



Long-term urologic outcome in patients with caudal regression syndrome, compared with meningomyelocele and spinal cord lipoma

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Abstract

Background/Purpose: The long-term urologic outcome in a large series of patients with neural tube defects was evaluated.

Methods: The following clinical parameters in 398 patients ranging from 1 to 37 years of age—69 with caudal regression syndrome (CRS), 244 with meningomyelocele (MMC), and 85 with spinal lipoma (SL)—were studied: congenital renal anomalies, renal function, vesico-ureteric reflux, upper tract dilatation, urodynamic pattern, and urinary continence.

Results: Single kidney was much more frequent in CRS (20.3%), compared with MMC (1.2%) and SL (0%). Vesico-ureteric reflux was found in 37.7% of patients with CRS, 43.0% of MMC, and 21.2% of SL. Patients with CRS had a higher risk of impaired renal function (8.7%), compared with MMC (5.3%) and SL (1.2%). Neuropathic bladder was found in 61% of patients with CRS, 98% of MMC, and 42% of SL. Among them, clean intermittent catheterization and drugs allowed 30% of patients with CRS, 45% of MMC, and 71% of SL to be dry for more than 4 hours.

Conclusions: Diagnosis influences the urologic outcome in neural tube defect. In CRS, the incidence of renal agenesis and vesico-ureteric reflux was unexpectedly high. The risk of renal damage and, in those with neuropathic bladder, of urinary incontinence, was similar to patients with MMC.

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Patients with neural tube defect (NTD) may have very different urologic outcomes. Meningomyelocele (MMC) is an open NTD, usually associated with a cascade of malformations as well as hydrocephalus, Chiari II, hydro-myelia, and low-placed spinal cord. Almost 100% of them have some degree of neurologic impairment. Spinal lipoma (SL) and caudal regression syndrome (CRS) are closed

NTDs, not always associated with neuropathic bladder (NB) but presenting different urologic patterns with a great variability [1]. Although prognosis is believed to be correlated with the type and severity of NTD, and in general, MMC is considered more severe than SL or CRS, there are no studies in the literature comparing the long-term urologic outcome in different populations of NTD treated by the same team. In particular, the urologic outcome in patients with CRS patients has not been studied yet in large series of patients. Moreover, urologic results in patients with NTD are

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obviously influenced by the combination of medical and surgical treatments received, so that it is difficult to establish the prognostic factor of the single NTD in the determination of the outcome. The aim of our study was to assess the risk of urologic impairment according to the diagnosis of NTD. In particular, we studied a large population of patients with CRS, and we compared the results with patients with MMC and SL.

1. Methods

In our institution, patients with NTD are followed-up after the diagnosis by a multidisciplinary team including, among others, neurosurgeons, pediatric urologists, and nephrologists. Only the cases with NTD diagnosed by magnetic resonance and with complete urologic assessment were included. The patients considered for this study were those with diagnosis of MMC, CRS, or SL; in our institution, the neurosurgical procedure is performed at birth on all patients with MMC; before 6 months, if possible, on patients with SL; and only in selected cases on patients with CRS. Few patients have been operated in other hospitals. We excluded from this study the patients who underwent augmentation cystoplasty or bladder neck surgical or endoscopic procedures, representing less than 5% of all patients. We included the cases treated surgically or endoscopically for vesico-ureteric reflux (VUR).

We studied 398 patients (196 males and 202 females): 244 with MMC (122 males and 122 females), 85 with SL (38 males and 47 females), and 69 with CRS (36 males and 33 females), ranging from 1 to 37 years of age. The mean age of the patients at the last control was 13 years and 5 months for MMC, 12 years and 7 months for SL, and 9 years and 4 months for CRS.

The follow-up ranged from 12 months to 14 years, with a mean of 7 years and 9 months. Every case was studied with clinical evaluation, renal function (plasmatic creatinine and urea, creatinine clearance calculated with the Schwartz formula [2], radio isotopic scan in most cases), ultrasounds, cystogram, and urodynamic studies. Clinical evaluation included weight and size, urinary tract infections, continence, voiding pattern, and use of drugs.

The following parameters in these 3 populations were studied: congenital renal anomalies, renal function, VUR, upper tract dilatation, urodynamic pattern, and urinary continence. Congenital renal anomalies considered were: single, horseshoe, and multicystic kidney.

Renal function was considered mildly compromised if multiple bilateral scars (or monolateral in a single kidney) were present at renal scan without reduction of creatinine clearance calculated with the Schwartz formula [2]. We considered as severely compromised renal function every case with reduced creatinine clearance with respect to the normal values for age, according to the Italian guidelines [3]. Patients were considered to have VUR if in at least

1 cystogram or scan, VUR of some degree was detected, independently from the eventual treatment and evolution. Patients with VUR were evaluated and divided into 2 groups: those with NB and nonneuropathic bladder (NNB).

The upper tract dilatation was evaluated by ultrasound and according to the guidelines of our institute: we considered the upper tract dilated if the anteroposterior diameter of the pelvis was more than 7 mm at less than 6 months of life or more than 10 mm after 6 months or if ureteral diameter was more than 10 mm.

Urodynamic and clinical pattern allowed us to differentiate between the patients with NB and those with NNB. The patients with normal continence, voiding pattern, and urodynamic study results were considered to have NNB. A bladder was considered NB if incontinent, with pathologic urodynamics and impaired bladder voiding.

From the clinical point of view, patients with NB were considered as "socially" continent if dry for at least 4 hours, independent of the use of clean intermittent catheterization (CIC) or anticholinergics (balanced NB). The others were considered as incontinent and divided into 3 groups: with mild, moderate, and severe incontinence if they were dry more than 3 hours, between 1 and 3 hours, and less than 1 hour, respectively. The evaluation of urinary continence was possible in children older than 3 years.

Collected data were analyzed with Fisher's exact test to verify a possible statistically significant difference ($P < .05$) between the 3 populations studied.

2. Results

Data are shown in Table 1.

We found significantly more frequent congenital anomalies of the kidney, in particular, of agenetic kidney, in patients with CRS. For both of these, $P < .01$.

No significant difference was observed in renal function impairment and upper tract dilatation among the 3 populations.

Regarding social incontinence among NB cases, patients with CRS were significantly more incontinent than the other patients ($P < .05$).

3. Discussion

In the evaluation and follow-up of a patient born with NTD, the urologic assessment is of utmost importance because of the possibility of renal damage mainly caused by voiding impairment, VUR, and infection. A precise diagnosis by magnetic resonance and classification of the NTD [4] must be the first step of the evaluation. Among NTDs, MMC is considered to have a worse prognosis than SL and CRS, but it is difficult to demonstrate it scientifically using clinical data. In the present study, we were able to

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