Tips for doing well in neonatology section of Pediatric Boards
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Some general points

• There are no negative marks and hence no questions SHOULD be left unanswered
• When the answers are not clear in the first reading of the question then and only then try the method of exclusion to get to the best possible answer

Rastogi’s Rule

• Common presentations of common diseases
• Rare presentations of common diseases
• Common presentations of rare diseases
• Rare presentations of rare diseases
FETAL WELL BEING

Biophysical profile

- NST, fetal body movements, breathing, fetal tone, amniotic fluid volume
- Scores of 10 is well fetus, 2 is certain fetal asphyxia, 4 or 6 needs frequent reevaluation for delivery
Electronic FHR monitoring

• **Normal** – FHRv of 6-15 bpm, basis of nonstress test-reactive/positive test is normal
• **Abnormal patterns**
  – Tachycardia, >180, infection
  – Bradycardia, <110, head compression
  – Loss of FHRv, sinusoidal variation

Decelerations

1. Early, mirror image of uterine contractions, head compression
2. Variable, irregular, umbilical cord compression
3. Late, occurs 10s after uterine contraction and last longer, uterine placental insufficiency

ACOG def. for types of tracings

• **Category 1:** normal - routine care
• **Category 2:** indeterminate - continuous surveillance
• **Category 3:** intervention
  – Absent baseline variability
    • Recurrent late dec
    • Recurrent variable dec
    • Bradycardia
  – Sinusoidal pattern
RESUSCITATION

Case

• FT baby is delivered vaginally has HR of 90/m and no respiratory efforts, central cyanosis, flaccid and has no reflexes.
• What is the Apgar score?
• What is the first step during resuscitation?
• What is the subsequent step?
Case continued

• After 1 min of IPPV the HR is 50/min
• What is the next step?

Question 1: Time for the saturation to reach (85-95%) expected normal is

1. 1 minute
2. 5 minutes
3. 10 minutes
4. 60 minutes

Case

• You are preparing to attend the vaginal delivery of a mother at 40 weeks gestation with no abnormal antenatal or perinatal history. AROM was done 8 hours ago, which revealed MSAF. There is a sudden prolonged brady with loss of baseline variability. What is the next step...
• Baby is born with no tone, poor respiratory efforts and HR of 60. What is the next step...
• Apgar are 2 and 4. What is the long term outcome...
NRP- important considerations

• Temp. control: normothermia and prevention of hyperthermia
• MSAF: No suctioning, presence of NRP trained person
• Delayed cord clamping in vigorous term and preterm babies
• HR monitoring best by cardiac monitor
• O2 management: ≥ 35 w start with 21%, in <35 w with 21-30%

• PPV: use of device for PEEP
  – algorithm for HR initial assessment
    • HR increasing - continue PPV 15 sec
    • HR stable not inc. – continue PPV 15 sec
    • HR decreasing and chest not moving – MR SOPA - chest moves - continue 30 sec
  – algorithm for 2nd HR assessment at 30 sec of PPV
    • If ≥100 continue
    • If 60-99 MR SOPA if needed and continue
    • If <60 Intubate/mask, 100% O2, CC
• CC: 2 thumb technique, preferable

INFECTIONS
### GBS

- The incidence of GBS sepsis has decreased from 3-4 to 0.5/1000 live births with 2-3% mortality
- Risk based guidelines (CDC 2010 guidelines)
  - Ill appearing-Rx
  - Maternal chorioamnionitis-Rx
  - GBS with ROM>18h and/or <37w-CBC,BCx
  - GBS with inadequate prophylaxis- 48h observation

### Multivariate risk assessment:
- Newborn EOS risk calculator
- Over 50% decrease in investigation and Rx

### Risk assessment based on newborn clinical condition:
- Investigating and Rx only those who appear ill
- Require 4-6hrly serial exam for 48h

### EOS in less than 34 weeks

- Incidence increased with decreasing GA
  - 6/1000 for <34 w to 32/1000 for =/<24 w
- Cannot use risk stratification
- Upto 75% mortality
- If treated with antibiotic: increased death, NEC and BPD
Clinical presentation
- Early (<3DOL, pneumonia > sepsis > CNS, ascending infection
- Late onset: less mortality but more CNS involvement and sequelae, deep infection as cellulitis, arthritis, osteomyelitis

