

## **GUIDELINES FOR MANAGEMENT OF A NEWBORN WITH MENINGOMYELOCELE**

**Note:** This document contains guidelines for five different types of encounters regarding management of meningocele:

- 1. Initial prenatal diagnosis by physicians or other providers of care for the mother.**
- 2. UW/CHRMC faculty providing prenatal counseling regarding management.**
- 3. Obstetric, perinatal and pediatric staff attending delivery.**
- 4. Infant ICU staff at CHRMC.**
- 5. UW/CHRMC Neurodevelopmental/Birth Defects Attending Initial and Followup.**

**Background:** These guidelines are based on a combination of evidence, expert opinion and clinical experience. New information may come along at any time. We will do our best to update these guidelines and welcome input.

Most fetuses with meningocele and hydrocephalus are now recognized by prenatal diagnosis. Prenatal diagnosis allows time for counseling and thoughtful consideration by each family about their decision and plan for each embryo or fetus. These guidelines are intended to assist health professionals in optimally managing the meningocele in fetuses whose parents have chosen to continue pregnancy.

As of February 2003, a national multicenter randomized trial of in utero meningocele repair began recruiting patients. Three centers around the country are involved in the study (Vanderbilt, Childrens Hospital of Philadelphia, and University of California at San Francisco). No center in the country will provide in utero repair outside the randomized trial in hopes of collecting data as rapidly as possible regarding the relative efficacy of in utero repair at 19-25 weeks compared to term. Infants delivered at or near term will be 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](http://www.spinabifidamoms.com) (good as of March 28, 2008).

All newborns with meningocele 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. The presence of other life-threatening anomalies may be reason to defer treatment indefinitely.

The evidence for a better motor level and ambulatory outcome following prenatal diagnosis, and delivery by C-section without rupture of membranes in fetuses that have a bulging lumbosacral meningocele and in utero motor function in the affected spinal cord segments is primarily derived from the ongoing series accumulated by our own group. These fetuses must be distinguished from those who have either a flat meningocele lesion or a large lesion with no in utero motor function below T12; these meningocele types often do not benefit from C-section delivery.

It is also important to recognize and distinguish fetuses with secondary neural tube defects: skin-covered lesions such as lipomeningoceles or myelocystoceles. 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 meningomyelocele. Such lesions may, on occasion, be large enough to require C-section because of obstruction of labor. There is, however, no evidence that C-section delivery is needed routinely or that any benefit will accrue in terms of motor function. Neonates with secondary (skin-covered) neural tube defects do not need to be managed under sterile conditions and usually do not require immediate neurosurgical closure.

The usual management approach is for one of the Neurodevelopmental/Birth Defects pediatric attendings to see the neonate on consultation in the newborn nursery to confirm a diagnosis and document other anomalies that may need attention. A Neurodevelopmental/Birth Defects pediatrics attending will then see the child in clinic at about two months of age in conjunction with Neurosurgery with plans to surgically intervene when the child is 4 to 8 months of age.

It is important to recognize that counseling about recurrence risks is totally different for secondary neural tube defects and that folic acid supplementation is not protective. As folic acid supplementation and prenatal diagnosis/termination reduce the incidence of meningomyelocele, but not secondary neural tube defects, the latter will occupy our attention more of the time. Ron Lemire was a world expert on the embryology and occurrence of secondary neural tube defects and he has left us a legacy of considerable experience in managing them. If there is ever a question about these issues, please feel free to contact the Neurodevelopmental/Birth Defects and Neurosurgery faculty.

### **1.) Guidelines for the obstetric provider who is the first to become aware of a fetus with meningomyelocele**

- Arrange for counseling, including information about fetal repair, delivery by C-section and other maternal care issues if the family is interested in continuing the pregnancy. This can be done through the Prenatal Genetics Clinic at UW (phone number 206-598-8130) or another perinatal center convenient to the family. Patients may be referred from other institutions directly to pediatric faculty in the Neurodevelopmental/Birth Defects program (usually Drs. Doherty or Shurtleff) for discussions that focus on the outcome of the infant in relation to obstetrical management and developmental outcome. Families wishing to obtain more information about the postnatal care and prognosis for children with meningomyelocele can also be referred to the Neurodevelopmental/Birth Defects program at CHRMC: 4800 Sand Point Way NE, A7938, Seattle, WA 98105, 206-987-2210. Some families may wish to have prenatal counseling with Neonatology, Dr. David Woodrum (206-543-3200) regarding relevant aspects of care.
  
- Collect all ultrasound data. Repeat studies as needed.

