

W7: Strategies for Incontinence Management in Congenital Pathologies from Childhood to Adulthood

Workshop Chair: Giovanni Mosiello, Italy

26 August 2013 14:00 - 18:00

Start	End	Topic	Speakers
14:00	14:15	Introduction and presentation of childhood pathologies requiring continence treatment during childhood: spina bifida, posterior urethral valves, bladder exstrophy	<ul style="list-style-type: none"> Giovanni Mosiello
14:15	14:30	GUIDELINES AND TREATMENT STRATEGIES	<ul style="list-style-type: none"> Mario De Gennaro
14:30	14:30	MEDICAL AND SURGICAL STRATEGIES FOR UNREMITTING SYMPTOMS	None
14:30	14:45	PHARMACOTHERAPY AND REHABILITATION	<ul style="list-style-type: none"> Laetitia De Kort
14:45	15:00	Mini-invasive treatment: role of botulinum toxin, sacral neuromodulation, endoscopic procedures	<ul style="list-style-type: none"> Giovanni Mosiello
15:00	15:15	SURGERY: AUGMENTATION, DERIVATION, SLING	<ul style="list-style-type: none"> Laetitia De Kort
15:15	15:30	CONSERVATIVE MANAGEMENT VERSUS SURGICAL MANAGEMENT : CLINICAL CONSIDERATION, ADVANTAGES AND DISADVANTAGES	<ul style="list-style-type: none"> Wendy Bower
15:30	16:00	Break	None
16:00	16:15	Questions	All
16:15	16:30	Adult life: the neuro-urologist point of view	<ul style="list-style-type: none"> Giulio Del Popolo
16:30	16:45	Adult life : the uro-gynecologist point of view	<ul style="list-style-type: none"> Pallavi Latthe
16:45	17:00	Discussion	All
17:00	17:45	CASE PRESENTATION AND SHORT BREAK	All
17:45	18:00	DISCUSSION AND TAKE HOME MESSAGES	<ul style="list-style-type: none"> Giovanni Mosiello

Aims of course/workshop

Spina bifida, bladder exstrophy/epispadia, 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 to ameliorate the treatment of continence lifelong.

Title: STRATEGIES FOR INCONTINENCE MANAGEMENT IN CONGENITAL PATHOLOGIES FROM CHILDHOOD TO ADULTHOOD

Category: **Pediatrics**
Chair : **Giovanni Mosiello**
Length: **180**
Level: **Advanced**

INTRODUCTION AND PRESENTATION OF CHILDHOOD PATHOLOGIES REQUIRING CONTINENCE TREATMENT DURING CHILDHOOD: SPINA BIFIDA, POSTERIOR URETHRAL VALVES, BLADDER EXSTROPHY.

GIOVANNI MOSIELLO , ITALY

Patients with complex congenital genitourinary pathologies (CCGUP) as spina bifida, bladder exstrophy, anorectal malformations, posterior urethral valves, require lifelong evaluation and treatment, especially for continence. Different treatment options in order to ameliorate continence can be considered after the first surgical treatment in the newborn period, 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, to discuss about the concerns for a correct choice in the management in childhood is related to individual condition of the single patient , because actually with the improvement in care of the pediatric urology, patients with CGUP survive into adulthood, and a correct choice means to avoid procedure that can impair in adult life aspects as fertility and pregnancy. Then summarizing there are some 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.

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SPINA BIFIDA

Pediatric Spinal cord lesions (SCL), can be classified considering congenital and acquired lesions. In the congenital lesion the most common type is spina bifida (SB) open or closed . SB is a congenital neural tube defects that actually in many countries is less common then.in the past, due to folic

acid supplement during pregnancy, as well as for prenatal diagnosis and related consequences, and rates of SB vary in the different countries approximately 8/10,000 live births are affected by SB.

The term SB includes: occulta SB, or occult spinal dysraphism (OSD) myelomeningocele (MMC), meningocele and lipomeningocele. The most common location of the malformations is the lumbar and sacral areas and MMC is the most significant form, often associated to hydrocephalus. Disability is mainly related to the level of lesion and open or closed defect.

While the diagnosis of MMC is in the newborn age, the diagnosis of OSD could be done in different ages, mainly for urinary tract infections, urinary incontinence, etc. with a typical natural history in infants children and adolescents.

SCL in pediatrics are not only related to congenital lesions, because we have to consider spinal cord injury (SCI) can be related to car accident, sports trauma, surgery and medical treatment. SCI in pediatric received scant attention due to its rarity. In Europe the incidence is documented in Portugal and Sweden (27 children/million children/year and 4.6 children/million children/year respectively). For the other countries, the estimated incidence of pedSCI varied from 0.9-21.2 children/ million children/year in the ages 0-14 years. The management differs between the countries depending on the age of the child and the local organization of health care. Knowledge of incidence and etiology of pedSCI is important in Ped SCI is unique with a high percentage of highcervical spinal cord injuries, very often associated to head trauma or politrauma. Particularly unique to children is the lap belt injury. Theoretically the neuro-urological approach is the same in children with spinal cord injury (SCI) and congenital lesions as spina bifida (SB), because urological goals are the same: to establish a satisfactory bladder emptying, to maintaining a safe bladder storage pressure, avoiding infections, in order to prevent upper urinary tract deterioration, with the final goal to reach continence. The importance of neurogenic bladder dysfunction (NBD) has been well described, renal damage remains a real risk and children require a careful evaluation and follow-up because NBD change during time. T

According to the nature of the neurological deficit, the bladder and sphincter may be in either an overactive or inactive state: the bladder may be overactive with increased contractions, and low capacity and compliance, or inactive with no effective contractions; the outlet (urethra and sphincter) may be overactive causing obstruction, or paralysed with no resistance to urinary flow; these conditions may present in different combinations t.

Hip subluxation/dislocation can occur especially in congenital SCL or in SCI injured before age 10. Contractures of the joints, most commonly the hips, elbows, knees, and ankles, occur in almost all patients with SCL. So in the treatment of NBD we have to consider the other pathologies related to SCL as scoliosis, hydrocephalus and another concern of SCL patients is latex allergy. According to the Spina Bifida Association of America (SBAA), over 73 percent of people with spina bifida develop a latex allergy from mild to life-threatening. The common use of latex in medical facilities makes this a particularly serious concern, and the best solution is to treat children in a latex free environment. In conclusion in our mind a child should undergo rehabilitation and treatment including surgery in a specialist paediatric spinal centre to obtain maximum functional and quality of life gain.

