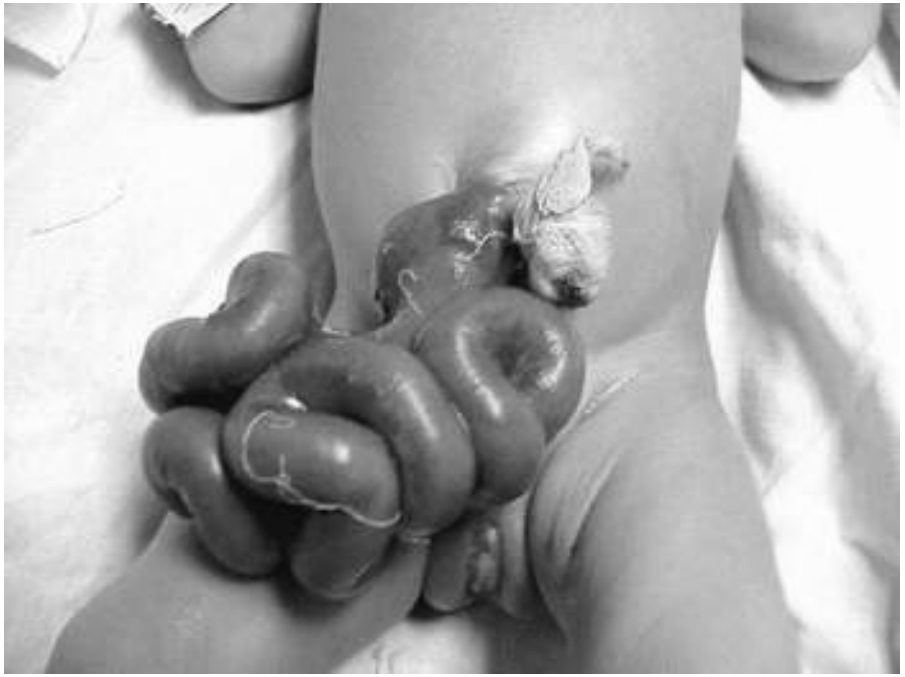
Abdominal Wall Defects



Sajani Shah MD
M&M Conference
SUNY Downstate
Feb 24, 2006

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.

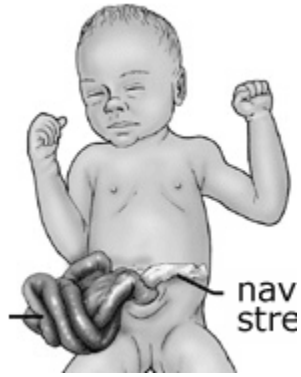
Hospital Course

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

Currently in NICU slowly tolerating feeds

Gastroschisis

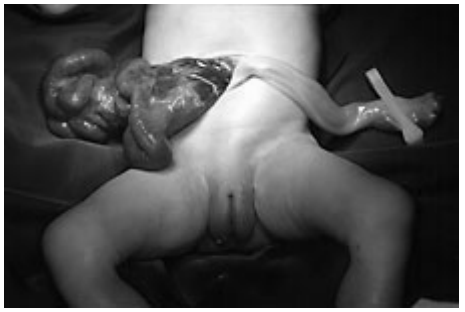
Gastroschisis



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



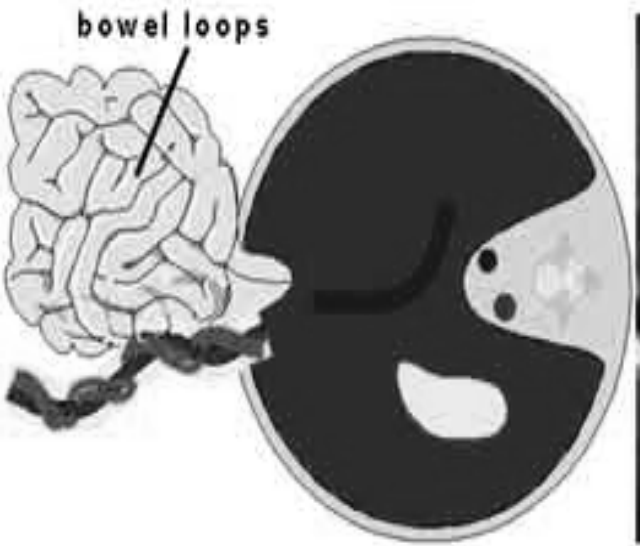
- Incidence 1 in 10,000
- Infrequent congenital malformations
- High association prematurity
- Herniated contents rarely liver
- Associated cryptorchidism – 30%
- Intestinal atresia bowel common- 10%