- ☑ Collect all available data regarding prenatal testing (AFP, other tests). A fetal karyotype is essential for complete counseling.
- ☑ Referrals are accepted from any referring obstetric provider, the patient's neurosurgeon or their primary care physician. Prepare a dictated summary in the medical record including the above information, clinical observations, and approximate EDC for the mother's chart and send to the Neurodevelopmental/Birth Defects office: CHRMC, 4800 Sand Point Way NE, A7938, Seattle, WA 98105. Phone 206-987-2210. Fax 206-987-3824.

**2.) Guidelines for UW/CHRMC faculty providing prenatal counseling**

- ☑ Send email notice to all the NDV attendings, the NDV Clinic nurses, the Neurosurgery ARNPs, the MCC service ARNPs, the MCC Service Medical Director (Ron Dick) and the NICU charge nurse (Sue Prueitt). The email should include the mother's name, her Children's hospital number, delivery route, delivery hospital and delivery date (when known), consult note date and provider, and a brief description of fetal findings.
- ☑ If delivery of the child will occur within three working days, contact the specific attendings on call for Neurosurgery, Neurodevelopmental/Birth Defects, and Neonatology.

**3.) Guidelines for the team of physicians (obstetrics, neonatology) attending birth**

- ☑ Notify BOTH the Neurodevelopmental/Birth Defects and Neurosurgery attendings on call at CHRMC (physician paging operator: 206-987-7777) when the delivery is scheduled; time and place of Cesarean section. This notification should occur even if the expected treatment plan is not to intervene. Many children who are not expected to survive follow a very different clinical course than the prenatal expectation. If a newborn is delivered with a previously unrecognized meningocele, please contact the Neurodevelopmental/Birth Defects and Neurosurgery attendings at the time the lesion is discovered. Please provide relevant medical history on the baby and the mother.
- ☑ Handle the infant with sterile, non-latex gloves and with sterile clothing and sheets.
- ☑ Institute latex allergy precautions.
- ☑ Cover the lesion with non-adhesive dressing wet with sterile Ringer's lactate solution or saline.
- ☑ 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.

- ☑ Cover with a sterile dressing surrounded by a ring of Curlex to prevent pressure on the sac.

Note: Packaged materials and instructions for the dressing described above are available from Patti Jason, RN, Director, CHRMC Transport Team, (206) 987-2632, pager 469-6336. There is also a more detailed set of instructions complete with illustrations on the Neonatology Division website at <http://neonatal.peds.washington.edu/NICU-WEB/mcelecov.stm>. Anyone unable to directly access this website could ask the IICU unit coordinators to print a copy.

- ☑ Position the infant in the side lying or prone positions only; avoid pressure on the sac or nerves.
- ☑ Arrange transport through normal channels for transport of critically ill newborns. Timing of transport depends on the stability of the infant and the availability of bed space. Notify the Neurodevelopmental/Birth Defects attending on call (paging operator at CHRMC 206-987-7777) for direct physician to physician communication on an urgent basis.
- ☑ On transfer to CHRMC, pre-term or unstable infants will be admitted to the Infant ICU. Term and stable (most) infants should be admitted to the ward on the Medically Complex Child Service. If the MCC service is capped, the team should consult with the NDV attending on call about the most appropriate attending assignment for the patient: general medicine or Neurodevelopmental.
- ☑ Ensure that the prenatal counseling summary is on the infant's chart.
- ☑ Add and complete an appropriate growth chart for head OFC, height and weight.

#### 4.) **Guidelines for the CHRMC Infant ICU Attending on Call**

- ☑ Term newborns with meningomyelocele can be admitted to the regular ward on the Medically Complex Child Service, or, if full, on the Neurodevelopmental/Birth Defects service.
- ☑ There is a meningomyelocele order set in CIS that should be used for all patients.
- ☑ Any newborn exhibiting a clinical picture of perinatal asphyxia should be evaluated for severe brainstem dysfunction associated with a Chiari II malformation since the initial presentation is quite similar.
- ☑ Preterm newborns with meningomyelocele usually need IICU respiratory care

in the same manner as other prematures. It is best if they can be nursed prone, or at least side-lying, but emergency intubation or related procedures may require a brief period of time supine. Try to keep the open sac sterile and protected from pressure if possible. Preterm newborns with meningocele are more likely to have congenital heart disease and/or syndromic conditions (e.g. Trisomy 13 or 18) than term newborns.