Posterior urethral valves

Posterior Urethral Valves - PUV) are the most common cause of bladder outflow obstruction in male infants, and the most common obstructive uropathy leading to chronic renal failure in childhood. PUV incidence is estimated at 1/5000 to 1/8000 male births

Actually with the term of PUV are described any congenital obstructive obstacle, membrane or diaphragm, folds, web of the posterior urethra. PUV present a wide spectrum of gravity and from the more severe obstructive forms, with significant consequences on the upper urinary tract and renal function, to the minimally obstructive forms with minimal consequences on bladder function: urinary tract infection and voiding dysfunction

The most common classification is by Young,

type I: are a pair of leaflets origin from the lower border of the verumontanum, extend laterally and distally on the lateral wall of the membranous urethra, to fuse anteriorly at 12 o'clock,

type II, are folds that arise from upper border of the verumontanum and proceed proximally towards the bladder neck,

type III, are a congenital urethral diaphragm with a central perforation, not related to the verumontanum.

During time different classification were suggested as by Dewan and Ransley a unique form of PUV, which they called "congenital obstructing posterior urethral membrane" (COPUM) or Hendren's classification in 2 forms. Anyway it is important to know that as a consequence of the early urethral obstruction due to PUV, we will found a lower urinary tract dysfunction during life as related to prostate disease in adult life because bladder outlet obstruction determines hyperplasia and hypertrophy of the detrusor muscle, which appears significantly thickened. Furthermore in PUV the urinary apparatus above the membranous urethra may develop incorrectly during pregnancy because of the presence of distal obstruction and high pressure, of course the posterior urethra, the bladder, ureters and kidneys may show different degree of damage according to the severity of the urethral obstruction. 50% of patients with PUV show vesico-ureteral reflux (VUR), In about 20% of PUV patients, a unilateral high grade VUR with significant renal dysplasia and a contralateral normal kidney is found: this condition is termed "VURD syndrome" (posterior urethral Valves, Unilateral Reflux, Dysplasia) in a "pop-off" mechanism, relieving the pressure in the urinary system and therefore protecting the contralateral kidney. The most severe consequence of PUV is renal dysplasia caused either by a developmental defect either by high pressure. The majority of patients with PUV are actually diagnosed in the prenatal age. Newborn with PUV may present with failure to thrive, or urosperis, abdominal distension, vomiting, desidratation, renal failure with increased creatinin, acidosis, iperkaliemia.

In older children PUV are generally minor forms presenting urinary incontinence, enuresis, urinary tract infections. Lower urinary tract symptoms not responding to anticholinergics and associated with postvoid residual at the ultrasound should be investigated. In every age the first step is Ultrasound and the VCUG that is the gold standard for the diagnosis of PUV. In older children with voiding symptoms, the sonographic report of increased bladder thickness and significant post void urine residual will require further diagnostic investigations (VCUG, uroflowmetry cystoscopy

Regarding VCUG this exam permits to identify presence of diverticula, VUR, to evaluate the urethra during micturition, when the posterior urethra typically appears enlarged with a reduced diameter distal to the obstruction, with sometimes reflux in the seminal vesicles. In older children with minor forms VCUG could be apparently normal and some authors suggested uretroscistocopy in all.

Bladder exstrophy – epispadias complex (BEEC)

In BEEC the defect of the lower abdominal wall and genitalia is related to an abnormal development of the cloacal membrane, which would prevent medial migration of the mesenchymal tissue and proper lower abdominal wall development. Some authors suggested that the timing of the rupture of this defective cloacal membrane determines the resulting variant of the

exstrophy-epispadias complex. Absence of migration, ascent, alignment of the allantois with the yolk sac with its persistence at the dome of the cloaca could be able to explain the bowel location in cloacal exstrophy. The exstrophy-epispadias complex ranges from simple distal epispadias, genital defect to a multisystemic defect of the cloacal exstrophy type, involving all the abdominal wall.

Classic Bladder Exstrophy The incidence of bladder exstrophy has been estimated to be 3.3 cases per 100,000 live births. The male-female ratio is reported to range between 5:1 to 6:1 .

Epispadias Isolated male epispadias has been reported to have an incidence of 1 in 120,000 males. The male-female ratio varies from 3:1 to 5:1. Female epispadias occurs in 1 of every 500,000 female patients.

Cloacal Exstrophy The reported incidence ranges from 1:200,000 to 1:400,000, the sex ratio may be 1:1.

In many cases a prenatal diagnosis is possible, no visible bladder and after appropriate counseling, arrangements can be made for delivery of the newborn in a specialized exstrophy center because this surgery must be considered as a neonatal emergency. The aims of surgical treatment are well established : to recreate 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 success. A 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.

GUIDELINES AND TREATMENT STRATEGIES

MARIO DE GENNARO, ITALY

SPINA BIFIDA

The management of neurogenic bladder dysfunction in children has changed over the years. The introduction of clean intermittent catheterisation (CIC) has revolutionised the management and today the conservative management is a very successful treatment option. The care of children with a neurogenic bladder requires constant observation and adaptation to new problems. Standard treatment is CIC with anticholinergics : children do not have upper tract deterioration when managed early with IC and anticholinergic medication. CIC should be started soon after birth in all babies, especially in those with signs of possible outlet obstruction Furthermore the early initiation of CIC in the newborn period makes it easier for parents and for children to accept it.

Early management results in reduced renal and bladder damage and in neurogenic bladders that are refractory to anticholinergics, injection of botulinum toxin into the detrusor seems to be effective to avoid or postpone during childhood major surgery. Sacral neuromodulation seems to be effective too in selected cases . Some patients require bladder augmentation., Autoaugmentation is a valid alternative associated to sling procedure , in order to increase bladder capacity..For Children with underactive sphincters, no medical treatment available has been validated to increase bladder outlet resistance and additional bladder outlet procedures is required when both the bladder and outlet are deficient. Bladder outlet procedures include bulking agents endoscopic injections and bladder neck reconstruction or other forms of urethral

reconstruction Continent stoma, as Mitrofanoff or Monti are utilised primarily after failure of previous bladder outlet procedure or when native urethra is not accessible. Children with neurogenic bladder have disturbances of bowel function as well as urinary function.

