CHAPTER 15

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Myelomeningocele

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Neural tube defects result from failure of the neural tube to close during embryogenesis. Although the incidence of neural tube defects has declined in recent decades, it remains the cause of chronic disability of between 70,000 and 100,000 individuals in the United States (1). Myelomeningocele, also referred to as spina bifida, is the most common neural tube defect and is the most severely disabling birth defect compatible with survival (1).

Myelomeningocele is a fluid filled cystic swelling, formed by dura and arachnoid. Myelodysplasia of the neural elements manifests in the vertebrae as a defect in the posterior elements. The sac protrudes through this defect and contains spinal nerve roots. Dysplasia of the spinal cord and nerve roots leads to bowel, bladder, motor, and sensory paralysis below the level of the lesion (2). Patients with myelomeningocele can also have concomitant lesions of the spinal cord, such as diastematomyelia or hydromyelia, or structural abnormalities of the brain, such as hydrocephalus or Arnold-Chiari malformation, which can also compromise neurologic function.

The survival rate for patients with myelomeningocele in the 1950s was only about 10%. Due to advances in the management of several important complications, a recent series reported at least 75% of children born with an open myelomeningocele defect can be expected to reach their early adult years (3). However, comprehensive treatment requires optimal care to prevent, monitor, and treat a variety of potential complications that can affect function, quality of life, and survival. This is best accomplished by a multidisciplinary team approach including specialists in orthopaedic surgery, neurosurgery, urology, rehabilitation, physical and occupational therapy, and orthotics. Access to nutritionists, social workers, wound specialists, and psychologists is also helpful.

As a result of the increased survival into adulthood, many patients with myelomeningocele now live long enough to eventually require transition to adult medical providers. This presents a great challenge since adult providers may lack the expertise necessary to manage these patients. Additionally, adults with myelomeningocele who failed to develop the skills they require to live independently remain dependent on aging family members who may be unable to care for them.

INCIDENCE

The incidence of infants born with neural tube defects shows regional and racial variations but is decreasing overall. The birth prevalence rate of myelomeningocele from 1983 to 1990 in the United States was 4.6 per 10,000 (4). Since that time, there has been a decrease in the number of new cases of myelomeningocele. This decrease can be attributed to two main factors: prenatal screening with elective termination of affected pregnancies and increased awareness of the importance of administration of folate to women before and during pregnancy. The United States Public Health Service recommends that all women of childbearing age who are capable of becoming pregnant should consume 400 μg of folic acid per day for the purpose of reducing their risk of having a pregnancy affected with myelomeningocele or other neural tube defects (5). Total folate consumption should be <1 mg per day because the effects of higher intakes are not well known (5).

An estimated 50% to 70% of neural tube defects can be prevented through the daily consumption of 400 μg of folic acid (5). The U.S. Food and Drug Administration mandated adding folic acid to all enriched cereal grain products by January 1998 (6). From October 1998 to December 1999, the birth prevalence rate of myelomeningocele in the United States decreased 22.9% compared with 1995–1996 (6). Notably, the prevalence of myelomeningocele remained higher among Hispanic women than among women in other racial/ethnic

populations (6). This finding may be attributable to differences in folic acid consumption, eating habits, or genetic factors. In addition, religious and cultural preferences may play a role in the persistently higher prevalence of myelomeningocele in Hispanic women who may be less likely to terminate an affected pregnancy.

Overall the trend of decreasing incidence of myelomeningocele in the United States has continued after the folic acid mandate. From the early postfortification period of 1999–2000 to the recent postfortification period of 2003–2005, the birth prevalence of myelomeningocele among infants born to mothers of all racial/ethnic populations decreased 6.9%, from 2.04 to 1.90 cases per 10,000 live births (6).

A similar trend of decreased incidence of myelomeningocele related to folic acid consumption has been reported in Europe. Two of the first European countries to develop a periconceptional folic acid supplementation policy were the United Kingdom (1992) and Ireland (1993) (7). In a population-based study examining the effect of folic acid supplementation on the prevalence of neural tube defects in 16 European countries, a 32% decrease was found when comparing the periods 1989–1991 and 1999–2001 in the United Kingdom and Ireland (7). A 17% reduction in prevalence of neural tube defects was found in countries with folic acid supplementation introduced by 1999. In contrast, a decrease of 9% was seen in countries with no supplementation policy by 1999.

ETIOLOGY

Myelomeningocele is believed to result from failure of fusion of the neural folds during neurulation, which occurs at 26 to 28 days of gestation. Conditions that result from abnormalities during the phase of closure of the neural tube, such as myelomeningocele and anencephaly, are referred to as neurulation defects. In contrast, conditions such as meningocele, lipomeningocele, and diastematomyelia arise from abnormalities that occur during the canalization phase from 28 to 48 days of gestation and are referred to as postneurulation defects.

The cause of this embryonic failure is not known but is suspected to be multifactorial in origin, involving both genetic and environmental factors. Folate deficiency is an important contributor to the cause of neural tube defects as evidenced by the decrease in incidence observed after folate supplementation. Other environmental factors have also been examined for a potential role in neural tube defects, including temperature; drug exposure; substance abuse; maternal infection; and other nutritional factors, such as vitamin B_{12} and zinc (8).

Genetic factors seem to play an important role in the development of myelomeningocele. Animal studies have shown as many as 100 mutant genes that affect neurulation, and almost all have homologs in humans (8). Studies have suggested a higher incidence of neural tube defects in siblings of affected children than in the general population. A positive family history has been reported in 6% to 14% of cases (9, 10). Overall, for a couple with a child with myelomeningocele, the chance that a subsequent sibling would be affected by a major malformation of the central nervous system is approximately 1 in 14 (11). Although association with single gene defects, increased recurrence risk among siblings, and a higher frequency in twins than in singletons indicate a genetic contribution to the etiology, the low frequency of families with a significant number of neural tube defect cases makes research into genetic causation difficult (8).

DIAGNOSIS

Prenatal screening for myelomeningocele and other neural tube defects involves biochemical testing of maternal blood for alpha-fetoprotein or the use of ultrasound evaluation. Maternal serum alpha-fetoprotein, a glycoprotein secreted by the fetal yolk sac and liver, has been used as a screening test for open neural tube defects for over 30 years. The detection rate for anencephaly is >95% and for open neural tube defects between 65% and 80% (12). Since closed neural tube defects do not increase alpha-fetoprotein, biochemical screening is not effective. Additionally, an increased serum alpha-fetoprotein is not diagnostic for open neural tube defects since it can also be associated with other abnormalities including gastroschisis, omphalocele, congenital nephrosis, and fetal demise.

With improvement in ultrasonographic techniques, prenatal diagnosis using ultrasound can be quite accurate. A recent report on prenatal screening in Europe found 88% of 725 cases of neural tube defects were detected prenatally using ultrasound at a median gestation of 17 weeks (13). The technique of three-dimensional ultrasound using multiplanar views can achieve diagnostic accuracy within one vertebral body in around 80% of patients (12).

When a diagnosis of myelomeningocele is suspected on ultrasound, careful evaluation of the entire spine and a search for other abnormalities is warranted as associated malformations are found in around 23% of patients (14).

ASSOCIATED CONDITIONS

Hydrocephalus. After repair and closure of the myelomeningocele defect, which is done in the first 48 hours of life, many infants will develop some degree of hydrocephalus. Using new protocols aimed at reducing shunt placement rates in patients with myelomeningocele, approximately 60% of infants with require a shunt (15). The incidence of hydrocephalus with need for cerebrospinal fluid diversion has been reported to correlate with functional level of the myelomeningocele lesion. Between 97% and 100% of patients with a thoracic level lesion require shunt placement compared to 87% of lumbar-level patients and 37% of sacral-level patients (15, 16).

Patients who do not require shunting may have a better prognosis in terms of upper extremity function and trunk balance as compared to patients who require shunting (17). One study comparing a group of 98 patients with myelomeningocele

and a shunt with a group of 63 patients with no shunt found that patients without a shunt were more independent in their ambulation at medium and longer distances (18). In addition, the authors noted patients with no shunt tend to walk at a significantly greater velocity and stride length as compared with those with a shunt (18).

Infection and obstruction of cerebrospinal fluid shunts are serious complications that have the potential to affect a patient's motor and intellectual development. In one study following a group of 61 patients with myelomeningocele and a cerebrospinal fluid shunt, 95% patients underwent at least one shunt revision (3). Data not yet published by the senior author of this chapter also show that patients with an incidence of shunt infection have a decrease in functional mobility in the school and in the community compared to patients who have not experienced an infection (Dias L., personal communication). Awareness of this information will allow caregivers to effectively counsel patients with myelomeningocele and a ventriculoperitoneal shunt regarding functional ambulatory expectations.

Chiari II Malformation. The Chiari II malformation is present in almost all patients with myelomeningocele (1). It is characterized by caudal displacement of the posterior lobe of the developing fetal cerebellum and medulla into the spinal canal. If the brainstem or spinal cord is compressed within the spinal canal, progressive dysfunction may result, manifesting as weakness or paralysis of the vocal cords, or difficulty feeding, crying, or breathing (1). However, these symptoms are nonspecific and may also result from a shunt malfunction, which should be excluded prior to surgical decompression for the Chiari II malformation.

Tethered Spinal Cord. Tethered cord syndrome is a stretch-induced functional disorder of the spinal cord with its caudal part anchored by an inelastic structure such as scar tissue (19). Magnetic resonance imaging of the spine will show signs of tethering in most patients with myelomeningocele; however, the clinical signs are present in only about 30% (17). The most common clinical symptom of tethered cord is progressive scoliosis (44%) (3). Especially concerning is scoliosis that develops before 6 years of age in the absence of congenital vertebral anomalies. Also, since scoliosis is not as common in myelomeningocele patients with low lumbar- and sacral-level involvement, when seen in this population, it may signify tethered cord syndrome. Other common symptoms are gait changes associated with loss of muscle strength (35%) and spasticity (26%) (3) especially in the medial hamstrings and ankle dorsiflexors and evertors. Additional common symptoms associated with tethered cord are loss of motor function, back pain at the site of the repaired spina bifida defect (20), or changes in urologic function. As with hydromyelia, a shunt malfunction must be ruled out first when tethered cord is suspected. When a diagnosis of tethered cord syndrome is made, surgical treatment by a skilled neurosurgeon with experience with this procedure is indicated to prevent further deterioration. Often, symptoms will stabilize or improve with surgical untethering (1).

Hydromyelia. Hydromyelia is an accumulation of cerebrospinal fluid in the enlarged central canal of the spinal cord. In patients with myelomeningocele, hydromyelia develops due to shunt malfunction or untreated hydrocephalus. Magnetic resonance imaging of the spine in a group of 231 patients with myelomeningocele revealed hydromyelia in 49% (21). However, not all patients develop symptoms that require treatment of the hydromyelia. Those who are symptomatic may present with progressive scoliosis, urologic problems, pain, and motor or sensory defects (1). Decreased grip strength and thenar atrophy are also reliable signs of hydromyelia (17).

Urinary Tract. Most patients with myelomeningocele have neurogenic bladder dysfunction and may go on to develop progressive deterioration of the upper urinary tract and chronic renal disease. Treatment to reduce bladder pressure and minimize urine stasis is important to prevent these complications. In addition, regular monitoring of urinary tract function is necessary in order to detect changes in bladder function that may indicate shunt malfunction or tethered cord syndrome (1). Management includes clean intermittent catheterization of the bladder, which is necessary in approximately 85% patients with myelomeningocele (3). In addition, antibiotic prophylaxis and anticholinergic medication to reduce vesicoureteral reflux may be beneficial. A variety of surgical options exist for those patients that fail medical treatment or in order to facilitate self-management. These include vesicostomy, a diversion of the bladder to the lower abdominal wall to facilitate catheterization, and bladder augmentation in which a segment of the ileum is added to the bladder to increase capacity and reduce bladder pressure.

Bowel Management. Innervation of the bowel and anus is affected in most patients with myelomeningocele leading to dysmotility, poor sphincter control, and often fecal incontinence. Decreased bowel motility can cause constipation and fecal impaction, which in turn may cause increased intra-abdominal pressure leading to ventriculoperitoneal shunt malfunction (1). The goal of bowel management is to achieve continence and avoid fecal impaction by prompting regular elimination of stool using oral laxatives, suppositories, and/or enemas. If these measures are not successful, the Malone antegrade continence enema (MACE) procedure is an option. The MACE is a surgical procedure in which the appendix and the cecum are used to create a catheterizable stoma through which the patient irrigates the colon. In one study evaluating the results of the MACE procedure in 108 patients with myelomeningocele, approximately 85% achieved continence (22).

GENERAL HEALTH ISSUES

Patients with myelomeningocele have a high incidence of latex allergic reactions since they are exposed to latex products as a consequence of repeated surgical procedures, implantation of latex-containing materials, and catheterization (23). Latex

allergy occurs in 18% to 40% of patients with myelomeningocele (3, 23, 24). The reaction may be a severe, life-threatening anaphylaxis in up to 26% (3). For this reason, it is imperative to avoid exposure to latex in myelomeningocele patients both in and out of the hospital environment. All surgical procedures performed on myelomeningocele patients should be done in a strictly latex-free setting.

Nutrition is an important issue in patients with myelomeningocele and appropriate counseling should begin at an early age. Childhood and adolescent obesity is common in patients with myelomeningocele and likely results from a variety of factors including energy intake and motor impairment. One study of 100 children and adolescents with myelomeningocele found 40% were markedly overweight, defined as Body Mass Index above the 95th percentile (25).

The psychosocial impact of myelomeningocele on the patient and the family should not be overlooked. Parents of children with myelomeningocele report more psychosocial stresses compared to parents of able-bodied children. One study found parents of myelomeningocele patients reported less parental satisfaction, less perceived parental competence, more social isolation, and less adaptability to change in comparison to a matched group of parents of able-bodied children (26). Another study looked at depressive symptoms in patients from 9 to 18 years of age with myelomeningocele compared to matched able-bodied patients. The authors found greater risk of depressive mood, low self-worth, and suicidal ideation in the myelomeningocele patients (27). The multidisciplinary care team should be aware of the potential for psychosocial issues and be prepared to refer or treat any concerns appropriately.

COMPLICATIONS

Skin Breakdown. The risk of skin breakdown and development of pressure sores is a significant problem in patients with myelomeningocele who lack protective sensation. Reported incidence of pressure sores varies in the literature from 17% to 82% of patients (28–32). The most common locations are over the sacrum, ischial tuberosity, greater trochanter, or on the feet (31). One study followed a group of 75 patients with myelomeningocele treated for pressure sores and determined that over the 13-year study period more than two million dollars were spent on their treatment (33). Another group reported 415 admissions at their institution between 1988 and 2005 for the treatment of pressure ulcers (31).

Care must be taken to aggressively prevent the development of pressure sores. All patients should be instructed from a young age to avoid walking without adequate foot protection, especially on rough or hot surfaces. Orthotic devices should be inspected on a regular basis, at least annually, to ensure proper fit and no pressure points or sharp edges. When casting, ample padding must be used and applied in a smooth fashion. Self-adhering foam pads can be used to supplement padding over pressure points such as the anterior knee, heel, or ankle malleoli. In addition, surgical arthrodesis within the foot should be strictly avoided since the resulting inflexibility in an insensate foot has been shown to be related to the development of neuropathic skin changes (30).

Fractures. Long bone fractures occur in up to 20% of patients with myelomeningocele and may involve the physis, metaphysis, or diaphysis (34, 35). The increased risk for fracture is thought to be related to a variety of factors including disuse osteoporosis, joint contractures, and postsurgical immobilization, especially spica casting. In addition, the level of neurologic involvement has been found to correspond to prevalence of fracture, with the higher the level of involvement, the higher the prevalence of fracture (34). This is thought to be attributable to osteopenia related to relative lack of mobility.

In myelomeningocele patients, fractures may result from minor trauma or physical therapy, and caregivers must have a high index of suspicion. In addition, caregivers need to be aware of the typical presentation of a metaphyseal or diaphyseal fracture in myelomeningocele patients who may not have pain due to lack of normal sensation. In this population, fracture should be suspected when a patient presents with a warm, swollen extremity (35). Other signs of fracture include erythema, temperature elevation >100°F, white blood cell count >10,000/mm^3, elevated erythrocyte sedimentation rate, general malaise, or nausea and vomiting (34–36). If not aware of this presentation for fracture, a mistaken diagnosis of cellulitis or osteomyelitis may be made and delay proper treatment. When a diagnosis of metaphyseal or diaphyseal fracture is made, healing often proceeds quickly. Most fractures can be treated nonsurgically, and immobilization is usually required for only 2 to 4 weeks (34, 35).

In contrast to metaphyseal and diaphyseal fractures, physeal fractures often have a different cause and clinical presentation. Physeal fractures are most common in ambulatory patients with low lumbar level of involvement (34, 35). Patients may complain of mild pain and often have warmth and swelling but may have only minimal increase in temperature, erythrocyte sedimentation rate, and white blood cell count (35). Radiographs may show a widened growth plate with an irregular and slightly widened metaphysis (34). These fractures heal at a slower rate and often require immobilization for up to 8 weeks.

Infection. Patients with myelomeningocele have an increased risk of postoperative infection that is likely multifactorial in origin. Contributing factors are lack of protective sensation, bladder paralysis, and poor soft-tissue envelope. With regard to spine surgery in particular, wound infection may occur in up to 50% of patients (4). Bacterial colonization of the urinary tract may occur due to bladder paralysis and its management. Infection rates in spinal surgery have been found to be higher in the presence of concurrent urinary tract infection; hence, some recommend obtaining preoperative urinary cultures (4).

PROGNOSIS FOR AMBULATION

Within the orthopaedic community, there is debate over whether or not working to achieve the goal of early ambulation in patients with myelomeningocele is worthwhile. Some attest early ambulation can provide physiologic and psychological benefits to a child with myelomeningocele even if that child will later become a sitter, while others dispute these benefits. One study compared a group of 36 high-level myelomeningocele patients who participated in a walking program with 36 matched patients who were prescribed a wheelchair early in life (37). At final follow-up, only 12 patients in the walking group retained the ability for effective ambulation. Despite this, patients who had walked early had fewer fractures and pressure sores, were more independent, and were better able to transfer compared to the wheelchair group (37).

Many factors influence the potential for ambulation in an individual patient with myelomeningocele. One of the most important is the motor level of involvement. Other contributing factors include sitting balance, upper extremity spasticity, obesity, age, and availability of appropriate orthotic support. Musculoskeletal deformity of the spine, pelvis, knees, and feet has also been shown to significantly influence ability to walk (38).

Neurologic level of involvement and the resulting muscle group strength plays a crucial role in achieving and maintaining ambulation. Asher and Olson studied the ambulatory status of 98 patients with myelomeningocele and found a notable difference in the ability to walk between patients with fourth lumbar level of involvement and third lumbar level. Most of the patients with fourth lumbar level involvement were functional household or community ambulators compared to the third lumbar level patients, who were mostly nonfunctional ambulators (38). In the same study, 20 of 21 patients with fifth lumbar or sacral level of involvement were community ambulators.

