primed

# Pediatrics

## NEONATOLOGY

FIRST EDITION
eyadprimed

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## NEONATOLOGY

eyadprimed
FIRST EDITION
2025
VERSION 2.2

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"Two men looked out from prison bars, one saw the mud, the other saw stars."

*Which one will you be?*

# ASSESSMENT & ROUTINE CARE OF THE NEWBORN

## Introduction

***Neonatal period:*** Extends from birth through the first month (first 4 weeks) of life.

**Neonatal mortality accounts for about 65% of deaths in the 1st year of life (highest during the 1st day after birth).**

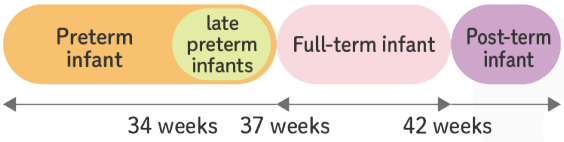
## Classifications of newborn infants

### 1. According to Gestational period

***a. Preterm:*** less than ***37 wk*** of gestation

***b. Full term:*** between ***37 and 42 wk*** of gestation

***c. Post term:*** more than ***42 wk*** of gestation.




### 2. According to Birth weight

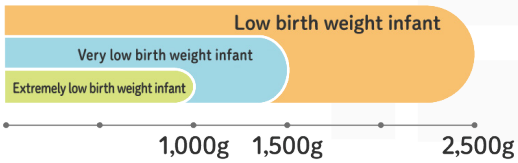
***a. Large birth weight (macrosomia):*** ***4000 gm*** or more after birth

***b. Normal birth weight (NBW):*** ***2500 to 4000 gm***

***c. Low birth weight (LBW):*** ***2500 gm*** or less

***d. Very low birth weight (VLBW):*** ***1500 gm*** or less

***e. Extremely low birth weight (ELBW):*** ***1000 gm*** or less




### 3. According to size for gestation

***a. Small for gestational age (SGA):*** birth weight less than ***10th percentile*** for gestational age

***b. Appropriate for gestational age (AGA):*** birth weight between ***10th and 90th percentile*** for gestational age

***C. Large for gestational age (LGA):*** birth weight more than ***90th percentile*** for gestational age

# Assessment for gestational age

## 1. Antenatal (obstetric calculation)

***a. History:*** date of last menstrual period

***b. Obstetric assessment*** for date of ***1st fetal heart sounds*** (10-12 wk) and ***fetal activity or quickening*** (16-18 wk).

***c. Ultrasonography.***

## 2. Postnatal

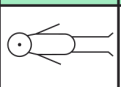
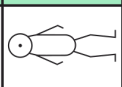
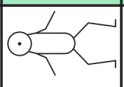
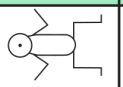
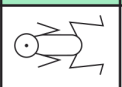
***a. Rapid delivery room assessment:***

| | Preterm | Full term |
|---|---|---|
| | **1. Measurements** | |
| **Weight** | < 2.5 Kg | 3–3.5 Kg |
| **Length** | < 47 cm | 50 cm |
| **Head circumference** | < 33 cm | 35 cm |
| **Chest circumference** | < 30 cm | 33 cm |
| | **2. Vitality** | |
| **Cry** | Feeble | Good |
| **Sucking power** | Weak | Good |
| **Muscle tone** | Poor | Good |
| **Muscle movement** | Slow, infrequent | Strong, frequent |
| | **3. Physical findings** | |
| **Scalp hair** | Fine, wool-like | Coarse, silky |
| **Ear lobe** | Pliable, shapeless, deficient in cartilage | Stiff, distinct ridges, thick cartilage |
| **Breast nodule** | Very small, < 1 cm (≥ 33 wk) | Well developed, > 1 cm |
| **Genitalia** | **Male:** Partially or undescended testis, small scrotum<br>**Female:** widely separated labia majora (female) | **Male:** Fully descended testis, large scrotum<br>**Female:** labia majora covering minora (female) |
| **Plantar creases** | Absent or only few on anterior 1/3 | Extensive, deep, allover |

***b. Accurate assessment:*** is made by combination of physical and neurological criteria in newborn e.g. "Dubowitz score", "New Ballard score", etc.

**New Ballard Score** assesses the ***Physical & Neuromuscular maturity*** of newborn infants:

## NEUROMUSCULAR MATURITY

| NEUROMUSCULAR MATURITY SIGN | SCORE | | | | | | | RECORD SCORE HERE |
|---|---|---|---|---|---|---|---|---|
| | -1 | 0 | 1 | 2 | 3 | 4 | 5 | |
| POSTURE | |  |  |  |  |  | | |
| SQUARE WINDOW (Wrist) |  >90° |  90° |  60° |  45° |  30° |  0° | | |
| ARM RECOIL | |  180° |  140 –180° |  110 –140° |  90 –110° |  <90° | | |
| POPLITEAL ANGLE |  180° |  160° |  140° |  120° |  100° |  90° |  <90° | |
| SCARF SIGN |  |  |  |  |  |  | | |
| HEEL TO EAR |  |  |  |  |  |  | | |
| | | | | | | | TOTAL NEUROMUSCULAR MATURITY SCORE | |

## PHYSICAL MATURITY

| PHYSICAL MATURITY SIGN | SCORE | | | | | | | RECORD SCORE HERE |
|---|---|---|---|---|---|---|---|---|
| | -1 | 0 | 1 | 2 | 3 | 4 | 5 | |
| SKIN | sticky friable transparent | gelatinous red translucent | smooth pink visible veins | superficial peeling & / or rash, few veins | cracking pale areas rare veins | parchment deep cracking no vessels | leathery cracked wrinkled | |
| LANUGO | none | sparse | abundant | thinning | bald areas | mostly bald | | |
| PLANTAR SURFACE | heel-toe 40–50 mm: -1 < 40 mm: -2 | >50 mm no crease | faint red marks | anterior transverse crease only | creases ant. 2/3 | creases over entire sole | | |
| BREAST | imperceptible | barely perceptible | flat areola no bud | stippled areola 1–2 mm bud | raised areola 3–4 mm bud | full areola 5–10 mm bud | | |
| EYE / EAR | lids fused loosely: -1 tightly: -2 | lids open pinna flat stays folded | sl. curved pinna; soft; slow recoil | well-curved pinna; soft but ready recoil | formed & firm instant recoil | thick cartilage ear stiff | | |
| GENITALS (Male) | scrotum flat, smooth | scrotum empty faint rugae | testes in upper canal rare rugae | testes descending few rugae | testes down good rugae | testes pendulous deep rugae | | |
| GENITALS (Female) | clitoris prominent & labia flat | prominent clitoris & small labia minora | prominent clitoris & enlarging minora | majora & minora equally prominent | majora large minora small | majora cover clitoris & minora | | |
| | | | | | | | TOTAL PHYSICAL MATURITY SCORE | |

**Reference**
Ballard JL, Khoury JC, Wedig K, et al: New Ballard Score, expanded to include extremely premature infants. J Pediatr 1991; 119:417–423. Reprinted by permission of Dr Ballard and Mosby—Year Book, Inc.

**SCORE**

Neuromuscular _____
Physical _____
Total _____

**MATURITY RATING**

| SCORE | WEEKS |
|---|---|
| -10 | 20 |
| -5 | 22 |
| 0 | 24 |
| 5 | 26 |
| 10 | 28 |
| 15 | 30 |
| 20 | 32 |
| 25 | 34 |
| 30 | 36 |
| 35 | 38 |
| 40 | 40 |
| 45 | 42 |
| 50 | 44 |

**GESTATIONAL AGE (weeks)**

By dates_____
By ultrasound_____
By exam_____

# Neonatal Examination

**Objectives of Neonatal Examination:**

1. Confirm normality
2. Identify any congenital abnormalities or birth defects
3. Detect problems of maternal or familiar diseases, or complications of labor
4. Detect any acute condition requiring urgent diagnosis and therapy
5. Provide initial health and educational advice

***Complete examination of a newborn consists of:***

- Perinatal history
- Physical examination
- Assessment of the findings

## A. Perinatal history

**Importance**

- The history will often identify clinical problems and suggest what clinical signs to look for during the examination.
- A general examination is not complete if a history is not taken.

### Maternal background:

- Mother's age, gravidity and parity.
- Number of infants (alive and dead), cause and age at death.
- Birth weight of previous infants.
- Any problems with previous infants, e.g. neonatal jaundice, preterm delivery, congenital abnormalities.
- Home and socioeconomic status.
- Family history of congenital abnormalities.

### The present pregnancy:

- Gestational age.
- Problems during the pregnancy, e.g. vaginal bleeding.
- Illnesses during the pregnancy, e.g. rubella.
- Smoking, alcohol or medicines taken.
- Blood groups.
- Assessment of fetal growth and condition.

### Labor and delivery:

- Spontaneous or induced onset of labor.
- Duration of labor.
- Method of delivery.
- Signs of fetal distress.
- Problems during labor and delivery.
- Medicines given to the mother.

### Infant at delivery:

- Apgar score & any resuscitation needed.
- Any abnormalities detected.
- Birth weight and head circumference.
- Estimated gestational age.
- Vitamin K given.
- Placental weight.

### Infant since delivery:

- Time since delivery.
- Feeds given.
- Urine and meconium passed.
- Any clinical problems, e.g. hypothermia, respiratory distress, hypoglycemia.
- Contact between infant and mother.

## B. Assessment of history

- Make an assessment of potential and actual problems after taking history and before examining the infant.
- This helps to look for important clinical signs that may confirm or exclude problems suggested by history.

## C. Physical Examination of a Newborn

**A complete physical examination is an important part of newborn care.**

- Each body system is carefully examined for signs of health and normal function.
- Look for any ***signs of illness or birth defects.***

### Requirements for neonatal examination

- Whenever possible the infant's mother should be present.
- A warm environment is essential
- A good light
- Wash your hands before examining the infant
- The infant should be completely undressed.

### The order of examination

- Measurements
- General inspection
- Regional examination
- Neurological status
- Examination of the hips
- Examination of the placenta (if available)

# Characteristics of the normal full term newborn

**Physical and physiological characteristics of a newborn are different from those of an older infant, child or adult.**

## Measurements and vital signs

***Length (average):*** 50 cm.

***Weight:*** 2,500-4,000 gm (average 3,400 gm)

***Head circumference:*** 33-35 cm. (***Molding:*** shaping of the head from passage through the birth canal.)

***Respiratory rate:*** 30-40/min.

***Pulse rate:*** 120-180 beats/min.

***Blood pressure:*** 70-85/40-60 mmHg.

***Temperature:*** at delivery, baby's body temperature is the same as his mother. A ***transient fall*** of temperature occurs after delivery and is ***restored within 4-8 hours.***

## Appearance

- Spontaneous physical activity, passive muscle tone, posture, level of consciousness and quality of the cry.
- During awake States: ***Quiet alert:*** best time for breast feeding/bonding
  ***Active alert;*** Crying.

## Body proportions

***Head*** is relatively large, face is round and mandible is relatively small.

***Chest*** is round rather than flattened antero-posteriorly.

***Abdomen*** is prominent, extremities are relatively short.

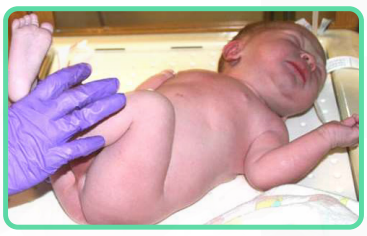
***Mid-point of the stature*** is the umbilicus (not symphysis pubis).

***Predominant posture*** is partial flexion.

## Color

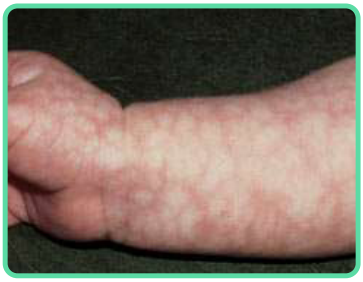
***Normal color is rosy pink.***

- ***Mongolian spots:*** purplish mottling over back & buttock (have no pathological significance).
- ***Cyanosis or pallor*** require careful examination to reveal the cause.

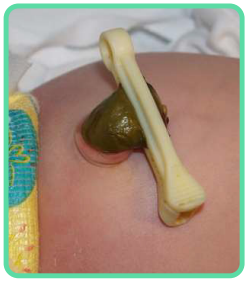


## Skin

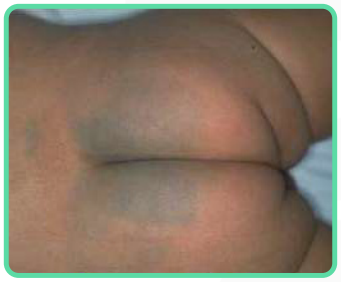
- ***Covered with vernix caseosa at birth:*** adherent cheesy white material.
- Normally ***dry skin with peeling at ankles.*** Extensive peeling and transverse fissuring over the abdomen denote dysmaturity.
- ***Lanugo hair*** cover the shoulders at birth: fine and soft.
- ***Milia*** are white spots over the nose and cheeks: distended sebaceous glands.
- ***Erythema toxicum neonatorum*** is a common rash usually between day 2–5 blotchy red spots on the skin with overlying white or yellow papules or pustules.
- ***Capillary hemangiomas*** are common over eye lids, forehead and back of neck.
- **Umbilical stump:** darkens, dries and shrinks, then ***falls by the end of the first week.***



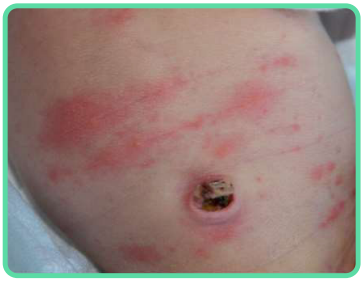
Vasomotor instability with Cutis marmorata



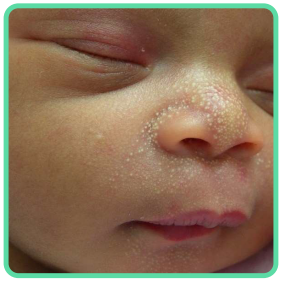
Meconium staining



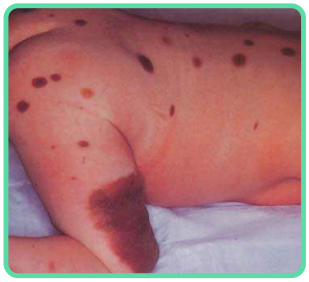
Mongolian spots

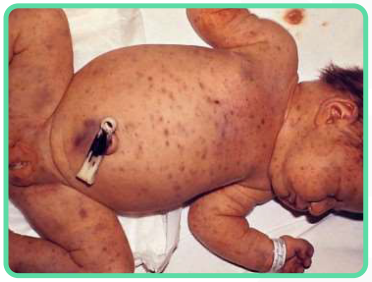


Erythema toxicum



Milia





Petechiae, Ecchymoses, Congenital Nevi

## Head & Neck

***Head:*** Size (OFC)

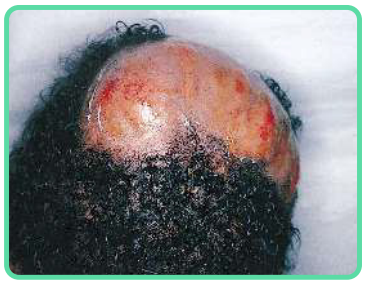
***Skull:*** Fontanels, Shape & Molding, Caput succedaneum, Cephalhematoma

***Neck:*** Clavicles & Sternomastoids, Webbing & Swellings.

***Face:*** Nevus simplex (Salmon patch), Port-wine, Strawberry and Cavernous hemangiomas

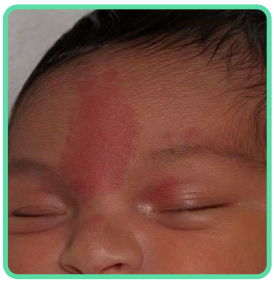
- ***Eyes:*** Discharge, Hemorrhage
- ***Ears:*** site, size & shape
- ***Mouth:*** Palate, Gums (Epstien pearls & natal teeth), Tongue (macroglossia & tongue-tie), micrognathia.

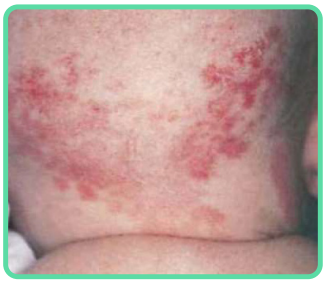
Caput succedaneum



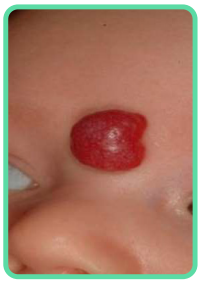
Cephalhematoma

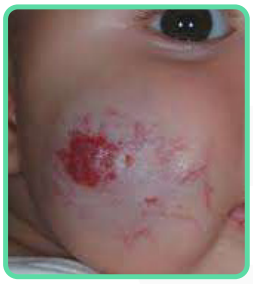
Caput succedaneum



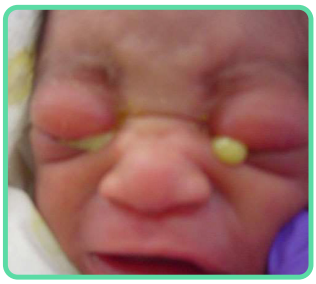


Nevus simplex (Salmon patch)



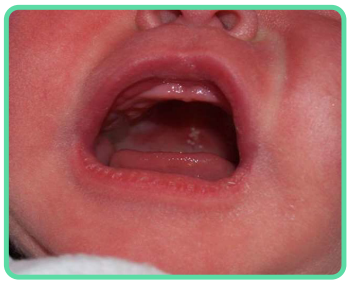


Port-wine, Strawberry and Cavernous hemangiomas

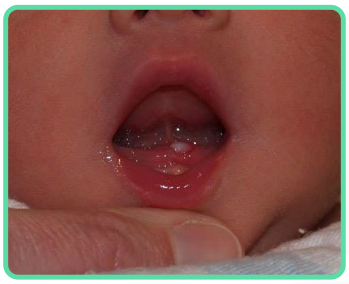


Discharge

Hemorrhage



Epstien pearls



Natal teeth

## Chest

- Shape, symmetry, position and development of nipples
- **Breast tissue size:** 10 mm at term.
- **Breast engorgement** (maternal hormones).
- **Normal respirations** 30-40 breaths/min, up to ~ 60 breaths/min

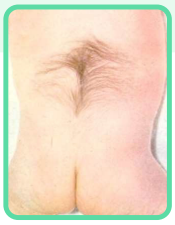
## Abdomen

- Palpate for masses/organs
- **Liver** may be palpable 2 cm below the right costal margin.
- **Both kidneys** should be palpable in the first day of life with gentle, deep palpation.
- Divarication of recti & Umbilical hernia
- Umbilical Cord Stump

## Extremities:

- Position & Movement
- Brachial plexus injury, Club feet
- Polydactyly, Syndactyly, absent fingers
- Palm & Sole Creases

## Back & Spine



- Assess for intact spine without masses or openings,
- indentation, skin pigmentation or hair tuft.
- Hair tufts, Sacral dimples, Meningomyelocele

## Anus & Rectum

- Assess rectal patency with 1st temp; lubricated thermometer.

## Cardiac signs

- Heart seems large in respect to chest shape. Apex is lateral to the left nipple line in the 4th intercostal space.
- Transient innocent ***cardiac murmurs*** may be audible which usually ***disappear later.***
- **Rate:** between 120 and 180 beats/min according to activity, crying, wakefulness, etc. A pulse rate of less than100 beats/min generally signifies low blood oxygen levels.

## Gastrointestinal activity

- The first stools are generally passed within 24 hours and consist of ***meconium stools (black).***
- By establishing breast feeding, meconium is replaced by ***"transitional stools" (greenish brown)*** on the 3rd/4th day. Later stools become golden yellow and may contain milk curds.
- **Frequency:** 3-5 motions/day by end of the first week.
- **Melena (altered blood in stools):**
  - ***Swallowed maternal blood*** (Hb A)
  - ***Hemorrhagic disease of the newborn*** (Hb F)
- Gut mucosa is immature and allows passage of macromolecules: Liability to develop allergic disease with early introduction of foreign protein into diet.

## Renal function

- First urination during 1St 24 hrs in more than 95% of normal term infants.
- Urination may be delayed 24-72 hours, depending on external atmosphere temperature.
- Kidney is not fully able to fully concentrate urine.
- ***Abundant urates*** give the urine a ***pink color and may stain the diaper.***
- ***Reduced glomerular filtration*** rate may lead to ***drug retention and toxicity.***

## Genitalia and mammary glands

- Enlargement and secretions of the breasts in both sexes (witch's milk).
- **Females:** Hypertrophy of both labia with bloody vaginal discharge may be encountered.
- **Males:** Hydrocele, Hypospadias.

**1st and 2nd conditions are secondary phenomena to the trans-placental passage of maternal estrogen, they subside spontaneously.**

### Hematological system

- Hb level averages ***17-19 gm/dL.*** Mild reticulocytosis & normoblastemia are evident.
- **Total leukocyte count (TLC):**
  - ***After 24 hrs:*** 25x$10^9$/L (with ***relative neutrophilia***).
  - ***After 1 week:*** 15x$10^9$/L (with ***relative lymphocytosis***).
- Little transfer of certain clotting factors from mother.
- Normal hemostatic mechanisms depend on establishment of normal intestinal flora and elaboration of vitamin K.

***High RBC mass leads to indirect bilirubin overload on liver → physiologic jaundice.***

***Decreased synthesis of coagulation factors II, VII, IX X (liver immaturity) and transient vitamin K deficiency → hemorrhagic disease of the newborn***

### Central nervous system

- If the newborn blinks at light, the pupillary light reflex is present.
- Blinking response to a loud sound can assess hearing.
- Nystagmus on rotation.
- Response to pain by withdrawal and crying.
- ***Local reflexes:*** ***superficial reflexes*** (abdominal) and ***deep reflexes*** (triceps and patellar) are normally present. ***Planter response*** is usually extensor.

## Neonatal reflexes

- These are ***inborn behavioral patterns that develop during uterine life.***
- They should be ***fully present at birth*** and are ***gradually inhibited*** by higher centers in brain during the first 3 to 12 months of postnatal life.

### Moro reflex (Embrace, mass or startle reflex)


- **Eliciting:** Baby is held supine with shoulders and head supported by examiner's hand, then, allow ***head to fall back suddenly*** for 10-15° in the examiner's hand, ***sudden loud sound or sudden withdrawal of blanket*** from underneath the baby.
- **Response:** Baby extends trunk, extends & abducts arms and legs, then flexes and adducts arms and legs back (embracing).
- **Appearance:** Birth (28 wks gestation).
- **Disappearance:** This reflex lasts up to ***5-6 months***
- **Significance:** ***Absence:*** ***Generalized:*** Cerebral injury

  ***Localized:*** Nerve inj., joint affection, bone fracture or focal CNS lesion

  ***Bilateral absence:*** UMNL, CNS depression by narcotics or anesthesia, Bilateral injury of brachial plexus, Premature baby

  ***Asymmetric response:*** Erb's palsy , fracture clavicle or humerus

  ***Persistence after 6 mo:*** Cerebral palsy, MR

## Glabellar tap reflex

- **Eliciting:** ***Sharp tap on the Glabella***
- **Response:** Momentary tight ***closure of the eyes***
- **Appearance:** Birth (32-34 wks gestation)
- **Disappearance:** Variable
- **Significance:** ***Absence*** indicates ***CNS depression or prematurity***

## Pupillary reflex

- **Eliciting:** ***Close one eye and expose the other to light***
- **Response:** ***Pupil constricts***
- **Appearance:** Birth(29-37 wks gestation)
- **Significance:** ***Persists***

## Rooting reflex

Rooting reflex

- **Eliciting:** ***Corner*** of baby's mouth is stroked or touched
- **Response:** ***Turns*** his/her head & ***open*** his/her mouth to follow "root" in direction of stroking
- **Appearance:** Birth
- **Disappearance:** Disappears at ***4 mo awake*** and ***7 mo asleep***
- **Significance:** Helps baby find ***breast or bottle to begin feeding***
  ***Absence*** indicates ***CNS depression, prematurity or bulbar palsy***

## Suckling and swallowing reflex

Suck-swallow reflex

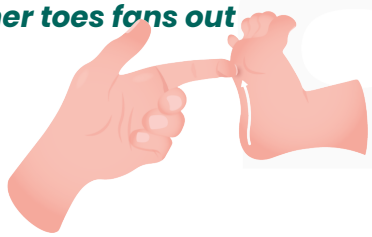
- **Eliciting:** ***Roof*** of baby's mouth is touched
- **Response:** Begins to ***suck***
- **Appearance:** Birth
- **Disappearance:** Disappears at ***4 mo awake*** and ***7 mo asleep***
- **Significance:** Premature babies may have weak or immature sucking

## Grasp reflex

Palmar grasp reflex

- **Eliciting:** ***Stroking palm*** of a baby's hand
- **Response:** ***close his/her fingers*** in a grasp
- **Lasts only 2 months**
- **Significance:** Stronger in ***premature babies***

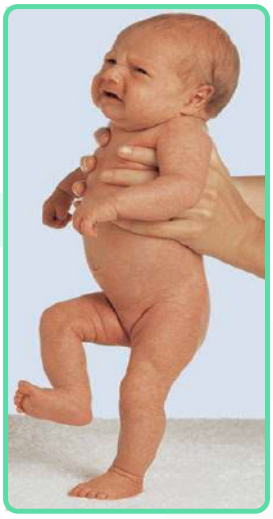
## Planter reflex (Babinski)



- **Eliciting:** ***Sole of foot*** is firmly stroked along lateral side
- **Response:** The ***big toe bends back toward top of foot and the other toes fans out***
- **Appearance:** Birth
- **Disappearance:** Up to ***2 years***
- **Significance:** A normal reflex up to about 2 years of age

### Placing reflex

- **Eliciting:** Baby is ***held upright***, dorsum of one foot touching under edge of a table
- **Response:** He flexes knee and hip to put the foot on table
- **Appearance:** Birth
- **Disappearance:** Disappears by ***6 weeks***
- **Significance:** ***Absent*** in ***CNS depression or LL paresis***



### Stepping reflex

- **Eliciting:** Baby is held in standing position, place his feet on a flat surface.
- **Response:** Stepping movement
- **Appearance:** Birth
- **Disappearance:** Disappears by ***6 weeks***
- **Significance:** ***Absent*** in ***CNS depression or LL paresis***

# Routine Care of the Neonate at Birth

## In the first hours of life (Delivery room care)

***The newly born baby is gently dried and wrapped in a warm, sterile towel.***

1. ***The time of birth is noted.***
2. ***Put the baby on a radiant warmer on his back*** with the neck in the neutral position by putting a towel beneath the baby's shoulders.
3. ***Rapidly assess the neonate's condition:***

| Proceed to routine care | Commence resuscitation |
|---|---|
| • Is breathing or crying spontaneously<br>• Has good muscle tone and responds when stimulated<br>• Has a heart rate above 100 beats/ minute<br>• Becomes pink rapidly | • Is not breathing spontaneously or has difficulty breathing<br>• Has poor muscle tone<br>• Has a heart rate less than or equal to 100 beats/minute<br>• Has persistent central cyanosis at 1 minute |

**Minimal Handling is a gold standard in neonatal manipulation.**

***APGAR score:*** A tool for monitoring adaptation to extra-uterine life.

In case of resuscitation, Apgar score is determined retrospectively.

