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Congenital Abdominal Wall Defects

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History

- 1 day old F ex-40 week gestation born via vaginal delivery
- Mother: 31 years old, G5P1
  - PMHx: multiple abortions
  - Denies smoking, drug use
- Fetal U/S @ 23 weeks: suspicious for umbilical cyst
  - No follow up studies obtained
- Apgar score 9/9

Physical Exam

- 99.7 BP 72/37 HR 160; birth weight 3208 grams
- No acute distress
- S1/ S2 RR
- Equal breath sounds b/l
- Abd: midline defect at umbilicus containing viscera c sac
- Genitourinary structures and anus grossly normal

Omphalocele

Imaging

- Upon arrival of pediatric team, omphalocele ruptured
- Pediatric surgery was called
  - Echo: small PDA, normal function
  - Renal U/S: negative
- Taken to OR urgently for exploration

Intraoperative Findings

- Ruptured omphalocele with everted ruptured bladder
- Partial cystectomy and two layer repair
- No other findings
- Primary fascial closure achieved
- Patient returned to NICU intubated

Postoperative Course

- Extubated POD#2
- Voiding freely
- Started on feeds POD#4, advanced as tolerated
- Patient was discharged home on POD#7

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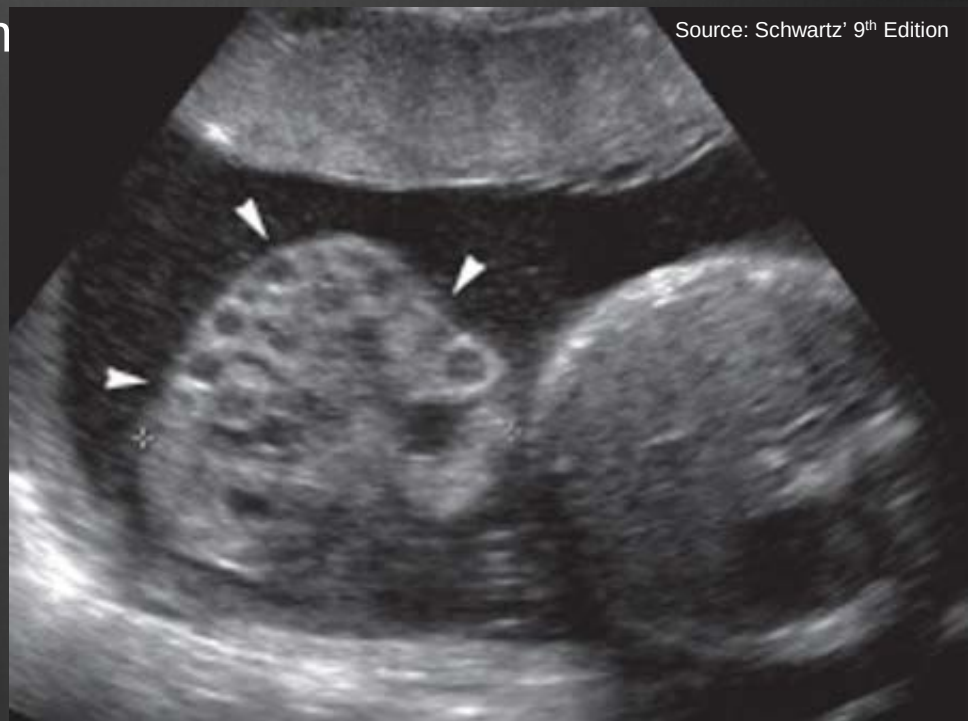
History

- 2 day old male, ex-38 week gestation born via c-section
  - Mother 24 years old, G4P2
  - PMHx: obesity, PCOS, fibromyalgia and anxiety
  - Meds: Cymbalta, Xanax prn
  - Denies smoking history
- Fetal U/S @ 25 weeks
- APGAR scores 4 & 7



Source: Schwartz' 9th Edition

Physical Exam

- Afebrile
- Intubated, equal breath sounds b/l
- S1/S2 RR
- Abd: bowel herniated through defect right of umbilicus
- Bowel matted, leathery in appearance

