Paediatric Guideline for Management of Neural Tube Defects

Title: Paediatric Guideline for Management of Neural Tube Defects			
Date effective from:	17/02/2025	**Review date:**	12/02/2029
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Approval Date:	02/12/2024		
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Target Audience:	Paediatric team, Neurosurgical team, Neonatology team, Nursing team and allied health teams		
Supersedes:	Version 1		
Keywords (min. 5):	Neural tube defects, pre-operative care, post-operative care, allied health input		

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Version Control

Date	Author	Version/Page	Reason for change
17/02/25	Dr Kaylita Chantiluke, Dr Vix Monnelly, Dr Helen Turner	V1	

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NHS Lothian

Contents

1.0 Purpose

This guideline aims to provide healthcare professionals with an understanding of neural tube defects and how to manage patients with myelomeningocele. It focuses on pre-operative and post-operative care as well as providing information on allied health input and discharge planning.

2.0 Scope

This guideline focuses on the management of babies with myelomeningocele.

Consequences of myelomeningocele are variable, but include:

- Motor and sensory loss caudal to lesion – to varying degrees
- Neuropathic bladder and bowel dysfunction
- Orthopaedic abnormalities secondary to disrupted motor innervation
- Chiari Malformation (Type II)
    - Present in majority of patients with myelomeningocele
    - Hindbrain herniation, small posterior fossa; displacement of medulla, lower pons and cerebellar vermis; and elongated 4th ventricle through the foramen magnum
    - Can cause obstructive hydrocephalus
- Hydrocephalus
    - Very common (up to 80%), but not always requiring treatment in the early neonatal period
- Co-existing CNS malformation
- Spinal deformity e.g kyphosis or scoliosis

3.0 Definitions

The term ‘Spina Bifida’ encompasses a group of disorders of spinal cord development which result from failure of the normal closure of the embryonic neural tube. The neural tube forms the brain and spinal cord, and its aberrant closure can lead to a wide range of anomalies.

The incidence of Spina Bifida is 6 per 10,000 in the UK (1), and myelomeningocele accounts for 75% of all cases of Spina Bifida (2). Spina Bifida can occur at any level along the spinal cord but is mostly commonly found in the lumbar and sacral regions. Prognosis depends on the level of the lesion, involvement of neural tissue in the defect, ascending infection and the presence of other structural defects.

A variety of detailed classification systems for Spina Bifida exist, with more recent systems being based upon the presumed point of origin in embryogenesis (3). These classifications are outside the scope of this guideline.

Spina Bifida can be broadly divided into two groups based on its cutaneous presentation – Spina Bifida Aperta and Spina Bifida Occulta:

Spina Bifida Aperta ('open'): This refers to patients who have neural tissue exposed, frequently leaking cerebrospinal fluid (CSF). In Spina Bifida aperta, neural tissue is not covered by skin, but is either completely exposed, leaking CSF, or covered by little more than rudimentary meningeal membranes. This is referred to as either a myelomeningocele or myelocele (3):

Myelocele: The neural placode is flush with the surrounding skin

Myelomeningocele: The neural placode is raised up by an accumulation of CSF in the underlying subarachnoid space

Open neural tube defects are pan central nervous system (CNS) malformations - this means that in addition the spinal cord anomaly, several structural abnormalities of the brain are usually seen. These abnormalities are encompassed by the term 'Chiari II malformation'. Hydrocephalus is also present in approximately 80% of cases (3).

Spina Bifida Occulta ('closed'): This covers all other forms of Spina Bifida, where the neural tissue is not visible but hidden due to intact overlying skin. The skin is frequently abnormal with cutaneous stigmata of Spina Bifida. Spina Bifida occulta is not a pan CNS malformation, therefore Chiari II malformation and hydrocephalus are not associated with it (3).

4.0 Roles and responsibilities

This guideline is applicable to all professionals looking after patients with Spina Bifida.

5.0 Main content

Antenatal Management

The aim of antenatal management is to provide information and choices for parents based on the best available clinical information, and to ensure that all teams likely to be involved in the care of the baby have all relevant details prior to birth

Myelomeningocele maybe detected antenatally at the anomaly scan (18-21wks).

