Say hi to Pediatrics

Colour code:

- Most common
- Most serious
- Gold standard
- First line
- specific & sensitive
- Diagnostic test

By: Sohib Shabatat

Topics:

Well newborn

* Neonatal screening:
→ From the infants heel on 3rd - 7th day of life
→ TSH, PKU, G6PD

* Primitive reflexes:
→ Mediated by spinal cord & brain stem
→ disappear at 4-6 months
1. Grasp 2. Suckling 3. Rooting
4. Moro:
- Absent: = Premature
= Birth trauma (ICH)
= CNS depression (narcotics)
- Asymmetric: = Erb's palsy
= Clavicular fracture
- Persestent: = CP (CNS damage)
= Mental retardation

* Delivery room management:
1. skin-skin contact: stabilize temp (30-60m)
2. Breast feeding ASAP within 1st hr
3. Newborn identification by bracelets
4. vit-k injection (IM) in the 1st hr

→ vit-k deficient bleeding:
time: after birth up to 2-12 weeks later
site: skin, mucosa, umbilical, ICH, circumcision site
Maternal risk: anti epileptic, anti-Tb
vit-k antagonist

* Newborn assessment:
1. Resuscitation
2. Gestational age and birth weight
3. Apgar score (done at 1, 5, 10 min)
4. Identification of infant
5. Anomalies
6. Growth assessment:
W: 2.5 - 4.25 kg (3.5 avg)
L: 46 - 56 cm (50 avg)
HC: 33 - 38 cm (35 avg)
→ weight loss in 1st week must NOT exceed 10-12 %
→ Birth weight regain after 2 weeks

* Apgar score: (N) 7-10 in 5 min
→ To asses infants transition from intra to extra-uterine life

	Sign	2	1	0
A	Appearance (skin color)	Normal over entire body	Normal except extremities	Cyanotic or pale all over
P	Pulse	>100 bpm	<100 bpm	Absent
G	Grimace (reflex irritability)	Sneezes, coughs, or vigorous cry	Grimaces تكشيرة	No response
A	Activity (muscle tone)	Active	Arms and legs flexed	Absent
R	Respirations effort	Good, crying	Gasping, irregular	Absent

* Stooling: Meconium Pass before 48 hrs
→ delayed: 1. imperforated anus
2. Hirschsprung
3. Meconium plug syndrome
* Urine: Pass before 24 hrs
4-6 times / day
→ don't discharge until he pass stool & urine
* V. discharge: withdraws bleed from maternal hormons
* U. cord: fall of in 10-14 days
* Delayed closure of AF: hypothyroid / Rickets hydrocephalus
* Delayed closure of PF: Down syndrome
* risk for hypoglycemia:
1. Mother with DM 3. Preterm
2. SGA or LGA 4. sick infant
* don't retract uncercumsized genitalia
* Nothing in sleep area

Neonatal sepsis

* sepsis : Bacteremia w/ SBS in a <28 day infant
* Bacteremia: +ve culture in a well looking baby

Types	early onset sepsis	Late onset sepsis (LOS)
Source of infection	Maternal birth canal	Hospital or community
Time of infection	Before or during delivery	After delivery
onset	Before 72 hrs	After 72 hrs
Commonest organisms	Group B strep (GBS) E-coli	Coagulase -ve staph Staph aureus Klepsella Pneumonia Psuedomonas } long time in NICU
Risk factors	- Maternal GBS colonization - Chorioamnionitis - PROM - Preterm birth - Multiple gestation	- prematurity - low birth weigh - delayed feeding - invasive procedures - Prolonged catheter
Admission	symptomatic: to NICU + Empiric abx + diagnostic tests well appearing: observe 48 hr	admission + diagnostic tests + empiric abx
Treatment	Ampicillin + Gentamycin - Cephalosporin if meningitis	Community: Ampicillin + gentamycin OR Ampicillin + Cefotaxime & 3rd cephalo if meningitis Hospitalized since birth: Vanco instead of ampicillin Meropenem for E-coli

* Clinical features:
→ non-specific (↑ index of suspicion)
1. hypothermia or fever
2. tachypnea or apnea
3. hypotension
4. Poor cry & Feeding, lethargy

* The most common reason for admission to NICU is to rule out sepsis

* Investigations:
1. Blood culture
2. WBC (Poor value)
3. Absolute Neutrophil count (ANC):
→ Neutropenia: < 1800 at birth
< 7800 at 12-14 hrs
4. Absolute Band count (immature Neutrophile)
→ > 10% diagnostic
5. ratio of immature to total (I/T):
→ > 0.2 is suspicious
6. CRP: late marker, not specific
useful to monitor response
7. Procalcitonin (PCT): early, specific for bacterial sepsis & correlates with severity
8. Neutrophil CD64: sensitive
9. LP: whenever B.culture is +ve
10. Urine culture: for LOS
11. X-ray: for apnea, distress, NEC

* Supportive Management:
1. thermo-neutral environment
2. O_2
3. Fluids
4. Fresh Frozen plasma & Packed RBC

* Complications:
1. spread of inf: meningitis, hepatitis..
2. septic shock: ↓BP, Poor perfusion
3. Acute renal failure
4. DIC

Key features of congenital infections*	
Toxoplasmosis	• Chorioretinitis • Hydrocephalus • Diffuse intracranial calcifications
Syphilis	• Rhinorrhea • Skeletal anomalies • Desquamating rash (palms/soles)
Rubella	• Cataracts • Heart defects (eg, PDA) • Sensorineural hearing loss
Cytomegalovirus	• Periventricular calcifications • Microcephaly • Sensorineural hearing loss
Herpes simplex virus	• Vesicular/ulcerative rash

*Nonspecific findings include growth restriction, jaundice, hepatosplenomegaly & blueberry muffin rash.
PDA = patent ductus arteriosus.

Prematurity

*Preterm: is the one delivered before 37 week

* Risk factors:

→ Commonly: Idiopathic or miscalculation

1. Maternal: - uterine anomalies (Bicornat)
   - cervical incompetence
   - PROM
   - Pre-eclampsia, DM
2. Fetal: - congenital anomaly
   - congenital infection
   - Multiple pregnancy

* characteristics:

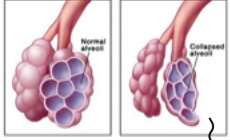
1. General: - Hypoactive, weak cry
   - Poor suckling
2. Skin: reddish with ↑ lanugo hair
3. hypotonia: - frog like position
   - head lag
4. underdeveloped ears
5. small breast bud, no nipple
6. Underdeveloped genitalia
7. Incomplete sole creases

* Complications:

1. CNS: - ICH
   - hypoxic ischemic encephalopathy
2. Eyes: - Retinopathy of prematurity
3. CVS: - PDA
4. RS: - RDS
   - Apnea
   - aspiration pneumonia
   - Broncho-pulmonary dysplasia
5. GI: - Poor suckling & swallowing
   - immature bowel function
   - recurrent constipation
   - NEC
6. Hemato: - Anemia
   - Bleeding
7. Metabolic: - hypoglycemia
   - hypocalcemia
   - hypothermia
   - hyperbilirubinemia

* start breast feed at 33-34 week as coordination between suckling and swallowing develop

*NB: Testicular descending occur between 31-34 week of guestation



Respiratory distress syndrome

** Type 1 RDS:

→ hyaline membrane disease

* Incidence:
common in premature newborn

* Pathophysiology:
→ Surfactant deficiency → alveolar collapse → right to left shunt → hypoxia & metabolic acidosis → P.HTN.

* Clinical presentation:
Grade 1: Tachypnea & nasal flaring
2: Intercostal & subcostal retraction
3: Expiratory grunting
4: cyanosis, ↓ consciousness

→ Auscultation: ↓ air entry

* investigation:
1. CXR (diagnostic): - Air bronchogram
- Ground glass
- white lung
2. CBC, ESR, CRP
3. ABG: hypoxemia (↓PaO2) & acidosis

* Management:
→ supportive: - IV fluid & inotrops
- NPO (aspiration)
→ CPAP
* surfactant need 36-48 hrs after birth to develop

* ↓O2 Sat in intubated child:
1) Displaced tube
2) Obstructed tube (by secretions)
3) Pnemothorax
4) Equipment failure

** Type 2:
→ Transient tachypnea of newborn

* Eitiology:
delayed clearence of lung fluids (wet lung syndrome) in C-section of Full term infants

* Clinically:
1. Tachypnea & mild retractions
2. resolve in 24 hrs

* CXR:
- ↑ perhilar vascularity
- Fluids in lung fissures
- Small Pluerat effusion

* Tx:
- suction, physiotherapy
- O2, NPO

Neonatal hyperbilirubinemia

* Causes:

1. Physiological

2. Unconjugated:
   - → ↑ Production: - hemolysis → Rh - ABO, H. sphero, G6PD
     - infection
     - Polycythemia
     - cephalohematoma
   - → ↓ uptake: Gilbert disease
   - → ↓ conjugation: - Crigler - najjar
     - hypothyroid
     - Breast milk jaundice
   - → ↑ enterohepatic: - Bowel obstruction
     - Breast feeding jaundice

3. Conjugated:
   - Neonatal hepatitis
   - TORCH
   - Sepsis
   - biliary atresia

	Physiological jaundice	Pathological jaundice
Onset	2nd or 3rd days of life	first day of life
Rate of rise/day	Less than 5 mg % per day	More than 5 mg / day
Maximum level	Less than 15 mg	More than 15 mg
Type of bilirubin	Unconjugated mainly	Conjugated or Unconjugated
Duration	7 days in full term & 14 in preterm	Longer duration (> 2 weeks)
Treatment	Reassurance	Of the cause ± Photo or Exchange

* Risk factors:

1. F H : hemolysis , Maternal illness , drugs
2. Instrumental delivery

* Clinical exam:

1. Skin colour : yellow , green , Palor
2. head : cephalohematoma , Microcephaly
3. Umbilical infection , hernia
4. NSM
5. Absent reflexes

* Investigations:

1. Bilirubin (total & direct)
2. Blood group
3. CBC , blood film , retic count
4. Coomb's test , enzyme assay
5. CRP , ESR

* kernicterus:

→ Brain damage due to deposision of free unconjugated bilirubin in basal ganglia

* Clinically:

1. Early : Poor feeding , hypotonia , ↑pitched cry
2. Intermediate : Hypertonia
3. Advanced : Opisthotonus , apnea , seizure , coma
4. late : choreoathetosis , deafness , MR

* Tx:

Types	1. Phototherapy	2. Exchang transfusion
Idea	1. 450 nm , 45 cm 2. Protect eyes & testes	
Indications	1. Bilirubin 15 - 20 2. Before and after exchange 3. Prophylactic in hemolysis	1. Bilirubin > 25 mg/dl 2. Early in hemolytic disease 3. Bilirubin ↑ > 0.5 - 1 / hr 4. F H of kernicterus or sever erythroblastosis
contraindication	Cholestasis (bronze baby syndrome)	
Side effect	1. Hyperthermia 2. Dehydration 3. loose stool 4. skin rash 5. retinal damage	1. Thrombosis , embolism 2. Infections 3. Hypo glycemia , Ca Hypo thermia Hyper kalemia 4. Volume over load → arrhythmia & HF

3. IvIg : reduce the need for exchange

Short stature

* Growth phases :

1) Intra-Uterine :
   → The most ↑ in length is in the 2nd TM
   → Birth size reflect intrauterine environment

2) Infantile :
   → 0-2 yrs
   → Rapid decelerating
   → depends on nutrition, insulin, ILGF

3) childhood :
   → 2-12 yr
   → slower, decelerating (4-6 cm/yr)
   → depends on Growth hormone & thyroxine

4) pupertal :
   → 12 y - Final height
   → accelerated
   → depends on sex steroid estrogen

Stage	Boys	Girls
1	childhood phase	childhood phase 4-6 cm/yr
2	still (slower than G1)	↑ height velocity 9cm/yr
3	↑ height velocity	Peak height velocity 10cm/yr
4	Peak height velocity	slowing (onset of menarche)
5	further growth → final height	Adult height

* Short stature : height < 2 SD

* growth failure : failure to maintain height velocity that is appropriate for age :

1) curve deviated down across 2 major centiles

OR : 2) Age 2-4 yr : HV < 5.5 cm/yr
4-6 yr : HV < 5 cm/yr
> 6 yr → boys < 4 cm/yr
→ girls < 4.5 cm/yr

* Adult height depends on :
1) Mid-Parental height 2) Bone age

* causes of short stature :

1) Normal genetic : - child short normal
- Parent short normal
- Not delayed Bone age
- become short adult

2) Constitutional :
- short normal but younger than his age
- Parent NOT short but maybe in their childhood
- Bone age IS delayed
- Final height in lower half of target

3) Dysmorphic syndrome : - Turner
- Noonan
- Down

4) SGA : Asymmetrical → @ Final height
→ catch up
symmetric → short adult
→ NO catch up

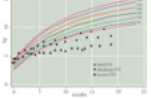
5) skeletal dysplasia : Achondroplasia
6) Chronic disease : celiac, CKD, CF
7) social & emotional
8) Endocrine : ↓ Growth hormone
↓ thyroxine
↑ cortisol
precocious puberty

* Investigations :

1) CBC, Film, ESR, LFT, KFT, lytes
2) karyotype
3) Thyroid, IGF-1, Prolactin, cortisol
4) TTG, urine analysis & culture
5) Bone age
6) Endocrine stimulation testing

* Growth hormone indications :

1) Growth hormone deficiency
2) Syndromes (Turner, Noonan..)
3) SGA no catch up in 4 years
4) CKD



Failure to thrive

*Growth Charts:

Chart	CDC	WHO
origin	National (US)	International (6 countries)
studies	Cross sectional	Longitudinal
cutoff value in centiles	5th and 95th	3rd and 97th
Recommendation	from 2-19 yrs	from 0-24 months
Note		Breast fed infants

*Failure to thrive:
→ in ability to maintain the expected rate of growth

Type	characters	Causes	Nutritional status
1	↓ weight Ⓝ Length Ⓝ Head circumference	1. Inadequate caloric intake 2. Exessive loss of calories 3. Increased metabolic demand	Wasting: BMI < 3rd
2	↓ weight ↓ length Ⓝ head circumference	1. constitutional growth delay 2. Genetic short stature 3. Hypothyroidism 4. Growth hormone deficiency 5. Hypopituitarism 6. Chronic malnutrition	Stunting: Height < 3rd BMI Ⓝ
3	↓ weight ↓ length ↓ Head circumference	1. Congenital infections 2. chromosomal abnormalities 3. Prenatal exposure to toxins	

*Nutritional rehabilitation:

→ we prefer GI tract over parentral
→ Most physiologic to least:
1. increase caloric density
2. oral supplement
3. gastric bolus
4. gastric continuous
5. jejunal continuous

*Complications of nutritional support:

1. Refeeding syndrome: - ↓ k
   - ↓ Mg
   - ↓ PO4
2. enteral support: - Tube malposition
   - Irritation or infection of tube site
3. Parentral support: - Infection (↑risk)
   - Metabolic derangement
   - Mechanical complications

CDC Growth Charts 5th and 95th percentile		
BMI-for-age	≥ 95th	Obesity
BMI-for-age	≥ 85th and < 95th	Overweight
BMI-for-age	< 5th	Underweight
Stature-for-age	< 5th	Short Stature

TABLE 5.1 Rules of Thumb for Growth
WEIGHT
Weight loss in first few days: 5–10% of birthweight
Return to birthweight: 7–10 days of age
Double birthweight: 4–5 months
Triple birthweight: 1 year
Daily weight gain:
20–30 g for first 3–4 months
15–20 g for rest of the first year
HEIGHT
Average length: 20 in. at birth, 30 in. at 1 year
At age 4 years, the average child is double birth length or 40 in.
HEAD CIRCUMFERENCE (HC)
Average HC: 35 cm at birth (13.5 in.)
HC increases: 1 cm per month for first year (2 cm per month for first 3 months, then slower)

SHORT STATURE

	Rate	Bone age
Familal	normal	normal
Constitutional	normal	abnormal
Chromosomal	abnormal	normal
Nutrition/GI/Endo	abnormal	abnormal

Developmental milestones

* Investigations for delay:
Hearing / vision / thyroid / karyotype / Brain MRI
Gene test / Fragile X test / EEG if seizes

	Vision	Fine motor	Gross motor	Social / behavioral	Language
1 month			rises head momentarly when prone	begining to smile	
2 months	Follows object 180°		rises head sustained when prone	social smile listen to voice	
3 months	Accomodation convergence	open hand spontanuous reach objects	rises head above plane of body	sustained social contact	aah, ngah
4 months		reach objects & put in mouth	support weight / full head control sits w/ full truncal support	laughs loudly excited at sight of food	
7 months		transfer from hand to hand	sits with pelvic support rolls over	enjoys mirros Prefers mother	Pollysylabisc vowels
10 months		thumb index grasp release object grasped by others	sits alone / crawl Pulls to standing position walks holding furnature (cruises)	waves bay bay plays peeka boo responds to sound of name	few words besides mama & dada
1 yr	visual acuity visual field	release objects on demand	walks with hand held	Plays simple boys game adjustment of posture to dressing	
15 m		tower of 3 cubes	walks alone / crawl upstairs	hugs parent indicate desire by Pointing	Follows simple comands name familiar objects
18 m		tower of 4 cubes / scribbles imitates virtical stroke	Runs stiffly walks upstairs one hand held	feeds self kisses parent with pucker	10 words, name pictures identify Parts of body
24 m		tower of 7 cubes imitates horizontal stroke	Runs well up and down stairs on step at a time	listen to stories with pictures Handles spoon well	Put 3 words together
30 m		tower of 9 cubes	upstairs alternating feets	Pretends to play	knows his full name
3 yr		draws circle / 10 cubes	tricycle / stand on 1 foot momentarly	plays with children / washes hand unbutton clothing / put on shoes	knows his age & sex
4 yr		draws cross, sequare, man	hops on 1 foot / throw ball	goes to toilet alone	tells story / count 4 pennies
5 yr		draws triangle	skips	dress & undress	count 10 pennies

Infant feeding & formula

* differences in 100 ml between cow's and breast milk

Type	Breast milk	cow's milk
Calories	67 kcal	67 kcal
Protiens	1.2gm whey : casien is 60 : 40 lactoferrin present	4.5 whey : casein is 20:80
carbohydrates	6.7gm lactose	4 gm lactose
Fats	3.5 gm small fat globule less volatile F A cholestrol and FA	4.5gm large fat globule High volatile FA lower cholestrol & FA
Minerals	1.5 : 1 Ca : PO4 better iron absorption	1 : 1 Ca : PO4
Vitamins	A and B Adequate C and D Deficient	A and B Adequate C and D Deficient
water	87.5 %	87.5 %

* Calculate the feeds:

1) Number : 6-8 times / day

2) Amount : Age : 1 month : 60-90 ml / feed
2-3 m : 120-130 ml
4-5m : 140-150 ml
6-7m : 160-170 ml
weight : 150 ml / kg / day → 25 ml / kg / feed

3) small spoon (4g) → 30 ml H_2O, large (8g) → 60 ml H_2O

formula type	Modifications	Indication / Recommendations
lactose free	lactose is replaced by sucrose or glucose	1. Galactosemia 2. lactose intolerence 3. Malabsorption following diarrhea
Iron fortified cow milk		1. Best for infant from 0-12 months (Normal healthy infants)
Soy-Based	1. Soy protion + methonine 2. Vegitable oil 3. vitamins, minirals, iron 4. Sucrose / corn syrup	1. Galactosemia 2. lactase deficiency 3. cow's milk protien allergy 4. Vegetarian
Extensively hydrolized protien	1. casein hydrolysate & Amino acids 2. starch, sucrose, corn syrup 3. Medium chain triglycerides	1. allergy to cow milk or soy protien 2. GI malabsorption
Elemental	1. Free amino acids 2. corn syrup solids 3. MCT & vegitable oils	1. sever protien allergy 2. sever GI impairment (CF)
Premature	↑ protien (whey) ↑ MCT ↓ lactose	1. < 1.8 kg 2. < 36 weeks gestation

* Formulas Not to be used:

1) added thickening agents → allergy, affect gastric emptying
2) Hypoallergic formula → currently available are NOT hypoallergic
3) whole cow milk → Not to be feed during 1st year of life :
1. ↓ iron, vit-E & ↑ Na, k, Cl, protien
2. Microscopic GI bleed
3. kidney stones
4. allergy
4) low fat (skim cows milk → kidney stones
Not to be feed in the first 2 years
5) Goat milk → 1. kidney stones
2. Metabolic acidosis (1st month)

Wheezing & Stridor

* Acute onset wheeze:

- Asthma: dry cough
  intermitent episodes
  atopic disease
  family history
  early inspiratory crackles
- Infections: - Bronchiolitis (<2 yrs)
  - RSV, RV, Parainfluenza
  - cough, rhinorrea
  wheese, tachypnea
  intermittent fever
  - Croup
  - atypical pneumonia
  - Bacterial tracheitis
- Foreign body: - sudden onset
  - unilateral wheese
  - unequal breath sound

* Chronic or recurrent wheeze:

- structural: - Anomalies (tracheomalacia)
  - Mediastinal mass
  - Retained foreign body
  - CVS disease
  - Vascular ring / TEF
- Non structural: - Asthma
  - Primary ciliary dyskinesia
  - Bronchopulmonary dysplasia
  - Vocal cord dysfunction → late onset
  - CF → test for it if nasal polyp
  - Aspiration syndromes

* Stridor:

* Acute onset:
- Croup: - Cough, rhinorrhea, fever
  - Resp distress at night
  - inspiratory stridor
  - hoarseness of voice
  - steeple sign on CXR

mild	moderate	sever	life-threat
No audible stridor at rest	audiable at rest	SpO2 < 93% or cyanosis	↓LOC lethargy

Tx: ABC, nebulised dexa 0.3 mg/kg
Nebulised adrenaline

Indications for admission:
- < 6 months old
- respond to > 1 adrenaline shot

- Bacterial tracheitis:
  - Fever, cough, toxic appearence

Tx: Airway / IV vanco & 3rd cephalo

- Epiglottitis: - Drooling, toxic
  - Hyperextended neck
  - thumb sign on CXR

Tx: Intubation, IV cephtriaxone
→ H. influenza - B

* Chronic:

- laryngomalacia:
  - inspiratory stridor (low pitched)
  - Peak 6-9 m
  - ↑ by activity, supine, viral illness
  - ↓ by rest, prone, sleep
- Retropharyngeal abscess:
  - Under 5 years
  - Polymicrobial

Tx: admission, IV abx, incision & drain

- Unilateral VC paralysis:
  - weak voice, weak cry
  - Aspiration of liquids

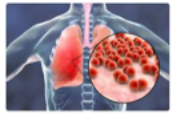
causes: Iatrogenic, Neurologic, birth trauma or idiopathic

Dx: laryngoscopy

- Bilateral VC paralysis:
  - Feeding difficulty, Normal voice
  - cyanosis, apnea, OSA, FTT

causes: Iatrogenic, Neurologic, birth trauma
Dx: laryngoscopy ,, Tx: tracheostomy

Pneumonia



* Its acute infection of pulmonary parenchyma.

* Classification:

1) Lobar pneumonia: - bronchi NOT affected (air bronchogram)
   → Causes: S.pneumonia, S.aureus, H.influenza, fungal
   ↳ Round pneumonia: S.Pneumonia / klebsiella

2) Bronchopneumonia: - bronchi affected (irregular distribution)
   → Causes: S.aureus, S.pneumonia, Mycoplasma
   ↳ Mycoplasma: the most common LRTI worldwide in ages 5-20 yrs
   → Symptoms but without x-ray changes
   ↳ Cavity lesion (Pneumatocele): S.aureus

3) Interstitial: diffuse inflammation & infiltration of lymphocytes & macrophages
   → Causes: RSV, Influenza, Parainfluenza, adeno

* Bacterial pneumonia:

→ 1st 2 months: klebsiella, E-coli
→ 3 m - 3 yr: S.pneumonia, H.influenza, staph
→ > 3 yr: Mycoplasma, S.pneumonia, staph, chlamydia

* Clinical features:

- Fever, dyspnea, Resp distress:

1. Tachypnea / tachycardia
2. Dyspnea
3. O2 sat < 90%
4. altered mental status
5. Agitation
6. Nasal flaring
7. Noisy breathing sound
8. Grunting
9. cyanosis
10. Pursed lips
11. Tripod position
12. Retractions
13. Accessory muscles

* Investigations:

1) CXR: - viral → diffuse bilateral infiltrates
   - S.Pneumo → lobar consolidation
   - staph → Pneumatocele
2) WBC: - viral → N or ↑ but < 20000
   - Bacterial → 15000 - 40000 + Neutrophilia
3) CRP & ESR & PCR

* Complications:

1) Pulmonary:
   - Pleural effusion / empyema - Pneumothorax
   - Pneumatocele (thin wall, No air fluid level)
   - lung abscess (thick wall, with air fluid level)
   - Broncho-pleural fistula - Resp failure
2) Mets:
   - Meningitis - CNS abcess
   - Pericarditis - endocarditis
   - osteomyelitis - septic arthritis
3) Systemic:
   - SIRS / sepsis - HUS

* Indications for hospital admission:

1) Resp distress or hypoxemia (< 90%)
2) < 6 month & suspected bacterial CAP
3) suspected virulent pathogen (MRSA)
4) If there's concern about home care

Tx:

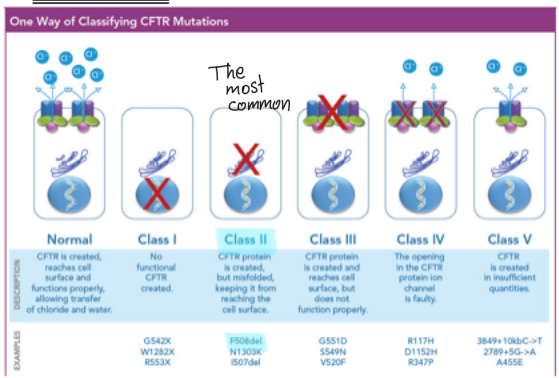
- Inpatient
  - typical → Ampicillin
    - alternative → ceftriaxone
      - ↓ MRSA → Vancomycin
  - atypical → Azithromycin
    - alternative → clarithro or Erythro
- Outpatient
  - typical → Amoxicillin or Amox-Cav

⇒ Supportive:

1. Hydration
2. Oxygenation
3. Antipyretic
4. Painkiller

Cystic fibrosis

* Autosomal recessive mutation in CFTR gene on chromosome 7
* Defect in Cl channels → Cl + Na doesn't get out of cells and so H_2O → No hydration of secretions in every channels & tubes
* Mutations:



* Systemic effect:

1) RS: Defective mucocilliary clearing → Mucus obstruction → infection + inflammation
⇒ Infection: S. aureus / late: Pseudomonas
⇒ Progressive obstucting lung disease

2) Pancreas: Pancreatic insufficiency → auto-digestion → CF related DM
⇒ steatorrhea (hard to flush, greesy, bulky, Pale)

3) Intestine: thickend mucus → obstruction

4) Biliary tree: focal biliary cirrhosis → chronic cholecystitis + Cholelithiasis
⇒ deficiency in Vit-K, A, E, D

* Clinically:

1) RS:
- Chronic sinusitis, Nasal polyp
- viscous purulent green sputum
- Pneumothorax, hemoptysis
- Clubbing, cor pulmonale

2) GI:
- Meconium illius (failure to pass stool)
- distal intestinal obstruction syndrome:
  - = RLQ Pain
  - = Loss of appetite
  - = emesis
  - = Palpable mass

3) GU: infertility: Male: Azospermia
Female: thick vaginal secretions
⇒ IVF is usefull

* Diagnostic criteria:

1) one of:
- typical Features (wet cough, mucus & wheese)

And
- Hx of CF in sibling
- Positive newborn secreening test

2) labs:
- 2 ↑ sweat chlorine in 2 separate time
- Identification of 2 CF mutations

* Investigations:

1) Newborn secreening (IRT)
2) sweat chlorine (in 2 separate ocations)
→ infant weight > 2 kg & ⩾ 36 week gues
→ Ⓝ < 30
grey zone: 30-59
CF: ⩾ 60

* Treatment:

1) Lung:
- Prophylactic abx for 1 y in newcase
  - ↳ staph: Flucloxacillin
  - ↳ Psudo: Cephalo + Aminoglycoside
  - ↳ arosolized abx
- DNAse
- Inhaled B-agonist
- Oral steroids

→ Atelactasis: physiotherapy & abx
→ Cor pulmonale: lung transplant

2) GI:
- Pancreatic enzyme (creon)
- Vit replacement
- Insulin for hyperglycemia
- liver transplant (end stage)

Asthma

* Inflammation → Bronchospasm → hyperinflation → hyperventilation → V/Q mismatch → hypoxia

* Viral infections (adeno & COVID) are the most important cause of asthma exacerbation

* Types of childhood asthma:
1) Chronic asthma associated with allergy
2) Recurrent wheeze triggered by viral infection
3) Asthma in female, obese, early puberty

* Asthma Predictor index:
1) One of: - Parent hx of asthma
- Positive skin test
OR
- Eczema
2) two of: - Eosinophilia ≥ 4%
- Wheeze unrelated to cold
- Allergic sensitization to food

* Signs & symptoms:
- Dry cough, wheeze, SOB, Chest tightness, sputum
- Diurnal variations (Nocturnal & early morning awaken)
- Resp distress, End expiratory wheeze
- Diminished breath sounds, Pulsus paradoxus
- Weight loss
- Erythematous turbinate or polyps
- atopic dermatitis or eczema

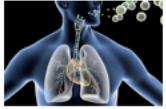
* Investigations:
1) PFT: ↓FEV1 >> FEV1/FVC < 0.8
Bronchodilator response ≥ 12%
Exersise challeng worsening ≥ 15%
2) skin Prick test
3) Fraction of exhaled Nitric oxide (FeNO)
4) Eosenophilia in sputum & Blood
5) Urinary leucotrienes
6) Total & specific IgE
7) CXR for: - atypical presentation
- Complicated

* High risk Patients:
1) sudden sever exacerbation
2) Prior intubation OR admission to ICU
3) ≥ 2 hospitalization / yr
4) ≥ 3 ER visits / yr
5) ≥ 2 cainsters of inhaled SABA / month
6) current use or withdrawl from systemic steroids

* Tx:
- Quick relief
↳ Rapid acting B-2 agonist
↳ systemic steroids
↳ Ipratropium bromide
- long term
↳ ICS: only for first 3 steps
↳ LABA: Not for < 5 yrs old
↳ LTRA: for < 5 yrs old
↳ systemic steroids
↳ LAMA
↳ Biologic

* Acute sever asthma:
- Breathless at rest, Resp distress
- Not interested in feeding
- Sits upright
- Talks in words not sentences
- agitated and confusion, cyanosis
- Tachycardia, O2 sat < 92%
- wheeze & Pulsus Paradoxus may disappear

* Treatment:
1) Oxygen (face mask → venturie → CPAP)
2) Albutereol (B2 agonist)
↳ continuous nebulization
3) systemic steroids
4) IV fluids
5) IV adrenaline if: - Anaphylactic
- life threat attack
6) Anticholinergic (ipratropium)



Upper respiratory tract infections

* The common cold:

→ Common cause is Rhinovirus

→ symptoms: Rhinorrhea, nasal obstruction
sore throat, cough

* Epidimiology:
→ children: 6-7 colds/yr
→ adults: 2-3 colds/yr
→ children in day care centres

N.B: change in color or consistency of nasal secretions OR PMN predominates does NOT indicate bacterial superinfection

Tx:
1) supportive: - Nasal spray (oxymetazoline)
↳ Not for child < 2 yr
- 1st gen antihistamine
- acetamenophen
2) Antiviral

* Complications:
1) Otitis media
2) Sinusitis
3) Asthma exacerbation

* Sinusitis:

* Cause: S. pneumonia, H. influenza
Moraxella, Staph

* Predisposing conditions:
- URTI
- Allergic rhinits
- Smoke exposure
- CF, GERD
- foriegn body
- Cilliary dyskinesia

* Clinically: Nasal congestion, discharge
fever, cough, facial pain, headache

* Dx: - Persistent RTI > 10-15 days
- Temp > 39° & purulent discharge 3-4 d
- Sinus aspiration & culture (not done)
- X-ray → air fluid level

* Tx: - Amoxicillin for 7 days
- Frontal sinus: ceftriaxone

* Complications:
- Periorbital & orbital cellulitis
- Meningitis, Cavernous sinus thrombosis
- epidural & subdural abscess
- brain abscess
- Osteomyelitis of frontal bone

* Acute pharyngitis:

* Causes: Viral & Bacterial
* Clinically: - sore throat - Fever
- headache - GI symptoms
- Red pharynx, tonsilar exudate
- Anterior cervical LAP
viral: coug + rhinorrhea + diarrhea

Dx: CBC, culture, anti SPOT
Tx: symptomatic & abx
Complications: - otitis media
- Parapharyngeal abscess
- PSGN
- Rheumatic fever

* Retropharyngeal & lateral pharyngeal abscess:

* Clinically: - Fever, irritiability
- ↓oral intake, drooling
- Neck stiffness, refuse to move neck
- sorethroat, muffled voice
- stridor, Resp distress

* Dx: - CT - culture
* Tx: - IV abx (3rd gen + ampicillin-sulbactam)
- Surgical drainage

* Indications for adenoidectomy:

1) Chronic nasal infection 2) Recurrent OM
3) Chronic sinus infection not responding to tx

Congenital heart diseases - Cyanotic

	Tetralogy of fallot TOF	Transposition of great arteries TGA
Components	1. large VSD 2. over-riding aorta 3. Pulmonary stenosis 4. Rt. ventricular hypertrophy	1) Aorta arise from RV 2) Pulmonary arise from LV
Hemodynamic	Blood from RV → Narrow Pulmonary (oligemia) → VSD → large VSD make pressure equal bw ventricles	two isolated circulations, incompatible with life → Patient need shunt (ASD, VSD, PDA)
Clinically	1) Central cyanosis: - delayed → PS progress gradually → PDA - at birth → sever PS (extreme fallot) - absent → mild PS (Pink fallot) 2) blue clubbing: after 6-12 months 3) FTT 4) Cardiac: - systolic thrill (P area) - ejection systolic murmur (P area) - Single accentuated S2 (aortic)	1) Central cyanosis: - at birth, deep - Not relieved by 100% O_2 2) Marked RV hypertrophy 3) Accentuated S2 → No thrill or murmur
Investigation	1) CBC: Hct > 65 « ↓MCV 2) ABG: hypoxemia, acidosis 3) CXR: Boot shaped heart, ↓ Pulmonary vascular markings 4) ECG, ECO, Cath	1) CXR: egg on string, ↑Pulmonary vasculobure 2) ECG, ECO: RVH 3) Cath
Treatment	1) Total repair 2) Iron 3) PG (early) Tet spells: knee-chest → oxygen → Phenylephrine also: IV fluid, BB, Morphine, Na-Bicarb	1) Prostaglandin 2) urgent shunt: Rashkind (ballon atrial septostomy) 3) switch operation (in 1st-2nd week of life)
Complications	1) Tet spells: - sever cyanosis, syncopy, convulsion (hyper cyanotic spells) - Causes: stress: infection, cry, hypoxia 2) Infective endocarditis 3) Polycythemia + hyperviscosity 4) Brain infarction or abscess 5) IDA	1) Heart failure, infective endocarditis 2) Recurrent chest infection 3) Thrombosis & Polycythemia 4) stroke & Brain abcess

* Indication for surgery in TOF:

1. O_2 sat < 80% 2. Hypercyanotic spells

* The most common cyanotic congenital heart disease → in Newborn → TGA
→ in general → TOF

* The most common congenital heart disease is VSD

Cardiac cyanosis	Respiratory cyanosis
- Soft tachypnea - Murmur	- Resp distress → dyspnea, retraction → harsh tachypnea → Grunting - Relive by 100% O_2

* Other cyanotic heart disease:

1) Total anomalous P. venous return
→ CXR: snow man appearence

2) Ebsteins anomaly:
→ CXR: very large heart

3) Tricuspid atresia

4) Truncus arteriosus

5) Hypoplastic left ventricle:
→ cyanosis + sepsis + Shock

* N.B: if newborn presents with sever cyanosis + Met. acidosis → initiate Prostaglandin (PGE1)

