Caring for the Child with Spina Bifida

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KEYWORDS

• Spina bifida • Myelomeningocele • Hydrocephalus • Chiari 2 malformation
• Transition of care

KEY POINTS

- Prenatal consultation with a specialty team, including maternal-fetal medicine specialists and neurosurgeons, can help parents understand what to expect and can reduce anxiety.
- Fetal myelomeningocele closure has been shown to reduce the rate of hydrocephalus in well-selected patients.
- Hydrocephalus occurs in approximately 80% of children with myelomeningocele. Signs and symptoms of hydrocephalus include headache, vomiting, lethargy, as well as other more subtle symptoms.
- Chiari 2 malformation is present in nearly all children with myelomeningocele. If brainstem symptoms develop, the first priority is to assure that hydrocephalus is adequately treated.
- Interdisciplinary clinics, including neurosurgery, orthopedics, rehabilitation medicine, urology, and other specialists, provide optimal care for children with spina bifida.

INTRODUCTION

Myelomeningocele (MMC) is one of the most complex congenital conditions that pediatricians will provide care for in their practice. It is also the most common, with an annual incidence of approximately 8 to 10 cases per 10,000 live births in North America. The incidence correlates directly with maternal red blood cell folate levels, so folic acid fortification remains an important public health opportunity in parts of the world where fortification still does not occur.

The characteristic lesion in MMC is the open blister–like defect over the spine, but the full spectrum of manifestations is broad and includes both cranial and spinal anomalies. More than 20 structural abnormalities of the nervous system characteristically accompany MMC. More importantly, the aberrant neurologic function arising from these anomalies imparts dysfunction of a variety of normal reflexes that

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Pediatr Clin N Am 68 (2021) 915–927
https://doi.org/10.1016/j.pcl.2021.04.013
pediatric.theclinics.com

predominantly but not exclusively affect the lower extremities and sphincters (bowel/bladder). As such there are a wide variety of clinical manifestations of MMC. Children often have multiple health concerns that require care from many medical disciplines, including Neurosurgery, Urology, Orthopedics, Physical Medicine and Rehabilitation, Developmental Pediatrics, and Sleep Medicine as well as Orthotists, Nurse Clinicians and Physical, and Occupational and Speech Therapists. Because of the complexity of care and the inherent interconnectedness of problems, care coordination is the central concept for the most highly effective clinics.

The pediatrician plays a crucial role in care of children with MMC. Although multidisciplinary clinics may be optimally designed to provide for the common needs that are specific to children with MMC, children with this condition also must have a primary care provider (PCP). Children with MMC and other forms of spina bifida require routine well-child care, as well as care for conditions that are outside the expertise of the specialists. Naturally, the PCP assures a holistic, patient-centered approach to care that can be at risk in highly subspecialized care models. The purpose of this chapter is to review the neurosurgical care of children with MMC, focusing on information that a PCP should be familiar with to serve as partner in the child's comprehensive care team.

DEFINITIONS

Spina bifida is a nonspecific term that refers to many distinct abnormalities of the spinal cord. Another collective term that is used for this family of conditions is spinal dysraphism. Fundamentally, all of these abnormalities are problems of the embryologic development of the spinal cord. The most severe type is MMC. This is caused by a failure of the primary closure of the neural tube, leading to an open neural placode on the infant's back, with no overlying skin coverage (Fig. 1). Neurologic function below the level of the lesion is lost—this is the most common type of spinal dysraphism. The term "spina bifida" is often used colloquially to describe MCC. However, for the purposes of this manuscript, the authors refer to open, non–skin covered spinal dysraphism as MMC.

Closed types of spinal dysraphism are caused by several different embryologic abnormalities. These abnormalities can be dramatic and obvious at birth, such as a large terminal myelocystocele (Fig. 2). Or they might show no outward signs of abnormality. Table 1 shows many different types of congenital abnormalities that fall under the

