

GUIDELINES FOR MANAGEMENT OF A NEWBORN WITH MENINGOMYELOCELE

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

As of February 2003, a national multicenter randomized trial of in utero meningomyelocele repair began recruiting patients. Three centers around the country with experience are involved in the study (Vanderbilt, Childrens Hospital of Philadelphia, University of California at San Francisco). No center in the country plans to 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 (good as of June 3, 2005).

All newborns will benefit from sterile management and consideration for closure of their open lesions within 24 to 48 hours of birth. A serious breach of sterility may be a reason to defer 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 a trial of labor in fetuses who have a bulging lumbosacral meningomyelocele 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 meningomyelocele lesion or a large lesion with no motor function below T12. These other types of myelomeningocele often do not benefit from C-section delivery.

It is also important to recognize and distinguish fetuses who have secondary neural tube defects. These individuals have skin-covered lesions. Such lesions may be large enough on occasion to require C-section because of obstruction of labor, but there is no evidence that C-section delivery is needed routinely or that any benefit will accrue in terms of motor function. 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 extrophy of the cloaca that do not occur with meningomyelocele. Neonates with secondary (skin-covered) neural tube defects do not need to be managed under sterile conditions and usually do not need immediate neurosurgical closure. The usual

management is to see the neonate on consultation in the newborn nursery to make a diagnosis, and look for other anomalies that may need attention. We see the child back in clinic at about two months of age with plans to surgically intervene when the child is 4 to 8 months of age. It is also very important to recognize that the 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 is a world expert on the embryology and occurrence of secondary neural tube defects and we have developed considerable experience in managing them. If there is ever a question about these issues, please feel free to contact the Neurodevelopmental/Congenital Defects and Neurosurgery faculty on the team.

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/Congenital Defects program (usually Dr. Shurtleff) for discussions that focus on the outcome of the infant in relation to obstetrical management. Families wishing to obtain more information about the postnatal care and prognosis for children with meningomyelocele can also be referred to the pediatric faculty in the Neurodevelopmental/Congenital Defects program at CHRMC: 4800 Sand Point Way NE, M2-8, Seattle, WA 98105, 206-987-2204. Some families may wish to have prenatal counseling with Neurosurgery (206-987-2544) or Neonatology, Dr. David Woodrum and others (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.
- Prepare a dictated/written summary including clinical observations, the above information and approximate EDC for the mother's chart, baby's record (if started), parents, and send to the Neurodevelopmental office at CHRMC, 4800 Sand Point Way NE, M2-8, Seattle, WA 98105, 206-987-2204, the patient's neurosurgeon, any referring obstetric provider or primary care physician, and send to the Infant OT/PT team in the Rehabilitation Dept. at CHRMC, 4800 Sand Point Way NE, 6I-1, Seattle, WA 98105, 206-987-2114.

2.) Guidelines for UW/CHRC faculty providing prenatal counseling

- Ask the transcriptionist to send copies of the consultation to ALL the Neurodevelopmental/Congenital Defects and Neurosurgery attendings at CHRC. Alternatively, send email notice with the mother's name and hospital number if the consult is generated at CHRC. (Please note the names of the Neurodevelopmental and Neurosurgery attendings in dictation.)
- If delivery of the child will occur within two weeks, contact the specific attendings on call for Neurosurgery, Neurodevelopmental/Congenital Defects, and Neonatology.

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

- Notify BOTH the Neurodevelopmental/Congenital Defects and Neurosurgery attendings on call at CHRC (physician paging operator: 206-987-7777) when the baby is scheduled for delivery; time and place of Cesarean section. If the newborn has a previously unrecognized meningomyelocele, please contact the Neurodevelopmental/Congenital Defects and Neurosurgery attendings at the time the lesion is discovered. Please provide relevant medical history on the baby and the mother.
- The infant should be admitted to the Neurodevelopmental/Congenital Defects service on the ward if term and stable (most infants) or to the Infant ICU (IICU) if pre-term or unstable.
- 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, CHRC 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 position 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/Congenital Defects attending on call (paging operator at CHRMC (206-987-7777 for direct physician to physician communication on an urgent basis).
- ☑ Add the prenatal counseling summary to 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 Neurodevelopmental/Congenital Defects service. Any such newborn billed as having had perinatal asphyxia should be evaluated for severe brainstem dysfunction associated with 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 meningomyelocele are more likely to have congenital heart disease and/or syndromic causes (eg Trisomy 13 or 18) than term newborns.
- ☑ Contact the Neurodevelopmental/Congenital 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 write a consult request.