Maintenance of walking ability as an adult also correlates with the functional level of involvement. A review of 29 adult myelomeningocele patients aged 20 to 43 years found 95% of patients with third lumbar level of involvement or lower remained ambulatory (39). In contrast, only 22% of patients with second lumbar level of involvement or higher remained ambulatory. The difficulty with maintaining the ability to walk as an adult relates to the high energy cost required to walk. Also, in patients with high level of involvement, there is a high incidence of spinal deformity requiring surgical treatment. Hip and knee flexion contractures are also common and prone to recurrence as an adult despite aggressive treatment during childhood (17).

Correlating with functional level of involvement, one of the most important physical factors for maintaining ambulation is the strength of the quadriceps and the hamstrings muscles (38, 40). Seitzberg et al. (40) looked at a group of 32 patients with myelomeningocele and found a significantly better chance for maintaining ambulation as an adult if quadriceps strength was at least grade 4 during childhood. They also found that overall patients with grade 3 or higher hamstring function during childhood had a significantly better chance for adult ambulation. However, they noted that hamstring function was not relevant in patients with normal quadriceps strength (40). Another study of 109 patients also found a correlation between quadriceps strength and ambulatory ability (41). In this group, 82% of patients with grade 4 or higher quadriceps power were community ambulators, whereas 88% of patients with grade 2 or less were not functional ambulators.

The strength of the iliopsoas muscle has also been shown to be important for ambulation. McDonald et al. (42) looked at a group of 291 patients with an average age of 14.5 years and found that 100% of the patients with symmetrical grade 4 or 5 iliopsoas strength were ambulatory. In contrast, 89% of the patients with iliopsoas strength grade 3 or less were nonambulatory.

Sitting balance is a factor that can be assessed at a young age and has also been shown to be predictive of ambulatory potential in patients with higher levels of involvement. The ability to sit without hand support indicates nearly normal functioning of the central nervous system. When hand support is needed for sitting, use of an orthosis and external support for ambulation is likely to be severely impaired (17). A study of 206 patients with myelomeningocele confirmed that sitting balance was an independent predictor of community ambulation (43). In this study, lumbar and sacral level patients with no sitting-balance deficit and sacral level patients with a mild sitting-balance deficit were likely to be independent ambulators.

FUNCTIONAL CLASSIFICATION

The best known and most widely used classification of myelomeningocele is based on the neurologic level of the lesion (43, 44). Four main groups are identified based on the level of the lesion and associated functional and ambulatory capability (Table 15-1).

Thoracic/High-Lumbar Level of Involvement. The first group includes the thoracic and high-lumbar level patients, which represents approximately 30% of patients with myelomeningocele. This group is defined by the lack of functional quadriceps activity and has a neurologic level of L3 or above (43). To achieve ambulation during childhood, patients in this group require bracing to the level of the pelvis with either a reciprocating gait orthosis (RGO) (Fig. 15-1) or a hip–knee–ankle–foot orthosis (HKAFO) (Fig. 15-2). The majority of patients in this group, between 70% and 99%, require a wheelchair for mobility as an adult (17, 45). The inability to maintain community ambulation in adulthood relates to the high energy cost required to achieve ambulation with either an RGO or an HKAFO.

Low-Lumbar Level of Involvement. The next group, approximately 30% of patients with myelomeningocele, has

TABLE 15-1 Functional Classification of Myelomeningocele

Group	Neurologic Level of Lesion	Prevalence	Functional Capacity	Ambulatory Capability	FMS
Thoracic/high lumbar	L3 or above	30%	No functional quadriceps (≤ grade 2)	During childhood, require bracing to level of pelvis for ambulation (RGO, HKAFO) 70%–99% require wheelchair for mobility in adulthood	1,1,1
Low lumbar	L3–L5	30%	Quadriceps, medial hamstring ≥ grade 3. No functional activity (≤ grade 2) of gluteus medius and maximus, gastrocsoleus.	Require AFOs and crutches for ambulation 80%–95% maintain community ambulation in adulthood	3,3,1
High sacral	S1–S3	30%	Quadriceps, gluteus medius ≥ grade 3 No functional activity (≤ grade 2) of gastrocsoleus	Require AFOs for ambulation 94%–100% maintain community ambulation in adulthood	6,6,6
Low sacral	S3–S5	5%–10%	Quadriceps, gluteus medius, gastrocsoleus ≥ grade 3	Ambulate without braces or support 94%–100% maintain community ambulation in adulthood	6,6,6

low-lumbar level of involvement. Functionally patients in this group have purposeful (grade 3 or higher) quadriceps and medial hamstring activity but lack purposeful activity (below grade 2) of the gluteus medius, gluteus maximus, and gastrocsoleus muscles. Hence, these patients require braces to control the position of the foot and ankle as well as crutches or a walker in order to ambulate. Between 80% and 95% of patients in this group maintain community ambulation in adulthood, but

FIGURE 15-1. Reciprocating gait orthosis (RGO) with a reverse walker.

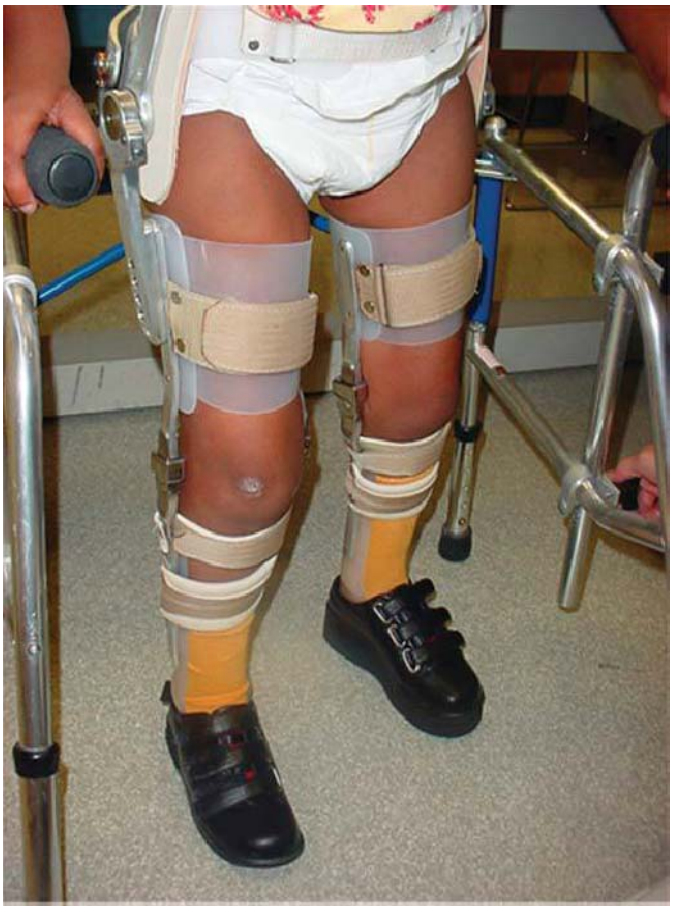

FIGURE 15-2. Hip–knee–ankle–foot orthosis (HKAFO).

most will use a wheelchair for long-distance mobility (43, 45). This group includes patients from L3 to L5 level of involvement, although patients with L3 level of involvement represent a transitional population and are included in this group only if they have evidence of strong quadriceps and medial hamstring function (43). Since medial hamstring function is needed for community ambulation, there is a significant difference in the ability to walk between children with L3 and L4 level of involvement (38). Because of this, children with L4 level of involvement have the most potential benefit from proper orthopaedic care of musculoskeletal deformities. Aggressive treatment of hip contractures; rotational malalignment of the tibia; and deformities of the knee, ankle, and foot are essential to maintain functional ambulation.

High-Sacral Level of Involvement. Patients with high-sacral level of involvement represent approximately 30% of patients with myelomeningocele. Patients in this group have functional activity in the quadriceps and gluteus medius (grade 2 or higher) but lack functional activity in the gastrocnemius-soleus. Patients with high-sacral level walk without assistive devices but do require an ankle–foot orthosis (AFO) (Fig. 15-3). These children have a characteristic gluteus lurch with excessive pelvic obliquity and rotation during gait.

Low-Sacral Level of Involvement. The last group of patients, approximately 5% to 10% of patients with myelomeningocele, has low-sacral level of involvement. These patients also have both quadriceps and gluteus medius function, but are distinguished from the high-sacral level patients based on the presence of gastrocnemius-soleus functional activity. Patients with low-sacral level of involvement walk without braces or assistive devices and have a gait pattern that is close to normal gait because they have normal gluteus medius and maximus function.

Between 94% and 100% of patients with sacral level involvement maintain community ambulation as adults (38, 45, 46). In this group, aggressive treatment of tethered cord syndrome; avoidance of arthrodesis in the foot; and treatment of deformities of the knee, ankle, and foot are important to promote functional ambulation.

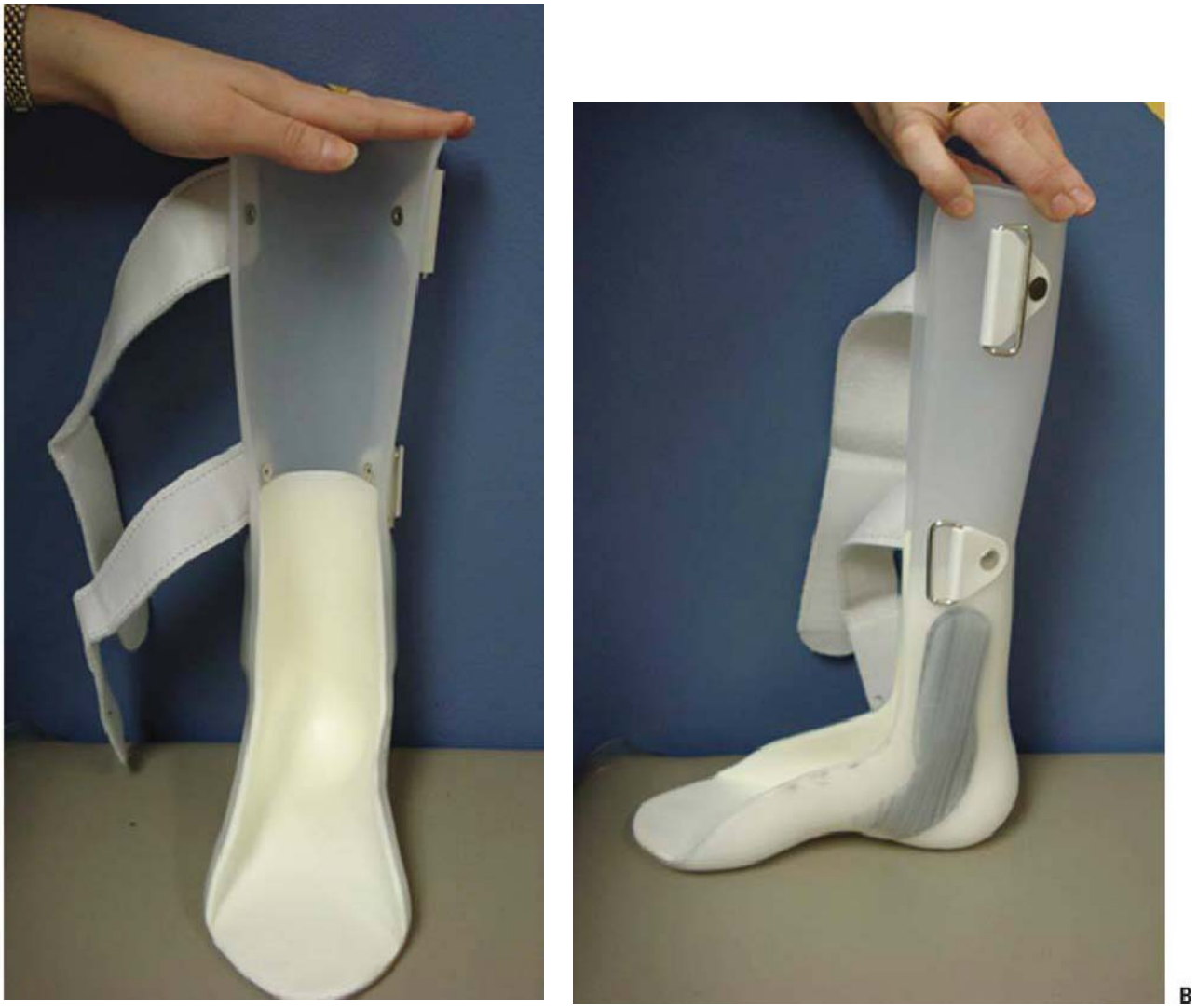

FIGURE 15-3. Ankle–foot orthosis (AFO), front view **(A)** and side view **(B)**.

FUNCTIONAL MOBILITY SCALE

Many instruments specific to the pediatric population exist to assess quality of life, health status, physical function, and mobility in patients with physical disabilities. However, many of these instruments, such as the Pediatric Outcome Data Collection Instrument and the Child Health Questionnaire, are time consuming to administer and analyze. Because of this, the Functional Mobility Scale (FMS) was described in 2004 as a useful, simple tool to describe the more focused issue of functional mobility in children with disabilities and to aid communication between orthopaedic surgeons and health professionals (47).

The FMS was initially devised to describe functional mobility in children with cerebral palsy, but the authors reported they had also successfully used it to assess children with myelomeningocele (47). Recently, the FMS was used in a study by Battibugli et al. (18) to compare function in groups of patients with myelomeningocele. The FMS is unique because it allows quick, practical scoring of mobility over three distinct distances representing mobility in the home (5 m), at school (50 m), and in the community (500 m). In this way, it is effective for distinguishing between groups of children with varying levels of disabilities and provides a means for standardized communication between health professionals (47). The FMS has also been found to be sensitive to detect change after operative intervention (47).

To apply the FMS, a child is given a score from one to six based on their walking ability for each of the three distances assessed (Table 15-2). A score of one is used when a child uses a wheelchair, two for a walker, three for the use of two crutches, four for the use of one crutch or two walking sticks, five for a child who is independent on level surfaces, and six for a child who is independent on all surfaces. Two additional possible ratings are C for a child who crawls for mobility in the home and N for a child who does not complete the given distance. For example, a child who ambulates with crutches at home and at school and uses a wheelchair for long distances but would be an FMS 3,3,1.

Use of the FMS allows for an accurate clinical picture of a given patient's functional status at a distinct point in time. Often parents or the patient may have difficulty choosing a single response to a question regarding function and will default to the highest level of function. This can impact interpretation of outcome studies if parents choose different responses at different time intervals when there has been little actual change in function (47). A major advantage of the FMS is its ability to account separately for distances representing home, school, and the community hence addressing the complexities of functional mobility in the real world.

TABLE 15-2 Functional Mobility Scale

Rating	Function
1	Uses a wheelchair
2	Uses a walker independently
3	Uses crutches independently
4	Uses one or two sticks independently
5	Independent on level surfaces
6	Independent on all surfaces
C	Crawls for the given distance
N	Does not complete the given distance

GAIT ANALYSIS

Gait analysis is defined as the systematic measurement, description, and assessment of quantities that characterize human locomotion (48). Clinical gait analysis has received a great deal of attention in regard to its application to the treatment of children with cerebral palsy. Increasingly gait analysis is also being recognized as a valuable component of the comprehensive orthopaedic evaluation of patients with myelomeningocele. Its use has been reported in the literature for assessing various manifestations of myelomeningocele including hip subluxation/dislocation, lower extremity contracture, and rotational abnormalities (49–53). Two main groups of patients with myelomeningocele can especially benefit from gait analysis: (a) patients with a low-lumbar lesion who walk with external support and a below-knee orthosis and (b) patients with sacral-level lesions who walk with no external support and AFOs (17). Studies have shown the average walking velocity for a patient with low lumbar-level involvement is 60% of normal (50). The average walking velocity for a patient with high sacral-level involvement is approximately 70% of normal (52).

The components of gait analysis may include kinematics, kinetics, electromyographic data, measurement of videotape recordings, energy expenditures, clinical observation, and foot pressure readings (48). The data obtained from these areas are presented as graphic and numerical data (for kinematics, kinetics, and ground-reaction forces), as electromyographic activity, and as videotape recordings. All of these data are then analyzed by a clinician with training in the interpretation of gait studies and a report of the gait analysis is generated. Several high-quality, commercial gait analysis systems are now available. The more comprehensive of these systems provide the clinician with three-dimensional kinematics and kinetics as well as dynamic electromyography (48). Three-dimensional gait analysis is especially useful for analyzing transverse plane deformities such as rotational problems. However, when a three-dimensional study is not available, the data obtained from a two-dimensional study have useful applications in the documentation of coronal and sagittal plane deformities such as crouch gait and foot deformities.

Kinematics describe the spatial movement of a body without consideration of the forces that cause the movement. These movements are linear and angular displacements, velocities, and accelerations. Kinematic data answer the question of what is happening at the level of each of the major lower extremity joints but not why it is happening (54). Kinematics are useful

in determining treatment outcome through the comparison of preoperative and postoperative gait analysis data.

Kinetics on the other hand describe the mechanisms that cause movement around a joint. Hence, kinetics answer the question of why a particular movement or gait deviation occurs (54). Kinetic data include ground-reaction forces, joint moments, and joint powers. In order to calculate kinetic data, simultaneous acquisition of joint motion and force-plate data is necessary (48). The study of kinetics leads to improved understanding and knowledge of the pathogenesis of gait patterns (54).

Gait analysis is useful in preoperative planning for ambulatory myelomeningocele patients because it allows accurate dynamic assessment of an individual patient's gait problems. Postoperatively gait analysis is used to obtain a much more accurate, objective, and quantitative assessment of outcome than was previously possible (54). Often a patient's true functional status differs from what would be expected based on information obtained during the static clinical examination. Moen et al. demonstrated this in a study examining crouch gait in myelomeningocele patients. They found significantly greater dynamic knee flexion during ambulation using gait analysis than what was measured on clinical examination. Gait analysis is a useful component of the comprehensive evaluation of ambulatory myelomeningocele patients, especially when surgical treatment is being considered.

With specific regard to patients with myelomeningocele, gait analysis is useful to assess the abnormal movements that occur as compensation for muscle weakness. For example, due to weakness of the gluteus medius and maximus muscles, compensatory movements at the pelvis and hip such as increased active pelvic rotation and stance phase hip abduction develop to facilitate forward progression of the limb and maintain independent ambulation. All children with low lumbar-level involvement show increased anterior pelvic tilt, but compensatory movements become less pronounced with lower levels of motor involvement (17).

Gait analysis is helpful in determining the course of treatment for patients with hip flexion-adduction contracture and low lumbar or sacral-level patients with unilateral hip subluxation or dislocation (50). Gait analysis has also proved useful in increasing the appreciation of the effects of rotational malalignment of the lower extremity. Specifically, it has helped with understanding the relationship between external tibial torsion and a significantly increased valgus stress at the knee joint (49). In addition, the information gained from gait analysis in regard to the coronal and transverse plane kinematics at the pelvis and hip and the coronal plane kinetics at the hip and knee is important in the prescription of effective orthotics and walking aids (53).

OVERVIEW OF ORTHOPAEDIC CARE

Over the past 30 years, the overall care of children with myelomeningocele has changed substantially in regard to all specialties including neurosurgery, urology, rehabilitation, orthotics, and orthopaedics. Specifically relating to orthopaedics, the advent of gait analysis in the late 1980s contributed to a better understanding of the underlying deformities and their effect on function. This has led to a shift in the focus of orthopaedic treatment from the goal of radiographic changes to functional improvement (55).

