Persistent Pulmonary Hypertension of the Neonate (PPHN)

- Also known as Persistent Fetal Circulation (PFC)
- Seen most frequently in term, post-term & in patients suffering from...
Pathophysiology

affected infants have severe, persistent pulmonary vasoconstriction

Pathophysiology

right heart pressure > left heart pressure
continuation of factors that allow fetal circulation pathways: blood shunting through f.o., d.a. & away from lungs
metabolic & respiratory acidosis hypoxemia which perpetuates pulmonary vasoconstriction

Etiology

• Underlying cause unknown
• Symptoms imply dysfunction of pulmonary vasoregulation resulting in abnormally high PVR
• Current theories include
  – Chronic uterine hypoxia
  – ↑ development of vascular smooth muscle
  – Perinatal factors that cause vasospasm
Diagnosis

• If hypoxemia is severe or worsening, think
  
• To differentiate (at bedside)
  – Hyperoxia test
  – Preductal vs. postductal test
  – Hyperoxia-hyperventilation test

Hyperoxia Test

• Give 100% oxygen x

• Do
  
• If PaO₂ < 100 mmHg =

Preductal vs. Postductal

• Measure preductal & postductal SpO₂ or PaO₂
  
• If preductal > postductal by 15-20 mmHg or more =
Hyperoxia-Hyperventilation Test

- Most accurate of the 3
- Patient hyperventilated to PaCO₂ of 20-25 mmHg & pH 7.50
- Alkalosis produces pulmonary vasodilation & systemic vasoconstriction → improves lung perfusion & O₂ content of arterial blood
- If PaO₂ <50 mmHg before test & rises to >100 mmHg after test =

Diagnosis

- Echocardiograms have advanced Dx of PPHN
- Will show
  -
  -
  -
  -

Treatment

- *Hyperventilation therapy*
  - Increases risk of barotrauma
  - Rates up to 150/min is recommended to allow ↓ inspiratory vent pressures
  - Make sure to allow enough
  - If unsuccessful →
Other

**Treatment**

- **INO**
  - Inhaled nitric oxide
  - Powerful, selective pulmonary vasodilator by relaxing smooth muscle
  - Mixed with oxygen then added to ventilator circuit
  - Half-life =
  - Criteria: 1) 2)

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**Treatment**

- **INO - Results**
  - Pulmonary vasodilation
  - Improved V/Q match
  - Increased PaO₂
    - **
  - Decreased PAP
    - **
  - No change in SVR

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**Treatment**

- **INO - Side-effects**
  - NO + Hgb ➔
    - Normal metHgb =
    - 5-6% =
    - Useless in
    - MetHgb has no
Treatment

• *INO* - Side-effects
  – NO + O₂ →
  •
  •

Administration of NO

• Mixed by special machine then introduced into inspiratory limb of vent circuit just proximal to ETT
• Mixed & added at last minute to minimize

Administration of NO

• Contraindications
  – Neonate that is dependant on
• Precautions
  – Rebound: abrupt DC can make
  – MetHgb formation:
  – NO₂ formation:
  – Drug interactions: has additive effects with
Dosage of NO

- 20 ppm x
- Decrease to 5 ppm for up to 14 days, then wean to 0 ppm
- Constant dose t/o resp cycle
- May use with
- Monitor
- –
- –
- –

Transient Tachypnea of the Newborn (TTN)

TTN

- = RDS Type II because of similarities in symptoms
- Etiology
  - Retention of lung fluid following birth
  - Occurs in term & near-term neonates with history of C-section or very fast deliveries
Diagnosis

• Within a few hours, baby shows

• May have
  • ↑ PaCO₂
  • CXR mimics early RDS

Diagnosis

• Made after all other potential problems have been ruled out
  – IRDS
  – **Pneumonia**

Treatment

• Treat symptoms
  – Warm, humidified O₂
  – Positive pressure:
  – Frequent turning
  – Gentle CPT
  – Broad-spectrum antibiotics (since often mistaken for pneumonia)
Other

Apnea

- True apnea = cessation of breathing for long enough to cause cyanosis &/or bradycardia
- Usually takes
- Classed as

<table>
<thead>
<tr>
<th>Etiology</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Respiratory</strong></td>
</tr>
<tr>
<td>RDS</td>
</tr>
<tr>
<td>Congenital upper airway anomalies</td>
</tr>
<tr>
<td>Airway obstruction</td>
</tr>
<tr>
<td>Post-extubation</td>
</tr>
<tr>
<td>CPAP</td>
</tr>
<tr>
<td>Pneumonia</td>
</tr>
<tr>
<td>Hypoxia</td>
</tr>
<tr>
<td><strong>Environmental</strong></td>
</tr>
<tr>
<td>Increased environmental temperature</td>
</tr>
<tr>
<td>Suctioning</td>
</tr>
<tr>
<td>Feeding</td>
</tr>
<tr>
<td><strong>Metabolic</strong></td>
</tr>
<tr>
<td>Hypoglycemia</td>
</tr>
<tr>
<td>Hypo- and hypernatremia</td>
</tr>
<tr>
<td>Hypocalcemia</td>
</tr>
<tr>
<td>Hypo- and hyperthermia</td>
</tr>
<tr>
<td><strong>Cardiovascular</strong></td>
</tr>
<tr>
<td>Congestive heart failure</td>
</tr>
<tr>
<td>PDA</td>
</tr>
<tr>
<td>Anemia</td>
</tr>
<tr>
<td>Tachycardia &amp; bradycardia</td>
</tr>
<tr>
<td>Sepsis</td>
</tr>
<tr>
<td>Polycythemia</td>
</tr>
<tr>
<td><strong>Central Nervous System</strong></td>
</tr>
<tr>
<td>IVH</td>
</tr>
<tr>
<td>Meningitis</td>
</tr>
<tr>
<td>Seizures</td>
</tr>
<tr>
<td>Pharmacologic sedation</td>
</tr>
<tr>
<td>Karlacteri</td>
</tr>
<tr>
<td>Immaturity of resp centers</td>
</tr>
<tr>
<td>Tumors</td>
</tr>
<tr>
<td><strong>Gastrointestinal</strong></td>
</tr>
<tr>
<td>NEC</td>
</tr>
<tr>
<td>Gastroesophageal reflux</td>
</tr>
</tbody>
</table>
Central Apnea