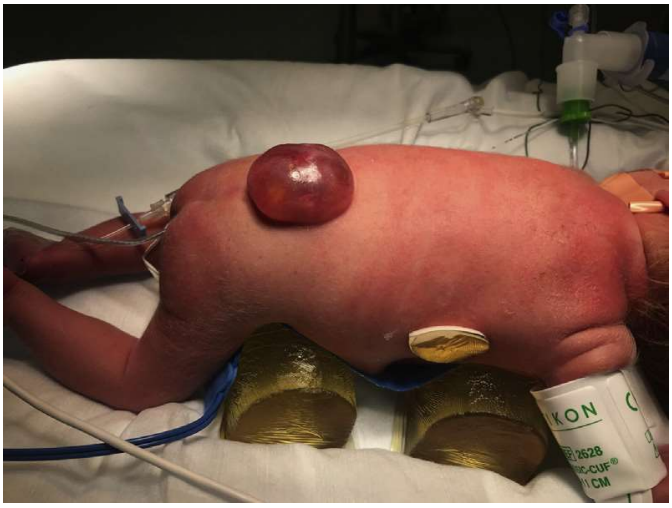

Fig. 1. A child with a cystic myelomeningocele.

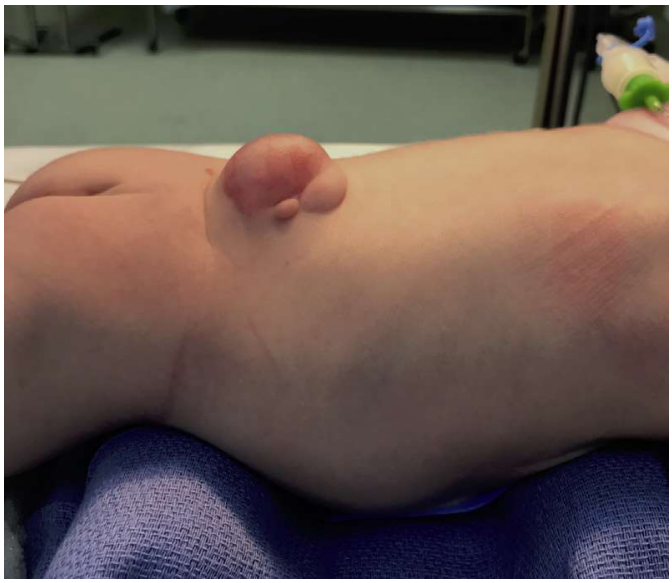

Fig. 2. A child with a skin-covered terminal myelocystocele.

broad term "spina bifida." Most of this chapter focuses on the care of children with MMC, with discussion of some specific issues surrounding other types of spina bifida. For a more detailed discussion of occult spinal dysraphism, the reader is referred to *Chapter 14: Cutaneous stigmata of the spine: a review of indications for imaging and referral*.

DIAGNOSIS

Prenatal Diagnosis

Initial diagnosis of MMC is usually triggered by elevated alpha-fetoprotein (AFP) level in maternal serum, performed for screening early in the second trimester. AFP is elevated in about 80% of cases. Diagnosis is confirmed on second trimester ultrasound, between the 18th and 24th week of gestation. High-resolution ultrasound is

Table 1
Types of spinal dysraphism

Diagnosis	Characteristics	Other Terms
Open Spinal Dysraphism		
Myelomeningocele	Open spinal cord, no skin covering	Spina bifida aperta
Closed Spinal Dysraphism		
Lipomyelomeningocele	Fatty mass, skin covered	Spinal lipoma
Split cord malformation	Duplicated spinal cord, focal hypertrichosis (hairy patch)	Diastematomyelia, diplomyelia
Terminal myelocystocele	Fluid-filled mass, skin covered	
Dermal sinus tract	Sacral dimple	Fibrovascular tract
Limited dorsal myeloschisis	Sacral dimple	Meningocele manqué
Neurenteric cyst	Ventral attachment to spinal cord, minimal cutaneous findings	
Spina bifida occulta	Incomplete closure of posterior bony arch of lower lumbar vertebrae. No clinical consequences	

often able to distinguish open MMC from other types of spina bifida, determine the anatomic level of the abnormality, and determine the presence of associated findings such as hydrocephalus. The Spina Bifida Association recommends that parents with a new diagnosis of MMC undergo prompt consultation with specialists who are dedicated to providing care to individuals with MMC and will be part of their long-term care team.[1] This consultation is often done with a collaborative, interdisciplinary approach to review the expectations for initial newborn management, including surgical closure of the defect as well as the long-term expectations for function.

Estimation of Lesion Level

Although it is impossible to predict a child's neurologic outcome before birth, there are generalizations that can be made based on the type of spina bifida and the anatomic level of the abnormality seen on ultrasound. Ultrasound-based estimation of the anatomic location of the lesion is typically accurate within 1 to 2 spinal levels. Therefore, estimations about future functions, such as ambulation and bowel and bladder function, can be provided during prenatal discussions. **Table 2** provides information about the likelihood of community ambulation for each functional lesion level for children with spina bifida.

