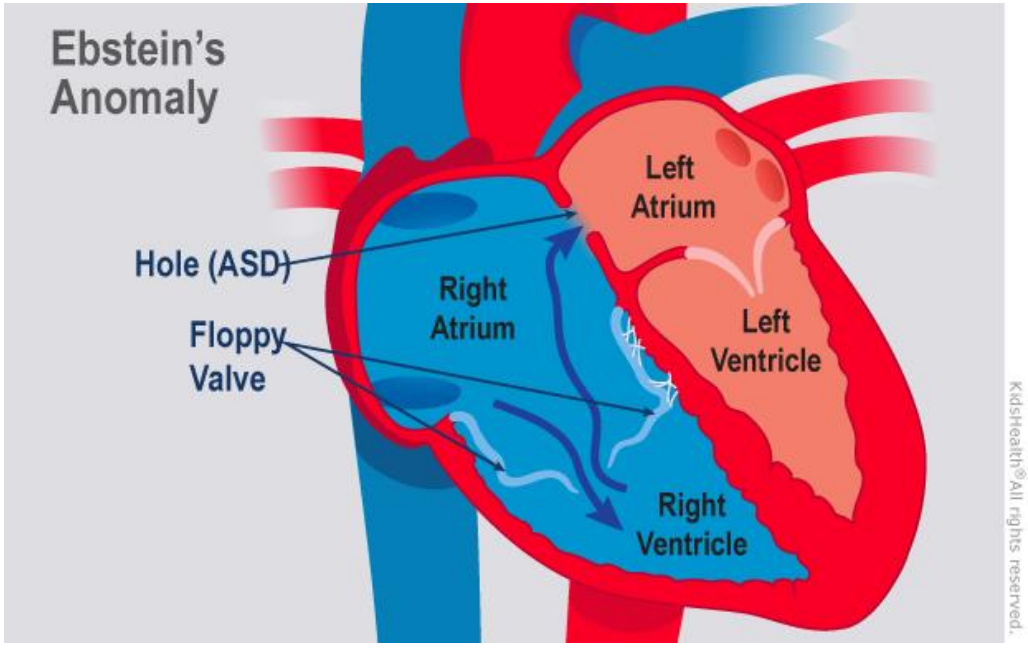
Snowman = TAPVR



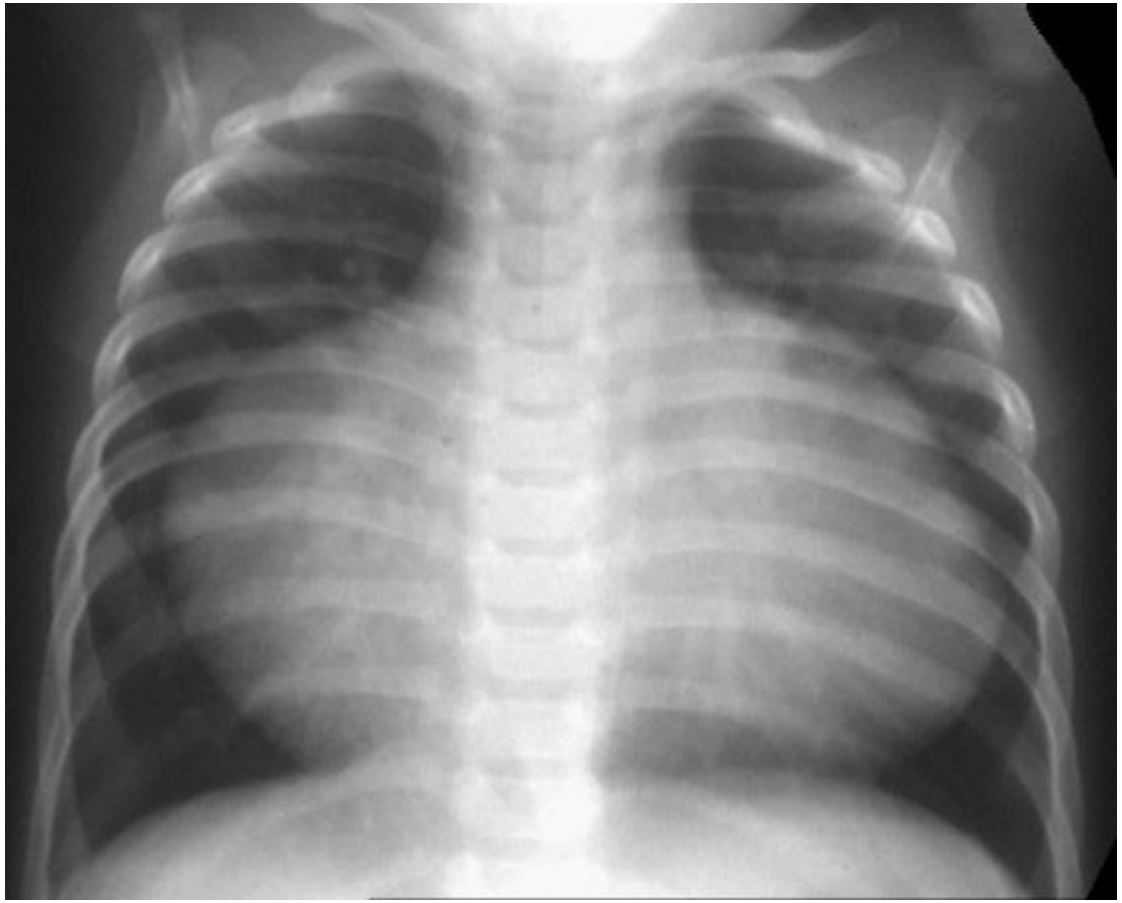
Ebstein anomaly

- **Ebstein anomaly** is a rare **heart defect** in which the tricuspid valve — the valve between the upper right chamber (right atrium) and the lower right chamber (right ventricle) of the **heart** — isn't formed properly. As a result, blood leaks back through the valve and into the right atrium.
- Dysplasia of the tricuspid valve

causing regurge into the right atrium



- In **Ebstein anomaly**, the tricuspid valve sits lower than normal in the right ventricle. This makes it so that a portion of the right ventricle becomes part of the right atrium, causing the right atrium to enlarge and not work properly(no enough blood flow to the pulmonary circulation)-oligemic lungs

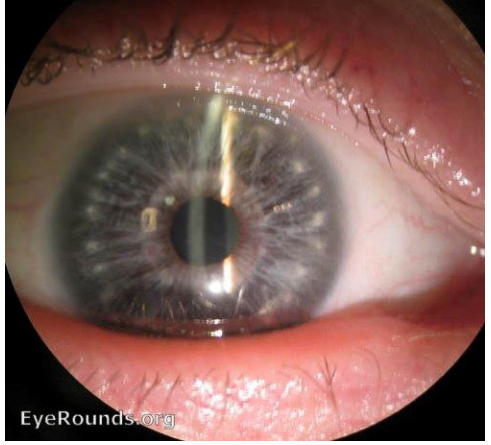


Severely dilated right atrium, dark (oligemic lungs), Ebstein anomaly of the tricuspid valve

Coarctation of the aorta

- Coarctation of aorta Difference in pulse between upper and lower extremities(radiofemoral delay), hypertension in upper extremities, congestive heart failure.
- The typical heart **murmur** that is associated with a **coarctation** is a systolic **murmur** that is loudest in the back below the left shoulder blade (scapula).
- Coarctation of aorta Heart failure, signs of **ischemia** to organs and extremities.(delayed capillary refill and cold extremities and weak peripheral pulses)

Syndromes associated with CHD

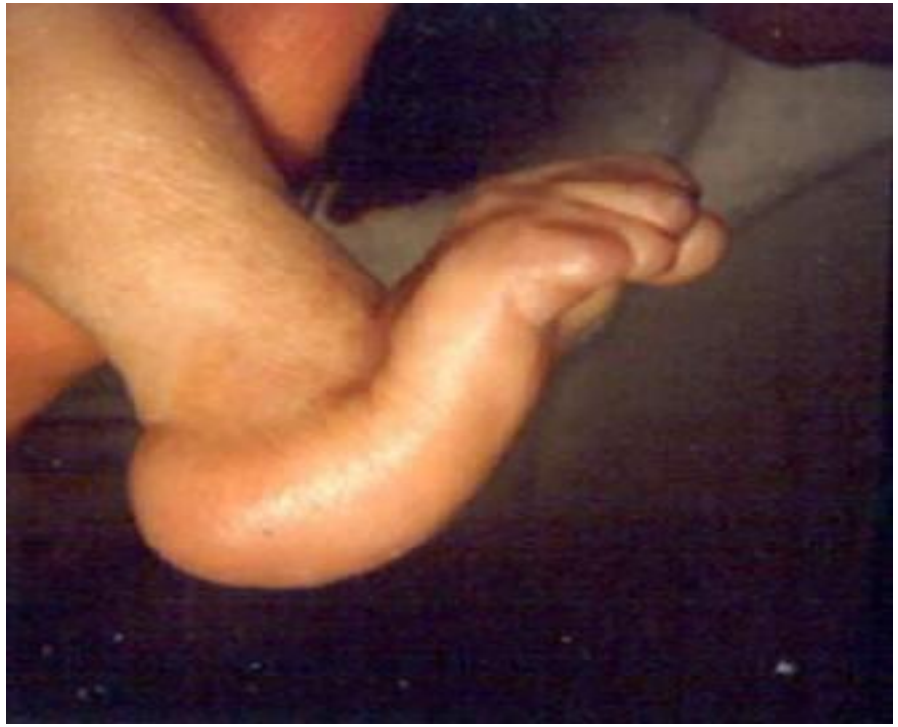


- Trisomies
  1. Trisomy 21;AV-canal
     - H&N findings (upslanting eyes,brush-field spots,webbed neck,low set ears,wide nasal bridge,epicanthal folds)
     - Hand findings; **Clinodactyly** curvature of a digit mainly pinkie, **Simian crease** is single transverse palmar **crease,short fingers.**
     - GI diseases; Duodenal atresia , annular pancreas , hirschsprung disease, imperforated anus.
  2. Trisomy 18;VSD,ASD(EDWARD syndrome)
  3. Trisomy 13;VSD,ASD,PDA
- SYNDROMES
  1. Digeorge
  2. Turner
  3. William's
  4. Noonan
- Atrioventricular septal defect (AVSD), or AV canal, is a heart defect that involves the valves between the heart's upper and lower chambers and the walls between the chambers. Other terms used to describe this problem include endocardial cushion defect and AV canal defect.

EDWARDS SYNDROME(trisomy 18)

- Findings;short sternum,widely spaced nipples,hypertelorism, Rocker bottom feet.
- Other non specific signs include;microcephaly,low set ears
- Cardiac deformity; AVSD. Or

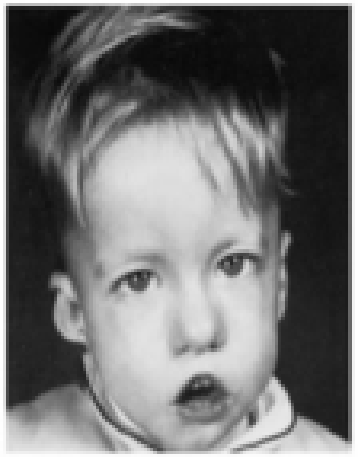
Endocardial cushion defect.

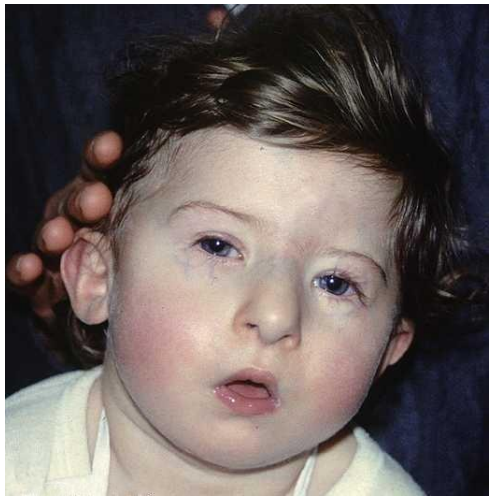


Genetic Abnormalities
DiGeorge Syndrome (22q11)

- Features vary widely
- **CATCH 22**
- **C**ardiac abnormality (interrupted aortic arch, truncus arteriosus, TOF)
  Abnormal facies
  Thymic aplasia
  Cleft palate
  Hypocalcemia/**H**ypoparathyroidism

DiGeorge Syndrome





The College of Osteopathic Medicine of the Pacific
The discipline of learning. The art of caring.

Cardiac abnormalities

Truncus arteriosus,

TOF.(because its 22 it

Has 2 Ts of the main

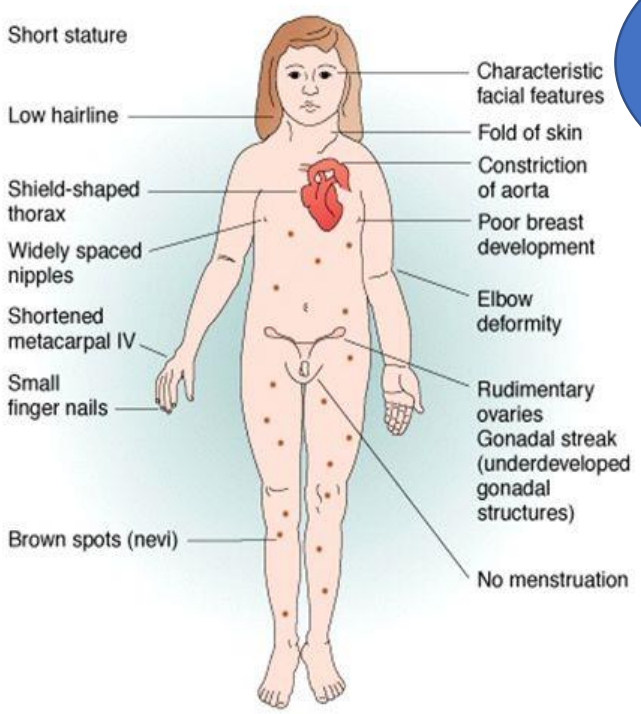
Cyanotic lesions)

Patients with thymic aplasia,the thymus is the primary organ for t cell maturation,so thymic aplasia patients have tcell deficiency,primary immune-deficiency

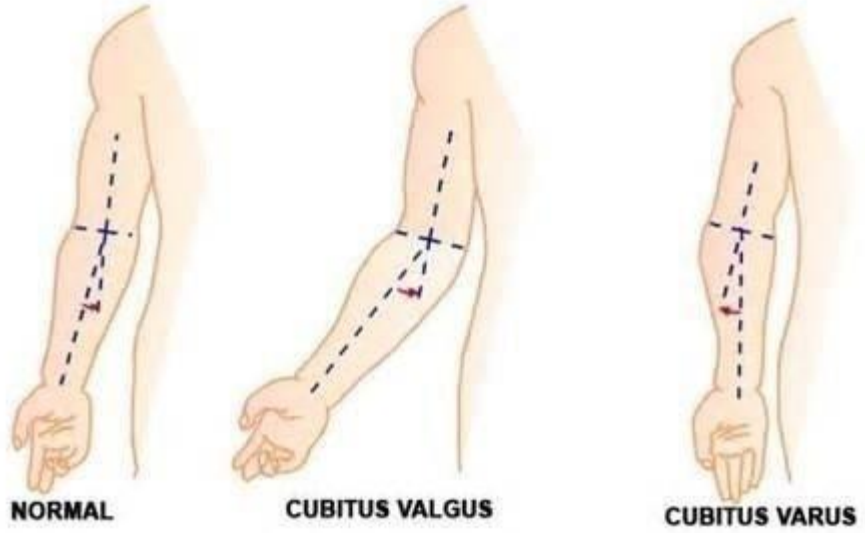
Turner syndrome(monosomy X)

"TURN THE AORTA ON"
Aortic anomalies; bicuspid aortic valve (BAV), **coarctation of the aorta**, and thoracic aortic aneurysm.

- Epicanthal folds
- Low posterior hair line
- Cubitus valgus
- Broad chest with widely spaced nipples
- Cardiovascular anomalies
- Hyperconvex finger nails
- Pigmented nevi
- Sex chromatin negative



Horse-shoe kidney

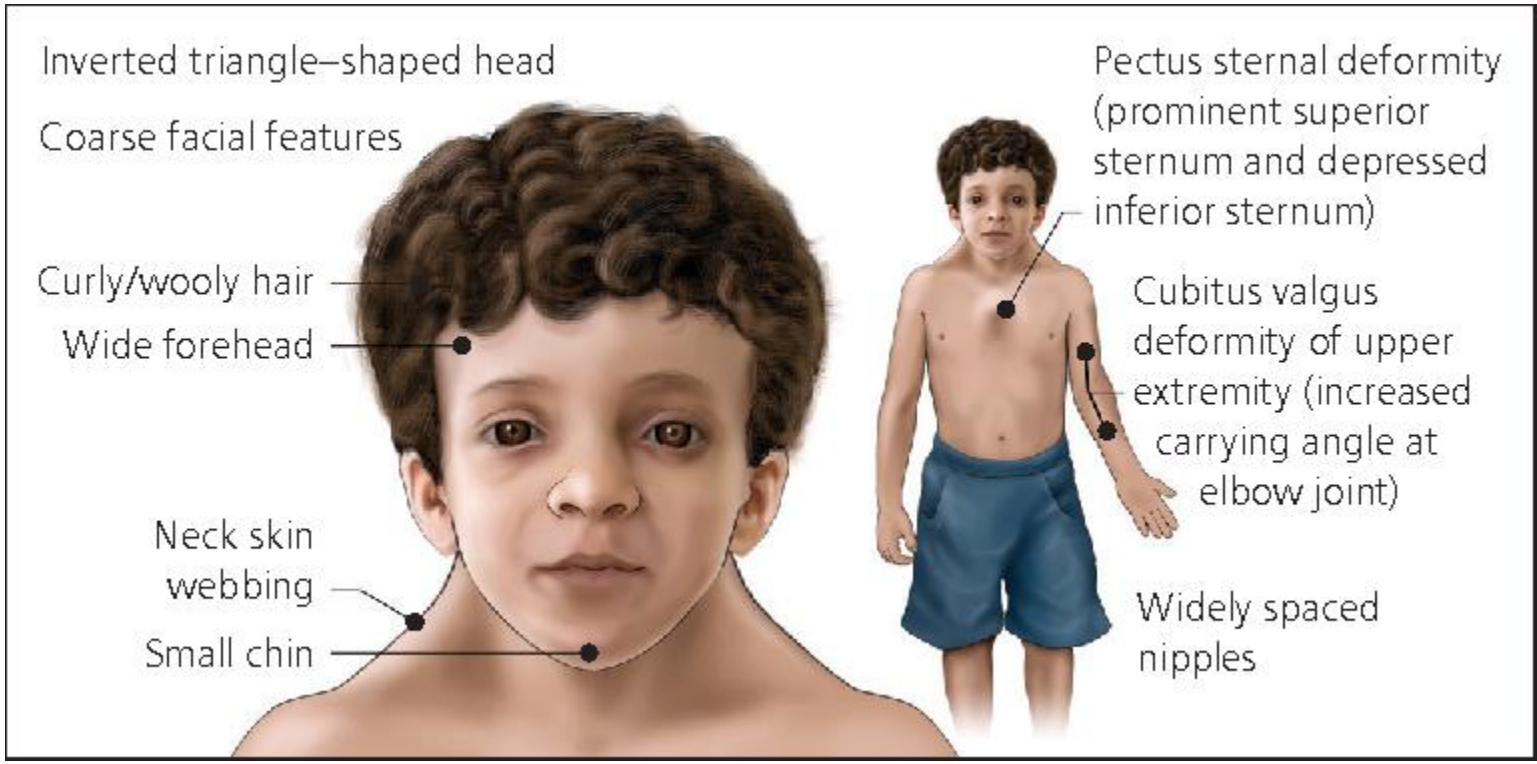


Williams syndrome(WIDE SMILE BUT STENOTIC LESIONS)

- Ear to ear smile/wide mouth
- Small and widespread teeth
- Cardiac defects;supra aortic stenosis,pulmonary artery stenosis.
- A small deletion of chromosome 7q11.

Noonan syndrome

- Widely spaced nipples
- Webbed neck
- Inverted triangle shaped face
- Cubitus valgus
- Autosommal dominant
- Heart defect; **dysplastic pulmonary valve**
- Sometimes; described as “the male version of Turner’s syndrome”.



Disease/feature	Chromosomal defect	General features	Type of heart defect
Trisomy 21(down syndrome)	3 pairs of chromosome 21(increased risk of ALL,AML,hypotonic)	Upslantin eyes,epicanthal folds,low set ears,webbed neck,brush-field spots in eyes	AVSD or endocardial cushion defect(AV-canal mainly),
Trisomy 18(EDWARDS SYNDROME)	3 pairs of chromosome 18	Short sternum,hypertelorism,microcephaly,rocker bottom feet	AVSD or endocardial cushion defect(ASD,VSD,PDA)
Trisomy 13	3 pairs of chromosome 13	-	AVSD or endocardial cushion defect(ASD,VSD,PDA)
DiGeorge syndrome	Deletion 22q11	CATCH-22,cleft palate,hypocalcemia and hypopara,thymic dysplasia	2 of the main cyanotic lesions.TOF,Truncus arteriosus
Truner's syndrome	One x chromosome,no other sex chromosome,monosomy x	Cubitus valgus,widely spaced nipples,low hair line,pigmented nevi,lymphedema,horse shoe kidney	TURN THE AORTA ON Coarctation of the aorta,thoracic aorta aneurysm,bicuspid Aortic valv
Williams syndrome	Deletion 7q11	Wide smile(ear to ear),small and widely spaced teeth	Wide smile but stenotic vessels,supra aortic stenosis,PA stenosis
Noonan syndrome(male	Autosomal dominant genetic	Cubitus valgus,webbed	Dysplastic pulmonary valve

Normal values

Age	RR	HR	BP(usually plotted on curves>95 hypertensive,<5 hypotensive,90-95 prehypertensive
Neonates	60	Abnormal above 220	>130/80 hypertension regardless the age
1 year	40	Max HR=220-age	
1-3 years	35		
3-5 years	30		
Early childhood	25		
adolescents	12-20		

Think about the possible etiologies HF in this patient! (Two month old at presentation)

CHD with increased pulmonary blood flow	CHD with systemic flow obstruction	Poor myocardial contractility	HF due to dysrrhythmia
• VSD (holosystolic murmur) • PDA (machinery murmur, wide pulse pressure,bounding peripheral pulses) • AV canal defect (Down syndrome) • Truncus arteriosus (mild desaturation, possible ejection click, wide pulse pressure)	• Aortic stenosis (ejection click with systolic murmur, radiation to the neck) • Coarctation of aorta (high blood pressure in upper extremities, poor femoral pulses, radiofemoral delay,systolic murmur)	• Dilated cardiomyopathy (family history) • Myocarditis (hx of viral infection)	• SVT (HR >220) • Bradycardia (complete heart block), congenital CHB presents earlier

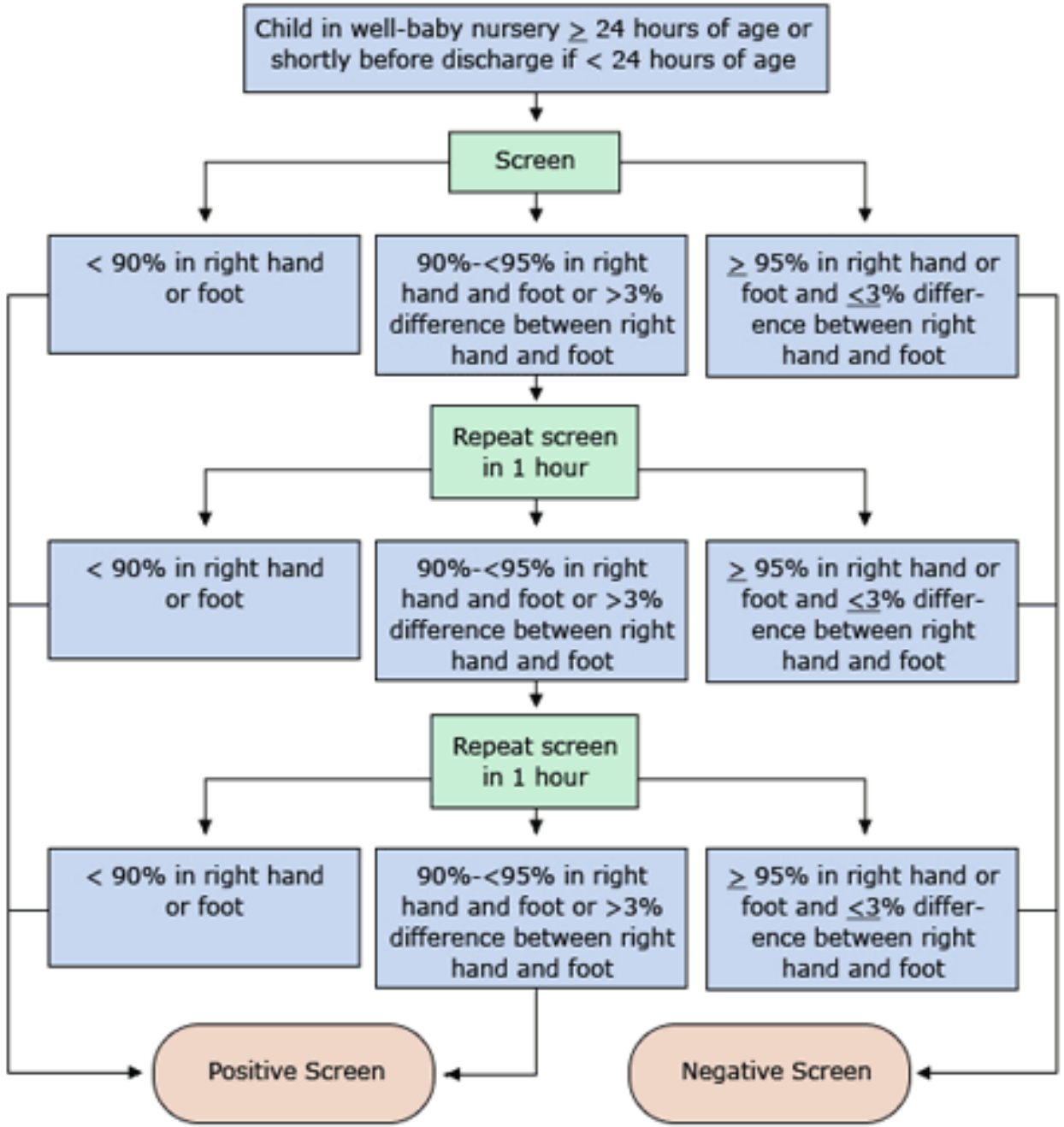
These are some examples of causes of heart failure in a two month old
The most common is CHD with increased pulmonary blood flow

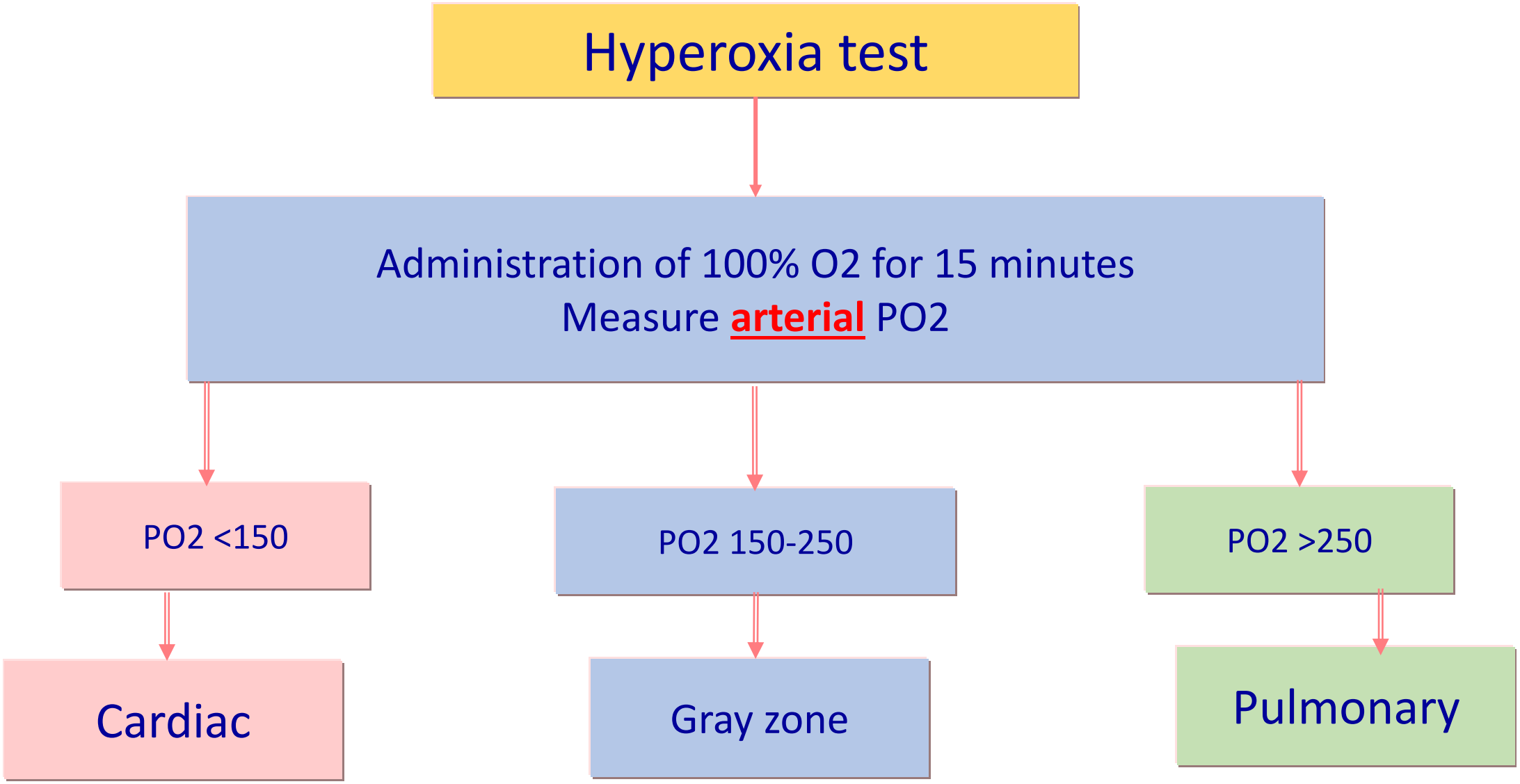
- Lesions with HF Sx: ASD,VSD,PDA,Truncus arteriosis.
- Lesions with cyanosis: TGA>TOF>TRICUSPID ATRESIA>TAPVR(can present with HF Sx).
- Lesion associated with TOF: VSD.
- Lesion associated with TGA:ASD,PFO if there's PDA a better condition and an indication to keep it patent for better mixing.
- Lesion associated with tricuspid atresia:born with a PFO,they need either a PDA or VSD to survive.
- TAPVR:they need a PFO to survive.
- Ebstein anomaly :dysplastic tricuspid valve.
- Lesions with oligemic lung field:TOF,pulmonary atresia(noonan syndrome associated with dysplastic pulmonary valve),ebstein anomaly,tricuspid atresia.

You diagnosed this infant with HF secondary to a large VSD. What are the lines of management?

Nutritional support	Increase caloric intake (fortified formulas, more frequent feeds, NG feeding if needed)
Medications	Diuretic (furosemide): decrease the congestion, improves respiratory distress Afterload reduction (captopril) ACE-inhibitor: decrease the amount of left to right shunting Inotrope (digoxin) increases the contractility: rarely needed, helps if systolic function is depressed
Surgery	Surgical paliation (Pulmonary artery constriction by a band for temporary relief of symptoms if correction cannot be done) Surgical repair (VSD closure) is the definitive therapy

- General notes;
  - Hypoglycemic seizures in babies of diabetic mothers is common.
  - Tachypnea and tachycardia are early signs of sepsis in addition to fever,hpotension is a late sign of sepsis(the baby is shocked)
  - If you suspect sepsis ask about associated rashes.
  - Note: pulse ox should be measured in right arm (preductal), and in lower limbs (post-ductal)-see next slide for assessment.
  - Cyanosis is not always apparent to eyes,so always do a pulse oxy,in addition anemia can make it less possible to be apparent,polycythemia makes it easier to be seen.
  - **Detection of cyanosis depends on the amount of de-oxyhemoglobin in blood. 3-5 grams of de-oxyhemoglobin are needed.**


Child in well-baby nursery ≥ 24 hours of age or shortly before discharge if < 24 hours of age
Screen
< 90% in right hand or foot
90%-<95% in right hand and foot or >3% difference between right hand and foot
≥ 95% in right hand or foot and ≤3% difference between right hand and foot
Repeat screen in 1 hour
< 90% in right hand or foot
90%-<95% in right hand and foot or >3% difference between right hand and foot
≥ 95% in right hand or foot and ≤3% difference between right hand and foot
Repeat screen in 1 hour
< 90% in right hand or foot
90%-<95% in right hand and foot or >3% difference between right hand and foot
≥ 95% in right hand or foot and ≤3% difference between right hand and foot
Positive Screen
Negative Screen


Hyperoxia test
Administration of 100% O2 for 15 minutes
Measure arterial PO2
PO2 <150
PO2 150-250
PO2 >250
Cardiac
Gray zone
Pulmonary

- Eisenmenger's syndrome is defined as the process in which a long-standing left-to-right cardiac shunt caused by a congenital heart defect (typically by a ventricular septal defect, atrial septal defect, or less commonly, patent ductus arteriosus) causes pulmonary hypertension[1][2] and eventual reversal of the shunt into a cyanotic right-to-left shunt.

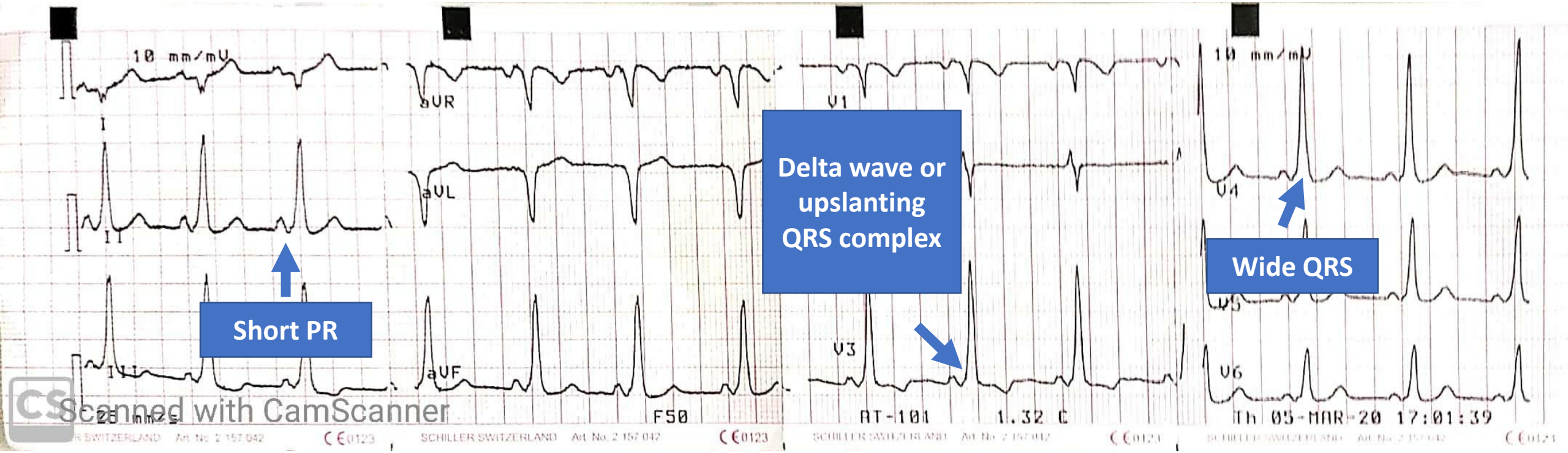
General quick tips when reading an ECG

- Small square is .04,large square is .2
- Rhythm;regular distance between r-r intervals
- Rate;300/number of large squares between r-r intervals
- Axis;lead 1 and 2
- PR interval(.12-.2 secs) 3-5 small squares,above or below abnormal
- Qrs(<.12,less than 3 small squares)
- QT interval should be corrected to the heart rate
  - Qt time in seconds/root square of r-r interval in secs
  - Normal value is .4-.44 secs
  - Rough estimation the qt should end before half way between r-r

Paroxysmal arrythmias

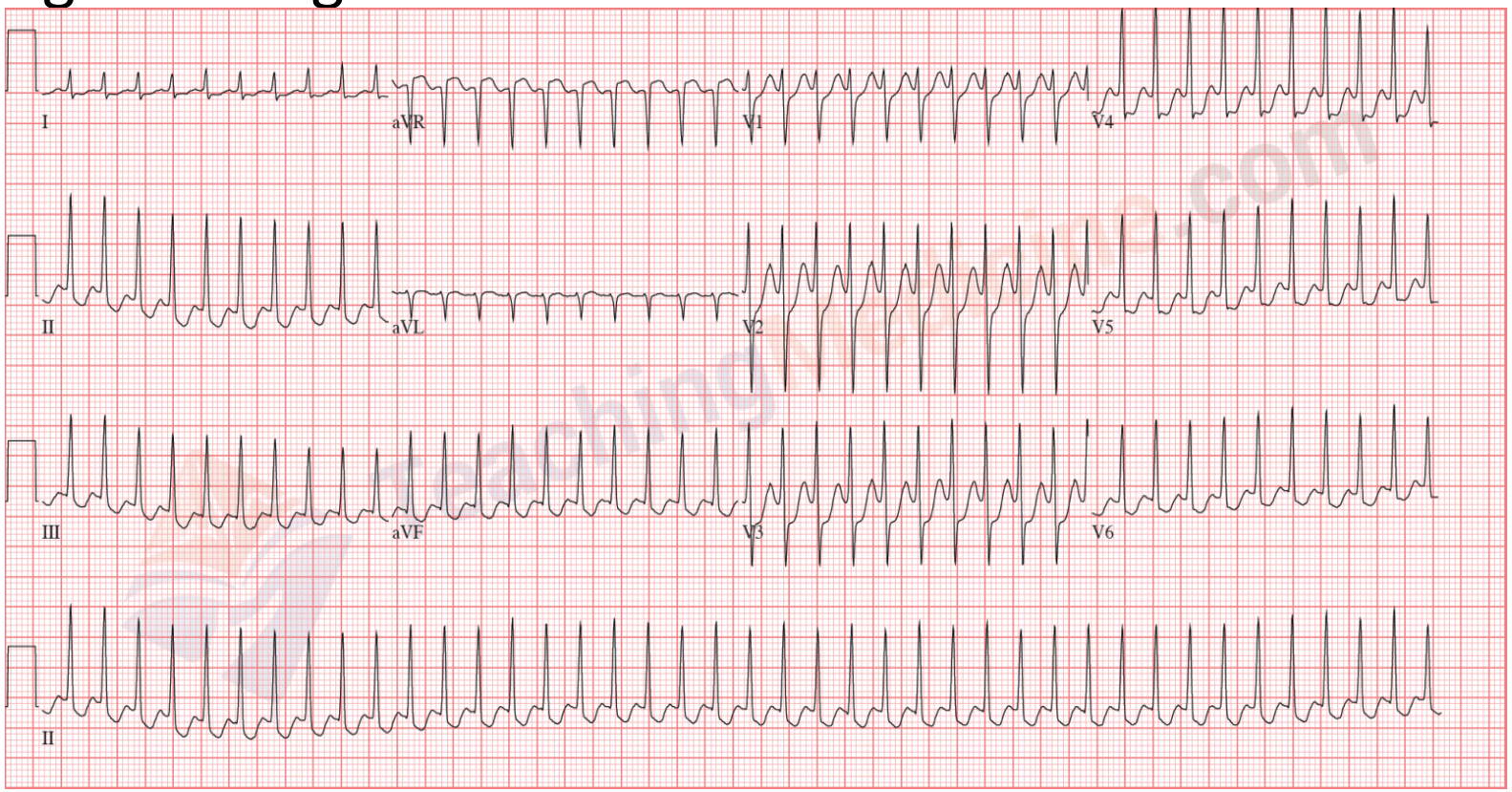
- Paroxysmal arrythmias are arrythmias that happen occasionally from time to time.(ask about asthma medications,recent surgery for bleeding-anemia,anxiety,exercise-related,symptoms of hyperthyroid or pheo,symptoms of anemia)
- What supports the history of 1ry arrythmia
    1. Sudden onset and sudden cessation
    2. Episodes occur at rest
    3. Presyncope /syncope
- What supports the history of cardiomyopathy?
    1. Family history of sudden cardiac death or diagnosed myopathies
    2. Symptoms of heart failure
    3. Related to exercise

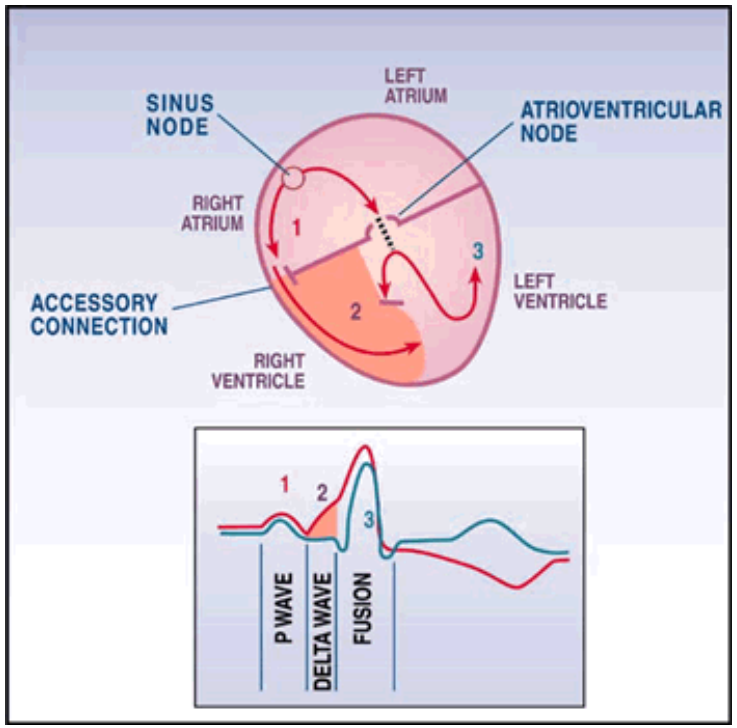
What is your impression? These signs only seen when in sinus rhythm,when they're in SVT there is no signs on the ECG



Wolffe-Parkinson-White (WPW) Syndrome

- WPW patients are usually in sinus rhythm,unless something happens and one of the pathways is blocked usually the accessory pathway,then the impulse will go back to the atria thru the accessory pathway(finished the refractory period and not blocked anymore),causing SVT.
- The ECG signs of WPW are only seen in the sinus rhythm,there are no signs during the SVT.



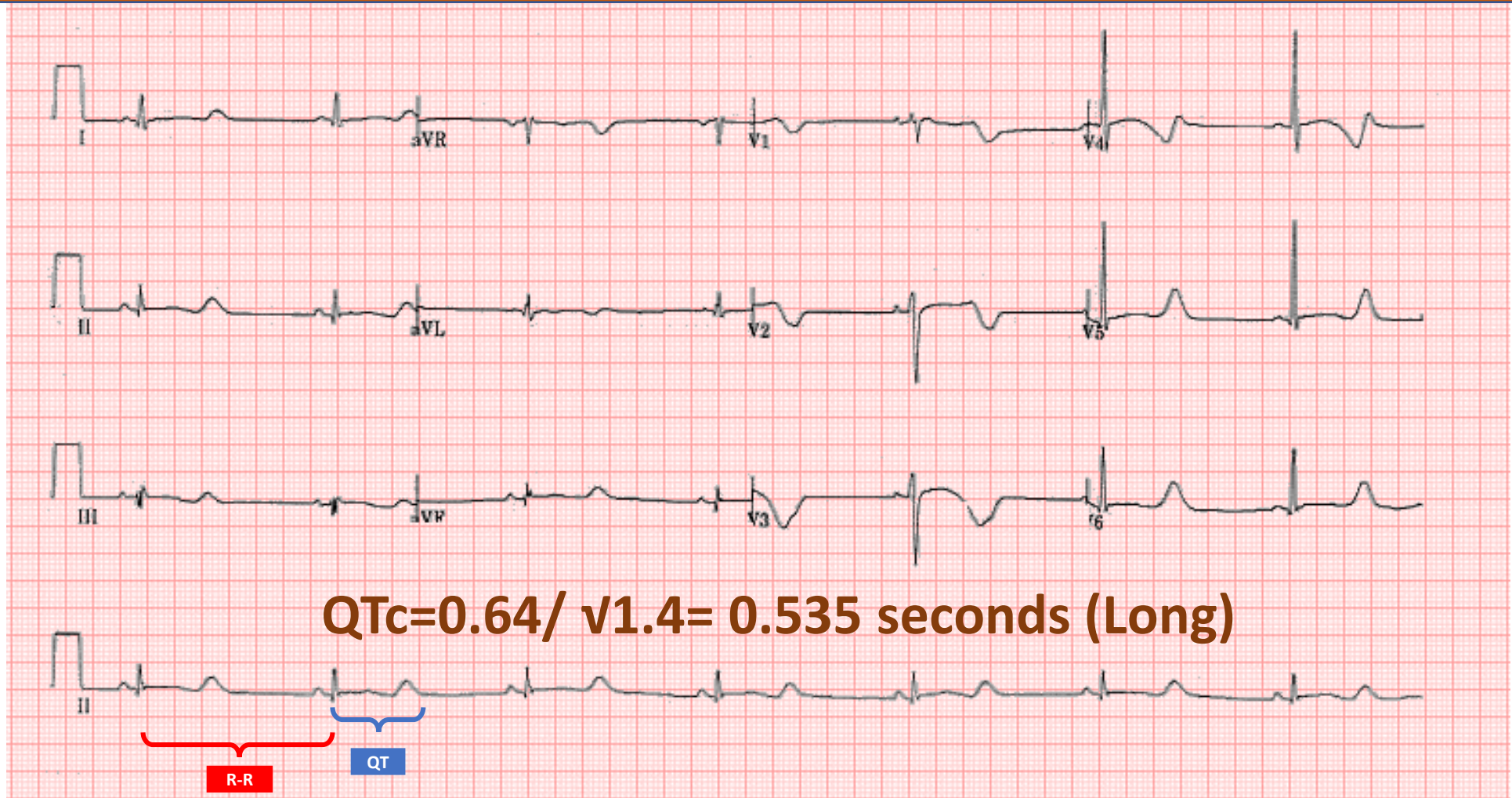


How to assess patient with tachycardia

1. Sinus rhythm tachycardia;doesn’t exceed 220 bpm,max heart rate=220-age.
2. SVT;narrow complex tachycardia(HR>220)
3. Ventricular tachycardia;wide complex(HR>220)

- Managing patient with tachy cardia;ABCS,give oxygen,IV access,12-lead ECG,assess the rhythm.
- Patient is hemodynamically stable; try vagal manevuers like Valsalva,IV adenosine,unstable give synchronized cardioversion(this applies for wide and narrow QRS,difference is in wide complex we can consider amiodarone)

If the child had history of palpitation and syncopal episode, with family history of sudden death, and this is the ECG. What is the diagnosis that explains the history?



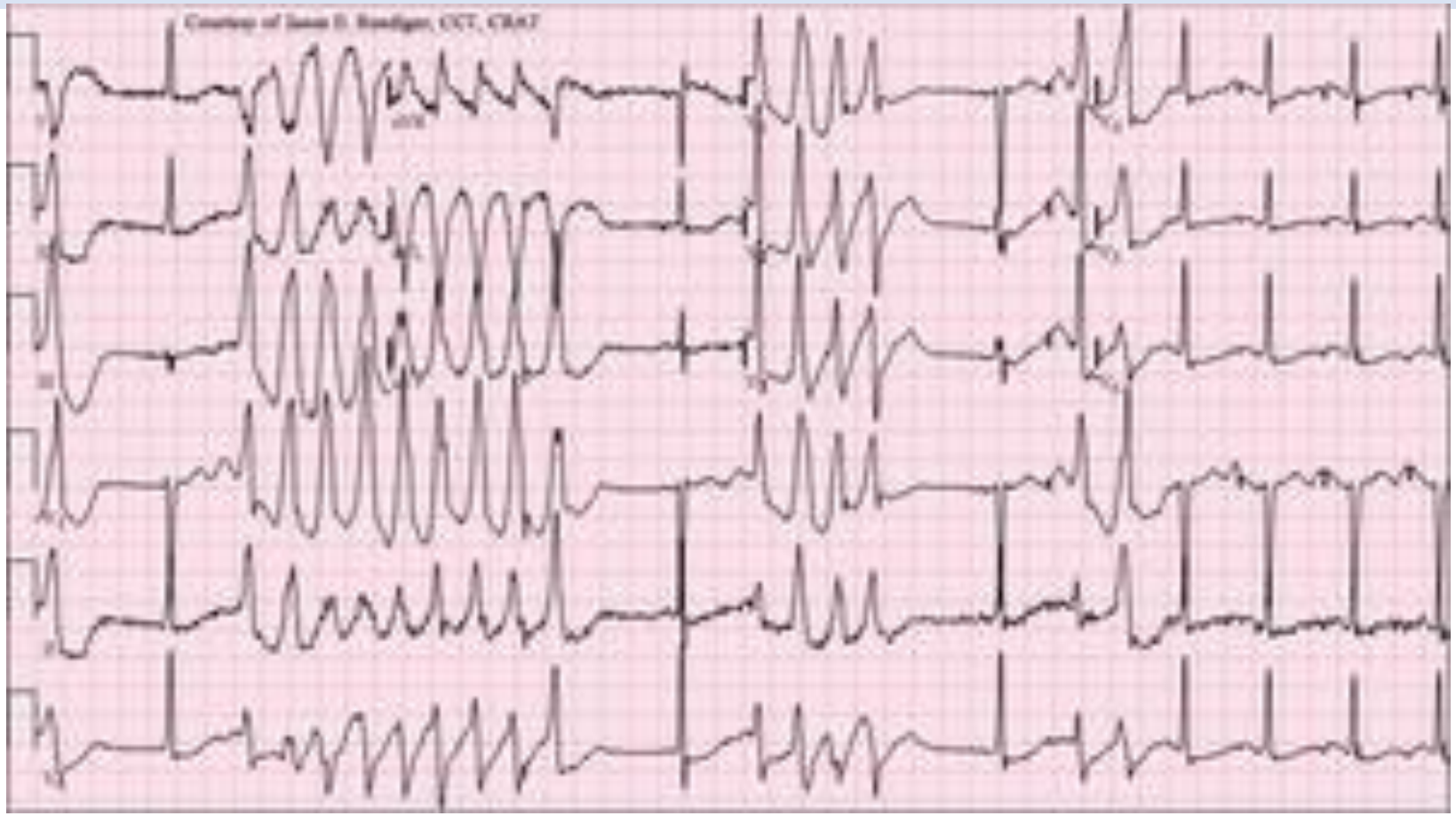
QTc=0.64/ √1.4= 0.535 seconds (Long)

Long QT Syndrome
How do you calculate the corrected QT?

Bazett formula
Corrected QT (QTc)= QT/√ R-R

How does long QT cause syncope, or sudden death?

- Episodes of Torsade de Pointe,polymorphic ventricular tachycardia



Approach to a heart murmur in children

Murmur due to structural heart disease (pathological murmur)

Any abnormality in the cardiac structure resulting in turbulence of flow. Examples:

- Septal defect
- Abnormal valve (stenotic, regurgitant)
- Abnormal vessel (stenotic vessel)
- Dynamic obstruction (as HOCM)

Functional murmur due to non-cardiac pathology (pathological murmur)

Any abnormality that results in increased flow across normal valves. Examples:

- Anemia
- Fever
- Sepsis
- Hyperthyroidism

(all are high output states)

Innocent murmur with no pathology

Murmur caused be normal flow in certain situations, very common in children.

Examples:

- Still's murmur (musical vibratory sound)
- Pulmonary or aortic flow murmurs (increase in exercise or anxiety)
- Venous hum
- Subclavian/internal mammary souffle in pregnancy

- Still's murmur is a common type of benign or "innocent" functional heart murmur that is not associated with any sort of cardiac disorder or any other medical condition. It can occur at any age although it is most common among children two to seven years of age and it is rare in adulthood.musical vibratory murmur that disappears in standing position.
- The venous hum, a continuous murmur usually of maximum intensity in the supraclavicular area, is a common auscultatory finding. Heard particularly in children, it is caused by vibration of the walls of the internal jugular veins.
- A **mammary souffle(internal mammary or subclavian murmur)** is a continuous murmur with a soft, humming quality. It is typically heard over the breast during late **pregnancy** and lactation and is thought to result from increased blood flow to the breast of no known pathological significance.

So, we have to tailor the history towards ruling out cardiac pathology, and high output states.

Cardiac Pathology

- Asking about symptoms of heart failure (exercise capacity, respiratory symptoms, Cyanotic heart disease symptoms- cyanosis)
- Asking about symptoms of HOCM (chest pain, syncope, palpitation), Hypertrophic obstructive cardiomyopathy.
- Family history of cardiomyopathies, sudden death, congenital malformations.. etc

High output states

- Anemia (Pallor, nutritional history, growth, hemolysis(family history of hemolysis, malabsorption, family history… etc.)
- Fever, sepsis, recurrent infections
- Hyperthyroidism symptoms

How does the story help you in differential diagnosis? The following is example of critical thinking

- **Structural heart disease?** Many structural heart disease produce symptoms early on during infancy and early childhood. Complex heart disease present in the neonatal period with either cyanosis or heart failure. However some MILD structural heart disease may not present with any symptoms. (examples: atrial septal defects, small ventricular septal defects, small ductus arteriosus, mild aortic or pulmonary valve stenosis)
- **Acquired heart disease and cardiomyopathy?** Acquired heart disease usually present with disease specific symptoms (endocarditis, myocarditis, cardiomyopathy). So it is unlikely to present as an incidental heart murmur. Keep in mind that HOCM may present with a heart murmur in an asymptomatic child, but it is still not a common disease at this age.
- **High output state?** Anemia is common in children, and sometimes it is asymptomatic if mild.
- **Innocent murmur?** These are very common in children, but the diagnosis should only be made by exclusion. So we cannot depend on the history only to diagnose innocent murmur. Physical exam is extremely important.

Evaluation of a heart murmur

- There are four objective points in evaluation of a heart murmur that **must** be described:
  - Location (where is the murmur best heard)
  - Radiation if present (neck, back, or axilla)
  - Grade (i.e loudness)→ (2/6: soft, 3/6 loud with no thrill, 4-6/6 loud with thrill)
  - Systolic, diastolic, continuous (systolic corresponds with pulse)
- Other features of the murmur can also be described, and may require some experience
  - Specific timing (early or late systolic, holosystolic, early or mid-diastolic)
  - Pitch (high pitch vs low pitch)
  - Shape (crescendo de-crescendo..)
  - Quality of sound (blowing, musical, harch)
- Evaluate the presence of associated additional cardiac sounds (S3, S4, Clicks, snaps, Splitting).
- Maneuvers: murmurs can be manipulated through several maneuvers that can help in the diagnosis

- **Question: Describe maneuvers and their effect on physiology and murmurs.**

The murmur of mitral regurge is pansystolic murmur with axillary radiation.(same as VSD but with radiation to the axilla)

Murmur of mitral stenosis is opening snap diastolic rumble murmur

aortic stenosis murmur is a midsystolic murmur with ejection systolic click best heard at the pulmonic area –right upper sternal border- and radiation to the coronary artery

- There is a soft 2/6 systolic murmur at the left upper sternal border, with no radiation. Murmur is midsystolic.
- The second heart sound is split, and it does not vary with inspiration or expiration.
- Rest of exam is normal

Diagnosis?

ASD

- There is a grade 4/6 systolic murmur at the apex, with no radiation. Holosystolic murmur.
- Thrill at the apex palpated
- Rest of the physical exam is normal

Diagnosis?

Small VSD

- There is a 3/6 systolic murmur at the right upper sternal border, radiates to the carotid artery. Murmur is midsystolic.
- There is ejection systolic click.
- Rest of exam is normal
- The aortic stenosis murmur is best heard at the aortic area,right upper sternal and is midsystolic with ejection click

Diagnosis?

Bicuspid aortic valve with aortic stenosis

- There is a soft 2/6 systolic murmur at the left sternal border, with no radiation. Murmur is musical, vibratory in nature
- The murmur becomes not audible in standing position
- Rest of physical exam in normal

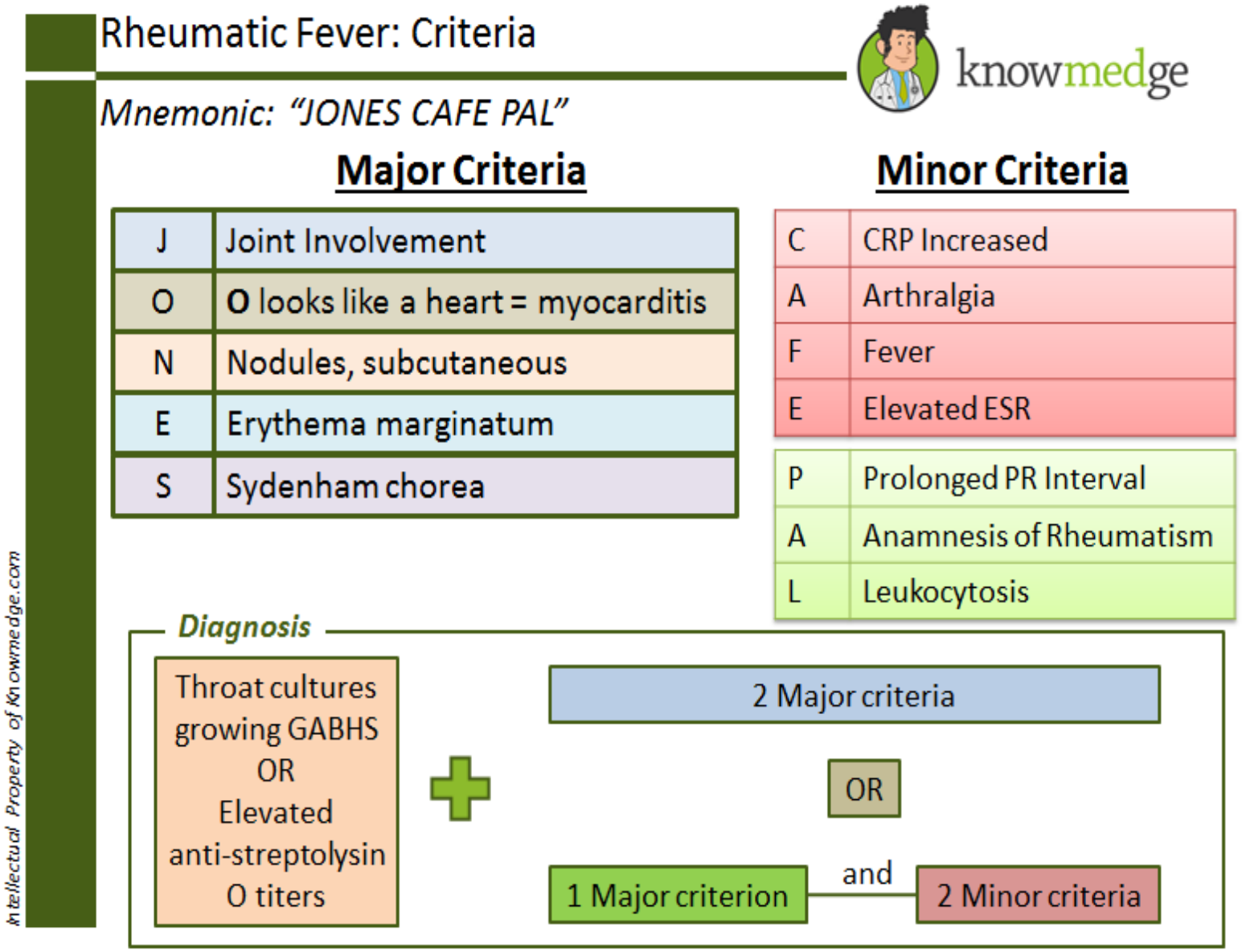
Diagnosis?