Posterior urethral valves

PUV are one of the few life-threatening congenital anomalies of the urinary tract found during the neonatal period. Antenatal treatment of PUV remains controversial. Open fetal surgery as reported by Harrison is experimental. The most common modality of antenatal treatment is very severe form at high risk for the fetus is the creation of a vesico-amniotic shunt. Amnioinfusion has also been suggested in case of PUV with oligohydramnios. Bladder drainage is mandatory after the birth; In a newborn with suspicion of PUV the first act is then to provide bladder drainage. This can be achieved by a 6 or 8 Fr. feeding tube or Tieman or Foley catheter inserted in the urethra. As an alternative, a suprapubic 5 Fr. epicycstostomy can be used. Today endoscopic valve ablation is the standard treatment with an endoscopic incision or resection with Valve ablation, and it is important in this maneuver to avoid extensive electrocoagulation, because the most common complication of this procedure is stricture formation. Cystoscopy is performed with a neonatal cystoscope, Valves ablation may be performed utilizing a 3 Fr. Bugbee electrode connected to a cut only monopolar electric cautery. A neonatal resectoscope may be utilized as well for valves ablation, some pediatric urologists prefer to incise the valves by a "cold knife", at 5 and 7 and 12 o'clock positions. It should be avoided to remove all the valves folds, while in extremely small premature babies, a small Fogarty catheter can be used. A "second look" is generally performed 2-4 weeks later and a VCUG is repeated after 3-4 months. Vesicostomy is used when the child is too small or in severe general condition. Otherwise, a cutaneous vesicostomy provides an improvement or stabilisation of upper urinary tracts although there has been concern that a vesicostomy could decrease bladder compliance or capacity. High diversion should be considered if bladder drainage is insufficient to drain the upper urinary tract. Life-long monitoring of these patients is mandatory, as bladder dysfunction is very common and continence is a major problem. In the newborn with PUV, chronic obstruction typically produces a small capacity, scarcely compliant bladder, often associated with upper tract dilatation. After successful valve ablation, progressive reduction of upper dilatation and improvement in bladder capacity and compliance are expected. However, not all bladders become "normal". Up to 75% of boys with PUV show abnormal bladder dynamic. Urodynamic studies allowed to identify four different patterns of bladder dysfunction changing during time in boys with PUV: bladder instability (overactivity); low compliance; high voiding pressure and myogenic failure. For the risk of high tract deterioration in the presence of high bladder pressures some authors recommend early aggressive anticholinergic therapy. High voiding pressures after valve ablation can be secondary to residual valve leaflets, urethral stenosis or bladder neck obstruction. Bladder neck is perceived as an important cause of bladder outlet obstruction in PUV patients. This led several authors to perform minimal bladder neck endoscopic incision; that are debated while encouraging results have been reported with the use of alpha-blockers. The use of botulinum toxin was suggested by some author as an alternative treatment. Overactive bladder will be treated with antimuscarinics. Oxybutinine has been widely used so far, but newer drugs such as Tolterodine have the advantage of fewer side effects. An hypocompliant bladder could improve with anticholinergis, but in case of failure a bladder augmentation could be required. High voiding pressure are now successfully faced with alpha blockers, while more complex is the approach to myogenic failure which may require scheduled double voiding, alpha blockers, nocturnal bladder drainage and intermittent clean catheterization. The indication to bladder

augmentation have progressively decreased in the past years in boys with PUV. First, the improved knowledge in valve bladder pathophysiology has shown that PUV bladder is in continuous evolution with spontaneous progressive increase in capacity and compliance over the years. Secondly, the greater attention paid to bladder rehabilitation and urotherapy and the more “aggressive” conservative have progressively reduced the indications to bladder augmentation in PUV patients. The incidence of end stage renal disease (ESRD) in patients affected by severe PUV is significant. In one third of cases ESRD develops in the newborn age, mostly due to renal dysplasia. The other two thirds reach ESRD in the late puberty or adulthood. In these latter patients renal function was satisfactory at birth; during the years a combination of urinary infections, bladder dysfunction, hyperfiltration and increased metabolic demand at puberty may lead to decompensation. A significant reduction in renal graft survival in PUV patients was still reported.

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, or complete repair.

1. The initial staged approach to functional bladder closure includes bladder, abdominal wall, and urethral closure in the newborn period with bilateral osteotomy, epispadias repair at 6 months to one year of age; and bladder neck reconstruction along with antireflux procedure later at 4 to 5 years. Different types of pelvic osteotomy have been suggested and osteotomy, is still debated for the different ages. Osteotomy anyway gives some advantages: reduces abdominal wall tension, placing the urethra in a deeper plane, reinforcing outlet resistance and supporting bladder neck.

2. Complete Repair Developed by Mitchell, this approach combines standard bladder closure with the “penile disassembly” technique for epispadias repair at the same time with the aim to reduce the number of procedures required for reconstruction and potentially improving continence without the need for formal bladder neck reconstruction. The urethral plate is dissected from the corporal bodies and the urethra bladder plate are moved posterior into the pelvis. This procedure leaves an high percentage of patient 60% with hypospadias. The long term results of the 2 techniques are difficult to compare because is very common that every single centre chooses one of these according to the training of the single urologist. Aim of surgical management of epispadias: is providing a satisfactory cosmetic appearance, as well as normal genital function and preservation of fertility, and functioning urethra. The critical point to obtain a cosmetic and functional penis are: dorsal chordee correction and penis mobilization and elongation, internal rotation, Cantwell-Ransley, of the cavernous bodies, Glanduloplasty. epispadias repair objectives include achievement of urinary continence with preservation the upper urinary tracts and the reconstruction of cosmetically acceptable genitalia. The surgical management of incontinence in penopubic epispadias is virtually identical to that in closed bladder exstrophy. In patients with complete epispadias and good bladder capacity, epispadias and bladder neck reconstruction (Young-Dees-Leadbetter bladder neck reconstruction) can be performed in a single-stage operation. Urethroplasty need to be performed after bladder neck reconstruction.

