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Bladder Exstrophy - A case report

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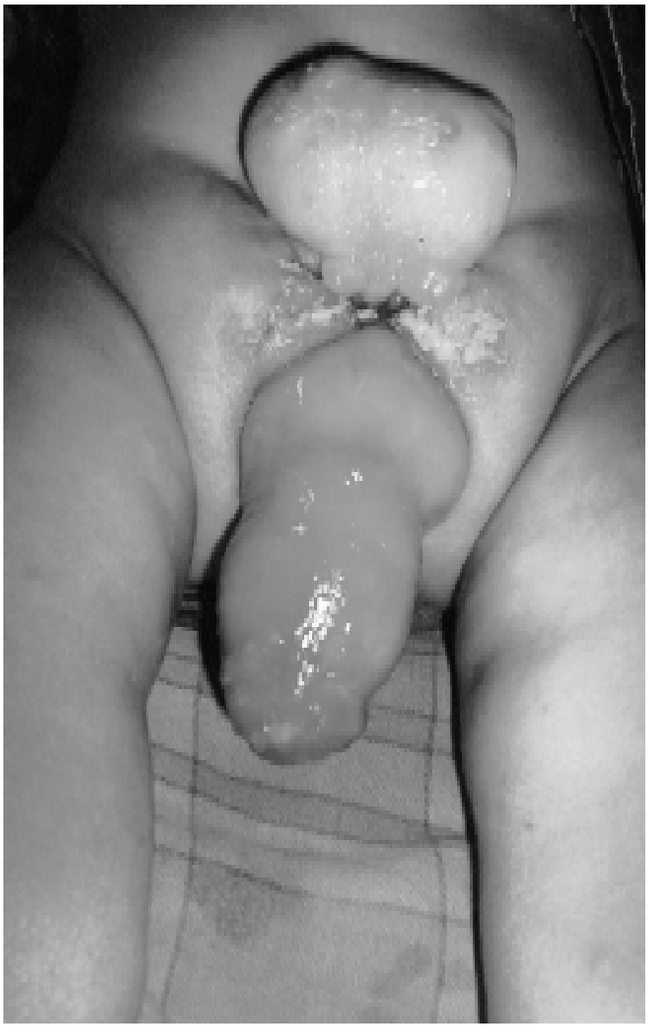
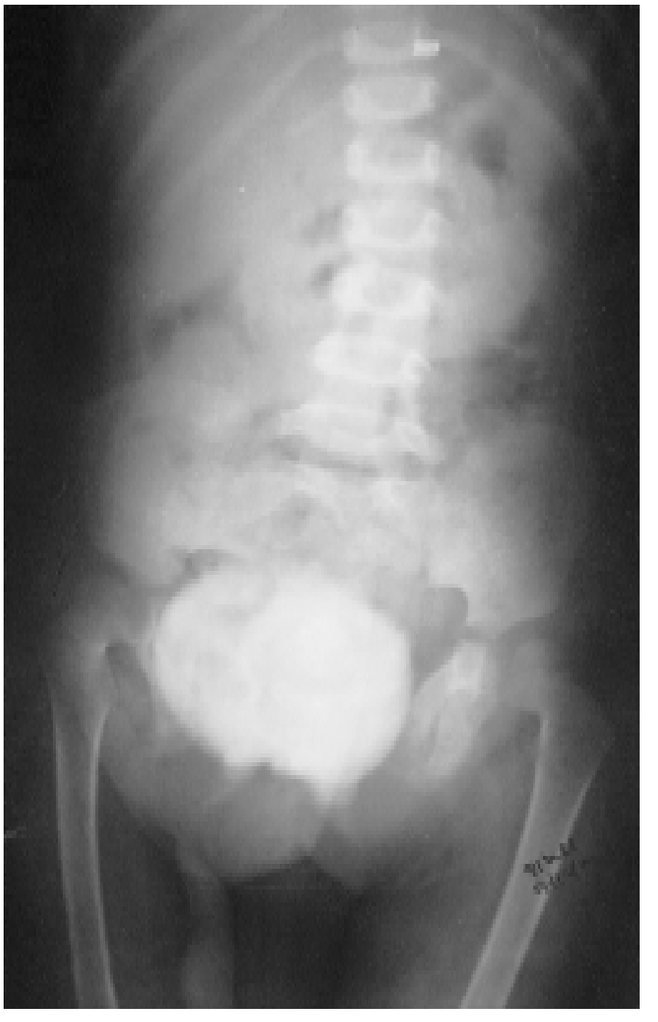
Introduction

Bladder exstrophy is a rare congenital birth defect that is the 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 correctly and the anus and vagina appear anteriorly displaced. Also, there is diastasis of the pubic bones.



