MYELOMENINGOCELE REPAIR

Surgical Specialty:	Neurosurgery
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Background:

- General Considerations
  - Failed closure of the neural tube during gestational weeks 5-6 results in a form of myelodysplasia which presents as a visible, fluid-containing sac.
  - Myelomeningocele (MMC) lesions contain CSF/meninges and neural tissue most commonly seen in the lumbosacral region.
  - Patients are at high risk of infection and tethering of the spinal cord which can result in paralysis, sensory loss, and bladder/bowel/sexual dysfunction. Subsequent Chiari malformation is common.
  - Diagnosis is possible as early as the first trimester of pregnancy with amniocentesis and ultrasonography. Postnatal diagnosis can be completed with physical examination and ultrasonography.
  - Early surgical intervention is essential to preventing further neurological deficits.
  - In human studies, intrauterine closure of fetal MMC has been shown to be more effective than postnatal surgery in improving and preserving neurological function as well as reducing the incidence of hindbrain herniation, Chiari malformation, and need for ventriculoperitoneal shunting.[1-2]
  - Postnatal surgery is usually done within 24 to 48 hours after birth to decrease the risk of infections[3,4] and mitigate further neurological sequalae.
- Patient Considerations
  - Disease Specific Considerations
    - Universal latex allergy precautions should be utilized.
  - Associated Comorbidities
    - Hydrocephalus and Chiari malformations may require shunting.
    - A cerebellar tonsillar herniation and caudal displacement of brain stem characterizes the condition.
    - Scoliosis (with resulting, restrictive lung disease), renal anomalies, intestinal malrotation, tracheoesophageal malformations, cardiac abnormalities, and syringomyelia are also associated with this pathology.[5]

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - A thorough and organized pre-anesthetic evaluation is critical to the identification of any associated comorbidities.
  - Early communication with the neurosurgical team is critical to planning a timely intervention.
  - Discuss anesthetic risks/benefits with family.
  - Additional labs/tests indicated during work-up
    - Type and Screen
    - Cardiac echo if concern for cardiac abnormalities
  - Discussions to have with surgeon/family
    - Code Status
- Specific or Unique Room Set-Up Requirements
  - Positioning
    - Induction can be conducted in the lateral or supine position. If the supine position is selected special consideration must be used to avoid any trauma/compression to the defect using a “doughnut” shaped cushion or other custom equipment.

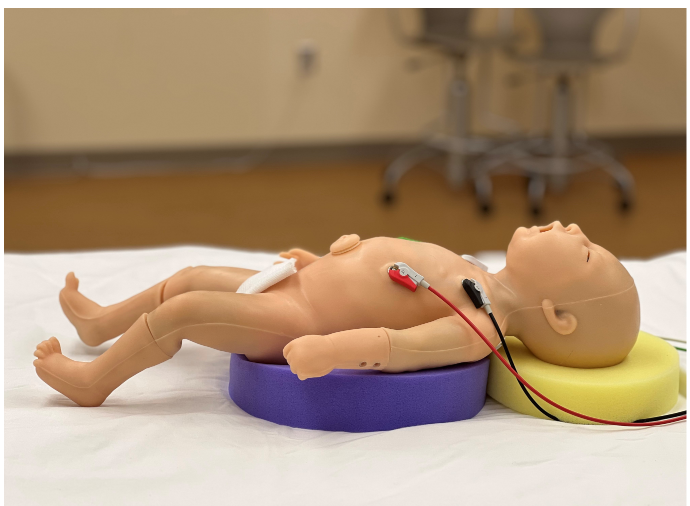

Figure 1. Supine position with utilization of doughnut pillows to keep myelomeningocele lesion free of compression with supporting foam for occiput. *Photo Credit: Elizabeth M. Putnam, MBBS FRCA, Assistant Professor, Anesthesiology, Director of Simulation for Anesthesiology, University of Michigan*

  - Airway
    - Patients with associated cranial abnormalities such as nasal encephalocele may be difficult to mask ventilate.
    - Endotracheal intubation is required. Video laryngoscopy is recommended particularly for intubation in the lateral position which is more challenging.
  - Access and Monitors
    - Standard ASA monitors.
    - Two peripheral intravenous points of access should be present and functioning for adequate resuscitation.

