Omphalocele

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Outline

1. Embryology
2. Epidemiology
3. Antenatal
4. Postnatal
5. Surgery
6. Outcome

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1. Embryology

Physiologic herniation fails to return to abdomen (6-10wk)

- **Sac** composition similar to umblical covering (amnion, Wharton jelly, peritoneum)
- **umblical cord** inserts into sac (sometimes at apex, sometimes closer to abd wall)
- **Contents** can include intestine (with non rotation), liver, bladder, stomach, gonads
- Poor diaphragmatic motion, narrow thorax and small lung area leading to **pulmonary hypoplasia**.

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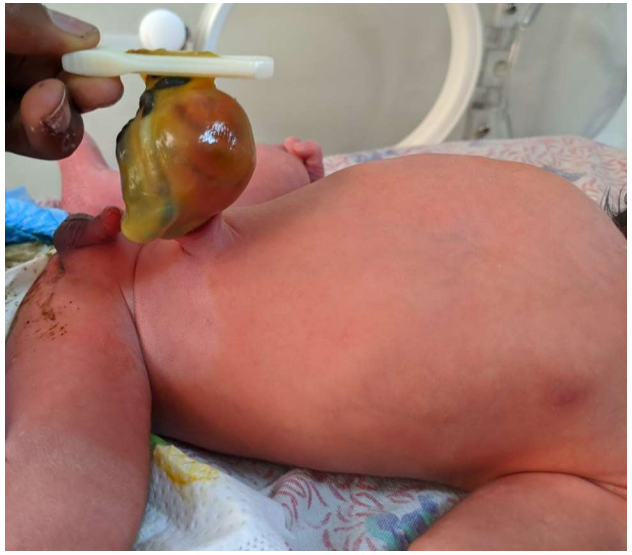

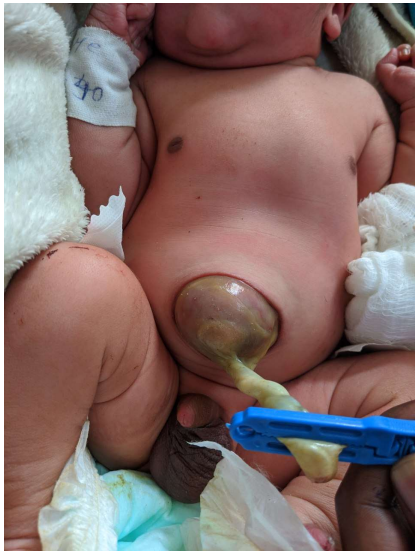

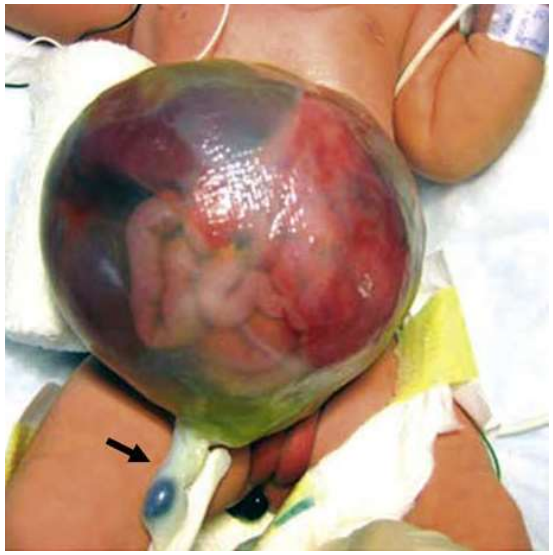

- **size-based classification** that separates omphaloceles into hernias of the cord (<1.5cm), small, medium, large, and giant defects but distinction not clear. Some categorize **based on liver herniation** as small (liver in) and giant (liver out). Others based on **weather it can be closed primarily.**

