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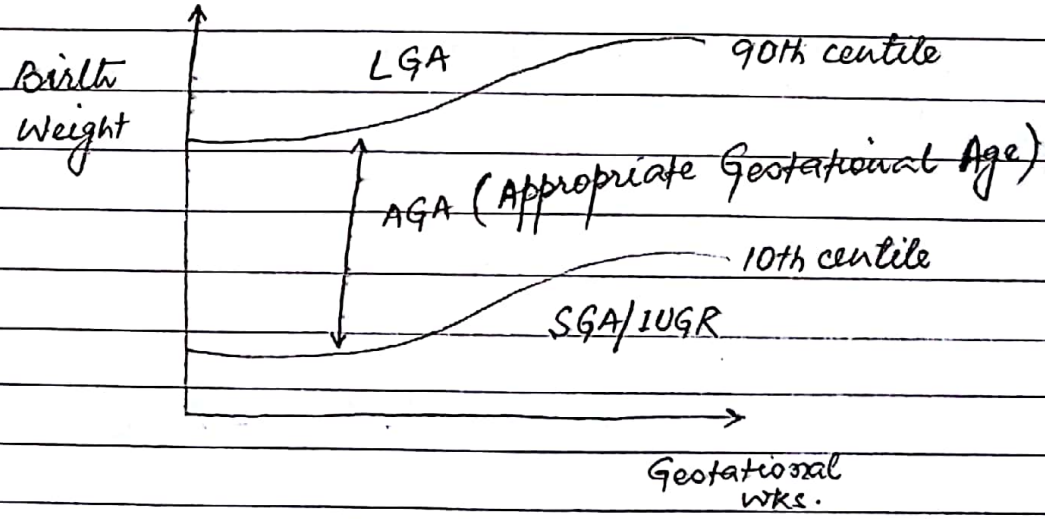
PEDIATRICS

Neonate – First 4 wk. of life.
↳ Early – 7 days, cause of death – Prematurity
Late – 7-28 days, – Sepsis.

Term – 37-42 wk gestation
Preterm – < 37wk
Post term – > 42wk

Normal wt → 2.5-4kg
LBW < 2.5kg
VLBW < 1.5kg
ELBW < 1kg
Macrosomia > 4kg.

Lubchencho chart:



Constitutional:
- LGA (large Gestational Age)
- Short stature.
- Delayed puberty.

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- All term baby has 6 frontanelle:
  - Anterior → ~~closes at 6 months~~
  - Posterior
  - 2 sphenoid
  - 2 Mastoid.

Posterior fontanel - Closes at birth.
open in 3% baby.

Anterior fontanel - diamond shaped.
2.5 X 2.5 cms.
At level
Pulsatile
closes at 18-24 months of life.

Craniosynostosis:
Early closure of all fontanel.

Complication:
- Microcephaly
- ICT ↑ → optic atrophy.
- cosmetic.

Rx - Craniectomy

Syndromes associated - Apert
Crouzon } AD
Pfiffer
Carpenter - AR

Delayed closure of fontanel:
- Rickets
- Down's
- Hypothyroidism
- Hypophosphatasia
- Cleido cranial dysostosis.

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Q. 2 wks baby, Hypotonia, hypothermia, umbilical hernia, constipation, physiological jaundice prolonged.

↳ Congenital hypothyroidism
(humeral head – Epiphyseal dysgenesis)
(46(h) epiphysis of bone)

Neonatal Screening:
- TSH ; T_4
  - Delayed rise of TSH
  - TBG deficiency (TSH(h) ; T_4↓)
  - Best time : after 48hrs (72hrs)

M/c/c of congenital hypothyroidism – 85% agenesis/ dysgenesis of thyroid gland.

Congenital hypothyroidism :
Prevalance = 1 : 2000
Girls > Boys.

Neonatal screening : Continue
TSH; T_4
- Cold temp^r
- Physiological TSH surge 48 hrs.
- APP > 48hrs to 6days.
- In OP Ghai → 3-5days

→ Sample obtained → Heelprick.
Safe area → Side prick.

- pt in shock, venous access can't get in 60sec.
- M/c other site – Near Tibial tuberosity.
  Upper end of tibia
  Lower end of femur.
- In shock → i.v. fluids ⇒ 20ml/kg bolus NS.

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Most easily assesible venous route – Umbilical Vein.

Phenylketonuria: Deficiency of Phenylalanine hydroxylase.

Phenylalanine
PKU ⊖ ≠ Phenylalanine hydroxylase.

Tyrosine
↓ Tyrosinase
DOPA
↓
Dopa quinone
↓
Melanin

↑ Phenyl alanine → Toxic to Brain → so in PKU High Phenylalanine (20 mg/dl)
- Child Mental Retardation.
  - Developmental delay.
  - Exaggrated Reflexes
  - Microcephaly.

Rx – PKU
- Supplement Tyrosine
- Restrict Phenylalanine
- Now Tyrosine essential.
- Aim: Serum Phenylalanine < 6 mg/dl.
- Lifelong.

Q. About PKU Rx, first step–
A) Stop the substrate of the enzyme
B) Supplement the enzyme.
C) Reduce the substrate of the enzyme
D) Provide deficient proteins.

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Maple Syrup Urine disease (MSUD):

- Deficiency of α-keto acid branched chain dehydrogenase.
- High Valine : Leucine : Isoleucine blood & CSF.
- Coma
- Severe acidosis.
- We do dialysis — To remove Valine : Leu : Isoleucine.

Neonatal Screening : Continue

- Tandem mass
  - Spectrophotometry → metabolic
- Cystic fibrosis
- Congenital adrenal hyperplasia
- G6PD deficiency.
- Biotin deficiency.

ANTHROPOMETRY : TERM BABY

Length — 50 cms
Head circumference — 35 cm
HC > CC but not more than 3 cm.

- HC > CC (>> 3cm) → Congenital hydrocephalus.
  → Asymmetric IUGR baby.

CC = HC → at 9-12 months
By 1yr ; CC >>> HC

Upper Segment : Lower segment (US : LS)
New born = 1.7 : 1
At 10 yrs = 1 : 1
Adults = 0.9 : 1

Achondroplasia : Short limb dwarf (US/LS ↑)
Hypothrodism : US/LS ↑ (disproportionate short stature)

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Meconium:

95% pass meconium at 24 hrs.
99% " " " 48hrs.

Cause of delayed passage meconium
- Imperforate anus
- Hirschprung / aganglionic
  ↳ Rectum biopsy.
- Meconium ileus → Cystic fibrosis.
  ↳ Small intestinal obstruction.

Q 48hrs baby has not passed meconium. Next Ix
A) CFTR gene test
B) Sweat Chloride
C) Manometry
D) Lower GI contrast study.
  ↳ △ Hirschsprung
  Treat meconium ileus.

Delayed Urine:
- B/L Renal agenesis
  ↳ Maternal oligohydramnios.
  ↓
Post. Urethral valve (also cause) ↓
  B/L pulm. hypoplasia
  ↓↓
  POTTER'S Sequence.

Potter's face – Nose pinched in
  Retrognathia
  Micrognathia
  Bag & mask → Pneumothorax.

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Q If a baby has not passed urine in 1st 48hrs, Next Ix
→ USG

Q 3 days old, c/o - Weak dribbling urine stream.
O/E - palpable distended bladder.
Δ → Posterior Urethral valve.
IOC : MCU (Micturating Cysto Urethrography).
Rx : Cystoscopic Fulguration

SGA / IUGR :
Complication : TORCH infection / Chromosal disorders.
Symptom - Asphyxia → causes MAS
↓ (Meconium Aspirated Syndrome)
PPHN (Persistent pulm. HTN of New born).
Severe IUGR - Pulmonary hemorrhage
Limited stores - Hypoglycemia, HypoCa ; HypoMg.
- Polycythemia
↳ bcoz in IUGR erythropoietin is very sensitive to hypoxia.
- Neutropenia
- Thrombocytopenia.

Q Full term small for date babies are more disposed to -
a) Hyper Ca
b) CNS infection
c) PDA
} common in preterm.
d) Hypoglycemia

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	Symmetric IUGR	Asymmetric IUGR
Cause →	Chromosal/ Torch	Maternal complications
cell no.	↓	(N)
Cell size	↓	↓
Brain	↓	(N) spared. HC > CC > 3cm
		Brain/liver ↑

PONDERAL'S INDEX:

$$\frac{Wt\ (gm)}{Length\ (cm)^3} \times 100$$

< 2 → Asymmetric IUGR
≥ 2 → AGA/ Symmetric IUGR

Normal Neonatal phenomenon:

Milia: Distended Sebaceous gland on face & nose.

Erythema Toxicum: Erythema on face & trunk 2-3 days of life.

Stork bites: Pinkish gray capillary hemangioma on back & buttocks.

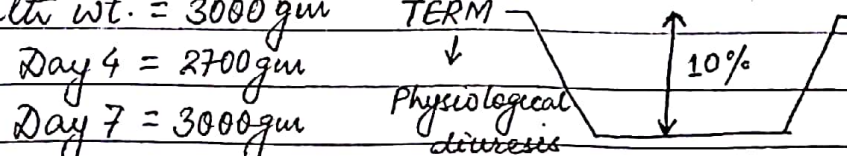
Epstein Pearl: Epithelial inclusion cyst on palate & prepuce.

Natal teeth (Pre deciduos teeth): lower incisor position.

Withdrawl vaginal bleeding: On 5th - 7th day.

Acrocyanosis (Peripheral Cyanosis): Limbs cyanosed Lips pink.

Birth wt. = 3000 gm
Day 4 = 2700 gm
Day 7 = 3000 gm



• ECFC

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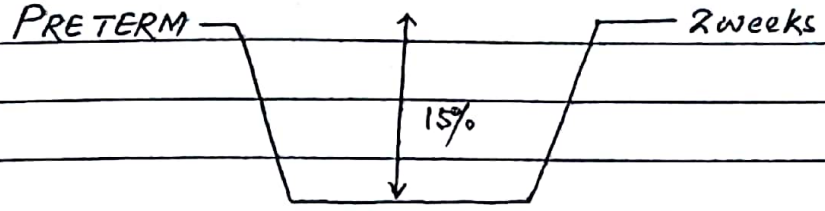

IUGR → do not lose wt (ECF compact)
↳ Wt. stable for 1-2 days then wt. gain.

Q. Not normal in a newborn?
A) Proteinuria] in ELBW
B) Glucosuria]
C) 1-2 pus cells/hpf
D) Bacteriuria

Neonatal Reflexes:

① Moro Reflex:
1st phase – Abduction of shoulder joint
Extension of elbow joint.
opening of fingers

2nd phase – Adduction & flexion.

- Appears 28-32 wks gestation.
- Adduction / complete → 36-38 wks.
- Disappears at around 2-3 months of life.
- Persistence beyond 6 months abnormal.
↓
Cerebral palsy.

- Asymmetric moro's
  - Brachial plexus injury.
  - # clavicle
  - # Humerus
  - Hemiplegia.

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Early hand preferance is always abnormal
- Hemiplegia at other side.
- 95% cases → Hemiplegia at Rt. side.

Exaggerated Moros → HIE -1

ATNR (Asymmetric Tone Neck Reflex):
Side of face – Extended
Side of occiput – Flexed.
Onset – 35 wks
Fully developed – 1 month
Duration – 6-7 month
Do not roll (Rolling start when this reflex disappear)
Disappears – 6 months

STNR (Symmetric tonic Neck reflex):
Neck extended
→ Tone ↑ UL
↓ LL

Neck flexed
↓
Tone ↓ in all limbs.

- Not present at birth.
- Appears 4-6 months of life.
- Disappears 8-12 months of life.
- Child start to crawl when this reflex disappears.

Parachute reflex:
- Not at birth
- Appears 6-7 months of life.

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- Well developed at 10-11 months of life.
- Persists life time.

LANDAU Reflex:
- Appears at 3 months of life
- Disappears at 1 year of life.

On ventral suspension – Spine strengthen / straighten.
Child get out of flexion attitude by this reflex.

GRASP Reflex:
- Appears 28 wks of gestation.
- Well developed 32 wks
- Disappears 3 months of life.

Sucking & Rooting reflex:
< 28 wks absent.
At 28 wks – Some sucking bursts.
32 wks – Appear
34 wks – Co-ordination

Q 31 wks; 1500gms; Feed – ?
A) Enteral → NG tube (Expressed Breast Milk)
B) Enteral + i.v. fluids
C) IV fluids.
D) TPN.

Q 33 wks; 1500gms; Feed – ?
↳ Enteral → Katori / Paladay / Spoon (Expressed Breast Milk)

Dr. Siddharth Sethi

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AIIMS NICU Protocol 2014:

> 34 wks – Breast feed
32 – 34 wks – Katori (Expressed breast milk)
< 32 wks – NG tube (" " ")
< 1200 gms – i.v. dextrose + minimal enteral feeds 10–15 ml/kg/day.

↓

Rapid enteral feed can cause – Necrotising Enterocolitis.

Fetal alcohol Syndrome:

- Skin folds at the corner of the eyes
- Low nasal bridge.
- Short nose
- Indistinct philtrum (groove b/w nose & upper lip)
- Small head circumferance (microcephaly)
- Small eye opening.
- Flat mid face / midfacial or maxillary hypoplasia.
- Thin upper lip.
- Septal defects < ASD, VSD.

Q. Ass. c Felal alcohol Syndrome except –

A) Microcephaly
B) Overgrowth
C) Flat face
D) Small palpabral fissure.

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PREMATURITY

Respiratory System: RDS (Respr distress Syndrome).
Chr. lung disease / Bronchopulm. dysplasia
(O_2 dependence on 4 wk of life)
Rx: Home O_2 therapy.
Prevention: Nasal CPAP.
Vit. A supplementation.
Furosemide
Keep underhydrated in ICU.

CNS: ① Apnea (>20sec); or any period if ass. c central cyanosis & bradycardia).

Q. M/c Sign of acute hypoxia in neonates
~~A)~~ Bradycardia
B) Tachycardia.

Types of Apnea: 3 types
① Central: asphyxia; preterm.
② Obstructive:
all newborn are obligate nose breathers till 4 months of life.

Q. Full term newborn
episodes of cyanosis — worsen when feed.
Seems better when crying.
↳ Choanal atresia → B/L; posterior.

③ Mixed (M/c)

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Apnea of prematurity:

Risk: [redacted] < 28wks → 100%

Onset: 1-2days ; never > 7days.

Rx:

1st step: Nasal CPAP

Methylxanthines – Aminophylline (Narrow therapeutic range)

Caffeine citrate (DOC)

↳ Wide margin of safety

Q Loading dose of Aminophylline: 5-6mg/kg.
followed by maintenance dose
1-2mg/kg every 6-8hrs.

Caffeine Citrate: loading dose → 20mg/kg
Maintenance dose → 1-5mg/kg/day

② Intracranial hemorrhage

(Only subdural hemorrhage is common in term baby).

– Capillaries in the subependymal germinal matrix is fragile; so they rupture.

Preterm; sudden pale; Shock, fontanel bulging; seizures – Intra ventricular hemorrhage (IVH)

↓

Risk IVH preterms – < 1500gm → 30%

[50% IVH ☐ in 24 hrs.
75% " " 72hrs.

IOC for newborn having seizure → Trans fontanel USG

Term; Breech → IVH

Instrumental delivery → IVH

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Prevention of IVH :
- Antenatal steroids
- Low dose indomethacin to baby.
- Prevent acidosis, infections in baby.

③ Asphyxia

Preterm	Term
↓	↓
Periventricular leucomalacia (PVL)	Parasagittal injury
Power : UL > LL	↓
↓	Spastic quadriplegic cerebral palsy.
Diplegia (upper limbs strong LL are weak)	MRI : Coronal Section - parasagittal injury.
↳ Spastic diplegia type of Cerebral palsy.	• Mentally Retarded

Q. M/c sequal of Periventricular leucomalacia in preterm
- Spastic diplegia

MRI : PVL (IOC)
- less white matter
- Shrinkage Ventricle.

- Mentally Retarded.

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Status marmoratus : diffuse neuronal loss following asphyxia.

APGAR Score : 1, 5, 10 minutes
- doesn't help in resusitation.
- 5' ; 7' ; 10' low → Worse neuronal outcome.

	0	1	2
A = Appearance	Central cyanosis/ Pale	Acrocyanosis	Pink.
P = Pulse (HR)	0	<100	>100
G = Grimace	No	Grimace	Crying
A = Attitude	Extended	Mid	Flexed
R = Respr effort	Apnea	Gasping	Crying.

NORMAL – 7-10

Moderate to severe asphyxia ; out of hospital CPR
↓
ischemia reperfusion injury
↓
Free radical damage.

Rx - Therapeutic hypothermia / Selective head cooling :
33.5°C c in 6hrs of life ; Keep for 72 hrs.
↓
preventive

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Hypoxic Ischemic Encephalopathy (HIE):
Injury to brain at severe asphyxia.

Signs	STAGE 1	STAGE 2	STAGE 3
• Loss of consciousness	Hyperalert	Lethargic	Stuporous; Coma
• Muscle tone	Normal	Hypotonic	Flaccid
• Posture	Normal	Flexion	Decerebrate
• Tendon reflexes/ clonus	Hyperactive	Hyperactive	Absent
• Myoclonus	+nt	+nt	-nt
• Moro reflex	Strong	Weak	-nt
• Pupils	Mydriasis	Miosis	Unequal, poor light reflex.
• Seizures	None	Common	Decerebration
• EEG finding	Normal	Low voltage changing to seizure activity.	Burst suppression to isoelectric
• Duration	< 24 hr if progresses; otherwise may remain (N)	24hrs – 14days	Days to weeks
• Outcome	Good.	Variable	Death, Severe deficit

DOC: Seizures in Newborn: Phenobarbitone.
(Bolus, 20 mg/kg)

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③ CVS : Hypotension ; PDA (Patent ductus arteriosus).

PDA : Preterms ⇒ Asphyxia → PGs

Term ⇒ Rubella infection → Vessel wall defect.

↓

Rx : Surgery

In preterm : Rx : NSAIDs

(Ibrufen > Indomethacin)

↓

Less nephrotoxic

If Medical management fails → Sx

C/F of PDA :

- 6-10 wks of life CHF
- Preterm baby, failure to wean off ventilator (hypoxia ; CO_2 retention)

O/E : • Bounding pulses & wide pulse pressure.

• Continuous machinery murmur at the upper left sternal border.

④ GI System : Necrolising Enterocolitis

⑤ Eye : ROP / Retrolental fibroplasia.

⑥ Hypothermia ; Hypo Glycemia ; Hypo Ca ; Hypo Mg.

⑦ Anemia ; jaundice ; Infection.

Necrolizing Enterocolitis (NEC):

R/F: ① Immature Gut.

NEC: Susceptibility of premature infants
- Reduced proteolytic enzymes
- ↑ Gastric PH
- ↓ peristalsis
- ↓ motility
- Altered epithelial membr & tight junction
- Altered bacterial flora.
- ↓ mucous coat
- Altered mucous protein
- ↑ epithelial permeability.

- Sepsis toxins
- Top fed (Cow milk)

② Mature Cocaine
③ PPIs; Anti H_2
④ Rapid advancement of feed.

Prevention: ① Antenatal steroids
- ↓ IVH
- ↓ PVL
- ↓ NEC
- ↓ RDS
- ↓ Neonatal mortality.

HUMAN MILK: ↑ proteolytic enzyme
↓ Gastric pH
↑ peristalsis
↑ motility
Less pathogenic bacterial flora

- Altered mucus coat (Improves)
- ↓ epithelial permeability.

② Trophic feeds – expressed milk
10-15 ml/kg/day.

③ Avoid PPIs ; anti H_2
④ Avoid rapidly feed advance.
⑤ Probiotics.

Q. 1 yr old infant; 10-12 episodes of watery stools/day for last 9 days. Along c Zn, which else should be advised —

A) ORS c antibiotics
B) ORS orally.
C) ORS c low lactose diet
D) ORS c low lactose diets & probiotics.

PROBIOTICS:

Prevents NEC – Lactobacillus acidophillus & Bifidobacterium infantis to VLBWs

– may use in Rotavirus:
Lactobacillus rhamnosus & Saccromyces boulardii

HMT = Hyponatremia, Metabolic acidosis, Thrombocytopenia.

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Modified Bell's Staging for NEC:

Stage Ia: Suspected NEC	; Distention ; ileus ; occult blood in stool
I_b: Suspected	Gross blood loss.
IIa: Definite	Focal pneumatosis
II_b: Definite	HMT, Diffuse pneumatosis ; Portal Venous Gas.
IIIa: Advanced	DIC ; Shock ; Peritonitis
III_b: Advanced- perforation	Pneumoperitoneum

Q. Neonate ; distended abdomen & B/L gas shadow under the diaphragm.

Δ - NEC

↳ Ia 90% preterm

2nd - 3rd wk of life.

Q Features of NEC are all except

A) Abd. distention

~~B)~~ ↑ Bowel sound

C) Pneumoperitoneum

D) Metabolic acidosis

Rx: NEC

- Stop all oral feeds
- TPN (Glucose, AA's, Lipids)
- Antibiotics (Cefotaxime ; Vancomycin ; Metronidazole)
- Stage III — may require Sx.

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Q. Child c NEC c perforation & poor general condⁿ is treated c :

A) Conservative t/t only.
B) Foark drain c glove
C) Laprotomy c resection anastomosis
D) Extracorporeal membr oxygenation.

Stage III < Stable — Laprotomy
Unstable — Peritoneal drain

NEONATAL SEPSIS:

= Symptoms + Bacteremia

	EARLY	LATE
	< 72 hrs	> 72 hrs.
R/F —	Maternal Fever < 7days	M/c/c — Nosocomial
	Foul liquor	M/c/c world — Coagulase -ve staph
	PPROM: Chorioamnionitis.	In India — Klebsiella, S.aureus.
M/c/c World:	Group B streptococcus, E.Coli (M/c)	Meningitis ⇒ CSF examⁿ
Rx —	Ampicillin + Gentamycin	— Cefotaxime + Amikacin

M/c/c India : Klebsiella, S.aureus

Sepsis screen for early diagnosis :

① TLC < 5000/cumm or > 20000
② ANC < 1500/cumm
③ PS for band cells / Immature neutrophils > 20% (I/T > 0·20) & toxic granules
④ Micro ESR (6min (N) - 3days of life) > 15mm fall in 1st hour
⑤ CRP ; Procalcitonin +ve
⑥ Lumbar puncture (In late sepsis)

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⑦ Chest X-Ray

ANC = Neutrophil + Band cells

Q Lab finding in Neonatal Sepsis except –
A) ↑ CRP
B) Leucocytosis
C) ↓ ESR
D) Toxic granulated multilobulated nuclei

Duration of Antibiotics in Neonatal Sepsis:
Bacteremia = 10-14 days
Meningitis = 21 days
Arthritis, Osteomyelitis = 4-6 wks

Temp^r regulation of newborn:
Non-shivering Thermogenesis –
Brown Fat
- Nape of Neck
- Interscapular
- Around kidneys & adrenal
- Around blood vessels (around mesentery).

