

# Congenital Medullary Malformations and Urinary Disorders: The Contribution of Urodynamic Assessment in the Diagnostic, Prognostic and Therapeutic Approach in the Light of Recommendations and The Literature

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## Abstract

## Original Research Article

Congenital medullary malformations or spinal dysraphisms are anomalies in the closure of the neural tube affecting the spinal column and the spinal cord. The cause is usually multifactorial. The main cause of morbidity and mortality in these children is renal deterioration. This is the main cause of neurological bladder in children. Urinary management must begin early, from the neonatal period, with the aim of preserving the urinary tract and renal function, as vesico-sphincter imbalance is the cause of renal deterioration and may therefore be life-threatening for these children. Urodynamic investigation is the key examination for analysing the functioning of the lower urinary tract and proposing a course of treatment based on the mechanisms of vesicosphincter dysfunction. There are still very few publications on urodynamics in children. Although urodynamic exploration techniques differ little from those used in adults, the pathophysiology of bladder and bowel function in infants is specific to this age group. This article reviews the literature to demonstrate the value of urodynamic testing in the diagnosis and treatment of urinary disorders in congenital medullary malformations.

**Keywords:** spinal dysraphisms, renal deterioration, vesicosphincter dysfunction, urinary disorders.

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## GENERAL

Spina bifida or spinal dysraphism is one of the neural tube closure anomalies affecting the spinal column and spinal cord, occurring during embryogenesis [1].

The prevalence of open spinal dysraphism according to this register is estimated in Europe between 2005 and 2015 at 5.1/10000 pregnancies (live births + foetal deaths + medical terminations of pregnancy), with 1.8/10000 live births and 3.2/10000 terminations of pregnancy.

The commonly used classification of spinal dysraphism distinguishes between open and closed dysraphism. In open dysraphism, there is a defect in dorsal closure not only of the vertebrae and the musculoaponeurotic plane but also of the skin.

There are several clinical forms of closed dysraphism depending on the presence or absence of a

subcutaneous mass. Closed spinal dysraphism is becoming the major cause of neurological bladder disease [2].

## OBJECTIVE

Our objective is to show the interest of urodynamic assessment in the diagnostic and therapeutic approach to urinary disorders in congenital medullary malformation through a review of the literature.

## METHODOLOGY

We review the contribution of urodynamic assessment through publications and recommendations from learned societies. We also consulted the registers and protocols of centres specialising in the management of bladder and bowel disorders in patients with spinal dysraphism.

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## RESULTS

Diagnostic, prognostic and pre-therapeutic benefits of urodynamic testing Urodynamics is the gold standard for assessing lower urinary tract function in patients with spinal cord disease. Vesico-sphincter disorders in spina bifida are polymorphous. They depend

on the site of the malformation, its extent, the form and degree of damage to the nerve structures at the site of the malformation and whether or not it is associated with another central nervous system malformation [3]. For the sake of clarity, we will outline four very schematic types of vesico-sphincter disorders [4, 5].

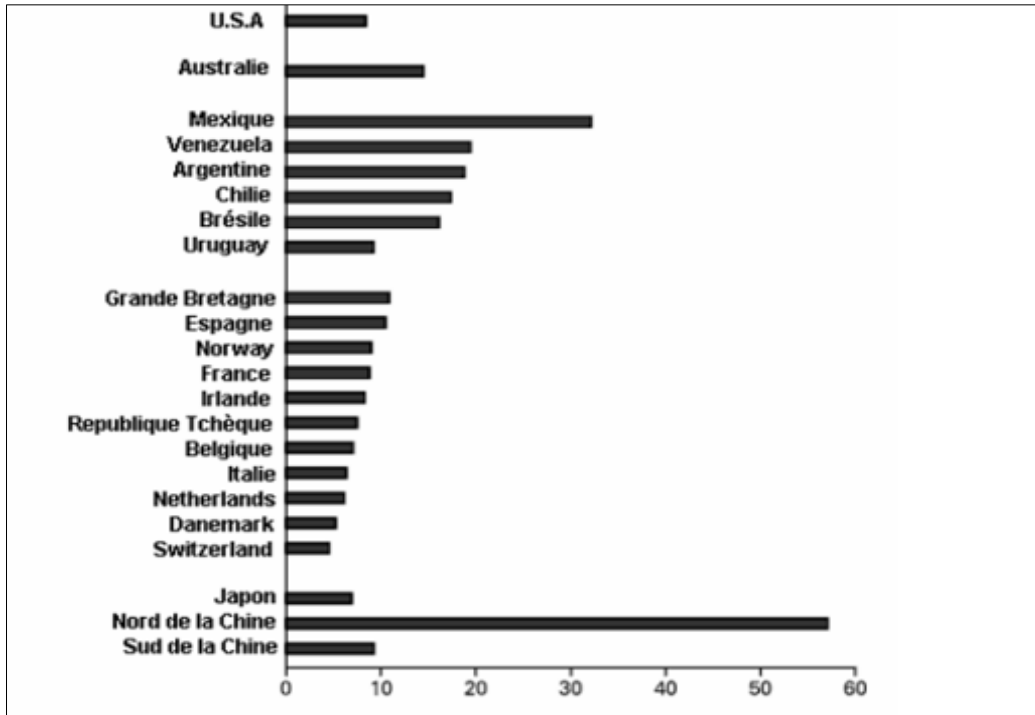
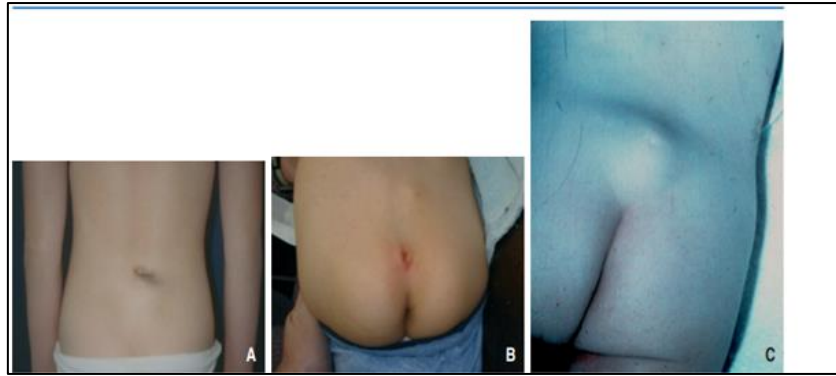


