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Guidance for the Clinician in Rendering Pediatric Care

CLINICAL REPORT

# Providing a Primary Care Medical Home for Children and Youth With Spina Bifida

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**KEY WORDS**
spina bifida, developmental disability, medical home, chronic condition, hydrocephalus, myelomeningocele, meningomyelocele

**ABBREVIATIONS**
NTD—neural tube defect
AFP—$\alpha$-fetoprotein
UTI—urinary tract infection

The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.



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## abstract

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The pediatric primary care provider in the medical home has a central and unique role in the care of children with spina bifida. The primary care provider addresses not only the typical issues of preventive and acute health care but also the needs specific to these children. Optimal care requires communication and comanagement with pediatric medical and developmental subspecialists, surgical specialists, therapists, and community providers. The medical home provider is essential in supporting the family and advocating for the child from the time of entry into the practice through adolescence, which includes transition and transfer to adult health care. This report reviews aspects of care specific to the infant with spina bifida (particularly myelomeningocele) that will facilitate optimal medical, functional, and developmental outcomes. *Pediatrics* 2011;128:e1645–e1657

## INTRODUCTION

Myelomeningocele is a complex chronic condition that affects the child and the family as well as the health care and related service providers. Although the occurrence of spina bifida* is decreasing, every year more than 1500 children are born with spina bifida in the United States. More than 160 000 Americans younger than 18 years are affected by spina bifida, and the 30-year survival rate has improved to nearly 90%.[1] Diagnosis of spina bifida and other neural tube defects (NTDs) is now often made early in pregnancy through $\alpha$-fetoprotein (AFP) screening and fetal ultrasonography, which provides time for decision-making and planning by families.

A child born with spina bifida faces a long and multifaceted path of medical and surgical care. The complexity and severity of spina bifida depends on the type and location of the defect as well as the occurrence of associated conditions. The average lifetime cost is more than $635 000.[2] Children 1 through 17 years of age with spina bifida have average medical expenditures 13 times greater than children without spina bifida.[3] The primary care management of spina bifida in the medical home provides an opportunity to optimize the child's outcomes with improved quality of care in a cost-effective, family-centered, coordinated system.

*Note: in this report, the term "spina bifida" is used interchangeably with "myelomeningocele."

## BACKGROUND

### Etiology and Risk

Spina bifida, anencephaly, and other NTDs occur as a result of defective neurulation or closure during the third week after conception of the embryonic neural fold, which becomes the neural tube. Failure of closure in a cephalic direction leads to the development of anencephaly; failure of closure in the caudal direction leads to spina bifida. Myelomeningocele arises as a failure of neurulation by approximately day 28 after conception. The primary defect in the neural tube then leads secondarily to the failure in the formation of portions of the spinal cord and the dorsal elements of the vertebral bodies and overlying tissues, which gives rise to the spina bifida sac. A lesion that involves elements of the spinal cord as well as the meninges within the sac is termed a myelomeningocele (or meningomyelocele). If the meninges but not the neuronal elements are involved, the lesion is classified as a meningocele. These 2 lesions represent open NTDs. Closed NTDs are those in which the overlying skin is intact and include lipomeningocele and lipomyelomeningocele as well as occult spina dysraphisms (Table 1).

The etiology of spina bifida includes genetic and environmental factors.[4] Spina bifida occurs worldwide and in all racial groups, although there are geographic and ethnic variations.[5] Genetic factors are likely to be related to the increased risk in some populations, notably the Irish, Scottish, and other northern Europeans.[6] This risk may be related to altered folate metabolism.[7] In the United States, a slightly higher rate occurs in families of Latin descent.[8] Chromosome disorders including trisomy 13 or 18 and microdeletion of 22q11 are linked to NTDs.[9] Exposure to some prenatal medications increases the risk of spina bifida. These medications include valproic acid, carbamazepine, isotretinoin, methotrexate and other folic acid antagonists, excess vitamin A or its analogs, and retinoic acid.[10] Maternal nutrition (especially folate deficiency), alcohol consumption, obesity, and fever, either attributable to illness or hot tub/sauna use, can increase risk, as does maternal diabetes mellitus.[11,12] Family history of previous NTDs is a significant risk factor, increasing the risk more than 20-fold (eg, from 0.1%–2.5%).[13,14]

### Occurrence

In the United States, the birth prevalence of spina bifida has decreased to less than 1 in 1000 live births.[15] This reduction might be related to im-

