Original Article

URINARY TRACT COMPLICATIONS IN PATIENTS WITH SPINAL DYSRAPHISMS

Mostafa Sharifian[1], Navid Anvaripour[2], Alireza Zali[3]

ABSTRACT

Objective: To determine the prevalence of urinary complications and to review the management of these complications in patients with spinal dysraphism.

Methodology: This is a cross-sectional descriptive study in which Ninety four patients with various types of spinal dysraphism were studied. The mean age of patients was 7.2 years (range: 0.1-35 years), 56.4% were males. Sixty eight patients (72.3%) had at least one episode of pyelonephritis, hydronephrosis in 34.1%, vesicoureteral reflux in 35.1%, renal atrophy in 13.9% and end stage renal failure in 3.2% cases.

Results: Among 61 patients aged four years or older, forty seven (77%) had urinary incontinence. Seventy six cases (80.9%) had paraclinic findings in favour of neurogenic bladder. Of these, forty five (59.2%) received medical treatments (CIC ± anticholinergic agents). Anti-reflux surgery was performed in nine patients (9.6%); subsequently, vesicoureteral reflux relapsed in six cases.

Conclusion: Our study revealed that urinary complications were common in patients with spinal dysraphism. Early appropriate management based on urodynamic studies and careful follow-up of patients with spinal dysraphism will significantly decrease the occurrence of urinary complications in these children.

KEY WORDS: Spinal dysraphism, Neurogenic bladder, Urodynamic study, Vesicoureteral reflux, Urinary incontinence, Detrusor-sphincter dyssynergia.

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1. Mostafa Sharifian,
Pediatric Infectious Research Center (PIRC),
2. Navid Anvaripour,
Department of Pediatrics,
3. Alireza Zali,
Department of Neurosurgery,
1-3: Mofid Children Hospital,
Shaheed Beheshti University of Medical Sciences,
Tehran, Iran

Correspondence

Mostafa Sharifian, MD
Pediatric Infectious Research Center (PIRC),
Shahid Behasti Uni. M.C.,
Mofid Children Hospital,
Shariati Ave, Tehran, Iran.
Email: mostafasharifian@yahoo.com

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INTRODUCTION

Defective closure of the caudal neural tube at the end of week four of gestation results in anomalies of the lumbar and sacral vertebrae or spinal cord called spina bifida or spinal dysraphism. These anomalies range in severity from clinically insignificant defects of the L5 or S1 vertebral arches (occult spinal dysraphism) to major malformations of the spinal cord that lies uncovered by skin or bone on the infants back (myelomeningocele).[1] Urinary system abnormalities are about the most important and critical complications of these defects. More than 90% of infants with myelomeningocele have a neurogenic bladder,

with 30% having lower motor neuron lesion.[2] The most important consequences of nerogenic bladder dysfunction are urinary incontinence, pyelonephritis, and upper tract deterioration. Failure of the urethral sphincter to relax during a bladder contraction (detrusor-sphincter dyssynergia) results in functional obstruction of the bladder outlet and subsequently high intravesical pressure, detrusor hypertrophy and terabeculation, vesicoureteral reflux (VUR), hydroureteronephrosis and renal damage. More than 95% of children with spinal dysraphism suffer from urinary incontinence.[3]

The advent of clean intermittent catheterization (CIC) in the early 1970s by Lapides,[4] refinements in techniques of urodynamic studies (UDSs) in children and development of surgical modalities to manage incontinence dramatically changed the way this group of children were traditionally managed. Most pediatric urologic centers now believe that functional assessment of the lower urinary tract is an essential element in the evaluation process and managment of these patients.[5]

According to the literature, proper management of patients with spinal dysraphism dramatically reduces the urinary complications of these children. Hence we studied patients with spina bifida to determine the prevalence of urinary complications and to review the management of these complications.

METHODOLOGY

In this descriptive study, we evaluated urinary complications in patients with various types of spinal dysraphism who attended out-patient clinics between July 1999 and March 2005. (Fig-1)

The type of spinal dysraphism, time of spinal reconstructive surgery, number of pyelonephritis episodes were determined. We determined anatomical abnormality in urinary tract by ultrasonography, VUR and bladder terabeculation by voiding cystourethrography and renal scarring by DMSA scan. Urinary incontinence was studied in children four years or older. Using UDSs, we also determined the prevalence of neurogenic bladder; paraclinic findings in favor of neurogenic bladder such as hydronephrosis, thickening of bladder wall and abnormal urinary residue in sonography, bladder terabeculation with or without VUR in voiding cysto-urethrography (VCUG) and bladder terabeculation in cystoscopy were determined. We also determined medical treatments (CIC and anticholinergic drugs) and surgical interventions used for the patients.