This is a time of high anxiety for families. Learning accurate information can ameliorate some of the fear surrounding the diagnosis. The care team during the prenatal period can also aid the family in determining if they are a good candidate for prenatal MMC closure.

PRENATAL CLOSURE OF MYELOMENINGOCELE

Publication of the Management of Myelomeningocele Study (MOMS) in 2011 was a watershed moment for neurosurgical care of children with MMC. MOMS was a randomized controlled trial comparing standard open MMC closure with fetal repair.[2] The primary outcome was a composite of death or development of hydrocephalus requiring surgical treatment. Final results showed significantly fewer children reaching the composite endpoint in the fetal surgery group versus the standard care group. More impressively, although 82% of children in the postnatal surgery group underwent ventriculoperitoneal shunt placement for hydrocephalus, only 40% in the fetal surgery group were shunted. Other observed benefits to fetal surgery included improvement in motor function, given the anatomic level of the MMC defect, less severe hindbrain herniation (Chiari 2 malformation), and improved cognitive function. Fetal surgery was also associated with a higher rate of preterm delivery and uterine dehiscence than postnatal repair.

Based on these largely favorable results, many programs have developed fetal surgery centers, and the technique for fetal surgery has continued to evolve.[2–5] On the

Table 2
Myelomeningocele lesion level definition and likelihood of community ambulation

Lesion Level	Definition	Likelihood of Community Ambulation (%)
Thoracic	Flaccid lower extremities	<1
High-lumbar	Hip flexion present	7
Midlumbar	Knee extension present	44
Low-lumbar	Foot extension (dorsiflexion) present	79
Sacral	Foot plantar flexion present	95

other hand, the MOMS trial had very strict inclusion criteria. At many current centers, patients who would not have been candidates for enrollment may be offered fetal surgery. In addition, long-term follow-up shows that there may be unintended consequences for mothers after fetal surgery, such as problems with subsequent pregnancies. Detailed discussion of the nuances of fetal surgery is beyond the scope of this text. However, it is crucial for pediatricians to know that the option for fetal repair exists and that parents who are considering fetal surgery should be referred for specialist consultation.

PERINATAL CARE OF MYELOMENINGOCELE

Obstetric Care

For the infant who is not undergoing fetal surgery, routine obstetric care is delivered throughout the third trimester. Current recommendation is for scheduled cesarean delivery at 37 or 38 weeks.[6] However, this remains an area of controversy, and vaginal delivery remains a viable option. Although strong practice preferences exist in obstetrics, high-quality evidence demonstrating clear superiority of one route of delivery over another does not exist. Parents should be encouraged to discuss these issues with their obstetricians.

Neonatal Care and Myelomeningocele Surgery

After delivery, the child with MMC is admitted to the neonatal intensive care unit. Screening for other congenital abnormalities is typically performed, including echocardiogram and renal ultrasound. Surgical closure of the MMC defect is performed within 24 to 48 hours of birth. Before surgical closure, the child should be positioned prone, and the exposed placode kept covered and moist. Technical details of surgical closure are beyond the scope of this text, but briefly, the goals are to close the dura and skin over the neural elements. With large MMC defects, various techniques may be necessary to achieve adequate coverage of the defect with skin closure.[7]

Children born with a skin-covered lesion or closed form of spina bifida often will not have an initial surgery while in the neonatal intensive care unit; however, it is recommended that these children undergo similar monitoring and education to establish future treatment plans. Bowel and bladder management should be discussed even in the cases where surgical intervention is delayed.

NEUROSURGICAL CARE FOR THE CHILD WITH MYELOMENINGOCELE

The neurosurgical care of children with MMC centers around 3 distinct conditions with potential surgical treatment: hydrocephalus, Chiari 2 malformation, and tethered spinal cord.