**TABLE 1** Spina Bifida: Types, Description, and Implications

| Spina Bifida Type | Description | Clinical Implications |
|---|---|---|
| Myelomeningocele | Open NTD posterior vertebral defect and extrusion of spinal cord elements into a meningeal sac; location: cervical, thoracic, lumbar, and/or sacral spine; leads to paraplegia and insensitivity below the lesion and neurogenic bowel and bladder; associated defects include structural brain anomalies (Chiari II malformation, hydrocephalus, brainstem dysfunction abnormalities of the cerebrum and corpus callosum, learning disabilities, dislocated hips and clubbed feet) | Complex multisystem disorder that requires ongoing monitoring by spina bifida team, enhanced primary care in the medical home with bidirectional communication and comanagement with the multispecialty spina bifida care team, early-intervention and special education services, physical therapy and adaptive equipment, and developmental and learning monitoring |
| Meningocele | Closed NTD posterior vertebral defect without extrusion of spinal cord elements into a meningeal sac; location: cervical, thoracic, lumbar, and/or sacral spine; motor deficits are less likely than with myelomeningocele; structural brain anomalies and Chiari II malformation are less likely | Early closure of spina bifida sac; follow-up by spina bifida team; additional monitoring in the medical home; ongoing monitoring for neurologic function; early-intervention monitoring; periodic monitoring for possible late onset of neurologic signs |
| Occult spinal dysraphism/lipomeningocele | Closed NTD posterior vertebral defect and with fatty tumor that might contain neuronal elements; location: typically lumbar and/or sacral spine; leads to motor deficits if neuronal elements are involved; associated defects include tethered cord; structural brain anomalies and Chiari II malformation are unlikely | Early neurosurgical intervention; follow-up by spina bifida team; additional monitoring in the medical home; enhanced primary care in the medical home and with follow-up by the multispecialty spina bifida care team; ongoing monitoring for neurologic deficits; early-intervention monitoring |
| Spina bifida occulta | Benign closed NTD posterior vertebral defect only without a meningeal sac; location: lumbar-sacral spine; usually asymptomatic but can be associated with occult spina dysraphism; usually no associated defects | Monitoring and reassurance within the medical home |
| Tethered cord | Traction injury to the distal spinal cord caused by anomalous attachment of the spinal cord, which causes subtle and progressive loss in neural function; can occur with any NTD at any time, but often occurs with growth spurts; might be precipitated by ventricular shunt failure | Monitoring of all children and youth with open or closed NTD for signs of tethering; changes in lower extremity strength, function, or sensation; urinary incontinence or enuresis; changes in bowel function; or worsening scoliosis |

Spina bifida is a general term for several malformations of the spine and its neural elements, which may be associated with neurologic, neuromotor, developmental, and orthopedic anomalies and are related to structural abnormalities of the brain and cranium.

proved prenatal nutrition, including folic acid supplementation for women of childbearing age and the mandatory enrichment of flour with folate as well as the termination of affected fetuses.[16,17] Despite folic acid fortification, infants continue to be born with NTDs and will need both immediate and long-term medical, surgical, and related interventions.[18]

## Clinical Manifestations

The effects of spina bifida relate to the location and size of the defect and the presence of hydrocephalus, brain abnormalities such as the Chiari II malformation, and other neurologic and orthopedic conditions. Thoracic and higher lumbar myelomeningocele lesions are more likely to be associated with significant motor and sensory deficits and structural abnormalities in the lower extremities than are those in the lower lumber or sacral regions.[19] Functional defects of the urogenital and lower intestinal tract are likely at all levels.[20]

Hydrocephalus occurs in ~85% of infants with myelomeningocele and, in most cases, necessitates ventricular shunt placement. Once placed, shunts need to be monitored regularly for potential malfunction. In addition, hydrocephalus is associated with significant problems in learning and cognitive function.[21] After birth, the open spinal lesion needs to be protected from trauma, which could cause additional neurologic damage. In addition, if the lesion is leaking cerebrospinal fluid, measures to protect against infection (meningitis) are required. Surgical closure within 72 hours reduces the risks of infection and additional spinal cord injury.[22,23]

In addition to the primary deficits in motor and sensory function, children and youth with spina bifida experience a range of comorbid conditions including learning disabilities, problems with attention and executive function, dysfunction of upper extremities, strabismus, and seizures. They are also subject to other functional complications such as limitations of movement and ambulation, scoliosis, joint instability, fractures, bowel and bladder dysfunction, altered growth including precocious puberty, and obesity. These children and teenagers will need ongoing prevention because of the increased risk of developing latex allergies.[24] The physical and psychological consequences of impaired mobility and independence, altered appearance, and the long-term needs of the condition also require identification and intervention. The child's physical and developmental disabilities, limited mobility, and chronic health conditions can be barriers to social integration that can have lifelong consequences

**TABLE 2** Systemic Effects of Myelomeningocele

| System | Effects |
|---|---|
| Neurologic | |
| Central | Hydrocephalus, Chiari II malformation, dysgenesis of the corpus callosum |
| Cranial nerves | Vision, strabismus, hearing, speech, stridor, swallow |
| Spinal | Tethered cord, progressive loss of motor and sensory function |
| Motor | Paraplegia, upper extremity hypotonia |
| Sensory | Loss of sensation and proprioception |
| Cognitive | Cognitive deficits including learning disabilities, executive function disorders, attention-deficit/hyperactivity disorder |
| | Lower cranium nerve dysfunction |
| Vision | |
| Acuity | Visual acuity problems, rarely severe |
| Alignment | Strabismus, oculomotor disorders |
| Oromotor | Oromotor dysfunction, swallowing disorder |
| Respiratory | |
| Central | Central ventilatory disorder, apnea, sleep apnea, hypoventilation |
| Pulmonary | Aspiration, restrictive and obstructive lung disease, pneumonia |
| | Stridor |
| Gastrointestinal | |
| Swallow | Swallowing dysfunction; oral, pharyngeal |
| Neurogenic bowel | Constipation, soiling, accidents |
| Urologic | |
| Neurogenic bladder | Bladder atonia/dystonia, increased bladder pressure, leakage/incontinence, reflux, renal failure |
| UTI | Bacterial colonization, acute/chronic UTIs |
| Genital | |
| Insensitivity | Lack of sexual sensation, erectile dysfunction, and retrograde ejaculation in males |
| Infertility | Early pubertal onset and increased risk of NTD in infants of women with NTD |
| Orthopedic | |
| Spinal | Scoliosis/kyphosis |
| Extremities | Decreased mobility, hip dislocation, clubbed feet, osteopenia, fracture, linear growth abnormalities |
| Growth | |
| Stature | Short stature resulting from limb-length disorders and precocious puberty in females |
| Weight | Overweight and obesity |
| Psychology | |
| Anxiety/depression | Increased anxiety and depression |
| Behavior | Social isolation, anxiety, depression, immaturity, risk-taking behavior, at increased risk for abuse |
| Social | |
| Inclusion | Social isolation, limited friendship, physical and transportation barriers |
| Employment | Limited work experience, physical/society barriers, loss of health coverage or benefits |

Myelomeningocele is the most serious and complex NTD and affects multiple body systems; the severity of these effects is related to the type and location of the NTD and the extent of neuronal injury.

