

REVIEW ARTICLE

EAU/ESPU guidelines on the management of neurogenic bladder in children and adolescent part II operative management

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Abstract

Background: Treatment in children and adolescents with a neurogenic bladder is primarily conservative with the goal of preserving the upper urinary tract combined with a good reservoir function of the bladder. However, sometimes—even in childhood—conservative management does not prevent the development of a low-compliant bladder or overactive detrusor.

Material & Methods: After a systematic literature review covering the period 2000-2017, the ESPU/EUAU guideline for neurogenic bladder underwent an update.

Results: In these patients, surgical interventions such as botulinum toxin A injections into the detrusor muscle, bladder augmentation, and even urinary diversion may become necessary to preserve the function of the upper (and lower) urinary tracts. The creation of a continent catheterizable channel should be offered to patients with difficulties performing transurethral clean intermittent catheterization. However, a revision rate of up to 50% needs to be considered. With increasing age continence of urine and stool becomes progressively more important. In patients with persistent weak bladder outlets, complete continence can be achieved only by surgical interventions creating a higher resistance/obstruction at the level of the bladder outlet with a success rate of up to 80%. In some patients, bladder neck closure and the creation of a continent catheterizable stoma is an option.

Conclusion: In all these patients close follow-up is mandatory to detect surgical complications and metabolic consequences early.

KEYWORDS

bladder augmentation, bladder neck reconstruction, EAU guideline, Mitrofanoff stoma, neurogenic bladder, sling procedure, urinary diversion

1 | INTRODUCTION

Within the European Association of Urology (EAU)/European Society for Pediatric Urology (ESPU) guidelines for the treatment of neurogenic bladder in children and adolescents, proactive management with clean intermittent catheterization (CIC) +/- anticholinergic medication starting in the first months of life is recommended. Starting treatment only once changes or complications have occurred, may have long-term implications for patients and their optimal management. Despite the fact that almost 12% of neonates with myelodysplasia show no signs of neurourological dysfunction at birth,¹ they have a high risk of developing this later on in childhood. In a recent series, around 15% of patients have impaired renal function at the age of 29 years.² In a recent systematic review concerning the outcome of adult patients with meningomyelocele this finding is confirmed, as 25% have some degree of renal damage and 1.3% have end stage renal failure.³

The main goals of treatment concerning the urinary tract are preservation/improvement of renal function and prevention of urinary tract infections (UTIs). During childhood, continence of urine and stool becomes progressively more important. Therefore, maintenance or development of continence is the then the third goal.

Herein, we will present the EAU/ESPU guidelines on the operative management of neurogenic bladder in children and adolescents.

2 | PROCEDURES TO INCREASE BLADDER CAPACITY

2.1 | Botulinum toxin A

In patients with a neurogenic bladder refractory to anticholinergics/antimuscarinics, botulinum toxin A injections into the detrusor muscle should be considered as the next step to restore a low pressure, high capacity reservoir.^{4,5} A recent systematic review in children with a neurogenic bladder, demonstrated that continence can be achieved in 32% to 100%, the maximum detrusor pressure is decreased by 32% to 54%, maximum cystometric capacity is increased by 27% to 162% and bladder compliance was improved (28-176%).⁴ Onabotulinum toxin A (maximum 300 IU injected at 20-30 different sites) seems to be more effective in bladders with detrusor muscle overactivity, whereas noncompliant bladders without detrusor overactivity are unlikely to respond.^{6,7} It has been shown that botulinum toxin A injections into the trigone seem to be safe with regard to reflux and upper urinary tract changes, but if it is advantageous compared with the conventional technique has not been investigated.⁸

Abobotulinumtoxin-A may also be used, but there are only a few studies performed so far in children with neurogenic bladder.⁹

