Neonatology

Neonate: 1st 4 weeks of life

Neonatal Period

Early
1st 7 days of life

Cause of death:
Prematurity
Infections -Septis

Late
7-28 days of life

Macrosomia: Birth wt >4 kg

Term Baby = 37-42 wks
Preterm <37
Post term >42 wks of Gestation

Percentile:

Lubchenko
arranged all newborn babies in order of wt:

3 wt

4 kg

90th

10th

8.5 kg

Gest.

26

37 wks

Small for Gest. Age = on 37 wks, baby wt <1.5 kg.
Large for Gest. Age = on 37 wks, baby wt >4 kg.
Appropriate for Gest. Age = baby should be btw 10th-90th percentile

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Large Gestational Age

 MCC of large baby is Constitutional

 MCC of short stature is Constitutional

 Constitutional: It is physiological delay, they can grow in late period.

 MCC of delayed puberty: Constitutional.

 Fontanelles

 All term babies have 6 fontanelles

 1. Ant
 1. Post
 2. Mastoid
 2. Sphenoidal

 Post fontanel:

 1) It closes at birth

 Only open in 3/4 of babies

 Ant fontanel:

 Diamond shaped

 - Diagonally 2.5 cm
 - It is open at birth
 - It is at bever
 - It is pulsatile
 - Closes at 18-24 month of life

 Causes of delayed closure of Ant. fontanel:

 MCC is Rickets, Hypothyroidism, Down’s syndrome,
 Hyperparathyroidism, Child abuse, traumatic skeletal dysplasia.
Early:

- CRANIOSYNOSTOSIS
  - Premature closure of skull sutures (one or all sutures)
  - Complications: Microcephaly
    - TICI cause: 2° optic atrophy
      - If one (coronal suture closed) is closed early then
      - Skull always grows into fuse suture
      - Cosmetic problem: abnormal skull shape
    - Rx: Cranietectomy (cutting of skull)

**4 Syndromes as & Craniostenostosis:**

1) APERT - AD
2) CROUZON - AD
3) LE CARPENTER - AR
4) PFIFER - AD

In which condition Ant. fontanelle is bulging:

- In ↑ ICT: Not pulsatile
- Infant is crying: Ant. fontanelle is bulging & still pulsatile
- Dehydratd: Ant. fontanelle is depressed

A child has 14 days old & has hypotonia, hypothermia & has umbilical hernia, constipation & also has ant. fontanel & all sutures are widely opened. He has Jaundice & that prolonged & exaggerated physiological Jaundice

**As is Cong. Hypothyroidism**

**Cong. Hypothyroidism:**

McC. preventable cause of mental retardation Cong. Hypot.

Early As is & Early thyroxine supplementation prevents

MR caused by Cong. Hypothyroidism.
McC of Cong. Hypothyroidism (overall):
- agenesis (85%) or dysgenesis of thyroid gland

- Every baby in world should get TSH screening test.
  Ideally we do T4, T3, TSH.
  In world, mostly TSH is done.

Q: Best time to do TSH - after 48 hrs to 6 days
- Cord Blood (6-9 days, 3-5 days)
  a) 48 hrs
  b) 72 hrs
  c) 24 hrs

- In all babies, in first TSH surge
- Physiological surge in first 48 hrs
- Physiological TSH 48 hrs surge

> Phenylketonuria:
  phenylalanine \(\xrightarrow{\text{Phenylalanine hydroxylase}}\) tyrosine \(\xrightarrow{\text{Tyrosine}}\) melanin

- Mon: Deficiency of enzyme phenylalanine hydroxylase
- GF - Pigment ab
  - Blue eyes
  - Brown hair
  - Fair skin
  - AR

- Phenylalanine is accumulated & it is toxic to brain so they develop seizures, developmental delay.

Rx: Tyrosine supplementation & phenylalanine
Tyrosine is non-essential A.A. but now it becomes essential A.A. for Phenylketonuria pt's.

Phenylalanine is an essential A.A.

Afro: To keep Sr. Phenylalanine ≤ 6 mg/dl level.

→ Check phenylalanine levels to prevent MR (mental retardation).

In adults 2 phenylketonuria, Restrict of phenylalanin & Tyrosine supplement is life long.

Q: Musty odor: due to phenylacetic acid

AllMS Nov 2010

Q. Rx of PKU, first step:

To reduce the substrate (phenylalanine levels ≤ 6 mg/dl) of the enzyme.

-- Maple Syrup Urine Disease

Deficiency of enzyme α-ketoadic Branched chain dehydrase case i.e. valine, leucine, isoleucine.

In plasma, CSF → ↑ levels of valine, leucine, isoleucine.

Rx: Restrict of these 3

[Neonatal Screening: In US, UK]

1) TSH

2) Tandem mass spectrometry – 1st most of metabolic (MS) disorders

3) Cystic Fibrosis (in west)

4) CAH (Congenital Adrenal Hyperplasia)

5) Gold screening – In India.

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For screening: Put 2 drops of blood by heel prick.

For heel prick: We do in Peripheral area.
Repeated heel prick in centre can cause Calcaneal Osteomyelitis.

→ ICMR - TMS is doing freely for 1st one lakh Population

Urine & Meconium

99% of Babies pass their 1st Urine in First 48 hrs
First Stool in 1st 24 hrs

⇒ Meconium:

Delayed meconium:
- Hirschsprung disease / Aganglionic
- Meconium ileus occurs in cystic fibrosis (8-10th)
- Imperforate Anus (Anorectal malformation)

Gold standard is to see Hirschprung is Rectal Biopsy

A 48 hr old baby has not passed meconium:
what is next to be done:
1) CFTR gene Test
2) Sweat Chloride
3) Manometry
4) Lower GI contrast study → To see Hirschprung
also Rx meconium ileus
Urine:

A baby didn't pass urine

After 20 wks of gestation - Amniotic fluid is baby's urine

Causes:

B/L Renal agenesis

↓

Oligohydramnios

↓

B/L Pulm Hypoplasia - POTTER'S SEQUENCE

Boys can have Post Urethral Valves

when child is going to micturate PUV obstruct midstream

Ixc for PUV → MCU (micturating cystourethrography)

--

Hydronephrosis

Thick wall bladder

Diverticuli

In MCU:

→ Thick wall trabeculated bladder & diverticuli

→ B/L hydronephrosis

→ Dilated Post urethra

Rx - Fulgurate of the Postuvalves

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Anthropometry

Term Baby:

- Length = 50 cm
- Head circumference = 35 cm

In newborn baby, HC > CC = 3 cm

In a baby, HC > CC > 3 cm - ase
  a) Hydrocephalus
  b) Asymmetric IVGE

CC < HC - Newborn
CC = HC - Around 9-12 months
CC > HC - At 1 year of life

Upper Segment (US)
Distance from head to pubic symphysis
Lower segment
Total length = US

In newborn = US : LS = 1.7 : 1
At 10 years = US : LS = 1 : 1
Adult = US : LS = 0.9 : 1

Thyroxine - helps for skeletal maturation
Bone ossification

In hypothyroidism, baby from 6 yrs he has hypothyroidism & 10 yrs he has US : LS = 1.4 : 1 which means short stature.

Hypothyroidism causes disproportionate short stature.
Temperature Regulation.

All newborn have non-shivering thermogenesis mediated by brown fat.

Brown fat (+) in - nape of neck around kidneys & adrenals interscapular area.

Fat liberates FA oxidaton → liberate heat. By blood vessels it is transported to various parts.

Core Body Temperature:

Temperature of middle ear, esophagus.

 spiritually

Core Body Temp. = 36.5 - 37.5°C

Term baby has warm & pink palms & soles

of baby is preterm - brown fat less so risk of hypothermia.

When we or baby exposed to cold

1st baby - cold stress - corresponds to temp 36-36.5°

dvelop

Hypothermia < 36°C

Severe Hypothermia < 32°C.

In premature baby:

By mother, to prevent hypothermia of baby she do kangaroo mother care. & out clothes in blow baby & mother. Baby kept on mother's chest in clothes.

A. All are following are components of KMC except:

a) Skin & skin care.

b) Early discharge & follow up.

C) Supplemnary nutrition.

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AIMS NICU Protocol:

1) Skin to skin contact

2) Early discharge & follow-up etc.

Hypothermia:

- Mechanism of Heat Transfer in overhead radiant warmers → Convector

  It is a → Convective warmer

Complications of IUGR Baby:

1) Most of babies have congenital, chromosomal or TORCH anomalies

   Trisomy - 18, 13, 21

2) Perinatal depression → Asphyxia

3) Meconium Aspirate Syndrome (MAS)

4) NEC of death in IUGR babies: Pulm. Haemorrhage
   Suddenly they bleed

5) Persistent Pulmonary HTN of Newborn due to Hypoxia Causing Persistent Fetal Circulation

6) Hypothermia, Hypoglycemia, Hypocalcemia due to limited reserve

7) ATN

8) Polycythemia (Premature have anemia due to prematurity & IUGR have polycythemia)

9) Thrombocytopenia

10) Neutropenia

- In 1st Trimester of pregnancy, our cells & brain are developed.
1. Chromosomal problem
   a) Maternal problem (APH, PPH)

2. Cell B
   \[ \text{N} \downarrow \text{N} \]
   \[ \text{BRAIN / Head sparing} \]
   \[ \text{Symmetric IUGR} \]
   \[ \text{Asymmetric IUGR} \]
   \[ \text{HC > CC > 3 cm} \]
   \[ \text{(Liver - small etc)} \]

3. To differentiate Symmetric IUGR & Asymmetric IUGR:
   \[ \text{Ponderal Index} = \frac{\text{WT (gm)}}{\text{LENGTH (cm)}^3} \times 100 \]
   \[ \begin{align*}
   <2 & \quad \text{Asymmetric IUGR} \\
   \geq 2 & \quad \text{AGA / Symmetric IUGR}
   \end{align*} \]

   Birth wt = 2.5 kg; length = 50 cm

   \[ \text{PI} = \frac{2500 \times 100}{(50)^3} = 2 \]

4. Full term, small for date babies are predisposed to
   a) Hypercalcemia
   b) CNS infection - Common in Preterm
   c) Hypoglycemia - Limited reserves
   d) PDA - Common in preterm

5. Notes from Jain Stationery
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Neonatal Reflexes

1) Moro's Reflex:
   1) Adduct \(^\text{\textsuperscript{1\textdegree}}\) at Shoulder Joint
   2) Extens \(^\text{\textsuperscript{\textdegree}}\) at elbow joint
   3) Opening of Fingers
   4) Adduct \(^\text{\textsuperscript{\textdegree}}\) at shoulder joint
   5) Flexion at elbow joint

   Appears as early as 28-32 weeks of gestation
   Adduct \(^\text{\textsuperscript{\textdegree}}\) phase seen at 37 wks of gestation
   In Preterm: - adduct \(^\text{\textsuperscript{\textdegree}}\) & extens \(^\text{\textsuperscript{\textdegree}}\) is alone.
   - Disappears at 3 months of life

   Persistence beyond 6 mon is ab(\textsuperscript{\textdegree}) Moro Reflex:
   It means there is brain injury
   eg: Cerebral Palsy.

Child has asymmetric Moro's: - Causes:
1) Erb's Palsy (Brachial plexus injury)
2) # humerus, clavicle #
3) Hemiplegia

1 yr baby left handed -> due to right side hand weakness

Handedness comes at 3 yrs always.
Early hand preference means -> Hemiplegia

An exaggerated Moro's seen in:
- HIE (Hyponic Ischemic Encephalopathy) - Brain Injury occurs during hypoxia.
2) **Grasp Reflex:** - By stimulating medial aspect of palm.
   - Oneset - 28 wks of gestation
   - Fully developed - 32 wks of gestation
   - Disappears at 12 wks of age

3) **Sucking & Rooting Reflexes:**
   - Roots for nipple & starts suckling
   - Appears at 32 wks of gestation
   - Coordination appears by 34 wks of gestation

   Child can breast feed at 34 wks of gestation (imp). 1 kg baby also breast feed. Gestation is important.

**Note:**
Baby's birth wt is 1400 gm. Gestation is 31 wks.

Baby is fed by:
   a) Parenteral Nutrition (TPN)
   b) Enteral Nutrition by NG Tube, we give Expressed Mother's Breast milk
   c) Enteral + IV Fluids
   d) IV Fluids

**Note:**
Baby's birth wt is 1400 gm. Gestation is 33 wks

Baby is fed by:
   a) TPN
   b) EN by Katori / Pataday, we give Expressed Mother’s Breast milk
   c) EN + IV Fluid
   d) IV Fluids

3) Baby wt <1000 gms. Give him → Enteral + IV Fluids.
   We build up the Enteral feeds in extremely low birth wt.

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4) **Asymmetric Tonic Neck Reflex:** (ATNR)

- **Onset:** 35 wks of gestation
- Fully developed 1 mon of life
- **Duration:** 6-7 mon
- **Side of Occiput is left:** Flexed
- **Side of Face is extended:**
- Disappears at 6 mon of life

**Child Roll on Bed (Supine to Prone & Prone to Supine)** at 4-6 mon of life.

5) **Symmetric Tonic Neck Reflex:** (STNR)

- **If you Extend the Neck:** Tone ↑ in upper limbs —
- ↓ in lower limbs
- **Flex the Neck:** Tone ↓ in all 4 limbs

→ **Elicitable at 2 mon of life.**
→ **Disappears at 6 mon of life.**

6) **Most difficult neonatal reflex to elicit:**

a) Moros
b) Grasp
c) AsSTNR
d) STNR

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6) Parachute Reflex:
- appears around 9-12 mon of life
- it never disappears

Normal Neonatal Phenomena

1) Milia: distended sebaceous glands on face & nose.
   - they are (1)
   - white spots (2) on nose of baby.

2) Erythema Toxicum: Erythematous patches on face & trunk
   - red patches on trunk & face
   - usually seen on day 2-3 of life

3) Stork Bites: Pinkish, greyish capillary hemangiomas.

4) Epstein Pearls: Epithelial inclusion cyst on palate & prepuce
   - white spot on palate

5) Natal Teeth / Premature Teeth - lower incisor position

6) Withdrawl vaginal bleeding - on 5th - 7th day.
   - 5 yr old girl & have menstrual bleed - it is (3)
   - we get hormones from mother
   - D5-D7, they decrease

Birth wt - 2.5 kg, & on 4 days = 2.2 kg, & on 7 day = 2.5 kg

All babies loose wt because their tubules are not developed much

Term baby loose 10% & regain birth wt 7-10 days

Preterm baby loose more (15%) of birth wt &
   Regain almost 1 week

Term

\[\text{10\%} \quad 7-10 \text{ days} \quad \downarrow \quad 15\% \quad \text{Preterm}\]
All babies lose wt by physiological diuresis. K gain wt from ECF.

What is not (a) in a newborn
a) Proteinuria
b) Glucosuria
c) wt loss
d) Bacteriuria
In extremely preterms
- Protein loss
- Glucose loss
- but Bacteria in urine is always Ab (a)

SEPSIS
Neonatal SEPSIS

SEPSIS: defined as c/e of sepsis + bacteremia.
It is of 2 types:

Early sepsis
Risk factors:
- Non-antimicrobial fever <4 days
- Maternal fever

Late sepsis
> 72 hrs
Risk factors:
- RIF→ nosocomial, hospital
- Acquired
- Foul liquor: most infect, are

Organism:
- Group B Streptococci
- E. coli (mc)
- New Group B Streptococci

Incidence is falling due to prophylaxis

Doc: Ampicillin + Gentamicin
which of the following can YOU Infect in ICU?
1) using prophylactic antibiotic inj.
2) using high grade antibiotic (meconam & vancomycin)
3) Hand washing

MCQ of sepsis in developing India - Klebsiella > Staph. Aureus

World - E. coli

Neonatal meningitis

Organism:
Group B, ß Streptococci; E coli

Doc - Ceftriaxone (3rd gen cephalosporin)

Late sepsis -

meningitis mc in late sepsis.

we always do Lumbar puncture & CSF in late sepsis.

Rx - Cefotaxime + amikacin

Gold Standard ABIS for Sepsis is

Sepsis screen for early ABIS:

1. TLC - <5000/cumm 0s > 20,000
   In severe sepsis - Leucopenia also may occur

2. Absolute Neutrophil count - <1500/cumm

3. Peripheral smear for band cells - > 20%
   [Immature neutrophil (IT ratio)] (IT > 0.2)
   & Toxic granules

4. Micro ESR (6mm - 3 days) - >15mm fall 1st hr
   1st 3 days of life fall is <3mm (0)
   In 1st hrs of fall is >15mm 5% of sepsis
5) CRP +ve
   Procalcitonin
6) Lumbar puncture - lately done in late sepsis
7) Chest X-ray - Pneumonia can have bacteremia
   Meningitis also have bacteremia
   Pneumonia seen in sepsis.
   Old culture is more imp.
8) Ratio of Blood to culture medium

ANC means:
   a) Neutrophils + Band cells
   b) N - Band cells
   c) N + Band/2

In sepsis - Duration of antibiotics:
   Give to Newman
   Bacteremia + → 10 - 14 days
   Meningitis + → 3 wks (21 days)
   Arthritis osteomyelitis → 4 - 6 wks

Q: Lab finding in neonatal sepsis is all except:
   1) ↑ CRP
   2) Leucocytosis
   3) ↓ ESR, ↑ ESR
   4) Toxic granulated multilobulated nuclei

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Fetal Alcohol Syndrome (FAS)

C/F:
1) Skin Folds at corner of eye
2) Low nasal bridge
3) Small head circumference - microcephaly
4) Small eye opening = Small palpebral fissures
5) Flat mid face (or mid facial or maxillary hypoplasia or small mid face)
6) Short nose
7) Indistinct or Absent Philtrum & thin upper lip
8) They have septal defects in heart like ASD, VSD

✓ All are associated to FAS except:
a) microcephaly
b) Overgrowth
c) Flat Face (Flat mid face)
d) Small palpebral fissures

PREMATURITY

Respiratory

Complications:
1) Most Imp: Complications is Respiratory is RDS
   30% of mortality in Neonates in India = RDS

b) Chronic disease - Bronchopulmonary dysplasia - refers to O₂ dependence at 4 wks of life
   Home O₂ Therapy

c) Apnea - Cessation of Respirations > 20 sec in newborn or at any period it is an "Central cyanosis/
   Bradycardia (even it is of 2 sec"
All Newborns are Nose breathers (obligate)

Apnea

Central Obstructive mixed

Occur in Asphyxia Premature

Q) A full term newborn baby having episode of cyanosis which are worse when he is attempting feed but he seems better (pink) when is crying most imp diagnosis:

a) Choanal atresia, B/L Post Choanal atresia which becomes symptomatic
b) Tricuspid atresia
c) Pulmonary atresia
d) Laryngeal atresia

Of all Choanal atresia - is asymptomatic

The type of apnea in Preterm is mixed

⇒ Apnea of Prematurity: Apnea because of Prematurity

< 28 wks: 100% Risk to develop Apnea.

Onset of apnea - 2 in 1-2 days
≠ never > 7 days

4 month old, apnea occur - It is not due to prematurity

It is may be due to meningitis, do L.P etc.

Rx: 1. Nasal CPAP 2. By prongs (Nasal prongs)
2. methylxanthines: Aminophylline
The loading dose of Aminophylline is

5-6 mg/kg

TID

Maintenance - 1-2 mg/kg every 6-8 hrs

dose

a) 50-75 mg/kg
b) 0.5-10 mg/kg
c) 2.0-3.5 mg/kg
d) 5-6 mg/kg

→ Doc for Apnea of Prematurity: Caffeine Citrate

→ loading dose of Caffeine Citrate = 20 mg/kg

maintenance dos: 5 mg/kg given OD

PREMATURITY

CNS

Intracranial haemorrhage:

All haemorrhages are common in preterm except

Subarachnoid.

