A ctive and ongoing surveillance for urological problems helps to minimise the impact of the major source of mortality and morbidity in spina bifida.

Key issues for clinicians
- Urological complications of spina bifida are a major source of morbidity and mortality.
- An understanding of the neurogenic bladder is central to the management of urological complications of spina bifida.
- Regular annual urological review helps prevent long term complications, especially renal failure.
- Recurrent urinary tract infections are a major source of long term morbidity and complications. Urinary tract infections demand close investigation and often require specialist follow up. Patients need to be educated for the early detection of urinary tract infections.

Understanding urological complications — the neurogenic bladder

Effective control of urinary incontinence in spina bifida needs to acknowledge the special management issues related to the presence of abnormal neurological bladder function, specifically those related to the neurogenic bladder.

Effective bladder training depends upon the ability to sense the presence of urine in the bladder and the passage of urine through the urethra. A lowered bladder sensation can cause decreased, altered or absent sensation, thereby interfering with effective continence control.

Urinary tract sensation may be decreased, and easily not noticed when the person is otherwise occupied such as while working, watching television or at other tasks. An abnormal anatomical distribution of the nerves may cause sensations to arise from inappropriate places. Absent sensation makes responding to a full bladder impossible, requiring other strategies to achieve continence control.

Faeces or flatus in the rectum can also alter bladder feeling, adding further confusion to sensation, which is important for continence control.

Neurogenic bladder and sphincter abnormal function patterns — the role of urodynamic studies

Not only is an understanding of bladder structure critical to successful continence control, but equally important is bladder and sphincter function. This is one of the main functions of urodynamic studies of the bladder. Fluoroscopic urodynamic studies help the urologist to:
- image the structure and function of the bladder and sphincter
- provide a prognosis for upper tract deterioration
- maintain surveillance for those at high risk of complications
Neurogenic bladder functional abnormalities

Neurogenic bladder functional abnormalities can be classified into three main types:
- hyper-reflexic bladders when the detrusor muscle is unstable or overactive
- areflexic when the bladder is lacking any muscle tone
- mixed picture where there are elements of the hyper-reflexic and areflexic patterns in the one bladder.

There are cases of normal bladder function in spina bifida, but this should only be determined after urodynamic studies in the symptomatic patient.

Sphincter functional abnormalities

Sphincter function can be classified as:
- nonfunctional when the sphincter does not work at all
- synergic when the sphincter control is coordinated with bladder emptying
- dysynergic when the bladder emptying is not coordinated with the sphincter relaxation. When the full bladder starts to contract, the sphincter remains closed.

Bladder function in spina bifida

The commonest neurogenic pattern in spina bifida is the areflexic bladder with a nonfunctional sphincter. This can lead to complications of the upper renal tract and can be a major source of morbidity and mortality. This often results in a high bladder pressure due to urinary retention. High bladder pressure can result in long term urinary sphincter damage.

The commonest urodynamic pattern in spina bifida is the areflexic bladder with a nonfunctional sphincter.

Renal failure

Renal failure is an endstage result of these complications that surveillance aims to prevent.

Aims of urological management

Urological complications are a major source of mortality and morbidity in spina bifida and their prevention and management is a large component of patient care. The main aims of urological management are to:
- preserve upper tract function
- restore low pressure storage
- ensure adequate emptying
- control continence
- minimise UTIs.

Managing UTIs and related urinary tract disorders — a critical issue

Urinary tract infections are common in the presence of abnormal urinary tract structure and function. Recurrent UTIs can seriously compromise renal function and cause permanent renal damage. Furthermore, they can be difficult to detect in the presence of abnormal sensation.

The need for referral

Recurrent UTIs are common in spina bifida and are a strong indication for referral. As hydronephrosis and hydroureter are common, those with recurrent urinary tract infections, or a person with spina bifida not receiving ongoing urological surveillance who develops a urinary tract infection, should be referred to a urologist.

Educating patients to increase their awareness of symptoms and signs of UTIs

While some patients with spina bifida will present with the classic symptoms of UTIs such as frequency, urgency and dysuria, all of these symptoms may be difficult to detect due to reduced or changed sensation resulting from decreased innervation secondary to the spina bifida lesion.

Altered sensation causes atypical presentation

A typical presentations of UTIs may include any (or none) of the following symptoms:
- smelly, offensive urine odour; often like ‘old fish’
- cloudy or bloody urine
- dysuria, although pain sensation may be altered by innervation
- abdominal or loin pain
- fevers
- nausea
Chapter 4: Urological management of spina bifida (including management of urinary tract infections)

- anorexia
- vomiting
- headache
- confusion
- malaise.

Clinicians should also note that UTIs often develop in the presence of constipation.

Educating patients to detect UTIs early can improve quality of life

Late or delayed presentations of UTIs can result in severe complications — including renal failure — and patients need to be educated to be aware of the signs and symptoms of UTIs.

Management of urinary tract infections

Clinicians should have a low threshold of commencing antibiotics when treating demonstrated or suspected urinary tract infections in spina bifida.

