Management of Neural Tube Defects in the Neonatal Period

Pathology:
The neural tube normally closes between 15 and 28 days post-conception: failure of normal closure results in a neural tube defect (NTD). Maternal folate deficiency, maternal anti-epileptic use and poorly controlled maternal diabetes are all associated with a higher incidence of NTDs. Currently in Wales prevalence is 13 per 10,000 births: in a unit such as UHW, we expect to see 7 cases per year.

<table>
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<th>Normal structures</th>
<th>Common anomaly consisting of a midline defect of the vertebral bodies without protrusion of spinal cord or meninges. Most often it is asymptomatic and of no consequence, but it can be associated with developmental abnormalities or tethering of the spinal cord.</th>
<th>Meningoceles are rare. The structure contains meninges and CSF only.</th>
<th>The most common type of neural tube defect, is a sac like structure which protrudes through a defect in the vertebral arches, containing meninges, CSF and spinal cord and/or nerve roots.</th>
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Other less common neural tube defects include lipomyelomeningocele and diastematomyelia.

The degree of neurological impairment depends on the position and extent of the lesion. There will usually be lack of sensation in the corresponding dermatome, complete/full paralysis of the skeletal muscles and often bladder and/or anal sphincter paralysis.

Clinical Presentation and Diagnosis:

Usually antenatally diagnosed:

Alpha fetoprotein (AFP) is a plasma protein produced by the developing fetus. Elevated AFP from amniotic fluid is very reliable unless the defect is closed. Maternal plasma AFP is less reliable, detecting only 50-90% of open anomalies, and is falsely positive in 5%.

Fetal anomaly scanning at 20 weeks: characteristic appearances include lateral displacement of the spinal peduncles, or the myelomeningocele may be visualised. A ‘lemon head’ shape may be seen on fetal anomaly scan.

Postnatally, the following clinical features may be noted:

- Open lesions are obvious and not covered by normal skin.
- Closed lesions may present with a visible abnormality of the back, such as asymmetric buttock fold or dimple, haemangioma, hair patch, or other cutaneous lesions.
- Abnormal neurology in lower limbs / asymmetrical or absent movement or spasticity in the lower limbs.

Immediate management in Labour Ward:

- Refer to Fetal Medicine folder. These patients may have been counselled antenatally.
- Prepare cling-film to cover the lesion i.e. place sheet on resuscitaire, and place baby onto cling film, wrapping around the abdomen. Employ a clean technique.
- Manage the airway, breathing and circulation as any other baby (see NLS guidance).
- If no resuscitation is required following birth, nurse in the lateral or prone position avoiding direct pressure on the lesion.
- Give Vitamin K as normal.

On going management:

- Weight, length and OFC to be measured on admission, and plotted on appropriate growth chart.
- Lesion care: Cling film dressing may be replaced with a non-adhesive dressing, wet with 0.9% Saline. Continue to nurse in side lying or prone position to avoid pressure on the sac or nerves.
- IV access, bloods for FBC, baseline U and E, CRP, group and save.
- IV antibiotics should be given to infants with open lesions i.e. when there is broken skin or evidence of CSF leak. Babies with closed lesions may also require antibiotics if there are risk factors for early neonatal sepsis. Antibiotics as per local protocol. If IV antibiotics are not indicated, commence prophylactic trimethoprim (2mg/kg nocte).
• Fluids and Feeding: Surgical correction usually occurs within 72 hours of birth, and infants will need to be nil by mouth for 6 hours prior to surgical intervention - IV fluids as per neonatal fluids guideline. No contra-indications to feeding if operative intervention is delayed.
• Monitor for evidence of renal impairment: monitor urine output and perform regular U and E as indicated. If renal function deteriorating, consider referral to paediatric nephrology.
• Commence clean intermittent catheterisation (CIC) 3-4 hourly. Refer to CIC Operational Guideline in nursing folder.
• Early, thorough baby check, in particular, record absence or presence of spontaneous movements in lower limbs.
• Orthopaedic abnormalities related to disrupted innervation of muscles i.e. dislocated hips, talipes are common. Refer to physiotherapy if there is evidence of these anomalies at birth. Arrange neonatal hip screening ultrasound scanning for 6 weeks postnatally. There may be an associated spinal deformity – kyphosis or scoliosis. Discuss with orthopaedics and radiology prior to imaging.
• 10% of infants may have an associated chromosomal defect – trisomy 18, triploidy, single gene disorders. Low threshold for genetic investigations if concerns about possible multiple anomalies (aCGH).

Cranial ultrasound scan:
• Cranial ultrasound is performed to look for evidence of ventriculomegaly. There may also be abnormal anatomy, particularly midline defects.
• If large ventricles are present, measure the ventricular index (VI) and plot on VI chart in guideline for Cranial Ultrasound scanning. The VI is measured as the falx to the lateral border of the lateral ventricle, in a coronal view taken in the plane of the third ventricle.
• The Chiari II malformation (herniation of the cerebellar vermis through the foramen Magnum) is present in 95% of children with myelomeningocele. Initial presentation of severe brainstem dysfunction is similar to the presentation of perinatal asphyxia.

Inform neurosurgical registrar on call at UHW and Paediatric Surgical Registrar of birth and admission to the NNU. These patients should be discussed at the Thursday Surgical Neonatal meeting.

Bladder and Bowel Management:
Children with meningomyelocele are likely to have a neuropathic bladder and bowel.
Early involvement of the Paediatric Specialist Continence Nurse is important (44627).

Clean intermittent catheterisation
• Start CIC with each feed (3-4 hourly), document clearly the catheter volumes as part of the bladder diary.
• Clear documentation of fluid input / urine output and passage of meconium/ stool.
• If urine output is poor, assess bladder volume using ultrasound scan: a random check of bladder volume in a newborn should be less than 30-45mls.
• Parents and carers will need to undertake training in CIC, arranged via the Paediatric Specialist Continence Service or taught by NICU staff where appropriate.
• Treat symptomatic bacteriuria according to local guidance. Patients on CIC often have asymptomatic bacteriuria, which does not need to be treated.

Medications
• Commence prophylactic Trimethoprim (2mg/kg nocte) on all patients with myelomeningocele (unless already on IV antibiotics).

Bowels
• Good skin care for the perineum is important - dribbling urine and stools. Topical treatment with Cavalon is recommended.

Imaging
• Arrange ultrasound scan of the renal tract within 48 hours.

Follow Up
• NICU to refer to local Community Paediatrician and Children’s Community Nursing Services for on-going co-ordination of care. NICU to arrange open access with local Children’s Assessment Unit.
• Referral to paediatric neurology specialist nursing team for serial OFC measurement (UHW ext 48268).
• Referral for all infants in South Wales to the Neuropathic Bladder (Y48/Y49) Clinic. To be seen within 4 weeks for discussion of anticholinergics and further imaging.
• Referral by Y48/Y49 clinic for DMSA scanning, and then to Joint Surgical Neuropathic Bladder Clinic (Joint 45) within 4 months.
• Urodynamics at 6 months to be arranged by Paediatric Continence Nurse Specialists and subsequent review at Joint 45 clinic.

References:

K Burke and S Barr January 2014, to be updated January 2017