Gastroschisis

Preoperative Workup

- Echo: normal function
- Renal U/S: no abnormalities noted
- PICC placed in NICU
- TPN started DOL 2
- Taken to OR for exploration on DOL 2

Intraoperative Findings

- Ex-lap via upper midline longitudinal incision
- Bowel inspected, no malrotation, anomalies encountered
- Reduced contents & closed original defect circular fashion
- No excessive tension noted on closure
- Taken to NICU intubated

Postoperative Course

- Extubated POD#4
- POD# 10 Started on oral 10% dextrose solution
- Return of bowel function
- POD# 18 half-strength elemental feeds started
- Remains in NICU tolerating feeds

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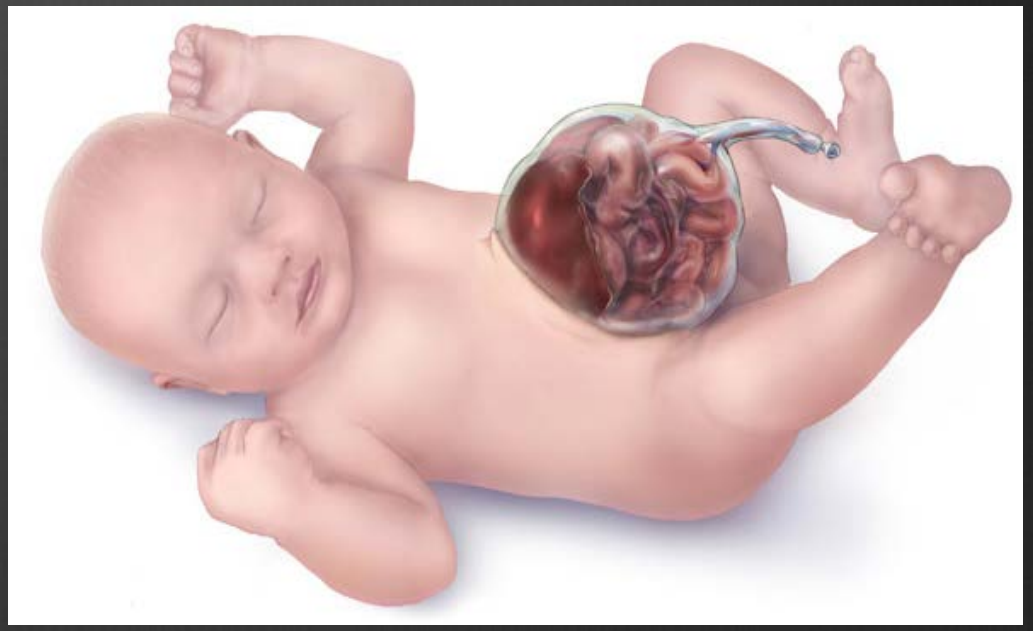
Congenital Defects

- Omphalocele
- Gastroschisis
- Umbilical cord hernia
- Ectopia cordis thoracis
- Cloacal Exstrophy
- Patent Urachus



Embryology

- Craniocaudal & mediolateral infolding at 4th week
- Lateral folds meet at midline constricting yolk sac
- By 6th week, intestinal growth leads to herniation
- By week 10 midgut returns to abdomen; bowel fixation
- Closure of the abdominal ring
- Failure leads to abdominal wall defects

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Omphalocele

- Incidence of 1 per 6-10,000, male predominance
- Results from incomplete closure of abdominal wall
- Central defect covered by sac (midgut & abd organs)
- 30-70% incidence of associated anomalies
- Large for gestational age (> 4 kg)

Risk Factors

- Male sex
- Twin or higher multiple order gestations
- Increased prevalence among Black mothers
- Both young and advanced maternal age
- Alcohol consumption in first trimester

