Children with Spina Bifida: Key Clinical Issues

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KEYWORDS

• Spina bifida • Myelomeningocele • Hydrocephalus • Treatment

Spina bifida is the most common of the neural tube defects (NTDs), which include myelomeningocele, encephalocele, and anencephaly. Spina bifida occulta is a common spectrum condition, present in approximately 5% of the population, in which one or more vertebral arches may be incomplete (**Fig. 1**). The spinal cord is normal, and there are usually no associated neurologic abnormalities. Spina bifida occulta may be accompanied by localized skin abnormalities (dermal sinus, dimples, and pigmented or hairy skin). If there is an associated skin-covered swelling, with an intact spinal cord, the terms meningocele or lipomeningocele are used. Meningoceles are almost always present in the lumbosacral area. A lipomeningocele may be entirely asymptomatic, but intraspinal lipomas can impinge on the cord and lead to progressive weakness and/or deformity.

When spina bifida is open and associated with a malformed spinal cord and a sac, the terms myelomeningocele or meningomyelocele are used. Myelomeningoceles arise as a consequence of incomplete or disrupted neurulation during the fourth week of gestation, when the embryo is only 3 to 5 mm in length (**Fig. 2**).[1] The myelomeningocele includes the splayed-open malformed cord (neural placode) as well as the meninges and fatty tissue (**Fig. 3**). Myelomeningoceles are often intact, with a meningeal sac enclosing cerebrospinal fluid (CSF), but in many cases the sac is disrupted and leaking CSF at birth. Myelomeningoceles vary in size and location, and the range of associated motor and sensory impairments depends on the level of the lesion.

SENSORY AND MOTOR IMPAIRMENTS

Spinal level is determined by careful examination of sensation and motor function, and is generally classified as thoracic, high lumbar (L1 or L2), midlumbar (L3), low lumbar

NORMAL ANATOMY   **SPINA BIFIDA OCCULTA**

Skin

Neural arch

Spinal cord

Vertebral
body

Intact skin   Swelling, birthmark,
Meninges   hairy patch
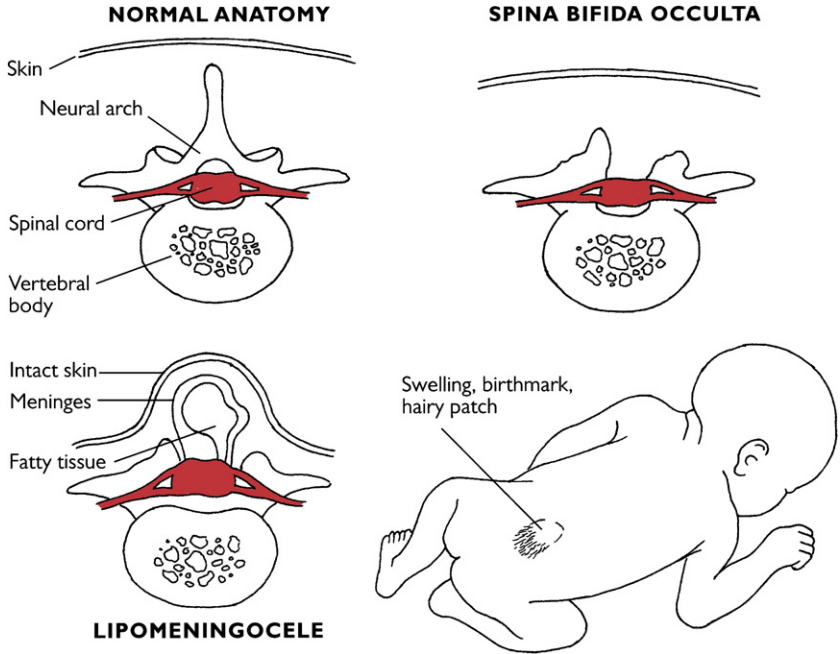
Fatty tissue

LIPOMENINGOCELE

Fig. 1. The spectrum of spina bifida occulta. Abnormalities of this kind are common and usually asymptomatic. (*From* Sandler A. Living with spina bifida: a guide for families and professionals Chapel Hill (NC): University of North Carolina Press; 1997. p. 15; with permission.)

