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Original Research Article

Bladder exstrophy: surgical management in older children and young adults: review of 29 cases in sub-Saharan surgical facility

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ABSTRACT

Background: Bladder exstrophy is a challenge to every surgeon in the world but it is more challenging in developing world. The condition is highly varied in nature and so are the procedures. Though rare it is associated with severe disability not to be able to have continence of urine and affect normal reproduction.
Methods: We retrospectively retrieved documents from hospital records
Result: We describe 29 cases of Bladder extrophy where 10 of which were treated with uretero-sigmoidostomy diversion. Bladder closures were done for 19 cases with 2 cases of bladder wall dehiscence. We also reported use of paraexstrophy skin to augment the contracted bladders in 9 older children.
Conclusions: Because of lack of awareness in the community, bladder exstrophy cases come late in the childhood or in adulthood. This has affected the result where many of them offered only diversion. In One third (9 cases) we used paraexstrophy skin to augment bladder capacity where we observed better outcomes.

Keywords: Bladder, Exstrophy, Urinary diversion

INTRODUCTION

Bladder exstrophy is also known as ectopic vesicae. Bladder exstrophy is a rare congenital malformation of the genitourinary system, with an estimated incidence of approximately 1 per 50,000 live births.[1] The exstrophy-epispadia complex represents a severe midline abdominal birth defect that causes wide separation of the pubic symphysis, an abdominal wall defect and an anteriorly positioned open bladder and urethra.[2] Although in rich countries, patients present early in infancy before one year of age, many cases in developing countries still come very late school age or even in young adulthood. This late presentation was because of a lack of awareness of the parents and the local health service centers that it could be surgically reconstructed.

Surgeons therefore find it difficult to reconstruct the condition according to the textbook description. On the side of patients and family, find the repeated procedures and possibly failure of reconstructions constraining with finance and time, we therefore had to do some improvement of older children bladder extrophy closure using the skin around it to augment capacity of bladder. Unfortunately, many of our patients come after the bladder mucosa has contracted and we had to excise it and urinary diversion in the form of uretero-sigmoidostomy had to be done.[4,5]

In general, bladder exstrophy is demanding both for the surgeon and for the patient. Our review indicates, that the cases are still at home because of lack of awareness and do not come at appropriate age of surgery i.e., before the age of 1 year.[6] It has also shown that the community health service provider should recognize the problem and execute early transfer to better centers.

The objective of this research is to make the surgeons in developing country who are facing this problem to use as

much paraexstrophy skin as much possible to augment the capacity of bladder in young adult patients.

METHODS

This is a retrospective study performed in University of Gondar Hospital during the period of August 2002 to December 2022. All 29 patients presenting with bladder extrophy and operated during the last 20 years at university of Gondar hospital were included. Medical records reviewed and described. Type of the problem, surgical procedures outcomes of procedures analyzed using a computer application software namely EPI info 6. The types of procedures used were as described below. All patients have given informed consent for both treatment and their data to be used for research

Bladder closure

Endotracheal tube general anesthesia with muscle relaxants. In older patients, the patient is placed in the supine position and a thorough hand scrub with chlorohexidine and povidone iodine is given. A circumferential incision is made around the exstrophied bladder just outside the mucocutaneous junction. Cephalad, a triangular piece of skin, is excised, and caudad, the incision, is carried on to the urethral plate on the epispadic shaft. The incision is deepened from the cephalad extent and a plane is created between the peritoneum and the bladder wall.[3] The ligaments of the bladder are divided. The urethral plate is then dissected off the corpora cavernosa an intersymphysial band of tissue is now seen and this is divided from the symphysis and mobilized for a short distance so that it can be brought to the midline. The distance between the mobilized urethral plate and the distal end of the incision on the epispadic shaft is measured and equal lengths of paraexstrophy in older children where the bladder mucosa is contracted, (Figure 1).[5] We took some of the skin circumferentially (Para exstrophy) with bladder wall to increase the capacity of bladder volume. As we took some skin to compensate capacity, there was even more need to flap sliding to cover abdominal wall defect which was managed by lateral relaxing incisions (Figure 2-5).