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2. Epidemiology

- **1 in 5000** (1 in 10,000 for large)
- Incidence has remained stable unlike gastroscisis
- **Associated problems**
  - Prematurity
  - Pulmonary hypoplasia – more common in giant omphaoceles
  - Chromosomal Anomalies 50% (trisomy 21,18,13)--- more common in small omphaloceles
  - Syndromes (beckwith-Wiedemann, cloacal exstrophy, pentalogy of cantrell)
  - Cardiac anomalies 20-40%, CNS anomalies
  - Short bowel in 10%

3. Antenatal diagnosis and management

- Diagnosis
  - elevated maternal serum **α-fetoprotein and Ultrasound**
  - Antenatal incidence higher ~1 in 1100 (**IUFD/termination**)
- Screening
  - **fetal echo** and **Karyotyping**
  - **MRI** is better in detecting pul hypoplasia (predicting neonatal ventilatory support)
- Delivery
  - There is no advantage of preterm delivery
  - **Most prefer C section** despite lack of data, b/c of fear of sac rupture & liver injury

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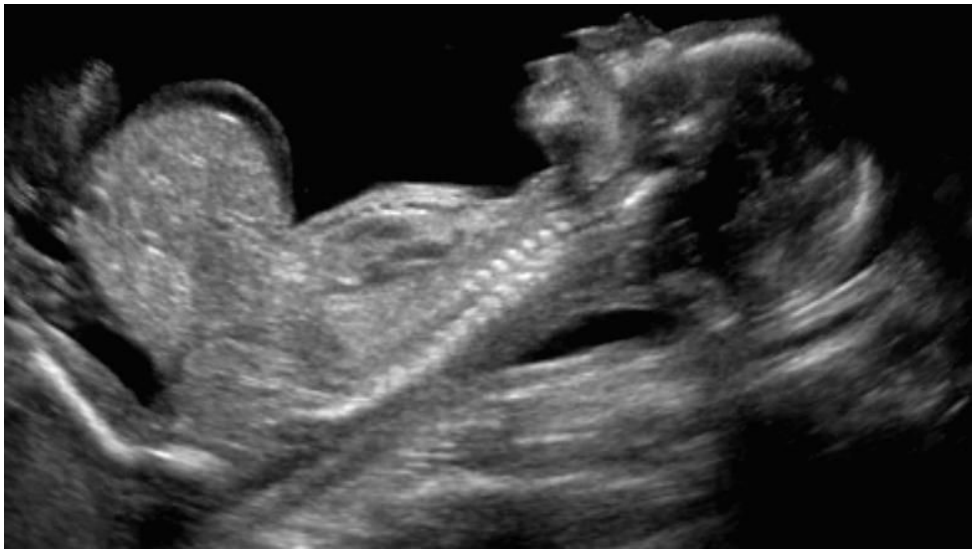

- **Diagnosis** can be made at 18-wk on 2D ultrasound. Finding bowel in umblical cord can be normal upto 12 wk. prenatal finding of a “giant” omphalocele has not been accurate in **predicting postnatal outcomes** but O/HC and O/AC ratios are being studied,

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4. Neonatal management

- **pulmonary hypoplasia** may require immediate intubation and ventilation
- Cover (saline soaked gauze) – minimize fluid/temp loss and dec risk of rupture
    - * if ruptured manage as gastroschisis (silo, antibiotic)
- Gastric decompression – NG tube drainage
- **Rule out cardiac anomalies** – clinical evaluation, CXR ± echo
- **Rule out renal anomalies** – Renal ultrasound
- **Neonatal hypoglycemia** – BW syndrome

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5. Surgery

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Considerations choosing repair type

- Size of defect
- Amount of herniated viscera
- Presence of associated anomalies
- Gestational age

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5.1. Primary closure

<u>Closure in a single procedure</u> = For small and medium sized omphaloceles

- **Sac excision:**
  - leave part of the sac on the liver and bladder,
    - Leaver may bleed (sac is densely adherent to gleason's & hepatic vein may also be underneath)
    - Bladder may be injured
  - Omphalomesenteric duct, allantoic cyst or ectopic liver tissue should be recognized and excised
- **Fascial closure:**