Innocent murmur

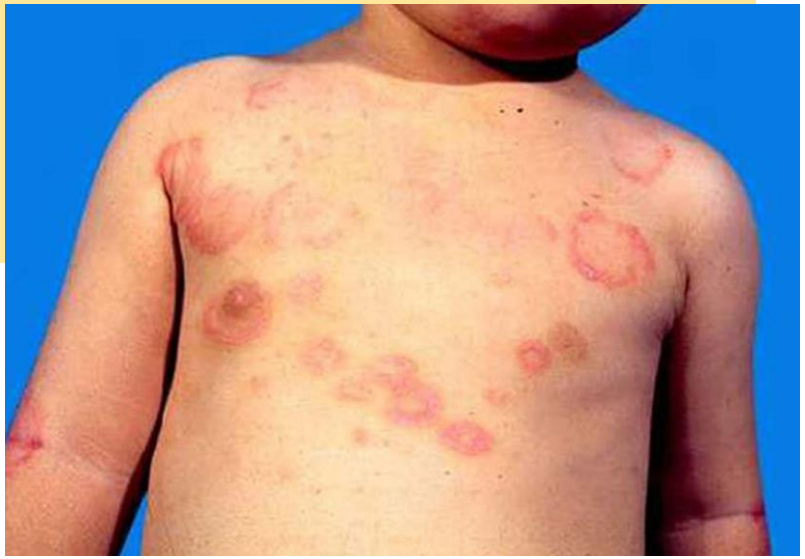
Differential diagnosis of joint pain and swelling

- **Septic arthritis is less likely due to more than one joint involvement.(number of joints involves?),goes with involvement of one joint.**
- **Reactive arthritis is a possibility, usually they are due to concomitant viral infection(recent viral infection?),associated with immune response to gastroenteric or genitourinary organisms(salmonella,shigella,yersinia,chlamydia),HLA-B27,Characterized by(conjunctivitis,arthritis,urethritis),can't pee cant see cant climb the tree**
- **Trauma is ruled out by history, hemarthrosis usually occur with previous history of recurrent manifestation of bleeding disorder, and with positive family history of hemophilia.(personal or family history of bleeding disorders?)**
- **Connective tissue disorders are not diagnosed with acute presentations. In addition, there are usually associated with extra-articular manifestations. (extra-articular organ involvement, including the skin, eye, heart, lung, renal, nervous and gastrointestinal systems?)**
- **Henoch-schönlein purpura is a disease in younger children and has to have skin rash in the lower extremities.(skin rash?)**
- **Rheumatic fever is a strong possibility based on the story, particularly with the past history of pharyngitis.**

Arthralgia (in the absence of arthritis) anamnesis of rheumatism; Previous rheumatic fever



- Arthritis: very painful, migratory, involving large joints, non-destructive.
- Carditis: presents as congestive heart failure, new murmur, friction rub of pericarditis.
- Subcutaneous nodule: non-tender nodules on the extensor surfaces of joints (very infrequent).
- Erythema marginatum: Long lasting skin rash on the trunk or limbs
- Chorea: can be very late in the disease



- ECG findings
  1. Bifid, biphasic p wave aka p.mitrale in lead 2

  and V1 consistent with left atrial dilatation or hypertrophy because of the mitral regure.

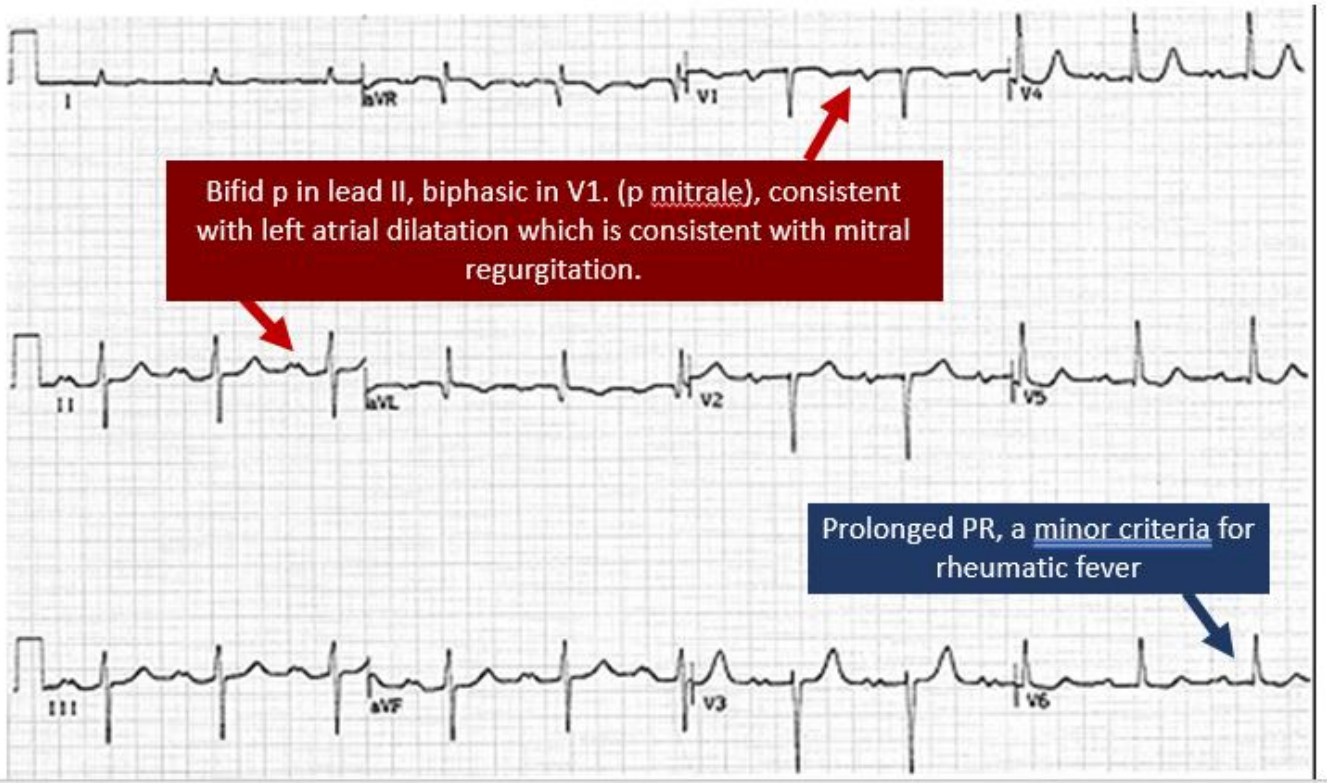
  1. Prolonged PR interval-one of the minor criteria

Carditis (50%)
Endocarditis
- significant murmur
- valvular dysfunction

Myocarditis
- may lead to heart failure and death

Pericarditis
- pericardial friction rub
- pericardial effusion
- tamponade

What are the two findings in this ECG, and how do you explain it in view of Rheumatic fever?



- Bifid aka biphasic p wave aka p.mitrale is consistent with left atrial defect,dilatation or hypertrophy
  remember the regurge is a volume overload it causes receiving hypertrophy and downward hypertrophy

•What is carditis in acute rheumatic fever?

- Inflammation can involve all cardiac layers, PANCARDITIS (endocardium, myocardium, and pericardium)
- Most likely affected valves: Mitral valve followed by aortic valve (usually mitral and aortic regurgitation)
- Mitral valve and aortic regurgitation may improve after treatment with anti-inflammatory medications
- With time mitral stenosis can develop many years after the acute rheumatic fever due to fibrosis. Follow up should be done on regular basis
- The murmur of mitral regurge is **pansystolic murmur with axillary radiation.**
- Murmur of mitral stenosis is **opening snap diastolic rumble murmur**

Cause and Pathophysiology of rheumatic fever

- Group A Beta hemolytic streptococcal pharyngitis
- ARF follows untreated pharyngitis by 2-6 weeks
- Most common age group 5-15 yrs
- No gender differences except for Sydenham's chorea → more females

- Antibody cross-reactivity (molecular mimicry)
- Cell wall of group A strept. Contains a highly antigenic protein (M protein)
- Anti- M proteins may cross react with cardiac and smooth muscle myosin inducing cytokine release and tissue destruction

Treatment

- Treat infection if active pharyngitis
- Anti-inflammatory (Aspirin (ASA), consider steroids if intolerant to aspirin
- Treat heart failure if present(decrease the preload by furosemide, decrease the afterload by an ACE inh., inotrope to increase contractility if there's left ventricular dysfunction by digoxin if present)
- For chorea, the use of valproic acid or haloperidol is usually effective

- Patients who had rheumatic fever should receive prophylaxis long acting penicillin injection every 3-4 weeks
  - If there is cardiac involvement➔ prophylaxis should be given well into adulthood
  - If there is no cardiac involvement➔ prophylaxis should be given for 5 years or till the age of 18 years

Infective endocarditis

1. Causes
    a. Bacterial infection, fungal infection
        i. MCC → Staphylococcus aureus then Streptococci viridans group & coagulase negative Staphylococci
2. Risk factors
    a. Valvular heart disease including rheumatic disease, congenital heart disease, artificial valves, hemodialysis, intravenous drug use, electronic pacemakers
3. Clinical signs
    a. General
        i. Fever
        ii. Anaemia and pallor → feeling tired
    b. MSS
        i. Skin
            1. Necrotic skin lesions
        ii. Arthritis/arthralgia
        iii. Hands
            1. Splinter haemorrhages in nail bed
            2. Clubbing (late)
    c. CNS

c. CNS
    i. Neurological signs from cerebral infarction
    ii. Eye → Retinal infarcts
d. CVS
    i. Changing cardiac signs → heart murmur
    ii. Splenomegaly
e. US → Haematuria (microscopic).

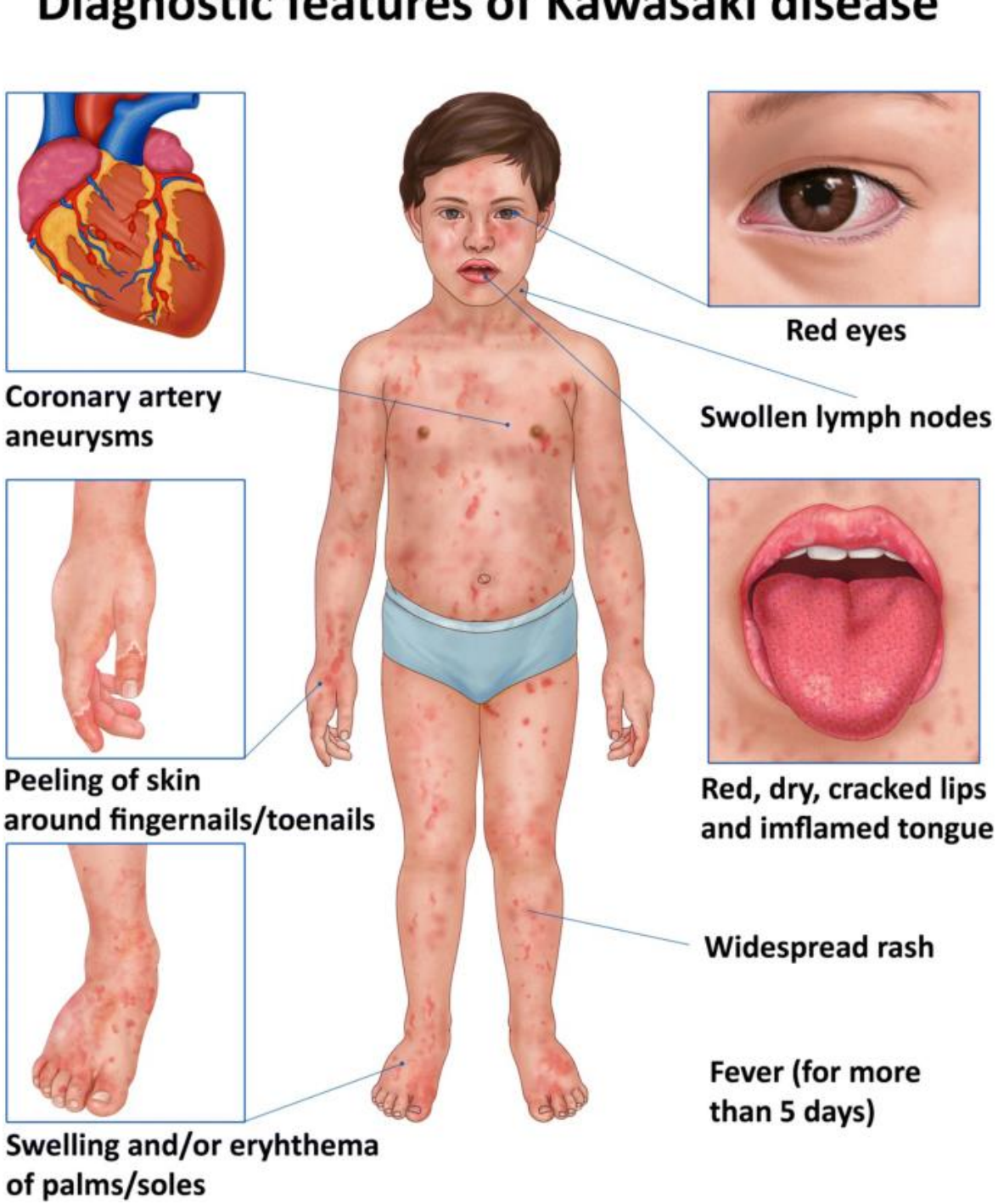
4. Diagnosis
    a. Multiple blood cultures should be taken before antibiotics are started
    b. cross-sectional echocardiography → vegetations
    c. Acute-phase reactants "ESR, CRP" → raised
5. Management
    a. Tx
        i. High-dose **penicillin** in combination with an **aminoglycoside**
        ii. Surgical removal→ If infected prosthetic material, e.g. prosthetic valves, VSD patches or shunts
    b. Prophylaxis
        i. Good dental hygiene in all children with congenital heart disease
        ii. Avoidance of body piercing and tattoos.
        iii. Antibiotic prophylaxis is no longer recommended
6. Complications → Valvular insufficiency, heart failure, stroke, kidney failure

Management is by high-dose penicillin in combination with an aminoglycoside, long term prophylaxis is no longer recommended, removal of the prosthesis or the foreign material if it is the cause, surgical fixation of the CHD if present.

- Findings in infective endocarditis from head to toe:
    - General:anemia,pallor,fever
    - CNS:cerebral infarcts,altered level of consciousness
    - Hands:clubbing,splinter hemorhhages
    - Head:retinal infarcts
    - Skin:necrotic skin lesions
    - CVS:heart murmur
    - Abdomen:Splenomegaly
    - Urogenital:hematuria
- Diagnosis is by blood multiple blood cultures before starting Abx, then start Abx,elevated ESR and CRP,Echo looking for vegetations.

Kawasaki disease

- **Kawasaki disease** (KD), or mucocutaneous lymph node **syndrome**, is an illness that causes inflammation in arteries, veins, and capillaries. It also affects your lymph nodes and causes symptoms in your nose, mouth, and throat. It's the most common cause of heart **disease** in children.



Kawasaki Disease Diagnosis Criteria

knowmedge

Mnemonic: "CREAM"

Criteria

Fever for greater than 5 days

+

4 out of 5 of the following

C	Conjunctivitis (non-exudative)
R	Rash (polymorphous non-vesicular)
E	Edema (or erythema of hands or feet)
A	Adenopathy (cervical, often unilateral)
M	Mucosal involvement (erythema or fissures or crusting)

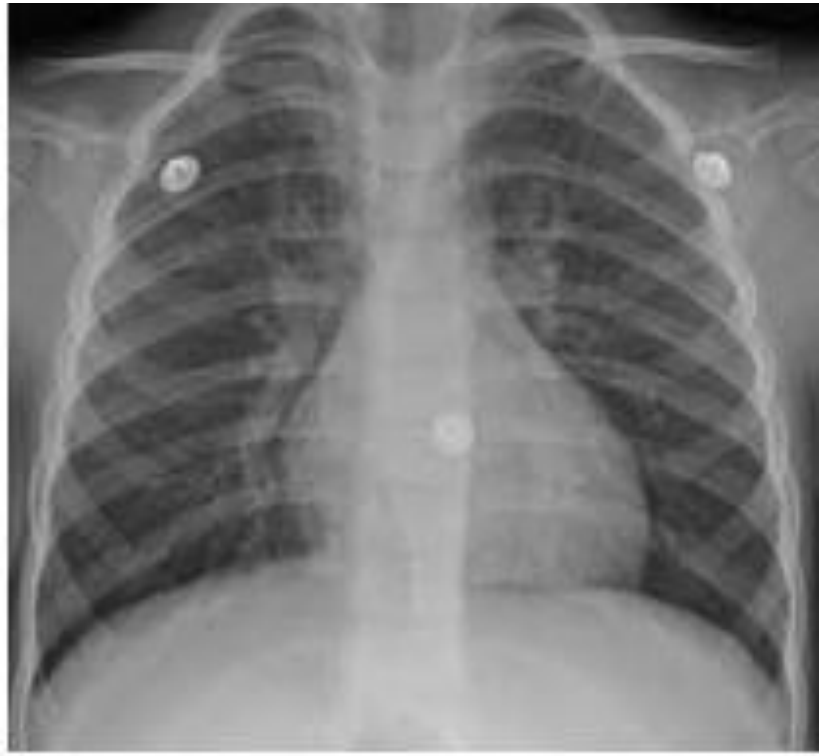
RESPIRATORY

Acute viral bronchiolitis

- RSV is the most causative agent (50% of cases)
- LRTIs include:bronchitis,bronchiolitis and pneumonia,so bronchiolitis is a LRTI,considered to be the most common LRTI,and most common indication for admission of infants in winter season.
- Definition:infection and inflammation in the bronchioles,it is most common in infants,most severe in infants 1-3months(smallest airways,under developed thoracic cage and respiratory muscles,weak immunity).
- Presentation;1.viral prodrome of URTI,nasal congestion or clear rhinorrhea,sneezing then low grade fever then gradual respiratory distress including wheeze cough SOB irritability(history)
- P/E findings;general finndings signs of respiratory distress depending on severity ranging from nasal flaring,tachypnea and tachycardia to retractions and increased work of breathing. Auscultatory findings;fine crackles(expiratory),wheezes(inspiratory),prolonged expiratory phase(because of the obstruction),hyperinflation of the chest because of obstruction so hyperresonant percussion note may be found.

- Risk factors for developing viral bronchiolitis(being of male gender,being in a crowded environment(siblings),non-breast fed because breast feeding provides passive immunity by passing Abs and probiotics)
- Babies are obligate nose-breathers so anything that causes obstruction to their nose will decrease the feeding.
- Patients with bronchiolitis are usually alert and interested in the surroundings,but may be agitated and irritated because of hunger or hypoxemia,but if they looks lethargic,tired,depressed this is an alarming sign that the baby is developing respiratory failure type 2(hypercapnic) because of prolonged hyperventilation,decreased breathing,co2 retention,so he got fatigued and now is retaining CO2.

- The diagnosis is clinically made,we don’t need radiology to confirm it,we only do a chest film in certain indications including; 1.the patient looks toxic for you to rule out pneumonia 2.very severe and needs ICU admission3.discrepancy of air entry between the two lungs.
- Usually they have excellent prognosis if we gave them the support within 4 days,but there are pathological risk factors that increase the seriouseness of the disease which are;1.premature babies(26-30 weeks)2.congenital heart disease3.chronic lung disease this also includes premature babies having bronchopulmonary dysplasia 4.cystic fibrosis.



- This xray in not specific but can represent the lungs of a baby with viral bronchiolitis.
- Findings are;hyperinflation of the chest, peribronchial inflammatory changes and increased vascular markenings.

Viral bronchiolitis

- Indications for admissions;
    1. Has signs of respiratory distress(nasal flaring,tachycardia,tachypnea,retractions,increased work of breathing)
    2. Cyanotic or 02 sat below 92%
    3. Apnic episodes
    4. High grade fever and you suspect pneumonia
    5. Decreased feeding, admission for IV fluids.
    6. RR>60
- Indications for ICU admissions;
    1. Failure to keep their o2 sat above 92% with oxygen supplementation or recurrent apnea.
    2. Infants with lethargy and exhaustion,shows signs of respiratory failure.(type 2 hypercapnic

- Diagnosis is made clinically so we don't need radiological or laboratory investigations but we can do an electrolyte check and ABGs(if alkalotic means theres compensation for the acidotic hypoxemia by hyperventilation and co2 wash out,if normal theres no compensation and the patient is in impending respiratory failure.)
- Management is mainly supportive
    1. Oxygen supplementation
    2. IV fluids
    3. Chest physiotherapy and hypertonic saline nebulizers to improve airway clearance.
- Palivizumab;preventive measure of bronchiolitis,given for high risk babies;it is IVIGs but very expensive and not stable needs infusion every 3-4weeks.
- The median duration of symptoms is 12 days,half the patients resolve and get back to normal baseline but the other half may remain symptomatic(wheeze and cough) 1 month after the episode.

STRIDOR

- Stridor is a sign of(large)upper airway obstruction.
- ***laryngomalacia*** is the most common cause of chronic stridor.(not to confuse with tracheomalacia which is an inborn abnormality of the tracheal cartilage and presents early in the first week of life)
- ***Croup*** is the most common cause of acute stridor.
- An inspiratory stridor suggests airway obstruction *above the glottis.*
- An expiratory stridor is indicative of obstruction in the lower trachea.(below the glottis)
- A biphasic stridor suggests a glottic or subglottic lesion.

Causes of acute stridor

1. Croup(laryngotracheobronchitis)*the most common cause of acute stridor
2. Epiglottitis
3. Bacterial tracheitis
4. Retropharyngeal abscess

Chronic stridor

1. Laryngomalacia*most common cause of chronic stridor.
2. Tracheomalacia(first week of life)
3. Vocal cord paralysis either uni or bilateral.

Laryngomalacia (chronic stridor)

- Is the *most common* cause of chronic stridor in children younger than two years.
- Male predominance.
- The condition is due to an *intrinsic defect or delayed maturation of supporting structures of the larynx*.
- The airway is partially obstructed during inspiration by the prolapse of the flaccid structures, The inspiratory stridor is usually worse when patient is supine,crying,agitated or when an URTI occurs.
- Summary : Immature development of supporting laryngeal structure thus prolapse of flaccid structures causing partial obstruction during inspiration,worse when the patient is supine,agitated,crying or when an URTI develops,male predominance<2yrs.

Laryngotracheobronchitis (Viral Croup) (acute stridor)

- The **most common** cause of acute stridor in childhood.
- The condition is caused most commonly by **parainfluenza virus**, but it can also be caused by influenza virus types A or B, RSV and rhinoviruses.
- Croup usually occurs in children **6 months to 6 years** of age, with a peak incidence in the second year of life.
- Slight male predominance.
- Is usually preceded by an upper respiratory tract infection of several days' duration.
- Then A low-grade fever, barking cough, inspiratory stridor and hoarseness then develop.
- Symptoms are characteristically worse at night and are aggravated by agitation and crying.

Epiglottitis (acute stridor)

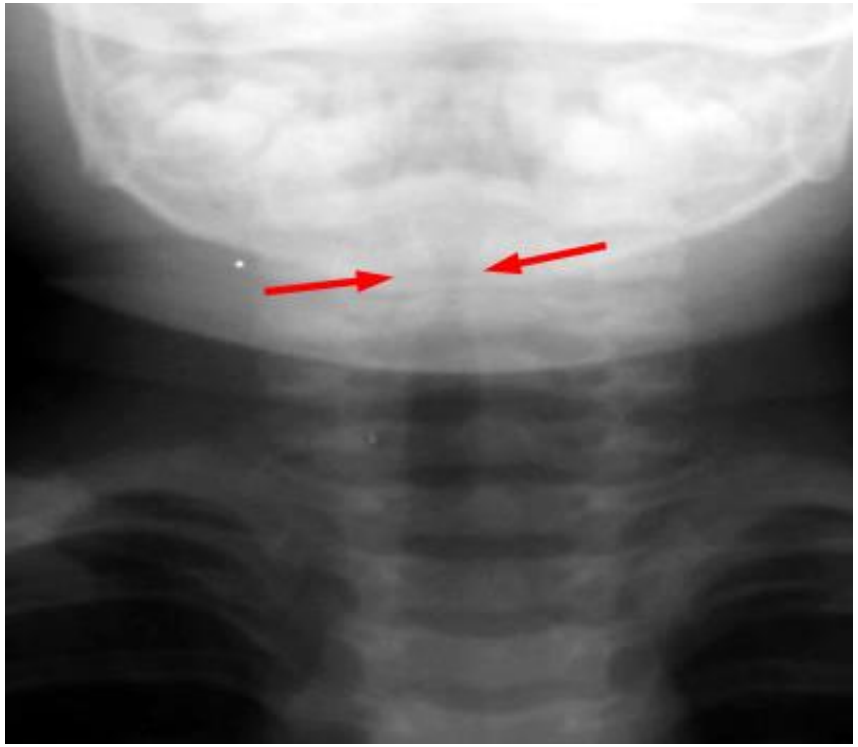
- True Medical Emergency
- In children, epiglottitis is almost always caused by **Haemophilus influenzae type b.**
- In recent years, the occurrence of epiglottitis has been reduced dramatically by the widespread use of the H. influenzae type b vaccine.
- Epiglottitis usually occurs in children *2-7 years* of age, with a peak incidence in three-year-olds.
- Male predominance.
- The disease is characterized by an abrupt onset of high fever, toxicity, agitation, stridor, dyspnea, muffled voice, dysphagia and drooling.

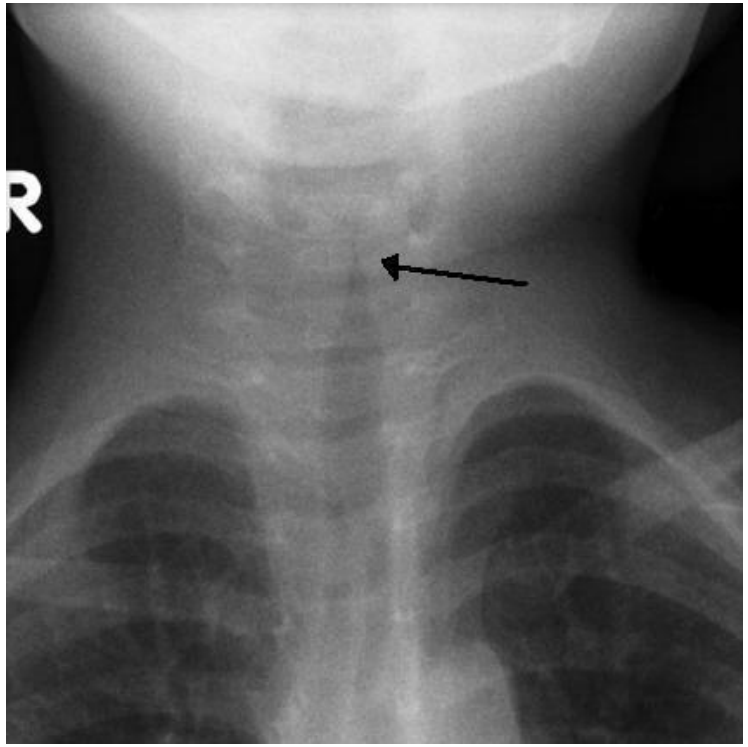
- The older child may prefer to sit leaning forward with the mouth open and the tongue somewhat protruding.
- *An edematous, cherry red epiglottis, visualized in a controlled environment, is the hallmark of epiglottitis.*

Most common causes of stridor	Age	Male;female	Cause	Aggrevating factor/relieving factors	presentation
1.laryngomalacia	less than 2 years(peak 4-6 weeks)	Male predominance	Intrinsic defect that caused immaturation of the supporting structures of the larynx	Worse when the patient is supine,crying or agitated or when an URTI happens	Most common cause of chronic stridor in patients. Inspiratory stridor because of prolapse of flaccid structures
2.CROUP(laryngotracheobronchitis)	6 months to 6 years Peak 2yrs	Male predominance	Commonly para influenza virus,influenza or RV or RSV can cause it.	Worse at night or when the patient is crying.	Starts with an URTI symptoms for several days then low grade fever,barking cough hoarseness and inspiratory acute stridor
3.epiglottitis	2-7 year with peak at 3 years	Male predominance	Hemophilus influenza type B	Patients prefer to sit leaning forward with the mouth open the tongue somehow protruded	Abrupt onset of high grade fever,toxicity,stridor ,dyspnea,dysphagia ,drooling,muffled voice,agitation

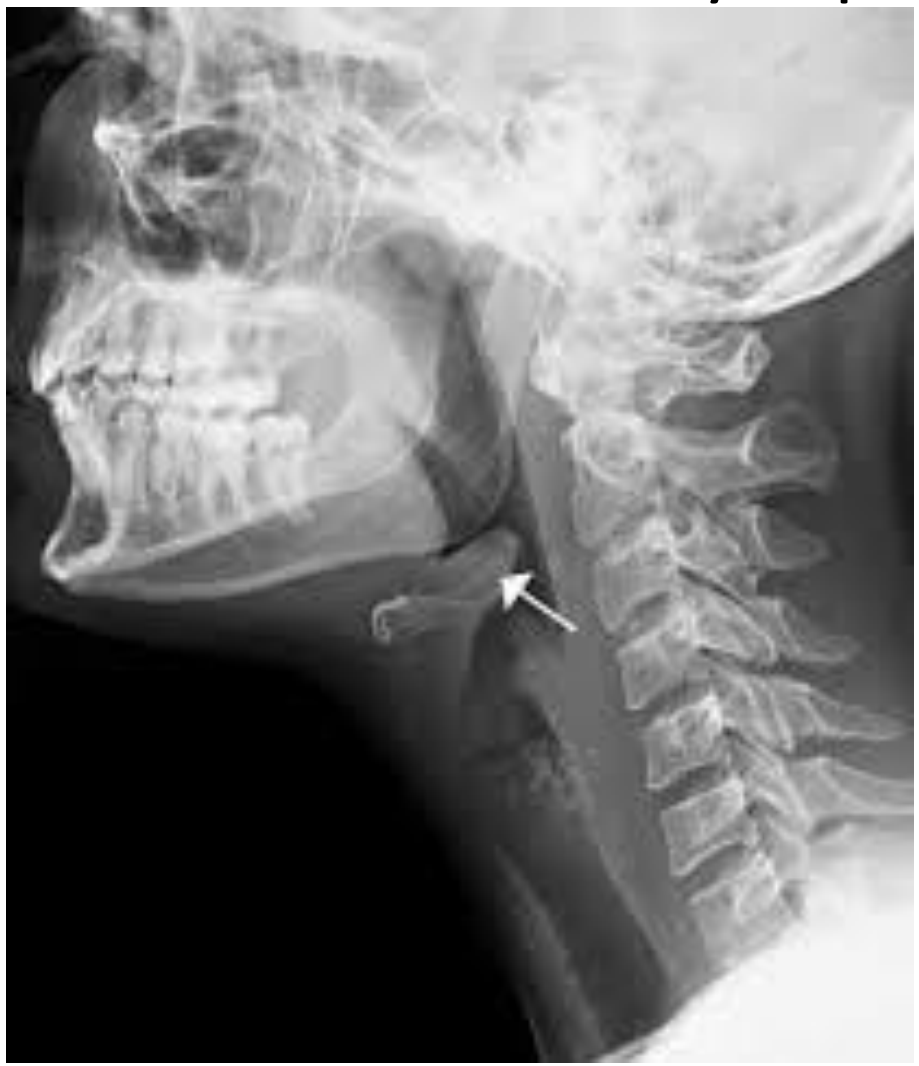
Less common causes of stridor	Cause/age	Presentation/age
1.Vocal cord paralysis(chronic stridor or more commonly hoarseness) -unilateral(more common on the left side because of long course of the recurrent laryngeal nerve) -bilateral	-uni. Usually birth trauma -bilateral usually central cause like hydroceph,Arnold-chiari,cerebral hemorrhage,birth asphyxia or attempts for endotracheal intubation,bulbar injury(congenital or syndromatic)	The stridor is usually biphasic, -in unilateral voice or cry is weak but no respiratory distress. -in bilateral voice is of good quality but there is marked signs of respiratory distress.
2.Tracheomalacia(chronic stridor)	Inadequate cartilaginous support of the trachea.	May not be present at birth but appears gradually during the first weeks of life as a chronic stridor worse in supine position agitation,URTI(just like laryngomalacia)
3.Bacterial tracheitis(acute stridor)	Staph aureus	Usually after an URTI,the patient then becomes toxic,high fever,respiratory distress,commonly in patients younger than 3 years.
4.Retropharyngeal abscess(acute stridor)	Complication of bacterial phayringitis	Sudden onset of fever,dysphagia,refuse feeding,extension of the neck,sore throat,respiratory distress,usually in patients less than 6 years.

Steeple sign(diagnostic of croup)





Thumb Sign (diagnostic of epiglottitis+the hallmark of edematous cherry epiglottis)



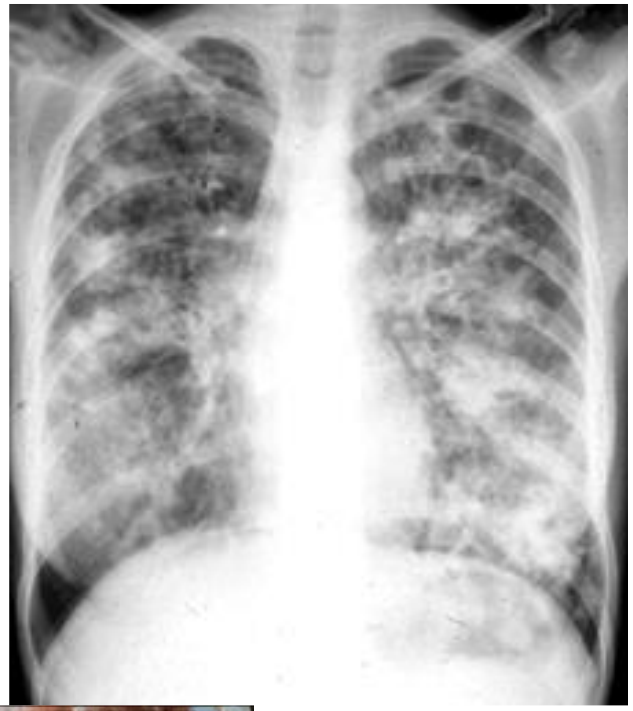
Cystic fibrosis

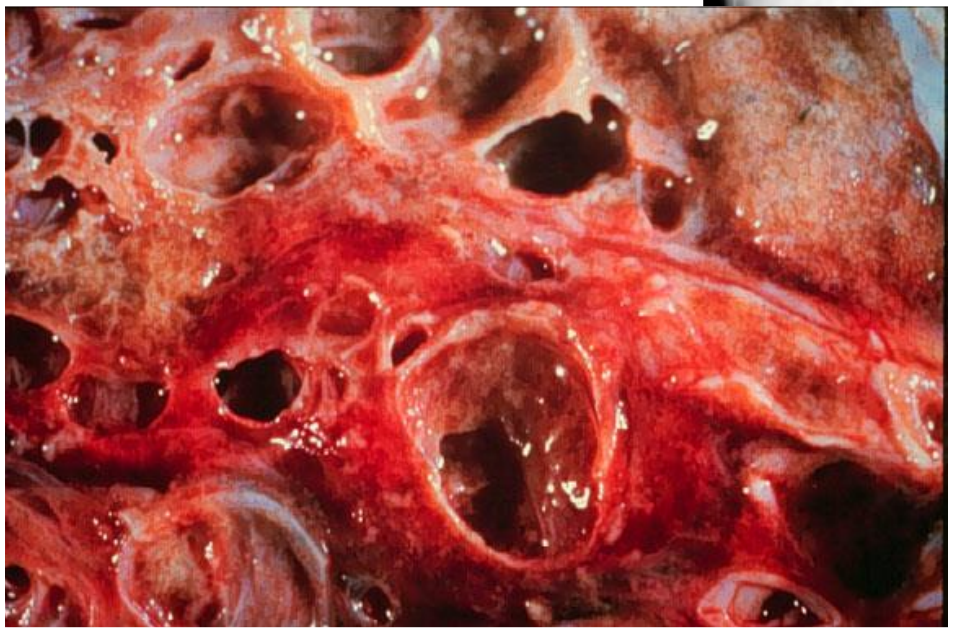
- CF is a life-shortening, multisystem genetic disease.
- Mutated gene is CFTR(cystic fibrosis transmembrane conductance regulator),regulates chloride conductance.
- Characterized by
    1. Sinu-pulmonary disease,sinusitis and nasal polyps,recurrent infections and bronchiectasis (chronic progressive obstructive lung disease-chronic cough)
    2. GI-diseases(meconium ileus,distal intestinal obstruction syndrome,rectal prolapse,GERD,intussusception,FTT)
    3. Pancreatic disease(exocrine insufficiency-enzymatic - leads to fat malabsorption thus KADE vitamin deficiencies&steatorrhea,protein malabsorption and hypoprotenemia and edema,DM-TYPE 2 due to progressive pancreatic damage leads to insulin deficiency and DM2)
    4. Hepatobiliary disease;cholestasis,liver cirrhosis.
    5. Genitourinary infertility and late onset puberty.

- **Pathophysiology is dysregulation of chloride movement and thus water,leads to dehydration of cells and accumulation of thick secretions.(mutation in the transmembrane conductance regulator of chloride)**
- **F508del most common mutation.**
- **class 2 is the most common type.**
- **Common organism colonizing the lungs staph. Aureus/HiB and later in the disease pseudomonas aeruginosa.**

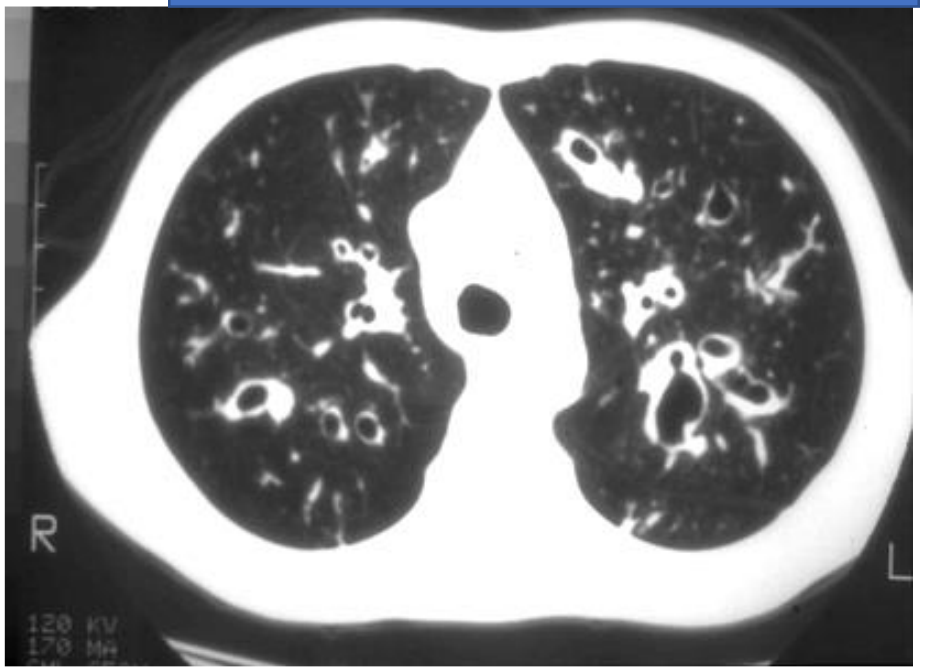
- Diagnosis 1 clinical feature + one laboratory evidence
- Clinical features for the sake of Dx;
    1. Typical clinical features.
    2. One sibling diagnosed with CF
    3. Positive neonatal screening test IRT(immunoreactive trypsinogen)
- Laboratory;
    1. 2 positive sweat chloride tests on 2 separate occasions>60 mmol/L(in children less than 6 months a value of <30 is negative,a value between 30-59 repeat the test until you get a value below 30 or above 60)
    2. Identification of 2 CF mutations.
    3. Nasal potential difference test.

Chest xray findings;bilateral diffuse infiltrates(always think of CF),hyperinflation
CT-findings
Dilatation of the airways signet ring appearance.

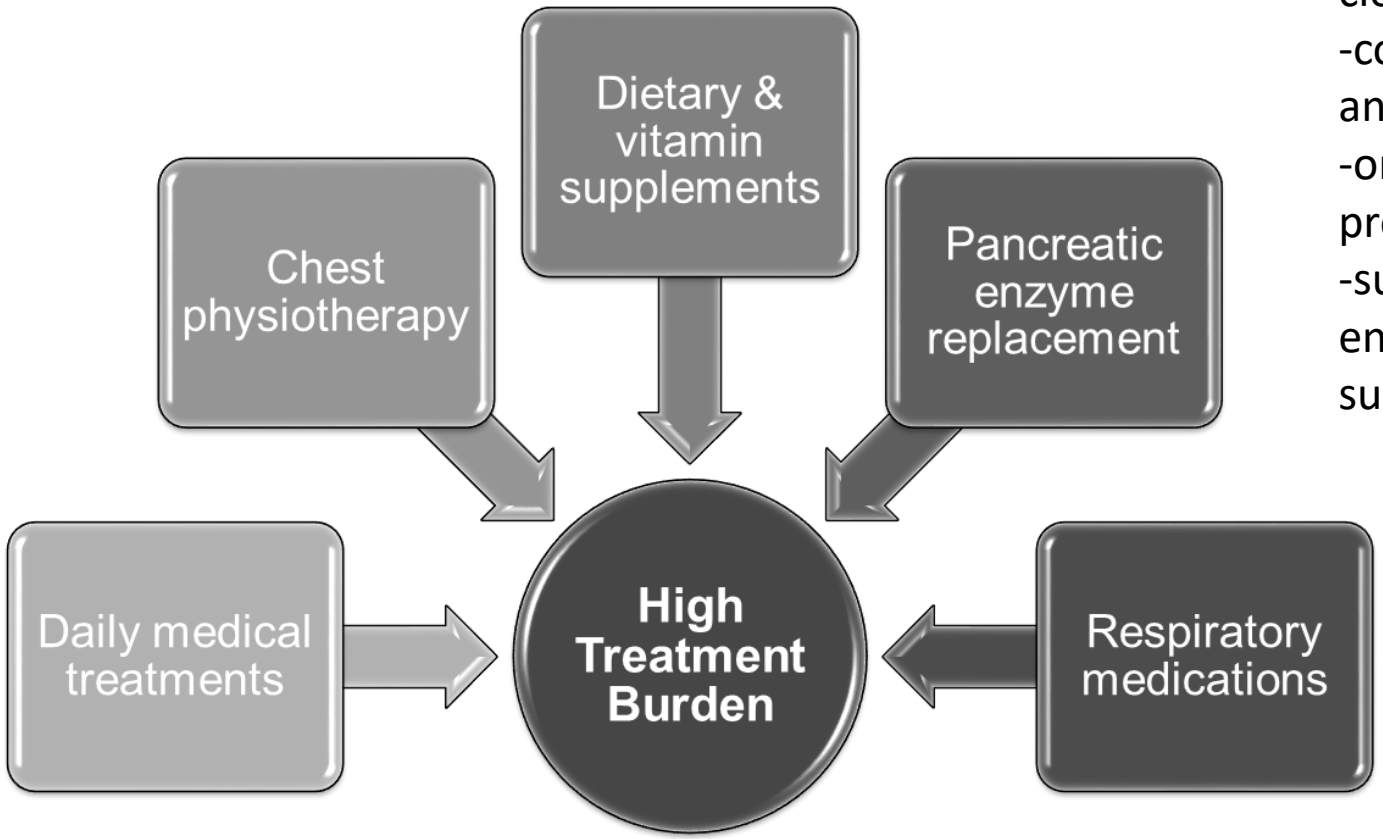




Complications;
Pneumothorax
Hemoptysis
Respiratory failure
Cor pulmonale
Digital clubbing



Complexity of CF Treatment



-Chest physiotherapy and hypertonic saline nebulizers to promote secretions clearance.
-control lung infection by giving antibiotics.
-oral laxatives and good hydration to prevent obstruction.
-support adequate nutrition by giving enzyme replacements and vitamin supplements.

Key questions to ask in a patient with wet cough

- 1.amount and colour (large amount and green colourleads towards bronchiectasis)
- 2.hemoptysis suspecting TB,immunization history,recent travel to an endemic area with TB or sick contact with TB patient
- 3.associated symptoms steatorrhea(loose frequent pale in colour bulky foul smelling stools hard to flush) leads to CF
- 4.gets better with antibiotics leads to CF
- 5.antenatal history,meconium ileus leads toward CF
- 6. poor weight gain or FTT, growth parameters

Differential diagnosis;
Cystic fibrosis
TB
Primary ciliary dyskinesia
Chronis sinusitis
Primary immune defciency

ASTHMA

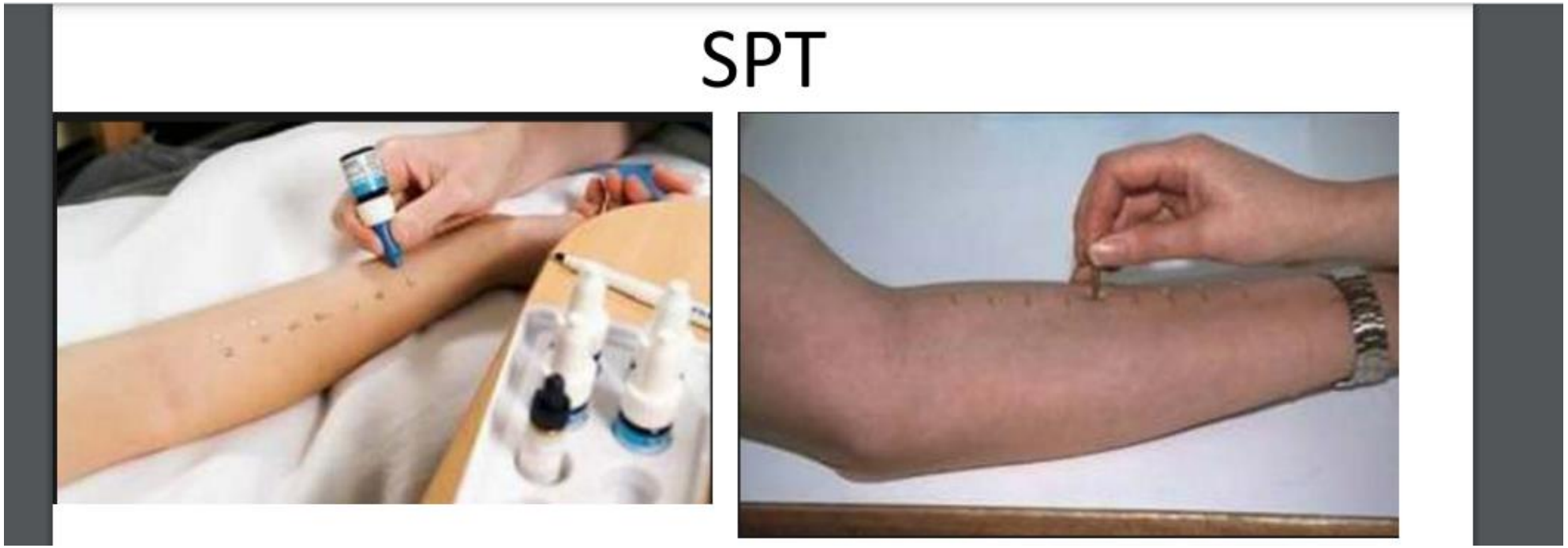
- Chronic inflammatory condition of the airways causing recurrent or persistent bronchospasm(early only contributor to the obstruction,then the inflammatory phase also contributes to the obstruction),bronchial hyperresponsiveness.
- Some patients will have irreversible subepithelial fibrosis that leads to remodelling and smooth muscle hypertrophy only 5%.
- Chronological pathophysiology;
    1. Early phase 15-30 mins after exposure(bronchospasm)
    2. Late phase (4hrs-12hrs)infiltration of inflammatory cells(eosinophils,mast cells and neutrophilic granulocytes) lead to mucosal edema and mucus plug

- Body compensation over time,hyperinflation;at first its beneficial it helps split the obstructed airways however with time it will have negative effect and increase the work of breathing.how?
- Whenever we have overdistention of the alveoli(hyperinflation) theres vasoconstriction causing shunting and hypoxemia,hypoxemia drives hyperventilation(increased work of breathing),at first the body compensates by hyperventilation(co2 wash out)-respiratory alkalosis-,but with time patient respiratory system gets tired and enters into a respiratory failure type 2 (hypercapnic) so whenever theres a patient with respiratory distress and ABGs are normal this is a warning sign.

- Check level of consciousness of the patient,agitated-hypoxia,lethargic-respiratory failure(co2 retention and acidosis or normal ABGs).
- Causes of asthma
    1. Genetic input cytokine imbalance towards t-helper 2, aerosensitization which is an igE mediated response.
    2. Environmental input –viral infection(RSV in infants <1year) they are not direct contributors -inhaled allergen.
- An interaction between two factors leads to asthma,in order to be labelled as asthma patients you need to be >5years, before that we label it as reactive airway disease.
- Most children with recurrent wheezy chest and cough won't have asthma as adults,most adults with asthma have the symptoms since childhood.
- Most children with asthma overgrow their problem as adults only a small percentage continue to be asthmatic as adults.

How to document aerosensitization? SKIN PRICK TEST

- Why it is important? for preventive measures to avoid the allergen,for prognostic factors usually atopic children continue to have asthma as adults,also for immunotherapy.
- Allergen vs irritant; allergen is an organic material, genetic predisposition to igE mediated response most common is the house dust mite, irritant is a non-organic material that generates a hyperresponsive response without a genetic predisposition,example smoking.



Clinical phenotypes of asthma

- 1.asthma in children <5years aka recurrent viral bronchiolitis; recurrent wheezy chest and cough they usually overgrow their problem as adults,labelled as reactive airway disease.
- 2.chronic or persistent asthma usually associated with atopy. Above the age of 5 years
- Other uncommon types of asthma(occupational asthma,female asthma in obese prepubertal females, samters triad of asthma-aspirin sensitivity&nasal polyps and asthma)
- Before diagnosing samter triad in nasal polyps and asthma rule out CF and primary ciliary dyskinesia, primary immune deficiency.

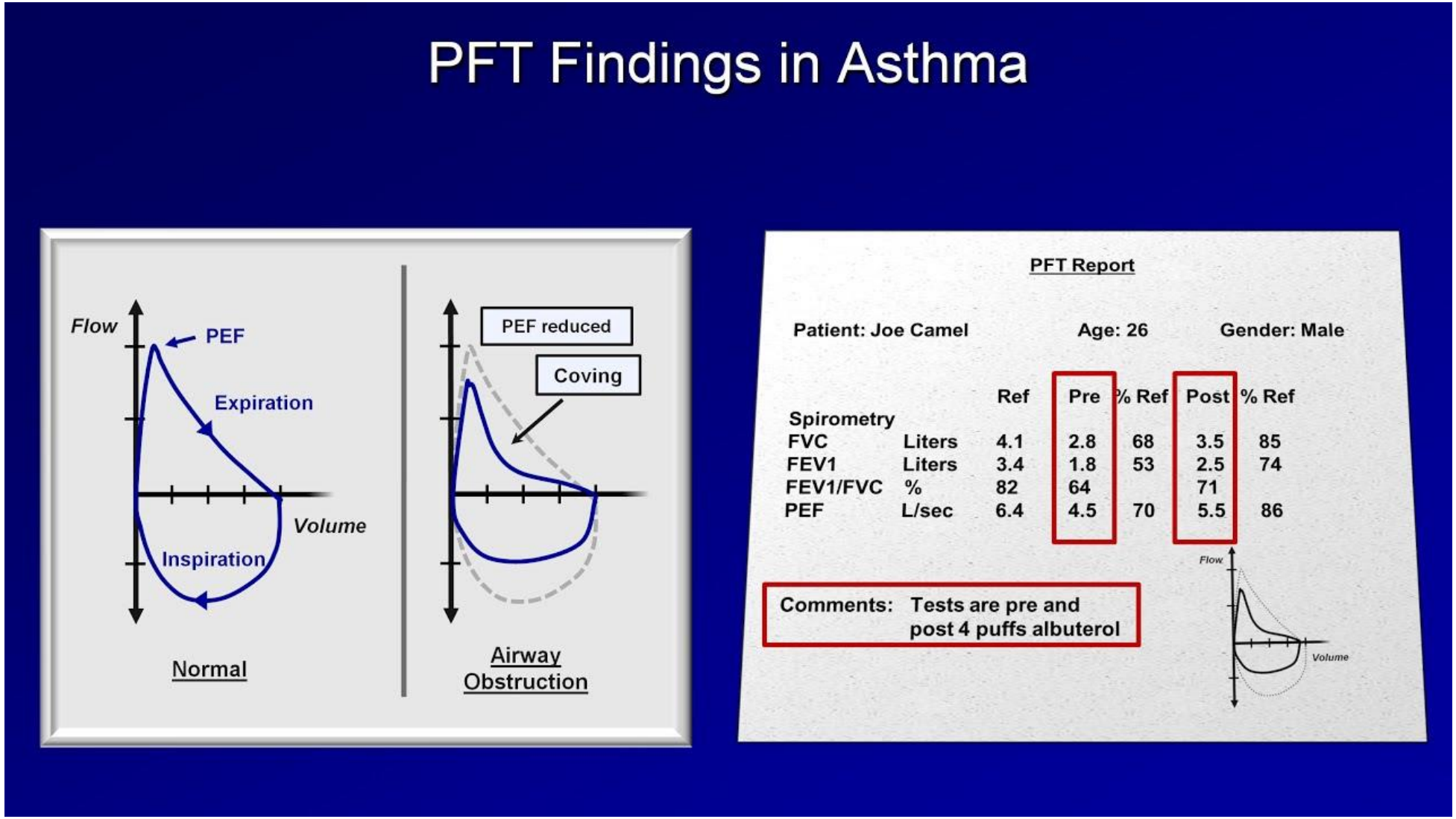
Clinical presentation

- Wheezy chest
- Cough in asthma(dry,gradual onset,worse at night and after exercise,related to certain exposures)-don’t ask them in the miniosce not key Qs-
- Factors supporting asthma Dx;food allergy,atopies,parents history-the maternal history is stronger-.
- What are other atopies?-atopic dermatitis-allergic rhinitis-allergic conjunctivitis-food allergy.
- How to document severity of asthma? frequency of episodes,night awakenings,post-tussive vomiting,cyanosis,use of risky medications SABA,previous hospitalization because of it.

Physical exam findings&investigations

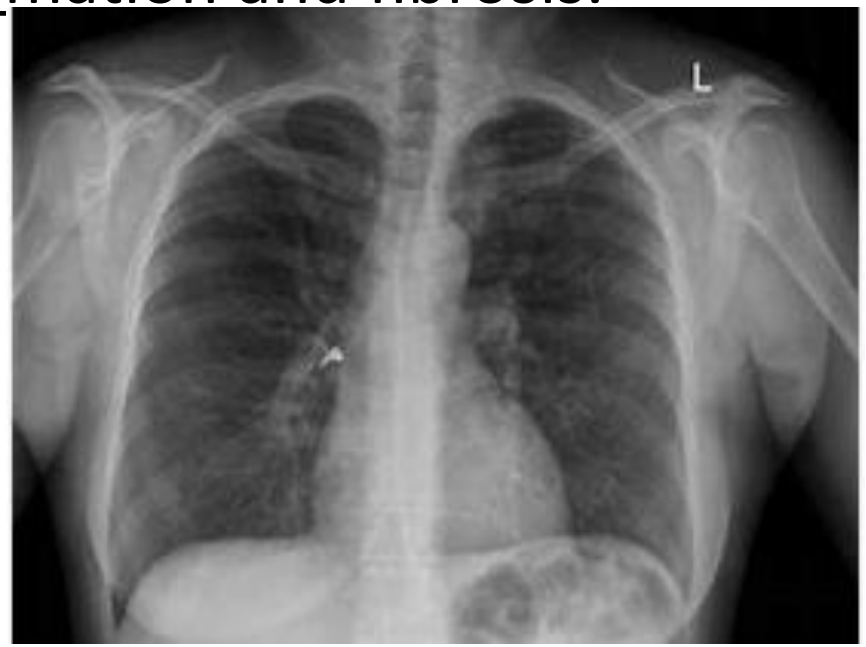
- General exam;vital signs check for signs of distress and level of consiouseness, usually irritable and agitated except when in impending RF they would be lethargic.
- End-expiratory wheezing or a prolonged expiratory phase /most commonly, although inspiratory wheezing can be heard.
- Diminished breath sounds and chest hyperinflation may be observed during acute exacerbations.
- They can also present with silent chest and become wheezy after treatment.
- Investigations/asthma biomarkes
    1. Spt for documentation of aerosensitization
    2. Total serum IgE
    3. Serum eosinophils (eosinophilia>4%)
    4. Spirometry(not important for diagnosis but to assess and monitor the disease progress)
    5. Fraction of Exhaled Nitric Oxide Testing.(FENO),high level would indicate inflammation of airways.
    6. Radiology
    7. Urinary leukotrienes
- And remember asthma is a clinical diagnosis you do investigations for preventive measures of allergens to assess and monitor the disease to rule out other diseases.

- Findings; Airflow limitation (low FEV 1 ,FEV 1/FVC < 0.8)
- Bronchodilator response : improvement in FEV1> = 12%
- Exercise challenge : worsening in FEV1 >=15%



Radiology findings

- Xray findings early in the disease;peribronchial thickening and hyperinflation(seen as flattening of both hemidiaphragms both are parallel,increased lucency of lung fields),also found in viral bronchiolitis.
- Later in the disease you can find severe hyperinflation/diffuse bronchiectatic changes,peribronchial inflammation and fibrosis.



- How to document severity and control of asthma?frequency of episodes,night awakenings,use of risky medications SABA, exacerbations requiring oral steroids,interference with daily activity.
- Refer High risk patients to specialist : • History of sudden severe exacerbations • History of prior intubation for asthma • Admission to an ICU because of asthma • Two or more hospitalizations for asthma in the past year.

ASTHMA MANAGEMENT

- Management of acute exacerbation of asthma in the ER/relief medications
  1. ABCS, securing patent airways and o2 supplementation.
  2. Short acting beta2-agonist (SABA)-Ventolin(albuterol).
  3. Ipratropium bromide(short acting muscarinic receptor antagonist SAMA),used as sandwich therapy Ventolin-ipratropium-Ventolin.-you can repeat this up to 3 times/hr, every nebulizer nearly needs 20 mins.
  4. Short course of systemic steroids.
  5. If patient is still not responding well we can try one of these in order, IM epinephrine, IV terbutaline, Magnesium sulfate, aminophylline(bronchodilator),non-invasive respiratory support(biPAP),last resort is intubation then ECHMO.
- Extra notes on relief medications:
  - Side effects of Ventolin is hypokalemia and hypophosphatemia, can be used in cases of hyperkalemia.
  - ECG progression of a pt with hypokalemia,prolonged t wave,biphasic t wave and then flattened t wave.
  - aminophylline has a very low therapeutic index.
  - Also note that if the patient is already on a controller you can keep it in your plan while he is acutely managed for the exacerbation.
  - In the hospital we prefer to give the patient the inhaled medication by a nebulizer, why? usually we give it with oxygen, provides more humidity since the patient will already be having dry airways, better delivery to the terminal alveoli.