Other bacterial infections
- E. coli/Klebsiella sp. No change incidence hence ↑ in % in neonatal sepsis, not in absolute numbers. Usual clinical presentation is shock
- Listeria: Gram + rod, in unpasteurized milk, cheese, raw vegetables and uncooked meat
  - Early onset: <7DOL, transplacental, chocolate colored amniotic fluid, preterm deliveries, sepsis/pneumonia
  - Late onset: >7DOL, nosocomial, meningitis with mononuclear cells

Initial drug of choice
- Ampicillin and Aminoglycoside
- Stop asap if culture negative to prevent dysbiosis related disease including neonatal mortality, NEC, BPD, obesity asthma, allergies etc
Other congenital infections

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>CMV</th>
<th>Rubella</th>
<th>Toxoplasmosis</th>
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<tr>
<td>LBW</td>
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<td>Petechiae</td>
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<td>CHD</td>
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<td>Cataract</td>
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<td>Periventricular</td>
<td>Cortical++</td>
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<tr>
<td>Microcephaly</td>
<td>++</td>
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<tr>
<td>Deafness</td>
<td>+</td>
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<td>Diagnosis</td>
<td>Oral swab/urine</td>
<td>IgG</td>
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Management of Syphilis

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<tr>
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<tr>
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Conjunctivitis

- **Time of onset** will give the clue to the cause:
  - Hours-chemical-silver nitrate
  - DOL 2-5-Nisseria gonorrhoeae-purulent, emergency, needs IV antibiotics
  - DOL 5-14-Chlamydia-bilateral, cough
Trisomy 21

**Types**
- 94% non-disjunction
- 3-5% translocation
- 2% mosaic.
  Commonest cause in both old and young mothers is non-disjunction

**Recurrence risk**
- If no translocation- 1% risk till mat. age is 37y
- if mat. translocation-10-15%
- if pat. translocation-5%

**Defects**
- Cardiac (40-50%)- Endocardial cushion defect, VSD
- Extremities-single palmer crease, 5th finger has hypoplastic middle phalange and clinodactyly
- Face-slaning palpebral fissure, Brushfield spots, epicanthic folds, short neck, flat occiput
- GI- duedenal atresia, Hirschsprung Disease
- Neurology- hypotonia, MR,
- Other –hypothyroidism, leukemia, hip dysplasia
Other chromosomal anomalies

- Trisomy 13 (Patau-MIDLINE deformities)
  - Holoprosencephaly, cleft lip/palate, coloboma, sloping forehead, cutis aplasia, VSD, polydactyly, hyperconvex nails, persistence of fetal Hb
- **Trisomy 18 (Edward)**
  - Cardiac (common, VSD, PDA, PS), clenched hand, overlap of 2nd over 3rd and 5th over 4th finger, rocker bottom feet, small mouth/eyes/palpebral fissures, short sternum, hernia, cryptorchidism


**PULMONARY**
Question 2

- A 28 w GA male infant weighing 1500 g experiences respiratory distress requiring ventilation soon after birth. Ventilator parameters are: SIMV of 40 /min, 22/5 pressures and FiO2 of 0.8 to maintain a PaO2 of 60 mm Hg. CXR shows...

Surfactant is administered. What is the most probable observation over next few hours?

1. Reduction in spontaneous respiratory rate
2. Decreased PIP requirement
3. Pulmonary hemorrhage
4. Pneumothorax

RDS-Surfactant Deficiency

- Clinical course: Peak-1 to 3 d and recovery starts with onset of diuresis and ↓ in vent. support
- Risk Factors: Low GA, male, , IDM, perinatal depression
- RDS in term: SPB def, IDM, Beckwith Weideman syndrome, congenital syphilis
- Pathology: Hyaline membrane (cellular debris in fibrinous matrix)
- Treatment-surfactant, supportive
- Complications: pneumothorax
- DD for reticuloalgranular CXR- GBS pneumonia, PAPVR
Case

• An infant is delivered by c/s at 34 wk GA. There is no h/o ROM, maternal fever, or abn. FHR. The infant requires IPPV with a bag-mask and 100% O2 in the delivery room. AS are 5 and 7. Umb. Art. sample pH was 7.23. He is admitted to the WBN, but develops respiratory distress within an hour of life. RA ABG shows 7.10/70/50 with CXR.