Following antenatal diagnosis, the following steps should be taken prior to birth:

1. Multidisciplinary team (MDT) discussion with the family including counselling around prognosis; discussion around the current treatments available (fetal surgery vs early postnatal surgery). The family may wish to discuss options including termination of pregnancy and in some cases, palliative care arrangements after birth. This is usually undertaken by a neurologist and a neurosurgeon once the diagnosis has been made. The Neonatal consultant on for post-natal should be contacted to support

discussions on page 4133 (dial 110 to page). Fetal Medicine Team takes the lead on referrals for fetal surgery.

2. Antenatal MRI and amniocentesis (required when referral for fetal surgery is considered) may be performed based on clinical need.
3. Encourage the family to contact a Spina Bifida charity such as, Spina Bifida Hydrocephalus Scotland (4). The neonatal team can offer a tour of the neonatal unit if the family wishes.
4. The plan for mode and date of birth will be shared with neonatal and neurosciences teams, if the pregnancy is continued.
5. Neonatal team to ensure the neonatal management plan is updated on TRAK.

Prenatal surgery for open fetal repair

Prenatal surgery with open fetal repair of the spinal lesion has been shown to improve short-term outcomes for the child including rates of ventriculoperitoneal shunting, frequency of hindbrain herniation and an earlier age of independent ambulation, with consequent risks of prematurity and maternal morbidity (5-6).

Referral for Open Fetal Surgery

Surgery is offered and performed according to internationally agreed criteria and protocols. University Hospitals Leuven in Belgium is currently the Fetal Surgery Centre (FSC) that provides open fetal surgery for all patients with open Spina Bifida who live in Scotland. Surgery is ideally performed between 23+0 and 25+6 weeks of gestation (7). Referral criteria for Open Fetal Surgery to treat fetuses with Open Spina Bifida can be found at: https://www.uclh.nhs.uk/our-services/find-service/womens-health-1/maternity-services/your-pregnancy/spina-bifida-open-fetal-surgery

Only after eligibility is established at the FSC will a prenatal operation be offered. Clinicians are encouraged to refer patients from 20 weeks' gestation to the service as soon as the diagnosis of fetal Spina Bifida is made, whilst awaiting the results of genetic tests. Patients who are referred must be less that 26+0 weeks gestation with normal genetic testing and a spinal lesion from T1 to S1 (including myelomeningocele or myeloschisis) with a radiological diagnosis of Chiari type II malformation (6).

Post-operative care following Open Fetal Surgery

Following surgery, patients are expected to return to their referring hospital for antenatal care and delivery. A post-operative protocol and provision of email support by the FSC is available to the units caring for these patients.

Following their birth through elective caesarean section, patients are admitted to the Neonatal Unit (NNU). The recommended neonatal protocol following open fetal surgery for Spina Bifida is shown in appendix 2 (8).

Management at Delivery

Planned postnatal closure

The aim is to protect the lesion while facilitating transition and enabling parental bonding.

Check neonatal management plan on maternal TRAK care record prior to delivery for any individualised management plan.	
Neonatal middle grade doctor/ANNP and nursing team leader to be present at birth.	Inform neurosciences team as soon as possible after delivery.
Provide airway, breathing and circulation management as per neonatal life support (NLS) guidelines if required.	Cardio-respiratory support is not normally required. If required, then attempt airway support and inflation breaths in lateral position in first instance. Supine position only if required for emergency procedures.
Position infant prone or in lateral decubitus position (side lying).	Avoids pressure on the sac/nerves and reduces the risk of trauma to lesion.
Document: 1. **Position and size of lesion** 2. **Any skin covering** 3. **If sac appears intact or ruptured prior to covering** 4. **If able, and parents are consenting, photographs of lesion to be taken prior to covering**	In lesions > 5cm, plastic surgery input is frequently required during surgical closure – the Neurosurgical team will advise if their input is required following their assessment of the patient
Cover exposed neural placode with a 0.9% sodium chloride soaked sterile gauze. Cover gauze with cling film ***(Image 1)***	Covering promptly reduces risk of traumatic damage, infection and prevents desiccation of the neural placode and further neurologic injury to exposed nerves /cord.

