MYELOMENINGOCELE IN NEONATES: PRE AND POST-OPERATIVE MANAGEMENT - GCNC - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Neonates with myelomeningocele require surgical repair of the spinal lesion within the first 24-72 hours of life.
- Pre-operative management for myelomeningocele includes lesion stabilisation, positioning, infection control, prevention of latex sensitivity, monitoring of skin integrity, monitoring of respiratory function, management of neurogenic bladder and bowel, maintenance of thermoregulation and the provision of family support.
- Post-operative management for myelomeningocele includes post-operative care, wound management, infection control, monitoring of skin integrity, observation of hydrocephalus, diagnostic assessment of Chiari II Malformation, nutritional management, introduction of oral feeds, prevention and treatment of urinary and bowel complications, positioning, lower limb splinting to avoid pressure areas and optimise lower limb position and the provision of family support and education.

Key Performance Indicators:

- Protection of lesion is routinely undertaken pre operatively
- Monitoring and implementation of bladder drainage pre and post operatively
- Maintenance of skin integrity throughout the patient's hospitalisation pre and post operatively
- Implementation of latex precautions from birth and the use of latex free gloves and products.
- Photograph and documentation of the lesion prior to surgery within the patient's medical records (and progress notes if possible)

This document reflects what is currently regarded as safe practice. However, as in any clinical situation, there may be factors which cannot be covered by a single set of guidelines. This document does not replace the need for the application of clinical judgement to each individual presentation.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st July 2022	**Review Period**: 3 years
Team Leader:	Nurse Educator	**Area/Dept**: GCNC - CHW

CHANGE SUMMARY

- Content updated with updated references.
- Addition to content to include section on skin integrity.

READ ACKNOWLEDGEMENT

- All clinical staff working in Grace Centre for Newborn Care are required to read and acknowledge they understand the contents of this document.

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Defining Statement

This guideline relates to Neonates who present with the spinal lesion myelomeningocele (MMC) that requires surgical repair within the first 24-72 hours of life. More minor lesions do occur that either remain occult or that may require repair at a later date. Ensuring the appropriate clinical management of the neonate during the pre- and post-operative period is important for achieving optimal long-term growth and developmental outcome[1, 2, 3]. Hydrocephalus and Chiari Malformation (ACM) can occur as a consequence of the MMC, and the neonate with MMC will require screening and management of these conditions should they be present[4].

Definition of Myelomeningocele

Neural tube defects (NTD) are a spectrum of congenital anomalies, affecting approximately one per 1000 pregnancies. NTD results from the disruption in normal development of the brain and spinal cord during the early stages of embryogenesis (AIHW National Perinatal Statistics Unit, 2011). MMC is a form of NTD with arrest of the normal closure and containment of the neural tube with internalisation of the spinal cord. In MMC, interruption in the formation of the neural tube inhibits normal neural tissue differentiation and causes abnormal development of the spinal cord, meninges and vertebral column. In MMC, a meningeal sac containing malformed spinal cord tissue forms, and remains externalised through this retained opening in the spine and the skin[1, 5].

While the exact cause remains uncertain, epidemiological studies have revealed that NTDs are multifactorial. Environmental risk factors include obesity, stress, alcohol consumption, low calorie diet, folic acid deficiency, malnutrition, maternal hyperthermia, diabetes, hyperinsulinaemia, obesity, stress, epilepsy, and anticonvulsant medications. A family history of NTD, Trisomy 13 and 18, and various genetic syndromes and chromosomal rearrangements are also associated with NTD[6].

Some degree of neurological impairment is inevitable in MMC, however, the degree of impairment is determined by the location of the lesion, the extent of neural tissue involvement and the severity of hydrocephalus and ACM. There is no cure for MMC. Nerve tissue that is damaged or lost cannot be repaired or replaced. The lesion however can be managed[1, 5]. Standard procedure is to repair the MMC promptly after birth however there is increasing evidence in support of inutero repairs internationally. The aim of surgery is to provide suitable skin coverage to exposed nerve tissue, which serves to protect the nerve tissue from further damage, prevent cerebrospinal fluid (CSF) leakage and minimise the risk of infection[7, 4].

Assessment

Following birth, on admission to the neonatal intensive care unit (NICU), the neonate must undergo careful assessment.

