Meningomyelocele

INTRODUCTION: Failure of closure of the neural tube during the third week of gestation leads to the constellation of defects observed in patients with meningocele (MMC). The open neural tube is continuous with the surface of the skin. For this reason, infants with MMC are at risk for bacterial meningitis due to the spinal defect. Leak of cerebrospinal fluid (CSF) leak is commonly observed. The major indication for early operative repair (within 48h of delivery) is prevention of infection. Although protection of the exposed neural tissue from trauma and drying is essential, the neurological deficit caused by MMC is fixed and rarely improves following repair. Deterioration, however, can occur.

DIAGNOSIS AND PRE-OPERATIVE MANAGEMENT:
• Currently, MMC is usually diagnosed prenatally by ultrasound during the second trimester. Positive screening for maternal serum alpha-fetoprotein may also prompt a fetal ultrasound. A select group of patients are being evaluated for inclusion in a randomized trial between conventional post-natal MMC repair and fetal surgery. Document if the mother is a participant in the clinical trial and, if she is, whether the fetus had prenatal repair and whether the procedure was associated with any complications. Contact the Nurses at the Fetal Treatment Center (476-0445) of the expected delivery; they will inform you whether the mother is in the clinical trial.
• Notify Neurosurgery of the expected delivery.
• Pediatric team should be present for delivery, which will almost always be by cesarean section.
• Use sterile non-latex gloves.
• After birth, position infant on side or on abdomen. Resuscitate as needed. Although all MMC patients have a Chiari II malformation (hindbrain herniation) visualized on MR imaging, only a minority will be symptomatic at birth. This may consist of stridor and upper airway obstruction.
• If infant did not undergo prenatal repair of MMC:
  - Carefully examine MMC to estimate the anatomic level of lesion and whether sac is intact. A small amount of CSF usually ‘weeps’ from the translucent edges of the neural placode. If the sac ruptures, it usually decompresses and drops to the level of the back.
  - Using sterile technique, cover lesion with sterile Telfa™ dressing soaked in bacitracin (50,000 units/ 1,000 mL of 0.9% NaCl) and apply transparent dressing (Do not use Silvadene™, Betadine™, or other anti-infective agents).
  - Once the dressing is in place and if repair is planned within 24-48h, do not change dressing unless it is soiled. If closure is delayed greater than 48h, change the dressing bid and keep it moist with bacitracin solution.
  - Avoid contamination of site and dressing from stool and urine.
• If infant did have prenatal repair of MMC:
  - Examine operative site for evidence of breakdown, leakage of CSF, or inflammation.
- If operative site is well healed, no special wound care is needed.
- If there is leakage of CSF, breakdown of site, or evidence of inflammation, cover lesion with sterile Telfa™ dressing soaked in bacitracin (50,000 units/1,000 mL of 0.9% NaCl) and apply transparent dressing.
- Consult with Neurosurgery and Pediatric Surgery regarding possible need for further surgery and/or special treatment.

• Perform a careful neurological examination to determine the levels of the sensory and motor defects. Note the presence of any orthopedic deformities such as clubfeet.
• Measure the head circumference and look carefully for clinical findings of hydrocephalus. Obtain a baseline head ultrasound. The decision to place a ventricuoperitoneal shunt is individualized for each patient. In general, symptomatic hydrocephalus, progressive increase in head size, or leakage of CSF from the repaired defect site are indications for shunt placement. Usually a shunt is placed several days after the initial repair, although infrequently this may need to be done at the same time as the repair.
• Keep infant flat and either prone or on side.
• Monitor infant closely for signs of meningitis.
• Check with Neurosurgery regarding further investigation (e.g., cranial and abdominal ultrasound, radiograph of spine), timing of surgical repair and whether feedings can be started.

POST-OPERATIVE MANAGEMENT:
• After surgical repair, a dry Telfa™ dressing should be applied to the incision daily or PRN if soiled. Gently clean the incision with sterile normal saline and apply a layer of bacitracin ointment. Place Duoderm™ around the incision and use paper tape to prevent skin breakdown. DO NOT use Tegaderm™ or Opsite™ post-operatively. Observe carefully for signs of wound infection or CSF leak.
• Discuss orders with Neurosurgery regarding positioning of infant, antibiotics, feeding, and timing of post-operative cranial ultrasounds.
• Obtain abdominal and hip ultrasound and request Urology and Orthopedic consults for evaluation of urinary function and associated orthopedic abnormalities.

Prior to discharge, arrange with Neurosurgical Nurse Specialist for patient to be enrolled in Spina Bifida Clinic.