BLADDER EXSTROPHY

Surgical Specialty:	General Abdominal and Urological Surgery
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Background:

- General Considerations
  - Bladder exstrophy is a congenital malformation and can be potentially life-threatening in the neonatal period, if not managed in a high-volume center of excellence.
  - It is a rare congenital anomaly involving the genitourinary tract, (1:50,000) more common in males compared to females (2.3:1).[1] The posterior bladder wall protrudes outside of the body through a defect in the anterior abdominal wall and the urinary tract is open. It is often associated with a separation of the pubic rami resulting in pelvic diastasis.[2]
  - Bladder exstrophy is caused by the rupture of the cloacal membrane during embryogenesis. This impairs mesenchymal growth in the abdominal wall, resulting in a defect.
  - The risk for recurrence within a family is approximately 1 in 100. Among the offspring of those individuals with exstrophy, occurrence rates are as high as 1 in 70 live births, which is almost 500-fold greater than the incidence in the general population.[3]
  - Diagnosis of bladder exstrophy can be made on prenatal ultrasound and/or at the time of delivery through visual inspection.
  - Bladder exstrophy is surgically repaired in a step-by-step process. Surgery is usually performed soon after delivery. The first step involves closing the open bladder within the abdominal wall with the goal of achieving urinary continence while maintaining adequate renal function and creating a cosmetic repair of the external genitalia.[4] In certain cases, initial repair may be delayed to stabilize the neonate, until adequate weight gain is achieved and the necessary work-up is performed.
  - Pelvic osteotomies might be required to allow midline rotation and approximation at the pubic symphysis when a pelvic gap is present. This step decreases the risk of bladder prolapse and wound dehiscence.[5]
  - Closure of the bladder has long been considered a neonatal emergency to be done within 48 hours after birth. However, some centers now perform a one-stage surgery a few weeks after birth.[6]
- Patient Considerations
  - Bladder exstrophy can be associated with a wide array of clinical manifestations known as the *exstrophy epispadias complex of genitourinary malformations*. It can range from isolated epispadias, bladder exstrophy and epispadias, bifid bladder to a

multisystem defect involving the genitourinary system, gastrointestinal system, and/or musculoskeletal systems.
    - Cloacal exstrophy is an associated condition that includes bladder exstrophy, anorectal malformation, omphalocele, and spinal defects such as tethered cord and spina bifida.[3]
    - Congenital cardiac anomalies may be present, and a thorough history and physical exam should be performed before surgery.[3]
    - Pediatric patients with bladder exstrophy are at an increased risk for developing latex allergy from repeated surgeries/urethral catheterizations.[3] Primary prevention starting with the first surgery is mandatory.

Anesthetic Planning:

- Pre-Anesthetic Evaluation/Consideration:
    - Physical exam: To rule out spinal cord/ vertebral anomalies such as dysraphisms and tethered cords.
    - Imaging: Ultrasound and MRI may be required to rule out spinal dysraphisms, to facilitate safe placement of thoracic epidural and/or caudal catheter. Possible CXR, echo for cardiac evaluation.
    - Labs: Based on physical exam and history: Complete blood count, blood typing and cross-matching, renal function test, and electrolyte levels are usually needed
    - Discussions should be had with the family regarding the length of surgery, positioning, complications, possible procedures such as posterior iliac osteotomies, blood products, and PICU/NICU recovery.
    - Discussion with the surgical team regarding plans for vascular access, postoperative disposition, postoperative extubation, and regional analgesia is also necessary.
- Specific or Unique Room Set-up Requirements
    - Airway: Cuffed Endotracheal tube, appropriately sized laryngoscopes, video laryngoscopes
    - Medications: Rocuronium, Propofol, Ketamine, and Fentanyl are some of the commonly used anesthetic medications.
    - Monitors: Standard ASA monitoring +/- CV line, arterial line.
    - Blood Availability (if indicated based on labs/discussion with the surgical team)
    - PICU/NICU Bed Availability after the procedure
    - Epidural/Caudal catheter for pain management

Intraoperative Considerations:

- Monitors:
    - Standard ASA monitoring +/- Arterial line. Arterial line is useful since major hemodynamic shifts are anticipated with these procedures and some of these infants have co-existing cardiac defects.
- Vascular access:
    - 2 large bore peripheral IVs, preferably in the upper extremity should be obtained for volume resuscitation. PICC/Central Line could be considered for long-term post-operative access, as they might require parenteral nutrition postoperatively
- Anesthetic induction:
    - Inhalational (sevoflurane) vs. Intravenous induction with propofol 1-2mg/kg and rocuronium 0.6 mg/kg

