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/B basic
Advanced

Suggested Learning before Workshop Attendance
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)
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 myelomeningocele. 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
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 extrophy. 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 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 incomplete 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.
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 miniminvasive 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.

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.
Posterior urethral valves
transition and long-term complications
Gundela Holmdahl
ICS
Gothenburg 2019

Secondary pathology
• Detrusor hypertrophy/bladder dysfunction
• Dilated upper urinary tracts
• Reflux
• Renal dysplasia/obstructive uropathy
• Oligohydramnios — lung hypoplasia

Newborn with PUV
• Stabilisation/neonatal care
• Bladder drainage
• Primary investigation with renal ultrasound and VCUG
• Follow creatinine and urinary output
• Resection of valves

Long term outcome PUV
• Renal status
  – 50% normal renal function
• Bladder dysfunction
  – LUTS twice as common (Jalkanen 2016)
  – Mostly emptying problems, seldom incontinence
• Sexual function and paternity
  – Probably normal, but depending on renal status (Taskinen 2012)

Bad prognostic factors
Elevated nadir creatinine (1 year of age) (Hennus 2012)
Early diagnosis
Respiratory problems neonatally
Bilateral reflux
Recurrent UTI
Day time incontinence at > 5 years of age
Polyuria

Treatment options
• Prenatal intervention
• Upper urinary tract drainage
• Vescostomy
• Early resection – cycling of the bladder
• Bladder treatment
Changing Bladder dysfunction in boys with PUV

In infants over activity with low capacity, dyscoordination 50% low compliance
With time change to under activity with emptying problems Few patients still low-compliant

The valve bladder syndrome
- Persisting upper tract dilatation
- Poor compliant bladder
- Emptying problems
  (Glassberg 2001)
Probably due to a insufficiently treated bladder in combination with polyuria

Bladder treatment designed for type of dysfunction

Bladder regimen (Hoa Doung 2010, 2013)
Pharmacotherapy
  Mostly anti cholinergics (Casey 2012)
  Alfa adreneg blockers (Abraham 2009)
Bladder neck incision (Kaplanzadeh 2007, Sarin 2013)
CIC (Holmdahl 2003)
Bladder augmentation (pre or post tx)
  Bowel (Jesus 2015) or ureter (Johal 2008)

Problems with CIC
- Disturbed anatomy
- Normal sensitivity
- Risk of strictures, epididymitis and UTI
- Compliance to treatment, especially during adolescence
- Other options:
  - Mitrofanoff
  - Vesicostomy button

Boy with PUV born 1990

- Valves, reflux, renal insufficiency from start
- CIC from 1 year of age
- 4 y of age bladder augmentation with ureter
- 5 y of age Tx
- Situation "under control" for about 5-10 years...

Cont...

- 15 y of age big problems non-compliance with CIC, decreasing renal function, polyuric, SP drainage
- 16 y of age Mitrofanoff, Pelvic abscess, respiratory problems, dialysis
- 17 y of age new Tx
- 18 y of age - transition to adult care, non-compliance with CIC, polyuric, decreasing renal function
Take home PUV

Control and treat renal/bladder dysfunction life long

- creatinine/s
- protein/u
- blood pressure

Bladder work up with invasive urodynamics and active treatment in case of decreasing renal function/transplantation
Neurogenic bladder dysfunction in Children and Adolescents

Kate Abrahamsson
Professor, Senior Consultant Pediatric Urology
The Queen Silvia Children’s Hospital
Göteborg

Gothenburg experience of continence surgery at Neurogenic Bladder Dysfunction (NBD)

Indication for incontinence surgery in children and adolescents with NBD

• High detrusor pressure
  • Nowadays very rare cause due to potent anticholinergic drugs
    • Oxybutynin (Ditropan® etc., Oxybutynin® etc.), Tolterodine (Detrusitol®), Solifenacin (Vesicare®) or Fesoterodin (Toviaz®)
    • Severe overactivity, Anticholinergics in combination with Mirabegron (Betmiga®)
  • Botulin; children need anesthesia
    • Good effect at overactivity
    • Worse effect at small capacity bladders and stiff detrusors
• Insufficient sphincter activity (individual need of Leak Point Pressure)

Why should we perform continence surgery in children/adolescents with NBD due to Spina bifida?

Using generic HRQoL measures, urinary/fecal incontinence and their medical management may not play a determinant role in HRQoL of persons with SB. However many other factors affect HRQoL in these patients.