The main goal of orthopaedic care of a patient with myelomeningocele is to make the musculoskeletal system as functional as possible. As discussed earlier, walking ability is highly dependent on the neuromuscular level of the lesion. Whether or not ambulation should be the goal for every child with myelomeningocele is controversial. The role of the orthopaedic surgeon is to assist the patient and the family in developing realistic individualized goals and to provide the necessary care to meet these goals. Additionally, providers must help families to avoid neglecting the child's total development while focusing on the use of the lower extremities. Emphasizing intellectual and personality development utilizing wheelchair mobility, wheelchair sports programs beginning in preschool, and educational mainstreaming can lead to dramatically increased independence (17).

Both congenital and acquired orthopaedic deformities are seen in patients with myelomeningocele. Congenital deformities are present at birth and include kyphosis, hemivertebrae, teratologic hip dislocation, clubfoot, and vertical talus. Acquired developmental deformities are related to the level of involvement (4) and are caused by paralysis, decreased sensation in the lower extremities, and muscle imbalance (34). For example, calcaneus foot and hip dislocation are two acquired orthopaedic deformities caused in part by muscle imbalance. Orthopaedic deformities may also be result from iatrogenic injury such as postoperative tethered cord syndrome. Accordingly, the orthopaedic surgeon must monitor spinal balance and deformity and assist with monitoring the neurologic status of each patient.

The newborn examination of a patient with myelomeningocele should include identification of the level of paralysis for each extremity. Any associated conditions such as clubfoot or hip or knee contractures should be recognized and treated appropriately. In addition, a manual muscle test should be performed by a skilled physical therapist to evaluate the neurologic level of function. This should be done before closure of the spinal defect, again 10 to 14 days after closure, and then on an annual basis. Since a given patient's motor level should remain the same throughout their lifetime, a change in muscle strength may be a sign of tethered spinal cord.

After the initial newborn examination, orthopaedic follow-up should occur regularly every 3 to 4 months during the 1st year of life. After that, patients are seen every 6 months until the age of 11 or 12 years after which time patients are followed annually. The follow-up periodic orthopaedic examination should include assessment and monitoring of motor and sensory function, spinal alignment, and skin integrity. Orthoses should be inspected on a regular basis to ensure appropriate fit with no areas of irritation or pressure points on the skin. Patients with myelomeningocele have multiple

medical comorbidities that must be considered as part of any orthopaedic treatment. Because of this, orthopaedic care should ideally be administered as part of a multidisciplinary team including neurosurgery, urology, and physiatry.

ORTHOPAEDIC MANAGEMENT

Spine. Spinal deformities such as scoliosis and kyphosis have a high prevalence in patients with myelomeningocele. Spinal deformity may present as a developmental deformity that is acquired and related to the level of paralysis, as a congenital deformity resulting from malformations such as hemivertebrae or unsegmented bars, or as a combination of both (4). The frequency of spinal deformity correlates with level of neurologic involvement. Hence, patients with a high-level lesion should have radiographs of the spine at least annually to evaluate any deformity. Patients with low-lumbar or sacral level of involvement have a low incidence of scoliosis; hence, any abnormal curvature in these patients should alert the caregiver to the possibility of an underlying tethered cord.

Scoliosis. Developmental scoliosis typically presents with a long, sweeping, C-shaped curve with the convexity often on the opposite side of the elevated pelvis (4). Overall, the prevalence of scoliosis in patients with myelomeningocele is reported to be between 62% and 90% (56–59). Many factors have been identified in patients with myelomeningocele that correlates to the development of scoliosis. One important factor is the functional level of involvement (4, 56, 59, 60). Trivedi et al. applied rigid criteria to a population of 141 patients in order to define incidence of developmental scoliosis defined as a Cobb angle >20 degrees. They found the prevalence of scoliosis to be 93% in patients with thoracic functional level, 72% in upper lumbar, 43% in lower lumbar, and <1% in sacral level patients (59). Other important factors in predicting development of scoliosis are ambulatory status (4, 56, 61) and the level of the last intact laminar arch (57, 59, 62). Less important predictive factors are hip dislocation/subluxation and lower extremity spasticity (4).

Scoliosis typically develops gradually in patients <10 years of age and then increases rapidly with the adolescent growth spurt. When a curve develops in a child younger than 6 years of age, it may be related to an underlying hydromyelia or a tethered cord syndrome. Muller et al. (61) found that curve progression was related to size of the curve with curves <20 degrees progressing slowly. In contrast, curves >40 degrees progressed severely and quickly at almost 13 degrees per year.

For a patient with curve magnitude <20 degrees, the recommended treatment is observation with follow-up radiographs every 4 to 6 months (4). When curve magnitude exceeds 20 degrees, brace treatment can be considered but has been controversial in patients with myelomeningocele. There is general consensus that brace treatment does not halt curve progression. However, the goal of using a brace in this population is not to correct the deformity but rather to support the trunk in a functional position and control the curve during growth hence possibly delaying the need for surgical stabilization (63). Bracing typically consists of a custom-molded thoracolumbosacral orthosis used during the day. Ideally, the brace should assist with sitting balance and free the hands for functional usage but not interfere with pulmonary function, lower extremity bracing, self-catheterization, or sitting (4). When an orthosis is prescribed for a patient with myelomeningocele, it is imperative to ensure proper fitting and counsel patients to have their skin assessed daily to prevent skin complications.

Indications for operative treatment of scoliosis in patients with myelomeningocele have not been strictly defined. Most agree that progressive curves with magnitude >50 degrees that interfere with sitting balance warrant surgical treatment. Given the high risk of complications in patients with myelomeningocele, such as infection and pseudoarthrosis, surgical treatment should be considered on an individual basis. The functional consequences of spinal surgery on ambulation, motor skills, and activities of daily living should be reviewed. Some studies specifically evaluating the effect of spinal fusion on ambulatory ability have suggested ambulation may be more difficult following surgery (64–66). Furthermore, multiple studies have also shown no significant difference in the ability to perform activities of daily living after surgical intervention (64–66). However, with newer instrumentation and changes in postoperative management, considerable improvement in final outcome is possible (55).

The goal of surgical treatment of spinal curvature in patients with myelomeningocele is to prevent further deformity and create a stable, balanced spine while avoiding complications (31). Multiple studies have established that combined anterior and posterior instrumented arthrodesis is the treatment of choice in most patients to achieve fusion and provide the best long-term correction (31, 67–71). In combination with the posterior approach, the anterior approach allows for diskectomy to improve curve flexibility and anterior interbody fusion to increase strength of the fusion mass (4). Use of the posterior approach alone for instrumented arthrodesis has shown high failure rates with hardware complications and subsequent loss of correction (71, 31). Anterior arthrodesis and instrumentation alone may be considered for a select group of patients with a thoracolumbar curve <75 degrees, compensatory curve <40 degrees, no increased kyphosis, and no syrinx (72).

When planning for correction, the fusion should include all curves and should extend from the upper thoracic vertebrae to the sacrum in nonambulators (4, 31).

DUNN-MCCARTHY PELVIC FIXATION An alternative to the Galveston type of pelvic fixation is that described by McCarthy et al. (73). In this technique, the ends of the Luque rods are prebent to fit over the sacral ala in the manner of large alar hooks (Figs. 15-4 to 15-6). This technique may be indicated particularly when the pelvis is very thin or small. It is mechanically at its best in the correction of kyphosis and is contraindicated in lordosis.

The end of the rod that is to fit over the sacral ala must be bent before the operation. The tight bends necessitate that the

Dunn McCarthy Pelvic Fixation (Figs. 15-4 and 15-5)

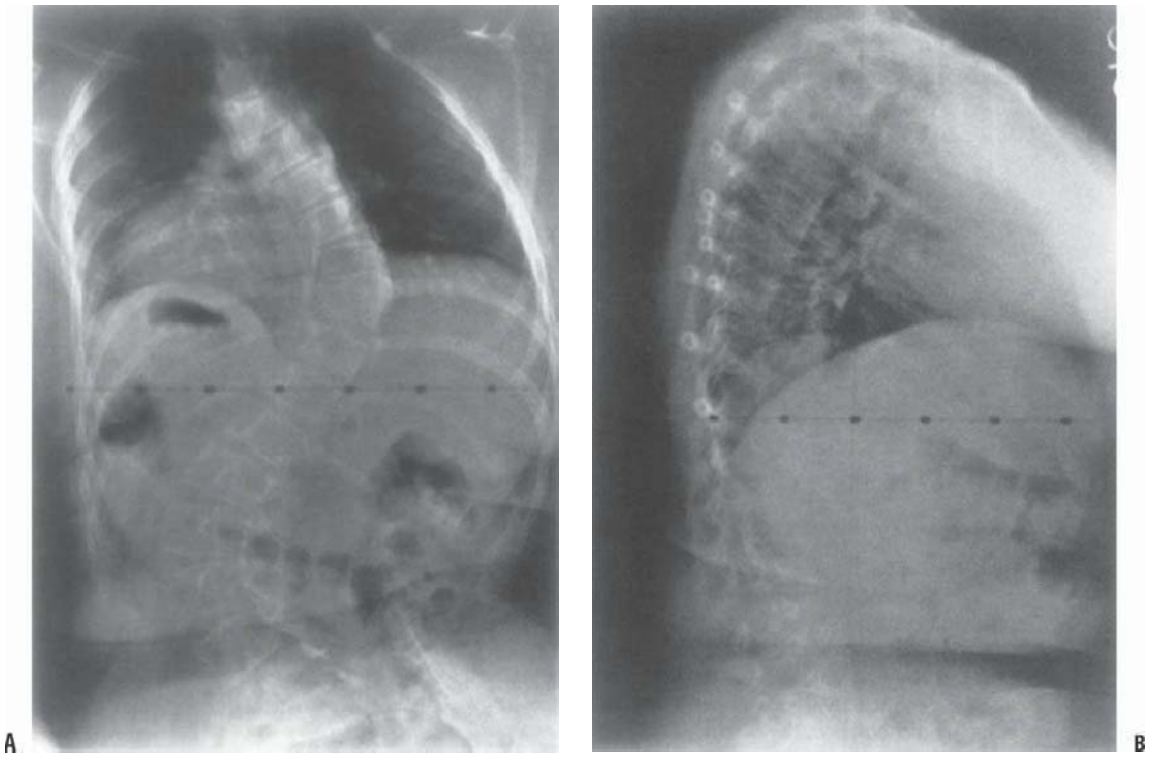

FIGURE 15-4. **Dunn McCarthy Pelvic Fixation.** Measurement from the preoperative radiographs aids in achieving the correct dimensions of the bends. The first consideration is that the midportion of the sacral ala is lateral to the midportion of the lamina. This amount of lateral offset in the rod **(A)** can be estimated by measuring the distance from the midportion of the L5 lamina to the midportion of the sacral ala. In the typical patient, this is about 1 to 1.5 cm. The width of the segment that is to go over the sacral ala **(B)** is measured from the lateral radiograph of the pelvis. This width is usually between 1 and 1.5 cm. When this procedure is used in the bifid myelodysplastic spine, careful preoperative planning is necessary to be sure that the rod lies in the desired position. At surgery, the sacral ala is cleaned as it would be for lumbosacral arthrodesis. It is important that the hook portion of the rod passes anterior to the alae, thus necessitating the dissection be carried out slightly more anterior than usual. Before seating the rod, it should be possible to pass a finger around the front of the alae.

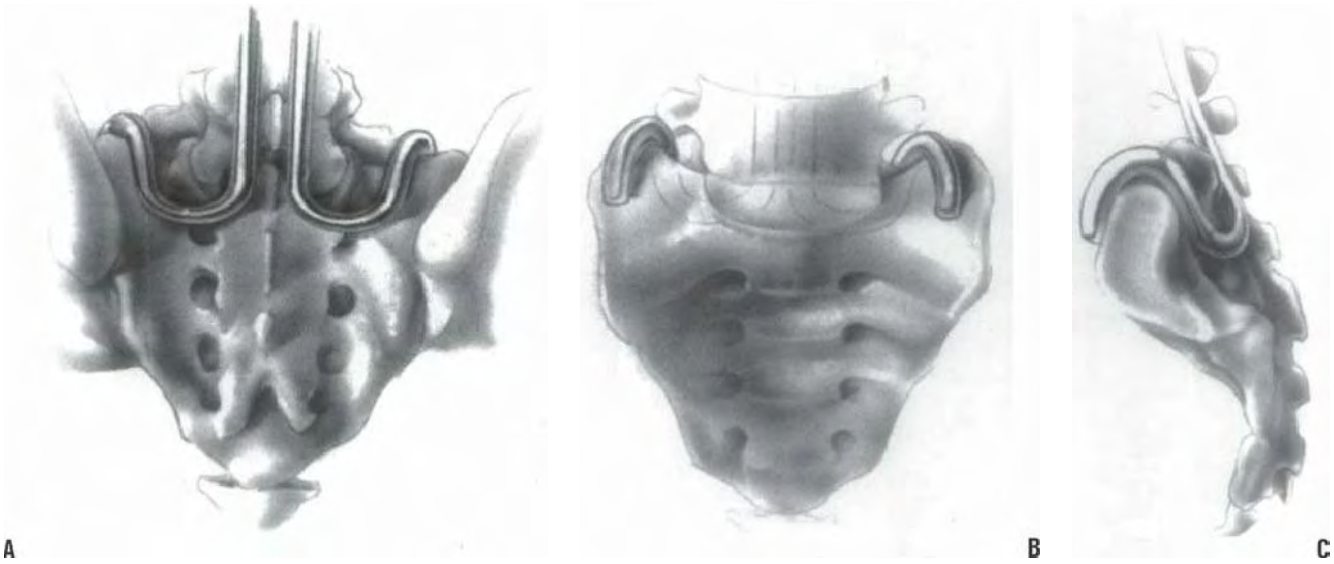

FIGURE 15-5. **A-C:** The prebent portion of the rod is hooked on the ala like a giant sacral hook. It does not penetrate the cortex. It is possible to make minor adjustments to the rods during surgery, but it is not possible to bend all of the necessary curves into the rod on the operating room. Contouring lordosis into the sacral segment of the rod positions it more firmly against the sacral alae. Use of the Texas Scottish Rite crosslinks on the spinal segment of the rods prevents movement of one rod in relation to another and provides a rigid construct. The rod is held in place over the sacral alae by the sublaminar wires.

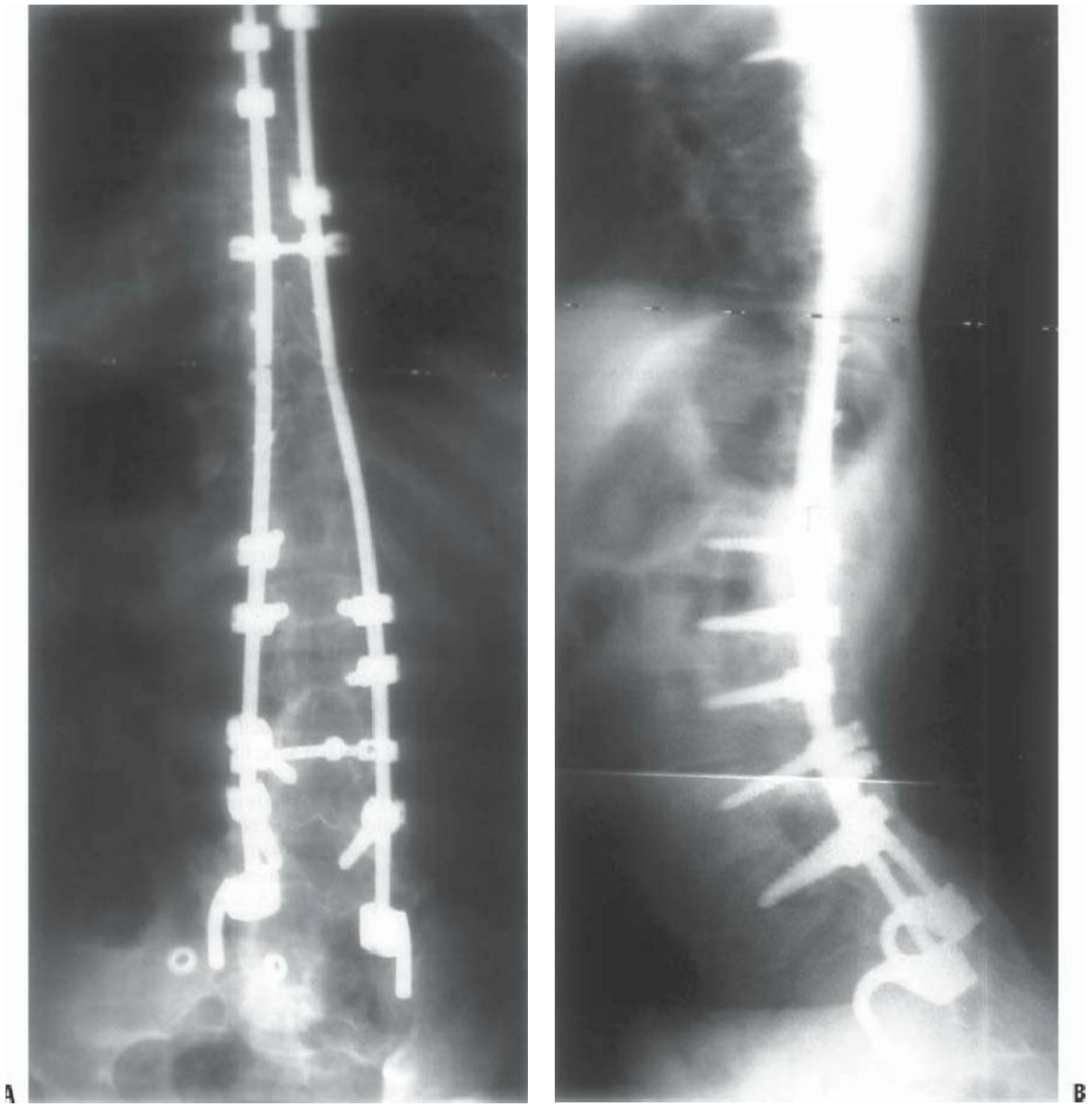

FIGURE 15-6. Anteroposterior **(A)** and lateral **(B)** radiographs after posterior arthrodesis and instrumentation with the Dunn-McCarthy technique in a child with myelomeningocele. Note the sacral alar hooks that connect independently to the rods. (Courtesy of Richard McCarthy, MD, Little Rock, AR.)

rods be heated over a flame to soften the metal before bending. Bends of two different dimensions can be made, one at each end of a long Luque rod. The end that fits less well can be cut off at surgery and discarded. It is necessary that the rods be bent so that they are mirror images of each other . These rods are available prebent.

A new variation of this method is now available using a special sacral hook that is made of titanium and designed to be used with a titanium rod. This allows for ease of use since contouring of the rod can be done independently of the sacral portion. It is necessary to use this hook in conjunction with a pedicle screw above (usually at least one screw at L4) so that distraction firmly seats the sacral hook in place. This technique can be used in a variety of neuromuscular spinal deformities in which fixation to the sacrum is required.