Axillary temp^r:
- Normal 36.5 - 37.5°C
- Cold stress / mild hypothermia (36 - 36.4°C)
- Moderate hypothermia (32 - 35.9°C)
- Severe (< 32°C)
- Hyperthermia > 37.5°C – mostly iatrogenic.

Prevention of hypothermic: KMC (Kangaroo mother care).

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Q. All components are of KMC except:
A) Kangaroo position
B) Kangaroo nutrition
C) Early discharge & follow up.
~~D)~~ Supplementary nutrition.

Q. When to stop KMC:
- When a child reaches 2500gm wt.

Q. Delivery room temp^r – > 72°F (AAP)
> 25°C (WHO)
↳ (25-28°C)

AIIMS NICU protocols
- < 28wks / < 1000 gms → put baby in Polythene bag.
↓
reduces convection
- Incubator → Reduces convection.

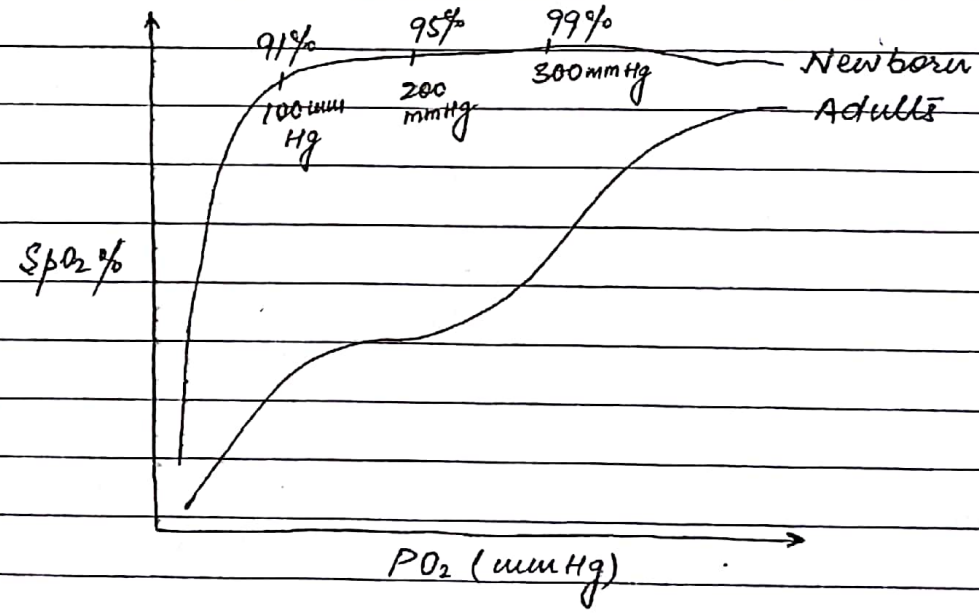
Q Mechanism of heat transfer in overhead radiant warmers :–
Radiation + ↓ convection loss.

Hypothermia:
CNS : Asphyxia
CVS : Asphyxia Cardiomyopathy.
Lungs : ARDS
Kidneys : Asphyxia ATN
Hypoglycemia ; Hypo Ca ; Hypo Mg.

Retrolental Fibroplasia / ROP (Retina of Prematurity):

R/F : Preterm.
High flow O_2.

- Proliferation, dilatation & tortuous vessels.
- Tractional retinal detachment.
- ROP stage I-V : 'plus' — Blindness.
  Stage IV : Incomplete tractional RD
  Stage V : Complete " "



WHO targets SpO_2 = 91-95% in preterm.

Inv. of ROP
BLINDNESS: Regular indirect ophthalmoscopy.

AAP ROP guidelines — (Risk <30 wks / <1500gms)

Gestation (wks)	1st visit to ophthalmologist Post menstrual (wks)	Interval (wks)
22	31	9
23	31	8
24	31	7

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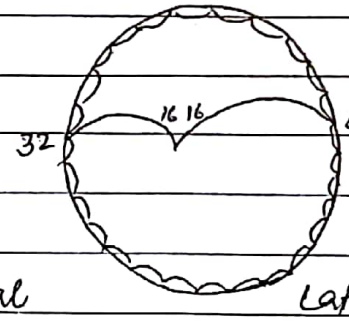
25	31	6
26	31	5
27	31	4
(28)	32	4
29	33	4
30	34.	4

Q. Pediatrician in a district hospital calls ophthalmologist for-
a) Newborn c Respiratory distress
b) New born 28 wks gestation
↳ After first visit, baby has to go to ophthalmologist, every 2wks till his/her eye look like term retina.

Retinal Vascularization: Left eye.



Vascularization of Retina starts at 16wks, finishes medially at 32 wks & laterally at 40wks.

Rx: ROP
- Laser Photocoagulation / Peripheral ablation
  - Type I ROP (all plus disease)
- Cryotherapy.
- Stage V → Retinal reattachment.

New drug — Bevacizumab → Anti-VEGF; resistant.

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Respiration :

Tachypnea : RR > 60/min.

Silverman Anderson Retraction Score :

Feature	Score 0	1	2
Chest movement	Equal	Respr lag (upper chest inspiration)	Paradoxical/ Seesaw Respr
Intercoastal Retraction	None	Minimal	Marked
Xiphoid Retraction	None	"	"
Nasal Flaring	None	"	"
Expiratory Grunt.	None	Audible c Stethoscope	Audible.

Downes scoring for Respiratory distress :

Feature	Score 0	1	2
RR (per min)	<60	60-80	>80/apneic episode
Cyanosis (Central)	None	In room air	in 40% O_2
Retraction	"	Mild	Moderate - Severe.
Grunting	"	Audible c Stethoscope	Audible c Stethoscope
Air entry (mid-axillary line)	Clear	↓ (Delayed)	Barely audible.

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M/c/c of RD → Surfactant deficiency

Surfactant → Phosphotidyl choline (65%) (Most imp.)
Phosphotidyl glycerol
Phosphotidyl inositol
Phosphotidyl ethanolamine
SP-A, SP-B, SP-C, SP-D.
Other protein.

↳ Homogenates 20 wks gestation
Amniotic fluid – 28 wk gestation
Mature levels – 95% 35 wks.

RDS risk ∝ Degree of prematurity.

Risk of RDS:
< 28 wks — 60-80%
> 37 wks — <5%
Infant of diabetic mothers, at term
Rare in IUGRs → Stress → Cortisol

Lecithin: Sphingomyelin ratio
>2 → lungs mature.
>3.5 infants of DM mothers.

CxR: RDS (lungs): Ground Glass appearance.
(White out lung).

Air bronchogram — Classic feature of RDS.
(N) CxR seen in Early RDS.

Rx : • Humidified O_2 (40-60%)
- We don't give 100% O_2 bcoz of ROP.
• Nasal CPAP : mild - moderate distress early in ELBW's
FiO_2 : Start c 40-60% ----> room air
PEEP : 5cm H_2O

• Intrabracheal surfactant - Severe ; as rescue.

- Intubate the baby — IN
Give Surfactant — SUR
Extubate the baby — E

Survanta - Bovine
Curosurf - Porcine
Infasurf - Calf.
Synthetic also available

Q. 32wks, Preterm baby in emergency C.S.
Grunting, RR = 70/min. Best management of choice
A) Humidified O_2 by hood
B) Mechanical ventilation.
C) CPAP
D) Surfactant therapy & mechanical ventilation.

Q. All occurs in RDS except:
A) Cyanosis
B) Occurs in preterm
C) More in IDM
D) Treated by 100% O_2

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Q. All true about CPAP except:
A) Initiated FiO_2 0.40 – 0.60
B) Used in apnea of prematurity.
C) Improves compliance.
Volume; FRC↑[RDS FRC↓[CV]
D) Used prophylactically in ELBW's
E) All true.

Q. Term female, Birth wt. = 3.5kg, uncomplicated delivery.
Respiratory distress after birth.
CxR – Ground glass appearance.
On ventilation & given surfactant.
But cond^n deteriorates & hypoxemia increases.
H/o sibling dying c in one week c similar complain. ECG & blood culture – (N).

↳ Δ = Neonatal Pulm. Alveolar proteinosis
↳ Autosomal Recessive
↳ Mutation in Protein B (Rarely in c)
Rx: Early lung transplantation

Postmortem Bx – Pink eosinophilic material in lung.

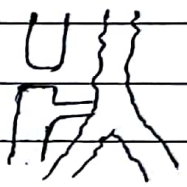
- Idiopathic: 90%
  Adult or acquired
  IgG antibodies to GM-CSF
- Secondary – 5-10%
  Hematological malignancy
  Inhalational lung ds.
  Silicon
  Titanium oxide.
- Congenital: 2%

CT scan — Crazy paving pattern
(Prominant intraalveolar septae).

℞ — Early lung transplant in neonates.
Adults → Broncho alveolar lavage.

Respiratory distress : Newborn

Tracheo-esophageal fistula (TEF)



M/c type — Type C
Distal TEF
Esophageal atresia
- Cyanotic newborn & frothing
- Aspirates gastric juice lead to pneumonia.
- Not a surgical emergency.

℞ - Keep him propped up.
- Suction catheter in upper blind esophageal pouch.

Diaphragmatic hernia:
85% cases — left.
pushed heart & trachea — to opposite side.
- Scaphoid Abdomen
- Barrel chest
- Mediastinal shift to right.
- Apparent dextrocardia
- Peristalsis on left chest.

Diaphragm — develops from septum transversum & pleuroperitoneal canals.
↳ fail to close on left side
(Bochadelek hernia)
- <5% are B/L

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Q. Cause of death in Congenital Diaphragmatic Hernia:
A) Septicemia
~~B)~~ Pulmonary hypoplasia – Left
C) Hemorrhage.

Bag & mask is absolutely c/i bcoz it will cause further abdominal distension.
↓
So elective intubation is done.

Baby born → Bag & mask c 100% O_2
↓
diaphragmatic hernia
↓
Intubate
Next step – NG tube to decompress the gut.

Born baby → Diaphragmatic hernia
↓
Intubated
↓
Heart further to right
Next Step – Remove the tube
& Reintubate.

Q. Most imp. Prognostic factor in Congenital diaphragmatic hernia:
~~A)~~ Pulm HTN – Persistent (PPHN)
B) Age
C) Time of Sx
D) Size of defect.

Prevention : PPHN
- Elective intubation
- HFOV > 300–600/min

Rx : PPHN
- iNO → Pulm. vasodilator.
- Sildenafil PDE-V inhibitor

New drug: Bosentan, Ambrisentan — Endothelin antagonist.
- PGI_2 ⇒ Iloprost
- Amlodipine ; ECMO (Extracorporeal membr oxygenation)

Transient Tachypnea of Newborn (TTNB):
R/F : • Term by C.S. (lungs are wet) ∵
• Macrosomia ∵
• Excessive maternal sedation.
• Precipitous labour.

CxR – a prominent horizontal fissure
↳ Most specific feature.

• Benign condn
• Self limited : 48–72hrs.
• FiO_2 requirement < 0.40
• Never require mechanical ventilator.

Meconium Stained Liquor :
Meconium is marker of Perinatal Hypoxia

Glottis open → meconium passed

Parasympathetic Stimulation → Anal sphincter relax, Peristalsis ++

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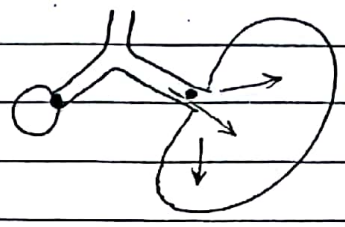
Perinatal hypoxia : Common in postterm
bcoz of UPI.

Meconium :

Physical –



– ball valve mechanism
– air leak 20-30%

Chemical – Irritant → pneumonia
↳ Impair surfactant func^{n}.

Biological – Good culture media

Meconium Stained liquor.
↓
Baby born
↓
Vigrous ← Tone is good
Resp^{r} effort is good
HR > 100/min
↓ yes / No
Transfer the baby to mother
Catheter in the nose
↓
PPV @ 100% O_2.

Q Sequence of Resuscitation
A) Mouth → Nose
B) Nose → Mouth
C) Mouth → Nose → Trachea

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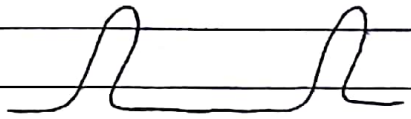
Intra uterine TORCH infections:
Others — HIV, HBV, Varicella, Syphilis.

Common features:
① - Asymptomatic

Rubella: Transmission - 2 peaks



10-12wks ↓ CRS (congenital Rubella Syndrome).

Delivery ↓ Rubella >16wks risk CRS <1%

② - If Symptomatic
- SGA; failure to thrive
- Anemia; thrombocytopenia
- Hepatosplenomegaly.
- Unexplained Rash & cholestasis.

△ - IgM (△stic of infection)
IgG persisting beyond 6-9 months

HIV in infants
- Infants can't make IgM-HIV or IgA-HIV.
- Maternal IgG-HIV can persist in the baby for 18 months.
- HIV < 18 month: Diagnosis.

Best: DNA & PCR
P24 assay
Culture is difficult.

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Intracranial calcification & Chorioretinitis] M/c/c – Toxoplasmosis – CMV

Toxoplasmosis	CMV
- 25-50%	- Periventricular Calcification.
- Choroid plexus calcification.	- Atrophy of brain. (Cortical).
- Subependyma & Caudate nucleus calcification.	- Hydrocephalus ex vacuo.
- ~~Hydrocephalus~~ Hydrocephalus	- M/c/c of Non syndromic SNHL
- Seizures.	- Seizures
	- Microcephaly
	- Mental Retardation.

Q. Pregnant lady; no complain. Mild cervical lymphadenopathy in 1st trimester. Prescribe Spiramycin (prevent Vertical Transmission) but she was non compliant. Baby born c hydrocephalus & intracerebral calcification.
↳ Toxoplasmosis

DOC: Rx: Pyrimethamine + Sulfadiazine.

Δ: Best: IgM Immunosorptive assay.

Sensitivity of ELISA: IgA >> IgM

Q. True about transplacental CMV infection:
– It is M/c/c of non-syndromic SNHL.

Q Doesn't stablish Δ of CMV in neonate –
A) Urine culture of CMV
~~B~~) IgG CMV antibodies in blood
C) Intra-nuclear inclusion bodies in hepatocytes (Owl-eye)
D) CMV viral DNA in blood by polymerase chain reac^{n}.

Best specimen – Urine culture & Saliva.

CMV disease: Retinitis, colitis, pneumonia.

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Rx : CMV

DOC : i.v. Ganciclovir (Severe, child, pregnant)

- Oral ; Prodrug → Valganciclovir
- Resistant to oral Foscarnet.

Congenital Syphilis :

EARLY	LATE
- C in 1st 2yrs of life.	- After 1st 2yrs.
- Mucocutaneous rash/rhinitis (Snuffles).	Hutchinson's Triad:
- Lymphadenopathy.	① Hutchinson teeth/ mulbery molars - 1st lower molars.
- Hematological (Autoimmune Anemia)	- Saddle nose, Frontal bossing, Olympian's brow, Higoumenaki's Sign (sternoclavicle prominence)
- Renal lesion	- Rhagades
- Skeletal (Osteochondritis, metaphysitis, Periostitis)	② Interstitial Keratitis
- Glaucoma	③ Nerve deafness. (SNHL)
- Pseudoparalysis of Parrot.	- Clutton's joints (painless joints) ↳ Risk of injury.

Pseudoparalysis :

M/c/c - Scurvy
- Early syphilis
- Osteomyelitis
- Septic arthritis.
- Hypokalemia → Hypotonia.

DOC - Penicillin G
→ 10-14 days

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Rubella Syndrome:
Triad — Microcephaly (Mental Retardness)
PDA
Cataracts.

M/c eye manifestation of Rubella — Salt & pepper fundus
Calaract
Glaucoma
Micro-ophthalmia

HEART — PDA
Peripheral pulm. stenosis
VSD
ASD (Rare).

Q. Rubella embryopathy except.
A) Deafness
B) MR
C) AS
D) PDA

Q. True about Rubella embryopathy except:
A) Diagnosed when IgM antibodies in child.
B) Infection after 16 wks results in major congenital anomalies.
C) Deafness, heart disease, Cataract.

Q. Hypoplastic limb — Varicella (Chicken pox) embryopathy during pregnancy.

Varicella Embryopathy —
- Skin Rash
- Optic nerve hypoplasia
- Brain – Cortical atrophy.
- LS plexus – Aplasia / hypoplasia limbs.

Mother get chickpox – 5 days before delivery
or, c in 2 days of delivery.
↓
Baby chicken pox illness.
↓
Prevention: Varicella Zoster Ig to the baby.
↓
even after 120 hrs of exposure.

Q Pregnant; HBsAg +ve : no gaundice
HBeAg +ve ⇒ 90% chance baby is carrier
& later in life Portal hypertension
↓
Ascites, Spleenomegaly, Varices

HBeAg -ve ⇒ Anti HBeAg +ve
then 10% chance of Vertical transmission

Give HBIg baby to 12 hr of life
HBV vaccine to baby c in 24 hrs of life.

Pricked by HBsAg +ve patient
Are you immunized?
~~Anti HBsAg~~ · Anti HBs Ab titre
Good >10 mIU/ml ; High risk >100

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Incomplete vaccination / Not know titre

↓

HBV + HBIg

a/c to CDC guidelines.

If titre is good — don't do anything.

'CDC' HBV DNA load > 1000 IU/ml

↳ Can't join Surgecal branch.

Neonatal jaundice

Heam

↓ Hemooxygenase → CO_2 ⊖ Et Co. — hemolysis.

Biliverdin → Metalloporphyrin

↓ B. reductase

Bilirubin

1gm of Hb = 34 mg of bilirubin

1gm/dl Albumin binds to 8 mg bilirubin

- Unconjugate bilirubin passes BBB & cause jaundice & Kernicterus.
- In 1st 2wks, BBB is not developed properly.
- (N) S. Albumin = 3.5-5.5 g/dl
- Healthy term baby can bind 24-25 mg/dl bilirubin.
- Sick, preterm, risk factors can go into early Kernicterus.
- In adults ammonia cross BBB in hepatic encephalopathy.

- Healthy Baby (< 1000g) → We start phototherapy (5-7 mg/dl) ← Bilirubin

↓

Sick baby → Bilirubin (4-6 mg/dl) — we start phototherapy.

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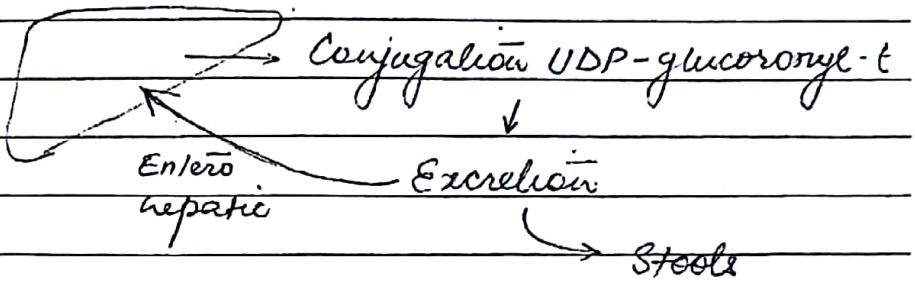
Bilirubin
↓
uptake by liver (Y-ligandin uptake)



> 2-2.5 mg/dl in adults ⇒ Yellow Sclera.

Kramer's zone:
progression of jaundice in new born is cephalo-caudal
(Bilirubin)

Zone I (5 mg/dl) →
Zone II (10 mg/dl) →
Zone III (12 mg/dl) →
Zone IV (15 mg/dl) →
Zone V (>15 mg/dl) →
↳ Danger Zone.

6 causes of physiological jaundice:

① Life span of RBC is less.
② Hematocrit more
③ Newborn deficient in Y-ligandin.
④ UDP glucuronyl-t deficient in newborn.
⑤ Excretion reduced.
⑥ Enterohepatic circulation ↑.

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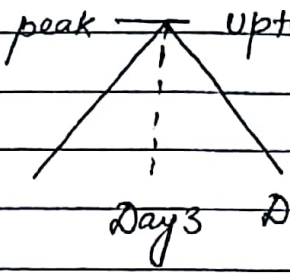
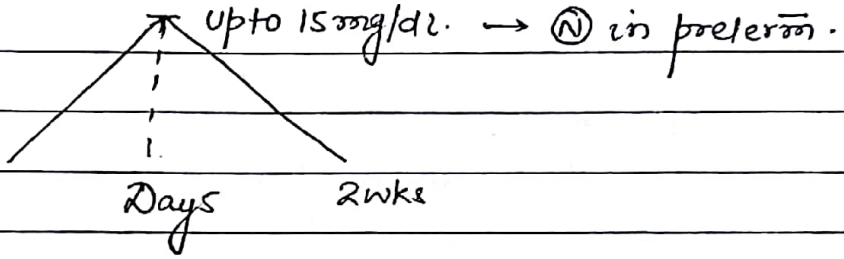
Physiological jaundice:

jaundice in term babies.



Preterms — have more jaundice.



Pathological jaundice:

- Hemolysis.

M/C/C — Incompatibility Rh/ ABO [O mother ; baby A/B]
- RBC membr defect.
- RBC enzyme defect.

Def: jaundice c in 24 hrs of life.

* Rh -ve mother ; previous abortions
we take cord blood samples.
- Rh status of baby. → If +ve
- Hb → 10 mg/dl
- Bilirubin → 5mg/dl
- Peripheral Smear (P/s) +ve
- Direct Coomb's test (DCT) → +ve

↓

means Severe hemolysis

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Rx → Exchange transfusion at birth.

