The online version of this article, along with updated information and services, is
located on the World Wide Web at:
http://pedsinreview.aappublications.org/content/26/2/50

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™

Neurosurgical Management of Myelomeningocele (Spina Bifida)

Mark S. Dias, MD, FAAP*

Objectives   After completing this article, readers should be able to:

1. Understand the outcomes regarding survival, cognitive abilities, ambulation capacities, and urologic functions for the child who has a myelomeningocele.
2. Appreciate the importance of folate in the pathogenesis of myelomeningoceles and the rationale for periconceptional folate supplementation in the prevention of neural tube defects.
3. Describe the initial neurosurgical care and rationale for neurosurgical treatment for the child who has a myelomeningocele.
4. Discuss the common presenting features of a shunt malfunction in the child who has a myelomeningocele.
5. Understand the significance and describe the management of the child who has signs or symptoms of shunt malfunction and no change in ventricular size on imaging studies.
6. Discuss the importance of considering shunt malfunction before initiating other neurosurgical treatments for the deteriorating child who has a myelomeningocele.
7. Describe the common signs and symptoms of spinal cord tethering, Chiari malformation, and syringomyelia, and the surgical options for each.

Introduction

Myelomeningocele (spina bifida) is the most common congenital neurologic malformation compatible with life. Therefore, every busy pediatric practice includes at least a few children who have spina bifida, and it is important for the primary care clinician to have a practical working knowledge about the unique and important problems that face this group of children. The incidence of myelomeningocele in the United States is between 0.41 and 1.43 per 1,000 live births (1) and is lower among African-Americans than Caucasians. Native Africans have a very low incidence of 0.1 per 1,000 live births. (2) The incidence of myelomeningocele in the United States and worldwide has declined significantly over the past 3 decades. (2) This decline has been attributed, in part, to improved maternal nutrition and periconceptional vitamin supplementation and to wider availability of prenatal diagnostic tests with a corresponding increase in the number of elective terminations. However, these factors account for only a portion of the decline, and other reasons are not yet known.

Myelomeningocele represents a localized failure of the embryonic neural tube (the forerunner of the brain and spinal cord) to close properly, likely due to a disruption of the interactions among many complex environmental, teratogenic, and genetic factors. A hereditary component is suggested by the higher incidence in families already having one or more affected children. The odds of the parents of a child who has myelomeningocele having a second affected child increases to 3% to 4%, and the odds for a third affected child among those already having two children who have the disorder is 10%.

One identified environmental factor in the embryology of myelomeningocele is the vitamin folic acid (folate) and its metabolites. Periconceptional administration of supplemental folate in both noncontrolled and randomized, placebo-controlled studies has reduced the recurrence rate among women who have had a previously affected pregnancy

*Associate Professor of Neurosurgery, Pennsylvania State University College of Medicine and Milton S. Hershey Medical Center, Hershey, Pa.

(at a dose of 4 mg or 4,000 mcg/d) as well as among women who never have had a child who has a myelomeningocele (at a dose of 0.4 mg or 400 mcg/d). However, studies of serum and red blood cell folate levels among mothers of infants who have myelomeningocele have produced inconsistent results. More recent attention has focused on the suggestion that neural tube defects are caused not by a deficiency of folate but by enzymatic abnormalities involving metabolic pathways that require folate and its metabolites tetrahydrofolate and 5-methyltetrahydrofolate, (3) abnormalities that could be overcome with folate supplementation.

Children who have spina bifida comprise one of the largest groups of patients for whom pediatric neurosurgeons care. Their management often is extraordinarily complex and may require the integrated efforts of caregivers from many disciplines, including neurosurgeons, orthopedists, urologists, physiatrists, physical and occu-

Periconceptual
administration of supplemental folic acid has reduced the recurrence rate among women who have had a previously affected child as well as among women who have never had such a child.

pational therapists, orthotists, gastroenterologists, endocrinologists, speech and swallowing experts, pulmonologists, and intensivists. Use of a spina bifida team, in which a core of dedicated clinicians and staff provide coordinated, multidisciplinary care, results in the best opportunity to minister comprehensively to the child who has spina bifida, with medical care both higher in quality and ultimately less expensive. (4)

Treatment of children who have myelomeningoceles has undergone a striking evolution over the past several decades. From the early 1900s, improvements in surgical techniques have allowed early closure of the back wound with no infection resulting. The introduction of effective cerebrospinal fluid (CSF) shunting systems in the 1950s led to dramatic improvements in the treatment of hydrocephalus. The introduction of clean intermittent catheterization (CIC) for the treatment of associated neurogenic bladder dysfunction dramatically reduced the

mortality from chronic renal failure. More recently, the following have led to improved long-term function and quality of life for many affected children: 1) treatment of the symptomatic Chiari malformation by posterior fossa and cervical decompression, 2) treatment of progressive spinal cord dysfunction by spinal cord untethering and shunting of syringomyelia, and 3) aggressive orthopedic management of spinal and lower extremity deformities.