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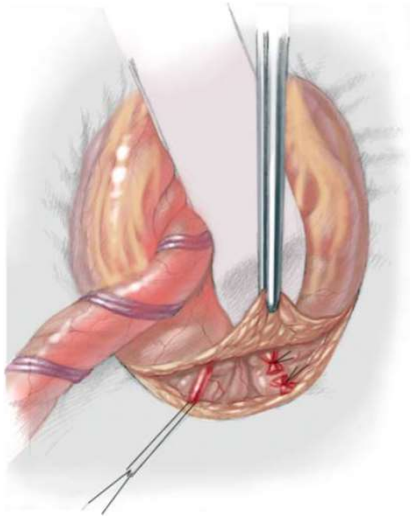

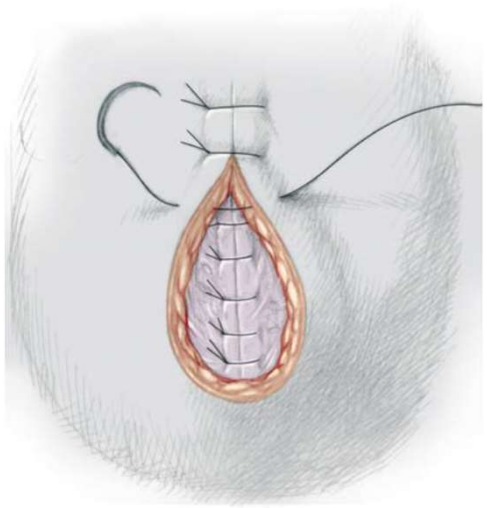

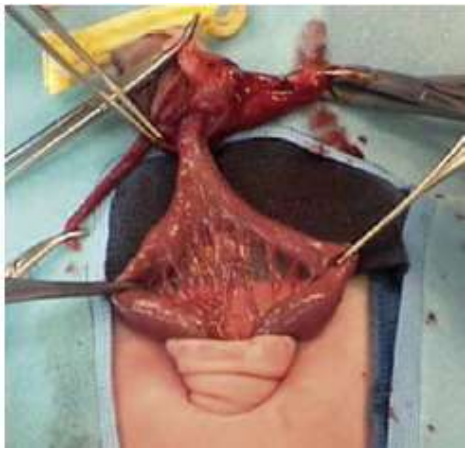

- **Sac excision and Primary closure**: The skin is undermined enough that a secure fascial closure can be accomplished. For very small defects, the umbilical cord can be left behind to give better cosmetic results. it is not unusual to find an omphalomesenteric duct remnant during repair of small omphalocele

5.2. Staged closure

Early closure with minimal compression = For Giant omphaoceles

- **Serial Reduction**
  - Abdominal wall skin Flap + Removal of sac **(Gross 1948).**
  - Placement of silastic sio + Removal of sac **(Schuster 1967)-** usually need hand sewn b/c of large size
  - sequential ligation of the sac itself **(Hong 1994)-** if thick sac with cord inserting near the apex
  - Vaccum assisted closure **(Aldrige 2016)**
- **Fascial closure**

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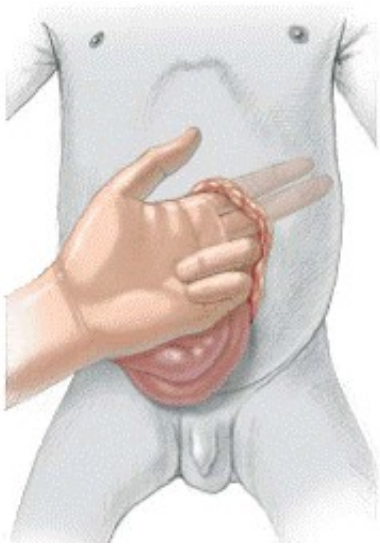