Routine antibiotic sensitivity tests ensure appropriate treatment.

Some patients may have had considerable or ongoing exposure to multiple antibiotics over time and, combined with the risks of repeated instrumentation such as during catheterisation, the risk of resistant organisms is high. For this reason, midstream urine (MSU) tests should always include sensitivity testing as well as culture to ensure appropriate antibiotic testing.

In view of reduced sensation, repeat MSUs are necessary to confirm that the infection has resolved.

Review of catheterisation techniques

Many patients presenting with urinary tract infection will be self-catheterising to empty their bladders (see Chapter 5 Controlling urinary incontinence). Many of these patients will have reusable catheters and should be advised to use single-use only catheters until the infection has resolved. Catheterisation techniques should always be reviewed after a urinary tract infection to ensure that the process is clean (see clean intermittent catheterisation in Chapter 5). A discussion from specialist clinics may be appropriate at this point.

Urinary tract infections — should I refer this patient on?

The appearance of recurrent UTIs in someone with spina bifida needs to be taken very seriously, as treatment of any underlying abnormality may prevent the development of renal failure.

An MSU culture that demonstrates a low number of mixed organisms is likely to be due to contamination. In the absence of symptoms and presence of otherwise normal symptoms, signs and investigations, this is unlikely to represent a UTI. These patients may not necessarily require referral, but if any doubts exist, clinicians should feel free to refer patients for further assessment.

Case study: Learning to read the signs of UTIs

Michael is a 25 year old man with spina bifida who presents with recurrent septic shock secondary to UTIs requiring multiple hospital admissions. Due to his spina bifida lesion, he has reduced pelvic sensation relating to typical presentations of UTIs. For him, the early signs of UTI included cloudy, smelly urine, fever and tiredness. Management included referral to a specialist facility for urodynamic review, increasing oral intake of fluids and reviewing catheterisation techniques.

In addition, Michael was taught to present to his GP for dipstick and MSU testing if any of his characteristic symptoms of a UTI were present. Treatment is now started at an earlier stage and the number of his UTI related hospital admissions have decreased from 10 per year to three per year.

Routine urological assessment of spina bifida

As the complications of spina bifida are an ongoing process, regular monitoring of the urinary tract is necessary, especially in high risk patients, in order to prevent and treat potential urological complications. Urodynamic and renal tests are especially critical when determining the optimal time for surgery and prevention of complications.

Lifelong surveillance — the role of the GP

General practitioners may see patients with spina bifida who have not had regular urological monitoring or assessment. When reviewing patients with spina bifida, GPs need to ensure that urological monitoring appropriate for the age group has taken place. Clinicians can order basic monitoring tests for patients who are currently not receiving urological surveillance, followed by referral to a urological or spina bifida centre.

Urological monitoring — the basics

As part of routine medical care, GPs and other clinicians need to:

- assess current and past urological history
- review past renal investigations
• ensure that annual creatinine, urine microscopy and culture and renal ultrasound are ordered.

**Key urological issues for each age**

**Assessment of newborn**

After the closure of the spina bifida defect, the initial evaluation of the newborn urinary tract involves renal ultrasound, residual urine measure, voiding cystourethrogram and urodynamic studies. This is performed within the context of a specialised paediatric unit.

**Children below five years of age**

Children below the age of five years are at the highest incidence of renal damage. In addition to renal function monitoring, these children often require annual or biannual urodynamic studies.

**School age**

Social issues relating to incontinence are critical in school age children as they can interfere with educational opportunities, self esteem and social development. These children may require urodynamic investigations to assist with continence control.

**Young people**

The teenage and young adult years are often times of poor compliance, especially with continence regimens. There may be many psychological, educational and social issues (see Chapter 2 Impact of hydrocephalus and other CNS conditions on care management). Additional urodynamic studies may be required to sort out continence issues.

**Adulthood**

Urological surveillance does not stop just because a person with spina bifida reaches adulthood. Patients require baseline renal ultrasound, renal function tests, annual nuclear scans for measuring glomerular filtration rate and other investigations to assist diagnostic interpretation of complications in the event of change in symptoms.

**Bladder and urinary tract management**

Common approaches to the management of bladder and other urinary tract problems in spina bifida involve a combination of the following:

• conservative management including pharmacological agents
• surgical intervention.

The procedures are described within the context of urinary continence management in Chapter 5 Controlling urinary incontinence.

**Urological surgery in spina bifida**

Urological surgery for the management of incontinence is an important option for people with spina bifida where other procedures, such as intermittent catheterisation, are not feasible. In addition, there are many urological reasons for surgical interventions, all of which impact upon the control of incontinence.

The decision to proceed with surgical intervention for the control of urinary incontinence in spina bifida is a highly complex area. Indications for surgical intervention of the urinary tract in spina bifida include persistent high urinary storage pressure, upper urinary tract deterioration, vesicoureteric reflux and incontinence.

For an overview of the indications and techniques for each of these procedures, see Chapter 5 Controlling urinary incontinence.

**References**