Critical Decisions

Perhaps the greatest change has occurred in attitude. In the past, many physicians considered treatment of children who had myelomeningocele to be cruel, believing it more merciful to allow them to die. In 1971, in an effort to decide better which patients should be treated, the "Lorber criteria" were developed to allow withholding of treatment from certain infants who had myelomeningoceles. (5) The Lorber criteria included: 1) the presence of severe hydrocephalus at birth, 2) total paraplegia, 3) spinal kyphosis at birth, and 4) any additional birth defect. However, none of these criteria, either in isolation or in combination, is an entirely accurate predictor of outcome. (6) Moreover, the outcome of children treated without regard to selection criteria (nonselective treatment) (7) compares favorably with those from Lorber's and others' series (8)(9)(10) in which selection was employed. A comparison of the unselected series by McLone (7) with the selected series of Lorber and Salfield (10) shows that the mortality rate for the unselected group was comparable to that of the "best" infants from the selected group. Overall mortality for the unselected group was 15% and that for the selected group (including those infants who were believed to be poor risks and, therefore, were allowed to die) was nearly 70%. Intelligence quotients (IQs) for the "best" infants from the selected group were only slightly higher than those in the unselected group (15% more children having normal IQs) and were more than offset by the tremendous increase in mortality. Examined from a different perspective, 60% of the children from the selected group who were allowed to die would have been "competitive" if they had been allowed to survive. (7) Finally, only 13 of 300 parents admitted to regret over their decision to treat their child; remarkably, 9 of the 13 regretted their initial decision not to treat their child. (6)

This is not to say that every child who has a myelome-

ningocele should be treated. Children who have multiple severe or potentially fatal associated malformations and those who have severe chromosomal abnormalities may not be chosen for treatment. However, the decision to withhold treatment from any child should be individualized and made only after a frank and realistic discussion with the parents about the chances for meaningful survival. The children for whom medical care should be withheld generally comprise fewer than 1% of infants born with myelomeningoceles.

The parents of children who have myelomeningoceles are overwhelmed initially with the complexity of the disorder that afflicts their infant. Confused and often frightened, they usually follow the suggestions of the treating physician. Accordingly, it is incumbent on physicians to present an accurate picture of the nature of the myelomeningocele; the likely sequelae of the malformation; and the outlook for intellectual, sensorimotor, urinary, sexual, and societal function. Although each clinician has an inherent bias based on personal, ethical, or religious principles, we must refrain from transmitting this bias to the parents because it is they rather than we who will shoulder the burdens and share the joys of caring for their child.

Prognosis

A few facts are important to transmit accurately to the parents. First, nearly all children who have myelomeningocele survive initially with proper treatment, (11)(12) although a few children succumb each year, most commonly to the Chiari malformation in infancy and to unrecognized shunt obstruction in later life. Hindbrain dysfunction is the most frequent cause of death in infancy because of associated stridor, apnea, and aspiration pneumonitis. (13) The long-term mortality can be as high as 35% to 50% by adulthood. (14)(15)(16) Approximately 75% of children who have myelomeningoceles have IQs higher than 80, and this figure is not affected significantly by the presence or absence of hydrocephalus. (13)(17)(18)(19)(20) However, among those whose intelligence is normal, 60% are learning disabled, (18)(21)(22) with the most common feature being a nonverbal learning disability. Verbal scores typically are better than performance scores, and particular deficits are seen in mathematics, sequencing of information, visual perceptual skills, and problem solving.

Approximately 60% to 70% of surviving children can ambulate well enough to function in the community.

(21)(23)(24) Ambulation obviously is highly dependent on and directly correlated with sensorimotor level. (23) Excluding children who have severe developmental delays and hypotonia (accounting for about 13% of patients), 89% of preadolescent children, including 100% of those who have low lumbar and sacral level lesions and 63% of those who have higher level lesions, are community ambulators when they receive aggressive multidisciplinary orthopedic and neurosurgical management. (21) After adolescence, however, community ambulation decreases to about 50% because it becomes more energy-efficient to use a wheelchair. (13) Orthopedic procedures are common; (24)(25) in one study of 206 unselected children who were followed for 6 months to 18 years, 64% underwent at least one orthopedic procedure and an average of 5.4 procedures were performed per patient. (24)

Urologic social continence (free of urinary incontinence in social situations with or without CIC) can be achieved in about 85% of school-age children. (11)(26)(27)(28) Most have chronic constipation, and bowel regimens are important to avoid obstipation. Although

Despite their handicaps, many children who have myelomeningoceles grow up to be contributing members of society.

almost all people who have myelomeningocele have abnormal genital sensation, about 66% of boys report at least some feeling, and 70% to 75% report the ability to achieve erections, although many are reflexive. An objective assessment of 15 selected male patients by Sandler and colleagues showed that 60% had objective evidence of at least nocturnal erections. (29) Ejaculation and reproduction are possible, although oligospermia (of uncertain cause) is reported.

Despite their handicaps, many children who have myelomeningoceles grow up to be contributing members of society. Some 82% of adult patients are independent in activities of daily living, and 30% attend or finish college. However, only 32% are gainfully employed. (30) An increasing problem for many is navigating the transition from childhood to adult care clinicians and maintaining access to appropriate health care in adulthood.

Therapy

From a neurosurgical perspective, essentially all children must have their myelomeningocele repaired at birth, and

Figure 1. Large myelomeningocele in a newborn. Arrows point to the exposed placode (open spinal cord). The surrounding tissue is dysplastic epithelium

80% to 90% ultimately require a shunt for control of hydrocephalus. (11)(13)(21) Further, 30% to 40% of those receiving shunts require at least one shunt revision during the subsequent year, 60% within 5 years, and 85% within 10 years. (13)(31) About 33% of children require spinal cord untethering, and 15% to 35% undergo occipitocervical decompression of the Chiari malformation for neurologic deterioration at some point in their lives.