Figure 1: Bladder exstrophy before reconstruction in older child of age 8.

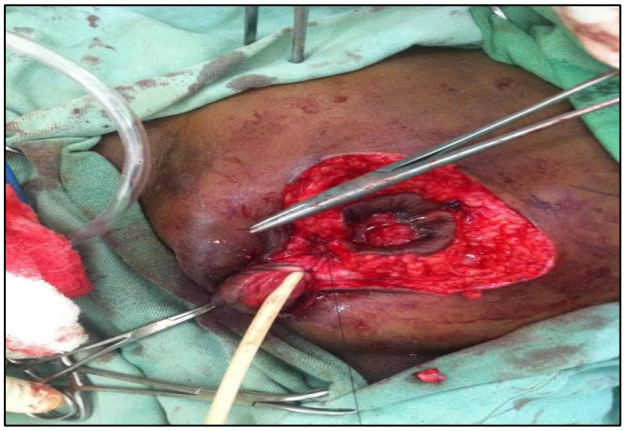

Figure 2: How para exstrophy skin mobillzed to increase bladder capacity.

Figure 3: Bladder closure technique using the para exstrophy skin.

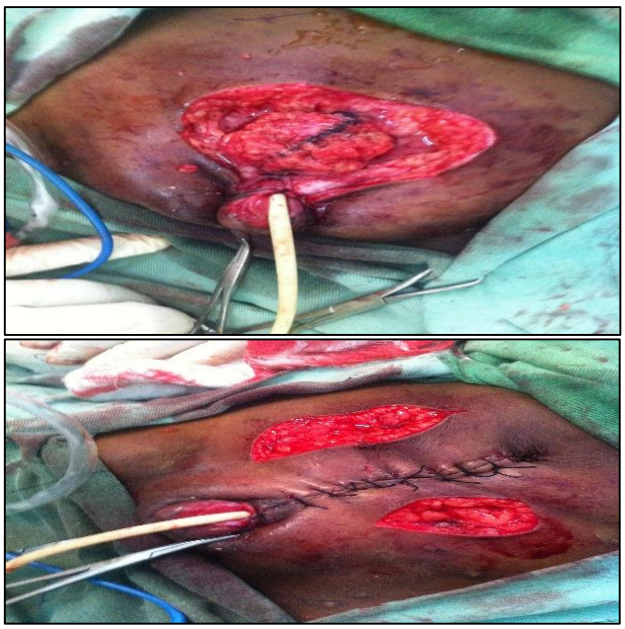

Figure 4: Abdominal wall closure with sliding flaps and relaxation incisions.

The two paraexstrophy flaps are sutured to each other in the midline in two layers using 4/0 or 5/0 polygycolic acid) sutures and the suture line is continued to approximate the proximal free edge of the paraexstrophy flaps to the urethral plate. Using the same suture material, the lateral edges of the paraexstrophy flaps are brought together in the midline and sutured to each other over an 8F tube feeding tube. The paraexstrophy flaps are thus tubularized.

Both the ureters are cannulated with 5-F infant feeding tubes which are anchored at the ureteric orifices with 4/0 chromic catgut. stents are brought out by lateral stab incisions, and are anchored at the skin with 4/0 silk sutures. The subcutaneous tissue and skin are closed in layers. The ureteric stents are removed after 2 weeks and the bladder catheters 3 weeks after surgery. We used sliding skin flap bilaterally and subcutaneous tissue to close in cases where circumferential skin is used to augment bladder capacity.[6]

Bladder neck repair

This procedure is undertaken at least 1 year after primary closure of the bladder exstrophy. Here we used standard procedures as described By many authors.[2,3] However the time is decided on by the parents when they are ready both financially and logistically. Many of the bladder neck procedures were done 3 months after the initial procedure as many of our patients are older children

RESULTS

A total of 29 cases of bladder extrophy of various types have been operated in Gondar university hospital. Classical bladder extrophy with no epispadia accounted for 12 cases, whereas exstrophy-epispadia complex occurred in 7 children. Adult cases of bladder exstrophy reported in 10 cases where age at presentation is above 18 years. All of them were female patients. None of them developed any cancerous or pre-cancerous patients in those adult patients (Table 1).

