W27: ICS Core Curriculum (Free): Children and Young Adults Workshop Committee on Transitional Care for Continence in Congenital Malformations.

Workshop Chair: Giovanni Mosiello, Italy
13 September 2017 14:30 - 16:00

Start	End	Topic	Speakers
14:30	14:35	Introduction	Giovanni Mosiello
14:35	14:50	An Overview From the Pediatric Urologist Prospective: Neurogenic Bladder, Posterior Urethral Valves, Bladder Exstrophy, Anorectal Malfromations, Guidelines	Jian Guao wen
14:50	15:05	Stoma: Indications and Concerns For Mitrofanoff and Malone	Mario De Gennaro
15:05	15:20	How to Manage Boys Operated Forposterior Urethral Valves and Severe Hypospadia	Selcuk Yucel
15:20	15:30	The Bladder Reservoir and the Outlet: Role of Bulking, Sling, Artiphicial Sphincter and Surgery for Continence	Mario De Gennaro
15:30	15:40	Surgery For Continence	Giovanni Mosiello
15:40	15:45	Bowel Management	Giovanni Mosiello
15:45	15:55	Questions	None
15:55	16:00	Take Home Message	Giovanni Mosiello

Speaker Powerpoint Slides

Please note that where authorised by the speaker all PowerPoint slides presented at the workshop will be made available after the meeting via the ICS website www.ics.org/2017/programme Please do not film or photograph the slides during the workshop as this is distracting for the speakers.

Aims of Workshop

Spina bifida, bladder exstrophy/epispadia, posterior urethral valves, hypospadia and anorectal malformations are surgically managed in childhood but all these patients require life-long urological care for the treatment of continence. There are critical aspects to define:
- Correct management in childhood to avoid procedure that impair adult life.
- Transition out of childhood, actually confusing.
- Lack of knowledge in paediatric and adult health care professionals, about adult life problems and congenital pathologies respectively.

The objectives of workshop are to offer an overview with practical suggestions for best practice management of bladder and bowel incontinence in the above pathologies.

Learning Objectives

- Correct management in childhood to avoid procedure that impair adult life aspects,
- Define common knowledges between pediatric and adult health care professionals about adult life problems and congenital pathologies respectively
- Focus on transition out of childhood: who should manage the mature pediatric urology patients?

Learning Outcomes

This workshop is defined for a multidisciplinary participation- different health care professionals involved in continence care of young adults operated for congenital malformations impairing daily life. The partecipants will receive suggestions to use in their clinical practice (university hospital, hospital, outpatient clinic,eg) in order to recognize what has been done previously, correct or not, and how to perform a correct approach to these patients based on EBM data.

Target Audience

Urologists, paediatric urologists, surgeons, pediatric surgeons, nurses, physiotherapists, paediatricians, continence advisors

Advanced/Basic

Advanced

Suggested Learning before Workshop Attendance

S. Tekgul, H.S. Dogan, E. Erdem, P. Hoebeke, R. Kocvara, J.M. Nijman, C. Radmayr, M.S. Silay, R. Stein, S. Undre. Management of neurogenic bladder in children. In Guidelines on Paediatric Urology, European Society for Paediatric Urology, European Association of Urology, chp, 3K, pp 36-41, 2015.

Nijman R., Tekgul S., Chase J., Bael A., Austin P., von Gontard. Diagnosis and management of urinary incontinence in childhood. In Incontinence., Abrams P., Cardozo L., Khoury S., Wein A., 5th ed., Ch. 9, 5th ed. pp. 729-825, 2013.

Suggested Reading

1) Consensus Review of Best Practice of Transanal Irrigation in Children.
Mosiello G, Marshall D, Rolle U, Crétolle C, Santacruz BG, Frischer J, Benninga MA.
J Pediatr Gastroenterol Nutr. 2016 Dec 13. [Epub ahead of print] PMID: 27977546
2.Button Cystostomy: Is it really a Safe and Effective Therapeutic Option in Paediatric Patients with Neurogenic Bladder?
Mosiello G, Lopes Mendes AL, Capitanucci ML, Zaccara AM, De Gennaro M.
Urology. 2016 Sep 29. pii: S0090-4295(16)30636-7. doi: 10.1016/j.urology.2016.09.025. [Epub ahead of print] PMID: 27693876
3.Prevalence of Spina Bifida Occulta and Its Relationship With Overactive Bladder in Middle-Aged and Elderly Chinese People.Wu JW, Xing YR, Wen YB, Li TF, Xie JF, Feng QD, Shang XP, Li YL, Feng JJ, Wang XX, Zhai RQ, He XF, Chen T, Liu XJ, Wen JG.Int Neurourol J. 2016 Jun;20(2):151-8
4Effects of botulinum toxin type a in the bladder wall of children with neurogenic bladder dysfunction: a comparison of histological features before and after injections. Pascali MP, Mosiello G, Boldrini R, Salsano ML, Castelli E, De Gennaro M., J Urol. 2011 Jun;185(6 Suppl):2552-7.
5 A Comparative Study of Outside-In and Inside-Out Transobturator Tape Procedures for Female Stress Urinary Incontinence: 7-Year Outcomes. Chun JY, Song M, Yoo DS, Han JY, Hong B, Choo MS.
Low Urin Tract Symptoms. 2014 Sep;6(3):145-50
6 Impact of transurethral resection on urinary flow rate in children with posterior urethral valve in short term follow-up.
Ipekci T, Akin Y, Gulmez H, Ates E, Yucel S. Saudi Med J. 2014 May;35(5):460-5.
7 Factors affecting complication rates of ureteroscopic lithotripsy in children: results of multi-institutional retrospective analysis by Pediatric Stone Disease Study Group of Turkish Pediatric Urology Society.Dogan HS, Onal B, Satar N, Aygun C, Piskin M, Tanriverdi O, Gurocak S, Gunay LM, Burgu B, Ozden E, Nazli O, Erdem E, Yucel S, Kefi A, Demirci D, Uluocak N, Aridogan IA, Turunc T, Yalcin V, Kilinc M, Horasanli K, Tan MO, Soygur T, Sarikaya S, Kilicarslan H, Turna B, Doruk HE, Tekgul S. J Urol. 2011 Sep;186(3):1035-40. doi: 10.1016/j.juro.2011.04.097.
8 Staged male urethroplasty transferring megalourethra tissue as free graft dorsal inlay to proximal urethral atresia in VACTERL association.
Bagrodia A, Yucel S, Baker LA. Urology. 2011 Dec;78(6):1417-9. doi: 10.1016/j.urology.2011.03.023.
9 Comparison of outcomes of tubularized incised plate hypospadias repair and circumcision: a questionnaire-based survey of parents and surgeon.
Snodgrass W, Ziada A, Yucel S, Gupta A. J Pediatr Urol. 2008 Aug;4(4):250-4. doi: 10.1016/j.jpurol.2007.12.007. Review
10 Transurethral injection of bulking agent for treatment of failed mid-urethral sling procedures.
Lee HN, Lee YS, Han JY, Jeong JY, Choo MS, Lee KS.
Int Urogynecol J. 2010 Dec;21(12):1479-83

An Overview From the Pediatric Urologist Prospective: Neurogenic Bladder, Posterior Urethral Valves, Bladder Exstrophy, Anorectal Malfromations, Guidelines

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Neurogenic bladder (NB), posterior urethral valves (PUV), bladder exstrophy (BE), anorectal malfromations (ARM) are most common confronted during the pediatric urological practice. Detailed urodynamic study (UDS) is vital to assess the results and to plan subsequent treatment for these entities. This paper provides an overview of these diseases from UDS prospective.

Neurogenic bladder

NB results from a variety of abnormalities of the central or peripheral nervous systems and contributes to various forms of lower urinary tract dysfunction (LUTD). In children, the spinal level and extent of congenital lesion are poorly correlated with the clinical outcome. UDS and functional classifications have therefore been more valuable for defining the extent of the pathology and planning treatment in children. The treatment protocol mainly depends upon an UDS. These urodynamic parameters include bladder capacity, the intravesical filling pressure, the bladder leakage pressure, the presence or absence of reflex detrusor activity; the competence of the internal and external sphincteric mechanisms, the degree of detrusor sphincter dysnergia (DSD), the voiding pattern and the post-voiding residual urine volume.

Posterior urethral valves

PUV is life-threatening congenital anomalies. Bilateral hydroureteronephrosis and a distended bladder during prenatal evaluation, a thick-walled bladder and a dilated posterior urethra are suspicious signs of PUV. Voiding cystourethrogram (VCUG) confirms a PUV diagnosis. A secondary reflux is observed in at least 50% of patients with PUV. Following surgical treatment, patients require close follow-up to detect and monitor for bladder dysfunction that may lead to renal injury by video UDS adds the benefit of fluoroscopy to simultaneously image the urinary system. The synchronous evaluation of structure and function

provides insight into the correlation and causation of detected anomalies. The utilization of VCUG and UDS is similar but not identical to VUDS.

Bladder exstrophy
BE is characterized by an infra-umbilical abdominal wall defect, incomplete closure of the bladder with mucosa continuous with the abdominal wall, epispadias, and alterations in the pelvic bones and muscles. Even before bladder neck reconstruction, UDS can be predictive for detrusor function and the ability of the bladder to increase in size without high intravesical pressures and also the application of anticholinergic therapy to enhance bladder volume. Following bladder neck reconstruction, urodynamic assessment provides an objective correlation with the clinical assessment of continence. It also helps in planning pharmacotherapy for elimination of uninhibited detrusor contractions, improving bladder compliance and reducing intravesical pressures.

Anorectal malformations
ARMs involves the distal anus and rectum as well as the urinary and genital tracts. Renal agenesis, vesicoureteral reflux, uninhibited detrusor contractions, poor compliance of the bladder or incomplete bladder emptying and NB are the most common urinary system malformations associated with ARM. Early and repeated UDS is mandatory to detect as earliest as possible the onset of deterioration before irreversible neurological damage has occurred. UDS are of limited value in the preoperative setting but are useful in the follow up of ongoing urological dysfunction postoperatively in patients with ARMs.