2.2 | Bladder augmentation

Bladder augmentation should be offered in cases where anticholinergic drugs and botulinum toxin A injections fail to maintain a low-pressure reservoir with a good capacity and bladder compliance. For augmentation, ileal and colonic segments can be used.¹⁰ Each bowel segment has its advantages and disadvantages, for example, ileum is more compliant than the colon, colon is directly located beside the bladder, ileum is associated with loss of vitamin B12 and bile acid (diarrhea), colon produces more mucus increasing the risk of stone formations.¹¹ In those who are not able to perform transurethral CIC, a continent catheterizable cutaneous channel can be offered. In this group of patients, the rate of surgical complications and revision is high. The 30-day all over event rate in one large review in patients with bladder augmentation +/- a continent catheterizable cutaneous channel was approximately 30% (23-33%) with a reoperation rate in this short time period of 13%.^{12,13} In these patients with a long-life expectancy, the complication rate clearly increases with the follow-up period.¹²⁻¹⁵ Bladder perforation, as one of the worst complications, occurred in 6% to 13%.¹⁵ Continent cutaneous catheterizable channels have been shown to have a relatively high revision rate due to stenosis or incontinence of the stoma (up to 50-60%), depending on the type of channel.^{16,17}

In addition to surgical complications the metabolic consequences of incorporating bowel segments into the urinary tract have to be taken into account; for example, disturbances of the acid-base balance, vitamin B₁₂ deficiency and loss of bone density. Stool frequency may increase and diarrhea can occur after the exclusion of bowel segments.¹¹ Last but not least, these patients have a lifelong increased risk of developing secondary malignancies.¹⁸⁻²⁰ Therefore, a lifelong follow-up of these patients is required, including physical examination, ultrasound, blood gas analysis, (pH and base excess), renal function and vitamin B₁₂ levels if ileum was used. Cystoscopy may be performed, although it is not clear at what frequency it can be recommended.

Adverse effects of intestinal cystoplasties can be avoided by the use of ureterocystoplasty. The combination of a small contracted bladder, associated with severe dilation of the ureter of a nonfunctioning kidney is quite rare. The technique was first described in 1973 by Eckstein²¹; the success rate depends on patient selection and the reaugmentation rate may be as high as 73%.^{22,23}

Autoaugmentation with partial detrusorctomy or detrusor myotomy creating a diverticulum avoids metabolic

complications as no intestinal segments are required. However, reports are conflicting especially in regard to the success rates; therefore, the patients and their parents should be informed of a potentially low success rate. Detrusorectomy may be used in very selected cases,²⁴⁻²⁷ which have a relatively good preoperative bladder capacity of 75% to 80% of the expected volume.^{28,29} Seromuscular cystoplasty has also not proven to be as successful as enterocystoplasty.³⁰

Tissue engineering, even if successful in vitro and some animal models, does not achieve results comparable to using intestinal segments.^{31,32} Therefore, these alternatives for bladder augmentation should be considered as experimental and should be used only in controlled trials.

3 | CONTINENT AND INCONTINENT CUTANEOUS URINARY DIVERSION

Incontinent urinary diversion should be considered in patients with neurogenic bladder dysfunction and high bladder pressures, who are unable or unwilling to perform CIC. In children and adolescents, the colonic conduit has been shown to have fewer complications compared with the ileal conduit.³³⁻³⁶

The indication for total bladder replacement in children and adolescents with myelodysplasia and neurogenic bladder dysfunction is extremely rare but may be necessary in some adults due to secondary malignancies or complications of previous urinary diversions. Any type of major bladder or bladder outlet reconstruction should be performed in centers with sufficient experience in the surgical technique, and with experienced health care personnel to carry out postoperative follow-up.³⁷⁻³⁹

4 | PROCEDURES TO INCREASE THE BLADDER OUTLET RESISTANCE

A weak or insufficient bladder outlet is a safe situation for the upper urinary tract but may become an increasing social problem due to persistent incontinence. It is a major challenge for the pediatric urologist to increase bladder outlet resistance. So far, no available pharmacological treatment has been validated to increase bladder outlet resistance.⁴⁰⁻⁴⁵

Using fascial slings with an autologous fascial strip or artificial material to increase bladder outlet resistance can achieve a continence rate between 40% and 100%. In

males, sling procedures are less successful compared with females. In most cases, this procedure is performed in combination with bladder augmentation.⁴⁶⁻⁴⁸ Catheterizing through a reconstructed bladder neck or a urethra compressed by a sling may not be easy, so many surgeons prefer to combine this approach with a continent catheterizable channel.⁴⁹ In contrast to autologous slings, artificial slings have a high complication rate in girls who perform transurethral CIC.⁵⁰

