Surgical Emergencies in Neonates

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Necrotizing Enterocolitis (NEC)

- Bowel wall necrosis of variable degree
- Perforation in approximately 30% of affected infants
- First described in 1888 by Paltauf
- Named in 1953 by Schmid and Quaiser
Epidemiology

- Primarily affects very low birth weight infants; infants < 1000 g highly susceptible
- Primarily affects premature infants (usually < 32 weeks)
- Incidence in NICU varies from 1-2% (Japan) to 28% (Hong Kong); Incidence in USA is 1-8%
- Overall mortality 10-30%
- Surgical mortality around 20-50% (down from 70%)
- 20-40% of infants with NEC require surgery
- Occurs postnatally only after feeding initiated
Full-term v Premature Infants

• Full-term
  – Presents DOL 1-3
  – Associated with hypoxic/ischemic events (asphyxia, CHD)

• Premature
  – 2-3 weeks after feeding initiated
  – No dramatic cardiorespiratory events
Etiology

• Intestinal hypoperfusion
  – Fetal hypoxia
  – Apnea/bradycardia
  – Arrhythmia
  – PDA/congenital heart disease
• Presence of pathogenic bacteria
• Excess protein in intestines
Diagnosis

• Abdominal distension (most common finding)
• General clinical deterioration usually present
• Feeding intolerance
• Gross or occult blood in stool
• Radiographic findings ranging from bowel edema to free air

Fulminant Course
• Acute onset of GI symptoms
• Coagulopathy/DIC
• CV collapse
Treatment

- **Medical**
  - NPO
  - GI decompression
  - Broad spectrum antibiotics
  - Fluids and transfusion as needed

- **Surgical**
  - Peritoneal drainage
    - <1500 gm
    - Medically unstable

- [Laparotomy with resection and colostomy]
Anesthetic Concerns

- Inhaled agents often poorly tolerated
- Adequate intravenous access
- Ventilatory support
- Avoiding hypothermia
- Volume status
- Metabolic (glucose, electrolyte disturbances)
- May have other associated problems like PDA
- Maintaining intravascular volume and hemoglobin is key
Abdominal Wall Defects

• **Gastroschisis:**
  – herniation of viscera via a lateral abdominal wall defect with no membrane; does not involve umbilicus

• **Omphalocele:**
  – herniation of viscera via a midline abdominal wall defects and covered by a membrane; involves umbilicus
Epidemiology

• Gasteroschisis
  – 1:10,000 live births
  – Isolated defect (<15-20% with other anomalies)
  – May be associated with abnormal GI function (malrotation)

• Omphalocele
  – 1:4000-7000 live births
  – Often (50-70%) associated with other anomalies
  – 20-30% chromosomal anomalies (trisomy 13, 15, 18, 21)
  – Often with normal GI function
Etiology

• **Gastroschisis** – failure of development of lateral abdominal wall

• **Omphalocele** – failure of abdominal contents to return to abdominal cavity during 10\(^\text{th}\) week of gestation
Omphalocele

- Central defect in umbilical ring
- Abdominal contents herniate into a sac
- Umbilical cord inserted into the sac

- Fascial defect > 4cm, can be as large as 10-12cm
- Stomach, intestines, liver (30-50%)
- Associated with other anomalies (Beckwith-Wiedemann, prune belly syndromes; congenital heart disease)
Gastroschisis

- Abdominal wall defect, usually to the right of umbilical cord
- Bowel exposed; matted, thickened, inflamed
- Umbilical cord normal and separate
- Defect around 2-5cm
- Involves small and large intestines
- Usually isolated defect
Diagnosis

- Ultrasound confirms diagnosis in about 95% of cases as early as 10-14 weeks of gestation
- Diagnosis at delivery is usually straightforward except for smallest defects
Treatment

Medical
• Adequate fluid support
• Maintain normothermia and protect viscera
• Decompress stomach
• Treat sepsis
• Assess for other anomalies

Surgery
• Gastrochisis repair is more urgent
• Primary closure (compartment syndrome)
• Staged closure (silastic membrane, spring-loaded silo)
Anesthetic Concerns