In ambulatory patients, preserving pelvic motion is important for function; hence, whenever possible, lumbosacral fusion should be avoided. Pedicle screw instrumentation offers some advantages in patients with myelomeningocele in terms of correcting scoliosis while preserving lumbar lordosis and lumbar motion in ambulatory patients (74). However, difficulties with pedicle screw instrumentation may be encountered in patients with small, tightly packed vertebrae in lordotic segments or with small, dysplastic, and rotated pedicles. In these cases, familiarity with other instrumentation constructs such as multihook systems or sublaminar wires is necessary.

An important consideration for patients with myelomeningocele and scoliosis is the increased risk of complications associated with spine surgery in this population. Complications encountered with frequency include hardware problems such as implant failure, dislocation, and pseudoarthrosis, infections, postoperative lower extremity fractures, and neurologic complications. Hardware problems have been reported in approximately 30% of patients and often lead to loss of correction (31, 75). Pseudoarthrosis rates have been reported as high as 76% and are dependent on the approach and instrumentation utilized, with the highest rates associated with isolated posterior fusion (37, 66, 67, 69, 71). Wound infection and incisional necrosis are common and correlate with the incision used. The triradiate incision has been associated with a 40% rate of skin necrosis and should be avoided (69). The risk of wound infection is increased by the presence of a concurrent urinary tract infection, common in this population. For this reason, preoperative urinary cultures should be obtained. Lower extremity fractures due to disuse osteoporosis have been reported in up to 29% of patients in the first 6 months after surgery (66). Neurologic deficit occurs with a low frequency but can be permanent (66, 69).

Kyphosis. In patients with myelomeningocele, an associated kyphotic deformity is present in 8% to 21% of patients and occurs most commonly in the upper lumbar or thoracolumbar region (76–80). Patients may present with a large, rigid curve at the time of birth, often exceeding 80 degrees (31, 78). Progression of the curve has been related to the level of the neurologic lesion (75) and ranges from 4 to 12 degrees each year (31, 77–79). The natural history of rigid congenital kyphosis is rapid progression, especially after the 1st year of life when the child begins to sit (4). The apex of the curve is usually located in the upper lumbar spine (78). Rigid curves may be associated with vertebral anomalies, a sharp apical angulation, and the potential for skin breakdown over the prominence of the deformity (81). Development of trunk control and sitting balance can lead to the development of compensatory thoracic lordosis in older patients (80).

Treatment of rigid kyphosis is indicated to prevent progression of deformity, correct abnormal sitting posture, and prevent skin breakdown over the apex of the deformity (4). Conservative treatment using bracing and/or modified wheelchair seating systems has been largely ineffective (82). Surgery has been recommended as the treatment of choice; however, absolute criteria have not yet been well defined in the literature for indications and timing of surgical treatment, extent of resection and fusion, or type of instrumentation. Kyphectomy with osteotomy and resection of the vertebral bodies and spinal fusion has been the standard surgical treatment (83). Kyphectomy has been one of the most challenging procedures for spine surgeons and has been associated with high complication and mortality rates (78, 31, 82, 55) (Fig. 15-7). Improvements in final outcome have been seen using newer techniques, such as early intervention, longer fusion, and the decancellation described by Lindseth and Stelzer (78). With this technique, the authors reported persistent correction with the potential for continued growth of the remaining lumbar vertebrae increasing the capacity of the abdominal cavity (78).

HIP

Deformity about the hip is very common in patients with myelomeningocele and may consist of hip joint contractures, subluxation, or dislocation. The development of hip deformity is related to the patient's neurologic level of involvement. For each type of hip deformity, treatment depends on the level of neurologic involvement, the type of deformity present, and the functional capacity of the patient (31).

Hip Contractures. Several factors contribute to the development of hip contractures in patients with myelomeningocele including muscle imbalance, positioning, and spasticity (31, 84). Muscle imbalance plays a major role, as seen in a patient with low-lumbar level of involvement who lacks normal strength in the gluteal muscles. In this case, the relatively greater strength in the hip flexors and adductors leads to deformity about the hip. The type and severity of contracture depends in part on the degree of muscle imbalance present (84). Positioning is a contributing factor especially in patients with high levels of involvement who rely on wheelchairs for mobility (31). Spasticity of the hip musculature may be seen in patients with tethered cord syndrome.

Hip contractures and the resultant loss of motion can affect a patient's function more than hip subluxation or dislocation. If not treated properly, pelvic obliquity and compensatory spinal deformity may result (34). In ambulatory patients, hip flexion contracture causes the patient to stand with increased lordosis leaning forward to use the arms for support resulting in greater energy cost (85). The effect of hip contractures on gait has been documented with gait analysis. Gabrieli et al. found that patients with unilateral hip flexion and/or adduction contractures had increased pelvic obliquity leading to asymmetric gait and compensatory scoliosis. The authors concluded that a symmetric gait pattern was related to absence of hip contracture or bilateral symmetric hip contractures but had no relation to hip dislocation. Current treatment goals based on studies of functional results focus on maintaining hip range of motion with contracture release, especially unilateral hip adduction and flexion contractures (50, 85–87).

The routine clinical examination of a patient with myelomeningocele should include the Thomas test to assess for hip flexion contracture. Because hip flexion deformity tends to decrease in the first 2 years of life, except in patients with high levels of involvement, treatment is rarely indicated in this age group. Specific treatment recommendations are based on a patient's functional level of involvement. In patients with thoracic or high-lumbar levels of involvement, flexion contracture of up to 30 to 40 degrees may be tolerated as long as it does not interfere with orthotic use and ambulation. In a high level patient attempting to walk with a RGO, more

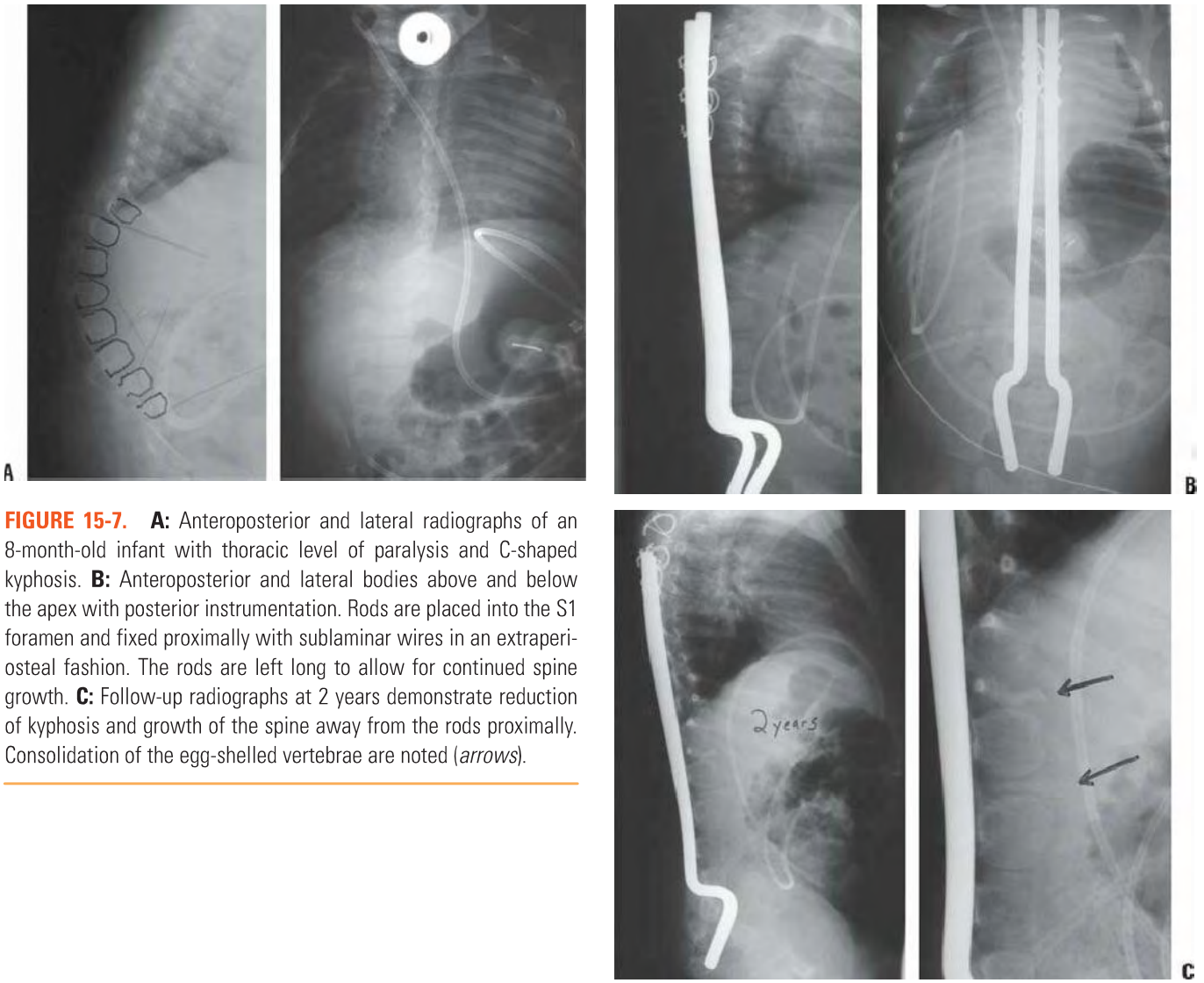

FIGURE 15-7. **A:** Anteroposterior and lateral radiographs of an 8-month-old infant with thoracic level of paralysis and C-shaped kyphosis. **B:** Anteroposterior and lateral bodies above and below the apex with posterior instrumentation. Rods are placed into the S1 foramen and fixed proximally with sublaminar wires in an extraperiosteal fashion. The rods are left long to allow for continued spine growth. **C:** Follow-up radiographs at 2 years demonstrate reduction of kyphosis and growth of the spine away from the rods proximally. Consolidation of the egg-shelled vertebrae are noted (*arrows*).

severe hip flexion contractures may cause a very short stride length and increased lumbar lordosis. Treatment is indicated to provide sufficient range of motion to allow the patient to sit comfortably in a wheelchair, lie supine in bed, and use an orthosis for standing and walking (31). Soft-tissue release is performed through an anterior approach and usually includes the sartorius, rectus femoris, iliopsoas, and tensor fascia latae. If needed, the anterior hip capsule can be divided as well. To prevent recurrence of contracture, physical therapy is necessary to maintain range of motion and a total body splint (TBS) can be used at night-time. In very severe cases with deformity >60 degrees, proximal femur extension osteotomy can be used, especially if pressure sores result from the hip deformity (31, 85).

For patients with low-lumbar level of involvement, lesser hip flexion contractures can result in major functional impairment. In such a patient who walks with AFOs and crutches, a hip flexion contracture of >20 degrees can lead to significant anterior pelvic tilt causing decreased walking velocity and increased demand on the upper extremities (50, 52). When surgical treatment is indicated in this group, care must be taken to preserve hip flexor power. For contractures >20 degrees that interfere with function, the tensor fascia latae and the rectus femoris are released. The sartorius is detached from the anterosuperior iliac spine and reattached to the anterioinferior iliac spine. If iliopsoas lengthening is necessary, it is done so in an intramuscular fashion above the pelvic brim.

When adduction contracture is present and interferes with function, treatment includes myotomy of the adductor longus and gracilis. The adductor brevis is included if necessary. A subtrochanteric valgus osteotomy of the proximal femur may be necessary in severe cases in order to achieve sufficient abduction to improve pelvic obliquity. Abduction contractures usually respond well to the Ober-Yount procedure (88, 89). Cast immobilization after release of hip contractures is unnecessary. A TBS is used full time for the initial 10 days followed by early mobilization and night-time usage of the splint.

Hip Subluxation/Dislocation. Hip instability affects up to one-half of patients with myelomeningocele during the first 10 years of life with either hip dislocation or subluxation

(17, 31). Treatment of this common and complicated problem remains a controversial issue. In the 1960s and 1970s, an aggressive treatment approach was advised, and the procedure of choice was transfer of the iliopsoas tendon (90, 91). Other approaches used included the external oblique transfer and varus osteotomy of the femur. The goal of treatment was anatomic reduction of the hip. Then, in 1978, Feiwell et al. (88) described the importance of a level pelvis and adequate range of motion of the hips rather than anatomic reduction of the joint. Since then, the focus has shifted from obtaining radiographic reduction of the hip to achieving maximal functional results (85, 86). Data from gait analysis support this approach (50). Modern treatment of hip instability is based on the patient's functional level of involvement and consists largely of maintaining hip range of motion with contracture release only.

With the earlier treatment approach for hip instability, reconstruction was offered to both ambulatory and nonambulatory patients. Reported rates of success or failure were often based solely on anatomic and radiographic results with little regard given to the functional consequences of surgical treatment. Subsequently, concerns developed over whether radiographically successful hip reduction led to decreased range of motion and pathologic fractures compromising functional results (92). Feiwell et al. (88) compared functional results in patients who had undergone hip reduction to those who had not and found no improvement in range of motion or ability to ambulate in those who had undergone surgery. In addition, they found surgery did not lead to decrease in pain or need for bracing.

Rather, multiple studies have demonstrated a high complication rate leading to decreased ambulatory function in patients who have undergone surgical reduction of hip dislocation. Sherk et al. (93) compared a series of patients who had undergone surgical treatment of dislocation to those who had not and found 36% in the surgically treated group had worsened ambulatory capacity as a result of surgical complications. Worsening neurologic deficit has also been reported after surgical treatment of hip dislocation (92). Another series reported a high complication rate in surgically treated patients with 29% incidence of loss of motion and 17% with pathologic fractures (87).

There is general agreement in the literature that ambulatory ability does not depend on the status of the hip, but instead the most important factor in determining ambulation is the level of functional involvement (39, 85, 87, 94, 93). Preserving muscle strength of the iliopsoas and quadriceps is more relevant to potential for continued ambulation in adulthood than the status of the hip joint.

For patients with thoracic and high-lumbar levels of involvement, the stability of the hip joint has little clinical effect on function (85, 87, 93). Treatment should be limited to contracture release to allow for proper sitting posture, perineal care, and facilitate use of orthoses for ambulation. Convincing evidence does not exist to support hip reduction in this group of patients.

There is a high incidence of hip instability in patients with low-lumbar levels of involvement because of underlying muscle imbalance. Using gait analysis, it has been shown that hip instability in this group of patients has minimal effect on gait symmetry (50). In addition, the walking speed of patients with a unilateral hip dislocation was 60% of normal, which corresponds to that of patients without hip dislocations in previous studies from the same center (50). Hence surgical relocation of the unilaterally unstable hip in patients with low-lumbar level of involvement is not recommended. As discussed above, unilateral soft-tissue contractures should be treated to maintain a level pelvis and flexible hips (50, 85).

Hip instability in patients with sacral level of involvement is relatively rare but presents a challenging treatment dilemma. Hip dislocation in a patient who walks with no support can lead to level arm dysfunction (50, 55). Patients may develop an increased lurch due to the loss of a fulcrum from the dislocated hip (95). Patients in this group place high demand on the hip and have functional hip abductor strength that can be compromised with hip instability (50). Careful consideration should be given to surgical reduction in this group as a concentric reduction may help to maintain independent ambulation into adulthood, prevent asymmetry in gait, and preserve the integrity of the hip joint (50). When surgical treatment is being considered, a computerized tomography scan of the hips with three-dimensional reconstructions may be useful for preoperative planning to better assess acetabular deficiency and select the most appropriate type of pelvic osteotomy. Capsular plication is indicated when laxity is present, and rotational malalignment of the femur should be corrected at the same time. Excessive varus should be avoided to preserve hip abductor function for stability during stance and foot clearance during swing. Further studies are necessary to better assess the results of surgical treatment of hip instability in this select group of patients.

Hip Stiffness. Severe stiffness of the hip joint in patients who have undergone attempted surgical treatment presents a major problem (85). One option for treatment is the Castle procedure that entails resection of the proximal femur past the level of the lesser trochanter (96). A capsular flap is then closed across the acetabulum, and the quadriceps muscle is sutured around the resected end of the femur. The goal of the procedure is to allow patients improved range of motion for function, but disadvantages include the need for postoperative traction and high risk of postoperative heterotopic ossification.

The McHale et al. (97) procedure is another option for treatment of this serious complication. This procedure consists of femoral head resection with a valgus subtrochanteric osteotomy of the femur. In the author's experience in patients with cerebral palsy, this allows good range of motion in flexion, extension, abduction, and adduction leading to improved sitting ability and ease of perineal care. Postoperatively, we use the TBS (Fig. 15-8) instead of cast immobilization to allow early range of motion and easier care of the patient.

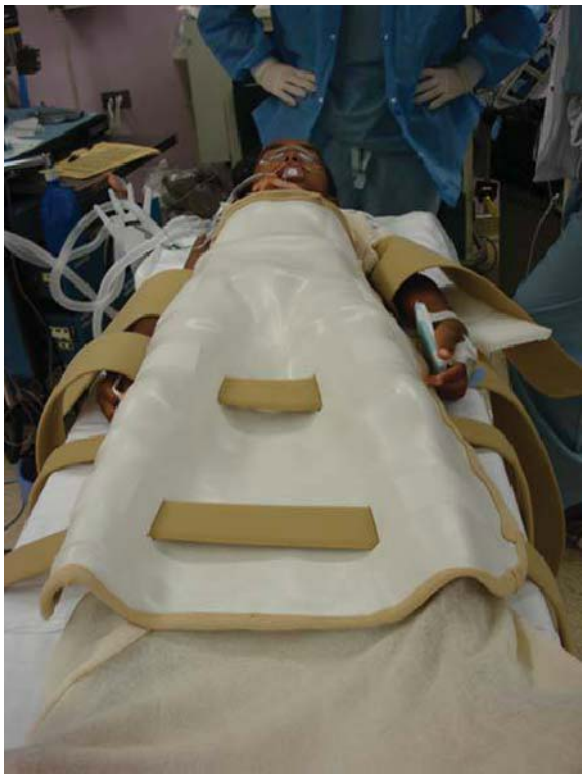

FIGURE 15-8. Total body splint.

KNEE DEFORMITIES

The two most common deformities about the knee in patients with myelomeningocele are knee flexion contracture and knee extension contracture. Other less commonly seen deformities are knee valgus deformity, knee varus deformity, or late knee instability with pain. Contractures occur most commonly in patients with thoracic and high lumbar level of involvement, and less often in patients with low lumbar level involvement (34). Deformity at the knee joint may occur as a result of many contributing factors, including static forces of positioning, fibrosis of surrounding muscles, muscle imbalance around the knee joint, and fracture malunion (95).

Knee Flexion Contracture. At birth, flexion contracture of the knee is a common finding in healthy newborns that often resolves during the first 6 months of life (98). This is in contrast to the fixed knee flexion contracture that can occur in both ambulatory and nonambulatory patients with myelomeningocele. Generally, more severe contractures are present in patients with thoracic level of involvement compared to those with lumbar level involvement (98–101). Early splinting can help to prevent knee flexion contracture in patients with high-level lesions.

