Aims of Workshop
Spina bifida, bladder exstrophy/epispadia, posterior urethral valves, hypospadias and anorectal malformations are surgically managed in childhood but all these patients require life-long urological care for the treatment of incontinence. 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

**Suggested Reading**


**An Overview From the Pediatric Urologist Prospective: Neurogenic Bladder, Posterior Urethral Valves, Bladder Extrophy, Anorectal Malformations, Guidelines**

Jianguo Wen, MD, PhD, Professor

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Zhengzhou, China

Neurogenic bladder (NB), posterior urethral valves (PUV), bladder extrophy (BE), anorectal malformations (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 hydrourereteronephrosis 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
Bladder extrophy
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 obstructive 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 caretaker. Both stomas’ have a common goal which is to keep the bladder ad bowel as empty as possible, to avoid damage to the kidneys, 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-urolologists 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, hydrourephrosis and lower urinary tract dysfunction. A proper posterior urethral valve ablation may improve the bladder cycling to prevent hydrourephrosis, 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 stream 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 un repaired 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 consider urodynamics not useful (Lotman).

The results are scant at the following up in all series anyway and the procedure is not recommended because of low success rates. Nevertheless, 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 augmentation 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 in 33% 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 catheterizable 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 minimally invasive 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 extrophy 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.
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 procedure. 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**

Transitional Care for Continence in Congenital Malformations.

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**GIOVANNI MOSIELLO, MD, FEAPU, FEBPS**

Affiliations to disclose:
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- Wellspect: consultant
- Coloplast: consultant
- Pfizer: PI in clinical trial
- Ipsen: PI in clinical trial
- Allergan: PI in clinical trial

Funding for speaker to attend:
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Introduction

➢ Neurogenic bladder (NB), Posterior Urethral Valves (PUV), Bladder Exstrophy (BE), Anorectal Malformations (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.

Urodynamic study

Cystometry

Synchronous bladder and urethral pressure measurement

Urinary incontinence

Bladder pressure and abdominal pressure simultaneously measured

Cystometry is more and more popular

Voiding at standing position

Voiding at supine position

Urethral instability

Wensen catheter fixator
Neurogenic Bladder (NB)

- NB results from a variety of abnormalities of the central or peripheral nervous systems contributing to various forms of lower urinary tract dysfunction.
- Myelodysplasia is a common reason.

**Guidelines for urodynamics and uro-neurophysiology tests in NB-EAU**

- Initial investigation is necessary to document the severity of the UUT (UI).
- The recording of a bladder diary is advisable.
- Indirect tests are mandatory before invasive urodynamics are planned.
- Flexible cysto-urography is the preferred method for invasive urodynamics in patients with MNU.
- If this method is not available, then a filling cystometry combining into a pressure-flow study should be performed.
- For renal urodynamics testing, a physiologic filling rate (see Table 1, e.g., not lesser than 20 ml/min) and body position should be considered.

Specific uro-neurophysiological tests and provocative maneuvers (e.g., filling cystometry with cooled saline in the supine test, coughing, straining, and anal stricture) are electroproctography (EPT).

NB

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

**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 of internal and external sphincteric mechanisms;
- Degree of coordination of detrusor and sphincter mechanisms;
- Voiding pattern and post-voiding residual urine volume;
- VUR
The European Association of Urology (EAU) - Madersbacher classification system.

**Voiding Cystometry**

- **Pdet** cmH2O
- **Pves** cmH2O
- **Pabd** cmH2O
- **EMG** uV

**Video UDS** provides more information of function and morphology.

### 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

Management guided by UD

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

Drainage and decrease the bladder pressure in a fetus
Bilateral hydroureteronephrosis distended bladder, a thick-walled bladder and a dilated posterior urethra are suspicious signs of PUV.

Voiding cystourethrogram (VCUG) is commonly used, but video urodynamics study (VUDS) is recommended to confirm PUV.

A secondary reflux is observed in at least 50% of patients.

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.

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.

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.

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.

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 frequently associated with ARM.

Anorectal malformations are congenital anomalies that occur in approximately 1 in 5000 live births.

