Other Diseases of Prematurity

Neonatal/Pediatric Cardiopulmonary Care

Other Diseases

Pulmonary Dysmaturity (Wilson-Mikity Syndrome)

Description

- Disease of functional & structural pulmonary changes seen in premature infants with no apparent underlying lung disease
Other Diseases of Prematurity

Pathophysiology

- Unknown
- Only common finding is prematurity with the majority of infants <
- 1 theory =
- May also be an association between

Diagnosis

- Chest Radiograph
  - Similar to Stage III & IV of BPD but infant has not been mechanically ventilated
- Lung biopsy
  - Early
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  - Late
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Clinical Signs & Symptoms

- Initial symptoms usually appear near the end of 1st week & are usually mild
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Clinical Signs & Symptoms

- Symptoms gradually worsen over following 2-6 weeks leading to acute phase of the disease
- Acute phase lasts from days to weeks & presents as
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- 2/3 survive this phase & begin gradual recovery
- Clearing of disease by age

Retinopathy of Prematurity (ROP)

- Initially described in 1942 as RLF
- Term literally means
- \textit{ROP} is a more descriptive term of the actual events neonate passes through leading to scar formation
- Epidemic in 1940-1950s so less O2 used
  - ↓ in ROP cases
  - ↑ in mortality & cerebral palsy
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History

- Studies have identified a definite link between development of ROP & O₂ use (specifically PaO₂)
- Other factors that contribute
  - Retinovascular immaturity
  - Circulatory & respiratory instability

History

- ROP occurs in
  - 25-30% of premies <35 wks with 40% vision impairment & blindness
  - 10 schools for the blind - 18% have Hx of ROP

Physiology of Developing Eye

- Retinal capillaries begin branching at 16 wks - from optic nerve outward
- Reach perimeter at

  - If premature -

  - Then if develop ROP -
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Pathophysiology

- If high PaO₂ → retinal vessels constrict causing necrosis of vessels “vaso-obliteration”
- In attempt to re-establish blood supply to retina → un-necrosed vessels multiply → may extend into liquid portion of eye → hemorrhage → formation of scar tissue behind retina → traction → detachment

Pathophysiology

- Damage can stop at any point
- Although ↑ PaO₂ implicated, so has:
  - Immaturity
  - Hypoxia
  - IVH
  - Apnea
  - Infection
  - PDA
  - NEC
  - Hypercarbia
  - Lactic acidosis
  - Genetic factors
  - Hypotension
  - Birth weight
  - Bright lights
  - Blood transfusion
  - Vit. E deficiency
  - Early intubation
  - Duration of O₂
  - Prostaglandin synthetase inhibitors
  - Prenatal complications

Diagnosis

- 5 stages of classification
  - Stage 1: A thin white demarcation line is seen separating the avascular retina from the vascularized retina.
  - Stage 2: A ridge is now formed and rises up from the plane of the retina. New vessels may be seen posterior to the ridge.
  - Stage 3: There is proliferation of extraretinal fibrovascular tissue. It is often seen posterior to the ridge or connected to the posterior aspect of the ridge.
  - Stage 4: There is subtotal detachment of the retina.
  - Stage 5: Continued traction and build up of fluids. The retina becomes completely detached.
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### Diagnosis

- Dx made by
- Appears between 35-45 wks gestation (time of retinal vascular development) and may progress through all stages in a few weeks

### Treatment

- Vitamin E (antioxidant) does not help
- Cryotherapy
  - Cold probe into eye -- freezes avascular portion preventing further abnormal vessel growth
  - Must be done at Stage III or less
- Complications
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- Laser therapy
  - Same result as cryotherapy but less invasive, less traumatic
  - New studies
    - 
    -
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Treatment

- Best treatment is still
  - Cautious use of oxygen
  - PaO\textsubscript{2} as low as possible ( )
  - Monitor
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Intraventricular Hemorrhage (IVH)

- Bleeding into cranium = major source of morbidity in premie population
- Can occur in many different areas
  - Subdural, subarachnoid
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  - Cerebellar tissue
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  - **Intraventricular**
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IVH
Pathophysiology

- Developing brain is
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  - Small bleeds are confined & usually no residual effects

Pathophysiology

- Severe hemorrhage
  - Blood enters brain tissue
  - Blood enters ventricles
  - Enlarge & compress brain tissue

Pathophysiology

- Most occur 1 to several days post-delivery
- Triggered by
  - Fluctuations in blood flow
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  - Premie babies of alcohol-consuming moms
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Signs & Symptoms

- Depends on severity of bleeding
- Range from severe, rapid deterioration to no apparent side-effects
- Signs & symptoms
  - Apnea
  - Hypotension
  - ↓ Hematocrit
  - Flaccid muscle tone
  - Bulging fontanelles
  - Tonic posturing

Classification

Complications

- Most serious = post-hemorrhagic hydrocephalus (PHH)
  - Caused by obstruction of CSF outflow & impairment of CSF return to brain → ↑ ICP
  - Treatment
    - Remove CSF by lumbar puncture
    - If persistent -
      - Surgical placement of ventricular-peritoneal shunt
      - Ventriculostomy with external drainage
Complications

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Treatment

• Best is to avoid factors that lead to IVH
• Carefully avoid factors that lead to fluctuations in cerebral blood flow
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• Low-dose indomethacin (Indocin) has been shown to decrease incidence & severity

Treatment

• Rx following IVH is mainly supportive (very little can be done to treat once bleed has occurred)
• Precautions to prevent further hemorrhage & damage
  • Osmotic diuretics
  • Monitor for increased bilirubin
  • Avoid hypotension in presence of ↑ ICP to avoid ↓ CPP