PHARMACOTHERAPY AND REHABILITATION

LAETITIA DE KORT, Netherlands

Pharmacotherapy

Bladder dysfunction in children with spina bifida, bladder exstrophy or posterior urethral valves may require pharmacotherapeutic intervention. Either detrusor overactivity, outflow obstruction or recurrent infection may be an issue in these patients.

Bladder pressure

Concerning bladder pressure, the aim is to keep pressure low to prevent both deterioration of the upper tract and incontinence. First line treatment of a bladder with high filling pressure is the administration of antimuscarinic agents. Dose should be adjusted to bodyweight; to maximize patient compliance once-daily medication is preferable to twice-daily administration. Although oxybutynin and propiverine are the only antimuscarinics that have been registered for paediatric use, all the newer medications shown to be efficacious in adults (such as tolterodine, darifenacin, solifenacin and fesoterodin) may be used in older adolescents who have achieved near-adult size. Higher doses of antimuscarinics appear to be needed in neurogenic patients with typical side effects, including dry mouth, constipation, headache, drowsiness, flushing, blurred vision, personality changes and compromised learning, being reported. Currently, there is insufficient proof of that one antimuscarinic drug is more beneficial than another.

Botulinum toxin A (BoNT/A) injections into the detrusor are a relatively new treatment option for neurogenic bladder overactivity. One systematic review analyzed 6 clinical studies totalling 108 patients, 93% of whom had meningomyelocele. After 30 injections of 10 U/ml (maximal dose of 300 U), under cystoscopic guidance and general anaesthesia 4 out of 5 studies reported decreased bladder pressure and significantly increased detrusor compliance. Incontinence improved in 65-87% of cases. The mean duration of effect varied from 4-42 weeks with repeat BoNT/A injections required after at least 6 months.

Replacing oral antimuscarinics with a BoNT/A injection could help adolescents who have trouble with a medication regimen. Whilst no serious adverse events have been associated with BoNT/A injections the appearance of antibodies in children and adolescents have been reported.

Outflow obstruction

For the treatment of bladder emptying problems in the management of neurogenic bladder and chronic lower urinary tract symptomatology dysfunction alpha antagonist working alone or in conjunction with anticholinergic medications will improve clinical symptoms, increase bladder compliance, and decrease detrusor overactivity.

Urinary tract infection

Patients with neurogenic bladder dysfunction, especially those with a poorly compliant bladder and vesicoureteral reflux, are at risk for recurrent UTIs. These may be symptomatic with abdominal pain, fever, or foul-smelling urine, or be asymptomatic and give rise to bladder stone formation, upper urinary tract scarring or bladder carcinoma. Currently there is no proof of that continuous chemoprophylaxis is beneficial in preventing, diagnosing and treating UTIs in children with neurogenic bladder dysfunction. If one decides to give prophylaxis, 50-100 mg once daily of nitrofurantoin or trimethoprim is recommended.

Rehabilitation

Adolescents on the one hand need to gain independence from their care givers and on the other hand to learn to cope with their constraints. Special programs exist for adolescents. The aim of rehabilitation is to learn or to retain proper bladder emptying, either spontaneously or by CIC, to strive for proper bowel emptying and to maintain adequate fluid intake.

CONSERVATIVE MANAGEMENT VERSUS SURGICAL MANAGEMENT : CLINICAL CONSIDERATION, ADVANTAGES AND DISADVANTAGES

WENDY BOWER Denmark

Lower urinary tract malformations, along with kidney anomalies, are the most common cause of end-stage renal disease in children and young adults requiring renal replacement therapy (Woolf 2013). Although routine surveillance has meant that more patients are surviving into adulthood, kidney and bladder function remain challenged by high grade reflux, a high pressure reservoir, incomplete emptying and urinary tract infections (de Kort 2012; de Lair 2007). The separate classification of over or underactivity in both the detrusor and the urinary sphincter determines the pattern of bladder dysfunction and directs tailored management (Tekgul 2009).

Conservative management of either or both the bladder and sphincter complex involve pharmacologic management, intermittent catheterization, bowel management and neuromodulation (Carr 2010). The objectives of these non-surgical strategies are to preserve renal tract function primarily, thence optimise quality of life and promote independence of self-care (de Jong 2008). Achievement of continence is of secondary importance to preservation of renal function. In the adolescent population discussion of sexuality, potential fertility problems and treatment to optimise sexual function is also important (de Kort 2012).

Conservative management to address Detrusor overactivity or decreased compliance during storage:

- Antimuscarinics (oral, slow-release, intravesical, transdermal) to increase bladder capacity and suppress detrusor overactivity in combination with clean intermittent catheterization (CIC) from newborn age (if spinal cord lesion) or when signs of outlet obstruction are noted (Lehnert 2012)
- Regular evaluation of adherence to medication and CIC
- Upper tract ultrasound surveillance whilst pdet <30cmH₂O and capacity near normal
- Periodic measurement of bladder wall thickness
- Early evaluation of pelvic floor activity to identify any sphincter dyssynergia
- Transcutaneous sacral or percutaneous tibial neuromodulation
- Repeated Botox A injections of bladder

Conservative management to address Detrusor underactivity during emptying:

- CIC with largest possible catheter: includes instruction and review of adequate hand hygiene, perineal hygiene, catheter cleaning, insertion of catheter without contamination, optimal interval between CIC (Hill 2013). Any mental impairment or physical difficulty limiting self-care should be considered.
- Overnight catheter drainage
- Intravesical neuromodulation
- Treatment and prevention of urinary tract infections: bacteriuria is usual in patients performing intermittent catheterization but may be ↓ by improved hydration and more frequent catheterizations (Hill 2013). Physical symptoms of infection accompanying bacteriuria >10⁵ from a catheter specimen, warrants treatment with antibiotics. There is no evidence that catheter technique, type or strategy change the incidence of UTI (Moore 2007).

