The British Association of Urological Surgeons

Spina Bifida

Physiology

Spina Bifida is a collective term used to describe a group of patients who have a congenital failure of closure of the spinal column and meninges. These abnormalities are also known as Neural Tube Defects and include Spina Bifida Occulta, Menigocele and Myelomenigocele.

The importance of these conditions to Urologists is that they commonly cause Neuropathic bladder dysfunction and can result, if poorly managed or unrecognised, renal failure and/or incontinence. The urodynamic effects of effects of Spina Bifida often include poor bladder compliance and a non-relaxing sphincter resulting in a high risk to the upper tract.

Spina Bifida is the result of a combination of genetic and environmental factors. Deficiency of Folate in early pregnancy and /or an inability to utilise this due to genetic defects are well known factors. Less well known are certain anti seizure medications (Sodium Valproate, Catbamazepine), poorly managed maternal diabetes and maternal obesity.

Paediatric management has also improved with close surveillance of the kidneys, frequent urodynamic examination, early intermittent catheterisation and the use of anticholinergics and Botox to control bladder pressures all protecting the kidneys.

More recently in utero closure of the spinal defect has been practiced and appears to improve mobility and hydrocephalus rates, although clear data on bladder outcomes is yet to emerge.

Management

Patients with spina bifida should be under lifetime urological surveillance under specialist care which should include holistic care for bowel and sexual dysfunction management (NICE 2012)

Occasionally patients will present with completely untreated Spina Bifida or problems related to a neuropathic bladder secondary to previously unrecognized Spina Bifida Occulta.

The aims of management remain the same at whatever age or stage of management the patient is:

1. Maintain and protect the renal function
2. Achieve and maintain urinary continence
3. Promote independence

The key to safe and effective bladder management is achieving good high volume low pressure urine storage combined with efficient and complete and regular bladder emptying. This can often be achieved by urethral self-catheterisation plus oral medication or Botox injections.

However if this is not effective – either in protecting the kidneys or achieving continence – then augmentation cystoplasty +/- anti incontinence surgery (e.g. rectus fascial sling in females or Artificial Urinary Sphincter in males) +/- creation of a Mitrofanoff continent catheterisable stoma may be required.

Alternatives may include an ileal conduit or a suprapubic catheter which may be more appropriate to the individual patients' abilities and circumstances.

Once safe and effective bladder management has been established there remains an absolute requirement for on-going follow up and surveillance.

Special Considerations

As a Urologist one is generally asked to take over the care if these patients as they transition into adulthood. This can be challenging time and is greatly helped by robust paediatric to adult transition arrangements including shared clinics.

In addition to the neuropathic bladder and bowel dysfunction these patients will often have hydrocephalus requiring drainage via a ventriculoperitoneal shunt and spinal deformity due to scoliosis. These will require care from expert neurosurgeons and / or spinal surgeons and can impact considerably on the Urological management.

With aging patients with Spina Bifida often present with more problems. They may become more obese and their scoliosis may deteriorate making managing their intermittent catheterisation or their ileal conduit more difficult. They may develop a para stomal hernia or renal stones.