Giant Omphaloceles

Overview, Physiological Sequela and the Dilemma of Surgical Management Options

The Hendren Project

Abdominal Defects Center Webinar

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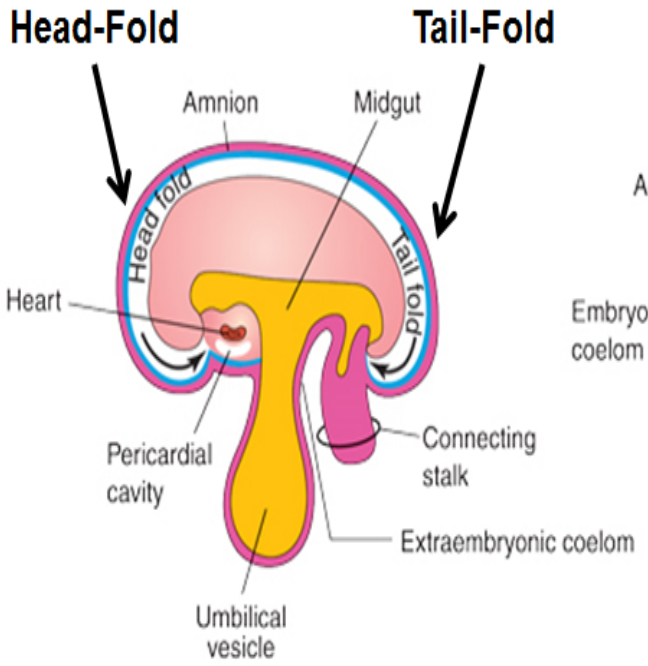
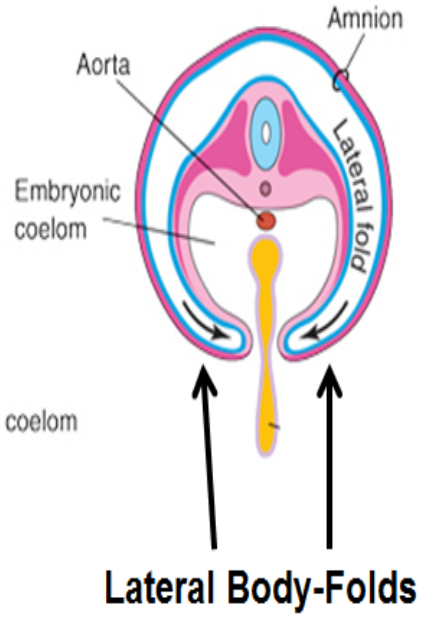
Embryology

- ✓Abdominal wall defect of variable size
- ✓Omphalocele results from defect in migration of the lateral folds very early in embryogenesis (~ 3 weeks) to form the umbilical ring
- ✓Always occurs at the umbilicus
- ✓Rectus muscles insert more laterally on costal margins
- ✓Due to early event, often accompanied by other defects





Comparison b/w Gastroschisis & Omphalocele

	Gastroschisis	Omphalocele
Location	Right of umbilicus	At umbilicus
Sac	No	Yes (may be ruptured)
Contents	Small intestine/colon – may be stomach	Liver, intestines, spleen, gonads
Etiology	Failure of umbilical coelom development -> rupture	Failure of lateral folds
Associated anomalies	Intestinal atresia	Chromosomal, cardiac, pulmonary hypoplasia, pulmonary and systemic hypertension
Outcomes	Good (as long as not significant bowel loss)	Dependent on size, pulmonary hypoplasia and hypertension and associated anomalies
Maternal age	Younger	Average
Mode of delivery	Vaginal	Cesarean preferred
Surgical Management	Urgent	Not urgent

Anatomy

- ✓ Covered by sac (peritoneum, Wharton's jelly, and amnion)
- ✓ Can be centered in the upper, mid or lower abdomen
  - Epigastric Omphalocele (cephalic defect) → Pentalogy of Cantrell
  - Central Omphalocele (lateral folds) → classic omphalocele
  - Hypogastric Omphalocele (caudal folds) → Lower midline syndrome (cloacal exstrophy)
- ✓ Umbilical cord is inserted eccentrically
- ✓ Omphaloceles may be small (hernia of the cord) or Giant
- ✓ Omphalocele sac may rupture in utero, during labor or after delivery
- ✓ Prenatal rupture occurs in 10-20% of cases

Associated Anomalies

- ✓Omphaloceles are associated with anomalies in 50-70% of cases
- ✓Chromosomal abnormalities in 20-30% (trisomy 21,18,13)
- ✓Beckwith Wiedemann Syndrome
- ✓Musculoskeletal abnormalities

A.D.A.M.