All preterm has fragile capillaries in germinal

day layer ruptures & bleeds.

Q: → Preterm baby became Suddenly pale, shock, Ant. Portosel

belging & has seizures

→ Diagnosis: Intraventricular Haemorrhage (massive)

(bleeding in ventricles)

children < 1500 gm - Incidence is 30%

Next Tx for seizure in newborn: USG (Transfontanelle

because + of window (AF)
Can a Term Baby have IVH?

In Breech presentation, Intracranial hemorrhage may occur. Later they develop MR.

Instrumental delivery can cause IVH.

Complications:

Asphyxia:

Baby is Preterm

TERM

Parasagittal Injury

Status Marmotatus

Diffuse Neuronal loss

Later in life, he develops Spastic Quadriplegia

Periventricular leukomalacia

Worst

More developmental delay, MR

UL are strong & LL are weak (UL > LL)

Commando-like crawl

He drags leg

Spastic diplegia (UL < LL) - Best

Type of Cerebral palsy

MC sequelae of periventricular leukomalacia in preterm infants is spastic diplegia.

MC: Loss of white matter vs. gray matter - MR.
**Hypoxic Ischemic Encephalopathy (HIE):**

**Sarnat & Sarnat - Staging**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>Sensorium Hyporeakitec lethargic coma</td>
</tr>
<tr>
<td>II</td>
<td>TONE</td>
</tr>
<tr>
<td>III</td>
<td>DTR</td>
</tr>
<tr>
<td>IV</td>
<td>MORAS</td>
</tr>
<tr>
<td>V</td>
<td>PUPILS</td>
</tr>
<tr>
<td>VI</td>
<td>SEIZURES</td>
</tr>
<tr>
<td>VII</td>
<td>EEG</td>
</tr>
</tbody>
</table>

**Doc:** - Phenytoin

**Neurodevelopmental Outcome:**

**Prematurity**

**CVS**

1. Hypotension caused by immature immaturity
2. PDA

**PDA**

- Common in preterms because of liberation of PG's
- In Term baby's - PDA due to Rubella vessel wall defect

<table>
<thead>
<tr>
<th>Preterms</th>
<th>Term</th>
<th>Sy</th>
</tr>
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<tbody>
<tr>
<td>↓</td>
<td>↓</td>
<td></td>
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</tbody>
</table>
| Asphyxia | NSAI
| NSAI    | [IV] |
| Indomethacin | Rubella |
| ↓     | O |
| PG's   | → PDA |

- PDA vessel wall defect
- Sympathetic Hyperactivity
- Ligation: ductus arteriosis

**All 8 Terms baby's are surgically Inserting coils & close it Operated.
As Compared to IBUPROFEN, Þ INDOMETHACIN
because it is IBUPROFEN is less nephrotoxic

→ Clinically: PDA Baby:

Presents Þ 6-10 wks of CHF
Can also present Þ Preterm newborn Þ Failure to
wean off from ventilator
If you wean off Þ $O_2$. L & $PCO_2$. **

Not seen in PDA:

- $CO_2$ wash out Þ Narrow pulse press.

→ O/E - PDE

1) Child has bounding pulses Þ Wide pulse press.

2) Continuous murmur Þ Machinery murmur (SBP < 80) is at upper left sternal border

PREMATURITY

· GIT

1) Necrotising Enterocolitis:

Child has limited reserves:

- Hypothermia, Hypoglycemia, Hypocalcemia

→ Child prone to infections

→ Prone to anemia, neonatal jaundice

→ Has risk of Retinopathy of Prematurity (RP) (Retinal Fibroplasia) - Back of lens - vessels are dilated, proliferative & tortuous in vitreous

→ Severe RP Þ Child have

- Trachoidal Retinal Detachment
"+" - dilatation of vessels at post-pole.

Pediatrician in a district hospital calls ophthalmologist for:

a) Newborn & respiratory distress
b) Newborn 28 wks gestation to receive high flow O2
c) Newborn & jaundice
d) Newborn birth wt 2.3 kg.

Most important risk factor of ROP - Prematurity.

ROP can cause blindness.

Best test to look periphery of retina is indirect opthalmoscopy.

We do regular indirect ophthalmoscopy; if ROP, then we do photocoagulation.

When will you send the child for first visit?

AAP first visit:

<table>
<thead>
<tr>
<th>Gestation</th>
<th>Interval weeks</th>
</tr>
</thead>
<tbody>
<tr>
<td>28 wks</td>
<td>32 wks → 4 wks</td>
</tr>
<tr>
<td>29 wks</td>
<td>33 wks → 4 wks</td>
</tr>
<tr>
<td>30 wks</td>
<td>34 wks → 4 wks</td>
</tr>
</tbody>
</table>

So after 30 wks incidence of ROP.

After this 2 weeks baby sent to ophthalmology.

Until retina looks like mature

Rx of ROP:

1) Photocoagulation
2) Cryotherapy
3) In stage V - Retinal reattachment sx
Q5 Most imp. factors of retrolental fibroplasia

a) Prolonged labour
b) Intrauterine infection
c) Meconium aspiration

- Low birth wt.

\[
\begin{array}{|c|c|}
\hline
\text{wt} & \text{ROP Risk} \\
< 1 \text{kg} & 80\% \\
1-1.2 \text{kg} & 65\% \\
\hline
\end{array}
\]

Rx for Threshold ROP = Laser Photocoagulation

PREMATURITY

\[\rightarrow \text{Necrotising enterocolitis: (NEC)}\]

Preterm \& Abdominal distensio

Risk Factors:

- Me \rightarrow Immature Gut

\[\text{Ischemia} \downarrow \text{maternal cocaine}\]

\[\text{Sepsis toxin} \rightarrow \text{Immature Gut} \uparrow \text{Top Fed}\]

Breast milk protects against NEC

Staging of NEC:

Modified Bell's staging NEC

\[\text{Suspected NEC} \rightarrow \text{distensio, ileus, occult blood stools}\]
a → Gross bid in stools, Focal pneumatosis

II

<table>
<thead>
<tr>
<th>a</th>
<th>DEFINITE NEC</th>
</tr>
</thead>
<tbody>
<tr>
<td>b</td>
<td>Diffuse Pneumatosis, HMT-H-Hypotonuria, M- Metacidosis</td>
</tr>
<tr>
<td>T</td>
<td>Thrombocytopenia, Portal venous Gas</td>
</tr>
</tbody>
</table>

III

<table>
<thead>
<tr>
<th>a</th>
<th>Impending perforation (Air cause Perforation)</th>
</tr>
</thead>
<tbody>
<tr>
<td>b</td>
<td>Perforation causing Peritonitis, Shock, Pneumoperitoneum (air in peritoneum)</td>
</tr>
</tbody>
</table>

Q X-ray appearance of NEC:

Air goes into wall of gut → Pneumatosis Intestinalis

Q Posterior Gas seen in Stage IIb of NEC

Q Newborn has air around portal vein or air in et-hyochordium basis → NEC

Aims

Features of NEC are all except:
1) Abd. distensio
2) ↑ Bowel Sounds, ↓ Bowel Sounds
3) met-acidosis
4) pneumoperitoneum

Aims.

q Probiotics are useful in:

1) NEC
2) Breast milk jaundice
3) Hospital acquired pneumonia
4) Cholera → Doc. Tetracycline/Doxycycline

Probiotics are commonsels
Can we prevent NEC:

1) Give maternal Betamethasone

2) Trophic Feeds: These feeds are not nutrient
   → Through NG Tube, minimal 10-20mL/kg/day
   → Breast milk
     - 500mL Breast milk for ½ kg Baby
     - This is for local effect on gut mucosa.

3) Probiotics Prevent NEC:

WHO Recommendations for Probiotics:

1) Antibiotics Associated Diarrhea:

2) Rotavirus
   - Saccharomyces Boulardii
   - Lactobacillus rhamnosus

3) Probiotics known to prevent NEC
   - Lactobacillus acidophilus
   - Bifidobacterium infantis
     - Given for VLBW
     - To prevent NEC

Best Study design → metaanalysis & systematic review.

- metaanalysis
- Systemic review

- (R)RCT → Triple Blinding
- Case-Control
- Case Series

- C Problem
   → E out problem
Rx of NEC:

1) NPO (Nil per oral) & TPN
2) Cefotaxime & Vancomycin (E. coli cause perforated peritonitis) + Metronidazole (for anaerobic organisms)
3) Stage III → may require Sx

Newborn

- Distended abdomen & B/L gas shadows under diaphragm: Probable perforation
- Gastric perforation

NEC - stage III-b
- Duodenal atresia
- Hirschsprung disease

A child with NEC has perforation & poor general condition is treated & death of shock Stage III-b + perforation peritonitis

- Conservative Rx only
- Frank drain & glove + vancomycin (cefotaxime) + metronidazole

- Laparotomy & resection + anastomosis → cause death on table
- Extra corporeal membrane oxygenation (ECMO) → artificial lung

- Of in lung, PPHN → Diffusion not done in lung then go for ECMO
- Massive hemorrhage in lung (eg: Wegner's granulomatosis)
  → Put him in ECMO

New Delhi Super Bug

NDM-1 = New Delhi metallo-β-lactamase 1

Enzyme which makes E.coli resistant to Carbapenems
Gene = Carbapenem Resistance gene Bla Non-t

Last drugs to use in E.Coli

Tigecycline
Colistin – (Nephrotoxin)

Antibiotics susceptible for E.Coli isolated from
UK, Chennai & Haryana.

RESPIRATORY DISTRESS
IN NEW BORN

Causes of RDS:

- Any preterm → RDS (MAS – rare)
- In term baby → Transient Tachypnea of Newborn (TTNB)
- Meconium Aspiration Syndrome (MAS)
- In Post term → MAS – common

Silverman Anderson Scoring for RDS

Score: upper chest lower chest xiphoid nasal grunt

Retract
Retract
Retract
Dilata

0: Synchronized None None None None
1: Lag on Inspiration: Just Visible: Minimal Minimal

2: See – Saw Marked Marked Marked
Paradoxical

Score > 6 = Impending Resp Failure

Downe score:
1. Resp Rate
3. Retract
4. Grunting
5. Air entry.

"Grunting is always severe distress"

A baby has marked flaring & marked grunting & NO retract

Score: 2 + 2 + 0 = 4

Diaphragmatic Hernia

8/10 by:
Scaphoid Abd

Barrel shaped chest

Trachea shift to opp. side (mediastinal shift)

Heart is also pushed to opp. (apparent dextrocardia)

Embryologically diaphragm develops from Septum Transversum. 2 canals & are pleuroperitoneal canals. 2 of these canals close at 8 wks of gestation. We do mostly left pleuroperitoneal canal fail to close. So most C diaphragmatic hernia is on left (85%) post-lateral

85% Left posterior lat. Bochdalek

C < 5% B/L

(\(R\)) sided hernia diagnosed late (6 mon)

\[\text{The baby is in distress because on the left side there is no place to develop so he has (1) pulmo hypoplasia}\]
Q: What is the cause of death in Congenital Diaphragmatic Hernia?

Pulm. Hypoplasia.

1. Bag & mask ventilation is absolutely C/I. We have to do electively intubate because if you do bag & mask ventilation, air goes into gut & gut is more distented so more ROS.

2. After Bag & mask ventilation, you should put NG Tube to decompress gut. (If you do Bag & mask ventilation.)

3. After Intubation, Heart is more shifted to (R) side. It means Intubation is done wrongly into esophagus. In this situation, you have to be intubate.

Most imp. prognostic factor in Cong. diaphragmatic hernia is

4) Pulm. HTN \( \rightarrow \) PPHN \( \rightarrow \) Hypoxia

\[ \text{Need ECMO} \]

2) Sildenafil used in Newborn in PPHN because it release NO - Vasodilator (Pulm.)

3) Bosentan is used for Pulm. HTN.

4) Survival rate is 60-70%.
Tracheoesophageal Fistula

Type C' TEF is MC in Babies
Esophageal atresia - distal fistula

We see
Dribbling saliva: he can't swallow
Aspirat'n pneumonia: he aspirate gastric juice in lungs

Baby is cyanosed & frothing - A5is TEF.

RDS

Disease of prematurity
Degree of Prematurity Risk
< 28 wks 60-80%
> 34 wks <5%

MC of Resp distress in Newborn - RDS:
Infant of Diabetic mother is also risk of RDS (Term)

Surfactant:

1. 65% of Surfactant - Phosphatidyl Choline (Most imp component)
2. Phosphatidyl Inositol
3. Phosphatidyl Glycerol
4. Other protein
5. SP - A
6. SP - B
7. SP - C
8. SP - D

Surfactant is homogeneous at 20 wks of gest
It appears in amniotic fluid in 28-35 wks
>95% of Surfactant seen in >35 wks of gest

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26 wks pregnant & bleeding we give
12 mg IM Betamethasone 24 hrs apart;
2 doses for baby lung maturer

Beneficial act of Betamethasone is 24 hrs to 7 days
so when Tocolytics are given

Betamethasone ↓ risk of Periventricular leukomalacia
where as Dexamethasone ↑ so Dexam is not
preferred.

Betamethasone [maternal]:
- INEC
  - ↓ Intra cranial haemorrhage
  - ↓ Neonatal mortality
  - ↓ Periventricular leukomalacia

28 wks pregnant, Single dose of Betamethasone is
given, next dose is not given in 32 wks because

Double course of Betamethasone not preferred
because there is development of behavioural
abnormality like attention deficit so single
course is preferred.

For see the lung maturity of baby we see
Leucithin/Sphingomyelin ratio
L/S > 2 = Lungs are mature

In infant of diabetic mother, L/S > 3.

X-ray appearance of RDS (A11)
2) Ground glass
3) White-out
4) Reticulonodular shadows
5) Air-Bronchogram (because there is a alveolar pathology)

1) RDS occurs 19 hrs of life

2) RDS never occur in IUGR because develop stress release Steroids so no Surfactant

Surfactant ↓ Surface tension & prevents collapse of alveoli during expiration.

If no Surfactant, alveoli collapse at end of expiration.

Rx of RDS:

1) nasal prong - CPAP: for mild - moderate
   - FiO₂: 50 - 70% → room 21%
   - PEEP: 5cm H₂O (Preterm requires PEEP)

2) Surfactant: when there is severe distress.
   By IntraESCO route
   Route: INSURE - Intubate → give Surfactant → extubate

a) Survanta - Bovine Surfactant
b) Curosurf - Porcine Surfactant

Giving Surfactant prophylactically to ELBW is controversial.
Preferred Surfactant is Rescue Surfactant.
All occur in RDS except

1. Cyanosis
2. Occurs in preterms
3. More in IOM
4. Treated ≥ 100% $O_2$ - because if you give this baby develop Retinoblastoma Optic atrophy

For Resuscitation - 100% $O_2$ is given. (Ideally)

a) It is outside by mouth to mouth
   b) Room air etc

All 5.
A 32 wk preterm baby is born = emergency CS. Secn. Child develops grunting Respirat° ≤ Resp. rate 70/min. Best mx of choice

a) Surfactant Therapy & mechanical ventilation
b) CPAP
   c) Humidified $O_2$ by hood
   d) Mechanical ventilation

IN RDS, FRC ↓; By CPAP → FRC ↑

All is true about CPAP except
1. Initiated at $Fio_2$ 0.5 - 0.7
2. CPAP IMPROVES LUNG COMPLIANCE OR VOLUME, FRC
3. Used in apnea of prematurity
4. CPAP prophylactically in ELBW XXX

Prophylactic CPAP in newborn are not given because it develops Pneumothorax

Q) A newborn term Female baby at birth wt 3.5 kg
Baby is on mech. ventilat & is given surfactant but the condition deteriorates & hypoxemia increases. There is a H/o full term sibling dying in one week & similar complaints. Echocardiogram bid cultures are normal. Most likelyosis is.

**Neonatal Pulm. Alveolar Proteinoses.**

- Neonatal Pulm. Alveolar Proteinoses:
  - AR
  - Mutation in Protein B/c /GM-CSF
  - Rx: lung transplant

**Transient Tachypnea of Newborn:***

Risks factors:

1. Term baby born by C-Sect - Baby develops inlet-lungs (fluid into lungs)
2. Precipitous labour
3. Excessive sedation to mother
4. Macrosomia

- It is a transient condition & resolves in 48-72 hrs
- It is benign condition
- O$_2$ requirement of this baby less & FiO$_2$ < 0.4
- Baby doesn't require mech. ventilat (because it is benign condition)

In X-ray: In (R) lung - Horizontal fissure becomes prominent - Specific
Meconium Aspiration Syndrome

Membrane is rupture, meconium like liquid & baby is not born.

Meconium is a marker of perinatal hypoxia

Perinatal hypoxia

Glottis is open

$\uparrow$

In newborn

Parasymp stimulation

$\downarrow$

Aspirates meconium

Anal sphincter relax

Peristalsis stimulates

Meconium aspiration is common in post terms because of uteroplacental insufficiency (placenta is calcified).

Meconium

1) Physical

Chemical

Biological

Problem

Block bronchus

meconium is a good culture

cause Ball valve mechanism

medium so

it causes

Sepsis

(Infected)

In about 20-30%

air leaks like

Pneumothorax

It is like chemical irritant so it can cause

Chemical Pneumonitis
Best Resp stimulus for baby - Suct\n
Meconium Stained Liquor

↓

Baby Born

↓

VIGOROUS

\text{Resp Effort (By cry we know this)}

\text{HR > 100/min}

We count HR for 6 sec in a child (6 sec = 10)

\begin{itemize}
  \item NO \text{ if baby is limp or Apnea or}
  \text{HR < 100/min}
\end{itemize}

YES

Transfer baby to Intratracheal Suct\n
(Indicate?)

mother

IN (a) Baby we do

\begin{itemize}
  \item T
  \item A \text{ Suct\ through mouth > nose}
  \item B
  \item C
\end{itemize}

A baby born by vaginal delivery has Resp Distress. One of:

- He has hyperinflated \( \text{t} \) upper lobe \& \( \text{t} \) mediastinal shift
  - a) \text{α-1 antitrypsin deficiency} \text{ occurs in old age people (we see emphysema)}
  - b) \text{Transient Tachypnea of Newborn}
  - c) \text{Pneumonia}
  - d) \text{Cong lobar emphysema} \text{ occurs hyperinflated in \( \text{t} \) upper \& middle lobe. In cystic}

\text{Notes from}

\text{Jain Stationery}

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Jaundice

Haem → metalloporphyrins (Chole)

Haemoglobinase → END TIDAL CO2 is marker of

1 mole of CO2 is liberated → Hemolysis

1 mole Haeme liberates 1 mole of CO2

BILIVERDIN (B)

B. Reductase

BILIRUBIN → 1 gm of Hb = 34 mg of Bilirubin liberated

→ Albumin is plasma protein. Bilirubin binds to Albumin

→ Upptake by Ylgrad → 1 gm of albumin binds to 8 mg of Bilirubin

→ Bilirubin CROSS BBB & cause KERNICTERUS

→ In adults, Ammonia CROSS BBB

→ In baby in 1st 2 wks of life

Excret by Urine

→ Unconjugated Unbound bilirubin crosses BBB

→ A term healthy baby (3kg) → don't develop

Kernicterus (even at 25 (8x3:24)

→ A prem & sick child can go to easily

Kernicterus even at low levels of Bilirubin

→ Incompatibility

Put him on Phototherapyl

→ There is no threshold for phototherapy

Hb of Newborn = 16-18 mg/ dl

Physiological Jaundice:

1) Heat ↑ (because Hb ↑)

2) Rapid Turnover of RBC (Fetal Hb life span is less)

3) Deficiency of Y-Ligand

4) Deficiency of UDP-glucoronoyl Transferase in newborns
6) Increased enterohepatic circulation

Kramer Zones: Given by Kramer

Dermal zone - Bilirubin (mg/dl)

1 - 5
2 - 10
3 - 12
4 - 15


Physiological jaundice: Physiological jaundice in terms

Peak in 3 days up to 12 mg/dl

Term

Peak on D5 up to 15 mg/dl

In Preterm:

Peak on D5 up to 15 mg/dl

(2 wks)

(4 wks)
Pathological Jaundice:

- Palms & soles are yellow

Risk factors:

MCC of Pathological Jaundice:

1. RH Incompatibility > ABO Incompatibility
   - O - mother
   - A/B - Baby

2. RBC membrane defect like Spherocytosis, elliptocytosis

3. RBC enzyme defect like G6PD def, Pyruvate kinase def.

Jaundice in 24 hrs of life is always abn

- Severe haemolysis (sclera of baby - yellow)
  ↓
  Rx - Exchange transfus

→ RH+ mother, Previous preg./abort

u) Coni-inoui - from maternal side
   - (cord bid Hb > 18-20 ml/dl)
   - Samples sent to lab are
     1. RH status
     2. Hb [in haemolysis, Hb ↓, Bilirubin ↑]
     3. Bilirubin
     4. Direct Coombs test in baby (Indct - in mother)
     5. Peripheral smear for haemolysis

Indications for exchange transfus at birth:

- Rh+ve Baby, Hb - 10 gm/dl, Bilirubin - 5mg/dl
- DCT+ve, Peripheral smear for haemolysis +ve

Indication of severe haemolysis - R. phototherapy

Jaundice is pathological:

Any Jaundice > 20 mg/dl is always abn
Persisting Jaundice
7-10 days - Terms 
>14 days in Preterms Ø is also AbØ
Jaundice ± in 1 day of life.