At the time of the child's birth, the myelomeningocele placode (Fig. 1) should be protected from external mechanical trauma and desiccation and kept as clean as possible because the malformation may contain functional neural tissue and frequently leaks CSF. The malformation is covered best with gauze dressings soaked in sterile saline solution. Iodine-containing compounds such as povidone-iodine should be avoided because they may injure the exposed tissues further. Plastic wrap may be placed atop the gauze to keep the dressings moist. Sensorimotor function can be assessed by observing the spontaneous movements of the legs and feet and the response to pinprick stimulation. In addition, muscle imbalance produces characteristic postures of the lower limbs and gives a good indication of sensorimotor level. Imbalances between iliopsoas and quadriceps in the infant who has an upper lumbar level lesion results in a flexed hip deformity, imbalances between quadriceps and hamstrings in the infant who has a mid-lumbar level lesion produces a characteristic knee extension deformity, and imbalances between foot dorsi- and plantar-

flexors in the infant who has a low lumbar level lesion produces a dorsiflexed ankle (rocker bottom) deformity.

The malformation should be closed surgically within the first 1 to 3 days after birth to avoid meningitis or other complications. Hydrocephalus is apparent in about two thirds of newborns and develops over days to weeks after the closure in most of the remainder. Symptoms and signs of hydrocephalus include macrocrania, a bulging fontanelle, split sutures, sunsetting eyes, poor feeding, vomiting, irritability or lethargy, apnea or bradycardia, and leakage of CSF from the closed repair site. Hydrocephalus also can promote or exacerbate symptoms from the Chiari malformation. Enlarged ventricles are identified readily on cranial ultrasonography, computed tomography (CT), or magnetic resonance imaging (MRI); serial imaging studies often show increasing ventricle size. Hydrocephalus is treated with a ventricular shunt. Those who have severe hydrocephalus at birth may be treated either with a shunt inserted contemporaneous with the myelomeningocele closure (with no demonstrated increased risk of shunt infection or other complications) or with temporary CSF diversion via external ventricular drainage to a closed collection system at the bedside for several days, followed later by a permanent shunt. For the remainder of patients, treatment generally is delayed until the need for a shunt becomes apparent (usually within days to weeks after the myelomeningocele repair). A ventriculoperitoneal shunt is employed most commonly; ventriculoatrial shunts now are used rarely because of the increased risk of significant complications such as sepsis, endocarditis, and shunt nephropathy.

Although 80% to 90% of children who have myelomeningocele ultimately require a shunt, a shunt commits both the patient and physician to a lifelong struggle to maintain shunt patency. Even a functioning shunt wreaks havoc on the patient and family because signs of shunt malfunction (eg, headache, nausea, vomiting, listlessness) are confused easily with a viral syndrome and can prompt another visit to the physician or hospital. Finally, most children who have myelomeningocele and shunted hydrocephalus are considered shunt-dependent for life. (32) Therefore, shunts should be provided only to those

children whose need has been demonstrated. Asymptomatic infants who have stable mild or moderate hydrocephalus and a cortical mantle of at least 3.5 cm may be observed safely for up to 5 months without shunting with no demonstrated deleterious effect.

The long-term follow-up of individuals who have myelomeningocele is accomplished best through a consistent partnership between the primary care practitioner and a multidisciplinary team of dedicated pediatric medical and surgical specialists who evaluate the child regularly. The evaluation of shunt function (and malfunction) might be the most critical part of this partnership. The evaluation of potential shunt malfunction is the most worrisome and time-consuming activity for both individuals who have myelomeningocele and their caregivers. The signs and symptoms of shunt malfunction in these individuals are legion and often can confuse even the most experienced clinician. The oft-quoted signs and symptoms of intracranial hypertension, including headache, nausea and vomiting, poor feeding, listlessness or lethargy, sun-setting eyes, or extraocular abduction palsies, although indicative of a shunt malfunction, may be absent. In addition, a shunt malfunction in this population may manifest as: 1) cognitive changes such as a decline in school performance or worsening behavior; 2) the onset of, or change in the frequency of, seizures without another demonstrable cause; 3) a change in motor performance such as a decrease in muscle strength, loss of previously acquired motor skills, or increase in spasticity in either upper or lower extremities; 4) a change in ambulation without another definable cause; 5) a change in urinary or bowel function (more frequent "accidents" or urinary tract infections or an increase in catheterization frequency); 6) a change in lower cranial nerve function suggestive of hindbrain dysfunction; 7) pain in the back, particularly at the myelomeningocele repair site; or 8) worsening scoliosis or lower extremity orthopedic deformities. Once a shunt is placed, any subsequent deterioration in neurologic, orthopedic, or urologic function should prompt an investigation of the shunt to exclude a malfunction before any other treatment is rendered. Finally, because papilledema may be the only sign of shunt malfunction in a few individuals, every physician encounter should include a funduscopic examination.

The evaluation of a child suspected of having a shunt malfunction usually includes CT to evaluate ventricular size. Most shunt malfunctions are accompanied by an increase in the ventricle size compared with baseline scans. Unfortunately, baseline scans are not always available, and the presence of "normal" ventricles on CT does not exclude a shunt malfunction. Moreover, up to 15% to 20% of patients who have shunt malfunction have little or no change in ventricle size, necessitating the diagnosis of shunt malfunction solely on clinical grounds. Parents' assessments of their children's clinical condition can be extremely important in these circumstances and never should be ignored. Any patient who has significant symptoms or signs suggesting shunt malfunction should be referred to a neurosurgeon. The need to consider the shunt first when assessing the child who has a myelomeningocele cannot be overemphasized; nowhere else in pediatric neurosurgery is clinical judgment so important and misjudgment so treacherous.

