Exstrophy Bladder or Ectopia Vesicae

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41.1 Introduction

The bladder exstrophy-epispadias-cloacal exstrophy complex is a constellation of ventral wall defects caused by a developmental abnormality 4–5 weeks after conception. Bladder exstrophy exists concomitantly with epispadias due to failure of lateral body wall folds to close in the midline, leaving the urethral meatus on the dorsum of the penis [1].

Bladder exstrophy /ectopia vesicae is a rare congenital malformation of the genitourinary system. The reported incidence is 1:10,000–50,000 live births, 2–3 times more common in male newborns. Basic anomaly is nonclosure of anterior bladder wall and anterior abdominal wall, exposing the bladder mucosa to the outside. This is associated with separation of pelvic bone, malrotation, so that the gap in the lower abdomen and pelvis is very wide. Urethra may not be closed properly with occurrence of epispadias. Management requires early closure of the bladder and abdominal wall along with pelvic osteotomy and approximation of pubic symphysis. Both intraoperative and postoperative management are very challenging both for the anesthesiologist and the surgeon. Postoperative pain management and pelvic and lower limb immobilization are imminent for several weeks until healing occurs. Babies have been operated in the neonatal period, but this is associated with high morbidity. This chapter will discuss the embryological abnormality responsible for the defect, various surgical components, and anesthetic management, and perioperative pain management [2].

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41.2 Embryology

The bladder and ureterovesical junction are formed during 4–6 weeks of gestation and arise from the primitive urogenital sinus following subdivision of the cloaca. The bladder develops through mesenchymal-epithelial interactions between the endoderm of the urogenital sinus and mesodermal mesenchyme.

Various theories have been proposed for the genesis of **bladder exstrophy** or the **exstrophy-epispadias complex (EEC)** including obstruction or failure of mesenchymal migration [3], premature rupture of cloacal membrane [4] (prior to the fusion of the uro-rectal septum and the cloacal membrane), abnormal cell-cell interactions, and alteration in cell death. Another theory is persistence of pubic diastasis and open bony pelvis. The pelvic musculature rests behind the hindgut and urogenital sinus. Pubic diastasis is normal in early gestation, and by 8–10 weeks, the pubic bones rotate, with approximation and closure of the pubic symphysis. In pubic diastasis, because of malrotation of the pelvic bones, as the fetus grows and posterior musculature develops, it exerts anterior pressure, preventing normal rotation and approximation of the pubic bone and symphysis, disrupting closure of the bladder and abdominal walls, consequent exstrophy [5]. Three types of defects (EEC) may occur [6, 7]:

1. Cloacal exstrophy,
2. Classical bladder exstrophy, or
3. Epispadias. **OEIS complex** (Omphalocele, Exstrophy of the cloaca, Imperforate anus, and Spinal defects) is the most severe form of **EEC**. It is extremely rare with an incidence of 1:200,000–400,000 live births, sporadic in occurrence, with strong association with genetic defects, spina bifida, intersex, environmental exposures, twinning, and in vitro fertilization. Babies have specific facial features, developmental delays, and heart, skeletal, genitourinary, and neurological defects [8].

41.3 Pathophysiology

41.3.1 Bone Defect

They have a widened **pubic symphysis**, short pubic rami, retroverted acetabulum, wide sacroiliac joints, and larger sacrum [9]. **Spinal and Neurological** defects are present in nearly all babies including spina bifida occulta, scoliosis, hemivertebrae [10] neural tube defects (NTD), and tethered cord [11], and they add on to the severity of already present urinary and bowel incontinence. Other bony defects include clubfoot, tibial malformations, and congenital hip dislocation [8]. **Pelvic floor** musculature is abnormal, anus and bladder are more anterior [9], and these predispose female patients to uterine prolapse [12]. The **ventral abdominal wall** fascia is absent, umbilicus is more caudal position (at the upper limit), and with a small or large omphalocele depending on the degree of maldevelopment [8].

41.3.2 Anorectal Displacement

Anorectal displacement predisposes the baby to fecal incontinence. GI defects are very frequent in the form of imperforate anus, rectal stenosis, rectal prolapse [13], rudimentary hindgut, malrotation, and short bowel [8, 14].

41.3.3 Genitalia

Genitalia is profoundly abnormal, e.g., dorsal urethral meatus, epispadias, and short penis in males, and short stenotic vagina, bifid clitoris, and divergent labia in females [15]. **Bladder** is open anteriorly, undeveloped, small, and poorly compliant. Ureters enter the bladder at an abnormal angle with vesicoureteral reflux. Horseshoe kidney is very common in EEC [16].

41.4 Diagnosis and Postnatal Care

EEC can be diagnosed during prenatal sonography in the early second trimester and confirmed at 32 weeks of gestation [17]. Sonography findings are absent bladder filling, low set umbilicus, wide pubic rami, anterior anus, small genitalia, and lower abdominal mass [18], genitalia defects, and constant urine seepage. Male babies have epispadias, undescended testis, and/or inguinal hernia. Female babies may have epispadias, bifid clitoris, separated labia, absent vagina, and/or bifid uterus. Prenatal diagnosis allows for parental counselling, prognosis, and treatment approaches.

Postnatal diagnosis is by the evident defect. Babies must undergo renal, cardiac, pulmonary, and neurosurgical evaluation soon after birth. The exposed bladder and bowel segments should be covered with non-adherent dressing, kept clean using saline, and changed frequently. Evaluation under anesthesia (EUA) is done to ascertain the extent of defects and formulate management strategy. These babies should be managed at a tertiary neonatal center [19].

