W22: ICS Core Curriculum (Free): Transitional care for continence in posterior urethral valves, bladder exstrophy, cloaca, neurogenic bladder

Workshop Chair: Giovanni Mosiello, Italy
04 September 2019 09:00 - 10:30

Start	End	Topic	Speakers
09:00	09:05	Introduction	Giovanni Mosiello
09:05	09:15	posterior urethral valves : concerns in transition and long- term complications	Gundela Holmdahl
09:15	09:25	Neurogenic bladder : concerns in transition and long- term complications	Kate Abrahamsson
09:25	09:35	Bladder exstrophy: concerns in transition and long- term complications	Giovanni Mosiello
09:35	09:45	Cloaca: concerns in transition and long- term complications	Giovanni Mosiello
09:45	09:55	Conservative and no invasive management of continence in transitional care	Magdalena VuMinh Arnell
09:55	10:05	Surgery for continence in Young adults with congenital malformations: tip and tricks	Rien Nijman
10:05	10:10	European reference network for rare and complex disease : could be this a valid worldwide model?	Kate Abrahamsson
10:10	10:15	Complex case presentation	Giovanni Mosiello
10:15	10:25	Questions	All
10:25	10:30	Take home messages	Giovanni Mosiello

Aims of Workshop

Spina bifida, bladder exstrophy, cloaca , posterior urethral valves are managed immediately after the birth with a surgical procedure . All these congenital malformations require lifelong urological care for the treatment of continence and there are critical aspects to define:
- Correct management in childhood, defining a correct timing for surgery for continence, transition out of childhood, the role of conservative management, indications for re-operation , complications in long-term
The objective of this workshop is to critically evaluate the treatment in childhood, defining concern in transition, exploring new modalities for a best practice treatment of incontinence life-long .

Learning Objectives

At the end of workshop delegates will be able to describe common treatment and evolution in childhood of posterior urethral valves, bladder exstrophy, cloaca, neurogenic bladder (spina bifida)

Target Audience

Urology, Urogynaecology, Bowel Dysfunction, Conservative Management

Advanced/Basic

Advanced

Suggested Learning before Workshop Attendance

-International Consultation on Incontinence 6th edition 2017 ICUD edition: P Abrams, L Cardozo,A Wagg, A Wein . Chapter 9 Diagnosis and management of urinary incontinence in childhood. Chair
Rien Nijman (THE NETHERLANDS)
Members
Serdar Tekgul (TURKEY) Janet Chase (AUSTRALIA)
An Bael (BELGIUM)
Dan Wood (UK)
Doug Canning (USA)
Paul Austin (USA)
Erik van Laeke (Belgium) Johan van der Walle (Belgium) Giovanni Mosiello (Italy) Alexander von Gontard (GERMANY)
-EAU/ESPU guidelines on Pediatric Urology EAU Guidelines on
Paediatric Urology S. Tekgül (Chair), H.S. Dogan, P. Hoebeke, R. Kocvara, J.M. Nijman (Vice-chair), C. Radmayr, R. Stein Guidelines Associates: E. Erdem, A.K. Nambiar, M.S. Silay, S. Undre

Introduction

Spina bifida, bladder exstrophy/epispadia,cloaca, posterior urethral valves are managed immediately after the birth with a surgical procedure .All these patients require lifelong urological care for the treatment of continence and there are critical aspects to define:
- Correct management in childhood to avoid procedure that impair adult life aspects as fertility and pregnancy
- transition out of childhood: who should manage the mature pediatric urology patients?
- Lack of knowledge in pediatric and adult health care professionals about adult life problems and congenital pathologies respectively.

The objective of this workshop is to critically evaluate the treatment in childhood, defining a best practice treatment of incontinence and a correct indications in children and adolescents of surgical procedure considering long-term effects.

Posterior urethral valves: concerns in transition and long- term complications
Gundela Holmdahl

Posterior Urethral Valves represent a congenital condition, with a prevalence of 2.48 per 10,000 live births, which very often deals with bladder problems. It is one of the main issue of Clinical Urodynamic in children.
A great proportion of patients suffer from urinary incontinence, and bladder dysfunction was seen in many of them (55%, 0-72%) after primary treatment. Despite early relief of obstruction, pathological changes in the bladder can cause bladder dysfunction and consequent progressive slow deterioration of the upper urinary tract drainage with renal failure, possibly leading to renal insufficiency over years. Thus, prevention and treatment of bladder dysfunction, based on urodynamic observations are mandatory in terms of long-term outcome. Over time abnormal urodynamic patterns of myogenic failure, detrusor overactivity and decreased compliance/small capacity (so-called 'valve bladder) may develop. In addition, urodynamic bladder function may change with time, leading with myogenic failure in some post-pubertal patients, most likely secondary to increased urine production and persistence of functional bladder neck outlet obstruction.impairment. Nowadays, differently than in the past when aggressive surgical approaches were claimed, a conservative management was shown more effective and safe, following primary neonatal valves ablation. An accurate and early toilet training program, a strict follow-up mainly based on not invasive urodynamic evaluations, and an advanced urotherapy program with associate appropriate drugs therapy are effective to prevent and treat bladder dysfunction in children with urethral valves. The cases leading with renal insufficiency, polyuria, upper urinary tract deterioration and, ultimately, kidney transplant need more aggressive investigations and treatments.

