CORRECTION OF BLADDER EXSTROPHY IN ADOLESCENT: 2 CASE REPORT

Burgio A., Di Maggio G., Ferrara F., Giannotti G., Pavone M., Bulotta A.L., Messina M.

Division of Pediatric Surgery, Department of Pediatrics, Obstetrics and Reproductive Medicine
University Of Siena

Introduction: bladder exstrophy is characterized by an incomplete development of anterior portion of bladder, urethra, abdominal wall and a median separation of pubic symphysis. We describe 2 clinical cases in adolescent treated with different surgical approach. ***Materials and Methods:*** two patients, one male and one female, respectively, 10 and 14 years old. Both cases had relapse: the bladder plate was performed in female when she was 2 years and in male when he was 6 years old. The male showed bladder exstrophy with epispadias; the female showed bifido clitoris, absence of minora labia with a small vaginal orifice. In both patients ano-rectal manometric and barium enema had normal parameters. The x-Ray basin showed diastasis of pubic bones, respectively, 7.5 cm in male and 10 cm in female. The male had closure of bladder and abdominal wall with transformation of epispadias to peno-scrotal hypospadias. The female had rectal reservoir (about 250cc), with amputation of the rectum with Hartmann's method and recanalization with Duhamel's method, anastomosis uretero rectal and vaginoplasty. ***Results:*** clinical and laboratory follow-up to date is 6 months. The female holds the urine about 2-3 hours during the day; in the night losses are not reported; the male shows dehiscence of abdominal and vesical wall with suprapubic fistula and loss of urine. The urethral peno-scrotal meatus appears patent. ***Conclusions:*** our experience, although limited in the number of cases and in the time of observation, is in agreement with the literature: we can say that the correction of bladder exstrophy in adolescents might be a urinary diversion with creation of rectal reservoir, useful for age of the patients and postoperative management. It is always necessary an adequate follow-up to check any complications.

INTRODUCTION

Bladder exstrophy is characterized by an incomplete development of anterior portion of bladder, urethra, abdominal wall and a median separation of pubic symphysis. Urine is dripping from the ureteric orifices on the bladder surface. Marshall and Muecke proposed a persistent cloacal membrane in the epispadias-exstrophy complex causing a "wedge effect", preventing the mesodermal tissue from closing in the midline (1). The incidence has been estimated to be between 1/30.000 – 50.000 live births; 2-3/1 ratio of male-to-female exstrophy births.

Due to high-resolution real-time ultrasound, prenatal diagnosis of bladder exstrophy is usually possible between the 15 th and 32 nd week of gestation, depending on the severity of the defect and the expertise of the sonographer. The index finding is the non-visualization of a normally filled fetal bladder during repeated careful ultrasound examinations. Though prenatal surgery is not necessary, early diagnosis allows optimal postnatal management (2).

In boys the anomaly consists of an exposed everted bladder in continuity with an epispadiac penis. The pubic bones are widely separated and the external sphincter complex is represented only by a fibrous interpubic bar and has no recoverable muscle function. The penis itself is short and with upwaed chordee. In exceptional cases it can be very small and sex assignment as female may be appropriate. In girls, the anomalies are similar with a bifid clitoris and a separate vaginal opening.

Regardless of surgical approach, successful initial closure of the bladder is critical to the development of adequate bladder capacity and ultimate continence in patients with bladder exstrophy (3). Failure of initial closure, and the need for subsequent re-closure, severely impacts the patient's potential for eventual continence (4).

Due to lack of medical care and public awareness in certain underserved localities, few cases not receive treatment during infancy and present during adolescence.

We describe 2 clinical cases in adolescent treated with different surgical approach.

CASE REPORT

We reported two cases: a ten year old boy and fourteen year old girl.

Case 1: AY ten year old boy was submitted to closure of bladder exstrophy, in another country, when he was 9 years old it relapsed after two weeks.