First trimester

Weeks 1 2 3 4

Head   an closed

Anterior   an
neuropore
(an)

Spinal
column

Posterior
neuropore   pn
(pn)   pn

View from above,   Side view,
21 days   28 days

Fig. 2. Neurulation occurs during the fourth week of gestation. The process probably involves several genes important in folate-dependent biosynthesis. (*From* Sandler A. Living with spina bifida: a guide for families and professionals Chapel Hill (NC): University of North Carolina Press; 1997. p. 13; with permission.)
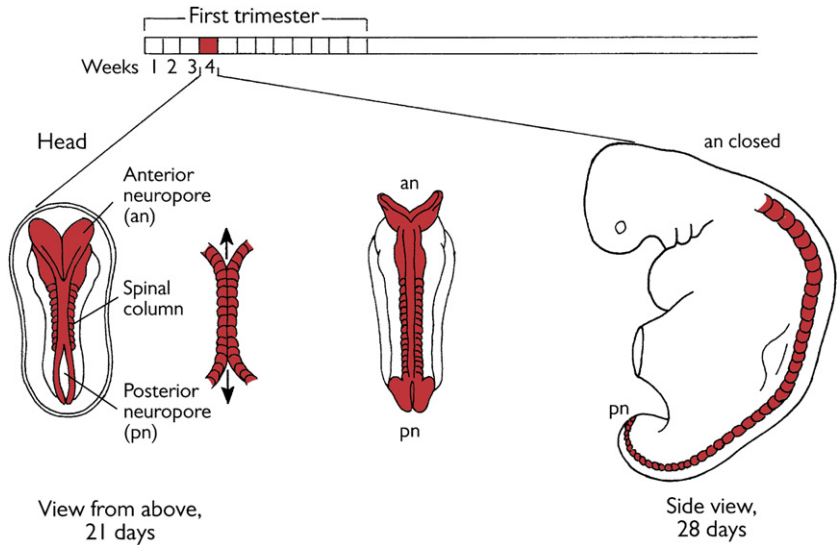
Fig. 3. Myelomeningoceles and open spina bifida. Neural elements, including malformed spinal cord, meninges, and fatty tissue are exposed. (*From* Sandler A. Living with spina bifida: a guide for families and professionals Chapel Hill (NC): University of North Carolina Press; 1997. p. 16; with permission.)
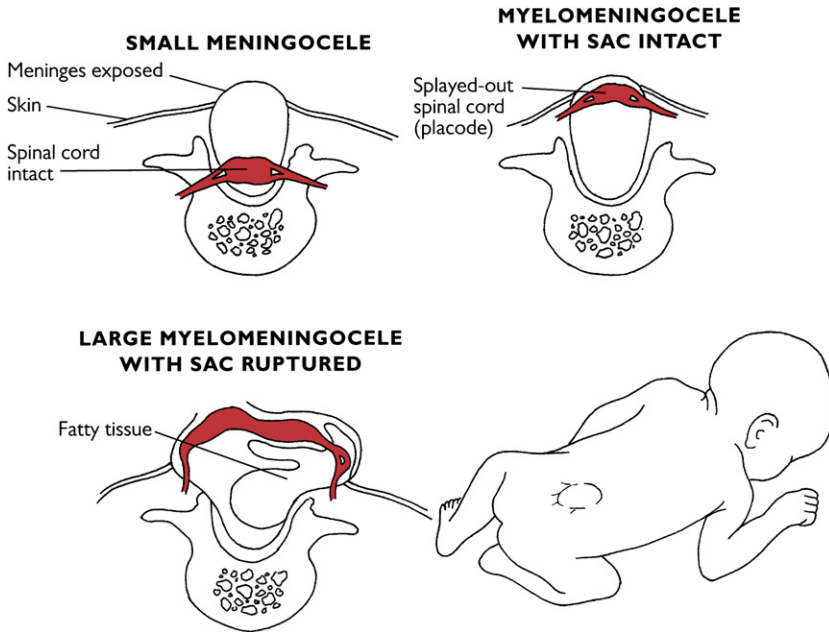
(L4 or L5), or sacral. Asymmetry of sensory loss or weakness is common. Most children with low lumbar (L5) or sacral myelomeningocele have absent sensation around the anus, perineum, and feet, but some individuals with lower sacral lesions may have no detectable sensory loss. Children with L1 or L2 lesions may have some hip flexion and adduction but no quadriceps strength to extend the knees. Those with L3 lesions may have knee flexion but paralysis of ankles and feet. Children with L4 and L5 lesions have quadriceps strength (for knee extension) and may have some hamstring (for knee flexion) and anterior tibialis strength (for ankle dorsiflexion). Those with S1 lesions may have functioning glutei (involved in hip extension) and gastrocnemii (involved in ankle plantar flexion). Assessment of motor function is important in predicting mobility and the need for bracing and also serves as useful baseline information in determining whether neurologic deterioration from tethering is occurring.

Functional mobility outcomes for different levels of spina bifida and the need for bracing have been reviewed extensively.[2,3] Children with sacral lesions usually walk by the age of 2 to 3 years and may require bracing at the ankles. Those with L3 paralysis usually require forearm crutches and bracing above the knees. Children with high lumbar or thoracic lesions may eventually stand upright and walk with extensive support of the hips, knees, and ankles. Most children with midlumbar spina bifida, who are able to ambulate with crutches and braces, rely increasingly on wheelchairs for mobility as they get older.