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 ARM.
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 predicting 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
Bambino Gesú Children Hospital
Roma, Italy

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

The Mitrofanoff principle
Continent diversion

Ileal Conduit - Stomal Complications

Stomal stenosis 0-15%
Stomal retraction 0-31%
Stomal bleeding 0-8%
**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**

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

---

**Monti**

---

**Keep easier bladder emptying**

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

---

**and if you don’t have appendix?**
- **Bowel care**
  - Antegrade colonic enema procedure

**MONTI-YANG RECONFIGURED ILEUM AS MITROFANOFF TUBE**

**IN SITU IMBRICATED APPENDIX AS MALONE TUBE**

-Macedo cecal flap for MACE (bowel)
Mitrofanoff complications

- catheterisation difficulties
  - 27% of Mitrofanoff and 60% of Yang/Monti
- 10-40% stomal stenosis
- 2% stomal prolapse
- 28% sacculcation
- 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

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 KU. BJU Int 2002; 89 (4): 496-497

Contra-Indications

- Insufficient dexterity
- Can be used in quadriplegia

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

Selcuk Yucel, MD
Professor in Urology and Pediatric Urology
Acibadem University School of Medicine, Istanbul, Turkey

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

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 Valve**

Very Conflicting Findings
No rest Valve but Poor Emptying

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

**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 UD's
- 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 syndrome 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 transplantation (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 cmH2O 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

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 colleagues 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 cystoscope 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 UDIs 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 plateau.
- 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

**BIG PROBLEM !!!**

Stream always gets better in time.

Post Surgery

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

**DON'T BE DECEIVED BY APPEARANCE !!!**

- 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

**COSMESIS**
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

Pelvic Floor Reconstruction

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

7 years
Anterior Osteotomy

Quality of Life in Adults With Bladder Exstrophy-Epispiadias Complex

Viviane Wittmeyer,* Estelle Auzel, Agnès Lendl-Zmuda, Philippe Grine, Philippe Reussse, Janvick Rict, Jacques Bierte and René Beacon

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

<table>
<thead>
<tr>
<th>Health Concepts</th>
<th>Mean ± SD DEEP</th>
<th>Mean ± SD Normative Population</th>
<th>p Value</th>
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<tbody>
<tr>
<td>General health</td>
<td>59.4 ± 24</td>
<td>66.1 ± 10.0</td>
<td>0.004</td>
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<tr>
<td>Physical functioning</td>
<td>76.6 ± 13.9</td>
<td>84.5 ± 21.2</td>
<td>0.400</td>
</tr>
<tr>
<td>Role-physical</td>
<td>76 ± 25.9</td>
<td>81.2 ± 32.2</td>
<td>0.4</td>
</tr>
<tr>
<td>Role-physical</td>
<td>89.2 ± 25</td>
<td>85.1 ± 32.2</td>
<td>0.21</td>
</tr>
<tr>
<td>Social functioning</td>
<td>89.5 ± 20.1</td>
<td>96.5 ± 21.4</td>
<td>0.0</td>
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<tr>
<td>Bodily pain</td>
<td>75.4 ± 25</td>
<td>73.4 ± 23.7</td>
<td>0.67</td>
</tr>
<tr>
<td>Vitality</td>
<td>58.6 ± 24.4</td>
<td>55.96 ± 10.05</td>
<td>0.74</td>
</tr>
<tr>
<td>Mental health</td>
<td>72.5 ± 25.3</td>
<td>68.5 ± 17.6</td>
<td>0.25</td>
</tr>
</tbody>
</table>

Bladder/urethral support

The application of pelvic osteotomy in adult female patients with exstrophy: applications and outcomes
Mohd F. Anuar, John P. Beartfort*, Reinwardt Maximiliaan Corvelloia* and Paul P. Hamberger* 2011 BJU INTERNATIONAL | 110, 209-212

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

PROCEDURES FOR CONTINENCE

OUTLET

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

• Bulking – vesicoscopy
• Bladder Neck - vesicoscopy
First Option: Bulking Agents

- moderate improvement
- decreasing with time
- repeatable
- allows to wait

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 – vescicoscopy
- • Bladder Neck - vescicoscopy

ARTIFICIAL SPHINCTER

BLADDER NECK CLOSURE

PROCEDURES FOR CONTINENCE

OUTLET

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

Fascial sling

A

Normal Urethra

Incompetent
Urethra

B

The sling
supports and
partially
compresses the
urethral lumen

C
**MINI SLING**

Reservoir & Outlet Surgery

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

**PROCEDURES FOR CONTINENCE**

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

- simultaneous ?
- 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)

ROBOTIC PAEDIATRIC UROLOGY: FOR CONTINENCE?
sling
Bladder neck
Reimplant
Mitrofanoff

Beyond Pediatric Incontinence: The Challenges of Transitional Care
June 6-8, 2013 | Toronto, Canada

THANK YOU
Grazie
Arrivederci a Roma

Bambino Gesù Children Hospital
Roma
Let’s meet in Rome!
SURGERY FOR CONTINENCE
Giovanni Mosiello

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

- 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 becomes poor, and the responsibility for identifying late complications often rests upon the primary care provider.