(Table 2).[25] Thus, children with myelomeningocele are like children with spinal cord injuries with the addition of neurocognitive impairments.

The primary care physician plays a central role in long-term comprehensive care through collaborative comanagement with multiple medical and surgical specialists. It has long been recognized that the provider of primary care in the medical home must be knowledgeable about the unique medical issues of spina bifida and its developmental, educational, and social consequences.[26]

### Prevention

The mandated supplementation of cereal flours with folic acid in 1998 in the United States and Canada has been linked to a reduction in the prevalence of NTDs.[27] Also, all women of childbearing age should ingest 400 μg (0.4 mg) of folate daily,[28] which may decrease the occurrence of NTDs by up to 70%. Women who have had a previous pregnancy with any NTD and women with spina bifida should take 10 times that dose or 4000 μg (4 mg) daily from the period before conception through the first trimester. On the basis of current American Academy of Pediatrics guidelines, the primary care physician should recommend folic acid to all sexually active females, not just those with a history of an NTD. Folate supplementation has not been the sole factor in reducing the occurrence of NTDs. Better overall nutrition, improved maternal health, and termination of affected pregnancies are additional factors.

## DIAGNOSIS AND INITIAL COUNSELING OF THE FAMILY

Prenatal screening by measuring maternal serum AFP concentrations at 15 to 20 weeks of gestation has become the standard of care.[29] The AFP concentration is increased when fetal cerebrospinal fluid is released, as occurs with open myelomeningocele and anencephaly. Thus, a closed NTD might not be detected by measurement of the maternal serum AFP level. Elevated concentrations of AFP can be found in other fetal conditions, including twin pregnancies, ventral wall defects, fetal tumors, congenital nephrosis, and incorrect dating of the pregnancy. Although maternal serum AFP concentration is a useful screen, the results should be interpreted with caution. Maternal serum AFP concentrations more than 2.0 multiples of the median during weeks 15 to 20 after conception are considered abnormal and should be repeated. Abnormal concentrations are followed by high-resolution ultrasonography or other evaluations such as amniocentesis.

Ultrasonographic examination is used to screen for NTDs.[30] Two-dimensional ultrasonographic examination during the first trimester detects more than 90% of cases of anencephaly and more than 80% of encephaloceles but fewer than half of spina bifida cases. Second-trimester ultrasonography can increase the detection of spina bifida to 90% to 95%.[31]

Ultrasonographic examination can also help to monitor the fetus over time. It can be used to characterize the architecture of the spinal defect and the presence of and changes in cerebral ventricles over the course of the pregnancy and to identify other structural abnormalities. Fetal MRI provides additional information to better characterize the NTD, Chiari II defect, and status of ventricles and can detect previously unidentified abnormalities.[32]

Amniocentesis may be warranted if there are concerns that the NTD is related to a genetic or chromosome disorder or to confirm findings from ultrasonography or maternal serum tests. Measuring amniotic fluid AFP and amniotic fluid acetylcholinesterase can be useful for detection of an open NTD.[33] Chromosome analysis (karyotype) and microarray testing can be performed to determine if the NTD is related to chromosome aneuploidy, microdeletion, or another genetic condition.[34]

The process of fetal screening can be confusing and stressful for the expectant mother and her significant other. Counseling and support should be provided to the pregnant woman and her family about the screening procedures and their risks, results, and interpretation. This counseling should be provided by a medical geneticist, genetic counselor, and members of the fetal diagnosis team who are familiar with the procedures and are knowledgeable about the prognosis of NTDs. Counseling should include general information about spina bifida, obstetric care, choices about the pregnancy and delivery, neonatal care, surgery, the likelihood of hydrocephalus and its treatment, and the potential for disabilities and complications. The family should also be made aware of the services, care plan, and types of support that will be provided to them and the child. The option of fetal surgery to potentially reduce the extent of neurologic damage of an NTD and to reduce the need for ventricular shunting should be discussed. Families interested in pursuing fetal surgery should be referred to centers that provide fetal surgery for NTDs.[35]

Families may turn to the pediatric primary care provider for information and guidance when the primary care provider has a previous relationship with the family. In this case, it is essential that the primary care provider have ongoing communication with the prenatal diagnostic team to ensure that reliable and uniform information is provided to fam-

ily members as they look for answers and plan for options and decisions for the pregnancy and subsequent care.

