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BLADDER EXSTROPHY WITH TREATMENT IN THE INDIAN SCENARIO- A REVIEW

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ABSTRACT

Bladder exstrophy is one of the most rare birth disorder. An incidence of around 3.3 per 1,000 live births is recorded for classical bladder exstrophy, which is more prevalent in males compared to females. The urinary system, genitalia, bony pelvis, spine, anus, and ventral body wall can all show signs of anomalies caused by bladder exstrophy. Bladder exstrophy often involves the displacement of all tissues anteriorly, including the bladder, urethra, vagina, and rectum. Bladder exstrophy is notable for the abnormally-shaped bony pelvis. Approaches and strategies have evolved in the years that followed. If we talk about treatrment, a new method for closing the abdominal wall has replaced pelvic osteotomy; it involves re-anchoring the recti to the pubic rami following midline approximation, rather than removing them. At the same time as a successful method for repairing the bladder neck was described.

KEYWORD: Bladder exstrophy, bony pelvis, pelvic osteotomy, bladder neck.

INTRODUCTION

The urinary system, genitalia, bony pelvis, spine, anus, and ventral body wall can all show signs of anomalies caused by bladder exstrophy, one of the most serious birth disorders.[1] The first written account of an exstrophy bladder is around 2000 BC.[2] Even more than a century after Trendelenberg first proposed surgical treatment, pediatric surgeons still face the formidable obstacle of bladder exstrophy. An incidence of around 3.3 per 1,000 live births is recorded for classical bladder exstrophy, which is more prevalent in males compared to

females. Third Although several hypotheses have been advanced to explain its etiopathogenesis, we still do not know what causes it. There are no additional tests needed to confirm a clinical diagnosis of bladder exstrophy.[3]

ETIOLOGY

With a male-to-female ratio of 1.5 to 1, bladder exstrophy is incredibly rare, with an estimated 1 instance per 30,000 to 50,000 live births. According to research, normal mesenchymal ingrowth cannot occur in the lower anterior abdominal area when the cloacal membrane is either overgrown or persists. The result is a convergence of the lower abdominal muscles and a caudal fusion of the genital ridges with the cloacal membrane. The rupture causes either urinary tract exstrophy (sometimes called epispadias or typical bladder exstrophy) or cloacal exstrophy (when the hindgut is sandwiched between the two halves of the bladder), depending on the degree of ingrowth of the anorectal septum.[4]

EPIDEMIOLOGY

Very few babies are born with bladder exstrophy. About 2 out of every 100,000 live births is the prevalence rate according to the biggest international dataset.[5] Approximately 3.3 out of every 100,000 live births in the US are attributed to classical bladder exstrophy. The third point is that the disease affects boys at double the rate of girls. The number of men is reportedly six times that of females, according to some research.[6]

Its occurrence is lower in Western areas, among people of low socioeconomic position, and among people of non-white race.[7] In addition, research has shown that irradiation and maternal smoking during the first trimester enhance the likelihood of severe types of Bladder-exstrophy-epispadias-complex (BEEC). Contrarily, the probability of these severe types is reduced by periconceptional folate.[8]

PATHOPHYSIOLOGY

The BEEC complex encompasses a spectrum of congenital abnormalities from mild cloacal exstrophy to severe epispadias. Classical bladder exstrophy is defined by anomalies affecting the anus, pelvic floor, lower urinary tract, genitalia, abdominal wall, and bony pelvis.[6]

Bladder exstrophy often involves the displacement of all tissues anteriorly, including the bladder, urethra, vagina, and rectum. Bladder exstrophy is notable for the abnormally-shaped bony pelvis. A good way to describe it is as the "open book configuration" of the pelvis.