If Apgar score is ≤ 4 at 1 minute or ≤ 6 at 5 minutes, should initiate necessary steps based on neonate's needs.

| Score | 0 | 1 | 2 |
|---|---|---|---|
| **Appearance (color)** | Blue/pale | Body pink, extremities blue | All pink |
| **Pulse (heart rate)** | Absent | < 100 | > 100 |
| **Grimaces** (response to catheter in nostril) | No response | Grimace | Cough or sneeze |
| **Attitude (tone)** | Limp | Some flexion of extremities | Active motion |
| **Respiratory effort** | Absent | Slow, irregular | Good crying |

***Apgar score at 1 minute:***

- A score of 8-10 is normal.
- A score of 4-7 means mildly to moderately depressed newborn.
- A score of 0-3 means severely depressed one.

***Apgar score at 5 minutes:*** indicates the probability of successfully resuscitating the infant.

- If the 5 minute Apgar score remains ***below or equal to 6*** extended Apgar score is done every 5 minutes i.e. at 10, 15 and 20 minutes.
- An Apgar core of 0-3 at 20 minutes reflects high risk of mortality and cerebral palsy in case of survival.

4. ***The upper airway is cleared by gentle suction of excess fluid and any inhaled vernix, blood or meconium:***

   Start by suctioning the mouth and pharynx since nasal stimulation by the suction catheter can induce a strong gasp with aspiration of oral and pharyngeal contents.

   Avoid too deep suctioning of pharynx since this will induce vagal stimulation → bradycardia.

5. ***If the baby starts to breathe regularly with his color changing to pink no further resuscitative steps are needed. If not look at step (7).***

6. ***Complete routine care by:***

   ***The mother should be encouraged to put her baby as early as possible on her breast*** so as to enhance maternal neonatal bonding by early skin-to-skin contact. Breastfeeding should be on demand, day and night

   ***The umbilical cord is clamped and cut***: ***Wait*** at least 1 - 3 min before clamping. ***Clamp*** with two forceps 10 cm from umbilicus and cut between. ***Tie off*** the cord (double ligature), leaving a 2 to 3 cm stump. ***Disinfect*** umbilicus: tip, stump and base of the cord.

   ***The baby is weighed*** and ***rapid clinical examination*** is done.

   ***The baby is dressed*** and put in the arms of his mother.

### *Thermoregulation*

- Dry with a clean, dry cloth; wrap in another
- Place against mother's (dried) body and cover with a dry cloth
- Perform a full clinical examination under an infant warmer.
- Cover head to reduce heat loss.
- Keep axillary temperature 36 - 37 °C, and pink, warm feet.
- Keep in a warm room (23 - 25 °C).
- Delay bathing until 24 hours after birth.

### *Assessment for risk factors for hypoglycemia*

- Birth weight < 2500 g or > 4000 g
- Maternal diabetes
- Difficulty breastfeeding

- ***If blood glucose is normal ( ≥ 45 mg/dl),*** observe that neonate is breastfed at least every 3 hours.
- ***Check blood glucose*** before each meal until there are 3 consecutive normal results.
- ***If blood glucose is < 45 mg/dl,*** hypoglycemia.

***Assessment for risk factors for neonatal infection:*** Prophylactic antibiotics for 48 hours

***Major risk factors:***

- Maternal fever (≥ 38 °C) before or during labor in preterm
- Prolonged rupture of membranes (PROM) ≥ 18 hours
- Foul-smelling, cloudy amniotic fluid
- Twin with clinical signs of infection

***Minor risk factors:***

- Preterm or birth weight < 2000 g
- Resuscitation at birth
- Meconium stained amniotic fluid
- Home delivery

## *Preventive treatments*

***Gonococcal conjunctivitis:*** Care of the eyes by application of antibiotic eye drops.

***Hemorrhagic disease of the newborn:***

- Administer ***vitamin K1*** IM within first few hours of life:
  - **Neonate weighing 1500 g or more:** 1 mg single dose
  - **Neonate weighing less than 1500 g:** 0.5 mg single dose.

***Rickets and vitamin D deficiency***

- Neonates particularly at risk and if possible all neonates should receive ***vitamin D for 6 months:*** Colecalciferol (vitamin D3) or Ergocalciferol (vitamin D2) PO:
  - Preterm or neonates high risk vitamin D deficiency: 600 to 1200 IU once daily
  - Term neonates: 400 to 800 IU once daily

***Neonatal vaccination:*** BCG, hepatitis B and polio vaccines

| | Dose/route of administration | Contra-indications |
|---|---|---|
| **Hepatitis B**<br>**Monovalent** | One dose = 5 to 10 ug<br>(follow manufacturer's instructions)<br>IM injection, anterolateral thigh | None but use only the monovalent vaccine |
| **Polio oral (Dose 0)**<br>**Bivalent (poliovirus types 1 and 3)** | One dose = 2 drops (approximately 0,1 ml)<br>Oral route | None |
| **BCG** | One dose = 0.05 ml<br>Intradermal injection, deltoid<br>(junction of lower 2/3 and upper 1/3 lateral aspect of upper arm) | Neonate whose mother has active tuberculosis* |

*Start the neonate on isoniazid preventive therapy, and administer the BCG vaccination when the isoniazid therapy is completed.

***7. If the baby is taking regular breaths but his color remains cyanotic*** he is in need for additional oxygen by facemask till color improves.

***8. If the baby fails in taking regular breaths or remains apneic*** do:

- Gentle stimulation by rubbing the back or slapping the soles of the feet.
- In many cases blowing oxygen on the nose will initiate breathing.

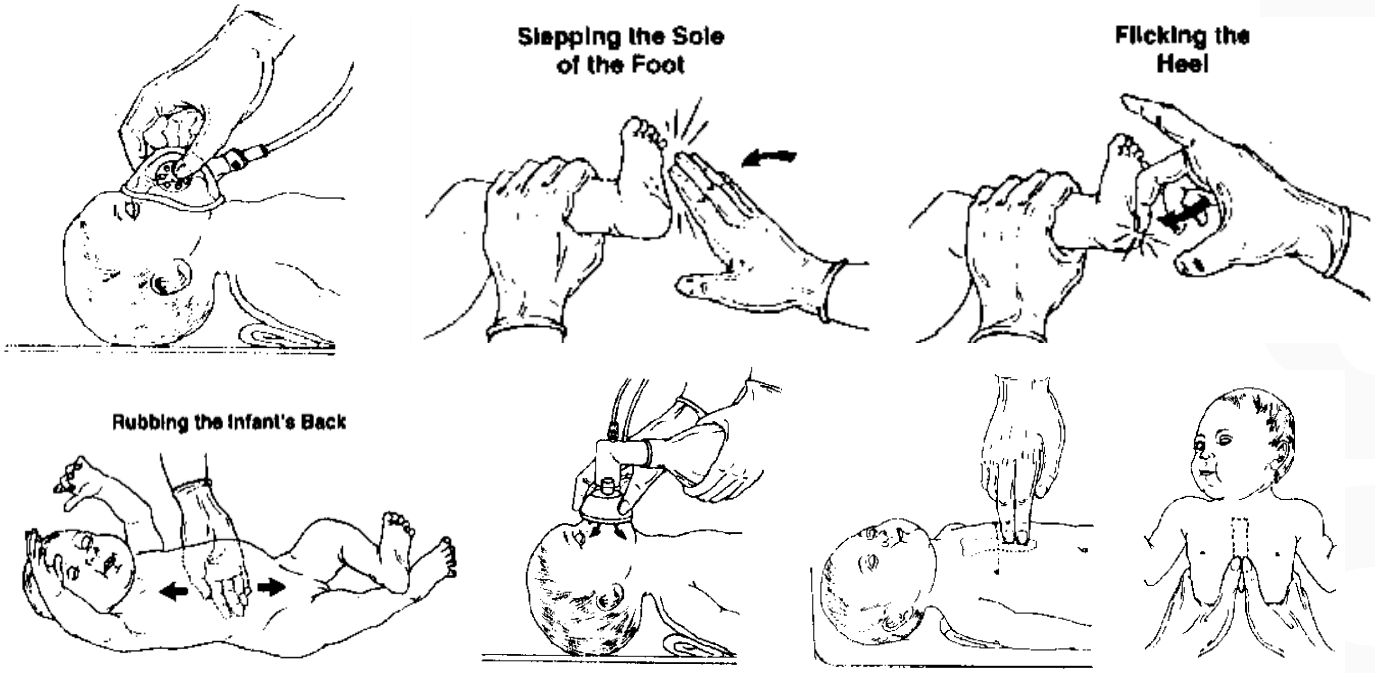
## *If all of these steps fail the next step is to:*

### *i- Expand and ventilate the lungs*

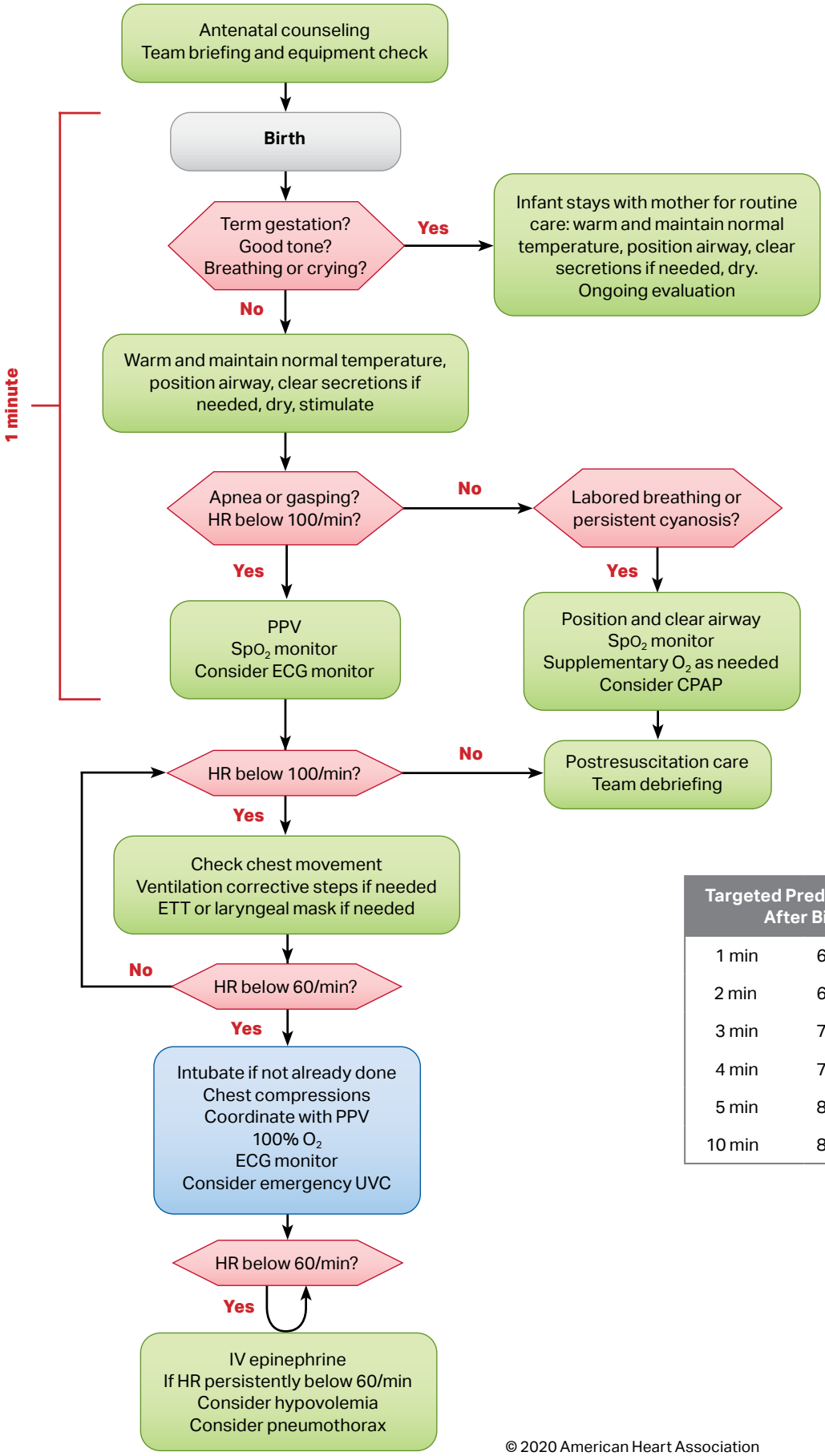
- With either bag and mask or bag and endotracheal tube.
- Often inflation of the lungs itself will initiate a gasp with spontaneous breathing.
- If it does not, the lungs should be ventilated at a rate between 30 and 50 inflations per minute with pressures below 30 cm H2O.
- The majority will quickly become pink and begin breathing within 2 to 5 minutes.
- ***Positive pressure ventilation may be appropriate in these circumstances to increase heart rate:***
  - if apnea and gasping is occurring
  - if the heart rate is below 100 beats per minute
  - if there is persistent cyanosis

**Positive pressure ventilation can be achieved with different types of bag-mask devices, which have different relative advantages and disadvantages.**

***ii- If the infant fails to become pink with good expansion of the lung*** then he is probably in shock. ***Cardiac massage*** using two-finger pressure on the lower half of the sternum at a rate of 140 taps per minute is occasionally necessary.



## Neonatal Resucitation Program Algorithm




| Targeted Preductal $SpO_2$ After Birth | |
|---|---|
| 1 min | 60%-65% |
| 2 min | 65%-70% |
| 3 min | 70%-75% |
| 4 min | 75%-80% |
| 5 min | 80%-85% |
| 10 min | 85%-95% |

# After the first hours of life

***Monitor:***

- Danger signs
- Temperature, heart and respiratory rate 2 times daily
- Weight once daily
- Urine and stool

***Keep cord clean, dry and exposed to the air (no dressing).***

***Observe breastfeeding.***

## Criteria for discharge from maternity hospital

- Appropriate management of neonatal infection
- Healthy neonate: good breastfeeding on demand, normal respiration and temperature, etc
- Weight > 1500 g
- BCG, hepatitis B and polio (0) vaccines administered
- Clinical record filled out
- Postnatal visit appointment given

## Information for the mother

- Breastfeeding
- Care for the baby
- Danger signs requiring a consultation

# 2 TERM & WEIGHT DISORDERS

## Disorders of duration

### 1. The Preterm newborn (Prematurity)

**A baby born before completing 37 weeks of gestation, counting from the first day of the Last Menstrual Period (LMP).**

#### Incidence:

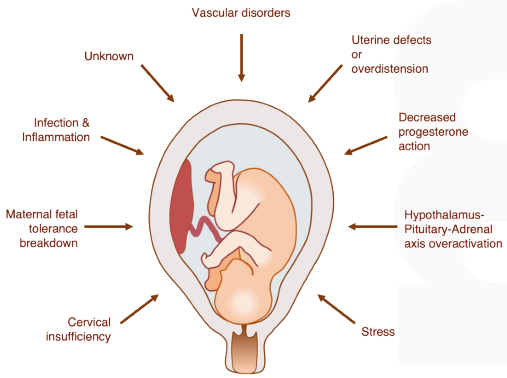
- Approximately 8% of live births are premature; almost 2% are less than 32 weeks gestation.

#### Etiology: (obstetrics)

***Unknown (idiopathic) in most cases.***

***Maternal:***

- Low socio-economic status.
- Mothers under the age of 16 or over 35 years.
- Maternal physical and/or mental stress.
- Acute or chronic maternal illness.
- Multiple-gestation births (in about half of all cases).
- Prior poor birth outcome (single strongest predictor).
- Inadvertent early delivery = incorrect estimation of gestational age.




***Obstetric factors:*** e.g. uterine trauma and malformations, placenta previa, placental abruption, cervical incompetence, and PROM.

***Fetal conditions:*** e.g. fetal distress, IUGR, and erythroblastosis fetalis.

#### Physical Characteristics

- ***Body measurements are decreased:*** Birth weight <2500 gm., length <47 cm. and head circumference <33 cm.
- Relatively large head and abdomen.
- Thin skin, little SC fat, abundant lanugo hairs, carpal and/or pedal edema.
- Small breast bud, soft ear pinna (almost without recoil).

- ***External genital organs:***
  - ***Males:*** smooth, poorly pigmented scrotum, un-descended testicles.
  - ***Females:*** separated labia majora, protruding labia minora.
- Generalized hypotonia, hypoactivity, and signs of neuro-muscular immaturity.

## Problems of prematurity

***Respiratory:*** Respiratory distress syndrome and apnea of prematurity.

***Temperature regulation:*** hypothermia.

***Neurologic:*** Intracranial hemorrhage and periventricular leukomalacia

***Cardiovascular:*** hypotension, patent ductus arteriosus (PDA).

***Gastrointestinal:*** Weak suckling and swallowing, increased demands, immature GIT, nutritional deficiencies, and necrotizing enterocolitis (NEC).

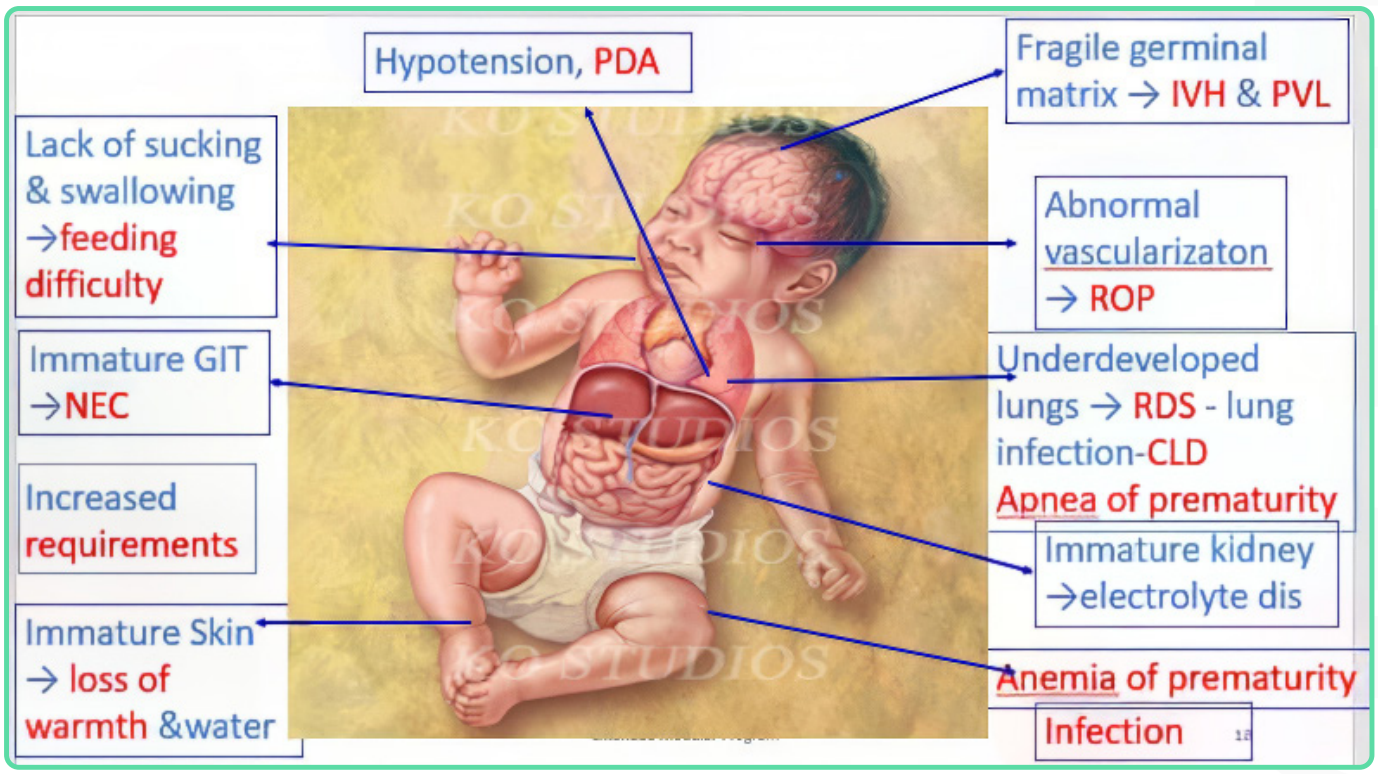
***Metabolic:*** decreased stores, hypoglycemia/hyperglycemia and hypocalcemia.

***Hematologic:*** Anemia of prematurity and poor clotting mechanism.

***Renal:*** Low GFR and renal tubular dysfunctions, electrolyte disturbances.

***Ophthalmologic:*** Retinopathy of prematurity (ROP).

***Immunologic:*** deficiency of both humoral and cellular immunity, high possibility of infection, rapid deterioration.



## 2. The Post-term newborn

**Defined as neonates born at or beyond 42 weeks gestation regardless birth weight**

### Etiology: (see obstetrics)

- In most cases cause is ***unknown***, ***inaccurate dating of pregnancy*** in some cases (inadvertent post term)

### Clinical characteristics:

- Dry cracked loose wrinkled skin with peeling, decreased subcutaneous fat, long nails, meconium staining of amniotic fluid then cord and nails.

### Morbidities: (Read Only)

- Meconium aspiration, persistent pulmonary hypertension, oligohydramnios, low Apgar score, birth injuries, hypocalcemia, hypoglycemia, fetal macrosomia, polycythemia, ± congenital anomalies.

### Management:

- Management of related morbidities

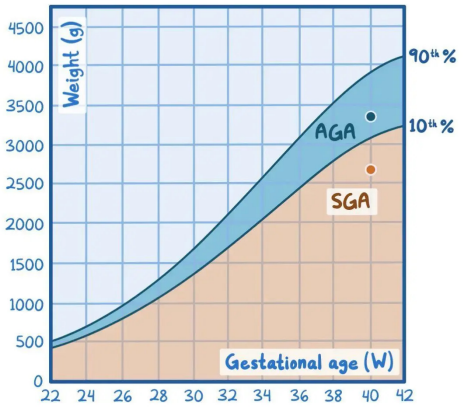
# Disorders of growth

## 1. The small for gestational age newborn (SGA)

**SGA describes neonate whose birth weight is <10th percentile for GA or <2 SD below the mean for the infant's GA.**

***Constitutionally Small babies***

***Intrauterine growth retardation (IUGR):*** diminished growth velocity in the fetus as documented by at least 2 intrauterine growth assessments due to pathological causes.




**Babies who are constitutionally small are at low risk to morbidities and mortality compared to those who are IUGR due to pathological causes**

### Etiology: (see OBSTETRICS)

### Physical characteristics:

- Wasted appearance, meconium staining, little subcutaneous fat
- IUGR neonates might be ***proportionate = symmetrical*** or ***disproportionate = asymmetrical*** (head sparing if growth restricted in later part of pregnancy)
- ***Alert, active and hungry***

### Problems and complications:

- • Polycythemia, thrombocytopenia, hyperbilirubinemia
- • Hypothermia, hypoglycemia (poor glycogen stores), hypocalcemia
- • Pulmonary hypertension, meconium aspiration, pulmonary hemorrhage
- • Congenital anomalies, infections, perinatal depression and necrotizing enterocolitis.
- • Dyslipidemia, delayed cognitive development, neurologic impairment (late complications)

### Management:

- ***Antenatal:*** delivery in an equipped hospital with NICU
- ***Anticipation and management*** of mentioned complications
- Special attention to ***temperature and blood glucose.***

## 2. The large for gestational age (LGA)

**Neonate with birth weight 2 SD above the mean for GA or above 90th percentile for his gestational age**

### Etiology:

- Constitutionally large (large parents)
- Infants of diabetic mothers (IDM)
- Post-term infant (some cases)
- Syndromes (Beckwith-Weidman)
- Hydrops fetalis

### Problems and complications:

- Birth trauma (Erb's palsy) and perinatal depression
- Hypoglycemia due to hyperinsulinism (IDM, Beckwith-Wiedmann, erythroblastosis fetalis)
- Higher morbidity and mortality

### Management:

- Delivery in an ***equipped hospital with NICU***
- ***Anticipation and management*** of the above complications according to the cause

# NEONATAL NEUROLOGY & BIRTH INJURIES

## Birth trauma

**Birth trauma is defined as injury to the infant resulting from mechanical forces (such as compression or traction) during the delivery.**

## Causes

### *Maternal causes*

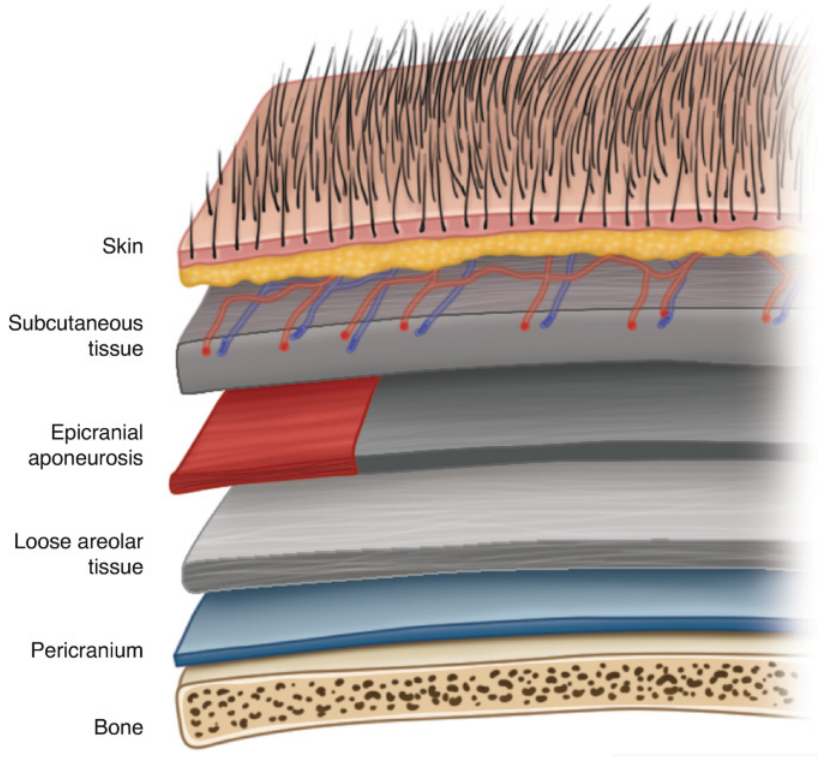
- Primipara
- Small maternal stature
- Maternal pelvic anomalies
- Oligohydroamnios

### *Fetal causes*

- Vacuum extraction
- Very low birth weight babies (VLBW)
- Fetal macrosomia
- Large fetal head
- Breech presentation
- Use of forceps

## Site/Types

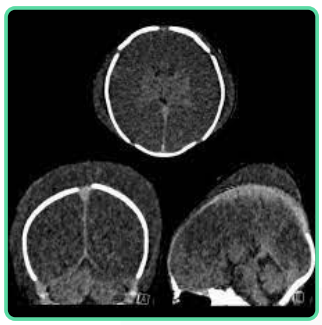
1. ***Head injuries***
2. ***Nerve injuries and spinal cord injuries***
3. Bone injuries
4. Intra-abdominal injuries
5. Soft tissue injuries



# A. Cranial Injuries

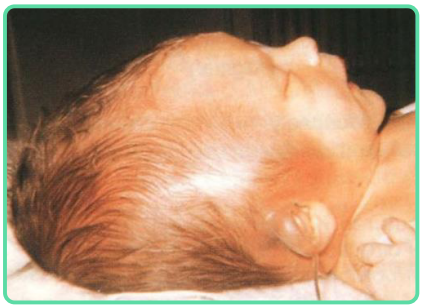
## 1. Caput succedaneum:

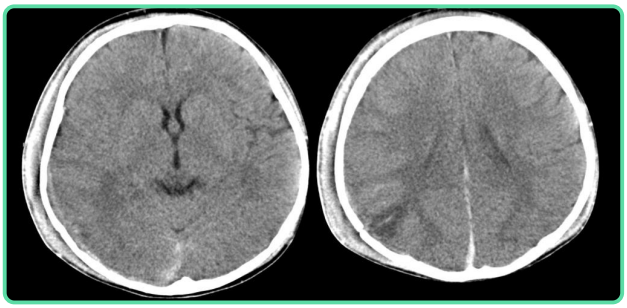
- ***A diffuse edematous swelling*** of the soft tissue of the scalp involving the presenting part of the fetus during delivery.
- It ***disappears within few days.***
- ***X- ray skull:*** shadow extracranial, crosses the sutures
- ***No ttt***



## 2. Subgaleal Hemorrhage

- Beneath the galea aponeurosis
- Continues after birth
- Serial head circumference measurements
- A boggy fluctuant mass, crosses the suture line and moves as the baby is repositioned
- Pallor, tachycardia
- CT or MRI

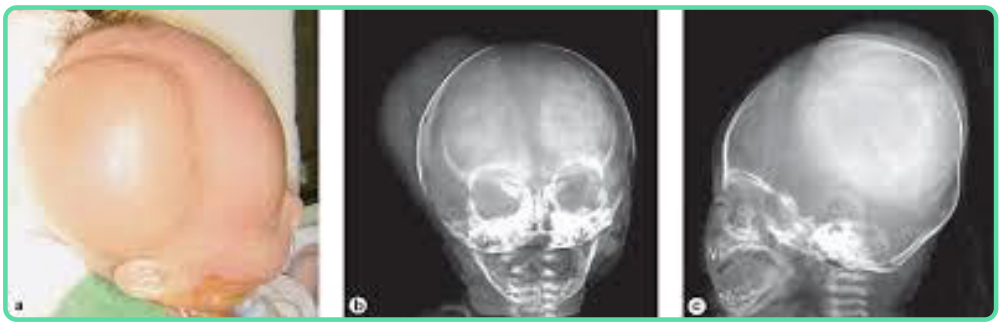




## 3. Cephalhematoma:

***It consists of subperiosteal hemorrhage (± fracture of skull), characterized by:***

- Limited to a surface of one cranial bone.
- May be unnoticed until several hours after birth.
- Gradually increases after birth…. Anemia, hyperbilirubinemia (if big)
- There may be an underlying skull fracture in 25%.
- A big cephalhematoma may cause hyperbilirubinemia, anemia or infection.
- ***X-ray*** swelling does not cross sutures
- ***ttt:*** conservative. ***DO NOT aspirate***



## 4. Intracranial hemorrhage (ICH):

### *Etiology*

- Bleeding tendency
- Prematurity
- Traumatic
- Hypoxic
- Congenital vascular malformation

### *Types*

- In the ***meninges*** (epidural, subdural, subarachnoid)
- In the ***parenchyma*** (e.g. intracerebral)
- In the ***ventricles*** (intraventricular)

### *Clinical manifestations*

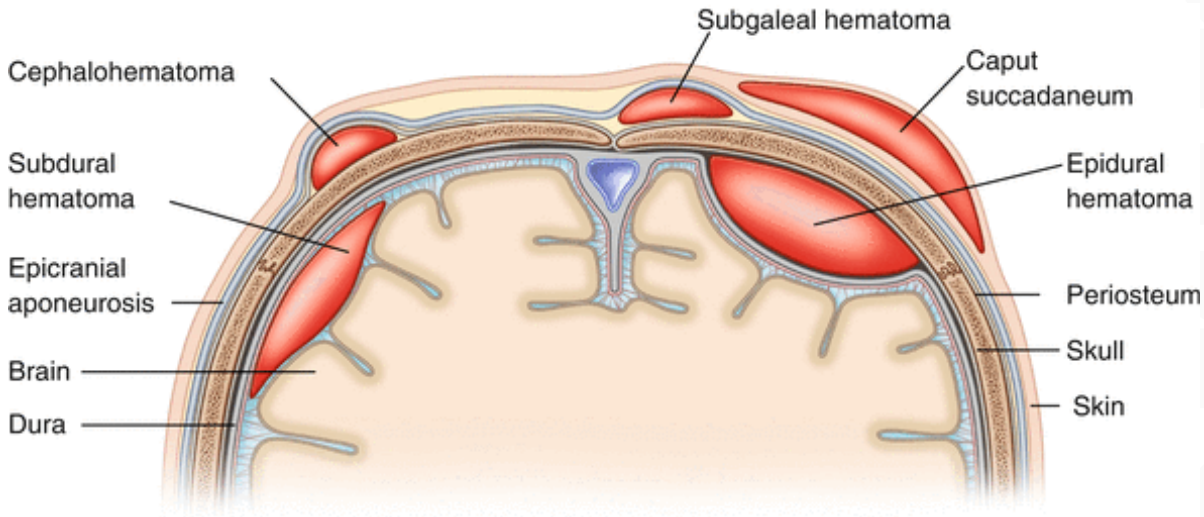
- It may manifest at birth or later on.
- ***The commonest symptoms are:*** lethargy, apnea, irritability, high pitched cry, bulging/ tense/plugging fontanelle, seizures and pallor.
- ***Other symptoms:*** of cause - Fussy, refusal of feeding - Coma - Cyanosis - Abnormal N reflexes - Unequal pupils - 6th nerve palsy

### *Investigations*

- ***Lab:*** CBC - PT/PTT
- ***Imaging:*** US - CT

### *Treatment*

- Neurosurgical consultation
- NICU admission, head up, minimal handling
- TPN/NG feeding
- ttt cause: Vit. K, FFP, PLT
- ttt manifestation & complications: anticonvulsant, BRBCs, dexamethasone, ventilation, AB



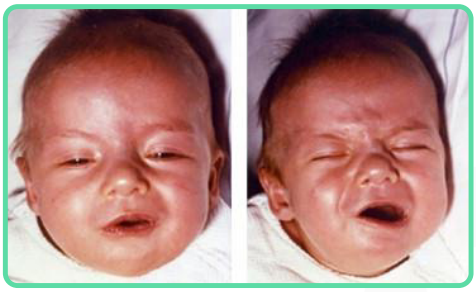
## B. Facial palsy / bell's palsy

**By direct pressure of the forceps blades or by haemorrhage & edema around the facial nerve.**

- Loss of movement of affected side.
- Affected eye side which remains open.
- Facial asymmtery.
- Absent of rooting reflex

### Treatment

- Aims at protecting the eye with antiseptic ointment, which remains open even during sleep.
- Neuroplasty (surgery to repair nerve tissue)
- The condition disappears within weeks.
- NG or OG feed.



DD: Hypoplasia of depressor anguli aris muscle

## C. Peripheral nerve injuries

### 1. Brachial palsy:

***a. Erb's paralysis:*** injury of the ***5th and 6th*** cervical roots.

- The affected upper limb is internally rotated at the shoulder, with extension of the elbow and pronation of forearm (policeman's tip position).
- The paralyzed limb is hypotonic with loss of ordinary movements, but the hand moves properly with asymmetrical Moro reflex.

***b. Klumpke's paralysis:*** injury of the ***7th and 8th*** cervical roots

- Leading to dropping of the shoulder and paralysis of the hand (Claw-hand).
- In some cases meiosis and ptosis occur if the sympathetic fibers are also injured (Horner's).

***c. Paralysis of the entire brachial plexus.***

***TTT: Position-Physiotherapy-Neuroplasty***

### 2. Phrenic nerve:

***All of the following + the manifestations of brachial palsy:***

- Injury cervical roots (3rd, 4th and 5th cervical nerves).
- Cyanosis and irregular labored respiration.
- Breath sounds are diminished on the affected side.
- Fluoroscopy: elevation of the diaphragm on the paralyzed side and sea-saw movements of the two sides of the diaphragm.

## D. Visceral injuries

***The important viscera which could be injured are:*** liver, spleen and adrenal glands.

***Examine for signs of blood loss*** (anemia, abdominal distension or shock).

# Neurological Problems of The Neonate

## Hypoxic-Ischemic Encephalopathy (HIE)

**Asphyxia is defined biochemically as having three components: hypoxemia, hypercapnia, and mixed acidosis (metabolic and respiratory).**

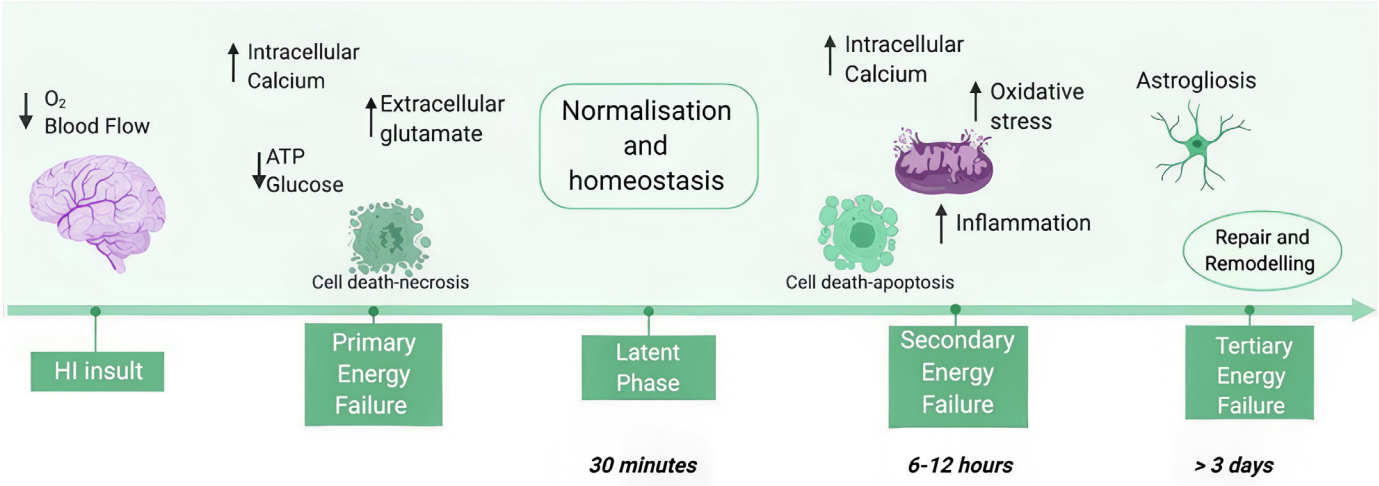
***A failure of function of at least two organs*** (may include lung, heart, liver, brain, kidneys and hematological) consistent with the effects of acute asphyxia.

### Pathophysiology of Hypoxic-Ischemic Encephalopathy:

***Perinatal asphyxia leads to primary and secondary events:***

***Primary neuronal damage:*** cytotoxic changes due to failure of microcirculation → inhibition of energy-production → cytotoxic edema and free radical formation → neuronal damage (Immediate neuronal necrosis).

***Secondary neuronal damage:*** May extend up to 72 hours or more after the acute insult and results in an inflammatory response and cell necrosis or apoptosis (reperfusion injury) (delayed neuronal apoptosis).




### Risk factors (see OBSTETRICS)

***The incidence of perinatal asphyxia is higher in complicated pregnancies:***

1. Pre-eclampsia, hypertension, severe maternal pulmonary or cardiac disease
2. Intrauterine growth restriction
3. Placental abruption, infarction, cord around neck, cord knot.
4. Fetal anemia, infection, hydrops, severe heart failure
5. Malpresentation, obstructed labour

***Antenatal:*** PE/ hypotension, placental causes, cord compression, post maturity.

***Natal:*** Prolonged/obstructed labor (malposition), induced labor.

***Postnatal:*** Shock, anemia, CCHD, Respiratory.

## Diagnosis of HIE

### *CP (criteria A&B):*

- ***Acute perinatal event*** (Preeclampsia, placental abruption, cord around the neck) ***(Causes of perinatal asphyxia)***
- ***Persistence of an Apgar score of 0-3 for longer than 5 minutes*** (≥10 minutes)
- ***Neonatal neurologic sequelae*** (eg, seizures, abnormal consciousness, hypotonia)
- ***Multiple organ involvements*** (kidneys, liver, heart, intestine)
- ***Meconium***

### *Investigations:*

- ***Metabolic derangement:*** ***Profound metabolic or mixed acidemia*** pH≤7.0 or a base deficit of ≥16 within first hour, ***hypoglycemia***, ***hyper $K^+$)***
- ***ABG***
- ***LFT & KFT***
- ***ECHO***
- ***EEG***
- ***Brain Imaging***

## Neurological examination (grading of HIE) (Sarnat and Sarnat, 1976):

***Grade 1:*** Mild encephalopathy with the infant hyperalert, irritable, and over-reactive to stimulation. (sympathetic over-stimulation with tachycardia, dilated pupils and jitteriness. The EEG is normal.

***Grade 2:*** Moderate encephalopathy with the infant displaying lethargy, hypotonia and proximal weakness. There is parasympathetic overstimulation with low resting heart rate, small pupils, and copious secretions. The EEG is abnormal and 70% of infants have seizures.

***Grade 3:*** Severe encephalopathy with a stuporous, flaccid infant, and absent reflexes. The infant may have seizures and has an abnormal EEG.

## Management

- Monitoring
- CUS/ECHO
- aEEG
- Labs
- Resuscitation (cord ABG, APGAR)
- Hypothermia
- Adjuvant lines (Stabilization, theraputic)
- New lines (stem cell)

**Hypothermia in HIE:**

Selective head or whole body hypothermia of a core temperature of 33.5 C° applied within 6 hours of birth for 48-72 hours is neuroprotective.

***Possible mechanisms include:***

1. Reduced metabolic rate and energy depletion.
2. Decreased excitatory transmitter release.
3. Reduced alterations in ion flux.
4. Reduced apoptosis due to HEI.
5. Reduced vascular permeability, edema, and disruptions of blood-brain barrier functions.

# 4 METABOLIC NEONATAL DISORDERS

## A-Neonatal Jaundice

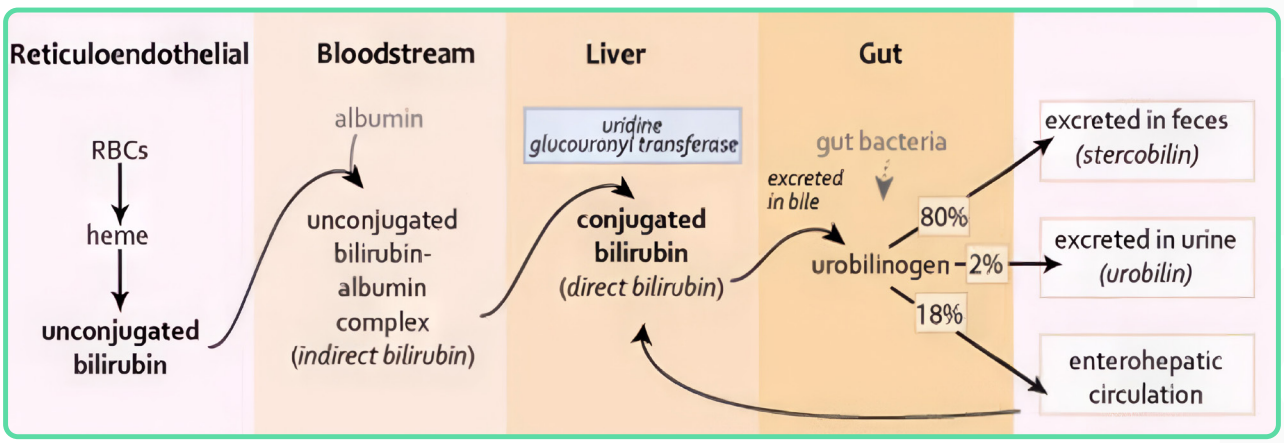
**Yellowish discoloration of skin and mucous membrane due to increased serum bilirubin above 7 mg% (normal serum bilirubin is 0.1 -0.8 mg %).**

- Jaundice is the visible manifestation of chemical bilirubinemia .
- In adults sclera appears jaundiced when serum bilirubin exceeds 2 mg/ dl.
- Icterus, however, becomes apparent on the skin when serum bilirubin reaches more than 5 mg/ dl.
- Almost all neonates (60% Term and 80% Preterm) will have bilirubin greater than 5 mg/ dl in the first week of life and about 6% of term babies will have levels exceeding 15 mg/dl.

## Metabolism of bilirubin

### Source of production:

- Bilirubin is derived from the breakdown of heme proteins which are present in hemoglobin, myoglobin and certain heme containing enzymes.
- Three fourths of the bilirubin comes from hemoglobin catabolism.
- One gram of hemoglobin results in the production of 34 mg of bilirubin.
- A normal term newborn produces about 6 - 10 mg/kg/ day of bilirubin.



## Steps from bilirubin production to excretion

### *(1) Reticuloendothelial system (RES)*

- Macrophages phagocytose sensescent erythrocytes
- Hemoglobin metabolism yields bilirubin
- Pathway: heme → biliverdin (green colored) → bilirubin (yellow colored)

### *(2) Bloodstream*

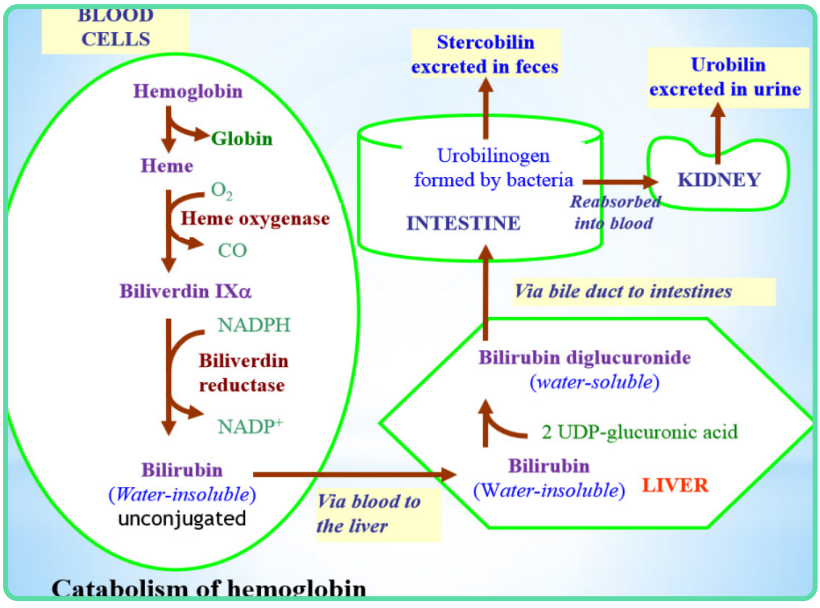
- Albumin binds bilirubin and complex is carried to liver
- Bilirubin albumin complex = ***indirect bilirubin (water insoluble)***

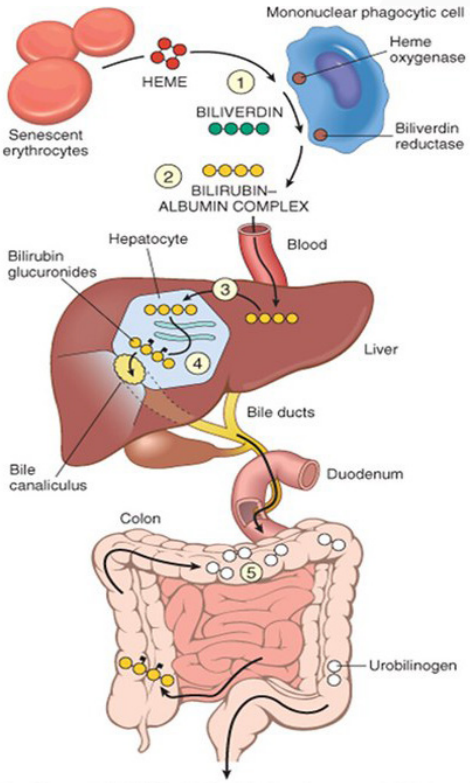
### *(3) Liver*

- Hepatocytes take up bilirubin
- Hepatic microsomes conjugate bilirubin with ***glucoronic acid***
  - Conjugation via ***UDP glucuronyl transferase;*** enzyme is synthesized slowly after birth, sometimes causing newborn jaundice
- ***Conjugated bilirubin = direct bilirubin aka water soluble***
- A portion of conjugated bilirubin is excreted in urine
- Remainder is secreted into bile and then into small intestine

### *(4) Gastointestinal tract*

- In terminal ileum and colon, bilirubin is deconjugated by bacterial enzymes and metabolized to urobilinogen
  - ***18% Of urobilinogen*** is absorbed via enterohepatic circulation and delivered back to liver
  - ***80% Of urobilinogen*** is converted to stercobilin and excreted in feces (stercobilin gives characteristic color of feces)
  - ***2% Of urobilinogen*** is converted to urobilin and excreted in urine (urobilin gives characteristic color of urine)



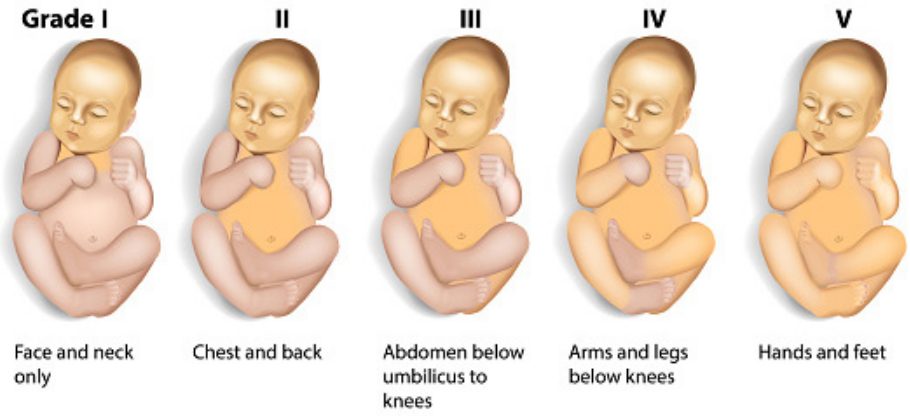



# Assessment of jaundice

## Clinical criteria:

- It is very widely used and utilizes the principle that clinical jaundice first becomes obvious in the face followed by a ***downward progression as it increases in intensity.***
- Assessment of jaundice should be done in natural light. The finger is pressed on the baby's skin, preferably over a bony part, till it blanches.
- The underlying skin is noted for yellow color. Extent of jaundice thus detected gives a rough estimate of serum bilirubin .
- Clinical estimation of bilirubin by experienced person, though reliable, has to be confirmed by laboratory methods.

| Area of body | Range of bilirubin (mg/100 ml) |
|---|---|
| Face | 4-8 |
| Upper trunk | 5-12 |
| Lower trunk & thighs | 8-16 |
| Arms & lower legs | 11-18 |
| Palms & soles | >15 |




# Differential Diagnosis of Neonatal Jaundice

**Hyperbilirubinemia in the first week of life is usually of the indirect variety.**

**Causes are usually classified based on the time of onset of jaundice. While referring a baby with jaundice make sure that either the mother is referred or mother's blood sample is sent.**

### *A. Jaundice appearing on the first day of life:*

1. STORCH-EB infection.
2. Hemolytic disease of the newborn.

### *B. Jaundice appearing on 2nd–3rd day:*

1. Physiological jaundice.
2. Criggler Najjar syndrome.
3. STORCH-EB infection.

### *C. Jaundice after 3rd day to 7th day of life:*

1. Absorption of hematoma (cephalhematoma).
2. Hemolytic anemias as: spherocytosis, G6PD.
3. STORCH-EB infection.
4. Neonatal sepsis, pneumonia, urinary tract infection.

### *D. Jaundice appearing after the 1st week of life:*

1. Hemolytic anemias as spherocytosis.
2. Breast milk jaundice.
3. Synthetic vitamin K. injection.

### *E. Persistent jaundice after the 1st month of life:*

1. Hypothyroidism (unconjugated).
2. Intestinal obstruction.
3. Biliary atresia.
4. Neonatal hepatitis.
5. Inspissated bile syndrome.
6. Choledocal cyst.
7. Metabolic as galactosemia.

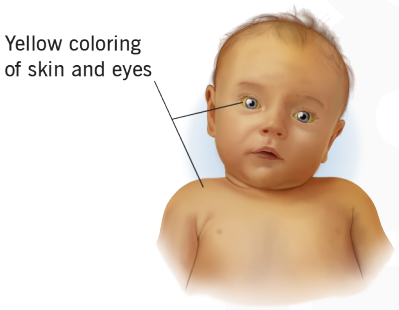
**Remember that**

- The age of appearance may overlap and the above mentioned grouping is only a general classification.
- Infection must be ruled out in jaundice appearing any time after third day of life.
- Even after extensive investigations, cause remains uncertain in over one third of cases.
- Neonatal jaundice may be multifactorial in origin

# I. Physiological Jaundice

**It is the commonest cause of neonatal jaundice.**

- First appears between 24-72 hours of age
- Maximum intensity seen on 4-5th day in term and 7th day in preterm neonates
- Does not exceed 15 mg/ dL
- Clinically undetectable after 14 days.




***No treatment is required but baby should be observed closely for signs of worsening jaundice.***

## Causes:

***Immaturity in bilirubin metabolism at multiple steps results in the occurrence of hyperbilirubinemia in the first few days of life. These are:***

- Increased bilirubin load on the hepatic cell
- Defective uptake from plasma into liver cell
- Defective conjugation
- Decreased excretion
- Increased entero-hepatic circulation

**It is indirect hyperbilirubinemia, all tests are normal and no treatment is needed.**

# II. Pathological Jaundice

***Presence of any of the following signs denotes that the jaundice is pathological:***

- Clinical jaundice detected before 24 hours of age
- Rise in serum bilirubin by more than 5 mg/ dl/ day
- Serum bilirubin more than 15 mg/ dl
- Clinical jaundice persisting beyond 14 days of life
- Clay/white colored stool and/or dark urine staining the clothes yellow
- Direct bilirubin >2 mg/ dl at any time

***One should investigate to find the cause*** of pathological jaundice.

***Treatment is required*** in the form of ***phototherapy or exchange blood transfusion.***

## A. Hemolytic Disease of the Newborn

**If an Rh +ve baby is born to an Rh -ve mother, she may get sensitized during labor, or abortion by passage of small amounts of Rh +ve blood, or by transfusion of Rh +ve blood to Rh -ve lady.**

***Rh immunization is less frequent because:***

a. Immunization occurs late in pregnancy so the 1st baby is usually not affected.

b. The father may be Dd so the child will be dd or Dd.

c. Some mothers are weak antibody producers.

d. Associated ABO incompatibility (will destroy RBCs before sensitization occurs).

***Clinical Picture***

1. ***Hydrops fetalis:*** Born still birth or die after labor: Pallor, huge hepatosplenomegaly, generalized edema and anasarca.
2. ***Icterus gravis neonatorum:*** Jaundice appears in 1st day of life, marked anaemia, hepatosplenomegaly, inspissated bile syndrome.
3. ***Rh. hemolytic anemia:*** Anemia with a peak in 3rd week of life, mild jaundice, positive Coombs' test.