The use of artificial urinary sphincters was introduced by Scott et al.⁵¹ Continence rates in the literature can be up to 85% in selected patients.⁵²⁻⁵⁵ Theoretically, good candidates would be postpubertal patients, who can void spontaneously, provided that they are manually dexterous to handle the sphincter-pump. In very selected patients CIC through the sphincter in an augmented bladder is possible. However, several steps are required to deactivate the artificial sphincter and empty the bladder mostly by CIC.⁵⁶ Erosion rates can be up to 1 in 5 and revision-rates up to 100% depending on the follow-up time.⁴⁶

Patients who underwent a bladder neck procedure only, have a greater than 30% risk of developing a low compliance bladder requiring an augmentation. Furthermore, 50% of these will develop upper urinary tract changes due to the onset of high bladder pressure.^{57,58} Even in patients with a good bladder capacity and compliance, there is a risk for postoperative changes in bladder function. Therefore, a very close follow-up of patients with neurogenic bladder and surgery at the level of the bladder neck is required to avoid upper urinary tract damage and chronic renal failure.

Bladder neck reconstruction (BNR) in children with the neurogenic bladder is less favorable compared with those in patients with bladder exstrophy.⁵⁹ In most patients, the creation of a continent catheterizable stoma is necessary due to difficulties performing urethral CIC. In one publication only 10% to 33% still able to perform transurethral CIC and the reoperation rate is between 67% and 79% after a median follow-up between 7 and 10 years.⁶⁰ The combination of a sling procedure with a urethral lengthening procedure or BNR may improve the continence rates.⁶¹ Bulking agents have demonstrated a low success rate (10-40%), which is only temporary in most cases.⁶²⁻⁶⁴ On the other hand, bulking agents at the level of the bladder outlet have shown not to adversely affect the outcome of further definitive surgical procedures.⁶⁵ Bladder neck closure with a continent catheterizable stoma is often seen as the last resort to gain urinary continence in those patients with persistent urinary incontinence.^{66,67} A complication rate of up to 33% has been reported, with vesicourethral/vesicovaginal fistula

occurring in up to 15%.⁶⁸ Furthermore, there is a high risk of bladder stone formation, bladder perforation, and deterioration of the upper urinary tract function, if the patient is not compliant with CIC and bladder irrigations.^{68,69}

5 | SURGICAL INTERVENTIONS FOR A VESICoureTERAL REFLUX

In patients with neurogenic bladder dysfunction, it is almost impossible to make a clear distinction between primary and secondary reflux, as the vesicoureteral reflux is mostly secondary to the detrusor-sphincter dysfunction and increases the risk for pyelonephritis. Therefore, treatment is primarily related to bladder dysfunction.^{70,71} On the other hand, patients with high-grade reflux before augmentation have a higher risk for persistent symptomatic reflux following enterocystoplasty.⁷² Simultaneous ureteral reimplantation in high-grade symptomatic reflux especially in patients with low-pressure high-grade reflux should be discussed; especially in patients with a bladder outlet procedure. Endoscopic treatment has a failure rate of up to 75% after a median follow-up of 4.5 years,⁷³ which is significantly higher than the failure rate of open techniques,⁷⁴ and may also have an increased risk of inducing obstruction.

6 | NEUROSTIMULATION/NEUROMODULATION

Intravesical electrical stimulation of the bladder,⁷⁵⁻⁷⁷ sacral nerve stimulation^{78,79} and transcutaneous neuromodulation⁸⁰ are still experimental in children with neurogenic bladder dysfunction and cannot be recommended outside the confines of clinical trials. The same is true for the intradural somatic-to-autonomic nerve anastomosis.^{81,82}