41.5 Surgical Management

Surgical repair of EEC is challenging because of the rarity and complexity of the defect. Basic repair is closure of the bladder, closure of anterior abdominal wall, approximation of the pubic rami, and epispadias and chordae correction. Bladder and abdominal wall closure should be done as early as possible, but pelvic osteotomy should be done only after 72 h age, when pelvis is still malleable.

41.5.1 Surgical Approaches Include

1. **CPRE- Complete Primary Repair of exstrophy**, preferred technique with low costless inflammation and fibrosis, improved bladder growth, and less need for urinary diversion [20– 24], and

2. **MSRE- Modern Staged Repair of Exstrophy** [25] that involves three surgeries: **The first stage** (within 2–3 days of life) includes bladder and abdominal wall closure, reconstruction of belly button, and osteotomy (>72 h age). **The second stage** (at 6–12 months of age) includes epispadias repair in boys, and urethral and labial repair in girls. **The third stage** (at 6–10 years age) for bladder neck reconstruction and bilateral ureteral reimplantation.

41.5.2 Other Surgical Approaches Described are

1. **Erlangen repair -** delayed complete one-stage repair at 8–10 weeks of age, allowing for stabilization and growth of the infant. Continence rates as similar to MSRE [26].
2. **Kelly repair -** an alternative staged repair, in which, instead of pelvic osteotomy, closure is accomplished through soft tissue mobilization from their attachment to the pelvic sidewall. Continence rates are similar to MSRE; however, the lower abdominal wall has an abnormal appearance [27].
3. **Warsaw approach -** is a two-stage approach, with early abdominal wall and bladder closure and lower extremity and pelvic immobilization, followed by additional surgeries at leisure [28].

41.5.3 Postoperatively

Postoperatively, these babies require immobilization of the pelvis and lower limbs for several weeks, especially if pelvic osteotomy has been done. This is extremely painful and continuous pain management for 4–6 weeks is essential. Immobilization can be achieved by:

(a) **External fixator-** Traction is adjusted according to the degree of pelvic diastasis as assessed on pelvic radiology, after 7–10 days of surgery and then at 4 weeks,
(b) **Spica cast application,** instead of painful external fixator, that creates problems with nursing care and feeding, immobilization can be achieved with similar results, or.
(c) **"Mummy wrapping"** the child's legs, though simple and less painful, is less secure and has poor results [29].

70% of these children can live without incontinence and with normal urethral voiding, and with minimal complication rate [30].

41.5.4 Preoperative Care

Preoperative care is directed towards less exposure of bladder and other viscera, and minimizing trauma and risk of infection. A non-adherent film dressing is applied,

kept wet and cleaned with saline, and changed frequently as it becomes soaked with urine.

41.6 Preanesthetic Assessment

Components of PAE are:

1. A thorough assessment of the severity of the EEC defect,
2. Other organ systems, especially cardiac, pulmonary, and renal function assessment,
3. Discussion with the surgeon as to the plan of surgery, Primary or Staged, and.
4. Parental counselling and informed consent.

41.7 Anesthesia Concerns

(a) CPRE approach, though preferred, increases the risk of surgery and anesthesia because of prolonged duration (4–6 h), increased blood loss, hypothermia, and high complication rate (wound dehiscence, bladder prolapse, vesicocutaneous fistula, penile loss [31–33], and urinary incontinence [34, 35].
(b) MSRE approach has the disadvantage that it takes 10 years for full repair, but improves the safety in the neonate.

Preoperative investigations include Hb and complete blood counts, blood urea, serum creatinine and electrolytes, blood group and cross matching, and urinalysis (routine, microscopic examination, culture, and sensitivity).

41.8 Anesthesia Management

General principles of neonatal anesthesia need to be followed. Surgery requires general anesthesia, endotracheal intubation, and controlled ventilation. Two IV lines should be secured. Standard noninvasive monitoring is sufficient and includes ECG, NIBP, SpO_2, $EtCO_2$, and temperature. Central venous catheters are useful and can be considered as they can be utilized postoperatively for fluids, drug, TPN administration, as well as for blood sampling.

Caudal epidural catheter by ultrasound or landmark guided technique can be placed to facilitate intraoperative and postoperative analgesia. Catheter should be tunneled subcutaneously as it must be kept for 4–6 weeks, the entire postoperative period. Local anesthetic with additives (clonidine or dexmedetomidine) is preferred to fentanyl (short acting), and to morphine (respiratory depression). Measures should be taken to prevent hypothermia by use of fluid warmers and forced air blankets.

Care must be taken to prevent reopening of intracardiac shunts and PFC (persistent fetal circulation) and risk of ROP with high FiO_2.

41.9 Postoperative Management

- Pain management for 4–6 weeks,
- Antimicrobial prophylaxis (risk of infection due to stents, drains, pelvic fixator pins, osteotomy, and urine spillage). Other reasons are potential wound dehiscence and infection, pyelonephritis, vesicoureteral reflux, and additional surgical interventions,
- The wound should remain dry and free from tension,
- Pelvic and lower limb immobilization,
- Care to prevent accidental drag and removal of abdominal and bladder drains (suprapubic catheter), urethral catheter, and ureteral stents for the entire duration of immobilization,
- Early feeds or TPN,
- Care in a thermoneutral environment, and
- Adequate hydration and maintenance of volume status.

41.10 Conclusion

Management of bladder exstrophy has significantly changed over the last few decades. Surgery includes bladder and abdominal wall closure, pelvic osteotomy, postoperative traction and immobilization, pain and sedation management, nutritional management, and advanced pediatric nursing care. A team of pediatric urologists, orthopedic surgeons, anesthesiologists, pediatricians, pediatric nursing staff, and other hospital staff working together can provide the child with the best chance for a functional and cosmetic result. Most children can lead a dry life with normal urethral voidance and minimum complications.

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