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Exstrophy – Epispadias complex - Single stage complete primary repair a case report

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Abstract
We presenting a case of Exstrophy epispadias complex in a 1 ½ year old child. Exstrophy-epispadias complex (EEC) represents a spectrum of genitourinary malformations ranging in severity from epispadias to classical bladder exstrophy and exstrophy of the cloaca. Prevalence at birth for the whole spectrum is reported at 1/10,000, ranging from 1/30,000 for classical bladder exstrophy to 1/200,000 for exstrophy of the cloaca, with an overall greater proportion of affected males. In our case report we attempted single stage primary repair compared to multiple surgeries.

Keywords: Exstrophy-epispadias complex, Complete Primary repair of Exstrophy, Multi Staged Repair of Exstrophy

Introduction
The Exstrophy-epispadias complex (EEC) is the most serious form of abdominal midline malformation. The characteristic defects of the EEC involve the urinary system, the musculoskeletal system, the pelvis, the pelvic floor, the abdominal wall, the genitalia and sometimes the spine and the anus [1] The EEC covers a spectrum with different severity levels, ranging from epispadias representing the mildest form, with lower and upper fissure, to the full picture of classical bladder exstrophy, and exstrophy of the cloaca. Plenty of postulates and animal model studies were done to find the etiopathogenesis of Exstrophy-epispadias complex. Embryologically this defect is thought to be due to failure of the cloacal membrane to be reinforced by the ingrowth of mesoderm [2]. The management of this defect is very complicated owing to the complex defect involved, associated anomalies that need to be addressed. Traditionally, this defect is treated with multi staged procedure [3, 4] or a novel single stage approach [5].

Case Report
1 ½ year old male child brought by their parents with complaints of passage of urine from lower abdominal wall since birth. No other associated complaints.
On examination, lower abdominal wall defect seen with bladder protrusion. Pubic diastasis with classical exstrophy was observed. Bladder plate appeared epithelized with skin. No evidence of inguinal hernia noted. Left testis was palpable in superficial inguinal pouch. Right testis was seen in scrotum (pic 1). On x-ray pelvis pubis diastasis noted (pic 2). Visualized vertebra and bones appeared normal. USG KUB showed bilateral mild pelviectasis. 2D Echo was normal.
Intra operatively, an omphalocele sac was observed above the bladder plate. Bladder plate was dissected all around closed primarily in two layers. Epispadias repair done using Mitchell's Technique. Omphalocele sac was used for secondary closure (pic 3, 4)
Post operatively in our case child developed a small bladder neck fistula, needing secondary correction later.

Discussion
The Exstrophy-epispadias complex (EEC) is the most serious form of abdominal midline malformation. The characteristic defects of the EEC involve the urinary system, the

musculoskeletal system, the pelvis, the pelvic floor, the abdominal wall, the genitalia and sometimes the spine and the anus (1) Prevalence at birth for the whole spectrum is reported at 1/10,000, ranging from 1/30,000 for classical bladder exstrophy to 1/200,000 for exstrophy of the cloaca, with an overall greater proportion of affected males [6]

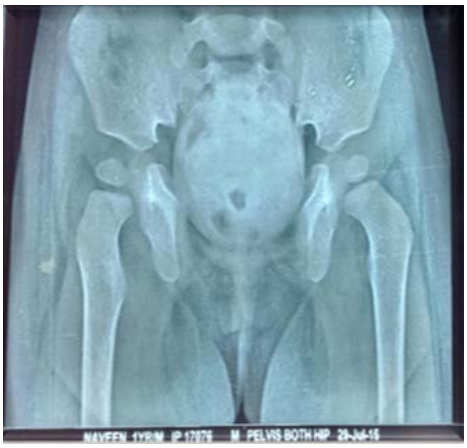
Boys with epispadias have a urethra that is extremely short and split and the opening is on the upper surface of the penis. Girls with epispadias have a urethral opening located between a split clitoris and labia minor.

Cloacal exstrophy is a severe birth defect in which there is usually a membrane-covered area on the abdominal wall that contains the abdominal contents (omphalocele). The bladder is divided in two halves and males have a penis split in two halves. Females have a clitoris divided in two halves and may have two vaginal openings. Other abnormalities are sometimes associated with the complex include a separation of the pubic bones, absence of the lower portion of the bladder causing lack of bladder control (incontinence) and abnormal position of the ureters causing back reflux in the kidneys.

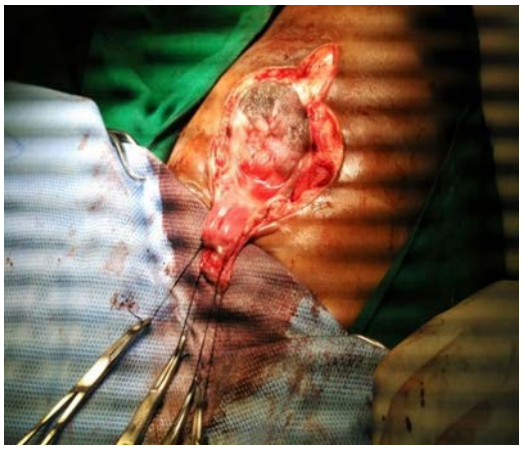
The management of this defect is very complicated owing to the complex defect involved, associated anomalies that need to be addressed. Traditionally, this defect is treated with a staged procedure called "Multi Staged Repair of Exstrophy" (MSRE) [3, 4]. However, presently a single stage approach called the "Complete Primary repair of Exstrophy" (CPRE) [5] is being done in selected children with the benefits of better bladder growth and continence in children treated with CPRE technique.

In our case report, we would like to share our experience in managing such a complex defect with the CPRE technique. In this patient the bladder plate was good with no other secondary changes. An omphalocele sac detached intra operatively, helped in reinforcing the bladder closure. Inspite of this, child developed a small bladder neck fistula, which needed a secondary surgery at later date.

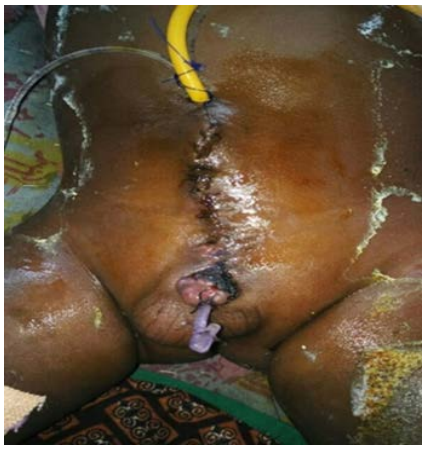
Picture 1



Picture 2



Picture 3



Picture 4

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