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Case Report

PRIMARY CLOSURE OF THE URINARY BLADDER, URETHRA AND ANTERIOR ABDOMEN OF CLASSICAL AND CLOACAL EXSTROPHY - A PRELIMINARY EXPERIENCE OF THREE CONSECUTIVE CASES

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Abstract

A 4 years old female and 1.5 months old male with classical exstrophy and a 2 days old male child with cloacal exstrophy were admitted in Paediatric Surgery Department of Sher-e-Bangla Medical College Hospital (SBMCH), Barisal. The mothers of all children had complaints of protrusion of fleshy mass in lower abdomen from birth. They always remain wet producing smell of urine.

The cloacal exstrophy case had a prolapsed ileum that was discharging stool. He had no anus too.

All of the cases underwent one stage closure of urinary bladder and urethra and lower abdomen without osteotomy and patient was discharged. After 8 months of first operation the female patient underwent repair of the wound dehiscence. At the time of discharge her urinary bladder capacity was 10 ml and remained dry for 5 minute after manual evacuation. The male patient with classical exstrophy developed urethrocutaneous fistula on 15th post-operative day. The cloacal exstrophy case was discharged with end ileostomy on 10th post-operative day.

Introduction

Classical exstrophy of the urinary bladder is a rare congenital anomaly. Prevalence of classical exstrophy is 1 in 40,000 and cloacal exstrophy is 1 in 200,000 live births[1]. Male to female ratio in both types is 2:1.

It results due to failure of the abdominal wall to close during foetal development and results in protrusion of the extrophied posterior bladder wall through the lower abdominal wall. So it is the spectrum of anomalies of lower abdominal wall, urinary bladder, anterior bony pelvis and external genitalia. Treatment is surgical correction of the defect. The aims of surgery are to obtain secured abdominal wall closure, achieving urinary continence with preservation of renal function, and adequate cosmetic and functional genitalia expected during childhood. Additional surgery might be needed to optimize bladder storage and emptying function[1], reconstruction of the continent anorectum ensuring faecal and good looking genitalia. Patient is considered socially continent if can remain dry for more than 3 hours during the day[2]. The purpose of the study is to identify the problems encountered by the patients and the parents that should be managed subsequently.

We are presenting consecutive three cases, which have been operated successfully in SBMCH.

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Case report

Case 1: A 4 years old female child was admitted in the Paediatric Surgery Department, SBMCH on 24-11-2008 with protrusion of fleshy mass in lower abdomen from birth. It always remain wet producing

smell of urine. There was a triangular gap in the hypogastric region occupied by extrophied wall of the urinary bladder plate including trigone. Right ureteric orifice was a golf hole appearance. Urethra was short. The mucosa of the Urinary bladder was thick and produced polyps due to chronic cystitis cystica. Upper urethelium was replaced by dermal epithelium (Fig.-11). There was a large sessile mass arising below the left ureteric orifice and this is due to cystitis cystica. There was bifid clitoris and labia minora with absence of mons pubis. Vaginal orifice was stenosed and anus was anteriorly placed. Patient had a small umbilical hernia and she had waddling gait. Live and spleen were not palpable and kidneys were not ballotable. Patient had no neurological deficit. She was mildly anaemic with normal pulse and blood pressure. Examination of urine, blood urea and creatinine were within normal range. USG shows moderate hydroureteronephrosis of the right kidney. Intravenous urogram indicated delayed excretion, dilated pelvicaleceal system and hydroureter of the right kidney. Her symphysis pubis is widely separated.

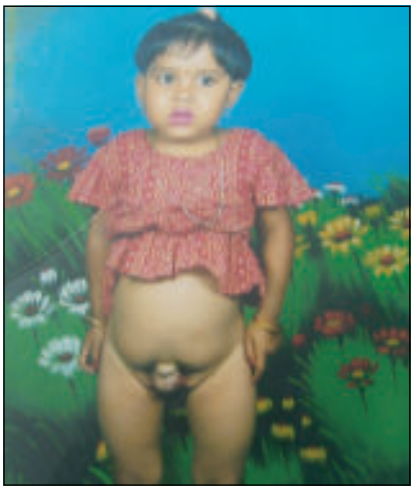

Fig.- 1: *Female child of 4 years of age with classical exstrophy*

Patient underwent primary closure of the urinary bladder, urethra, and lower abdomen without osteotomy of the pubis. The rectus abdominis muscles were dissected subperiosteally at their insertion and apposed in the midline. Caudal wound dehiscence occurred on 14th post-operative day (Fig.-2). After 8 months of the first operation patient admitted for secondary repair of the wound. Urethro-vesical closure done successfully (Fig.-3) with reinforcement of the gap between skin and underlying urinary bladder using surrounding soft tissue. She was discharged on 22nd post-operative day. Before discharge post-operative intravenous urogram (Fig.-4) showed well