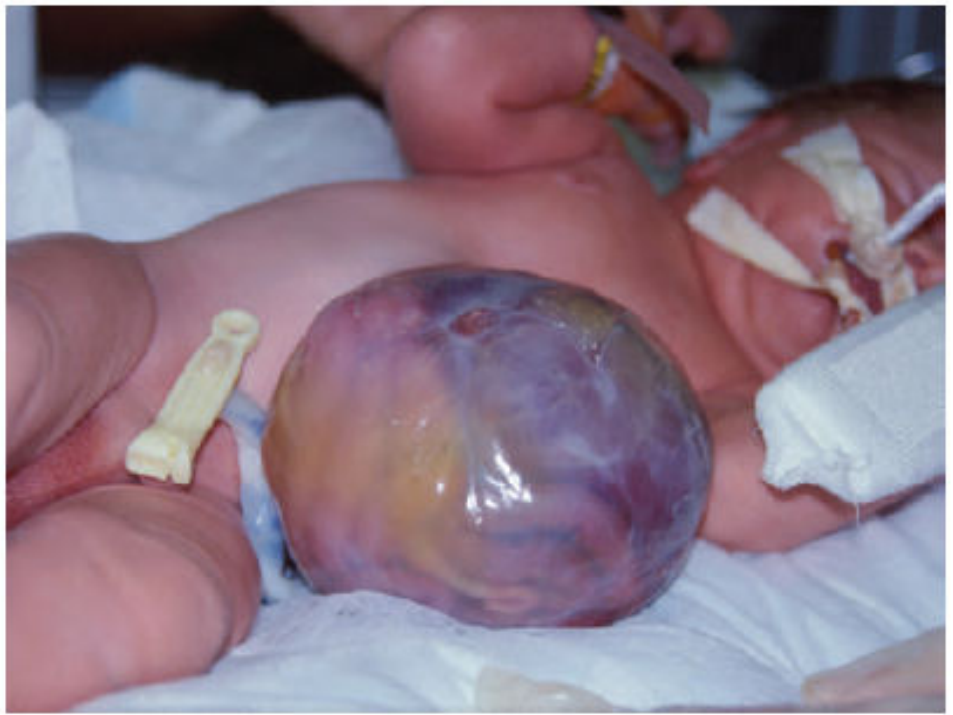
Mac Bird, T et al. Demographic and environmental risk factors for gastroschisis and omphalocele in the National Birth Defects Prevention Study, J Pediatr Surg 2009

Associated Abnormalities

- Chromosomal abnormalities in ~50% (trisomy 13, 18, 21)
- Cardiac abnormalities (ASD/VSD, coarctation of aorta)
- Pulmonary hypoplasia, respiratory distress

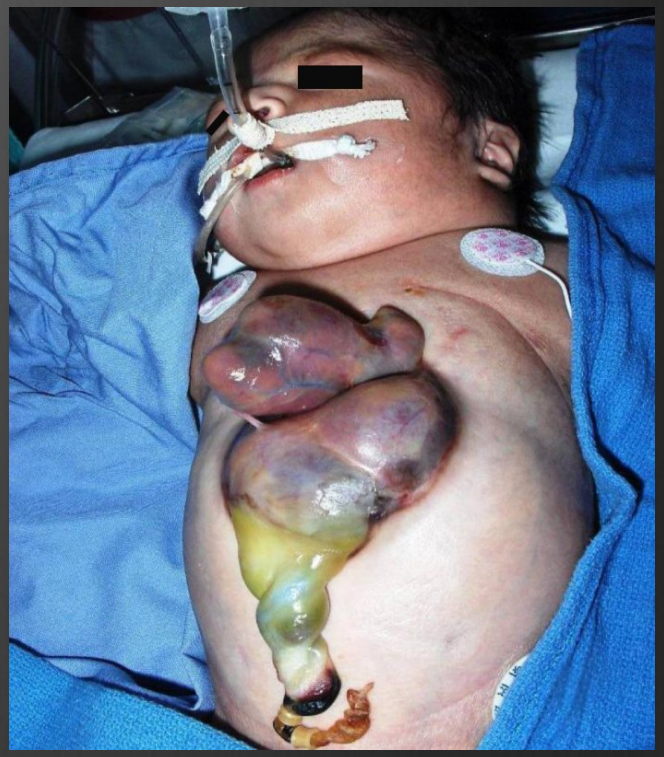
Associated Malformations

- Cephalic fold defect
- Caudal fold defect
- Beckwith-Wiedemann Syndrome



Cephalic Fold Defect

- Pentalogy of Cantrell
  - Thoracoabdominal ectopia cordis
  - Central tendon diaphragmatic defect
  - Upper midline omphalocele
  - Cardiac defect (VSD, LV diverticulum)
  - Apical pericardial defect