Congenital heart disease - Acyanotic

	VSD	ASD	PDA	A. Coarctation	Aortic stenosis	Pulmonary stenosis
Types / description	1) Membranous (80%): don't close 2) Muscular (20%): close with aging	1) Ostium secondum (upper part) 2) Ostium premium (lower part) 3) Common A-V canal (Down synd.)	- Congenital rupella - Premature	Narrowing of descending aorta usually distal to left subclavian artery	- Valvular: Bicuspid - Supra or subvalvular	- Valvular stenosis - Supra or sub valvular
hemodynamic	1) Left to right shunt during systole ↳ volume overload → LVH ↳ Pressure overload → PHTN 2) late → ↑PHTN → Rt to lt shunt → cyanosis (eisenmenger)	1) left → Right shunt ↳ lung plethora ↳ Rt. V. Hypertrophy 2) ostium primium usually associated with mitral regurge	1) Physiological closure in 24 hrs 2) Blood shift from Aorta → Pulmonary 3) Lt. atrial & Ventricular enlargement	1) upper limb: ↑BP, ↑B. flow strong radial pulse 2) lower limb: ↓BP, ↓ B. flow weak Femoral pulse 3) Lt. V. Hypertrophy	1) obstruction of blood flow from Lt. V to aorta 2) LVH	1) obstruction of blood flow from R.V to pulmonary 2) RVH
clinically	1) Small → Asymptomatic 2) Feeding difficulty, Forehead sweating Failure to thrive, Exertional dyspnea 3) Recurrent chest infections: fever, cough 4) Cardiac: - apex shifted outward & down - Systolic thrill on left parasternal - Accentuated S2 - Harsh pansystolic murmur on left parasternal (max)	1) secondum: Asymptomatic 2) Primium: forehead sweating feeding difficulty Exertional dyspnea Recurrent chest infection S2 wide fixed splitting Biventricular enlarge Murmur (MR → axilla)	1) Small: Asymptomatic 2) Forehead sweating Feeding difficulty Exertional dyspnea, FTT Recurrent chest infections 3) Big pulse volume (water hammer pulse) 4) Lt. V. Hypertrophy Continuous (Machinery) murmur systolic thrill accentuated S2 (PHTN) Gallop rhythm	Mild: Asymptomatic sever: Headache, Blurring vision Feeding difficulty, FTT Forehead sweating, Fatigue Heart: - strong apical pulse (LVH) - Systolic thrill (Lt. Parasternal) - Accentuated S2 (HTN) - Harsh systolic murmur in left sternal border (Max: inter scapular area) - Radio-Femoral delay	1) Mild: Asymptomatic 2) Feeding difficulty, FTT Forehead sweating Exertional dyspnea 3) easy fatigability syncopal attack fainting 4) Recurrent chest infection 5) - Apex shifted ↓→ - Systolic thrill on Lt parasternal - weak S2 - harsh systolic murmur (aortic) - Small pulse volume - ↓BP	1) Asymptomatic diagnosed accedentaly 2) ejection systolic murmur over Pulmonary area 3) Muffling of S2
Investigation	- CXR: cardiomegaly - ECG, Eco, Cath *Note: Lt. V. enlarge before Rt. V *Cyanosis → Peripheral (HF), Cardiac (shunt reversal), Chest (infections)		- CXR: Lt. V enlargement lung plethora - ECG, ECO - cath	- CXR: Cardiomegaly lung congestion rib notching (Rosler's sign) - ECG, ECO - spiral CT		
Treatment	1) surgical closure after ↓PHTN by banding the P. artery 2) Small: close spontaneous	1) Secundum: occlusion devise 2) Primium: surgical correction ↳ in 3-6 yrs 3) in 3-6 months for CAVC	1) Indomethacin (PG ⊗) 2) surgical ligation 3) cath insertion of intravascular coils before 1st yr age	1) Coarctectomy 2) Intravascular stent 3) Balloon angioplasty	- Ballon dilatation - Valve replacement	- Balloon valvoplasty if sever
Complications	1) HF, infective endocarditis 2) PHTN, shunt reversal (cyanosis) 3) FTT	1) HF, A-fib, Rt BBB 2) PHTN 2) shunt reversal (eisenmenger)	1) HF, IE 2) PHTN 3) Shunt reversal ↳ lower limb cyanosis	- HF, IE, HTN - Intracranial hge	- HF, IE - Sudden death	- HF, IE - Pulmonary oligemia ↳↑ risk of Tb

Heart failure & Kawasaki disease

* Inability of heart to supply tissues w/ blood

* Etiology:

1) Preload: - Hypervolemia (ARF, fluid overinfusion)
- shunt lesions (ASD, VSD, PDA)

2) Afterload: - HTN
- obstructive lesions (AS, coarctation)

3) Pump: - Myocarditis (viral: Coxaki, toxins)
- Cardiomyopathy
- -ve inotrope: hypoxia, hypoglycemia

4) Arrhythmia: - sever tachycardia > 220 BPM
- Heart block

* Clinically:

1) Pulmonary congestion: - Dyspnea - orthopnea
- Paroxysmal nocturnal D.
- Pulmonary edema
↳ coarse crepitations
- Cough + Chest infections

2) systemic congestion: - Congested neck vein
- Puffy eyes (infants)
- Tender hepatomegaly
- lower limb edema
- Sacral edema
- Diaphoresis
- irritability

3) ↓ out put: - dizziness, syncopy
- Chest pain
- oliguria
- Cool periphry (peripheral cyanosis)
- easy fatigue
- Gallop rhythm & S3
- Pulsus alterens
- Pulse dificit
- ↓ capillary refil

* Investigations:

- CXR: cardiomegaly, lung congestion
- ECG: Arrhythmia - ECO
- CBC, ESR, CRP, ASOT, Troponin I, CPK

* Treatment:

1) Preload: Diuretics
2) Afterload: Dilators (captopril)
3) Inotrope: Dopamine, Dobutamine, Digitalis
4) supportive: - Oxygen
- semi-sitting position
- Salt & Fluid restriction

* Diuretics side effect: ↓k, ↓Na, dehydration
↑Ca, hyperurecemia
hyperglycemia

* kawasaki disease

* Vasculitis of small-medium sized vessles in children 6 months - 4 yrs
→ Acute Febrile vasculitis

* Clinically:

1) Fever > 5 days, ≥ 38° + 4 of:
2) Conjuctival injection
3) lip cracking, strawberry tounge
4) Anterior cervical lymphadenopathy
5) Erythema of hands and feet
6) skin rash
others: abd. Pain, diarrhea, arthritis

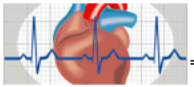
* Complication: coronary artery aneurysm

* Investigations:

- Eco: at diagnosis & 2-3 weeks of illness
- CBC: Anemia, Marked thrombocytosis (milions)
- CRP & ESR ↑↑
- sterile pyuria
- ↑ serum transaminase + Plasma levels

* Treatment:

1) IVIg: 2g/kg over 10 hrs for 1 day
2) Aspirin → No coronary effect: 6-8 weeks
↳ Coronary effect: continued
↳ Anticoagulant



Arrhythmias

* ECG Parameters in Pediatrics:

1) Rate : 60 ÷ R-R (in sec), SS = 0.04 sec, LS = 0.2 sec

2) P wave: P. pulmonale → Rt. atrial hypertrophy
P. mitral → Lt. atrial hypertrophy

3) P-R : < 0.16 sec (4 small sequares)
- long : 1st degree heart block
- Elongate till A-V block : Mobitz 1
- variable : AV dissociation
- short : Wolf-Parkinson white

4) QRS: - Narrower in neonates
- +ve in AVF, -ve in I (axis ↙)
- tall R in V1 : RVH
- tall R in V6 : LVH

5) Q wave: Ⓝ in lead II, III, AVF (> 3 mm)
- Deep in I, aVL : Anomalous Lt coronary from pulmonary A
- Deep in V5, V6 : LVH, septal hypertrophy

6) ST : Pericarditis ↑, ischemia ↑ or ↓

7) T : in V1 : Newborn: upright
Fewdays - adolecent : inverted
Adults : upright
↳ elvated if > 2/3 QRS (ex: hyper K)

8) U wave: hypokalemia

9) QT : must always be corrected
↳ QTc = QT / √R-R , Ⓝ ≤ 450 msec

* Indications for ECG in Pediatrics:

1) episodes of LOC or Palpitations
2) Congenital or structural heart diseases
3) Incidental finding of abnormal rhythm
4) Child with chest pain

* Bradycardia:

- sinus bradycardia (non cardiac cause)
- Sinus node dysfunction (sick sinus syndrome)
  - ↳ after congenital heart surgery
- Heart Block

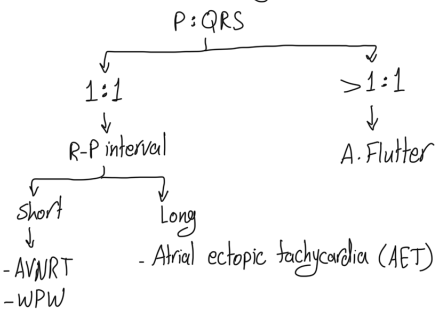
* Irregular rhythm:

- Sinus arrhythmia : Normal variation of HR between inspiration & expiration
- Second degree heart block : regularly irregular
- A-Fib : Irregularly irregular (tachy)
- Premature contractions (PACs and PVCs)
  - ↳ missed beat on auscultation

* Tachyarrhythmias:

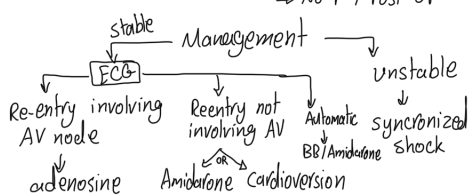
- Sinus tachy : - Maximum rate = 220 - age
- Ventricular tachy: - wide complex QRS
  - occational sinus beats (Narrow)
  - occational normal P waves
    - ↳ P < QRS (< 1:1)
- ↳ Supraventricular tachy: - narrow or wide QRS
  - rate > limits of sinus > 200

*Types: 1) AV-reentry tachy (AVRT)
2) AV-nodal re-entry tachy (AVNRT)
3) Atrial tachy (AT)



* Mechanism of SVT:

1) Accessory pathway mediated : - WPW (delta wave)
2) Non-AP mediated : - AVNRT
- A. Fib
- A. Flutter
3) Non-reentry : - AET (Polymorphic P)
- Junctional ET
  - ↳ No P, Post op



Acute diarrhea

* Change in normal stool (↑Frequency) or (↑Fluidity) for 7 days and no >14 days
* Peak in 6-11 months (enteral feeding)
  ↳ before it: - Formula milk & unsterile bottle or mother's hands

* Causes:
1) Bloody: salmonella, shigella, C. jejuni
   Entero-invasive E-coli, Entamoeba histolytica
2) watery: Rota. V, Entero-toxogenic E-coli
   Giardia, staph aureus
3) lactose intolerence, cow milk allergy

* Complications of diarrhea:
1) Dehydration & shock
2) Electrolytes imbalance (↓K, ↓Ca, ↓Na)
3) Metabolic acidosis
4) Convulsions: 1. Fever
   2. ↓Na (Brain edema)
   3. Hypocalcemia
   4. toxins
5) Malnutrition (if recurrent)
6) sepsis & DIC
7) ARF & HUS (↑ with shigella)

* Indications for investigations:
1) if < 3 months (septic workup)
2) Patient with chronic diseases (DM)
3) very sick „ Travel hx
4) Bloody diarrhea

* Investigations:
1) stool analysis & culture
2) Septic workup: CBC, ESR, CRP, B. culture
3) KFT & Electrolytes, Urine Na
4) ABG (sever or complicated cases)
5) coagulation profile (for DIC)
6) Urine Na, Osmolarity, specific gravity
   ↳ in dehydration

* Treatment:
1) Oral rehydration solution (mild-moderate)
   ↳ 75 ml/kg over 3-4 hrs
   ↳ 100 ml for each loose stool
   ↳ contents: Na: 90 mmol, Cl: 80
   K: 20 „ glucose: 110
   citrate (bicarb): 30
2) Abx, antipyretic
3) Antiemetic (odansteron)
4) Zink (10-20 mg/day) (infants > 6 months)

* Signs and degrees of dehydration:

Symptom	Mild	Moderate	Sever
Mental status	conscious alert	Drowsy or irritable	lethargic unconscious
weight loss	children 5% infants 5%	7% 10%	10% 15%
HR, Pulse	N	↑, weak	↑↑, very week
BP	N	N to ↓	Shock
Fontanels	N	sunken	deeply sunken
RR	N	Fast	Fast & deep «acidosis»
Eyes	N	sunken	deeply sunken
Tears	Present	Decreased	Abscent
Tounge	Moist	Dry	very dry
skin fold	elastic	retract slowly	Retract very slowly (tent)
Extremities	N	Cool	Cold & cyanotic
UOP	N	decreased	Anuric
Cappillary refill	N	2-5 sec	>5 sec

* NB: Use lactose free milk for 2 weeks after invasive (bloody) diarrhea
* NB: Tachycardia and delayed cappillary refill Precedes hypotension & oliguria

Dehydration

* Isotonic dehydration :

Factors affecting insensible water losses

Increased losses	% Change	Decreased losses	% Change
Prematurity	100–300	Enclosed incubator	25–50
Radiant warmer	50–100	Humidified air	15–30
Phototherapy	25–50	Sedation	5–25
Hyperventilation	20–30	Decreased activity	5–25
Increased activity	5–25	Hypothermia	5–15
Hyperthermia	12%/°C		

* Management :

1) Bolus of 20 ml/kg in 20 min up to 3 doses.
   ↳ Normal saline
2) Maintaince : 100 ml/kg for the first 10 kg
   50 ml/kg for the second 10 kg
   20 ml/kg for the rest of weight
   ↳ half saline (GS 0.45%)
   → use his original weight
   → Do NOT include the bolus
3) Deficit : 10 x weight x degree of dehydration
   ↳ half saline (GS 0.45%)

* Maintainence + dificit : half in the first 8 hrs and half over 16 hrs

* Hyponatremic dehydration :

Etiology of hyponatremia

Circulating volume	Urinary Na (mEq/L)	
	≤ 20	≥ 20
Decreased	Burns	Adrenal insufficiency
	Cystic fibrosis	Diuretics – early
	Diuretics – late	Salt wasting (CAH)
	Gastroenteritis	
Normal or Increased (fluids in interstitial)	Cardiac failure	Renal failure
	Hepatic cirrhosis	SIADH
	Nephrotic syndrome	Water intoxication

* Symptoms :

1) seizures (Brain edema)
2) Nausea, malise, lethargy

* Corrected by 10-12 mmol/day
* Use hypertonic saline (3%)
* Na mmol = (desired - actual) x wt. x 0.6

* Hypernatremic dehydration :

* Symptoms : - irritability
- ICN, cerebral thrombosis
- hypocalcemia
- hyperglycemia
- Renal vein thrombosis

* Causes :

1) Una < 20 : diarrhea
   inadequate water intake
2) Una > 20 : - Renal : Diabetis insipedis
   Hyperglycemia
   diuretics
   intrensic renal diseases
   - ↑ salt gain : ↑ oral ingestion
   ↑ minaralocorticoid
   ↑ IV saline
   Rapid correction by diuretics, dialysis

* Treatment :

1) 1.25 - 1.5 x Maintaince over 24 hrs
   ↳ GS .3% or GS .45%
2) correct over 48-78 hrs

Chronic diarrhea

* DDx:

1) Functional diarrhea:
- Toddlers age group
- otherwise well child

2) Intractable diarrhea of infancy:
- Loss of digestive absorptive capacity
- Ex: lactose intolerence:
  - Failure to gain weight
  - Abscence of breastfeeding

3) Milk & soy protien intolerence:
↳ first 3 m : Enterocolitis (bloody diarrhea)
↳ >6 m : protien losing enteropathy
occult blood loss

Tx: Protien hydrolysate formula

4) Pancreatic insufficiency:
- Greasy, Foul smelling stool
- Pale, bulky stool

→ Cause: CF
→ Dx: Fecal fat / sweat chlorine / stool elastase
→ Tx: creon (pancreentic enzyme supp)

5) Celiac disease:
- Bloating, diarrhea, or constipation
- lactose intolerence
- Anemia, ↓ Vit-D
- Chronic fatigue, joint & muscle pain
- Dermatitis herpitiform
- Associated Autoimmune diseases:
  ↳ DM1, RA, thyroid, AI liver disease

Dx: anti-TTG / Biopsy (Marsh score)
Tx: Gluten free diet

6) IBD:
- weight loss, abdominal pain
- anal fistula (crohns)
- +ve FH
- Anemia, ↓ Albumin, ↑ ESR, CRP
- Uveitis, Erythema nodosum
- Growth failure, development delay

Dx: upper & lower endoscopy
MRI or small bowel follow throw

Tx: Anti-inflammatory
Immunosupressent
Biologic
Surgery

* Clues in the history:
1) Onset: - after birth: NEC
- at weaning: celiac
2) Stool volume & character
3) Associated symptoms: Fever, Nocturnal awake
4) Diet, FH, Antinatal, Drugs, rash

* Investigations:
1) Stool electrolyte & osmotic gap:
OG = 290 − 2 (stool Na + K)
↳ >100 : osmotic (lactose intolerence)
2) Stool elastase
3) Fecal calprotectin
4) stool α-1 antitrypsin: protien loosing enteropathy
5) stool reducing substances and PH
6) stool C. difficle (not for <1 yr)
7) Stool microscopy and culture
↳ RBC, WBC, fat globules, Giardia

* Red flags:
1) Hematochezia or melena
2) weight loss or growth arrest
3) Anemia
4) Persistent fever

Cholestasis

* Choly: bile + Stasis: ↓ movement
* Hyperbilirubinemia where direct is > 20%

* Causes:

1) Idiopathic (neonatal hepatitis) (Giant cell hepatitis)
2) Extra-hepatic biliary atresia (2nd most common) EHBA
3) Cholydochal cyst (cyst of the common bile duct)
4) Infections: TORCH, neonatal septicemia
5) Metabolic diseases:
   - Galactosemia
   - Tyrosinemia
   - α-1 antitrypsin deficiency
   - CF
6) Familial syndromes:
   - Alagille syndrome
   - Progressive familial intrahepatic cholystasis

* Consequences:

1) Fat malabsorption (steatorrhea)
2) Fat soluble vit deficiency (A, K, E, D)
3) Progressive liver damage → cirrhosis

* Clinically:

1) Persistent jaundice > 2 w (olive green)
2) Dark urine & Pale stool
3) Hepatomegaly
4) Pruritis
5) Bleeding & Bone disorders
6) if Alagille: Deep eyes, large ears, VSD or PS, vertebral arch defict (butterfly arch)
7) wheeze: if CF or α-1 antitrypsin deficiency
8) cataract or mental retardation: if Galactasemia

* Investigations:

1) serum bilirubin (↑ total & ↑ direct)
2) liver enzymes (↑ ALT, ↑ AST, ↑ Alkaline phosphatase)
3) Albumin serum (↓)
4) PT, INR (for vit-k assesment)

For the cause:

→ sepsis: CBC, CRP, ESR, culture
→ Galactosemia: reducing substances in urine
→ Tyrosinemia: urine succinyl acetone (Make urine black)
→ α-1 antitrypsin: serum level
→ Cholydochal cyst: US, CT
→ ToRCH: Total IgM (> 20 mg)

To differenciate idiopathic from EHBA:

→ Biopsy → Giant cell transformation → idiopathic
→ Biopsy → expantion of portal area w/ fibrosis → EHBA
→ HIDA scan → No dye excretion → EHBA

* Treatment:

1) Abx if sepsis
2) Galactosemia → lactose free diet
3) Cholydochal cyst → surgical correction
4) EHBA → Kasai operation
5) Medium chain triglyceride
6) Vit - replacement
7) liver transplant:
   - if Kasai Failed (EHBA)
   - end stage liver disease

** liver cirrhosis:

Causes:

1) Drugs (MTX) and alcohol
2) NAFLD
3) Chronic hepatitis (HBV, HCV)
4) Metabolic: wilsons, α-1 antitrypsin
5) Auto immune hepatitis, Biliary cirrhosis

* Metabolic syndrome: 3/5 of:

1) obesity 2) HTN 3) hyperglycemia
4) Triglycerides > 150 5) ↓ HDL (M < 40, F < 50)

→ associations with metabolic syndrome:

1) Fatty liver 2) OSA

* Fatty liver → steatohepatitis → Fibrosis → cirrhosis
  (vit-E (anti oxident))

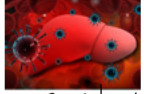
* NB: synthetic function of the liver:

1) Albumin (HL: 3 weeks) 2) PT (Factor 7) (HL: 7hrs)

* NB: AST and ALT in cirrhosis are abnormally Normal as theres No hepatocytes to produce them
* NB: in cirrhosis, liver shrunk in size and become impalpable

* Complications:

1) Portal HTN
2) Hepatic encephalopathy
3) Carcinoma
4) Growth failure



Viral hepatitis

* Types :

1) HAV, HEV : RNA , Acute , Fecal oral
Incubation : 2-6 weeks

2) HBV, HDV : DNA , Acute - chronic , Parentral
sexual & vertical , IP : 2-6 months
Not transmitted by breast milk

3) HCV : RNA , Acute - Chronic , Parentral
sexual , vertical , IP : 1-5 months

→ Vaccines presents for HAV & HBV

* Clinically :

1) HAV : - Fever , Vomiting , abdominal pain , anorexia
- Dark urine , Pale stool
- Jaundice , Hepatomegaly & tenderness
- severly ↑↑ aminotransferase

2) HBV, HCV : - similar to A
- gradual ↑ aminotransferase

* Investigations :

1) Bilirubin (↑ total early , ↑ direct later)
2) ↑ ALT & AST (10 folds)
3) urine analysis (Bilirubin present)
4) PT & INR
5) ↑ Blood ammonia (the main toxin) (↑ encephalopathy)
6) serum albumin (↓ after 3 weeks !)