The Chiari II Malformation

Some 98% of children who have myelomeningoceles have an associated Chiari II malformation (Fig. 2) con-

Spinal cord tethering is a well-recognized cause of neurologic deterioration in children who have a myelomeningocele.

sisting of caudal displacement of the cerebellar tonsils and vermis, caudal medulla, and variably the fourth ventricle into the cervical spinal canal. The Chiari II malformation also incorporates a number of other brain malformations (eg, callosal dysgenesis, abnormalities of neuronal migration and brain sulcation) that likely are responsible for the learning disabilities faced by affected children. Chiari II malformation, therefore, is considered a "pancerebral malformation." Although anatomically present in nearly every child who has a myelomeningocele, it results in significant problems severe enough to require surgical treatment in only 15% to 35% of patients. Symptoms may develop at any age but are most common during infancy. Disorders of swallowing, representing disordered lower cranial nerve or brainstem function, are the most frequent clinical manifestations and may include choking on foods (particularly liquids), nasal regurgitation during drinking, and frequent vomiting or significant gastroesophageal reflux. (33) Among the other signs of lower brainstem dysfunction are repeated
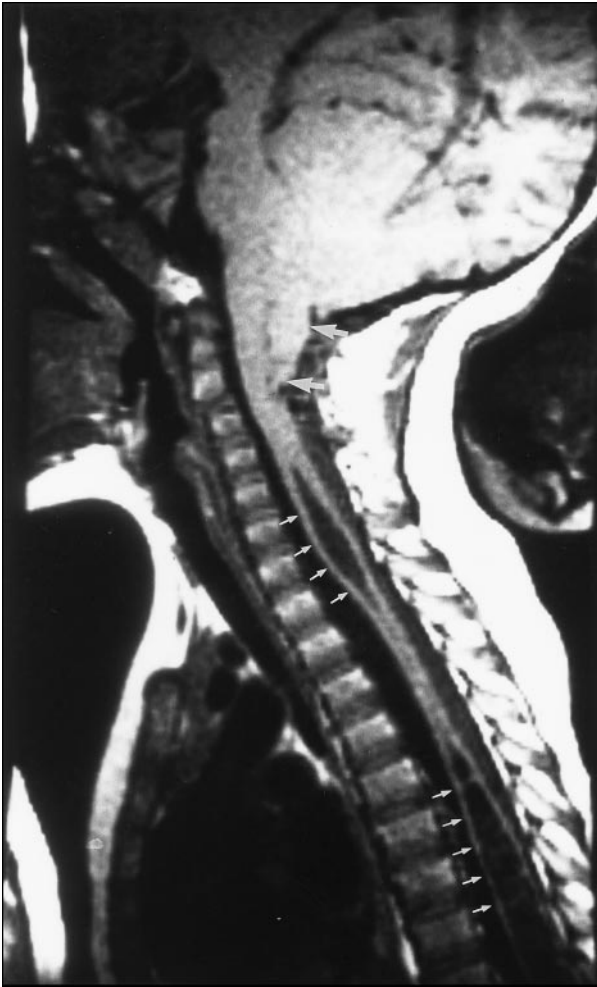
Figure 2. Sagittal T1-weighted MRI of the posterior fossa and cervical and thoracic spinal cord of a child who has a myelomeningocele. A Chiari II malformation is evident, with the inferior cerebellar vermis (large white arrows) and medulla projecting well below the level of the foramen magnum. There also is a syringomyelia (small white arrows) that spans C6 to T2 superiorly and from T5 inferiorly, extending to the bottom of the figure.

aspiration pneumonia, dysarthria, apnea or cyanotic spells, inspiratory stridor, and a hoarse or high-pitched cry. Newborns who have symptomatic Chiari II malformations may have soft, feeble cries and, therefore, may be described as "good" babies by nursery personnel. (34) Other signs and symptoms of the Chiari II malformation, particularly in older children and adults, may include weakness or spasticity of the upper extremities, pain (headache or neck pain), cerebellar dysfunction, oculomotor changes, and scoliosis.

The Chiari II malformation is evaluated anatomically with MRI. Associated hydrocephalus, syringomyelia, and spinal cord tethering may be evaluated simultaneously. The decision to treat the Chiari II malformation surgically depends on the magnitude, duration, and type of symptoms. For example, the parents of most children who have myelomeningocele describe some sort of "food intolerance" in their children, who have a more sensitive gag reflex, intolerance for certain food textures, and frequent vomiting when eating certain foods. Many of these symptoms improve or resolve with time and do not require treatment. Similarly, infants who have mild, nonprogressive symptoms may be followed expectantly; many improve spontaneously. On the other hand, children who have vocal cord weakness or paralysis, significant stridor, apnea, aspiration, or sensorimotor deterioration are considered potential surgical candidates. The surgical approach to these patients entails initially addressing any associated hydrocephalus, either by providing shunts for infants who have hydrocephalus and do not yet have a shunt or by excluding a shunt malfunction in those who have one. In many cases, effective treatment of the hydrocephalus can eliminate symptoms. For the remainder or patients, direct surgical repair of the Chiari II malformation is undertaken.

The surgical approach involves decompression of the medulla and upper cervical spinal cord followed by insertion of a dural patch graft (duraplasty) to expand the cervical dural sac. Other procedures, such as opening or stenting the fourth ventricle (in the absence of fourth ventricular enlargement) or plugging the obex, have not proven to be of significant additional benefit and largely have been discarded. Cervical laminectomy is carried out to below the most inferior level of the cerebellar tonsils, and a generous patch graft (of periosteum, cadaver dura, or bovine pericardium) is sewn to the dural edges to create a more capacious dural sac. Surgical outcome depends on the patient's age and presentation. Patients whose symptoms are obvious at the time of birth generally fare poorly; postmortem studies often demonstrate a disorganized and chaotic brainstem architecture. (35) Many of these newborns require tracheostomies, Nissen fundoplications, and gastrostomy feeding tubes because of persistent swallowing difficulties, vocal cord palsies, repeated episodes of aspiration pneumonia, and gastroesophageal reflux despite surgical decompression. In contrast, older infants and children often improve following decompression, particularly if it is performed before severe and irreparable damage to the brainstem has occurred. (33) Overall, swallowing and other brainstem abnormalities improve in about 50% of patients;

upper extremity weakness, cerebellar dysfunction, and pain improve in more than 80%.

