Omphalocele Clinical Guideline

CHOC Children's
Children's Hospital of Orange County

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.

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(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

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Surgical Management:

Small or Medium sized defect

Primary Closure in the OR when safe for neonate early in life

Large Defect
(≥ approx. 5 cm)

"Paint and Wait" Technique

- *Most commonly* used method
- Goal: promote granulation and epithialization of sac using escharotic agent (product containing silver) and gauze wrap
  - Daily dressing changes with silver sulfadiazine (silvadene), xeroform gauze and gauze wrap
  - In some cases Mepitel AG may be used in place of silver sulfadiazine and less frequent dressing changes may be needed
- 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 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

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Post-Operative Care Management: remember to complete the VON QI OR Handoff Tool

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 revaluate 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

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 anomolies)
  - OEIS complex (omphalocele, exstrophy of the bladder, imperforate anus, spinal anomolie)
  - Beckwith-Wiedemann Syndrome (macroglossia, hemihypertrophy, hypoglycemia, organmegaly)
  - Trisomies 13-18
- Giant omphaloceles associated with higher frequency of anomalies
- All infants are malrotated
- Monitor TFT's in extended exposure to betadine
- Long term complications include: pulmonary hypoplasia and hypertension, chronic lung disease, GERD, malrotation with volvulus, feeding difficulities, failure to thrive
- If going home with omphalocele prior to abdominal closure, consider a custom protective shell device to avoid sac rupture.

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