Abdominal Wall Defects

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Case Presentation

HX: Pt is a 36 week gestational age male born via C-section delivery with a dx of gastroschisis made on a prenatal ultrasound at 15 weeks.

Maternal Hx: xx year old female G 0010, denies any medical problems.

Meds: Tylenol for pain. Denies aspirin, ibuprofen or any OTC meds. Denies smoking or illicit drug use.
Case Presentation

PE:
Vitals: T 99 P 140 RR 45
CVS: S1, S2 no murmur
Lungs: CTA B/L
Abd: Soft, 4 cm defect to the right of the umbilical cord containing bowel. The bowel is thick, matted and edematous.
Ext: no deformity
Hospital Course

- Delivered via C-section
- Intubated with NGT Decompression
- Saline soaked gauze wrapped around intestine.
- OR for placement of spring loaded silo
- Taken to NICU
  - IV Abx
  - IVF
  - Incubator
Hospital Course

- **DOL#1** Surgical placement of spring loaded silo
- **DOL#1-8** Placed in incubator, TPN, bowel reduction, mildly sedated
- **DOL#9-11** Paralyzed baby. Freed up filmy adhesion of bowel to fascia
- **DOL#12** Taken to OR. Silo removed and fascia closed with silastic patch.
Hospital Course

- DOL#44 Taken back to OR for removal of mesh and closure of skin

Currently in NICU slowly tolerating feeds
Gastroschisis
Gastroschisis

**Definition:**

- Deformity caused by involution of secondary umbilical vein and results in a full thickness defect of the abdominal wall to the right of the umbilical cord.

- Herniation of small bowel and large bowel

- The loops of intestine lie uncovered in the amniotic fluid and become thickened, edematous and matted.
Gastroschisis

- Incidence 1 in 10,000
- Infrequent congenital malformations
- High association with prematurity
- Herniated contents rarely liver
- Associated cryptorchidism – 30%
- Intestinal atresia bowel common – 10%
Risk Factors

- 4X more common in women < 20 years of age
- Smoking
- Stressed and undernourished mothers
- Over the counter meds: Vasoactive properties pseudoephedrine, aspirin, ephedrine
- Multifactorial
Main Consequences of Gastroschisis

- Perivisceritis
- Short Gut Syndrome
- Intestinal Atresia
Prevalence of Gastroschisis in the US

Prevalence is calculated by dividing the number of affected people by the total population.

Data from the Metropolitan Atlanta Congenital Defects Program, 2004.
Gastroschisis by Race

Gastroschisis by Maternal Age

Prevalence per 10,000

Diagnosis of Gastroschisis

**Before Birth**
- Excess amniotic fluid
- Ultrasound
- Maternal serum alpha-fetoprotein levels elevated

**After Birth**
- Visible
Fetal Ultrasound

Fetal ultrasound showing bowel protruding from abdominal wall defect.
Treatment

• Immediately after birth exposed part it wrapped in sterile saline soaked gauze.

• Tx: plastic “silo” slowly compressed over the next week

• Surgical closure of the defect.

• Infants are feed TPN for 2-5 weeks until normal bowel function begins.
Postnatal Care

- Consideration for third-space fluid losses
- Nutrition - albumin
- Risk of Infection - Antibiotics
- Intestine often thickened:
  - Damage form amniotic fluid
  - Ischemia from constricting wall defect
- Incubator - temperature control
- Respiratory Compromise
Surgical Options for Treatment

- Primary Closure
  - +/- Mesh

- Staged closure with
  - Spring loaded Silo
  - Dacron reinforced Silastic Sheet- Sutured to medial aspect of rectus fascia
Silo Placement

Silastic silo to conserve heat and prevent infection.
Surgical Management for Intestinal Atresia

- Intestinal Atresia
  - Bowel placed into abdomen and plan for reoperation
  - Distal atresia perform a proximal diverting stoma
  - Proximal atresia- Nasogastric Decompression
  - Primary Anastomosis- not advised
Complications

- Deceased venous return
- Abdominal Compartment Syndrome
- Decreased Pulmonary Compliance
- Renal Failure
- Necrotizing Enterocolitis
“The Gentle Touch”

- Treated 52 babies with gastroschisis
- Used “gentle touch” approach vs manual reduction

- Fascial Closure 5.5 days vs 7 days
- Feeding 11 to 24 days vs 12 to 30 days
- TPN cessation in 23 days vs

- Conclusions: Gravity reduction of intestine in babies with gastroschisis is both gentle and effective

“Gentle Touch” Protocol

- Vaginal Delivery and ET Intubation
- Gastric and Colonic Decompression
- Broad Spectrum IV Abx
- Sedation
- Placement of Silo
- Gravity Based Bowel reduction
- Delayed Primary Fascial Closure
- TPN until bowel function returns

Omphalocele
Omphalocele

- Incidence 2.5 in 10,000

- Results from failure of normal embryonic regression of the mid-gut from the umbilical stalk into the abdominal cavity.

