OMPHALOCELE & OTHER CONGENITAL ABDOMINAL WALL DEFECTS

AN INTERESTING CASE FROM KINGS COUNTY HOSPITAL CENTER

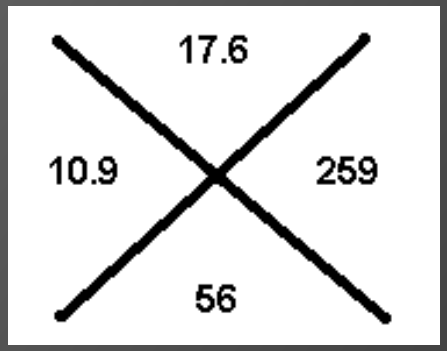
MADHURI RAO, MD, PGY-3

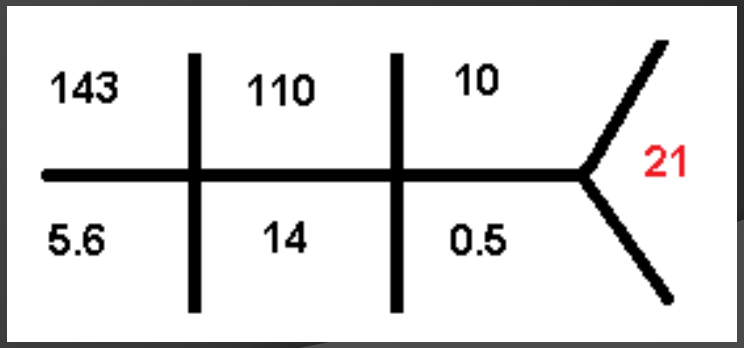
CASE PRESENTATION

- Baby – male, 4000gm, 39 week gestation
- Mother – 26 yo G4P2 with inadequate prenatal care at an outside institute
- Prenatal sonogram suggestive of omphalocele. s/p amniocentesis- normal karyotyping
- Normal vaginal delivery

SOON AFTER BIRTH…

- No acute distress
- Macroglossia, macrosomia, ear creases
- 3.5 cm defect in abdominal wall at the umbilicus with bowel covered by a sac
- Labs – Hypoglycemic FSG 30 mg/dL





IMMEDIATE MANAGEMENT

- Wet gauze over omphalocele by NICU team and pediatric surgery consulted
- Baby placed in warmer
- OGT inserted
- Rectal exam – meconium evacuated
- Omphalocele reduced at bedside and dressing placed over it
- NPO
- IVF and IV Abx started

SURGICAL MANAGEMENT

PRESURGICAL TESTING AND OPTIMIZATION

- Echocardiogram – PDA (1.7mm), PFO
  Rpt. Echo prior to discharge- normal
- Renal U/S – left hydronephrosis

Repair in the operating room planned for DOL 2

SURGICAL MANAGEMENT (Continued)

OPERATIVE DETAILS

- General anesthesia
- Excision of sac
- Ligation of umbilical vessels
- Dissection of fascia
- Closure of fascia with multiple figure of 8s with 3-0 PDS suture
- Closure of skin with circumferential inverted 4-0 PDS suture

POST OPERATIVE COURSE

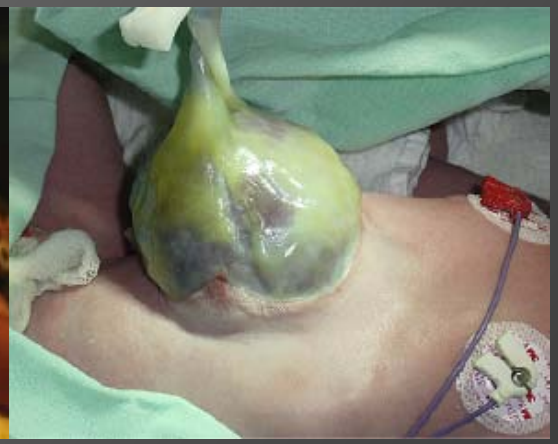
- POD 1 – Extubated
- POD 2
  - BMs +ve
  - PO feeds
- In house work up of associated abnormalities (r/o Beckwith-Wiedemann syndrome)
  - Syphilis –ve
  - Chromosome karyotype normal
- Discharged home on POD 8 (DOL 10)

