Chapter

44

Bladder Exstrophy

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A two-month-old, 5.5 kg infant presents for complete primary repair of classic bladder exstrophy. The infant was born to a 35-year-old G2P1 at 40 5/7 weeks of gestation with good prenatal care. Prenatal bladder exstrophy was diagnosed during an anatomical ultrasound at 22 weeks gestational age.

What Is Bladder Exstrophy?

The bladder exstrophy-epispadias complex consists of rare congenital defects involving the genitourinary and gastrointestinal systems, abdominal wall muscles, pelvic structures, and sometimes the spine and anus. The combined incidence of exstrophy-epispadias complex is approximately 2.15 per 100,000 live births with twice (and up to six times) as many males being affected. Reduction in the expression of p63, a member of the p53 tumor suppressor family, may be a risk factor for development of exstrophy-epispadias complex. During the first weeks of fetal life, exstrophy-epispadias complex results from a derangement in mesodermal layer fusion. The defect in the abdominal wall is widely believed to be the result of overdevelopment of the cloacal membrane which prevents medial migration of the mesenchymal tissue towards midline. The rupture of the cloacal membrane results in herniation of the lower abdominal components to the surface of the abdominal wall.

The severity of the exstrophy-epispadias depends on whether the rupture occurs before or after the separation of the genitourinary and gastrointestinal tracts. The exstrophy-epispadias complex covers a range of severity from epispadias (least severe) to classic bladder exstrophy to cloacal exstrophy (most severe). Classic bladder exstrophy has an incidence of 1 per 10,000 to 50,000 live births and is characterized by an open bladder, exposed dorsal urethra, diastasis of the pubic symphysis, anteriorly displaced anus, inguinal hernia, and genitalia defects. Classic male and female exstrophy is demonstrated by Figure 44.1.

Only 25% of exstrophy-epispadias complex cases are diagnosed prenatally with fetal ultrasound between 15 and 32 weeks of pregnancy, and the remainder are diagnosed during the postnatal examination. Prenatal diagnosis warrants referral to a specialized center with expertise in managing this complex anomaly. Likewise, any infants diagnosed at birth should be sent expeditiously to these centers for experienced evaluation and treatment. Nelson et al. described improved clinical outcomes, lower hospitalization costs, and lower morbidity and mortality rates when patients were treated at specialized hospitals.

What Other Anomalies May Be Associated with Bladder Exstrophy?

While epispadias occurs in isolation, cloacal exstrophy usually occurs with other anomalies. These include malformations of the gastrointestinal, musculoskeletal, and central nervous systems, known as the OEIS complex (omphalocele, exstrophy, imperforate anus, spinal abnormalities). Additionally, patients with classic exstrophy may have skeletal and limb deformities such as clubfoot, congenital hip dislocations, and tibial malformations. Horseshoe kidney and other renal anomalies are more common in children with classic bladder exstrophy (CBE). CBE are rarely associated with omphalocele, imperforate anus, and rectal stenosis. Approximately 7% of CBE patients may have spinal abnormalities such as spina bifida, scoliosis, and hemi-vertebrae.

What Are the Current Treatment Options for Classic Bladder Exstrophy?

Historically, bladder exstrophy patients underwent a cystectomy with high morbidity in the first year of life due to complications from renal failure. Advanced

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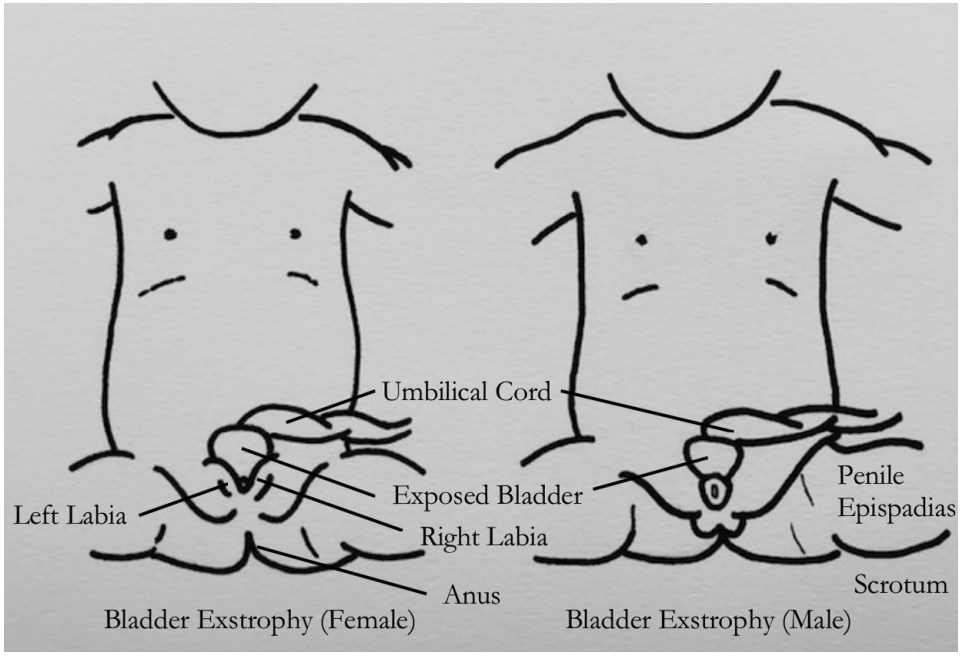

