

PEODGATPCS

PG-NOTES

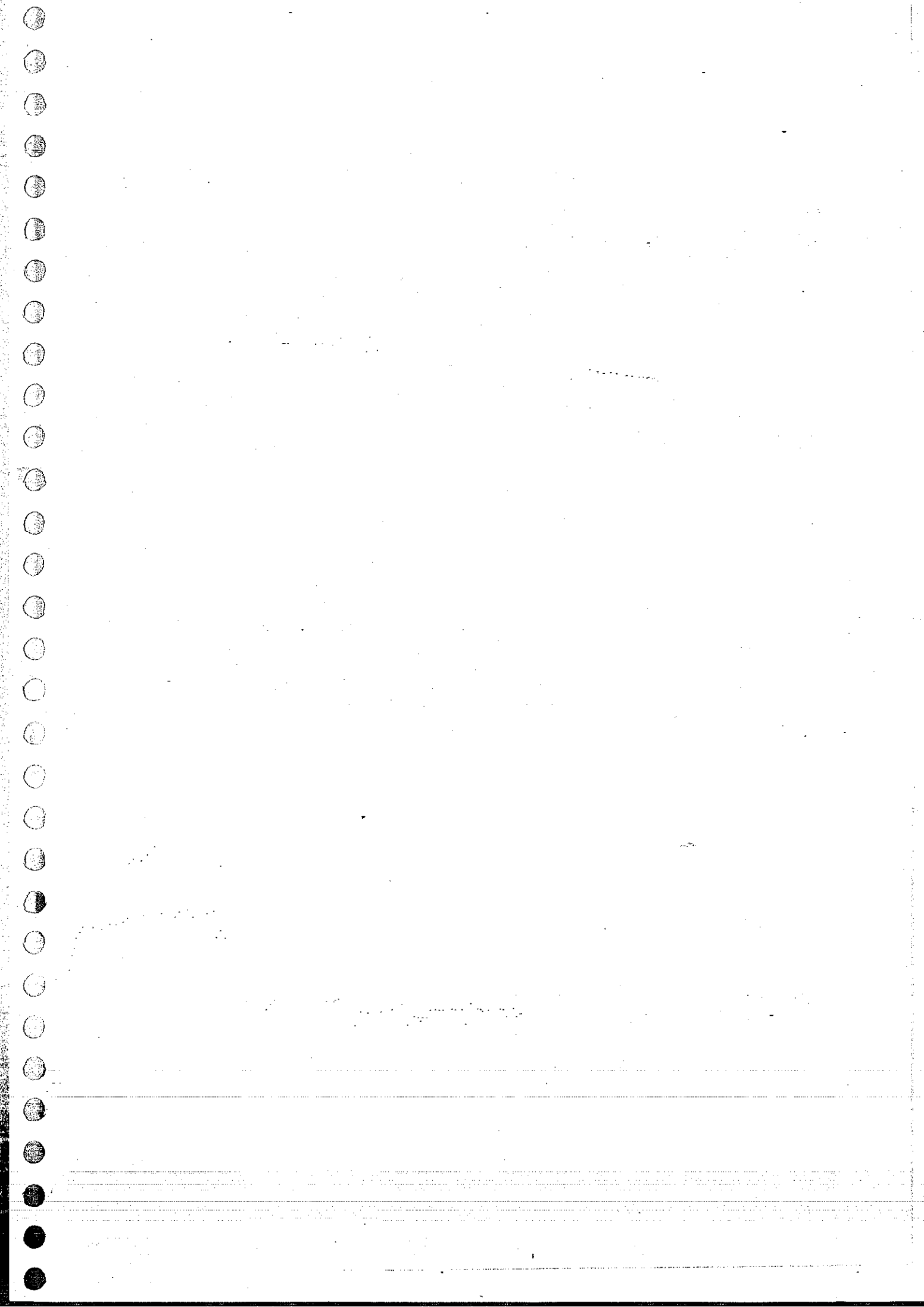
2017

RENOVATION

JAN STATIONERY

PH: 9654691327

9811982449



→ NEONATOLOGY →

8th JULY 2017

Normal Bth wt: 2.5-4kg

Macrosoma: > 4k

LBW: < 2.5kg

VLBW: < 1.5kg

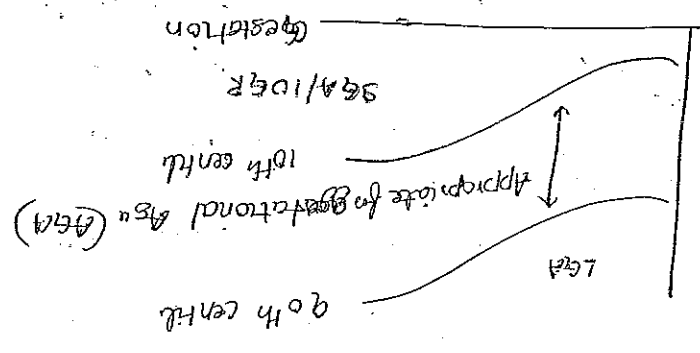
ELBW: < 1kg

Term baby: 37-42wks

Preterm: < 37 weeks

Postterm: > 42 weeks

→ Lubchenko Chart



⇒ Large for gestational age (Large baby)

Large baby: m/c constitutional

constitutional:

* LGA

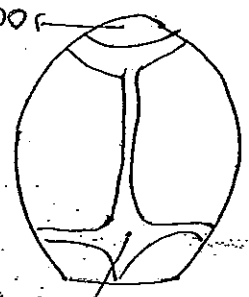
* AI 07 Short stature

* AI 08 delayed puberty



All term babies have 6 fontanelles

Anterior fontanelle (diamond shape, 2.5 x 2.5 cm, closed at birth, at level, pialatal)
 closes at 8-24 months



closes at birth

⇒ Delayed closure of fontanelle

* Hypothyroidism

* Hypophosphatasia

* Cleidocranial dysostosis

* Bones

* Unilateral hernia

Constipation

Prolonged physiological jaundice

Longer interval Hypothyroidism

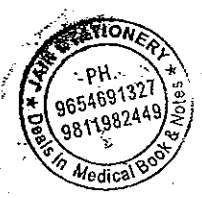
→ Lower femur delayed epiphysis → Epiphyseal degeneration at the level of head of humerus } Hypothyroidism

→ Neonatal screening @ Tandem mass spectrophotometry

① TSH T4 ② acylcarnitine ③ galactose ④ CAH ⑤ Biotinidase

Best time to do TSH: Am) in 1st 48 hrs the baby have

TSH Surges



Amman AP

* 24-48 hrs to 6 days best time is 72 hrs

* 72 hrs after birth

* OP4 3-5 days life

* m/c of hypothyroidism: agenesis / dysgenesis of thyroid gland

CRANIOSTENOSIS:

→ Premature closure of skull suture (one or all sutures)
• complication: ① Microcephaly

↑
Intracranial tension ↑

↓
optic atrophy

② Abnormal shape

Cranio stenosis → Rx: craniotomy

Syndr of cranio stenosis: Apert synd

AD

→ "Dragon"

→ "Carpenter" → Autosomal recessive

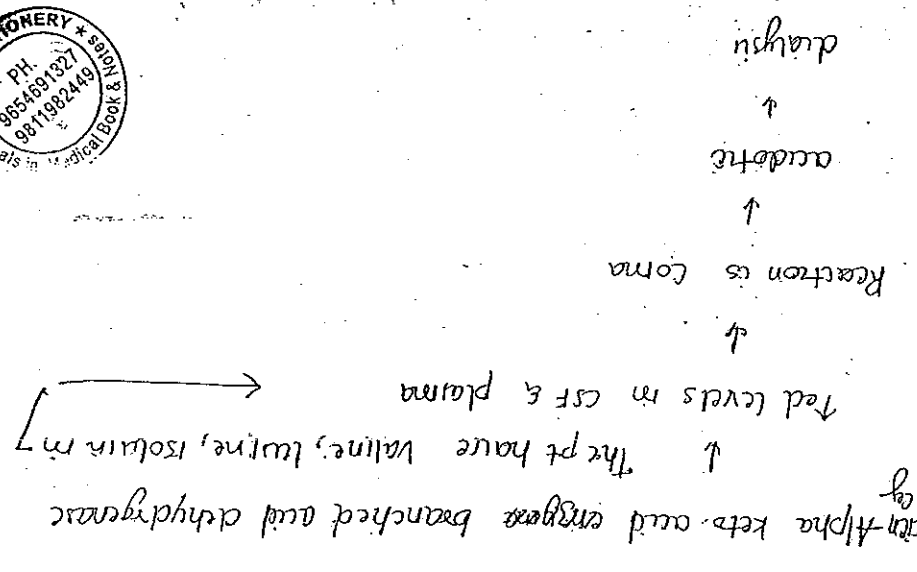
→ "Puffer"

In dehydration the fontanelle depressed
In raised ICP the font "bulging"
Crying infant → bulging and pulsatile





For this a screening test is present = Tandem Mass spectrometry
used to see early



Maple syrup urine disease

* Aim: serum Phe < 6mg/dL

* New tyrosine is essential

* Restrict phenylalanine

* Supplement tyrosine

Melanine

DOPA

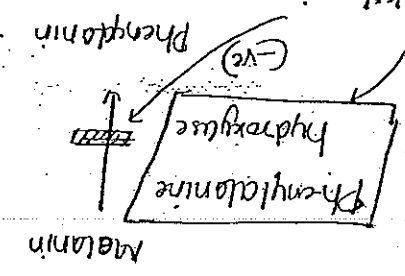
Tyrosine

Tyrosine

toxic

Phenyl ketouria

Phenylalanine → toxic



URINE AND MECONIUM

↓
Rt meconium in 24 hr
1st urine in 48 hr

→ MECONIUM ←

⇒ Delayed meconium passed

• Hirschsprung / Aganglionic

• Imperforate anus

• cystic fibrosis = Meconium in ileum

• old question) 48 hr old baby not passed meconium

what is the next investigation

Ans) lower GI contrast

↓
uscdm [Asu] of Hirschsprung

↓
Candido / treat Hirschsprung

→ URINE ←

Child has B/L renal agenesis

↓
Rt urethral valve

↓
oligohydramnios (amnionic fluid of mother has

crine of baby after 20 wks)

↓
B/L hypoplastic Pulm hypoplasia

↓
fetus goes in POTTER'S SEQUENCE

Potter's facies: pinched in nose

retrognathia

B/L hydronephrosis





(short stature)

in Hypothyroidism: $UB:LS$ is $\nearrow$ ed (disproportionate)

Adults 0-9:1

At 10 yrs 1:1

Newborn 1.7:1

Ratio

Upper segment = Head to pubis symphysis
Lower segment = total length - upper segment

By 1yr, $CC > HC$

HC becomes equal to CC at 9-12 months

b) Asymmetric IUGR

a) Congenital Hydrocephalus

If more than > 3 cm

$HC > CC$ not more > 3 cm

Head circumference 35 cm

length 50 cm

ANTHRAPOMETRY

Rx $\rightarrow$ Gynaecologic evaluation

$\rightarrow$ Maturing cytology

ACHONDROPLASIA

* short, limb dwarfism

Limbs are short

US/LS ratio < 1

→ complication of IVGR @ small babies →

* Chromosomal / TORCH infections

* Have asphyxia = can also cause meconium aspiration

* asphyxia = PPHN

synd (MAS)

Persistent Pulmonary HTN @ New born

* Limited stores = Hypoglycemia

Hypothermia

Hypocalcemia

Polycythemia

Neutropenia

thrombocytopenia

⇒ infection and PDA more common in preterm

* IVGR √ symmetric





De apperance 3 month of life

Adduction / complete 36-38 wks

& Apperance in 28-32 wks gestation

adduction & flexion

↓

↓

↓

genuine of firm

extreme at elbow joint

Moro reflex & Abduction of shoulder joint

REFLEX ES

$$\frac{\text{wt(gm)}}{\text{Length (cm)}} \times 100 = \text{if } < 2 \text{ Asymetric logR} \\ > 2 \text{ symmetric logR (w) More}$$

Ponderal & Index

Brain / Liver ↑

HC > CC >>> 8cm

↓

Brain spared

small

Brain size less
cell size less
cell-n less

Normal

Placental

Chromosomal

Asymmetric

Symmetric

Asymmetric Moro's

Brachial plexus

~~HLT~~ # device: # future

→ Hemiplegia (more in (R) hand)

flaccid it will away: appara atrophy

exaggerated Moro's HIE-1, toy sack

(Hypoxic ischaemic encephalopathy)

Grasp reflex

x Apper 28 cks gestation

+ fully appears fully developed by 32 weeks

disappeared 3 months

SOCKING AND FOOTING

Apper 32 cks gestation

Coordinated w/ - 34 cks

well delayed 36 weeks

After 1 month of life, rooting in low

prominent





seen in newborn & well elicited at 2 months of life

2nd phase = 1/2 neck u. flexed, tone ↓ in all limbs

1st phase = Neck extended, tone ↑ in upper limb, ↓ in LL

→ Symmetric tonic neck reflex

After 6 months baby rolls

* disappear 6 to 7 months of life

* fully developed in 5th month of life

* appear at 35 weeks of gestation

Neck

* extension of one arm & flex at other arm

→ Asymmetric tonic neck reflex →

Internal: Keton / paladog - expressed breast milk

33 wks gestational, 1400 gm fed?

ATP4

External: Naga, 2000 gm fed? still

31 wks gestational, 1400 gm fed?

ATP4

Racinate reflex:

Not in a newborn

appear at 7-8 months

well develop at 10-11 months

Results for life

MILIA: distended sebaceous glands on face and nose
↓
Reassure the mother

ERYTHEMA TOXICUM:

erythematous patch on face & trunk

usually seen on day 2 - day 3 of life (Reassure)

ACROCYANOSIS: Normal in newborn

[Cyanalgia is always abnormal everywhere]

STORK BITES:

pinkish greyish papillary haemangioma

EPSTEIN PEARL: white spot on palate

epithelial inclusion cyst on palate & peripose

Natal teeth: (per deciduous teeth) lower incision position

Midline vaginal bleed: on 5th and 7th days (N) condition

due to mother hormones in child with breast

(Question) Birth wt 3000gms (after birth all babies less wt

4 days 2700gms

10% of birth wt and regain
the birth wt by 7 days of life)





extremities are cold but soles warm

* Cold stress 36.5°C to 36.5°C

* (N) baby should have pink & warm palm and soles

36.5°C - 37.5°C

* (N) core temp is :
→ Core body temperature: temp of middle ear & oesophagus

* Around blood vessels [cardiac, mesenteric]

* adrenal, kidney

* INTERSCAPULAR

* Nape of the neck

Non-shivering thermogenesis

New born cannot shiver bcoz brown fat

→ TEMPERATURE REGULATION →

Bacteraemia → Abnormal

1-2 pm cells / hpf

Protonuria

glucosuria

(Question) which is not normal in new born

less water more and in lot

the tubules of babies are immature and

↓
Bcoz: Physiological diuresis

[Preterm child loss 15% and regain weight]

* Hypothermia $< 36^{\circ}\text{C}$

Severe $< 32^{\circ}\text{C}$

↓
so should do KMC kangaroo mother care

* improve breast feed

* bed infection

* bed hospitalization

NEET '15 In KMC all are KMC component except:

kangaroo position

" Nutrition

Early discharge, followup

xv supplementation nutrition

NEET '16 when to stop KMC = for 2500gms

temp of NICU & labour room to prevent hypothermia

u $26-28^{\circ}\text{C}$

hypothermia: Brain ischemia

Heart - aphyxial cardiomyopathy

Lungs ARDS

hypoglycemic, hypoa: Hypoing

kidney: Aphyxian tubular necrosis

⇒ heat in a Radiant warmer

baby less heat by convection

if baby $< 1000\text{gms}$ put him in polythene cover





which reduces the convective heat loss

⇒ Fetal alcohol syndrome

thin upper lip.

Features:

① skin folds on lower eyelid

② absent furrow

③ microcephaly

④ small eye openings

⑤ flat & small mid face (Mid facial hypoplasia/Micrognathia)

⑥ short nose

⑦ low nasal bridge

⑧ septal defect

⑨ small palpable fissures

PREMATURITY →

* Respiratory: RDS; chronic lung disease/

Broncho pulmin dysplasia

Dependence at 4 weeks of life

[Vita to ELBW prevents Bronchopulmin dysplasia]

CNS [Apnoea]

↑
cessation of resp in newborn > 20 min (and)

as central cyanosis and bradycardia

3 types of apnoea: ① central apnoea

↑
Extreme preterm, asphyxia

② obstructive apnoea

[all newborn are nose breathers till 4 months]

question) Mother when child feeds = blue
↑
cyan = pink color

classical H/o of

8/7 choanal atresia

↓
vll is asymptomatic

③ Mixed apnoea (M/c apnoea) in preterms

Apnoea of prematurity: bcos of prematurity

(Risk 100% in < 28 wks)





the meninges

a seizes classical 4/10 of intraventricular

(Reaction): Reterm: sudden pole, shock, frontal bulging

Subdural common in term babies

→ In preterm all hemorrhage are common

CNS [INTRACRANIA HAEMORRHAGE]

1.5 mg/kg once daily

20 mg/kg [Loading Dose]

↑ because it has wide margin of safety

↓ (Am) caffeine citrate

↓ AIP 2007: Doc. for apnoea prematurity is:

every 6-8 hours

↓ 1/6 maintenance dose u 1-2 mg/kg

↓ (Am) 5-6 mg/kg

↓ All M's 2006: Loading dose of aminophylline

③ caffeine citrate

② methylxanthines < theophylline

treatment: ① Nasal CPAP

never after 7 days

upto 7 days

onset: Most 1-2 days

Risk < 1500 gm = 30%

IVH occurs in
 50% in pt 24 days
 75% in 1st 72 hrs

term baby and breech

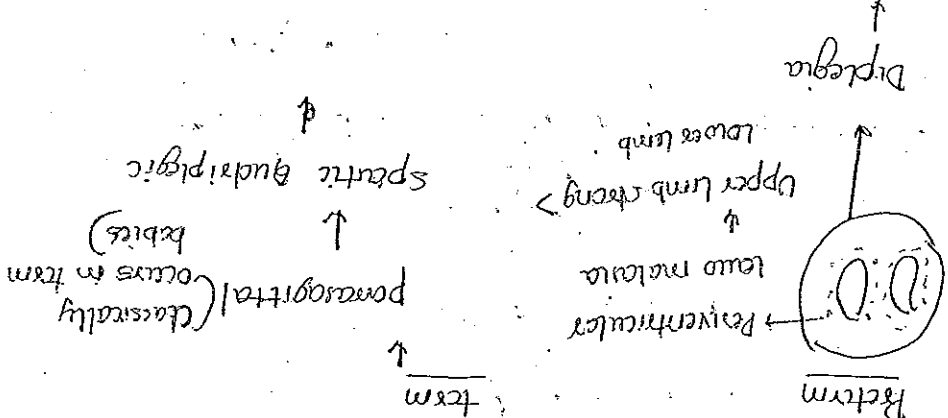
common for IVH

in preterm (capillary are fragile in germinal matrix in brain)

⇒ All intracranial hemorrhage cause ICH

CNS [Asphyxia = lack of O_2]

(question) Baby got Asphyxia < term (a) preterm



parasagittal (classically occurs in term babies)
 ↑
 sporadic quadriplegic

spastic diplegia (type of cerebral palsy)
 Atoms) m/c sequel of PV leucomalacia
 spastic diplegia
 " Quadri
 Seizures
 Mental retardation



shrinking of ventricle }
 passive of white matter }
 PVL

IOC: MRI

Goldstrand: MRI (for AVM)

→ Status Marmoratus: diffuse neuronal loss

→ CPR (outside the Hspt) / Asphyxia

Pathogenesis of HIE: Ischemia reperfusion injury
 ↓
 free radicals

Rx: Therapeutic Hypothermia /
 ↓ selective head cooling 33-35°C

↓ brain metabolism

↓ free radical release

↓ injury

→ Hypoxic Ischemic Encephalopathy (due to Hypoxia)

↳ MLC/ensure in a neonate

staging of HIE: 3 stages
 ① ② ③

stage 1: hyperalert child

tone (N)

reflex hyperactive

pupils Mydriatic

fixed progress





**** Stage 2:**
consciousness: lethargic
pupils: Miosis
reflex: hyperactive
Seizures: seen
variable prognosis
Stage 3:
↓
Baby will die
stupor, coma
flaccid
Decelerate
seizures: Deceleration
severe neurological damage
Aims: stage 2 & neonatal seizure; phenobarbital
DOC: phenobarbital
Cardiovascular system
CVS [Hypotension]: [PDA] ↑
common in preterms
→ PDA: Patent Ductus Arteriosus
↑
prostaglandin release
Rubella synd
* H & a Vessel Wall Defect
→ PAS → asplenia
→ rubella
term