Conservative management to address Sphincter overactivity during emptying:

- Early urodynamic evaluation to classify sphincter and pelvic floor activity
- Antimuscarinics and CIC
- Surveillance during puberty as bladder capacity, maximum detrusor pressure and leak point pressure may all ↑ after puberty

Conservative management to address Sphincter underactivity during storage:

- Bladder neck laxity protects the upper tracts but impairs continence → need for appropriate urine containment products along with toileting and fluid management (Hill 2013; de Jong 2008)

Conservative management to address Bowel Management:

- Surveillance to identify faecal impaction and faecal incontinence: bowel diary, rectal diameter on ultrasound, constipation score
- Chronic laxative use: stool softeners and oral stimulant laxatives (de Kort 2012)
- Program of retrograde colonic enemas

- Transcutaneous thoracic interferential electrostimulation (Clarke 2009)

Indications for surgical therapy in place of a trial of conservative management

- Combination of high bladder pressure and reflux at a young age
- Previous spinal fusion or limitations in arm function that preclude self CIC
- Physical weight of child makes wheelchair transfers difficult, necessitating a catheterizable stoma
- Stoma to preserve patient privacy; young people and where caregiver is not a parent

MINI-INVASIVE TREATMENT: ROLE OF BOTULINUM TOXIN, SACRAL NEUROMODULATION, ENDOSCOPIC PROCEDURES

GIOVANNI MOSIELLO, Italy

The management strategies of incontinence in these children will be not defined by urodynamics studies and urinary tract imaging only, and in our experience a multidisciplinary approach is mandatory, with a team involving at minimum an urologist, urotherapist, psychologist . Lower urinary tract dysfunction have to be related to coexisting pathologies too, as well as quality of life. The children's physical limitations, patient's emotional status, as families social needs may play a critical role determining a different therapeutic strategy in the same clinical situation.. In any case before any surgical reconstruction all available medical management options as well as all endoscopic or minimally invasive procedures should be exhausted. Today the mini-invasive treatment approach is the gold standard in neurogenic bladder in children and the same approach has to be considered in the other congenital pathologies as Bladder exstrophy or posterior urethral valves.

Today is well established that urological and nephrological treatment of neurogenic bladder dysfunction (NBD) should start as soon as possible to prevent renal damage For this reason in pediatric urology practice newborns with spina bifida start clean intermittent catheterization (CIC) from birth with anticholinergics .When NBD is associated to untreatable severe vesico-ureteral reflux (VUR),surgery is often required.In the past yearswe suggested a combined endoscopic injections of botulinum toxin type A (BoNTA) and dextranomer/hyaluronic acid (DxHA) as an effective minimally invasive treatment of VUR and NBD not responding to CIC and anticholinergics . Anyway these patients have to perform CIC. and when patients, or their caregivers, are unable to catheterize the native urethra surgical alternatives have to be considered: urinary continent derivation according to Mitrofanoff's principle, vesicostomy or ileal conduit, especially when cerebral function is bad. In the past years it was demonstrated as button cistostomy is an effective

form of bladder drainage , and recently good results were reported with button cistostomy endoscopic insertion. We decide to include this procedure in the treatment of VUR and NBD using BoNTA and DxHA in 12 children, unable to perform CIC, in a extended combined endoscopic approach. The surgical procedure was performed in general anaesthesia: first of all endoscopic suburethral injection of DxHA (0.5-1 ml/side) and bladder BoNTA injections (10 IU/Kg, max 300 IU) were performed and then as cistostomy a Mic-Key gastrostomy, button (Kimberly-Clark/Ballard,USA) was inserted endoscopically according to original Subramaniam's technique. The mean overall operative time was 40 minutes, no surgical complications were reported., At the mean follow-up of 11,8 months urinary tract infections was solved in all, as well as VUR , but in one renal unit.. All the buttons are in situ, well working. No peristomal urine leakage , wound infection, erosion granuloma, were observed. Buttons were changed every 3 months and all relatives and patients are satisfied about urinary drainage , opening the button and connecting it the feeding tube.for intermittent bladder emptying that is regularly performed in all 4-5 times daily, as for CIC, with 1 patient performing nocturnal continuous emptying. BoNTA endoscopic injection was repeated after 9-12 months to maintain a low pressure bladder and a good continence status This cumulative endoscopic treatment seems to be a valid alternative to derivation, augmentation and reimplantation in the short and medium term period, helping child maturation and family education and a correct definitive management choice , improving children status ,avoiding psychological pressure on relatives in patients with associated severe mental disability, where major surgical reconstruction could be considered an overtreatment.

This treatment could be very useful to postpone major surgery in adult life

Another very promising treatment option is Sacral neuromodulation (SNM), that was used extensively in adults.It has been suggested that sacral root stimulation facilitates bladder recovery, we have experienced sacral neuromodulation (SNM) in children with incomplete injury and SNM seems to be a promising therapeutic modality for NBD in selected ones. SNM has been suggested in children previously operated for bladder exstrophy A total of 14 patients with NBD with a mean age of 16.1 yr (range 10-21) were implanted between 2008-2012. There were 10 boys and 4 girls of which 6 had a congenital disorder and 8 had an acquired NBD.

The implant was in all time placed in the side which was identified being the best during preliminary testing. No post-operative complications were observed.

The average time of the intervention was 53 minutes for the first operation, and 40 minutes for the second one. All implants are in place and functioned well at last control. The clinical and urodynamic results are shown in table 1.

The evaluation is done by dividing patients in two groups: responders and non responders. Patients were considered responders if: they were satisfied and had one or more of the following criteria: <50% incontinence episodes, <50% post voiding residual, <50% of necessity for CIC, >50% voided volume.

In order to get a better understanding of the optimal indications for SNM, we systematically searched the PubMed, EMBASE and Cochrane databases Language restrictions were applied to other languages than English. No date restrictions were applied. The last search was performed on ... 2012. We hand-searched the reference list of all included studies and any relevant review articles.

Abstracts of all identified studies were independently reviewed by two authors and any study reporting on SNM for the treatment of neurogenic bladder in pediatrics was reviewed in full text. (Adult studies were reviewed if they reported on children as well if they were well described). We aimed to include any original SNM study that reported efficacy and/or safety data on tested

and/or permanently implanted patients suffering from neurogenic bladder. Non original articles, studies not published as full-text articles, and those including adults were excluded.