CONGENITAL ABDOMINAL WALL DEFECTS

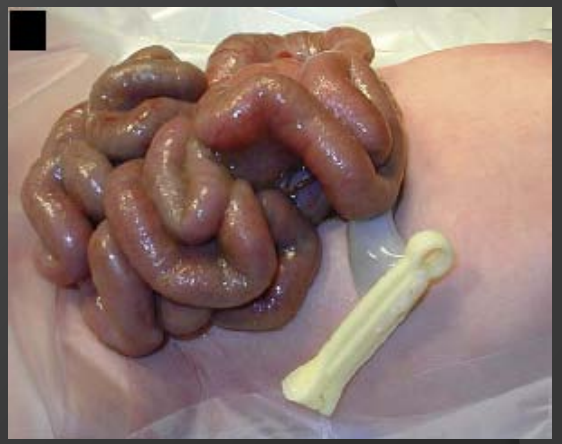
DEFECT	SITE	CONTENTS	ASSOCIATED ANOMALIES
Omphalocele – lateral fold	Umbilicus	Liver, intestine, spleen, gonad	Chromosomal, cardiac
Pentalogy of Cantrell-cephalic fold	Superior umbilicus	Liver, intestine	Cardiac, sternal cleft, pericardial defect, central tendon diaphragm defect
Cloacal exstrophy-caudal fold	Inferior umbilicus	Intestine	Bladder exstrophy, imperforate anus, epispadias
Umbilical cord hernia	Umbilicus	Intestine	Uncommon
Gastroschisis	Right umbilicus	Intestine	Intestinal atresia
Ectopia cordis thoracis	Midline sternum	Heart	Cardiac

Grosfeld: Pediatric Surgery, 6th ed.

