Spinal dysraphism and neurogenic bladder: Still a relevant topic

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Abstract

Background: The spinal dysraphism (SD) is a common disease in children. It remains a current topic because of its bladder and sphincter disorders’ (BSD) impact involving the vital and functional prognosis. Myelomeningocele is the most severe form of spina bifida (SB). Objective: To highlight the BSD in SD patients and their features. Materials and methods: We conducted a retrospective study including patients followed in our Physical and Rehabilitation Medicine (PRM) Department and having BSD associated with SD. We collected epidemiological and clinical characteristics, such as neurologic level, orthopedic deformities, assistive device use, and level of ambulation in addition to biological, radiological and urodynamic features, care and follow-up. Results: Fifty patients, 30 girls and 20 boys, average age 10.33 ±1.28 years, mean length of follow-up 5.8 years. The most common type of SD was myelomeningocele, 30 had an independent ambulation. Urinary symptoms were dominated by urinary incontinence. The serum creatinine was disrupted in 18 cases including one with end-stage renal disease performing hemodialysis. Urine tests showed a urinary tract infection in 35 patients. The radiographic abnormalities were hypertonic bladder in most cases. Urodynamic exploration objectified an overactive bladder in most cases. The preferential micturition way was intermittent catheterization associated, in most cases, with anticholinergic treatment and in one case the intra-detrusor botulinum toxin. Conclusion: The BSD in children with SD remain a major issue requiring early and multidisciplinary care to preserve the integrity of the upper urinary tract and to improve the quality of life.

Keywords: Spinal Dysraphism, Urinary Bladder Diseases, Long-term care, Rehabilitation.

INTRODUCTION

Neuropathic bladder caused by SB remains an important cause of chronic renal failure in developing countries [1, 2]. In contrast, recent reports from western countries showed that children born with SB can avoid such complication if they are provided adequate urological intervention [3]. Early therapy with clean intermittent catheterization to decrease intravesical pressure is the preferred treatment, and antimuscarinic agents to counteract detrusor instability [4, 5] help in safe guarding renal function for such children. Optimal therapy is not usually available for children living in developing or underdeveloped countries [6].

SB is caused by a failure of the caudal neural tube to fuse normally in early development. A variety of neurological deficits can be seen, depending on the severity of the fusion abnormality and location of the lesion. Variable impact on the somatic, parasympathetic and sympathetic innervation of the bladder affects the ability to store and empty urine, and can ultimately cause chronic kidney disease due to poor bladder dynamics. Urological issues can be a significant source of morbidity and mortality, and are implicated as a cause of death in almost a third of patients with open SB followed long term [7].

In this study, we aim to investigate the bladder and sphincter disorders (BSD) in SB children who were followed up at our department, and their features.

MATERIALS AND METHODS

We conducted a retrospective study between 2000 and 2016 including all patients followed in our Physical...
and Rehabilitation Medicine (PRM) Department and having BSD associated with spinal dysraphism (SD).

We have collected data including age, gender, history of ventriculoperitoneal shunting, neurologic level (S2 below, sacral; L5/S1, lumbosacral; L3/L4, mid-lumbar; L1/L2, high lumbar; thoracic), spinal and foot deformities, hip problems (including hip subluxation, dislocation, and surgery), use of assistive devices, use of lower extremity orthotics or walking aids, complications from assistive device use, and ambulation status. In addition, we have studied the type of treatment, antimuscarinic agents, clean intermittent catheterisation (CIC), and antibiotic prophylaxis. Renal function, ultrasound, micturatingcystourethrogram (MCUG), serum creatinine, and bladder function (urodynamic studies) were evaluated.

RESULTS

Data of 50 children with spinal SD were evaluated. 30 girls and 20 boys with average age 10.33 ±1.28 years. The mean length of follow-up was 5.8 years. The SD most common type was myelomeningocele in 32 cases (64%)(Fig. 1). The distribution of neurologic and ambulation levels is listed in Table 1. Most subjects (62%) were with a low neurologic level (sacral plus lumbosacral) and paraplegic (Fig. 2) with hollow feet (Fig. 3). Ankle-foot orthoses and walkers were the most commonly used assistive devices (Fig. 4, Fig. 5, Fig. 6, Fig. 7). Moreover, 32% of the studied population had an aligned spine (Fig. 8, Fig. 9, Fig. 10), 38% of cases were diagnosed with hip dislocation, 52% had hollow feet and 84% used orthosis. Hydrocephalus was present in 14% of cases who did a ventriculoperitoneal shunting (Fig. 11, Fig. 12). Urinary incontinence was found in 28 patients (56%)(Fig. 13).Urinary complications are listed in Table 2. The serum creatinine was disrupted in 18 cases including one with end-stage renal disease performing hemodialysis. Urine tests showed a urinary tract infection in 35 patients. Twenty (40%) patients diagnosed to have hypertonic bladder(Fig. 14, Fig. 15), vesicoureteral reflux in 10 cases (Fig. 16) and uretero-hydronephrosis in 5 cases. The preferential micturition way was intermittent catheterization associated, in most cases, with anticholinergic treatment and in one case the intra-detrusor botulinum toxin (Fig. 17).