① jaundice c in 24 hrs of life.
② Reaching 20mg/dl
③ Rate rise > 0.2mg/dl/hr.
④ Persisting beyond
- 1 wk terms
- 2-3 wks preterm.
⑤ Clay stools.

Phototherapy:
- Any jaundice on day 1 of life start phototherapy.

Serum bilirubin cut off.

	Phototherapy		Exchange transfusion	
	Healthy babies	Babies c R/F	Healthy babies	Babies c R/F
Day 1	← Any visible jaundice →		260(15)	220(10)
Day 2	260(15)	170 (10)	425(25)	260(15)
Day 3	310(18)	250 (15)	425(25)	340(20)

R/F ① Gestation < 35 wks / wt. < 2kg.
② Sepsis
③ Hemolysis
④ Asphaxia
⑤ Sick baby.

Principle:
- Structural isomerazation
  - Bilirubin → LUMIRUBIN
- Photo isomerization
  - 4Z 15Z ↔ 4Z 15E (Soluble).
- Minor pathway → photo-oxidation.

40 cms away, falls @ 4-6 mg/dl/day.

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- Phototherapy occurs at 425-475 nms of blue green light.
- Irradiance → 6micro Watt/cm²/nm
- Intensive > 30 micro watt/cm²/nm.

Q. AIIMS Nov. 2013
Which does not effect the efficacy of phototherapy?
A) Types of phototherapy lamp.
B) Skin pigmentation.
C) Spectral radiance of incident light
D) Initial bilirubin levels.

Complication:
- Hyperthermia; insensible losses
- Hypocalcemia ♀
- Diarrhoea.
- Cover eyes & genitals
  - Retinal damage
  - Mutations.
- Phototherapy is C/I in conjugated jaundice
  ↓
  Bronze baby Syndrome.
  (Skin, Urine).

Exchange Transfusion:
- Double volume exchange transfusion.

Blood vol. of new born = 80 ml/kg ♀
→ 2 x 80 ml/kg = 160 ml/kg.

- Transfuse fresh. (<7 days) whole blood.
  - Reduces bilirubin by 85%
    ↓
    Not 100% because bilirubin is also in tissue.

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• Albumin ↑ ~~efficiency~~ efficiency.

Complications:
- Infections
- ACD (Acid citrate dextrose)

↓

Bicarbonates

1 mole of citrate ⟶ 3 mole of Bicarbonate.

- Metabolic alkalosis
- Hypokalemia, hypocalcemia.
- Old blood → Hyperkalemia, metabolic acidosis.

Persistent jaundice:

Cause — Hypothyrodism
Breast milk jaundice
Hematoma (Cephalhematoma, IVH)
C Clay colour stool (Cholestasis)
Crijjler - Najjar Syndrome type II (Milder form)
↳ deficiency of UDP-glucuronyl-t

Crigglер-Naggar Syndrome type 1 — Very Severe
↳ Absent UDP-glucuronyl-t.

Pathological jaundice

Breast milk jaundice	Breast feeding jaundice.
- Onset → day 14	- Onset → Day 3.
- Some mother have pigment B ⊆ inhibit conjugation.	- In primigravida.
- Day 14 = 20-30 mg/dl	- Starvation stimulates entero-hepatic circulation.
- May Kernicterus.	
- Persists 4-6 wks.	

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Rx: Temporary interrupt 48-72hrs. Meanwhile give formula milk.

Rx: Ensure feeding.

Q Ture about jaundice in newborn (neonates) is –
A) Can be seen after Ventouse delivery.
B) Physiological jaundice seen c in 48hrs of birth.
C) Increased conjugated bilirubin leads to kernicterus
D) Breast milk jaundice is maxm in 7days of birth

Neonatal cholestasis:
Neonate, jaundice & clay stools.
– Direct bilirubin > 2mg/dl or >20% total bilirubin

Medical – Common.
Neonatal hepatitis → CMV
Sepsis
Galactosemia
α-1 antitrypsin deficiency.
Neonatal hemochromatosis.

Surgical – Extra hepatic biliary atresia (EHBA)
Rx – Kasai's surgery c in 8wks of life otherwise 80% die.
↓
M/C/C indication of liver transplant in babies → EHBA.

Q Which is an ominous sign in a 10day old newborn?
A) Unconjugated hyperbilirubinemia
B) Conjugated "
C) Failure to gain wt.
D) Doll's eye reflex
↳ Normal in 1st 10 days of life.

GGT → Gamma Glutamyl transferase.

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Medical – Common

Surgical – EHBA
- GGT ten times higher in Surgical causes.
- DO USG → Shows Intra hepatic biliary radicle (IHBR) are dilated.
- Triangular cord Sign

- HIDA nuclear Scan:
HIDA dye not seen in gut, even in delayed images; white in hepatitis excretion of dye.

Best test – Liver biopsy.
↓
Shows dilatation & proliferation of intrahepatic bile duct.

Before doing HIDA Scan we have to give phenobarbitone 2 to 3 days before test.

Gold Standard – Preoperative Cholangiography.

Alagille Syndrome –
- Cholestasis
- [redacted] AD
- Bile duct paucity Syndrome.
- M/c heart disease → Peripheral pulm. stenosis.
↑
Rubella

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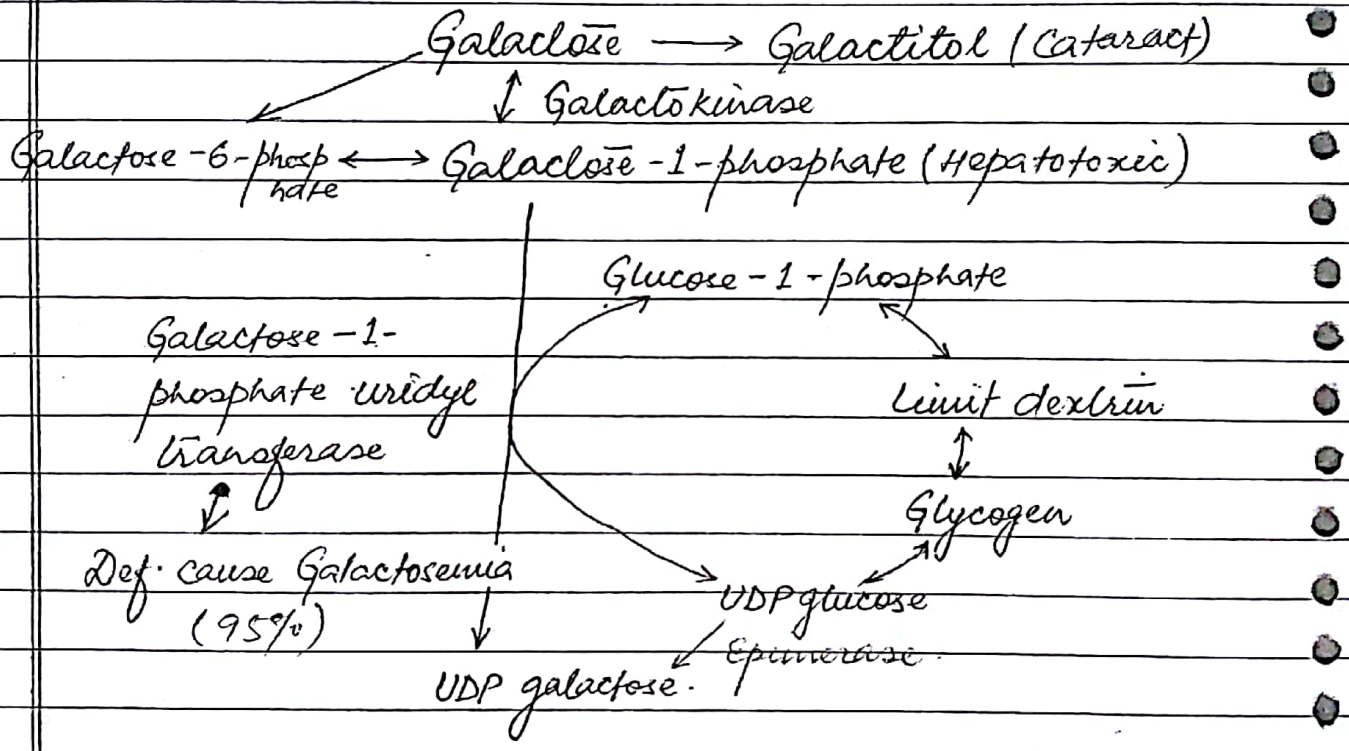
Q. 1 month old child present c conjugated bilirubinemia & intrahepatic cholestasis. On liver biopsy staining c PAS, red coloured granules were seen inside the hepatocytes. Probable diagnosis is —

A) α-1-antitrypsin deficiency.
B) Congenital hepatic fibrosis
C) Hemochromatosis
D) Wilson's ds.

CMV — Intranuclear "Owl eye" inclusion bodies.



- He get jaundice, bleeding, hypoglycaemia, PTT↑.
- Galactitol cause cataract.
- Rx — Lactose free milk.

- Galactokinase deficiency → Only cataract
  No liver failure
- Fructokinase def. → Causes Benign Fructosuria
  ↳ No symptom.

Fructose-1-phosphate leads to liver failure.

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Q. M/c/c of Neonatal Cholestasis:
A) EHBA
B) Neonatal hepatitis
C) Choledochal cyst.
D) Physiological

Q. Neonatal cholestasis seen in -
A) Chronic hepatitis (>6month)
B) Hep. B
C) Galactosemia
D) Rh incompatibility (cause ● unconjugated jaundice)

Q. Pregnant lady, HBsAg +ve, No jaundice.
↓
next step → HBeAg

- HBeAg +ve (90%)

Carrier HBsAg → Portal HTN
↓
Triad - Ascites
Splenomegaly
Varices.

- HBeAg -ve
anti HBe Ab ⊕
↳ 10%

Immunization [HBIg - baby c in 12hrs of life
HBV - baby in 24hrs of life
↓
prevent Vertical transmission

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CDC guidelines:

Pricked by HBsAg ⊕ patient

↓

Are you immunized?

↓

Anti HBsAG titre

Good > 10 mIU/ml

High risk > 100 mIU/ml.

Don't know titre → Incomplete

↓

HBV + HBIg

If Good titre → Nothing is to be done

Q.

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~~New born, Respiratory distress:~~

Neonatal Seizures:

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Neonatal hypoglycemia:

Causes:

- Limited stores → Preterm, IUGRs
- Stress → Sepsis
- Polycythemia
- Galactosemia – liver failure.
- Low cortisol – CAH (Congenital Adrenal Hyperplasia)
  Hypopituitarism.
- Low ACTH; Low LH/FSH → Micropenis.
- Hyperinsulinism – In infants ☆ diabetic mother.
  ↳ Foetus ☆ β-cell hyperplasia.
  ↓
  Foetus Blood glucose ↓
  ↑
  Pedersons hypothesis
- Insulinoma; Nesidioblastosis
- Beckwith weidman syndrome.
  ↳ Hemihypertrophy of limb
  ↳ Macroglossia
  ↳ Risk of wilms
  ↳ Hyperinsulinism

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Q. A term baby to a diabetic mother, few hours after birth was lethargic & his blood glucose was 38mg%.
What should be done next -
A) Give 10% dextrose orally.
B) 10% dextrose i.v. - Bolus 2ml/kg → Glucose drip
C) Give expressed breast milk — GIR ↓ 6-8 mg/kg/min
D) DO exchange transfusion.

Neonatal hypoglycemia:

Symptomatic	Asymptomatic
→ Bolus 2ml/kg 10% dextrose f/by Glucose drip 6-8 mg/kg/min	→ Blood glucose < 20mg/kg ↓ Glucose drip (6-8mg/kg/min)
	→ Blood glucose = 20-45 mg/dl ↓ Breast feed ; 1hrly.

A baby of glucose drip & he gets seizures
how to ↑ glucose rate
- Upto 12.5% through peripheral vein.

Max^{m} glucose infusion rate (GIR) = 12mg/kg/min.
Emergency drug = i.m. Glucagon.
for hypoglycemia ↓
Glycogenolysis + Gluconeogenesis.

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Q. Glucagon is effective for Mx in persistent hypoglycemia in all except.

A) Large date for baby.
B) Nesidioblastosis
C) Galactosemia
D) Infant of diabetic mother.

Q. 1 yr old, hypoglycemia & hepatomegaly
No jaundice.
Hypoglycemia doesn't respond to glucagon.
↓
Δ → Von Gierke's (Glycogen storage ds - I)

Glycogen —— --- Glucose-6-PO_4

Von Gierke's ⊖ → Glucose-6-phosphatase.
↓
Glucose

Types of Glycogen storage disorder:
V = Von Gierke's → Liver primary
P = Pompe's ds → Heart primary; Cardiomegaly. Large QRS complexes.
C = Cori → Debranching enzyme.
A = Anderson → Branching " deficiency
M = Mc Ardles → Muscle phosphorylase deficiency.
Harding =
Tom =

jaundice never occurs in Glycogen storage ds.

Rx pompe's : Enzyme replacement therapy.

Enzyme absent in pompe's → lysosomal α-1,4-glucosidase
— Also called Acid & neutral maltase.

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Muscle affected in GSD:

Calcium:

(N) S. Ca^{2+} → 9-11 mg/dl.

Neonatal hypocalcemia:

- S. Ca^{2+} < 7mg/dl
- ~~Basis~~ Best index of body calcium
  ↓
  Ionized < 4 mg/dl (or) < 1 mmol/L

- Tetany is rare in infants.
- Tremors, seizures, jitteriness.
  ↳ Tremulousness is stimulus sensitive.
  ↳ Can stop on passive restrain.

Early Hypo Ca^{2+}	Late Hypo Ca^{2+}
Causes – Prematurity	Cause – Feeding c phosphate
Asphyxia	rich milk (cow milk)
Infant of DM mother.	
↳ Test:	
Blood glucose	
+ S. Ca^{2+} & S. Mg^{2+}	

Good Ca^{2+} Supplement ($Ca^{2+}/P > 2$)

100 ml	Cow milk	Breast milk
Ca	118mg	34 mg
PO_4	100mg	15mg

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Advantages of Breast milk:
- protects against late onset hypocalcemia.
- protect against pneumonia
  NEC (Necrotizing Enterocolitis).
  Allergy, Eczema, asthma.
  Rota virus diarrhoea.
  Bronchiolitis (IgA-RSV)

Q. Milk deficient in:
a) Iron & Vit. C

Q Breast milk has enough iron & Vit. C for 6 months.
↓
∴ Scurvy never occurs in 1st 6 month of life.

Q. If baby on exclusive breastfeed for 2yrs.
↓
Iron deficiency anemia.

Q APP recommends Vit. to all infants — Vit. D drops.
- RDA Vit. D infant 400 IU/day
- Breast milk 25 IU/L.

M/c/c of Hypo Ca^{2+} in infants = Maternal deficiency of Vit. D.

25 [OH] Vit. D = Status.

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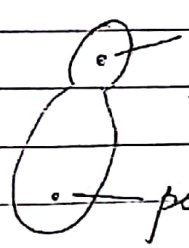
Infant of DM mother:

- Can be stillborn; preterm
- Macrosomia
  - ↳ Bcoz of Hyperinsulinism
- Linear growth in utero depend upon insulin & or Insulin like growth factor.
- IUGR → White's classification class F/R
  mother → Placental vasculopathy.
- Hypoglycemia
- Hypo Ca^{2+}, Hypo Mg^{2+}
- Neonatal jaundice
- Polycythemia → Renal Vein Thrombosis (RVT)

Q. ~~All~~. Not seen in infant of diabetic mother:
↓
Hyperglycemia.

Anomaly:
(M/c)- CVS - 8.5% [VSD; HOCM] Assymetrical septal hypertrophy.
- Neural tube defect → 5%
- Lazy left colon syndrome → Pseudoobstruction of colon
- Sacral agenesis / Caudal regression Syndrome
  - ↳ Most specific.

NEURAL TUBE DEFECT (NTD)



Ant. neuropore → closes at 25 days of IUL
Failure of closure Causes NTD
- Anencephaly
- Encephalocoele

post. neuropore → closes at 27 days of IUL
- Spina bifida
- Meningocoele
- Meningomyelocoele
  - M/c site → lumbo-sacral

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Anencephaly:

- Ant. neuropore fails to close.
- No brain, absent of part of hind brain.
- Earliest abnormality diagnosed by USG (10-12 wks) of gestation.
- Most severe NTD
- Don't resuscitate
- Mostly post-term.

Herniation of brain tissue — Encephalocoele.

Lumbo-sacral myelomeningocoele:

Complication:

- Paraplegia / Paraparesis
- Neurogenic bladder → CKD.
- Constipation (Severe)
- Associated hydrocephalous. (obstructed)

↓

Bcoz of Arnold-Chiarri type II malformation

⇓

MRI Brain

Ruptured myelomeningocoele:

Best test — Blood culture.

- Cover c Normal saline soaked gauze.
- 95% of neonatal meningitis have Leukemia.

M/c/c of meningomyelocoele — Folic Acid deficiency.

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Folic acid deficiency:

- −1/+3 months conception − start folic acid.
- 400 mcg given.

Recurrence:

1 child − 3.5% chance
2 child − 10%
3 child − 25%

To prevent recurrence → Folic acid 4mg
↓
reduces risk by 75%.

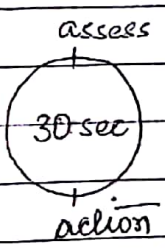
△ : ① USG
② Amniocentesis
↳ Acetylcholinesterase
& α-fetoprotein are markers.

α-fetoprotein in mother serum is marker of Neural tube defect.

Resuscitation:

NRP Guidelines 2015:

T = Temp^r
A = Airway : position neck, suction
B = Breathing
C



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Indication of Bag & mask c 100% O_2:

- Apned / gasping ~~~~ after initial steps.
- HR < 100/min after 30 sec PPV.
- Central cyanosis despite 100% O_2.

- Chest compression if HR < 60/min, falling after 30 sec PPV.
- Chest compression : Bag mask = 3:1

In 1 minute = 120 events
90 Chest compression & 30 bag & mask.

Compression to Ventilation ratio:

- Children / Infants — Single rescuer 30:2
  2 rescuer 15:2

- Adults — 1 or 2 rescuer 30:2

CPR sequence → CAB

Drugs for resuscitation:

1) 0.9% Nacl 20ml/kg bolus → shock
2) 1:10,000 epinephrine 0.1 - 0.2ml/kg
   ↓
   If HR = 0 or falling.
3) I.V. $NaHCO_3$ — documented metabolic acidosis.
4) I.V. Naloxone — mother opioid addict.

Targeted preductal SpO_2 after birth:

1 min = 60% - 65%
2 " = 65 - 70 %
3 " = 70 - 75%

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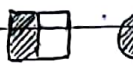
4 min = 75 - 80%
5 min = 80 - 85%
10 min = 85 - 90%

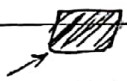
GENETICS

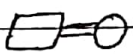
 → Heterozygotes for AR carrier

 → Carrier for sex-linked recessive

 → Death

 → Abortion of stillbirth
Sex unspecified.

 → Proband.

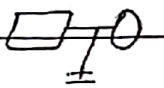
 → Consanguinous marriage

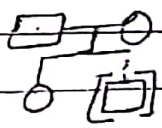
 → Dizygotic twin

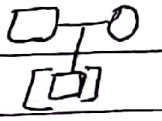
 → Monozygotic twin

 → azoospermia

 → Endometriosis

 → Infertility.

 → Adopted in

 → Adopted in

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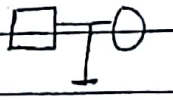
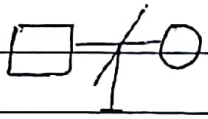
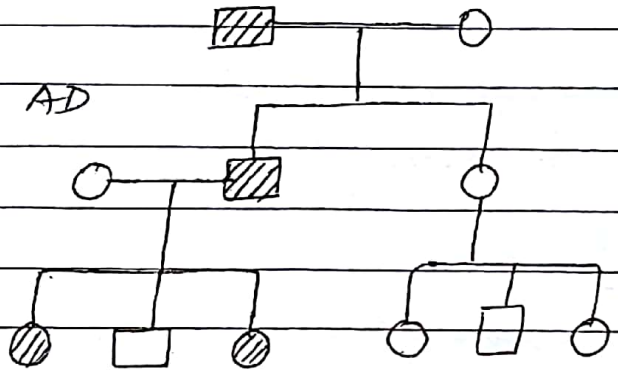
 → No children for choice or reason unknown

 → Vasectomy

 → Tubal

 → Divorce.



Autosomal dominant:

D = Dystrophy myotonic → distal myopathy.
O = Osteogenesis imperfecta
M = Marfan Syndrome
I = Intermittent porphyria
N = Noonan Syndrome → Turner phenotype XX & XY.
A = Adult PKD, Achondroplasia
N = NF (Neurofibromatosis)
T = Tuberous sclerosis
VH3 = Von Willibrand Syndrome
Hutington chorea
Familial Hypercholestrolemia
Hereditary spherocytosis.

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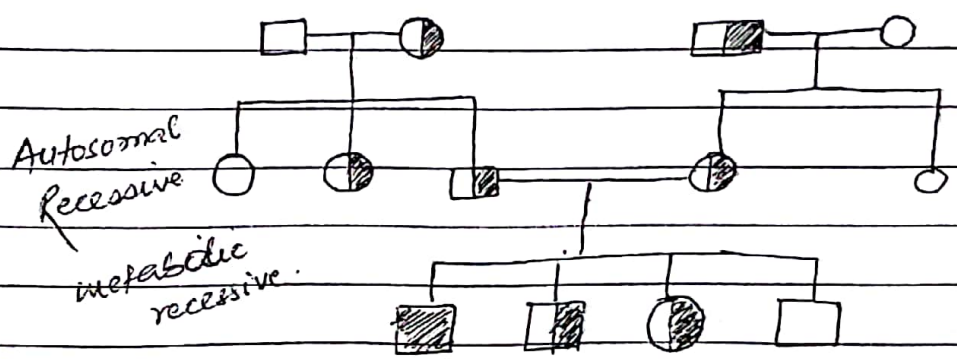
Turner Syndrome	Noonan Syndrome.
- 60% case - XO	AD
- Webbed neck	XX = XY
- Cystic hygroma	
- Lymphedema of hand & feet.	
- Primary amenorrhoea. during puberty.	
- Streak ovaries	
- Cubitus valgus.	
- MR rare.	- 25% MR
- M/c heart ds - Bicuspid aortic valve ∴ out stenosis ↳ Half to one-third. 20% coarctation.	- Valvular pulm. Stenosis. HOCM ASD.
- Girls infertile	- Girls are mostly fertile. - Boys - Cryptorchidism. - Clotting factor deficiency.