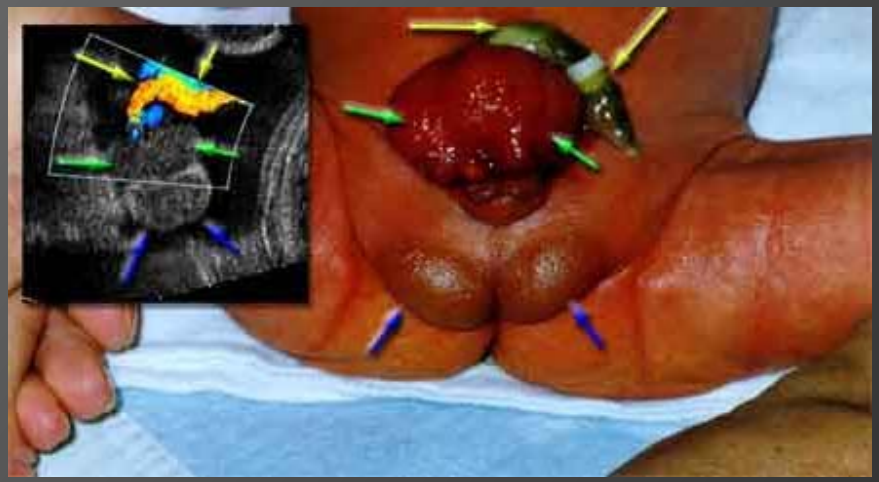
OMPHALOCELE



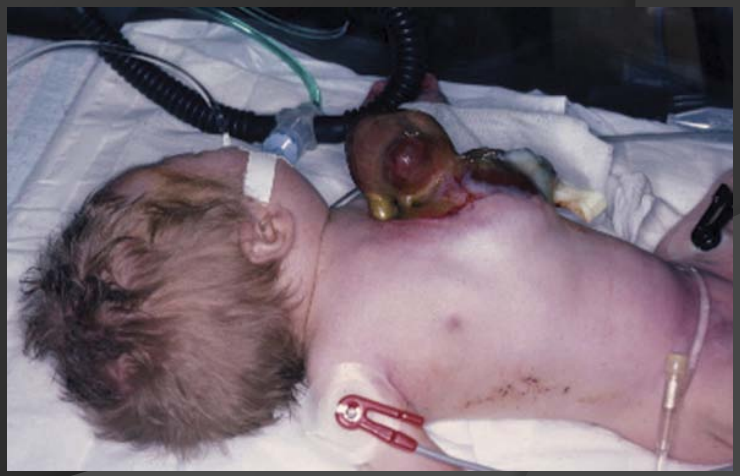
GASTROSCHISIS



CLOACAL EXSTROPHY



ECTOPIA CORDIS THORACIS



HISTORY

- Abdominal wall defects – first century AD by Aulus Cornelius Celsus, fifth century by Paulus Aegineta
- Omphalocele
  - 16th century by Ambrose Par
  - First successful repair by Hey in 1802
  - Stage reduction with prosthetic material by Schuster in 1967
- Gastroschisis
  - 16th century by Lycosthenes
  - Confused with ruptured omphalocele
  - Moore and Stokes described it as a separate entity
  - First successful repair by Visick in 1873

INCIDENCE

- 1 in 2000 live births
- Omphalocele – 1-2.5 per 5000 births, M > F
- Gastroschisis – 1 in 4000 births
- Increased detection with increasing antenatal U/S, ms AFP testing

EMBRYOLOGY

- **Week 3-5**

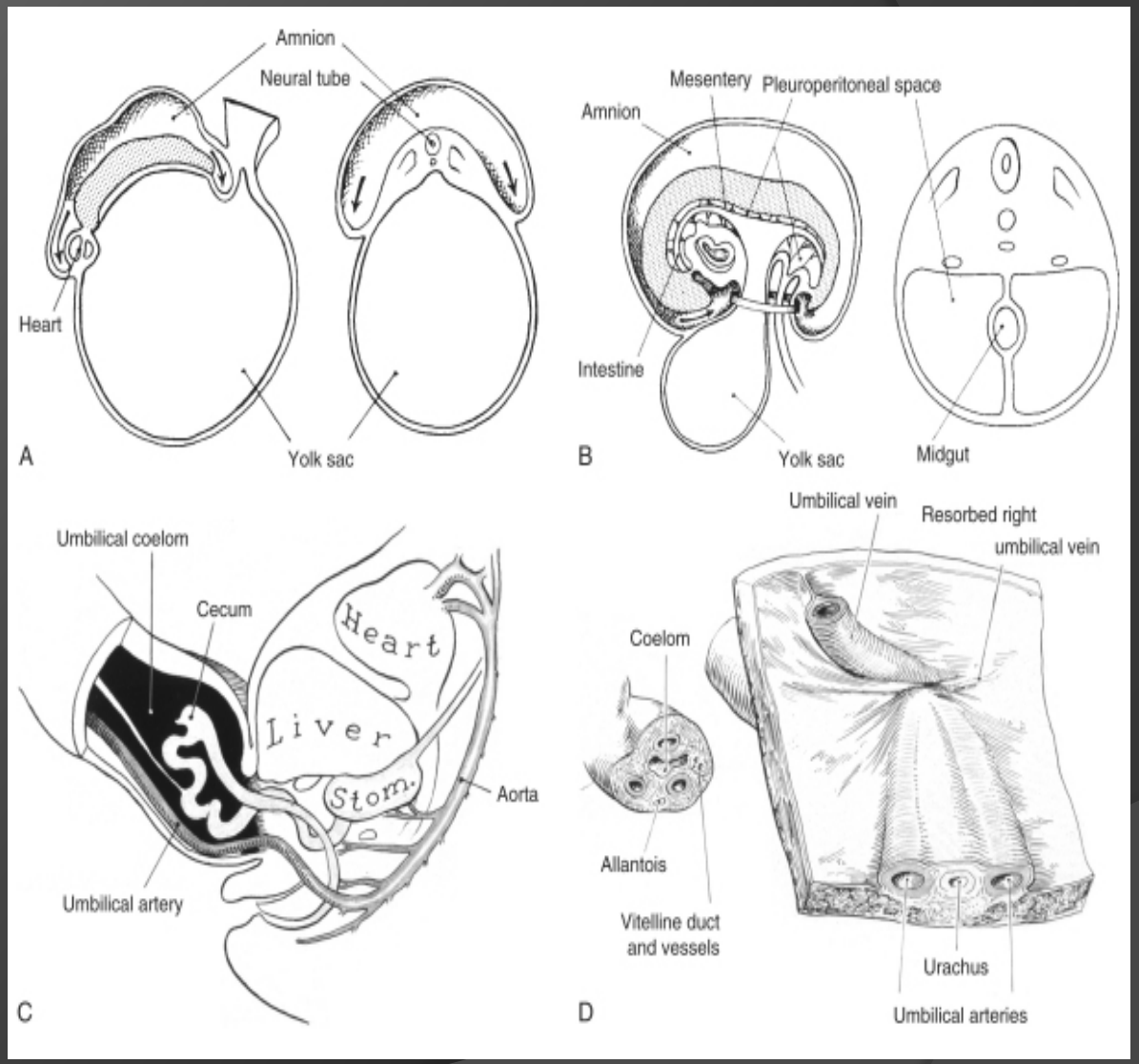
  Folding of embryonic disk in 4 planes converging at the umbilicus. Rapid growth of intestines.