## THE ROLE OF THE PRIMARY CARE PROVIDER IN ONGOING CARE IN THE MEDICAL HOME

The optimal care of the infant, child, or youth with spina bifida is best provided by a multispecialty team and a primary care provider who collaborate to provide comprehensive and coordinated care and support to the child and family.[36] The spina bifida team typically includes a clinical nurse specialist or nurse practitioner; pediatric specialists in neurosurgery, orthopedics, urology, developmental pediatrics, and physical medicine; physical therapists; orthotists; psychologists; social workers; and health education professionals. All these specialists might not be available to all teams and for all clinic settings. The care team's goal should be to meet the individual needs of each child and family by providing comprehensive and coordinated care, support, and education to the patients and families and support and assistance to the primary care provider, the child's school or early-intervention program, and other service providers.[37]

The primary care provider's interventions may begin even before birth and continue through transfer to adult health care. Families that have a preexisting relationship with a primary care provider may turn to him or her for guidance and information at the time of prenatal diagnosis. In other cases, the family may select a primary care provider before or at the time of delivery. The primary care provider's confidence in caring for an infant with an NTD might vary depending on knowledge base and previous experience. The spina bifida team can be a valuable resource for the primary care provider to ensure optimal care. An in-depth understanding of the child's needs and those of the parents will develop through contacts during scheduled well-child visits and visits for acute illnesses or other concerns.

The primary care provider usually has *first contact* with the family, whether for routine or acute care. The care of the individual with spina bifida requires the primary care provider to be able to recognize and treat issues unrelated to the spina bifida (such as gastrointestinal or respiratory tract infections) while at the same time identifying and rapidly referring for conditions (eg, headaches or new-onset weakness) that might indicate serious problems (such as ventricular shunt malfunction). This ability requires knowledge about spina bifida and optimal communication. Specialists should routinely communicate with primary care providers by using a combination of technologies such as e-mail, telephone, and fax. Primary care providers should communicate acute symptoms or concerning changes in their patients (eg, headaches or urinary symptoms) to the specialty team.

### Newborn Period

For the fetus identified with an NTD, ongoing prenatal planning and care are important, as is the timing, location, and method of delivery (ie, in a tertiary medical center). However, despite the availability of prenatal diagnosis, infants continue to present at delivery with previously unidentified open and closed NTDs. The obstetric and infant care teams should be prepared to address anticipated or unexpected birth of an infant with an open or closed NTD. At birth, a physician, using sterile nonlatex gloves, should assess the open lesion to document its location and size and to determine if it is leaking cerebrospinal fluid.[38] The lesion should be covered and protected using a sterile technique.[22] The dressing should not be removed or disturbed except by the neurosurgical team. The infant should be placed in a prone or lateral position to avoid pressure on the lesion. After stabilization in the delivery room, the infant should be shown to the parent(s) before being transported to the nursery. Admission to the NICU or transportation from an outlying hospital to a NICU should be promptly arranged. Initial neonatal examination of the infant with an open or suspected closed NTD should be performed shortly after birth and should include all the usual elements of a typical newborn physical examination but with particular attention for spinal irregularities, hemangiomata, hair tuft or abnormal pigmentation, dimples, or pits. Assessment of head circumference and tenseness of the anterior fontanel for signs of hydrocephalus, movement of the lower extremities, level of sensation, and deep tendon reflexes and anal wink should be noted. Abnormalities of the lower extremities and flexion or extension contractures of hips, knees, and ankles should be noted. Other physical or neurologic or congenital abnormalities, including structural anomalies of the heart, airway, gastrointestinal tract, ribs, and kidneys, should be assessed. Ventricular size should be evaluated soon after birth by ultrasonography, computed tomography (CT), or MRI; ultrasonography and MRI are better than CT scanning at identifying a Chiari II malformation.

Neurosurgical consultation should be obtained shortly after birth, and arrangements for surgical closure should be made within 72 hours after birth; this step further decreases the risk of central nervous system infection.[23] After repair of the myelomeningocele, head circumference should be measured regularly and carefully to

monitor for increase at a rate greater than the normal curve. On the basis of clinical evidence, the newborn infant should be monitored for the next few weeks to determine if a ventricular shunt is indicated. Serial neuroimaging using ultrasonography can identify the progression of hydrocephalus. The patient is reassessed every 3 to 10 days depending on the level of concern. Progressive hydrocephalus may cause the infant to develop irritability, vomiting, stridor, or poor feeding; 85% or more of infants with myelomeningocele will require a ventricular shunt.[39]

The newborn infant should be evaluated by a urologist before discharge. A baseline sonogram of the kidneys and bladder is typically performed to determine if the infant already has renal involvement or congenital abnormalities. Measurement of serum urea nitrogen (SUN) and creatinine concentrations, a voiding cystourethrogram, and direct measurement of the pressure within the bladder may be indicated as well.

Discharge planning should begin soon after birth and should ensure that all primary and specialty care is arranged before discharge. Communication is open among the specialists, spina bifida team, and support programs, including the early-intervention program, the primary care provider, and the family. A written discharge plan that focuses on the infant's individual and unique needs related to the NTD should be provided to all clinical and support programs and the family. Early follow-up with the primary care provider and the spina bifida team should be arranged.

Primary care of the infant and child with spina bifida within the medical home should ensure that all routine care, services, intervention, and immunizations are provided in line with American Academy of Pediatrics recommendations in *Bright Futures*, with addition of the specific recommendations for patients with spina bifida provided by the Spina Bifida Association of America and others (see "Resources").

## Hydrocephalus

Because hydrocephalus develops in most children with open spina bifida (myelomeningocele) and in some children with closed lesions, head growth should be carefully monitored in all infants with spina bifida. Changes in head circumference percentile, the tension of the anterior fontanel, changes in mental status, vomiting, and changes in extraocular movement, such as strabismus or the sunset sign should be discussed with the neurosurgical team. Later, when the cranial sutures and fontanels close, the functional status of the ventriculoperitoneal shunt is determined by signs such as headache, irritability, changes in mental abilities, and vomiting.[40] Shunt failure can occur at any time—days, weeks, or even years after insertion. The primary care provider needs to become familiar with the signs and symptoms of shunt failure and should be in communication with the neurosurgical team.

