APPENDICOVESICOSTOMY (MITROFANOFF) MALONE ANTEGRADE CONTINENCE ENEMA (MACE)

Surgical Specialty:	Urology
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Background:

- General Considerations
  - Both procedures use the appendix to create a catheterizable channel between the skin and bladder (Mitronanoff) or bowel (MACE).[1]
    - Improves the quality of life of the patient by establishing bladder and bowel continence via catheterizeable channels.
    - If the appendix is not available, due to a prior appendectomy, the surgeon may use a piece of the gastrointestinal tract (usually the ileum) to create the channel. [6]
    - The skin opening is positioned in the abdominal wall or in the umbilicus
  - Each procedure can be performed alone or together in children with neurogenic bladder (Mitrofanoff) and neurogenic constipation (MACE).
  - Indications for surgery are patients with spinal dysraphism (i.e., myelomeningocele) or an anorectal malformation who fail conservative management in treating their incontinence or to perform antegrade continence enemas in case of constipation/ recurrent fecal impaction.
  - Often performed as an open procedure, although robotic and laparoscopic techniques are described. [6]
  - Perioperative pain control can be challenging due to the presence of abnormal spinal anatomy precluding neuraxial regional techniques. Additionally, these patients often have chronic constipation therefore minimizing opioid pain medication is desirable.
  - Enhanced recovery after surgery (ERAS) protocols are commonly utilized for these major reconstructive surgeries.[2,3]
  - Patient/parent education sessions are organized a few weeks after the procedure when the wound-healing process is complete. In the meantime, the channel is often catheterized with a Foley-like device to avoid closure. Thereafter, the stoma is equipped with a button device.
- Patient Considerations
  - Spinal dysraphism
    - Abnormal anatomy precluding epidural placement.
      - Consider alternative techniques, such as erector spinae plane catheter or abdominal wall block (e.g. TAP).

        - Decreased ambulation, possibly wheelchair-bound.
            - Succinylcholine is relatively contraindicated.
        - Sensation below the level of the spinal lesion may be reduced.
            - Document dermatomal level of sensation in physical exam prior to performing any regional techniques.
    - Chronic renal insufficiency as a result of chronically increased bladder pressures.
    - Chronic bacterial contamination of urine due to urinary stasis in the bladder
    - Patients with myelomeningocele often have Chiari malformation and sequelae associated with chronic hydrocephalus (VP shunt, h/o surgery).
    - A history of previous appendectomy may alter the surgical plan.
    - Associated Comorbidities/Syndromes
        - Co-existing genetic syndromes

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Conduct a thorough pre-anesthetic evaluation
        - Neurology recommendations, seizure history, review prior anesthetic records,
        - Renal/abdominal ultrasound, CBC, chemistry, urine culture and sensitivity
        - Discuss antibiotic and DVT prophylaxis plan
    - Discussions to have with surgeon/family
        - Code Status
- Specific or Unique Room Set-up Requirements
    - Airway
        - Endotracheal tube
    - Drugs/Infusions
        - If following an ERAS protocol patients may receive a carbohydrate drink 2 hours prior to surgery as well as acetaminophen, pregabalin vs gabapentin, and often a scopolamine patch
        - Regional anesthetic as indicated
            - Allows for decreased use of intraoperative opioids, improved hemodynamics, and potentially reduced anesthetic requirements resulting in improved hemodynamics
            - Low-thoracic epidural (usually around T10 level) may be considered in cases with normal spinal cord anatomy
            - Bilateral erector spinae plane catheters (T10) are gaining popularity as an alternative to epidural in patients with spina bifida anatomy.[4]
            - An abdominal wall block can also be considered (e.g. TAP)
            - Continuous infusion of local anesthetic in the wound has also been shown to reduce postop opioid consumption.[5]
        - Consider ketamine or dexmedetomidine infusions
    - Intravenous fluids can be run via an infusion pump at a standard maintenance rate to minimize excessive fluid administration unless otherwise requested by the surgical team
        - Acetaminophen and ketorolac (if renal function is normal) administration is typically given towards the conclusion of surgery unless contraindicated
        - Dexamethasone and ondansetron for PONV prophylaxis

        - A benzodiazepine, such as diazepam, can be helpful for muscle spasms in the PACU
    - Monitors
        - Standard ASA monitors
        - Arterial-line can be considered on a case-by-case basis but often is not required unless patient-specific conditions warrant placement
        - Two peripheral intravenous lines are desirable
    - Blood Availability (if indicated)
        - An up-to-date type and cross should be available but transfusion is rarely required intraoperatively
    - PICU Bed Availability (if indicated)
        - Usually not required
    - Other Indicated Resources (ex. ECMO standby)
        - Unlikely to be needed

Intraoperative Considerations:

- General
    - Anesthetic and Physiologic Implications of the Surgical Procedure
        - The goal is to provide a smooth intraoperative course while maintaining hemodynamics, preserving gut motility, minimizing opioid and fluid administration, and utilizing a multimodal pain control strategy that optimizes that patient for recovery.[2,3]
- Induction
    - Inhalation vs. intravenous induction as indicated.
    - According to local protocol and regulation, methadone may be given on induction for pain control throughout the case and into the postop period.
    - Ketamine vs. dexmedetomidine bolus followed by infusion should be considered.
    - Appropriate antibiotic prophylaxis
- Positioning
    - Supine.
    - Careful padding of all pressure points.
        - Turn the head every hour to minimize pressure injury risk.
- Maintenance
    - Sevoflurane supplemented with ketamine vs. dexmedetomidine infusion.
    - Opioid only as needed with the goal to use as little as required.
- Hemodynamic/Physiologic goals
    - Maintain normothermia.
    - Maintain euvolemia.
        - The use of a fluid pump can minimize excessive fluid administration.
    - Minimize opioids
        - May exacerbate pre-existing neurogenic bowel dysmotility.
    - Avoid nasogastric tubes and surgical drains as these have not been shown to improve outcomes
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
    - Lengthy procedure, requires padding of pressure points and periodic repositioning during the case.
    - Muscle relaxation is helpful for surgical dissection.
    - Blood loss is minimal to moderate, transfusion is often not required.

    - Creation of an appendicovesicostomy, intraoperative urine output is not able to be accurately monitored due to the bladder being open.

- Emergence/Disposition
    - Given the long nature of the procedure, there is a risk of a prolonged emergence therefore careful planning and timing of discontinuing infusions and inhalation agents is important.
- Post-op Care
    - Patients recover in PACU and then to a surgical floor.
    - Pain control can be challenging if unable to employ regional and other ERAS techniques.
    - Early postoperative physical therapy is desired.
    - Thromboprophylaxis in adolescents with limited mobility

Case-Specific Complications/Pitfalls

- Regional catheter unable to be placed due to anatomy
    - Will require rescue pain medications such as an opioid PCA
- Pressure injury
    - Ulnar nerve injury, hair loss, heel sore
- Deep vein thrombosis
    - Prophylaxis as indicated
- Postoperative VP shunt dysfunction
    - High level of suspicion in case of headache, vomiting, decreased consciousness, bradycardia

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Created: 8/10/22; Last revised: 9/5/23