- **Week 6**

  Physiological midgut herniation

- **Week 10-12**

  Reduction of PMH into abdomen for rotation and fixation



EMBRYOGENESIS OF DEFECTS

OMPHALOCELE	GASTROSCHISIS
Failure of extra embryonic gut to return into abdominal cavity – simple omphalocele Failure of lateral body folds to migrate medially – larger defect with liver	In-utero vascular accident • Right umbilical vein involution → necrosis in abdominal wall →defect • Premature right vitelline artery involution → weakening of abdominal wall →rupture a/w intestinal atresia
Covered by 2 layer amniotic peritoneal membrane	Not covered by membranes
Defect at the umbilicus	Defect to the right of the umbilicus
Occurs early in embryogenesis	Intestinal damage later in pregnancy

CLINICAL FEATURES

OMPHALOCELE	GASTROSCHISIS
Full term	Likely premature
Central abdominal wall defect	Defect to right of umbilicus
Sac	No sac
Contains midgut, liver and other organs	Contains midgut and sometimes gonad
<u>Associated conditions</u> • Cardiac (45%) – VSD most common • Chromosomal (40%) • Cryptorchidism (30%) • Macrosomia • GERD • Musculoskeletal • Neural tube defects	<u>Associated conditions</u> • Intestinal atresia • Small for gestational age • GERD (16%) • Cryptorchidism (15%)

OMPHALOCELE - ASSOCIATED SYNDROMES

SYNDROME	FEATURES
Beckwith-Wiedemann	Most common Omphalocele, macroglossia, hypoglycemia, nevus flammeus
OEIS	Omphalocele, Exstrophy, Imperforate anus, Spinal anomalies
Gershoni-Baruch	Omphalocele, diaphragmatic hernia, cardiovascular anomalies, radial ray defects
Donnai-Barrow	Diaphragmatic hernia, exomphalos, absent corpus callosum, hypertelorism, myopia, sensorineural deafness (autosomal recessive)
Fryns	Diaphragmatic hernia, coarse facies, acral hypoplasia, omphalocele
Trisomy 13, 18, 21	

MANAGEMENT – PRENATAL

- Antenatal U/S
  - Omphalocele – Sac +ve, Liver +/-, sensitivity = 75%
  - Gastroschisis – Sac -ve, Liver -ve, sensitivity = 83%
  - Look for other associated anomalies
- Amniotic fluid – elevated AFP, AChE
- Maternal serum – elevated AFP