***Investigations***

1. ***In Rh -ve mother,*** do antibody titre anti-D, if higher than 1/64 proceed to amniocentesis, but if 1/64 or below follow-up is indicated every 2 weeks.
2. ***Spectrophotometric study of amniotic fluid*** is done and blotted against Lilly chart, a result in high zone II or zone III needs urgent intervention.
   - ***If pregnancy is 34 weeks or more*** à induction of labor and management of hyperbilirubinemia and prematurity is done.
   - ***If pregnancy is less than 34 weeks***, intrauterine intra-umbilical cord blood transfusion with O -ve blood compatible with the mother's serum.
3. ***After labor:*** cord bilirubin, hemoglobin, Coombs' test and reticulocytic count (which must be high in hemolysis) are done to assess the severity of the condition.

## Treatment

### I. Exchange transfusion

#### Indications:

1. Cord bilirubin of 5 mg% or more.
2. Rise of serum bilirubin of more than 1 mg/hour.
3. Cord Hb of 10 gm% or less.
4. History of previous neonatal death or previous kernicterus
5. Any sign of kernicterus.
6. If serum indirect bilirubin reaches level of exchange according to curves.

**Type of blood:** Group O Rh -ve fresh blood compatible with the baby's and maternal serum of citrate phosphate dextrose anticoagulation system.

**Volume:** Double volume = 85 x 2 x wt (Kg).

**Time:** One hour.

**Technique:** Insert an umbilical vein catheter, remove about 20 cc of infant's blood and replace by 20 cc of the prepared blood. Give I.V. Ca, glucose during the procedure and monitor HR, RR of the baby.

#### Complications of exchange transfusion:

##### Acute complications:

- Transient bradycardia with or without calcium infusion.
- Cyanosis.
- Transient vasospasm.
- Thrombosis.
- Apnea with bradycardia.
- Infections: CMV, HIV, and hepatitis.
- Necrotizing enterocolitis.

##### Late complications:

- Anemia and cholestasis.
- Late anemia.
- Graft versus host reaction.
- Portal vein thrombosis.

**Indications of exchange transfusion in neonates**

Exchange transfusion is indicated for severe indirect (unconjugated) hyperbilirubinemia to avoid bilirubin neurotoxicity when other therapeutic modalities have failed or are not sufficient.

The procedure may also be indicated in infants with erythroblastosis fetalis (HDN) presenting with severe anemia, hydrops, or both, even in the absence of high serum bilirubin levels.

#### Other indications for exchange transfusion:

- Neonatal septicaemia
- Neonatal polycythemia
- Severe anemic heart failure
- RH disease
- DIC
- Drugs as toxic doses of phenobarbitone
- Metabolic as maple syrup urine disease
- Polycythemia

## Extras for exchange transfusion

***Volume:***

- Blood Volume = 70 90 ml/kg for term and 85 110 ml/kg for preterm infants
- One blood volume removes 65% of baby's red cells.
- Two blood volumes removes 88%
- Thereafter the gain is small
- Meticulous care must be taken with volume balance, the rate of the exchange, the vital signs and any signs of air in the lines.
- During the exchange ensure volume in/volume out balance does not exceed
  - 5ml < 1000g baby
  - 10ml > 1000g baby
  - 15ml > 2000g baby

**It is still the most effective and reliable method to reduce serum bilirubin.**

**Anticipation and early referral to a higher center is indicated.**

***Choice of blood for exchange blood transfusion***

- In ABO incompatibility: Use O cells of same Rh type, ideal is to have O cells suspended in AB plasma.
- In Rh isoimmunization: In emergency use O -ve blood. Ideal is O -ve cells suspended in AB plasma. One may use baby blood group but Rh -ve blood also.
- Other conditions: Baby's blood group.

***Complications***

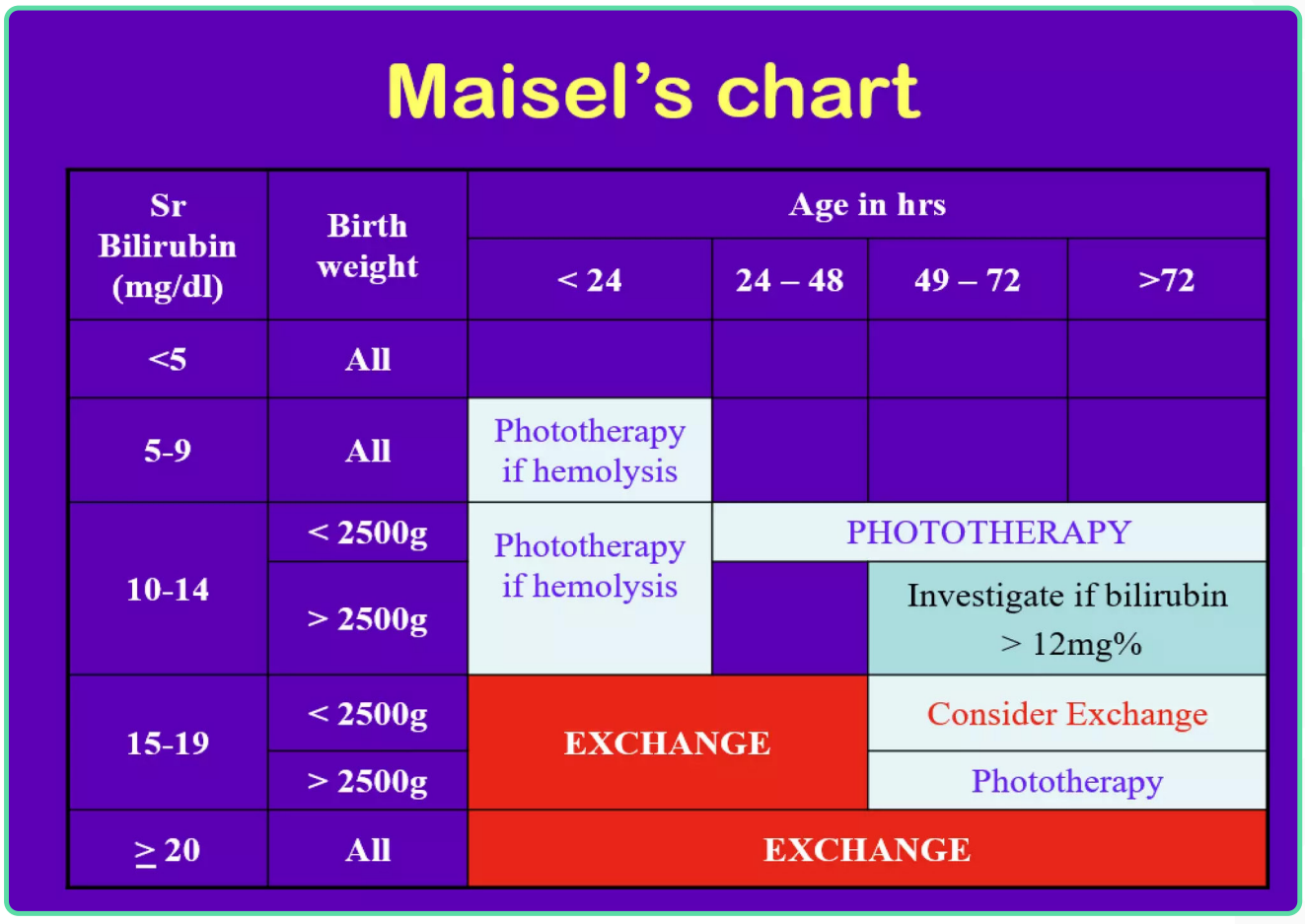
- **Air embolus:** Ensure the lines are correctly set up. - Watch the lines continuously for air. - Turn the line off instantly if air is seen. - Never have a 3 way tap open to air and the baby - Be very careful if there are large swings in intrathoracic pressure.
- **Volume imbalance:** The nurse is responsible for recording the volume balance throughout the exchange.
- **Arrhythmias** Can occur from a variety of causes. - Set the monitor to have an audible QRS complex.
- **Acidosis:** Blood for exchange transfusion is preserved in CPD (citrate, phosphate, dextrose) and can be quite acidotic. - Check the baby's blood pH before, during ( usually half way), and after the exchange - Check more frequently for a sick, unstable or small baby.
- **Respiratory distress:** Monitor respiration and SpO2 constantly.
- **Hyperkalaemia:** CPD blood can have high potassium [K+] levels. - Check [K+] at the start of each bag. - Monitor the QRS complex. (for arrhythmia, widening QRS) - Monitor K+ with each blood gas.
- **Hypernatraemia:** CPD blood has a high [Na+]. - Monitor [Na+] with each gas.
- ***Hypocalcaemia:*** Monitor calcium [Ca++] and give replacement - Ca++ in IV fluids as per clinical guidelines
- ***Hypoglycaemia:*** Unlikely during the exchange as CPD blood has 19mmol/L [glucose]. - However rebound hypoglycaemia may occur afterwards. - Commence a 10% glucose infusion post exchange, or if the exchange is interrupted.

- **Thrombocytopenia:** Very common, and more severe after more exchanges (due to increased platelet consumption). - Recovers in a few days. - Monitor platelets serially for a week post exchange.
- **Coagulopathy or neutropenia:** More likely the multiple transfusions
- **Anaemia/Polycythaemia:** From poorly mixed or packed blood. - Check the PCV of each blood bag. - Agitate the bag every 15 minutes.
- **Infection:** Prophylactic antibiotics are not indicated. - Observe closely for signs of infection.

## Maisel's chart

**It is used for taking decision regarding treatment in cases of pathological jaundice. In presence of any of the following, treat as in next higher bilirubin category.**

- Perinatal asphyxia
- Respiratory distress
- Metabolic acidosis
- Hypothermia
- Low serum protein
- Birth weight <1500 g
- Signs of clinical or CNS deterioration



### II. Phototherapy

Exposure of the newborn to blue light (450 - 460 nm) à formation of an isomer of indirect bilirubin which becomes water soluble and pass with the urine.

It reduces serum bilirubin about 1-3 mg in 8-12 hours of exposure.

***Fibreoptic phototherapy***

- These devices use a standard light source, usually a quartz halogen bulb. The light from the bulb may then be passed through a filter before being channelled down a fibreoptic bundle into a pad of woven optic fibres.
- ***In conventional fibreoptic phototherapy the light source could be:*** Fluorescent lamps - Quartz halogen lamps - Gas discharge tubes - Light emitting diodes (LEDs).

***Complications:***

1. Increased insensible water loss (dehydration): Provide more frequent and for longer duration extra breast feeding.
2. Loose green stools: weigh often and compensate with breast milk.
3. Skin rashes: Harmless, no need to discontinue phototherapy;
4. Bronze baby syndrome: occurs if baby has conjugated hyperbilirubinemia. If so, discontinue phototherapy;
5. Hypo or hyperthermia: monitor temperature frequently.
6. Harm to the eyes and genitalia.

### III. Phenobarbitone:

- Enzyme inducer needs 3-7 days to act (level of glucuronyl transferase).
- Long latent period and has sedative effect.(not used now)

### IV. Packed RBCs transfusion in anemia

### V. Intravenous immunoglobulin:

- In recent years, IVIG has been used for numerous immunologically mediated conditions.
- In the presence of Rh, ABO, or other blood group incompatibilities that cause significant neonatal jaundice, IVIG significantly reduce the need for exchange transfusions.
- Dose range from 500-1000 mg/kg. (only in immune hemolysis with positive coombs test)

## B. ABO Incompatibility

***A milder form of hemolytic disease of newborn with the following characteristics:***

- Mild anemia, and jaundice
- Mother is O + baby is A or B.
- Occurs in 25% of all newborn
- The 1st baby is affected.
- Coomb's test usually negative
- Spherocytosis + anaemia + high retics
- Usually need phototherapy only.

## C. Crigler-Najjar Syndrome

***Familial cause of indirect hyperbilirubinemia (glucuronyl transferase 1A1 deficiency).***

***Type I:***

- Severe form of glucuronyl transferase deficiency with autosomal recessive inheritance.
- No enzyme is detected.
- The baby is deeply jaundiced and ends by kernicterus in spite of treatment.

***Type II:***

- Milder form, autosomal dominant (AD).
- Enzyme activity is 20-30% of normal.
- Responds well to phenobarbitone.
- Phototherapy (exchange may be needed).

## D. Gilbert's Syndrome

- Congenitally deficient enzymes and acceptors (Y, Z).
- Jaundice is mild (Bilirubin: 1-3 mg %).
- Provoked by low caloric intake.

## E. Rotor & Dubin Johnson Syndromes

- Cause direct hyperbilirubinemia due to defective excretion of bilirubin to the bile duct system.

## F. Breast Milk Jaundice

- Due to 3a - 20b pregnanediol which passes with the milk in breast fed babies à inhibition of glucuronyl tansferase enzyme.
  - This condition may persist as a prolonged physiological jaundice or it may appear for the first time after the first week.
  - It is common in solely breast fed babies and the intensity is maximum between 10 14 days of life.
  - The bilirubin levels are never high enough to require exchange though phototherapy may occasionally be necessary.
  - If bilirubin is less than 15 mg/ dl at 3 weeks one need not worry. But if bilirubin is > 15 mg/dl at 3 weeks, cessation of breast milk for 48 hours will decrease bilirubin levels dramatically and breast milk can be resumed without any risk of recurrence of jaundice.
  - However, more frequent breast feeds without cessation results in improvement in many.
- ***Occurs in 2nd week of life.***
- ***Unconjugated (indirect bilirubin), No kernicterus*** is reported.
- ***Treatment:*** stop breast feeding for 48 hours at least, and then start again.

  Note: according to lecture, do NOT stop BF and continue as usual.

## G. Congenital Biliary Atresia (refer to hepatology)

## H. Neonatal Hepatitis and Inspissated Bile Syndrome (refer to hepatology)

**Prolonged indirect jaundice**

***The following conditions may lead to prolonged indirect hyperbilirubinemia:***

- Crigler Najjar Syndrome
- Breast milk jaundice
- Hypothyroidism
- Pyloric stenosis
- Ongoing hemolysis, malaria

## Risk factors of jaundice

***A simple pneumonic for risk factors is JAUNDICE:***

- ***J***aundice within first 24 hrs of life
- ***A*** sibling who was jaundiced as neonate
- ***U***nrecognized hemolysis
- ***N***on optimal sucking/nursing
- ***D***eficiency of G6PD
- ***I***nfection
- ***C***ephalhematoma /bruising
- ***E***ast Asian/North Indian

## Approach to a jaundiced baby

***The following four questions need to be answered***

- What is the birth weight?
- What is the gestation?
- What is the postnatal age in hours?
- Is the jaundice physiological or pathological?

**If the jaundice is physiological and baby is well only observation is necessary.**

**In deeply jaundiced newborn one must also evaluate for presence or absence of bilirubin toxicity ( kernicterus ).**

**Kernicterus is identified by lethargy and poor feeding, poor or absent Moro's reflex, opisthotonus or convulsions.**

# Workup for pathological jaundice

## 1. History

- Review maternal and perinatal history
- Family history of jaundice, liver disease
- Previous sibling with jaundice for blood group incompatibility
- Maternal illness during pregnancy
- Previous history of malaria
- Traumatic delivery, delayed cord clamping, oxytocin use
- Birth asphyxia, delayed feeding, delay in meconium passage
- Breast feeding

## 2. Physical examination

- Prematurity
- Small for gestation: polycythemia, hepato splenomegaly, cataract, rash.
- Extravascular bleed: cephalhematoma
- Pallor: hemolysis, blood loss
- Petechiae: sepsis, TORCH infections
- Hepatosplenomegaly: Rh isoimmunization, sepsis, TORCH infections

## 3. Laboratory tests (must in all*)

- Serum bilirubin total and direct*
- Blood group and Rh for mother and baby*
- Direct Coomb's test on infant
- Hematocrit*
- Peripheral smear for RBC morphology, evidence of hemolysis and, reticulocyte count
- Sepsis screen
- Liver and thyroid function tests in cases with prolonged jaundice
- TORCH titres

### Indications for referral to hospital

- Onset of jaundice within 24 hours of life
- Rapidly rising TSB of greater than 6 mg/dL/day (103 µmol /L/day)
- Clinical jaundice below umbilicus (if till the soles of the feet urgent referral for possibility of ET)
- G6PD deficiency (if not previously hospitalised)
- Clinical symptoms/signs suggestive of sepsis.

# Causes of conjugated Hyperbilirubinemia

**This is uncommon during the neonatal period.**

**It is defined as a direct serum bilirubin level of > 2 mg/dL.**

**It is important to document the cause as it is never physiological.**

- ***Idiopathic*** neonatal hepatitis
- ***Infections:*** Hepatitis B, TORCH, Sepsis
- ***Malformations:*** Biliary atresia, choledochal cyst, bile duct stenosis.
- ***Metabolic disorders:*** Galactosemia, Hereditary Fructose intolerance Alpha-l antitrypsin deficiency, Tyrosinemia, Glycogen storage disease type IV
- ***Total parenteral nutrition***

## Common

- Hyperalimentation cholestasis
- CMV infection
- Other perinatal congenital inf. (TORCH)
- Inspissated bile from prolonged hemolysis
- Neonatal hepatitis
- Sepsis

## Uncommon

- Hepatic infarction
- Cystic fibrosis
- Biliary atresia
- Choledochal cyst
- $\alpha_1$-Antitrypsin deficiency
- Neonatal iron storage disease
- Alagille syndrome (arteriohepatic dysplasia)
- Inborn errors of metabolism (galactosemia, tyrosinemia)
- Byler disease

# Kernicterus

**It is bilirubin encephalopathy, occurs at high levels of indirect bilirubin deposition in cells of basal ganglia à necrosis of neurons.**

***Indirect hyperbilirubinemia; usually at 20 mg% and at lower levels in preterm:***

- ***Stage I:*** Irritability, lethargy, poor Moro reflex, high pitched cry.
- ***Stage II:*** Convulsions, coma, oculogyric crises and may end by death.
- ***Stage III:*** Apparent improvement.
- ***Stage IV:*** Late sequelae: cerebral palsy, deafness, choreoasthetosis, MR.

***Treatment:*** Prevention and treatment as cerebral palsy

### *BIND score*

BIND score, which was first introduced by Johnson et al. in 1999 (as shown in **Table 2**) quantifies the severity and progression of ABE.

**Table 2. Bilirubin-Induced Neurologic Dysfunction**

| Clinical Signs | BIND Score | Date:<br>Time: | Date:<br>Time: |
|---|---|---|---|
| **Mental Status** | | | |
| Normal | 0 | | |
| Sleepy but arousable; decreased feeding | 1 | | |
| Lethargy, poor suck and/or irritable/jittery with strong suck | 2 | | |
| Semi-coma, apnoea, unable to feed, seizures, coma | 3 | | |
| **Muscle Tone** | | | |
| Normal | 0 | | |
| Persistent mild to moderate hypotonia | 1 | | |
| Mild to moderate hypertonia alternating with hypotonia, beginning arching of neck and trunk on stimulation | 2 | | |
| Persistent retrocollis and opisthotonus - bicycling or twitching of hands and feet | 3 | | |
| **Cry Pattern** | | | |
| Normal | 0 | | |
| High pitched when aroused | 1 | | |
| Shrill, difficult to console | 2 | | |
| Inconsolable crying or cry weak or absent | 3 | | |
| **TOTAL BIND SCORE** | | | |

**Advanced ABE** (score 7 - 9): urgent bilirubin reduction intervention is needed to prevent further brain damage and reduce the severity of sequelae
**Moderate ABE** (score 4 - 6): urgent bilirubin reduction intervention is likely to reverse this acute damage
**Mild ABE** (score 1 - 3): subtle signs of ABE

Note: An abnormal or 'referred' Auditory Brainstem Response (ABR) is indicative of moderate ABE. Serial ABR may be used to monitor progression and reversal of acute auditory damage and could be indicative of the effectiveness of bilirubin reduction strategy.

**Adapted:** Johnson L, Bhutani VK, Karp K, et al. Clinical report from the pilot USA Kernicterus Registry (1992 to 2004). J Perinatol. 2009 Feb;29 Suppl 1:S25-45

# B- Neonatal Hypoglycemia

**The most common metabolic problem in newborns.**

***Neonatal hypoglycemia is defined as a serum glucose:***

- ***Less than 35 mg/dl*** during the ***1st 3 hours.***
- ***Less than 40 mg/dl*** between ***3-24 hours.***
- ***Less than 45 mg/dl*** after ***24 hours.***

***Alternatively:***

- Less than 30 mg/ dL in the first 24 hours
- Less than 45 mg/ dL thereafter

## Major long-term sequelae

***Neurologic damage*** resulting in mental retardation, recurrent seizure activity, developmental delay, and personality disorders.

Some evidence suggests that severe hypoglycemia may ***impair cardiovascular function.***

## Causes & Risk Factors

***Babies who are more likely to have hypoglycemia include those with:***

***1- Inadequate substrate or enzyme function as in:***

- Intrauterine growth retardation
- Prematurity
- The smaller of discordant twins
- Infant of toxemic mothers
- Infants with placental abnormalities

***2- Hyperinsulinism as seen in:***

- Infants of diabetic mothers
- Erythroblastosis fetalis
- Functional B cell hyperplasia
- Panhypopituitarism
- Beckwith syndrome (macrosomic baby with intractable hypoglycemia, macroglossia, visceromegaly, exomphalos and facial nevus)

***3- Increased metabolic needs: (Born under significant stress)***

- Cases of RDS
- Perinatal asphyxia
- Polycythemia
- Hypothermia
- Systemic infections
- CHD (cyanotic)
- Infants with heart failure

***4- Metabolic diseases and inborn errors of metabolism as in:***

- Galactosemia
- Glycogen storage diseased
- Methyl malonic acidemia

## Clinical Picture

***1- It may be completely asymptomatic.***

***2- The onset of symptoms varies from a few hours to a week after birth.***

***3- The presenting symptoms are:***

- ***General findings:*** weak high-pitched cry, poor feeding, hypothermia, diaphoresis.
- ***Neurologic signs:*** Tremors, jitteriness, irritability, hypotonia, lethargy, seizures.
- ***Cardiorespiratory signs:*** tachypnea, apnea, cyanosis, severe pallor and cardiac arrest.

## Laboratory studies

***Serum or plasma glucose levels.***

***Serum insulin (insulin/ glucose ratio):***

- When blood glucose is less than 40 mg/ dL , plasma insulin concentration should be less than 5 and no higher than 10 µU/ mL.
- This testing may not be available in the emergency department.

***Urine:***

- Obtain a first voided urine dipstick for ketones.
- Failure to find large ketones with hypoglycemia suggests that fat is not being metabolized from adipose tissue ( hyperinsulinism ) or that fat cannot be used for ketone body formation (enzymatic defects in fatty acid oxidation).
- Send urine for organic acid analysis.

***Screening for metabolic errors:***

- Electrospray ionization tandem mass spectrometry in asymptomatic persons allows earlier identification of clearly defined inborn errors of metabolism.
- These disorders include aminoacidemias, urea cycle disorders, organic acidurias, and fatty acid oxidation disorders.
- Earlier recognition of these inborn errors of metabolism has the potential to reduce morbidity and mortality rates in infants with these conditions. conditions.
- This testing may not be available in the emergency department.

***Angiography:*** The detection of adenomas by celiac angiography has had limited success. The chance of detecting a tumor blush must be balanced against the potential risk of causing vascular trauma in infants younger than 2 years.

# Treatment

**Treatment is a stepwise process depending on the presence or absence of symptoms and signs, and the response of the infant at each step.**

***Early feeding*** of the newborn with breast milk or formula is encouraged. For those unable to drink, a nasogastric tube can be used.

***For those who cannot protect their airway or are unable to drink,*** nasogastric, intramuscular, intraosseous, or IV routes can be employed.

***For symptomatic infants,*** it is recommended to administer parenteral glucose. Therapy should be started while awaiting laboratory confirmation. Begin with an intravenous (IV) bolus of dextrose (200 mg/kg) given over 5 minutes (2 mL/kg of 10 percent dextrose in water).

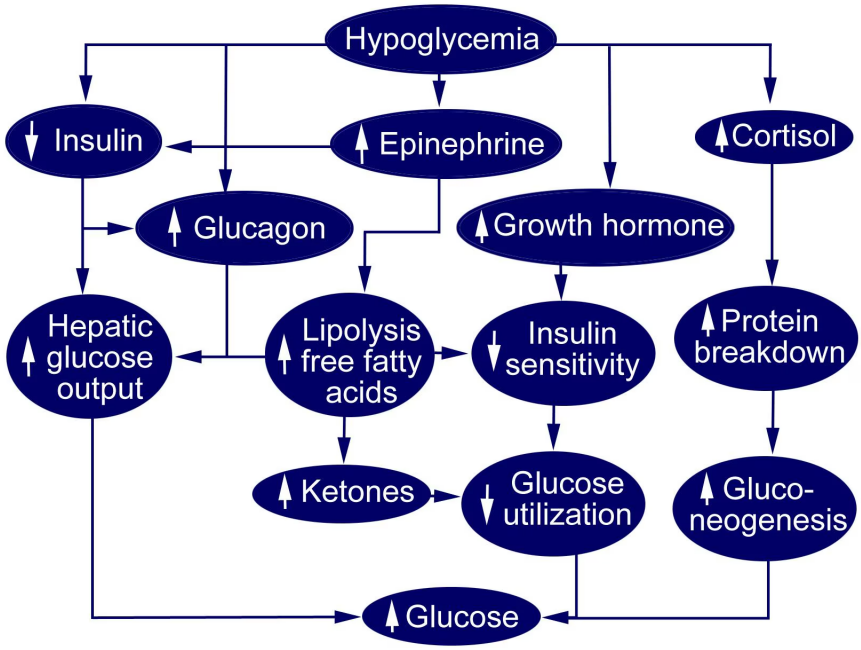
***This is followed by*** the continuous administration of parenteral glucose infusion at an initial rate of 6 to 8 mg/kg per minute.

***If hypoglycemia is persistent,*** glucose infusion rates should be increased as needed.

***In the rare patients*** who fail to maintain target blood glucose levels despite maximal glucose infusion rates, it is suggested to administer ***glucagon*** at an initial dose of 10 to 20 mcg/kg per hour as a continuous infusion.

***Other drugs*** used to raise glucose levels: dextrose, diazoxide, and octreotide. Case reports have shown that nifedipine may help to maintain normoglycemia in children with PHHI.

***Cortisol should not be used,*** **because it has minimal acute benefit and may delay the diagnosis of the cause of hypoglycemia. Cortisol stimulates gluconeogenesis and causes decreased use of glucose, which leads to overall elevated blood glucose and may mask the true cause of hypoglycemia.**



# 5 RESPIRATORY NEONATAL DISORDERS

## Apnea

**Apnea is cessation of respiration for more than 20 seconds or associated with cyanosis or bradycardia.**

***Periodic breathing*:** is a normal phenomenon found in premature and full term infants: pauses in breathing for no more than 10 seconds at a time followed by a series of rapid, shallow breaths, then breathing returns to normal without any stimulation or intervention).