Lemelle, Lottmann, Mouriquand Qual Life Res 2006

Participation in social life and physical intimacy

• 25 adolescents 16-18 yrs (all in the age range in Western Sweden)
• 17/25 continent (9/17, continence surgery)
• 8/25 incontinent

Arnell Vu Minh, M et al J Pediatr Urol 2019

Participation in social life and physical intimacy

Arnell Vu Minh, M et al J Pediatr Urol 2019
Participation in social life and physical intimacy

<table>
<thead>
<tr>
<th></th>
<th>Sex (M, F)</th>
<th>Urine continent</th>
<th>Spend free time with friends</th>
<th>Sleep overnight</th>
<th>Partner</th>
<th>Sexual debut</th>
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<td>M</td>
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<td>F</td>
<td>Y (6)</td>
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</tr>
</tbody>
</table>

Szymanski et al. J Urol 2018

Urinary incontinence decreases HRQoL from the age of 10 yrs

Results after continence surgery at NBD in Gothenburg Period 2000-2017

- 67 patients
- Mean age at surgery 11.5 yrs (range 5-20)
- Follow-up; mean 4.9 yrs (2-11)
- Diagnose – NBD
  - Myelomeningocele 50
  - Occult spina bifida 13
  - Spinal cord trauma 2
  - Cerebral Palsy 1
  - Trisomy 1

Methods

- Surgery
  - Low pressure reservoir
  - Basler augmentation
  - Increased outlet resistance
  - Basler neck plasty
  - Alternative CIC-channel
  - Mitrofanoff/Monti

- Urotherapy
  - Motivation
  - Instructions
  - Continuity
  - Availability

Benchmarking:

Neuropathic Bladder and Augmentation Cystoplasty, Review
Joshua D. Roth, MD*, Mark P. Cain, MD
Ileocystoplasty

Side effects
- B12 deficiency
- Infection
- Perforation
- Stone formation

ICS 2019

Bladder augmentation 53/67 patients

- Complications in Göteborg
  - Stones 6/53 patients 11%
  - Perforation 1/53 patients 2%
  - Ileus 2/67 patients 2%

- International results
  - Stones 6-52%
  - Perforation 6-14%
  - Ileus 3-11%

ICS 2019

Alternative CIC channel

- Mitrofanoff
- Appendicovesicostomy
- Monti (from small intestine when appendix is unusable)

- Always needed at ileocystoplasty and bladder neckplasty

ICS 2019

CIC-channel 66 patients, 69 channels

- Mitrofanoff 52 (75%)
- Monty (spiralmonty) 17 (25%)

ICS 2019

Redo-surgery of the CIC-channel

Göteborg (66 pat)
- Total 6 pat (9%), 9 episodes (13%)
  - Stoma revisions Mi 4 (7,6%) Median 0,7 yrs
  - Subfascial revisions 5 (7,7%) 2 Mo (17,2%) Median 2,3 yrs

Indianapolis (500 pat)
- Stoma revisions Mi 13,7% Median 1,2 yrs
  - Subfascial revisions Mi 0,5% Median 2,2 yrs
  - Subfascial revisions Mi 16,0% Median 2,3 yrs

Szymanski et al J Pediatr Urol 2015

ICS 2019

Bladder neck plasty; Young Dees

- YD
  - Urethral elongation and narrowing
  - Leakage at high abdominal pressure
  - Not in use at NBD after 2005
Kropp procedure

Elongation of urethra
Implantation in the posterior bladder wall
Completely continent

Increased outlet resistance; 42 patients

- Total 5/42 ruptures (12%)
- YD 12
  - Spontaneous rupture 2/12 (17%)
- Kropp 28
  - Spontaneous rupture 1/28 (3.6%)
  - Rupture due to high detrusor pressure 2/28 (7.2%)
- (Isolated sling 1)
- (Bladder neck closure 1)

Results from experiences in Göteborg

- Continence surgery at NBD is advanced with a high complication rate
  - ~80% of the patients – no need for redo-surgery during childhood
  - ~10% redo-surgery at one occasion or more

Suggestions from experiences in Göteborg

- To decrease the complication rate, an experienced team (urotherapist/urologist) that works closely together seems to be of importance
- Intensive preventive care needs to be performed mainly by the urotherapists
- HRQoL and social functioning are improved by urinary continence

"Take home" consideration

Would a closer surveillance by uro-therapists prevent complications in adults with NBD after continence surgery provided during childhood?
BLADDER EXSTROPHY CONCERNS

