POSTERIOR SAGITTAL ANORECTOPLASTY (PSARP)

Surgical Specialty:	Colorectal Surgery
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Background:

- General Considerations
  - Anorectal malformations (ARM) can range from simple membranous obstructions, with or without urethral involvement, to stenosis, agenesis, or complex cloacal deformities. Urgent surgery, either lower ARM repair or colostomy, must be performed for any infant unable to pass meconium due to ARM.
  - Simple malformations can be treated with a single surgery in the neonatal period, with staged operations for more complicated lesions. The three-stage operation consists of a colostomy during the neonatal period, ARM repair around 3-6 months (PSARP), followed by colostomy closure.[2]
  - Posterior sagittal anorectoplasty (PSARP) is a procedure to correct ARMs, creating an anus within the sphincter muscle and separating the digestive tract from all urogenital structures. It usually involves a midline sagittal incision from the sacrum to the anal dimple with dissection of muscle structures behind the rectum and potential perineal work depending on the complexity of the ARM.[1]
  - Rectovestibular fistula is the most common defect in females, whereas rectourethral fistula is the most common defect in males.[3]
  - A pediatric surgeon typically performs PSARP, but involvement from urology and gynecology may be necessary depending on the parts of the urogenital tract affected by the malformation.
- Patient Considerations
  - Patient presentation can vary based on the severity of ARM and the presence of associated comorbidities such as chromosomal anomalies (trisomy 13, 18, or 21) or other syndromes associated with ARM.
  - May be part of VATER syndrome/VACTERL association. All patients with confirmed ARM should have undergone evaluation for VACTERL (vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb) abnormalities.[2,3]
  - Approximately 50% of patients with ARM have additional congenital anomalies.
    - Sacral agenesis or anomalies of the spinal cord, such as tethered cord or lipoma, may be present.
    - Approximately 33% of patients have cardiac anomalies, with atrial septal defects, patent ductus arteriosus, tetralogy of Fallot, and ventricular septal defects being the most common cardiac lesions.[2,3]

        - Limb defects are common in patients with ARM – polydactyly, syndactyly, hypoplastic thumbs, and radial aplasia.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Detailed history and physical with careful review of pertinent imaging for additional congenital anomalies.
        - Echocardiography to assess associated cardiac defects (if not performed as part of the VACTERL workup perinatally).
        - Assess limb abnormalities in the context of ease of vascular access.
        - Review sacral imaging if neuraxial analgesia is planned.
    - Discussions to have with the surgeon/family
        - Surgeon – preferred patient positioning during case and any planned changes to positioning intraoperatively (prone to supine/lithotomy). Also, ascertain if the lower extremities will be prepped in the surgical field.
- Specific or Unique Room Set-Up Requirements
    - Airway
        - Endotracheal intubation is generally recommended due to prone positioning, procedure length, and intraoperative position changes.
            - Consider additional steps to secure the endotracheal tube as needed (Tegaderm reinforcement, liquid adhesive, nasotracheal intubation, etc).
        - If associated with a repaired tracheo-esophageal fistula(TEF), careful placement of the endotracheal tube (ETT) is required, avoiding insertion into the previous fistula track.
    - Drugs/Infusions
        - Surgeons may use muscle stimulation to identify key muscle groups during dissection.[3,4] Nondepolarizing neuromuscular blockers (NMB) can abolish the ability to identify muscle groups with a stimulator, so qualitative monitoring of NMB is ideal during this portion.[5]
        - Several options exist for induction and maintenance of anesthesia and may be used at the discretion of the clinician.
            - Sevoflurane
            - Propofol
                - Infusion (100-300 mcg/kg/min)
            - Dexmedetomidine
                - Bolus (0.5-1 mcg/kg) or infusion (0.2-1 mcg/kg)
            - Ketamine
                - Bolus (0.5-1 mg/kg) or infusion (0.1-0.5 mg/kg/min)
            - Remifentanil
                - Infusion (0.05-2 mcg/kg/min)
        - Generally, this procedure is done within the first year of life, so postoperative nausea/vomiting prophylaxis is not indicated.
        - Regional/neuraxial analgesia may be considered provided spinal anatomy is reassuring.
            - Single-shot caudal with or without re-dose at procedure end and epidural placement are the most reported techniques.[6]