## Development

Most people with myelomeningocele have intelligence within the normal range, but most experience significant learning disabilities. These disabilities include nonverbal learning disorder, poor executive skills, attention deficits, and memory problems.[41,42] These cognitive difficulties might delay the child's maturation and impede his or her ability to acquire the skills needed to live independently, which in turn affects family members.[43] Recognition of these issues and interventions to optimize learning and independence are critical for optimizing learning and social participation.

Cognitive development is affected by several factors, including shunt infection, Chiari II malformation, hydrocephalus with repeat shunt replacements, and neuronal migration disorders (eg, hypoplasia of the corpus callosum). These factors distinguish myelomeningocele from other disorders that are limited to the spinal cord, such as lipomeningocele and traumatic paraplegia. Developmental and learning disorders may become evident at any time from infancy through adolescence; therefore, developmental surveillance should be part of all routine well-child visits. Particular attention should be paid to language and communication, and formal testing should be ordered if concerns develop. Because there is an increased risk of vision and hearing problems in children with spina bifida and hydrocephalus, testing and evaluation should be performed on a regular basis.

The primary care provider should remain in close contact with the child's early-intervention and school program to ensure that information is shared. The child's developmental status, with strengths and weaknesses noted, should be formally evaluated to ensure that appropriate needs are recognized and addressed. The primary care provider might need to provide information to the school program and, at times, advocate for the child and family for specific educational services. An emergency plan for the school should be developed and reviewed annually; it may contain information on medical conditions (eg, if the child develops signs of increased intracranial pressure) as well as a plan for evacuation from the school for the child with limited mobility.

## Bowel Function

Almost all people with myelomeningocele have disorders of innervation of the lower intestinal tract and anus

with lower tract dysmotility and poor or absent sphincter control, which lead to fecal incontinence. Fecal incontinence can seriously impair social relations, limit independence, and lower self-esteem.

The management of incontinence is time-consuming; yet, initiating a bowel-management program early in development helps both the family and the child to develop a routine, effective program of management and self-care. The goals of a bowel-management program are to achieve continence and promote regular elimination of stool, which can be achieved with an individualized program that may use timed toileting, changes in diet, and oral laxatives, suppositories, and enemas, singly or in combination. At the initiation of a functional bowel program, constipation and fecal impaction may necessitate a bowel clean-out.

As children become older and capable of self-management, they might benefit from a surgical procedure called antegrade continence enema (ACE).[44] In the Malone antegrade continence enema (MACE) procedure, the appendix and cecum are used to create a catheterizable stoma.[45] In the ACE procedure, the cecum is brought up to the abdominal wall and an ostomy tube is inserted, which saves the appendix for urologic procedures. With either procedure, the patient is able to clean out the colon by irrigating it through the ostomy tube. The irrigation is performed daily or every other day; it may take as long as 2 hours to complete. The ACE/MACE can achieve fecal continence in ~85% of patients.

## Urinary Tract Function

Almost all people with myelomeningocele have a neurogenic bladder. They may not be able to store urine in the bladder or may be unable to empty it. Failure to completely empty the bladder and elevated pressure within the bladder increase the risk of urinary tract infection (UTI) and renal tract injury resulting from urinary reflux and can lead to progressive damage to the upper urinary tract and kidneys, with the risk of progressive renal damage, which still occurs in youth and adults with spina bifida. Early evaluation of urinary tract structure and function by ultrasonography, radiographic imaging, and urodynamic testing beginning in infancy can help determine the status of the kidneys and bladder. Urologic treatment is aimed at normalizing pressure within the bladder, providing continence, and minimizing infection and reflux. Close attention to urinary tract function is important, because changes may indicate UTI or tethered cord, including tethering attributable to shunt malfunction. Some children or youth benefit from bladder augmentation or urinary diversion procedures to increase bladder capacity, improve urinary continence, and reduce the risk of progressive renal injury.

Management of the urinary tract typically begins with the early introduction of clean intermittent catheterization (CIC) beginning even in infancy and close monitoring for changes in bladder function. If CIC is unable to optimize pressures within the bladder, a vesicostomy (a hole placed in the abdominal wall into the bladder) may be indicated. Many patients are also maintained on medication to reduce vesicoureteral reflux. These and other interventions improve bladder pressures and urinary continence, which permits the child with myelomeningocele to function better in school and other social settings. The child/teenager is monitored over time for the occurrence of UTIs; some people with repeated infections may benefit from chronic antibiotic prophylaxis to reduce the risk of reinfection, although this is practiced less often now than in the past. Differentiating UTI that requires antibiotic treatment from bacteruria, which is often present but does not need to be treated, may be difficult and require consultation with a urologist.

The primary care provider should be aware of and monitor the child's adherence to the catheterization program while maintaining contact with the urology team regarding medications and interventions. Communication from the primary care provider to the specialist regarding UTIs and other urinary problems is critical. In the case of fever, nonspecific symptoms, or changes in urinary tract function, the primary care provider should maintain a high index of suspicion for UTI. The child should have urinalysis and urine culture obtained if there is a likelihood of bladder or kidney infection. Patterns of bacterial resistance to antibiotics should be carefully monitored. Oral and, occasionally, intravenous antibiotics will be required to treat UTIs in these children. Open and active communication between the primary care provider and the pediatric urology team is essential for maintaining optimal renal function and establishing urinary continence. Blood pressure should be monitored to identify hypertension.