The variables assessed included year of publication, level of evidence, study type, number of patients, gender, age, (duration of the underlying neurologic disease), success rate (total, according to the neurologic disease/injury causing neurogenic bladder, according to the type of LUTD) of test phase and permanent SNM, duration of test phase, length of follow-up, adverse event (total, type of adverse event), and surgical intervention (total, type of surgical intervention) of test phase and permanent SNM. (In accordance with the literature (see article), an improvement of >50% in bladder diary variables (number of leakages, pad use, number of voids, number of catheterisations) was considered a success.

We compared the results of our own study group with the results of studies found in literature.

We evaluated the results using a subgroup analysis comparing acquired with congenital NBD. The results of our own series showed an overall success rate of 71.4%, versus 70.4% in the literature studies. Considering different pathologies, incomplete spinal cord injury (SCI) presented the highest success rate; 100% in the literature and 75% in our own experience. The lowest rate was reported in complete SCI and myelomeningocele (0%).

A total of 71 children with NBD treated with SNM with a mean age of 14,3 years (range 5-21) were included: 14 children (mean age 16.1 yr, range 10-21) of our own experience and 57 patients (mean 13.9 yr, range 5-21) from 11 studies identified by an electronic search on PubMed (6 pediatric studies and 5 adult studies). The clinical response was evaluated on the basis of patient satisfaction and one or more of the following criteria: <50% incontinence episodes, <50% post voiding residual, <50% of necessity for CIC, >50% voided volume, defining responders from non-responders. All clinical data of 71 patients were independently extracted and evaluated by 2 reviewers, both not directly involved in the surgical procedure.

Our study together with the literature found a favorable outcome for acquired versus congenital NBD. Subgroup analysis shows that the predictive value of the positive response of SNM depends on the specific indications. Well-designed, pathology specific, multi-centric studies could be useful as well as investigations on PUV and BEEC.

Anyway the most important urological advancement in NBD treatment in these years remains the use of onabotulinum toxin A that has revolutionized the treatment of NBD . In the recent years onabotulinum toxin A has changed dramatically the treatment of NBD in adults and pediatrics in many centers the indications to bladder augmentation were dramatically reduced, and children have been treated with success and during time, avoiding bladder augmentation. In our experience onabotulinum toxin A confirmed its efficacy and safety as well as in the experiences of other colleagues We have treated 91 patients with NBD, aged 2-18 years old, with 281 procedures (1-9 treatments). No severe complications were observed maybe due to our pre-intra and postoperative protocol treatment and only in 3 cases we observed persistent hematuria 24 hours longer. Botulinum toxina A safety was confirmed regarding the effects on bladder wall after repeated injections While this treatment seems to be more effective in bladders with detrusor muscle overactivity, scant data are reported on low-compliant high pressure bladders The most commonly used dose of botulinum toxin is 10 U/kg with a maximum dose actually still of 300 units, that of course will be reduced as in adults to 200 UI. Of course it is unclear how many times this treatment can be repeated, although repetitive treatment has been found to be safe in children and in adults Histological studies have not found ultrastructural changes after injection

In the guidelines of European Urological Association for Neurogenic Lower Urinary Tract Dysfunction the role of onabotulinumtoxin A is presented in a different way respect to ICCS document. Onabotulinum toxin could be a treatment alternative in other pathologies as PUV.. Regarding bladder outlet insufficiency treatment, this may be well managed by bulking agents. Of course this a temporary treatment with a short term efficacy useful in pediatric population in order to postpone major surgical procedure. For the same reason children with bowel dysfunction are well managed using advanced trans anal irrigation, and Malone procedure indications were reduced during time.

SURGERY: AUGMENTATION, DERIVATION, SLING

LAETITIA DE KORT, Netherlands

Bladder augmentation

When medication has failed to lower elevated 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. Ileocystoplasty is more commonly performed, but carries the risk of postoperative intestinal obstruction, mucus retention, increased rate of stone formation, and electrolyte imbalance.

The risk of secondary malignancy of the augmented bladder is increased, although less than 20 cases have been described worldwide.

Augmentation may be combined with ureteral reimplantation, bladder neck tightening (sling suspension, bladder neck reconstruction, artificial sphincter implantation) or the creation of a continent catheterizable urinary stoma (Mitrofanoff, Monti). As bladder augmentation lowers bladder pressure, diminishing or abolishing vesicoureteral reflux, ureteral reimplantation should only be performed in cases where high grade reflux occurs at low bladder pressure. Similarly, as bladder augmentation will improve continence, only patients with low leak point pressure need reinforcement of the bladder outlet. Urodynamic testing will determine surgical options.

Derivation

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

Slings

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%. In boys preservation of erectile function after a fascial sling procedure can be expected. Synthetic suburethral slings can only be used in a tension free mode, due to risk of erosion. In neurogenic stress incontinence a firmer suspension is needed, making synthetic slings inappropriate. Currently, there are no reports describing long-term

results of the synthetic suburethral slings, suggesting its use in a very young population should be tempered. Injection of a bulking agent into the bladder neck area as a primary treatment of bladder outlet incompetence is not recommended because of low success rates.

Following insertion of the artificial urinary sphincter, efficacy rates for complete dryness between voids vary between 56% and 91%. The revision rate is high, about 1/3 require reoperation and 19% device removal due to erosion. Longterm survival (>10 years) of the prosthesis is up to 60%. Approximately half of the individuals able to empty before insertion of the artificial sphincter can do so afterwards, however, bladder dynamics can change postoperatively. Up to 5 years later augmentation cystoplasty may be required in 33% of patients in order to minimize the effect of this change on kidney drainage and function.

Catheterizable channels

Creating an abdominal continent catheterizable stoma into the bladder is a good option when urethral catheterization is impeded. Up to 21% of patients will have problems related to stoma

leakage or stenosis within the first 2 years of its creation and require minor revision. An antegrade continence enema stopper effectively eliminates stomal stenosis. Patients with good manual dexterity and fine motor ability gain a greater ability to self-care and appreciate the privacy gained from not having to expose their genital area to a caregiver. In wheelchair-bound girls dependant on CIC, a catheterizable channel obviates transfers in the bathroom. Stoma creation in male patients only follows difficulty with urethral catheterization. As extreme weight gain can cause adjacent skin to partially obscure a catheterizable channel, postpubertal patients should be educated in nutrition and portion control.