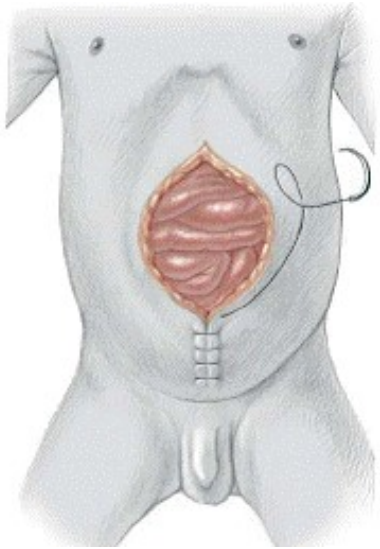

- **Abdominal wall skin flap**: can dissect lateraly upto ant axillary line. Patients are left with ventral hernia. Some surgeons close the fascia with patch at the same time.

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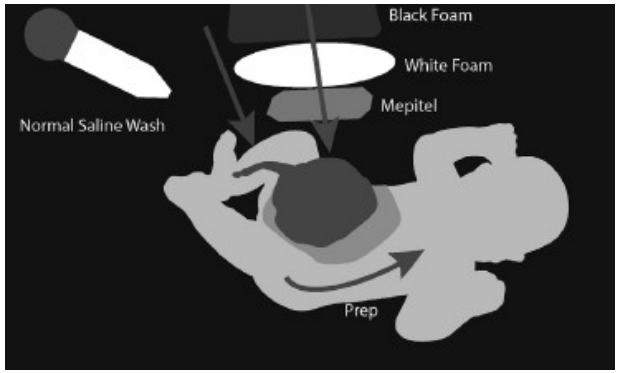

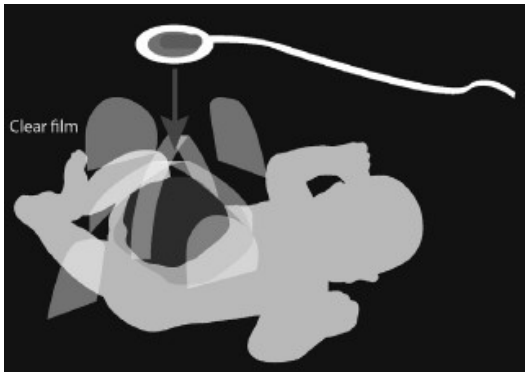

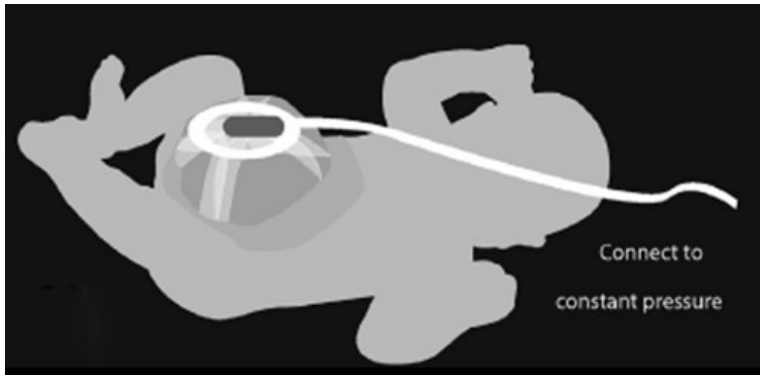

- **Wound VAC (Vacuum assisted closure):** negative pressure wound therapy; using a suction pump, tubing and a dressing to remove excess exudate and promote healing