Pubic diastasis, anterior segment shortening, and posterior segment outward rotation are the most common bony pelvic anomalies.[9]

At birth, a kid with exstrophy has a pubic diastasis of around 4 cm; by the age of 10, it has grown to roughly 8 cm. On the other hand, the average human maintains a distance of less than 1 cm throughout their lives. One possible explanation is that the symphyseal ligaments have been torn. Second, there is a 30% reduction in length in the bony pelvis's anterior parts. The back portions of the bony pelvis, however, are within the usual range of measurement. Additionally, the bony pelvis's anterior and posterior halves are also externally rotated.[10]

HISTORY

Nearly four thousand years ago, the Assyrians recorded the earliest mention of bladder exstrophy on their tablets. A growing number of publications have been published since then. Based on what we know about the past two hundred years, nearly every surgical procedure has been documented at some point. In our opinion, the forefathers invested much in this area and brought both theoretical understanding and practical surgical expertise to the present day. We need to hold on to this priceless asset and enhance the treatment of bladder exstrophy patients in the future by incorporating the advantages of new technologies.[2]

TREATMENT IN THE INDIAN SCENARIO

A preliminary study (1974–1985) out of AIIMS, New Delhi[11] recommended waiting till after the neonatal phase to do the surgical repair. Repairs were always done in stages. To close large spaces, surgeons performed posterior iliac osteotomies, and to stretch the urethra, they employed para exstrophy flaps. When primary closure failed, permanent diversion via aureterosigmoidostomy, ileal conduit, or colonic conduit became an attractive alternative. Approaches and strategies have evolved in the years that followed. A new method for closing the abdominal wall has replaced pelvic osteotomy; it involves re-anchoring the recti to the pubic rami following midline approximation, rather than removing them.[12] At the same time as a successful method for repairing the bladder neck was described[13], other alternatives for enhancing continence and protecting the upper tracts were added, including bladder augmentation.[14] Since para-exstrophy flaps make self-catheterization more difficult, they have also been mostly abandoned. Also being practiced at the moment is a one-stage total repair during the newborn period.[15] From Vellore, we hear of a comparable shift in strategy and methodology.[16] Nowadays, a graduated functional reconstruction is the go-to method for fixing the issue, rather than urinary diversion. Augmentation cystoplasty and a catheterized

stoma, with or without bladder neck closure, are the recommended techniques due to the native bladder's inefficiency as a storage organ and the prevalence of upper tract deterioration throughout phased restoration.

From Vellore, we hear of a comparable shift in strategy and methodology.[17] Urinary diversion is no longer used, and a graded functional reconstruction is used to fix the problem. Since the native bladder is thought to be an inefficient storage organ and upper tract degradation is prevalent during staged restoration, the preferred methods are augmentation cystoplasty and a catheterized stoma with or without bladder neck closure. Ureterosigmoidostomy, bladder-conserving exstrophy repair, and bladder-enhanced exstrophy repair are still options that are used in select centers.[18] Patients who have a ureterosigmoidostomy often report an improved quality of life after the procedure. Patients whose repairs have failed may also benefit from total cystectomy or permanent diversion. The year 19 In another place, researchers found that a method that combined anterior iliac osteotomies with transverse bladder closure yielded better outcomes. In[19], The alternative treatments are bladder augmenting exstrophy repair, bladder preserving exstrophy repair, and ureterosigmoidostomy, which is still done in select centers.[20] Patients who have a ureterosigmoidostomy often report an improved quality of life after the procedure. Patients whose repairs have failed may also benefit from total cystectomy or permanent diversion.The year 19 In another place, researchers found that a method that combined anterior iliac osteotomies with transverse bladder closure yielded better outcomes.[21]

From Mumbai comes a novel theory on the embryogenesis of bladder exstrophy, and from this idea comes a new method. Sections[22–24] This technique elevates skin from the clitoris and labia majora in women, and the penis and scrotum in males, to a more optimal position for perineal healing. Lucknow has revealed yet another significant contribution to the treatment of bladder exstrophy. Methods for constructing an umbilicus and closing the abdominal wall that does not include pelvic osteotomy have been detailed. 24 and 25 Additionally, this facility has documented successful one-stage exstrophy restoration using a muscle stimulator to identify the pelvic diaphragm muscles.[25] One of the most important factors in the satisfactory urological result of exstrophy, abnormalities is reliable anterior pubic fixation. In this regard, researchers in Mumbai have documented a novel three-loop method for anterior pubic fixation.[26]

