Neuroscience NICU

Myelomeningocele Guidelines

(Last Revised November 2018)

This guideline provides recommendations regarding the prenatal, perinatal, and perioperative management myelomeningocele, a type of open neural tube defects. Infants with skin-covered neural tube defects should be managed differently and would be excluded from these guidelines.

Prenatal counseling

- Most myelomeningoceles are currently diagnosed by ultrasound prenatally
- Ideally, mothers carrying a fetus with a myelomeningocele should receive prenatal counseling with their obstetrical provider as well as with postnatal providers (neonatology, neurosurgery, urology) through the Fetal Care Center
- Fetal consultation should ideally occur prior to 23 weeks of gestation in order to allow for discussion of referral for fetal repair

Initial postnatal management

- Latex allergy precautions
- Handle with sterile, latex-free gloves and sterile towels at delivery
- Place side-lying or prone to ensure no pressure is placed on the lesion
    - **If infant requires positive pressure ventilation or intubation:**
        - Place infant supine with lesion positioned within a donut hole
        - Support the head with fluidized positioner or sponge donut center
- The lesion should be dressed to minimize the potential for injury during transport:
    - Place infant prone, carefully dry upper body and legs, avoiding pressure on the lesion
    - Wrap lesion with sterile gauze soaked in warm saline
    - Cover lesion and abdomen with layers of occlusive plastic wrap
- For lesions without skin covering, the infant should be started on prophylactic ampicillin and gentamicin until 48 hours after surgical closure of the sac
- If not delivered at NMC or the FCC, transfer to CH&MC NICU should occur promptly to allow for repair within the first 48 hours after birth
- Maintain the infant prone throughout transport

Admission to the NICU

- See admission checklist
- On arrival, the following services should be consulted/notified:
    - Neurosurgery
    - Urology
    - OT/PT (to see 48 hours post-operatively)

Bedside care:

- See skin care sheet
- Monitor for symptoms of Chiari II (apnea, bradycardia, hypoventilation, stridor, swallow dysfunction)
- Monitor daily head circumference
- Maintain NPO with appropriate IV fluids (D10W or starter TPN) upon admission until operative plan has been established
- Most infants with isolated myelomeningocele should be able to feed orally, so if OR is not imminent, should be allowed to attempt PO until necessary to make NPO pre-operatively
- Initiate clean intermittent catheterization protocol[1]:
    - Start with q6h and continue through peri-operative period
    - Once stable post-operatively, if volumes ≤30 mL for ¾ catheterizations for 24 hours, increase interval to q8h, then q12h, then q24h
    - May stop catheterizing if:
        - Residuals ≤30 mL for 3 days
        - RUS ≤ grade 2 hydronephrosis
    - At any point if residuals >30 mL, return to frequency that maintains residuals ≤30 mL and maintain that frequency through discharge
    - If RUS demonstrates > grade 2 hydronephrosis, maintain at q6h frequency regardless of residual volumes
    - Note: Catheterization must include aspiration of the catheter to ensure that emptying of low-pressure bladder is achieved

Imaging:

- Obtain HUS upon admission
- Obtain echocardiogram unless negative prenatal echo
- Renal and bladder US within first 24 hours if prenatal ultrasound with hydronephrosis, otherwise obtain post-operatively
- VCUG prior to discharge

Examination:

- Assess for other malformations and possible syndromes that could affect management
- Determine lower extremity strength and sensory level

<u>Post-operative management</u>

- Continue antibiotics for 48 hours after closure unless instructed otherwise by neurosurgery
- Continue side-lying or prone positioning until cleared by neurosurgery for supine
    - Starting no earlier than 24 hours after surgery, parents may hold infant in horizontal position, taking care to avoid any pressure on the wound site
- Infant may resume oral feeding once awake and cueing after surgery

Neuroimaging and Follow-up for Hydrocephalus

- Hydrocephalus is common in infants with myelomeningocele
- Continue daily head circumference until discharge
- Post-operatively, infants should receive head ultrasound daily on POD #1 and 2, and then as needed
- Post-operative MRI should be completed prior to discharge

Neuroimaging After Shunt Placement (if applicable)

- POD #1, all infants should have:
  - Shunt series
  - Rapid brain MRI (or full brain MRI if infant is close to discharge to replace the post-operative MRI "prior to discharge" described above)

Outpatient Follow-up

- These infants should be discharged in a conventional car seat, unless a car bed is required for other indications (e.g. desaturation or bradycardia during angle tolerance test) or if neurosurgery has not lifted the prone/side-lying restrictions prior to discharge
- All infants with myelomeningocele should follow up with
  - Neurosurgery 2 weeks after surgery for a wound check and head US
  - Urology one month after discharge with renal and bladder US
  - CDC clinic 3-6 months after discharge
  - TIPS 3 (EDN referral by social work)
- If club feet or musculoskeletal anomalies, ensure that outpatient orthopedic follow-up is scheduled prior to discharge for 1-2 weeks after discharge