7 | OPERATIVE MANAGEMENT OF FECAL CONSTIPATION

If retrograde transanal irrigation is difficult or impossible due to anatomic or social circumstances antegrade irrigation can be performed by using a Malone Antegrade Continence Enema (MACE)-appendicostomy.^{83,84} In a long-term study of 105 patients, 69% had successful bowel management, however, stomal complications occurred in 63% (infection, leakage, and stenosis), 33% required surgical revision, and 6% eventually required diverting stomas.⁸⁵ In addition, patients need to be informed, that

the antegrade irrigation is also time-consuming with durations of at least 20 to 60 minutes. However, in most patients, bowel cleansing once every 2 days will be sufficient to remain continent for stool in between the irrigations. A cecostomy-button is gaining more and more popularity, especially in the USA as an alternative to an ACE stoma.⁸⁶

8 | SURGICAL PROCEDURES FOR TEMPORARY SOLUTIONS

8.1 | Urethral dilatation

This can lower the pop-off pressure by lowering the detrusor leak point pressure by dilatation of the external sphincter under general anesthesia up to 36 Charrière. Some older studies have shown the procedure to be safe and effective in selected patients, especially females.⁸⁷⁻⁹⁰

8.2 | Incontinent vesicostomy

An incontinent vesicostomy—preferably a Blocksom stoma⁹¹—is an option to reduce bladder pressure in children/newborns, if the parents are noncompliant with CIC and/or CIC through the urethra is extremely difficult or impossible due to anatomical or social circumstances.⁹²⁻⁹⁴ Also, in patients with high-grade reflux and recurrent febrile UTI's a temporary incontinent diversion (eg, refluxing ureterocutaneostomy) can be considered. A cystostomy button may be an alternative, with a complication rate (mostly UTI) of up to 34% within a mean follow-up of 37 months.⁹⁵

8.3 | Sphincteric botulinum toxin A

Onabotulinum toxin A injection into the urethral sphincter has been shown to be effective in decreasing urethral resistance. However, the evidence is still too poor to recommend its use to decrease bladder outlet resistance, but it may be considered as an alternative in some patients, who do not wish to or are incapable of performing CIC.^{96,97}

8.3.1 | Follow-up

Neurogenic bladder patients require lifelong multidisciplinary follow-up, not only for urological aspects but also for neurological and orthopedic aspects.