Giovanni Mosiello
Rome, Italy

EXSTROPHY-epySpadia

1906 TRENDELEMBURG
First attempt of reconstruction

1950 JEFFS RD
STAGED APPROACH:
1) Bladder closure
2) Bladder neck (BN), residual epispadias

1999 MITCHELL M
COMPLETE PRIMARY REPAIR
OF EXSTROPHY (CPRE)

Exstrophy: perineal anomalies

- congenital
- iatrogenic
  - puborectal muscles
  - connective tissue
  - tendineal tissue
  - sphincter innervation

FOLLOW-UP EXSTROPHY-EPYSPADIA

<table>
<thead>
<tr>
<th>Patient</th>
<th>% CONTINENCE *</th>
<th>% MICTURITION</th>
</tr>
</thead>
<tbody>
<tr>
<td>% CHILDREN</td>
<td>80</td>
<td>84</td>
</tr>
<tr>
<td>% ADULTS</td>
<td>40</td>
<td>14</td>
</tr>
</tbody>
</table>

* Dry > 2 hours

- BLADDER AUGMENTATION 0-70% CHILDREN
- CANCER RISK

WOODHAUSEN CR, North AG, Gehrheit JP
Sedating test of time: long-term outcome of reconstruction
of the exstrophy bladder
World J Urol. 26: 344-8, 2006

FOLLOW-UP EXSTROPHY-EPYSPADIA

ADULT AGE

<table>
<thead>
<tr>
<th>Year</th>
<th>Patient</th>
<th>% CONTINENCE</th>
<th>% MICTURITION</th>
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<tbody>
<tr>
<td>1997</td>
<td>Lottmann HB</td>
<td>57</td>
<td>67</td>
</tr>
<tr>
<td>2001</td>
<td>Capeloschi G</td>
<td>59</td>
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<tr>
<td>2003</td>
<td>Surier I</td>
<td>68</td>
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</tr>
<tr>
<td>2005</td>
<td>Mouriquand PD</td>
<td>80</td>
<td>45</td>
</tr>
</tbody>
</table>

GIOVANNI MOSIELLO, MD, FEAPU, FEBPS

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

Funding for speaker to attend:

Self-Funded

Institution (non-industry) funded

Sponsored by:
**Pelvis reconstruction at birth**

**EXSTROPHY: Skeletal Defects**

- Diastasis of the symphysis pubis
- External rotation ilium (mean 12°)
- External rotation anterior pelvis (mean 18°), shortened 30%

- Acetabulum: retroversion
- Increased distance between the triradiate cartilage

**EXSTROPHY: CONTINENCE OUTCOME**

Percentage of children Born with exstrophy who are continent (principle series over the last century)

**Complex primary Repair**

- CPRE (Mitchell J Urol 162:1415, 1999)
  - Bladder deeply into pelvis
  - Bladder neck reconstruction
  - Proximal urethra curve (genital ducts into post urethra)
  - Corpus spongiosus conserved
  - Urethra under corpora

**In one shoot**
Bladder capacity following CPRE

Prospective Follow-up in Patients After Complete Primary Repair of Bladder Exstrophy

J Urol, Oct 2008

❖ 32 CPRE, 1994-2007
❖ 16/32 antenatal diagnosis
❖ 6 BN surgery, 3 bulking
❖ no bladder augmentation
❖ 8/21 continence >2 hours
❖ PPCR= Percent Predicted BC/age

Patients:
- 20 patients (18 boys), closure mean 9.9 mos, osteotomy, total TPN 10/12 pts > 2000, mean hospital stay 6.3 wks
- Results: average follow-up 7.4 years: 100% successful

Indication for delay (n)
- Small bladder template 15/20 (75)
- Prematurity 2/20 (10)
- Late referral 2/20 (10)
- Neural tube defect 1/20 (5)

8/21 continence >2 hours

Patients: 20 patients (18 boys), closure mean 9.9 mos, osteotomy, total TPN 10/12 pts > 2000, mean hospital stay 6.3 wks
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- Late referral 2/20 (10)
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Cervellione RM, Bianchi A, Fishwick J, Gaskeff SL, Dickson AP
Salvage procedure to achieve continence after failed bladder exstrophy repair
J Urol. 179:1393-6, 2008

ESTROFIA-EPISPADIA
Surgery for continence after "staged repair"

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Continent</th>
<th>Incontinent</th>
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<tr>
<td>Augmentation + CIC</td>
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<td>0</td>
</tr>
<tr>
<td>Augmentation + BN reconst + CIC</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Substitution + CIC</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Augmentation + BN closure + CIC</td>
<td>10</td>
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<tr>
<td>Derivation</td>
<td>4</td>
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<tr>
<td>Main ii pouch</td>
<td>2</td>
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<tr>
<td>Totals</td>
<td>20 (90%)</td>
<td>3 (10%)</td>
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</table>