Dryness may also be achieved by closing the bladder neck combined with a catheterizable stoma. Complications after a bladder neck closure have been reported in up to 31% of cases, with 15% developing vesicourethral fistula. Persistent leakage, more UTIs, stone formation, bladder perforation, and deterioration of the upper urinary tract have also been reported after bladder neck closure especially when CIC regularity is neglected. To date, patient compliance with long-term CIC appears to be good and is associated with preservation of the upper urinary tract.

ADULT LIFE : THE NEURO-UROLOGIST POINT OF VIEW

GIULIO DEL POPOLO, Italy

Neurogenic bladder patients require lifelong care of continence , monitoring of renal function . Periodic investigation of upper tract changes, renal function and bladder status is mandatory in spina bifida as well as in PUV and BEEC . Patients operated on with reconstructive procedures using intestine should be regularly followed up for complications such as infection, stones, metabolic changes, and neoplasm, that has been found to occur in 0.6-2.8% of patients during median follow-up of 13-21 years . In a study including 153 patients with a median follow-up time of 28 years , malignancy was found in 4.5%. Who is involved in this lifelong care? The answer is the Neuro-Urologist, that is an the urologist with expertise in the diagnosis and management of congenital and acquired pathologies of the urinary tract related to a neurogenic damage . So the clinical interest of Neuro-Urologist will be very close and sometimes overlapped to the Adolescent Urologist to continue the management of continence in older children and young adults previously treated for major congenital anomalies of the genitourinary system. Advances in paediatric medical care and surgical management resulted in improved survival and quality of life for children with complex congenital anomalies of the urogenital tract as BEEC , PUV , Spina Bifida. Every urologist could be considered a NeuroUrologist? The answer is no. NeuroUrologist involved in the life-long care of continence, fertility, sexuality from childhood through adolescence and into adulthood must be competent in reconstructive urology – ureter, bladder, urethra and genital; trained in paediatric and adult urology, competent in urodynamics, and finally to be a part of a specialized team with nephrologist, urotherapist nurse, uro-radiologist, endocrinologist, gynecologist , psychologist, etc.that meets on a regular basis to formulate patient treatment plans. In the past 25 years there has been an enormous development in the paediatric urology. Many

surgical techniques have changed dramatically, and Paediatric urology has totally changed and where in the past the consideration was survival or not of a patient, today care has progressed such that continence is an expectation rather than rarity, with increased survival and better functional outcomes in term of quality of life (QoL) too, with the expectations of our patients , related to social life and sexuality too. If in the past it could be difficult to measure QoL, today we have more instruments as Qualiveen-30 that is a neurological urinary disorder -specific health-related quality of life (HRQL) instrument, recommended in the European Association of Urology guideline by 2008.

Sexuality and spinal cord injury (SCI) :there are very scant data on analysis on the sexual impact of medical and psychological treatments related to SCI. Literature reports that some co-morbidities are more prevalent in women with SCI compared with able-bodied women but data on sexual functioning are missing. To improve sexual rehabilitation services, sexual issues and response require evaluation during periodical check-ups using validated questionnaires administered by a physician 'guide' who coordinates professional operators thus providing personalized programmable interventions. as we have done investigating a possible correlation between sexual hormonal status and the presence of female sexual dysfunction (FSD) using the Female Sexual Function Index (FSFI) in females with spinal cord injuries (SCI), in selected 39 SCI fertile-aged women. Very interesting appear the results of SNM . Over the last few years, sacral neuromodulation (SNM) has become an established treatment option for lower urinary tract symptoms (LUTS). In our study we have evaluated if SNM improves sexual function in females, measuring the Improvement in sexuality by the Female Sexual Function Index (FSFI) and the Female Sexual Distress Score (FSDS). We included 31 women, 17 of whom were neurogenic with permanent SNM Both questionnaires indicated a clinically significant improvement in sexuality that was maintained up to the final visit for 4 out of 11 neurogenics with sexual dysfunctions: The positive effects regarding sexuality may be due either to enhancement of LUTS or to the direct stimulation of the sacral roots (S3). SNM may be beneficial in men too with selected incomplete SCI . 22 subjects (42.3%) showed erectile impairment (14 were neurogenic). In the first visit post-SNM, five retentionists of neurogenic origin and two with overactive bladder syndrome of idiopathic origin achieved noticeable erectile improvement and their median IIEF-5 score shifted from 14.6 to 22.2, and 15.5 to 22.5, respectively. Future research should test SNM in a larger sample of subjects, exclusively with sexual dysfunctions, in order to better understand the mechanism of action of SNM on erectile function. Another treatment option as Literature suggests is oral PDE5 inhibitors that represent a safe and effective treatment option for ED caused by SCI. Further research is needed on head-to-head comparative trials and SCI patient preference for these drugs; their impact on ejaculation and orgasm function, their early use after SCI for increasing the recovery rate of a spontaneous erection, and their effectiveness and tolerability in the long-term are still to be investigated. Prostate volume and PSA levels are lower in SCI patients and are inversely related to the patient age at lesion onset. Whether this effect is mediated directly or indirectly by a impaired nerve supply to the prostate remains to be determined. Despite the present observation of reduced prostate disease, as during the last twenty years life expectancy in SCI patients has improved significantly, the need to screen these patients for the occurrence of prostate disease should not be disregarded. Neurogenic population is most investigated respect to BEEC and PUV because spinal cord lesion effects on sexual life were studied extensively and study on their concerns on ejaculation, orgasm, erectile dysfunction, etc are very useful for adult with BEEC and PUV treatment.

In BEEC male fertility was documented in two large exstrophy series, in which 3 of 68 and 4 of 72 patients had successfully fathered children. In another very large series of 2500 exstrophy and epispadias patients, there were 38 males who had fathered children.

Stein reported the conclusion that male patients with genital reconstruction and closure of the urethra demonstrated a high risk of infertility. Anyway sexual function and libidus in exstrophy patients are normal. The erectile mechanism in patients who have undergone epispadias repair appears to be intact, because 87% of boys and young men in the Hopkins series experienced erections after repair of epispadias. However ejaculation may be poor or absent ejaculation after genital reconstruction. Complete absence of ejaculation is rare, but the emission that does occur may be slow and may last for several hours; Most patients reported satisfactory orgasm; half of the men described intimate relationships as serious and long term.