- ☑ Contact the Neurodevelopmental/Birth Defects attending on call about the infant after stabilization if no direct contact has occurred prior to the infant's arrival. Ask the IICU resident to initiate a formal consult request in CIS.

### **5.) Guidelines for Neurodevelopmental/Birth Defects Attending on Call**

- ☑ Notify Admitting for admission to the medical floor unless the infant is unstable and requires admission to the IICU and provide the name of the neurosurgeon on call and your name with how to contact each. Request a call from the ward clerk as soon as the infant arrives on the floor.
- ☑ Notify neurosurgical service and identify yourself and how to contact you. In general, the children will initially be on the Medically Complex Child Service or the Neurodevelopmental/Birth Defects service, then on Neurosurgery post operatively (typically one or two days) and then back to the original service. It is expected that the two services will work collaboratively at all times, regardless of who is the current attending.
- ☑ Evaluate the infant for contraindications to surgery, confirm or modify the prenatal prognosis, and advise on treatment of other issues, etc.

Note: The IICU resident may need to be reminded to write an order for a Neurodevelopmental/Birth Defects consultation. The IICU attendings have always appreciated contributions to management from the Division. In view of current billing and documentation issues, a documented consult request is important. It is even more important to take the initiative to go see the child regardless of consult status.

- ☑ Examine the infant for other malformations and syndromes that could alter the prognosis or management plan.
- ☑ Record muscle strength examination and sensory level.
- ☑ There is a meningocele order set in CIS that should be used for all patients. The details discussed in this section are in the order set but are reviewed here.
- ☑ Request PT and OT meningocele evaluation consults via online orders from the Infant OT/PT Team Referral Line and leaving an email at:  
Alice Crandall, PCC, Rehabilitation Medicine, available 8:00 am to 5:00 pm

[alice.crandall@seattlechildrens.org](mailto:alice.crandall@seattlechildrens.org) (206) 987-3371

It is sure to cause delay in involving OT/PT if direct contact is not made.

Contact information for each Infant OT/PT Team member, *use only after contacting Referral Line in OT/PT at 206-987-2113*:

Lynn Wolf [lynn.wolf@seattlechildrens.org](mailto:lynn.wolf@seattlechildrens.org) 206-987-3131

Gail Bonato [gail.bonato@seattlechildrens.org](mailto:gail.bonato@seattlechildrens.org) 206-987-1320

Jane Mason [jane.mason@seattlechildrens.org](mailto:jane.mason@seattlechildrens.org) 206-987-3130

Susan Hutchinson [susan.hutchinson@seattlechildrens.org](mailto:susan.hutchinson@seattlechildrens.org) 206-987-1467

Nan Street [nan.street@seattlechildrens.org](mailto:nan.street@seattlechildrens.org) 206-987-3148

- Notify Neurodevelopmental/Birth Defects Nurse Line (206-987-2184). The nurses are available from 8:00 am to 4:00 pm Monday through Friday. The Neurodevelopmental/Birth Defects nurse can provide the floor staff nurses with consultation and instructions as to the infant's needs. The floor nurse, medical team coordinator or Neurodevelopmental/ Birth Defects nurses will provide the family with information on how to obtain a Care Notebook and information about meningomyelocele.
- Discuss the management plan with the house staff.
- Institute latex allergy precautions. See hospital protocol.
- Obtain an ultrasound examination of urinary tract, head for ventricle size, and an echocardiogram on all patients before surgery. Cardiology consultation should be requested, if possible, prior to closure surgery if an anomaly is present.
- Request urology and orthopedic consultations if needed. Virtually all infants will have a neurogenic bowel and bladder and will need management during the newborn hospitalization. Many infants will have musculoskeletal deformities requiring orthopedic consultation, though rarely on an emergent basis.
- Request social work consultation. The social worker assigned to the Medically Complex Child service and Neurodevelopmental/Birth Defects service is available weekdays through the paging operator (206-987-2000).
- If the infant presents with cardio-respiratory distress, apnea, bradycardia, laryngeal stridor or aspiration with swallowing, institute a symptomatic Chiari II malformation work-up, which may include blood gases and oximetry, an MRI of the brainstem, sleep study, clinical feeding assessment by OT, videofluoroscopy of swallowing and ENT evaluation. The workup should be individualized. Neurosurgery should already be following the case.
- Spend adequate time with family explaining all aspects of care to inform them and begin to develop the therapeutic alliance that will allow optimal care of the

child for the next 21 years.

