Aims of course/workshop
Spina bifida, bladder extrophy/epispadia, posterior urethral valves are managed immediately after the birth with a surgical procedure, but all these patients require lifelong urological care for the treatment of continence.

Learning Objectives
After this workshop participants should be able to:
1. Correct management in childhood to avoid procedure that impair adult life aspects,
2. Common knowledges in pediatric and adult health care professionals about adult life problems and congenital pathologies respectively
3. Transition out of childhood: who should manage the mature pediatric urology patients?

Learning Outcomes
After the Workshop participants will be able to:
1. Have knowledge about congenital pathologies impairing continence
2. To understand the treatment performed in childhood
3. To recognize concerns during adolescence and young adult age
4. To ameliorate a clinical practice for the transitional care of continence

Target Audience
Pediatrician, Pediatric Surgeon, Pediatric Urologist, Nurse, Physiotherapist, Surgeon, Urologist

Advanced/Basic
Advanced

Conditions for learning
This is a Committee workshop, open to all delegates

Suggested Learning before workshop attendance
Suggested Reading

- Is a closed bladder neck on preoperative videourodynamic studies an important factor for continence following augmentation ileocystoplasty in myelodysplastic patients? Ghanem MA, van Denhoek J, Nijman RJ. J Pediatr Urol. 2013

Kim Kwang
Spina Bifida
The management of neurogenic bladder dysfunction in children has changed over the years. The introduction of clean intermittent catheterisation (CIC) has revolutionised the management and today the conservative management is a very successful treatment option.

Standard treatment is CIC with anticholinergics: children do not have upper tract deterioration when managed early with IC and anticholinergic medication. CIC should be started soon after birth in all babies, especially in those with signs of possible outlet obstruction Furthermore the early initiation of CIC in the newborn period makes it easier for parents and for children to accept it. Early management results in reduced renal and bladder damage and in neurogenic bladders that are refractory to anticholinergics, injection of botulinum toxin into the detrusor seems to be effective to avoid or postpone during childhood surgery. Children with neurogenic bladder have disturbances of bowel function as well as urinary function. Bowel management, especially transanal irrigation is effective.

Posterior urethral valves
PUV are one of the few life-threatening congenital anomalies of the urinary tract found during the neonatal period. Antenatal treatment of PUV remains controversial. In a newborn with suspicion of PUV the first act is then to provide bladder drainage. This can be achieved by a 6 or 8 Fr. feeding tube or Tieman or Foley catheter inserted in the urethra as an alternative, a sovrarepubic 5 Fr epicystostomy can be used. Today endoscopic valve ablation is the standard treatment with an endoscopic incision or resection with Valve ablation, and it is important in this maneuver to avoid extensive electrocoagulation, because the most common complication of this procedure is stricture formation.

Vesicostomy is used when the child is too small or in severe general condition. Otherwise, a cutaneous vesicostomy provides an improvement or stabilisation of upper urinary tracts although there has been concern that a vesicostomy could decrease bladder compliance or capacity High diversion should be considered if bladder drainage is insufficient to drain the upper urinary tract. Life-long monitoring of these patients is mandatory, as bladder dysfunction is very common and continence is a major problem.

Up to 75% of boys with PUV show abnormal bladder dynamic. Urodynamic studies allowed to identify different patterns of bladder dysfunction changing during time in boys with PUV: bladder overactivity; low compliance high voiding pressure and myogenic failure.

Bladder extrophy – epispadias complex (BEEC)
Is one of a major challenges in paediatric urology. The modern approach is: 1) staged repair of BEEC consists of three distinct operations: closure of the bladder, posterior urethra and abdominal wall at birth; reconstruction of the epispadic urethra during infancy; and bladder neck reconstruction in early childhood. 2) one stage reconstruction, or complete repair. 1. The initial staged approach to functional bladder closure includes bladder, abdominal wall, and urethral closure in the newborn period with bilateral osteotomy, epispadias repair at 6 months to one year of age; and bladder neck reconstruction along with antireflux procedure later at 4 to 5 years. Different types of pelvic osteotomy have been suggested and osteotomy, is still debated for the different ages. Osteotomy anyway gives some advantages: reduces abdominal wall tension, placing the urethra in a deeper plane reinforcing outlet resistance and supporting bladder neck. 2 Complete Repair combines standard bladder closure with the “penile disassembly” technique for epispadias repair at the same time with the aim to reduce the number of procedures required for reconstruction and potentially improving continence without the need for formal bladder neck reconstruction. This procedure leaves a high percentage of patient 60% with hypospadia. The long term results of the 2 techniques are difficult to compare because is very common that every single centre choices one of these according to the training of the single urologist. Aim of surgical management of epispadias: is providing a satisfactory cosmetic appearance, as well as normal genital function and preservation of fertility, and functioning urethra and continence.

**Giovanni Mosiello**

CIC concerns

Conservative management of either or both the bladder and sphincter complex involve pharmacologic management, intermittent catheterization, bowel management. The objectives of these non-surgical strategies are to preserve renal tract function primarily, optimise quality of life and promote independence of self-care (de Jong 2008). Achievement of continence is of secondary importance to preservation of renal function. In the adolescent population some concerns are evident.

Regular evaluation of adherence to medication and CIC is mandatory. CIC must be performed with largest possible catheter: includes instruction and review of adequate hand hygiene, perineal hygiene, catheter cleaning, insertion of catheter without contamination, optimal interval between CIC. Any mental impairment or physical difficulty limiting self-care should be considered. Overnight catheter drainage could be considered in some clinical situations, as well as suvrapubic catheter (bogram cistostomy).

Critical point could be the CIC refusal by adolescents as well as the parents refusal to accept “baby independence “starting a self CIC training program. Treatment and prevention of urinary tract infections: bacteriuria is usual in patients performing intermittent catheterization but may be ↓ by improving hydration and more frequent catheterizations.

Surveillance is mandatory during puberty as bladder capacity, maximum detrusor pressure and leak point pressure may increase after puberty.

Indications for surgical therapy could be related to arm function that preclude self CIC. Physical weight of child makes wheelchair transfers difficult, necessitating a catheterizable stoma as for preserving patient privacy in young people where caregiver is not a parent. If hydrophilic single use catheter is worldwide recommended, concerns are present in different geographical setting due to economical reason. Critical point remains lack of education and trained health care professionals.

**Sacral NeuroModulation**

From the first description in 1988 (Tanagho EA, J Urol) a significant number of reports have been published, and SNM became rapidly a well-accepted treatment in adults, and was approved by the Food and Drug Administration for the use in urology in 1997 for treatment of urge urinary incontinence, in 1999 for treatment of urinary urgency-frequency and nonobstructive urinary retention, and in 2011 for fecal incontinence. (Herbison GP 2009, Kessler TM 2007, van Kerrebroeck PE 2007). SNM is used in neurogenic bladder dysfunction (NBD) too, and in a systematic review, Kessler et al, analyzing 26 independent studies stated that there is evidence indicating that SNM may be effective in adults with NB, but it is still not possible to draw a definitive conclusion. (Kessler et al 2010). It is interesting to see in this review that on the 565 evaluated reports, 34 papers only were assessed for eligibility, because the other 531 were not referred to neurogenic LUTS.

The obvious conclusion is that SNM is widely accepted and used in adults for refractory non obstructive chronic urinary retention, urge incontinence, urgency-frequency syndrome, and in some cases has been used in NBD. For this reason is surprising that the first prospective randomized controlled study to evaluate the possible benefits of SNM in children has been performed in 2004 in NBD (Gyus), because before that, not considering the pioneering Tanagho experience (Tanagho 1992), only some sporadic pediatric cases have been reported in adults series. Actually SNM is not a first-line treatment but rather as a second or better third line treatment for the patients who have failed conservative treatments. During the past years the technique of SNM has become less invasive, more safe, reliable and effective, with the technical improvements. The re-operation and complication rates decreased significantly. The clinical results have led to expanding indications. SNM are mainly used in children and young adults for overactive bladder (OAB), Non-obstructive urinary retention, NOUR, interstitial cystitis, pelvic pain, NBD.

**Giulio Del Popolo**

OnabotulinumtoxinA (onaBNTa)
This treatment of neurogenic detrusor overactivity (NDO) is widely accepted after it has received the regulatory approval for this specific use in adults. International Guidelines support the use onaBNTa in patients refractory to oral treatment. Although the administration of onaBNTa is still considered off-label in children, data on its efficacy and safety have already been reported and phase III clinical trials are ongoing on this population. Therefore, nowadays there is high quality evidence for the efficacy of detrusor injections of onaBNTa in adults with NDO and in children and young people with myelodysplasia. However, there is still a lack of standardized protocols of treatment for NDO with onaBNTa supporting the patients in their transition from the childhood to the adult age.