Stoma: Indications and Concerns For Mitrofanoff and Malone

Mario De Gennaro

Continent stomas gained favor in pediatric and adolescent management of urinary and fecal incontinence, as well as in cases of neuropathic bladder/bowel and of chronic obstinate constipation (obstipation). The relatively easy-to-create conduits, utilize segments of the large or small bowel, (appendix for the Mitrofanoff's and for Ace-Malone and small bowel for the Monti's stoma), which are intended to be continent and easy to access for the patient or, in case of severe limitation in dexterity, for the care-taker. Both stomas' have a common goal which is to keep the bladder ad bowel as empty as possible, to avoid damage to the kidney, to the bowel wall and to reduce the risk of urinary leak, soiling of feces and/or increase in the severity of the fecal retention. The Mitrofanoff or Monti stomas, are created, in the large majority of cases, during a bladder augmentation enterocystoplasty, or more rarely not in association with the augmentation. The catheterizable channel is positioned at the level of the groin or, at the level of the umbilicus. The latter position is preferred by teen-agers and even more so, by female patients. The Ace/Malone is usually positioned at the level of the RIF. The utilization of suprapubic catheters for intermittent emptying and of the "spring" percutaneous cecostomy, for the application of the same emptying protocol guaranteed by the Mitrofanoff and by the Ace/Malone techniques, have been considered and utilized during the last 2 decades, to avoid major open or laparoscopic surgery, but have never gained great interest by neuro-urologists or pediatric surgeons, after the initial hopes and enthusiasm. Whilst continent stomas are and remain a main tool for the management of urinary incontinence or bladder emptying especially in cases of Neuropathic Bladder, valve bladder, bladder extrophy, after bladder neck closure, in recent years the number of Ace-Malone procedures in children and adolescence has reduced, thanks to the improvements in the conservative management of fecal incontinence and obstipation or fecal impaction with oral disimpaction and rectal wash-out systems. Moreover, continent stomas' are not complication-free, (e.g. perforation, dehiscence, stenosis, infection; etc.). One of the most common complication of the continent stomas is to not be continent from the time of its creation, or soon after. Medium or long-term occurrence of urinary or fecal leak also affect the final outcome and quality of life of the patients. The use of bulking agents and ultimately, a re-do procedure have to be performed in a timely fashion, to avoid returning completely to the initial condition. Dermatitis, strictures, psychological compromise and changes of the perception of self, are at risk in patients undergoing the mitrofanoff's procedure. The recent introduction of laparoscopic urological procedures in pediatric practice has found interest in renal and vesical procedures. It is foreseen that less invasive techniques will become more and more present, in the near future, providing more satisfactory emptying of the bladder and bowel reservoirs, while guaranteeing no urine or fecal leak and an acceptable quality of life.

How To Manage Boys Operated for Posterior Urethral Valves and Severe Hypospadias

Selcuk Yucel, MD

Congenital urological diseases may affect the future adult life depending on the nature of the disease and the treatment. Posterior urethral valve and hypospadias are two major congenital urological diseases that may significantly alter the physical and psychological well-being of an adult.
Posterior urethral valve can be diagnosed with renal failure, urinary tract infections, hydronephrosis and lower urinary tract dysfunction. A proper posterior urethral valve ablation may improve the bladder cycling to prevent hydronephrosis, high residuals in urinary bladder. However, worsening the renal , bladder storage and voiding function may be due to rest urethral valve, urethral stricture, bladder neck hypertrophy, hostile bladder, aperistaltic upper urinary system, renal dysplasia and valve bladder syndrome. Control cystoscopy to check for valve and stricture presence is very critical before starting urodynamics. Identifying a hostile bladder with voiding problems is another critical step to start clean intermittent catheterization through urethra and catheterizable channels and medication/bladder surgery to decrease bladder pressure and increase bladder volume.

It is common to observe an underactive bladder with high bladder residuals in posterior urethral valve adolescents related with long term anticholinergic use or myogenic failure. Children with renal dysplasia can be prepared for renal transplantation with a very comparable long term renal function. Posterior urethral valve is a very distinctive disease requiring specific medical attention during transitional age.
Hypospadias is a disease with a relatively higher incidence compared to four decades ago. Particularly posterior hypospadias is a very challenging disease requiring surgical skills and experience. Obstructed voiding may be associated with a cosmetically very acceptable penis following hypospadias repair. However, urofowmetrics with poor sream may improve after puberty in repaired hypospadias. High bladder pressures should be suspected in poor streamers and should be followed up properly to prevent detrusor and renal problems. Improper skin flaps and grafts may also end up with stones or hair in the urethra. Hematuria, urinary tract infections and dysuria symptoms should raise suspicion on such pathologies. Self-esteem and other psychologic problems are also not uncommon in adolescent and adult repaired or unrepaired hypospadias.

The Bladder Reservoir and the Outlet: Role of Bulking, Sling, Artiphicial Sphincter and Surgery for Continence
Mario De Gennaro

Injection of a bulking agent into the bladder neck area as a primary treatment of bladder outlet incompetence is a very commonly performed procedure in the past year . Some authors suggest to perform urodynamics before the treatment in order to select patients (De Gennaro) , other considered urodynamics not useful (Lotman).
The results are scant at the follow-up in all series anyway and the procedure is not recommended because of low success rates. Neverthless bulking agents in bladder neck are used and could be useful in selected patients and permits to gain time until puberty in mild form.

Many surgical approaches have been described for increasing bladder outlet resistance to achieve continence, however long-term results are lacking. Stress incontinence due to sphincter incompetence is most commonly managed with an abdominoperineal puboprostatic autologous fascial sling procedure in boys and a transvaginal autologous fascial sling procedure in girls. The success rate for dryness or improved continence is variable, 25–100% according to different series where in some cases bladder augmentation or Mitrofanoff procedure have been associated. Synthetic suburethral slings can only be used in a tension free mode, due to risk of erosion. Some authors report good results either in male either in female. Anyway in neurogenic stress incontinence a firmer suspension is needed, making synthetic slings inappropriate. Currently, there are no reports describing long-term results of the synthetic suburethral slings, and its use in a very young population should be avoided and performed after puberty.

Several Bladder neck reconstruction procedure have been described . The results are high in terms of continence , where more long is the reconstructed bladder neck , higher is effective for continence. Anyway surgeons must consider that this increase the risk for upper tract and bladder augementation is often required. Bladder neck reconstruction can be performed today with laparoscopic technique , as described by Chrzan with “U2 t be dry procedure “ and with robotic.

Artiphicial sphincter (AMS) has been considered as the most effective solution, and AMS seems really effective in selected patients, efficacy rates for complete dryness between voids vary between 56% and 91%. following insertion of the artificial urinary sphincter, the revision rate is anyway very high, about 1/3 require reoperation and device removal due to erosion is commonly described in children. Approximately half of the individuals able to empty before insertion of the artificial sphincter can do so afterwards, however, bladder dynamics can change postoperatively. Actually, AMS is suggested only in adolescents, post puberty or young adults. Up to 5 years later augmentation cystoplasty may be required in33% of patients in order to minimize the effect of this change on kidney drainage and function.

Dryness may also be achieved by closing the bladder neck combined with a cathetarizable stoma. Complications after a bladder neck closure have been reported in up to 31% of cases, with 15% developing vesicourethral fistula. Persistent leakage, more UTIs, stone formation, bladder perforation, and deterioration of the upper urinary tract have also been reported after bladder neck closure especially when CIC regularity is neglected.

Surgery for Continence
Giovanni Mosiello

In some cases the treatment of urinary incontinence requires a surgical treatment.
In patient with congenital malformations or neurogenic bladder the treatment is often tailored on the own clinical situation of the patient . The advantage to define a specific best treatment in different clinical situation present anyway the disadvantage that is difficult to compare the different series, resulting in scant evidence results. First worldwide accepted criteria is to perform surgery always after failure of all conservative treatment failure. Second one surgery must be mini-invasive as possible respecting anatomy and avoiding major surgery in very young children, this according to the physiological amelioration of continence after puberty, then as obvious consequence is better to avoid some continence procedure before puberty as artiphicial sphincter. Third one the surgical procedure must can improve continence or preserve renal function but the clinical results are not always related to resulting quality of life.

Last but not the least as in hydraulic the surgical procedure on the outlet will increase the bladder pressure with risk for the upper urinary tract, and a careful patient selection must be performed considering surgery for major reconstruction in order to avoid unnecessary surgical procedure as well as the need of new surgery after few years in order to preserve upper tract.
Surgery for continence could be performed:

- To increase reservoir
- To increase outlet resistance
- To permit catheterization
- To derivate

Bladder augmentation

When medication has failed to decrease elevated end filling detrusor pressure, or creates troublesome side effects, bladder augmentation may be indicated. Detrusor myectomy, or detrusorectomy,''auto-augmentation,'' shows a success rate of approximately 50% with respect to bladder compliance and capacity in neurogenic bladders. This procedure has been very popular in the past but was replaced , as miniinvasive procedure by botulinum toxin injection. Recently gained new popularity thanks to laparoscopic procedure. Ileocystoplasty is more commonly performed, but carries the risk of postoperative intestinal obstruction, mucus retention, increased rate of stone formation, and electrolyte imbalance. The risk of complication or effectiveness is the same either with ileum or with sigma and the choice is related to surgeon's preference and experience. The risk of secondary malignancy of the augmented bladder is increased, although less than 20 cases have been described worldwide.
Augmentation may be combined with ureteral reimplantation, bladder neck tightening (sling suspension, bladder neck reconstruction, artificial sphincter implantation) or the creation of a continent catheterizable urinary stoma (Mitrofanoff, Monti). As bladder augmentation lowers bladder pressure, diminishing or abolishing vesicoureteral reflux, ureteral reimplantation should only be performed in cases where high grade reflux occurs at low bladder pressure. Similarly, as bladder augmentation will improve continence, only patients with low leak point pressure need reinforcement of the bladder outlet. Urodynamic testing will determine surgical options. Bladder replacement instead of augmentation may be appropriate in cases of bladder exstrophy where use of native bladder tissue is impossible. The use of tissue engineering is still far from a clinical use and this treatment can not be considered in the next 10 years.
Derivation
Ileal conduit ('wet deviation') is no longer indicated except in case of severe mental disability or severe renal dysfunction and no options for bladder reconstruction.

Bowel Management
Giovanni Mosiello

Bowel dysfunctions, constipation and incontinence, are common conditions in paediatric population resulting often in a negative impact on Quality of life . Different bowel management program are used ranging from conservative treatment (dietary, laxatives, rectal suppositories) to mini-invasive (transanal irrigation) until invasive treatment (Malone anterograde continence enema or sacral neuromodulation). The treatment of bowel dysfunction has been changed in the past 10 years by the reintroduction in daily clinical practice of a transanal irrigation (TAI

Patient population:

Anorectal malformation
Patients with ARM necessitate a bowel reconstruction. ARM is often associated to spine defect and BD could be the result of both. To insure continence and bowel emptying, a regular program for colon cleaning is necessary for most of patients either temporarily or life-long. Peña first introduced a specific BM, mainly based on the irrigation of colon with different solutions. This procedure is time consuming and variably tolerated by patients and caregiver. For these patients, the effectiveness of BM is significantly related to the improvement of quality of life (QoL).

Spina Bifida
Children with congenital or acquired spinal cord injury (SCI) present commonly with BD, requiring a life-long prolonged BM: in myelominingocele (MMC) abnormal defecation is present in 68% . Thirty-nine percent of patients with SCI reported BD has a negative impact on their QoL . Until now the care of BD has been underestimated and still empirical compared to the management of bladder dysfunction.

Options for Bowel Management
Fluid intake and diet
Physical activity
Medications
Anal/rectal stimulation and Manual evacuation

Suppositories and micro-enemas
Anal plug
Abdominal massage and Biofeedback

TAI
Sacral nerve modulation (SNM), Tibial nerve stimulation (PTNS), Transcutaneous electrical nerve stimulation (TENS), Surgical options

Conclusions

The treatment of BD significantly affects the family and caregivers quality of life and has a high social cost. Before the use of TAI surgical procedures were common: percutaneous cecostomy, bowel resections, permanent stomas and Malone procedur. This procedure is effective in the long-term management of neurogenic bowel but the complications and re-exploration rates are high The percutaneous cecostomy was widely used before the introduction of laparoscopic Malone. The permanent stoma is the most invasive and definitive alternative, but its' use should be limited only to selected children.
TAI is an effective method for bowel management. It is easy to perform and inexpensive. However can be associated with abdominal cramps, shivering, electrolyte imbalance, and with more severe but rare complications: systemic reactions to irrigation solutions and rectal perforation . Different solutions, probes and modalities have been described. In the last teen years new systems providing advanced, more effective, and safe irrigation have been introduced. This advanced TAI revolutionized the BM in adults and children. The critical point remains the selection and indication. In order to improve the efficacy TAI treatment should be individualized to each patients . A structured standardized approach to the treatment of constipation and fecal incontinence improves symptoms, quality of life and decrease hospital readmissions. The BM should be tailored in each individual patient. A correct BM remains multidimensional in TAI era too.