=> Phototherapy:

a) Principle: - Phototherapy converts less soluble to more soluble bilirubin.

b) It is a very specific blue light - 425 - 475 nm

c) It is a specific lux 6 - 12 μW/cm²/s (lux is a
spectral radiance of incidence light)

d) NOF doesn't affect the efficacy of phototherapy.

a) Type of lamp
b) Skin pigmentation --
c) Spectral radiance of light --lux

d) Initial levels of bilirubin.

=> Change the tube lights every 1000 hrs.

Mechanism by which Bilirubin Fall in Phototherapy:

1. Photoisomerism: - 4Z 15Z → 4Z 15E (Reversible and)
E → Z again

2. Best - Structural Isomerization

Bilirubin → LUMIRUBIN (Irreversible)

Keep child around 40cm away from light by at
which bilirubin falls at ≥ 4 - 6 mg/dl in a day.

Side effects of phototherapy:

1. Dehydration, Fever
Hypocalcemia
Whenever child is put on phototherapy we cover eyes & genitalia because phototherapy cause.

Retinal damages

Mutated in genitalia

Bile pigments in gut → cause diarrhea

Phototherapy C/I in Conjugated Jaundice becoz it can cause Bronze baby syndrome.
Phototherapy is given in uncon Jaundice.

Pathological
Breast milk Jaundice
Because of pigment in breast milk which is conjugated.

Breast Feeding Jaundice
Occurs around Day 4 of life → It occurs in D3 of life

It persists till 4-6 wks

If Temporary Interrupt is done → it disappears
Rx: Ensure Feeding

True about Jaundice in neonates is:
1. Can be seen after venouse delivery → occur hematoma → Jaundice occur

2. Physiological Jaundice is seen 6 in 1st hrs of birth (occurs in D3)

3. Conjugated Bilirubin leads to kernicterus (uncon)
d) Breast milk jaundice is max in 7 days from birth (14 days)

For all cephal hematomas -> drain should be done.

Neonate, Jaundice; clay stools. As: Neonatal Cholestasis

1) Later require Liver Transplant.

Neonatal Cholestasis:

Direct Bilirubin > 2mg/dl or Direct Bilirubin is forming > 15% of Total bilirubin.

Cause:

1) Surgical - Extrahepatic Biliary Atresia (EHBA)

4 Sx -> Kasai's Sx

- Should be done < 8wks of life otherwise

80% mortality

- Indicated for Liver transplantation in children

a) Fulminant Hepatic Failure (FHF)

b) a failed Kasai's

→ which is an ominous sign in an 10-day old newborn

a) Unconj. Hyperbilirubinemia

b) Conj. Hyperbilirubinemia -> sx has to be done (ExBA)

c) Failure to gain weight

d) Doll's eye reflex - in 1st 4wks of life

2) MCC of Neonatal Cholestasis is Medical

3) Others: Sepsis

Galactosemia

NOTES FROM
JAIN STATIONERY

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To differentiate causes of neonatal cholestasis:

whether it is medical vs surgical

1) GGT enzyme is 10 times ↑ in surgical
2) USG in EUSA - shows dilatation of intrahepatic biliary radicals

Triangular cord sign seen in USG of EUSA

HIDA nuclear sign - HIDA dye not seen in gut.

Gold standard test for biliary atresia is:

Perioperative/intraoperative cholangiography

If dye is not coming into gut then do:

Kasai x ray done MRC is also done

Liver Bx:

- 2, 4, 9, 10 protein C & S are carbohydrate in liver

Liver Bx in biliary atresia → Dilatation & proliferation of intrahepatic bile ducts.

In hepatitis: Distorted lobular architecture is seen

Specific for CMV: Intranuclear "owl-eye" type of inclusion bodies

Specific for neonatal haemochromatosis: Prussian blue stain +ve

In α1AT deficiency: PAS +ve

diastase resistant granules are seen
Galactosemia.

In Galactosemia why we get Jaundice?

Gal-1 Phosphate

Galactose 1-phosphate

Gal-1 Phosphate Transferase

UDP galactose

So Gal-1 phosphate accumulate in liver, which is hepatotoxic & this child has Jaundice, Bleeding, Hypoglycemia, G. Cataract. Galactitol cause Cataract.

Breast milk is c/I in Galactosemia (absolute c/I) because In breast milk lactose is present & damage to liver ↑.

Tx: - TMS

Rx: - Lactose free milk.

In baby, Galactokinase deficiency, is a mild disease - only cataract & NO liver failure occur.

Q: A 9 day old full term infant presented to you with fever, lethargy & increasing Jaundice. Lab result - AID sugar - longa physical exam - Hepatosp. megaly.

Total Bilirubin - 15mg/dl, Direct - Bilirubin - 7mg/dl

Liver enzymes - AST 700 ALT - 650

Bld culture +ve for Gram -ve Rod. This is classic picture of E. coli. Galactosemia prone to E coli sepis.

In Galactosemia babies Immune defect also occurs
In Galactosemia, always babies present E. coli sepsis.

→ Child of Vasanthi was weaned from breast milk on D5 of life & was started on Sugar cane juice. Child develop hypoglycemia & hepatomegaly.

Lab: Hypophosphatemia & Reducing substance in urine. Child is suffering from deficiency of enzyme Aldolase B.

As is Hereditary Fructose Intolerance (HF1)

→ Fructose + phosphate is hepatotoxic.

→ Fructokinase deficiency has no symptoms (symptoms k/A benign fructosuria (mild))

In HF1 - Jaundice & Bleeding is also seen.

→ Me of neonatal cholestasis is
a) EHBA
b) Neonatal hepatitis - CMV - TORCH Infectious

c) Choledochal cyst

d) Conjugate

→ The chief mechanism in phototherapy is
a) Photo oxidat0
b) Photo - Isomerisat0

c) Structural isomerisat0 - Luminrubin

d) Conjugate

→ Neonatal cholestasis is seen in
a) Chronic Hepatitis - > 6 months ab< in liver

b) Hepatitis B

c) Coli - 1-Pou hepatic x
8) Pregnant female & report HBsAg +ve, No Jaundice. 

Next Ix 18

HBsAg

Will become 

Baby HBsAg Carrier → develop Portal HTN

Triad of Portal HTN:
1) Ascites
2) Varices
3) Splenomegaly.

Pregnant lady & HBsAg

\[ \text{HBsAg}^+ \quad \text{HBsAg}^- \quad \text{Anti-HBsAB}^+ \]

90% Risk
10% Risk she can spread to
She can spread her baby
to her baby

To prevent vertical Transmission give

- HBIG 2 hrs & [Passive &
HBV vaccine 5 in 24 hrs [Active Immunization]

& we give 0 dose of Hep B (In our India)

→ Person pricked by HBsAg +ve Patient

vaccine 0,1,6

R U IMMUNIZED AGAINST HEP B

Anti-HBsAB titre >10mIU/ml (Recommended for Health workers, doctors, nurses etc.)

"CDo" Good - Nothing can be 
done

HBIG + HBV

Next 5-10 yrs titre should be

>100 mIU/ml
In America: Mantoux +ve \( \rightarrow \) \( \Delta^{15}TB \)

↓

CXR

→ of Pickled by HCV +ve

↓

Check HCV RNAq, PCR, (RNA Replicat^n).

↑ Increase

↓

Rx: Give Interferon

→

Health check up

Doctor

↓

According to CDC Protocol

HBEAg +ve

↓

HBV DNA load > 1000 IU/ml

↓

Cannot join SX

TORCH

0 - Others: HBV, HIV, varicella, syphilis

These are Intrauterine infections.

Occur in Intrauterine period.

Most of these infections occur during delivery.

Transmission of Rubella:

\[ \text{Mother } \rightarrow \text{Low-births delivery} \]
of mother gets rubella > 16 wks - risk of CRS is < 1%.

→ During delivery:
  → Most of TORCH Infections are asymptomatic.
  → If mother get TORCH Infection in 1st Trimester - TORCH Symptomatic
  TORCH Infection Babies also look alike - ie

Common: - Symptom
1) SGA
2) Unexplained anemia / Thrombocytopenia / rash
3) Hepatosplenomegaly
4) Cholestasis

TORCH : -
1) IgM⁺ - Ac for Infect⁻
2) Newborn +ve for IgG

If baby has 6 - 9 mon persisting IgG → shows Infect⁻ (TORCH
of IgG is from mother, titer should fall
Persistence of IgG is abn)

HIV

→ Infants cannot make Ig M - HIV & Ig A - HIV

→ Maternal Ig G - HIV can persist in baby for 18 mon
  (In Baby - Ig G persisting >18 mon - +ve HIV)

→ In New borns For ΔSis (Tests):
  a) q PCR - Best (DNA)
  b) p24 assay
  c) Culture (difficult to culture HIV)

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Risk of perinatal Transmission of HIV:

a) Out any intervention
   1) Out Breast Feeding \( (BF) \) 15 - 30%
   2) \( e \) BF \( \rightarrow \) risk by 5 - 15%

b) In intervention
   Anti Retroviral prophylaxis + 'C' sect + NO BF (in America)
   \( \rightarrow \) risk \( < 2\% \)
   Best milk is Formula milk
   \( \rightarrow \) Breast milk is whey based protein
   In India:
   - of mother not giving milk to baby, baby die due to diarrhoea.
   "NACO": Infant Feeding in HIV
   Affordable
   Accessible
   Acceptable "Mixed Feeding is bad"
   Feasible \( \uparrow \) risk of transmission
   Safe Cow milk lead to ulcers in gut
   Sustainable
   Give exclusive
   Breast milk \( \rightarrow \) Feasible Formula

\( \rightarrow \) Chorioretinitis
Intracranial calcifications are seen in
Toxoplasmosis \( \rightarrow \) CMV
CMV is most common cause of sensorineural hearing loss.

Toxoplasmosis

Calcifications are seen in
  Peri-ventricular calcification?
  Choroid plexus, caudate
  Nucleus, Subependymas
  Calcium cause
  Hydrocephalus ex-vacuo
  SNHL (+ve CMV Infected)

Hydrocephalus, Seizures
  Macrophagy

A preg lady had no complaints but mild cervical lymphadenopathy
in 1st trimester. She was prescribed spiramycin but she
was non complaint. Baby was born & hydrocephalus &
Intra-cerebral calcification. Which of these is likely cause?

1) Rubella
2) Toxoplasmosis
3) CMV
4) Herpes

Toxoplasmosis -
  → Spiramycin decrease vertical transmission
  → IgA ELISA > IgM ELISA most sensitive
  Best IgM ISAGA (Immuno absorptive agglutination assay)
  Rx: pyrimethamine + sulfadiazine

w/o F doesn't establish a diagnosis of cong CMV infection in
a neonate

1) Urine culture of CMV & saliva - Best specimens
2) IgG CMV AB's in blood
3) Intranuclear inclusion bodies in hepatocytes
4) CMV viral DNA in blood by polymerase chain react
  positive antigenemia q/PCR

PCR best test for CMV

DOC for CMV: i/v gancyclovir
Absolute Indications to Rx CMV:

1) CMV cause colitis (diarrhea)
2) CMV Retinitis

Best drug for CMV in children, severe, pregnant
- IV Ganciclovir
- Oral drug: Prodrug of Ganciclovir
- Valganciclovir
  - Same bioavailability like Ganciclovir

S/s of Ganciclovir: Anemia
  - Thrombocytopenia
  - Bone marrow suppression

Ganciclovir Resistant: Oral Foscarnet

Cong. Syphilis

Early
- Early syphilis:
  1) mucocutaneous rash / thinitis
  2) Lymphadenopathy
  3) Haematological like autoimmune anemia
  4) Renal lesions
  5) Skeletal lesions [osteochondritis, metaphysitis
    Periostitis - bone lesions]
  6) Glaucoma

Pseudoparalysis of parrot: sun in syphilis
  - Generalized tenderness
  - Hypokalemia, septic arthritis

"AFP is diagnosed if we rule out pseudoparalysis."
Late Syphilis:

Hutchinson's Triad:

1) Hutchinson's Teeth / mulberry molars - 1st lower molars
   Saddle nose, frontal bossing, ophthalm's brow (prominence of
   Higoumenakis sign (sternoclavicle) prominence of sternal
   clavicular joint

2) Interstitial keratitis
   Rhashade - Rash on Face

3) Nerve deafness (SNHL)
   Clutton joints - Painless Joints - prone to Injury
   In diabetes, peripheral neuropathy

Rx: Penicillin G for 10-14 days

Syphilis:

Rubella

Triad of Rubella Syndrome:

Microcephaly

PDA (deafness SNHL)

Cataract

Eyes - mc eye manifestation of Rubella is Salt & Pepper Fundus

Cataract, glaucoma

Microphthalmia

Hear - PDA, Peripheral pulm. stenosis

VSD

PDA & "ASD" is rare in Rubella.

8) All of following occur in Rubella embryopathy except

a) Deafness

b) Mental retardation (MR)

c) Aortic stenosis

d) PDA

Notes from

Jain Stationery

Gautam Nagar

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True about Rubella embryopathy except ( )

a) Occurs when IgM Ab's in child
b) Occurs when IgG persists in child for > 6 mo

c) deafness, heart disease, cataract

d) Infection after 16 wks results in major cong anomalies.

Explain: "If mother gets rubella after 16 wks risk of CRS < 1%"

**Varicella Embryopathy**

**Chicken Pox**

It involves

1) Skin - Rash

2) Brain - Cortical atrophy

3) Optic Nerve - Optic nerve atrophy & hypoplasia

4) Plexus - Lumbosacral -> aplasia of limb bud hypoplasia of limb

might terminate the pregnancy - 1st Trimester

Mother has chicken pox

2 in 2 days before delivery / baby has chicken pox like

3 in 5 days after delivery / illness

Indicate of VZ Ig (vaccine) to baby to prevent chicken pox like illness.

**Hypoglycemia**

Def: Bid glucose < 40 mg/dl (R)

plasma glucose < 45 mg/dl

Causes:

1) Limited Reserve: Preterm, IUGR

2) Stress like - Sepsis
4) Galactosemia (Gal-1-P: 1-day hepatotoxic substance)
5) Hyperinsulinism - infant of a diabetic mother.

**HYPERINSULINISM - PEDERSON'S HYPOTHESIS**

<table>
<thead>
<tr>
<th>mother</th>
<th>Placenta</th>
<th>Fetus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glucose</td>
<td>Glucose</td>
<td>Glucose</td>
</tr>
</tbody>
</table>

- Stimulate
- β cell hyperplasia

2) Occur in insulinoma
3) Nesidioblastosis - β-cell pancreatic tumor
4) Beckwith Weidmann syndrome - Hemihypertrophy of limbs
- Macroglossia
- Risk of Wilms' tumor

**A Term infant born to a diabetic mother, few hrs after birth was lethargic & his bid glucose was 30mg%. What should be done next?**

**ASRS: Symptomatic Hypoglycemia. → can cause MRI**

1) Give 10% dextrose orally
2) 10% dextrose IV - 2ml/kg → keep on Glucose Infusion Rate (GIR)
3) Give expressed breast milk → 4-6 mg/kg/min
4) Do exchange transfusion → Glucose IR max 12 mg/kg/min.
   (If baby got seizure after 4-6mg/kg)

"Hypoglycemia has a bad prognosis"

In all above, monitor glucose level

max % of dextrose given through peripheral vein = 12.5%
otherwise if you give more, he/she develops thrombophlebitis of vein.
In emergency, I/m Glucagon can be given to cause glycogenolysis, glucogenesis.

Glucagon is effective for many in persistent hypoglycemia in all except:
1) Large for date baby
2) Neosdiomblastosus
3) Galactosemia (Gal-1-p may be hepatotoxic & Glucagon needs liver)
4) Infant of diabetic mother

A 1-yr-old boy has hepatomegaly & hypoglycemia & there is no jaundice. Hypoglycemia doesn’t respond to Glucagon.

- Glycogen Storage Disease Type 1: Vangiers
  Glu-6-P → Glycogen, def. of G-6-Phos Phatase
  G-6-phosphate
  Glucose (last step of glycogenolysis)

Glucagon helps in diagnosis.

Hypocalcemia

- Serum Calcium = 9-11 mg/dl

Def.: Sr. Ca < 7 mg/dl (R)
  Ionized Ca < 4 mg/dl or < 1 m mole/l
  Best index of calcium = Ionized calcium
  Free

Infants: Tetany is rare in infants
  → Have Tremors, jitteriness (R) Seizures
  → Tremulousness (Tremor +)
mcc of jitteriness in infants = Hypocalcemia.

Hypocalcemia → 

Early
< 48 hrs

Late
> 48 hrs

Causes
- Prematurity
- Feeding in phosphate rich milk in cow's milk
- Infant of diabetic mother
- Birth asphyxia:
  - Look for Ca/Phosphate
  - Good ratio = Ca/P > 2

100 ml Breastmilk Cow's milk
Ca 34 mg 118 mg
P04 15 mg 100 mg

Cow milk is not a Ca+ supplement it cause hypocalcemia.

Breast milk is good Ca+ Supplement & prevents baby from late hypocalcemia.

Breast milk protects against:
- a) NEC
- b) Late onset Hypocalcemia
- c) Pneumonia
- d) Rotavirus diarrhea
- e) Allergy/eczema
- f) RSV Bronchiolitis (IgA antibodies against RSV)
- g) Breast milk has long fatty acids, Docosahexaenoic acid (DHA) important for vision & myelination in 1st month of life.

American Academy of Pediatrics (AAP) recommends all infants should get Vit D

- a) A
- b) B
- c) C
- d) D
→ Scutvy occurs in 6-18 mon.
→ RDA vit D in infants - 400 IU/day.
  Breast milk - 25 IU of vit D/litre.
→ Till 1 year of life give 1 ml vit D drops.
→ 25(OH) vit D tells vit D level.
→ In America, fortified vit D is given.