On admission in our clinic the patient showed exstrophy-epispadias complex, urethral plate covered the whole dorsum of the penis from the open bladder to

Correspondence to:
Prof. Mario Messina, Division of Pediatric Surgery, Dept of Pediatrics, Obstetrics and Reproductive Medicine.
University of Siena. Policlinico "Le Scotte" - Viale Bracci - 53100 - Siena - Italy
Telefono: +39 577 586501 - Fax: +39 577 586174
E-mail: messinam@unisi.it

the glandular grove (fig.1). Both corpora cavernosa were located beneath the urethral plate. The penis appeared shorter than normal and dorsally located. Absence of the umbilical scar. The normal-sized testes were located in the scrotum. Anal region were normal.

Routine laboratory and assessment of baseline renal function were performed and were normal; ano-rectal manometric and barium enema had normal parameters; basin x-Ray showed diastasis of pubic symphysis approximately 7.5 cms. No associated spinal, orthopedic and gastrointestinal anomalies.

The patient was submitted to closure of bladder exstrophy and partial correction of epispadias. Ureteral catheters were placed. The bladder was mobilized preserving dissection laterally and toward the pubic bones. Urethral plate was separated from corpora cavernosa; the uretra plate detached from the corpora allowing a more effective correction of glans by rotation of the corpora with transformation of epispadias to peno-scrotal hypospadias. Bladder was closed and the uretra was placed ventrally. The patient showed dehiscence of abdominal and vesical wall with suprapubic fistula and loss of urine. The urethral peno-scrotal meatus appears patent.

Case 2: AH a fourteen year old girl was submitted to closure of bladder exstrophy, in another country, when she was 2 years old and failed. When she was ten years old she was submitted to surgery for right renal lithiasis.

On admission in our clinic the patient showed bladder exstrophy, bifido clitoris, absence of minora labia with a small vaginal orifice (fig.2). The patient showed low self-esteem and lack of social communication. Routine laboratory and assessment of baseline renal function were performed and were normal; ano-rectal manometric and barium enema had normal parameters; basin x-Ray showed diastasis of pubic symphysis approximately 10 cms and abdomen ultrasound showed first grade hydronephrosis on the left kidney. No associated spinal, orthopedic and gastrointestinal anomalies. The patient had closure of bladder exstrophy, rectal reservoir (about 250cc), with am-

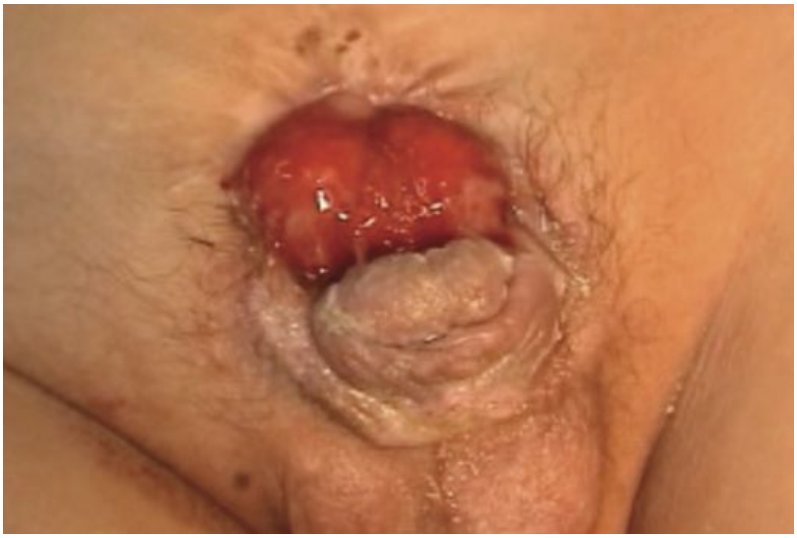

Fig.1: Exstrophy-epispadias complex in the male (image before surgery)

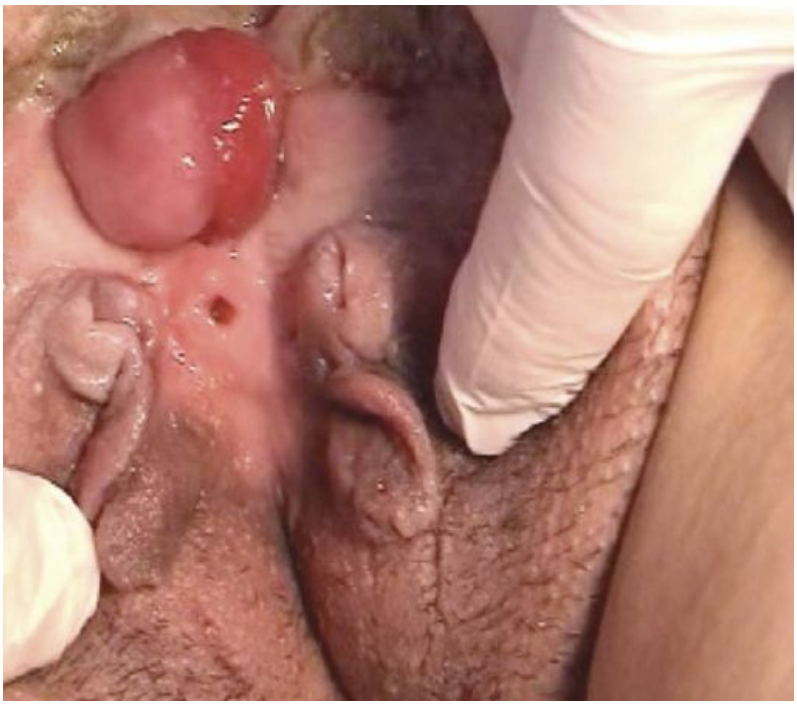

Fig.2 (above): Bladder exstrophy with bifido clitoris, absence of minora labia and small vaginal orifice(image before surgery)

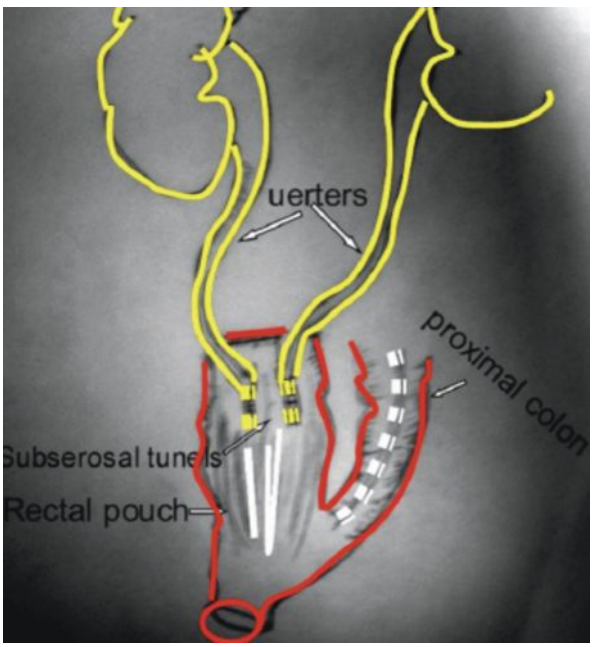

Fig.3 (right): Image of surgery: Duhamel pullthrough with ureterorectostomy

putation of the rectum with Hartmann's method and recanalization with Duhamel's method, anastomosis uretero rectal and vaginoplasty.
The sigmoid colon was divided above the rectum. The colon was passed behind the rectum, anastomosed to the anal canal and the posterior rectal wall above the dentate line and internal sphincter. The two ureters were detached from the bladder and were implanted in a subserosal tunnel on the lateral rectal wall below the upper edge of the rectum. Transanastomotic ureteric catheters were passed into the rectal space and were accessible through the anus. The proximal rectum was closed in two layers and abdomen closed leaving a pelvic drain. Blood transfusion was not necessary, the patient began act kinesis of the pelvic floor at 15 days and she also showed urinary continence during the daytime with an emptying frequency of 5-6 times, losses are not reported in the night. Following repair, the patient showed great satisfactory with their body image and improvement in their social interaction and self-esteem. The patient was discharged after 25 days. Two mounths after surgery abdomen ultrasound demonstrated normal appearance and blood gases, serum electrolytes, BUN and creatinine serum were estimated with maintenance of values within the normal ranges. The patient is still in follow-up.

DISCUSSION

Reconstruction of exstrophy- epispadias complex remains one of the greatest challenges facing the paediatric urologist and the results are as yet uncertain. The goals of therapy include urinary continence with preservation of renal function, and reconstruction of functional cosmetically acceptable genitalia (5).
In developing countries where early surgical reconstruction is not done routinely and there are limited technical resources, there is a high incidence of failure of bladder reconstruction for children with bladder exstrophy. Many, either primarily or following failed surgery, will come to consideration of diversion. Also the social taboos and culture in our locality often do not favour an external stoma and add to the need for an internal continent urinary diversion (6).
Pahernik et al. report their long-term results of conversion from a colonic conduit into a continent anal urinary diversion, as after conduit urinary diversion in childhood, some patients wish to have a later conversion to a continent diversion to avoid external appliances and to improve their quality of life. Specific advantages such as an improved body image and a more independent lifestyle without external appliances seem to justify these more complex procedures (7). The past 15 years have seen an increasing interest in continent diversion of the upper urinary tract. The concept of refashioning the bowel to serve as a urinary reservoir rather than a conduit is based on the original pioneering observations by Goodwin and others in the development of the augmentation cystoplasty (8). The distruction of peristaltic integrity and refashioning of the bowel has led to the development of many innovative urinary reservoirs constructed from bowel, using antireflux procedures to avoid upper tract sepsis and additional surgical techniques to achieve urinary continence (9).
Many of the 40 or so variants of continent urinary diversion presently used represent modifications of the parent procedures.The multiplicity of procedures reflects the fact that most remain less than satisfactory and the "best" continent diversion has yet to be devised. It is however becoming apparent that certain procedures are associated with lower early and late complications . Mohamed A. et al. used the principles of the Duhamel pullthrough technique to internally divert urine for children who have failed to establish acceptable functional bladders after several operations for bladder exstrophy. This procedure is associated with minimal pelvic dissection and achieves a reasonable rectal reservoir. There is the theoretical advantage of avoiding a mix of urine and faeces, where the upper part of the rectal pouch will serve as a reservoir for urine and the faecal stream will come from the proximal bowel to the lower part of the rectum with preservation of the anal sphincter (10), (fig.3).

CONCLUSION

Our experience, although limited in the number of cases and in the time of observation, is in agreement with the literature: we can say that the correction of bladder exstrophy in adolescents might be a urinary diversion with creation of rectal reservoir, useful for age of the patients and postoperative management. It is always necessary an adequate follow-up to check any complications.

REFERENCES

1.Marshall, V. F. And Muecke, E. C.: variations in exstrophy of the bladder. J Urol, 88: 766, 1962.
2.Anne –Karoline Ebert, Heiko Reutter, Michael Ludwig and Wolfang H Rosch. Orphaned Journal of Rare Disease 2009, 4:23
3.Oesterling JE, Jeffs RD. The importance of a successful initial bladder closure in the surgical management of classical bladder exstrophy: analysis of 144 patients treated at the Johns Hopkins Hospital between 1975 and 1985. J Urol 1987; 137:258-62.
4.Gearhart JP, Ben Chaim J, Sciortino C, Sponseller PD, Jeffs RD. The multiple reoperative bladder exstrophy closure: what effects the potential of the bladder? Urology 1996; 47:240-3.
5.Benson MC, Olsson Ca. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ, editors. Campbell's urology. 7th ed. Philadelphia, PA: WB Saunders; 1998. P.3190-227.
6.Elcoat C. Coping with stoma care in the community. Practitioner 1989;22(1469):776-9.
7.Pahernik S, Wiesner C, Gilltzer R, Stein R, Thurof JW. Conversion from colonic conduit into recto-sigmoid pouch (Mainz pouch II).BJU Int 2006;97:157-60.
8.Goodwin WE, Harris AP, Kaufman JJ, Beal JM. Open transcolonic ureterointestinal anastomosis: new approach. Surg gynaecol Obstet 1953;97:295-300.
9.Bastian PJ, Albers P, Haferkamp A, Schumacher S, Muller SC. Modified ureterosigmoidostomy (Mainz Pouch II) in different age groups and with different techniques of ureteric implantation. BJU Int 2004; 94(3):345-9.
10.Mohamed A., Baky Fahmy., abo Zid Aoud Mansour , Alaa Mazy. Ureterorectostomy as a continent urinary diversion for complicated bladder exstrophy in children by using a modified Duhamel procedure: a case series. International Journal of Surgery (2007) 5, 394-398.