Giant Omphalocele Repair: A 26-Year Single Surgeon Consecutive Case Series

Sameer Shakir MD, Mychajlo Kosyk BA, Sammy Othman MD, Saïd C. Azoury MD, Holly Hedrick MD, David W. Low MD

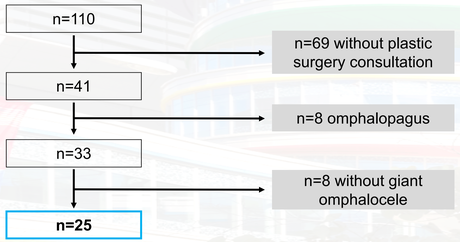
Divisions of Plastic Surgery and General Surgery, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA

Background

- Omphalocele represents the most common congenital defect of the abdominal wall and is associated with a mortality rate between 20-70%
- Giant omphalocele, resulting in abdominovisceral disproportion (AVD), is characterized by an abdominal wall defect >6 cm in diameter and/or liver herniation
- We provide the longest known consecutive case series of a senior surgeon's experience with giant omphalocele repairs in a multidisciplinary fashion over nearly 3 decades
- We hypothesized staged reconstruction of giant omphaloceles using a combination of extra-abdominal tissue expansion and bridging mesh is associated with low postoperative morbidity and hernia recurrence rates

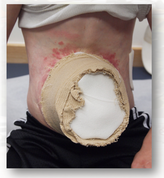
Methods

Study design: Retrospective cohort study of all consecutive subjects undergoing giant omphalocele repair at a single quaternary-care institution



Treatment Protocol:

- Neonates with giant omphalocele undergo gradual reduction of the extraperitoneal viscera using preformed silos with the application of topical agents to promote epithelialization

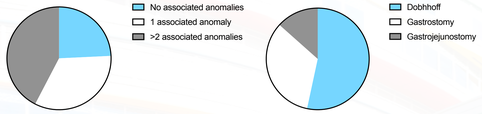


- Reconstruction of the resultant large ventral hernia involves a staged approach with extra-abdominal tissue expansion and mesh reinforced fascial repair
- Abdominal wall reconstruction is performed in conjunction with general surgery to minimize visceral injury
- Postoperative dietary advancement is dictated by general surgery

Results

Defining the cohort:

- N=25 subjects met inclusion
- Born at median gestational age 37.0 weeks, weighing 2,736 grams, via Cesarean section

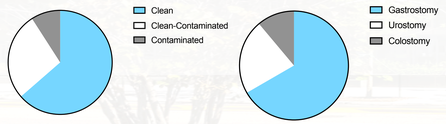


- Median presenting age – 3.3 years
- Median fascial defect length x width – 14 cm x 11.5 cm
- 36% presenting with ostomies

Tissue expansion:

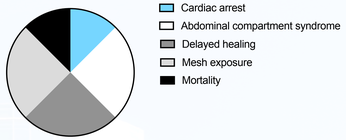
- 56.5% required expansion prior to definitive closure
- ASA class – II (28.6%), III (71.4%)
- Median age at expander placement – 5.9 years
- Median expansion volume – 250 mL
- Median time to fascial repair – 360 days
- Complication rate – 30.8%
- Complications – seroma, infection, extrusion
- Repeat expansion rate – 16.7%

Abdominal wall reconstruction:



Wound Class at Abdominal Wall Reconstruction

- Fascia repair – primary (14.3%), bridging "sandwich" (85.7%)
- Anterior components separation – Unilateral (9.1%), Bilateral (13.6%), None (77.3%)
- Mesh type – Strattice™ resorbable (38.1%), Bard® nonresorbable (9.5%), Vicryl® resorbable + Bard® nonresorbable (52.4%)
- Skin closure – primary (95.5%), use of wound VAC (4.6%)
- Complication rate – 34.8%

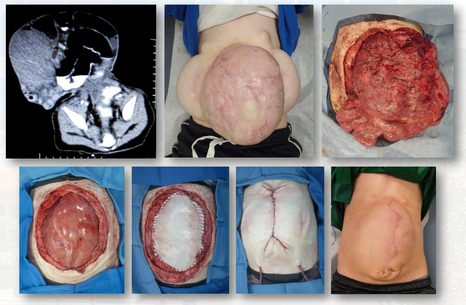


Complications Following Abdominal Wall Reconstruction

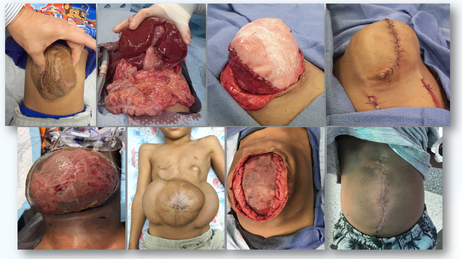
- Reoperative rate – 13.0%
- Hernia recurrence rate – 8.0%

Postoperative care:

- Length of ventilatory support (d) – 6 [3, 20] (median, [IQR])
- Tracheostomy – 17.4%
- Hospital length of stay (d) – 15 [11, 21]
- Length of TPN (d) – 5 [2, 12]
- Time to full feeds (d) – 5 [1, 10]
- Survival to hospital discharge – 91.3%
- Length of follow-up (y) – 3.5 [2.6, 4.6]



Staged reconstruction of giant omphalocele. Abdominovisceral disproportion (UL). Placement of extra-abdominal tissue expanders at age 9 years (UM). Removal of hernia sac with resulting visceral block (UR). Bridging Vicryl® mesh inset to prevent visceral adherence to overlying Strattice™ mesh (LL). Primary closure of soft tissues with 3-year postoperative result (LR).



Abdominal compartment syndrome. 4-year-old male presenting with giant omphalocele without significant soft tissue deficiency. Note the extra-abdominal liver and visceral contents. Initial repair was performed using a bridging Strattice™ mesh and primary closure of non-expanded overlying soft tissues. Postoperative course was complicated by life-threatening liver compression requiring emergent takedown of repair. Following recovery, he underwent staged expansion followed by "sandwich" repair using Vicryl® and Bard® bridging mesh. Three years following staged reconstruction, he is doing well without complication.

Discussion

- Despite a high incidence of complications, giant omphaloceles may be successfully reconstructed with a combination of tissue expansion and mesh implantation
- The amount of expansion needed may be estimated by the trunk circumference above and below the omphalocele (i.e., surgical scar to surgical scar around the posterior trunk)
- Anterior components separation does not allow for reliable primary fascial repair in the midline of giant omphalocele
- When compared to biologic mesh repair, use of bridging Vicryl® mesh as a deep, "peritoneal" layer with an overlying permanent mesh may be preferable given lower rates of seroma, tissue attenuation, bulge, and hernia recurrence
- The lifetime risk associated with permanent mesh reconstruction remains unknown, although we have not observed mesh infections or enterocutaneous fistulae to date

Conclusions

- Reconstruction of large omphalocele defects using tissue expansion and mesh implantation offers a durable solution, with low recurrence
- Although rare, families should be counseled regarding potential devastating complications in this high-risk group