• Maternal cocaine

Top fid
spec. toxins
[Ischemia to Gut
→ immature Gut

Risk factor of NEC: only in preterm

[NEC] erythrogenic - retrocolitis

Gastrointestinal system

continuous mechanical

* Upper ① sternal border ↑

Allms) what type of Pulse present in PDA: wide

* O/E: bounding pulse & wide pulse pressure

CO₂ wash out

Allms) what is not seen in PDA is

CO₂ ↑, O₂ ↓

↓
Failure wean off from ventilation

* It can also present in a preterm

* Present 6-10 wk & CHF

* less nephrotoxic than ↓

↑ IBraun (c) Endomethasone

Medical Mx

Simple Sx: Interventional

↓
Toc: Surgical Mx - (superior)

May use Probiotics in rotavirus diarrhoea

Lactobacillus rhamnosus
 Saccharomyces boulardii
 only rotavirus

NEC staging: Modified Bell's staging: 3 stages

1 stage 2 stage 3 stage

- Ia. Definite → focal pneumatosis
 - Ib. Definite → Distuse pneumatosis
 - IIa. Advance → Distuse impending perforation, shock
 - IIb. Advance → Perforation, shock
 - IIIa. Advance → Peritonitis, Pneumoperitonium
 - IIIb. Advance → Peritonitis, Pneumoperitonium
- AIIIS: portal venous gas
 is only seen; HMT
 Hyponatremia
 Metabolic acidosis
 Thrombocytopenia

⇒ Pneumatosis intestinalis → air in gut wall

AIIIS) Neonate come distended abd and perforation

is due to: NEC

[90% of neonates in picture]
 2-3 wks of life

Management:

① NPO

② PPN (Total parenteral nutrition)

③ Antibiotics against E-coli

cause perforations pneumonitis

Cefotaxime, vancomycin, metronidazole



④ stage III: NR child & perfusion & poor general condition (shock)

stage III
 stable → laparotomy
 unstable → portocaval drain

Frank drain & glove

EYE [retinopathy, ROP, retrolental fibroplasia]

Vessels are proliferating

dilatation, tortuous behind

the lens

these vessels cause tractional

Retinal detachment (stage V)

↓
 complement detachment (stage IV)

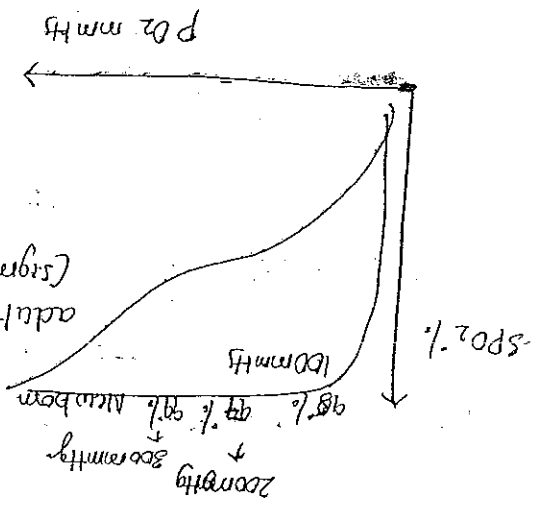
High flow O₂

Risk factor: only in Retin

at post pole

↓
 Plus disease

ROP - I to IV



Regular indirect ophthalmoscopy

Prevent blindness



* AAP ROP screening guideline

Gestation (wks) 1st visit (wks) Interval (wks)

22	31	9
23	31	8
24	31	7
25	31	6
26	31	5
27	31	4

Attn: Baby born in 28 wks gestation: Eye checkup

* 28	* 32	* 4
29	33	4
30	34	4

⇒ After 1st visit go to ophthalmologist every 2 to 3 weeks upto baby appear term.

⇒ According to AAP at risk ~~28~~ < 30 gest age / < 1500 gm eye check up. at 28 weeks

treatment:

- ① laser photocoagulation
- ② cryotherapy

③ V. retinal reattachment

④ Bevacizumab (anti VEGF) [use this drug in resistant ROP]



* limited Reserves : Hypothermia, Infection, Jaundice,

Anemia of Prematurity, Hypoglycemia, Hypocalcemia

→ NEONATAL SEPSIS →

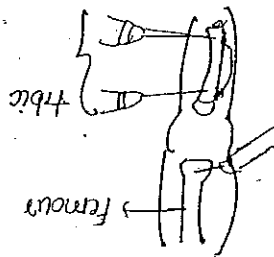
clinical features of sepsis + bacteraemia

Gold standard to dx sepsis is: Blood culture

Neonatal sepsis
 Early (1st 72 hrs)
 Late (1st after 72 hrs)

peripheral heel prick:

if we prick in centre it cause calcaneal osteomyelitis



Early sepsis: ① Risk factors are maternal chorioamnionitis

↓
 to mother ① foul smelling liquor
 ② fever
 ③

② Gp B strept, E coli
 overall avoid ward ↓

treatment: ampicillin + gentamycin



Late Neonatal sepsis (Nosocomial)

① cause: in India: *Klebsiella*, *Staph aureus*

World: *E. coli*

Overall India and developing countries:

Klebsiella > *Staph aureus*

② Meningitis is very common in late

so do CSF

③ treatment: cefotaxime + amikacin

↑
Sepsis

④ Sepsis screen for early asu

⑤ TLC [Leucopenia also suggest sepsis]

$< 5,000 / \text{cumm}$ (co) $> 20,000$

⑥ ALC $< 1500 / \text{cumm}$

Absolute
Neutrophil
cell

⑦ Rf for band cell $> 20\%$ immature total neutrophils

and toxic granules $I/T > 0.2$

⑧ Micro ESR (6mm-3 days) $> 15 \text{ mm fall 1st hour}$ Ristive

Red in infection

⑨ CRP, Procalcitonin better than CRP

↑
more specific for infections

⑩ Lumbar puncture (commonly done in late)

⑪ Chest Xray (for pneumonia)



⇒ In AHC we should include band cells

* Duration of antibiotic

Bacteraemia : 10 - 14 days

Meningitis : 21 days

Arthritis, osteomyelitis - 4-6 weeks

→ RESPIRATORY DISTRESS SYND →

Silverman - Anderson scoring system

Feature	Score 0	Score 1	Score 2
Chest movement	Equal	Resp lag	Scarred Resp
Intracostal Retraction	None	Minimal	Marked
Xiphoid retraction	None	Minimal	Marked
Naal Flaring	None	Minimal	Marked
Expiratory grunt	None	Audible	Audible
Stethoscope			

Dawson's Scoring for RDS

- ① Rep Rate
- ② Central cyanosis
- ③ Retraction
- ④ Grunting



⑤ Atrial entry (mid axillary line)

→ SURFACTANT ←

↓
(it reduce surface tension and prevent alveolar collapse)
impo component: Phosphatidylcholine

other component: Phosphatidylinositol

Phosphatidylethanolone

Phosphatidylglycerol

* In amniotic fluid after 28 weeks

mature level after 35 weeks

homogenate: 20 weeks gestation

* Atr bronchogram: specific feature on X ray in RDS

* RDS always occurs in 1st 6 hours

↓

so max neonatal mortality in 1st 6 hours

* RDS Rx: ① Nasal CPAP

[Mild to moderate RDS]

Early CPAP to ELBW babies

FIO₂ fraction inspired 0.2 to 0.6

and then room

PEEP - 5 cm H₂O

End expiratory pressure

⑥ Intra tracheal surfactant (insure approach)

In - Intubate

Sur - Surfactant

E → Extrubate



Survivator: Boine
 Unsurf: Percine

Infusing: calf lung extract

* Risk < 28 wks → 60-80%

* Risk 28-37 wks → 5%

* RDS never seen in 104R

104R → stress → cortisol

* Term, infant to diabetic mother

L/s ratio worse than 2

Lungs are matured

delivery can be done

L/s ratio > 3.5 → term, infants of diabetic mother

Question: All is true for CPAP except

1) Initiated EtO₂ 0.40-0.6

2) Used in apnea of prematurity

3) Improves compliance, vol, FRC

Functional Resp capacity

4) Early CPAP in extremely LBW

All are true

Question: Early lung transplantation

→ Neonatal Pulm Alveolar Reticulosis

Prominent interlobular septa [Crazy paving pattern]



SUNDAY 9/7/2011

Type C Tracheo oesophageal Fistula: } both & cyanosed child

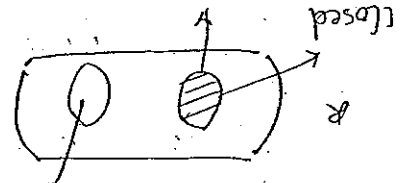
(Location) Saphoid Abd, Band shaped chest, heart pushed to the Right, Acute Bowel on L side, tracheal shift

Bag & Mask ventilation: Absolute C/I

⇒ Born
⇒ Diaphragmatic Hernia
Intubated
Heart shifted to its side
Reintubate

↓
intubated (Not stop)
↓
Mesogastric tube
(To decompress)

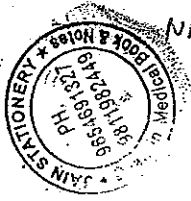
⇒ Pleuro Peritoneal Canals
85% cases open in
posterior Bochdalek
<5% are b/l
N close at 8 week
(D) Diaphragm are late associated



Pulmon hypoplasia (m/c of death in D hernia)

⇒ Most imp prognostic factor in D hernia: development of PPHN
persistent pulm HTN & newborn

Hypoxia



To prevent PPHN in CDH

* Elective intubation

* HFOV: High frequency oscillatory ventilation

To treat PPHN

* iNO inspired Nitric acid

* sildenafil

* Bosentan, Ambrisentan
[Aims Mags]

endothelin antagonists

* PGI₂ [iloprost]

* -mlodapine

* ECMO

m/c of distress in a term: Transient Tachypnea of New Born

(TNG)

(or)

→ Risk factor

Wet lung

term, cesarean (AI¹⁴)

* prominent horizontal fissure on R side

* Macrosomia (AI)

* precipitous labour

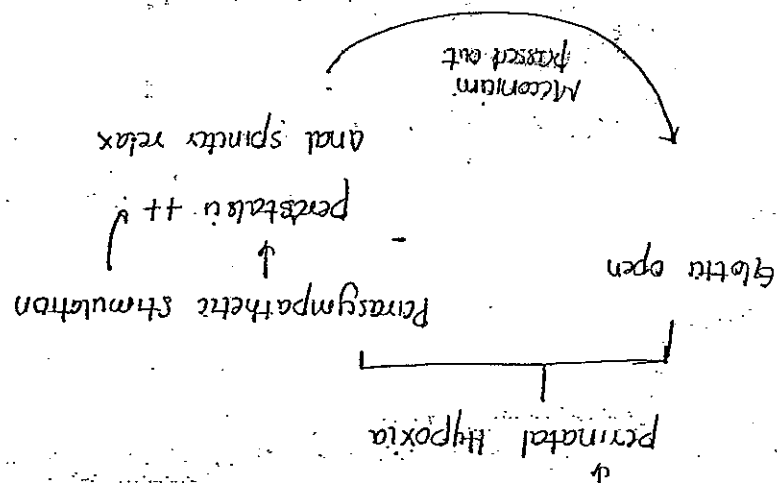
→ Benign; resolves 48-72 hrs

PO₂ > 0.4 (AI¹⁴)



→ MACONIUM ASPIRATION ←

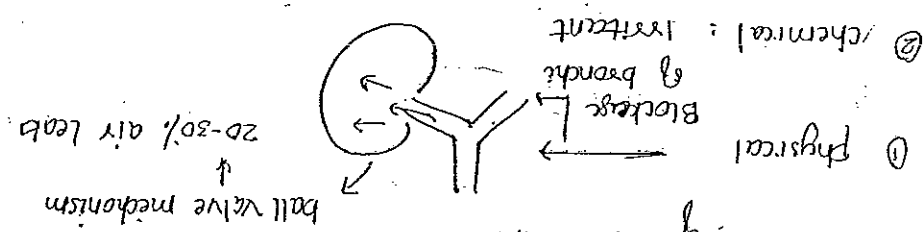
* Meconium stained liquor



+ It is common in post-term babies the placenta

become old and

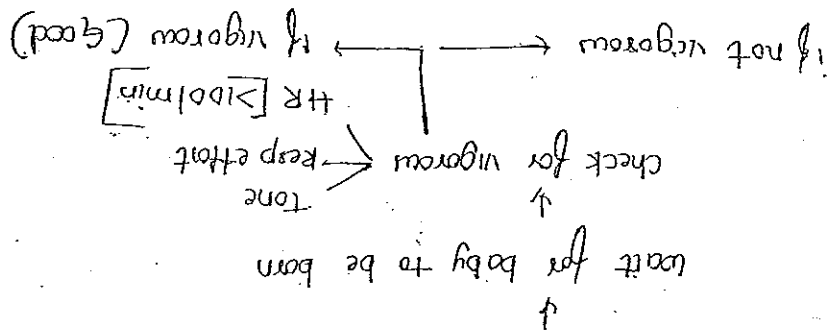
* Results of Meconium



③ Biological: Good culture medium (more prone to sepsis)

So they required antibiotic

* Management of meconium stained liquor



HR in newborn should be checked for 6 sec
 if 6 sec = 10 beats
 if 6 sec = 2100 beats

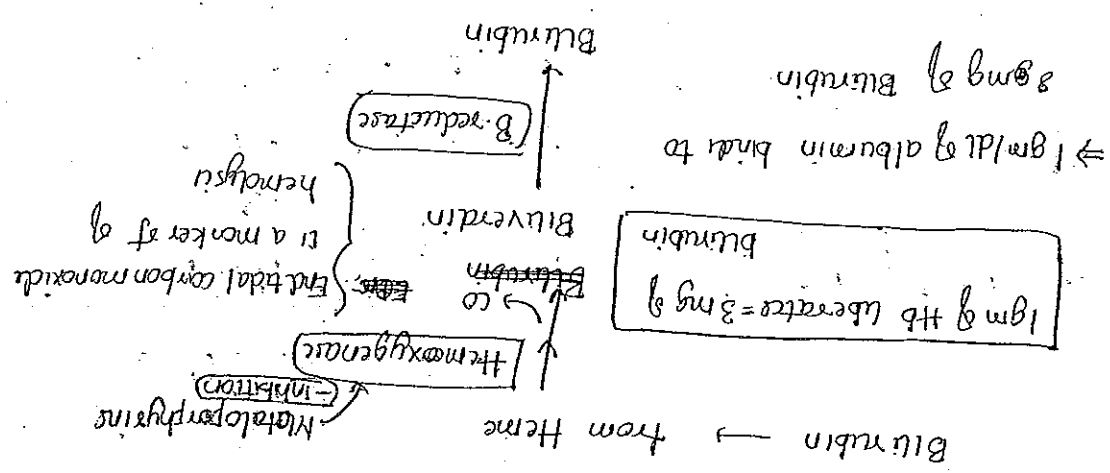
NRP 2015

If not vigorous
 (a) limp
 (b) apnea
 (c) <100/min

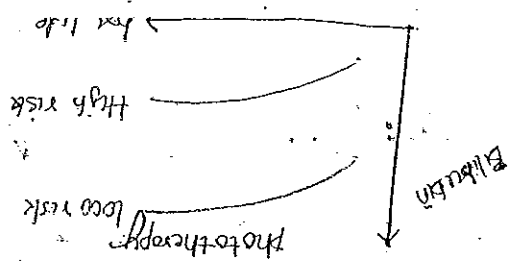
Start PPV (+ve pressure ventilation)

Intra tracheal suction (in obstruction)

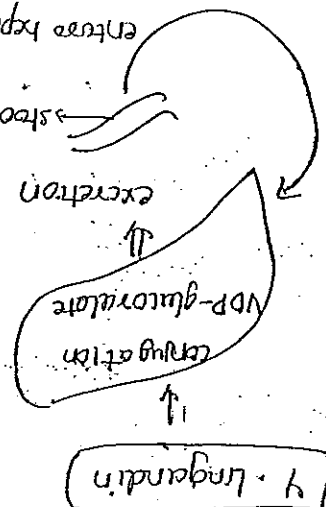
NEONATAL JAUNDICE



⇒ Unconjugated, unbound Bilirubin crosses the blood brain barrier
 ⇒ s. Bilirubin = 3.5 to 5.5 mg/dl
 ⇒ term, healthy i.e. no risk factor does not go for kernicterus
 ⇒ even at 25 mg/dl bilirubin
 ⇒ Preterm, sick, & risk factors go to kernicterus early

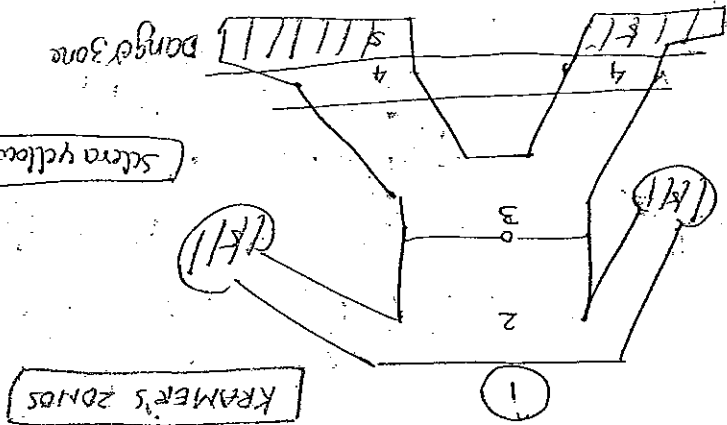


Bilirubin is taken up by the liver by a

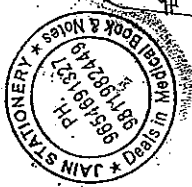


→ Physiological Jaundice

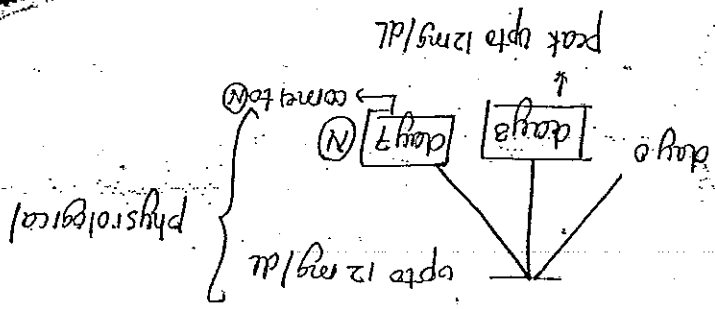
- Hemolysis of it is more
- Rapid turnover RBCs
- Y-Ligandine deficient
- UDP glucuronyl deficient
- Excretion def
- enterohepatic circulation def



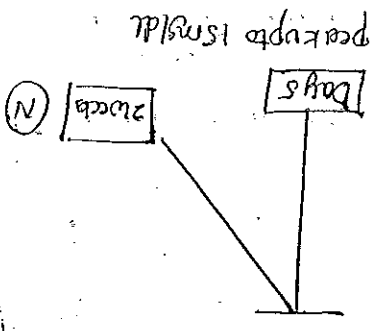
1 = 5
2 = 10
3 = 12
4 = 15
5 = >15 mg/dL



Jaundice in a term baby



Preterm baby



Pathological Jaundice: [Jaundice $\geq$ in 24 hrs] * $\Rightarrow$ severe haemolysis $\Rightarrow$ exchange transfusion

- Haemolysis
- Incompatibility Rh / ABO $\rightarrow$ [o mother, AB baby]
- Rbc memb defect
- Rbc enzyme defect $\rightarrow$ 46 PD Def

Pyruvate kinase Def

(ques) Mother Rh-ve, previous abortions
Cord Blood $\rightarrow$ (Hb 18-20 gm/L)

check: Rh status of baby Rh+ve
Hb (10 gm/dL)
Bilirubin $\geq$ 5 mg/dL
peripheral smear for haemolysis (+ve)
DCT Direct comb test (+ve)
Indication for exchange transfusion at Birth





=> Reaching 20 mg/dl bilirubin

Pre-entrant beyond 1) 7 days in term
2) 14 days in preterm

③ Hypokalaemia

② Hypocalcaemia (due to atax bind to Ca)