- Fluid loss and shifts must be accounted for
- Effects of increased intra-abdominal pressure
  - Respiratory
  - Hemodynamic
  - Renal perfusion
  - Perfusion to lower extremities
- Decompress/suction stomach
- Inhaled (no N\textsubscript{2}O)/IV anesthetics, NMB
- Monitoring (A-line/CVP may be helpful)
- Prolonged postop ventilation may be needed
Pyloric Stenosis

• First described in 1627 by Fabricious Hildanus
• Defined as gastric outlet obstruction due to hypertrophy of pyloric sphincter and overlying mucosa
Epidemiology

- Occurs in approximately 2-5/1000 live births
- Typically manifests between 2 and 6 weeks of age
- Male:female ratio 4:1
- Increased risk in children of affected parents
- Near 0% mortality with appropriate treatment
Etiology

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Diagnosis

- Early diagnosis is key to minimize complications
- Should be suspected in neonates and young infants with nonbilious vomiting
- “Olive” may be felt on physical exam
- Ultrasound has become invaluable for early diagnosis
- Barium swallow is no longer necessary
Diagnostic Criteria

- 4 mm thickness
- 16 mm length
- If both criteria met:
  - 89% sensitivity
  - 100% specificity
Metabolic Derangements

- Loss of acid
- Loss of chloride
- Retention of $\text{HCO}_3$
- Loss of potassium
- Hypoglycemia
- Postoperative apnea?
- Decreased $\text{HCO}_3$ and acidosis in late stages
Treatment

• Medical management is more urgent than surgery
• Hypochloremic alkalosis (in 2/3) and volume depletion must be corrected before surgery
• Hypoglycemia may need to be corrected
• NPO, antispasmodics and slow feedings with clear liquids are occasionally successful on their own
• Surgery with slow resumption of feedings is standard treatment in developed countries
Anesthetic Concerns

- Gastric emptying prior to induction
- Awake intubation versus rapid sequence induction
- High index of suspicion for residual electrolyte/metabolic imbalances
- Analgesia
  - Local infiltration with local anesthetics
  - Non opioid analgesics (acetaminophen)
- Awake extubation
Tracheoesophageal Fistula (TEF)

• Definition
  – Connection between the esophagus and airway
  – May be associated with esophageal atresia or tracheal atresia (rare)
  – May be congenital or acquired

• Associated with other congenital abnormalities, in particular VACTERL
  – Vertebral abnormalities
  – Imperforate anus
  – Congenital heart disease
  – Tracheoesophageal fistula
  – Renal abnormalities
  – Limb abnormalities
Anatomy

Figure 1. Classification of EA/TEF.17,18 Top left, A: EA (Gross classification A, Vogt classification 2, approximate frequency 5%). Top center, B: Proximal TEF with distal EA (Gross classification B, Vogt classification 3A, approximate frequency 0.8%). Top right, C: Distal TEF with proximal EA (Gross classification C, Vogt classification 3B, approximate frequency 88.5%). Bottom left, D: Proximal TEF and distal TEF (Gross classification D, Vogt classification 3C, approximate frequency 1.4%). Bottom right, E: TEF without EA or "H"-type TEF (Gross classification E, approximate frequency 4%).
Epidemiology

- Occurs in 1/3000 live births
- VATER, VACTERL, and other combinations of anomalies (30-50%)
- No increased incidence by gender or ethnic background
- Increased incidence in siblings
- Associated with low birth weight
- Survival approaches 100% but may have long-term complications
Etiology

• Failure of complete separation of foregut and laryngeotracheal structures during 4th and 5th weeks of development
• Exact mechanism is controversial
• Other structures that develop at this time are often abnormal
Diagnosis

• May be cause of polyhydramnios
• Ultrasound may aid prenatal diagnosis (absent stomach bubble)
• Excessive secretions and regurgitation of feeding
• Inability to pass orogastric tube (esophageal atresia)
• Distended/scaphoid abdomen
• Radiography with radio opaque catheter
• Bronchoscopy/esophagoscopy if above studies equivocal or for late presentation
Treatment