Review Article
Adult Care of Children From Pediatric Urology
Christopher M. Wiedemann, Gyan P. Bahl, Michael M. Youssef

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

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

Long-term continence care of Complex Congenital Malformations

- Spina Bifida and Neurogenic bladder (M and F)
- Anorectal Anomalies (M and F)
- Exstrophies/Episipadias (M and F)
- Urogenital Sinus –CAH (F)
- Ambiguous Genitalia (M and F)
- Vaginal Anomalies (F)
- Urethral anomalies and trauma (M)
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: baclofen pump,
  Ventriculo-peritoneal derivation

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/COLONS/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’


---

**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

---

**Surgical complications of bladder augmentation: comparison between various enterocystoplasties**

Ricardo Gonzalez 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

---

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

2000: 792 Ac
2009: 595 Ac

**WHY? BOTOX effects?**


---

**Surgical Techniques in Urology**

Pediatric Laparoscopic Ileal Cystoplasty: Complete Intracorporeal Surgical Technique

R. Lopez-Diaz, A. H. Paynter, and M. O. Baskin

Surgical Techniques in Urology
Pediatric Robotic-assisted Laparoscopic Augmentation Ileocystoplasty and Mitrofanoff Appendicostomy: Complete Intracorporeal—Initial Case Report

Ricardo Gonzalez, Michael K. sq, W. Stuart Hendren, and Steven R. F. D. L. B.

---

2017: We don’t know anything about complications of lap and robotic
Preoperatively, nearly three-quarters of the patients had upper tract changes consisting of VUR and/or hydronephrosis.

Augmentation alone or with reimplantation???

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

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

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

The good short-term results of detrusorectomy generally remain unchanged at longterm follow-up.

Detrusorectomy reduces the need for augmentation and use of antimuscarinics in children with neuropathic bladders.
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, and filling pressure decreases.


Level of evidence 4
Grade of recommendation C

Ureterosigmoidostomy
Level of evidence : 3
Grade of recommendation: B

This type of continent urinary reconstruction may be utilized in reconstruction for bladder extrophy, 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

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.

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

Conclusions: This study demonstrated that local restorative bladder capacity, maximum detrusor pressures and detrusor side pressure increase significantly in patients with myelomeningocele following puberty. The increase in bladder capacity could be attributed to increasing bladder outlet resistance resulting in decreased voided urine to males and unilateration in females. A significant number of patients spontaneously achieve continence at puberty, and continence becomes more likely when increased local cystometric bladder capacity is not associated with an increase in maximum detrusor pressure. Finally, no significant postpubertal open urinary tract incontinence was observed in our series.
More neourethra is longer and narrower, more likely patients will be dry but they will have problems catheterizing neourethra.

If we change the outlet we change the bladder.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Number of Patients</th>
<th>Lower tract changes (%)</th>
<th>Upper tract changes (%)</th>
<th>CKD (%)</th>
<th>Upper tract changes, subsequent (%)</th>
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</thead>
<tbody>
<tr>
<td>BNR</td>
<td></td>
<td>14 (38)</td>
<td>6 (17)</td>
<td>17 (42)</td>
<td></td>
</tr>
<tr>
<td>BNR + AC</td>
<td>14 (38)</td>
<td>13 (36)</td>
<td>13 (36)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>BNR + UD</td>
<td>20 (56)</td>
<td>20 (56)</td>
<td>18 (50)</td>
<td>10 (26)</td>
<td></td>
</tr>
<tr>
<td>BNR + UD</td>
<td>17 (48)</td>
<td>17 (48)</td>
<td>17 (48)</td>
<td>11 (29)</td>
<td></td>
</tr>
<tr>
<td>BNR + UD</td>
<td>28 (75)</td>
<td>18 (48)</td>
<td>10 (26)</td>
<td>13 (36)</td>
<td></td>
</tr>
</tbody>
</table>

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

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.

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 minimally 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

Defecation disorders in children
- Constipation / fecal incontinence
  - Functional: 95%
  - Organic: 5%
- Constipation: > 90-95%
- Non-retainive fecal incontinence: <5-10%
- Anorectal malformations
- Spinal problems
- Miscellaneous

Background
- There is evidence of long-life treatment of BD in these pediatric patients: Spina Bifida, Anorectal Malformations, Hirschsprung Disease.

BBD
- Altered bowel and bladder control is common in children, mainly, in those with disabilities. In these patients, management of the two problems has 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.

Anorectal Malformation
- Genitourinary Anomalies: 40%
- Sacral anomalies: 25%
- LUT Dysfunction: 15%
NB in Children
Bowel Dysfunction - Treatment

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

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 children and young people with constipation or fecal incontinence due to NBD (n=346 children). Transanal irrigation is reported as improving quality of life and family’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.

(Choi EK Spinal Cord 2013)

(Choi EK Spinal Cord 2013)

(Choi EK Spinal Cord 2013)

(Choi EK Spinal Cord 2013)
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

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 QoL of patients and care givers

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