GUIDELINES FOR MANAGEMENT OF NEWBORNS WITH MYELOMENINGOCELE

1. **Providers making the initial prenatal diagnosis**
2. **UW/SCH faculty providing prenatal counseling.**
3. **Obstetric, perinatal and pediatric staff attending delivery.**
4. **Admitting SCH attending.**
5. **Guidelines for NDV consultant and other SCH providers.**

Important phone numbers

SCH paging operator	206-987-7777
SCH NICU desk	206-987-2041
UW NICU	206-598-4606
UW Maternal and Infant Care Clinic (MICC)	206-598-4070
SCH Prenatal Diagnosis and Treatment Program	206-987-6255
Infant OT/PT Team referral line	206-987-2113
PCC Alice Crandall alice.crandall@seattlechildrens.org	206-987-3371
SCH Infant Transport Team	206-987-2632
Neurodevelopmental Clinic	206-987-2210
Neurodevelopmental Clinic Nurses	206-987-2184

Background: These guidelines, based on a combination of evidence, expert opinion and clinical experience, are intended to assist healthcare providers to provide optimal care to babies with myelomeningocele, as well as pregnant women carrying fetuses with myelomeningocele. In addition, several SCH policies address the surgical and nursing management of patients with myelomeningocele. These complementary documents are located in the hospital policy section of the CHILD website at SCH.

Most fetuses with myelomeningocele and hydrocephalus are now recognized by prenatal diagnosis. Ideally, women carrying fetuses with myelomeningocele should receive prenatal counseling from both an obstetrician/perinatologist and a postnatal provider experienced in the care of babies with myelomeningocele. The SCH NDV program provides comprehensive prenatal counseling for myelomeningocele through the SCH Prenatal Diagnosis and Treatment Program (206-987-6255). This counseling includes information on the pathophysiology of myelomeningocele, the MOMS trial of in utero closure surgery, the expected pregnancy course, the recommended delivery route, associated medical complications, medical/surgical treatment, recurrence risk, prevention with high-dose folic acid and prognosis with respect to mobility, cognition, bladder/bowel function, independence and quality of life. When appropriate, reproductive decision-making support is provided.

While not universally accepted, our program feels that cesarean section delivery prior to rupture of membranes improves motor outcomes for fetuses with lumbosacral lesions that protrude >1cm off the back and expectation of somewhat intact lower extremity motor function. Cesarean section is unlikely to be of benefit after rupture of membranes, for fetuses with flat lesions and for fetuses without lower extremity movement.

All newborns with myelomeningocele benefit from sterile management and preparation for closure of their open lesions within 24 to 48 hours of birth. A serious breach of sterility may be a reason to defer surgical treatment, although this is rare. The presence of other life-threatening anomalies may be reason to defer treatment indefinitely.
As of February 2003, a national multicenter randomized trial of in utero versus postnatal myelomeningocele repair began recruiting patients. Three centers around the country are involved in the study (Vanderbilt, Children's Hospital of Philadelphia, and University of California at San Francisco). No other center in the US will provide in utero repair outside the randomized trial. Participants are

delivered by C-section without a trial of labor at one of the study sites. For more details, see the study website: www.spinabifidamoms.com.

Fetuses with skin-covered lesions such as lipomeningoceles or myelocystoceles do not routinely require cesarean section delivery. Such fetuses can often be recognized by the absence of any brain involvement (no hydrocephalus, no Chiari II malformation), the location and characteristics of the mass itself and/or the presence of abdominal wall defects such as exstrophy of the cloaca that do not occur with myelomeningocele. Negative amniotic alpha fetoprotein (AFP) and/or acetylcholinesterase testing can also distinguish skin covered lesions.

Neonates with skin-covered lesions do not need to be managed under sterile conditions and usually do not require immediate neurosurgical repair. Neurosurgery and Neurodevelopmental should be consulted about these infants who may require additional work-up and/or monitoring for neurogenic bowel and bladder. Neurosurgical intervention usually occurs at 4-8 months of age, unless there is progressive neurological impairment. Recurrence risk for isolated skin covered lesions tends to be lower than for myelomeningocele and folic acid supplementation is not preventive.

1) Guidelines for the obstetric provider who is the first to become aware of a patient carrying a fetus with myelomeningocele

- ☑ Ensure that the patient is offered appropriate diagnostic work-up including serial ultrasounds to confirm the defect and evaluate for other findings as well as amniocentesis for karyotype, AFP and acetylcholinesterase.
- ☑ Ensure that the patient receives appropriate counseling, ideally from both an obstetrician/perinatologist and a postnatal provider experienced in the care of babies with myelomeningocele. The SCH NDV program (usually Dr. Doherty) provides comprehensive prenatal counseling for myelomeningocele through the SCH Prenatal Diagnosis and Treatment Program (206-987-6255).
- ☑ Ensure that medical records, imaging, and laboratory data are made available to the consultants