(due to high citrat)

complication: ① Metabolic alkalosis
② Mammie blood transfusion (complication like q)

so it a a Mammie blood transfusion
before Rebound after Rebound

so again transfuse the blood

Rebound Bilirubin

Bilirubin from tissues comes to blood

but after the sometime

only in blood

* it reduces the bilirubin by 85%

* fresh whole blood (to avoid Hyperkalemia)

$$80 \text{ ml/kg} \times 2 = 160 \text{ ml/kg}$$

$$\text{Blood of term baby} = 80 \text{ ml/kg}$$

* Double vol exchange transfusion

phototherapy

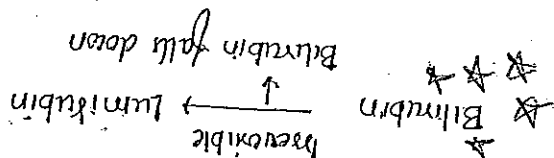
* 425-475 nm

* LUX 6-12 microwatt/cm²/mm
(unit of light exposure)

lux should be less than 6 @ change the light

* skin pigmentation does not effect efficacy of the phototherapy

* Principle: structural isomerization



Bilirubin falls down

(2) Photo isomerization

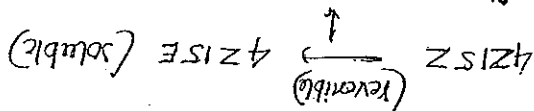


Photo oxidation (minor)

⇒ baby should be kept 40cm away

→ complication

* Fever, dehydration (so feed frequently)

* Hypocalcaemia (mecha Not known)

* Diarrhoea (Bile pigments in the Gut)

* ~~eyes~~ liver & genital

damage to retina

Mutation in gonads

* phototherapy C/I in conjugated jaundice bcos cause bronze baby synd: skin & urine



2 imp Pathologic Jaundice

Breast Milk Jaundice

• It occurs at day 14 of life
• pigment in the breast milk
• that inhibit conjugation

• Day 14 (20-30 mg/dl)

• cause kernicterus

• so temporarily interrupt

the breast milk for

48-72 hrs [treatment]

• Breast for 4-6 weeks and

slowly goes off

It is very difficult to see

Breast feeding Jaundice

• Occurs in day 3 of life

• occurs in primigravida

* (Shavation) (not giving milk)

↓

stimulate enterohepatic circulation

↓

He loses lot for 20%

↓

Jaundice occurs

↓

[treatment] ensure breast feeds

slowly goes off

It is very difficult to see

(quet) have about jaundice in neonate

(Am) can be seen after ventous delivery / forceps delivery

↓

they cause hematoma

↓

Hb come out

↓

cause jaundice

(quet) Meconium clay coloured stools and jaundice

⇒ this condition is called neonatal cholestasis

↓

Direct bilirubin $2\text{mg/dl (or)} >15\%$

↗

total bilirubin

extrahepatic biliary atresia : kasai's operation

done in 8 wks of life (otherwise 50% mortality)



Kasai's operation failed (or) late beyond 6 weeks

Liver transplantation is to be done

Medical, cause of Jaundice cholestasis

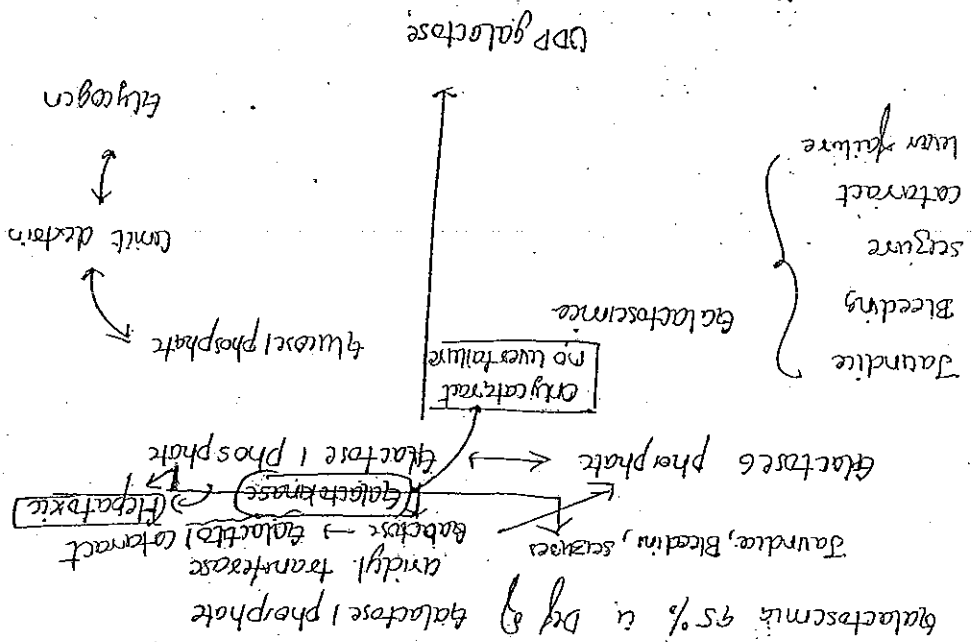
① N. Hepatitis

② sepsis

③ galactosemia [can cause Jaundice]

④ AI AT dy

⑤ N. hemochromatosis



Glycogen storage disorder - no liver failure

(Glycogen) can occur in stage 4

hereditary fructose intolerance

due to Aldolase B deficiency - leads to

Jaundice

fructose 1 phosphate (hepatotoxic)

fructose loading test $\Rightarrow$ Baby is given glucose

fructose kinase $\Rightarrow$ asymptomatic; as a mild def

$\rightarrow$ Benign fructose uria

Metabolic and surgical cause of cholestasis

* ~~not~~ Dying from medical ex

(*) G6P is 10% higher in ex

(2) * Vitamin: EHA Δ E cond sign

A cord like parallel to ^{structure} portal vein

IHR dilation induces UDP glucosyl transferase

(3) * HIDA nuclear scan (byscan give phenobarbitone)

3 day before
in biopsy where the HIDA dye is not seen in the gut

Delay image dye not seen in gut

* But test: liver biopsy

will show dilation & proliferation of IHRD

(5) Gold standard: Peroperation (Intraoperative Bile Duct)

choolangiography

of ducts, normal or abnormal
in or the liver filled with dye and shrink it coming out



↑
Atlagilla synd: & cause, cholethraesi

we have peripheral Pulm stenosi

* matation in Jagged - I

ques) Omious sign in a 10 day old child
(Am) conjugated hyperbilirubinaemia

⇒ Bilis eye u @ in 1st 4-6 wks of life

Alima's) Hepatocytes & PAS +ve Red coloured granules
(Am) Alpha 1 Antitrypsin deficiency (AR)

⇒ Russian blue stain used in haemochromatosis

↓
cause fibrosis

↓

Iron

⇒ CAIV: inclusion bodies; owl's eye

Cytomegalovirus

→ INTRAUTERINE TORCH INFECTIONS →

O-HBV, HIV, Varicella; syphilis

* They are transmitted during labour

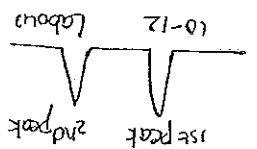
↓

So no symptoms

* Rubella - 2 peaks of transmission

1 peak = 10-12 wks (congenital rubella synd)

2nd peak = labour



mother gets rubella (after 16 wks) & fetus to body > 1%



* Symptom:

SGA; failure thrive

Anemia; thrombocytopenia

Hepatosplenomegaly

Rash; cholelithiasis

* How to test TORCH

Diagnosis:

IgM is diagnostic of TORCH

IgG (cross placenta) → 6-9 months

persistent

HIV:

Diagnosis:

Infant cannot make IgM @ IgA against HIV

Maternal IgG against HIV

Maternal IgG HIV can persist in the baby (>18 months)

Diagnose children less than 18 months

but: DNA PCR

other test: p24 assay

Culture HIV (but difficult)

⇒ Intracranial calcifications; fundus showing scar

chorioretinal scar

1st Ass: toxoplasmosis

CMV



- Periventricular calcification
 - Hydrocephalus ex vacuo [ventricles apparently enlarged]
 - Deafness (sensory-neural hearing loss) 7% infected
 - Seizures
- atrophy of the brain & ventricles apparently looking normal



CMV
 ⇒ Periventricular calcification in CT

DOC: Pyrimethamine + sulphadiazine
 Spermogram: the vertical transmission of Toxo
 ELISA: ser IgG is better than IgM

Diagnosis: IgM } But
 ISAGA

Intracranial calcification
 - Amn) Toxoplasmosis
 Central ventricular calcification in CT scan

in 1st trimester: and baby has hydrocephalus &
 pregnant 2 no H10 but has cervical lymph node enlargement
 Allm May 11
 (que), it
 calcification cause: Hydrocephalus
 seizures

toxoplasmosis: slight intracranial calcification
 more in choroid plexus
 caudate nucleus
 subependyma
 calcification cause: Hydrocephalus

Diagnosis:

Don't not establish a dx of CMV in a neonate

Urine and saline are best to culture CMV. PCR

Amn, IgG CMV antibodies in blood

PP6 PCR

treatment

(99% all babies are CMV - IgG +ve)

We don't treat infection

We treat disease

CMV disease: ① CMV chori

CMV retinitis

CMV, pneumonia

DOC: ~~the~~ Ganciclovir IV

Vangacyclovir ORAL

IV in severe disease

Foscarnet ORAL (for resistant)

CONGENITAL SYPHILIS

Severe mucocutaneous Rhinitis

Cracks at the chin

Bluetongue rhinitis

(snail)

Osteochondritis, Metaphysitis

Parositis

Glucosoma

Pseudoparasitosis of parent

normal limb; refuse to move



Painful Limbs

* Septic arthritis

* Scurvy

* Early syphilis

* hypokalaemia

* osteomyelitis

b) sensory neural hearing loss (SNHL)

c) interstitial keratitis

wide spaced notched incisors, and Hairy molar

(d)

② Saddle Nose, Frontal bossing, Oxyphoric brow

Hygumenecki sign (prominent sternoclavicular joint)

Rhago [cracks at chin & angle of mouth]

③ cutaneous joint: no pain at joints

Doc for syphilis: Always penicillin

Rubella synd

Microcephaly {
PDA
cataract
SNHL
Δ third eye
Rubella



PDA
cataract



⑤ Renal lesion

⑥ Autoimmune anemia

Late syphilis:

① Hutchinson's triad:

a) Hutchinson teeth



* m/c eye manifestation of Rubella

Eg: salt and pepper fundus

cataract

glaucoma

Microphthalmia

PDA

Heart: PDA

Peripheral Pulm stenosis

VSD

→ ASD is rare in Rubella

→ what is not seen in Rubella: Aortic stenosis

*** (True about Rubella except

Ans) Infection after 16 weeks results in major

congenital anomalies

↓
after 16 weeks the risk is only 1%

(Que) Baby has a Rash & hypoplastic limb

which infection to mother at pregnancy

Ans) Chicken Pox

& congenital Varicella embryopathy

involves brain → causes: cortical N atrophy, optic atrophy
Skin → cause: rash

Lumbosacral plexus → hypoplasia of limb

Aplasia of a limb

Ques) If mother gets chicken pox before 5 days after 2

day around delivery the child will also have

chicken pox : indicated V-Z immunoglobulin to baby

chicken pox vaccines : 2 doses

6-8 weeks apart

8 in 72 hr after exposure to 120 hr

post exposure prophylaxis

Ques) Pregnant women come & report HBSAg +ve

Next step

HBSAg investigation

If it +ve

90% the baby will become carrier

portal hypertension (head of Portal HTN : Abatto splenomegaly Varices)

If HBSAg (-ve)

developed Anti HBe

only 10% chance to spread to baby

⇒ Prevention : HBSAg 2 in 12 hr to baby

HBV vaccine 2 in 24 hr to baby



• Cause of hypoglycaemia

a) limited stores, Reborn, 104R

b) stress: sps

c) Polycythemia: RBC contain sugar: hypopituitarism

d) Galactosemia: Galactose 1 po4 [Hepatotoxic]

e) hyperinsulinism: Mother Placenta Fetus

↑ uncontrolled

↓ glucose

↑ hyper

↓ B cell hyperplasia

⇒ Insulinoma; Nesidioblastosis

↓ Pcell secreting

• Beckwith-Wiedemann: Fetal hypotrophy of limbs

Macroglossia

Risk of Wilms

hypoglycaemia

Ques) Baby born to diabetic Mother: after some hrs

birth became lethargic & his blood sugar was

30gm/l

symptomatic

↓

10% dextrose bolus

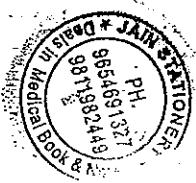
↓

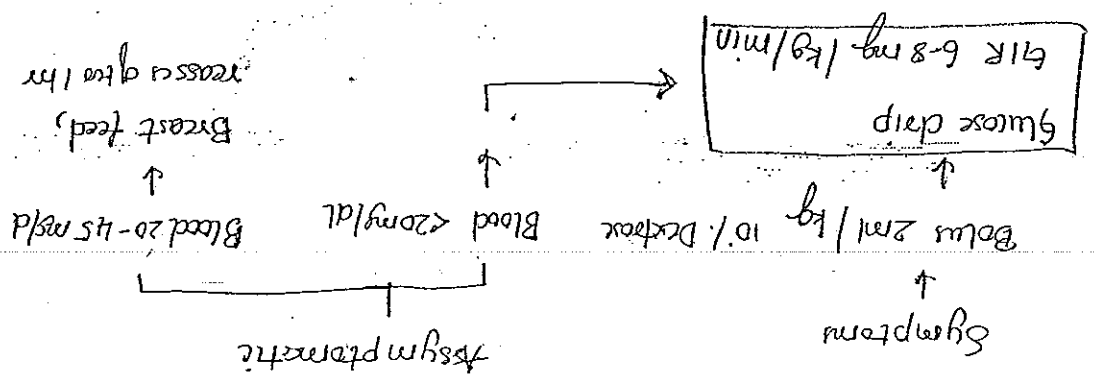
(Next) IV glucose drip

AllIMS NICU Protocol 2014

↓ Asymptomatic hypoglycaemia

Summative





If the drip stopped we use a emergency drug

Glucose → Glycogenolysis
IM
↓
Glucagon
↓
Liver. Needed

⇒ glucose cannot act in galactosemia

↓
glucose pol accumulation
↓
Hepatotoxic

Reassess hypoglycemia: $< 12 \text{ mg/kg/min}$ infusion rate

How to ↑ infusion rate

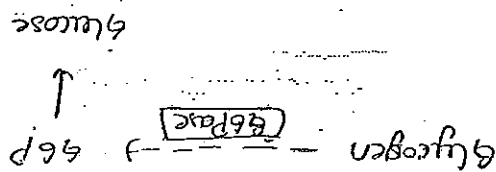
more fluid
↓
Conc can be red
Peripheral vein: 12.5%
↓
but can cause overload
↓
central preferred vein
⇒ 11.5% Dextrose given through peripheral vein

glut) 1yr child: has hypoglycemia } No Dextrose
Hepatoemegaly }

But no hypoglycemia does not respond to glucose
↓
Dextrose: 4SD - I Von Gierkes



Deficiency of enzyme glucose 6 phosphatase



→ CALCIUM →

• (N) S. cal: 9-11 mg/dL

• Neonatal hypocalcaemia: $S < 7 \text{ mg/dL}$

• 50% of cal is bound to diff protein

• Best index of cal is Ionized cal = $< 4 \text{ mg/dL (OR)} < 1 \text{ mmol/Lit}$

• INFANTS: tetany rare in infants

but seizures, tremors, jitteriness

↓
can be stopped on passive restraint

mild hypocalcaemic

2 types

Early: 1st 72 hrs
late: after 72 hrs

late

phosphate rich milk: cal

Early

Prematurity

Infant of diabetic mother

Asphyxia

hypophylactic &
and monitoring

Good cal supplement: Ca/P ratio > 2



100 ml	cow	Mother
Ca	118 mg	34 mg
PO ₄	100 mg	15 mg

⇒ Breast milk protects from

• NEC

• allergy, eczema, asthma

• late onset hypocalcaemia

• Pneumonia

• Beta virus diarrhoea

• Infection

• IgA - RSV - Bronchitis

⇒ AAP endorses which vit should be given to all infants

Vit D

→ Vit C causes scurvy which is present in breast milk

⇒ Milk is deficient in: Vit C & Iron

⇒ Breast milk has sufficient Vit C & Iron for 1st 6 months of life

So use complementary feed

⇒ PDA vit D infant 400 IU/day

⇒ Breast milk 25 IU/L

1 ml drops = 400 IU

⇒ m/c of hypocalcaemic Bezura: Vit D

Infants of Diabetic Mothers

• Preterm, stillborn

• Macrosomia



linear growth for fetus: insulin

hyperinsulinism $\rightarrow$ macrosomia

IUGR $\rightarrow$ if mother have advanced DM $\downarrow$

class F / class R $\rightarrow$ Diabetes in fundus $\downarrow$

placental vasodilatation $\downarrow$

placental insufficiency

RDS [Lis ratio >3.5]

22% hypocalcaemia, hypogly

19% neonatal jaundice

At risk of polycthemia $\rightarrow$ cause renal vein thrombosis (RV)

[Allias] (yes) complication that can not be seen in infant of Diabetic mother

hypoglycaemia

anomalie

m/c anomaly: CVS (8.5%) [VSD] + TGA; HCLM/asymmetrical sept

Neural tube defects (5%) $\downarrow$ hypertrophy

easy left adon/pseudo oblique
DOC: Propionol

sacral agenesis

m/spectic anomaly: sacral agenesis / caudal regression

56/100 mother + DM



⇒ Muscular Disease differentiating left colon

by biopsy

Encephalocele

Folic Acid intake from 1 month before pregnancy

of the pregnancy

FA deficiency cause

Neural tube defects

baby born with clefting

Indication for Do Not Resuscitate

2nd indication: placental synd

Early anomaly seen by usg in

10-11 weeks

neurogenic bladder (severe constipation)

paraparesis/paraplegia

myelomeningocele

site → lumbosacral

Encephalocele: exomphalic (m/c)

herniation of brain tissue

MRI brain

⇒ Povidine Iodine { Neurotoxic Saline → Non neurotoxic

⇒ Reputed myelomeningocele

Bat: Blood culture (they can have bacteraemia)

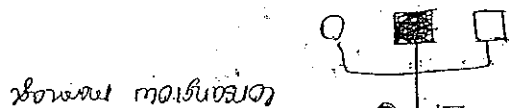
⇒ Risk of recurrence is by 3.5% for every pregnancy to prevent recurrence dose of FA is 10 times the (N)

dose 4mg → 4g (reduce risk by 75%)





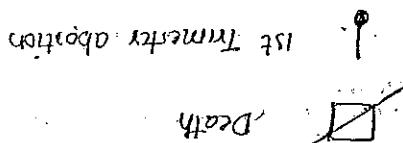
divorced



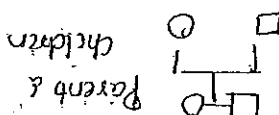
consanguineous marriage



Rebared
(case under evaluation)



Autosomal Recessive
carrier
→ only female carrier
affected
Death



Parent & children

Male
female
lacking

→ GENETICS →

(MSAFP)

Maternal serum α-fetoprotein

Acetylcholinesterase & AFP

AFP (Marker of neural tube defect in amniotic fluid)

3 child → 2%

2 child → 10%

1 child → 3-5%



Hereditary spastic paraplegia

Hypercholesterolemia

VH3 → WVD, Huntington chorea

T - Tuberculous sclerosis

N - NF

A - Adult PKD, Atrophoplasia

N - Noonan synd → looks like Turner but the karyotype is (N)

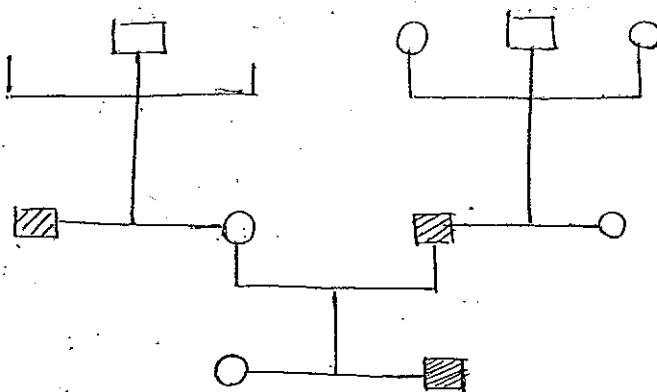
I - Intermittent Rophyma

M - Marfan synd

O - Osteogenesis Imperfecta

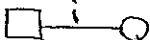
D - Dystrophy myotonic → involves distal muscles more