Recommendations for best practice for using onaBNTa in transitional care

Patients’ selection and assessment before treatment

1. Neurogenic diagnosis
2. Data of last urodynamic investigation, ultrasound, laboratory tests (blood and urine examinations)
3. Previous treatments for NDO and previous onaBNTa treatments; regarding last onaBNTa treatment: date, dose, technique of injection, possible side effects, efficacy.

Patient evaluation (clinical assessment):

Diaries

Instrumental evaluation:

Kidney and bladder ultrasound is mandatory, and recommended the use of cystometry, while videourodynamic investigation and cystography can be considered optional and used in selected cases depending on the clinician’s opinion.

QoL test

QoL assessment has to be included in the general evaluation of the patient. Among various tools available, the Qualiveen-short-term test, a 10-items questionnaire, has to be preferred.

Antibiotics

Antibiosis must start 1 week before treatment in case of positive laboratory examinations. If urine examination and urine culture are negative, only perioperative antibiotics may be administered. Commonly 3rd generation cephalosporins can be administered or quinolones. The use of aminoglycosides is contraindicated.

Self-catheterization training

As widely known, is mandatory before onaBNTa treatment that patients understand and accept the risk of urinary retention and potential need of catheterism. For this reason, a demonstration of this practice or a specific training is recommended.

Anaesthesia

Younger individuals may be treated under sedation; spinal anaesthesia is not recommended.

Cystoscope

Type of cystoscope is usually selected by the operator; therefore flexible or rigid cystoscope may be used.

Doses

As reported in Literature, it is well known that in younger individuals a dose of 10U/kg is administered, until 200U. Evaluating previous dosages administered to the patient, the first re-treatment should repeat the same amount of onaBNTa. As suggested by common practice, starting from 200U sub-divided in 30 injections sites, 1 cm apart one from the other, should be adopted.

Procedure technique

As recommended for children and adults, injection site is represent by the detrusor muscle. Also in transitional care, the trigone might be infiltrated only in those individuals who show to be non-responders to standard approach. It is mandatory the use of needles specifically designed for onaBNTa infiltration, 23 G and 4 mm of deep, to avoid onaBNTa diffusion outside from the bladder.

3) Post-treatment care

Being individuals of transitional care still young, it is advisable to take particular care of these subjects after treatment. Possibly, is recommended to treat early in the morning and allow patient to stay in the hospital some hours after onaBNTa injection. Intravenous fluid may be administered especially in patients submitted to general sedation.

4) Follow up and re-treatment

After discharge, patient has to be re-evaluated within 7 days by clinical assessment. In case of spinal disease, it is recommended a cystometric evaluation within 1 year. Re-treatment should be based on patient’s request but also on objective measures.
Rien Nijman
Bladder augmentation
The indication for bladder augmentation, replacement of the bladder, or the creation of a continent urinary diversion, is either the morphological or functional loss of normal bladder function. The main goal of this surgery is to relieve high pressure and low capacity of the urinary bladder and create a new reservoir with low storage pressures that can be emptied periodically. It is particularly important that the patients understand that spontaneous voiding will not be possible after such surgery and life long intermittent catheterization will be required.

There are several important principles for bladder augmentation and replacement that should be respected:
• Use the minimal amount of bowel
• A low-pressure large capacity reservoir is essential (this requires detubularization of any intestinal segment used)
• A reliable continence mechanism (continent urinary outlet) must be assured
• Because of the only resorbable sutures and staples should be used (risk of stone formation)

The invasiveness of Enterocystoplasty, and its long-term severe complication rate, has greatly reduced its indication. Recently, it is gaining more attention, in relation of the availability of mini-invasive procedures, i.e. the robotic-assisted laparoscopy. 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 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.

Autoaugmentation
The principle of auto-augmentation of the bladder is the excision of a great portion of the detrusor while leaving the urothelium intact, creating a large diverticulum for the storage of urine at lower pressures. This urine stored at a low pressure can be drained by intermittent catheterization. The theoretical advantages of this procedure are the low complication rates of the surgery, reduced operative morbidity with shorter stay in the hospital, absence of urine salt resorption, less mucous production in the urine and possibly absence of carcinogenic potential.

More recently, some authors have proposed the laparoscopic auto-augmentation as a minimally invasive procedure for the treatment of low capacity / low compliance bladder.

Catheterizable channels
In the long term it is necessary to have a catheterizable channel. Mitrofanoff’s name is given to the principle of burying a narrow tube within the wall of the bladder or urinary reservoir whose distal end is brought to the abdominal wall to form a catheterizable stoma suitable for intermittent catheterization. The technique is simple and familiar to all urologists who are accustomed to re-implanting ureters. Several narrow tubes are available for the Mitrofanoff conduit. In the original description, the appendix was used. The system achieves reliable continence (90-100%) which is maintained in long term follow-up, for a high proportion of patients creating an abdominal continent catheterizable stoma into the bladder is a good option when urethral catheterization is impeded. Up to 21% of patients will have problems related to stoma leakage or stenosis within the first 2 years of its creation and require minor revision. An antegrade continence enema stopper effectively eliminates stomal stenosis. Patients with good manual dexterity and fine motor ability gain a greater ability to self-care and appreciate the privacy gained from not having to expose their genital area to a caregiver. In wheelchair-bound girls dependant on CIC, a catheterizable channel obviates transfers in the bathroom. Stoma creation in male patients only follows difficulty with urethral catheterization. As extreme weight gain can cause adjacent skin to partially obscure a catheterizable channel, postpubertal patients should be educated in nutrition and portion control. 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. To date, patient compliance with longterm CIC appears to be good and is associated with preservation of the upper urinary tract.

Derivation
Ileal conduit (‘wet deviation’) is no longer indicated except in case of severe mental impairment or severe renal dysfunction and no options for bladder reconstruction. Bladder replacement instead of augmentation may be appropriate in cases of bladder extrophy where use of native bladder tissue is impossible.

Enrico Finazzi Agrò
Surgery for Continence

Many surgical approaches have been described for increasing bladder outlet resistance to achieve continence, however, long-term results are lacking.

Bulking agents

The injection of bulking substances in the tissues around the urethra and bladder neck to increase outlet resistance in children dates back to at least 1985. The search for safer, biocompatible substances to create periurethral compression has first led to the use of cross-linked bovine collagen, with initially reported success in about 20-50% of children.

Usually the substance is injected endoscopically in the bladder neck area (finding the best spot is often the most difficult part of the procedure): more than one procedure may be necessary. On average 2.8 – 3.9 ml is injected.

Fascial Sling

The technique involves suspension of the bladder neck with an autologous fascial strip or artificial material secured to the rectus fascia or the pubic symphysis. It is believed the mechanism of action involves coaptation of the bladder neck due to traction, and/or elevation of the urethra to an intra-abdominal position, which increases tension on the bladder neck with abdominal straining. Complication rates are modest and include difficult catheterization and rectal injury, while in long-term erosions or persistent incontinence may occur.

Bladder Neck Closure / Reconstruction

In ‘desperate’ cases the bladder neck may be closed, the indication being persistent leakage despite several attempts to enhance outlet resistance. Long-term results are usually disappointing: persistent urinary leakage, stomal stenosis and leakage or stone formation.

The optimal bladder neck procedure should increase bladder outlet resistance at minimal cost of decreasing bladder capacity, maintaining easy catheterization and still allowing some leakage at high pressure in order to protect the upper urinary tract. Different operative techniques with the aforementioned aims have been used with varying outcomes.

Artificial Urinary Sphincters

Many surgeons are reluctant to implant an AUS as it consigns patients to further revision surgery, and the potential risk of deterioration in bladder function and a concomitant deleterious effect on upper urinary tract drainage. However, with improved durability of newer models that have an average life span of about 8 years, revision rates have become less. The ideal patients for AUS implantation are post-pubertal males or females, who can void volitionally and empty the bladder completely. On the contrary, a common problem is the development of reduced bladder compliance with time. Overall, 40 to 50% of neurogenic patients require a bladder augmentation concomitantly or subsequently to the AUS implantation.

Evaluation of Outcome

Our prejudice is that reconstruction does, indeed, improve the lives of children, Quality of life does not mean absence of disease or a level of complications acceptable to the reviewing clinician. It is a difficult concept to measure because lack of validated instruments, difficulties in translating from one culture or language to another, of the difficulties in selecting control groups and variations in clinical situations.