W27 Transitional care

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15:45 **Questions**

15:55 **Take Home Message**

GIOVANNI MOSIELLO, MD, FEAPU, FEBPS

Affiliations to disclose:

Medtronic: consultant
Wellspect: consultant
Coloplast: consultant
Pfizer: PI in clinical trial
Ipsen: PI in clinical trial
Allergan: PI in clinical trial

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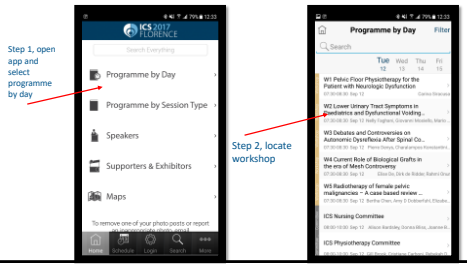
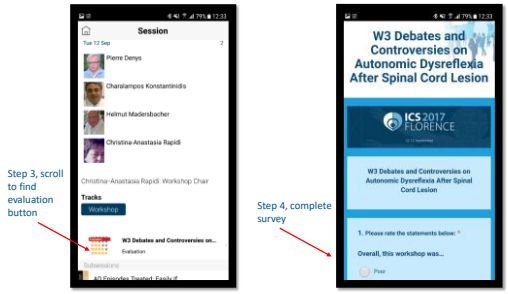


W2 Luts in pediatrics



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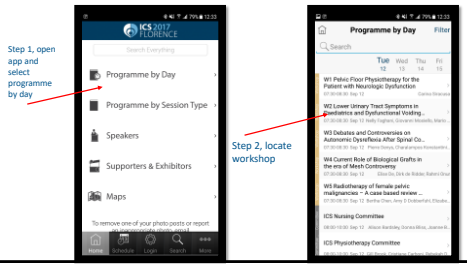

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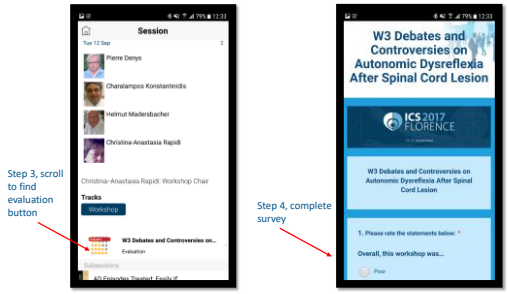

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Jian Guo Wen

Affiliations to disclose†:

† All financial ties (over the last two years) that you may have with any business organization with respect to the subjects mentioned during your presentation

Funding for speaker to attend:

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- [ ] Institution (non-industry) funded
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ICS Teaching Module

W27: ICS Core Curriculum (Free): Children and Young Adults Workshop Committee on Transitional Care for Continence in Congenital alformations.

An Overview From the Pediatric Urologist Prospective: Neurogenic Bladder, Posterior Urethral Valves, Bladder Exstrophy, Anorectal Malfromations, Guidelines

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Zhengzhou University, China
2. Neuro-Urology Clinical and Research Unit, Paediatric Hospital Bambino Gesù, Rome, Italy

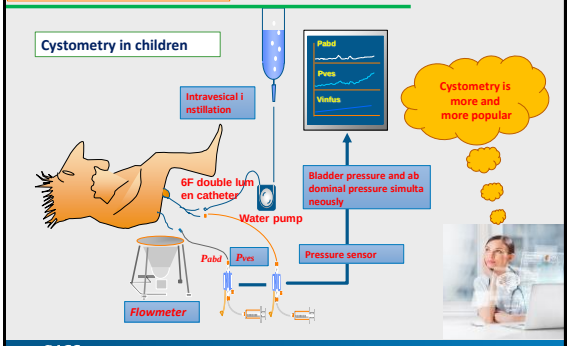
ICS International Continence Society
Teaching Module

Introduction

- Neurogenic bladder (NB) , Posterior Urethral Valves (PUV), Bladder Exstrophy(BE), Anorectal Malfromations (ARM) are most common confronted during the pediatric urological practice.
- Bladder dysfunction in these entity shows more or less the voiding dysfunction. This lecture provides an overview of these diseases from the urodynamic prospective.

ICS Teaching Module

Urodynamic study — Cystometry

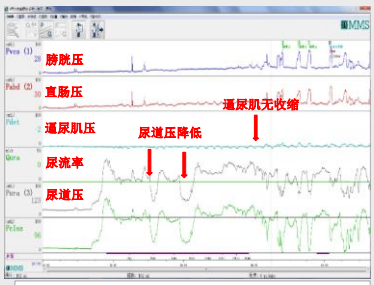


ICS Teaching Module

Synchronous bladder and urethral pressure measurement

Voiding at standing position

Voiding at supine position



Urethral instability

ICS Teaching Module

Synchronous bladder and urethral pressure measurement

Wensen catheter fixator

ICS Teaching Module

Video Urodynamic St udy

overview: outline

- Neurogenic Bladder
- Posterior Urethral Valves
- Bladder Exstrophy
- Anorectal Malfromations

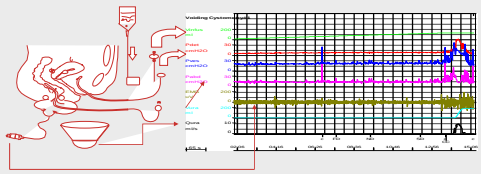
Neurogenic Bladder (NB)

- NB results from a variety of abnormalities of the c entral or peripheral nervous systems contributes t o various forms of lower urinary tract dysfunction.
- Myelodysplasia is common reason.

Spinal Dysraphi sm

NB

- The spinal level and extent of congenital lesion are poorly correlated with the clinical outcome
- *Urodynamic studies* are very valuable for defining t he extent of the pathology and planning treatment in children.



NB

Guidelines for urodynamics and uroneurophysiology tes ts in NB-EAU

Guidelines for urodynamics and uroneurophysiology tests	GR
Urodynamic investigation is necessary to document the (dys-)function of the LUT [10].	A
The recording of a bladder diary is advisable.	B
Noninvasive testing is mandatory before invasive urodynamics are planned.	A
Video-urodynamics are currently the preferred method for invasive urodynamics in patients with NLUTD. If this method is not available, then a filling cystometry continuing into a pressure-flow study should be performed.	A
For standard urodynamic testing, a physiologic filling rate (see Table 1; eg, not faster than 20 ml/min) and body-warm fluid must be used.	A
Specific uroneurophysiologic tests and provocative manoeuvres (eg, fast-filling cystometry with cooled saline [the ice-water test], coughing, tapping, and anal stretch) are elective procedures [10,12].	C

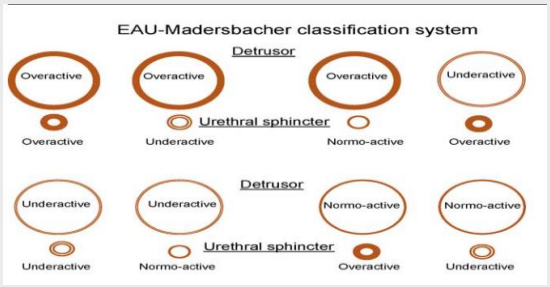
GR = grade of recommendation; LUT = lower urinary tract.

NB

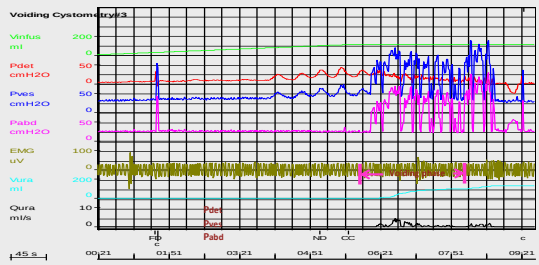
- Important urodynamic parameters
  - Bladder capacity and intravesical filling pressure;
  - Intravesical pressure at the moment of urethral leakage;
  - Presence or absence of reflex detrusor activity;
  - Competence internal and external sphincteric mechanisms;
  - Degree of coordination detrusor and sphincteric mechanisms;
  - Voiding pattern and post-voiding residual urine volume;
  - VUR

NB

The European Association of Urology (EAU)–Madersbacher classification system.



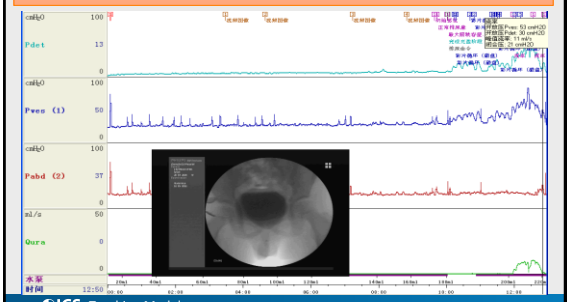
NB



Acontractile detrusor in a child with neurogenic bladder; urination achieved by abdominal pressure

NB

Video UDS provides more information of function and morphology.



NB

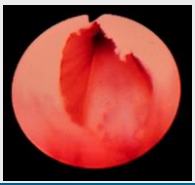
Management guided by UD

- anticholinergic drugs
- Electrical stimulation
- Injection of botulinum toxin
- Toilet training
- *clean intermittent catheterization*
- Surgical procedures

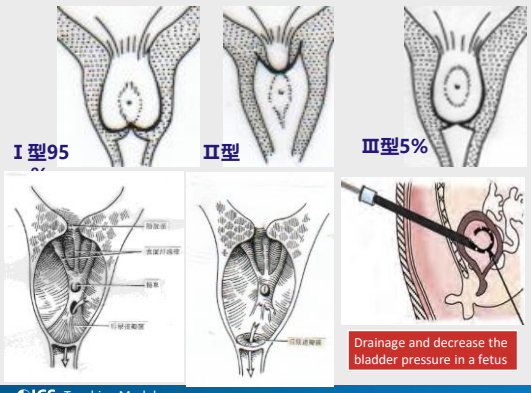
Posterior urethral valves (PUV)

- PUV is life-threatening congenital anomalies.
- PUV are found in 1 in 1,250 in a population undergoing foetuses in US

Type 1



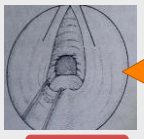
type3



PUV: Diagnosis and Treatment

- Bilateral hydroureteronephrosis distended bladder, a thick-walled bladder and a dilated posterior urethra are suspicious signs of PUV.
- Voiding cystourethrogram (VCUG) is common used, but video urodynamic study (VUDS) is recommended to confirms PUV.
- A secondary reflux is observed in at least 50% of patients.





Electrosurgical scalpel Valve ablation and Bladder drainage

PUV

PUV: Follow up by VUDS

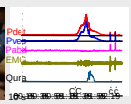
- Following surgical treatment, close follow-up to detect and monitor the bladder dysfunction that may lead to renal injury.
- The synchronous evaluation of structure and function of VUDS provides insight into the correlation and causation of detected anomalies.