Male Bladder Exstrophy

Pelvic Floor Continence

Penile length and genitalia ducts

Male Exstrophy/Epispadias:
Long-term Genital Function

- Semen: scarce
- Ejaculation: 75%
- Paternity: 50% who wished, successful
- Sexuality: normal desire
Genital and Reproductive Function in Males After Functional Reconstruction of the Exstrophy-Episadias Complex—Long-Term Results

- Single-stage reconstruction (CPRE)
- consequent placement of the collicolus seminalis in the posterior urethra
- normal ejaculation in 94.1%
(but severely impaired sperm quality and hormonal problems)

Long-term Follow-up (18-35 years) of Male Patients with History of Bladder Exstrophy (BE) Repair in Childhood: Erectile Function and Fertility Potential Outcome

**Patients:**
- 30, 18-35 (avg 26) years,
- 10 diversion, 15 staged repair, 5 complete primary

**Results:**
- Erections 28/30 (IIEF); penile length - (7.65cm)
- 10/30 married; 4/10 children (1/4 assisted reprod)
- 20/30 not married: 6 inadequacy feeling, 10 cosmetic
- retrogr ejac 16/30, low volume 8/30, anejaculation 6/30

**Epispadia female**

- Labia introitus low abdominal wall
- Anus in the position of the vagina
Exstrophy: gynaecological concerns

- **50% uterine prolapse**
  - In many cases amounting to procedentia
  - Introitus narrow
  - After surgical enlargement, girls have normal fertility

- **16 women, 8 married, 5/8 delivered 8 children, 10 required uterine prolapse repair**

- **as the repair involves a sling around the cervix, further pregnancy may be difficult**

- **Woodhouse CRJ, The gynaecology of exstrophy – BJU, 1999**

Pregnancy after lower urinary tract reconstruction for congenital abnormalities

- **20 women/28 babies**
  - 7 Exstrophies
  - 7 Spinal Dysraphism
  - 2 Sacral agenesis
  - 1 Cerebral palsy
  - 1 Imperforate anus
  - 1 Small bladder/VUR

- **TABLE 4 The risk of prolapse**

<table>
<thead>
<tr>
<th>Prolapso</th>
<th>Syntesis closure</th>
<th>Open pelvis</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>No</td>
<td>11</td>
<td>3</td>
<td>14</td>
</tr>
<tr>
<td>Yes</td>
<td>1</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>12</td>
<td>7</td>
<td>19</td>
</tr>
</tbody>
</table>

*Statistically significant, P < 0.02

- **COMPLICATIONS**
  - 15 UTI
  - 3 UT obstruct.
  - 2 Mitrofanoff difficulties
  - 1 pyelonephritis

- **DELIVERY**
  - 10 vaginal
  - 7 emergencies
  - 12 cesarian

Quality of Life in Adults With Bladder Exstrophy-Episadipsis Complex

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

<table>
<thead>
<tr>
<th>Health Concepts</th>
<th>Mean ± SD</th>
<th>Mean ± SD</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>DEC</td>
<td>NORMATIVE</td>
<td></td>
</tr>
<tr>
<td>General health</td>
<td>56.4 ± 24</td>
<td>69.1 ± 10.0</td>
<td>0.006</td>
</tr>
<tr>
<td>Physical functioning</td>
<td>76.6 ± 3.9</td>
<td>84.5 ± 21.2</td>
<td>0.001</td>
</tr>
<tr>
<td>R wel-physical</td>
<td>8.6 ± 29</td>
<td>81.2 ± 32.2</td>
<td>0.4</td>
</tr>
<tr>
<td>Rest-physical</td>
<td>30.3 ± 16</td>
<td>82.1 ± 32.2</td>
<td>0.21</td>
</tr>
<tr>
<td>Social functioning</td>
<td>80.5 ± 28.1</td>
<td>66.6 ± 21.4</td>
<td>0.01</td>
</tr>
<tr>
<td>Bodily pain</td>
<td>70.4 ± 29</td>
<td>73.4 ± 23.7</td>
<td>0.67</td>
</tr>
<tr>
<td>Vitality</td>
<td>53.8 ± 34.4</td>
<td>59.9 ± 18.05</td>
<td>0.74</td>
</tr>
<tr>
<td>Mental health</td>
<td>72.5 ± 25.8</td>
<td>68.5 ± 17.8</td>
<td>0.25</td>
</tr>
</tbody>
</table>
CLOACA CONCERNS