Air leak syndromes

• Pneumothorax
  – Air between parietal and visceral pleura
  – Risk Factors: Aspiration (MAS), parenchymal diseases (RDS), PPV (high PIP)
  – Presentation can vary from
    • Tension: severe RD, bradycardia, apnea, hypotension with mediastinal shift
    • Large leak: respiratory distress
    • Small leak: usually accidental finding
  – Complication: IVH by ↓ venous return, SIADH
  – Asymmetrical air entry, transillumination
  – Rx: EMERGENCY if tension-needle aspiration followed by chest tube, supportive

Air leak Syndromes

• Pneumopericardium: air in pericardial sac
  – If large: muffled HS, venous congestion, decreased CO
  – Rx if symptomatic-pericardial aspiration, high mortality
• **Pneumomediastinum**: air in mediastinum
  – Usually after IPPV or difficult intubation, high PIP
  – Muffled heart sounds, CXR: sail sign
  – Usually needs supportive treatment

• **Pulmonary Interstitial Emphysema**: air in interstitial space in the lung tissue
  – Usually preterm with RDS and on ventilation
  – Rx: decrease MAP, if unilateral: selective intubation/blocking of bronchus
**Broncho-Pulmonary Dysplasia**

- BPD - 36 wk of corrected GA with oxygen requirement
- Mechanical trauma (Baro- and Volu-trauma) to susceptible lungs leading to inflammation
- Injury is increased in PT due to low levels of antiproteases and antioxidants
- Poor compliance, increased WOB, pulmonary hypertension, RVH
- Radiographic: honeycomb appearance
- Rx: nonspecific as supportive care, inc. calories (120-150 cal/kg/day), diuretics, bronchodilators, steroids

**Apnea of Prematurity**

- Cessation of air flow for >20s or associated with cyanosis/bradycardia
- Types: Central (no effort, no airflow), Obstructive (no airflow despite effort), Mixed
- Cause: prematurity (usually after 12h of life), infection, maternal med. (narcotics, magnesium), infant med. (prostaglandin), CNS disorders (IVH)
- Treatment: Treat underlying disease, methylxanthines (caffeine), CPAP, mechanical ventilation
**Transient Tachypnea of Newborn**
- It is a diagnosis of exclusion
- Cause: delayed absorption of lung fluid
- Risk factors: elective c/s, IDM, perinatal depression, precipitous delivery
- Radiograph: Fluid in the minor fissure
- Rx: supportive, resolves 2-3 days

**Meconium aspiration syndrome**
- Definition: MSAF+RD+CXR changes
- Clinical: usually post-term, severe respiratory distress
- Complications: pulmonary hypertension, airleak syndromes
- CXR: snow storm appearance
- Rx: respiratory support, correcting acidosis, antibiotics, surfactant, ECMO
Case

- A FT infant is delivered vaginally to mother with DM. He develops cyanosis and respiratory distress immediately following birth that requiring IPPV with 100% oxygen. There is no improvement and is admitted to the NICU. His BW is 4.5kg, has saturation of 70 in the right hand and 45 in the left leg, with precordial lift and a loud S2. CXR reveals decreased pulmonary blood flow. The MOST likely cause of respiratory distress in this infant…

Pulmonary Hypertension

- **Cause**
  - Maladaptation: normal vasculature but vasoconstriction (hypoxia, hypothermia, polycythemia, pneumonia)
  - Maldevelopment: abnormal structure of pulmonary vascular bed (chronic intrauterine hypoxia, pulmonary hypoplasia)
- **Rx** respiratory support (no hyperventilation), correcting acidosis, iNO, sildanefil, ECMO

* FT baby boy presented mild hydrops, after birth as RD due to lung hypoplasia and scaphoid abdomen. CXR showed…
Congenital diaphragmatic hernia

- **Types:** Posterolateral thru Foramen of Bochdalek (L>>R) and central thru Foramen of Morgagni
- **Complications:** related to lung hypoplasia and pulmonary hypertension
- **Treatment:** IMMEDIATE intubation, correcting pH and delayed surgical repair. May require ECMO

CARDIOLOGY

Congenital Heart Disease-Some Facts

- Incidence 8/1000 live births (excluding PDA in PT newborns) with 25% have other associated abnormalities
- **Suspect:** cyanosis with minimal respiratory distress
Congenital Heart disease - some facts

- **VSD** - commonest CHD
- **TGA** - commonest cyanotic HD in first week of life
- **HLHS** - 2nd commonest cyanotic HD and commonest cause of cardiac mortality in first week of life
- **TOF** - commonest cyanotic HD beyond neonatal period

Presentation of CCHD

- **5T’s, DO, ESP** - TGA, TOF, TAPVR, Tricuspid atresia, Truncus arteriosus, DORV, Ebstein’s Anomaly, Single ventricle and Pulmonary atresia
- **HLHS Vs. sepsis**: usually HLHS presents after the duct is closed by 48-72 h and neonate presents with cardiac failure to the ER with no murmur. If no high risk factors for infection, always consider the diagnosis of HLHS