Giant Omphalocele

Historically, they have been defined by various criteria: diameter of sac, diameter of abdominal wall defect, inability to primarily close the defect, tissue defect > 5cm, liver containing sac and volume disproportion b/w abdominal viscera and cavity

✓ Definition: Lack of consensus .. Defect >5cm or > 50% liver herniation

Physiological & Anatomical Sequelae

- ✓ Pulmonary Hypoplasia
- ✓ Pulmonary Hypertension
- ✓ Systemic Hypertension
- ✓ Inguinal hernias and undescended testicles
- ✓ GERD

Table 2 Results of spirometry measurements in GO children

	% Predicted[a]	*P* value[b]
FVC	61.1 ± 18.5 (36-102)	**.03**
$FEV_{0.5}$	65.9 ± 21.3 (44-119)	**<.01**
$FEV_{0.5}$/FVC	108.5 ± 13.4 (77-123)	.27
$FEF_{25\%-75\%}$	84.9 ± 29.15 (46-145)	.61

Bold indicates statistical significance.
[a] Data presented as mean ± SD (range).
[b] Compared with previously published normative values [25].

Danzer E., et al., 2012

Giant Omphalocele

Physiological & Anatomical Sequelae

✓Pulmonary Hypertension

Table 2
Clinical outcomes.

	No pulmonary hypertension ($n = 34$)	Pulmonary hypertension ($n = 20$)	p Value
NICU length of stay[a]	57.3 ± 35.2	152.3 ± 97.4	<0.0001
Staged closure	22 (64.7%)	11 (55%)	0.3934
Duration of mechanical ventilation[a]	16.5 ± 14.9	45.3 ± 28.9	0.0008
Requirement for HFOV[a]	3 (8.8%)	7 (35%)	0.0281
Requirement for tracheostomy[a]	2 (5.9%)	8 (40%)	0.0032
Requirement for home oxygen[a]	5 (14.7%)	12 (60%)	0.0004
Reactive airway disease[a]	13 (38.2%)	15 (75%)	0.0239
Gastroesophageal reflux	25 (73.5%)	19 (95%)	0.1308
Requirement for feeding tube at time of NICU discharge[a]	15 (44.1%)	16 (80%)	0.0243
Nissen fundoplication	1 (2.9%)	4 (20%)	0.0572
Nitric oxide therapy[a]	0 (0%)	13 (65%)	<0.0001
Chronic pulmonary vasodilator therapy[a]	0 (0%)	9 (45%)	<0.0001
Survival[a]	34 (100%)	17 (85.0%)	0.0460

[a] Indicates parameters found to be statistically significant ($p \leq 0.05$).

Table 3
Prenatal imaging features predict pulmonary hypertension.

	No pulmonary hypertension ($n = 34$)	Pulmonary hypertension ($n = 20$)	p Value
Contents of hernia sac: liver	34 (100%)	20 (100%)	1.0000
Contents of hernia sac: bowel	32 (94.1%)	19 (95%)	1.0000
Contents of hernia sac: stomach[a]	13 (38.2%)	12 (60%)	0.0322

[a] Indicates parameters found to be statistically significant ($p \leq 0.05$).

E.A. Partridge et al. / Journal of Pediatric Surgery 49 (2014) 1767–1770

✓Systemic Hypertension

Systemic hypertension in giant omphalocele: An underappreciated association

William H. Peranteau *,[1], Sasha J. Tharakan [1], Emily Partridge, Lisa Herkert, Natalie E. Rintoul, Alan W. Flake, N. Scott Adzick, Holly L. Hedrick

Table 2
Hypertensive episodes.

	meet criteria for htn	% htn pts w systolic htn	% htn pts w diastolic htn	% hospital days w systolic htn	% hospital days w diastolic htn	% of htn pts w preop htn	% htn pts w postop htn
Schuster	82% (27/33)	96% (26/27)	81.5% (22/27)	18.3%	4%	0% (0/27)	100% (27/27)
Delayed	67% (8/12)	100% (8/8)	50% (4/8)	31.1%	3%	75% (6/8)	80% (4/5)[a]
All GO	78% (35/45)	97% (34/35)	74% (26/35)	21.3%	8.3%	17% (6/35)	97% (31/32)[a]
Control	15% (3/20)	100% (3/3)	0% (0/3)	17%	0%	33% (1/3)	100% (3/3)
P (Schuster vs. delayed)	0.23			0.35	0.34		
P (All GO vs. Control)	<0.001			0.38	N/A		

% hospital days with htn only evaluates pts that met criteria for htn and indicates the number of days out of the entire hospital stay with an event.
htn, hypertension; pts, patients.
[a] 3 patients in the delayed group did not have post-op blood pressures recorded thus the denominator is different.