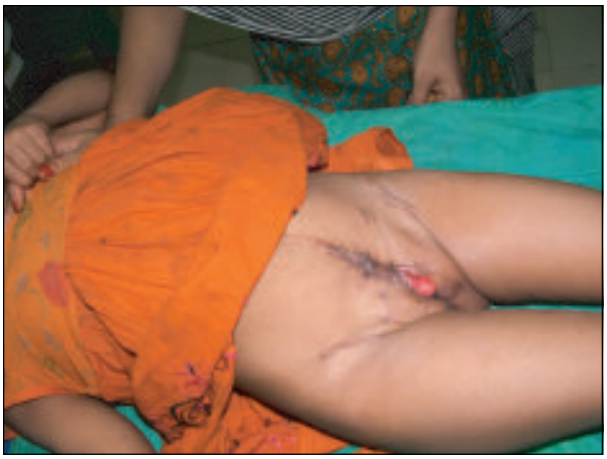

Fig.- 2: *Case 1 after 6 month of first operation.*

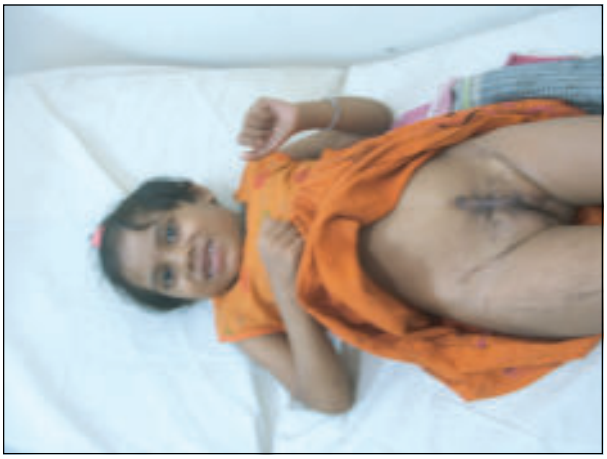

Fig.- 3: *Case 1 one month after second operation.*

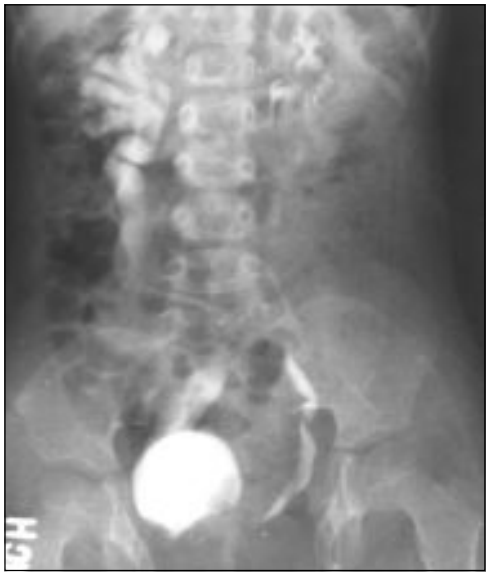

Fig.- 4: *Postoperative intravenous urogram showing small urinary bladder, hydroureteronephrosis of the right kidney and left renal partial duplication of pelvicaleceal system*

excretion of contrast by both the kidneys, right sided gross hydroureteronephrosis, and duplication of left upper ureter. Ultrasonogram of the urinary bladder indicates 10 ml capacity. At that time patient remain dry for 5 minutes after evacuation of the bladder.

Case 2: A male child of 1·5 months (Fig.- 5) got himself admitted in the paediatric surgery ward on 20-10-2009 with the complaints of a fleshy wet area at the middle part of the lower abdomen with smell of urine and a short divided penis.

