OMPHALOCELE

Surgical Specialty:	Pediatric Surgery
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Background:

- General Considerations
  - An omphalocele is a midline congenital abdominal wall defect that results from a failure of the extraembryonic gut to return to the abdominal cavity during the fifth to tenth week after conception.[1]
  - The partial or total extrusion of abdominal contents is covered by a membranous sac.
  - There is an increased risk of heat loss, fluid loss, and infection.
- Patient Considerations
  - An omphalocele is not a surgical emergency, and neonates should be medically stabilized in the NICU before surgical management. This includes appropriate fluid resuscitation, intubation and ventilatory support for respiratory failure, full-stomach precautions, and adequate nutrition, i.e. total parentral nutrition (TPN).
  - Patients commonly present with cardiac anomalies such as patent ductus arteriosus (PDA), patent foramen ovale (PFO), or ventricular septal defect (VSD).
  - Patients with congenital cardiac lesions may need a Pediatric Cardiology consult, transthoracic echocardiogram (TTE), inotropes, or hemodynamic support before surgical management of the omphalocele.
  - Large omphaloceles can contribute to lung hypoplasia or abnormal thoracic development.
- Associated Comorbidities/Syndromes
  - Often associated with congenital anomalies such as Trisomy 21, Beckwith-Wiedemann syndrome, and Pentalogy of Cantrell.
    - Beckwith-Wiedemann syndrome: Hypoglycemia, macroglossia, congenital heart disease, omphalocele
    - Pentalogy of Cantrell: Sternal defect, congenital cardiac malformation, absence of pericardium, diaphragmatic defect, omphalocele
  - Other associated defects include the following: oral clefts, polyhydramnios, growth restriction, neural tube defects, and GI abnormalities (intestinal atresia, stenosis, malrotation).[1-3]

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - A thorough review of medical conditions, comorbidities, vital signs, labs, directed history, physical exam, and optimization of comorbid conditions.
    - Evaluate the patient's respiratory status, ventilation settings (if intubated), lines, and medications.
        - Establish adequate IV access preoperatively (in NICU) or intraoperatively. Line access can be difficult, and umbilical lines are unavailable.
    - Discuss the risks, benefits, and alternatives with the surgeon and parents before proceeding.
    - Additional labs/tests indicated during work-up
        - Chest X-ray can help identify the extent of pulmonary hypoplasia
        - ABG in mechanically ventilated patients
        - Consider TTE as these patients are at high risk for congenital cardiac lesions
    - Discussions to have with surgeon/family
        - Code Status
        - ECMO Candidate
- Specific or Unique Room Set-Up Requirements
    - Room
        - Warm room temperature to a goal of 85°F
        - Forced air warmer, heat lamp
    - Equipment
        - Various airway devices available, including video laryngoscopy
        - Multiple sizes of endotracheal tubes (ETT)
        - Suction catheter
        - Nasogastric (NG) or orogastric (OG) tubes
    - Drugs/Infusions
        - Availability of calcium gluconate, dopamine infusion, syringe infusion pump
    - Monitors
        - Pre- and post-ductal pulse oximetry can be helpful as many of these patients present with a PDA or PFO.
        - An arterial line can monitor hemodynamics, volume, and acid-base status. The comorbidities and size of the omphalocele should be considered when deciding to place an arterial line.
        - A rectal temperature probe is ideal due to NG or OG tube in place.
    - Blood Availability (if indicated)
        - Blood should be typed and crossed on standby, although it is usually not needed.
        - Warmed blood tubing
    - NICU Bed Availability
        - Patients often come from the NICU and should return to the NICU postoperatively.
    - Other Indicated Resources
        - ECMO standby may be necessary for patients with decompensating congenital cardiac disease or severe respiratory failure.

Intraoperative Considerations:

- General
  - The major concerns are fluid management, metabolic stability, temperature preservation, ventilation, and avoiding abdominal compartment syndrome.
  - Dramatic fluid shifts can occur after surgical removal of the membranous sac. Primarily extracellular fluid (ECF) is lost due to fluid transudation and exudation.
  - Establish adequate IV access. Volume resuscitation should occur with isotonic crystalloids. Lower extremity PIV can be less useful with high intraabdominal pressures.
  - Ventilation can be compromised with abdominal closure.
  - High intraabdominal pressures can alter drug metabolism due to portocaval connections that bypass the liver.
  - Poor renal perfusion can reduce the excretion of drug metabolites.
- Induction
  - If breathing spontaneously, consider rapid sequence induction for larger omphaloceles due to the risk of aspiration.[2]
  - Decompress the stomach with an NG or OG tube. Patients from the NICU typically arrive with either one. Consider leaving it in to help identify tracheal passage and for NICU management.
- Airway
  - Patients can arrive mechanically ventilated or breathing spontaneously.
    - NICU patients frequently arrive with an uncuffed ETT. May need to exchange to cuffed ETT due to high airway pressures during the surgery.
    - If patients arrive with cuffed ETT, confirm ETT position, cuff leak, and ensure adequate oxygenation and ventilation.
  - Patients with Beckwith-Wiedemann syndrome have a large tongue and can be a potentially difficult airway. Consider backup airway modalities, including video laryngoscopy (C-MAC, Glidescope) and fiberoptic.[3]
- Positioning
  - Usually supine with the airway at the head of the bed.
- Maintenance
  - Drugs/Infusions
    - Maintenance
      - Sevoflurane is generally used, with MAC 0.5-1.0 depending on the hemodynamics.
        - For MAC < 0.7, consider supplementing with IV analgesics and ensure adequate paralysis.
      - Avoid nitrous oxide as it can cross the intestinal wall and expand the intestines in a short period of time. This will make abdominal closure difficult.
    - Analgesia
      - Fentanyl 0.5-1 mcg/kg re-dosed every hour as necessary. Clinical judgment and caution are advised due to the risk of apnea.
      - Avoid long-acting opioids like morphine if extubation is anticipated.
    - Paralytics
      - Short to intermediate-acting paralytics such as rocuronium is recommended to help facilitate abdominal wall closure.
      - Most neonates return to the NICU where they are kept sedated and paralyzed for staged closure.