2) Guidelines for UW/SCH faculty providing prenatal counseling

- ☑ Provide counseling regarding the pathophysiology of myelomeningocele, the MOMS trial of in utero closure surgery, the expected pregnancy course, the recommended delivery route, associated medical complications, medical/surgical treatment, recurrence risk, prevention with high-dose folic acid and prognosis with respect to mobility, cognition, bladder/bowel function, independence and quality of life. When appropriate, provide reproductive decision-making support.
- ☑ Usually, counseling is performed through the Prenatal Diagnosis and Treatment Program in the Springbrook Professional Center near SCH (206) 987-6255. The Prenatal Program staff have experience coordinating prenatal and postnatal care and can assist in all tasks described in this section.
- ☑ Upon completion of the prenatal counseling session, the patient will be added by the Prenatal Clinic staff to the Prenatal Forecast Tool. The Forecast Tool is a spreadsheet distributed electronically every Monday to provider groups, including the NDV attendings, NDV clinic nurses, Neurosurgery ARNPs, MCC Service ARNPs, MCC Service Medical Director, NICU charge nurses, and neonatologists. The Forecast Tool contains the pregnant woman's SCH MR#, anticipated delivery date, delivery hospital, delivery route, fetal findings, prenatal consult date and provider name.
- ☑ If the delivery date is not known at the time of prenatal consultation, contact the Prenatal Clinic at 206-987-6255 once delivery date is finalized. The date will be added to the Forecast Tool.
- ☑ If the child will be delivered within three working days, contact the attendings on call for Neurosurgery, NDV and Neonatology by phone, in addition to notifying the Prenatal Clinic.

3) Guidelines for the providers (obstetrics, neonatology) attending delivery

In addition to the usual care of a newborn, the following measures are required for infant with myelomeningocele:

- ☑ Handle the infant with sterile, non-latex gloves and with sterile clothing and sheets.
- ☑ Institute latex allergy precautions.
- ☑ Dress the lesion to minimize the chance of physical damage and infection
  - o Cover the lesion with non-adhesive dressing wet with sterile Ringer's lactate solution or saline.
  - o Fill a syringe with the same solution, add a sterile, silastic tube to the tip of the syringe with the syringe taped to the baby's back and the distal tip of the tube in the center of the dressing.
  - o Cover with a sterile dressing surrounded by a ring (doughnut) of Kerlix to prevent pressure on the sac.

  Note: Packaged materials and instructions for the dressing described above are available from the SCH NICU Charge Nurse (206) 987-2477. A detailed set of instructions with illustrations is available at http://neonatal.peds.washington.edu/NICU-WEB/mcelecov.stm and in the Policies and Procedures section of the internal Seattle Children's Hospital website CHILD (Stabilization of the Unrepaired Myelomeningocele Lesion).
- ☑ Keep the patient in side-lying or prone position to avoid pressure on the sac or nerves.
- ☑ If the patient will be transferred to Seattle Children's Hospital (SCH), notify the Neurodevelopmental, Neurosurgery and Neonatology attendings on call (physician paging operator: 206-987-7777).
- ☑ Term newborns with myelomeningocele should be transferred to the SCH NICU whenever possible, to ensure that they are ready for back closure within 48 hours after birth. If the NICU is full, stable patients can be admitted to the regular ward on the Medically Complex Child Service, or, if full, on a ward team with a NDV or house attending.
- ☑ Ensure that the prenatal counseling summary is in the infant's transfer paperwork. It is in the SCH electronic medical record under the mother's name and MR#.

4) Guidelines for the admitting SCH attending

- ☑ Term newborns with myelomeningocele should be admitted to the NICU whenever possible to ensure that they are ready for back closure within 48 hours after birth. If the NICU is full, stable patients can be admitted to the regular ward on the Medically Complex Child Service, or, if full, on a ward team with an NDV or house attending.
- ☑ The NDV Meningomyelocele Admit Orderset should be used for all new patients with myelomeningocele. Pre-op work-up includes cranial ultrasound, renal ultrasound, echocardiogram, OT/PT spina bifida assessment as well as consults from Neurosurgery, NDV and Plastic Surgery.
- ☑ Keep the patient prone or side-lying, unless supine positioning is required for emergent treatment.
- ☑ Monitor closely for Chiari II symptoms including apnea, bradycardia, hypoventilation, stridor and swallow dysfunction. Notify the Neurosurgery and NDV attendings about concerning symptoms and consider a brain MRI, sleep study, clinical feeding assessment by OT, videofluoroscopic swallow study and/or ENT evaluation.
- ☑ Provide meticulous skin care for the perineum in the presence of dribbling urine or stool.
- ☑ See additional information in the next section.

5) Guidelines for NDV consultant and other SCH providers (residents and attendings).

Coordination of care:

- ☑ Confirm that the NICU, Neurosurgery and Plastic Surgery (if appropriate) attendings have been informed about the patient.
- ☑ Confirm that the admitting resident knows to use the NDV Meningomyelocele Admit Orderset.
- ☑ Discuss the need for pre-op antibiotic treatment and head CT with the neurosurgical team.

