Newborn Critical Care Center (NCCC) Clinical Guidelines

Myelomeningocele (MMC) Guidelines

Myelomeningocele is a complex chronic condition which requires lifetime care coordination. The myelomeningocele sequence includes the open neural tube defect (ONTD), hydrocephalus and equinovarus deformities. The NCCC goal of care is to optimize the outcome for the infant. Care needs will vary according to the level of the lesion as well as the timing of the repair. Some prenatally identified patients will undergo in–utero repair of the lesion. The goal of fetal surgery is to decrease the risk of subsequent hydrocephalus that would require ventriculo-peritoneal (VP) shunt placement.[1] Some patients identified prenatally will have consulted with Pediatric Neurosurgery (Drs. Elton/Quinsey), Pediatric Urology (Dr. Ross), Neonatology, and Physical Medicine and Rehabilitation (Pediatric Medicine and Rehabilitation Attending) prior to delivery. Since 90-95% of infants with MMC have neurogenic bladder and renal damage which can begin within the first six months of life; this contributes significantly to their morbidity, and an aggressive urological approach is suggested.[2]

GUIDELINES FOR INITIAL CARE

1. Admission and stabilization of the infant.
2. **Position prone.**
3. Dressing:
    a. Open lesions (Wet): non-adherent pad (Telfa) moistened with normal saline, covered by a 4x4 and stabilized with loose gauze bandage roll (Kerlix gauze) wrapped around the infant (NO other dressing, NO Vaseline gauze)
    b. Prenatal closures (Dry): non-adherent pad (Telfa) covering taped at edges until evaluated by Neurosurgery
4. Begin Ampicillin and Gentamicin for infants with open lesions and continue 24 hours post-operatively. Prenatal closed repairs do not generally require antibiotics
5. Consult Pediatric Neurosurgery.
6. **Insert indwelling catheter and maintain until infant permitted to lie supine (discuss with Pediatric Neurosurgery and Pediatric Urology).**
7. Initiate Latex precautions.
8. Assess degree of neurological involvement.
9. Monitor for signs of increased intracranial pressure.
10. Birth and daily head circumference
11. Begin UTI prophylaxis with Amoxicillin 20 mg/kg/day (to start after discontinuing initial Ampicillin). Continue until VCUG is complete and the results are discussed with Pediatric Urology.
12. Strict intake and output.

13. Imaging:
    a. For open lesions, obtain head ultrasound (HUS) between 1-3 days post-operatively for baseline. No imaging is needed in the pre-operative period.
    b. For prenatal closures, obtain HUS at birth.
    c. A brain MRI is not necessary in the neonatal period unless there are extenuating circumstances.
14. Consult other services (timing may vary) as appropriate: Pediatric Urology, Physical Medicine and Rehabilitation, Orthopedics, Genetics – see below.

SURGICAL REPAIR - OPEN MYELOMENINGOCELE

1. **Anticipate OR on DOL #0-2 for repair (closure). "Mud flap" (3M Steri-Drape™ / plastic drape with adhesive strip) will be placed in the OR.**
2. Assess surgical site daily and notify Pediatric Neurosurgery team if there are concerns.
3. Infant to remain FLAT, either prone or on their side for the first 48 – 72 (confirm time with Pediatric Neurosurgery) hours post-op.
    a. Do not raise the head of the bed until cleared to do so by Pediatric Neurosurgery.
4. Infant may be held in the lateral position to breast feed, if applicable.
5. After 48-72 hours post op (confirm time with Pediatric Neurosurgery), once the infant is stable from NCCC viewpoint, infant may be held by the parents (flat, prone or lateral).
    a. Use a pillow to transfer and facilitate comfort while holding.
6. A "mud flap" will be placed in the OR and should be maintained for 72 hours. It is attached to the skin horizontally below the surgical incision and above the buttocks to prevent stool from contaminating the incision. Hang it out over the diaper. It can be removed once the surgical dressing is discontinued. The back of the diaper should not cover the incision.