VUDS for PUV cases during follow up

Bladder exstrophy (BE)

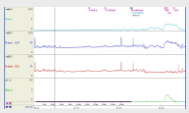
- BE is characterized by an infra-umbilical abdominal wall defect, incompleteclosure of the bladder with mucosa continuous with the abdominal wall, epispadias, and alterations in the pelvic bonesand muscles.
- Before bladder neck reconstruction, urodynamic assessment can be predictive for detrusor function and the ability of the bladder to increase in size without high intravesical pressures and also the application of anticholinergic therapy to enhance bladder volume.

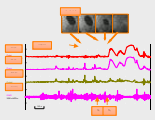


BE

- Following bladder neck reconstruction, urodynamic assessment provides an objective correlation with the clinical assessment of continence.
- It helps in planning pharmacotherapy for elimination of uninhibited detrusor contractions, improving bladder compliance and reducing intravesical pressures

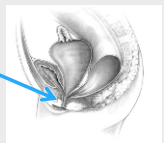




Anorectal malformations (ARMs)

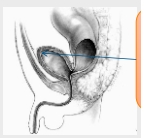
- ARMs involve the distal anus and rectum as well as the urinary and genital tracts. Renal agenesis, VUR, uninhibited detrusor contractions, poor compliance of the bladder or incomplete bladder emptying and NB are frquentely associated with ARM.
- Anorectal malformations are congenital anomalies that occur in approximately 1 in 5000 live births.

the urinary tract, vagina, and rectum all meet in a common channel and the baby has a cloaca



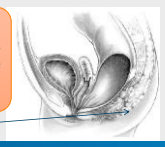
ARMs

- Early and repeated urodynamic evaluation is mandatory to detect as earliest as possible the onset of deterioration before irreversible neurological damage has occurred.
- Urodynamic studies are of limited value in the preoperative setting but are useful in the follow up of ongoing urological dysfunction postoperatively in patients with ARMs.



Recto-bladder neck fistula.

Recto-vestibular fistula in females



summary

- NB, PUV, E, ARM are very common in pediatric urological practice.
- Urodynamic study (UD) is important in assessing the bladder function before and after treatment in these diseases.
- UD is also play an important role in predicatiing the development of disease as well as preventing the renal deterioration of secondary to the bladder dysfunction.

Stoma: Indications and Concerns For Mitrofanoff and Malone

Mario De Gennaro

Urology & Urodynamics
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1

Mario De Gennaro

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Affiliations to disclose†:

No disclosure

† All financial ties (over the last year) that you may have with any business organisation with respect to the subjects mentioned during your presentation

Funding for speaker to attend:

- ☐ Self-funded
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Ileal Conduit - Stomal Complications

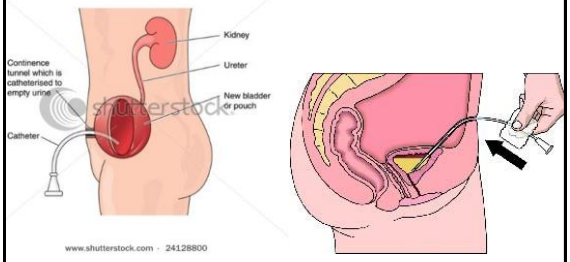
Stomal stenosis 0-15%

Stomal retraction 0-31%

Stomal bleeding 0-8%

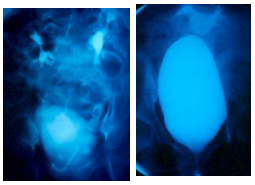
3

The Mitrofanoff principle Continent diversion



CASE REPORT - AUTONOMY

15 year-old lovely young lady: spina-bifida, neurogenic bladder and neurogenic bowel. Paraplegic and wheelchair user. Good student and supported by an excellent family



Normal upper urinary tract and normal renal function
Low-compliance, but good capacity bladder, open bladder neck, and no VUR

- She was on ICC through the native urethra + Oxy
- CIC was done by the mother 6 times daily (about 15' at home). She was also on daily rectal enema, again done by the mother (30 minutes long procedure)
- Despite this management, she was still on diapers day and night, wetting between ICC and having fecal incontinence
- She was particularly interested to become independent in the management of her condition

Patient, Family, Doctor choice:

•Bladder Neck Wrap and Suspension using the rectal muscle sheath;
•Autoaugmentation

•MACE with the appendix with an external lower right abdominal stoma (VQZ);
•Mitrofanoff conduit with a Full-Monti (ileum) with an external umbilical stoma;

-- 10 years after --

100% independent, 100% continent for urine and stool

She is doing self-CIC through the umbilical stoma, which is easy and fast (5 minutes) 4 to 5 times daily (no CIC at night)

She is doing alone also the bowel management with an antegrade enema 3 times weekly, with 700 cc of water plus a teaspoon of common salt. The procedure needs 20 to 30 minutes and is performed in the toilet, early in the morning introducing a catheter in the right abdominal stoma

Mitrofanoff umbilical stoma

MACE abdominal stoma

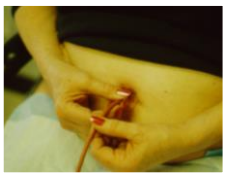
- ***Mitrofanoff***

- 1st described in 1980
- Continent supravesical catheterisable channel
- Appendix on vascular pedicle

10

Keep easier bladder emptying

- To be considered:
  Catetherizable urethra
  Hands dexterity/Mobility
  Motivation/Support
  Progression of the disease
  Age



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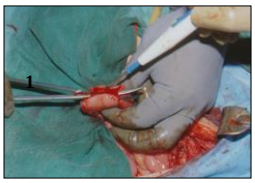
and if you don't have appendix?

- ***Monti***



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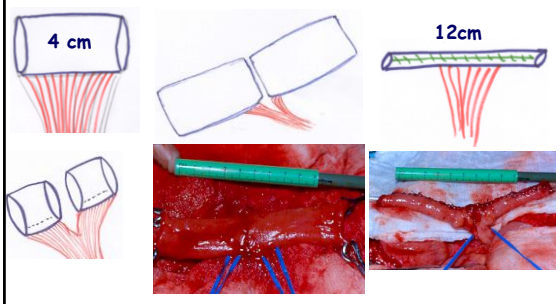
MONTI-YANG RECONFIGURED ILEUM AS MITROFANOFF TUBE



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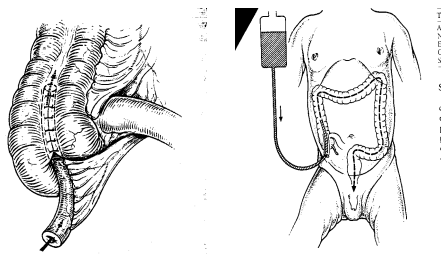
- **Bowel care**
  - **Antegrade colonic enema procedure**



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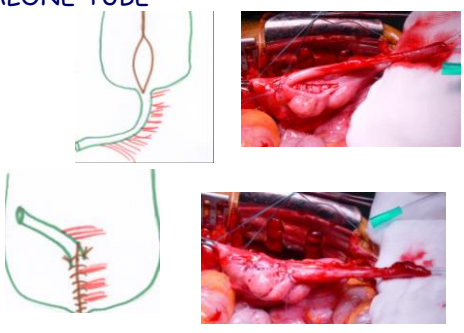
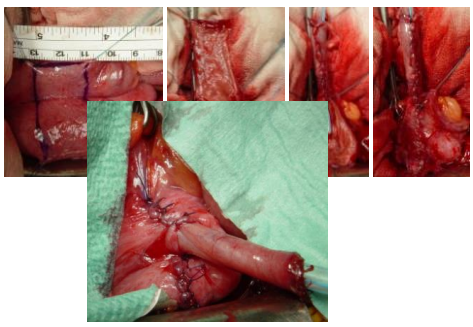
MACE

IN SITU IMBRICATED APPENDIX AS MALONE TUBE



-Macedo cecal flap for MACE (bowel)



Laparoscopic ACE-Malone

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Into adulthood

Mitrofanoff complications

- catheterisation difficulties
  - 27% of Mitrofanoff and 60% of Yang/Monti
- 10-40% stomal stenosis
- 2% stomal prolapse
- 28% sacculation
- 16-50+% revision rate

Sahedevan K et al. Is continent diversion using the Mitrofanoff principle a long term viable option for adults requiring bladder replacement. BJU Int 2008; 102: 236-240

21

Mitrofanoff complications

- 40% UTI
- 40-100% stones (higher rate both Mitrofanoff and cystoplasty)
- high patient acceptance
- patients with both urethra and Mitrofanoff prefer Mitrofanoff

Fishwick JE, Gough DC, O' Flynn KJ. BJU Int 2002; 85 (4): 496-497
Woodhouse CR, Lennon GN. Eur Urol 2001; 39 (3): 253-259
Horowitz M, Kuhr CS, Mitchell ME. J Urol 1995; 153 (3 Pt 1): 771-772

22

Contra-Indications

- Insufficient dexterity
- Can be used in quadriplegia

Harmon EP, Hurwitz G. South Med J 1994; 87 (10): 1005-1006

23

How to Manage Boys operated for Posterior Urethral Valve and Severe Hypospadias

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Affiliations to disclose†:

none

† All financial ties (over the last year) that you may have with any business organisation with respect to the subjects mentioned during your presentation

Funding for speaker to attend:

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References Related to PUV

The Valve Bladder Syndrome: 20 Years Later. Glassberg KI., J Urology,166:1406-1414;2001

Impact of transurethral resection on urinary flow rate in children with posterior urethral valve in short term follow up. Ipekci T., et al., Saudi MedJ,35(5):460-465;2014

Long term Bladder Dysfunction and Renal Function in Boys with PUV Based on Urodynamic Findings. Ghanem MA., et al., J Urology ,171:2409-2412;2004

Normal empty bladder management: Effecrtive therapy for the Seevere Valve Bladder. Koff SA., Br J Urol, 85 (suppl) : 18: 2000

Pre-transplant management of valve bladder: A critical literature review. Jesus LE and Pippi Salle JL., J Ped Urol., 11: 5-11; 2015

PUV Associated Problems

BO Obstruction due to Valve
BO Obstruction due to Bladder Neck
Bladder Dysfunction
Incontinence and/or Poor Emptying
Upper System Obstruction due to BOO
Upper System Obstruction due to Bladder Wall/Dysfunction
Upper System Obstruction due to Severe Hydronephrosis
Polyuria
Primary UTI or Surgery Associated UTI
Renal Dysplasia

Is Aggressive Management Worthed ?

YES!!!

we can avoid/postpone renal transplantation
we can avoid/postpone augmentation surgery
we can avoid/postpone upper system diversion
we can avoid/postpone CIC
we can avoid treatment related complications or morbidities

in selected cases….

Poor Prognostic Factors

Prenatal Severe Findings (oligohydro, bil severe hydro)
Postnatal Cr > 1 mg/dl
Dysplastic Kidneys
Severe Hydroureteronephrosis
Incontinence

BOO due to Valve

Make sure that Valves are Ablated Efficiently

Residual valves (10-80%)

- VCUG
- Endoscopy
- Uroflowmetrics and PVR
- Urodynamics

BOO due to Valve

- VCUG
  - Posterior urethra/penile urethra ratio
- Endoscopy
  - 12 o'clock rest valve, ant valve, strict
- Uroflowmetrics and PVR
  - Qmax <15 ml/sec and no change in PVR
- Urodynamics
  - High voiding pressures

BOO due to Valve

Who are in Greater Risk for Rest Valves ?