Giovanni Mosiello
Rome, Italy

Persistent Cloaca: common channel, different length

Surgery

Common channel > 3 CM
need to separate vagina/urethra
common channel to urethra, vaginal reconstruction/substitution
69% CIC

Common channel < 3 CM: posterior sagittal anorecto
vagino urethroplasty (total urogenital mobilization)
19% will require CIC

Concerns similar to Uro-Genital Sinus

Location of the vaginal confluence
determined endoscopically - RINK

Distance from the bladder neck to the confluence
Distance from the confluence to the external meatus (UGS length)
Concerns Adolescents

1. UTI: urinary retention
2. Incontinence
3. Stenosis
4. Fistula

ASSOCIATED MALFORMATION

- Vesico ureteral Reflux
- Renal Agenesia/Hypoplasia
- Hydronefrosis
- Duplex system
- Vaginal malformation: 40% two vaginas, 15% agenesis
  Spina Bifida

Table 2
Urinary incontinence in adolescent and adults with CHD

<table>
<thead>
<tr>
<th>Reference</th>
<th>N.</th>
<th>Age (range)</th>
<th>Time of follow-up (range)</th>
<th>No. continent Total No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Davies et al (2010)</td>
<td>80</td>
<td>14-30</td>
<td>2-8</td>
<td>80/87 (85.7)</td>
</tr>
<tr>
<td>Couchman et al (2010)</td>
<td>80</td>
<td>14-30</td>
<td>2-8</td>
<td>80/87 (85.7)</td>
</tr>
<tr>
<td>Rintala (2015)</td>
<td>47</td>
<td>10-30</td>
<td>2-8</td>
<td>47/53 (85.7)</td>
</tr>
</tbody>
</table>

Continence after TUM and PUM Palmer B 2012, J Urol

Prognosis and Channel Length, Levitt M, Sem Ped Surg 2010
Long-term urological outcomes in cloacal anomalies
Brian T. Caldwell, MD*, Duncan T. Wilkie, MD, MBBS

Constipation often requires intermittent catheterization and in some cases, bladder augmentation. The complexity of cloacal malformations and associated anomalies make long-term multidisciplinary follow-up imperative.

Conclusions
Persistent cloaca is still a challenge for pediatric surgeons and urologists. Despite optimal operations that are performed by very experienced surgeons, significant functional disturbances remain in many adolescent or adult patients with cloaca. Approximately, half of the patients gain spontaneous fecal continence as they grow through childhood. The remaining half require adjunctive measures to stay clean. Adolescents and adults report similar spontaneous urinary continence rates; the rest of the patients stay dry mainly by continent urinary stomas or intermittent catheterization. Genital tract problems are common in puberty, many patients require secondary procedures to establish patient genital tract that enables uncomplicated maturations and sexual life.
Lost in transition? from a urotherapeutic point of view

Magdalena Vu Minh Arnell
R.N.
Ped.Urotherapist
Urotherapeutic unit
Sahlgrenska University Hospital
Queen Silvias Children’s Hospital
Regional Rehabilitation Centre

Urotherapeutic unit specialized on neurogenic bladder- and bowel dysfunction in Gothenburg

Outpatient clinic: 100 children with Spina Bifida 0-18 yrs
A team: R.N. pediatric urotherapist
pediatric urologist
pediatric neurologist
pediatric colorectal surgeon
occupational therapist
rehabilitation assistant

Spina Bifida; incidence in Sweden

- In 2016
  - a total of 121500 children were born
  - 29 (2.39/10000) were born with Spina Bifida
  - 24 fetuses with Spina Bifida were aborted

Neurogenic bladder and bowel dysfunction in individuals with Spina Bifida

Insufficient emptying of bladder/bowel
No automatic emptying - injured nerve function
Urotherapists/nurses - 2 main focus- in need of lifelong follow-up !!

Pediatric care

All children and adolescents are followed according to a national structured protocol
No cases of reduced renal function at the age of 18 year

Family-centered nursing perspective to reach the best possible treatment results
The contact with the family is based on:
- continuity, “personal” urotherapist
- respect
- cooperation
- support

GOAL for Adolescent care to become independent in bladder and bowel treatments

Conversation with the urotherapist without parent
Education about the body and the treatments
Discuss friendship, integrity, sexuality
Raise the level of awareness of being a young person with a disability
Good knowledge about your self is a prerequisite for independence
**Definition of transition**

- **Transfer** is described as the time when the responsibility for care transfers from one medical unit to another.

- **Transition** involves significantly more than the physical transfer between the clinics.