- The neurosurgical team, Kids Rehabilitation team and neonatologist need to be informed of the neonate's arrival.
- Commence continuous cardiorespiratory monitoring. Assess and record baseline vital signs[8, 9].
- Examine the MMC to determine the level and condition of the lesion.
- Perform a head to toe examination to assess motor function, movement and reflexes, verify the presence of any dysmorphic features, and identify any additional spinal or orthopaedic deformities.
- The Kids rehab physiotherapist will review and assess the lower limb function and alignment. It is common for babies with MMC to have impaired motor function and reduced or absent sensation over affected dermatomes.
- A photograph of the lesion, with parental consent, must be taken by the medical photographer and documented in the neonate's medical record and progress notes. If there is a delay in taking photographs, the neurosurgical team could take photographs when the baby is positioned prone in theatre.
- Head circumference is recorded on the growth chart – daily[9].
- Assess the bladder and ensure passage of urine – this is particularly important to remember in infants nursed prone [10, 11] Record passage of meconium since birth and assess stooling function[12].
- Please see local guidelines on *Admitting a Neonate for Newborn Intensive Care*.

Pre-operative Care

Positioning and Handling

- The neonate must be nursed prone or in a side-lying position, and remain unclothed, with a loose nappy. Avoid placing clothing or blankets on or near the lesion to maintain integrity of the lesion to ensuring there is no undue pressure or risk of injury to the lesion and exposed nerve tissue.
- If supine positioning is unavoidable (e.g. for urgent intubation) a foam mattress with cut-out area should be available for use temporarily in these scenarios.
- Ensure all handling is gentle.
- Monitor skin integrity over pressure points as neonates with MMC are limited in how they can be positioned and may have lower limb immobility, contractures or weakness[13, 14].

Lesion Stabilisation and Monitoring

- The lesion must remain covered by a moistened, non-adherent gauze and non-permeable dressing (Bactigras), to prevent fluid loss and drying of the lesion, minimise evaporative heat loss, and protect the CSF from becoming contaminated.
- The dressing must be applied in a sterile non-touch technique, with sterile latex-free gloves, and must not be removed or redressed without a medical order.
- Observe and document the size, colour and perfusion of the lesion with a description of the membrane.
- Observe for tears in the membrane and monitor leaking of CSF. Inform the neonatal registrar and neurosurgical team of any tears in the membrane or excessive fluid leakage[13, 14].

Infection Control

Contamination of CSF can lead to infection of the brain and spinal cord which will delay shunt insertion and management of hydrocephalus and so must be prevented at all costs.

- Hand hygiene and infection control must be meticulous. Stool must be prevented from soiling the lesion as much as possible. Change nappies as soon as stooling occurs[13, 14]. Ensure that nappies are checked regularly for voiding. If not voiding, bladder scan should be performed and if the residual above upper limit then insert an IDC.
- For guidance on the use of the bladder scanner, see here.
- The neonate will be prescribed prophylactic broad spectrum antibiotics from birth until the time of surgical repair. There is insufficient evidence to support post-operative continuance of prophylactic antibiotics. Administration is therefore at the discretion of the treating teams[14, 15].

Latex Sensitivity

Neonates with MMC have a high risk of developing a latex sensitivity. Research has shown repeated early exposure to latex allergens is a major risk factor for sensitisation, and the avoidance of latex dramatically decreases the likelihood for an allergy developing[16, 17, 18].

- Nurse in a strictly latex-free environment. All forms of contact (direct, intravenous and intraoperative), must be avoided[16, 18].
- Use clean latex-free gloves when handling bodily fluid. Most stock in GCNIC is latex free (GloveOn Coats gloves, Nitrile).
- Use sterile latex-free gloves during sterile procedures such as catheterisation or dressing changes[18].
- The need for latex precautions must be clearly documented in the neonate's medical record and care plan, and this should be verbally handed over to all interdisciplinary teams involved in the patient's care[16]. Preferably the children should be first on an operation list as this will limit the exposure in theatre to latex.

Thermoregulation

Thermoregulation is of prime importance both before and after surgery. The neonate with MMC is at increased risk of hypothermia owing to the immaturity of their thermoregulatory system, their inability to generate heat from muscle movement in their lower limbs, lesion related evaporative heat and fluid losses, and having to be nursed bare[14].

- Nurse the neonate in a neutral thermal environment to maintain axillary temperature between 36.5-37.5º C.
- Skin temperature monitoring must be continuous and axillary temperature should be measured regularly. Skin temperature should read 0.5ºC less than the axillary temperature[19].
- Providing an external heating source such as baseplate mattress heating and/or a radiant warmer may be required.
- Dressing in a beanie and booties may help[8].