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

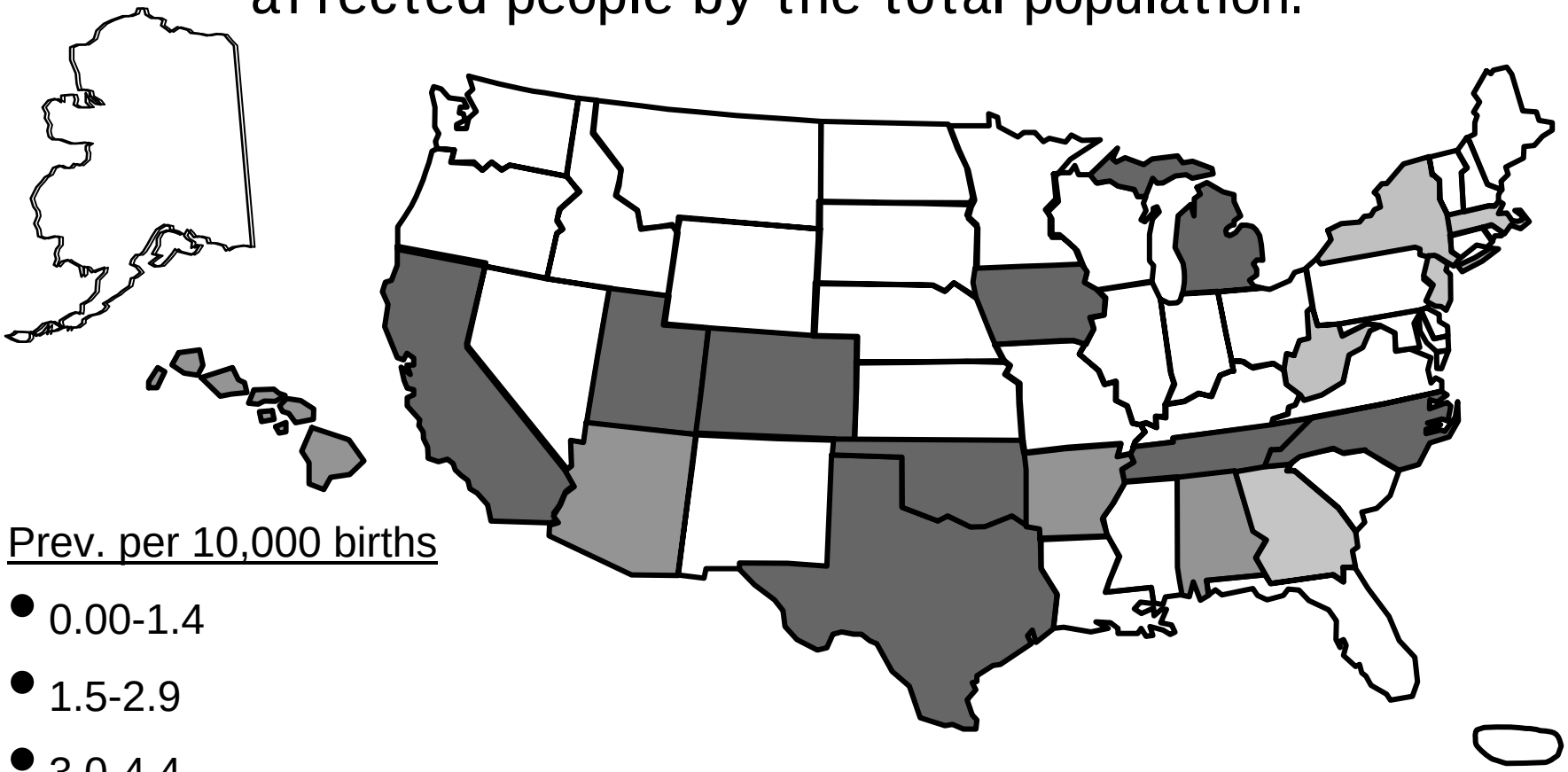
Main Consequences of Gastroschisis

- 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.