Neurogenic bladder : concerns in transition and long- term complications
Kate Abrahamsson

A damage a part of the central nervous system involved in LUT function may result in the neurogenic bladder. The most common cause in childhood is myelomeningocoele. The incidence has been decreasing due to folic acid supplementation during pregnancy. Before the discovery of clean intermittent catheterisation (CIC) children with this condition usually had repeated febrile UTIs and progressive renal failure due to these and to high intravesical pressure.. Treatment is directed to preservation of bladder and kidney function. As long as bladder volume with low pressures is achieved, kidney function will remains normal. Low pressures are possible by using antimuscarinic medication or botulinum toxin or in some cases operations. Intermittent catheterization is needed to ensure proper bladder evacuation. In most cases it is possible to become dry in between catheterization, or perhaps some incontinence remains during transfers or straining. Neurogenic bladder is joined with a neurogenic bowel. Colonic washouts have improved the quality of life dramatically: almost all patients can obtain pseudo continence for stool. Attention for tethering and scarring of kidneys is the mainstay of the treatment. Children with neurogenic bladder and bowel dysfunctions (NBBD) require lifelong evaluation and treatment, especially for continence. Different treatment options in order to ameliorate continence can be considered lifelong including rehabilitation , pharmacotherapy, endoscopic procedures and surgery reconstructions. For this reason is very important for all health care professionals involved in the treatment of these patients to know about the effects in adult life of the procedure performed in childhood, in order to define a correct timing of management, discussing how the concerns for a correct choice in the management of the single patient are related to individual condition of the single person. Actually with the improvement in care, children with spina bifida (SB), that is the most common cause of NBBD in pediatrics, survive into adulthood, and a correct treatment choice permits to increase quality of life (QOL), preserve renal function and to avoid procedure that can impair in adult life aspects as fertility and pregnancy.

Bladder exstrophy: concerns in transition and long- term complication
Giovanni Mosiello

Bladder exstrophy – epispadias complex (BEEC) is one of a major challenge 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 , The aims of surgical treatment are well established across all reconstructive technique: a functional urinary reservoir, preserved renal function, urinary continence, and cosmetically pleasing and functional genitalia . Successful primary

closure is the most important determinant of potential bladder growth and eventual continence. The timing of initial closure is an important predictor for successA higher percentage of female patients achieved complete continence compared to males that requires many surgical procedures including repeated bulking agents, bladder augmentation , bladder neck reconstruction and derivation, in order to achieve continence during life.. ; urogynecologist and neurourologist , discussing in interactive way some concenrs. Some cases will be presented . At the end of the workshop the aim is that all the partecipants will be able do manage correctly these patients through all the different ages.

Cloaca: concerns in transition and long- term complications
Giovanni Mosiello

Patients born with an ARM usually necessitate a bowel reconstruction to insure continence and bowel empty. ARM is associated with a high frequency of spine defect and in these patients the bowel dysfunction could be the result of these two malformations. Literature data shown that, even if successfully reconstructed, a regular program for colon cleaning is necessary in about two thirds of these patients for variable period of time or, in the worst cases for all their life. Now a day, the most efficient modality to clean the colon in these patients seems to be by transanal irrigation (TAI). The Bowel Management introduced by Peña, consists in irrigating daily the colon with a probe and different solution, amount and frequency according to the age of patient and type of malformation. The quality of life of these patients depends on efficacy of the procedure and time consuming. Other most conservatives modalities of BM such as assistive intervention, demonstrated a good results even if, in case of manual evacuation may be unacceptable by patients and care givers. The possibility to attend school, sport, and, later, to be able to have a job or social, and sexual relationship are the goals for a correct bowel management, that most of the patients are willing to reach to improve the QoL.

Conservative and no invasive management of continence in transitional care
Magdalena VuMinh Arnell