Preparation for Surgery

Most neonates will have surgery within the first 24-72 hours. Surgery may be scheduled earlier if the lesion is ruptured or there is increased intracranial pressure with hydrocephalus[20].

- Please see local guidelines on Transfer of a Neonate to Operating Theatre and other Hospital Investigative Departments.

Post-operative Care

Immediate Post-operative Assessment:

On returning to NICU/CICU from the operating theatre:

- Inform the medical team of the neonate's return to the unit.
- Perform a head to toe assessment.
- Assess airway support and document[9].
- Monitor temperature closely as the neonate may have become hypothermic intra-operatively[8].
- Perform a pain assessment and document score[14]. See also local Clinical Practice Guideline: *Pain Management of the Newborn Infant*.

Wound Care and Handling

- Refer to instructions and post-operative orders made by the neurosurgeon in the operative notes.
- Neonates are to continue to be nursed prone or side-lying for 7-10 days, to prevent pressure on the suture lines and to promote wound healing.

- Observe wound site and document appearance, colour, dressing and perfusion. If wound or surrounding areas are dusky in appearance inform the NICU/CICU registrar and neurosurgical team, and have the neonate reviewed.
- The wound must remain covered with a transparent dressing and should be assessed with cares for signs of leakage, dehiscence and infection. If soiling is noted, the dressing should be changed to minimise risk of wound infection in consultation with the medical and nursing team leaders.
- Soiling of the wound must be avoided until it is properly healed[14, 15].

Chiari Malformation

CM is a constellation of abnormalities in the brain almost always present in babies with MMC. The malformation is characterised by herniation of the cerebellum and medulla down into the spinal canal and into the middle fossa. CM is commonly associated with hydrocephalus[4] which can lead to cranial nerve dysfunction and raised intracranial pressure. Signs of this include dysphagia, absent gag reflex and poor breathe, suck, and swallow coordination and cardiorespiratory compromise[2,].

- The neonate must remain on continuous cardiorespiratory monitoring.
- Observe for signs of cardiorespiratory compromise such as bradycardia, tachycardia, hypotension, insufficient breathing, apnoea and inspiratory stridor[2,8].
- Monitor for signs of swallowing difficulty, aspiration of saliva or oral feeds, hoarse voice, facial weakness, progressive weakness in the lower extremities, or any sudden changes in bladder and bowel habits. Promptly inform the NICU/CICU Registrar of such changes[5].
- Measure head circumference daily [21].

Hydrocephalus

Hydrocephalus is a prominent issue in major MMC, affecting up to 90% of neonates either at birth or after lesion repair at 2-3 weeks of age[1, 22]. CSF is normally continuously produced by the choroid plexus in the ventricular system and flows freely around the brain and spinal cord and is reabsorbed in the arachnoid granulations. In MMC, hydrocephalus results from obstruction to this flow into the subarachnoid space around the spinal cord in the presence of CM and hindbrain herniation[4]. Hydrocephalus and the subsequent increased intracranial pressure and will require drainage to prevent brain injury.

- Perform neurological observations at fourth hourly intervals (or more frequently if clinically indicated)[1, 22].
- Measure head circumference daily (or more frequently if clinically indicated).
- Monitor for signs of raised intracranial pressure such as a rapidly expanding head circumference, an enlarged head, a tense or bulging fontanelle, separation of the cranial sutures, episodic apnoea and bradycardia, stridor, irritability, seizures, a high pitched cry, bulging or sunset eyes, altered equality or reactivity of the pupils, swallowing dysfunction or feed intolerance[21].
- The neonate who exhibits acute neurological changes such as stridor, swallowing dysfunction or central apnoea requires prompt review by the unit registrar or fellow[2].

- A head ultrasound, and/or magnetic resonance imaging will be part of the evaluation of the ventricular size[2, 9].
- CT scans should be limited where possible as each scan delivers a high dose of radiation which can lead to increase of metastatic disease later in life[23].

Urinary Tract Complications

There is a high incidence of neurogenic bladder among neonates with MMC[24, 10].