## Mobility

Mobility should be addressed from infancy through adulthood. Creeping or crawling gives an infant experiences with control and competence. Mobility aids for toddlers, whether wheeled or upright, continue this process. Being safely mobile in their homes and communities will help children and teenagers become more independent. Walking or using a wheelchair not only promotes confidence and independence but also helps with overall fitness and weight control. In prescrib-

ing wheelchairs or other assistive devices, the focus should be on optimizing age-appropriate functionality and independence for the child or teenager. Many teenagers with spina bifida can safely drive a car but need modifications with hand controls. Driving rehabilitation specialists can be helpful in identifying potential challenges to safe driving, providing appropriate driver training, and suggesting changes needed to the car to make it accessible.

### Orthopedic Problems

Orthopedic problems occur in most children with myelomeningocele and tend to be more common and more complex with higher-level lesions; they affect activity more with increasing age and in obese patients.[46] Clubfeet, hip subluxation and dislocation, and knee instability or contractures may be present at birth, especially in patients with lumbar and thoracic lesions. Unbalanced muscle function leads to worsening of existing joint problems or to the onset of new orthopedic disorders. Correcting deformities, maintaining posture, and promoting ambulation to maximize function and independence are generally handled by orthopedists and physical therapists. There is also a need to diagnose and treat fractures, which are more common in children with myelomeningocele, who often have osteopenia. Fractures in insensate limbs may present with redness and swelling without pain and should be suspected by the primary care provider.

Scoliosis occurs in most children with lesions above the second lumbar vertebra (L2). Examination of the back and monitoring of the progression of scoliosis should be performed regularly by the spina bifida team and primary care provider, even in preschool-aged children. Progression of scoliosis should prompt a thorough neurologic evaluation and imaging to identify correctable causes such as tethered spinal cord, hydromyelia, or shunt malfunction and to maintain function. Motor and sensory function and bowel or bladder patterns should be part of the evaluation. The pediatric care provider should continue to monitor lower extremity function and identify any deterioration.

### Skin Care

The loss of normal skin sensation and autonomic response to pressure or irritation places people with an NTD or spinal cord injury at risk of breakdown of skin as well as the formation of decubitus ulcerations. Injury to insensate skin can occur at any site of pressure or irritation, including the pelvic area from prolonged sitting, casting, or braces or even tight clothing; abrasion from seating or positioning equipment; or thermal injuries from any of a variety heat sources. Injury to insensate skin can be permanent damage that requires long-term, complicated, and costly treatment and causes additional disabilities.[47] Careful monitoring of skin integrity and for potential sources of injury should begin in infancy and continue along the life span. All sign of injury or breakdown of insensate skin should be treated promptly and vigorously to minimize the risk of complications.[48]

### Adolescence

Teenagers face many physical, mental, emotional, and social changes. They must develop their own identities and interests and strive for greater independence. This transition period can be challenging, especially for teenagers with spina bifida. It is important for the parents, teenager, educational system, and health care providers to take an active role in encouraging growth, self-confidence, independence, and competency even before adolescence.

Fitness and well-being should be encouraged from an early age. Physical activity is especially important for those with spina bifida. Physical activity improves general health, reduces obesity, and improves confidence and self-image. Children and teenagers with spina bifida should be encouraged to engage in physical activities with friends and to participate in adapted sports. Attending summer camp or using accessible recreational facilities can also help with well-being and the development of self-care skills such as dressing, bathing, toileting, and mobility. The teenager should be educated to inspect areas of insensate skin while bathing and dressing and to report any signs of irritation or ulceration to prevent decubitus ulceration or other serious injury to the skin. By midadolescence, teenagers should make their own doctor appointments and participate in their individualized education plan (IEP) or 504 plan, if they have one. They should be encouraged to advocate for themselves in school and report problems such as teasing or bullying.[49,50]

The primary care provider should include children and youth in the discussion of their health and related issues from the early school years. Even before adolescence, children should be encouraged to participate actively in health care visits and report their concerns and accomplishments. By early adolescence, some time during each visit should be set aside for private discussions with the child. This time should be increased until the teenager has essentially private office visits with the doctor. Beginning in early adolescence, transition-related issues, such as education about spina bifida, treatments and medications, general health issues, and transition and transfer to adult health care, should be discussed. For some youth with significant cognitive impairment, the goal

of independent decision-making might not be achievable, and planning for personal and financial guardianship might be necessary. These and other topics can be introduced during health maintenance, preventive care, and acute care visits.

The primary care provider should develop an understanding of the mental health risks associated with spina bifida, including social isolation, learned helplessness, and low self-esteem. Youth with spina bifida are at increased risk of both anxiety and depression.[51] The primary care provider should evaluate these conditions and identify the patient's risk of suicide. Interventions such as increasing physical activity and teaching relaxation techniques may be beneficial. Counseling and support should be given to the teenager and family members by the primary care provider. Counseling through school- or community-based mental health professionals may be indicated before anxiety or depression worsens. Whenever these conditions persist or interfere with school or social activities, the adolescent should be referred to a mental health professional who is familiar with the mental health challenges of teenagers with disabilities.

## Sexuality

Issues of sexuality in teenagers and young adults with spina bifida are often overlooked or actively avoided. For patients with spina bifida, a number of significant issues related to sexual maturation require attention. Precocious puberty is common in girls with myelomeningocele and hydrocephalus; girls with hydrocephalus have menarche at an average age of 10.9 to 11.4 years rather than the more typical 12.7 years for the average American girl. Precocious puberty affects the physical, physiologic, and emotional outcomes of sexual maturation, including linear growth.[52] The progression of puberty can be delayed by using leuprolide (Lupron), which provides additional time for emotional maturation and improved linear growth; this treatment is usually best managed by a pediatric endocrinologist. Topics of discussion that are important for girls with spina bifida include fertility, pregnancy, and contraceptive choices. Folate supplementation needs to be stressed. Female teenagers who are lonely or naive or have learning disabilities are at increased risk of being taken advantage of sexually or even subject to sexual assault.

