

Original article

İ. Ulman
O. Ergün
A. Avanoğlu
A. Gökdemir

The Place of Mitrofanoff-Neourethra in the Repair of Exstrophy-Epispadias Complex

Department of Pediatric Surgery, Division of Pediatric Urology, Ege University, Faculty of Medicine, İzmir, Turkey

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Summary

The authors present their experience with seven patients having exstrophy-epispadias complex who had undergone *Young-Dees-Leadbetter* bladder neck reconstruction with an adjuvant *Mitrofanoff* neourethra. All the patients, but one, were male with a mean age of 7.4 ± 3.9 years. Six of them had had previous primary closure of their exstrophied bladders accompanied by anterior diagonal iliac osteostomies, and one having pure epispadias had an incompetent bladder neck and a very low bladder capacity. Four of these patients had breakdown of their reconstructed bladder necks due to clean intermittent catheterization (CIC) via urethra. Ileocystoplasties were performed to all of the patients for their unacceptably low bladder capacities (Mean 20.9 ± 12.9 ml) with an adjuvant *Mitrofanoff* neourethra. The patients were

put on CIC every 3–4 hours via *Mitrofanoff* channel. Six were totally continent, and one had to use a pad during daytime. There were no complications related to the *Mitrofanoff* stoma except two patients complained about temporary difficulty on catheterization. The authors conclude that incorporating a *Mitrofanoff* stoma to bladder neck reconstruction procedures in exstrophy-epispadias complex, prevents injury to the bladder neck during CIC, thus lowers the risk and incidence of failure.

Key words: Bladder exstrophy – Epispadias – Bladder neck reconstruction – Appendix – Appendicovesicostomy – Clean intermittent catheterization – Continence

Résumé

Les auteurs présentent leur expérience au sujet de 7 patients porteurs d'exstrophie-épispadias chez qui une reconstruction du col vésical par *Young-Dees-Leadbetter* a été pratiquée avec un néo-urètre selon *Mitrofanoff*. Tous les patients, sauf un, étaient de sexe masculin avec un âge moyen de $7,4 \pm 3,9$ ans. Six d'entre eux ont eu une fermeture primitive antérieure de leur extrophie vésicale accompagnée par des ostéotomies iliaques diagonales antérieures et un, porteur d'un épispadias pur, était porteur d'un col vésical incompetent et d'une vessie de très basse capacité. Quatre de ces patients ont présenté une désunion du col vésical reconstruit à cause d'un sondage intermittente par l'urètre (CIC). Des iléocystoplasties ont été pratiquées chez tous les patients atteints d'une vessie de capacité très basse inacceptable (moyenne $20,9 \pm 12,9$ ml) avec un néo-urètre accessoire de *Mitrofanoff*. Les patients durent subir un drainage intermittente chaque 3 à 4 heures par le canal de *Mitrofanoff*. Six ont été complètement continents et un devait utiliser un tampon pendant la journée. Il n'y a pas eu de complications en relation avec l'abouchement de *Mitrofanoff* excepté chez deux patients qui avait des difficultés de cathétérisme. Les auteurs concluent que l'utilisation d'un abouchement de type *Mitrofanoff* au cours des reconstructions du col vésical dans l'exstrophie-épispadias et susceptible d'éviter des lésions du col vésical pendant CIC de sorte que le risque d'échec se trouve diminué.

Mots-clés: Extrophie vésicale – Epispadias – Reconstruction du col vésical – Appendice – Appendicovesicostomie – Sondage intermittent – Continence

Zusammenfassung

Die Autoren stellen ihre Erfahrungen mit 7 Patienten mit einer Blasenexstrophie oder einem Epispadie-Komplex vor, bei denen eine *Young-Dees-Leadbetter*-Blasenhalsrekonstruktion mit einer Neourethra nach *Mitrofanoff* durchgeführt worden war. Alle bis auf ein Kind waren männlich bei einem Alter von 7,4 bis 3,9 Jahren. Bei 6 war ein primärer Verschluss der Blasenexstrophie mit gleichzeitiger anteriorer, diagonaler Osteotomie der Iliacae durchgeführt worden. 1 Kind hatte eine Epispadie mit inkompetentem Blasenhalss und sehr kleiner Blasenkapazität. Bei 4 der rekonstruierten Blasen kam es zu einer Nahtruptur im Rahmen der postoperativen, transurethralen, intermittierenden Katheterisierung.

Bei allen Patienten wurde aufgrund der geringen Blasenkapazität (Durchschnitt $20,9 \pm 12,9$ ml) eine Ileozystoplastik vorgenommen. Gleichzeitig wurde ein *Mitrofanoff*-Stoma angelegt. Durch dieses Stoma wurde alle 3–4 Stunden eine intermittierende Katheterisierung vorgenommen. Sechs Kinder sind jetzt vollständig kontinent, 1 Patient benutzt tagsüber eine Abdeckung zum Schutz. Nur 2 Patienten klagten über vorübergehende Schwierigkeiten beim Katheterisieren, sonst waren keine Komplikationen bzgl. des *Mitrofanoff*-Stomas zu beobachten.

Die Autoren schließen hieraus, daß das *Mitrofanoff*-Stoma bereits bei der Rekonstruktion des Blasenhalsses angelegt werden sollte, um Verletzungen des Blasenhalsses während des intermittierenden Katheterisierens zu verhindern.

Schlüsselwörter: Blasenexstrophie – Epispadie – Blasenhalss – *Mitrofanoff*-Stoma – Appendicovesicostomie – Intermittierendes Katheterisieren – Urinkontinenz

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Introduction

Achieving a capacitant and continent lower urinary tract in patients with exstrophy-epispadias complex has been a surgical challenge. As an adjunct to bladder neck reconstruction, repairing of the lower urinary tract in this anomaly sometimes requires bladder augmentation for a sufficient reservoir capacity and acceptable continence. A competent bladder neck can be achieved by one of the previously described well-known procedures, *Young-Dees-Leadbetter* as the most commonly performed. However the current status, as spontaneous voiding without residual the ultimate goal, is still far from being satisfactory, resulting either in continence without spontaneous voiding, or voiding with more or less incontinence. The more realistic management protocols in the recent years tend to draw back from spontaneous voiding, at least temporarily, in favor of continence (2). This has been mostly possible by the placement of an adjuvant *Mitrofanoff* neourethra in patients undergoing bladder neck reconstruction (9). This approach, by allowing an alternative channel for intermittent catheterization, might avoid the surgeon having to compromise the bladder neck reconstruction in order to achieve length and narrow diameter, and also prevent injury to the neourethra during catheterization especially in the early postoperative period. This study is a review of our experience regarding the placement of an adjuvant *Mitrofanoff* neourethra in bladder exstrophy patients undergoing *Young-Dees-Leadbetter* bladder neck reconstruction.

Materials and methods

Six children with exstrophy-epispadias complex and one with incontinent epispadias were subjected to *Mitrofanoff* procedure in this department between 1994-1996. The mean age of the patients was 7.4 ± 3.9 , with all, but one, male. All the patients had had primary closure of their exstrophied bladders with anterior diagonal iliac osteotomies, four had had previous failed bladder neck reconstruction procedures (*Young-Dees-Leadbetter*). Ileocystoplasty was performed on all of the patients due to their unacceptably low bladder capacities (mean capacity 20.9 ± 12.9 ml) following primary closure and a reasonable follow-up period. Bilateral ureteroneocystostomy (*Politano* in 2, *Cohen* in 5) was routinely performed usually at the time of bladder neck reconstruction and/or bladder augmentation. A right-sided *Mitrofanoff* channel was created using pedicled vermiform appendix in all patients using the technique described elsewhere (9). No attempt was made in any patient to close the bladder neck completely as in the original description of the *Mitrofanoff* procedure, instead, a long and narrow, but patent urethra was created over a 6-8 F stent for bladder neck reconstruction (5). The *Mitrofanoff* stoma was placed in the right lower abdominal quadrant at 2-3 cm distance from both midline and the groin. To prevent stomal stenosis, an insertion of V skin flap was used when constructing the stoma. Intermittent catheterization of the bladder via appendicocutaneousostomy was started on the 21st postoperative day.

The patients were followed monthly for the first three months, and every three months thereafter. The continence status, and any complication related to *Mitrofanoff* procedure was noted. Continence was defined as having at least a 4-hour dry interval.

Results

Follow-up ranged from 6 to 28 months (mean 18 ± 10 months). There were no complications related to the *Mitrofanoff* procedure.

Two patients described temporary difficulty in catheterization that resolved spontaneously. Patient and parent compliance and satisfaction were adequate. All the patients were on clean intermittent catheterization (CIC) every 3-4 hours, although one defined spontaneous voiding. Six patients were totally continent, and one required a pad during daytime. This single patient also had moderate stoma leakage which might necessitate refashioning of the conduit in future. The ultimate bladder capacities of the patients were not less than 250 ml (mean 332 ± 72 ml).

Discussion

Clean intermittent catheterization has been one of the most significant contributions to the management of bladder exstrophy, following its initial description in neurogenic bladder patients (3, 4, 6). The use of catheterizable stomas further developed the continence parameters for exstrophy cases. The *Mitrofanoff* procedure was first described in 1980 as an alternative access to urinary reservoir (5). Although it was originally used together with complete closure of the bladder neck, it is currently offered as an alternative to urethral catheterization for patients with neurovesical dysfunction, or reconstructed bladder neck in bladder exstrophy (1, 7, 10). Closure of the bladder neck is not preferred today, because it is of vital importance acting as a pop-off valve preventing the perforation of the augmented bladder in case of unexpected pressure increase. Closure of the bladder also appears to increase the risk of stone formation, particularly with bladder augmentations incorporating bowel mucosa (1).

Unaided spontaneous voiding has been the ultimate goal of surgical treatment in exstrophy patients. However, apart from sporadic reports, the incidence of spontaneously voiding and continent patients following surgical repair is still unsatisfactory. Criticizing the definition of continence as dry intervals less than 4 hours, some authors advocate creating a bladder neck that would not leak at the expense of spontaneous voiding (2). This approach unsurprisingly gives excellent continence but low voiding ability rates. What obtained with this type of management gives immediate parent satisfaction. This is observed clearly in this study. However, we think and stress that, the expense of spontaneous voiding for continence should not lead to the expense of the safety valve or pop-off mechanism necessitating a patent urethra. Techniques depending on flap valve mechanism over the intravesical urethra are strictly dependent on CIC intervals which can easily be shorter following excessive fluid intake. An inadvertently performed very narrow or strictured *Young-Dees-Leadbetter* type bladder neck behaves also in that manner. So, examining the urethra to verify its patency is essential in the first 12 to 18 months following bladder neck reconstruction if the patient is continent and using the *Mitrofanoff* stoma for CIC. This can be accomplished during urodynamic studies, or rarely by urethro-cystoscopy.

The indications put forward by the authors that combine appendicovesicostomy with bladder neck reconstruction have been; the facilitation of catheterization in wheelchairbound and obese patients, and difficulty in catheterization per urethram observed frequently following successful bladder neck reconstructions. Four patients in this series had experienced satisfactory continence in the early postoperative period. However, they developed incontinence within a few weeks after they started CIC per urethram. Cystoscopy revealed breakdown of the bladder neck sutures due most likely to catheterization trauma. This finding led us to extend the indication of appendicovesicostomy. Thus, it

protected the neourethra from catheter injury in the postoperative period by offering an alternative channel for catheterization. The urethras of all patients are patent, but the patients are so much adapted to a *Mitrofanoff* stoma that the families, even the oldest patient's, do not prefer urethral catheterization and seem to enjoy continence with appendicovesicostomy.

The results of the present study led us to make some changes in our policy regarding continence in exstrophy-epispadias complex. We believe that; the first aim in the management of patients with bladder exstrophy-epispadias complex should be to achieve urinary continence with the ability to void spontaneously without residual. This can be obtained, although not very frequently, with *Young-Dees-Leadbetter* bladder neck reconstruction procedure. Since the success is unpredictable, every patient deserves it as the operation of choice. If it fails, or intermittent catheterization becomes a necessity, a *Mitrofanoff* stoma added to the first procedure later on or combined with a second bladder neck reconstruction will serve as an alternative way of catheterization for a safe period for the neourethra to heal. The family preference should determine the fate of the stoma in the future.

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ibrahim Ulman, M.D.

Department of Pediatric Surgery
Ege University
Faculty of Medicine
35100 İzmir
Turkey

EKSTROFİ-EPİSPADİAS KOMPLEKSİNİN ONARIMINDA MITROFANOFF NEOÜRETRASININ YERİ

Yazarlar ekstrofi-epispadias kompleksi nedeniyle Young-Dees Lead-Better mesane boynu rekonstrüksiyonu ve adjuvan Mitrofanoff prosedürü uygulanan 7 hasta ile ilgili tecrübelerini sunmaktadırlar. Bir hasta dışında tüm hastalar erkek olup ortalama yaş $7,4 \pm 3,9$ 'dur. Altı hasta ekstrofi vezika hastası olup daha önceden ekstrofinin primer kapatılması ve anterior diagonal iliak osteotomi uygulanmıştı ve bir hastada pür epispadias, inkompetan mesane boynu ve çok düşük mesane kapasitesi mevcuttu. Dört hastada daha önceden onarılan mesane boyunları üretra yoluyla uygulanan temiz aralıklı kateterizasyon (TAK) nedeniyle açılmıştı. Mesane kapasitesi çok düşük olan (ort. $20,9 \pm 12,9$ ml) tüm hastalara ileosistoplasti ve adjuvan Mitrofanoff stoması uygulanmıştır. Tüm hastalar Mitrofanoff yoluyla 3-4 satte bir TAK uygulamasına geçmişlerdir. Altı hasta tamamıyla kontinan olup 1 hasta gün içerisinde pet kullanmak zorunda kalmıştır. İki hastada geçici kateterizasyon güçlüğü dışında Mitrofanoff stomasına yönelik hiçbir komplikasyona rastlanmamıştır. Ekstrofi-epispadias kompleksinde mesane boynu rekonstrüksiyonuna ek olarak Mitrofanoff stomasının eklenmesi TAK sırasında mesane boyununun yırtılmasını ve dolayısıyla operasyonun başarısının azalmasını önlemektedir.

Pediatric Surgery

Prof. Dr. A. M. Holschneider, Kinderchirurg. Klinik des
Städt. Kinderkrankenhauses, Amsterdamer Str. 59, D-5000 Köln 60

Dr. I. Ulman
Department of Pediatric Surgery
Ege University Fac. of Medicine
35100 IZMIR
Türkei

Professor Dr. med. A. M. Holschneider
Direktor der Kinderchirurgischen Klinik
des Kinderkrankenhauses der Stadt Köln
Amsterdamer Straße 59
D-5000 Köln 60 (Rhein)
Telefon (0221) 7774-200 oder 201

Dear Doctor Ulman,

Cologne, 22th of February 1997

Thank you very much for your interesting manuscript entitled:

The place of Mitrofanoff neourethra in the repair of extrophy - epispadias complex.

We are glad to be able to confirm that your manuscript has been accepted by the editorial board and our advisers for publication in the European Journal of Pediatric Surgery. We have sent your paper to the publishers and thank you very much again for having let us see your paper.

With kind regards
Yours sincerely



(Prof. Dr. A. M. Holschneider.)