- Continuous suction of oropharynx
- Avoid bag-mask ventilation; intubate early
- Treat associated problems
- Gastrostomy tube to vent the stomach
- Central line for parenteral nutrition
- Surgery via thoracotomy as medical condition allows
Anesthetic Concerns

- Airway management is key; may require novel technique (see next slide)
- Inhalational induction and spontaneous ventilation until trachea is secured
- Aspiration and atelectasis are common
- Constant monitoring of tube position to avoid inflating stomach
- Ensure pulmonary inflation via auscultation
- A-line for BP monitoring and blood gases
- Postoperative course depends upon type of repair needed and associated problems (other congenital anomalies, prematurity)
Airway Management

Fig. 3. Placement of the bifurcated tube under fiberoptic guidance. A fiberscope is inserted into the bifurcated tube and advanced into the left main bronchus (a). The tube is then advanced over the fiberscope until the tip splays to sit astride the carina (b).

Anesthesiology, V 100, No 3, Mar 2004
Diaphragmatic Hernia

• May be congenital or acquired
• Defect in the diaphragm allowing abdominal viscera to herniate into the thorax
  – 80% Bochdalek (5:1 L:R)
  – 2% Morgagni (anterior, small)
  – 15-20% Paraesophageal (small, may be asymptomatic)
  – Eventration
• Affects development of both alveoli and pulmonary vasculature
Epidemiology

- Incidence approx. 1/5000 births
- 35% mortality (not counting in utero deaths)
- Estimated cost per case is $250,000; total cost/year in the US is $264,000,000 (1995)
- 40-50% incidence of associated malformations, including cardiac abnormalities
Etiology

- Pleuroperitoneal separation occurs in 9th week of fetal life
- Premature return of abdominal contents may give rise to hernia
- Failure of separation may give rise to hernia
Diagnosis

- May be associated with polyhydramnios
- 50% now diagnosed by ultrasound
- Postnatal findings
  - Cyanosis, respiratory distress
  - Barrel chest/scaphoid abdomen
  - Heart sounds shifted (usually to R)
  - Bowel sounds in chest
- Bowel in chest on x-ray
- Small hernias may go undiagnosed
Treatment

- Medical stabilization is a priority
  - Intubate with minimal bag-mask ventilation
  - Decompress gut
  - Ventilate to maintain $\text{SpO}_2 > 85\%$
  - Assess cardiovascular status; look for persistent fetal circulation
  - May require ECMO

- Surgical management when stable
  - Best timing is unclear
  - Severe hypoxia/acidosis may be contraindication for surgery
  - Outcome depends on degree of pulmonary hypoplasia and associated abnormalities
Anesthetic Concerns

- CVP and art line in addition to usual monitors
- Very sick patients may not tolerate anesthetic drugs other than muscle relaxants
- Nitrous oxide contraindicated
- Minimize peak inspiratory pressures
- Fluid shifts may occur
- Increased intraabdominal pressure may occur after reduction
Meningomyelocele

- Herniation of meninges via bony defect in the spine
- Results in sac containing
  - CSF
  - Dysplastic neural tissue
- May be sacral, lumbar, thoracic, or cervical
Epidemiology

- Incidence in the United States varies from 1-10 per 10,000 births
- Varies with ethnicity
  - Highest in whites
  - Lowest in blacks
- Incidence decreasing with improved prenatal care
- Mortality is low for isolated meningomyelocele
Etiology

• Failure of neural tube closure during 4th week of gestation
• Incidence related to folate deficiency
• May be associated with other birth defects
Diagnosis

• May be diagnosed prenatally
  – Ultrasound
  – Alpha fetoprotein
• Diagnosis usually obvious at birth
Treatment

- Urgent surgery to avoid CNS infections and further damage
- Likely to have long-term problems of varying severity
  - Hydrocephalus
  - Urological problems
  - Paralysis/orthopedic problems
- Intelligence is usually normal
Anesthetic Concerns

- Anesthetic care usually straightforward
- Typically repaired during first few days of life
- Potential fluid/blood loss
- Positioning
- Heat loss