- 78% of GO patients demonstrate evidence of systemic hypertension
- HTN may be related to increased abdominal pressure with stimulation of the renin angiotensin system

Giant omphaloceles: surgical management and perinatal outcomes

Akinkuoto A.C. et al., JOURNAL OF SURGICAL RESEARCH 198 (2015) 388–392

- GO defined as liver herniation > 50%
- Patients classified as isolated, omphalocele with minor or major associated anomalies
- Major anomalies: cardiac, genitourinary or anorectal defects requiring immediate management, CDH, alveolar capillary dysplasia, and/or chromosomal aneuploidy or duplication

Table 1 – Outcomes of fetuses and neonates with omphalocele.

Outcomes	Isolated	Minor	Major	*P* value
Total patient number	23	41	31	N/A
Live born	19	39	27	N/A
GO, *n* (%)	11 (52)	25 (61)	23 (74)	N/A
Postnatal mortality, *n* (%)	0 (0)	7 (17)	11 (41)	0.006
Duration of mechanical ventilation, d median (IQR)	0 (0–2)	0 (0–5)	36 (22–89)	0.04
Hospital LOS, d median (IQR)	18 (9.5–38.5)	28.5 (13–32.5)	157 (48.5–185)	<0.001
O_2 at 30 DOL, *n* (%)	2/16 (12.5)	9/33 (27.3)	16/18 (88.9)	<0.001

DOL = days of life; IQR = interquartile range; LOS = length of stay; N/A = not applicable; O_2 = oxygen.

Table 2 – Characteristics and outcomes of patients with GO managed with early *versus* delayed closure.

Characteristics and outcomes	Early repair ($n = 5$)	Delayed repair ($n = 35$)	P value
GA at birth (wk), mean ± SD	37.9 ± 2.2	36.4 ± 3.1	0.306
Birth weight, g mean ± SD	2693 ± 540	2695 ± 836	0.864
Fetal center evaluation, *n* (%)	1 (5)	25 (71.4)	0.043
Chromosomal anomalies, *n* (%)	0 (0)	4 (11.4)	1.00
Minor anomalies, *n* (%)	3 (60)	14 (40)	0.634
Major anomalies, *n* (%)	2 (40)	18 (51.4)	1.00
Length of intubation, days median (IQR)	12 (6–27)	0 (0–3)	0.932
Length of stay, d median (IQR)	73 (50–143)	33 (21–103)	0.857
Oxygen at 30 DOL, *n* (%)	2/5 (40)	16/29 (55.2)	1.00
Mortality at 6 mo, *n* (%)	0/5 (0)	8/34 (23.5)	0.563

DOL = days of life; GA = gestational age; IQR = interquartile range; SD = standard deviation.

There was no difference between early and delayed repair except for fetal center evaluation that resulted in more delayed repair

Management of giant omphaloceles: A systematic staged surgical vs. nonoperative delayed closure☆

Bauman B., et al Journal of Pediatric Surgery 51 (2016) 1725–1730

Table 2
LOS (days), time to full feeds (days), and mortality presented as the range and mean of the studies listed in Table 1.

Therapy	LOS (range, mean, (SD))	Time to Full Feeds (range, mean, (SD))	Mortality (% range, mean, (SD))
Topical Silver (n = 4)	(n = 3) 20–78, 40 (32.9)	(n = 2) 6–8, 7 (1.4)	(n = 4) 9.5–20, 7.4 (9.5)
Topical Betadine (n = 3)	(n = 3) 14–34, 27 (11.27)	(n = 2) 8.5–33, 20.7 (17.3)	(n = 3) 16–25, 21.5 (4.8)
Manuka honey (n = 1)	66	13	20
2% Aqueous eosin (n = 1)	21	NR	25
Negative pressure wound therapy (n = 1)	70	19	none
Surgical silo (n = 3)	(n = 2) 42–42, 42 (0)	(n = 2) 12–35, 23.5 (16.3)	(n = 3) 25–46, 37 (10.8)
Interposition mesh (n = 1)	10.2	NR	none

Not all studies within each therapy provided data on all three endpoints.
N = number of studies; NR = not reported; LOS = length of stay (days).

Table 3
Comparison of endpoints between surgical and delayed closure methods.

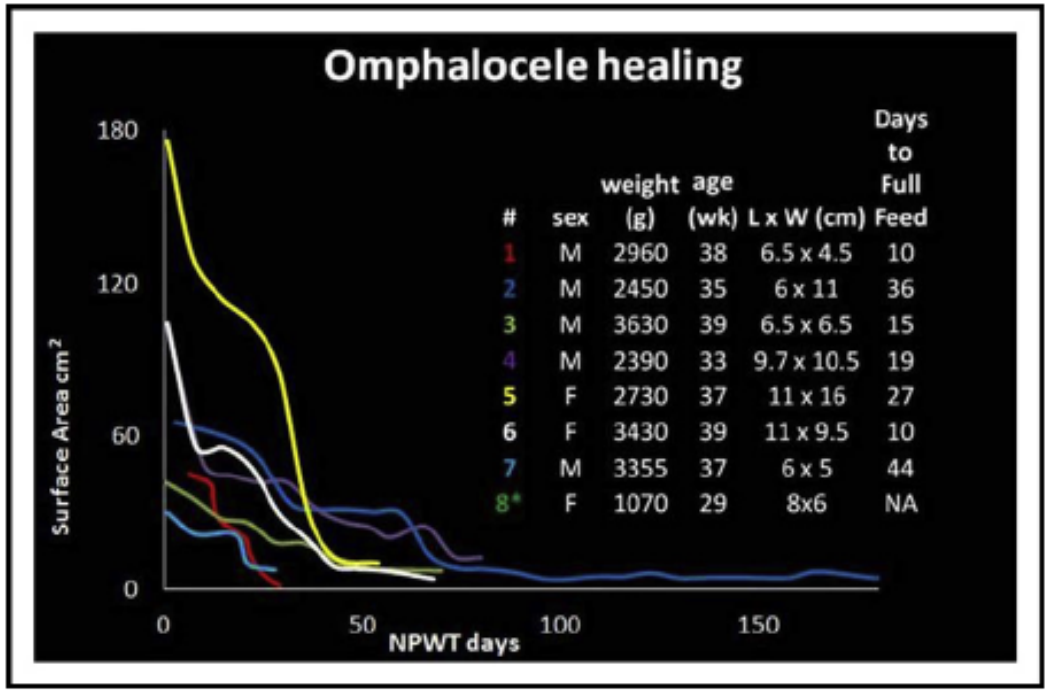
Endpoint	Surgical closure	Delayed closure
LOS	31.8 (17.7) (n = 3)	39.5 (24.3) (n = 9)
Time to full feeds	23.5 (16.3) (n = 2)	14.6 (10.2) (n = 6)
Mortality	15/64 (23.4%) (n = 4)	62.25/286 (21.8%) (n = 10)

LOS and time to full feeding represent mean values of reported data with their associated standard deviation and the number of studies involved in the calculation.
N = number of studies; LOS = length of stay.

Negative pressure wound therapy for initial management of giant omphalocele

Aldridge B., et al., The American Journal of Surgery (2016) 211, 605-609

- No complications or deaths secondary to treatment
- No wound infections, no fistulas and no sac rupture
- Median time to 1st enteral feed was 19 days
- Median length of hospital stay 70 days
- TTT can be begin on the first day of life and is usua complete by 2 months



#	sex	weight (g)	age (wk)	L x W (cm)	Days to Full Feed
1	M	2960	38	6.5 x 4.5	10
2	M	2450	35	6 x 11	36
3	M	3630	39	6.5 x 6.5	15
4	M	2390	33	9.7 x 10.5	19
5	F	2730	37	11 x 16	27
6	F	3430	39	11 x 9.5	10
7	M	3355	37	6 x 5	44
8*	F	1070	29	8x6	NA

Figure 2 Decrease in surface area over time with NPWT with associated patient data. (*) Patient died after 7 days of treatment of unrelated causes.