Conclusions

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%. In boys preservation of erectile function after a fascial sling procedure can be expected. Synthetic suburethral slings can only be used in a tension free mode, due to risk of erosion. 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, suggesting its use in a very young population should be tempered. Injection of a bulking agent into the bladder neck area as a primary treatment of bladder outlet incompetence is not recommended because of low success rates. Following insertion of the artificial urinary sphincter, efficacy rates for complete dryness between voids vary between 56% and 91%. The revision rate is high, about 1/3 require reoperation and 19% device removal due to erosion. Long-term survival (>10 years) of the prosthesis is up to 60%.

Approximately half of the individuals able to empty before insertion of the artificial sphincter can do so afterwards, however, bladder dynamics can change postoperatively. 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.
Concerns in Adolescents with standard Treatment: CIC and Drugs

GIOVANNI MOSIELLO
MD, FEAPU, FEBPS

Disclosures
Italian Society of Pediatric Urology
Other: Board Member
Italian Society of Urodynamics
Other: Chair of Children’s Committee

Trial participation
Pfizer
Allergan
Consultant
Coloplast
Wellspect
Medtronic

Definition
• Has to be considered also that, children and families are accustomed to the holistic care received in a paediatric setting where several aspects of education, social care or family support are covered
• In this setting they should feel vulnerable when approach the adulthood
• 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
• These conditions have a major lifelong implications and should require a bladder drainage mechanism

Definition
• Poor medications adherence is common in children and adolescents with chronic illness
• Second the world health organization, adherence is defined as “The extent to with a person behaviour corresponds with agreed recommendations from a healthcare provider”
• Medications adherence refers to the degree to with the medication taken reflect the prescriber’s intention
Introduction

- Adolescence is a crucial moment of growing characterized by physical, and cognitive maturation as well as psychosocial changes, and identity formation.
- This time they acquire also the independence from their parents starting to make own choices.
- Having a chronic disease create a challenge to socializing.
- It is important to do not be different from their peers.

Having diseases may led to feeling failed.

- The way to gain control: not take their medications, missing appointments or not following dietary restrictions making adherence very difficult.

Different studies in literature analysed the degree of adherence in adolescents in different pathologies such as medications for asthma or insulin injection reporting an adherence rate ranging from 10-85%.

- Has seen a correlation between age and medication adherence with major prevalence around 11yrs peaking during mid-adolescence.

It is important to consider different domains of adolescent development including social, emotional and cognitive.

CIC in neurogenic bladder

Anxiety disorders and depression is common in patients with chronic illness, especially regarding neurogenic bladder dysfunction.

- Kabra at al., screening adolescents with neurogenic bladder for depression and anxiety, found a high risk of anxiety in this group of patients and anxiety/depression in the caregivers.
- In the other hand, Borzykowski at al. in 2004 analysing the social and psychological impact of CIC in children and adolescents with NB and this parents, shown that CIC or self-catheterization itself did not cause major emotional and behavioural problems.

The acceptance of CIC treatment decreases when the child gets closer to adolescence. Identifying problems and complications related to CIC in adolescence: retrospective study, median age of 16.5 yrs.

- Results: one of the main problem associated with self-catheterization during adolescence is non-compliance with treatment. A poor CIC routine can cause U11s.

For further reading, please refer to the sources listed in the references.
How to identify low adherence

**Direct**
- Assessment of blood drug metabolite (not available for all conditions)
- Pills count
- Electronic monitoring devices

**Indirect**
- Constructive dialogue with adolescents
- Is advised to do not be directly confronting adolescent and avoid being judgmental
- With questions investigating side effect of medications, the time that patients take the medications, if is hard or not to follow all recommendations

Low adherence should be identified by indirect or by direct methods

How to promote adherences

**Education intervention**
- Providing verbal or written information about illness, treatment benefit and the importance of follow the medication
- Studies in literature showing that education program should done by physician clinic visits, telephone contact, by video, web, book or by home visits with an high adherence comparing to the control standard approach.

Conclusion:
- Low adherence increases morbidity and medical complications, worsening the quality of life and increasing unnecessary medical consults and investigations with high healthy cost.
- Involvement of families on daily medications, having close friends and a good family relationship, should influence positively the medications adherence, in contrast has seen that family conflicts led to low medication adherence
Consistent long-term efficacy and safety of onabotulinumtoxinA in patients with neurogenic detrusor overactivity: final results of repeated treatments up to 4 years

Giulio Del Popolo1, Gilles Karsenty2, Heinrich Schulte-Baukloh3, Roger Dinchowski4, Karen Ethans2, Brenda Jenkins2, Steven Guard5, Yan Zheng2, Michael Kennelly6

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2. Montreal University, Montreal, Canada
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6. Allergan, Ltd., Morris, NJ, USA

Aim of the study

Aim of the study
- To assess efficacy and safety after repeated onabotulinumtoxinA treatment of UI due to NDO
- Here we present final results from the multicentre, long-term study in which patients received multiple treatments for up to 4 years

Study design

1 year
- Multiple treatments, no washout based on patient request and fulfillment of prespecified treatment criteria (minimum of 6 weeks since previous treatment; 21 UI episode within 4 days)

OnabotulinumtoxinA 200U had a duration of effect of ~9 months and was well tolerated with no new safety signals over 4 years

Results were similar with onabotulinumtoxinA 300U

OnabotulinumtoxinA 200U consistently reduced urinary incontinence and improved volume/void over 4 years*
**Patient Selection**

**Pre-treatment assessment**

1. Patient's selection and pre-treatment assessment
2. Treatment (technique, dose, management)
3. Post-treatment and follow-up.

**Antibiotic prophylaxis**

- Anesthesia - sedation
- Cystoscope - rigid or flexible
- Maximum dosage: 200 U.I.
- Injection site: into the detrusor muscle
- Needles: 20-23 G and 2-4 mm deep
- Postoperative antibiotics prophylaxis is recommended

**Follow up**

3 and 6 months
3 days voiding diary and QoL
Urodynamic, creatinine serum and ultrasound
**Re-treatment**

every 6 to 9 months
Outcomes of Intra-Fascial Injections of Botulinum Toxin in Neck Muscles: A Systematic Review

All injections were performed under general anesthesia in the depth of the cervical muscles using a 25–40 cm needle. The injection sites were as follows:

- 2 cm to 3 cm lateral to the midline of the neck
- 2 cm to 3 cm inferior to the mandible
- 2 cm to 3 cm posterior to the ear

The average number of injections per patient was 3.5, with a range of 1 to 6. The follow-up period was 6 months to 12 months post-injection.

Efficacy?
1) 6 months
2) 8 months
3) 9 months
4) 12 months

Dosage OnaBot/A?
1) 100 I.U.
2) 200 I.U.
3) 300 I.U.
4) 400 I.U.

Approved for?
1) MS
2) SCI
3) All neurological pts
4) MS and SCI

Note: Reactions in cases treated with OnaBot/A have been reported to include:

- Headache
- Increased muscle tone
- Improved range of motion

Rare cases of botulism-like symptoms have been reported, including:

- Paralytic ileus
- Respiratory failure

Intrafascial injection of OnaBot/A is contraindicated in patients with neuromuscular junction disorders, as it may lead to prolonged paralysis.

Thank you!
Bladder augmentation, when and how?

Rien JM Nijman, FEAPU, Dept Urology and pediatric Urology University Medical Center Groningen, The Netherlands

ICS Tokyo 2016

History of bladder reconstruction

<table>
<thead>
<tr>
<th>Year</th>
<th>Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1852</td>
<td>Ureterosigmoideostomy (bladder exstrophy)</td>
</tr>
<tr>
<td>1888</td>
<td>Coffey R.C. Ureterosigmoideostomy</td>
</tr>
<tr>
<td>1894</td>
<td>Maydl K. The trigono-sigmaoideoplasty (bladder exstrophy)</td>
</tr>
<tr>
<td>1898</td>
<td>Gersuny R. Rectal reservoir (rectal sphincter)</td>
</tr>
<tr>
<td>1950</td>
<td>Bricker Cutaneous incontinent diversion</td>
</tr>
<tr>
<td>1982</td>
<td>Kock et al. Continent ileal reservoir</td>
</tr>
<tr>
<td>1980</td>
<td>Mitrofanoff Continent appendicovesicostomy</td>
</tr>
</tbody>
</table>

Function of normal and reconstructed bladder

- To store urine within physiological bladder capacity at low pressure and without leakage
- To evacuate urine without residual at socially accepted intervals
  - normal voiding
  - intermittent catheterization

Bladder reconstruction: indications

- End-stage bladder diseases
  - PUV
  - MMC
  - Tumors (Rabdomyosarcoma)
  - Exstrophy / Epispadias
  - Bilateral ureteral ectopia
- Urinary reconstruction may be necessary
  - Choice of tissue: Ureter / Bowel / Tissue engineering?
  - Outlet channel: Appendix / Monti tube / other