In addition to the disruption of motor and sensory nerves, myelomeningocele affects the sacral parasympathetic nerves that supply the muscular walls of the bladder, urethra, and rectum, and are critically important in sexual functions. Sympathetic nerves controlling the bladder outlet, which originate in the lumbar region of spinal

cord, are also typically involved. Bladder and bowel dysfunctions are present in almost all children with myelomeningocele along with varying degrees of sexual dysfunction.

HYDROCEPHALUS AND CHIARI MALFORMATION

Most babies with myelomeningocele have a complex brain malformation, Chiari type II, with associated hydrocephalus.[4–6] Among patients with Chiari II malformations, 80% of those with sacral lesions and more than 90% of those with higher-level lesions receive a shunt. The Chiari II malformation consists of downward displacement of the cerebellum, elongation and upward displacement of the medulla and fourth ventricle, dysgenesis of the corpus callosum, a small posterior fossa, and associated hydrocephalus (Fig. 4). This complex anomaly arises in the fifth week of gestation as a consequence of abnormal neurulation. The Chiari malformation is commonly asymptomatic, but may present with a spectrum of symptoms and signs related to brainstem compression and lower cranial nerve dysfunction. About 30% of infants with myelomeningocele have mild symptoms, including feeding difficulties and gastroesophageal reflux, whereas 5% have more severe symptoms, including stridor, weak cry, failure to thrive, apnea, and cyanosis ("Chiari crisis").

Abnormal CSF dynamics leads to hydrocephalus, which may be present prenatally (the so-called lemon sign on fetal ultrasonography). Neonatal signs include large or rapidly enlarging head circumference, bulging anterior fontanel, and split sagittal suture. Ventriculoperitoneal (V-P) shunts are placed in newborns with myelomeningocele and hydrocephalus, allowing control of CSF pressure and ventricular volumes and prevention of progressive hydrocephalus.
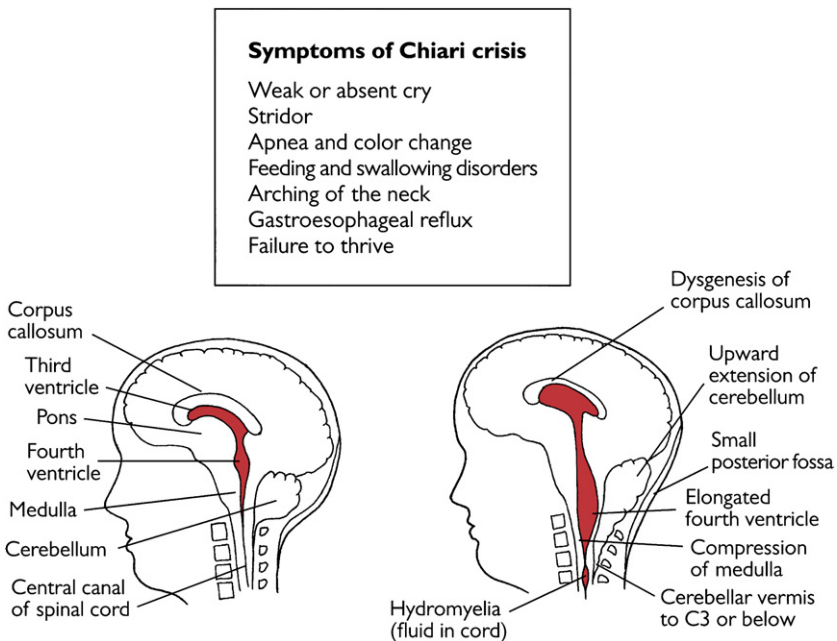

Fig. 4. Features of the Chiari II malformation, compared with normal anatomy (*left panel*). Symptoms of Chiari crisis may occur in infancy because of brainstem compression. (*From Sandler A. Living with spina bifida: a guide for families and professionals Chapel Hill (NC): University of North Carolina Press; 1997. p. 68; with permission.*)

ASSOCIATED NEUROLOGIC DISORDERS

Approximately 15% to 20% of children with spina bifida have seizures in childhood.[7] Seizures are more likely in children with shunts, especially in those with previous shunt infections, and the onset of a seizure may indicate a shunt malfunction. The seizures are usually generalized tonic-clonic type and respond well to antiepileptic medications.