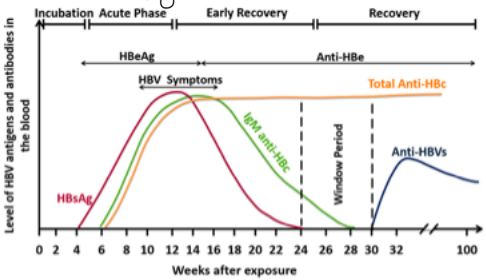
* Serologies :

1) HAV : - Anti-HAV-IgM : Acute infection
- Anti-HAV-IgG : Recovery OR immunity

2) HBV : HBsAg : +ve in acute & chronic infection
HBsAb : +ve in immunity
(HBsAg & HBsAb never +ve together)
window period between ↓HBsAg and ↑HBsAb
HBcAb : if +ve never become -ve
IgM : infection < 3 months
IgG : infection > 3 months
HBeAg : Infectivity (↑ viral load)
HBcAg : Not in blood (biopsy)

دلالة انه الفايروس دخل الجسم بقالة حتى لو طلع زي الوشم

* No HBsAg = No virus now



3) HCV : PCR
4) HDV, HEV : - HDV-Ag
- Anti-HDV and Anti-HEV (IgM)
→ HBVAg + Abs are +ve in D infection

** Hepatic encephalopathy :

* Toxins (Ammonia/GABA) → BBB → Brain edema
* Causes of ↑ toxins :
1) Failing liver
2) Bypassing liver (pre hepatic Portal HTN)

* Stages of encephalopathy :
1) Abnormal behaviour, irritable, sleep disturbance
2) Confusion, agression, Slurred speach, Astrexis
3) disoriented
4) Coma, decerebrate or decorticate

* Tx : ↓ ICP (of edema) :
1) Mannitol 2) bed at 45°
3) Hyperventilation to wash CO_2
4) ↓ protien intake (↓ ammonia)
5) Abx (if inf) & Dextrose 10 (for hypoglycemia)
6) laxatives

* Manifestations of liver cell failure :

- Encephalopathy , Jaundice
- Spider navi , bruising , epistaxis
- Ascitis , caput medusa , spleenomegaly
- Clubbing , Palmar erythema , Flabbing tremor

Other GI topics

**GERD:

*Causes:

1) Physiologic: (1st yr)
- Immature lower esophagial sphincter
- Small stomach capasity
- Short intra-abdominal esophagus
- large volume feeding (supine position)

2) Pathologic: Hiatal hernia, scleroderma

*Clinically:
- infants: regurgitation (resolve by 6 months)
- Children: Pain, dysphagia, FTT, vomiting, weight loss

*Complications:
1) Bleeding (IDA) & Peptic stricture
2) Apnea (vagal stimulation) & aspiration
3) sudden infant death syndrome (vagal stimulation)

*Investigations:
1) 24 hrs PH monitor
2) Barium study (hiatal hernia, esophagial strictures)
3) Endoscopy (Esophagitis, strictures)

*Tx:
1) life style: (Thickening formula by cerials, avoid acids)
2) Antacids, H_2 receptor blockers, PPI
3) Neissen fundoplication:
- sever esophagitis
- complicated case
  - ↳ Recurrent aspiration
  - ↳ strictures

**Celiac disease:

→Intestinal intolerence to gliadin fraction of Gluten
→Immune mediated in genitecally predisposed

*Pathology: villous atrophy, Crypts hyperplasia epithelial damage, lymphocyte infiltration

*Clinically:
- onset 6 months - 2 yrs (solid food)
- Diarrhea, Abdominal distention, FTT
- Pale, loose, offensive stool
- later childhood anemia (IDA)
- Arthritis, chronic fatigue
- Dermatitis herpetiform

*Associations: DM, Thyroiditis, Addisons

*Investigations:
- anti-TTG ab
- anti-endomysial ab
- Endoscopy with biopsy

*Tx: Gluten free diet (oat is allowed)

**DDx of upper GI bleeding:

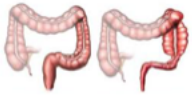
1) Esophagitis, Gastritis, Peptic ulcer
2) Hemorrhagic disease, coagulopathy disorder
3) Cow milk allergy
4) Esophagial varices, AV malformation
5) Forign body, corrosive

**DDx for lower GI bleeding:

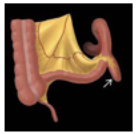
1) Bacterial enteritis / Amebic dysentry
2) Cow milk allergy
3) Intussusception
4) Henoch-Shonllien purpura
5) Anal fissure, Colonic polyps
6) IBD, NEC
7) Meckel's diverticulitis

**DDx of constipation:

1) Functional: voluntary withholding due to painful defication (Anal fissure)
2) Imperforated anus, intestinal stricture
3) Cieliac, CF, Hirschsprung
4) Neural tube dificit, trauma
5) ↓K, ↓thyroid
6) ↑Ca, ↑ Vit-D
7) Myopathy (Duchene muscular)



Hirschsprung disease & Meckel's diverticulitis



* Hirschsprung disease:

→ Congenital aganglionic megacolon

Pathogenisis: - abscence of ganglionic cells in the myenteric & submucosal plexuses of rectum → Failure of relaxation (obstruction)

Extent: Recto-sigmoid (80%)

Clinically: - Neonate: delayed passage of meconium
- Childhood: - Chronic constipation
- Abdominal distension
- Bilious vomiting
- Malnutrition
- Enterocolitis
- PR: Narrow segment, empty rectum
Gush of fluids and gases

Investigations: Barium enema, Anorectal manometry
suction rectal biopsy

Tx: - a primary pull-through procedure
- Colostomy: if associated enterocolitis

* Meckel diverticulitis:

→ 2% of individuals
→ 2 inch from ilio-cecal valve
→ 2 types of mucosa (Pancreatic OR Gastric)

Clinically: - Painless rectal bleeding
- Intussusception
- Diverticulitis (if pancreatic → amylase, lipase)
↳ clinically as appendicitis

Investigations: Meckel scan (IV tech-99m)

Tx: surgical excision

Hematuria

** Nephritic syndrome:

* Causes:

1) Post-strept glumerulonephritis
2) SLE, Henoch-Shonlien purpura (HSP)
3) Membrano proliferative glumerulonephritis

* PSGN:

→ Cause: B-hemolytic strep (tonsilits / scarlet fever)

* Clinically: - Mild edema (periorbital → LL)
- ARF (oliguria / Azotemia)
↳ < 1cc/kg/hr
- Hematuria (>5 RBCs)
- Hypertension (↓GFR, ↑Na-H2O)

* Complications: - HF, RF (↑K)
- Hypertensive encephalopathy
- Dialysis (Rapidly progressive GN)

* Investigations:

- Urine analysis: Hematuria, Protienuria, RBC cast
- Blood: KFT, ↓C3
+ve ASOT & anti-DNA ab
- Renal US
- Biopsy if atypical features:
= @C3 = HTN or ↓GFR > 2 weeks
= Sever anuria = Rapidly progressive
= -ve ASOT

* Tx:
- ↓Fluid, K, Phosphorus
- Diuretics (Frusimide)
- ACEI (captopril)

* NO ROLE FOR STEROIDS HERE

* NB: Risk for PSGN after GABHS is 15%

* NB: Abx doesn't prevent PSGN

* Other causes of 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

* Alport syndrome:

→ 80% x-linked

* Clinically: - RF
- Sensorineural deafness
- Ocular changes
- Microscopic hematuria w/ URTI
- Protienuria, HTN (late age)

* Dx: EM: Thinning of glomerular basement membrane

* Tx: ACEI delay progression to ESRD

* IgA nephropathy:

* clinically: - Recurrent macroscopic hematuria
- loin pain 1-2 days after URTI
- ± Protienuria

* Dx: ↑IgA, light microscope

* Complications: ESRD (if HTN or heavy protienuria)

* Tx: ↓Na, ACE or ARB, steroids

* Henoch schoenlein purpra (IgA vasculitis):

* Criteria: - Purpura on lower limbs + 1/4
1) Acute onset diffuse abdominal pain
2) IgA dominant immune deposits
3) acute onset arthralgia or arthritis
4) hematuria or Protienuria
↳ The MC renal involvement

* Risk for Intussuception and ICH

* Dx: stool occult blood, ↑IgA

* Tx: spontaneous recover (if sever → steroids)

* Hemolytic uremic syndrome:

* Triad of: Acute hemolytic anemia, ↓GFR, ↓ Platelet

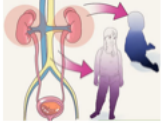
* Causes: Infections, hereditary, Malignancy, Drugs

* Diarrhea (bloody) + HUS: shigella, shiga toxin E-coli
↳ ↑Risk: Age < 5yr, WBCs > 13000, antimotility

* Clinically: Bloody diarrea, Palor, oliguria, colitis hepatitis, Pancreatitis, Glucose intolerence seizures, irritability, HTN, renal cortical necrosis → 75% dialysis

* Dx: CBC ↑LDH, schistocytes, Fragmented RBCs, KFT↑ ↑ liver enzymes, hematurea, Protienuria

* Tx: Transfusion (sever hemolysis), Platelets (bleeding), Fluid



Nephrotic syndrome

* Characteristics:
1) Heavy protienuria
2) Hypoprotienemia
3) Hyperlipidemia
4) Generalized edema

* Eitiology:
1) Idiopathic: - Minimal change disease
- Focal segmental sclerosis
- Mesangial proliferation

2) secondary: - Immune (SLE, HSP)
- HBV, HCV
- DM
- Drugs (penicillamine)

* Minimal lesion nephrotic syndrome:
→ Child 2-6 yrs, M:F : 2:1

* Clinically: - Pitting, generalized edema
↳ 1st around eys (morning)
↳ Scrotal/labial swelling
↳ Ascitis/Plueral effusion (Anasarca)
- Resp distress
- Frothy urine
- Abdominal pain, vomiting

→ NO HTN, NO Hematuria, NO Fever

* Complications:
1) Infections: - Spontanuous bacterial peritonitis
- Pneumonia
- Pyelonephritis
- septicemia
↳ Staph, Pnemococci

2) Thrombosis: Hemo concentration
↓ Protien C & S, ↓ antithrombin 3

3) Hypovolumic shock
4) Hyperlipidemia

* Investigations:
1) Urine analysis: Protienuria +++, ++++
Hyaline cast
↳ Albumin/creatinine: > 2
↳ N < 0.2, Protienuria 0.2-2
↳ Protien loss: > 1gm/day

2) Blood: - Albumin < 2.5 gm/d, ↓ Ca
- ↑↑ Cholesterol, ↑ TG & lipoprotiens
↳ KFT, C_3, C_4 are NORMAL

3) Renal biopsy if atypical:
- age < 1 yr or > 12 yr, FH of renal disease
- persistent HTN or hematuria
- abnormal KFT or C_3 ↓
- Skin rash
- No steroid response (4w)

* Treatment:
1) supportive: - ↑ carbs, Protiens, Ca^{+2}, Vit-D
- ↓ salts, Fat
- Salt free albumin IV then diuritic

2) Specific: - Steroids
- Immunosupressive
- ACEI (to ↓ Protienuria)
↳ afferent arteride V.C

3) For complications: - Abx & vaccination
- anticoagulant

* Complications of steroids:
1) HTN
2) Muscle wasting
3) Peptic ulcer
4) Osteoporosis

NB: Hypoalbuminemia:
< 2.5: Edema
< 1.5: Anasarca
< 1-1.2: Scrotal/labial edema

* Normo-albuminuria: < 30
* Micro albuminuria: 30-300

AKI

* Decline in GFR
→ definitions of AKI in peds (pRIFLE & KDIGO):

pRIFLE stage	Estimated creatinine clearance (eCCl)	Urine output
R = Risk for renal dysfunction	eCCl decreased by 25 percent	<0.5 mL/kg per hour for 8 hours
I = Injury to the kidney	eCCl decreased by 50 percent	<0.5 mL/kg per hour for 16 hours
F = Failure of kidney function	eCCl decreased by 75 percent or eCCl <35 mL/min per 1.73 m^2	<0.3 mL/kg per hour for 24 hours or anuria for 12 hours
L = Loss of kidney function	Persistent failure >4 weeks	
E = End-stage renal disease	Persistent failure >3 months	Ⓝ 1 ml/kg/hr

* Risk ↑ in PICU, mechanical ventilation & vasopressors

* Classification:

1) Pre renal: Volume depletion: - bleeding
- intestinal loss
- ↑ cautaneues loss (burn)
↳ ↓GFR, Ⓝ Tubular function (reabsorption) → Oliguria

2) Intrinsik renal: - Prolonged hypoperfusion (ATN)
- Sepsis
- Glomerular disease (PSGN, HUS)
- Acute interstitial nephritis
- Nephrotoxins

3) Post renal: - Stones - Clots
- strictures - PUV
- Neurogenic bladder

* Clinically:
- Edema, ↓UOP, hematuria, HTN
- Fever, arthritis, rash
- Vomiting, diarrhea

→ oliguria: UOP < 1ml/kg/hr infants, < 0.5 ml/kg/hr children > more than 6 hrs

* Investigations:

1) KFT: Cr: Ⓝ (Not specific) Newborn 0.3 - 1
Infant 0.2 - 0.4
Child 0.3 - 0.7
adolecent 0.5 - 1

2) Electrolytes: ↑K, ↑PO4, ↓Na, ↓Ca

3) CBC, VBG

4) Urine analysis: Hematuria
Protienuria
WBCs
Specific gravity

5) C3, C4, ASOT, Uric acid

6) US, Biopsy

Measurement	Prerenal AKI	Intrinsic AKI
Urine specific gravity	>1.020 concentrated	<1.012 diluted
Urine/plasma creatinine	>40	<20
Urine Na (mEq/L)	<20 ↓	>40 ↑
FENa	<1 percent	>2 percent
FEUrea	<35 percent	>50 percent

* New specific markers:

1) Neutrophil gelatinase associated lipocallin (NGAL)
2) Kidney injury molecule-1 (KIM-1)
3) IL-18
4) Cystatin C
5) liver type fatty acid binding protien

* Management:

1) Fluid (NS): 10-20 % fluid overload is a critical threshold for outcome
↳ Hypovolemia: NS bolus → No UOP → U-cath
↳ Euvolemic: ½ NS every 6 hr over 6 hr
↳ Hypervolemia: Furosemide
↳ Fluid overload > 20% → renal replacement T.

2) Hyperkalemia: causes:
- ↓GFR, ↓ tubular secretion of K
- Metabolic acidosis
- tissue break down (Tumor lysis, rhabdomyolysis)

→ ECG changes when K > 7:
- tall peaked T, Prolonged PR
- Flat P, wide QRS
- V. tach and Fib

Tx: abnormal ECG: Ca-gluconate
Glucose + insulin
Frusimide " dialysis

3) Wide AG met. acidosis: Na-bicarb

4) HTN: respectively: 1) Diuretics
2) CCB
3) ACE

* RRT: Fluid overload or ↑K unresponsive
BUN 80 - 100
sever met. acidosis not responsive
life threatening complications

CKD

* Evidence of structural or functional kidney abnormalities for at least 3 m with or WITHOUT ↓ in GFR
→ Not for children < 2 yrs

* Causes:

1) Obstructive uropathy
2) Renal hypoplasia and dysplasia
3) Reflux nephropathy
4) Nephritis (FSGS)
5) PKD

* Consequences:

1) kidney failure & ESRD (70% in 20 yrs)
2) Cardiovascular diseases
3) Growth & development retardation

* Most common cause of death:

1) Cardiovascular disease
2) Infections

* Staging of CKD:

Table 1. Stages of CKD[a]

Stage	Description	GFR (mL/min/1.73 m²)
1	Kidney damage with normal or GFR	≥ 90
2	Kidney damage with mild GFR	89-60
3A	Mild to moderate GFR	59-45
3B	Moderate GFR	45-30
4	Severe GFR	30-15
5	Kidney failure	< 15 or dialysis

* Complications:

1) Anemia: develops early
Tx: EPO & iron

2) Renal Bone Metabolic disease:
* ↑PO4, ↓Ca, ↓Vit-D, ↑PTH
↑FGF-23 (fibroblast growth factor 23)

clinically: Fractures
Skeletal deformities
Poor growth
Vascular & soft tissue calcifications

Tx: ↓PO4 intake & PO4 binders
active vit-D when PO4 controlled

3) HTN: ACE-ARB

4) Met. acidosis: in stage 4
Tx: Na-bicarb

5) CVS: LVH due to HTN and Anemia

6) Volume overload & Hyperkalemia

7) Nausia, vomiting, anorexia

8) short stature, FTT

9) CNS: lethargy, seizures, coma

* RRT:

1) Hemodialysis
2) Peritoneal dialysis
3) Continuous renal replacement therapy
→ None is superior in term of outcome

* special indications:

2) Neonates & small infants
3) Hemodynamic instability
↳ Multiorgan dysfunction
↳ continuous management of fluid overload
↳ ICU

Dysuria

* Burning or pain upon urination.