Asthma control management

- Step 1: very low dose ICS.1*2
- Step 2: low dose ICS.1*2
- Step 3: medium dose ICS.1*2
- Step 4: we add another controller choices are LABA(long acting beta-2 agonist)-salmetErol i.e- or montelukast which is an LTRA(leukotriene receptor antagonist); but this one per se is not preferred in pediatrics because it was reported to have associated psychiatric side effects and nightmares.
- Step 5: high dose ICS +/- oral steroids, or LAMA or biological therapy (OMALIZUMAB) which is an anti-igE monoclonal antibody.

Technique :
MDI + Spacer (metered dose inhaler+valve holding chamber)

DPI(dry powder inhaler)

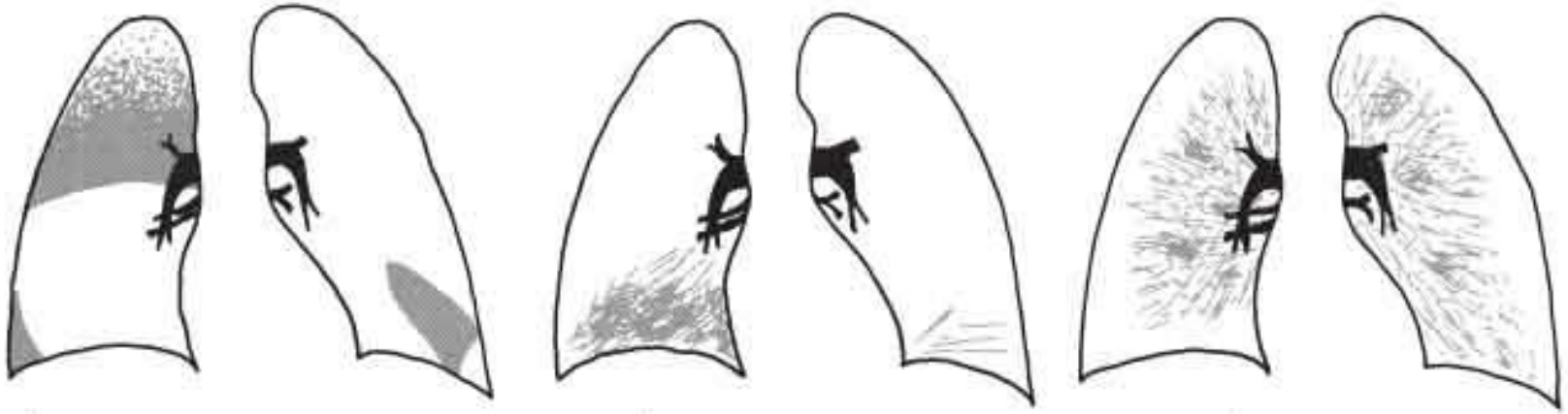
Requires effort

Key questions to ask in history of asthma

- Family history of asthma
- Personal history of atopies(allergic rhinitis or conjunctivitis,food allergy and atopic dermatitis).
- Interval free symptoms.
- Gets better with Ventolin.

Pneumonia

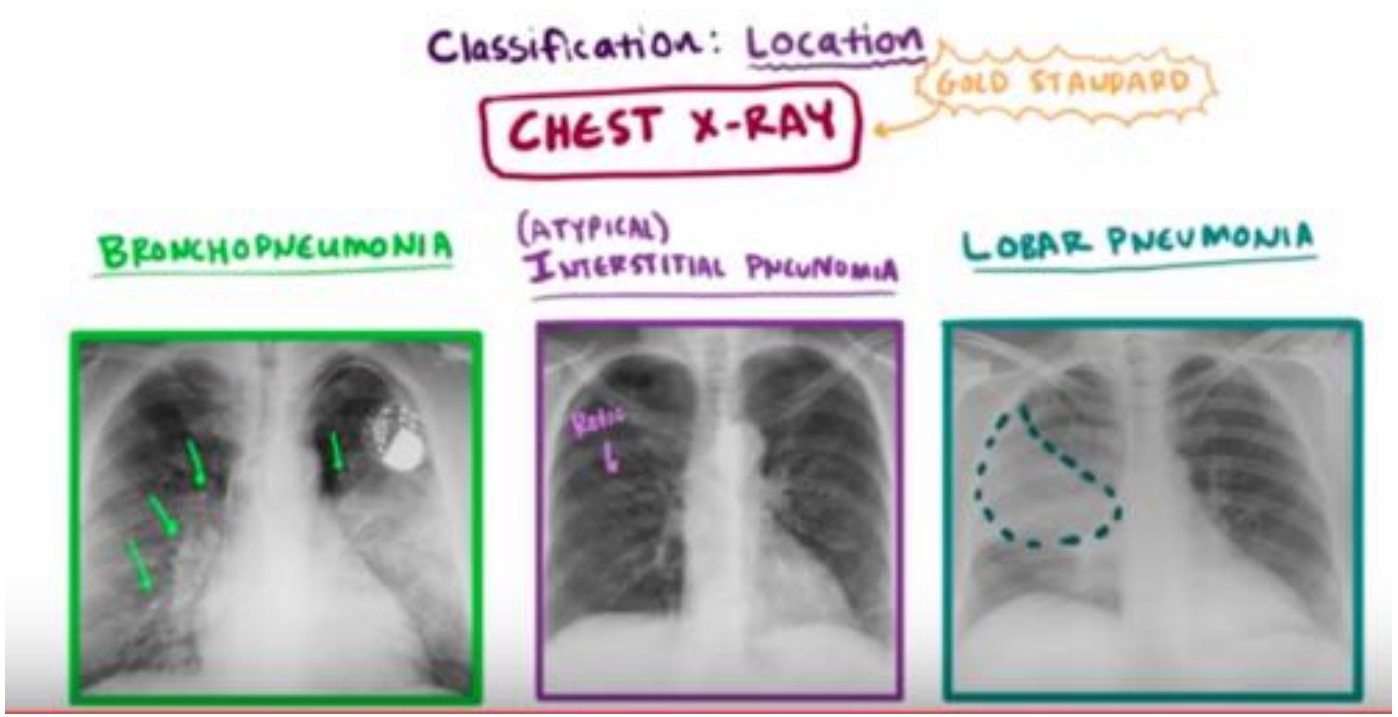
- Pneumonia is classified according to either anatomy or etiology.
- According to anatomy:
  1. Lobar pneumonia/segmental consolidation: lung opacity in the distribution of one lobe with air bronchogram and accompanied with pleural effusion.no volume loss; bronchi patent.
  2. Bronchopneumonia: multifocal patchy opacities in a a basal distribution, no air bronchogram, volume loss because bronchi are filled with fluids.consider mycoplasma in pts >5yrs.
  3. Interstitial pneumonia: reticular pattern of opacity mainly in a central distribution.
  4. Special types of pneumonia: atypical pneumonia with mycoplasma, pneumatocele(cavitatory lesions), round pneumonia.
- According to etiology: its either viral or bacterial.



Lobar pneumonia:
Lobar and/or segmental consolidation with air bronchogram and accompanying pleural effusion

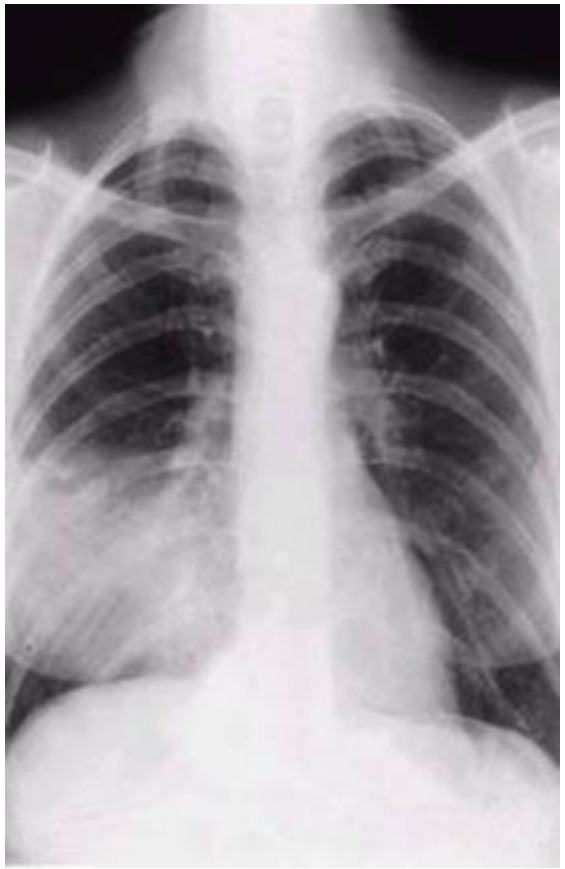
Bronchopneumonia:
coalescing areas of consolidation in a predominantly basal distribution

Interstitial pneumonia:
reticular pattern in a predominantly central distribution



	Lobar/segmental	bronchopneumonia	Atypical/interstitial pneumonia
Most common organism	Strep.pneumoniae	Staph.aureus	mycoplasma
Distribution of lung infiltrates/opacities	Anatomy of the lobe	Multifocal patchy opacities in a basal distribution	Reticular pattern in a central distribution(peribronchial, perivascular)
	Strep also is the most common cause of round pneumonia	Staph also is the most common cause of pneumatocele	

Lobar Pneumonia AKA segmental consolidation



- Mainly infection and inflammation of the alveoli
- Radiology: opacity that involves the anatomy of one lobe, with air bronchogram(fluid filling the alveoli) accompanied with pleural effusion

Pathogens

S. pneumoniae

Others

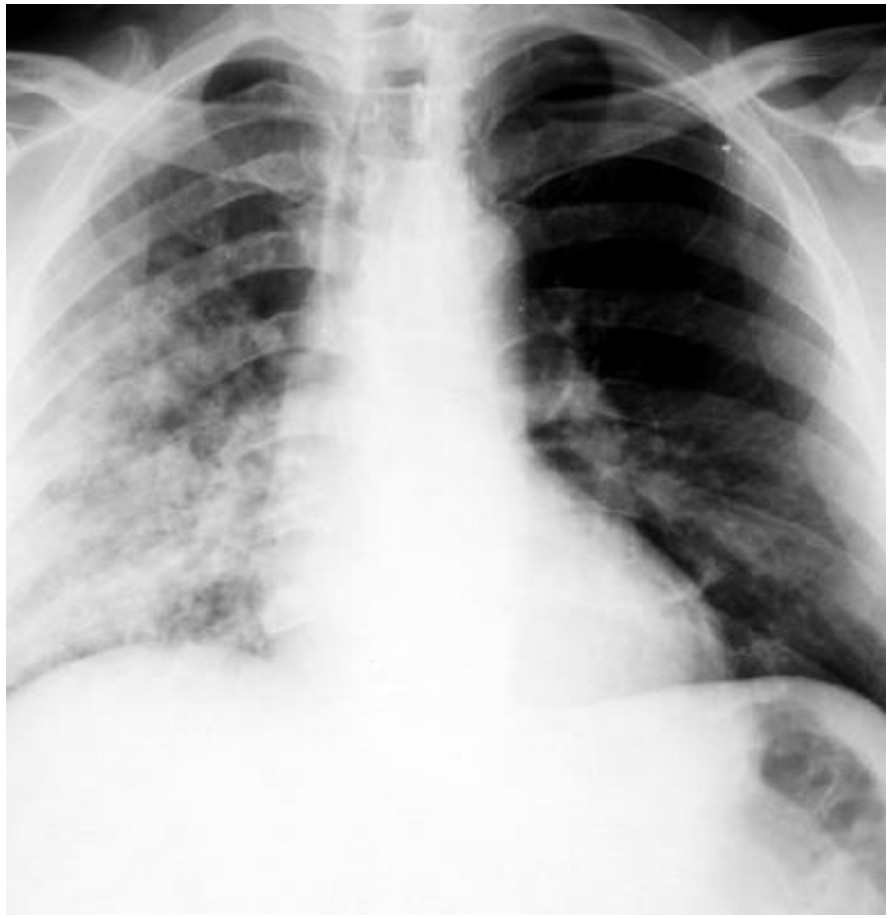
S. aureus

H. influenzae

Fungal

- Bronchi not primarily affected and remain air filled ->; generally no volume loss
- Less commonly seen due to early treatment
- DDx: Aspiration and Pulmonary Embolus

Bronchopneumonia



- Coalscing areas of consolidation in a predominantly basal distribution.
- Primarily affects bronchi and adjacent alveoli -> multifocal patchy opacities.

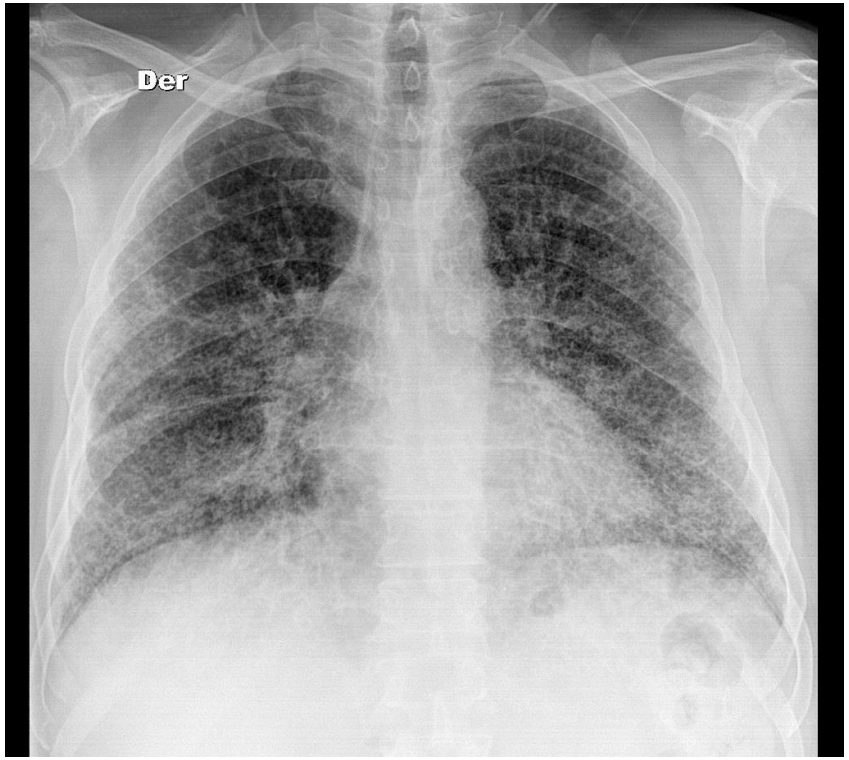
- Volume loss may be present as bronchi filled with exudate.

Pathogens

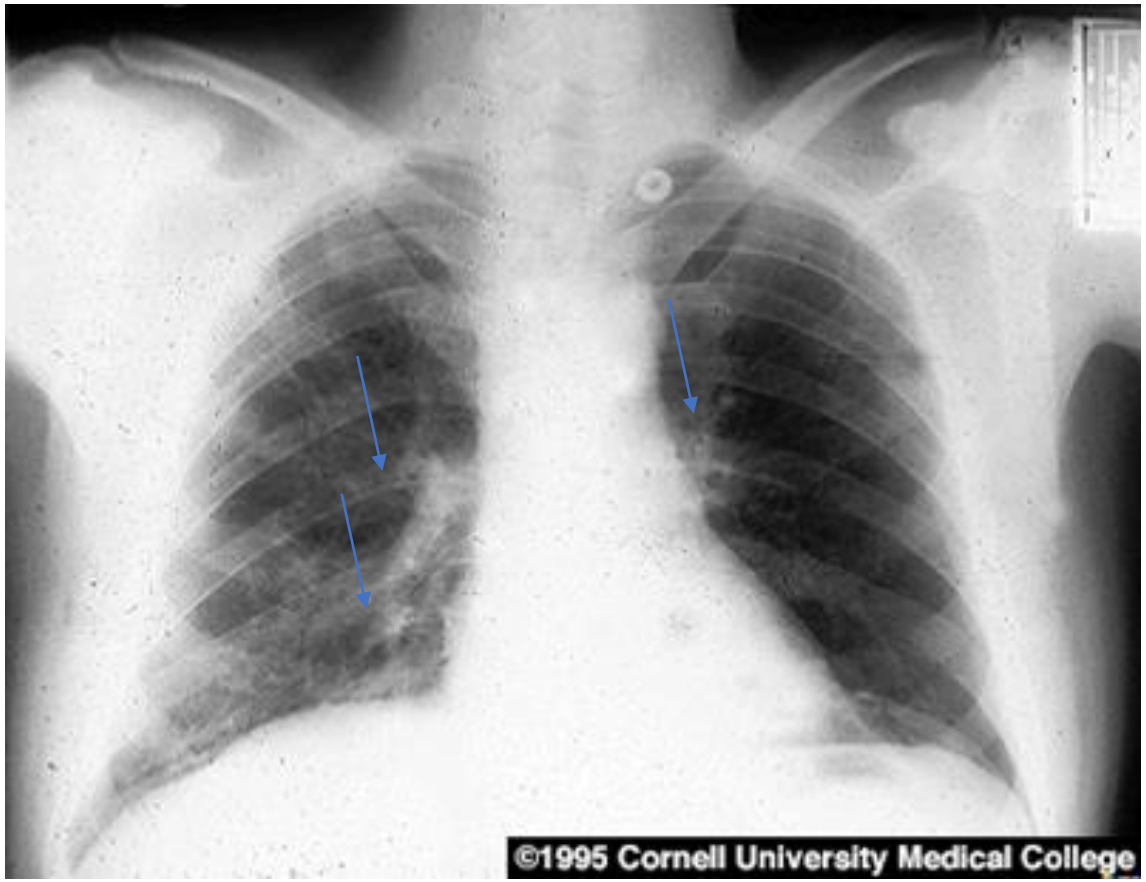
S. aureus
S. pneumoniae
Gram negatives
Mycoplasma>5years of age

Interstitial pneumonia

- Mainly involves the interstitium.
- Reticular pattern in a predominantly central distribution.



Mycoplasma



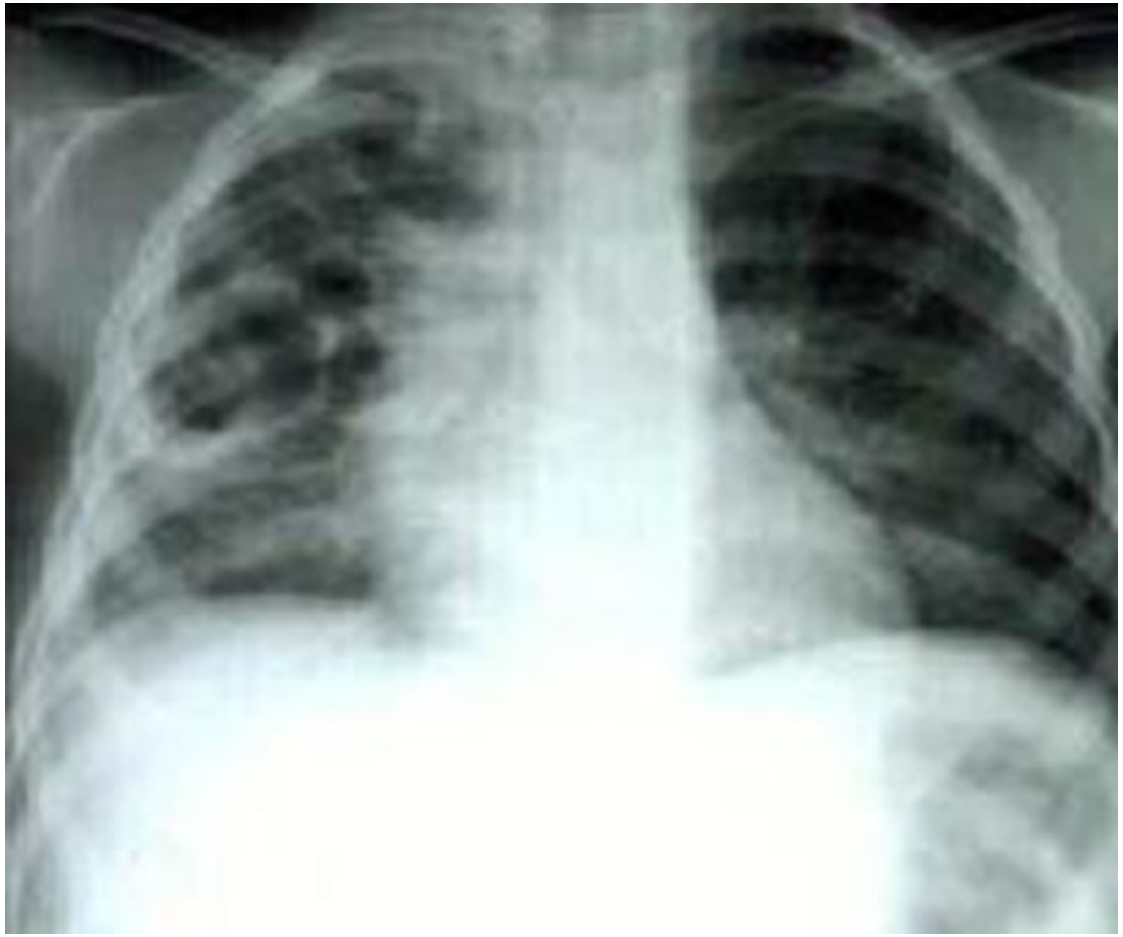
- Most common cause of interstitial pneumonia.
- Among the most common lower respiratory infections worldwide.

Ages 5-20 yrs

Gradual onset of headache, malaise, fever, sore throat, and cough

Bilateral peribronchial perivascular interstitial infiltrates in central and middle lung zones.

Cavitary lesions- pneumatocele



Can occur in 50% of Children

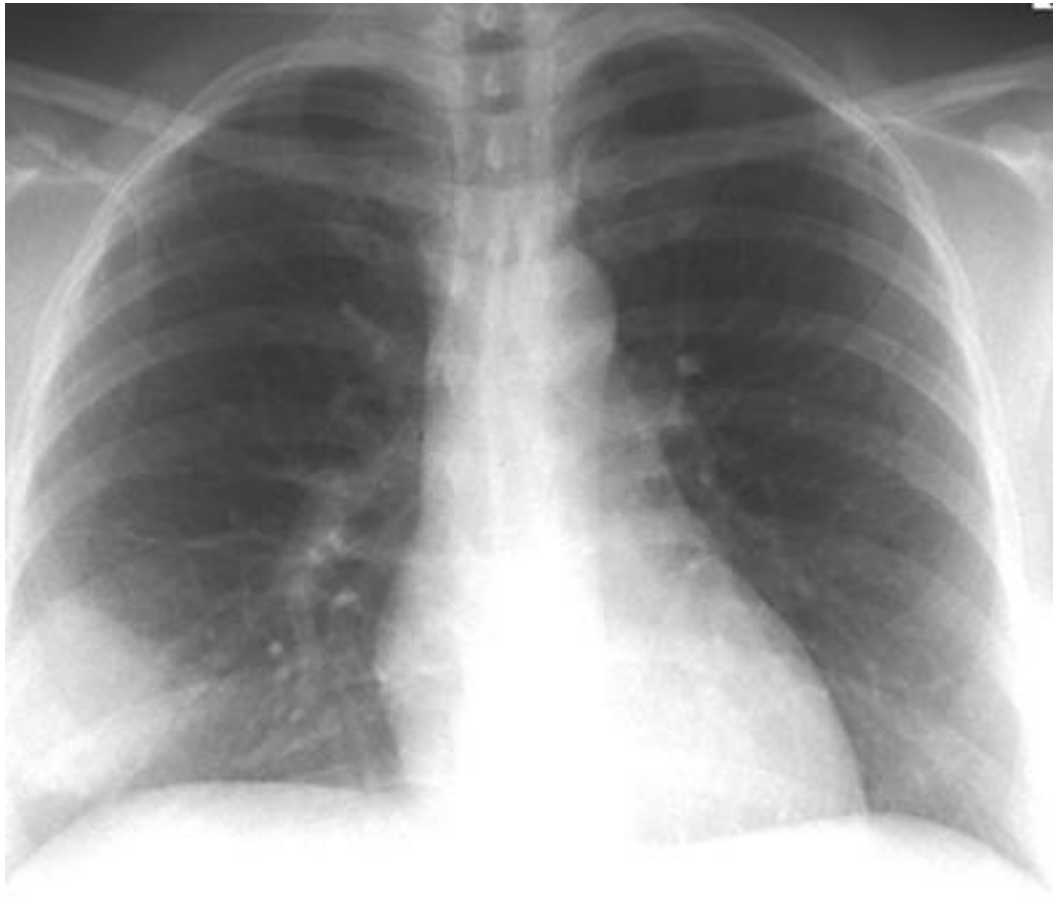
Pathogens

S. Aureus most common

S. pneumoniae

H. influenzae

Gram negative
TB & fungal



Round Pneumonia

S. pneumoniae
Klebsiella
Any pneumonia in children
Atypical Measles

Bacterial:

- *1st 2 months of life*: the common agents include: *klebsiella, E. Coli and staphylococci.(vaginal flora),GBS.*
- *>3months*: *S pneumonia, H influenza and staphylococci*.

Viral: RSV, Influenza, parainfluanza or adenovirus

- Clinical features; **Onset:** May be insidious starting with URTI or may be acute with high fever, dyspnea and respiratory distress.
- Can present with acute abdominal pain, referred from the pleura.
- **P/E:** signs of respiratory distress,signs of consolidation;bronchial breathing,Increased tactile vocal fremitus , dull percussion note.

Diagnosis:

- **CXR:** confirms the diagnosis and may indicate a complication such as pleural effusion or empyema.
- **Viral pneumonia:** hyperinflation with bilateral interstitial infiltrates and peribronchial cuffing.
- **Pneumococcal pneumonia:** confluent lobar consolidation.
- **Staphyloccoci:** Cause pneumatocele as well.

Bloods:

- **Peripheral WBC count**: differentiates viral from bacterial.
- *Viral pneumonia*: WBC count can be NL or elevated but not higher than 20.000 with lymphocytosis.

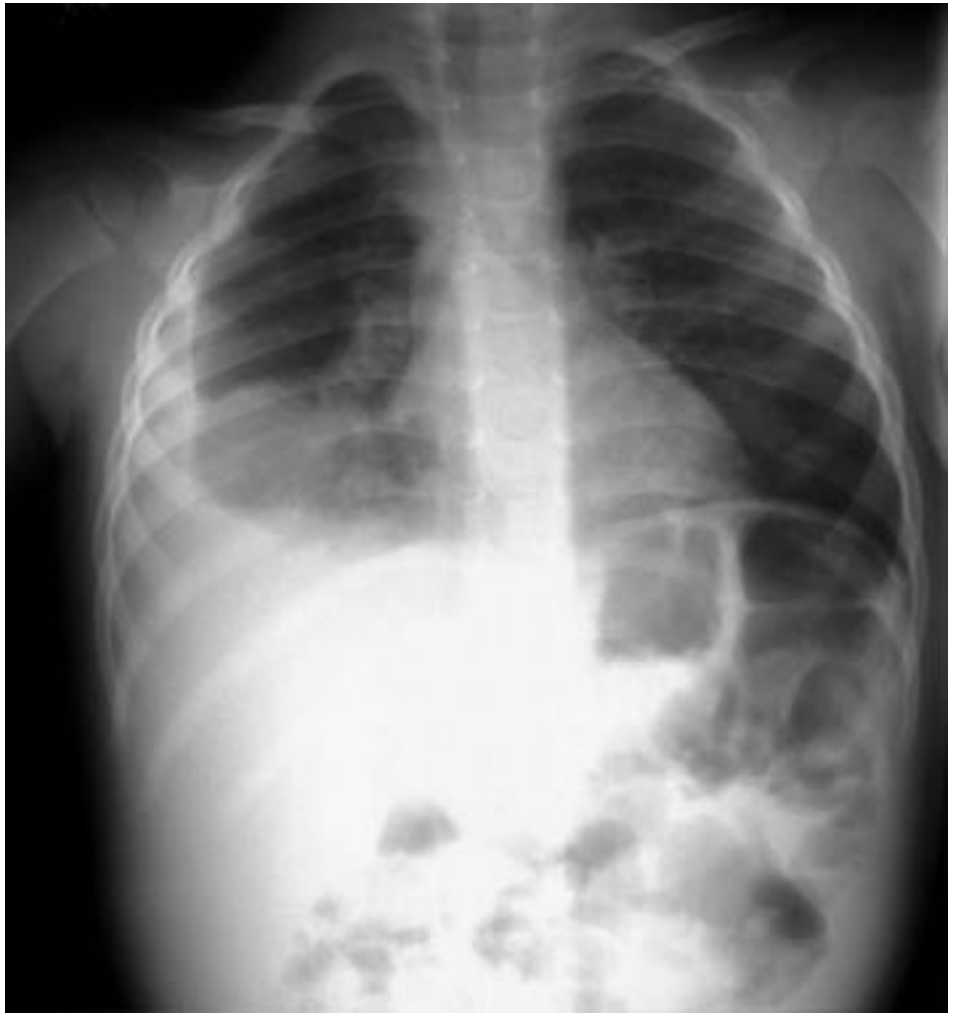
- *Bacterial pneumonia*: is often associated with an elevated WBC count 15,000 – 40,000 with neutrophilia.
- **Blood CX:** should not be routinely performed in nontoxic, fully immunized children, used to rule out sepsis.
- should be obtained in children who fail to demonstrate clinical improvement and in those who have progressive symptoms or clinical deterioration after initiation of antibiotic therapy.
- **CRP and ESR(elevation).**

Complications Associated With Pneumonia

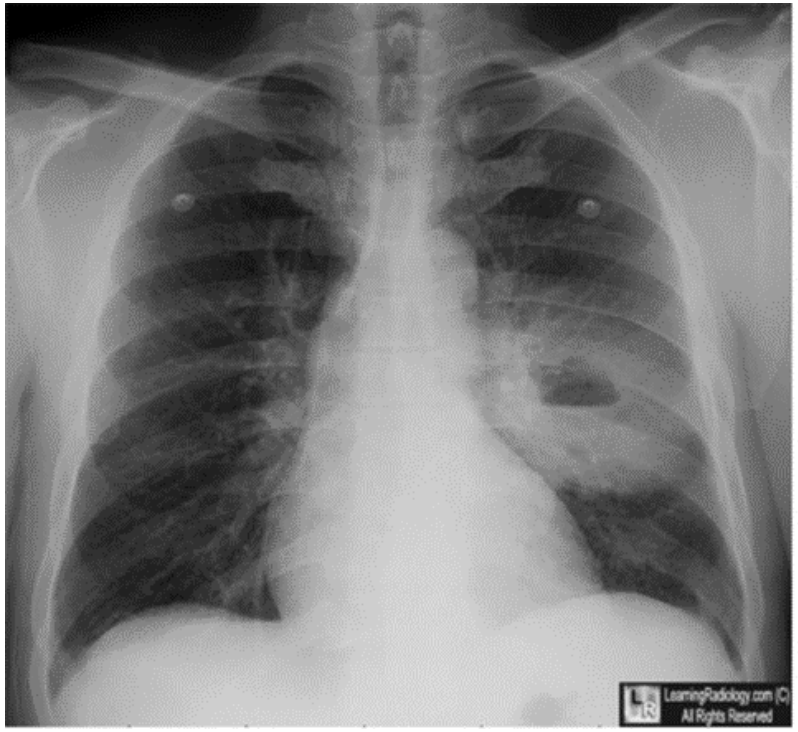
Pulmonary

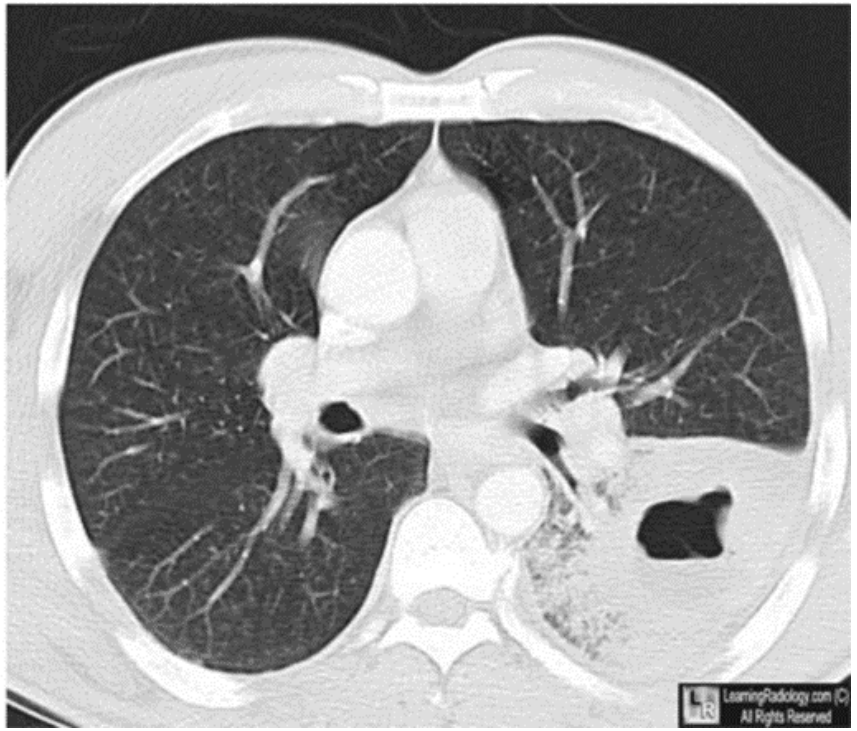
- Pleural effusion or empyema
- Pneumothorax
- Pneumatocele: often resolve spontaneously or may lead to pneumothorax.
- Lung abscess
- Bronchopleural fistula.
- Necrotizing pneumonia.
- Acute respiratory failure

Pleural effusion



Lung abscess





In-Patient management

- Adequate Hydration
- Oxygenation
- Antipyretics and pain control
- Monitoring of :
  - . RR
  - . WOB
  - . Temperature
  - . HR
  - . Oxygen saturation
- Antiobiotics;ideally penicillin G(given IV or IM) or ampicillin(IV OR IM),or ceftriaxone(3rd generation cephalosporin),azithromycin if patient is old enough>5 years to suspect mycoplasma,vancomycin if you suspect MRSA(IV)

Recurrent Pneumonia

- **Two episodes of pneumonia within the same year or 3 or more episodes over any period of time but with complete resolution of clinical and radiological findings between acute episodes.**
- **Causes;**
  1. **FB aspiration(history of chocking)**
  2. **Swallowing abnormalities(feeding &weight gain,swallowing symptoms)**
  3. **Tracheo-esophageal fistula**
  4. **Tracheomalacia**

  Ask about the timing of the symptoms in relation to feeding and the change in position.

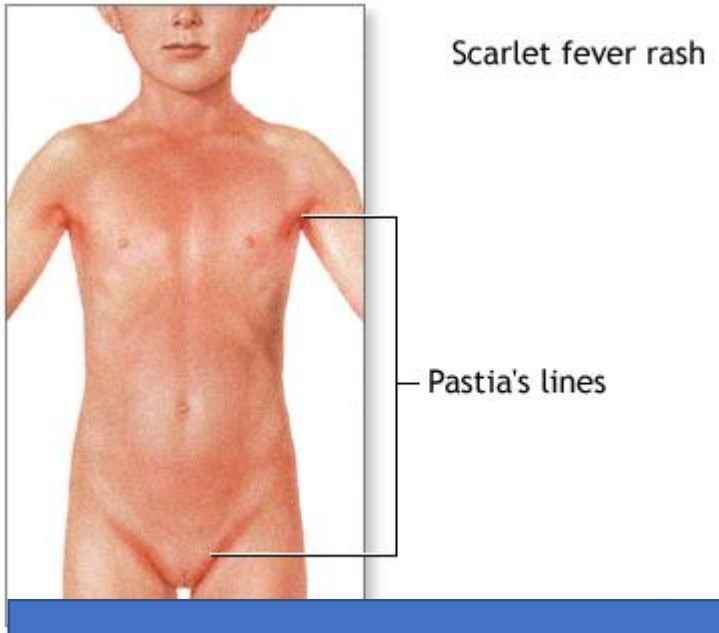
  1. **Immunosuppressive therapy or primary immune-deficiency.(other signs of infection in other symptoms,previous hospitalizations)**

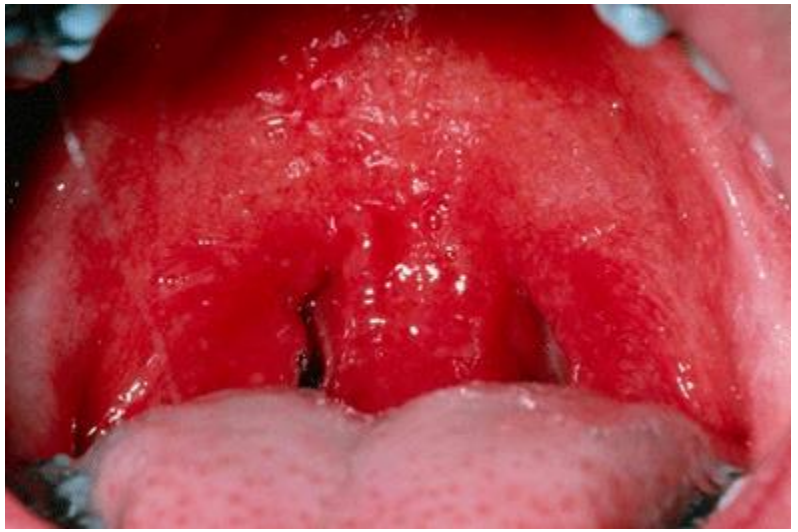
URTI

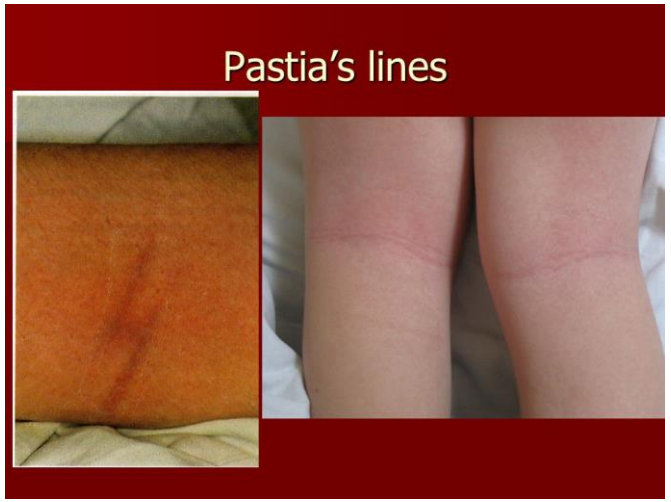
Pharyngitis

VIRAL	B hemolytic group A strep./streptococcus pharyngitis
*more gradual onset of symptoms of URTI(rhinorrhea ,cough ,diarrhea),sore throat with erythematous pharynx but no puss. Variants: 1.adenovirus;pharyngoconjunctival fever. 2.Coxsackie virus may present as *harpangina(greyish vesicles or punched out ulcers on the posterior pharynx) *acute lymphonodular pharyngitis. * hand-foot-mouth disease.(see infectious part) 3.EBV presents as infectious mononucleosis (kissing tonsils and exudative,cervical lymphadenopathy,hepatosplenomegaly,fatigue, MACULOPAPULAR SMALL PINKISH TANNED TO BROWNISH IN COLOUR RASH,the rash can be a bit raised appears following treatment with amoxicillin.dx by heterophile antibody test,atypical lymphocytes seen in peripheral smear. 4. PRIMARY HERPES SIMPLEX ,young children ,high fever ,gingivostomatitis.	*abrupt onset of URTI,high grade fever,SORE THROAT WITH ENLARGED TONSILLS with yellow,**blood tinged exudate,petechiae** on the palate and posterior pharynx,red swollen uvula and **cervical LAP.** ***uncommon below 2-3 years of age**. *scarlet fever:Scarlet fever ,GABHS produce Strep.pyogenies.exotoxin. FINDINGS;circumoral pallor,pharyngitis,strawberry tongue ,fine red papular rash ‘sand paper’,Thompson’s lines or pastia’s lines.

Scarlet fever

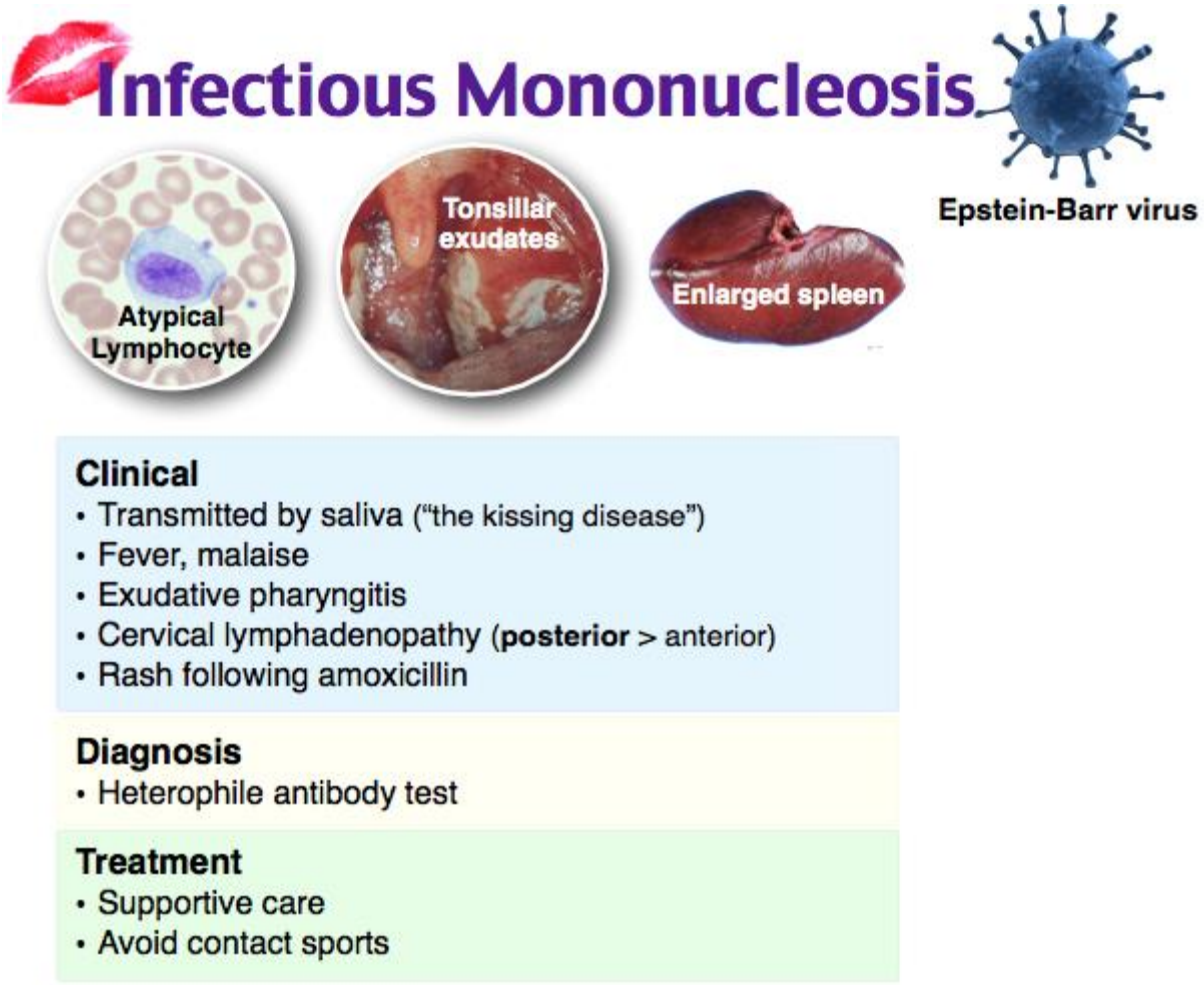


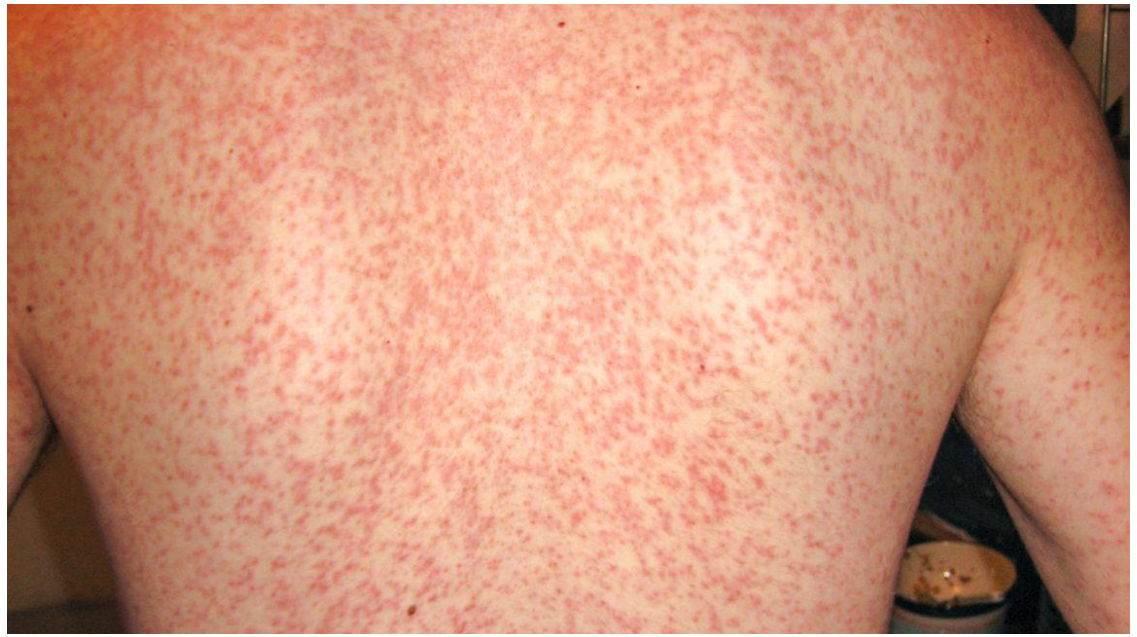


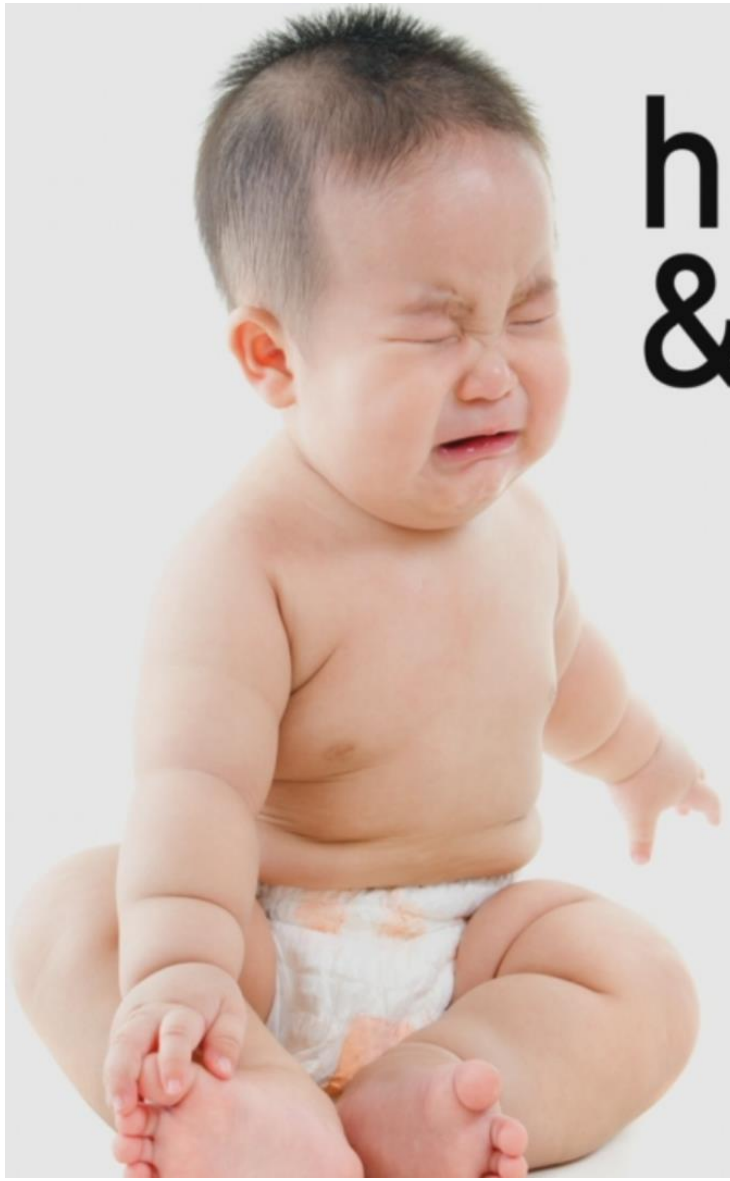


*Pastia's lines or Thompson's lines.
*Scarlet fever rash fine popular erythematous rash(sandpaper rash)
*red pharynx
*stawrberry tongue

INFECTIOUS MONONUCLEOSIS







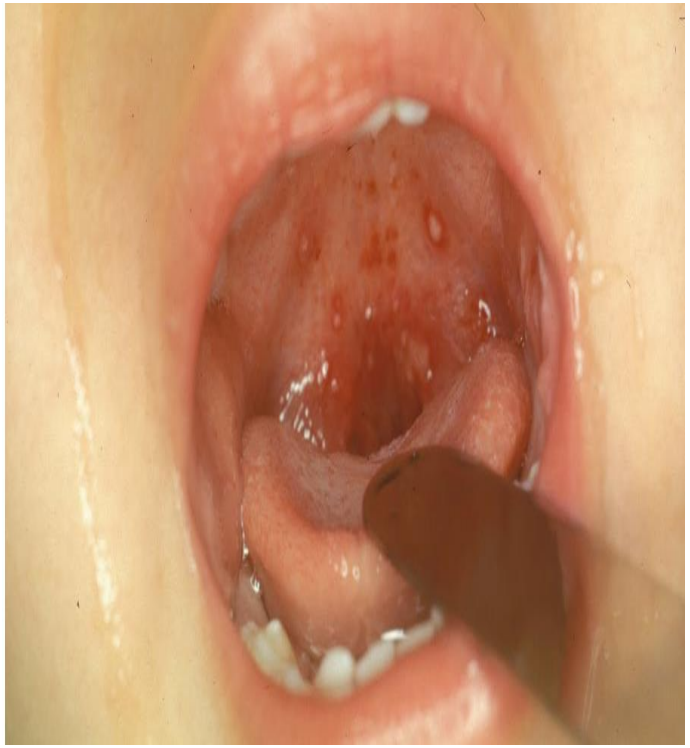
hand, foot & mouth disease

source: American Academy of Pediatrics

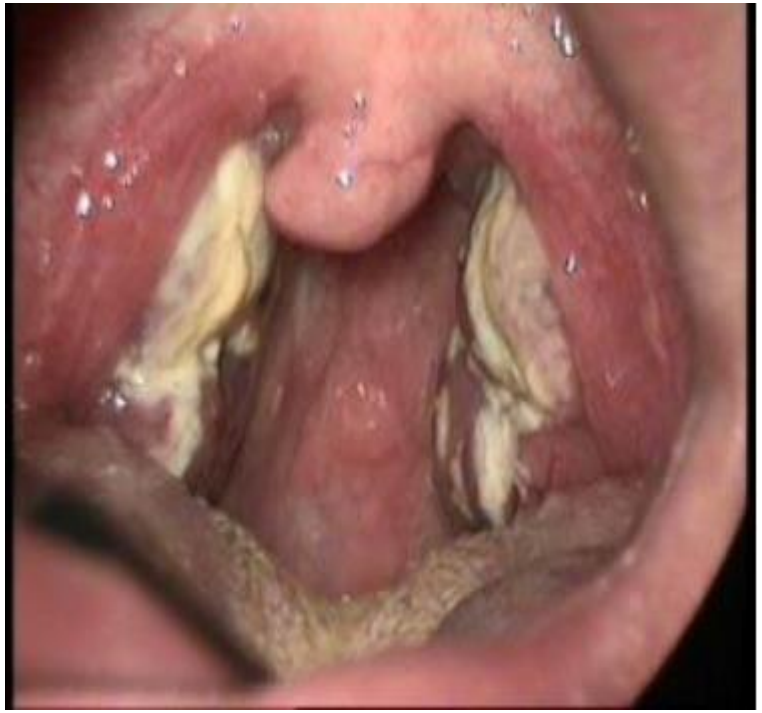
SYMPTOMS

fever
sore throat
sores on tongue, gums or inside the cheek
red rash or blisters
Irritability in infants & toddlers
loss of appetite

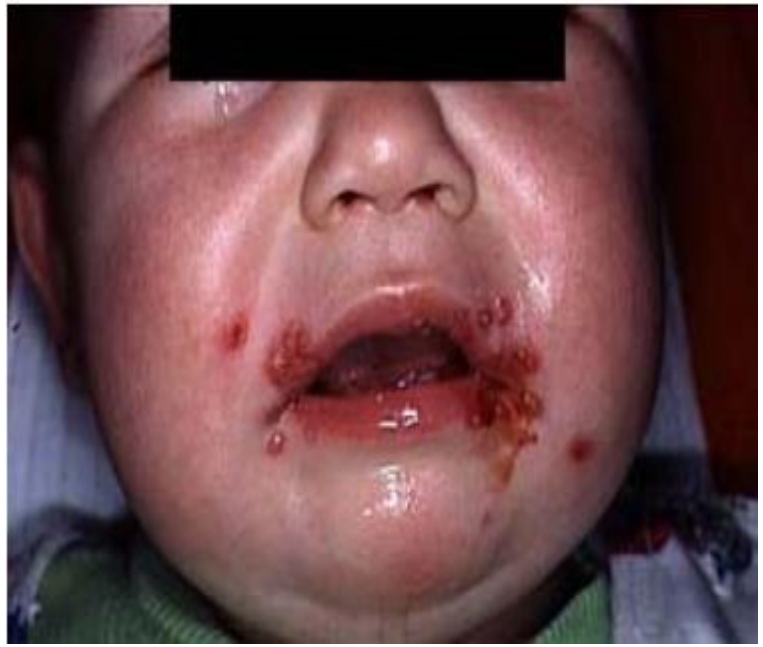
Herpangina



IM



Primary herpetic gingivostomatitis



Diagnosis and treatment

- Dx;by throat swab and culture to identify GABS,CBC with differential,high wbcs count and neutrophilia.
- Tx;we treat the GABS by antibiotic early to prevent acute rheumatic fever by penicillin V(given orally) or if allergic erythromycin,also symptomatic treatment with antipyretics and analgesia(Acetaminophen,avoid aspirin to prevent reye syndrome)
- Recurrent infection could be because of relapse(same bacteria didn't resolve because of non-compliance with medications or resistance),reinfection due to new exposure.
- Tonsillectomy is indicated in children with recurrent infection>7times in the past year or enlargement >5inch.

Common cold or rhinorrhea

- Definition; viral illness in which symptoms of rhinorrhea and nasal obstruction, sore throat are prominent but systemic s&sx are absent or mild such as myalgia and fever in contrast to flu.
- Causative agent rhinovirus, less common adeno and RSV.
- Why reinfection? multiple serotypes, host immunity is not protective, virus is able to change antigenic part and behave as if there were multiple serotypes.
- ddx;allergic rhinitis(itching&sneezing), foreign body(unilateral&foul-smelling discharge), sinusitis(headache&facial pain), pertussis(onset of paroxysmal cough), strep nasopharyngitis(nasal discharge that blocks the nares)
- Investigations not needed but could be necessary to rule out certain conditions; PCR,nasal smear for eosinophilia if you suspect allergic rhinitis, bacterial culture if you suspect strep rhinopharyngitis.
- Treatment is mainly supportive and symptomatic relief;decongestants mainly topical(oxymetazoline,phenylephrine), rhinorrhea & cough 1st generation anti-histamines, analgesia use paracetamol.

Sinusitis

- Usually comes with an URTI,rhinosinusitis,self-limiting and resolves by its own.
- Could be more serious and isolated caused by a bacteria,the most common causative agent is strep pneumoniae.
- We don’t suspect sinusitis before the age 4-5 years because complete maturation(pneumatization) of the **maxillary sinuses occurs at age 4-5,7 for frontal sinuses.**
- Clinical presentation;nasal congestion, discharge, fever, cough, headache, facial pain, decreased sense of smell.
- Diagnosis; symptoms of URTI that persist for at least 10 days without improvement.
- Tx;amoxicillin(GIVEN ORALLY)

Dx is Based on history ; one of the following :

1-persistant hx of RTI including cough ,nasal discharge for>10-15 days without improvement .

2-OR severe symptoms ;temp.>39 *C and purulent nasal discharge for 3-4 days.

3-Worsening URTI symptoms after initial improvement .

- Indications for antibiotic use in acute sinusitis;-Usually self limiting , no specific treatment needed).