    - Arterial access may be indicated based on patient comorbidities and the need for close blood pressure or lab monitoring. In some instances, central intravenous access should be considered for patients with difficult intravenous access or large lesions which may result in increased blood and 3rd space losses when skin grafting is required.
    - Patients are at high risk for hypothermia based on their size which can be further exacerbated by large defects. Thus, vigilance should be maintained in temperature monitoring. The operating room temperature should be closely regulated. Additionally, use of radiant warming and forced-air heating pads is recommended.
    - Latex-free urinary catheters are recommended for monitoring urine output.
  - Drugs/Infusions
    - A combination of IV (midazolam and/or ketamine boluses) and volatile anesthetic can provide adequate depth of anesthesia. Hemodynamics of these very young patients are sensitive to volatile anesthetic; therefore, it should be titrated carefully.
    - Opioids: fentanyl boluses or a remifentanil infusion may be utilized in small doses as sensation at the level of the lesion is diminished. A long-acting narcotic such as morphine is not recommended due to the risk of postoperative apnea and neonatal organ immaturity.
    - Paralysis: Discuss the use of NMB with the surgeon. Usually, these agents are not contraindicated.
    - Prophylactic antibiotics can be administered per institutional policy or surgical preference.
    - IVF: There may be large insensible losses related to the size of the lesion. Isotonic crystalloid solutions are optimal for resuscitation. Dextrose-containing fluids should be continued to maintain normoglycemia. Albumin may be indicated in larger resuscitations.
  - Blood Availability (if indicated)
    - Transfusion may be required in cases of large lesions.
  - NICU Bed Availability
    - Recommended for close respiratory and neurologic monitoring.
    - Patients are at risk for respiratory compromise most commonly due to neonatal age (particularly if there is prematurity) and possible abnormal brain stem responses to hypoxia and/or hypercarbia. In cases of large defects, increased fluid resuscitation and high tension on the abdomen from skin closure may also affect pulmonary compliance.
    - Patients require close neurological monitoring for signs of persistent or new deficits and signs of increased ICP which may require shunting.

Intraoperative Considerations:

- General
  - General anesthesia is performed to facilitate primary closure of the neural tube defect.
  - General neonatal considerations should be utilized: use appropriately sized equipment, maintain normothermia, and anticipate organ immaturity.
- Induction
  - IV or inhalational induction can be performed. Intubation with video laryngoscopy should be considered. The lesion should be kept free from compression. Utilize a doughnut pillow around the lesion when supine.

- Positioning
    - Close attention to keeping the lesion free of compression is required. Following induction and intubation patient should be placed in the prone position. All pressure points should be padded and free of compression. Prone pillows or padding used for the face and other areas of the body must be adjusted to avoid compression of the eyes and abdomen. The nose and forehead should be elevated enough to avoid contact with the bed surface.
- Maintenance
    - Usually completed with volatile agents, however, there is no contraindication to total intravenous anesthetic.
- Hemodynamic/Physiologic goals
    - Minimize FiO_2 and peak airway pressures as tolerated.
    - Normotensive BP goals.
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
    - Typically, neuromonitoring is not utilized and muscle relaxation provides optimal conditions for airway and surgical management.
- Emergence/Disposition
    - Emergence: awake extubation in the supine position for a neurological exam at the end of the procedure. Large resuscitations or patient instability may require the patient to remain intubated.
    - Direct transport to NICU is most common.
- Post-op Care
    - Close monitoring during transport and arrival to ICU for respiratory compromise due to neonatal physiology and for signs of increased ICP.

Case-Specific Complications/Pitfalls

- Respiratory Compromise should be treated with supportive management which may include invasive airway management
- Signs and symptoms of increased ICP require immediate neurosurgical evaluation and consideration of ventricular shunting.

References

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