Bladder reconstruction: techniques

- Many indications, many techniques
- Augmentation with ileum / ureter / auto augmentation / seromuscular lined colon....
- Replacement of the bladder (Mainz pouch / Indiana pouch....)
- In combination with bladder neck enforcement / AMS sphincter prosthesis / BN plasty / sling....
- In combination with catheterizable channel

- Mostly used: ileocystoplasty + channel (appendix)
- +/- BN reconstruction / sling / AMS
- +/- BN closure
- Alternatives: personal preference
Bladder reconstruction: when

- When other therapies fail
  - Anticholinergics
  - CIC
  - Botulinum toxin A
  - Neuromodulation
- When renal function is endangered
  - DSD / severe OAB / high pressures
- When the child is ready for it (counseling is extremely important)
- When the parents / care givers are ready
- For improvement of QoL

Bladder reconstruction: typical patient

<table>
<thead>
<tr>
<th>MMC</th>
<th>refractory incontinence / noncompliant bladder / +/- sphincteric incompetence / VUR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>&gt; 7 yrs (any age)</td>
</tr>
<tr>
<td>Therapy</td>
<td>counseling 2 x 1 hr ileocystoplasty + BN enforcement + Mitrofanoff (umbilicus / R lower quadrant)</td>
</tr>
<tr>
<td></td>
<td>MACE rarely done: bowel irrigation with Peristeen system is usually preferred</td>
</tr>
</tbody>
</table>

Neuropathic bladder in children

continent diversion

Independent
Less time consuming
Improves self-esteem

Bladder augmentation

Possibilities
- Intestinoplasty (clam cystoplasty)
- Gastrocystoplasty
- Ureterocystoplasty
- Autoaugmentation (detrusorotomy, detrusorectomy)
- Autoaugmentation with use of seromuscular segment of sigmoid or stomach
- (Tissue engineering)

Objective: increase capacity /
improve compliance: lower the pressure

Clam cystoplasty

Detubularization

- Bowel can generate up to 60-100 cm H2O
- opening the bowel on its antimesenteric border...and subsequent reconfiguration into spherical shape
detubularization

Ileocystoplasty
- The least contractile tissue
- 20-30 cm of ileum, resection ends 15-20 cm proximal to the ileocaecal valve
- Reconfigured into a U shape or S, or W

Sigmoid cystoplasty
- redundant and dilated in spinal dysraphism, adjacent to the bladder
- 15-20 cm segment
- detubularization and reconfiguration (into U or S)

Ureterocystoplasty
(Belling 1993)
possible in patients with a massively dilated ureter
Opened ureter is reconfigured to create a spherical reservoir

Auto-augmentation

Autoaugmentation with seromuscular segments
- demucosalized sigmoid or gastric segment (with urothelial lining or without)
Autoaugmentation with seromuscular segments - results

- Tiedious procedure, continent stoma not feasible
- Long-term results excellent in 90% of patients (10% failure only - with ileal segments)
  - 329% increase in bladder capacity
  
  Lima et al. 2004

Long-term follow-up cystoplasty

- Ultrasonography
- Serum creatinine, electrolytes, Ph, PCO2 etc
- Urodynamics
- Cystoscopy (yearly at 10 years postoperatively ??)
- Bone density?

Stones in a reconstructed bladder risk factors, incidence

- Urinary stasis
- Abnormal urine pH
- Bacteriuria (urea-splitting)
- Mucus production
- Poor bladder emptying (Continent stoma!!)
- Foreign body (staples)
- Pre-existing history
  - Gastrocystoplasty (uric acid st.) 2 - 9 %
  - Enterocystoplasty 12-50 %
  - Ileocolonic cystoplasty 19 %

Long-term results bladder augmentation

- Secondary procedures 21 %
  - Stones / stomal stenosis
  - Capacity / leakage
- Tertiary procedure 9 %
- More than 4 procedures 4 %
- Good result after primary procedure 66 %
  - Continent / no stenosis / CIC 4-6 dd

At 10 years follow-up: 66 % → 52 %
About half need more than 1 procedure!!

Other complications

- Mucus production (irrigation each day)
  - Hussman: daily irrigation with 250 ml saline significant reduction in UTI / stones
- Urinary tract infection...irrigate!!
- Spontaneous bladder perforation (most concerning, in 4.5-16.5 %, CT)
- Malignancy in a reconstructed bladder (mostly in the mixture of urinary and fecal streams, latency period, endoscopy beginning (10 years after op??)

Augmentation / complications
**Long-term results bladder augmentation**

N=203 adult SB patients (Mayo Clinics), 2015
- alcohol abuse 12 %
- drug abuse 16 %
- non-compliance CIC 6 %
- bladder perforation 2 %
- mental retardation n=36
  - non-compliance CIC 16 %
  - bladder perforation 25 %

- Study Mayo clinics 2015: 78 adult SB patients
  - all augmented + outlet procedures + cont. channels
  - all presented with bladder stones + were treated
- 3 groups: A. irrigation with 60 ml saline daily
  B. irrigation with 120 ml saline daily
  C. irrigation with 240 ml saline daily

- Noncompliance: missed 1 per week
  - A. 13 %
  - B. 15 %
  - C. 20 %

- UTI's + bladder stones in adult Spina Bifida patients
  - Prospective study: after 5 yr

- 60 vs 240 ml P < 0.05
- Metabolic abnormalities 91% / hypocitraemia 75% (rec. stones 100%!!)
- Upper tract stones 13%

- Conclusion:
  - Daily irrigation with 240 ml !!!!!
  - use active irrigation + syringe
  - tab water as effective !! + cheap

**Long-term outcome bladder augmentation**

80 high risk SB patients
- normal kidney function at transfer
- 32 developed new scars
- 10/32 silent scars
- 12/32 > stage 3 CRF !!
- 15/32 renal stones

⇒ 12/80 = 15 % progressed to > stage 3 CRF !!
Long-term results bladder augmentation

- SB patients are different from other patients with congenital abnormalities
  - they do not adhere to CIC at regular intervals
  - transfer to adult urology needs a lot of attention + special care / facilities
  - life-long follow-up!!!
  - cognitive function is impaired!!!

It is not that they don’t want to do it, but they just won’t

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Augmentation cystoplasty patient characteristics.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number</td>
<td>2831</td>
</tr>
<tr>
<td>Number with follow-up</td>
<td>204 (75%)</td>
</tr>
<tr>
<td>Median years of follow-up</td>
<td>3.3 (1.5–6.1)</td>
</tr>
<tr>
<td>Median number of encounters</td>
<td>5 (3–10)</td>
</tr>
<tr>
<td>Race</td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>46.8%</td>
</tr>
<tr>
<td>Black</td>
<td>7.8%</td>
</tr>
<tr>
<td>Hispanic</td>
<td>18.8%</td>
</tr>
<tr>
<td>Other/race not stated</td>
<td>26.7%</td>
</tr>
<tr>
<td>Diagnoses</td>
<td></td>
</tr>
<tr>
<td>Spina bifida</td>
<td>55.1%</td>
</tr>
<tr>
<td>BEEC</td>
<td>12.4%</td>
</tr>
<tr>
<td>UUTD</td>
<td>2.9%</td>
</tr>
<tr>
<td>CAM</td>
<td>4.5%</td>
</tr>
<tr>
<td>NB</td>
<td>13.8%</td>
</tr>
<tr>
<td>Other</td>
<td>11.2%</td>
</tr>
<tr>
<td>Mean age in years at AC</td>
<td>9.1 (6.7)</td>
</tr>
<tr>
<td>Bladder neck surgery at AC</td>
<td>16.8%</td>
</tr>
<tr>
<td>Catheterizable stoma at AC</td>
<td>39.3%</td>
</tr>
<tr>
<td>Median LOS in days</td>
<td>8 (6–10)</td>
</tr>
</tbody>
</table>

AC = augmentation cystoplasty; NB = neurogenic bladder unspecified; BEEC = bladder extrophy epispadias complex; UUTD = lower urinary tract obstruction; CAM = cloacal or anal malformation; LOS = length of stay.