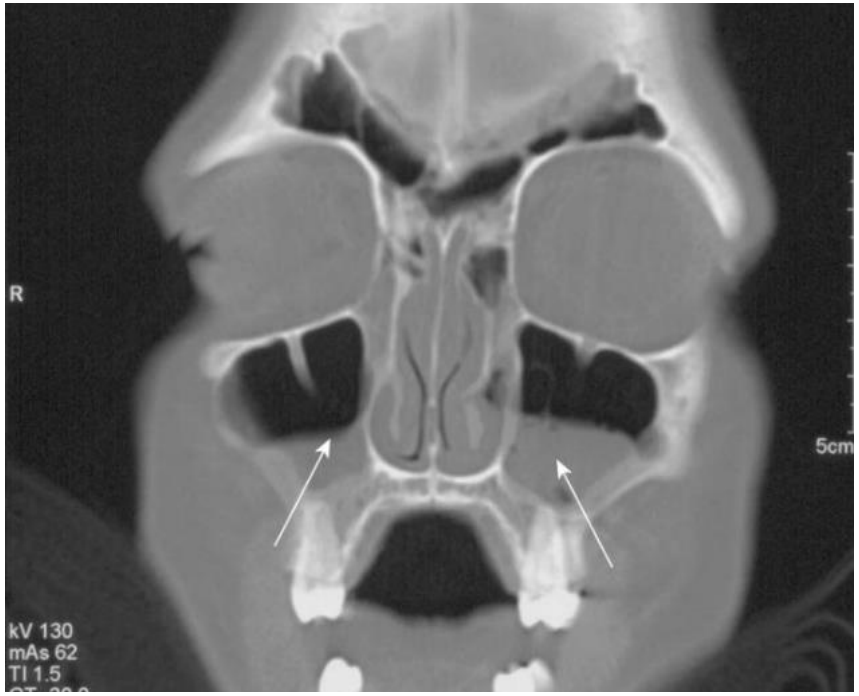
1-Persistent acute sinusitis. cough and congestion>10 days

2-Severe acute sinusitis. Toxic appearing child with high fever or worsening of sx after initial management

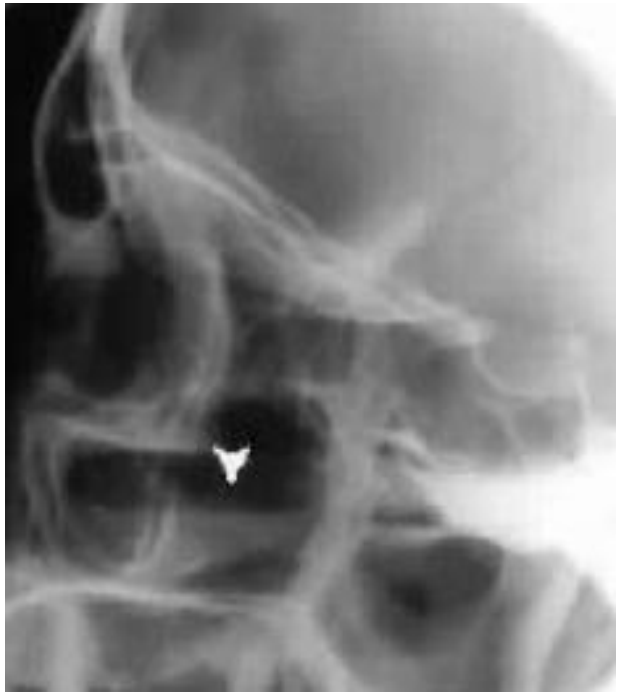
3-Toxic child with suspected complications

Air-fluid level in acute sinusitis

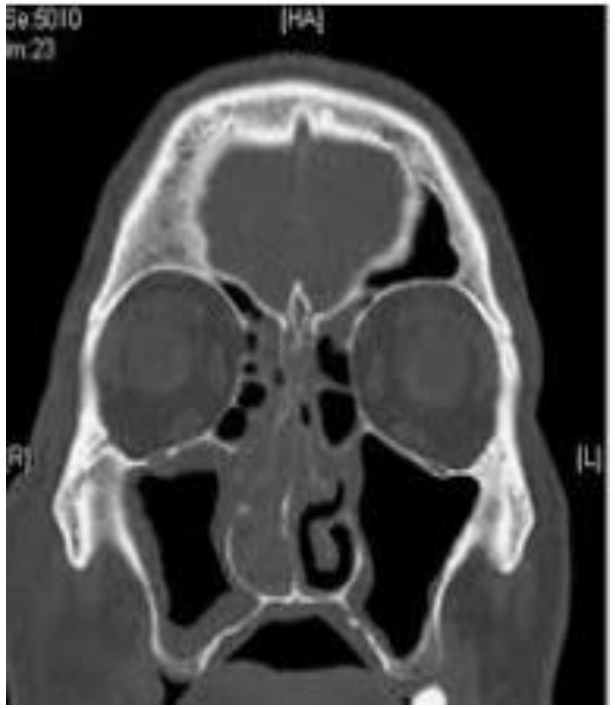
CT

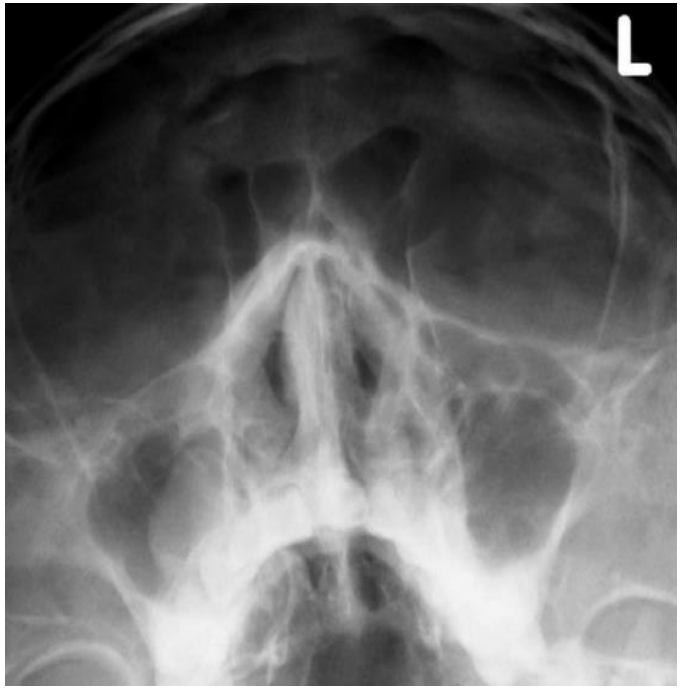


Plain film



Mucosal thickening in chronic sinusitis



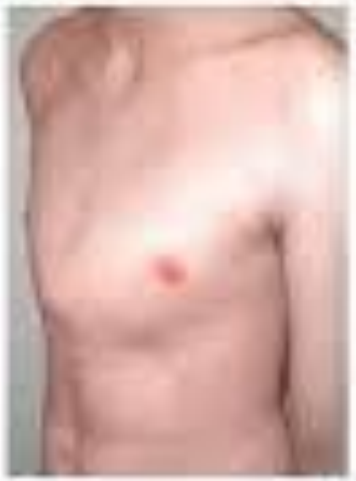


Complications of URTI

1. LOCAL
   - If viral superinfection by bacteria
   - Spread downwords to cause LRTI(RSV,CORONA,INFLUENZA,ADENOVIRUS)
   - Abscess, otitis media ,mastoiditis.
   - Orbital complications;preseptal cellulitis.
   - Cranial complications;cavernous sinus thrombosis,meningitis.
2. Systemic
   - Rheumatic fever,post-strep glomerulonephritis,igA nephropathy.

Chest deformities

Pectus carinatum



Pectus excavatum

Infectious

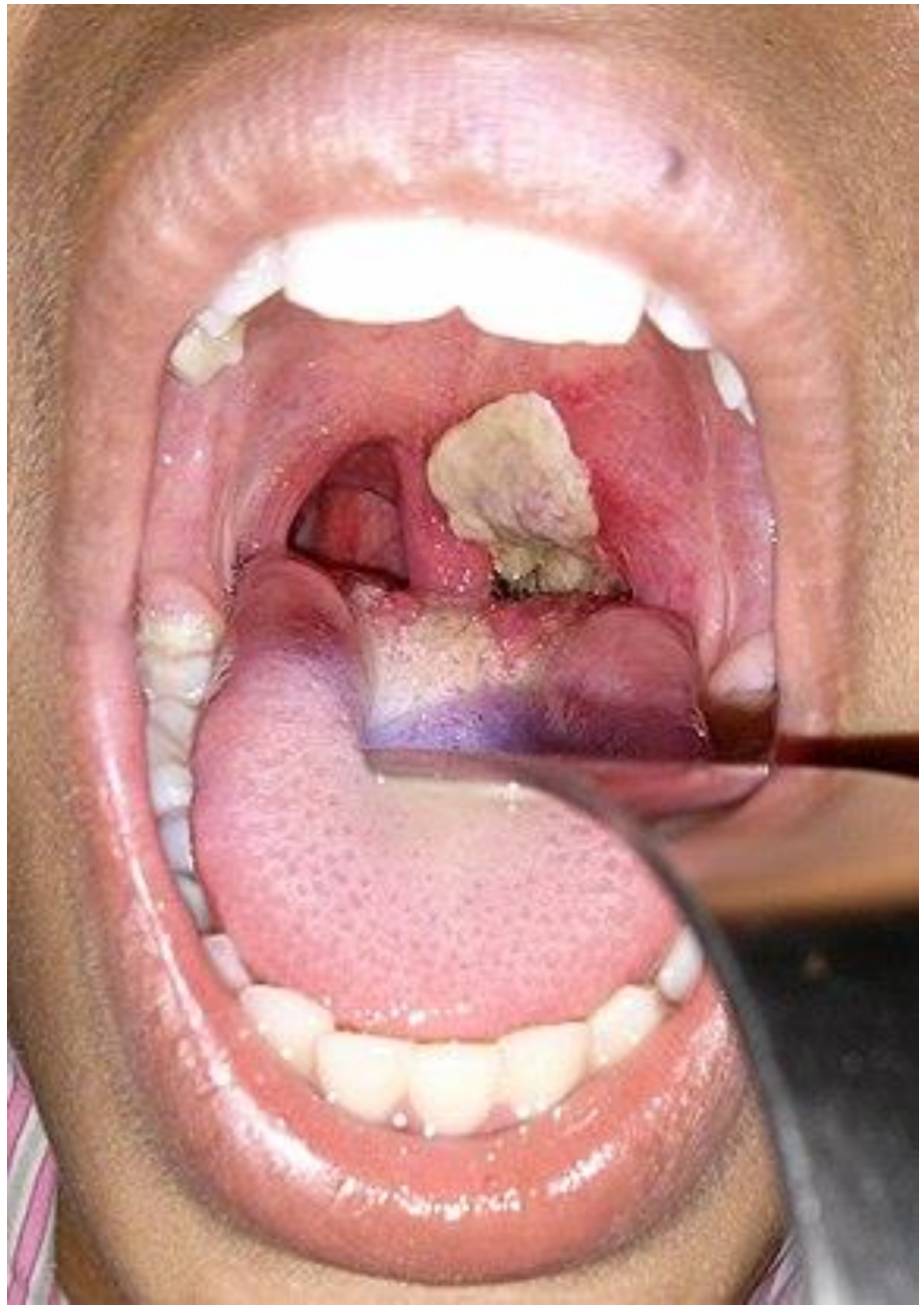
Pertussis aka whooping cough or 100 day disease

- Causative organism; Bordetella pertussis(gram negative bacilli)
- Most susceptible <5 years.
- Transmitted by droplets,incubation period 6-14days(1-2 weeks)
- Clinical picture(3 phases of the disease)
    1. Catarrhal phase(preparoxysmal)-catarrh is inflammation of the mucous membranes,this phase include;low grade fever,wheeze,cough,rhinorrhea,excessive lacrimation,conjunctival injection.(1-2 weeks)
    2. Paroxysmal phase characterized by recurring intense episodes of coughing,a paroxysm is a series of coughs in rapid succession with increasing in its intensity and at the end theres a large inspiration producing the whoop sound.(2-4 weeks)
    3. Convalscent phase(1-2 weeks);the cough decreases in frequency and intensity.

- Diagnosis; history of sick contact,incomplete immunization history(2,3,4 months a booster at 18 months,preschool age)
- Defenitive diagnosis is by isolation and culture or fluorescent AB stain.
- Complications; pneumonia, pneumothorax, atelactesis, post cough vomiting and seizure,anoxic brain damage.
- Tx;erythromycin
- Prevention for contact;if less than 7 years we give booster of erythromycin if >7 years erythromycin for 7-14 days.

Diptheria

- Droplet born,incubation period 2-4 days.
- Causative organism is corynebacteria diptheria(gram negative non spore forming)
- Clinical presentation;sore throat and hoarseness,stridor,pseudomembrane formed in the pharynx and larynx(gray-green membrane),cervical lymphadenopathy.
- Diagnosis;culture from the membrane(tellurite culture media),incomplete vaccination hx,sick contact,lymphocytosis.
- Complications;neuritis(peripheral neuropathy of the palate,eye,extremities),myocarditis.
- Treatment;anti-toxin within the first 48 hrs,penicillin +erythromycin,you stop the antibiotic after 3 negative cultures.



Tetanus

- Causative agent;clostridium tetani(spore forming anaerobe)
- Incubation period 2-4 days
- Clinical picture;refusal of feeding and 2 days later,locked jaw,stiffness of all muscles and board like rigidity,opisthotonous stiff limbs(Arching backward),stiffness of the facial muscles(risus sardonicus),clenched fists,convulsions,involvement of laryngeal muscle causes cyanosis.
- Causes of death;prolonged laryngeal spasm(cyanosis and hypoxemia),obstruction by laryngeal secretions,pneumonia.
- Treatment;anti-toxin/penicillin.

Gastroenteritis

1. Most frequent cause of gastroenteritis in developed countries is rotavirus infection, which

accounts for up to 60% of cases in children under 2 years of age, particularly during the winter and early spring.(most common cause of sporadic severely dehydrating diarrhea in young children-peak from 3-15months),other viruses are norovirus,astrovirus,adenovirus.(feco-oral transmssionn)

2. Bacterial causes (food poisoning)→ less common in developed countries but may be suggested by the presence of blood in the stools.

- Campylobacter jejuni infection, the most common of the bacterial infections in developed countries→ associated with severe abdominal pain,transmitted by dogs,cats with diarrhea.
- Salmonella-transmitted by undercooked eggs.
- Shigella (3rd most common)→ dysenteric type of infection(diarrhoea containing blood or mucus), with blood and pus in the stool, pain and tenesmus. Shigella infection may be accompanied by high fever.,dysentric type can cause HUS(microangiopathy of the kidneys)
- Cholera and enterotoxigenic Escherichia coli infection → **profuse, rapidly dehydrating diarrhoea.**
- Hemolytic uremic syndrome (**HUS**) is a condition that affects the blood and blood vessels. It results in the destruction of blood platelets (cells involved in clotting), a low red blood cell count (anemia) and kidney failure due to damage to the very small blood vessels of the kidneys

3.Parasites

- Giardia
- Cryptosporodium

- In gastroenteritis there is a sudden change to loose or watery stools often accompanied by vomiting.There may be contact with a person with diarrhoea and/or vomiting or recent travel abroad.
- All of these viruses tend to spread on hands that have touched either an infected person's stool or surfaces contaminated with infected stool. For this reason, young children – especially those just starting to learn good hygiene – are particularly vulnerable to viral gastroenteritis.
- Bacterial or food-poisoning; Food that hasn't been prepared or stored properly can grow bacteria on its surface, and these bacteria sometimes produce irritating chemicals called toxins. If a child eats the germ-filled food, symptoms of gastroenteritis are triggered either by the bacteria themselves or by their irritating byproducts.
- Intestinal parasites – Intestinal parasites can be spread to children on dirty hands, on the soiled surfaces of toys and bathroom fixtures, and in contaminated water or food.

- Conditions that can mimic gastroenteritis **(DDx)**

a. Systemic infection → Septicaemia, meningitis

b. Local infections →Respiratory tract infection, otitis media, hepatitis A,UTI

c. Surgical disorders→ Pyloric stenosis, intussusception, acute appendicitis, necrotizing enterocolitis, Hirschsprung's disease.

d. Metabolic disorder→ Diabetic ketoacidosis, adrenal insufficiency

e. Renal disorder→ Haemolytic uraemic syndrome.

f. Other→ Coeliac disease, cow's milk protein allergy, lactose intolerance.

- Dehydration leading to shock is the most serious complication and its prevention or correction is the main aim of treatment.

- **Investigations**

a. Stool culture → (indications)if child appears septic, if there is blood or mucus in the stools, or the

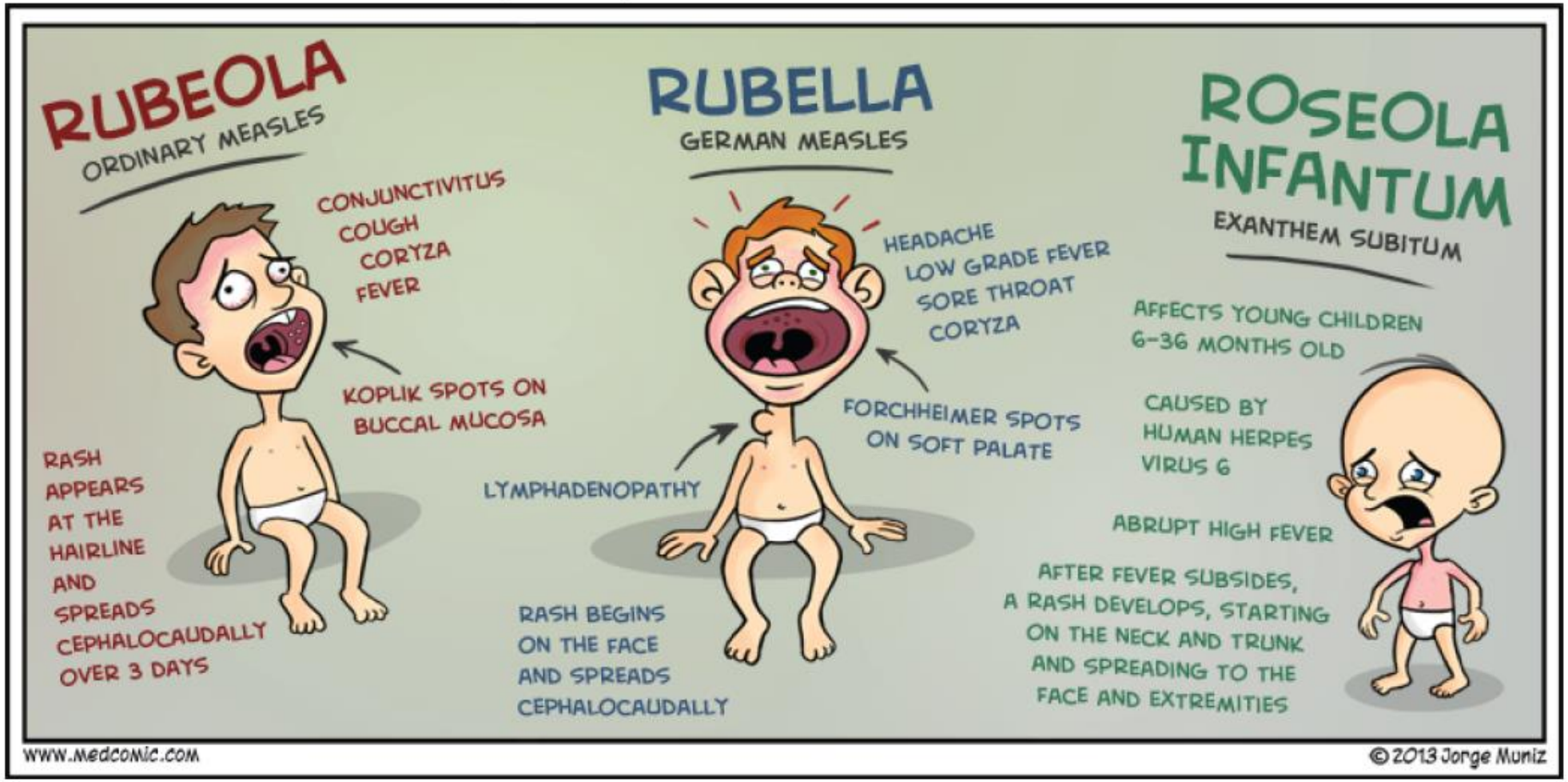
child is immunocompromised, after recent travel, if not improved by day 7.

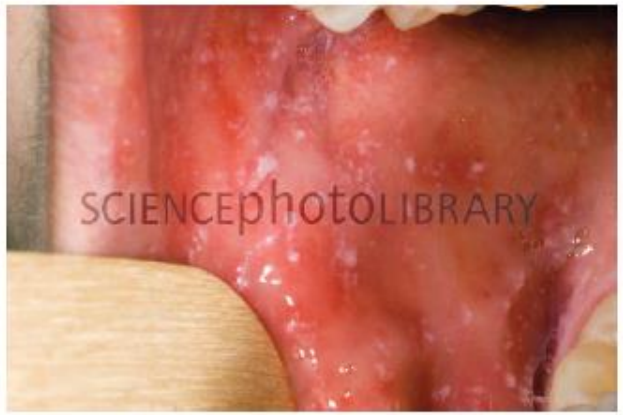
b. Plasma electrolytes, urea, creatinine(KFT)and glucose.(to rule out the ddx mentioned before and to assess the hydration level and its electrolyte disturbances)

c. blood culture(if you suspect sepsis)

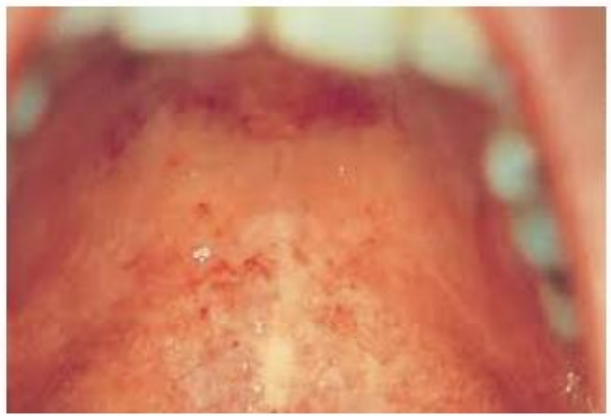
- **Treatment;oral rehydration or IV, consideration of antidiarrheal if E.coli O157:H7,c.difficle is not suspected,antibiotics only in selected cases,if vomiting is severe and surgical conditions has been excluded an antiemtic can be considered(ondansetron safe in children).**
- **Tx if the case is c.difficle stop the abx use. Antibiotics are ineffective against toxins,it can prolong the shedding duration of salmonella.**
- **Indications for abx;contact with a known case of bacterial gastroenteritis.**

Study common child exanthems and enanthems from your printed slides.

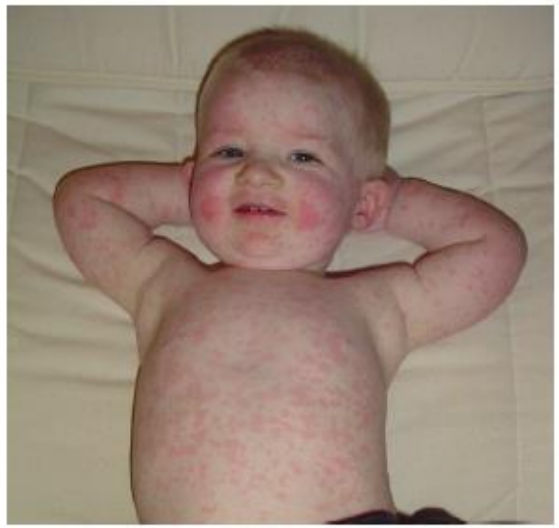




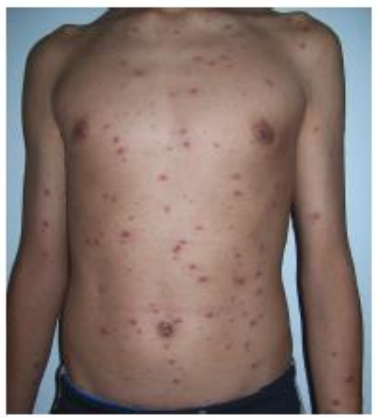
Koplik's spots on the third pre-eruptive day



forschheimer spots

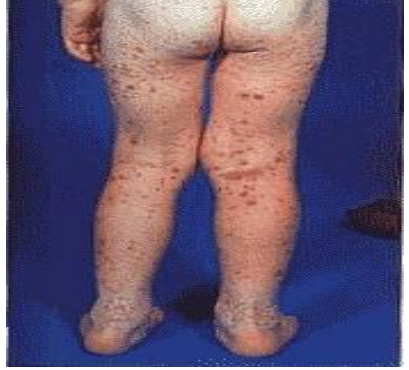


Erythema infectiosum (5th disease,slapped cheek)



Chickenpox

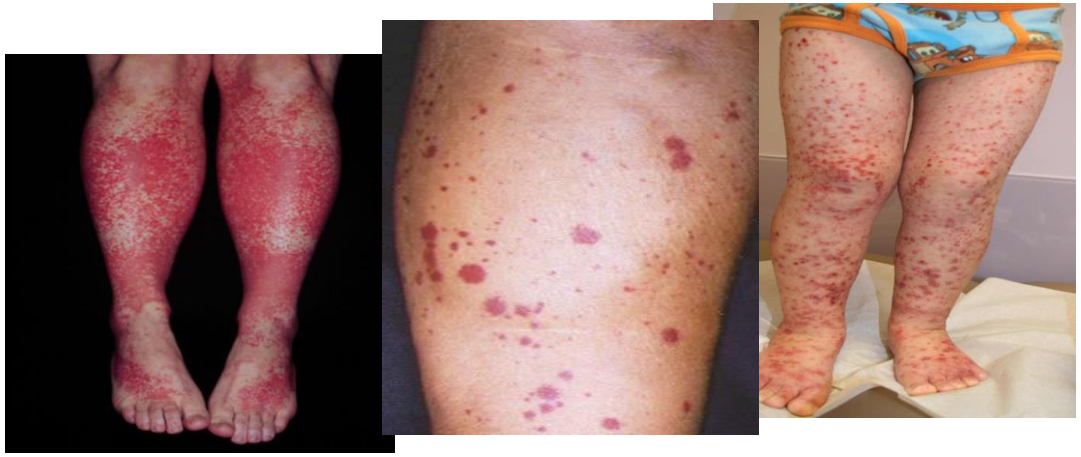
HSP(IgA vasculitis)-small blood vessels

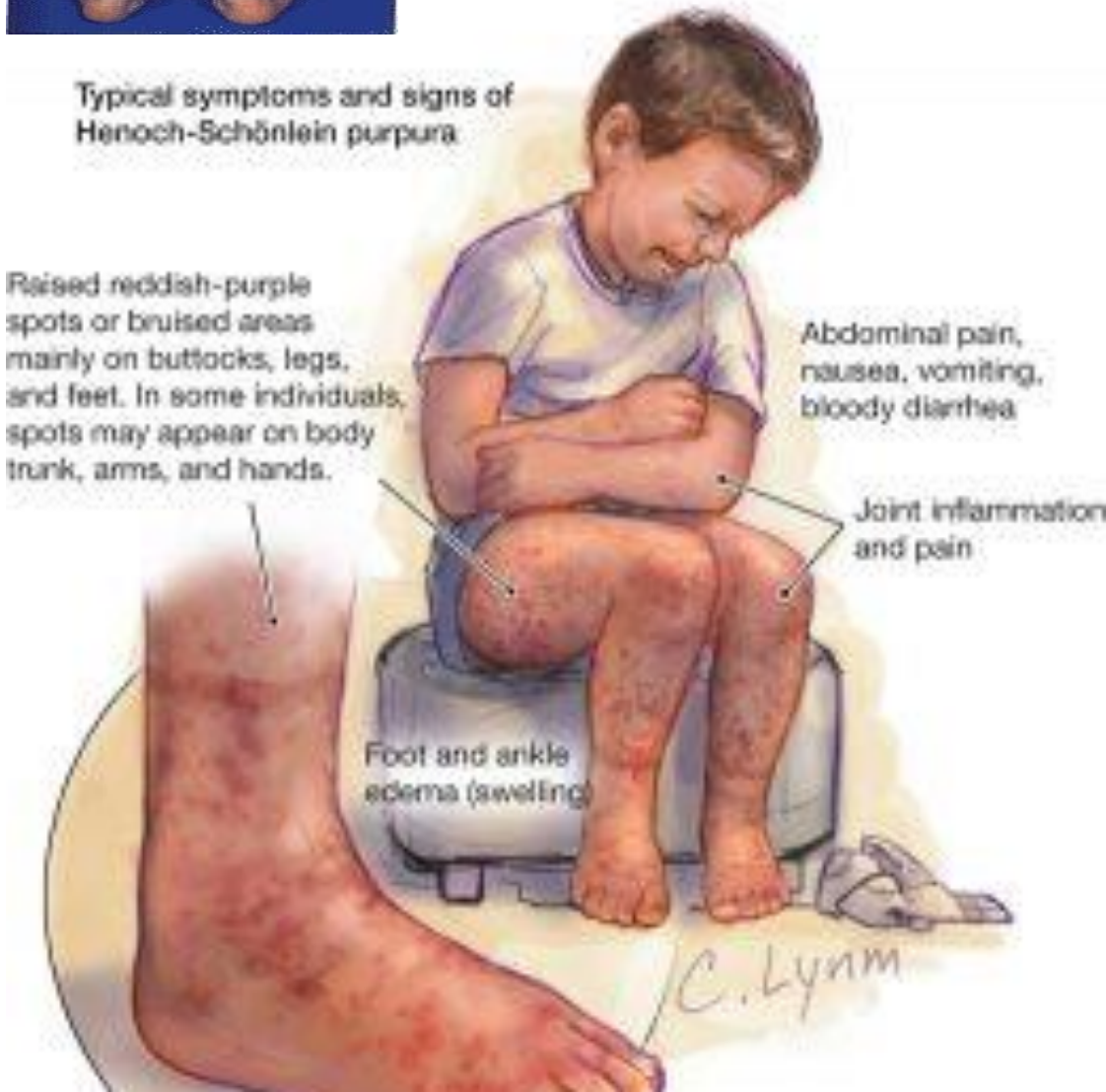


.It started on the foot and over the three days the rash went up to cover thigh

.The rash is reddish-purple raised and not itchy lesions starts on foor,legs and ascend to the thighs and buttocks.

There is a history of abdominal pain around the umbilicus.no diarrhea or vomiting.There is a history of joint swelling&pain in both knees and there is limping.There is history of red urine.No dysuria or frequency.No change in color of stool





- Occasions for vaccines
  1. First month of life(BCG)
  2. 2 months(DaPT,HiB,HBV),IPV
  3. 3 months(DaPT,HiB,HBV),IPV+OPV
  4. 4months(DaPT,HiB,HBV),IPV,OPV
  5. 9 months measles alone,OPV
  6. 12 months or one year(MMR is given alone at 1 year of age).
  7. 18 month,3 Boosters of DaPT,MMR(final MMR),OPV.
  8. Preschool age(4-6 years)dT,OPV(مع صف عاشر زيOPV)
  9. 15 years(10th grade)-dT
- Polio vaccine starts as IPV at 2 month then given IPV+OPV at 3 months&4 months (المطعوم الخماسي)then continues with every vaccine occasion as OPV to 6 years except when the MMR given.OPV is given on 5 occasions 3,4,9,18,preschool.
- MMR: measles, mumps, and rubella (German measles): لقاح الحصبة والنكاف والحصبة الألمانية:اللقاح الثلاثي.(given at age 1 year and 18 months)
- pneumococcal is given with rota at ages 2,3,4 or 2,4,6 depending on the country and a booster at age of 18 months.
- Varicella and hepatitis A vaccine is given at age of 1 year,varicella has a booster at age 18 months with the pneumococcal booster.

Schedule of the NIP and UNRWA in Jordan

Time of vaccination	Vaccine (s)	Comments
Within the first month of life	BCG	Only 1 dose
2 months of age (60+ days)	(DTaP, IPV, Hib: الخماسي المحسن), HepB, RotaV	pneumococcal
3 months (90+ days)	(DTaP, IPV, Hib), HepB, RotaV, OPV	pneumococcal
4 months (120+ days)	(DTaP, IPV, Hib), HepB, RotaV, OPV	pneumococcal
9 months	Measles, OPV	Monovalent measles
12 months	MMR	Not in NIP : Varicella, HepA
18 months	MMR, OPV[b], DTP[b]	Pneumococcal,varicella
6 years, first grade	OPV[b], Td	Reduced diphtheria vaccine
10th grade	Td	Not in NIP: At 11 y: HPV, MCV4

Additional vaccines outside the schedule

- Pneumococcal vaccine against streptococcus pneumonia at 2,3,4 months of age and 18 months.
- Meningococcal vaccine(MCV) against Neisseria meningiditis for travelers to crowded areas military,al-hajj.
- Rotavirus vaccine(optional expensive vaccine given at 2,3,4 months orally).
- Hepatitis A given after 1 year of age(& a booster at 18 months)
- Varicella recommended to give 2 doses after 1 year of age(12 and 18 months)

Types of vaccines

- **Live-attenuated:** **BCG, MMR, OPV, Rota,** Varicella, (oral typhoid, yellow fever), Nasal Influenza virus
- **Inactivated:** **DTaP** (toxoids and inactivated components) **(Tdap, Td, DTP), IPV, Hib** (polysaccharide conjugate), Hepatitis A (inactivated), Meningococcal, pneumococcal (polysaccharide conjugate or polysaccharide), Influenza virus (inactivated)
- **Genetically engineered** (recombinants antigens): Hepatitis B, HPV
- Live-attenuated vaccines are contraindicated in cases of cell-mediated immune defects and pregnancy. *OPV is the only vaccine contraindicated when household contains an immunocompromised member.*

Types of vaccines

Live attenuated(BMV OR nasal yellow typhoid)

- BCG
- MMR
- VARICELLA
- OPV
- ROTA VIRUS
- NASAL INFLUENZA
- YELLOW FEVER
- TYPHOID(oral).

Killed inactivated BB Dora Puts Makeup Always RIP.

- HiB
- HBV
- DaPT
- PCV(PNEUMOCOCCAL)
- MCV(MENINGOCOCCAL)
- Hep A
- Rabies
- Influenza
- POLIO(IPV)

General contraindications for vaccines

- Live vaccines for immunocompromised or pregnant but you can give killed inactivated to immunocompromised, remember breastfeeding is not a contraindication.
- Moderately to severly ill,remember mild illness is not a contraindication.
- Influenza and yellow fever to people with egg allergy.
- Previous severe anaphylactic shock or severe allergic reaction is always a contraindication(remember egg allergy is not a contraindication for MMR)
- Live-attenuated vaccines are contraindicated in cases of cell-mediated immune defects and pregnancy. *OPV is the only vaccine contraindicated when household contains an immunocompromised member.*

Special considerations

- Polysaccharide vaccine not before 2 year-give it conjugated with protein.
- MMR not before 1 year
- Hep A after 1 year, safe in liver disease.
- OPV don’t give to contacts of immunocompromised or patients taking prednisone.
- Rotavirus not after 8 months.
- Hib not before 6 weeks of age also not to a healthy child after 5 years of age.
- Pure **polysaccharide vaccines** are available for three diseases: pneumococcal disease, meningococcal disease, and Salmonella Typhi.

- Mention 3 moderate side effects for DTP
  - **Seizure**
  - Non-stop crying for 3 hours or more
  - High fever

Possible side effects to all vaccines

- Local reactions to injectable vaccines
- Anaphylaxis to the vaccine or one of it’s components (contraindications for further similar doses)
- Syncope
- fever

Not a contraindication!

- The followings are not contraindications to vaccine administration:

❑Mild illness with or without fever

❑Breast feeding

❑Local rxns or fever after previous vaccine

❑Preterm birth

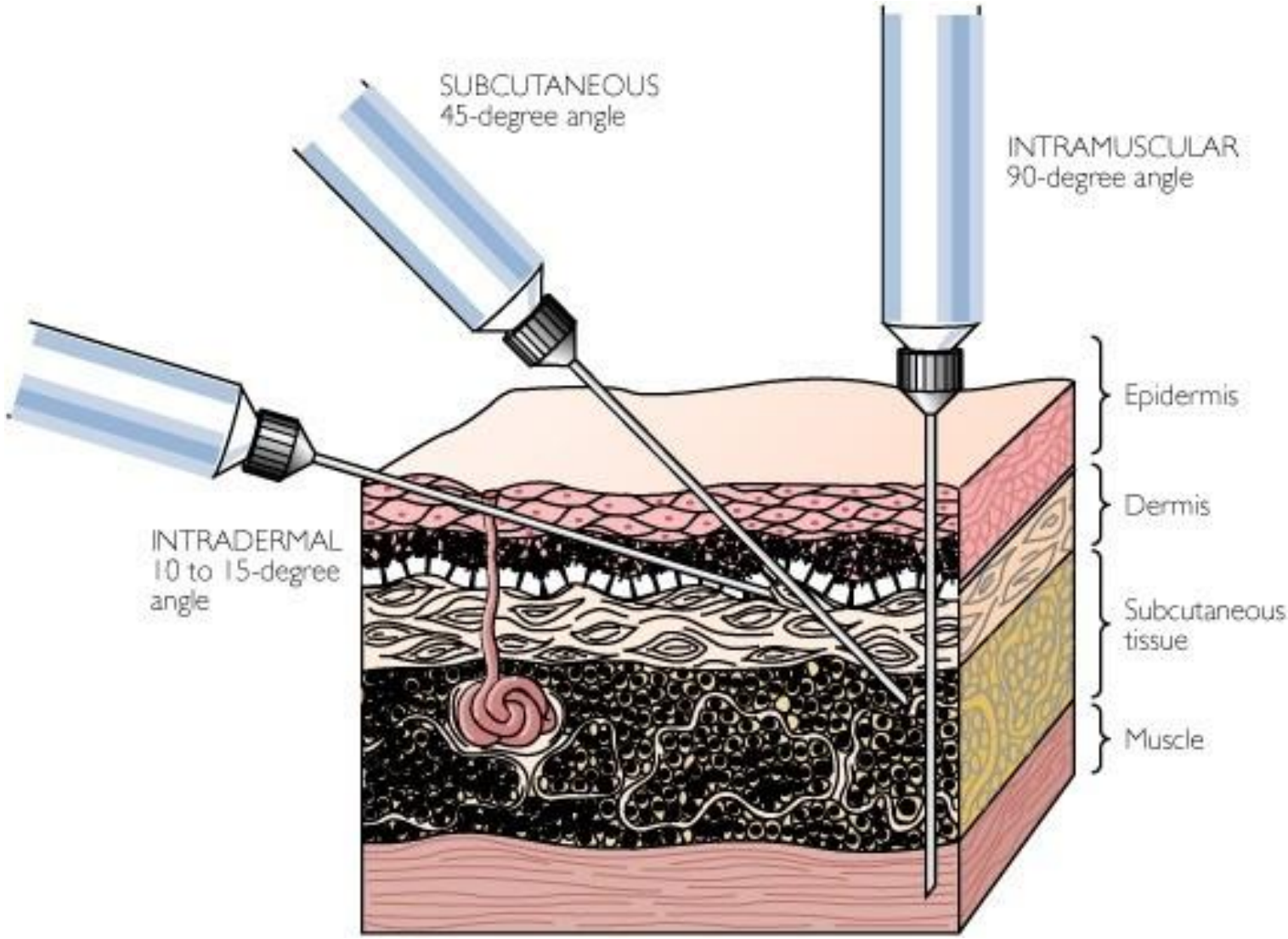
❑Penicillin allergy

❑Concurrent antibiotics use

❑Family history of seizure, controlled seizures

administration

- Most: IM (Ant-lat thigh or deltoid). Exceptions:
- Intradermal (ID): BCG (or SC)
- SC: MMR, Varicella, Polysaccharide vaccines, IPV (or IM)those given at 1 year of age except the IPV.
- Intranasal: Nasal influenza vaccine
- Oral: OPV, Rota



Meningitis

- General considerations;
- Route of entry: mucosal surfaces of respiratory and GI tract → viral replication in regional LN's→ viremia seeds the CNS.

WBC is high	**normal up to 5 and all lymphocytes**	**0**	**1**	**2**
Protein is normal	normal up to 40 mg/dl	0	1	**2**
Sugar is normal	normal more than than two third of serum	0	1	**2**

CSF findings beyond Neonatal period

CSF finding	Normal	Viral	Bacterial	Comment
WBC per HPF	0-5 all Lymphocytes	5-500 mainly lymphocytes.	Usually >1000 mainly PMNs.	In neonates, up to 25 cells is considered normal
Glucose	CSF:Serum ratio >2/3	Normal.	CSF:Serum ratio <1/2(bacteria consumes glucose)	
Protein	<40 mg/dl	Normal or mildly elevated.	Usually >100	
Appearance	Clear	Clear	Turbid	

Aseptic meningitis

- Infectious causes;viral infections;
  - Nonpolio-EV serotypes: Coxsackie A, B, echoviruses parechoviruses, Fecal-oral route(Conjunctivitis, pharyngitis, rash, herpangina, hand-foot-mouth disease).
  - Others: EBV, CMV, VZV, Arboviruses, HHV6, Influenza A and B, mumps
  - Clinical manifestations of meningitis ; Nuchal rigidity, bulging fontanel, manifestations of particular viruses (e.g., rash, conjunctivitis, pharyngitis, diarrhea)
  - In older children: +positive meningeal signs(meningeal signs cannot be seen before the age of 2)
  - ***Clinical features cannot reliably differentiate viral from bacterial meningitis; and CSF profiles may overlap, particularly during early bacterial meningitis.***

✖***Non-infectious causes;*** **Drugs:** Ibuprofen, Azathioprine, IVIG,**Malignancy:** Lymphoma, Leukemia,**Autoimmune:** Sarcoid, Behcet's disease, SLE,**Other causes:** Epidermoid cyst, Heavy metal poisoning, Intracranial hemorrhage.

✖Dx: CSF PCR.

Bacterial meningitis: Common causes

- <3 months – Group B streptococcus(maternal vaginal normal flora) (39 %), gram-negative bacilli (30% percent), *Strep pneumoniae* (14 %), *N. meningitidis* (12 %)

- ≥3 months and <10 years – *S. pneumoniae* (nasopharynx normal flora)(47 %), *N. meningitidis* (32 %)

- Adolescents: *N. meningitidis* (nasopharynx normal flora)(55 %)

Bacterial meningitis: CLINICAL FEATURES

- ✖ Fever and symptoms and signs of meningeal inflammation, often preceded by symptoms of upper respiratory infection

- ✖ In infants (none specific): fever, hypothermia, lethargy, respiratory distress, jaundice, poor feeding, vomiting, seizures, irritability, or bulging fontanel

- ✖ Petechiae and purpura with any of the bacterial pathogens but most commonly seen in *N. meningitidis*

Meningeal signs:

- 1- Nuchal rigidity: active movement or passive neck flexion
- 2- Kernig sign
- 3- Brudzinski sign
- Signs are present in 80 percent of children > 2y of age with bacterial meningitis at the time of presentation.

Meningeal signs:

- **Kernig's** is performed by having the supine patient, with hips and knees flexed, extend the leg passively. The test is positive if the leg extension causes pain. The **Brudzinski's sign** is positive when passive forward flexion of the neck causes the patient to involuntarily raise his knees or hips in flexion.

Bacterial meningitis: Evaluation

- **Blood cultures** are positive in 50% of patients with bacterial meningitis

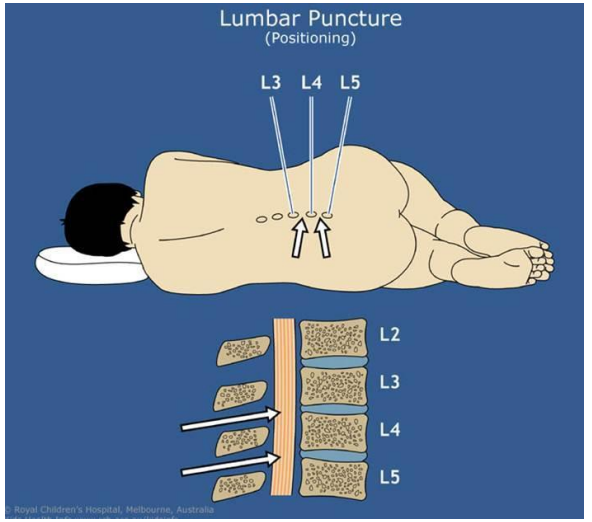
- **CSF examination: Lumbar puncture**
- **Contraindications:** cardiopulmonary compromise, signs of increased ICP, papilledema, focal neurologic signs, and skin infection at site for LP.

- ***It is essential that antimicrobial therapy not be delayed if there is a contraindication to or inability to perform an LP, or if the LP is delayed by the need for cranial imaging***

- Send CSF for(4 tubes): cell count and differential, glucose and protein, Gram stain and culture. Send viral PCR as appropriate

ASIS same level of l4,go one space above or one space below

- **Gram stain:** + in 90% of pneumococcal meningitis and 80% of meningococcal meningitis.
- Gram-positive diplococci suggest *S. pneumoniae*
- Gram-negative diplococci suggest *N. meningitidis*
- Gram-negative coccobacilli suggest Hib.
- Gram-positive cocci suggest GBS.



- **Rapid diagnostic tests and latex agglutination test,not recommended because its not spenicifc nor sensitive.**
- **Negative blood culture doesn’t rule out bacterial meningitis.**

Bacterial meningitis: management

- ABC (hypovolemic or septic shock?)
- IV fluids

- Administration of first dose of empiric antibiotics

- Complete CNS examination daily to assess progress.
- After csf culture treat specifically.

- Head circumference daily if <18 months.

Bacterial meningitis: management

- **Major pathogens** : *S. pneumoniae* and *N. meningitidis*
- third-generation cephalosporin (e.g., cefotaxime, ceftriaxone) and vancomycin.
- **Use of dexamethasone** and timing

Complications of bacterial meningitis

1. Focal disability such cranial nerve palsies or hearing loss
2. Developmental delay
3. Hydrocephalus
4. SIADH
5. Intracranial abcess
6. Seizures,ataxia

Meningococcal Disease

Neisseria meningitidis, also known as meningococcus, is a bacterium, that causes **meningitis and/or septicaemia (meningococcemia)**

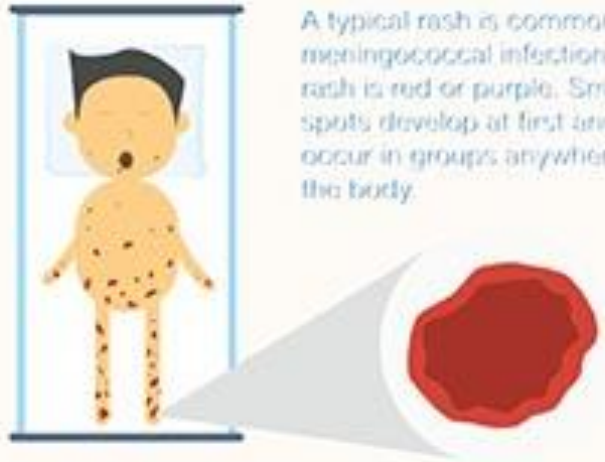
Signs and symptoms

High temperature

Vomiting

Headache

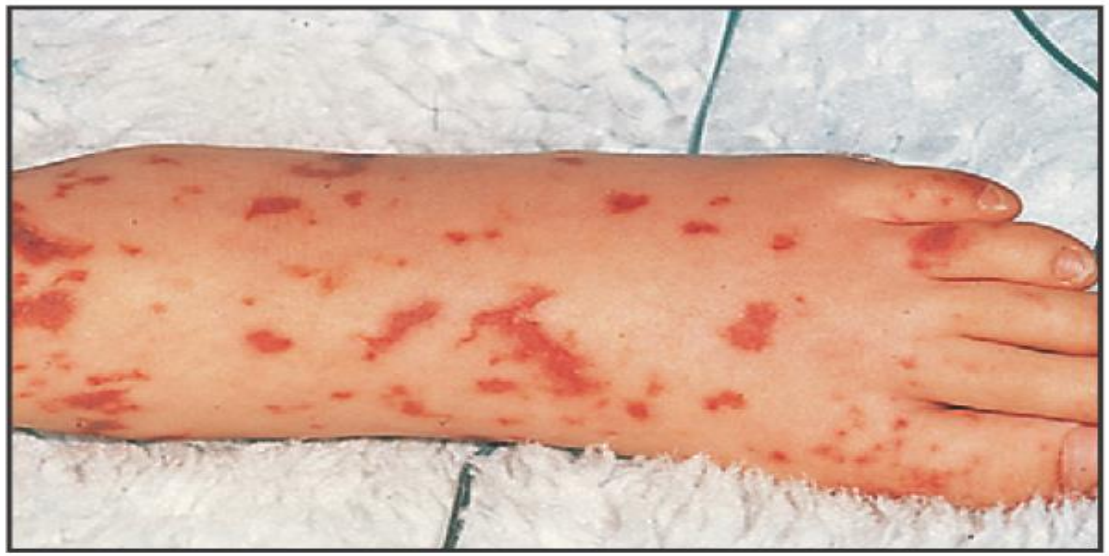
Meningococcemia

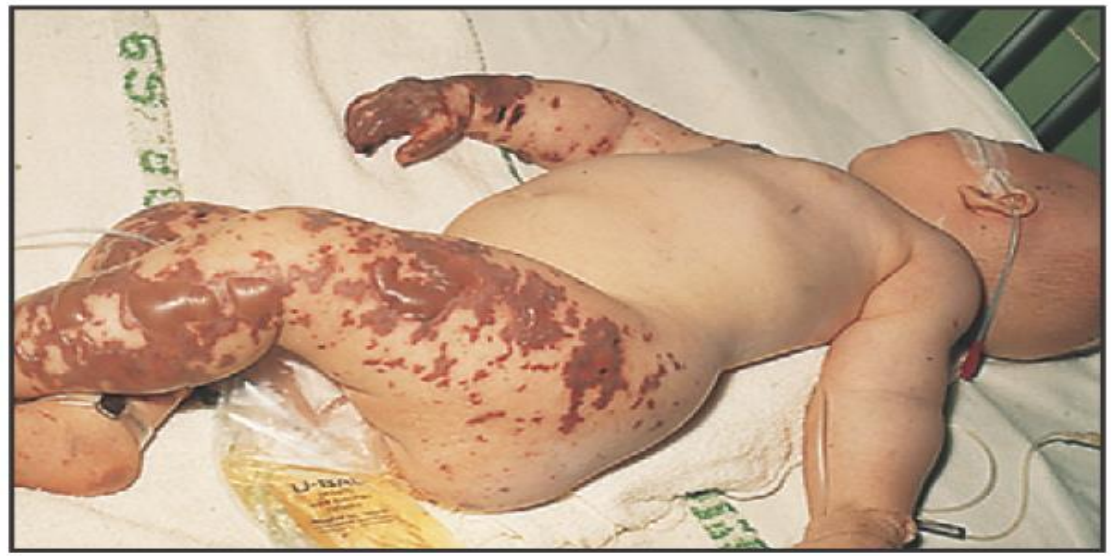


A typical rash is common with meningococcal infection. The rash is red or purple. Small spots develop at first and may occur in groups anywhere on the body.

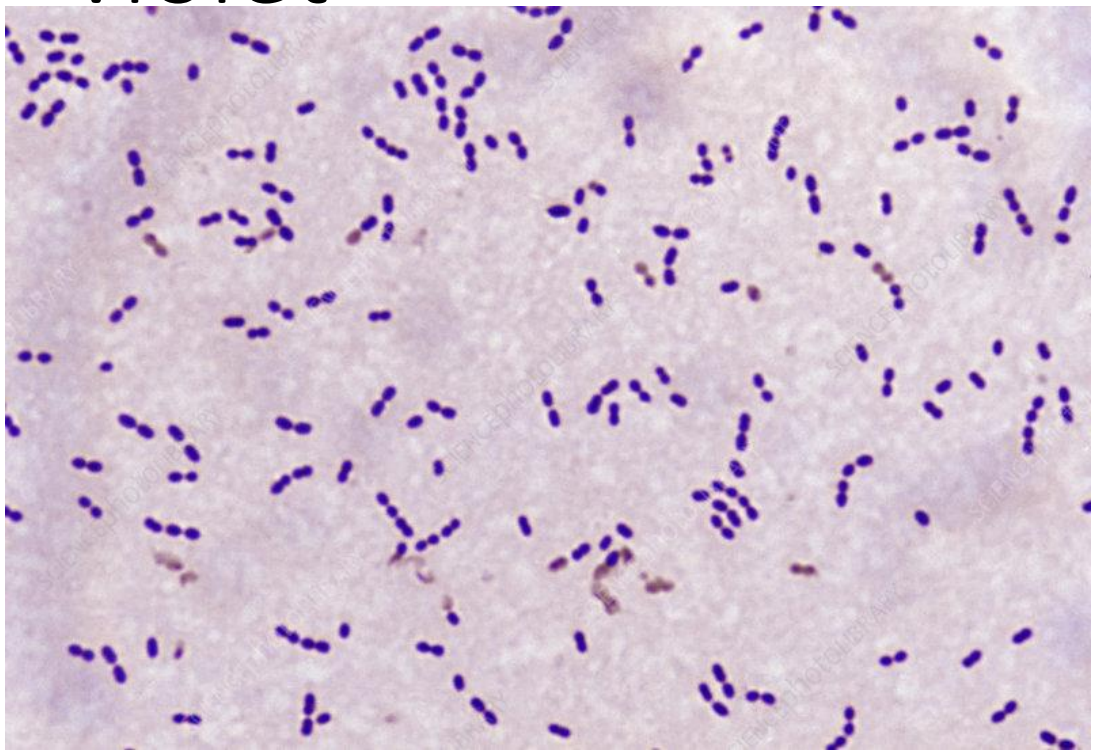
Call to emergency!

If you suspect meningitis or septicaemia - **get medical help immediately**

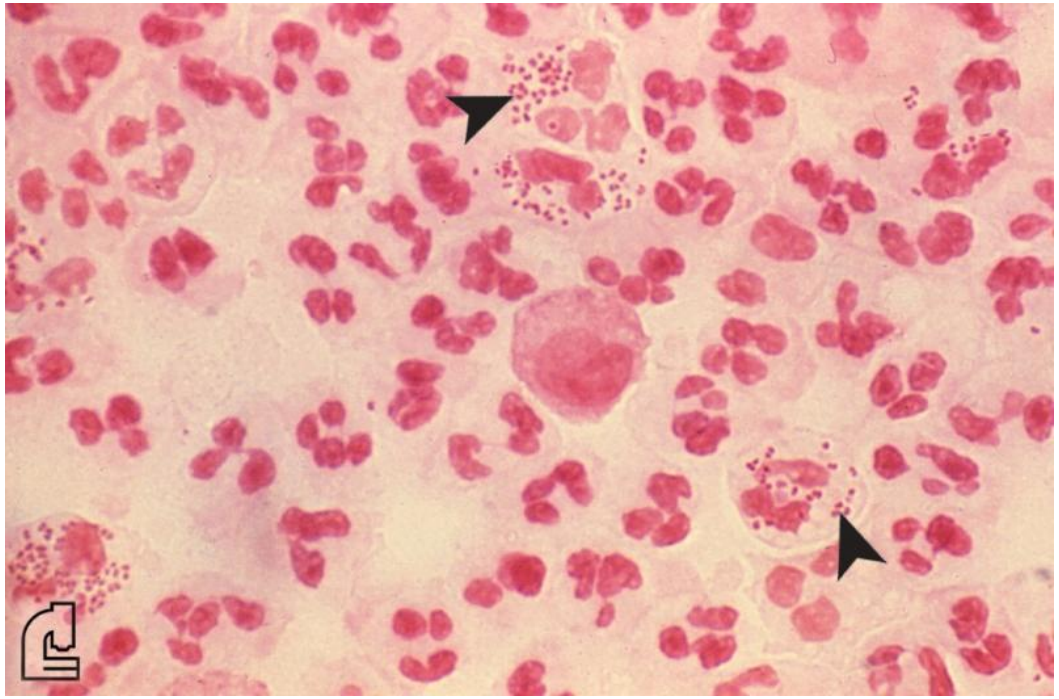




Gram stain-negative is red and positive is violet

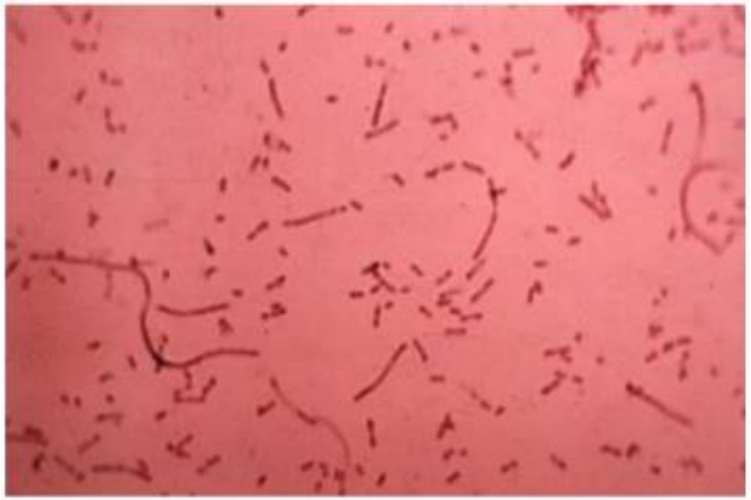


Gram positive diplococci is strep.pneumonia
Vancomycin+3rd generation cephalosporin(ceftriaxone) 10-14 days



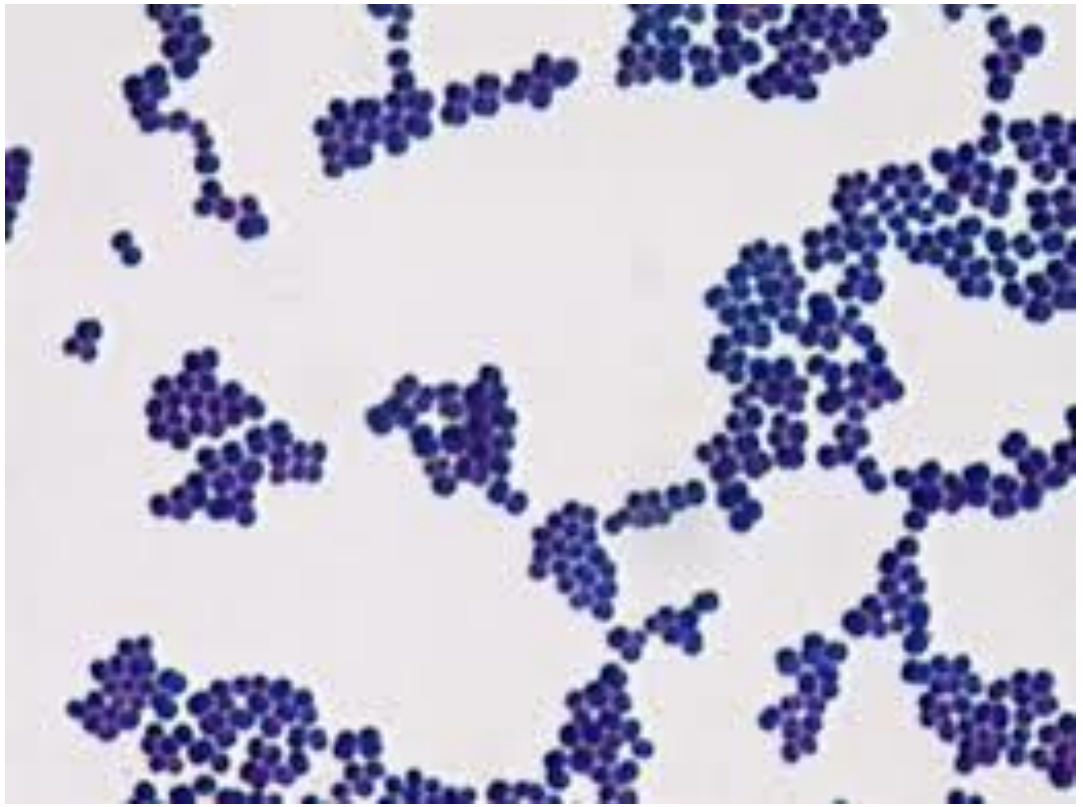
Gram negative diplococci is Neisseria meningiditis
Penicillin or 3rd generation cephalosporin(ceftriaxone) 5-7 days

Gram Negative Coccobacilli



Haemophilus influenzae

Gram positive coccibacilli is hemophilus influenza
3rd generation cephalosporin(ceftriaxone) 7-10 days



Gram positive cocci is GBS
Penicillin or 3rd generation cephalosporin(ceftriaxone) 7-10 days

- Sunset eyes
- Increased head circumference
- Dilated scalp veins
- Bulging anterior fontanele

herpetic meningo-encephalitis(HSV-1,HSV-2)

- Findings of csf is of that for viral (mild elevation of wbcs,differential shows more lymphocytes,glucose csf:serum ratio normal,protein mild elevation,rbcs found(80% of cases),RBC's were crenated (ie not fresh) and there was xanthochromia(due to cell lysis)in the CSF sample.
- Dx; by viral pcr
- Tx;give acyclovir
- **crenation** describes the formation of abnormal notched surfaces on cells as a result of water loss through osmosis.
- Xanthochromia:is the yellowish appearance of cerebrospinal fluid that occurs several hours after bleeding into the subarachnoid space caused by certain medical conditions, most commonly subarachnoid hemorrhage.

Exanthems

- Refer to dr Dawood Hamed slides on the e-learning website, the lecture is only 20 slides and it’s a high-yield topic.