Transition is defined by Blum as “a purposeful, planned movement of adolescent and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems.”


**ICS september 2019**

- Transition may involve medical, psychological, and social support.

- Each child’s journey through transition is unique.

- Parents’ role is to be part of the process but to start to step back so that the child gets increasing experience and independence to make own health care decisions.


- **ICS september 2019**

**Two studies**

<table>
<thead>
<tr>
<th>Adults 27-50 yrs</th>
<th>Teenagers 16-18 yrs</th>
</tr>
</thead>
<tbody>
<tr>
<td>69 individuals (all in the age-range)</td>
<td>25 individuals (all in the age-range)</td>
</tr>
<tr>
<td>37 f + 32 m</td>
<td>10 f + 15 m</td>
</tr>
<tr>
<td>Median age 34 yrs</td>
<td>Median age not specified</td>
</tr>
</tbody>
</table>


- *Urinary continence appears to enhance social participation and bladder and bowel management* *J Pediatr Urol.* 2019

**ICS september 2019**

**Elimination/Continence**

<table>
<thead>
<tr>
<th>Adults 27-50 yrs</th>
<th>Teenagers 16-18 yrs</th>
</tr>
</thead>
<tbody>
<tr>
<td>70% CIC</td>
<td>100% CIC</td>
</tr>
<tr>
<td>35% - urinary continent</td>
<td>68% - urinary continent</td>
</tr>
<tr>
<td>80% - used pads</td>
<td>44% - used pads</td>
</tr>
<tr>
<td>14% - urotherapy support</td>
<td>100% - urotherapy support</td>
</tr>
<tr>
<td>0% - support for fecal elimination</td>
<td>100% - support for fecal elimination</td>
</tr>
</tbody>
</table>

**ICS september 2019**

**Transition from pediatric care to adult care**

- Pediatric care could begin the transition earlier and the transition to adult care should be individually based.

- Raise the competence about how to support these patients.

- Urotherapy educated and interested nurse - the link to the urologist

- Follow-up programs even for adults (coming in fall 2019)

- A wish: “Young adult transition unit” 18-25 years, we are working together pediatric care and adult care when the individuals, based on their potential and maturity – are well prepared for transition, then it is time…. 

**ICS september 2019**

- Diapers not pads.

- Same catheter size as when they were kids

- Fecal incontinence - no enema (manual removal, not independent)

- Ends with CIC (hard to get the right products)
Thanks for your attention!

magdalena.vuminh-arnell@vgregion.se
Surgical procedures in adolescents with SB, extrophy etc.
tips and tricks

Prof Rien JM Nijman FEBU
Dept of Urology and
Paediatric urology
University Medical Centre
Groningen
The Netherlands

ICS workshop, 2019

Tips and tricks

• It is all about the bladder, the sphincter + CNS
• It is absent or not working properly
• Epispadias / extrophy / cloacal malformations
  - Normal dexterity / normal cognitive function
• Spina bifida
  - Wheelchair bound
  - Abnormal dexterity
  - Abnormal cognitive function
  ➔ individualized care + plan

Tips and tricks

But they all have in common
- To become as independent as possible
- To go to school
- To go to “work” if possible
- To go without a diaper
- To look as normal as possible / good cosmesis

➔ To improve quality of life

Tips and tricks

If cognition fails
- The caregivers + surroundings + health care system
determine what you can do safely
- What we can do in US/Europe etc. can not be done
  in sub-Saharan Africa etc.

Our patients become older
- They may end up in retirement homes
- They may become completely dependent again
- At that stage a continent diversion may become
dangerous....

Tips and tricks

Objectives:
• 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: 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
Bladder reconstruction: techniques

- 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 / adolescent is ready for it (counseling is extremely important)
- When the parents / care givers are ready
- For improvement of QoL

Tips and tricks

- Epispadias in females: urethral + BN reconstruction in one procedure
  - Ventral approach
  - Do not separate urethra from the vagina
  - Reconstruct pelvic floor as in exstrophy
- Normal voiding + continence can be achieved in 80 %
- In all female patients who have had BN reconstruction
  - Pregnancy is on the agenda
  - A vaginal delivery may compromise the BN → CS

Tips and tricks

- Exstrophy reconstruction of the pelvic floor is important to
  - prevent uterine prolapse in females
  - improve continence
- Closure of the pelvic ring helps
  - continence
  - preventing uterine prolapse
  - lengthening the penis
  - improve cosmesis
Sometimes a Mainz 2 pouch is better than a continent diversion