The etiology of knee flexion contracture is multifactorial and may result in part from the typical supine positioning of patients with the hips abducted, flexed, and externally rotated and the knees flexed. Another factor relates to underlying quadriceps weakness combined with prolonged time spent in a sitting position that leads to a gradual contracture of the hamstrings and biceps femoris and eventually contracture of the posterior knee capsule. Spasticity and contracture of the hamstrings may also result from tethered cord syndrome. In ambulatory patients, quadriceps weakness combined with paralysis of the gastrocnemius-soleus and gluteus muscles leads to flexion at the knee. Finally, flexion deformity at the knee may be exacerbated by fracture malunion (102).

In most nonambulatory patients, knee flexion contracture does not have a major impact on mobility or ability to transfer. However, in ambulatory patients, knee flexion contracture causes crouch gait, which has a high energy cost. Increased knee flexion during ambulation leads to increased oxygen cost and less efficient ambulation (51). Flexion deformity of >20 degrees has been shown to interfere with orthotic fitting, which can prevent the patient from being upright and ambulating (99). Gait analysis is useful in quantifying the amount of knee flexion during ambulation, which can differ from that seen on a static clinical examination. Using computerized gait analysis, one study found the degree of actual knee flexion during gait was significantly greater than the degree of clinical contracture (51). This information is useful in evaluating patients and planning proper treatment.

Because of the increased energy cost of a crouched gait, surgical treatment of knee flexion contracture is indicated when contracture exceeds 20 degrees in a patient with ambulatory potential (51, 99). Contracture release may also be indicated in nonambulatory patients if the fixed flexion position interferes with sitting balance, standing to transfer, or transfer from chair to bed (100). Treatment consists of radical knee flexor release including the hamstrings, gastrocnemius, and posterior capsule. It is also important to correct any hip flexion contracture at the same time, if present.

The knee release is done using a transverse incision located approximately 1 cm above the posterior flexor crease extending from medial to lateral. In a patient with thoracic or high-lumbar involvement, all of the medial and lateral hamstrings tendons are divided and resected. Lengthening of the tendons can be done in patients with lower level of involvement to preserve some flexor power. After this, the origin of the gastrocnemius tendon is released from the medial and lateral femoral condyles allowing exposure of the posterior knee articular capsule. An extensive capsulectomy is then performed leaving the posterior cruciate ligament intact. After closure of the wound with nonabsorbable suture, a long leg cast is placed with the knee in extension taking care to pad the patella to prevent pressure. If full extension is achieved at the time of surgery, the cast is left in place for 3 weeks. If complete extension is not achieved, a cast change may be performed 1 week later in order to achieve further correction. After 3 weeks, a knee immobilizer is used at night-time to maintain correction.

In rare instances, a supracondylar extension osteotomy of the femur may be necessary to achieve full extension of the knee if radical knee flexor release is not successful. This is primarily used for older patients who maintain the ability for

community ambulation but are limited by a fixed knee flexion contracture.

In most cases, radical knee flexor release is successful in correcting the knee flexion deformity. Dias (99) reported a series of 23 knees undergoing radical flexor release. At final follow-up of 38 months, 21 of 23 knees maintained correction with flexion contracture of <10 degrees. In another study, a prospective review of 45 knees treated with radical flexor release found the mean knee flexion contracture decreased from 39 to 5 degrees after surgical release (100). The final average knee flexion contracture at follow-up of 13 years was 13 degrees (100). The authors noted a higher rate of recurrence of knee flexion contracture in patients with thoracic level of involvement compared to those with lumbar or lumbosacral. They also noted functional improvement in terms of walking ability in patients with L3/4 and L5/S1 levels of involvement.

Knee Extension Contracture. Knee extension contracture is much less common than a flexion deformity. In most cases, knee extension contracture occurs bilaterally and is present at birth (99). Knee extension is frequently associated with other congenital anomalies such as dislocation of the ipsilateral hip, external rotation contracture of the hip, and equinovarus deformity of the foot (99, 103) (Fig. 15-9). Other causes of fixed extension contractures are unopposed quadriceps function with weak hamstrings, extensive bracing in extension, malunion after supracondylar fracture of the femur, and iatrogenic after surgical treatment of flexion contracture (34, 101).

Initial treatment entails a serial casting program with the goal of achieving at least 90 degrees of knee flexion. In most young patients, casting followed by physical therapy is successful. Surgical treatment is indicated when persistent extension contracture interferes with gait, sitting, using a wheelchair, or performing transfers (101, 104). The preferred procedure is V–Y quadriceps lengthening with anterior capsulotomy as needed to obtain 90 degrees of flexion at the time of surgery (99, 101, 104). This is done using an anterior oblique incision beginning superomedially below the lesser trochanter and extending distally and laterally (99, 103). The extensor mechanism is divided superior to the patella with an inverted V incision. If needed, the anterior capsule is divided transversely to the medial and lateral collateral ligaments. The quadriceps is then sutured with the knee held in 45 degrees of flexion. The knee is then immobilized in a long leg cast with 45 degrees of knee flexion for 2 to 3 weeks. Physical therapy with active and passive motion starts after 2 to 3 weeks. Results with quadriceps plasty have been positive in terms of improving gait and sitting. Parsch and Manner (101) reported very good results after quadriceps plasty in 9 out of 10 patients. Dias (99) reported 13 of 15 patients treated with quadriceps plasty maintained at least 90 degrees of flexion at 43 months follow-up.

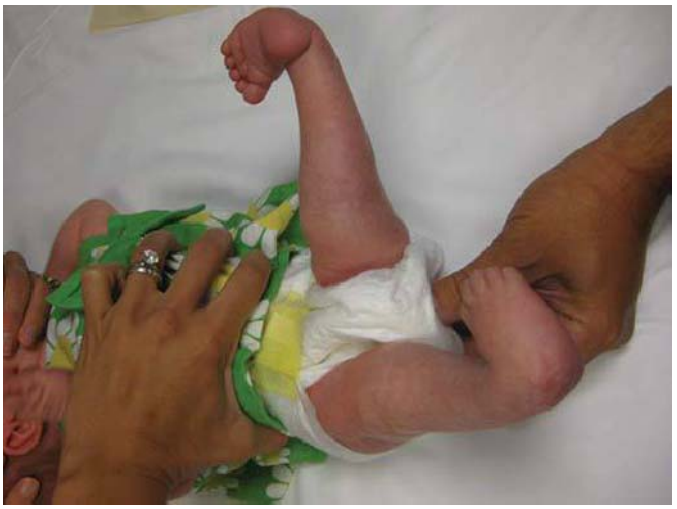

FIGURE 15-9. Newborn patient with myelomeningocele and knee extension contracture with ipsilateral hip dislocation and equinovarus deformity of the foot.

In nonambulatory patients without normal quadriceps function, another treatment option for knee extension contracture is tenotomy of the patellar tendon (104, 105). Sandhu et al. (105) reported a successful result in five out of eight patients with no further surgery required at 4 years follow-up. The authors achieved 50 to 70 degrees of knee flexion with tenotomy of the patellar tendon and 90 degrees or more of flexion with division of the medial and lateral retinacula as well. However, the authors stress that patellar tenotomy is recommended only for patients without normal quadriceps function and would otherwise recommend a formal quadricepsplasty.

Knee Valgus Deformity and Late Instability. Valgus deformity of the knee, seen especially in patients with low-lumbar and sacral level of involvement, leads to instability, pain, and arthritis in adulthood. A specific gait pattern has been identified in symptomatic patients who have weakness of the hip abductors and gastrocsoleus muscles. The characteristic gait, described by Williams et al. (106), is an abductor lurch with the knee deforming into valgus and flexion during stance, followed by a swivel push-off on a fixed pronated foot. This gait pattern leads to increased stress on the knee ligaments and articular surfaces. Williams et al. (106) reported a series of 72 community ambulators over the age of 23 years and found 17 (24%) had significant knee symptoms.

The use of gait analysis has contributed to the understanding of abnormal valgus stress at the knee by allowing the identification of multiple factors leading to this stress. This includes rotational malalignment of the femur, femoral anteversion in association with excessive external tibial torsion, excessive trunk and pelvic movement, and knee flexion contracture (46, 55, 107, 108). Surgical treatment of excessive rotational deformities can decrease valgus stress at the knee and is indicated in patients over 6 years of age (55). Correction of rotational deformities leads to a significant improvement in knee stress and pain and may prevent the onset of late degenerative

changes (49, 108). In addition, if knee valgus is associated with knee flexion contracture or hindfoot valgus, these deformities must be addressed at the same surgical setting (95). Patients found to have valgus stress at the knee should be encouraged to use an AFO and forearm crutches to decrease pelvic obliquity and rotation and hence increase stance-phase stability and decrease stress at the knee joint (46, 107).

ROTATIONAL DEFORMITY

Rotational deformities of the lower extremities develop commonly in both ambulatory and nonambulatory patients with myelomeningocele. The femur may be involved with an external rotation deformity of the hip that occurs due to contracture of the posterior hip capsule and short external rotator muscles. In addition, with the abnormal gait and activity levels in children with myelomeningocele, the normal newborn femoral torsion does not reliably decrease with growth (34). Even more common in patients with myelomeningocele are torsional deformities involving the tibia. Internal tibial torsion is a congenital deformity and is frequently associated with clubfoot. External tibial torsion, often associated with a shortened fibula and valgus deformity of the ankle, is an acquired deformity resulting from muscle imbalance. In certain ambulatory patients, the persistent proximal swivel motion of the pelvis and hip over the planted stance foot induces external tibial torsion (17).

In nonambulatory patients, rotational deformities are mainly a cosmetic problem. Treatment is indicated in ambulatory patients whose gait is impacted by the deformity, such as with internal tibial torsion, which can cause significant intoeing causing patients to trip and fall. Initially, treatment should be conservative utilizing twister cables attached to an AFO brace. In patients older than 5 to 6 years of age with severe femoral or tibial rotational deformity, surgical treatment indications include labored gait, difficulty with orthotic fitting resulting in skin ulceration, and pain (49). A detailed assessment of the patient's gait pattern utilizing three-dimensional gait analysis when available should be done to determine the extent of deformity correction necessary. The goal of treatment is to minimize bracing requirements while achieving as normal a gait pattern as possible (109).

Femoral Torsion. In ambulatory patients with myelomeningocele, both excessive hip external and internal rotation can occur and impact gait. An internal rotation deformity of the hip can cause severe valgus stress at the knee when associated with external tibial torsion. External rotation at the hip joint can contribute to severe out-toeing when associated with external tibial torsion. Careful physical examination, including three-dimensional gait analysis if available, is necessary to ensure all the components of rotational deformity affecting gait are identified. As mentioned above, the initial treatment is with a twister cable attached to an AFO brace. If the deformity and resulting gait problem persist past the age of 5 to 6 years, femoral osteotomy is indicated. In this case, the osteotomy is performed at the subtrochanteric level, and the distal segment is rotated to bring the foot into a position of neutral rotation (109). An AO dynamic compression plate, either 5 hole or 6 hole, is used for fixation. Postoperatively either a TBS or abduction wedge is used for 4 to 6 weeks until sufficient healing is present to allow mobilization and weight bearing.

Internal Tibial Torsion. Internal tibial torsion requires surgical treatment when the resulting intoeing causes significant gait disturbance with frequent tripping. At the time of surgery, it is important to recognize any associated muscle imbalance. For instance, a spastic anterior tibialis muscle may require tenotomy with tendon excision at the same time as correction of the rotational deformity.

External Tibial Torsion. Excessive external tibial torsion can also affect gait, cosmesis, and cause difficulty with orthotic fit. External rotation of the tibia places the medial malleolus more anteriorly leading to rubbing against the AFO and may cause a pressure sore on the medial aspect of the ankle (109). Improving external tibial torsion will not only alleviate skin issues, but also improve the effectiveness of the AFO brace in achieving knee extension. Even in the absence of a fixed knee flexion contracture, external tibial torsion >20 degrees can lead to a crouch gait pattern because the AFO is unable to improve the extension of the knee during stance phase (110). Hence, internal rotation osteotomy should be considered when the amount of external torsion exceeds 20 degrees in order to improve knee extension during stance phase (110). When planning for surgical correction, the patient's entire lower extremity should be carefully examined with particular attention to the hindfoot as hindfoot valgus may occur in association with external tibial torsion. In this case, both deformities require treatment in order to achieve a successful result (95).

When surgical treatment is indicated for either internal or external tibial torsion, the procedure of choice is a distal tibia and fibula derotation osteotomy (111). However, in patients with myelomeningocele, rotational osteotomies of the tibia are known to have a high rate of complications, such as delayed union and wound infection (112), so careful attention must be paid to the technical details of the procedure. The osteotomy should be performed just above the distal tibial physis, and the distal fibula osteotomy should be performed through a separate incision. The osteotomy should be created using multiple drill holes and a corticotomy in an attempt to decrease the thermal insult to the bone and preserve healing potential. An AO dynamic compression plate, usually a 5-hole plate, is used to provide stable fixation. The wound is then closed over a drain with interrupted, nonabsorbable sutures, and a short leg cast is placed. For the first 3 weeks, no weight bearing is allowed. After that time, a cast change is performed and the sutures are removed. The patient is allowed to weight bear in a walking cast for an additional 3 weeks or until sufficient healing is present. Utilizing this approach in a series of 10 osteotomies, there were no incidences of nonunion (49).

Using lower extremity osteotomies to treat rotational deformities has resulted in successful outcomes in terms of gait parameters and range of motion in 80% to 90% patients (109, 112). With regard to excessive external tibial torsion, derotation osteotomy will improve knee extension during stance phase. Corrective osteotomy may also delay or prevent the onset of late degenerative changes about the knee (49). Dunteman et al. used three-dimensional gait analysis to examine eight patients with external tibial torsion. They found increased valgus knee stress in 100% of the patients. After derotational tibial osteotomy, a significant improvement in the abnormal knee moment was seen along with improvement of knee extension during stance phase (49). In order to avoid the increased risk of complications such as delayed union and wound infection in patients with myelomeningocele, meticulous attention to technical details is important.

FOOT/ANKLE DEFORMITY

Foot deformity is present in almost all patients with myelomeningocele (34, 113). The spectrum of foot deformities seen includes calcaneus, equinus, varus, valgus, clubfoot, and vertical talus. Foot deformities can preclude effective bracing to allow ambulation, cause difficulty with shoe wear, create cosmetic problems, or lead to pressure sores. The common goal of treatment is a plantigrade, braceable foot with maximally preserved range of motion. Serial manual muscle testing is important for the detection of subtle muscle imbalance, which can lead to more significant deformities. Early intervention with casting, bracing, or surgical treatment may prevent fixed bony deformities. Surgical principles include the use of tendon excisions that are more reliable than tendon transfer or lengthenings. For bony deformities, osteotomies provide correction while preserving joint motion. Surgical arthrodesis should be strictly avoided because the stiffness that results combined with an insensate foot has been shown to result in the development of neuropathic skin changes (30, 114). After surgical treatment, an AFO brace should be used to maintain correction and prevent recurrence.

Clubfoot. Clubfoot is the most common foot deformity in patients with myelomeningocele and has been reported in 30% to 50% of patients (31, 34, 115). Incidence of clubfoot varies with neurologic level of involvement. It occurs in approximately 90% of patients with thoracic or lumbar levels of involvement and 50% of patients with sacral level involvement (31). The clubfoot deformity in patients with myelomeningocele is quite different from the idiopathic clubfoot. In myelomeningocele, the clubfoot is often severely rigid (Fig. 15-10), similar to that seen in patients with arthrogryposis. Many patients also have severe internal tibial torsion.

Traditional teaching has been that nonsurgical management is rarely successful, and extensive soft-tissue release surgery is necessary for correction. However, two recent studies have reported promising early results using the Ponseti method of serial manipulation and casting in clubfeet associated with myelomeningocele (116, 117). Gerlach et al. (116) reported that initial correction was achieved in 27 of 28 clubfeet. Relapses occurred in 68% of the clubfeet but were treated successfully without extensive soft-tissue release surgery in all but 4 ft. Similarly, Janicki et al. reported initial correction with the Ponseti method in 9 out of 9 clubfeet. Five feet had recurrences and three of these required extensive soft-tissue release. They did note skin breakdown in 2 of the clubfeet. The Ponseti method can be useful in decreasing the need for extensive soft-tissue surgery, but families should be counseled about the high risk of recurrence, potential for need for further treatment, and risk of skin breakdown and fractures.

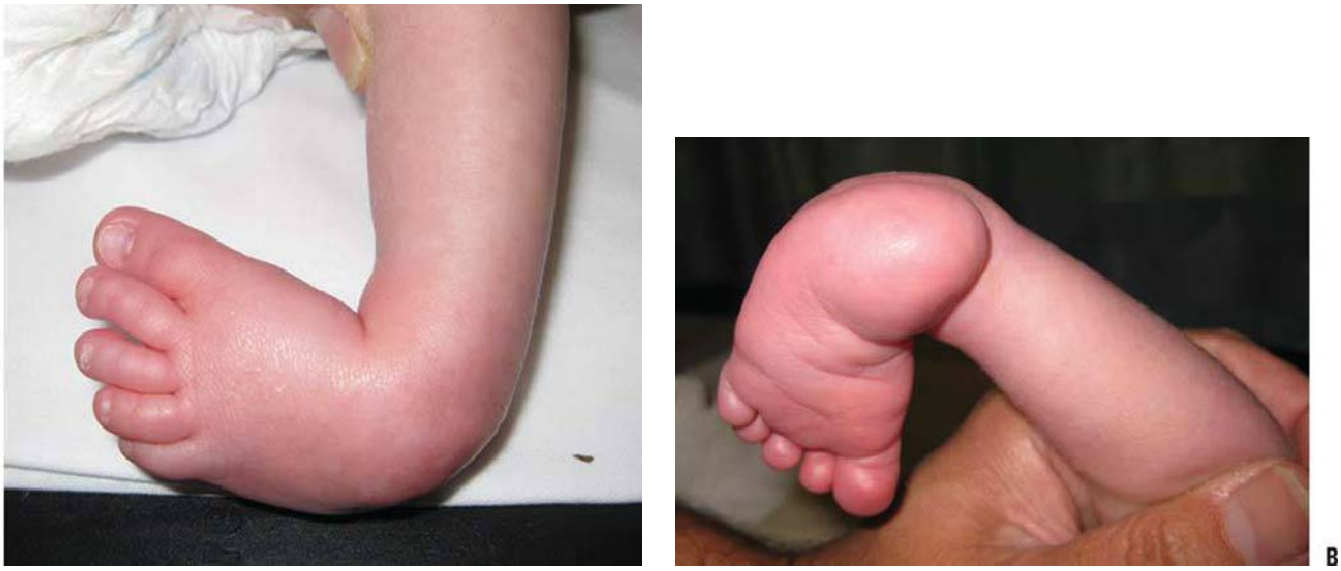

FIGURE 15-10. Rigid clubfoot in infant with myelomeningocele, anterior **(A)** and posterior **(B)** views.