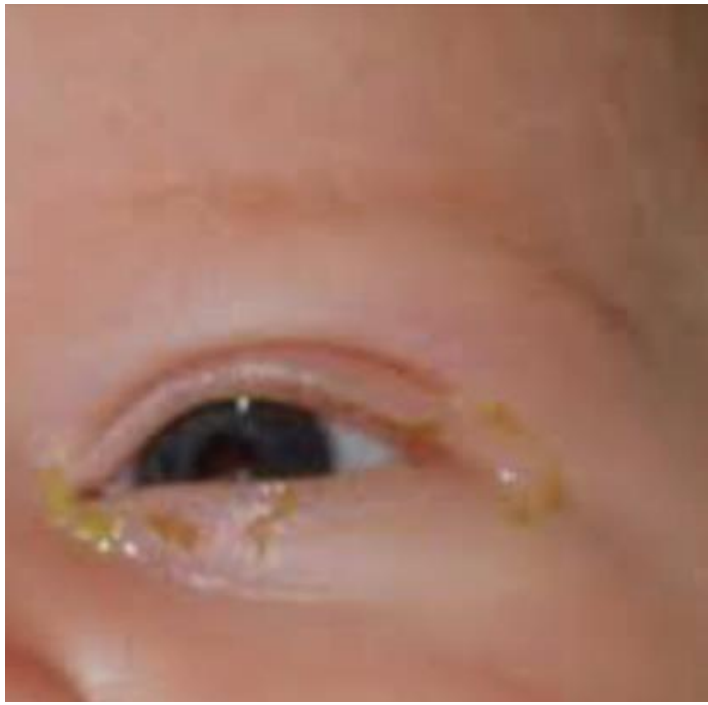
Neonatology

Common eye problems

- Congenital nasolacrimal duct obstruction(CNLDO).
- Happened in 20% of infants in the first year of life
- More prevelance in prematures
- Mostly resolve by 1 year(90%) of cases.if no improvement we can do propping

Management

- simple observation
- Clean eye
- massage of the lacrimal sac (Crigler)
  - 10 massage s each time 3 times /day
- Application of topical antibiotics when a bacterial superinfection occurs

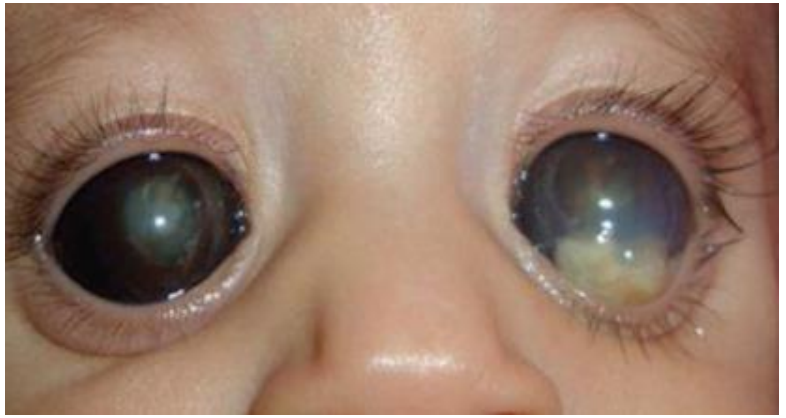


When to see a pediatric ophthalmologist?

- If you’re not sure of the diagnosis,differentials include;
  - Primary congenital glaucoma(PCG)
  - Foreign body
  - Corneal infection

Primary congenital glaucoma

- Improper development of the trabecular meshwork and the anterior chamber angle,causes inadequate drainage of the aqueous humor leads to increased IOP and stretching of the sclera causing enlargement of the globe aka bupthalmus/primary congenital glaucoma.
- **usually bilateral.**
- **symptoms include;**
    1. **Large cornea;**normal horizontal **corneal** diameter is 10.5 mm,measurements greater than 12 mm,are highly indicative of glaucoma.
    2. **Bupthalmus**,large eye globe because of scleral stretching
    3. **Blepharospasm**;involuntary eye twitching or blinking
    4. **Watery tearing**
    5. **Photophobia**
    6. **Cloudniess** of the cornea
- Complications;optic nerve damage
- Management;needs urgent surgery



Risk factors

1. Family history of PCG,pathogenic variant in the family(mode of inheritance could be autosomal dominant or recessive)
2. Congeintal rubella syndrome
3. Sturge-weber syndrome if the facial port-wine stain involves the upper and lower eyelids.

- Not very common happens in 1:10,000
- Ddx: infantile glaucoma **Infantile glaucoma** develops between the ages of 1-24 months.

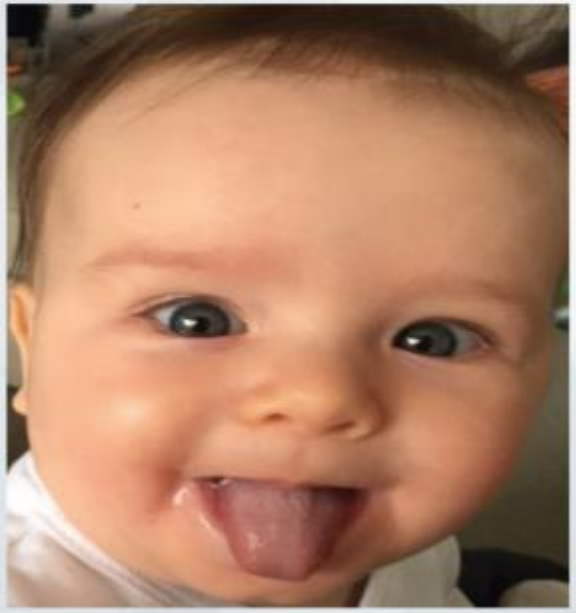
Differentials include;1.psedostrabismus which is an optical illusion because of wide nasal bridge,we can differentiate it by presence of symmetrical light reflex
2.transient neonatal strabismus which is a transient strabismus that goes away intermittently happens because of immature eye muscles,resolves by 2-4 months

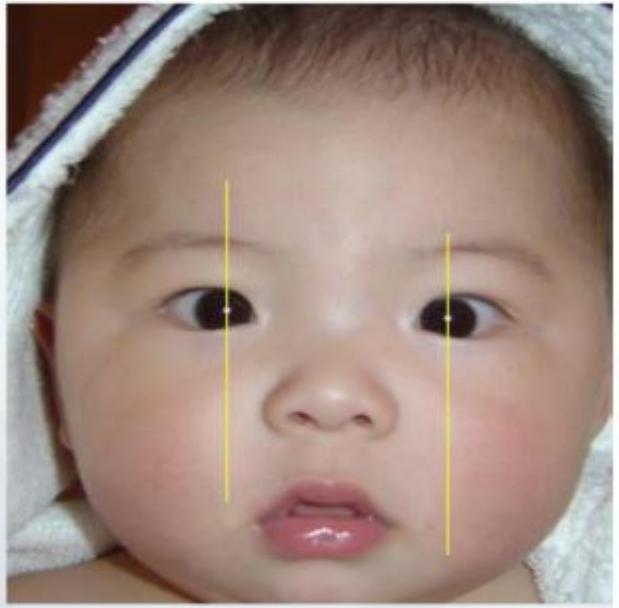
Strabismus – "squint that goes away"

Transient neonatal strabismus

- *NORMAL ocular alignment*
- intermittent
- ***Resolves by 2-4 months[1,2]***

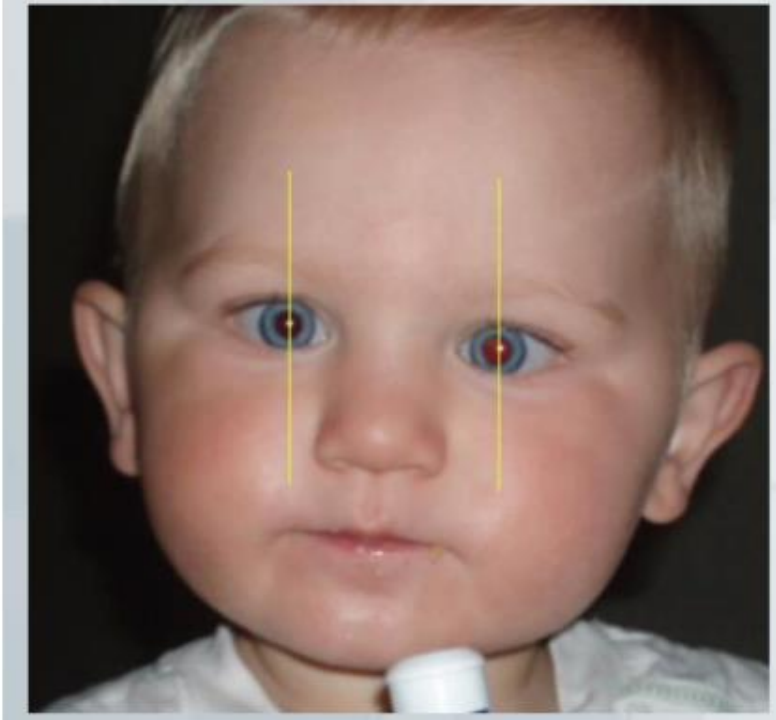
Pseudo-strabismus: Optical Illusion

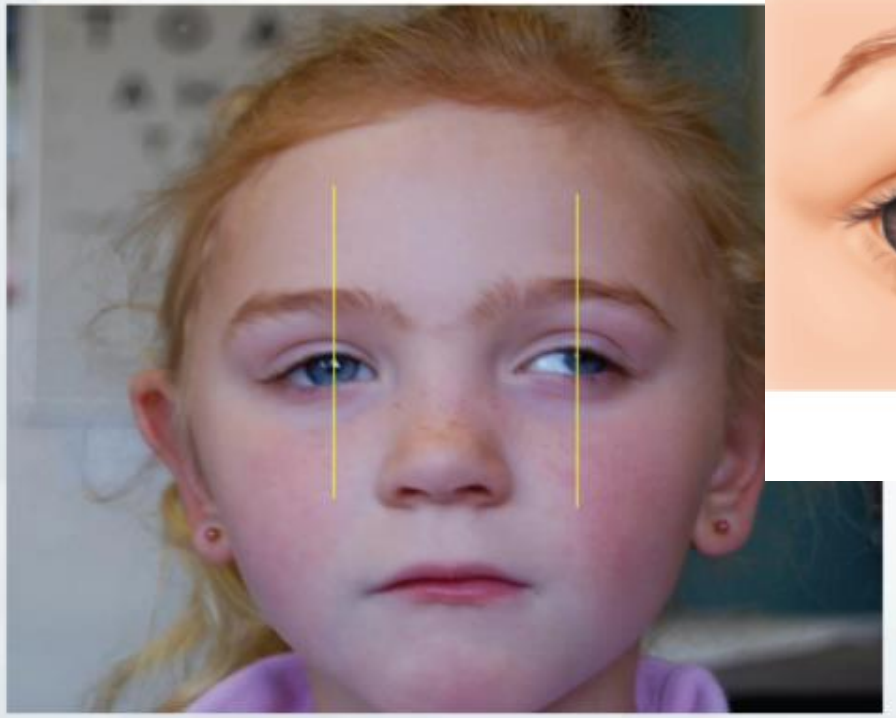


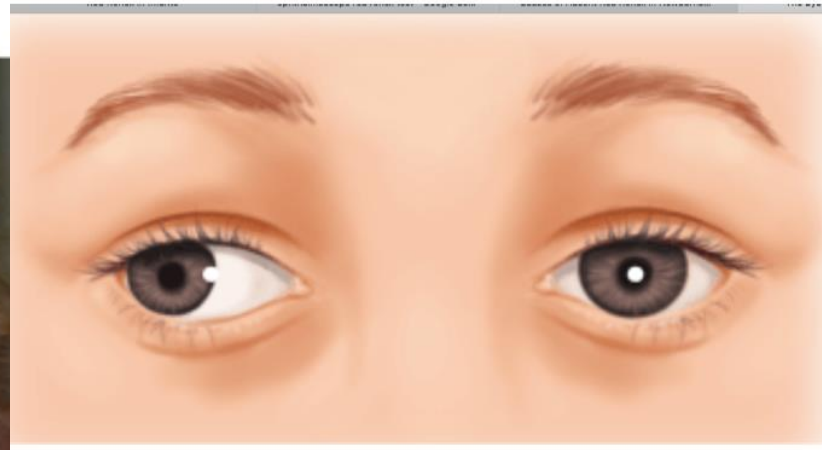


- Wide nasal fold/bridge of nose
- Intermittent – looking sideways
- "see both ears"
- Corneal light reflex - symmetry

True strabismus – variable direction, size and frequency







Asymmetrical pupillary reflex

Consider:

- CAUSE? – *secondary cause until proven otherwise*
- EFFECT ON VISION DEVELOPMENT – AMBLYOPIA

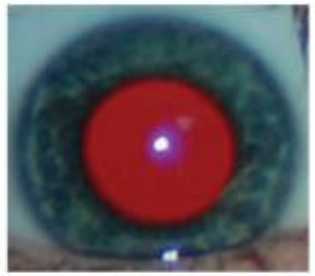
- **True trabismus is when theres asymmetrical light reflex needs referral to know the cause to avoid the amblyopia complication,in different directions.**
- **Amblyopia**, also called lazy eye, is a disorder of sight in which the brain fails to process inputs from one eye and over time favors the other eye

Red reflex

Should be done before newborn discharge

Good Red Reflex

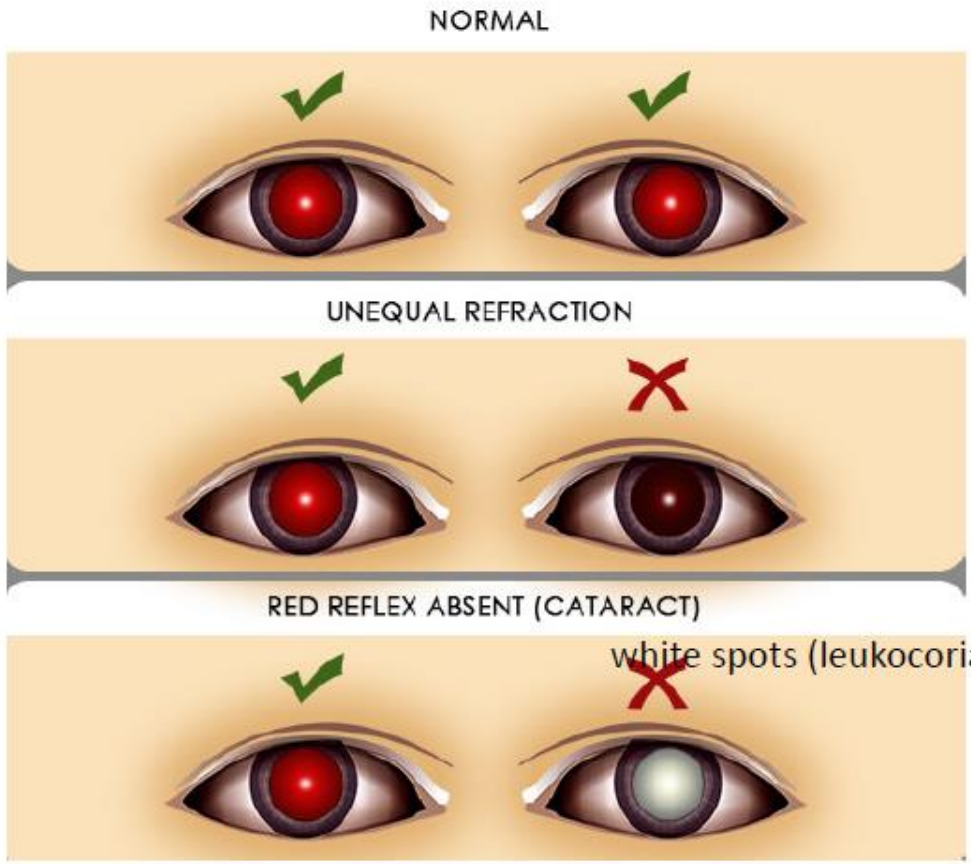
- Bright red
- Symmetric
- How to acquire the reflex?
  - Dim room
  - Direct ophthalmoscope
  - Simultaneous viewing of both eyes at arm's length



Red reflex should be equal in both eyes

Use direct ophthalmoscope from 2 to 3 ft away from the patient in a darkened room. The infant will usually be interested in the light and look directly toward it.

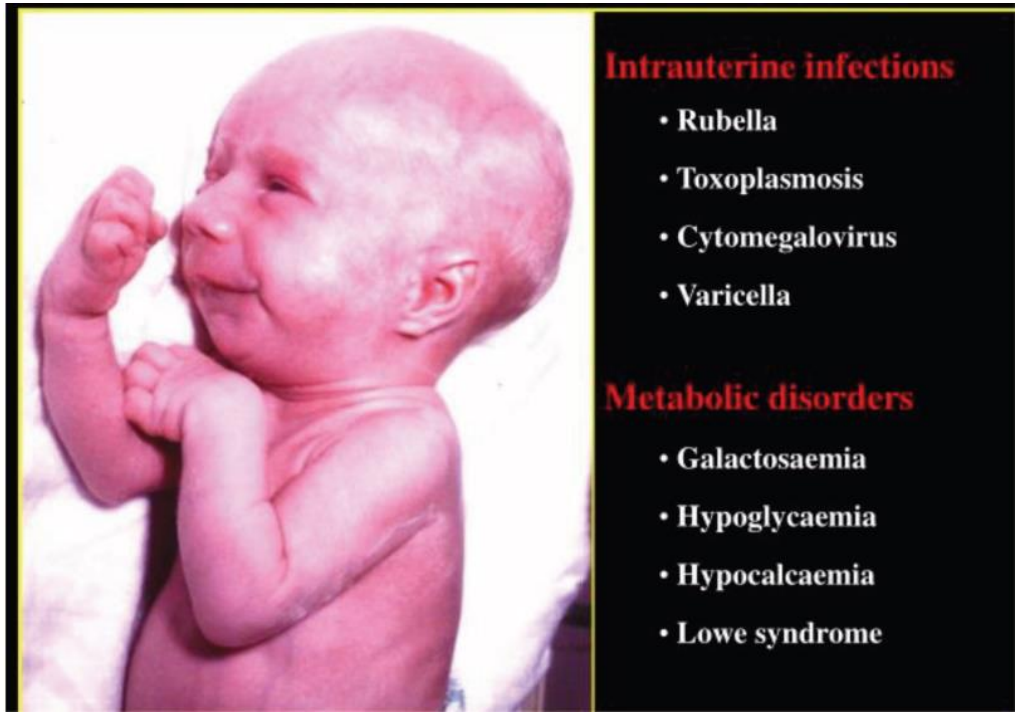
Interruption of Abnormal red reflex



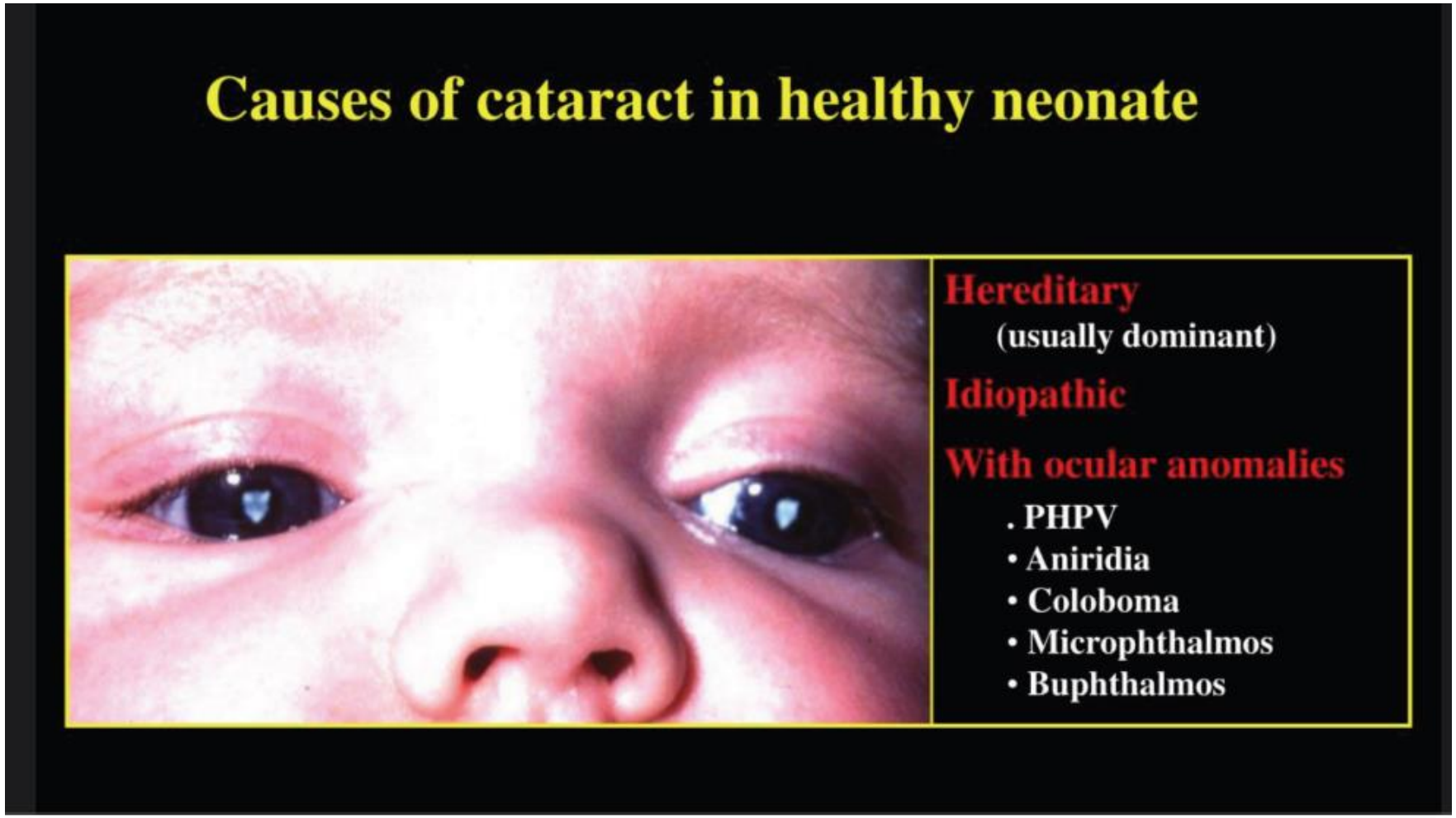
white spots (leukocoria)

Leukocoria

- Causes;
  1. Retinoblastoma 1 in 18-30,000 live births.
  2. Congenital cataracts
     - **Systemic causes**;metabolic such as galactosemia and galactokinase deficiency,hypocalcemia,hypoglycemia,lowe syndrome,infections such as congenital rubella syndrome,TORCH infections,syndromes;trisomies 13,18,21.
     - **Non-systemic causes**; isolated hereditary cataracts could be AD AR or X-linked,better prognosis than other systemic abnormalities**.**

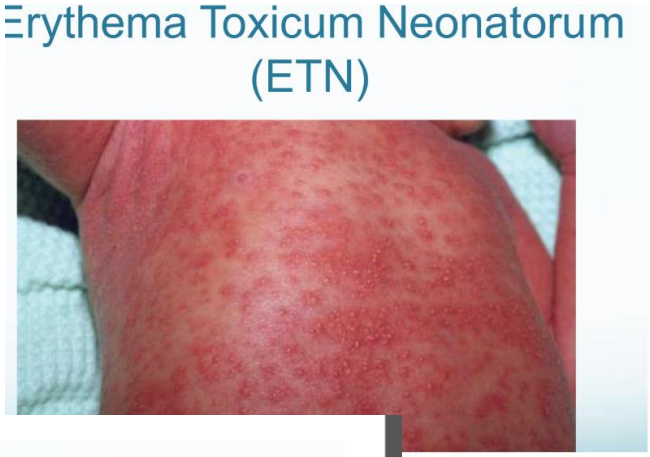


Congenital Cataracts and retinoblastoma are urgent but not an emergency the only emergency is the PCG.



Common skin manifestations

- ERYTHEMA TOXICUM NEONATORUM

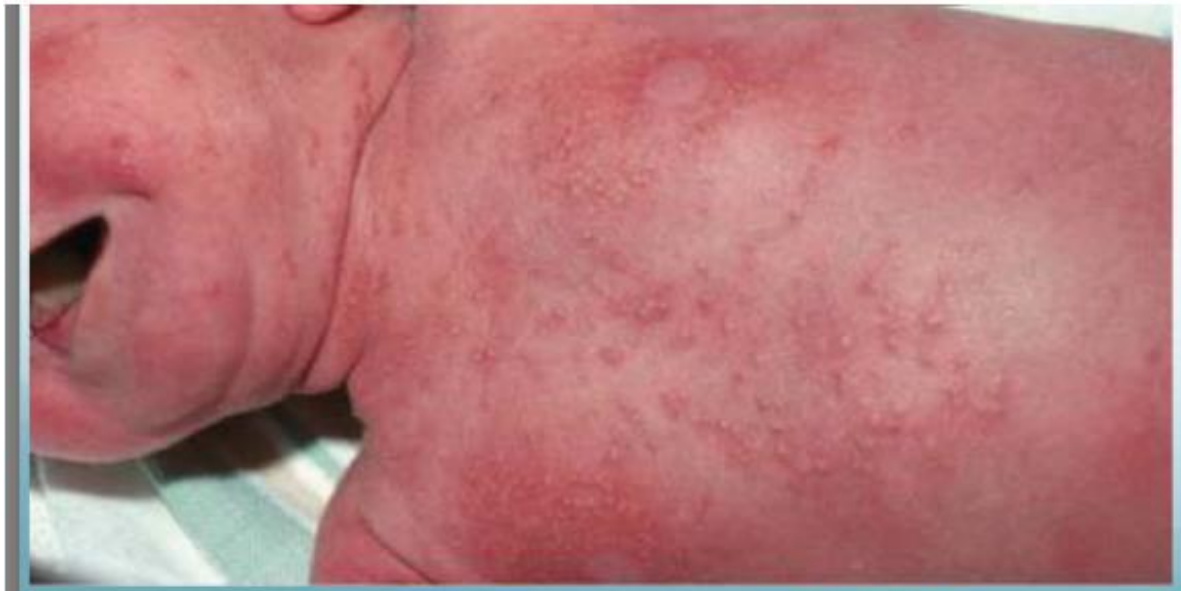


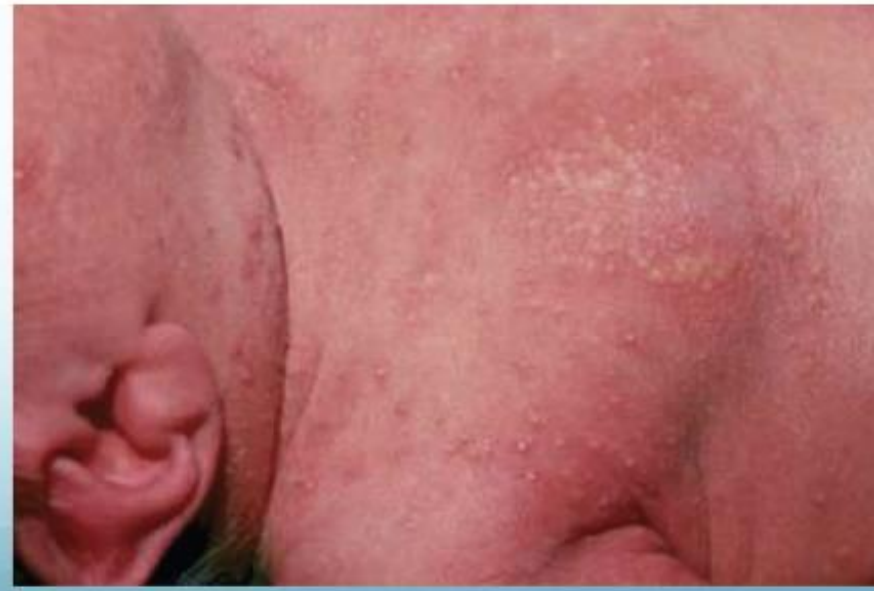
Erythema Toxicum Neonatorum

- Occurs in 50% or more of healthy normal newborns
- 1st-3rd day of life
- Resolves spontaneously ~2 weeks
- Classic eruption:
  - Erythematous blotchy macules, papules or pustules
  - Mainly on trunk, face and proximal limbs

Etiology of ETN

- **Etiology**: Unknown
  - GVH against maternal lymphocytes
  - Immune response to microbial colonization through hair follicles
- **Dx**: Clinical appearance alone
  - Wright/Giemsa stain→sheets of eos w/ few scattered neuts.
  - Skin Bx is rarely needed
- **Tx**: Parental reassurance





ETN

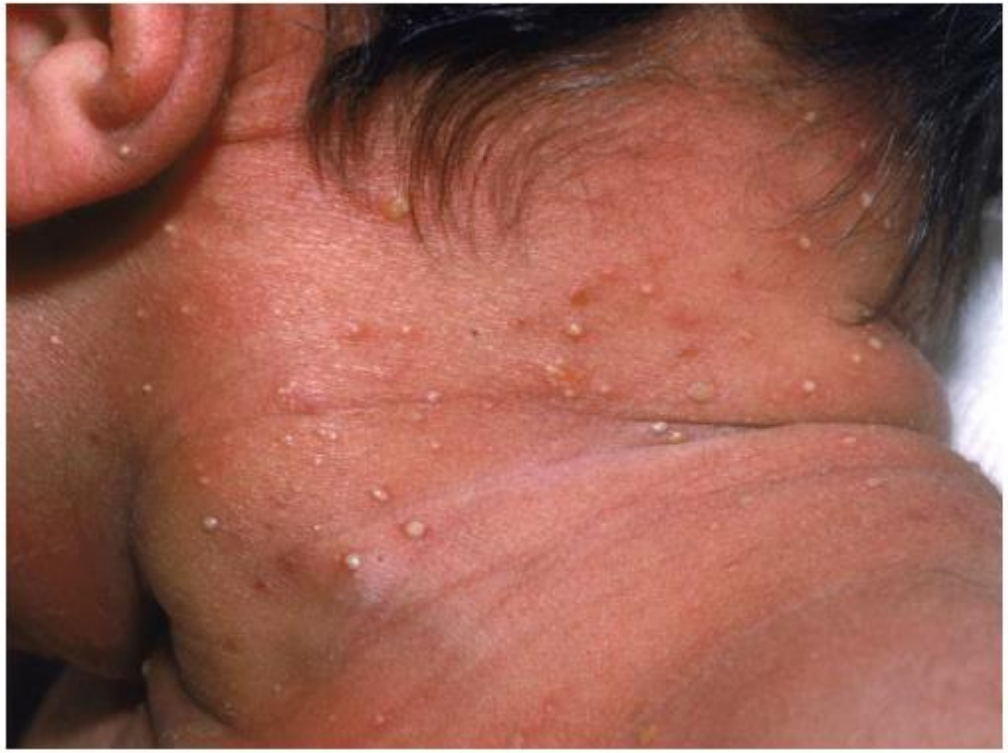
- Appears 1st on **FACE** → trunk & extremities or anywhere on the body **EXCEPT** palms/soles
- **Histologically**:
  - Subcorneal pustule filled with eosinophils and occasional neutrophils
- 15% peripheral **eosinophilia**

TRANSIENT NEONATAL PUSTULAR MELANOSIS

Transient Neonatal Pustular Melanosis

- The etiology is unknown.
- No familial predisposition
- More common in term-gestation infants
- More in African
- present at birth
- benign, asymptomatic

Ref:
https://emedicine.medscape.com/article/1112258-overview#a3

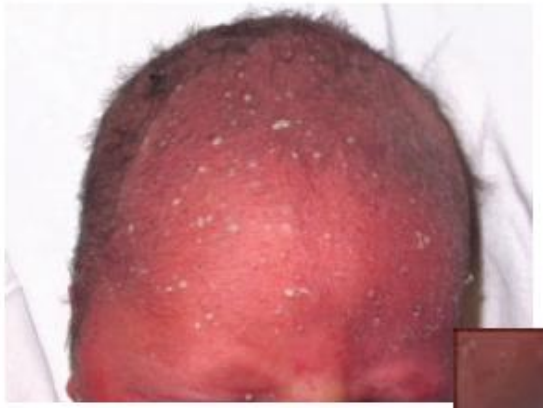


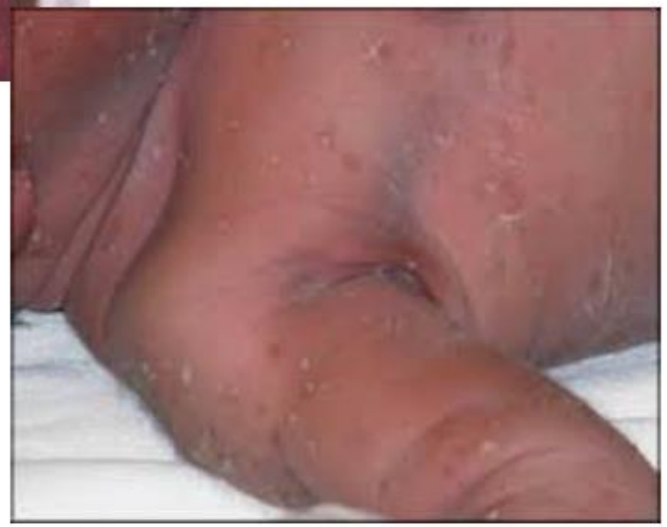
PUSTULES& vesicles THAT RESOLVE WITHIN 48 HOURS THEN MACULES THAT FADE WITHIN 3-4 WEEKS MAY PERSIST FOR MONTHS,COMMON IN TERM-GESTATION INFANTS AND IN AFRICANS

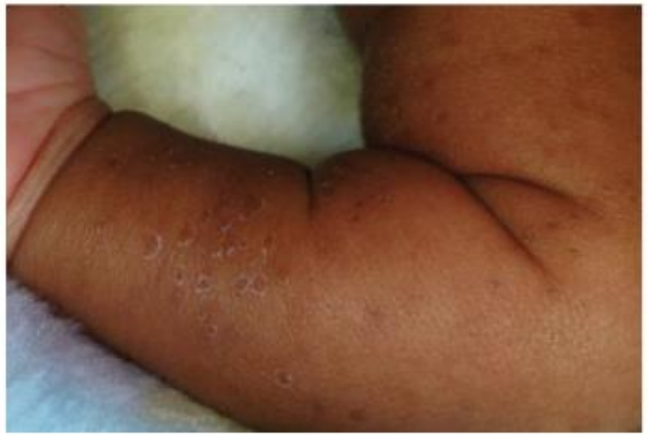
Transient Neonatal Pustular Melanosis

The vesicles and pustules usually resolve within 48 hours

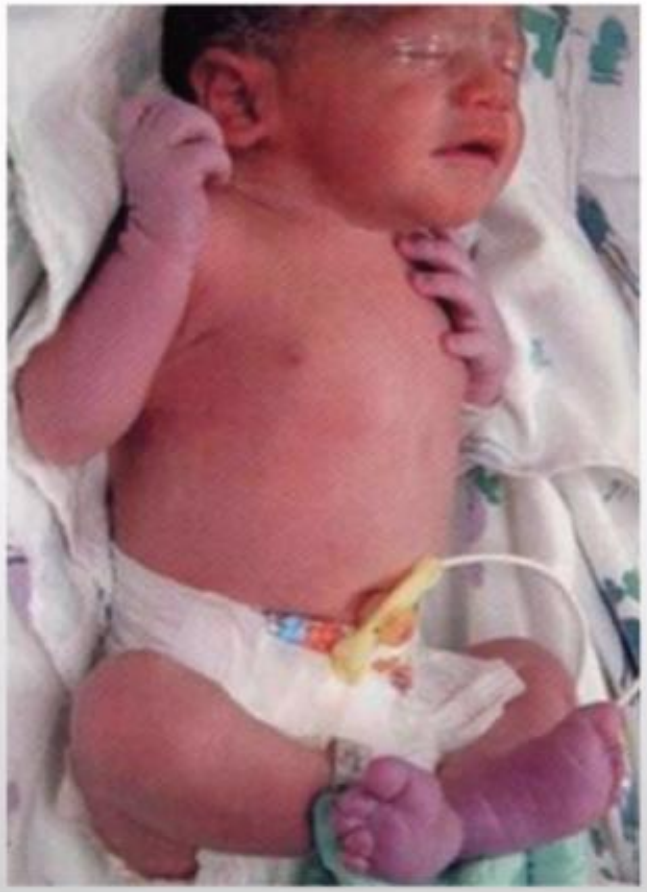
brown macules usually fade over 3-4 weeks but may persist for several months





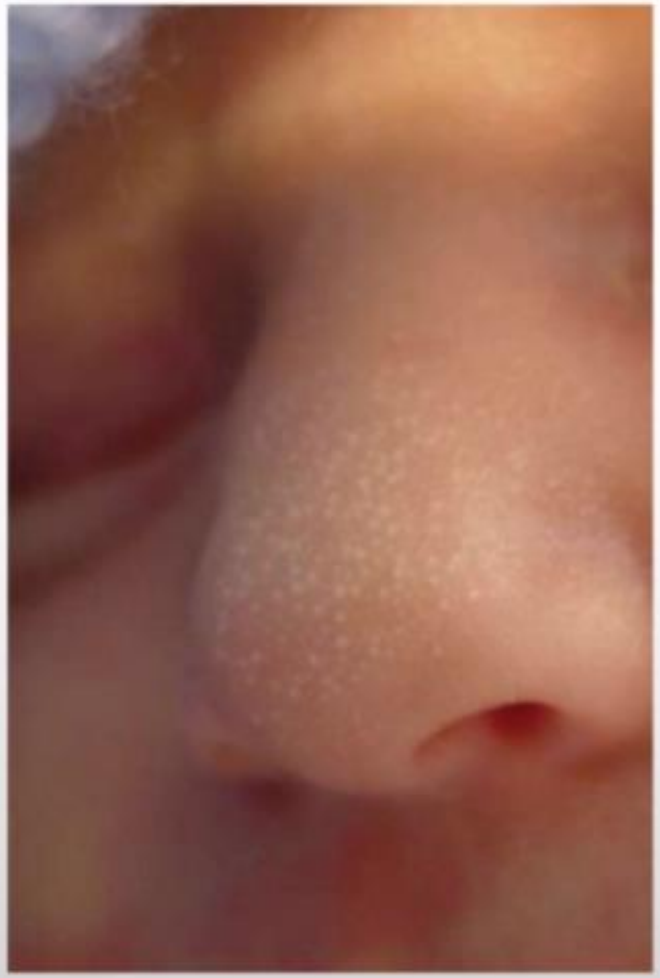


ACROCYANOSIS (PERIPHERAL CYANOSIS)



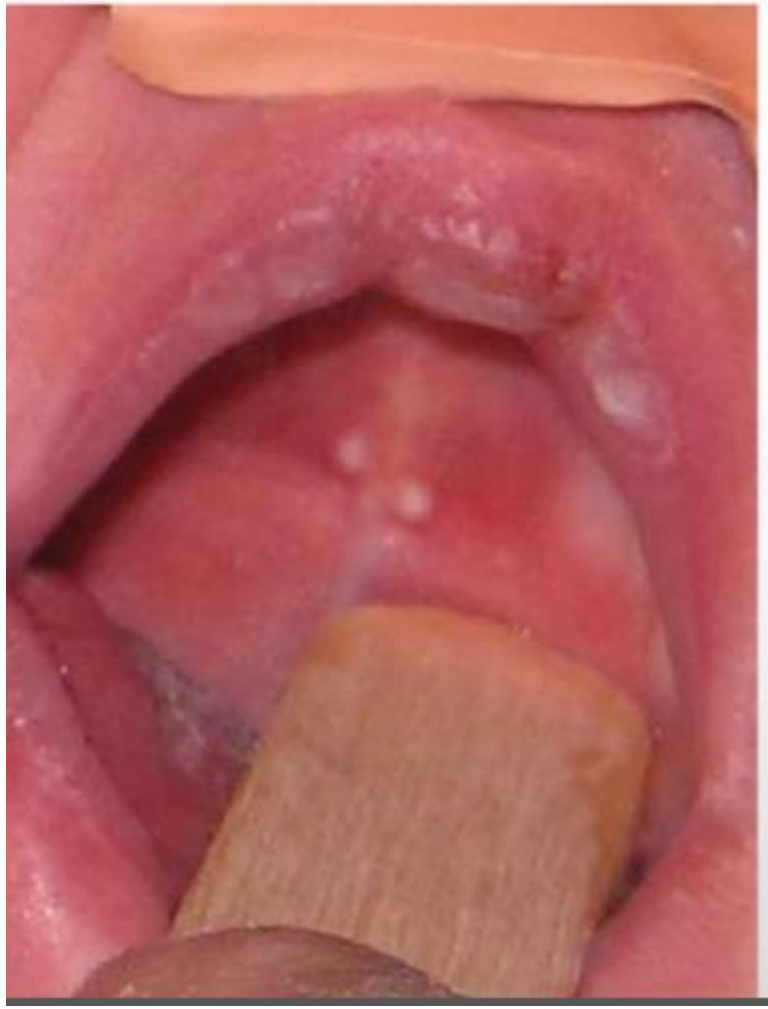
- Functional peripheral vascular disorder characterized by bluish discoloration of skin
- Caused by vasospasm of the small vessels of the skin in response to cold
- Usually particularly marked on the palms, soles and around the mouth
- Absence of cyanosis of warm central parts
- Resolves with warming of the skin
- Recurrence unusual after 1 month of age

SEBACEOUS GLAND HYPERPLASIA



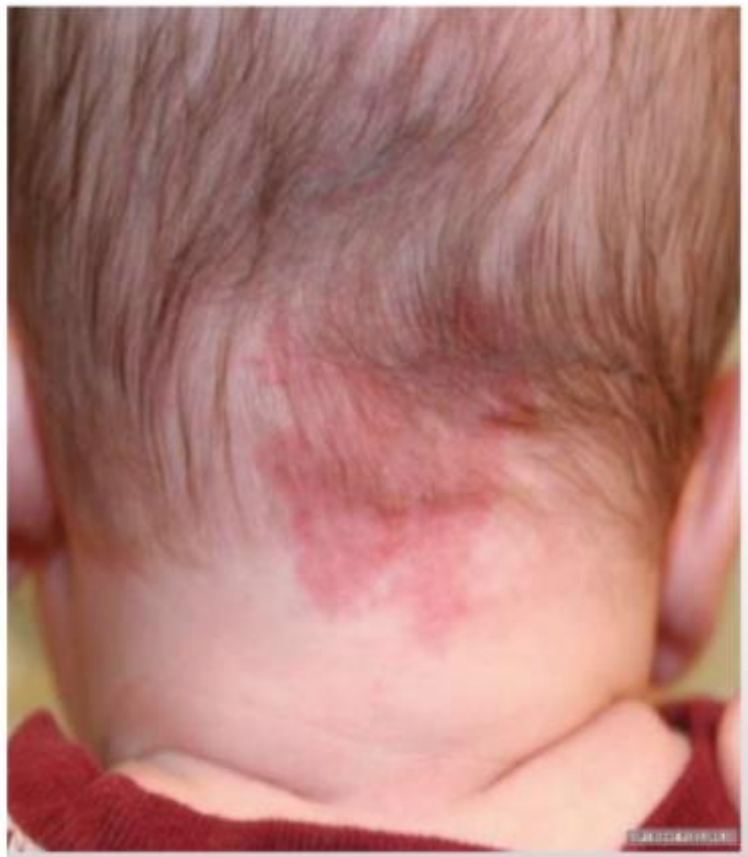
- Common benign proliferation of the sebaceous glands seen during the first weeks of life
- Result from maternal androgenic stimulation of sebaceous gland
- Multiple, uniform, pinpoint, yellowish papules 1–3 mm in diameter most prominent on the nose, cheeks, upper lip and forehead
- **Treatment** - Resolves within few weeks

EPSTEIN PEARLS



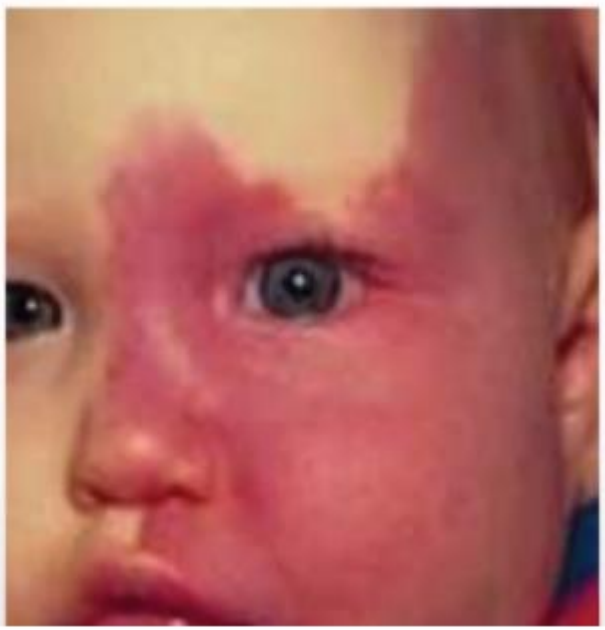
- Yellowish white, keratinous cysts, 1–2 mm diameter,
- Seen in up to 85% of all neonates
- Site - Along the alveolar ridges and/or in the midline at the junction of the hard and soft palate
- **Treatment** - Generally disappear without treatment within a few weeks

SALMON PATCH (NEVUS SIMPLEX)



- Most common vascular birthmark of infancy. Seen in 40% of all newborns
- Cause: Area of superficially dilated capillaries
- Appears as irregular dull, pinkish – red macules with poorly defined borders
- Site - On the face (angel kiss), nape of neck (stork bite)
- Become more intense in colour when child is crying
- Most of these lesions spontaneously disappear

PORT WINE STAIN (NAEVUS FLAMMEUS)

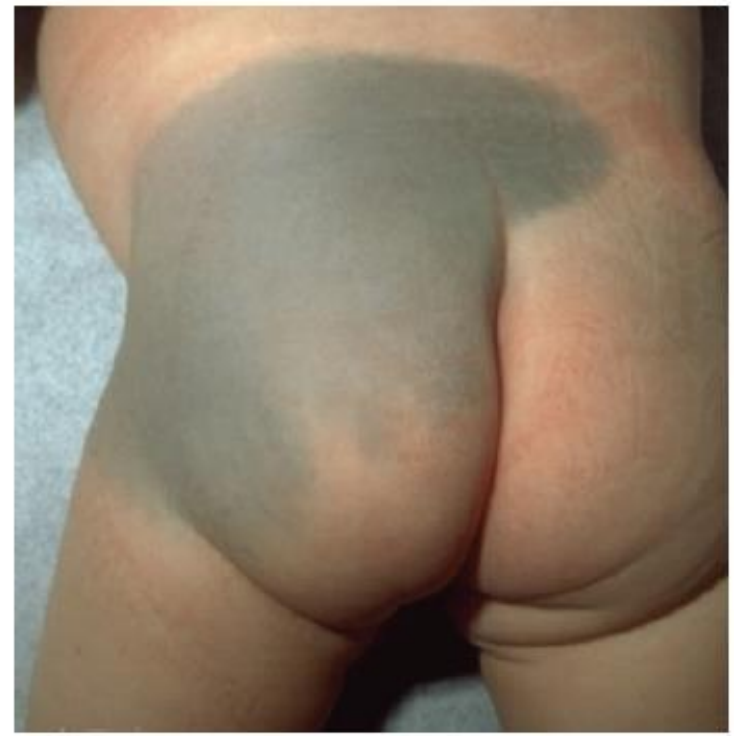


- Vascular birthmark, about 0.3% of newborns
- Large, irregular, deep red or purple macule with wel defined borders
- Usually unilateral, often on the face.
- Represents a vascular malformation involving mature capillaries.
- Lesions do not enlarge but persist throughout life

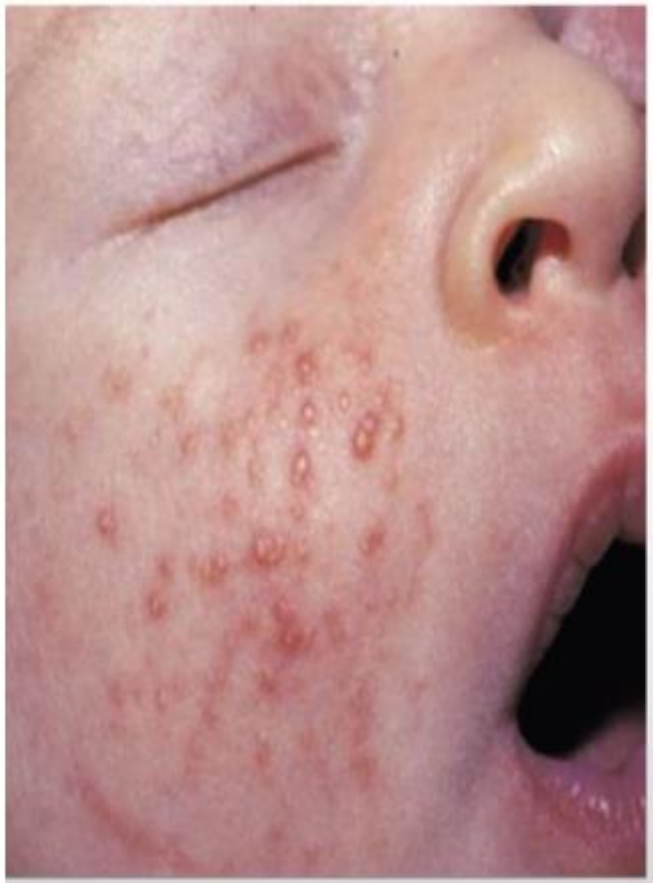
Abnormal neonatal skin disorder

MONGOLIAN SPOTS

- Blue-gray, poorly circumscribed, single or multiple, macular lesion of various sizes
- Entrapment of melanocytes in dermis of developing embryo, the cells fail to reach their proper location in the epidermis
- Usually present at birth or appears within the first weeks of life
- Most commonly over lumbosacral region
- Common in asian, black and hispanic infants
- Most fade during first two years of life



NEONATAL ACNE



- Prepubertal acne can be divided into five subgroups: neonatal, infantile, midchildhood, preadolescent and adolescent
- Thought to be due to androgens (maternal & infant)
- May affect up to 20% of neonates, more common in boys
- Presents at or shortly after birth with erythematous papulopustular lesions, *and comedones*
- Site - commonly on cheeks, chin and forehead

The well new-born

- Vitamin K given to all babies after delivery in an intramuscular Injection has been shown to prevent both early and late forms of bleeding.
- All babies are born with low levels of vitamin K because of low levels of transplacental passage,breast milk is a poor source of vitamin K,synthesized by gut flora and they lack gut flora,their liver cannot store vitamin K.
- Vitamin-K deficient bleeding formerly known as the hemorrhagic disease of newborn present as;skin bruising, mucosal bleeding, bleeding at the umbilicus and circumcision site, or even fatal intracranial hemorrhage.
- In general, weight loss of >10% to 12% in the first postnatal week is a cause for concern and necessitates a thorough evaluation.
- It is typically taught that newborns should regain their birth weight by 2 weeks after the birth, although many newborns reach this value much sooner if feeding is well established.
- Weight loss of 10-12% of birth weight is common in first week of life because of skin losses and breast milk is still colustum.
- Average weight gain after the first week of life is 20-30g/day
- They double their birth weight by 4 months and triple by 1 year
- Umbilical cord stamp falls off at 10-14 days

Table 1. **The Apgar Score**

The Apgar Score	0	1	2
Heart rate	Absent	<100 beats per min	>100 beats per min
Respiratory effort	Absent	Weak cry; hypoventilation	Good cry
Muscle tone	Flaccid	Some flexion	Active motion/Well flexed
Reflex irritability	No response	Grimace	Cry/Cough/Sneeze
Color	Blue/Pale	Acrocyanotic	Completely pink

APGAR
APPEARANCE: PALE OR BLUE/ACROCYANOSIS/PINK
PULSE:ABSENT/<100/>100 beats/min
GRIMACE:ABSENT/GRIMACE/CRY OR COUGH OR SNEEZE
ACTIVITY: FLACCID/SOME FLEXION/WELL FLEXED
RESPIRATION:ABSENT /WEAK CRY/GOOD CRY

- A 5-minute Apgar score of 7 to 10 is considered normal.
- Apgar scores can be helpful in assessing an infant's transition from intrauterine to extrauterine life
- It may reflect neonatal resuscitation efforts
- It t should not guide these resuscitation efforts.
- Apgar scores should not be used to predict neurologic outcomes or development of infants

- You reviewed the bay delivery Notes. His birth weight was 3 Kg, with no maternal Illnesses and his fetal record was normal.

Looking at Abgar score in the newborn medical record you found that it was 7 at one minutes

At 5 minutes of life the description of Abgar score was:

- Pulse rate was 130bpm,
- Breathing is well and has good cry
- Cyanotic Hands and feet
- Good Muscle tone
- and strong grimace

Q5. How you calculate Abgar score?

Q6 . Calculate his Abgar score at 5 min ?

Table 1. The Apgar Score

The Apgar Score	0	1	2
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Color	Blue/Pale	Acrocyanotic	Completely pink

Stooling pattern&urination

- **Meconium**
  - The infant typically passes a first meconium stool shortly after birth, often within the first hours and typically before 24 to 48 hours
  - These **black, tarry, and sticky stools**
  - **Most neonates pass meconium in the first 24 hrs**
- **Transition Stool**
  - Occurs as the mother's human milk production increases.
  - typically occurs in a pattern, often from **green/brown to a seedy, loose, mustard yellow appearance.**
- The infant's first urination nearly always occurs in the first 24 hours.
- Hard to detect because it cannot be seen,or because of frequent meconium passage.
- How to detect;review the records or look at the diaper with a strip or use a cotton ball or catheterization.
- scant and darkly colored,Can be "brick dust" this is urate crystals (often termed "brick dust") confused with blood in urine,completely benign.

Jaundice apparent if TSB>5 mg/dl,(1.2 milligrams per deciliter (mg/dL)

- Severe neonatal hyperbilirubinemia ; Defined as a Total serum Bilirubin>25 mg/dL in Term Newborns .
- Bilirubin-Induced Neurologic Dysfunction (BIND) -Acute signs = Acute Bilirubin Encephalopathy (ABE) include:poor feeding, lethargy, hypertonia.
- Kernicterus is the chronic and permanent sequelae of BIND.
- How to know bilirubin value,total serum bilirubin or transcutaneous bilirubinometer (unreliable above 14 mg/dl).
- Risk factors for exaggerated jaundice;cephalohematoma or bruising,male gender,breast-feeding,low caloric intake,dehydration and weight loss.
- Breast-feeding jaundice or lactation failure jaundice happens because of inadequate feeding and insufficient caloric&fluid intake,affected babies are dehydrated and have weight loss causes include maternal causes,cracked nipples or difficult sucking/first week of life.(colostrum milk)
- Breast-milk jaundice during the second week of life,adequate feeding, The exact cause of breast milk jaundice isn't known. However, it may be linked to a substance in the breast milk that prevents certain proteins in the infant's liver from breaking down bilirubin

Neonatal jaundice

Pathologic jaundice	Physiologic jaundice
• Clinical jaundice before **24 hours.** • Rise in serum bilirubin by more than **5mg/dl/day** • Serum bilirubin more than **15 mg/dl** • Clinical jaundice persisting beyond **14 days of life** • Clay/white colored stool and /or dark urine staining the cloths yellow. • Direct bilirubin **>2mg/dl** at any time.	• First appears after **24 hours** of age. • Does not exceed **15 mg/dl** • Clinically undetectable after **14 days** • Maximum intensity seen **4-5 day in term and 7th day in preterm**.