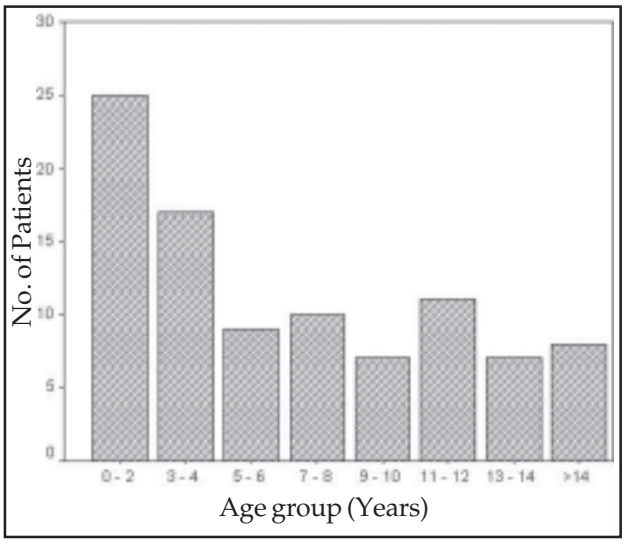

Figure-1: Age distribution of patients.

The collected data were analyzed using SPSS 9.0 and Exel 2003 software.

RESULTS

We included in our study 94 patients with spinal dysraphism. Of these 70 patients, seventy (74.5%) had myelomeningocele, two (2.1%) lipomeningocele, one (1.1%) meningocele, one (1.1%) sacral agenesis and 20 patients (21.3%) were affected by other types of occult spinal dysraphism. Of the 20 patients with occult spinal dysraphism (OSD), the spinal lesions were diagnosed in only seven patients (35%) until the age of three years.

The mean age of spinal surgical repair in patients with myelomeningocele was 86.4 days (ranged from one day to two years). Out of 70 patients with this lesion, only nine cases (12.9%) underwent surgical repair during the first 72 hours of life. Fifty seven patients (81.4%) underwent reconstructive surgery between three days and one year after birth.

Based on DMSA scan, pyelonephritis occurred in 68 children (72.3%); 23 cases (24.5%) had one episode, 22 cases (23.4%) had two, 10 patients (10.6%) had three, and 13 patients (13.8%) had more than three episodes of febrile UTI.

According to the ultrasonographic studies, 68 patients (72.3%) had abnormal findings; renal atrophy were reported in 13 patients (13.9%) which was bilateral in three cases (3.2%). Thirty two patients (34.1%) had hydronephrosis, of which 20 cases (21.3%) presented on both sides. In 51 (54.3%) patients, an increased thickness of bladder wall was present. Thirty eight patients (40.4%) had abnormal post voiding residue in the bladder.

Seventy five patients (79.8%) had abnormal findings in VCUG study. We found VUR in 33 patients (35.1%) which had involved both sides in 20 cases (21.3%); two patients (2.1%) with grade I, three (3.2%) with grade II, nine (9.6%) with grade III, thirteen (13.8%) with grade IV, and six patients (6.4%) with grade V. Furthermore, bladder trabeculation was found in 71 cases (75.6%); twenty nine patients (30.9%) had moderate and 23 cases (24.5%) had severe bladder trabeculation.

DMSA scan was performed in 89 patients. Renal scar was reported in 32 cases (35.9%) (bilateral in 66.3% of patients). Decreased cortical function in one or both sides was also present in 59 patients (66.3%).

Twenty three patients underwent cystoscopic studies. It was abnormal in 21 cases; nineteen patients had some degrees of bladder trabeculation, 14 cases had a decreased bladder capacity and nine patients had abnormal urethral orifices.

Out of 61 patients aged four years or older, 47 cases (77%) suffered form urinary incontinence. Medical treatment was started in 19 patients, in nine cases it was unsuccessful; these patients later on needed surgical interventions to achieve continence (cytosplasty in eight cases and cystosplasty plus bladder neck reconstruction with Young-Dees method in one patient). In 16 cases, surgical intervention (cystoplasty) was performed without any previous medical treatment.

For evaluating the lower urinary function UDS was performed in 16 patients (17%). The mean age of patients at the time of these studies was 7.9 years (ranged six months to 22 years). Neurogenic bladder was found in all of these cases.