- ☑ Ensure that OT/PT has been consulted and notified by phone at the Infant OT/PT Team Referral Line 206-987-2113.
- ☑ Notify the NDV nurses at 206-987-2184. The NDV nurses can provide the floor staff nurses with consultation and instructions as to the infant's needs.

Evaluation:
- ☑ Examine the infant for other malformations and syndromes that could affect the prognosis or management plan.
- ☑ Record information about lower extremity muscle strength and sensory level.
- ☑ Discuss the management plan with the medical team.

Social:
- ☑ Spend adequate time with family to discuss overall care, answer questions and develop a therapeutic alliance.
- ☑ Confirm that the family has been given: 1) "Living with Spina Bifida: A Guide for Families and Professionals" book by Adrian Sandler, MD (University of North Carolina Press), 2) the Hydrocephalus pamphlet published by the Hydrocephalus Association, 3) a SCH Care Notebook and 4) the NDV folder with additional information about myelomeningocele. If parents do not have these items, notify the NDV nurses or social worker who can provide them.
- ☑ Confirm that social work is involved. For families who may qualify based on income, encourage application for SSI (Supplemental Security Income) as soon as possible.

Neurogenic bladder:
- ☑ An indwelling catheter is preferred during the acute post-closure period and may be required beyond that time in certain clinical situations.
- ☑ Clean intermittent catheterization (CIC) should be implemented 2-3 times daily, as soon as the patient is stable enough. A true post-void residual is obtained by CIC within a few minutes of a detected void and should be less than 5cc. Random CIC volumes should be less than 30cc.
- ☑ Parents should be trained to perform CIC, as early as possible since children may be discharged home on CIC.
- ☑ Urology consultation should occur prior to discharge.
- ☑ Prophylactic antibiotics, Ditropan and phenoxybenzamine are not be routinely used in the bladder management of newborns with myelomeningocele.

Hydrocephalus:
- ☑ Monitor daily head size, plotted on an appropriate head-size chart and ensure that follow-up cranial ultrasounds are performed (typically twice weekly in unshunted infants). In premature infants, the brain is more compliant so the ventricles may expand with little change in head size.
- ☑ For patients with shunts, ensure appropriate positioning to prevent skin breakdown over the shunt valve or chamber (have a doughnut under the head.)

Other:
- ☑ Ensure that nutrition has been consulted to optimize nutritional status.
- ☑ Ensure that a neonatal hearing evaluation is performed.
- ☑ If the patient has club feet or other musculoskeletal abnormalities, confirm that outpatient orthopedic follow-up is scheduled prior to discharge.
- ☑ Give recommendations about discharge needs (such as catheters) and outpatient NDV follow-up.
- ☑ Notify the NDV/Birth Defects patient care coordinators, preferably in writing or by e-mail, of the plan for clinic appointments. These appointments need to be tailored to the needs of each infant and family. Work with the discharge planners and others unfamiliar with our clinic to minimize uncoordinated appointments in many separate clinics.

For questions or feedback on this document, please contact Dan Doherty and/or Bill Walker. In addition, any of the following faculty will be glad to answer questions about specific patients: Jeff

McLaughlin, William Walker, Chuck Cowan, Dan Doherty, Gwen Glew or Sam Zinner. Contact the SCH paging operator (206-987-2000) to determine who is on call.

Doctor	Phone Number	Pager Number	Email Address
Chuck Cowan	206-987-2210	206-469-5369	charles.cowan@seattlechildrens.org
Dan Doherty	206-987-2210	206-540-5892	dan.doherty@seattlechildrens.org
Gwen Glew	206-987-2210	206-469-0668	gwen.glew@seattlechildrens.org
Jeff McLaughlin	206-987-2210	206-469-6704	john.mclaughlin@seattlechildrens.org
William Walker	206-987-2210	206-469-3579	william.walker@seattlechildrens.org
Samuel Zinner	206-685-1290	206-469-5157	szinner@u.washington.edu

Disclaimer: These guidelines have been developed by the Division of Developmental Medicine, Department of Pediatrics, UW, to assist physicians and other healthcare professionals. The recommendations may not be appropriate for use in all circumstances, and any decision to adopt a particular recommendation must be made by the practitioner based upon available facts and circumstances presented by individual patients.

Distribution:

Developmental Medicine attendings, nurses, fellows, PCCs
Developmental Medicine website c/o CHDD webmanager
Neurosurgery attendings, ARNPs, residents, PCCs
Neonatology: Dr. Craig Jackson & website
NICU head nurse, SCH
NICU head nurse, UW
Prenatal Diagnosis and Treatment Program, SCH
Maternal and Infant Care Clinic, UW
SCH Housestaff office and website
OT/PT Department, SCH
Director, Surgery, SCH: Dr. Robert Sawin