* Causes:
1) Infections: pyelonephritis, Cystitis, urethritis
2) Vulvovaginitis (white discharge)
3) Chemical irritation (soap, poor hygine)
4) stones
5) others: sexual abuse, trauma, labial adhesion

* UTI:
→ M>F as neonate, more in uncircumsized

* Classification:
1) Cystitis: urinary symptoms, low grade fever
2) Pyelonephritis: loin Pain, ↑Fever (scar)
3) Asymptomatic bacteriauria: No treatment

NB: sterile pyuria (WBCs, -ve culture):
- Nephritis (all types)
- Viral infection
- Kawasaki
- Cx after starting abx (3rd cephalo)
- IBD

* Clinically:
- Neonate: Fever, sepsis (Poor feeding, lethargy) Hypoactivity, FTT, Prolonged juandice
- Children: Fever, vomiting, abdominal pain Urinary symptoms, hematuria

* Dx: Urine culture & microscopy
↳ > 50 000 CFU
Urine analysis: Pyuria (>5 WBCs) microscopic hematuria
Urine gram stain
leukocyte esterase (not specific)
Nitrite (specific, not sensitive)
ESR, CRP, WBC

→ ways of collection:
SPA, cath, clean catch, midstream

→ Organism:
E-Coli, klebsiella, Psudomonas, Enterobacter

* Scaring: lead to Protienuria, HTN, CKD
↳ DMSA scan (4 month after UTI)

Risk:
- Young age
- High grade VUR
- Delayed Tx of UTI
- Recurrent UTI
- Bacterial virulence
- Bowel and bladder dysfunction

* Imaging: US: if Fever with UTI
VCUG: if Hydronephrosis, scarring atypical or complex cases

* Treatment:
1) Pyelo: 3rd cephalo, ampicillin & aminoglycoside
2) Previous culture or extended spectrum B-lactam: Aminoglycosides or carbapenem
3) Cystitis: 2nd or 3rd cephalo OR Amox-Calv
→ No difference bw. oral vs IV
→ Follow up by urine test (not culture)
→ Use IV abx if: too young, vomiting
→ Prophylactic abx prevent UTI but NOT scarring

* Stages of VUR:
1) Grade 1: reflux to ureter
2) Grade 2: reflux to renal pelvis
3) Grade 3: Grade 2 + dilation
4) Grade 4: More dilation
5) Grade 5: Tortousity & conver calysis

* NB: Nitrite +ve → diagnostic of UTI
Leukocyte estrase +ve & Nitrite -ve culture

* NB: Fever maybe the only presentation of UTI in infants

Hypotonia

* Floppy infant : Generalized hypotonia presents at birth

* Presenting features :
- ↓Fetal movement , Abnormal presentation Polyhydramnios , Frequent C-section
- Arthrogryposis اعوجاج المفاصل
- Poor resp effort (develop pectus excavatum)
- Poor suck & swallow (choking , leaking milk) Poor weight gain
- Frog like posture
- Traction response (head lag)
- vertical response (Ⓝ suspend elbow, elevate shoulder)
- Ventral suspension (arched back)

* Types :
1) Cerebral hypotonia : - TORCH
- Down , Prader Willi
- HIE
- Cerebral malformation

↳ Abnormalities of other brain function
Fisting of the hand
Normal or brisk tendon reflex
scissoring on vertical suspension
Movement through postural reflexes

2) Motor unit :
- Anterior horn cell : Spinal muscular atrophy
- Peripheral nerve : Gullian Barré (rare in neonate)
- Neuromuscular junction : myasthenia gravis
- Muscular dystrophy and congenital myopathies

↳ Absent or depressed tendon reflexes
Fasciculation (tounge)
Muscle atrophy
Failure of movement on postural reflexes

* Spinal muscular atrophy (SMA) :
→ Degeneration of anterior horn cells
→ Deletion or mutation on Ch. 5

1) SMA-1 (werding hoffman disease) :
→ Presents from birth to 6 months
→ AR
→ Die before 1 yr from resp. failure

2) SMA-2 :
→ largest group
→ AR
→ Normal till 6 months, Present before 18 months
→ Proximal muscle weakness
→ life span from 2 yrs to 3rd decade

3) SMA-3 :
→ Mild form , Normal life span
→ After 18 months
→ AR , AD , X-linked
→ Symmetrical muscle weakness

* Investigations :
1) Electromyography
2) Gene testing

* Tx : Gene therapy
Supportive

Meningitis

* Pathogenisis:

1) Hematogenous: Immune deficiency
   - Aseplenic
   - Neonate
   - Complement↓ (N. meningitis)
   - Hypogammaglobulinemia
   - HIV
   - DM, steroid use
2) Direct: - sinusitis, mastoiditis
   - Trauma, Neurosurgery, CSF leak
   - CSF shunt, choclear implant

→ Predisposing factors: - Immune deficiency
- Anatomical defict
- Recent infection
- Recent exposure
- travel

* Causes:

1) Bacterial: < 3 m: GBS, E.Coli, listeria
   > 3 m: S.Pneumonia, HiB, N.meningitis
2) Viral: Enterovirus (coxsackie), CMV, EBV
3) Fungal: ↓ immune
4) Tb: extra pulmonery Tb

* Clinically:

1) Neonate: - Bulging fontanel
   - Poor Feeding, vomiting
   - Fever OR Hypothermia, Resp distress
   - irritability, seizures
   - lethargy, ↓ consciousness
   - meningococcal rash (blanchable)
2) Children: Fever, neck rigidity, ↓consciousness
   - Brudzinski, kerning signs
   - Headache, vomiting, Papilledema
   - Cushing: ↑BP, ↓ HR, irrigular breathing
   - Confusion, convulsion, coma
   - Focal paralysis, Photophobia

* Investigations:

1) CBC, Blood culture, CRP, Procalcitonin
2) kFT, electrolytes, glucose, PT, PTT, INR
3) LP: CSF culture, Gram stain & PCR and:

Parameter	Normal	Bacterial	viral	Tb
cell count	1 m: 30 2-3m: 9 >3m: 5	↑↑↑	↑↑	↑
cell type	lymphocyte	Neutrophil	lymphocyte	lymphocyte
Protiens	20-40	↑	Ⓝ or ↑	↑↑
Glucose	2/3 blood G. 40-80	↓	Ⓝ or ↓	↓↓

* Contraindications for LP:

1) CVS-RS unstability
2) ↑ICP (Papellidema, Focal neurologic sign)
3) Skin infection at site of LP
4) Bleeding disorders

* Indications for neuroimaging:

1) signs of ↑ICP, coma
2) Hx of hydrocephalus or present of CSF shunt
3) Hx of CNS trauma or surgery

* Management:

1) supportive: - Oxygenation, ↑head of bed
2) Abx: < 3m: Ampicillin + cephtriaxone
   > 3m: Vancomycin + cephtriaxone
3) steroids if: - HiB (↓risk of deafness)
   - Brain edema
   - younge children < 6 yrs
   - Aseplinic, unimmunized
   ↳ Not proved for neonates
   ↳ given under umbrella of abx
4) Prophylaxis for contact: Rifampicin + Vaccine

* Complications:

1) subdural effusion/empyema
2) Hearing dificit/Blindness
3) ↓IQ
4) seizures (if > 3 days ↑ risk for epilipsy)
5) Hemiparesis & neurological deficit
6) Sepsis, Shock, DIC

Epilepsy

* Types of epilepsy:

1) Absence : precipitated by hyperventilation
2) Myoclonic
3) Tonic
4) Tonic-clonic : followed by deep sleep
5) Atonic : sudden fall to the floor

* Causes:

1) Structural : trauma , tumor , vascular
2) Genitic : trisomy , kleinfelters , tuberous sclerosis
3) Infections
4) Metabolic : Glut-1 deficiency
5) Immune : Rasmussen syndrome

* Epilepsy syndromes:

1) Lennox Gastaut :
   - < 8 yr
   - Multiple types (tonic, myoclonic, absence)
   - Very abnormal EEG
   - Tx : Valoprate , lamotrigine

2) Infantile spasm:
   - infancy
   - spasms (arms abducted , neck flexed, leg extend)
   - EEG : Hypsarrhythmia
   - developmental delay
   - Tx : Steroids or Vigabatrin

3) Childhood abscence :
   - 4-8 yrs
   - Abscence
   - EEG : generalized 3Hz spike waves
   - Stimulated by hyperventilation
   - Resolves by adolecent
   - Tx : Ethosuximide , lamotrigine

4) Benign Rolandic epilepsy : in Jordan
   - 4-11 yr
   - Focal (ipsilateral face & tounge)
   - Resolve by puberty
   - Tx : Carbamazepine

5) Juvenile myoclonic epilepsy :
   - > 12 yr
   - Myoclonic (kelog's) (drops what in hands)
   - EEG : fast spike waves 4-6 Hz
   - Provoked by sleep deprivation & Flash light
   - Tx : Life Long Lamotrigine , valoprate

* Status epilepticus : > 5 min

Tx : 1) ABC

2) Anticonvulsant :
   - 0-5 min : Benzo (1. lorazepam / 2. Diazepam 3. Midazolam)
   - 5-10 min : 2nd dose benzo
   - 10-15 : Fosphenytoin , phenobarbital
   - 15-30 : Phenobarbital , valoprate , Pyridoxine
   - >30 : Anesthesia consult

* Febrile seizure :

→ > 6 months → rapid ↑ temp or ↓ temp
→ simple : < 15 min , Generalised , don't recur in 24 hrs
→ Complex : > 15 min , Focal , recure in 24 hrs
→ Tx as seizures acutely but No daily prophylactic
→ Antipyretics do NOT Prevent seizure

* Neonatal seizures : ↑ in ↓ Birth weight

* Types : - subtle (transient eye deviations)
  - Clonic , tonic , spasm , myoclonic
* Causes : - HIE , IVH , Drug withdrawl
  - Infections , kernicterus , Metabolic
* Tx : Phenobarbital , Phenytoin , levtriacetam
  ↳ ↓ receptors of bnzos

* Epilepsy imitators :

1) Vaso vagal syncopy : last sec , trigger , +ve FH
2) Breath holding attacks : start with crying
   Deep cyanosis + syncope
   Common in IDA children
   Resolve spontaneous
3) Long QT & cardiac syncopy : triggered by exercise
   strong FH
   sensoryneural deafness
4) Reflex anoxic seizures : sudden stimulus
   vagal stimulation → Asystole
   LOC & Pale

Headache

* Primary headache:

1) Migraine: last 2-72 hrs
   Unilateral but NOT sidelocked
   ↑ by activity
   Nausea, Photophobia, Phonophobia
   - ↳ with aura: vertigo, diplopia, nystagmus
   - ↳ Chronic: ≥ 15 day/month, ≥ 8 migrainous headache
   - ↳ Complications: - Status migrainous > 72 hr
     - Persistent aura
     - Migrainous infarction
     - seizures
   - ↳ Tx: Abortive: Analgesia, triptans, Ergotamine
     Preventive: Amitriptyline, topiramate, propranolol

2) Tension: Pressing or tightening (min - days)
   - ↳ Infrequent: ≥ 10 episodes in < 1 day/month
   - ↳ Frequent: ≥ 10 episodes in 1-14 day/month
   - ↳ chronic: ≥ 15 day/month

* Red flags:

1) side locked
2) worse with laying down, valsalva, exercise
3) worse with standing upright
4) Awakening pt. from sleep
5) New onset
6) Focal neurological symptoms
7) Immune compromised

* Secondary headache:

1) Brain tumor
2) Brain bleeding
3) Idiopathic intracrainial hypertension
4) cerebral veinous sinus thrombosis
5) concussion
6) Infections
7) Sinusitis

Other neurological diseases

→ All are autoimmune
→ All typically preceded by infections.

* Autoimmune encephalitis:

* Symptoms: encephalopathy, Psycosis, seizure
Focal deficits, movement disorders

* Criteria: - subacute onset
- ≥1 Neurological symptoms or MRI features
- Reasonable exclusion of other causes

* Investigations: CSF (↑ protiens, ↑ lymphocytes, ↑IgG)
Anti NMDA & anti-GAD 65
EEG & MRI

* Tx: Steroids IV, IVIg

* Acute disseminated encephalo myelitis:

→ In white matter → Prepubertal children

* Clinically: - Fever & acute onset encephalopathy
- Hyperreflexia, clonus

* Investigations: CSF (↑ protiens, ↑ lymphocytes)
MRI

* Tx: IV steroids, IVIg
→ Typically monophasic but (MOG ab) ↑ risk of relapse

* opsoclonus myoclonus ataxia syndrome:

* opsoclonus, myoclonus, ataxia, neuroblastoma
* Investigation: CSF & serum ab, MRI (cerebellar atrophy)
* Tx: IV steroids, IVIG

* Transverse myelitis:

→ demyleneation of spinal cord
→ Bimodal (<5 yr, >10 yr)

* Clinically: acute onset motor, sensory, autonomic dysfunction
First presentation is backache
initially hyporeflexia but then hyperreflexia
sensory level

* Investigation: CSF (↑ protiens, ↑ lymphocytes)
MRI (common in cervical)

* Tx: IV steroids, IVIg, Plasmapharesis

* Poor outcome: younge, rapid onset, complete Paraplasia, assissted ventilation

* Acute Flaccid myelitis:

→ Polio like (affect anterior horn cells)
↳ Asymmetric, Favors upper limb & proximal muscles
→ Hyporeflexia
Dx: MRI (↑ in cervical) (restricted to grey matter)
Tx: supportive (10% only achive full recovery)

* Guillian Barre syndrome:

* trigger: Campylobacter, CMV, EBV, Mycoplasma
* Clinically: symmetric ascending parasthesia
Flaccid paralysis, abscent reflexes
* Investigation: CSF (↑ Protien, Ⓝ cells)
Antibodies
Nerve conduction study (block)
MRI (nerve root enhancement)

*Tx: IVIg
Plasmapheresis

* Charcot marie tooth:

* Clinically: distal weakness, abscent reflexes
High arched foot, Hammer toe
Distal muscle atrophy

* Investigation: NCS, EMG, DNA testing (heriditary)
dupplication on Ch.17 ↵

* Infantile botulism:

* Cause: C.botulinum in: spores in dust & soil
Honey

→ 1 week - 1 yr

* Clinically: Descending paralysis, hypotonia
Constipation, Poor feeding, hyporeflexia

* Dx: spores and botulinum toxin in stool
* Tx: Botulism Ig

* Myasthenia Gravis:

→ Post synaptic NMJ

* clinically: Ptosis, diplopia (↑ by activity, ↓ by rest)
* Dx: Ice pack test, Antibody test (anti AChR & MuSK)
* Tx: Supportive, Pyridostigmine, immunotherapy
Thymectomy

* Duchene muscular dystrophy:

→ X-linked ressesive

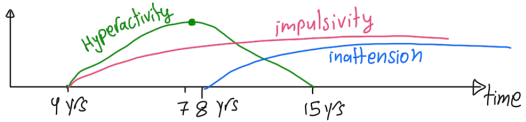
* Clinically: Proximal muscle weakness, cardiomyopathy
Cognetive dysfunction
+ve Gower's sign, Psudohypertrophy in calf

* Dx: CK, DNA test
* Tx: Steroid, Gene therapy, supportive

ADHD & ASD

* ADHD:

* Hyperactivity, impulsivity ± Inattention
→ Most common disorder of childhood
→ 1/3 before 6 yrs, common in males



* Diagnostic criteria:

1) <17 yr: ≥6 symptoms of hyperactivity & impulsivity
OR ≥6 symptoms of inattension

2) ≥17 yr: 5 symptoms each

And:

1) Present in more than one setting
2) At least persist for six months
3) Present before age of 12 yrs
4) Impair function

* Tx:

1) <5 yrs: Behavioural therapy
2) ≥6 yrs: stimulant drugs: Methylphenidate
Amphetamines
Atomoxetine
α-2 agonist
Behavioral therapy

* Autism spectrum disorder:

* Characteristics:
1) deficits in social communication & interaction
2) Restricted repetitive pattern of behaviour, interest & activities
→ Common in males

* Associated conditions:
1) Mental retardation (50-75%)
2) Seizures (10-40%)
3) Feeding disturbances (20-25%)
4) Sleep disturbances

* Impaired social communication: (hallmark)
1) Social reciprocity deficits
2) Inability to develope or maintain relationships
3) Impaired non-verbal communication

* Restricted repetitive behaviour:
1) stereotyped behaviours, Ecolilia
2) self injurious behaviours
3) Need the same routiene identically everyday
4) Restricted interests
5) Intellectual impairment

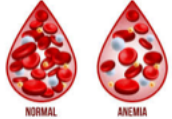
* Treatment:

1) Behavioural & educational interventions
2) Drugs: Risperidone & Aripiprazole
→ Start with low doses then increase slowly

* Poor outcome:

1) lack of joint attention by 4 yrs
2) lack of functional speech by 5 yrs
3) IQ < 70
4) seizure
5) sever ASD symptoms

Anemia



* Hb or Hct below the normal range for age.
* Normal : RBCs : at birth 15-20 gm/dl
  1st two months : physiologic anemia
  WBC : neonate : 10000 - 20000
  children : 4000 - 11000
  Platelets : 150000 - 450000

* Structure of Normal hemoglobin :

Hb	composition	at birth	6-12 months
HbF	$2\alpha + 2\gamma$	70%	2%
HbA1	$2\alpha + 2\beta$	30%	96%
HbA2	$2\alpha + 2\delta$	<1%	2%

* Symptoms : - easy fatigue , tiredness
  - ↓ concentration , ± syncopy
  - exertional dyspnea & palpitation

* Signs : - Pallor (lips , conjuctiva, Palms , nails)
  - water hummer pulse (↓ viscosity)
  - HF

* Types of anemia :

Anemia
- Decreased Red cell Production :
  - Red cell aplasea : - Black fan diamond
    - Parvovirus B19 infection
    - Fanconi anemia
    - Aplastic anemia
    - Leukemia
  - Ineffective erythropoiesis : - IDA
    - Folic acid deficiency
    - Chronic renal failure
- Increased Red cell destruction (hemolysis) :
  - RBCs membrane defict : - Hereditary spherocytosis
    - Elliptocytosis
  - RBCs Enzyme disorders : - G6PD deficiency
    - Pyruvate kinase deficiency
  - Hemoglobinopathies : - Thalassemia
    - Sickle cell anemia
  - Immune hemolysis : - AI hemolytic anemia
- Blood loss : - Feto maternal bleeding
  - Inherited bleeding disorders
  - Internal or external losses
  - Chronic GI blood loss