- ☑ Assure that basic information on each infant is submitted to the myelodysplasia database (PDMS). The OTs, PTs and nurses should have the permanent information data entry form if they are notified about the infant. A backup procedure is to notify Dr. Shurtleff (206-987-2058, e-mail david.shurtleff@seattlechildrens.org) or Sharon Duguay (e-mail sharon.duguay@seattlechildrens.org) directly.
- ☑ Any of the following faculty will be glad to answer questions: Jeff McLaughlin, William Walker, Chuck Cowan, Dan Doherty, Gwen Glew or Sam Zinner. Contact the CHRMC paging operator (206-987-2000) to determine who is on call.

Doctor	Phone Number	Pager Number	Email Address
Jeff McLaughlin	206-987-2210	206-469-6704	<a href="mailto:john.mclaughlin@seattlechildrens.org">john.mclaughlin@seattlechildrens.org</a>
Gwen Glew	206-987-2210	206-469-0668	<a href="mailto:gwen.glew@seattlechildrens.org">gwen.glew@seattlechildrens.org</a>
William Walker	206-987-2210	206-469-3579	<a href="mailto:william.walker@seattlechildrens.org">william.walker@seattlechildrens.org</a>
Chuck Cowan	206-987-2210	206-469-5369	<a href="mailto:charles.cowan@seattlechildrens.org">charles.cowan@seattlechildrens.org</a>
David Shurtleff	206-987-2058	206-469-5485	<a href="mailto:david.shurtleff@seattlechildrens.org">david.shurtleff@seattlechildrens.org</a>
Samuel Zinner	206-685-1290	206-469-5157	<a href="mailto:szinner@u.washington.edu">szinner@u.washington.edu</a>
Dan Doherty	206-987-2210	206-469-6331	<a href="mailto:dan.doherty@seattlechildrens.org">dan.doherty@seattlechildrens.org</a>

#### 6.) Guidelines for Neurodevelopmental/Birth Defects Attending Follow-up

- ☑ Consult with neurosurgeons as to antibiotic management, nutrition and any other appropriate medical considerations during the hospitalization.
- ☑ The infants will come to the Medically Complex Child Service, or, if that service is capped, to the Neurodevelopmental/Birth Defects Service. NDV / BD attendings should provide consultation to the MCC team. Preferable placement is with nurses experienced in the management of newborns with neural tube defects.
- ☑ A Renal/Bladder Ultrasound will already have obtained as part of the preoperative evaluation. A VCUG is not required prior to discharge regardless of the findings on the renal / bladder ultrasound. Follow urine post voiding residual for temporary or permanent postoperative retention or infection. A true post-void residual obtained by in and out catheterization within a few minutes of a detected spontaneous void should be less than 5cc. A random check of bladder volume in a newborn should always be less than 30 - 45cc. 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 once the indwelling catheter is removed. Parents should be trained in this technique as early as possible as all children will be discharged home on some frequency of

daily CIC. Prophylactic antibiotics, Ditropan and Phenoxybenzamine are not be routinely used in the bladder management of newborns with meningomyelocele.

- ☑ Organize discharge plan for parents and the local health care provider. Begin early in the hospitalization and involve the unit / team discharge planner if complexity warrant (e.g. need for home health care, special equipment, etc).
- ☑ Follow carefully for positioning to prevent head weight bearing on the shunt valve or chamber (have a doughnut under the head.)
- ☑ Manage skin care for perineum in the presence of dribbling urine or stool.
- ☑ Record daily head size, plot on appropriate head size chart and obtain follow-up cranial ultrasounds (typically twice weekly in unshunted infants). In premature infants, the brain is more compliant and ventricles may expand with little change in head size!
- ☑ Every infant should have a nutrition consult.
- ☑ Every infant should have a neonatal hearing evaluation.
- ☑ Notify the Neurodevelopmental/Birth Defects patient care coordinators, preferably in writing or by e-mail, of the plan for clinic appointments. These 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.

These guidelines were developed with the assistance of many people, notably David Shurtleff, MD. Please send any questions or suggestions about these guidelines to Jeff McLaughlin, MD at [john.mclaughlin@seattlechildrens.org](mailto:john.mclaughlin@seattlechildrens.org) or 206.987.2210 or page.

In addition, there are several policies regarding surgical and nursing management of meningomyelocele in the hospital policy section of the CHILD website at CHRMC. These guidelines complement each other.

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

**It is not the intention in promulgating these guidelines to interfere with the provider/patient relationship, nor are these guidelines intended to represent the standard of care in any given circumstance. These guidelines are recommendations to be used at the sole discretion of the provider and are not meant to dictate the**

**manner or style of clinical practice employed in rendering services to a particular patient.**

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