Infant of Diabetic Mom:
→ Risk
  → Preterm
  → Stillborn
  → Macrosomia - because of Insulin & IGF.
  → IUGR due to uteroplacental insufficiency → class F/R.
  While "Classification" of Diabetic mother
  → Mother of class F/R.
  → Baby is at risk of RDS [L/S ratio > 3.5]
  → Hypoglycemia (because they get hyperinsulin by explained by Pederson's hypothesis)
  → Hypocalcemia, Hypomagnesemia (22%)
  → Can have Jaundice (19%)
  → Can have polycythemia
    ↓
    → Cause renal vein thrombosis (due to viscosity
        of bid)

All the following are complications in newborns:
  of a diabetic mother except:
  1. Hyperbilirubinemia
  2. Hypoglycemia X Hyperglycemia due to hyperinsulinemia
  3. Fluorinemia
Cong. Anomalies in infants of diabetic mother:
Mc congenital anomaly is
a) CVS - 8.5% of infants (VSD, TGA, HOCM) → Asymmetrical
b) Neural tube defects - 5%
   - Septal Hypertrophy

c) Sacral agenesis/caudal regression syndrome
   - Doc: β blockers
   - Propionate

d) Lazy left colon [Pseudoobstruct of intestine]
   - Most specific anomaly: Sacral agenesis/caudal regression syndrome

Lazy left colon
   - Hirschprung
   - Ganglion AD but Aganglionic
   - Not working

Both are differentiated by Rectal Bx.

Neonatal Seizures
   → Mc type of seizure in neonate: HIE in 1st mon of life

Causes:
   a) HIE

b) other MC - metabolic causes:

   Mc type of seizure = Subtle - Nonspecific - Blank stare/pawn

   In adults - GTCS

   Synaptogenesis is completed at 2 yrs
   Neurons are arborized

   DOC: Phenobarbital (not Diazepam)
   Diazepam has preservative Benzylalkonium chloride

   Blonds to albumin so Bilirubin is Free

   a) Thiamine  b) Vit C  c) Cyanocobalamin  d) Pyridoxine

   b) WOF is related to occurrence of Neonatal Seizure
Pyridoxine deficiency

↓

GABA Inhibitory

GABA level ↓

So there is an imbalance of the stimulatory vs inhibitory mechanism.

⇒ Pyridoxine dependency disorder

⇒ AR

⇒ rarest

Neural Tube Defects (NTD)

Embryology:

0 → anterior neuropore - close at 25 day

⇒

0 → posterior neuropore - close at 27 day

Failure of closure of ant. or post. neuropore develop NTD

If ant. neuropore is not closed - baby develop Anencephaly (Absence of brain & part of hindbrain)

Anencephaly:

⇒ Indications not to do Resuscitate

1) Anencephaly

2) Child looks like Trisomy 13 (Patau)

NTD babies are born post-mature due to absence of hypothalamic-pituitary axis
Encephalocele: herniation of brain tissue.

Post-neuropore not closed - Baby develops
spina bifida

meningocoele (herniation of meninges)

meningo(myelo)coele (herniation of meninges & nerves)

Lumbo-sacral area is most site for myelomeningocele.

Myelomeningocele:
- Has Paraplegia
- Ch. constipation
- Neurogenic bladder

In all these children do MRI because 80% of them have Obstructive Hydrocephalus & they have an associated Arnold-Chiari Type II malformation.

Ruptured myelomeningocele:
- Cover & gauge
- Gauge should be soaked in Normal Saline.

Betadine
Povidone is non-toxic
Spirit

Meningitis:

To diagnose meningitis in a ruptured myelomeningocele:

a) Swab culture - Coagulase -ve Staph
b) Blood culture (Bacteria in blood when there is injection)
c) Urine culture

Stool culture
NTD

1) maternal deficiency of folic acid
   - zn
   - malnutrition

2) maternal diabetes

3) alcohol

4) radiation

5) antiepileptic drugs like phenytoin, valproic acid.

6) Safest drug antiepileptic given in preg → carbamazepine (CBZ)

Valproate cause → Hypoplastic left heart syndrome
Phenytoin → also toxic

Folic acid - should be start taken

1 mon before & 3 mon after concep^n.

Folic acid given = 400µg. (1st Trimester)

Recurrence:

1 child → 3.5%
2 children → 10%
3 children → 25%

To prevent recurrence Folic Acid given is

Folic acid: 4000µg/mo → 4mgs

→ Marker of NTD in amniotic fluid (C)

a) Phosphatidyl esterase
b) Pseudo cholinesterase
c) Acetyl cholinesterase & AFP
d) Butyryl cholinesterase

In mother's serum → MSAFP

Maternal serum Alpha fetoprotein
Genetics

- male
- female

Heterozygote for AR

Parental carry of sex-linked recessive (female carrier)

Parents & Children

1 boy & 1 girl (in order of birth)

1st trimester

About 10% of stillbirth sex unspecified

Dizygotic twins

Proband (case under investigation)

Monozygotic twins

Method of identifying persons in a pedigree: Here the proband is child 2 in generation II

No. of children of sex indicated

Affected individuals

Consanguineous marriage

Autosomal recessive

Autosomal dominant

- Adopted child

They usually present late

For e.g. ADPKD present at 3rd to 4th decade of life (late)

JAIN STATIONERY
09654691327
Autosomal Dominant

D - myotonic Dystrophy (adult m's affect more)
D - Osteogenesis Imperfecta
M - Marfan's
I - Intermittent Porphyria
N - Noonan's Syndrome  → Disproportionate, short stature
A - ADPKD Adult PKD, Acholeplasia (short limb dwarfism)
N - Neurofibromatosis
T - Tuberosus Sclerosis
VH3 - V- Von Willebrand disease
H - Hereditary spherocytosis
H - Huntington's chorea
H - Hypercholesterolemia (familial)

---

Noonan's Syndrome:

- Look like Turner's phenotype
- AD (equally occurs in male)
- XX = XY
- XO = 60%
- MC heart defects in Noonan's: Valvular Aortic Stenosis > HOCM, ASD

MC heart defect in Turner's Syndrome is

- Y(2)-yard have Bicuspid Aortic
- 20% Coarctatio Aorta

---

Turner's
- XO = 60%
- AD, XX = XY
- Heart 1/3 - 1/2 Bicuspid aortic valve = valvular PS, HOCM, ASD
- 90% CoA
Turners

TVOonans

Infertile girls

Girls: delayed puberty & mostly they are most fertile.

Boys: Cryptorchidism

The chance of child being not affected if both parents are affected in Achondroplasia:

- a) 0%
- b) 25%
- c) 50%
- d) 100%

AD are mostly heterozygous - Mendel

\[ AA^c \quad AA^c \]

\[ AA \quad A^cA \quad A^cA \quad A^cA^c \]

Autosomal Recessive (Presents Early)

Parents are carrier

Children are affected

Both males & females are affected

"Consanguinity precipitated AR trait

most of metabolic disorders are AR

Autosomal Recessive

Cystic Fibrosis

\[ x^c \text{ AT deficiency} \]

Wilson's disease

Haemochromatosis

Friedrich's ataxia

Gaucher's disease

Neimann's Pick's

Tay Sachs'
British guy

British lady

XY carriers

XX carriers

Baby can have cystic fibrosis??

[In europe/UK - 1 in every 25 have carriers for cystic fibrosis.]

\[
\frac{1}{25} \times \frac{1}{25} = \frac{1}{625} \text{ Chance have CF}
\]

\[\begin{array}{c}
A^C A^C \\
A^C A^C \\
A^C A^C \\
A A
\end{array}\]

1 in every 1500 have CF (ask nearest relative UK/Europe)

British guy

British lady

my brother died of CF

\[\begin{array}{c}
\text{Chance of baby can have CF = } \frac{1}{150}
\end{array}\]

\[
\frac{1}{25} \times \frac{3}{25} \times \frac{3}{4} = \frac{1}{750}
\]

\[\begin{array}{c}
A^C A^C \\
A^C A^C \\
A A
\end{array}\]

ENZYME REPLACEMENT

which was first disease for which enzyme replacement was successful -

Gaucher's (β-glucocerebrosidase) - Genzyme

CER-enzyme

Others - Pompe's (GSD-1) - Rx: Enzyme Replacement

- Fabry's XLR (lysosomal: cherry red spot)

- mucopolysaccharidosis type I - Hunter

- MPS type II - Maroteaux-Lamy
Hurler's Syndrome (Gargoulism) - It is MPS

Deficiency of L-Iduronidase.

1. None at birth
2. Developmental delay
3. Hepatosplenomegaly, Kyphosis
4. Coarse facies
5. Clouding cornea
6. Umbilical & inguinal hernia
7. MR
8. Stiff, immobile joints, death in teens
9. Chronic rhinitis

Gene Therapy
First successful in XLR - SCID. Seven Combined Immunodeficiency

Virus DNA → Enzyme + Adenosine deaminase enzyme

Transduction.

X-Linked Recessive (Poor Boys)

Females - Carriers
Affected - Boys

H/o on maternal side.
A carrier female \( \square \) male

\[
\begin{array}{c}
XX^C & \rightarrow \text{carrier}\text{ (affected)} \\
XY & \\
XY^C & \times \times & X^C Y & \times Y
\end{array}
\]

\( \frac{1}{2} \) of daughter carrier

\( \frac{1}{2} \) sons affected

XLR disorders: Poor boys!!

1) Duchenne's muscular dystrophy
2) Haemophilia A & B
3) G6PD deficiency
4) Wiskott-Aldrich syndrome
5) Color blindness
6) Lesch-Nyhan syndrome
7) Ch. granulomatous disease \( \rightarrow \) an immunodeficiency disorder \( \rightarrow \) NADPH oxidase deficiency: NBT dye

Duchenne's muscular dystrophy

\( \rightarrow \) Mc. Hereditary neuro muscular disease

\( \rightarrow \) Pseudohypertrophy of calf muscle because of fat infiltration

\( \rightarrow \) Proximal muscle weakness

\( \rightarrow \) Ine Gower's Sign ↑

"Human genome has 30,000 genes largest genome is Dystrophin"

Dystrophin

\( \square \) in Skeletal m's, Brain, Heart

- Died in teenage life because of Recurrent chest infections & cardiomyopathies
- \( \frac{1}{3} \) of Duchenne's have MR
- \( \frac{1}{3} \) of Duchenne's also occur because of New
CPK is very high in Duchenne's (10,000 etc)

Wiskott–Aldrich Syndrome

Trait:

- Eczema
- Thrombocytopenia
- Immunodeficiency

Lesch–Nyhan Syndrome

- Purine defect
- HGPRTase deficiency
- Self mutilating (1st nose to palate)

1) A mentally challenged child with dysphagia & ophisticotonic spasms. He also have choreoathetoid movements &
   self mutilating behavior & the family. H/o: WOF T3 is suggested

2) Sr. uric acid > 8 mg/dl => hyperuricemia

3) Sr. alkaline phosphatase

4) Sr. lactate DHase

A male child (2) Fanconi Syndrome & nephrocalcinosis has variant of Dent disease. All are true except:
   a) Hypercalcinia → Nephrolithiasis → X-IR
   b) Proteinuria → LMW Proteinuria, BA mOglobulinuria
   c) Similar presentation in Father/mother
   d) Rickets

Fanconi (- generalized proximal tubule defect.

Proximal tubule absorb:

\[
\begin{align*}
\text{Na}^- & \quad 65\%  \\
\text{HCO}_3^- & \quad > 85\%  \\
\text{P}\text{O}_4^- & \quad > 88\%
\end{align*}
\]

AA & glucose - 100%

↓ loss cause (Phosphaturia)

Rickets
LOWE SYNDROME

Inheritance of Lowe syndrome - XLR

Oculo-cerebro-renal syndrome

\[ \rightarrow \] FANCONI SYNDROME

Cataract  MR

Glaucoma  microcephaly

- 3yr 6mon/ male
- Bilateral cataract
- Global developmental delay, Hypotonia
- Polyuria, polydipsia
- Rickets

\[ X - \text{LINKED DOMINANT} \]

I \( x^Y \)

II

III

IV

Father Transmits his disease to all of his daughters

No male - male transmission

\[ X \text{ linked dominant} \]

- Familial hypophosphatemic Rickets
- Urea cycle defect due to Ornithine Transcarbamylase deficiency
- Incontinentia pigmenti \( \} \) only seen in girls
- Rett's syndrome \( \} \) (boys die - it is very severe
  girls live)
RETT Syndrome

It is one of pervasive developmental disorder (PDD) 
PDD are 1) Autism - common in boys < 3 yrs 
2) RETT 
3) Asperger's syndrome - common in boys 
↓
- Have good IQ, no MR

KFA "LITTLE PROFESSORS"

Girls & Rett's are born till 6-18mon of life 
[after later in life they develop]

↓
- Head growth, microcephaly, MR
- Seizures
- Stereotypical hand movements
- Repetitive behaviour

RETT's due to mutation in MECP2 mutation gene

AOF are characteristics of Rett syndrome except:

a) Incidence of MR
b) Seizure
 c) Abnormal dendritic morphology in cortical pyramidal cells
    * Seen in post mortem brain by
 d) Macrocephaly

Q) MCC of death in girls to RETT (die suddenly)
   a) Seizures
   b) Arrhythmias (cardiac)
Congenital Heart Disease (CHD)

Nada's criteria:
1. Major criteria
   1. Systolic murmur grade III or more at time
   2. Diastolic murmur
   3. Central cyanosis
   4. CHF

   In child, we see physiologic/flow innocent
   murmurs; systolic < grade III normally.

Minor criteria
systolic murmur < grade III
Ab (S) S2
Ab (S) ECG
Ab (N) X-ray
Ab (N) BP

CoA:
- weak femoral pulse, differential BP.

According to AAP
- In every growing child, check femoral pulse.

Ab (N) ECG

All newborns are born E RVH & RAP,
E in 1st month of life they develop like an
adult.

For this look ECG

V1, V2, V3, R

ECG of newborn:
- T waves -n "up" on
After 1st 48 hrs T waves become "-ve"

QRS axis " - Take any 2 lead

In Newborn = +150°, +180° line head

2nd Heart sound = due to closure of A2 Pulm. a.

→ Split varies with Resp. Freq. because

During inspiration → V1 → delay in closure of pulm. valve to split

IN ASD = 2nd HS is wide & Fixed

ASD = -- RE L -- VR, V2, V3

→ Vol. overload of Rt. ventricle

→ RVH (RV Hypertrophy)

→ RAD (LT axis deviation)

→ 2nd HS Fixed because RV is already overloaded

VSD: 2nd HS wide & variable

TOF: A2

only A2 is heard

P2 is not heard
**SYNDROMES due to**

**Maternal:**
- Lithium → **Ebstein's anomaly**
- Mumps → **endocardial fibroelastosis** *(LV obstruction of newborn)*
- Penicillamine → **cutis laxa** *(skin overhanging from mouth)*
- *Naftafin* → **Chondrodysplasia punctata**
  & ab® nose
- SLE → **Complete heart block in newborn** *(because of anti-Ro Ab's in mother)*
  Pacer makes insert® needed in baby.
  mc in Sjogren syndrome
- *Thalidomide* → **Phocomelia** *(now Thalidomide use in Methyl®)*

---

**Fetal Circulation**

Placenta oxygenates blood
- Pulm. a → A® ′communal′ → Ductus Arteriosus
- Ductus joins A® distal to origin of @Subclavian.

2 umb. a & 2 veins
- Rt umb. vein disappears
- Lt umb. vein remains.

Umbilical a bring impure blood to placenta →
↓ O-venous

Placenta 30-35 mm
↓ umbilical vein

Oxygenated ↓
↓ IVC. PO₂ is around 28-30
↓ Lt side
↓ Rt ventricle
During fetal life, blood doesn't go lung. Some part of blood that went to RV goes to descending aorta equilibrated.

Closure Order

- Umb. a. close
- Umb. v. close
- Ductus venosus closes
- Ductus arteriosus closes

Ductus arteriosus
2 closures

Physiological
Anatomical:

At Birth
Due to vasoconstrict

Lumen closes
It closes
Lumen closes by 10-21 days

Classification of CHD
Acyanotic

Lt → Rt: Shunts

1) ASD at level of ventricle
2) VSD - at level of ventricle
3) PDA at level of D. Arteriosus

4) Pulm. Bid. Flow ↑ kla Plethora
5) Failure to Thrive
6) Recurrent pneumonia
7) Feeding difficulty kla Feeding Diaphoresis
8) CHF as early as 6-10 wks of life

When CHF occurs, symp. stimulat occur to ↑ contractility
- Tachycardia, Sweating
- Cardiomegaly
Cyanotic

Based on Pulm-Bld Flow

↑ (Plethora) ↓ (Oligemia)

0) TAPVC
1) Cardiomegaly (cm)
2) TGA + VSD
3) Cardiac anomaly cm
4) Ebstein's anomaly (cm)
5) Truncus Arteriosus

It can be a size heart or ↓

Single G2 (HS)

1) TOF
2) Double-outlet RV & pulm. stenosis (PS)
3) TGA & VSD or PS
4) Tricuspid atresia
5) Single ventricle or PS

TAPVC

Pulm. vein joins Rt. side.
Supra cardiac - Pulm. vein join to SVC
Cardiac - Pulm. vein joins to coronary sinus
Baby is Cyanotic & PLETHORA (↑ Pulm. bid flow).

All Pt. w/ TAPVC have ASD (to survive) otherwise they cannot survive.

Most TAPVC are Supracardiac (50%) in this Heart looks like "Fig of 8" or Snowman or Cottage Loaf appearance.

Most Severe is TAPVC is Infracardiac present at Cyanosis at birth because of Obstructive Pulm. venous hypertension. In Infracardiac TAPVC,

CXR: Kerley B lines

Ground glass appearance (also seen in RDS)

Only Heart disease that worsen by PGs infusion is Infracardiac TAPVC (or pulsus paradoxus)

Truncus arteriosus

Persisting TA causes Cyanosis & Plethora.

CHD & plethora are all except (PGs)

a) TGA & VSD
b) Tricuspid atresia
c) TOF
d) TAPVR

WOCHD doesn’t produce Cyanosis

a) Persistent Truncus arteriosus
b) PS (Isolated PS) → acyanotic
c) Single Ventricle
d) TAPVR
Critical PS → cyanotic at birth

PS murrum radiates to back

ASD

MC → Secundum ASD

Syndromes of ASD

1) Down Syndrome
   1. 40-50% of children have CHD
   2. CHD is also one of death in down's
   3. MC CHD in down's is endocardial cushion defect or Ostium primum ASD or AV canal defect

2) Holt-Oram Syndrome

   AD, familial
   Its an ASD secundum
   VSD, 1° block, AF seen in pts.
   Bony defects
   Thumb ab □ seen → absent thumb, rudimentary thumb

Very large Lt → Lt shunt
Pulm bid flow ↑ (plethora)
Eg. of plethora.
CHF never occurs in TOF + Cardiomegaly.

\( r_x = \text{Pollicisat}^0 \) of Index finger (Index finger as Thumb)

Defect in single gene leads to multiple effects is k/n
a) pleiotrophy T8x5 mutation - Holt-Oram Syndrome
b) Penetration Hand \& Heart Syndrome
  \( \text{Common Transcript}^0 \) Factor
c) Dominance
  \( \text{Chimeraism} \)
d) None of the above

In Holt-Oram Syndrome all of following defects are seen except:

a) PDA
b) ASD
c) VSD
d) 1st Heart block
e) AF

3) Lutembacher Syndrome: - ASD +MS--
4) Ellis van Creveld Syndrome: - ASD + Polydactyly---

ASF Secundum
During childhood \( \text{child is asymptomatic} \)
Wide \& Fixed \( s_2 \) (2nd /3s)

\[ \text{On ECG} \rightarrow \text{RAD} \]

Adult: - RV Failure

AF (chorea) \( \rightarrow \) CVA
Reverse shunt \( R \rightarrow L \)
Eisenmenger Syndrome (cyanotic)

\( \text{If} \) ASD Secundum must close 3-5 yrs by Occluxon patch

If ASD Secundum \( < 3 \text{mm} \) \( \text{it can be closed by nature} \)
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD PFOAM</td>
<td>Baby has ASD &gt; LAD</td>
</tr>
<tr>
<td>VSD</td>
<td>Baby also have atrial septum</td>
</tr>
<tr>
<td>Big cut in septum</td>
<td>Baby also have one atrial septum</td>
</tr>
<tr>
<td>Turbulent flow</td>
<td>Vegetations</td>
</tr>
<tr>
<td>Damage wall</td>
<td>Vegetations</td>
</tr>
</tbody>
</table>

- **ASD PFOAM**: Small ASD (mids) R - L
- **VSD**: Large, small, medium
- **Large Size**: VSD and small ASD
- **Small ASD**: Always require surgery

**VSD** is less of infective endocarditis.

**ASD PFOAM**: Least chance of infective endocarditis is.

It is rare in small ASD secundum.
Small  medium  Large (RV Pres > LV Pres)

asympotomatic  Shunt  murmur absent
Small extra systolic murmur (no murmur heard)

→ In medium to large size of VSD, LA enlargement occurs first.
→ VSD is vol. overload of left heart so LVM is seen.
→ Delayed diastolic murmur at apex (due to large amount of blood crossing mitral valve) in large VSD.