Helps prenatal counseling and planning

MANAGEMENT – DELIVERY

- Plan for delivery at a tertiary care center
- Involvement of obstetric, NICU and pediatric surgical team
- No benefit of C-section over vaginal delivery * (practiced in some cases of giant omphalocele because of potential liver injury or sac rupture)
- Plan for repair ASAP
  - Worsening bowel edema in gastroschisis
  - Enlarging liver with omphalocele

• Segel SY, Marder SJ, Parry S, et al: Fetal abdominal wall defects and mode of delivery: A systematic review. Obstet Gynecol 2001; 98:867
• Singh SJ, Fraser A, Leditschke JF, et al: Gastroschisis: Determinants of neonatal outcome. *Pediatr Surg Int* 2003; 19:260.

MANAGEMENT – NEONATAL

Principles of surgical management

- Safely reduce evisceration
- Cosmetic closure of defect
- Identify and treat associated anomalies
- Nutritional support
- Management of abdominal, wound, or bowel complications

MANAGEMENT – NEONATAL (cont.)

Immediate care

- OGT/NGT
- Rectal exam
- Maintain body temperature
- Ventilatory support
- Antibiotics

Omphalocele

- Dressing to minimize heat loss
- Maintenance IV fluids
- Look for associated anomalies

Gastroschisis

- Heat preservation with bowel bag
- Resuscitative IV fluids
- Plan for immediate OR

MANAGEMENT – NEONATAL (cont.)

OPERATIVE MANAGEMENT – OMPHALOCELE

- Primary vs. staged vs. delayed closure based on size and condition of the neonate

- PRIMARY CLOSURE
  - Reduce abdominal contents with intact sac
  - Circumferential dissection to rectus
  - Excise, invert and ligate sac with vessels and urachus
  - Fascial closure with mattress suture
  - Purse string suture for skin to recreate umbilicus

MANAGEMENT – NEONATAL (cont.)

Larger defects – techniques to facilitate primary fascial closure

- Creation of flaps
- Component separation
- Bioabsorbable mesh
- Tissue expanders

MANAGEMENT – NEONATAL (cont.)

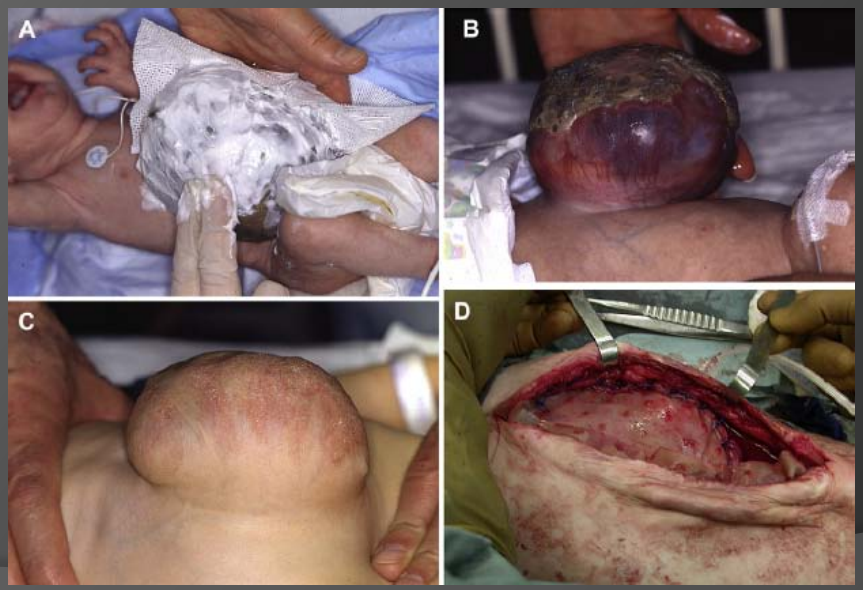
STAGED CLOSURE

- Sac excised and silo bag sewn to fascia
- Can use sequential ligation of a thick sac
- Slow reduction over 7-8 days
- Delayed fascial closure in the OR

MANAGEMENT – NEONATAL (cont.)