Pay close attention to thermoregulation.	Increased risk of hypothermia due to immaturity of thermoregulation system and lesion related evaporative losses.
Encourage parents to hold their baby while protecting the placode. **Support an early attempt at breastfeeding if mother wishes to breastfeed and ensure that she is supported in expressing within the first hour and with ongoing lactation until her baby is ready to breastfeed.**	

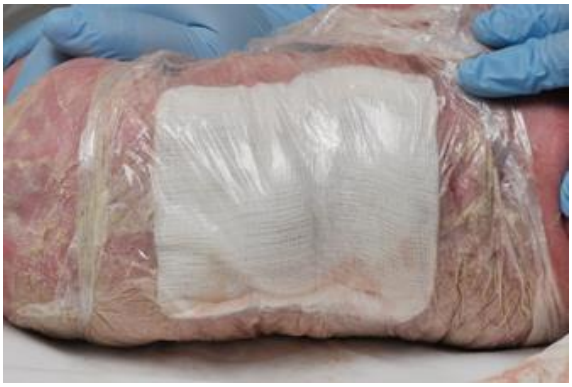


Image 1: Sterile gauze soaked in 0.9% sodium chloride placed over the placode and subsequently covered by cling film.

Management on Admission to Neonatal Unit (NNU)

The aim of management is to protect the lesion while maintaining cardiorespiratory stability and supporting parents.

General Care	**Nurse unclothed in an incubator and maintain normothermia.**	
	Continue to nurse in prone or lateral decubitus position (side lying).	Prone is preferred position, side lying can be used

		intermittently if position change needed.
	Ensure vitamin K is given.	Must be given pre-operatively.
	Obtain IV access and take baseline pre-operative bloods.	**Haematology:** - FBC, coagulation screen, group and save **Biochemistry:** - U&Es, calcium, phosphate, magnesium, glucose
Infection	**In straight forward cases, prophylactic antibiotics may not be required** [9,10] **Use clinical judgement in specific cases: e.g., prematurity, PROM, baby born out with the Simpson Centre for Reproductive Health.** **Please discuss with Neurosurgical team and/Microbiology if in doubt.**	Follow local neonatal antibiotic guidelines
Clinical Assessment	**Initial assessment should include neurological examination and assessment for any associated congenital anomalies.**	Motor evaluation with a record of lower limb muscle strength to indicate functional level of myelomeningocele - this provides a baseline for ongoing surveillance
	Occipitofrontal circumference (OFC) should be measured and plotted daily on TRAK and Badger.	Due to risk of hydrocephalus.
	Monitor for urine output and passage of meconium.	Due to risk of neuropathic bladder and bowel.

		May require urinary catheter due to risk of urinary retention.
Care of lesion	**Keep lesion covered and moist with sterile gauze soaked with sterile saline.**	Ensure gauze remains well soaked. Gauze can either be replaced every 2-3 hours with fresh sterile gauze or can infiltrate with fresh saline every hour, and changed gauze 12 hourly.
Fluids and feeding	**Aim for enteral feeding, unless other pathology or clinical condition means enteral feeding not appropriate; or surgery is not within next 4-6 hours.**	Care should be taken not to disrupt lesion during breastfeeding and bottles given in side-lying position.
Imaging	**Perform cranial ultrasound scan (CrUSS) including measurements of ventricular indices if antenatal ventricular dilatation, or any evidence of cardiovascular or respiratory instability.**	Cardiovascular or respiratory instability can be caused by hydrocephalus. Further imaging will be arranged by neurosciences team.
	Request medical photography of lesion if time allows prior to transfer to Royal Hospital for Children and Young People (RHCYP).	Try to time this to avoid uncovering lesion unnecessarily.
Parents	**Ensure parents are updated with adequate information and explanation regarding the progress of their baby after delivery.**	
Transfer to RHCYP	**This should occur when safe to do so, and in consultation with neurosciences team.**	Safe neonatal care takes priority over urgent transfer.

Pre-Operative Care

The Neurosurgical team is responsible for deciding on optimal timing for operative repair.

