CHAPTER 25

Disorders of the Spinal Cord

Myelomeningocele

INTRODUCTION

Management of the child with myelomeningocele is one of the most challenging tasks that pediatric orthopaedic surgeons face.* Typically, patients with myelomeningocele are referred to as having "spina bifida," but a more specific definition of terms is in order. Myelomeningocele is one of the more severe forms of *spinal dysraphism,* which also includes meningocele, lipomeningocele, and caudal regression syndrome (or lumbosacral agenesis). *Neural tube defects* is another collective term, encompassing the disorders of anencephaly, myelomeningocele, and encephalocele. *Spina bifida occulta* is the mildest form of spinal dysraphism. This condition can be as simple as a radiographic curiosity or an incidental finding of incomplete formation of the posterior arch of the spinal column, usually identified in the lower lumbar or sacral spine. Typically neither the overlying skin nor the underlying neural elements are affected, and the patient is otherwise completely normal on both clinical examination and imaging studies. In occasional patients, spina bifida occulta may be associated with an overlying sinus, fatty deposit, or hemangioma. In such cases there may be associated myelodysplasia, which will need to be investigated with further imaging of the spinal cord (typically magnetic resonance imaging, or MRI). *Meningocele* is a condition in which the meninges are exposed in a saclike protrusion, almost always posteriorly, but very rarely anteriorly or laterally. Because of the risk of breakdown of the meninges with secondary infection of the central nervous system (CNS), surgical repair is usually required. This lesion may be present in the cervical, thoracic, lumbar, or sacral spine. When located at the base of the skull, it is more often referred to as an *encephalocele.* In meningocele, typically there is no involvement of the neural elements (i.e., there is no myelodysplasia), and thus there is usually no associated bowel, bladder, or lower extremity paralysis. Affected patients do have a higher than average risk for congenital vertebral anomalies, progressive noncongenital scoliosis during growth, or the development of tethered cord syndrome, presumably because of scarring in the meninges, so they will require monitoring during growth. *Myelomeningocele* ("spina bifida" or sometimes "meningomyelocele") is a severe developmental anomaly characterized not only by exposure of the meninges but also by myelodysplasia of the underlying neural elements and CNS malformation. The dysplasia of the spinal cord and nerve roots results in bowel, bladder, motor, and sensory paralysis distal to the malformation in most cases. Patients with myelomeningocele often have other lesions of the spinal cord, such as diastematomyelia and hydromyelia. These lesions may be found at sites remote from the myelodysplastic lesion itself. Furthermore, structural abnormalities of the brain causes hydrocephalus in most cases, potentially compromising neurologic function at yet another level.

Spina bifida is a multisystem disorder that demands a coordinated approach from numerous health disciplines in order to maximize each patient's potential. The orthopaedist should remember that the neurologic dysfunction is rarely limited to that corresponding to the level of spinal column dysraphism. Untreated hydrocephalus, Arnold-Chiari malformations, ventricular shunt revisions, CNS infections, and scarring of the residual spinal cord (tethered cord syndrome) may all compromise what would otherwise be considered a stable neurologic disorder. The orthopaedic surgeon should always document the level of neurologic function. Any loss of function should be appropriately evaluated.

INCIDENCE

The incidence of myelomeningocele varies around the world.* The regional and national variations are possibly due to the different genetic compositions of different populations as well as to environmental factors. Epidemiologic reports usually include the rates for both anencephaly and myelomeningocele, which may vary according to the gestational age from which the study begins. The overall incidence of anencephaly and myelomeningocele for all gestational ages in 14 Texas-Mexico border counties was 14.6 per 10,000 live births during the period 1993 to 1995.[105] Included were 87 cases of anencephaly, 96 cases of myelomeningocele, and 14 cases of encephalocele. The incidence of these defects was slightly higher in Hispanic women (14.9) than in Anglo women (10.6). Excluding cases that achieve a gestational age of less than 20 weeks (as some epidemiologic studies do) alters the rates significantly: in the same study, the rate of anencephaly was 4.9 per 10,000 live births and the prevalence of spina bifida was 6.7 per 10,000 live births

* See references 17, 66, 100, 122, 129, 168, 180, 232.

* See references 3, 43–45, 52, 58, 61, 81, 105, 133, 149, 228, 260.

when only cases reaching a gestational age greater than 20 weeks were included. Racial predilection is also evident in neural tube defects. In a California study covering the years 1990 to 1994, Hispanic women were 45 percent more likely than Caucasian women to carry a fetus affected by anencephaly and Asian women were less than half as likely as Caucasians to carry a fetus affected by myelomeningocele.[81] The birth-prevalence rate of myelomeningocele during the period 1983 to 1990 for 16 states in the United States was reported as 4.6 cases per 10,000 live births. Prevalence by individual state varied from 3.0 per 10,000 in Washington to 7.8 per 10,000 in Arkansas. The ratio of affected females to males was 1.2:1; this slightly increased predilection for females has been noted in other studies.

The incidence of infants born with neural tube defects is decreasing.* Some of this decrease may be due to natural or unidentified causes.[44,58,260] Two identifiable factors also appear to play a role. The more important factor is *prenatal screening* using ultrasonography, measurement of maternal serum alpha-fetoprotein (AFP), or both, and elective termination of the pregnancy. AFP is a protein normally present in fetal tissues and amniotic fluid from weeks 6 to 14 of gestation. With closure of the abdominal wall anteriorly and the neural tube posteriorly, AFP is no longer released into the amniotic fluid, so that amniotic AFP decreases to undetectable levels. If the neural tube or the abdominal wall remains open, AFP remains detectable in amniotic fluid and maternal serum. In Scotland, a maternal serum AFP screening program reduced the birth incidence of neural tube defect by 80 percent.[58,270] In South Australia during the time period of 1966 to 1991, the overall prevalence of neural tube defects was stable at 2 per 1,000 live births. During this period, however, the birth prevalence of anencephaly decreased by 96 percent and the birth prevalence of myelomeningocele decreased by 82 percent as a result of screening by maternal serum AFP levels or midtrimester ultrasonography (or both). This screening program identified 90 percent of anencephaly- and 75 percent of myelomeningocele-affected pregnancies. In France, the overall prevalence of neural tube defects has been stable at approximately 10 in 10,000 live births, but a program of ultrasonography and elective termination of pregnancy reduced the birth prevalence of anencephaly by 100 percent and of myelomeningocele by 60 percent between 1979 and 1994.[6] The second factor has been the administration of folate to the mother before and during pregnancy,† as recommended by the U.S. Public Health Service.[42] Worldwide, the impact of folate administration has been difficult to document, perhaps in part because in the West, 50 percent of pregnancies are unplanned.[83,228]

EMBRYOLOGY

In the embryo, the CNS begins as a dorsal focal thickening caused by proliferation of ectodermal cells. These cells increase in number and in height, ultimately forming a layer of pseudostratified epithelium. As the cells proliferate, a groove forms in the sagittal plane of the cell mass. This groove deepens, bringing the lateral portions of the neural plate toward each other. Contractile proteins located within the superficial margin of these cells are thought to be responsible for the actual contraction and drawing together of the neural folds. Progressive flexion brings the peripheral edges of the neural folds into contact. On about the 21st day, cell adhesion occurs at the point of contact, fusing the neural folds into the neural tube. Initially, fusion occurs near the center of the embryo at a point destined to become the craniovertebral junction. Fusion then proceeds longitudinally in both directions, forming the long neural tube. The cephalic (brain) end of the embryo closes first.

As the neural folds fuse together to form the neural tube, the superficial ectoderm separates from the underlying (now fused) neural ectoderm and fuses with itself across the midline to close the back. The separation of superficial and neural ectoderm creates a plane into which mesenchymal cells migrate. This mesenchyme gives rise to the neural arch of the vertebrae and to paraspinal muscles. Closure of the neural ectoderm into a tubular structure and separation of the neural tube from the superficial ectoderm are critical events in the development of the CNS, and are completed by 4 weeks after fertilization (Fig. 25–1).

The embryonic origin of myelomeningocele likely stems from developmental abnormalities occurring at 26 to 28 days of gestation, during the phase of closure of the neural tube. Abnormalities that develop during this process are referred to as *neurulation defects* and include myelomeningocele and anencephaly. Abnormalities arising in the next phase (canalization), from 28 to 48 days of gestation, are termed *postneurulation defects* and include meningocele, lipomeningocele, and diastematomyelia.[100]

Morgagni is often credited with developing the theory that myelomeningocele results from rupture of the distal end of the neural tube, in 1769.[199] According to his theory, when cerebrospinal fluid (CSF) cannot escape from the ventricular pathways, it flows instead into the central canal of the neural tube, distends the tube, and bursts it open at the distal end of the neural tube, creating the myelomeningocele. It appears unlikely that Morgagni actually developed this theory, because the pathophysiology of CSF flow was not understood at that time. Morgagni's real contribution was to note an association between hydrocephalus and spina bifida. A different mechanism for the development of myelomeningocele was postulated by Gardner and Paget.[94,215] They thought that intrauterine hydrocephalus caused the distal end of the neural tube to rupture, producing myelomeningocele.

It was von Recklinghausen who postulated that myelomeningocele resulted from failure of the neural tube to close.[282] This view was supported by Patten, who showed that overgrowth of the neural tube in myelomeningocele embryos implied lack of closure or interference with the closure of the neural tube.[218]

As has been proved over time, myelomeningocele can be produced both by interference with closure of the neural tube and by rupture of the already closed neural tube.[259] Distention and rupture of the developing spinal cord in mouse embryos can be caused by poisoning the pregnant mouse with vitamin A. Thus, primary failure of the neural tube to close and secondary rupture of the once closed neural tube are both possible causes of myelomeningocele.

* See references 3, 43–45, 52, 58, 133, 228, 260, 270.

† See references 27, 83, 135, 197, 200, 255, 281.

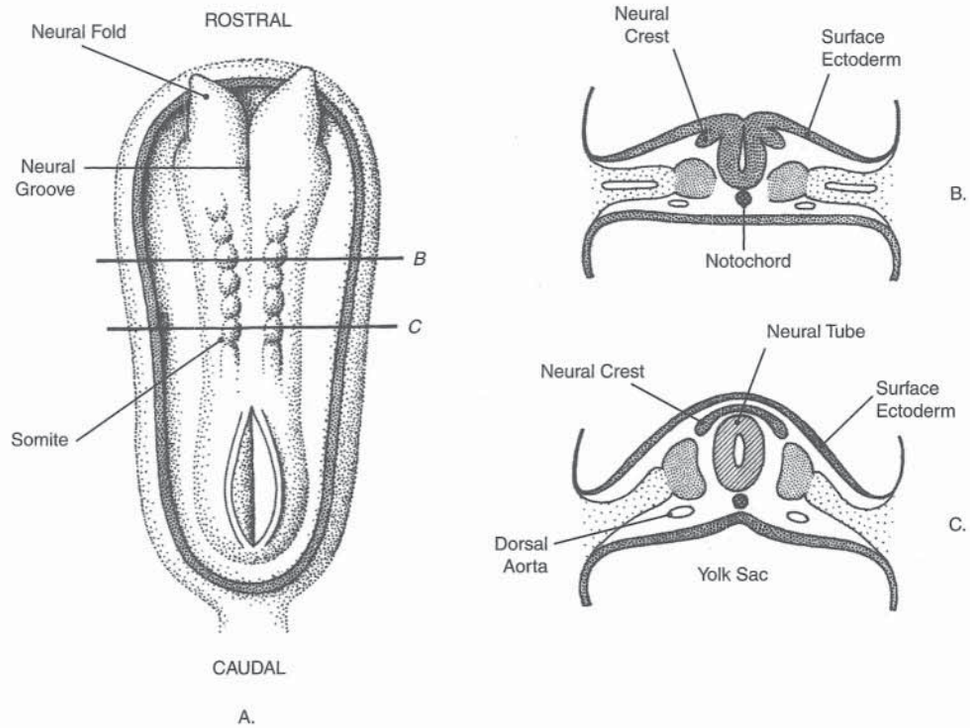


FIGURE 25–1 Embryologic development of the spinal cord. Diagrams demonstrate the formation of the neural crest with infolding of the neural plate into the neural tube. Embryonic appearance at approximately 22 days. **A,** The neural tubes have fused opposite the somites but are widely spread out at both ends of the embryo. Closure of the neural tube occurs initially in the region corresponding to the future junction of the brain and spinal cord. **B,** Cross-section at level *B* (of part A) demonstrating formation of the neural tube and its detachment from the surface ectoderm. **C,** Cross-section at level *C* (of part A). Note that some neuroectodermal cells are not included in the neural tube but remain between it and the surface ectoderm as the neural crest. These cells first appear as paired columns but soon break into a series of segmental masses. (Adapted from Moore KL: The Developing Human, p 4. Philadelphia, WB Saunders Co, 1988.)

ETIOLOGY

The cause of myelomeningocele remains obscure. Genetic factors play a role (discussed under Heredity). Experimentally, minor fluctuations in incubator temperature can prevent normal closure of the neural tube in chicks.[211] Exposure to the fungus *Phytophthora infestans* by excessive consumption of potatoes was at one time postulated to be an etiologic factor,[226] but other studies have refuted this hypothesis.[50] The most important etiologic factor identified has been the association of folate deficiency in the pregnant female with an increased risk for the development of neural tube defects, including myelomeningocele, in the offspring. One study that compared mothers of children with neural tube defects, mothers of children with other abnormalities, and mothers of normal children found no difference in folate intake during pregnancy among the groups.[196] Most studies,* however, have demonstrated a 60 to 100 percent reduction in the risk of neural tube defects with the administration of adequate levels of folate to pregnant women. The U.S. Public Health Service recommends that all women of childbearing age in the United States who are capable of becoming pregnant should consume 0.4 mg of folic acid per day to reduce the risk of having a pregnancy affected with spina bifida or other neural tube defect.[42] Total folate consumption should normally be less than 1 mg/day.

HEREDITY

Genetic factors appear to play an important role in myelomeningocele. There is a significantly greater incidence of neural tube defects, including myelomeningocele, in the siblings of children affected with anencephaly or myelomeningocele than in the general population.[69,115,153,157,254,293] The familial incidence of major neural tube defects has been reported as 6 to 8 percent.[69,115,153,157,254,293] Lorber and Salfield studied the family histories of 722 infants with myelomeningocele.[154] Of the 1,256 siblings, 85 (6.8 percent) had CNS malformations: myelomeningocele in 54, anencephaly in 22, and simple hydrocephalus in 9. For a couple with an affected infant, the risk of subsequent siblings incurring a major malformation of the CNS is approximately 1 in 14.

The exact nature of this increased familial incidence is not understood. The risk is higher in larger families and in specific socioeconomic and geographic groups. Lorber found an increased incidence of spina bifida occulta in the parents of children with myelomeningocele (overall prevalence of 21.4 percent, compared with 4.5 percent in adult controls).

* See references 27, 135, 197, 200, 202, 255, 281.

Thus, families with a history of neural tube defects should be counseled regarding the potential for this development, and pregnancies should be screened (see discussion under Incidence, above).

PATHOLOGY

A thorough description of the pathologic findings of myelomeningocele was given in 1888 by von Recklinghausen, who accurately dissected out both the spinal cord and the meninges in cases of myelomeningocele and recognized every variety of spina bifida.[282]

Lesions may occur at any level along the spinal column but predominate in the lumbosacral area. They are next most frequently found in the cervical spine area (usually as an encephalocele or meningocele only), and a smaller number of lesions are scattered along the thoracic spine. The great majority of the lesions are posterior, but very rarely an anterior or lateral meningocele may be encountered. The anterior cysts protrude through the vertebral bodies, not through the vertebral arches.

The basic deformity of myelomeningocele is an open neural placode, which represents the embryologic form of the caudal end of the spinal cord. A narrow groove passes down the placode in the midline. This represents the primitive neural groove and is directly continuous with the central canal of the closed spinal cord above (and occasionally below) the neural placode. CSF passes down the central canal of the spinal cord and discharges from a small pit at the upper end of the placode to bathe the external surface of the neural tissue. This fluid does not indicate rupture of the myelomeningocele.

Skin. Skin over a myelomeningocele sac is almost always absent. Between the skin and the neural placode is a zone of thin epithelium. At points, skin may actually reach the edge of the neural placode. In the usual type of lesion, there is a raised mass on the back that is covered laterally at its base by normal skin, but the apex of the mass is devoid of skin (Fig. 25–2). It is covered by a tissue-paper-thin membrane (arachnoid) through which one may see nerve roots. Within a day or two, this tissue breaks down to an ulcerated granulating surface. The lesion may heal over completely by epithelial growth from the periphery. More often, however, the mass will slough from secondary infection, which in turn usually leads to meningitis and death without intervention. Hemangiomatous or other pigmented lesions are frequently seen in the skin surrounding the sac.

Meninges. Underlying the neural placode is the arachnoid sac and subarachnoid space. Because the superficial (dorsal) surface of the neural placode represents the everted interior of the neural tube, the deep (ventral) surface represents the entire outside of what should have been a closed neural tube. Thus the ventral nerve roots and the dorsal nerve roots arise from the deep (ventral) surface of the neural placode and pass through the subarachnoid space to their root sleeves. Because the placode is everted, the two dorsal roots are lateral to the two ventral roots.

Within a few millimeters of the edge of the skin is the junction between skin and dura mater. Outside the dura mater is a true epidural space that contains epidural fat. The underlying vertebral bodies are flattened and widened.

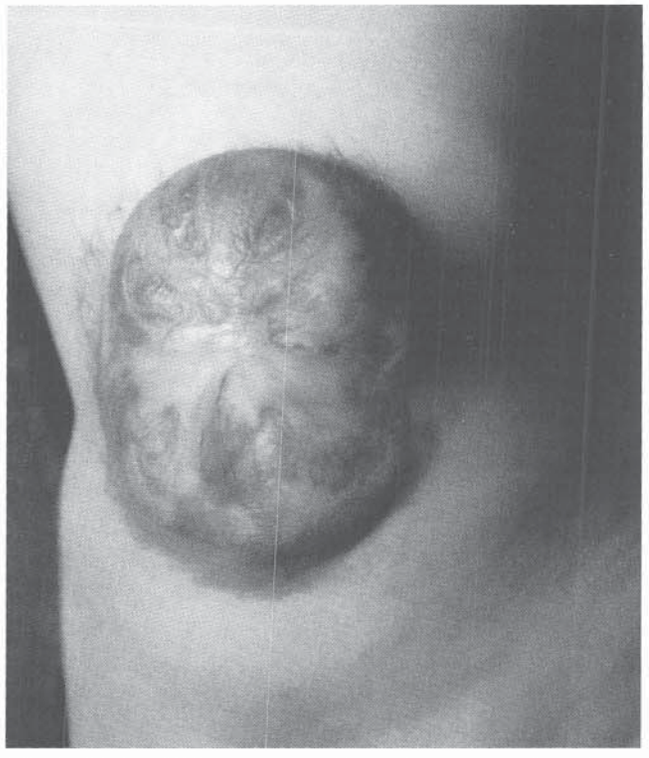


FIGURE 25–2 Clinical appearance of untreated myelomeningocele sac. Note large protrusion of the meninges, without protective skin. Breakdown of the sac usually occurs and is followed by further neurologic injury, meningitis, and potentially encephalitis.

The pedicles are everted and lie nearly horizontal in the coronal plane. The laminae are hypoplastic and often are also everted. The spinous processes are by definition absent. The paraspinal muscle masses are present but are everted with the pedicles and laminae. Thus, they lie anteriorly and often act as flexors and not extensors of the spine. The muscles may be markedly attenuated owing to the lack of innervation from the CNS.

The size of the sac on the child's back at the time of birth depends on the amount of spinal fluid that is collected ventral to the neural placode.

Spinal Cord. Dysplasia of the spinal cord is invariably present. The cord may be (1) cystic or cavitated, (2) solid but degenerated and disorganized, or (3) grossly proliferated. Frequently all these features are found together in varying degrees.

Peripheral Roots. Peripheral nerve development is not affected in myelomeningocele. At surgery and on dissection of the postmortem specimen, normal peripheral roots have been found in every case. However, inside the dura mater the roots appear to have tenuous connections with the cord itself and on occasion are hard to identify.

Vertebrae. The principal defect is the arrest of development of the laminae. The spectrum of failure varies from failure of the laminae and vertebral spines to fuse posteriorly to, at the other extreme, total failure of formation of laminae, the pedicles alone being present. The intraspinal canal is widened as a result of lateral displacement of the pedicles on the vertebral bodies.

Brain. In myelomeningocele there may be associated anomalies of the cerebellum and brain stem, such as a Chiari type II deformity, in which the posterior lobe of the cerebellum, the medulla, and the fourth ventricle have herniated through the foramen magnum into the cervical spinal canal; in the more severe Chiari type III malformation the entire cerebellum and lower brain stem are inferior to the foramen magnum (Fig. 25–3). Hydrocephalus develops from obstruction of CSF flow at the roof of the fourth ventricle by dislocation of the ventricle, by occlusion of the subarachnoid space at the site of herniation, by occlusion of the same space at the tentorial level by adhesive arachnoiditis, or by an associated aqueduct stenosis. Other causes of hydrocephalus in myelomeningocele are the Dandy-Walker malformation, which consists of marked distention of the fourth ventricle due to occlusion of the foramina of Luschka and Magendie; "forking" of the aqueduct of Sylvius, in which the aqueduct is represented by two narrow channels situated in a sagittal plane; and aqueduct stenosis. Radiologic studies of CSF dynamics in children with hydrocephalus have shown increased production of CSF. Secondary changes in the brain develop as a result of increased pressure due to the hydrocephalus.

TREATMENT

General Principles

NATURAL HISTORY. Prior to the introduction of the Holter valve for the shunting of hydrocephalus and adequate closure of the myelodysplastic lesion, death frequently occurred in infancy because of the hydrocephalus or sac breakdown followed by meningitis; any survivors usually succumbed to renal failure. A study by Rickham and Mawdsley[1a] found that only two of 57 infants who were not treated surgically survived, one each with meningocele and encephalocele. All patients with myelomeningocele died within 6 months of birth. Other studies found similar results.[150] Shunting of the hydrocephalus combined with sac closure led to a significant increase in survival. Unfortunately, this commonly resulted in a large number of severely handicapped children.[150,190] This led to the introduction of criteria for selecting infants for aggressive surgical care.[149–152,154] Specific criteria mitigating against aggressive surgical treatment of patients born with myelomeningocele were proposed by Lorber after a review of 524 cases.[150] He found that the presence of severe paralysis (upper lumbar or higher), head circumference at or above the 90th percentile, congenital kyphosis, or the presence of other major congenital anomalies such as heart disease and severe birth injury were associated with a significantly greater likelihood of nonsurvival in infancy or severe handicap in survivors. These factors became known as "Lorber's criteria," and their presence at birth has been taken as a contraindication to aggressive surgical intervention of such affected children.[47,150–152] A follow-up study by Lorber and Salfield in which these criteria were applied to 120 children found that all 71 children presenting with adverse criteria and not treated surgically died, 90 percent of them within 6 months of birth.[154] Seven had meningocele, were treated, and survived without handicap. Of 42 infants with myelomeningocele treated actively, 36 were alive 3 to 9 years later. The authors felt that the quality of life in the survivors was much greater than in their prior assessments when selection criteria were not used. Menzies and colleagues,[190] however, noted that more than one-third of 27 infants cared for in the home and not offered aggressive surgical treatment lived more than 2 years, and 30 percent of them lived long enough to attend school. Surana and colleagues[266] reported that the level of paralysis was not an indicator of survival in patients not treated surgically, and that infants who survived more than 3 months were far less likely to die in the first year. Charney and colleagues assessed the influence of timing of surgical intervention on survival, developmental delay, and loss of motor function.[47] They found comparable survival rates (90 to 100 percent) for infants treated within 48 hours of birth, between 3 and 7 days of age, and between 1 week and 10 months of age, without evidence of loss of motor function or increased developmental delay for the later treatment groups. They concluded that surgical treatment need not be "emergent." Swank and Dias assessed the outcome of 206 children treated by closure of the spinal defect within 48 hours of birth without selection criteria.[267] Of these 206 patients, 192 required shunts for hydrocephalus. Apparently, all survived. At the present time, in most U.S. centers, specific criteria for early surgical treatment are not set, and the parents of all affected infants are offered surgical closure of the sac, followed almost invariably by ventriculoperitoneal shunting.[232]

Long-Term Outcome

GENERAL. Laishram and colleagues reviewed the functional status of 179 patients with myelomeningocele born in Newfoundland or Labrador between 1967 and 1990.[130] Thirty-five percent were wheelchair bound and the rest were ambulatory. The majority of schoolage children attended regular schools. The authors concluded that most of the patients attained a surprisingly high quality of life.

AMBULATION. Factors affecting both short- and long-term potential for ambulation have been addressed in a number of studies.* The single most important prognostic factor for maintaining ambulation in adulthood seems to be the strength of the quadriceps muscle.[7,14,66,113,237,264] Barden and colleagues, in a review of 29 adult patients with myelomeningocele, ages 20 to 43, found that only two of nine thoracic/upper lumbar level patients walked, whereas 19 of 20 lower lumbar/sacral level patients walked.[14] The status of the hip did not correlate with the ability to walk in their study. Asher and Olson evaluated the ambulatory status of 98 patients ages 5 to 31 years.[7] Twenty of 21 patients with sacral and L5 level paralysis were community ambulators and most patients with L4 level paralysis were community ambulators, but most patients with L3 level paralysis or above were not functional ambulators. Additional factors in nonambulation were obesity, hip deformity, scoliosis, foot and ankle deformity, and age. Hoffer and colleagues reviewed the ambulatory status of 56 patients who were at least 5 years old and found that no patient with thoracic level paralysis was a community ambulator but all patients with sacral level paralysis were.[108] This study found no significant difference in ambulatory status in patients with different levels of lumbar

* See references 7, 14, 31, 46, 66, 73, 79, 80, 87, 103, 108, 113, 136, 161, 171, 176, 206, 212, 221, 237, 252, 264, 268.

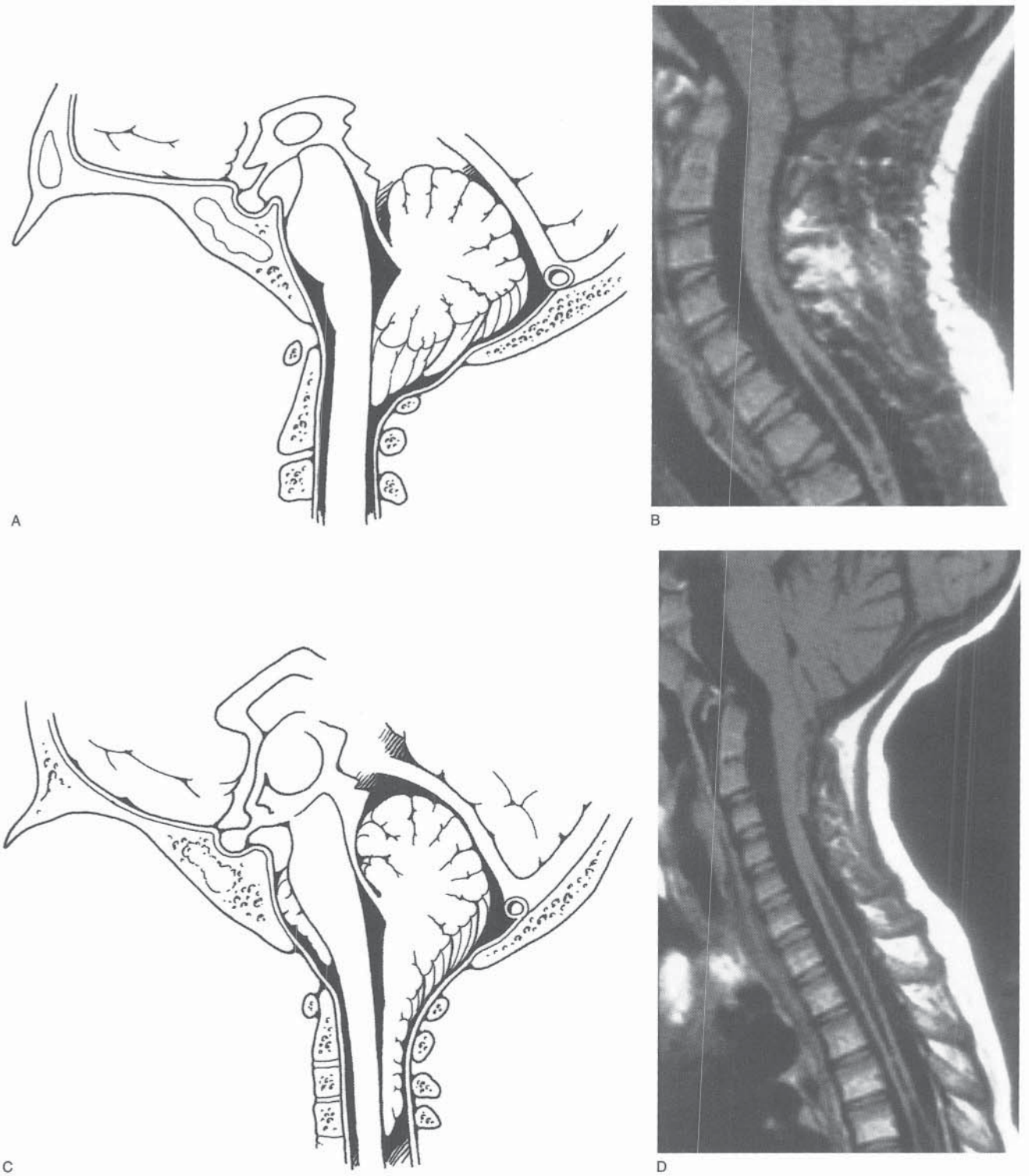


FIGURE 25–3 Arnold-Chiari malformations of the brain stem. **A,** Type I Arnold-Chiari malformation: cerebellar tonsillar herniation only. **B,** Appearance of type I Arnold-Chiari malformation on MRI. Note associated cervicothoracic syringomyelia. **C,** Type II Arnold-Chiari malformation: more extensive herniation of the cerebellum and brain stem through the foramen magnum. Type II malformations are more commonly seen in patients with myelomeningocele. **D,** Appearance of type II Arnold-Chiari malformation on MRI. Note associated cervicothoracic syringomyelia.

level paralysis. Of a group of 40, 14 were community ambulators, 5 were household ambulators, 2 were nonfunctional ambulators, and 19 were nonambulatory. Schopler and Menelaus reviewed the maintenance of quadriceps muscle strength longitudinally in a group of 109 patients with myelomeningocele.[237] They found that only 4 of 51 with normal quadriceps strength in the first 3 years of life demonstrated deterioration in strength over time. Twenty-one of 22 patients initially assessed as having at least grade 4 strength improved during follow-up, but none of 36 patients assessed as having less than grade 4 strength improved. Quadriceps strength was strongly correlated with ambulation ability: 98 percent of those with grade 4 or 5 quadriceps strength were at least household ambulators and 82 percent were community ambulators, whereas 88 percent of patients with grade 0 to 2 quadriceps strength were nonambulatory. McDonald and colleagues reviewed the relationship between lower extremity muscle strength and ambulatory status in 291 children.[176] They noted that variations from the classic neurosegmental pattern as described by Sharrard[247] were often identified, and that there was considerable variability in ambulatory status of patients with lesions at the same neurosegmental level, especially in the midlumbar level. Specific muscle strength by group predicted 86 percent of the mobility outcome. They found that all patients with iliopsoas strength of grade 3 or less relied on wheelchairs for some or all of their mobility, whereas none of those with iliopsoas strength of grade 4 or 5 relied *solely* on wheelchairs. Strong quadriceps were an important determinant of household ambulation in patients with weak iliopsoas muscles. Patients with good iliopsoas and quadriceps strength and antigravity gluteal strength could be expected to ambulate without the need for a wheelchair, and those with grade 4 or 5 gluteal and tibialis anterior strength usually walked without aids or braces. Swank and Dias found that achieving sitting balance was an important predictor of ambulation potential.[268] When the sitting balance factor was combined with motor level function, the ability to walk could be predicted in 92 percent of 206 children (ages 0 to 10 years) with myelomeningocele.

HEALTH PROBLEMS OF THE MYELOMENINGOCELE PATIENT

General/Universal Problems. A trap awaiting the inexperienced physician is the perception that myelomeningocele represents a congenital lower extremity paralysis that can be characterized by the "level of the lesion," with a readily definable border between functioning and nonfunctioning motor and sensory root levels and a predictable lower extremity and total patient function to match. In fact, myelomeningocele is a complex congenital anomaly that is often dynamic and changing in the neuromuscular components that influence the patient's mobility capabilities and the orthopaedic surgical requirements. In addition, patients typically have bowel and bladder paralysis, CNS anomalies (especially hydrocephalus), and congenital anomalies of the spine and lower extremity, all of which confound the clinical picture. Neurologic function can change over time as a result of unchecked or complicated hydrocephalus or scarring of the spinal cord. The most important organ systems requiring management in these patients (in addition to the musculoskeletal system) are the neurologic system, the gastrointestinal system, and the genitourinary system.

Upper Extremity Function in Myelomeningocele. Upper extremity function is often disturbed in patients with myelomeningocele.* Turner[277] found only two of 33 and Jansen's group[119] only two of 25 patients with myelomeningocele whose hand function was normal. Upper extremity dysfunction is secondary to neurologic impairment by hydrocephalus, brain stem compression by the Arnold-Chiari malformation, hydromyelia involving the cervical spinal cord, and cerebral insult consequent on the placement of ventricular shunts or infection of these shunts. This dysfunction can take the form of spasticity, ataxia, dyspraxia, or a combination of these. The presence of spasticity may be particularly important, since Mazur and colleagues found that patients with upper extremity spasticity were less likely to be independent in activities of daily living.[172] Decreased grip strength is common.[134,201] Mazur and colleagues found that patients with higher lesions (thoracic or upper lumbar) and patients who had undergone more than three shunt operations were more likely to have abnormal hand function;[169] Turner, however, found no correlation with the level of the myelodysplastic lesion.[277] Several authors have noted that hand function can improve over time in schoolage children.[119,277] An assessment of hand function by therapists and the orthopaedist is important to establish appropriate goals for mobility aids, activities of daily living, and classroom performance and activities.

Puberty. Girls particularly are at risk for the development of early or precocious puberty.[93,98,223] Proos and colleagues found that 20 of 32 girls had breast development and pubic hair by age 9.[223] They correlated the risk of precocious puberty with increased intracranial pressure and a higher incidence of shunt malfunctions and revisions. Furman and Mortimer also noted that girls with myelomeningocele began menstruating at an average age of 10 years and 3 months, significantly younger than their mothers, siblings, and the U.S. mean.[93]

Cognitive Problems. Cognitive learning difficulties are regularly reported in patients with myelomeningocele, particularly those requiring shunts.[67,84,111] Thus, difficulties at school should be assessed and addressed by the health care team caring for patients with myelomeningocele. One study correlated school performance with walking ability, suggesting that energy consumption devoted to ambulation as opposed to using a wheelchair impaired intellectual performance in school.[84] Performance level tends to improve with increasing age, emphasizing the importance of monitoring the overall health and neurologic function of the child.[210] Children with myelomeningocele do have difficulty adjusting to their peers without disability;[297] interestingly, children with myelomeningocele and normal IQs had a higher rate of psychosocial maladjustment than mentally disabled children in mainstream schools. Rüdeberg and associates have emphasized the importance of coordinated, aggressive rehabilitation in the ability of the child to attend normal schools.[230]

Psychosocial Implications of Myelomeningocele. The impact of the diagnosis of myelomeningocele on the patient,

* See references 119, 134, 169, 172, 201, 212, 277.

family, and the community health care system is significant.* Appleton and colleagues found that a population of children with myelomeningocele, ages 9 to 18, were at greater risk of depressive mood, low self-worth, and self-blame on psychological testing, and that perceived parental social support had a direct effect on depressed mood, particularly in girls.[5] A study of the families of a large Scandinavian myelomeningocele population found that families with a myelomeningocele child coped surprisingly well, compared with control families.[137] They did not have significantly different one- and two-parent family distributions compared to controls and did have a similar mean number of children. However, responsibility for the care of the disabled child fell largely to the mothers, who reported feeling they were receiving adequate support less often than controls; both parents reported more frequent absences from work secondary to illness in the child than controls. Mothers of myelomeningocele children were significantly less likely to work outside the home or did so less than they desired because of the necessity of looking after the disabled child. These findings were not related to the severity of the child's disability. A study by Holmbeck and colleagues found that families with the least physically impaired children reported the most family difficulties.[111] An important study by Rüdeberg and colleagues compared two groups of patients, one group treated from 1970 to 1980, is which patients did not receive early therapeutic intervention, and a second group of patients treated from 1981 to 1992, who did.[230] The authors found that 100 percent of patients in the second group achieved independent locomotion by age 5 years, compared to 35 percent of the first group. Patients in the second group underwent many more orthopaedic procedures to achieve this locomotion, but drastically fewer urologic procedures secondary to early urodynamic rehabilitation. Moreover, 76 percent of the second group achieved normal schooling, compared to 54 percent of the first. Coordinated medical care and physical therapy were very important factors in the quality of life of these children.

Specific Problems by Level

THORACIC LEVEL. Patients with thoracic level lesions essentially have flail lower extremities and, based solely on the total lower extremity flaccid paralysis, would be expected not to develop muscle imbalance–induced lower extremity deformities. In fact, a "frog-leg" deformity is frequently present in these patients at birth, characterized by hip flexion, abduction, and external rotation contractures. There may be in addition knee flexion and ankle equinus contractures. These may respond to judicious passive manipulation, but the hip contractures often will not respond adequately to this treatment, and the surgeon will be faced with the decision of releasing the contractures to allow the lower limbs to be placed in a position for upright mobility. Occasionally, these patients may develop secondary flexion deformities from "spasticity" in the lower extremities, which presumably is really involuntary reflex motor function below the level of the myelodysplastic cord lesion.

The most frequent deformities encountered by the orthopaedic surgeon in this patient group are spinal: congenital scoliosis, developmental scoliosis, and progressive congenital deficiency kyphosis.

Most patients with thoracic level lesions can achieve exercise or household ambulation as children.[46] All require extensive bracing above the hip and upper extremity aids (walker or crutches), and will ambulate with a swing-through gait using their upper extremities and abdominal muscles. Physician and family must be aware that, for most such patients, this is a temporary capability, and, because of the energy expenditure required for such ambulation, they will ultimately choose to use a wheelchair full-time except for transfers.* Charney and colleagues found that compliant parents, physical therapy, and the absence of mental retardation were the most important factors predicting community ambulation in children with thoracic level lesions, whereas scoliosis or hip surgery were not factors.[46] In Swank and Dias's series of 206 patients between the ages of 1 month and 13 years, 71 (34 percent) had thoracic level paralysis.[267] Twenty-four percent of these were community ambulators, 41 percent were household ambulators, and 35 percent were nonambulatory (accounting for all but one of the nonambulators in the entire population). Seventy percent of this group had associated orthopaedic defects at birth, most commonly clubfeet, kyphosis, hip dislocation, and knee flexion deformity.

UPPER LUMBAR LEVEL. Patients with upper lumbar level lesions have hip flexor power and some adductor power, but no motor control of the knees or feet. Their ambulation potential and needs parallel those of patients with thoracic level function for the most part, but theoretically they may be more efficient walkers as children because their hip flexor and adductor strength can be recruited to provide a better swing-through gait, or, with the use of a reciprocating gait orthosis, a reciprocating gait (see subsequent discussion under Orthotic Management). This iliopsoas strength, however, is usually not enough to tip the balance in adolescence and adulthood, when the natural history resembles that in patients with thoracic level lesions in that these patients rarely continue to ambulate as adults.† Hoffer and colleagues, however, did not find differences in ambulation between adult patients with upper and lower lumbar level lesions.[108] Patients in this group experience significantly more paralytic hip dysplasia and dislocation because of the presence of imbalance at the hip, with hip flexors and adductors present, but no hip extensors or abductors.

LOWER LUMBAR LEVEL. Patients with lower lumber level lesions have greater hip adductor strength and, more important, quadriceps power to provide active knee extension. Those with L5 functioning will have a functioning tibialis anterior, and they may have medial hamstring function as well. The hip strength is usually adequate to allow these patients to walk with the hips unbraced, that is, with knee-ankle-foot orthoses (KAFOs). Their gait will exhibit a compensatory combined maximus-medius lurch (the limb in external rotation, and a backward and lateral lean of the trunk over the hip to stabilize it in stance). Although theoretically these patients could walk with AFOs only, in practice this is not commonly the case, since strength around the knee is not

* See references 5, 110, 111, 121, 137, 148, 175, 210, 214, 230, 297.

* See references 7, 14, 46, 66, 108, 113, 170, 171, 176, 237, 268.

† See references 7, 14, 66, 113, 237, 267, 268.

completely normal, and the weakness of the foot, ankle, and hip abductors and extensors leads to the lurching gait, which imposes a great deal of stress on the unbraced knee.[139,272,288] These patients, too, are at high risk for the development of progressive hip subluxation and dislocation. It is in this group that surgical treatment of the hip is most controversial in terms of its influence on long-term ambulation preservation.* In the childhood population studied by Swank and Dias, 33 (92 percent) of 36 were community ambulators, and the other three (8 percent) were household ambulators.[267] Clubfeet and hip dislocation were also frequent in this group.

SACRAL LEVEL. Patients with sacral level myelomeningocele will have near normal knee function, and more stable hip, foot, and ankle function. The partial paralysis and insensate skin still lead to a number of foot problems, including cavovarus deformity, clawtoes, and neurogenic ulcers.† Hip subluxation can occur but is less frequent than in the previous two groups. Knee problems can occur associated with torsional or angular stress with ambulation.[74,139] Excessive ankle dorsiflexion or external rotation may make ankle orthotics difficult to fit for stabilization or ineffective in stabilizing the ankle. In theory, most sacral level patients could ambulate without orthotics. In practice, weak gastrocnemius and foot intrinsics result in abnormal foot and ankle function, just as problems with knee stability are frequent in patients with lower lumbar level paralysis. Gait studies demonstrate that even patients with sacral level myelomeningocele ambulate most effectively with AFOs and crutches because of stresses at the knee and weakness in the foot and ankle.[139,216,280]

Long-term reviews of patients with sacral level paralysis are a sobering reminder of the multifactorial risk of loss of neurologic function and mobility in myelomeningocele.[31,240] Brinker and colleagues reviewed outcomes in 36 patients at an average age of 29 years.[31] Thirty-five patients had been community ambulators and one had been a household ambulator. At review, the ability to walk had declined in 11 of the 35 community ambulators, and the household ambulator had become nonambulatory. Fifteen patients had developed osteomyelitis, and 11 had undergone amputations. Selber and Dias reported better results in 46 slightly younger patients (average age, 23 years).[246] At the final follow-up, 41 patients were still community ambulators. Thirty-nine patients had undergone a total of 217 orthopaedic procedures, and 12 had undergone tethered cord release. The authors concluded that aggressive management of tethered cord syndrome and avoidance of arthrodeses in the foot were major factors in these more favorable results.

Neurosurgical Treatment. The neurosurgeon is an important member of the health care team involved in the management of children with myelomeningocele. The initial challenge faced by the neurosurgeon is closure of the myelomeningocele sac; in 70 to 90 percent of patients sac closure will be closely followed by the need for venticuloperitoneal shunting. In follow-up, the neurosurgeon will be actively involved in the identification and treatment of shunt malfunction, shunt infection, brain stem compression by the Chiari II (or Arnold-Chiari) malformation, the development of hydromyelia within the spinal cord, and tethering of the distal nervous system tissue in scar, producing the so-called "tethered cord."

Closure of the Myelomeningocele Sac. Early closure of the myelomeningocele sac (within 48 hours) is a cornerstone in the management of children with myelomeningocele.[47,60,82,132,181,239] Prior to this protocol, death in infants born with myelomeningocele was virtually universal secondary to meningitis and ventriculitis. Depending on the extent of the dermal defect and the underlying bony deformity (specifically congenital kyphosis), closure can be achieved by direct approximation of the skin over the defect, with or without undermining of the skin, local rotational flaps, or musculocutaneous latissimus dorsi or gluteus maximus flaps. Defects greater than approximately 18 cm^2 are much more likely to dehisce after primary direct closure, and consultation with a plastic surgeon for the purposes of providing coverage of the skin defect with a flap is generally indicated.[60,99,239]

Hydrocephalus. The Chiari II malformation, characterized by herniation of the cerebellum and brain stem, is almost universally associated with myelomeningocele.[164] This deformity, especially after closure of the myelomeningocele sac, produces an obstructive hydrocephalus, resulting in the need for ventriculoperitoneal shunting in approximately 70 to 90 percent of infants. These shunts must be periodically reevaluated by the neurosurgeon for continued function and absence of infection. Despite the presence of a shunt, the developmental delay and learning difficulties frequently seen in patients with myelomeningocele are still presumed to be secondary to hydrocephalus.

Interestingly, although many patients have enlarged ventricles at birth, symptomatic hydrocephalus usually develops only after closure of the myelomeningocele sac. Thus, many infants undergo closure of the sac within 48 hours of birth, develop hydrocephalus, and then undergo ventriculoperitoneal shunt placement. A study comparing staged with simultaneous closure and shunting found that patients treated by the simultaneous technique had a significantly reduced incidence of wound leakage at the closure site and no deleterious effects with respect to shunt failure, hydrocephalus, or CSF infection.[195]

There is increasing evidence that the neurologic deficits in myelomeningocele patients are due to a combination of the primary myelodysplasia compounded by the exposure of the neural elements to amniotic fluid in utero.[38,191,192] This has led to interest in the development of fetal surgical techniques to close the sac in utero, in the hope of limiting the secondary neurologic injury. Preliminary reports indicate that in a small group of patients so treated, there was a reduction in the need for ventriculoperitoneal shunting, from 90 percent to 60 percent.[35] However, this fetal surgery was associated with maternal and pregnancy complications, premature birth chief among them. Further investigations will be necessary before this approach can be universally adopted.

Brain stem compression, presumably by the Chiari II malformation, can lead to respiratory obstruction and apnea.[109] Sleep disturbances related to air hunger, dyspnea, and squeaky voice may all require an assessment by the neurosurgeon as to the possible presence of brain stem compression as the etiology of these complaints.

* See references 4, 34, 38, 72, 79, 80, 85, 87, 103, 136, 221, 252.

† See references 31, 66, 90, 165, 216, 233, 240, 280.