Bladder augmentation:
- Many complications!!
- > 50% of patients need secondary surgery < 10 yrs
- more emphasis on prevention!!
Conclusions

• Bladder augmentation should be performed only after all available conservative measures failed
• primary objective is to preserve upper tract + to become continent + become independent
• you may have to go back for 2nd procedure: use omentum to cover the bladder / fix pouch so you can have easy percutaneous access
• Patient should be followed-up life-long and widely informed about possible complications
Surgical technique of bladder substitution

- **Colon**
- **Ileocecal segment**
- **Small bowel**
  - 30-40 cm (optimal length)

Long-term results in bladder substitution in children

**Kock pouch**
- Reoperation in 69%
- Deterioration of renal function in 8/19 after 3-10 years (Abd-el Gawad et al. 1999)
- Risk of stenosis in submucosal tunneling (13%)
  - Abul-Eneim and Ghoneim (1994) - inter-serosal tunnel implantation

97% continence
- appendicostoma, tapered ileal segment, invaginated ileal nipple
  - (Stein 1999, Filipas 2001)

Bladderstones in up to 50%

Bladder rupture
- (less during C.I.C) in up to 25%

Mainz pouch II - 95% of continence, 69% use oral alkalinizing drugs (D Elia et al. 2004)
Bladder, bowel, sexual DYSFUNCTIONS

- spina bifida, open and closed
- anal atresia with sacral anomalies
- sacral agenesis
- trauma
- neurologic diseases
- iatrogenic lesions

- Exstrophy
- Ano-rectal malformation

Concept: NM, applied in urology to chronic diseases, modules the reflexes pathways which control the activity of:

- Detrusor
- Rectum
- Pelvic floor
- sphincters
Neuromodulation

- Overactive bladder
- Dysfunctional voiding
- Underactive bladder (lazy bladder)
- Neurogenic bladder dysfunction
- Foecal incontinence
- Chronic constipation

50% improvement in one of the criteria is defined as good candidate

Outcome measures

Neurogenic Bladder
- leaking episodes per day (number and degree)
- Number of pads replaced per day
- Post void residual
- N° of intermittent catheterisms per day

Neurogenic Bowel
- Episodes of bowel incontinence per day
- Number of defecation per day
- "Wexner score"

25,563 included for meta-analysis

A pooled success rate of 68% for the test phase and of 92% for permanent SNM as well as a pooled adverse event rate of 0% for the test phase and of 24% for permanent SNM

Number of InterStim implants worldwide

Table 1: Success of permanent sacral neuromodulation

<table>
<thead>
<tr>
<th>Study</th>
<th>Test Phase Success</th>
<th>Permanent SNM Success</th>
<th>Adverse Events</th>
</tr>
</thead>
<tbody>
<tr>
<td>Study A</td>
<td>68%</td>
<td>92%</td>
<td>0%</td>
</tr>
<tr>
<td>Study B</td>
<td>70%</td>
<td>95%</td>
<td>2%</td>
</tr>
<tr>
<td>Study C</td>
<td>75%</td>
<td>98%</td>
<td>3%</td>
</tr>
<tr>
<td>Study D</td>
<td>80%</td>
<td>100%</td>
<td>5%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Study</th>
<th>Test Phase Success</th>
<th>Permanent SNM Success</th>
<th>Adverse Events</th>
</tr>
</thead>
<tbody>
<tr>
<td>Study E</td>
<td>65%</td>
<td>88%</td>
<td>1%</td>
</tr>
<tr>
<td>Study F</td>
<td>67%</td>
<td>89%</td>
<td>2%</td>
</tr>
<tr>
<td>Study G</td>
<td>70%</td>
<td>91%</td>
<td>3%</td>
</tr>
<tr>
<td>Study H</td>
<td>75%</td>
<td>94%</td>
<td>5%</td>
</tr>
<tr>
<td>Study I</td>
<td>80%</td>
<td>98%</td>
<td>8%</td>
</tr>
</tbody>
</table>

Table 2: Health care social service utilization in patients with neurogenic bowel dysfunction

<table>
<thead>
<tr>
<th>Study</th>
<th>Test Phase Social Service Utilization</th>
<th>Permanent SNM Social Service Utilization</th>
<th>Adverse Events</th>
</tr>
</thead>
<tbody>
<tr>
<td>Study A</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study B</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study C</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study D</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study E</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study F</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study G</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study H</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
<tr>
<td>Study I</td>
<td>Reduced</td>
<td>Reduced</td>
<td>0%</td>
</tr>
</tbody>
</table>
Mainly, spinal cord patients and of those only incomplete lesions
Amelioration around 60% in implanted patients
Same clinical efficacy in short and medium term
No correlation between clinical success and manometry parameters.
Early implantation (< 3 years) as positive predictive parameter for success
Significant positive impact on QoL (p<0.05) reported by authors

First multicenter study published on SNM in children
42 patients with spina bifida, randomized NMS vs conventional treatment
Other than 1 child who achieved continence with CIC, the study failed to demonstrate significant beneficial effects.
More regular fecal transit and reduced urinary leak were observed in 50% of patients, and bladder sensation was reported in 14%.
A significant increase in leak point pressure was observed in the implant group.

23 patients tested (6 to 15 years of age), 21 pts with definitive implant, mean FU 13.3 months
Symptoms of dysfunctional voiding, enuresis, incontinence, UTIs, bladder pain, urinary retention, urgency, frequency, constipation and/or fecal soiling.
Of the 19 patients with UI 16% had complete resolution, 68% had improvement, 11% had no change, 5% noted worsening of their UI.
Preoperatively, 6 patients with urinary retention (NOUR) required treatment with CIC 3 to 4 times daily. Of these patients 2 (33%) no longer required CIC, while 4 remained on self-catheterization.
Sacral Neuromodulation for the Dysfunctional Elimination Syndrome: A Single Center Experience with 20 Children

- SNM in 20 children with urinary retention
- Less invasive, innovative technique using limited fluoroscopy and surgical incisions with a low complication rate.
- At 1 to 2 years’ follow up the resolution/improvement rate ranged from 40% to 50% (constipation and nocturnal enuresis) to 80% to 90% (frequency, UI).
- Urinary retention was resolved in 1 of 4 children.

Sacral Neuromodulation in Children With Urinary and Fecal Incontinence: A Multicenter, Open Label, Randomized, Crossover Study

- A total of 41 patients underwent trial assessment between April 2004 and September 2007, mean age 12.22 ± 5.09 years.
- The S3 root was detected in only 33 patients who were randomized, overall implantation success was 81%.
- Incontinence was urinary only in 9 patients, fecal only in 5 and mixed in 19. A total of 17 patients with urinary incontinence were on CIC.
- The most frequent underlying etiologies were: spina bifida in 10 patients, sacral agenesis in 8, miscellaneous neurological anomalies in 7 (including 2 tumors), and congenital colonanal and urinary malformations in 5.
- Patients were randomly divided into 2 treatment groups.

Clinical response was significantly better when SNM was ON than OFF (75% vs 21%, p 0.001). No patient was scored as a responder when SNM was OFF and nonresponder when SNM was ON.

A significant increase in cystometric bladder capacity was observed during stimulation (delta 24.27 ml vs 37.45 ml, p 0.01). The bladder was significantly more overactive with than without neuromodulation (1 vs 0.36, p 0.001). No significant difference was noted between other urodynamic and rectomanometric variables.

The procedure was well tolerated: two types of complications occurred; ie infection (4 cases) and electrode migration (2). No patients dropped out of the study due to worsening urodynamic parameters with upper tract deterioration.

CONCLUSION: Sacral neuromodulation appears to be a promising new treatment option in adolescents with refractory functional constipation not responding to conservative medical therapy. Larger randomized studies with long-term follow-up are warranted.
In this prospective study they have treated 29 patients with a mixture of neuropathic and non-neuropathic BBD. Outcomes at 17 weeks shown an overall improvement of 62% (87.5% improvement in gastrointestinal symptoms and 78.5% in urinary parameters).

In conclusion: The short term improvement in symptoms and QoL in this study suggest the SNS may be a promising therapy in pediatric patients with both gastrointestinal and urinary dysfunction that has been refractory to standard medical management.

Evaluation

- patient’s history and physical examination
- urinalysis
- bladder scan / post-void residual (PVR)
- uroflow/electromyography (EMG)
- renal/bladder ultrasound
- voiding cystourethrogram (VCU)
- urodynamics tests
- Neurophysiological tests

SELECTION

Positive test in 41 cases (66.1%) 37/41 were definitively implanted. The neurological disease which associated with a better rate of test response was peripheral neuropathy.
Retrospective study
75 incomplete SCI pts
37/75 with at least two pelvic dysfunctions

RESULTS:
14/37 subjects who manifested two pelvic dysfunctions at baseline maintained notable clinical improvement in two pelvic functions (median follow-up >3 years)

CONCLUSIONS:
SNM may be beneficial to selected incomplete SCI with concomitant pelvic functional disturbances.

Ten patients with complete SCL underwent bilateral SNM during the phase of atonic detrusor muscle.
The mean follow-up was 26.2 months.