- Pre-operative care usually occurs on the paediatric neurosciences ward if the baby is clinically well and weighs more than 3kg.
- If pre-term, low birth weight, or any other concern about general health then pre-operative care may occur in the NNU or the High Dependency Unit (HDU)/Paediatric Intensive Care Unit (PICU) at the RHCYP. This should be a joint discussion involving the neonatal team and the neurosciences team.

If pre-operative care occurs in NNU then care will be shared with the neurosciences team and referral is required to the following teams:

At time of birth	Pre-Operatively	Post-Operatively (<24hrs)	Pre-Discharge	If Relevant
- Neurosurgery - Neuroscience	- Physiotherapy	- Urology	- General surgeons - Orthopaedics	- Plastic surgeons - Clinical genetics

Post-operative Management

If the baby is term, then post-operative care is usually provided at RHCYP, but occasionally a baby may require transfer back to the NNU. Care will be shared with neurosciences and Neurosurgery team, and baby may also have input from other specialties.

Nurse baby prone or in lateral decubitus position (side lying).	Can have short period of supine care after discussion with the Neurosurgical team
Daily nursing wound check.	If leakage, bleeding or signs of infection, please inform Neurosurgical team urgently.
Daily clinical examination including OFC and anterior fontanelle checks, assessing for signs of hydrocephalus	Hydrocephalus requiring treatment develops in >50% of children post-operatively. After the first 3 days post-operatively, if OFC remains stable then this can be done two to three times per week, then weekly based on the clinical progress.

	OFC values should be plotted on the head circumference chart to monitor for change – if concerns, inform Neurosurgery
Daily checks of lower limb function – ensuring no deterioration in movement relative to their post-operative baseline	Rarely, children can develop syringomyelia post-operatively (fluid collecting within their spinal cord) – if concerns, inform Neurosurgery
Regular observations including temperature, heart rate, respiratory rate and blood pressure for the duration of admission.	Specifically focus on signs of infection and fluid balance Bulbar symptoms are rare but can develop in patients with severe Chiari II malformations, signs include apnoeas and bradycardia – if concerns, inform Neurosurgery
Ensure adequate nutrition and hydration	Monitor for vomiting and/or aspiration and feeding difficulties (bulbar symptoms) – if concerns, inform Neurosurgery
Ensure routine newborn screening tests are performed	This includes the newborn physical examination, newborn hearing test and the Guthrie/blood spot card. If the baby is premature, please follow local NNU guidelines for any additional screening tests they may require.
Physiotherapy	Essential to determine both sensory and motor functional levels. They will document deformities and liaise with Community teams.

Check bladder and bowel function	Monitor for constipation.
A post-operative cranial ultrasound must be completed within 72 hours. **MRI for neural axis abnormalities should be completed prior to the patient being discharged home.**	These examinations provide baselines for future comparison. More frequent cranial ultrasounds may be advised by the Neurosurgical team based on their clinical assessments.
Ensure continued parental support and open communication	
Discharge planning and timing should be discussed with the neurosciences team.	

Sub-Specialist and Allied Health Input

Urology

- Indwelling catheter to be inserted at the time of the surgery and to remain in-situ until urology clinical nurse specialist review.
- Email referral to Eleni Papageorgiou and urology clinical nurse specialist Fiona Cropley & Aileen Connor.
- Urology clinical nurse specialist will then assess the neonate and advise on clean intermittent catheterisation and the use of prophylactic trimethoprim.
- Renal tract USS and baseline renal function bloods (urea, electrolytes and creatinine) to be organised by the unit where the baby is admitted (eg NNU/HDU/neurosciences). These tests should be performed on **day 7 of life**.
- Check with urology clinical nurse specialist prior to discharge regarding the results of renal tract USS and future appointments.
- For further information on urology input up until the first year of life please see appendix 1

Orthopaedic

- All babies with Spina Bifida should be referred for secondary neonatal hip screening from the newborn examination. Referral should be sent to NeonatalHipReferrals@nhslothian.scot.nhs.uk
- If there are any concerns regarding a structural deformity of the feet (clubfoot, calcaneovalgus/congenital vertical talus) please refer to the specialist physiotherapy team at loth.rhcypclubfootphysio@nhslothian.scot.nhs.uk

