# Congenital neuropathic bladder

Congenital neuropathic bladder

Neurogenic lower urinary tract dysfunction (NLUTD) refers to the spectrum of bladder dysfunction that can arise from congenital or acquired abnormalities of those parts of the nervous system that are responsible for normal bladder func - tion ( Table 83.3 ). The most common congenital cause of NLUTD is - abnormal development of the spinal canal (neural tube defects). The neural tube develops in early gestation (closure of the spinal canal is complete by day 35) and maternal folic acid deficiency is one of the primar y risk factors for incomplete closure. Spina bifida, the most common neural tube defect, ranges in severity from mild (spina bifida occulta), in which there is only mild separation of the spinal vertebrae but no neurological involvement, to severe (myelomeningocele), in An essay on the shaking palsy in 1817.

- 

Central nervous system tumours In /f_l ammatory/infective conditions of the central nervous system (encephalitis, transverse myelitis) Vascular conditions affecting the central nervous system (infarct, haemorrhage) Spinal cord injury Neurodegenerative and demyelinating diseases (e.g. multiple sclerosis, Parkinson’s disease) Other encephalopathy (e.g. cerebral palsy) Iatrogenic – pelvic/spinal/cerebral surgery Lesions of the peripheral nervous system (e.g. diabetes)

and exposed onto the skin of the lower back. Clinical features The clinical features of myelomeningocele can be variable, depending on which nerves have been everted in the menin gocele sac. Infants will have a visible cutaneous abnormality overlying the lower spine. In cases of spina bifida occulta, any suspicion of lower spinal cutaneous abnormality warrants further investigation with spinal ultrasound or MRI. In some cases, cutaneous lesions may be absent and, as the child grows with increasing age, tethering of the cord (fixation of the lower spinal cord due to scarring from surgery , lipoma or deep skin dimples, leading to stretching of the cord with growth of the child) can lead to the development of symptoms. Therefore, spinal investigation should be considered in any infant present ing with bladder or bowel dysfunction, failure to toilet train or lower extremity weakness, as this may be a sign of occult spinal dysraphism. An associated Arnold–Chiari malformation with hydro cephalus is commonly seen with myelomeningocele, resulting in developmental brain abnormalities. Infants may develop urinary infections, dribbling of urine and incomplete bladder emptying. End-stage renal disease (ESRD) is the commonest cause of death in infants with spina bifida and so ear ly identification, surveillance and treatment of those at risk for ESRD is the cornerstone of management. Investigation Myelomeningocele requires surgical closure of the spinal defect immediately after birth, and so urological investigations are delayed until the patient has recovered from surgery . /uni25CF Renal tract ultrasound and postvoid residual urine mea surement are required. /uni25CF Video urodynamics should be performed as

soon as feasi ble (usually in the first 2–3 months of life) to assess bladder function. /uni25CF In the presence of VUR, a dimercaptosuccinic acid (DMSA) renal scan is recommended at 3 months to pro vide accurate measurement of renal function. /uni25CF In those with risk factors for renal deterioration (hydro nephrosis, elevated postvoid residual, poor compliance, detrusor overactivity and DSD) early treatment should be initiated. All infants should undergo lifelong surveillance, initially with 6-monthly renal tract ultrasound and post void residual, and yearly urodynamics f or the first 2 years to detect any deterioration in renal drainage and bladder function. Treatment Bladder management aims to prevent deterioration in renal function. This is dependent on achieving low-pressure storage and voiding, with complete bladder emptying. Julius Arnold , 1835–1915, Professor of Pathological Anatomy , the University of Heidelberg, Heidelberg, Germany , described this condition in 1894. Hans Chiari , 1851–1916, Austrian, Professor of Pathological Anatomy , Strasbourg, Germany (Strasbourg was returned to France in 1918 after the end of the First World War), gave his account of this condition in 1891. The Arnold–Chiari malformation refers to a structural defect in the cerebellum characterised by ventral herniation of the cerebellar tonsils through the foramen magnum of the skull. findings ( Table 83.4 ). The management of incomplete blad - der emptying, high-pressure storage, detrusor overactivity and high-pressure voiding is centred ar ound clean intermittent self-catheterisation (CISC) in combination with antimuscarinic - therapy to reduce bladder pressure. If this fails , then intraves - ical botulinum toxin A (BTX-A) is injected into the bladder wall. Augmentation enterocystoplasty , in which the bladder is bivalved and enlarged using a segment of ileum, is reserved f or those with ongoing risk factors for renal deterioration despite the above treatments. Patients, or parents, who are unable to perform urethral CISC should be considered for a continent urinary diversion with appendicovesicostomy , in which the appendix is used as a channel to connect the bladder with the skin of the umbilicus through which the patient can perform - self-catheterisation. These management options are discussed in more detail later in this chapter. Disorders that cause NLUTD often also lead to neuro - pathic bowel dysfunction (constipation or faecal incontinence) - and so this should also be addressed in all children presenting with NLUTD. Summary box 83.2 Congenital neuropathic bladder /uni25CF /uni25CF /uni25CF /uni25CF - /uni25CF - /uni25CF /uni25CF - -

The pattern of NLUTD depends on the site, severity and type of neurological lesion The bladder and sphincter may be overactive, normoactive or underactive The combination of an overactive bladder and overactive sphincter represents the highest risk of renal deterioration An overactive bladder is treated with pharmacotherapy, intravesical BTX-A, sacral neuromodulation or augmentation enterocystoplasty An underactive bladder is managed primarily with CISC, or if urethral catheterisation cannot be performed, then appendicovesicostomy An overactive sphincter is primarily treated with intrasphincteric BTX-A or sacral neuromodulation Bladder and bowel dysfunction often coexist and should be addressed together

---

Revision #1
Created 2025-12-31 15:30:07 UTC by Omar Ayman
Updated 2025-12-31 15:30:07 UTC by Omar Ayman