- Airway:
  - Endotracheal intubation, using a straight blade direct laryngoscope/video laryngoscope
- Positioning and other considerations:
  - The posterior iliac osteotomies are performed in the prone or supine position. Prone positioning implies a position change during surgery with its own complications (extubation, dislodged IV line, arterial line). The osteotomies are the most hemorrhagic part of the surgery. In the case of cloacal exstrophy, more than one positional changes may be necessary.
  - Avoidance of latex in the OR is mandatory.
  - Surgery typically lasts for several hours and it is important to maintain normothermia, by optimizing OR temperature and using a forced-air warming blanket device and intravenous, and irrigation fluid warming devices.
- Anesthetic Maintenance:
  - Sevoflurane/isoflurane/rocuronium/propofol/Ketamine; Caudal/thoracic epidural catheter
- Prophylactic antibiotics should be administered in consultation with the surgeon, as per institutional policy
- Anesthetic adjuvants:
  - Ondansetron and dexamethasone might not be necessary, as the risk of PONV is minimal in infants
- Intraoperative pain management:
  - IV Opioids: fentanyl/remifentanil infusion
  - Ketamine infusion 0.3-1 mg/kg/hr
  - In order to not interfere with the surgical field in case of posterior iliac osteotomies, one may consider performing a single shot caudal block at induction (e.g. ropivacaine 0.2% 1ml/kg + 1 mcg/kg clonidine or dexmedetomidine) and placing a lumbar epidural catheter at the end of surgery.
    - The epidural can be tunneled, as it might be required to stay in place for more than a week to facilitate early extubation in the postoperative period.
  - Continuous epidural local anesthetic infusion (Ropivacaine 0.4-0.5 mg/kg/hr or chloroprocaine 8-10 mg/kg/hr)
  - NSAIDs: Given the possibility of co-existing renal anomalies, intraoperative blood loss, and inability to monitor urine output, NSAIDs are typically avoided, especially in neonates
- Postoperative management: Many of these neonates require ICU level of care for post-operative recovery and stay intubated and sedated to facilitate wound healing and pain control. Early extubation is achievable in 48 hours with a functioning epidural catheter. A multimodal postoperative analgesic regimen could include the following medications:
  - Acetaminophen 15mg/kg/6H
  - Regional caudal catheter LA infusion (Ropivacaine 0.4-0.5 mg/kg/hr or chloroprocaine 8-10 mg/kg/hr)
  - Methocarbamol for relief of muscle spasms
  - Diazepam for relief of bladder/skeletal muscle spasms
  - Oxybutinine for relief of bladder spasms
  - PRN opioids

Case-Specific Complications/Pitfalls

- Major blood loss should be anticipated,
    - Blood and products, along with warming devices should be readily available. Serial ABGs help with transfusion goals, as well as maintaining acid-base levels and calcium correction with blood product administration in infants.
- Allowable blood loss in neonates is very small. Given the possibility of major fluid shifts, early PRBC administration in anticipation of surgical steps like pelvic osteotomy is essential.
- PRBC administration can cause rapid and substantial hypocalcemia that can impact cardiac function. Blood gas and electrolyte monitoring and calcium correction should be done when necessary.
- Since it is hard to monitor intraoperative urine output, other indicators like ABG, lactate, blood pressure, and pulse pressure variation can guide fluid management.
- Major blood loss and the requirement of vasopressor support might warrant ICU care and in some cases, postoperative mechanical ventilation.
- Wound dehiscence, surgical site infection, and urinary infection can occur a few days after surgery.

Post-op pain management

- Depending on the extent of the surgical procedure, the patient may be placed in vertical traction for stabilization and closure of the pelvis up to 4 weeks post-surgery.
    - Continuous epidural analgesia, along with the combination of opioids can provide pain relief post-surgery while the pelvis is braced.
    - A multimodal approach to analgesia has been shown to minimize opioid-related adverse effects.[5]
        - Other adjuvants include valium and methocarbamol for skeletal muscle spasms.
        - Oxybutinine is also used by urology to control bladder spasms.
- In patients with contraindications to epidural catheter placement, a multi-modal intravenous approach with opioids (nurse-controlled analgesia) and low-dose ketamine infusion may be used for pain control and to assist with sedation in intubated patients.[5]
- The inability to place a regional catheter for any reason might warrant postoperative mechanical ventilation, at least for the first 24-48 hours, to facilitate opioid infusion for pain control.
- Delayed return of bowel function might pose a challenge to oral nutrition and subsequently, introducing an oral analgesic regimen.

References

1. Prenatal diagnosis of anterior abdominal wall defects: Pictorial Essay. https://www.researchgate.net/publication/239743158_Prenatal_diagnosis_of_anterior_abdominal_wall_defects_Pictorial_essay. Accessed August 29, 2022.
2. Gearhart JP, Rink RC, E. MPD. *Pediatric Urology*. Saunders/Elsevier; 2010.
3. Shapiro, E., Lepor, H., & Jeffs, R. D. (1984). The inheritance of the exstrophy-Epispadias Complex. *Journal of Urology*, *132*(2), 308–310. PubMed Link
4. Mourtzinos A, Borer JG. Current management of Bladder Exstrophy. *Current Urology Reports*. 2004;5(2):137-141. doi:10.1007/s11934-004-0027-5 PubMed Link
5. Davis PJ, Cladis FP. Anesthesia for General Abdominal and Urological Surgery. In: *Smith's Anesthesia for Infants and Children*. Elsevier; 2017
6. Hofman A, Haider M, Promm M, Neissner C, Badely G, Rösch WH. Delayed primary closure of bladder exstrophy without osteotomy: 12 year experience in a safe and gentle alternative to neonatal surgery. J Pediatr Surg 2022 ; 57 : 303-308 PubMed Link

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