Haemolytic anemia

* Causes:

1) Corpuscular (intrinsic) (chronic) (Hereditary):
- Hereditary spherocytes, elliptocytes
- Thalassemia, Sickle cell anemia
- G6PD, Pyruvate kinase deficiency

2) Extracorpuscular (extrinsic) (acute) (Acquired):
- ABO or Rh incompatibility
- AI hemolytic anemia, SLE
- Sepsis, HUS, DIC
- Hypersepleenism (Pancytopenia + reticulocytosis)

* Clinically:

1) Signs and symptoms of anemia
2) Hemosidrosis (↑ iron)
3) ↑ Bilirubin (dark urine, Juandice ± Stones)
4) HSM (abdominal distension)
5) Mongoloid features + Reticulocytosis

** Thalassemia:

1) Alpha thalassemia:
- ↳ silent carrier (1 gene)
- ↳ α-thalassemia trait (2 genes)
- ↳ HbH disease (3 genes)
- ↳ Hydrops fetalis (4 genes)

2) Beta thalassemia:
- ↳ minor (trait): - one abnormal gene
  - mild (microcytic hypochromic anemia)
  - ↑↑ HbA2 (4-8%)
- ↳ Intermedia: - ↓ HbA1, ↑ HbF
  - Need transfusion after age 2yr
- ↳ Major (Cooly's anemia):
  - AR, abnormal 2 genes on Ch.11

* Clinically:
- onset: after 6 months of birth

* Complications:
1) Hemochromatosis: liver cirrhosis
   HF, DM
   Hyperpigmentation
   Delayed growth & sexual maturation
2) of transfusion: infections (hepatitis), volume overload
3) others: hypersplenism, gallstones

* Investigations:
1) Blood film: - microcytic, hypochromic
   - Reticulocytosis 5-10%
   - Anisocytosis, Poikilocytosis, target cells
2) Blood chemistry: - ↑ unconjugated bilirubin
   - ↑ iron & Ferretin, ↓ TIBC
3) X-ray skull: hair on end appearence
4) Hb electrophoresis: ↑ HbF (10-90%), ↓ or absent HbA1

* Treatment:
1) Packed RBCs (q 4-5 week), Follic acid (5 mg/day)
2) Deferasirox (Ex-Jade): iron chelator
3) Spleenectomy: - Hypersplenism (pancytopenia)
   - Huge (pressure symptoms)
4) BM transplantation // Gene therapy

** Sickle cell anemia:

→ AR disorder

* Inducers of sickling: hypoxia, dehydration, cold
  acidosis, infections

* Types:
1) SC trait: HbS (40%), asymptomatic
2) Sickle-β thalassemia: HbS (70%), HbA2 (↑), ↓ MCV
3) SC Anemia: HbSS

* Clinically:
- onset after 6 months of birth
- thrombotic symptoms, crisis:
1) Vaso-occlusive crisis: vascular thrombosis - infarction
   ↳ Abdomenal pain, Autosplenectomy (5-6y)
   chest pain (PE), strokes
   Painful swelling of hand & foot (dactilitis)
   Tx: analgesia, Fluid, O2, Abx, transfusion
2) Aplastic crisis: Parvo-B19, Pancytopenia, ↓ Retics
3) Sequestration crisis: Blood is pooled in spleen
   hypovolumic shock, spleenomegaly
4) Hyperhemolytic crisis: sever acute anemia (↑ Retics)

* Investigations:
- anemia + reticulocytosis, ↑ iron, ↑ billirubin
- RBCs sickle shaped
- HbS (90 - 100%), NO HbA1
- sickling test: sickling by exposure to hypoxia

* Tx:
- Packed RBCs, Follic acid
- Deferasirox
- Hydroxy urea (↑ HbF & ↓ HbS ⇒ ↓ VO crisis)

Haemolytic anemia & IDA

* Hereditary spherocytosis:
→ AD cell membrane defect

* Clinically:
- onset in neonatal period (neonatal juandice)

* Investigations:
- anemia, ↑Retics, ↑iron, ↑Ferretin
- Blood film: spherocytes
- Osmotic fragility test (rapid rupture)
- Acidified glycerol lysis test (rapid rupture)

* Tx:
- spleenectomy

* G6PD deficiency:
→ X-linked recessive
→ MCC of acute hemolysis in children

* Clinically:
- Neonatal juandice (first 3 days)
- Pricipitators: - Abx (sulphonamide, quinolones)
  - Aspirin
  - Antimalarial (chloroquine)
  - vit-k
  - viral hepatitis, sepsis
  - Fava beans
  - idiopathic

* Investigations:
- anemia (normocytic, normochromic) & ↑Retics
- Bite cells (fragmented RBCs), Heinz bodies
- Hemoglobinuria
- Enzyme activity (2-3 w after the attack)

* Treatment:
1) Packed RBCs (avoid whole blood or inotropes)
2) O2

* Autoimmune hemolytic anemia:
1) warm: - SLE, Leukemia
   - IgG (at normal body temp)
   - Spleenomegaly
   - Spherocytes

2) Cold: viral inf. (CMV, EBV, meseles, influenza, adeno)
   Bacterial (Mycoplasma)
   IgM-IgG (cold temp)

* Investigations: Coombs test
* Tx: steroids, IVIg, spleenectomy, Azathioprine

* Iron deficiency anemia:
→ MCC of anemia

* Causes:
1) ↓ iron intake: - Delayed weaning
   - Poor diet (cow's milk)
2) ↓ Absorption: chronic diarrhea
3) ↓ stores: Maternal IDA
   Premature baby
4) ↑ loss: Acute, recurrent, chronic blood loss
5) ↑ demand: menstruation

* Clinically:
- Anorexia & irritability, Pica
- Angular stomatitis, koilonychia

* Investigation:
- Microcytic hypochromic anemia, (N) or ↓ Retics
- ↓Ferretin, ↓iron, ↑TIBC

* Tx: Iron (for 6w after serum iron is (N))

* Follow up:
1) 1st effect: ↑ appetite (1st day)
2) 2nd effect: ↑ Retics

* Aplastic anemia:
- Pancytopenia, ↓ Retics

* Fanconi anemia: microcephaly
  cafe au lait spots
- hypocellular BM

Tx: Androgens
steroids
BM transplant
immunosupressive

Indication of BMT
1- Severe Aplastic anemia.
2- CML
3-AML in CR1 or CR2
4-ALL in CR2
5-SCID
6-B-Thalassemia
7- Lymphoma with relapse

Bleeding disorders

* Petechia: Pinpoint hge less than 2 mm
* Purpura: hge into skin & MM (2mm-1cm)
* Ecchymosis: subQ hge > 1 cm
* Ⓝ Platelet lifespan: 7-10 days

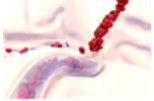
Clinical character	Purpuric disorder	Coagulation disorder
Petechia	Characteristic	Absent
Eccymosis	Small/characteristic	Common/large
Soft tissue hematoma	Rare	characteristic
Joint hemorrhage	Rare	characteristic
Delayed bleeding	Rare	common
Bleeding from superficial skin abrasions	common & persistent	Uncommon
Family history	Rare	Common
sex of Pt	Female	Male

* Investigations:

1) CBC / Blood Film
2) PT: sensitive to alteration in vit-k factors especialy: 10, 7, 2
3) aPTT: for intrinsik pathway 8, 9, 11, 12
   less sensitive for common pathway
4) Thrombin time: time for fibrenogen → Fibrin
   Ⓝ 11-15 sec
5) Bleeding time / Platelet function analyser

Bleeding disorders

- Defects of platelets:
  - Number: - ↓ Production:
    - Thrombcytopenia with absent radius (TAR)
    - Congenital amegakaryocytic thrombocytopenia
    - leukemia
    - Aplastic anemia (congenital or acquired)
  - ↑ destruction:
    - ITP / SLE
    - Wiskott Aldrich syndrome
    - HUS / DIC
    - TTP
    - Infections / Hyperspleenism
    - Drugs (ampicillin)
  - Function: - Primary:
    - Glanzmann syndrome
    - Bernard-Soulier syndrome
  - Secondary:
    - Drugs (NSAIDs / Aspirin)
    - SLE
    - Von-willibrand disease
- Coagulation defects:
  - Extrinsic pathway:
    - Factor 7 deficiency
    - warfarin
  - Intrinsic pathway:
    - Hemophila (A/B/C)
    - Von-willibrand
    - Heparin
  - Common pathway:
    - liver disease
    - vit-k deficiency
    - DIC
- Vessle wall defects:
  - immune vasculitis (SLE / HSP)
  - infections: meningococcemia / DIC
  - vit-C deficiency
  - Ehler danlos



Bleeding disorders

** Platelet disorders:

→ Mucocautanuous bleeding is the hallmark of platelet disorders
→ Thrombocytopenia is the MCC of acquired bleeding diasthesis
→ Risk of spontanuous bleeding when platelets are <20000

* ITP:

* Cause: Auto immune following viral inf by 1-2 weeks
→ Peak age is 1-4 yrs

* Clinically:
- Skin: Generalized petechial hge, not raised, don't blanch
- MM: Epistaxis, Melena, bleeding gums
- Internal organs: ICH (the cause of death) (1%)
- NO HSM /abdominal distension

* Investigations:
1) CBC (↓Platelet, ↓ RBCs, ↑WBC (viral))
2) BM biopsy: ↑ megakaryocyte but defictive budding
3) anti-Platelet ab: +ve in 60% of cases

* Tx:
1) Mild: No tx, avoid trauma, aspirin, Procedures
↳ if platelets ≥ 30 000
2) Moderate-sever:
- Steroids (BM study before it)
- IVIG (1gm/day for 2days)
- Immunosupressive (Rituximab)
- Spleenectomy (chronic/Resistent)
- Platelet transfusion & Plasmaphresis both have transient effect

NB: MCC of non thrombocytopenic purpura is HSP

* Wiskott aldrich syndrome:

→ X-linked
→ Hypogammaglobinemia, Eczema, Thrombocytopenia
→ Blood Film: Small platelets

* TTP:

→ Platelet consumption

* Glanzmann thrombosthenia:

→ AR
→ ↓ Platelet aggregation
→ deficient glycoprotien IIb/IIIa on platelet surface

* Bernard Solyer:

→ AR
→ ↓ Platelet adhesion (absent glycoprotien Ib)
→ Sever hemorrhage
→ blood film: large platelets > RBCs

** Coagulation disorders:

* Hemophilia A:

→ X-linked ressesive, deficiency of factor 8
* Clinically: "aggressive bleeding"
1) bleeding early in life or in infancy
2) Prolonged bleeding after circumcision
3) Hemarthrosis (the hall mark) → Joint damage
4) Easy bruising /skin eccymosis
5) Muscular hge (Psoas hematoma) → Maybe fatal
6) Bleeding from other orfices (epistaxis, rectal)
7) Hematuria & ICH → Maybe fatal

* Investigations:
1) PTT: prolonged (PT & platelet count Ⓝ)
2) Factor 8 assay:
Ⓝ: > 60%
carrier: 30-60%
mild: 5-30% (bleed after surgery)
moderate: 1-5% (bleed after minor trauma)
sever: < 1% (spontanuous bleed)

* Tx:
1) Factor 8: Cryoprecipitate (1, 8, 13, vWF)
IV infusion of purified factor 8
FFP (all factors)
Desmopressin (↑ factor 8 production)

* Hemophilia B:

→ Factor 9 deficiency (x-linked ressesive)
→ Same clinically to 8, Tx: FFP

* Hemophilia C:

→ Factor 11 deficiency (AR), mild bleeding

* Von-willibrand disease:

→ MC hereditary bleeding disorder (AD)
* Function: Platelet adhesion & carries factor 8
* Clinically: Bleeding from mucocautanuous surfaces
Bruising after surgery or trauma
Hemarthrosis is NOT usual
* Dx: ↓vW Protien, ↓F8, Prolonged PTT & bleeding time
* Tx: FFP / cryoprecipitate

DM

* Hyperglycemia due to defective insulin secretion and/or action

* Types of DM:

1) Type 1 DM: Not diagnosed before age of 6 m
2) Type 2 DM
3) MODY
4) DM associated with other diseases or drugs

* Clinically:

- Polyurea, Polydipsia, Polyphagia
- weight loss
- Fatigability
- DkA as first presentation → school age (infection) / puberty (hormons)

→ symptoms appear when 10% only normal B-cells are still functioning

* Investigations:

1) Fasting Plasma glucose: ⩾ 126 mg/dl
2) Random PG: ⩾ 200 mg/dl + symptoms
3) 2 hr PG in a 75g OGTT: ⩾ 200 mg/dl
4) HbA1c: longterm control (2-3m). (N) < 5.4, DM > 6.4

* Insulin:

1) ↑ lipogenisis + ↓ lipolysis
2) ↑ protien synthesis + ↓ Protienolysis
3) ↓ glycogenolysis + ↓ gluconeogenisis

→ Abscence:

1) ↑ lipolysis → ketones / Acidosis / weight loss
2) ↑ protienolysis → weight loss / muscle waste

* Counter regulatory hormones:

1) Epinephrin: ↓ secretion & action
2) Cortisol + GH: ↓ action
3) Glucagon: ↑ glycogenolysis & gluconeogenisis & lipolysis (No direct effect)

* DKA:

* Clinically:

- Abdominal pain
- vomiting
- ↓ level of consciousness
- Polyuria, Polydypsia, weight loss
- Difficulty breathing
- Signs: kussmaul breathing / dehydration

* Dx:

1) Glucose ⩾ 200 mg/dl
2) PH < 7.30
3) ketonuria or ketonemia: - acetone
   - Acetoacetate
   - B-hydroxy buteric acid
4) serum bicarb < 18

* Management:

1) ABC
2) NS 10 ml/kg
3) decrease NS to 5-7 ml/kg with KCl
4) IV insulin 0.1 unit/kg/hr
5) if acidosis improve & BG < 270 or falls > 90 swich to D5/NS w/ k and ↓ insulin

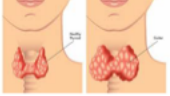
* Complications of DKA:

1) Arrhythmia / cardiac arrest
2) Venous thrombosis
3) Pulmonary edema (ARDS)
4) ARF
5) Bowel ischemia
6) cerebral edema, Risk:
   a) age < 5 yr
   b) New onset DM
   c) ↑ initial serum urea
   d) ↓ initial $PaCO_2$
   e) Rapid administration of hypotonic F
   f) IV bolus of insulin
   g) early IV insulin (1st hr)
   h) use of bicarb

* Screening for retinopathy & nephropathy:

1) Anually from age 11 yr or after 5 yrs of diagnosis, whichever is sooner

Hypoglycemia & Hypothyroidism



* Wolfram syndrome:

⇒ DM, Diabetes insipidus, optic atrophy & deafness (DIDMOAD)

→ other manifestations:
- Neurogenic bladder w/ hydroureteronephrosis
- Neurodegenerative illness (ataxia)
- Hypogonadism

* Hypoglycemia:

* Clinically:
1) Sweating
2) Pale
3) Blurred vision
4) Headache & extreme mood change
5) Hunger
6) Dizziness

* Dx: Blood sugar < 72
↳ in neonates no cutoff point (symptomatic)

* Tx: Dextrose 10%, 2-4 ml/kg bolus

* Sick day management:

→ During URTI or GE (↑ blood glucose)
→ Maintain hydration, control glucose level avoid ketoacidosis
→ Do NOT omit insulin

* MODY:

→ also called HNF4A
→ AD
→ Onset 9-25 yr
→ deficit in insulin secretion

* Diagnostic criteria:
1) DM in at least 3 generations
2) Diagnosis before age of 25yr in at least 1 affected subject

* Congenital hypothyroidism:

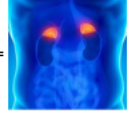
* Clinically:
1) head circumference slightly ↑
2) Prolonged physiological jaundice
3) ↓ activity
4) Feeding difficulty / constipation
5) Resp difficulty
6) sub normal temp / slow pulse
7) stunted growth / short extremities
8) AF & PF are widely opened
9) Protrusion of large tounge
10) Coarse features
11) Dry scaly skin
12) muscle hypotonia

* Causes:
1) Permanent: Primary: Thyroid dysgenesis
↓T4 Goiter
↑TSH Thyroperoxidase TSH resistent
central: Pituitary or hypothalamus disorder
↓T4 ↓TSH Inactivating mutation
2) Transient:
- sever iodine deficiency
- acute iodine overload (rare)
- maternal antithyroid drug (clears 3-4 days after birth)
- Transplacental transfer of TSH receptor blocking ab
- Hypothyroxemia of prematurity

* Complication: Development delay

* Tx: levothyroxine

Adrenal disorders



* Diurnal rhythms of ACTH & cortisol:

- → CRH Peaks at 4 am
- → ACTH Peaks at 4-6 am
- → Cortisol Peaks at 8 am

* Causes of adrenal insufficiency:

1) Primary: - Autoimmune adrenalitis - sepsis
- Congenital adrenal hypo/hyper plasia

2) secondary: - steroid therapy withdrawal
- Hypopituitarism
- Hypothalamic tumors
- CNS irradiation or surgery

* Addisons disease:

- → Acquired primary adrenal insufficiency
- → Autoimmune (80%)

* Clinically:

1) Fatigue / weakness
2) weight loss / poor appetite
3) Apathy / convulsion
4) N+V + abdominal pain / salt craving
5) Hyperpigmentation
6) Hypotension / weak pulse
7) loss of axillary & pupic hair (women)

* Investigations:

1) ↓Na, ↑K, hypoglycemia, low voltage ECG
2) 8:00 am cortisol level
3) ACTH & ACTH stimulation test
   - ↳ 1 mcg: Baseline & 30 min Cortisol level useful in central AI
   - ↳ 250 mcg: Baseline, 30 & 60 min level
4) Urine analysis for steroids
5) Imaging & gene testing

* Tx: stress dosing: infant: 25 mg
small children: 50 mg
large children: 100 mg
↳ continuous, Divided q 6-8 hrs
- Maintainence: Daily glucocorticoids
Daily mineralocorticoid

- → strongest Glucocorticoid: Dexamethason
- → steroid w/ ↑mineralocorticoid effect: Fludrocortisone

* Cushing Syndrome:

→ Hypercortisolism → Pit. overproduction of ACTH → Cushing disease
↳ other sources → ectopic ACTH synd.
↳ Adenoma / carcinoma / hyperplasia

* Clinically:

- Hirsutism - Facial flushing - striae
- Buffalo hump - HTN - Puberty arrest
- obesity and growth arrest (imp.)
- Bone undermineralisation - weight gain