Other Spinal Cord Abnormalities. Patients with myelomeningocele are subject to a number of other spinal cord lesions, in addition to the more obvious myelodysplastic one, that may require assessment or treatment by the neurosurgeon, specifically, hydromyelia, diastematomyelia, and tethered cord syndrome.* Hydromyelia (sometimes termed hydrosyringomyelia) is a dilation of the central canal of the spinal cord. This lesion is often detected as an asymptomatic finding on MRI, but it has been implicated in upper extremity weakness or spasticity in some patients; thus, patients with these clinical findings should undergo MRI and neurosurgical evaluation.[25,30,102] Diastematomyelia is a congenital anomaly of the spinal cord and column consisting of a central "splitting" of the spinal cord by a fibrous, cartilaginous, or bony spicule occupying the central portion of the spinal canal (diastematomyelia is further discussed in Chapter 11, Scoliosis). Myelomeningocele patients with congenital vertebral anomalies in addition to the open spinal canal may have an associated diastematomyelia, which should be investigated by MRI if there are symptoms of lower extremity weakness, spasticity, or back pain or if corrective spinal surgery is planned.

Tethering of the spinal cord in scar tissue at the site of repair of the initial myelodysplastic lesion may be the source of significant symptoms as the child with myelomeningocele grows.† Symptoms that have been attributed to the presence of clinically significant tethered spinal cord include back pain (especially at the site of sac closure), progressive lower extremity weakness, lower extremity spasticity, progressive scoliosis, and changes in bladder habits and function. Since a low-lying conus suggesting spinal cord tethering will be demonstrated on MRI in virtually all patients (Fig. 25–4),[25,26] the diagnosis of tethered cord is usually based on the presence of one or more of the aforementioned symptoms or signs, typical MRI findings, and the exclusion of hydromyelia or shunt malfunction as an alternative cause of the patient's signs or symptoms. Evidence of deterioration in somatosensory-evoked potentials (SSEPs) of the posterior tibial nerve has been used by some to document deterioration of lower extremity function and reversal of this deterioration with surgical untethering.[29,104] With respect to the impact of surgical untethering of the spinal cord in symptomatic patients, Sarwark and colleagues, in a review of 30 patients with low lumbar lesions, found that back pain resolved in all cases.[233] Curves stabilized or improved in 60 percent of patients with scoliosis, and 78 percent of patients with lower extremity weakness stabilized or improved. Spasticity was the finding least affected by surgical untethering, improving in only 43 percent, but stabilizing in the remainder. McLaughlin and colleagues found that intraspinal rhizotomy and distal cordectomy were effective in ameliorating symptoms and lower extremity deformities due to spasticity in patients with thoracic level lesions.[179] This treatment, however, is indicated only in patients with no voluntary lower extremity function and in whom symptoms of spasticity cannot be controlled with lower extremity bracing or surgery.

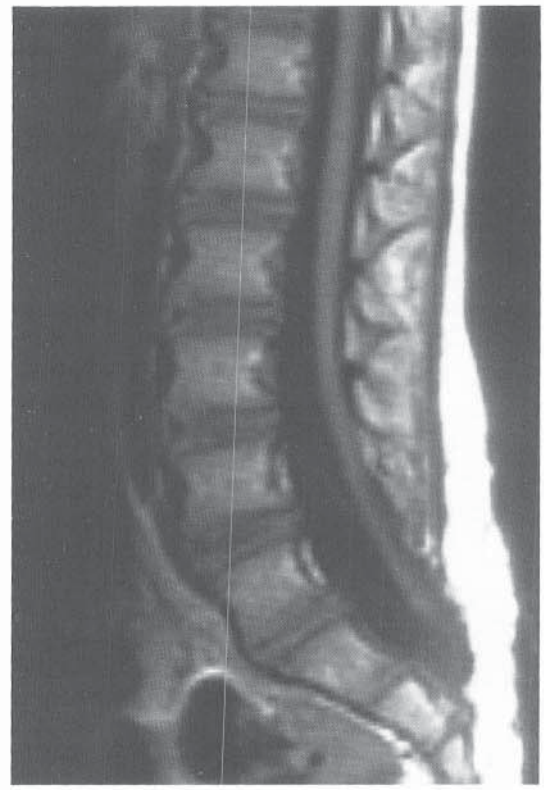


FIGURE 25–4 Appearance of tethered cord on MRI in a patient with myelomeningocele. Normally the conus should end at L1. See text for further discussion of tethered cord in myelomeningocele.

Urologic Treatment. Bladder paralysis and its attendant medical and social problems are a significant source of health and disability issues for the affected child and family.* Bladder paralysis is virtually universal in the myelomeningocele patient population.[138,160] At birth this paralysis is usually flaccid, manifesting as uncontrolled, constant dribbling of the urine. Uncontrolled, spasmodic bladder contractions and bladder neck obstruction commonly develop and can produce overflow dribbling, a smaller, less compliant bladder, and vesicoureteral reflux. Hydronephrosis results, with risk of injury to the renal parenchymal tissue from urinary obstruction or an exacerbating upper urinary tract infection.[253] Lower urinary tract infections are also frequent. Chronic renal failure or fulminant infections of the urinary tract were the most common causes of delayed mortality in patients with myelomeningocele prior to more modern management of the urinary tract.

The goals of urologic management should be to make these patients continent, keep them free of lower and upper urinary tract infection, and preserve renal function. The mainstay of management is to teach caretakers, and ultimately the patient, clean intermittent catheterization.† Such a program helps prevent the development of hydronephrosis and maintain bladder compliance and capacity. The institution of a clean intermittent catheterization program before 1 year of age resulted in fewer patients requiring bladder augmentation to correct lost of bladder compliance in one short-term study.[292] Total continence is not achieved in most

* See references 18, 25, 26, 30, 102, 104, 106, 182, 225, 233, 273.

† See references 18, 25, 26, 104, 106, 182, 225, 233, 273.

* See references 24, 124, 138, 140, 141, 160, 194, 219, 279.

† See references 24, 124, 138, 140, 141, 160, 219, 279, 292.

adult studies, but a reduced need for pads and preservation of upper urinary tract function still result from clean intermittent catheterization. Patients will also need routine evaluation of the lower urinary tract for evidence of infection, reduced bladder compliance and capacity, and hydronephrosis. Screening examinations consisting of voiding cystometrography and renal ultrasonography performed every 6 to 12 months suffice in most patients. Abnormalities may require more thorough urodynamic investigation.

The surgical treatment of spinal deformities may influence urinary tract management or function. In one study, eight of nine patients who underwent cordectomy with kyphectomy had improved bladder compliance and capacity postoperatively, but the ninth patient had poorer function secondary to the development of bladder spasticity requiring surgery.[131] In another study, six of 16 patients who underwent spinal surgery had urologic problems postoperatively, including one female patient who could no longer self-catheterize because of a change in body posture.[23] Thus, patients undergoing major spinal procedures should have baseline urologic evaluation, with reevaluation postoperatively as necessary.

ORTHOPAEDIC TREATMENT

Goals of Orthopaedic Management. The goal of orthopaedists participating in the care of children with myelomeningocele should be to serve as a partner in the health care team seeking to maximize function and minimize disability and illness. Over time, the specifics of the requirements for achieving that goal will change, based on the child's needs and abilities and changes in neurologic health. One of the major goals of the orthopaedist is to correct or prevent deformities that would otherwise keep the patient from being as comfortable and mobile as possible. Nearly all patients will need orthoses to replace muscle strength and joint stability so that they can stand and walk. Similarly, most children, irrespective of the extent of deformity and paralysis, can be enabled to walk at a young age with a combination of deformity correction, bracing, an upper extremity aid, and instruction. Thus, one of the primary functions of the orthopaedist is to correct foot and hip deformities that prevent the patient from using orthotics to ambulate in childhood. Many patients, especially those with thoracic or upper lumbar level paralysis, will not be able or willing to maintain the same level of independent ambulation as adults, since the extent of bracing and the energy consumption required for community ambulation is too great or onerous for the adult. Patients with myelomeningocele should be prepared for independent, self-sufficient *living*, and they will not be able or willing to devote a very substantial portion of their energy and time solely to walking for its own sake. Excessive emphasis on ambulation over the use of a wheelchair may even adversely affect academic achievement.[84]

The orthopaedic surgeon must monitor spinal balance and deformity in the myelomeningocele patient. There is a very high incidence of both congenital and neurologically related scoliosis and kyphosis, conditions that can jeopardize posture or sitting comfort or increase the likelihood of development of pressure sores.

Finally, the orthopaedic surgeon must assist in monitoring the neurologic status of the growing patient, since hydrocephalus, hydromyelia, or tethered cord syndrome secondary either to diastematomyelia or other anomaly or to scarring at the original level of myelodysplasia, can occur. Any of these conditions can result in subtle deterioration in a patient's intellectual function and upper or lower extremity function.

Physical Examination of the Newborn. The aim of the orthopaedic surgeon examining a newborn with myelomeningocele for the first time should be to identify the level of the paralysis for each extremity and to screen for associated deformities. Sphincter control, the presence of hydrocephalus, and the condition of the myelomeningocele sac are also important to note. Commonly, the orthopaedist is consulted after closure of the sac and shunting for hydrocephalus have been completed. The infant should be examined in a quiet, warm environment to allow the best assessment of joint range of motion, sensory preservation, and evidence of spinal deformity. A stimulated or crying infant, however, allows the examining surgeon to better appreciate the child's voluntary lower extremity muscular function. Sharrard has described the neurosegmental function of the lower extremity,[247,248] and this root-by-root assessment has become the mainstay of the description of preservation of lower extremity function and the basis for establishing a prognosis for long-term ambulation and the nature of secondary deformities likely to develop during childhood. The level of spinal cord lesion as visualized on prenatal ultrasonography has been positively correlated with the level of postnatal paralysis noted on physical examination, so that this information, if available, may be helpful.[51] Caution is advised, however, in that at times, determining the precise level of function can be difficult, and the level may change over time, may be asymmetric, or may not correspond exactly to the neurosegmental scheme as presented by Sharrard.[168,176,177] McDonald and colleagues found in a longitudinal, serial evaluation of 308 patients over the age of 5 that quadriceps strength correlated with iliopsoas strength, medial hamstring function could be present without tibialis anterior function, gluteus medius and maximus strength correlated strongly with each other and with tibialis anterior strength, and muscle weakness was most frequently noted in the gastrocnemius-soleus group.[177]

During the examination, the orthopaedist should first develop a sense of the child's overall vigor, as lack of vigor may suggest CNS depression, such as might result from untreated hydrocephalus. Whenever expedient, the examiner should turn the infant prone on the mother's lap, an examining surface, or the palm of the examiner's hand. The task now is to determine at what level the myelodysplastic deformity is located, how extensive it is, and the state of the skin overlying it, especially if the orthopaedist is conducting the examination after neurosurgical closure of the myelodysplastic lesion. The infant should also be assessed for obvious spinal deformities of congenital scoliosis or kyphosis associated with the myelodysplastic lesion. The examiner now looks for more subtle evidence of spinal dysraphism at other levels, specifically for discoloration or hemangiomas, hairy patches, or dimpling along the spinal column remote from the obvious myelodysplastic lesion. The entire spinal

column is palpated, with the examiner looking for curvature or defect.

With the child supine, neck mobility and upper extremity formation and function are assessed; these are most often normal in the myelomeningocele patient. Next, the examiner visually inspects the posture of the lower extremities, which will give a clue to the extent of the paralysis. For example, the child with a thoracic level lesion will most often lie supine with the legs in a flopped-open, "frog-leg" position with no spontaneous movement. Patients with lower levels of paralysis will exhibit spontaneous movement of the lower extremities; if necessary, the examiner can stimulate the child to observe such movement. Specifically, the examiner should look for hip flexion and adduction, knee extension and flexion, and ankle dorsiflexion and plantar flexion. A note of caution: observed toe movements should not be taken as an indication of volitional control of the digits, as movement of the toes most often results from root sparing below the myelodysplastic lesion and is not under volitional control. The examiner should try to assess the level of preservation of sensation by gently stroking the skin, beginning distally, and observing the infant's facial and lower extremity muscular response. The examiner checks for the usually obvious foot deformities, such as clubfoot and vertical talus. Range of motion of the hips is assessed, with specific noting of abduction, adduction, external rotation, or flexion contractures. The hips are assessed for concentricity and stability as well. Knee flexion contractures and their extent should be documented. The examiner strokes the patient's legs on both sides individually, from distal to proximal, by dermatome, to identify the level of sensory preservation. Finally, the examiner checks for the almost invariably present patulous anus and urinary dribbling suggesting bowel and bladder paralysis.

The physical examination should be supplemented with good AP and lateral radiographs of the entire spine. These radiographs should be carefully inspected for the level of the last closed posterior element, any congenital spinal deformity (particularly one remote from the myelodysplastic level), and pedicular widening, especially with associated congenital vertebral anomalies, which may suggest underlying diastematomyelia. In general, the level of paralysis noted on physical examination should correspond to the first open level of the spine; a substantial discrepancy may suggest that other deformities of the spinal cord or proximal CNS are contributing to the paralysis. Ultrasonography or MRI of the spinal cord may be indicated in such cases. Radiographs of the lower extremities may be obtained, but in general they merely confirm what has been determined from the physical examination.

This examination will give the orthopaedic surgeon a good understanding of the lower extremity anomalies present, the extent of lower extremity paralysis, and the presence of spinal vertebral anomalies that will need to be monitored with growth. The sum of these findings will allow the surgeon to provide the patient's parents with a reasonable general outline of what is required to correct the deformities and what the parents should expect in the future in terms of bracing needs and mobility expectations.

Periodic Assessment. Patients with myelomeningocele require periodic reassessment throughout growth, usually on a semiannual or annual basis. These assessments typically are carried out in a multidisciplinary clinic, which can reduce the number of physician visits families have to make and allow the health care team to provide a comprehensive evaluation and coordinated treatment plan when interventions are required. The ideal clinic with this purpose in mind will consist of an administrative or RN coordinator to function as a patient advocate, coordinating the disciplines that should be evaluating the patient, scheduling ancillary investigations, and securing results of them. This coordinator should also be sure that all of the patient's needs are being addressed over time, which will include educational, vocational, and sexual counseling. Other health care workers who will need to be involved with patient or parents include the orthotist, to provide and repair lower extremity and spinal orthoses; the physical therapist, to aid in lower extremity functional assessment, bracing needs, and instructions in range of motion exercises and mobility; the occupational therapist, to assess upper extremity function and adaptations for activities of daily living and educational modifications as needed; a nurse, to teach the parents and subsequently the child skin care and self-catheterization; a psychologist, to aid the parents in coping with the many challenges and stressors related to the child's disabilities, to ameliorate self-destructive or hostile behavior associated with these disabilities, and to address the low self-esteem and peer adjustment commonly required in patients with visible disabilities and limited mobility; a urologist, to monitor genitourinary function and maximize bladder control; a neurosurgeon, who, after closure of the myelodysplastic lesion and ventriculoperitoneal shunting in infancy, will need to monitor for shunt dysfunction and evidence of tethered cord development; a social worker, to assist the family in identifying financial support for its health care needs and in obtaining educational and vocational counseling; and ideally an experienced neurodevelopmental pediatrician, to oversee the whole process and provide a general assessment of the child's health.

Within this complex and massive generalized screening and treatment program, the orthopaedist should specifically assess at each routine visit that

1. The patient's motor and sensory function have remained stable.
2. The patient's mobility and bracing needs have remained stable.
3. Orthoses and upper extremity aids are appropriate to the patient's requirements, provide the desired effect of maximizing mobility, are in good repair, and are not causing any undue pressure points on the patient's lower extremities.
4. The range of motion of the patient's lower extremity joints is stable and is sufficient to allow the patient maximum mobility based on preserved motor strength.
5. The patient's upper extremity function is stable.
6. Spinal deformity is either stable or not present.
7. The patient's skin is in good condition over the spinal deformity, in the peroneal and ischial areas, at pressure points under orthoses, and over the knees and around the feet, where careless or excessive abuse of the skin may occur with crawling, walking, or swimming without braces or other protection.

These evaluations may be accomplished with the aid of the nurse, pediatrician, therapist, and orthotist. Periodic radiographic assessment of the spine and hips will often be required as well, as only the rare patient will have no evidence whatsoever of either spinal or hip deformity on physical examination.

Multidisciplinary Care. Because the health problems of patients with myelomeningocele encompass so many organ systems, the management of which must be integrated to treat the whole child and provide the families with support as needed, children with myelomeningocele are best assessed and treated in multidisciplinary clinics.* Kinsman and Doehring reviewed the costs and indications for 353 hospital admissions of 99 adults with myelomeningocele admitted over an 11-year period.[126] The average number of admissions per patient was 3.6 and the average length of stay for all admissions was 11.2 days. One hundred sixty-six of the admissions (47 percent) were deemed to be due to potentially preventable secondary conditions such as serious urologic infections, renal calculi, pressure ulcers, and osteomyelitis. This study emphasizes the need for coordinated care of the adult with myelomeningocele and the importance of continuing education of this population in self-care to prevent avoidable health problems. Kaufman and colleagues specifically assessed the impact of the disbanding of a multidisciplinary clinic on the myelomeningocele population.[125] Despite the availability of specialty care in the same area, fully 66 percent of patients did not regularly see a physician, and the authors recorded a serious increase in morbidity in the affected patient population, including amputation and nephrectomy. With the disbanding of the multidisciplinary clinic, neither the patient, nor the family, nor a local physician assumed the duties of coordinator of care.

GENERAL PRINCIPLES OF ORTHOPAEDIC MANAGEMENT OF MYELOMENINGOCELE

Latex Risk. Patients with spina bifida are at risk for the development of a serious allergy to latex.† Contact with latex in sensitized patients may produce local rashes or mucosal irritation. Cardiovascular collapse during major thoracic or abdominal surgery is the most serious manifestation of latex allergy.[10,123,297] Some 10 to 15 percent of studied patients report definite allergy to latex.[166,276] Risk factors for the presence of latex allergy include a history of prior allergic reactions and multiple previous surgeries.[166] Sensitivity to latex may be ascertained by a latex skin prick test or assaying for latex-specific IgE in serum. However, current clinic practice is to perform surgery and other invasive procedures only in a latex-free environment in all patients with myelomeningocele. Performing surgery and other invasive procedures in a latex-free environment can prevent sensitization, and may over time reduce sensitization in patients who were previously sensitized.[53,54] All personnel involved in the management of the myelomeningocele patient, including parents, nursing staff, anesthesiologist, and surgeon, must be cognizant of the risk of latex allergy or inducing it in this patient population. Patients should be operated on and should recover in a latex-free environment.

Infection. Patients with myelomeningocele have a higher rate of complications, including postoperative infections, for almost all orthopaedic surgical procedures than patients undergoing similar procedures who do not have myelomeningocele. The reason for the higher rate is multifactoral, with major factors being bladder paralysis and absence of protective pain perception. The former usually leads to the presence of bacteria in the urinary tract secondary to the bladder paralysis and its management. The diminished pain perception and skin insensitivity lead to more frequent wound breakdown and subsequent infection, either from unrecognized direct compromise of the wound under a cast or from excessive swelling in patients who move, ambulate, or otherwise challenge the operated part in ways that a patient with normal sensation would not.

Pressure Sores. Patients with myelomeningocele invariably have loss of protective sensation of the lower extremities corresponding to the level of the lesion and, even more important, of the buttocks and sacral areas. As a consequence, these patients are prone to the development of pressure sores. These sores may occur on the soles of the feet as a result of walking on bony exostoses or other prominences secondary to deformity or of walking on rough or hot surfaces without adequate protection for the feet. Patients who crawl may get pressure sores on the dorsum of the foot from similar trauma, especially those with paralytic, uncorrected, or recurrent equinovarus deformity, or in the prepatellar area. The medial malleolus is a common site of pressure sores in patients with valgus deformity of the distal tibia using AFOs or KAFOs that do not or cannot adequately accommodate the medial malleolar prominence. Intra-articular fusions of the foot may make the patient more susceptible to pressure sores even when the foot is in a plantigrade position, owing to loss of flexibility of the foot.[165] Thus, triple and subtalar arthrodeses should be avoided in ambulatory myelomeningocele patients whenever possible.

Patients who are primarily sitters are especially prone to pressure sores. These can develop in the sacrococcygeal area or are the ischial tuberosities or the greater trochanters. Patients who cannot or will not stay dry and clean of urine are particularly susceptible to the development of recalcitrant pressure sores, as are patients with pelvic obliquity secondary to asymmetric hip deformity or lumbosacral spinal deformity. Patients whose pelvic obliquity is fixed, such as after spinal fusion to the pelvis, are at relatively higher risk for the development of sores, and the surgeon must be very careful in the early postoperative period especially to guard against this complication. Patients with insensate skin over a kyphotic deformity may also develop sores over the apex of the deformity, either from internal pressure necrosis or from rubbing of the skin against the back of the wheelchair. Many children will require special adaptations to their wheelchair, such as custom-molded back supports or Roho cushions (manufactured by Roho, Inc., 100 N. Florida, Belleville, IL 62221, 800-850-7646) to distribute weightbearing forces and prevent excess skin pressure over bony prominences.

The management of pressure sores involves educating the child and family in prevention, careful postoperative management in at-risk patients, correcting deformities that cause recalcitrant lesions, appropriate brace modifications to prevent the brace from serving as the source of skin

* See references 19, 125, 126, 129, 130, 235, 261.

† See references 1, 10, 53, 54, 123, 166, 276, 296.

breakdown, and, to the degree possible, achieving a bowel and bladder management protocol that keeps particularly the wheelchair-dependent child dry and clean. Patients and their families must be educated to guard against the unprotected skin's contacting rough or hot surfaces, to inspect orthotics and wheelchairs for pressure points, and to shift and relieve body weight regularly while sitting. Good perineal hygiene is a must. Established pressure sores need prompt and aggressive treatment with weight relief and correction of the source of excessive or constant pressure. Pressure sores not treated in this manner can lead not only to extensive soft tissue breakdown and scarring but also to deep, recalcitrant osteomyelitis requiring repeated surgical debridements.

Surgeons placing patients in casts postoperatively must do so with great care and expertise. Cast padding must be evenly and smoothly applied, with bony prominences carefully protected. The casting material must similarly be carefully and evenly applied without any pressure points inadvertently created by fingers indenting the cast or changing the position of the limb after the padding and casting material have been applied. Lower extremity casts should extend beyond the toes but leaving the toes still visible, to protect them when and if the patient crawls or strikes them against some hard surface (Fig. 25–5). Similarly, the surgeon must educate the family to watch for sores developing on the dorsum of the toes in a child permitted weightbearing in a cast. As the plantar surface of the cast softens with ambulation, the toes or dorsum of the foot will be pushed against the dorsal surface or edge of the cast. Any undue swelling, erythema, odor, or unexplained systemic reaction is reason to remove a postoperative cast completely and inspect the surgical wound and limb for evidence of skin breakdown.

Fractures. Patients with myelomeningocele are susceptible to pathologic fractures of the lower extremities. Risk factors include inattention toward or abusive treatment of insensate parts, either by the patient or by caretakers, joint contracture, postsurgical cast immobilization, and higher levels of paralysis.* Newborns with higher levels of paralysis and joint contractures are susceptible to birth fractures.[28] Quan and colleagues assessed bone mineral density of the distal radius in an unselected group of patients with myelomeningocele.[224] They found that all patients had a lower bone mineral density than the normal population, and patients with a history of fracture had densities lower than patients without a history of fracture.

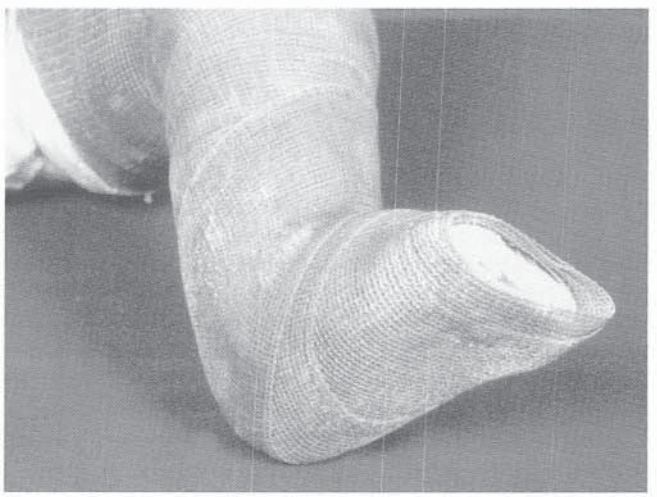


FIGURE 25–5 Proper casting of the feet in patients with myelomeningocele is important to prevent pressure sores. The casts should be well padded and should extend beyond the toes (with the toes visible) to prevent sores at the ends of the toes as the foot is dragged along the ground. The foot should be in a neutral position anatomically.

Several precautions should be taken to prevent fractures in this patient population. Caretakers and ultimately the patient must be educated on safe transfer techniques. Any passive manipulation of joint contractures must be gentle and taught by an experienced therapist or physician. Patients who must be immobilized in a cast postoperatively should, as much as possible, have the affected extremity placed in a functional position (avoiding plantar flexion or excessive knee flexion particularly). Mobilization from the postoperative cast into removable splinting should be undertaken as soon as feasible. Both physician and caretakers should be alert to the development of signs and symptoms of fracture after cast removal.

Fractures will manifest with localized erythema, heat, and swelling. Crepitus and deformity occur only with displaced fractures. The warmth and swelling and the frequent absence of a specific history of trauma will often cause the inexperienced physician or caretaker to suspect infection rather than fracture, and this impression may be fueled by a low-grade fever in the patient. Although hematogeneous osteomyelitis can certainly occur in patients with myelomeningocele, in the absence of direct contamination of the bone by longstanding or extensive pressure sore or surgical intervention, the diagnosis is almost always fracture in this clinical scenario. Fractures in patients with myelomeningocele tend to heal rapidly, with abundant callus. The surgeon should not be unduly alarmed by the extensive callus (Fig. 25–6). Fractures do not, however, invariably heal without incident, as malunion, delayed union, and physeal growth disturbance have all been reported.[71,77,128,287] Therefore, adequate maintenance of alignment and immobilization is required. Physeal fractures may be slow to heal, and require reevaluation to detect subsequent growth disturbance.[57,128,217,287]

Immobilization of the limb, whether after fracture or postoperatively, should be of minimal extent and duration and, as much as possible, in a position of function. Protective orthotics should be available when the cast is removed, and cautious range-of-motion and weightbearing exercises begun under supervision. Failure to follow these principles can lead to a prolonged and frustrating clinical sequence of mobilization after fracture or surgery → juxta-articular fracture → immobilization → increased osteopenia and joint contracture → mobilization → repeat fracture.

MANAGEMENT OF SPECIFIC PROBLEMS

Foot/Ankle Problems. Both congenital and developmental foot deformities are very commonly encountered by the surgeon in the management of the child with myelomeningocele.[33,90,244] Frawley and colleagues reported foot deformity in 263 of 348 feet in 174 patients with lower lumbar or

* See references 28, 57, 71, 77, 118, 128, 146, 217, 287.

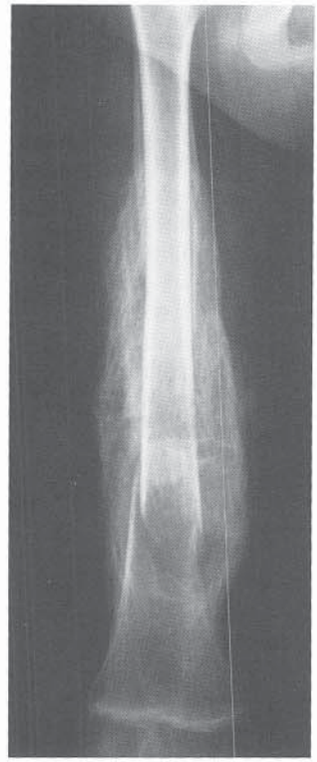


FIGURE 25–6 Fractures in spina bifida frequently manifest with asymptomatic swelling and erythema. Radiographically, there is typically exuberant new bone formation from excessive movement secondary to the lack of pain.

sacral level myelomeningocele, 64 percent of which required surgery.[90] Calcaneus deformity was most common, followed by equinus, valgus deformity, clubfoot, and vertical talus. Foot deformities will very quickly interfere with effective bracing for ambulation or lead to pressure sores in the ambulatory patient. Broughton and colleagues found that almost 90 percent of the feet in 124 children with thoracic or upper lumbar level myelomeningocele had deformity.[33] Equinus deformity was the most frequent, followed by calcaneus, valgus deformity, clubfoot, and vertical talus. The acquired deformities could not be accounted for solely by spasticity or muscle imbalance. Even the nonambulatory patient will have concerns with the cosmetic appearance of the feet and difficulty wearing normal shoes, and in Broughton's series, 78 percent of feet underwent corrective procedures.[33]

In general, foot deformities in the infant should undergo a trial of gentle passive manipulation. This trial must be very judicious, as pressure sores or fractures may otherwise result. Even with early correction, recurrence is common, and surgery is ultimately needed on most feet. Deformities that may respond to passive manipulation alone include equinus contractures, mild or positional talipes equinovarus, and calcaneal positional deformity. Only the true clubfoot deformity that is not completely rigid should be casted between passive manipulations, and then only with great caution by an experienced physician, since pressure sores are likely otherwise.[244] Even rigid clubfeet should be treated by gentle passive manipulation on a frequent daily basis by the family as instructed by the physician or therapist when casting is not feasible. Surgical correction, when required—and it almost invariably is for clubfoot and vertical talus deformities—should be delayed until the child is developmentally ready to be in the upright position. All major foot deformities in children with myelomeningocele have a high frequency of recurrence. Early recurrence of the deformity can be minimized by the surgeon's ensuring that after the removal of postoperative casts, well-fitting orthoses are immediately available for application, and that the child is capable of and encouraged to stand or walk in them. Proceeding with surgery for foot deformity before the patient is ready to be fitted with orthoses for standing or walking will result in incurring a needless risk of recurrence before the child even begins to walk.

EQUINUS. Pure equinus contractures in patients with myelomeningocele are common[33,90,244] but are not due to voluntary muscle imbalance, since the majority of patients have either flail feet or, in low lumbar level patients, tibialis anterior functioning. Positioning deformity, in utero or postnatally, and gastrocsoleus spasticity account for some of the equinus contractures seen, and in some patients equinus will develop after tibialis anterior tendon transfer to the calcaneus (see discussion under Calcaneus Deformity, below).

Patients with positional neonatal equinus contractures associated with higher-level paralysis can initially be treated with *very* gentle passive manipulation. If the equinus deformity persists when the child is ready for initial orthoses for standing and ambulation, percutaneous or open lengthening of the heel cord may be carried out. Percutaneous heel cord lengthening can be performed in the outpatient clinic if the patient is insensate in this area. Some patients will have long toe flexor contractures as well, which should also be divided. Otherwise, persistent toe flexion deformities can result in pressure sores on the ends of the toes when the child is placed in shoes. Careful postoperative casting for a few weeks should be followed by fitting of the orthoses required and standing or ambulation.

EQUINOVARUS. Clubfoot deformity is a common associated anomaly in patients with myelomeningocele, irrespective of the level of myelodysplasia.* This deformity in myelomeningocele patients, however, is truly teratologic in that the deformity is nearly always more rigid, with less propensity to respond to conservative treatment; requires extensive surgery to correct; and is more likely to recur, even after excellent correction combined with resection of the tendons that would presumably be the source of recurrence.†

Patients with myelomeningocele and clubfoot deformity can be managed as other patients with idiopathic clubfoot deformity (see discussion under Congenital Talipes Equinovarus (Clubfoot) in Chapter 22, Disorders of the Foot). However, the treating physician should be very experienced and comfortable with both manipulation and casting techniques, insofar as absence of the pain response or of protective sensation makes it very difficult to avoid pressure sores or fractures. This problem is compounded by the relatively poor response to these methods and resistance of the deformity to correction by these methods. Casting and manipulation must be stopped at least temporarily if swelling or skin necrosis develops. Most often, such treatment will not

* See references 15, 33, 59, 65, 90, 238, 244, 250, 283.

† See references 15, 33, 59, 65, 90, 165, 238, 244, 250.

prevent the patient from requiring surgical correction, and some surgeons defer all treatment until the time of surgical release. Surgery should be delayed until the patient has developed to the point of being ready for brace fitting and ambulation or standing. Crawling with the feet dragged behind and rotated internally, as is typical of infants with myelomeningocele, creates a deforming force that promotes recurrence. Surgery should not be performed prior to this neurodevelopmental point, since preventing recurrence of the deformity depends at least partly on adequate brace fitting and weightbearing.

The surgical correction of clubfeet does not differ from correction in patients with idiopathic deformity. However, extensive release is usually required, and the surgeon will very often find that ancillary procedures such as lateral column shortening will be required much more frequently than in patients with idiopathic clubfeet. In patients with lower lumbar lesions, the tibialis anterior may be lengthened or transferred to either the midline or the heel (see discussion under Calcaneus Deformity, below). In patients with upper lumbar level lesions or higher, tendons are resected rather than lengthened. Only in patients with virtually complete preservation of lower extremity function is typical tendon lengthening performed rather than resection. Furthermore, much as in patients with arthrogryposis, the surgeon may find that myelomeningocele clubfeet may require naviculectomy, talectomy,[65,238,250,251] or talar enucleation (Verebelyi-Ogston procedure)[91] to effect correction. In the most severe deformities, vascular compromise in bringing the foot into a corrected position has required the author to combine clubfoot correction with tibial and fibular shortening. Difficulty with wound closure is common, and rotational flaps have been described to provide primary closure of the surgical incision.[283] We have found, however, that the most effective management is to leave the wound open as much as necessary with the foot in the corrected position (provided the circulatory status is not impaired in the corrected position) and to change the cast or window it as necessary for dressing changes. An alternative is to close the skin loosely and bring the foot into a corrected position with a few cast changes in the first 2 weeks after surgery. We do not favor the latter technique in patients with myelomeningocele, as typically these patients will fling their casts off with movement of the limbs, forcing the foot into more equinus in the slipping cast. This in turn frequently leads to pressure sores of the heel and dorsum of the foot, to further add to the surgeon's aggravations. Inadequate skin coverage for surgical correction of recurrent clubfoot deformities has also been addressed with the preoperative use of soft tissue expanders, but with only limited success (two of seven cases).[15]

Postoperative casting must be meticulous, as described above. Excessive swelling, erythema, or systemic reaction must be investigated by removing the cast and inspecting the foot. Wound necrosis and pressure sores are frequent even in the most experienced and attentive of hands, and the families should be so warned. Bracing of at least the foot will be required indefinitely after cast removal, so the surgeon should be sure that the required orthoses are ready to be applied when the cast is removed. The postoperative casting protocol may be shortened compared to that in patients with idiopathic clubfeet, in favor of the braces.

De Carvalho Neto and colleagues[59] reported that radical posteromedial and lateral release produced an overall good or fair result in 77 percent of 63 feet, but Sharrard and Grosfield[244] noted a 22 percent reoperation rate for patients with myelomeningocele and clubfeet. Recurrence of deformity may be treated by primarily bony procedures, including the Verebelyi-Ogston procedure, talectomy, or triple arthrodesis. Talectomy usually corrects hindfoot deformity[65,238,250] but does not address forefoot deformity.[65,238] On long-term follow-up, weightbearing forces are not evenly distributed on the sole of the foot after talectomy, predisposing to the development of neurotropic ulcers.[251] Similarly, triple arthrodesis, even when the foot is clinically plantigrade, predisposes to the development of pressure sores on the foot in the myelomeningocele patient.[165]

A variation of equinovarus deformity may be seen in patients with lower lumbar level paralysis with a functioning tibialis anterior. In some of these patients, a deformity consisting of primarily forefoot dorsiflexion and supination gives the surgeon the impression of either a deformity due solely to the unopposed action of the tibialis anterior, or a very mild clubfoot. As a warning, in infants with such a deformity, posterior or lateral transfer of the tibialis anterior alone often will not correct all components of the deformity, and frequently a limited posteromedial release is required to fully correct the deformity, in combination with a lateral or posterior transfer of the tibialis anterior.

CALCANEUS DEFORMITY. Calcaneus deformity may be seen as a birth contracture or as a delayed deformity secondary to the unopposed action of the tibialis anterior in patients with lower lumbar level paralysis.* Not all developmental calcaneal deformities can be explained solely on the basis of muscle imbalance or spasticity.[33,90] Calcaneus deformity of the foot may make orthotic fitting more difficult and less effective, and predisposes the patient to the development of a neurotrophic heel ulcer. The latter can be very difficult to eradicate and may progress to a recalcitrant calcaneal osteomyelitis (Fig. 25–7). Patients with progressive calcaneal deformity or exhibiting a propensity toward ulcer development should be aggressively managed to prevent this from occurring. In one study, a delay in surgical treatment of this deformity resulted in a tenfold increase in the prevalence of calcaneal ulcers, from 3 percent to 30 percent of ambulatory patients with this deformity.[86]

Patients with mild calcaneal deformities from birth (with the possible exception of those with tibialis anterior–sparing involvement) may respond to gentle passive stretching of the foot into plantar flexion and splinting of the foot in a neutral weightbearing position by means of a small cloth or bandage wrapped around the foot. Patients with persistent or progressive calcaneal deformities associated with unopposed tibialis anterior function will frequently require anterior release of the deformity, usually combined with posterior transfer of the tibialis anterior to the calcaneus to facilitate brace fitting and prevent the development of calcaneal plantar ulcers.† This procedure is described in Plate 25–1. A few important points should be made with reference to this surgery. First, in patients with low lumbar level le-

* See references 13, 21, 33, 86, 90, 95, 227, 244, 278.

† See references 13, 21, 86, 95, 227, 244, 278.

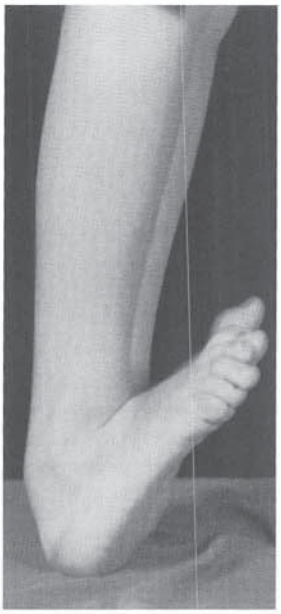


FIGURE 25–7 Calcaneal foot deformity in a patient with a low lumbar level myelomeningocele. In addition to making bracing difficult, this deformity places the patient at risk for the development of neurotrophic ulceration of the heel and calcaneal osteomyelitis.

sions, a posterior transfer that is performed in the hope of providing enough ankle stability to make braces unnecessary will usually not succeed, because this transfer is not of sufficient power to do so; continued ankle-foot orthotics will be necessary for the patient to achieve maximum mobility.[265] Second, the transfer should be positioned with the foot in a neutral position, and postoperatively the foot should be immobilized in a neutral position in a cast, *not* in equinus. If the foot is positioned in a plantar flexed position, the patient may sustain a distal tibial metaphyseal fracture after cast removal when the foot is dorsiflexed with weightbearing. Finally, excessive tightening of the transfer in equinus may result in the development of an equinus deformity that will require release, particularly when the transferred tibialis anterior is not under volitional control.

VERTICAL TALUS. Congenital vertical talus occurs with greater frequency in patients with myelomeningocele than in the general population, although it is much less common than clubfoot and other deformities of the foot.[33,90,244] The rigid planovalgus position of the foot, the prominence of the talar head medially, and the insensate skin combine to make satisfactory fitting of these feet with orthotics very difficult. Thus, patients with this deformity will usually require surgical correction. Surgical management is as for any other patient with congenital vertical talus (see the discussion under Vertical Talus in Chapter 22, Disorders of the Foot). The principles of timing and postoperative management raised in the discussion of managing clubfoot in patients with myelomeningocele apply here as well. Specifically, surgery should be delayed until the patient is neurodevelopmentally ready for orthotic fitting and ambulation, postoperative casting must be meticulous and hold the foot in a neutral functional position, and bracing and weightbearing should be instituted as soon as the casts are removed. In contradistinction to the management of clubfeet, however, there is no role for passive stretching or casting.

VALGUS DEFORMITY OF THE FOOT AND ANKLE. Valgus deformity at the ankle is a common deformity in ambulatory patients with myelomeningocele, irrespective of the level of paralysis.* The deformity may arise from the distal tibia, the subtalar joint, or both,[159] and may be compounded by an external rotation deformity of the tibia. The most common sequela of this deformity is skin irritation or breakdown over the medial malleolus from excessive pressure against the orthosis (Fig. 25–8). Important considerations for the orthopaedic surgeon are (1) determining the precise location of the clinical valgus (ankle or subtalar); (2) ascertaining whether the patient is skeletally mature or, if immature, approximately how much growth remains in the distal tibia; and (3) deciding whether the extent of deformity requires immediate correction because it is unbraceable or whether more gradual methods of correction can be used because the deformity is progressive but still braceable. Thus, assessment by the physician will require physical examination of the patient, consultation with the orthotist regarding the interim management of medial malleolar pressure areas, and AP radiographs of the ankle to determine the source of the valgus and the state of the physis. In skeletally immature patients, a scanogram and radiographs of the hand and wrist for estimation of bone age may be necessary to assess how much growth remains in the distal tibia, if distal tibial epiphysiodesis techniques are being considered.

Ankle. Surgical options for the management of distal tibial valgus deformities include distal tibial and fibular osteotomy,[2,159,245] distal tibial medial hemiepiphysiodesis or stapling,[37,159] and Achilles tendon–distal fibular tenodesis.[263] A distal tibial valgus deformity which causes pressure sores that cannot be corrected by adjustment of orthotics or occurring in skeletally mature patients will require a distal tibial osteotomy and varus realignment (Fig. 25–9). Skeletally immature patients with deformities that are progressive but not in need of immediate correction are candidates for either medial tibial hemiepiphysiodesis or Achilles tendon–fibular tenodesis. The medial growth arrest may be effected by direct curettage, stapling of the medial side of the distal tibia, or insertion of a screw percutaneously from the medial malleolus proximally across the physis. Fibular-Achilles tenodesis is indicated in young patients with milder distal tibial valgus deformities who are considered too young for an epiphysiodesis of the distal tibia.

Distal tibial osteotomy: Fixation may be done with crossed Steinmann pins, staples, an external fixator, or internal fixation with a dynamic compression plate. This osteotomy may be complicated by delayed union, nonunion, or infection, particularly in adolescents. Recurrence of the deformity is also relatively common in the skeletally immature patient. Postoperatively, patients should be kept nonweightbearing initially, since weightbearing with diminished pain perception can lead to excessive swelling and motion. The patients should also be counseled not to crawl in postoperative casting. If possible, the knees should not be flexed excessively in long-leg casts to avoid rehabilitation difficulties after cast removal.

* See references 2, 6, 11, 33, 37, 90, 159, 244, 245, 263, 289.

Technique of Tibialis Tendon Transfer to the Calcaneus to Prevent or Correct Calcaneus Deformity

This procedure is indicated in patients with unopposed voluntary or involuntary ankle dorsiflexion producing dynamic and potentially fixed calcaneal deformity of the ankle. The patient is most conveniently positioned in the lateral decubitus position with the affected side up, allowing ready access to the anterior and posterior aspect of the ankle and lower leg. Alternatively, especially when the deformity is bilateral, the patient may be positioned either supine (with the leg rotated to allow somewhat awkward access to the heel) or prone (with the knee flexed for the anterior portions of the procedure).

A, The anterior tibial tendon is exposed at its insertion through an incision on the dorsomedial aspect of the foot. The tendon is sharply resected from its insertion on the base of the first metatarsal, and a Bunnell-type suture of heavy absorbable suture material is placed at its distal end. The tendon is mobilized to the retinaculum. A second incision is made over the anterior compartment of the lower leg, exposing the tibialis anterior tendon and retracting it into the proximal incision. If soft tissue contractures of the long toe extensors or ankle capsule prevent plantar flexion of the foot into a neutral position, these structures can be divided through an extension of this proximal incision.

B, With displacement of the tibialis anterior superiorly and retraction of the contents of the anterior compartment laterally extraperiosteally away from the tibia, the interosseous membrane is identified and incised to allow passage of the tibialis anterior tendon through it and into the posterior compartment.

C, The Achilles tendon and superior aspect of the calcaneus are exposed through an incision paralleling the lateral border of the Achilles tendon.

D, The tibialis anterior tendon is brought through the interosseous membrane into the posterior aspect of the leg. A drill hole is made in the calcaneus from its superior surface just anterior and lateral to the insertion of the Achilles tendon.

E, The tibialis anterior tendon is passed through the calcaneus using straight Keith needles and is sutured over a pad and button on the sole of the foot. We prefer that the transferred tendon be sutured with the foot in a neutral position rather than plantar flexion. The anterior tibial tendon can be sutured to the Achilles tendon for additional reinforcement of the transfer. The surgeon must carefully avoid excessive pressure of the pad and button on the heel pad to avoid pressure necrosis in this area.

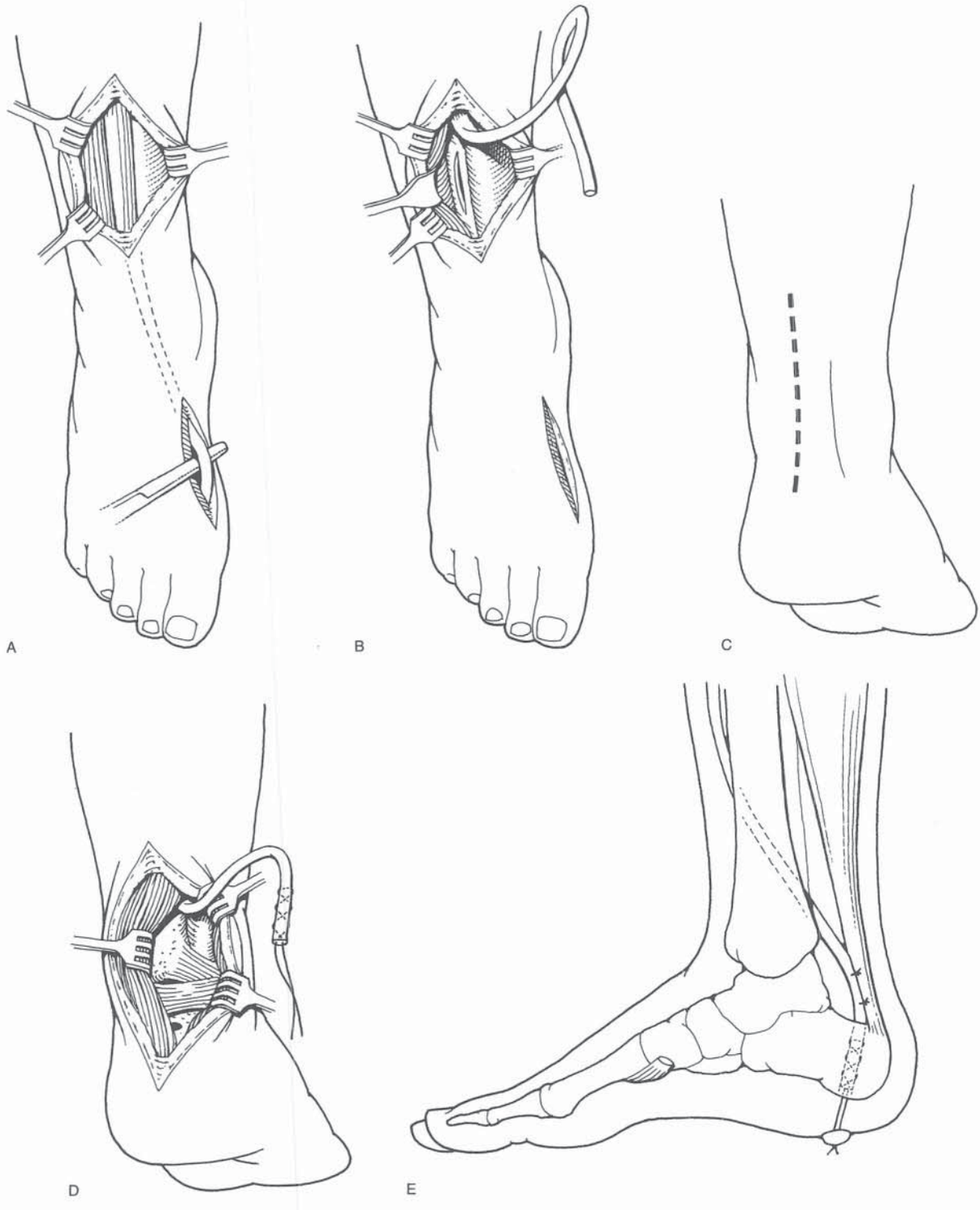
The wounds are irrigated and closed after the tourniquet has been released and hemostasis achieved. A short-leg cast is applied with meticulous attention to prevent wrinkles and pressure points.

