Neonatal Surgical Emergencies

James Moore, MD, PhD
Associate Professor of Pediatrics
UT Southwestern Medical Center
Medical Director
NICU Children's Medical Center Dallas, Dallas, TX

The speaker has signed a disclosure form and indicated he has no significant financial interest or relationship with companies or the manufacturer(s) of any commercial product/service that will be discussed as part of this presentation.

Session Summary

During this session multiple neonatal anomalies and conditions that require emergent surgery will be presented in case format and discussed from a presentation/stabilization and management focus.

Session Objectives

Upon completion of this presentation, the participant will be able to:

- discuss CDH in detail, as well as evidence for current treatment;
- understand multiple GI emergencies in case-based format with emphasis on stabilization/treatment;
- recognize airway/pulmonary emergencies and stabilization.

References


Session Outline

See handout on the following pages.
Neonatal Surgical Emergencies

FANNP's 23rd National Neonatal Nurse Practitioner Symposium October 18, 2012

James Moore M.D. Ph.D.
Associate Professor of Pediatrics
UT Southwestern Medical Center
Medical Director NICU: Children’s Medical Center Dallas

Objectives

› To discuss in detail CDH, and evidence for current treatment
› Multiple GI emergencies in case based format with emphasis on stabilization/treatment
› Airway/Pulmonary emergencies and stabilization

Issues in the Management of CDH: Topics that have Biologic Plausibility

› Should we use steroids prenatally?
› Should we use Surfactant after birth?
› Does iNO help with the Pulmonary HTN of CDH?
› What mode and strategy of ventilation should be employed?

Congenital Diaphragmatic Hernia

› Pulmonary hypoplasia
  - Severe on ipsilateral side
  - Variable on contralateral side
  - Immature, abnormal lung
› Reactive and Fixed Component to PHHN
› High mortality up to ≈ 50%
› ECMO survival = 51%

CDH Epidemiology

› CDH occurs 1:3,000 births
› In the US, 1600 newborns/year born with CDH
› 10-30% affected by chromosomal abnormalities
› 40% have associated anomalies (some of which have no affect on survival and others that greatly impact survival)
› Survival is center-dependent (ELSO)
  - Overall CDH survival: 67%
  - CDH survival after ECMO: 51%

CDH Pathophysiology

› Defect or complete absence of a diaphragm
› Displacement of stomach, intestines, liver, and/or spleen into the chest
› Pulmonary Hypoplasia
› Pulmonary Hypertension
  - Fixed (decreased surface area)
  - Reactive (vessel hypertrophy)
    • May respond to low CO2 and high PO2
Antenatal Steroid Use for CDH Infants

Question #1

- Do CDH infants exposed to antenatal steroids compared to placebo have improved
  - Survival?
  - Need for ECMO?
  - # Ventilator Days?
  - Length of Stay?

Biologic Plausibility For Using Steroids

- Nitrofen-induced CDH Rat Model

Assessment of Scientific Evidence

- 10 animal studies (most from same research lab) demonstrate benefit of antenatal steroids on CDH animal models on
  - Surfactant production, pulmonary vascular structure, lung growth, endothelin gene expression, etc.
- 1 small case series of 3 infants showed benefit
- 1 retrospective cohort study showed no difference in mortality, need for ECMO, or pulmonary outcomes between CDH infants
- 1 small RCT of 32 CDH infants found no benefit of antenatal steroids

Therefore the evidence supports: That Antenatal Steroids should not be routinely mandated for all mothers of infants with CDH

Question #2 Should we use Surfactant in patients with CDH?

- Compared to CDH babies that do not receive surfactant, does surfactant:
  - Reduce mortality?
  - Decrease the need for ECMO?
  - Decrease the # of ventilator days?
  - Decrease the length of stay?

Key Studies

- 2 large retrospective reviews show worsen outcome with the use of surfactant
  - Lally et al 2004
    - N=424
    - 209 surfactant treated and 215 non-surfactant treated
    - Results
      - Decreased survival OR: 2.17 (95% CI: 1.5-3.2; p<0.01)
      - Trend toward worse survival if given surf < 1HOL and if immediate respiratory distress
  - Van Meurs 2004
    - N=522
    - 192 surfactant treated and 330 non-surfactant treated
    - Results
      - Increased use of ECMO and CLD in Surfactant treated group
      - Lower survival rate
**Question #3 Should we monitor PPHN in CDH patients?**

- Pulmonary arteries and veins of all sizes from CDH patients demonstrate increases in muscularization and wall thickness.
- Animal CDH models suggest that pulmonary vascular shows abnormal vasoconstrictor response.
- Alveolar capillary cross sectional area of CDH lungs is reduced predisposing to PAH.
- Presence of PAH confers a worse prognosis.