- Younger age at Ablation
- Poor emptying/Severe Hydronephrosis
- Sepsis/Severe UTI
- Diverted (Vesicostomy)
- Low volume centers

BOO due to Bladder Neck

Very Conflicting Findings

- No rest Valve but Poor Emptying

- Generally VCUG and UD findings
- Alpha blockers seems to work
- Botox
- Bladder neck incision

Kajbafzadeh AM., et al., J Urol; 178: 2142-2149, 2007
Keihani S., et al., Urology; 99: 278-280, 2017

Bladder Dysfunction

75% -90% abnormality

- Postvalve ablation UD is essential.

Basically Three Abnormal Types

- Low Compliance/Detrusor Hyperactivity
  (w high voiding pressure)
  assoc . upper tract dilat ((Full) Valve Bladder)
  by Mitchell, 1982
  by Duckett, 1997
- Normocompliance/Detrusor Hyperactivity
- High Compliance and Acontractile Bladder
  (Myogenic Failure)

Bladder Dysfunction

Valve Bladder is associated with ESRD

- Proactive anticholinergic and CIC

Myogenic failure is unknown

- Anticholinergics
- Age related disease nature

- Better outcome if recognized early
- Continous follow up

Incontinence and/or Poor Emptying

Incontinence is associated with Valve Bladder
- Careful work up VCUG and UDs
- Retention ?
- Small capacity ?

Poor emptying
- Rest valve
- Bladder Neck
- Psuedo-residual

by Glassberg, 1982

- Acontractile bladder

Upper System Obstruction due to BOO

A good valve ablation can drain the whole system
- Less need for diversions
- Bladder neck ???
- Alpha blockers ???

Upper System Obstruction due to Bladder Wall/Dysfunction

Thick bladder wall very rarely obstr ureters

Bladder cath in babies fail to drain but rather aggravates
- best option for vesicostomy
- early ablation no cath and give some time

A good bladder therapy can drain the whole system
- Less need for diversions

Upper System Obstruction due to Severe Hydronephrosis

Never Accept the residual dilatation is due to severe hydro

Good bladder therapy with good emptying will decrease the dilatation in most cases

Double or triple voiding may help continence but not the dilatation

CIC or vesicostomy can be tried

Polyuria

Patients with valve bladder not only have obstructed ureters but even nephrons
by Canning DA, 2001

Drained system will cause a severe diuresis (nephrogenic DI)

Drainage must be adapted to polyuria

Full valve bladder sydrome can be prevented with overnight catheterization

Koff SA, 2000

Primary UTI or Surgery Associated UTI

UTI
Primary
- Related to VUR
- Prepuce
- Poor emptying
- Severe Hydro

Surgery related
- Stricture
- Augmentation
- Mucus
- Stone

Renal Dysplasia

PUV is 1-15% of renal transplatation (RT) in children
Outcomes are similar with other causes if valve bladder management is satisfactory

Small bladder due to oliguria will grow with RT
Small bladder will grow with age
PVR will increase with age
No rush for Augmentation

Renal Dysplasia

Augmentation increases the UTI, graft loss and mortality risk if done prior or at RT

If bladder therapy fails for a compliance <20 cmH20 and >60% EBC, a prior Augmentation is justifiable

Always get ready for CIC before RT

CIC can be difficult sometimes in PUV, cath channel ????

RT is possible even in diverted cases (ileal conduits etc.,)

References Related to Hypospadias

The Prostatic Utricle: An under recognized condition resulting in significant morbidity. Hester AG and Kogan SJ., J Pediatr Urol, 2017

Urinary flow patterns in infants with distal hypospadias. Olsen LH., J Pediatr Urol, 7(4):428-432;2011

Normalized Urinary Flow at Puberty after TIP urethroplasty for hypospadias in Childhood. Andersson M., J Urol, 194(5): 1407-1413, 2015

Treatment of Adults with Complication from Previous Hypospadias Surgery. Myers JB., J Urol., 188(25): 459-463, 2012

Long term follow up of hypospadias: Functional and Cosmetic Results. Rynja SP., J Urol., 182 (4): 1736-1743, 2009

Long term functional outcomes of distal hypospadias reapir: a single center retrospective comparative study of TIP, MAGPI and mathieu. Hueber PA., J Pediatr Urol., 11(2): 68.e1-7, 2015

Hypospadias Associated Problems

Giant Utricle

Bladder Dysfunction

Voiding

Surgery related

- Short Term Success
- Long term Success
- Complications

Is Long Term Follow up Worthed ?

ABSOLUTELY !!!

- Complication rate increases in time
- Time is the best tester
- Fistula may appear late , very late…
- Surgeons observes other colleauges results
- Witnessed ESRD related to posthypos strictures

- Cosmesis related Social/Psychol Problems
- Marriage or Relationship Problems

Giant Utricle

Proximal hypospadias can be associated with

- Always cath before starting surgery
- Always keep the pediatric cystocope at the table
- Beware if there is UTI and full bladder during PE
- Having a prior USG is a smart move

Bladder Dysfunction

No satisfactory evidence to do UDs for all hypospadias or severe ones

If emptying problem, first rule out utricle

If postsurgery, first rule out urethral strict or path

Voiding

Hypospadias children generally weak voiders

BUT

- Curve is generally plateu
- Almost never with PVR
- If emptying problem, first rule out utricle
- If postsurgery, first rule out urethral strict or path

Post Surgery

Short term
- Retention
- Dysuria
- UTI
- Dribbling
- Slow Stream

Post Surgery

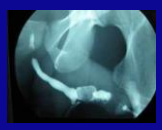
Long term
- Retention
- Dysuria
- UTI
- Dribbling
- Slow Stream

BIG PROBLEM !!!

Stream always gets better in time

DON'T BE DECEIVED BY APPEARANCE !!!




Post Surgery

Long term
- Stricture
- Stone

Good Urethroplasty
BUT
Ejaculation problems

Cosmesis

Self esteem

PENILE LENGTH

- Penis is short in prox cases
- Length is an issue in postpubertals
- Postsurgical shortening can be managed

Transitional Care for Continence In Congenital Malformations ICS 2017, Florence

COSMESIS

Transitional Care for Continence In Congenital Malformations ICS 2017, Florence

The Bladder Reservoir and the Outlet: Role of Bulking, Sling, Artificial Sphincter

Mario De Gennaro

Urology & Urodynamics
Bambino Gesù Children Hospital
Roma, Italy

PROCEDURES FOR CONTINENCE

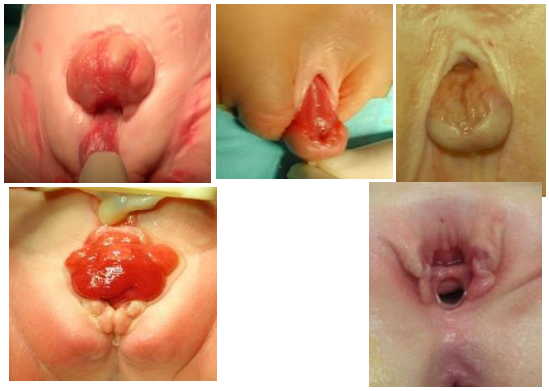
RESERVOIR

- Botox
- Auto Aumentation (laparoscopic)
- Ileo-Cystoplasty (robot assisted-lap)

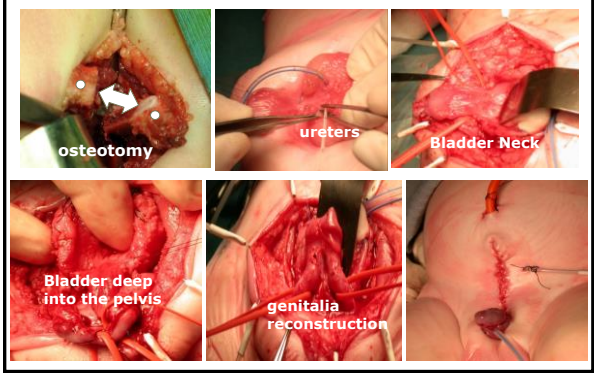
OUTLET

- Bulking (endoscopic)
- Sling
  - rectum fascia
  - eterologous
- (Artificial Sphincter)
- (Bladder neck surgery)

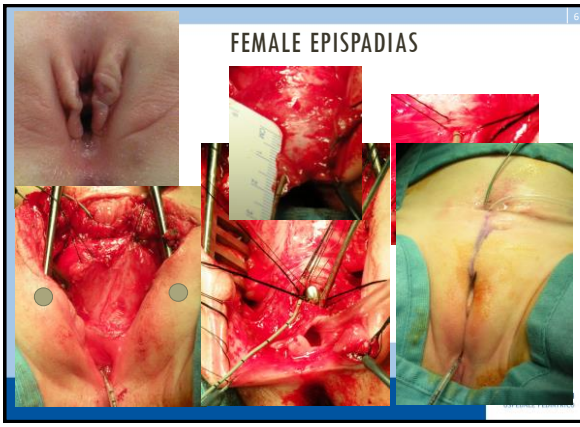
- Bulking – vesicoscopy
- Bladder Neck - vesicoscopy



EXSTROPHY: COMPLETE PRIMARY RECONSTRUCTION



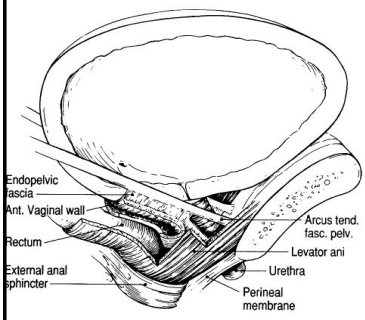
FEMALE EPISPADIAS

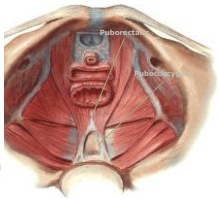


Epispadias Exstrophy

from Children to Adult FEMALE

Bladder/urethral support





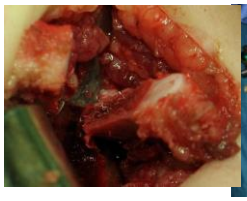
The pelvic floor (levator ani)

8

Pelvic Floor Reconstruction

G.C.
Exstrophy closure without osteotomy
Deiscence of bladder/abdominal wall

7 years
Anterior Osteotomy



The application of pelvic osteotomy in adult female patients with exstrophy: applications and outcomes

Mohd S. Ansari, John P. Gearhart*, Raimondo Maximilian Cervellione* and Paul D. Sponseller*
2011 BJU INTERNATIONAL | 108, 908–912

- **Patients:** 6 women (18-26 yrs) uterine/vaginal prolapse , bilateral innominate osteotomy. 3 previous suspension, 1 prior 5-attempts
- **Results:** 6/6 reduction pubic diastasis, no prolapse recurrence , 5/7 sexually activee

Quality of Life in Adults With Bladder Exstrophy-Epispadias Complex

Viviane Wittmeyer,* Estelle Aubry, Agnès Liard-Zmuda, Philippe Grise, Philippe Ravasse, Jannick Ricard, Jacques Biserte and Rémi Besson

- **Patients:** 25 (9 women, 16 men), 10 diversion
- **Results:** 2 married/partner, 22 high school, 18 (6 women) intercourse, 3women/7men had children (13), QOL scores < norm based

