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Bladder exstrophy with pubic symphysis diastasis in a male toddler: A case report

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ABSTRACT

Bladder exstrophy is a rare birth defect in which the anterior wall of the abdomen over the bladder does not close during the intrauterine period, leaving the child with an exposed bladder. Bladder exstrophy affects one out of every 50,000 births. Possible complaints include exposed bladder, incontinence or urine leakage, genital and pelvic bone defects, as well as intestine and reproductive organ defects. The majority of patients have their bladder exstrophy repaired as infants. If it is not addressed, as it was in this case, staged surgery will be required. A three-year-old male child with congenital bladder exstrophy was admitted to the Paediatric Surgeon's Outpatient Department in this case. Orthopaedic surgeons were consulted due to the pubic diastasis. At the age of three, an acetabulo-pelvic surgeon recommends using an external fixator on the pelvic wings to reduce tension on the reconstructed bladder wall and inter-pubic ligament. The patient was treated for undiagnosed bladder exstrophy. The reconstruction of the abdominal wall and urethra, as well as the use of an external fixator, was completed.

Keywords: Bladder exstrophy, External fixation in pelvic wings, stage wise surgery in neglected bladder exstrophy

1. INTRODUCTION

Bladder exstrophy is a complex, rare disorder that manifests itself early in a foetus's development in the womb (Ludwig et al., 2009). The abdominal wall does not fully form as the bladder develops, leaving the pubic bones separated and the bladder exposed to the outside skin surface through an opening in the lower abdominal wall (Shian and Larson, 2018). The bladder cannot store urine because the bladder and urethra are not closed. This open space is where urine from the kidneys drains. One out of every 50,000 live births are affected by bladder exstrophy, which is slightly more common in men. Different people experience the disorder in different ways and it can affect other organs such as the bowel, external genitalia and pelvic bones. Surgery is used to treat bladder exstrophy. The goal of treatment is to improve urinary control, maintain normal kidney function and improve the appearance and function of the external genitalia. If left untreated, normal urinary continence and sexual function are jeopardised. In this patient with

bladder exstrophy, the local abdominal wall is disrupted and the exposed inner surface of the bladder requires early closure. Pelvic osteotomies are required in severe cases where the bladder cannot be closed with soft tissue sutures alone. We used a light, Ilizarov external fixator to perform gradual positioning of bone fragments in a three-year-old. Following X-ray and CT analysis of pelvic deformity correction, volumetric capacity was gradually reduced.

2. CASE DISCUSSION

With complaints of an exposed bladder, incontinence or urine leakage, genital and pelvic bone defects, as well as intestinal and reproductive organ defects, a three-year-old boy and his parents visited the paediatric surgeon OPD (Figure 1). The baby was born in a rural setting at 8 months gestation and bladder exstrophy was not picked up on ultrasound. The majority of patients undergo bladder exstrophy repair surgery as infants, but because of a lack of funding, it has been neglected. Blood tests showed that everything was fine after the patient was admitted to the paediatric ward. Due to a midline defect, the bladder could not be seen in its normal anatomical position during abdominal and pelvic ultrasonography. A CT scan was done and the results showed bilateral undescended testicles, urinary bladder exstrophy and pubic diathesis. Both the paediatric surgeon and the orthopaedic surgeon were consulted regarding the osteotomy to reposition the bladder. The patient's suitability for surgery was determined following a preoperative examination. Under general and epidural anaesthesia, the full primary repair of bladder exstrophy, urethroplasty and glansplasty were carried out. The bladder was sealed following abdominal wall reconstruction (Figure 2). In order to treat bladder exstrophy, an oblique anterior pelvic osteotomy is performed and in order to treat pelvic diastasis, an external fixator is used. With a 5 cm incision on each side, the anterior approach (Iliofemoral or Smith-Petersen) was used. Over the lateral iliac crest and continuing to the anterior superior iliac spine, a five-centimeter skin incision is made. Dissection of soft tissues is finished. The ileum and iliac crest can be seen. To identify the osteotomy site, two K-wires were inserted. To reach the sciatic notch, oblique anterior pelvic osteotomies were done on both sides. In the right iliac blade, one proximal and one distal schanz pins were inserted. Two proximal and distal schanz pins were inserted into the iliac blade on the left side. Compression is finished using connecting rods. External Fixator is now finished. A thorough washing was performed using regular saline. Vicryl and ethilon were used in a layered closure. Lower limbs had mermaid costumes. The procedure was well tolerated by the patient. After the procedure, the patient is moved to a paediatric intensive care unit where their vital signs are carefully watched. For two weeks, analgesics and antibiotics were administered to treat the pain. The placement of suprapubic tubes, urethral catheters and a full course of antibiotics were also performed. Following surgery, total parenteral nutrition was frequently given and enteral feedings started around day 5. For three weeks, the patient was confined to a bed. The patient underwent physical therapy and received different day dressings. On day 2 (Figure 4) and three weeks later, both hips were post-operatively X-rayed, showing healed osteotomies with pins not penetrating hip joints and only a few complications. The external fixator was taken off after 35 days and a pelvic binder was suggested. In Figure 5, the patient was hemodynamically stable after 40 days and was released with a 15-day follow-up.