A two and half year old female child presented with defect in the lower anterior abdominal wall since birth with protrusion of a globular soft tissue swelling from the defect. Confluent with this swelling, in the inferior aspect, were two openings, from where urine dribbled continuously, suggesting ureteral openings, with exposed posterior wall of the bladder.



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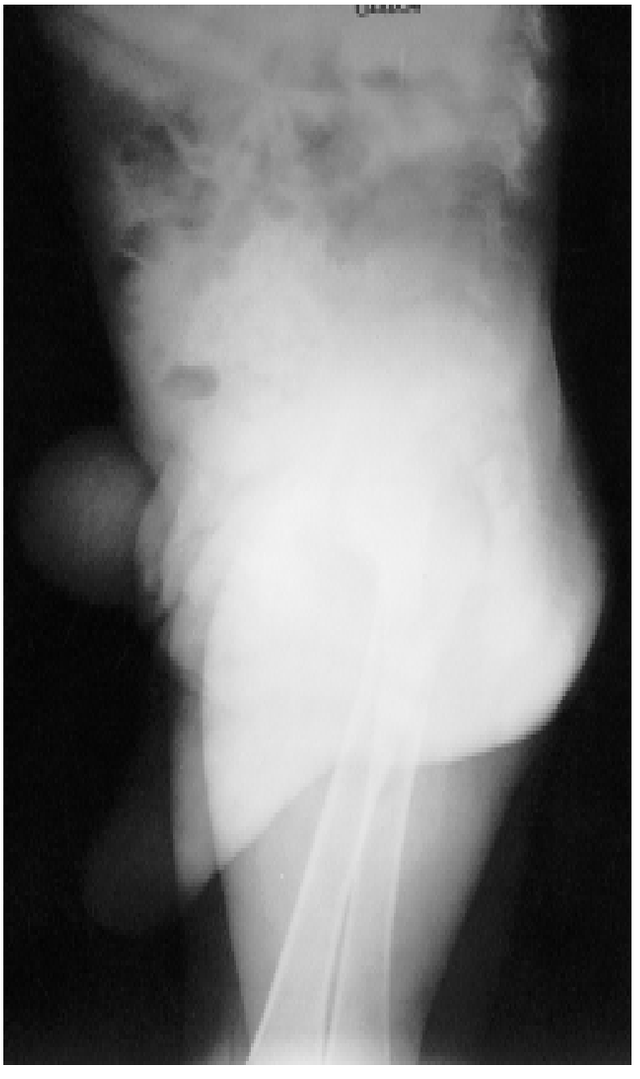
Umbilicus was not seen separately.

Patient had another prolapsing elongated soft tissue inferior to the globular swelling, with an opening through which patient used to defecate, suggesting prolapsed rectum, which was somewhat anteriorly placed than normal individuals. Labia were widely separated and clitoris was bifid [Fig 1].

