

# The spectrum of urological disease in patients with spina bifida

RA Cahill, EA Kiely

Department of Urology, Cork University Hospital, Ireland

## Abstract

**Background** Urological complications are the major cause of ill health during childhood and adult life of patients with spina bifida but the significance of urinary tract disease on the individual and the healthcare services is underemphasised.

**Aim** To assess the effects of spina bifida on the individual and the healthcare services.

**Methods** A retrospective review was performed to assess the frequency and significance of urological conditions requiring hospital attendance in patients with spina bifida currently attending a specialised multidisciplinary clinic over a period of six months.

**Results** Urinary sepsis accounted for the majority of admissions (62%), while 38 of 62 patients required 60 surgical procedures. Targeting the primary urological abnormality (the dysfunctional and usually poorly compliant bladder) allows implementation of effective treatments, including regular intermittent bladder catheterisation (52%) in order to preserve upper renal tract function. Associated postural abnormalities complicated both conservative and interventional therapies.

**Conclusion** This study highlights the surgical commitment for units caring for patients with spina bifida, the important considerations for the future healthcare services, and the range and severity of urological diseases encountered by these patients.

## Introduction

Spina bifida, or myelomeningocele, occurs when there is failure of fusion of the posterior aspect of the vertebral column. Its significance lies mainly in the associated spinal cord deformity, which may result in disturbance of neurological function to a greater or lesser degree. Following the perinatal period when neuropathology is the greatest cause of morbidity and mortality, the secondary problems in the urinary tract (including recurrent sepsis, stone formation and renal impairment) become one of the most frequent causes of ill health.<sup>1,2</sup> While recognition of the importance of addressing poor bladder compliance early can defer the problems of chronic renal failure until adulthood in the majority of recently born patients,<sup>3</sup> at present, over 50% of the population with spina bifida demonstrate signs of significant upper urinary tract dysfunction.<sup>4</sup> The particularly high incidence of neural tube defects in this country,<sup>5</sup> along with the increasing life expectancy of these patients,<sup>6</sup> means that these individuals require considerable medical input throughout their lives.<sup>4</sup>

This study describes a snapshot view of patients attending a multidisciplinary spina bifida clinic over a period of six months and review their morbidity in order to determine the incidence, significance and spectrum of urological disease in this group.

## Patients and methods

This study is a review of the urological complications of 62 patients who are currently attending a combined medical/urological clinic for individuals with spina bifida over a six-month period. This clinic has been in existence prior to the opening of the hospital in 1978 — it was held at St Finbarr's Hospital prior to this. Any patient with documented spina bifida (including mild forms) can access this clinic on referral from

either their family practitioners or any other specialist involved in their care. Many patients are life-long attendees, while others are referred later in childhood.

Complete and accurate data on all aspects of their clinical care were available for these 62 patients, while an additional 18 patients were excluded from the review due to incomplete documentation. In addition to medical notes, the resources utilised to provide the information for this study include the Hospital Inpatient Enquiry (HIPE) system and theatre logbooks as well as radiological records and databases.

## Results

Of the 62 patients, 33 were female and 45 were wheelchair bound. The spinal abnormality predominantly involved the lower vertebral segments (see Table 1 and Figure 1). The mean age of specialist urological assessment was 3.3 years (range 1 month to 10 years), with their care being managed by a dedicated paediatrician prior to this. The mean age of the patients is now 19 years (range 20 months to 58 years).

**Table 1. Level of spinal column defect in the 50 patients**

| Level of lesion     | No |
|---------------------|----|
| Cervicothoracic     | 1  |
| Thoracic            | 4  |
| Thoracolumbar       | 13 |
| Thoracolumbrosacral | 4  |
| Lumbar              | 22 |
| Lumbrosacral        | 3  |
| Sacral              | 3  |



Figure 1. Plain radiograph of the torso of a patient with spina bifida showing failure of fusion of the sacral vertebrae.

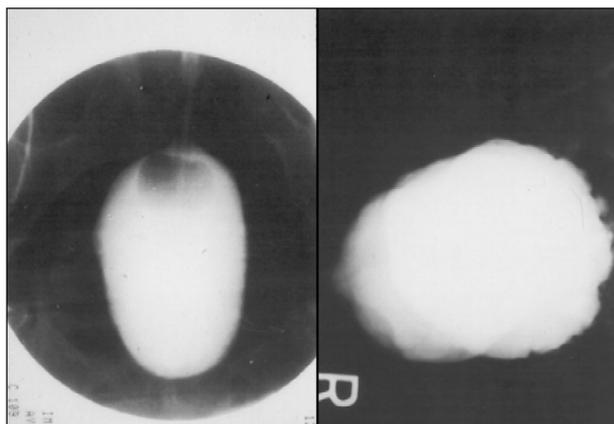


Figure 2 (a). MCUG showing a small, contracted hyperreflexic bladder. (b) MCUG showing a hypocompliant, areflexic bladder.