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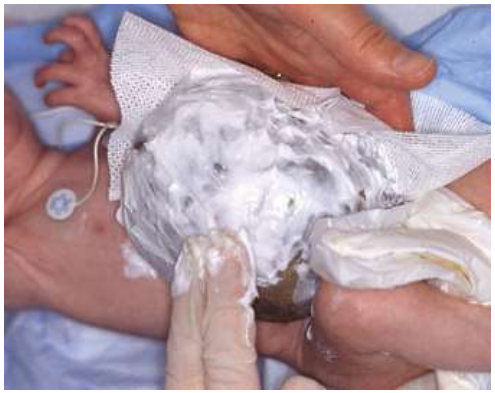
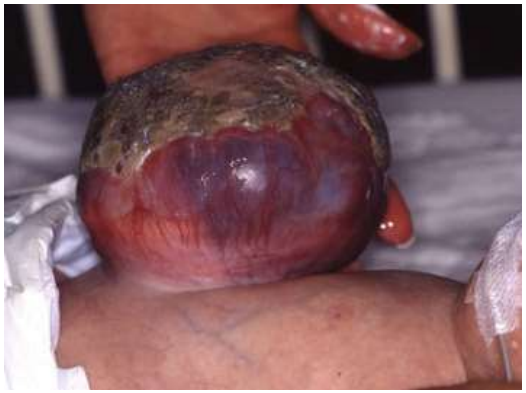
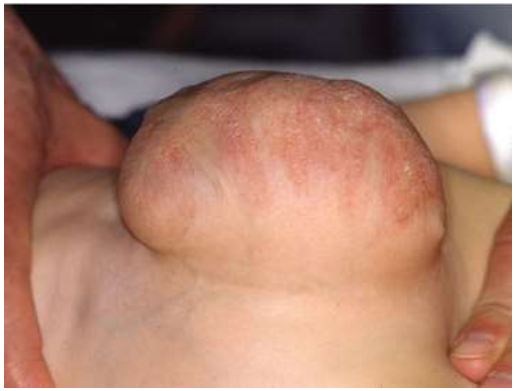
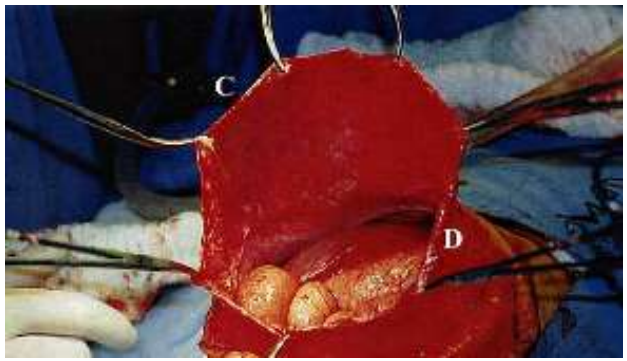
5.3. Delayed closure

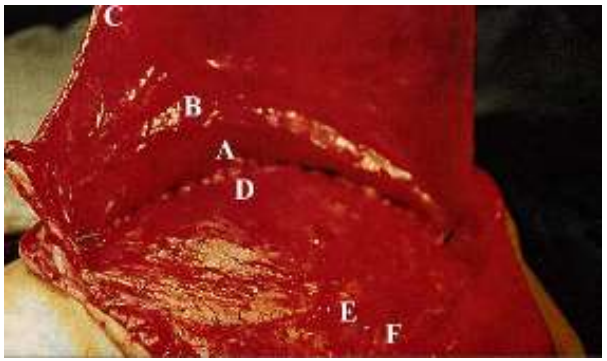
Initial treatment with topical therapy = for those who can't tolerate inc abd pressure (premature, pul htn, CHD)

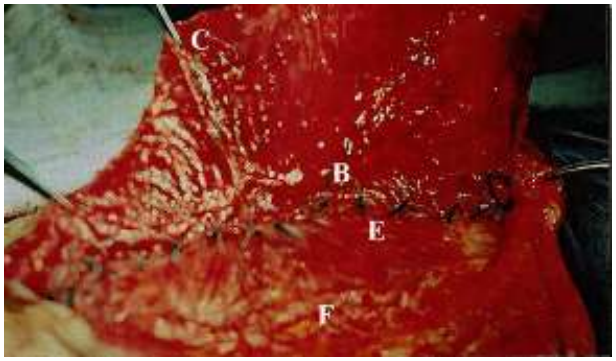
- **Escharotic therapy** (Ahlfeld 1899)
  - gradual sac epithelialization (Complete neo-epithelialization takes 2 to 3 months)
    - mercurochrome = also a disinfectant; but deaths due to mercury poisoning (abandoned)
    - silver nitrate - because of its hypotonicity, leads to hyponatremia
    - (Betadine) Povidone-iodine = systemic absorption with transient hypothyroidism (needs monitoring)
    - (Silvadene) Silver sulfadiazine = frequent dressing changes, which can damage granulation tissue
    - **1% Gentian violate** = cheap and studies from developing countries show its safe & effective
    - Other = acacia, manuka honey, neomycin, and polymixin/bacitracin
- **Fascial closure =** 1-5 yr of age