Maternal conditions and CHD

- **Association with maternal drugs**
  - Aspirin/Indomethacin-PH/PDA closure
  - Lithium-Ebstein’s anomaly
  - Ethanol-VSD
- **Association with maternal diseases**
  - Lupus-Cong. Heart block (anti Ro/La Ab)
  - Diabetes (commonest-VSD, TGA; most specific- asymmetrical ventricular hypertrophy)
**sCalp Injuries**

- **In Subcutaneous tissue:** *caput succedaneum*, soft, crossed midline/sutures, usually with molding, resolves over several days.
- Beneath Galea Aponeurotica in Loose areolar tissue: *subgaleal*, can move to neck and behind ear, can cause anemia, hypotension, jaundice, resolves in 2-4 wk.
- **Subperiosteal cephalhematoma**, confined to suture lines, firm, 10% have skull fracture, jaundice, resolve in weeks to months.
A 2 day old preterm infant who was born at GA of 26 wk and BW of 650g on RA CPAP. Her blood pressure has dropped acutely, and developed seizures at 2DOL. There B/L equal breath sounds, no murmur, hypotonia, a bulging anterior fontanelle, and lethargy. Lab. Shows severe anemia, metabolic acidemia, and hyperglycemia. This is most probably related to…

1. Severe hemolysis
2. Adrenal hemorrhage
3. Intraventricular hemorrhage
4. Sepsis/Meningitis

Intraventricular Hemorrhage
- From: germinal matrix
- Incidence: ↑ with ↓GA
- Timing: 50% in 24h and 90% in 72h
- S/S of anemia + CNS involvement.
- Prognosis: worsening with increasing grades
- Complication: PHH/PVL

Birth Asphyxia
- Definition (ACOG)
  - pH<7, AS<3 at 5min, neurological sequelae (HIE), multiple organ dysfunction
- HIE staging (Sarnat Stages)
  - Stage 1: hyperactive CNS with sympathomimetic activity, 100% normal
  - Stage 2: decreased CNS activity, parasympathomimetic activity, seizures, 80% normal
  - Stage 3: variable presentation, seizures rare, burst suppression EEG, 100% severe sequelae
Cerebral palsy

- Non progressive neurological deficit
- Incidence: 2-5/1000
- Clinico-pathological patterns:
  - Selective neuronal necrosis- HIE, diffuse damage, Quadriplegia, MR, seizures
  - Parasagittal cerebral injury- ↓ perfusion, necrosis in watershed areas, weakness of proximal muscles, U>L
  - Focal or multifocal ischemia- meningitis, trauma, thrombotic syndromes, as hemiplegia, seizures, cognitive defects
  - Status Marmoratus- kernicterus, basal ganglia, choreiform movements

Brachial plexus injury

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<tr>
<th></th>
<th>Erb-Duchenne</th>
<th>Klumpke</th>
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<tr>
<td>Roots</td>
<td>C5-7</td>
<td>C8-T1</td>
</tr>
<tr>
<td>Incidence</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Typical S/S</td>
<td>Waiters tip</td>
<td>Ape hand</td>
</tr>
<tr>
<td>Differentiate</td>
<td>Palmar grasp +</td>
<td>-</td>
</tr>
<tr>
<td>Associated</td>
<td>C4/5 (phrenic nerve), C7 (scapular winging)</td>
<td>T1 (Horner’s syndrome)</td>
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Hydrocephalus

- 2 types:
  - Obstructive- common, commonest cause is post hemorrhagic HC, others are aqueductal stenosis, Dandy-Walker Syndrome (cystic dilatation of 4th ventricle with hypoplasia of vermis)
  - Communicating- usually after bleeds, infections, NTD, Arnold Chiari malformation
- Treatment: repeated LP, shunts
- Shunt complications- block, infections
Case

- Mother is rushed in for stat c/s for abruption. Apgars are 2, 3, 8; improving after IPPV, chest compression and fluid resuscitation. Admitted to NICU. Mother had uneventful antenatal course. Baby has not passed urine and serum sodium is 125. He develops generalized tonic clonic seizures and bradycardia after 12h treated with phenobarb.
- What is the most likely cause for these seizures…

