Newborn Critical Care Center (NCCC) Clinical Guidelines

Myelomeningocele (MMC) Guidelines

Myelomeningocele (MMC) 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 patients identified prenatally 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, Pediatric Urology, Neonatology, and Physical Medicine and Rehabilitation 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. Antibiotics:
    a. Open lesions: Begin Ampicillin and Gentamicin and continue 24 hours post-operatively
    b. Prenatal closures: Do not generally require antibiotics
5. Measure size and determine location of defect, take a picture of the defect and upload into the EPIC media tab
6. Consult Pediatric Neurosurgery
7. Assess degree of neurological involvement with detailed neurologic, skin, and spine exam
8. Monitor for signs of increased intracranial pressure via vital signs, fontanelle exam, and neurologic exam
9. Obtain birth and daily head circumference

10. Insert indwelling catheter and maintain until infant permitted to lie supine (discuss with Pediatric Neurosurgery and Pediatric Urology)
11. Consult Pediatric Urology
12. Initiate Latex precautions
13. Begin UTI prophylaxis with amoxicillin 10 mg/kg/day (to start after discontinuing initial ampicillin), and continue until VCUG is complete and the results are discussed with Pediatric Urology.
14. Follow strict intake and output
15. 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
16. Consult other services (timing may vary) as appropriate: Pediatric Rehabilitation Medicine, Pediatric Orthopedics, Genetics – see below

SURGICAL REPAIR - OPEN MYELOMENINGOCELE

1. Anticipate OR on DOL #0-2 for repair (closure).
2. A “mud flap” (3M Steri-Drape™ / plastic drape with adhesive strip) will be placed in the OR and should be maintained by NCCC nursing for 72 hours
    a. It is attached to the skin horizontally below the surgical incision and above the buttocks to prevent stool from contaminating the incision (should hang over the diaper with the back of the diaper NOT covering the incision)
    b. It can be removed once the surgical dressing is discontinued
3. Assess surgical site daily and notify Pediatric Neurosurgery team if there are concerns
4. 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
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. Infant may be held in the lateral position to breast feed, if applicable

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

To Begin After Closure of the Back

1. Monitor strict intake and output
2. Obtain renal and bladder ultrasound (RBUS) at >48 hours of life for baseline (for long-term NCCC infants, a repeat RBUS may be indicated)
3. Once patient moved from prone begin clean intermittent catheterization (CIC) every 6 hours for those without indwelling catheter to find residual bladder volumes
    a. If volume <30 mL for 3 out of 4 catheterizations in 24hr interval, then increase interval to every 8 hours
    b. If volume <30 mL for 2 out of 3 catheterizations in 24hr interval, then increase interval to every 12 hours
    c. If volume <30 mL for 1 out of 2 catheterizations in 24hr interval, then increase to every 24 hours
    d. If volume <30 mL for 1 out of 1 catheterizations in 24hr interval, then stop catheterizations
    e. If fails any step of the CIC protocol (3a-d) then decrease catheterization interval in a stepwise fashion up to CIC every 4 hours
    f. For long term NCCC patients NOT undergoing CIC, a second CIC trial may be warranted prior to discharge
4. Teach the family how to perform CIC, if applicable
5. Order CIC equipment for home prior to discharge if needed (see complex discharge order set)
6. Male infants should be offered circumcision prior to discharge. This will be covered by Medicaid as a medical indication – use “spina bifida, neurogenic bladder” as indication for circumcision
    a. Discuss with pediatric urology the timing/type of circumcision to be performed if patient has atypical anatomy
7. Expect the infant to have voiding urodynamic studies (VUDS) at ≥ 3 months of age with Urology follow-up. If unable to have VUDS then obtain voiding cystourethrogram (VCUG) to rule out vesicoureteral reflux (VUR) and reach out to urology to schedule VUDS for next available spot.
8. Future medication guidelines upon urodynamics findings:

a. 0.2 mg/kg oxybutynin PO TID for hostile bladder or significant trabeculation on US or VCUG
b. Antibiotics for patients with vesicoureteral reflux, grade 3 hydronephrosis, or a hostile bladder. Amoxicillin 10mg/kg PO daily until age 2 months. Treating physician will then choose between daily trimethoprim/sulfamethoxazole (2 mg/kg) or nitrofurantoin (1 to 2 mg/kg) suspensions.

All myelomeningocele infants must have Pediatric Urology follow-up. (Dr. Ross and Dr. Arora) Separate Pediatric Urology appointments are necessary as pediatric urology does not participate in spina bifida clinic at this time. Check with Dr. Ross or Dr. Arora 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 PM&R (Pediatric Rehabilitation Medicine Attending) for in-hospital evaluation
   a. Open lesion: contact PM&R post-operatively when infant is stable and has freedom of movement
   b. Prenatal closures: contact PM&R following delivery
2. Obtain Occupational Therapy (OT) consult for positioning (and splinting when indicated)
3. Schedule follow-up in the Spina Bifida Clinic by calling 984-974-9747. The attending will indicate follow-up appointment time in their initial consult.

Pediatric Orthopedics

1. Consult Pediatric Orthopedics for assessment for talipes equinovarus and/or 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.
3. Routh, J. C., Cheng, E. Y., Austin, J. C., Baum, M. A., Gargollo, P. C., Grady, R. W., Herron, A. R., Kim, S. S., King, S. J., Koh, C. J., Paramsothy, P., Raman, L., Schechter, M. S., Smith, K. A., Tanaka, S. T., Thibadeau, J. K., Walker, W. O., Wallis, M. C., Wiener, J. S., & Joseph, D. B. (2016). 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. The Journal of urology, 196(6), 1728–1734.

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