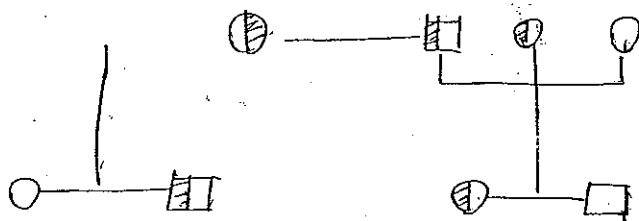
Autosomal Dominant



[O]

adoption



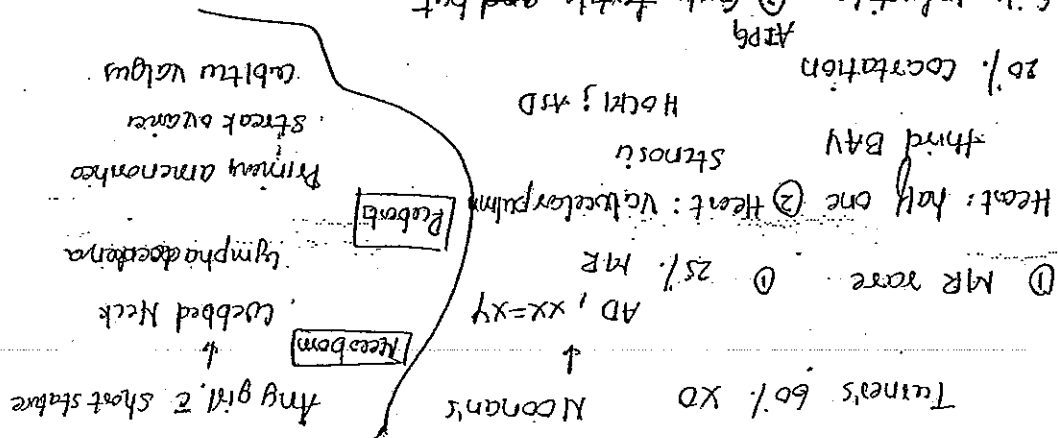


$AA \times Aa$
 $Aa \times Aa$
 $Aa \times Aa = 25\%$

the chance of the child not getting affected
 Both parent are affected $\rightarrow$ Achondroplasia

④ Nothing factor deficiency

Boys $\rightarrow$ infertile
 delayed puberty
 ③ Girls infertile but



(31)



$$= \frac{1}{2500}$$

$$\frac{1}{25} \times \frac{1}{25} \times \frac{1}{4}$$

Male Female child

⇒ In UK & Europe: 1/25 carrier of cystic fibrosis

Adenosine deaminase deficiency

↓

XLR severe combined Immuno deficiency

[7/11/15] (qua) gene therapy not time successful

⑤ X-linked recessive Fibrosis (only seen in boys)

④ GBD II pompe [Heart]

③ MPS VI maroteux lang

② Hunter MPS-I

① Gaucher's synd: [B-glucocerebrosidase] CER-enzyme

Enzyme replacement therapy

clouded cornea clear cornea

Autosomal Recessive X-linked recessive

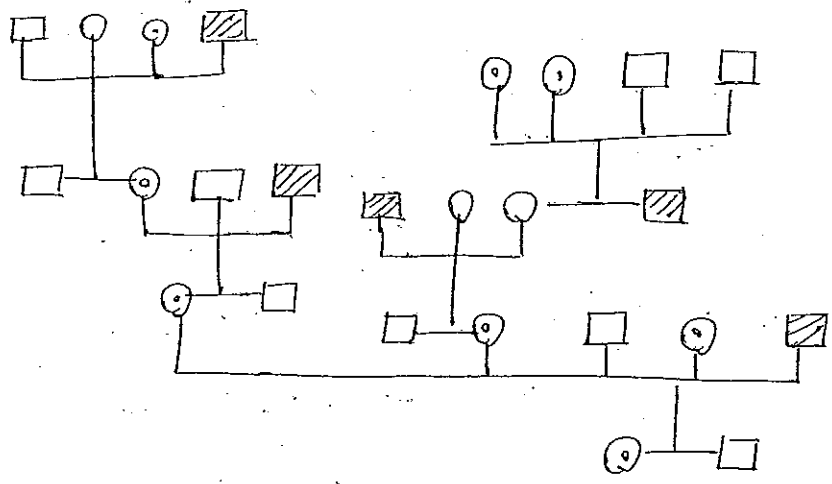
MPS-I MPS-II

Hunter Hunter

- Duchenne muscular dystrophy
- Hemophilia A & B
- 66 PD of
- Wiskott - Aldrich synd
- Leish - Nohan synd
- Chronic granulomatous disease



→ X linked Recessive



⇒

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

die $\text{AAC} \text{AAC} \text{AAC} \text{AAC} \text{AAC}$
 $\frac{2}{3}$

chance child can have CF?

British lady
 my brother
 died of CF

⇒ British guy



- Duchenne Muscular dystrophy →
 - * m/c hereditary neuromuscular disease
 - ↓
expressed in skeletal muscle
 - * Have pseudo hypertrophy of calf muscles
 - * Proximal muscle weakness
 - * Gowers' sign ↓
 - * all of them die in teenage life
 - * Recurrent chest infection
 - * Cardomyopathy
 - * 1/3 Mental retardation
 - * 1/3 Duchenne can also occur due to New mutation
 - * LK level in 10,000 (very high)
 - * Becker's muscular dystrophy: Milder variant
 - present late
- Wiskott Aldrich's synd →
 - * Xlinked Recessive
 - * triad of Eczema, Thrombocytopenia, Immodeficiency
- Chronic granulomatous disease →
 - * XLR
 - * NADPH oxidase deficiency
 - * Immodeficiency
 - * Test: NBT dye test

→ Leach Hyhan synd →

* self mutilation * Mental Retardation

* XL recessive * diphygia

* start 2 Nse; * ophthalmone spasms

* choroidoid movement
palate
fingers

* Urine detect: HPPRAE del

hypoxanthine guanine phospha

⇒ AIPg → investigation: serum uric acid level $> 6.5 \text{ mg/dL}$

Alims (que) M child 2 Fanconi synd 2 nephrocalcinosis

has a variant of dent disease: All are true except

↓
(the) same in father

(XLR) (XLR)

hypercalcaemia: 24 hr uric acid
↓
24mg/kg
↓
nephrocalcinosis
nephrocalcinosis

Proteinuria 1: 1000 B2 microglobulinuria

Losses

Proximal = Tubular =

Na⁺ → 6%
H₂O₃ → >85%

defect
PO₄ → >88%

loss leads to
aminoacids
glucose
↓ 100%

Rickett





⇒ low synd

* AI 06

+ XLR

* OCL 1 gene is mutated

* Oculo-cerebro-renal disease → Fanconi synd
↓
cataract
glaucoma
Fanconi's synd
↓
retinopathy
HFR

* Treatment: Na and PO₄ supplementation

⇒ Fanconi's synd can be of many cause

(1) genetic - XLR Dent's

XLR lowcs

AR - cystinosis

(2) Metabolic: galactosemia

HFI

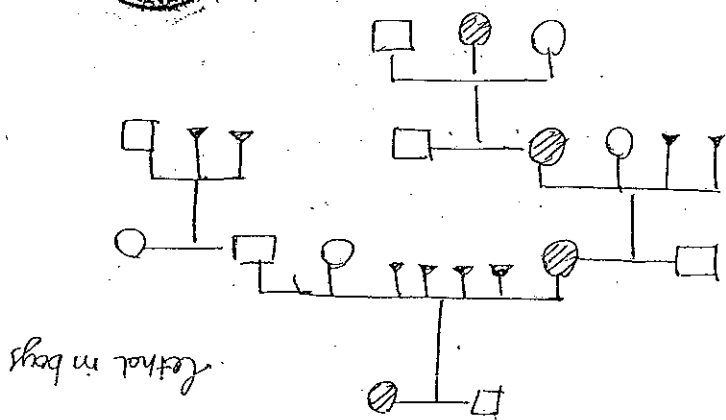
Net DNB
Thyroidism [Ex: Mithunone]

(3) Acquired: tetracycline exposed to b

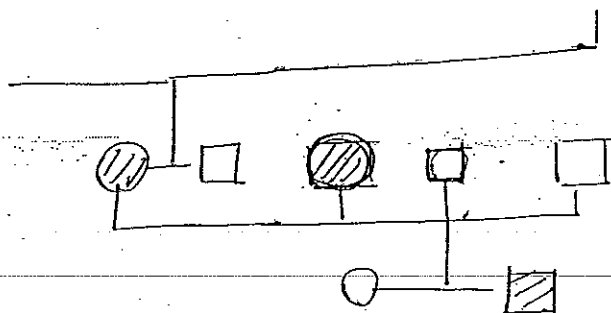
⇒ X-linked Dominant

Father x y
↓
to son
to daughter

Affected father
↓
affect to all his daughter
none of his sons



- * X linked dominant : severe in Boys
- * Familial hypophosphatemic rickets
- * Uræa cycle defect due to OTC def
- * Incontinentia pigmenti are only seen in girls
- * Retts synd



→ pervasive developmental disorder

• Autism

• Asperger

• Rett's

→ Autism: m/c in boys

less than 3 yrs

→ Autism what is not in Autism: (And) Visual impairment

→ Asperger: m/c in boys

good IQ

little profluser (called)

→ Rett's syndrome: m/c in girls

XLD

McCP2 gene mutation

(N) till 6-18 months

↓ in head growth

Macrocephaly; Mental retardation

stereotypical hand movement

Repetitive behaviour

Allms: Not seen in Rett's: (And) Macrocephaly

↓

Post mortem brain biopsy
↓
dendritic morphology in cortical
sheets abn pyramidal cell



⇒ In Rett's the girl die due to Arrhythmia

Most common urea cycle disorder is

Alam 2015

Ornithine transcarbamoylase : urine orotic acid

urine urea

Detection :
Evida that synd

SP deletion : cat cry synd

Partial trisomy 22 : cat's eye synd

Vertical pupil

supraventricular AS, hypertension
peripheral PS, Ectm in face

Prader-willy synd (15q11-13-)

or Georges synd (22q11-)

absent thymus, absent parathyroid

hypoplasia of ~~gale~~ thyroidal pouch

⇒ Micro deletions are Asc'd by a FISH

cross locations ~~↔~~ heterozygous

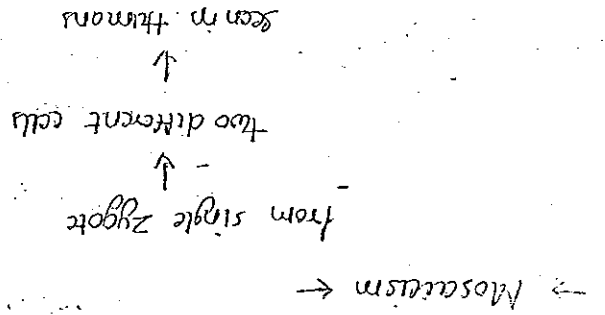
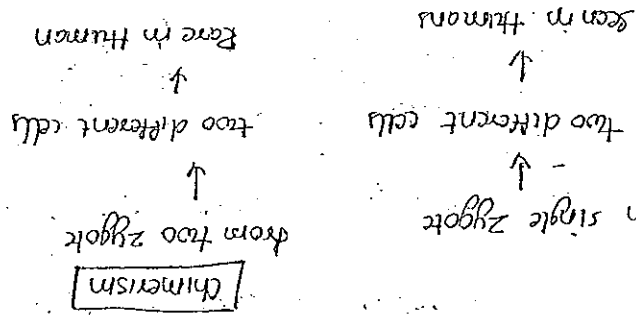
TRANSLOCATION →

balanced ~~↔~~ unbalanced

total genetic material

remains same





D 13, 14, 15
G
D-6
G-4

Some cells are 46 and 47

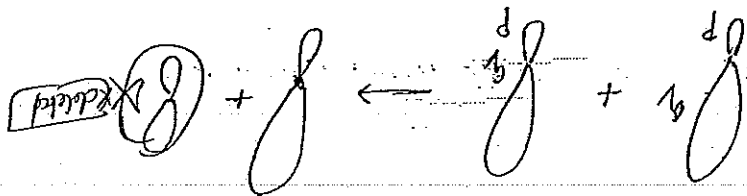
* 1-2% Mosaic downs

* 3-4% Robertsonian translocation

* 95% Trisomy - 21 [maternal meiotic non dysfunction] AIPG 200

Down synd →

Robertsonian Translocation



Larsen's synd

Cytogenetics: 6b/1.45X0

15% mosaic xx/xo

10% Isochromosome Xq or Xp

10% 46 X deletion

5% Mosaic xo/xy

Isochromosome: one arm lost and

duplication of the other arm

Klinefelter's 80% XXY

b) they are also mosaic: XXY/XXY

Somatic mosaicism: it will not be transmitted

germline, mosaicism: transmitted

cannot be used by blood DNA

Question

Anticipation: for every successive generation

the disease will present early and is more

severe.

only occurs in trinucleotide repeat disorder

-tg: fragile X CAG

F. Ataxia GAA

Hypotonic Dystrophy - CAG, CCG

Spinobulbar, muscular atrophy - CAG

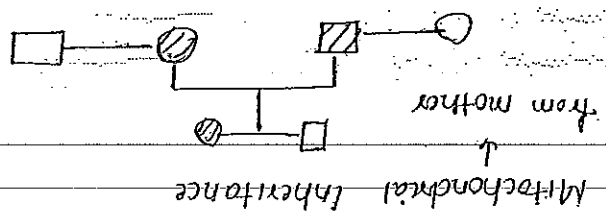
Huntington's CAG

Spinocerebellar Ataxia - CAG/CTG





- * Recusen syndrome : Pancreatic insufficiency + Remyotopia
- * NARP
- * Kearns - Sayre synd (KSS)
- * Leigh / disase
- * Leber hereditary optic neuropathy (LHON)
- * and lactic acidosis (MLAS)
- * Mitochondrial encephalopathy, stroke like episodes
- * Myoclonic epilepsy and ragged fibers (MERRF)



22 JULY 2017

NADA's criteria

MAJOR $\rightarrow$ for congenital heart disease

④ loud systolic murmur

② Diastolic Murmur

③ Central cyanosis

④ CHF

MAJOR

① systolic murmur

② Abn S2

③ Abn ECG

④ Abn X ray

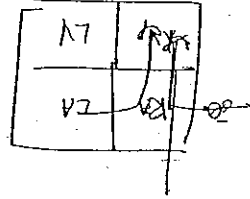
⑤ Abn BP MAJOR MEET

$\frac{1}{2}P_2 \rightarrow$ split (varies w resp)

$A_2 - P_2$ (expre)

$A_2 - P_2$ (inspre)

ASD : $A_2 - P_2$ } wide & fixed
 $A_2 - P_2$ } (inspre = cprc)



ASD = RV Vol overload



Physiological } systolic & diastolic flow murmur

VSD = wide & variable

A_2 ----- P_2 (exp1)

A_2 ----- P_2 (inspire)

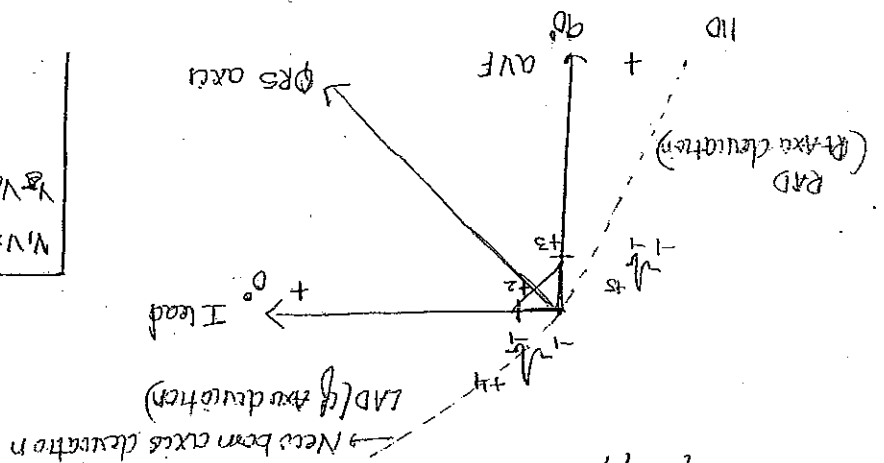
* All New born are born with Rt ventriculopathy (RVH)
Rt Ant D (RAD)

* Axis like adult > 1 month life

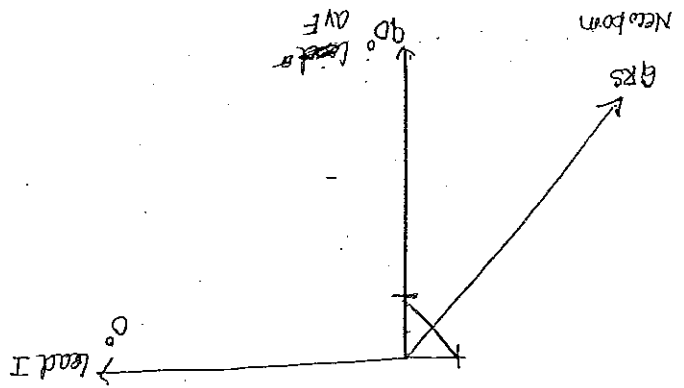
* T waves - V1, V3R, V4R

① T waves are +ve

they appear UP + < 48 hr life



$V_1 V_2 = \text{Rt side leads}$
 $V_5 V_6 = \text{Lt}$



② After 1st 48 hrs they become -ve
they are -ve still adolescence



③ In adult life they again become +ve

Incidence of CHD:

Incidence = 0.8 to 1%

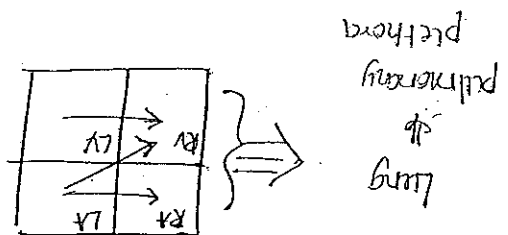
Recurrence = 2-6%

Synd: ① Down's synd = 40-50% of children = three CHD = M/L² death

Endocardial cushion / AV canal / O primum ASD

→ severe type of ASD

Endocardial cushion (left to R shunt)



② Turners = half to one third BAV; 20% coarctation

③ Noonan's - AD; XX = XY; Valvular pulm stenosis

HOCM, ASD

④ Congenital Rubella synd → PDA; peripheral pulm stenosis, VSD

Imp → Peripheral pulm stenosis

seen in: ① Alagille synd
Rubella

Williams





AIIMS Nov 2010: Phocomelia

Thalocamide

Maternal: Lithium, Causu:

fbstch's anomaly

2. Mumps: endocardial fibroelastosis /

severe LV obs hibern

3. Pentallamine: utis laxa

4. Warfarin: chondrodysplasia punctata

Abn. Nose

in the place we use Heparin (in 1st trimester)

from 2nd trimester again use Warfarin

5. SLE: complete heart block → pacemaker

Ant Ro Antibodies

there are more found in SJOGEREN'S synd

SJOGEREN'S >>> SLE

FETAL CIRCULATION:

Ductus venosus: connect umbilical vein to inf vena cava

PO₂: 30-35 mmHg

PO₂: 28-30 mmHg

umbilical vein 100% blood

50% → liver

70% → ductus venosus



→ Classifi

Physiological closure : 10-15 hrs of life
Anatomical closure : 10-21 days of life

→ Ductus Arteriosus Closure

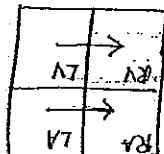
First breath : Umbilical Artery → L Vein →
↓
Ductus venosus → Ductus arteriosus
sequence of closure

Inf vena cava blood → RV
↓
only min blood to
upper body & brain → descending
aorta
↓
lower part body
lung from RV



* Present in CHF 6-10 weeks life

- * Excessive sweating = CHF $\rightarrow$ tachycardia $\rightarrow$ sympathetically $\rightarrow$ cardiac output $\rightarrow$ feeding dephoradi = sick rest sick cycle
- * Recurrent Pneumonia
- * Failure to thrive
- * PBF $\uparrow$: pulmo plethora
- * ASD, VSD, PDA



shunts $\rightarrow$ (LH) $\rightarrow$ (RH)

ACMNIOTIC

CYANOTIC HEART DEFECTS

Pulm Blood Flow

(increased)

(decreased)

PLETHORA

• Pulm Arteriosclerosis

↓ tx of cyanosis & plethora

• TGA + VSD

• TAPVC (Total Anomalous)

Heart → Boxed shaped
↑ Balloon shaped

• Ebstein

(Massive cardiomegaly)