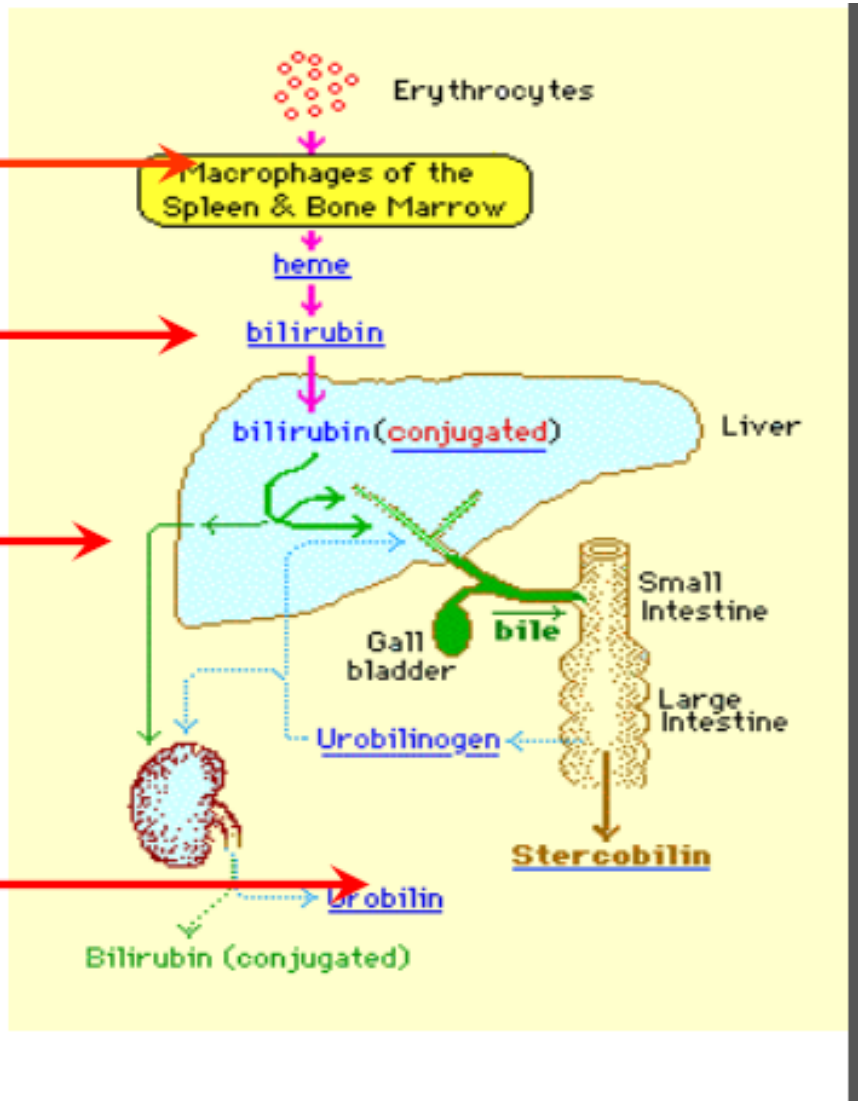
Why physiological jaundice happens

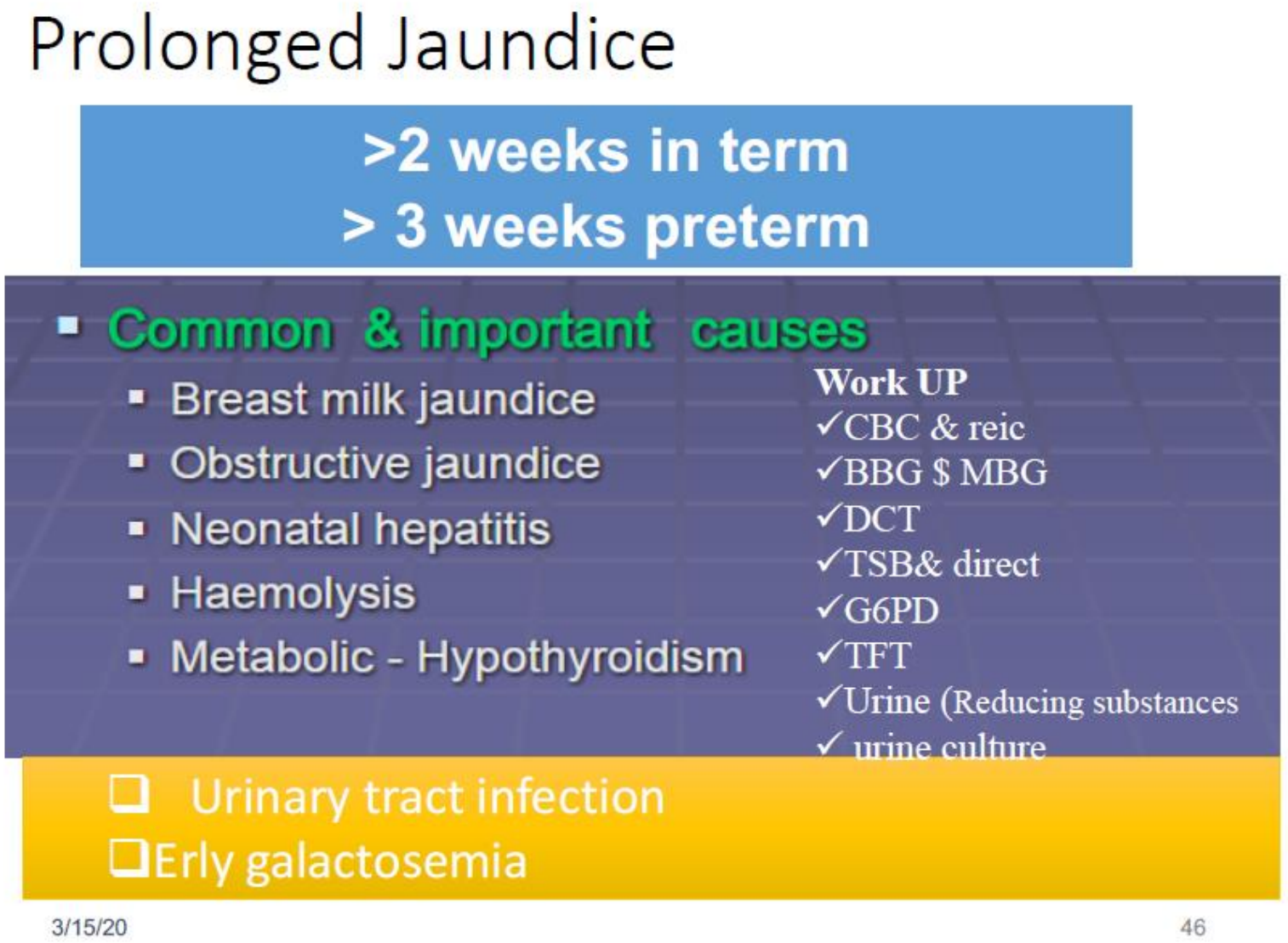
Increased rbc's
and ineffective erythropoiesis

Shortened RBC lifespan

Immature hepatic uptake &
conjugation (paucity of ligandin
and decrease UGTA1)

Increased enterohepatic
Circulation
(Increase B glucuronidase)





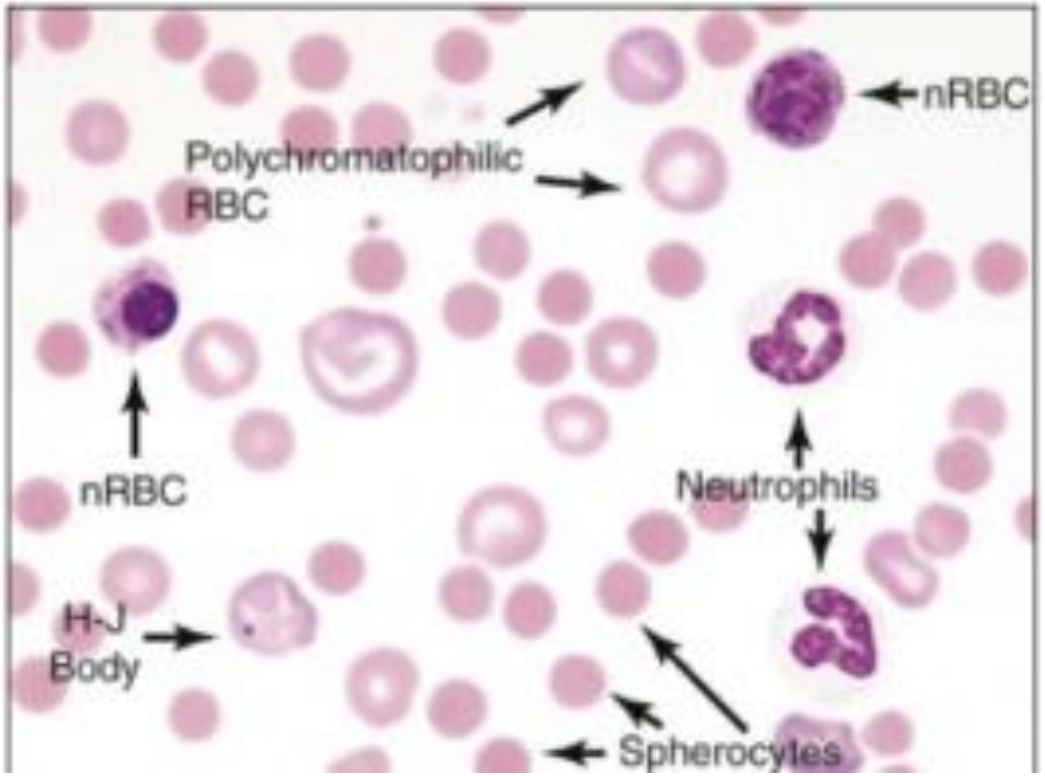
- The **direct Coombs test** is used to **test** for autoimmune hemolytic anemia.(ABO incompatibility)

Causes of pathological jaundice

Hemolysis	Increased RBCs	Sepsis	Galactosemia
*hemoglobinopathies,immune-mediated;Rh or ABO incompatability or minor blood group incompatibility. *inherited membranopathies such as hereditary spherocytosis. *enzymopathies;G6PD	*polycythemia *cephalohematoma *ineffective erythropoiesis.		

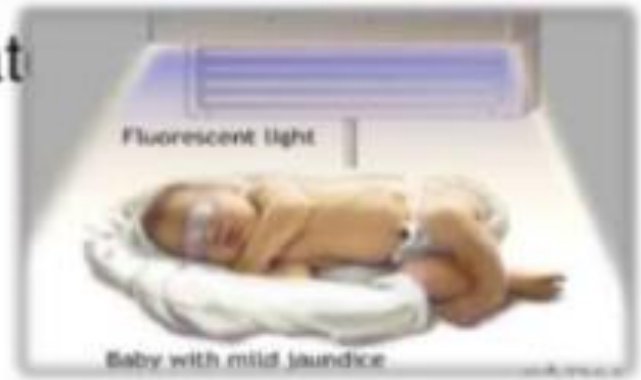
ABO Incompatibility

- Early onset jaundice – within 24 hour after birth
- Baby blood group A or B, Mother blood group O
- Direct Coomb's test +ve
- Blood smear show increase spherocytes
- Usually can be controlled with phototherapy



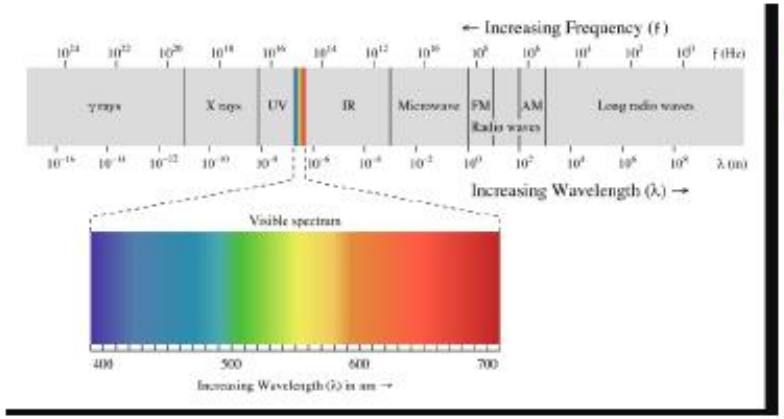
Therapeutic Options

- Phototherapy for neonat
with mild jaundice



- Exchange transfusion i
Severe cases

Intravenous Immune globulin



Phototherapy

- Goal: to treating neonatal hyperbilirubinemia and prevent related neurotoxicity
- Decreases the need for exchange transfusion
- Exposure of the skin of the jaundiced baby to blue or cool white light of wavelength 425-475 nm
- Toxic bilirubin molecule isomerizes to non-toxic product

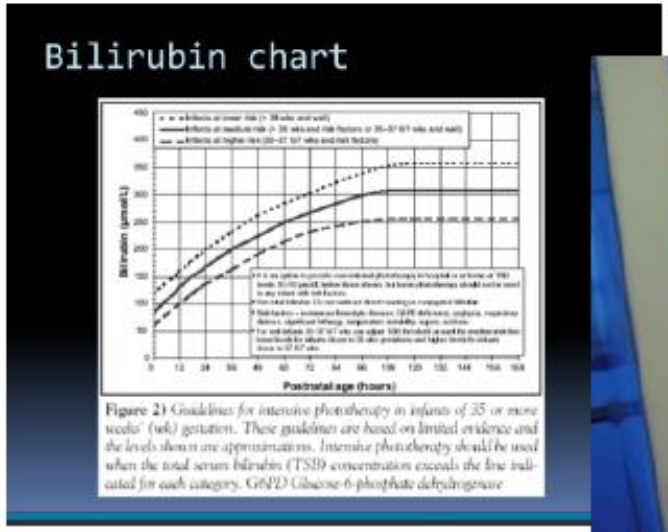


Figure 2) *Guidelines for intensive phototherapy in infants of 35 or more weeks' (wk) gestation. These guidelines are based on limited evidence and the levels shown are approximations. Intensive phototherapy should be used when the total serum bilirubin (TSB) concentration exceeds the line indicated for each category. G6PD Glucose-6-phosphate dehydrogenase*

Indications for exchange transfusion

- bilirubin levels >25 mg/dL, severe hyperbilirubinemia.
- those who are not responding to phototherapy.
- those with evidence of acute bilirubin encephalopathy.(BIND)

IVIGS/Indications:

- infants with **isoimmune hemolytic** disease
- **TSB level is rising despite phototherapy(before trying transfusion)**
- or is within 2 or 3 mg/dL of the threshold for exchange transfusion

❖NOTE; There are two types of **Coombs tests**. The **direct test** looks for antibodies that are stuck to red blood cells. The **indirect test** looks for antibodies floating **in the** liquid part of your blood, called serum.

❖Direct coombs test is used to test for hemolytic anemias(autoimmune),indirect is used to detect any abs in the serum,usually mother&infant blood group incomaptibility

❖Normal retic count for a

1 day	3.0-7.0 %

Hemolytic disease of newborn

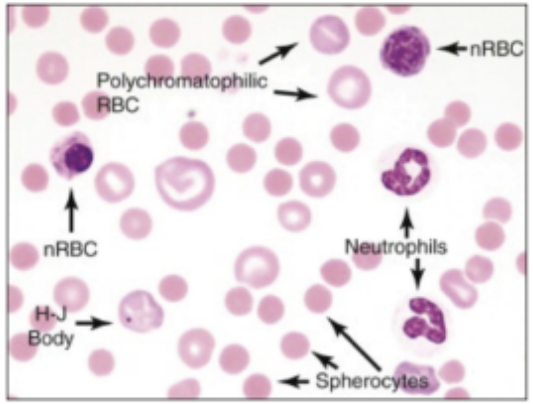
Isoimmune hemolytic anemia

- Is diagnosed by
- a positive direct antiglobulin test result
- low hemoglobin level
- high reticulocyte count
- signs of hemolysis on peripheral smear
- and indirect hyperbilirubinemia

Investigations

What other test

- Blood film



1. Spherocytes
2. Increase number of nucleated RBC
3. Polychromasia
   - Abnormally high number of immature red blood **cells** found in the bloodstream as a result of being prematurely released from the bone marrow during blood formation)

Neonatal GI emergencies

- Meconium passage is only affected by passage of meconium in utero,who passed in utero pass meconium earlier.
- Term babies pass meconium earlier than preterm babies.
- Breast milk,yellow stool,large and more frequent stool during the 1st week then decreases in the 3rd week.
- Formula feed babies have a persistent green stool.

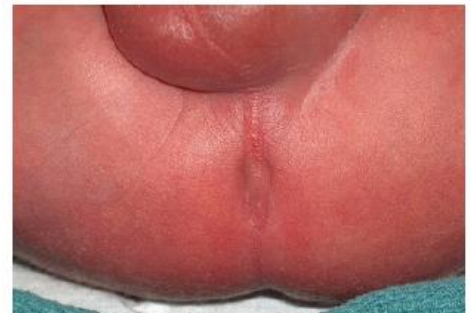
Case 1: Why Prenatal Fetal US is important

Possible findings	Diagnosis
- Polyhydramnios and no visualization of normally visible fluid-filled structures, such as the stomach or dilation of structures, such as the stomach and duodenum	• Esophageal atresia
. - Fetal Double-bubble sign	• Duodenal atresia
- Bowel wall thickness greater than 3 mm(echogenic bowel)	• Bowel obstruction
- Intraluminal and/or abdominal calcifications	• Meconium peritonitis and other causes
- other abnormalities	. VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association

Differential diagnosis for delayed meconium passage.

- Ano rectal malformation
- Meconium ileus
- Meconium plug
- HD
- Left small colon
- Hypothyroidism
- Acute Colonic Pseudoobstruction (Acute Megacolon, Ogilvie Syndrome

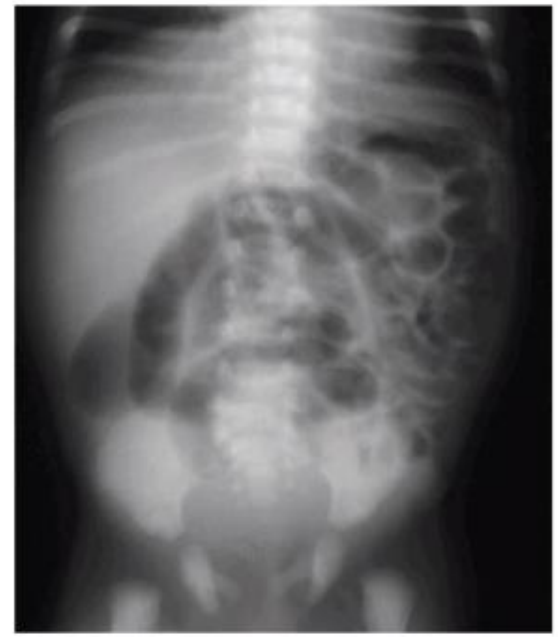
- Ano rectal malformation



Meconium Plug syndrome

- functional colonic obstruction
- is a transient disorder of the newborn colon
- characterized by delayed passage (>24-48 hr) of meconium and intestinal dilatation.
- The incidence is one in 1,000 births
- May be it is associated with Hirschsprung disease in 40% of cases and cystic fibrosis in 40% of cases.

[https://emedicine.medscape.com/article/410969-overview]

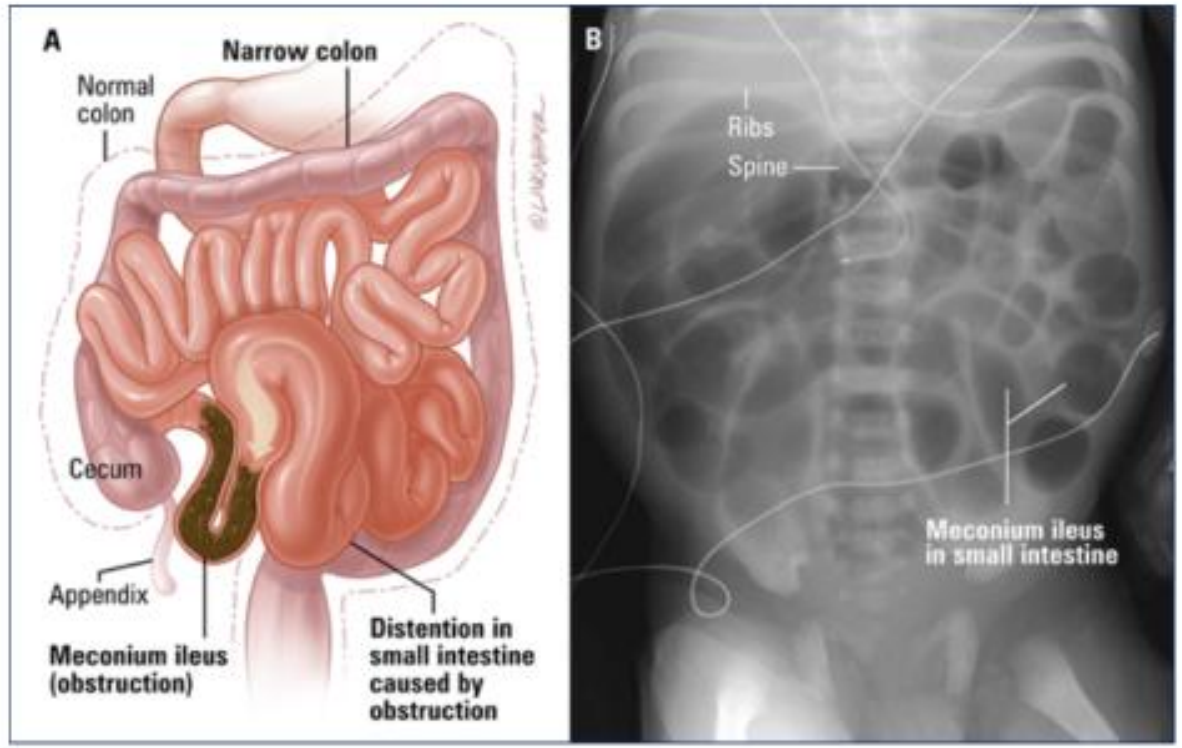


Supine frontal view of the abdomen in a newborn with meconium plug syndrome demonstrates multiple dilated loops of bowel but no rectal gas.

View Media Gallery

Case 1: What are the DDX

- Meconium ileus (MI)
  - meconium is thicker and stickier than normal.
- The most common cause is cystic fibrosis
- Around 20% of CF present with (MI)
- Most often at level of ileocecal valve in neonates
- Complication
  - Intestinal atresia
  - Volvulus
  - Meconium peritonitis
  - Perforation

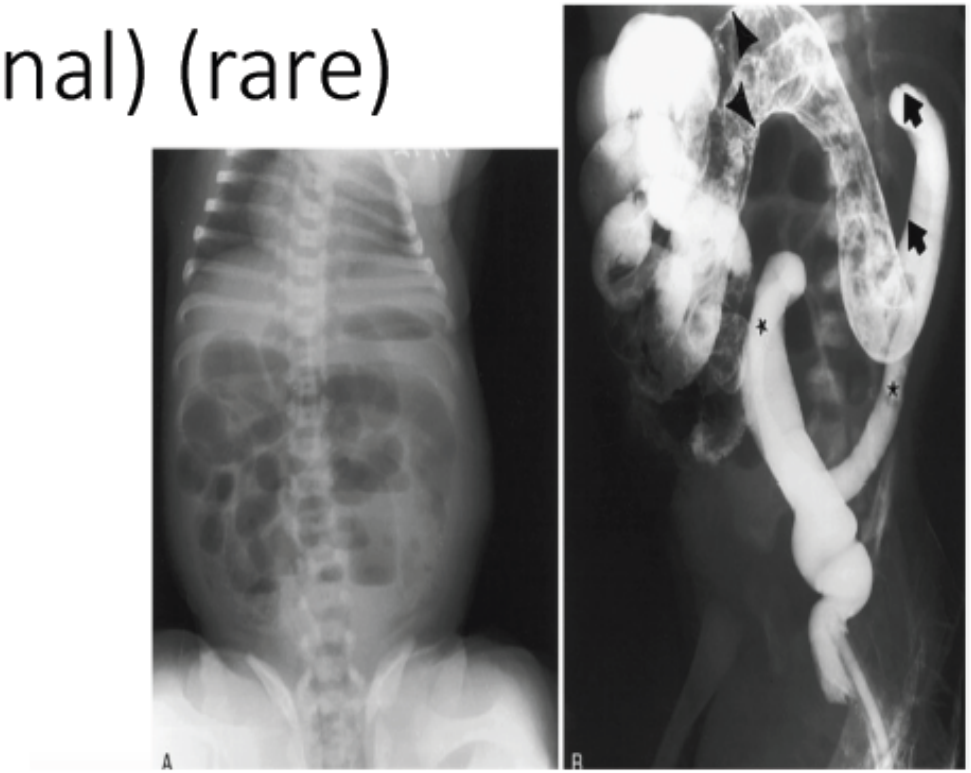


A. Illustration of intestine blocked by meconium. B. Abdominal x-ray of a newborn infant with meconium ileus showing dilated loops of bowel.

Other DDX
Left small colon (dysfunctional) (rare)

- transient and resolves 24 to 48 hours after birth
- History of maternal diabetes (40%)
- association with hypothyroidism, hypermagnesaemia
- Asses maternal use of psychotic medication that Dysmotility in the descending colon

The obstruction is typically partial and involves the descending colon distal to the splenic flexure.

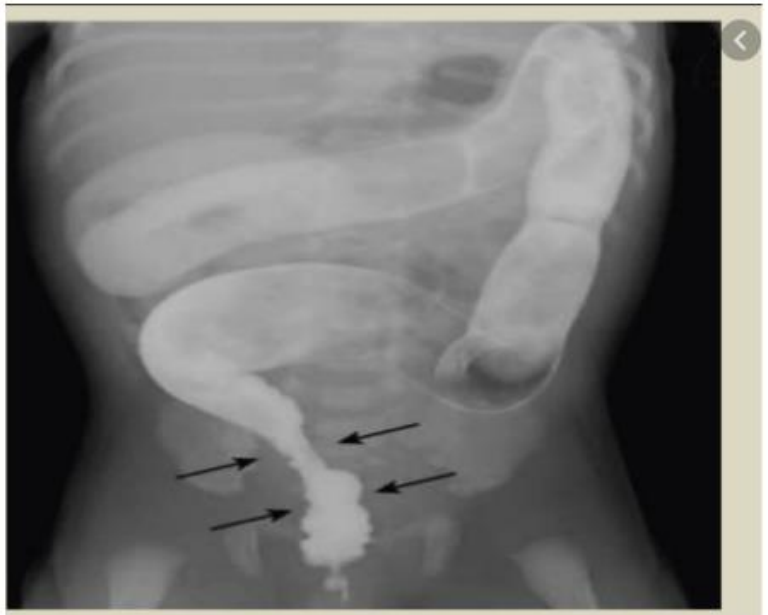


Barium enema radiograph shows typical findings of the small left colon syndrome. Note the distal microcolon (*asterisks*), the splenic flexure transition (*arrows*), and the dilated transverse colon proximal to the point of obstruction (*arrowheads*).

Heirschpring disease-diagnostic study barium enema show sawtooth appearance

What Is the next Diagnostic studies

- Contrast enema



rung disease in an infant. Frontal radiograph demonstrates the diameter of t rectum (arrows) to be smaller than the diameter of the sigmoid colon, an abnormal rectosigmoid ratio. Note the saw tooth appearance of the abnormal contracted segm

- Gold-standard for diagnosis is rectal biopsy we can start with suction biopsy if the results turned to be equivocal we do full-thickness biopsy
- Multiple hypertonic contrast enema to treat meconium plug or meconium ileus.
- Surgical resection is the definitive treatment for HD

Key questions in the history of a patient with delayed meconium passage

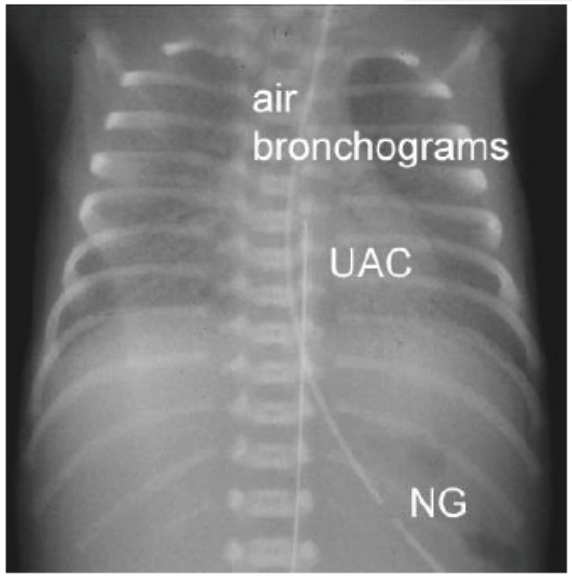
- Associated symptoms such as vomiting,distended abdomen,jaundice.
- Constitutional symptoms such as decreased feeding,poor sucking,fever.
- Family history of HD,CF,duodenal or jejunal atresia
- Pre-natal U/S findings.
- Shock symptoms such as pallor.

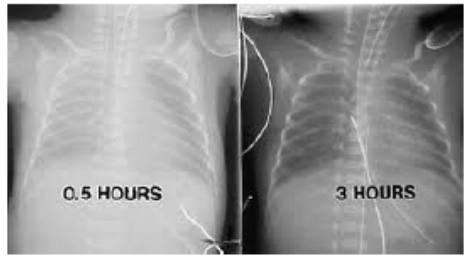
Complications of prematurity

- Respiratory management of RDS
- 1- **SURFACTANT**
- 2- Respiratory support:
- - invasive CONVENTIONAL
- (MECHANICAL VENTILATION, HFO,)
- - None invasive (CPAP, NPPV, High flow
- NASAL CANNULA)

Types of Respiratory problems

1- Respiratory distress syndrome





Hyaline membrane disease

=

respiratory distress syndrome.

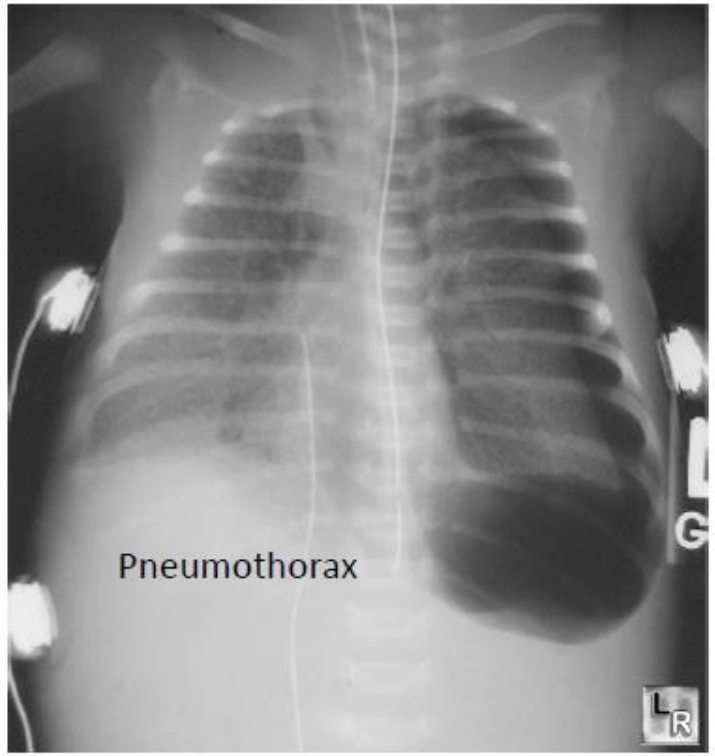
-

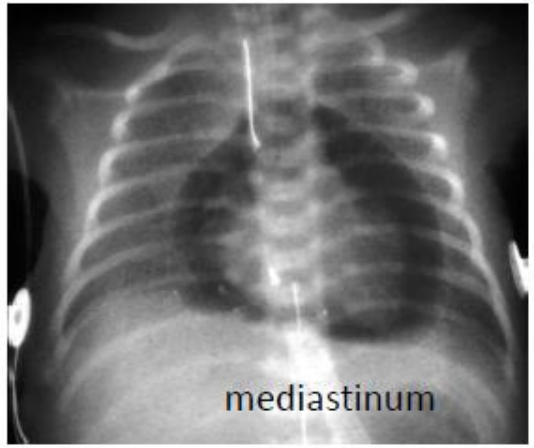
a condition in which the air sacs cannot stay open due to lack of surfactant in the lungs.

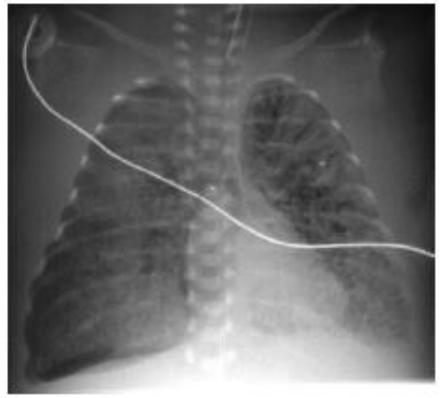
Resp complication of Respiratory distress syndrome

Air leaks

Air leaking out of thel lung spaces into other tissues







Pulmonary interstitial emphysema

- 2.PDA+3.infections(other complications of prematurity)

Complications of prematurity?

4 **- Metabolic problems**

- fluid loss through skin
  - (thin skin, no Keratin, Rapid Respiratory rate, from warmer and large Surface area)
- Have immature kidney
  - that cannot concentrate or regulate electrolytes and the buffer well)
- Na Imbalance
- Ca Imbalance
- K imbalance
- Risk of hypo and hyperglycemia

Complications of prematurity?

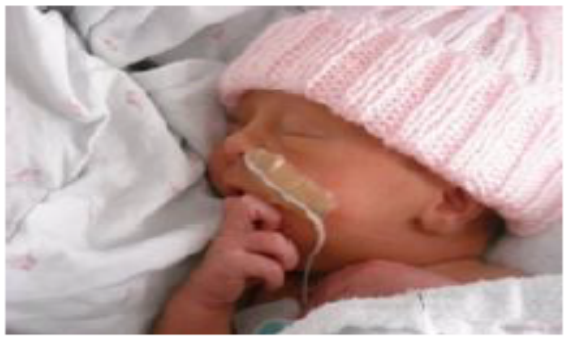
5-Skin care

Has: Fragile, thin transparent skin

9- Gastro intestinal problems

Feeding problems

- Difficulty in self feeding
- In coordination of sucking and swallowing
- Abdominal distension
- Regurgitation and aspiration

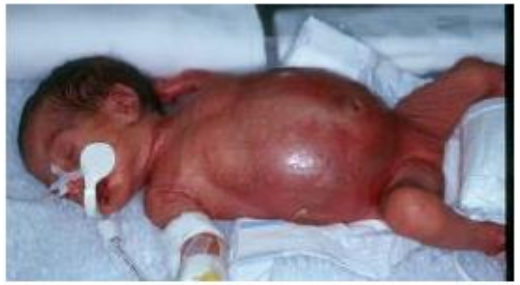


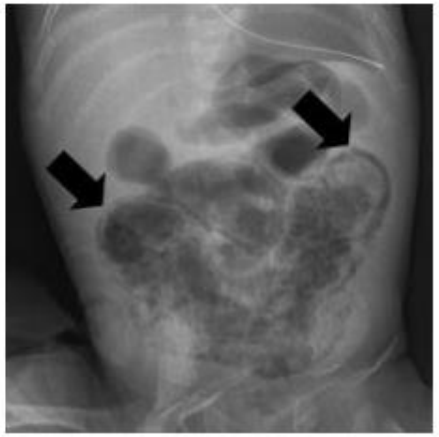
➢**UNABLE TO COORDINATE SUCK AND SWALLOW BEFORE 34 WEEKS GESTATION.**

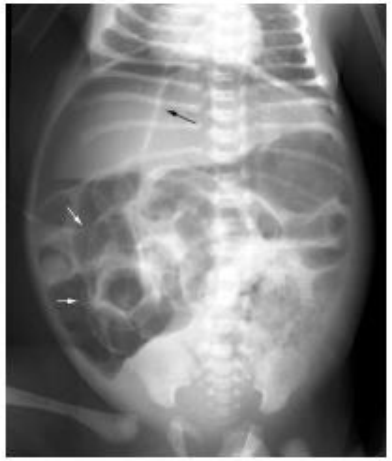
Necrotizing enterocolotis

GI problems

- NEC

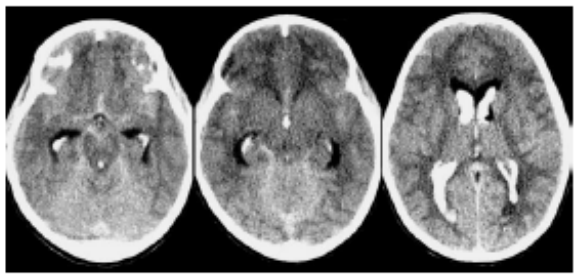






Neurologic complications :

– intrventricular hemorrhage IVH -.

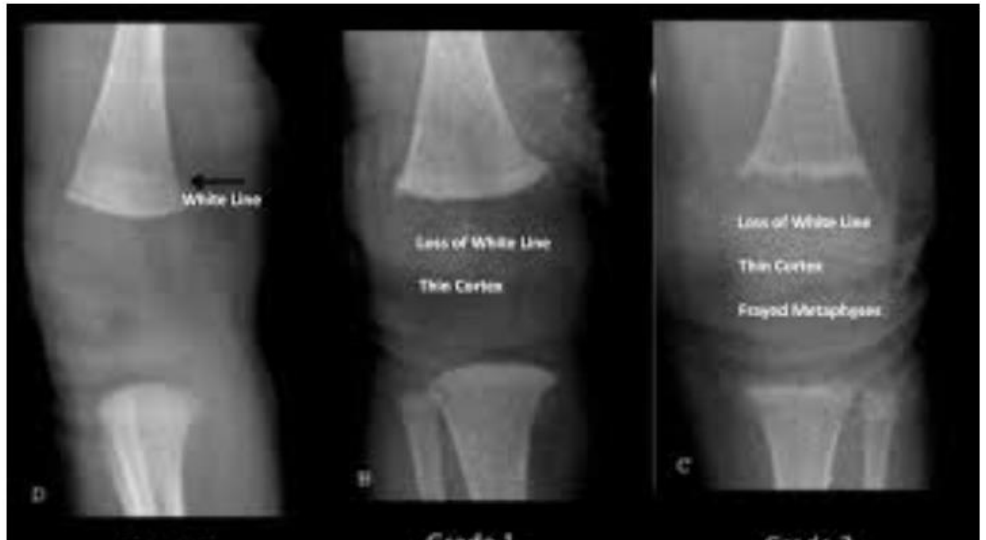


✓ Can lead to PHH :post hemorrhagic hydrocephalus

- ROP is retinopathy of

Prematurity

- Chronic lung disease is bronchopulmonary dysplasia.



Later Problem when the baby is stabilized

- ROP
- Anemia of prematurity
- Chronic lung disease
- Osteopenia of prematurity
- Infection
- Post hemorrhagic hydrocephalys
- PVL

ENDOCRINOLOGY

DKA

DKA – History:

- Polyuria , polydipsia, weight loss
- Abdominal pain
- Vomiting
- Confusion
- Tiredness
- Difficulty breathing

- Patients can come with hyperglycemia only or hyperglycemia and ketosis or hyperglycemia,ketosis and acidosis DKA.
- Progression may be accelerated by intercurrent illness or stress.

DKA

- Remember insulin is an anabolic protein in enhances anabolic processes in the body (lipid synthesis,protein synthesis,glycogen synthesis,and inhibits catabolic pathways such as lipolysis,glycogenlysis and gluconeogenesis.)
- If insulin is deficient lipolysis is stimulated by counter regulatory proteins as a defence mechanism causing release of Fas from adipocytes to the blood,they will be metabolized/oxidized into ketone bodies(acetone,acetoacetate,beta-hydroxybuteric acid which is the most abundant one)-this is the ketotis state caused by hyperglycemia and insulin deficiency(this causes polydipsia and polyuria and weight loss)
- This causes excess acid(which means more hydrogen ions this leads to binding with bicarb and bicarb consumption),the buffer system tries to compensate buy acid(co2) wash out by respiratory hyperventilation(this causes altered mental status,abdominal pain,nausea&vomiting,palpitations)

DKA – Clinical signs:

- Kussmaul breathing
- Lethargy
- Dehydration
- Signs of infection

- **Kussmaul breathing** is a deep and labored **breathin g** pattern often associated with severe metabolic acidosis.

Diagnosis of DKA:

- Glucose > 200 mg/dL
- pH < 7.3
- Ketonuria or ketonemia
- Serum Bicarbonate < 18 mmol/L

- Normal bicarb is 22-26 mmol/L.
- Base excess : It is defined as the amount of acid required to restore a litre of blood to its normal pH at a $PaCO_2$ of 40 mmHg. The **base excess** increases in metabolic alkalosis and decreases (or becomes more negative) in metabolic acidosis

DKA – Investigations:

- Capillary glucose STAT
- Venous blood – glucose, gases, electrolytes, urea, creatinine
- Ketones in urine or blood

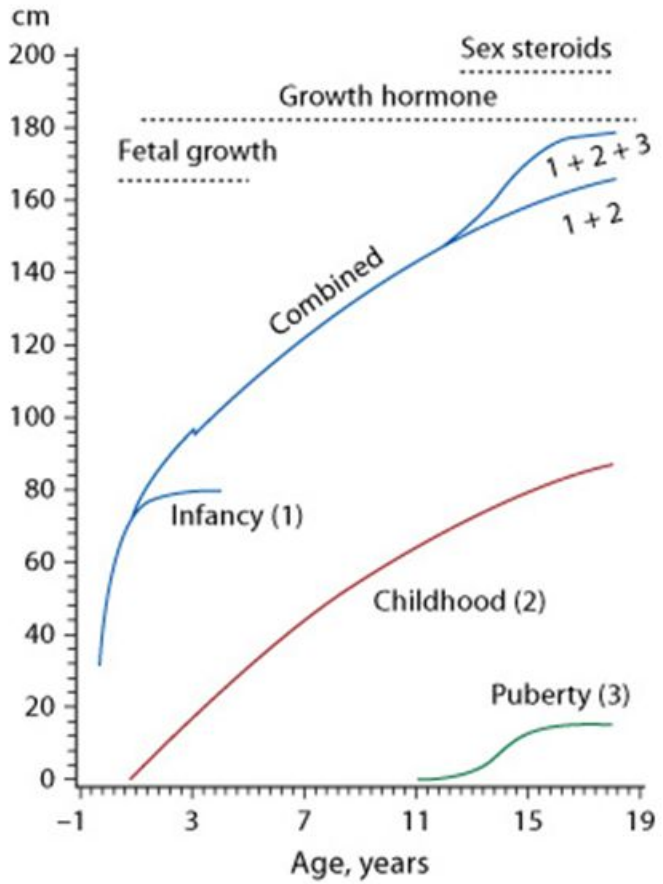
Management of DKA

1. IV fluid replacement by normal saline at a rate of 10 mL/kg .
2. Check levels of potassium(they could be falsely high(not indicating total body stores)but depleted intracellularly because of extracellular shifting by k-h pump to compensate acidosis or could be low because it was lost by urine by osmotic diuresis),so if it was low correct it then give insulin,if it was high give insulin and KCL together.
3. Insulin IV infusion at a rate of 0.1 units/kg/hr.
4. Admission to ICU and close monitoring of the fluid status,electrolytes,glucose,ABGs and acid-base status.
5. Know the triggering cause and treat it.

Short stature

University of Glasgow NHS Greater Glasgow and Clyde

ICP model of growth



In utero: Maternal/placental factors

Infancy: Nutrition

Childhood: Growth hormone

Puberty: Growth hormone + sex steroid

Height below 2 standard deviations from the mean. (3rd centile)

Causes of Short Stature

- Normal (genetic and constitutional delay)
- Small for gestational age
- Dysmorphic syndromes
- Skeletal dysplasias
- Chronic disease
- Endocrine
- Dire social circumstances!
- Idiopathic short stature

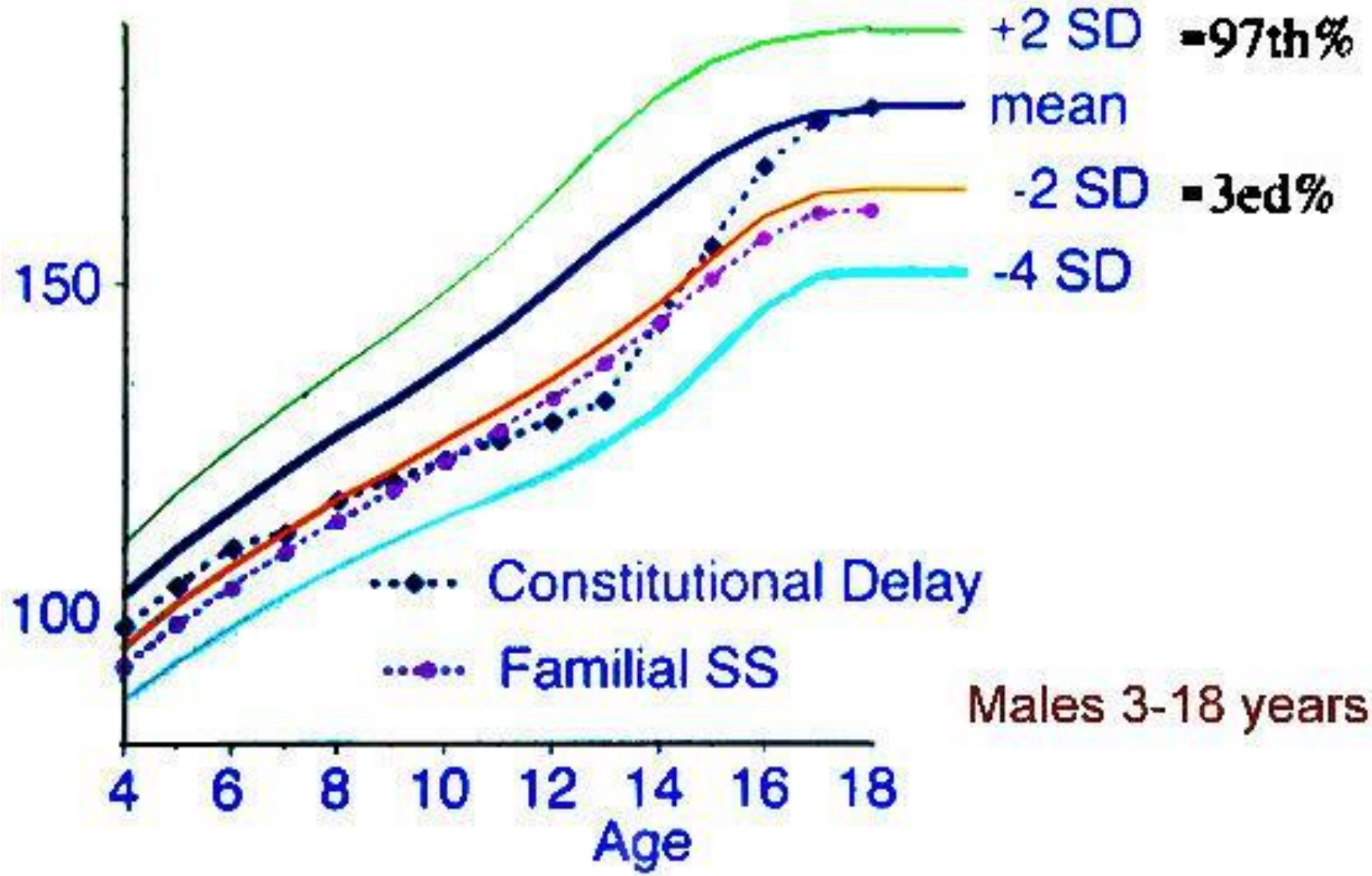
Normal genetic short stature

- Child short and normal
- Parent(s) short and normal
- Bone age not delayed
- Child destined to become short adult

when calculating the midparental height he is following the curve.

Constitutional delay

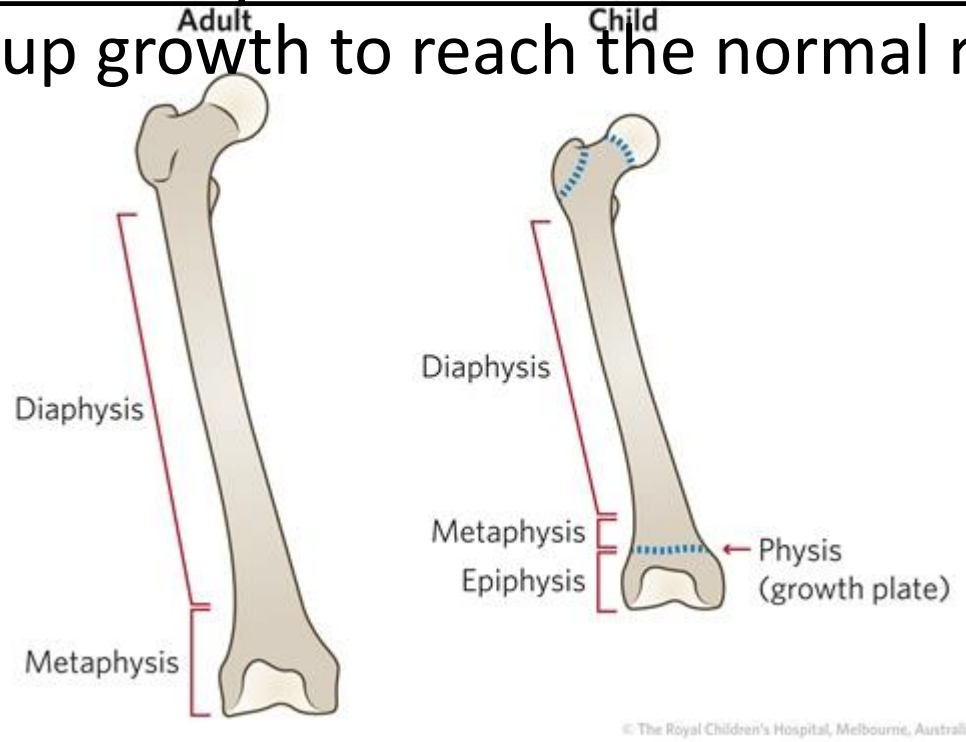
- Child short, normal but looks younger than chronological age.
- Parent(s) not short, but may have been so in childhood(positive family history of delayed bloomer)
- Bone age delayed
- Late puberty and catch-up growth
- Final height usually in lower half of target range


+2 SD
=97th%
mean
-2 SD
=3ed%
-4 SD
150
100
Constitutional Delay
Familial SS
Males 3-18 years
4
6
8
10
12
14
16
18
Age

Intrauterine growth retardation (IUGR)

- Asymmetric (HC>L>Wt)
  3rd trimester malnutrition; catch-up growth during infancy; normal childhood growth and final height.
- Symmetric (HC=L=Wt)
  prolonged intrauterine malnutrition; catch-up growth in infancy partial/absent; short stature in childhood despite normal growth rate; short adult.

- Most infants(90%)born small for gestational age (SGA) experience catch-up growth by two years of age.
- About 10 % of SGA infants, particularly those born with more severe SGA, do not experience catch-up growth to reach the normal range by two years of age.



Skeletal dysplasias

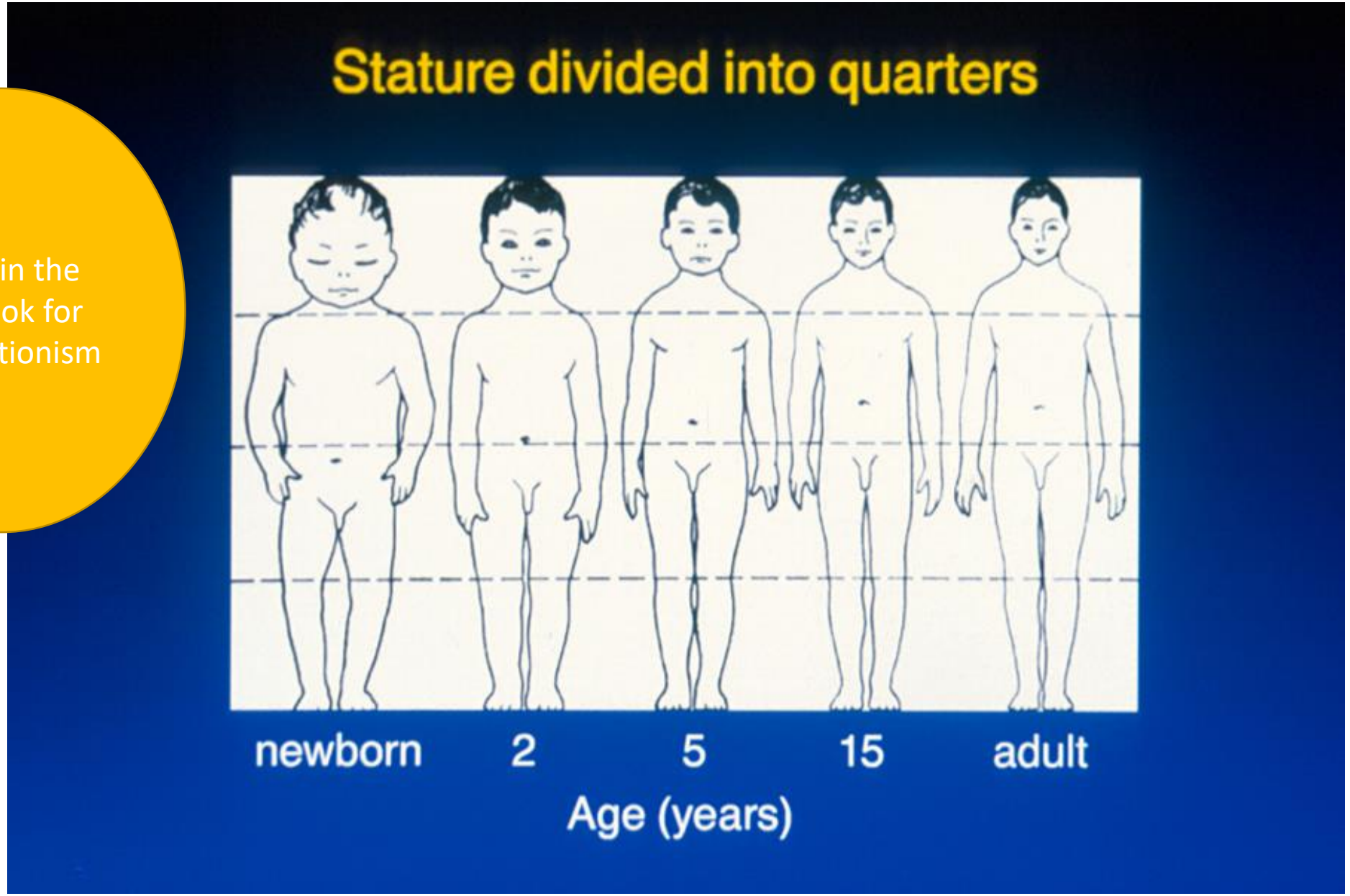
- What is **skeletal dysplasia**? **Skeletal dysplasia** describes a category of rare genetic disorders that affect bones and joints and hinder children's growth and development. The disorder causes abnormally shaped bones, especially in the head, spine and long bones of the arms and legs.

Skeletal dysplasia

- What is **skeletal dysplasia**? **Skeletal dysplasia** describes a category of rare genetic disorders that affect bones and joints and hinder children's growth and development. The disorder causes abnormally shaped bones, especially in the head, spine and long bones of the arms and legs.
- Frank skeletal abnormality affecting
  - epiphyses (epiphyseal dysplasia)
  - metaphyses (metaphyseal dysplasia)
  - spine (spondyleal dysplasia)

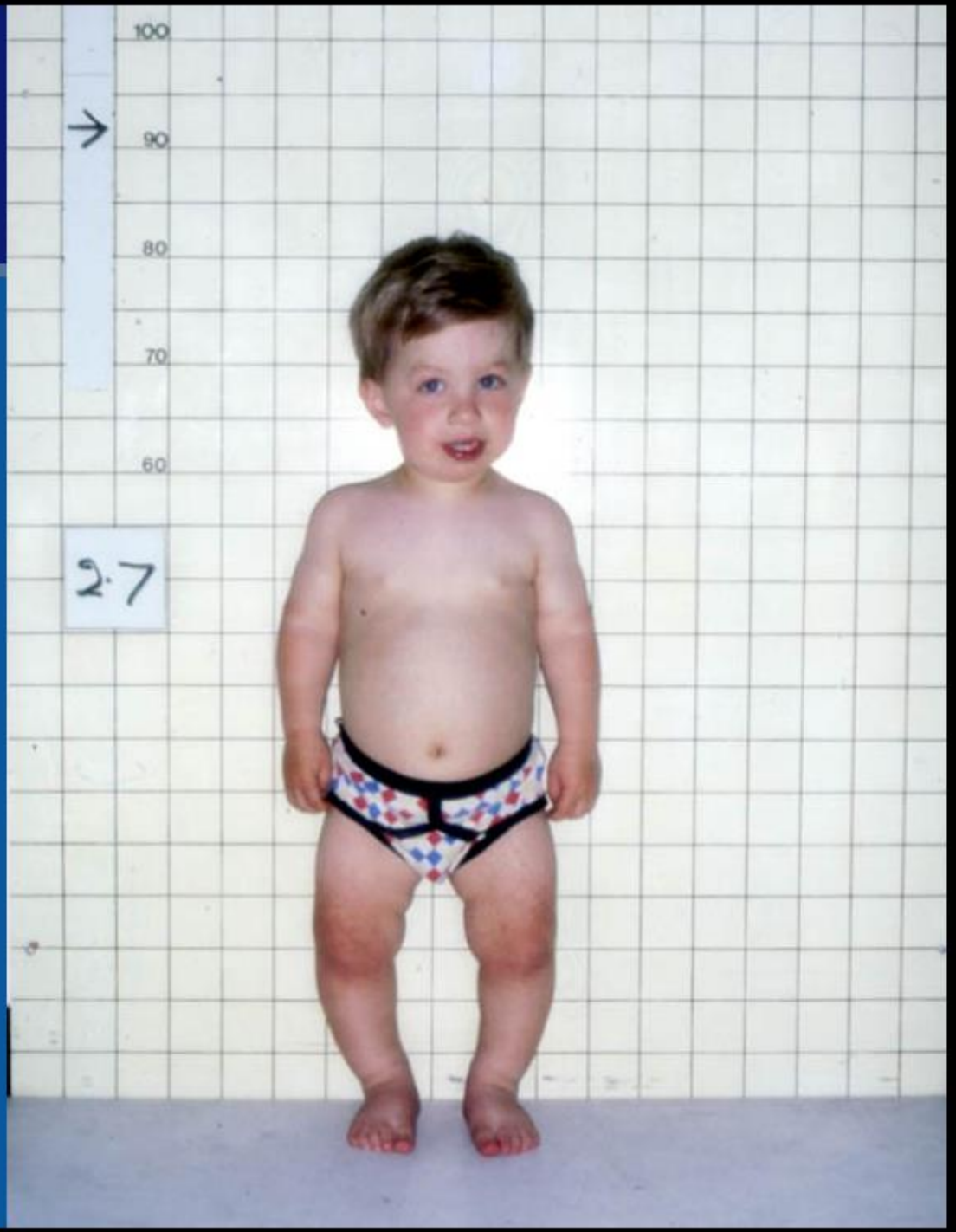
  in various combinations (e.g. spondylo-epiphyseal dysplasia)
- Bone age tends to be advanced.
- Adult height SDS < Childhood height SDS

HINT in the P/E;look for proportionism



Achondroplasia is a genetic disorder whose primary feature is dwarfism. In those with the condition, the arms and legs are short, while the torso is typically of normal length.

Achondroplasia



Chronic systemic disease

Many different types

Any chronic condition (e.g. GI, cardiac, respiratory, renal, metabolic, CNS, joints) can cause short stature and/or slow growth

Chronic renal disease and GI disorders (e.g. coeliac disease) can be silent, and present with short stature,that’s why we do KFT,LVT,CBC,COELIAC SCREEN.

Social and emotional short stature

- Children from poor communities tend to be shorter than those from affluent areas
- Severe short stature with growth failure may result from emotional abuse/neglect (psychosocial deprivation)

Endocrine disorders

- Growth hormone deficiency and resistance.
- Thyroxine deficiency
- Cortisol excess
- Precocious puberty
- Idiopathic short stature(a diagnosis of exclusion)

Classification of growth hormone deficiency

- anterior pituitary protein secreted by somatotrophs.
- Secretion is pulsatile.
- Stimulated be sleep and exercise, inhibited by free fatty acids.(thats why levels of GH is not beneficial to test for)

- cause (congenital or acquired)
- nature (true permanent deficiency or functional/temporary insufficiency)
- severity (complete, severe or partial)
- whether isolated or part of multiple anterior pituitary deficiency

Idiopathic short stature

- Definitions vary according to different centres
- One definition is significant short stature (< -2.5 SD-below the third percentile) not attributable to normal familial short stature or constitutional delay or to other causes of short stature .
- Heterogenous condition which may include partial GH resistance, mild skeletal dysplasias etc.

Approach to a patient with short stature

History:

- When was this first noticed? (congenital vs acquired causes)
- Previous measurements (growth velocity)
- Detailed review of systems(chronic diseases, brain tumors, irradiation)
- Birth hx (gestational age, weight & length) (if it was small for gestational age)
- Neonatal hypoglycemia, prolonged jaundice (signs of neonatal hypopituitarism)
- Nutritional hx
- Parents heights and puberty (genetic and or constitutional short stature).
- Headache , visual disturbances (central causes of GH deficiency)

On examination

- Reliable measurements of ht and wt , Proper PLOTTING
- Proportionism (skeletal dysplasias)
- Dysmorphism (syndromes vs GH deficient)
- Midline defects
- General exam to rule out signs of chronic diseases.
- Pubertal (Tanner)stage(precocious puberty)

Investigations

1. Screening of short stature ;CBC and blood film, KFT,LFT, free T4 TSH celiac screen TTG-IgA antibodies(tissue transglutaminase IgA) , consider blood gas, IGF1+BP3, prolactin, cortisol (to rule out central pituitary causes)

    - Bone age(delayed in constitutional delay,advanced in skeletal dysplasias and normal in genetic short stature)

2. If the screening tests do not yield a diagnosis and/ or we have a low IGF1 we proceed to :

Dynamic (stimulation) testing for GH.

Karyotyping if female.

MRI brain after diagnosing GH deficiency to look for a cause (e.g mass or congenital defects).

Conditions where growth hormone therapy is recommended

- Growth hormone deficiency
- Turner syndrome/Noonan Syndrome/SHOX
- Prader-Willi syndrome.
- SGA with no catch up by 4 years
- Chronic renal insufficiency.
- ISS(idiopathic short stature,controversial)

- Females ,stage 1 is childhood size,stage 2 is breast budding, stage 3 is aerolar enlargement,stage 4 is secondary mount and contouring,stage 5 is the mature adult size and shape.
- Males,stage 1 is childhood size,stage 2 is testes and scrotum beginning to grow,stage 3 penis begins to increase in size,stage 4 scrotum darkens both scrotum and penis continue to grow,stage 5 is the mature adult size.
- For both pubic hair;stage 1 is no pubic hair,stage 2 is long straight sparse hair,stage 3 is curly coarse and increase,stage 4 is continue to grow and darken,stage 5 extends to mid-thigh(mature adult).

I
II
III
IV
V
3
4
10
16
25
<2,5
2,5-3,2
3,6
4,1-4,5
>4,5

I
II
III
IV
V

Precocious puberty

What is definition of precocious puberty?

Secondary sexual development more than 2.5 standard deviations earlier than the median or mean age. For female before the age of 8 years and males before age of 9 years.

- Causes are divided into gonado-tropin dependent,gonadotropin-independent.

Gonadotropin-dependent precocious puberty (GDPP) - Causes

- Idiopathic
- Central nervous system (CNS) tumors
- CNS infection
- Head trauma
- Iatrogenic
    - Radiation
    - Chemotherapy
    - Surgical

- CNS malformations such as hydrocephalus,arachnoid cyst.
- Genetic.