Figure 3. MCUG demonstrating gross vesico-ureteric reflux (VUR).

### Congenital abnormalities

These patients had a higher than usual incidence of congenital urological abnormality (23% in this series). There were four cases of phimosis, three of bilaterally cryptorchidism and three of inguinal herniation (bilaterally in one individual). Four had renal malformations (two renal dysgenesis, one agenesis and one duplication).

### Hospital admissions

In an eight-year period since HIPE was introduced to this hospital (between January 1991 and December 1998, allowing accurate determination of the frequency and length of inpatient stays) there were 15<sup>1</sup> hospital admissions for urological reasons. The mean length of stay was 7.2 days (range 1-32). Urinary tract infections were the most common reason for hospitalisation (62%), resulting in 35 patients being admitted 97 times and accounting for 542 bed days.

### Urodynamic studies

Twenty-seven patients had urodynamic studies performed to assess bladder function. Seventeen patients were found to have high pressure, hypocompliant bladders (see Figure 2a), with nine also having detrusor hyperreflexia. Ten had areflexic bladders (see Figure 2b). In patients with hydronephrosis who had urodynamic studies performed (n=18), 16 demonstrated elevated bladder pressures (as indicated by bladder leak-point pressures >40mmHg) and hyperreflexia (four having associated detrusor instability), one had areflexia, while another had a normal study.

### Vesico-ureteric reflux

Twenty-nine children had micturating cysto-urethrograms. Vesico-ureteric reflux (VUR) was demonstrated in 14 children, in five of whom it was present bilaterally (see Figure 3). Eight of the patients with reflux had urodynamic studies performed, which revealed high bladder pressures in seven. In addition, five of the seven had definite detrusor hyperreflexia. Seven patients with reflux underwent a submucosal teflon injection (STING) procedure during the early years of this review. This eliminated the reflux in one patient (clean intermittent catheterisation [CIC] was also instituted for this individual), but failed to correct reflux in the other six, although four of these

had short-term success. This latter group subsequently underwent urodynamic studies and in each case a poorly compliant, high pressure bladder was revealed.

### Clean intermittent catheterisation

In total, 32 patients (52%) use CIC (15 self-catheterise) either to assist bladder emptying and/or to achieve continence. The mean age of commencing this therapy was 10.5 years (range 3 months to 36 years) while the fact that those requiring assistance (predominantly in 15 cases by their mothers) had a mean age of 8.2 years (range 12 months to 16 years) indicates the degree of dependency of some of these patients. Of the 18 patients with hydronephrosis, 10 showed radiological improvement (see Figure 4a, b), while six remained stable and two deteriorated despite CIC. In those using CIC to manage their incontinence, five also required anticholinergic medication.

Additionally, two patients underwent a bladder neck incision. Five patients suffered complications from the repeated instrumentation of their urethrae (three developed urethral strictures, one developed epididymo-orchitis requiring admission, while in another a urethral tear occurred). However, similar problems also developed in two patients who had never undertaken CIC (one patient required an optical urethrotomy and subsequent dilatation to treat an urethral stricture while another developed a significant scrotal infection).

### Stone disease

Six patients developed urological calculi at a mean age of 24 years (range 8-30). Four of these were staghorn stones (see Figure 5), the other two had bilateral calyceal stones. Extracorporeal shock wave lithotripsy (ESWL) was performed in four patients, but was only successful in one. Three ultimately required nephrectomy for this benign condition.

### Postural difficulties

Spinal deformity (see Figure 6) prevented three patients from successfully performing CIC (in one case this was due to the back brace). ESWL was rendered impossible for two cases, while in one patient a satisfactory renal ultrasound was unobtainable. Prior spinal surgery (done via an anterior approach) caused intraoperative difficulties in one case.



Figure 4 (a). Intravenous pyelogram (IVP) showing marked hydronephrosis (left>right). (b) IVP in the same patient as (a) showing improvement in radiological appearance after institution of regular clean intermittent catheterisation (CIC).



Figure 5. Plain radiograph showing a left staghorn calculus.