## Types of apnea

### 1- Apnea of prematurity:

- It occurs in ***absence of identifiable predisposing diseases*** in a premature newborn infant.
- Due to ***immature respiratory center (central)*** or ***collapse of pharyngeal wall (obstructive)*** or ***both (combined)***

### 2- Symptomatic apnea:

***It occurs in preterm or full term neonate due to a specific pathology as:***

- ***CNS:*** drugs, seizures, hypoxic injury, intracranial hemorrhage
- ***Respiratory:*** pneumonia, obstructive lesions, severe HMD, pneumothorax.
- ***Infections:*** sepsis, necrotizing enterocolitis, meningitis.
- ***Gastrointestinal:*** oral feeding in some preterm babies, gastro-esophageal reflux, and intestinal perforation.
- ***Metabolic causes:*** hypo and hypernatremia, hypo and hyperthermia, hypoxia, hypoglycemia, hypocalcemia.
- ***Cardiovascular:*** anemia, hypovolemia, hypo and hypertension.

## Investigations

- Complete blood count, CRP & electrolytes and blood sugar.
- Cranial ultra sound & abdominal X-ray.

## Treatment

***1- Infants at risk for apnea*** should be monitored with apnea monitors

***2- Gentle cutaneous stimulation*** is often adequate.

***3- Idiopathic apnea of prematurity:*** should be treated with caffeine 20mg/Kg loading to be followed by 5 mg/Kg given every 24 hours orally or IV. Other drugs could be used as Aminophillyn and Doxapram.

***4- Symptomatic apnea:*** Treat underlying disease as anaemia, sepsis, reflux, or hypoglycemia.

***5- CPAP (continuous positive airway pressure), -/+ mechanical ventilation.***

# Neonatal Respiratory Distress & Failure

***It is the most common emergency related to the newborn infants. It is classified into:***

1. Central respiratory failure.
2. Peripheral respiratory difficulty.

## Central respiratory failure

***This originates in the central nervous system due to:***

1. Narcosis due to: morphine, barbiturates or from maternal anesthesia.
2. Prenatal or perinatal anoxia.
3. Intracranial hemorrhage (anoxic or traumatic).
4. Congenital anomalies of CNS.
5. CNS infection.

### Clinical Manifestations & Management

- Apnea
- Cyanosis.

### Management:

- ***As secondary apnea:*** treat the cause, tactile stimulation and in severe cases mechanical ventilation.

# Peripheral respiratory difficulty

## Etiology:

### *A. Extra pulmonary causes:*

- Choanal atresia (unilateral or bilateral).
- Congenital goiter
- Retro-micrognathia: pierr robin syndrome
- Diaphragmatic hernia.
- Tracheo-esophageal fistula.
- Neonatal sepsis (septicemia).
- Vocal cord paralysis or laryngomalacia.
- Abdominal distension/ascites.
- PDA & heart failure
- Metabolic acidosis

### *B. Pulmonary causes:*

- Congenital lobar emphysema.
- Transient tachypnea of neonate
- Hyaline membrane disease (IRDS).
- Meconium aspiration.
- Congenital/ neonatal Pneumonia.
- Pulmonary hemorrhage.
- Pneumothorax.
- Heart failure (PDA/myocarditis)
- Sepsis.

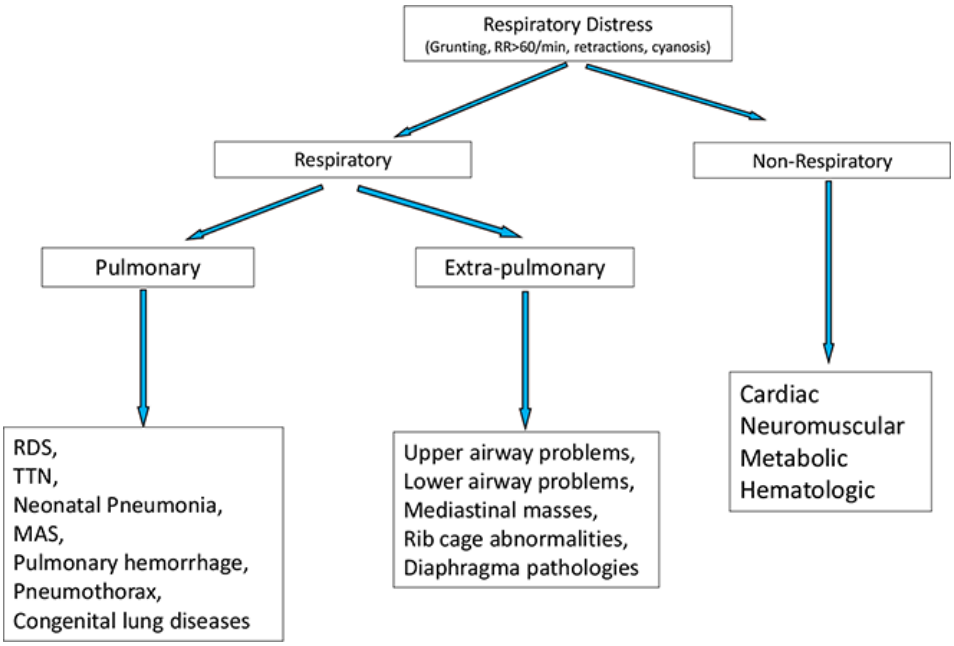
## Clinical manifestations

- ***Tachypnea:*** respiratory rate > 60/minute.
- ***Retractions*** (intercostal, subcostal and xiphoid retraction).
- ***Grunting:*** it is expiration against partially closed glottis.
- ***Flaring*** of ala nasi
- ***Cyanosis*** may develop in severe cases.

## Treatment:

- Give oxygen, CPAP, Ventilation.

***Specific measures:*** as surfactant for RDS, antibiotics for pneumonia or indomethacin for PDA.



# Hyaline Membrane Disease (HMD)

# Infantile Respiratory Distress Syndrome (IRDS)

**The term describes a syndrome of neonatal respiratory distress in which hyaline membrane with atelectasis are the principal findings at autopsy.**

The condition is the major cause of death in the neonatal period.

The disease occurs early in premature and the incidence is inversely related to thegestational age and weight.

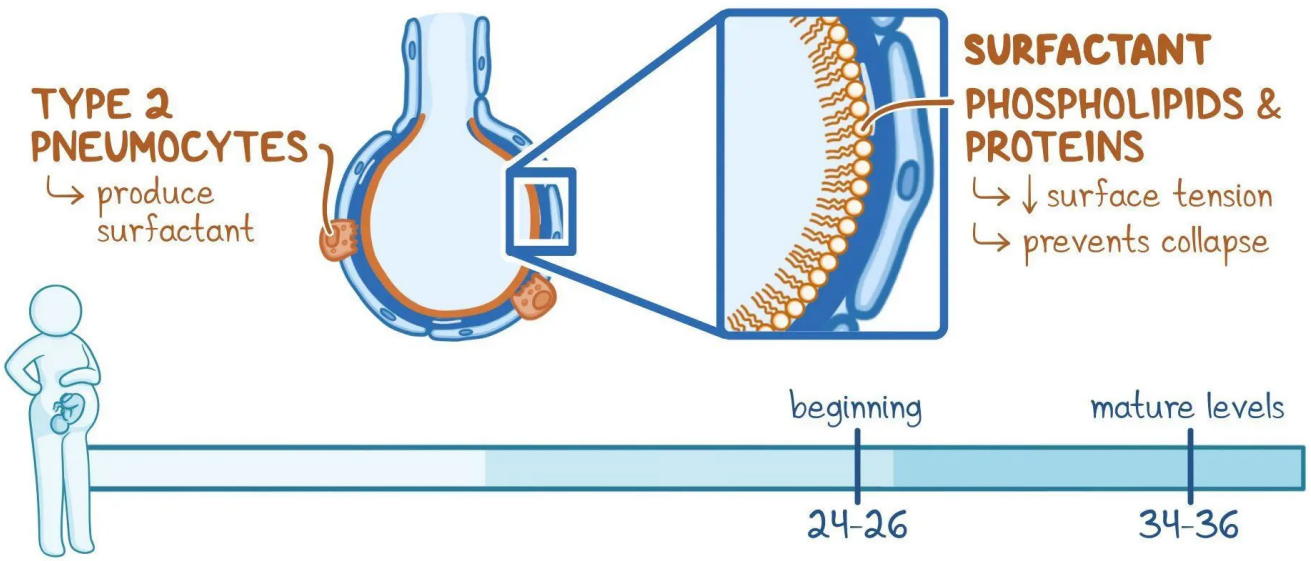
It is rare at term except in the infant of diabetic mother, those delivered with C.S. and those with history of asphyxia at birth.

## Predisposing factors:

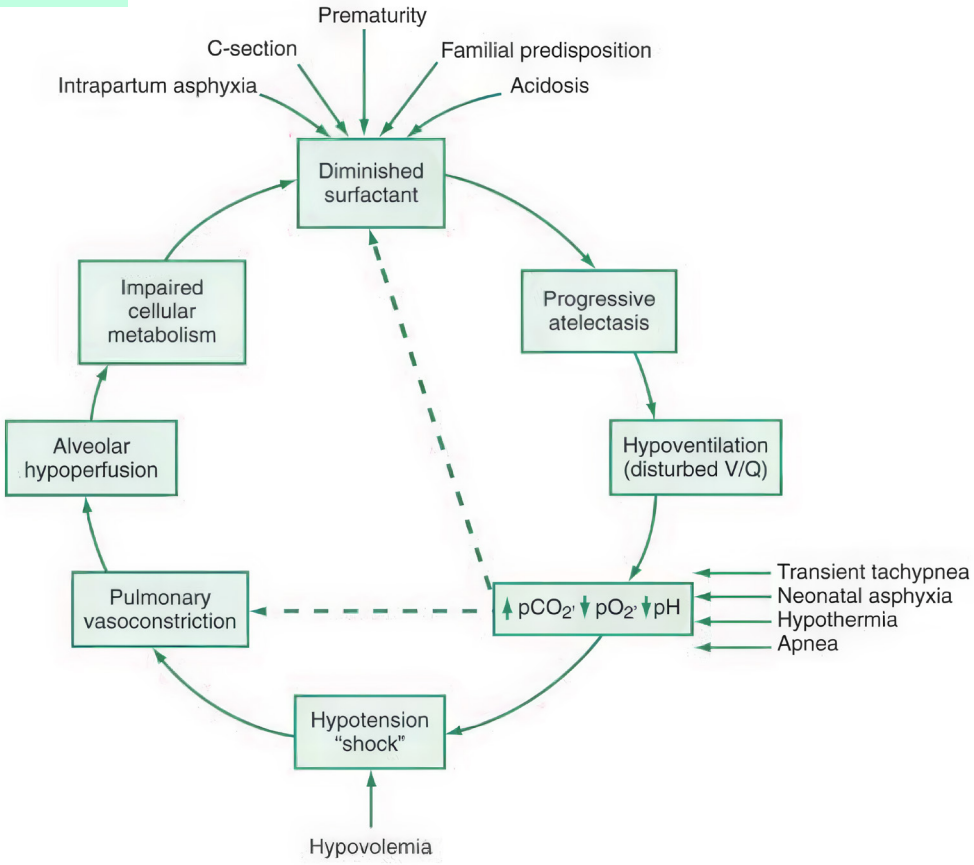
- Prematurity, diabetic mother and C.S.
- Asphyxia, hypoxemia, hypovolemia.
- Hypotension and cold stress may suppress surfactant synthesis.

## Etiology:

- ***Surfactant deficiency,*** together with small respiratory units result in atelectasis, increased airway resistance, insufficient alveolar ventilation with asphyxia, hypoxemia and acidosis.
- Surfactant is a dipalmitoyl-phosphatidyl choline and is a phospholipid, which prevents alveolar collapse by reducing alveoli surface tension. It is produced by type-II pneumocytes and is seen at about 24 weeks' gestation. It causes an increase in lung compliance.



## Pathogenesis



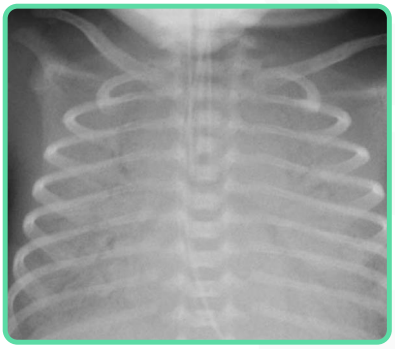

Contributing factors in the pathogenesis of hyaline membrane disease. The potential "vicious circle" perpetuates hypoxia and pulmonary insufficiency.

## Clinical manifestations

- • Signs of hyaline membrane disease usually appear within minutes after birth but they may not be recognized for several hours until rapid shallow respiration develops (RR more than 60/min.) together with grunting, flaring of ala nasi, chest retraction and cyanosis which manifest in severe cases.
- • Chest auscultation reveals diminished air entry on the affected side+/-fine rales.

## Diagnosis is confirmed by

***1. X-ray chest:*** reticulo-granular appearance, prominent air bronchograms, total obscuration of the cardiac silhouette and widespread ground glass opacity.



***2. The laboratory findings*** are characterized by ***progressive hypoxemia*** (↓ O2), hypercarbia (↑ CO2) and ***metabolic acidosis.***

## Prevention

1. Prevention of prematurity.
2. Transfer of the mother at risk for preterm delivery to equipped center.

**The best transportation of the preterm neonate is in the gravid uterus.**

3. Antibiotics should be given to mothers with preterm prelabour rupture of the membranes as this reduces the risk of preterm delivery
4. Avoidance of unnecessary C.S.
5. Prenatal administration of betamethazone before delivery (33 weeks gestation or less) reduces the incidence of IRDS (best if given 3-4 days before delivery).
6. Early treatment of hypoxia, hypothermia, and hypoglycemia.

## Treatment:

**Treatment of hyaline membrane disease requires *careful and frequent monitoring* of heart and respiratory rate, oxyhemoglobin saturation, PO2, PCO2, bicarbonate, electrolytes, blood sugar and temperature. This should be carried out in *neonatal intensive care unit* by *experienced and skilled personnel.***

***The main lines of treatment include:***

**1. Incubator care to regulate temperature and humidity.**

**2. Calories and fluid**

- Should be provided IV for the 1st 48 hrs.
- 10% glucose should be given at a rate of 65 - 80 ml/kg per day.
- Also 2.5-3gm/kg amino acids should be given to avoid negative nitrogen balance.

**3. Oxygen administration:**

- The ideal oxygen therapy is to give ***warm humidified $O_2$*** at a concentration to keep oxygen saturation by pulse oximeter placed over the right hand between 89 and 94%.

**4. Assisted ventilation:** It aims at increasing the functional residual capacity.

***Indications:***

a) Arterial blood pH less than 7.2.
b) Arterial blood CO2 more than 60 mm Hg.
c) Arterial blood O2 less than 50 mm Hg at O2 concentration of 60-70 %.
d) Persistent or recurrent life threatening apnea.

***Methods:***

a) The simplest and most physiological method is the use of nasal continuous airway pressure (nCPAP).

b) Positive pressure ventilator with endotracheal tube in case of CPAP failure.

c) If the neonate is in need for intubation, the administration of surfactant through the endotracheal tube to be followed by extubation and use of CPAP as early as possible is advised.

**5. Other additional treatments include:**

- Antibiotics to treat or to prevent pneumonia till receiving the results of cultures.
- Enteral feeding using breast milk as early as possible.
- Close monitoring of blood pressure and correct hypotension.

# Complications

***of hyaline membrane disease and intensive care management:***

1. Encephalopathy and intraventricular hemorrhage.
2. Complications of ET intubation and ventilation such as pneumothorax and pneumopericardium.
3. Complications of umbilical vessels catheterization as sepsis, necrotizing enterocolitis and portal vein thrombosis.
4. Complications of oxygen toxicity as retinopathy of prematurity and broncho- pulmonary dysplasia (Chronic lung disease).
5. Anemia due to prematurity, frequent blood sampling and nutritional deficiency.

# Prognosis

***Depends on early diagnosis and management in well-equipped NICU by skilled personnel.***

- In good centers, about 50% of those less than 1 kg survive and the morbidity progressively decreases at higher weights to over 95% if more than 2.5 kg.
- The long-term prognosis for normal pulmonary function in most infants surviving hyaline membrane disease is excellent.

**Extubation as early as possible with institution of CPAP minimizes lung injury from barotrauma and oxygen toxicity and reduces the risk of chronic lung disease.**

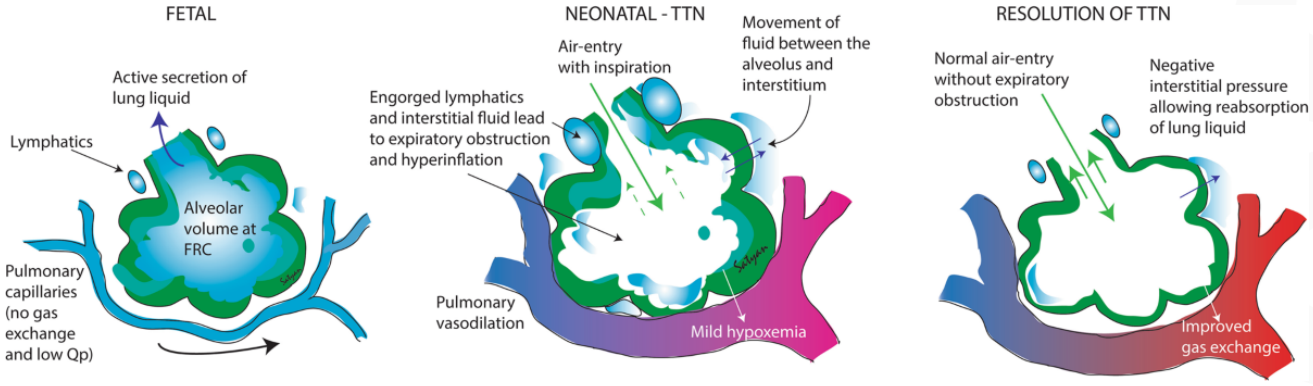
# Transient Tachypnea of the Neonate (TTN)

**Respiratory distress** with transient and mild tachypnea in the term and near term neonate especially after CS due to delayed clearing of lung fluids.

- The baby ***improve in 48 hours.***
- CBC, CRP shows ***no evidence of sepsis.***

## Pathophysiology

- Disruption or delay in clearance of fetal lung fluid withtransient pulmonary edema.
- Increased fluid volume with ↓ lung compliance and ↑ airway resistance




## Risk Factors

- Elective cesarean section
- Macrosomia and IDM's
- Prolonged labor
- Excessive maternal sedation
- Maternal fluid overload, esp. with oxytocin infusion
- Delayed clamping of the umbilical cord

# Clinical Manifestations

***Infant:*** usually near-term or term

***Onset:*** within 6 hrs after delivery

***Degree:*** mild to moderate RD

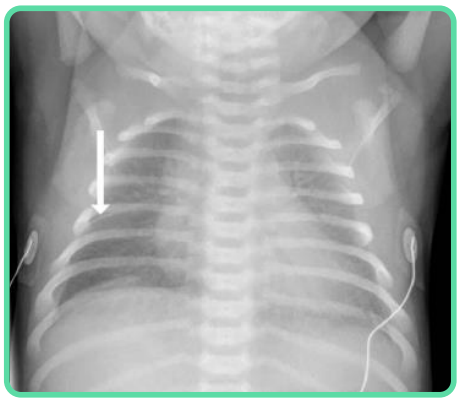
***Auscultation:*** usually good air entry +/- crackles

***Persists usually for*** 12-24 hrs (up to 72 hrs)

***Exclude other causes*** of RD in the first 6 hrs of life

**Spontaneous improvement is an important marker of TTN**

## Investigations



***CBC and CRP***

***Blood gas analysis***

***Chest x-ray:***

- Prominent perihilar streaking
- Fluid in the fissure
- Prominent pulmonary vascular markings

**Chest x-ray usually shows evidence of clearing by 12-18 hrs with complete resolution by 48-72 hrs**

## Management

***General management of RD***

***Antibiotic therapy,*** depends on history and infant's clinical status; can be stopped at 48-72 hrs if cultures are -ve.

***No role for diuretics***

## Prognosis

- The disease is ***self-limiting***
- No risk of residual pulmonary dysfunction

# Meconium Aspiration Syndrome

- Fetal distress or asphyxia during birth will lead to passage of meconium and provoke vigorous respiratory efforts.
- Meconium and blood, will be inhaled into the upper airway.
- The baby has chemical pneumonitis, and obstruction of air way with emphysematous chest.

### Clinical picture:

- Severe respiratory distress with hyperinflated chest

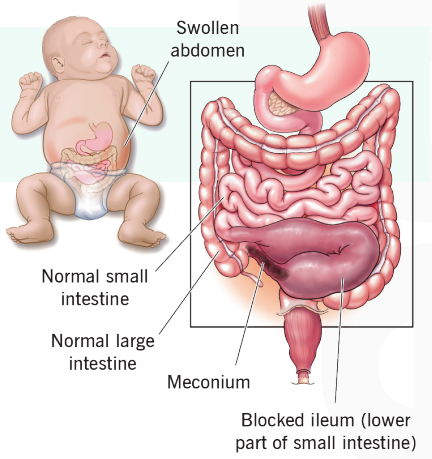
### Management:

- Endotracheal suction at birth if the baby is NOT VIGOROUS.
- Antibiotics, Oxygen and CPAP if needed
- Surfactant endotracheally (meconium deactivate endogenous surfactant)

# 6 GASTROINTESTINAL NEONATAL DISORDERS

## Meconium ileus

**Meconium ileus, or impaction of inspissated meconium in the distal small bowel, causing obstruction of the small intestine at the level of the terminal ileum.**




- It ccounts for up to 30% of cases of neonatal intestinal obstruction.
- It is common in patients with ***cystic fibrosis (CF)*** in whom the lack of fetal pancreatic enzymes inhibits digestive mechanisms, and meconium becomes viscid.
- Approximately 10 percent of patients with CF present as neonates with MI; in most cases.

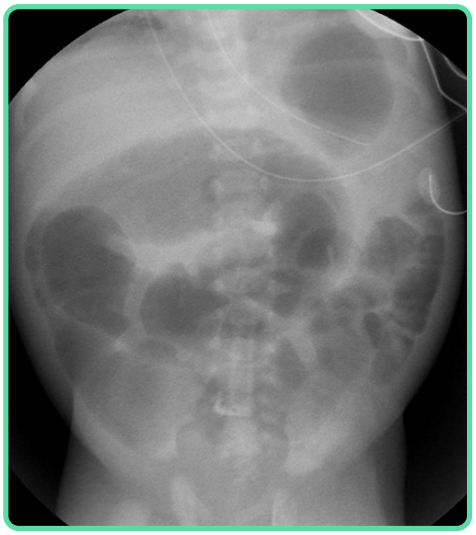
### Presentation and diagnosis

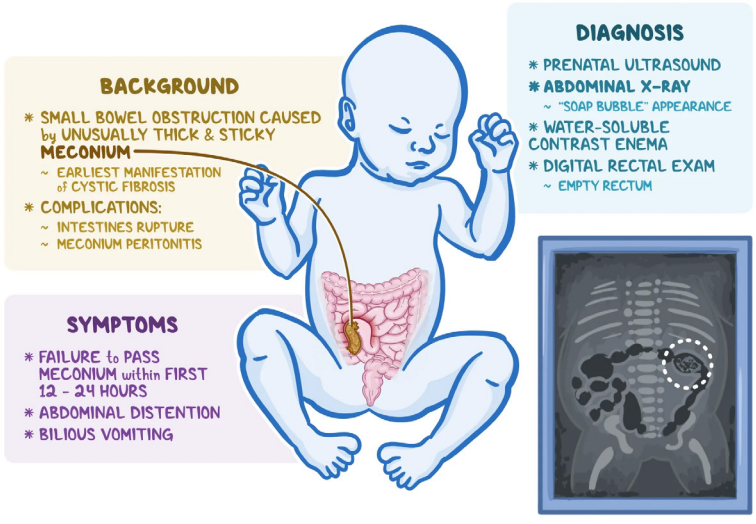
- Infants with MI generally present during the ***first three days of life*** with ***abdominal distension*** and ***failure to pass meconium***, ***with or without vomiting.***

### Treatment

***For simple meconium ileus*** a high-osmolarity gastrografin enema.

***If the procedure is unsuccessful or perforation of the bowel wall is suspected,*** a laparotomy is performed and the ileum opened at the point of largest diameter of the impaction.





# Neonatal Necrotizing Enterocolitis (NEC)

**NEC is characterized by varying degrees of mucosal or transmural necrosis of the intestine of the preterm neonate of unknown etiology.**

- It is the ***most common life-threatening emergency*** of the gastrointestinal (GI) tract in the newborn period.

## Risk Factors:

- The greatest risk factor for NEC is ***prematurity.***
- NEC rarely occurs before the initiation of enteral feeding and is much less common in infants fed human milk.
- Aggressive enteral feeding may predispose to the development of NEC.
- Although nearly ***90% of all cases of NEC occur in preterm infants,*** the disease can occur in full-term neonates.
- ***NEC in term infants is often a secondary disease***, seen more frequently in infants with history of birth asphyxia, Down syndrome, congenital heart disease, rotavirus infections, gastroschisis, and Hirschsprung disease.

## Pathogenesis:

- Rapid advancement of milk feed in small preterm babies especially if artificial formula in a septic hypotensive preterm.

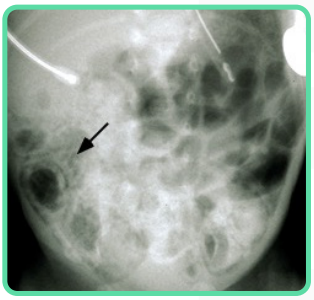
## Clinical manifestations

- Preterm baby not doing well with ***abdominal distension ± bloody stool.***
- Symptoms and signs of ***sepsis.***

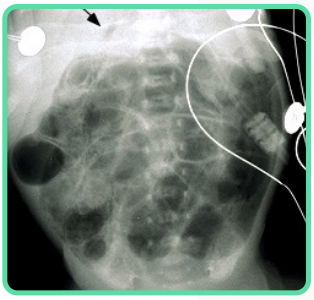
## Diagnosis:

### *Plain abdominal X-ray:*

- Plain abdominal radiographs are essential to make a diagnosis of NEC.
- The finding of ***pneumatosis intestinalis (air in the bowel wall)*** confirms the clinical suspicion of NEC and is diagnostic.
- Portal venous gas is a sign of severe disease, and pneumoperitoneum indicates a perforation.



***Ultrasound with Doppler flow assessment*** may be useful to evaluate for free fluid, abscess, and bowel wall thickness, peristalsis, and perfusion.