↑ failure's physiology

• TGF

• DORV + PS

Double outlet RVo

+ Pulm stenosis

• Single vent + PS

• TGA + VSD + PS

• LVH → tri cuspid

• Atrial





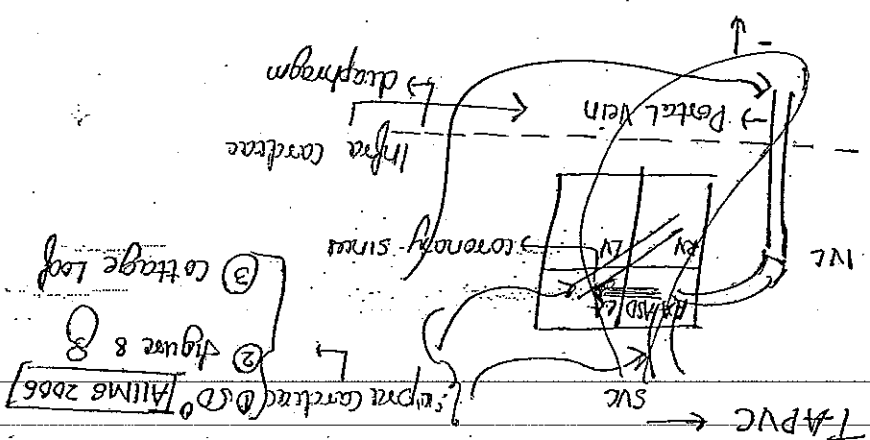
* Atrial is connected condition by PGE1 $\rightarrow$ basically is a good
 text in all other conditions
 $\downarrow$
 More bleed to lungs
 $\downarrow$
 $\uparrow$ Pulm pressure
 $\downarrow$
 $\uparrow$ Pulm hypertension

Only pediatric cardiac surgery emergency

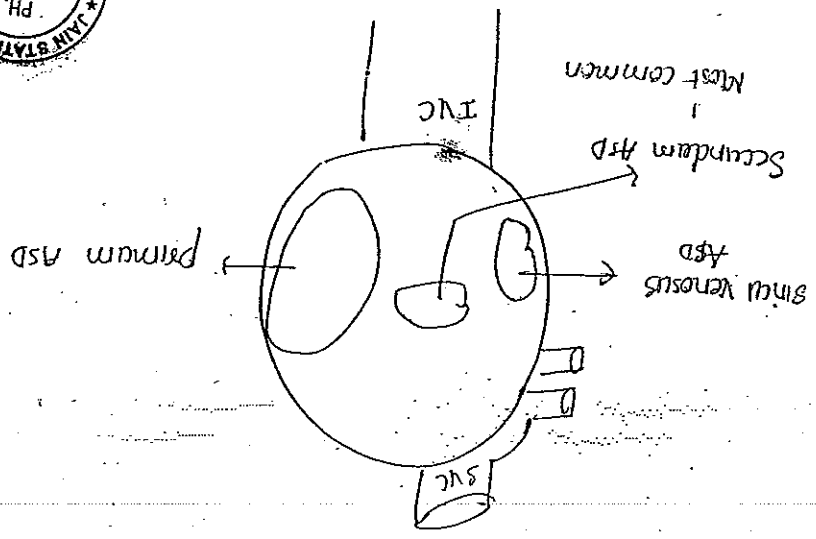
* White lungs / glass ground / kerry B line
 $\uparrow$
 Portal vein at the level of diaphragm
 ok pulm venous hypertension of the

* Most severe cyanose = Present at the birth itself

bleed to pure $\downarrow$ = plethoria



→ ATRIAL SEPTAL DEFECT →



ASD syndromes

1. Down's : endocardial cushion synd

AV canal

Primum ASD

2. Holt ORAM synd : familial

→ distally place thumb
↓ ASD (secundum)
↑ ASD
↓ Hand treat
ASD secundum synd

→ Tripharyngeal thumb
② Picrothyry
→ Rudimentary thumb
↓ TBX mutation
Atrial fibrillation

Surgery : Polysistation of Index finger factor
like thumb
PDA is not seen

TAR synd : Platelet count

Bone marrow : (Fanconi anemia)

Echocardiography [Holt ORAM synd]



* Absent Radium $\Rightarrow$ Holt's syndrome

* Karyotyping - has no role

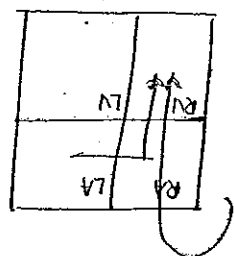
③ Lutenbacher: ASD + MS

④ Ellis Van Creveld: ASD + polydactyly

$\rightarrow$ ASD secundum $\leftarrow$

+ child = asymptomatic

= wide fixed S₂



* ECG: Rt Axis deviation (RAD)

* Adult life complications

• Rt Ventricular failure

• At risk arrhythmias: Atrial Fibrillation

$\downarrow$

thrombus

$\downarrow$

stroke

$\downarrow$

CVA

Reversal [R > L] $\therefore$ Eisenmenger syndrome

$\downarrow$

Child develops cyanosis & clubbing



* Natural course

• $< 3mm$ = all close

• $28mm$ = unlikely to close

we go for surgery

surgery: • All symptomatic

• Asymptomatic $\rightarrow \boxed{Qp/Qs > 2}$

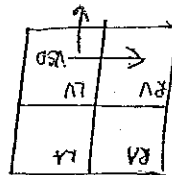
Q = blood flow

P = pulm

S = systemic

• this is a simple sx

VSD



thraspid infective

vegetation

→ ASD Primum ←

* Downs

+ Also. has mitral regurgitation

+ Wide fixed S_2 + S_1 == S_2 (par systolic mormour)

apex $\rightarrow$ axilla and back

* Conduction defect $\rightarrow$ ECG $\rightarrow$ LAD $\Rightarrow$ cause LAD

* RSR' = Defect of conduction in the RV

MR also cause LAD



MCA: small VSD: M/c & IE
Small ASD: M/c secundum



* Loud marmur [small hole VSD]
 ↓
 at loud pan systolic
 at low sternal border
 normal

Maldie de Rogers

* Small ejection systolic murmur

* No marmur

* Compare $\frac{2}{3}$ ratio of aorta & $\frac{1}{3}$
 → Small VSD
 → Medium VSD
 → Large VSD
 → 6-10 weeks of life

* Size: small
 Medium
 Large

* 70% perimembranous

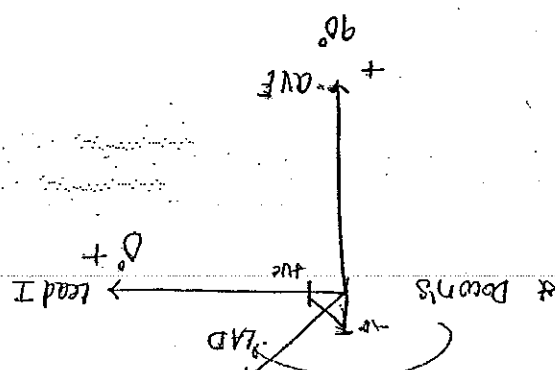
30% VSD

* M/L of CHD

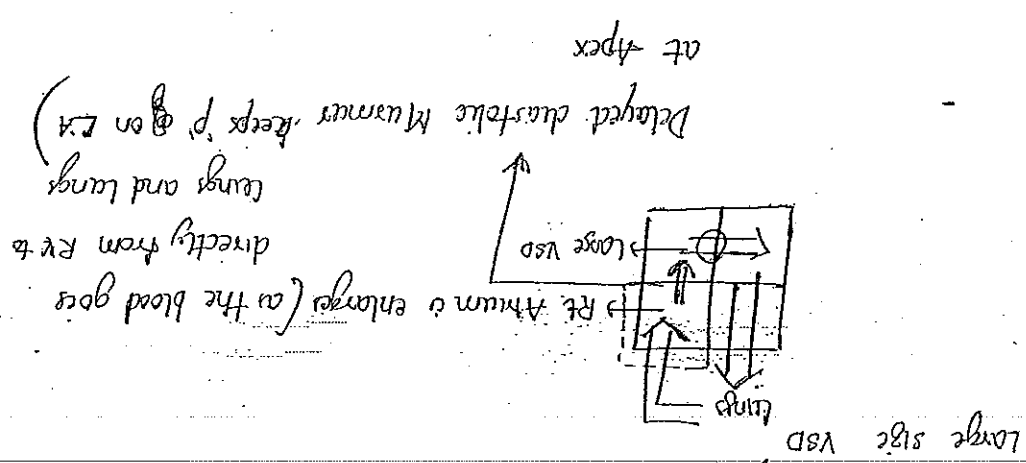
→ Ventricular septal defect →

* Early surgery

* CHF 6-10 weeks of life



* Which chamber of the heart enlarges 1st in ~~small~~ medium to large size VSD



* Natural course

- 80% Perimembranous
- 50% Muscular VSD

• close by 4 yrs

• Indications of ex in c/o VSD

- Failure of Medical therapy
- $Q/P/Q_s > 2$, asymptomatic

- Swiss-cheese VSD (Multiple VSDs at the apex)
- Supracardiac VSD (VSD in ventricle outflow)

* Complications: IE, CHF, reversal;

• Large $L \rightarrow R$ shunt

PBF increase

Reversible pulmin microvascular changes (Pulm HTN)

RV 'p' test $\Rightarrow$ shunt is $R \rightarrow L$ (reversal - Eisenmenger synd)
significantly developed





→ Ductal Dependent Blood flow ←

- bc in both
- NO
- sildenafil [phosphodiesterase II inhibitor]
- Bosentan, Ambrisentan [endothelin antagonists]
- Iloprost [P412]
- Amiloridine

* Pulm HTN Rx (reversal = secondary Pulm HTN)

Heart & lung transplant

we should go to care

sx & c/I

Eisenmenger's synd = irreversible damage

Reversal at adult age

Small PDA

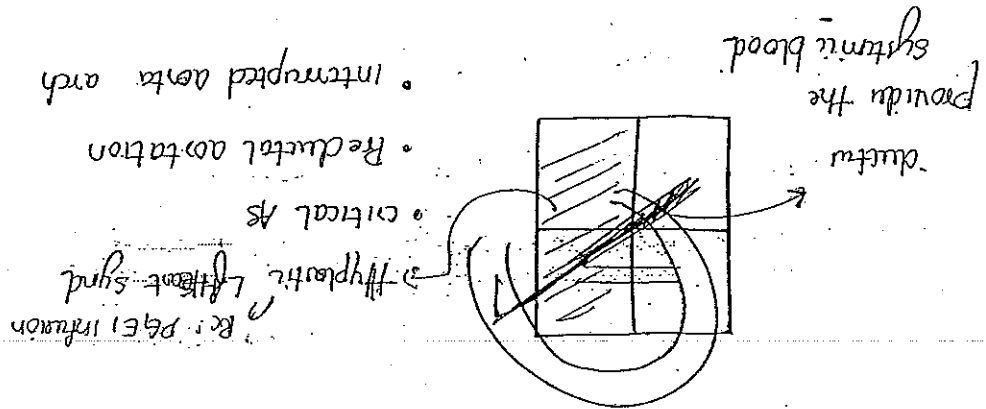
Large PDA: only reversed

than fingers

more in toe (more impure blood, go to toe)

* Differential clubbing & cyanosis

→ DUCTAL DEPENDENCE BLOOD FLOW ←



I. Systemic Blood flow dependence (L side heart Problem)

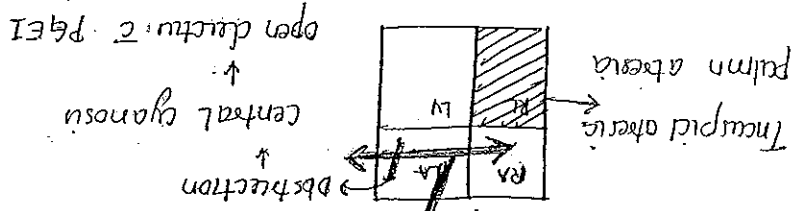
* shock (when ductus closed)

↓

then open the ductus by

Rx: PGE1 infusion

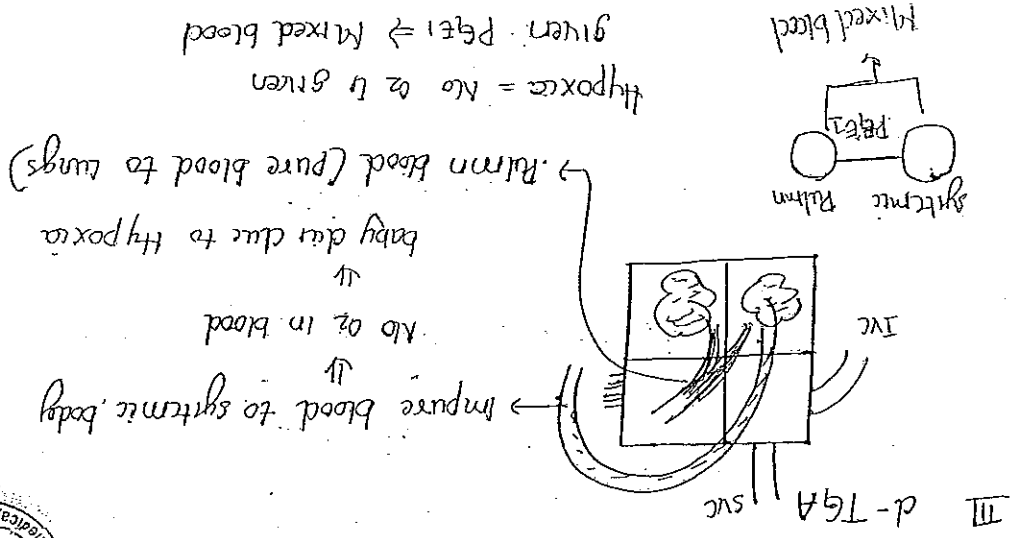
II. Pulmonary Blood flow (Rt heart)



central cyanosis

↓

open ductus: PGE1



III. d-TGA

IVC

SVC

Impure blood to systemic body

↓

No O2 in blood

↓

Pulm blood (pure blood to lungs)

↓

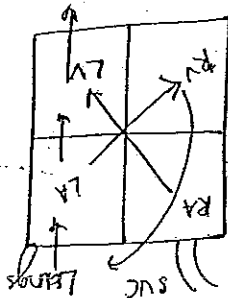
hypoxia = No O2 given

given: PGE1 ⇒ Mixed blood





Aorta = Rare blood to system



* Normal
* Associated IVC
* Abn are present
like spleen

→ L - TGA = corrected TGA

child is cyanotic at birth

* Worst TGA = TGA + intact septa > TGA + ASD
+ 50% of TGA has VDA [VDA is useful for mixing]

2 weeks of life

so switching should be done early in life

* Definitive surgery: Jettens atrial switch

* Emergency surgery: Rashkind's Balloon Atrial septostomy

* M/c cyanotic heart disease in infants is TGA

* Truncus Arteriosus → Ductal independent



Intact aortic ECG helpful in Ebstein's

Ventricular = ECG

* Pressure is like RA

* Box / Balloon shape heart (massive cardiomegaly)

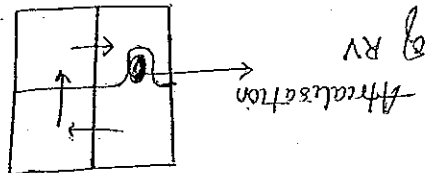
* Systolic, diastolic; Multiple heart sounds = Quadriple rhythm

* Maternal pre-eclampsia Lithium

* Cyanosis $\downarrow$ PBF

* down displacement of tricuspid into RV

They have mostly ischaemia
 Arrhythmia
 WPW synd
 SVT synd



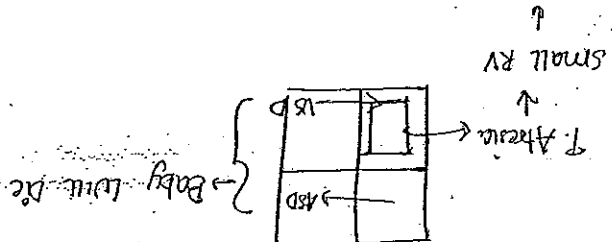
→ EBSTEIN'S ANOMALY →

* LV Hypertrophy; LAD

* PBF $\downarrow$

* Cyanotic [Cyanosis $\propto$ LVH = Tricuspid Atresia]

Hypoplastic RV $\Rightarrow$ RV blood flow $\downarrow$ ed = Aegemia



→ TRICUSPID ATRESIA →

→ ETIOLOGY OF PALLOD ←

- * M/c cyanotic heart disease
- * M/c cyanotic heart disease beyond infancy presentation
- * Infundibular Atrialms os
leads to RVH
- Aorta override the VSD
- * VSD is perimembranous VSD
- * becomes suprasystolic - R to L shunt
- * Example of cyanosis & oligemia
- * The pink to F becomes blue T of
- * Features of T of

• Cyanosis

• In Infancy : there is cyanotic spells
hypoxic

tet spells

↳ like to squat* (squatting older)

• He will have hypoxia : polycythemia

Erythropoietin
↓
↑
lead to Iron def

lead to renal vein thrombosis

Hb is red

* complication : In children < 2 yrs : thrombosis

> 2 yrs : haematogenous brain abscess
unexplained fever
↓
persistent hypoxia
↓
for child



* pathogenesis of cyanotic spells

3 Reason: ① Rt → Lt shunt increase

↓
1st reason At the time of spell ↓

② Hypoxia (P_{O2} falls ⇒ resp center +++)

↑
Resp center (stimulated)

↓

P_{O2}

2. Dynamic pulmo stenosis

3. Child is crying

treatment: oxygen

• IV NaHCO₃

• s/c Morphine = ↑ P_{O2}

• IV propranolol = (↓ dynamic pulmo stenosis)

• knee chest position (= ↑ systemic vascular resistance)

Drugs: IV ketamine

IV phenylephrine

Alims Nov 16: How do you identify the spell clinically

Any Murmur disappear

Breathless

More cyanosis

Surgery: Palliative shunt

opposite to aortic arch

④ Modified blalock taussig (subclavian artery to Pulm artery)

② Watson (ascending aorta → RPA)

③ Rott (descending aorta → LPA)



* After 3-6 months → definitive sx can be done

* TOF children have Rt. Aortic arch Atkins

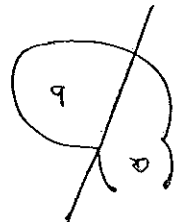
$\left. \begin{array}{l} 25\% \text{ TOF} \\ 50\% \text{ Transcath aortic} \end{array} \right\} \text{ both have Rt. Aortic arch}$

* Cardiomegaly $\left. \begin{array}{l} \text{CHD} \\ \text{Never seen in TOF} \end{array} \right\}$

Rt Heart Never falls b/w of septal defect

* Cardiomegaly = Cardio Thoracic Ratio > 0.6 infants > 0.55 older

$$\left(\frac{a+b}{c} \right) \left(\frac{a}{b} \right)$$



pericardium

the ratio is NOT changed if there is one sided cardiomegaly

MCQ ALCAPA → Bypass

Abnormal origin of the coronary artery

Anterolateral wall MI in ECG
 deep Q wave I & AVL

* on angiography → (Rt) coronary falls b/w (L) coronary

M/C ^{old} Medium sized Vasculitis ^{Thrombus}
 Kawasaki's disease = coronary artery aneurysm
 is from palm eating



kawasaki

* Fever > 5 days on 4 days

* change in extremities (eg: erythema, edema, desquamation)

in palms & soles
on 2nd & 3rd wk

* Bttx B/c conjunctivitis (not on 2 exudate)

* Polymorphous rash (not vesicular)

* ~~on~~ v/c cervical lymph nodes > 1.5cm
pathy

* change in lips & oral cavity

Tr: pharyngeal erythema

strawberry tongue

dry / fissured swollen lips

* vasculitis which is not seen in adult: kawasaki

sh/c vasculitis in adult: susacs synd

* treatment: immunoglobulin

kawasaki = rare in < 3 month infants

immunoglobulins from mother in pregn

* Prevent aneurysms in kawasaki = 0.14 Ig 2gm/kg

acute phase not > 4-6%

② Aspirin 100mg/kg/d x 2 weeks

③ 15% resistant to IV Ig - repeat &

- add steroids

④ 1-3% Recurrence



* coronary aneurysm

Small size = 50% resolve over next 2 years

Aspirine 3-5mg/kg/day

low dose aspirine: Antithrombotic dose

Medium - large = add warfarin + surgery

* Non Classic features: Arthralgia

Arthritis

urethritis

Thrombocytosis

Intractable: aseptic meningitis

Death: M/C death - Myocarditis

Henoch - Schonlein Purpura: (leucocytoclastic

(HSP) M/C vasculitis

(wbc invade into vessel wall) (dermu)

(4) uterine lesion - palpable purpura

(5) non thrombocytopenic palpable purpura

(6) Immune fluorescence shows IgA1 HSP

IgA: ~~all~~ galactosylate

Absent galactosylation of IgA leads to deposition in Mesangium glomerulus (kidney)



and more

* HSP.