**Key Study**

- Persistence of pulmonary hypertension increases risk for mortality.

<table>
<thead>
<tr>
<th>Week</th>
<th>%</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>60</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>3</td>
<td>36</td>
<td>&lt;.0007</td>
</tr>
<tr>
<td>6</td>
<td>0</td>
<td>&lt;.02</td>
</tr>
</tbody>
</table>


**Key studies**

- Randomized controlled trials show iNO provides no benefit in reducing mortality in CDH infants with refractory hypoxemia respiratory failure OI >25.

**Key studies**

- Randomized controlled trials show iNO may increase need for ECMO in CDH with refractory hypoxemic respiratory failure OI>25.

**PAH Management Recommendations**

- iNO should not be routinely used in patients with CDH.
- iNO should be reserved for possible trial if the patient is failing to stabilize.
- But we have Knowledge Gaps: What is the role of other pulmonary vasodilators (e.g., sildenafil, endothelin-1 receptor antagonists, and EGF) in the treatment of PAH in CDH?

**Question #4 Should We Use Protective lung strategies in CDH**

- Do protective lung strategies (permissive hypercapnea and avoidance of barotrauma and volutrauma) reduce the need for ECMO and improve survival?
Lungs of infants with CDH are hypoplastic and underdeveloped
- Excessive pressure delivered by mechanical ventilation results in barotrauma
- Excessive volume delivery results in volutrauma and air leak
- Management of CDH with focusing on tolerating elevated PCO₂ (permissive hypercapnea) and minimizing barotrauma/volutrauma might improve outcomes

Supporting evidence table

<table>
<thead>
<tr>
<th>Variable</th>
<th>Bagolan (Rome)</th>
<th>Boloker (Columbia)</th>
<th>Finer (San Diego)</th>
<th>Wilson (Boston)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Quality</td>
<td>Mode of ventilation</td>
<td>PIP &gt; 25</td>
<td>CMV Š PIP 20, IT 0.5, PCO₂ &gt; 60 or SaO₂ &lt; 80 Š HPPV or HFOV</td>
<td>CMV → HFOV</td>
</tr>
<tr>
<td>Max PIP</td>
<td>M A P H FO V 20</td>
<td>M A P H FO V 12</td>
<td>M A P H FO V 20</td>
<td>M A P H FO V 12</td>
</tr>
<tr>
<td>PEEP</td>
<td>3-4</td>
<td>5</td>
<td>3-4</td>
<td>5</td>
</tr>
<tr>
<td>pH</td>
<td>PCO₂ &lt; 65</td>
<td>60-65</td>
<td>&lt; 60</td>
<td>60-65</td>
</tr>
<tr>
<td>PO₂</td>
<td>SpO₂ (pre or post)</td>
<td>&gt; 85 (pre)</td>
<td>Pre ductal target 90, tolerated &gt; 80</td>
<td>Pre &gt; 90, post ignored</td>
</tr>
<tr>
<td>Comments</td>
<td>ECMO only if unable to maintain pre ductal SpO₂ &gt; 85</td>
<td>Wung protocol.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Summary Statement
- No randomized control trials evaluating permissive hypercapnea vs. hypocapnea/normocapnea
- All retrospective studies using historical controls found that protective lung strategies improved outcomes relative to historical controls
- Based on preponderance of literature, the use of protective lung strategies should be considered “standard of care”

Knowledge Gaps For “Gentilation”
- No RCT comparing lung protective strategies to traditional ventilator strategies
- Lack of long term neuro-developmental follow-up

Case Study
- You are called to the newborn nursery to evaluate a term infant for non-bilious emesis with feeding attempts.
- When you arrive, you note: increased secretions, and a distended abdomen.
- To prove her point, the nurse attempts to feed the infant, and the infant promptly gives back all the formula it just took in.
- What is your next step? What is your differential diagnosis?
Do you see anything?

Dilated proximal esophageal pouch

Tracheoesophageal Fistula (TEF)

- **EPIDEMIOLOGY**
  - 1:1,000 births
  - 30 - 40% with additional anomalies
  - Can be associated with VACTERL
    - **V** = vertebral (small, hypoplastic vertebrae or hemivertebra)
    - **A** = anal (anal atresia or imperforate anus)
    - **C** = cardiac (VSD)
    - **T** = tracheoesophageal fistula
    - **E** = renal (incomplete formation of one or both kidneys or urologic abnormalities)
    - **R** = limb (radial (absent or displaced thumbs, forearm defects such as radial aplasia))
  - **ETIOLOGY** - abnormal formation during the 4th week of gestation