INFANTS WITH CLOSED LESIONS REPAIRED PRENATALLY

1. Consult Pediatric Neurosurgery
2. Follow initial care guidelines generally these infants will not require antibiotics
3. Obtain head ultrasound (HUS) at birth
4. These lesions may or may not be healed and may require wound care/dressing. This should be determined with Pediatric Neurosurgery.

SUB-SPECIALTY CONSULTS

Pediatric Urology

1. Monitor strict intake and output
2. UTI prophylaxis: Amoxicillin 20 mg/kg/day until VCUG obtained and the results are discussed with Pediatric Urology (see initial care).

3. Obtain renal ultrasound (RUS) at > 48 hours life for baseline. For long-term NCCC infants, a repeat RUS prior to discharge may be indicated.
4. Obtain voiding cystourethrogram (VCUG) to rule out vesicoureteral reflux (VUR) prior to discharge.
5. Initiate clean intermittent catheterization (CIC) after birth for those who do not have an indwelling catheter.
    a. Volume < 30 mL on two separate measurements discontinue CIC
    b. Volume > 30 mL, continue CIC every 4 hours except overnight (2300-0700) unless infant wakes then perform CIC
    c. For long term NCCC patients NOT undergoing CIC, a second CIC trial may be warranted prior to discharge
6. Determine post-void residual measurement prior to discharge.
7. Teach the family how to perform I & O catheterization if indicated.
8. Order CIC equipment for home prior to discharge (see complex discharge order set).
9. Male infants should have circumcision prior to discharge. This will be covered by Medicaid as a medical indication – use “spina bifida, neurogenic bladder” as indication for circumcision.
10. Obtain baseline creatinine and BUN prior to discharge.
11. Expect the infant to have urodynamic studies at >/= 3 months of age with Urology follow-up. These studies may need to be scheduled prior to discharge. Discuss with pediatric urology prior to discharge the coordination of the urodynamic studies.

All myelomeningocele infants must have Pediatric Urology follow-up. Dr. Ross attends the Spina Bifida Clinic, so a separate Pediatric Urology appointment is not always indicated. Check with Dr. Ross about the timing of urology follow-up and schedule appropriately.

Pediatric Genetics

*Consult for any of the following**:***

1. MMC sequence only and no prenatal genetic counseling or parents have questions about recurrence risk or maternal preconception folic acid guidelines with future pregnancies – *specify genetic counseling*
2. Infant has additional malformations/dysmorphic features not in MMC sequence
3. Infant has abnormal genetic testing

Physical Medicine and Rehabilitation (PM&R)

1. Consult Pediatric Medicine and Rehabilitation Attending for in-hospital evaluation
    a. Prenatal closures: contact PM&R Attending following delivery

b. Open lesion: contact PM&R post-operatively when infant is stable and has freedom of movement

2. Obtain Occupational Therapy (OT) consult for splinting/positioning
3. Schedule follow-up with PM&R Attending, in the Spina Bifida Clinic. The attending will indicate follow-up appointment time in their initial consult.

Pediatric Orthopedics

1. Assessment for talipes equinovarus
2. Assessment for dislocated hips

DISCHARGE PLANNING

1. All infants should be referred to CDSA (Children's Developmental Service Agency) so home therapy services can begin after discharge.
2. Some infants may require a Home Health referral if they are in need of dressing changes, have a gastrostomy tube and/or other medical equipment.
3. If a car bed is necessary, UNC Hospitals will provide.

References:

1. Adzick NS et al. A Randomized Trial of Prenatal versus Postnatal Repair of Myelomeningocele**.** *N Engl J Med* 2011; 364:993-1004.
2. Kessler TM, Lackner J, Kiss G, Rehder P, Madersbacher H. Early proactive management improves upper urinary tract function and reduces the need for surgery in patients with myelomeningocele. *Neurourol Urodyn* 2006; 257: 758–762.