Table 1: Distribution of neurologic level and ambulation level

<table>
<thead>
<tr>
<th>Neurologic level</th>
<th>n (%)</th>
<th>Ambulation level</th>
</tr>
</thead>
<tbody>
<tr>
<td>No</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td>S</td>
<td>12 (24)</td>
<td></td>
</tr>
<tr>
<td>LS</td>
<td>19 (38)</td>
<td></td>
</tr>
<tr>
<td>ML</td>
<td>9 (18)</td>
<td></td>
</tr>
<tr>
<td>HL</td>
<td>8 (16)</td>
<td></td>
</tr>
<tr>
<td>T</td>
<td>1 (2)</td>
<td></td>
</tr>
<tr>
<td>C</td>
<td>1 (2)</td>
<td></td>
</tr>
<tr>
<td>Community</td>
<td>24 (48)</td>
<td></td>
</tr>
<tr>
<td>household</td>
<td>6 (12)</td>
<td></td>
</tr>
<tr>
<td>Non-ambulator</td>
<td>20 (40)</td>
<td></td>
</tr>
</tbody>
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No = no neurologic deficit; S = sacral; LS = lumbosacral; ML = midlumbar; HL = high lumbar; T = thoracic; C = cervical.

Table 2: Urinary complications

<table>
<thead>
<tr>
<th>Urinary symptoms</th>
<th>Yes (%)</th>
<th>No</th>
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<tbody>
<tr>
<td>Renal failure</td>
<td>18 (36%)</td>
<td>32</td>
</tr>
<tr>
<td>Urinary tract infection</td>
<td>35(70%)</td>
<td>15</td>
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Figure 5: Urodynamics abnormalities

Figure 6: Micturition way

Figure 7: Lumbar surgical scar

Figure 8: Ventriculoperitoneal shunting scar

Figure 9: Conflicting Knee-Ankle-Foot-Orthosis

Figure 10: Knee-Ankle-Foot-Orthosis with abduction bar

Figure 11: X-ray of the spine showing kyphoscoliosis and ventriculoperitoneal shunting

Figure 12: X-ray of the spine showing kyphosis

Figure 13: X-ray of the spine showing L3 level SD
Our results demonstrate that our studied spina bifida children developed a considerable BSD. 36% of our cohort had renal failure, while Dik and al have reported recently that only 6 out of 144 (4%) children with spina bifida had evidence of renal scars at the age of six years [3]. Lewis and al in 1994, reported that the prevalence of renal parenchymal damage was 19.4% with higher prevalence of parenchymal damage in children over 10 year of age (27.3%), twice that of the 13.3% under 5 years of age [9]. Remarkable incidence of renal damage could be explained by the delay in proper management, as regular emptying of the bladder might be not commenced early and anticholinergic drugs might be not instituted in some children. Furthermore, we insist in appropriate practices and management including early diagnosis and treatment of acute pyelonephritis that could also have contributed to the worse outcome in these patients. Thus, multi-disciplinary specialized spina bifida clinics should be available where children have an easy access. Establishing such clinics, would help to reduce this observed delay in commencing the appropriate management to protect the kidneys. However, our cohort is rather advantaged group as many of them had CIC associated with anticholinergic treatment. Early investigation and management of neurogenic bladder is crucial to protect the kidneys [3, 10]. Early start of CIC is the most important factor to avoid renal damage. It was reported that the prognosis of children with upper renal tract changes at birth did not seem to be any worse than children developing changes later in life [11]. Five of the 6 patients with renal scarring in Dik and al report, were started on therapy with intermittent catheterization and antimuscarinic therapy several months after birth [3]. CIC is not accepted by many families as modality of therapy. It has psychosocial impact on the treated children and their families [12, 13]. Furthermore, advice from the practitioners and health care professionals, and parental guidance to promote the CIC was also a major factor. Patients, who were not receiving CIC, had a higher incidence of vesicoureteral reflux, urinary tract infections and renal scars. One patient progressed to renal failure with hemodialysis. This further stresses the need for an aggressive approach for optimal care of the bladder to protect already compromised kidneys.

Accurate prenatal diagnosis of various forms of SD by ultrasound with determination of severity and prognostic factors is vital, due to a potentially high termination rate (up to 65%) [14]. Ultrasound screening improves prenatal diagnosis and can also characterize the anatomical nature of the lesion, allowing identification of the level of the lesion, with low spinal lesions being associated with increased bladder dysfunction [15]. Although expensive, fetal MRI differentiates unique anatomical features. Besides influencing the decision regarding termination, prenatal diagnosis also allows for consideration of prenatal closure of the defect. Prenatal vs postnatal closure was evaluated in a multicentral, randomized, controlled trial [14]. Initial
results revealed a decreased need for ventriculoperitoneal shunting and improved lower extremity motor outcomes. However, these benefits were partly offset by an increased incidence of preterm delivery and uterine dehiscence in those who underwent prenatal intervention. While awaiting the urological results, several groups have evaluated the results of prenatal intervention on urological outcomes using case-control methodology and patient cohorts that were mostly closed\[17, 18\]. They have noted no significant differences between prenatal and postnatal closure regarding need for CIC, rate of urinary incontinence, need for reconstructive urological surgery or urodynamic parameters. Carr found that patients undergoing closure prenatally had 18.5% success with toilet training, compared to 8.3% of patients in a historical group who had normal urodynamics after undergoing closure postnatally\[19\]. While it is unclear if prenatal closure will significantly improve bladder outcomes, delaying neurosurgical closure past 72 hours has been shown to increase the incidence of febrile urinary tract infections, vesicourethral reflux, hydronephrosis and worse urodynamic parameters. Surgical intervention within 24 hours improves bladder capacities and decreased detrusor leak point pressure\[20\].

CONCLUSION
We have shown that an early therapy of BSD resulted in a better prognosis in children born with SB. Children with poor compliance had higher incidence of VUR and UTI. There is a need of more awareness about the importance of starting proactive treatment to prevent renal damage in children with SB.

Conflict of Interest
The authors declare that they have no conflict of interest.

Informed consent
Informed consent was obtained from all individual participants included in the study.

REFERENCES