• Skin Rash

• Pain abd

• Testicular pain

• Arthritis

• Kidneys

40-50% cases
Microscopic haematuria

Proteinuria

1-2% acute renal failure: crescentic GN

↑
proliferation of parietal epithelial

cell

Rc: IV Methyl prednisolone

→ RHEUMATIC FEVER ←

* Caused by B haemolytic streptococci

* M/frequent b/w 5-15 yrs
↑
types: 1, 3, 5, 6 & 18

* Latent period 3 weeks (after throat infection)

* Autoimmune phenomenon

↑
Molecular mimicry
Myocardial cells

* DDC: Anti-inflammatory: Aspirin^(w)

prednisolone

* 2015: Modified Jones criteria

Define risk areas

Echo cardiography





Bed rest
crystalline Penicillin G x 10 days
to eradicate the colonisation

12 wts
Prednisolone : severe carditu/CHF

Treatment : high dose Aspirin for 12 wts

* Prolonged PR interval, after accounting for age
* Feasibility (unless carditu is a major criteria)

* ESR ≥ 30 mm/h and CRP ≥ 3.0 mg/dl
* Fever $\geq 38^\circ\text{C}$

Minor * Monoarthralgia

* Polyarthralgia
* Subcutaneous nodules
* Monoarthritis (or) polyarthritis * Erythema marginatum

Major * subclinical cardiac
* chorea

Recurrent of RF : 2 major / 1 major + 2 minor / 3 minor
- **

$\Rightarrow$ 2 major / 1 major + 2 minor

② prevalence of RF > 1 per 1000 popuⁿ/year
school children

low risk area : ① Rheumatic fever is > 2 per 1 lakh

Prevention: 1st prevention

2nd prevention → ① Benzathine Pen G

6,000,000 IU for children with < 60 lb

1-2 million IU weighing > 60 lb every 3 weeks

② oral penicillin V = 250 mg BID

③ oral sulfadiazine = 0.5 mg OD < 60 lb
sulfisoxazole 100 mg OD > 60 lb

Allergy to PEN → Macrolides

RF 2 out condit: 8 y (or) 21 y of age

5 y or longer

RF 2 condit but

T but residual HD: 10 y (or) until 21 y of age

5 y or longer

RF 2 condit & residual HD

(Persistent valvular disease)

Life long

10 y to 40 y of age



satnday

USUS SAVES

19th Aug 2013

RESPIRATORY SYSTEM

M/c of URI: Rhinovirus

ARI control program

No pneumonia (cough & cold) → Hemoremedi

Pneumonia ⇒ RR/min Age → or trimoxazole (A)

Severe pneumonia
 60 or more < 2 months
 50 " " 2-12 "
 40 " " 12-60 months
 amoxicillin

→ N/im penicillin

Very severe pneumonia → IV chloramphenicol

IV ceftriaxone

8) All in Nov 14 = cough & cold = /Ans) No pneumonia

Neonate ⇒ group B streptococci > E.coli (worldwide)

⇒ In India = klebsiella m/c in newborn



3 wks - 3 month = Resp syncytial virus > streptococci pneumonia > H influenza (m/c bacteria)

4 month - 4 years = Resp syncytial virus

> 5 years = M. pneumoniae > S. pneumoniae > Chlamydia

Viral pneumonia = • Not lobar

• Diffused, B/L infiltrate

* Intestinal

Doc for RSV = Ribavirin

Pandemic

• phase 1-3 = animal, occasional human

• phase 4 = May 27, 2009; Mexico = human-human transmission

• phase 5 = May 29, 2009; Mexico = USA

• phase 6 = June 11, 2009

VIRUS

1. H1N1

• ssRNA, enveloped

• family = orthomyxoviridae

• size = 80 - 200 nm

• type = A, B, C

• Influenza type A surface antigen

H (haemagglutinin)

N (neuraminidase) → inhibitor = oseltamivir
zanamivir



Symptoms:

complicata in high risk groups

1. $< 1 \text{ yr}$; $> 65 \text{ yrs}$ (extreme of age)

2. pregnancy

3. chronic illness = liver

kidney

throat

4. Post transplant

5. Nephrotic synd

Indications of giving Oseltamivir

• All suspected cases

• confirmed cases = throat swab

Realtime PCR (RT-PCR) Nasopharyngeal swab

• to contacts = household

Doctor

• Dose = 75 mg capsule BD for 5 days

• Prevention = 75 mg cap OD x 7-10 days till the last day of contact

⇒ Zanamivir

Inhalational drug

• R_x = two 5mg inhalation

(total 10 mg) per day for 5 days

Propylhexu = 10 mg = once per day



~~not the agent~~

Phase of settlement

step pneumonia:

Incubation = 1-3 days

in resistant to ceftriaxone

Wacane agant s. pramona

(pneumococcal polysaccharide vaccine)

DPIT 24.7

ସତ୍ୟ ୩୧ '୦୧ '୨

Booster 15-18 months

comes by the name of pneumonia

• Routine: vaccine of cat



• GAVI = global alliance for vaccine & immunization

↑ Made by Bill Gates (founder) and his wife
Held for free in Pakistan

Staph aureus:

• High in mortality (10-30%)

• most dangerous

• pneumotoxole = can also be seen in Klebsiella
↓
kerosene oil poisoning

there can rupture and form pneumothorax

• emphysema in children

• txb = cloxacillin

only for MRSA = vancomycin

ceftriaxone is Doc for influenza

Atypical pneumonia

• Rare < 45 years; > 5 years

• Persistent cough

• interstitial

• org are atypical like Mycoplasma

Chlamydia



INFECTIONS OF UPPER AIRWAYS

Fever, and stidor



upper airway obstruction

~~tracheo~~
Croup / laryngeal bronchitis

⇒ Croup / laryngotracheal Bronchitis



• it is viral

• 75% caused by parainfluenza

• Prodrome = stidor

• Barking cough (croup)

• X Ray = steeple sign

• Doc = Mild croup = High dose Dexamethasone 0.6 mg/kg

moderate - severe = nebulised racemic epinephrine

Epidemiology

Fever; stidor; Crouping = Bact = 1. Epiglottitis

infant 2. Diphtheria



Vaccine eradicated disease



• Rx = Macrolide



There is an trapping

$$R = \frac{1}{4}$$

Artery resistance is inversely proportional to $\frac{1}{R}$ (radius of artery)

Pituitary, chronic lung disease

(1) to (2) shunt

Smoking mother

Males: Toppled

Risk factor

50% of it is caused by RSV

Inflammatory obstruction of smaller airways

→ BRONCHITIS →

Ceftriaxone + Sulbactam (ESBL)

It is Airway

India = H. Influenza

S. Aureus

S. pneumoniae

S. pyogenes

World

Epiglottitis

(expiratory sound)

Viral prodrome = wheeze/rhonchi

• x-ray = hyperinflation

• tx = 1. Bronchodilation have no role

2 Doc = humidified O₂

3. Nebulised ribavirin (to ↓ to ↓ shunt)

(return to lung disease)

4. Palivizumab

is also given as prophylaxis to prevent

further attacks

Prognosis: excellent (only 1% mortality)

long term → Persistent wheeze during infancy

is a risk factor for reactive airway disease

(Blue) A 2 yr old child with acute severe asthma

Management: Oxygen

Nebulised 2 Salbutamol

Nebulised 2 Ipratropium bromide

9/c terbutaline

IV hydrocortisone

Terbutaline infusion

IV aminophylline

50% mg/kg IV



Foreign Body aspiration: persistent vit hyperinflation

Next step: Bronchoscopy

Remove FB

Classification of Bronchial Asthma

Day time
Night
Peak expiratory flow rate
symptoms
time symptoms
≤ 2 times a month
asymptomatic: < 2 times a month
Day time
Night
Peak expiratory flow rate
symptoms
time symptoms
≤ 2 times a month
asymptomatic: < 2 times a month

Mild persistent: > 1 time a week but < 1 time a day
Moderate persistent: > 2 times a week
Severe persistent: daily attacks
daily attacks
off attacks

continuous! frequent
limited physical activity
variable > 30%
predictable
variable > 30%

Intermittent: < 2 times a week
Mild persistent: > 2 times a week but < 1 time a day
Moderate persistent: > 2 times a week
Severe persistent: daily attacks
daily attacks
off attacks

Treatment: SAs beta-2 agonist

Mild persistent: ++++ inhaled steroids

[Budesonide, fluticasone, beclomethasone]

Moderate persistent: +++ montelukast / salmeterol
(long acting β agonist)



Severe persistent: +++ Oral, low dose, long term,
 alternate day prednisolone

← RECURRENT PNEUMONIA

2 episodes of radiographic pneumonia in 1 year and
 3 episodes in any time frame

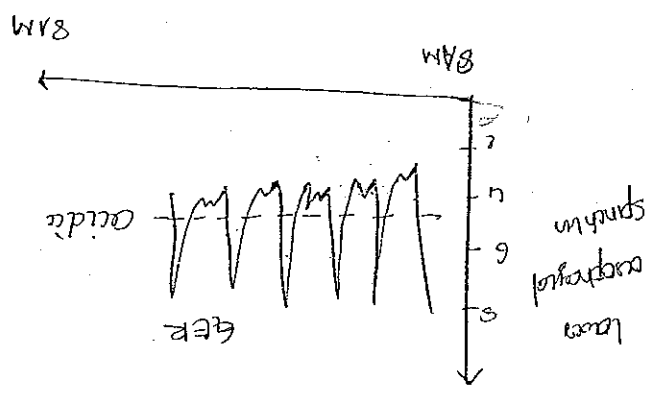
Persistent pneumonia: persistence of symptoms
 and radiographic abnormality for more than 1 month

DD

1. L to R shunt

2. Immunodeficiency (Eg: HIV)

3. Gastro oesophageal reflux disease



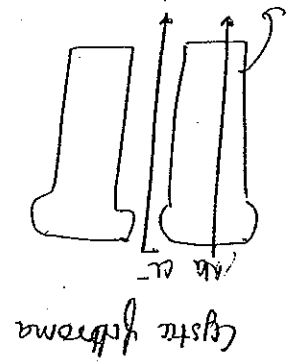
Feeding blo is more imp in recurrent pneumonia



→ CYTIC FIBROSIS ←

• CFTR gene on 7q
 2500 UK
 met

↓
 code for a chloride channel



more observed = children wont be hydrated

① Respiratory = pneumonia, >5yr

• m/c organism associated cystic fibrosis = Pseudomonas

<5yrs: S. Aureus, H. influenza

5-18yr = S. Aureus, pseudomonas

>18yr = Pseudomonas, S. aureus

Burst holderna: rare

2-5%

↑
 DDC = cithromoxazole specific

fatal



• Aeromonas
• achromobacter } rare

Anti pseudomonal antibiotic

peperasiline

Tecarsiline

Inhaled antibiotic

Tobramycin

Astreanam

Colistin

ciproflox

Amikacin

levofloxacin

② Cystic fibrosis = affects exocrine pancreas

↓
Steatorrhea < 5yr
↑
R_x foul smelling bulky stool

supplement pancreatic enzyme = Lipase capsules

m/c pancreatic insufficiency in children = cystic fibrosis

③ Endocrine 25% DM after 35 years

④ 4/11 : meconium-illness, intussusception, colon mucosal thickening
Dios (Distal intestinal obstruction synd) * appear in old pts





* New drug Investigator

* single gene CFTR have 1800 mutations

$\Delta F508 = \text{class II mutation}$ causes trafficking defect (phenylalanine)

3 nucleotides are deleted in $\Delta F508$ mutation (TTT code for

3. Seen in 25-35% of Indian children

2. 70% in Caucasian

Deletion

1. Delta F508 $\rightarrow$ m/c mutation $\rightarrow$ No position $\rightarrow$ Phenylalanine

Diagnosis of cystic fibrosis

① Absent, var is very common in cystic fibrosis men

• Azospermia
• Nasal polyps

Next investigation $\rightarrow$ Astic of Humpback diem
Lower GI contrast study $\rightarrow$ treat meconium illness

② 48 hr old boy does not pass meconium

Δ F508 → affects



Nucleotide

binding domain

Diagnostic criteria

Sweat chloride more than 60 meq/L on 2 occasions

(b)

Two known CFTR mutation

Best test: diagnostic, nasal electrode, potential difference

MCQ: Dysphagia lusoria = 1° duodenal baby

↓
vascular ring compresses the esophagus

Bar swelling (abdomen @ subcutaneous artery)

MCQ: H type of Tracheo oesophageal fistula (H-TOF)



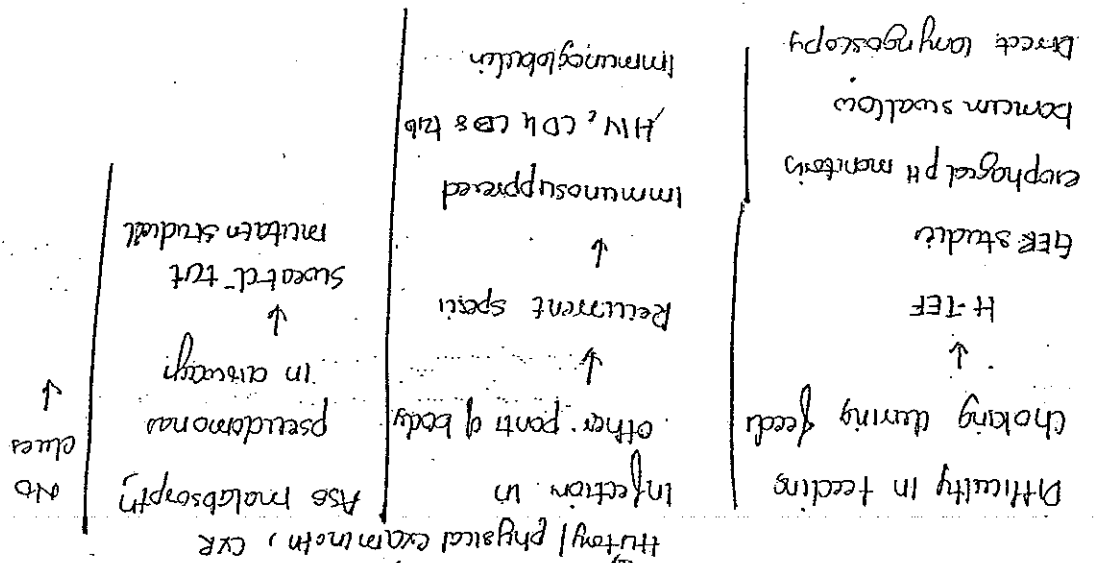
- Rare fistula
- Recurrent pneumonia & feeding difficulty
- Tracheobronchitis

malacia

IX = Tracheo Bronchoscopy

* Bronchoscopy and BAL

Approach to PT recurrent/persistent pneumonia



CENTRAL NERVOUS SYSTEM

Microcephaly = Head circumference > 3SD below mean

Head circumference

- 35 cm at birth
- 8 month - 40 cm
- 12 month - 45 cm
- 2 year - 48 cm
- 12 year - 52 cm

Cause of Microcephaly

Genetic cause

Downs

Edward's

Turner (3,18,21)

Cri-du-chat SP

Cornelia de Lange

Acquired cause

Mother / baby

TORCH

hyperphenylalanine

hypothyroid

Meningitis

Encephalitis

hypoglycemia

radiation

DM

alcohol



Macrocephaly : > 2 SD above the mean

Seen in

Neurodegenerative disorders = grey matter

Regression of milestones

Anemia, hepatosplenomegaly

Unexplained Deaf, Blind, seizures

* Cherry Red spot in Macula

Macrocephaly

1. GM 1 gangliosidosis

(β galactosidase absent)

develops neurodegenerative symptoms

baby dies in 3 yrs.

2. Gauchers

(β glucocerebrosidase deficiency)

3. Niemann pick

(sphingomyelinase variant)

Huge splenomegaly

Tay-Sachs :

• Def of β hexosaminidase A

• AR; Askenazi Jews

• 6 months - exaggerated startle reflex/hyperaesthesia



(β hexose aminidase A deficiency)

Sandhoff

(β hexose aminidase A deficiency)

1. GM 2 [Tay Sachs]

Macrocephaly

Seizure

Cherry-red spot macula

No. organomergly [to sandhoff in which all are same but organomergly seen]

WHITE MATTER

1. frequent fall
2. incoordinated movement
3. upper motor neuron sign

Normal/microcephaly

a. kabbes

b. XLR adreno leucodystrophy

adrenal gland dysfunction

ACTH excessively produced

hyperpigmentation

Asu: Serum Very long chain

Fatty acid

degenerath shat from perito

occipital area

Rx: Early Bone marrow transplantation

Metachromic leucodystrophy



eg white matter

Canavan = Diffuse thickening

area + Basal ganglion

from frontal periventricular

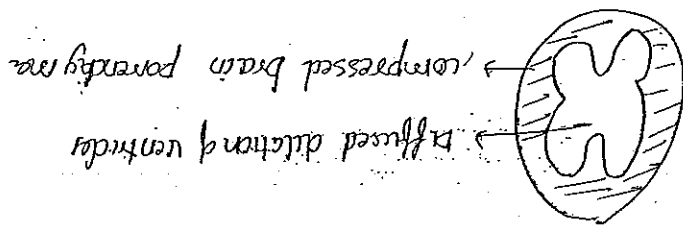
degeneration shat from

b. Alexander disease

a) Canavan's disease

Macrocephaly

CONGENITAL HYDROCEPHALUS



CSF production

• choroid plexus (25%) lateral, III & IV

• Ectra choroidal plexus (25%) capillary endothelium in brain

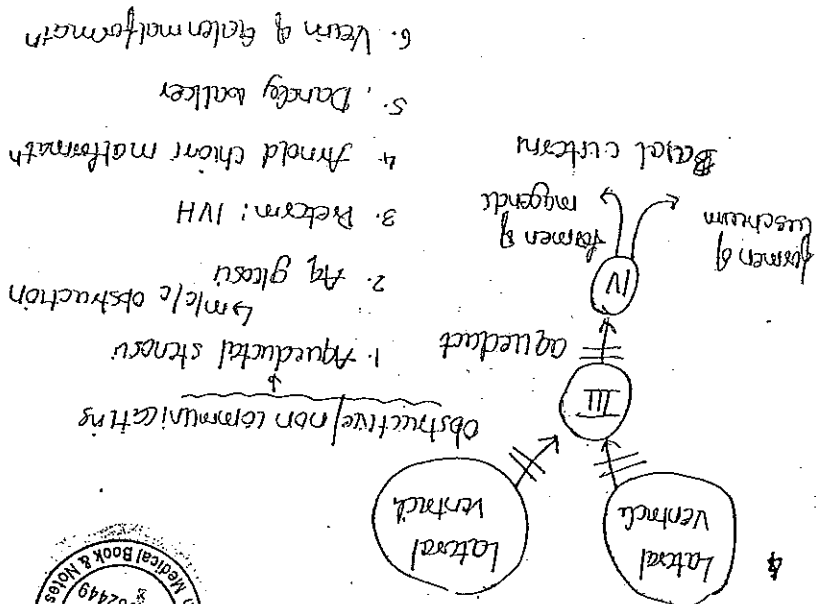
parenchyma

• Rate of CSF production : 20ml/hr

• Total vol in infants : 50 ml

• Adults : 150 ml

CSF flow



1. Aqueductal stenosis
2. Arq. gliosis
3. Porence: IVH
4. Arnold choroid malformation
5. Dandy walker
6. Vein of Galen malformation

Hydrocephalus = Enlarged ventricles $\rightarrow$ out $\rightarrow$ in intracranial tension

$\Rightarrow$ Arnold-Chiari malformation:

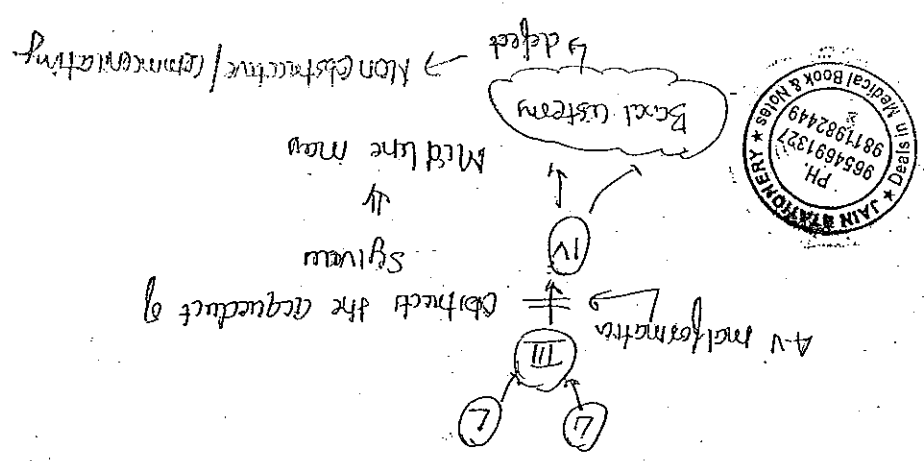
downward displacement + hypoplasia of cerebellum
 leads to obliteration of cisterna magna
 CSF outflow is also obstructed

