Update and News From Mayo Clinic in Minnesota

Update on Spina Bifida Management at Mayo Clinic

Mayo Clinic offers a full range of services for rare and common congenital spinal cord defects. Two areas of that program are prenatal open spina bifida surgery and repair in tethered cord syndrome.

Prenatal Open Spina Bifida Surgery
Mayo Clinic’s practice in prenatal surgery for repair of myelomeningocele (Figures 1 and 2), the most common form of spina bifida, is now up and running. In May 2012, Mayo Clinic in Rochester, Minnesota, performed the first surgery of this type in the Upper Midwest. “It was a complete success,” says Nicholas M. Wetjen, MD, the neurosurgeon on the team. Ten weeks after the prenatal repair, the infant was born at 36 weeks’ gestation without complication or motor impairment and is continuing to thrive.

Forty medical specialists contributed to the procedure, which was led by surgeons Norman P. Davies, MD, in obstetrics and gynecology, Abdalla E. Zarroug, MD, in pediatric surgery, and Dr Wetjen in neurosurgery. “This is the kind of surgery that Mayo is set up for, with a team of collaborating surgeons and numerous medical disciplines involved,” says Dr Wetjen. “The patient was identified, and within three weeks we were performing the operation.”

Having encountered prenatal surgery in an animal model as an undergraduate, Dr Wetjen says the surgical experience had such an impact on him that it was a major factor in his decision to become a pediatric neurosurgeon. Prenatal repair of myelomeningocele in humans was pioneered in the 1990s, and in 2003, a randomized, prospective clinical trial for the procedure was initiated. The results, reported in The New England Journal of Medicine in 2011, showed that when adjusted for lesion level, prenatal surgery improved outcomes, despite the fact that infants in the prenatal group had more severe lesions than those in the postnatal surgery group and that nearly 13% of them were delivered prematurely (eg, before 30 weeks’ gestation). The significant findings included reduced need for shunting at 12 months of age, improved scores on mental and motor function at 30 months of age, and decreased incidence of hindbrain herniation associated with Chiari II malformation.

The surgery is not without risks to maternal and infant health, which is one reason why it is performed at so few medical centers. Not all infants improve, and risks must be weighed against benefits. Patient selection criteria are critical. Obesity, for example, was an exclusion criterion in the clinical trial.

Having waited for the results of the clinical trial, the Mayo team went to the University of California, San Francisco, one of the three participating study centers, for specialized training. “We have organized and adapted the surgery in a way we feel is most efficient for Mayo,” Dr Wetjen explains. Some of the critical aspects of the surgery are anesthesia for mother and infant and continuous irrigation and replacement of the amniotic fluid. “The neurosurgical repair is very much like what we do in a postnatal open spina bifida repair,” he says. “Depending on the age of the fetus, the tissue planes may be easier to define, just more friable because the patient is so fragile.”

Repairing Tethered Cord Syndrome
Occult spinal dysraphism, or tethered cord syndrome (TCS), can take several forms, but
generally the terms refer to a malformation of the spinal cord in which the cord is tethered to what is usually a developmental abnormality at the base of the spine. The cord malformation may not be able to be repaired; however, by untethering it, further neurologic deficits can be prevented. TCS may be suspected when there is abnormal hair or discoloration of the skin at the tethering site. A fat pad or dimple above the gluteal cleft, especially one with fluid discharge, is highly suspicious for an underlying abnormality. Early diagnosis is critical, and definitive diagnosis requires an MRI scan. Dr Wetjen points out that pediatricians may be reluctant to refer an infant for MRI because it requires anesthesia, but he adds that Mayo’s pediatric anesthesiologists are very experienced in conducting MRI in infants and children.

If neurologic symptoms are not present at the time of diagnosis, patients are monitored over time in Mayo’s Cerebral Palsy/Spina Bifida Clinic. The clinic includes physicians and other specialists from neurology, neurosurgery, urology, and physical medicine and rehabilitation.

When symptoms are present—and, in some cases, as a preventative measure—surgery is required to untether the cord. If the dural spinal tract is leaking fluid, emergency surgery is performed to prevent bacteria from entering the spinal canal. Dr Wetjen states that apart from emergency situations, surgery is best conducted when the child is between six and 12 months of age, and this is one reason why early diagnosis is so important.

In addition to seeing referred patients, Dr Wetjen occasionally reviews the medical history, MRI scans, and videos sent by parents who are considering adopting children with confirmed spinal abnormalities. “It is a type of complimentary electronic consult,” he explains. A father of adopted children, Dr Wetjen feels a special bond with adoptive parents and understands the many unknowns they face.

Mayo Clinic in Minnesota has a multidisciplinary team to manage the ongoing neurologic, bowel, and bladder issues and chronic pain that may be associated with spina bifida into adulthood. With the expansion into prenatal surgery for myelomeningocele, Mayo is well prepared to manage spinal cord abnormalities across the life span.

Figure 2. Prenatal open myelomeningocele repair. A, Ultrasonography of the abdomen and uterus is used to plan the incision that exposes the myelomeningocele defect of the fetus. B, The uterus is opened, the defect is exposed, and, with continuous infusion of warm irrigation to replace the amniotic fluid, the neural placode is exposed, dissected, and closed under microscopic magnification. C, The dura is closed in a watertight manner or, in some cases, is grafted because native dura is inadequate for closure. D, The dysplastic skin edges are trimmed, the lumbar myofascia elevated, and the skin closed over the neural placode and dural closure before closing the hysterotomy and abdominal incision.