

# Omphalocele Clinical Guideline



**Inclusion Criteria:** Any neonate born with an omphalocele regardless of size or gestation

## Available Resources

- Omphalocele PFE
- Omphalocele wrap video

## Prenatal Recommendations

### Antepartum Care:

- Associated with elevated maternal serum alpha-fetoprotein level
- Ultrasound suspicious for omphalocele: refer to Maternal-Fetal Medicine for detailed ultrasound exam
  - MFM ultrasound to include evaluation for other abnormalities, description of organ involvement, and preliminary counseling/consultation
- Consider need for fetal MRI to further evaluate anatomy and lung volumes
- Referral to Pediatric Cardiology for fetal echocardiogram at approximately 22 weeks
- Referral to Genetics with discussion of amniocentesis
- Referral to Pediatric Surgery
- Multidisciplinary care meeting to involve OB, MFM, Neonatology, Genetics and Pediatric Surgery

### Delivery:

- Recommended delivery at a medical center with a Level IV NICU
- Vaginal delivery *may* be possible in small omphaloceles. Cesarean deliveries warranted for giant omphaloceles to prevent omphalocele rupture and trauma to enclosed organs, specifically liver
- Encourage full-term delivery; delivery may be warranted earlier for fetal and/or maternal indications

## Delivery Room Anticipation and Resuscitation

### Pre-briefing:

- Team huddle to discuss plan of care and clearly define team member roles
- Advanced preparation of supplies including equipment for intubation, 8 Fr (preterm) and 10 Fr (term) Salem sump (SS), bowel bag, and potential normal saline fluid boluses and resuscitative medications

### Delivery/Resuscitation:

- Placement of 8 Fr (preterm) and 10 Fr (term) Salem sump (SS) orogastric or nasogastric tube to low intermittent suction (LIS)
- Assess respiratory status. Small omphaloceles may not require additional support, whereas large omphaloceles may require CPAP or intubation
  - Giant omphaloceles are more likely to have pulmonary hypoplasia and often respond to low volume and rapid rate ventilation

## (Continued) Delivery Room Anticipation and Resuscitation

### Maintain integrity of omphalocele sac:

- Utilize sterile gloves when handling
- Place neonate in bowel (Lahey) bag lined with small amount of warm sterile saline solution
- Position neonate side lying while supporting the omphalocele with blanket rolls to optimize perfusion and prevent compression of blood vessels

### Antibiotics:

- Ampicillin and Gentamicin if needed for sepsis risk factors or in event of sac rupture

## Upon NICU Arrival Monitoring

### Respiratory:

- Lung hypoplasia and decreased lung volumes often require respiratory support
- Risk of pulmonary hypertension in patients with giant omphaloceles
  - Monitor pre and post ductal saturations

### Cardiovascular:

- Echocardiogram to evaluate for cardiac anomalies and assess for pulmonary hypertension

### IV Fluids and Access:

- PICC for long-term central venous access in patients with giant omphalocele
- PIV for patients with small omphalocele with anticipated early primary closure
- If sac is intact: Initiate D10W at 80 mL/kg/day
- If sac is ruptured: Initiate D10W, may need up to 120 mL/kg/day and provide NS boluses for replacement fluids
- Hypoglycemia often seen in neonates with Beckwith Wiedemann Syndrome

### Antibiotics: Clinical use of antibiotics not empirical

- May consider 48 hr sepsis rule out antibiotic treatment in presence prenatal risk factors, symptomatic patient, or ruptured omphalocele

### Gastrointestinal:

- NPO and MOPs (Milk oropharyngeal) until hemodynamically stable
- Continue SS to LIS
  - Consider replacement of high-volume SS output (>15 mL/kg/shift) with ½ NS (replace 1 mL/mL output over 4 hours)

### Genetics:

- Consult on admission
- Anticipate sending chromosomal microarray analysis
- Consider AFP level if suspicion of Beckwith-Wiedemann Syndrome

### Skin:

- Skin wound ostomy team (SWOT) consult on admission
- See management options section below

## Surgical Management

### Small or Medium sized defect

Primary closure in the OR when safe for neonate early in life Grade A

### Large Defect (≥ approx. 5 cm)

- “Paint and Wait” Technique: *Most commonly* used method Grade A
- Goal: promote granulation and epithelization of sac
  - Obtain SWOT consult
  - Daily dressing changes with xeroform gauze and Aquaphor
  - Consider using Dakin’s solution or betadine if sac infection is suspected.
- Grade B
- Abdominal wall closure/ ventral hernia closure later in life

## Surgical Preparation

- Pre-operative labs completed within 24 hours prior to surgery and evaluated:
  - CBC with differential, BMP at 12 - 24 hours of life.
  - Blood gas
  - Type & Cross (if not already completed)
- Pre-operative echocardiogram Grade B
- Order desired blood products to be on hold for the OR
  - With large defect closures: Packed red blood cells, platelets, FFP (20 mL/kg of each)
- Ensure adequate IV access (2 PIV’s or 1 PIV and 1 PICC) for administration of blood products and medications
- Fluids per pre-operative checklist
- Postoperative pain medications using pain guideline 2
- Anesthesia to administer pre-operative antibiotics within 1 hour of incision

## Post-Operative Care Management

### Monitoring:

- Monitor for signs and symptoms of compartment syndrome: decreased distal pulses, abdominal distention, decreased urine output, skin discoloration Grade A

### Gastric Decompression:

- 8 Fr or 10 Fr SS tube to LIS Grade A
- NPO and MOPs

### Diagnostic Studies/Labs:

- CXR immediately post-operatively
- Temperature, blood gas and glucose level within 1 hour post-operatively
- CBC / BMP in the AM post-operative day #1
  - Or earlier if clinically warranted