References

1. **Routh JC, Cheng EY, Austin JC, et al. Design and Methodological Considerations of the Centers for Disease Control and Prevention Urologic and Renal Protocol for the Newborn and Young Child with Spina Bifida. J Urol 2016;196:1728-34.**

	Pre-op	Post-op			Pre-op Shunt	Post-op Shunt	Discharge
		Day 0	Day 1	Days 2-3			
Vitals/ Monitors	Per NICU Protocol	Per NICU Post-op Protocol	Per NICU Protocol			Per NICU Post-op Policy	
Neuro Checks	Check anterior fontanel q4h Head circ daily						Include DC head circ in DC summary
Imaging	HUS on admission, echo unless normal prenatal echo, RUS if fetal hydronephrosis		HUS POD 1, renal US	HUS on POD 2	HUS PRN	POD1: Shunt series and rapid brain MRI (or full brain MRI if close to DC)	VCUG and full brain MRI prior to discharge
Diet / IVF	Per NICU Team, NPO prior to surgery per anesthesia	Diet / IVF per NICU Team			Per NICU Team, NPO prior to surgery per anesthesia	Diet / IVF per NICU Team	
Activity / Positioning	Prone for ≥48 hours with Z-Flo under abdomen, chest, and head. HOB Flat	Prone or side-lying (no holding) with Z-Flo under abdomen, chest, and head. HOB Flat	If repair complex (w/ plastics), NS will clear for activity, otherwise may hold 24 hrs post-op				Must observe for at least 48 hours in supine position prior to discharge
Wound / Skin Care	Transport: Sterile gauze with sterile warm saline. Wrap with Saran wrap, diaper below defect, + mudflap	Site covered x 48 hrs post-op, notify NS resident for dressing changes due to soiling. Keep mudflap at all times.			Soap and water bath night before and morning of shunt surgery. CHG bath per policy.	Any dressings will be removed POD 1 by NS; Daily wound washing with soap and water starting POD 2	

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		Day 0	Day 1	Days 2-3			
Antibiotics	Amp / Gent from delivery until repair	Amp / Gent x 48 hrs post-op			Ancef (40mg/kg) on call to OR	Ancef x 24 hours post-op	
Pain	Per NICU Team; avoid narcotics if possible	Scheduled Tylenol x 24 hrs; minimize narcotic use	Tylenol PRN; minimize narcotic use			Scheduled Tylenol x 24 hrs; minimize narcotic use	Tylenol PRN; minimize narcotic use
Urologic Management	Q6H intermittent caths	Follow cath protocol					
Consults	Neurosurgery Urology OT/PT						
Follow Up							Neurosurgery, urology, CDC, TIPS 3. If club feet, ensure ortho follow-up

Admission Checklist

Consult:

- Neurosurgery
- Urology
- PT/OT
- TIPS 3

Medications:

- Ampicillin (until 48 hours post-op)
- Gentamicin (until 48 hours post-op)
- Vitamin D
- D10W or starter TPN at 80 ml/kg/day

Admission labs:

- CBC with differential
- Type & Screen
- No other labs are indicated specifically for myelomeningocele – order labs as otherwise clinically indicated

Imaging:

- Renal ultrasound
    - Within first 24 hours if prenatal ultrasound with hydronephrosis, otherwise obtain post-operatively
- Head US on admission
- Echocardiogram unless normal fetal echo

Procedures:

- NPO (may place feeding orders, if appropriate, once surgical plan in place)
- Latex precautions
- Activity restrictions: maintain prone
- Dressing instructions
- Initiate clean intermittent catheterization protocol

Skin care for the myelomeningocele patient

Diaper care from admission:

1. Wash perianal area with Cetaphil gentle cleanser BID. Pat, do not rub when able
2. If skin reddened at all, increase above wash to include with stools
3. Apply A& D ointment as barrier if reddened at all. Otherwise, initiate the Sensicare/ stoma powder regimen.

Post-operative incisional care:

1. Place Duoderm flap just above the intergluteal cleft, ensuring no contact with the incision
2. Place cotton ball at the cephalic tip of the cleft for absorption and to protect the integrity of the Duoderm.
3. Follow all postoperative neurosurgery guidelines for incisional care. Notify if there is increased redness, stress along the suture line, possible dehiscence, or leakage of any fluid.
    a. If infant returns from OR with dressing, it should remain in place for two days
    b. If glue was used to close wound, it will remain open without dressing
4. If fluid is observed, protect the surrounding skin with a barrier (Cavilon and then Duoderm).

Home care:

1. No immersion baths for 6 week post-operatively. Sponge baths only.
2. Continue with Cetaphil as soap for cleansing.
3. If perianal area is reddened despite the above regimen, add a tablespoon of baking soda to baths