For male teenagers, altered sensation, erectile dysfunction, and incontinence affect sexual behaviors, including intercourse.[53] The issues of sexual performance may require referral to a professional who is knowledgeable in male sexual disorders. Treatment with sildenafil (Viagra) or other medications may be beneficial. Sexuality is affected by self-esteem and self-image and might be an issue not addressed by the spina bifida team; the primary care provider might be the only one to address these issues and should initiate treatment or refer to specialty services when indicated.[54]

## Transition to Adult Health Care

Transitions for adolescents and young adults with spina bifida should include the orderly transfer to accessible adult medical care as well as transitions in social, educational, vocational, and recreational areas.[55] As part of health care transition, all young people need to develop an understanding of their health care needs and begin to take responsibility for their health care decisions, as their cognitive and learning skills allow. Transition planning should focus on building competencies in self-care, fostering independence and self-advocacy, and assisting with supports and strategies for transfer to adult care providers.[56,57]

The medical home should be part of the transition process, which should include exploring educational and vocational options, obtaining skills for work, finding safe and dependable transportation, deciding on living arrangements, developing healthy social and personal relationships, obtaining medical insurance, and identifying both primary and specialty care providers who are knowledgeable in the care of adults with spina bifida. Transition planning and discussions within the medical home beginning early in adolescence can help to make these changes easier and outcomes more successful. Web sites such as the New York State Institute for Health Transition Training for Youth With Developmental Disabilities Ages 14–25 Years (http://healthytransitionsny.org), the Adolescent Health Transition Project (http://depts.washington.edu/healthtr), the Health Care Transition Initiative (http://hctransitions.ichp.ufl.edu), and the National Health Care Transition Center's Got Transition? page (http://gottransition.org) can provide guidance for families and the primary care provider.

## Family Support

Spina bifida has a profound effect on the family, beginning with the prenatal diagnosis; at birth, the families have to face issues such as deformities and complex surgical interventions for closure of the spinal lesion and shunt placement for hydrocephalus. Later, the extent of physical and developmental disabilities becomes clearer. Families must deal with the increased demands for care, including procedures such as catheterization and range-of-motion exercises, more frequent medical appointments, and other interventions, all of which disrupt the family's

**TABLE 3** Care of the Child and Youth With Spina Bifida: Potential Roles of the Pediatric Primary Care Provider

| Time | Concerns | Actions |
|---|---|---|
| Fetuses (prenatal) | Diagnosis | Consult with diagnostic team |
| | Lesion: type and location | Review obstetric plan |
| | Other central nervous system findings (hydrocephalus, Chiari II malformation) | Review findings and implications with family |
| | Other physical findings (orthopedic, other) | Discuss options and plan |
| | Maternal and family stress and anxiety | Referral to spina bifida team |
| | Family choices and plan | Update primary care provider and other provider knowledge base |
| | | Family education and support |
| | | Communicate among family and all professionals |
| Newborns | Stabilization | Consult obstetric and neonatal teams, neurosurgery, others specialists |
| | Protection of lesion | Family support |
| | Examination | Monitor head growth |
| | Surgical closure | Neonatal screening and newborn hearing assessment |
| | Hydrocephalus | Discharge planning |
| | Motor function | Primary care and specialty follow-up |
| | Bowel and bladder function | |
| | Orthopedic conditions | |
| | Maternal/family anxiety and depression | |
| Infants | General health status | Health care per *Bright Futures* recommendations |
| | Growth | Referral to spina bifida team and support group |
| | Development | Referral to early-intervention program |
| | Immunizations | Monitor head growth and shunt function |
| | Spina bifida-specific concerns: hydrocephalus and shunt function, bowel and bladder function, UTI | Assess for feeding or swallow problems |
| | | Discuss latex precautions |
| | | Supplemental Security Income, Medicaid, and Medicaid Waiver |
| Preschool-aged children | General health status | Health care per *Bright Futures* recommendations |
| | Growth and development | Follow-up with spina bifida team and support group |
| | Spina bifida-specific concerns: vision and strabismus, motor function and mobility, hydrocephalus and shunt function, bowel and bladder function, UTI | Obtain early-intervention assessment, individualized family service plan |
| | | Developmental team evaluation |
| | | Discuss social inclusion and activities |
| | | Bracing and ambulation |
| | | Activity and weight management |
| | | Dental referral |
| | | Toileting |
| School-aged children | General health status | Health care per *Bright Futures* recommendations |
| | Growth and development | Follow-up with spina bifida team |
| | Weight control and healthy diet | School transition, service and support, Section 504/individualized education plan |
| | Spina bifida-specific concerns: motor function and mobility, hydrocephalus and shunt function, bowel and bladder function, tethered cord | Screen for attention-deficit/hyperactivity disorder, learning disabilities, and executive function disorders |
| | | Bullying and safety |
| | | Self-care and independence |
| | | Physical activities and social group participation |
| | | Neurology and physical therapy follow-up |
| Teens | General health status | Adolescent health care per American Academy of Pediatrics recommendations |
| | Weight control and healthy diet | Confidentiality and private visits |
| | Sexuality | Birth control and folic acid |
| | Psychosocial stressors | Health care transition |
| | Spina bifida specific concerns: motor function and mobility, tethered cord | Self-care and independence |
| | Developing independence | Driving and transportation |
| | | Spina bifida teen support group |
| | | Educational and vocational planning |
| Adults | General health status | Identify health care resources |
| | Weight control and healthy diet | Health insurance, Social Security Disability Income |
| | Spina bifida specific concerns: motor function and mobility, bowel and bladder management, skin care and pressure sores | Education and employment |
| | | Living situation |
| | | Spina bifida adult support group |
| | | Transfer to accessible adult health care |

Concerns and actions are not intended to be limited to a single time period but should be addressed and readdressed as part of ongoing health care across the life span.

hopes and plans for both the child and themselves as a family unit.

