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:
- 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
- Referral to Pediatric Cardiology for fetal echocardiogram @ approximately 22 weeks.
- Consider need for fetal MRI for further evaluation of anatomy and lung volumes
- Genetics consultation 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 Level IV medical center
- 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
- Encouragement of full term delivery but delivery may be warranted earlier for fetal and/or maternal indications

Delivery Room Anticipation and Resuscitation:
Pre-briefing:
- Team huddle with discussion of plan of care and clearly defined team member roles
- Advanced preparation of supplies including equipment for intubation, 8 fr (preterm) and 10 fr (term) salem sump, bowel (lahey) bag, and potential normal saline fluid boluses and resuscitative medications.

Delivery/ Resuscitation:
- Placement of 8 fr (preterm) and 10 fr (term) salem sump orogastric or nasogastric tube to low intermittent suction
- Assess respiratory status. Small omphaloceles may not require additional support, whereas large omphaloceles may require CPAP or intubation.
  - Giant omphaloceles more likely to have pulmonary hypoplasia and often respond to low volume and rapid rate ventilation
- PIV access. No umbilical lines
- Dextrose stick.
- Initiate IV fluids D10W at 80 ml/kg/day
  - Assess need for additional NS fluid boluses; most often needed in events of sac rupture

Approved by Evidence-based Medicine Committee 9/19/18
Reassess the appropriateness of Care Guideline as condition changes and 24 hours after admission. This guideline is a tool to aid clinical decision making. It is not a standard of care. The physician should deviate from the guideline when clinical judgement so indicates.
(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 sidelying 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 if early primary closure not possible
- If sac is intact: total fluid limit of 80 ml/kg/day
  - If sac is ruptured: may need up to 120 ml/kg/day and 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 until hemodynamically stable
- Salem sump to low intermittent wall suction
- Consider replacement of high volume salem sump output (≥ 30 ml/kg/day)
  - ½ ml replacement to 1 ml output with 0.45 NaCl
Genetics:
- Genetics consult at admission
  - Anticipate sending chromosomal microarray analysis
  - Consider AFP level if suspicion of Beckwith Wiederman Syndrome
Skin:
- Wound care consult at admission
- See management options section below
### Surgical Management:

<table>
<thead>
<tr>
<th>Small or Medium sized defect</th>
<th>Large Defect (≥ approx. 5 cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary Closure in the OR when safe for neonate early in life</td>
<td>“Paint and Wait” Technique</td>
</tr>
<tr>
<td></td>
<td>Most commonly used method</td>
</tr>
<tr>
<td></td>
<td>Goal: promote granulation and epithelialization of sac using escharotic agent (product containing silver) and gauze wrap</td>
</tr>
<tr>
<td></td>
<td>Daily dressing changes with silver sulfadiazine (silvadene), xeroform gauze and gauze wrap</td>
</tr>
<tr>
<td></td>
<td>In some cases Mepitel AG may be used in place of silver sulfadiazine and less frequent dressing changes may be needed</td>
</tr>
<tr>
<td></td>
<td>Abdominal wall closure/ ventral hernia closure later in life</td>
</tr>
</tbody>
</table>

### Surgical Preparation:

- Pre-operative labs completed within 24 hours prior to surgery and evaluated:
  - CBC with differential
  - BMP at 12-24 hrs of life
  - Blood gas
  - Type & Cross (if not already completed)
- Pre-operative echocardiogram
- Order desired blood products to be on hold for the OR
  - Packed red blood cells, platelets, FFP (20 ml/kg of each) with large defect closures
- Ensure adequate IV access (2 PIV’s or 1 PIV and 1 PICC) for administration of blood products and medications.
- Replace TPN with D10 ½ NaCl or D5 ½ NaCl to avoid electrolyte imbalances.
- Anesthesia to administer pre-operative antibiotics within 1 hour of incision
- Make appropriate post-operative pain control plan and pre-order appropriate medications
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 an 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:

- Pentalogy of Cantrell (abdominal wall defect, ectopia cordis, sternal cleft, diaphragmatic hernia, cardiac anomalies)
- OEIS complex (omphalocele, exstrophy of the bladder, imperforate anus, spinal anomaly)
- Beckwith-Wiedemann Syndrome (macroglossia, hemihypertrophy, hypoglycemia, organmegaly)
- Trisomies 13-18

Giant omphaloceles associated with higher frequency of anomalies

All infants are malrotated

Post-Operative Care Management:

Monitoring:

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

Gastric Decompression:

- 8 fr or 10 fr salem sump tube to low intermittent suction
- NPO with salem sump until full return of bowel function

Diagnostic Studies/Labs:

- CXR immediately post-operative
- Temperature, blood gas and glucose level within 1 hour post-operative
- 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
  - Omphalocele closure includes bowel surgery: cefoxtin
  - Omphalocele closure does not include bowel surgery: ancef

Pain Management: as clinically indicated

- IV acetaminophen Q 6 hrs for 24 hrs and reevaluate pain management plan daily
- Utilize PRN dosing of narcotics prior to initiating narcotic infusion
  - Morphine or fentanyl infusion, versed PRN if indicated

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 are 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

Diagnostic Studies/Labs:

- CXR immediately post-operative
- Temperature, blood gas and glucose level within 1 hour post-operative
- 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
  - Omphalocele closure includes bowel surgery: cefoxtin
  - Omphalocele closure does not include bowel surgery: ancef

Pain Management: as clinically indicated

- IV acetaminophen Q 6 hrs for 24 hrs and reevaluate pain management plan daily
- Utilize PRN dosing of narcotics prior to initiating narcotic infusion
  - Morphine or fentanyl infusion, versed PRN if indicated

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 are 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
References