DELAYED CLOSURE WITH ESCHAROTIC THERAPY

- Severe congenital anomalies preventing closure
- Gradual epithelialization of the omphalocele sac – Silvadene, Povidone-iodine, Mercurochrome (not used)
- Repair of formed ventral hernia after epithelialization



MANAGEMENT – NEONATAL (cont.)

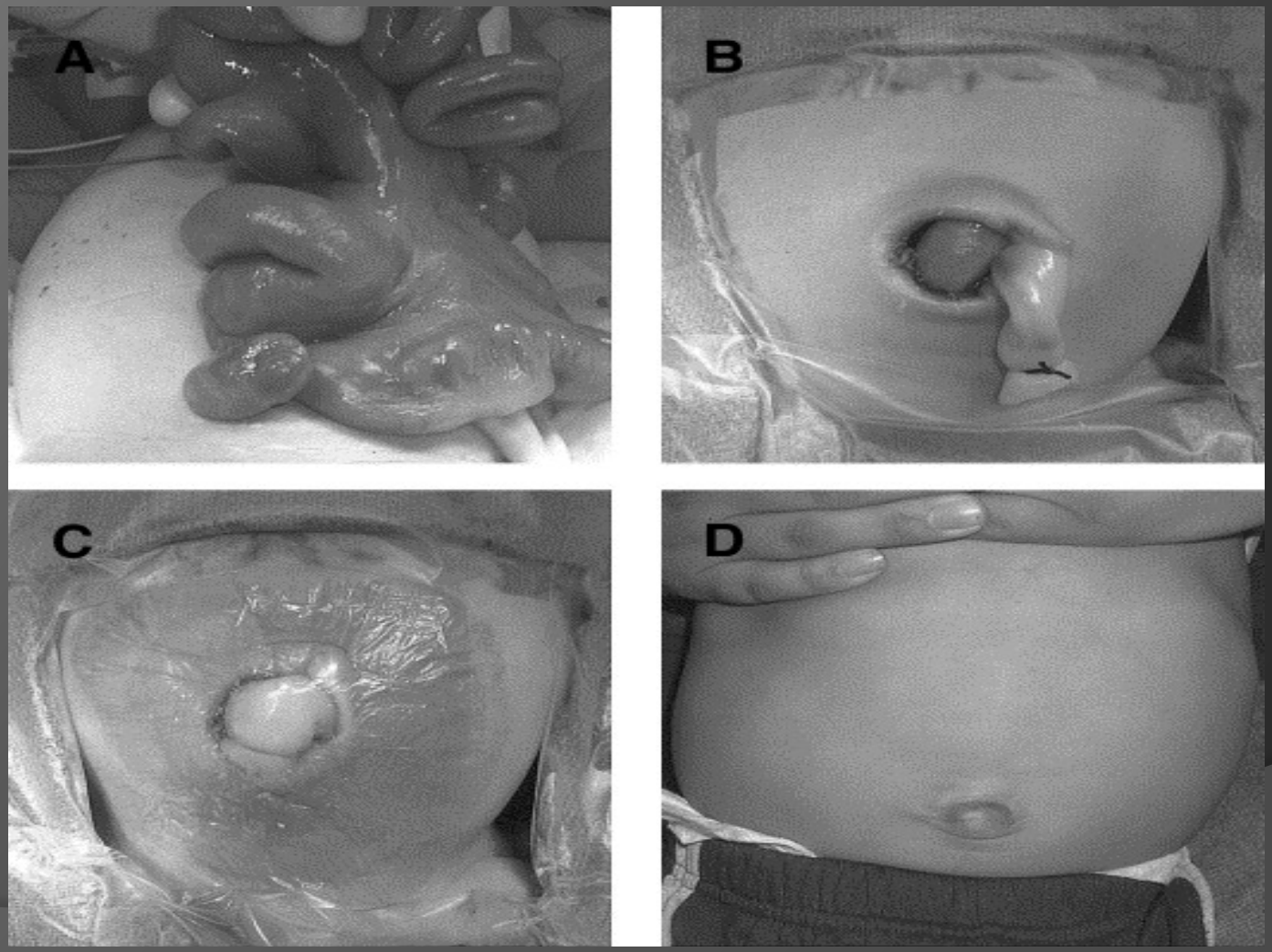
OPERATIVE MANAGEMENT – GASTROSCHISIS

- Primary reduction with operative closure of fascia
- Silo placement, serial reductions and delayed fascial closures
- Primary or delayed reduction without fascial closure
- ‘Plastic’ sutureless closure

Sandler, J. Lawrence and J. Meehan, *et al.* A “plastic” sutureless abdominal wall closure in gastroschisis. *J Pediatr Surg*, **39** (2004), pp. 738–741

MANAGEMENT – NEONATAL (cont.)

Sutureless closure



MANAGEMENT – NEONATAL (cont.)

POSTOPERATIVE CONSIDERATIONS AND COMPLICATIONS

SHORT TERM

- Ventilatory support
- Compartment syndrome from tight closure
  - Monitor ventilatory pressures, IAH, urine output and distal perfusion
- Delayed bowel function
  - Need for TPN/PPN
- NEC – risk factors of IAH, prematurity, birth weight, delayed feeding
- Wound failure
- Infection and sepsis

MANAGEMENT – NEONATAL (cont.)

POSTOPERATIVE CONSIDERATIONS AND COMPLICATIONS

DELAYED

- Ventral hernia (repaired after age 1 year)
- Need for umbilicoplasty
- Adhesive SBO – 25% of gastroschisis and 13% of omphalocele repairs

van Eijck Floortje C, et al. The incidence and morbidity of adhesions after treatment of neonates with gastroschisis and omphalocele: a 30 year review. J Pediatr Surg 2008;43:479-83.

PROGNOSIS

OUTCOME MEASURES

- Survival
  - Associated anomalies
  - Intestinal length and function