The Tethered Cord

Spinal cord tethering (Fig. 3) is another well-recognized cause of neurologic deterioration in children who have myelomeningoceles and may produce delayed and progressive neurologic deficits, urologic dysfunction, or orthopedic deformities. (36)(37)(38)(39)(40)(41)(42)(43) Although anatomically present and visible on imaging studies in virtually every child who has myelomeningocele, only about one third require surgical treatment. Untethering is reserved for those having progressive symptoms or signs of tethering. Growth and development of the vertebral column may stretch the tethered spinal cord progressively; the increased incidence of the tethered cord syndrome during periods of rapid growth (37)(44)(45)(46) supports this concept. In addition, intermittent and transient traction on the tethered spinal cord may increase with flexion movements of the pelvis or spine (47) and contribute to repetitive neurologic injury. Several studies have documented improvement or stabilization of neurologic function, (37)(41) (44)(48)(49)(50) urologic disorders, (38) and orthopedic deformities (39)(42)(43) after untethering.

It should be emphasized that tethered cord syndrome in the patient who has a myelomeningocele is diagnosed on clinical rather than on radiologic grounds. A vigilant eye; keen clinical judgment; and objective, accurate, and reproducible means of evaluating patients (eg, serial manual muscle testing, urodynamics, scoliosis radiographs) are necessary to evaluate and follow the child who has a tethered spinal cord. Signs and symptoms of spinal cord tethering may include: 1) pain, either in the back or legs; 2) motor deterioration, manifested as a decrease in muscle strength or an increase in tone (spasticity); 3) a change in sensory level; 4) a change in bowel or bladder function (clinical change or changes on urodynamic studies); 5) a deterioration in gait; and 6) progressive orthopedic deformities, either in the lower extremities (hip dislocation, pes cavus, equinovarus) or spine (scoliosis).

Once tethering is suspected clinically and hydrocephalus or shunt malfunction convincingly excluded as a cause of the deterioration, further evaluation with manual muscle testing, urodynamics, or scoliosis radiographs may be necessary to document changes in function objectively. Radiologic evaluation includes MRI to confirm tethering and define the anatomy radiographically and to identify any associated lesions (such as lipomas, syringomyelia, or other developmental malformations) that

Figure 3. Sagittal T1-weighted MRI of a child who has myelomeningocele. The spinal cord extends down to the level of the sacrum, where it is attached to the site of the previous closure. This represents radiographic evidence of spinal cord tethering. Also, a long syringomyelia extends to the level of L1. This type of "haustral" appearance is relatively common in syringomyelia, and despite the appearance, this pattern usually does not represent multiple isolated compartments.

could contribute to neurologic deterioration. Untethering is accomplished by reopening the previous wound, exposing the dura, and dissecting the scarred portion of

the spinal cord (the myelomeningocele placode) from the overlying dural closure. The caudal end of the placode is inspected for tethering lesions because a few patients have an unrecognized (and previously untreated) thickened filum terminale that also can tether the spinal cord. Other associated tethering lesions identified on the preoperative MRI are repaired simultaneously.

Syringomyelia

Syringomyelia represents a dilatation of the central canal of the spinal cord with CSF (Figs 2, 3). Syringomyelia is visible on MRI in up to 80% of patients who have myelomeningocele, (21)(51)(52)(53) but becomes symptomatic in only 2% to 5%. Common presenting features include upper extremity weakness or loss of function, back pain, scoliosis, and worsening spasticity or ascending motor loss in the lower extremities. Bowel and bladder dysfunction are unusual. The classic "dissociated sensory loss" (loss of pain or thermal sensation with preservation of proprioception) (34) is uncommon among these patients. Although syringomyelia and Chiari I malformation are recognized as causes of scoliosis in children who do not have myelomeningocele, the contribution of the Chiari II malformation or syringomyelia to scoliosis in children who have myelomeningocele is less clear because the genesis of scoliosis in this population is multifactorial and complex. Finally, extension of the syringomyelia into the brainstem (syringobulbia) can cause lower cranial nerve and brainstem dysfunction.

Because syringomyelia, like the Chiari II malformation and spinal cord tethering, is present in so many asymptomatic individuals who have myelomeningocele, its treatment is based largely on clinical symptoms and signs and requires considerable clinical judgment to determine which patients might benefit from treatment. Symptoms from syringomyelia may be caused by hydrocephalus or an unrecognized ventricular shunt malfunction (54)(55) and may resolve after shunt insertion or revision. In the absence of a shunt malfunction, the initial treatment of symptomatic syringomyelia in these patients is controversial. A Chiari decompression is recommended by some, based on the premise that restriction of dynamic CSF flow from the Chiari II malformation promotes or causes the syringomyelia. (56) However, this procedure may be less effective than direct shunt of the CSF within the syrinx to either the peritoneum or pleural cavity.