Oculomotor disorders, such as difficulty with visual tracking, are common in spina bifida and may be related to the effects of Chiari malformation and hydrocephalus on midbrain gaze centers. Strabismus occurs in 20% of the patients and may require surgery. Even in the absence of hydrocephalus and Chiari malformation, fine motor function may be impaired, probably because of cerebellar and cervical cord abnormalities.[8]

Parents and clinicians need to remain vigilant for signs of neurologic deterioration in children with spina bifida. The signs of shunt failure (rapidly enlarging head circumference, swelling or redness along the shunt track) are usually clear in the infant and toddler. In young children, shunt failure may present acutely with headache, irritability, lethargy, and vomiting, but signs of shunt malfunction may be subtle and insidious, including mild drowsiness and impaired attention and coordination. Chiari II malformation or cervical hydromyelia may present with neck pain, progressive spasticity, or ataxia.

Another important cause of neurologic deterioration is tethered cord.[9] During normal growth, the spinal cord ascends within the canal so that the conus moves from L4 to L2 between birth and puberty. In spina bifida, the abnormal cord may be tethered to the scar tissue or bony deformities, leading to ischemic damage. Associated spinal cord anomalies such as hydromyelia and cord lipomas may also cause neurologic problems. Clinical signs are most common around the age of 6 to 12 years, including deterioration of walking, back pain, leg pain, spasticity, increasing scoliosis, progressive foot deformity, and deterioration in bladder and bowel function. Progressive weakness over time on manual muscle testing and changes in bladder function are key clinical findings. The back pain is typically worsened with activity and relieved by rest. Tethering is generally diagnosed on clinical grounds, although magnetic resonance imaging of the spine, urodynamics, and electrophysiologic testing may provide additional data. Surgical release of the tethered cord effectively relieves pain and may arrest neurologic deterioration.[10]

ORTHOPEDIC IMPAIRMENTS

Muscle weakness leads to abnormal positioning in utero. Consequently, 50% of the babies with myelomeningocele have significant foot deformity at birth ("clubfoot"), including calcaneovalgus, equinovarus, and vertical talus (**Fig. 5**). During early childhood, further deformity may occur from ongoing muscle imbalance, postural effects of gravity, and growth. A plantigrade foot in neutral position is essential for optimal walking, and a well-positioned foot may protect against skin breakdown. Hence physical therapy, casting, subcutaneous releases, and postoperative splinting are commonly needed in infancy. More definitive surgical reconstruction, including releases, tendon transfers, and bony surgery, may be required at the age of around 12 to 24 months, and bracing is usually required to maintain alignment and improve mobility.[11]

Hip flexors and adductors are innervated by L1 and L2, whereas hip extensors and abductors are innervated by L5 and S1. Hence, muscle imbalance and hip instability are common in spina bifida. Subluxed or dislocated hips occur in 25% to 50% of
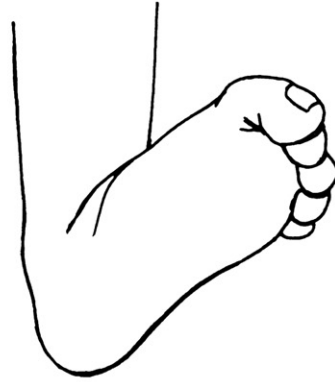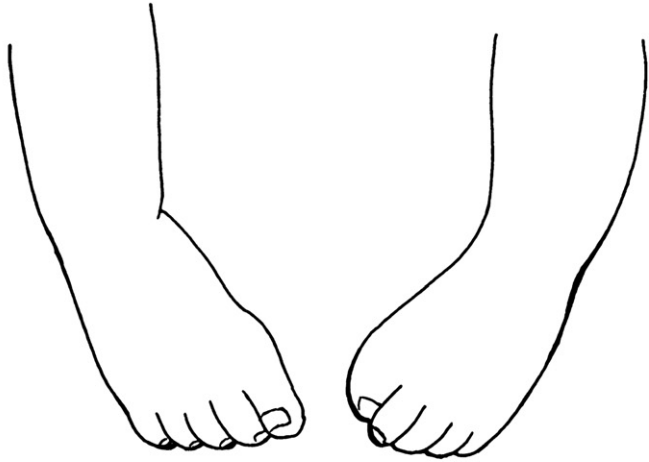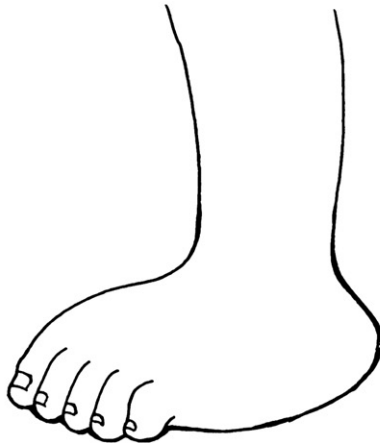
Fig. 5. Typical appearances of foot deformities in spina bifida, including calcaneovalgus, equinovarus, and vertical talus. (*From* Sandler A. Living with spina bifida: a guide for families and professionals Chapel Hill (NC): University of North Carolina Press; 1997. p. 137; with permission.)

newborns with high or midlumbar lesions, and another 25% become unstable during early childhood. The main concern is not the effects on walking but that asymmetric hips and associated pelvic obliquity may cause scoliosis, seating problems, and pressure sores.