- Measure urine output and monitor fluid balance accurately.
- Observe for signs of 'neurogenic bladder' such as continuous dribbling, squirting voiding habit, only voiding on abdominal stimulation or lack of regular voiding such as dribbling of urine or anuria.
- Urinary investigations should be performed in all babies. This includes external examination, renal USS, MCUG, bladder scanning at set intervals. A DMSA scan may be requested whilst in the NICU, but is usually performed after discharge.
- Monitor for signs of urinary tract infection such as, temperature instability, lethargy, irritability, pain and increased respiratory support [25,26]. Occasionally the urine maybe noted to be dark, cloudy or foul smelling but this is rare in the newborn period.
- The administration of anticholinergic medications and/or prophylactic antibiotics may be considered to improve storage and reduce bladder pressure and minimise the likelihood for infection[10].
- Clean intermittent catheterisation (CIC) may be required to properly empty the bladder. Initiation of CIC and frequency of CIC will usually be determined by the Kids Rehab team or urologist[10, 11].
- An indwelling urinary catheter is not recommended for extended periods of time due to the high risk of infection[10].

Bowel Complications

Neurogenic bowel can also occur in neonates with MMC.

- Closely monitor and document stool consistency and regularity.
- Observe for signs of neurogenic bladder, as well as constipation with overflow. Report any sudden changes in bowel patterns to the unit registrar/fellow[12].
- Monitor skin integrity around anus and buttocks and observe anal tone. A patulent anus may leak stool continuously.
- Change nappies and clean area in a timely manner to promote perianal skin integrity and prevent soiling of the surgical wound.
- An emollient barrier cream will be required to promote perianal skin integrity[14].

Nutrition

Ensuring the neonate receives adequate nutrition is fundamental to support wound healing, postoperative recovery, growth and development.

- Preoperatively, it is appropriate that the neonate is kept nil by mouth or receives parenteral nutrition, as surgery is usually performed in the first 48-72 hours of life.
- Postoperatively, parenteral nutrition should be administered until the neonate is clinically stable and it is appropriate to commence and grade up on enteral feeds[5, 14].

Oral Feeding

- The neonate may breast or bottle feed in a side-lying position, with care taken to ensure pressure is not placed on the surgical wound.
- Most neonates with MMC will feed vigorously and do not have suck, swallow coordination problems.
- Alternate side-lying position reduces the likelihood the neonate will begin to favour one side. Liaise with the Occupational Therapist for positioning devices if required[14].
- Assess for signs of aspiration with oral feeding. Signs of dysphagia, absent gag reflex and poor breathe, suck, swallow, coordination are serious signs of untreated CM.
- Observe for signs of gastro-oesophageal reflux, which may be due to the CM but also related to the positioning required during feeding neonates with MMC.
- Enteral feeding is preferred to parenteral nutrition. Consider placing a nasogastric tube for tube feeding if the neonate does not achieve a sufficient oral intake[5].
- The involvement of a nutritionist, speech pathologist and lactation consultant is appropriate and encouraged[14].

Family-Centred Care

Ensuring parents and families are sufficiently supported is an important priority in the pre and postoperative care of neonates with MMC.

- Orientate parents to the ward environment and routines, such as medical rounds and visiting hours.
- Ensure adequate time is taken to provide parents with accurate explanations. MMC is the lesion that leads to Spina Bifida (SB), however Spina bifida (SB) is what the child will live with post management of the MMC. Spina bifida is the constellation of symptoms that the child is left with to manage throughout life. The parents will need to understand how their child's life will be impacted by SB, what supports there are and what their role will be in supporting a child with SB with regards to treatments, surgery, complications, prognostic expectations[27] as well as the likelihood of recurrence in future pregnancies[28]. The majority of the families will have had antenatal counselling regarding their child's predicted functional risks by the Kids Rehabilitation team and it is useful for the clinical team at the bedside to understand this counselling to be able to address gaps in predictions and what is the postnatal reality for their child. Parents may experience a period of shock whilst they come to terms with their child presenting as better or worse once they are born and the team at the bedside will need to be sensitive and supportive through this transition for the families.
- Encourage parental involvement in care. Provide education about nappy changes, feeding, care practices and precautions, medication administration and catheterisation.
- Encourage cuddles and kangaroo care to support attachment and bonding[29].

- Many parents will have met with the Kids Rehab team in the antenatal period which includes a social worker, OT, physiotherapist, clinical nurse consultant and medical consultant. This team should be involved early to help parents adjust to the birth of their baby and the ensuing challenges.
- Involve the Social Worker and Discharge Liaison Nurse early on in patient care. The Social Worker is aware of ongoing support networks for the parents and the neonate. The Discharge Liaison Nurse can assist parents in preparing for taking their baby home and can help coordinate common services in the community[27].

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