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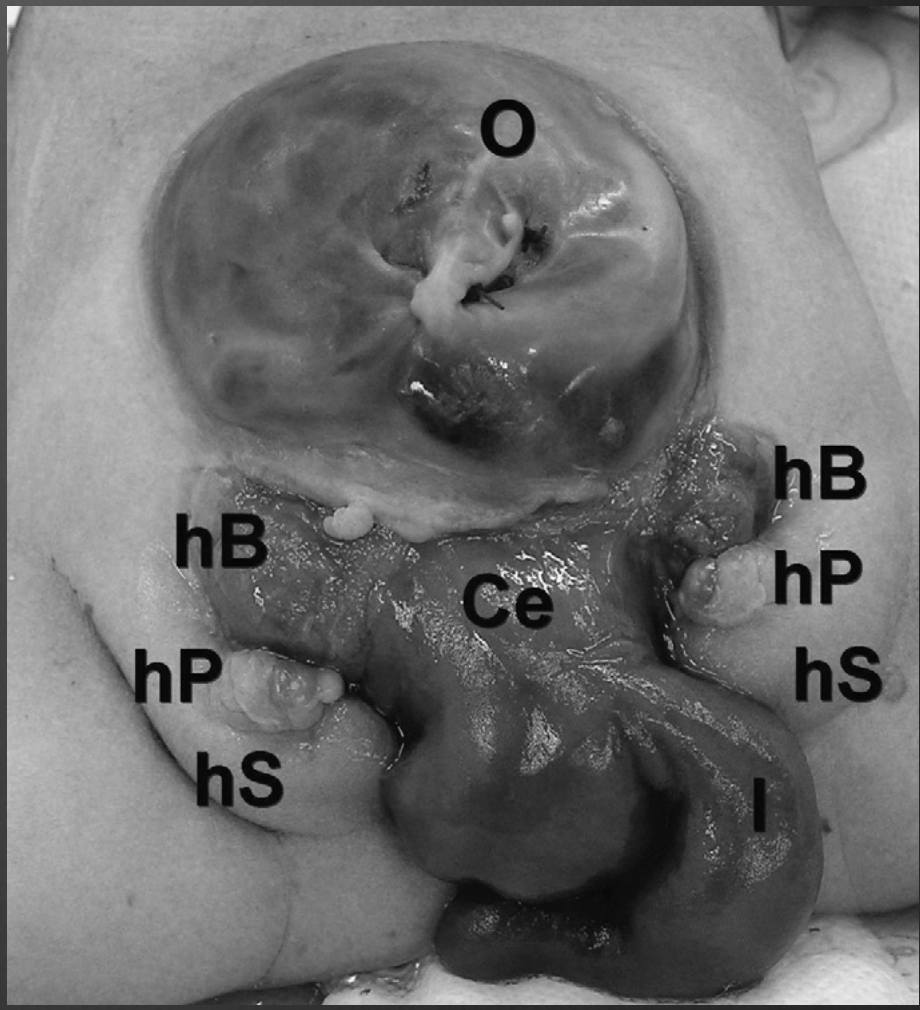
Caudal Fold Defect

- OEIS Complex
  - Omphalocele
  - Exstrophy
  - Imperforate anus
  - Spinal defect