Scoliosis may be congenital or acquired. Congenital scoliosis occurs in 15% to 25% of newborns with spina bifida, most commonly with thoracic lesions. Acquired (or "paralytic") scoliosis is usually first noted in early school age, and the condition may progress rapidly, especially during puberty. Tethering and hydromyelia, pelvic obliquity, and asymmetric motor function may cause progressive scoliosis. Severe kyphosis may be present at birth, most commonly associated with lumbar myelomeningoceles, posing a challenge to the surgeons performing the primary closure. Severe scoliosis and kyphosis may interfere with sitting and walking, and may compromise respiratory function. Lightweight molded orthoses may prevent progression and/or delay the need for spinal fusion and stabilization with rods.

BLADDER AND BOWEL DYSFUNCTION

Myelomeningocele is almost always associated with neurogenic bladder.[12] Despite normal urinary tracts in 90% of the children on ultrasonography, 1 in 3 newborns has a "hostile bladder" and is at risk for developing hydronephrosis and renal scarring. Urodynamics (also known as video urodynamic studies) is valuable in the diagnosis of bladder dysfunction, detecting those with hostile features, such as high bladder pressures and outlet resistance (detrusor-sphincter dyssynergia). Clean intermittent catheterization (CIC) and anticholinergics, or vesicostomy, may be needed in the neonatal period to prevent renal complications. Monitoring the urinary tract with ultrasonography every 6 to 12 months is important to detect pelvicaliectasis or hydronephrosis. Also, catheterized urine samples are important to detect bacteriuria, and a voiding cystourethrogram may be needed to rule out vesicoureteral reflux. The incomplete emptying of a neurogenic bladder predisposes to bacteriuria, but this condition is usually asymptomatic and does not require treatment. However, in the presence of high bladder pressures and/or vesicoureteral reflux, the kidneys are at risk for pyelonephritis. Hence, babies with reflux should be on prophylactic antibiotics and CIC.

Children with neurogenic bladder are unable to perceive bladder fullness and lack coordination between detrusor contraction and sphincter relaxation. For some, this lack of coordination poses a risk to their kidneys, but for all it presents a challenge of continence. Lapides and colleagues[13] revolutionized the management of spina bifida with the advent of CIC. Detailed accounts of the techniques are available elsewhere.[2] In addition to CIC, pharmacologic management includes the use of anticholinergic medications to inhibit detrusor contractions and increase storage volumes. With CIC 4 or 5 times daily, anticholinergic medications, and good clinical care, 70% to 90% of children are reliably dry or have only occasional episodes of wetting. Some children with very small capacity bladders may require surgical augmentation with the cecum (enterocystoplasty). One variation is the Mitrofanoff procedure, in which the appendix is used to connect the augmented bladder to the abdominal wall at or near the umbilicus and the continence ostomy is then used for CIC.

Most children with spina bifida have a neurogenic bowel, developing constipation because of decreased bowel motility. Efforts to control constipation in early childhood can help to prevent urinary tract infections. Initiation of a bowel program by the age of 3 to 5 years with regular assisted evacuation of stool is an important habilitation goal. The social acceptance of the young child at school entry is enhanced by having predictable bowel movements and few episodes of incontinence. The choice of bowel

management methods should be based on individualized assessment and include education to enhance the family's motivation and treatment adherence.[14] Methods for bowel management include habit training, digital stimulation, suppository or mini-enema daily (or every other day), cone enema (with colostomy irrigation kit), and Malone antegrade continence enema (ACE). The ACE has become accepted as an important salvage procedure for intractable constipation and fecal incontinence.[15]

COGNITIVE IMPAIRMENTS

Individuals with spina bifida frequently have below-average cognitive abilities, and mild intellectual disability is not uncommon.[16] Abstract reasoning, visual perceptual abilities, and visual motor integration are typically weak. Verbal reasoning score tends to be higher than that for nonverbal reasoning. Higher-level lesions are associated with lower IQ, although this association does not have predictive usefulness. The presence of hydrocephalus is not in itself a major risk factor for intellectual development. Indeed, many children with myelomeningocele and without hydrocephalus have a neuropsychological profile similar to those with hydrocephalus. Among those with hydrocephalus and shunts, ventriculitis and other major shunt complications may lead to acquired brain injury and more severe intellectual disability. Attention, organization, and executive functions may be impaired, and many children may meet the criteria for attention-deficit/hyperactivity disorder. Children of school age may show relative strengths in social skills, expressive language, reading, and spelling. Problem solving, writing, and math skills are typical areas of weakness.[17]

The combination of paralysis and cognitive impairments are major challenges to the development of independent living skills. Doting parents and other family members can unwittingly behave in ways that encourage dependency. An important goal of habilitation is for the child with spina bifida to assist in self-care and to participate in activities of daily living. Young children should be encouraged to assist in diaper changes, putting on braces, bathing, dressing, and catheterization. Beginning this process early helps to prevent learned helplessness, dependency, and severe impairments in adaptive function.