  - Viscero-abdominal disproportion (influences risk of complications)
- Nutritional autonomy
- Cosmesis

PROGNOSIS (cont.)

- Omphalocele
  - 94% overall survival
  - Mortality from cardiac and chromosomal anomalies
  - Long term problems including GERD, pulmonary insufficiency, failure to thrive in 60% of infants

    Biard JM, et al. Prenatally diagnosed giant omphaloceles: short- and long-term outcomes. Prenat Diagn 2004;24:434-9.
- Gastroschisis
  - 99% overall survival
  - Prognosis depends on intestinal atresia and degree of irreversible bowel damage

LITERATURE REVIEW

Review of the evidence on the closure of abdominal wall defects

Mortellaro VE, et al. Department of Surgery, Children's Mercy Hospitals and Clinics, Pediatric Surgery International, Dec 2010.

Management of omphalocele

- Based on case reports and small case series
- Results of a retrospective review from Kings College Hospital, London (1995-2002)
  - 35 infants: 11 (minor) – primary closure, 13 (major) – primary closure, 11 (major) – staged closure
  - Infants with staged closure needed significantly longer periods of ventilation, longer hospital stay and more delayed bowel function

LITERATURE REVIEW

Reports on techniques for repair of large omphalocele

Table 1 Reports on techniques for staged repair in large omphaloceles

Method	Reference, year	No. of patients	Survival	Complications
Alloderm	Kapfer 2006, JPS 41:216 [62]	4	4 short term, 2 long term	Deaths from cardiopulmonary issues
	Alaish 2006, JPS 41:e37 [57]			
VAC	Kilbride 2006, JPS 41:214 [58]	3	3	None
Tissue expanders	Martin 2009, JPS 44:178 [59]	6	6	Infection, extrusion in one
	Foglia 2006, JPS 41:704 [74]			
	De Ugarte 2004, JPS 39:613 [75]			
	Bax 1993, JPS 28:1181 [76]			