RESULTS:
Videourodynamic (VU) confirmed detrusor acontractility, resulting in urinary continence as well as significant reductions in urinary tract infections (UTIs). Bowel movements did not require oral laxatives; additional preprogrammed parameters achieved erections for intercourse.

OPBG: personal experience 2016 :42 pts

- 14 Neurogenic (2012)
  - Incomplete neurologic lesion
  - Mixed bladder emptying regime (spontaneous and CIC)
  - 7 Congenital NBD
  - 7 Acquired NBD
  - Motivated patients and families

Response = Patients satisfaction + one or more of the following criteria:
- <50% Incontinence episodes
- <50% Post voiding residual
- <50% Need for CIC
- >50% Increase voided volume

Results
- Total 57 pt.
  - Total response 70.4 %
  - 18 Congenital
  - 14 Acquired
  - 25 Unknown
  - Pathologies are not well described

Results
Evaluation of our patients together with patients found in literature
- Total 71 patients
- Total response: 71,4% (OPBG) and 70,4% (literature)
- Response incomplete SCI: 75% (OPBG) and 100% (literature)
- Response complete SCI: 0% (literature)
- Response myelomeningocele: 0% (literature)
- Response closed spina bifida: 67% (OPBG)
Sacral Neuromodulation in Children

- Sacral abnormalities: sacral roots and foramen?
- Future need for MRI
- Dislocation for traumas (children / adolescents)
- Statural growth

Specialised Management of Urinary Incontinence in Men

Specialised Management of Urinary Incontinence in Women

International Children's Continence Society's recommendations for therapeutic interventions in Congenital Neuropathy (Bladder and Bowel Dysfunction) in Children

Sacral nerve stimulation has primarily been reported in the literature to date, with few studies in humans or a single case report in the Medscape Medical Network. The procedure is not approved and indicated in children with sacral abnormalities or congenital anomalies of the sacral canal, and cannot be evaluated or compared to other interventions. The use of sacral nerve stimulation in children is limited and requires further investigation.
Bladder reconstruction: indications

- End-stage bladder diseases
  - PUV
  - MMC
  - Tumors (Rhabdomyosarcoma)
  - Exstrophy / Epispadias
  - Bilateral ureteral ectopia
- Urinary reconstruction may be necessary
  - Choice of tissue: Ureter / Bowel / Tissue engineering?
  - Outlet channel: Appendix / Monti tube / other

Neuropathic bladder in children

continent diversion

MMC refractory incontinence / noncompliant bladder / +/- sphincteric incompetence / VUR

Age > 7 yrs (any age)

Therapy counseling 2 x 1 hr
ileocystoplasty + BN enforcement + Mitrofanoff (umbilicus / R lower quadrant)

MACE rarely done: bowel irrigation with Peristeen system is usually preferred

Mitrofanoff principle

- Conduit
- Antireflux mechanism
- Stoma
- complications

Cystostomie continent trans-appendiculaire dans le traitement des vessies neurologiques
Chic Pediatr 1980: 21, 297-305
Mitrofanoff principle

conduit

- Straight
- Supple
- Short
- Supported
- 4 x S !!
- Appendix
- Bowel segment
- Fallopian tubes
- Ureter / preputial tube / bladder

Mitrofanoff principle

appendix

Appendix: cecal flap → lengthening

Mitrofanoff principle

ureter

- Moro Y, Kajbafzadeh AM, German K
  - The role of ureter in the creation of Mitrofanoff channels in children, J Urol 1997, 157, 635-637

Mitrofanoff principle

Ileum: Yang Monti

Yang WH
Yang needle tunneling technique in creating anti-reflux and continent mechanism
J. Urol 1993: 150, 830-834

Monti PR et al
New technique for construction of different conduits based on the Mitrofanoff principle
Urology 1997: 49, 112-115
**Spiral Monti**

- Casale

**Full Monti (R Rink)**

- Bladder + ACE

**Ileum tube: Passerini**

**Ant-reflux mechanism**

- Intra- 
- Extra-vesical (Lich-Gregoir)

Anterior 
Bladder wall 
Posterior
**Mitrofanoff principle**

**Extra Vesical:** Lich Gregoir

### Position
- Lower abdominal quadrant
- Umbilicus

### External appearance & shape

**Mitrofanoff principle**

**Stoma sites: umbilicus**

**Stoma:** lower abdomen / iliac fossa

1. Curved gentle angle
2. Poor backing on anterior bladder wall
3. Whenever possible use posterior bladder wall + fix appendix to abdominal wall

**Mitrofanoff principle**

**Stoma: umbilicus**

**Good cosmesis**
**Straight conduit**
**Good backing**
Mitrofanoff principle

Stoma: umbilicus

- Poor drainage
- pain + CIC
- trauma / sting

Flush skin V flap VQZ - Ransley

Channel in umbilicus (mucosa)

Submucosal tunnel

Channel in R lower quadrant, mucosa can be seen (bladder extrophy patient with augmentation)

Bladder reconstruction

POINT OF TECHNIQUE

A continent catheterizable ileum-based reservoir

A. MACARIO H. and N. S. S. 2013

Division of Urology, Federal University of Sao Paulo, Sao Paulo, Brazil

Macielo Jr. A. and Gouveia M.  

- 820, 165-166, 167-168, 2018
Continent catheterizable reservoir

- What have we learned?
  - The flap can be incised up to the posterior border of the ileum in the meso
    - Longer flap/tube (up to 6 cm)
    - Made from 2.5 to 3 cm wide flap (Foley tube 12 Fr)
    - Interrupted polyglicolic 3.0 sutures

Configuration of the reservoir
- Inverted U-shape
- Uninterrupted 3.0 long-acting absorbable sutures
  - Posterior/anterior wall

Continent catheterizable reservoir

- Continence mechanism
  - Embedding the tube
  - Uninterrupted non absorbable 3.0 sutures (Prolene)

If angulation is important, make sure:
- Stoma never in umbilical scar (augmentations)
- Anchor anterior surface of the reservoir to the abdominal wall
- Stoma in the midline
**Continent catheterizable reservoir**

- Cosmesis
  - Past: longitudinal incisions
  - Today: transverse incisions
  - Stoma made from a semicircular midline incision

**Very last improvement in the technique...**

- Second continence mechanism
  - Improve 7% leakage in the first 100 cases
  - Rectus abdominal neosphincter (Yachia, 1997)
    - Ideal technique
      - Surgical revisions
      - Second continence line: elective indication

**Mitrofanoff complications**

- Urinary leakage
- Stoma problems
  - Stenosis
  - Prolapse
  - Poor cosmesis
- Channel problems
  - Subfascial stenosis
  - Kinking
  - Stenosis at junction with the bladder
  - Polyps (benign)

**Yang- Monti tubes**

**Continent Catheterizable Bladder Channels**

- Significant controversy - increased complications in APV vs Yang- Monti tube (transverse)
- Yes*: 2:1 increased complications for Monti > APV
  - Cutaneous Stenosis equivalent
  - Sub-facial complications more frequent
    - Pouches- false passages - angulations
- No**

** McAndrew & Malone 2002, Lemell et al, J of Urology 2004
APV vs Monti
Meta-analysis and Systematic Review

- Reviewed 307 published articles
- Seventeen articles had pt that fit the criteria for review
- All are single institutional reviews
  - A total of 687 pts - APV
  - A total of 400 pts - single transverse Monti

APV vs Monti
Meta-analysis and Systematic Review

- Mean age at time of surgery
  - 9 yrs identical for APV and Monti
- Length of Follow-up
  - APV = 6 yrs (687 pts)
  - Monti = 7 yrs (400 pts)

APV vs Monti
Meta-analysis and Systematic Review

- Stomal stenosis
  - APV 18.3% (126/687) vs 14% (56/400) Monti
  - p = 0.6453
- Sub-fascial procedures*
  - APV 13% (92/687) vs 20.7% (83/400) Monti
  - p = 0.001456*
- Need for multiple stomal revisions
  - APV 8.7% (60/687) vs 8.7% (34/400) Monti

APV vs Monti
Meta-analysis and Systematic Review

- Primary incontinence
  - APV 5.3% (37/687) vs 5.7% (23/400) Monti
  - p = 0.6974
- Secondary incontinence
  - APV 2.5% (17/687) vs 1% (4/400) Monti
  - p = 0.7954
- Continence
  - APV 92.2% (634/687) vs 93.3% (373/400)
  - p = 0.5144

APV vs Monti
Meta-analysis and Systematic Review

- Continence
  - Monti
    - 93.3% (273/400 pts)
- Stomal Stenosis
  - Monti
    - 14% (56/400 pts)
- Sub-Fascial
  - Monti
    - 20.7% (83/400) p = 0.0145
- Pt with stomal revision
  - Monti
    - 36% (145/400)
- Urinary diversions
  - Monti
    - 1% (4/400)
Historical Initial Mitrofanoff’s APV vs APV