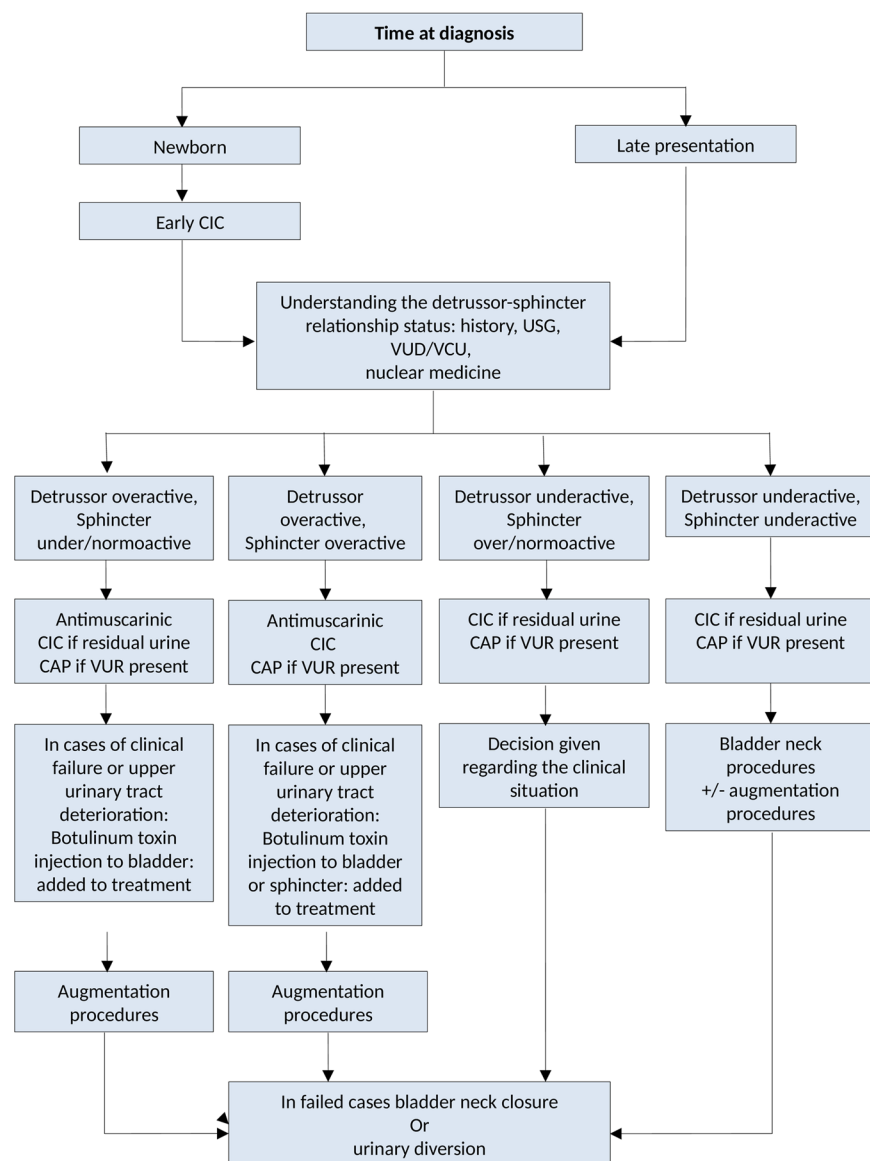


FIGURE 1 Algorithm for the management of children with myelodysplasia with a neurogenic bladder.¹⁰⁷ CAP, continues antibiotic prophylaxis; CIC, clean intermittent catheterization; USG, Ultrasonography; VCU, Voiding cysturethrography; VUD, Videourodynamic; VUR, Vesicoureteral reflux

Regular investigation of the upper and lower urinary tracts is mandatory. Bladder stones, as one of the most frequent complications after augmentation, can be detected by ultrasound as well as during cystoscopy.

In those patients who underwent urinary tract reconstruction using bowel segments, regulatory investigations concerning renal function, acid-base balance, and vitamin B₁₂ status is essential to avoid metabolic complications.

There is an increased risk for secondary malignancies in patients with neurogenic bladder with and even without enteric bladder augmentations.^{19,20,98-104} Therefore, patients need to be informed about this risk. The most significant symptom of developing malignancy is macroscopic

hematuria as well as changes in the upper urinary tract. Although there are poor data on follow-up schemes to diagnose secondary malignancies, after a reasonable follow-up time (eg, 10-15 years), an annual cystoscopy can be considered. It is important to note that cystoscopy has not been shown to be effective in the early detection of secondary malignancies, due to the aggressiveness of the tumors.²⁰ Cytology is not useful after incorporating intestinal segments into the urinary tract.^{105,106}

As the overall prognosis of patients with myelodysplasia and neurogenic bladder dysfunction is good, the transition of follow-up into adult care should be well-coordinated in close cooperation with experienced adult urologists (Figure 1).

Summary of evidence	LE
Injection of botulinum toxin into the detrusor muscle in children who are refractory to anticholinergics/muscarinergics has been shown to have beneficial effects on clinical and urodynamic variables.	2a
Recommendations	Strength rating
Treatment of faecal incontinence is important to gain continence and independence. Treatment should be started with mild laxatives, rectal suppositories as well as digital stimulation. If not sufficient transanal irrigation is recommended, if not practicable or feasible, antegrade continence enema (ACE) stoma should be discussed.	Strong
Ileal or colonic bladder augmentation is recommended in patients with therapy-resistant overactivity of the detrusor, small capacity and poor compliance, which may cause upper urinary tract damage and incontinence. The risk of surgical and nonsurgical complications and consequences outweigh the risk for permanent damage of the upper urinary tract)+/- incontinence due to the detrusor.	Strong
In patients with a neurogenic bladder and a weak bladder outlet, a bladder outlet procedure should be offered to gain continence. In most patients this should be done together with a bladder augmentation due to the risk of a high pressure postoperatively.	Weak
Creation of a continent cutaneous catheterizable channel should be offered to patients who have difficulties in performing a CIC through the urethra.	Weak

Note: Summary of evidence and recommendations has been established by the EAU/ESPU guideline panel after reviewing and discussing the current literature.¹⁰⁷

Abbreviations: CIC, clean intermittent catheterization.

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