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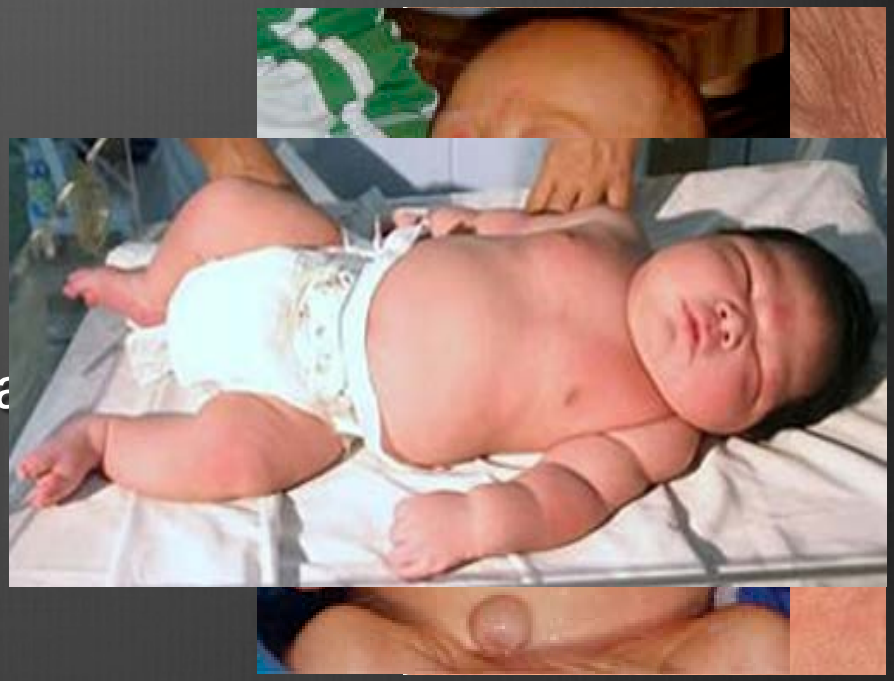
Beckwith-Wiedemann Sx



- Macrosomia
- Macroglossia
- Omphalocele/Umbilical hernia
- Ear pits or creases
- Neonatal hypoglycemia

- Increased likelihood of childhood cancers (Wilms, HB)

Initial Care

- Prompt neonatal examination, resuscitation
- Bowel decompression
- Evaluate for associated anomalies
  - CXR
  - Echo
  - Renal U/S
- Pediatric surgery consult for immediate/planned repair

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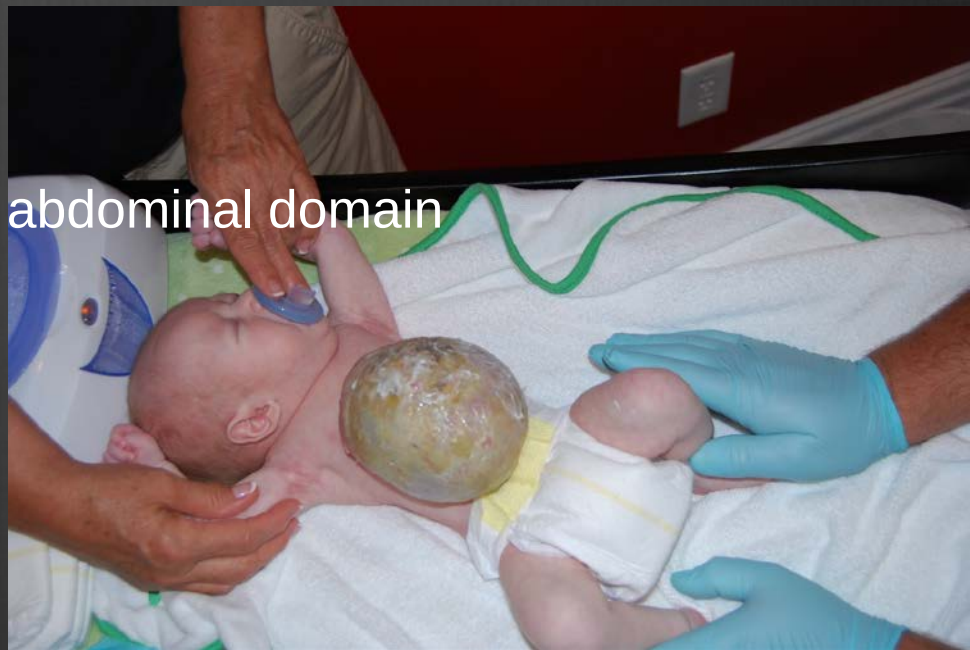
Surgery