Complications of VSD:
1. Most complication of VSD is Infective Endocarditis.
2. Pt. has CHF 6-10 weeks of life.

Rx:
- By natural course - VSD closed by:
  - Perimembranous VSD - 80%  
  - Muscular VSD - 30%  

Indications of Rx in a case of VSD:
1. Failure of medical therapy.
2. Swiss cheese VSD - multiple VSD.
3. Supracristal VSD - large VSD at ventricular outflow.
4. QP > 2 (even if there is no symptoms)

QP Pulm bid Flow
QS Systemic bid flow

Large VSD:

[Diagram of pulmonic and systemic blood flow]

Lung Heart Transplant

PAPERSDTO develop Eisenmenger (R→L) Reversal Shunt

RVH (RV Press ↑)
NO & Bosentan (Endothelin antagonist) given in Pulm. HTN.

B) Sequence of events in Eisenmenger's Syndrome:
   a) L → R shunt → RVH → R → L shunt → Pulm. HTN
   b) L → R shunt → Pulm. HTN → RVH → R → L shunt
   c) L → R shunt → RVH → Pulm HTN → R → L shunt
   d) R → L shunt → LVH → Pulm HTN → L → R shunt

**PDA & Eisenmenger's Syndrome**

a) Differential Clubbing / Cyanosis
b) more impure blood goes to Toes as compared to Fingers

In children: 2 Down's syndrome → endocardial cushion defect can cause Eisenmenger syndrome. In early mon's 80% do EchoCG of life.

**Ductus dependent lesions**

1. The following cardiac defects are characterised by ductus dependent bid flow except:
   a) Transposition of great arteries: intact septum → mixing doesn't occur
   b) Interrupted aortic arch - Systemic bid flow dependent
   c) Truncus arteriosus - always cyanotic - ductal independent
   d) Hypoplastic left heart syndrome (HLHS) - Systemic bid flow dependent

1. Systemic bid flow dependent

   e.g. Interrupted aortic arch
      b) Pseudocal Coarctation
      c) HLHS
      d) Critical AS

2. True child present is shock Feeble pulses (as child grows, DA closes)
MC cyanotic Heart disease in Infants. TGA

Cool peripheries:

6) A child is admitted on 7 days of life with severe RD & shock. He was discharged 3 days back healthy. what could be probable diagnosis?
   a) VSD large
   b) HLHS \rightarrow Rx: Give PGE1 infusion

Pulm. Bld flow dependence:
   1. Tricuspid atresia
   2. Critical PS

These child present w/ "central cyanosis" in 1st 7-10 days.

3 Oxygen dependence:
   eg. TGA

TGA at birth - Cyanotic.
TGA child died at birth due to hypoxia.
"TGA dependent on ductus for oxygen."

Infra cardiac TAPVC worsen to PG's.
Rx of TGA:
Emergency operator in TGA is Rashkind balloon atrial septostomy (more mitral - more oxygenate?)
\rightarrow definitive sx is Jatene's arterial switch (changing und...
Cyanotic

TGA

RVH → TDF
LVH → Tetralogy

Switch should be done early and in the first 2 weeks of life.

- 50% of TGA are present with VSD and they don't have any problem except mild cyanosis.
- Complete TGA is more TGA.

Tricuspid atresia

- Atresia of Tricuspid valve.
- Can only survive if he has both ASD & VSD.
- It is cyanotic.
- Hypoplastic RV.

→ Cyanotic c ↓ Pulm bid flow
→ LVF
→ LAD

Ebstein's Anomaly

- ECG shows it is like tricuspid valve into right ventricle.
- Hemodynamics is to tricuspid atresia.

1. Cyanosis c ↓ Pulm bid flow
2. Maternal lithium
3. Systolic murmur, diastolic murmur
   Quadriple rhythm
4. Box shaped heart/Balloon shaped heart

5. Discordance b/w intracardiac ECG & intracardiac pressures recording
MC Cyanotic heart disease beyond infancy is TOF

1. They have arrhythmias like SVT, WPW

TOF

1) Infundibular PULM STENOsis: It's like a Subvalvular narrowing of PULM valve.

2) Perimembranous VSD: VSD - Opening b/w LV & RV

3) RVH
   - RV press becomes Supra Systemic
   - Shunting is R→L
   - Cyanosed, Clubbing
   - Oligemic lung due to Polyhydramnios
   - Polyhydramnios due to IDA
   - Ferritin levels are low

4) Overriding of aorta

During infancy, → Cyanotic / Tet spells / Hypoproteinemic spells

Squatting episodes

2 Complications of TOF:

MC complication in Children < 2 yrs → Thrombosis

> 2 yrs → Haematogenous Brain abscess

Notes from Jan Stationery

Gautam Nagar
09654691327
Shunt is done in infants
Definitive repair is done adults.

Pathogenesis of Cyanotic Spell:
During the Time of Spell
1. $R \rightarrow L$ Shunt $↑$
   $↓$
   $↓ P_O_2$ (hypoxia)
   $↓$ stimulate
   Rep centre. (+++)

During Spell
a) $↓$ Systemic vascular resistance ($R \rightarrow L$ Shunts go easily)
b) Dynamic pulmonary stenosis ($RV$ press $↑$, $R \rightarrow L$ shunt $↑$)
c) Crying (before crying, Take breath $↑ RV$, $R \rightarrow L$ shunt)

Rx of Spell

1) $I/v$ NaHCO$_3$, $I/v$ Oxygen inhalation $→$ to $↑ P_O_2$
2) Subcutaneous morphine, Suppress Rep. Centre
3) $I/v$ Ketamine, Phenylephrine $↑$ SVR
   Whenever we squat, Femoral a. etc compressed
   $↑$ SVR
   In emergency conditions, We put him in knee chest position
4) $I/v$ Propomolar $→$ Dynamic PS

Management of Cyanotic Spells include all except
1. Oxygen Inhalation
2. Knee chest
3. Phenobarbitone / CaCl$_2$
4. Morphine

Rx
Types of Palliative Shunts

1. Blalock-Taussig Shunt:
   Anastomose Subclavian to Pulm. a. (Joined Pulm. a.
   which is off to aortic arch.)


Blalock-Taussig Shunt:
Some child of TOF have Rt. aortic arch.
Rt. aortic arch most commonly &
A. Tetralogy of Fallot (25%)
B. Transcatheter atresia (50%)

After 3-6 mon of shunts. we do definitive Sx.

Pott's Shunt:
Rt. subclavian a. to Rt. Pulm. a.
Descending aorta to Lt. Pulm. a.

Trilogy

A. Tetralogy

Overriding of aorta is absent

CHF never occur in TOF

C. Cardiomegaly

RT Heart never fails because R→L Shunt.

Q) A) Murmure present & recurrent abd. pain, restlessness, LV failure - LV is ischemic.
   Instability & diaphoresis. - on feeding Cardiac auscultation reveals a non specific murmur.
   He is believed to be at risk of MI. ECG shows Q waves - V1, V2, aVR.
   ST elevation & T wave depress. The most likely diagnosis is ASD.
2) VSD
3) TOF
   ALCAPA
   1. Lt. coronary coming from pulm. a.
   2. LAD branch
5) Anomalous origin of coronary a. (C.A come from aorta)
6) LV severely ischemic.
   Any infant or antenatal MI is ALCAPA

We do Angiography → RE. coronary fills because it is
   coming from pulm. a.
   pxz = Kawasaki Disease

8) A 2yr old boy is brought into emergency room
   with a complaint of fever for 6 days &
   development of limp. O/E, he is found to have
   an erythematous macular exanthema over his
   body, ocular conjunctivitis, dry x cracked lips
   a red throat, x cervical lymphadenopathy.
   There is grade II/VI vibratory systolic
   ejection murmur at lower left sternal border.
   A WBC & differential show predominant
   neutrophils 2 ↑ platelets on smear.
   Osis: Kawasaki disease.

Mc acquired disease → UK, Chandigarth, Japan
   is Kawasaki

Mc acquired disease in India is Rheumatic heart
disease.

→ WOF vasculitis doesn't occur in adults.
   a) Kawasaki disease - 85% children <5 yrs age
   b) Susac's syndrome - vasculitis in adult females, rhinal a. involved
Kawasaki Disease

It is the most common vasculitis.

MC complication of Kawasaki disease is:

20-25% coronary artery aneurysm development → Thrombosis → die due to aneurysmal MI

Pathogenesis:

- Occurs because of Staph or Streptococcal super Ag's
  - New pathogenicity: this Ag's present in air (cold wind movements in trophic zone)
  - Seasonal disease
  - It affects only children

Kawasaki Disease k/a mucocutaneous lymph syndrome

Diagnostic Criteria:

1. Fever > 5 days & any 4 of these 5:
   1. Changes in extremities (e.g., erythema, edema, desquamation on palms & soles seen on 3rd week of illness)
   2. Bilateral conjunctivitis (not abscess & exudates)
   3. Polymorphous rash (not vesicular) - Sandpaper rash
   4. Cervical lymphadenopathy, U/L > 1.5 cm
   5. Changes in lips & oral cavity (e.g., pharyngeal erythema, dry, fissured or swollen lips, strawberry tongue)

Drug that prevents aneurysms:

- IV Ig 2 gm/kg (acute phase)
- Aspirin 100 mg/kg/day x 2 weeks to ↓ risk by 4-6%.

Kawasaki → Aneurysm

Medium - large size

Aspirin 3-5 mg/kg/day + Aspirin + WARFARIN (anti-thrombolytic)
mcc of death in Kawasaki disease
in acute phase is myocarditis
→ Extracardiac infections: aseptic meningitis, irritable arthritis, urethritis

**Blood Pressure**

2yr old child, unconscious, BP - 86/60 mm Hg,
HR - 180/min, capillary filling time - in 4 sec
= degree compensated shock.

Shock: BP < 10th percentile for that age & sex.
HTN: BP > 95th percentile for that age & sex

10th percentile Systolic BP

\[
\text{4yr: 40 mm Hg} \\
\text{>1yr: 70 + (age x 2) mm Hg}
\]

[Capillary filling time = 2 sec.

HR ↑ because of stimulation of symp system

Rx: Normal saline 20 ml/kg

Check perfusion

HR

CFT

**HTN**

BP > 95th percentile for age & sex.

- Essential HTN: 10% obese ↑

In adults - essential HTN -

In child it is not

- *2° HTN: MCC HTN in children - renal (Reflex nephro...*)
Renovascular major Renal atherosclerosis, R.Ven thrombias small HUS

b) Cardiac: - Post ductal Co A (don't have problems because they develop collaterals, they present late)

Endocrine: - Hyperthyroidism, Pheochromocytoma, Cushing's, 1st hyperaldosteronism, Cong. Adrenal hyperplasia & Hypertension, 11-B hydroxylase

Check 4 limbs to r/o Co A

End organ damage:

Renal: al proteinuria
Fundus: Retinopathy
Heart: Concentric LVH

Rx of HTN: - ACE 1 / ARB's
C/I in GFR < 30 ml/min/1.7 m² because of hyperkalemia

GFR < 30 → Doc is CCB's like Amlodipin.
Mostly HTN is asymptomatic
Sometimes HTN becomes emergency

Emergency → seizures
LVF; S3 gallop
basal crepts

1) A 4 yr old boy is brought to pediatric emergency & unconscious following seizures. The BP in all 4 limbs is 150/110 mm of Hg. The most appropriate drug from him is: Aasis: Hypotens' Emergency.

a) 1/2 Diazepam
Nitroprusside is very sensitive to light. 
Check BP every 30 min.

2) Propranolol 1/IV
3) Aminophylline 1/IV, 1/IV labetalol, 1/IV esmolol
4) Phenobarbitaline 1/IV

We should not give Nitroprusside > 48 hrs
because it causes Cyanide poisoning (Poisoning)

Takahayasu Arteritis
A 12 yr old boy presented w/ seizures &
BP 200/140 & femoral pulses were not
palpable. 

a) Takahayasu arteritis - Pulseless disease
b) Grand mal seizures

C) Fibromuscular dysplasia - Renal a. stenosis in adult females
d) Renal parenchymal defect

In Japan & USA - TA a.ffect - Arch of aorta
In India - TA affects - Abd. aorta

Tachyarrhythmias

Based on QRS

VT / VF

MC Tachycardia

In children:

SVT:
- Stable: Adenosine stable, Pulseless arrest
- VT:
  - Should be given
  - Brisk P waves absent:
  - Inverted
  - PR > 220 in infants
  - 180/min in older children

1) Vagal maneuvers
2) Bulbar massage
3) Valsalva maneuvers
4) Ice packs over eyes
**GENETICS**

**DELETIONS**

<table>
<thead>
<tr>
<th>Major</th>
<th>Micro</th>
</tr>
</thead>
<tbody>
<tr>
<td>5p⁻ (5th chromosome short arm deleted)</td>
<td>Williams Syndrome (7q23)</td>
</tr>
<tr>
<td>Cri du chat Syndrome</td>
<td>Prader-Willi Syndrome (15q11-13)</td>
</tr>
<tr>
<td>Cry-like cat</td>
<td>DiGeorge Syndrome (22q11-)</td>
</tr>
<tr>
<td>Abnormal larynx</td>
<td>ASIS: FISH (Flourescence In situ Hybridization)</td>
</tr>
</tbody>
</table>

**DiGeorge Syndrome**

- Hypoplasia of 3rd & 4th pharyngeal pouch
- Absent thymus & absent parathyroids

**William's Syndrome**

- Supravalvular Aortic Stenosis
- Hypercalcemia
- Also have peripheral PS
- They have ELFIN FACIES

**TRANLOCATIONS**

<table>
<thead>
<tr>
<th>Balanced</th>
<th>Unbalanced</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total amount of genetic material remains same after translocat°</td>
<td>Robertsonian Translocates b/w 2 acrocentric chromosomes therunbalance</td>
</tr>
</tbody>
</table>

\[
\text{Unbalanced: } p^+ + q^+ \rightarrow q + \text{lost}
\]

**Down's Syndrome**

- MCC of mental retardation is Down's syndrome
- 95% of Down's is Trisomy
- Maternal meiotic non disjunction
  - (Risk of Down's increases with maternal age)
Risk of Down's in young mother is

1) 3-4% by Robertsonian Translocation (acrocentric chromosome)

- Group D: 13, 14, 15
- Group G: 21, 22

Down's occurs by: D-G, G-G Translocation

2) 1-2% by mosaic = 46/47, 47/46

Mosaicism

- 2 different cells from a single zygote

Chimera: different cells from different zygotes
- Rare in humans, occurs in cattle

In Turner's - 60% are XO
(Downs)

Rest are mosaic - XO/XX
XO/XY

Somatic mosaicism

- Not transmitted to next generation

Germ line mosaicism

- Transmitted to next generation

We can't diagnose germ line mosaicism by blood (because in blood is X)

- e.g.: Osteogenesis Imperfecta
- Risk of recurrence 1/2
A couple has 2 children affected with Tuberculous Sclerosis. On detailed clinical & lab evaluation (including molecular studies) both parents are normal to explain 2 affected children in this family.

- Non penetrance
- Uniparental disomy
- Genomic imprinting
- Germline mosaicism

**GENOMIC IMPRINTING**
(Chr 15q 11-13)

\[ \text{Delet}^n \]

Prader-Willi Syndrome
Angelman Syndrome

Genetically defined as differential expression of genetic defect based on parent

Prader-Willi Syndrome
Obese
Severe neonatal hypotonia
Small hands & feet
Mental retardation
Unusual behavioral profile

Angelmann Syndrome
"Happy Puppet"

- At birth but subsequently develops
- Seizures, mental retardation, ataxia

Prader-Willi Syndrome is an X-linked disorder

Ghrelin: appetite hormone, mostly from fundus, eats more
FSH
LH
GH
Uniparental Disomy

AA

Cystic Fibrosis

AA

Carrier

Unequal Crossing Over

Child acquires two ab genes from affected parent.

E.g. Some Cystic Fibrosis, Sickle cell anemia
- Russel-Silver Syndrome
- Beckwith-Wiedemann Syndrome
- Neonatal DM
- 25-29%, Prader-Willi Syndrome & 5%, Angelmann

Uniparental disomy
Uniparental disomy

Mitochondrial Inheritance

Maternal Inheritance

E.g.-
1. Myoclonic epilepsy & Red Ragged Fibers (MERRF)
2. Mitochondrial encephalopathy, stroke like episodes,
   & Lactic acidosis (MEALS)
3. Leber hereditary optic neuropathy
4. Leigh disease
5. Kearns-Sayre Syndrome (chronic progressive
   ophthalmopigia)
6. NARP: Neuropathy, ataxia
   Retinitis pigmentosa.
Anticipatory

Grandfather - 60yrs, Father - 40yrs, Child - 10yrs
every successive generation disease is more severe & presentin
carries an Anticipatory.

Eg: Trinucleotide repeat disorders are eg's of anticipatory.

a) Fragile X - CGG
b) F Ataxia - GAA
c) Myotonic dystrophy - CTG, CCTG
d) Spinobulbar muscular atrophy - CAG
e) Huntington's chorea - CAG
f) Spinocerebellar ataxia - CAG/CTG

DEVELOPMENTAL MILESTONES

Neck holding - 3 mos

3 mos - Neck holding
5 mos - Sitting & Support
4-6 mos - prone to supine, supine to prone (roll in bed)
8 mos - Sitting & out support
9 mos - Crawling
10 mos - Creeping (Stand & support)
12 mos - Standing & out support, walking & out support
2 yrs - Walk up stairs & 2 feet at each step
3 yrs - Upstairs & one foot at each step, ride tricycle

4 yrs - Hops on one foot
5 yrs - Bicycle

NOTES FROM
JAIN STATIONERY
GAUTAM NAGAR
09654691327
Anticipation

Grandfather 60 yrs, Father 40 yrs, Child 10yrs
Successful every generation disease is more severe & presenting
causing K/A Anticipation.

eg: Trinucleotide repeat disorders are eg's of anticipation.

a) Fragile X - CAG
b) F Ataxia - GAA
c) Myotonic dystrophy - CTG, CCTG
d) Spinobulbar muscular atrophy - CAG
e) Huntington's Chorea - CAG
f) Spinocerebellar ataxia - CAG/CTG

DEVELOPMENTAL MILESTONES

Neck holding - 3 mon

3 mon - Neck holding
5 mon - Sitting & support
4-6 mon - Prone to supine, supine to prone (roll in bed)
8 mon - Sitting with support
9 mon - Crawling
10 mon - Creeping (stand & support)
12 mon - Standing 2 out support, walking 2 out support
2 yrs - Walk up stairs 2 2 feet at each step
3 yrs - Upstairs 2 one foot at each step, ride tricycle
4 yrs - Hops on one foot
5 yrs - Bicycle
   Skips on 2 feet
Fine motor

12 weeks - Grasp reflex disappears
4 mon - Goes for objects
5 mon - Bidextrous grasp
6-7 mon - Transfer object, palmar grasp
9 mon - Pincher p grasp, mature at 1 yr
13 mon - Casting
15 mon - Self feed & a spoon
18 mon - Self feed & a cup
24 mon - Turns pages of a book one at a time

Fine Hand Skills

3 yrs →  ○ Scribbles horizontally & vertically → 2 yrs

4 yrs → +

4 yrs → Resist

5 yrs → Δ

6 yrs → X

7 yrs - Diamond □

8 yrs → +

9 yrs - Cylinder □

11 yrs - Cube □

15 yrs - Tower of 2 blocks
18 mo - Tower of 3 blocks.