Hydrocephalus

Incidence

Hydrocephalus is an abnormal accumulation of cerebrospinal fluid in the ventricles of the brain. It is the most common condition treated by pediatric neurosurgeons. Although there are many causes of hydrocephalus that differ in prevalence, MMC is one of the most common causes of hydrocephalus. Published data from small series note that hydrocephalus develops in 50% to 90% of children with MMC. In a national sample of more than 4000 children, enrolled in the National Spina Bifida Patient Registry (NSBPR), 80% of children had hydrocephalus requiring surgical treatment.[8] Furthermore, children with higher lesion level (thoracic or upper lumbar functional level) were found to have greater odds of having hydrocephalus compared with lower

levels. It is very uncommon for closed spinal dysraphism (non-MMC forms of spina bifida) to be associated with hydrocephalus.

The primary outcome from the MOMS trial of fetal MMC closure was a complex composite outcome including death, cognitive test scores, and meeting criteria for the treatment of hydrocephalus. The trial was stopped early due to significantly better results in the fetal surgery group. A simple examination of the proportion of children who were treated for hydrocephalus in each group illustrates the important point: 40% of children with fetal surgery later underwent shunt placement for treatment of hydrocephalus compared with 82% in the postnatal surgery group.[2] Therefore, one of the primary potential benefits of fetal surgery is reduction in the need for treatment of hydrocephalus.

Signs and symptoms in infants

In infants, before closure of the cranial sutures, hydrocephalus produces characteristic signs, most importantly accelerated head growth and bulging of the anterior fontanelle (Fig. 3). Additional signs and symptoms may include impaired eye movements, especially paralysis of upward gaze, apnea or bradycardia, lethargy, and vomiting. The most severe symptom of hydrocephalus in newborns with MMC is stridor, which may exist in isolation or may accompany other signs of bulbar dysfunction, such as poor swallowing or secretion control. Inspiratory stridor in the newborn with MMC is a clinically urgent sign of brainstem distress. The acute threat is laryngeal dysfunction and respiratory obstruction from relaxation of the vocal cords. Hydrocephalus is a global stressor for the compromised nervous system, which is thought to tip a tenuous system out of balance and result in brainstem failure. This brainstem failure becomes manifest as stridor, a sirenlike wailing cry, and impairment of management of normal oral secretions. Therefore, stridor in an infant with MMC is considered an urgent indication for hydrocephalus treatment.

Signs and symptoms in older children

In older children, with fused cranial sutures, the primary symptoms of hydrocephalus are headache, nausea/vomiting, and lethargy. Importantly, children with MMC may have additional symptoms of hydrocephalus that are not common in children without MMC. These can include lower cranial nerve dysfunction, sleep apnea, swallowing dysfunction, or leakage of cerebrospinal fluid (CSF) from the MMC closure site. As stridor, lower cranial nerve dysfunction, swallowing problems, and sleep apnea are likely consequences of vulnerability of the brainstem, which is a result of the Chiari 2 malformation. When a child with MMC displays these symptoms, the first line of treatment is directed at hydrocephalus. Surgical decompression of the posterior fossa (Chiari 2

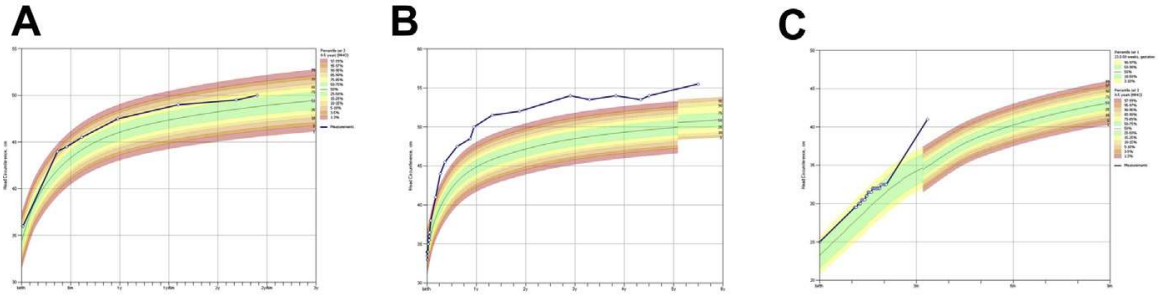

Fig. 3. (*A*) A normal head growth curve. (*B*) A macrocephalic but reassuring head growth curve. Note that although this represents a larger-than-average head, the growth trajectory is parallel to the normal curve. (*C*) A concerning head growth curve, with head growth trajectory crossing percentiles.

decompression) is only indicated once adequate treatment of hydrocephalus has been assured. In an older child or adolescent with MMC, inadequately treated hydrocephalus may also present as neck or back pain (Table 3).