Associated Anomalies

VACTERL: vertebral, anal, cardiac, TE, renal, limb

- Congenital heart disease is associated with higher mortality
  - VSD is most common
  - ASD
  - Tetralogy of Fallot
  - PDA

- Echocardiogram – important to determine position of aortic arch

Types of TEF

<table>
<thead>
<tr>
<th>Types of TEF</th>
<th>Associated Anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>EA (upper pouch)</strong> with distal TEF just above carina</td>
<td>90% Distended abdomen, Hypersalivation, reflux gastric secretions into lung</td>
</tr>
<tr>
<td>Isolated EA with no tracheal communication</td>
<td>90% Scaphoid abdomen, Hypersalivation</td>
</tr>
<tr>
<td>Isolated TEF (&quot;H&quot; type)</td>
<td>4% Often asymptomatic coughing during feedings, Aspiration pneumonia</td>
</tr>
</tbody>
</table>

Types of TEF

<table>
<thead>
<tr>
<th>Types of TEF</th>
<th>Associated Anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Double TEF, separate attachments with upper and lower esophagus</td>
<td>15% Distended abdomen, Increased salivation into lungs, Potential to reflux gastric secretions into lungs</td>
</tr>
<tr>
<td>Upper pouch fistula</td>
<td>15% Scaphoid abdomen, Hypersalivation, Saliva may enter lungs</td>
</tr>
</tbody>
</table>

Tracheoesophageal Fistula (TEF)

- **CLINICAL PRESENTATION**
  - **Prenatal**
    - fluid-filled stomach not visualized by fetal ultrasound
    - Polyhydramnios (hydramnios) 40%
  - **Postnatal**
    - Excess salivation
    - **Non-bilious** vomiting
    - Feeding intolerance
    - Respiratory distress with feedings
    - Intermittent periods of cyanosis
    - Abdominal distention (if fistula)
Case Study

You are called to evaluate an infant with bilious emesis.
Upon arrival you note that the infant has physical features that are consistent with Trisomy 21.
Green stains on blanket.

Duodenal Atresia

EPIDEMIOLOGY

- 1/10,000 births
- High rate of associated disorders:
  - Trisomy 21 (31%), malrotation (20%), congenital heart disease (30%), esophageal atresia (10%), genitourinary anomalies (11%), annular pancreas (20%)

ETIOLOGY

- Failure of recanalization during the 8th to 10th week of gestation after obliteration of the lumen by epithelial proliferation (6th to 7th weeks’ gestation)
- Usually occurs in the second part of the duodenum
**Jejunal-ileal Atresia**

- **EPIDEMIOLOGY**
  - Overall incidence greater than duodenal or colonic atresias
  - Male = female
  - Usually single atresia (multiple atresias occur in 6% to 20%)
  - Distal ileum: 36%
  - Proximal jejunum: 31%
  - Distal jejunum: 20%
  - Proximal ileum: 13%

- **ETIOLOGY**: In utero mesenteric vascular occlusion

---

**How would you describe this film?**

"cork-screw"

---

**Malrotation**

- **CLINICAL**
  - 2/3 with symptoms in the first month of life
  - If concurrent volvulus, may have bilious emesis, bloody stools, and/or shock

- **DIAGNOSIS**
  - Plain abdominal radiograph: ranges from nonspecific ruminations to complete obstruction with dilated bowel loop(s), air fluid levels; and decreased air distal to the obstruction
  - Barium enema: cecum seen in the right upper quadrant rather than the usual location of the right lower quadrant; corkscrew appearance of the proximal jejunum

- **MANAGEMENT**: Surgery- Ladd’s procedure

---
**What are the findings?**

- Multiple dilated loops of bowel
- No air in distal or rectal region
- Amount of bowel gas suggest lower versus upper obstruction

**Consider dx of lower GI obstruction:**

- Meconium ileus
- Microcolon
- Meconium plug
- Hirschsprung disease
- Colonic atresia
- Anorectal malformations

**Microcolon**

- **ETIOLOGY**
  - Functional immaturity of the ganglion cells
  - Primarily affects the descending and rectosigmoid colon
  - Transient functional obstruction
  - Note: can be a descriptive term as well for obstruction proximal to colon (meconium ileus)

- **EPIDEMIOLOGY**
  - Associated with:
    - Maternal diabetes (most common)
    - Maternal hypothyroidism
    - Maternal toxemia (maternal magnesium exposure)

**Hirschsprung’s Disease**

- **EPIDEMIOLOGY**
  - 1/5,000 births
  - If one child has Hirschsprung’s disease, there is a 3% to 5% risk to next child
  - 1/3 have a relative with Hirschsprung’s disease
  - 80% are male
  - Associations: trisomy 21, heterochromia, Waardenburg syndrome, congenital deafness, 13q deletion, pheochromocytoma, neurofibromatosis, neuroblastoma
Hirschsprung's Disease

- Diverting colostomy proximal to transition zone with definitive treatment at age 1.0 to 1.5 years or weight > 10 kg
- Daily anal dilations followed by laparoscopy-soave pull-through of colon through anus
- Monitor for signs of enterocolitis
- Hearing screen (suggests syndromic etiology, ex. Waardenburg syndrome)

What are the findings?