POSTOPERATIVE MANAGEMENT

The patient is not permitted to bear weight in the cast while the button is in place. The cast can be removed 2 to 3 weeks after surgery to inspect the wounds and remove the button by transecting the absorbable suture at the skin. We use this clinic visit to cast the patient for new ankle-foot orthoses. A new short-leg walking cast is applied and the patient is allowed to bear weight as tolerated. The parents must be educated to watch for evidence of skin irritation or excoriation at the edges of the cast and the tips of the toes, and to report breakdown of the sole of the cast. The cast is removed 6 weeks after surgery and the patient is placed in AFOs or KAFOs as needed.

Excessive swelling after removal of the cast may be due to a fracture of the distal tibial metaphysis. This complication is most easily avoided by immobilizing the foot and ankle in a neutral position in the cast, rather than in plantar flexion, and by ensuring a smooth transition from cast to orthosis postoperatively.

PLATE 25–1. Technique of Tibialis Tendon Transfer to the Calcaneus to Prevent or Correct Calcaneus Deformity



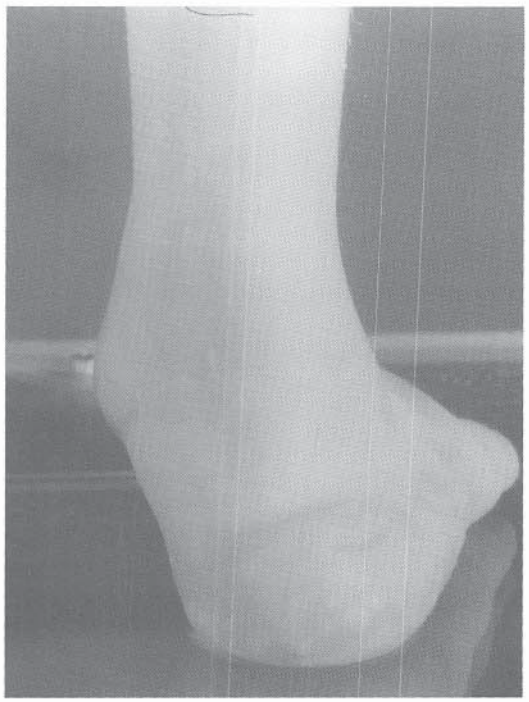
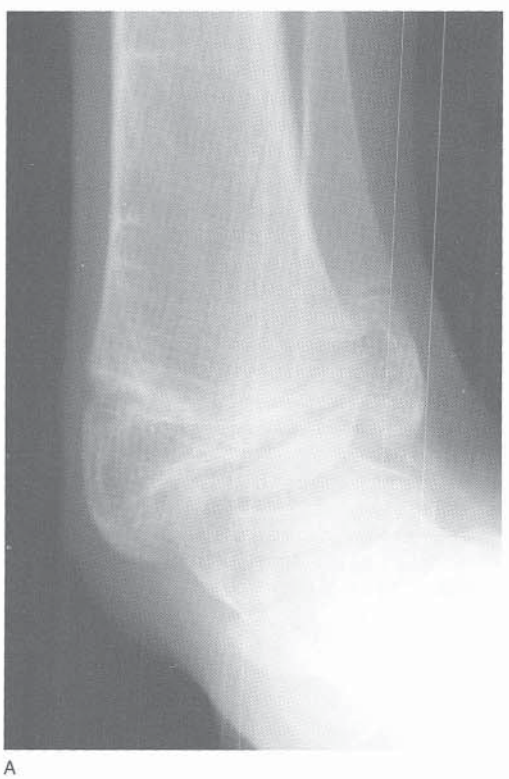


FIGURE 25–8 Valgus deformity at the ankle in myelomeningocele. This deformity may lead to ulceration over the medial malleolus or head of the navicular from rubbing against the AFO component of the patient's orthosis.

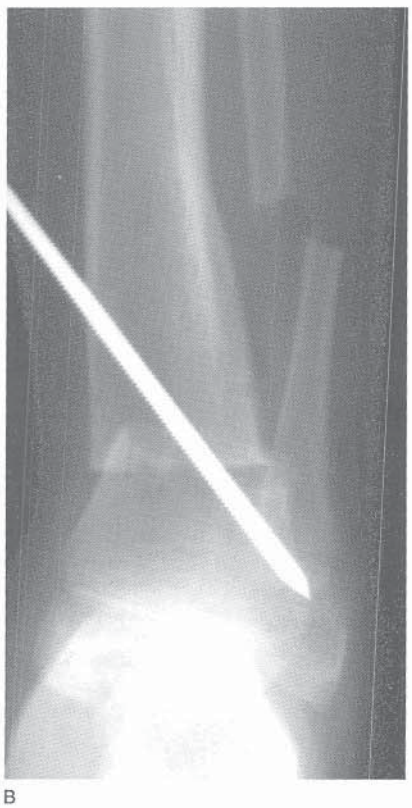
Distal tibial medial hemiepiphysiodesis: If the patient is skeletally immature with a deformity not demanding full and immediate correction, a medial hemiepiphysiodesis can be considered. The medial tibial physis can be closed with direct surgical ablation, stapling,[37] or insertion of a medial malleolar screw.[262] The advantage of this technique is that usually, immediate weightbearing can be allowed and external immobilization is not necessary.

Achilles tendon–fibular tenodesis: Stevens and Toomey have described the surgical technique of tenodesing a portion of the Achilles tendon to the distal fibula above the distal fibular physis.[263] The rationale is that the valgus deformity is secondary to lateral compartment paralysis with subsequent underdevelopment of the fibula, and that this lack of growth stimulation can be compensated for by tenodesis of a slip of the Achilles tendon to the fibula. With weightbearing and ankle dorsiflexion, the tenodesis pulls downward on the fibula, leading to gradual correction of the deformity. The surgical procedure is outlined in Plate 25–2. The procedure, similar to a distal tibial hemiepiphysiodesis, is indicated in skeletally immature patients with a progressive deformity that does not yet require complete correction. It is particularly suited to younger patients in whom hemiepiphysiodesis is not appropriate.

Subtalar Joint. When radiographs reveal that most of the valgus deformity is in the subtalar region, treatment should be with subtalar arthrodesis.[6,11] Aronson and Middleton reported satisfactory results in 20 feet treated by internal screw fixation across the subtalar joint and extra-articular iliac crest bone grafting.[6] The reader is cautioned that, as described



A



B

FIGURE 25–9 Distal tibial osteotomy for ankle valgus. **A,** Preoperative radiographic appearance. **B,** Postoperative radiographic appearance. Note displacement of the distal fragment laterally to prevent excessive prominence of the medial malleolus. The fibular osteotomy should be placed as distally as possible to prevent excessive prominence of the distal fragment on the lateral side of the ankle.

by Maynard and co-workers, fusions in the foot, even when clinically plantigrade, predispose the patient to neurotrophic ulcers of the foot on long-term follow-up.[165] Thus, triple arthrodeses and subtalar fusions should be avoided whenever possible.

Rotational Deformities (Internal/External). Rotational deformities of the lower extremities are frequent in both ambulatory and nonambulatory patients.[64,88,290] In nonambulatory patients and in most ambulatory patients, the problem is largely a cosmetic one. Extreme internal rotation may interfere with ambulation if the child is catching the foot on the contralateral extremity during swing. Internal rotational deformities are usually either dynamic secondary to medial hamstring dominance, or fixed secondary to internal tibial torsion. When dynamic internal rotational deformity is interfering with gait, Dias and colleagues reported good results with transfer of the semitendinosus to the biceps and head of the fibula.[64] We have no experience with this procedure. Internal tibial torsion may be treated by osteotomy of either the proximal or distal tibia and fibula. We prefer to perform tibial osteotomy distally with fixation with staple, crossed Steinmann pins, or a dynamic compression plate.

Marked external rotation, in addition to being cosmetically displeasing to the family or patient, may also indirectly interfere with ambulation by making fitting or function of the AFO component of bracing more difficult. The external rotational deformity places the medial malleolus in the line of progression of the limb and may lead to constant skin breakdown due to rubbing against the AFO in this area. This problem is aggravated if there is valgus deformity of the ankle or hindfoot as well. In addition, the calcaneus-preventing action of the orthotic, particularly ground-reaction orthoses, may be rendered ineffective if the external rotational deformity moves the foot sufficiently out of the line of progression of the patient's gait (Fig. 25–10). An external rotational deformity may come from the hip or the tibia. Treatment, if required, should be by internal rotational osteotomy of the affected segment (usually the tibia).

The surgeon must assess the nature of the patient's gait carefully preoperatively before recommending rotational osteotomy and the extent to which deformity should be corrected. Patients who ambulate with little knee motion—that is, who advance their limbs primarily by hip flexion or adduction with the hip externally rotated (typical of patients with upper lumbar level paralysis)—will have difficulty clearing a foot in swing that points directly in line of progression. Such patients should not have the external rotational deformity corrected if no gait or bracing problems are present. If rotational osteotomy is undertaken, the surgeon should aim to have the angle of foot progression be in a more acceptable position of external rotation. Satisfactory results with distal tibial osteotomy have been reported in 80 to 90 percent of cases.[64,88] However, Fraser and Menelaus also reported significant complications in seven of 21 patients treated by tibial rotational osteotomy, including six with delayed union (averaging 6 months to union), three with wound infections, and one with 6 months of persistent swelling for unknown reasons.[88] This experience parallels our own, and we do not recommend a rotational osteotomy of the tibia in a patient with myelomeningocele without serious consideration of the potential sequelae, weighed against the extent to which the rotational deformity is creating problems for the child.

Knee. The knee is not prone to many congenital anomalies in patients with myelomeningocele and is a surprisingly hardy joint in general in these patients in youth. However, long-term studies in ambulatory patients with low lumbar or sacral level lesions suggest that knee instability with or without pain is present in about 25 percent.[288]

CONGENITAL KNEE FLEXION CONTRACTURE. Patients can be born with flexion contractures of the knee. Flexion contractures of less than 10 degrees will resolve by the time the patient is ready for ambulation, either spontaneously or with judicious passive stretching, even when the patient has no motor function across the knee. Knee flexion deformity may subsequently recur, particularly in patients with higher levels of paralysis.[290]

CONGENITAL KNEE HYPEREXTENSION/DISLOCATION. Congenital knee hyperextension or dislocation may also occur in patients with myelomeningocele, most frequently in patients carried in the full breech position. Simple hyperextension deformity may respond to careful passive stretching and splinting. Congenital knee dislocation will require surgical treatment. This treatment should be performed well before the child reaches walking age, so that the postoperative knee-flexed position can be resolved before orthotics are required for ambulation. In patients with myelomeningocele, treatment of congenital knee dislocation usually results in some extension contracture, persistent hyperextension at the knee, and/or multiplanar instability. The treatment of congenital knee dislocation is discussed further under Congenital Dislocation of the Knee in Chapter 20, Disorders of the Knee.

DEVELOPMENTAL KNEE FLEXION CONTRACTURE. In both ambulatory and nonambulatory patients, knee flexion contractures can develop during growth. The development of such contractures does not appear to correlate with ambulatory status, level of paralysis, or the presence of spasticity.[290,291] Normally, knee flexion deformities of 20 degrees or less are well tolerated in the ambulatory patient, either with or without bracing across the knee; nonambulatory patients will usually tolerate even more flexion contracture without interference in mobility status or transfers. If more deformity is present, careful thought must be given to the patient's ambulation level and the extent to which ambulation is being impeded by the flexion contracture. Contractures that interfere with ambulation or transfers respond well to radical knee flexor release,[63,162] which hold up well over time.

KNEE EXTENSION CONTRACTURE. Another common problem is extension contracture, although it is not as common as one might predict from the number of patients with at least some quadriceps function but no hamstring function. The deformity is most often seen as a consequence of extensive bracing or other immobilization in extension, particularly after repeated fractures of the femur. Extension contractures may also be seen after treatment for flexion contracture or congenital knee dislocation. Extension contractures do not in general provide impediments to bracing or ambulation, but they are difficult for the patient to cope with in the sitting position and can inpede independent sit-to-stand transfers. In ambulatory patients, observation is usually the wisest course. When the extension contracture is problem-

Technique of Distal Fibular–Tendo Achillis Tenodesis for Mild Ankle Valgus in Skeletally Immature Patients

The patient is placed prone on the operating table for easiest exposure of the posterior aspect of the lower leg and heel for this procedure.

A, A long vertical incision is made paralleling the lateral border of the Achilles tendon. Through this incision the Achilles tendon, peroneal tendons, and the posterior aspect of the fibular shaft proximal to its physis are exposed.

B, A distally based slip (approximately 1 cm wide) of Achilles tendon is fashioned. The remaining Achilles tendon is lengthened if necessary.

C, A Bunnell-type suture is passed through the proximal free end of the slip of the Achilles tendon, tubulating the tendon as needed. A trough is made in the posterior distal fibular shaft, and if it is stout enough, drill holes are made proximal to the trough to receive the suture.

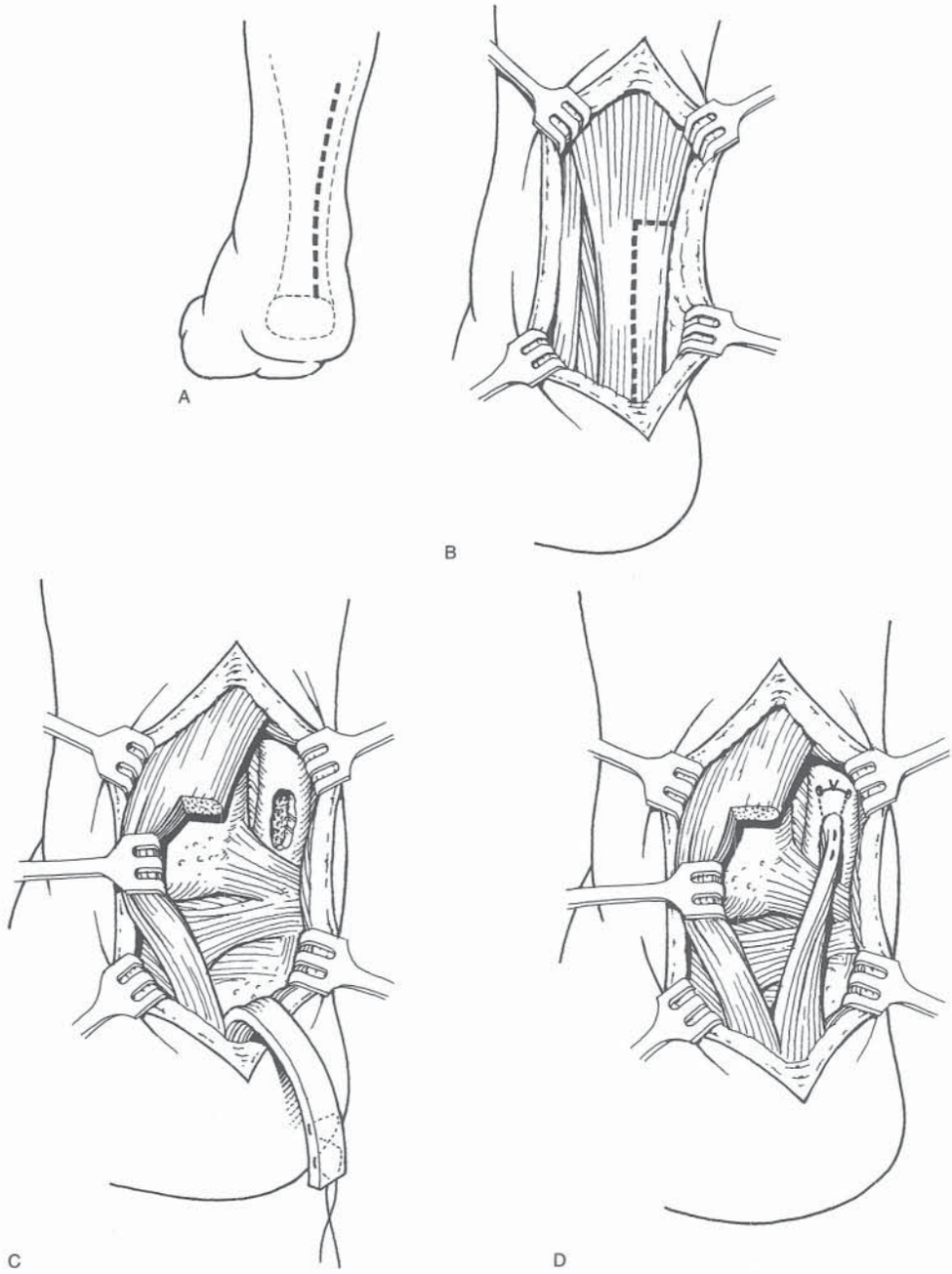
D, The free slip of Achilles tendon is then sutured into the trough in the distal fibula. The transferred portion of the Achilles tendon should be tensioned so as to be snug in a neutral ankle dorsiflexion. If the fibula is too small for holes to be drilled in the cortex, the suture may be passed around the shaft of the fibula, or the slip of Achilles tendon may be wrapped around the fibula and sutured to itself.

The wounds are irrigated and closed. A well-molded and padded short-leg walking cast is applied with the foot in a neutral position.

POSTOPERATIVE MANAGEMENT

The patient is allowed to bear weight in the cast. The child and parents are educated to watch for evidence of skin irritation or excoriation at the edges of the cast and tips of the toes, and to report breakdown of the sole of the cast. The cast is removed 6 weeks after surgery and the patient is placed in AFOs or KAFOs as needed.

PLATE 25–2. Technique of Distal Fibular–Tendo Achillis Tenodesis for Mild Ankle Valgus in Skeletally Immature Patients



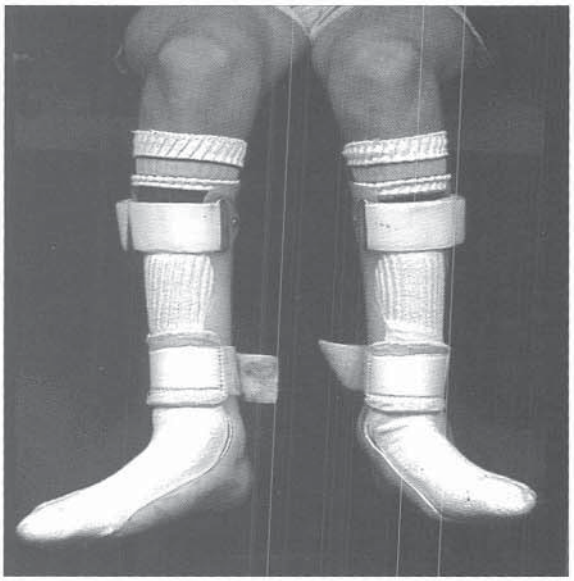


FIGURE 25–10 Extreme external tibial rotation takes the foot out of line of weightbearing, making stabilization of the foot and ankle for weightbearing in a ground-reaction orthosis ineffective.

atic in ambulatory patients, a V-Y quadricepsplasty is effective.[63] In nonambulatory patients, adequate flexion can usually be gained by simple transection of the patellar ligament.[231] Intra-articular release is usually not required. With either procedure, the patient should be immobilized in knee flexion only as long as necessary for the soft tissues to heal. This should be followed by a program of daily gentle passive and (if possible) active range-of-motion exercises of the knee.

KNEE INSTABILITY/INTERNAL DERANGEMENT. Patients with myelomeningocele frequently present with unexplained swelling of the knee. The surgeon must first be sure that there is no infection or intra-articular fracture. When these conditions are excluded, the precise explanation of the effusion may be difficult to determine. Usually the problem is synovial irritation from multiplanar instability or excessive movement of the knee, which frequently develops in the adult patient.[288] Patients who are ambulatory with AFOs likely should be converted to KAFOs, at least temporarily, to protect the knee. The physician and parents should also review the patient's activities and mode of activity, looking for those that might be placing undue stress on the knee (such as incautious transfers or aggressive activities out of orthoses). At least one study found that the use of KAFOs did not provide protective benefit to the knee in patients who were able to ambulate effectively in AFOs alone.[139] Thus, in general, patients who ambulate effectively in AFOs should not be prescribed KAFOs solely in the hope of preventing long-term instability of the knee.

Hip. No part of the orthopaedic management of patients with myelomeningocele is more controversial than the proper management of the hip joint.* Specific deformities encountered by the orthopaedist include abduction/external rotation contractures, hip flexion contractures, developmental dysplasia of the hip present at birth, and progressive paralytic subluxation and dislocation of the hip, usually with attendant hip flexion and adduction contractures. The most frequent and vexing of these problems is paralytic subluxation and dislocation of the hip. The presence of unopposed hip flexor and adductor muscle function in the growing child, as seen in patients with upper lumbar lesions and, to a lesser extent, in those with lower lumbar level lesions, leads almost inevitably to progressive hip subluxation and dislocation. The nature of the problem is perhaps best understood by evaluating the treatment of developmental dysplasia of the hip in patients with myelomeningocele compared to treatment in neuromuscularly intact patients. In the latter, Pavlik harness and closed reductions are the mainstay of treatment in patients before walking age. In patients with myelomeningocele, because of the associated muscle imbalance, such reduction inevitably is followed by recurrence of the dislocation, since there is no spontaneous improvement in the structural abnormalities of femoral and acetabular deformities which contribute to the dislocation. Rather, in myelomeningocele, muscle imbalance of intact flexors and adductors with weak or absent extensors and abductors will drive the hip back into a dislocated position, with accentuation of the structural deformity. Therefore, in patients with myelomeningocele, efforts to reconstruct a dislocated or subluxed hip must follow the principles of all paralytic hip dislocation treatment: obtain a concentric reduction (usually by means of an open reduction); correct the bony abnormality (of femoral anteversion and valgus, and acetabular insufficiency, usually posterior), since there is no propensity for spontaneous correction; and seek to balance the flexor/adductor : extensor/abductor imbalance that created the deformity initially, by transfer or release. Thus, paralytic hip surgery from the outset in myelomeningocele patients will be extensive and will involve transfers or muscle releases, which by their nature will result in *diminished* muscle strength, even though balance may have been achieved. When to this mix is added the higher incidence of complications from any surgery in patients with myelomeningocele, such as hip stiffness, fracture, heterotopic bone formation, against the backdrop of diminishing mobility in these partially paralyzed patients, the battleground of the controversy is clear (although the answers are not!).

ABDUCTION/EXTERNAL ROTATION CONTRACTURE. This deformity is typically seen in patients with thoracic and upper lumbar level lesions, either as a congenital or as a developmental deformity. Occasionally it develops from poor positioning and passive manipulation of the lower extremities, with the affected patient's limbs always allowed to be in a position of flexion, abduction, and external rotation. Initial management consists of gentle passive manipulation that draws the limbs into a position of neutral hip flexion, adduction, and internal rotation. Persistent deformity preventing bracing above the hips that is present when the child is neurodevelopmentally ready for upright positioning should be treated surgically. Release of the tensor fascia lata, rectus femoris, sartorius, and gluteal muscles from the anterior and lateral pelvis will usually achieve neutral positioning of the hips. Postoperatively, the lower limbs are maintained in a neutral position with a removable narrow foam "adduction" splint

* See references 4, 7, 14, 16, 34, 38, 39, 46, 55, 66, 72, 73, 79, 80, 85, 87, 103, 108, 113, 116, 117, 136, 176, 178, 186, 187, 189, 207, 208, 221, 237, 248, 249, 252, 256, 264, 267, 268, 275, 286.

or by wrapping the legs together in Ace bandages. The parents are taught gentle passive manipulation of the hips to maintain the ability to bring the hips into neutral adduction, extension, and internal rotation as soon as the wound has healed adequately. Heterotopic bone formation, hip subluxation or dislocation, femoral fracture, and recurrence of deformity are the most common complications associated with this procedure. Parents and therapists should have a clear understanding that the purpose of such release is for the achievement of upright positioning and ambulation with HKAFOs and upper extremity aids subsequently. Such patients will typically choose the wheelchair as the predominant method of mobility in adolescence and adulthood.

FLEXION DEFORMITY. Pure hip flexion deformity is usually seen in conjunction with hip subluxation or dislocation (discussed below), secondary to involuntary hip flexion or spasticity (often with knee flexion contracture as well), or as a simple contracture due to unopposed preserved hip flexor power. In the ambulatory patient who requires only KAFOs, usually the extent of contracture does not require release or extension osteotomy for its own sake. However, if the patient has a concurrent troublesome knee flexion contracture, greater hip extension may be desirable to aid the postoperative management of such patients. Nonambulatory patients are not usually troubled with hip flexion contracture. Patients who remain good walkers with HKAFOs are the main group requiring treatment of hip flexion contracture. Usually 20 to 30 degrees of contracture can be accommodated by the patient and the orthotist. When deformity is greater than this, hip flexor release (or, rarely, extension osteotomy of the proximal femur) should be carried out. Frawley and colleagues reviewed the results of hip flexor release in 57 hips in 38 children at an average of 9 years after surgery.[89] A good outcome was achieved in 43 hips, in four the contracture recurred after initially good results, and ten had a poor outcome (persistence of flexion contracture of greater than 30 degrees). Six hips required a repeat release, which was successful in two. The success of the procedure could not be correlated with the patient's age or neurologic level, but did correlate with walking ability.

HIP SUBLUXATION/DISLOCATION. The orthopaedist is frequently required to decide the management of paralytic hip subluxation and dislocations. Radiographic deformity is virtually inevitable in patients with preserved hip adductor and flexor power without hip extensor and abductor power. This imbalance characterizes upper and lower lumbar myelomeningocele patients almost by definition. The first question is whether to treat such deformity. Second, if treatment is undertaken, the principles of paralytic hip surgery must be adhered to, that is, the surgeon must obtain reduction, correct bony deformity, and balance the deforming muscle forces to prevent recurrence.

Broughton and colleagues reviewed the natural history of hip deformity in 802 children with myelomeningocele.[34] Hip dislocation had occurred by the age of 11 years in 28 percent of thoracic level patients, in 30 percent of upper lumbar level patients, in 36 percent of patients with L4 functioning, in 7 percent of patients with L5 functioning, and in 1 percent of patients with sacral level lesions. Hip dislocation was not inevitable, even with maximal muscle imbalance about the hip. Hip flexion contractures were much more common in patients with thoracic and upper lumbar level paralysis than in the other groups. Broughton and colleagues thus challenged the concept of muscle balance restoration as a principal aim in the management of the hip in children with myelomeningocele, since the presence and extent of imbalance were variable.

Reduction almost always means anterior open reduction with capsulorrhaphy. Correcting the femoral bony deformity requires a proximal femoral varus osteotomy, often with external rotation to correct the associated femoral anteversion.

Acetabular deformity may be corrected by Pemberton osteotomy, Dega osteotomy, the shelf procedure, the Steel triple innominate osteotomy, or the Chiari osteotomy.[4,29,48,161] Most surgeons believe that a Salter innominate osteotomy is *not* indicated in myelomeningocele patients, since this osteotomy redirects the acetabulum to face posteriorly, which is the direction of hip dislocation initially, and may thus result in recurrent posterior instability.

Muscle balancing procedures include simple release of the iliopsoas tendon with adductor release, posterior transfer of the adductor muscle mass on the ischium to convert it into more of a hip extensor, transfer of the iliopsoas tendon posterolaterally to convert it to a hip abductor (Sharrard procedure), and transfer of the external oblique to the trochanter to recruit a hip abductor from the anterior abdominal wall.

Transfer of the Iliopsoas Tendon in Myelomeningocele. Mustard[208] in 1952 and Sharrard[246] in 1959 described transfers of the iliopsoas tendon to gain abduction and restore muscle balance about the hip. The Mustard procedure entails mobilization of the iliopsoas tendon with the lesser trochanter into the pelvis, rerouting it through a large trough in the ilium, through the gluteal muscles, and reinserting the tendon into the greater trochanter (Fig. 25–11). In a follow-up study of 50 patients (43 had residual poliomyelitis, and only one had myelomeningocele) Mustard reported generally favorable results, although he did note decreased quadriceps strength in one patient, which he thought might be secondary to traction on the femoral nerve intra- or postoperatively.[208] Cruess and Turner reported that the functional results of 13 Mustard procedures in 11 patients with myelomeningocele were very good in none, good in eight, and poor in five.[55] The failures were due to progressive subluxation despite the transfer, and loss of hip flexor strength. Only one patient had a decreased need for bracing and upper extremity aid.

Sharrard's procedure (which he described as a posterior transfer of the iliopsoas tendon,[248] rather than the anterolateral transfer of Mustard) consists of transfer of the iliopsoas muscle through a hole in the ilium underneath the femoral nerve, with reinsertion of the tendon into the posterior greater trochanter through a drill hole and reattachment of the iliacus portion to the outer wall of the ilium (Fig. 25–12). The results of this procedure in the treatment of myelomeningocele hips has received more attention in the literature.* Bunch and Hakala found that all 32 hips in 17 children were reduced at 4- to 14-year follow-up and that all but three of the transfers were active.[36] The radiographic results were improved by combining transfer with varus osteotomy.

* See references 32, 36, 38, 116, 117, 241, 248, 249, 264, 295.

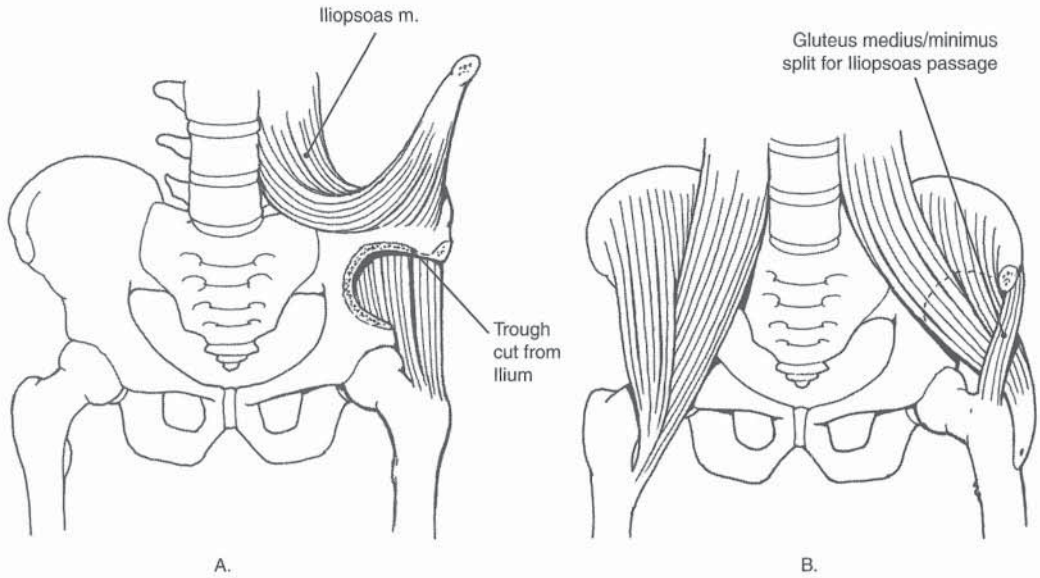


FIGURE 25–11 Mustard transfer of the iliopsoas. **A,** The iliacus muscle is mobilized with a portion of the lesser trochanter, and a trough is made in the ilium. **B,** The muscle is transferred to the greater trochanter through a split in the gluteus medius and minimus.

However, Jackson and colleagues found an overall success rate of only 50 percent in 42 hips in 24 children, and they recommended that the procedure be performed only in children less than 5 years old with a normal acetabular index.[117] Sherk and Ames found on follow-up that 13 patients who had undergone transfer were community ambulators, 13 were household ambulators, and 10 were nonfunctional ambulators or nonambulators.[249] Nineteen of 36 hips lost reduction, and the transferred muscle did not act as a functional abductor in any case. Furthermore, three patients had loss of hip motion, and one required a hip fusion secondary to heterotopic bone formation. Stillwell and Menelaus also found that the power of the transferred muscle was poor; they advised against early surgery and recommended it only in patients ages 2 to 4 years who were likely to walk in short-leg braces as adults.[264]

Transfer of the External Oblique Muscle. An alternative tendon transfer to restore active hip abduction is transfer of the external oblique abdominal muscle to the greater trochanter (Fig. 25–13).[221,295] This technique was described by Thomas and Thompson.[271] Phillips and Lindseth reviewed the results of combined transfers of the adductors, external oblique, and tensor fascia lata muscles in 41 patients with myelomeningocele.[221] Thirty-seven had improved gait pattern (which included increasing the number of patients able to ambulate independently from seven to 21), 17 had a lesser need for bracing, and 21 had a lesser need for assistive devices.

Varus Osteotomy with Tendon Transfer. A variation of this technique includes varus osteotomy (the McKay hip stabilization procedure). Tosi and colleagues reviewed the results of this procedure performed on 66 hips in 34 children with L3 or L4 level of paralysis.[275] The procedure was successful in 37 of 51 hips in children who remained neurologically stable and in three of 15 hips in children who had progressive loss of neurologic function during the follow-up period. The authors recommended performing the procedure only when hip instability was documented. Weisl and colleagues compared posterior iliopsoas transfer with varus osteotomy and adductor and psoas release and found that 19 percent of hips were subluxated or dislocated on follow-up after iliopsoas transfer, compared to 12 percent after varus osteotomy and soft tissue release.[286] However, 80 percent of the transfer patients and 61 percent of the osteotomy patients were wheelchair dependent despite the surgical success of hip stabilization.

Pelvic Osteotomy. Chiari's osteotomy[48] is a favored osteotomy in paralytic dislocations since concentric reduction is not required and the osteotomy does not redirect the acetabulum posteriorly (in the direction of paralytic dislocations) (Fig. 25–14).[4,29,48,161] Mannor and colleagues, however, found on 10-year follow-up of 11 patients treated by 13 Chiari osteotomies that the Chiari osteotomy alone did not produce long-term hip stability in most patients.[161]

Another osteotomy to consider is the Dega osteotomy, as described by Dega and colleagues.[62] With this procedure, the volume of the acetabulum is diminished, and the acetabulum may be redirected anteriorly to some extent by placing larger bone wedges posteriorly. No reports of the Dega osteotomy in the myelomeningocele population have been published in the English literature to date.

Postoperative Hip Stiffness. One of the complications of extensive hip surgery is loss of mobility secondary to heterotopic bone formation. In the nonfunctional or nonambulatory patient, this loss of mobility can create significant morbidity due to loss of the ability to sit comfortably because of loss of adequate hip flexion or fixed pelvic obliquity. Taylor reported that resection of the proximal femur failed in most cases, and he recommended repositional osteotomy (i.e., flexion osteotomy) as an alternative when improved hip positioning is required.[269]

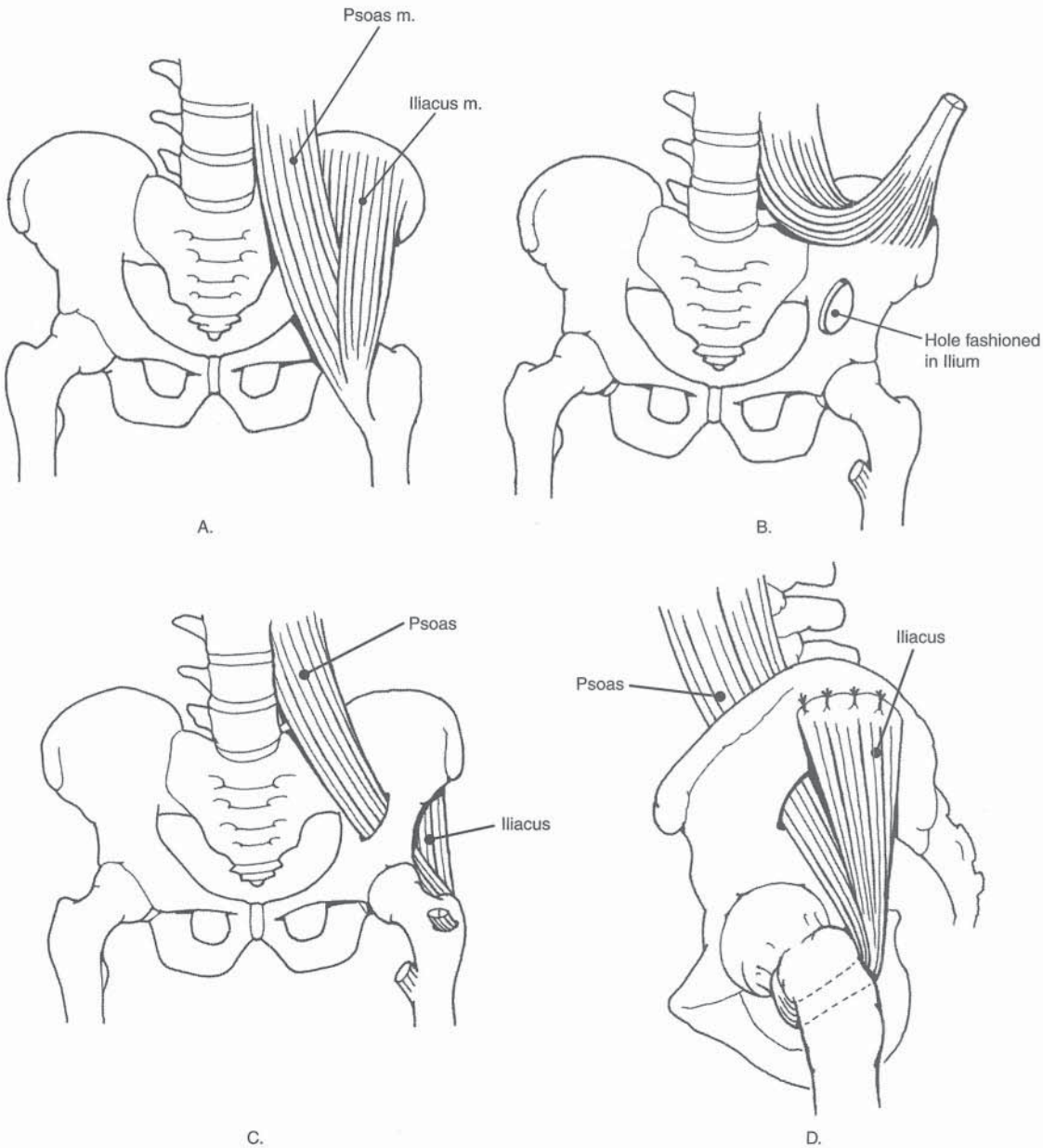


FIGURE 25–12 Sharrard transfer of the iliopsoas. **A,** Normal anatomy of the iliopsoas muscle on the anterior aspect of the hip. **B,** The iliopsoas muscle is mobilized from the lesser trochanter, and a hole is cut in the posterior ilium in preparation for the transfer. **C,** The psoas muscle is transferred through the hole, and the iliacus muscle is transposed to the outer aspect of the ilium. **D,** The iliacus muscle is attached to the outer wing of the ilium, and the iliopsoas tendon is inserted into the greater trochanter via a hole directed posteroanteriorly.

Summary of Management of Paralytic Hip Subluxation and Dislocation in Myelomeningocele. The indications for and the effectiveness of surgical treatment of paralytic hip subluxation and dislocation remain controversial.* Direct application of paralytic surgical techniques relatively successfully employed in patients with poliomyelitis without an adequate understanding of the confounding variables in patients with myelomeningocele has resulted in excessive hip surgery in myelomeningocele patients.[79,80,187] Nevertheless, some patients may gain from the added stability of successful hip surgery in myelomeningocele.[4] One of the problems is that *successful* surgery of the hip may be difficult to accomplish: redislocation, hip stiffness, and loss of active hip flexion are common complications* and may actually make the patient worse.[7,71,252] Unfortunately, a review of the literature does not clarify the indications for surgery, even in the good ambulator. Some authors contend that the patient who is most likely to benefit from hip stabilization procedures has a low lumbar level lesion, is neurologically stable, and has proved to be an excellent ambulator.[4,136,186] Others have found no difference in function between low lumbar level lesion

* See references 4, 34, 38, 39, 55, 72, 79, 80, 85, 87, 89, 103, 161, 187, 221, 249, 252, 264, 275, 286, 295.

* See references 4, 72, 79, 80, 87, 103, 136, 187, 252.

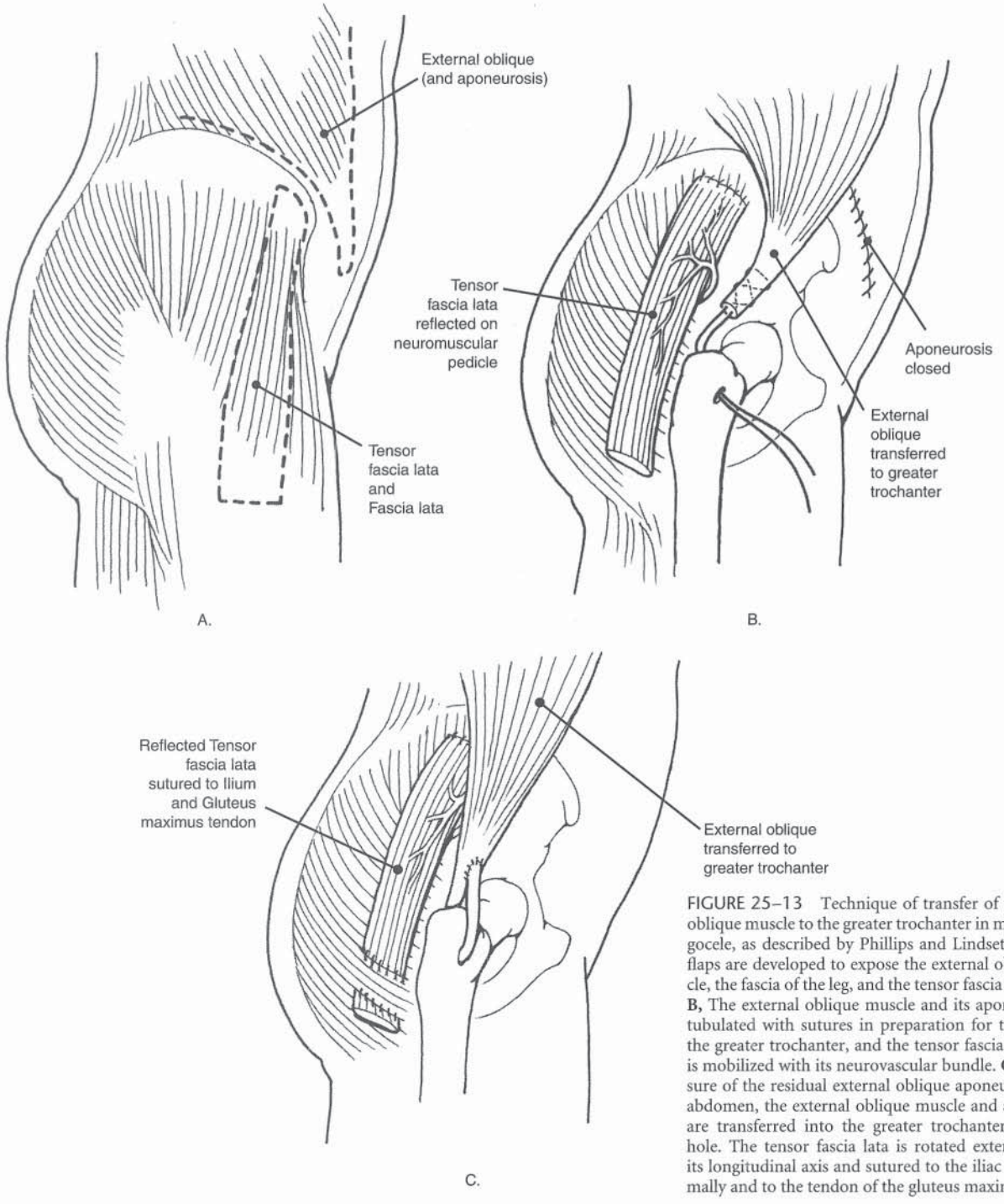


FIGURE 25–13 Technique of transfer of the external oblique muscle to the greater trochanter in myelomeningocele, as described by Phillips and Lindseth.[221] **A,** Skin flaps are developed to expose the external oblique muscle, the fascia of the leg, and the tensor fascia lata muscle. **B,** The external oblique muscle and its aponeurosis are tubulated with sutures in preparation for transfer into the greater trochanter, and the tensor fascia lata muscle is mobilized with its neurovascular bundle. **C,** After closure of the residual external oblique aponeurosis in the abdomen, the external oblique muscle and aponeurosis are transferred into the greater trochanter via a drill hole. The tensor fascia lata is rotated externally along its longitudinal axis and sutured to the iliac crest proximally and to the tendon of the gluteus maximus distally.

patients with dislocated hips and those with surgically stabilized hips.[74,79,80,87,103,252] Some authors believe that unilateral dislocations in the excellent ambulators further sharpen the indication for stabilization of the hip to prevent limb length inequality, pelvic obliquity, and scoliosis.[85] Others have found no such benefit.[74,252] Fortunately, hip pain is unusual in myelomeningocele patients, and thus its "prevention" cannot be used to justify surgical reduction or stabilization of the hip. Areas that are not controversial are (1) the primary goal of the orthopaedic surgeon should be to maintain flexibility in the hip, and (2) patients with thoracic and upper lumbar level lesions do not benefit from hip stabilization. Furthermore, there is rarely, if ever, an indication to perform iliopsoas transfer in patients with myelomeningocele, since those with higher lesions will not benefit and those with lower lesions risk loss of both active and passive hip flexion. The reader should also know that this author did not perform hip stabilizing procedures during 15 years of managing

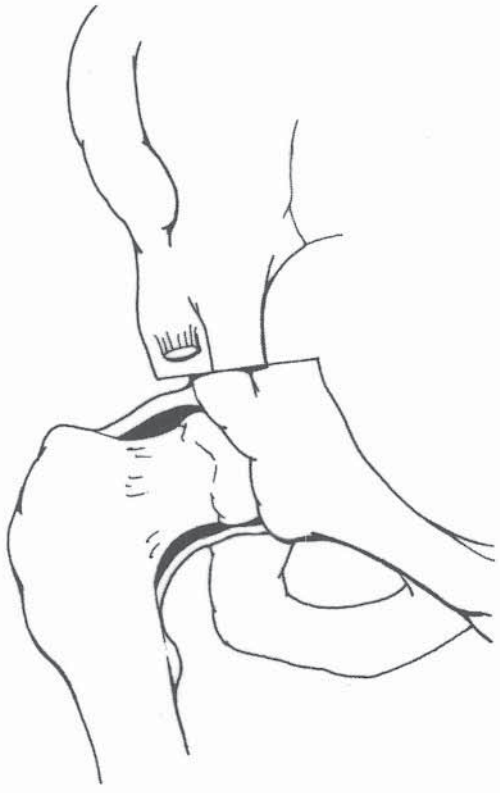


FIGURE 25–14 Chiari osteotomy for paralytic hip dislocation or subluxation in myelomeningocele.

the orthopaedic needs of a large myelomeningocele population, since a clear indication that a particular patient would benefit from such intervention did not arise.