- Small defects more amenable to primary closure
- ‘Giant’ defects managed with expected ventral hernia
  - Silvadene topically to promote granulation
  - Skin flaps and grafting
  - Tissue expanders increase abdominal domain



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Gastroschisis

- 1 in 3-8,000 live births
- Increasing incidence over past two decades
- Right paramedian defect without sac
- Prematurity, low birth weights in 40%
- Increased α-FP
- Classified as simple & complex gastroschisis
  - Presence of associated abnormality

Etiology

- Theories:
  - Failure of mesoderm to form anterior abdominal wall
  - Failure of lateral folds to fuse midline leaves defect
  - In utero rupture of omphalocele
  - Resorption of right umbilical vein leads to weakness

Risk Factors

- Maternal age < 21 years
- Stressed, under-nourished mothers
- Smoking
- Preterm delivery more common in gastroschisis
- Use of vaso-constrictive drugs, ephedrine, cocaine

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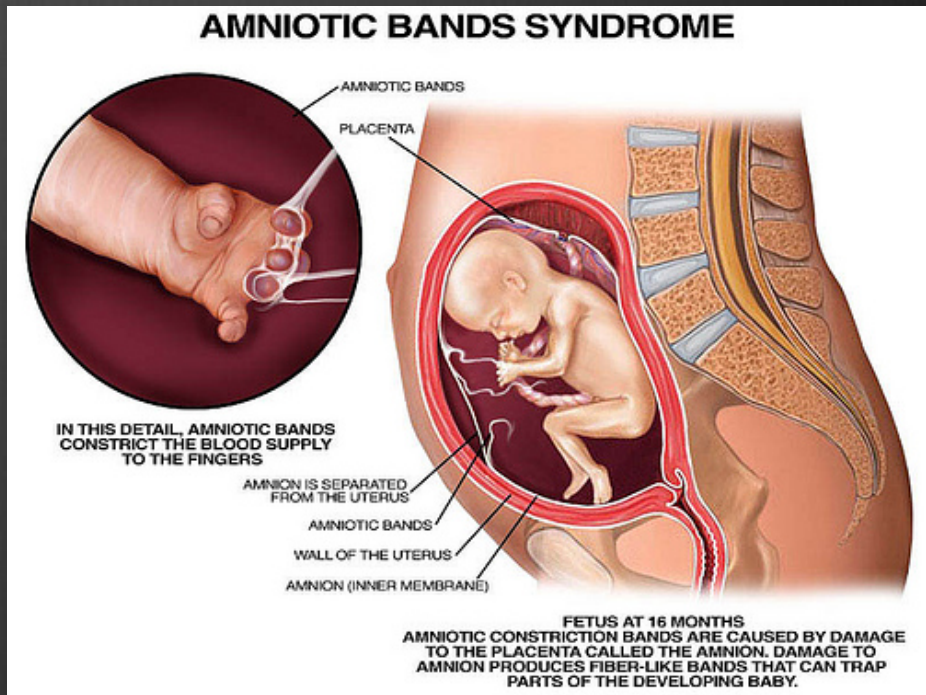
Associated Abnormalities

- Concomittant bowel atresias in 6.9-28% (jejunal/ileal)
- Cryptorchidism 10-15%
- Amniotic Band Syndrome
  - Thoracic Wall abnormalities
  - Limb abnormalities
  - Intestinal atresias
  - Abnormal genitalia
  - Meningocele
  - Umbilical cord abnormalities



Initial Care

- Prevent hypothermia & volume depletion; respiratory support
- Inspect and cover exposed bowel
- Position on side to prevent mesenteric kinking
- Establish central venous access early
- Rule out associated anomalies (rare)
- Rapid transfer to OR for attempted primary closure