- Multiple dilated loops of bowel
- No air in distal or rectal region
- Amount of bowel gas suggest lower versus upper obstruction
- Consider dx of lower GI obstruction:
  - Meconium ileus
  - Microcolon
  - Meconium plug
  - Hirschsprung disease
  - Colonic atresia
  - Anorectal malformations

What do you see?

Small colon, so not Hirschsprung

"Filling defects"

Meconium Plug

- Colonic obstruction caused by viscous, congealed mass of meconium (contrast with meconium ileus)
- Either be spontaneously passed or may create an obstruction
- Known association between an underlying diagnosis of cystic fibrosis (CF), Maternal diabetes, and prematurity are known risk factors
- Majority of cases are thought to be caused by an immaturity of the ganglion cells of the colon, leading to a transient impairment of colonic function
Imperforate Anus

- **Epidemiology**
  - 1/5,000 births
  - Includes wide spectrum of anorectal abnormalities (anal stenosis, genitourinary abnormalities)
  - Associated with other anomalies: genitourinary (28% to 50% if high imperforate anus), gastrointestinal (13%), cardiac (7%), skeletal or central nervous system (6%), VACTERL
  - Usually no familial pattern, but there have been reports of autosomal recessive or dominant inheritance

- **Clinical**
  - High lesions and intermediate lesions often without anal opening
  - If rectal atresia: anal opening appears normal
  - Rocker bottom perineum suggests sacral agenesis and has a poor prognosis for fecal continence
  - Urinary reflux if rectourethral fistula
  - In high lesions: 10% tethered cord

Airway/Respiratory Emergencies

- **Neck Masses**
  - Cystic Hygromas
  - Tracheal anomalies

- **Thoracic masses/pulmonary lesions**
  - Congenital lobar emphysema
    - Overdistension of one or more lobes (not histological lung)
  - Congenital cystic adenomatous malformation
    - Multicystic mass of lung tissue, proliferation of bronchial structures at the expense of alveoli
  - Pulmonary agenesis
    - Absence of lung
  - Congenital diaphragmatic hernia
  - Tracheoesophageal fistula

Cystic Hygroma

Persistent cloaca

Rectoperineal fistula
Cystic Hygroma

- Multiloculated cystic spaces lined by endothelial cells
  - Separated by fine walls containing numerous smooth muscle cells
  - Result of maldevelopment of lymphatic spaces

Incidence about 1 in 12,000 births
- 50–65% appear at birth, 85–90% appear by age 2
- Neck~75%, Axilla 20%; can be seen in mediastinum, retroperitoneum, pelvis, groin
- Nuchal/post cervical CH’s have been associated with chromosomal abnormalities—high mortality rate

Complications
- Respiratory—large hygromas can extend into oropharynx and trachea
- Inflammation/Infection
- Hemorrhage

Treatment
- Dependent on size, location, symptoms/complications
- Some pts require emergent surgery due to airway compromise
- Best treatment is complete excision
- Aspiration typically not effective due to rapid refilling of fluid
- Sclerotherapy—Bleomycin, OK-432 (no longer available in US), doxycycline, fibrin glue

Congenital Lobar Emphysema

- Postnatal overdistension of one or more lobes of histologically normal lung
  - Probably due to cartilaginous deficiency in the tracheobronchial tree
  - Obstruction causing the overdistension may be due to
    - 1—chondromalacia of bronchi
    - 2—extrinsic pressure on bronchus by anomalous pulmonary vein or abnormally large PDA
    - 3—idiopathic

Location
- LUL 47%, RML 28%, RUL 20%; lower lobes <5%; Bilat rare

Diagnosis
- Usually can be made by plain CXR; Chest CT and V/P scans may be helpful

Treatment
- May require urgent surgical decompression with lobectomy
- Selective bronchial intubation
- Sometimes see spontaneous resolution—need close observation

Summary

- Evidence for State of the Art CDH care
  - No evidence for Prenatal Steroids
  - No evidence for Surfactant
  - No evidence for iNO, but consider for decompensation
  - Use of Gentilation has improved outcomes

- Review of Many Surgical Emergencies, however can not cover all in 45 min

A11a: NEONATAL SURGICAL EMERGENCIES