Gonadotropin-independent precocious puberty (GIPP)

- CAH
- Testosterone/estrogen-producing tumors
- Ovarian cysts
- McCune-Albright syndrome
- Familial male–limited precocious puberty
- hCG-producing tumors
- Exogenous exposure to androgen/estrogen
- Hypothyroidism

How to approach a patient with precocious puberty

- By history
- Onset • Progression • Other associated pubertal changes • Neurological symptoms(headache,blurred vision,syncope,seizures) • History of previous CNS insult • Abdominal pain(to rule out CAH) • Symptoms of hypothyroidism(constipation,cold intolerance,weight gain) • Growth velocity • Family History • Drug History
- By physical examination
- Growth Parameters • Tanner Staging • Dermatological exam • Neurological exam • Thyroid exam

Investigations

- Bone Age • TFT(rule out hypothyroidism) • LH,FSH • Estradiol/Testosterone • GnRH stimulation test • Pelvic ultrasound(to rule out hormone-releasing tumors) • Brain MRI(to rule out pituitary causes and CNS malformatios and tumors) • Others: IGF-1, cortisol, DHEAS, 17-OH progesterone.

- Management of gonadotropin dependent precocious puberty depends on:

- etiology

- Pace of sexual maturation

- Predicted adult height

- Psychosocial ?

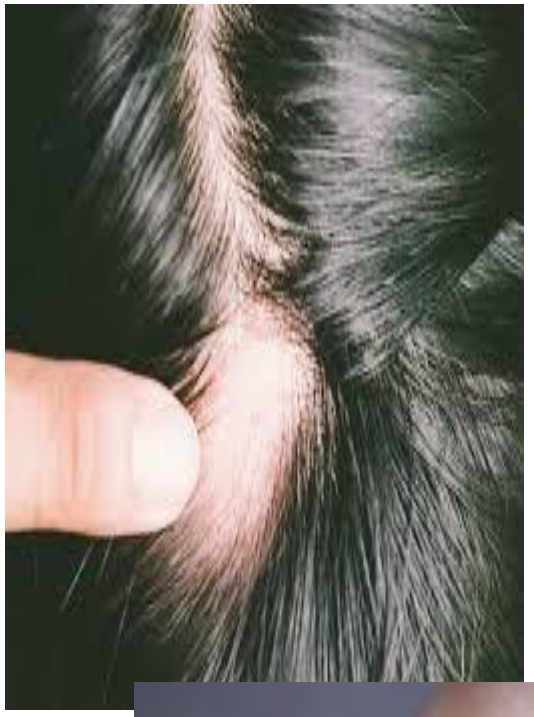
- Management for dependent types include GnRH agonist,slows pubertal acceleration,can be stopped when it is safe for puberty to proceed.

Hashimoto’s thyroiditis

- The term “Thyroiditis” refers to “inflammation of the thyroid gland”. There are many possible causes of thyroiditis . Hashimoto’s thyroiditis, also known as chronic lymphocytic thyroiditis, is the most common cause of hypothyroidism in the United States. It is an autoimmune disorder involving chronic inflammation of the thyroid. This condition tends to run in families.
- may not have any symptoms early on, even when the characteristic thyroid peroxidase (TPO) antibodies are detected in blood tests. TPO is an enzyme that plays a role in the production of thyroid hormones. If Hashimoto’s thyroiditis causes cell damage leading to low thyroid hormone levels, patients will eventually develop symptoms of hypothyroidism.

Diagnosis

- Thyroid function test showing elevated TSH with or without a decrease in T4.
- Thyroid ultrasound showing,increased blood flow indicating inflammation.
- TPO antibodies/THYROGLOBULIN Abs.
- Associated with alopecia areata(other autoimmune patchy hair loss disease)
- Complications;progression into alopecia totalis including the eyebrows,autoimmune polygladular syndrome-APS-.



Auto-immune Polyglandular syndrome

- Check antibodies against different endocrine organs;thyroid,parathyroid,adrenals,pancreas,gonads,GI.
- They may develop hypothyroidism,diabetes,primary adrenal insufficiency(treatment respectively,thyroxine 50 mg,insulin and diabetes education,substitution hydrocortisone and emergency pass)
- Presenting with one autoimmune endocrine disorder might be the first manifestation of an autoimmune poly-glandular syndrome that takes place over time.

Congenital Hypothyroidism:

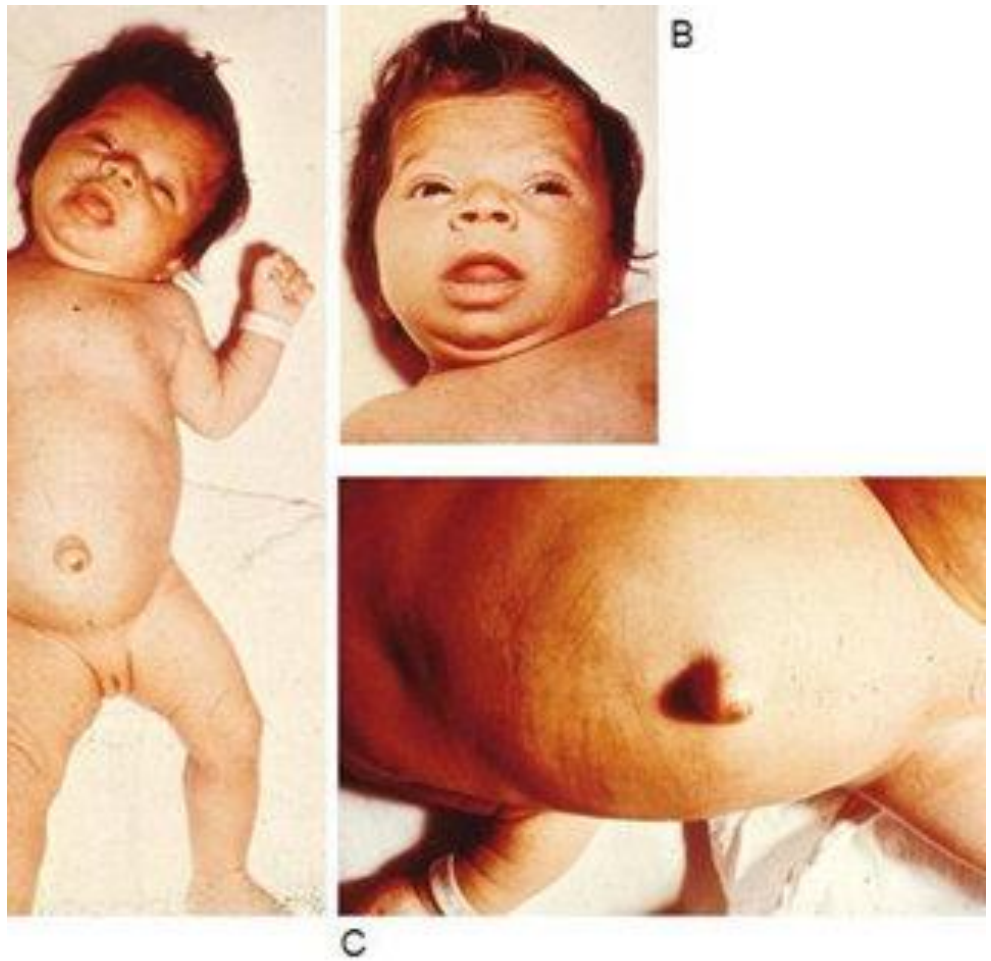
- Detection and treatment of neonates with hypothyroidism is a pediatric emergency.
- If therapy is not started soon after birth , developmental delay will result within few weeks to few months.
- Neonatal screening is essential due to the difficulty in making a clinical diagnosis early enough.

Clinical manifestations of congenital hypothyroidism:

- Most infants with congenital hypothyroidism are asymptomatic at birth.
- Birth weight and length are normal, but head size may be slightly increased.
- Prolongation of physiological jaundice .
- Decrease activity.
- Feeding difficulties.
- Respiratory difficulties.
- Constipation.
- Subnormal temperature .
- Slow pulse .

- If undetected , there will be progression which will lead to retardation of physical and mental development over the following months.
- By 3-6 months of age the clinical picture is fully developed.

- Stunted growth. Short extremities.
- Wide AF and PF.
- Coarse features.
- Protrusion of large tongue.
- Dry, scaly skin.
- Coarse, brittle and scanty hair.-
- Hypotonia.



- Major congenital anomalies occur in 3% of the normal population, but in up to 10% of newborns with congenital hypothyroidism .

- The most common associated defect is **a congenital heart anomaly.**

- A **hearing problem** is reported in up to 20% of infants with congenital hypothyroidism, so patient should undergo screening hearing test.

- Management

- Starting immediate tx of levothyroxine dose: 10–15 g/kg once daily

•Monitoring serum free T4 and TSH:

- At 2 and 4 wk after initiation of L-T4 treatment

- Every 1 to 2 months during the first 6 months of life

- Every 2 to 3 months between 6 months and 3 yr of age

-Every 6 to 12 months until growth is complete

•4 wk after a change in L-T4 dosage

Calcium metabolism and bone cases

- Calcium is regulated by the PTH,hypocalcemia stimulates PTH release,PTH has immediate effect on the distal tubules of the kidney causing increased calcium reabsorption and phosphate excretion,direct effect on the bone;stimulates osteoclasts for bone resorption,increases the activity of alpha 1 hydroxylase in the kidneys to produce vitamin d which in turn increases calcium reabsorption from the gut.

Biochemical Stages of Nutritional Rickets

Stage	Ca	P	Alk P	PTH	Events
Stage 1	N to Low	N	N to high	N to high	Poor Ca absorption from gut(radiologic finding is osteopenia)
Stage 2	N	Low	High	High(because of hypocalcemia)	Secondary hyperPTH, bone loss(alk.phosphatase elevated in bone disease) (radiologic findings rachitic changes-cupping-+1)
Stage 3	Low	Very low	Very high	Very high	Ca stores depleted, severe hypocalcemia, seizures

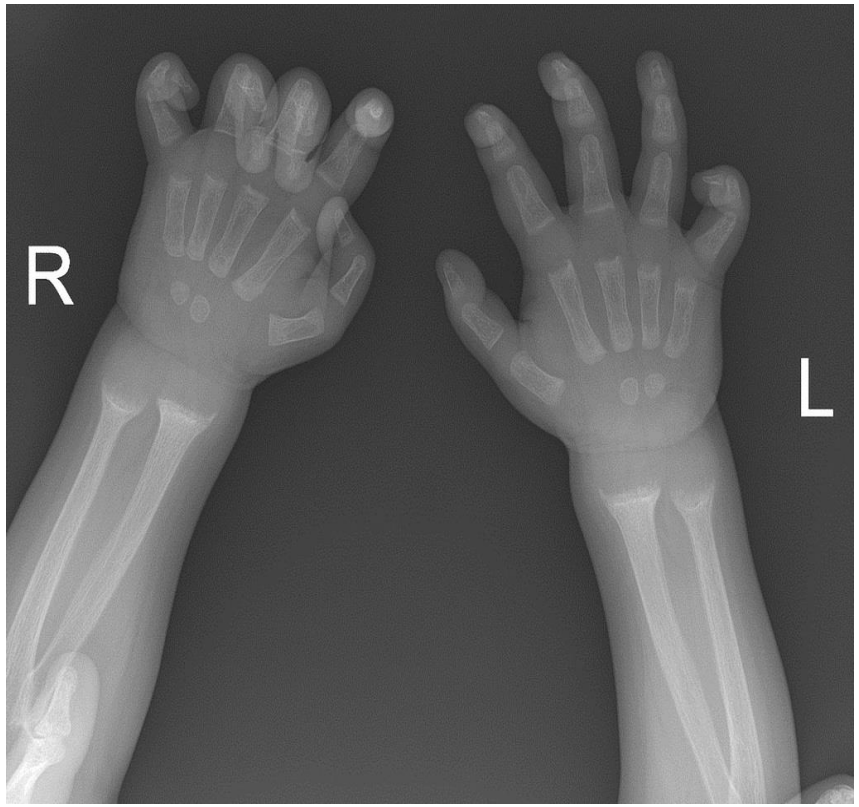
- Ca (normal range 8.5-10mg/dl)
- Phosphate(2.5-4.5 mg/dL)
- Mg (normal 1.7 to 2.2mg/dl)
- ALP (nl 20-140 iu/L)
- PTH (10-55 pg/mL)
- Albumin (3.4 to 5.4)
- 25 (OH)D (20 to 50 ng/mL)

Case 1

- Ca 6.8 mg/dL
- P 7.6 mg/dL
- Mg 3.7 mg/dl
- ALP 333 U/L
- PTH 4 pg/mL (low)
- Albumin 3.7 mg/dl
- 25 (OH)D 55 ng/ml
- Hypocalcemia,hyperphosphatemia,normal vitamin d,low PTH.
- +clinical picture of dysmorphismic features diagnosis is congenital hypoparathhyroidism due to sanjad sakati syndrome

3 year old with delayed walking and bowing of lower limbs

- Ca 6.5 mg/dL(8.5-10)
- P 2.3 mg/dL
- Mg 2.5 mg/dl
- ALP 720 iU/L
- PTH 500 pg/mL.high
- Albumin 3.7 mg/dl
- 25 (OH)D 3 ng/ml
- Hypocalcemia,hypophosphatemia,low vitamin D levels and high PTH diagnosis is nutritional rickets

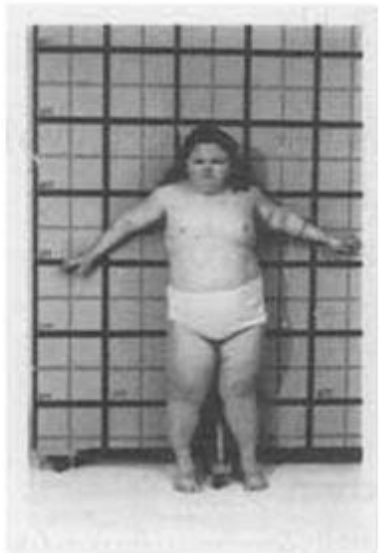


- Ca 6.5 mg/dL
- P 7 mg/dL
- Mg 2.5 mg/dl
- ALP 220 U/L
- PTH 500 pg/mL (high)
- Albumin 3.7 mg/dl
- 25 (OH)D 38 ng/ml
- Hypocalcemia,hyperphosphatemia, and normal vitamin d, high PTH.
- pseudohypoparathyroidism.(peripheral resistance to PTH)
- Same values as congenital hypopara with sanjad sakati syndrome but PTH is high(peripheral resistance)

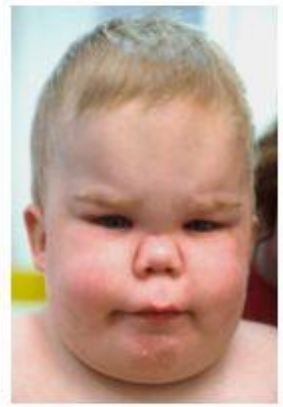
Pseudohypoparathyroidism (PHP)

Pseudohypoparathyroidism (PHP) is characterised by **hypocalcaemia**, **hyperphosphataemia** and elevated levels of serum **parathyroid hormone** (PTH).

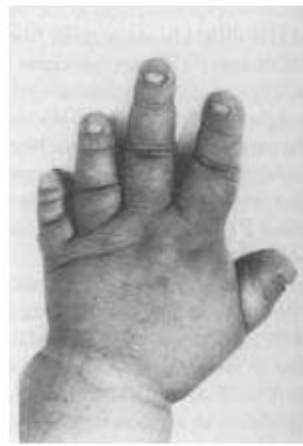
Besides PTH resistance, affected individuals may show distinctive but variable features. These clinical findings are termed **Albright's hereditary osteodystrophy** (AHO).



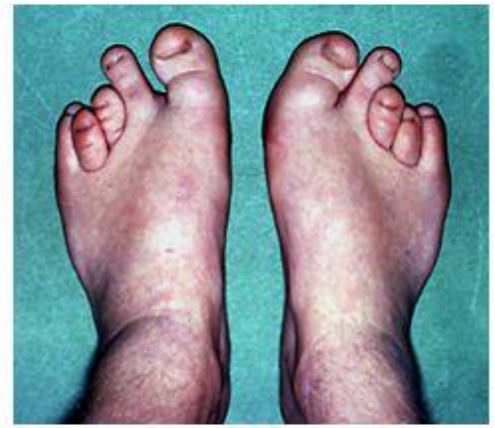
short stature, obesity, short limbs

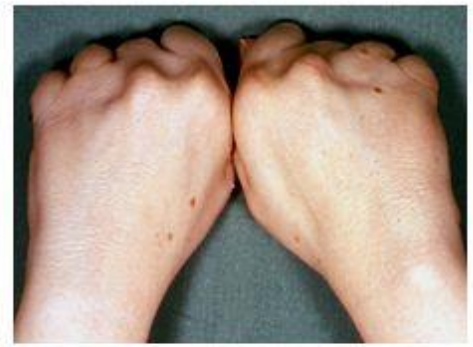


round face, mental retardation



Brachydactyly- hands/feet





Characteristic 'dimpling' replacing the knuckles

Case presentation:

8month old female baby presented with:

- poor appetite
- decreased activity
- Preference of water over milk
- Polyurea and polydipsia
- symptoms increased in severity in the last 2months
- Incidental finding of hypercalcemia
- (ca=16.5 mg/dl)

P/E:

- V/S :

HR 126 RR 35 BP90\50

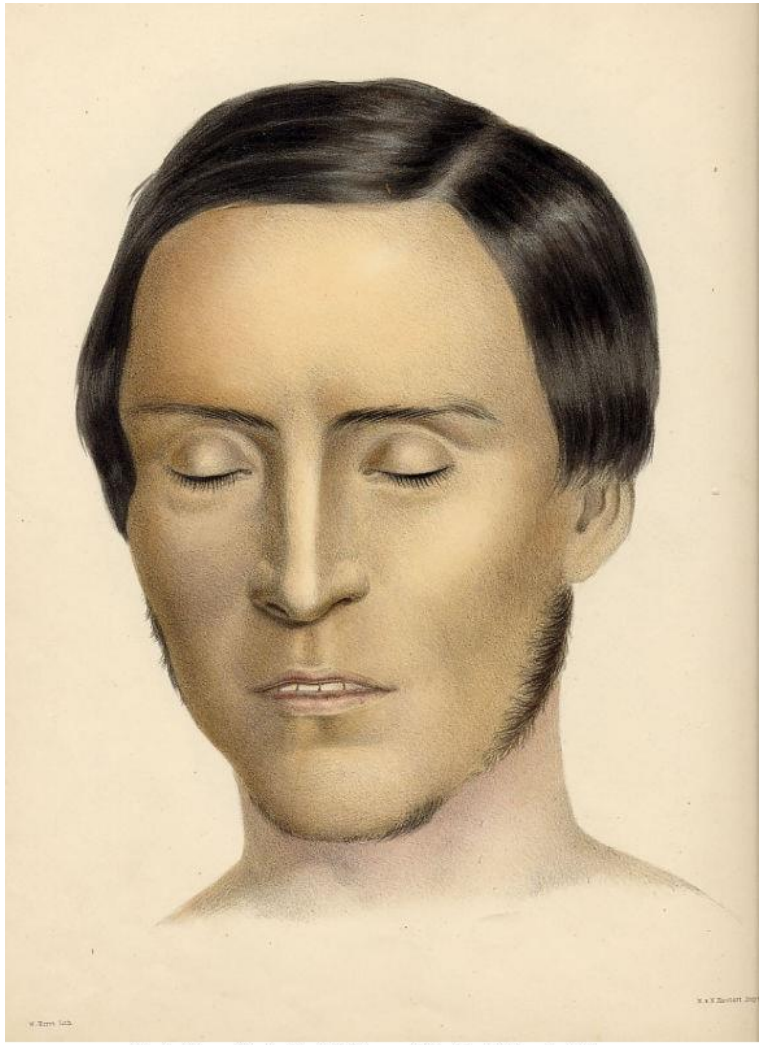
- Growth parameter

HT 65 10th centile WT 6.8 kg 10th centile

- Sunken eyes, dry lips
- GAEB with normal heart sounds
- Soft lax abdomen
- No organomegaly
- Problem is hypercalcemia and dehydration.

- The hypercalcemia was due to the fact that the patient has Williams syndrome.
- Hypercalcemia can occur in these patients and it usually is transient and mild, but can also be severe as in this patient.

Primary adrenal insufficiency: Symptoms



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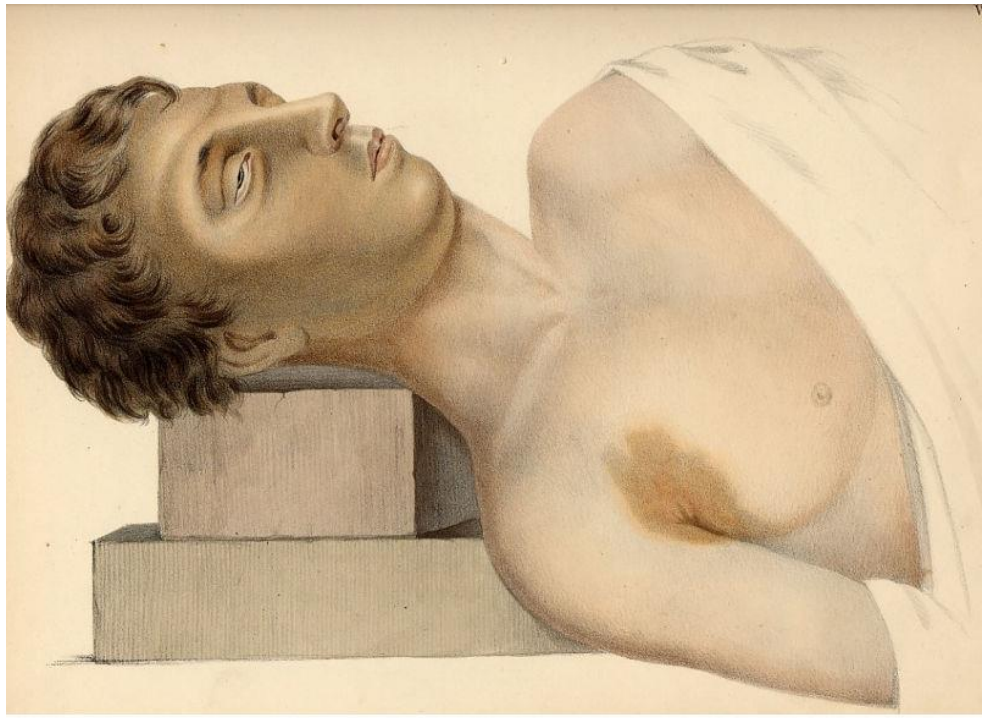
- The problem is within the adrenal cortex,very thin layer so more than one layer will be affected (low cortisol,aldosterone and androgens)-only the 1ry type.
- General Fatigue&Weakness.
- Orthostatsis
- Weight loss
- Poor appetite
- Neuropsychiatric
  - Apathy
  - Confusion
- Nausea, vomiting
- Abdominal pain
- Salt craving

Primary adrenal insufficiency: Physical findings

- Hyperpigmentation
- Hypotension
- Orthostatic changes
- Weak pulses
- Shock
- Loss of axillary/pubic hair (women)



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Primary adrenal insufficiency: Laboratory findings

- Hyponatremia(low aldosterone and low cortisol) and hypoglycemia(because of low cortisol)
- Hyperkalemia(aldosterone causes na reabsorption and k excretion)
- Narrow cardiac silhouette on CXR
- Low voltage EKG
- Diagnosis ;
    1. 8 am cortisol levels
    2. Acth serum levels
    3. Acth stimulation test
- Treatment is divided into acute management and maintenance;
    - Acute(adrenal crisis) is the stress dose of hydrocortisone ,fluid replacement, dextrose for hypoglycemia,potassium usually normalizes with fluid.
    - Maintenance is by replacement of mineralocortecoids and corticosteroids and patients education about Emergency steroid administration IM hydrocortisone.

Congenital adrenal hyperplasia

- A common cause of primary adrenal insufficiency,the most common cause for CAH is 21-hydroxylase deficiency.
- This enzyme deficiency causes unability to produce cortisol and aldosterone because this enzyme is needed in the pathway,but not in the androgenic pathway,also the accumulation of substrates that cannot be converted into end products because of enzyme deficiency wil be shifted into the androgenic pathway,causing excess androgen,also the constant low levels of cortisol will keep stimulating ACTH secretion causing excess androgens and hyperplasia of the gland itself.
- Excess androgen leads to virulization in girl,or early puberty.

Cushings syndrome

- Acth-dependent: (hight ACTH)
  - Cushing disease(pituitary ACTH secreting tumor)
  - Ectopic secretion of ACTH (small cell lung cancer)
- ACTH-independent:
  - Gluco-corticoid therapy
  - Adrenal adenoma

- Diagnosis ;
  - Acth serum levels
  - 24hr urine free cortisol
  - Salivary cortisol or 8am&8pm cortisol levels.
  - Dexamethasone suppression test

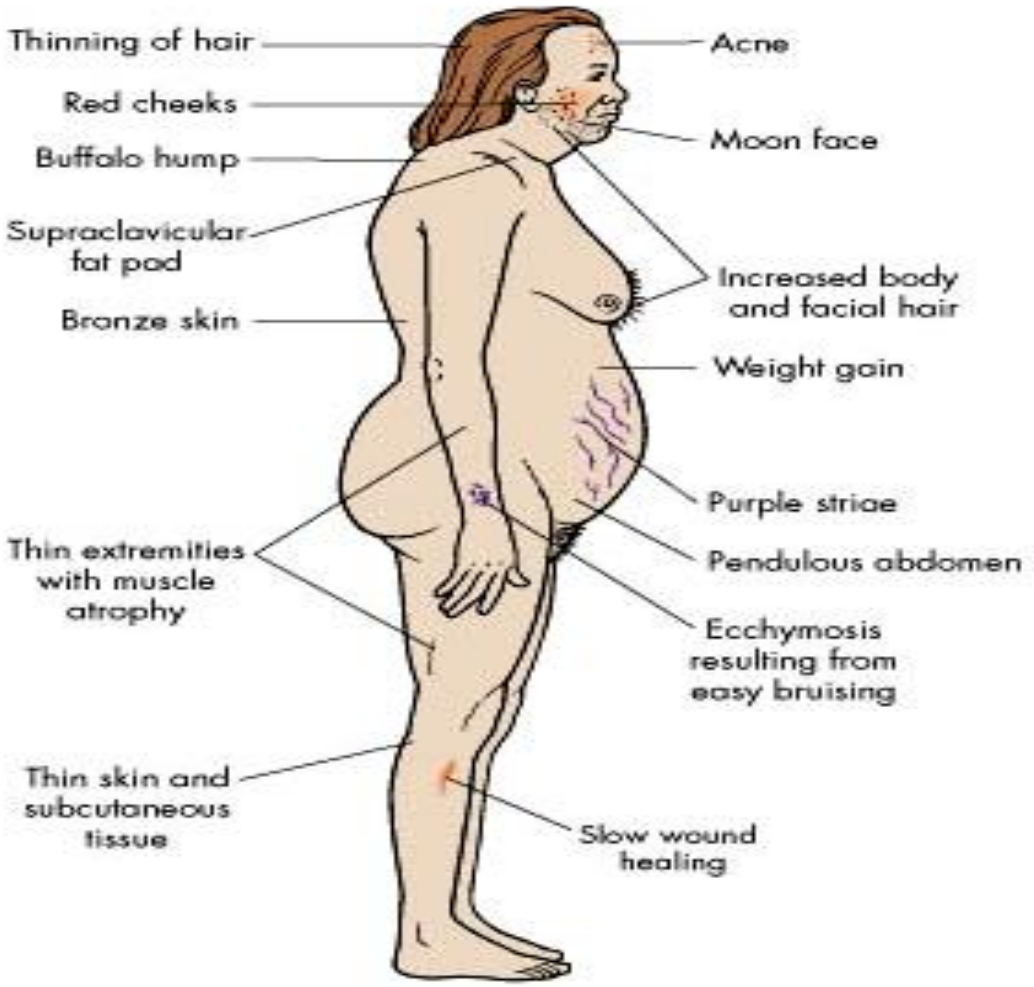


Figure 47-9 Common characteristics of Cushing's syndrome.

Pheochromocytoma or catecholamine secreting tumor

- Triad of episodic headache, diaphoresis and tachycardia ±hypertension.
- **Diagnosis:**
  - Measurement of catecholamine metabolites (metanephrines) –urine/blood
  - Localization by CT imaging and MRI
- Treatment is surgical

Neurology

Approach to developmental delay

- **screen** children for general development using **standardized,** validated tools at 9, 18, and 24 or 30 months
- for autism at 18 and 24 months
- **or at any point when** a caregiver or the clinician has a concern.
- Screening is assessment of asymptomatics with no previous history/family history or risk factors.
- Survillence is assessment of asymptomatics with previous history or family history or risk factors as a follow up for early detection.
- Screening is not intended for diagnosis but rather for more rapid identification of individuals who require further evaluation.
- Children identified via screening as positive for a particular disorder can be referred for further evaluation, which usually entails more comprehensive history, testing, and examination to elucidate diagnoses more clearly.(referred for surveillance)

- global developmental delay because she has delay in 2 developmental milestones or more.
- Most developmental clinicians and researchers correct for prematurity for the first 24 months after birth
- You correct to gestational age of **40 weeks**
- The incidence of visual and hearing impairments is higher in preterm than term children

  Due to increased risk for retinopathy of prematurity, jaundice, cortical hemorrhages, infections, and extended hospitalization.
- Unrecognized visual or hearing impairment can distort performance on cognitive and behavioural testing.
- Children born preterm are also at greater risk than their term peers for: intraventricular hemorrhage +possible cerebral palsy which can also affect performance on assessments of motor function.
- Importance of early detection early referral to therapeutic services and higher chance for catch up.

Approach to autism

- Autism spectrum disorder is a neurodevelopmental disorder.
- All children should be screened for ASD at age of 18 months and 24 months in order to detect as many cases as possible,early detection,early referral for rehabilitation and better outcomes.
- characterized by impairments in two major domains:

  1) deficits in social communication and social interaction

  2) restricted repetitive patterns of behavior, interests, and activities
- The presentation can be as a delay in development early on (9-10 months) in 50% of patients,or they may remain normal until 18-24 months and present with regression in social and language skills.
- Motor skills are usually normal and not affected.

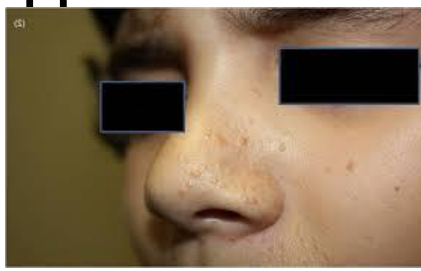
Some of the symptoms they may have

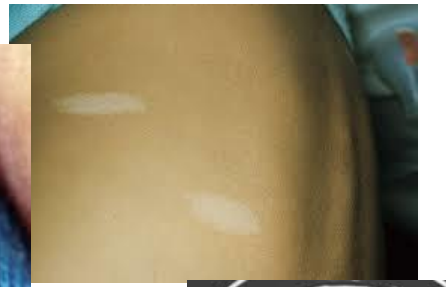
1. Repetitive patterns;rhythmic movements,echololia meaningless repetition of words
2. Social separation from parents may elicit a lack of eye contact,absence of typical responses to pain or physical injury,difficulties in social interaction,doesn’t imitate parents.
3. Language;language delay and speech abnormalities
4. Pointing;protodeclarative pointing using the index finger to indicate an intrest in an item to another person,absence of this is predictive of ASD.
5. Play;fascinated with a certain toy,interest in part of an object instead of a functional use of the whole object,doesn’t pretend in play.
6. Environmental stimuli;unusual response to external stimuli either lack of response or exaggerated response,this is indicative of ASD,item number 12 in the MCHAT if YES the child fails in this item.
7. Self-injurious behaviour;skin-picking,self-biting,slapping..

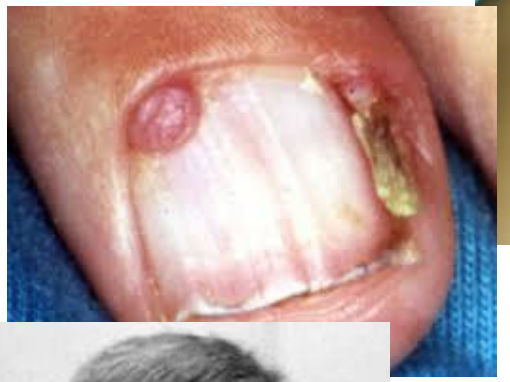
- What screening tool is used for early detection of ASD?
  - M CHAT(modified checklist for autism in toddlers)
- How to use the checklist and interpret the data?
  - The child receives one point for each failed response.
  - For all items answer with no is indicative for ASD,for items 2,5,12 answer with yes in indicative for ASD.
  - Score 0-2:low risk,child passed the screener,if child is younger than 24 months repeat the test after the 2nd birthday,no further action required until surveillance indicates high risk for ASD.
  - Score 3-7:medium risk,child is required to follow up for further evaluation in depth using the MCHAT-R/F.
  - Score 7-20:high risk,you can refer the child to an interview for further evaluation to gain clarity,or you can skip this an refer the patient to diagnostic evaluation and early intervention services.
- What are the 2,5,12 items?(if answers yes he fails the items and scores 1 point)
  - 2,have you ever wondered if your child is deaf?
  - 5,does your child do unsual finger movements?
  - 12,exaggerated response to external stimuli or lack of response?

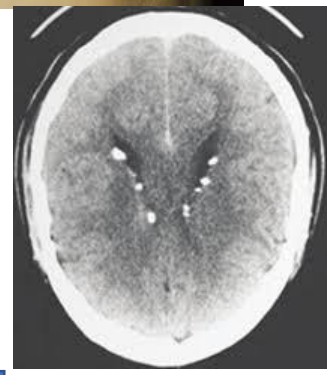
Next step is look for signs of dysmorphism or signs for syndromes that come with autism

- Syndromes that come with autism
  1. Tuberous sclerosis a rare disease causing Tumors, or growths, in the brain and other organs (periungal or subungal fibromas,ash leaf spots,angiofibromas,ventricular calcifications.)
  2. Fragile x s a genetic disorder characterized by mild-to-moderate intellectual disability.
  3. PKU;phenylketonuria
  4. Angelman syndrome
  5. Down syndrome
  6. Neurofibromatosis(café au lait Spots, neurofibromas)

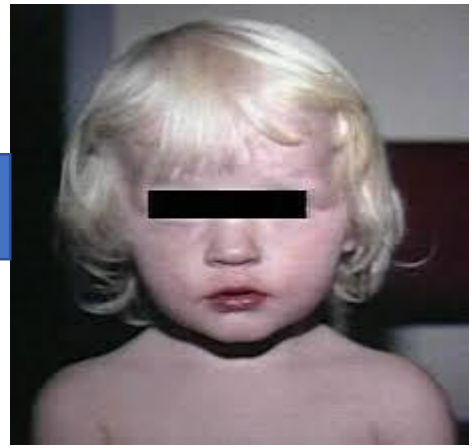






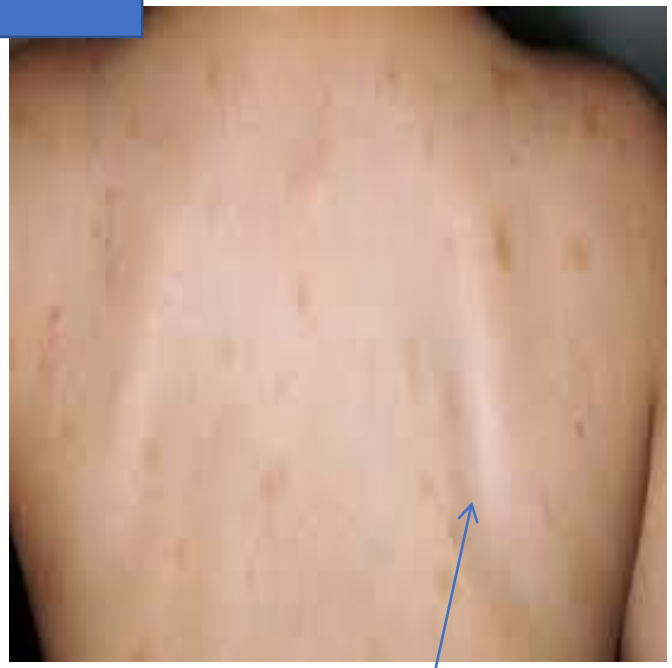


PKU



Fragile X

Down syndrome



Neurofibromatosis 1

Angelman syndrome

Next step is rule out co-occurring disorders and do investigations

- Co-occurring conditions;anxiety,OCD,ADHD,depression.
- Investigations:

  Hearing assessment, high resolution microarrays to rule out chromosomal abnormalities, whole genetic sequencing, metabolic workup, FMR1 to rule out fragile x. EEG and MRI not routinely done only if seizures.
- Treatment :
  - There's no cure for autism, and there's no "one-size-fits-all" treatment
  - **Behavioral and communication therapies** : ABA
  - **Drug therapies :** for associated conditions
  - **Complementary and alternative therapies:** needs to be discussed with parents
  - Needs prolonged and skilled effort
  - Discuss with parents : dietary changes (gluten free), vitamins, chelation, hyperbaric oxygen

Further elaborations

- Severity : each case is decided alone
- Prognosis : continues to develop slower than expected but does not deteriorate, intelligence is correlated with prognosis.
- Factors associated with better outcome/worse outcome
  - Presence of joint attention/absence
  - Functional play skills
  - Higher cognitive abilities/co-morbidities or seizures.
  - Decreased severity of autism symptoms/severe autistic symptoms
  - Early identification
  - Involvement in intervention
  - A move toward inclusion with typical peers
- Risk of recurrence depends on underlying aetiology (if identified) ,Identification rate of an underlying aetiology is 5-20 % .
- In general (if no aetiology could be identified) :the risk of recurrence is up to 25 %.

Approach to a patient with 1st seizure

- What he was doing before the attack,crying or angry to rule out breath holding spell(a period of breath cessation that causes loss of consciousness),eating to rule out aspiration,jumping to rule out trauma.
- Detailed description of the event; duration,type of movement,parts of the body involved(if more than one which one came first),preceeding stare,eyes,frothy secretions from the mouth,urination and defecation,associated apnea or cyanosis,post-ictal weakness.
- Causes could be : infection,autoimmune,trauma,iatrogenic,hereditary/congenital malformation,neoplastic,be systematic ask one by one in the history.

- Infection: fever, signs and symptoms of meningitis or any other source of infection from other systems,endocrine disorders to rule out electrolytes disturbances(hypocalcemia,hypomagnesemia,hyponatremia).
- Trauma:recent head trauma or injury.
- Autoimmune:rule out vasculitis and other rheumatological conditions by asking about joint swellings,pain or rashes.
- Iatrogenic:any drugs used,ingestion of toxins or food poisoning.
- Neoplastic:ask about focal neurological signs,history of worsening headache,signs of increased intracranial pressure,and constitutional symptoms.
- Hereditary or familial: parents consanguinity,family history of seizures,perinatal history any insults during pregnancy,birth weight,NICU admission or prolonged jaundice.
- **P/E:**Perform a complete general and neurological examination,Look for dysmorphic features,Look for focal neurological signs ,Look for skin lesions that may orient you towards a specific neurocutaneous disorder(NF,tuberous sclerosis)

Differential diagnsosis of a child with seizure

- Seizure
- Epilepsy (2 unprovoked seizure,occurring at least 24hrs apart)
- Febrile seizure associated with fever>1 month of age usually 6m-6years (peak 18 m) in the absence of cns or electrolyte imbalance.
- Epileptic encephalopathy :epileptiform abnormalities cause it,associated with developmental delay.
- Seizures are classified according to onset into :focal(aware or unaware) including focal to bilateral,generalized onset,unknown onset.
- Another classification depends on etiology.could be unknown

Investigations

- LP:because the patient was a febrile
- Electrolyte screen
- Toxicology screen
- Brain MRI,sleep EEG,positivity rate 60% the standard,sleep is 90%
- Management:
  - No need for treatment since the recurrence rate for 1st seizure is 30-50% only council about if recurrence happens,lateral position,keep safe,nothing inserted into the mouth,if seizure continues for more than 5-10 mins refer to nearest hospital or give valium per rectum..
  - Recurrence of 2nd seizure is 80%.
  - The recurrence is usually between the 6months-2 years period,after 2 years from the seizure recurrence is rare.

Approach to a child with headache

- History
- O:onset and duration
- C:character or headache
- R:radiation
- A:associated symptoms
- T:timing,when does it happen and how frequent,course and progression in terms of intensity and frequency
- E:exacerbating and relieving factors.triggers and what makes it better
- S:severity in terms of awakening from sleep

Table 4. Red Flags for Secondary Headache

- Progressive pattern of the headache: becoming more severe and/or more frequent
- Increased headache with straining, coughing, or sneezing
- Explosive or sudden onset of severe headache (<6 mo duration)
- Systemic symptoms: fever, weight loss, rash, and joint pain
- Secondary risk factors: immunosuppression, hypercoagulable state, neurocutaneous disorder, cancer, genetic disorder, and rheumatologic disorder
- Neurologic symptoms or signs: altered mental status, papilledema, abnormal eye movements, or other abnormalities or asymmetries on neurologic examination
- New or different severe headache, change in attack frequency, severity, or clinical features
- Sleep-related headache, headache waking the patient from sleep, or headache always present in the morning

ddx

Episodic recurrent headaches
- Tension-type headache
- Migraine with or without aura
- Fasting/eating disorders
- Recurrent toxic exposures: alcohol, toxins, illicit drugs, medications
- Recurrent sinus disease
- Seizure-associated headache
- Mitochondrial disease
- Trigeminal autonomic cephalalgias

- Migraine : with or without aura
- Tension headache : band like
- Chronic headache : >15 days /month
- Trigeminal autonomic cephalgia : accompanied by autonomic symptomsThe **Trigeminal Autonomic Cephalalgias** (TACs) are a group of headache disorders characterised by attacks of moderate to severe unilateral pain in the head or face, with associated ipsilateral cranial **autonomic** features such as lacrimation, conjunctival injection, rhinorrhoea, nasal congestion, eyelid oedema and ptosis

Table 6. **SMART Headache Management**

Sleep	Regular and sufficient sleep
Meals	Regular and sufficient meals, including breakfast and good hydration
Activity	Regular (but not excessive) aerobic exercise
Relaxation	Relaxation, stress reduction, and management
Trigger avoidance	Avoid triggers such as stress, sleep deprivation, or other identified triggers

Approach to flaccid paralysis

- ACUTE FLACCID PARALYSIS (AFP):The sudden onset of generalized flaccid weakness in the absence of symptoms of encephalopathy… implicates the motor unit,AFP is an emergency.
- The initial complaint could be weakness,proximal expressed as difficulty getting up from a chair or difficulty climbing up or going down the stairs or distal weakness tripping by nothing or it could be abnormal gait.
- Older children will complain of specific disabilities Limb weakness often associated with weakness of muscles of the head and neck Inquire about diplopia, drooping eyelids, difficulty chewing and swallowing, facial expressions, and voice changes.
- Physical exam:Look for atrophy or hypertrophy,Fasciculations,Palpate muscles for tenderness and texture,Joint contractures, myotonia,Strength and tendon reflexes,Watch the child sit, stand, and walk Neck flexion/extension,Particular attention to oculomotor exam, speech (nasal speech), cough,Assess the presence of facial weakness,Sensory exam.
- Ddx:ACUTE FLACCID PARALYSIS
  - Guillain Barre Syndrome
  - Transverse Myelitis
  - Poliomyelitis
  - Botulism

	syndrome)			
Presentation	It is an autoimmune inflammatory demyelinating polyneuropathy typically comes after a respiratory tract infection or gi infection,usually presents with symmetrical weakness that ascends over hours or days,in severe cases may involve the respiratory muscles,cranial nerves involvement mainly facial nerve and ocular muscles,no pupillary abnormalities ,absent or diminished reflexes,bladder and bowel dysfunction are not persistent.	Immune mediated spinal cord inflammation,clinical presentation varies depending on the level of injury and usually resembling mass occupying lesions(myelopathy) so its an emergency for MRI spine,asymmetrical weakness,sensory neuropathy,early bladder involvement and back pain,should be considered with any ascending weakness especially if without bulbar involvement,reflexes can be +or -	Poliovirus,causing motor neuron damage in the brain stem and spinal cord,purely motor and no sensory component,starts with a viral prodrome then meningism then weakness muscle atrophy diminished or absent reflexes and paralysis.	Caused by c.botilinium Acute onset of bilateral cranial neuropathies associated with symmetric descending weakness,preceeded by classic pentad: dry mouth, nausea/vomiting, dysphagia, diplopia, and fixed dilated pupils.
Prognosis and treatment	Prognosis is excellent if given the needed care and respiratory support Tx include plasma exchange and IVIGs with or without	Prognosis 50% recover completely and 10% do not recover others partial recovery,tx IV steroids.	Treatment is mainly supportive,physiotherapy,ventilation &pain management.	Antitoxin and antibiotics

Nephrology

- Approach to acid base disorders:
  1. Check PH,<7.35 acidemia,>7.45 alkalemia
  2. Know the primary cause by looking at pco2(respiratory) and hco3(metabolic),the abnormal value is the primary cause.
  3. What's the primary process? Acidosis if Pco2>45 its respiratory,if hco3<20 its metabolic ,alkalosis if pco2 is <35 its respiratory,if hco3>28 its metabolic.
  4. See the compensation,if metabolic process calculate the expected pco2 value if they're not equal +/- there is a secondary process,if respiratory calculate the expected hco3 acute and chronic compensation(same mechanism)
  5. Calculate the anion gap for metabolic acidosis,if normal calculate the urine anion gap.

Normal values and formulas:

- Ph=7.35-7.45
- Paco2=35-45 mEq/L
- Hco3-=18-25 mEq/L
- Anion gap = sodium – (cl + hco3) normal 11-14-for metabolic acidosis.
- Urine anion gap = na + k – cl normally zero-calculated when normal anion gap
- Expected pco2= 1.5(hco3)+8 (+/-2)
- Metabolic compensation for respiratory acidosis(الارقام الصغيرة): acute(within minutes) for each 10 mEq pco2 increase there is 1 mEq hco3 increase.chronic for each 10 pco2 increase there's 3.5 hco3 mEq increase.
- Metabolic compensation for respiratory alkalosis(الارقام الكبيرة):for each 10 pco2 decrease there's 2 hco3 mEq decrease,and for each 10 pco2 decrease there's 5 hco3 mEq decrease.

Causes

- Respiratory acidosis(conditions causing co2 retention):
  1. Emphysema,COPD,bronchiectasis,asthma.
  2. Upper airway obstruction laryngospasm,bronchospasm,aspiration.
  3. Lower causes such as pneumonia and pneumothorax.
- Respiratory alkalosis(conditions that increase co2 wash out,hyperventilation)
  1. Anxiety related.
  2. High altitudes.
- Metabolic acidosis:
  1. Increased excretion of hco3(diarrhea,RTA)-normal anionic gap its zero normally(hyperchloremic state) do urine anionic gap if +ve its distal RTA-1,if –ve diarrhea(anus is low) or procimal RTA-2
  2. Decreased excretion of acid(Renal failure)-high anionic gap
  3. Increased acid production lactic acid or ketoacid(ischemia and DKA)-high anionic gap
  4. Acidic drugs or poisons(ASA)-high anionic gap
- Metabolic alkalosis(volume depletion):
  1. Chloride responsive(urine chloride<15)management is fluids(nacl)-vomiting,CF,diuretics,pyloric stenosis
  2. Chloride resistant (urine chloride>20)management is by blocking the excess of mineralocortecoids depending on the cause.(barter,liddle,gitelman)

Both cause chloride resistant metabolic alkalosis along with liddle syndrome

Some Differences Between Bartter Syndrome and Gitelman Syndrome

Feature	Bartter Syndrome	Gitelman Syndrome
Location of kidney defect	Ascending loop of Henle (mimics effects of loop diuretics)	Distal tubule (mimics effects of thiazides)
Urinary calcium excretion	Normal or increased, commonly with nephrocalcinosis	Decreased
Serum magnesium level	Normal or decreased	Decreased, sometimes greatly
Renal prostaglandin E2 production	Increased	Normal
Usual age at presentation	Before birth to early childhood, often with intellectual disability and growth disturbance	Late childhood to adulthood
Neuromuscular symptoms (eg, muscle spasms, weakness)	Uncommon or mild	Common

Definite approach to metabolic problems

- Ph<7.35,Hco3 is low(metabolic acidosis)
  - Do anion gap, if high its either lactic acidosis,dka,drugs and poisons(ethylene glycol or methanol),renal failure, if normal(hyperchloremic type) do urine anion gap(normally zero) if+ve distal RTA-1,- ve diarrhea(anus is low) or proximal RTA-2.
  - Failure to thrive suggests chronic metabolic acidosis.
- Ph>7.45,hco3 is high(metabolic alkalosis)
  - Urine chloride,if urine chloride<15 its chloride responsive its either vomiting or diuretics or CF(volume depletion) management is by fluids(nacl),if urine chloride>20 its chloride unresponsive(block the action of excess mineralocortecoids)

Distal RTA TYPE 1

- Causes metabolic acidosis with normal anion gap and hypokalemia.
- Urine anion gap is positive.
- Hypercaliurea&nephrocalcinosis(positive ions)
- Ph cannot fall below 5.5(لانه الاول)
- Can be secondary to metabolic disorders such as cystinosis.

Proximal RTA TYPE 2

- Causes metabolic acidosis with normal anion gap and hypokalemia.
- Hypophsphatemia.
- Urine anion gap is negative.
- Aminoacidnuria&glucosuria(negative ions)
- Ph can fall below 5.5
- Can be familial or secondary to autoimmune disease

Nephrotic syndrome

- Presentation is edema,sites include: periorbital worse at the morning, chin of the tibia, ascites, scrotum, in infants look for sacral edema.
- Things to ask in the history to a patient with edema:

1. Difficulty breathing or history of cardiac diseases to rule out cardiac problems.
2. Jaundice or chronic diarrhea to rule out GIT causes of ascites.
3. Skin rashed with angioedema to rule out allergies.
4. Family history of renal disease and drug history.
5. Frothy urine,decreased urine output,blood in urine.
6. Other sites of edema

Theories explaining edema in nephrotic syndrome

- Underfill theory,the kidneys are losing too much protein than daily synthesis,causing hypoalbuminemia,this decreases the oncotic pressure and this drives fluid out of the intravascular compartment causing 3rd spacing,also drives RAAS activation causing sodium and water reabsorption causing low urine sodium,low serum sodium is dilutional.
- Overfill theory,primary sodium retention because of distal tubular injury leading to resistance to ANP(causes sodium excretion)this leads to sodium and water retention overfilling the intravascular compartment and causing edema(increased intravascular hydrostatic pressure)

- What to monitor a patient admitted for edema in the nephrology department?
    1. Daily weight
    2. Input and urine output(number of ccs in a certain duration/weight/number of hours in that duration) ml/kg/hour(normal urine output is more than 1 ml/kg/hour).
- Other labs needed:

1. Electrolytes mainly na,k(can have dilutional hyponatremia).
2. Kidney function test(creatinine and urea).
3. Serum albumin(hypoalbuminemia)
4. Urinanalysis:wbcs above 5cells/hpf is pyuria,rbs above 5 cells/hpf is hematuria,protein 3+ and above is nephrotic range.we quanitify the proteinuria by doing the spot protein-creatinine ratio,above 2 is nephrotic range.(low urine sodium)
5. Lipid profile:shows elevated glycerol and triglycerides because of the liver response to hypoalbuminemia by increased synthesis of lipoproteins,complement is normal.

- **Microscopic hematuira can be seen in nephrotic syndrome and it is usually transient.**
- **the lipid profile will normalize when albumin is back to normal.**

Kidney biopsy is not needed as most cases of nephrotic syndrome are minimal change disease.

- So we can start treatment and assess the response.
- However if the child was less than 1 year or older than 10 year and has gross hematuria/Hypertension/impaired renal function then he needs a biopsy or resistant to steroid therapy.
- <u>Management :</u>
  - Start steroids for 4 weeks,protein will be negative after 10-14 days.
  - Supportive therapy includes:sodium restriction and no excess fluid intake,high protein diet has no role.
  - Oliguria is caused by intravascular depletion we can solve it by giving the patient albumin and furosemide to resolve the Edema.
  - Not to give the live attenuated vaccines,advice the mom to give the yearly influenza vaccine and the added vaccines pneumococcal and chicken pox

Prognosis and complications

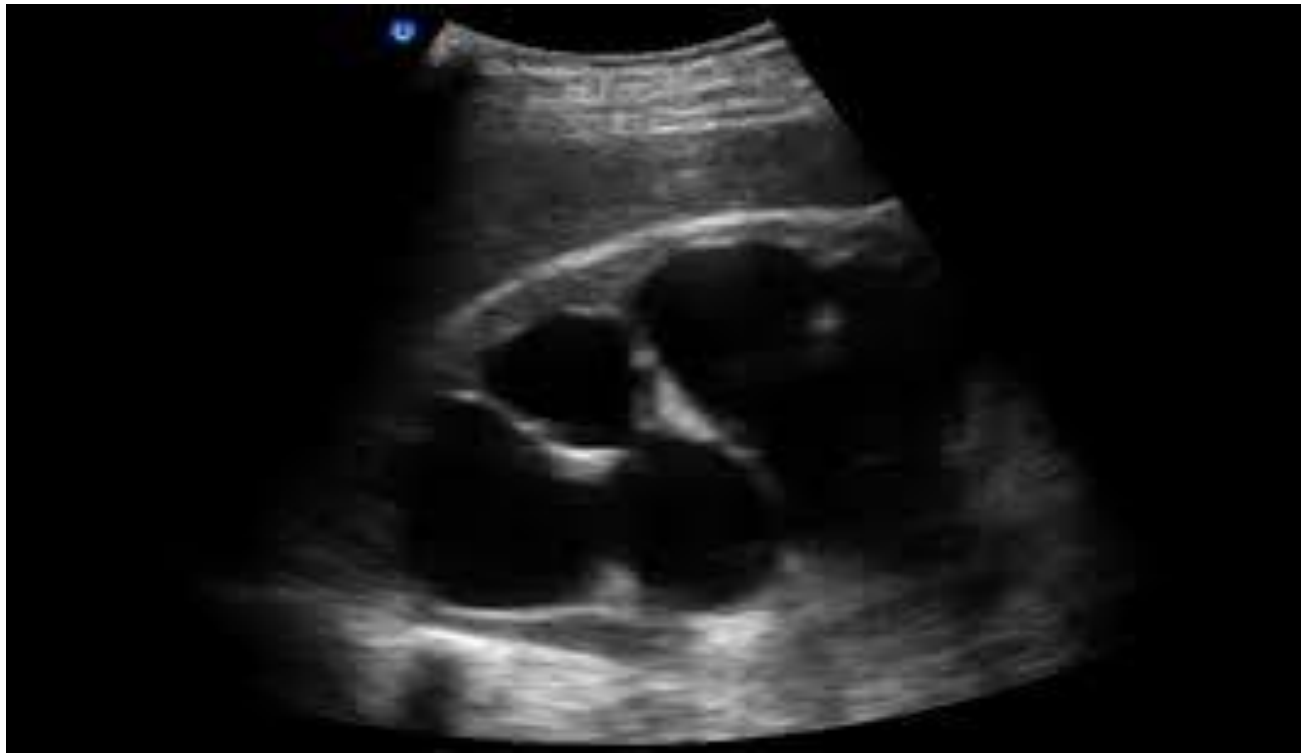
- 90% of patients are expected to relapse by having edema and nephrotic range proteinuria,usually after an URTI,a relapse is defined as edema and proteinuria 3+ and above for three days.
- Complications
  - Child is at risk of infection because of lost immunoglobulins in the urine,impaired opsonization especially the encapsulated bacteria pneumococcal
  - At risk of thrombosis because of low AT3.
- Complications of long term use of steroids:
  - Weight gain and increased appetite.
  - High bp and glucose readings.
  - More nervous.
  - Osteopenia:keep a balanced diet rich with vitamin D and calcium.
- Minimal change disease doesn't lead to renal failure, except if he is resistant to steroid therapy then a renal biopsy is indicated and he might be having steroid resistant nephrotic syndrome and this can progress into CKD.

•**STERIOD RESISTANCE** :Failure to respond after initial 4-8 weeks of steroids
•**STERIOD DEPENDANT:**Two consecutive relapses during steroid therapy or within 14 days of ceasing therapy
•Remission is zero on dipstick for 3 days.