When soft-tissue release surgery is indicated, the optimum time for treatment is at approximately 10 to 12 months of age. The surgical treatment consists of a radical posteromedial–lateral release using a Cincinnati incision (see Chapter 29). All tendons are excised rather than lengthened, including the anterior tibialis tendon. The subtalar, calcaneocuboid, and talonavicular joints are completely released. A separate plantar release may be needed through a plantar incision. Improved results have been shown with the use of a temporary Kirschner wire (K-wire) to derotate the talus in the ankle mortise (Fig. 15-11)

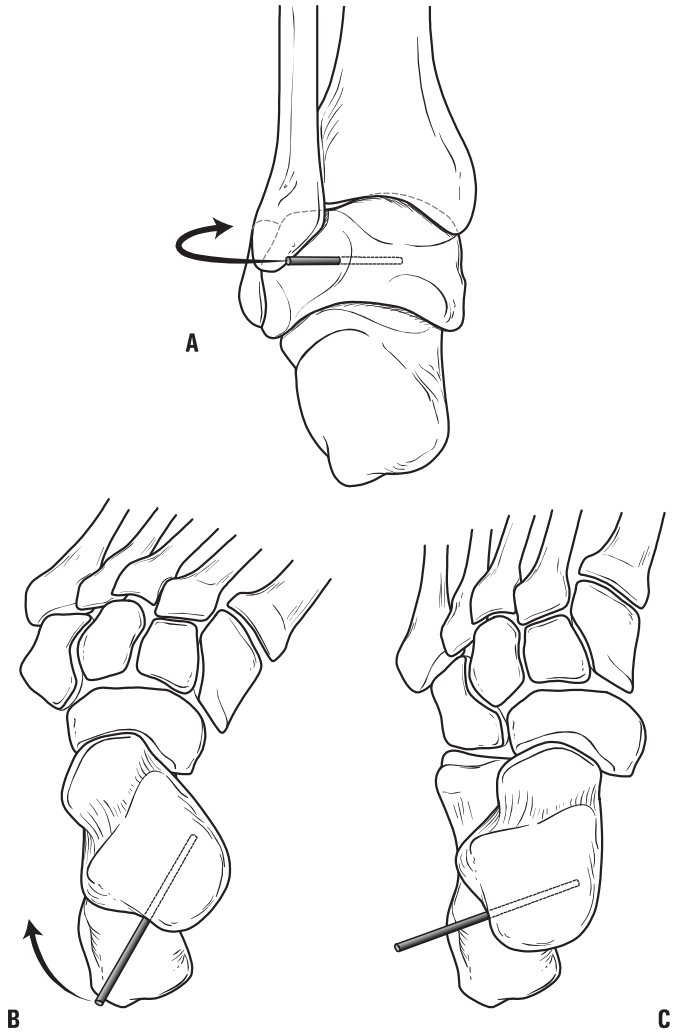

FIGURE 15-11. Temporary K-wire inserted into the posterolateral aspect of the talus to derotate the talus medially in the ankle mortise. **A:** Posterior view of the ankle and talus. The K-wire is inserted in the posterolateral surface of the talus. Note the external rotation of the talus in the ankle mortise. **B:** The abnormal rotation of the talus is seen. The K-wire is used to derotate the talus to its normal position. **C:** With the talus in a normal alignment and the talonavicular joint reduced, a second K-wire is then used to maintain this correction. (Reprinted from de Carvalho Neto J, Dias LS, Gabrieli AP. Congenital talipes equinovarus in spina bifida: treatment and results. *J Pediatr Orthop* 1996;16:782–5, with permission.)

(115). The K-wire is placed into the posterolateral aspect of the talus to rotate the talus medially, and the navicular is reduced on the talar head. A second K-wire is driven through the body of the talus into the navicular to hold the reduction and the temporary K-wire is then removed. Another K-wire is used to maintain the proper alignment of the talocalcaneal joint. Postoperatively, a long leg posterior mold splint is used with the foot in slight equinus to decrease tension on the interrupted sutures used for skin closure. After 2 weeks, the patient is changed to a long leg cast with the foot held in the corrected position. This remains in place for 6 weeks. After casting day and nighttime AFOs are used to maintain correction.

Good results after surgical release have been reported in 61% to 83% of patients (31, 115, 118). Outcome varies with motor level of involvement. de Carvalho Neto et al. (115) reported 50% poor results in patients with thoracic and high-lumbar level of involvement compared to only 11% poor results in patients with low-lumbar and sacral levels of involvement. The recurrence rate after surgical treatment is higher than in patients with idiopathic clubfoot and may be due in part to the lack of normal muscles around the ankle joint and lack of weight bearing (115). For this reason, it is important that at the time of cast removal, a standing A-frame is prescribed as well as the AFO.

Partial or complete recurrence occurs in 20% to 50% of patients after primary surgical correction (31). Patients with partial recurrence often develop adduction deformity, which may result from growth imbalance between an elongated lateral column and a shortened medial column. If bracing is not successful, surgical correction consists of a combination of lateral column shortening and medial column lengthening (see Chapter 29). This is done with the "double osteotomy," which consists of a closing wedge osteotomy of the cuboid with an opening wedge osteotomy of the medial cuneiform (Fig. 15-12) (119). Good results have been shown using this technique in children older than 4 years of age (119).

When complete recurrence occurs, the best procedure for achieving a plantigrade foot is talectomy (Fig. 15-13) (92, 120). Using an Ollier incision, an attempt is made to remove the talus as one piece. The tibiotalar, subtalar, and talonavicular joints are identified and opened widely. If contracture and scar make dissection difficult, needles can be used with intraoperative imaging to confirm location of the joints. To avoid recurrence, it is important not to leave any fragments of the talus remaining. Once the talus is removed, the calcaneus is thrust posteriorly in the ankle mortise and held in position with a K-wire. A short leg cast is then applied for at least 6 weeks. Dias and Stern (120) reported good results in 82% of feet treated with talectomy. The authors noted that severe forefoot deformities are not corrected by the talectomy; hence, any residual adduction deformity must be treated separately with concomitant closing wedge osteotomy of the cuboid.

Equinus. Equinus deformity occurs more commonly in patients with thoracic and high-lumbar levels of involvement but has been reported in patients with all levels of involvement

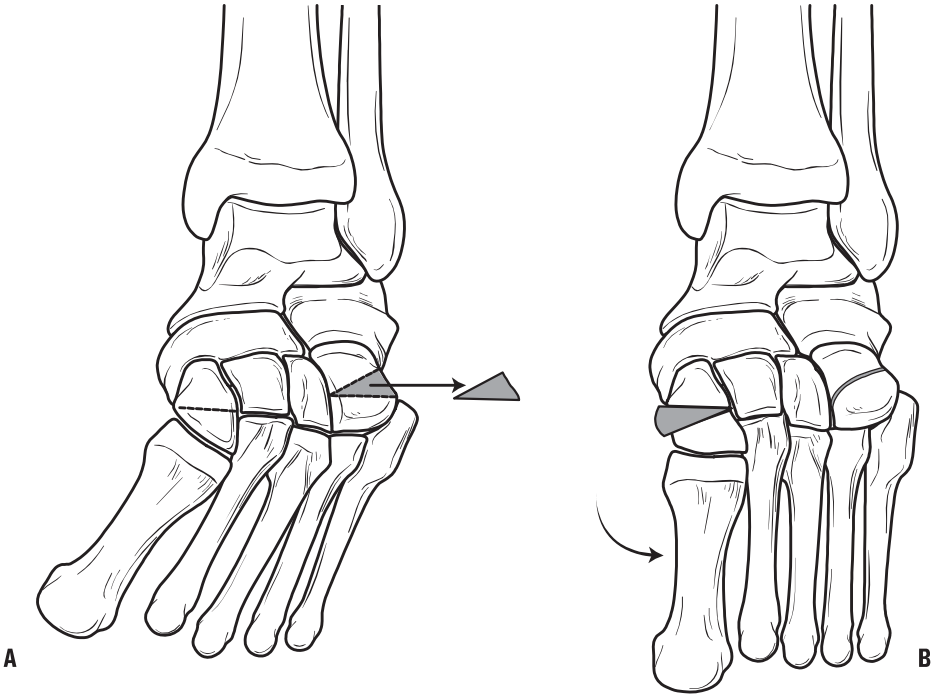

FIGURE 15-12. "Double osteotomy" to correct forefoot adduction. **A:** closing wedge osteotomy of the cuboid. **B:** opening wedge osteotomy of the medial cuneiform. (Reprinted from Lourenco AF, Dias LS, Zoellick DM, et al. Treatment of residual adduction deformity in clubfoot: the double osteotomy. *J Pediatr Orthop* 2001;21:713–8, with permission.)

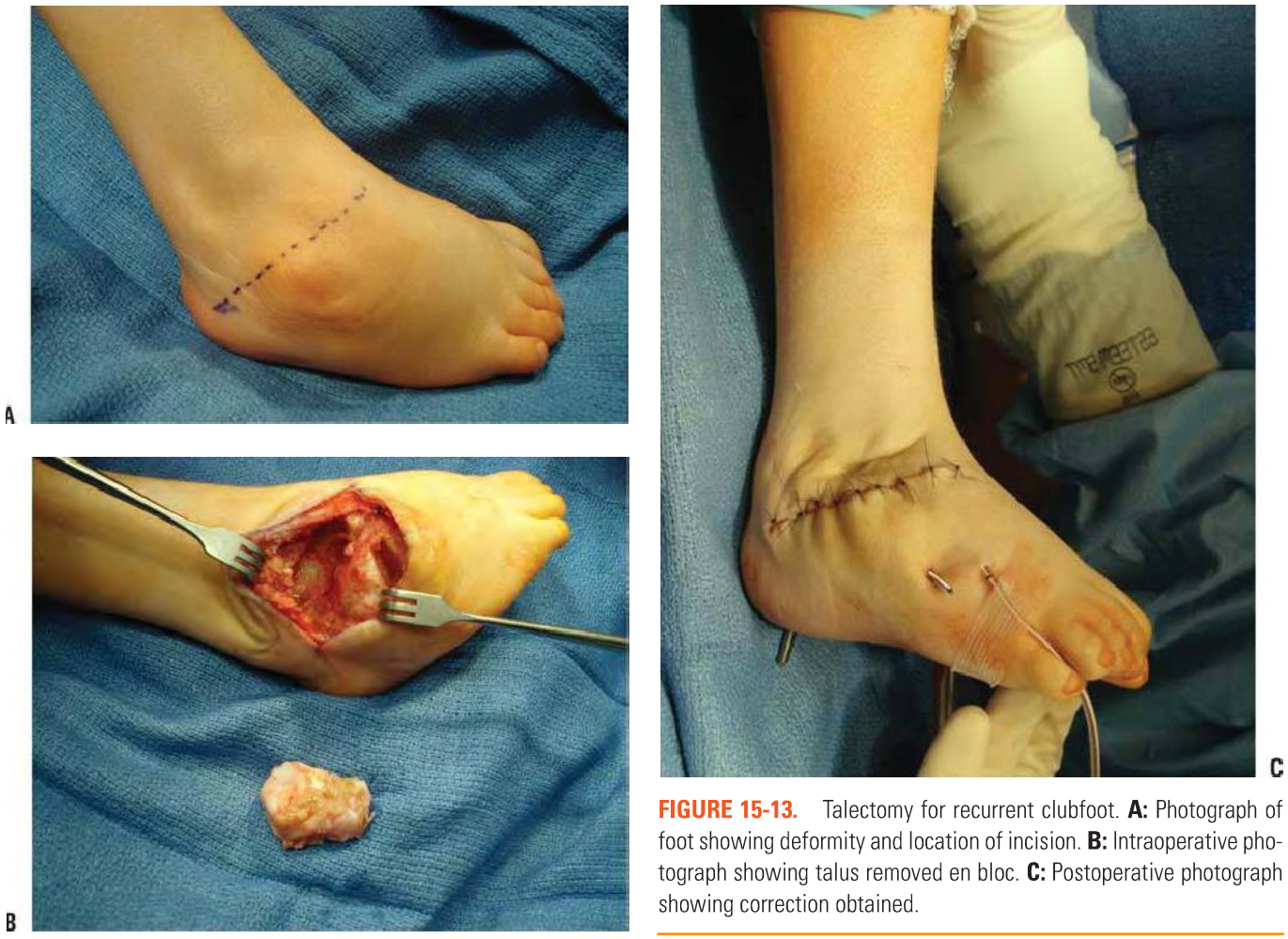

FIGURE 15-13. Talectomy for recurrent clubfoot. **A:** Photograph of foot showing deformity and location of incision. **B:** Intraoperative photograph showing talus removed en bloc. **C:** Postoperative photograph showing correction obtained.

(121). An AFO may be used to attempt to prevent equinus. Surgical treatment is indicated to achieve a plantigrade, braceable foot. The type of surgical procedure selected depends on the severity of deformity. Mild deformities respond to simple Achilles tendon excision. More severe contractures require a radical posterior release including the posterior tibiotalar and talocalcaneal joints. The authors prefer to use a limited Cincinnati incision and excise all tendons. The calcaneofibular ligament must be divided to achieve full correction. A K-wire may be used in the talocalcaneal joint to maintain neutral hindfoot alignment. A short leg cast is used for at least 6 weeks postoperatively followed by an AFO during the day and night.

Vertical Talus. Vertical talus deformity occurs in approximately 10% of patients with myelomeningocele (34) and is characterized by a rigid rocker-bottom flatfoot deformity with malalignment of the hindfoot and midfoot. The talus is nearly vertical and the calcaneus is in equinus and valgus. The navicular is dislocated dorsally and laterally on the talus. Vertical talus occurs in two forms in patients with myelomeningocele, either congenital that is more common, or developmental. The goal of treatment is to restore the normal relationship between the talus, navicular, and calcaneus and provide a plantigrade weight-bearing surface (122). Traditional treatment has been with complete posteromedial–lateral and dorsal release when the patient is between 10 and 12 months of age. However, a new technique of serial manipulation and cast immobilization followed by open talonavicular pin fixation and percutaneous tenotomy of the Achilles tendon has been reported in idiopathic congenital vertical talus with excellent short-term results (123). The authors have begun using this method for initial correction of vertical talus in newborns with myelomeningocele with good initial success (Fig. 15-14).

When extensive soft-tissue release is necessary, good results have been reported with single-stage surgical correction addressing both the hindfoot and the forefoot (122). Using a Cincinnati incision, the Achilles tendon is z-lengthened, and the posterior capsules of the tibiotalar and subtalar joints are opened. The posterior and anterior tibial tendons are detached from their insertions and tagged for later repair. After this, the medial and dorsal aspects of the talonavicular joint, and the medial and lateral aspects of the subtalar joint are released. If necessary, the calcaneocuboid joint is released as well. Next a small K-wire is placed into the posterolateral aspect of the talus and used as a joystick to elevate the talus into a reduced position while plantarflexing the navicular and the forefoot (Fig. 15-15). Both the talonavicular and subtalar joints are then pinned in a reduced position, and if needed the extensor and peroneal tendons can be lengthened.

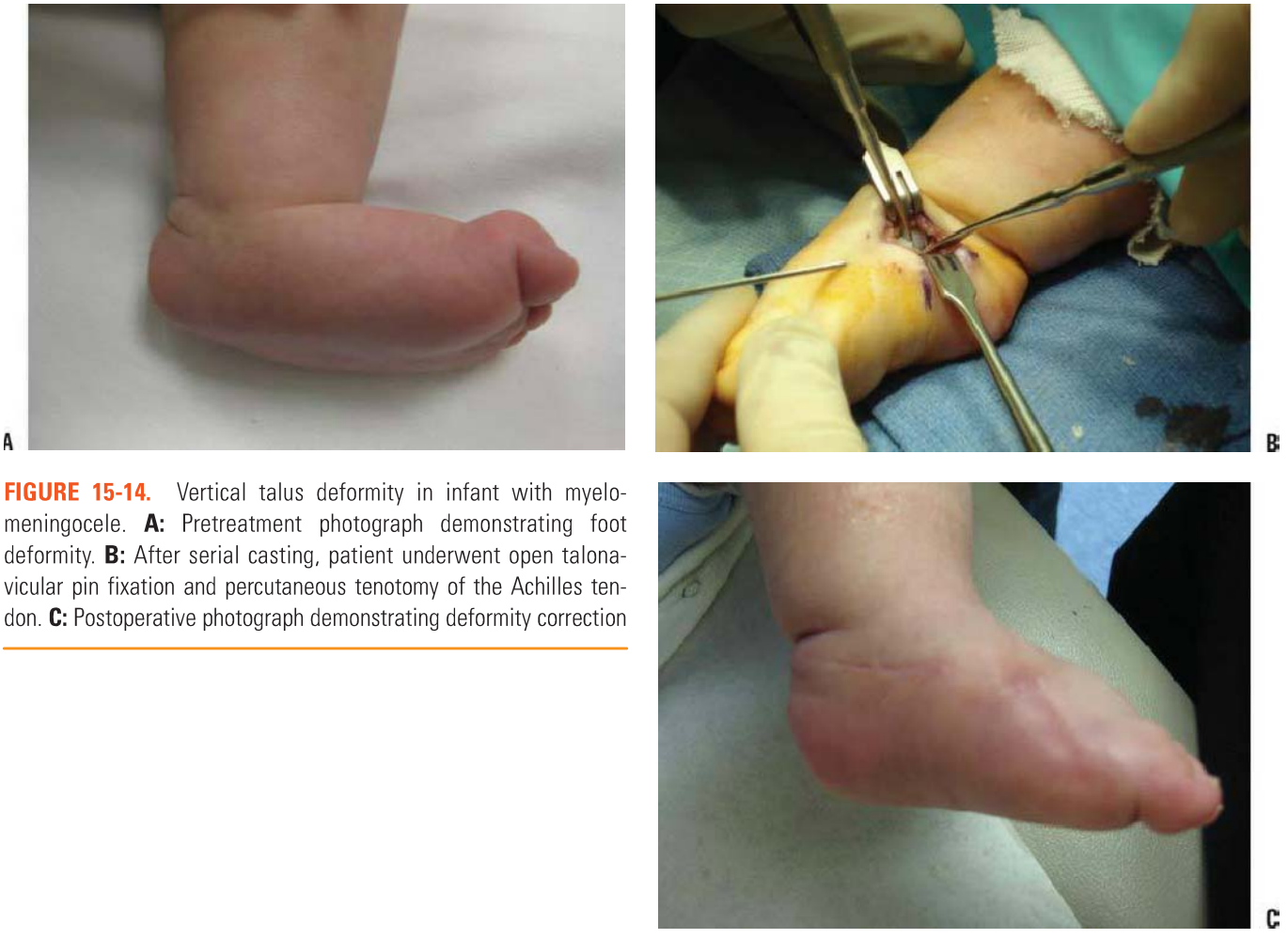

FIGURE 15-14. Vertical talus deformity in infant with myelomeningocele. **A:** Pretreatment photograph demonstrating foot deformity. **B:** After serial casting, patient underwent open talonavicular pin fixation and percutaneous tenotomy of the Achilles tendon. **C:** Postoperative photograph demonstrating deformity correction

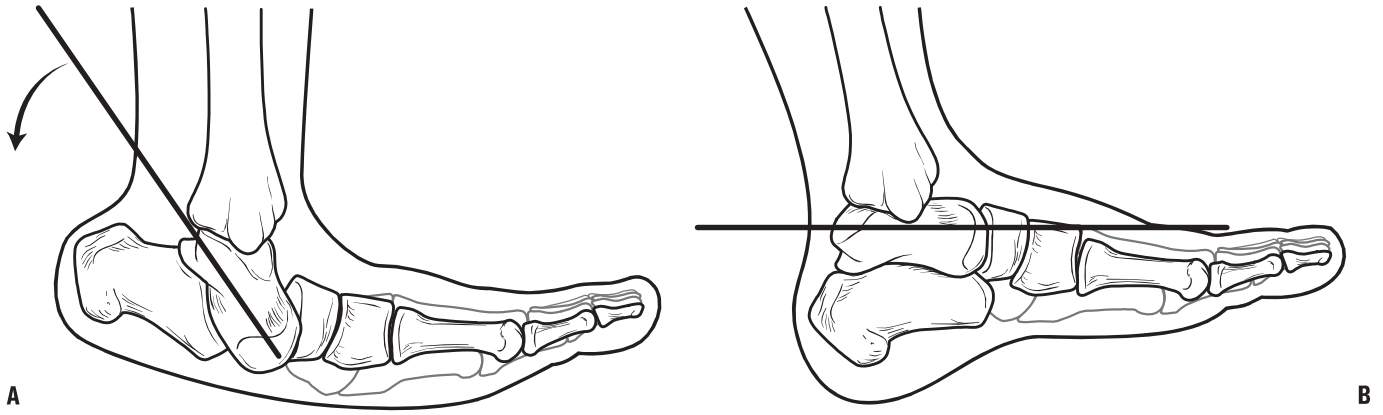

FIGURE 15-15. **A,B:** K-wire placed into posterolateral aspect of talus and used as joystick to elevate talus into reduced position while plantarflexing the navicular and forefoot. (Reprinted from Kodros SA, Dias LS. Single-stage surgical correction of congenital vertical talus. *J Pediatr Orthop* 1999;19:42–8, with permission.)