Neonatal Seizures

- Types
  - Subtle - most frequent, oral, facial ocular activity, may be associated with changes in HR, resp. BP and sa.
  - Multifocal clonic - one limb migrating to another
  - Focal clonic - may represent focal disease
  - Tonic - change in posture, more in preterm
  - Myoclonic
- Many causes - asphyxia, metabolic, infection, trauma, malformation

Hearing Screening

- Discussing neonatal hearing screening with medical students on rounds in WBN. The statement that you are MOST likely to include in your discussion is that
  - A. an infant should be tested while asleep
  - B. intervention in children who have hearing impairment should begin at 12 months of age
  - C. normal neonatal hearing screening results should be confirmed by repeat testing at 6 months of age
  - D. otoacoustic emission is the definitive procedure for testing hearing in newborn
  - E. visual reinforcement audiometry currently is used as a screening test in newborns
Inborn errors of metabolism

- **When to suspect**: just about any S/S specially if the initial usual diagnosis e.g. sepsis is not responding to the usual forms of treatment e.g. antibiotics.

- **Specific smells**
  - Sweaty feet - Isovaleric acidemia/Glutaric aciduria
  - Male cat urine - Glycinuria
  - Maple syrup odor - Branched chain aa
  - Musty odor - PKU

Hyperammonemia-confirm

A. Metabolic acidosis and Hypoglycemia

- No ketosis
  1. Fatty acid oxidation defects
  2. Organic acidurias

- Normal lactate
  1. Butyric acidosis
  2. Glutaric aciduria

- Ketonuria

- High lactate
  1. Congenital lactate acidosis
  2. MMA, PA, hPA
  3. Multiple carboxylase def.
Hyperammonemia-confirm
B. Normal pH and glucose → Plasma and urine aa

Citulline
Normal-range
1. Congenital lysine intolerence
2. Rett syndrome
Abnormal
Trace
Test Orotic acid

Mildly High
Arginosuccinic acidemia 2.

Very High
1. Citrullinemia
Argininosuccinate synthetase def.

Low-CPS def
High-GTP def.

Temperature Regulation
- Neonates more prone to heat loss as
  - ↓skin thickness- radiant+ conductive loss
  - ↓subcut. fat
  - ↓peripheral vasoconstriction leading to ↓heat conservation
  - Immature autonomic nervous system
  - ↑BSA to wt.-radiant heat loss
- Convective Incubators-large radiant loss( ↓by double wall) + small evaporative loss( ↓by inc. humidity) and small conductive heat loss (↓by rubber mattress)
- Radiant Warmer-large convective and evaporative loss, ↓by covering by Saran wrap

Hypothyroidism
- Commonest cause-thyroid dysgenesis
- Early presentations-prolonged jaundice, large post. fontanelle
- Others-umbilical hernia, macroglossia, hypotonia, goiter
- Diagnosis- by newborn screening-low T4 and high TSH
- Rx-levothyroxine

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• Rx-levothyroxine
Case

- Baby delivered after difficult vaginal delivery to a mother with gestational diabetes poorly controlled on insulin. Baby was 4.3kg and was send to WBN where he developed tachypnea and jitteriness.
- What is the Differential Diagnosis for tachypnea and jitteriness for this baby?

Hypoglycemia- IDM

- Hypoglycemia is commonest presentation of IDM and can present as tachypnea. Other IDM related causes for tachypnea are RDS, TTN, CHD (VSD), birth asphyxia, birth trauma and hypocalcemia.
- Other common presentations of IDM are polycythemia and jaundice.
- Specific malformations for IDM- Hypertrophic Obstructive Cardiomyopathy d/t asymmetrical ventricular septal hypertrophy and caudal agenesis syndrome.

Hypocalcemia

- Types
  - Early (<3DOL): maternal causes (DM, hyperparathyroidism), perinatal causes (prematurity, asphyxia, infections)
  - Late (>3DOL): hypoparathyroidism, hypomagnesemia, vitamin D def.
- S/S: If symptomatic as jitteriness, high pitched cry, Chvostek/Trousseau sign, seizures, prolonged QTc
- Rx: Underlying cause, Ca, Vit. D, low phosphate formula.
Congenital Adrenal Hyperplasia

• Commonest cause: 21 hydroxylase def. (followed by 11beta hydroxylase def.)
• S/S with 21OH-salt wasting in 2nd week of life with ↑K, ↓Na and hypotension with pseudohermaphroditism in females and males may have precocious puberty.
• Diagnosis: ↑17OHP in amniotic fluid or serum (Newborn Screen)
• Rx: Antenatal-maternal glucocorticoid, Postnatal-replacement of GC/MC