Figure 1 Patient came with Bladder exstrophy

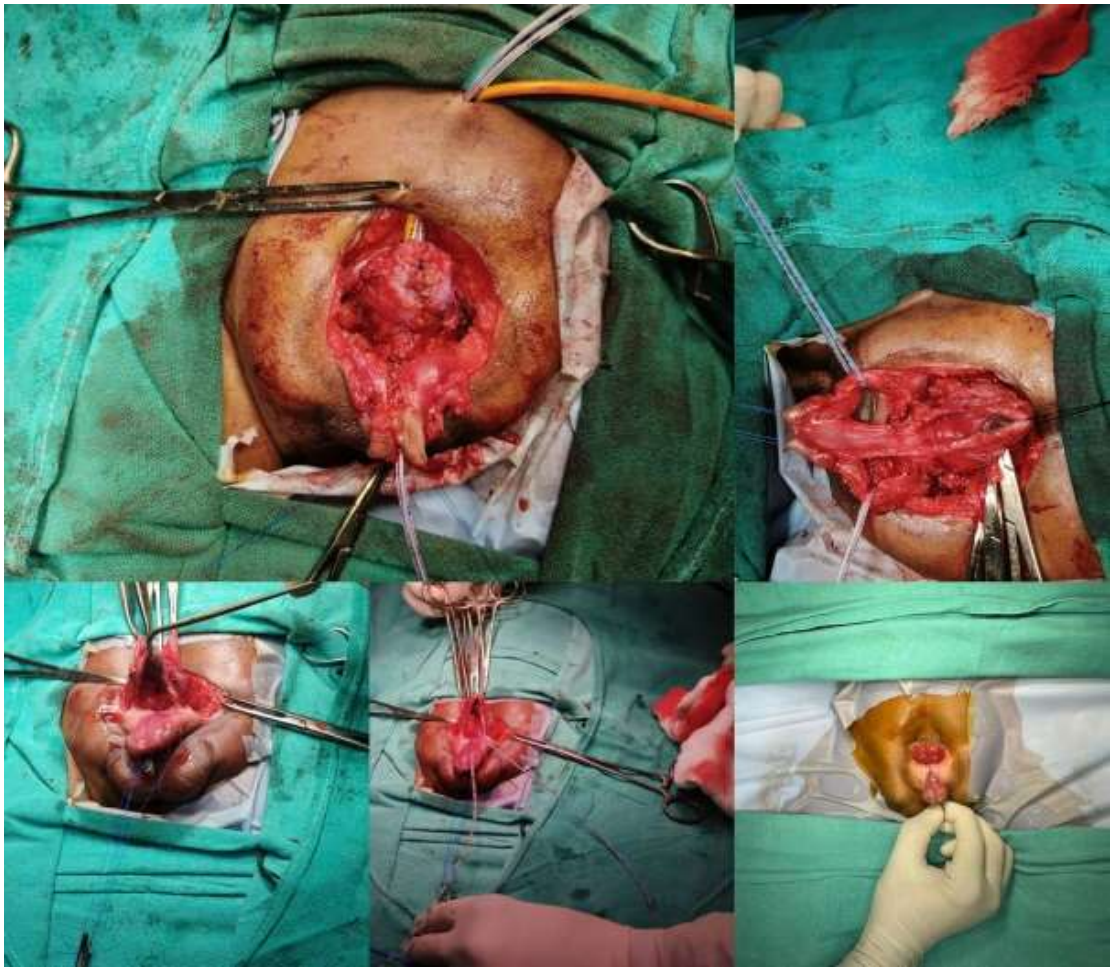

Figure 2 Intraoperative abdominal wall reconstructions for bladder

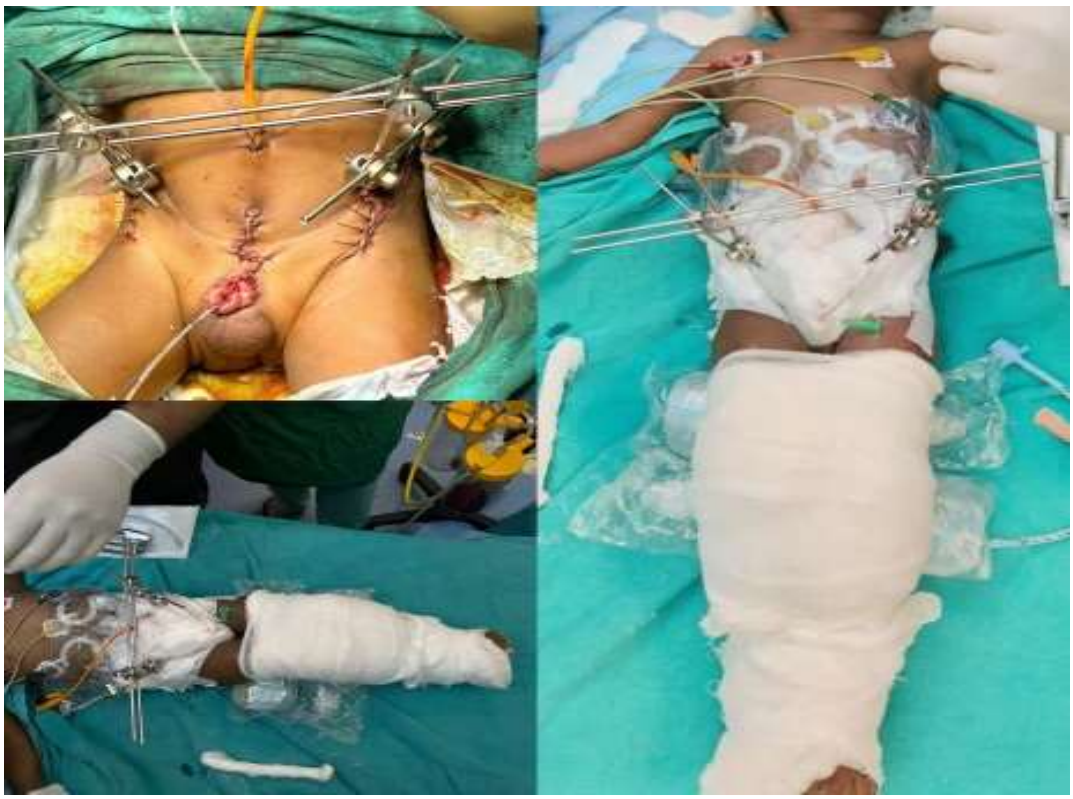

Figure 3 External fixator applications with post-operative clinical photograph

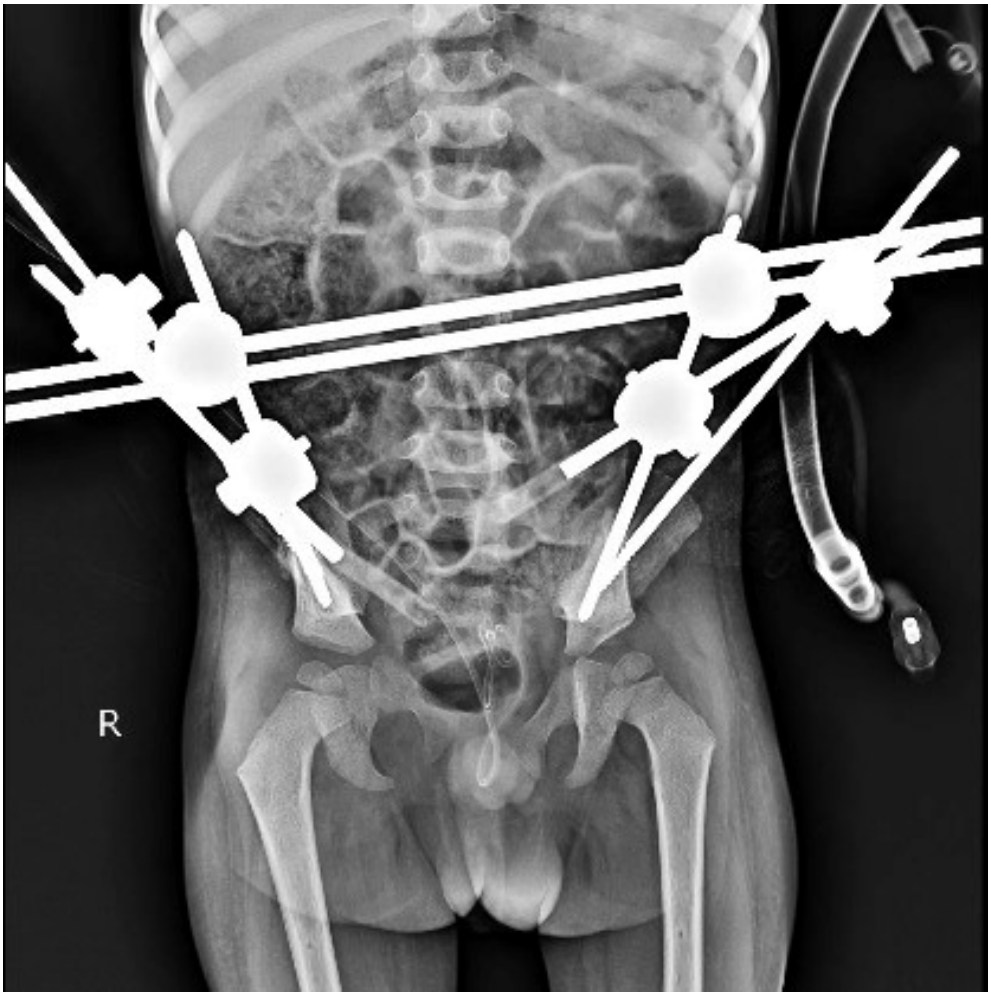

Figure 4 X-ray Pelvis with both hips post operatively on day 2

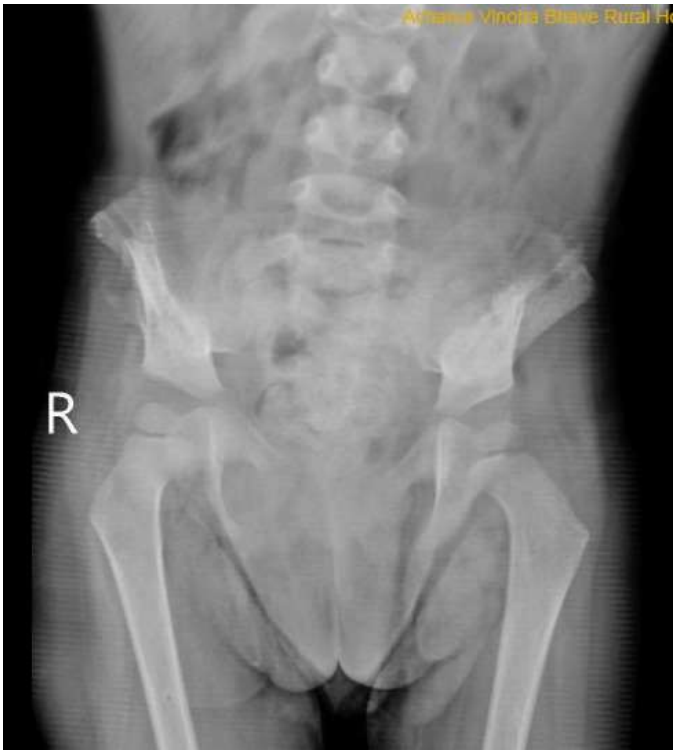

Figure 5 X-ray Pelvis with both hips post operatively after removing external fixator

3. DISCUSSION

A rare birth defect known as bladder exstrophy causes the bladder to develop outside of the womb. Due to the exposed bladder's inability to hold urine or perform normally, urine leakage occurs (incontinence) (Kajiwara et al., 2004). Issues with bladder exstrophy can range in severity. They can be problems with the intestine, reproductive system, pelvic, genital and bladder bones. During pregnancy, a routine ultrasound may reveal bladder exstrophy. However, the flaw might not be apparent until after the