Conclusion

The single greatest long-term neurosurgical problem for children who have myelomeningoceles is the maintenance of proper shunt function. Most shunt malfunctions can be managed by prompt and timely recognition of signs and symptoms. Particular heed should be paid to the patient's and parent's concerns, especially if they suggest that the present symptoms or signs are similar to those that accompanied previous shunt malfunctions. If primary care and other physicians can be educated about the signs and symptoms of shunt malfunction in this population and understand that the ventricular size on CT may not change in the face of shunt malfunction, we will have advanced the care of these children considerably. Finally, delayed neurologic deterioration in affected children is not simply the natural history of this disorder; in most cases, it is due to a treatable cause. Diligent multidisciplinary follow-up and prompt surgical management of symptomatic Chiari II malformation, spinal cord tethering, and syringomyelia can stabilize or improve the status of these patients. With further advances, the outlook for these children is ever more hopeful.

References

1. Elwood JM, Little J, Elwood JH. Epidemiology and control of neural tube defects. *Monographs in Epidemiology and Biostatistics.* Volume 20. Oxford, England: University Press; 1992
2. Shurtleff DB, Lemire RJ. Epidemiology, etiologic factors, and prenatal diagnosis of open spinal dysraphism. *Neurosurg Clin North Am.* 1995;6:183–193
3. Scott JM, Wier DG, Molloy A, McPartlin J, Day L, Kirke P. Folic acid metabolism and mechanisms of neural tube defects. In: Bock GMJ, ed. *Neural Tube Defects.* CIBA Foundation Symposium Number 181. Chichester, England: John Wiley and Sons;1994: 180–191
4. Kaufman BA, Terbrock A, Winters N, Ito J, Klosterman A, Park TS. Disbanding a multidisciplinary clinic: effects on the health care of myelomeningocele patients. *Pediatr Neurosurg.* 1994;21:36–44
5. Lorber J. Results of treatment of myelomeningocele. An analysis of 524 unselected cases, with special reference to possible selection for treatment. *Dev Med Child Neurol.* 1971;13:279–303
6. McLone DG. Treatment of myelomeningocele: arguments against selection. In: Little JR, ed. *Clinical Neurosurgery.* Baltimore, Md: Williams and Wilkins; 1986:359–370
7. McLone DG. Results of treatment of children born with a myelomeningocele. *Clin Neurosurg.* 1983;30:407–412
8. Charney EB, Weller SC, Sutton LN, Bruce DA, Schut LB. Management of the newborn with myelomeningocele: time for a decision-making process. *Pediatrics.* 1985;75:58–64
9. Gross RH, Cox A, Tatyrek R, Pollay M, Barnes WA. Early management and decision making for the treatment of myelomeningocele. *Pediatrics.* 1983;72:450–457
10. Lorber J, Salfield S. Results of selective treatment of spina bifida cystica. *Arch Dis Child.* 1981;56:822–830

11. McLone DG. Continuing concepts in the management of spina bifida. *Pediatr Neurosurg*. 1992;18:254–256

12. Reigel DH, McLone DG. Myelomeningocele: operative treatment and results. In: Marlin AE, ed. *Concepts in Pediatric Neurosurgery*. Basel, Switzerland: S Karger; 1988:41–50

13. McLone DG, Dias L, Kaplan WE, Sommers MW. Concepts in the management of spina bifida. In: Humphreys RP, ed. *Concepts in Pediatric Neurosurgery*. Basel, Switzerland: S Karger; 1985:97–106

14. Hunt GM, Poulton A. Open spina bifida: a complete cohort reviewed 25 years after closure. *Dev Med Child Neurol*. 1995;37: 19–29

15. Hunt GM. Open spina bifida: outcome for a complete cohort treated unselectively and followed into adulthood. *Dev Med Child Neurol*. 1990;32:108–118

16. Bowman RM, McLone DG, Grant JA, Tomita T. Spina bifida outcome: a 25-year prospective. *Pediatr Neurosurg*. 2001;34: 114–120

17. McLone DG, Czyzewski D, Raimondi AJ, et al. Central nervous system infections as a limiting factor in the intelligence of children born with myelomeningocele. *Pediatrics*. 1982;70: 338–342

18. Shaffer J, Wolfe L, Freidrich W, et al. Developmental expectations: intelligence and fine motor skills. In: Shurtleff DB, ed. *Myelodysplasias and Extrophies: Significance, Prevention, and Treatment*. New York, NY: Grune and Stratton; 1986:359–372

19. Soare PL, Raimondi AJ. Intellectual and perceptual motor characteristics of treated myelomeningocele children. *Am J Dis Child*. 1977;131:199–204

20. Hunt GM, Holmes AE. Some factors relating to intelligence in treated children with spina bifida cystica. *Dev Med Child Neurol*. 1974;35(suppl):65–70

21. McLone DG. Myelomeningocele. In: Youmans JR, ed. *Neurological Surgery*. 4th ed. Philadelphia, Pa: WB Saunders; 1996: 843–860

22. Fletcher JM, Francis DJ, Thompson NM, et al. Verbal and non-verbal skill discrepancies in hydrocephalic children. *J Clin Exp Neuropsychol*. 1992;14:593–609

23. Findley TW, Agre JC, Habeck RV, Schmalz R, Birkebak RR, McNally MC. Ambulation in the adolescent with myelomeningocele: I. Early childhood predictors. *Arch Phys Med Rehab*. 1987;68: 518–522

24. Swank M, Dias L. Myelomeningocele: a review of the orthopaedic aspects of 206 patients treated from birth with no selection criteria. *Dev Med Child Neurol*. 1992;34:1047–1052

25. Brinker MR, Rosenfeld SR, Feiwell E, Granger SP, Mitchell DC, Rice JC. Myelomeningocele at the sacral level. *J Bone Joint Surg*. 1994;76-A:1293–1300