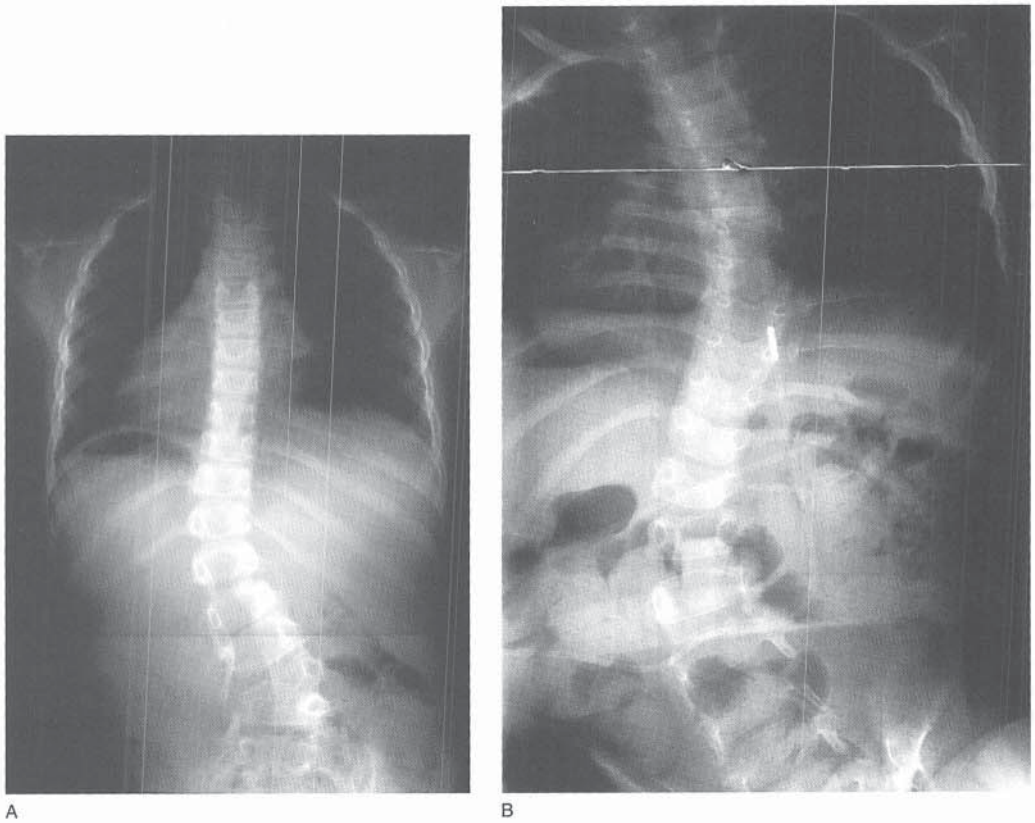
Spine

GENERAL. Spinal deformity in patients with myelomeningocele occurs frequently, can be complex, and often requires treatment.* Deformities can be congenital or acquired, specific to myelomeningocele or similar to deformities seen in other conditions. Congenital spinal anomalies include scoliosis secondary to vertebral malformations, congenital kyphosis related to posterior dysplasia, and intrathecal anomalies such as diastematomyelia. Acquired deformities include idiopathic-like scoliosis, pelvic obliquity–related scoliosis, and neuromuscular curves secondary to spinal muscle asymmetry, hydrocephalus, or tethered cord from any cause (Fig. 25–15). Deformities occur with any level of paralysis and without regard to ambulation ability or history. Problems created by spinal deformity include unstable skin over the deformity in the case of kyphosis, pressure sores or interference with sitting balance in wheelchair-bound patients, and pulmonary compromise secondary to compression from the diaphragm or rib deformity. Although generalities can be made, treatment in each case must be individualized, based on the etiology, severity, and risk of progression of the deformity, the patient's age and ambulatory status, and the impact of the deformity on the patient's well-being.

* See references 8, 9, 12, 23, 40, 41, 49, 68, 75, 76, 78, 92, 101, 107, 112, 120, 142-144, 153, 156, 163, 167, 173, 182-185, 193, 198, 203-206, 213, 222, 225, 234, 242, 257-259, 274, 284, 285.

Patients with myelomeningocele who undergo spinal surgery are particularly likely to sustain peri- and postoperative complications. Experienced and attentive treatment by the health care team is essential. Even under the care of an experienced team, pressure sores, urinary tract infections, wound breakdown, deep infections, pseudarthrosis, and progression of the deformity are much more frequent than in most other patient populations. The treating surgeon must ensure preoperatively that the patient's shunt function is stable, there is no ongoing urinary tract infection, the weightbearing skin of the pelvis and upper thighs is free of pressure sores, and the skin over the portion of the spine to be operated on is healthy. Postoperatively, the wound must be carefully monitored for evidence of infection, and promptly debrided if there is evidence of either superficial or deep infection or tissue necrosis. The patient's urinary tract must be kept clean. The patient's perineal skin must be carefully monitored as the patient resumes sitting: the load of weightbearing will have changed anatomically because of the deformity correction, and there is always some loss of lumbopelvic movement with fixation to the pelvis, which likely increases the pressure applied in the areas that are loadbearing in the sitting patient. Finally, when the patient performs independent transfers with flail or nearly flail extremities, the surgeon must observe these transfers preoperatively to determine whether fixation to the pelvis will tolerate these movements postoperatively. We often allow only one- or two-person assisted transfers for 6 to 8 weeks postoperatively to prevent excessive lumbopelvic movement through the limbs with independent transfers. If necessary, a spinal-thigh orthosis or cast can be used to protect the lumbopelvic junction from excessive movement during this period.

KYPHOSIS. Kyphosis of the lumbar spine is a very common deformity in myelomeningocele patients (Fig. 25–16). Carstens and co-workers found that 20 percent of over 700 myelomeningocele patients had lumbar kyphosis on lateral radiographs;[40] other series report prevalences as high as 46 percent in the myelomeningocele population.[209] Kyphotic deformities have been described as *paralytic, sharp-angled,* and *congenital.* In the review of Carstens and colleagues, paralytic kyphosis (less than 90 degrees at birth) was most common (44 percent of their cases), followed by sharp-angled kyphosis (90 degrees or more at birth; 38 percent).[40] Both kinds of kyphotic curves progressed steadily during growth at a rate of 2 to 5 degrees per year. Progression averaged 4 degrees per year in the series of Doers and colleagues[68] and more than 6 degrees per year in the series of Mintz and colleagues.[198] True congenital kyphosis was the least common (14 percent) in Carstens's series, and progression was variable during growth. Kyphosis in myelomeningocele patients is most commonly seen in patients with thoracic and upper lumbar levels of paralysis; in Carstens's series, the most common level of paralysis was lower thoracic (ranging from upper thoracic to L5). In that series, the apex of the deformity varied from T12 to L5 but was most commonly at L2 (Fig. 25–17). Progressive kyphosis is usually associated with a compensatory thoracic lordosis (Fig. 25–18).[68] In the series of Mintz and colleagues, progressive kyphosis was associated with loss of any prior preserved lower extremity function.[198]



A B

FIGURE 25–15 Patterns of spinal deformity in myelomeningocele. **A,** Congenital scoliosis. Note L4 hemivertebrae. **B,** Paralytic scoliosis.

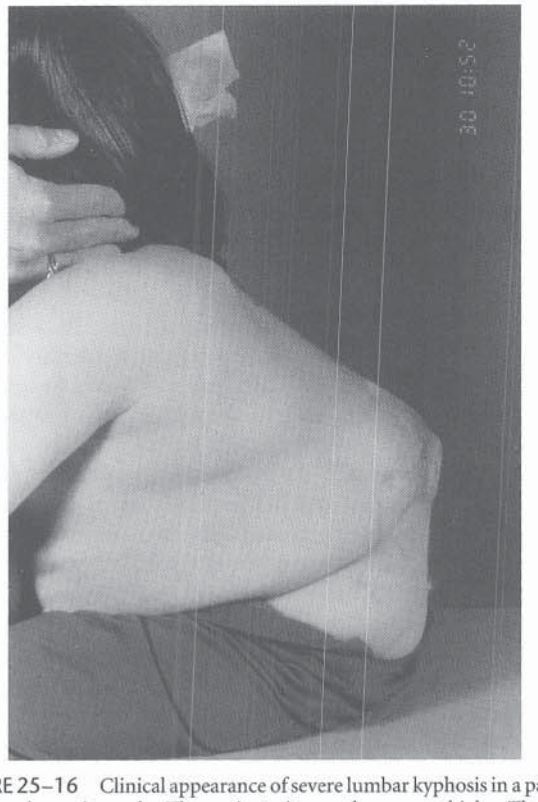


FIGURE 25–16 Clinical appearance of severe lumbar kyphosis in a patient with myelomeningocele. The patient sits on the upper thighs. The skin over the kyphotic area is easily traumatized.

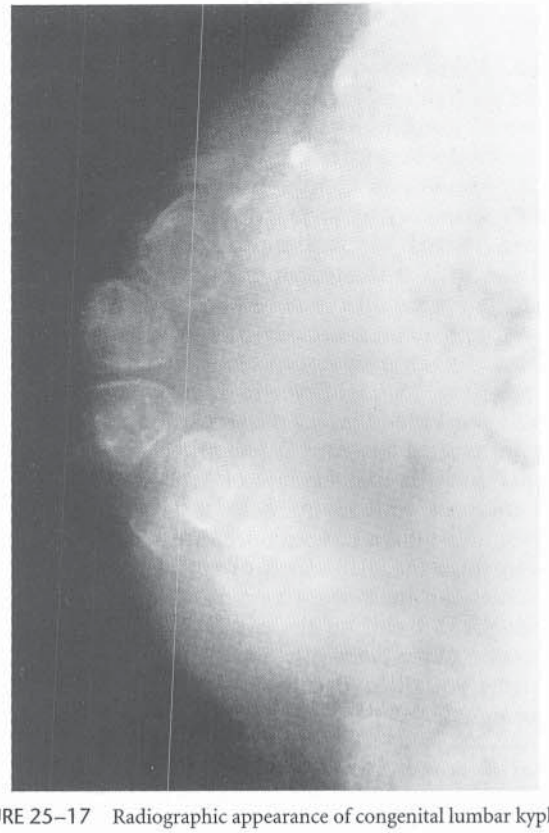


FIGURE 25–17 Radiographic appearance of congenital lumbar kyphosis.

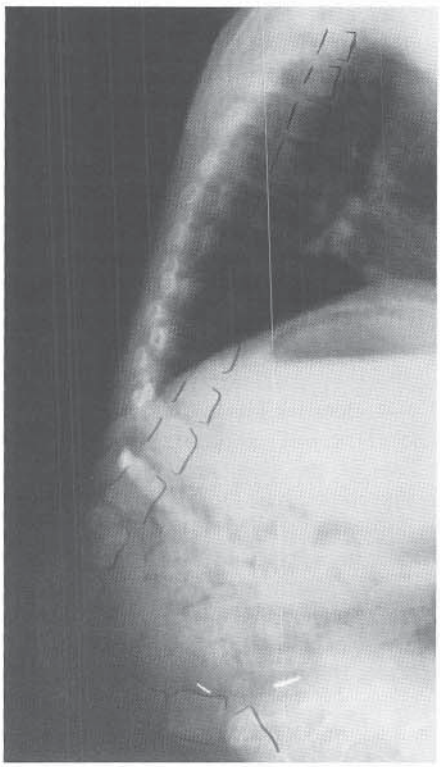


FIGURE 25–18 Patients with lumbar kyphosis typically have a compensatory thoracic lordosis.

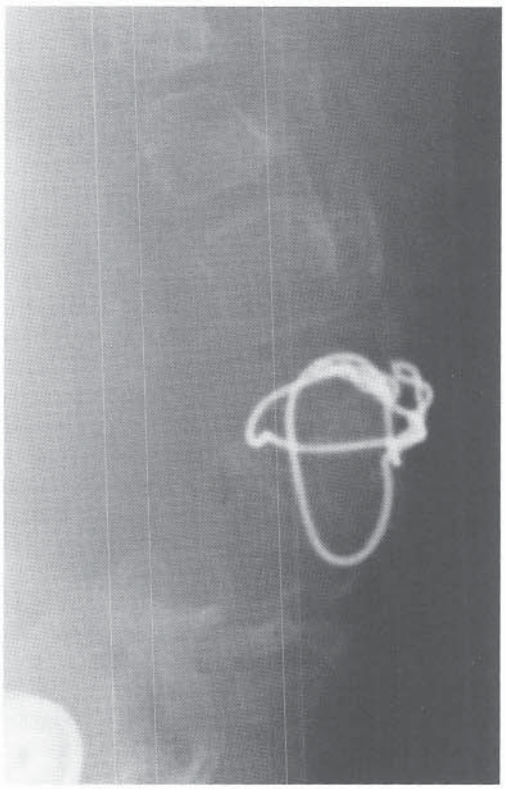


FIGURE 25–20 Congenital kyphosis can be treated by partial vertebrectomy at birth if necessary to close the skin over the myelomeningocele defect. Typically, the deformity will recur with growth, and further procedures may be necessary.

Lumbar kyphosis can be problematic from birth, causing difficulty closing the skin and meningeal defect or later difficulties with skin breakdown with sitting, sitting balance problems, or even pulmonary compromise from pressure on the thoracic cavity from the collapsing abdomen and diaphragm. Chronic skin breakdown can leave the neural elements and the spinal column exposed and at risk for infection (Fig. 25–19).

If the kyphosis creates difficulty with skin closure at birth, one or two largely cartilaginous vertebral bodies can be enucleated and the remaining spine held together with suture or cerclage wire (Fig. 25–20).[49] Unfortunately, partial vertebrectomy in the newborn probably does not prevent the subsequent development of kyphosis.

The treatment of myelomeningocele-related kyphosis is never easy. There appears to be little or no role for bracing to control or correct the deformity. Patients with skin breakdown over a stable kyphosis that does not itself need treatment should first have their wheelchair support and activities carefully evaluated and any irritants found to be causing

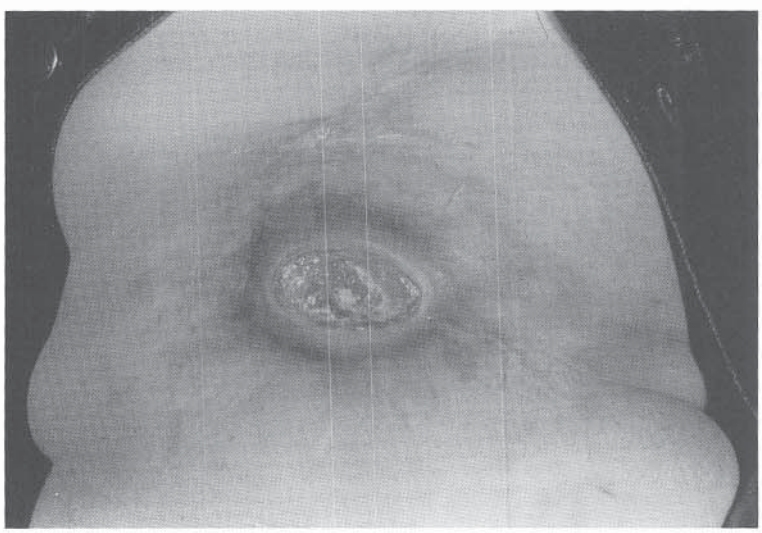


FIGURE 25–19 The consequence of severe kyphotic deformity is often unstable skin prone to repeated breakdown over the kyphotic deformity.

the breakdown removed. If these efforts are unsuccessful, rotational or free flaps may be used to cover the kyphotic area with thicker, more stable skin. Soft tissue expanders have been used for this purpose as well, independently and in conjunction with spinal deformity correction.

The definitive management of kyphosis consists of kyphectomy and posterior spinal fusion and instrumentation.* This is one of the most challenging procedures in orthopaedics. Patients in whom the apex of deformity lies below the level of neurologic function are typically treated by cordectomy, vertebral body resection, and instrumentation. Careful preoperative assessment is necessary. The function of the shunt must be known, and if it is nonfunctioning in a patient who is shunt dependent, it must be replaced before surgical treatment of the kyphosis is carried out. The skin over the kyphosis must be as stable as possible, and if poor, consultation with a plastic surgeon should be obtained to assess methods to improve the skin by tissue expanders or rotational flaps. The patient should be treated for any urinary tract infection preoperatively, and the renal function should be known. The aorta typically bridges the area of kyphosis and thus is not at great risk during vertebrectomy. However, the kidneys are often nestled within the kyphotic area and may be inadvertently injured during surgery.[92,147] Interestingly, cordectomy may result in improved bladder function, as evidenced by increased bladder compliance and capacity. This occurred in eight of nine patients tested by Lalonde and Jarvis;[131] one patient, however, did develop a spastic bladder requiring urologic surgery to correct. Rarely, a patient will have neurologic function below the apex of kyphotic deformity, and in these patients the spinal sac must be carefully protected from injury or devascularization.

Surgical Technique of Kyphectomy. Sharrard in 1968 to 1969 was the first to describe the technique of vertebrectomy in the management of kyphotic deformity, both in the newborn[242] and in the older child.[243] At surgery, the neural elements are dissected away from the posterior spinal elements (Plate 25–3). In patients with no function below the level of resection, the nerve roots and cauda equina remnants may be resected by tying the roots, elevating the distal cord, and transecting it. The meninges should be dissected free of the neural elements, resected distal to the elements, and sutured closed. The spinal cord should *not* be tied, as acute hydrocephalus may result, which in turn can cause sudden death. After resection of the cord, the lumbar spine is dissected extraperiosteally from the posterior approach to the anterior aspect of the vertebral bodies. Two or more vertebral bodies are then resected through their midportion, so that the kyphosis can be reduced. Fixation to the pelvis is then carried out. A number of instrumentation techniques have been described, including fixation with Harrington compression instrumentation, Luque-Galveston instrumentation to the iliac crests, the Dunn-McCarthy modification of Luque instrumentation to the sacrum, vertebral body plates, and figure-of-eight wire loops around the pedicular remnants with immobilization in a cast or brace (Fig. 25–21).† Our preferred technique is to use Dunn-McCarthy rods with posterior pedicular wire loops or vertebral body screws and to place sublaminar wires from the lowest level at which the posterior elements are intact to the T2 or T3 level proximally. In very young patients, postural reduction with the application of a double-pantaloon body cast and prone positioning postoperatively may be used; in such patients, however, we find that there are many problems with pressure sores within the cast or around the kyphectomy site from excessive pressure or gradual recurrence of the deformity.

Results. Martin and co-workers reported improved skin condition and sitting posture in all ten patients treated by vertebrectomy, figure-of-eight wire fixation, and postoperative cast immobilization at an average age of 5 years.[163] Six patients had postoperative lower extremity fracture, two had delayed wound healing, and three had identified pseudarthroses. Two of the pseudarthroses united after repeat surgery, so that nine of the ten patients eventually had solid fusions on follow-up.[163] The average degree of deformity was 90 degrees preoperatively, 40 degrees postoperatively, and 60 degrees at follow-up averaging 5 years.

Warner and Fackler found that eight of 21 patients in whom kyphosis was stabilized with Harrington compression instrumentation but none of 12 patients treated by the Dunn-McCarthy technique and instrumentation had recurrence of kyphosis on follow-up.[285] McCall[173] and Heydemann and Gillespie[107] have also reported improved results in deformity correction using the Dunn modification of segmental fixation. In McCall's series, preoperative deformity averaged 110 degrees, postoperative deformity averaged only 15 degrees, and loss of correction averaged only 5 degrees on follow-up. Eight of 16 patients had complications, and blood loss averaged 1,100 mL.[173]

The reader is cautioned that most authors agree that kyphectomy and instrumentation is a major surgical procedure: intraoperative blood loss is usually well in excess of 1,000 mL, perioperative deaths have occurred, and postoperative complications, including skin breakdown, infection, loss of fixation, and recurrence of deformity, are more frequent than after most other orthopaedic procedures.*

SCOLIOSIS. Scoliosis in patients with myelomeningocele may be congenital, idiopathic-like, or related directly or indirectly to the spinal dysraphism and associated paralysis (associated intrathecal anomalies such as tethered cord, hydromyelia, or diastematomyelia; paralytic pelvic obliquity; asymmetric paralysis). Scoliosis is one of the most common musculoskeletal deformities requiring treatment in patients with myelomeningocele.† Muller and Nordwall found that nearly 70 percent of 131 patients had scoliosis, most by the age of 6.[204] In their series the incidence of paralytic-related scoliosis depended on the level of paralysis, being present in 94 percent of patients with thoracic level lesions and 20 percent of patients with sacral level lesions. Ambulatory status also correlated strongly with the development of scoliosis. Scoliosis was more likely in nonambulatory and limited ambulatory patients. Piggott, in a study of 250 patients with myelomeningocele, found that only 10 percent had no evidence of scoliosis, and anticipated that 50 percent of patients would

* See references 49, 76, 78, 101, 107, 112, 142-144, 156, 163, 173, 242, 257, 274, 285.

† See references 49, 76, 78, 101, 107, 112, 142, 144, 156, 163, 173, 242, 257, 274, 285.

* See references 101, 107, 144, 156, 163, 173, 274, 285.

† See references 9, 12, 41, 142, 167, 182, 183, 203–206, 213, 222, 225, 257, 258, 284.

require surgical correction.[222] Noncongenital scoliosis in young myelomeningocele patients is highly likely to progress. In a longitudinal study by Muller and colleagues, noncongenital scoliosis increased an average of 5 degrees per year.[205] The severity of the curve and the age of the patient were risk factors for progression: curves of more than 40 degrees were much more likely to progress, and curves progressed only slightly after the age of 15.

Spinal orthotics such as the Boston brace have a role in the management of noncongenital scoliosis in patients with myelomeningocele. In a study of 21 Swedish children by Muller and Nordwall, curves in six patients progressed to the point of requiring spinal fusion, but only one patient out of 14 with a curve of less than 45 degrees at the initiation of bracing required fusion during the period of study.[203] One patient did develop a pressure sore. Practically speaking, spinal orthotics such as the Boston brace may be difficult to incorporate into the overall management of the child with myelomeningocele, especially an ambulatory child, because the spinal orthosis may be hot, uncomfortable, and cumbersome. Those patients requiring a pelvic band for lower extremity bracing must have the band accommodate the brace; those who sit exclusively may have pressure problems under the brace, or over the anterior thigh in the sitting position.

Spinal fusion with correction of scoliotic deformity can have a positive effect on pulmonary function in myelomeningocele patients. Carstens and colleagues found in a series of 13 patients that ten had restrictive changes in pulmonary function preoperatively.[41] Despite an anterior procedure in eight, eight patients had improved vital capacity when evaluated an average of 1 year postoperatively, and in six the forced expiratory volume had increased. In another study, Banta and Park found improved pulmonary function in the majority of ten myelomeningocele patients who had undergone anterior spinal procedures, and postulated that this was secondary to improved thoracic mechanics after spinal stabilization.[12]

Extensive spinal fusion, as is necessary to treat progressive noncongenital scoliosis in myelomeningocele, has potential negative impacts on the child's overall mobility. Mazur and colleagues found that whereas sitting balance improved in 70 percent of 27 patients treated by anterior and posterior fusion for paralytic scoliosis, ambulatory ability was adversely affected in 67 percent, unchanged in 33 percent, and improved in none.[167] Muller and colleagues found that only one of 14 patients who had undergone spinal fusion for scoliosis maintained the same level of mobility postoperatively: four lost the ability to pull to a sitting position, seven lost the ability to ambulate with or without braces, three could no longer transfer into or out of a wheelchair independently, and nine who had been able to stand supported at a standing table preoperatively could no longer do so postoperatively.[206] Thus, the decision to perform anterior and posterior fusion, especially to the pelvis, must be carefully weighed against the potential impact on the child's mobility and independence.

Activities of daily living, including self-dressing and self-catheterization, may also be adversely affected by extensive spinal fusion.[23,206] Boemers and colleagues found that 6 of 16 patients had alteration in bladder function after spinal fusion, including loss of ability to self-catheterize.[23] Thus, the wise orthopaedic surgeon will seek the counsel of therapists and the urologist regarding the potential impact of spine fusion on these activities before proceeding with fusion in these patients. Finally, the incidence of pressure sores in sitting weightbearing areas may actually be increased by spinal fusion to the pelvis, irrespective of whether there is residual pelvic obliquity or not; presumably, the loss of flexibility of the lumbar spine and the lumbosacral junction combined with altered locations of weightbearing in the sitting position are the causes of this increased incidence of pressure sores.

GENERAL MANAGEMENT OF THE SPINE IN PATIENTS WITH MYELOMENINGOCELE. Radiographic evaluation of the entire spinal column should be carried out in infants with myelomeningocele. The physician should specifically look for the presence, location, and severity of kyphosis, the last level of posterior element closure, and any evidence of congenital spinal deformity (Fig. 25–22). The latter includes failures of formation or segmentation, as with any congenital spinal anomaly (see discussion under Congenital Scoliosis in Chapter 11, Scoliosis), and pedicular widening or secondary posterior element incompleteness, which may indicate the presence of diastematomyelia. Routine physical examination and periodic radiographic screening for evidence of scoliosis should be performed in all patients with spina bifida, since the prevalence of this deformity is so high.

Congenital spinal deformities are managed as in any other patient: if the deformity is progressive, local anterior and posterior spinal fusion is carried out (see discussion under Congenital Scoliosis in Chapter 11, Scoliosis).

Progressive, noncongenital (and thus presumably neuromuscular) curves are treated according to their severity, evidence of progression, and the patient's skeletal maturity. First, the overall health of the patient's neurologic system should be evaluated, particularly in patients with newly evident or rapidly progressive deformities. Shunt function should be assessed, and the spinal cord evaluated for evidence of tethering, hydromyelia, or diastematomyelia. Curves between 25 and 45 degrees in skeletally immature patients may be considered for total contact orthoses. Bracing in ambulatory patients dependent on extensive lower extremity braces, particularly HKAFOs, can be challenging to parents, child, and physician, but spinal orthoses do seem to at least delay the rate of progression of deformity[20,204] and thus should always be considered in each individual, and probably recommended in patients who are quite young, in whom deferral of spinal fusion is warranted. As for patients with idiopathic scoliosis, spinal fusion should be considered for curves of more than 45 degrees.

Spinal Fusion for Scoliosis in Myelomeningocele. Several aspects of myelomeningocele make considerations regarding surgery for scoliosis unique in this condition. Foremost among these is the presence of a posteriorly deficient spinal column, presenting challenges to both fixation and fusion. Second, because the curves are neuromuscular, the treatment of many curves will entail fusion to the pelvis. Not only does this imply fusing across the posterior deficiency, but fusion to the pelvis may negatively impact mobility and self-care.[23,167,206] There will invariably be significant scarring around the neural elements, and distraction correction in patients with useful lower extremity function must be done

Text continued on page 1288

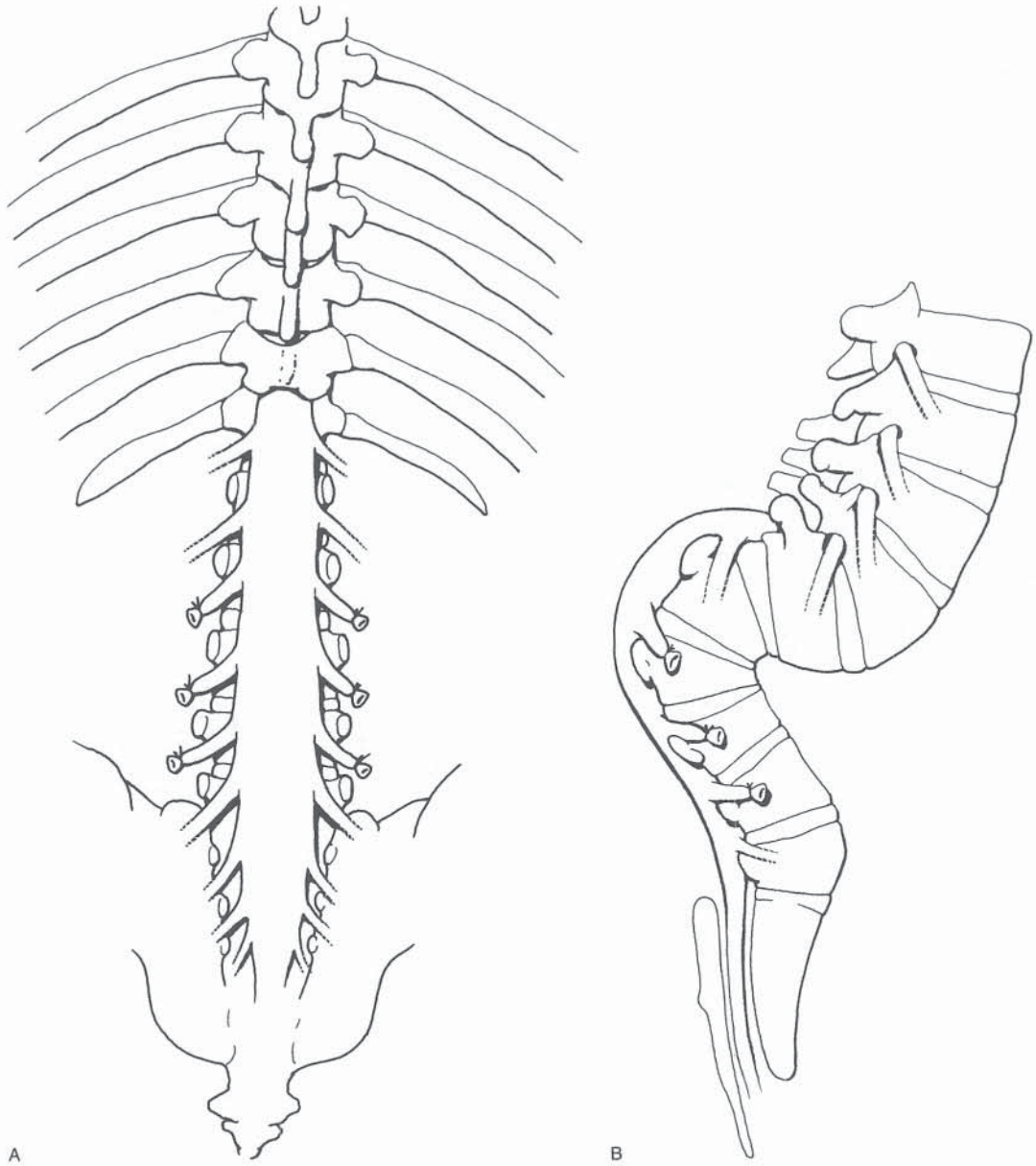
Technique of Lumbar Kyphectomy in Myelomeningocele Patients with Fixation to the Pelvis Using the Dunn-McCarthy Technique

Before this major surgical procedure is undertaken, every effort must be made to provide stable soft tissue coverage of the kyphotic area. The patient's ventriculoperitoneal shunt must be determined to be patent and functioning, or the patient must be known to be shunt-independent. The patient is positioned prone on rolls or a Hall-Relton or similar frame. Meticulous attention must be given to padding bony prominences and avoiding excessive arm abduction. An indwelling Foley catheter, large intravenous lines, and central venous pressure monitoring are essential minimum anesthetic requirements. The entire posterior aspect of the trunk to the buttocks is draped into the surgical area.

A, The posterior skin is mobilized from the myelomeningocele sac remnants. Usually a midline incision is effective for this purpose, but this may be modified, depending on the state of the patient's soft tissues and the need for any concomitant rotational flaps. After mobilization of the skin, the posterior spinal elements are exposed proximal to the dysraphic area. The sac is then mobilized by identifying and freeing it from the spinal column distal to the intended level of kyphectomy. The sac is transected distally and then mobilized proximally by identifying and tying off in sequence the nerve roots as they exit the sac. Because of previous scarring, dissection is usually more tedious than the illustrations might suggest, and the vessels between the sac and the spinal column or associated with the nerve roots can cause considerable bleeding.

B, Mobilization of the sac continues proximally until it can be elevated off the spinal column proximal to the intended level of kyphectomy, usually the point where the posterior elements are intact, or the base of the thoracic spine.

PLATE 25–3. Technique of Lumbar Kyphectomy in Myelomeningocele Patients with Fixation to the Pelvis Using the Dunn-McCarthy Technique



A

B

Technique of Lumbar Kyphectomy in Myelomeningocele Patients with Fixation to the Pelvis Using the Dunn-McCarthy Technique

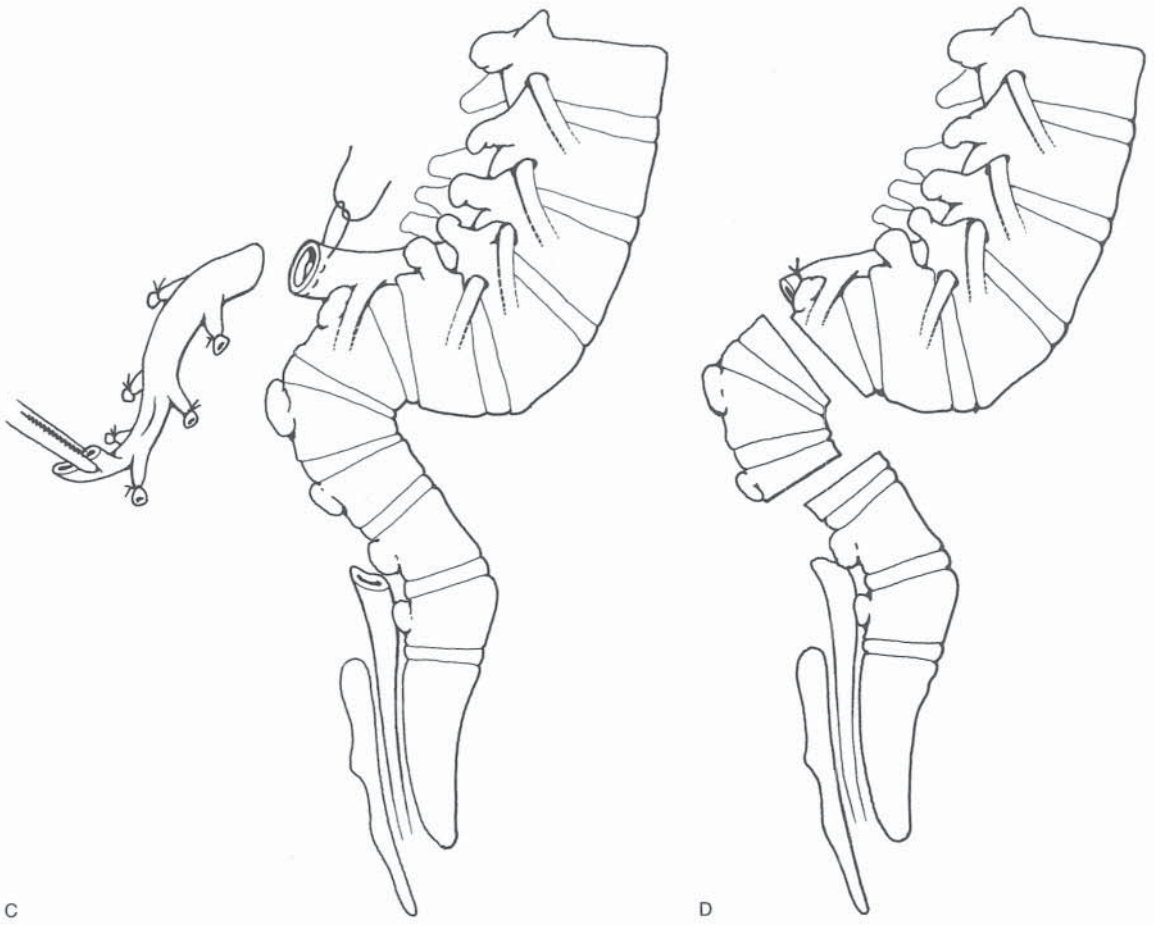
Continued

C, While the sac is held by its distal stump, the cord and sac remnants are transected proximal to the level of kyphectomy. The meninges only are tied with a purse-string suture at the level of resection. It is *essential* that the spinal cord not be sutured along with the meninges. Tying of the spinal cord may cause central canal obstruction and acute hydrocephalus, and has been associated with sudden perioperative death.

D, The lumbar spine is then exposed in either a subperiosteal or extraperiosteal fashion, as desired, exposing the kyphotic area of the spine circumferentially and allowing the paraspinal muscles, psoas muscle, retroperitoneum, and abdominal contents to fall anteriorly with the dissection. The extraperiosteal plane is usually more easily mobilized from the spine; segmental vessels will usually need to be ligated as the dissection proceeds. Subperiosteal dissection provides a stout protective membrane between the spinal column and the retroperitoneal area, but dissection will normally be more tedious and vascular.

To correct deformity, typically two vertebral body equivalents spanning three segments will need to be resected. The surgeon should carefully plan which vertebrae are to be resected to provide optimum realignment of the residual spinal column, ideally by making tracings of preoperative radiographs and studying the effect of vertebral body resection at different levels. Proximal thoracic lordosis and/or a milder kyphotic deformity of the spine below the major kyphotic deformity may shift the optimum area of resection (usually proximally). However, the surgeon should avoid centering the area of resection near the thoracolumbar or lumbosacral area; approximation and fixation of the residual spinal column is more difficult when resection has been carried out in these areas.

PLATE 25–3. Technique of Lumbar Kyphectomy in Myelomeningocele Patients with Fixation to the Pelvis Using the Dunn-McCarthy Technique



C D

Technique of Lumbar Kyphectomy in Myelomeningocele Patients with Fixation to the Pelvis Using the Dunn-McCarthy Technique
Continued

E, Fixation and fusion of the spinal column is then performed using the Luque technique. Loops of wire are passed under the laminae where they remain intact. We prefer to extend the fusion proximally into the upper thoracic spine to optimize spinal fixation and prevent the development of a midthoracic kyphotic deformity, which can result from shorter fusions. In the dysraphic area, wires can be looped around the transverse process/pedicle remnants or passed through drill holes in the pedicles if they are stout enough. Bone screws inserted directly into the vertebral bodies or figure-of-eight loops of wires passed around the bases of pedicles above and below the area of vertebral body resection are alternative methods of fixation in this area.

Optimum fixation to the pelvis has been provided, in our experience, by the Dunn-McCarthy modification of the Luque instrumentation. Prefabricated right and left S-shaped stainless steel rods are contoured to the residual deformity (ideally lordotic, but in actuality usually straight or slightly kyphotic). The lower S portion of each rod is passed around the front of the sacral alae and secured to the spine below the kyphectomy area. Both rods are then pushed anteriorly toward the proximal spinal segment, thereby reducing the kyphotic deformity, and the wires of the proximal spinal segment are gradually tightened to the rod.

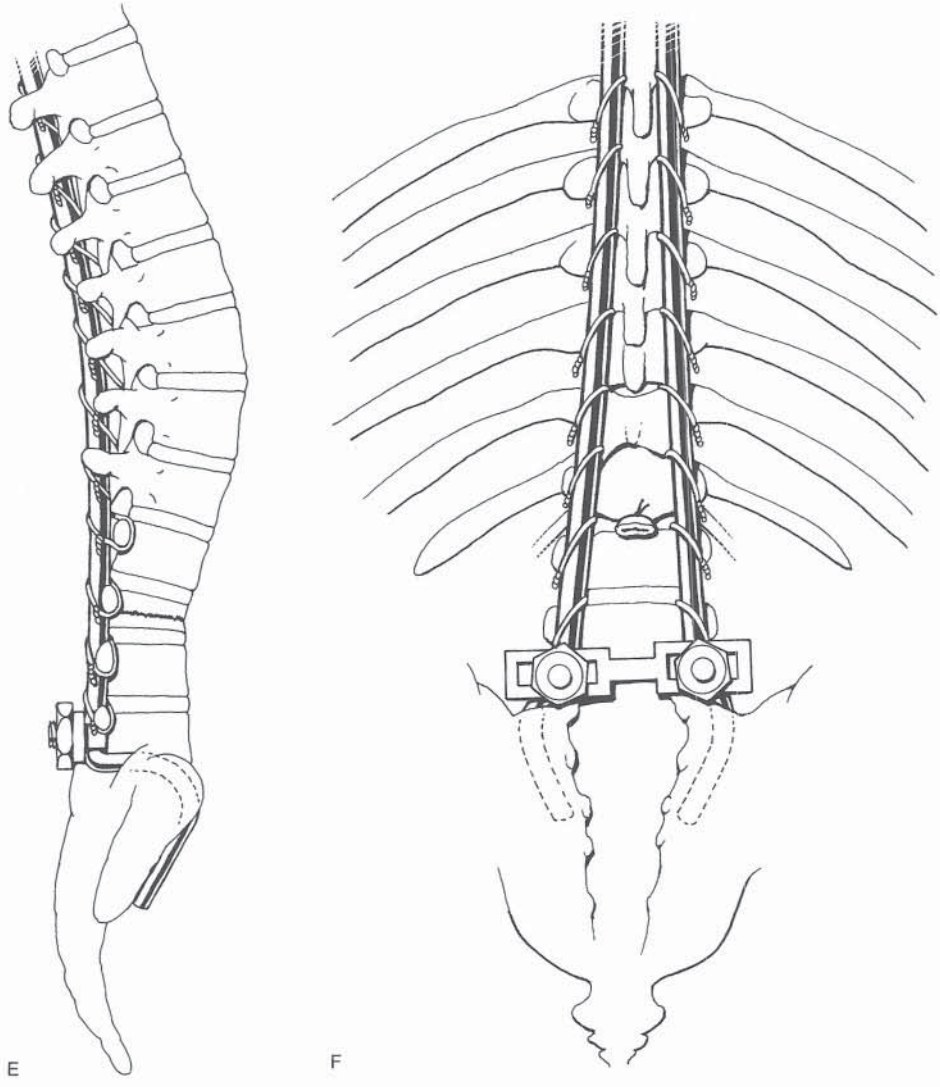
F, After completion of fixation, bone graft is placed around the rods from the sacrum to the upper limit of the fusion. The rods should be cross-linked to provide maximum construct stability. The wound is then closed over drains.

POSTOPERATIVE MANAGEMENT

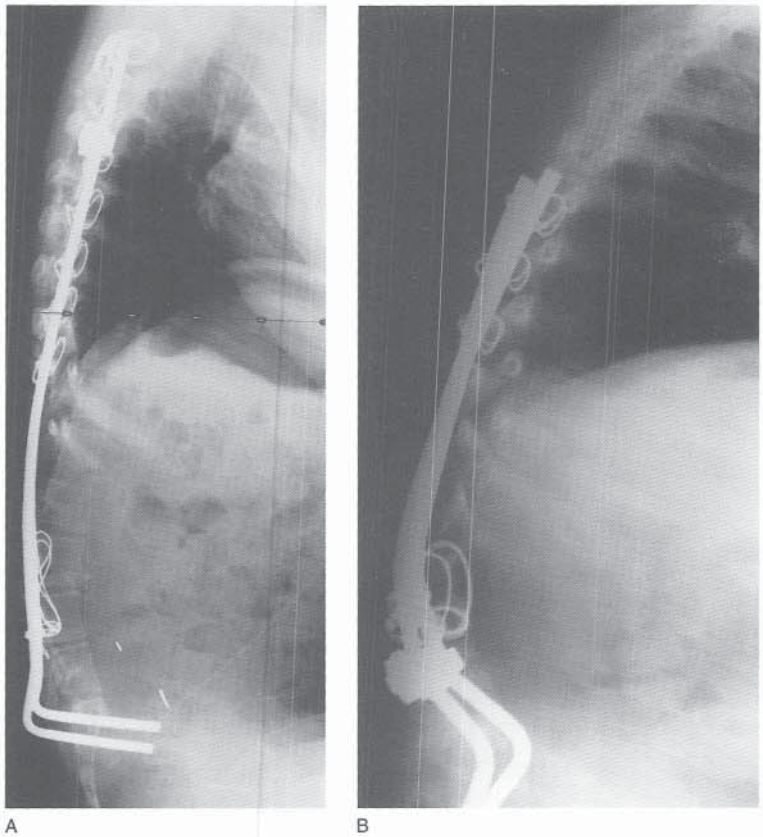
The patient should be initially nursed in bed, either prone or supine. We prefer to have the patient sitting as soon as feasible, depending on the adequacy of spinal fixation. If extremely secure fixation has been accomplished, external immobilization is not necessary. However, for 6 weeks, the patient should not be allowed to pull him- or herself into a sitting position or to self-transfer. Nursing staff and caretakers should be taught to transfer the patient to and from the bed, chair, and toilet in an effective sitting position, that is, with the legs and torso moved simultaneously without torque on the lumbosacral spine or hips.

If fixation is not secure or if compliance is not assured, the patient can be immobilized in a custom-molded thoracolumbospinal orthosis with thigh extensions fixed or hinged to allow the sitting position. This will provide external support in the sitting position and minimize movement between the legs, pelvis, and spine during transfers. If fixation is completely inadequate, the patient can be nursed prone or supine in bed until satisfactory healing has occurred; this course is to be avoided if at all possible.

