Bladder Exstrophy

Ashwin V Apte, *Rajat Saran

Department of Surgery, *Department of Orthopedics, People’s College of Medical Sciences and Research Centre, People’s campus, Bhanpur, Bhopal-462037 (M.P.)

Abstract:
Bladder extrophy, more properly, the exstrophy-epispadias complex is a rare congenital anomaly occurring once every 10,000-50,000 live births with a 2.3:1 as male & female ratio. The diagnosis involves a spectrum of anomalies of the lower abdominal wall, bladder, anterior bony pelvis and external genitalia. It occurs due to failure of the abdominal wall to close during fetal development and results in protrusion of the posterior bladder wall through the lower abdominal wall. We report a case of bladder extrophy managed by us.

Key Words: Bladder extrophy, Exstrophy-epispadias complex, cloacal extrophy.

Introduction:
The exstrophy-epispadias complex of genitourinary malformations can be as simple as a glanular epispadias or an overwhelming multisystem defect such as cloacal extrophy.

Bladder extrophy is a rare congenital malformation of the bladder and urethra in which the bladder is turned inside out. Bladder is flattened and exposed to outside the body. Bladder neck fails to form properly and the anus and vagina appears to be displaced anteriorly. Also, there is diastasis of the pubic bones.

The incidence of bladder extrophy has been estimated as being 1 in 10,000 to 1 in 50,000 (Lattimer & Smith, 1966) live births. The male-to-female ratio of bladder extrophy derived from multiple series is 2.3:1 (Shapiro et al, 1984). The risk of recurrence of bladder extrophy in a given family is approximately 1 in 100 (Ives et al, 1980).

Spectrum of anomalies

The typical manifestation of exstrophy-epispadias complex is:

- Bladder everted through a midline lower abdominal wall defect
- Widening of the pubic symphysis
- Epispadias in male (dorsal cleft in the penis, exposing the urethral mucosa)
- The anus and vagina appears to be displaced anteriorly
- The testicles may not be fully descended
- Bifid clitoris in female, with a short “urethral strip” indistinguishable from bladder mucosa.

The spectrum of disease extends from spade penis and epispadias on one hand, to extrophy with cloaca (also known as cloacal extrophy) on the other hand.

Case report:

We report a 7 year old child presented to us for the first time with classical extrophy of the urinary bladder. The child was second of the four siblings, born by full term normal vaginal delivery out of non consanguineous marriage, with no significant antenatal history. There was no history suggesting recurrent urinary tract infections and other noticeable illness in past. The child was of normal built and weight for his age.

Gross examination of the child showed opened bladder plate at the lower part of abdomen with urine dribbling all the time and a typical uriniferous odour. Minute observations of the local area showed that the bladder mucosa is having small nodular pattern and two projections on both sides denoting the two ureteric openings (Fig I). The bladder neck was open and could be noticed well on pulling the splayed penis downwards. There was pubic diastasis, demonstrated by pre-
operative X-ray (Fig.II) and the testis was situated in the superficial inguinal pouch on both sides with less developed scrotal sacs. The anal opening was anteposed with short perineal distance and a normal sphincter muscle tone. Investigations of the patient revealed normal haemogram, urine analysis and renal functions.

Sonography of abdomen showed normal kidney on both the sides; both testes were seen in superficial inguinal pouch.

Surgery was planned and bladder turn-in with bladder neck reconstruction and urethroplasty (Fig.III) was done in a single sitting along with bilateral iliac osteotomies (Fig.IV). Repair after reconstruction was taken up very well (Fig.V) and the pubic bone approximation was demonstrated on post-operative X-ray (Fig.VI). Bilateral ureteric cannulation and bladder

---

Fig. I: Photograph showing bladder mucosa with small nodules. Epispadiac short penis, bilateral undescended testis with poorly developed scrotal sac, absent umbilicus.

Fig. II: X-ray showing wide public diastasis and bony changes in the pelvis.

Fig. III: Bladder turn-in and neck reconstruction done & ureteric catheters in situ.

Fig. IV: Operative picture showing exposure of iliac crest for iliac osteotomy.

Fig. V: Post-operative picture showing well formed penis and umbilicus and adequate skin closure.

