Gastroschisis vs. Omphalocele

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Take Home #1

• Prior to the late 1960’s, survival for infants with gastroschisis was rare.
• Three major advances have led to marked improvement:
  – Parenteral nutrition
  – Silo closure
  – Advances in perinatal (NICU) care
Embryology

- Embryo unfolds in four different directions:
  - Cranial (thoracic & epigastric walls)
  - Caudal (hindgut, bladder, hypogastric wall)
  - Lateral (midgut and lateral abdominal wall)
- Midgut grows rapidly from 6-10 weeks
- Midgut elongates faster than the abdominal wall and herniates through the umbilical ring
Embryology

• Normal development:
  – Midgut rotates and returns into abdominal cavity by week 10-12
  – Allantois, vitelline duct, vessels obliterate
  – Umbilicus with vein and two arteries remains
  – Umbilical ring closes around these structures
Pathogenesis

- Poorly understood
- Depends on timing of embryologic abnormality:
  - The earlier the event, the more complex the anomaly
4-6 Weeks: Body Stalk Defect

- Head
- Spine
- Extremity abnormalities
6-7 Weeks: Gastrochisis

- Believed to be caused by vascular compromise of the right abdominal wall
- Persistence of right umbilical vein or loss of omphalomesenteric artery on right side
7-8 Weeks

- *Pentalogy of Cantrell*
  - Thoracoabdominal ectopia cordis
  - Cardiac defects
  - Omphalocele
  - Cleft sternum
  - Diaphragmatic, pericardial defects

Figure 9: The fetus (*Pentalogy of Cantrell*).
7-8 Weeks

- **Cloacal Exstrophy**
  - Extrophy of the bladder
  - Imperforate anus
  - Omphalocele
  - Lower neural tube defects
7-8 Weeks

• Omphalocele
  – Improper migration of fusion of the lateral folds resulting in failure of closure of the umbilical ring = persistent herniation of midgut
  – Umbilical vessels insert on to the sac and radiate around it (important … remember this)
Etiology of Defects

- Younger mothers <20
- Folic acid deficiency
- Hypoxia
- Salicylates, acetaminophen, ibuprofen, pseudoephedrine
- Marijuana, cocaine, alcohol
Genetics

**Gastroschisis**
- Hirschsprung’s disease
- Oromandibular limb hypogenesis
- (sporadic)

**Omphalocele**
- Trisomy 13
- Trisomy 14
- Trisomy 15
- Trisomy 18
- Trisomy 21
Incidence

**Gastroschisis**
- 1 in 6,000
- Male = female
- Incidence increasing

**Omphalocele**
- 1 in 4,000
- Male = female
- Incidence is static
Take Home #2

• The physiologic impact of omphalocele is related to its size and associated anomalies:
  ➔GI FUNCTION IS NORMAL! ⇐

• In gastroschisis the bowel does not function properly but the child rarely has other serious issues.
# Remember This Slide!

<table>
<thead>
<tr>
<th></th>
<th>Gastrochisis</th>
<th>Omphalocele</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Defect</strong></td>
<td>Open</td>
<td>Membrane covered</td>
</tr>
<tr>
<td><strong>Defect size</strong></td>
<td>2-5 cm</td>
<td>2-15 cm</td>
</tr>
<tr>
<td><strong>Umbilical cord</strong></td>
<td>Left of defect</td>
<td>Center of membrane</td>
</tr>
<tr>
<td><strong>Bowel</strong></td>
<td>Inflamed</td>
<td>Normal</td>
</tr>
<tr>
<td><strong>Alimentation</strong></td>
<td>Delayed</td>
<td>Normal</td>
</tr>
<tr>
<td><strong>Anomalies</strong></td>
<td>10%</td>
<td>60%</td>
</tr>
</tbody>
</table>
Associated Conditions

**Omphalocele**
- Cardiac
- Chromosomal
- Down’s syndrome
- Macrosomia
- GERD
- Cryptorchidism
- Musculoskeletal
- Neural tube defects

**Gastroschisis**
- Intestinal atresia
- Small for gestational age
- Prematurity
- GERD
- Cryptorchidism
Gastroschisis

- Small defect (usually less than 4 cm)
- Occurs to the right of the umbilicus
- Normal abdominal wall and musculature
- No sac or remnant of a sac!
- Midgut herniated through defect
  - Possibly duodenum, stomach, colon or gonad
Associated Conditions

- Intestinal atresia 10%
- GERD 16%
- Undescended testicle 15%
  - Usually corrects spontaneously
Initial Care of Gastrochisis

• Premature patient
  – Heat preservation
  – Respiratory support
  – Large surface area of exposed intestine
  – Increase fluid loss

• Place patient in bowel bag
  – Controls heat & evaporative loss
  – Allows monitoring of bowel
Initial Care of Gastroschisis

• Goal: Primary reduction if possible!
• Cesarean delivery is not mandatory but allows team to be assembled
• If no respiratory distress is noted:
  – Start IV line
  – Place monitoring equipment
  – Place NGT (evacuate stomach)
  – Intubate then paralyze patient
Gastroschisis Pearls

• Close primarily if possible
• Silo reduction is good alternative
• Don’t make the closure too tight
• Wait to feed the patient (14 days)
• Slow advances on feeds
• Expect high residuals and bilious aspirates
• Rectal stimulation
Postop Problems

- Necrotizing Enterocolitis
- Short gut syndrome
- Malabsorption
- Obstruction
- TPN-associated cholestasis
- Developmental delay
High Risk Gastrochisis

• The following factors are associated with a more complicated post-operative course:
  – Intestinal perforation
  – Atresia
  – Necrotic Segments
  – Volvulus

• Characterized by: prolonged ileus & time to tolerate feeds, longer length of stay, increased infectious complication & mortality.
Omphalocele
Omphalocele

• At birth look for other anomalies
  – Echocardiogram
  – CXR
  – Renal ultrasound

• Other more severe associated anomalies take precedence over surgical repair
  – Exception: disruption of the sac
Omphalocele

• If severe pulmonary hypoplasia is present:
  – Closure of abdomen may be impossible
  – Treat with topical bacteriocidal agent
    • Wait for epithelialization
    • Fix hernia at later date
  – Current agents used (some toxicity):
    • Silvadene
    • Betadine
    • Triiodomethane-petrolatum gauze
Small Omphalocele

- Usually closed primarily
- Very similar to gastroschisis closure
- Watch abdominal pressure
- Ligate umbilical vessels
- Neoumbilicus can be created
Large/Giant Omphalocele

• Options include:
  – Short-term silo reduction (2 weeks)
    • Followed by closure with mesh
  – Long-term silo reduction (2-6 weeks)
  – Staged closure
    • Close skin flaps over amnion
    • Fix hernia at 6-12 months of age
Postop Management

• Respiratory support is critical
  – Intubation with paralysis

• Nutritional support
  – TPN initially

• Meticulous wound care
  – Prophylactic antibiotics
  – Watch for cellulitis, fascitis
REMEMBER …

• Omphalocele has normal GI function but bad associated anomalies/defects … survival about 60%

• Gastroschisis has horrible GI function but few associated anomalies/defects … survival about 85-90%