HEMATOLOGY

Isoimmunization-Rh

• Mother is Rh- i.e. dd (Rhesus An has 3 components C, D, E with D as the major component). Baby is Rh+ i.e/DD or Dd.
• Second pregnancy/ Rh+ antigen induces IgG which crosses placenta easily causing hemolysis of fetal RBC.
• Prevented by giving Anti D antibody(Rhogam) to Rh- mother at 28 wk GA and at birth of Rh+ baby.
Isoimmunization-ABO

- Incidence not influenced the number of pregnancies
- Mothers with group A or B produce IgM antibodies and that of O produce IgG which easily crosses placenta
- Usually mild as the antigen is on types of cell and they capture the antibodies
- Has spherocytosis with B-O incomp.

Phototherapy

- Mechanism of action
  - Configurational Photo-isomerization: 4Z-15Z to 4Z-5E
  - Structural Photo-isomerization: lumibilirubin
    - Photo-oxidation
- Blue light- effective wavelength (710-780nm) and penetrates skin well.
- If phototherapy given to baby with high direct bilirubin -bronze baby syndrome

Jaundice related to breast feeding Vs Breast feeding jaundice

- Jaundice related to BF
  - Usually exaggerated physiological jaundice due to decreased intake
- BF jaundice
  - Prolonged with peak of 20-30mg/dl by 2 wk and then normalize over 4-12 wk
  - Rapid decrease after cessation of breast feeding for 24 h and rises 2-4mg/dl after resuming BF
  - Can cause kernicterus
Thrombocytopenia

- Sick or well
  - Sick
  - Sepsis/DIC
  - Alloimmune
    - Antiplatelet antibodies as that of lupus, ITP
    - Maternal and newborns platelets are low
  - Autoimmune
    - Transplacental transference of maternal antibodies, with normal maternal platelets
    - Severe, can have IC bleed, death in 20%

Question 4:
BB delivered at home with precipitous delivery was admitted to WBN and discharged with mother. Was exclusively breast fed. Develops fresh bleeding per rectum on DOL 4. There is no other site of bleeding or pertinent history. Clinical examination is normal. What is the next step…

1. To order GI series
2. Start antibiotics after CBC/BCx
3. Administer a dose of phytadione
4. Emergency laparotomy

Hemorrhagic Disease of Newborn

- Vitamin K needed for carboxylation reaction which activates the clotting factors
- Newborns have ↓Vit.K: ↓bacterial flora in intestine to produce Vit.K, immaturity of hepatic synthesis
- Types:
  - Early: <24h, maternal drugs anticonvulsants, warfarin or ATT, decreased transplacental transference
  - Classic: 2-7d, exclusive breast feeding
  - Late: 2w-6m, hepatobiliary disease, IC Bleed
Esophageal atresia

- 30-40% with associated abnormalities as in VATER and VACTERL associations
- 4 varieties with type 3 being the commonest: upper end of esophagus is atretic and lower end has fistula with trachea
- Type 4 or H type is rare but commonest one from the examinations perspective: S/S as cough during feeding and recurrent aspirations
- Rx-surgical: primary or delayed

Case

- Baby is delivered to mother with history of polyhydramnios. Mother had irregular antenatal care. Baby did not tolerate feeds and started to have non-bilious vomiting. OG tube could be passed to the stomach. AXR was ordered and shows...
Double bubble sign
- Associated with duodenal atresia
- High rate of association with trisomy 21, malrotation and CHD.

Case
- 24 week PT AGA BG has had relatively uneventful course in NICU. At 8 weeks of life when tolerating full enteral feeds developed abdominal distention and residuals. Feeds were stopped and AXR shows. Was treated medically. What is the commonest sequelae?

Necrotizing Enterocolitis
- Up to 10% of those born <1500g
- Predisposing factors: prematurity, feeds, infection, poor perfusion
- AXR: pneumatosis intestinalis
- Outcome: high mortality and morbidity such as small gut syndrome (s/p surgical resection) or strictures (s/p medically treated)
Congenital Hyperplastic Pyloric Stenosis

- 3/1000 births, male x 5
- Related to decreased NO production
- Hypochloremic, hypokalemic, metabolic alkalosis
- Barium-string sign, US-bull's eye sign
- Rx-Pyloromyotomy

Case

- 2 day old FT baby in WBN develops distention and has not passed meconium since birth. PMD orders an AXR which shows large dilated stacked loops with absence of air in the recto-sigmoid region.
- What is the next step...