Fig. VI: X-ray picture after operation showing approrximation of pubic symphysys with steel wire.
Bladder Exstrophy ———————————————————————————————————— A V Apte, R Saran.

donation was done for two weeks. Post-operative
course of the patient was uneventful and the patient
was on follow-up for six months after surgery. During
follow-up visit it was noticed that the glanuloplasty
sutural disruption occurred. The child was able to hold
urine for about one hour during day time. Post-operative
investigation showed the bladder capacity to be
approximately 50 ml with normal kidneys.

Anatomical Considerations:

Bladder exstrophy, cloacal exstrophy, and
epispradias are variants of the exstrophy-epispadias
complex. The cause of this complex is thought to be
the failure of the cloacal membrane to be reinforced
by ingrowth of mesoderm (Muecke, 1964). The cloacal
membrane is a bilaminar layer situated at the caudal
end of the germinal disc that occupies the abdominal
wall below umblicus. Mesenchymal ingrowth between
the ectodermal and endodermal layers of the cloacal
membrane results in the formation of lower abdominal
muscles and pelvic bones. The cloacal membrane is
subject to premature rupture; depending on the extent
of the infra-umbilical defect and the stage of
development during which the rupture occurs, bladder
exstrophy, cloacal exstrophy, or epispadias results
(Ambrose & O’Brien, 1974).

Exstrophy of the bladder is part of a spectrum
of anomalies involving the urinary tract, the genital tract,
the musculo-skeletal system and sometimes the
intestinal tract. In classical bladder exstrophy, most
anomalies are related to defects of the abdominal wall,
bladder, genitalia, pelvic bones, rectum and anus.

Sponseller et al (1995) found that patients with
classic bladder exstrophy have a mean 12 degree
external rotation of the posterior aspect of the pelvis
on each side, retroversion of the acetabulum and a mean
18 degrees of external rotation of the anterior pelvis
along with 30% shortening of the pubic rami, in addition
to the diastasis of the symphysis pubis.

The triangular defect caused by the premature
rupture of the cloacal membrane is occupied by the
exstrophy bladder and the posterior urethra. The fascial
defect is limited inferiorly by the intra-symphysial band,
which represents the divergent urogenital diaphragm.
This band connects the bladder neck and posterior
urethra to the pubic ramus. The anterior sheath of the
rectus muscle has a fan-like extension behind the
urethra and bladder neck which inserts into the intra-
symphysial band.

The perineum is short and broad and the anus
is situated directly behind the urogenital diaphragm; it
is displaced anteriorly and corresponds to the posterior
limit of the triangular fascial defect. The anal sphincter
mechanism is also anteriorly displaced and it should be
preserved intact in case internal urinary diversion is
required in future management

Rectal prolapse frequently occurs in untreated
exstrophy patients with a widely separated symphysis.
It is usually transient and easily reduced. Silver et al
(1997) found that the anterior corporal length of male
patients with bladder exstrophy was almost 50% shorter
than that of normal controls, the penis appears short
not only because of the diastasis of the pubic symphysis
but also because of marked congenital deficiency of
anterior corporal tissue.

The vagina is shorter than normal, hardly greater
than 6 cm in depth, but of normal caliber. The vaginal
orifice is frequently stenotic and displaced anteriorly,
the clitoris is bifid and the labia, mons pubis and clitoris
are divergent. The uterus enters the vagina superiority
so that the cervix is in the anterior vaginal wall. The
fallopian tubes and ovaries are normal. The frequent
occurrence of indirect inguinal hernias is attributed to a
persistent processus vaginalis, large internal and external
inguinal rings, and lack of obliquity of the inguinal canal.

The urinary tract is usually normal, but
anomalous development may occur. Horseshoe kidney,
pelvic kidney, hypoplastic kidney, solitary kidney and
dysplasia with megaureter are all encountered in these
patients. The ureters have an abnormal course in their
termination. The peritoneal pouch of Douglas between
the bladder and the rectum is enlarged and unusually
deep, forcing the ureter down laterally in its course
across the true pelvis. The distal segment of the ureter
approaches the bladder from a point inferior and lateral
to the orifice and it enters the bladder with little or no
obliquity. Therefore, reflux in the closed exstrophy
bladder occurs in 100% of cases and subsequent
surgery usually is required at the time of bladder neck
reconstruction.