SKIN PROBLEMS

The most common cause of skin injury is pressure from prolonged sitting in one position. Persistent redness over the ischial tuberosity may progress to blistering and skin breakdown. Pressure sores can be extraordinarily slow to heal, and exact an enormous cost both in health care expenditures and in loss of function. Other common injuries include burns and trauma to insensate feet, emphasizing the importance of health education and anticipatory guidance in prevention of secondary disability.

OTHER PEDIATRIC HEALTH ISSUES

The motor paralysis of spina bifida leads to decreased caloric expenditure and obesity in at least 20% of school-aged children. Obesity may seriously compromise mobility and pose additional challenges to optimal health and habilitation. Changes in body mass index and skin fold thickness are useful indicators of obesity in the clinic. Nutrition education, motivational interviewing, and family-based support may help to prevent obesity.

Short stature is common in spina bifida, affecting 80% of those with lesions at or above L3. Small lower limbs, spinal deformities, and scoliosis may contribute, but differences in frequency and amplitude of growth hormone secretion have been

reported. Other neurosecretory abnormalities have been implicated in the development of precocious puberty in children with spina bifida and hydrocephalus. Referral to a pediatric endocrinologist may be needed for control of rapidly progressing puberty with leuprolide acetate (luteinizing hormone receptor inhibitor).

After a report of a cluster of cases of intraoperative anaphylaxis in children with spina bifida in 1989, it was estimated that 18% to 40% had latex allergy. Latex allergy is more common in children with a history of allergies and asthma and in those who have had multiple operations. The Nursing Council of the Spina Bifida Association has spearheaded efforts worldwide to raise awareness of latex allergy and institute latex precautions for primary and secondary prevention, thereby decreasing morbidity and mortality from this common complication.[18]

THE HISTORY OF CARE OF THE CHILD WITH SPINA BIFIDA

The history of health care of children with spina bifida has included several revolutionary and dramatic advances. In 1956, engineer John Holter's only son Casey was born with a myelomeningocele. Holter's efforts to save his son's life led to the development of the Spitz-Holter valve and other effective shunt valves.[19] In the 1960s, ventriculoatrial shunting was commonly done, but V-P shunting soon became the standard treatment (**Fig. 6**). Although much progress has been made in the prevention of ventriculitis, there continues to be a high incidence of unpredictable shunt failure. In the past 10 years, alternative techniques for treating hydrocephalus have been available, including the endoscopic third ventriculostomy.[20,21]

In 1972, Lapides and colleagues[13] revolutionized the management of spina bifida using CIC. This simple procedure allowed complete bladder emptying, thereby protecting the kidneys from infection and preventing or reversing hydronephrosis. This technique remains the mainstay of urological management of the neurogenic bladder and has prevented countless deaths from renal failure.

Before the 1960s, most newborns with spina bifida in the United States and United Kingdom were not treated surgically because the prognosis was thought to be poor. In 1971, Lorber[22] and other investigators published criteria based on prognostic factors for selecting which infants to treat and which to allow to die. Until the early 1980s, many nurseries used these criteria to select infants for treatment or nontreatment, although some centers, most notably the Children's Memorial Hospital in Chicago, questioned the appropriateness of selection and published large series on consecutive nonselected patients with myelomeningocele.[5] Based on the birth of a girl (Baby Jane Doe) with spina bifida and hydrocephalus in 1983, whose parents declined surgery, the US Congress adopted Baby Doe rules as amendments to the Child Abuse and Neglect Funding Requirements for the United States, mandating the provision of life-sustaining medical treatment to seriously ill infants. Since then almost all newborns with spina bifida in the United States have been treated.

In 1970, Hide and Semple[23] described a comprehensive multidisciplinary outpatient clinic in Oxford, England, attended by a pediatrician, orthopedic surgeon, pediatric surgeon, urologist, physical therapist, nurse, and social worker. Similar spina bifida clinics were developed in the United States and were functional in almost every state during the 1980s and 1990s. During the past 10 to 15 years it appears that financial pressures, and perhaps declining prevalence of spina bifida, have affected outpatient programs for children with spina bifida, affecting the participation of some specialists and the extent of care coordination.