PLATE 25–3. Technique of Lumbar Kyphectomy in Myelomeningocele Patients with Fixation to the Pelvis Using the Dunn-McCarthy Technique



E F



A B

FIGURE 25–21 Fixation options for definitive surgical management of kyphosis. **A,** Luque segmental wire and Galveston instrumentation fixation to the pelvis. **B,** Luque wire with Dunn-McCarthy fixation to the pelvis. This is our preferred stabilization technique.

carefully to avoid the potential loss of neurologic function. Finally, these challenges, combined with scarring of the posterior soft tissues, make for a much higher than average incidence of wound healing and deep infection complications.*

In many patients, the combination of posterior element deficiency and relative skeletal immaturity mandates anterior or combined anterior and posterior spinal fusion.† Internal fixation may be anterior, with vertebral body screws and rod, or posterior, with rods, hooks, wires, and pedicle bone screws with fixation to the pelvis.‡

Postoperative Management After Spinal Fusion. Urinary tract infections (which threaten both the urinary tract and the posterior spinal fusion site), wound infections, pressure sores, implant failure, and pseudarthrosis are all postoperative problems unique or more frequent in the myelomeningocele patient after extensive spinal fusion, particularly with instrumentation.

The patient's urinary tract should be managed by prompt return to the preoperative management (usually clean intermittent catheterization) with postoperative urine cultures, and prompt aggressive treatment of early postoperative urinary tract infections. The surgical wound must be kept covered with a sterile dressing until healed, and regularly inspected for evidence of inflammation, necrosis, hematoma, CSF collection, or drainage. If present, these conditions should be managed aggressively as well, with surgical debridement as required.

Fusions to the pelvis with instrumentation must be carefully protected during early union (the first 6 to 12 weeks). During this time, we do not allow independent transfers by the patient, and we teach the parent and other caretakers to move the patient's spine, pelvis, and lower extremities as a unit to prevent excessive force on the instrumentation through the lumbosacral junction or the hips. If necessary, a spinal orthosis with thigh extensions is fabricated to protect these areas. The patient's skin must be carefully monitored for evidence of irritation or impending breakdown in the new weightbearing areas of the sacrum, buttock, and thighs. Sitting should be resumed gradually, with assessment of these areas after the initial 20 minutes of sitting postoperatively, and periodically thereafter. Adjustments to the wheelchair cushion and back support are almost always necessary. Finally, in follow-up, the surgeon must monitor for evidence of deep infection or pseudarthrosis as indicated by implant failure or progressive deformity.

Results of Spinal Fusion for Scoliosis in Myelomeningocele. A review of early reports of spinal fusion in myelomeningocele patients is instructive: the surgeon simultaneously realizes how much experienced multidisplinary care and improved surgical techniques and instrumentation have impacted the results of spine fusion and is sobered by the frequency and gravity of postoperative complications in this patient population. Sriram and colleagues found that of 33 patients treated by a variety of methods (including Harrington rod instrumentation in 26), seven developed deep wound infections, 14

* See references 9, 142, 173, 183, 213, 257, 258, 284, 285.

† See references 9, 142, 183, 213, 258, 284, 285.

‡ See references 9, 22, 142, 183, 213, 234 284.

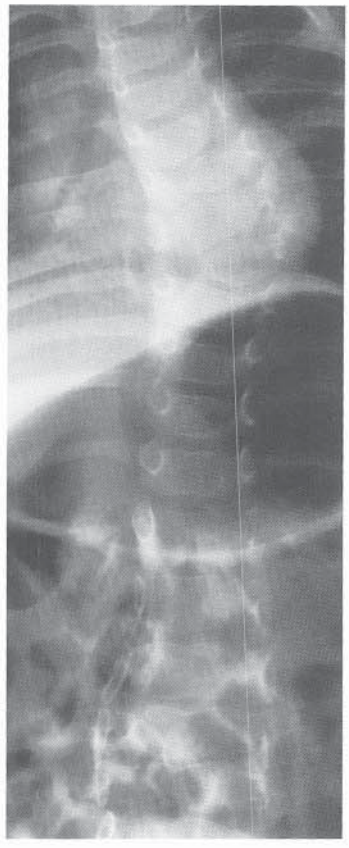


FIGURE 25–22 Congenital abnormalities of the spine associated with myelomeningocele. Spinal dysraphism and congenital vertebral anomalies are often best appreciated on infant films. In all infants with myelomeningocele, radiographs of the entire spine should be obtained and studied for evidence of these deformities. This radiograph demonstrates opening of the posterior spinal elements from the L1 level, a hemivertebra at T8, and a butterfly vertebra at T6.

had at least one pseudarthrosis, and in two the neurologic deficit worsened postoperatively.[257] Drummond and colleagues in a review of 18 patients treated surgically by a variety of methods, found that solid fusion was eventually achieved in 15; there were three cases of deep infection, ten of pseudarthrosis, ten fractures, and five patients developed pressure sores with plaster immobilization.[72]

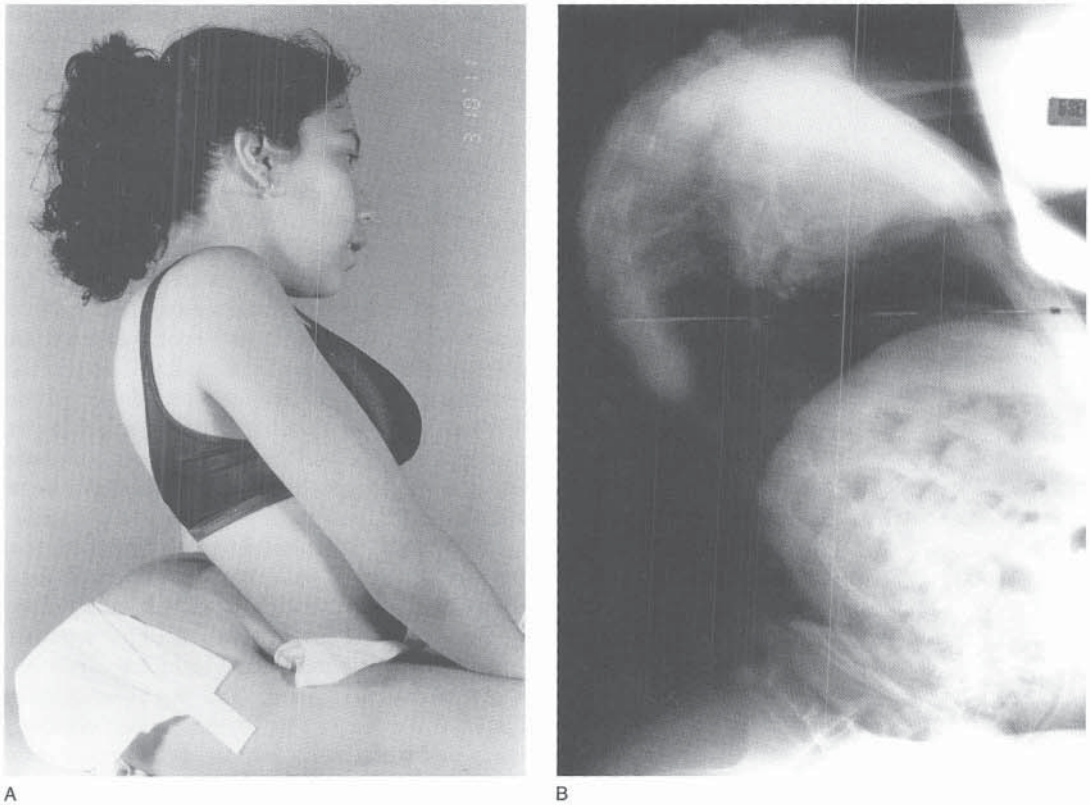
Posterior fusion or anterior fusion alone appears to be inadequate for most patients with paralytic scoliosis in myelomeningocele.[213,258] Stark and Saraste found that five of eight patients treated by anterior fusion and Zielke instrumentation had progressive spinal deformity above the area of anterior surgery requiring further posterior surgery despite solid fusion from the first procedure.[258] Osebold and colleagues found that combined anterior and posterior fusion resulted in a reduction in the pseudarthrosis rate to 23 percent from 46 percent for posterior fusion alone.[213] Ward and colleagues found that seven of 14 patients treated by isolated anterior or posterior fusion with or without instrumentation developed pseudarthrosis, compared to only two of 24 patients treated by combined anterior and posterior fusion with a variety of instrumentations.[284] McMaster reported that 21 of 23 patients treated by staged anterior spinal fusion with Dwyer instrumentation followed by posterior spinal fusion with Harrington instrumentation had improved posture and function; however, one patient died of cardiorespiratory failure, four had wound necrosis, two had deep wound infections, and one had a lumbosacral pseudarthrosis.[183]

The best results of spinal fusion for paralytic scoliosis in myelomeningocele patients occur in patients treated by combined anterior and posterior fusion with stable segmental fixation achieved by a combination of sublaminar wires, pedicular remnant wires, and/or pedicular screws.[9,284] Banta reported the results of combined anterior and posterior fusion with Luque rods and sublaminar and pedicular remnant wires in 50 patients with scoliosis or kyphosis.[9] In four patients deep wound infections developed, necessitating rod removal in one. In six patients pseudarthrosis developed, in three at the thoracolumbar junction, requiring supplemental posterior fusion, and in three at the lumbosacral junction, which were asymptomatic and did not require repair. Overall, the addition of anterior fusion to posterior fusion and instrumentation resulted in greater correction of spinal deformity and pelvic obliquity and improved the fusion mass over that achieved with posterior fusion alone.

Our preferred surgical treatment for scoliosis in patients with myelomeningocele is a single-stage, combined anterior spinal release and fusion and posterior spinal fusion with Luque instrumentation to the pelvis. We use sublaminar wires under the intact lamina, pedicular wires or screws in the area of posterior element insufficiency, and either Luque-Galveston or Dunn-McCarthy fixation to the pelvis (Plate 25–3).

HYPERLORDOSIS. A less common but potentially difficult to treat spinal deformity in patients with myelomeningocele is hyperlordosis, with or without associated scoliosis.[8,259] Hyperlordosis can lead to difficulty sitting, intertriginous skin breakdown, and difficulty with self-catheterization in females because of the posterior rotation of the perineum (Fig. 25–23). In the past, this deformity was associated with lumboperitoneal shunting,[259] but this method of shunting is rarely used currently. Treatment, when required, is by a combination of anterior and posterior spinal release and posterior instrumentation; in severe rigid deformities, postural reduction in traction after spinal release prior to definitive instrumentation may improve the deformity.[259]

HEMIMYELODYSPLASIA. A relatively rare manifestation of myelomeningocele is characterized by significantly asymmetric involvement of the lower extremities, with one leg being significantly affected and the contralateral leg being normal or nearly normal. This condition is referred to as "hemimyelodysplasia" or "hemi-spina bifida." The affected extremity shows all the typical manifestations of myelomeningocele—motor and sensory paralysis, congenital deformities, and deformities developing secondary to the motor paralysis. In addition, significant limb length inequality develops in most patients because of paralysis-induced growth inhibition on the affected side. A review of ten such patients by Maguire and colleagues found that scoliosis and limb length inequality had developed in all patients, and six of the ten had congenital renal anomalies.[158] Many of the patients in their report had congenital vertebral anomalies and intrathecal anomalies, so these patients require thorough assessment of the spinal column, spinal cord, and urinary system. Limb



A B

FIGURE 25–23 Hyperlordosis in myelomeningocele. **A,** Clinical appearance. Excessive lordosis can interfere with self-catheterization, particularly in girls. **B,** Radiographic appearance.

length inequality will require orthotic management, epiphysiodesis, or lengthening, as clinically indicated (Fig. 25–24).

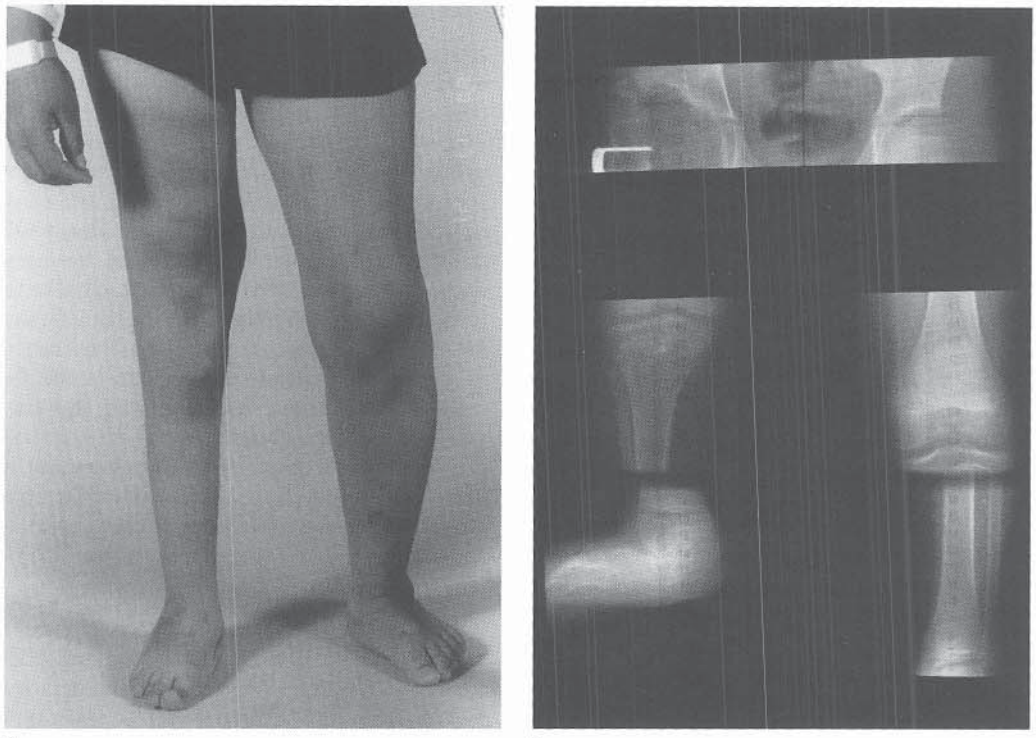
ORTHOTIC MANAGEMENT

General Principles of Bracing/Rehabilitation. A principal component of the management of patients with myelomeningocele is the prescription of orthoses to stabilize joints in the absence of lower extremity muscle function to facilitate weightbearing and ambulation.* Only rarely will a patient with a low sacral level lesion not require bracing at all; the huge majority of children will need lower extremity braces to accomplish upright positioning and ambulation. It behooves the orthopaedist to be familiar with basic orthotic principles, as he or she often provides the prescription for orthoses, checks their fit, and confirms that the prescription is appropriate and achieves the desired maximal mobility with minimal intrusion.

The purpose of lower extremity orthotics is to substitute for lower extremity muscle function, which, when present normally, stabilizes joints during weightbearing and powers advancement of the limbs in ambulation. Lower extremity orthoses can also provide protection for insensate skin. The initial orthotic fitting should be carried out when the child is neurodevelopmentally ready to walk and capable of working with a physical therapist, rather than at a specific age (such as 12 months). Patients with thoracic or upper lumbar level lesions should have achieved enough trunkal balance and strength to sit independently before lower extremity bracing is considered. Patients with lower lumbar or sacral level lesions usually have enough lower extremity strength to pull themselves into a standing position, and this is the time to fill the first prescription. As a generality, this neurodevelopmental stage is achieved somewhat later than in children without myelomeningocele, usually in the 18- to 24-month range. If surgery such as hip abduction contraction release or surgical release of clubfoot deformity is required to enable the orthotist to fit the patient, this surgery should be timed so that the patient can be placed in lower extremity braces upon removal of the postoperative casting, and standing and ambulation can begin immediately. Proceeding with surgery or brace fitting for the purposes of standing or walking before the child has reached these milestones to placate anxiety that a 12-month-old child "is not yet walking" serves only to create frustration for parents, child, and health care workers alike, and may deflect attention away from the fact that unlimited independent ambulation is not a realistic goal for many patients.[7,16,66,188,268]

The first prescription provided should usually extend one joint level beyond that predicted to be necessary in the long term, based on the childs's level of muscle paralysis. The child who is just beginning to assume upright mobility and who is compensating for muscle weakness with relatively heavy and restrictive lower extremity orthotics can be expected to experience muscle fatigue, joint instability, rotational instability, and lack of confidence. All of these factors tend to unmask less than normal strength and joint stability,

* See references 56, 84, 96, 97, 127, 145, 155, 171, 174, 203, 220, 229, 236, 280, 290.



A B

FIGURE 25–24 Clinical and radiographic appearance of a patient with hemimyelodysplasia involving the right lower extremity. **A,** Clinical appearance. Typically there is pronounced asymmetry between the limbs in function, and leg length inequality. **B,** Radiographs demonstrating an 8-cm leg length inequality. The patient had previously undergone a varus osteotomy of the right hip.

particularly in the hip and knee. Furthermore, any orthotist will readily attest to the fact that reduction of HKAFOs to KAFOs and KAFOs to AFOs is infinitely more easily and quickly achieved than trying to extend an orthosis one level higher than originally fit. Thus, patients with good quadriceps function who might be expected to require AFOs in the long run should be fitted with KAFOs initially, and those with less than good quadriceps strength should be fitted with HKAFOs initially. We believe that the purpose of the initial fitting is to gain upright mobility, and that orthotic adjustments should be minimal and prompt, to maintain focus on that initial goal. Once the precise extent of bracing an individual child needs has been confirmed and the initial goal of independent upright positioning and ambulation has been achieved, more elegant, lightweight orthotics made of polypropylene and more sophisticated joint components can be prescribed.

Crutches or walkers are integral components of ambulation for children with myelomeningocele, even those with lower lumbar and sacral levels of paralysis.[280] Their use allows the child to transfer some weightbearing to the upper extremity. Many children will adopt a swing-to or swing-through gait pattern using crutches and any form of lower extremity bracing. This provides the child with a rapid but not necessarily energy-efficient means of ambulation.

Specific Protocols for Levels

THORACIC AND UPPER LUMBAR LEVEL PATIENTS. Patients with thoracic or upper lumbar levels of paralysis will not be able to pull themselves to stand, and thus this neurodevelopmental milestone cannot be relied on to determine when a lower extremity orthotic prescription should be initiated. Stable independent sitting balance without the need for constant upper extremity support is usually taken as a good indication that these patients are ready for upright positioning. Full lower extremity bracing (with HKAFOs) is likely to be required permanently, and the long-term outlook for maintenance of ambulation, other than for transfers or exercise only, is not good. The family and health care team should both be aware that wheelchair mobility is the most likely end result. However, some highly motivated patients with this extensive level of paralysis will maintain some ambulatory potential, so that in general, providing the opportunity to ambulate with braces as children is appropriate.

When the patient appears ready for upright positioning, these patients do best with standing frames or similar full upright positioning devices (Fig. 25–25). "Wheeled standers" will allow children a significant amount of mobility on flat surfaces at home or school (Fig. 25–26). Once the patient has become comfortable with the upright position and not resistive to it, consideration can be given to converting to HKAFOs; these braces, when the joints are locked, will allow both standing support (although without the broad base of support that a standing frame or similar device will provide) and ambulation with the orthotics and an upper extremity aid, specifically a walker or perhaps crutches. Patients who do not tolerate or who are afraid of the upright position should not be forced into it; attempts should be made later, when the child has developed further. Satisfactory function of the upper extremities is a prerequisite for consideration

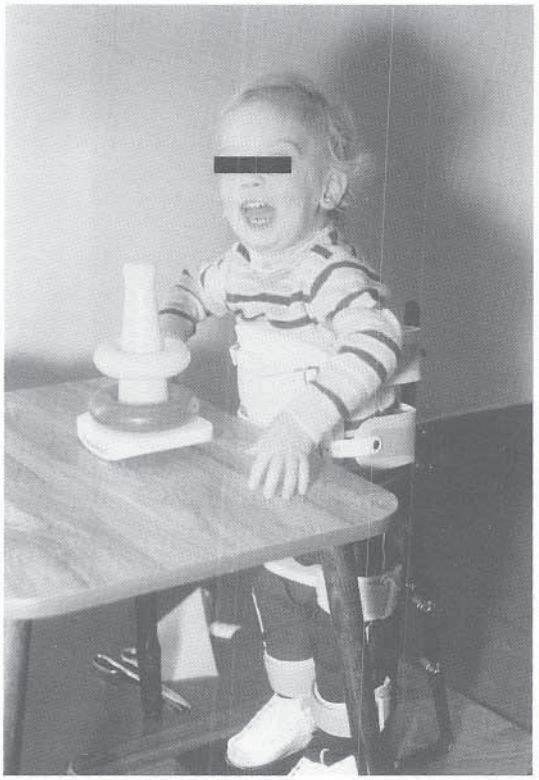


FIGURE 25–25 Standing frame for patients with significant lower extremity paralysis. This device may be used to introduce the child to upright positioning and requires relatively little adjustment for proper fitting.

of HKAFOs. After the child has demonstrated good acceptance and adaptation to upright mobility, reciprocating gait orthoses (see below) should be considered. Patients with some quadriceps function and strong adductors may ultimately ambulate in KAFOs.

LOWER LUMBAR AND SACRAL LEVEL PATIENTS. Typically, patients with lower lumbar or sacral level paralysis will have good to excellent quadriceps function, and for the most part they should be able to function with AFOs. The first prescription should be for KAFOs in most children, since their initial efforts to ambulate, coupled with the need to compensate for lower extremity weakness with braces, may manifest as insufficient knee stability for maximum mobility in AFOs alone. The consequences of fitting a child with AFOs when KAFOs are required include recurrent falls, knee effusions, and an unwillingness to ambulate. Usually, the timing for fitting the initial orthosis is more easily determined than in patients with upper lumbar or thoracic levels of paralysis, since the majority of patients will at least attempt to pull to stand. Surgery for congenital foot or knee deformities should be timed such that the patient can be fitted with the initial orthotic prescription on removal of the postoperative casting.

Polypropylene orthoses may be considered for the first prescription, since the most complex subsequent adaptation after initial fitting is to reduce KAFOs to AFOs once the lack of need for knee support has been documented in practice. Frequent falls, loss of confidence, or recurrent knee effusions indicate the need to reinstitute KAFOs.

Specific Braces

ANKLE-FOOT ORTHOSES. Polypropylene AFOs are a mainstay of lower extremity bracing in myelomeningocele patients (Fig. 25–27). Only rarely will a patient need not even these braces to maintain maximum endurance and comfort. The primary purposes of AFOs are to provide protection to the foot and toes during weightbearing and to stabilize the flail or poorly motored ankle. AFOs are ideally suited for patients with sacral level lesions, and often the only type of brace these patients require. A variation suitable for patients with lower lumbar level lesions is the ground-reaction AFO (Fig. 25–28). The proximal anterior tibial component is meant to counteract calcaneus moment at the foot by pressing against the shin to more effectively prevent ankle sag into dorsiflexion with weightbearing.[114,272] Because of this tendency for ankle dorsiflexion in any patient with inadequate gastrocsoleus function, articulated AFOs are usually not beneficial to patients with myelomeningocele.

KNEE-ANKLE-FOOT ORTHOSES. Knee-ankle-foot orthoses are common to many myelomeningocele patients, since they are required for patients with upper lumbar level lesions and are the recommended first prescription for most patients with lower lumbar level lesions. The orthosis consists of an AFO component, a thigh cuff, and some type of knee hinge (Fig. 25–29).

KAFOs must be aligned to the patient's limb, taking into account knee flexion deformity, varus or valgus deformity at the knee, and rotational deformities in the tibia, ankle, or foot. Usually, approximately 20 degrees of knee flexion deformity is readily tolerated by the patient without significantly affecting walking ability. Greater deformities will usually obligate the patient to walk with the knee hinges locked.

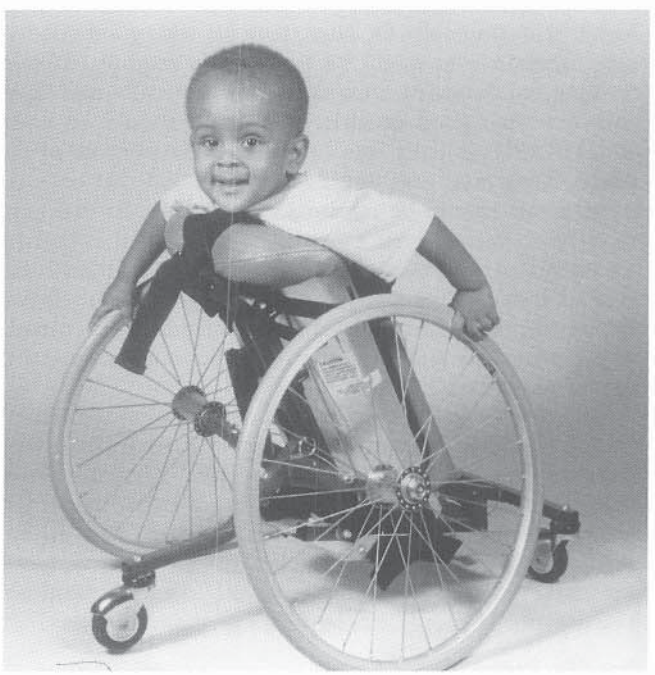


FIGURE 25–26 The "wheeled stander" gives the child mobility in the equivalent of the standing frame. This device is suitable for young children with significant lower extremity paralysis who otherwise are unable to stand or move about.

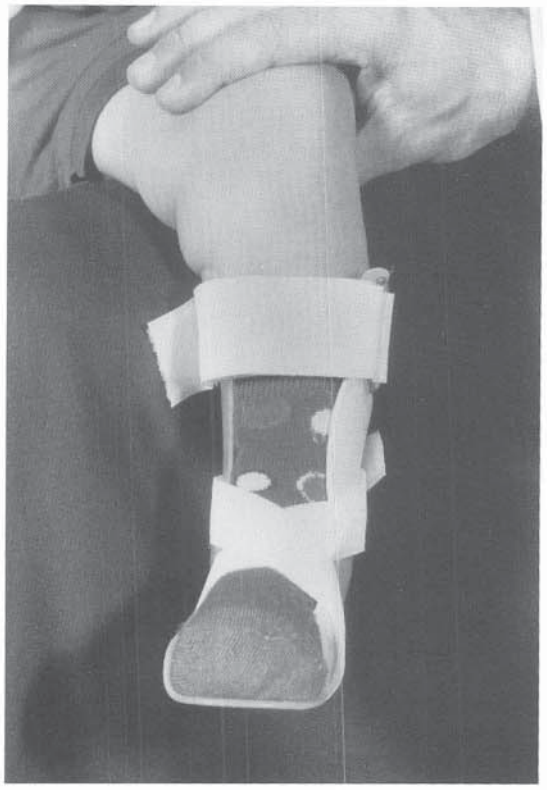


FIGURE 25–27 A solid-ankle polyethylene ankle-foot orthosis. This orthosis may be appropriate for patients with sacral level myelomeningocele and for the occasional patient with lower lumbar myelomeningocele and minimal knee and foot deformities.

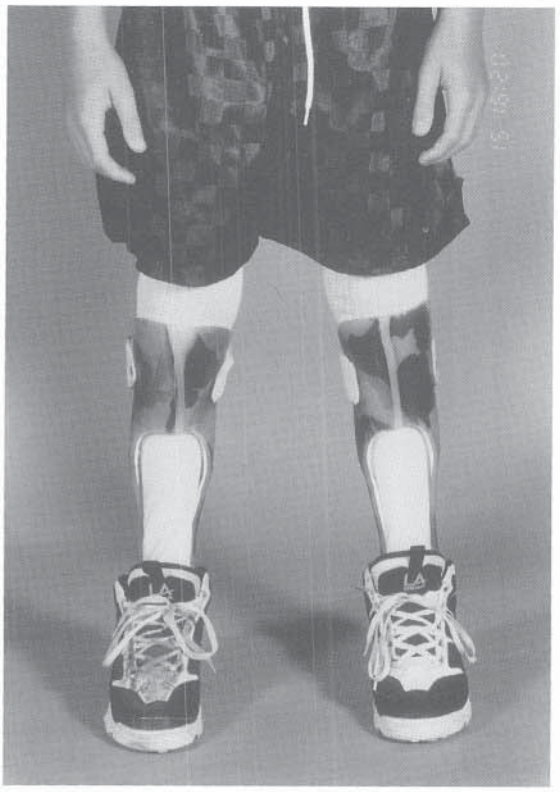


FIGURE 25–28 Ground-reaction ankle-foot orthoses. The anterior portion of the brace presses against the shin with dorsiflexion moment at the foot to prevent excessive dorsiflexion of the ankle and flexion at the knee.

Ambulation in such a position will usually be labored, and usually the patient will adopt a swing-to or swing-through gait strategy, depending on the abdominal and hip flexor muscle strength. If the knee hinges are not positioned to reflect the extent of knee flexion deformity in the knee-locked position, pressure areas will develop at the top of the cuff in the back. This area is often troublesome even when the fit is correct for the amount of knee flexion deformity, since patients often tend to lean backward over the top of the orthoses when standing (Fig. 25–30), or the orthoses may be pushed into this area when the patient is sitting.

HIP-KNEE-ANKLE-FOOT ORTHOSES. Hip-knee-ankle-foot orthoses are extensive braces required in all patients without adequate hip strength or stability to allow weightbearing in stance in KAFOs alone. Patients with any strength less than good quadriceps strength are usually best fitted with HKAFOs initially, for the reasons discussed above. These orthotics typically consist of two long-leg braces (KAFOs) as described above, connected by a pelvic band and a free hip joint (Fig. 25–31). The free hip hinge provides medial-lateral stability in both standing and walking, and will provide rotational stability. The most significant difference between HKAFOs and KAFOs for the orthotist is that limb length inequality (true or apparent) and rotational, flexion, and adduction or abduction deformities through the hip must be taken into account in aligning the orthotic. Thus, extending a prescription for KAFOs to HKAFOs by the addition of a pelvic band is often a significant undertaking

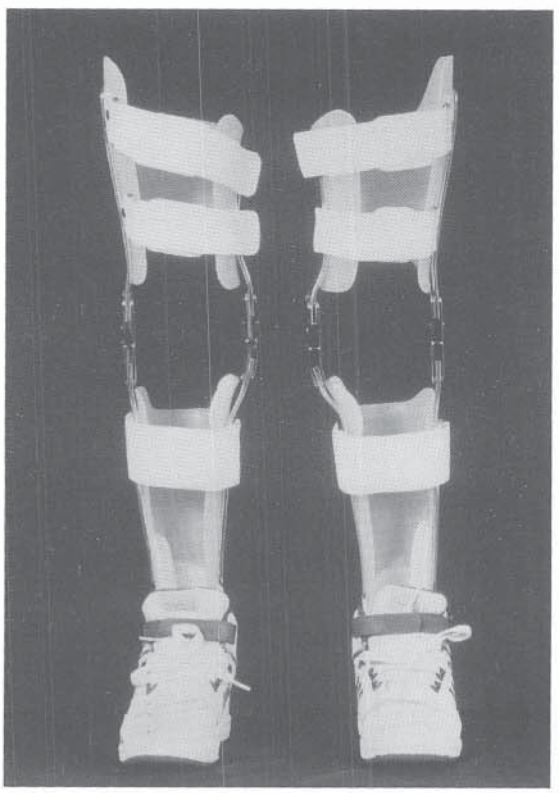
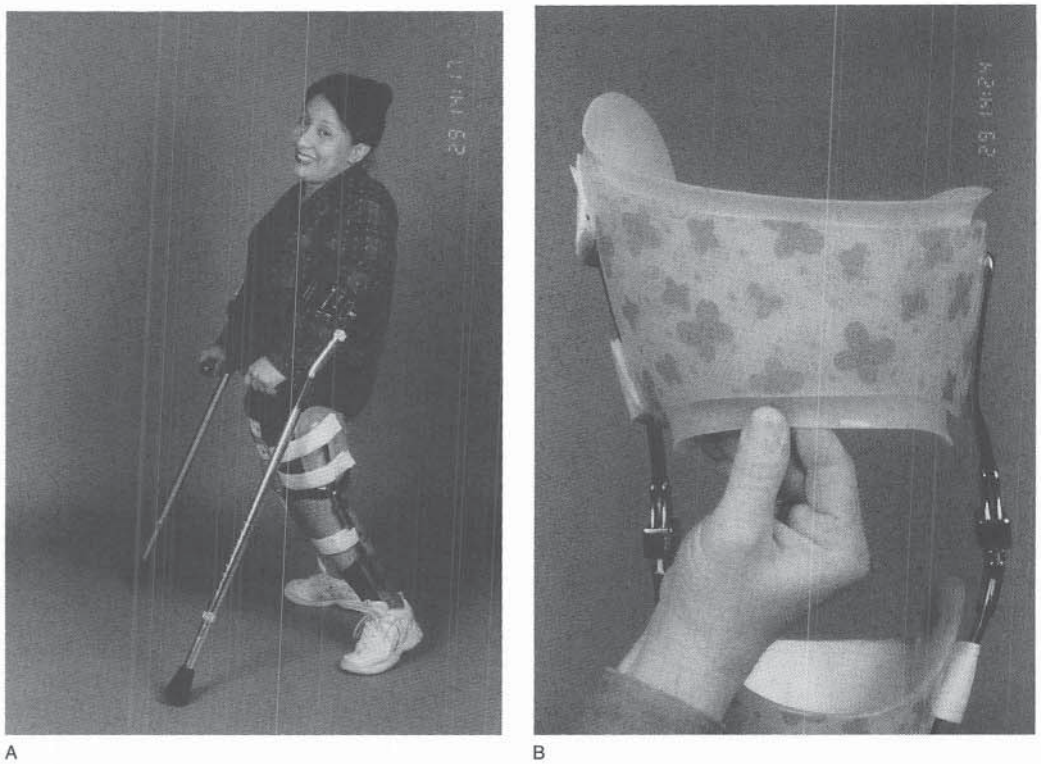


FIGURE 25–29 Standard knee-ankle-foot orthoses have plastic thigh and AFO components connected with a knee hinge (typically a free, drop-lock hinge).



A B

FIGURE 25–30 Orthoses must be made to fit the patient's deformities. **A,** Long-leg braces that do not accommodate knee flexion deformity will dig into the posterior aspect of the child's upper thigh, and can produce pressure sores in this area. **B,** Milder deformities may be accommodated by inlaying a softer plastic insert into the thigh portion of the KAFO to prevent excessive pressure in the posterior-superior thigh area.

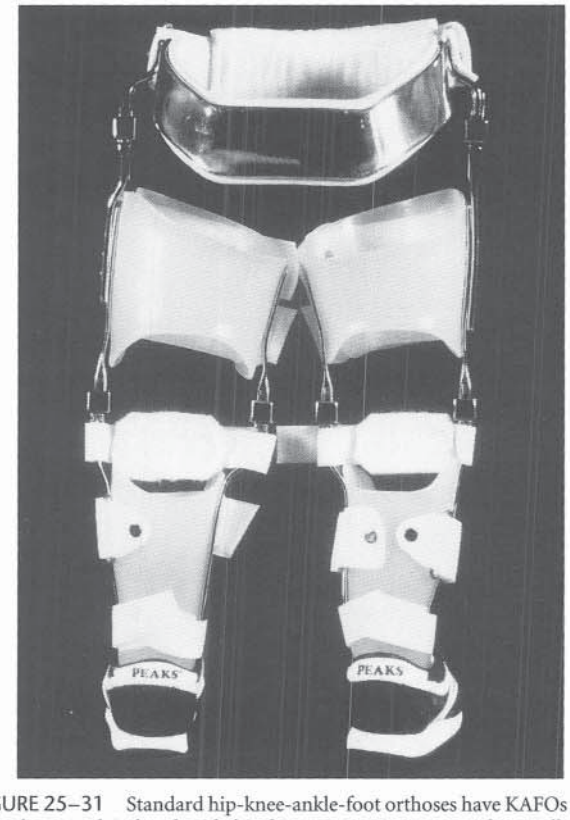


FIGURE 25–31 Standard hip-knee-ankle-foot orthoses have KAFOs connected to a pelvic band with hip hinge joints incorporated. Usually free or drop-lock hinges are used.

for the orthotist; removing an unneeded pelvic band, however, is a very simple procedure.

HKAFOs represent very significant bracing and make transitional movements very awkward. This fact, combined with the implied extensive muscular weakness of the lower extremities, makes this mode of ambulation impractical in most adolescents and adults. Patients requiring HKAFOs for ambulation as children will usually, as adolescents or adults, favor full-time wheelchair use, or alternatively will use KAFOs to assist with transfers only.

RECIPROCATING GAIT ORTHOSES. The concept of reciprocating gait orthoses (RGOs) was originally introduced at the Ontario Crippled Children's Treatment Center and refined by Douglas and colleagues.[70] These braces are a sophisticated form of HKAFOs; they consist of long-leg braces with a connecting pelvic band. The unique feature of the orthosis is that the two long-leg components are connected by spring-loaded cables (Fig. 25–32). Flexion of the hip with advancement of the limb produces passive contralateral hip extension through the springlike cable. A longer, more energy-efficient stride is thus obtained. Ideal candidates for this orthosis are patients with upper lumbar level lesions (i.e., with hip flexor power only) who have demonstrated motivation for ambulation and no major contractures or deformity preventing fitting of the long-leg components of the braces. In such patients, gait laboratory and clinical studies have demonstrated a more efficient, less energy-consuming ambulation compared to ambulation in conventional HKAFOs.[56,96,171,174,220,294] However, many children will

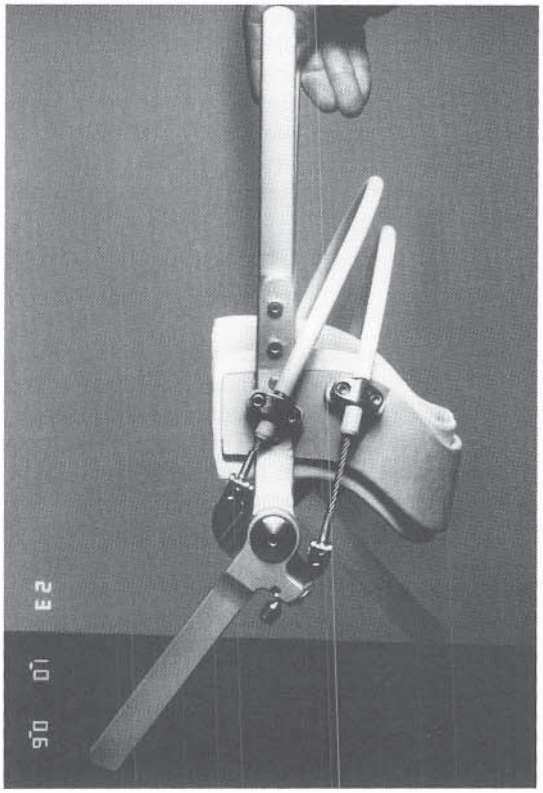


FIGURE 25–32 The pelvic component of a reciprocating gait orthosis. The KAFO components are connected by a spring-loaded cabling apparatus. When the patient advances one limb (by flexion of that hip), dynamic extension is produced in the contralateral limb.

adopt a swing-to or swing-through gait pattern with crutches and HKAFOs.[56,171,174,294] This pattern of ambulation is faster than reciprocating gait, irrespective of whether the child is fitted with RGOs or conventional HKAFOs. RGOs have also been prescribed in patients with thoracic or lower lumbar levels of paralysis, but their value in these patients is less obvious.[174]

Other Mobility Aids

STANDING FRAME. Standing frames consist of a simple base (such as a partial sheet of plywood) with an A-frame-like upright support to which the child can be strapped. No actual mobility is provided with these devices, but the patient can be placed in an upright position in them. They are good initial test equipment when upright mobility is being considered in more severely impaired patients, as they are simple to adjust and can accommodate relatively severe deformities. They allow patients to interact with peers of their age group at eye level, and allow the parents, therapists, and physician an opportunity to assess the child's reaction to being in an upright position, as a precursor to ambulation in HKAFOs. Occasional patients react poorly, especially if they are capable of crawling, and resist being "locked" in one upright location. The child should not be forced to accept it. Also, a child in this upright position must be carefully monitored by a responsible adult. Usually, the child will stand at a table surface in the standing frame, and one must be careful that the child not learn to push himself away from the table, pitching backward in the apparatus.

PARAPODIUM. These devices are the equivalent of "mobile standing frames." Simple lockable hip and knee hinges are incorporated into the device. The patient can be held upright with the hinges locked, or sit with the joints unlocked. Walking can be accomplished with an upper extremity aid, usually a walker. Fitting is simpler than with HKAFOs because of less intimate contact between the limbs and the orthotic, but they are heavy and bulky. Their use is limited to patients with thoracic level lesions motivated as children to be in an upright position with limited mobility, and in this respect they have an advantage over a standing frame. They are not useful long-term or adult mobility aids.[145] At our institution, we prefer to use wheeled standers as a mobility aid for this patient group.

WHEELCHAIRS. Many variations of wheelchairs are currently available to the paraplegic population. The wisest course for the physician is to recruit the assistance of a knowledgeable physical therapist, occupational therapist, or other mobility specialist to provide the ideal prescription for each child based on the child's needs. The majority of patients with myelomeningocele will use a wheelchair as adolescents and adults, even those with lower lumbar level lesions, since wheelchairs are an energy-efficient means of transportation. Parents of patients who have achieved walking are often reluctant to fill a prescription for a wheelchair for their older child or adolescent out of fear that the child will "never walk again." They should be educated that *useful* ambulation or standing transfers will not be neglected by the child, and that the wheelchair is simply the most energy-efficient mobility device. If a child chooses never to walk again, it is because upright mobility with lower extremity bracing and an upper extremity aid was not useful to the child in any daily activity. The health care team should help guide the child and family to the many enjoyable endeavors possible with wheelchair mobility such as racing, basketball, tennis, and similar adapted recreational activities.

Almost all patients with myelomeningocele are excellent candidates for the use of manual wheelchairs, since their upper extremities are relatively unaffected and manual powering of the wheelchair provides the patient with much-needed exercise. Some controversy exists as to whether patients with higher levels of paralysis given wheelchairs as their sole method of mobility become more efficient wheelchair users than those who take to a wheelchair subsequent to exercise or limited ambulation, or alternatively, whether such patients incur fewer medical problems such as decubiti, joint contracture, bone fragility, hydronephrosis, or obesity if they are taught upright mobility. Studies indicate that neither position is, in general, correct.[145,170] Thus, the health care team in consultation with the family must individualize decisions about the timing of introduction of wheelchairs and the extent to which efforts at upright mobility are made, based on each child's needs and desires.

GENERAL CARE OF THE MYELOMENINGOCELE PATIENT

Skin. Insensate skin represents a constant source of potential skin breakdown in the majority of patients with myelomeningocele, and parents, caretakers, and the patients themselves must be carefully taught how to prevent pressure

sores. Initially, parents must be cautioned to prevent contact with excessively hot or cold surfaces or excessive exposure to sunlight or cold. Bath water must always be checked by the parent before the child is immersed in it. The immobile child must be turned regularly and positioned carefully in bed. The diaper should be changed frequently to keep the perineal area clean, and diaper rashes should be treated promptly with appropriate creams and exposure to air. When shoes are put on, they must be opened fully to be sure that the child's toes do not curl up in the toebox, causing sores here.

When the child gains crawling mobility, the lower legs must be protected from rough surfaces such as concrete to prevent excoriation. A common source of foot ulceration is from contact with the rough surfaces of a pool; protective "wading shoes" and education can help prevent this occurrence.

After the child has been fitted with lower extremity braces for ambulation, the braces must be inspected every day for broken components or evidence of undue pressure on the child's skin. The most common sources of undue pressure are in the instep with a planovalgus foot deformity; around the malleoli secondary to growth of the child or to a rotational or angular deformity affecting the position of the ankle relative to the orthotic; and at the top of the thigh portion of a long-leg brace posteriorly when the presence of a knee flexion deformity has not been accounted for in the alignment of the long-leg brace.

The sitting child must be fitted with a wheelchair which has good cushioning (such as eggcrate or Roho cushion); patients with pelvic obliquity or bony deformity about the pelvis or lower spine may need custom inserts to protect bony prominences from becoming excoriated. The child must be taught to stay dry and clean in the perineal area to prevent contact dermatitis leading to skin breakdown. Intermittent relief of the buttocks must be achieved by regularly performing a "pushup" in the chair; children and adolescence may need watch alarms or similar reminders to ensure that this occurs throughout the day. The perineal, posterior spinal, and ischial areas should be inspected every day as well, and any increased pressure marks reported to the physician.

Counseling. In managing children with myelomeningocele, it is important not to neglect the long-term goals of the patient as an individual in an effort to achieve short-term goals such as walking and prevention of urinary complications. Although these short-term goals are important in themselves and can have some impact on the long-term goals, we as caretakers and parents should realize that the long-term goal for any child with the intellectual ability to comply is to develop into a self-sufficient individual, able to look after him- or herself in activities of daily living and to seek gainful employment. Thus, the child's general education is very important. Learning to look after bowel function, paralyzed bladder function, and insensate skin is more important than the ability to take a few steps in the physical therapy department. As young adults, educational, employment, and sexual counseling are required. The young child with an upper lumbar level of paralysis may be able to achieve reasonable mobility with the aid of crutches, long-leg braces, and the encouragement of parents and therapists. That child will grow into either an independent or dependent adult according to whether he or she is well-educated, has been taught self-care, remains healthy by avoiding shunt, skin, and urinary tract problems, and has been guided toward realistic employment training. That child will *not* earn a living by walking, and the efforts and time devoted to that goal must be carefully weighed against the global needs of the child.

SUMMARY

Management of the child with myelomeningocele is one of the most challenging and rewarding areas that the pediatric orthopaedist will face. The surgeon will encounter in this patient population virtually all of the deformities for which we have been trained: congenital foot, knee, hip, and spinal deformities, fractures, contractures, muscle imbalance, and scoliosis. Correction of these deformities can be very beneficial to the child, and thus rewarding to the physician. However, the complication rate is higher and the success rate is lower in this patient population for virtually any given standard technique, which can be a source of frustration and discouragement to physician, parent, and child alike. Management of these children begins from birth and continues throughout growth. Nowhere in orthopaedics is the adoption of a team approach, spearheaded and coordinated by a developmental pediatrician or similar individual, more important to the delivery of health care to patients. The orthopaedist is an integral component of that team, and should devote energy to the correction of deformity, maximizing the child's mobility, and screening for loss of neurologic function or the development of deformities (especially spinal). These efforts must always be guided by the long-term goal of developing healthy, happy, employable, and self-sufficient young men and women.