Table 1: Age distribution and outcome of bladder exstrophy reconstructive surgical operations at university of Gondar hospital between AUG 2002 TO Dec. 2022.

Age range (years)	Number of cases with bladder closure in 2 staged procedures (failures*)	Percentage of success (%)	Number of bladder closure in extrophy-epispadia complex (Failures)	Percentage of success (%)	Uretero-sigmoidostomy and excision of the bladder wall (Failures)
2-5	6 (2)	67	4 (1)	80	0
5-10	2 (0)	100	2	100	0
10-18	4 (0)	100	1	100	0
Above 18	0		0		10 (0)
Total	12 (2)		7 (1)		10 (0)

Failures* defined when there is total dehiscence of bladder wall repair.

Closure of the bladder in classical extrophy patients were done12 patients after the necessary investigation completed. Out of which 10 had successful closure with frequency of urination approximately every 3 hour. Requirement of diaper was only during the night for those children under the age of 5 years. Two of them had bladder dehiscence due to concomitant presence of symphysis pubis diathesis. Osteotomy was not performed in any of the cases.

There were seven patients with bladder extrophy-epispadia complex who underwent staged procedures. We were able close the bladder in all of them, while one of the patients had his epispadia dehisced and was rendered incontinent. He was then referred to another center.

In the last 20 years, we operated up on 10 adult patients who were all females and had classic hypospadias. The bladder mucosa was atrophied and bladder closure was not possible. They were counseled for urinary diversion (uretero-sigmoidostomy) and bladder excision followed by abdominal wall approximation. Bladder wall excision and approximation of bladder wall was done few months after urinary diversion. No major electrolyte and acid base abnormality was detected following diversion surgery. Ileal or colonic conduits were not used as there were no appliances available on the market. On follow up, all of them reported that they were dry day and night and had no fear of participating in social and public activities.

DISCUSSION

As in many developing centers our patients were presented to the care very late in childhood or early adulthood.[6] These presentations had posed more technical difficulties to the reconstructions as the bladder walls were to certain extent contracted. In older children, we included some skin from the abdominal wall which is adjacent to the bladder mucosa used to augment the capacity.[5] We found this technique very useful in terms of both increasing capacity of the bladder as well as taking sutures when reconstructed. Using the adjacent skin to make up bladder capacity is also mentioned in the literature.[5] We recommend that in those young adults

presenting late with bladder extrophy to use this technique.

Bladder neck repair was done at least 3 months after the initial surgery as most of them are young children and were able to tolerate major surgical problems. Therefore, as it is mentioned in many reports, the second stage bladder neck repair or epispadias repair or any revision can be carried out if the cases are above 5 years old, we suggest.[2,3] Diversion using uretero-sigmoidostomy in 10 patients performed. They had very much contracted bladder which were not amenable to bladder closure. After thorough consultation the 10 patients underwent the procedure and the result was found satisfying.[6-8] The most important factor is being dry all the day and night and were happy that they could socialize, go to work and sleep together with family without fear of isolation.[9,10] However, due to the limited data, some of the recommendations we mentioned should be reproduced with larger data and controlled study.

CONCLUSIONS

With all the scarcity and the problem of African surgical facility, it was possible to help these individuals to make them live with the society without fear by diverting the urine to the sigmoid colon. When parents are not informed about the condition, they tend to present very late in childhood or late in puberty. The inclusion of some paraxial skin to augment the capacity is something worth to consider, though it is reported in 30% urethral stricture complication if used for urethral reconstruction. There was even more need to flap sliding to cover the abdominal wall defect which was managed by lateral relaxing incisions

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