- Barium Enema revealed gradual narrowing of the sigmoid
- The likely diagnosis is...
- Confirmed by…
**Hirschsprung's Disease**

- 1:5000, usually male, 80% rectosig. only
- Can be associated with trisomy 21
- Failure of cranial to caudal migration of neural crest cell -parasymp. innervation
- Diagnosis: AXR, biopsy (absent ganglion cells)
- Complication: Acute bacterial colitis
- Rx: single stage pull through or initial colostomy followed by correction

**Meconium plug vs. ileus vs. peritonitis**

- **Plug**
  - benign variation of Hirschsprung’s disease
  - Delayed passage of meconium
  - Usually has small colon, IDM
- **Ileus**
  - 90% have CF
  - bilious vomiting, obstruction, AXR-bubbles in the intestinal lumen
  - enema successful in 60%
- **Peritonitis**
  - In utero perforation
  - Secondary to ileus, atresia, volvulus, gastroschisis
  - Usually seal spontaneously or can require surgery

**Omphalocele vs. Gastrochisis**

<table>
<thead>
<tr>
<th></th>
<th>Omphalocele</th>
<th>Gastrochisis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Incidence</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Chrom. Abn.</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Midline</td>
<td>Yes</td>
<td>No (80% on R)</td>
</tr>
<tr>
<td>Covering</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Umb. cord</td>
<td>Involved</td>
<td>Normal</td>
</tr>
<tr>
<td>Assoc. Abn.</td>
<td>More</td>
<td>Less</td>
</tr>
<tr>
<td>M/M</td>
<td>More</td>
<td>Less</td>
</tr>
</tbody>
</table>
DERMATOLOGY

ERYTHEMA TOXICUM
- Most common 30-70%
- Onset DOL 2-3
- 1-3mm erythematous macule/papule-pustule
- Fades in 5-7 days
- Benign, has eosinophils

PUSTULAR MELANOSIS
- More in dark skinned
- 3 stages-non-inflammatory pustule, ruptured pustule with scale, hyperpigmented macule (up to 3mm)
- Benign, has neutrophils
- No Rx
Question 5

- Baby boy born with oligohydramnios, Potters Sequence, and hypoplastic lungs requiring ECMO. Had B/L hydronephrosis diagnosed by antenatal US and the voiding cystourethrogram shows…

The likely diagnosis is

1. Oligohydroamnios
2. Atonic bladder
3. Posterior Urethral Valves
4. Congenital Marion’s disease

25% 25% 25% 25%

ADDITIONAL MATERIAL
Case

- A FT boy is born to a woman who has known multiple drug abuse problems. Her urine drug screen was positive for barbiturates, benzodiazepines, and opioids. The infant is delivered NSVD with AS of 8 and 9. You are asked to evaluate the infant for early discharge at 23 hours of age. Findings on physical examination are normal, with the exception of jitteriness, and the infant is not breastfeeding well. Should you clear for discharge?

Timing of Drug Withdrawal

- Depends on the half life of the drug, the time before delivery drug was abused and the severity often depends on the duration of abuse during pregnancy
- Usually with most of the drugs withdrawal is within 1-2 days but with methadone/subutex can be delayed till 5-7 days after birth due to long half life

Late Preterm Infants

"Late Preterm" and "Early Term" Definitions

- "Late Preterm" infants are born between 34 and 36 weeks of gestation
- "Early Term" infants are born between 37 and 40 weeks of gestation

- Late Preterm
- Early Term
- Full Term
- Post-term
RSV prophylaxis guidelines (AAP 2009)

- Who
  - All <29w and <12 months old
  - All <32w and <12 months old with CLD
  - All with CLD on O2 6 months before RSV season and <24 months old
  - Pulmonary abn, neuromuscular disease and <12 months old
  - All hemodynamically significant CHD on med, requiring surgery, mod.-severe PH and <12 months old
  - Post Cardiac transplant and <24 months old
  - Immunocompromised and <24 months old
  - Downs and CF- if other criteria met
RSV

- When: From November for 5 doses (MAX) - covers 6 months
- Stop if breakthrough RSV positive infection
- No use as treatment and in hospital outbreaks