* Cushing disease: - microadenoma (80-85%)
- Tx: trans-sphenoidal surgery

* other sources: - ACTH is 10-100 times in cushing
- ACTH is NOT suppressed by dexa. supp. test

* Adrenal tumors:

- → in young children
- → Adenoma mainly secretes cortisol
- → Carcinoma mainly secrete cortisol & androgen

* Pheochromocytoma:

→ Headache, diaphoresis, tachycardia ± HTN (resistent)

Dx: metaneprins in urine and blood
CT / MRI

Tx: control BP + surgical removal

* Congenital adrenal hyperplasia:

→ Defective conversion of 17-hydroxyprogesterone to 11-deoxycortisol by 21-hydroxylase due to mutation in CYP 21A2 gene which make the enzyme deficient

* Salt loosing crisis:
→ sever ↓Na and dehydration, ↑K, met. acidosis

Tx:
1) IV bolus of 10-20 ml/kg NS
2) IV bolus of 2-4 mg/kg Dextrose 10% if hypoG.
3) Glucose + Insulin for K
4) stress dose of hydrocortisone (IV bolus)
5) daily cortisone IV 50-100 mg/m² q 6 hrs
- → stress dose continue untill pt. is stable and feeds normally
- → No need for mineralocorticoid replacement
- → ↑K may normalise with fluid and cortisone stress dose
- → IF K > 6: cardiac monitor
- → if K > 7: Ca-gluconate or IV insulin+glucose

Precocious puberty

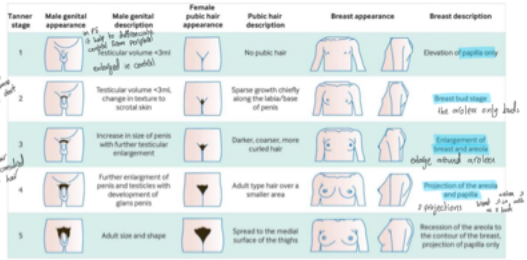
* Appearence of 2ndary sexual characteristics before: F: 8yr, M: 9yr (2.5 SD of mean)

* Pathophysiology:

→ kisspeptin: modulate the -ve feedback on GnRH secreation
  ↳ Gain of function → central precocious puberty
  ↳ Loss of function → AR, idiopathic hypogonadotropic hypogonadism

* Stages of Puberty:

1) Thelarche (breast bud)
2) Pubarche
3) Adrenarche ↳ DHAS ↳ Androstenidione
4) Menarche



* Classification:

1) Central PP (Gonadotropin dependent PP)
  ↳ iso sexual (F→F / M→M)
2) Peripheral PP (Gonadotropin independent PP)
  ↳ iso sexual
  ↳ contrasexual (F→M 2nd sexual character)

1) central PP:
- Idiopathic
- CNS tumors (↑ risk)
- CNS infection
- head trauma
(↑ risk) - Iatrogenic (radio/chemo/surgical)
- Malformation (hydrocephs)
- Genetics

2) Peripheral PP:
- Congenital adrenal hyperplasia
- Adrenal carcinoma or adenoma
- Granulosa or Theca cell tumor
- leydig cell tumor
- Ovarian cyst
- McCune Albright syndrome
- Familial male-limited PP
- hCG producing tumors
  ↳ choriocarcinoma / dysgerminoma hepatoblastoma / teratoma
- exogenous androgen / estrogen

* Premature thelarche:

→ befor 2 yrs, isolated, unilateral
→ Bone age (N), Breast regress by time

* Pelvic US:

Prepubertal:
- Ovaries show microcystic structure.
- Uterine shape is tubuler.
- Thin endometrium.
- Ovaries are smaller than 2 ml.
- Uterine length is smaller than 4 cm.

Pubertal:
- Ovaries are macrocystic/follicular
- Uterine is pear shape
- Thick endometrium.

* Investigations:

- Bone age
- TFT
- LH, FSH (>5)
- Estradiol / testosterone
- GnRH stimulation test
- Pelvic US
- Brain MRI
- IGF-1
- cortisol

* Treatment: if ↓ age or Bone age > 2 yrs

1) central: GnRH agonist (IM q month)
  ↳ Leuprolide acetate / Triptorelin / Histrelin
2) Peripheral:
  ↳ Tumors → surgery
  ↳ hCG tumors → surgery / radio / chemo
  ↳ defict in steroidogenisis → glucocorticoid

* McCune-Albright syndrome:

* Triad: Peripheral PP
  Cafe au lait spot (irrigular border)
  Fibrous dysplasia of bone
→ May present with vaginal bleeding
others: gigantism, cushing, thyrotoxicosis, adrenal hyperplasia, hepatitis, intestinal polyp, arrhythmia

* Tx: Aromatase inhibitor (Anastrazol)
  Anti-estrogen (tamoxifen) → V. bleeding
  Bisphosphonate → fibrous dysplasia

* Familial male-limited precocious puberty:

→ AD, Present at 1-4 yrs
→ Activating mutation in LH receptor → ↑ testosteron
Tx: - Spironolactone + Aromatase inhibitor
- ketoconazole (hepatotoxic)
- Bicalutamide (anti androgen) + Anastrazole
  ↳ ↓ Growth velocity
  ↳ ↓ 2nd sexual characteristics
  ↳ No serious side effects

Fever

* Acute fever: <5 days
* Rectally: ≥ 38 °C
* Fever of >7 days without focus is Fever of unknown origin FUO

* MC source is Resp (in all age groups)
* Most serious source is meningitis and meningoencephalitis

* High risk group:
1) Pale or blue colour
2) Appears ill
3) Doesn't awake or if aroused doesn't stay awake
4) Weak or high pitched cry
5) Tachypnea, grunting, sever retractions
6) Reduced skin turgor
7) Age < 3 months
8) Temp ≥ 38°C
9) Non blanching rash
10) bulging fontanel
11) Neck stiffness
12) Focal neurological signs / seizures

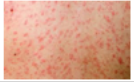
* SIRS:
→ widespread inflammatory response ± Infection
* Criteria:
one of: - Temp > 38.5 or < 36
- leukocyte count ↑ or ↓ or >10% immature neutrophils
And one of: - Tachycardia or Bradycardia
- Tachypnea or mechanical ventilation

* Sepsis: SIRS + infection
* Pathogens: - Group B strep (neonate)
- Strep pneumonia
- N. meningitis
- Staph including MRSA
- Viral (RSV)
- Candida
- Ricktesia

* Septic workup:
1) WBC & ANC
2) Procalcitonin, CRP
3) culture: Blood, urine, CSF, sputum
4) Molecular assay (PCR)

* Management:
1) ABC
2) Admission
3) Fluids, Glucose & electrolytes
4) Abx:
< 3 months: Ampicillin + cefotaxime
> 3 months: - looks well: ceftriaxone
- looks ill: ceftriaxone + Vancomycin
with No focus

* Fever of unknown origin:
* Causes:
1) Infection: Bacterial: Tb, brucellosis, salmonellosis, liver abcess, endocarditis
Viral: hepatitis, EBV
Parasite: malaria, toxoplasmosis
2) Rheumatic: Rheumatic fever, RA, SLE
3) Malignant: leukemia, lymphoma, Neuroblastoma
* Investigations:
1) CBC, CRP, ESR, LFT, KFT
2) Urine & blood culture
3) Chest x-ray, abdominal US, ECO
4) Bone marrow study
Tx: supportive / avoid empiric abx



Skin rash

	Measles (Robeola)	Rubella (German measles)	Roseola infantum	scarlet fever
Cause	Rubeola virus (RNA)	Rubella virus (RNA)	HHV-6 & HHV-7	Streptococci
Incubation P.	10 days	2-3 weeks	10 days	4 days
Prodroma	↑Fever, corysa, cough, conjuctivitis	mild fever & 3C	Fever only (40-41°C)	Fever, headache, malise sorethroat, Anorexia
Rash	Maculopapular Face (behind ears) → trunk → limbs Fades in the same order onset: day 4 Duration: 6 days Fever rises to 40° with rash onset	maculopapular Face → Neck → trunk onset: day 2 Duration: 3 days	Maculo papular Trunk onset: day 4 Duration: 1 day only	Maculopapular Axilla, neck, groin, cubital fossa onset: day 2 Duration: 3-6 days
Important Signs	koplik's spots (2nd day - 3rd day) (Pathognomonic) ↳ white macules opposite to lower posterior molar tooth	Posterior cervical & occipital lymphadenopathy ↳ characterstic not Pathognomonic	No localizing signs	Sand paper like white strowberry tounge: 1st day Red strowberry tounge: 3rd day Circum oral pallor Peeling skin (Palms & fingers)
Complications	Pneumonia Encephalitis Entritis otitis media ↳ subacute sclerosing pan encephalitis (behavioural changes mental retardation, seizure, deafness 5-10 yr later) ↳ Fatal	Congenital rubella syndrome - Microcephaly - Cataract - PDA ↳ if infected in 1st TM ↳ Fate: - Abortion - Born with manifestation - develop sym. later on	Febrile convulsions	spread of infection Rheumatic Fever (HF) PSGN
Treatment	supportive + vitamin-A	supportive + Vit-A	Paracetamol	Penicillin

Other viral infections

	Chicken Pox (varicella)	Infectious mononucleosis	Erythema infectosum (5th disease)	HSV infection
Cause	Varicella zoster virus	EBV	Parvovirus B19	HSV-1 & HSV-2
Incubation P.	2-3 weeks	4-14 day		
Clinically	Mild fever, headache, anorexia Rash: onset: day 2 red itchy Papules → vesicles → crust duration: 7 days simultaneously present Trunk → Face & limbs	Malise + Anorexia ↑Fever Pharyngitis Multiple enlarged LN groups HSM Macular rash (5%) ↳ 80% if ampicillin given	mild fever, malise, headache - Erythema on face (slapped cheeks) - Trunk & limbs maculopapular rash - Rash fades giving lacy reticulated appearence	- Gingivostomatitis - Eczema herpeticum - conjuctivitis - keratitis (dendritic ulcer) - Temporal lobe encephalitis
Investigations	CBC PCR	- WBC: 20-40% atypical lymphocyte ↳ large w/ nucleus inside nucleus and RBCs adhere to it - Viral capsid antigen (IgG & IgM) - EB nuclear antigen antibodies		
Complications	- 2ry Bacterial infection - Pneumonia (2nd MC) - Encephalitis - DIC	- Pancytopenia - Meningitis, encephalitis - Gullian Barre syndrome - Myocarditis, Pericarditis - Pneumonia	Transient aplastic crisis Fetal hydrops & death	
Treatment	Acyclovir, Paracetamol Anti-Pruritic	supportive, Acyclovir steroids (Resp obstruction / hemolysis)		Acyclovir

Immunization

*Types of immunization:

1) Passive: - Immune deficiency
- Post exposure prophylaxis
- Theraputically

2) Active: Vaccines:
- live attenuated: BCG, Flu
MMR, Varicella
oral Polio, Rota
- killed/inactivated: Pertussis, Influenza
Injectable Polio, Rabies
cholera, Hep-A
- Toxoid: tetanus, diphtheria
- Part of organism: acellular pertussis
HBV, HPV
↳ polysaccharide capsul: Pneumococcal
meningococcal
sallmonella typhi
↳ conjugated capsul: HiB
Pneumococcal
meningococcal

*Immunization schedule 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), HepB, Rota	
3 months (90+ days)	(DTaP, IPV, Hib), HepB, Rota, OPV	
4 months (120+ days)	(DTaP, IPV, Hib), HepB, RotaV, OPV	Final doses of Hib, HepB and RotaV.
9 months	Measles, OPV	Monovalent measles
12 months	MMR, Hep A	
18 months	MMR, OPV, DTP, , Hep A	Final MMR.
6 years, first grade	OPV, Td	Reduced diphtheria vaccine
10th grade	Td	

→ dose: 0.5 ml
→ site: < 18 month: Anterolateral thigh
Adolecent: Deltoid

* BCG vaccine:

→ Intra dermal (Deltoid)

* CI: - Immune deficiency
- steroid use
- significant fever
- septic skin
- Preterm
- Mother has HIV

*DPT:

CI: - FH of convulsions, sudden infant death
- CP or seizurs (CNS disorders)
↳ use DT or DaPT

* HiB:

CI: Anaphylaxis

* HBV:

CI: - sever allergy to previous dose

* If mother is HBsAg +ve: Vaccine + IVIg
then test infant for HBsAg + HBsAb

* Pneumococcal:

CI: - sever allergy to previous dose

*Measles:

CI: Immune deficiency, neomycin resistence

*MMR:

CI: - sever allergy to previous dose
- Immune deficiency, Pregnancy

* Rotavirus:

→ Risk of intussuception if 2nd dose given after 8 months of age

*Influenza:

CI: Allergy to chicken eggs

* Meningococcal:

CI: sensitivity to mercury, hx of GBS

*Varicella:

CI: Gelatin allergy, Preg, steroids, chemo

oral
- OPV
- Rotavirus

Nasal
- Influenza

intradermal
- BCG

sub-Q
- Pneumococcal
- Measles
- MMR
- Meningococcal
- Varicella

I.M
- IPV
- DPT/DPaT
- HBV
- H. influenza
- Pneumococcal (conjugated)
- Influenza
- HAV

Shock

* Tissue hypoperfusion of oxygenated blood
→ Normal central venous pressure (CVP) = 5-8 mmhg

* Types of shock:

1) Hypovolumic: - sever dehydration
- sever hemorrhage
- sever burn
↳ BP & CVP are low

2) Obstructive: - Cardiac tamponade
- Pericardial effusion
- Tension pneumothorax
- Massive Pulmonary embolism
↳ CVP is elevated
↳ Fluids & Inotrops contraindicated

3) Cardiogenic: - sever acute HF (sever hemolysis)
- Myocarditis or cardiomyopathy
↳ CVP is elevated
↳ Fluids are contraindicated

4) Distributive: - Neurogenic shock
↳ spinal cord trauma
- Anaphylactic shock
↳ CVP is low
↳ Dilated blood vessles

5) Dissociative: - Cyanide poisoning
- CO Poisoning

6) Septic: - sepsis
↳ Mixed cardiogenic & distributive
↳ Second most common type

* Hemodynamics in shock:

Physiologic variable	Preload	Pump function	Afterload	Tissue perfusion	Tissue perfusion
Clinical measurement	Clinical signs[a] or central venous pressure (if measured)	Cardiac output or index[b]	Systemic vascular resistance	Capillary refill time[a]	Mixed venous oxygen saturation[b]
Hypovolemic	↓	↓	↑	↑	Low
Cardiogenic	↑	↓	↑	↑	Low
Distributive	↓ or ↔	↑	↓	↓ initial	High
Obstructive	↑	↓	↑	↑	Low

	CO	SVR	MAP	Wedge	CVP
Hypovolemic	↑	↑	↔ Or ↓	↓↓↓	↓↓↓
Cardiogenic	↓↓	↑↑↑	↔ Or ↓	↑↑	↑↑
Obstructive	↓	↑	↔ Or ↓	↑↑	↑↑
Distributive	↑↑	↓↓↓	↔ Or ↓	↔ Or ↓	↔ Or ↓
Septic: Early	↑↑↑	↓↓↓	↔ Or ↓	↓	↓
Septic: Late	↓↓	↑↑	↓↓	↑	↑ or ↔

* Grading:

1) Tachycardia & Tissue hypoperfusion:
- Cold extremity, Peripheral cyanosis
- Core-Peripheral temp difference (>2°)
- Capillary refill >5 sec
2) Hypotension
3) Multi-organ system failure
4) Refractory metabolic acidosis / Death

* Investigations:

1) Vital signs and urine output
2) CVP, Blood culture
3) ABG, lytes, blood sugar (stress hyperglycemia)
4) CXR, ECO

* Management:

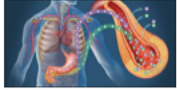
1) IV Fluid bolus:
↳ hypovolumic, distributive, septic
↳ up to 4-5 boluses

2) Inotrops: Dopamine, Dobutamine, adrinaline
vassopressors: Noradrenaline
Afterload reduction: Na nitroprussiode
↳ cardiogenic

3) Adrenaline:
↳ Distributive

4) of precipitating factors:
O_2 → hypoxia
IV Nabicarb → acidosis
Atropine → brady arrhythmia

5) specific:
surgery to control bleeding
chest tube for tension pneumothorax
Pericardiocentesis for tamponade
Abx for sepsis



Genetic and metabolic disorders

** X-linked dominant:

* Rickets: (vit-D resistent)
  * Clinically: Bowing of limbs
    Dental abnormalities
    Missing incisors
    Rossary sign
    Delayed closure of AF
  * X-ray: cupping and fraying
    widening of distal ulna & radius
    wide joint space
    Green stick fractures
  * Electrolytes: Ca + PTH are (N), ↓ PO4
    ↑ Alkaline phosphatase
  * Tx: one alpha, vit-D, Phosphate (↑dose)
  * Vit-D dependent rickets is AR

** X-Linked recessive:

* Anhidrotic ectodermal dysplasia:
  * Clinically: Fever of unknown origin
    swollen lips, big teeth
    scanty eye brow
    Never sweat

* Autosomal Dominant:

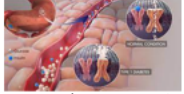
* Hyperinsulinoma:
  * Clinically: hypoglycemia
    Hypoplastic genitalia
    Micropenis
    No hepatomegaly
* Hyperlipidemia:
  * clinically: xanthoma & xanthelasma
    Abdominal pain (pancreatitis)
  * labs: lipids in thousands
* Waardenburg syndrome:
  * Clinically: Heterochromia iridis
    eyes widely spaced (hypertelorism)
    Deafness
* Neurofibromatosis-1
  * Clinically: café au lait spots (> 6)
    Axillary freckling
    hyperpigmented spots in the axilla
    Acoustic neuroma (in type-2)
* Tuberos sclerosis:
  * Clinically: Neurocutaneous:
    seizures
    Angiofibroma (sever acne doesn't go)
    Ash leaf macule (hypopigmented patch)
    Periventricular calcification on CT

* Achondroplasia:

→ The MC congenital skeletal dysplasia
→ Short stature

** Autosomal recessive:

* Glycogen storage disease:
  * Clinically: Hypoglycemia
    Hepatomegaly
    lethargy
    ↑ weight
    ↑ hair & eyelashes
    ↳ Tx: IV glucose
* Zink deficiency (Acrodermatitis enteropathica):
  * Clinically: chronic diarrhea
    Dermatitis (Napkin)
    Alopecia
* Ataxia & telangectasia:
* Mucopolysaccharidosis:
  * Clinically: Coarse facial features
    Hirsutism & thick hair
    widening of the wrist
    HSM & Umbilical hernia
    Racoon eye



Metabolic disorders

* Bickwith wiedman syndrome: AD
  - → Also called Hyperinsulinemia
  - * Triad of: Macrosomia or overgrowth
    Hypoglycemia (difficult to control)
    -ve ketones in urine
  - * other signs: Macroglossia
    Facial hemangioma
    Umbilical hernia