Urotherapy is described as a therapy for patients with Lower Urinary Tract Syndrome (LUTS) in order to improve the dysfunction of the bladder. It is a combination of cognitive, behavioral, and physical training. The purpose of the urotherapy is to normalize voiding pattern and prevent functional damage. Therefore is easy to observe that urotherapy alone can't be effective mostly in children with anatomic or neurogenic causes of LUTS. Anyway urotherapy can be as a basic guidance useful as explanation of the bladder function and dysfunction. Understanding of the problem helps to further motivation for treatment. Account shall be taken of the total process of clean fill to be emptying the bladder. The program is divided into standard therapy and specific interventions, such as pelvic floor training, behavior training and biofeedback training, clean intermittent catheterization, neuromodulation. Explanation and instruction, in combination with drug treatment of constipation and urinary tract infection, physical therapy and feedback can be part of the treatment. CIC: During the 1970's an American urologist Jack Lapides (1914-1995) developed clean intermittent catheterization (CIC) to treat persistent infections in the urinary tract. In the 1980s CIC was widely applied to evacuate urine in neuropathic bladder patients with amazing results to prevent kidney damage, as well as for posterior urethral valves, cloaca, bladder exstrophy. A child of 6 years old can learn how to self-catheterize themselves well. This is recommended because self-catheterization is less painful and makes the child less dependent. Adolescents tend to ignore good advice, low adherence and they are prone to postpone intermittent catheterizations especially when they can retain their urine because successful bladder neck surgery and/or bladder augmentation surgery , for this reason they can present suddenly an increased risk for the upper urinary tract. Despite successful valve ablation a significant proportion of children still present with voiding disorders due to detrusor overactivity or low compliance or increased activity of the perineal floor that mimic DV. As already mentioned in rehabilitation in DV one case we treated with EMG and uroflow BFB after urethral valve ablation was then able to relax perineal floor and avoid abdominal contraction during voiding but still with a fractionated flow due to detrusor underactivity. In a recent study 30 children still with LUTS after posterior urethral valves, showing at urodynamic evaluation detrusor overactivity or low compliance in 77%, non relaxing perineal floor in 23% and obstructed bladder neck in 7% were submitted to bladder and PFM BFB . Children with neurogenic bladder are poor candidates for BFB even in uncomplete lesion. Antibiotic prophylaxis and sometimes antimuscarinics can be useful in the initial approach to obtain earlier good results and improve motivation but should then be dismissed during follow up. Some Authors prefer a labor intensive program, even on an inpatient basis, others prefer short sessions and homework. Sacral neuromodulation delivered via implantable electrodes offer efficacy in improving neurogenic bladder dysfunction. Up to date, case series reported benefits in paediatric OAB, dysfunctional elimination syndrome, and Fowler syndrom. Selective stimulation of the sacral and pudendal nerves has led to significant improvement in overactive bladder in patients who have had stimulators implanted The role of spinal stimulation in children is still to be clearly defined. The use of TENS involving the S3 region with surface electrodes made this technique applicable in children. The mechanism for neuromodulation is not well understood. The hypothesis is that the electrical current directly affects the central nervous system by artificially activating neural structures, facilitating both neural plasticity and normative afferent and efferent activity innervating the low urinary tract.

Surgery for continence in Young adults with congenital malformations: tip and tricks
Rien Nijman

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

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

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

Bladder augmentation

When medication has failed to decrease elevated end filling detrusor pressure, or creates troublesome side effects, bladder augmentation may be indicated. Detrusor myectomy, or detrusorectomy,''auto-augmentation,'' shows a success rate of approximately 50% with respect to bladder compliance and capacity in neurogenic bladders.This procedure has been very popular in the past but was replaced , as miniinvasive procedure by botulinum toxin injection. Recently gained new popularity thanks to laparoscopic procedure . Ileocystoplasty is more commonly performed,but carries the risk of postoperative intestinal obstruction, mucus retention, increased rate of stone formation, and electrolyte imbalance. The risk of complication or effectiveness is the same either with ileum or with sigma and the choice is related to surgeon's preference and experience.The risk of secondary malignancy of the augmented bladder is increased, although less than 20 cases have been described worldwide.

Augmentation may be combined with ureteral reimplantation, bladder neck tightening (sling suspension, bladder neck reconstruction, artificial sphincter implantation) or the creation of a continent catheterizable urinary stoma (Mitrofanoff, Monti). As bladder augmentation lowers bladder pressure, diminishing or abolishing vesicoureteral reflux, ureteral reimplantation should only be performed in cases where high grade reflux occurs at low bladder pressure. Similarly, as bladder augmentation will improve continence, only patients with low leak point pressure need reinforcement of the bladder outlet. Urodynamic testing will determine surgical options. Bladder replacement instead of augmentation may be appropriate in cases of bladder exstrophy where use of native bladder tissue is impossible. The use of tissue engineering is still far from a clinical use and this treatment can not be considered in the next 10 years.

Derivation
Ileal conduit ('wet deviation') is no longer indicated except in case of severe mental disability or severe renal dysfunction and no options for bladder reconstruction.

European reference network for rare and complex disease : could be this a valid worldwide model?
Kate Abrahamsson

Rare and complex urogenital conditions can require surgical correction, often during the neonatal period or in childhood. Individuals affected require life-long care provided by multidisciplinary teams of experts who plan and perform surgery, and provide post-operative physiotherapy and psychology support. eUROGEN will provide independently-evaluated best practice guidelines and improve the sharing of outcomes. It will, for the first time, offer the capacity for tracking long-term outcomes for patients over a 15 to 20-year period. The network will collect data and materials where they are lacking, develop new guidelines, build evidence of best practice, identify practice variation, develop education programmes and training, set the research agenda in collaboration with patient representatives, and share knowledge through participation in virtual multidisciplinary teams. Ultimately, the network seeks to advance innovation in medicine and improve diagnostics and treatment for patients

Complex case presentation
A girl of 9 years old has been previously operated several times for anorectal malformation (vestibular rectal fistula) ectopic ureters in bilateral duplex system, occult spinal dysraphysm, and is presenting with a severe urinary incontinence . Furthermore she has been then operated for bladder neck reconstruction.The girl and her family require a definitive solution.