        - Patients with smaller omphaloceles and few or no comorbidities may be a candidate for extubation.
            - Paralytics should not be spared during abdominal closure – however, one should practice judicious administration of paralytics throughout the surgery. Plans for extubation should be discussed with the surgeon.[2]
        - Consider cisatracurium bolus or infusion as patients can have poor renal perfusion from high intraabdominal pressure.
    - TPN
        - Continue during case
    - Lipids
        - May continue or pause infusion
    - Fluids
        - Isotonic crystalloids
        - Consider 5% Dextrose 0.45% NS for maintenance fluids (utilizing the 4-2-1 rule) if the patient is not on TPN.
    - Hemodynamic support
        - Medications to support hemodynamics should be readily available, including calcium gluconate, and dopamine infusion.
- Hemodynamic/Physiologic goals
    - Maintain euvolemia with isotonic crystalloids and colloids
        - Excessive fluid administration with high peak airway pressures can result in a right-to-left shunt across a PDA or PFO, resulting in desaturation.
    - Goal saturation on pulse oximetry >92%
    - Avoid 100% O_2 due to the risk of retinopathy of prematurity
    - Avoid hyperventilation and hypocarbia
    - Maintain mean arterial pressure appropriate for age (typically 40-60 mmHg for full-term neonates)
    - During abdominal closure, closely monitor airway pressures and blood pressure
        - Ventilation should be maintained at a constant peak pressure with measurement of tidal volumes (TV) or keep a fixed TV and measure the peak pressures.
        - The goal is to keep peak plateau pressure < 25 mmHg or peak inspiratory pressure < 35 mmHg.[2,3]
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
    - For small omphaloceles, primary repair may be feasible with a single surgery.
    - Moderate to large omphaloceles are at a higher risk for respiratory compromise. Airway pressure, oxygenation, and ventilation should be closely monitored during surgical closure.
    - Moderate to large omphaloceles increase the risk of abdominal compartment syndrome with circulatory compromise to the kidneys, intestines, and lower extremities. Patients can have lower extremity edema and hypertension. Compression against the IVC can result in significant hypotension.
    - Staged bedside closure may be necessary if neonates are unstable from a cardiopulmonary standpoint.
- Emergence/Disposition
    - Fully awake on emergence
    - Avoid aspiration

    - The majority of patients remain intubated due to respiratory insufficiency and/or staged closure.[4]
- Post-op Care
    - Monitor for signs of increased abdominal pressure: decreased urine output, lower extremity edema or cyanosis, hypotension.

Case-Specific Complications/Pitfalls

- Hypothermia
    - Extensive radiant heat loss can occur through the omphalocele.
    - Use forced warm air convection, heat lamp, and warm the room.
- Hypovolemia
    - Large fluid shifts occur with the removal of the membranous sac. Hypovolemia often persists into the postsurgical period.
    - For boluses, use isotonic crystalloids.
    - For maintenance fluid, consider 5% Dextrose 0.45% NS. Monitor for hyperglycemia.
- Respiratory insufficiency
    - Large omphaloceles and progressive abdominal closure can function as a restrictive lung physiology. Patients require mechanical ventilation during the pre and postoperative course.
- Pneumothorax
    - Abdominal closure can result in elevated airway pressures with subsequent barotrauma.
    - Monitor airway pressures during abdominal closure. The goal is to maintain peak inspiratory pressure < 35 mmHg.
- Pulmonary edema
    - Large-volume resuscitation can result in pulmonary edema. Resuscitation should be balanced between crystalloids and colloids.
- Abdominal compartment syndrome
    - Abdominal closure with an elevated abdominal pressure >20 mmHg can affect multiple organs.
    - Surgeons will perform a staged closure if there is a concern. Patients should be monitored for signs of organ failure after each staged closure.
- Intestinal ischemia
    - Can occur as a sequela of sepsis or abdominal compartment syndrome
- Sepsis and metabolic acidosis
    - Patients are at a higher risk of bowel infection after amniotic membrane removal.

References

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