Figure 6. Plain radiograph showing severe scoliosis in a patient with spina bifida.

### Surgical procedures

In all, 38 patients have required 60 urological operations (see Table 2). Five suffered post operative complications (four developed lower respiratory tract infections, while one had a wound infection). Ten patients had a permanent urinary diversion performed; six with an ileal conduit and four by ureterostomy. In each case, however, renal function continued to deteriorate following surgery. The eight patients who have had bladder augmentation surgery (seven ileocystoplasties, one ureterocystoplasty) have been followed-up for a mean of two years (range 1-3). Three out of five patients with incontinence showed improvement (one transiently), while the degree of hydronephrosis (in seven patients) decreased dramatically in two patients, remained unchanged in three and progressed in two.

### Discussion

While the various individual urological maladies suffered by patients with spina bifida are well known,<sup>7</sup> their combined significance to both this group and to the medical profession is infrequently reported. The considerable demand on the health service in general is magnified in countries like Ireland which have a relatively high incidence of myelomeningocele<sup>8</sup> — up to 6.5 per 1,000 births versus two per 1,000 births worldwide.<sup>5</sup> The introduction of folate supplementation may reduce these figures by 50%,<sup>9,10</sup> but this has yet to be effectively implemented.<sup>11,12</sup> The pressure exerted on the urological speciality in particular is compounded by this country having the lowest ratio of consultant urologist to population of all the European Board of Urology affiliated countries.

While centralisation of paediatric surgical services as is currently being considered<sup>14</sup> may have benefits in certain areas, further reducing the availability of specialist centres would have a large impact on this dependent group. Their frequency of hospital attendance from an early age means that local access is of great importance both to the patients and to their families. Furthermore, the quality of care available to the former paediatric patient presenting to the urologist in adulthood will be diminished by the obvious gap in training and experience should the major paediatric units monopolise this area.

The high level of interrelated co-morbidities associated with spina bifida makes the provision of multidisciplinary clinics desirable in caring for those with this condition. Our clinic is held once-monthly and attended by a dedicated paediatrician, a

neurosurgeon and an orthopaedic surgeon, as well as an urologist. Furthermore, there is easy access to an interested plastic surgeon. Such a facility allows the patients to 'multi-consult' the various specialities at a single visit and so simplify their hospital involvement. In addition, it facilitates interdisciplinary communication and co-ordination and, thereby, can accelerate clinical management decisions and actions. Despite the value of early and frequent specialist urological assessment and care, the late age of referral to a specialist urologist (mean 3.3 years) reflects the fact that, especially in patients with near normal mobility — lesions at the sacral level may spare lower limb neural function, bladder dysfunction may not be apparent before toilet training is attempted.<sup>16</sup>

The patients described in our review have attended over a lengthy time scale and hence their care may not reflect best modern practice. Indeed, their experiences highlight how our improved understanding of the relevant pathologies associated with spina bifida have allowed more effective intervention. Bladder dysfunction is now known to have a central role in the urological morbidity and, in particular, the renal dysfunction associated with this condition.<sup>17,18</sup> As well as increasing the risk of urinary sepsis due to incomplete bladder evacuation, detrusor hyperreflexia and dyssynergia with sphincter activity (as reflected by urodynamically-measured high detrusor leak point pressures [DLPP]) precipitates upper tract deterioration due to the impeded ureteric delivery of urine consequent on high intravesical pressure and leads to the direct transmission of this pressure to the renal parenchyma via the standing column of intraureteric fluid.<sup>19</sup> This results in reduced glomerular filtration and renal clearance mechanisms<sup>20,21</sup> and seems most likely to occur when the DLPP (the pressure required to induce involuntary urinary leakage) exceeds 40cm of water.<sup>22</sup> The associated phenomenon of VUR is now realised to be only of secondary importance and, indeed, it usually resolves only when the associated high intravesical pressure has been corrected.