### *Clinical picture*

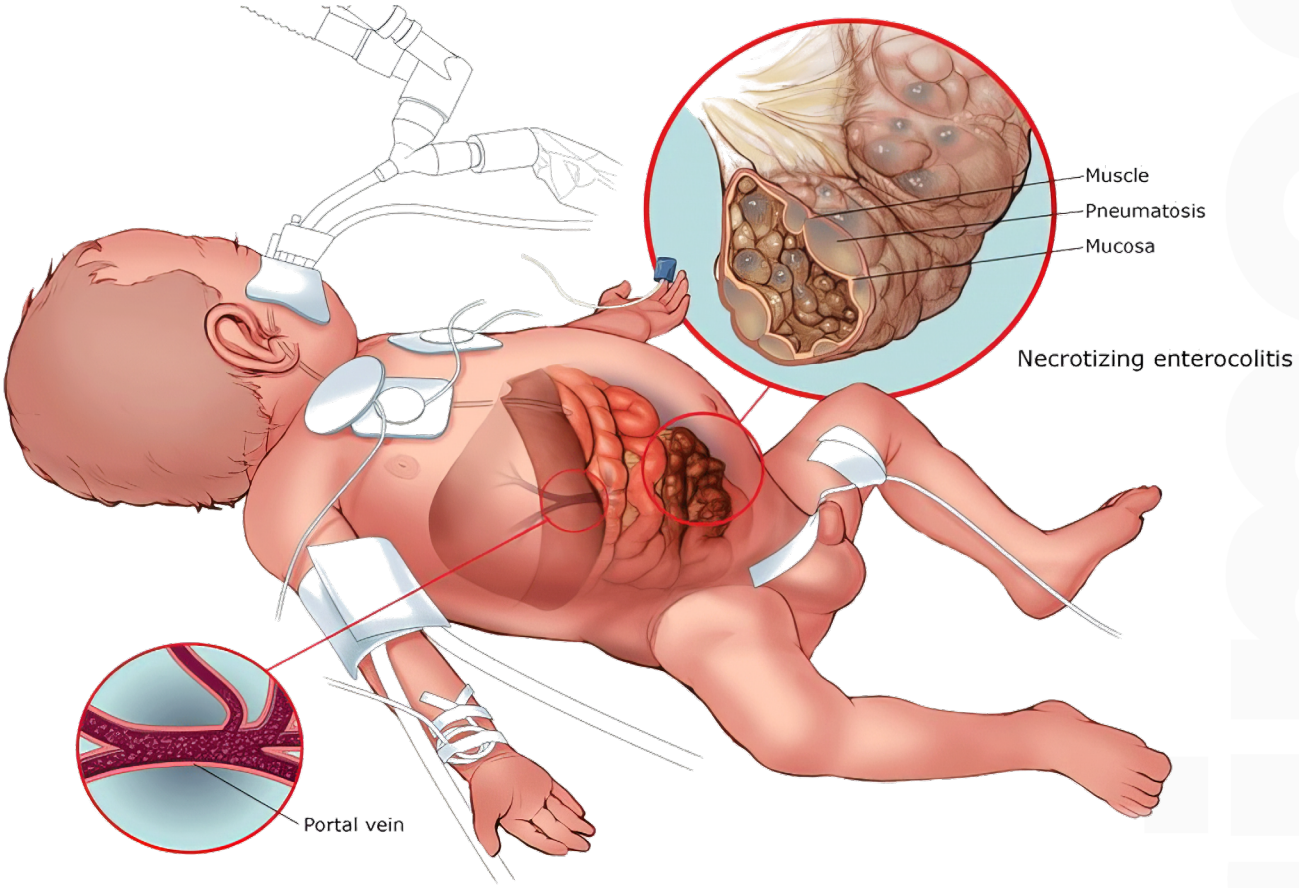
### *Thrombocytopenia*

## Treatment

***There is no definitive treatment for established NEC, so therapy is directed at providing supportive care and preventing further injury:***

- Cessation of feeding, nasogastric decompression, and administration of IV fluids.
- Careful attention to respiratory status, coagulation profile, and acid-base and electrolyte balances are important.
- Systemic antibiotics.
- A surgeon should be consulted early in the course of treatment.

**The only absolute indication for surgery is evidence of perforation on abdominal radiograph (pneumoperitoneum)**



# HEMATOLOGICAL NEONATAL DISORDERS

## Anemia

### Normal newborn

- Hblevel averages 17 – 19 gm/dL
- Mild reticulocytosis & normoblastemia
- High RBC mass → indirect hyperbilirubinemia → physiologic jaundice
- **Total leukocyticcount (TLC):**
  - After 24 hrs → $25x10^9$/L with relative neutrophilia
  - After 1 week → $15x10^9$/L with relative lymphocytosis

### Types

#### 1) Physiologic anemias in neonatal period

##### *i. Physiologic anemia of infancy*

- In utero, the fetal aortic oxygen saturation is 45%, erythropoietin levels are high, and RBC production is rapid.
- After birth, there is a rapid fall in Hb, starting by the end of the 1st week of life and progressively declining reaching a nadir (lowest level) at 6-8 weeks after birth.
- ***This is a physiologic phenomenon called physiologic anemia of infancy and is due to:***
  1. Shorter life span of the RBCs containing the fetal hemoglobin.
  2. Abrupt cessation of erythropoiesis at birth (increased oxygen saturation).
- Once the nadir is reached, RBC production is stimulated (gradual rise in Hb).

##### *ii. Anemia of prematurity*

- It is an exaggeration of the normal physiological anemia.
- In addition, small premature infants develop rapidly vitamin E deficiency.
- Such vitamin E deficiency causes more shortening of the life span of the RBCs.
- Nadir occurs earlier and reaches lower levels of Hb

### 2) Pathologic anemias in the neonatal period by time of occurrence:

#### *Early neonatal period (at birth or in 1st 24 hours):*

1. Fetal hemorrhage.
2. Feto-maternal transfusion.
3. Twin to twin transfusion.
4. Fetal hemolysis (Rh incompatibility, ABO, alpha thalassemia).
5. Early cord clamping.
6. Intrauterine infection.
7. Neonatal hemorrhage.

#### *Anemia in the 1st week (2nd-7th days):*

1. Hemolytic disease of the newborn.
2. Improperly tied umbilical cord.
3. Subcapsular hepatic hemorrhage.
4. Large cephalhematoma.
5. Suprarenal hemorrhage.
6. Septicemia.
7. Hemorrhagic disease of newborn.

#### *Anemia after 1st week:*

1. Congenital hypoplastic anemia.
2. Vitamin E deficiency.
3. Chronic hemolysis as enzyme defects, alpha-thalassemia and cong. infection.

## Investigations

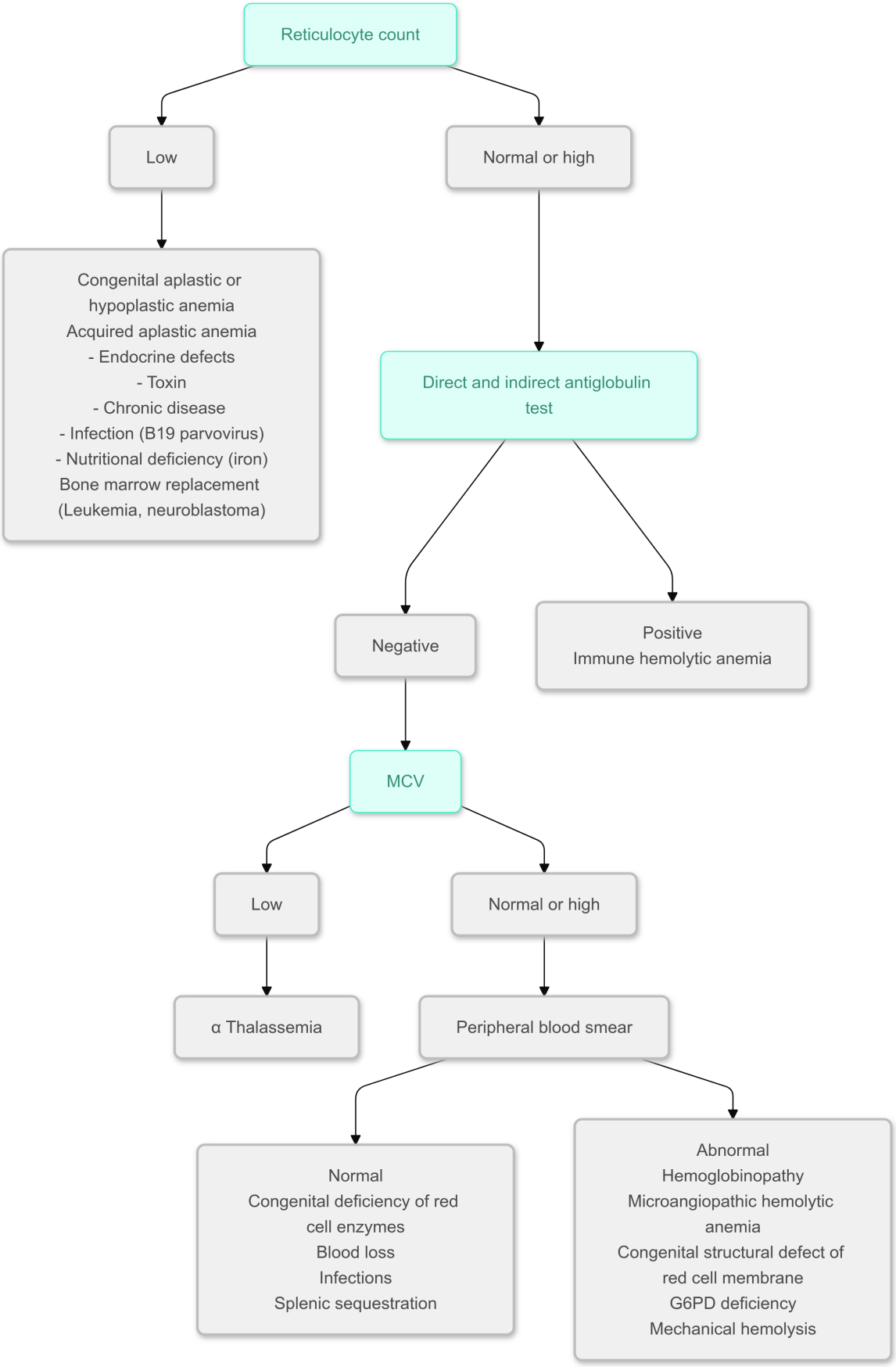
- Complete blood count: Hb% + indices.
- Reticulocytic count / bilirubin level
- Coomb's test.
- G6PD assay / Osmotic fragility.
- Sepsis workup
- STORCH-EB screening.

## Treatment

***Treatment of underlying cause.***

***If anemia is severe*** transfusion of cross matched blood or RBCs.

***Recombinant human erythropoietin (r-HuEpo)*** can prevent and treat many cases of chronic neonatal anemia.




Laboratory algorithm for the diagnosis of neonatal anemia. When used in conjunction with a complete history and physical examination, the cause of the anemia often can be identified. In some cases, ancillary laboratory testing will be of assistance.

# Polycythemia

**A central venous hematocrit of > 0.65**

- Mostly asymptomatic
- Hyperbilirubinemia
- Lethargy, poor feeding, hypotonia
- Stroke, renal vein thrombosis
- Tachypnea, PPHN

## Causes

1) Placental red cell transfusion → delayed cord clamping / stripping / uterine contractions before clamping
2) Materno-fetal hge / twin-to-twin transfusion
3) Placental insufficiency
4) IDM / LGA
5) Dehydration / maternal propranolol

## Treatment

***Increase fluid intake & repeat Ht after 4 to 6 hours***

***Partial exchange transfusion*** using normal saline

- ***Volume of exchange:***

$$\frac{\text{Blood volume (80} \times \text{weight)} \times \text{(Observed} - \text{desired hematorcit)}}{\text{Observed hematocrit}}$$

# Hemorrhagic disease of the newborn

**It is spontaneous bleeding tendency from 2nd-6th day post-natal.**

## Etiological factors

- Vitamin K deficiency due to absent intestinal flora.
- Liver immaturity ➔ decreased factors II, VII, IX and X (vitamin. K dependent).

## Clinical picture

- GIT, nose bleeding (common), umbilical stump bleeding.
- Intracranial and pulmonary hemorrhage (rare), it is more common in preterms.

## Types:

***Early:*** First 24hr / severe / GIT or ICH/ maternal medications (anticonvulsants)

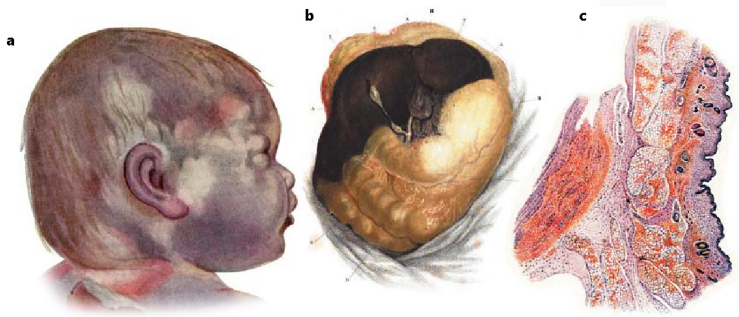
***Classic:*** 2 – 7 days old / breast-fed or poor feeding / TPN / AB

***Late:***

- Between 2 – 8 wk (up to 12 weeks) ➔ biliary atresia, malabsorption, breast-fed.
- It ***commonly presents with ICH***

## Laboratory investigations

- ***CBC:*** normal platelet count.
- ***Bleeding time:*** normal.
- ***Clotting time:*** prolonged
- ***Platelet count:*** normal.
- ***PT:*** prolonged
- ***PTT:*** prolonged.
- ***Decreased levels of factors II, VII, IX, X.***




## Prevention:

- ***Vitamin K 1 mg IM*** to all newborns immediately after birth.

## Treatment:

- ***Vit. K (Konakion) 5 mg IM.***
- ***Fresh frozen plasma in severe cases*** 10 ml/Kg.

# DD of hemorrhage in a newborn

### *1- Hemophilias: (inherited coagulation disorders)*

- Hemophilia A, B.
- Von Willebrand hemophilia.
- There is +ve family history in most cases in hemophilias, PT is normal and PTT is prolonged.
- ***Treatment*** is transfusion of the deficient clotting factor.

### *2- Disseminated intravascular coagulopathy (DIC):*

- Sick newborn: weak suckling, weak cry- hypothermia, apnea
- Due to sepsis or anoxia, acidosis
- Bleeding at puncture sites, bad general condition.
- Consumption of platelets and clotting factors (Platelet is decreased, PT and PTT are prolonged + Increased fibrin-degradation products =FDPs).
- ***Treatment:*** treatment of underlying ***causes*** + ***FFP*** 10mL /Kg.

### *3- Neonatal thrombocytopenia: platelet count <150.000/mm3*

- Isoimmune thrombocytopenia
- Thrombocytopenia absent radius syndrome
- Congenital infections
- Maternal ITP
- Maternal SLE
- Neonatal sepsis

### *4- Thrombasthenia*

- ***Hereditary:*** Glanzmann, Bernard-Soulier
- ***Acquired:*** maternal anti-platelet drugs

### *5- Swallowed maternal blood syndrome:*

- The infant may swallow the mother's blood during delivery or from a fissure in her nipple. Later, blood is passed in the stools or is vomited.
- In these cases the blood contains mainly adult hemoglobin. The latter is not alkali resistant while fetal hemoglobin is.
- Differentiated by Apt test (Alkali denaturation test) or Kleihauer-Betketest (acid elution test)..

# How to investigate bleeding in newborn

- CBC (especially platelet count)
- PT, PTT
- BT
- Clotting factor assay
- CT

| Clinical Evaluation | Platelets | PT | PTTK | Likely diagnosis |
|---|---|---|---|---|
| Sick | ↓ | ↑ | ↑ | DIC |
| | ↓ | N | N | Sepsis, NEC |
| | N | ↑ | ↑ | Liver disease |
| | N | N | N | Stress bleed |
| Healthy | ↓ | N | N | IT, occult infection, thrombosis, B.M. hypoplasia or infilteration |
| | N | ↑ | ↑ | Hemorrhagic disease of newborn ( vit. K deficiency ) |
| | N | N | ↑ | Hereditary clotting factor deficiencies (Hemoplilia A/B, VWD) |
| | N | N | N | Bleeding due to local factor ( trauma, anatomic anomalies); Factor XIII deficiency |

PT = prothrombin time; PTT = partial thromboplastin time; ↓ = decreased; ↑ = increased; DIC = disseminated intravascular coagulation; N= normal .

## Treatment

- Treatment of underlying cause
- FFP in hemophilias, hemorrhagic disease of newborn and liver disease.
- Platelet transfusion in thrombocytopenia if severe.
- Replacement of deficient factor in inherited clotting defects (hemophilia).
- Packed RBCs for hemorrhagic anemia.

# Thrombosis

***Naturally-occurring anticoagulants:*** Protein C & S (vitamin K dependent proteins)

### Thrombophilia

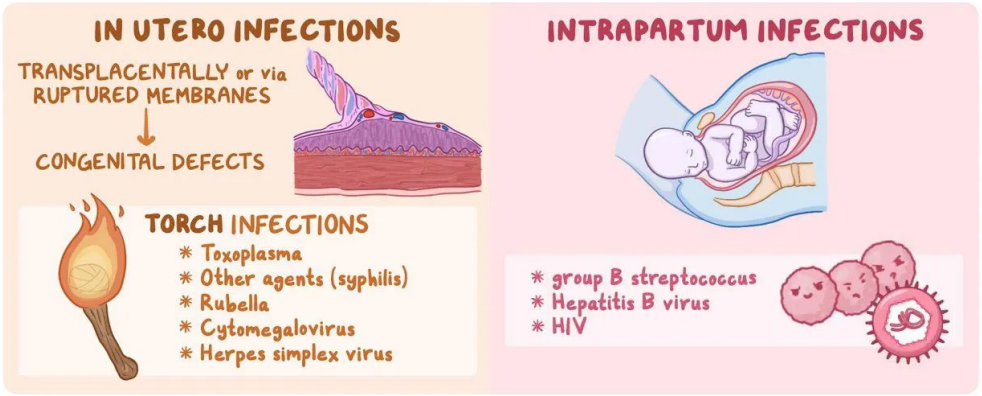
- Defect in anticoagulation or fibrinolytic pathways
- Arterial or venous
- Treated by tPA (tissue plasminogen activator)
- Indwelling catheters
- Color change in abdomen or limb

# 8 FETAL & NEONATAL INFECTIONS

## Fetal (congenital) infections

**Congenital infections can develop in the neonate transplacentally or perinatally (from vaginal secretions or blood). They are major causes of maternal and fetal morbidity and mortality.**

| In utero (transplacental) infections | Infections acquired during birth (hematogenous or genital route) |
|---|---|
| • Toxoplasmosis<br>• Rubella<br>• Cytomegalovirus disease<br>• Parvovirus B19<br>• Varicella-zoster virus (VZV)<br>• Human immunodeficiency virus (HIV)<br>• Listeria monocytogenes<br>• Syphilis, congenital tuberculosis | • Gonococcal ophthalmia<br>• Chlamydia trachomatis<br>• Pneumonia and meningitis<br>• Herpes simplex infections (HSV)<br>• Hepatitis B virus<br>• HIV<br>• HZV |




### Consequences of Fetal Exposure to Infection:

***Infection of the fetus can result in:***

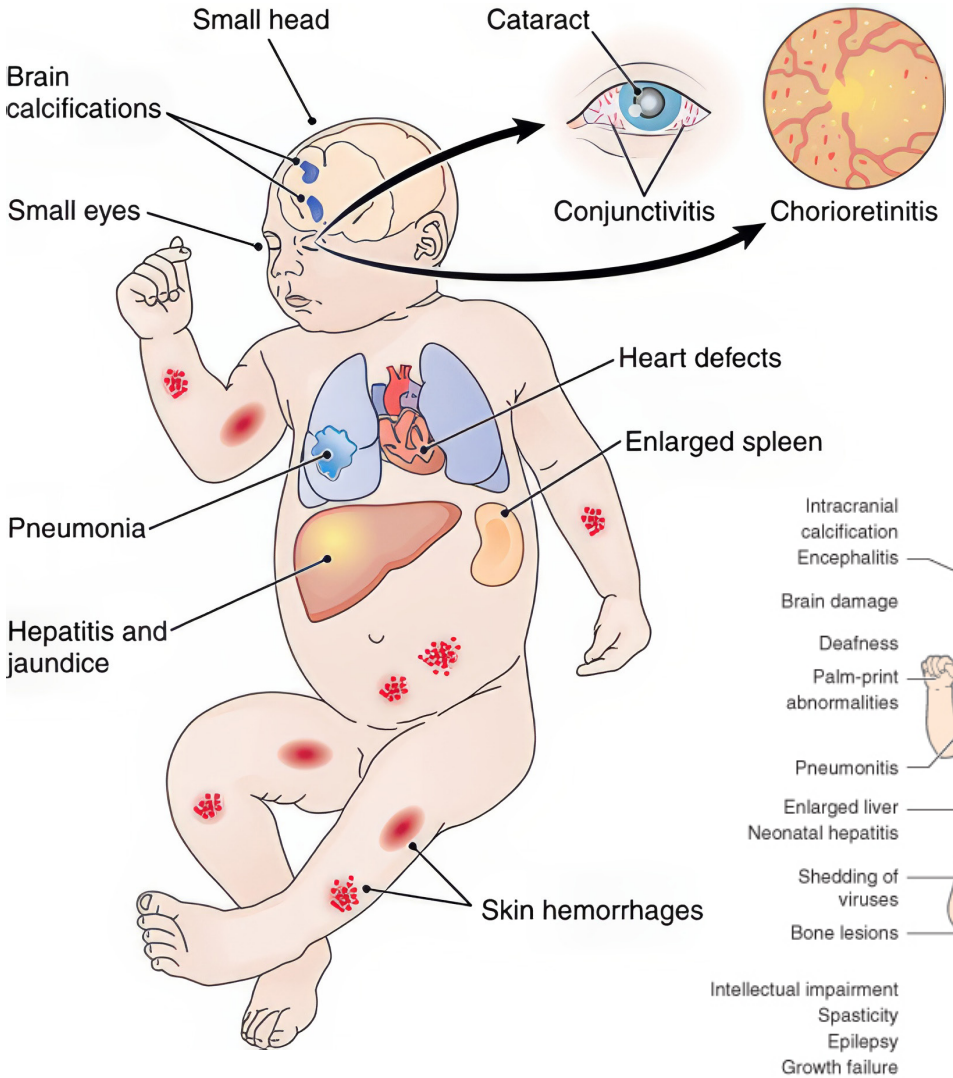
- Death of embryo and abortion/Intrauterine fetal death/Stillbirth
- Premature labor
- Manifestations of infection at birth
- Clinically free at birth, clinical manifestations develop afterwards
- Free from infection

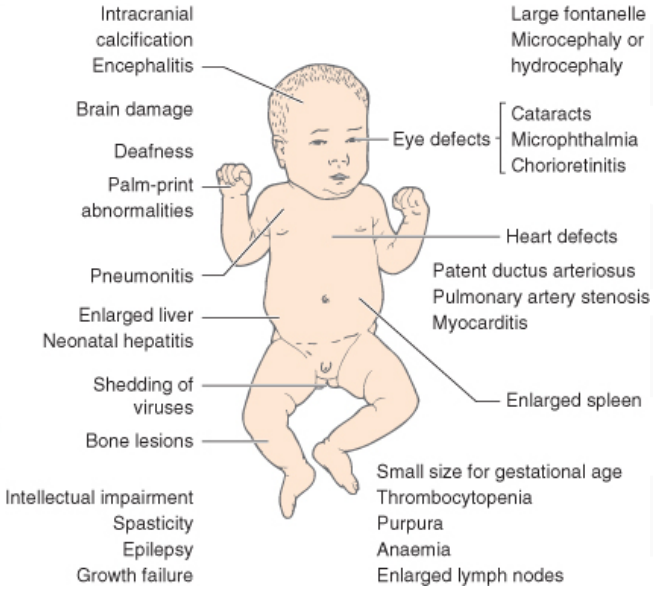
## Clinical Manifestations of congenital Infections

***Are rarely disease specific but include:***

- ***Prematurity, IUGR, or failure to thrive.***
- ***Ocular abnormalities:*** Microophthalmia, anophthalmia, cataract, chorioretinitis, or keratoconjunctivitis.
- ***Neurological abnormalities:*** Microcephaly or hydrocephalus, seizure, mental retardation, or intracranial calcification.
- ***Cardiac*** (PDA and pulmonary stenosis)
- ***Hepatosplenomegaly***
- ***Skin rash:*** Vesicular, maculopapular, or petechial hemorrhage, purpura.
- ***Hematological abnormalities:*** Thrombocytopenia, anemia, or neutropenia.
- ***Other organ involvement:*** Pneumonitis, myocarditis, nephritis, hepatitis (hepatomegaly & jaundice) or non- immune hydrops.

**The combination of IUGR with any of these features should prompt investigation for congenital infection.**



# Congenital Toxoplasmosis

**Congenital toxoplasmosis is caused by transplacental acquisition of Toxoplasma gondii.**

- 85% of congenitally infected infants appear normal at birth.
- Maternal infection in first trimester less likely to infect fetus (10%) but if it does the damage is more severe.
- Mid or late trimester infection more likely to infect fetus (30-50%) but the effects are milder.

## Manifestations

- Prematurity, IUGR, jaundice, hepatosplenomegaly, myocarditis, pneumonitis, rash, microcephaly and seizures: ***The classic triad of findings*** consists of ***chorioretinitis, hydrocephalus***, and ***intracranial calcifications.***
- Some infants have a fulminant course with early death, whereas others have long-term neurologic sequelae (which may not develop for several years).

**Pregnant women should avoid contact with cat litter boxes and other areas contaminated with cat feces**

**Congenital toxoplasmosis**
TORCH fact sheets

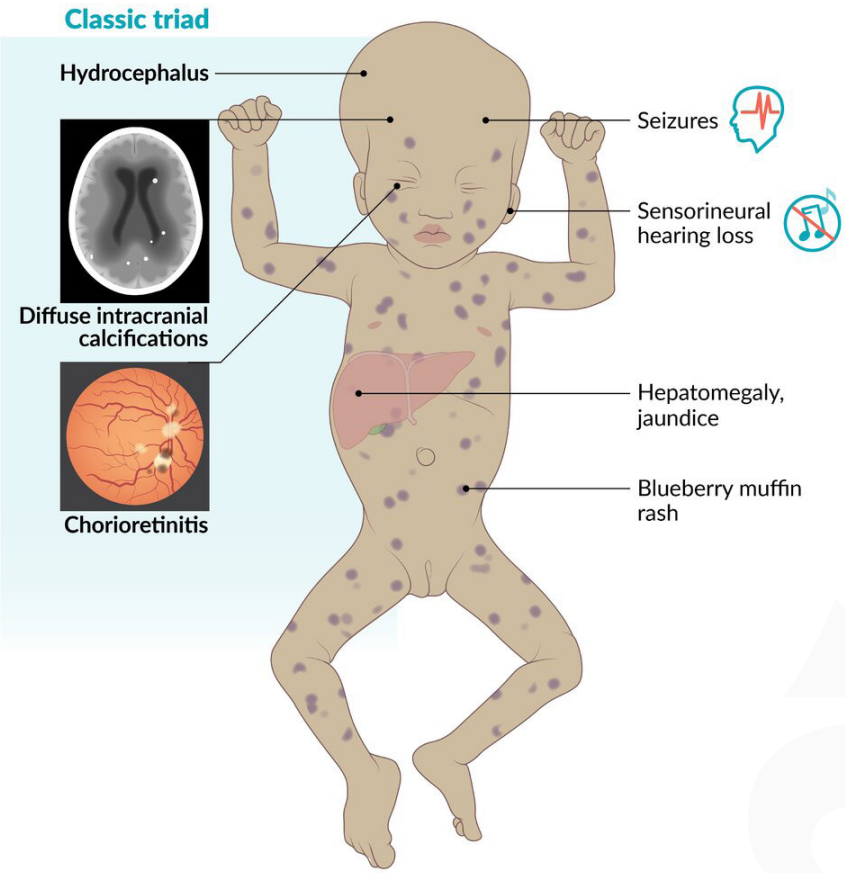
**Pathogen**
*Toxoplasma gondii*

**Transmission**
- Mother: cat feces, undercooked meat
- Fetus: transplacental

**Diagnostics**
Serological testing, PCR of spinal fluid

**Treatment**
Pyrimethamine, sulfadiazine, and folinic acid

**Prevention**
Maternal screening and exposure avoidance



# Fetal Rubella Syndrome

**Rubella is one of the more teratogenic viruses.**

- Ninety percent of infants present with some finding of congenital rubella if infection occurs within the first 12 weeks, and 20% present with congenital disease if the infection occurs between weeks 12 and 16

## Congenital rubella syndrome

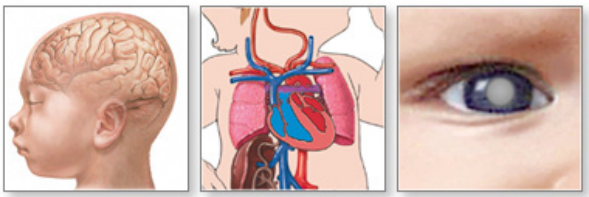
***Characterized by:***

- ***IUGR, intracranial calcifications, microcephaly, cataracts, deafness***
- ***Cardiac defects*** (most commonly patent ductus arteriosus or pulmonary arterial hypoplasia)
- ***Neurologic disease*** (with a broad range of presentations, from behavior disorders to meningoencephalitis)
- ***Osteitis***
- a ***"blueberry muffin" appearance*** caused by purpuric skin lesions
- ***Hepatosplenomegaly.***

***Infection of the fetus is CHRONIC,*** so congenitally infected infants will shed virus at high titre for many months.

## No available treatment.

## Prevention is by vaccination.




# Herpes simplex virus

**Although both HSV-1 and 2 may cause neonatal herpes, HSV-2 is responsible for 70%**

- Ninety percent of neonatal herpetic infections are perinatally transmitted in the birth canal.
- Approximately 10% of infections are acquired transplacentally or via an ascending infection from the cervix.
- Intrauterine infection is associated with IUGR, preterm labor, and miscarriage

***Three distinct syndromes:***

1. ***The most common (45%)*** is localized skin, eye, or mouth disease.
2. ***30% of cases*** manifest as central nervous system (CNS) disease, including meningitis or encephalitis
3. ***25% of cases*** manifest as disseminated disease that involves multiple organs.