Health Concepts	Mean ± SD BEEC	Mean ± SD Normative Population	p Value
General health	56.4 ± 24	69.1 ± 18.6	0.006
Physical functioning	76.6 ± 33.9	84.5 ± 21.2	0.001
Role-physical	76 ± 29.9	81.2 ± 32.2	0.4
Role-emotional	90.3 ± 25	82.1 ± 32.2	0.21
Social functioning	80.5 ± 29.1	80.6 ± 21.4	0.8
Bodily pain	75.4 ± 29	73.4 ± 23.7	0.67
Vitality	58.8 ± 24.4	59.96 ± 18.05	0.74
Mental health	72.5 ± 25.3	68.5 ± 17.6	0.25

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PROCEDURES FOR CONTINENCE

OUTLET

- **Bulking (endoscopic)**
- Sling
  - rectum fascia
  - eterologous
- (Artificial Sphincter)
- (Bladder neck surgery)

- Bulking – vesicoscopy
- Bladder Neck - vesicoscopy

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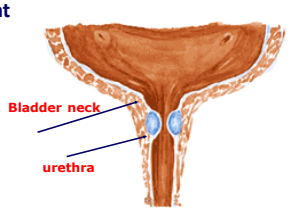
First Option: Bulking Agents

moderate Improvement

decreasing with time

repeatable

allows to wait



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OUTLET

- **Surgery for Bladder Neck**
  - not satisfactory in neurogenic patients
  - necessary (but not very effective) in exstrophies
  - to obtain real results complete closure may be required
- **Artificial Sphincter**
  - male, after puberty, very selected cases
  - late complications
  -

• Bulking – vesicoscopy

• Bladder Neck - vesicoscopy

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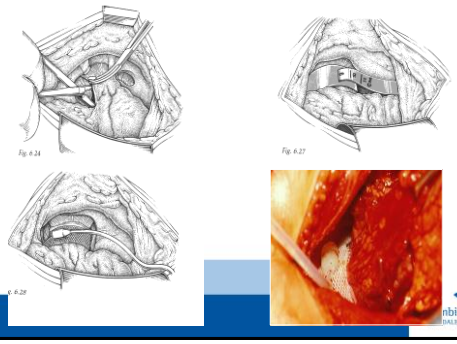
ARTIFICIAL SPHINCTER



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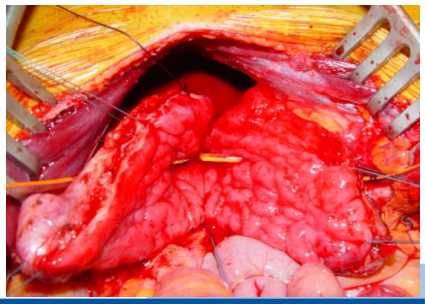
BLADDER NECK CLOSURE



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PROCEDURES FOR CONTINENCE

OUTLET

- Bulking (endoscopic)
- **Sling**
  - **rectum fascia**
  - **eterologous**
- (Artificial Sphincter)
- (Bladder neck surgery)

• Bulking – vesicoscopy

• Bladder Neck - vesicoscopy

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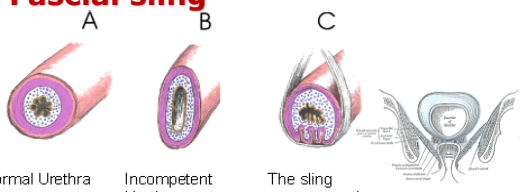
Fascial sling



Normal Urethra

Incompetent Urethra

The sling supports and partially compresses the urethral lumen

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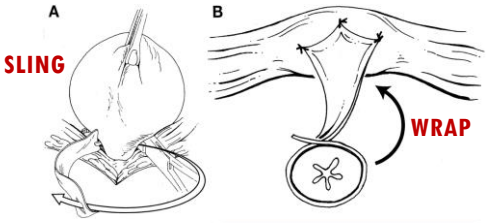
FASCIAL SLING / WRAP



BUGG, C. E. JR.; JOSEPH, D. B.
J Urol: 170(4, Part 2 of 2) October 2003 pp 1501-1504

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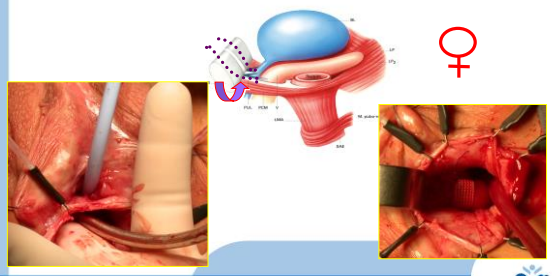
Sling procedure

Tension-free trans-Vaginal Tape (TVT)



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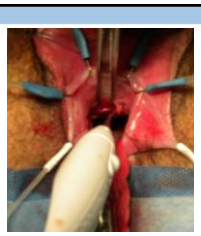
MINI SLING



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PROCEDURES FOR CONTINENCE

Reservoir & Outlet Surgery

- simultaneus when required, to prevent UUT and renal function damage
- nowadays Outlet (bladder neck/sling) before & Bladder if required (UDS sourveillance)

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J Urol. 2015

Long Term Outcomes of Bladder Neck Reconstruction without Augmentation Cystoplasty in Children.

Grimsby GM1, Menon V1, Schlomer BJ1, Baker LA1, Adams R2, Gargollo PC3, Jacobs MA4

109 patients Outlet surgery without AC (mean 8.5 yrs)

At 5 years f-up:

54% (59/109) additional continence surgery, (20/109) AC)
46% (50/109) developed VUR or hydro, and 21% (23/109) had onset or worsening renal scarring.

At time of AC: 13/18 had UUT changes, 15/18 continued incontinence, and 11/18 had EFP >40 cm H2O.

Reservoir & Outlet Surgery

- simultaneus ?
- Outlet surgery & Bladder if required ?

After Outlet only:

- the estimated 10 year cumulative incidence of AC is 30%
- other continence procedures 70%
- ***upper tract changes >50%, and CKD 20%***

careful patient selection and close follow up is essential if considering BO procedures without AC.

Surgery for Continence in Children

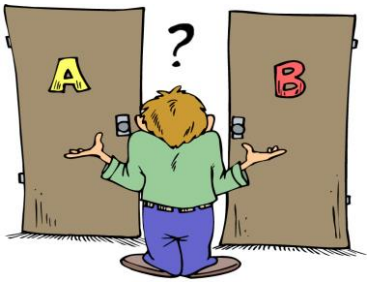
Surgical Intervention (principles)

The mainstay of current NBD in children is non-surgical

a small group who fails other treatments

no specific universal procedure for everyone (age, medical history, social status, disabilities)

explain different options



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ROBOTIC PAEDIATRIC UROLOGY: FOR CONTINENCE ?

sling

Bladder neck

Reimplant

Mitrofanoff

Bambino Gesù
OSPEDALE PEDIATRICO

Beyond Pediatric Incontinence: The Challenges of Transitional Care

June 6-8, 2013 | Toronto, Canada

Sponsored by

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ICS International Continence Society

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SAVE THE DATE

ICCS 2018
Rome
26-29 September
Meeting Chair
Mario De Gennaro

defoe Meeting Info | +39.0523.338391 | iccs2018@defoe.it

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THANK YOU

GRAZIE

ARRIVEDERCI A ROMA

Bambino Gesù
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ICCS
2018
Rome
26-29
September
Meeting Chair
Mario De Gennaro
Let's meet
in Rome!

SURGERY FOR CONTINENCE
Giovanni Mosiello

GIOVANNI MOSIELLO, MD, FEAPU, FEBPS

- Affiliations to disclose:

Medtronic: consultant
Wellspect: consultant
Coloplast: consultant
Pfizer: PI in clinical trial
Ipsen: PI in clinical trial
Allergan: PI in clinical trial

Funding for speaker to attend:

[x] Institution () funded

:

Long-term follow-up and late complications following treatment of pediatric urologic disorders

Akhavan A., Stock JA Med. Clin.N. Am., 95: 15-25,2011

- Long-term sequelae of reconstructive urology surgery can affect children for years and even decades after surgery.
- The need for life-long surveillance is critical in ensuring effective identification and management of late complications.
- As children mature into adolescence and adulthood, follow-up with the treating urologist become poor, and the responsibility for identifying late complications often rests upon the primary care provider.

ReviewArticle

AdultCareofChildrenFromPediatricUrology

ChristopherR.J.Woodhouse,*GuyH.Neild,RichardN.YuandStuartBauer

FromtheCentreforUrology,UniversityCollegeLondonHospitalsandtheCentreforNephrology,UniversityCollegeLondonRoyalFreeCampus,London,UnitedKingdom,andtheDepartmentsof Urology,Children'sHospitalBoston,Boston,Massachusetts

- Paediatric urology conditions requiring management in adulthood, including congenital anomalies on the genitourinary tract such us, renal disease**, congenital obstructive uropathy (PUV), spinal cord anomalies with neurogenic bladder or iatrogenic** causes , **bladder exstophry**
- **These conditions have a major lifelong implications and should require a bladder drainage mechanism**

Long-term continence care of Complex Congenital Malformations

- Spina Bifida and Neurogenic bladder (M and F)
- Anorectal Anomalies (M and F)
- Exstrophies/Epispadias (M and F)
- Urogenital Sinus –CAH (F)
- Ambiguous Genitalia (M and F)
- Vaginal Anomalies (F)
- Urethral anomalies and trauma (M)

Initial considerations

- few longitudinal studies (small series)
- old patients (adults) had been treated differently than new series (more conservative approach)
  - *change in knowledge*
  - *advanced nephrological treatment*
  - *sophisticated technology*
- different modalities/strategies in different series

Pediatric Surgery for Continence

Considered different options for:

- age, etiology, prognosis
- Patient and relatives motivation and desire
- Disability (all aspects) and Comorbidity
  - Mobility / Hand function
- Social and economic situation / caregiver
- Previous surgery

Different patients

Anesthesiology risk is increased for :

Neurogenic damage and sequelae

Scoliosis and reduced joint mobility

Pressure sores

Malnutrition

Drugs

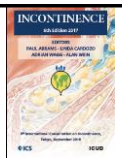
Gastrostomy ,Tracheostomy

Device : baclofene pump,

Ventriculo-peritoneal derivation

Surgeon

Results are highly dependent on the skills and experience of the individual surgeon.



Therefore graded recommendations for specific procedures cannot be provided. There are no randomized controlled trials (level 1 and 2 evidence).

Based on the available literature most studies have a level of evidence 3-4 and grade of recommendation C or D.

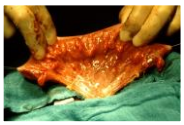
Surgical strategies

- **1) Improve Bladder Storage**
  - a. Detrusor characteristics
  - b. Outlet competence
- **2) Facilitate Emptying**
  - Intermittent v. Continuous
  - Urethral v. suprapubic (abdominal)
- **3) Supravesical Diversion**
  Conduit v. Continent

AUGMENTATION

Therapy-resistant overactivity of the detrusor, or small capacity and poor compliance, will usually need to be treated by bladder augmentation.

A simple bladder augmentation using intestine may be carried out if there is any bladder tissue, a competent sphincter and/or bladder neck, and a urethra that can be catheterised.



