State-of-the-Art Reconstructive Surgery for Bladder Exstrophy at The Johns Hopkins Hospital

John P. Gearhart, MD, Robert D. Jeffs, MD

• Bladder exstrophy is a congenital anomaly that has been difficult to correct. Advances in reconstructive surgery, some of which have originated at The Johns Hopkins Hospital, Baltimore, Md, have impacted on the care of these patients. We reviewed the progress made in patient care and the results of state-of-the-art treatment.

(*AJDC*. 1989;143:1475-1478)

Reconstructive surgery for serious congenital anomalies has shown tremendous advances in the 100 years since Johns Hopkins endowed the hospital that bears his name. Bladder exstrophy is a devastating defect that illustrates this progress in treatment. The advance was given impetus by Hugh Hampton Young, the founder of the Brady Urological Institute, The Johns Hopkins Hospital, Baltimore, Md, and by other innovations from the same institution.

Early attempts at bladder closure to produce a urine receptacle failed due to dehiscence and infection or failed to produce continence. Trendelenburg,[1] in 1906, first attempted osteotomy as an adjunct to closure of the bladder, but the first sporadic success was reported by Burns[2] in 1924.

The lack of success in reconstruction led to many alternatives for urinary control, beginning with Magdl[3] in 1894 who successfully transplanted the trigone into the rectum. The ureterosigmoidostomy performed by Coffey[4] and improved by Nesbit[5] and Leadbetter and Clark[6] became a popular alternative to closure, but ascending infection, hyperchloremic acidosis, and imperfect rectal continence prevented it from being a perfect solution. Adenocarcinoma at the ureterocolic anastomosis has also dampened enthusiasm for its use. The Boyce-Vest procedure,[7] Heitz-Boyer-Hovalacque procedure,[8] ileal conduit,[9] colon conduit,[10] and colocolostomy with internal diversion have all been used to treat incontinence when bladder closure was impossible or failed.

External and internal diversion may still be required for classical exstrophy when the bladder is represented by a fibrous patch and other complicating anomalies prevent closure or subsequent growth of the bladder. The ideal reconstruction is directed to accomplish closure of the abdominal wall and bladder with subsequent correction of reflux, bladder neck revision for continence, and anatomical and functional repair of the epispadias.

Young[11] described the first female patient with urinary continence following repair for exstrophy in 1942 at The Johns Hopkins Hospital. Failure to correct reflux, recurrent infection, and intermittent incontinence marred what had seemed at first a great success. Dees,[12] in 1949, described a method of reconstructing the bladder neck in the related condition of complete epispadias with incontinence. These early contributions might be said to have stimulated others in the field of surgery to renew efforts toward successful reconstruction in this condition.

Contributions to the management of exstrophy reads like a list of prominent urologists, pediatric urologists, and pediatric surgeons who have practiced during the last 35 years. The names of Williams and Keaton,[13] Shultz,[14] Cendron,[15] Marshall and Muecke,[16] Lattimer et al,[17] Culp,[18] Ansell,[19] Johnston and Kogan,[20] and Chisholm[21] are among early contributors to reconstruction and the understanding of exstrophy.

The Johns Hopkins Hospital has had special interest in the exstrophy problem during the past 15 years, with publications addressing many facets of this problem. These include factors in successful closure,[22] cause of wound dehiscence,[23] muscarinic cholinergic receptors,[24] inheritance of classical exstrophy,[25] incidence of malignancy,[26] urodynamic assessment during bladder neck plasty,[27] augmentations in failed closure,[28] and the effect of preliminary epispadias repair on bladder volume.[29]

Contributions from many centers have addressed epispadias repair,[30,31] urodynamics in exstrophy,[32] factors in success,[33] staged closure using colon,[34] and the use of artificial sphincter in exstrophy.[35]

The great interest, the diversity of opinion, and the variety of techniques attest to the difficulty of reaching an ideal solution in every patient. In general, the advances that have led to improved success include (1) early closure with better preservation of bladder epithelium and underlying muscle, (2) technical improvement in initial closure including penile lengthening and osteotomy when necessary, (3) management of reflux after initial closure and at the time of bladder neck reconstruction, (4) staged repair allowing for bladder growth and increased volume before bladder neck plasty for incontinence, (5) Leadbetter[36] trigonal tubularization after ureteral relocation, (6) preliminary epispadias repair to encourage bladder adaptation at low pressure to increase volume,[29] and (7) adjuncts to the surgical plan and technique.[37]

This latter category cannot be minimized when one considers that antibiotics permit the contaminated bladder to be closed as a sterile wound. Monofilament and polyglycolic acid sutures have reduced reaction in wounds and helped to prevent foci of infection and wound reaction to gut sutures. Anticholinergics, sedatives, analgesics, and im-

Accepted for publication June 2, 1989.

From the Division of Pediatric Urology, James Buchanan Brady Urological Institute, The Johns Hopkins Hospital, Baltimore, Md.

Reprint requests to Division of Pediatric Urology, The Johns Hopkins Hospital, Baltimore, MD 21205 (Dr Jeffs).

proved drainage techniques have kept the bladder and ureter at rest while healing occurred. Immobilization by traction or external fixation is important to maintain approximation of the anterior pelvic and fascial closure.

STAGED APPROACH

Not all patients present at birth with large bladders and an adequate phallus. The Table presents the options suggested by various ages and stages of presentation. Provision is made in the table both for the good bladder leading to successful closure and for the small bladder or others that fail to allow completion of the ideal plan.

THE ANOMALY

"In classical exstrophy no structure would seem missing," said Muecke in 1964. The defect in this anomaly (Figure) can be considered a midline separation with foreshortening of the distance between umbilicus and anal opening, with varying degrees of underdevelopment of bladder, urethra, and phallus. Classical exstrophy is the most common condition in a spectrum, ranging from penile epispadias to cloacal exstrophy. Classical exstrophy occurs in approximately 1 in 30 000 births. Shapiro et al[25] estimated the risk of recurrence in siblings as 1 in 300 and in offspring of patients with exstrophy as 1 in 70. Associated problems frequently seen in exstrophy are diastasis of the pubis, vesicoureteral reflux after closure, inguinal hernia, anterior displacement of the anus and sphincter mechanism, and short urethral groove. Gonadal and ductal abnormalities in both male and female patients do not appear to be part of the complex.

OSTEOTOMY

The newborn pelvis can be molded to allow midline closure of pubis and rectus fascia. This closure is sometimes difficult to hold, as the defect may be so wide that closure is difficult. Consideration should be given to each case to determine the need for osteotomy. Shultz[14] described the posterior iliac osteotomy, but more recently Sponseller et al[38] demonstrated the effectiveness of anterior transverse innominate osteotomy in promoting apposition of either initial closure or reclosure after partial or incomplete dehiscence. Firm anterior closure of the intersymphyseal bar provides support and stability for the subsequent bladder neck plasty and may improve the efficacy of puborectalis and levator ani muscles.

Management Plan for Bladder Exstrophy

Age	Problem	Possible Solution
	Initial Presentation of Patients With Bladder Exstrophy	
0-72 h	Classic exstrophy with capacity and moderate symphyseal separation; long urethral groove; mild dorsal chordee	I. Midline closure of bladder, fascia, and symphysis to level of posterior urethra; no osteotomy
0-72 h	The above findings with short urethra and severe chordee	II. Close as above, adding lengthening of dorsal urethral groove by paraexstrophy skin
0-72 h or late presentation	The above with very wide separation of symphysis or late presentation of patient (beyond 72 h to 1-3 y) for initial treatment	Iliac osteotomy and closure as in I or II
0-2 wk	Male penis duplex or extremely short	Consider female sex of rearing and closure as in I or II
0-2 wk	Very small nondistensible bladder patch	Prove by examination under anesthesia then nonsurgical expectant treatment awaiting internal or external diversion; experimental early augmentation by bowel and other material has been tried
	The Incontinent Period After Initial Closure	
1 mo to 3 y	Infection with residual due to outlet stenosis	Urethral dilatation; occasional meatotomy or bladder neck revision
	Infection, grade III reflux with pliable outlet resistance	Continuous antibiotic suppression with plan for early ureteroneocystostomy
	Partial dehiscence at bladder neck or partial prolapse of bladder (both prevent bladder capacity increase)	Reclosure of bladder neck with or without osteotomy
	Continence and Epispadias Repair	
2½-5 y	Closed bladder with incontinence; normal intravenous pyelogram; bladder capacity of 60 mL or more under general anesthesia; bilateral reflux; good penile size and length of urethral groove	Plan bilateral ureteroneocystostomy; bladder neck reconstruction, epispadias repair followed by 3 wk suprapubic drainage; prepare 5 wk and 2 wk before surgery with two 2-mg/kg injections of testosterone in oil
2½-5 y	As above with small bladder capacity (<60 mL)	Epispadias repair only after preparation with testosterone; bladder capacity will improve postoperatively

Continued on p 1477.

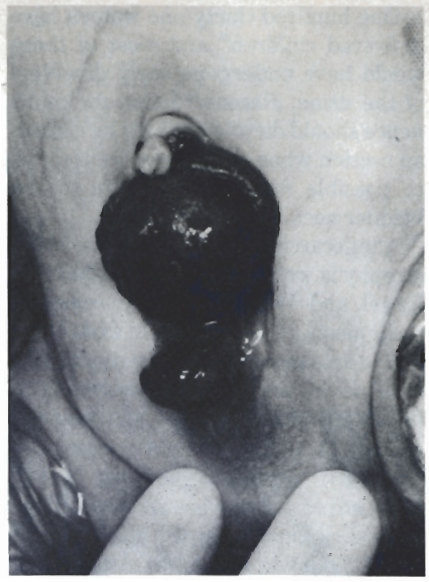
RECONSTRUCTION OF THE BLADDER NECK

The initial stage of exstrophy management involves closure of bladder without attempts to produce continence. In the subsequent interval, free drainage aids in preventing upper tract infection and minimizes the problems from reflux. The uninfected bladder can then adapt at low pressure to urine that may accumulate at rest. The adaptation process may be encouraged by preliminary epispadias repair at 2 years of age. A capacity of 60 mL as judged under general anesthesia is desirable before proceeding with bladder neck plasty. The procedure[39] is a modification of the Leadbetter procedure using the Cohen[40] antireflux operation to reimplant and relocate the ureters, followed by tubularization of the trigone and suspension of the bladder neck. Urinary diversion and minimization of bladder spasms is required to allow firm healing of the repair. Voiding trials are begun after 3 weeks of diversion. Voluntary detrusor contractions may not exist at first, gentle dilation by catheter may be required, and weeks or months may be needed to develop a useful dry interval and nocturnal control.

RESULTS

First-stage closure of the exstrophied bladder is not always successful as seen in the review of 20 patients treated for wound dehiscence after primary closure.[23] Although reclosure is possible, subsequent result of reconstruction may be disappointing. Primary closure was selected in 28 of 29 consecutive patients referred to The Johns Hopkins

Management Plan for Bladder Exstrophy (cont)		
Age	**Problem**	**Possible Solution**
	Epispadias repaired; bladder capacity greater than 60 mL	Proceed to bladder neck plasty and ureteroneocystostomy
	Epispadiac penis short with severe chordee before or after bladder neck reconstruction	Correction of chordee, lengthening of urethral groove, and epispadias repair; prepare with testosterone; consider osteotomy to aid in achieving penile length
3+ y	Complete repair of bladder, bladder neck, and epispadias with dry interval but wet pants	Patience, biofeedback, oxybutynin hydrochloride (Ditropan), imipramine hydrochloride, and time
	Above, with marked stress incontinence	Wait; may require bladder neck revision or injection
	Small-capacity bladder unchanged by time, epispadias repair, or attempted bladder neck reconstruction	Consider augmentation cystoplasty and bladder neck reconstruction; acceptance of intermittent catheterization will be necessary
3-7 y	Small bladder unsuitable for bladder neck plasty	Consider temporary diversion by colon conduit with plan for continent undiversion to bladder using bladder to form urethra and conduit for augmentation; in patients over 7 y, artificial sphincter can be considered
3-7 y	Small closed exstrophy unsuitable for bladder neck reconstruction or augmentation, or late presentation of untreated exstrophy, unsuitable for closure	Consider permanent external or internal diversion or continent urinary diversion, internal diversion direct by ureterosigmoidostomy or indirect colocolostomy; evaluate day continence of anal sphincter and nighttime seepage prior to internal diversion
5-15 y	Closed exstrophy with epispadias repaired with uncontrolled stress or dribbling incontinence	Consider revision, injection, augmentation and revision, continent urinary diversion, and artificial sphincter with omental wrap
10-20 y	Closed or diverted exstrophy with inadequate penis	Consider penile lengthening, urethral reconstruction with augmentation using free graft, pedicle grafts, and tissue transfer



Male neonate with bladder exstrophy. The umbilicus, exstrophied bladder, and complete epispadias associated with this condition are shown.

Hospital without prior treatment. One patient underwent urinary diversion because of a very small bladder. Mild bladder prolapse occurred in 2 patients who required distal revision of the closure. The remainder had satisfactory initial closure.[41]

Marshall and Muecke[16] in 1970 reviewed 329 functional bladder closures reported in the literature between 1906 and 1966 and determined that urinary continence with preservation of renal function was achieved in only 16 (5%) of the cases.

When staged reconstruction is used similar to what is described above, better continence results are obtained as reported by Chisholm[21] (45% continent), Ansell[19] (43% continent), and Mollard[42] (69% continent).

Oesterling and Jeffs,[43] in review of the The Johns Hopkins Hospital experience of 144 patients seen between 1975 and 1985, selected 50 patients who had completed staged reconstruction. Two groups of patients were compared: group A (39 patients) had had successful initial closure while group B (11 patients) had required reclosure of the bladder after partial or complete dehiscence. Group A patients had a continence rate of 92%, while in group B only 55% became continent.

In follow-up, 88% of these groups had normal upper tracts, while 6% had persistent reflux requiring additional treatment, and 6% (3 patients) required additional treatment for hydronephrosis.

When continence is not achieved in patients with adequate bladder capacity, additional repeated bladder neck plasty may be performed or injection of polytetrafluoroethylene (Teflon) or collagen can be considered.

THE FAILED RECONSTRUCTION

When continence is not achieved, when the bladder remains too small to proceed with bladder neck revision, or when hydroureteronephrosis develops, techniques of salvage are well established. Twelve such patients treated at The Johns Hopkins Hospital are described by Gearhart and Jeffs.[28] All had augmentation cystoplasty and used intermittent catheterization; in addition, repeated bladder neck plasty in four patients, artificial sphincter in three, and Mitrofanoff procedure in one were required. Artificial sphincter is usually reserved for the older patient where omentum can be used to protect the urethra from the sphincter cuff.

LONG-TERM FOLLOW-UP

Newer techniques for correction of chordee and repair of epispadias can achieve reasonable penile length and erection. Details of epispadias repair will not be discussed in this presentation. However, all patients followed up in our local experience have erections and older patients have a normal libido. Penile length and chordee correction are problems in some patients. Fertility in the male patient has been documented by sperm count[44] and in the report of Shapiro et al,[25] 38 men had fathered children. Possible injury to the ejaculatory ducts and retrograde ejaculation are a concern in the individual patient who has undergone reconstruction.

One hundred thirty-one women have delivered children[25] and most of these would have undergone early diversion of the urine. Reconstruction for continence should dictate delivery by cesarean section when pregnancy occurs, due to possible danger of damage to the bladder neck.

Adenocarcinoma[45] in untreated exstrophy is well known in older patients. Engel and Wilkinson[26] have reviewed malignancy of the bladder in exstrophy, and to date there are three patients who have developed rhabdomyosarcoma in the exstrophied bladder and two patients who underwent late closure have developed squamous cell carcinoma, while no patient who has undergone early closure has developed epithelial tumor.[46] However, long-term surveillance is mandatory in these patients.

When urinary diversion is chosen, malignancy of the bowel wall is a concern; in ureterosigmoidostomy, 10% of patients after 10 years may develop adenocarcinoma.[47]

Social adjustment in a series of older patients with exstrophy reviewed by Lattimer et al[17] was considered good in 13 of 17 patients. Woodhouse et al[48] found 55 of 64 patients gainfully employed and well adjusted. The recent series from The Johns Hopkins Hospital indicates that patients who underwent reconstructed exstrophy appear to have normal interactions at home and at school. Of course, long-term follow-up is required.

CONCLUSION

Early successful closure, careful management of the incontinent period, and a good technical bladder neck plasty can result in a patient with normal control and normal upper tracts. The small bladder removed from external irritation will increase in size and can be helped by preliminary epispadias repair if needed. Bladder neck reconstruction should not be attempted until the bladder reaches a capacity of 60 mL. Epispadias repair has many variations and these have not been detailed here. Lengthening of the urethral groove is often required and stimulation with testosterone is helpful in preparation for penile surgery.[49]

Augmentation,[28] intermittent catheterization, Mitrofanoff procedure,[50] and continent diversion[51] are salvage techniques that provide capacity for the small or unusual bladder and for those whose reconstruction is unsuccessful.

This report was submitted in honor of The Johns Hopkins Hospital 1889 to 1989 centennial.

References

1. Trendelenburg R. The treatment of ectopia vesicae. *Ann Surg.* 1906;44:281-284.
2. Burns JE. A new operation for exstrophy of the bladder. *JAMA.* 1924;82:1587-1598.
3 Magdl K. Uber die Radikaltherapie der blasen Ectopie. *Wien Med Wochenschr.* 1894;45:1113-1117.
4. Coffey RC. Transplantation of the ureter into the large intestine in the absence of a functioning bladder. *Surg Gynecol Obstet.* 1921;32:383-389.
5. Nesbit RM. Ureterosigmoid anastomosis by direct eliptical connection: a preliminary report. *J Urol.* 1949;61:728-734.
6. Leadbetter WF, Clark BG. Consideration of problems incident to performance of ureteroenterostomy: report of a technique. *J Urol.* 1950;73:62-82.
7. Boyce WH, Vest SA. A new concept concerning treatment of exstrophy of the bladder. *J Urol.* 1952;67:503-517.
8. Taccinoli M, Laurenti C, Racheli T. Sixteen years' experience with the Heitz-Boyer-Hovalacque procedure for exstrophy of the bladder. *Br J Urol.* 1977;49:385-390.
9. Jeffs RD, Schwartz GR. Ileal conduit urinary diversion in children: computer analysis follow-up from 2 to 16 years. *J Urol.* 1975;114:285-288.
10. Hendren WH. Exstrophy of the bladder: an alternative method of management. *J Urol.* 1976;115:195-198.
11. Young HH. Exstrophy of the bladder: the first case in which a normal bladder and urinary control have been obtained by plastic operation. *Surg Gynecol Obstet.* 1942;74:729-733.
12. Dees JE. Congenital epispadias with incontinence. *J Urol.* 1949;62:513-522.
13. Williams DI, Keaton J. Vesical exstrophy: 20 years' experience. *Br J Surg.* 1973;60:203-207.
14. Schultz WG. Plastic repair of exstrophy of the bladder combined with bilateral osteotomy of the ilia. *J Urol.* 1958;79:453-458.
15. Cendron J. Treatment of bladder exstrophy. *Ann Chir Infant.* 1982;12:974-979.
16. Marshall VF, Muecke EC. Functional closure of typical exstrophy of the bladder. *J Urol.* 1970;104:205-208.
17. Lattimer JK, Beck L, Yeaw S, et al. Long-term follow-up after exstrophy closure: late improvement and good quality of life. *J Urol.* 1978;119:664-666.
18. Culp DA. The histology of the exstrophied bladder. *J Urol.* 1964;91:538-548.
19. Ansell JS. Surgical treatment of exstrophy of the bladder with emphasis on neonatal primary closure: personal experience with 28 consecutive cases treated at the University of Washington Hospitals from 1962 to 1978: techniques and results. *J Urol.* 1979;121:650-653.
20. Johnston H, Kogan SJ. Exstrophic anomalies and their surgical reconstruction. *Curr Probl Surg.* 1974;9:1-39.
21. Chisholm TC. Exstrophy of the urinary bladder. In: Kiesewetter WB, ed. *Long-term Follow Up in Congenital Anomalies.* Pittsburgh, Pa: Pittsburgh Children's Hospital; 1979;6:31-35.
22. Jeffs RD, Guice SL, Oesch I. The factors in successful exstrophy closure. *J Urol.* 1982;127:974-976.
23. Lowe FC, Jeffs RD. Wound dehiscence in bladder exstrophy: an examination of the etiologies and factors for initial failure and subsequent closure. *J Urol.* 1983;130:312-315.
24. Shapiro E, Jeffs RD, Gearhart JP, Lepor H. Muscarinic cholinergic receptors in closed exstrophied bladders. *J Urol.* 1985;134:308-310.
25. Shapiro E, Lepor H, Jeffs RD. The inheritance of classic bladder exstrophy. *J Urol.* 1984;132:308-311.
26. Engel RM, Wilkinson HA. Bladder exstrophy. *J Urol.* 1970;104:699-704.
27. Gearhart JP, Williams KA, Jeffs RD. Intraoperative urethral pressure profilometry as an adjunct to bladder neck reconstruction. *J Urol.* 1986;136:1055-1056.
28. Gearhart JP, Jeffs RD. Augmentation cystoplasty in the failed exstrophy reconstruction. *J Urol.* 1988;129:790-792.
29. Gearhart JP, Jeffs RD. Bladder exstrophy: the small bladder after closure. *J Urol.* 1989;142:525-562.
30. Johnston JH. The genital aspects of exstrophy. *J Urol.* 1975;113:701-705.
31. Duckett JW. Use of paraexstrophy skin pedicle grafts for exstrophy and epispadias repair. *Birth Defects.* 1977;13:171-178.
32. Toguri AG, Churchill BN, Schillinger JF, Jeffs RD. Gas cystometry in cases of continent bladder exstrophy. *J Urol.* 1978;119:536-537.
33. Husmann DA, McLorie GA, Churchill, BN. Closure of the exstrophic bladder: an evaluation of the factors leading to its success and its importance on urinary continence. *J Urol.* 1989;142:522-524.
34. Rapp S, Giron A, Degoe JN. Complete reconstruction of bladder exstrophy. *Urology.* 1976;7:413-416.
35. Light JK, Engelmann UH. Reconstruction of the lower urinary tract: observations on bowel dynamics and the artificial urinary sphincter. *J Urol.* 1985;133:594-597.
36. Leadbetter GW Jr. Surgical correction of total urinary incontinence. *J Urol.* 1964;91:261-266.
37. Gearhart JP, Jeffs RD. Complications of exstrophy and epispadias. In: Smith RB, Ehrlich RM, eds. *Complications in Urologic Surgery.* Philadelphia, Pa: WB Saunders Co; 1989.
38. Sponseller PC, Gearhart JP, Jeffs RD. Anterior iliac osteotomy in the exstrophy patient. Presented as a poster exhibit at the American Urological Association; June 10, 1988; Boston, Mass.
39. Jeffs RD, Lepor H. Management of the exstrophy-epispadias complex. In: Walsh PC, Gittes RF, Perlmutter AD, Stamey TA, eds. *Campbell's Textbook of Urology.* Philadelphia, Pa: WB Saunders Co; 1986:1882-1919.
40. Cohen SJ. Ureterozysloneostomie: ein neue antireflux Technik. *Aktuel Urol.* 1975;6:24-27.
41. Lepor H, Jeffs RD. Primary bladder closure and bladder neck reconstruction in classical bladder exstrophy. *J Urol.* 1983;130:1142-1145.
42. Mollard P. Bladder reconstruction in exstrophy. *J Urol.* 1980;124:525-529.
43. Oesterling JE, Jeffs RD. The importance of a successful initial bladder closure in the surgical treatment of classical bladder exstrophy: analysis of 144 patients with bladder exstrophy treated at The Johns Hopkins Hospital from 1975 to 1985. *J Urol.* 1987;137:258-262.
44. Hanna MK, Williams DI. Genital function in males with vesical exstrophy and epispadias. *Br J Urol.* 1972;44:169-174.
45. Kandzari SJ, Majid A, Ortega AM, Milam DF. Exstrophy of the urinary bladder complicated by adenocarcinoma. *Urology.* 1974;3:496-498.
46. Smerdjian HS, Texter JH, Yawn DH. Rhabdomyosarcoma occurring in repaired exstrophic bladder: case report. *J Urol.* 1972;108:354-356.
47. Spence HM, Hoffman WW, Fausmyer PP. Tumors of the colon as a later complication of a ureterosigmoidostomy for exstrophy of the bladder. *Br J Urol.* 1979;51:466-470.
48. Woodhouse CR, Ransley PC, Williams DI. The exstrophy patient in adult life. *Br J Urol.* 1983;55:632-635.
49. Gearhart JP, Jeffs RD. The use of parenteral testosterone in genital reconstructive surgery. *J Urol.* 1987;138:1077-1078.
50. Mitrofanoff P. Cystostomie continente trans-appendiculaire dans le traitment: des vessies neurologiques. *Chir Pediatr.* 1980;21:297-302.
51. Rowland RG, Mitchell ME, Bihrle R. The cecoileal continent urinary reservoir. *World J Urol.* 1985;3:185-190.