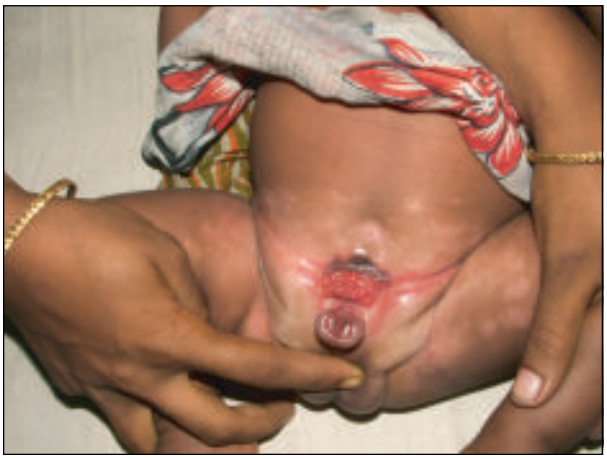

Fig.- 5: *One and a half months old male of case 2.*

Mother gave history of vaginal delivery after a full term pregnancy. Patient's birth weight was 2·75 kg. There was a triangular gap in the hypogastric region occupied by exposed urinary bladder with well-defined small trigone and two ureteric orifices. Urethra was extrophied and short. Penis is short and wide with a deep navicular fossa. Patient had right-sided inguinal hernia. There was diastasis of the symphysis pubis. Examination of other system appeared normal Patient's total and differential WBC count, bleeding and clotting time, routine and microscopic examination of urine, blood urea and creatinine were within normal limit. Intravenous urogram indicated functioning kidneys with normal ureters.

Patient was treated with one stage closure of the urinary bladder and urethra and anterior abdominal wall without osteotomy. On 15th postoperative days he developed small urethrocutaneous fistula at the level of the proximal part of the shaft of the penis (Fig.- 6). At the time of discharge he was advised to treat fistula after 6 months.

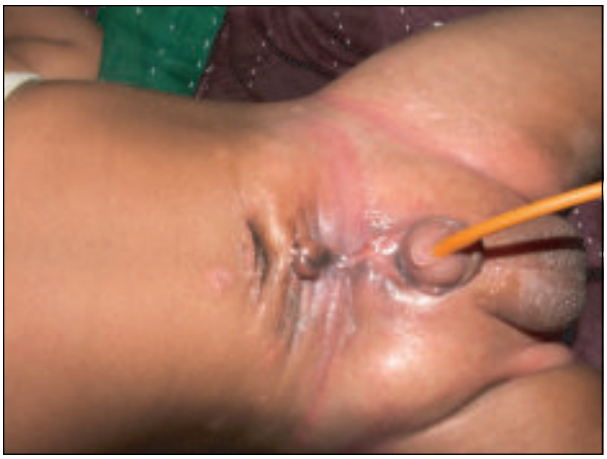

Fig.- 6: *Case 2 on 15th postoperative day with urethrocutaneous fistula.*

Case 3: A male child of 2 days weighing 2·4 kg was admitted on 19-01-2010 in the Paediatric Surgery Word with the complaints of red fleshy mass in the lower abdomen with bad smell of urine and stool. He was otherwise normal. There was a large triangular gap at the lower abdomen with absence of umbilicus. Structures of the umbilical cord were coming out at the apex of the gap (Fig.-7). Small intestine was prolapsing through an oval gap of the extrophied posterior wall of the urinary bladder and caecal plate.

Normal looking ureteric orifices is in the two helves of the extrophied urinary bladder mucosa. Trigon cannot be identified. Extrophied urethra is short and divided, and attested to the rudimentary bifid penis. The scrotum is normal looking and contain left testis. Right testis is in the inguinal region. Symphysis pubis is widely separated. Perineum is short with absence of anus. Other abdominal organs appeared normal. Examination of the other systems appeared normal

Fig.- 7: *A male child of 4days old with cloacal exstrophy.*

and did not have any associated anomaly. Patient underwent emergency operation with primary reconstruction of the urinary bladder and abdominal wall repair. Colon and rectum was absent. A 5 cm long pelvic appendix extending from behind the caudal end of the urinary bladder the cranial end of which was dilated and its lumen was empty. Terminal ileum was dilated and kinked at the junction of the caecal plate. The caecal plate was excised along with the kinked ileum and appendix and end ileostomy done. The ileostomy started functioning from 3rd post-operative day. The discharged stool was well formed. After primary closure, urinary bladder drained adequate urine (Fig.-8). On 4th post-operative day the frustrated mother and the grandmother wanted to take the child at home discontinuing treatment. The guardians gave up the idea after counseling. Patient was discharged on 10th post-operative day (Fig.-9) after explaining the possibility of problems and future plan of treatment with advice of regular follow-up examination protocol.