$\Rightarrow$ Dandy-Walker syndrome

large post fossa cyst communicating to 4th ventricle
 Vermin aplasia
 cerebellar hypoplasia
 appearance of Bat wings

$\Rightarrow$ Vein of Galen malformation

m/c arterial venous malformation
 Hydrocephalus is seen



Non obstructive / communicating

1. Basal excoelatus

- TB

- cryptococcus

treatment of Hydrocephalus

1st use by Drugs : Furosemide

acetazolamide

Alt sx : Ventriculoperitoneal shunts (VP shunt)

complication : Blockage

infection (coagulase -ve staph)

CNS

also called staph epidermid

Seizures / Epilepsy

Abn electrical activity of brain = seizure

Recurrent unprovoked seizure = epilepsy

Not related to fever

(a)

acute cerebral acidosis / Insult

m/c/c : seizure in newborn : subtle seizure

cause HIE

m/c/c seizure in child = febrile seizure



Typical febrile seizures

41°C

36 months - 5 yrs

1/ febrile synd

< 10 minutes, one episode

Risk factor for recurrence in febrile seizures

MAJOR

Age < 1 year

Duration of fever > 24 hr

Fever 38-39°C (100.4 - 102.2°F)

MINOR

F/H of febrile seizures

Family history of epilepsy

Complex febrile seizure

Daycare

Male gender

Lower serum sodium at time of presentation

* No risk factor = 12%

* If there 1. Risk factor = 25-50%

2 " = 30-59%

> 3 " = 78-100%



Risk factor

Risk for subsequent epilepsy

Simple febrile seizure 1%

Recurrent " 4%

Complex " 6%

Fever < 1 hr before febrile seizure 11%

Family H/o epilepsy 18%

Complex febrile seizure (focal) 24%

Neurodevelopmental abn 33%

treatment of febrile seizure

1. per rectal diazepam via: Nasogastric tube for

2cm into rectal canal

2. Prevention

prophylaxis: No need

concerned/ recurrent

Intermittent prophylaxis

only when seizures
oral clobazam (as)

oral diazepam for 48-72 hr of fever

Prolonged febrile seizure children have: Altered temporal

structures in MRI



→ Epilepsy in children →

(Nelson; OP Gal)

Partial

• simple partial (No loss of awareness)

• complex partial (loss of awareness)

— aura

• automatisms (involuntary orpharyngeal movements)

• treatment carbamazepine

carbamazepine

Ring enhancing lesion (Partial epilepsy)

Neurocysticercosis
↳ Tuberculoma

• usually large; >20mm

• Multiple

• irregular margin

• Peritumoral edema

• Mid line shift

Antihelminth
{ Dexamethasone
(c) Prednisolone

treatment: steroid 3-5 days

• SCOLEX

• solitary

then given Albendazole



Generalized, Epilepsy

• Tonic

• Clonic

• GTCS [aura GTCS, post ictal phase]

↓

opening of eyeballs

tongue bite

both

incontinence

• Myoclonic seizure

• Atonic

treatment: For all generalized seizure

Doc: Valproate < 2yr → Hepatotoxicity

* Generalized 3 Hz spike = Absence seizure

↓

• Lasts < 30 sec

• No aura

• No post ictal phase

• > 5 yr

• Hyperventilation, Provokes

• Female > male

• Doc: Ethosuximide 4/b

Valproate



Atypical Absence seizures

also the myoclonic component

Doc: Valproate

⇒ INFANTILE SPASMS / SALAM / WEST SYND

4-8 months

Flexor, symmetrical

1. idiopathic / cryptogenic type = Good prognosis

Secondary

- HIE

- structural malformation of brain

Seizure activity

- Recons synd

- Tuberculous sclerosis? pts = Doc = Vigabatrin

EEG a very specific = Hypsarrhythmia

↓
generated spike of high voltage, slow waves

- Doc = ACTH to suppress CRH



* M/c generalized epilepsy in ~~adult~~ adolescents & young adults
 11 (20-30 yrs)

Juvenile myoclonic epilepsy - JAZZ synd

• 12-16 yrs

• Morning - myoclonic jerks

drop things

started getting generalized seizure (90%)

• only 1/3 of them have absence seizure

• Bad news: life long disease; No remission

• Good news: Valproate: good response

EEG • 4-6 Hz generalized spike + photic stimuli

STATUS EPILEPTICUS child

• Convulsion > 30 min (∞)

• No regain of consciousness b/w convulsions

• Area of brain which is vulnerable to hypoxia

Hippocampus

Amygdala

thalamus

subcortical

• treatment: DOC: IV lorazepam (fast onset)
 (large + 1/2 life)





Ques) 6 months old child comes with few & seizure
• Meningeal signs are rare in > 18 months
• Every child < 12 months: 1st episode few & seizure
↓
then do lumbar puncture (CSF)
↓
to exclude meningitis

Repeat
General anaesthesia: Thiopentone
• IV levetiracetam 20 mg/kg (Acutrus)
• IV Midazolam 2-20 mg/kg/min
• IV Valproate 20-30 mg/kg
• Phenobarbitone 20 mg/kg → 10 mg/kg
• Repeat 20 mg/kg → 10 mg/kg
short t_{1/2} life so combined

↓
In hospital: IV Midazolam + Phenytoin (Bolus) 20 mg/kg

→ INFECTIONS ←

MENINGITIS

INDIA

WORLD

Group B & D streptococci

Klebsiella

E. coli

2 months - 3 years

H influenza

Strept pneumoniae

type B

> 3 years Strept pneumoniae

S. pneumoniae

USA = Neisseria meningitidis = bug there is no vaccine to Neisseria

Acute Bacterial Meningitis

95% cases occur b/w 1 month to 5 y

Defects of complement system C3 and properdin

system meningococcal infection

T-lymphocyte defects (e.g. AIDS/chemotherapy) →

ultra monocytes

* Longitudinal / acquired defect across mucocutaneous barrier → pneumococci

• Lumbosacral meningo myeloid and dural sinus → Staph & enteric bacteria



ceftriaxone + vancomycin



Resistance

treatment of Meningitis: ceftriaxone

Streptococcus

carbapenem 1st #

Recurrence meningitis in CSF leak

chicken pox vaccine = live vaccine = 1 month before

be done before splenectomy

but vaccination should

all have vaccine
↓
pneumococcal
↓
H-Influenza
↓
Neisseria
↓
we get capsulated infection

↓

asplenia

due to side cell dysfunction

↓

In splenectomy / autosplenectomy

coagulase -ve staph

Penetrating CNS trauma / CSF shunt infection →

Neurological sequel of meningitis

- SNHL (sensorineural hearing loss)

infection = SNHL via aqueduct
↓

Prevention = Dexamethasone 0.15 mg/kg

30-60 min

Before antibiotic

antibiotic prophylaxis to the contacts to

household / occupational contacts

DOC: 1m ceftriaxone single dose

Rifampicin for 2 days

~~ceprolox~~

Fluoroquinolone: ciproflox

→ ENCEPHALITIS →

AP4 2010, 2011

M/c 80% enterovirus

sporadic = HSV

• In CT = B/L temporal Hypodense: HSV
• In MRI = temporal one hyperintense





Traumatic neuritis = inflammation
in gluteal region in para
sacral nerve

Paralytic polio

AFP can be Asymmetrical AFP (vll)

Hypothalamic lesion Hypo
thalamia
early syphilis

Spetic arthritis

causes: osteomyelitis

provided we should Rule out pseudoparalysis

In children < 15 years

Acute onset < 6 weeks

→ ACUTE FLACID PARALYSIS ←

DOC: Doxycycline

TYPHOID ENCEPHALITIS:

Untreated HSV = 70% mortality

But fast to HSV = QFT PCR

DOC of HSV = IV Acyclovir

Localized temporal discharges: HSV

- Symmetric AFP (B/L)

Transverse myelitis: Urinal = HSV

Myeloma
Ventricles

level: Motor

sensory

thoracic (mostly)

Treatment: Dec: IV methylprednisolone

* m/c of AFP worldwide = Guillain Barre synd / AIDP
(acute inflammatory demyelinating)
(polyneuropathy)

- Campylobacter Dysent

- after 10 days weakness sets in

- It can also occur after Mycoplasma

Strept pneumoniae

- Areflexic [DTR absent]

- Ascending

- Symmetric

- Involves Diaphragm

Diagnosis: CSF

1st week

2nd week

cells

10/hpf

10/hpf

protein - 50mg/dL

50mg/dL

Classically seen in 2nd week $\Rightarrow$ Albuminocytological dissociation



② Nerve conduction studies

Bilateral symmetrical demyelinating disease

treatment: IV Ig

↓ if fail

plasma pheresis

* All ventricular dilatation = communicating hydrocephalus

↓

Scan m. TB

Meningitis

* Indication of IV Ig

Immunodef = hypogammaglobulinemia

↓

B cell don't work

↓

pt has to keep left leg IV Ig

Kawasaki

Gillian Barre synd [AIDP]

ITP

Rh Isoimmunization





* Indication of plasma pheresis

• Removal

- anti GBM (palm hemorrhage)

- Guillain Barre synd

- dialysis dependent ANCA +ve crescentic GN

[Wegener, microscopic polyangitis]

- Deficiency: Factor H mutation HUS

~~anti~~ f

antibodies against factor H

Atypical HUS

NEPHROLOGY

GFR at birth = $15-20 \text{ ml/min/1.73 m}^2$

At 3 months = $2/3$ of adult

Adult values = 2 gm

Tubular concentration reaches adult value by 1 yr of age

Nephrogenesis is complete at 36 wks of gestation

Child born preterm @ 104R = develop HTN in later life

less no of nephrons

GFR maturation

Schwartz formula (in children) = depends on height

↓
Muscle mass

↑
Creatinine comes from muscle

$$e \text{ GFR} = \frac{\text{Creatinine (mg/dL)}}{K \times \text{height (cm)}}$$

K = constant

3. Jaffe's reaction method

Best method: HPLC/arginine assay

Value of K in Schwartz

LBW infant = 0.33

(N) infant 0-18 months = 0.45

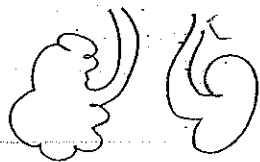
Girl 2-16 yrs = 0.55

Boys 2-13 yrs = 0.55

Boys 13-16 yrs = 0.70



M/c abd mass in newborn: Multicystic dysplastic kidney



- 80% v/l
- non functional
- "bunch of grapes"

If both the kidneys do not work = Potter face

B/L kidney agenesis

→ OLIGURIA →

• If u the urine output $> 400 \text{ ml/m}^2/\text{day}$

New urine output $< 1 \text{ ml/kg/hr}$

• In acute kidney injury Very common

" " Glomerulonephritis very common

Non oliguric AKI

Aminoglycoside (Gentamycin)

damage the mitochondria of PCT

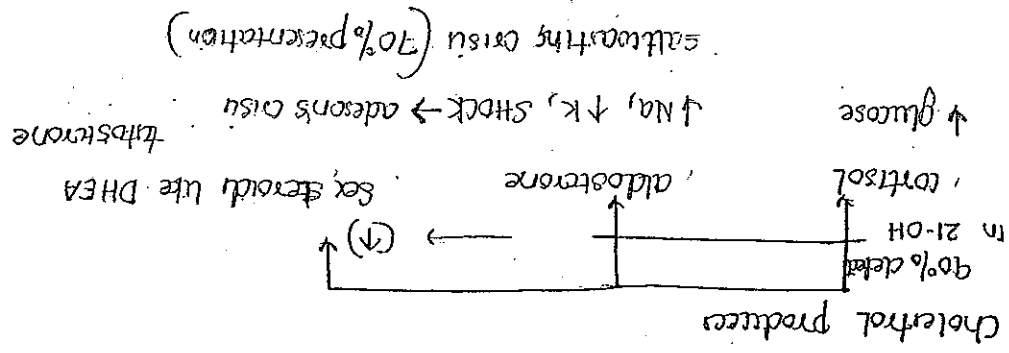
Neonatal renal failure

AKI = polyuric phase (in resolving phase)



CONGENITAL ADRENAL HYPERPLASIA (CAH)

M/c imp/case of pseudohypoparathyroidism
 ↑
 karyotype is xx but externally
 Baby appear as ♂



2 weeks child = creatinine = 0.2 to 0.4 mg/dL

Boys with CAH = have more testosterone = precocious puberty

Girls with CAH = " " " " = cutaneous hyperpigmentation = looks like penile

ACTH ↓ cause hyperpigmented labia = look like scrotum

treatment: supplement glucocorticoids

Mineralocorticoids

Rx: Hydrocortisone 15-20 mg/m²/day
 ↓
 8 hourly

Fludrocortisone 0.15 mg/day

Girls with CAH = they require reconstruction sex



Boys: $\downarrow$ ACTH = $\downarrow$ pigment, precocious puberty, hypoparathyroidism, ($\uparrow$) renin

testosterone = cause premature epiphyseal closure
 CAH is a cause of short stature

→ AMBIGUOUS →

ambiguous genitalia

Boy = penoscrotal hypospadias + empty scrotum

when 21-OH is deficient

17-OHP is accumulated

17-Hydroxyprogesteron = every child is tested for it

level of 17 OHP in CAH = $> 600 \text{ ng/ml}$

usually in 1000's

Deoxymethasone: cross the placenta

prevent the virilization of next baby

20 mcg/kg of pre-pregnancy wt

17 OH deficiency cause HTN in CAH

$\downarrow$

DOCA is potent mineralocorticoid

$\downarrow$

it is accumulated = cause HTN

fludecort = used in ASU of CAH



Hypokalemic metabolic alkalosis = Aldosterone defect (leads to) Male looks as female in 17 α hydroxylase deficiency

SO $\downarrow$ testosterone

AT aldosterone & testosterone

21 testosterone $\downarrow$ 1 = mean = high

17 Aldosterone $\downarrow$

11 both $\uparrow \downarrow$

Aldosterone synthase deficiency = leads to

N $\uparrow$ $\downarrow$ dehydration, hyperkalemia, hyponatremia

→ HAEMATURIA ←

Blood in the urine

M/c/cause = stone

two types = 1. Gross

2. Microscopic

Microscopic haematuria = > 5 RBC/hpf centrifuged

Haematuria = glomerular

Extraglomerular

Glomerular

IgA, MPGN

Post streptococcal GN

Dysmorphic RBC

Extraglomerular

stone, haemorrhagic cystitis

UTI, ischaemic adenoma

Idiopathic hypercalcaemia

A common observation



24 hr urine ca. $> 4 \text{ mg/kg} = \text{hypercalcaemia}$

Bright red
cola colored urine
painless, proteinuria

Recurrent gross haematuria

m/c worldwide = IgA Nephropathy

other causes = MPGN

Idiopathic hypercalcaemia

Alport's syndrome: X linked dominant $> AR > AD (5\%)$

Present like recurrent gross haematuria

third eye = anterior lenticonus (very specific for Alports)
ear = deafness (SNHL)

kidney = recurrent gross haematuria

$\rightarrow 75\%$ boys required kidney transplant by they reach 30yr
 $\rightarrow$ Boys have severe symptoms

Alpha 5 domain of collagen IV affect Basement memb

in Alports syndrome

collagen type IV (Collegraphy)
Anti GBM = $\alpha 3$ domain of collagen IV
Thin GBM = $\alpha 3-\alpha 5$ " "
Bengal familial Haematuria = $\alpha 3-\alpha 5$ domain of collagen IV



Feature of Alports on: Etchen Microscopy

Irregular Basement memb = some are thick

" " thin

Basket weave appearance = lamina densa of

Basement memb is

splitted

→ ACUTE KIDNEY INJURY →

Now a day we do not ^{say} kidney failure

Biomarker : present in urine

1. Best Biomarker in world for AKI = NGAL

(Neutrophil gelatinase associated lipocalin protein)

NGAL = rises 2 in min; creatinine = rises in 2 days

2. Urine IL-8

3. Urine KIM-1 (kidney injury molecule)

4. Urine β -FABP (Leucocyte fatty acid Binding protein)

5. Serum cystatin - C

AKI = prerenal

renal

postrenal

Prerenal (common in New born)

hypoxia

asphyxia = early organ involved = kidney



• Hypertension

• Hypovolemia

• Burns

Renal : HTN ; HUS

Post renal : obstruction by stone, tumour & clot

In severe diarrhoea = arginase feature
↓
give Ringer lactate

Indices

Pre renal

Renal

Urinary sodium (mg/L)

< 20

> 40

Urinary osmolality

> 5000

< 300

B-urea/creatinine Ratio

> 20:1

< 20:1

Fractional excretion of Na%

< 1

> 1

$\frac{\text{Urine Na}}{\text{Serum Na}} \propto \frac{\text{Urine Cr}}{\text{Serum Cr}}$

↑
(49% Na observed in tubules in renal cause)

50 1% in urine excretion

• M/c/cause of AKI in child & Adult world wide : ATN

(Acute tubular necrosis)

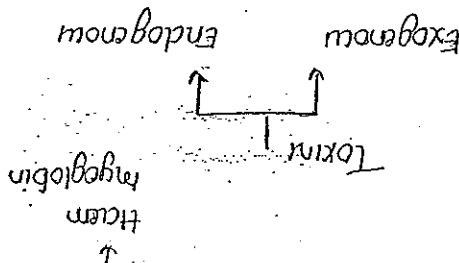
• Tubular Necrosis



Tubular Necrosis

Muscle crush injury = cause AKI

Drugs



myoglobin } soluble in alkaline
 hem } medium

sodium bicarbonate

in acidic medium = ppt in tubules

Hemolytic Uremic syndrome (HUS)

Triad: Microangiopathic hemolytic anemia

Thrombocytopenia

AKI

90% follows diarrhoea

Developed = E coli O157; H7 → burger disease

countries

↓
 E.coli on burger

Developing countries = Shigella Dys I

Germany June 2011 = E.coli O104: H4 on "sprout from Egypt"

Shiga toxin = endothelial injury

↑
 cause platelet aggregation

also called as

↓
 TMA (thrombotic microangiopathy) → Thrombus formation



schistocytes in peripheral smear is due to breakage of RBC

Thrombocytopenia (PNI) DOC in Paroxysmal nocturnal hemoglobinuria

DOC = Eculizumab (C5) new humanized monoclonal Antibody
 world costly drug (CR) against C5
 Plasma pheresis → (in India)

HUS may be diarrhoeal Negative

(HUS D-)

Mitomycin C

gemtazabine (pancreatic ca)

Tacrolimus, cyclosporine

Eclampsia

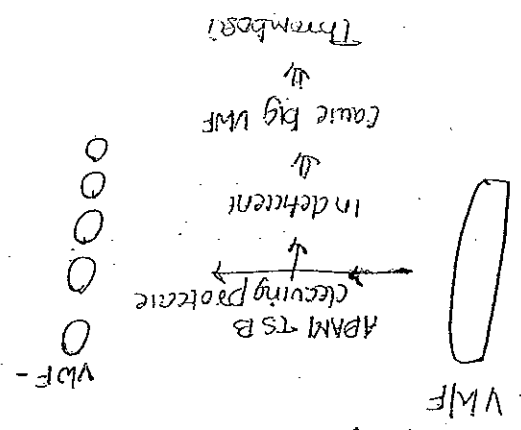
HIV

SLE

genetic = factor H mutation

anti H factor Antibodies

Adults: TTP = multi-systemic thrombus formation
 = fever and neurological symptoms



1.73 m² = Adult Body surface area

Complication of AKI

- Fluid overload
 - ↳ Rx: Restrict fluid insensible loss 400 ml/m² + replace urine output
 - ↳ Rx: Insulin + Dextrose
 - ↳ Metabolic alkalosis
 - ↳ Transcellular shift of potassium to cell
- Hyperkalaemia
 - ↳ Rx: Insulin + Dextrose
 - ↳ Metabolic alkalosis
 - ↳ Transcellular shift of potassium to cell
- Dilutional hyponatraemia
 - ↳ Rx: IV Furosemide (Distal tubular Na⁺ diuretic)
 - ↳ IV NaHCO₃ so that it excreted
- Dilutional anaemia
 - ↳ Rx: Resin: koxylate-polystyrene
- HypoPO₄ → hypocalcaemia
 - ↳ Rx: Resin: koxylate-polystyrene
- Metabolic acidosis
 - ↳ Rx: Dialysis
- Hypertension
 - ↳ Rx: IV calcium gluconate