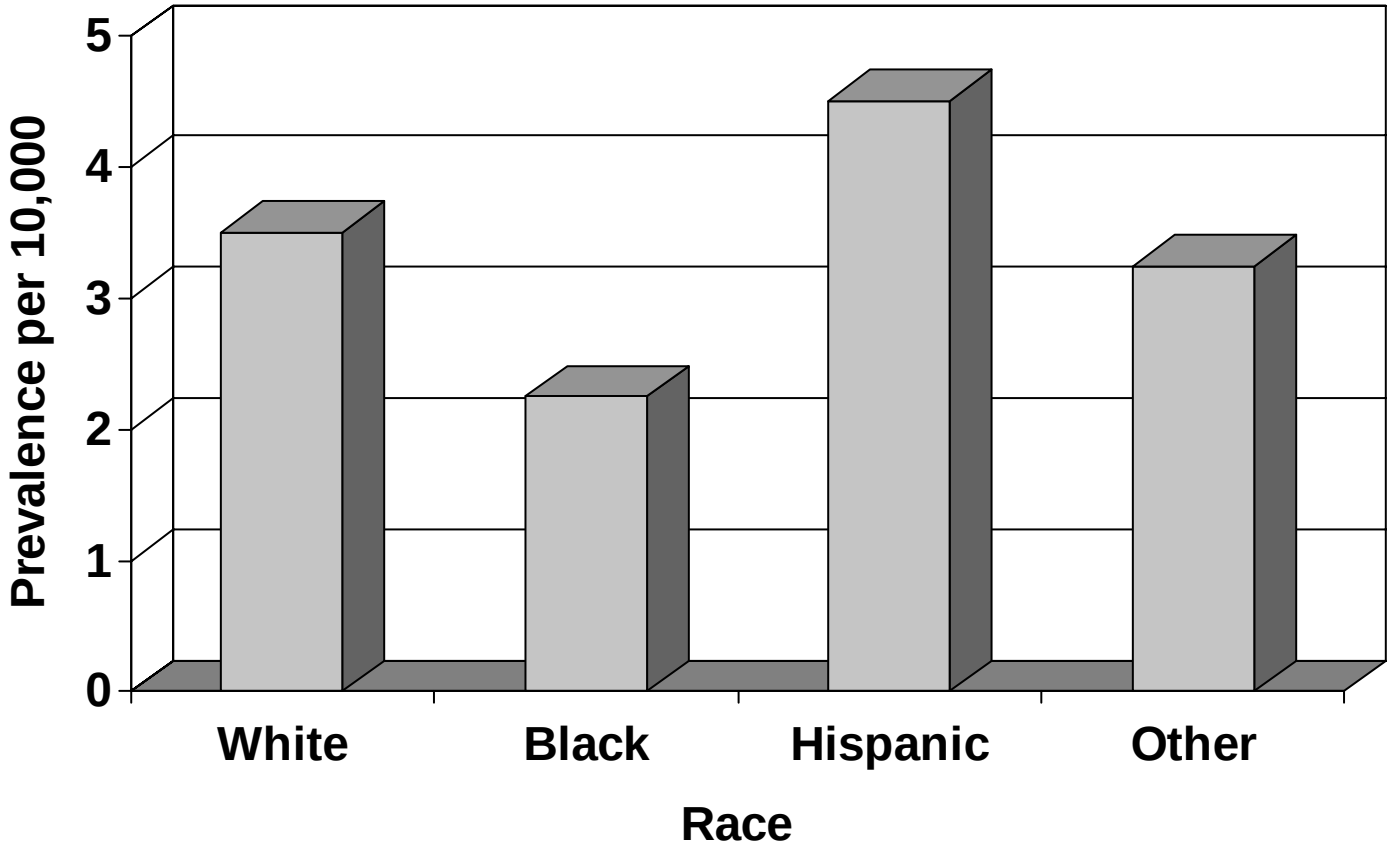


Prev. per 10,000 births

- 0.00-1.4
- 1.5-2.9
- 3.0-4.4
- 4.5+

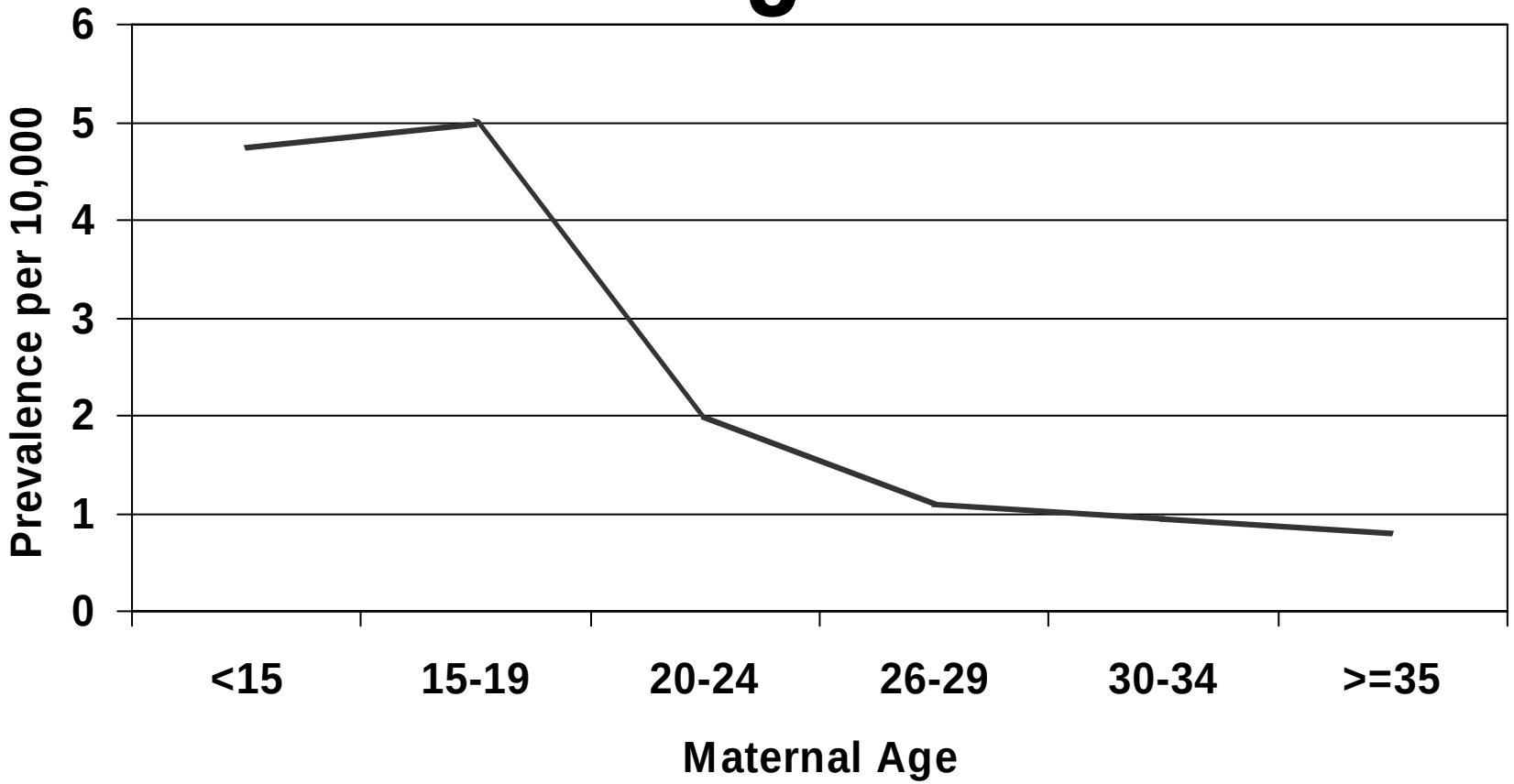
Data from the Metropolitan Atlanta Congenital Defects Program, 2004.

Gastroschisis by Race



Source: 2003 Congenital Malformations Surveillance Report: A Report from the National Birth Defects Prevention Network. Birth Defects Research (Part A): Clinical and Molecular Teratology. New York: Wiley-Liss; 2003.

Gastroschisis by Maternal Age



Source: The Metropolitan Atlanta Congenital Defects Program, 2004.