Figure 44.1 Classic features of bladder exstrophy by gender. Illustration by Sandra Perez

surgical techniques have greatly improved survival rates to nearly 100%. The primary reconstructive goal is closure of the bony pelvis, bladder, and abdominal wall defects, followed by epispadias repair; however, long-term goals include preserving renal function, obtaining urinary continence, achieving sexual function, and cosmesis.

The immediate management of bladder exstrophy consists of covering the extruding viscera with sterile silicon gauzes and occlusive dressing to prevent dehydration of the exstrophic plaque and keeping the areas clean with saline washes to reduce risk of infection. There are two main surgical approaches to bladder exstrophy repair: complete primary repair of exstrophy (CPRE) versus modern staged repair of exstrophy (MSRE). As the name implies, the staged repair involves several stages of repair. The first stage is closure of the abdomen and bladder during the newborn period. The second stage is closure of the urethral epispadias. The third stage is a continence procedure and delayed until between ages five to nine, so that the patient achieves an adequate bladder capacity. The complete primary repair combines the first two stages of MSRE. Proponents of CPRE suggest that combining the first two stages stimulates bladder growth and reduces cost by decreasing the number of operations.

PREOPERATIVE EVALUATION

What Preoperative Workup Should Be Considered?

Assessment of all organ systems to ensure absence of associated anomalies is a crucial part of preoperative preparation. Patients with cloacal exstrophy should have cardiac, spinal, and renal ultrasounds to evaluate for associated anomalies. For patients with classic bladder exstrophy, the minimum workup should include a basic metabolic panel to assess baseline renal function before any urinary tract reconstruction. Complete blood count is usually obtained and type/cross-match is recommended if pelvic osteotomies are necessary. For surgical planning, a renal ultrasound screen for other congenital anomalies of the kidney and urinary tract and an abdominal plain X-ray measures the degree of pubic diastasis.

What Preoperative Metabolic Derangements Would Be Expected?

Generally, patients with classic bladder exstrophy have normal kidneys and renal function. For cloacal exstrophy, an inherent short gut syndrome can result

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in significant fluid and electrolyte losses (sodium, chloride, calcium, and magnesium) from the terminal ileum. Patients with short gut syndrome may have significant fat, fat-soluble vitamins, and bile acid malabsorption.

INTRAOPERATIVE MANAGEMENT

What Monitors and Access Would You Require for This Case?

This infant is presenting for complete closure of abdominal and bladder defects, repair of epispadias, and pelvic osteotomies. The complexity and prolonged duration of the procedure with potential for large blood and insensible losses would require large bore peripheral intravenous catheter, arterial catheter, and central venous catheter along with the routine American Society of Anesthesiology (ASA) monitors. Arterial catheterization allows for sampling of electrolytes, blood glucose, arterial blood gases, and hematocrit levels. Due to the open bladder, urine output is not easily measured. Current noninvasive hemodynamic monitoring systems have not been validated for infants. Central venous pressure monitoring allows for continuous assessment of fluid status and postoperatively, the catheter is used for infusion of parenteral nutrition. Arterial and central line placement can be particularly challenging in small infants. Ultrasound guidance is recommended for insertion of arterial and central venous catheters. Special consideration should be given to pressure point padding given the long procedure, especially the sacrum.

What Intraoperative Fluids Would You Deliver? The Infant Was Breastfed Four Hours Prior to Surgery

Determination of intraoperative fluids requires calculation of the maintenance, replacement of ongoing losses, and correction of any specific electrolyte abnormalities. Maintenance is calculated using the "4-2-1 rule" which was first developed by Holliday and Segar in 1957. The formula is derived by calculating water maintenance based on energy expenditure. For decades, their recommendations for the maintenance fluid of choice has been the use of dextrose 5% with 0.2% normal saline. For this patient under 10 kg, the calculated hourly rate equals 22 mL/h (4 mL/kg $\times$ 5.5 kg). Large abdominal surgery with exposed bladder or peritoneum requires 10–50 mL/kg/h of nonglucose-containing isotonic crystalloid to compensate for the insensible losses. Either normal saline or lactated Ringer's solution is routinely used for replacement of volume deficits or blood losses.