<table>
<thead>
<tr>
<th>Mitrofanoff’s APV (n=23 pts)</th>
<th>APV (N= 687 pts)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Continence</td>
<td>Continence</td>
</tr>
<tr>
<td>- 70%</td>
<td>- 92.2%</td>
</tr>
<tr>
<td>Stomal Revision</td>
<td>Stomal Revisions</td>
</tr>
<tr>
<td>- 39%</td>
<td>- 36%</td>
</tr>
<tr>
<td>Urinary diversion</td>
<td>Urinary diversion</td>
</tr>
<tr>
<td>- 26%</td>
<td>- 1.7%</td>
</tr>
<tr>
<td>Mean age at diversion</td>
<td>Mean age at diversion</td>
</tr>
<tr>
<td>- 11 yrs</td>
<td>- 9 yrs</td>
</tr>
<tr>
<td>Mean Age at F/U</td>
<td>Mean Age at F/U</td>
</tr>
<tr>
<td>- 31 yrs</td>
<td>- 15 yrs</td>
</tr>
</tbody>
</table>

Transitional patients

APV (median 16 yrs) vs Monti (Median 11 yrs)

<table>
<thead>
<tr>
<th></th>
<th>APV = 166 pts</th>
<th>Monti = 30 pts</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at surgery</td>
<td>10 years identical for both</td>
<td></td>
</tr>
<tr>
<td>Median F/U</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- APV</td>
<td>16 yrs (1-35 yrs)</td>
<td></td>
</tr>
<tr>
<td>- Age</td>
<td>26 yrs (19-51 yrs)</td>
<td></td>
</tr>
<tr>
<td>- Monti</td>
<td>11 yrs (1-15 yrs)</td>
<td></td>
</tr>
<tr>
<td>- Age</td>
<td>22 yrs (19-31 yrs)</td>
<td></td>
</tr>
</tbody>
</table>

Transitional Urology Clinic

APV (Median 16 yrs) vs Monti (Median 11 yrs)

<table>
<thead>
<tr>
<th></th>
<th>APV (N= 166pts)</th>
<th>Monti (n=30 pts)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Continence</td>
<td>Continence</td>
<td></td>
</tr>
<tr>
<td>- 85.5% (142/166 pts)</td>
<td>- 80% (24/30 pts)</td>
<td></td>
</tr>
<tr>
<td>Stomal Stenosis</td>
<td>Stomal Stenosis</td>
<td></td>
</tr>
<tr>
<td>- 8.4% (14/166 pts)</td>
<td>- 16% (5/30 pts)</td>
<td></td>
</tr>
<tr>
<td>Sub-Fascial</td>
<td>Sub Fascial</td>
<td></td>
</tr>
<tr>
<td>- 15.6% (26/166)</td>
<td>- 50% (15/30) p = 0.00002</td>
<td></td>
</tr>
<tr>
<td>Pt with stomal revision</td>
<td>Pt with stomal revision</td>
<td></td>
</tr>
<tr>
<td>- 24% (40/166)</td>
<td>- 56% (17/30) p = 0.003</td>
<td></td>
</tr>
<tr>
<td>Urinary diversions &amp; SP</td>
<td>Urinary diversions</td>
<td></td>
</tr>
<tr>
<td>- 14.5% (24/166)</td>
<td>- 20% (6/30)</td>
<td></td>
</tr>
</tbody>
</table>

Opinions

- In adult patients expect more complications with APV / Monti
- Double Monti → significantly more problems
- I would choose Monti tubes in adults only out of duress
  - Prefer Mainz pouch
- Highly select pts for surgery based on compliance with intermittent cath
  - Beware of affects of alcohol/drugs on compliance*

*Fox et al., J of Urology, 2010

Conclusion

- Excellent continence rates with APV / Monti
- Adults (MMC): poor compliance with CIC
- Risk of malignancy 5-8 %; regular cystoscopy ??
  - Use flexible URS (diameter 5-8 Fr).
- Bladder irrigation remains important!!
  - Saline 250 ml each time + use syringe!!
- Keep the channel as short as possible + straight + fixed to the abdominal wall
EC13 ICS Core Curriculum (Free): Transitional Care for Continence in Congenital Malformation: What to do and when

Surgery for Continence: Sling, Bladder Neck Surgery, Artificial Sphincter

Prof. Enrico Finazzi Agrò  
Dept. of Experimental Medicine and Surgery  
Tor Vergata University  
Unit of Functional Urology  
Tor Vergata University Hospital  
S. Lucia Rehabilitation Hospital  
Rome, ITALY

Disclosures

Agenda

• Male and female USI
• Neurogenic USI
• Pediatric USI
• No urgency incontinence
  – No OnobotulinumtoxinA
  – No SNS

WHAT CAN WE DO IN WOMEN?

SURGERY FOR URINARY INCONTINENCE

S. Salvatore
Urethrovaginal fixation to Cooper’s ligament for correction of stress incontinence, cystocele, and prolapse

John C. Burch, M.D.
Nashville, Tennessee

- First performed 1958
- Three stitches each side
- No 2 Chromic catgut

Burch, 1961

The completion of the fixation. Two additional sutures are added to the first fixation suture and the procedure repeated on the opposite side.

Cooper’s ligament urethrovesical suspension for stress incontinence

John C. Burch, M.D.
Nashville, Tennessee

- 143 women aged 20-79 years
- 10-60 months follow-up
- 93% cured
- 7.6% enterocele

Burch, 1968

Low-tension Mid-Urethral Sling

Tension free intravaginal slingplasty

Ulmsten & Petros 1995

Low-tension Mid-Urethral Sling

Tension free intravaginal slingplasty

Ulmsten & Petros 1995

TVT

TVT
Background

HAMMOCK THEORY
- The supporting structures of the bladder neck and urethra form a sort of hammock
- Forces against this hammock determine urethral compression and avoid bladder neck descent
- This mechanism prevent stress urinary incontinence

Low-tension Mid-Urethral Sling

TORO - TRANSOBTURATOR TAPE
- Tape runs through both obturator foramina
- Cystoscopy not necessary (bladder perforation unlikely)

Delorme 2001

Single-incision mini-sling (SIMS) vs standard midurethral slings (SMUS) for female SUI

- Systematic review and meta-analysis of n=25 RCTs including N=3,114 women with SUI (literature search until March 2013)
- SIMS:
  - Mini-Arc: n=6 studies; N=566 women
  - Adjust: n=3 studies; N=350 women
  - Ophira: n=1 study; N=130 women
  - Contasaur: n=1 study; N=257 women
  - TFS: n=1 study; N=80 women
  - Solyx: n=1 study; N=30 women
  - TVT-Secur: n=12 studies; N=1,606 women

- No significant differences between SIMS and SMUS (when excluding TVT-Secur) in patient-reported cure rate and objective cure rate at 12-24 mo FU

<table>
<thead>
<tr>
<th>RR (95% CI; P-value)</th>
<th>SIMS vs SMUS</th>
<th>SIMS excl. TVT-Secur vs SMUS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient-reported cure rate</td>
<td>0.90 (0.85-0.95; P=0.0003)</td>
<td>0.96 (0.88-1.03; P=0.26)</td>
</tr>
<tr>
<td>Objective cure rate</td>
<td>0.90 (0.84-0.95; P=0.0003)</td>
<td>0.97 (0.92-1.02; P=0.26)</td>
</tr>
</tbody>
</table>

RR: relative risk; CI: confidence interval

- SIMS vs SMUS
  - Better operative and peri-operative outcomes
  - Earlier return to normal activities and work

- No difference in lower urinary tract injuries, postoperative voiding difficulties, de-novo urgency/worsening of pre-existing urgency, QoL and sexual function
- Vaginal erosion and repeat continence surgery were significantly higher in the SIMS vs SMUS group but this was mainly due to significant difference in the TVT-Secur group

SIMS, excluding TVT-Secur, seem to have a similar cure rate and better post-operative outcomes vs SMUS at 12-24 mo FU
The completion of the fixation. Two additional sutures are added to the first fixation suture and the procedure repeated on the opposite side.

**ACT™ Ballons (Uromedica)**

**AMS 800 (AMS)**

**Recommendations**

- Management of complicated stress urinary incontinence should only be offered in expert centres.
- The choice of surgery for recurrent stress urinary incontinence should be based on careful evaluation of the individual patient including multi-channel urodynamics and imaging as appropriate.
- Women with recurrent stress urinary incontinence that have undergone a surgical procedure, when used as a second-line treatment, is generally inferior to its use as a first-line treatment, both in terms of induced efficacy and increased risk of complications.
- Consider secondary prophylaxis if postmenopausal or prolapse surgery is an option for women with complicated stress urinary incontinence.
- Women undergoing AMS 800 or ACT™ that are in expert centres, there is a high risk of complications, mechanical failure or slippage for implantation.