Physiotherapy

- Ensure paediatric neurology physiotherapy team are aware of an admission or if the family have antenatal counselling, we are aware of EDD (as per current usual communications or email: PhysiotherapyNeuroRHCYP@nhslothian.scot.nhs.uk)
- Physiotherapy will strive to complete a pre-op initial assessment (day and stability of patient dependent), wherever the baby is within RIE/RHCYP.
- Detailed post operative assessment from Day 1 post-op focusing on:
    - Establishing the motor level of lesion on clinical examination
    - Muscle imbalance/contractures and deformities/orthopaedic concerns
    - Liaise with the neonatal orthopaedic physiotherapy team if required
- Teaching the parents/carers about the motor and sensory components of the condition, the effect of muscle imbalance, potential challenges with developmental progress and a home exercise programme, handling and positioning the child in preparation for discharge home
- Provide parents with an information leaflet containing exercises and advice
- Liaison with community physiotherapy teams with a detailed handover
- Follow up at Spina Bifida clinic with liaison, advice, and recommendations for ongoing management

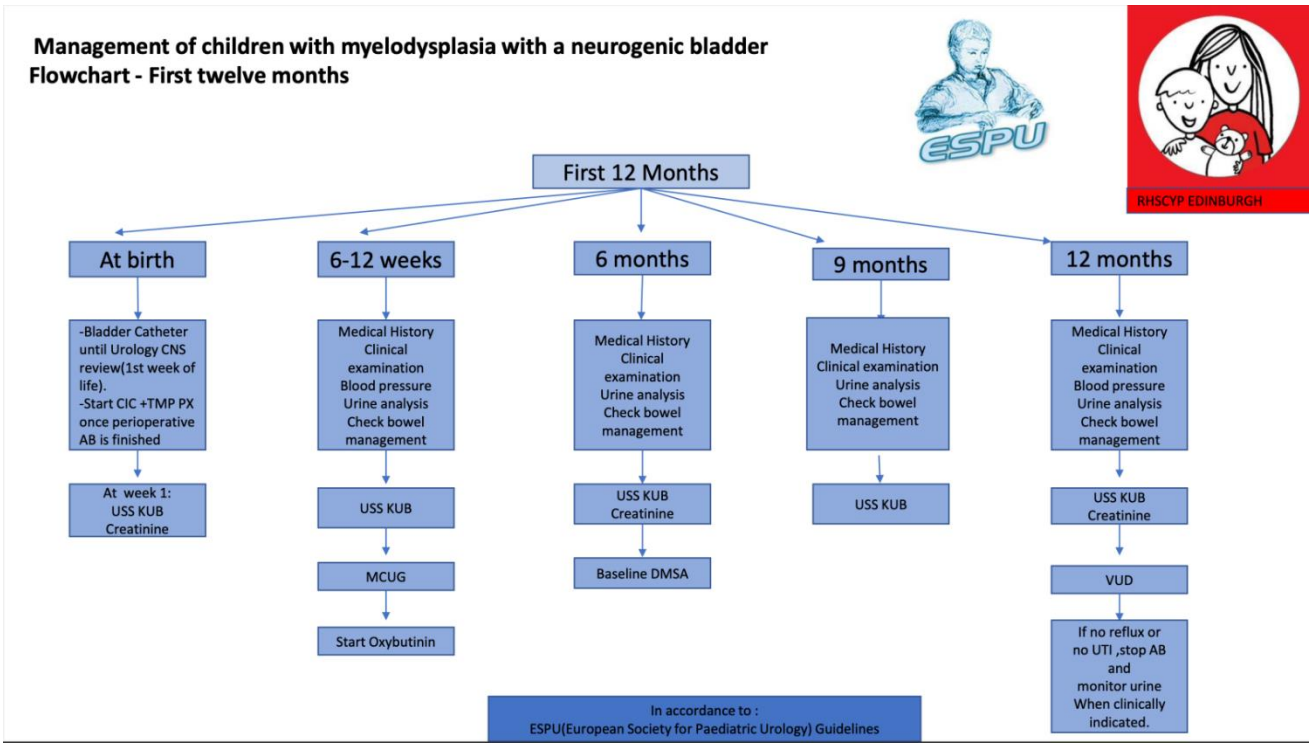
Discharge and follow up

- Ensure the level of the myelomeningocele is documented along with a full list of relevant problems such ventriculomegaly, Chiari malformation etc.
- Neurosurgical team should advise frequency of OFC measurements following discharge – this can be performed either by the health visitor or a children's community nurse.