Urinary continent diversion

- Uretosigmoidostomy
- Mainz pouch II / Sigma pouch
  - Consistent series (128 patients)
  - Continence rate: 95%
  - Renal preservation rate: 92%
  - Long-term follow-up (10 years)

Exstrophy / epispadias in adolescents

- Before pelvic reconstruction
- Following pelvic reconstruction
Exstrophy / epispadias in adolescents

18 yrs old male
Continent diversion +
incontinence + hernia 8 cm

Exstrophy / epispadias in adolescents

Reconstruction using mesh
- Risk of infection
- Risk of recurrence

Tips and tricks

Neurogenic bladders
- Early start of CIC + anticholinergics
  → 70% will be dry
- Need for augmentation decreases
- Different methods to treat bladder and sphincter surgically
  • Augmentation
  • BN reconstruction / plasty
  • Catheterizable channel

Neuropathic bladder in adolescents

continent diversion

Independent
Less time consuming
Improves self-esteem

Tips and tricks

Augmenting the bladder
• Provide reservoir of at least 400 ml
• Use ureter or ileum
• Irrigate bladder at least once a day with 250 ml (saline or water), use active irrigation
  → Less stones / less UTIs
• Close and life-long follow-up is mandatory!!
• Cave secondary malignancies / stones / metabolic abnormalities

In wheelchair bound patient always consider continent catheterizable channel

Tips and tricks

Augmentation

Surgical Complications: 20 – 50%
### Tips and tricks

#### Augmentation Ileum / Colon
- Compliance: Ileum < Colon
- Contraction: Ileum < Colon
- Mucous: Ileum < Colon
- Relation to the Bladder: Ileum < Colon
- Ureter re-implantation: Ileum < Colon
- Met. Acidosis: Ileum < Colon
- Bile Acid: Ileum > Colon
- Vitamin B₁₂: Ileum > Colon

#### Ureterocystoplasty
- Severely dilated Ureter & non-functioning kidney
- Re-Augmentation up to 81% (21/26)
  - Careful selection of the patients

### Tips and tricks

#### Catheterizable channel
- Keep it as short as possible
- Make it straight
- Give good support
- Fix it to abdominal wall
- Use the native bladder back wall to re-implant
- Make it suitable for Fr 12-14 catheter

#### Mitrofanoff principle
- Straight
- Supple
- Short
- Supported
  - 4 x S !!
- Appendix
- Bowel segment (Monti)
- Ureter / preputial tube / bladder tube

#### Continent Vesicostomy / Ileal Nipple
- 10 - 12 cm
- Stable Continence Mechanism
  - e.g. disabled patients (fine motor skills - AC-II Malformation)

#### Cave diverticula and kinking in the long-term follow-up

---

Yang 1993, Monti 1997, Ghoneim 2005

Stein 2000, Thüroff 2005
Tips and tricks

- Continent Cutaneous Diversion
  - Continence: > 90%
  - Complication: 30% - 50%
  - Stomal stenosis
  - Ureterointestinal Obstruction
  - Stone formation
  - Incontinence

Stomal principle

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

Avoid kinking

Mitrofanoff principle

Channel in umbilicus

Submucosal tunnel

Percent of Patients Without Stomal Revision

APV

Monti

p = 0.003
Opinions

- In adult patients expect more complications with APV / Monti
- Double Monti → significantly more problems
- In adolescents and adults use appendix if possible or go for a Mainz pouch / Sigma pouch
- Highly select pts for surgery based on compliance with intermittent catheterization
  - Beware of affects of alcohol/drugs on compliance*

  *Fox et al, J of Urology, 2010

conclusions

- primary objective is to preserve upper tract + to become dry + become independent + improve QoL
- only do the operation, if the patient is ready for it
- you may have to go back for 2nd procedure: use omentum to cover the bladder / fix pouch so you can have easy percutaneous access (stones)
- Patient should be followed-up life-long and widely informed about possible complications

Not everything that is possible is useful for the patient

Individual and interdisciplinary concepts

thank you
What we hope for!

"The best medical centres and faculties of Europe are connecting in 24 networks composed of nearly a thousand of centres of expertise or highly specialized healthcare providers. The newly established ERN will have a substantial impact on medical practice. In short, it marks the start of a new era in cross-border cooperation and in healthcare organisation"


What is an European Reference Networks (ERN)?

