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COVER

“The Knitting Lesson” (ca 1860) by Jean Francois Millet (1814–1875). Renowned for his peasant paintings, Millet in this painting illustrates the cycles of life and the passing on of skills from one generation to another. One of the major tasks of pediatricians is to teach parents and children skills to promote health. May we do it as gently and lovingly as this mother teaches her daughter knitting. (From the Museum of Fine Arts, Boston, Massachusetts.)

ANSWER KEY

Myelomeningocele


Myelomeningocele (MM) occurs in 1 in 1000 newborns in the United States and is the most common physically disabling birth defect in humans. Caused by failure of fusion of the caudal portion of the neural tube (the embryonic structure that gives rise to the central nervous system), MMs most commonly occur in either the lumbar or the sacral portions of the spine. Because of the neurologic deficit that occurs distal to the site of MM, affected children often require intervention by many members of a diverse group of medical and surgical specialists, including neurosurgeons, orthopedic surgeons, urologists, rehabilitation and developmental medicine specialists, and the general pediatrician. It is the pediatrician who plays a key role in the early management of these children. Recognition of problems early in life, with appropriate evaluation of the newborn and counseling of the parents and family, must be performed in a timely and sensitive manner.

Management of children affected with MM must be geared toward the anomalies that are known to occur commonly in association with this disorder. During the immediate neonatal period, after the condition has been diagnosed by visual inspection, the infant should be kept in a prone position with the sac covered with sterile gauze soaked with warm saline, and manipulation should be kept to a minimum. Generally, all newborns who have a MM undergo a neurological procedure to close the back within 48 h of birth; occasionally, surgery may be delayed if the infant is medically unstable.

After surgery, the child is kept in a prone position with the surgical site covered with a sterile drape to minimize the risk of infection, and the head circumference is monitored daily. Hydrocephalus, due to Arnold-Chiara malformation, occurs in 70% to 90% of children who have MM. If the head circumference is noted to be enlarging rapidly, or if symptoms or signs of increased intracranial pressure are noted, ultrasonography or computed tomographic scan of the head should be performed. If hydrocephalus is confirmed, a ventriculoperitoneal shunt is placed.

In addition to these neurosurgical problems, infants and children affected with MM are at risk for urologic and orthopedic complications. Urologic dysfunction, resulting from neurogenic bladder, is a serious, potentially life-threatening, and common associated feature. Children who have neurogenic bladders can be divided into two categories: those who experience urinary retention and overflow and those with total urinary incontinence. In patients with the former problem, increased sphincteric activity, resulting in persistent retention of urine after voiding, ultimately leads to bladder thickening, upper urinary tract dilatation, vesicoureteral reflux of urine, and recurrent urinary tract infections. In patients found via postvoid catheterization to have high urinary residual volumes, clean intermittent catheterization, coupled with frequent imaging (including sonography of the kidneys and voiding cystourethrogram) and urodynamic studies, is essential.

During the newborn period, the bladder of the infant with MM should be catheterized after voiding to assess the presence of residual urine. Infants who demonstrate total incontinence and in whom constant dribbling of urine is found require careful and regular monitoring of postvoid residual volumes to assess indications for clean intermittent catheterization.

Owing to the high frequency of orthopedic dysfunction associated with MM, it is essential that an orthopedic surgeon, working in conjunction with a physiatrist and an orthotist, follow affected infants and children. Because the fetus with MM may be unable to move the lower extremities as a result of the neurologic lesion, newborns often are
as brief a period as possible (6 mo to 1 y) that covers the period of greatest likelihood of recurrence.

We give every family a detailed description of the risks and benefits described previously so they can make an informed decision. Each parent is taught how to manage the occurrence of another seizure. In some instances, we will give the family a prescription for rectal diazepam and teach them how it should be given if the child has a prolonged seizure. This provides the family with a sense of control over future seizures.

When we educate families about the natural history of febrile seizures and their consequences and the alternative of daily anticonvulsant medication and its consequences, most of our parents elect not to have their child started on anticonvulsant medication following a first febrile seizure. Most also are relieved not to be forced to medicate their child even if the child has a second and sometimes a third seizure. Many parents whose children have been started on medication after a first febrile seizure are delighted to be “allowed” to discontinue it. We see each of these decisions as being in accord with the principles of rational decision making.

**SUGGESTED READING**
A list of references and additional readings is provided at the end of the accompanying article on "Decision Making and the Child with Afebrile Seizures."
PIR QUIZ

10. Statements supporting the overwhelming importance of careful history-taking and clinical observation in the diagnosis, classification, and management of afebrile seizures include each of the following, except:
   A. History and clinical observations may support a diagnosis of epilepsy even if the electroencephalogram results are normal.
   B. Abnormal electrical events should not be treated as seizures in the absence of clinical manifestations.
   C. Partial seizures are more likely than generalized seizures to be treatable by surgery.
   D. Partial and generalized seizures often respond best to different anticonvulsants.
   E. In contrast to partial seizures, generalized seizures are usually difficult to control.

11. Regarding the long-term consequences of afebrile seizures, it is
   A. often difficult to determine whether anticonvulsants are needed.
   B. generally not known whether anticonvulsants are needed.
   C. generally not known whether anticonvulsants are needed.
   D. generally not known whether anticonvulsants are needed.
   E. generally not known whether anticonvulsants are needed.

12. A 6-y-old boy experiences the onset of recurrent partial complex seizures. Your initial choice of anticonvulsant is carbamazepine. In the interest of maximizing benefits and minimizing expense and side effects, you do each of the following, except:
   A. Monitor the boy closely for headache, drowsiness, and dizziness during the first few weeks of therapy.
   B. Discontinue the medication when the white blood cell count is found to be 3500/mm³ (with 40% neutrophils) during a visit for cough and fever.
   C. Obtain a drug level when seizures continue despite a reasonable dose.
   D. Settle on a dose that controls the seizures without toxicity, no matter what tests of serum drug level reveal.

13. A 10-y-old girl has been treated for 1 y with sodium valproate for idiopathic partial complex seizures. She has attained excellent control without signs of toxicity. She has gained 7 lb during the past 12 mo. Her mother asks you about adequacy of the current dosage and when it will be safe to take her daughter off medication. You reply with each of the following statements, except:
   A. The seizure threshold generally increases with age, independent of medication use.
   B. If the girl remains free of seizures for 2 y, she has an excellent chance of staying free of seizures even if off valproate.
   C. A normal or greatly improved electroencephalogram will increase the likelihood that she will remain free of seizures once off medication.
   D. Given her weight gain, the dosage of medication should be increased to prevent breakthrough seizures.
   E. Idiopathic seizures are much less likely to recur without therapy than are seizures of known cause.