Figure 1: Graph illustrating variations in the rate of neural tube closure defects in different countries around the world



Figure 2: Large lumbar myelomeningocele in a newborn baby



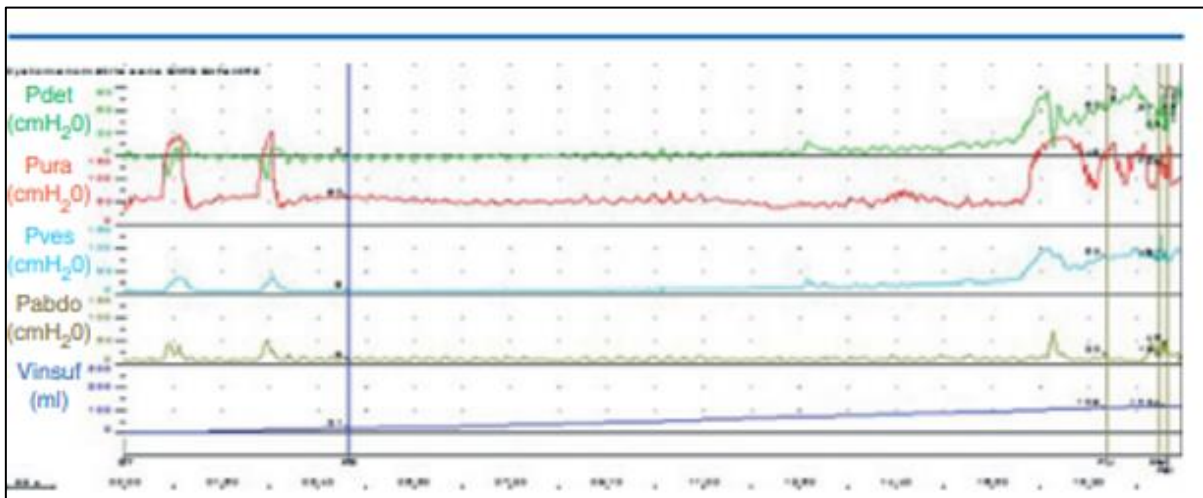
**Figure 3. Occult dysraphism with pigmented spot and hairiness on the dorsolumbar median raphe (A) demic sinus(B) deviation of the gluteal fold and retro sacral lipomatous mass(C)**

**Table 1: The distribution of these four types of vesico-sphincter disorders**

Type	Hyoactive bladder	Hyperactive bladder	Normal bladder	Total
Sphincter hypotonia	44	20		65
Sphincter hypertonia	26	85		111
Normal sphincter			13	13
Total	70	105	13	188

The impact of vesico-sphincter disorders is twofold: social, through its consequences on continence, and vital, through its consequences on the upper

apparatus and the risk of degeneration of the bladder wall [6].



**Figure 4. Detrusor overactivity on urodynamic assessment**

**After Dysraphism Surgery**

Studies show that routine urodynamic testing is necessary after surgery. According to the HAS, there is no recommendation or consensus on the time frame for urodynamic testing. In some studies, it has been postponed for several weeks to eliminate the consequences of spinal shock following neurosurgical closure of the neural tube [7].

Medical management of bladder and bowel disorders Analysis of the National Spina Bifida Patient Registry from 2009 to 2015, involving 5,250 patients, showed that 92.6% of patients received treatment for neurological bladder disease [8].

This bladder damage predisposes to the risk of chronic renal failure [9]. Our analysis focused mainly on the following recent international recommendations:

Brazilian, published in 2021; European, published in 2019; British, published in 2016 and International (International Children's Continence Society), published in 2012. These recommendations are based on systematic reviews of the literature. There is no national or international consensus on management, and there is considerable variability in the diagnostic and therapeutic management of urological disorders associated with dysraphism [10].