[Insulin] ↓ K⁺ in 10-15 min

[Ca²⁺] stabilises the cardiac memb (cardioprotective)

* Cal gluconate is only given when: ECG changes in hyperkalaemia

→ sine wave pattern

Caution with

* Indications of dialysis

• Uremia = encephalopathy, pericarditis

• Refractory hyperkalaemia

• Refractory metabolic acidosis

• dysnatremia in renal failure

fluid overload



→ CHRONIC KIDNEY DISEASE ←

Causes in children

< S_{crea} = hypoplasia

dysplasia

PUV (post urethral valve)

> S_{crea} = acquired = GN/HUS

Features CKD

A Azotemia

A acidosis metabolic

A Anaemia (normogytic normochromic) sub/utancor
rh EPO

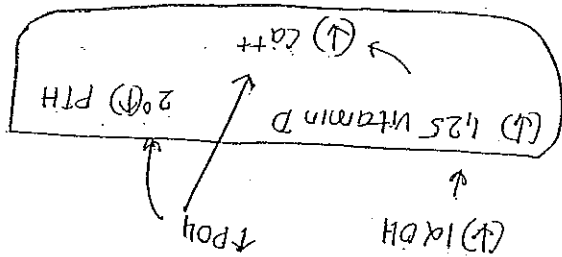
B Bone

C Cardiovascular calcification CaXP

E Growth failure
m/c cause of death in adults

When GFR fall 40-60

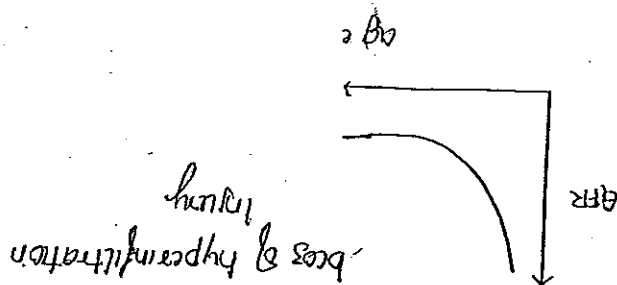
Renal osteodystrophy



Hypertension injury

If kidney injury occur then remaining nephron leads to hyperfiltration injury

ESRD: $GFR < 10 \text{ mL/min} / 1.73 \text{ m}^2$



treatment: life long renal replacement

(dialysis) Transplantation

But txt: * living related donor renal transplantation

→ NEPHROTIC SYNDROME ←

Proteinuria $> 40 \text{ mg/m}^2/\text{hr}$ (a) 2g/day

hypoproteinaemia

Edema: S. Albumin $< 2.5 \text{ g/dL}$

hyperlipidaemia

Cholesterol $> 200 \text{ mg/dL}$

children: minimal change disease (a)

electromicroscopy Foot process podocytes one apposed



Older > 16 yr : F545
 african american

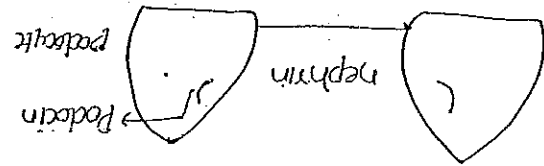
DOC: prednisolone steroid sensitive

* Sodium and water retention is m/c cause of edema
 in Nephrotic syndrome

* In nephrotic synd = lost

- Albumin
- Ig G → infectious SBP spontaneous bact peritonitis
- Thyroid Binding globulin TBG; VDBG
- transferrin; ceruloplasmin
- protein C, S; AT III lost → Procoagulant state

Fibrinogen $\uparrow\uparrow$ (normal = $< 150 \text{ mg/dL}$)



Gene Protein disease

NPHS I (chr 19) → nephrin → Finnish congenital nephrotic (< 3 months)





KIT 1 (tumour suppressor gene)
→ (11 p 13) → Frauer syndrome
KIT 1 (Xy) Male pseudohermaphrodite + Psgs
↑
ultraecond = undescended testis
↑
cause gonadoblastoma
↑
gonadectomy

HTN
post capsular cataract
short stature
impaired glucose intolerance
Doc = oral cyclophosphamide

steroid dependent and steroid toxic
- cyclosporin
#16
Tazelum (better drug)

Rx: Doc = calcineurin inhibitor
Psgs

NPHs2 (Ch 1) → Podocin → steroid resistant

URINARY TRACT INFECTIONS

Definition: symptom of UTI + urine culture $> 10^5$ CFU/mL

m/c/ symptom: fever

m/c/ organism: E. coli

common in ♀ (bags short urethra)

> 1 yr boys

Haematuria = > 1 yr boys

Ascending infection

Asymptomatic bacteraemia = NOT to treat

10^6 CFU/mL

Best urine sample = Suprapubic aspiration

No E. coli colony

E. coli colony present in urethra

cause of m/c/ UTI in children = Vesicourethral reflex (VUR)

Investigation of choice

MCV/VGUG

pelvic scans = DMSA nuclear scan



Function of kidney: MAA3 / DTPA nuclear scan

→ GLOMERULO NEPHRITIS →

• Edema

• HTN

• Haematuria

• m/c infection in India: Acute post streptococcal GN

↓

Infection of throat (serotype 12) / skin

(serotype 49) is nephritogenic

Strain of *B haemolytic streptococcus*

• Age 5-12 years

• Acute phase resolves 4-6 wks but

abnormality in urine in 1 year

• 95% Resolves; only 5% CKD

PSGN

4-6 wks

Edema

X

Proteinuria

X

Hypertension

X

Haematuria col X

urine RBC → 1 year

G → 1-2 wks





cytoglobulinemia

SLE

Shunt nephritis

Factor H mutation

C3 & low in = I.E

Presistent decrease in C3 = MPGN, C3 glomerulopathy

(anti DNA or B testing)

Pyoderma = 4-6 wks = to develop PSGN

(Also testing)

Scar throat = 1-2 wks = to develop PSGN

GENERAL PAEDIATRICS

GROWTH

Weight gain

Average wt gain in 4 months

30 gm/day - 1st 4 months

20 gm/day - 5-8th months

10 gm/day - 9-12 months

X 2 = 5 months

X 3 = 1 year

X 4 = 2 years

X 5 = 3 years

X 6 = 5 years

X 7 = 7 years

X 10 = 10 years

Length

Growth velocity

At birth 50 cm } 25 cm

1 year 75 cm

2 year 90 cm } 15 cm

4 1/2 year 100 cm

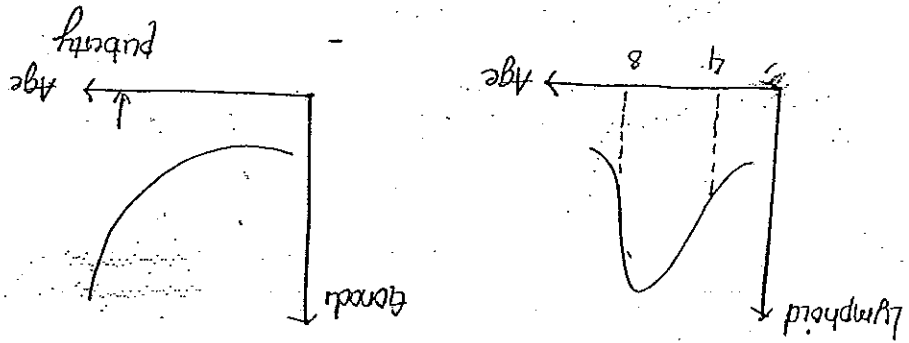
Add 6 cm / year till puberty

Age

Somatische



School going children = 6-8 yrs / year Ideal



~~Adolescent~~ Tanner's = Sexual Maturation Rating

Boys : Genitalia, Pubertal hair
Girls : Breast, Pubertal hair

SMR grading : 1 = No character

5 = completely developed

* girl gets menarche at SMR stage 4

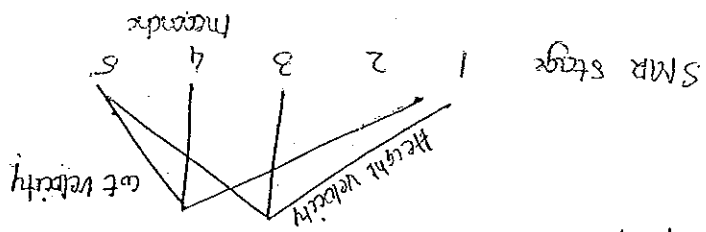
growth spurts → Thelarche → Pubarche → Menarche

1st clinical sign

* Boys

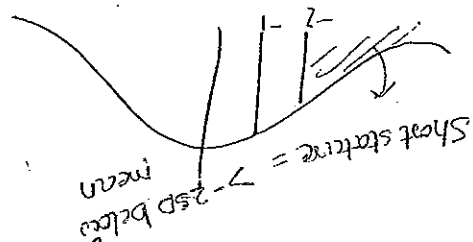
Testes → Penis → Pubic hair → Axillary hair

girl





- Mid parentile height
- as a adult predicted height
- Boy = average - 1.65 cm
- Girl = average - 0.5 cm



Height < 3rd centile for age & sex

SHORT STATURE

craniopharyngioma

Tubercular meningitis

Organic: hypothalamic hamartoma

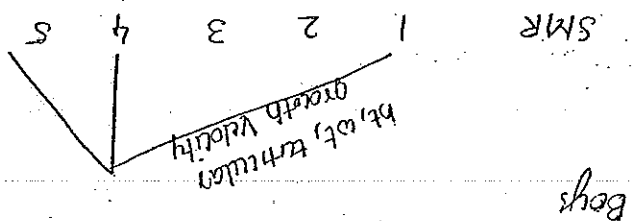
♂ = MRI Brain

♀ = Idiopathic

Precocious puberty = gonadotrophin dependent & central

1. M/c of delayed puberty = constitutional

ABNORMALITIES OF PUBERTY



• No LH, FSH → micropenis, hypoglycaemia
 • Hoarse voice, doll like face } panhypopituitarism

- Bone age delayed
- Age 1-3 yr
- No birth wt & length

↓
 • Growth hormone deficiency

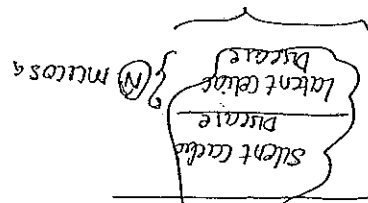
→ Cushing

hypothyroid

→ +ve ecology

DG², DG⁸

Genetic susceptibility



- Celiac disease ↓
- Gluten free diet = ht ↑ (✓)

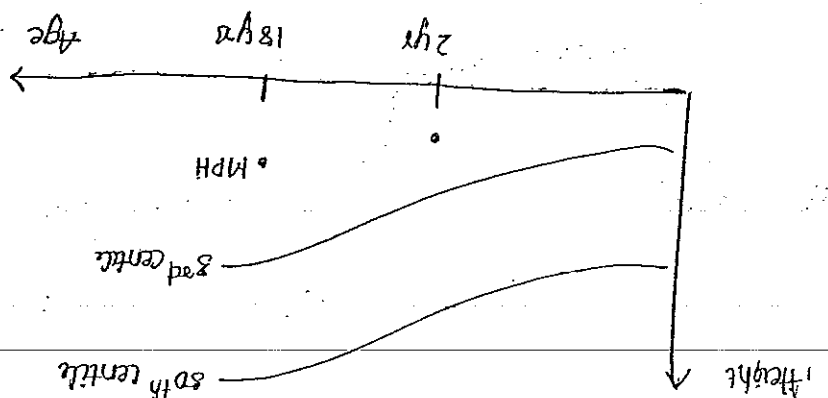
⇒ Celiac lobes
 → Symptomatic
 → Short stature
 → Mucosal mandible

- Chr illness
- Malnutrition

Pathological

• Physiological & Familial
 • Constitutional

Causes:



Ass of GH deficiency

growth hormone stimulation test

Basal level

stimulated level = 2 stimuli

L-arginine }
clonidine } act as stimuli
Insulin

↑
after stimuli

↑
no response

↑
GH deficiency

• IGF BP3 protein : not specific

Indication

gle high indication us FDA

given to GH deficiency

Turner

Prader Willi

CPD

HT > 2.25 below mean

Disproportionate SHORT STATURE

us/ls used . Achondroplasia, Ricket,

Hypothyroid



US/LS ved

• MPS IV morbus

TB spine

* m/c/cavc of short stature = Physiological

Physiological short stature
↑ Constitutional ↑
↓ Familial ↓

Birth length Normal

Lag 6-12 months

Growth velocity Normal

Final height Normal/subnormal

Puberty Delayed

Bone age Delayed

Qued MF < 3rd centile

Growth velocity (N)

↓
Constitutional



→ DEVELOPMENTAL MILESTONES ←

Gross motor
Fine motor

Social

CROSS MOTOR

3 months Head holding

5 " sitting & support

4-6 " rolls in Bed;

8 " sitting & out support

9 " crawling

10 " creeping (stand & support)

12 " standing & out support; walking & out support

18 yr walk up stairs & two feet on each step

3 yr walk up stairs & one foot; not tripping

4 yr hops on one foot

5 yr skips on two feet

FINE MOTOR

3 month = moros & grasp disappear

12 ~~month~~ ^{weeks} = grasp reflex disappear

4 month = bow for objects



5 month bidegrou grasp

6-7 " Transfer object, palmar grasp

9 month Pincer grasp "native, neat" $\downarrow$ 1 year

13 month: casting

15 " self feed & spoon

18 " " " cup

24 " turn page of a book one at time

15 month tower of 2 blocks

18 month train

2 year

3 year bridge

2 year chain of chimney
3 year (tower of 9)

4 year = gate
5 year = steps $\rightarrow$

Fine hand skills

3 yr (lwd)

4 yr +

rectangle

4 1/2 yr





Alarm therapy (Bat~~R~~)

Bat~~R~~ Rx = Behavioural +ve reinforcement

At 14yr old $\bar{c}$ Nocturnal enuresis = bat Rx

Nocturnal enuresis > 5 years (IS)

Bladder control = 85% by 5 years

PqE1 $\bar{c}$ out supervision = 5 years

dress / lunchen himself (under supervision)

dry by night

Dry by day

stranger anxiety

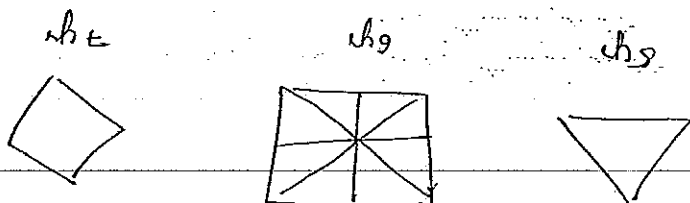
waves bye bye

smile at mirror image "enjoy"

recognize mother

Social smile

Social



Drugs: Dermopressive



only when alarm therapy fails

treat of alarm therapy = 5-8 weeks

Que) 6-yr old boy - enuresis

urine examination, ultrasound (N)

Next step?

counsel and re-eval after 6 months

ENCOPRESIS:

stools > 4 ym

cause: chronic constipation

Que) DUB II = Meaning of Death understood by child

> 3 ym no idea

3-9 ym idea

> 9 year = entity, irreversible, himself

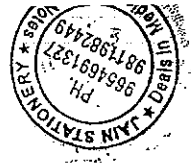
object permanence;

constancy = 9 months

~~Levelling~~

Peak a boy = 9 month





Cross a buy Road

10-12 years

Play

parallel play = 3 yrs

start, domestic role play = 4 yrs

Developed full domestic role play = 5 years

Language

1 month head turns to sound

3 month cooing

6 month babble; monosyllables (ma, ba)

9 month Bisyllables (mama, baba)

1 yr 2 word & meaning
15 mont = 4-5 words
18 month vocabulary of 10 words
2 yr simple sentence & 2 words

3 yrs = tells age and sex, uses pronouns, handedness

4 years = tells story

5 years = known 4 colour (he can name them)

Vocabulary of 50-100 yem = 2 yem

Blood Pressure

Ques) 4 ym, unconscious, HR = 180/min

LFT = 4 sec. BP = 80/60 mmHg

Diagnosis = compensated shock

⇒ Shock BP < 10th centile of that age & sex

10th percentile systolic BP

1 yr = 70 mmHg

> 1 yr = 70 + (age x 2) mmHg

treatment = Bolus 20ml/kg 0.9% NaCl

Response should be checked

↓

HR

capillary filling time

↓

No response

Rpt bolus 60ml/kg total

↓

CVP = Central venous pressure
keep CVP line



if CVP low = more fluid

CVP (N) / High = inotropes

Fluids: isotonic = (N) saline = 154 meq/L Na and Cl

finger lactate = urine plasma

↓
130 meq/L Na

4 meq/L K⁺

109 meq/L Cl⁻

28 meq/L lactate

3 meq/L Ca

HTN:

> 95th centile for sex & age

Adult

child

essential

10% essential, ↑

• Secondary hypertension

1. Renal Parenchymal - VUR

2. Renovascular Major: RAS, RVT

Minor: HUS

3. Cardiac = postductal coarctation

4. Endocrine = hyperthyroid



7

phocochromocytoma, CAH

II B / 13 alpha OH deficiency

s. end organ damage

Fundus

concentric LVH

urine protein

4 umb BP should be checked in children to rule out

coarctation

hypertensive emergency

LVEF 33; gallop; basal crepts

seizure

SOB: infusion

micardipine infusion

Camold "

labetalol "

giving for 48 hrs ↑
granule toxicity

↓ because is photosensitive
we should cover it

Sed mitroparside

1. end organ damage

2. Severe HT2 < 94th

Rx: ACEIS / ARBS

Indication:

wk style modification

obese HTN



(Ques) ALEI c/I in GFR < 30

hypokalemia: Doc ?
CCB (amlodipine)

Takayasu aortitis: (pulse len duar)

USA / Japan
↓
and of aorta affected
Abd aorta affected
Rec: stent

Tachycardia ECG

↑
QRS
VT/VF
(broad)

HR > 220/min
Narrow
> 180 m older children
P wave absent
inverted

↑
Vagal maneuver
(a)
Vagotonic
stable
Vagotonic
amiodarone

↑
Defibrillation 0.5-1 J/kg

↑
Stable / Infant
CHF / shock
synchronised
cardioversion



Important in Diarrhoea (NEET 2013)

↑ Na⁺ / ↓ K⁺

Mmol/L
 into ORS
 Re 36 Mal
 300
 osmolality Des 245
 Sodium 90-100
 45
 75 Retardation

Potassium 20 40 80-100 mmol/L

glucose 111 125 75

chloride 80 70 65

citrate 10 7 10
 + Mg, Zn, Cu

ORS packet
 NaCl 2.6 gm
 KCl 1.5 gm

C₆H₅NaO₇ · 2H₂O 2.9 gm → trisodium citrate
 glucose anhydrous 13.5 gm



WHO Dehydration

No dehydration - replace ongoing losses - 5-10mg/kg
 Some 0.5-7.5 mL/kg 4 hrs
 Severe IV Ringer lactate

> 1yr	0.5 hr	30 mL/kg
< 1yr	1 hr	70 mL/kg

WHO Zinc in Acute diarrhea

⇒ 2X RDA x 2 weeks

> 6 months	10 mg	RDA
< 6 months	5 mg	Diarrhea

→ Acrodermatitis enteropathica ←

↓ zinc deficiency

cause: growth retardation

hypogonadism

anorexia

alopecia

acral dermatitis

acrodermatitis enteropathica



behavioral changes

infection

pregnancy: preterm, IUGR, NTO

→ SAM ←

* children - 6-59 months

↓ wt for height (Acute malnutrition)

below -3 SD (SD on Z score)

2) visible severe wasting

3) presence of b/l edema

4) mid upper arm circumference

Height for age (chronic malnutrition)

wt for age (Both Acute & chronic)

Management:

SAM complication management:

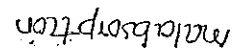
Sugar: WHO blood $>54 \text{ mg/dL}$ (mmol/L) $<3 \text{ mmol/L}$

H Hypothermia (WHO rectal temp $<35.5^\circ\text{F}$ / $<35.5^\circ\text{C}$)

I Infection TB, Malaria, UTI

E Electrolyte hypokalemia





exemplum

စာအုပ်အမျိုးအစား

They have G/T abn

overlap of fingers

Rocky Bottom Jet

But comment history
after down's synd

Relative to maternal age

→ Edward (Trisomy 18) ~~the~~ →

aplastic

Holo prosoctophaly

fused, lateral ventricle

fused frontal lobes

→ PATANS →

Polydactyl

cleist up, cleist palat

hypotetorism

→ Patan thronomy 1813 →