In 1972, Brock and Sutcliffe[24] reported the association between increased amniotic α-fetoprotein (AFP) and NTDs. This knowledge opened the door to maternal
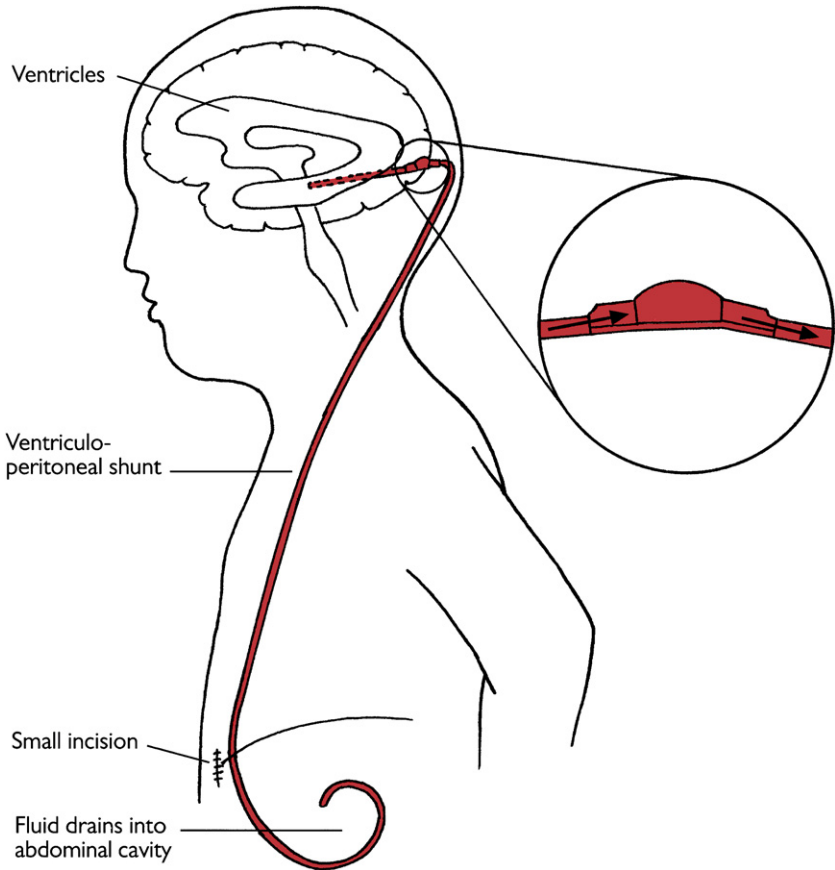
Fig. 6. V-P shunt. Excess CSF drains from the ventricles through a ventricular catheter and pressure-sensitive valve into the peritoneal space. (*From* Sandler A. Living with spina bifida: a guide for families and professionals Chapel Hill (NC): University of North Carolina Press; 1997. p. 66; with permission.)

serum AFP screening between 15 and 20 weeks of gestation. Comprehensive screening programs that include counseling, AFP screening, high-resolution ultrasonography, and amniocentesis have been an important breakthrough in the management of NTDs.

In 1981, the first attempts to surgically treat hydrocephalus in the human fetus were reported, but subsequent case series showed an unacceptably high rate of morbidity and mortality. In 1997, in utero repair of myelomeningocele via hysterotomy began, based on animal models and evidence that neurologic function deteriorates during gestation.[25] By 2003, 234 women had fetal repair of myelomeningocele, and preliminary evidence showed that only 49% of infants had a shunt placed by the age of 1 year.[26] The Management of Myelomeningocele Study (MOMS) is a randomized trial of prenatal versus postnatal myelomeningocele repair funded by the National Institute of Child Health and Human Development, which is nearing completion at 3 clinical centers. The trial has 2 primary end points: the need for shunting by 12 months and neurodevelopmental outcome (Bayley scores and functional motor level) at 30 months. To date, more than 150 of the planned enrollment of 200 subjects have been

randomized, and it remains to be seen whether prenatal surgery represents another breakthrough in the treatment of spina bifida.

ETIOLOGY OF NTDS: RECENT ADVANCES

Folates are cofactors for one-carbon transfer reactions that are important in the biosynthesis of methionine and proteins. Deficiency in 5,10-methylene tetrahydrofolate reductase, an enzyme important in the conversion of homocysteine to methionine, is a risk factor for NTDs. Folate deficiency is associated with elevated homocysteine levels and is also a risk factor for NTDs. Numerous mutant mouse models attest to the ease of disruption of neurulation due to genetic variants, and recent evidence implicates a variety of genetic variants in folate and vitamin B_{12} metabolism that may affect risk for NTDs.[27] Other factors associated with NTDs include chromosomal disorders, maternal exposure to valproic acid, hyperthermia during early pregnancy, and maternal diabetes.