VUR(vesicouretral reflux)

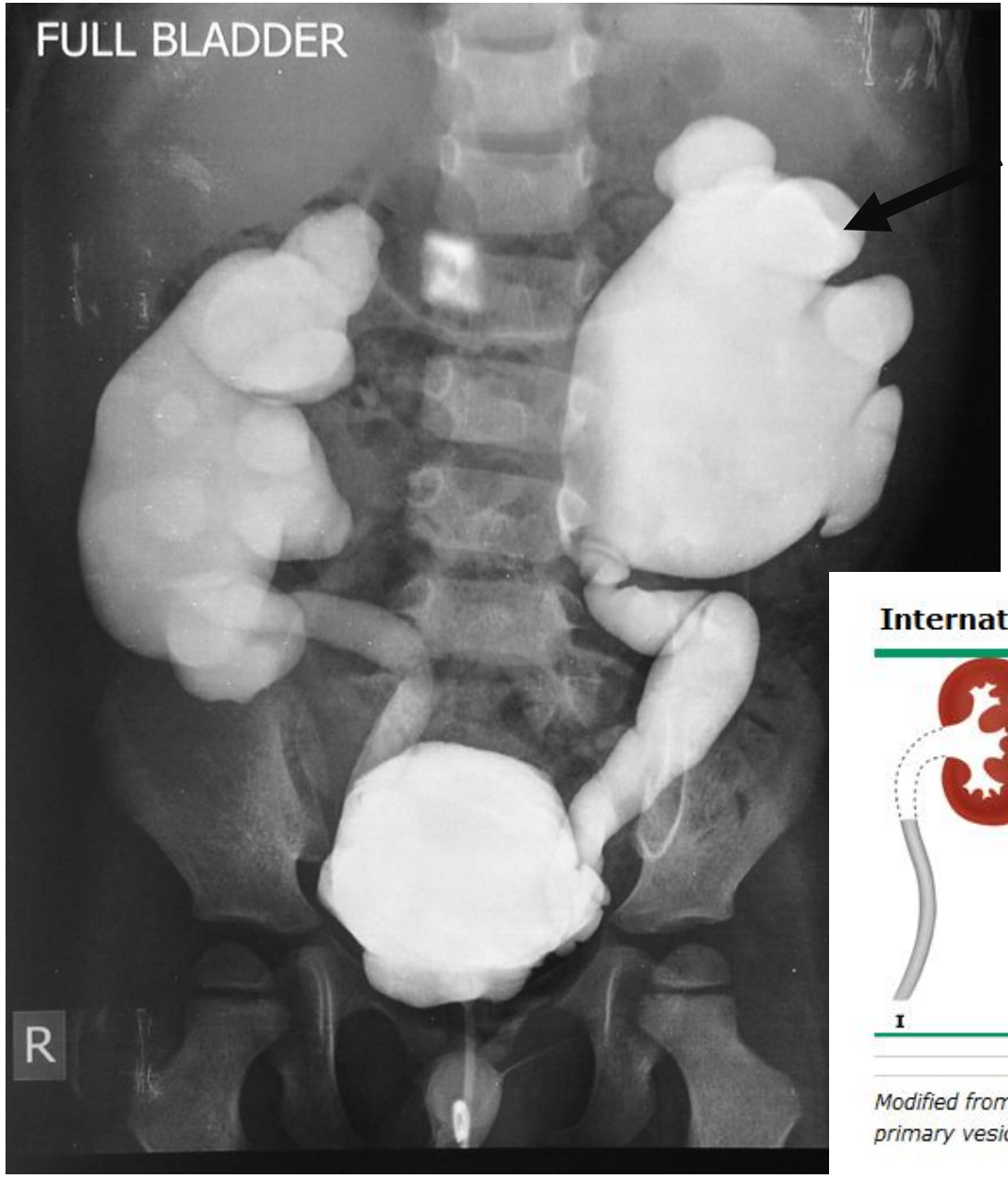
- Complaint:recurrent UTI or antenatal us showed hydronephrosis.
- Investigations in order:ultrasound,VCUG.
- Causes:primary present at birth and theres a defect in the development of the kidney/valves and it’s the most common cause in children,secondary to posterior urethral valve or neurogenic bladder.
- **Posterior urethral** valves (PUV) are obstructive membranes that develop in the urethra close to the bladder.
- **Neurogenic bladder**, the nerves that carry messages back-and-forth between the **bladder** and the spinal cord and brain don't work the way they should.
- Other investigations include: baseline DMSA scan to assess function&KFT.
- Scarring on DMSA can be the result of dysplastic kidneys or recurrent uti

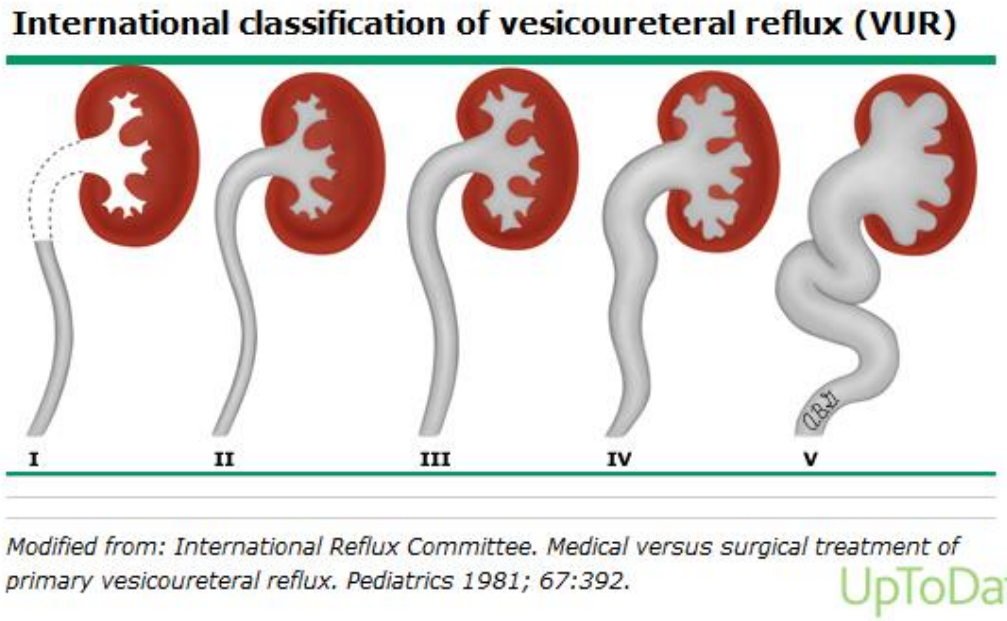
Renal US showing hydronephrosis



- Ddx:
- Bilateral VUR.
- Posterior urethral valve.
- Next step is do VCUG/MCUG
- It showed bilateral severe hyronephrosis with dilated ureter and cortical thinning and thickened and trabeculated bladder

GRADE 5 ,because of the tortous dilated ureters and calyceal blunting as the arrow shows



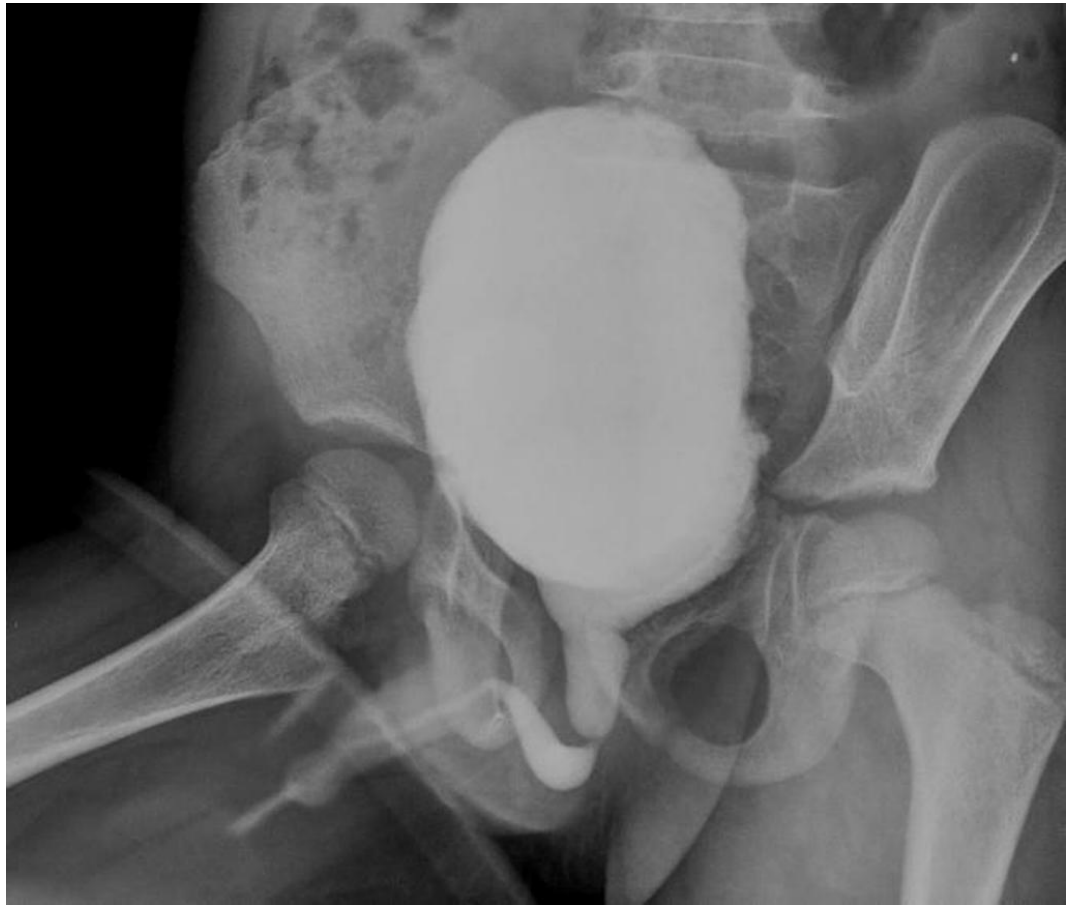


Management

- Prophylactic Abx.
- Follow up creatinine.
- Surveillence for UTIS.
- Surgical correction of the high grade VUR because its unlikely to resolve byitself.
- Treat bowel and bladder dysfunction.

Posterior urethral valve

- Poor stream,drippling or antenatal us showed hydronephrosis .
- Do bilateral ultrasound then VCUG
- Findings:elongated bladder and trabeculated outline normally its round and smooth,also narrowing in the urethra and dilatation proximal to the narrowing.
- Management is surgical either through valve ablation by cystoscopy or vesicostomy for drainage.

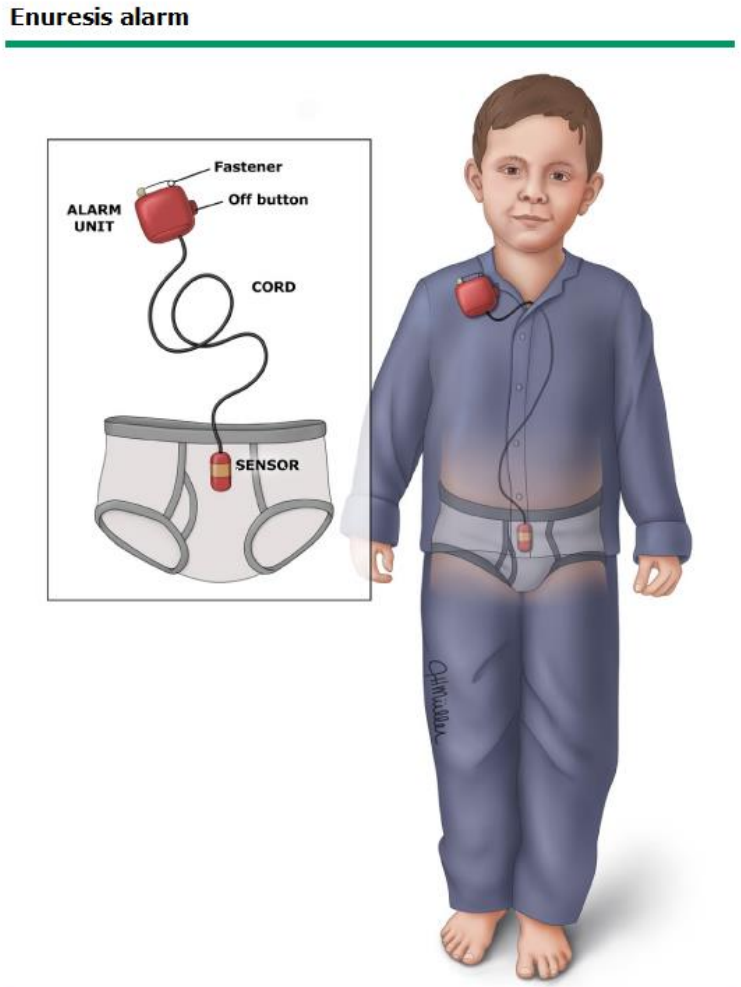


Primary enuresis(wetting the bed)

- **Enuresis** is considered **primary** when bladder control has never been attained and secondary when incontinence reoccurs after at least six months of continence.
- Ask about day time symptoms of wetting,urgency,frequency,if not present it is called mono-symptomatic enuresis,if present its called non-monosymptomatic enuresis(bladder dysfunction with various types)
- Ask about constipation,fluid intake in the night,family history of enuresis and presence of dysuria to rule out UTI.
- In the physical focus on general exam,abdomen exam looking for masses&tenderness,look in the back for dimpling which indicated bladder dysfunction.

Management

- Mono-symptomatic enuresis will resolve spontaneously but 1% will remain wetting as adults.
- Urotherapy:decrease fluid intake in the night,decrease salt and sugar intake,avoid caffeinated drinks.
- Use star chart with days of being dry to motivate the child.
- If previous measures fail we can try desmopressin which is ADH.
- Enuresis alarm.



Assessing a patient with CKD

- Calculating the eGFR by the schwartz formula (.41*height/creatinine),see the KDIGO chart to know the stage.
- What do you need to investigate in them :
- Growth parameters
- Signs of anemia,pallor,murmurs,earthy colour of CKD(low erythropoietin)
- Hypertension.
- Electrolytes(hyperkalemia refer to a dieticien,hyperphosphatemia due to decreased excretion managed by dietary restrictions or calcium bicarb,hypocalcemia,hyperparathyroidism because of low calcium and low vitamin d alpha 1 hydroxylase deficient,needs replacement with one alpha vitamin d,hyponatremia may need salt replacement.
- Look for metabolic acidosis and treat with sodium bicarb.

UTIs - a child with dysuria

- In neonatal period males are more affected because of uncircumsission.
- Later on females are more affected.
- Ask about :
  - Associated urinary symptoms: urgency,frequeny,hematuria,abdominal or loin pain,fever
  - Family history of stones,dietary habits
  - History of vaginal discharge,itching.
  - Bowel habit.
- Physical exam:general for fever,renal angle tenderness and suprapubic tenderness,genitalia for stenosis,adhesions,discharge,back for dimple.
- Differentials:utis including urethritis,cystitis,pyelonephritis,vulvovaginitis,chemical irritation and stones.

Presentation

neonates: fever , sepsis,hypoactivity,Failure to thrive,prolonged jaundice

■In children :fever ,vomiting,abdominalpain

■Urinary symptoms:dysuria,frequency,urgency,new onset day or night time incontinenance,hematuria,smelly urine.

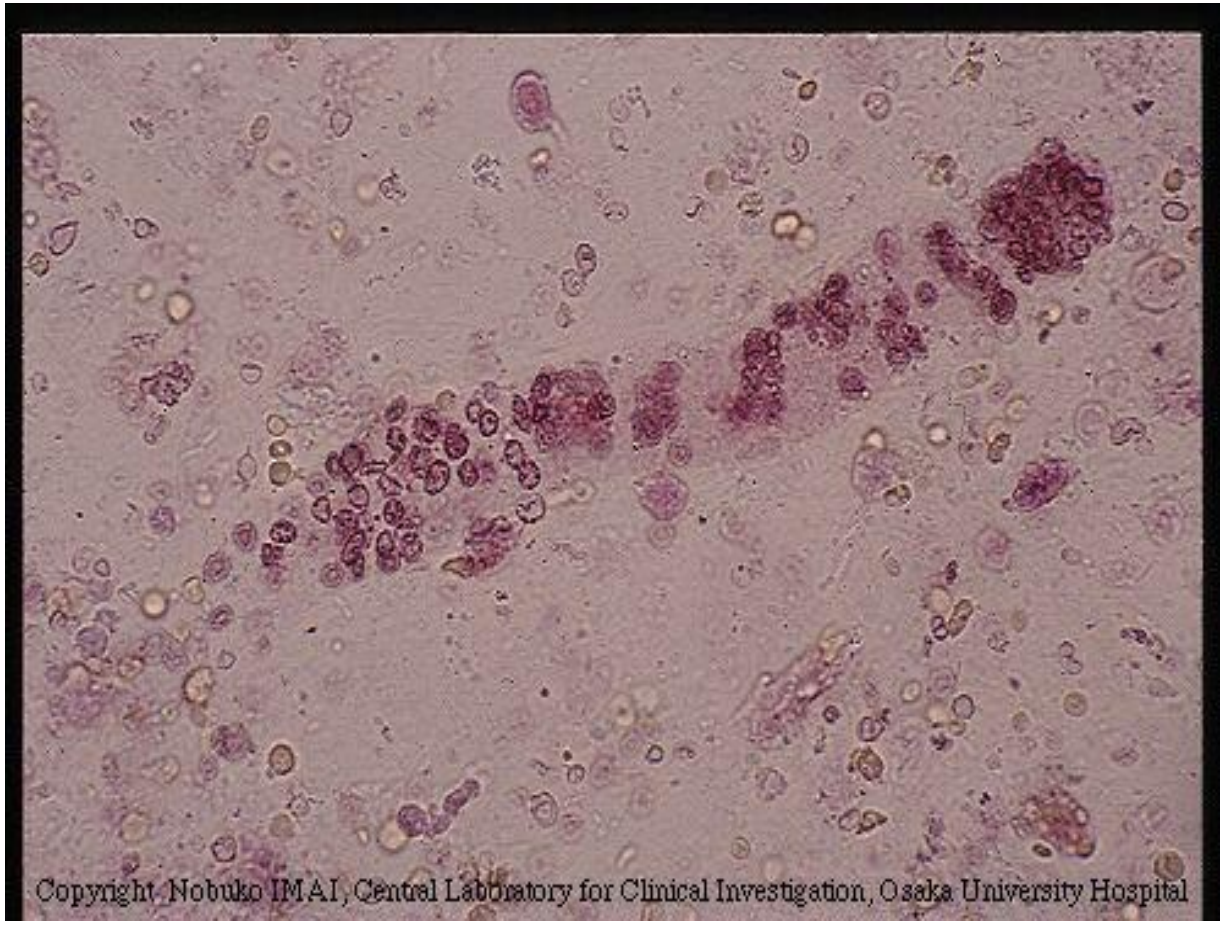
- 1-Cystitis urinary symptoms ,Low grade fever
- 2.Pyelonephritis :Loin pain, high grade fever,raised inflammatory markers,DMSA is gold standard,showsdecreased uptake
- Does this child have UTI?Temp>39,for>48 h,with no focus
- Diagnosis is by culture and microscopy,negative urinanalysis with symptoms doesn't rule out UTIs
- Dipstick for nitrite :(specific but not sensitive),if negative doesn't rule out UTI
- CRP, ESR,WBC,indicate pyelonephritis
- Most causative organism is E.coli
- Treatment is empirical antibiotic,3rd generation cephalosporin

Approach to red urine

- Causes of red urine(RBCS,hemoglobin,myoglobin,drugs foods and certain dyes)
- Things to ask in the history :
- Colour,bright red indicates non-glomerular causes,tea coloured indicates glomerular causes.
- Timing in association to voiding, at the beginning or early its urethral cause at the end or terminal its bladder cause
- Presence of clots indicates non-glomerular cause,absence of clots indicates glomerular cause
- Presence of lower urinary symptoms,flank or abdominal pain to rule out UTIs.
- History of seizure or trauma to rule out rhabdomyolysis.
- History of jaundice or pallor to rule out intravascular hemolysis.
- History of URTI(if latency period 2-3 weeks indicates PSGN,2-3 days IgA nephropathy)
- History of frothy urine,periorbital edema,decreased urine output to rule out glomerular causes.
- History of skin rash or joint swelling to rule out systemic diseases that cause hematuria such as SLE,HSP.
- Personal previous similar history.
- Family history of renal diseases,hematuria or deafness.
- History of taking any drugs,certain food or pigments porphyria,beets,rifampicin

RBC casts

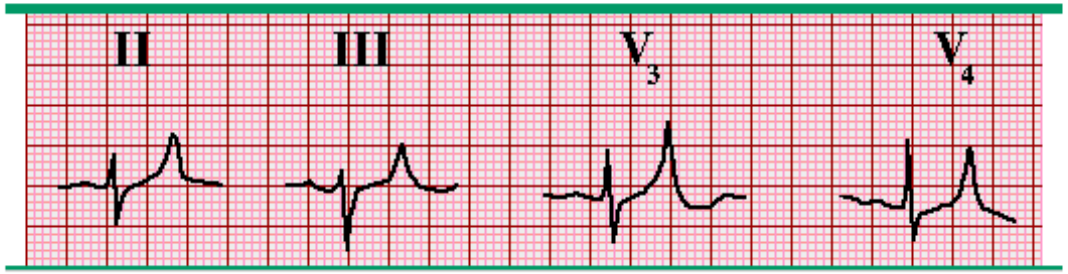
- Casts is not the same as clots,clots are for nonglomerular causes but rbcs casts are glomerulonephritis until proven otherwise.
- PSGN:manifested as nephritic syndrome(hematuria,edema, oliguria,HTN,azotemia)



Diagnosis&Managemet

- Diagnosis of PSGN: low c3,high ASO titers
- Manage the hypertension which is caused by volume overload by furosemide.
- Fluid restriction.
- Managing the hyperkalemia (calcium gluconate,potassium restriction,if its is resistant then dialysis is indicated).
- If metabolic acidosis present treat with sodium bicarb,if resistant dialysis is indicated.

ECG showing peaked T waves in hyperkalemia



A tall peaked and symmetrical T wave is the first change seen on the electrocardiogram (ECG) in a patient with hyperkalemia.

- Prognosis
  acute kidney normalizes within a week, proteinuria normalizes in few months,microscopic hematuria resolves in few months, complement goes ack to normal in 6-8 weeks,rare recurrence and there is no long term sequelle.

Distinguishing extraglomerular from glomerular hematuria

	Extraglomerular	Glomerular
Color (if macroscopic)	Red or pink	Red, smoky brown, or "Coca-Cola"
Clots	May be present	Absent
Proteinuria	Usually absent	May be present
RBC morphology	Normal	Dysmorphic
RBC casts	Absent	May be present

RBC: red blood cell.

Gastroenterology
return to the modules in the dossier

- Milk formulas:
- Cows milk protein-semilac,Enfamil AR
- Soy:prosobee
- Casein hydrolysate:nutramigen,alimentum,pregestimil
- Aminoacid:neocate

Hepatitis b

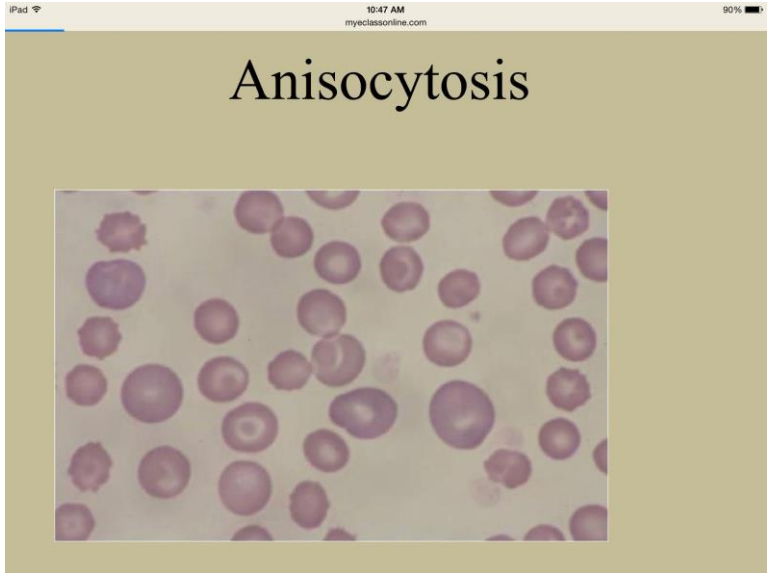
	HBsAg	Anti-HBs	Anti-HBc
Susceptible	Negative	Negative	Negative
Vaccinated	Negative	Positive	Negative
Past Infection	Negative	Positive	Positive
Acute Infection	Positive	Negative	IgM Positive
Chronic Infection	Positive	Negative	IgG Positive

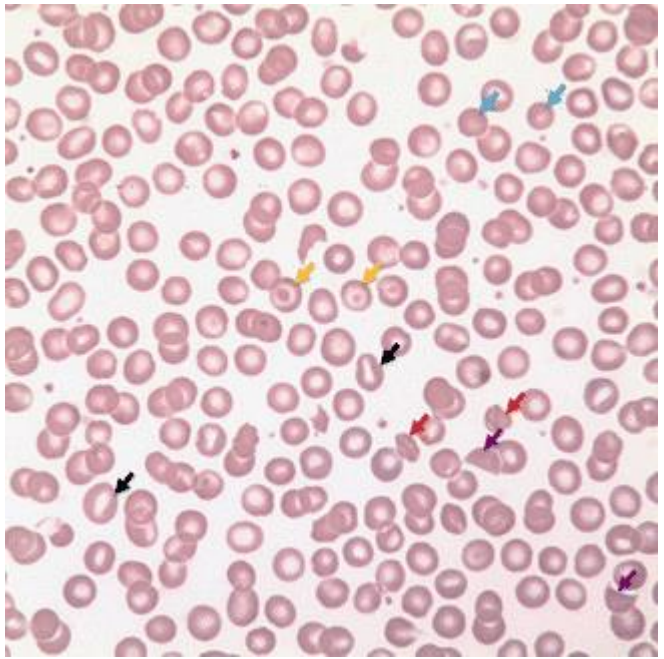
HEMATOLOGY

- Hg low
- See MCV normal is 80-100
- MCV low(microcytic anemia)<80,differentials include:iron deficiency anemia and thallassemias.
- MCV normal differentials include:hemolysis,sickle cell disease,anemia of chronic disease.
- MCV high >100(macrocytic anemia differentials include:folate deficiency and b12 deficiency.

Iron deficiency anemia(hypochromic microcytic anemias)

- RBC indices: low Hgb but high rbcs number,low MCH,low MCV,but high RDW
- Iron studies shows:low serum iron,low ferritin,but high TIBC.
- Peripheral blood smear shows anisocytosis(different in size),poikilocytosis(abnormally shaped RBCs).



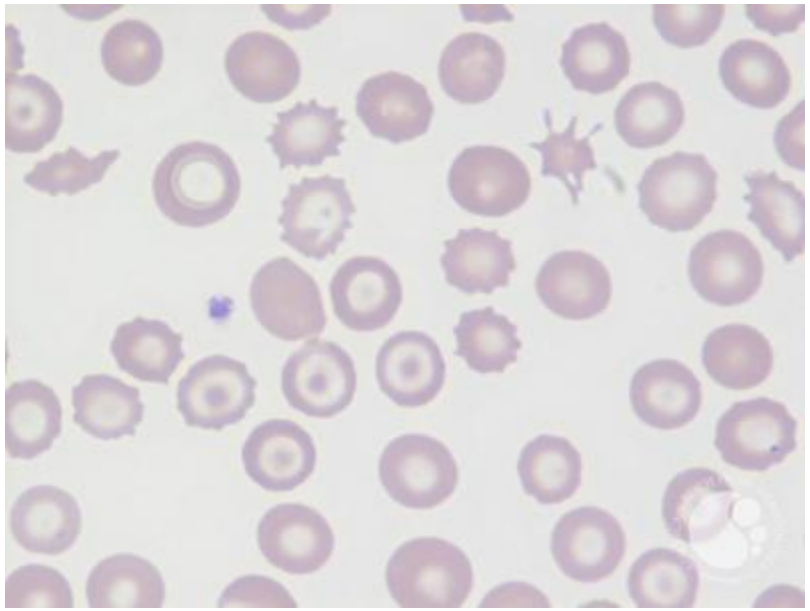


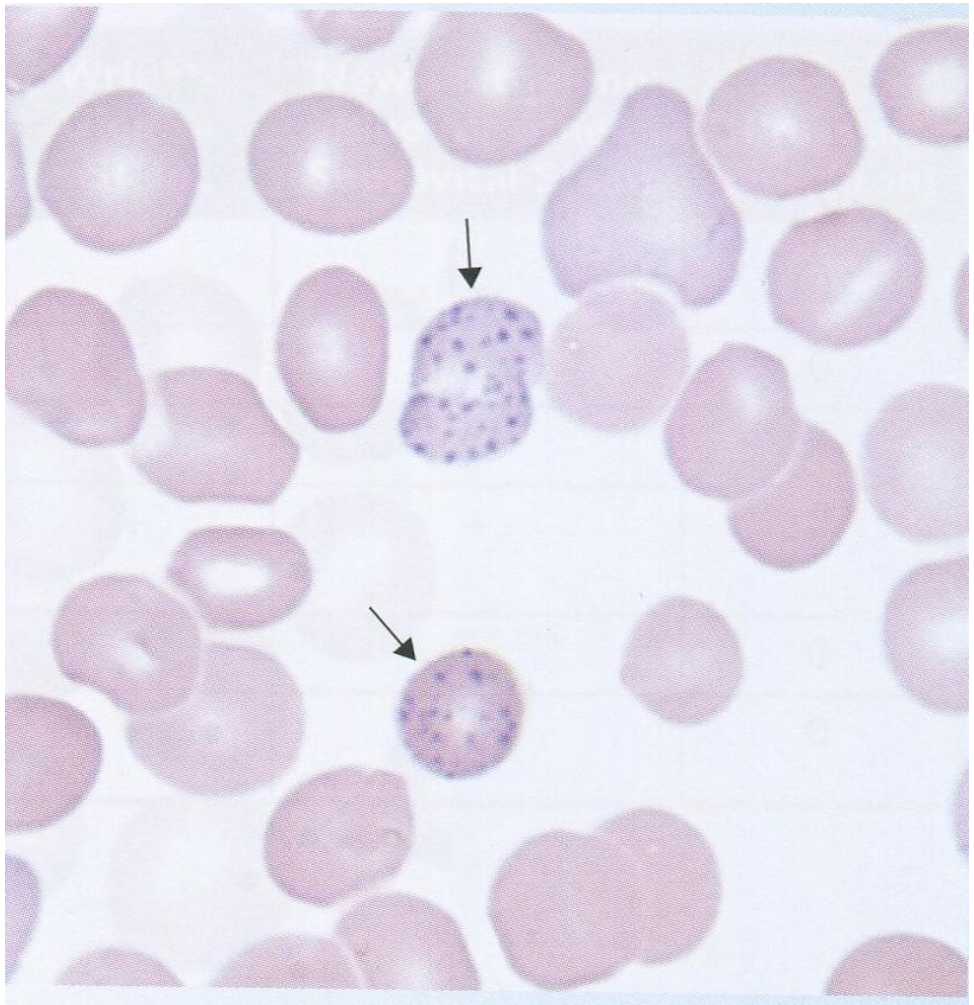
Thalassemias(hypochromic microcytic anemia)

- RBC indices:low MCH,low MCV,and low or normal RDW.
- As a general rule MCV/RBCS above 13 its iron deficiency anemia,below 13 its thalassemia.
- Alpha thalassemias,no alpha chains(diagnosis is by genetic sequencing)**,HgH disease 3 gene deletion** they have **extravascular hemolysis**(elevated LDH,splenomegaly,indirect hyperbilirubinemia) **or Hgb barts 4 gene deletion hydrops fetalis**.
- Beta thalassemia,no beta chain,diagnosis is by electrophoresis,increased HgF,HgA2.

Beta thalassemia major or cooley's anemia

- Findings on blood smear aren't specific
- Basophilic stippling
- Target cells





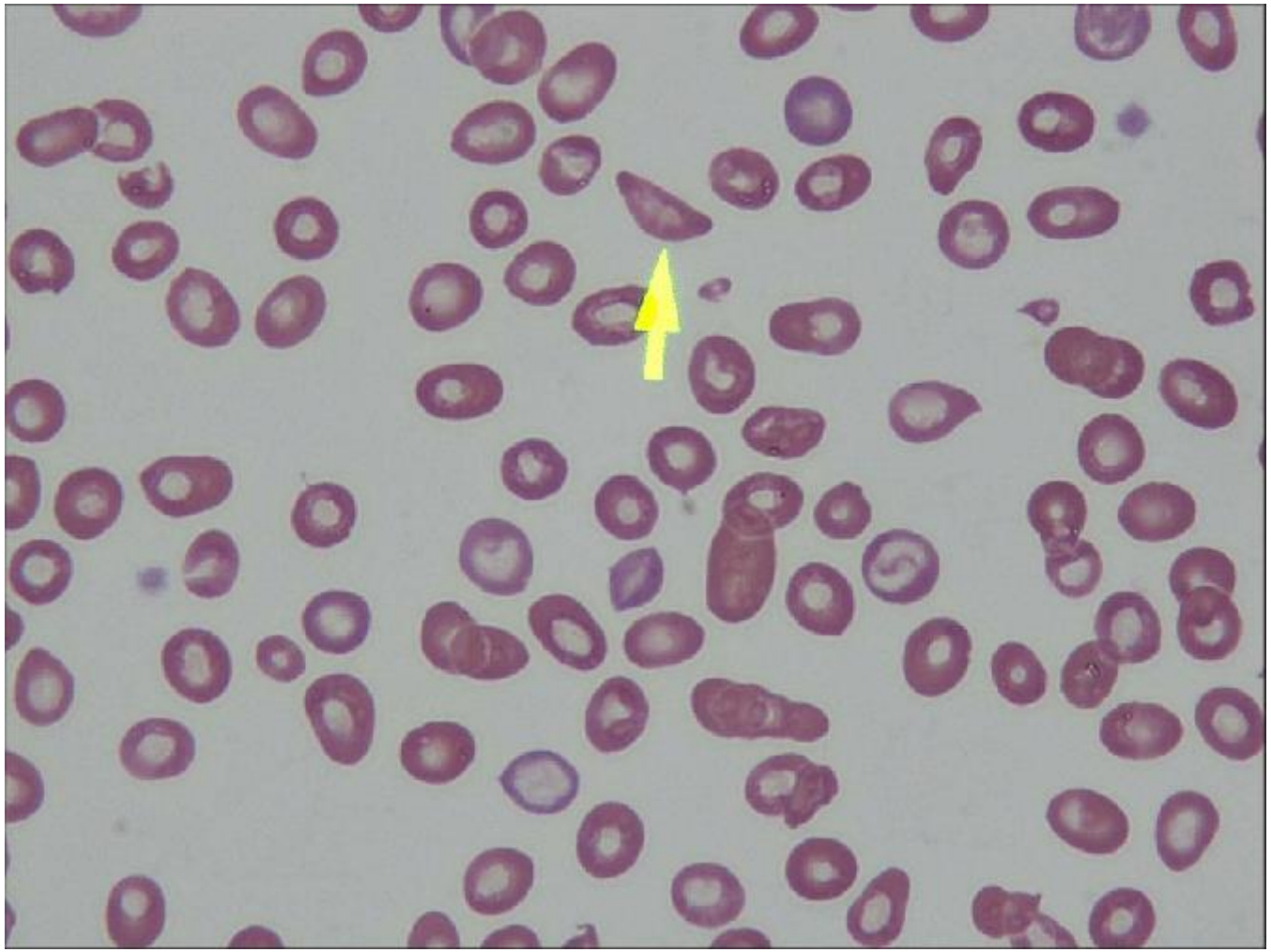
Anemia of chronic diseases (normocytic normochromic anemia-nonhemolytic)

- RBCs indices:normal MCV and MCH.
- Iron studies:low serum iron,normal TIBC,high ferritin.
- If the normocytic normochromic anemia + low erythropoietin its renal failure,+abnormal bone marrow its aplastic anemia.
- Nofindings on blood smear.

Normocytic normochromic anemia-hemolytic

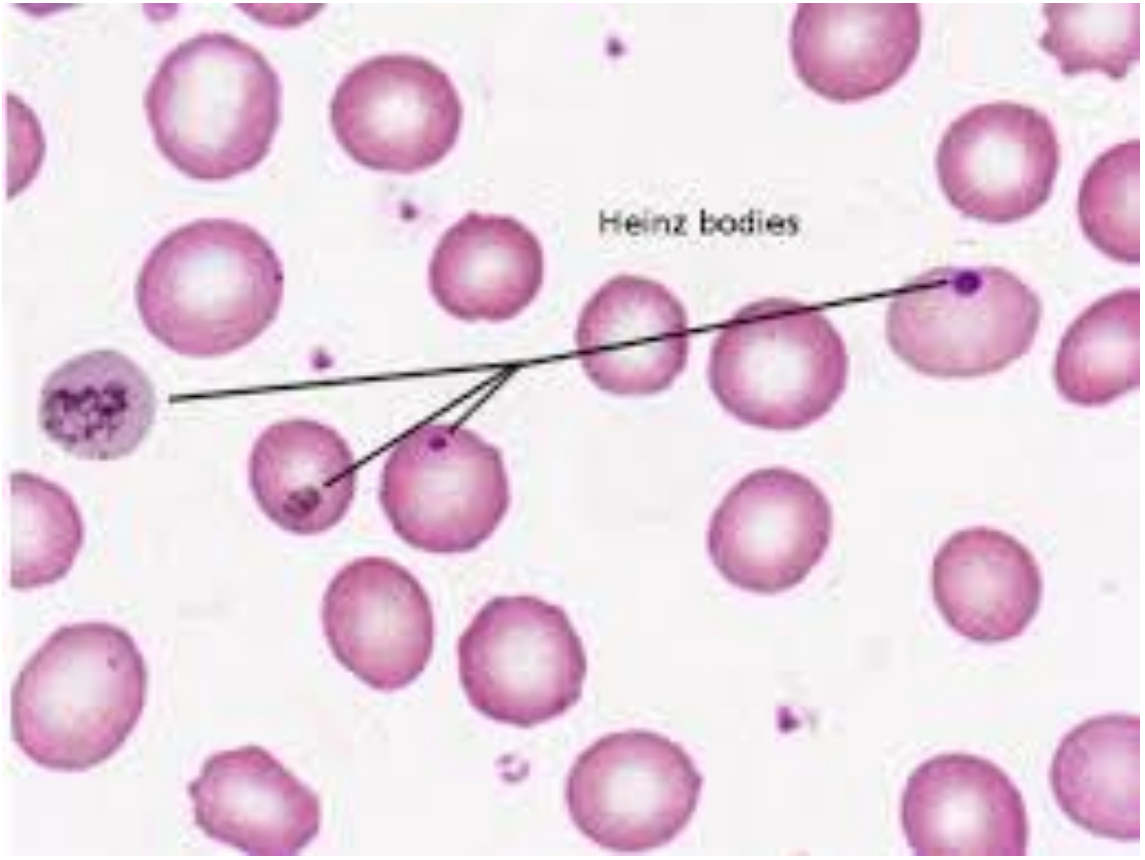
- Intrinsic hemolysis: G6PD,SPHEROCYTOSIS,SICKLE CELL DISEASE,HbC disease.
- Laboratory Findings :
  - Normocytic anemia
  - High LDH
  - Urine positive for Hgb and hemosiderin(intravascular)
  - High indirect bilirubin
  - High reticulocyte count
  - Low haptoglobin(because its function is to bind to plasma free Hgb)

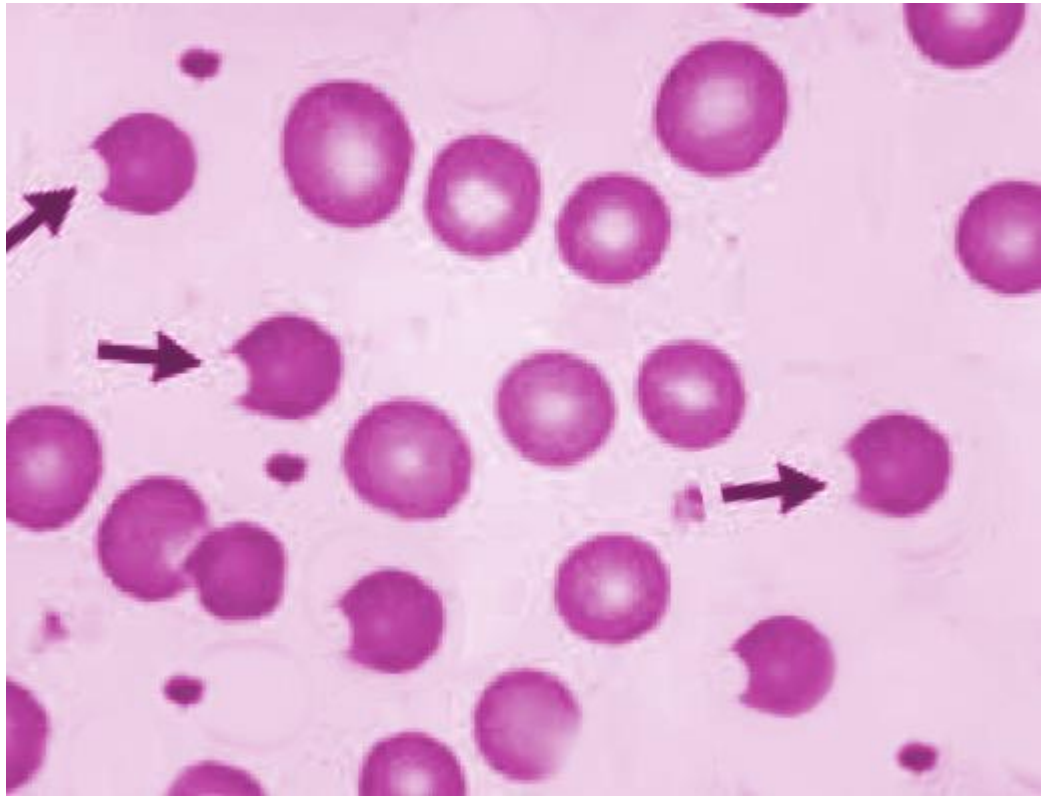
Sickle cell anemia



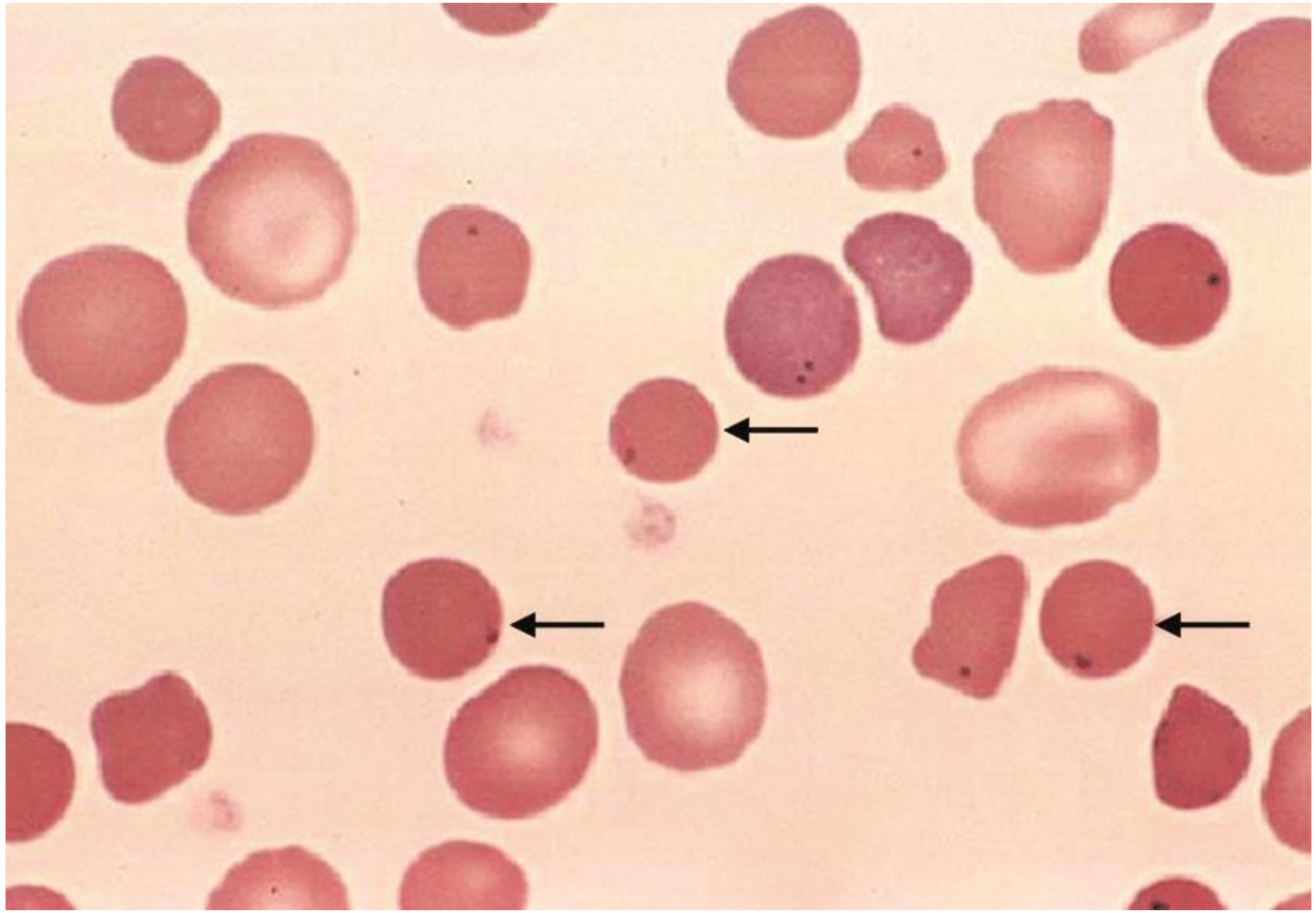
G6PD

- HEINZ BODIES,DEGMACYTES(BITE CELLS)



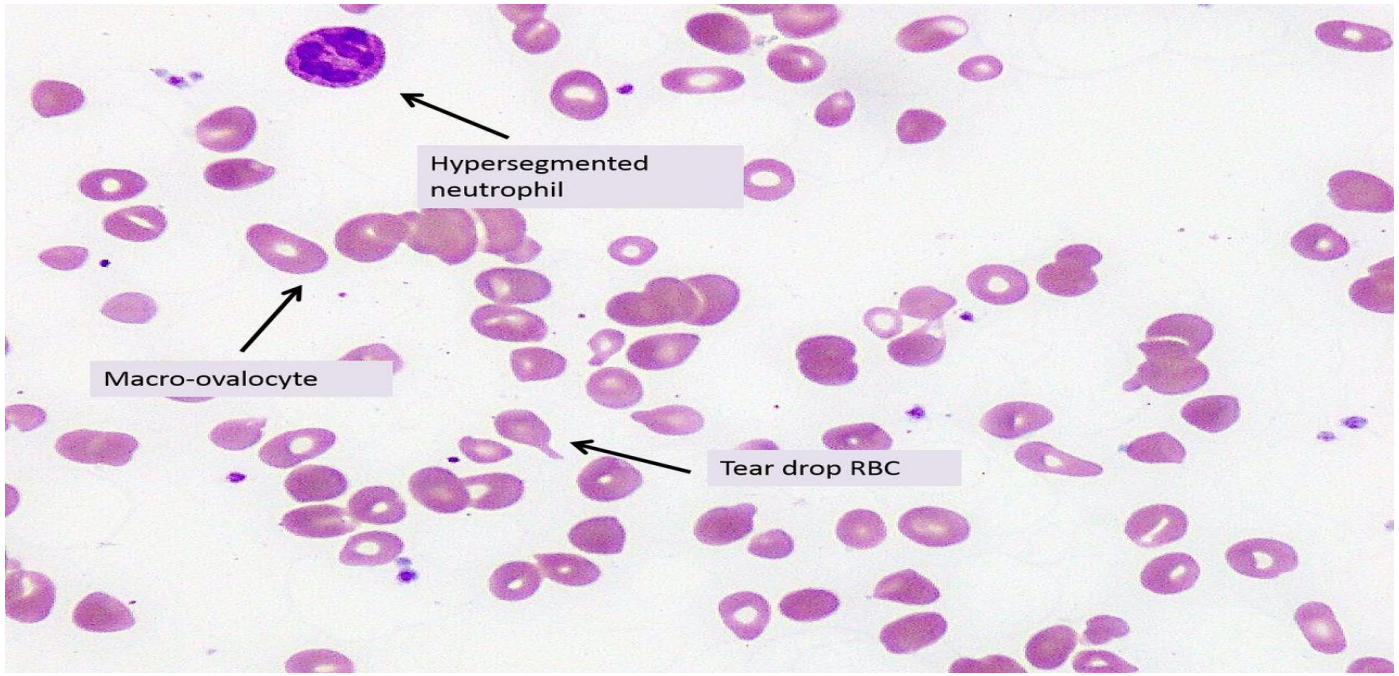


Spherocytosis



Macrocytic anemias

- B12 deficiency :High homocysteine,high MMA(methlmalonic acid),neurological symptoms.
- Folate deificiency:high homocysteine,but low MMA and no neurological symptoms.
- This blood smear is a finding of all megaloblastic anemias.



Dehydration & fluid therapy

Dehydration & Fluid therapy

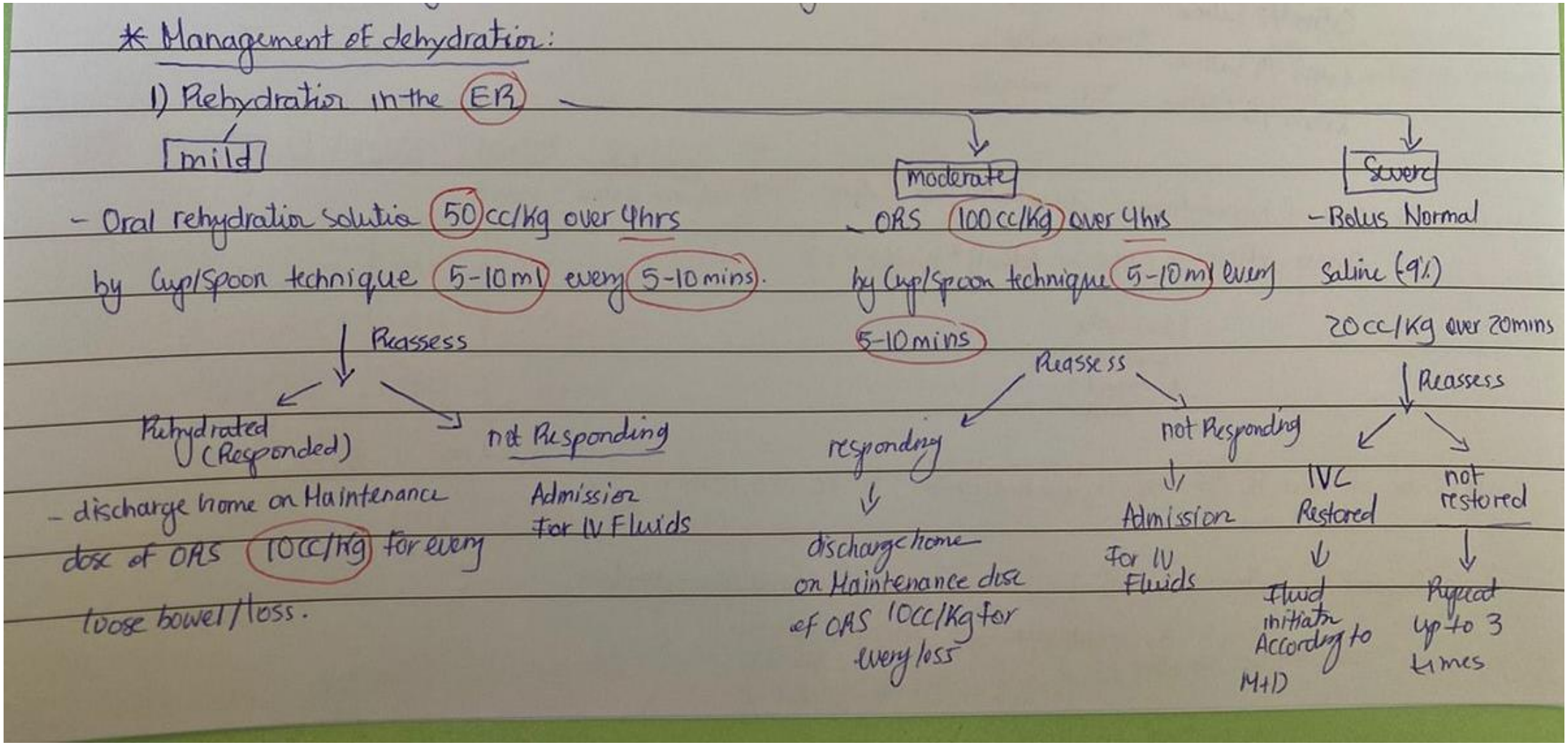
* dehydration is divided clinically into

	mild	moderate	Severe
	5%.	10%.	15%. → Young infants
& According to Sodium loss into	5%.	7%.	10%. → older children
	hyponatremic (hypotonic)	Isonatremic (Isotonic)	hypernatremic (hypertonic)

* Signs of Assessment of dehydration degree: (From head to toe)

① Vital Signs (hypotension[1], tachycardia[2], delayed pulse[3], ↓ Pulse Volume[4])
② LOC
③ eyes < ↓tears, Sunken eyes
④ dry mucus membranes
⑤ skin < ↓temp, delayed skin turger, Pale colour
⑥ delayed Cap refill
⑦ ↓Urine output.

mild → shows no signs only Sx (thirsty), history of losses.
moderate → any of the aforementioned signs except BP & CapreFill (nl)
Severe → hypotensive & Cap refill > 4seconds + over Signs are more prominent (e.g: mucus membranes, cracked)
In severe dehydration → BP could be very low or unrecordable


* Management of dehydration:
1) Rehydration in the ER
mild
- Oral rehydration solutio 50 cc/kg over 4hrs
by Cup/Spoon technique 5-10ml every 5-10 mins.
Reassess
Rehydrated (Responded)
- discharge home on Maintenance dose of ORS 10cc/kg for every loose bowel/loss.
not Responding
Admission for IV Fluids
Moderate
- ORS 100 cc/kg over 4hrs
by Cup/Spoon technique 5-10ml every 5-10 mins
Reassess
responding
discharge home on Maintenance dose of ORS 10cc/kg for every loss
not Responding
Admission for IV Fluids
Severe
- Bolus Normal Saline (9%)
20cc/kg over 20mins
Reassess
IVC Restored
Fluid initiate According to M+D
not restored
Repeat up to 3 times

* Indications for hospitalization:

1) Severe dehydration
2) mild/moderate dehydration not responding to ORS.
3) ~ ~ not tolerating Oral intake.
4) Underlying condition/disease complicating the management
5) not certain about the Dx.

* Calculating IV Fluid needed + type of solution

Maintenance

	Water	Na^+	K^+
	1st 10kgs 100	2-3 mEq/Kg/day	1-2 mEq/Kg/day
	2nd 10Kgs 50		
	Rest of weight 20		

Deficit

Water	Na^+	K^+
= weight * % of dehydration * 10	84 mEq/L	30mEq per 1000cc loss

doesn't do anything with choosing the solution, cause its found in Ampullis Added to the IV line.

Deficit is 85mEq with every 1000cc lost

20% of K^+ found intracellularly average of 150

$\frac{20}{100} \times 150 = 30$ mEq/L

60% of Na^+ found in the extra-cellular compartment in an average of 140

$\frac{60}{100} \times 140 = 84$ mEq/L

* What determines our solution choice is the Sodium needed in the First (8hrs) and not the 24hrs

* Sodium Content of Solutions :

NS	154 mEq/L
G5W 1/2 Saline	77 mEq/L
G5W 1/4 Saline	38 mEq/L
G5W 1/5 Saline	33 mEq/L

* In the case of hyponatremic dehydration, for Sodium we also calculate

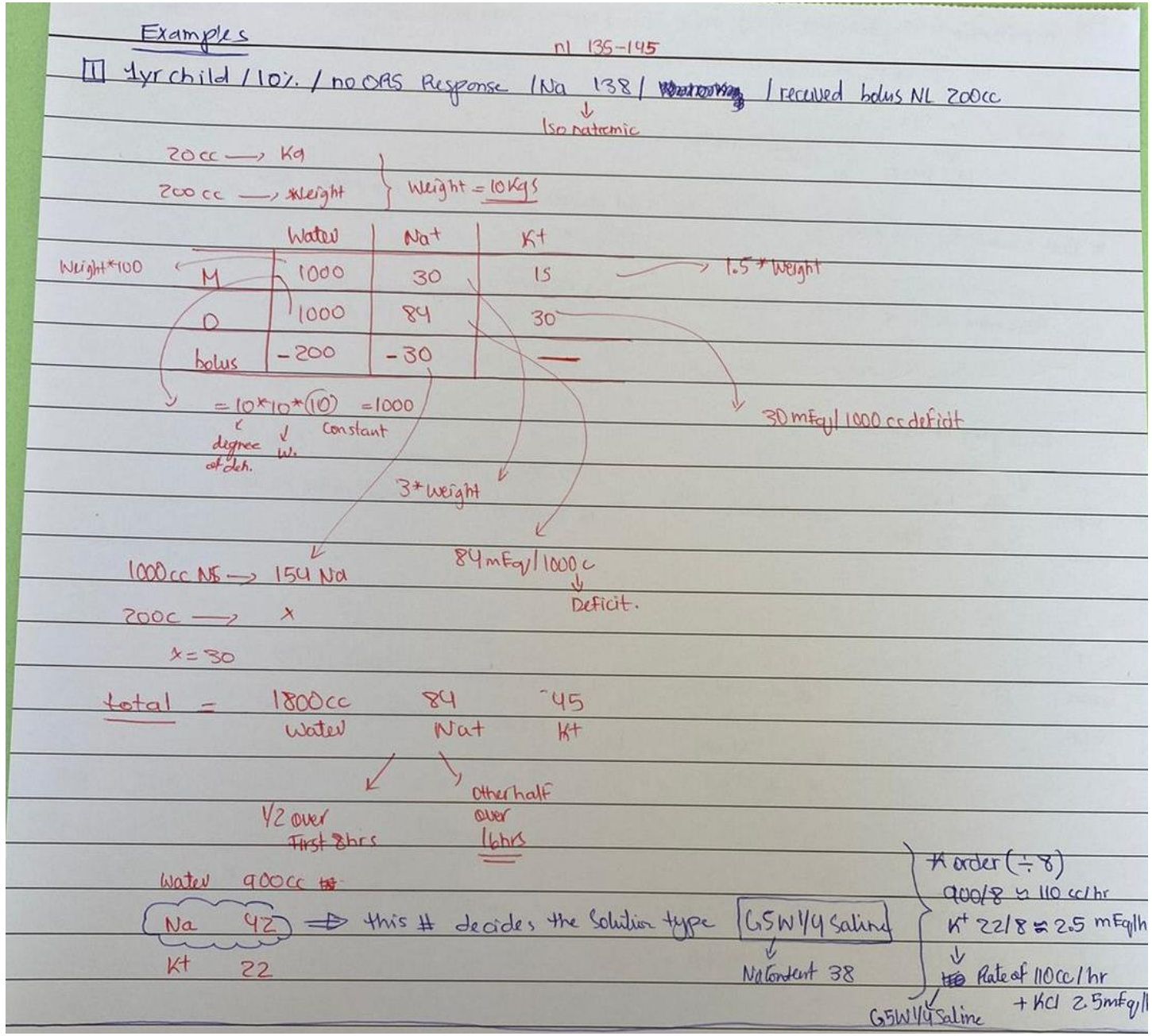
Excess deficit = (130 − Actual) * Weight * .6

(130 ↙ Which is the lower limit to correct Sodium)

* Rate of Correction for hypo/hyper natremia is 12 mEq/24hrs = .5 mEq/1hr. ✓

* Rapid Correction of hyponatremia ⟶ Central pontine demyelination.

~~ ~ hypernatremia ⟶ Cerebral Edema.


Examples
nl 135-145
1 1yr child / 10% / no ORS Response / Na 138 / [redacted] / received bolus NL 200cc
Isonatremic
20cc → Kg
200cc → Weight
Weight = 10Kgs
Water | Na+ | K+
M 1000 | 30 | 15 → 1.5 * Weight
O 1000 | 84 | 30
bolus -200 | -30 | —
Weight*100
= 10*10*(10) = 1000
degree of deh. Wt constant
3*weight
30 mEq/1000 cc deficit
84 mEq/1000 c
Deficit.
1000cc NS → 154 Na
200c → x
x = 30
total = 1800cc Water 84 Nat 45 Kt
1/2 over First 8hrs
other half over 16hrs
Water 900cc
Na 42
Kt 22
this # decides the Solution type
G5W1/4 Saline
Na Content 38
order (÷8)
900/8 ≈ 110 cc/hr
K+ 22/8 ≈ 2.5 mEq/h
Rate of 110cc/hr
+ KCl 2.5mEq/l
G5W1/4 Saline

G5W1/4 Saline + KCl 2.5mEq/hr.

[2] 1yr child / 10kg / Na 115 / received 200 cc bolus NS

	water	Na+	K+
M	1000	30	15
O	1000	84	30
bolus	-200	-30	—
		+90	

hypo (Excess Na deficit

Excess deficit = (130-115)*10*.6 = 90

total 1800 / 174 / 45

~~First 8hrs~~

Water 900 cc
Na ~~87 mEq~~
K+ 22

G5W1/2 Saline (77)

order
900/8 = 110
~~87/8~~
22/8 = 2.5

G5W1/2 Saline
Rate of 110cc/hr
+ KCl rate of 2.5mEq/hr.

3 hypernatremic dehydration is the most severe & serious dehydration, the least one that shows signs

↳ should be managed in PICU

* equation 1.5*M
or
M + 1/2 D
} Same equation.

* Best Solution for this kind is G5W1/2 saline (but you can change solution depending on your reassessment)

Remember Rate of correction should be [12 mEq/24hrs / .5mEq/1hr] to Avoid Complications

Examples on Reassessment

↓

6pm Na = 170
10pm Na = ↑ 170

Correct Rate = 0/hr } either change to a less tonic solution (G5W1/4 Saline)
or ↑ Rate of infusion (more water / dilution)

↙

6pm Na = 170
10pm Na = 160

Rate of correction
2.5/hr
10 → 4 hrs
x → 1hr
} Change to a more tonic solution (NS)
or ↓ Rate of infusion (less dilution)

↙

6pm Na = 170
10pm Na = 168

Rate of correction
.5/hr ✓

→ Continue with the same solution & Same Rate