Plain KUB

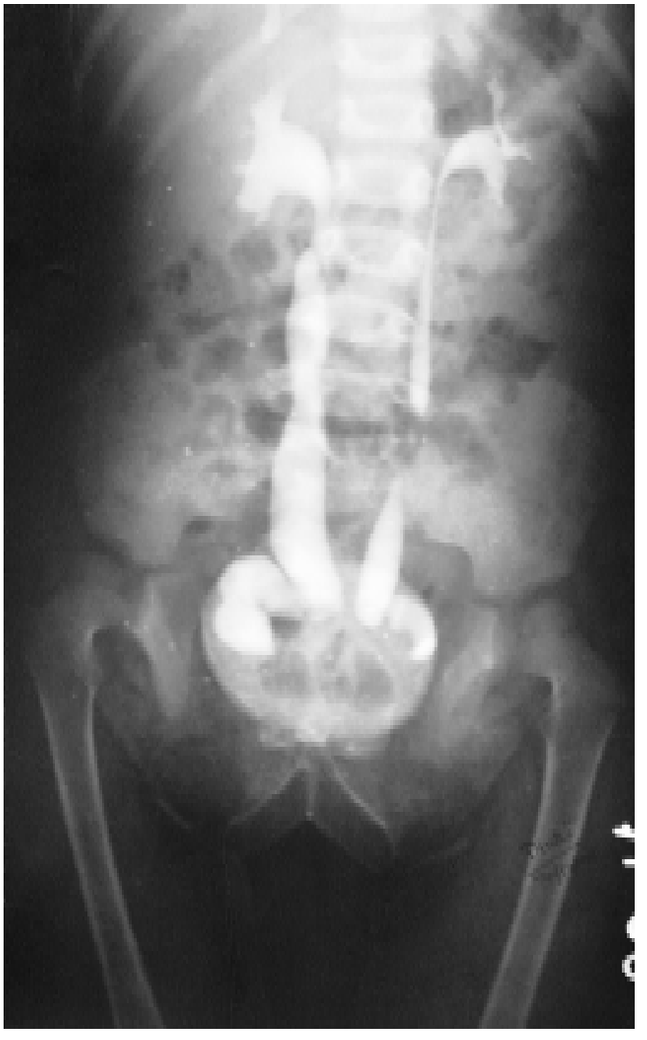
Radiographs were taken in antero-posterior and lateral projections [Fig 2,3] which revealed a globular soft tissue shadow arising from the anterior aspect of lower abdomen and pelvis.



Another elongated soft tissue opacity was seen below it arising from the pelvis and directed caudally.
There was evidence of diastasis of pubic bones.
Coccyx was absent.

Intravenous Urography

IVP [Fig 4] was done by injecting 15 cc of 76% Urografin intravenously.



There was prompt excretion of contrast from both kidneys. There was e/o fullness of right renal pelvis with gross dilatation and tortuosity of both ureters, that of the right side being affected more than left, with abrupt tapering at their distal ends.

Non filling of bladder was noted with leakage of contrast through ectopic ureteral openings s/o partial obstruction at distal ends of both ureters.

Diagnosis was put as exstrophy of bladder with bilateral ectopic ureteral openings with distal partial strictures with back pressure changes in the form of bilateral hydroureter, more on the right side, with rectal prolapse and omphalocoele.

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 [1,2,4]. The risk of having sibling with bladder exstrophy is 1% [7]. 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 medial mesenchymal ingrowth, causing the abdominal wall to remain lateral and the posterior bladder wall to be exposed to the external surface [2, 7]. Trigone of the bladder and ureteral openings are exposed and sometimes there is mild prolapse. The anterior abdominal wall defect involves the entire urethra and bladder neck [4].The pubic symphysis is always widened [3] with diastasis of rectus abdominis. Umbilicus is low set [4, 7] . Frequently there is omphalocoele [4,7] which is confluent with exstrophic bladder. [4]

In males the penis is short, stubby, curved upwards and is drawn into the exstrophic area [2,4]. Unilateral or bilateral cryptorchidism may be present [2,4]. Inguinal hernia may be associated [2,4].

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 unicornuate uterus may be present [2, 4].

Anus is anteriorly placed and may be patulous, and this is more commonly seen in girls [4].

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. [4].

In untreated patients, due to continuous dribbling of urine, there can be mucosal abrasion, infection, squamous metaplasia resulting in acquired VUJ obstruction. The detrusor muscle may become fibrotic and scarred. Instances of adenocarcinoma of bladder have been reported in untreated adult patients. [4].

Diagnosis of exstrophy - epispadias complex can be made antenatally. Antenatal USG findings include [5,6].-

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

In neonates, exstrophy of bladder is diagnosed on clinical examination; workup includes baseline RFT 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. [7].

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

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 SI joints. Later reconstruction of bladder neck and sphincter is done [2].

Complications of closure is the failure to reach adequate capacity and thus augmentation or reconstruction may be necessary [2].

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 [1,2,3].Complications include stricture at the site of anastomosis, increased chances of adenomas and adenocarcinomas at the site of ureterocolic anastomosis [3], and hyperchloraemic acidosis.

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