### Fluid Management:

- Continue pre-operative management of fluids

### Antibiotics:

- 24-hour postoperative prophylaxis in the absence of any complications or symptomatic patient grade B
- Omphalocele closure includes bowel surgery: Cefoxitin
- Omphalocele closure does not include bowel surgery: Cefazolin

### Pain Management:

- Pain medications using surgical pain guideline 2 Grade A

### Skin care:

- Use of negative pressure wound vac used in some cases.
- Notify surgery of any signs of erythema, drainage, bleeding, or wound concerns
- If sutures placed, contact surgery for removal plan / date
  - After sutures removed or surgical site has healed, apply Mepitel One (preemies) / Mepitac (post-term) to surgical sites once healed for scar therapy
  - Change or re-apply after each bath

## Post-Operative Feeding

- Surgical team clearance and return of bowel function (stool and tolerance of SS removal); in patients with significant pulmonary HTN and/or cardiac anomalies assure adequate bowel perfusion and acid-base balance prior to feeding \*
- Use human milk (maternal/parent's or pasteurized donor), PO vs gavage per CGA/respiratory status
  - Surgical Feeding Guideline 1: Consider for preterm infants (< 34<sup>0/7</sup> weeks GA and/or < 2 kg) or infants with significant dysmotility or bowel resection \*
  - Surgical Feeding Guideline 2: Consider for infants > 34<sup>6/7</sup> weeks GA and uncomplicated course
  - If unable to progress to full oral feeds, gastrostomy tubes are contraindicated and home NGT may be necessary (consult GI)

## Considerations for Management

- Incidence is 1 in 4,000 - 6,000 births
  - 3:1 increased prevalence in males
- Cardiac anomalies occur in 30 - 50% of infants with omphalocele. Most commonly seen are Tetralogy of Fallot and ASD
- Extreme precaution should be taken to reduce risk of sac rupture. Minimize sac contact and ensure proper wrapping techniques are used.
- Associated anomalies (more common in patients with giant omphalocele):
  - Pentalogy of Cantrell (abdominal wall defect, ectopia cordis, sternal cleft, diaphragmatic hernia, cardiac anomalies)
  - OEIS complex (omphalocele, exstrophy of the bladder, imperforate anus, spinal anomalies)
  - Beckwith-Wiedemann Syndrome (macroglossia, hemihypertrophy, hypoglycemia, organomegaly)
  - Trisomies 13 -18
- All infants are malrotated
- Long term complications include: pulmonary hypoplasia and hypertension, chronic lung disease, gastroesophageal reflux disease, malrotation with volvulus, feeding difficulties, failure to thrive
- If going home with omphalocele prior to abdominal closure, consider a custom protective shell device to avoid sac rupture and special car seat to accommodate omphalocele

## Omphalocele Clinical Guideline *References*

- Ayub, S. S., & Taylor, J. A. (2019). Cardiac anomalies associated with omphalocele. *Seminars in Pediatric Surgery*, 28(2), 111-114. <https://doi.org/10.1053/j.sempedsurg.2019.04.002> (Level III)
- Bauman, B., Stephens, D., Gershone, H., Bongiorno, C., Osterholm, E., Acton, R., Hess, D., Saltzman, D., & Segura, B. (2016). Management of giant omphaloceles: A systematic review of methods of staged surgical vs. nonoperative delay closure. *Journal of Pediatric Surgery*, 51(10), 1725-1730. <https://doi.org/10.1016/j.jpedsurg.2016.07.006> (Level I)
- Duggan, E., & Puligandla, P. S. (2019). Respiratory disorders in patients with omphalocele. *Seminars in Pediatric Surgery*, 28(2), 115-117. <https://doi.org/10.1053/j.sempedsurg.2019.04.008> (Level II)
- Hutson, S., Baerg, J., Deming, D., St Peter, S. D., Hopper, A., & Goff, D. A. (2017). High Prevalence of Pulmonary Hypertension Complicates the Care of Infants with Omphalocele. *Neonatology*, 112(3), 281-286. <https://doi.org/10.1159/000477535> (Level II)
- Partridge, E. A., Hanna, B. D., Panitch, H. B., Rintoul, N. E., Peranteau, W. H., Flake, A. W., Scott Adzick, N., & Henrick, H. L. (2014). Pulmonary hypertension in giant omphalocele infants. *Journal of Pediatric Surgery*, 49(12), 1767-1770. <https://doi.org/10.1016/j.pedsurg.2014.09.016> (Level III)
- Peranteau, W. H., Tharakan, S. J., Partridge, E., Herkert, L., Rintoul, N. E., Flake, A. W., Adzick, N. S., & Hedrick, H. L. (2015). Systematic hypertension in giant omphalocele: An underappreciated association. *Journal of Pediatric Surgery*, 50(9), 1477-1480. <https://doi.org/10.1016/j.jpedsurg.2015.02.051> (Level I)
- Verla, M. A., Style, C. C., & Olutoye, O. O. (2019). Prenatal diagnosis and management of omphalocele. *Seminars in Pediatric Surgery*, 28(2), 84-88. <https://doi.org/10.1053/j.sempedsurg.2019.04.007> (Level II)
- Wagner, J. P., & Cusick, R. A. (2019). Paint and wait management of giant omphaloceles. *Seminars in Pediatric Surgery*, 28(2), 95-100. <https://doi.org/10.1043/j.sempedsurg.2019.04.005> (Level II)