Delhi, Lucknow, Kochi, Hyderabad, and other cities have hosted live surgical workshops on bladder exstrophy and epispadias correction. Professionals from India and around the world have showcased their expertise and cutting-edge methods. Also, if you look around in the literature online, you may find a plethora of case studies detailing novel approaches to treating uncommon forms of bladder exstrophy and exstrophy variations. So, when it comes to surgically fixing the exstrophy-epispadias combo,[18] Indian pediatric surgeons are top-notch. What happens to these individuals beyond the first repair is not well documented, though. While I can't promise to cover every single reason here, I will say that there are likely several. Contamination, opening up, widening of the upper tract accompanied by degradation, fistulas, development of stones, incontinence, and a host of other significant problems are among the many occurrences that affect the postoperative period, according to the literature. Closure of fistulas, epispadias repair, restoration of the bladder neck, and bladder closure are common surgical procedures that often necessitate a second operation.[19]

Very few have documented long-term follow-ups, even though multiple authors/centers are operating for bladder exstrophy. These patients' dire circumstances need frequent hospital visits; the absence of long-term follow-ups indicates either poor documentation or unimportant findings. Only a small number of institutions have produced papers and given talks on topics unrelated to surgical procedure.[27-31]

DIAGNOSIS

In the case of **pseudoexstrophy**, the bladder's covering is only skin. Divergent recti and pubic diastasis are two of the observable musculoskeletal disorders. The bladder protrudes like a hernia when it is enlarged.[32]

A **superior vesical fissure** allows the skin to enter the bladder, causing the normally formed bladder to prolapse. The absence of musculoskeletal anomalies in patent urachus highlights the need to differentiate between the two diseases.

A normal phallus and normal bladder are present in a case of duplicate exstrophy. A suprapubic exstrophy mucosal plate is located next to a normal bladder. Due to the absence of ureters, the exstrophic bladder plate remains dry. This abnormality is thought to be caused by a superior vesical fissure that eventually fuses.[31-32]

The presence of a single ectopic intestinal segment distinguishes covered exstrophy from pseudo exstrophy. You may find it on the lower part of your belly. **Covered exstrophy** differs from other types in that it is characterized by anomalies of the external genitalia.[14]