Discussion:

Exstrophy of bladder is a rare condition with
incidence of 1 per 30,000 - 50,000 live births with male
to female ratio ranging from 1.5:1 to 5:1 (Ben-Chain
et al, 1996; Russell et al, 2000; De Bruyn et al, 2001). The
risk of having sibling with bladder exstrophy is 1%
(Silverman & Kuhn, 1993). The condition is intermediate
in severity between epispadias and cloacal exstrophy. The condition is thought to be caused by incomplete development of the infra-umbilical part of the anterior abdominal wall, associated with incomplete development of the anterior wall of the bladder owing to delayed rupture of the cloacal membrane. Persistence of the cloacal membrane prevents mesenchymal ingrowth, causing the abdominal wall to remain lateral and the posterior bladder wall to be exposed to the external surface (Russell et al, 2000; Gearhart & Jeffs, 1998). Trigone of the bladder and ureteric openings are exposed and sometimes there is mild prolapse. The anterior abdominal wall defect involves the entire urethra and bladder neck (Silverman & Kuhn, 1993). The pubic symphysis is always widened with diastasis of rectus abdominis (De Bruyn et al, 2001). Umbilicus is low set and frequently there is omphalocoele (Silverman & Kuhn, 1993; Gearhart & Jeffs, 1998) which is confluent with exstrophic bladder (Ben-Chain et al, 1996). In males the penis is short, stubby, curved upwards and is drawn into the exstrophic area, unilateral or bilateral cryptorchidism may be present and may be associated with inguinal hernia (Russell et al, 2000; Silverman & Kuhn, 1993).

In females, the urethra is short, often buried in the exstrophied bladder. The clitoris tends to be bifid. The labia are also widely separated. The vagina is short and orifice may be stenotic. Uterine prolapse or unicorionate uterus may be present (Russell et al, 2000; Silverman & Kuhn, 1993). Anus is anteriorly placed and may be patulous; this is more commonly seen in girls. Distal ends of ureters are slightly dilated and curve laterally, then medially and slightly upwards in the shape of a hook before entering the bladder (Silverman & Kuhn, 1993). In untreated patients, due to continuous dribbling of urine, there can be mucosal abrasion, infection, squamous metaplasia resulting in acquired vesico ureteric junction (VUJ) obstruction. The detrusor muscle may become fibrotic and scarred. Instances of adenocarcinoma of bladder have been reported in untreated adult patients. (Silverman & Kuhn, 1993).

**Prenatal Diagnosis and Management:**

Diagnosis of exstrophy - epispadias complex can be made antenatally. Antenatal ultrasonography (USG) findings (Hamada et al, 1999; Pinnette et al 1996) included:

- Repeated failure to visualize the bladder.
- Lower anterior abdominal wall mass.
- Low set umbilicus with omphalocoele.
- Abnormal genitalia.
- Increased pelvic diameter.
- Associated renal anomalies, myelomeningocele and limb anomalies which are more common in cloacal exstrophy.

In neonates, exstrophy of bladder is diagnosed on clinical examination; workup includes baseline renal function test before complex reconstruction of the urinary tract. Renal USG is done to rule out renal agenesis, hydronephrosis and ectopic kidney. After bladder reconstruction is done, USG is done to look for upper urinary tract deterioration which may result from increased bladder pressure or repeated infection.

Spinal USG, radiography and MRI is done to exclude myelodysplasias or vertebral anomalies. (Gearhart & Jeffs, 1998).

An early assessment by examination under anaesthesia should be carried out in a center experienced with the condition.

**Surgical Procedure:**

The goals of surgery are to close the bony pelvic ring, close the bladder, posterior urethra and close the anterior abdominal wall defect and reconstruct the genitalia. In the first year of life, the bladder is closed following osteotomy of both iliac bones just lateral to sacro-iliac joint. Later reconstruction of bladder neck and sphincter is done. Complications of closure is the failure to reach adequate capacity and thus, augmentation or reconstruction may be necessary (Russell et al, 2000).

Another option is urinary diversion if continence is poor following bladder reconstruction. This can be done by ureterosigmoid anastomosis or formation of ileal conduit, colonic conduit or continent urinary diversion (Ben-Chain et al, 1996; Russell et al, 2000; De Bruyn et al, 2001). Complications include stricture at the site of anastomosis, increased chances of adenomas and adenocarcinomas at the site of ureterocolic anastomosis and hyperchloraemic acidosis (Russell et al, 2000).

In an effort to decrease costs, decrease the morbidity associated with multiple operative procedures, and possibly affect continence, there has been a recent interest in performing single-staged reconstruction or combining procedures in appropriately selected patients.
Bibliography: