

EFFECTS OF TOTAL UROGENITAL MOBILIZATION ON LOWER URINARY TRACT FUNCTION IN CHILDREN WITH CONGENITAL ADRENAL HYPERPLASIA.

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INTRODUCTION AND AIM OF THE STUDY

Congenital adrenal hyperplasia (CAH) is the most common cause of female pseudo-hermaphroditism and ambiguity of external genitalia. Surgical treatment is possible; aims of the surgical intervention are clitoroplasty, formation of labia minora and urogenital sinus (UGS) repair. Different techniques were described through the years to correct UGS and perform vaginoplasty. Former procedures used posterior perineal flap or vaginal cut-back. Even if simple, these techniques do not reproduce normal female anatomy but shorten the perineum, bring hair bearing skin into the vagina and preclude mucosal lining of the posterior vaginal fornix. Moreover, the position of the urethra is not modified and the urethra remains in a hypospadiac position. Surgical complications, as vaginal stenosis or urethrovaginal fistula, and lower urinary tract symptoms have been reported in adult females who underwent older feminizing procedures¹. Therefore, new techniques of urogenital sinus repair were developed. Recently, the procedure of total urogenital mobilization (TUM) used by Pena to correct cloaca malformation has been performed by Rink in patients with UGS due to CAH². In this technique, UGS is circumferentially mobilized without separation of the vagina from the sinus and brought down to perineum; the redundant tissue of the common sinus forms a mucosal lining of the vestibule. In case of long UGS, division of pubourethral ligaments anteriorly and the endopelvic fascia may be necessary to allow caudal displacement. Therefore, critical issue is raised regarding the possibility of a stress urinary incontinence after TUM. Since vaginal separation from the urethra is unnecessary and a mucosal line forms the vestibule, cosmetic results of TUM are good and vaginal stenosis and urethrovaginal fistula rare. Nevertheless, consequences of this technique on lower urinary tract function (LUTF) are poorly understood. Few data on continence status

after TUM seem to indicate that the presence of UGS longer than 3 cm is a negative prognostic factor for continence³. We prospectively followed children with CAH, who underwent TUM during infancy, in order to evaluate genital anatomy, LUTF and continence status.

MATERIALS AND METHODS

Seven patients with CAH underwent TUM in infancy (average age: 14.8 ± 12.6 months; range 8-43 months). The average length of the UGS was 1.78 ± 0.9 (range, 1-3.8) cm; UGS was longer than 3 cm in one patient. All patients were prospectively evaluated after operation. Examination under anaesthesia and cystoscopy were performed one year after the operation and at toilet training age in order to assess anatomy. Cosmetic and anatomical genital appearance was evaluated according to Creighton criteria. At toilet training age, all children underwent bladder diary and urodynamics to evaluate LUTF; urodynamic study will be planned every 2 or 3 years from the first evaluation.

RESULTS

All children have had a follow-up of at least 2 years (range, 24-59 months; average: 39.7 ± 18 months). We did not observe short- and long term complications requiring surgical revision. According to Creighton criteria, cosmetic results were good in 6/7 children and satisfactory in the other one. At toilet training age all but one patient achieved day-time and night-time continence. Bladder diary was abnormal only in one patient with UGS longer than 3 cm.

In this patient a daytime frequency lower than 4 void per day, with small voided volumes, and severe incontinence episodes were recorded. Urodynamics was normal in 4/7 patients.

Out of the 3 children with abnormal pattern, 1 had detrusor overactivity which normalized during follow-up, 1 showed dysfunctional voiding without post-voiding residual urine and the remaining had stress urinary incontinence. Stress urinary incontinence was found only in the patients with UGS longer than 3 cm.

DISCUSSION

Our series did not show surgical complications after TUM. Cosmetic appearance of external genitalia was good in the majority of patients. Clinical and urodynamic data seem to confirm that the presence of UGS longer than 3 cm is a negative prognostic factor for continence.

CONCLUSION

TUM may provide a better cosmetic appearance of external genitalia, a good vaginal opening and length, with satisfactory bladder-sphincter function in patients with UGS shorter than 3 cm. Further investigations are needed to verify if such assumption may be valid also for patients with longer UGS requiring more extensive dissection.

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PELVIC FLOOR TRAINING IN THE TREATMENT OF DYSFUNCTIONAL VOIDING IN CHILDREN WITH LUTS.

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INTRODUCTION AND AIM OF THE STUDY.

Pelvic-floor therapy seems to be a useful component in the treatment of bladder dysfunction in which urethral sphincter hyperactivity or pelvic floor contraction plays a role during the voiding phase in non neurogenic pediatric patients. To evaluate the efficacy of the outpatient urotherapy in children with LUTS, the AA consider their own experience in 45 cases of dysfunctional voiding.

Material and methods

A total of 45 patients with LUTS, 41 girls and 4 boys, aged between 5 and 15 years, were enrolled in this study. LUTS at the time of presentation are described in Tab 1. All patients were studied at first with non-invasive urodynamic approach: bladder diary, pre-post voiding renal-bladder ultrasound, uroflowmetry associated with perineal surface electromyography (FLW-EMG), three investigations at least, which documented dysfunctional voiding. The FLW-EMG patterns at the diagnosis in all pts are illustrated in Tab 2. Twenty-four patients also had H₂O Cystometry (Cys) for associated symptoms of voiding and storage phases: 14 showed lower detrusor compliance, 15 detrusor hyperactivity, 2 augmented compliance with detrusor hyperactivity and 2 were normal. Voiding cystourethrography (CUM) was performed in 26 pts for recurrent UTI, which documented RVU in 4 patients and spinning top urethra in 22 female pts. The treatment was standard urotherapy, behavioral modifications, pelvic floor training and FLW-EMG biofeedback in all patients, associated with anticholinergic therapy in 24 pts, alpha-antagonist in 2, desmopressine in 10 and antibiotic prophylaxis in 22.

The therapy protocol comprises a weekly half-an-hour session, for a period of three months with a follow up at 6, 12, 24 months.

Tab. 1- LUTS at presentation in 45 Pts

IVU ± pain	23
Urgency ± urge incontinence	26
Increased voiding frequency	25
Decreased voiding frequency	10
Enuresis	25
Giggle incontinence	2
Stipsis, encopresis	7

Tab. 2 FLW-EMG patterns at diagnosis

Increased pelvic floor activity	45
Bell shaped curve	6
Staccato flow curve	17
Interrupted flow curve	9
Tower shaped curve	10
Plateau shaped curve	3

RESULTS

The criterion to evaluate the success of the therapy was the improvement of the subjective symptoms and the patterns of bladder diary, ultrasound and FLW-EMG curves. Resolutions of TUI and RVU were also taken into account. Tab. 3 shows results related to LUTS, while Tab. 4 documents FLW-EMG patterns after therapy.

Tab. 3 - LUTS after treatment in 45 pts

IVU ± pain	3 -13%
Urgency ± urge incontinence	3 -11.5 %
Increased voiding frequency	2- 8 %
Decreased voiding frequency	1- 10%
Enuresis	3- 12%
Giggle incontinence	1
Stipsis, encopresis	2

Tab 4 - FLW-EMG patterns after treatment

Normal pelvic floor activity	39 (86,7 %)
Increased pelvic floor activity	6 (13.3 %)
Bell shaped curve	40*
Staccato flow curve	1
Interrupted flow curve	4
Tower shaped curve	0
Plateau shaped curve	0

DISCUSSION According to the data, urge incontinence, urgency and enuresis resolved in 88% of the pts while the voiding frequency improved in 90% of the cases.

Pelvic floor activity became normal in 39 pts (86,7%) while 6 did not show any improvement and remained stable with their hyperactivity. The FLW patterns improved in 87% of the cases. All pts except three resolved their TUI. RVU disappeared in three pts: one patient had an endoscopic subureteral macropastique injection for persisting unilateral RVU III, after an eight-month period of anticholinergic therapy.

CONCLUSION

In our experience the results achieved with the urotherapy, often combined with pharmacological therapy in pediatric patients with dysfunctional voiding, are encouraging. The simultaneous treatment of the filling disturbances, when present, played probably an important role.

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THE “MINIMAL APPROACH” FOR VESICoureTERAL REFLUX IN NEUROGENIC BLADDER.

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INTRODUCTION AND AIM OF THE STUDY

Neurogenic bladder (NB) overactivity can cause severe and irreversible renal damage and bladder-wall destruction years before incontinence becomes an issue. Clean intermittent catheterization (CIC) combined with anticholinergics is the standard therapy for NB. In patients not responding to medical treatment botulinum toxin injections (Botox) into the detrusor can be performed. Previous papers showed increased capacity, decreased pressure and increased bladder compliance¹; differently from what previously suspected, Botox is proven that does not induce *de novo* vesicoureteral reflux (VUR) in adult patients with idiopathic overactive bladder². In addition, simultaneous treatment with Botox on external urethral sphincter and dextranomer/Hyaluronic acid (Deflux) has been reported to improve endoscopic correction of vesico-ureteral reflux (VUR) in bladder dysfunction (BD). We report our results with simultaneous subureteral Deflux injection and Botox detrusor injections, to treat reflux in paediatric neurogenic bladder.

MATERIALS AND METHODS

Seven patients, 4 boys and 3 girls (average age: 10.8±8.1 years) with NB and VUR were considered, with a total of 12 refluxing renal units treated. All patients were under CIC and were evaluated before endoscopic treatment with videourodynamics (VUD) and renal ultrasounds (US). All patients with mental disorders and/or previous urological surgery were excluded. Inclusion criteria were: VUR grade 2°- 4° and neurogenic bladder overactivity not responding to anticholinergics. All patients were admitted to the study after a written consent was obtained. Treatment consisted of endoscopic subureteral injections of Deflux (1 ml for each ureter) and detrusor endoscopic Botox injections (10 U.I. /Kg), simultaneously. All patients were clinically followed-up every 6 months and with cystography at 6-12 months.

RESULTS

VUR disappeared in 10/12 renal units. In the remaining 2 units (one girl with previous 4° grade VUR) it decreased to 3° grade. No relapses were observed through the subsequent

follow-up of 12.7 (range: 7-30) months. Cystometric bladder capacity always increased, with an average increase of 77 ml; detrusor overactivity reduced in all cases, with an average reduction of the amplitude of the contractions of 22 cmH₂O.

DISCUSSION AND CONCLUSION

Botox has been reported to treat non responding patients with neurogenic and non-neurogenic overactive bladder³, but it has been shown that the effects of Botox are temporary (10-12 months)¹. We demonstrated the efficacy of simultaneous ureteral Deflux and bladder Botox treatment in children with NB, which could be applicable in the management of difficult cases, without adverse effects. The minimvasive approach and the easy procedure encourage to resolve reflux and to treat high bladder pressure, in a single shot.

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A LESS-INVASIVE APPROACH TO PERFORM RECTUS FASCIAL SLING IN BOYS WITH NEUROGENIC INCONTINENCE.

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INTRODUCTION AND AIM OF THE STUDY

Among children with neurogenic bladder dysfunction (NBD), the majority are treated by anticholinergics and clean intermittent catheterization (CIC), while some of them need manipulation of the bladder neck to achieve socially acceptable urinary continence. Regarding this aspect, options for increasing bladder outlet resistance include: injection of bulking agents around the bladder neck, artificial urinary sphincter implantation, bladder neck tubularization. Because absolute success rates have been elusive for most of the above procedures, the use of a rectus fascial sling is still an attractive alternative with good success in boys^{1,2}. Even if it has been proven that synthetic slings are effective also in neurogenic adult female patients, in children autologous (rectus fascial, bladder strips), or at least heterologous tissues are highly preferred, in order to prevent long-term complications, such as infection and erosion. Moreover, many patients require open surgery for bladder augmentation in conjunction with the sling procedure.

The rectus fascial sling procedure, via an abdominal-perineal approach, consists in suspending the bladder neck by placing a simple U-shaped rectus abdominous fascial strip, secured at the posterior surface of the pubic symphysis. The perineal approach is performed to prepare the passage of the sling alongside the prostate using blind digital dissection.

Potential complications of this procedure include bladder neck, urethral and rectal injury. The blunt finger-dissection around the bladder neck may be difficult in young male patients; the periprostatic neurovascular bundles may be damaged with resultant impotence, requiring a preventive lateral opening of the endopelvic fascia in order to pass the fascial sling laterally to the prostate.

Thus, to reduce possible damage due to blunt finger dissection, we modified the sling procedure, performing a mini-invasive passage of the rectus fascial strip through the perineum, by means of the use of the curved carrier devices for tension-free vaginal tape. In this paper we report the outcome of our series of rectus fascial sling in male children,

and we present technical details of rectus fascial sling procedure in boys with neurogenic incontinence.

MATERIALS AND METHODS

We reviewed the charts of 5 walking spina bifida boys (average age: 15 ± 6.8 years, range: 6-24 years), who underwent rectus fascial sling procedure for neurogenic incontinence from 2000 to 2007. Of those, the older four ones walk without or with orthotic use, while the youngest one walks with the aid of major orthoses. Clinical and videourodynamic indications were: an open bladder neck during filling at low detrusor pressure and evidence of stress urinary incontinence. Two boys had normal bladder capacity for age and normal compliance, not requiring additional bladder surgery; while the remaining 3 patients needed bladder augmentation procedures (two detrusorectomy and one sigmoido-cystoplasty). Four patients regularly used CIC and one emptied his bladder by abdominal straining. All patients were followed by clinical evaluation for continence (bladder diary and pad test), upper urinary tract ultrasound (US) and urodynamics, with mean follow-up of 2.6 (range 0.5 -7) years.

RESULTS

Two patients, aged 21 and 24 years, achieved urinary (day and night-time) continence soon after surgery; 2 boys, aged 14 and 20 years, reached only night-time continence with improvement of daytime after sling, and subsequently repeated bulking (dextranomer/Hyaluronic acid) agent injection at the site of the sling, finally achieving continence; the remaining 6-year boy, with difficult ambulation requiring orthoses, remained wet at night, and achieved only an improvement of dryness between CIC, waiting for further procedures. The age-related bladder capacity improved in all patients with a good compliance in all cases: average capacity pre- and post-surgery is 222 and 467 ml respectively. No early or late surgical complications were found and upper urinary tract remained stable in all cases.

DISCUSSION AND CONCLUSION

In our series, good results regarding the urinary continence with no complications have been obtained in those difficult to treat patients. In fact, the very poor bladder outlet resistance and the fact that those boys walk with orthotic use, make surgery for continence a real challenge. The procedure modification, using curved carrier devices may reduce possible damaging of the anatomical structures of the perineum and periprostatic tissues.

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LAPAROSCOPIC COLPOSACROPEXY IN ROKITANSKY SYNDROME

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INTRODUCTION AND AIM OF THE STUDY

Mayer-Rokitansky syndrome (MRK) consists in various degrees of vaginal atresia associated with other mullerian anomalies and normal ovarian activity.

We report a rare case of a 16-year old girl affected by MRK, presenting vaginal prolapse after sexual intercourses.

MATERIALS AND METHODS

We applied the same surgical technique used in the reconstructive phase of colposacropexy in older patients affected by hysterocele or pelvic organs prolapse: this video shows no uterine or mullerian remnants; a lateral mobilization of sigmoid colon and incision of posterior peritoneum with access to presacral space and exposition of sacral bone is performed; a cranio-caudal extension of posterior peritoneal incision with identification of right ureter is completed; an "S" plicature of prosthetic material allows anterior and posterior vaginal vault fixing to polypropylene prosthesis with prolene suture; the prosthesis is fixed to sacral bone and complete reperitoneization is performed.

RESULTS

Operating time was 50 minutes and hospitalization time was 48 hours. Vaginal prolapse was treated completely and early recovery of sexual function was obtained. No prolapse recurrence or pelvic pain was observed at one month follow-up.

DISCUSSION

The absence of uterine remnants didn't hold the possibility to achieve the restoration of menstrual function but on the other hand, in this case the anatomical characteristics allowed a rapid and safe laparoscopic treatment due to the absence of vesico-rectal

prolapse and low grade of vaginal aplasia: vesicovaginal and rectovaginal dissection were not needed. As usual, a complete re-peritoneization was performed in order to avoid intestinal adhesions, prosthesis infection or prolapse recurrence. At one month of follow-up sexual function recovery was obtained, in absence of pelvic pain.

CONCLUSION

Laparoscopic approach is a safe and mini-invasive procedure that allows early recovery of sexual function without traction devices.

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LOW AND SHORT-LIVED DETRUSOR CONTRACTILITY, DETRUSOR OVERACTIVITY AND BLADDER OUTLET OBSTRUCTION IN FEMALE PATIENTS WITH MULTIPLE SCLEROSIS.

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INTRODUCTION AND AIM OF THE STUDY

Patients with multiple sclerosis (MS) often suffer from lower urinary tract symptoms (LUTS), detrusor overactivity (DO) and elevated post-void residual urine (PVR)¹. Objective of the study was to investigate detrusor contractility in female patients with MS and in control subjects using the power factor².

MATERIALS AND METHODS

Female patients with MS and LUTS referred to our outpatient clinic from the Department of Neurology between January and June 2007 and undergoing pressure-flow study were enrolled. All studies were performed in duplicate using a 6 Ch transurethral double lumen catheter and an 8 Ch transrectal catheter. Bladder was filled at 50 ml/min rate. The examination was carried out according to ICS guidelines. Detrusor contractility was investigated using the CLIM software and power factor (W) parameters were derived. A random sample of female patients with lower urinary tract symptoms, no detrusor overactivity or bladder outlet obstruction undergoing pressure-flow study was used as control. A random sample of patients with Stress Urinary Incontinence (SIU) was also used as control.

RESULTS

Eighteen MS patients, 34 controls and 26 patients with SUI were evaluated. MS patients were characterised by a unique sensory function and voiding dynamics. Volume at first sensation, normal desire to void and cystometric capacity were

lower than in controls. Small voided volumes were produced with poor flow rates, high detrusor pressures and residual volumes.

Detrusor contractility was low in absolute terms and force decreased toward the end of the voiding phase. A significant difference was observed between urodynamic parameters measured in MS patients compared to control subjects and patients with SUI (Table I).

Table I: mean values in controls, patients with MS and SUI.

	Controls	MS	SUI	*p<
Age (yrs)	57	42	65	0.01
Volume at 1 st sensation (ml)	125	88	124	n.s.
Volume at normal desire (ml)	236	157	223	n.s.
Cystometric capacity (ml)	345	230	365	n.s.
Qmax (ml/s)	21	6.9	16.3	0.001
PdetQmax (cmH ₂ O)	24	32.5	19.6	0.001
Voided volume (ml)	387	76	307	0.001
Post void residual (ml)	0	94	20	0.001
Wmax	13.3	7.6	8.9	0.001
W20	11.7	4.8	7.8	0.001
W80	6.8	6.4	3.8	0.001
W80-20	-5.4	0.7	-4.2	0.001

*One-Way ANOVA

DISCUSSION

No information is available as to the natural history of detrusor contractility in female MS patients and its relation to LUTS. The combination of urgency with or without urge incontinence and high residual urine volumes is associated with poor and short-lived detrusor contractility as well as bladder outlet obstruction. In the absence of anatomic obstruction, the condition appears to be functional and possibly dependent upon a poorly coordinated pelvic floor. Empirical management of MS patients suggests that pelvic floor training by an experienced physiotherapist may be helpful and that the combination of alpha-blockers and antimuscarinics may be useful in patients with increased residual urine following antimuscarinics monotherapy treatment. Notwithstanding MS patients maintain a good perineal function longer than expected, the detrusor muscle shows a significant damage by the time patients refer LUTS. The

demonstrated detrusor underactivity in female MS patients, pleas for a different therapeutic approach to detrusor overactivity with drugs that do not impact on smooth muscle contractility. Further research is needed on the effect of pelvic floor rehabilitation on functional bladder outlet obstruction in female MS patients.

CONCLUSION

LUTS, and particularly OAB symptoms, in female patients with MS have a different pathophysiology from other patient categories and require a specific therapeutic approach.

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