Urodynamic investigations are recommended in newborns with dysraphism, but the time taken to perform them varies according to the situation. According to European recommendations:

- in the case of MMC with postnatal repair, they should be carried out at a distance from the surgery in order to limit the effects of the latter (at 2/3 months of life) [11].
- in the case of MMC with antenatal repair, they can be performed in the first few days of life
- for all other types of dysraphism, urodynamic investigations may be carried out if there is a clinical or radiological suspicion of a neurological bladder.

According to Brazilian recommendations: Urodynamic testing should be carried out within the first 6 months of life.

The following data should be determined:- detrusor leak point pressure- start and end of filling pressure- bladder capacity If urodynamic investigations are normal, they may be repeated at 1 year of age.

The time frame for initiating intermittent urinary catheterisation according to the recommendations:

- European recommendations [12], systematic, from the first days of life
- British recommendations [13], the experts do not decide between an expectant attitude with delayed initiation or a proactive attitude with immediate initiation.
- Brazilian recommendations [14], systematic, from the first days of life
- Recommendations of the International Children's Continence Society [15], to be introduced from the first days of life if the

newborn does not empty the bladder spontaneously (at least until urodynamic investigations) or if there is high-grade vesico-ureteral reflux. They should be repeated 5 to 7 times a day [16].

The introduction of intermittent bladder catheterisation during the initial hospital stay will help to educate the parents, who should be able to leave the hospital independently.

Overall, it is difficult to conclude whether one approach is superior to the other. However, it should be noted that in the case of "reactive" management, the child should be closely monitored.

L. B Johnston carried out a study in 1998 on 51 children with an average age of 3 years presenting with occult spinal dysraphism. He found that the most frequent urodynamic abnormalities were detrusor overactivity (82%) and bladder-sphincter dyssynergia (43%) [17].

L. T Lavalée carried out a study in 2013 on a cohort of 121 children with occult spinal dysraphism with an average age of 11 months, mixing patients operated on by spinal cord release (33%) and not (67%). In this study, he found that the most frequent urodynamic abnormalities were bladder hyperactivity (35%), low capacity (33%) and low compliance (30%). In this study, bladder-sphincter dyssynergia was not studied [18].

Agnès diagnosed vesico-sphincter dysfunction in 75% of cases. The most frequent urodynamic manifestations were vesicosphincter dyssynergia (53%) and detrusor overactivity (45%), and 15% of cases of sphincter insufficiency based on urodynamic criteria.

**Table 2: Urodynamic abnormalities in occult spinal dysraphism**

Autor	L.B Johnston 1998	Nogueira 2004	Lavalle 2013
Population	51 children mean age 3 years	Population 16 children under 3	years 123 mean age 11 months
Detrusor overactivity	82%	25%	35%
Low compliance	50%	Non reinstated	30%
Vesico spherical dyssynergia	43%	25%	Non reinstated

Urodynamic assessment before and after dysraphism surgery.

A review summarising 17 articles published over the last 25 years. It showed that spinal cord release has a positive effect on urological symptoms and urodynamic parameters.

The publications reported different levels of success with spinal cord release surgery: urodynamic

improvement concerned between 5% and 93% of patients, depending on the study.

This review also showed that early intervention significantly prevents traction on the spinal cord, thereby avoiding irreversible neurological damage [19- 21]. All the studies found a significant improvement in capacity and compliance after spinal cord release surgery.

**Table 3: Comparison of urodynamic results after spinal cord release in the literature**

	<b>Khoury 1990, 31 children</b>	<b>Noguerira 2004, 54 children</b>	<b>Guerra 2006, 24 children</b>	<b>Yenner 2015, 40 children</b>
Pathology	Occult dysraphism	Dysraphism and attached medulla	Dysraphism and attached medulla	Dysraphism and attached medulla
Urodynamic improvement		From 40% to 100% depending on the sub-group	48%	54%
Urodynamic improvement	0%		8%	12%
Disappearance of bladder hyperactivity	59%		59%	Non su
Improved compliance	66%		Average improvement of 44ml	41%
Increase in bladder capacity				10%
Reduction in post -micturition residue				25%

## CONCLUSION

The management of vesico-sphincter disorders in patients with spinal dysraphism is complex, and requires knowledge of both the pathophysiology of the spinal cord malformation and the management of the neurological bladder in children.

Patients with spina bifida have a high risk of damage to the upper urinary tract. The proportion of patients suffering from urinary incontinence in this population is very high. Urodynamic assessment appears to be able to identify patients at greatest risk of upper urinary tract damage.

The treatment of neurological urinary incontinence remains a difficult problem. No indication can be accepted without a multidisciplinary approach and without taking account of the family environment of these children. Regular monitoring of these patients is essential to prevent urological complications.

The value of early urodynamic assessment in the case of spinal dysraphism is to understand the mechanism of vesico-sphincter dysfunction, to identify children at high renal risk, and to anticipate the child's future continence potential.

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