Q. Chance of child being not affected if both parents are affected ∴ Achondroplasia is -

A) 0%
~~B) 25%~~
C) 50%
D) 100%

$AA^c \times AA^c$

(AA) AA^c A^cA A^cA^c



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Autosomal Recessive:
- Cystic Fibrosis
- α_1 AT defficiency
- Wilson's ds
- Haemochromatosis
- Friedrich's ataxia
- Gaucher's ds
- Niemann's pick ds.
- Tay Sach ds.
- Hurler's Syndrome.

Mucopolysaccharoidosis (MPS)
- Child c noisy breathing
- Coarse facies.
- Ab (n) accumulation of glucose aminoglycans.
- Gargolism
  - ↳ Chronic rhinitis.

Mucopolysaccharoidosis type I → K/A HURLER's Syndrome
↓
defficiency of L-Iduronidase.
Rx - Enzyme replacement therapy.

HURLER	HUNTER Syndrome.
- MPS-I	- MPS-II
- AR	- XL recessive
- Corneal clouding	- Cornea clear.

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Enzyme Replacement therapy:

1st to be treat → ① Gaucher's ds
(β glucocerebrosidase)
↓
Company name — GENZYME
CER-zyme.

② Pompe's
③ Hurler (MPS-I)
④ MPS-VI (Maroteux-Lamy)
⑤ X-linked recessive Fabry's
(CNS & kidney problem)
↓
Lysosomal disorder.
Cherry red spot macula seen.

Gene therapy:

1st to be treated by Gene therapy → X-linked recessive
Severe combined immunodef. (SCID)
(Adenosine deaminase deficiency)

British guy British lady
Chance of child having cystic fibrosis

(Uk/ Europe/ askenazi jews → 1/25 carrier cystic fibrosis.

→ $\frac{1}{25} \times \frac{1}{25} \times \frac{1}{4} = \frac{1}{2500}$

$AA^c \times AA^c$

$AA^c \cdot A^cA \; AA^c \; AA$

1/4

British guy British lady (brother died of cystic fibrosis)

Chance of child to have cystic fibrosis

$\frac{1}{25} \times \frac{2}{3} \times \frac{1}{4} = \frac{1}{150}$

$AA^c \times AA^c$

$A^cA^c \; A^cA \; AA^c \; AA$

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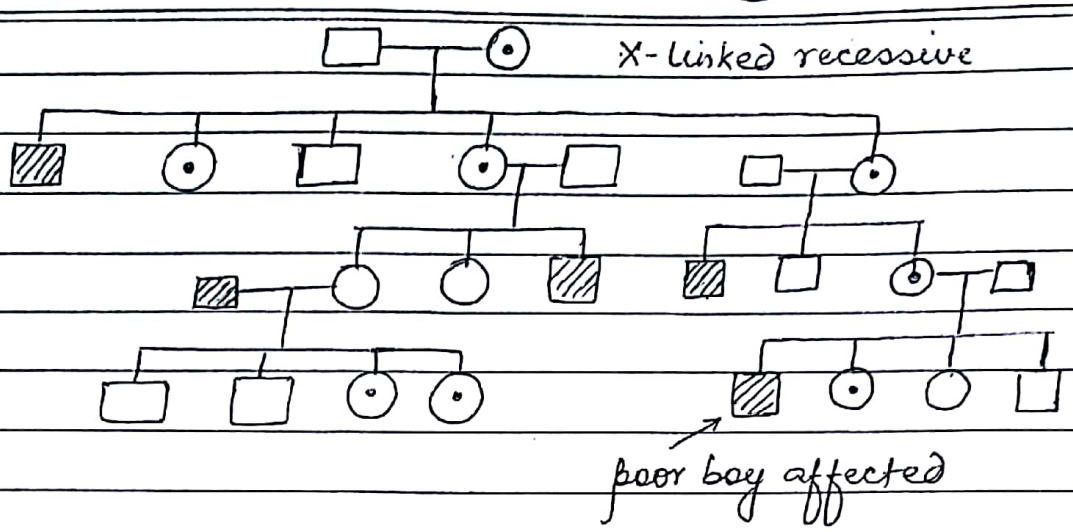

$X X^c$ $X Y$

XX XX^c $X^c Y$ XY

X-linked Recessive: (Poor ~~boys~~ boys)
- Duchenne muscular atrophy. (M/c hereditary Neuromuscular dz)
- Hemophilia A & B.
- G-6PD deficiency.
- Wiskott-Aldrich syndrome.
- Colour blindness.
- Lesch-Nyhan syndrome.
- Chronic granulomatous disease.

Duchenne muscular hypertrophy (DMH):
- Pseudohypertrophy of calf muscle bcoz of fat deposition.
- Proximal muscle weakness
- Gower's Sign → Not specific.

△ → CPK = 10,000 IU

Valley Sign ⇒ Hypertrophy of Supraspinatus
Atrophy of Infraspinatus
↳ Specific for boys who don't have calf hypertrophy.

↓
Also seen in DMH.

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Human Genome:
- 30,000 gene.

Largest gene – Dystrophin
↳ Skeletal muscle
↳ Heart – Cardiomyopathy.
↳ Brain → 1/3 cases MR.

Boys – Duchenne – die teens due to recurrent chest infection.

Duchenne → XLR, 1/3 denovo mutation.

Becker's dystrophy:
- Similar to duchenne
- Mild form & present late.
- X-linked recessive.

Wiskott-Aldrich:
- X-linked recessive
- Eczema
- Thrombocytopenia
- Immunodeficiency.

Chronic granulomatous dz –
- X-linked recessive
- Immunodeficiency.
- ~~Hexo~~ NADPH oxidase deficiency
- Dx → NBT dye test

Lesch-Nyhan Syndrome:
- X-linked ~~Sporadic~~ recessive
- Purine defect
- HGPRT ase deficiency.
- Hyperuricemia > 6.5 mg/dl
- Self-mutilation → nose, palate, fingers.

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Q. A mentally challanged child c dysphagia & opisthotonic spasms. He is also having choreoathetoid movements & self mutilation behaviour c +ve family history. Which of the following investigation is suggested?

A) Serum uric acid. → Leesh Nyhan Syndrome.
B) S. ALP
C) S. LDH
D) Lead level in blood

Q. A male child c Fanconi Syndrome c nephrocalcinosis have a variant of dent disease. All true except

A) Hypercalciuria → 24hr urine Ca^{2+} > 4mg/kg → Urolithiasis, Nephrocalcinosis
B) Proteinuria → LMW (β2 microglobinuria)
C) Similar presentation in father
D) Rickets

Fanconi Syndrome — Proximal tubular defect

Na^+	HCO_3^-	PO_4^-	AA, Glucose
60%	>85%	>88%	100%

Lowe Syndrome — X-linked recessive.
↳ Oculo-cerebro renal gene. (OCRL gene).

Congenital cataract, " glaucoma | Microcephaly, MR | Fanconi's Syndrome.

Fructose-1-phosphate, Glucose-1-phosphate] proximal tubular toxic.

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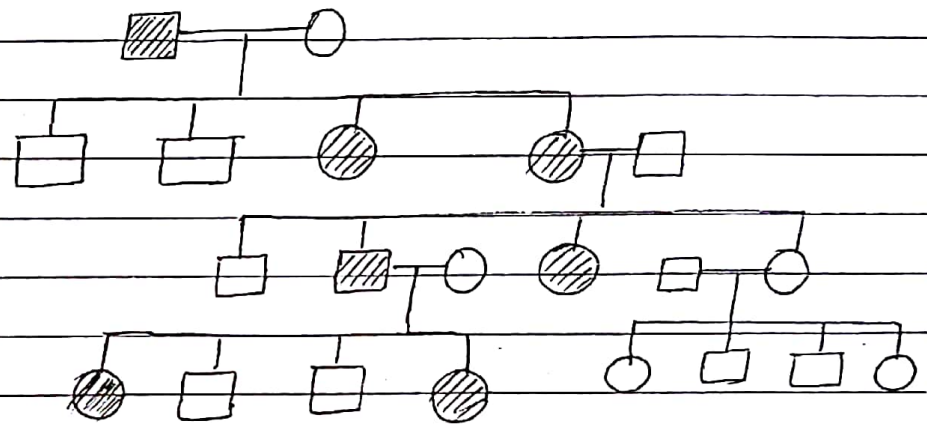
Fanconi's Syndrome:
- Genetic
  ↳ X-linked recessive – lowe
  XLR Dent
  AR
  ↳ Cystinosis.
- Metabolic – Galactosemia, HF-1
  Tyrosinemia (Rx – Nitisinone).
- Acquired – expired tetracycline



X-linked dominant father to all daughters – none sons

X-linked dominant: Males are more severely affected
- Familial hypophosphatemic rickets.
- Urea cycle defect due to OTC deficiency.
- Incontinentia pigmenti] Only seen in girls.
- Rett's syndrome.]

↓
bcoz boys die.
(lethal in male fetus).

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Rett's Syndrome:

- Pervasive ~~pervasive~~ developmental disorders.
  - ↳ Autism (Common in boy < 3yrs)
  - Asperger
  - Rett's

Q. Not seen in autistic ~~dis~~ disorder –

A) Social avoidance
B) Visual impairment.
C) Interest in one self
D) Introvert person.

Asperger's Syndrome: Common in boys
Very good IQ.

Rett's Syndrome – Common in girls
X-linked dominant.
Normal till 6-18 months
Decrease in head growth.
Microcephaly; MR
Hand movements
Repetative behaviour
$MeCP_2$ gene mutation

- ↳ Macrocephaly not seen.
- ↳ Abnormal dendritic morphology in cortical pyramidal cells (postmortem brain biopsy).
- ↳ Seizures.

Cause of death → Arrhythmias (Cardiac)
↓
Sudden death.

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Ornithine transcarbamoylase deficiency:

Orotic aciduria → URACIL
→ X-linked disorder.
- The mother of these child have also high Uracil level in urine.

Deletions:

Major deletion	Microdeletion
→ Cri-du-chat	→ Williams (7q23-) Syndrome
↳ Deletion of 5p chr.	→ Prader-willi Syndrome (15q11-13-)
- Cry like cat due to abn. larynx.	→ Di-george's Syndrome (22q11-)

Δ → by FISH.

Williams Syndrome:
Supravalvular aortic stenosis
c peripheral pulm. stenosis.
- Hypercalcemia
- Elf in facies.

Di-George's Syndrome:
- Hypoplasia of 3rd & 4th parapharyngeal pouch.
- Absent < Thymus
Parathyroid.

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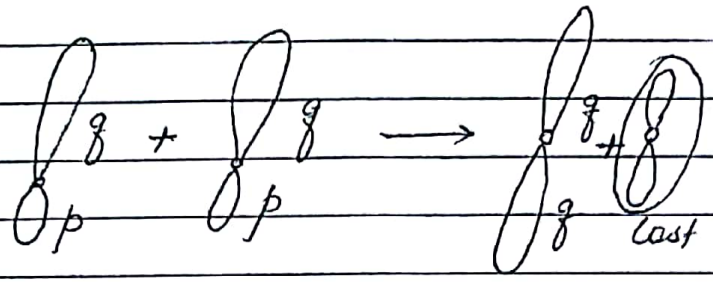
TRANSLOCATION:



- Unbalanced
- Robertsonian Translocation:
  Translocation b/w two Acrocentric chromosome.

M/c/c (Genetic) of MR = Trisomy 21 (Down's Syndrome)
↓
Extra chromosome is of females.

Trisomy 21 – In 95% cases: Maternal meiotic non-disjunction.
- 3-4% – Robertsonian translocation
- 1-2% Mosaic 47/46 ↓
  carrier of RT look normal but they can have abn children.

MOSAICISM:

A single zygote giving rise to different cells.

CHIMERA – different zygote giving rise to different cells
↳ Rare in humans.

- Mosaicism Seen in humans
- 1-2% are downs

Klinefelter Syndrome → 80% XXY; XY/XXY; XY/XXXY.

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Turner's Syndrome:

Cytogenetics

60% 45-XO

15% Mosaic XX/XO

10% Isochromosome Xq or Xp. — Mentally Retarded.

10% 46X deletion.

5% Mosaic XO/XY ⟶ Risk of Gonadoblastoma.

loss of one arm & duplication of other

↓

Isochromosome.

Mosaicism { Somatic — Not transmitted
Germline — Transmitted
↳ Blood DNA is normal.
eg: ~~Germline~~ Osteogenic imperfecta.

Q. Couple has two children c tuberous sclerosis. On detailed clinical & lab evaluation (including molecular studies) both parents are normal. ∴ one of the following explains the 2 affected children in this family —

A)

B)

C)

D) Germline mosaicism.

Maternal inheritance ⟶ Mitochondrial.

Mitochondrial inheritence:

- MERRF (Myoclonic epilepsy & red ragged fibres.
- Mitochondrial encephalopathy, stroke-like episodes, & lactic acidosis (MELAS)
- Leber hereditary optic neuropathy (LHON)

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- Leigh disease
- Kearns - Sayre Syndrome (KSS) (ophthalmoplegia)
- NARP (Neuropathy, Ataxia, Retinitis pigmentosa).
- Chronic progressive ophthalmoplegia.
- Pearson's Syndrome : Pancytopenia + Pancreatic insufficiency

Anticipation : Severely of genetic disorder ↑ in every successive generation.
More repeats — more problem.

eg: All Trinucleotide repeat disorders
- Fragile X — CGG repeats
- F. Ataxia — GAA "
- Myotonic dystrophy — CTG, CCTG
- Spinobulbar muscular dystrophy — CAG
- Huntington's — CAG
- Spinocerebellar ataxia — CAG/CTG.

Fragile X : genetic
- 2nd m/c cause of MR in boys.
- X-linked
- Large face
- Large ears
- Prominent jaw
- Large testis.
- CGG repeats > 1500 repeats

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Genomic imprinting:
Chr. 15q 11-13 deletion.
↳ Paternal inheritance cause Paderwilli Syndrom
↳ Maternal " " Angelmann "

- Differential expression of genetic appearance depending upon parents.

Prader-Willi Syndrome
- Severe neonatal hypotonia.
- Obesity
- Small hands & feets. (Rx - GH treatment)
- Unusual behaviour
- MR.
- High Ghrelin → So obese. (Rx - Anti Ghrelin)
(apetite hormone for fundus.)

Angelmann Syndrome:
- (N) at birth
- K/A happy puppets.
- Subsequently develop seizures
- MR
- Ataxia

Genomic imprinting → (1) Prader Willi Syndrome (70%)
↓
Almond shaped eyes.

(2) Angelman Syndrome (70%)
(3) Neonatal DM
(4) Beckwith Weidman
(5) Seckle
(6) Temple
(7) Wang
(8) Pseudohypoparathyroid Ib.

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Q. Father carrier of cystic fibrosis – AA^c

Mother – (N) → AA

↓

Yes there is a chance of cystic fibrosis in child

ACA^c

↓

Uniparental bisomy.

- In some cystic fibrosis
- In some sickle cells.

- 30% Prader Willi – Unimaternal disomy.
  5% Angelmann – Uniparental disomy.

PRADER WILLI :

70% – Paternal inheritance deletion.
30% – Unimaternal disomy
↳ Maternal silencing gene.

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Congenital Heart ds:

NADA's CRITERIA:

1 Major & two minor criteria at least.

Major:

1. Systolic murmur grade III or more ass. c thrill.
2. Diastolic murmur
3. Cyanosis (central)
4. CHF

Minor:

1. Systolic murmur < grade III
2. Abn S_2
3. Abn ECG
4. Abn X-Ray
5. Abn BP

& =

A_2 ------- P_2 Expire

A_2 ---------- P_2 Inspire

ASD – Wide & fixed S_2.

A_2 ---------- P_2 Expire

A_2 ---------- P_2 Inspire.

- Volume overload in right ventricle.

VSD – Wide & Variable S_2.

A_2 ---------- P_2

A_2 ---------- P_2

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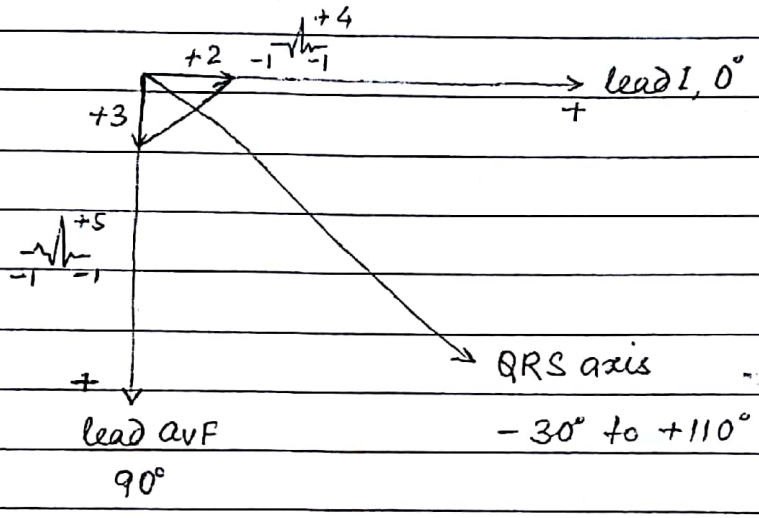
TOF – Single S_2 (A_2).

All newborn has RVH & RAD.
- Axis like adult > 1st month life.
- T-wave V_1 ; V_3R ; V_4R
  - 'up' first 48 hrs.



- Negative after 48 hrs
- Never be +ve < 6yrs
- > 6yrs → positive.

Prevalance of CHD:
- Prevalence = 0.8 – 1%
  Recurrance = 2–6%

M/C CHD = VSD (30–35%).
2nd M/C CHD = ASD (Secundum) – 6–8%
> PDA (6–8%)
> Co-arctation of Aorta (5–7%)
> TOF (5–7%)
> Pulm. Valve stenosis (5–7%)
> Aortic " " (4–7%)

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M/C Syndrome in CHD = Down's Syndrome.

Down Syndrome:
M/C → Complete AV septal defect (CAVSD) / AV canal defect / Endocardial cushion defect
Ostium primum ASD.
(37%) — M/C/C of death.
- VSD (31%)
- ASD (15%) → Secundum
- Partial AV septal defect (PAVSD) — 6%
- TOF → 5%
- PDA → 4%
- Miscellaneous → 2%

Turner's → half to One third bicuspid aortic valve; 20% coarctation.
Noonan's → Turner's phenotype.
AD; XX = XY
Valvular pulm. stenosis.
HOCM; ASD.
Rubella → PDA; Peripheral pulm. stenosis; VSD.
↓
Rubella
Alagille Syndrome
William's "

Maternal lithium → Ebstein's anomaly.

Maternal mumps → Endocardial fibroelastosis / LV obstruction newborn.
Maternal penicillamine → Cutis Laxa.

Maternal SLE << Maternal Sjogren Syndrome
- New born complete heart block.
pacemaker insertion (anti ~~Sm~~ Ro).

Maternal warfarin:
Chondrodysplasia punctata.

Maternal thalidomide: Phocomelia.

Foetal circulation:
1 Umbilical Vein (left)
2 umbilical artery.

Ductus venosus →

PO_2 in umbilical vein = 30-35 mm of Hg
PO_2 of IVC in fetus = 28-30 mm of Hg.

As soon as child takes his 1st ≠ breath
- Umbilical artery constricts.
↓
U. vein closes
↓
Ductus venosus closes.

Ductus arteriosus:
- Physiological ↓ in 10-15 hrs.
- Anatomical → 10-21 days.

Foramen ovale:
Functionally closes by 3 months.
Anatomically 10-15% open.

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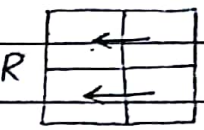
CHD

Acynotic Cynolic.

Acynolic : L→R Shunt

ASD, VSD, PDA.



Pulm. blood flow ↑ (Plelhora)

↳ lung vascularity is good.

C/F : Failure to thrive

Recurrent pneumonia

Feeding diaphoresis

CHF in 6-10 wks of life.

Suck-rest-suck cycle

↑ sweating. (d/t sympathetic stimulation).

Tachycardia] in every CHD.

Cardiomegaly]

Cyanotic

PBF

↑ ↓

Plelhora	Oligemia	
- Persistant Truncus arteriosus (Cyanosis + Recurrent pneumonia)	Massive Cardiomegaly.	→ Tricuspid Atresia - LVH Ⓝ Heart Single S_2
- d TGA + VSD	• Ebstein's (Box/Ballon heart)	FALLOT'S • TOF • DORV + PS • d TGA + VSD + PS
- TAPVC		• Single Ventricle + PS.

physiology.

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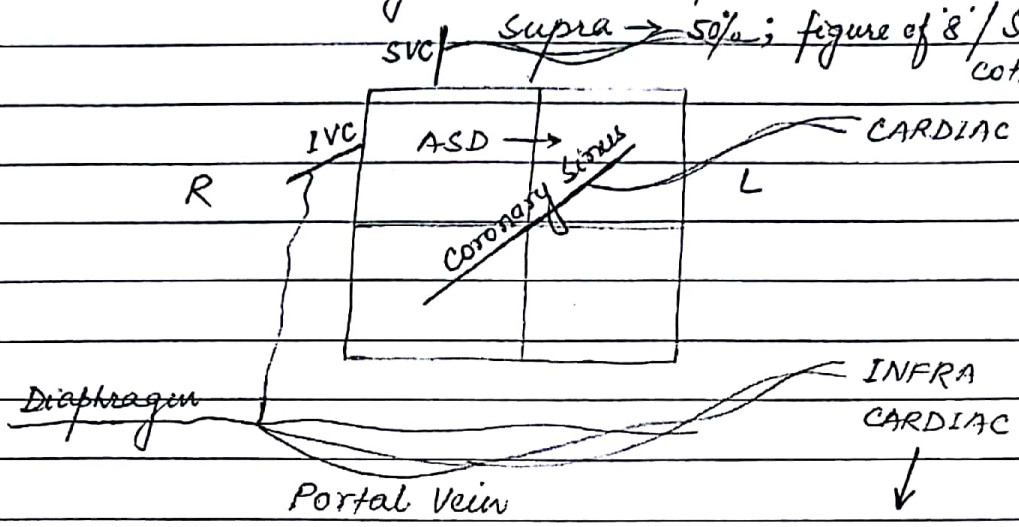
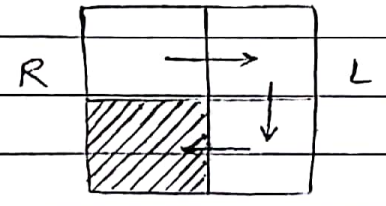
TAPVC (Totally anomalous pulm. venous connection):



More severe TAPVC.