2 yr old child - make a Train

2½ yr old - make a Train & chimney.

3 yr old - Bridge

3 yr - Tower of 9

4 yr - Gate

6 yr - Steps

Social

2 mon - social smile

3 mon - recognizes mother

6-7 mon - Smiles at mirror image, keeps

9 mon - waves bye-bye

6 mon - Stranger anxiety

2 yrs - Dry by day

3 yrs - Dry by night

dress / undress himself, needs supervision

5 yrs - dress / undress himself with supervision.

Bladder control (85%)

>5 yrs - If baby wet's bed then it is Nocturnal enuresis.

Best Rx for Nocturnal enuresis:

1) Behavioural Therapy (i.e. reinforcement)

2) "Alarm" Therapy

3) E:

Encopresis: - stools in undergarments > 4 yrs age.

Rx: - Laxative
**Language**

- 1 mon: head turns to sound
- 3 mon: cooing
- 6 mon: babbles, monosyllables (ma, ba)
- 9 mon: disyllables (ma-ma, baba) object permanence, peek a boo constantly
- 1 yr: 2 words & meaning
- 18 mon: vocabulary of ten words
- 2 yrs: simple sentences & 4 words (2 phrases, poodle, snow man)
- 3 yrs: tells age & sex, uses pronouns, handedness, identify colors
- 4 yrs: tells story
- 5 yrs: knows colors, 4 colors
- 15 mon: 4-6 words
- 2 yrs: 50-100 words
- <3 yrs: child understand "DEATH"
- 3-9 yrs: doubt that can
- > 9 yrs: understand happen to him

**Respiratory System**

**Viral Infections**

**Rhinovirus:** mc

**Pneumonia**

Mc of Pneumonia is RSV in children

Others are adenovirus,

Influenza, ParaInfluenza

Viral Pneumonia is B/L & Interstitial infiltrate

These are not lobes

Rx for RSV: Ribavirin

Doc for Influenza: Tamiflu, Relenza

H_1N_1 -> H_1N_1 Type A Influenza: Neuamidine θ

Coxal drug: Oseltamivir (in pandemic)
**H1N1 Virus**

**Family:** Orthomyxoviridae

**Phases of Pandemic:**
- **Phase 1 to Phase 3:** Pandemic only in Animals
- **Phase 4:** May 27, 2009, Mexico
  - Human to Human Transmission at a community level in single country
- **Phase 5:** May 29, 2009, USA
  - Human to Human Transmission in a single zone US
- **Phase 6:** June 11, 2009
  - India & USA
  - in 2 different zones

**H1N1 Virus**

- ssRNA virus, enveloped
- Viral Family: Orthomyxoviridae
- Size: 80-200nm or 0.08-0.12 μm in diameter
- Three types: A, B, C

**Surface Ag's:**
- H (Hemagglutinase): binds to host cells & enters host cell
- N (Neuraminidase): Infects vision, escape out of cell & spread infection

- Oseltamivir & Neuaminidase

- H → 16 Types
- N → 9 Types

- H1N1 → Human, Swine, Ducks
  - Virus got reassorted in swine

**Virus has segments**

1. North American swine virus get reassorted
Complications:
- Pneumonia
- ARDS like illness

Risk factors:
- High risk of groups
  1) Extreme of age like <1 yr or >65 yrs
  2) Pregnancy
  3) Heart
  4) Chronic illness
  5) Immunosuppression, post transplant, nephrotic syndrome

Doc is Oseltamivir: for H1N1

Indications of Oseltamivir:
1) To all suspected cases
2) To all confirmed cases
   • Throat swab
   • Nasopharyngeal swab
   • Report in 8 hrs
3) Contacts, household, occupational

Dose of Oseltamivir:

Adults: Rx Prophylaxis

Adults: 75mg cap. 75mg cap od
   2 x 5 days till 10 days after last contact

Children 15 kg or less: 30 mg od
   60 mg/day divided into 2 doses
   15-23 kg: 90 mg/day 45 mg od
   divided into 2 doses
Doc. for H. Influenza - Ceftriaxone

25-40kg: 150mg/day
60mg OD
divided into 2 doses

> 40 kg: 150mg/day
75mg OD
divided into 1 doses

Oseltamivir dose remain same, when we are using anti TB because it is not metabolised by liver, it is excreted by kidneys. So in renal failure, dose of Oseltamivir is reduced.

Oseltamivir is not Teratogenic.
Oseltamivir is Teratogenic only in cats.

IMNCI Program For Resp. System

8/5x
Classification Therapy Where to Treat

1) Cough or cold
   No fast breathing No Pneumonia Home remedies Home
   No chest drawing
   or indicators of severe illness

2) Resp. rate Pneumonia Cotrimoxazole Home
   RR/minute age
   60 or > 60mm
   50 or > 2-12mm
   40 or > 12-60mm

3) Chest indrawing Severe pneumonia IV Imipenem Hospital Ceftriaxone

4) Cyanosis (central) Very severe P I/IV Chloramphenicol Hospital
A 10 month old child weighing 5 kg & 68 cm 55
in ht. present cough RR 48/min
no chest indrawing & grunt. The baby is
NO PNEUMONIA.

1) WOF statement is not True
6 wk old child has Fever & cough RR 44/min
wheezing + on auscultation
1) Treat Fever
2) Treat wheezing
3) Antibiotics not required

This is Pneumonia NOT Pneumonia. It is
Viral Bronchiolitis

**Bacterial Pneumonias**
causes of Pneumonia
1st 2 mon of life - klebsiella, E.coli, staph
Blw 3 mon = 3yrs - Pneumococci, H.Influenza
staphylococci
After 3 yrs - Pneumococci, staphylococci

**Neonatal Pneumonia**
Community acquired Hospital acquired
Ampicillin + Gentamycin occurs in Hospital
occurs in home. Cefotaxime + Amikacin

**PNEUMOCOCCI**
Causes lobar Pneumonia
I.P => 1-3 days
=> Lobar Pneumonia is severe pneumonia.
Dr. for Pneumococci is Ceftriaxone
McC of Pneumonia, meningitis (all over world) — Penicillin

Doc for resistant Pneumococci — Vancomycin

Vaccination against Pneumococci: - PPV - 23 only given to children > 2 yrs
Children < 2 yrs - can't mount Ab's against Ag's
- So conjugated vaccine is given
  PCV - 7/11 (PREVANCE/WYETH)
given in 6, 10, 14 wks
  15-18 mon Booster
  DPT vaccine is conjugated vaccine
  Conjugated to Ag
  Again, we have to give booster dose.
  Single dose of PCV - 7/11 is costly so not given to child in India.

GAVI — Global Alliance for Vaccines & Immunization
  By Bill Gates & malindagotees
  GAVI is given to every child in Pakistan free of cost

Most Pneumonia & max. mortality is Staphylococci
  of 10-30%
  Staph

Pneumatocele:
  Air filled cavities in lung parenchyma
  If pneumatocele rupture cause pneumothorax
  Also cause by Klebsiella
  Also due to kerosene oil poisoning (kerosene - hydrocarb

Staph. is MCC of Empyema —pus in pleural cavity

Empyema Rx: — I.C.D. Tube drainage
Atypical Pneumonia

1. Symptoms: a persistent dry cough
2. rare < 4 yrs
3. This is Interstitial, B/L
4. chlamydia, mycoplasma cause this

Doc: macrolides (Azithromycin, Erythromycin)

Upper airway Infections

Child has fever & Stridor

Mucous IT's is Group B Haemophillus.

1. Group B:
   - 75% caused by Parainfluenza
   - In prodrome period → Stridor
   - Barking Cough
   - X-Ray: Steeple sign

Rx of Group B:
- mild-moderate: Dexamethasone 0.6 mg/kg
- severe: Nebulized racemic epinephrine

A 2yr old child brought to emergency at 3:00 am. He has barking cough, Stridor only during crying. Hydration OK. RR 36/min. Temp 39.6 degrees. Satur 96%. The next step is high dose of Dexamethasone.

→ A 6yr old boy is brought to emergency room 7 a 2Hr of 3Hr of fever to 39.5°C (103°F).
- Sore throat. Child appears anxious & toxic. He inspiratory Stridor x drooling
- you should immediately prepare to establish an airway (Tracheostomy)
EPIGLOTTITIS

2.

Epiglottitis:

World: S. pyogenes
S. pneumoniae
S. aureus

India: H. influenzae

Toxic: Drooling, Stridor, Stridor,
Fever, Impending Resp Failure:

Bacterial:

DOC: Ceftriaxone + Salbactam (B-lactamase)
Caused by ESBL Organisms

Laryngeal diphtheria: have same symptoms:

Toxic, drooling, Stridor, Fever, Imp Resp Fail

SMALLER airway Infections

Broncholitis:

- Inflammatory obstruct of smaller airways
- Caused by RSV (50%)
- Occurs in children < 2 yrs

Risk Factors:

* Preterms, CHD, Lung disease, Cho,

Top fed (Breast milk IgA - RSV) protects against
Broncholitis

Rα1/4 Leads to air trapping

(like in asthma)

X-Ray: Interstitial spaces wide

Hyper Inflation of lungs

→ After a prodrome, This child has wheezing / Rhonch
Doc for Bronchiolitis is Humidified O2
- For high risk groups like Preterm, CHD pt. we give nebulized Ribavirin i.e. Palivizumab - humanized monoclonal AB against RSV

- Prognosis: Excellent
  1% mortality

Persisting wheezing during infancy is
Reactive airway disease (Asthma)

Foreign body aspirat

2 year old child, acute severe asthma (status asthmaticus), Rx T O2, nebulised salbutamol.
No recovery then nebulised Ipratropium Bromide
No recovery then 8/c Terbutaline not recovered
I/v Hydrocortisone given no recovery
Terbutaline infusion is given continuosly no recovery
I/v-cromophylline given if no recovery 50% mgsq is given no recovery.

- If a baby has wheeze in 1st episode think about foreign body aspirat

- Do bronchoscopy for foreign body aspirat

Rx

ASTHMA

Notes from Jain Stationery

Gautam Nagar
09654691327
<table>
<thead>
<tr>
<th>Step</th>
<th>Sympt</th>
<th>Night Time Sympt</th>
<th>PEFR, (N = 78%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Intermittent</td>
<td>&lt; 1 time a week</td>
<td>≤ 2 times a month</td>
<td>&gt; 80% predicted</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Variability &lt; 20%</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Repeatability</td>
</tr>
<tr>
<td>2. Mild</td>
<td>&gt; 1 time a week</td>
<td>&gt; 2 times a month</td>
<td>&gt; 80% predicted</td>
</tr>
<tr>
<td>Persistent but &lt; 1 time a day</td>
<td></td>
<td></td>
<td>Variability 20-30%</td>
</tr>
<tr>
<td>3. Moderate</td>
<td>Daily use β-agonist</td>
<td>&gt; 1 times a week</td>
<td>&gt; 60% &amp; ≤ 80% pred</td>
</tr>
<tr>
<td>Persistent daily attacks affect activity</td>
<td></td>
<td></td>
<td>Variability &gt; 30%</td>
</tr>
</tbody>
</table>

**Severe**: Continuous, Limited Frequent ≤ 60% predicted

**Persistent**: Physical activity

**Classification**

--- Long term prevent

Intermittent

Inhaled Short acting β-agonist as required for symptomatic relief

If needed > 3 times a week move to step 2.

Mild Persistent

Inhaled Short acting β-agonist as required + inhaled Budesonide

Fluticasone or Bexomethasone 100-200 mg

Inhaled Steroid or Cromolyn

Na or Sustain release Theophyllin

Moderate Persistent

Salbuterol + Sustain release Theophylline + above

Severe Persistent

Oral low dose Prednisone on alternate days + above
Recurrent Pneumonia: 2 episodes of radiographic pneumonia in 1 yr or 3 episodes in any time frame.

Persistent Pneumonia: persistence of symptoms. Radiographic (x-ray) abnormality for > 1 mon.

Recurrent Pneumonia:
- Causes:
  - L → R shunts
  - GERD
  - Immunodeficiency
  - GERD: 24 hr pH monitoring.

\[ \text{pH} \]

\[ \begin{array}{c}
\text{8} \\
\text{6} \\
\text{4} \\
\text{2}
\end{array} \]

\[ \text{am} \rightarrow \text{am} \]

\( \rightarrow \) \text{Feeding H}_{2} \text{O}_{2} \)

MC of Recurrent Pneumonia is Cystic Fibrosis

1/2500 in UK, chr. +79.

1/15 in Ashkenazi Jews — carriers.

MC lethal genetic disorder in Caucasian is Cystic Fibrosis — MUCous.

1) Gene: CFTR

\( \wedge \) codes for chloride channel
mutat° in CFTR → mucus accumulates

↓

Cause Problem

mc presentation of CF [↑ Glycogen] → Pneumonia

most imp. system affected in CF is resp system (inflamed)

1) mc organism Pneumonia in CF is Pneumonia.

2) Endocrine Pancreatic Insufficiency

presents as Steatorrhea (Fat malabsorption)

foul smelling bulky stools - mc in children < 5 yrs

Rx: Supplementation of enzymes (Amylase lipase)

A 1 yr old girl presents with fever since 24 hrs a day, coughing, she is passing foul smelling, bulky stools, had 4 attacks of bronchitis in past. arieri - CF

NOD statement not true about CF

a) AR

b) AB° CFTR which leads to defective Transport

active Cl° in CFTR requires ATP.

c) mutat° in CFTR

3) Endocrine: - 25% IDDM after 35 yrs.

4) meconium ileus

5) Azospermia

6) Nasal polyps

A 40 yr old male presents with infertility. His testis are in shape but was not palpable. Semen analysis revealed azospermia. Detailed examination showed low vol.

low pH, low viscosity with high clast empathy time.
1) Burkholderia Cepacia cause fatal (sepsis) organ - 2.5%.
2) Achromobacter - 4.5% cases

Rx: Ceftazidime
Piproz
Atofpazone
In America: Inhaled antibiotics like Tobramycin, Aztreonam
inhaled; nebulised colistin, Ciproflox, Amikacin, levoflox
Dornase a (Pulmozyme) breaks mucus bonds.


Asthma Criteria for Asthma CF:

1. Cystic Fibrosis Criteria for CF:

1) Sweat chloride > 60 meq/l on 2 occasions (8) < 40 meq/l

2) Finding 2 known CTR mutations

3) Best - finding nasal electrode - potential difference (NEPD)

A child presents raised chloride 32 meq/l & 42 meq/l

- Suspicion of CF. Other test that is used to exclude

- Repeat sweat chloride

- DNA analysis for AF508 mutation

- NEPD

- Fat in stool for next 72 hrs

- Disphagia Lusoria

- Male, 10 mon boy, Down Syndrome & Recurrent pneumonia

- Aberrant 2 subclavian a. Compressing esophagus like

- Vascular ring - Barium swallow

- H-Shaped TE Fistula

- 18 mon boy, recurrent pneumonia & feeding difficulty

- Chocking

**Stomach**
In CF - vas is absent in 98-100%
In infertile males - vas is absent in 1%

→ In CF - 26000 pt. effected in USA
CFTR gene on 7q
There are 1800 mutatn phenylalanine
MC mutatn in CF is AF 508 in 70% Caucasians
Phenylalanine mutatn Seen only in 25-30% in
508 Indians children

→ 3 nucleotide deleted in CF
(TrTr)

ΔF 508 mutatn is class II type mutatn
it cause Trafficking defect

LUMECAT: CFTR Corrector
IVA CAT: CF Potentiator

M2 organism are cystic fibrosis

Pseudomonas (non mucoid)

b) Burkholderia, Capacea

CF & Pseudomonas is severe in female
(Pseudomonas undergoes mucoid transformatn)
in presence of estrogen

Cystic Fibrosis:
< 5yrs - S. aureus > H. Inf.
5-18 yrs: - S. aureus > Pseudomonas
> 18 yrs: - pseudomonal > S. aureus

Pseudomonas in Throat becomes mucoid (40-50%)

F/6
I. X is: TracheoBronchoscopy. For TEF

2) A 4-year-old E Hx of TEF. Presents with cough. The cough has persisted since he was discharged from hospital following TEF repair. Cough - dry, barking & occasionally asis & expiratory wheezing. ASIS

a) Subglottic stenosis
b) Tracheomalacia - do bronchoscopy. We see Stridor. Wheeze

c) Sinusitis

d) Cough variant asthma

Imp in Diarrhea

<table>
<thead>
<tr>
<th>Mmol/L</th>
<th>WHO-ORS</th>
<th>osM (molal)</th>
<th>LOR</th>
<th>ORS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>311 mosm/kg</td>
<td>(a hydrion salt</td>
<td>Low Osmolality</td>
<td></td>
</tr>
<tr>
<td>Na (mEq/L)</td>
<td>90 (cholera)</td>
<td>45</td>
<td>75</td>
<td></td>
</tr>
<tr>
<td>K+ (mEq/L)</td>
<td>20</td>
<td>40</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>Glucose</td>
<td>111</td>
<td>125</td>
<td>75</td>
<td></td>
</tr>
<tr>
<td>Chloride</td>
<td>80</td>
<td>70</td>
<td>65</td>
<td></td>
</tr>
<tr>
<td>Citrate</td>
<td>10</td>
<td>-</td>
<td>10</td>
<td></td>
</tr>
</tbody>
</table>

NaCl 2.6 gms
KCl 1.5 gms
Citrate (C6H4(NO3)3) 2.9 gms
Glucose anhydrous 13.5 gms

Total 20.5 gms

Notes From
JAIN STATIONERY
09654691327
Diarrhea.

No dehydration  Some dehydration  Severe dehydration

Rx: For every lost stool  75 ml/kg ORS over  Ringer lactate 10 ml
    use 5-10 ml/kg ORS  4 hrs

30 ml/kg 70 ml/l

< 1 yr  1 hr  5 hrs
> 1 yr  1/2 hr  2.5 hr.

Every diarrheal child should be given Zn (acute phase)

Zn = 2 RDA X 2 wks

Recommended allowance

DNB RDA Diarrhea

> 6 mn 10 mg  20 mg/day
< 6 mn 5 mg  10 mg/day for 2 weeks

Fluids

Isotonic

0.9% NaCl  Ringer lactate (mEq/l) - It is like plasma

154 mEq/L Na⁺  Na⁺ → 150
154 mEq/L Cl⁻  K⁺ → 4
Cl⁻ → 109
lactate → 28
Ca²⁺ → 3

Maintenance Fluids

Type of fluid we use:

N/3 or N/5 + 5% Dextrose + 20 mEq/L K⁺

50 ml 30 mEq/L (15 mEq/100 ml)
At 5 mon - child doubles his wt.