Treatment

Ventriculoperitoneal shunting. The goal of hydrocephalus treatment is to maximize the growth and development potential of the child's brain. The mainstay of treatment is the ventriculoperitoneal (VP) shunt. A VP shunt consists of a ventricular catheter, a valve that regulates CSF flow and a distal catheter placed into the peritoneum. The VP shunt was invented and first used in the 1950s and has revolutionized the care of children with hydrocephalus. Before shunting, 90% of children born with hydrocephalus could be expected to die before age 10 years, and the other 10% were at high risk for severe developmental delay. Now, the goal for a newborn with hydrocephalus is their brain development and function to be as close to normal as possible (although possibly limited by other conditions). It should be noted that for many children with MMC, even when hydrocephalus is adequately treated, there are learning challenges. Nonverbal learning disorders, problems with fine motor skills, and behavioral difficulties are common.[9] Pediatricians should be aware of these commonly associated conditions, as children may benefit from focused academic assistance.

Although shunts are highly beneficial overall, they have shortcomings. Most importantly, shunts can become obstructed or disconnected, known as shunt failure. For reasons that are not entirely clear, shunt failure is more common in younger children.[10] However, shunt failure can occur at any age, throughout the lifetime. Shunt failure leads to symptoms of elevated intracranial pressure, as described earlier. Symptoms of shunt failure differ from one child to the next, even for multiple shunt failures in the same child. For some children, shunt failure may manifest with subtle symptoms, such as mild headache or decline in school performance. For others, shunt failure may be life threatening, with coma onset within minutes or hours. Most commonly, in a child who has multiple shunt failures, the failures will have similar symptoms. However, it is possible that a child will have different symptoms from one failure to the next. Therefore, providers caring for these children must be vigilant for all signs of shunt failure.

Table 3
Signs and symptoms of hydrocephalus

Infants	• Increasing head circumference • Vomiting/poor feeding tolerance • Irritability • Lethargy • Bulging fontanelle • Separated cranial sutures • Impaired eye movements (upgaze palsy) • Apnea or bradycardia
Older Children	• Headache • Nausea/vomiting • Lethargy • Bradycardia/hypertension • Eye movement abnormalities
Symptoms Unique to Myelomeningocele	• Stridor (infants) • Neck pain • Back pain

For most children, if the shunt fails, the cerebral ventricles will become larger. However, in some cases, there is no change in ventricle size. In these cases, the clinical presentation alone must guide decisions about surgical exploration of the shunt. A more detailed discussion of hydrocephalus can be found in Chapter 8: *Hydrocephalus and the Primary Care Provider.*

Endoscopic third ventriculostomy. Endoscopic third ventriculostomy (ETV) is an alternative procedure for treating hydrocephalus that does not require implanted hardware. ETV involves the use of a neuroendoscope to make a fenestration of the floor of the third ventricle, allowing passage of CSF from the ventricle into the prepontine cistern. Traditionally, ETV was used only as a treatment of obstructive hydrocephalus, such as that caused by posterior fossa tumors or stenosis of the cerebral aqueduct. The most important limitation of ETV is that it does not successfully treat hydrocephalus in all patients. Younger children and those whose hydrocephalus is caused by intraventricular hemorrhage or MMC are less likely to be adequately treated by ETV.[11] Recently, with the addition of choroid plexus cauterization (CPC), the combined procedure (ETC/CPC) has been shown to be successful in up to 70% of children with MMC.[12] Again, a more detailed discussion of ETV and ETV/CPC can be found in Chapter 8: *Hydrocephalus and the Primary Care Provider*. Children treated with ETV are not cured of their hydrocephalus, but rather they depend on the patent ETV channel. Therefore, they could become symptomatic if the ETV were to close. Although this is less common than shunt failure, it is still important to consider hydrocephalus as a possible problem, should these children develop symptoms that would be concerning for shunt failure.

Clinics Care Points

- Pediatricians should be aware of the signs and symptoms of hydrocephalus, in particular those that are unique to children with MMC, such as stridor.
- Pediatricians should recognize that these children are always at risk for recurrence of symptoms.
- Any signs or symptoms that are possibly related to hydrocephalus should be evaluated by a neurosurgeon.