***Prevention:*** elective caesarean section for active genital herpes

# Cytomegalovirus (CMV)

**CMV represents the most common congenital viral infection.**

- ***Primary, reactivation, or recurrent CMV infection*** can occur in pregnancy and can lead to congenital CMV infection. Vertical transmission can occur at any stage of pregnancy.
- ***Most (90%) of infected infants are asymptomatic*** but may have ***later deafness or learning disability.***

***Approximately 10% of infants will have one or more sequelae of intrauterine infection:***

- IUGR, sensorineural hearing loss, intracranial calcifications, microcephaly, hydrocephalus, thrombocytopenia, jaundice with hepatosplenomegaly, delayed psychomotor development, chorioretinitis and/or optic atrophy.

***Follow up:*** Long-term serial audiology and developmental assessment, head circumference and ophthalmology.

**Congenital CMV infection**
TORCH fact sheets

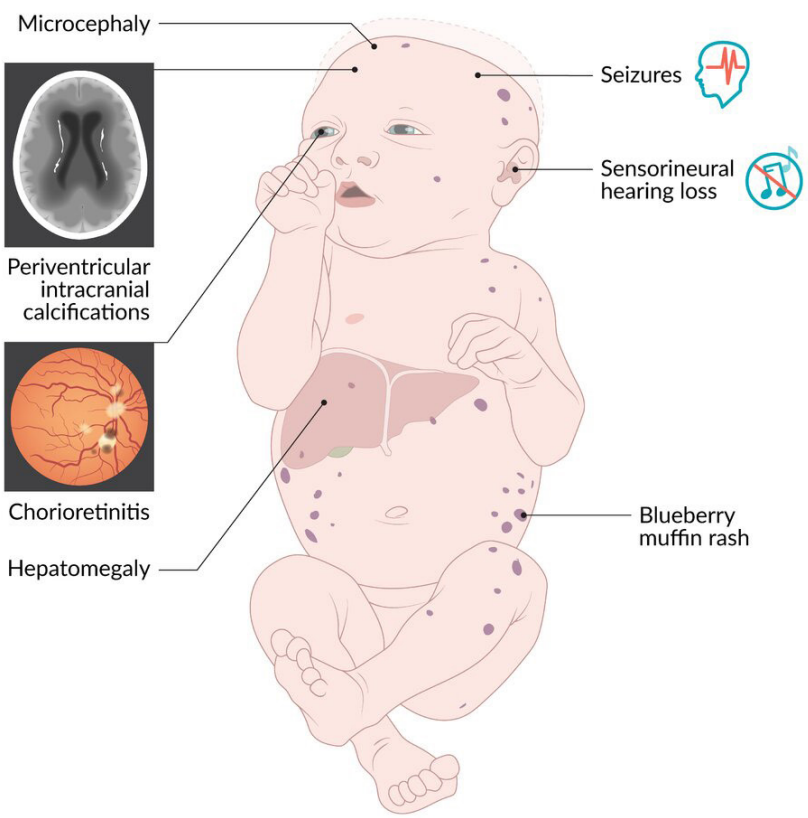
**Pathogen**
Cytomegalovirus

**Transmission**
- Mother: bodily fluids
- Fetus: transplacental

**Diagnostics**
PCR or viral culture of urine or saliva

**Treatment**
Ganciclovir, valganciclovir, or foscarnet

**Prevention**
Maternal avoidance of exposure to bodily fluids, especially with young children (e.g., avoid sharing food, maintain rigorous hand hygiene after changing diapers)




# Parvo virus B19 (B19V)

- It causes **erythema infectiosum (fifth disease).**
- ***The effects of this virus on the fetus*** are much greater and include miscarriage, fetal anemia, hydrops fetalis, myocarditis, and/or intrauterine fetal death.
- B19V infection accounts for 15%-20% of cases of ***non-immune hydrops fetalis.***

## Varicella-zoster virus (VZV)

- **Congenital varicella syndrome (CVS)** results in spontaneous abortion, chorioretinitis, cataracts, limb atrophy, cerebral cortical atrophy, and/or neurological disability.
- Acquisition of infection by the mother in the perinatal period, specifically 5 days prior to delivery or 2 days afterward, poses a risk of severe neonatal varicella, which carries a mortality rate of 30%. (No time for transfer of transplacental maternal IgG antibodies).

# Neonatal infections

## A. Systemic Infections (Neonatal Sepsis)

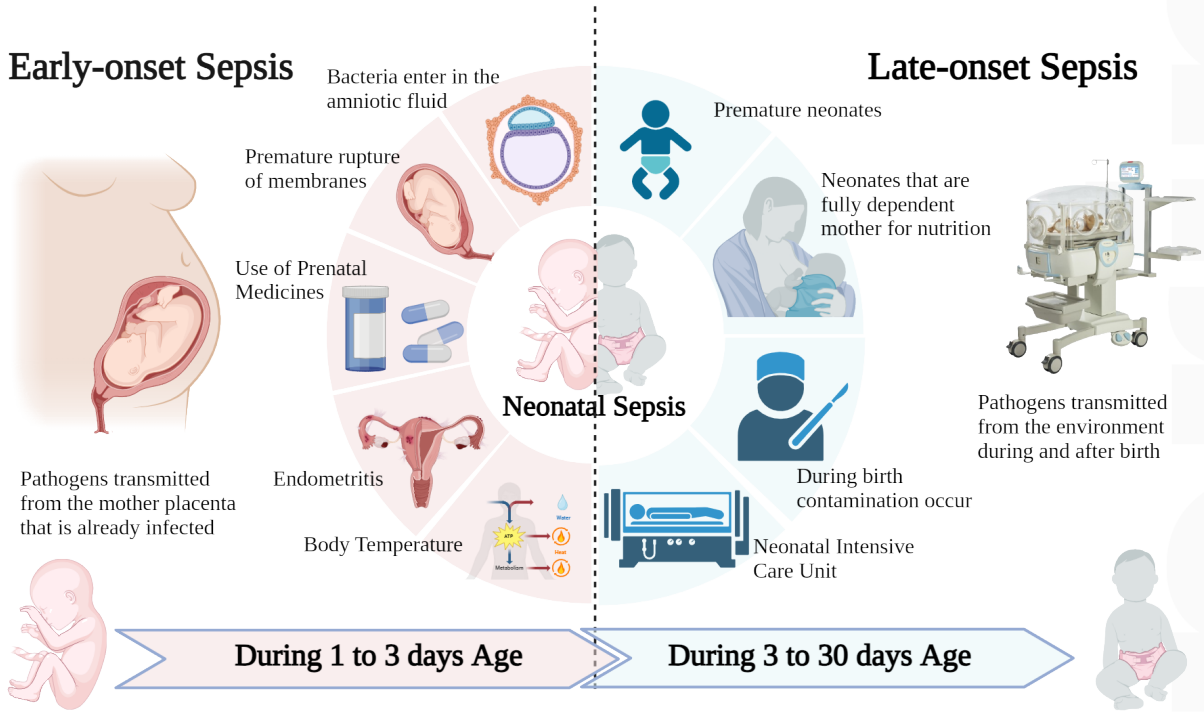
**Signs of neonatal sepsis are generalized and often non-specific to a particular system**

- Apparently trivial infections can rapidly lead to systemic sepsis, especially in the preterm
- ***Serious infections:*** septicaemia, meningitis, pneumonia, UTI
- Neonatal sepsis is associated with ***high mortality & significant morbidity***

### Common pathogens

***Early infection (0-4 days of age):*** ***Group B Streptococci and E coli (60-70%),*** Listeria monocytogenus, gram negative organisms (Klebsiella, Enterococcus), etc

***Late infection (> 4 days of age):*** Staphtlococcus aureus, group B Streptococci, E coli, Klebsiella, Pseudomonas, Staph. epidermidis, H. influenza, etc



## Predisposing factors

***Maternal:*** PROM - Preterm labor - Fever, UTI - Chorioamnionitis

***Neonatal:*** Intrapartum asphyxia - IV catheters - Intrauterine monitoring - Health care acquired infection.

## Symptoms & Signs:

***Presentation may be subtle or non-specific:*** Infant goes off his feed - Has an unexpected rise or fall of temperature - Develops unusual respiratory pattern - Irritability or lethargy - Fits or apneic spells - Poor perfusion (mottling), abdominal distension, jaundice, - Bleeding, petechiae - Seizures - Bulging fontanel is a very late sign of meningitis

***Neonatal meningitis:*** Symptoms sometimes can be subtle so every patient less than 28 days with suspected sepsis should always be screened for meningitis.

***Neonatal Herpes*** can cause CNS infection and early is important for early treatment with acyclovir to decrease mortality.

## Investigations

***A sepsis work-up:***

- ***Complete blood counts*** (White count is very nonspecific and platelets are often low in infection), repeat in 5 hours
- ***Blood and urine cultures.***
- ***CRP***
- Ear, nose and throat ***swabs***, swabs from any obvious sites of infection.
- ***Lumbar puncture*** (should be used quite readily.)
- ***Chest x-ray***
- ***Blood gases, U&Es.***
- ***Associated lab findings:*** hypo or hyperglycemia, hypocalcemia, hyponatremia and DIC.

## Treatment

***Should be tailored to age of onset, clinical setting and initial findings.***

- ***START IMMEDIATELY*** empiric antibiotic combination parentrally, While awaiting results of investigations.
- ***Supportive care*** of different systems are critical

# B. Superficial infections

## 1) Conjunctivitis (Ophthalmia neonatorum) (ON):

- ***Refers to any conjunctivitis occurring in the first 28 days of life***
  - Neisseria gonorrhoeae was the most common cause of infective ON in the past but now accounts for less than 1% in developed countries
  - ***Chlamydia trachomatis*** took over as the most causative agent
  - However both pathogens acquired from birth canal declined over recent decades
  - Non sexually transmitted bacteria such as staphylococcus aureus, streptococcus pneumoniae, pseudomonas, haemophilus species and other gram negative bacteria make up most of cases
  - Viral infections are less common as herpes simplex, adenovirus or enterovirus
  - May also occur as reaction to chemical irritant (silver nitrate), usually mild and self limiting
- **Presentation:** Purulent or mucopurulent or mucoid discharge from one or both eyes within first month of life, may show injected conjunctiva or lid swelling and may be associated with systemic infection (as pneumonitis, rhinitis, otitis in chlamydia)
- **Investigations:** Conjunct. swab & culture may be needed (+ chlamydia culture if suspected)
- **Treatment:**
  - ***Chlamydia:*** erythromycin or azithromycin orally (ophthalmologist)
  - ***Gonorrhoeal:*** hospitalization, ceftriaxone, ttt mother and father
  - ***Other bacterial infections:*** topical antibiotics
  - ***Herpes simplex:*** acyclovir IV, hospitalization, topical antivirus
  - ***Prophylaxis:*** varies according to countries, topical antibiotics

## 2) Septic spots & blisters;

- Are commonly due to ***Staphylococcus infection.***
- **Treatment:** Wash with antiseptic soap containing antibacterial agent. If extensive treat with systemic antibiotics

## 3) Umbilical stump infection (Omphalitis):

- Typically presents as a ***superficial cellulitis*** that can spread to involve the entire abdominal wall and may progress to systemic disease.
- Usually caused by ***streptococci***, staphylococci or gram negative bacteria
- **Standard ttt** is hospitalization, antibiotics

## 4) Oral thrush:

- Infection of the buccal cavity by Candida albicans. Adherent white painful plaques on buccal membrane & tongue. Can cause nipple fissures to the nursing mother,
- **Treatment** by local antifungal.

## 5) Candidal diaper dermatitis:

- Rash with red areas around anus and in the groin with characteristic satellite papules and involvement of intertriginous folds, **treatment** by topical antifungal ointment

# 9 NEONATAL EMERGENCIES

- Neonatal emergencies are not uncommon problems.
- They appear either at the time of birth, during the in-hospital post-birth period, or at home within several weeks of discharge.
- In all instances they present significant diagnostic and treatment challenges to the clinician, and must be taken seriously.

## Recognizing critically-ill neonates

- Poor perfusion / Shock – Respiratory distress / acidotic breathing / apnea – Cyanosis – Disturbed level of consciousness – Seizures – Bleeding – Trauma – Surgical Abdomen

### "THE MISFITS"

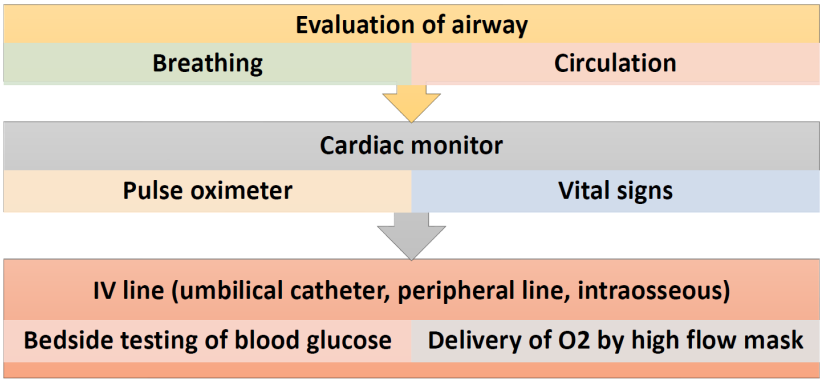
**This mnemonic gathers the most important disorders that can present as an emergency during neonatal period, and each of them presents clinically with one or more of the above mentioned symptoms and signs:**

- ***T***rauma - ***H***eart - ***E***ndocrine
- ***M***etabolic - ***I***nborn errors (metabolic emergencies) - ***S***epsis - ***F***ormula mishaps - ***I***ntestinal catastrophes - ***T***oxins - ***S***eizures

## Acute life threatening event

- ***ALTE is defined as*** "an episode that is characterized by some combination of ***apnea*** (centrally or occasionally obstructive), ***color change*** (usually cyanotic or pallid, but occasionally erythematous or plethoric), ***marked change in muscle tone*** (usually marked limpness), ***choking,*** or ***gagging.***"

# Systematic approach



# Recognition and Approach of the Critically Ill Neonate

## Clinical history

- Prenatal (PROM, Maternal fever)
- Natal (Apgar score, need for resuscitation)
- Postnatal ( feeding and weight gain, urination and bowel movement)

## Physical examination

- Oxygen saturation and vital signs
- Infant's appearance, work of breathing, Circulation

## Laboratory Investigations

- Electrolytes, serum glucose, and calcium level.
- Complete blood cell count (CBC), CRP, Blood culture.
- Pro-thrombin time (PT); partial thromboplastin time (PTT).

**Note that:**

- Hypoglycemia in neonates is defined when blood glucose is below 40 mg/dl.
- Saline boluses may be given. Start with 5-10 ml/kg then check on vital and clinical signs of perfusion.
- Laboratory tests should be ordered once there is IV access. Basic tests are withdrawn and other tests may be considered depending on differential diagnosis.

# Trauma

- Any neonate ***presenting with apparent life-threatening event (ALTE)*** and ***no obvious cause*** should be ***evaluated for head trauma*** either accidental or non-accidental.
- The baby should then be ***stabilized (ABCD),*** with proper ***monitoring*** of temperature and blood sugar.
- ***Laboratory investigations*** must include a bleeding screen and imaging should include CT brain and skeletal survey. ***Ophthalmologic examination*** is also needed.

# Heart

***Cardiac emergencies in neonates include:***

## 1) Duct-dependent pulmonary circulation

***These are congenital heart defects with obstruction to pulmonary blood flow; for instance:***

- Pulmonary atresia with or without Fallot tetralogy, Tricuspid atresia, Critical pulmonary stenosis, and TGA with intact interventricular septum.

### 2) Duct-dependent systemic circulation

***These are congenital heart defects with obstruction to systemic circulation; for instance:***

- Coarctation of the aorta, Interrupted aortic arch, Hypoplastic left heart syndrome, and Critical aortic stenosis.

### 3) Congestive heart failure

- Congenital heart diseases → PDA, Truncus arteriosus, Total anomalous pulmonary venous connection.
- Severe anemia
- Sepsis
- Metabolic derangements
- Thyrotoxicosis / Arrhythmias

### 4) Arrhythmias

- Severe uncontrolled Sinus or Supraventricular tachycardia (SVT)
- Bradycardia as in complete heart block in infants of lupus mothers.

### 5) Arteriovenous malformations.

### Management

***If suspecting duct-dependent heart defects:*** start prostaglandin infusion

***If congestive heart failure:*** fluid restriction, diuretics, inotropes, supportive ventilation, and correction of anemia.

***if SVT:*** vagal maneuvers, pharmacologic treatment, and consultation of pediatric cardiology

# Neonatal shock

**Circulatory insufficiency leading to tissue hypoperfusion**

- The hypoperfusion will be initially compensated for by tachycardia and vasoconstriction (compensated shock), then compensatory mechanisms fail leading to uncompensated shock and eventually tissue damage.

### Causes of neonatal shock:

***Circulatory insufficiency could be caused by:***

1. ***Hypovolemia***
2. ***Heart failure (cardiogenic shock)***
3. ***Distributive shock*** due to vasodilation and pooling of blood in peripheral vessels
4. ***Obstructive shock*** as seen in tension pneumothorax or persistent pulmonary hypertension of the newborn.

# Endocrine

***This group of neonatal emergencies includes:***

### 1) Congenital adrenal hyperplasia (CAH):

- An Autosomal Recessive disorder caused by defective synthesis of cortisol by the adrenal gland leading to overproduction of ACTH and stimulation of adrenals.
- The most common enzyme deficiency in this disorder is ***21-hydroxylase deficiency.*** It presents clinically with ***vomiting, diarrhoea, hypoglycemia, hypotension, and shock.***
- Electrolyte disturbance in the form of hyponatremia and hyperkalemia are characteristic.
- ***Blood gas analysis*** reveals metabolic acidosis that should be corrected along with the hyponatremic dehydration. Supplementation with mineralocorticoids and glucocorticoids is life-saving.

### 2) Neonatal thyrotoxicosis

- This causes high ***output heart failure and tachyarrhythmias.***
- It is usually transient in infants born to mothers with Grave's disease, who receive TSH-receptor stimulating antibodies.
- ***Antithyroid drugs*** are usually needed together with ***Propranolol*** (only used if neonate is not in failure).

### 3) Growth hormone deficiency

- This disorder presents by ***hypoglycemia that persists beyond the first 4 to 5 days of life*** and needs ***immediate correction of blood glucose levels.***

### 4) Syndrome of inappropriate antidiuretic hormone secretion (SIADH)

- Excessive uncontrolled secretion of antidiuretic hormone leading to water retention in the body ***(water intoxication).***
- It occurs in central nervous system disorders like infections, sinus thrombosis, or hypoxic ischemic encephalopathy, and sometimes in pneumonia.
- ***Laboratory abnormalities*** include elevated urine osmolality, decreased plasma osmolality, hyponatremia, and hypouricemia.
- ***Treatment*** essentially is directed to correction of hyponatremia, water restriction, and sometimes loop diuretics are used.

# Neonatal sepsis (see before)

# Formula over dilution or over concentration

**It results in hypo/hypernatremia**

## Metabolic derangements

***Hyperkalemia***

- This results from acute tubular necrosis (as in hypoxia ischemia), excessive hemolysis, old blood transfusion, sepsis, and extreme prematurity.
- It must be ***treated immediately*** as it can cause fatal ***arrhythmia.***
  - Emergency treatment includes calcium gluconate injection (cardio-protective), β2-agonist nebulization, NaHCO3 infusion, diuretics, or dialysis. Glucose-Insulin infusion can be used as well.

***Hypokalemia*** can also cause ***arrhythmias*** and is treated by infusing the calculated ***K deficit.***

***Hyper/hyponatremia*** should be corrected ***very gradually.***

***Hyper/hypocalcemia*** should be corrected to ***avoid resultant arrhythmias as well.***

## Inborn Errors of Metabolism

***These disorders are caused by*** inherited enzymatic deficiency or dysfunction in certain metabolic pathways

***They can present as*** an emergency during neonatal period through causing intoxication and/or energy failure, and some of these disorders cause dysmorphism or organ dysfunction.

***Their symptoms are*** mostly ***non-specific*** such as poor feeding, failure to thrive, vomiting, irritability, encephalopathy, apnea, lethargy, seizures, metabolic acidosis, or hypoglycemia.

***An example of intoxication disorders*** are the organic acidemias and urea cycle defects causing hyperammonemia. Hypoglycemia occurs in glycogen storage disorders and fatty acid oxidation defects.

***Emergency treatment of hyperammonemia*** includes sodium benzoate, protein restriction, and sometimes dialysis is needed.

## Toxins/Poisons

- Toxic ingestions are uncommon. Occasionally the result of a maternal ingestion in a breastfeeding mother, homeopathic remedies, or overuse of accepted medications.

# Intestinal catastrophes

## a- Neonatal intestinal obstruction

***The most common clinical findings are;*** vomiting (Bilious), abdominal distension and sometimes failure to pass meconium (depending on the level of the obstruction)

***Plain abdominal radiograph*** shows dilated air-filled loops proximal to the obstruction and no air distal to it. (Normal neonates show rectosigmoid air in a plain abdominal radiograph after 12-24 hours.)

***Neonatal intestinal obstruction can be classified into:***

- ***High (upper) obstruction:*** Proximal to mid ileum (involve stomach, duodenum, jejunum and proximal ileum). Plain radiographs reveal one, two, or a few dilated air-filled bowel loops
- ***Low obstruction:*** Involve distal ileum or colon. Radiographs show multiple dilated air-filled bowel loops. These neonates need a contrast enema.

### Examples of high intestinal obstruction in neonates

***Duodenal obstruction:***

- Major associated anomalies are present in about 50 % of the patients.
- Approximately 30 % have Down syndrome. Annular pancreas may cause duodenal obstruction.
- Plain Xray shows double bubble sign.

***Malrotation and Midgut Volvulus:*** The sudden onset of bilious emesis in a neonate who has been normal for the first few days of life should be considered to be due to a midgut volvulus until proven otherwise.

### Examples of low intestinal obstruction in neonates

***Ileal atresia*** is an important cause for low intestinal obstruction, representing approximately 50 % of small bowel atresias.

***Hirschsprung disease*** is a form of low intestinal obstruction caused by the absence of normal myenteric ganglion cells in the submucosal and intermuscular myenteric plexuses of a segment of the colon.

## b- Necrotizing Enterocolitis (discussed previously)

## c- Pneumoperitoneum

- Is usually the result of a hollow viscus perforation e.g. NEC, Hirschsprung disease.
- It may be iatrogenic e.g. by rectal thermometer, enema or mechanical ventilation.

## d- Incarcerated inguinal hernia

- When the contents of inguinal herniae get blocked at the neck and cannot easily be reduced.

# Neonatal Seizures

## Etiology:

1. Hypoxic ischemic encephalopathy.
2. Infarctions and hemorrhage
3. Brain malformation.
4. Infections (meningitis, encephalitis, intrauterine infection).
5. Metabolic disorders (hypoglycemia, hypocalcemia).
6. Inborn error of metabolism as pyridoxine dependency and nonketotic hyperglycinemia.
7. Genetic causes.

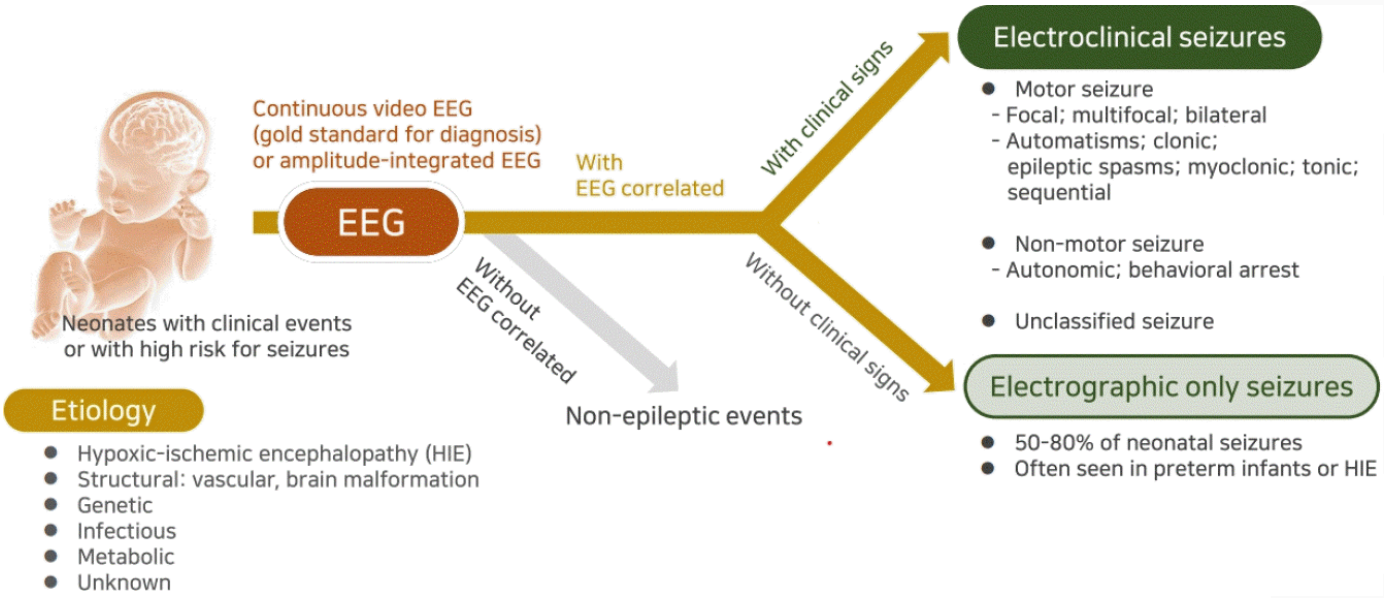
## Diagnosis:

***Laboratory investigations*** as serum glucose, calcium, magnesium and sodium. CBC and CRP.

***Brain imaging*** using cranial ultrasound, CT scan or MRI. CSF analysis. EEG.

## Management:

- ***First line*** is ***benzodiazepines***; ***second line*** is ***phenobarbital*** and ***third line*** is ***phenytoin***.