Ileal or colonic patches are frequently used for augmenting the bladder,
Alternative techniques for augmentation cystoplasty.
J Urol 1998

Before deciding on what type of procedure can be performed some significant factors must be addressed. These are

- Physical and mental capacity of the patient to do CIC
- Previous surgery (on urinary tract and bowel)
- Renal function status (including acid base state)
- Absence or presence of reflux
- Outlet resistance
- The need for a catheterizable channel

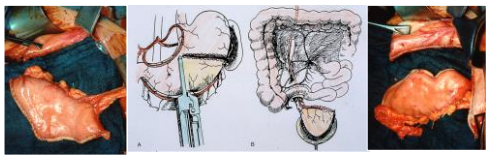
Bladder augments. ILEUM/COLON/STOMACH

Patients with low-capacity/high-pressure bladders

- intestinal neo-bladder
- consequences for renal function
- consequences for electrolyte balance
- life-long urological patients
- stones and infections
- malignancy

Stomach

Today is not more used as an augmenting patch because of the associated complications, but it has been widely used in 80'- 90'



Nguyen DH, Mitchell ME. Gastric bladder reconstruction. Urol Clin North Am 1991 Nov;18(4):649-57.

Augmentation cystoplasty is widely used in the surgical management of neurogenic bladder in patients with spina bifida, although ileal loop diversion is still performed

USA : 1998-2005 Spina Bifida Patients

Bladder augmentation was performed in 3,403 pts
ileal loop diversion in 772 pts

J Urol. 2011 Wiener JS et al

The estimated number of ACs performed on children is decreasing

Pediatric patients undergoing AC in the United States (US) for trends over the 2000s.

2000 : 792 Ac
2009 : 595 Ac

WHY? BOTOX effects?

J Urol. 2013 Schlomer BJ, Saperston K, Baskin L
National trends in augmentation cystoplasties in the 2000s and factors associated with patient outcomes.

Surgical complications of bladder augmentation: comparison between various enterocystoplasties
Ricardo González **Urology** 2000

79% required additional procedures

56% bladder neck procedures
23% continent stomas

Continence was achieved in 95%.

Calculi :developed more frequently in patients with continent stomas, no difference between segments

SBO : Sigmoid colon showed a trend of a lower rate of SBO
Perforation : no difference between segments

No difference at all

Surgical Techniques in Urology

Pediatric Laparoscopic Ileal Cystoplasty: Complete Intracorporeal Surgical Technique

Armando J. Lorenzo, José Cerveira, and Walid A. Farhat

Pure laparoscopic enterocystoplasty in children is an advanced procedure that is technically demanding. Although it appears feasible and provides a minimally invasive option to bladder augmentation, its equivalency or superiority over laparoscopic assisted or conventional open techniques remains to be demonstrated. UROLOGY 69: 977–981, 2007. © 2007 Elsevier Inc.

Surgical Techniques in Urology

Pediatric Robotic-assisted Laparoscopic Augmentation Ileocystoplasty and Mitrofanoff Appendicovesicostomy: Complete Intracorporeal—Initial Case Report

Mohan S. Gundeti, Michael K. Eng, W. Stuart Reynolds, and Gregory P. Zagaja

This preliminary first successful report suggests that robotic-assisted ileocystoplasty and appendicovesicostomy is feasible. A reasonable outcome with early recovery, resumption of normal activities, and excellent cosmesis can be achieved in selected patients. However, whether a robotic-assisted approach provides any significant advantages over conventional open procedures is yet to be determined with a large case series. UROLOGY 72: 1144–1148, 2008. © 2008

2017 : We don't know anything about complications of lap and robotic

Congential Anomalies

The Urologist's Role in the Management of Spina Bifida: A Continuum of Care

UROLOGY 76: 32–38, 2010

Douglass B. Clayton and John W. Brock, III

Preoperatively, nearly three-quarters of the patients had upper tract changes consisting of VURand/or hydronephrosis.

Augmentation alone or with reimplantation???

After augmentation, hydronephrosis resolved in all patients and VUR resolved in 76% without need for reimplantation.

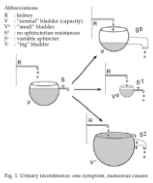
NEUROGENIC BLADDER IN CHILDREN: BASIC PRINCIPLES, NEW THERAPEUTIC TRENDS

J.M. Guys, G. Hery, M. Haddad, C. Borrionne

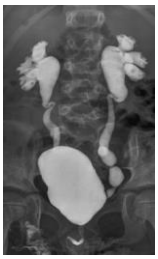
Scandinavian Journal of Surgery 100: 256–263, 2011

At the beginning endoscopic treatment combining anti reflux procedure, injection of the bladder neck and botulinum toxin can be considered as a "total endoscopic management" and should be our first line.

Or to postpone it



VESICO - URETERAL REFLUX TREATMENT IN NEUROGENIC BLADDER



A Minimally Invasive Approach in the Treatment of Vesicoureteral Reflux in Neurogenic Bladder in Children.
G Mosiello.Eur.Urol 2009

this approach was suggested 2 years before by our group

M

The Current Management of the Neurogenic Bladder in Children with Spina Bifida

Pediatr Clin N Am 59 (2012) 757–767
doi:10.1016/j.pcl.2012.05.006

Dominic Frimberger, MD[a,*], Earl Cheng, MD[b], Bradley P. Kropp, MD[c]

MINIMAL INVASIVE TREATMENTS AND NEUROMODULATION
Botulinum Toxin

The intravesical injection of botulinum toxin (Botox) is a good temporary measure to enhance bladder capacity and decrease intravesical pressures.[29] Botox is injected into the detrusor muscle endoscopically using a cystoscope under anesthesia. It is performed as an outpatient procedure, is generally well tolerated, and the effects

Detrusorectomy

Despite some advantages (e.g. avoiding mucus, decreased malignancy rate and fewer complications), **autoaugmentation cystoplasty, has not proven to be as successful as standard augmentation with intestine**

Detrusorectomy

49 detrusorectomies were performed (mean follow-up: 9.6 years) in 20 years

CONCLUSIONS:
The good short-term results of detrusorectomy generally remain unchanged at long-term follow-up.

Detrusorectomy reduces the need for augmentation and use of antimuscarinics in children with neuropathic bladders.
Chrzan R, et al , 2013. J Ped Urol

Detrusorectomy Lap

- 25 children , age of 9.3 years (range 0.9 to 14.2)
- Median follow-up was 6.8 years (range 0.1 to 15.6).
- Bladder autoaugmentation in children with NBD offers, after a transient decrease in bladder capacity, a long lasting increase in capacity and compliance, end filling pressure decreases

. EL Hansen,…. LH Olsen . J Urol 2013

Level of evidence 4
Grade of recommendation C

OTHER : Ureter ???

Ureterocystoplasty is rarely used but could be an useful and metabolically neutral alternative to bowel segments

Aktuelle Urol. 2010. Fisang C, Hauser S, Müller SC.

Ureterosigmoidostomy

Level of evidence : 3
Grade of recommendation: B

This type of continent urinary reconstruction may be utilized in reconstruction for bladder exstrophy, an incontinent urogenital sinus or the traumatic loss of the urethral sphincter.

As this reconstruction is totally dependent on the normal function of the anal sphincter, contraindications include incompetence of the anal sphincter, anal prolapse, previous anal surgery, and irradiation.

Because of the potential for electrolyte resorption, renal insufficiency is also a contraindication

Neurogenic Bladder : Surgical strategies

- 1) Improve Bladder Storage
  - a. Detrusor characteristics
  - b. Outlet competence
- 2) Facilitate Emptying
  - Urinary derivation

REVIEW PAPER

FUNCTIONAL UROLOGY

A review of prospective Clinical Trials for neurogenic bladder: The place of surgery, experimental techniques and devices

Cristian Persu[1], Emmanuel Braschi[2], John Lavelle[3]

73 full papers analyzed /580

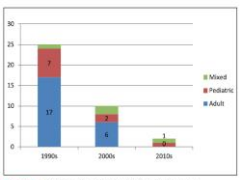


Figure 1. *Number of trials for each ten year period.*

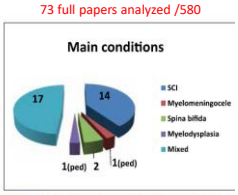


Figure 2. *Main conditions behind the neurogenic bladder.*

patients included in surgical trials is 3453, out of which 59% are males. The papers include a total of 369 children (21.2%), essentially looking at all the techniques that are also used in adults.

Conclusions There is still a lot of work to be done in order to obtain a significant level of evidence in the field of surgical procedures used in neurogenic bladder patients.

Postpubertal Urodynamic and Upper Urinary Tract Changes in Children With Conservatively Treated Myelomeningocele

Fayez Almodhen, Jean Paul Capolicchio, Roman Jednak and Mohamed El Sherbiny*
From the Division of Pediatric Urology, Montreal Children's Hospital and McGill University Health Center, Montreal, Quebec, Canada

When to perform surgery for continence?Before or after puberty?
Spontaneous increasing in Bladder Outlet resistance

Conclusions: This study demonstrates that total cystometric bladder capacity, maximum detrusor pressure and detrusor leak point pressure increase significantly in patients with myelomeningocele following puberty. The increase in bladder capacity could be attributed to increasing bladder outlet resistance resulting from prostate gland enlargement in males and estrogenization in females. A significant number of patients spontaneously achieve continence at puberty, and continence becomes more likely when increased total cystometric bladder capacity is not associated with an increase in maximum detrusor pressure. Finally, no significant postpubertal upper urinary tract deterioration was observed in our series.

JOURNAL OF LAPAROENDOSCOPIC & ADVANCED SURGICAL TECHNIQUES
Volume 23, Number 9, 2013
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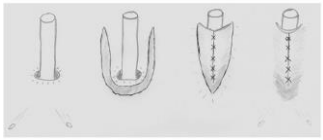
DOI: 10.1089/lap.2013.0018

Vesicoscopic Bladder Neck Procedure in Children: What We Have Learned from the First Series

Ratel Chrzan, MD, PhD, Pieter Dik, MD, PhD, Aart J. Klijn, MD, Caroline F. Kuijper, MD, Marleen M.C. van den Heijkant, MD, and Tom P.V.M. de Jong, MD, PhD



The declining long-term success rate indicates that the bladder neck region becomes damaged over time. The use of a full-thickness tube instead of a mucosal tube may be an important amelioration to protect the bladder neck from destruction. The construction of a continent channel for CIC can improve the outcome when anterior bladder neck plasty is performed.

Long-Term Outcome of the Pippi Salle Procedure for Intractable Urinary Incontinence in Patients with Severe Intrinsic Urethral Sphincter Deficiency

Shigeru Nakamura,* Taiju Hyuga, Shina Kawai and Hideo Nakai

THE JOURNAL OF UROLOGY®

Table 2. *Results of Pippi Salle procedure*

	Short-Term Results		
Pt No.—Age—Sex	Continence Status	Postop Complications	Additional Procedure
1—10—F	Wet	–	BA
2—15—F	Wet	–	BA
3—33—M	Dry	–	–
4—62—F	Wet	–	BA
5—12—F	Wet	–	BA
6—9—F	Dry	Difficulty catheterizing	TUI
7—16—M	Dry	–	–
8—11—M	Dry	–	–
9—45—F	Dry	Difficulty catheterizing	TUI
10—15—M	Dry	–	–
11—16—M	Dry	Difficulty catheterizing	TUI
12—12—M	Wet	–	BA

More neourethra is longer and narrower, more likely patients will be dry but they will have problems catheterizing neourethra.