Vol. of maintenance fluid:

1 - 10 kg → give 100 ml/kg/day → 40 ml/hr
11 - 20 kg → 1000 ml
+ 50 ml/kg addition >10 kg
21 kg onwards: 1000 ml - 1st 10 kg
+ 500 ml - 2nd 10 kg
+ 20 ml/kg additional >20 kg

(For 25 kg - 1600 ml - given) [Note: 500 ml (20x3)]

GROWTH

Avg. wt. gain in 4 mon

90 gm/day - 1-4th mon
20 gm/day - 5-8th mon
10 gm/day - 9-10th mon

wt. multiple

X2 - 5mon
3 - 1 yr
4 - 2 yrs
5 - 3 yrs
6 - 5 yrs
7 - 7 yrs
10 - 10 yrs

Length

At birth: 50 cm growth velocity
1 yr - 75 cm → 25 cm
2 yr - 90 cm → 15 cm
4 yrs - 100 cm

Add 5 cm/yr till 10 yrs puberty

These will be pubertal spurt
Somatic growth:

Sigmoid shape: → Pubertal spurt

In school going children, avg. ht. velocity/yr is

a) < 5 cm
b) 5-8 cm
c) 9-12 cm
d) 13-16 cm

Head Circumference

At birth → 35 cm
3 mon → 40 cm
1 yr → 45 cm
2 yr → 48 cm
12 yrs → 52 cm

90% of brain growth occurs at 2 yrs.

Puberty

GONADAL GROWTH occurs at puberty.

LYMPHOID GROWTH:

Physiological lymphoid hyperplasia occur by 4-8 yrs
Transitions don't occur. It
Hyperplasia is ⑨

NOTES FROM
JAIN STATIONERY
GAUTAM NAGAR 09654691327
PUBERTY (behaviour - paradoxical)

ADOLESCENCE - 10 - 19 yrs.

Early adolescence - 9 - 13 yrs. (Tanner's sexual maturation age 1 - 2)
- 2nd sexual characters

Mid adolescence - 14 - 15 yrs. (SMR - 3 - 5)
- Menarche
- Peak of growth vel.
- Acne
- Sexual drives

Late 16 - 19 yrs. (SMR - 5)

SEQUENCE OF PUBERTY

Girls:
- Thelarche → Pubarche → Menarche

Boys:
- Testis enlargement → Penis → Pubic Hair → Pubic Hair
- Axillary Hair

Girls:
- Breast development
- Pubic Hair

Boys:
- Genitalia & Pubic Hair
- Breast Not developed

Tanner Stage 1 - Testis Small Size
- NO Growth

Tanner Stage 5 - Mature Testis (Adult Size)
- Complete Growth
- Mature Breast

Imp. Points - SMR
- Stage 1 - No Sexual Growth
- Stage 5 - Complete Growth

1st Sign of Sexual Maturity

Boys - Enlargement of Testis
Girls - enlargement of breasts
Males - Proader's orchidometer for testicular size.
Testis volume:

Stage 1 → 1.4 mL
Stage 5 → 20-25 mL.

Girls:

h-t velocity

wt

SMR

1

2

3

4

5

menarche

Boys:

h-t velocity

wt


4

Most rapid h-t found in Tanner smr stage 4 in girls & stage 5 in boys.

Abnormalities of Puberty:

Delayed Puberty

No 2° sexual characters
MCC of delayed puberty is constitutional growth
14 yrs - boys
14 yrs - girls; no menarche by 16 yrs.

Precocious Puberty

Premature appearance of 2° sexual characters
Puberty occurs by pulsatile release of GnRH hormone
Mostly precocious puberty is due to gonadotropin dependent / central
Idiopathic
Craniospinal irradiation → Endocrine eff... is Panhypopituitarism.

- mals - (mri) organic disorders
  - Hypothalamic hamartoma
  - Craniopharyngioma, hydrocephalus
  - TB meningitis.

**SHORT STATURE**

We compare child to 50th percentile

> 2SD > 2 SD below mean & < 3rd percentile

1. Mid parental ht. = adult predicted ht

Boys = avg parents + 6.5 cm
Girls = avg parents - 6.5 cm

2. Types:
   1) Physiological → a) constitutional
      b) familial
   2) Pathological → a) malnutrition

PAPERISTO
of child has GH deficiency, check for other hormones may be hypopituitarism.

b) Chronic illness - liver, heart, kidney.

Certain causes of short stature in girls - Turner's

1) Endocrine Causes:

1) Hypothyroidism: → US/LS ↑ to that age.
   - Ins Thyroid helps for skeletal maturity
   - Disproportionate growth in hypothyroidism

2) Cushing syndrome: Birth wt. & wi Birth length (N)
   - GH ↓ At around 1-2 yrs he develop lag.
   - Bone age is delay
   - Doll like face
   - Hoarse voice

Tests: GH Stimulate test

- Insulin
- Clonidine
- Arginine

- After any stimulii, take 2 samples.
  - 1. Basal sample

Most specific test to use GH deficiency - IGFBP

US FDA: Subcutaneous GH Recombinant GH

Indicated to give GH:

1) GH deficiency
2) Turner's
3) Prader-Willi
4) Crohn's failure
5) HT > 2.25 SD below mean.

MC S/E of GH administration is fluid retention.

Physiological causes of short stature:

1) Constitutional
2) Familial
Bone age in puberty is (○) in familial short stature.

**PHYSIOLOGICAL**

**CONSTITUTIONAL**

- A. wt. (N)
- A. length

- Log. 6-12 mon
- Growth velocity (N)
- Ultimate ht. (N)

* Bone age delayed
* Puberty delayed

1) AOF statements: Familial short stature are
   True except:
   a) Shape of growth curve is (○)
   b) Ultimate ht. is below avg.
   c) Growth retardation is (○) from early childhood
   d) Bone age is markedly retarded (○)

2) Normal growth velocity (N)

Ht below 3rd percentile in:
   a) Familial short stature
   b) Constitutional short stature
   c) Genetic short stature
   d) Primordial short stature: These are dwarfs also
      seen in Russell-Silver skeletal syndrome

**DISPROPORTIONATE SHORT STATURE**

- US/LS
  - ↑ in Hypothyroidism, achondroplasia, rickets

  - TB spina, mucopolysaccharidosis type IV, pseudo-hyppalbuminemia

**NOTES FROM**

JAIN STATIONERY

Gautam Nagar
09654691327
**NUTRITION**

**CELIAC DISEASE**

![Diagram showing the progression from latent to symptomatic celiac disease with manifestations of mucosal lesion and genetic susceptibility.

**Genetic Susceptibility**

DQ-2, DQ-8

**Positive Serology**

Gastrointestinal manifestations (classic)

- Age of presentation: 6-24 months
- Abd. pain
- Chv. or recurrent diarrhea
- Abd. distensio
- Anorexia
- Irritability
- Failure to thrive or wt. loss

Rarely: Celiac crisis like sepsis, hyponatremia

Non-gastrointestinal manifestations:

- Age of presentation: older age of child to adult
- Dermatitis herpetiformis
- Iron deficient anemia
- Dental enamel hypoplasia

**Marsh Criteria:**

- Villous atrophy & crypt hyperplasia seen in active celiac disease

- If you restrict gluten, mucosa of intestine is (N)
2x Test: IGA antireticulin AB's
↓
IGA tissue transglutaminase

Rx: Only Rx for Celiac disease is gluten-free diet
- Strict, life-long diet
- Avoid: wheat, rye, barley, oat (contaminated with wheat)
  Gluten should be given <50 mg/day.

Plasma Osmolality = 2(\[Na^+\] + Glucose + BUN)
= 290 mosm/kg @ 18 / 2.8

Na = 125, glucose = 108, BUN = 140 mg/dL
Ar = 306 mosm/kg

PEM
Classification:

1) Indian Academy of Pediatrics, classified by
   Wt/age > 80%
   I  41-80%
   II 61-70% "kwashiorkor"
   III 51-60%
   IV ≤ 50%

2) WHO classification based on
   moderate       severe
   wt. for ht. 70-79% ≤ 70%
   > 80% acute/wasting
   ht. for age 86-89% ≤ 80%
   > 90%

PAPERS gunning
1) All symmetric edema
2) ""
KWASHIORKOR (deprived child)

1) Edema
2) Hepatomegaly
3) Flaky paint dermatosis
4) Flag sign — alternate pigmentation of hair
5) Not good appetite (V)

MARASMUS

K/A WASTING
Last muscle to last fat is buccal mucosa.
Sensorium is alert.
Appetite is voracious.

Age-Independent Criteria for malnutrition

Kenacathi & McLaren Index: midarm circumference

mid-arm circumference 1-5 yrs

Name of Index calculator: 2 value value in malnutr

Kanawathi & McLaren

Rao & Singh
Dugdale
Quaker arm circumference
Measuring stick (Quac stick)
Tietze’s ratio — Ratio <1 in a child (1yr)

S.A.M.

(Severe Acute Malnutrition)

Among children — 6-59 men of age is defined
by WHO & UNICEF as any of following:
with growth reference:

2) visible severe wasting
3) Presence of bipedal edema
   mid arm circumference below 11.5 cm

This classification is used to identify children at high-risk of death. Children having SAM require urgent attention in hospital.

- In a child below 6 months of age, MUAC can't be used & SAM should be assessed in presence of:
  I, II, or III (any)

- In Africa, Ready to Use Therapeutic Feed (RUTF) should drink:
  >10 kg → at least 500 ml

Complications of malnutrition (WHO):
Sugar: <54 mg/dL or <3 mmol/L (hypoglycemia)
Hypothermia: Temp <95.5°F, <35.5°C
Infections
Electrolytes: Resomal ↓Na ↑K
Dehydration
Deficiency of vitamins & minerals

Complications occur 0-7 days of infection
After 7 days mx

Begin Feeds:
- Type: milk based
- Roux-NG route, 75-100 kcal/kg/day (1.5-3 g/kg)
- Energy dense: after 7 days, 250-300 kcal/kg, 2-3 g protein/kg

'Stimulation'
Tender, love & care

JAIN STATIONER: 07654691327
Criteria for discharge:

- wt/ht reaches 90th%
- antibiotics completed
- immunization completed
- appetite good
- edema -ve

Now he can go home

CNS

Microcephaly

Head circumference > 3.5 SD below mean

Macrophaly

HC > 1.5 SD above mean

Microcephaly:

Causes:

- Genetic
  - Cri du chat syndrome
  - Trisomy 13, 18, 21

- Acquired
  - Maternal
    - TORCH, DM
    - HIE
  - Alcohol
  - PKU
  - Radiat
  - Malnutrition
  - Hyperphenylalaninemia
  - Meningitis
  - Encephalitis
  - (>6 mg/dl)
  - Phenytoin

Macrophaly:

- Macrophaly is seen in
  - a) metachromatic leukodystrophy
  - b) adreno leukodystrophy
  - c) Canavan's disease
  - Neurodegenerative disorders
Neurodegenerative disorders

Grey matter disorders  white matter disorders

Grey matter Degeneration

Appears (N)
They have some gradual problems in later life
Regression of milestones:
They can be deaf, blind, unexplained anemia,
hepatosplenomegaly, cherry red spot on macula
microcephaly  macrocephaly

1) Gm1 gangliosidosis GM2
- galleria
- die of seizure
- Tay-Sach's (brain has ab @ grey)
- Sandhoff's

- Gaucher's: - B-glucocerebroside
- Neisamm Picks: - Sphingomyelinax

--- Tay-Sach's Sandhoff
- AR, Asklenazi - Jews
- B-hexosaminidase - A def. def. B-hexosaminidase A & B
- 6mon ----> exaggerated startle
- hyperacusis - cherry red
- no organomegaly Organomegaly V

White matter Degeneration

Presents C:
Frequent Falls
Gait incoordinated
UMN Signs
- micro\'N  macrocephaly
1) KRAEPELE'S disease 2) Canavan's disease (ab with)
2) XLR: adenosinekaryodystrophy
3) Alexander's disease

PAPERISTO - Class H: hyperpigment
3) metachromatic leukodystrophy.

Hydrocephalus

Enlarged ventricles or out increase in intracranial tension.

CSF Product:
- 75% of CSF - lateral, III & IV ventricles
- 25% of CSF - Extrachoroidal - Capillary endothelium in brain parenchyma.

Rate of CSF production - 30 mL/h.

CSF volume:
- Infants - 50 mL
- Adults - 150 mL

CSF Flow:
- Lateral ventricles
  - Frontal
  - Temporal
  - Occipital
- III ventricle
- Aqueduct
- IV ventricle
- Luschka's
- Magendie
- Basal cisterns

Obstructive hydrocephalus

1) Aqueductal stenosis - MCC
2) Aqueductal gliosis
3) Arnold-Chiari malformation - downward displacement of hyperplasia of cerebellum

- Obliteration of cisterna magna.

Type I
- Adults
Type II
- Newborns (Lumbosacralmyelomeningocele)

adolescents
MCC of hydrocephalus (Overall)
Post Inflammatory Obstruction of Basal Cistern

3) Dandy-Walker Syndrome

- Large cyst in post fossa. - Communicating "BAT WING APPEARANCE" in ventricles (vermis)
- Vermis aplasia or Cerebellar hypoplasia

- A newborn presents with congestive heart failure
  - Bulging fontanelle
  - Bruit on auscultation
  - Transfontanelle USG shows a hypoechoic midline mass & dilated lateral ventricles

  a) Medulloblastoma
  b) Encephalocele
  c) Vein of Galen malformation (AVM)
  d) Pneumocystis cyst

4) Vein of Galen cause Obstructive Hydrocephalus
   - IX of choice - MR angiography.
   - Vein of Galen drains into straight sinuses

Non Obstructive / Communicating Hydrocephalus

Ventricles are communicating

- TB have basal
- Cryptococcal meningitis exudates
- Periventricular disease
- Basal Cisterns
All ventricles are dilated.
Enhancing basal exudates in basal cisterns.

WoF can distinguish hydrocephalus due to aqueductal
stenosis when compared to Dandy Walker malformation

Posterior fossa vol.

b) In ventricle size
c) Lat. ventricular size
d) Head circumference

Rx: Drugs - Furosemide, Acetazolamide.
   If drugs are not effective - Subcutaneous Shunt
   Ventriculoperitoneal Shunt.
   (Excessive CSF from ventricles → goes to peritoneum)

Indications of Shunts:
- Paucihypothalamic thinning
- Periventricular oozes

Complications of Shunt:
- Blockage
  - Infected
  - MC Organism that affects Shunt: Staph. epidermidis
    (Coagulase -ve)

EPILEPSY

Recurrent unprovoked unprovoked seizures not r/t fever
or acute cerebral insult.

Partial → Generalized

Simple complex

→ Loss of consciousness
  → aura, automatisms (involuntary orofacial movements)
Doc for Partial seizures is
- Ox Carbamazepine or Carbamazepine
  - Ring enhancing lesions

Necrocytis / Tubercoloma
- Usually they are solitary
- These are usually large >20 mm
- Multiple, irregular
- Perilesional edema
- Mid-line shift
  
Rx:
- Prednisolone (dexa x 3-5 days)
- Albendazole

Generalized Epilepsy

a) Tonic (↑ tone)
b) Clonic (rhythmic movements)
c) GTCS → Typical GTCS has aura, frothing, uprolling of eyelids
  
Post ictal phase

d) Atonic (sudden loss of tone
  - Myotonic (rhythmic jerks all over body)

Rx: Valproate
- Caution: <2 yrs, valproate is hepatotoxic drug

Absence Seizure

- Blank stare <30 sec (typical Absence seizure lasts <15 sec)
- No aura
- No post ictal phase
- Hyperventilation provokes
- Common after 5 yrs
- Common in females
Generalized.

Every second 3 spikes \( \rightarrow [3 \pm 1] \) spike.

Rx: ethosuximide (in children)

valproate

A boy was noticed to have persistent blank lasting 30 sec to 1 min several times a day.

most likely cases:

a) Absence Seizures

b) Day dreaming \( \rightarrow H_x \) Present

Absence seizures: True

Seizures brief, lasting < 30 sec.

typical age onset 1-2 yrs of age 5 yrs

Infantile Spasms:

SALAAM SEIZURES/ WEST SYNDROME

SeeN to 4-8 mon.

B/L Flexure Symmetrical contractions of head, trunk & extremities.

Idiopathic (cryptogenic) \( \rightarrow [3] \)

Good prognosis

very bad prognosis

85 - 90% chance this baby TO develop m. retardation.

\( \rightarrow \) develop:

a) HIE

b) Structural ab of brain

c) a° to Down syndrome

d) a° to Tuberculous sclerosis

EEG] is very imp - Hypsarhythmia.

Generalized chaotic pattern of high voltage slow wave

occur because of CRH hormone
Juvenile myoclonic Epilepsy  

1) 12-18 yrs  
2) In morning - myoclonic jerks occur usually  
3) Present c GTCS - in 90% children, only 10% Absence seizure  
4) This is life long disease, no remission  
5) Rx: DOC - VALPROATE - excellent response  
6) Complete remission is rare  
7) Response to anticonvulsant is v. good  

FEBRILE SEIZURES  

MC seizure in childhood  

Typical Febrile Seizure  
Atypical  
9 mos - 5 yrs  
GTCS  
last < 10 min  
have only one seizure/febrile episode  
RISK OF EPILEPSY IS ~ 33%  
Partial, complex  
> 15 min  
multiple, recurrent  
Risk of epilepsy - 1%  
Recurrence risk - 30-50% in atypical  

→ Predisposition to get epilepsy in late life  

after febrile convulsions are all except  
1) Complex febrile convulsions  
2) Family Hx of epilepsy  
3) Age of onset → risk of recurrence  
4) Mental retardation - 33%  
5) Seizure T in 1 hr of fever  
6) Recurrent  

Prevention of febrile seizures:  
1) Intermittent prophylaxis  
   Oral Pcm + Oral diazepam (PO)  
   Oral clobazam  
2) 48-72 hrs
TO abort febrile seizures in home:

1) A-B-C

2) Per rectal diazepam

**STATUS EPILEPTICUS**

Continuous convulsions lasting > 30 min (R)
Convulsions b/w c there is no return to consciousness

→ Apex somnolent, most vulnerable area of brain to hypoxia
   is Hippocampus, Amygdala, Thalamus & Subcortical areas

Mx & C

→ ABC

→ i/v lorazepam - DOC
   Faster onset of act, longer act

- (R)

i/v diazepam + phenytoin 20mg/kg
   ↓ refractory

Repeat phenytoin 10mg/kg → 10mg/kg
   ↓ refractory

i/v Phenobarbital 20mg/kg → 10mg/kg
   ↓ refractory

Midazolam infusion 2-20 μg/kg/min
   ↓ refractory

i/v valproate 20-30mg/kg
   ↓ refractory

i/v Levetiracetam
   ↓ refractory

General consultation (Gthiopental, Thio)
Meningeal signs are rare < 18 mon

- All are given in Status asthmaticus except
  - a) Phenytoin
  - b) Chlorapam
  - c) Thiopentone
  - d) CBZ

A baby 6 mon, presents 2 fever & seizure

* Infants < 1 yr → fever + seizure → do LP → CSF examination

**MENINGITIS**

- 1st 2 mon of life → Group B & D strep: coxillb, E.coli
- 2 mon → 12 yr → Nowadays S. Pneumonia
- Inf

N meningitides (VS)

( H influenzae is NOT the MC now)

- B/w 3 mon → 3 yrs (in India) → H influenzae

**Acute Bacterial meningitis**

1. 95% of cases occur b/w 1 mon to 5 yrs
2. Defects of complement system → c5-8 &
   properdin system → meningococcal infect
3. T lymphocyte defects (eg AIDS / chemotherapy) →
   monocytophagocytic → Cryptococcal
4. Congenital / acquired defects across mucocutaneous
   barriers → Pneumococci → C. acnes → CSF meningitis
5. Lumbosacral myelomeningocele & dural sinus
   → staph & enteric bacteria
6. Penetrating CNS trauma / CSF shunt infection →
   coagulase → Staph.
Has Splenectomy or autosplenectomy. - Capsulated Pneumococcal Infection → H. Influenzae
Sickle cell dysfunction / asplenia → Pneumococcal Infection → Neisseria

Child is immunized before 2 wks before going to Splenectomy.
Recurrent meningitis in CSF, leak, patients. Me due to:

a. E. coli
b. Pneumococcus: All children who are going to
c. meningococcus: Cochlear implant should be
d. Yersinia: Immunized against pneumococcal

(Pneumococcal Infections) Risk: 30 hrs

Doc → Ceftriaxone

Ceftriaxone resistant → Vancomycin.