5.) Guidelines for Neurodevelopmental/Congenital 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.

Note: The IICU resident may need to be reminded to write an order for a Neurodevelopmental/Congenital Defects consultation. The IICU attendings have always appreciated contributions to management from the Division. Dr Lemire recommends we provide the same management recommendations as if

on our service and not wait to be consulted. In view of current billing and documentation issues, a documented consult request is important.

- ☑ Notify floor and request a call as soon as the infant arrives. Evaluate the infant for contraindications to surgery, confirm or alter the prenatal prognosis, advise on treatment of other issues, etc.
- ☑ Notify neurosurgical service and identify yourself and how to contact you. In general, the children will be on the medical Neurodevelopmental/Congenital Defects service initially, then on Neurosurgery perioperatively (typically one or two days) and then back on Neurodevelopmental/Congenital Defects service. It is expected that the two services will work collaboratively at all times, regardless of who is attending at the moment.
- ☑ 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 Psychology, available 8:00 am to 5:00 pm 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:*

Lynn Wolf lynn.wolf@seattlechildrens.org 206-987-3131

Gail Bonato gail.bonato@seattlechildrens.org 206-987-1320

Jane Mason jane.mason@seattlechildrens.org 206-987-3130

Susan Hutchinson susan.hutchinson@seattlechildrens.org 206-987-1467

Nan Street nan.street@seattlechildrens.org 206-987-3148

- ☑ Record muscle strength examination and sensory level.
- ☑ Notify Neurodevelopmental/Congenital Defects Nurse (206-987-2184). The nurse is available from 8:00 am to 4:00 pm Monday through Friday. The Neurodevelopmental/Congenital 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/ Congenital Defects nurses will provide the family with information on how to obtain a Care Notebook and information about meningocele.
- ☑ Examine the infant for other malformations and syndromes that will alter the prognosis.
- ☑ 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 early if an anomaly is present.

- ☑ Request urology and orthopedic consultations if needed. Most infants will have a neurogenic bowel and bladder and will need urology 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 Neurodevelopmental/Congenital Defects 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. The OTs, PTs and nurses should both have the permanent information data entry form to initiate if they are notified about the infant. A backup procedure is to notify Dr. Shurtleff (206-987-2058, 425-454-1893, 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, Nora Davis, William Walker, Chuck Cowan, David Shurtleff, Dan Doherty 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	206-987-2204	206-469-6704	john.mclaughlin@seattlechildrens.org
Nora Davis	206-987-2590	206-469-6411	nora.davis@seattlechildrens.org
William Walker	206-987-2204	206-469-3579	william.walker@seattlechildrens.org
Chuck Cowan	206-987-2204	206-469-5369	charles.cowan@seattlechildrens.org
David Shurtleff	206-987-2058	206-469-5485	david.shurtleff@seattlechildrens.org
Samuel Zinner	206-685-1290	206-469-5157	szinner@u.washington.edu
Dan Doherty	206-987-2489	206-469-6331	dan.doherty@seattlechildrens.org

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

- ☑ Consult with neurosurgeons as to antibiotic management, nutrition and any

other appropriate medical considerations.

- ☑ Accept the infant in transfer if requested by the neurosurgeons or IICU staff. Preferable placement is on a medical floor with nurses experienced in the management of newborns with neural tube defects.
- ☑ 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 below 45cc.
- ☑ Consider use of phenoxybenzamine for relief of bladder sphincter spasm. Start with 1 mg P. O. q 24 hours as one dose and advance until residual urine is less than 5 cc, or side effects such as stuffy nose or drowsiness occur. Note that the normal urinary bladder capacity of a neonate is 30 cc.
- ☑ Organize discharge plan for parents and the local health care provider. Begin early in the hospitalization and involve the unit 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. Vinegar is an excellent agent for protecting the skin from alkali burns.
- ☑ Record daily head size, plot on appropriate head size chart and obtain follow-up cranial ultrasounds as needed. In premature infants, always obtain cranial ultrasounds since 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/Congenital 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. DO NOT leave this up to unit assistants or others unfamiliar with our clinic. To do so often results in 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 or 206.987.2204.

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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