Urologic Care of the Neurogenic Bladder in Children

Warren T. Snodgrass, MD*, Patricio C. Gargollo, MD

If we change the outlet we change the bladder

Table 1
Bladder neck reconstruction for neurogenic incontinence

Authors	Number of Patient (Male/Female)	Mean Follow-up	Number of "Dry" (%)	New Vesicoureteral Reflux (%)	Augmentation: Prior/ Simultaneous	Augmentation: Subsequent	Total (%)
Nili et al[37]	24 (10/14)	1.5–7 y	20 (83)	10 (42)			*a||*
Belman, Kaplan[34]	18 (10/8)	ns	14 (78)	4 (22)	16	1	17 (94)
Mollard et al[35]	16 (0/16)	12–36 mo	13 (81)	ns[a]	7		7 (44)
Snodgrass[38]	22 (13/9)	ns	20 (91)	9 (50)[b]	19		19 (86)
Salle et al[34]	17 (7/10)	26 mo	12 (70)	2 (12)[c]	12	1	13 (76)
Hayes et al[35]	28 (12/16)	28 mo	18 (64)	ns	23		23 (82)

Journal of Pediatric Urology (2014) 10, 380–391

Journal of Pediatric Urology
ELSEVIER

Long-term fate of the bladder after isolated bladder neck procedure

Benjamin Whittam *, Konrad Szymanski, Rosalie Misseri, Aaron Carroll, Martin Kaefer, Richard Rink, Mark Cain

BNR could be performed alone if UD showed an adequate bladder . In 45 % of these patients anyway a BA is required during time!

Table 3 Indications for augment and time to delayed augment.

Patient	Bladder neck	Time to augment (m)	Indications for augment
1	Bladder neck sling	3	New Incontinence Refractory contractions
2	AUS	8	Worsening compliance New Incontinence
3	AUS	10	Worsening compliance Poor capacity (<50%EBC)
4	AUS	13	Worsening compliance New VUR
5	Bladder neck sling	14	Worsening compliance New Incontinence
6	AUS	19	Poor capacity (<50%EBC) Worsening compliance New Incontinence
7	Bladder neck sling	19	New hydronephrosis New VUR
8	YD + BNS	29	Worsening compliance New hydronephrosis
9	Bladder neck sling	34	Poor capacity (<50%EBC) Worsening compliance New Incontinence
10	AUS	38	Worsening compliance Poor capacity (<50%EBC)
12	Krapp	55	New VUR New renal scarring
12	Bladder neck sling	88	New hydronephrosis New renal scarring Worsening renal function
13	Bladder neck sling	117	Worsening compliance New VUR

Note. AUS = artificial urinary sphincter; BNS = bladder neck sling; EBC = estimated bladder capacity; VUR = vesicoureteral reflux; YD = Young Dees.

Long Term Outcomes of Bladder Neck Reconstruction without Augmentation Cystoplasty in Children,
GM Grimsby et al, J Urology

109 patients underwent BNR without AC mean age of 8.5 years old.

Following BNR without AC, the estimated 10 year cumulative incidence is:

AC 30%
upper tract changes >50%
CKD 20%.

Because of these risks, careful patient selection and close follow up is essential if BNR without AC.

Bladder Neck Closure in Children: Long-Term Results and Consequences

Sara Hernandez-Martin[1] Pedro Lopez-Pereira[2] Sergio Lopez-Fernandez[1] Rubén Ortiz[1]
Mercedes Marcos[1] Roberto Lobato[2] Maria Jose Martinez-Urrutia[2] Enrique Jaureguizar[2]

Table 2 Main complications

Complications	No. of patients (%)
Fistula	4 (20)
Bladder stones	8 (40)
Catheterizable conduit	7 (35)
Stenosis	4 (20)
Leakage	3 (15)
Bladder perforation	2 (10)
Upper tract dilatation	0 (0)

Conclusions BNC is an effective approach to incontinence when other procedures have failed. In the long term, the most frequent complications are those related with catheterizable stoma and stones. The high incidence report of a low fertility index and erectile dysfunction meant further study in a larger cohort.

Surgery for continence in children :

- Surgery , especially if mini-invasive can improve QOL, but non surgical management is always the best
- Exhaust conservative measures
- Careful selection mandatory
- Multidisciplinary approach for choosing the best timing for surgery

Thank You

BOWEL MANAGEMENT
Giovanni Mosiello

ICS

GIOVANNI MOSIELLO, MD, FEAPU, FEBPS

Affiliations to disclose:

Medtronic: consultant
Wellspect: consultant
Coloplast: consultant
Pfizer: PI in clinical trial
Ipsen: PI in clinical trial
Allergan: PI in clinical trial

Funding for speaker to attend:

	Self-Funded
x	Institution (non-industry) funded
	Sponsored by:

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Defecation disorders in children

Constipation / fecal incontinence

- **Functional** 95%
  - **Constipation** > 90-95%
  - **Non-retentive fecal incontinence** <5-10%
- **Organic** 5%
  - **Anorectal malformations**
  - **Spinal problems**
  - **Miscellaneous**

Di Lorenzo C, Benninga MA, Gastroenterology 2004

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BACKGROUND

•There is evidence of long-life treatment of BD in these pediatric patients: Spina Bifida, AnoRectal Malformations , Hirschsprungs Disease.

ICS

BBD

Altered bowel and bladder control is common in children, mainly, in those with disabilities. In these patients management of the two problem as to be tailored as a consequence of associated comorbidities.

Bladder Bowel dysfunction (BBD)

- Lower urinary tract dysfunction
- Bowel Dysfunction

Several studies in literature reports the relationship between bowel and bladder and the terminology BBD is a clear message that these two entities have to be treated synergically

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Anorectal Malfromation

Genitourinary Anomalies	40%
Sacral anomalies	25 %
LUT Dysfunction	15 %

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Journal of Pediatric Surgery 52 (2017) 463–468

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Outcome in adults with anorectal malformations in relation to modern classification — Which patients do we need to follow beyond childhood?

Johan Danielson [a,c,*], Urban Karlbom [b,d], Wilhelm Graf [b,d], Tomas Wester [e]

Results: The ARM-patients had an inferior outcome ($P < 0.05$) for all incontinence parameters, 8 of 10 parameters for constipation, 2 of 6 for urogenital function and 7 of 13 quality of life parameters. Patients with rectobulbar and vestibular fistulas had the worst statistical outcome but patients with cloaca and rectoprostatic/bladder-neck fistula had worse outcome in absolute numbers. Forty-four patients (32%) reported incontinence of stool at least once a week and 16 (12%) had a permanent colostomy.

Conclusions: The functional outcome and quality of life in adults with anorectal malformations are closely related to the type of malformation. A large proportion of the patients have persistent fecal incontinence, constipation and sexual problems that have a negative influence on their quality of life. Structured multidisciplinary follow-up of adults with anorectal malformations by pediatric and colorectal surgeons, as well as urologists and gynecologists is therefore advocated.

Seminars in Pediatric Surgery 25 (2016) 112–116

journal homepage: www.elsevier.com/locate/sempedsurg

Congenital cloaca: Long-term follow-up results with emphasis on outcomes beyond childhood

Risto J. Rintala, MD, PhD*

	Continent (%)	Continent stoma/CIC (%)	Incontinence (%)	Renal failure (%)
Davies (2010)[12] ($n = 15$)	20	n.r	80	n.r
Couchman et al.[13] ($n = 19$)	47	53	n.r	21
Rintala (2015) ($n = 27$)	63	19	15	11

NB in Children

Bowel Dysfunction - Treatment

- normal healthy diet
- laxatives
- rectal suppositories
- (digital stimulation)
- **transanal irrigation**
- transrectal stimulation / sacral NMD
- anal plug
- Surgery

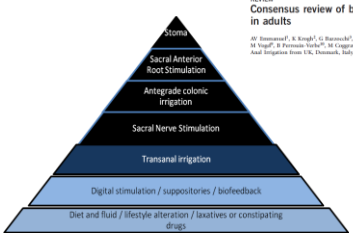
TAI USE IN ADULTS has been WELL DEFINED IN A STEP-WISE APPROACH

OPEN

REVIEW

Consensus review of best practice of transanal irrigation in adults

AV Emmanuel[1], K Krogh[2], G Bazzocchi[3], A-M Lund[4], A Bremner[5], D Leder[6], D van Rappenvelt[7], G Mosiello[8], M Vogt[9], B Perrouin-Verbe[10], M Coggrave[11], P Christensen[12] and Members of the working group on Trans Anal Irrigation from UK, Denmark, Italy, Germany, France and the Netherlands



Emmanuel AV, Krogh K, Bazzocchi G et al. Spinal Cord 2013; 51: 732-8

PEDIATRIC STUDIES

Transanal irrigation, increased independence from the caregiver and improved QoL in paediatric patients with Spina bifida and anorectal malformations.
(Midrio P. Colorectal Dis 2015)

Transanal colonic irrigation is a valid alternative to invasive surgical procedures and should be considered the first line of treatment for bowel management in children with soiling where simple pharmacological maneuvers failed to be effective.
(Pacilli M. J Pediatr Surg 2014)

Transanal irrigation has proven to be safe and effective in promoting intestinal emptying
(Marte A Minerva Pediatr 2013)

"We recommend that this simple therapeutic method be considered as a safe and valid choice for treatment of chronic constipation and fecal incontinence".
(Choi EK Spinal Cord 2013)

Transanal irrigation appears to be safe and effective bowel management system, which improves bowel function and QoL in children with faecal incontinence as a result of chronic idiopathic constipation, Hirschprung's disease and anorectal malformations.
(Nasher O. Int J Pediatr 2014)

Transanal irrigation is an effective, safe, non operative alternative to MACE.
(Corbett P. J Pediatr Urol 2014)

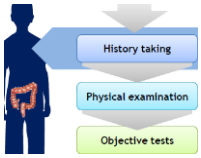
An evidence-based review of the use of transanal irrigation in children and young people with neurogenic bowel.

Studies (16) indicate that transanal irrigation can be an effective treatment for c hildren and young people with constipation or faecal incontinence due to NBD (n=346 children). Transanal irrigation is reported as improving quality of life and f amily's satisfaction with their bowel management...

Suggestion in Conclusion

Evidence-based review of the use of transanal irrigation in children and young people with neurogenic bowel may be considered as a first line treatment option for children and young people with neurogenic bowel but further research is required to prove the efficacy and acceptability of this procedure.

(Bray L. Spinal Cord 2013)



ICS

ACE-Malone (laparoscopic)

Surgery?

Cecostomy button
(temporary)

ICS

14

Consensus review of best practice of transanal irrigation (TAI) in children

- **Created by 6 international pediatric clinicians from different specialties: Urologists, Gastroenterologists and Surgeons + 1 Coloplast specialist**
- **An international consensus on aspects like indication, training, troubleshooting and follow-up of pediatric patients in need for TAI**

Giovanni Mosiello, David Marshall, Udo Rolle, Célia Crétolle, Bruno G. Santacruz, Jason Frischer, Marc A. Benninga
J Pediatr Gastroenterol Nutr 2017

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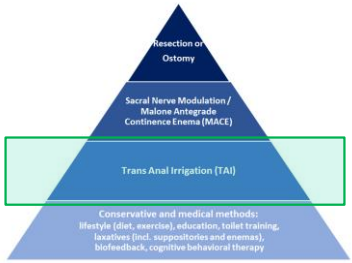
Treatment pyramid for defecation disorders in children



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Conclusions

The main objective of bowel management is to ensure a complete bowel evacuation to prevent complications.

Ensuring of a good bowel management is important to guarantee the best QoI of patients and care givers

paediatric bowel dysfunction but must be performed in a step- wise approach

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