Chiari 2 Malformation

The Chiari 2 malformation was originally described by Hans Chiari, along with his descriptions of other forms of hindbrain abnormalities that now bear his name.[13] The Chiari 2 malformation is fundamentally different from the Chiari 1 malformation, which is discussed elsewhere in this text (*Chapter 7: Chiari malformation*). According to the unified theory originally proposed by McLone, the open neural tube in MMC allows for venting of CSF during gestation. Therefore, the posterior fossa does not expand to its normal proportions, resulting in displacement of hindbrain tissue (brainstem and cerebellum) outside of the foramen magnum.[14] In addition to hindbrain herniation, there are numerous anatomic abnormalities that have been described as part of the Chiari 2 malformation, including beak shape of the midbrain tectum, interdigitation of the cerebral gyri, and disproportionate dilation of the posterior horns of the lateral ventricles (colpocephaly). It is typically thought that, although the degree of hindbrain herniation may vary, all children with MMC are born with some degree of Chiari 2 malformation present.

Signs and symptoms

The clinical manifestations of the Chiari 2 malformation can generally be attributed to abnormalities of the function of the brainstem. Symptoms include stridor, vocal cord

paralysis, opisthotonos, sleep-disordered breathing (SDB), swallowing difficulties, absent gag reflex, or other lower cranial nerve palsies. In general, the severity of hindbrain herniation seen on imaging studies does not correlate with the severity of symptoms. Therefore, attention to the clinical situation is far more important than the imaging appearance of the posterior fossa.

In infants with MMC, one of the most important clinical signs to recognize is inspiratory stridor. Stridor in this setting is thought to occur because of impairment of vocal cord function, leading to adducted vocal cords on inspiration. There are many causes of stridor in infants.[15] Pediatricians and pediatric emergency medicine physicians may be accustomed to evaluating an infant with stridor. However, in the setting of MMC, stridor is typically considered a harbinger of potentially life-threatening brain stem dysfunction. Urgent neurosurgical evaluation and treatment is indicated.[16]

Treatment

First-line therapy for brainstem-related symptoms in children with MMC is always directed at assuring adequate treatment of hydrocephalus. The presence of elevated intracranial pressure from undertreated hydrocephalus is thought to place additional stress on the brainstem and lead to Chiari 2 symptoms. If symptoms persist, despite adequate hydrocephalus treatment, direct decompression of the Chiari 2 malformation can be performed, which involves surgical removal of the bone of the posterior foramen magnum, the posterior arch of the C1 vertebra, and sometimes additional removal of more caudal lamina. There is ongoing debate about whether expansile duraplasty in addition to bony decompression is beneficial. In a national sample of more than 4000 children with MMC, approximately 10% had undergone Chiari 2 decompression surgery.[17] More rostral lesion levels (thoracic level MMC, more than lumbar level, more than sacral level) were more likely to have undergone Chiari 2 decompression. This study, based on a large sample, also showed that children who underwent Chiari 2 decompression when younger than 2 years were more likely to also require tracheostomy. One interpretation of this finding is that some children with MMC have a congenitally dysfunctional brainstem, leading them to require tracheostomy for airway protection regardless of other interventions. Nevertheless, the treatment algorithm for Chiari 2 symptoms in modern use remains as follows: (1) treat hydrocephalus, (2) decompress Chari 2 malformation, and (3) consider additional support such as tracheostomy as needed.

It has recently been recognized that children with MMC may be at high risk for SDB[18,19]; this is another likely manifestation of the Chiari 2 malformation. Pediatricians should be alert for symptoms such as snoring, excessive daytime sleepiness, or irritability. However, even in the absence of these symptoms, because of the high prevalence of SDB among children with MMC, referral for screening polysomnography may be indicated.

Clinics Care Points

- Pediatricians should be aware of the manifestations of the Chiari 2 malformation, such as stridor, cranial nerve dysfunction, swallowing difficulties, or sleep apnea.
- There should be a low threshold for referral to neurosurgery if any of these symptoms are noted.

Tethered Cord

Definition

Tethered spinal cord, or tethered cord, refers to the concept that the spinal cord naturally exists within the thecal sac, bathed in CSF, and without any significant

attachment to the surrounding tissue. Any type of spinal dysraphism can act as an attachment of the spinal cord to the surrounding structures, thus "tethering" the cord. The prevailing theory is that a differential between spinal growth or lengthening and somatic growth then results in traction on the tethered spinal cord, leading to a loss of function. When this manifests clinically, it is usually with back pain, leg pain, or loss of neurologic function in the legs and/or bladder.[20,21] In the case of MMC, the initial closure of the back is typically performed within 1 to 2 days of birth, but scar at the site of closure can result in spinal cord tethering later in life.