Calcaneus and Calcaneovalgus. Calcaneus deformity occurs in approximately 30% of patients with myelomeningocele. It is most common in patients with L4 or L5 level of involvement due to strength or spasticity of the ankle dorsiflexors combined with weakness of plantar flexion (31, 34, 124). Calcaneovalgus results from imbalance between the ankle evertors and the invertors. If the deformity is not rigid, an AFO may be useful to maintain the foot in neutral position. When the deformity is rigid, it can be very difficult to treat conservatively or surgically.

If left untreated, calcaneus deformity causes loss of normal toe-off and a crouch gait (31, 124). Persistent weight bearing on a calcaneus deformity leads to a bulbous heel prone to pressure sores and secondary osteomyelitis (31). External tibia torsion frequently develops in association with calcaneovalgus but can be avoided by early correction of the muscle imbalance (17). Surgical treatment with anterolateral release including tenotomy of all ankle dorsiflexors and the peroneus brevis and longus can achieve a plantigrade, braceable foot. Rodrigues and Dias (125) reported a series of 76 patients treated with anterolateral release and achieved a good result in 82%. The poor results were due to either recurrence requiring a second release or equinus deformity requiring release of the Achilles tendon. The authors have found the anterolateral release to be a simpler procedure than the anterior tibial tendon transfer to the os calcis with similar results. However, Park et al. (124) recently reported a series of 31 calcaneus feet treated with anterior tibialis tendon transfer with concomitant osseous surgeries in 12 ft. They noted no recurrence or worsening of the deformity in any patient and no other type of foot deformity developed after the surgery.

In older patients who have developed significant bony deformity, surgical correction requires not only release of all the extensor tendons and peroneals if needed, but also bony correction. A closing wedge osteotomy of the calcaneus with a plantar release can improve hindfoot alignment. If calcaneal valgus is present, a lateral opening wedge osteotomy of the cuboid may be necessary to achieve complete correction.

Ankle and Hindfoot Valgus. Valgus deformities of the hindfoot and ankle are common in ambulatory patients with myelomeningocele. Successful treatment depends on identifying the precise anatomical location of the deformity that can arise from the distal tibia, hindfoot, or both. Valgus deformities tend to become more pronounced as a child matures, begins ambulation, and gains weight (114). Valgus deformity (Fig. 15-16) is common in patients with low lumbar levels of involvement due to muscle imbalance, weight bearing, and the effects of gravity. When flexible, these deformities are initially managed with a rigid AFO to provide stability. Often as the hindfoot progresses into more valgus, skin irritation and breakdown over the medial malleolus and talar head result from excessive pressure against the brace. Surgery is indicated for severe, rigid deformities causing pain, difficulty with brace wear, or ulceration (114). Treatment options include distal tibia osteotomy, hemiepiphyseodesis of the distal tibia, or medial displacement osteotomy of the calcaneus.

For ankle valgus due to deformity in the distal tibia (Fig. 15-17), surgical treatment depends on the severity of the deformity and the amount of growth remaining. Hemiepiphysiodesis is indicated for mild deformities with sufficient growth remaining. Temporary growth arrest of the medial physis with continued growth of the lateral physis allows gradual correction of the valgus tilt. Use of a single cannulated screw has been reported in a series of 50 ft with satisfactory improvement of ankle valgus, low morbidity, and no incidence of permanent physeal closure (126). To avoid permanent closure of the physis, the screw should be removed within 2 years of its insertion. For more severe ankle valgus or in an older child with little growth remaining, a distal tibia osteotomy is indicated. Osteotomies of the distal tibia are associated with a high incidence of complications such as delayed union, nonunion, wound infection, and loss of correction. However, the authors have had good success with the transphyseal osteotomy described by Lubicky and Altiok (127). Care should be taken to create the osteotomy with multiple drill holes connected by an osteotome rather than with power instruments. If concomitant

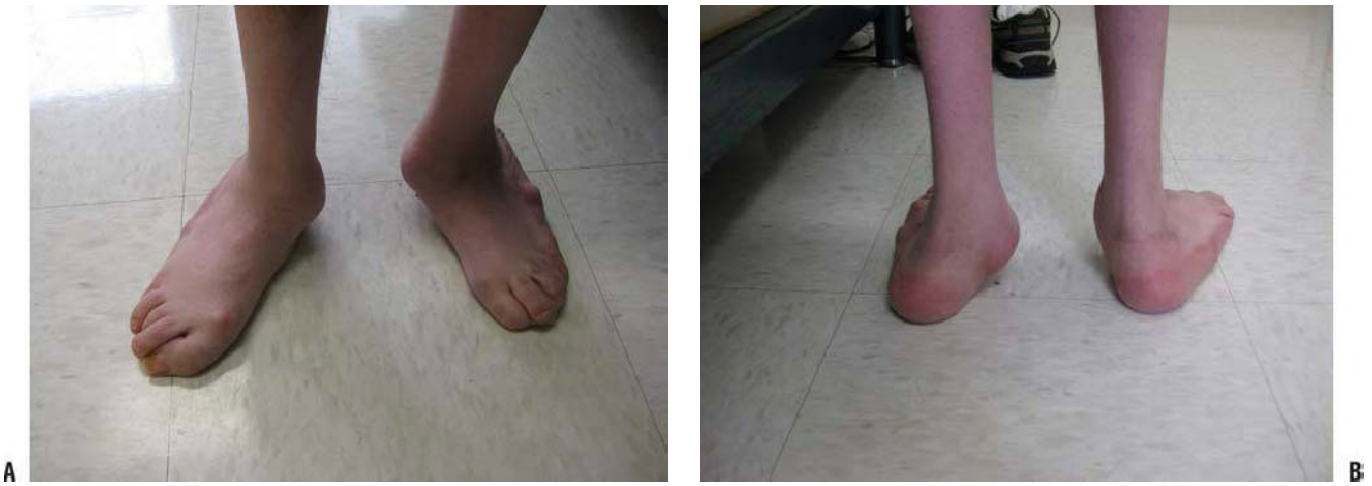

FIGURE 15-16. Valgus deformity in patient with myelomeningocele. **A:** Anterior view demonstrating bursa formation due to irritation over medial malleolus from brace. **B:** Posterior view demonstrating hindfoot valgus.

external tibial torsion is present as is often the case, internal rotation of the distal fragment should be done at the same time.

Surgical treatment for valgus deformity of the hindfoot consists of medial sliding osteotomy of the calcaneus in an effort to preserve subtalar motion while correcting the deformity. This procedure was initially described as a treatment for idiopathic flatfoot by Koutsogiannis (128) but has been also been reported in a series of patients with myelomeningocele (114). Using a lateral L-shaped incision to provide adequate exposure, full-thickness flaps are elevated to allow extraperiosteal dissection of the calcaneus. An oblique osteotomy is made, and the amount of displacement of the distal fragment required for correction is usually 50% of the width of the fragment (114). A threaded K-wire should be used for internal fixation that is left in place for 3 weeks. After 3 weeks, the K-wire is removed and the patient is allowed to begin weight bearing in a short-leg walking cast. Using this procedure in 38 ft in patients with myelomeningocele, good results were obtained in 82% (114). In this series, three of the poor results were due to unrecognized concomitant distal tibia valgus deformity.

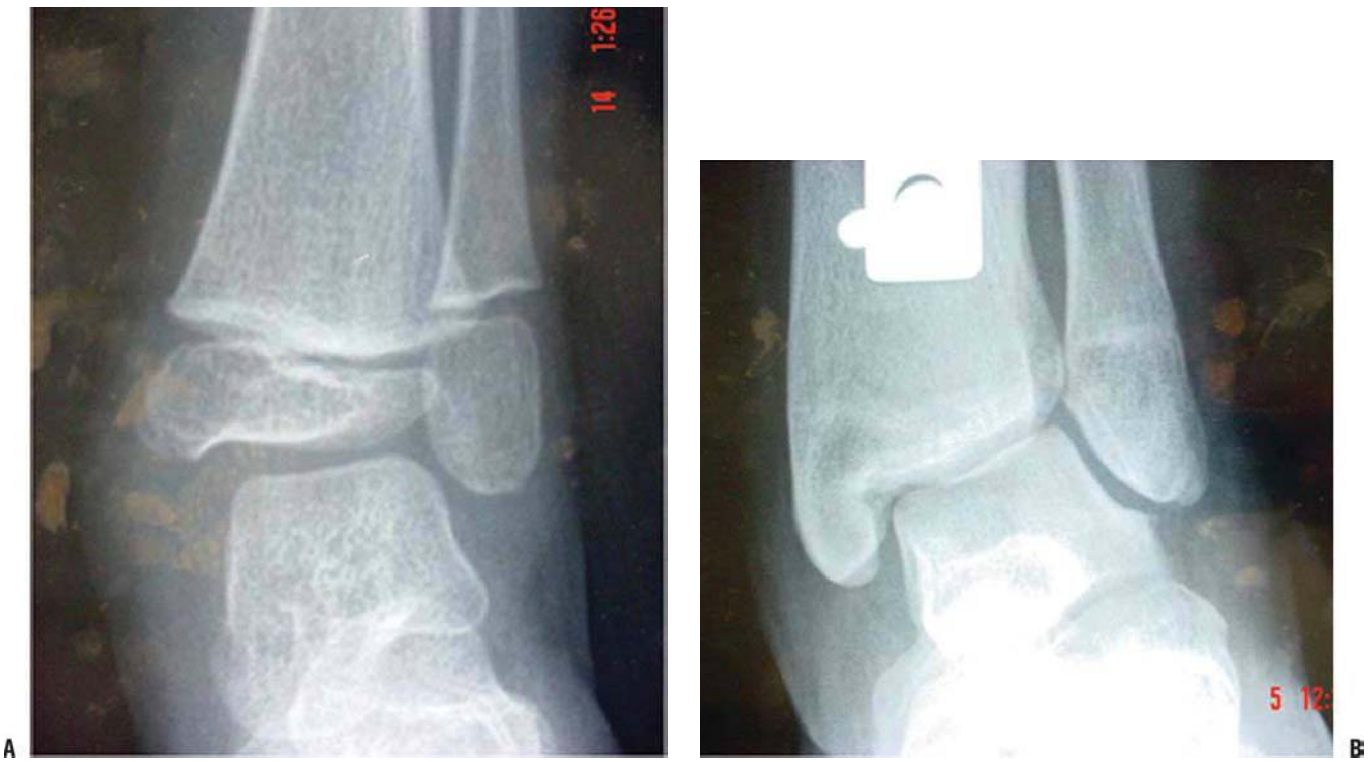

FIGURE 15-17. Distal tibia valgus deformity. **A:** Mild valgus in a skeletally immature patient. **B:** Severe valgus in a skeletally mature patient.

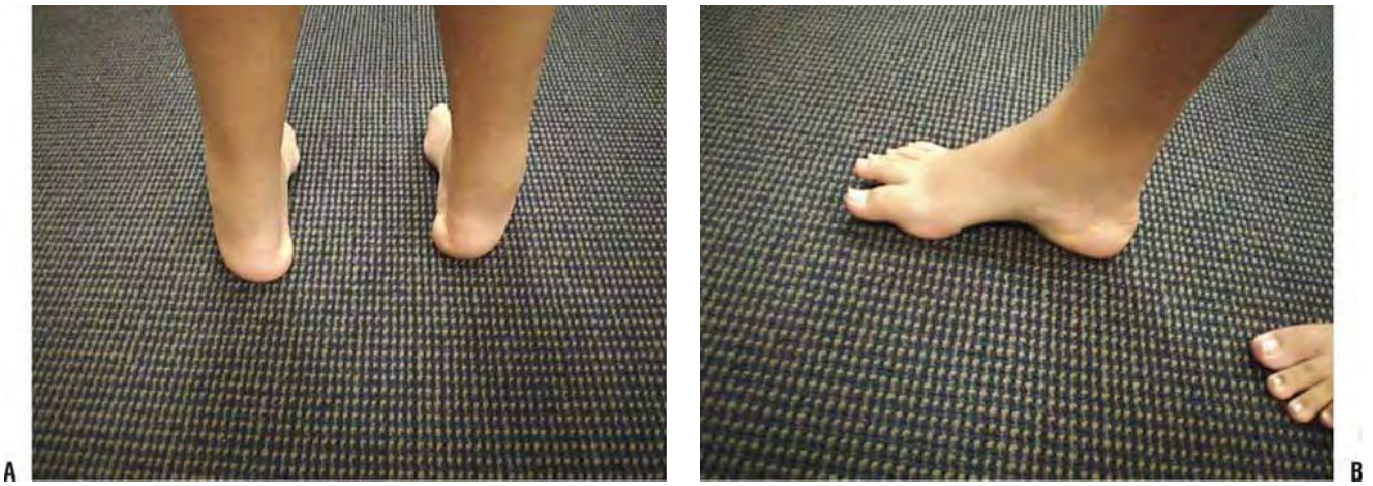

FIGURE 15-18. Cavovarus deformity in patient with myelomeningocele. **A:** Posterior view demonstrating hindfoot varus. **B:** Side view demonstrating cavus.

Cavus, Varus, and Cavovarus. Cavovarus deformity (Fig. 15-18) occurs in patients with sacral level myelomeningocele and in patients with lipomeningocele. The primary deformity is cavus and varus is caused by the muscle imbalance between the posterior tibialis and the peroneal muscles as well as intrinsic muscle weakness. Treatment is based on the flexibility of the deformity. The Coleman block test can be used to determine whether the hindfoot deformity is flexible or fixed (129). When the hindfoot varus is flexible, treatment is limited to the forefoot and consists of a radical plantar release. If the hindfoot varus is rigid, correction involves both the forefoot and the hindfoot (130). Muscle imbalance must be corrected at the same time. Mubarak and Van Valin (131) have described the use of selective, joint-sparing osteotomies to address deformity correction. They recommend a closing wedge osteotomy of the first metatarsal, opening plantar wedge osteotomy of the medial cuneiform, closing wedge osteotomy of the cuboid and if necessary a sliding osteotomy of the calcaneus and osteotomies of the second and third metatarsals. They also performed plantar release and peroneus longus-to-brevis tendon transfer when needed. In a series of 20 ft in patients with varying underlying etiologies, 95% had good or very good outcomes with this protocol (131). Triple arthrodesis should be avoided in this patient population with impaired sensation (130).

POSTOPERATIVE CARE

Patients with myelomeningocele are at a higher risk for certain postoperative complications compared to the general population. Care must be taken to prevent these complications, including skin breakdown, nonunion, and fractures. With regard to choice of immobilization, whenever possible a total body spica cast should be strictly avoided. A removable custom-molded TBS is a useful alternative to spica casting (Figs. 15-7 and 15-19). It provides adequate immobilization even for patients who have undergone bony surgical procedures while allowing for easier care and comfort of the patient. In addition, the TBS can be removed for gentle range of motion once adequate healing is present to avoid stiffness and contracture. Another benefit of the TBS is that it can be used at nighttime after the initial postoperative immobilization period to provide additional stretch in order to augment the effects of surgery and prevent recurrence of deformity. While immobilized after surgery, it is important to educate the patient's family and caregivers to avoid pressure on the posterior aspect of the patient's heels. We instruct them to use a small towel rolled up under the distal calf to keep the heel floating freely in order to avoid creating a pressure sore.

The use of rigid internal fixation with plate and screw fixation of osteotomy sites instead of K-wire fixation has many advantages. Rigid fixation allows a shorter period of immobilization

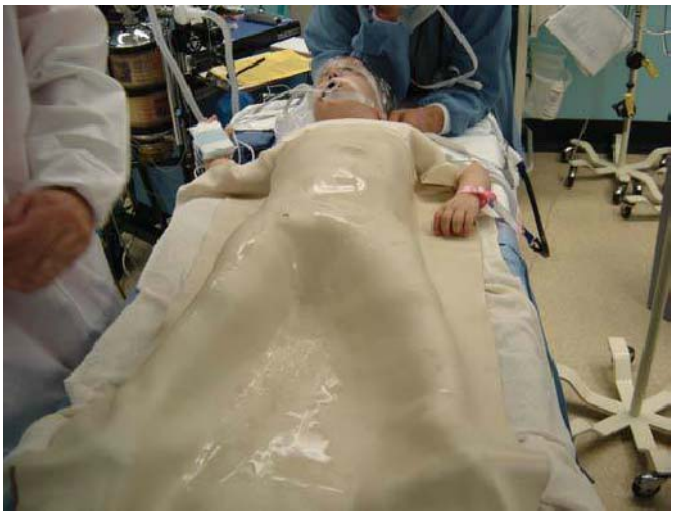

FIGURE 15-19. Intraoperative molding for custom TBS.

with earlier range of motion and weight bearing. In addition, rigid fixation helps to decrease the risk of nonunion.

The surgeon must properly educate the patient and family on how to avoid certain postoperative complications. Especially important is to strictly forbid crawling for at least 3 to 4 weeks after immobilization is discontinued. Crawling places a large amount of stress at the supracondylar region of the femur, which is a common location for postimmobilization fracture. The family is more likely to adhere to postoperative instructions if educated on the reason behind the recommendation.

Postoperative therapy should begin early—as soon as surgical wounds are stable and adequate healing is present. Goals of physical therapy should be tailored to the individual patient but often include preventing contractures with active and passive range of motion, strengthening program, early weight bearing, and gait training.