REFERENCES

Myelomeningocele

1. Task Force on Allergic Reactions to Latex, American Academy of Allergy and Immunology: Committee report. J Allergy Clin Immunol 1993;92:16.
2. Abraham E, Lubicky JP, Songer MN, et al: Supramalleolar osteotomy for ankle valgus in myelomeningocele. J Pediatr Orthop 1996;16:774.
3. Alembik Y, Dott B, Roth MP, et al: Prevalence of neural tube defects in northeastern France, 1979–1994: impact of prenatal diagnosis. Ann Genet 1997;40:69.
4. Alman BA, Bhandari M, Wright JG: Function of dislocated hips in children with lower level spina bifida. J Bone Joint Surg 1996;78-B:294.
5. Appleton PL, Ellis NC, Minchom PE, et al: Depressive symptoms and self-concept in young people with spina bifida. J Pediatr Psychol 1997;22:707.
6. Aronson DD, Middleton DL: Extra-articular subtalar arthrodesis with cancellous bone graft and internal fixation for children with myelomeningocele. Dev Med Child Neurol 1991;33:232.
7. Asher M, Olson J: Factors affecting the ambulatory status of patients with spina bifida cystica. J Bone Joint Surg 1983;65-A:350.
8. Baker RH, Sharrard WJ: Correction of lordoscoliosis in spina bifida by multiple spinal osteotomy and fusion with Dwyer fixation: a preliminary report. Dev Med Child Neurol Suppl 1973;29:12.
9. Banta JV: Combined anterior and posterior fusion for spinal deformity in myelomeningocele. Spine 1990;15:946.
10. Banta JV, Bonanni C, Prebluda J: Latex anaphylaxis during spinal surgery in children with myelomeningocele. Dev Med Child Neurol 1993;35:543.

11. Banta JV, Nichols O: Autogenous fibular subtalar arthrodesis in myelodysplasia. J Bone Joint Surg 1973;55-A:1317.
12. Banta JV, Park SM: Improvement in pulmonary function in patients having combined anterior and posterior spine fusion for myelomeningocele scoliosis. Spine 1983;8:765.
13. Banta JV, Sutherland DH, Wyatt M: Anterior tibial transfer to the os calcis with Achilles tenodesis for calcaneal deformity in myelomeningocele. J Pediatr Orthop 1981;1:125.
14. Barden GA, Meyer LC, Stelling FH III: Myelodysplastics: fate of those followed for twenty years or more. J Bone Joint Surg 1975;57-A:643.
15. Bassett GS, Mazur KU, Sloan GM: Soft-tissue expander failure in severe equinovarus foot deformity. J Pediatr Orthop 1993;13:744.
16. Bax M: Walking [editorial; comment]. Dev Med Child Neurol 1991;33:471.
17. Beaty JH, Canale ST: Orthopaedic aspects of myelomeningocele. J Bone Joint Surg 1990;72-A:626.
18. Begeer JH, Schreinemachers AL, Terwisga PH: Growth and the tethered cord syndrome. Eur J Pediatr Surg 1992;2(suppl 1):23.
19. Begeer JH, Staal-Schreinemachers AL: The benefits of team treatment and control of adult patients with spinal dysraphism. Eur J Pediatr Surg 1996;6(suppl 1):15.
20. Berned Muller E, Nordwall A, von Wendt L: The influence of scoliosis brace treatment on function in children with myelomeningocele. Acta Paediatr 1992;81:925.
21. Bliss DG, Menelaus MB: The results of transfer of the tibialis anterior to the heel in patients who have a myelomeningocele. J Bone Joint Surg 1986;68-A:1258.
22. Boachie-Adjei O, Bradford DS: Vertebral column resection and arthrodesis for complex spinal deformities. J Spinal Disord 1991;4:193.
23. Boemers TM, Soorani-Lunsing IJ, de Jong TP, et al: Urological problems after surgical treatment of scoliosis in children with myelomeningocele. J Urol 1996;155:1066.
24. Bomalaski MD, Teague JL, Brooks B: The long-term impact of urological management on the quality of life of children with spina bifida. J Urol 1995;154:778.
25. Bono R, Inverno M, Botteon G, et al: Clinical features and MR imaging in children with repaired myelomeningocele. Ital J Neurol Sci 1993;14:553.
26. Boor R, Schwarz M, Reitter B, et al: Tethered cord after spina bifida aperta: a longitudinal study of somatosensory evoked potentials. Childs Nerv Syst 1993;9:328.
27. Bower C, Stanley FJ: Dietary folate as a risk factor for neural-tube defects: evidence from a case-control study in Western Australia. Med J Aust 1989;150:613.
28. Boytim MJ, Davidson RS, Charney E, et al: Neonatal fractures in myelomeningocele patients. J Pediatr Orthop 1991;11:28.
29. Breed AL, Healy PM: The midlumbar myelomeningocele hip: mechanism of dislocation and treatment. J Pediatr Orthop 1982;2:15.
30. Breningstall GN, Marker SM, Tubman DE: Hydrosyringomyelia and diastematomyelia detected by MRI in myelomeningocele. Pediatr Neurol 1992;8:267.
31. Brinker MR, Rosenfeld SR, Feiwell E, et al: Myelomeningocele at the sacral level: long-term outcomes in adults. J Bone Joint Surg 1994;76-A:1293.
32. Broome HL, Basmajian JV: Survival of iliopsoas muscle after Sharrard procedure: an electromyographic study. Am J Phys Med 1971;50:301.
33. Broughton NS, Graham G, Menelaus MB: The high incidence of foot deformity in patients with high-level spina bifida. J Bone Joint Surg 1994;76-B:548.
34. Broughton NS, Menelaus MB, Cole WG, et al: The natural history of hip deformity in myelomeningocele. J Bone Joint Surg 1993;75-B:760.
35. Bruner JP, Tulipan N, Paschall RL, et al: Fetal surgery for myelomeningocele and the incidence of shunt-dependent hydrocephalus. JAMA 1999;282:1819.
36. Bunch WH, Hakala MW: Iliopsoas transfers in children with myelomeningocele. J Bone Joint Surg 1984;66-A:224.
37. Burkus JK, Moore DW, Raycroft JF: Valgus deformity of the ankle in myelodysplastic patients: correction by stapling of the medial part of the distal tibial physis. J Bone Joint Surg 1983;65-A:1157.
38. Canale G, Scarsi M, Mastragostino S: Hip deformity and dislocation in spina bifida. Ital J Orthop Traumatol 1992;18:155.
39. Carroll NC, Sharrard WJ: Long-term follow-up of posterior iliopsoas transplantation for paralytic dislocation of the hip. J Bone Joint Surg 1972;54-A:551.
40. Carstens C, Koch H, Brocai DR, et al: Development of pathological lumbar kyphosis in myelomeningocele. J Bone Joint Surg 1996; 78-B:945.
41. Carstens C, Paul K, Niethard FU, et al: Effect of scoliosis surgery on pulmonary function in patients with myelomeningocele. J Pediatr Orthop 1991;11:459.
42. [Centers for Disease Control]: Recommendations for the use of folic acid to reduce the number of cases of spina bifida and other neural tube defects. MMWR Morb Mortal Wkly Rep 1992;41:1.
43. [Centers for Disease Control]: Spina bifida incidence at birth—United States. JAMA 1992;268:708.
44. [Centers for Disease Control]: Spina bifida incidence at birth—United States, 1983–1990. MMWR Morb Mortal Wkly Rep 1992;41:497.
45. Chan A, Robertson EF, Haan EA, et al: Prevalence of neural tube defects in South Australia, 1966–91: effectiveness and impact of prenatal diagnosis. BMJ 1993;307:703.
46. Charney EB, Melchionni JB, Smith DR: Community ambulation by children with myelomeningocele and high-level paralysis. J Pediatr Orthop 1991;11:579.
47. Charney EB, Weller SC, Sutton LN, et al: Management of the newborn with myelomeningocele: time for a decision-making process. Pediatrics 1985;75:58.
48. Chiari K: Medial displacement osteotomy of the pelvis. Clin Orthop 1974;98:55.
49. Christofersen MR, Brooks AL: Excision and wire fixation of rigid myelomeningocele kyphosis. J Pediatr Orthop 1985;5:691.
50. Clarke CA, McKendrick OM, Sheppard PM: Spina bifida and potatoes. BMJ 1973;3:251.
51. Coniglio SJ, Anderson SM, Ferguson JE II: Functional motor outcome in children with myelomeningocele: correlation with anatomic level on prenatal ultrasound. Dev Med Child Neurol 1996;38:675.
52. Cragan JD, Roberts HE, Edmonds LD, et al: Surveillance for anencephaly and spina bifida and the impact of prenatal diagnosis—United States, 1985–1994. Morb Mortal Wkly Rep CDC Surveill Summ 1995;44:1.
53. Cremer R, Hoppe A, Kleine-Diepenbruck U, et al: Longitudinal study on latex sensitization in children with spina bifida. Pediatr Allergy Immunol 1998;9:40.
54. Cremer R, Kleine-Diepenbruck U, Hoppe A, et al: Latex allergy in spina bifida patients: prevention by primary prophylaxis. Allergy 1998;53:709.
55. Cruess RL, Turner NS: Paralysis of hip abductor muscles in spina bifida: results of treatment by the Mustard procedure. J Bone Joint Surg 1970;52-A:1364.
56. Cuddeford TJ, Freeling RP, Thomas SS, et al: Energy consumption in children with myelomeningocele: a comparison between reciprocating gait orthosis and hip-knee-ankle-foot orthosis ambulators. Dev Med Child Neurol 1997;39:239.
57. Cuxart A, Iborra J, Melendez M, et al: Physeal injuries in myelomeningocele patients. Paraplegia 1992;30:791.
58. Davis CF, Young DG: The changing incidence of neural tube defects in Scotland. J Pediatr Surg 1991;26:516.
59. de Carvalho Neto J, Dias LS, Gabrieli AP: Congenital talipes equinovarus in spina bifida: treatment and results. J Pediatr Orthop 1996;16:782.
60. de Chalain TM, Cohen SR, Burstein FD, et al: Decision making in primary surgical repair of myelomeningoceles. Ann Plast Surg 1995; 35:272.
61. De Wals P, Trochet C, Pinsonneault L: Prevalence of neural tube defects in the province of Quebec, 1992. Can J Public Health 1999; 90:237.
62. Dega W, Krol J, Polakowski L: Surgical treatment of congenital dislocation of the hip in children: a one-stage procedure. J Bone Joint Surg 1959;41-A:920.
63. Dias LS: Surgical management of knee contractures in myelomeningocele. J Pediatr Orthop 1982;2:127.
64. Dias LS, Jasty MJ, Collins P: Rotational deformities of the lower limb in myelomeningocele: evaluation and treatment. J Bone Joint Surg 1984;66-B:215.
65. Dias LS, Stern LS: Talectomy in the treatment of resistant talipes equinovarus deformity in myelomeningocele and arthrogryposis. J Pediatr Orthop 1987;7:39.
66. Diaz Llopis I, Bea Munoz M, Martinez Agullo E, et al: Ambulation in patients with myelomeningocele: a study of 1500 patients. Paraplegia 1993;31:28.
67. Dise JE, Lohr ME: Examination of deficits in conceptual reasoning

abilities associated with spina bifida. Am J Phys Med Rehabil 1998;77:247.
68. Doers T, Walker JL, van den Brink K, et al: The progression of untreated lumbar kyphosis and the compensatory thoracic lordosis in myelomeningocele. Dev Med Child Neurol 1997;39:326.
69. Doran PA, Guthkelch AN: Studies in spinal bifida cystica. I. General survey and reassessment of the problem. J Neurol Neurosurg Psychiatry 1961;24:331.
70. Douglas R, Larson P, D'Ambrosia R, et al: The L.S.U. reciprocation gait orthosis. Orthopaedics 1983;6:834.
71. Drummond DS, Moreau M, Cruess RL: Post-operative neuropathic fractures in patients with myelomeningocele. Dev Med Child Neurol 1981;23:147.
72. Drummond DS, Moreau M, Cruess RL: The results and complications of surgery for the paralytic hip and spine in myelomeningocele. J Bone Joint Surg 1980;62-B:49.
73. Duffy CM, Hill AE, Cosgrove AP, et al: Energy consumption in children with spina bifida and cerebral palsy: a comparative study. Dev Med Child Neurol 1996;38:238.
74. Duffy CM, Hill AE, Cosgrove AP, et al: Three-dimensional gait analysis in spina bifida. J Pediatr Orthop 1996;16:786.
75. Dwyer AF, Newton NC, Sherwood AA: An anterior approach to scoliosis: a preliminary report. Clin Orthop 1969;62:192.
76. Eckstein HB, Vora RM: Spinal osteotomy for severe kyphosis in children with myelomeningocele. J Bone Joint Surg 1972;54-B:328.
77. Edvardsen P: Physeo-epiphyseal injuries of lower extremities in myelomeningocele. Acta Orthop Scand 1972;43:550.
78. Eyring EJ, Wanken JJ, Sayers MP: Spine ostectomy for kyphosis in myelomeningocele. Clin Orthop 1972;88:24.
79. Feiwell E: Surgery of the hip in myelomeningocele as related to adult goals. Clin Orthop 1980:87.
80. Feiwell E, Sakai D, Blatt T: The effect of hip reduction on function in patients with myelomeningocele: potential gains and hazards of surgical treatment. J Bone Joint Surg 1978;60-A:169.
81. Feuchtbaum LB, Currier RJ, Riggle S, et al: Neural tube defect prevalence in California (1990–1994): eliciting patterns by type of defect and maternal race/ethnicity. Genet Test 1999;3:265.
82. Fiala TG, Buchman SR, Muraszko KM: Use of lumbar periosteal turnover flaps in myelomeningocele closure. Neurosurgery 1996; 39:522.
83. Forman R, Singal N, Perelman V, et al: Folic acid and prevention of neural tube defects: a study of Canadian mothers of infants with spina bifida. Clin Invest Med 1996;19:195.
84. Franks CA, Palisano RJ, Darbee JC: The effect of walking with an assistive device and using a wheelchair on school performance in students with myelomeningocele. Phys Ther 1991;71:570.
85. Fraser RK, Bourke HM, Broughton NS, et al: Unilateral dislocation of the hip in spina bifida: a long-term follow-up. J Bone Joint Surg 1995;77-B:615.
86. Fraser RK, Hoffman EB: Calcaneus deformity in the ambulant patient with myelomeningocele. J Bone Joint Surg 1991;73-B:994.
87. Fraser RK, Hoffman EB, Sparks LT, et al: The unstable hip and mid-lumbar myelomeningocele. J Bone Joint Surg 1992;74-B:143.
88. Fraser RK, Menelaus MB: The management of tibial torsion in patients with spina bifida. J Bone Joint Surg 1993;75-B:495.
89. Frawley PA, Broughton NS, Menelaus MB: Anterior release for fixed flexion deformity of the hip in spina bifida. J Bone Joint Surg 1996;78-B:299.
90. Frawley PA, Broughton NS, Menelaus MB: Incidence and type of hindfoot deformities in patients with low-level spina bifida. J Pediatr Orthop 1998;18:312.
91. Freeman JM: Practical Management of Meningomyelocele. Baltimore, University Park Press, 1974.
92. Fromm B, Carstens C, Niethard FU, et al: Aortography in children with myelomeningocele and lumbar kyphosis. J Bone Joint Surg 1992;74-B:691.
93. Furman L, Mortimer JC: Menarche and menstrual function in patients with myelomeningocele. Dev Med Child Neurol 1994;36:910.
94. Gardner WJ: Myelomeningocele, the results of rupture of the embryonic neural tube. Cleve Clin Q 1960;27:88.
95. Georgiadis GM, Aronson DD: Posterior transfer of the anterior tibial tendon in children who have a myelomeningocele [published erratum appears in J Bone Joint Surg 1990;72-A:792]. J Bone Joint Surg 1990;72-A:392.
96. Gerritsma-Bleeker CL, Heeg M, Vos-Niel H: Ambulation with the reciprocating-gait orthosis: experience in 15 children with myelomeningocele or paraplegia. Acta Orthop Scand 1997;68:470.
97. Glancy J: A dynamic orthotic system for young meningomyeloceles: a preliminary report. Orthot Prosthet 1976;30:3.
98. Greene SA, Frank M, Zachmann M, et al: Growth and sexual development in children with meningomyelocele. Eur J Pediatr 1985;144:146.
99. Gullestad HP, Bretteville G, Lundar T, et al: Tissue expansion for the treatment of myelomeningocele: case report. Scand J Plast Reconstr Surg Hand Surg 1993;27:149.
100. Hahn YS: Open myelomeningocele. Neurosurg Clin North Am 1995; 6:231.
101. Hall JE, Poitras B: The management of kyphosis in patients with myelomeningocele. Clin Orthop 1977;128:33.
102. Hall PV, Campbell RL, Kalsbeck JE: Meningomyelocele and progressive hydromyelia: progressive paresis in myelodysplasia. J Neurosurg 1975;43:457.
103. Heeg M, Broughton NS, Menelaus MB: Bilateral dislocation of the hip in spina bifida: a long-term follow-up study. J Pediatr Orthop 1998;18:434.
104. Heinz ER, Rosenbaum AE, Scarff TB, et al: Tethered spinal cord following meningomyelocele repair. Radiology 1979;131:153.
105. Hendricks KA, Simpson JS, Larsen RD: Neural tube defects along the Texas-Mexico border, 1993–1995. Am J Epidemiol 1999;149:1119.
106. Herman JM, McLone DG, Storrs BB, et al: Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord: presentation, management and outcome. Pediatr Neurosurg 1993;19:243.
107. Heydemann JS, Gillespie R: Management of myelomeningocele kyphosis in the older child by kyphectomy and segmental spinal instrumentation. Spine 1987;12:37.
108. Hoffer MM, Feiwell E, Perry R, et al: Functional ambulation in patients with myelomeningocele. J Bone Joint Surg 1973;55-A:137.
109. Holinger PC, Holinger LD, Reichert TJ, et al: Respiratory obstruction and apnea in infants with bilateral abductor vocal cord paralysis, meningomyelocele, hydrocephalus, and Arnold-Chiari malformation. J Pediatr 1978;92:368.
110. Holmbeck GN, Faier-Routman J: Spinal lesion level, shunt status, family relationships, and psychosocial adjustment in children and adolescents with spina bifida myelomeningocele. J Pediatr Psychol 1995;20:817.
111. Holmbeck GN, Gorey-Ferguson L, Hudson T, et al: Maternal, paternal, and marital functioning in families of preadolescents with spina bifida. J Pediatr Psychol 1997;22:167.
112. Huang TJ, Lubicky JP: Kyphectomy and segmental spinal instrumentation in young children with myelomeningocele kyphosis. J Formos Med Assoc 1994;93:503.
113. Huff CW, Ramsey PL: Myelodysplasia: the influence of the quadriceps and hip abductor muscles on ambulatory function and stability of the hip. J Bone Joint Surg 1978;60-A:432.
114. Hullin MG, Robb JE, Loudon IR: Ankle-foot orthosis function in low-level myelomeningocele. J Pediatr Orthop 1992;12:518.
115. Ingraham FD, Swam H: Spina bifida and cranium bifida. I. A survey of five hundred forty-six cases. N Engl J Med 1943;228:559.
116. Jackson RD, Padgett TS, Donovan MM: Iliopsoas muscle transfer in myelodysplasia. J Bone Joint Surg 1979;61-A:40.
117. Jackson RD, Padgett TS, Donovan MM: Posterior iliopsoas transfer in myelodysplasia. J Bone Joint Surg 1979;61-A:40.
118. James CC: Fractures of the lower limbs in spina bifida cystica: a survey of 44 fractures in 122 children. Dev Med Child Neurol Suppl 1970;22:88.
119. Jansen J, Taudorf K, Pedersen H, et al: Upper extremity function in spina bifida. Childs Nerv Syst 1991;7:67.
120. Kadic MA, Verbout AJ: Treatment of severe kyphosis in myelomeningocele by segmental spinal instrumentation with Luque rods. Acta Orthop Belg 1991;57:45.
121. Kalucy M, Bower C, Stanley F: School-aged children with spina bifida in Western Australia: parental perspectives on functional outcome. Dev Med Child Neurol 1996;38:325.
122. Karol LA: Orthopedic management in myelomeningocele. Neurosurg Clin North Am 1995;6:259.
123. Karol LA, Richards BS, Prejean E, et al: Hemodynamic instability of myelomeningocele patients during anterior spinal surgery. Dev Med Child Neurol 1993;35:261.
124. Kaufman AM, Ritchey ML, Roberts AC, et al: Decreased bladder

compliance in patients with myelomeningocele treated with radiological observation. J Urol 1996;156:2031.
125. Kaufman BA, Terbrock A, Winters N, et al: Disbanding a multidisciplinary clinic: effects on the health care of myelomeningocele patients. Pediatr Neurosurg 1994;21:36.
126. Kinsman SL, Doehring MC: The cost of preventable conditions in adults with spina bifida. Eur J Pediatr Surg 1996;6(suppl 1):17.
127. Knutson LM, Clark DE: Orthotic devices for ambulation in children with cerebral palsy and myelomeningocele. Phys Ther 1991;71:947.
128. Kumar SJ, Cowell HR, Townsend P: Physeal, metaphyseal, and diaphyseal injuries of the lower extremities in children with myelomeningocele. J Pediatr Orthop 1984;4:25.
129. Kupka J, Geddes N, Carroll NC: Comprehensive management in the child with spina bifida. Orthop Clin North Am 1978;9:97.
130. Laishram H, Kennedy R, Maroun F, et al: The impact of spina bifida on the medical services of Newfoundland and Labrador. J Pediatr Surg 1993;28:1098.
131. Lalonde F, Jarvis J: Congenital kyphosis in myelomeningocele: the effect of cordotomy on bladder function. J Bone Joint Surg 1999; 81-B:245.
132. Lanigan MW: Surgical repair of myelomeningocele. Ann Plast Surg 1993;31:514.
133. Lary JM, Edmonds LD: Prevalence of spina bifida at birth—United States, 1983–1990: a comparison of two surveillance systems. Morb Mortal Wkly Rep CDC Surveill Summ 1996;45:15.
134. Latcha CM, Freeling MC, Powell NJ: A comparison of the grip strength of children with myelomeningocele to that of children without disability. Am J Occup Ther 1993;47:498.
135. Laurence KM, James N, Miller MH, et al: Double-blind randomised controlled trial of folate treatment before conception to prevent recurrence of neural-tube defects. BMJ (Clin Res Ed) 1981;282:1509.
136. Lee EH, Carroll NC: Hip stability and ambulatory status in myelomeningocele. J Pediatr Orthop 1985;5:522.
137. Lie HR, Borjeson MC, Lagerkvist B, et al: Children with myelomeningocele: the impact of disability on family dynamics and social conditions. A Nordic study. Dev Med Child Neurol 1994;36:1000.
138. Lie HR, Lagergren J, Rasmussen F, et al: Bowel and bladder control of children with myelomeningocele: a Nordic study. Dev Med Child Neurol 1991;33:1053.
139. Lim R, Dias L, Vankoski S, et al: Valgus knee stress in lumbosacral myelomeningocele: a gait-analysis evaluation [see comments]. J Pediatr Orthop 1998;18:428.
140. Lindehall B, Claesson I, Hjalmas K, et al: Effect of clean intermittent catheterisation on radiological appearance of the upper urinary tract in children with myelomeningocele. Br J Urol 1991;67:415.
141. Lindehall B, Moller A, Hjalmas K, et al: Long-term intermittent catheterization: the experience of teenagers and young adults with myelomeningocele. J Urol 1994;152:187.
142. Lindseth RE: Myelomeningocele spine. In Weinstein SL (ed): The Pediatric Spine: Principles and Practice, p 1043. New York, Raven Press, 1994.
143. Lindseth RE, Stelzer L Jr: Vertebral excision for kyphosis in children with myelomeningocele. J Bone Joint Surg 1979;61-A:699.
144. Lintner SA, Lindseth RE: Kyphotic deformity in patients who have a myelomeningocele: operative treatment and long-term follow-up. J Bone Joint Surg 1994;76-A:1301.
145. Liptak GS, Shurtleff DB, Bloss JW, et al: Mobility aids for children with high-level myelomeningocele: parapodium versus wheelchair. Dev Med Child Neurol 1992;34:787.
146. Lock TR, Aronson DD: Fractures in patients who have myelomeningocele. J Bone Joint Surg 1989;71-A:1153.
147. Loder RT, Shapiro P, Towbin R, et al: Aortic anatomy in children with myelomeningocele and congenital lumbar kyphosis. J Pediatr Orthop 1991;11:31.
148. Loomis JW, Javornisky JG, Monahan JJ, et al: Relations between family environment and adjustment outcomes in young adults with spina bifida. Dev Med Child Neurol 1997;39:620.
149. Lorber J: Incidence and epidemiology of myelomeningocele. Clin Orthop 1966;45:81.
150. Lorber J: Results of treatment of myelomeningocele: an analysis of 524 unselected cases, with special reference to possible selection for treatment. Dev Med Child Neurol 1971;13:279.
151. Lorber J: Spina bifida cystica: results of treatment of 270 consecutive cases with criteria for selection for the future. Arch Dis Child 1972;47:854.
152. Lorber J: Selective treatment of myelomeningocele: to treat or not to treat? Pediatrics 1974;53:307.
153. Lorber J, Levick K: Spina bifida cystica: incidence of spina bifida occulta in parents and in controls. Arch Dis Child 1967;42:171.
154. Lorber J, Salfield SA: Results of selective treatment of spina bifida cystica. Arch Dis Child 1981;56:822.
155. Lough LK, Nielsen DH: Ambulation of children with myelomeningocele: parapodium versus parapodium with Orlau swivel modification. Dev Med Child Neurol 1986;28:489.
156. Lowe G, Menelaus M: The surgical management of kyphosis in older children with myelomeningocele. J Bone Joint Surg 1978;60-B:40.
157. MacMahon B, Pugh TF, Ingalls TD: Anencephalus, spina bifida, and hydrocephalus. Br J Soc Med 1953;7:211.
158. Maguire CD, Winter RB, Mayfield JK, et al: Hemimyelodysplasia: a report of 10 cases. J Pediatr Orthop 1982;2:9.
159. Malhotra D, Puri R, Owen R: Valgus deformity of the ankle in children with spina bifida aperta. J Bone Joint Surg 1984;66-B:381.
160. Malone PS, Wheeler RA, Williams JE: Continence in patients with spina bifida: long term results. Arch Dis Child 1994;70:107.
161. Mannor DA, Weinstein SL, Dietz FR: Long-term follow-up of Chiari pelvic osteotomy in myelomeningocele. J Pediatr Orthop 1996;16:769.
162. Marshall PD, Broughton NS, Menelaus MB, et al: Surgical release of knee flexion contractures in myelomeningocele. J Bone Joint Surg 1996;78-B:912.
163. Martin J Jr, Kumar SJ, Guille JT, et al: Congenital kyphosis in myelomeningocele: results following operative and nonoperative treatment. J Pediatr Orthop 1994;14:323.
164. Masters CL: Pathogenesis of the Arnold-Chiari malformation: the significance of hydrocephalus and aqueduct stenosis. J Neuropathol Exp Neurol 1978;37:56.
165. Maynard MJ, Weiner LS, Burke SW: Neuropathic foot ulceration in patients with myelodysplasia. J Pediatr Orthop 1992;12:786.
166. Mazon A, Nieto A, Estornell F, et al: Factors that influence the presence of symptoms caused by latex allergy in children with spina bifida. J Allergy Clin Immunol 1997;99:600.
167. Mazur J, Menelaus MB, Dickens DR, et al: Efficacy of surgical management for scoliosis in myelomeningocele: correction of deformity and alteration of functional status. J Pediatr Orthop 1986;6:568.
168. Mazur JM, Menelaus MB: Neurologic status of spina bifida patients and the orthopedic surgeon. Clin Orthop 1991;264:54.
169. Mazur JM, Menelaus MB, Hudson I, et al: Hand function in patients with spina bifida cystica. J Pediatr Orthop 1986;6:442.
170. Mazur JM, Shurtleff D, Menelaus M, et al: Orthopedic management of high-level spina bifida: early walking compared with early use of a wheelchair. J Bone Joint Surg 1989;71-A:56.
171. Mazur JM, Sienko-Thomas S, Wright N, et al: Swing-through vs. reciprocating gait patterns in patients with thoracic-level spina bifida. Z Kinderchir 1990;45(suppl 1):23.
172. Mazur JM, Stillwell A, Menelaus M: The significance of spasticity in the upper and lower limbs in myelomeningocele. J Bone Joint Surg 1986;68-B:213.
173. McCall RE: Modified Luque instrumentation after myelomeningocele kyphectomy. Spine 1998;23:1406.
174. McCall RE, Schmidt WT: Clinical experience with the reciprocal gait orthosis in myelodysplasia. J Pediatr Orthop 1986;6:157.
175. McCormick MC, Charney EB, Stemmler MM: Assessing the impact of a child with spina bifida on the family. Dev Med Child Neurol 1986;28:53.
176. McDonald CM, Jaffe KM, Mosca VS, et al: Ambulatory outcome of children with myelomeningocele: effect of lower-extremity muscle strength [see comments]. Dev Med Child Neurol 1991;33:482.
177. McDonald CM, Jaffe KM, Shurtleff DB, et al: Modifications to the traditional description of neurosegmental innervation in myelomeningocele [see comments]. Dev Med Child Neurol 1991;33:473.
178. McKibbin B: The use of splintage in the management of paralytic dislocation of the hip in spina bifida cystica. J Bone Joint Surg 1973;55-B:163.
179. McLaughlin TP, Banta JV, Gahm NH, et al: Intraspinal rhizotomy and distal cordectomy in patients with myelomeningocele. J Bone Joint Surg [Am] 1986;68:88.
180. McLone DG: Care of the neonate with a myelomeningocele. Neurosurg Clin N Am 1998;9:111.
181. McLone DG, Dias MS: Complications of myelomeningocele closure. Pediatr Neurosurg 1991;17:267.
182. McLone DG, Herman JM, Gabrieli AP, et al: Tethered cord as a cause

of scoliosis in children with a myelomeningocele. Pediatr Neurosurg 1990;16:8.
183. McMaster MJ: Anterior and posterior instrumentation and fusion of thoracolumbar scoliosis due to myelomeningocele. J Bone Joint Surg 1987;69-B:20.
184. McMaster MJ: The long-term results of kyphectomy and spinal stabilization in children with myelomeningocele. Spine 1988;13:417.
185. McMaster WC, Silber I: An urological complication of Dwyer instrumentation. J Bone Joint Surg 1975;57-A:710.
186. Menelaus MB: Dislocation and deformity of the hip in children with spina bifida cystica. J Bone Joint Surg 1969;51-B:238.
187. Menelaus MB: The hip in myelomeningocele: management directed towards a minimum number of operations and a minimum period of immobilization. J Bone Joint Surg 1976;58-B:448.
188. Menelaus MB: Orthopaedic management of children with myelomeningocele: a plea for realistic goals. Dev Med Child Neurol Suppl 1976:3.
189. Menelaus MB, Broughton NS: Dislocation of the hip in myelomeningocele: the McKay hip stabilization [letter; comment]. J Bone Joint Surg 1997;79-A:1750.
190. Menzies RG, Parkin JM, Hey EN: Prognosis for babies with meningomyelocele and high lumbar paraplegia at birth. Lancet 1985;2:993.
191. Meuli M, Meuli-Simmen C, Hutchins GM, et al: The spinal cord lesion in human fetuses with myelomeningocele: implications for fetal surgery. J Pediatr Surg 1997;32:448.
192. Meuli M, Meuli-Simmen C, Yingling CD, et al: In utero repair of experimental myelomeningocele saves neurological function at birth. J Pediatr Surg 1996;31:397.
193. Meuli-Simmen C, Meuli M, Adzick NS, et al: Latissimus dorsi flap procedures to cover myelomeningocele in utero: a feasibility study in human fetuses. J Pediatr Surg 1997;32:1154.
194. Mevorach RA, Bogaert GA, Baskin LS, et al: Lower urinary tract function in ambulatory children with spina bifida. Br J Urol 1996; 77:593.
195. Miller PD, Pollack IF, Pang D, et al: Comparison of simultaneous versus delayed ventriculoperitoneal shunt insertion in children undergoing myelomeningocele repair. J Child Neurol 1996;11:370.
196. Mills JL, Rhoads GG, Simpson JL, et al: The absence of a relation between the periconceptional use of vitamins and neural-tube defects. National Institute of Child Health and Human Development Neural Tube Defects Study Group [see comments]. N Engl J Med 1989; 321:430.
197. Milunsky A, Jick H, Jick SS, et al: Multivitamin/folic acid supplementation in early pregnancy reduces the prevalence of neural tube defects [see comments]. JAMA 1989;262:2847.
198. Mintz LJ, Sarwark JF, Dias LS, et al: The natural history of congenital kyphosis in myelomeningocele: a review of 51 children. Spine 1991; 16:S348.
199. Morgagni JB: The Seats and Causes of Disease Investigated by Anatomy. London, A Millar & J Caldwell, 1769.
200. MRC Vitamin Study Research Group: Prevention of neural tube defects: results of the Medical Research Council Vitamin Study. Lancet 1991;338:131.
201. Muen WJ, Bannister CM: Hand function in subjects with spina bifida. Eur J Pediatr Surg 1997;7 Suppl 1:18.
202. Mulinare J, Cordero JF, Erickson JD, et al: Periconceptional use of multivitamins and the occurrence of neural tube defects [see comments]. JAMA 1988;260:3141.
203. Muller EB, Nordwall A: Brace treatment of scoliosis in children with myelomeningocele. Spine 1994;19:151.
204. Muller EB, Nordwall A: Prevalence of scoliosis in children with myelomeningocele in western Sweden. Spine 1992;17:1097.
205. Muller EB, Nordwall A, Oden A: Progression of scoliosis in children with myelomeningocele. Spine 1994;19:147.
206. Muller EB, Nordwall A, von Wendt L: Influence of surgical treatment of scoliosis in children with spina bifida on ambulation and motoric skills. Acta Paediatr 1992;81:173.
207. Mustard WT: A follow-up study of iliopsoas transfer for hip instability. J Bone Joint Surg 1959;41-B:289.
208. Mustard WT: Iliopsoas transfer for weakness of the hip abductors. J Bone Joint Surg 1952;34-A:647.
209. Naik DR, Lendon RG, Barson AJ: A radiological study of vertebral and rib malformations in children with myelomeningocele. Clin Radiol 1978;29:427.
210. Nielsen HH: A longitudinal study of the psychological aspects of myelomeningocele. Scand J Psychol 1980;21:45.
211. Nogami H, Ingalls TH: Pathogenesis of spinal malformations induced in the embryos of mice. J Bone Joint Surg 1967;49-A:1551.
212. O'Connell DG, Barnhart R, Parks L: Muscular endurance and wheelchair propulsion in children with cerebral palsy or myelomeningocele. Arch Phys Med Rehabil 1992;73:709.
213. Osebold WR, Mayfield JK, Winter RB, et al: Surgical treatment of paralytic scoliosis associated with myelomeningocele. J Bone Joint Surg 1982;64-A:841.
214. Oyewole A, Adeloye A, Adeyokunnu AA: Psychosocial and cultural factors associated with the management of spina bifida cystica in Nigeria. Dev Med Child Neurol 1985;27:498.
215. Paget DH: Spina bifida and embryonic neuroschisis: a causal relationship. Definition of postnatal confirmations involving a bifid spine. Johns Hopkins Med J 1968;128:233.
216. Park BK, Song HR, Vankoski SJ, et al: Gait electromyography in children with myelomeningocele at the sacral level. Arch Phys Med Rehabil 1997;78:471.
217. Parsch K: Origin and treatment of fractures in spina bifida. Eur J Pediatr Surg 1991;1:298.
218. Patten BM: Overgrowth of the neural tube in young embryos. Anat Rec 1952;113:381.
219. Peeker R, Damber JE, Hjalmas K, et al: The urological fate of young adults with myelomeningocele: a three decade follow-up study. Eur Urol 1997;32:213.
220. Phillips DL, Field RE, Broughton NS, et al: Reciprocating orthoses for children with myelomeningocele: a comparison of two types. J Bone Joint Surg 1995;77-B:110.
221. Phillips DP, Lindseth RE: Ambulation after transfer of adductors, external oblique, and tensor fascia lata in myelomeningocele. J Pediatr Orthop 1992;12:712.
222. Piggott H: The natural history of scoliosis in myelodysplasia. J Bone Joint Surg 1980;62-B:54.
223. Proos LA, Dahl M, Ahlsten G, et al: Increased perinatal intracranial pressure and prediction of early puberty in girls with myelomeningocele. Arch Dis Child 1996;75:42.
224. Quan A, Adams R, Ekmark E, et al: Bone mineral density in children with myelomeningocele. Pediatrics 1998;102:E34.
225. Reigel DH, Tchernoukha K, Bazmi B, et al: Change in spinal curvature following release of tethered spinal cord associated with spina bifida. Pediatr Neurosurg 1994;20:30.
226. Renwick JH: Hypothesis: anencephaly and spina bifida are usually preventable by avoidance of a specific but unidentified substance present in certain potato tubers. Br J Prev Soc Med 1972;26:67.
227. Rodrigues RC, Dias LS: Calcaneus deformity in spina bifida: results of anterolateral release. J Pediatr Orthop 1992;12:461.
228. Rosano A, Smithells D, Cacciani L, et al: Time trends in neural tube defects prevalence in relation to preventive strategies: an international study. J Epidemiol Community Health 1999;53:630.
229. Rose GK, Sankarankutty M, Stallard J: A clinical review of the orthotic treatment of myelomeningocele patients. J Bone Joint Surg 1983; 65-A:242.
230. Rüdeberg A, Donati F, Kaiser G: Psychosocial aspects in the treatment of children with myelomeningocele: an assessment after a decade. Eur J Pediatr 1995;154:S85.
231. Sandhu PS, Broughton NS, Menelaus MB: Tenotomy of the ligamentum patellae in spina bifida: management of limited flexion range at the knee. J Bone Joint Surg 1995;77-B:832.
232. Sarwark JF: Spina bifida. Pediatr Clin North Am 1996;43:1151.
233. Sarwark JF, Weber DT, Gabrieli AP, et al: Tethered cord syndrome in low motor level children with myelomeningocele. Pediatr Neurosurg 1996;25:295.
234. Savini R, Cervellati S, Bettini N, et al: Surgical treatment of vertebral deformity due to myelomeningocele. Ital J Orthop Traumatol 1991; 17:55.
235. Sawyer SM, Collins N, Bryan D, et al: Young people with spina bifida: transfer from paediatric to adult health care. J Paediatr Child Health 1998;34:414.
236. Schiltenwolf M, Carstens C, Rohwedder J, et al: Results of orthotic treatment in children with myelomeningocele. Eur J Pediatr Surg 1991;1(suppl 1):50.
237. Schopler SA, Menelaus MB: Significance of the strength of the quadriceps muscles in children with myelomeningocele. J Pediatr Orthop 1987;7:507.

238. Segal LS, Mann DC, Feiwell E, et al: Equinovarus deformity in arthrogryposis and myelomeningocele: evaluation of primary talectomy. Foot Ankle 1989;10:12.
239. Seidel SB, Gardner PM, Howard PS: Soft-tissue coverage of the neural elements after myelomeningocele repair. Ann Plast Surg 1996;37:310.
240. Selber P, Dias L: Sacral-level myelomeningocele: long-term outcome in adults [see comments]. J Pediatr Orthop 1998;18:423.
241. Sharrard WJ: Long-term follow-up of posterior iliopsoas transplantation for paralytic dislocation of the hip. Dev Med Child Neurol Suppl 1969;20:96.
242. Sharrard WJ: Spinal osteotomy for congenital kyphosis in myelomeningocele. J Bone Joint Surg 1968;50-B:466.
243. Sharrard WJ, Drennan JC: Osteotomy-excision of the spine for lumbar kyphosis in older children with myelomeningocele. J Bone Joint Surg 1972;54-B:50.
244. Sharrard WJ, Grosfield I: The management of deformity and paralysis of the foot in myelomeningocele. J Bone Joint Surg 1968;50-B:456.
245. Sharrard WJ, Webb J: Supra-malleolar wedge osteotomy of the tibia in children with myelomeningocele. J Bone Joint Surg 1974;56-B:458.
246. Sharrard WJW: Congenital paralytic dislocation of the hip in children with myelomeningocele. J Bone Joint Surg 1959;41-B:622.
247. Sharrard WJW: Neuromotor Evaluation of the Newborn. St. Louis, CV Mosby Co, 1972.
248. Sharrard WJW: Posterior iliopsoas transplantation in the treatment of paralytic dislocation of the hip. J Bone Joint Surg 1964;46-B:426.
249. Sherk HH, Ames MD: Functional results of iliopsoas transfer in myelomeningocele hip dislocations. Clin Orthop 1978;137:181.
250. Sherk HH, Ames MD: Talectomy in the treatment of the myelomeningocele patient. Clin Orthop 1975;110:218.
251. Sherk HH, Marchinski LJ, Clancy M, et al: Ground reaction forces on the plantar surface of the foot after talectomy in the myelomeningocele. J Pediatr Orthop 1989;9:269.
252. Sherk HH, Uppal GS, Lane G, et al: Treatment versus nontreatment of hip dislocations in ambulatory patients with myelomeningocele [see comments]. Dev Med Child Neurol 1991;33:491.
253. Sillen U, Hansson E, Hermansson G, et al: Development of the urodynamic pattern in infants with myelomeningocele. Br J Urol 1996; 78:596.
254. Smith ED: Spina bifida and the total care of spinal myelomeningocele. Springfield: Charles C. Thomas, 1965.
255. Smithells RW, Nevin NC, Seller MJ, et al: Further experience of vitamin supplementation for prevention of neural tube defect recurrences. Lancet 1983;1:1027.
256. Somerville EW: Paralytic dislocation of the hip. J Bone Joint Surg 1959;41-B:279.
257. Sriram K, Bobechko WP, Hall JE: Surgical management of spinal deformities in spina bifida. J Bone Joint Surg 1972;54-B:666.
258. Stark A, Saraste H: Anterior fusion insufficient for scoliosis in myelomeningocele. 8 children 2–6 years after the Zielke operation. Acta Orthop Scand 1993;64:22.
259. Steel HH, Adams DJ: Hyperlordosis caused by the lumboperitoneal shunt procedure for hydrocephalus. J Bone Joint Surg 1972;54-A:1537.
260. Stein SC, Feldman JG, Friedlander M, et al: Is myelomeningocele a disappearing disease? Pediatrics 1982;69:511.
261. Stellman-Ward G, Bannister CM, Lewis M: Assessing the needs of the adult with spina bifida. Eur J Pediatr Surg 1993;3(suppl 1):14.
262. Stevens PM, Belle RM: Screw epiphysiodesis for ankle valgus. J Pediatr Orthop 1997;17:9.
263. Stevens PM, Toomey E: Fibular-Achilles tenodesis for paralytic ankle valgus. J Pediatr Orthop 1988;8:169.
264. Stillwell A, Menelaus MB: Walking ability after transplantation of the iliopsoas: a long-term follow-up. J Bone Joint Surg 1984;66-B:656.
265. Stott NS, Zionts LE, Gronley JK, et al: Tibialis anterior transfer for calcaneal deformity: a postoperative gait analysis. J Pediatr Orthop 1996;16:792.
266. Surana RH, Quinn FM, Guiney EJ, et al: Are the selection criteria for the conservative management in spina bifida still applicable? Eur J Pediatr Surg 1991;1(suppl 1):35.
267. Swank M, Dias L: Myelomeningocele: a review of the orthopaedic aspects of 206 patients treated from birth with no selection criteria. Dev Med Child Neurol 1992;34:1047.
268. Swank M, Dias LS: Walking ability in spina bifida patients: a model for predicting future ambulatory status based on sitting balance and motor level [published erratum appears in J Pediatr Orthop 1995; 15:278]. J Pediatr Orthop 1994;14:715.
269. Taylor LJ: Excision of the proximal end of the femur for hip stiffness in myelomeningocele. J Bone Joint Surg 1986;68-B:75.
270. Thom H, Campbell AG, Farr V, et al: The impact of maternal serum alpha fetoprotein screening on open neural tube defect births in northeast Scotland. Prenat Diagn 1985;5:15.
271. Thomas LJ, Thompson TC, Straub LR: Transplantation of the external oblique muscle for abductor paralysis. J Bone Joint Surg 1955; 32-A:207.
272. Thomson JD, Ounpuu S, Davis RB, et al: The effects of ankle-foot orthoses on the ankle and knee in persons with myelomeningocele: an evaluation using three-dimensional gait analysis. J Pediatr Orthop 1999;19:27.
273. Toet M, van Gool J, Witkamp T, et al: Spina bifida aperta and the tethered cord syndrome. Eur J Pediatr Surg 1991;1(suppl 1):48.
274. Torode I, Godette G: Surgical correction of congenital kyphosis in myelomeningocele. J Pediatr Orthop 1995;15:202.
275. Tosi LL, Buck BD, Nason SS, et al: Dislocation of hip in myelomeningocele: the McKay hip stabilization [see comments]. J Bone Joint Surg 1996;78-A:664.
276. Tosi LL, Slater JE, Shaer C, et al: Latex allergy in spina bifida patients: prevalence and surgical implications. J Pediatr Orthop 1993;13:709.
277. Turner A: Hand function in children with myelomeningocele. J Bone Joint Surg 1985;67-B:268.
278. Turner JW, Cooper RR: Posterior transposition of tibialis anterior through the interosseous membrane. Clin Orthop 1971;79:71.
279. Uehling DT, Smith J, Meyer J, et al: Impact of an intermittent catheterization program on children with myelomeningocele. Pediatrics 1985;76:892.
280. Vankoski S, Moore C, Statler KD, et al: The influence of forearm crutches on pelvic and hip kinematics in children with myelomeningocele: don't throw away the crutches. Dev Med Child Neurol 1997; 39:614.
281. Vergel RG, Sanchez LR, Heredero BL: Primary prevention of neural tube defect with folic acid supplementation: Cuban experience. Prenat Diagn 1990;10:149.
282. von Recklinghausen F: Untersuchungen über der Spina bifida. Arch Pathol Anat 1886;105:243.
283. Walker G: The early management of varus feet in myelomeningocele. J Bone Joint Surg 1971;53-B:462.
284. Ward WT, Wenger DR, Roach JW: Surgical correction of myelomeningocele scoliosis: a critical appraisal of various spinal instrumentation systems. J Pediatr Orthop 1989;9:262.
285. Warner WC Jr, Fackler CD: Comparison of two instrumentation techniques in treatment of lumbar kyphosis in myelodysplasia. J Pediatr Orthop 1993;13:704.
286. Weisl H, Fairclough JA, Jones DG: Stabilisation of the hip in myelomeningocele. Comparison of posterior iliopsoas transfer and varus-rotation osteotomy. J Bone Joint Surg 1988;70-B:29.
287. Wenger DR, Jeffcoat BT, Herring JA: The guarded prognosis of physeal injury in paraplegic children. J Bone Joint Surg 1980;62-A:241.
288. Williams JJ, Graham GP, Dunne KB, et al: Late knee problems in myelomeningocele. J Pediatr Orthop 1993;13:701.
289. Wiltse LL: Valgus deformity of the ankle: a sequel to acquired or congenital abnormalities of the fibula. J Bone Joint Surg 1972; 54-A:595.
290. Wright JG, Menelaus MB, Broughton NS, et al: Lower extremity alignment in children with spina bifida. J Pediatr Orthop 1992;12:232.
291. Wright JG, Menelaus MB, Broughton NS, et al: Natural history of knee contractures in myelomeningocele. J Pediatr Orthop 1991; 11:725.
292. Wu HY, Baskin LS, Kogan BA: Neurogenic bladder dysfunction due to myelomeningocele: neonatal versus childhood treatment. J Urol 1997;157:2295.
293. Yen S, MacMahon B: Genetics of anencephaly and spina bifida. Lancet 1968;2:623.
294. Yngve DA, Douglas R, Roberts JM: The reciprocating gait orthosis in myelomeningocele. J Pediatr Orthop 1984;4:304.
295. Yngve DA, Lindseth RE: Effectiveness of muscle transfers in myelomeningocele hips measured by radiographic indices. J Pediatr Orthop 1982;2:121.
296. Zerin JM, McLaughlin K, Kerchner S: Latex allergy in patients with myelomeningocele presenting for imaging studies of the urinary tract. Pediatr Radiol 1996;26:450.
297. Zurmohle UM, Homann T, Schroeter C, et al: Psychosocial adjustment of children with spina bifida. J Child Neurol 1998;13:64.