- Cyanosis at birth.
- Obstructive pulm. venous hypertension.
- Ground glass: Kerley B lines.
- Worsen by PGE1 infusion.
- Only pediatric cardiac Sx emergency.

Tricuspid Atresia:



- Cyanosis + ↓ PBF
+
LVH
LAD

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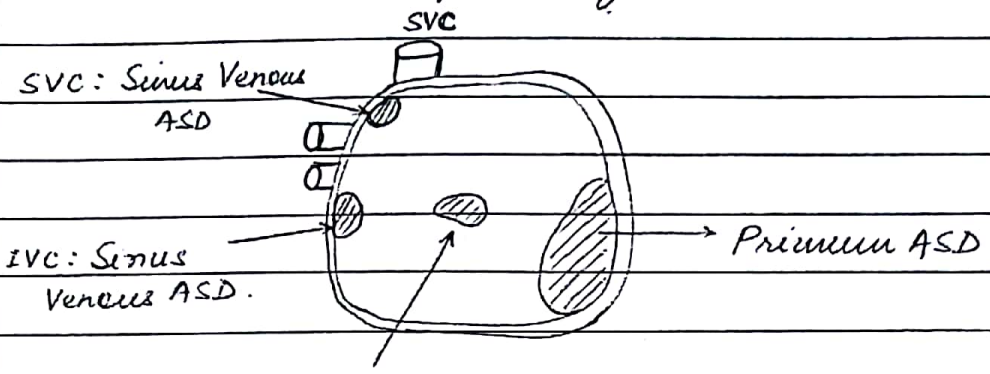
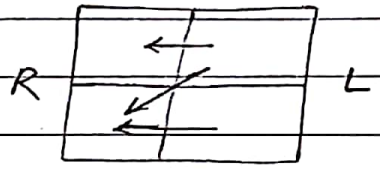
ASD (Atrial Septal defect):



M/C syndrome ass. = Down's Syndrome.

Endocardial cushion defect.



- Very large L-R shunt.
- PBF ↑
- Pulm. plethora

ASD Syndrome:

- HOLT ORAM : • Familial, AD,
  • ASD Secundum; VSD; 1 degree heart block;
  • AF + Bone defect (Absent Radius).
  • Distally placed thumb / Rudimentary thumb / Triphalangeal thumb.
  • A/k/A Hand-heart Syndrome.
  • TBX5 mutation → Pleiotrophy.
  ↳ Common transcriptional factor for hand & heart.

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Absent Radius associations:
- ECG (Holt-oram)
- Platelet (Thrombocytopenia ; AR).
- Bone marrow biopsy (Fanconi's anemia)
  ↓
  Congenital aplastic anemia
- Rarely Karyotyping (Edward Syndrome)

ASD Syndromes:
- Down's Syndrome
- Holt oram
- Lutembacher → ASD + Mitral Stenosis
- Ellis Van Creveld → ASD + polydactyly.

ASD Secundum:
- Child → asymptomatic, wide & fixed S_2.
- ECG → RAD (Right axis deviation).
- In adult life → Complications
  ↓
  - RV failure
  - Arrhythmias ; AF → CVA
  - Reversal : R → L (Eisenmenger Syndrome)

Natural history:
If < 3 mm → close itself
> 8 mm → Unlikely to close ; Require Sx.

Indication of Sx in ASD Secundum:
- All symptomatic.
- $Q_p / Q_s > 2$; even if no symptoms.
  ↳ Pulm blood flow / Systemic blood flow.

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Q. Least chances of Infective endocarditis is seen c/c ?

A) Small VSD → M/c/c of IE

~~B)~~ Small ASD secundum → Rare

C) Mild AS

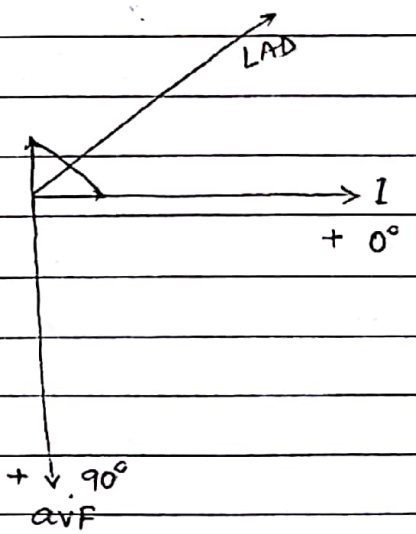
D) Mild AR.

ASD Secundum doesn't require any prophylaxis t/t before going to Sx.

ASD primum + Mitral Regurgitation:

- Wide & fixed S_2 +
  S_1 = = = = S_2 apex → axilla & back.
- 6 – 10 wks presents c/c CHF.
- Conduction defects ; ECG → LAD.
- Common in Down's Syndrome baby.
  ↓
  LAD d/t endocardial Cushion defect.

Down's
AVSD



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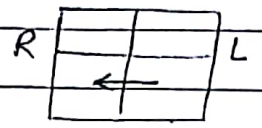
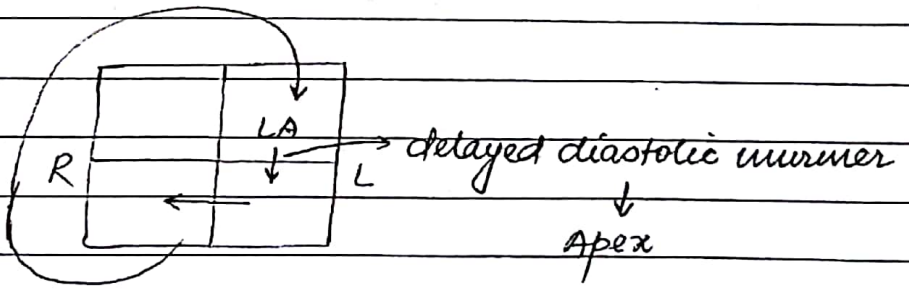
VSD (Ventricular Septal defect):

70% → Perimembranous

Small VSD	Medium VSD	Large VSD
• Root of aorta < 1/3		• > 1/2
• < 3 mm		
• Called Maladie de Roger's defect.	← CHF at 6-10 wks of life →	
• Loud murmur (S_1) (Pansystolic) ↓ lower left Sternal border.		• No murmur/ • Ejection systolic murmur. (S_1 & S_2).
• Asymptomatic		

In medium to large VSD, Left Atrium enlarges first.
In Small to medium VSD, left Ventricular hypertrophy. due to blood overload.



Natural course of VSD closure:

- 80% perimembranous
- 50% muscular VSD's close.
- By 4 yrs.

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Indication of Sx in VSD:

- Failure of Medical therapy.
  - ↳ Digoxin
  - ↳ Diuretics
  - ↳ Dilators
  - ↳ ACE i (Remodelling of heart).
- Qp/Qs > 2 ; even if no symptoms.
- Swiss cheese VSD. (Multiple ; Apex)
- Supracristal (outflow).

Large L → R shunt
↓
Pulm. blood flow ↑
↓
Irreversible changes pulm. microvascular.
[Pulm. HTN]
↓
RV pressure ↑ ; RVH
↓
Reversal ; R → L (Eisenmenger Syndrome).
↓↓
Sx is C/I !

Differential clubbing : Toes > fingers
↓
PDA + Reversal.
(also differential Cyanosis).

Down's Syndrome baby c Endocardial cushion defect undergoes Eisenmenger Syndrome.

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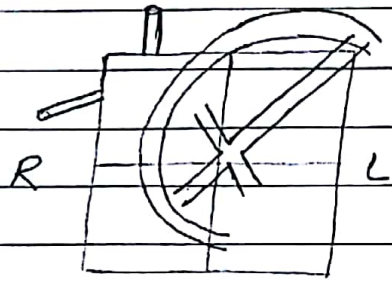
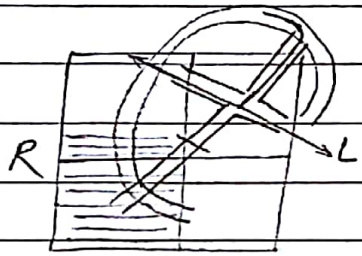
Ductal dependent lesion:



- HLHS ⇒ Rx - (PGE1) infusion. → S/E - Apnoea.
- Critical AS
- Preductal coarctation
- Interrupted aorta
  ↳ Shock.

I) Systemic blood flow dependence.

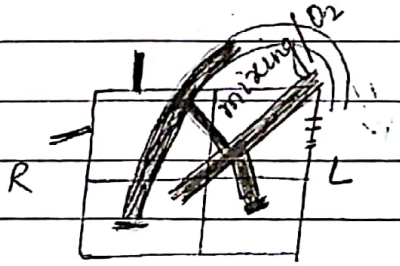
II) Pulm. blood flow dependence.
- Tricuspid Atresia
- Pulm. Atresia

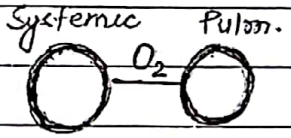


↓
Central cyanosis.

In emergency condn → PGE_1 infusion.

III) d-TGA





R_x - PGE_1 infusion

d-TGA → dependent for mixing.

Truncus Arteriosus is Ductal independent.

Emergency Sx in TGA → Rashkind's Ballon atrial septostomy.

Definitive Sx → Jatene's Arterial Switch.
↳ Best time: ē in first 2 weeks of life.

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M/c Cyanotic Heart ds:

Overall — TGA

In Infants — d TGA (<1yr)

> 1yr — TOF

Hyperoxia test in cyanotic newborns —

- 10L O_2 to cyanotic newborn
- pO_2 > 150 mm Hg ⟶ excludes heart ds.

① 50% d-TGA + VSD ⟶ Mild

② d-TGA + intact septa (complete d-TGA) — Birth.

③ d-TGA + patent foramen ovale ⇒ At birth.

① < ③ < ②

TOF (Tetralogy of Fallot):

M/c cynotic heart ds beyond infancy.

① Narrowing of the pulm. valve (Infundibular).
② RV hypertrophy.
③ Overriding of aorta over VSD.
④ VSD — opening b/w left & right ventricles. (perimembranous).

Pink child become a blue TOF.

- Cyanosis; clubbing.
- Polycythemia → R/F of Renal vein thrombosis (RVT)
- Hematuria, [redacted], Flank mass, Anemia.
- Infant — Cyanotic/ tet/ hyperpneic spells → Older Squats
- Complications:

① < 2yrs = Thrombosis

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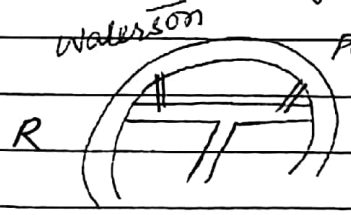
(11) > 2yrs = Brain Abscess.
↓
in the territory of middle meningeal artery
(Parieto-Temporal)

Infants → Cyanotic/tet/hyperpneic spells — older squat
(Rx: Knee chest position; i.v. Ketamine; Phenylephrine)
- R → L Shunt ↑ — Murmur disappears.
- PO_2 falls (Rx: O_2; i.v. $NaHCO_3$).
- Respiratory centre +++ (Rx: S/c Morphine)
- Dynamic Pulm. stenosis (Rx: i.v. Propanolol)
- Crying (Bcoz of ↑ venous return).

In TOF — loud Holosystolic murmur at the left 2nd-3rd ICS due to pulm. stenosis.

Palliative Shunt
- Modified Blalock-Taussig → Subclavian to Pulm. artery (opposite to Aortic Arch)
- Waterston
- Potts (Descending Aorta to left pulm artery)



25% TOF ⇒ has Right aortic arch.
↓
M/C c/c 50% of Truncus arteriosus

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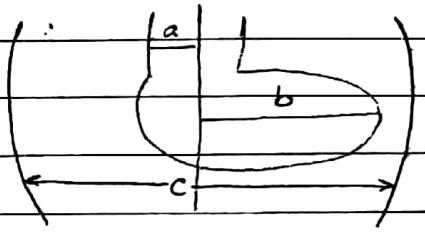
Pentalogy = +ASD

Trilogy = Overriding of aorta absent; VSD absent; ASD present.

- CHF never seen in TOF
- Cardiomegaly never seen in TOF.

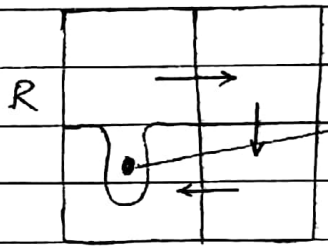
↓

Cardiothoracic ratio > 0.6 infant
> 0.55 in older infant.



"Boot shaped heart"
"Coer en Sabat"

Ebstein's anomaly:



- Cyanosis ↓ PBF
- Maternal lithium
- Atrialisation of RV.
  - Pressure = RA
  - Ventricular = ECG
  - → disconcordence b/w pressure & ECG.
- Intracardial ECG helpful in Δ.
- Systolic; diastolic murmurs.
  Quadruple rhythm
- Box/Ballon; Massive heart.
- SVT; WPW Syndrome.

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Q A neonate present c recurrent abdominal pain, restlessness, irritability & diaphoresis on feeding. Cardiac auscultation reveals a non-specific murmur. He is believed to be at risk of MI. The most likely Δ is —

A) VSD

B) ASD

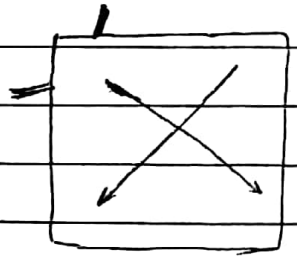
C) TOF

D) Anomalous origin of the left coronary artery = ALCAPA

- Q wave in Lead I ; aVL
- LAD branch absent.
- Ischemic LV → Antero lateral MI.
  On Aortography → Rt coronary artery fills.
- Lateral MI in infant.

Rx : Bypass Sx.

l-TGA → Corrected TGA.
Normal Heart.
Atrio-Ventricular discordance.



[RA connected to LV
  LA " " RV

No problem to baby.

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KAWASAKI'S DS:

- M/c acquired heart ds in US, japan, & Chandigarh.
- Fever > 5 days & ↓.
- Development of a limp.
- Erythmalous macular exanthem over body
- Ocular conjunctivitis.
- Dry & cracked lips.
- Red throat & cervical lymphadenopathy.
- Grade II/IV vibratory systolic ejection murmur at lower left sternal border.
- Predominant Neutrophils c ↑ platelet.

- M/c medium sized vasculitis.
- 20-25% causes develop aneurysm in future.

Q. Which of the following vasculitis not occur in adults?

A) Kawasaki ds (85% are < 5yrs)
B) Susac's Syndrome — Seen in adult females.
C) Giant cell arteritis
D) HSP

Peeling of palm & soles is classically seen in 2nd - 3rd wk of life.
Rare in ~~8~~ before 3 months of life.
DOC : Iv Ig.

Δ: Fever > 5 days & any 4 of these 5

① Changes in extremities (eg: Erythma, edema, desquamation).
② B/L conjunctivitis (not ass. c exudates)
③ Polymorphous rash (not vesicular)
④ Cervical lymphadenopathy.

⑤ Changes in lips & Oral cavity (eg: Pharyngeal erythma; dry/fissured or swollen lips, Strawberry tongue).

Non-classical feature of Kawasaki:
Arthralgia; Arthritis;
Thrombocytosis; urethritis;
aseptic meningitis (Irritable)

M/c/c of death in Kawasaki —
Overall — Coronary artery aneurysm
Acute phase — Myocarditis.

Rx: IvIg 2g/kg in acute phase reduces risk 4-6%
Aspirin 100mg/kg/day x 2weeks.

If Resistant to IvIg: 10-15% cases
↳ Add steroid (Methyl prednisolone).
↳ Repeat IvIg
↳ TNF blockers → Infliximab; Eternecept.
↳ IL-1 inhibitor → ANAKINRA

Recurrance Rate → 1-2% cases.

Mx of aneurysm:

Small → 50% resolve over 1-2yrs.
Aspirin 3-5mg/kg/day.

Medium to large → Add Warfarin
Sx (may be bypass)

HSP (Henoch-Schonlein Purpura):
M/c vasculitis (small vessel) Overall.
M/c leucocytoclastic vasculitis.
- Palpable non-thrombocytopenic purpura.
- IgA, deposition of vessels in dermis.

Pathogenesis: Abberrant Galactosylation
Mesangio-proliferative disorder.

Small vessels → Skin
Arthritis
Arthralgia
GIT – mesentric ischemia
Kidneys – HSP Nephritis (40-50%).
↓
84% develop in 4 wks
91% " " 6wks
97% " " 6months.

- Microscopic hematuria
- Proteinuria
- 1-2% RPGN (over days to week).
↳ On Biopsy crescent seen
↳ ∴ Crecentric GN
↓
proliferation of parietal epithelial cells.

Rx: i.v. Methylprednisolone.

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Rheumatic Fever:

- M/c acquired heart ds in India / Developing country.
- Due to Group A β-hemolytic streptococci strains M-type 1, 3, 5, 6 & 18.
- Most frequent b/w 5-15 yrs.
- Latent period 3 weeks.
- Autoimmune: Molecular mimicry

Affects < Myocardium
Muscle.

Modified Jones (2015):

- Era of Echocardiography → Subclinical AR/MR.
- A/c to Risk area.

Low Risk — defined as having an ARF incidence <2 per 100000 school-aged children (usually 5-14 yrs old) per year or an all age prevalance of RHD of ≤1 per 1000 population per year (Class IIa; level of evidence C).

Criteria = 2 major or 1 major + 2 minor
+ Essential criteria.

Recurrance = 2 major or 1 major + 2 minor
(OR)
3 minor

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Major Criteria:

Low risk populations:

- Carditis (Clinical &/or Subclinical)
- Arthritis (Polyarthritis only).
- Chorea
- Erythema marginatum
- Subcutaneous nodules.

Moderate to high-risk populations:

- Carditis (Clinical &/or Subclinical)
- Arthritis
  - Monoarthritis or polyarthritis
  - Polyarthralgia ⊕
- Chorea
- Erythma marginatum.
- Subcutaneous nodules.

Minor Criteria:

Low risk populations:

- Polyarthralgia
- Fever (≥ 38.5°C)
- ESR ≥ 60 mm in the first hour &/or CRP ≥ 3.0 mg/dl.
- Prolonged PR interval, after accounting for age variability.

Moderate to High risk populations:

- Monoarthralgia
- Fever ≥ 38.5°C
- ESR ≥ 30 mm in first hour or CRP > 3 mg/dl.
- Prolonged PR interval – Also in MR & MS.

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℞:
- Aspirin 100mg/kg/day X 12 wks.
- Prednisolone X 12 wks.
  - 2mg/kg X 3wks → taper
  - Severe carditis or CHF.
- Crystalline Penicilline G X 10 days
- IE prophylaxis.

Mitral valve → M/c involved in RF
↳ Recurrance cause Mitral stenosis.
↓
Primary prevention
↓
In India 2° prevention
↳ Penicillin G
Benzathine (i.m.)
- 600,000 IU for children wt ≤ 60lb
1.2 million IU for children wt > 60lb
every 4 wks.

Or, Penicillin V (Oral)
↳ 250 mg twice a day

or, Sulfadiazine or Sulfisoxazole (oral)
↳ 0.5g once a day for pt. wt ≤ 60 lb
1.0g once a day for pt. wt > 60 lb.

For pt. allergic to penicillin & Sulfa group.
↓
Microlides are given orally.

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CATEGORY	DURATION
- Rheumatic fever c out Carditis	5yr or until 21 yrs of age whichever is longer.
- RF c carditis but c out residual heart ds (No valvular ds)	10yr or until 21 yr of age, whichever is longer.
- RF c carditis & Residual heart ds (persistent Valvular ds).	10yrs or until 40 yrs of age whichever is longer. Sometimes lifetime prophylaxis.

Q
- Blood pressure = 86/600 mm Hg
- 4 yr, unconsiousness.
- HR = 180/min
- CFT = 4sec

Δ = ? → Compensated Shock.

Rx = 20 ml/kg 0.9% Nacl.

SHOCK = BP < 10th centile for age & Sex.

Criteria for Shock:

Criteria for Hypotension by age.

Age	Systolic BP
Term neonates (0 to 28 days)	< 60 mm Hg
Infants (1–12 month)	< 70 mm Hg
Children (1–10 yrs)	< 70 + (age in yrs X2)
Children > 10 yrs	< 90 mm Hg

Compensated Shock: Rx – 20 ml/kg 0.9% Nacl
Repeat upto 60 ml/kg
↳ CVP line (next step).
• Low – fluid given.

• Normal → Cold epinephrin
=/ Warm: Nor-epinephrin.

Hypertension BP > 95th centile
- Essential – 10%; increasing
- Secondary HTN
  ↳ Renal parenchymal – Reflux Nephropathy
    VUR
  ↳ Renovascular
    • Major → RAS; RVT
    Minor → HUS
  ↳ Cardiac – post ductal coarctation (Turner)
  ↳ Endocrinal – • Hyperthyroid
    • Cushing
    • Pheochromocytoma.
    • CAH 11 beta / 17 alpha hydroxylase deficiency

End organ damage in HTN:
• Fundus
• Echocardiography concentric LVH.
• Any adolescent BP > 130/80
  In children > 10 yrs → 120/80 → HTN
• Urine protein.

Rx: HTN
• Life style modification.
• Pharmacological therapy – end organ; Symptoms; Severe > 99th centile.
  ↓
  ACEi / ARBs.
  ACEi C/I if GFR < 30 → causes hyperkalemia
  ↓
  Rx → Amlodipine (CCB's)

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Hypertensive Emergency:

- LVF ⇒ S_3; Gallop; Basal crepts.
- Seizures.

Rx:

Best: i.v. Nicardipine infusion

Rx: Sodium Nitroprusside i.v. /
Esmolol /
Labetalol i.v.