According to the finding of UDSs, ultrasonography, cystography and cystoscopy, 76 patients (80.9%) had finding in favour of neurogenic bladder. From 76 patients with neurogenic bladder, 45 cases (59.2%) received medical treatments for decreasing the complications of this disorder such as urinary incontinence and VUR; four patients (5.3%) treated with CIC, 18 cases (23.7%) with anticholinergic agents (including oxybutinin) and twenty three (30.3%) with combination of CIC and anticholinergic agents. The mean age of patients at the onset of treatment was 4.8 ± 4.4 years. (Table-I)

From 94 patients, three cases (3.2%) ended in end stage renal disease (ESRD) and all of them underwent renal transplantation. Six cases (6.4%) underwent vesicostomy because of severe hydronephrosis and urinary retention. Surgery was performed in two other patients (2.1%) to improve urinary incontinence secondary to bladder or uretheral insufficiency; bladder neck reconstruction with Young-Dees method in one patient and Burch colposuspension in the other. Creating a continent catheterizable stoma using appendix (Mitrofanoff Principle) was performed in two patients (2.1%). Furthermore, cystoplasty

Table-I: Treatments used in patients with neurogenic bladder

Treatments	*No. of Patients*	*%*	*Cumulative %*
CIC*	4	5.3	5.3
Anticholinergic Agents	18	23.7	29
CIC+Anticholinergic Agents	23	30.3	59.2
No	31	40.8	100.0
Total	76	100.0	

*CIC: clean intermittent catheterization

was performed in 33 patients (35.1%); ileocystoplasty in 26 cases, sigmoid cystoplasty in three, bladder autoaugmentation in three and gastrocystoplasty in one patient.

Surgical treatment of VUR was performed in nine patients (9.6%); cystoplasty and anti-reflux surgery was done at the same time in two patients. Five patients underwent anti-reflux surgery alone which was followed by relapse of VUR and finally anti-reflux surgery plus cystoplasty was performed in all of them. Endoscopic injection of Teflon was used in one patient that failed and the patient underwent anti-reflux surgery and cystoplasty. Later on in one patient, anti reflux surgery was done alone, but he did not come back for a follow up visit to evaluate the outcome of operation. (Fig-2)

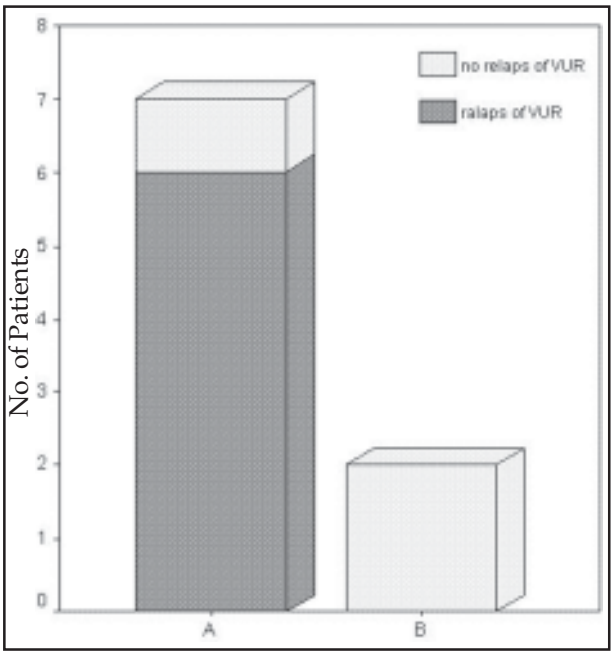

Figure-2: Type of surgical intervention in patients with vesicoureteral reflux.
A: only antireflux surgery.
B: antireflux surgery plus cystoplasty

DISCUSSION

Spinal dysraphism may be associated with serious and often irreversible complications such as urinary tract abnormalities. Early appropriate management can almost always prevent these complications. In our study, the mean age of patients at the time of myelomeningocele repair was 86.4 days after birth. According to the literature, this procedure can be performed safely up to 72 hours after birth[6] and any delay after 72 hours markedly increases the chance of meningitis or ventriculitis.[7] If for any reason the repair must be delayed beyond 72 hours after birth, cultures from the lesion should be obtained and confirmed to have no growth before the myelomeningocele repair. If cultures are positive, the patient should be treated with external ventricular drainage and antibiotics until the infection clears.[8] However in this review, only 12.9% of all cases underwent the procedure in first 72 hours after birth.

When the patients with OSD are evaluated in the newborn period or early infancy, the majority have a perfectly normal neurologic examination.[9] Urodynamic testing, however, reveals abnormal urinary tract function in about one third of babies younger than 18 months of age.[10] In contrast, practically all individuals older than three years of age who have not been operated or in whom an OSD has been lately diagnosed have either an upper or lower motor neuron lesion or a combination thereof on urodynamic study.[9,11] When such children were observed expectantly from infancy after the diagnosis was made, 58% deteriorated within two years.[12] Correction of the lesion in infancy has resulted not only in stabilization but also in improvement in the neurologic picture in many instances[13] in older children there is a less dramatic change after surgery.[10,11] According to our study, from 20 patients with OSD, only six (31.6%) were diagnosed before three years of age (the mean age of patients at the time of diagnosis 4.8 years) and none were diagnosed in newborn period or infancy. In more than 90% of patients with OSD there is a cutaneous abnormality overlying in the lower spine which may be easily detected on a careful clinical examination after birth.[14]