ORTHOSES

Almost all children with myelomeningocele will require orthotic support to achieve ambulation. The exception to this is some patients with low-sacral level of involvement. With regard to ambulation, the goal of orthotic treatment is to facilitate independent mobility while minimizing restrictions. The type of brace required depends on the motor deficit present and trunk balance. There are many other indications for the use of orthoses in patients with myelomeningocele aside from ambulation. These include maintenance of proper alignment and prevention of deformity, correction of flexible deformity, and protection of the insensate limb (95).

Nighttime bracing may be indicated to prevent orthopaedic deformities. As an example, a patient with thoracic level involvement may benefit from a nighttime TBS to prevent hip flexion and external rotation contractures, knee flexion contractures, and equinus. For this usage, the TBS is molded with the hips in fifteen degrees of abduction, knee extension, and the ankle in neutral (17). For patients with lower levels of involvement, an AFO can be used at night to prevent equinus contracture. Whenever nighttime splinting is utilized, the patient and the family should be carefully educated on skin care and proper fit in order to prevent areas of pressure irritation.

In patients with thoracic and high-lumbar level involvement, orthoses are needed for upright weight bearing and mobility. A standing frame (Fig. 15-20), which is a prefabricated trunk–hip lower extremity brace, allows the child to stand without hand support. This is usually prescribed for children aged 12 to 18 months or once the child demonstrates adequate head and neck control. It should be used up to 3 hours a day, divided into periods of 20 to 30 minutes.

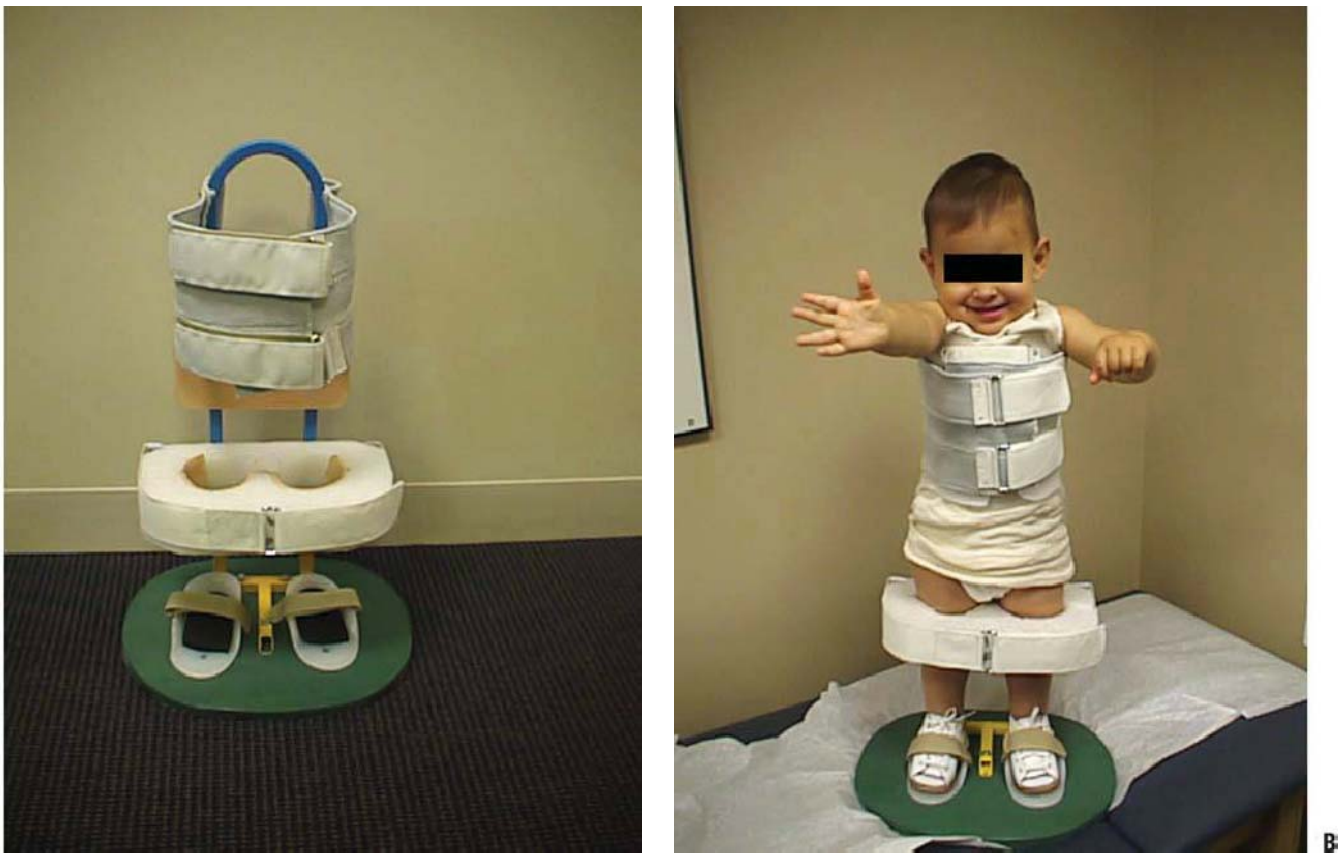

FIGURE 15-20. Standing frame, **(A)** alone and **(B)** with patient.

To achieve mobility, patients with thoracic and high-lumbar level involvement will require an orthosis that crosses the hip in order to control the trunk over the pelvis and lower limbs (95). Examples of this are the RGO (Fig. 15-1) and HKAFO (Fig. 15-2). The RGO, often used with a walker, is indicated for a child around 24 months of age with good sitting balance without hand support and good upper extremity function. An RGO is contraindicated in patients with severe scoliosis, hip flexion contracture >30 to 40 degrees, or severe visual deficit. As an alternative to the RGO, a parapodium (Fig. 15-21) is indicated for a child with poor trunk balance or upper extremity spasticity.

The HKAFO can be used for a patient with high-lumbar level involvement who has achieved swing-through ambulation with crutches. It is important however for providers to understand that most patients with higher levels of involvement will eventually opt to use a wheelchair for mobility. The wheelchair allows an energy-efficient means to achieve independent mobility. Several factors should be considered in the design of the wheelchair. In regard to the seat, special cushions may be needed to offload pressure areas and prevent decubitus ulcers over the ischium or the sacrum. Trunk supports should be added to the back rest as needed, and detachable arm rests allow for easier transfer in and out of the chair.

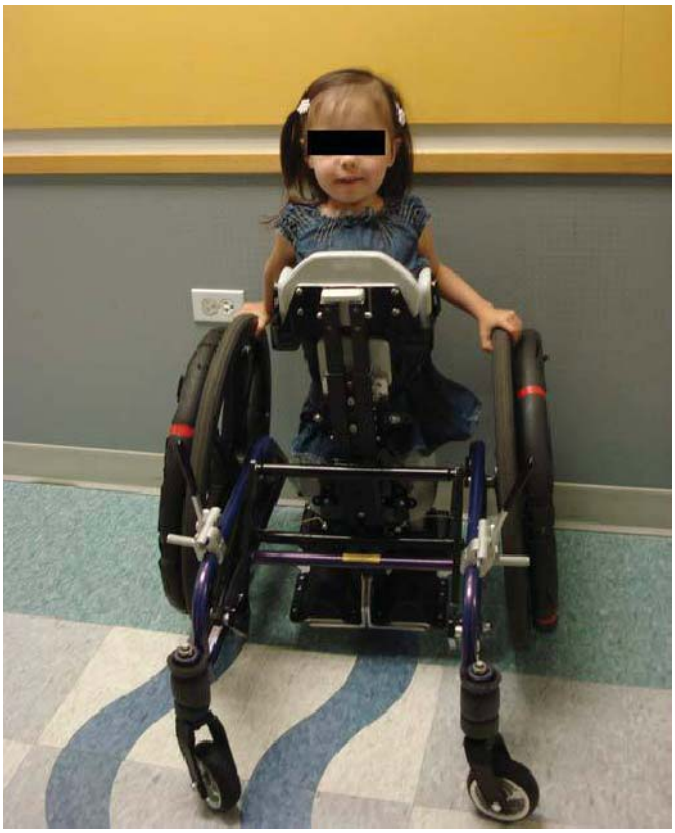

FIGURE 15-21. Parapodium.

Patients with sacral or low-lumbar level of involvement will require a solid AFO (Fig. 15-3) to compensate for muscle weakness below the knee (52). The AFO acts as a substitute for weak or absent ankle plantar flexors and dorsiflexors. The AFO should be designed to be rigid enough to provide ankle and foot stability while maintaining the shank-ankle angle at 90 degrees to prevent excessive dorsiflexion leading to crouch at the knee. Carbon reinforcement is often needed in the older child. In addition, special padding may be necessary over pressure points such as the medial malleolus and head of talus to prevent pressure sores. Certain patients with a tendency for crouch gait will benefit from use of a ground reaction AFO (Fig. 15-22) to assist with knee extension during stance.

Occasionally, a patient with low lumbar level of involvement will benefit from a knee–ankle–foot orthosis (KAFO) to prevent excessive valgus stress at the knee if the patient is too young for derotation osteotomy. Whether using KAFOs or AFOs, patients with low lumbar or high sacral level of involvement who have weakness of the hip extensors and abductors may benefit from the use of crutches to improve pelvic and hip kinematics. In this instance, crutches allow the upper extremities to share in weight bearing decreasing the stress on the lower extremity musculature and allowing a more functional gait pattern (52). Patients who are introduced to crutches at a young age are more receptive to their use compared to adolescent and young adult patients.

Rotational malalignment is common in patients with low lumbar and high sacral level of involvement. AFOs with twister cables are useful to correct either intoeing or out-toeing gait until an appropriate age for surgical correction is reached. These may be introduced as early as 2 years of age.

ADULT CARE

As overall care for patients with myelomeningocele improves and more and more patients are surviving into adulthood, increased attention is required to the issues unique to adult patients with myelomeningocele. It can be difficult for adult patients to find appropriate providers as few adult physicians have experience with the detailed care of patients with myelomeningocele. Ideally, adult care should be provided in a multidisciplinary setting similar to that for the pediatric patient.

Orthopaedic issues in adult patients tend to correspond to the patient's functional level of involvement. Thoracic level patients usually have an FMS of 1,1,1 or 2,2,1 and very occasionally 3,3,1. They have a high incidence of spinal deformity requiring surgical treatment and hip and knee flexion contractures. Even despite aggressive treatment during childhood, some amount of recurrence of contracture as an adult is common. Patients with low lumbar lesions are likely to maintain the ability to ambulate as adults (39) and often have an FMS of 3,3,1. To assist patients with maintenance of

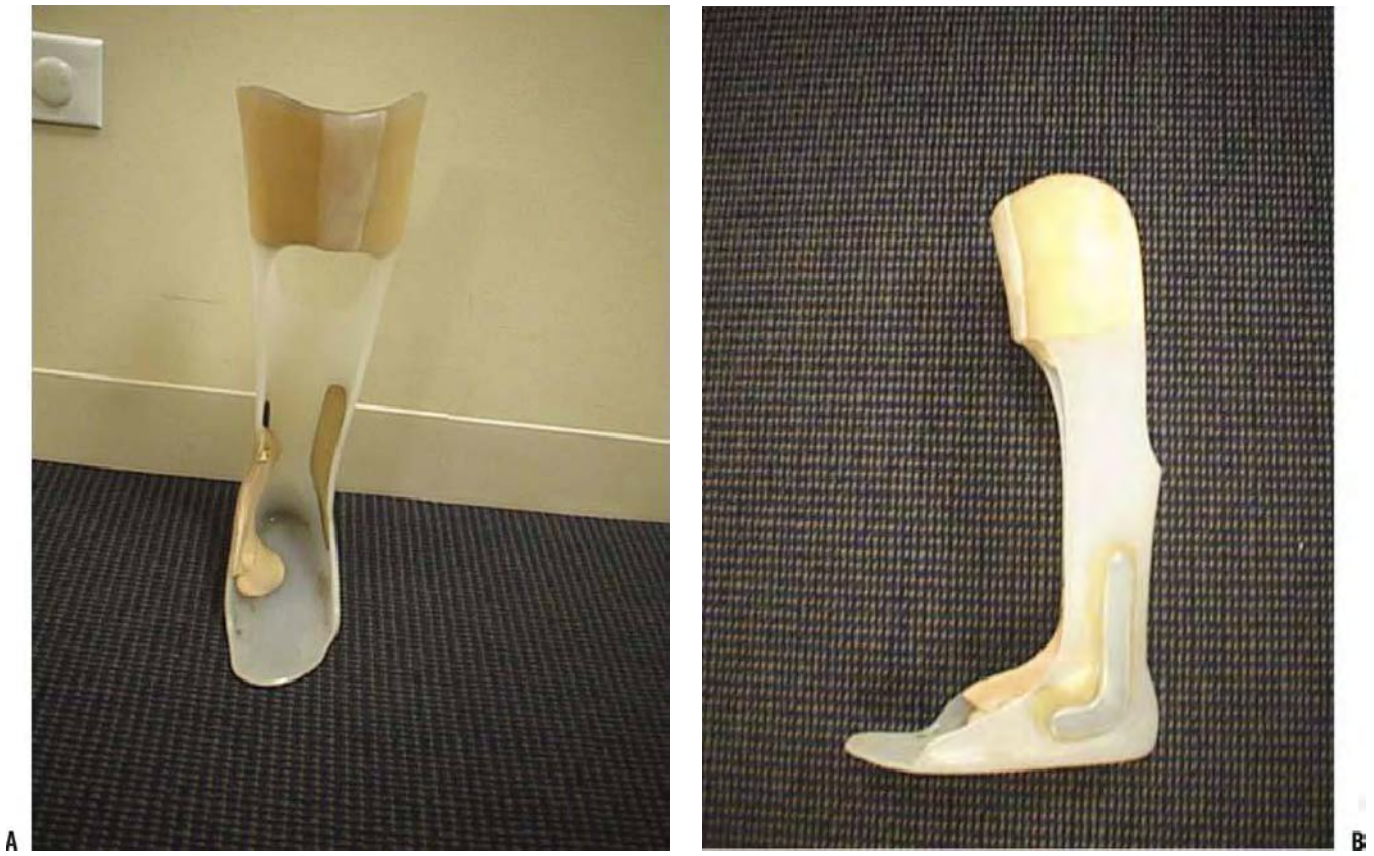

FIGURE 15-22. Ground reaction ankle foot orthosis (GRAFO). **A:** Front view. **B:** Side view.

ambulation, hip contractures should be treated aggressively as should any deformity of the knee, ankle, foot or rotational malalignment. The majority of patients with sacral level of involvement will maintain community ambulation as adults (38, 46) with an FMS of 6,6,3. As with low lumbar level patients, any rotational malalignment or deformity of the knee, ankle, or foot should be corrected. In addition, tethered cord syndrome should be treated aggressively and arthrodesis at the level of the foot should be avoided (46, 30).

Pressure sores are a major problem in adult myelomeningocele patients. In one study of 87 adult patients, 82% had experienced a pressure sore within the past 5 years (32). In this study, the sores were mainly located on the feet in areas of impaired sensation. The authors identified a significantly higher risk for pressure sores in patients with memory deficit, Arnold-Chiari malformation, and a history of previous sores. Patients with these conditions should be monitored closely and educated on a program of personal skin inspection and care.

In addition, we have observed a number of adult patients with thoracic or high-lumbar level of involvement who have developed severe lower extremity lymphedema. This causes problems with brace fitting and pressure sores leading to a functional decline. Prevention and treatment is with carefully fitted elastic compression stockings. If available, referral to occupational therapy or a lymphedema treatment clinic is beneficial.

The long-term outcome of adult patients with sacral level involvement has been evaluated in the literature (46,Brinker). Brinker et al. looked at a group of 36 patients ranging in age from 19 to 51 years followed for an average of 10 years. Although 97% patients were initially community ambulators, only 69% remained so at final follow-up. The authors also found a decrease in plantar sensation in 42% patients with skin breakdown in 75% patients. In addition, 64% patients had developed soft-tissue infections on the plantar surface of the metatarsal heads and heel. Forty-two percent of patients developed osteomyelitis necessitating a total of 14 amputations at various levels. In all 33 patients had undergone a total of 371 orthopaedic procedures.

Selber and Dias evaluated a group of 46 adult patients ranging in age from 18 to 38 years. In all 39 patients underwent 217 orthopaedic procedures. However, in contrast to Brinker et al, they found 89% patients maintained community ambulation at final follow-up, of whom 70% required no external support. In addition, only two amputations were performed. Selber and Dias attributed these results to aggressive treatment of tethered cord syndrome, surgical correction of musculoskeletal deformities, and avoidance of arthrodesis at the foot.

INTRASPINAL LIPOMA

Lipoma associated with the spinal cord, occurring in 1 in 4000 births, is the most common type of occult spinal dysraphism (132). Lipomeningocele is a subcutaneous lipoma connected to the conus medullaris by a vertebral and dural defect that can result in a tethered spinal cord and is the most common type. Other possibilities are intradural lipomas or lipoma of the filum (fatty filum terminale). In all, lipomas of the lumbosacral spine account for 25% to 35% of the cases of tethered cord syndrome (132).

Intraspinal lipoma is a separate entity from myelomeningocele with different embryogenesis, clinical presentation, and prognosis. Unlike with myelomeningocele, folate supplementation has not been shown to have an effect on reducing the incidence of intraspinal lipomas (133). Patients with intraspinal lipoma do not develop hydrocephalus or Chiari malformation and have normal intelligence (134). Also, the neurologic deficits resulting from tethered cord are asymmetric and can skip adjacent dermatomes (132).

Although most patients with intraspinal lipomas have normal neurologic function at birth, neurologic deterioration can occur at any age into adulthood. When not detected and treated appropriately, this can cause severe lower extremity dysfunction. With proper treatment and care, most patients with intraspinal lipomas maintain an FMS of 6,6,6. Rarely some patients have an FMS of 3,3,3.

Since the defect in patients with intraspinal lipomas is closed, the two main findings that prompt evaluation are cutaneous markers and neurologic deficits (133). The cutaneous manifestations of spinal lipoma include sacral dimples, masses, dermal sinuses, hemangiomas, and hairy patches in the lumbosacral area (132, 133). Neurologic deficits result from spinal cord tethering or compression of the cord and often occur during periods of rapid height or weight gain. Muscle imbalance caused by tethering of the cord leads to orthopaedic deformities, especially of the foot, which often require surgical correction.

Once tethering of the spinal cord causes muscle imbalance, surgical excision of the lipoma with cord untethering often does not lead to complete recovery (17). Hence, early aggressive neurosurgical treatment is indicated. In addition, since retethering occurs in approximately 30% patients (132), lifetime follow-up with manual muscle strength testing is recommended to facilitate early detection and intervention.

A recent review of 151 patients found acquired foot deformity was the most common orthopaedic manifestation occurring in 75% feet in patients with lipomeningocele (132). Of these, the most common deformity was cavovarus, followed by cavus with or without claw toes. Surgical correction (see "Cavus, Varus, and Cavovarus") was required in approximately 30% patients with lipomeningocele. Congenital foot deformities were also noted including clubfoot, vertical talus, and hypoplastic foot. Other common orthopaedic manifestations included scoliosis, which occurred in 20% of patients, none of whom required surgical treatment. Limb-length discrepancy was present in 11% patients. The shorter side was the side with weakness or deformity.

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