**W27: ICS Core Curriculum (Free): Children and Young Adults**

**Workshop Committee on Transitional Care for Continence in Congenital Malformations.**

**Workshop Chair: Giovanni Mosiello, Italy**

13 September 2017 14:30 - 16:00

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**Speaker Powerpoint Slides**

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

**Aims of Workshop**

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

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

**Learning Objectives**

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

**Learning Outcomes**

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

**Target Audience**

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

**Advanced/Basic**

Advanced

**Suggested Learning before Workshop Attendance**


**Suggested Reading**


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

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Neurogenic bladder (NB), posterior urethral valves (PUV), bladder exstrophy (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 (DD), the voiding pattern and the post-voiding residual urine volume.

**Posterior urethral valves**

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

How To Manage Boys Operated for Posterior Urethral Valves and Severe Hypospadias
Selcuk Yucel, MD

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

Hypospadias is a disease with a relatively higher incidence compared to four decades ago. Particularly posterior hypospadias is a very challenging disease requiring surgical skills and experience. Obstructed voiding may be associated with a cosmetically very acceptable penis following hypospadias repair. However, uroflowmetrics 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 unrepaird hypospadias.

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

**Mario De Gennaro**

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

Artificial 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
artificial 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.

Bowel Management
Giovanni Mosiello

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

Patient population:

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

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

Options for Bowel Management
Fluid intake and diet
Physical activity
Medications
Anal/rectal stimulation and Manual evacuation
Suppositories and micro-enemas
Anal plug
Abdominal massage and Biofeedback

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

Conclusions

The treatment of BD significantly affects the family and caregivers quality of life and has a high social cost. Before the use of TAI surgical procedures were common: percutaneous cecostomy, bowel resections, permanent stomas and Malone 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.