Surgical Management

- Return the bowel to the abdominal cavity
  - Minimize damage and intra-abdominal hypertension
  - Central venous access for TPN, dysmotility common
- Surgical Techniques
  - Silo, serial reductions
  - Primary reduction with operative closure
  - Primary reduction with umbilical closure

Preformed Silo

- Originally used for staged repair
- Newer devices obviate need for fascial suture
- Bowel gradually reduced 1-2x daily as silo ligated
- Length of time to closure usually 1-14 days
- Decreased ventilator, vasopressor time
- Fewer ventral hernias

Postoperative Care

- Prolonged ileus is typical in gastroschisis
- Benefit from early central venous access, TPN
- Early oral stimulation; sucking-swallowing reflex lost
- Small bowel series if no function after 6-8 weeks
- 60% of patients have social stress without umbilicus

Controversies

- Optimal mode of delivery has been debated
  - No difference in outcomes of c-section vs vaginal
- Early delivery may prevent exposure to amniotic fluid
  - Literature is mixed, 1 RCT: shorter LOS, earlier feeding
  - Low birth weight (<2 kg) have longer course
- Silo vs primary fascial closure?

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Study

- Examined two cohorts: primary closure vs silo
- 190 patients in closure technique analysis
- Lower Apgar score in preformed silo group (*p=0.01*)
- Higher hernia rates among primary closure (*p=0.0006*)
- NICU stay longer in silo group *(p=0.002)*
- Retrospective cohort study, bias inevitable
- Included skin only closures as part of primary closure cohort
- To date, only 1 randomized prospective trial: **no difference**

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Summary

Omphalocele

- Sac
- Common anomalies
- Defect at umbilicus
- Surgery non-urgent

Gastroschisis

- No sac
- Rare anomalies
- Defect R of umbilicus
- Urgent surgical repair (?)

Summary

- Prevention of excessive heat and fluid loss is **key**
- Evaluate all pts with abd wall defects for anomalies
- Early primary fascial closure when possible
- ‘Giant’ omphaloceles require careful surgical planning
- Early TPN in gastroschisis, expect prolonged ileus
- Silo closure is also a reasonable option

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References

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- Ashcraft's Pediatric Surgery, 5th ed.
- Townsend: Sabiston Textbook of Surgery, 19th ed.
- Greenfield's Surgery: Scientific Principles and Practice, 5th Edition
- Schwartz' Principles of Surgery 9th Edition
- Weil B et al. The jury is still out: changes in gastroschisis management over the last decade are associated with both benefits and shortcomings. J Pediatr Surg 2012; 47: 119-124

With regard to defects of the abdominal wall, which statement is correct?

a) In gastroschisis, the herniated bowel contents are covered by a membrane.

b) Gastroschisis is frequently associated with cardiac malformations.

c) Chromosomal abnormalities are often present with omphalocele.

d) Treatment of abdominal wall defects is immediate surgical closure of the fascial defect.

e) In omphalocele, a silo bag is placed to cover the exposed intestine.

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Gastroschisis

(a) is usually associated with other anomalies

(b) occurs in babies born to older women

(c) is located on the left of the umbilical cord

(d) repair is followed by prolonged ileus

(e) occurs most often in infants > 4 kg

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You are a pediatric surgery consult resident called by NICU in regards to a 1 month old male infant presenting with flaccid abdominal wall, bilateral undescended testes and ureteral dilatation. Mom says she changes wet diapers several times a day. What is your plan?

a) “Call GU, they cover this sort of thing”

b) Bilateral orchiopexy and ureteral stent placement

c) Cystoscopy

d) “Observe, no acute surgical intervention at this time.”

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Prune-Belly Syndrome

- Also known as Eagle-Barrett or Triad Syndrome
- Extremely lax abdominal wall
- Significant comorbidities; pulmonary hypoplasia
- Higher incidence in males
- Predisposed to UTIs
- Abdominoplasty at 6-12 months