Linezolid, Nitroprusside & Amphotericin B should be covered.

Q. A 12yr old boy c seizure.
BP = 200/140.
Femoral pulses not palpable.
Δ^z ?

A) Takayasu aortoarteritis (Rx - Prednisolone).
B) Grand Mal Seizures.
C) Fibromuscular dysplasia (FMD)
D) Renal parenchymal defect.

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Tachyarrhythmias

- Based on QRS

Wide > 0.09 sec (VT/VF)

- Pulseless
- Arrest.

Rx – Defibrillation 0.5–1 J/kg

- Stable

Rx – Lignocaine
Amiodarone.

Narrow

- Reentrant SVT
- HR > 220 infant; > 180 older
- P-wave absent; inverted

Vagal maneuvers

Stable

Rx – Adenosine (fastly given c saline flush)
↓
as close to the heart as possible.

CHF; Shock

Rx – Synchronized Cardioversion

Respiratory

Cough & cold caused by Rhinovirus.

IMNCI G/L

Age = 2m – 12 months = RR ~> 50 or more.
Age 12m – 60m = RR ~> 40 or more.

• Chest is indrawing = Pneumonia.
or Fast breathing.

Rx :- Give oral Amoxicillin for 5day.

Any general danger sign ⇒ severe Pneumonia.
or Stridor in calm child or very severe dz

Rx
→ Give 1st dose of an appropriate A/b.
→ Refer to hospital.

Q. 18 m old child wighing 11.5 kg comes to PHC.
with c Resp. difficulty. O/E Lethargic RR = 46/min.
no Chest Retraction = Nex Step.
Ans (B) Prescribe oral Antibiotic & Refer to higher centre.

→ Signs (< 2 months)
- convulsions or
- Fast breathing (≥60)
- Severe Chest Indrawing.
- 10 or more skin pustules.
or a big boil or
If axillary temp 37.5°C
or above or less than 35.5°C
- OR Lethargic/Unconscious.
- less than (N) limit.

Next step
Serious = Give i.v.
Bacterial ⇒ Gentamicin.
Inj (B)
↓
Refer to higher centre.

- Umbilicus red or draining → local (= Give oral
- Pus/Pus discharg from: Back, Buttock
ear or <10 skin pustules.

- Causes of Pneumonia Age wise in India & world
- Neonates : Grp. B Streptococcus, E. coli.
<3 weeks.
- 3wk - 3m = Respiratory Syncytial virus (RSV)
S. Pneumonia, H. Influenza.
- 4m - 4yr = RSV, S. pneumonae, H. Influenza.
>5yr = M. pneumonae., S. pneumonae.
- H. Influenza → Vaccination (dlt) → ↓ in Incidence.

Viral pneumonia
- Prodrome.
- Diffuse, B/L
- Not lobar pneumonia
- Interstitial infiltrate

T/t : ① DOC for RSV = Ribavarin.
② Influenza = Amantadine, Rimantadine
H1N1 = Influenza A → Neuraminidase Inhibitor.
↳ oral Oseltamivir.
↳ Inhaled zanamivir.

Pandemics :-
Phase 1 - 3 Animals.
Phase 4 - May 27, 2009 (Mexico)
↳ Hum-human transmission at community
Phase - 5 - 29/5/09 USA.
Human - Human transmission in one zone

Phase 6 : June 11, 2009, US & India.

→ Human - human transmission in two zones

H1N1 virus :-
- SS RNA.
- Belongs to orthomyxoviridae.
- Size = 80 - 200 nm.

Types :- A, B, C.

Surface Ag = H (Hemagglutinin)
N (Neuraminidase)

As the reassortment occurs in swine flu.
= Swine flu.

D/t Antigenic shift & drift - No vaccines made

Symptoms = Flue like.

Complication of H1N1 → in High risk groups.
⇒ Pneumonia (viral)
⇒ Bacterial superinfection.
⇒ ARDS like features.

High Risk groups :- Comorbidity.
→ They were associated with same ~~mortality~~
→ Pregnancy
→ Nephrotic syndrome, Chronic illness, post transplant
- <1 yr., >65 yrs.

Indications of oseltamivir
① Give it to all suspected cases of H1N1
② Give it to all confirmed case.
Dx is by throat swab & Nasopharyngeal.
Swab is send for Real time PCR.
⊗ H1N1 influenza.
③ Give it to all household contacts, occupational (Post exposure in doctors)-

Adults
Dose = 75 mg Cap BD - 5d.
Prophylactic dose = 75 mg OD → 7-10 days.

Page No.: 106 VVI
Date:

Zanamavir.

T/t dose = Two 5mg Inhalations (10mg total) twice/day x 5 days.

Prophylaxis dose = two x 5 mg Inhal® - OD.

Q:- H_1N_1 10 wks pregnant.
→ Give Oseltamivir [Not teratogenic
↳ only in cats]
→ TE®

Q:- Pt on Rifampicin induces hepatic CYP450.
gets H_1N_1, sy®
→ What happen to dose of oseltamivir
Ans = Remain same ~~but~~ Renal exc®
Q. Dose in hemodialysis
= oseltamivir dose is reduced.

Pneumonia

- St. pneumonia = Causes severe lobar pneumonia.
- Incub/ Pd. = 1-3 days
℞ → Ceftriaxone DOC.

→ Resistant Variety = DOC ⇒ Vanco + ceftriaxone

Vaccines :- > 2 yr. Children = PPV - 23.
< 2 yr = PCV - 7/11 = conjugated.
6, 10, 14 weeks, ƒ/B booster at 15-18 months.

GAVI :- Global Alliance for Vaccines & Immun®
- founded by bill gates & Melinda Gates
- Free vaccines in poor country

Staph. Aureus

- Max Mortality (10-30 %)

Ch: — Air filled cavity (Pneumatocele)
. also seen in dkt.
1) Klebsiella.
2) Kerosene oil poisoning

→ Pneumatocele can Rupture & develop pneumothorax

• S. aureus = M/c/c of empyema in children.
(Plus in Pleural cavity)

T/t = ICDT.

DOC for S. aureus = cloxacillin.
Vancomycin for MRSA.

• H. influenza = Usually part of S. sepsis.
- can have arthritis, meningitis.
Rx = ampicillin & chloramphenicol.
20-40 %. are resistant.
Rx. (Ceftriaxone) → DOC.

⊛ Atypical pneumonia
→ Rare - <4 yrs = ~~common~~. >5 yrs. !
- Symptoms - dry cough.
- Interstitial pneumonia
- Organism = Mycoplasma Chlamydia.
Rx :- Macrolides

15 month

Motor → Walks alone, crawls up stairs

Adaptive → Makes tower of 3 cubes, makes a line with crayon, inserts raisins in bottle

Language - Jargon; follows simple commands, may name a familiar object (eg. ball) responds to his/her name.

Social → Indicates some desires or needs by pointing, hugs parents.

18 month

Motor - Runs stiffly, sits on small chair, walks up stair with 1 hand held, explores drawers & waste baskets.

Adaptive → Makes tower of 4 cubes, imitates scribbling, imitates vertical strokes, dumps raisin from bottle.

Language - 10 words (average), names pictures, identifies 1 or more parts of body.

Social → feeds self, seeks help when in trouble, may complain when wet or soiled, Kissed parent with pucker.

24 month

Motor → Runs well, walks up & down stairs, 1 step at a time, opens door, climbs on

furniture, jumps.
Adaptive → Makes tower of 7 cubes (6 at 2 month) scribbles in circular pattern, imitates H₂ stroke, folds paper once imitatively
Language → Puts 3 words together (subject, verb, object)
Social → Handles spoon well, often tell about immediate experiences, helps to undress, listens to stories when shown pictures

30 months
Motor → Goes up stairs alternating feet
Adaptive → Makes tower of 9 cubes, makes VH & horizontal strokes, but generally will not join them to make cross, imitates circular stroke, forming closed figure
Language → Refers to self by pronoun 'I' Knows full name
Social → Helps put things away, pretends in play

36 months
Motor → Rides tricycle, stands momentarily on 1 foot
Adaptive → Makes tower of 10 cubes, imitates construction of bridge of 3 cubes

copies circle, ~~imitates cross~~

Language - Knows age & sex, counts 3 objects correctly, repeats 3 numbers or a sentence of 6 syllables, most of speech intelligible to strangers.

Social → Plays simple games (in parallel with other children) helps in dressing (unbuttons clothing & puts on shoes) washes hands.

48 months

Motor → Hops on 1 foot, throws ball overhead, uses scissors to cut out pictures, climbs well

Adaptive → Copies bridges from ~~parental~~ model, imitates construction of gate of 5 cubes, copies cross & square, draws man with 2-4 parts besides head, identifies longer of 2 lines.

Language - Counts 4 pennies accurately, tells story

Social → Play with several children, with beginning of social interaction & role-playing, goes to toilet alone

60 monthy.

Motor - Skips

Adaptive → Draws △ from copy, names heavier of 2 weights

Language - Names 4 colours, repeats sentence of 10 syllables counts 10 pennies correctly

Social → Dresses & undresses, asks questions about meaning of words, engages in domestic role playing.

Date / /
Page

Fever & Stridor Inspiratory Sound in upper airway.

Croup / LTB

- 75% parainfluenza
→ Prodrome → Stridor
Barking Cough.
Subglottic narrowing
k/A : Stippel sign on x-ray.
↳ Doc = for mild : dexa : 0.6mg/kg.
mod & Severe
↳ Nebulisation : Racemic epinephrine.
↳ Westley Croup Score, Criteria to dx it.

Epiglottitis

In world – S. pyogenes.
S. pneumoniae
S. aureus
In India – H. Influenza.
dat X-Ray (Neck) → Thumb Sign.
Rx → Airway [Emergency tracheostomy]
Ceftriazone + Sulbactam (ESBL)

BRONCHIOLITIS:

- Inflammatory obstruction of Smaller airway.

M/C organism – RSV (50%)
↳ Respiratory Syncytial Virus.

R/F :
- Males, Topfed.
① - Preterm ; Chronic lung disease.
② - L → R shunt.
- Smoking mothers.

Airway resistance, $R = \frac{1}{r^4}$ → air trapping

Prodrome → Wheeze / ~~and~~ Ronchi.

X-Ray chest shows Hyper inflation.

Rx : Antibiotics has no role.
Humidified O_2.
For 2 High Risk group – Nebulised Ribavarin
Palivizumab

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Long term — Persistent wheeze infancy
↓
Reactive airway disease.

ASTHMA

Classification a/c to severity.

	Step	Symptoms	Night symptoms	Peak expiratory flow rate.
I.	Intermittent.	< 1 time a week; asymptomatic & (N) PEFR b/w attacks	≤ 2 times a month	≥ 80% predicted; Variability < 20%.
II.	Mild persistent.	> 1 time a week but < 1 time a day	> 2 times a month	≥ 80% predicted; Variability 20-30%
III.	Moderate persistent.	Daily use β-agonists; daily attacks affect activity.	> 1 times a month	> 60% & < 80% predicted; Variability > 30%
IV	Severe persistent.	Continuous limited frequent activity.	Frequent.	≤ 60% predicted; Variability > 30%

For intermittent → SOS β_2-agonist
Mild persistent → +++ inhaled beclomethasone, Fluticasone, Budesonide.
Moderate persistent → +++ Salmeterol / Sustained release theophylline.
Severe persistent → ++ Oral low dose, long term, alternate day prednisolone.

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2 yrs → Acute severe asthma
↓
Oxygen; Nebulise salbutamol
↓
Nebulised ipratropium bromide
↓
i.v. hydrocortisone
↓
s/c Terbutaline
↓
Terbutaline infusion
↓
50% $MgSO_4$ → aminophylline.

Foreign body:
CxR → Persistent inflation
Ball valve inflation.
Rx → Bronchoscopy & remove foreign body.

Recurrent pneumonia:
2 episodes of radiographic pneumonia in 1 year
(OR)
3 episodes in any time frame.

Persistent pneumonia:
Persistence of symptoms & radiographic abnormalities for more than 1 month.

Recurrent pneumonia:
Cause → · L → R shunt
· GERD d/t aspiration.
Immunodeficiency.

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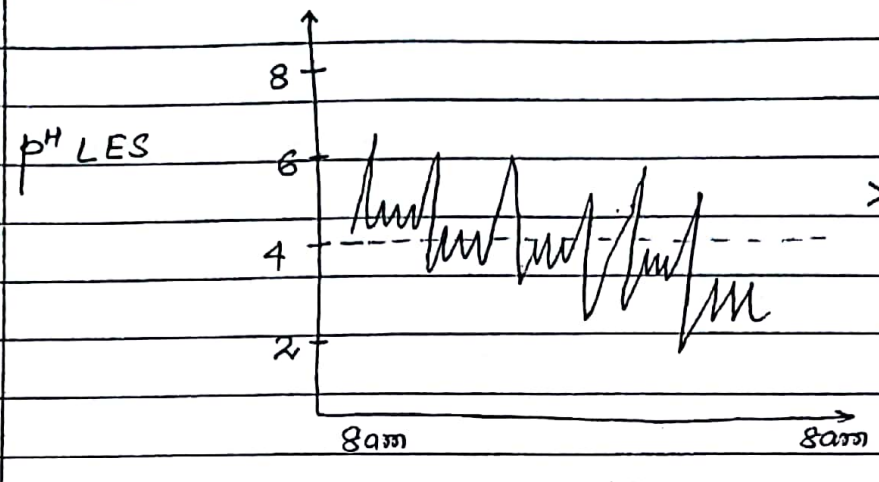

24 hr pH LES

M/c/c of Recurrent pneumonia in U.S. = Cystic fibrosis.

Cystic fibrosis

- Incidence : 1 in 2500 in UK

Gene → CFTR gene (7q 31.2 locus)

↓

CAMP regulated chloride channel.

- In CF more Sodium goes to lumen & to mucosa than chloride.
- Autosomal Recessive disorder.
- Respiratory : Pneumonia (>5yrs of life).

M/c organism ass. c cystic fibrosis — Pseudomonas aeruginosa.
(Mucoid > Non Mucoid)

Rx : Inhaled antibiotics
- Tobramycin
- Aztreonam
- Colistin
- Ciproflox

- Amikacin
- Levofloxacin

Respiratory : Pneumonia (>5yrs)
- <5yrs — S. Aureus ; H. influenzae.
- 5-18yrs — S. Aureus ; Pseudomonas
- >18yrs — Pseudomonas ; S. aureus.
  Acromonas ; Achromobacter ;
  Burkholderia capacia
  ↳ specific ; fatal
  ↳ Rx Mild — Colrimoxazole
  ↳ Severe — Meropenem + Colrimoxazole/Doxycycline.

- Exocrine Pancreas — Stealorrhoea (<5yrs)
  foul - bulky stools.
  ↓
  Rx — Stealorrhoea in CF
  Lipase 1000 IU/kg supplement.

- Endocrine → 25% DM after 35 yrs.
- GIT → Meconium ileum.
  Aldolescents — Distal intestinal obstruction Syndrome.

48hrs old baby has not passed meconium.
Ix → Lower GI contrast study
- Diagnostic
- Therapeutic in meconium ileum.

- GIT : Diffuse pain abdomen
  Colonic mucosal thickening.
  Intussusceptions.
- Nasal polyps ; Azoospermia

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- Absent Vas : 1% infertile men ; common in CF males.

Q. Which glands are not obstructed in CF ?

A) Cervix → Infertile
B) Pancreas → Insufficiency.
C) Sweat glands

M/c mutation in CF = ΔF508 mutation (Class II mutation) → Causes "Trafficking Defect"
(Seen in 70% caucasions)
↓ (25-30% Indian)
M/c lethal genetic disorder in caucasions.

CFTR gene has 1800 mutations.

How many Nucleotide deleted in CF ?
3 → TTT = Phenylalanine.

℞ : For Trafficking Defect
↓
DOC → LUMACAFTOR (Trafficking corrector drug)
+
IVACAFTOR (Potentiator drug).
↓
opens CFTR 'Cl' channel.

Lumacaftor — induce hepatic cyp 450
↓
∴ New Corrector → TEZACAFTOR
↓
doesn't induce Cyp 450.

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Q. Which of the following is a calcineurin inhibitor?

A) Tacrolimus
B) Sirolimus → m-tor inhibitor
C) Everolimus → m-tor inhibitor
D) Cyclosporine

Diagnostic Criteria of CF:

① Sweat Chloride > 60 meq/L on 2 occasions
[N < 40 meq/L]

OR, ② Two known CFTR mutations.

OR, Best – ③ Diagnostic nasal electrode potential difference.

Newborn screening for CF

IRT test → Immuno reactive Trypsinogen Assay test.
(Sensitive test)

Q. Male, 10 month boy, Down Syndrome & Recurrent pneumonia

Filling defect → In Ba-swallow
↳ in middle of esophagus.
↳ Abbarrent right subclavian artery vascular ring
↓
Dysphagia Lusoria.

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Approach to a pt. c recurrent / persistent pneumonia:

↓

History, physical exam, CxR
— Rule out TB

↓

Difficulty in feeding; choking during feeds.	Infection in other parts of body	Associated Malabsorption. Pseudomonas in airway	No clues
↓	↓	↓	
GER studies, Esophageal PH monitoring, Barium Swallow, Direct laryngoscopy.	Immunoglobulin CD4, CD8 NBT, HIV test.	Sweat Cl⁻ test Mutation Studies.	

Isotonic fluids:
- 0.9% Nacl — 154 meq/L Na & Cl.
- Ringer lactate = Plasma.
  - 130 meq/L Na
  - 109 meq/L Cl
  - 28 meq/L Lactate⁻
  - 4 meq/L K
  - 3 meq/L Ca

Maintenance fluid:

Need:
- Insensible water loss.
- Energy
- To prevent catabolism.

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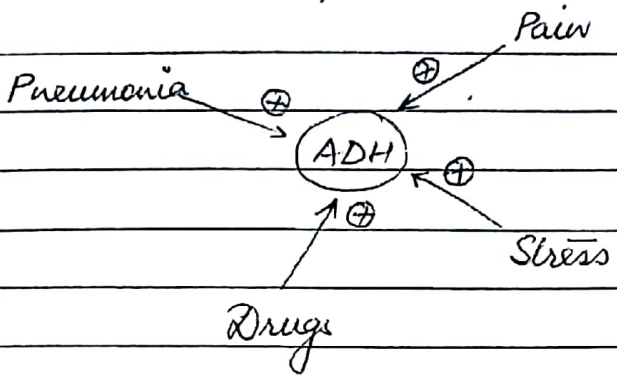
Type?
- Adult: 5% DNS
- Children 5% Dextrose + N/2 or NS.



Amount?
A/C to Holiday & Segar
< 10 kg – 100 ml/kg/day.
10 kg – 1000 ml/day. (40 ml/hr).
11–20 kg → 1000 ml (for 10 kg) + 50 ml/kg additional kg above 10 kg.
> 20 kg → 1000 ml + 500 ml + 20 ml/kg additional above 20 kg.

Patau's Syndrome (Trisomy 13):
- Cleft lip & palate.
- Polydactyly
- Hypotelorism (Eye sepreted wide).
- Abortus looks like Cyclops
  ↳ Holoprosencephaly. (Single eye)
  ↳ Fused frontal lobes + lateral ventricles.
  ↳ Aplasia cutis (problem of cleavage of skin).

~~- Rocker bottom~~

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Edward Syndrome (Trisomy 18)

- Rocker bottom feet
- Overlapping of fingers.
- 2nd M/C trisomy.
- Maternal age
- GIT anomaly are common
  - ↳ Atresia gut
  - ↳ Exomphalos
  - ↳ Malabsorption.

TTNB (Transient Tachypnea of New Born): Wet lungs.

R/F:

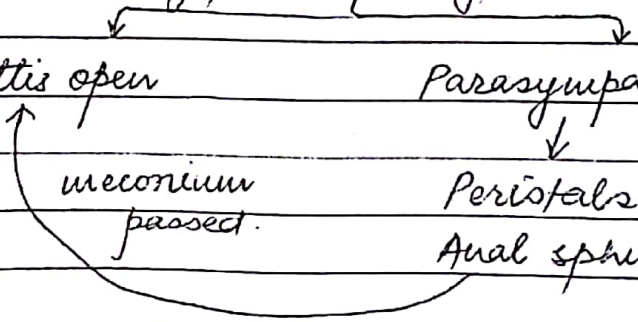
- Term ; Cesarean Section
- Macrosomia
- Precipitous labour.
- Maternal Sedation

C×R – Prominent horizontal fissure.

- It is a benign, self limiting condⁿ ⊆ resolves in 48–72 hrs.
  - FiO_2 requirement < 0.4
  - Never require mechanical ventilation.

MSL (Meconium Stained Liquor):

- Marker of perinatal hypoxia (common in Post term)



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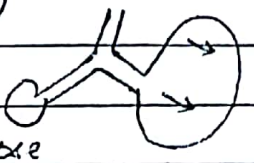
Complication of Meconium:

Physical:  Ball valve.

collapse

air leaks 20-30%.

Chemical: Irritant.

Biological < impairs surfactant function (Surfactant given)
Good culture media (Given Antibiotic)

℞: Meconium stained liquor

↓

Born

Vigrous < Tone.
HR > 100/min
Resp^{r} effort.

NO | YES

limp or, HR < 100 or, Apnea. → PPV × 100% O_2.

Transfer to mother

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GROWTH

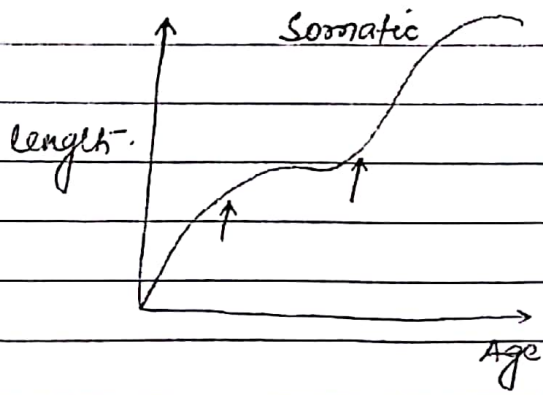
1-4 months : Weight gain 30gm/day
5-8 month : wt. gain @ 20gm/day
9-12 month : wt. gain @ 10gm/day.

Weight multiples:

Wt. × 2 = 5 months
× 3 = 1 yr
× 4 = 2yrs
× 5 = 3yrs
× 6 = 5yrs
× 7 = 7yrs
× 10 = 10yrs.