- Ensure follow-up in the Spina Bifida clinic is arranged.
- Ensure that the following teams are aware of discharge:
    - Neurosurgeons - confirm if any outpatient neuroimaging needs to be organised
    - Urology
    - Orthopaedics
    - General surgeons
    - Health visitor
- Encourage the family to contact Spina Bifida charity for support
- Ensure parents are aware of symptoms and signs to monitor for and that they must seek urgent medical advice if they are noted:
    - Wound healing issues - swelling/leaking
    - Increased prominence of scalp veins
    - Sunsetting sign
    - Unexplained irritability/drowsiness
    - Poor feeding
    - Frequent aspiration
    - Episodes of cyanosis
    - Deteriorating limb strength

- Changes in bladder or bowel habits

6.0 Associated materials

Appendix 1



Appendix 2

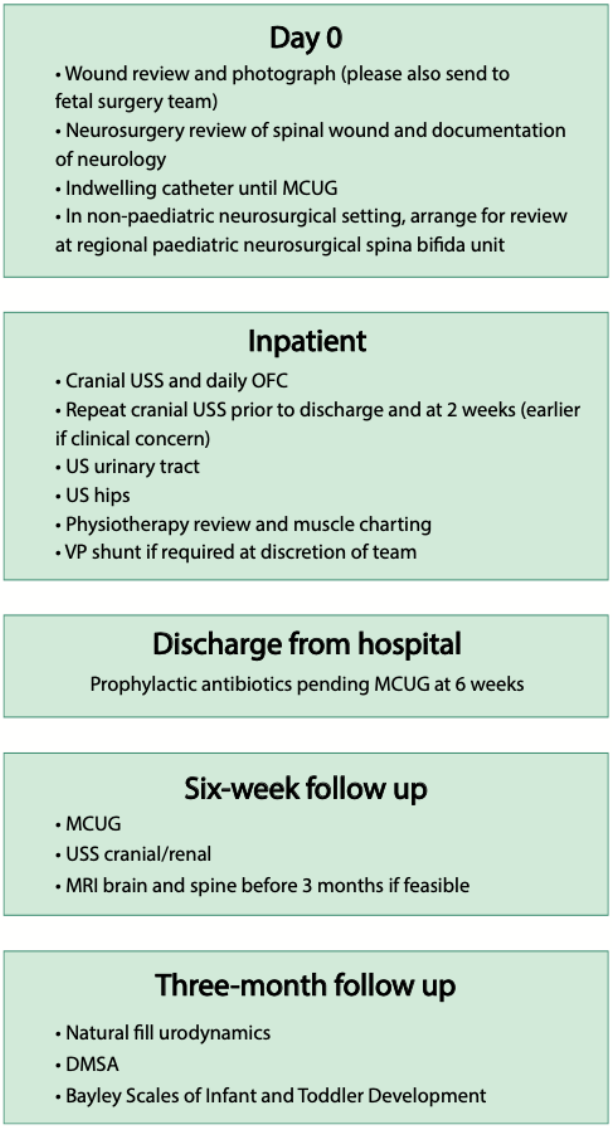


Figure 7. Neonatal protocol following open fetal surgery for spina bifida. DMSA = dimercaptosuccinic acid radionuclide scan; MCUG = micturating cystourethrogram; MRI = magnetic resonance imaging; OFC = occipito-frontal circumference; SCBU = special care baby unit; USS = ultrasound scan; VP = ventriculoperitoneal

7.0 Evidence base

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8.0 Stakeholder consultation

The guideline was drafted and circulated widely amongst various teams before finalising.

9.0 Monitoring and review

This guideline will be review on the 12th February 2029.