Now Ceftriaxone & Vancomycin is used.

→ MC neurological sequelae: - SNHL (Deafness)

→ Infection reaches cochlea via cochlear aqueduct.

We can prevent SNHL by giving Dexamethasone 0.15mg/kg
30-60 min before antibiotics usage.

Post Exposure prophylaxis to contacts & H. Influenzae:
- Rifampicin (A)
- I/M Ceftriaxone
- PQ's (Doxycycline)

ENCEPHALITIS:

1. MCC of meningocencephalitis: - 80% Enterovirus
2. MCC of sporadic encephalitis: - HSV-1 (Herpes)
   MC virus causing encephalitis in America. - West Nile Virus.
3. 5 yr old boy, fever & partial seizures since last
ON CT - B/L Temporal hypodense.
MRT - B/L Temporal lobes are hypointense.
EEG - Localised Temporal lobe affected
Δ&θs; - Herpes (This is Treatable)

DOC: i/v acyclovir
Test → CSF sent for HSV-PCR
In this CSF is haemorrhagic - seen in herpes.

HSV-1
- affects brain
HSV-2
- neonatal HSV
- He gets from mother
- affects genitalia

Acute Flaccid Paralysis
Flaccid paralysis of acute onset in children < 15 yrs

R/o → Pseudoparalysis
→ Early syphilis
→ Osteomyelitis
→ Septic arthritis
→ Hypokalemia (after diarrhea)
→ Scurvy

AFP:-

Asymmetric Paralytic Polio: - asymmetric polio
Traumatic neuritis: Injec'tn site pain (gluteal)

Symmetrical:

1) Transverse myelitis: Viral, varicella, herpes/mycoplasm
   a) Level - sensory/motor level
   b) Doc - high dose steroids 1/2 methylprednisolone
2) Guillain Barre Syndrome: - (AIDP)
   Acute Inflammatory Demyelinating Polyneuropathy
rarely axonal

→ Classically have diarrhea by campylobacter jejuni, mycoplasma, Enterobacteriaceae

↓ 10 days

→ Weakness, areflexic

(DTR absent)

→ Asy. Ascending

→ Symmetrical → causing Diaphragmatic paralysis

CSF in 1st week

CSF in 2nd week

CSF 1st week 2nd week

Cells 10/µL 10

Protein 50 mg/dL 500 - Albuminocytological

Dissociation occurs in 2nd week illness

Doc: IV Ig, plasma exchange

Q) About 12 days after mild ORE, a 10-year-old boy complains weakness in lower extremities several days after weakness progressed over trunk. DTR are absent in lower extremities. CSF shows elevated proteins.

GB (classical) - Rarely they can have sensory & autonomic features.

Not true about GB

a) Ascending paralysis

Albuminocytological

Flaccid paralysis

✓ Preserved reflexes (DTR-)

Ix OC for calcifications - CT scan
**Endocrinologic Disorders**

**CAH Compulsory Lab**

- mcf of female pseudohermaphrodite
- karyotype - xx
- phenotype - male

Cholesterol $\rightarrow$ DHEA $\rightarrow$ Testosterone $\rightarrow$

90% of due to

- CAD
  - 11 beta Cortisol
  - DOC
    - very imp. mineralocorticoid
    - produce from an adreno
    - aldostere ton in one of the

- 11-hydroxylase Cortisol Aldosterone
- no Cortisol $\downarrow$ Na, $\uparrow$ K
- $\downarrow$ glucose, $\downarrow$ shock

- labia are $\uparrow$ ACTH
- pigmented like scrotum

"Acute adrenal insufficiency"
"Salt wasting crisis"
"Addison's crisis"

**Case Study**

- Following discharge he had failed to gain wt. & began vomiting 3 days ago.
- This had persisted & worsened over past 2 days. Nappies are wet 2 times a day.

**ASIS**: CAH
- O/E Temp - 36.2°C, (Tympanic)
- HR - 160/min (Thready pulse), No murmur.

**ASIS** of CAH by:
- 11 Hydroxyl progesterone

- Males of CAH get precocious puberty
- Females get virilized
  1) Girls have ambiguous genitalia
  2) Hypertrophic clitoris
  3) $\uparrow$ ACTH $\rightarrow$ labia pigmented like scrotum
  4) Penis like structure
CAH: My 2 supplement Gc's & mineralocorticoids.

[Hydrocortisone & Fludrocortisone]

Girls & CAH require clitoral reducto surgery or clitoroplasty before marriage. They don't marry because of H T Testosterone.

WOF: axial CAH

- Hypopigmentation
  a) Delayed puberty
  b) Precocious Puberty in Boys
  c) Hypopigmentation
  d) ACTH ↑ pigmentation
  e) Hyporeninemia, Renin ↑
  f) Premature epiphyseal closure.

Short stature: Premature pubertal spurt (caused by Testosterone) in CAH.

CAH can cause prepubertal, conrasexual puberty.

11-β hydroxylase deficiency can cause HTN in CAH.

A 5 yr old boy has precocious puberty BP: 130/80.

Estimation of WOF help in ASS.

11-Deoxycortisol levels 11-β hydroxylase deficiency.

- DOCAS - un reliable
- 17-Hydroxy progesterone
- aldosterone

3 mon old female has normal genitalia (No KT)

Present to emergency department with Severe dehydration, hyperkalemia, hyponatremia. Measurement bid levels of which of following will be helpful.

- 17 Hydroxy progesterone cong. aldosterone deficiency
- Renin
- Cortisol
- Aldosterone synthase
GFR

1) At birth - 15-20 ml/min/1.73 m²
2) At 3 mon - 2/3 rd of adult
3) Adult values - 8 yrs
4) Tubular concentrating capacity reaches adult values at 1 yr.
5) At 36 wks of gestation → nephrogenesis is completed
   In preterm - less no. of nephrons

Q: Creatinine estimation:
Schwartz Formula 1972:

\[ GFR = \frac{k \times \text{ht (cm)}}{\text{Creatinine (mg/dl)}} \]

GFR depend on ht, muscle mass

↑ muscle mass = ↑ creatinine,
Creatinine depends on muscle mass.

→ Jaffe's reaction = creatinine < m.mass. →
best method to estimate creatinine is
enzyme assay of high performance liquid chromatography:

value of k in Schwartz Formula:
  Low birth wt infant → 0.33
  Normal infants 0-18 mon → 0.45
  Girls 2-16 yrs → 0.55
  Boys 2-13 yrs → 0.55
  Boys 13-16 yrs → 0.70

→ Schwartz method of determining of GFR in children is independent of

a) Body mass (Creatinine)
MC malignancy in child - leukemia - ALL.

- Age - (K)
- Renal Function
  - Method of estimation of creatinine (Jaffe's)
  - Estimated GFR = 4 yrs, Creatinine 1 mg/dl, ht-looms
    \[ \text{GFR} = \frac{55}{\text{For 4 yrs - creatinine 1 mg/dl is not good}} \]
    - Inulin clearance is best method to estimate GFR

MC abd. mass in neonates:
  a) Multicystic Renal Dysplasia
    - U/L - 80%, non-functional kidney
  b) Neuroblastoma
  c) Wilm's Tx
  d) Fibrosarcoma

Wilm's Tx:
- MC asymptomatic renal mass
  - 1-5 yrs age:

Oliguria
- Urine output < 400 ml/m²/day
  - Newborn < 1 ml/kg/hr

This is imp sign in GN, ARF
- Non oliguric ARF causes:
  1) Aminoglycosides - damage mitochondria of proximal
     Tubule
  2) Neonatal Renal Failure
  3) Resolving acute Tubular Necrosis as oliguria.
Adenovirus cause Haemorrhagic Cystitis

Hematuria

- Bid in urine
  - Glomerular
  - Extraglomerular
  - IgA, MPGN, PSGN
  - Stones, Cystitis
  - Idiopathic Calcium
  - Dicyclic RBC
  - Dysmorphic RBC
  - Isomorphic RBC
  - Pain, Bright-Rad
  - (RBC undergo osmotic damage when passing through glomeru)
  - COLA Color
  - Painless
  - Proteinuria

Recurrent Gross Hematuria (RGH)

- mc. is IgA nephropathy
- Others: MPGN, Idiopathic hypercalciuria

→ Recurrent gross Hematuria + poor vision due to

- Cataract + SNHL → Alport's
- Alport's: 85% - X-linked

1. Eye - ant. lenticonus - 30-40% in boys
2. Ear - SNHL - 50% in boys
3. Kidney - RGH
- Severe in males
- 75% of Alport's goes to renal failure before age of 30 yrs
- It is a Type 4 collagen α5 domain
- mostly X-linked, - AD, - AR
- only be diagnosed by Electron microscopy

- Irregular Basement membrane
- splitting of lamina densa z electron
- Basement Walny Basket weave appearance
ACUTE KIDNEY INJURY

Biomarkers:
1. Urine NGAL → Neutrophilic gelatinase act.
   Lipo calcin
2. Urine KIM-1 (kidney injury marker)
3. Urine IL-18
4. Urine L-FABP (Fatty acid binding protein)
5. Serum cystatin-C

3 Types of AKI

Pre-renal → Renal → Post-renal

No perfusion (hypoxia)
Hypotension
Hypovolemia (decreased volume)

Reversible

Indices:
Pre-renal → Renal (Tubules act on)

a) Urine Na (mEq/L) < 20 → > 40
b) Urinary osmolality < 500 → > 300

c) U/a creatinine ratio > 20:1 → < 20:1

d) Fractional excretion of Na% < 1 → > 1

Renal Causes:

Ischemic ATN → Newborns

AKI → Children, adults

ATN → Ischemia
Hypotension
Drugs
mec of death in earthquakes is

Caudal injury causing Rhabdomyolysis

Drug that can be given to every earthquake victim is NAC (Na,bicarbonate)

AKI children mec ATN > HUS

HUS /Thrombotic microangiopathy

Microangiopathic Hemolytic anemia

Thrombocytopenia

Acute Renal Failure

→ In HUS 90% develop diarrhea

Developed countries: E.coli 0157: H7

Developing countries: Shigella dysentriae I

→ In Germany, June 11, 1000 people got affected E coli

renal failure because the strain of
E.coli 0104: H4 & in sprouts

Shiga toxin cause endothelial injury

cause Thrombus: RBC breaks due to

Shear stress

Dox = Plasma exchange

(All immunopharma) Eculizumab + (5%) (both classic or alternative pathway)

worlds costliest drug

Complications of AKI:

1) Fluid overload → causes dilutional hyponatremia

2) dilutional anemia

3) Hyperkalemia cause anything else

4) Metabolic acidosis

5) ↑ PO4 → ↓ Ca
Fluid overload Rx: Restrict: Insensible losses 400 ml/m²
(sweating, etc)

HYPERKALEMIA

1) Nebulise child & salbutamol
2) Insulin + Dextrose
3) I/v Frusemide
4) I/v NaHCO₃
5) Oral Resins - KAEXYLATE - go to Colon blind + K⁺
   Polystyrene resins
6) Dialysis
7) I/v Ca gluconate

A 7 yr old girl 3 end stage renal disease require hemodialysis
4) E proxi. m. weakness, Sr. K⁺ is 7.8 meq/L × EKG
   Shows peaked T waves. WOF agents would lower her Sr. K⁺
   conc. most quickly.
   Ca gluconate I/v

- 4) Glucose + Insulin I/v - 10-15 min act.
- 4) KAYEXLATE in Sorbitol orally - 4-6 hrs
- 4) NaHCO₃ I/v 30-60 min

WOF should not be given in hyperkalemia except
   ECG changes (no "Only" given to ECG changes
   Ca gluconate I/v
1) Glucose + insulin I/v
2) Salbutamol nebulisation
3) NaHCO₃ I/v

NOTES FROM
JAIN STATIONERY
GAUTAM NAGAR
09654691387
Chronic Kidney Disease

Etiology:
- < 5 yrs - Anatomic Abnormalities
  - hypoplasia, dysplasia, obstructed PUV, malformations
- > 5 yrs - Acquired glomerular diseases (GN, HUS)

Hyperfiltration Theory:

\[ \text{GFR} \quad \text{Time} \quad \text{GFR Falls} \]

ESRD: End Stage Renal Disease
- GFR < 10 ml/min/1.73 m²
- Survival not possible. Out life long, dialysis or renal transplant.

\[ \Rightarrow \text{The best Rx for ESRD is} \]

- a) Long term peritoneal dialysis
- b) Living related donor renal transplant
- c) Long term hemodialysis
- d) Cadaveric renal transplant → In America

Before any transplant, blood group compatibility should be done; AB - Universal Recipient, O - Universal Donor.

Best cadaveric donor is a KTA victim because internal organs are O.
A cadaver can save 16 lives

Jain Stationery
09654691327
**NPHROTIC SYNDROME**

1. Proteinuria > 40mg/m²/h or > 2g/24hr.
2. Protein is mainly albumin.
3. Hyperproteinemina.
4. Edema - S. Albumin < 2.5 g/dl.
5. Hyperlipidemia
6. Cholesterol > 200mg/dl.

On LM – There is minimal changes.
E/LM – Effacement of foot processes of podocytes

Edema is due to Na+ and H2O retention.

![Diagram of kidney with ANP and other labeled parts.]

DOC → Prednisolone.

Lost in Urine:
- Albumin → Edema
- IgG → Infections
- Transferrin
- Ceruloplasmin
- TBG
- VDBP

Protein C, S, antithrombin III, lost (Hypercoagulable state)
Hyperfibrinogenemia.

8) All proteins are low except:

a) Albumin
b) Transferrin
c) Ceruloplasmin
d) Fibrinogen
**Gene** | **Protein** | **Disease**
---|---|---
NPHS-1 | Nephrin | Finnish Congenital < 3 mon nephritic
NPHS-2 | Podocin | Steroid Resistant FSGS

Doc: Calcineurin
Tacrolimus Oral
Azb
Cyclosporin

Doc in Steroid Tropic / Steroid Dependent - oral cyclophosphamide for 42 wks.

**UTI Infections**

1) **Def:** Symptoms of UTI + Urine culture showing $>10^5$ CFU/mL (significant)
2) Fever / Out Focus
3) MC Organism $\rightarrow$ E. coli
4) More common in $\varphi$ (<1yr of life, males)
5) Ascending route ($<1$ mon, hematogenous
6) Asymptomatic bacteriuria ($10^6$ bacteria) not to be treated
7) Best Urine Specimen: Suprapubic aspiration. Even a single colony of E. coli is significant
8) MC of UTI in children - VUR A sed by voiding cystourethrogram / MCU
9) Polar Scans caused by UTI $\rightarrow$ DMSA scan
10) For Function - MAG-3 Scan / DTPA
**IMP. POINTS**

- **Disorders**
  - Tocol
  - GERD
  - Achalasia Cardia
  - Zenker's Diverticulum
  - Hiatal Hernia
  - Ca of Esophagus
  - Diffuse Esophageal Spasm

- **Tests**
  - 24 HR pH monitoring
  - Esophageal Manometry
  - Barium Swallow
  - Endoscopic USG (EUS)
  - PET/CT > CECT
  - Endoscopic Bx
  - Manometry

**Overall Staging for Ca of Esophagus**

**Notes from Jain Stationery**

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Gautam Nagar
Wernicke encephalopathy - involves mamillary body of CNS

Equatorial diameter of lens - 9mm

Least sex virus in Ca of Genit - HPV-19

Germicidal efficacy of a disinfectant. Rideal Walker coefficient is measured by J or Phenol coefficient (C.)

meo of 2° amenorrhhea in India - PCOS

JAIN STATIONER

GAUTAM NAGAR

09654691387
Condition → Doc

1. Severe preeclampsia → Labetolol
2. Visceral & cutaneous → No
3. Rash → No
4. Ca of Endometrium → Progestins (mpa)
5. ADHD → Methylphenidate
6. ADHD + Tic/Tourette → Atomoxetine
7. ADHD adults → Atomoxetine
8. Doc. Tourette alone → D2 blocker Haloperidol

NOTES FROM
JAIN STATIONERY
GAUTAM NAGAR
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1. Bird Facies → Pierre-Robin Syndrome
2. Chipmunk Facies → β-Thalassemia
3. Leonine Facies → Leprosy
4. Adenoid Facies → Adenoid Hypertrophy
5. Torpid or myxedematous Facies → myxoedema
6. Mask like or → Parkinsonism
7. Parkinsonian Facies
8. Acromegalic Facies → Acromegaly
9. Cushingoid Facies → Cushing Syndrome
10. Gargoyle Facies → Hunter's Syndrome
11. Thyrotoxic Facies → Thyrotoxicosis
12. Hatchet Facies → Myotonia Atrophica
13. Flat Facies → Down Syndrome
14. Mitral Facies → Mitral Stenosis
15. Snoring Facies → Myasthenia Gravis
16. Elf-in Facies → William's Syndrome
17. Bourne Facies → Crouzon Syndrome
18. Bell's Palsy → Facial N. Damage
19. Amiodarone Facies → Caused by intake of amiodarone drug

1) Size of dormant or primordial Follicle → 0.02 mm
2) Size of 1° Follicle → 0.1 mm
3) Size of 2° Follicle → 0.2 mm
4) Size of 3° Follicle → Keeps changing
5) Size of mature Follicle → 20 mm

NOTES FROM
JAI N STATIONERY
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09654691383
Dripping candle wax sign \( \rightarrow \) melorheostosis

flowing candle wax sign

NOTES FROM
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Carriers are not shown by: Tularaemia phenomenon not shown.
- Tetanus
- Whooping cough
- Rabies
- Measles
- Rubella

**Herd Immunity** is not shown by:
- Tetanus
- Rabies

**IPV (Inactivated Polio Vaccine)**

**Isolation** is not beneficial in:
- Typhoid
- TB
- Leprosy
- STD's
- Poliomyelitis
- HepA

**HDIL**

- Life expectancy at birth
- Infant mortality rate
- Income (Real GDP)
- Knowledge
- Literacy rate

1st level:
- Health Education
- Specific protection
- Immunization
- Nutritional

2nd level:
- Access to Rx
- Disability Limited
- Pap smear
- Mammography
- Screening
- Special schools

3rd level:
- Iodine salt supplementation
<table>
<thead>
<tr>
<th>Primary:</th>
<th>Primary:</th>
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</thead>
<tbody>
<tr>
<td>Prevent of emergence</td>
<td>a) Health promotion</td>
</tr>
<tr>
<td>of Risk factors</td>
<td>Insecticide spray</td>
</tr>
<tr>
<td></td>
<td>Portable, safe water supply</td>
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<tr>
<td></td>
<td>Life style modification</td>
</tr>
<tr>
<td></td>
<td>Health education</td>
</tr>
<tr>
<td></td>
<td>Food fortification</td>
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<tr>
<td></td>
<td>Environment modification</td>
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<tr>
<td>b) Specific protect</td>
<td></td>
</tr>
<tr>
<td>- Chemoprophylaxis:</td>
<td>CP for TB</td>
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<tr>
<td>- Vit A for children's prophylaxis</td>
<td></td>
</tr>
<tr>
<td>- Wearing of goggles by widows</td>
<td></td>
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<tr>
<td>- Wearing of seat belt by car drivers</td>
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<tr>
<td>- Provision of 3 doses of OPV in early infancy</td>
<td></td>
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<tr>
<td>- Chemoprophylaxis of meningococcal disease</td>
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</tbody>
</table>

**Red Eye Causer:** GO SICK

G - Glaucoma
D - Orbital disease
S - Scleritis
U - Uveitis
C - Conjunctivitis
K - Keratitis

**NOTES FROM**

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