ADDED REFERENCE

Myelomeningocele

1a. Rickham PP, Mawdsley T: The effect of early operation on the survival of spina bifida cystica. Dev Med Child Neurol 1966;suppl 11:20.

Other Forms of Spinal Dysraphism

The term spinal dysraphism refers to a complex group of developmental abnormalities of the spine and neural axis in which there is nerve tissue anomaly, usually combined with bony anomalies of the vertebral column. This category of developmental affection encompasses a wide spectrum of conditions: at one extreme is myelomeningocele, as presented in the previous section. Other, generally milder forms of spinal dysraphism to be described here include lipomeningocele, tethered filum terminale, diastematomyelia, spinal bifida occulta, and caudal regression syndrome (including sacral and lumbosacral agenesis).

Cutaneous lesions may be present in patients with more subtle forms of spinal dysraphism. Such lesions include midline hemangiomas, sacral dimples, and local hairy patches. Sacral dimples associated with spinal dysraphism are always in the midline; these dimples may be connected to a sinus tract (dermal sinus) or a fibrous band leading to bone or dura. The clinician should be highly suspicious of skin dimples proximal to the fifth lumbar level, particularly when associated with spina bifida occulta. The location of the cutaneous lesion does not always correlate with the level of intraspinal lesion.

In spinal dysraphism, *musculoskeletal deformity* may be absent or present at birth; it may manifest as talipes equinovarus, congenital convex pes valgus, cavus, or simple equinus. There may be shortening or atrophy of the lower limb or hip subluxation or dislocation. When an infant presents with lower extremity deformity, clinical assessment of the spine at least is required to rule out spinal dysraphism.

Neurologic deficit may manifest at any time during a child's growth and development. One foot may be smaller than the opposite side, or calf or thigh atrophy may be noted. *Sensory dysfunction* is variable and may be difficult to detect in an infant or young child. Loss of sensation usually manifests as skin ulceration or pressure areas. In spinal dysraphism there may be loss of tactile, hot-cold, vibratory, and position sense. *Bladder dysfunction* in the form of frequency, dribbling, or hypotonic bladder may be present.

The diagnostic evaluation of patients suspected of having any form of spinal dysraphism should include a thorough clinical examination of the neurologic and musculoskeletal systems and plain radiography of the entire spine. If suspicion is still present after this assessment, ultrasonographic evaluation of the spine can be carried out before age 3 months to look for spinal cord or other intrathecal anomalies.[18,21] MRI of the spine should be performed in most patients with suspected spinal dysraphism to define the nature and extent of the spinal cord and other intrathecal anomalies. CT of the lumbosacral spine can be helpful in assessing associated congenital vertebral anomalies.

LIPOMENINGOCELE

Lipomeningocele is a dysraphic condition of the spine that is characterized by incorporation of subcutaneous fat into the distal part of the spinal cord. The lesion is often skin-covered, but the posterior elements of the vertebral column frequently are defective. The neuropathways are often intact; the neurologic deficit is caused primarily by tethering of the spinal cord to the surrounding fixed structures. Weakness in one or more lower extremities or disturbances in bowel or bladder function may be present from birth or may develop over time.

Clinical Features. Most cases of lipomeningocele are asymptomatic during infancy. The only physical finding may be the presence of a lumbosacral soft tissue mass (Fig. 25–33). Alternatively, there may be a variable degree of muscle weakness, sensory changes, or lower extremity skeletal anomalies such as hip dislocation or subluxation, knee deformity, and clubfoot or other foot deformity. The MRI findings in a patient with lipomeningocele are shown in Figure 25–34.

Treatment. The natural history of patients with lipomeningocele is variable. The late development of neurologic deficit may be due either to a mass effect from the lipomatous tissue acting as an expansile tumor within the central canal compressing the conus or to tethering of the spinal cord to subcutaneous tissue or dura. Neurosurgical debulking is usually indicated when neurologic deficit is noted, and is definitely indicated when progressive neurologic deficit is present. Secondary tethering of the spinal cord as a consequence of the scarring produced by this procedure may occur in the growing child.

Orthopaedic treatment entails treating the lower extremity deformity and weakness as indicated by their severity and symptoms. The most common orthopaedic abnormality is an equinocavus foot deformity (Fig. 25–35). Treatment should be undertaken as described earlier in this chapter in the discussions of myelomeningocele and cavus foot. During

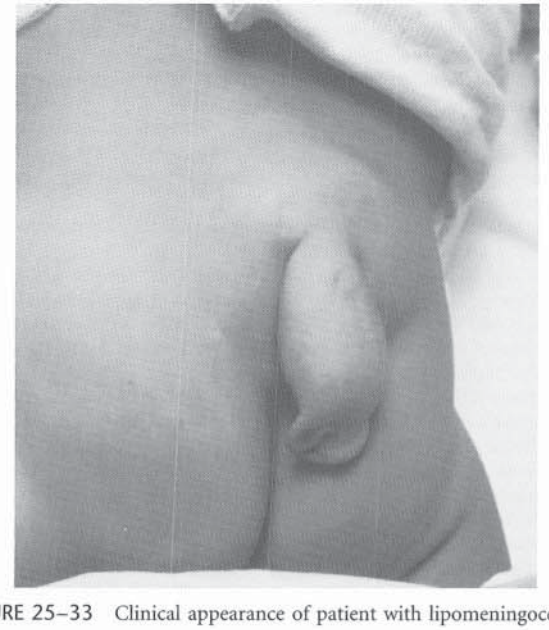


FIGURE 25–33 Clinical appearance of patient with lipomeningocele.

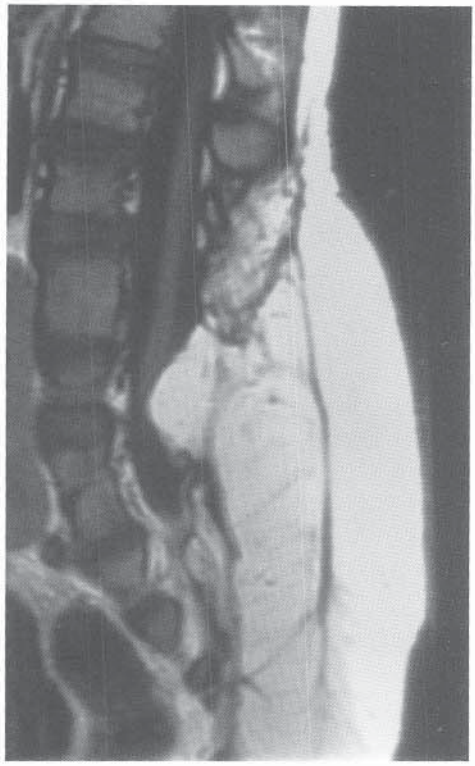


FIGURE 25–34 Appearance of lipomeningocele on MRI. Note lipomatous mass involving the conus medullaris.

growth, the patient should be periodically reassessed to screen for the development of associated scoliosis and to confirm stable neurologic function. In our experience, the more severely affected patients are for some reason more likely to develop Charcot joints, such as of the ankle (Fig. 25–36). These joints can be difficult to treat. Initial management entails protective bracing. Fusion may be attempted for symptomatic, deformed Charcot joints, but can be difficult to achieve.

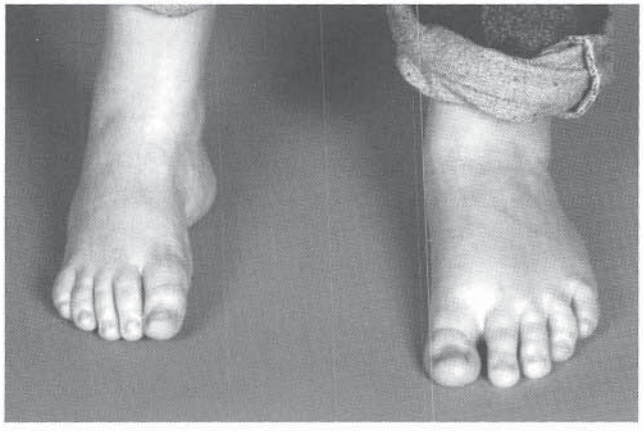


FIGURE 25–35 Patient presenting with a unilateral (equino) cavovarus foot deformity. Lower extremity weakness and back pain were present. MRI of the lumbar spine revealed a tethered cord secondary to a taut, thickened filum terminale.

TETHERED CORD (THICKENED FILUM TERMINALE)

Tethering of the spinal cord within the thecal sac preventing normal migration and maintenance of the conus to the level of the first or second lumbar vertebra[4] may occur in association with diastematomyelia (see below), lipomeningocele, a taut or thickened filum terminale, other congenital anomalies of the filum terminale, or secondary to scarring associated with intrathecal procedures such as closure of the myelomeningocele sac.[5,7,11,16,17,20,29] The radiographic hallmark of spinal cord tethering is the presence of the conus below its normal level of L1 or L2 after age 2 months (Fig. 25–37). When the cause is a thickened, taut filum terminale, this will be evident on MRI as well.

Affected patients may complain of back pain, perhaps radiating into the posterior thighs, lower extremity weakness, muscle tightness, particularly in the hamstrings, foot deformities, including equinus and equinocavus, and occasionally bowel or bladder problems. On examination, hemangioma, sacral dimpling, or hairy patches may be present. Treatment consists of neurosurgical release in the symptomatic patient. Most commonly, any associated orthopaedic deformities will persist and require treatment as indicated.

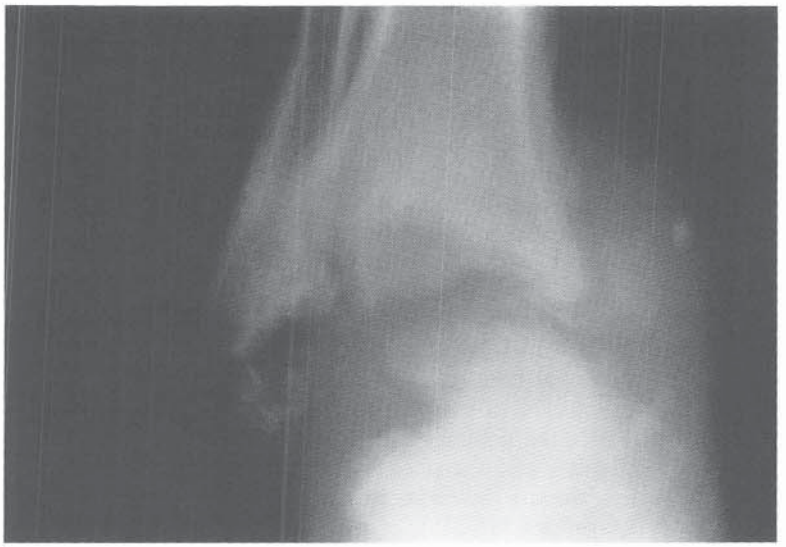
DIASTEMATOMYELIA

Diastematomyelia is a congenital malformation of the neural axis in which there is a sagittal division of the spinal cord or its intraspinal derivatives; often it is associated with a projection of an osseous, fibrocartilaginous, or fibrous spur that is attached anteriorly to one or more vertebral bodies and posteriorly to the dura (Figs. 25–38 and 25–39).[13,33] There may be an associated tethering of the spinal cord secondary to a thickened filum terminale.

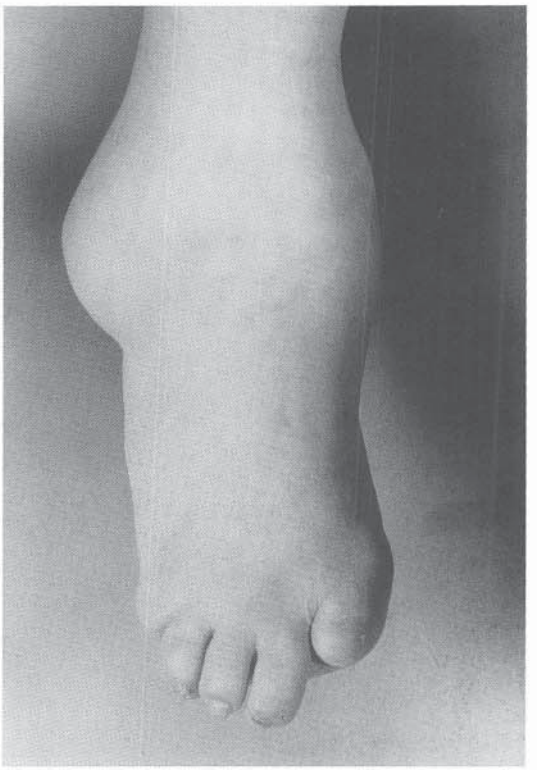
The pathogenesis of diastematomyelia is unknown. It appears that during the organization of the neural tube from the primitive neuroectoderm, aberrant mesodermal cells protrude into the neural tissue on its anterior surface instead of becoming arranged entirely around its periphery. They persist in this location, developing into a bony and dural septum. Associated multiple congenital anomalies of the vertebrae with some degree of incomplete spinal fusion are often present. The osseous septum transfixes the spinal cord or cauda equina, checkreining its normal ascent during growth of the vertebral column.

Clinical Picture. Abnormalities of motor function in the lower limbs are not ordinarily detected at birth. Hallmarks of the condition are the various types of skin defects that are found near the midline at the level of the lesion. The cutaneous abnormalities include abnormal tufts of hair, dimpling of the skin, ill-defined subcutaneous fatty tumors, and cutaneous vascular malformations. Back and lower extremity symptoms are highly variable and depend on the extent of interference with cord function.

Radiographic Findings. The hallmark of diastematomyelia on plain radiographs of the spine is a widening of the interpedicular distance in the region of the diastematomyelia itself. Often the posterior elements are incompletely formed in the region of the diastematomyelia. There may be associated congenital vertebral anomalies at or remote from the



A



B

FIGURE 25–36 Gross degenerative joint disease (Charcot joint) resulting from lack of protective sensation. **A,** Radiograph demonstrating significant destructive changes of the tibiotalar joint. This disorder occurs more commonly in patients with lipomeningocele than in patients with myelomeningocele. **B,** Clinical appearance. Note swelling of the ankle. Clinically, the joint is unstable, but the patient has only mild pain.

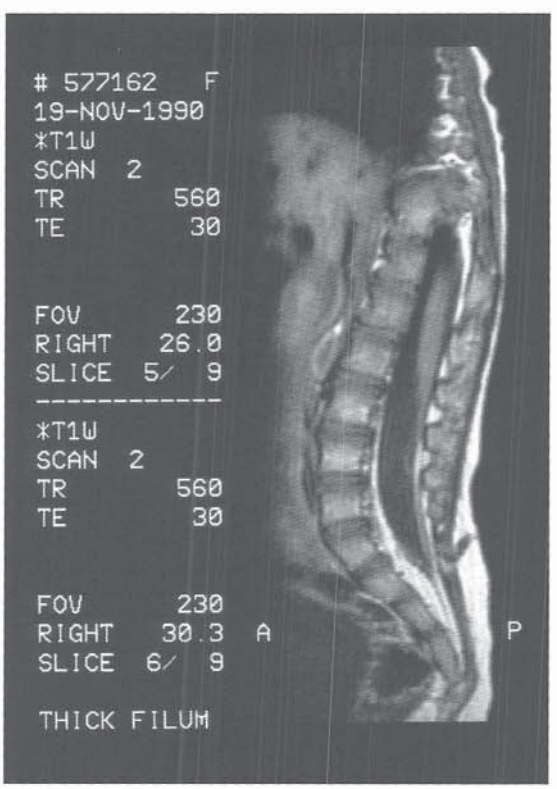


FIGURE 25–37 MRI of a patient with back pain and lower extremity weakness. The conus medullaris normally ends at L1 or L2 after 2 months of age. In this patient, the filum terminale is thickened, appearing to "tether" the conus medullaris distally.

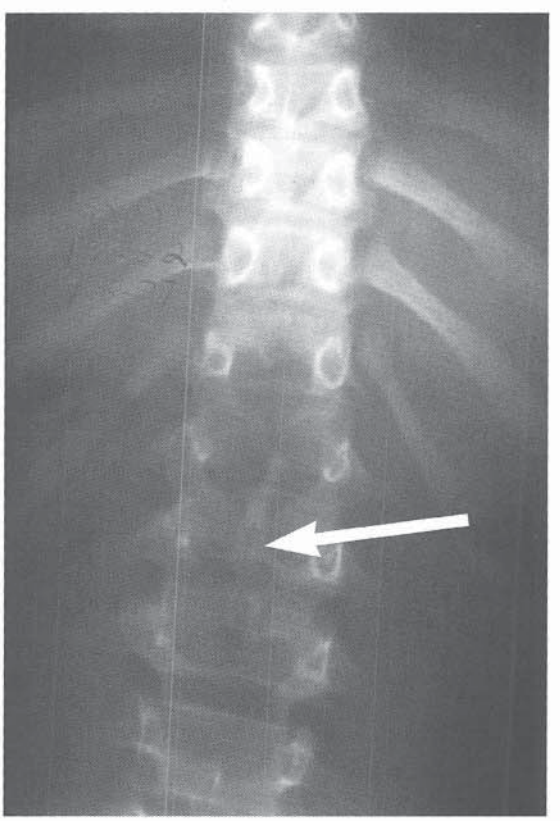


FIGURE 25–38 Plain radiographs of a patient with diastematomyelia in the thoracic spine. Note the interpedicular widening around the level of the diastematomyelia. The diastematomyelia spur itself is visible as a spicule of bone in the spinal canal (*arrow*).

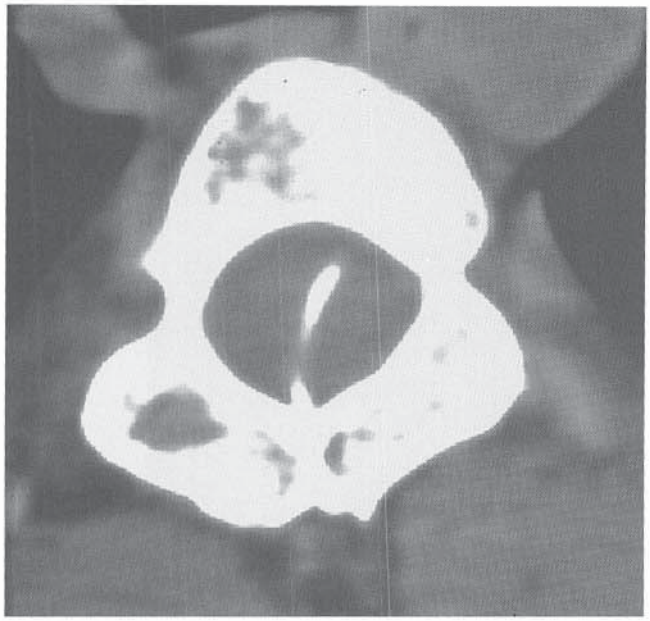


FIGURE 25–39 CT scan of a patient with diastematomyelia.

level of the diastematomyelia. CT and MRI will allow accurate localization and depiction of the lesion (Fig. 25–40).

Treatment. The purpose of surgery in diastematomyelia is to prevent progressive neurologic deficit. Patients in whom there are no symptoms, neurologic abnormalities, or orthopaedic physical findings may be carefully observed with serial examinations. Neurosurgical excision of the septum is recommended when there is a progressive neurologic deficit or the recent development of neurologic dysfunction. Neurosurgical resection of the lesion is also normally indicated when surgical correction of associated spinal deformity is planned, to prevent acute deterioration of spinal cord function during correction. Finally, in some patients, prophylactic excision may be warranted if the risk of development of neurologic dysfunction is deemed to be high.

SPINA BIFIDA OCCULTA

Failure of fusion of one or more of the vertebral arches is perhaps the most common congenital anomaly of the spinal column, and in the majority of individuals it is a normal variation. Spina bifida occulta is most commonly noted in the fifth lumbar and first sacral vertebrae; it may also be seen in the cervical, lower thoracic, and lower sacral spine of affected individuals.

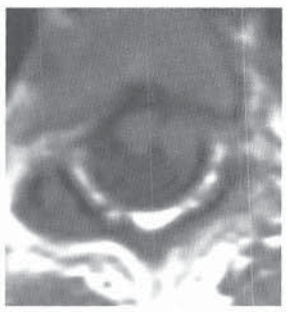


FIGURE 25–40 MRI of a patient with diastematomyelia. Note the splitting of the spinal cord ("diastematomyelia") around the osteocartilaginous spicule.

Clinical Features. The presence of spina bifida occulta radiographically is of no clinical significance in the majority of patients. The diagnosis is most typically made incidentally from radiographs of the spine obtained for some other purpose. Provided there are no symptoms or physical abnormalities in the spine or lower extremities of such patients, no further investigation or management is required (Fig. 25–41).

A very small percentage of individuals have an intrathecal anomaly such as lipoma or thickening of the filum terminale associated with the radiographic finding of spina bifida occulta. The physician should search for evidence of skin abnormality over the midline posterior spine and any lower extremity deformity or neurologic abnormality. If present, MRI evaluation of the spinal cord and consultation with a neurosurgeon should be obtained.

CAUDAL REGRESSION SYNDROME (SACRAL AND LUMBOSACRAL AGENESIS)

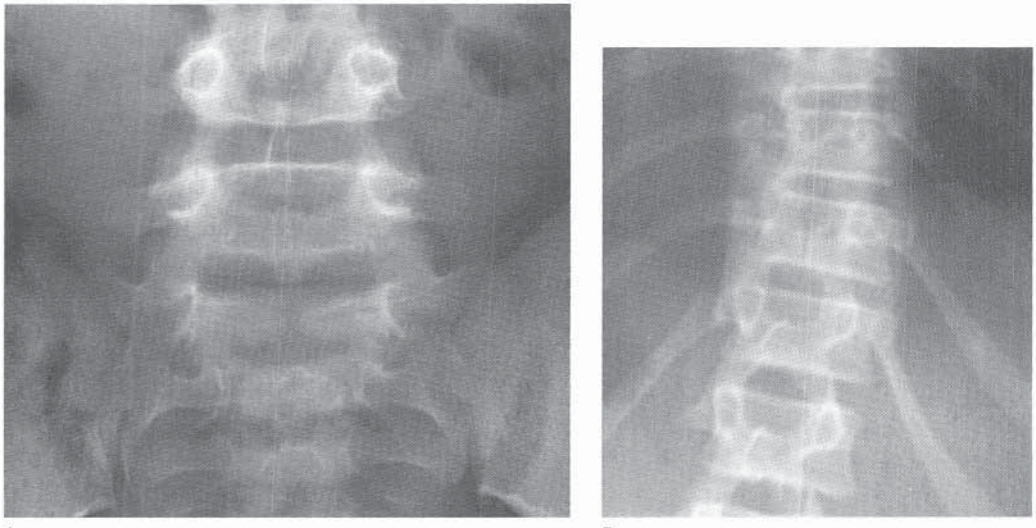
Caudal regression syndrome (lumbosacral or sacral agenesis) is a severe axial skeletal and neural deficiency characterized by absence of variable amounts of the sacrum and lumbar spine and associated neural elements. There are concomitant anomalies of the viscera, particularly of the genitourinary and lower gastrointestinal systems, with resultant impairment of bladder and bowel function.[1,6,8,14,22,23,25,30]

The failure of development ranges from mere absence of the lower coccygeal segment to complete aplasia of vertebrae below the 12th thoracic segment. Lesser degrees of involvement, such as absence of the lower coccygeal segments, often go unnoticed and are usually recognized fortuitously during unrelated radiographic studies, whereas the more extreme degrees of involvement may be incompatible with life.

The first known case was described in 1852, and since then a number of review articles and case reports have appeared in the literature.* The etiology of lumbosacral agenesis is unknown. A higher than normal incidence of diabetes mellitus in the mother has been noted repeatedly.[23] In a rare hereditary form of the condition, the location of the genetic defect has been determined.[19]

Pathologic Findings. The gross and histologic findings depend on the level of the lesion. In total absence of the lumbosacral spine, normal muscle tissue is replaced by large globules of soft, deep, yellow fat. Tendons are found as thin filaments, but have normal configuration. There is an abnormal nerve root pattern at the end of the spinal cord. The femoral vessels are very small. The femoral nerves are represented as gross fatty tissue adjacent to the small vessels. Afferent tracts are usually fairly well preserved, whereas efferent motor neuron pathways are impaired or missing. The spinal cord does not exhibit any lumbar enlargement or lumbosacral plexus and terminates at a higher level than usual, for example, at T7 when the vertebral column ends at the second lumbar segment; otherwise the anatomy of the spinal cord is normal above the lesion. Unlike in myelo-

* See references 1–3, 12, 14, 15, 22, 25, 26.



A B

FIGURE 25–41 Appearance of spina bifida occulta noted incidentally on spinal radiographs. If there are no complaints or physical abnormalities, further investigation is unnecessary. **A,** The most common level of spina bifida occulta is at L5 or S1. **B,** Spina bifida occulta of T11 noted incidentally on spinal radiographs taken to evaluate scoliosis. Spina bifida occulta may also be seen in the lower cervical spine.

meningocele, CNS deformities above the level of spinal cord lesion do not occur.

Classification. Caudal regression syndrome has been classified by Renshaw into four types.[26] *Type I* is either total or partial unilateral sacral agenesis (Fig. 25–42). *Type II* is partial sacral agenesis with a partial but bilaterally symmetric defect and a stable articulation between the ilia and a normal or hypoplastic first sacral vertebra (Fig. 25–43). This is the most common type. *Type III* is variable lumbar and total sacral agenesis, with the ilia articulating with the sides of the lowest vertebra present (Fig. 25–44). *Type IV* is variable lumbar and total sacral agenesis, the caudal end-plate of the lowest vertebra resting above either fused ilia or an iliac amphiarthrosis (Fig. 25–45).

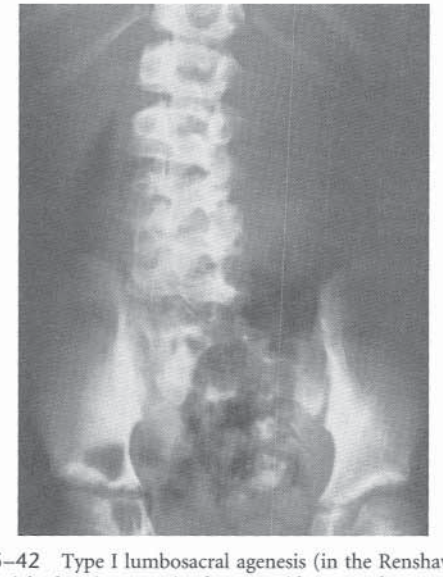


FIGURE 25–42 Type I lumbosacral agenesis (in the Renshaw classification). The right hemisacrum is absent, evident on the anteroposterior radiograph. (From Renshaw TS: Sacral agenesis. J Bone Joint Surg 1978;60-A:373.)

Clinical Findings. The appearance of the patient depends on the extent of spinal involvement and the degree of concomitant neurologic deficit.

TYPE I. In total or partial unilateral sacral agenesis the pelvic ring and lumbosacral junction are intact; therefore, the vertebropelvic articulation is usually, but not always, stable. The unilateral absence of the sacrum results in an oblique lumbosacral joint and lumbar scoliosis. The scoliosis ordinarily is not progressive and does not require orthotic or surgical treatment. Hips and knees are usually normal; there may, however, be a calcaneovarus deformity of the foot. There is sensory loss corresponding to the distribution of the involved sacral roots.

TYPE II. The vertebropelvic junction is stable unless there is associated myelomeningocele. In some patients with associated myelomeningocele, progressive kyphosis and paralytic scoliosis develop, requiring surgical stabilization to facilitate sitting. There may be associated congenital anomalies of the spine (such as hemivertebrae), which may cause progressive congenital scoliosis and require treatment by spinal fusion. There may be associated rib anomalies in the form of fusion of adjacent ribs or absence of ribs.

Motor paralysis is present and corresponds within one level with the vertebral defect. *Sensation* is usually intact. There may be anesthesia at S4 and distally. In patients with associated myelomeningocele the level of paralysis may be higher than the level of vertebral deficit, and the sensory loss may be more extreme. *Hip dislocation* occurs in type II agenesis and may be unilateral or bilateral. The pathogenesis of dislocation appears to be dynamic imbalance of muscles controlling the hip—absence or weakness of hip abductors-extensors versus normal motor strength of hip adductors-flexors. Foot deformities and flexion deformity of the knee

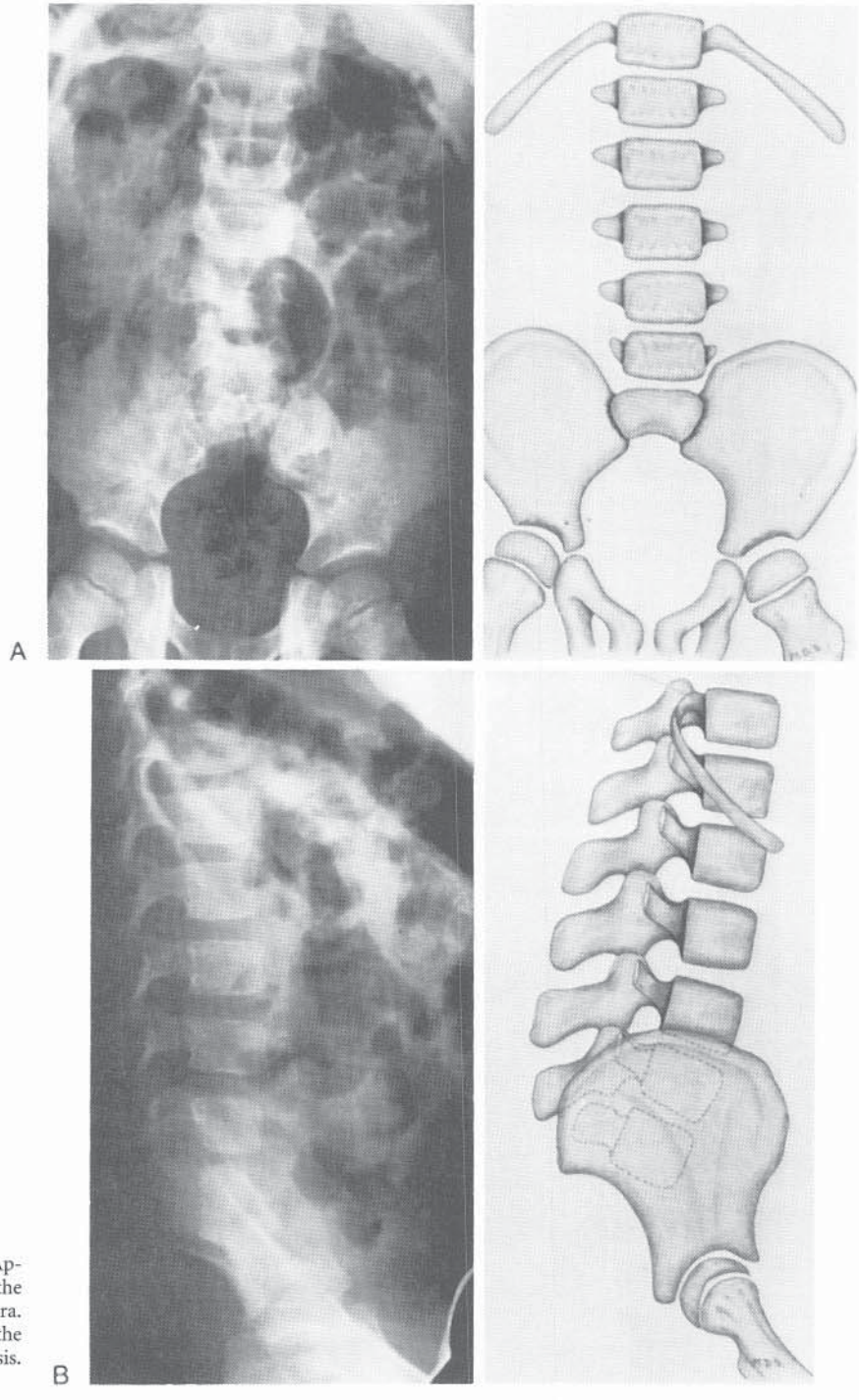


FIGURE 25–43 Type II lumbosacral agenesis. **A,** Appearance on AP radiographs. Note the absence of the lower sacrum and the hypoplastic first sacral vertebra. **B,** The lumbopelvic articulation is evidently stable on the lateral radiograph. (From Renshaw TS: Sacral agenesis. J Bone Joint Surg 1978;60-A:373.)

are usually not marked. Most patients with type II lumbosacral agenesis are ambulatory.

TYPE III. The lumbopelvic junction is relatively stable in this type despite absence of the sacrum and, in some, of L5. Progressive kyphosis and scoliosis may develop in these patients. The level of motor paralysis parallels the level of vertebral deficit within one segment. Sensation is characteristically intact at least down to the S4 nerve root level. In total absence of the sacrum, the buttocks are flattened, the intergluteal cleft is shortened, and there is dimpling of each buttock lateral to the cleft. The normal posterior convexity of the sacrococcygeal region is lost; on rectal examination the concavity of the sacrum and coccyx is absent. Hip dislocation, knee contracture, and foot deformity are common in type III agenesis and may require treatment. Patients with type III agenesis are unable to stand and walk without appropriate orthotic support or crutches.

TYPE IV. This type represents the classic, fully manifested form of lumbosacral agenesis. The patients are of short stature and have a characteristic cross-legged Buddha-like

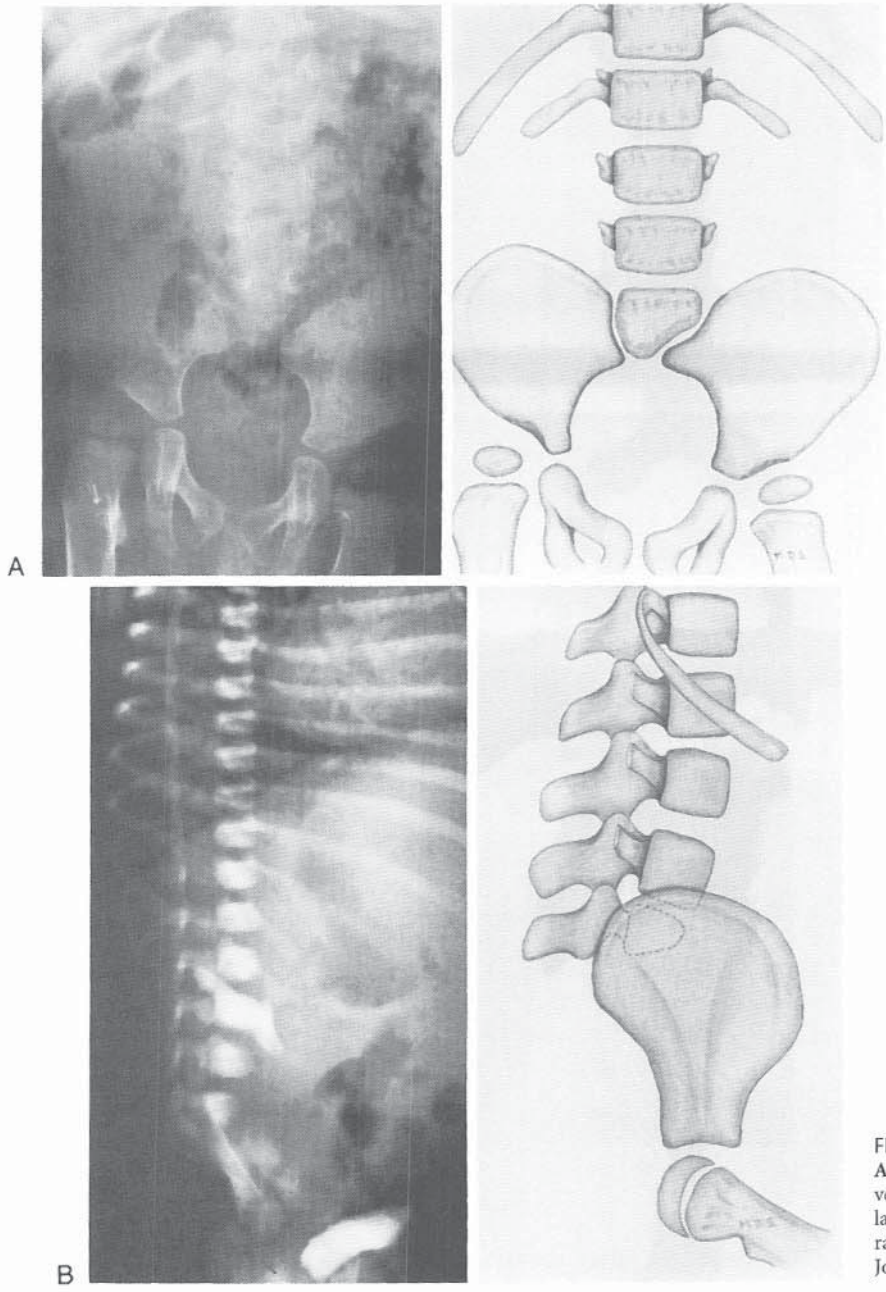


FIGURE 25–44 Type III lumbosacral agenesis. **A,** On the AP radiograph, the sacrum and lower lumbar vertebrae are absent. The third lumbar vertebra articulates with the pelvis. **B,** Appearance on the lateral radiograph. (From Renshaw TS: Sacral agenesis. J Bone Joint Surg 1978;60-A:373.)

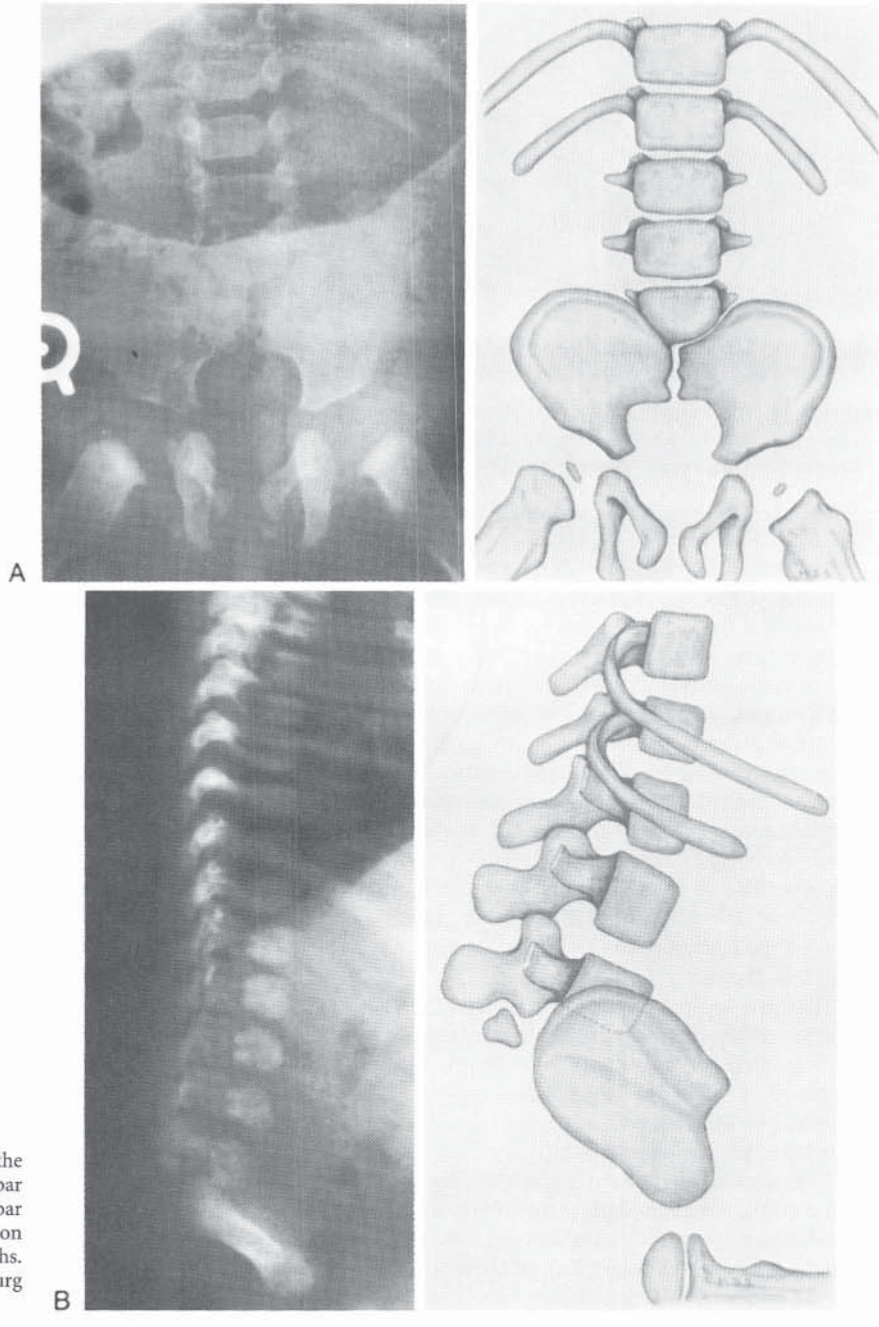


FIGURE 25–45 Type IV lumbosacral agenesis, the most severe type. The entire sacrum and lower lumbar spine are absent, and the articulation between the lumbar spinal remnant and pelvis is unstable. **A,** Appearance on AP radiographs. **B,** Appearance on lateral radiographs. (From Renshaw TS: Sacral agenesis. J Bone Joint Surg 1978;60-A:373.)

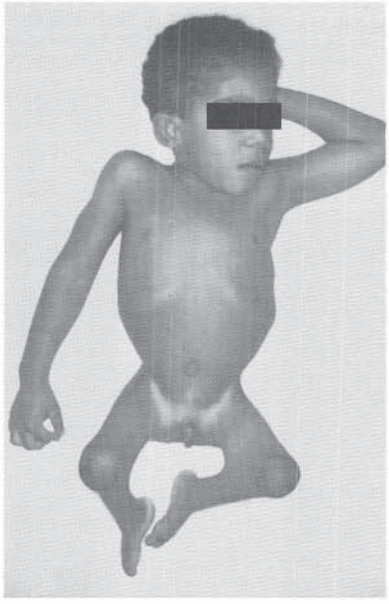


FIGURE 25–46 Clinical appearance of a 6-year-old child with type IV lumbosacral agenesis. Note the tapering of the lower extremities, and the "Buddha-like" posture secondary to the flexion contractures and webbing of the popliteal fossa.

attitude (Fig. 25–46). The T12 vertebra is often prominent posteriorly. There is marked disproportion between the thorax and the pelvis. The narrow, flat buttocks exhibit a depressed dimpling 2 or 3 inches lateral to the gluteal cleft. The normal convexity of the sacrococcygeal region is lost, and the anus is horizontal. The pelvis is very unstable under the spine—it tends to roll up under the thorax and drop forward, seeming to rest anterior to the thoracic spine, so that the patient tries to sit unsupported and is forced to support himself on his hands. In the occasional case in which there is also absence of the T12 vertebra, the opposing ribs articulate in the midline posteriorly. Almost all patients with type IV lumbosacral agenesis develop progressive spinopelvic kyphosis and scoliosis; they may require stabilization of the spine by spinal fusion.[10,24,25,27,32]

The hips typically have severe restriction of motion in a position of hip flexion-abduction contracture. The hips may be dislocated but in most cases are not. Knee flexion contractures of 50 to 60 degrees are typical, often with large popliteal webs. Fixed deformities of the feet (calcaneus or equinovarus) are usually seen.

There is muscle paralysis and atrophy of the lower limbs that is complete at and below the knees, with no voluntary or involuntary motor or reflex movement. Because of marked muscle atrophy, the lower half of the body develops a cone-shaped appearance. An extreme (and extremely rare) variation of this deformity is a single lower extremity, or "mermaid" configuration, known as sirenomelia.[14,31]

Peculiarly, sensory function is relatively intact compared to motor deficit, and is usually present at least to the knees. Distally, there may be spotty areas of hypesthesia or anesthesia. Unfortunately, rarely do patients have bladder or bowel control.

The severe pelvic outlet deformity may obstruct the lower intestinal tract, requiring colostomy. In order to ambulate, patients with lumbosacral agenesis may require spinopelvic stabilization, extension-producing releases or osteotomies at the hips and knees, and extensive orthotic support.

Treatment. The treatment of patients with caudal regression syndrome must be individualized based on the severity of the lower extremity deformity, motor paralysis, and motivation. The absence of CNS deformities such as hydrocephalus and Arnold-Chiari malformations and the preservation of protective sensation in the lower extremities can make efforts to reconstruct the lower extremities to facilitate ambulation rewarding. However, the severe lower extremity contractures associated with limited joint range of motion and extensive motor paralysis may militate against significant functional gains in mobility by lower extremity surgery for more severely affected individuals.

Some authors[9,28] have reported the results of treatment by bilateral subtrochanteric amputation and fitting with hip disarticulation prostheses. This procedure may be indicated in patients with severe, uncorrectable knee flexion deformities who simultaneously require spinopelvic stabilization. However, we have not found extensive limb-ablative surgeries beneficial to the mobility of our patients with caudal regression syndrome in the long term, and we prefer to concentrate on wheelchair mobility and spinal stabilization as necessary in such patients.