- Can include intestines, liver or spleen covered by a sac of parietal peritoneum and amnion which can rupture

- Herniates into the base of the umbilical cord.
Embryology

- 6\textsuperscript{th} wk – midgut loop elongates and herniates out through umbilical cord
- Midgut rotates 270 degrees
- Returns to abdomen by 10\textsuperscript{th} wk
- Anterior abdominal wall progressively closes leaving only umbilical ring
Etiology

Three Theories:

1. Persistence of the primitive body stalk
2. Failure of the bowel to return to the abdomen,
3. Failure of complete lateral-body fold migration and body wall closure
Omphalocele

- Associated Abnormalities in 60%
- Cardiac, Renal, Limb and facial anomalies
- Genetic Syndromes- Pentalogy of Cantrell, Beckwith-Wiedemann
- Associated with Trisomy 13, 14, 15, and 18
- Small Omphalocele without liver-
  
  Chromosomal abnormalities
Diagnosis

- Ultrasound
- Maternal Serum AFP
- Most omphaloceles occur sporadically
- Small Omphalocele < 5cm
- Large Omphalocele >5cm:
  - Pulmonary Hypoplasia -
  - High mortality
Evaluation

- An amniocentesis for karyotype is performed on all fetuses with omphalocele

- Search for other anomalies takes priority over repair of omphalocele

- Chest xray, echo, renal US

- Pulmonary Hypoplasia may preclude closure
ASSOCIATED MALFORMATIONS
With Omphalocele

- Upper Midline Syndrome
- Lower Midline Syndrome
- Beckwith-Wiedemann Syndrome
UPPER MIDLINE SYNDROME

- Pentalogy of Cantrell
- Sternal defect
- Ectopia cordis
- Pericardial and cardiac defects
- Diaphragmatic defect
- Omphalocele
LOWER MIDLINE SYNDROME

- Vesiculointestinal fistula
- Imperforate anus
- Colonic agenesis
- Bladder extrophy
- Omphalocele
Beckwith-Wiedemann Syndrome

Clinical Features

Somatic Gigantism
Hemihyperplasia
Macroglossia
Visceromegaly
Omphalocele
Ear creases/pits
Hypoglycemia
Tumours
Surgical Repair

- Small defects (<2 cm) can generally be managed by primary direct closure.

- Medium to large defects require a staged procedure.

- Primary closure reduces the risk of bacterial contamination, sepsis, acidosis, and hypothermia.
For moderate to large defects (2 to 9 cm) place a Dacron-reinforced silastic silo as a temporary cover for the bowel.

The silo can be reduced gradually over 3 to 7 days in the intensive care unit, after which the infant is returned to the operating room for final closure of the abdominal wall.
- Reducing a large defect or one that contains liver can be done with Doppler ultrasound guidance.

- Escharification:
  extremely large (>10 cm) lesion or a premature infant having respiratory difficulties, a topical sclerosing agent can be used as a temporary measure until definitive therapy can be performed on a more stable patient.
Prognosis

- **Gastroschisis**: Overall survival is 90%
  
  Low survival if associated with intestinal atresia

- **Omphalocele**: Mortality increased if associated with chromosome syndrome or cardiac defect.

  Giant Omphalocele associated with pulmonary hypoplasia: worse prognosis
## Conclusions

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<thead>
<tr>
<th>Defect</th>
<th>Gastrochisis</th>
<th>Omphalocele</th>
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</thead>
<tbody>
<tr>
<td>Defect Size</td>
<td>2-5cm</td>
<td>1-15cm</td>
</tr>
<tr>
<td>Umbilical Cord</td>
<td>Left of defect</td>
<td>Center of membrane</td>
</tr>
<tr>
<td>Bowel</td>
<td>Serositis, edema</td>
<td>Normal</td>
</tr>
<tr>
<td>Alimentation</td>
<td>Delayed</td>
<td>Normal</td>
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<tr>
<td>Assoc Anomalies</td>
<td>10%</td>
<td>60%</td>
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