Length:

At birth – 50cm] Growth Velocity → +25 (first year)
1 yr – 75 cm]
2yr – 90 cm → +15 → second year.
4 ½ yr – 100cm
Add 6cm / year till puberty.



Q. In school going children; the avg height velocity is
→ 5-8 cm

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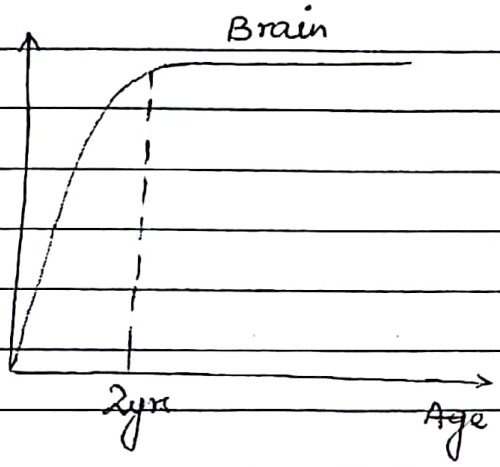
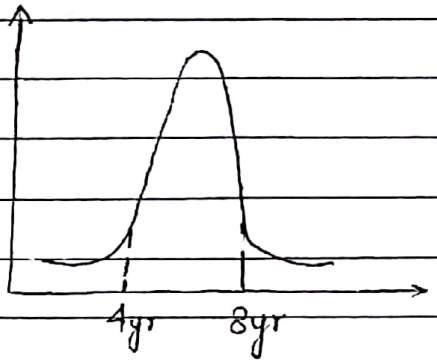
Head circumference:

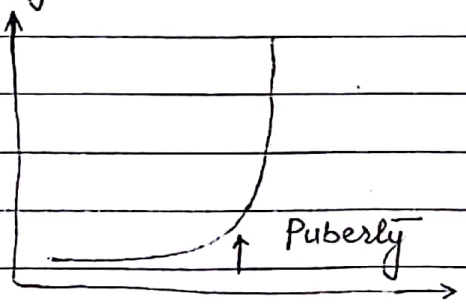
- 35cm at birth
- 3 months → 40cm
- 12months → 45cm
- 2yrs → 48cm (90% of brain grows)
- 12yrs → 52cm.



Lymphoid growth:



Gonad growth:



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Adolescent — 10 to 19 yrs.

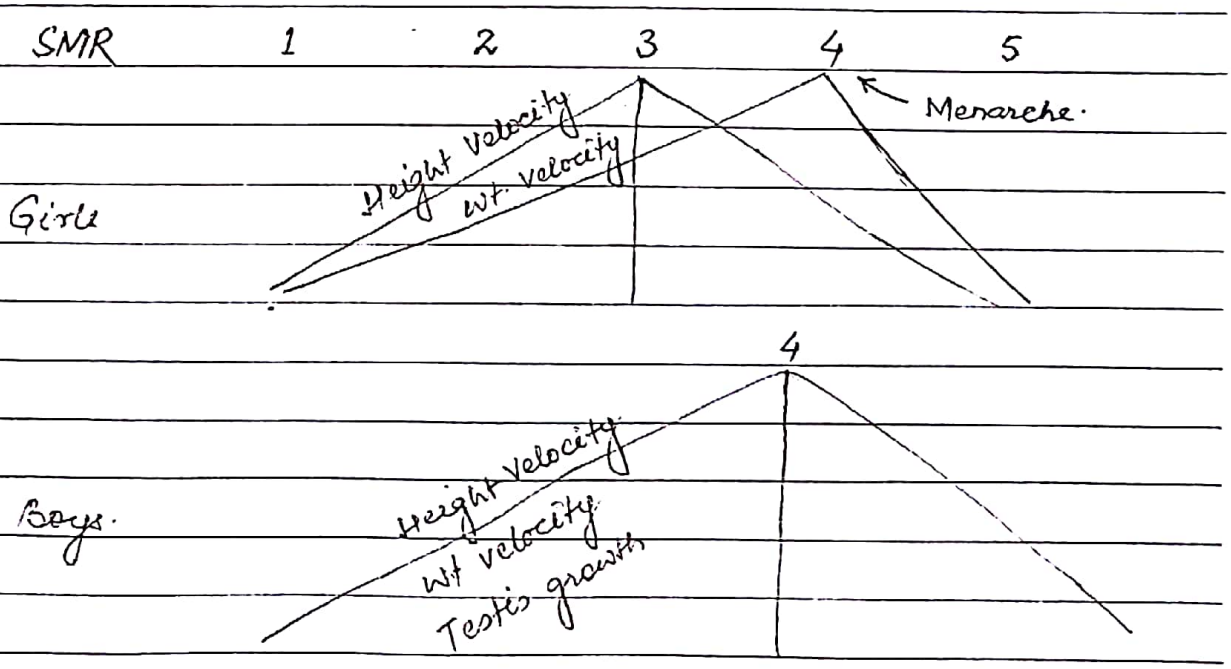
Tanner's Sexual Maturity Rating :
SMR Stage I - V
Stage I — No character
Stage V — Completely developed.

Menarche → SMR-IV

Sequence of puberty :
Girls :
Growth Spurt → Thelarche, Pubarche, Menarche.

Boys :
→ Testis - Penis - Pubic hair, Axillary hair.

Girls : BP (Breast — Pubic hair)
Boys : GP (Genitalia - Pubic hair).



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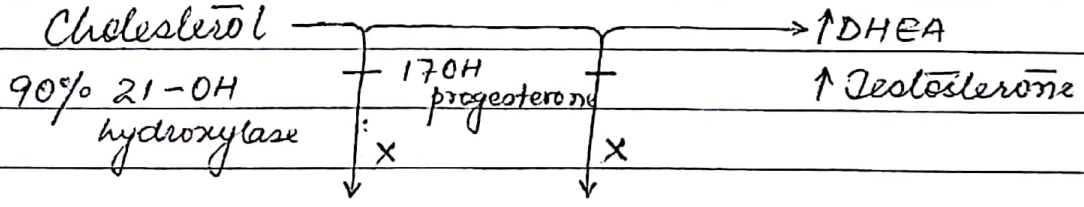
Most rapid ↑ in height → Phase 3 in girls & Phase 4 in boys.

Puberty – comes by pulsatile release of GnRH.

Precocious Puberty:
- Gonadotropin dependent / Central
- Females – Idiopathic.
- Males – Organic
  (Hypothalamic Hamartoma, Craniopharyngiomas, Hydrocephalus, TBM).

CAH (Congenital Adrenal hyperplasia):
M/c/c of female hermaphrodite.
Karyotype – XX
But looks like Male.



Cortisol
glucose ↓

Aldosterone
Na^+ ↓
Salt-Wasting Crisis (70%)
↑ K^+
Shock

– Cause of precocious puberty in boys.

Genitalia → Ambiguous
Labia is pigmented d/t ACTH.
{ Virilization
Hypertrophy of clitoris

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- Penoscrotal hypospadias
  + Empty Scrotum.

Due to deficiency of 21-OH hydroxylase lead to accumulation of 17-OH progesterone.

Q Level of 17-OH progesterone in CAH
A) < 150
B) 150 - 300
C) 300 - 500
~~D)~~ > 600 (> 3500)

CAH associated c Premature epiphyseal closure.
↓
Short Stature.

Rx : CAH
- Supplement of Hydrocortisone & Fludrocortisone
  ↓ Glucocorticoids: 15-20 mg/m²/day, Thrice daily.
  ↓ Mineralocorticoids: 0.15 mg/day.

- Girls require Sx
  - Clitoroplasty
  - Reconstruction Sx.
  - Never get married.

Cholesterol → DHEA → Testesterone.
Cholesterol → 11-deoxycortisol → Cortisol
Cholesterol → DOCA : potent mineralocorticoid → Aldosterone
11β-OH deficiency → HTN, Hypokalemic metabolic alkalosis.

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Q. 5yrs old boy has precocious puberty. BP = 130/80
Estimation of ♀ help> diagnosis?

A) 17-OH progesterone
B) 11-deoxy cortisol
C) Aldosterone
D) DOCA

Deficiency of 17 α-Hydroxylase → ↑ Aldosterone
↓
HTN, Hypokalemia, metabolic alkalosis.

→ X Cortisol
↓ glucose.

→ X Testosterone
↓
Male look like ♀.

(Mineralocorticoid) M T (Testosterone).

2 ↑

↑ 7

↑ ↑

3β Hydroxysteroid dehydrogenase deficiency (3β HSD def.)
↓
Causes ambiguous genitalia in Both sexes.

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Q. Mother; previous child CAH

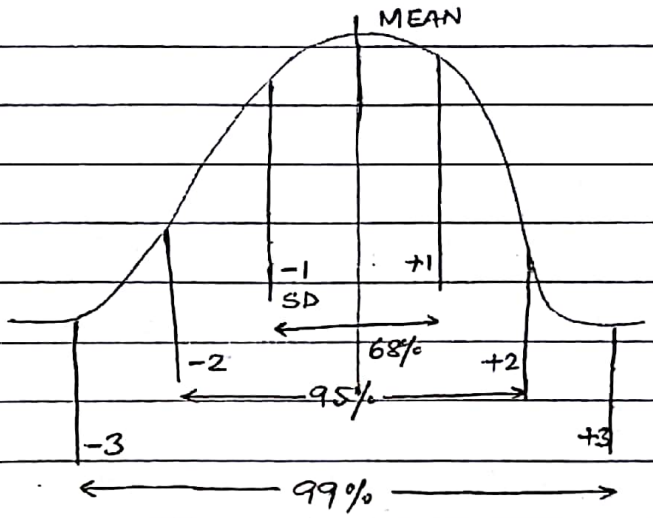
Next pregnancy — To prevent female virilization of foetus

↓

Dexamethasone
(20 mcg/kg prepregnancy wt.)

↓

Inhibits ACTH.



Short height > -2 SD below mean or < 3rd percentile

Mid parental height = Adult predicted height

Boys = average parents height + 6.5 cms
Girls = " " - 6.5 cms.

Causes of Short Stature:

- Physiological
  - Constitutional (M/C)
  - Familial
- Pathological
  - Malnutrition
  - Chronic illness
  - Celiac disease (Present at 6-18 months as mother introduces wheat now)
  - Genetic → Turner

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℞ : Celiac ds
- Restrict Wheat, Rye & Barley for lifetime.
- Gluten-free diet.
- Also avoid oats.

↓

- Endocrinal → Hypothyroid
Cushing
CAH
GH deficiency.

GH deficiency :
- Birth weight & length normal.
- lag at 1-2 yrs of life.
- Bone age delayed.
- Doll face
- Micropenis } Panhypopituitrism
- Short stature

Micropenis (↓LH, ↓FSH)
↓ACTH → Hypoglycemia

- Hoarseness of voice.

INV: GH level ⇒ IGF, BP3

GH stimulation test – Best
↳ Basal level GH
↳ Stimulated level – 2 stimuli
(clonidine ; Insulin ; L-arginine).

℞ – Recombinant GH (US, FDA)

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Indication: rGH

- GH deficiency
- Turner Syndrome
- Prader villi
- Chronic kidney ds.
- SGA height -2.25 SD below mean

When to stop GH?

- Height reaches 50th centile
- Epiphysis fuses.
- Pseudotumor cerebri
- Slipped capital femoral epiphysis.

Disproportionate short stature:

US/LS ↑ → Achondroplasia, Rickets, Hypothyroidism.

US/LS ↓ → TB spine, Mucopolysaccharoidosis IV (Morquio's ds)

Physiological Short Stature:

	CONSTITUTIONAL (M/C)	FAMILIAL
Birth length	(N)	IUGR
Lag	6-12 months	
Growth velocity	(N)	Less
Final Height	(N)/Sub(N)	Less
Puberty	Delayed	(N) ♀
Bone age	Delayed.	(N) ♀

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Developmental milestones:

Neck holding → @ 3month.

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Gross motor milestones:

3 months — Head holding; Neck holding.
5 months — Sitting c support.
4-6 months — Prone to supine, supine to prone (Roll in bed).
8 months — Sitting c out support.
9 months — Crawling
10 " — Creeping; stand c support
12 " — Standing c out support; walking c out support.
2yr — Walk up stairs c two feet at each step.
3yr — Upstairs c one foot at each step, rides tricycle.
4yr — Hops on one foot
5yr — Skips on two foot

Fine motor:

12wks → Moro's reflex disappears
Grasp reflex disappears
4 month — Goes for objects
5 months — Bidextrous grasp.
6-7 " — Transfer object, palmar grasp.
9 " — Pincer grasp; mature neat.
13 months — Casting.
15 " — Self feed c a spoon
18 " — Self feed c a cup.
24 " — turns pages of a book one at time.

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Social Milestone:

2 months — Social smile
3 " — Recognizes mother.
6-7 " — Smiles at mirror image.
9 months — Waves bye-bye.
6 months — Stranger anxiety.
2yrs — Dry by day
3yrs — Dry by night
Dress / undress himself (Supervision)
↓
c out supervision — 5yrs.

18 month — Seperation anxiety / Clinginess / Reprochment.

Language Milestones:

1 month → Head turns to sound
3 months → Cooing
6 months → Babbles; Monosyllables (ma, ba)
9 months → Bisyllables (mama; ba-ba)
1 yr → 2 words c meaning
18 months → Vocabulary of ten words.
2yrs → Simple sentences c 2 words; Phrase.
3yrs → tells age & sex; uses pronouns, handedness; identify colours.
4yrs → tells story
5yrs → Knows colour

15 months: 4-5 words
2yrs → 50-100 words

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When can a child understand death =

< 3yrs — No idea
3-9 yrs — Idea
>9yrs — Entity ; Irreversible ; himself

Object permanence / Constancy — 9 months.
Cross a busy road — 10-12yrs.
Tie shoe lace — 5yrs.
Bladder control — 85% by 5yrs.
Nocturnal enuresis — >5 yrs.
↳ more than 2/week x 3months.

Q. 14 yrs old child c Nocturnal enuresis
Rx — • Behavioural — +ve reinforcement.*
• Alarm therapy (Best)
↳ 85% relapse free rates.
• Drugs → Desmopressin

Encopresis : > 4yrs
Cause → Chronic constipation.

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CNS

MICROCEPHALY:

→ HC > -3SD below mean

Causes:	GENETIC	ACQUIRED
	- Trisomy 13, 18, 21	- Baby: HIE; Hypoglycemia, PKU, Meningitis, encephalitis.
	- Cri-du-chat 5p deletion	- Mother → TORCH, Hyperphenylalanine > 6mg/dl Alcohol, DM, Radiation.

MACROCEPHALY

HC > 2SD above mean.

Q Neurodegenerative disorders:

GREY MATTER:

- Normal at birth.
- Regression milestones.
  - ↳ Disappear as they come orderly.
- Deaf, blind, seizures
- Anemia, Hepatosplenomegaly.
- Cherry red spot macula.

Microcephaly	Macrocephaly
- GM1 gangliosidosis.	- GM2 gangliosidosis (Tay Sach's ds)
- Gaucher's ds (β-gluco)	- Sandhoff
- Niemann Pick's ds	

Huge spleen

VLCFA = Very long chain fatty acid.

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Tay Sach's ds –
- AR
- D/t deficiency of β-hexose aminidase A.
- 1/25 askenazi jew are carrier.
- 6 months exaggerated startle reflex.
- Cherry red spot macule
- Organomegaly not seen.

Organomegaly + Tay Sachs = Sandhoff
↓
Def. of β-hexose aminidase A & B.

WHITE MATTER DISORDERS:
- All the tracts are white matter.
- Frequent fall, incoordination
- Upper motor neuron sign.

Microcephaly / (N)
- Krabbe's
- XLR adrenoleukodystrophy.
↓
degeneration starts from parieto-occipetal area.
Δ → ↑ VLCFA levels
Rx → Early bone marrow Tx.
Lorenzo oil
- Meta chromatic leukodystrophy

Macrocephaly
- Canavan ds.
- Alexander ds.
→ MRI → Diffuse white matter thickening.
→ MRI → Degeneration starts from frontal periventricular area.

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Hydrocephalus:

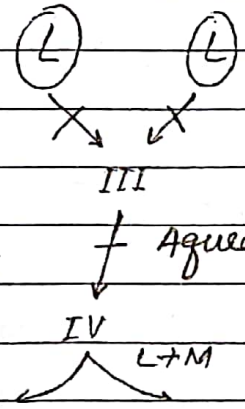
- Enlarged ventricles c or c out ↑ in CT.

CSF production:

- Choroid plexus (75%) → Lateral, III & IV ventricles.
- Extrachoroidal (25%) → Capillary endothelium in brain parenchyma.

Rate of CSF production → 20 ml/hr.
CSF volume in infants = 50 ml
adults = 150 ml.

CSF flow:



Obstructive / non-communicating.

↓

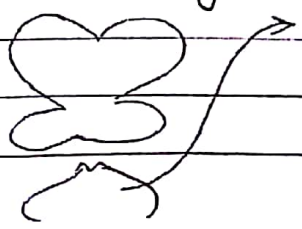
① Aqueductal stenosis (M/c)
② Aqueductal gliosis
③ Arnold chiari Malformation

↓

Downward displacement & hypoplasia in cerebellum leading to obliteration of cisterna magna.

Arnold-Chiari Type I – Adolescent/Adult
Type II – Newborn/c
lumbosacral myelomeningocele.

④ Dandy-Walker Syndrome:



large cyst in posterior fossa communicating c 4th ventricle.
– child has cerebellar hypoplasia

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⑤ Vein of Galen malformation:
- M/C arterio venous malformation in brain
- Sinus Venosus ASD.
- Obstructs aqueduct.
- Midline mass c dilated lateral ventricle.

Non-obstructive / communicating:
- Basal exudates
  - ↳ TB
  - ↳ Cryptococcal meningitis.

IOC for congenital Hydrocephalous — MRI.

Rx: Drugs → Furosemide
Acetazolamide.
VP shunts → anastomose ~~bags~~ ventricles to peritoneum.
- ↳ Gross hydrocephalus
- ↳ Thinned parenchyma.
- ↳ Complication — Blockage
  Infection (Coagulase -ve staph).

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Febrile Seizures:

- M/c seizure during childhood.
- Between 6month – 5yrs.

R/F for recurrance:

Major:

Age < 1yr
Duration of fever < 24 hr.
Fever 38–39°C (100.4 – 102.2°F)

Minor:

Family H/o Febrile seizures.
Family H/o epilepsy.
Complex febrile seizures.
Daycare
Male gender.
Lower serum Na^+ at time of presentation.

Having no risk factors carries a recurrance risk of approx 12%

If 1 R/F → 25–50%
2 " → 50–59%
≥3 " → 73–100%.

R/F	Risk for Subsequent Epilepsy
Simple febrile Seizure	1%
Recurrent " "	4%
Complex " " (>15 min duration or recurrent c in 24 hr)	6%
Fever <1 before febrile seizure	11%
Family H/o epilepsy	18%
Neurodevelopmental abnormality (Mental Retardation).	33%

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Complex febrile seizure complication:
- Prolonged febrile seizures.
MRI → Mesial temporal sclerosis
Temporal lobe + Hippocampal lobe.

Epilepsy in children:
Partial:
- Simple
- Complex - aura; automatisms.
℞ — Oxcarbamazepine &
Carbamazepine.

Ring Enhancing lesions.

Neurocysticercosis	Tuberculoma.
- Solitary	- Large > 20mm
- SCOLEX	- Multiple
℞ — Albendazole (DOC)	- Irregular margins
↓	- Perilesional edema
Before giving Albendazole 3–5 days of steroids given.	- Midline shift.

Generalised epilepsy:
- Tonic
- Clonic
- GTCS [aura – GTCS – post ictal phase]
↓
Drowsy, unconscious, frothing, tongue bite uprolling, incontinence.
- Atonic
- Myotonic

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℞ – Sodium Valproate.

↓

In < 2yrs → It is hepatotoxic.

Absence Seizure:

- Blank stare < 30 sec.
- No aura / post ictal phase.
- Hyperventilation provokes.

℞ : Ethosuximide (DOC)
Valproate

Atypical Absence seizure:

- Myoclonic component

℞ – Valproate.

JME (Juvenile Myoclonic epilepsy):

- 12–18 yrs.
- Myoclonic jerks morning.
- Drops things.
- Gene 6p gene
- Family History.
- GTCs seizures 90%
- 1/3 Absence seizures.

EEG of JME → Generalised 4–6 Hz spike + photic stimulation.

℞ – Valproate (lifelong) → Excellent.

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Infantile spasms / Salaam / West

- Flexor contractions of Head, trunk & extremities.
- 4-8 months
- EEG → HYPSARRYTHMIA
  ↳ Generalised chaotic · high volume slow wave.
- Idiopathic / Cryptogenic → Good.
- Secondary → HIE, Structural malformation, Down's Syndrome, Tuberous sclerosis.

℞ :— ACTH → inhibit CRH. (DOC).
Vigabatrin → In Tuberous Sclerosis

STATUS EPILEPTICUS:

- Convulsion > 30 min

(OR)

Continuous b/w no regain of consciousness.

Vulnerable to hypoxia — Hippocampus, amygdala. Thalamus, subcortical areas.

℞ : — i.v. Lorazepam (DOC) — longer t½
0.05 mg/kg

- Midazolam + Phenytoin (20 mg/kg)
  Repeat 10 mg/kg — 10 mg/kg
  Phenobarbitone 20 mg/kg — 10 mg/kg
  i.v. Valproate 20-30 mg/kg
  Midazolam infusion 2-20 mcgf/kg/min.
  i.v. ~~Levetiracetam~~ Levetiracetam 20 mg/kg.
  GA propofol ; Thiopentone.

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M/c/c of Status epilepticus → Febrile Seizures.

↓

℞ → per rectal diazepam / Buccal midazolam

Prevention of Febrile Seizures:

- No need
- Risk of Recurrance / concerned parents

↓

Intermittent prophylaxis Oral CLOBAZAM / DIAZEPAM

↳ New BZD

for 48-72 hrs of fever.