FOLIC ACID AND PREVENTION OF NTDS

The 1991 United Kingdom Medical Research Council trial showed that 4 mg of folic acid around conception and in early pregnancy reduced recurrence risk among women with previous NTD-affected pregnancy from 3.5% to 1%.[28] Several subsequent trials have demonstrated that supplementation with folic acid reduced the risk of NTDs in the general population by 50% to 70%.[29–31]

Natural food folates have poor bioavailability and stability, so prevention efforts have focused on periconceptional supplementation and fortification of foods. Mandatory folic acid fortification of enriched grain products marketed in the United States since 1998 has been associated with an important decline in prevalence of NTDs of between 35% and 78%, or approximately 1000 fewer NTD-affected pregnancies per year.[32] Brent and Oakley[33] and other researchers have argued that folic acid fortification levels are too low and that prevention efforts could be more effective in the United States and elsewhere. To date, there are no proven adverse effects of folic acid fortification. There is debate on the extent to which folic acid–preventable NTDs are being prevented and whether racial/ethnic variation in prevalence is due to underlying ethnic differences in susceptibility or differences in case ascertainment.[27,34] One of the major challenges in primary prevention of spina bifida is how to reach Latin American women, who have higher risk of NTDs and lower folic acid intakes than non-Latin American women. Current public health strategies include adding folic acid to corn masa flour.

EVIDENCE-BASED PRACTICE IN SPINA BIFIDA: CURRENT CHALLENGES IN QUALITY CARE

It is hardly surprising that such a complex and multisystem birth defect continues to challenge clinicians in their efforts to provide evidence-based answers to important clinical questions and dilemmas. In 2003, the National Institutes of Health, Centers of Disease Control and Prevention, Spina Bifida Association, and others sponsored a multidisciplinary symposium to highlight key questions and priorities for further clinical research.[35] A few clinical questions highlighted at that symposium are listed to illustrate the scope of the challenge and opportunity for further important research:

- What is the optimal treatment of hydrocephalus?
- What is the best method for evaluating and monitoring shunt function?

- Which patients would benefit most from tethered cord release?
- What is the optimal management of hip dislocation?
- How to prevent and manage osteoporosis?
- How can animal models be developed to explore neuromodulation of bladder function?
- What is the optimal management of reproductive issues in women with spina bifida?
- What are the pathophysiologic factors that affect skin breakdown?
- What are the instructional and developmental interventions that facilitate learning?
- How best to promote optimal psychosexual development and sexual adaptation?
- How to effectively teach and promote self-care?

Further advances in spina bifida health care will require a commitment to outcomes-based clinical research and would be greatly facilitated by translational research, multicenter trials, and development of a registry. This need is illustrated by the following 3 cutting-edge areas of clinical research: management of hydrocephalus, innovative approaches to enhance bladder function, and transition to adult health care.

Approximately 40% of new shunts fail within a year, and 80% within 10 years. It is troubling that outcomes of treatment of hydrocephalus have changed little in recent decades despite advances in shunt technology. This lack of improvement has led to a renewed focus in neurosurgery on understanding the pathophysiology of hydrocephalus and developing more effective treatment for this condition.

DISCUSSION OF CONSERVATIVE SHUNT PLACEMENT (THE CHICAGO EXPERIENCE)

Another new area of research is the procedure described by Xiao and colleagues[36] for rerouting functioning lumbar motor nerves to sacral root motor neurons in an effort to innervate the lower urinary tract. Several children who have undergone this procedure in recent years have been able to initiate a reflex bladder contraction by scratching or rubbing the ipsilateral sensory dermatome, but questions about effectiveness and concerns about transient weakness remain unresolved.

Transition to adult health care remains a major challenge. Much of the progress in management of spina bifida has come from the pooled experience of comprehensive multidisciplinary clinics in pediatric settings. Young adults leaving these pediatric settings struggle to find continuing sources of health care. The burdens are especially acute because of the lack of care coordination and insurance coverage in this age group.

A FUNCTIONAL HOLISTIC DEVELOPMENTAL PERSPECTIVE

This article summarizes some of the key clinical issues in the care of children with this complex birth defect. The need for a coordinated approach among clinicians of different subspecialties is clear. In addition to the core pediatric surgical specialists (neurosurgeons, orthopedic surgeons, and urologists), nursing and allied health professionals (including physical therapists, occupational therapists, and psychologists) help to enhance family education and the child's functional outcomes. A pediatric developmental perspective emphasizes a child's growth and changing capacities, assessing a child's strengths and weaknesses and helping to anticipate

challenges and opportunities. Optimal management of the whole child with spina bifida involves more than the sum of the subspecialty care of his or her parts. Instead, it calls for a flexible and dynamic partnership between the clinicians, the parents, and the child; one that continually strives to enhance health, self-care, learning, and participation in activities and ultimately secures a life of quality.

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