In line with the above principles, there is an increased emphasis on the need for establishing low bladder pressures through regular, adequate emptying by means such as anticholinergic medications and CIC<sup>23</sup> and/or by increasing the bladder capacity (by operative augmentation).<sup>24,25</sup> Although there may be complications with its use, particularly after

**Table 2. Surgical procedures performed in 38 patients**

| Endoscopic             | 17 |                                  |   | Open                        | 43 |
|------------------------|----|----------------------------------|---|-----------------------------|----|
| STING                  | 7  | Orchidopexy                      | 5 | Bladder neck reconstruction | 1  |
| Bladder neck incisions | 2  | Orchidectomy                     | 1 | Bladder augmentation        | 8  |
| Urethral dilatations   | 2  | Inguinal herniotomy              | 6 | (Ileocystoplasty)           | 7) |
| Optical urethrotomies  | 5  | Circumcision                     | 4 | (Ureterocystoplasty)        | 1) |
| Bladder stone removal  | 1  | Urethro-cutaneous fistula repair | 1 | Urinary diversions          | 10 |
|                        |    |                                  |   | (Ileal conduit)             | 6) |
|                        |    |                                  |   | (Ureterostomy)              | 4) |
|                        |    | Ureteric re-implantation         | 1 | Nephrectomy                 | 6  |

prolonged usage,<sup>26</sup> patients persist with and benefit (both subjectively and objectively) from CIC.<sup>27</sup> There is a recent trend toward the prophylactic use of CIC from the early neonatal period in patients with spina bifida<sup>28</sup> rather than waiting until urological problems have become apparent, a time when the older child may be resistant to its introduction.

Permanent urinary diversion is now only rarely performed in patients with myelomeningocele due to its limited benefits (the 'diverted patients' in our study were from the early part of the review).<sup>29</sup> However, the realisation that during voiding, detrusor pressure is more a reflection of urethral resistance than bladder contraction per se has focussed attention on therapies such as anticholinergic therapy (in particular oxybutynin chloride),<sup>30</sup> as well as urethrotomy and external sphincter dilatation,<sup>31</sup> in order to reduce bladder outlet resistance.<sup>32</sup> The former medications act by inhibiting the involuntary bladder contractions and increase the intravesical volume at which such contractions occur, however, whether taken orally or intravesically, they can result in side-effects such as dry mouth, constipation and cognitive impairment (although the latter route is better tolerated).<sup>33,34</sup>

Early use of diagnostic modalities such as ultrasound, nuclear renography and urodynamics can identify abnormalities at a subclinical stage in those with apparently normal voiding habits and neurological examination<sup>35,36</sup> and can predict future urinary tract dysfunction and deterioration.<sup>37</sup> It was only in the latter part of our series in which we have had the benefit of urodynamics and we believe this should be performed initially early in life in all myelomeningocele patients and then repeated when clinical deterioration occurs or when bladder reconstruction is being considered.<sup>38,39</sup>

The ability to accurately measure and follow upper tract function is often limited in patients with spina bifida as associated kyphoscoliosis, constipation and the presence of spinal rods can markedly reduce the ability of ultrasound and intravenous pyelography to assess renal status. While radionuclide (DMSA) scanning has been considered to be the investigation of choice, recently much interest has been generated by the potential of magnetic resonance urography (MRU) imaging as a non-invasive means of multiplanar imaging that avoids the use of ionising radiation. While it appears comparable to DMSA scanning in the diagnosis of renal scarring<sup>40</sup> and can give additional information about complex and congenital abnormal systems as well as the condition of the renal cortex, pelvicalyces and ureters,<sup>41</sup> entry into the magnet may be prevented in up to 50% of patients by their spinal deformity as well as an unwillingness to tolerate the claustrophobia induced inside the scanner in those with mental retardation. The development of open MR imaging machines may obviate these problems. While the usefulness of helical

computerised tomography has not been studied specifically in myelomeningocele patients, it has a more open gantry and is more rapidly performed than MRU and therefore should be considered for the evaluation of individuals without metallic rods. Standard transverse CT images may be less useful than the coronal slices obtained by excretory urography and MRU in the evaluation of congenitally anomalous and operatively altered urinary systems. MRU is superior in determining the condition of the renal parenchyma.<sup>42</sup>

Other noteworthy findings of our study include the clear illustration of the heavy predisposition of this patient group to early stone formation, precipitated by the combination of urinary sepsis and stasis. This may jeopardise renal function, all the more so because first line treatment strategies are often unsuccessful. Associated postural deformity can hinder diagnostic and therapeutic measures as well as complicating these young patients' recovery from surgery. The increased incidence of congenital urological abnormalities in this series has been described before in similar proportions.<sup>43,44</sup>

In summary, urinary tract disease is a common and continuing cause of significant morbidity in patients with spina bifida, and may occur to an advanced degree at an early age. Urological assessment and early institution of appropriate management is therefore essential to maximise these patients' well-being, but it entails a major surgical commitment from units entrusted with the care of these patients, and, therefore, carries considerable implications for the planning and provision of healthcare resources.

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Correspondence to: Dr Ronan Cahill; email: cahillronan@hotmail.com