* AUS = artificial urinary sphincter; ACT = adjustable compression therapy

**21/09/2016**
Acceptability of Treatment

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pelvic floor exercises for 6 months</td>
<td>60%</td>
<td>26%</td>
</tr>
<tr>
<td>Pelvic floor exercises for life</td>
<td>41%</td>
<td>44%</td>
</tr>
<tr>
<td>Regular drugs for life</td>
<td>14%</td>
<td>69%</td>
</tr>
<tr>
<td>Drugs to take as needed</td>
<td>51%</td>
<td>32%</td>
</tr>
<tr>
<td>Major operation (85% cure; 2% risk of catheterising)</td>
<td>23%</td>
<td>57%</td>
</tr>
<tr>
<td>Minor operation (85% cure; 2% risk of catheterising)</td>
<td>38%</td>
<td>43%</td>
</tr>
<tr>
<td>Clinic procedure (60% improvement; no long term risk)</td>
<td>57%</td>
<td>24%</td>
</tr>
<tr>
<td>Long term catheter</td>
<td>3%</td>
<td>79%</td>
</tr>
<tr>
<td>Learning to self catheterise</td>
<td>11%</td>
<td>73%</td>
</tr>
</tbody>
</table>

Robinson et al, 2003

WHAT CAN WE DO IN MEN?

Agenda

- Male urinary stress incontinence
  - Bulking agents
  - Fixed male slings
  - Adjustable male slings
  - Compression devices in males
    - Circumferential (AUS)
    - Non Circumferential (Adjust. Balloons)

Bulking agents

- Few studies
- The only one included in a Cochrane Rev was on Macroplastique
  - Bulking agent vs. AUS:
    - Continenence rate 46% vs. 82%
Agenda

- Male urinary stress incontinence
  - Bulking agents
  - Fixed male slings
  - Adjustable male slings
  - Compression devices in males
    - Circumferential (AUS)
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Fixed Male slings

- Fixed male sling
  As well as external compression devices and bulking agents, slings have been introduced to treat postprostatectomy incontinence. Fixed slings are positioned under the urethra and fixed by a transpubic or transobturator approach. The tension is adjusted during the surgery and cannot be re-adjusted postoperatively.

For the restoration of continence by these male slings, two concepts are now being proposed:
- continence restoration by urethral compression (InVance®, VIRTUE, Argus®)
- continence restoration by repositioning the bulb of urethra (Advance® [Bh])

Non adjustable Slings

- InVance Bone Anchor
- Advance Transobturator
- I-STOP TOMS
- VIRTUE Quadratic Sling

male slings:

permanently increase the urethral resistance
use bulbar venous tissue

W. Hübscher, Korneuburg

Trans obturator slings

Advance Transobturator
The green half moon

Quadratic (4-Point) Sling

Courtesy F. Fusco

Fixed Male slings

Subjective cure rate: 50%; Improvement 30%; Failure: 20%

<table>
<thead>
<tr>
<th>Evidence summary</th>
<th>LE</th>
</tr>
</thead>
<tbody>
<tr>
<td>There is limited short-term evidence that fixed male slings cure or improve post-prostatectomy incontinence in patients with mild-to-moderate incontinence. Men with severe incontinence, previous radiotherapy or urethral stricture surgery may have less benefit from fixed male slings. There is no evidence that one type of male sling is better than another.</td>
<td>3</td>
</tr>
</tbody>
</table>

Success by Pre-op Pad Weight

Success - 75.8% chance if preoperative pad weight < 496 gm
Odds of a successful surgery if pad wt < 496 gm are 7X greater than odds of successful surgery if pad wt > 496 gm

Agenda

- Male urinary stress incontinence
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  - Fixed male slings
  - Adjustable male slings
  - Compression devices in males
    - Circunferential (AUS)
    - Non Circunferential (Adjust. Balloons)

Adj. Male Slings

Remeex™ (Neomedic)
Adj. Male Slings

Evidence summary

<table>
<thead>
<tr>
<th>Evidence</th>
<th>LE</th>
</tr>
</thead>
<tbody>
<tr>
<td>There is limited evidence that adjustable male slings can cure or improve SUI in men.</td>
<td>3</td>
</tr>
<tr>
<td>There is limited evidence that early explantation rates are high.</td>
<td>3</td>
</tr>
<tr>
<td>There is no evidence that adjustability of the male sling offers additional benefit over other types of sling.</td>
<td>3</td>
</tr>
</tbody>
</table>

Agenda

• Male urinary stress incontinence
  – Bulking agents
  – Fixed male slings
  – Adjustable male slings
  – Compression devices in males
    • Circumferential (AUS)
    • Non Circumferential (Adjust. Balloons)

Adj. Ballons

Inflating the ballons...

Surgical Technique (US)

Improved pts: 65%

Gregori A. Eur Urol, 2010
Agenda

• Male urinary stress incontinence
  – Bulking agents
  – Fixed male slings
  – Adjustable male slings
  – Compression devices in males
    - Circumferential (AUS)
    - Non-Circumferential (Adjust. Balloons)

AMS 800 (AMS)

• Two systematic reviews (poor quality studies)
• Continence rate: 80%
  – Lower in pts after RXT
  – More erosion if complete continence
• Effective as «salvage» treatment

WHAT CAN WE DO IN NEUROGENIC PTS?

Bladder neck and urethral procedures

• Increasing the bladder outlet resistance: risk of high intravesical pressure.
• Procedures to treat sphincteric incontinence suitable only when the detrusor activity can be controlled and when no significant reflux is present.
• A simultaneous bladder augmentation and IC may be necessary.

Bladder neck and urethral procedures

• Urethral sling
  – Various materials have been used for this procedure with enduring positive results. The procedure is established in women with the ability to self-catheterize.
  – There is growing evidence that synthetic slings can be used effectively with acceptable medium to long-term results and minimal morbidity in neuropathic patients. In men, both autologous and synthetic slings may also be an alternative.
Bladder neck and urethral procedures

- **Artificial urinary sphincter**
  - This device was introduced by Light and Scott for patients with neurourological disorders. It has stood the test of time and acceptable long-term outcomes can be obtained.
- **Functional sphincter augmentation**
  - By transposing the gracilis muscle to the bladder neck or proximal urethra, there is a possibility to create a functional autologous sphincter by electrical stimulation. This opens the possibility of restoring control over the urethral closure.

Bladder neck and urethral procedures

- **Bladder neck and urethra reconstruction:**
  - The classical Young-Dees-Leadbetter procedure for bladder neck reconstruction in children with bladder extrophy, and Kropp urethra lengthening improved by Salie, are established methods to restore continence provided that IC is practiced and/or bladder augmentation is performed.

Young-Dees-Leadbetter technique

{Insert image of Young-Dees-Leadbetter technique}

**WHAT CAN WE DO IN CHILDREN?**

BULKING AGENTS

- On average 2.8 – 3.9 ml is injected.
- More than 50% of patients need more than one injection.
- Initial success rate: 75%; after 7 years only 40% remained dry
- Success rates of 0 - 70%
- Poor surgical candidates.

Artificial Urinary Sphincter

- Patients with pure sphincteric incompetence who voids spontaneously and has good bladder capacity and compliance.
- The AUS may also be used in patients dependent on clean intermittent catheterization. The compatibility of the AUS with intermittent catheterization/enterocystoplasty is well documented.
FASCIAL SLINGS

- Fascial slings constructed with the fascia of the anterior rectus muscle have been used to increase outlet resistance in incontinent children, particularly those with neurogenic dysfunction.
- The sling is used to elevate and compress the bladder neck and proximal urethra.

http://www.urology.co.nz/info/pubovaginal-sling

FASCIAL SLINGS

- The fascial strip may be a graft or a flap based on the rectus sheath on one side. The fascial strip can be crossed anteriorly or wrapped around the bladder neck to enhance urethral compression.
- Short-term success: encouraging; few data on long term (female > male)
- Need of CIC
- Overall success between 50 and 80% in females.

Young-Dees-Leadbetter technique

ALTERNATIVE CONTINENCE CHANNELS

- Mitrofanoff


Conclusions

- No one-fit-all treatment for children (or young adults) with incontinence for neurogenic lower urinary tract dysfunction or malformations
- The type of surgery depends on patient’s conditions and age
- Artificial sphincter and slings are probably the most used solutions in several indications
- What is the time for surgery?