- virtual networks involving healthcare providers across Europe
- one healthcare provider is the Network Coordinator, and the main responsible center
- aim is to tackle complex or rare medical diseases or conditions that require highly specialized treatment and a concentration of knowledge and resources

Treatment of patients with rare or complex diseases

ERNs are not directly accessible to individual patients

- With the patients’ consent and in accordance with the rules of their national health system, the patient’s case can be referred to the relevant ERN member in their country by their healthcare provider
ERNs are an exciting new form of cooperation at European level between healthcare providers with specialised expertise with the aim of improving care for patients with rare diseases or complex conditions to:

- Improve diagnosis
- Allow more equitable access to high-quality treatment and care

**eUROGEN** is one of the 24 European Reference Networks (ERNs) approved and supported by the European Commission in 11 European Member States.

**Thematic areas of expertise:**
- Urorectal/anorectal malformations
- Bladder exstrophy/epispadias
- Posterior hypospadias
- Complex genital reconstructions (DSDs)
- Non-syndromic, urogenital tract malformation
• HCP Interests: CPMS

**Cleacal Exstrophy – 14 mths old boy**

- Generalised:
  - Renal hypoplasia both side
  - Portal hepaticovenous stenosis, portal hypertension
  - Anorectal malformations, mega urethra, bifid scrotum
  - Imperforate bladder/rectum

  * MRI requisition:
  - 1. Bladder and rectal abnormalities
  - 2. Uterine and vaginal abnormalities

  * Anatomy:
  - Dorsal hypoplasia of the urethra with no tunica propria
  - Double diverticula (2:1)
  - Internal meatal symmetrical

  * Surgery:
  - Urethral reconstruction
  - Bladder augmentation
  - Vaginal and rectal repair
  - Urethrocutaneous neostomostomy
  - Full thickness skin grafts

**On arrival: 6 months of age**

- Chest X-ray: hemivertebra on D4
- Pelvic X-ray:
  - AP Sacrum Ratio 1.2
  - Pubic diastasis 45mm
  - Dysmorphic S3, S4 e S5

**Total body MRI: spinal dysraphism, myelomeningocele, caudal regression**

### Surgical procedures

- At 7 months: Right ureterocutaneostomy
- At 8 months: Bladder exstrophy repair (1 step)
  - Exstrophic elephant trunk dissection and ileo-colic reconstruction, terminal colostomy
  - Previous ileostomy closure
  - Two hemi-bladder approximation in one bladder plate
  - Bilateral inguinal orchidopexy
  - Partial abdominal wall closure with Gore-tex patch

**Post-operative / step complications**

- Sepsis
- Wound dehiscence with abdominal patch exposure

- Daily medications → IV therapy → Wound sterilisation → Advanced medications → Partial wound healing
Bladder extrophy repair (II step) at 13 months

- Anterior bilateral osteotomy
- Bladder reconstruction
- Pubic dyastasis approximation
- Complete abdominal repair with no patch
- Spica cast (immobilization)

Post II step complications (at 14 months)

- Wound infection
- Partial bladder prolapse
- Progressive left ureteronephrosis
- Urinoma
- Partial recurrence of pubic dyastasis (50mm)

Daily advanced medications
- Left ureteronephreectomy (at 15 months)

Summarizing today

At 18 months:
- Terminal colostomy (firm stools)
- Bowel management (clean for 12 hours)
- Bilateral ureterostomy (no UTI)
- Partial bladder prolapage with good anatomical bladder capacity
- Persistent pubic dyastasis
- Caudal regression
- Inguinal testicular pull
- Complete epispadias

Questions

- What's the usual age for surgery?
- What's the usual age for bowel management?
- What's the usual age for bilateral ureterostomy?
- What's the usual age for partial bladder prolapage?
- What's the usual age for persistent pubic dyastasis?
- What's the usual age for caudal regression?
- What's the usual age for inguinal testicular pull?
- What's the usual age for complete epispadias?

At birth

At discharge: 18 months

- Does surgery require blood draw?
- Does surgery require a parent to stay overnight?
For the urological part I would be careful with the reconstructions and also consider an incontinent diversion. There is also a neuropathic bladder component. The bladder might not be safe. There is already a loss of kidney function.

One could consider if feasible with the abdominal wall a second stoma for urine diversion. The bladder could be closed and an epispidias repair if feasible and orchidopexy without ureteral reimplantation.

The prostate should have external opening.

I do not see the need for additional osteotomy. Happy to discuss in video conference.

Wait with epispidias till after puberty and check at that time length and decide at that time for reconstruction of phalloplasty.

**Conclusions**

We treasured these tips, though different in this very complex and rare case 1:150,000...

ERN and CPMS: an exciting new form of cooperation between healthcare providers with specialised expertise.

Can we imagine to translate it worldwide?