Newborn Critical Care Center (NCCC) Clinical Guidelines

Myelomeningocele (MMC) Guidelines

Myelomeningocele (MMC) is the most severe form of open neural tube defect and is a complex medical condition that requires lifetime care coordination. MMC affects 0.8 to 1.0 of 1,000 live births worldwide; and 0.2 to 0.4 per 1,000 live births in the US.[1] MMCs are believed to form due to failed closure of the caudal end of the neural tube during primary neurulation in embryogenesis.[2] This defect leads to herniation of the meninges and spinal cord through a vertebral defect and causes neurologic impairment below the level of the lesion. The most common location for defect is the lumbosacral region. MMC is frequently associated with Chiari II malformation (herniation of the vermis, medulla, and fourth ventricle into the spinal canal) and hydrocephalus.[3] The American Academy of Pediatrics emphasizes coordinated, multidisciplinary care beginning in the neonatal period.

Neurulation is normally completed by approximately day 28 post-conception. The most strongly associated risk factor is low folate levels during pregnancy.[2] In MMC, failure of neural tube closure results in exposed neural tissue that is vulnerable to ongoing injury in utero due to mechanical trauma and amniotic fluid exposure, referred to as the 2-hit hypothesis.[4] Conditions associated with MMC include hydrocephalus, neurogenic bladder and bowel dysfunction, and orthopedic abnormalities such as clubfoot and hip dysplasia.

A pivotal advance in the field came from the Management of Myelomeningocele Study (MOMS).[5] This randomized controlled trial compared the safety and efficacy of prenatal repair of myelomeningocele with that of standard postnatal repair. The MOMS Trial found that fetal surgery significantly reduced the need for ventriculoperitoneal shunting and improved motor outcomes at 30 months of age. The trial was stopped early for efficacy. However, these benefits came at the cost of increased risks, including preterm delivery, uterine dehiscence, and maternal morbidity. Complications related to prenatal surgery included oligohydramnios, chorioamniotic separation, placental abruption, and spontaneous membrane rupture. Fetuses that were treated prenatally were born at an average gestational age of 34.1 weeks and 13% were delivered before 30 weeks gestation.

Delivery should be planned at a tertiary care center with access to neonatology, neurosurgery, and urology. 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.[6] Prognosis varies widely and depends largely on the level of the lesion, the size of the lesion, and the presence of associated conditions such as hydrocephalus.[2] Higher spinal lesions are associated with greater motor impairment. The NCCC goal of care is to optimize the outcome for the infant through careful, multidisciplinary care.

INITIAL CARE: ADMISSION and STABILIZATION

1. Position prone
2. Use foam DONUT to offload the defect for any procedures that require supine positioning.
3. Myelomeningocele (sac and surrounding skin) should *not* be manipulated at any time due to risk of rupturing. Any contact/touching of the skin around the MMC should occur with a pair of sterile gloves.
4. 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). Do not change this dressing unless grossly soiled.
    b. Prenatal closures (Dry): non-adherent pad (Telfa) covering taped at edges until evaluated by Neurosurgery
5. Antibiotics:
    a. Open lesions: Begin Ampicillin and Gentamicin and continue 24 hours post-operatively
    b. Prenatal closures: Do not generally require antibiotics for MMC alone.
6. Measure size and determine location of defect, take a picture of the defect and upload into the EPIC media tab
7. Consult Pediatric Neurosurgery and Pediatric Urology
8. **DO NOT BATHE infant**. Diaper should not be near defect, tuck top of diaper as necessary or leave diaper open.
9. Assess degree of neurological involvement with detailed neurologic, skin, and spine exam
10. Monitor for signs of increased intracranial pressure via vital signs, fontanelle exam, and neurologic exam
11. Obtain birth and daily head circumference
12. Insert indwelling catheter and maintain until infant permitted to lie supine (discuss with Pediatric Neurosurgery and Pediatric Urology)
13. Initiate Latex precautions
14. Begin UTI prophylaxis with amoxicillin 10 mg/kg/day (to start after discontinuing initial ampicillin), continue prophylaxis until VCUG is complete and the results are discussed with Pediatric Urology.
15. Follow strict intake and output
16. Imaging:
    a. For open lesions, obtain head ultrasound (HUS) on admission and between 1-3 days post-operatively for baseline
    b. For prenatal closures, obtain HUS on admission
    c. A brain MRI is not necessary in the neonatal period unless there are extenuating circumstances

17. Consult other services (timing may vary) as appropriate: Pediatric Rehabilitation Medicine, Pediatric Orthopedics, Genetics – see information 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
    c. Change mudflap if grossly contaminated
3. Assess surgical site **Qshift** and notify Pediatric Neurosurgery team if there are concerns
4. Infant to remain with HOB 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) on admission
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 PM&R attending will indicate follow-up appointment time in their initial consult.

Pediatric Orthopedics

1. Consult Pediatric Orthopedics prior to discharge to assess for talipes equinovarus and/or dislocated hips and /or fractures of the lower extremities

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 one. The Infant Care Coordinators (ICCs) can facilitate a car bed test.

References:

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5 Adzick, N. S. *et al.* A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med* **364**, 993-1004 (2011). https://doi.org/10.1056/NEJMoa1014379

6 Kessler, T. M., 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* **25**, 758-762 (2006). https://doi.org/10.1002/nau.20304