Diagnosis of Gastroschisis

Before Birth

Excess amniotic fluid

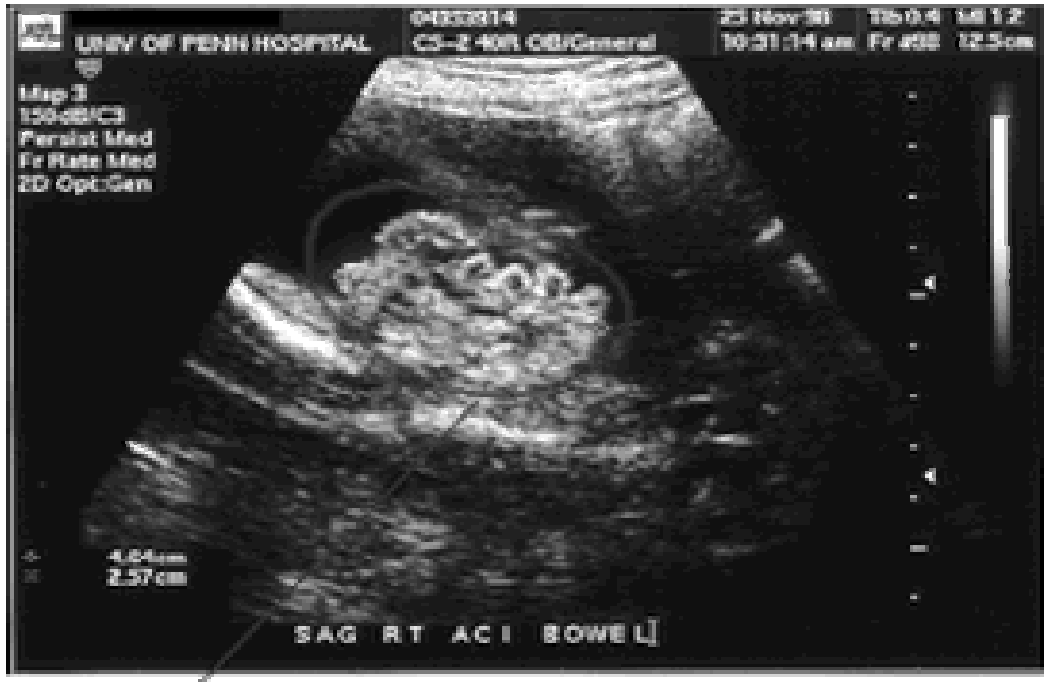
Ultrasound

Maternal serum alpha-fetoprotein levels elevated

After Birth

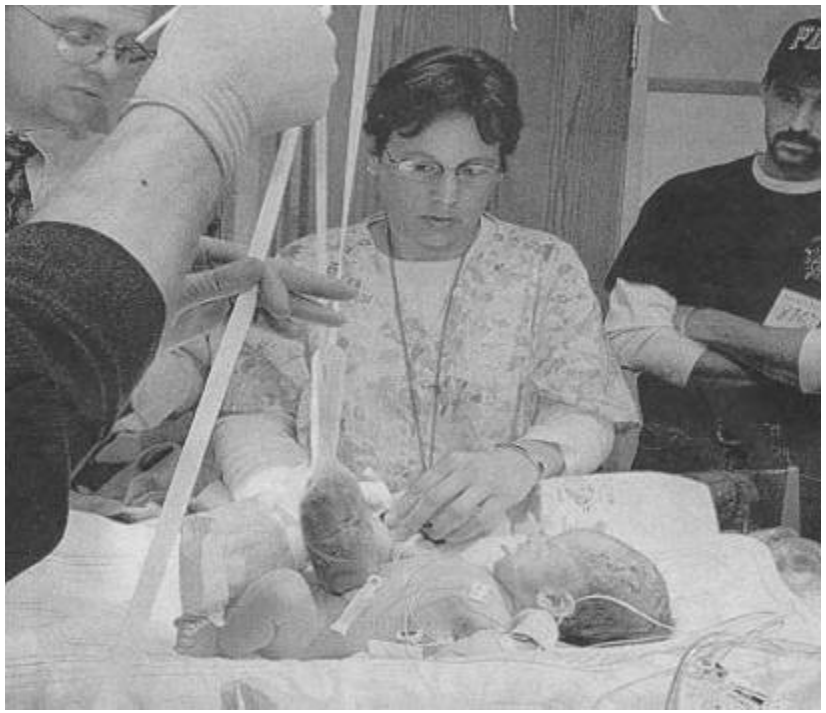
Visible

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**

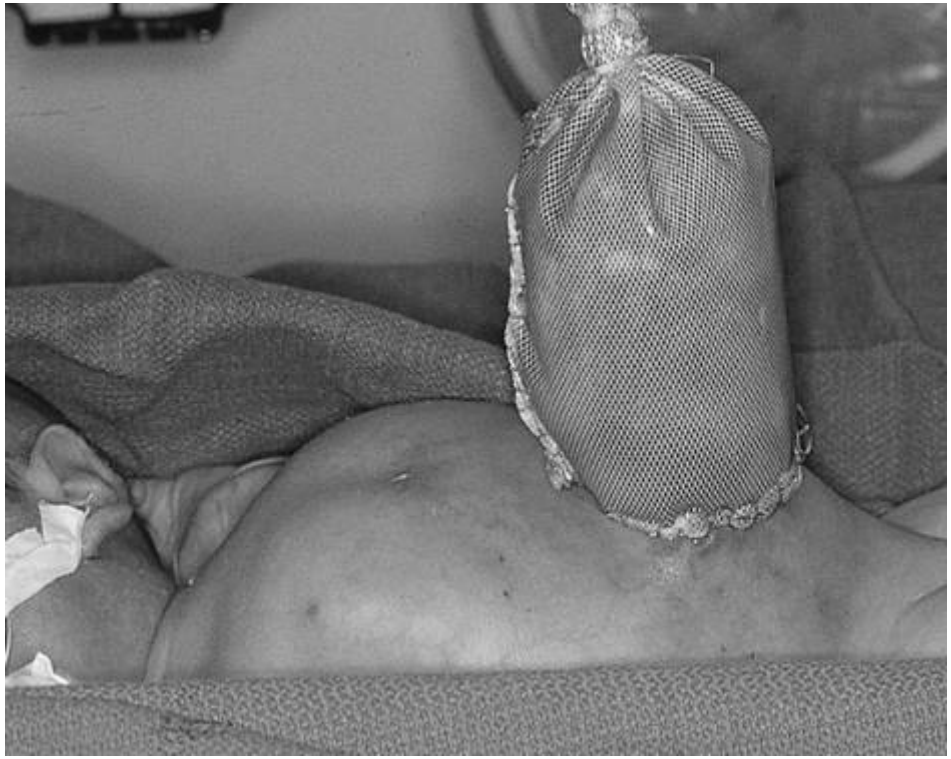
Postnatal Care

- 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

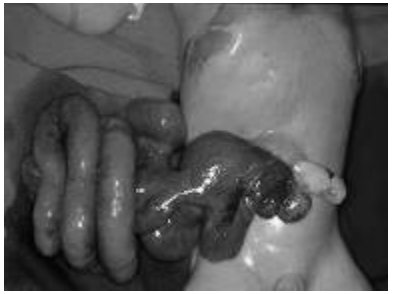
- Primary Closure
  - +/- Mesh
- Staged closure with
  - Spring loaded Silo
  - Dacron reinforced Silastic Sheet- Sutured to medial aspect of rectus fascia

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

Jona JZ. The 'gentle touch' technique in the treatment of gastroschisis. J Pediatric Surg 2003; 38:1036

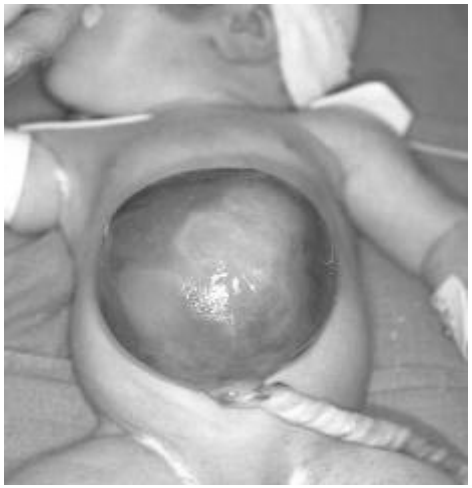
"Gentle Touch" Protocol

- 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

Jona JZ. The 'gentle touch' technique in the treatment of gastroschisis. J Pediatric Surg 2003; 38:1036