Table 2 Reports on techniques for delayed repair in large omphaloceles

Method of epithelialization	Reference, year	Number of patients	Number of patients who survived	Complications
Mercurochrome	Used in 1960–1980s primarily [2]	n/a	n/a	Mercury poisoning, not used currently
Povidone-iodine	Whitehouse 2010, JPS [67]	6	5 (CHD)	Transient increase TSH, not clinically significant
Silver sulfadiazine/ neomycin/bacitracin	Lee 2006, JPS 41:1846 [63]	22	19	Two ruptured sacs, one bleeding sac
	Pereira 2004, JPS 39:1111 [69]	11	11	

LITERATURE REVIEW

Management of gastroschisis

- Grade C evidence – no survival difference between immediate and delayed closure
- Routine use of a SILASTIC spring-loaded silo for infants with gastroschisis: a multicenter randomized controlled trial.

  Pastor AC, et al. Pediatric General Surgery, Hospital for Sick Children, Canada, Journal of Pediatric Surgery 2008 Oct;43(10)

  - 27 infants – primary closure, 27 infants – spring loaded silo and elective closure
  - No significant difference in LOS, time on TPN, sepsis, NEC

LITERATURE REVIEW

Management of gastroschisis

- Some evidence for using IAP to guide closure*
  - Limit of 20mmHg for primary closure
  - Also used to guide sedation, paralysis and silo reduction

Olesevich M, et al. Gastroschisis revisited: role of intraoperative measurement of abdominal pressure, J Pediatric Surg 40(5):789-792

Lacey SR, et al. Bladder pressure monitoring significantly enhances care of infants with abdominal wall defects: a prospective clinical study, J Pediatric Surg 28(10):1374-1375

SUMMARY

- Spectrum of congenital abdominal wall defects (Omphalocele and gastroschisis most common)
- Embryology and embryogenesis
  - Omphalocele – failure of migration of lateral fold
  - Gastroschisis – various theories (intrauterine ischemic event)
- Difference in clinical features between omphalocele and gastroschisis
  - Omphalocele – looks better but associated with more lethal anomalies
  - Gastroschisis – looks worse but has a better prognosis

SUMMARY (cont.)

- Management
  - Prenatal diagnosis may influence timing and location of delivery
  - Neonatal resuscitation focused on normothermia, fluid resuscitation, GI decompression, and ruling out associated anomalies
  - Surgical management – primary vs. staged closure

    Goal is to minimize damage to bowel and obtain cosmetically acceptable closure
- Prognosis
  - Omphalocele – associated anomalies
  - Gastroschisis – degree of bowel injury

REFERENCES

- **Grosfeld: Pediatric Surgery, 6th ed.**
- **Contemporary postnatal surgical management strategies for congenital abdominal wall defects**

  Marven S, et al. Sheffield Children's Hospital NHS Foundation Trust, Western Bank, United Kingdom; Leeds Teaching Hospitals NHS Trust, Leeds General Infirmary, Leeds, United Kingdom. Seminars in Pediatric Surgery (2008) 17, 222-235
- **Neonatal abdominal wall defects**

  Christison-Lagay ER, et al. Division of General and Thoracic Surgery, Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada, Department of Pediatric Surgery, Massachusetts General Hospital for Children, Boston, MA, USA
- **Review of the evidence on the closure of abdominal wall defects**

  Mortellaro VE, et al. Department of Surgery, Children's Mercy Hospitals and Clinics, Pediatric Surgery International, Dec 2010.
- **Etiology of intestinal damage in gastroschisis. II. Timing and reversibility of histologic changes, mucosal function, and contractility**.

  Langer JC, et al. J Pediatr Surg, ***25*** *(1990), pp. 1122–1126*

THANK YOU