* Goucher disease: AR
  - → Also called lysosomal storage disease
  - → The MC lipid storage disease
  - * Clinically: Abdominal distension
    Everted umbilicus
    Massive HSM
    - ↳ Tx: Palliative splenectomy

* osteogenisis imperfecta: AD
  - → Collagen-1 defict
  - * clinically: Transparent sclera
    Blue sclera

* Galactosemia: AR
  - * Clinically: Hypoglycemia
    Abcent red reflex
    Hepatomegaly
    - ↳ Breast milk is CI

* Sturge weber syndrome: Not inherited
  - * clinically: Port wine hemangioma
    seizures
    hemiplagia contralateral to port wine
    ↑ Risk of glucoma

* Maple syrup urine disease: AR
  - * Clinically: very thin (skin & bone)
    cannot walk, bed sores
    Bad smell of urine
    - ↳ ↑ valine, leucine & isoleucine

* Phenylketonuria: AR
  - * Clinically: blond with blue eyes vs their parents
    Mental retardation
    seizures

* Marfan syndrome: AD
  - * Clinically: very long stature / thin
    Arachnodactyly
    Myopia
    Aortic dissection/anurysm

Down syndrome

* The most common autosomal trisomy (47ch)

* Genetic types:

1) Non disjunction: in maternal meosis
↑ with ↑ maternal age
Extra ch. to pair 21
Non Familial

2) Translocation: 46 ch., one is compound
Not related to maternal age
Familial

3) Mosaic type: Non disjunction of zygot mitosis
Post fertalization
Some cells have 46, others 47
Clinical features are mild

* Clinically:

1) Head: Microcephaly, Flat occiput
Wide Anterior Fontanel

2) Eyes: Medial epicanthal fold, squint
cataract

3) Ears & Nose: low set ears
Flat nasal bridge

4) Mouth: Small oral cavity, mandible hypoplasia
Large & Fissured tounge (scrotal tounge)

5) CVS: Endocardial cusion defect, VSD, PDA

6) CNS: Floppy in the first 2yrs

7) Limbs: short & broad hands
simian crease & plantar crease
5th finger curved inward
wide gap bw 1st & 2nd toes

8) Short stature

9) Genitalia: small penis, Undescended
testes, hernia

10) Global developmental delay:
Motor, language, social

* Complications:

1) CNS: Motor & mental retardation
2) CVS: HF (cause of death), CAVC, CHD
3) Hearing impairment: secretory OM
4) Visual impairment
5) RS: Recurrent chest infection
6) Blood: Leukemia
7) ECS: DM, Hypothyroid
8) Anomalies: - Endocardial cusion defect
- VSD, PDA
- Duedenal atresia
↳ Double bubble sign
- Anal atresia
- Polycystic kidney

* Investigations:

1) karyotyping
2) CXR, abdominal x-ray & US, ECO
3) Hand x-ray (abscent 2nd phalanx)
4) CBC (Leukemia)
5) TFT, Blood glucose

* Prenatal diagnosis:

* indication: Maternal age >35 yr
Previous baby with down

1) Triple test: at 15-18 week
↑ hCG
↓ a-FP
↓ unconjugated estradiol

2) Chorionic villous sampling: 9-12 week

3) Amniocentesis: 14-15 week

4) Fetal US: short femur
↑↑ Nuchal fold thickness

* Tx: symptomatic:

1) physiotherapy & early rehab
2) Correction of congenital anomalies
3) Fertility & libido

Other chromosomal abnormalities

* Turner syndrome:

→ Monosomy (45 XO)

* Clinically:

1) Short stature: Maybe the only feature
2) Head: hearing loss (otosclerosis)
   Normal intelligence
3) Neck: webbing of neck
   Thyroiditis (hypothyroid)
4) Chest: widely spaced nipples
   Coarctation of aorta
   bicuspid aortic valve
5) Abdomen: horseshoe kidney, ectopic kidney
6) Genitalia: ↓development of 2ry sexual character
   Gonadal dysgenesis (primary amenorrhea)
7) UL: Cubitus valgus, spoon shaped nails
8) LL: Dorsal edema of both feet
   ↳ present at birth

* Investigations:

1) Karyotyping
2) ECo, abdominal US
3) Thyroid profile
4) ↑FSH & LH
5) Amniocentesis / chorionic villous sampling

* Tx: Growth hormone

Estrogen for 2ry sexual characters

* Klinefelter syndrome:

→ Non disjunction XXY

* Clinically:

1) Tall stature (upper segment > lower)
2) Normal intellegence
3) Narrow shoulders, wide hips
4) Gynecomastia / lack of facial hair
5) Small testes / Hypospermia (infertility)

* Investigations:

1) Karyotyping
2) ↑↑ FSH, LH, ↓↓ testosterone
3) Gonadal biopsy (dysgenesis w/ oligospermia)

* Tx: Testosterone

→ Apart from infirtility, can live ∞ lives

* Noonan syndrome:

→ Male turner, yet affects males & Females
→ AD

* clinically:

1) General: Short stature, developmental delay
2) Head: Low set - Posterior rotated ears
   Blue - green iris
   wide spaced eyes w/ epicanthal fold
3) Neck: webbed nech
4) chest: Pulmonary stenosis, low set nipples
5) Gonads: Cryptochidism // 6) Coagulation defect

* Edwards syndrome:

→ Trisomy 18
→ 2nd MC autosomal trisomy

* Clinically:

1) IUGR, microcephaly, Prominant occiput
2) Hypertonia, Clenched Fist
3) Rocker bottom feet, overlapping fingers
4) VSD, ASD, PDA
→ MCC of death is central apneea

* Prenatal screening:

↓ a-FP, ↓ hCG, ↓ estriol

* Prognosis:

* 50% die in the 1st week of life
* 90% die by 1 yr of age

* Patau syndrome:

→ Trisomy 13

* Clinically:

1) Polydactyly, clenched hand
2) small head, abcent eyebrow
3) cleft lip ± Palat, dysplastic ears
4) Undescended testes
5) CVS, CNS, kidney malformations
→ 80% die within the 1st yr

* Dx: US + a-FP

Other inherited syndromes

** Fragile-X syndrome:
→ X-linked recessive
→ Fragile site of distal part of long arm of x chromosome

* Clinically: (« Male child »)
1) Mental retardation
2) The « large syndrome » ☺ ين ←81
- long face
- large everted ears
- Prominant forehead
- Prominant mandible
- Macro-orchidism (Big testes)
  ↳ After puberty
3) Mitral prolapse

* Diagnosis:
- Trinucleotide repeat expansion (CGG)
  ↳ More repeats → worse condition

* In females:
- less sever due to x-inactivation
- variable degrees of mental retardation and learning disability

N.B: The MCC of inherited MR → Fragile-X
The MCC of congenital MR → Down

** Prader willi:
→ Paternal mutation

* Clinically:
1) At birth: Floppy neonate
Feeding difficulty
Failure to thrive
2) 2nd yr: hyperphagia & rapid wt. gain
3) CNS: Mental delay
4) General: Short stature
small hand and feet
* Dx: DNA methylation analysis

** Angelman syndrome:
→ Maternal mutation

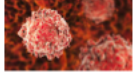
* Clinically:
1) CNS: sever intellectual disability
Microcephaly
Seizure & ataxic gait
2) Feeding difficulty, protruding jaw
3) Paroxysms of laughter
* Dx: DNA methelation analysis

** Poly-X Female:
→ 47 XXX or XXXX

* Clinically:
1) Mental retardation, increased with increased number of X
2) Infertility

** Poly-Y Male:
→ 47 XYY

* Clinically:
1) Tall male
2) Aggressive behaviour
3) Normal mentality



Haematological malignancies

* Leukemia is the MC childhood malignancy
  ↳ ALL is the MC
* Leukemias affects mainly Bone marrow although can infiltrate tissues
* Lymphoma develops peripherally but they can still involve the bone marrow

** Leukemias:

* Myelodysplastic syndrome:
  → Every MDS eventually becomes AML
  → Usually secondary to prior therapy or associated with genitic syndrome
  → Not considered Leukemia until it count for 20% of bone marrow
  → Presentation is about 7 yrs
  → Monosomy 7 or 5q deletion

Tx: stem cell transplantation

* Acute myeloid leukemia (AML):
  * Clinically: Fatigue, Pallor, Fever
    Bone pain, Bleeding (↓Platelets)
  * Predisposing conditions:
    - Congenital BM failure syndromes:
      Fanconi anemia, Black fan diamond congenital amegakaryocytic thrombocytopenia
    → Leukemia 2ry to radiation or chemo has poor prognosis
    → Down syndrome related AML has excellent prognosis
  * Genetic abnormalities:
    t(8:21), t(15:17), inv(16) & MLL
    ↳ All have good prognosis except MLL-r

Tx:
low risk of relapse: chemo: Doxorubicin
Fludarabine
High risk of relapse: BM transplant

* Chronic myelogenous leukemia CML:
  → Philadelphia chromosom t(9:22)
    ↳ BCR-ABL1
  * Phases:
    1) Chronic: weight loss, fatigue, night sweat
    splenomegaly (abdominal distension)
    ↳ CBC: Anemia, ↑WBCs, ↑Platelets
    2) Accelerated: Clonal evolution starts
    3) Blast crisis: development of AML or ALL

Tx: Tyrosine kinase inhibitor (Imatinib)

* Acute lymphoblastic leukemia:
  * Risk ↑ with: Down synd, immunodeficiency
    Ataxia telangectasia, neurofibromatosis
    → Peak incidence 2-4 yrs
  * Down ↑ risk of AML, yet ALL remains the MC leukemia in patient with down
    ↳ Here it has Bad Prognosis
  * ↑ Risk features: T-cell leukemia, age <1yr or >10
    initial WBC count 50000
  * Clinically: Bone Pain, HSM
    CNS: headache, CN Palsy, seizures, neck pain
    testes: hard, Painless mass
  * Tx: Chemo: steroids, MTX, Vincristine, mercaptopurine

** Lymphoma:

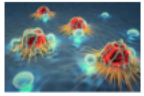
* Hodgkin:
  → Common in age >10 yr

↑Risk: FH, HIV, ataxia telangectasia, sarcoid
→ MC Type is Nodular sclerosing
* Clinically: Painless lymphadenopathy
  B-symptoms (F, NS, wt loss)
* Tx: Multiagent chemo & radiotherapy
↑ Risk of secondary cancer in survivors (AML/MDS) & (in Female breast CA)

* Burkitt lymphoma:
  → very aggressive yet responsive to chemo
  → Abdominal mass or jaw mass
  → t(8:14)
  → with Tx watch for ↓ Ca^{+2}

* Anaplastic large cell lymphoma: t(2:5)

Solid tumours



* Brain tumors:

→ the MC solid tumors
→ The 2nd MC malignancy in peds
* Clinically: N+V, headache, weight change
behavioural & sleep cycle changes
Developmental delay
Papilledema
Bulging fontanels
Cranial neuropathies, seizures
Imbalance & loss of coordination

* Alarming sign:- preference of one hand before age of 2 yrs
- change of handedness

* Commonly infratentorum (medulloblastoma)
↳ diagnosed before age of 10 yrs
* Tx: surgical excision, radio & chemo
* Complications: Neurodevelopment delay
Endocrinopathies
* diffuse intrinsic Pontine glioma: rare
↳ in brainstem, Poor response to chemo
* Pilocytic astrocytoma: Tx: total resection
* Glioblastoma multiform: Poor prognosis

* Neuroblastoma:

→ the 3rd MC
→ Tumor of sympathetic nervous system
↳ Classical presentation is suprarenal mass
* Clinically: Horners syndrome
Racoon eye
* Paraneoplastic: VIP secreting tumor
opsoclonus myoclonus ataxia
↳ Good Prognosis
* Poor Prognosis: advanced stage disease
age > 18 month
↑ risk histological features
MYC oncogen
* Investigation: catecholeamines metabolite in urine (HVA, VMA)
* Tx: surgery, radio, chemo (Poor prognosis)

* Wilms tumor (nephroblastoma):

→ MC Peds renal malignancy
* Clinically: unilateral, associated with:
aniridia, hemihypertrophy
GU- malformation
→ ↑ risk with Beckwith-Wiedmann syndrome
→ tumor has excellent prognosis
* Presentation: Abdominal mass
HTN, Anemia, hematuria
* Tx: low grade: surgery
high grade: Chemo
Radio for lung mets

* Osteosarcoma:

↑ risk: exposure to radio (10 yr Post exposure)
Hereditary retinoblastoma
→ Tp53 mutation
* Clinically: Pain, mass (in metaphysis)
* MC site: distal femer > Proximal tibia
* X-ray: Codman triangel, sunburst patteren
* Tx: surgery & Chemo (No radio)

* Ewing sarcoma:

→ Affect axial skeleton (MC Pelvis)
* Clinically: Pain, Fever, weight loss
* X-ray: onion skin appearence
* Tx: surgery, radio, chemo

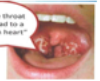
* Retinoblastoma:

→ Age of onset < 5 yrs
* Clinically: leukocorea, strabismus, nystagmus
glucoma, Periorbital cellulitis
Proptosis & Buphthalmus
* Tx: surgery, chemo, radio, laser photocoagulation
→ associated with osteosarcoma

* Hepatoblastoma:

↑ risk: Prematurity, low birth weight
→ associated with Familial adenomatos polyposis
* Clinically: Abdominal mass, weight loss
loss of appetite, ↑ AFP level
* Tx: surgery, chemo, liver transplant
* Mets: lung

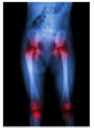
Rheumatic fever



* Auto-immune inflammatory disease involves Heart, large joints, CNS, Skin
→ Follows URTI with B-hemolytic strep
→ FH maybe +ve
→ Age: 5-15 yr
* Pathogenesis: Ab cross reactivity
* Diagnostic criteria:
2 major
1 major, 2 minor
+ Evidence of strep infection
* Major criteria:
1) Mono or Poly arthritis
2) Polyarthralgia
3) Carditis (clinical or subclinical)
4) Chorea
5) Sub Q nodules / Erythema marginatum
* Minor criteria:
1) Fever 38°
2) Monoarthralgia
3) ↑ESR, ↑CRP
4) Prolonged P-R interval on ECG
* Evidence of strep infection:
1) ASOT
2) Antistreptokinase
3) Anti hyaluronidase
4) Anti-DNase

* Notes on criteria:
→ All are reversible except carditis
→ Evidence of strep is NOT req in chorea
→ Prolonged PR is NOT considered if there's carditis
→ Arthralgia is NOT considered if there's arthritis
* Arthritis:
→ Painful, red, hot, swollen, limited mobility
→ large joints (knee, ankel, wrist, elbow)
→ Migratory
→ leaves joint with NO damage, No deformity
→ Dramatic response to salicylates
* Pan-carditis:
→ Pericarditis, myocarditis, endocarditis
→ endo: mid-diastolic murmur
Mitral regurge, MS
chronic: Mitral regurge, MS
→ Subclinical: detected by eco, No murmur
* Chorea:
→ Maybe the only manifestation
→ Abnormal movement
→ Hypotonia: - Tounge can't be protruded >5ecs
- Dysarthria
- inability to maintain grasp
- Boat shaped hands
- Pendular knee jerk
→ Emotional changes
* Erythema marginatum: Red macule, Pale centre
Not affecting face, Not itchy

* Sub Q nodules: small, hard, Painless
overlies bony prominence
* Complications:
1) Recurrence (damage heart valves, myocardium)
2) Rheumatic heart disease: incompetent valves
stenosis (adolescence)
3) Infective endocarditis
4) HF, Arrhythmia
* Investigations:
1) CBC (Anemia, ↑WBC), ↑ESR, ↑CRP
2) CXR, ECG, Eco
* Treatment:
1) Prevention of rheumatic activity:
- Benzathine penicillin IM q 2-3 weeks
2) Prevention of infective endocarditis:
- Oral amoxicillin (single ↑dose) 1 hr before tooth extraction
3) Antipyretics, Analgesia
4) Specific:
- Arthritis: Salicylates
- Carditis: Prednisone, Salicylates
- Chorea: Haloperidol
Phenobarbitone
Carbamazepine or valproic acid



Juvenile rheumatoid arthritis

* Etiology:
→ Environmental factor: Parvovirus, EBV
→ Genetic factor: HLA DR4, HLA DR8

* Pathogenesis:
→ Ag-Ab reaction → immune complex formation

* Criteria:
1) Age is < 16 yr
2) Duration is > 6 weeks
3) Arthritis in more than 1 joint
4) swollen, Pain, limitation of motion, NO Red

* Classification:
1) Poly-articular type:
- Joints: 5 or more
  small & large joints, symmetrical
  young girl
- sites: Temporo-mandibular joint
  cervical spines
  Limbs & interphalangeal joints
- systemic: low grade fever
  leukocytosis, ↑ESR, ↑CRP

2) Oligo or Pauci-articular type:
- Joint: 4 or less joints
  large joints (knee, ankel)
  Asymmetrical
- Type-1: young girl
  Iridocyclitis (inf. of iris & ciliary b.)
  ANA +ve (30%)
- Type-2: young boys
  Ankylosing spondylitis
  HLA B27 +ve

3) systemic onset:
- Joint: No symptoms or occur late
- General: spiking feve, macular rash
  ↑↑ ESR, ↑CRP
- Internal organs: HSM, lymphadenopathy
- Serositis: Pancarditis, pleurisy
- CBC: ↑↑ WBC, thrombocytosis, anemia
* Complication: Macrophage activating syndrome
  → Pancytopenia, ↓ ESR

4) Enthesitis-related arthritis:
→ Tenderness at insertion of tendon, ligament
→ At least 2 of:
- Hx of sacroiliac joint tenderness
- Acute anterior uveitis
- Male > 6 yrs
- Presense of HLA-B27 antigen
- FH of Ankylosing spondylitis or anterior uveitis

* Treatment:
1) NSAIDs → in oligo arthritis
2) steroids → intra articular → single joint
   → Topical → Iridocyclitis
   → oral → sever systemic disease
3) Methotrexate → Polyarticular
4) Biological: TNF-α blocker, Anti-IL-1
   Anti-IL-6 → systemic (after steroids)

** septic arthritis:
→ common in children < 2 yrs
→ knee is MC affected

* Organisms: staph aureus
  H. influenza (in young children)

* Clinically: Fever, anorexia, toxic look
  Red, hot, tender, ↓ range of motion
  joint effusion

* Investigations: CBC (↑ WBC)
  ESR, CRP (↑)
  Aspiration of joint space under US guide
  US for deep joints (effusion)

* Treatment: Broad spectrum antibiotic
  Joint aspiration & drainage

Say bye to Pediatrics

Sohib Shabatat