Omphalocele

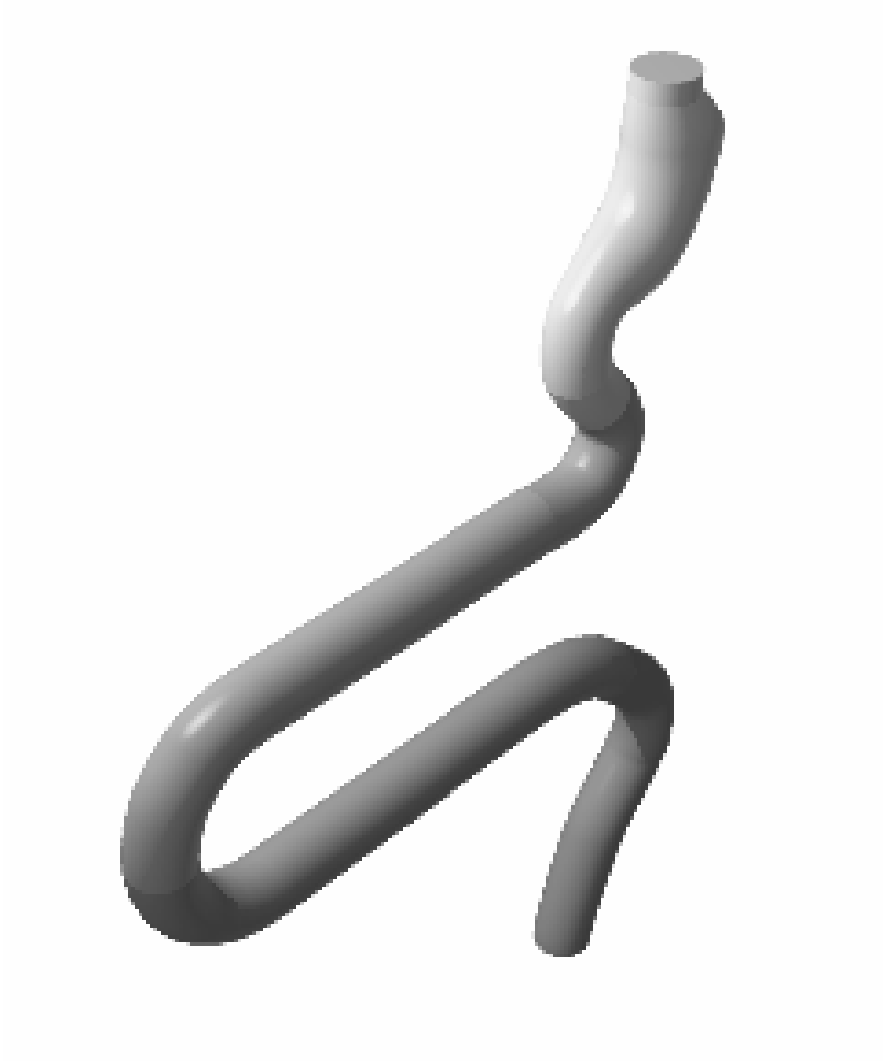
Omphalocele



- 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

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



Etiology

Three Theories:

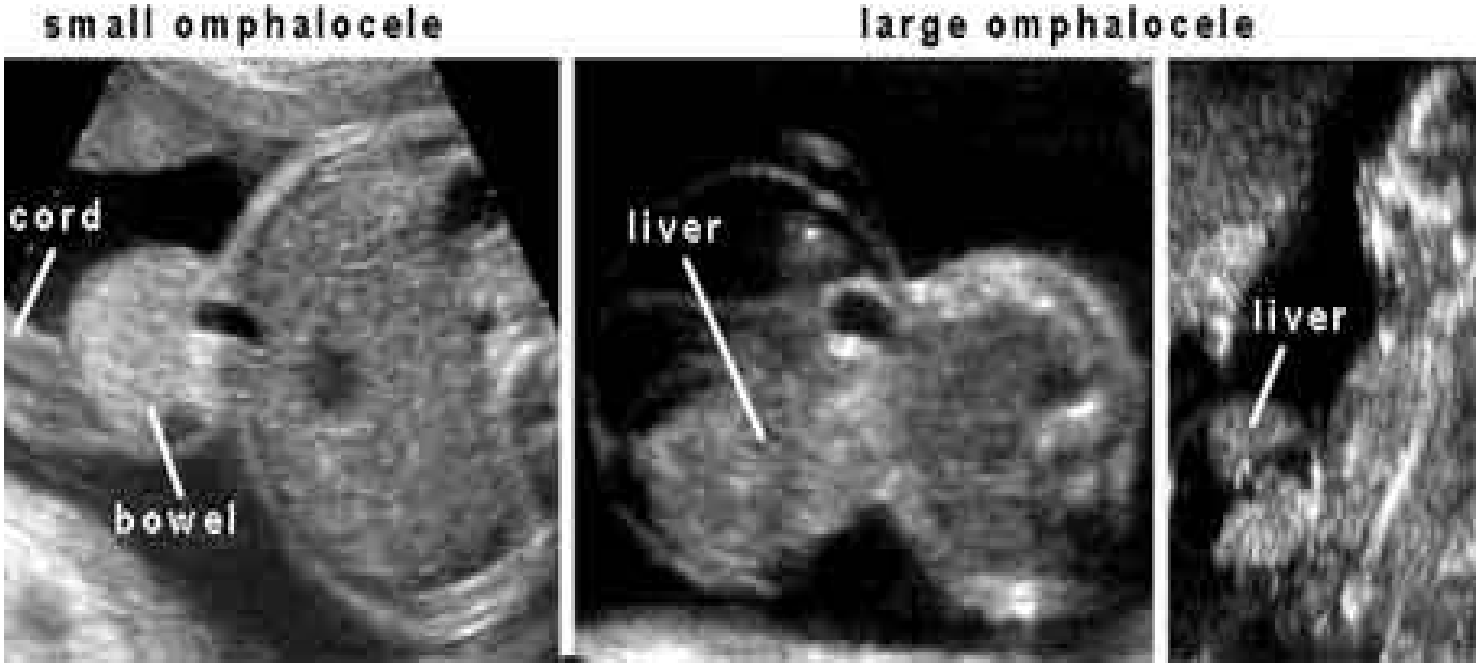
1. Persistence of the primitive body stalk
2. Failure of the bowel to return to the abdomen,
3. Failure of complete lateral-body fold migration and body wall closure