Knee flexion and foot deformities can be corrected by surgical releases or shortening, corrective osteotomy. A plantigrade foot can be provided for weightbearing with orthotic support. Unfortunately, recurrence of knee flexion deformity is frequent. Stabilization of the residual spine to the pelvis is indicated in the rare patient who is unable to sit comfortably secondary to the lumbopelvic instability. Stabilization may be achieved with rod or plate fixation of the pelvis to the residual spine, with bone grafting between the pelvis and spine.[10,25,27,32]

REFERENCES

Other Forms of Spinal Dysraphism

1. Adra A, Cordero D, Mejides A, et al: Caudal regression syndrome: etiopathogenesis, prenatal diagnosis, and perinatal management. Obstet Gynecol Surv 1994;49:508.
2. Andrish J, Kalamchi A, MacEwen GD: Sacral agenesis: a clinical evaluation of its management, heredity, and associated anomalies. Clin Orthop 1979;139:52.
3. Banta JV, Nichols O: Sacral agenesis. J Bone Joint Surg 1969;51-A:693.
4. Barson AJ: The vertebral level of termination of the spinal cord during normal and abnormal development. J Anat 1970;106:489.
5. Berbrayer D: Tethered cord syndrome complicating spina bifida occulta: a case report. Am J Phys Med Rehabil 1991;70:213.
6. Boemers TM, van Gool JD, de Jong TP, et al: Urodynamic evaluation of children with the caudal regression syndrome (caudal dysplasia sequence). J Urol 1994;151:1038.
7. Bono R, Inverno M, Botteon G, et al: Clinical features and MR imaging in children with repaired myelomeningocele. Ital J Neurol Sci 1993; 14:553.
8. Cama A, Palmieri A, Capra V, et al: Multidisciplinary management of caudal regression syndrome (26 cases). Eur J Pediatr Surg 1996; 6(suppl 1):44.
9. Douglas DL, Sharrard WJ: Treatment of lumbosacral agenesis by through-knee amputation and femoral osteotomy. J R Coll Surg Edinb 1988;33:219.
10. Dumont CE, Damsin JP, Forin V, et al: Lumbosacral agenesis: three cases of reconstruction using Cotrel-Dubousset or L-rod instrumentation. Spine 1993;18:1229.
11. Fitz CR, Harwood-Nash DC: The tethered conus. AJR Am J Roentgenol 1975;125:515.

12. Frantz CH, Aitken GT: Complete absence of the lumbar spine and sacrum. J Bone Joint Surg 1967;49-A:1531.
13. Goldberg C, Fenelon G, Blake NS, et al: Diastematomyelia: a critical review of the natural history and treatment. Spine 1984;9:367.
14. Guidera KJ, Raney E, Ogden JA, et al: Caudal regression: a review of seven cases, including the mermaid syndrome. J Pediatr Orthop 1991;11:743.
15. Harlow CL, Partington MD, Thieme GA: Lumbosacral agenesis: clinical characteristics, imaging, and embryogenesis. Pediatr Neurosurg 1995; 23:140.
16. Heinz ER, Rosenbaum AE, Scarff TB, et al: Tethered spinal cord following meningomyelocele repair. Radiology 1979;131:153.
17. Herman JM, McLone DG, Storrs BB, et al: Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord: presentation, management and outcome. Pediatr Neurosurg 1993;19:243.
18. Kadziolka R, Asztely M, Hansson T, et al: Ultrasonic measurement of the lumbar spinal canal: the origin and precision of the recorded echoes. J Bone Joint Surg 1981;63-B:504.
19. Lynch SA, Bond PM, Copp AJ, et al: A gene for autosomal dominant sacral agenesis maps to the holoprosencephaly region at 7q36. Nat Genet 1995;11:93.
20. McLone DG, Herman JM, Gabrieli AP, et al: Tethered cord as a cause of scoliosis in children with a myelomeningocele. Pediatr Neurosurg 1990;16:8.
21. Miller JH, Reid BS, Kemberling CR: Utilization of ultrasound in the evaluation of spinal dysraphism in children. Radiology 1982;143:737.
22. Miller P, Herndon WA, Yngve DA: Lumbosacral agenesis. Orthopedics 1985;8:1297.
23. Perrot LJ, Williamson S, Jimenez JF: The caudal regression syndrome in infants of diabetic mothers. Ann Clin Lab Sci 1987;17:211.
24. Perry J, Bonnett CA, Hoffer MM: Vertebral pelvic fusions in the rehabilitation of patients with sacral agenesis. J Bone Joint Surg 1970;52:288.
25. Phillips WA, Cooperman DR, Lindquist TC, et al: Orthopaedic management of lumbosacral agenesis: long-term follow-up. J Bone Joint Surg 1982;64-A:1282.
26. Renshaw TS: Sacral agenesis: a classification and review of twenty-three cases. J Bone Joint Surg 1978;60-A:373.
27. Rieger MA, Hall JE, Dalury DF: Spinal fusion in a patient with lumbosacral agenesis. Spine 1990;15:1382.
28. Russell HE, Aitken GT: Congenital absence of the sacrum and lumbar vertebrae with prosthetic management. J Bone Joint Surg 1963; 45-A:501.
29. Sarwark JF, Weber DT, Gabrieli AP, et al: Tethered cord syndrome in low motor level children with myelomeningocele. Pediatr Neurosurg 1996;25:295.
30. Sparnon AL, Ahmed S: Urological anomalies in the caudal regression syndrome. Aust NZ J Surg 1984;54:365.
31. Towfighi J, Housman C: Spinal cord abnormalities in caudal regression syndrome. Acta Neuropathol (Berl) 1991;81:458.
32. Winter RB: Congenital absence of the lumbar spine and sacrum: one-stage reconstruction with subsequent two-stage spine lengthening [see comments]. J Pediatr Orthop 1991;11:666.
33. Winter RB, Haven JJ, Moe JH, et al: Diastematomyelia and congenital spine deformities. J Bone Joint Surg 1974;56-A:27.

Spinal Muscular Atrophy

Spinal muscular atrophy (SMA) is found in a hereditary group of diseases. It is characterized by degeneration of the anterior horn cells of the spinal cord and occasionally of the motor neurons of the cranial nerves (fifth to 12th). The classic infantile form of SMA was first described by Werdnig in 1891,[29] while the less severe form was described by Kugelberg and Welander much later, in 1956.[14] SMA manifests with progressive hypotonia and weakness involving the lower extremities to a greater degree than the upper extremities and the proximal muscles more so than the distal musculature. Sensation is normal, as is intelligence. The incidence of SMA is approximately 1 in 15,000 to 1 in 20,000 live births,[7] and the prevalence of the carrier state is 1 in 80.

GENETIC STUDIES

SMA is usually inherited in an autosomal recessive pattern. It is the second most common disease inherited in an autosomal recessive pattern to affect children, after cystic fibrosis.[7] The genetic locus for spinal muscular atrophy has been identified on chromosome 5q.[7] Deletions or mutations at this location have been demonstrated in up to 98 percent of patients with SMA.[16] A less common form of the disease is inherited in an autosomal dominant pattern, and its genetic locus is also on 5q13.[5,23] The 5q locus gene product is the "survival motor neuron" protein, also known as SMN protein, which is deficient in patients with SMA.[15,16] Although the SMN protein has been linked with RNA metabolism, its precise role is under investigation.[19] With molecular genetic technology, first trimester prenatal diagnosis is now possible.[17,30]

CLASSIFICATION

There are three different types of spinal muscular atrophy: type I, acute infantile SMA or Werdnig-Hoffmann disease; type II, chronic infantile SMA; and a milder type III SMA, also known as Kugelberg-Welander disease. These forms are genetically similar but differ in age of presentation and in clinical course. As a rule, the younger the age at onset of the disease, the worse the prognosis is. There is some overlap among the various types.

Type I, Acute Infantile SMA. Type I SMA is usually identified between birth and age 6 months. Abnormal inactivity of the fetus in late pregnancy is reported by most mothers of affected babies. The newborn is floppy and inactive, with little to no movement of the arms or legs. The hips assume a characteristic posture, flopping into abduction, flexion, and external rotation, with flexion of the knees (Fig. 25–47). Finger and toe movement is usually present, as the distal musculature is relatively spared. Head control is absent, and the infant cannot lift his or her head. The growing infant is unable to roll over or sit.

Physical examination reveals lack of deep tendon reflexes. Tongue fasciculations are characteristic. The diaphragm is unaffected, but the intercostals are involved, leading to paradoxical respiratory movements. The face may appear expressionless, because the facial muscles are very weak.

The clinical course of type I SMA is one of relentless progression, usually leading to death by age 2 years secondary to pneumonia.[13] Longer survival has been seen more recently.[22] Poorer survival has been linked to age at presentation of less than 2 months.[26]

Because of the poor medical prognosis in acute Werdnig-Hoffmann SMA, orthopaedic treatment is rarely indicated. Birth fractures may occur and heal readily with splinting.[8] Medical trials are ongoing.

Type II, Chronic Infantile SMA. Type II SMA is diagnosed between the ages of 6 and 12 months. Muscular weakness is seen and is greater in the lower limbs than in the arms. The patellar reflex is absent, but the bicep and tricep reflex may be preserved in the very young. Tongue fasciculations and upper extremity tremors may be noted.

The infants achieve head control, and approximately three-fourths of affected children will develop sitting bal-

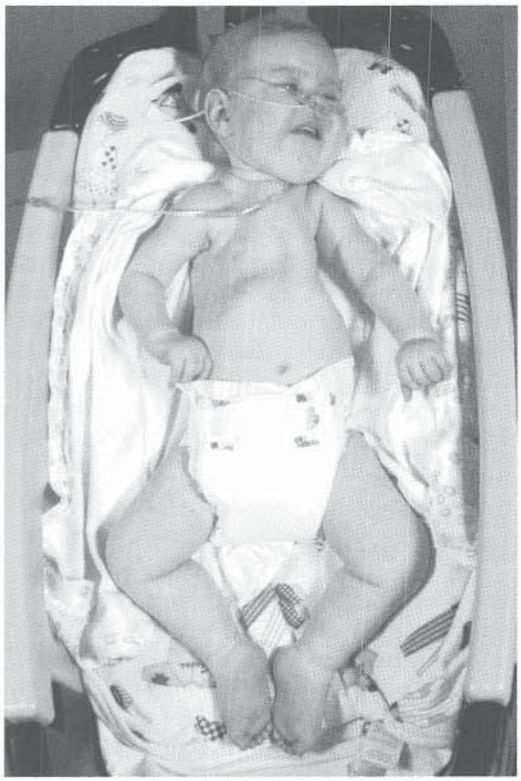


FIGURE 25–47 Infant with type I spinal muscular atrophy. The hips are held in abduction, flexion and extension rotation. There is obvious respiratory compromise. (Photograph courtesy of Susan Iannacone, M.D.)

FIGURE 25–48 Seven-year-old boy with type II spinal muscular atrophy. He has bilateral hip dislocation and diffuse weakness, and uses a wheelchair for mobility.

ance. They do not develop the ability to walk, and require wheelchairs for mobility (Fig. 25–48). Survival is better than in type I SMA. Some patients live into their fifth decade.

Type III, Kugelberg-Welander SMA. Type III SMA is usually diagnosed between the ages of 2 and 15 years. The presenting symptoms are of those hip extensor weakness, such as difficulty rising from the floor and climbing stairs. Type III SMA in males may initially be confused with Duchenne's or Becker's muscular dystrophy. Pseudohypertrophy of the calf may further confound the diagnosis. These patients do walk early in life, and most maintain their ambulatory skills into adolescence but become wheelchair bound in adulthood. The life span is nearly normal.

A functional grading of SMA has been proposed by Evans and colleagues.[9] This grading scale focuses on the highest function achieved, rather than on the age at onset, and is helpful prognostically in planning the orthopaedic treatment of the child with SMA (Table 25–1).

DIFFERENTIAL DIAGNOSIS

Infantile spinal muscular atrophy must be distinguished from cerebral palsy, which may present with hypotonia and flaccidity in the young infant. Congenital muscular dystrophy may resemble spinal muscular atrophy as well. Transverse myelitis may result from obstetric delivery and produce flaccid paralysis of the limbs and trunk. The characteristic presence of tongue fasciculations, which occur in 56 percent of patients with spinal muscular atrophy but are absent in all other neuromuscular diseases of infancy, can assist the clinician in the diagnosis.[13] The differential diagnosis of hypotonia in infants and children is given in Table 25–2.

DIAGNOSTIC EVALUATION

Serum creatine phosphokinase (CPK) and aldolase levels are usually normal in infants with spinal muscular atrophy. Mild elevation in muscle enzyme levels can be seen in children with later onset of SMA.

Electromyography in patients with SMA shows denervation, with fibrillation potentials and increased amplitude of response. Nerve conduction velocities are normal.

Muscle biopsy shows atrophy of the fiber group, with all fibers having a uniformly small diameter. This is distinctly different from the muscle biopsy appearance in muscular dystrophy, which shows variation in fiber size and degenerating muscle fibers. Muscle biopsy in hypotonic cerebral palsy is normal.

ORTHOPAEDIC TREATMENT

The orthopaedic treatment of children with SMA addresses three concerns: the development of contractures of the lower extremities, hip subluxation or dislocation, and scoliosis.

TABLE 25–1 **Spinal Muscular Atrophy: Functional Grading**

Grade I	Cannot sit independently, poor head control, early scoliosis
Grade II	Head control present, sitting possible, unable to stand or walk
Grade III	Can pull to stand and walk with support
Grade IV	Walk and run independently

TABLE 25–2 **Clinico-Anatomic Classification and Differential Diagnosis of the Hypotonic Child Syndrome**

	Cerebral Hypotonic Diplegia	Infantile Muscular Atrophy	Acute Infective Polyneuritis	Juvenile Myasthenia Gravis	Progressive Muscular Dystrophy	Polymyositis	Benign Congenital Hypotonia
Site of lesion	Cerebrum	Anterior horn cells	Peripheral nerves	Myoneural junction	Skeletal muscle	Skeletal muscle	Skeletal muscle
Inheritance	None	Recessive	None	Not defined	Sex-linked	None	None
Sex preponderance	None	None	None	Females	Males	Females	None
Limb musculature involved	Distal more than proximal	Generalized	Distal more than proximal	Generalized	Proximal	Proximal	Generalized
Cranial muscle pareses	Facial and pseudobulbar	Bulbar in late stages	Facial often, bulbar less often	Eyelids very often, ocular, facial, bulbar	None	Pharyngeal 50%	None
Respiratory paralysis	None	Common	Less common	Common 43%	Late	Occasional	Mild
Muscle fasciculations	None	Common	Less common	None	None	None	None
Muscle atrophy	Moderate	Severe	Moderate to severe	Mild	Severe	Mild (with tenderness)	Moderate
Pseudohypertrophy	None	None	None	None	Characteristic	Occasional	None
Deep tendon reflexes	Brisk or normal	Absent	Absent	Normal	Variable, usually depressed	Variable, usually depressed	Normal or depressed
Sensory defect	Cortical	None	Frequent	None	None	Usually none	None
Mental defect	Severe	None	None	None	Moderate	None	None or moderate
Muscle biopsy	Normal	Grouped atrophy	Atrophy	Lymphorrhages	Degeneration, variation in fiber size	Degeneration, and inflammatory cells	Small fibers
Electromyography	Normal	Fibrillations; sparse, giant action potentials	Fibrillations; sparse action potentials	Decline in amplitude of potentials	Short, low-amplitude potentials	Fibrillations; short, low-amplitude potentials	Normal or low-amplitude potentials
Serum creatinine kinase, SGOT, aldolase	Normal	Normal	Normal	Normal	Increased	Increased	Normal
Spinal fluid protein	Normal	Normal	Often high	Normal	Normal	Usually normal	Normal
Specific therapy	None	None	Steroids (?)	Rest and anticholinesterase drugs	None	Steroids	None
Course	Nonprogressive	Rapid	Acute or subacute	Prolonged, remittent	Chronic	Acute, subacute, or chronic	Nonprogressive
Prognosis	Severe chronic disability	Fatal, usually within 2 yr (4 wk to 20 yr)	Recovery in 80%	Complete remission in 25% within 6 yr; fatal in 5%	Fatal within 20–30 yr	Remission in 80%	Gradual improvement

From Millichap JG: The hypotonic child. Reproduced from Brennerman's Practice of Pediatrics, vol IV, chap 16. Hagerstown, MD, Medical Department, Harper & Row, Publishers, Inc, 1966.

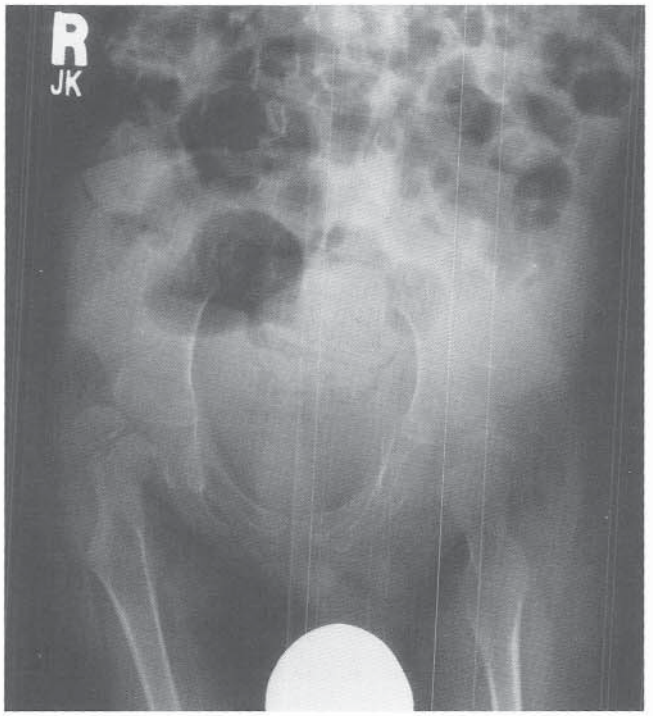


FIGURE 25–49 Bilateral hip dislocation in a 3-year-old boy with chronic infantile SMA.

Flexion contractures of the hips and knees invariably develop in children with SMA who are unable to walk. Physical therapy for gentle range of motion of the joints should be instituted. The role of surgical releases for correction of contractures is controversial. Rarely will the function of the nonwalking child be improved by surgical tenotomies, and recurrence of contractures in sitters is universal. Equinus contractures can occur in nonambulatory patients and may rarely develop in walking patients with SMA. Cavovarus foot deformities have also been described in ambulatory patients.[9]

Hip subluxation and dislocation are quite common in the nonambulatory patient with SMA (Fig. 25–49).[24] Proximal muscle weakness predisposes to coxa valga, which leads to uncoverage of the hips. Granata and colleagues found that radiographs of the hips were normal in only 31 percent of nonambulatory patients. The incidence of hip subluxation or dislocation in ambulatory patients in this series was 50 percent.[11] In the nonambulatory patient, unilateral dislocation leads to pelvic obliquity and uneven seating pressure. Bilateral dislocation does not predispose to pelvic obliquity but can accentuate lumbar hyperlordosis. Although the problem of hip instability is well recognized in spinal muscular atrophy, the treatment remains controversial.

Passive stretching exercises with release of contracted muscles has been advocated by Evans and colleagues for the treatment of hip dysplasia.[9] Surgical reconstruction of a unilateral subluxated or dislocated hip in a nonambulatory patient, consisting of varus derotation femoral osteotomy with or without capsulorrhaphy, has been advocated by Shapiro and Specht (Fig. 25–50).[25] Thompson and Larsen published a small series of four hip reconstructions for dislocation in SMA and found that all had poor results due to redislocation.[27] The need for reducing hip dislocations in these children remains unsettled.

Scoliosis is universal in nonambulatory patients with SMA, and is quite prevalent in children with the Kugelberg-Welander form of the disease as well. Granata and associates found that all but one of their patients with the infantile form of SMA developed scoliosis, with the mean age at which a curve was discovered being 4 years 4 months. All patients with mild SMA who lost the ability to ambulate also developed scoliosis, although at a slightly older age of 9 years 10 months. Twelve of 19 ambulatory patients developed scoliosis while able to walk, but the curves tended to progress more slowly than in sitting patients.[12] Evans and associates found that scoliosis manifested at a later age and was less severe in patients who were able to walk.[9]

Long, sweeping curves are most common in SMA, with a predominance of thoracolumbar curves (Fig. 25–51). Shapiro and Specht, in a meta-analysis of existing studies of scoliosis in SMA, found 12 percent thoracic curves, 62 percent thoracolumbar curves, 10 percent lumbar curves, and 16 percent double major curves.[25] Kyphosis is present in association with scoliosis in approximately 30 percent of patients.[12]

The nonoperative treatment of scoliosis in SMA is difficult. Orthoses can be prescribed to make sitting easier but are ineffective in preventing curve progression or altering the need for surgery.[9,12,18] Curves in nonambulatory patients were found to increase at a rate of 8 degrees per year despite brace use.[18] Respiratory function may be significantly depressed in patients with severe SMA, and rigid orthoses can further tax the patients' compromised respiratory status. A soft custom-made TLSO may be tolerated in young children with flexible curves between 20 and 40 degrees.[4]

Posterior spinal fusion with segmental spinal instrumentation is the treatment of choice for scoliosis in patients who can tolerate surgery. Fusion should include the entire thoracic and lumbar spine, and should extend to the pelvis. Luque-Galveston instrumentation and unit rod fixation have been used in these children (Fig. 25–52).[3,6] The goal of the surgery is to obtain a balanced trunk over a level pelvis to facilitate comfortable seating (Fig. 25–53). Anterior fusion is contraindicated in the vast majority of patients, as the risk of surgery in the chest and involving the diaphragm outweighs the potential problems from continued anterior growth and the crankshaft phenomenon.

Shapiro and Specht compiled the results of several studies of posterior spinal fusion in patients with SMA. The average age at surgery was 13.5 years. The average deformity was 90 degrees, and the average correction achieved after surgery was 43 percent. Based on their findings, they recommend earlier surgery to allow for less residual curve following instrumentation and fusion. They propose that surgery be considered in patients more than 10 years old with curves greater than 40 degrees and forced vital capacities greater than 40 percent of normal.[25]

Although curve correction and stabilization can be achieved through spinal arthrodesis, the patient often pays a price, as the ability to carry out activities of daily living declines following surgery. Specifically, the rigid and upright spine creates difficulty in self-feeding, drinking, and self-hygiene, as the patient cannot bring the hands against gravity to the face owing to upper extremity proximal muscle weak-

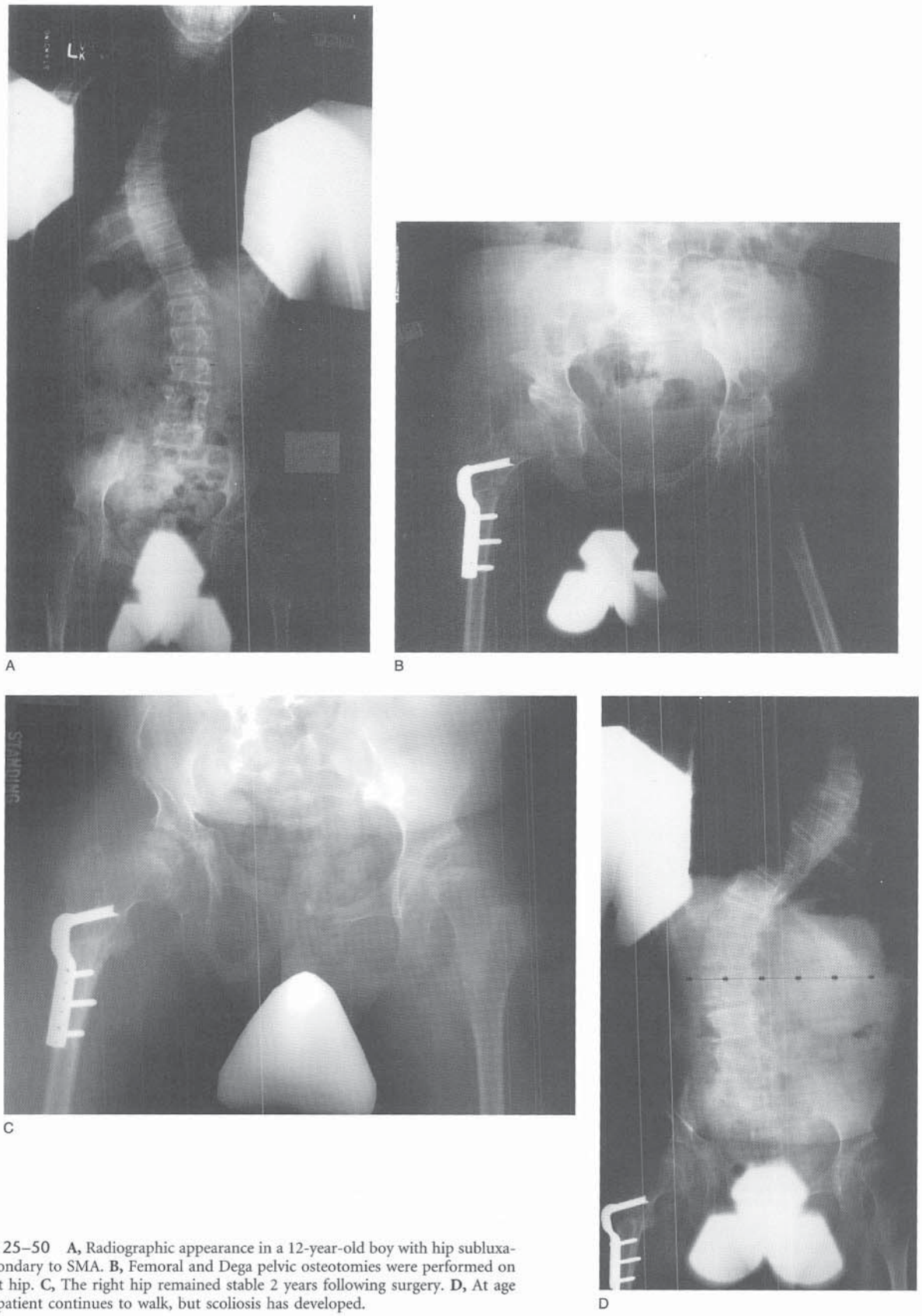


FIGURE 25–50 **A,** Radiographic appearance in a 12-year-old boy with hip subluxation secondary to SMA. **B,** Femoral and Dega pelvic osteotomies were performed on the right hip. **C,** The right hip remained stable 2 years following surgery. **D,** At age 14, the patient continues to walk, but scoliosis has developed.

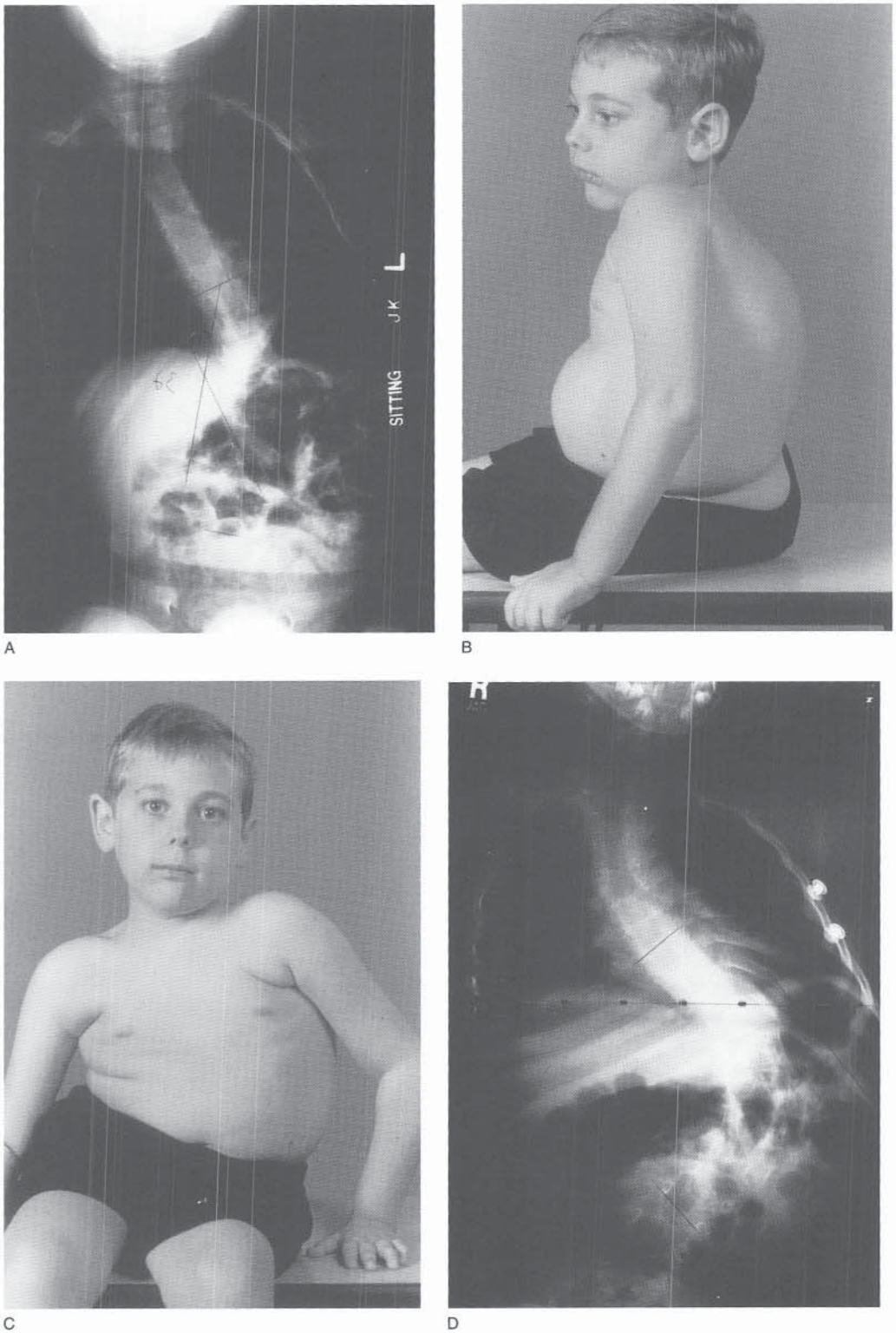


FIGURE 25–51 **A,** Thoracolumbar scoliosis in a 3½-year-old boy with type II SMA. **B** and **C,** Clinical appearance at age 9. The boy propped himself up with his upper extremities to maintain sitting balance. **D,** The magnitude of the thoracolumbar curve has significantly increased.

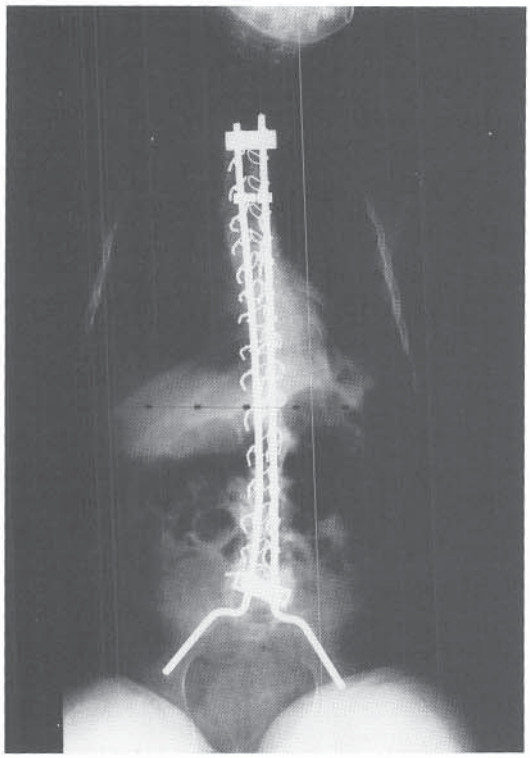


FIGURE 25–52 Posterior spinal fusion with Luque-Galveston instrumentation to the pelvis in the 9-year-old boy shown in Figure 25–51.

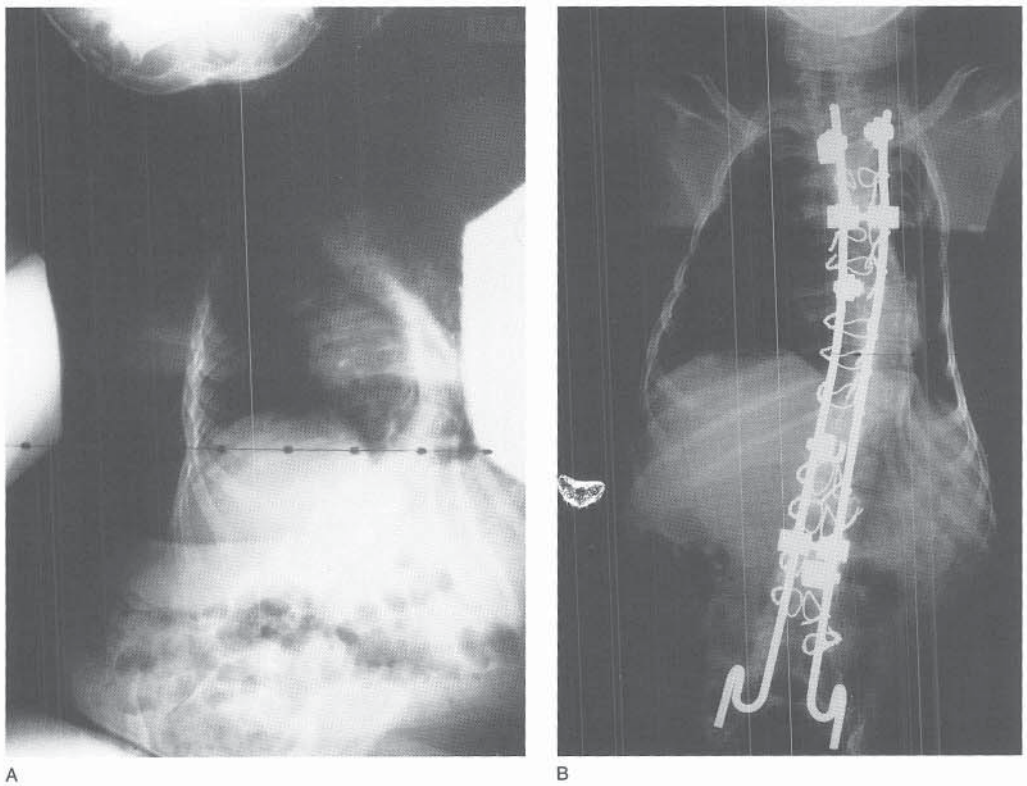
ness.[6,10] Counseling the patients and their families prior to surgery is essential. Occupational therapists should evaluate the children for the appropriateness of adaptive equipment following surgery.

There are conflicting reports in the literature regarding the effect of scoliosis surgery on pulmonary function. There is agreement that the decline in respiratory status does correlate with increasing severity of the scoliosis.[21] Some authors have found that the respiratory parameters measured improve following spinal arthrodesis,[21] while others have found a continuous decline in pulmonary function.[1,20] Nonetheless, aggressive pulmonary perioperative care must be provided to avoid respiratory complications such as pneumonia and the need for prolonged mechanical ventilation.

Scoliosis in the ambulatory patient poses a dilemma for treatment. Up to 50 percent of walking patients develop scoliosis. Often, lumbar lordosis and pelvic motion compensate for the proximal muscle weakness of the lower extremities and are essential to these patients' gait. The ability to walk may be lost in some patients following spinal arthrodesis.[9,10] For this reason, the surgical treatment of scoliosis should be delayed when reasonable in ambulatory patients.[25] When necessary, fusion may be stopped proximal to the pelvis (Fig. 25–54).

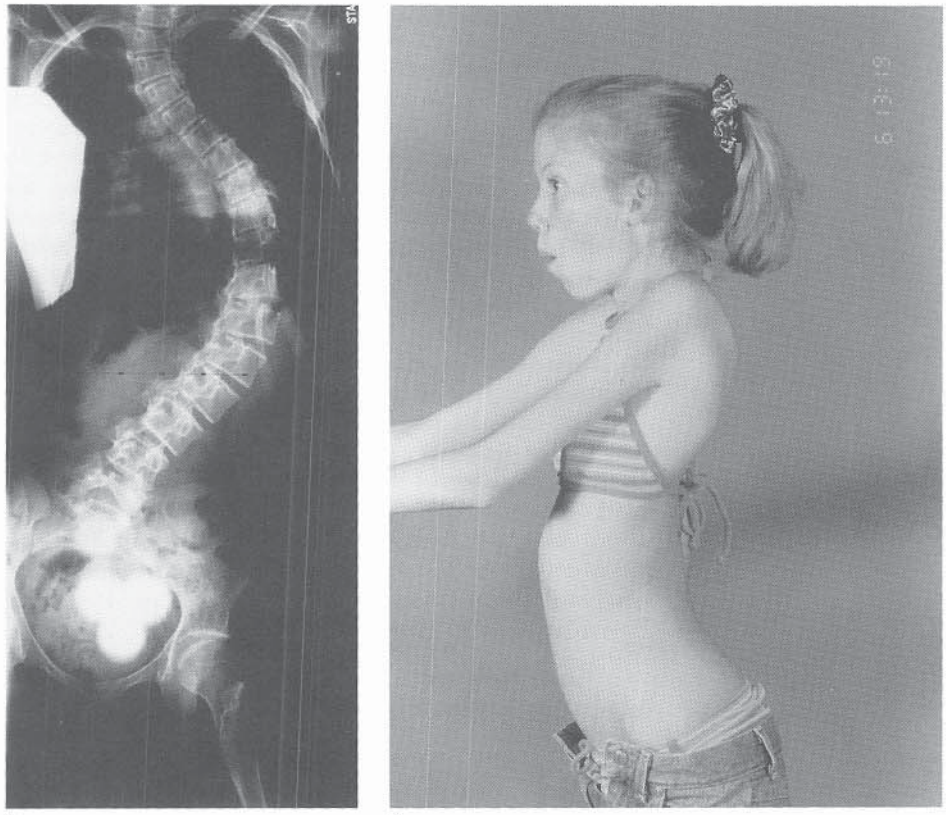
MEDICAL TREATMENT

There is currently no effective drug treatment for SMA. The life span of affected individuals has recently been significantly expanded in a few reported cases by the use of inter-

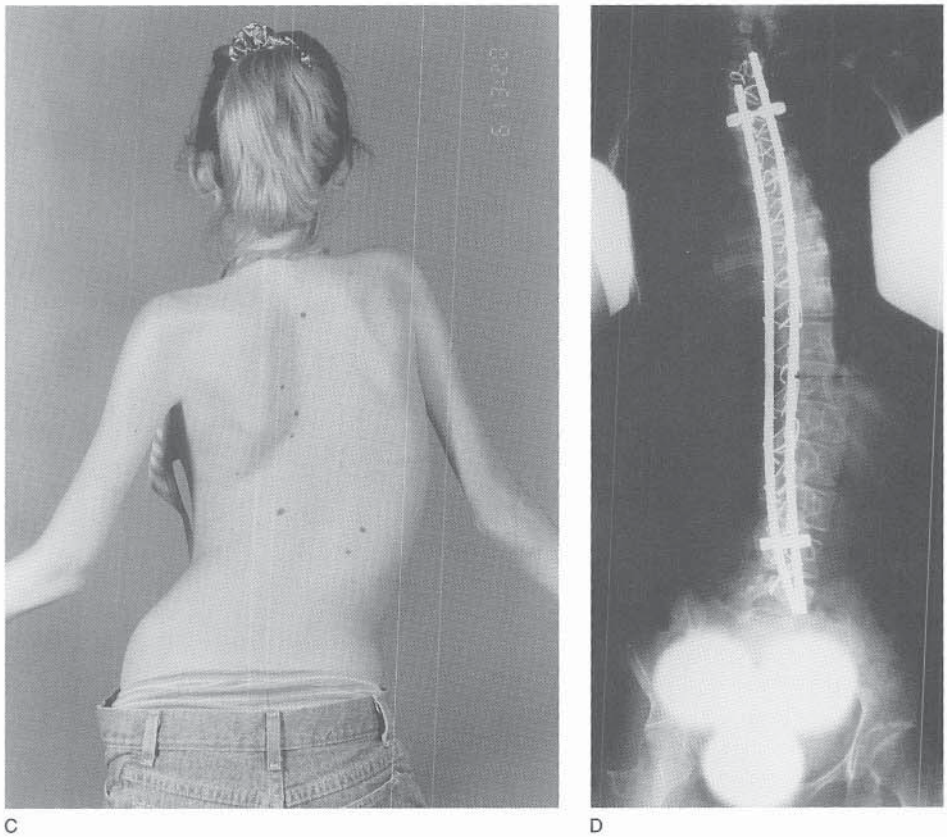


A B

FIGURE 25–53 **A,** PA spinal radiograph of a 12-year-old boy with type II SMA. **B,** Sitting balance has been improved by posterior spinal fusion with Dunn-McCarthy instrumentation to the pelvis.



A B



C D

FIGURE 25–54 **A,** PA standing spinal radiograph of an ambulatory 13-year-old girl with type III SMA. **B** and **C,** Clinical appearance of the girl. Note the anterior pelvic tilt, hip flexion, and excessive spinal lordosis. **D,** Posterior spinal fusion was performed using Luque instrumentation to L5 to allow pelvic motion during gait. Two years after surgery, she remains a limited ambulator.

mittent positive pressure ventilation, used both with and without tracheostomy.[2,28]

REFERENCES

Spinal Muscular Atrophy

1. Aprin H, Bowen JR, MacEwen GD, et al: Spine fusion in patients with spinal muscular atrophy. J Bone Joint Surg 1982;64-A:1179.
2. Bach JR, Wang TG: Noninvasive long-term ventilatory support for individuals with spinal muscular atrophy and functional bulbar musculature. Arch Phys Med Rehabil 1995;76:213.
3. Bell DF, Moseley CF, Koreska J: Unit rod segmental spinal instrumentation in the management of patients with progressive neuromuscular spinal deformity. Spine 1989;14:1301.
4. Bowen JR, Forlin E: Spinal muscular atrophy. In Weinstein SL (ed): The Pediatric Spine: Principles and Practice, p 1025. New York, Raven Press, 1994.
5. Boylan KB, Cornblath DR, Glass JD, et al: Autosomal dominant distal spinal muscular atrophy in four generations. Neurology 1995;45:699.
6. Brown JC, Zeller JL, Swank SM, et al: Surgical and functional results of spine fusion in spinal muscular atrophy. Spine 1989;14:763.
7. Brzustowicz LM, Lehner T, Castilla LH, et al: Genetic mapping of chronic childhood-onset spinal muscular atrophy to chromosome 5q11.2–13.3. Nature 1990;344:540.
8. Burke SW, Jameson VP, Roberts JM, et al: Birth fractures in spinal muscular atrophy. J Pediatr Orthop 1986;6:34.
9. Evans GA, Drennan JC, Russman BS: Functional classification and orthopaedic management of spinal muscular atrophy. J Bone Joint Surg 1981;63-B:516.
10. Furumasu J, Swank SM, Brown JC, et al: Functional activities in spinal muscular atrophy patients after spinal fusion. Spine 1989;14:771.
11. Granata C, Magni E, Merlini L, et al: Hip dislocation in spinal muscular atrophy. Chir Organi Mov 1990;75:177.
12. Granata C, Merlini L, Magni E, et al: Spinal muscular atrophy: natural history and orthopaedic treatment of scoliosis. Spine 1989;14:760.
13. Iannaccone ST, Browne RH, Samaha FJ, et al: Prospective study of spinal muscular atrophy before age 6 years. DCN/SMA Group. Pediatr Neurol 1993;9:187.
14. Kugelberg E, Welander L: Heredo-familial juvenile muscular atrophy simulating muscular dystrophy. AMA Arch Neurol Psychiatry 1956; 75:500.
15. Lefebvre S, Burlet P, Liu Q, et al: Correlation between severity and SMN protein level in spinal muscular atrophy. Nat Genet 1997;16:265.
16. Liu Q, Fischer U, Wang F, et al: The spinal muscular atrophy disease gene product, SMN, and its associated protein SIP1 are in a complex with spliceosomal snRNP proteins. Cell 1997;90:1013.
17. Lo Cicero S, Capon F, Melchionda S, et al: First-trimester prenatal diagnosis of spinal muscular atrophy using microsatellite markers. Prenat Diagn 1994;14:459.
18. Merlini L, Granata C, Bonfiglioli S, et al: Scoliosis in spinal muscular atrophy: natural history and management. Dev Med Child Neurol 1989;31:501.
19. Novelli G, Calza L, Amicucci P, et al: Expression study of survival motor neuron gene in human fetal tissues. Biochem Mol Med 1997;61:102.
20. Piasecki JO, Mahinpour S, Levine DB: Long-term follow-up of spinal fusion in spinal muscular atrophy. Clin Orthop 1986;207:44.
21. Robinson D, Galasko CS, Delaney C, et al: Scoliosis and lung function in spinal muscular atrophy. Eur Spine J 1995;4:268.
22. Russman BS, Iannacone ST, Buncher CR, et al: Spinal muscular atrophy: new thoughts on the pathogenesis and classification schema. J Child Neurol 1992;7:347.
23. Schrank B, Gotz R, Gunnersen JM, et al: Inactivation of the survival motor neuron gene, a candidate gene for human spinal muscular atrophy, leads to massive cell death in early mouse embryos. Proc Natl Acad Sci USA 1997;94:9920.
24. Schwentker EP, Gibson DA: The orthopaedic aspects of spinal muscular atrophy. J Bone Joint Surg 1976;58-A:32.
25. Shapiro F, Specht L: The diagnosis and orthopaedic treatment of childhood spinal muscular atrophy, peripheral neuropathy, Friedreich ataxia, and arthrogryposis. J Bone Joint Surg 1993;75-A:1699.
26. Thomas NH, Dubowitz V: The natural history of type I (severe) spinal muscular atrophy. Neuromuscul Disord 1994;4:497.
27. Thompson CE, Larsen LJ: Recurrent hip dislocation in intermediate spinal atrophy. J Pediatr Orthop 1990;10:638.
28. Wang TG, Bach JR, Avilla C, et al: Survival of individuals with spinal muscular atrophy on ventilatory support. Am J Phys Med Rehabil 1994;73:207.
29. Werdnig G: Zwei fruhim famile hereditare Fall von progressiver Muskelatrophie unter dem Bild der Dystrophie, aber auf neurotischer Grundlage. Arch Psychiatr Nervenkr 1891;22:237.
30. Wirth B, Rudnik-Schoneborn S, Hahnen E, et al: Prenatal prediction in families with autosomal recessive proximal spinal muscular atrophy (5q11.2–q13.3): molecular genetics and clinical experience in 109 cases. Prenat Diagn 1995;15:407.