- nonobstructive apnea
- Common type is apnea of prematurity (incidence is $1/\alpha$ to gestational age)
- absence of airflow and respiratory effort
- Many causes ----

Causes of Central Apnea

- ↓ peripheral chemoreceptor sensitivity
- ↓ arousal response (adults wake up if $\text{PaCO}_2$ ↑, $\text{PaO}_2$ ↓)
- ↓ stimulation of airway reflexes
  - Adults: something in airway (i.e. gastric reflux) →
  - Infants: something in airway →

Causes of Central Apnea

- Dysfunction of respiratory centers
- Dysfunction of ventilatory muscles
- Dysfunction of peripheral nervous system
  - Diseases affecting neurotransmission
  - Toxins (botulism)
  - Drugs that inhibit NM junction
  - Trauma
Causes of Central Apnea

• Others
  – Thermal instability
  – Metabolic disorders
  – PDA
  – Shock
  – Anemia
  – Sepsis
  – NEC

Treatment of Central Apnea

• Drugs that stimulate respiratory centers
  – CMV

Obstructive Apnea

• = absence of airflow with ventilatory effort
• Airway obstructs during inspiration
Causes of Obstructive Apnea

•
•
•
•
•
•

Diagnosis

• Dx with polysomnogram
• Monitors
  • Chest wall motion (impedance plethysmography)
  • Airflow (nasal)
  • $P_{ETCO_2}$
• ECG
• HR
• $SpO_2$
• pH

Nasal airflow

Vent efforts

Mouth open

Central apnea

Obstructive apnea

Hypopnea

EEG

HR
Rx for Obstructive Apnea

- Drugs (to reduce airway narrowing)
- Surgery
- Nasal CPAP during sleep

Diaphragmatic Hernia

- Incomplete embryological formation of diaphragm → herniation of abdominal contents into thorax
- Occurs mostly on left side through Foramen of Bochdalek
- 1/2,200 births
Diaphragmatic Hernia

- Stomach & intestines enter thorax compressing lung & pushing mediastinum to the right

- Prenatal: Lung on left does not develop
- Postnatal: Abdominal contents compress lung

Symptoms

- Chest X-Ray

Chest X-Ray
Other

Chest X-Ray

Treatment
- Mortality is very high
- Immediately upon diagnosis
  - Rates ≥
  - Hypoplastic lung
    - Is very stiff & susceptible to barotrauma

Treatment
- UAC
  - For ABG & BP
- Surgical repair
- Post-op
  - Vent x ≥
  - ↑ rates, ↓ PIP
  - Paralyze to ease ventilation
  - Dopamine & colloids if Qt is low
  - Wean as tolerated
Other

Patent Ductus Arteriosus (PDA)

Review

• Pulmonary artery blood is shunted away from fetal lungs through the ductus arteriosus
• Patent in fetus due to
  –
  –

Review

• Closure following delivery caused by
  – ↑ PaO₂ causing pulmonary vasodilation (↓ PVR)
  – ↑ PaO₂ causing systemic vasoconstriction
  – ↓ levels of circulating prostaglandins
  – Recent research says low pH at birth helps
• Functional closure -
Pathophysiology
• If d.a. doesn’t close as pulm pressures fall & aortic pressure rises -
  - Blood shunted from aorta to PA (L→R)
  - Hyperfusion & engorgement of pulm vessels
  - Hypoperfusion to all postductal organs & tissues
  - Pulm pressure ↑
  - Right heart pressure ↑

Pathophysiology
• If PAP exceeds aortic pressure
  - Shunt switches R→L

Pathophysiology
• PDA is not always undesirable
  - In presence of certain heart defects (Transposition of Great Vessels): PDA may be only connection between systemic & pulmonary circulation
  - To keep PDA open -
Diagnosis

- Most common indication = loud Grade I-
  Grade III systolic murmur heard at upper
  left sternal border
- Positive ID
  
  -
  
  -

Diagnosis

- Oxygen & noninvasive monitoring
  
  - R→L
    
    - Low PaO₂ that does not change with increases in
      FiO₂ (15 mmHg)
    
    - Preductal PaO₂ higher
      than postductal PaO₂

Diagnosis

- Oxygen & noninvasive monitoring
  
  - L→R
    
    - Signs of CHF & pulmonary edema
    
    - CXR - cardiomegaly with
      increased pulmonary
      vascularity
Other

Treatment

• If asymptomatic
  – Fluid restriction
    • <120 ml/kg/day
    • If murmur continues unimproved or deteriorating, then
      • Diuretic therapy

• If infant symptomatic & <1000 g - closure of PDA is required
  – Indomethacin (Indocin)
    • Blocks prostaglandin production → constricts systemic smooth muscle
    • Side-effects
      – Constriction of renal vessels →
      – ↓ in platelet adhesion →
  – Surgery

• If symptomatic & >1000 g
  – Fluid restriction x
  – If worsens or no improvement
    •
    •