ADULT LIFE : THE URO-GYNECOLOGIST POINT OF VIEW

LATTHE PALLAVI, United Kingdom

Bladder Exstrophy

- The word *Exstrophy* is derived from the Greek word *ekstriphein*, which literally means to "turn inside out." Bladder exstrophy is a malformation of the bladder, in which the bladder and related structures are turned inside out. The skin of the lower abdominal wall that normally covers the bladder also does not form properly and is separated, thus exposing the inside of the bladder to the external world. If you imagine a balloon that has been split and opened up so that the inside of the balloon is visible, you will have a picture of what has happened.
- **What other abnormalities are associated with bladder exstrophy?**
A baby with this condition will usually also have all, or some, of these other associated abnormalities:
 - **Epispadias:** the urethral opening is located between a split clitoris and labia minora
 - **Absence of a bladder neck and sphincter**
 - **Small bladder capacity**
 - **Abnormally positioned ureters causing reflux**
 - **Separation (diastasis) of the pubic bones**
 - **An anus positioned further forward than usual:** This positioning is typically not accompanied by problems with bowel function.
 - **A low positioned belly button (umbilicus):** The umbilicus is not usually visible and is removed when the bladder is surgically closed.
- **At what stage of prenatal development does bladder exstrophy occur?**
The condition of exstrophy occurs during the development of the embryo very early in the pregnancy, about 4 to 5 weeks after conception. The cause and nature of the faulty development is not exactly certain. One theory suggests that something goes wrong during this early folding and separation, causing the cloacal membrane to fail to close, leaving the bladder outside of the abdominal wall. A second theory proposes that the layer of skin which forms over the bladder at this stage is thin and unable to hold in the bladder. It pulls apart, again leaving the bladder inside out.
- **What determines the extent of the exstrophy?**
The extent of the exstrophy depends on how small or how large the opening is. One can think of exstrophy as a spectrum of conditions that ranges from the milder form,

epispadias (only a urethral defect or opening), to the most severe form, cloacal exstrophy (defect of the urethra, bladder and bowel), with classic bladder exstrophy (defect of urethra and bladder) being in the middle.

- **What is the likelihood that a child will be born with bladder exstrophy?**
Although bladder exstrophy occurs in only 1:30,000 to 40,000 births in the general population, it may be more likely to recur in a family but the overall risk is still <1% to 1 in 250 births (Campbell urology, 2008). Exstrophy of the bladder also occurs 2-3 times more frequently in males than in females.

- **What are the fertility issues?**

- Almost all boys with exstrophy produce healthy sperm. However, some may have difficulty fathering children through traditional sexual intercourse. The potential difficulty is in the delivery of the sperm to the egg. Difficulties may be a result of backward flow of semen during ejaculation (also known as retrograde ejaculation), the inability to ejaculate at all or low semen volume. If necessary, men may be able to use Assisted Reproductive Techniques (ART) such as intrauterine insemination and *in vitro* fertilization.
- If girls with bladder exstrophy have a problem with sexual function and infertility, it's most often a result of an anatomical concern. In some, the vaginal opening may be too narrow and may require surgical enlargement. When necessary, this procedure should allow for normal sexual intercourse and achieving pregnancy.
- Some women with bladder exstrophy may develop uterine prolapse, in which anatomical support for the uterus is lacking and the uterus may protrude into the vagina during the later stages of pregnancy. If this happens, a woman may need surgery, but she can still have babies by vaginal delivery or Caesarean section (C-section), although the latter is often recommended so that her urinary continence isn't affected during vaginal delivery.

Spina Bifida

Prevalence and incidence

Spina Bifida is a neural tube defect that affects approximately 3,000 pregnancies each year. Although the occurrences appear to be decreasing, Spina Bifida occurs in approximately seven of out every 10,000 live births in the United States. According to the Spina Bifida Association of America, it is estimated that more than 70,000 people in the United States are living with this birth defect.

What is effect of SB on sexual function?

The two are inseparable because the same nerves that control urinary function also control sexual function. Therefore, if one has poor sensation, the other will also. It is very important to have an understanding partner and to communicate clearly about the possibility of urine or stool leakages during sex. The other issue might be low self esteem

Fertility

Pregnancy is possible for almost all women with Spina Bifida so appropriate contraception is strongly recommended if pregnancy is not currently desired.

Indications and cautions similar to general population

Pre-pregnancy

Pre-pregnancy counselling ideal to optimise health prior to conception is recommended

Spina bifida is best prevented by taking 400 micrograms (mcg) of folic acid every day. Studies have shown that if all women who could become pregnant were to take a multivitamin with the B-vitamin folic acid, the risk of neural tube defects could be reduced by up to 70%.

Women who have experienced a pregnancy affected by a NTD like Spina Bifida need a higher intake of folic acid i.e 4 milligrams (mg) and is available by prescription from a health care provider.

Antenatal

Pregnant women with Spina Bifida should also see their urologist and neurosurgeon in addition to their gynecology team to make sure the pregnancy is not adversely affecting their shunt or kidneys.

Recurrent UTIs- Risk of pyelonephritis, SGA

Chronic hypertension if renal scarring- superimposed pre-eclampsia

Catheterisation in third trimester if performing CISC

Type of delivery – vaginal or C-section?

The type of delivery depends on multiple physical factors, including sensation level, ability to push with pelvic muscles, size of the pelvis, and flexibility around the hips and knees. The more conducive these factors are to the birthing process, the more likely a vaginal delivery can be performed.

If it appears the baby will not be able to be pushed safely through the pelvic area and down the birth canal, a Cesarean section will be recommended. If a C-section is chosen, the urinary tract is evaluated for re-implantations, diversions, or conduits to make sure they are avoided during the delivery incision.

Women with SB have successfully and safely received epidural anesthesia for their delivery. Because the anatomy of the spine is different, the epidural may need to be placed using ultrasound guidance. There are reports of successful epidurals for most myelomeningocele lesion levels, with or without scoliosis.

As with all pregnancy issues in Spina Bifida, monitoring the effects of the epidural is trickier than in usual pregnancies. Therefore, it is best to deliver with an obstetrician and hospital experienced in high-risk pregnancies.

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