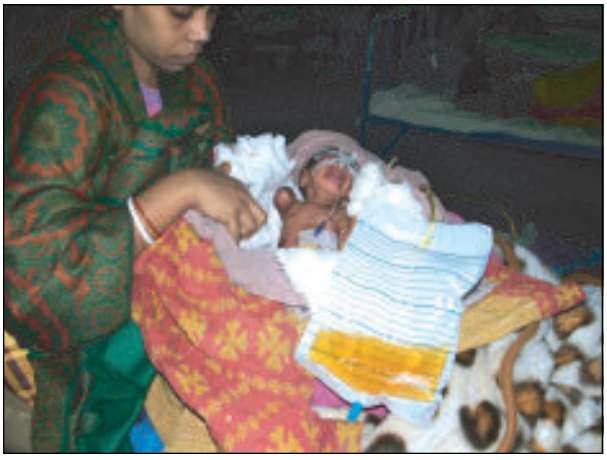

Fig.- 8: *Case 3 on third postoperative day with adequate urine drained.*

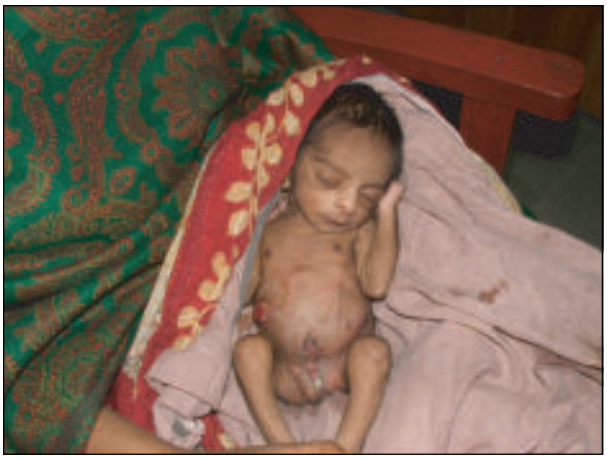

Fig.- 9: *Case 3 on tenth postoperative day before discharge.*

Discussion:

This classical exstrophy is commonly treated by staged surgical reconstruction of urinary bladder, bladder neck, urethra and genitalia that improves the quality of life of the patient. Initial experience from these three cases indicated that factors like continence, bladder capacity, and appearance of genitalia should be the main issue to be addressed in future to improve the quality of life of these patents. The reproductive and sexual function of the female patient should be possible with long-term multidisciplinary care. However, the female patent of case 1 has complications of vesicoureteric reflux, bladder augmentation and intermittent self-catheterization should be adopted to prevent further deterioration of the renal status. Her bladder template was not in a good condition and capacity after primary closure is small, she might be in need of bladder neck reconstruction. Purves et al also suggested such protocol [3]. Metrofenoff procedure is indicated if intermittent self-catheterization found difficult. Second male patient might achieve acceptable continence after epispadias repair and bladder neck reconstruction, which was also suggested by Baird et al [4]. In another series, long-term follow-up for urinary continence after reconstruction of classical bladder exstrophy indicated unpredictable outcome [2, 5].

Cloacal exstrophy represent abnormal embryogenesis involving the cloacal membrane. Theory yet to be established to explain the prolapse of the ileum, the blind end of the colon, and imperforated anus. Some suggested strangulation of the mid- or hind-gut. The aim of treatment is to have functional reconstruction

of the musculoskeletal, urological, gastrointestinal, and genital systems to facilitate nursing care and social support[6]. Cloacal exstrophy might require several staged procedures including urological, imperforated anus, genital, neurosurgical, orthopaedic, and plastic surgery. The result of bladder neck reconstruction in cloacal exstrophy significantly differs from that of classical exstrophy (25% versus 82%). Presence of neurological abnormality significantly hinders reconstruction of functional bladder [7]. Patient might be continent to faeces if associated anomaly is less severe[8].

Conclusion

One stage primary closure of the Urinary bladder and urethra without osteotomy and a soft tissue covering of the distal triangular gap is a better procedure for classical and cloacal exstrophy. Cloacal exstrophy was associated with initial high mortality in let 20th century. This high mortality was due to urosepsis and intestinal obstruction along with difficult care and social unacceptence. Now a day the survival rate is nearly 86%. Improved survival is due to recent advancement in surgical techniques, neonatal and paediatric intensive care, and improved modalities of monitoring of anaesthesia. Early primary one-stage closure is important because it laid the foundation for subsequent sequences of operation needed to improve the quality of life.

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