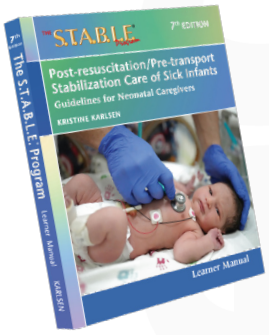


# Newborn Transport

**Medical transport of sick newborns requires skilled medical personal and specialized equipment.**

- Transport of critically ill and high-risk newborns to a ***tertiary care neonatal center*** occurs through ground or air transport.
- Transported newborns are called ***"out-born"*** neonates while those who were delivered in the same tertiary center are called ***"in-born".***
- The ***outcome*** of an ***out-born*** neonate with major medical or surgical problems remains ***worse than for an inborn infant.*** These emphases the importance of prenatal diagnosis and in-utero (maternal) transfer if possible.

### *Stabilization of Sick Newborns Before and during Transport:*




- We use the mnemonic ***S.T.A.B.L.E.*** to optimize learning, retention and recall of information.
- This is based on STABLE program, which is a post-resuscitation/pre-transport of sick neonates.
- ***S.T.A.B.L.E.*** stands for the six assessments of: ***S***ugar and Safe care, ***T***emperature, ***A***irway, ***B***lood pressure, ***L***ab work, and ***E***motional support.

# Sugar and Safe Care

**Transient hypoglycemia is common in healthy term infants after birth due to changes from continuous transplacental supply of glucose from the mother to an intermittent supply from feeds.**

### *Causes of hypoglycemia in newborns:* see before

### *Sugar Stabilization:*

- ***For at-risk neonates without a suspected or confirmed genetic hypoglycemia disorder,*** the goal is to maintain plasma glucose >50 mg/dL in the first 48 hours of life, and >60 mg/dL after 48 hours of life.
- ***In neonates with a suspected or confirmed genetic hypoglycemia disorder,*** the goal is to maintain plasma glucose >70 mg/dL.

### *Symptoms, Signs and Treatment of neonatal hypoglycemia:* see before

# Temperature Stabilization

**Neonatal hypothermia increases oxygen consumption and metabolic demands, which can impair resuscitative efforts.**

- Maintaining temperature control is important to avoid the detrimental effects of hypothermia.
- Heat loss occurs by conduction, convection, radiation, and evaporation.
- Newborns with risk of hypothermia are preterm, small for gestational age, infants with open skin defects (eg; gastroschisis).
- Preterm infants have more risk to develop hypothermia because of their large body surface area relative to their mass, thin skin, and decreased subcutaneous fat.

***Temperature should be maintained 36.5 - 37.5 C.***

- To prevent hypothermia in ***preterm infants***, it is recommended to use ***polyurethane bags or wraps in infants with birth weights less than 1500 grams.***
- For infants who require ***respiratory support***, ***humidified and heated air*** decreases hypothermia.

## Therapeutic Hypothermia for HIE

- Therapeutic hypothermia is the only proven neonatal intervention for neonatal encephalopathy that reduces the risk of cerebral palsy.
- For newborns who meet the criteria for therapeutic hypothermia, stabilization by support of respiratory and cardiovascular systems is done first. Then cooling is done by using therapeutic head cooling or whole-body cooling for 72 hours at 33 to 35C started in the first 6 hours after delivery.
- Low- tech therapeutic hypothermia is done with cooling-bags, packs might be beneficial for treatment of neonatal hypoxic ischemic encephalopathy in low- or middle-income countries.
- Therapeutic hypothermia started within one hour of post-natal life leads to reduction in seizures in newborns with neonatal encephalopathy

# Airway Stabilization

**Most critically ill neonates who require transport to tertiary neonatal intensive care have respiratory distress or failure.**

***To determine the severity of respiratory distress we have to evaluate:***

- Respiratory rate
- Work of breathing
- Presence of cyanosis
- Oxygen saturation
- Oxygen requirement
- Blood gas
- Chest x ray
- Neurologic status

# Blood Pressure Stabilization

**It is important to diagnose and manage shock in the newborn before and during transportation.**

- Shock is defined as physiologic state characterized by ***tissue hypoxia*** due to reduced oxygen delivery and/or increased oxygen consumption or inadequate oxygen utilization.
- ***Physical findings*** are those of decreased tissue perfusion (eg., cold extremities, acrocyanosis, and poor capillary refill), hypotension, and metabolic acidosis.
- Shock is usually ***reversible initially*** but if not treated ***progresses to irreversible*** organ dysfunction.

***Causes of Neonatal Shock:*** see before

***Stabilization of hemodynamic status in the newborn:***

- Stabilization of the airway
- Vascular access
- Initial fluid resuscitation
- Administration of empiric antibiotics

***Initial diagnostic evaluation:*** Basic laboratory tests.

# Laboratory work for stabilization of sick newborn

***The "4 Bs" lab tests should be obtained before transport of the sick newborn if possible:***

1. Blood count (complete blood count, CBC) with white blood cell, WBC differential and platelet count.
2. Blood culture; using sterile technique.
3. Blood glucose
4. Blood gas

***Maternal risk factors for neonatal sepsis:*** see before

***Clinical manifestations of sepsis: are nonspecific:***see before

***Neutrophil indices used to determine the possibility of infection:***

- ***I/T ratio***: immature to mature neutrophil ratio. A normal I/T ratio can help rule out sepsis.
- ***ANC;*** Absolute neutrophilic count.

# Emotional support

## Stabilization of Parents Emotions before and after newborn transport

- Try to answer parent's questions about their baby. Keep explanations simple.
- Provide illustrations and written materials if possible.
- If the baby has been given a name, call the infant by his name. Use the correct gender when referring to the infant.

### *What SHOULD you send with the newborn?*

- Complete ***maternal records*** including prenatal and labor data.
- Complete ***neonatal records*** including physician orders and notes, laboratory results and radiographs or other diagnostic tests.

# 10 NEONATAL SCREENING

**Screening of all newborns for an occult disease or the potential for disease**

**It is the process of testing newborn babies for some serious, but treatable, conditions**

***The goal*** is to detect disorders that are threatening to life or long-term health before they are symptomatic.

Early treatment of these disorders may significantly reduce mortality and morbidity in affected patients.

***Time:*** 3rd – 6th days.

***Decisions about what conditions to be included in newborn screening are based on:***

1. Incidence and severity of the condition within the targeted population.
2. Degree to which detection in the newborn period leads to better outcomes than usual routine clinical care.
3. Availability of an accurate screening test.
4. Resources for screening, diagnosis, and treatment.

***Screening test characteristics:***

1. The disease should be serious and common, so that lack of treatment would result in significant morbidity or death.
2. Effective treatment should be readily accessible.
3. Early detection of the disease in the presymptomatic or early symptomatic stage should lead to better outcomes (treatment and prognosis).
4. Has low false-negative rate.
5. Must be simple and in-expensive.
6. The results must be available on a timely base so intervention would be possible.
7. The neonate should be normal at birth
8. Follow-up mechanisms must be available.

**The screening tests must be easy to perform and interpret, and have acceptably, low cost, with high sensitivity and specificity.**

## Process of screening tests

***During birth Hospitalization:*** Screening for hearing loss and for critical congenital heart disease.

***Blood spot specimen:*** Should be obtained as close as possible to hospital discharge.

**In Egypt blood spot specimen is done on the fifth day after birth.**

## Screening the newborn for hearing loss

**Significant hearing loss occurs in 1 to 3 newborns per 1000 live births. Infants cared for at neonatal intensive care units [NICUs] are at-risk for sensorineural hearing loss (SNHL) and auditory neuropathy (AN).**

- Newborn screening detects hearing loss at an early age by ***universal neonatal hearing screen (UNHS)*** results in ***early intervention*** (eg, use of hearing devices and access to language [speech or sign]). ***Earlier detection and intervention*** (before 10 months of age) improves ***language and reading development.***
- Two electrophysiologic techniques, automated auditory brainstem responses (AABR) and otoacoustic emissions (OAE), are routinely used as screening tests. Both tests are portable, automated, and inexpensive, making them well suited to newborn screening. However, OAE does not detect AN and AABR should be used to screen infants who are at risk for AN.
- The two types of UNHS include one-stage, which utilizes a single screening test (ie, AABR or OAE), or two-stage, which utilizes two screening tests or repeats the same test. In the two-stage UNHS, only patients who fail the initial test receive a second screening study, and only patients who fail both tests are referred for audiologic assessment.
- Although UNHS has improved the early recognition of hearing loss, there are still infants with hearing loss who are not detected by UNHS. Therefore, a normal hearing screen should not be completely reassuring.
- Infants who ***pass*** the neonatal screening ***but have a risk factor*** for hearing loss should have ***audiologic assessment*** performed by a pediatric audiologist.
- ***Monitoring of infants for hearing loss should continue*** during routine primary care visits, particularly in those who are at risk.

# Screening For Critical Congenital Heart Disease

## Using Pulse Oximetry

- Congenital heart disease (CHD) is the ***most common congenital disorder in newborns.***
- Critical CHD, defined as requiring surgery or catheter-based intervention in the first year of life, accounts for approximately 25 percent of all CHD.
- In infants with critical cardiac lesions, the risk of morbidity and mortality increases when there is a delay in diagnosis and timely referral to a tertiary center with expertise in treating these patients.
- The goal of critical CHD screening in newborns is to reduce mortality and morbidity associated with delayed diagnosis by identifying newborns with critical CHD in a timely manner.
- There is evidence that universal newborn pulse oximetry screening (POS) improves the identification of patients with critical CHD compared with physical examination alone and may lead to decreased infant morbidity and mortality from critical CHD.
- Screening is performed at >24 hours after birth or as late as possible if early discharge is planned. Oxygen saturation (SpO2) should be measured in the right hand (preductal) and either foot (postductal).

### *Criteria for a positive screen:*

- SpO2 <90 percent in either extremity
- SpO2 90 to 94 percent in both the right hand and a lower extremity on two to three measurements, each separated by one hour
- SpO2 difference ≥4 percent between the upper and lower extremities on two to three measurements, each separated by one hour

### *Assessment of newborns with positive screens:*

- A neonate with hypoxemia should not be discharged from the hospital without ***excluding*** potentially life-threatening conditions. Newborns with positive screening results should ***undergo evaluation*** to identify the cause of hypoxemia.
  - ***If a noncardiac cause*** of the hypoxemia cannot be identified, then evaluation of critical CHD as the cause should include high-quality echocardiography, with interpretation by a clinician with expertise in the diagnosis of CHD.
  - ***In newborns in whom an alternative cause*** (other than critical CHD) is identified and treated, an echocardiogram may not be needed if the hypoxemia resolves.
  - ***If critical CHD is identified*** on echocardiography, urgent consultation with a pediatric cardiologist and/or transfer to a medical facility with pediatric cardiology expertise is warranted. Newborns with ductal-dependent lesions are at increased risk for death and significant morbidity unless interventions are initiated to maintain patency of the ductus arteriosus, ensure adequate mixing of deoxygenated and oxygenated blood, and/or relieve obstructed blood flow.

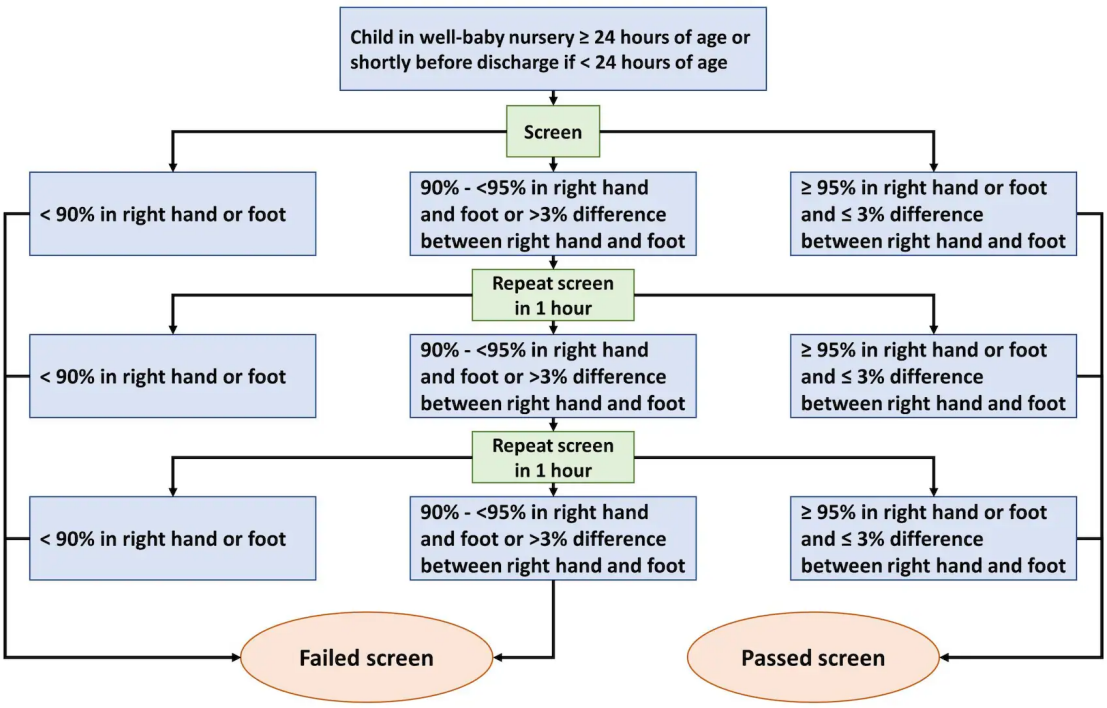
### *Common noncardiac causes of hypoxemia identified through newborn POS include:*

- Sepsis
- Pneumonia
- Transient tachypnea of the newborn (TTN)
- Respiratory distress syndrome (RDS)
- Persistent pulmonary hypertension of the newborn (PPHN)
- Meconium aspiration syndrome (MAS)
- Pneumothorax

### *Targeted lesions by pulse oximetry screening (POS)*

- ***They are defects that typically:***
  (A) require intervention in the first year of life,
  (B) present with hypoxemia some or most of the time.
- ***These include but are not limited to the following defects:***
  - Hypoplastic left heart syndrome
  - Tetralogy of Fallot
  - Total anomalous pulmonary venous connection
  - Transposition of the great arteries
  - Tricuspid atresia
  - Truncus arteriosus
  - Coarctation of the aorta

**While POS improves early identification of infants with these defects, some affected newborns (particularly those with coarctation of the aorta) may pass the POS.**



# Blood spot specimens for newborn screening

**Blood samples are collected on filter paper from puncture of the newborn's warmed heel, which are sent to a central laboratory for analysis.**

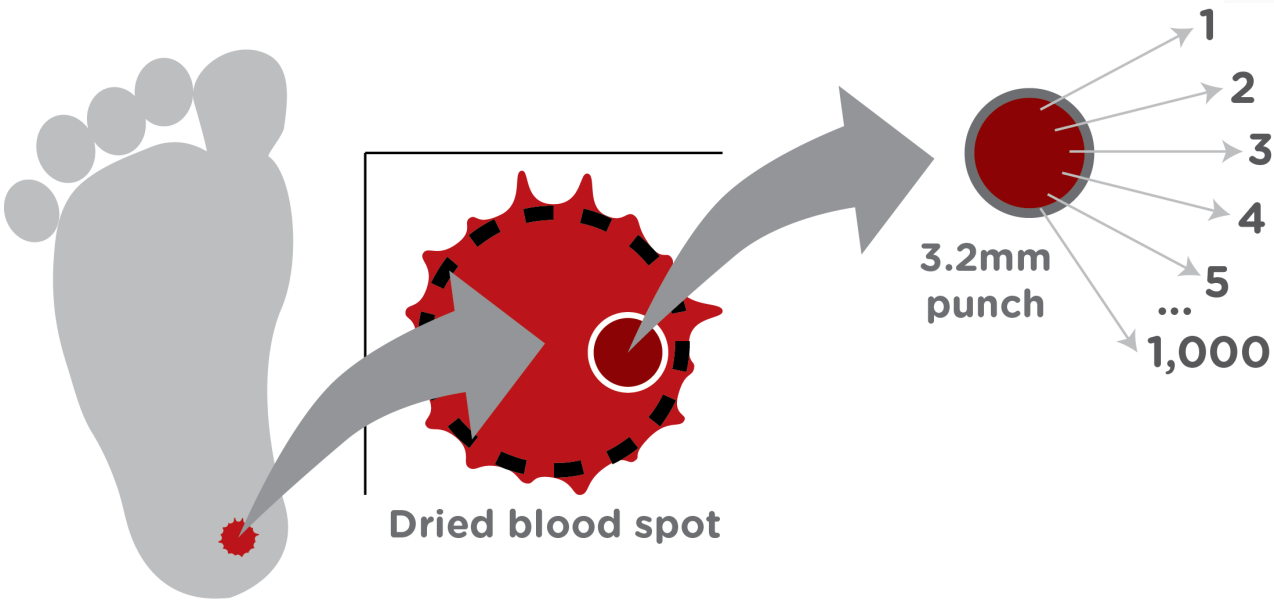
- Positive results from newborn screening should have a ***confirmatory test*** performed as quickly as possible.

***When the result is positive:*** It is possible that the child has a treatable ***genetic condition***, but it is much more likely that the baby is healthy and the test result is a ***"false positive".***

***In Developed countries the most important disorders screened for are:***

- Phenylketonuria, primary congenital hypothyroidism, cystic fibrosis, the galactosemias, medium chain acyl-CoA dehydrogenase deficiency, glutaryl-CoA dehydrogenase deficiency and congenital adrenal hyperplasia, together with several other disorders detectable by tandem mass spectrometry.

***In Egypt*** we used to screen for congenital hypothyroidism and phenylketonuria. But recently the Egyptian National Neonatal Screening Program screens for ***19 diseases.***




# Other screenings

## A) Clinical neonatal screening

1. CHD.
2. Imperforate anus
3. Cong. hip dislocation.
4. Esophageal atresia & TOF

## B) Neonatal screening for endocrine disorders

***Cong. Hypothyroidism.***

***Cong. adrenal hyperplesia.***

## C) Neonatal screening for inborn errors of metabolism

- ***Phenyl ketonuria:*** Irreversible MR
- ***Galactosemia:*** Cataract – Jaundice – FTT – Vomiting
- ***MSUD:*** Convulsions – Acidosis – Urine odour
- ***Homocystinuria:*** Marfanoid appearance
- ***Heriditary Fructose intolerance:*** Acute hypoglycemia, hepatic & renal dysfunction

## D) Neonatal screening for blood diseases

- ***Hemoglobinopathies:*** Sickle cell – Hb F – Hb e.
- ***G6PD deficiency.***

## E) Other disorders considered for neonatal screening

- ***Cystic fibrosis.***
- ***$\alpha_1$ antitrypsin deficiency***
- ***Duchenne's muscular dystrophy.***
- ***Neuroblastoma.***
- ***Familial ureteric reflux.***
- ***Liver diseases & biliary atresia.***
- ***Neonatal hearing screening.***
- ***Screening for congenital infections.***

# Questions

**1. Which one of the following is the most important cause of jaundice presenting in the first 24h of life?**

A. Prematurity.
B. Haemolysis
C. Breastfeeding.
D. Physiological jaundice
E. Early onset sepsis

**2. Which one of the following is the major contributor to the development of physiological jaundice?**

A. Breastfeeding.
B. Decreased hepatic bilirubin excretion.
C. Immature hepatic enzymes.
D. Enterohepatic circulation.
E. Increased bilirubin production

**3. Which of the following is most predictive for the development of kernicterus?**

A. Hyperbilirubinemia within the first 24 hours of life
B. Hemoglobin level immediately after birth
C. Peak conjugated bilirubin level
D. Peak unconjugated bilirubin level
E. Duration of Hyperbilirubinemia

**4. All of the following statement are true about congenital rubella except?**

A. It is diagnosed when the infant has IgM antibodies at birth.
B. It is diagnosed when the infant has IgG antibodies persist for more than 6 months.
C. Most common congenital defects are deafness, cardiac malformation and cataract.
D. Infection after 16 weeks of gestation results in major congenital defects.
E. Microcephaly , seizure and mental retardation can OCCUR.

**5. One statement is incorrect concerning oral moniliasis:**

A. Fungal infection caused by Candida albicans
B. Infection is usually acquired during delivery
C. May cause painful stomatitis with difficult suckling
D. Removal of plaques leaves punctate areas of bleeding
E. Treated by broad spectrum antibiotics

E

**6. Microcephaly may be caused by any of the following except:**

A. Chromosomal abnormality
B. Hypoxic ischemic brain injury
C. Bilirubin encephalopathy
D. Congenital aqueductal stenosis
E. Congenital infection

D

**7. As regards neonatal reflexes:**

A. Normally persist in childhood
B. Should be present at birth
C. Gradually exaggerates as they grow up
D. Their absence at birth indicates CNS maturity
E. Must disappear by the end of the first month

B

**8. Modes of treatment of neonatal jaundice include all the following EXCEPT::**

A. Light bulbs at home
B. Phototherapy
C. Exchange transfusion
D. IVIG
E. Adequate feeding

A

**9. Physiological jaundice is characterized by:**

A. Starts during the first 24 hours of life
B. Rises by more than 5 mg/dl/hour
C. Clinically undetectable after 14 days of life
D. Associated with clay colored stool
E. Direct bilirubin level is more than 2 mg/dl

C

**10. Physiological anemia in neonatal period is characterized by all the following, EXCEPT:**

A. Nadir is reached at 6-8 weeks
B. Progresses gradually till it reaches the nadir
C. In preterm it's milder than in full terms
D. It's due to shorter life span of fetal RBCS
E. Due to abrupt cessation of erythropoietin at birth

C

**11. As regards TTN:**

A. Defined as cessation of respiration in a newborn
B. It's a common problem in preterms
C. CBC and CRP show positive evidence of sepsis
D. Usually follows cesarean section
E. Improves within 2 weeks

D

**12. A healthy newborn baby boy may have all the following EXCEPT:**

A. Have erythema of the umbilical skin extending to the abdomen
B. Produce breast secretion
C. Have a single palmar crease
D. Open anterior fontanelle
E. Bluish discoloration on lower back (sacral vertebrae)

A

**13. The neonatal examination should be carried:**

A. In a warm room
B. In a dark room
C. In a cool room
D. One day after delivery
E. Only for preterm baby

A

**14. Which of the following does not increase the risk ofdeveloping NEC?**

A. Very low birth weight
B. Breastfeeding
C. RDS
D. Premature birth
E. Formula feeding

B

**15. A 28 hour old newborn has an abdominal x-ray due to delayed passage of meconium. A double bubble sign without distal bowel gas is seen. What is the most likely diagnosis?**

A. Oesophageal atresia
B. Anal atresia
C. Duodenal Atresia
D. Cystic fibrosis

D

**16. If the protruding organs have a protective membrane covering them, which diagnosis is more likely?**

A. Omphalocele
B. Cloacal exstrophy
C. Congenital diaphragmatic hernia
D. Gastroschisis

A

**17. How is a preterm birth defined?**

A. Before 28 weeks
B. Before 32 weeks
C. Before 35 weeks
D. Before 37 weeks
E. Before 38 weeks

D

**18. Extremely low birth weight (ELBW), define as birth weight less than ..**

A. 2500 gm
B. 2000 gm
C. 1500 gm
D. 1000 gm

D

***History***

Ahmed is a 4-day-old baby who is referred by his midwife to the pediatric day unit because of jaundice. His mother thinks that the jaundice may have commenced within the first 24 hours of life, but she was told by the first midwife that she saw that the baby was fine. The child has also been a little sleepy and has not breast-fed as well as previously. His birth weight was 3.70kg at term. His mother had a splenectomy after falling off a horse as a teenager. This is her first pregnancy. There is no family history of jaundice.

***Examination***

The sclera are markedly yellow and the infant is somewhat lethargic. He is well perfused and apyrexial. There is no hepatosplenomegaly and there are no other signs. He weighs 3.40 kg.

***Investigations***

CBC: HB 12.1 g/dL, WBC 27.7 * 10 ^ 9 / L Platelets 361 * 10 ^ 9 / L

Reticulocytes Blood film 12 %, Occasional spherocytes

Na 143, K 5, Urea 37.2, Creatinine 1.43

Bilirubin 32, Albumin 3,

ALT 48, ALP 347 (N), CRP 5

Baby's blood group A+

Maternal blood group 0+

Direct antiglobulin test (DAT) Positive

(G6PD) N

Urine dipstick No leucocytes or nitrites

## 1) What are the causes of jaundice in a neonate?

***Early onset (first 24 hours, hemolytic jaundice)***

- Rhesus hemolytic disease
- ABO incompatibility
- G6PD deficiency (commonest in those of African, Asian, or Mediterranean Descent)
- Hereditary spherocytosis

***Normal onset***

- Physiological (all newborns get a degree of jaundice peaking at 4-5 days)
- Bruising Polycythemia
- Causes of early jaundice

***Late onset (>14 days, prolonged jaundice)***

- Persistence of a pathological earlier jaundice
- Breast milk jaundice
- Neonatal hepatitis
- Biliary atresia
- Hypothyroidism
- Glactosemia

***Jaundice can also be a non-specific marker of neonatal infection at any stage.***

## 2) Why is neonatal jaundice potentially dangerous?

Neonatal jaundice can be dangerous, as unconjugated bilirubin can cross the blood-brain barrier. Very high levels can lead to kernicterus which can cause deafness and choreo- athetoid cerebral palsy.

## 3) What is the cause in this infant?

The cause of the jaundice in this infant is ABO incompatibility.

His mother is 0+ and will therefore have anti-A and anti-B antibodies in her blood.

This is further confirmed by the positive DAT (though, on occasion, this test can be negative).

The decreased hemoglobin and raised reticulocyte count provide further evidence of hemolysis.

The presence of a few spherocytes is common in ABO incompatibility.

The normal temperature, normal inflammatory markers and normal urine dipstick make infection unlikely.

Unlike Rhesus hemolytic disease, it can occur with the first pregnancy and does not get worse with successive pregnancies.

## 4) What is the treatment?

This infant's bilirubin level is well above the exchange transfusion line.

Phototherapy should be commenced immediately and blood taken for cross-match (for O+ blood to minimize further hemolysis).

The fall in birth weight of 8 per cent and the raised urea and creatinine suggest a degree of dehydration which could be exacerbating the jaundice.

Maintenance intravenous fluids should therefore be commenced and breast-feeding can be continued.

Appropriate lines should be inserted to enable the exchange transfusion to take place.

The blood for the transfusion usually has to be ordered from the regional blood transfusion center.

It is possible that the bilirubin will have fallen markedly by the time the blood has arrived, in which case the exchange transfusion may be avoided.

Following discharge, the hemoglobin should be monitored, a hearing test arranged and development followed up.

primed

# Pediatrics