26. Spindel MR, Bauer SB, Dyro FM, et al. The changing neurologic lesion in myelodysplasia. *JAMA*. 1987;258:1630–1633

27. Pike JG, Berardinucci G, Hamburger B, Kiruluta G. The surgical management of urinary incontinence in myelodysplastic children. *J Pediatr Surg*. 1991;26:466–470

28. Rudy DC, Woodside JR. The incontinent myelodysplastic patient. *Urol Clin North Am*. 1991;18:295–308

29. Sandler AD, Worley G, Leroy EC, Stanley SD, Kalman S. Sexual function and erection capability among young men with spina bifida. *Dev Med Child Neurol*. 1996;38:823–829

30. McLone DG, Naidich TP. Myelomeningocele: outcome and late complications. In: McLaurin RL, Schut L, Venes JL, Epstein F, eds. *Pediatric Neurosurgery*. Philadelphia, Pa: Saunders; 1989: 53–70

31. Steinbok P, Irvine B, Cochrane DD, Irwin BJ. Long-term outcome and complications of children born with meningomyelocele. *Child Nerv Syst*. 1992;8:92–96

32. Rekate HL. To shunt or not to shunt: hydrocephalus and dysraphism. *Clin Neurosurg*. 1984;32:593–607

33. Pollack IF, Pang D, Kocoshis S, Putnam P. Neurogenic dysphagia resulting from Chiari malformations. *Neurosurg*. 1992;30: 709–719

34. Oakes WJ. Chiari malformations, hydromyelia, syringomyelia. In: Wilkins RH, Rengachary SS, eds. *Neurosurgery*. New York, NY: McGraw-Hill; 1985:2102–2124

35. Gilbert JN, Rorke LB, James HE, Jones KL, Chernoff GF. The pathological basis for the failure of surgery to relieve the symptomatic Arnold-Chiari malformation. *Concepts Pediatr Neurosurg*. 1988;8:70–75

36. Begeer JH, Meihuizen de Regt MJ, HogenEsch I, Ter Weeme CA, Mooij JJ, Vencken LM. Progressive neurological deficit in children with spina bifida aperta. *Zeit Kinderchir*. 1986;41(suppl 1):13–15

37. Tamaki N, Shirataki K, Kojima N, Shouse Y, Matsumoto S. Tethered cord syndrome of delayed onset following repair of myelomeningocele. *J Neurosurg*. 1988;69:393–398

38. Kaplan WE, McLone DG, Richards I. The urological manifestations of the tethered spinal cord. *J Urol*. 1988;140:1285–1288

39. McLone DG, Herman JM, Gabrieli AP, Dias L. Tethered cord as a cause of scoliosis in children with a myelomeningocele. *Pediatr Neurosurg*. 1990–91;16:8–13

40. Vernet O, Farmer J-P, Houle A-M, Montes JL. Impact of urodynamic studies on the surgical management of spinal cord tethering. *J Neurosurg*. 1996;85:555–559

41. Herman JM, McLone DG, Storrs BB, Dauser RC. Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord. *Pediatr Neurosurg*. 1993;19:243–249

42. Reigel DH, Tchernoukha K, Bazmi B, Kortyna R, Rotenstein D. Change in spinal curvature following release of tethered spinal cord associated with spina bifida. *Pediatr Neurosurg*. 1994;20: 30–42

43. Rotenstein D, Reigel DH, Lucke JF. Growth of growth hormone-treated and nontreated children before and after tethered spinal cord release. *Pediatr Neurosurg*. 1996;24:237–241

44. Hendrick EB, Hoffman HJ, Humphreys RP. The tethered spinal cord. *Clin Neurosurg*. 1982;30:457–463

45. Hoffman HJ. The tethered spinal cord. In: Holtzman R, Stein BM, eds. *The Tethered Spinal Cord*. New York, NY: Thieme-Stratton; 1985:91–98

46. Moufarrij NA, Palmer JM, Hahn JF, Weinstein MA. Correlation between magnetic resonance imaging and surgical findings in the tethered spinal cord. *Neurosurg*. 1989;25:341–346

47. McLone DG, Naidich TP. The tethered spinal cord. In: McLaurin RL, Venes JL, Schut L, Epstein F, eds. *Pediatric Neurosurgery*. Philadelphia, Pa: WB Saunders; 1989:71–96

48. Bakker-Niezen SH, Walder HAD, Merx JL. The tethered spinal cord syndrome. *Zeit Kinderchir*. 1984;39(suppl 2):100–103

49. Chaseling RW, Johnston IH, Besser M. Meningoceles and the tethered cord syndrome. *Child Nerv Syst*. 1985;1:105–108

50. Scott RM. Delayed deterioration in patients with spinal tethering syndromes. In: Holtzman R, Stein BM, eds. *The Tethered Spinal Cord*. New York, NY: Thieme-Stratton; 1985:116–120

51. Cameron AH. The Arnold-Chiari and other neuro-anatomical malformations associated with spina bifida. *J Path Bact*. 1957;73: 195–211

52. Mackenzie NG, Emery JL. Deformities of the cervical cord in children with neurospinal dysraphism. *Dev Med Child Neurol.* 1971;13:59–67

53. La Marca F, Grant JA, Tomita T, McLone DG. Spinal lipomas in children: outcome of 270 procedures. *Pediatr Neurosurg.* 1997; 26:8–16

54. Hall PV, Campbell RL, Kalsbeck JE. Meningomyelocele and progressive hydromyelia. Progressive paresis in myelodysplasia. *J Neurosurg.* 1975;43:457–463

55. Hall P, Lindseth R, Campbell R, Kalsbeck JE, Desousa A. Scoliosis and hydrocephalus in myelocele patients. The effects of ventricular shunting. *J Neurosurg.* 1979;50:174–178

56. Oldfield EH, Muraszko K, Shawker TH, et al. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. *J Neurosurg.* 1994;80:3–15

Commentary

Orthopedic problems associated with myelomeningocele involve the spine and lower extremities. The incidence and severity of orthopedic problems depends on the level of involvement of myelomeningocele. Children who have thoracic level myelomeningocele are likely to develop significant spinal and lower extremity deformity compared with a child who has sacral level involvement and is unlikely to have spinal deformity with little lower extremity involvement other than foot deformity and weakness.