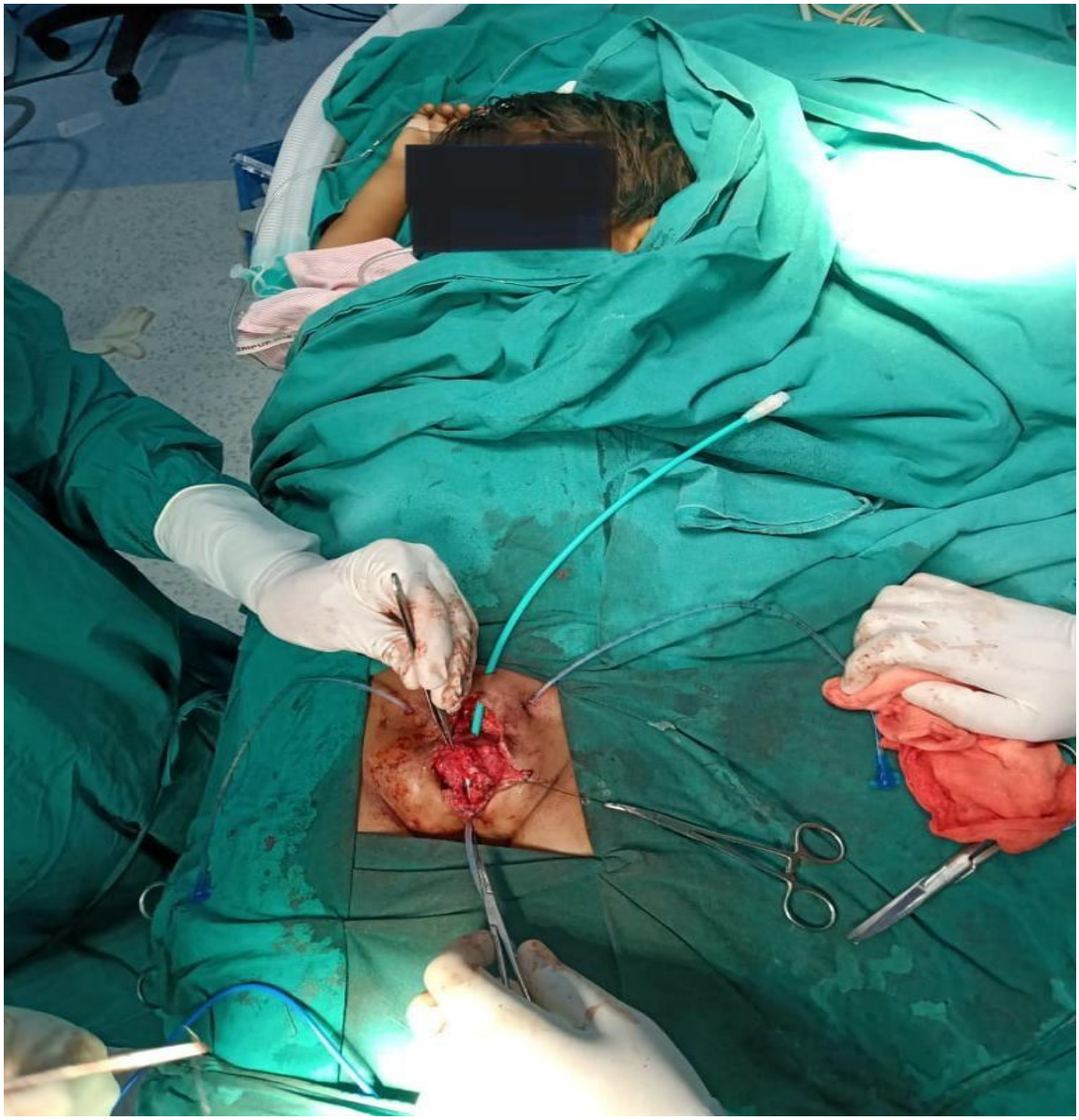

Fig. 1: Neck reconstruction as a surgical treatment for bladder exstrophy.

COMPLICATIONS

Unpleased micturition or dysuria: this consequence has decreased in frequency due to the decreased use of para exstrophy flaps; it is prevalent after open bladder procedures and can be caused by bladder spasms, wound infections, and urethral blockage. The first thing to do when urethral blockage or stricture happens is urethral dilatation. Urinary tract stricture, hemiglans/corpora loss, chronic dorsal chordee, and urethrocutenous fistula are frequent complications following epispadias treatment. The risks of bladder neck repair include UTIs,

bladder stones, and persistent incontinence. Risk of cancer as a consequence of osteotomy complications: Cancer of the bladder can develop in 1% to 2% of cases. The majority of people diagnosed with bladder exstrophy will likely have adenocarcinoma as their primary cancer. This can occur in bladders that have not undergone repair, according to the research.[32]

POSTOPERATIVE AND REHABILITATION CARE

Thorough postoperative care for children undergoing bladder exstrophy surgery is of the utmost importance. First, it contains parenteral medicines and postoperative analgesia. Epidural analgesia is used to control the pain that is experienced immediately. The youngster may experience pain from the immobilization and traction, therefore intravenous analgesics may need to be administered for an extended period. The youngster can begin receiving enteral feeding after they are completely awake. The lower extremities must be kept immobilized at all times. Furthermore, the majority of pediatric surgeons favorimmobilizing the lower limbs regardless of whether an osteotomy is done or not. The standard recommendation for external fixation after osteotomy is four weeks.[33]

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