Management of Hepatitis B

<table>
<thead>
<tr>
<th>Mat. status</th>
<th>Newborn &gt;2kg</th>
<th>Newborn &lt;2kg</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>HBV</td>
<td>HBIG</td>
</tr>
<tr>
<td>+</td>
<td>3 doses, 1&lt;12h</td>
<td>1 dose, &lt;12h</td>
</tr>
<tr>
<td>unknown</td>
<td>3 doses, 1&lt;12h</td>
<td>1 dose, &lt;7d</td>
</tr>
<tr>
<td>–</td>
<td>3 doses, 1st at 1-2M</td>
<td>None</td>
</tr>
</tbody>
</table>

PDA

- Normal course
  - physiological closure 12-15h, anatomic closure several months,
  - about 4% of term, 10% of 30-37 wk and 50% of <30 wk do not close by 72 h and considered PDA
- S/S-
  - Term-asymptomatic, systolic or machinery murmur, bounding pulse, CHF
  - Preterm can also have ↓bf to the gut can cause NEC, pulmonary h'inge, and BPD
- Treatment
  - Fluid restriction, maintaining hematocrit, ibuprofen (indomethacin, acetaminophen), surgical correction
Case

- You receive a telephone call from the mother of one of your patients, who tells you that she is 27 weeks pregnant and that her obstetrician has diagnosed a fetal arrhythmia. In discussion with the obstetrician, you learn that the fetal heart rate is 240 beats/min and that there is a 1:1 relationship between the atrial and ventricular contraction. What advice would you give to the mother regarding the management of the baby after delivery?

Fetal arrhythmias

- Bradyarrhythmias: Heart blocks-following clinically delivering if there is development of hydrops
- Tachyarrhythmias: Supra-ventricular Tachycardia-treated with giving digoxin to the mother

Names of some surgical procedures for CHD

- Rashkind balloon septostomy:TGA
- Blalock Taussig Shunt:TOF
- Glenn procedure:TA/SV with PS
- Jantene:TGA
- Fontan:TA/SV
- Norwood:HLHS-Stage 1-3
Norwood for HLHS

- Stage 1
  - Atrial septectomy
  - PA to Asc.aorta
  - BT shunt
- Stage 2
  - Glenn
  - Remove BT
- Stage 3
  - Fontan

Glycogen storage disease

- 8 types, types 1,2 and 3 are commonest
- Type 1 – von Gierke (liver, renal, GI). Glucose 6 PO4ase def., lactic acidosis, hepatomegaly, diarrhea, bleeding disorder, poor prognosis
- Type 2 – Pompe (muscle, nerves), lysosomal glucosidase def., muscle weakness, cardiomegaly, CHF, poor prognosis
- Type 3 – Forbes (liver, muscle), low glucose, hepatomegaly, muscle fatigue, onset after neonatal period, good prognosis

MPS and Lipidoses

- MPS- dysostosis multiplex, Alder Rieley bodies in WBC and urine MPS
  - Hunter-iduronidase def., onset-1y, cloudy cornea, HSM, coarse features, short stature, kyphosis
  - Hunters-iduronidase sulfatase def., onset 1-2y, X-linked, only MPS with retinal abn.
• Lipidoses
  – Gaucher’s-glucocerebrosidase def., Gaucher cell in bone marrow, normal retina
    • type I-normal CNS, onset any age
    • type II-profound CNS involvement, onset 1y
  – Niemann Pick-sphingomyelinase def., foam cells in bone marrow
    • type A cherry red spot, profound CNS involvement, onset 1m
    • type B normal retina, normal CNS, onset any age

Galactosemia
• AR, galactokinase or Galactose-1-PO4ase uridylytransferase def.
• Presents when feeds are introduced as lethargy, hepatomegaly, liver failure, renal tubular acidosis
• Can have cataract at birth
• ↑ risk of infection specially E.coli
• Lab- ↑LFT’s, galactose in urine (reducing substance positive with negative glucose oxidase test)
• Rx- elimination of all galactose and lactose in diet

PKU and Homocystinuria
• Classic PKU-AR, def. of phenylalanine hydroxylase, mousy/musty urine odor, severe MR and seizures if untreated, diagnosed by NBS, Rx by low phenylalanine diet
• Homocystinuria-AR, def. of cystathianine synthetase, usually asymptomatic in neonatal period, has downward dislocated lens (D:D Marfan’s), myopia, osteoporosis, scoliosis, arachnodactyly, ↓ joint mobility (D:D Marfan’s), MR, seizures, thrombotic episodes, Rx ↓methionine, supplement cysteine, folate, pyridoxine
AAP guidelines for bili management

• To measure bilirubin in hours of life
• High intensity phototherapy and specific follow up depending on the zone in the hourly bilirubin charts