Diagnosis

For children with MMC, imaging studies are not useful in determining whether a spinal cord is tethered. Essentially all MRI scans of children who have a history of MMC closure will seem to be "tethered." Thus, the determination of whether a patient is experiencing a clinically relevant tethered cord must be made based on signs and symptoms alone. As noted earlier, typical symptoms are back pain, leg pain, and neurologic deficit referable to the caudal spinal cord, typically leg weakness or sensory loss or loss of bladder function (**Box 1**).

Treatment

Surgical treatment of tethered cord is referred to as tethered cord release (TCR). This procedure has wide variability in risk profile, depending on the complexity of the tethering lesion. Therefore, decisions about whether to undertake a TCR in any given patient must consider the severity and progressive nature of the symptoms as well as the anatomy of the tethering lesion. Furthermore, there must be clear understanding between surgeon and patient/family about the goals of surgical treatment. In general, surgical untethering is effective at relieving back and leg pain and for arresting progression of deficit but not effective at reversing neurologic deficit. Therefore, outside of surgeries performed because of pain, the TCR is a prophylactic operation, performed to reduce the risk of worsening function. Careful explanation of these factors is crucial for appropriate shared decision-making when considering a TCR in a patient.

Clinics Care Points

- Tethering of the spinal cord can occur after initial closure of MMC.
- Surgical release of the tethered cord can be performed to alleviate pain or prevent worsening neurologic function.

Interdisciplinary Spina Bifida Care

The Spina Bifida Guidelines produced by the Spina Bifida Association places a strong emphasis on interdisciplinary lifespan care for individuals with spina bifida.[22] Lifespan care can be characterized as a mechanism for delivering care where the provider embraces the idea that, although they might not be involved in the individual's care throughout their entire lifespan, the care that is delivered will affect the patient across the lifespan. Therefore, every effort should be made to promote independence and overall wellness and to empower the patient to reach their highest potential and quality of life. This care is optimal when delivered longitudinally across a continuum that begins during the prenatal consultation and continues through pediatric and then eventually to transition into adult years.[23]

A multidisciplinary approach is recognized as best practice among spina bifida clinics throughout North America. Most clinics include Neurosurgery, Urology, Orthopedics, Physical Medicine and Rehabilitation, and Development Pediatrics. A collaborative interdisciplinary approach to care has been linked to improved health

Box 1
Signs and symptoms of tethered spinal cord

- Back pain
- Leg pain
- Leg weakness
- Worsening of bladder function
- Decreased leg sensation
- Scoliosis

outcomes, decreased morbidity and mortality, and decreased cost of care for individuals with spina bifida.[24] A central component of multidisciplinary care includes the access to individualized care coordinator. Quality care coordination is an essential part of the multidisciplinary care team. The coordinator is often the link between the community pediatrician and the specialist medical team.

Transitional Care

In the United States there are 1500 new births of children with spina bifida every year, and 75% of those children will now survive well into adulthood.[25] Transition of care is not unique to individuals with spina bifida. There are many individuals with chronic conditions of childhood surviving into adulthood, which has resulted in the establishment of transition as a core outcome in the Healthy People Maternal Child Health Bureau (MCHB), as well as a focus of the American Academy of Pediatrics (AAP). The AAP has embraced the importance of good transitional care and has provided guidance and leadership under the "Got Transition?" initiative. Transition of care is a process that begins early in adolescence and involves more than transferring care and/or changing providers. Health care transition is defined as the organized process of supporting youth in acquiring independent health care skills, preparing for an adult model of care, and transferring to new providers without disruption of care. The goal of health care transition is to optimize health and assist youth in reaching their full potential.

This process of health care transition involves working with youth and their families/caretakers beginning by age 13 to 14 years. The care changes from a "pediatric" model of care where the parents make most of the decisions to an "adult" model of care where youth being taking responsibility for their decision-making and self-care. Transition of care has evolved and grown into a separate domain in Pediatric care for which the details exceed the scope of this chapter. However, a recognition that all efforts expended by the patient, family, and provider team results in an adult with successful strategies and capabilities to participate in the full range of adult spina bifida care gives rise to an awareness of the overarching importance of transition readiness throughout the scope of care.

DISCLOSURE

The authors have nothing to disclose.

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