MENINGITIS:

Cause:

	India	World
< 2 months	Klebsiella E. coli	Gr. B / D Streptococci E. coli
2 months - 3yrs	H. influenzae type B	S. pneumoniae.
> 3yrs	S. pneumoniae	S. pneumoniae Nisseria

Acute Bacterial meningitis:

- 95% cases occurs b/w 1 month to 5yrs.
- Defect of complement system C_5-C_8 & properdin system
  - meningococcal infection.
- T-lymphocyte defects (eg - AIDS/Chemotherapy)
  - Listeria monocytogenes / cryptococcus.

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- congenital/acquired defects across mucocutaneous barrier → Pneumococci d/t cribriform plate.
- Lumbosacral meningomyelocele & dermal sinus – staph. & enteric bacteria.
- Penetrating CNS trauma/CSF shunt infection – coagulase -ve staph.

Recurrent meningitis in CSF leak pt. M/c d/t pneumococcus.

Autosplenectomy (sickle cell dysfunction/asplenia)
→ Pneumococcal infection
H. influenzae
Nisseria
} capsulated.

Splenectomy vaccination time – 2wks before.

℞: Ceftriaxone (DOC)
DOC in Resistant pneumococci = Vancomycin + Ceftriaxone.

M/c Neurological sequel of meningitis:
- SNHL: via aqueduct cochlear.
- Can we prevent SNHL?
  - Dexamethasone
  - 0.15 mg/kg
  - 30-60 mins before antibiotics.

Post exposure prophylaxis to contacts & doctors –
H. influenzae & Nesseria → i.m. Single dose ceftriaxone.
- Rifampicin X 2 days
Doctors → Fluoroquinolones.

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Q. 3 yrs old diagnosed to have HiB meningitis. Ix done before discharge → BERA.
(Brainstem evoked response audiometry)

ENCEPHALITIS:
M/c/c = Enteroviruses (80% cases).
M/c sporadic = HSV-1
M/c aseptic meningitis in unimmunized children — mumps.

Q. Child c fever & coma
Focal seizures.
CSF: Hemorrhagic
CT: Temporal hypodense ; MRI → Hyperintense
— HSV-1 infection.

Localised temporal spike → HSV encephalitis.
↓
DOC: i.v. Acyclovir.

Mortality rate of untreated herpes = 70%.

AFP (Acute Flaccid paralysis):
- Acute onset < 6 weeks
- < 15 yrs
- Rule out pseudoparalysis
↳ Septic arthritis
Osteomyelitis
Scurvy ; early syphilis
Hypokalemia → Hypotonia.

- Asymmetrical AFP:
• Paralytic polio.

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• Traumatic neuritis → i.m. injection (d/t)

– Symmetrical AFP :

• Transverse myelitis
↳ Herpes, Varicella, mycoplasma
Level → Thoracic area.
Rx : high dose i.v. Methyl prednisolone.

• Guillian Barre Syndrome (AIDP)
↳ Demyelinating.
– Diarrhea (By Campylobacter jejunii).
– Weakness occurs after 10 days.
– Areflexia (DTR absent)
– Symmetric
– Ascending → diaphragm involved.
– Plateaus → 4wks.
– Sensory & autonomic changes.
– Also d/t Mycoplasma; salmonella; S. pneumoniae.

CSF	1st wk	2nd wk
Cells	10/hpf	10
Proteins	50 mg/dl	500

→ Albuminocytological dissociation

– B/L symmetric demyliniating illness.

IOC for calcification : CT scan.

Rx : i.v. Ig (2g/kg)
↓ fails
Plasmapheresis.

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IV Ig indications:

- ↳ Kawasaki
- ↳ AIDP (Acute inflammatory Demyelinating Polyradiculo neuropathy.
- ↳ Hypogammaglobulinemia
- ↳ Rh isoimmunisation.

General Pediatrics:

- Diarrhea
- ORS
- Zinc
- Malnutrition:

ORS:

ReSoMal	Mmol/L	Old (WHO-ORS)	New/Universal
45	Sodium	90 (Cholera stool loss)	75 (Rotavirus 50-70)
40	Potassium	20	20
125	Glucose	111	75 (for facilitate diffusion of Na in cell)
70	Chloride	80	65
7	Citrate	10	10
+Mn, Zn, Cu		311	245
300			Low osmolarity ORS.

Citrate improves the self life of ORS.

Resomal → Rehydration sol^{n} for malnutrition.

Hyperkalemia Hypernatremia.

Substrate concentration of components of ORS sol^{n}:

NaCl — 2.6gms
KCl — 1.5gms
Trisodium citrate — 2.9gm
Glucose, anhydrous — 13.5gm

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WHO dehydration:
- No
- Some → Skin pinch slowly; thirsty
- Severe → skin pinch very slowly; lethargic; oligurea.

Rx: No dehydration:
- Replace ongoing losses.
- 5-10 ml/kg.

Some dehydration:
- ORS 75 ml/kg over 4 hrs.

Severe dehydration:
- i.v. Ringer lactate 100 ml/kg.

	30 ml/kg	70 ml/kg
>1 yr	0.5 hr	2.5 hrs
<1 yr	1 hr	5 hrs.

WHO Zn in Acute Diarrhea:
- 2 X RDA for 2 weeks.

Zn dose	RDA	Diarrhea
>6 months	10 mg	20 mg.
< 6 months	5 mg	10 mg

Acrodermatitis enteropathica (periorificial dermatitis)
- d/t Zn deficiency.
- Nutritional
- Genetic (AR) → Intestinal Zn transporter defect.
- Low Zn levels.

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- Improve on Zn supplement.

℞ — Zn (3mg/kg) — elemental Zn.

Malnutrition:

	Marasmus	Kwashiorkar
Muscle Wasting	++	++
Edema		++
Hepatomegaly		++
Low albumin		++
Pigment		++
Apetite	Voracious	Poor
Sensorium.	Alert	Lethargy.

Weight/Age Criteria (Indian Academy of Pediatrics)

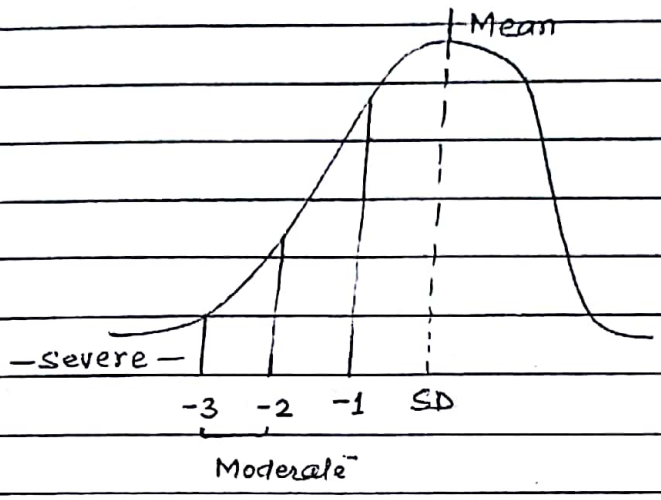
- Normal >80% refrence.
- Grade I → 71-80%
- Grade II → 61-70%
- Grade III → 51-60%
- Grade IV → ≤ 50%

for Edema ⇒ Add K
↳ Bad marker.

WHO

	Moderate	Severe
Weight/Height	71-79%	≤ 70
Acute (wasting)		
(N) > 80% reference		
Height/age		
Chronic (stunting)	86-89%	<85%
(N) >90% reference		

Symmetrical Edema ++++



Severe Acute Malnutrition:

Among children — 6-59 months of age.

Any of the following:

① Weight for height below -3 Standard deviation (SD or Z scores) of the median WHO growth references.

② Visible severe wasting.

③ Presence of bipedal edema.

④ Mid-arm circumference below 11.5 cm.

Below 6months → Mid-arm circumference can't be used.

Criteria for admission — If child fails apetite test.

Criteria for passing Apetite test.

Body wt. (kg)	Min^m amount of RUTF (Ready to use therapeutic factor) to be consumed for passing Apetite test (mL or grams)
< 4kg	15 mL
4-6.9	25 mL

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7 – 9.9	35 ml
10 – 14.9	50 ml

Mx:

Complications of SAM

Mx c in 1st 2 day
- S = Sugar (Blood Sugar < 54 mg/dl (OR) < 3 mmol/L)
- H = Hypothermia (< 95.5F (OR) < 35.5°C)

c in 1st wk
- I = Infections (TB, Malaria, UTI)
- EL = Electrolyte (Hypokalemia & Hypernatremia)
- DE = Dehydration
- D = Deficiency of Vitamins & minerals.

	Step	PHASE: STABILIZATION Day 1-2	Days 3-7	REHABILITATION Weeks 2-6
1.	Hypoglycemia	→		
2.	Hypothermia	→		
3.	Dehydration	→		
4.	Electrolytes	→		→
5.	Infection	→		→
6.	Micronutrients	No iron		c iron →
7.	Cautious feeding	→		
8.	Catch up growth.		→	→
9.	Sensory stimulation	→		→
10.	Prepare for follow up.		→	→

Feeding Rehabilitation: Cautious feeding
- F-75 containing 75 kcal/100 ml & 0.9 g protein/100 ml.

B = Begin feeds
E = Energy dense feeds.
S = Stimulation
T = Tender love & care

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Days	Frequency	Vol/kg/feed	Vol/kg/day
1-2	2hrly	11 ml	130 ml
3-5	3hrly	16 ml	130 ml
6-7	4hrly	22 ml.	130 ml

Energy dense feeds:

After the transition give:

- Frequent feeds (at least 4-hrly) of unlimited amounts of a catch-up formula.
- 150 - 220 kcal/kg/day.
- 4-6gm protein/kg/day.

Criteria for discharge:

- Weight for height > 80% of Reference standard.
- Edema should be absent for 2wks.
- MAC > 12.5cm.
- Weight gain > 5g/kg/day X 3days.
- Apetite is good.
- Complete antibiotics.
- Care taker should have learnt, motivated.

Weight for age < Acute (Wasting)
Chronic (Stunting)

Age independent criteria — MAC (>12.5cm)
6/w 1 to 5yrs.

Age independent index — Kanawati & McLaren index
Rao & Singh index
Dugdale index.
Quac stick index
Jeliffe ratio

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Osmolality = 2 X[Na] + [Glucose]/18 + [BUN]/2.8.

Pediatric Nephrology:
- Development
- Oliguria ; polyuria
- Hematuria
- AKI
- CKD
- Nephrotic, Nephritic
- UTI

} Topic

Development:

GFR:
- Newborn → 15 - 20 ml/min/1.73 m^2
- 3 months → 2/3rd adult
- Like adults → 2yrs of life.

- Tubular conc^n:
  - Adult morning urine osmolality >800 mosm/kg.
  - like adult 1yr.

- Nephrongenesis complete @ 36 wk of gestation.

- Barker's hypothesis:
  preterm & IUGR → hypertension in 2nd to 3rd decade
  ↳ Bcoz they have less no. of nephron.

- M/c asymptomatic abd. mass in 1-5yrs — Wilms tumour
- M/c abd. mass in newborn — Multicystic dysplastic kidney.
  ↓
  non-functional mass → bunch of grapes
  - 80% U/L.

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OLIGURIA:
- U.O. < 1 ml/kg/hr
- Common in AKI & Acute GN.

Causes of Non oliguric AKI — Aminoglycosides
— Neonatal renal failure.
— Resolving ATN (polyuria)

POLYURIA:
- U.O. > 4-5 ml/kg/hr. [Polydipsia ; Polyurea]

Approach —
— Blood glucose [RBS > 200 mg/dl
or FBS > 126 mg/dl]
↳ DM
— Venous Blood Gas.
(i) Hypokalemic hypochloric metabolic alkalosis
↓
Barter Syndrome ; Gittle's [illegible] Syndrome.

Bartter's Syndrome	Gitelman Syndrome.
— Severe ; Early.	— Mild ; Well preserved.
— Infancy	— Older child
— Antenatal → Polyhydramnios	— Hypo Mg
→ Nephrocalcinosis	— Hypocalciuria.
— Hypercalciuria	

(ii) Hyper Cl, hyper K, Normal anion gap metabolic acidosis
↓
Renal tubular acidosis.
(iii) Normal < Psychogenic] Water deprivation in psychogenic → Urine osmolality doubles
DI

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DI → Central / XLR Nephrogenic
- On Vasopressin challange, urine osmolality doubles by 100% in central. Hence differentiated from XLR Nephrogenic.

Q 4 yr; Polyuria & Polydipsia
Venous blood gas normal
Urine Osmolality
- Baseline 50 mosm/kg.
- On water deprivation = 60 mosm/kg
- On Vasopressen challange = 70 osm/kg.

Δ = XLR Nephrogenic DI
DOC = Thiazides.

Best method of GFR estimation = Insulin clearance.

Schwartz eGFR formula:

$$= \frac{K \times \text{height in cm}}{\text{creatinine (mg/dl)}}$$

Formula depends on:
- Height, muscle mass.
- Method of estimation of creatinine. [Jaffe's reaction].

S. creatinine is accurately measured by
- Enzyme assay / HPLC.

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Value of k in Schwartz formula:

Low birth weight infant	— 0.33
Normal infant 0-18 months	— 0.45
Girls 2-16yrs	— 0.55
Boys 2-13 yrs	— 0.55
Boys 13-16yrs	— 0.70

Schwartz method is independent of — Renal function.

HEMATURIA:

Red urine:
- Beet root
- Phenolphthalin
- Rifampicin

Q. 5yrs old, Red urine. Urine 1-2 RBCs/hpf — Intravascular hemolysis.

If Red urine → look for RBC's.

−	+
↓	↓
Hemoglobinuria	Hematuria
Myoglobinuria.	

Hematuria:
- Gross
- Microscope → >5 RBCs/hpf on centrifuged urine

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	Glomerular	Extra glomerular
Cause —	- IgA ; MPGN; PSGN.	- Cystitis; Stones; Idiopathic hypercalciuria ↓ 24hr urine Ca^{2+} > 4mg/kg.
Dysmorphic RBC	++	
Colour	Cola	Bright Red.
Pain	Painless	Painful.
Protein in urine	Proteinuria	

Recurrent Gross Hematuria:
- IgA
- MPGN
- Idiopathic hypercalciuria.

PSGN doesn't reoccur.

Alport's Syndrome:
Triad:
Eye : Ant. Lenticonus
Ear : SNHL
Kidney : 75% Boys ESRD before <30yr.

- 80% X linked > AR 15% > AD 5%
- Collagen IV → α-5 domain (in GB membr)

Good pasture Syndrome — α 3 domain of collagen IV abnormality.

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INV: Electron Microscopy.
↳ Basement membr is irregular.
↳ Splitting of the lamina densa
↳ Lamellation
↳ Straited GBM
↳ Basket weave appearance (classical)

Slit lamp examn → Keratoconus
Leukoconus

Acute Kidney Injury (AKI):
Best Biomarker → ① Urine NGAL
[Neutrophilic gelatinase-associated lipocalcin].
② Urine IL-18
③ Urine KIM-1 (Kidney injury molecule)
④ Urine L-FABP (Fatty acid binding protein).
⑤ Serum cystatin-C

Types:
- Prerenal
- Renal
- Post renal

PRERENAL — Hypotension, Hypoxia
Newborns → hypovolemia, burns, diarrhea.
RENAL — ATN > HUS
POST RENAL — Obstruction.

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Indices	Pre renal	Renal
Urinary Sodium (meq/L)	< 20	> 40
Urine osmolality	> 500	< 300
B. urea / Creatinine ratio	> 20 : 1	< 20 : 1
Fractional excreation of Na%	< 1	> 1

$= \frac{\text{Urine Na}}{\text{Serum Na}} \times \frac{\text{Serum Cr}}{\text{Urine Cr}}$

Cause of AKI:

M/C/C in Children / Adults

Prerenal → ATN

↳ Hypoxia

Hypotension

Drugs ← Exo ← Toxins

Sepsis

Hemoglobinuria ← Endo

Myoglobinuria

Hemolytic Uremic Syndrome (HUS):

- Microangiopathic hemolytic anemia
- Thrombocytopenia
- AKI

- 90% follows diarrhea.
- Developed : E. coli 0157 : H7.
- Developing country : Shigella dysentaie type I.
- Germany June 2011 : E. coli 0104 : H4

SHIGA toxin → Cause Endothelial injury

↓

TMA (Thrombotic microangiopathy).

On PBS → Schistiocytes are Dstic of HUS

Rx: ECULIZUMAB [DOC] → for PNH
↳ drug against C5.
If not available → Plasmapheresis.

Complications: (↱ Insensible losses – 400ml/m²)
- Fluid overload → Rx → Fluid restriction
- Hyperkalemia → cause arrythmia / Sudden death.
- Dilutional hypo Na. Rx – Restrict fluid
- Dilutional anemia Rx – PRBCs if Hb < 6
- Metabolic acidosis.
- Hyper PO4 → Hypo Ca
- HTN.

Hyperkalemia:
Rx – Glucose & insulin intravenously
↓
c/ in 10–15 min.

- Ca^{2+} stabilises cardiac memb^{r} potential.
↳ only given in ECG changes in Hyperkalemia.
↓
Calcium Gluconate i.v.

~~Mechanism~~: Hyperkalemia Rx:
Transcellular shift into cell.
- Insulin c/ Dextrose.
- Nebulised salbutamol.
Cardioprotective → i.v. Calcium Gluconate.
↑ delivery Na to distal
• Furosemide
• i.v. $NaHCO_3$
Dialysis . .

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· K-binders → Kaexylate polystyrene.

CKD (Chronic kidney disease):
Causes in Children —
- < 5yrs — Hypoplasia
  Dysplasia
  Posterior urethral valves (Boys)
- > 5yrs — Acquired
  ↳ GN / HUS.

Features:
A = Azotemia
Acidosis (metabolic)
Anemia (Normocytic Normochromic)
↳ Rx — S/C rh EPO (Erythropoietin).
B = Bone ds
C = Cardiovascular complication ↑
G = Growth failure ← Multifactorial causes.

When GFR ↓ — 40-60 ml/min/1.73m²

↓ ↓

↑ PO4 ↓ 1 α-OH

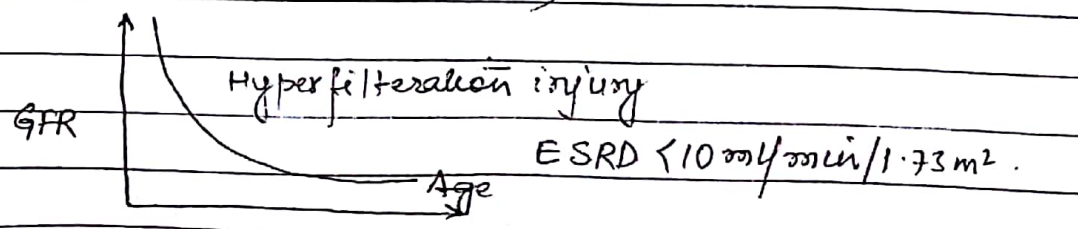
↓ ↓

↑ PTH ↓ 1, 25 vit·D.

↓ ↓

↓ Ca^{++}

Osteodystrophy
(CKD - MBD)



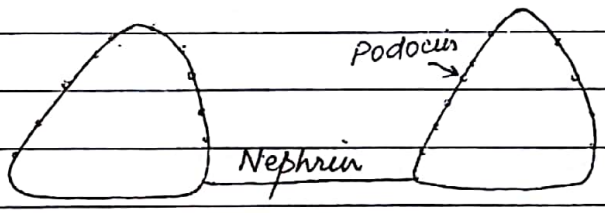
Nephrotic Syndrome:

- Proteinuria > 40 mg/m²/hr or > 2g/24hr
- Hypoproteinemia (Hypoalbuminemia)
- Hyperlipidemia (cholesterol > 200 mg/dl)
- Edema : S. Albumin < 2.5 g/dl.

On light microscopy — Minimal changes.
On Electron " — Effacement of foot processes of podocytes.

DOC : Prednisolone

Cause of Edema in NS → Na^+ & H_2O reabsorption.



Gene	Protein	Disease
NPHS1 (chr 19)	Nephrin	Finnish Congenital nephrotic (< 3 months) Rx : Nephrectomy.
NPHS2 (chr 1)	Podocin	Steroid resistant FSGS Rx : Calcineurin inhibitors ↳ Tacrolimus > Cyclosporine

Steroid toxic ; Steroid dependent

- Cushingoid
- HTN
- Post. subscapular cataract
- Impaired glucose tolerance
- Short

Rx – Oral cyclophosphamide for 12 wks.

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UTI (Urinary Tract Infection):

Definition: Symptoms + Urine culture $>10^5$ CFU/ml

- Symptom — Fever
- E. coli
- Females [Boys <1yr]
- Ascending [<1yr → Hematogenous]

Best urine specimen — Suprapubic aspiration.

- Asymptomatic bacteriuria → Shouldn't be treated.

- M/c/c of UTI in children → VUR
  IOC for VUR → MCU

VUR:
- Polar scarring → DMSA nuclear scan
- Function → MAG3 / DTPA nuclear scan.
- Prophylaxis → Antibiotic of choice
  ↓
  Cotrimoxazole.

Sx → Reimplantation of both ureter.
↳ Indication:
- Breakthrough UTIs
- Deterioration of Renal function.

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GLOMERULONEPHRITIS:

- Hematuria
- Edema
- HTN

M/c/c of GN → Post streptococcal GN.

Acute post streptococcal GN:
- follows infection of throat (serotype 12) / skin (serotype 49) c nephritogenic strains of β-hemolytic streptococci.
- Age → 5-12yrs
- Acute phase resolves in 4-6wks but urine normalises in 1yr.

	4-6wks
Edema	X
Cola Gross hematuria	X
HTN	X

C3 — 6-8wks normalese.

In urine – shows microscopic hematuria
↓
6-12 months. to resolve.

Sore throat → Δ by ASO titre (1-2wks)
Pyoderma → Δ by anti DNAase B (4-6wks)
95% PSGN resolves
5% PSGN → CKD

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Kidney Bx → Endothelial & mesangial cell proliferation & obliteration of capillary lumen.
- Neutrophil infiltration

Immunofluorescence — Granular deposits of IgG & C_3
"STARY SKY"

Good pasture Syndrome has linear deposits of IgG & C_3.

Sub epithelial humps are seen in PSGN.

Q Persistently ↓ C_3 found in all except:
a) Post streptococcal GN (normalise after 6 weeks)
b) Mesangio capillary GN
c) Cryoglobulinemia
d) SLE
e) IE
f) Shunt nephritis
g) Factor H- mutation → HUS.