Hypotrophic or absent posterior elements of the spine (spinous process, interspinous ligaments, lamina, and facet joints) are inherent in myelomeningocele. This anatomic defect can lead to abnormal kyphosis in the normally lordotic lumbar spine. The kyphosis may be present at birth and progress during the first few postnatal years. Brace treatment is of little value and may contribute to soft-tissue ulceration over the prominent kyphosis. Severe kyphosis may make sitting impossible without using the hands for balance, which is functionally limiting. Treatment of progressive rigid kyphosis is surgical, usually involving resection of vertebrae and placement of spinal rods.

Scoliosis also is common, especially in children who have thoracic myelomeningocele. Bracing is of questionable value and may decrease mobility and pulmonary function. Curves greater than 50 degrees may require surgical fusion. Any significant spinal deformity in a low lumbar myelomeningocele, new onset of back pain, or quickly progressive spinal deformity may be red flags for an underlying change in the neural elements such as an Arnold-Chiari, syrinx, or tethered cord and may warrant MRI of the entire spine.

The potential for independent ambulation is related closely to the level of involvement. Traditional teaching is that independent ambulation is possible if the quadriceps are strong (L3 or L4 level myelomeningocele). Long-term studies suggest that ambulation usually degenerates over a lifetime because the individual's muscle strength does not keep up with increasing body size, and neurologic deterioration is possible. Bracing often is helpful. For example, for a child who has weak quadriceps, a knee-ankle-foot orthosis helps the knee remain straight and bear weight during ambulation, often with the use of a walker. Contractures of the hips, knees, and ankles are common and require vigilant therapy, positioning and braces, and at times, surgery.

Hip dislocations are common in children who have thoracic and high lumbar myelomeningoceles. In general, the orthopedic community has moved away from aggressive surgical relocation of hips in children who have myelomeningoceles because dislocated hips are not painful and do not limit ambulation or general function. Knee pain is common and often unexplained, although it is believed to be due to abnormal forces during ambulation resulting from muscle imbalance and a Trendelenburg gait.

Feet tend to go into equinus, although a variety of deformities are common. A goal of orthopedic treatment is a braceable, plantigrade foot. Foot infections are particularly reticent to care and may lead to an amputation. Because the feet are insensate, particular care must be taken to recognize the early development of ulcers or skin breakdown, which occur frequently.

David L. Skaggs, MD
Editorial Board

PIR Quiz

Quiz also available online at www.pedsinreview.org.

5. Intelligence is normal (IQ, >80) in 75% of children who have myelomeningocele and does not appear to be affected by the presence or absence of hydrocephalus. Of this group, a learning disability (usually nonverbal) will be present in:

A. 15%.
B. 30%.
C. 45%.
D. 60%.
E. 75%.

6. A 2-year-old boy who has myelomeningocele develops symptoms of difficult swallowing, inspiratory stridor, and a hoarse cry. These symptoms are *most* consistent with a diagnosis of:

A. Chiari II malformation.
B. The natural history of the disorder.
C. Shunt malfunction.
D. Spinal cord tethering.
E. Syringomyelia.

7. A 9-year-old girl who has myelomeningocele describes new symptoms of bladder incontinence, deteriorating gait, and scoliosis. These symptoms are *most* consistent with a diagnosis of:

A. Chiari II malformation.
B. The natural history of the disorder.
C. Shunt malfunction.
D. Spinal cord tethering.
E. Syringomyelia.

8. A 13-year-old boy who has myelomeningocele complains that he cannot play baseball as well as he did last year. His arms are "stiff," but his hands feel "weak." These symptoms are *most* consistent with a diagnosis of:

A. Chiari II malformation.
B. The natural history of the disorder.
C. Shunt malfunction.
D. Spinal cord tethering.
E. Syringomyelia.

9. A 10-year-old girl who has myelomeningocele always has been a good student. Her latest report card shows declining grades, poor handwriting, and behavioral changes. These symptoms are *most* consistent with a diagnosis of:

A. Chiari II malformation.
B. The natural history of the disorder.
C. Shunt malfunction.
D. Spinal cord tethering.
E. Syringomyelia.

Neurosurgical Management of Myelomeningocele (Spina Bifida)

Mark S. Dias

Updated Information & Services	including high resolution figures, can be found at: http://pedsinreview.aappublications.org/content/26/2/50
References	This article cites 44 articles, 4 of which you can access for free at: http://pedsinreview.aappublications.org/content/26/2/50#BIBL
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): **Fetus/Newborn Infant** http://pedsinreview.aappublications.org/cgi/collection/fetus:newborn_infant_sub **Rheumatology/Musculoskeletal Disorders** http://pedsinreview.aappublications.org/cgi/collection/rheumatology:musculoskeletal_disorders_sub
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: /site/misc/Permissions.xhtml
Reprints	Information about ordering reprints can be found online: /site/misc/reprints.xhtml

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™