Urodynamic studies (UDSs) are the most sensitive and accurate test for detecting the urinary tract abnormalities. Bladder outlet

obstruction due to detrusor-sphincter dyssynergia (DSD) is a major contributor to the development of urinary tract deterioration in patients with myelodysplasia; bladder tonicity plays an important role in this regard, especially when outlet resistance exceeds 40 cmH2O.[15] Within the first three years of life, 71% of newborns with DSD had urinary tract deterioration on initial assessment or subsequent studies; CIC alone or in combination with anticholinergic agents, when detrusor filling pressures exceed 40cmH2O and voiding pressures are higher than 80 to 100 cmH2O, resulted in an incidence of urinary tract deterioration of only 8% to 10%.[16,17] As a result, urodynamic evaluation in the neonatal period is now recommended at most pediatric centers in the United States. Ideally it would be best to perform UDS immediately after birth, but the risk of spinal infection and the exigency for closure have not made this a viable option. Therefore, renal sonography and measurement of residual urine are performed as early as possible after birth; UDS are delayed until it is safe to transport the child to the urodynamic suite. If the infant cannot empty the bladder after spontaneous void or with a crede' maneuver, CIC is begun even before UDS are conducted. Other tests that should be performed in the neonatal period include a urine analysis and culture, serum creatinine, and a careful neurological examination.[5] In our study, UDSs performed in 17% of patients (16 patients) for evaluating the lower urinary tract function. Furthermore, 47.9% of patient received medical treatment and the mean age of the patient at the time of medical intervention was 4.8 years (7 children received an appropriate management at infancy). As a result, high prevalence of urinary complications was predictable in these patients which mostly could be prevented with appropriate management.

Vesicoureteral reflux occurs in 3 – 5% of newborns with spinal dysraphism, usually in association with detrusor hypertonicity or DSD[18]; if left untreated, the incidence of VUR in these infants at risk increases with time until 30 - 40% are afflicted by five years of age.[19] Prophylactic treatment that lowers detrusor filling and voiding pressures with oxybutynin and empties the bladder by means of CIC significantly reduces this rising incidence of reflux.[16] In our study, VUR was present in 35.1% of patients.

In our study, urinary incontinence was present in 61 patients (77%) aged four years or older. Of these 19 cases received medical treatment, in nine patients it was unsuccessful and these patients underwent surgical procedure. Cystoplasty was performed in 16 patients without history of previous medical treatment. According to the literature, urinary incontinence in patients with myelpdysplasia can result from total or partial denervation of the sphincter, bladder hyperreflexia, poor bladder compliance, chronic urinary retention, or a combination of these factors.[3] Initial attemps at achieving continence include CIC and drug therapy (anticholinergic agents or á-sympathomimetic agents as indicated) designed to maintain low intravesical pressures and a reasonable level of urethral resistance. Surgery becomes a viable option when drug therapy fails to achieve continence; these include augmentation cystoplasty, bladder neck reconstruction methods, using an artificial sphincter, endoscopic injection of bovine collagen around the bladder neck or creating a continent catheterizable stoma. Initiation of medical treatment according to urodynamic findings in neonatal period and infancy prevents irreversible detrusor muscle destruction and preserves capacity and compliance of bladder. These will significantly reduce the incidence of urinary incontinence and the need of surgical interventions.[5]

The identification and treatment of secondary causes of VUR brings about its spontaneous resolution, unless the ureterovesical junction has been irreparably damaged. In addition, failure to do so significantly jeopardizes any surgery that might inadvertently be done in an attempt to correct the deformity.[5] In our study, of the seven patients who underwent urinary reflux surgery without treatment of underlying causes, VUR relapsed in six cases

(85.7%). These six patients underwent cystoplasty (for removal of underlying cause) and revision anti-reflux surgery. Anti-reflux surgery was performed for the seventh patient alone in his last admission, so we could not evaluate VUR in this case. (Fig-2)

It has been documented that the neurologic lesion in myelodysplasia is a dynamic disease process in which changes take place throughout childhood.[20] Sequential urodynamic testing on a yearly basis beginning in the newborn period and continuing until the child is five years old provides a means of carefully monitoring these children. In addition, urinary tract ultrasonography should be performed annually until the age of seven years. In the presence of recurrent urinary infections, hydroureteronephrosis or DSD, cystography is necessary for evaluation of VUR.[5]

We conclude that the prevalence of urinary tract complications among patients with spinal dysraphism is high. The common complications include urinary tract infections, obstructive uropathy with vesicoureteral reflux, urinary incontinence and even renal failure. With the high prevalence of such complications, careful and periodic follow up is important in the treatment of these patients which would significantly improve the quality of life of these patients.

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