- **Escharotic (**silver sulfadiazine**) for delayed closure**. **Compression dressing**, helps in sequentially reducing the contents into the abdomen and facilitates subsequent closure. Rarely, operative closure may not be needed







- **Lazaro da Silva's technique for delayed closure of ventral hernia:** bilateral longitudinal fibroperitoneal aponeurotic transposition, resulting in 3 different layers of closure. Longitudinal midline opening of the hernial sac produces 2 flaps. Formation of the 4 other flaps from the rectus sheath (ant & post)

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Fascial closure Techniques

if pressure is felt unacceptable during fascial closure, different techniques can be used.

- interposition mesh: Patch in the abdominal wall and close the skin over the patch
  - Non absorbable (Marlex, Gortex, Prolene) risk infection, need interval removal, herniation
  - bioabsorbable materials (alloderm, surgisis) Can apply topical therapy over it for granulation
- Component separation dissection of subcutaneous tissue, muscle and fascia and **(Levy 2013)**
- Intraperitoneal Tissue expanders with volume added overtime **(Bax 1993).**

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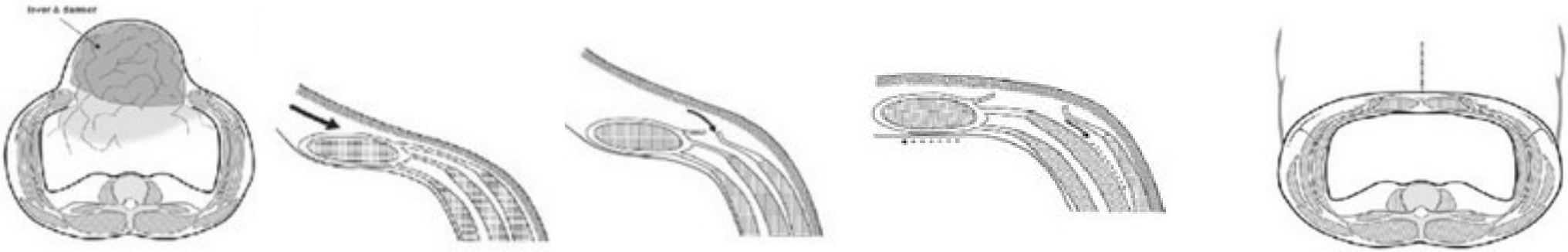

- **Component separation** : Dissection of the skin and subcutaneous fat from the abdominal wall muscles. Incision of the aponeurosis of the external oblique muscle 1 cm lateral of the rectus sheath over its full lenth including rib attachement. Separation of the external and internal oblique muscles upto mid-axillary line. After this closure of the abdomen is possible (rectus can be shifted medially 5 cm), the external oblique muscle is retracted laterally. **Complications** include skin necrosis, and haematoma).

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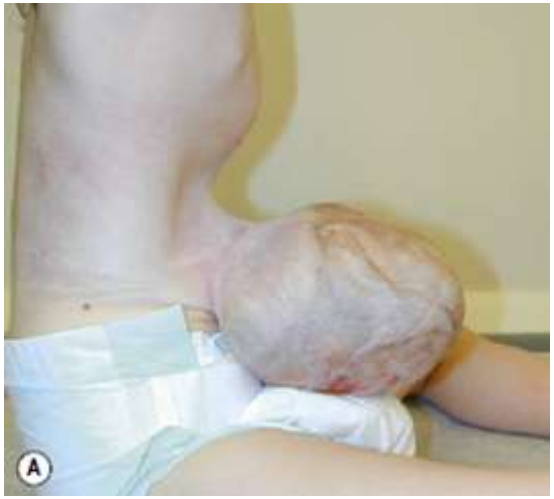