        - An epidural catheter placed at the beginning of the case may be challenging to secure as this area will be prepped into the surgical field, and there is a risk of dislodging the catheter with intraoperative position changes. Placement at the procedure's end may be preferable.
    - Monitors
        - Standard American Society of Anesthesiologists monitors: pulse-oximetry, electrocardiogram, non-invasive blood pressure, capnography, and temperature monitoring.
            - Reliable monitoring of core temperature (nasopharyngeal/esophageal) is essential, given the potential for the lower half of the body to be exposed in the surgical field. Consider starting with a warmed room and/or using radiant warming techniques, especially during surgical skin antisepsis application, as well as a forced-air convective warmer (such as a Bair Hugger) once surgical draping is complete.
        - Invasive blood pressure monitoring is indicated, depending on the presence of associated comorbidities or congenital anomalies.
            - Consider the use of pre- and post-ductal pulse oximetry in patients with congenital cardiac abnormalities.
    - Intravenous Access
        - PSARP is often associated with a 2-4 day hospital admission at a minimum,[3] so consider the use of ultrasound for intravenous (IV) placement given the likely need for extended IV access.
        - IV access is often limited to the upper extremities
        - For children likely requiring long-term nutrition postoperatively, consider placement of a peripherally inserted central catheter during the anesthetic.
    - Blood Availability
        - Blood loss is generally expected to be minimal.[3]
    - PICU Bed Availability
        - Not indicated unless the presence of extensive co-morbidities or congenital anomalies warrants a higher level of care.[3]

Intraoperative Considerations:

- General
    - The surgical team may request to turn the patient from prone to supine.
        - A secure ETT and high suspicion for mainstem or dislodgement are essential.
    - The total operating time of the procedure is estimated to be 2-3 hours; however, this is dependent on the complexity of malformations and associated comorbidities.[1-3]
    - A Foley catheter is usually placed after induction.
- Induction
    - Inhaled and/or intravenous agents may be used to induce anesthesia
    - Consider a rapid sequence induction if there is a concern for bowel obstruction.
    - A small amount of NMB may be used to facilitate airway management/intubation. This may be reversed once IV/arterial access and surgical positioning are achieved.
        - Ideally, use rocuronium or vecuronium, given the availability of molecular container NMB reversal agents (sugammadex) that can reliably and quickly reverse even dense neuromuscular blockade.
- Positioning
    - Prone, slightly jack-knifed position with a roll placed under the hips is preferred for initial open dissection.[4]

        - Ensure neutral positioning of the neck with face, abdomen, and thorax free from compression.
    - Mid-procedure position changes between the prone and supine positions are common.
        - Both anterior and posterior lower body areas may be prepped to allow for this.[4]
- Maintenance
    - Balanced anesthetic - Inhaled agents, total intravenous anesthesia (TIVA), short-acting opioid infusions, or opioid bolus as deemed appropriate, with potential analgesia via neuraxial technique.
    - Avoid additional doses of NMBs after intubation.
    - Avoid nitrous oxide due to the possibility of worsening bowel distension.
- Hemodynamic/Physiologic goals
    - Maintenance of normotension, euvolemia, and euthermia.
    - Foley catheter output can provide information about volume status, provided the patient does not have renal or heart failure.
    - Forced-air convective underbody warming and plastic drape for the upper body/head can be very helpful for temperature maintenance.
- Surgical Considerations
    - Surgical teams typically use muscle stimulation to ensure the orientation and appropriateness of pelvic floor muscle dissection so the rectum can be properly implanted into the anal sphincter. Minimal/absent neuromuscular blockade is important during this time.
- Emergence/Disposition
    - Extubation at the case's end is typical. Standard extubation criteria are appropriate.
    - Patients who have undergone PSARP typically require inpatient admission for 2-4 days, usually without ICU requirement.[3]
- Post-op Care
    - Post-operative pain management is usually effectively achieved with non-opioid medications such as acetaminophen or non-steroidal anti-inflammatory drugs.[5]
        - Patients older than 5 years or with a prior/current history of opioid use may have higher analgesic requirements.[5]
    - Patients without a colostomy remain *nil per os* and receive total parenteral nutrition to support healing, while early re-induction of enteral feeding is typical in those with a colostomy.[5]
    - The Foley catheter is usually kept in place for 5-7 days postoperatively.[3]

Case-Specific Complications/Pitfalls

- Frequent position changes from prone to supine and back are a risk for airway displacement and loss of vascular access, as well as potential pressure injury.

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