baby is born. Surgery will be required to correct the problems in infants with bladder exstrophy. When an ultrasound reveals that the baby's bladder is not filling and emptying normally, bladder exstrophy is suspected. To confirm the diagnosis, foetal imaging specialists will look for a low umbilical cord with an abdominal bulge below the cord insertion. Rarely are chromosomal or genetic syndromes, additional ultrasound findings or bladder exstrophy linked (Mardy et al., 2009). However, an amniocentesis may be advised for gender identification. If bladder exstrophy is not discovered while the mother is pregnant, the bladder defect is visible once the baby is born. In our case, bladder exstrophy was not identified during the antenatal period due to the rural setting and the patient's lack of follow-up. After the baby was delivered, it was determined that our patient had bladder exstrophy. In our case, the hip joint rotates outward and the pelvic cavity volume decreases. With lateral iliac osteotomy or horizontal iliac osteotomy, internal rotation of the distal bone fragment is used to treat these deformities. However, the latter is more likely to experience a significant blood loss because of the venous plexus at the location of the osteotomy. In addition, bladder exstrophy was treated and the pelvis' transverse diameter and cavity volume were both decreased. The two surgical complications for bladder exstrophy that happen most frequently are wound and bladder dehiscence. Wound and bladder dehiscence are the two surgical complications for bladder exstrophy that occur the most frequently. Bladder prolapse may be the result of complete dehiscence. Osteotomy, which relieves pressure on the bladder plate and abdominal wall during bladder turn-in and proper flap mobilization, can prevent it (Gearhart and Baird, 2005).

4. CONCLUSION

Patients with bladder exstrophy typically require long-term follow-ups, necessitating the involvement of multiple disciplines for their optimal care. The neonatologist, paediatrician and orthopedician play critical roles in the pre-operative management of these children. An early repair generally eliminates the need for an osteotomy, so a surgical consultation should be sought as soon as possible after birth. For osteotomy, a consultation with a trained paediatric surgeon and orthopedician is required. Long-term upper tract functional assessment requires the consultation of a nephrologist. As these patients enter adolescence and adulthood, certain psychosocial and sexual issues must be addressed properly. As a result, during their transitional care, they will need to see a psychiatrist as well as a urologist/andrologist.

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Informed Consent

Informed consent was obtained from the patient.

Author's contribution

All the authors contributed equally to the case report.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data collected during this study are available upon reasonable request from the corresponding author.

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