Neurogenic Bladder and Spina Bifida

Jameela Kari

The incidence of spina bifida worldwide still ranges from 0.3–4.5 per 1,000 births with higher incidence in the developing countries. The incidence of renal damage is nearly 100% in patients with an overactive pelvic floor (detrusor/sphincter dyssynergia; DSD) when not adequately treated.

Renal damage starts early in life, within the first 6 months. However renal function can be preserved provided that treatment starts early with adherence to the treatment regime,

Dryness for urine could be achieved by the time they go to primary school. To obtain such results, it is mandatory to treat detrusor overactivity from birth onward.

Patients with spina bifida should be treated from birth by clean intermittent catheterization and pharmacological suppression of detrusor overactivity. Urinary tract infections, when present, need aggressive treatment, and in many patients, permanent

prophylaxis is indicated. Later in life, therapy can be tailored to urodynamic findings. Children with paralyzed pelvic floor and hence urinary incontinence could be offered surgery around the age of 5 years to become dry. Rectus abdominis sling suspension of the bladder neck is a good -choice as procedure. In children with detrusor hyperactivity, detrusorectomy can be performed as an alternative for ileocystoplasty provided there is adequate bladder capacity. Wheelchair-bound patients can manage their bladder more easily with a continent catheterizable stoma on top of the bladder. This stoma provides them extra privacy and diminishes parental burden. Bowel management is important and could be achieved by retrograde or antegrade enema therapy.

Management of steroid-resistant nephrotic syndrome (SRNS)

Jameela Kari

Steroid-resistant nephrotic syndrome (SRNS) is defined as not achieving remission after 4 weeks' treatment with daily prednisolone. Renal histology in most patients shows presence of focal segmental glomerulosclerosis, minimal change disease, and rarely mesangioproliferative glomerulonephritis. A considerable percentage of children with SRNS show mutations in one of the key podocyte genes. The younger the age of presentation, the higher possibility that a causing mutation will be found

The remaining cases of SRNS are probably caused by an undefined circulating factor. Treatment with calcineurin inhibitors (ciclosporin and tacrolimus) is the recommended treatment for non-genetic SRNS, and approximately 70% of patients achieve a complete or partial remission with satisfactory long-term outcome. Additional treatment with drugs that inhibit the renin-angiotensin axis is recommended for hypertension and for reducing remaining proteinuria.

SRNS is the second commonest cause of end stage kidney disease (ESKD) in children as patients who do not respond to treatment and do not achieve remission will show declining kidney function and are at risk of ESKD.