What Is Your Plan for Perioperative Pain Management?

Regional anesthesia techniques, especially epidural catheter placement, combined with narcotic and non-narcotic analgesics and sedatives are often utilized. Oftentimes, these infants are managed in the pediatric intensive care unit for the first few postoperative days due to respiratory depressive effects of analgesics and sedatives. Children with continuous epidural analgesia required six- to ten-fold lower doses of intravenous morphine intra- and postoperatively than those without an epidural. Non-narcotic analgesics such as acetaminophen and ketorolac may be helpful alternatives to avoid respiratory depression. Additionally, opioids can reduce GI motility and delay oral feeding. Although ketorolac has been used safely in infants of this age, renal function and postoperative bleeding should be monitored. Despite a well-positioned epidural catheter, additional analgesia or sedation with opioids, diphenhydramine, and benzodiazepines may be required for optimal pain management.

POSTOPERATIVE MANAGEMENT

What Are Some Unique Considerations for Postoperative Pain Management?

The use of pelvic osteotomies during primary closure improves the success of the closure by reducing the tension on the approximation of the pelvis symphysis and abdominal wall and results in deeper placement of the bladder into the pelvis. Fixator pins and external fixation devices are left postoperatively for four to six weeks and the patient is kept immobilized during that time period using modified Buck's traction. Plain film X-rays are obtained to check the degree of pubic diastasis seven to ten days after surgery. If pubic diastasis is significant, the external pins are adjusted gradually to reapproximate the pubic symphysis. A spica cast has been used for pelvic and lower extremity immobilization, but it is associated with

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lower primary closure success rates and higher skin breakdown. The mean length of stay among bladder exstrophy patients is approximately 30 days.

Effective analgesia is critical to the success of this procedure; however young infants are more vulnerable to the respiratory and hemodynamic effects of opioids and sedative drugs. Epidural anesthesia has multiple benefits as alluded to earlier. Risks of epidural analgesia in very young infants include local anesthetic toxicity, and bacterial colonization of the catheters. Short-term non-tunneled catheters have been demonstrated to be colonized with *Staphylococcus epidermidis* at a rate of 9–11% in the lumbar region versus 29% in the caudal region. The duration of the indwelling epidural catheter can be extended by tunneling the catheter subcutaneously, thereby reducing the risks of bacterial colonization and accidental dislodgement. Kost-Byerly and colleagues reported a case series where tunneled epidural catheters were maintained for up to 30 days in infants undergoing bladder exstrophy repair without increased risk. The lumbar or caudal epidural catheter can be easily tunneled using an 18-gauge angiocatheter or Tuohy needle to the flank of the infant. The entry point of the catheter is covered with steri-strips or topical skin adhesive device. It is recommended to examine the site twice a day to ensure no localized infection or fecal contamination.

What Local Anesthetic Dose and Rate Would You Use for the Epidural?

Bupivacaine and ropivacaine 0.1% to 0.2% are the most commonly used local anesthetics for epidural analgesia in pediatrics. The steady state volume of distribution for amides is greater in children, which prolongs the elimination half-life, and thus, the risk of drug accumulation with repeated doses and/or continuous infusion is increased in infants and children. Bupivacaine must be used with caution as rising serum levels between 1 and 24 hours have been demonstrated with infusions of 0.2 mg/kg/h in neonates when infusing up to 48 hours. In pharmacokinetic studies of ropivacaine in infants under 1 year of age, Bosenberg et al. demonstrated ropivacaine plasma concentrations reached steady state within 24 hours for infusion rates of 0.2 mg/kg/h for up to 72 hours with no adverse side effects.

There is scant data on the safety of epidural beyond 72 hours in infants. A safe level of ropivacaine is considered a maximum serum of 4 mcg/mL or an infusion rate less than 0.4 mg/kg/h. Titration by the measurement of ropivacaine blood levels is not possible as these samples require a lengthy analysis. One recent case report has described the postoperative management of an infant with bladder exstrophy using a tunneled lumbar epidural infusing ropivacaine 0.1% at 3 mL/h (0.4 mg/kg/h) for 26 days. The ropivacaine levels for all samples remained below 4 mcg/mL. For this patient, 0.1% ropivacaine at 2.2 mL/h (0.4 mg/kg/h x 5.5 kg) would be an appropriate rate.

Serum lidocaine levels can be easily measured in most hospital's laboratories and used to guide therapy, alerting clinicians of potential toxic levels. Epidural lidocaine infused at a rate 0.8 mg/kg/h through a tunneled caudal epidural was used safely in neonates with bladder exstrophy. The rate for this patient is calculated at 4.4 mL/h (0.8 mg/kg/h x 5.5 kg) of 0.1% lidocaine.

Suggested Reading

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