The ongoing care of the child with spina bifida strains each family member. It alters roles and expectations and decreases opportunities (eg, the parent who cannot go back to work or school). It adds anxiety and stress and even a sense of guilt, which can become the source of depression and friction; it increases the risk of family dysfunction and instability.[57] Adolescence often increases family stress.[58]

Spina bifida affects not only the patient but also his or her siblings, who are subject to stress and feelings of loss, guilt, frustration, and anger. These feelings have consequences for the sibling, for his or her relationship with the child with spina bifida, and for the entire family.[59,60] The psychosocial and emotional consequences of spina bifida on the patient, siblings, and family should be part of the overall care of the child with spina bifida. These factors should be addressed by both the spina bifida team and the primary care provider in the medical home. A family-centered approach to care and anticipation of potential problems will help to identify and address stressors early.

## SUMMARY

The standard of care for children and youth with spina bifida remains a program of comprehensive and multidisciplinary support, and the medical home plays a central role. The pediatric primary care provider has frequent contact with the child, including well-child and acute health care visits, and by partnering with the child, parents, and specialty program, builds trust and can help address the medical, developmental, and psychosocial issues of childhood over time. The primary care provider also helps to manage ongoing care of the child's multisystem disorders and the unique nonmedical

**TABLE 4** Primary Care Interventions Typically Provided to Children and Teenagers With Spina Bifida

| |
|---|
| Fetuses (prenatal) |
| Counsel families in planning and decision-making |
| Provide information on spina bifida |
| Options and assistance with family choices |
| Consult with obstetric, neonatal, and neurosurgical teams |
| Prenatal (fetal)/postnatal surgery |
| Discuss postnatal planning and treatment |
| Family support |
| Newborns |
| Postnatal care and stabilization |
| Surgical repair of spinal lesion |
| Monitoring and surgery for hydrocephalus |
| Family support |
| Referral to multidisciplinary spina bifida term |
| Primary and specialty follow-up |
| Infants |
| Provide early and frequent follow-up |
| Monitor hydrocephalus |
| Provide routine and diagnostic-specific primary care |
| Give family and sibling teaching and support |
| Discuss recurrence risk and prevention |
| Refer to early-intervention program |
| Communicate and coordinate with the spina bifida team |
| Toddlers |
| Preventive and well-child care |
| Developmental monitoring |
| Mobility |
| Growth and weight management |
| Preschool-aged children |
| Transition from early-intervention program to preschool program |
| Ambulation and mobility |
| Bowel and bladder program |
| Social inclusion |
| School-aged children |
| Identify and characterize learning abilities |
| Ensure appropriate school-based services, Individualized Educational Plan or 504 plan |
| Monitor secondary conditions including latex allergy |
| Encourage physical activities, friends, and household responsibilities |
| Plan for educational transition |
| Encourage independent self-care and toileting |
| Preteens |
| Begin health care transition planning |
| Advocate for physical activities, recreation and community inclusion, |
| Monitor progress in school |
| Address bullying and safety |
| Monitor growth and puberty |
| Encourage independence and self-advocacy |
| Develop and maintain friendships |
| Teens |
| Continue transition and transfer process to adult care, activities and social participation |
| Educate regarding spina bifida and self-care |
| Provide private health care visits |
| Provide anticipatory guidance regarding sexuality and reproduction |
| Encourage independence in health care decision-making |
| Monitor growth and vital signs (blood pressure) |
| Encourage physical activities |
| Manage weight and nutrition |
| Encourage cardiopulmonary health and fitness |
| Young adults |
| Transfer care to a provider of routine adult health care |
| Provide resources for specialty care: neurosurgery, orthopedics, urology, and others |
| Monitor weight and physical fitness |
| Provide information regarding finances such as Social Security Disability Income and health insurance |
| Monitor education and employment |
| Help to build and maintain friendships and social support groups |
| Monitor for deterioration and late-onset complications |

issues related to spina bifida and is a first-line resource for patients and families in identifying and addressing strengths, problems, needs, and services.[37] The goals of primary care for children and youth with spina bifida should be to promote optimal health and well-being, to prevent secondary conditions and disabilities, to build on individual strengths and abilities, to help in the development of independence, and to promote social competence and inclusion—in short, to help patients become capable and contributing adults despite the challenges of their chronic health conditions.

The "Guidelines for Spina Bifida Health Care Services Throughout the Lifespan" and "Health Guide for Parents of Children Living With Spina Bifida" and other publications of the Spina Bifida Association (see www.sbaa.org) are useful resources for medical home providers. These and other resources can help in developing a care plan covering from birth through transition and transfer to adult health care, with focus on specific needs at each age and stage. Tables 3 and Table 4 outline some of these activities.


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American Academy of Pediatrics, Medical Home Initiatives for Children With Special Needs Project Advisory Committee. The medical home. *Pediatrics*. 2002;110(1 pt 1):184–186. Reaffirmed May 2008

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