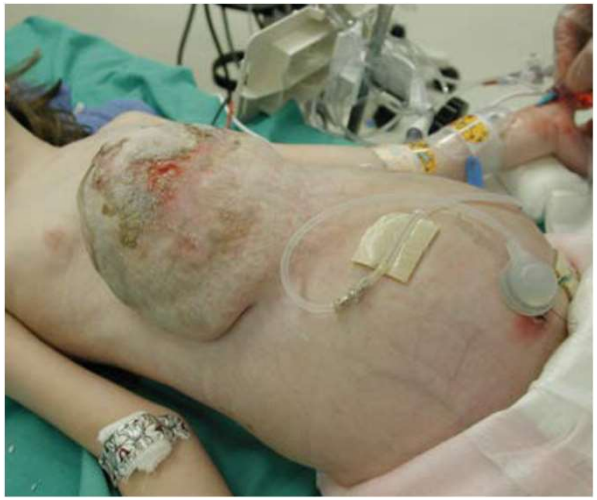

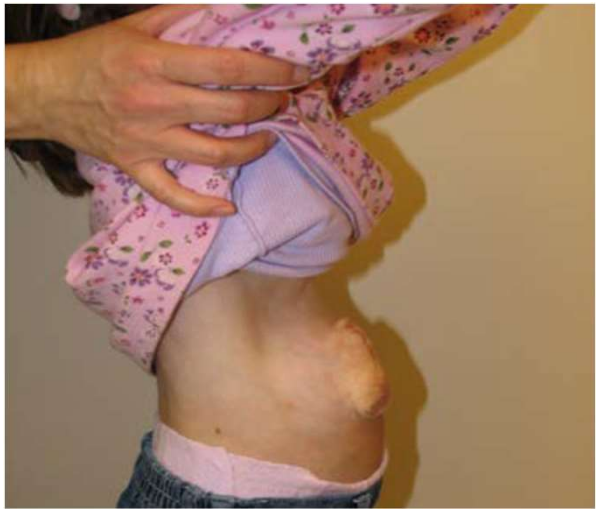

- **intraperitoneal tissue expander**: to correct abdominovisceral disproportion

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Post op care (after closure)

- **mechanical ventilation**: need in most for a few days until edema dec, especially if liver hernaition
- **monitoring:** urine output, tissue perfusion, and intraabdominal pressure
- **Gastric decompression**
- **Parentral nutrition**

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6. Outcome

- **Prognosis** – depend on severity of associated anomalies (20% do not live beyond 1st yr of life)
- Abdominal compartment = acute hepatic congestion, renal failure, and bowel infarction
- Escharotic = Early feeding and dec need of MV; But inc hospital stay, daily dressings, and wound infection
- Herniation rate = delayed closure (9%) >> staged closure (18%) >> primary closure (58%)
- Long term complications
  - Gastrointestinal - reflux, feeding difficulty and failure to thrive
  - Respiratory (60%) - pulmonary insufficiency, recurrent lung infections, or asthma
  - Psychological - cosmetic appearance of the abdomen (umbilicus, scar)
  - Neurodevelopmental delays

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Nebiyu Shitaye, Belachew Dejene. *Ethiop Med J, 2017, Vol. 55, No. 3*

ORIGINAL ARTICLE

MAJOR ABDOMINAL WALL DEFECTS AND OUTCOME OF MANAGEMENT AT A REFERRAL HOSPITAL.

- 23 patients over 4 years
  - 13 patients **GV paint**, 3 died (20%)
  - 9 patients **primary closure**, 2 died (20%)
  - 1 patient **staged closure**, 0 death (0%)
- Omphalocele **mortality 20%** (5/23) -- better than gastroschisis (70% mortality)

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References

- **Hollcomb and Aschraft Pediatric Surgery** 7th edition, 2020
- **Pediatric Surgery: General principles and newborn surgery**, 2020