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 ompahlocele
- 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

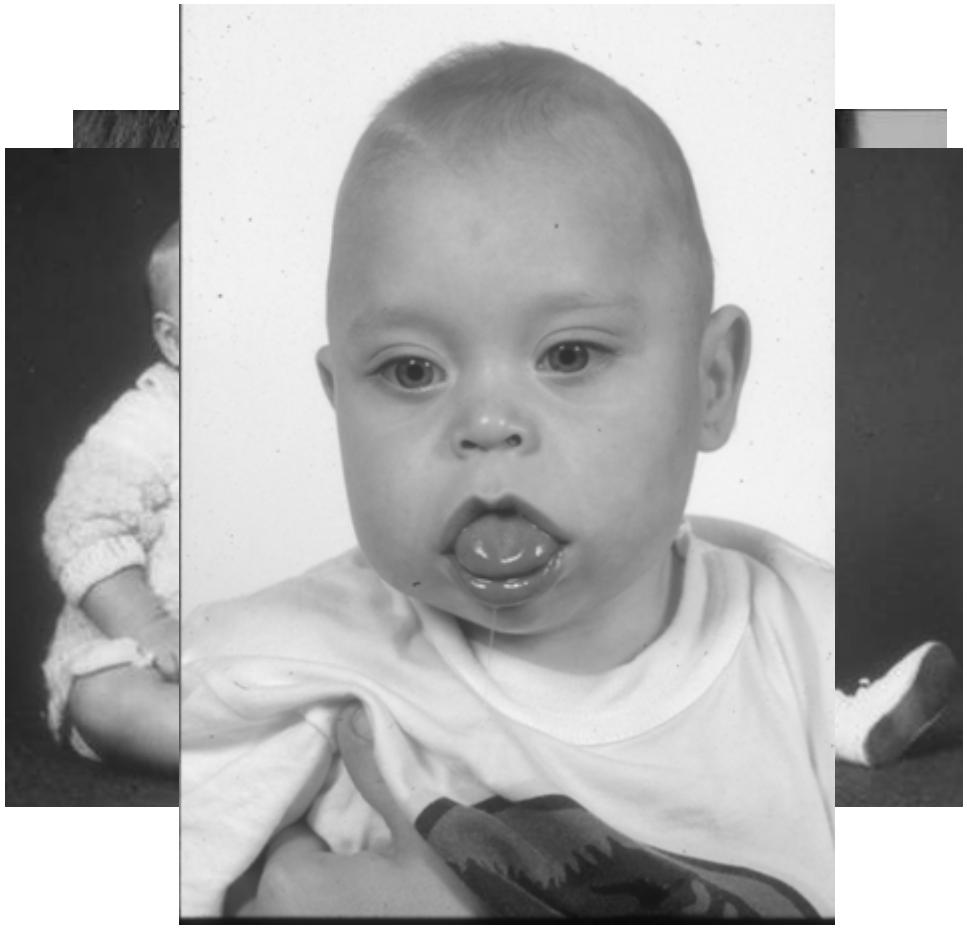
LOWER MIDLINE SYNDROME

- 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

Surgical Repair

- 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

	Gastroschisis	**Omphalocele**
Defect	Open	Membrane Covered
Defect Size	2-5cm	1-15cm
Umbilical Cord	Left of defect	Center of membrane
Bowel	Serositis, edema	Normal
Alimentation	Delayed	Normal
Assoc Anomalies	10%	60%

The End