CHAPTER 14

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# Cerebral Palsy



## INTRODUCTION

Cerebral palsy (CP) is the most common cause of physical disability affecting children in developed countries. The prevalence is about 2 per 1000 live births and is not decreasing. Children with CP have complex needs and are usually managed by a multidisciplinary team. The medical literature dealing with CP is extensive and varies from level I (Randomized Clinical Trials [RCTs]), to cohort studies and case reports (levels IV and V). In this chapter, the most recent and the highest level of evidence will be cited, where possible. However, randomized trials in CP are difficult to perform and relatively few have been published, especially on the orthopaedic aspects of CP. When RCTs are not available, cohort studies, with long-term follow up and objective outcome measures, will be referenced.

## CEREBRAL PALSY: DEFINITION

Cerebral palsy was described in 1861 by the English Physician William Little, who recognized a link between difficult births and the development of deformities (1). For many years, CP was known as "Little's Disease." Little popularized tenotomy to correct deformity in CP and was the first to bridge the gap between neurology and orthopaedics. Although his understanding of the *link* between brain injury and deformity has stood the test of time, his views on the *causation* of CP have been superseded. It is now accepted that only 10% to 20% of CP is related to perinatal events. The influence of "difficult births" may have been much greater in Little's era, when maternal health was poor, maternal and infant mortality was high, and obstetric services were primitive.

The term cerebral palsy was also used by Sir William Osler in 1889 in a book titled "The Cerebral Palsies of Children" (2). Freud considered CP to be caused not just at parturition but also earlier in pregnancy because of "deeper effects that influenced the development of the foetus" (3). Many other definitions have been proposed and debated since then (4).

## THE 2007 REVISED DEFINITION AND CLASSIFICATION OF CEREBRAL PALSY

The revised definition of CP, published in 2007, is as follows:

*Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems* (5).

The new definition has been widely accepted and is recommended as the most useful current operational definition of CP. However, it should be remembered that there is no test, genetic, metabolic, immunologic or otherwise, that demonstrates the existence or absence of CP. There is no specified cause such as cerebral pathology or even type of motor impairment, only that motor impairment exists resulting from nonprogressive cerebral pathology, acquired early in life (6).

## BRAIN DEVELOPMENT AND GROSS MOTOR FUNCTION

During the first trimester of pregnancy, the growth of the brain is rapid and the brain differentiates into a recognizable cerebrum, cerebellum, brain stem, and spinal cord at a very early age of fetal development. During this time of explosive growth, the developing brain is highly susceptible to genetic influences, exogenous toxins, nutritional deficiencies and other insults, some of which can be characterized by meconium analysis (7). Neuronal development peaks in the second trimester. Neurons differentiate from neural stem cells around the periventricular regions and migrate centrifugally toward the surface of the cerebral cortex. This results in functional activity in neurons by 7 weeks of differentiation with reflex movements detectable in the fetus by the 15th week of gestation (8). By the end of the second trimester, the majority of neurons have been formed. Loss of neurons can be accommodated by neuronal plasticity but not by the generation of new neurons. The third trimester is characterized by extensive synaptogenesis and remodeling with glialization commencing in the second trimester and continuing at least until the age of 2 years. Myelination of neurons begins late in the third trimester, reaches a peak in the early years of childhood, and continues into adolescence, following a well-defined pattern (9). The myelination of complex pathways results in the progressive elimination of primitive reflexes, during the first 6 months of neonatal life as normal postural reflexes appear and the acquisition of gross motor skills occurs. In the typically developing infant, head control is achieved by age 3 months, independent sitting by 6 months, crawling by 8 months (usually accompanied by pulling to stand) and independent walking by the age of 12 months. However, even typically developing infants may take 3 to 6 months longer than these mean figures and still be considered to have typical development (10).

## MOTOR CURVES AND CEREBRAL PALSY

The development of gross motor function in children with CP can be described by a series of curves that were derived from longitudinal measurements of gross motor function, using the Gross Motor Function Measure (GMFM) (11, 12) (Fig. 14-1). The curves show rapid acquisition of gross motor function in infants with a progressive separation of the curves especially between the ages of 2 and 4 years. The curves plateau between the ages of 3 and 6 years. The five gross motor curves constitute the five levels of the Gross Motor Function Classification System (GMFCS) (11–15).

Understanding the position of a child's development in relation to their gross motor curve provides a rational basis for the understanding of management strategies, goal setting, and long-term gross motor function. For example, a 2-year-old child GMFCS level II with signs of spastic diplegia is treated with a physical therapy program, ankle-foot orthoses (AFOs), and injections of Botulinum toxin A (BoNT-A) to the gastrocsoleus and hamstring muscles. Within 3 months, the child is noted to have progressed from standing with support to independent walking. While the intervention may well have contributed to these gains in gross motor function, the child is at the stage of rapid acquisition of gross motor function with or without intervention (12). This underlines the need for intervention studies in the first 6 years of life to be controlled. The popularity of many forms of intervention in early childhood in children with CP is the mistaken attribution of improvements in gross motor function to the intervention, when natural history has an undoubtedly much greater effect. Association is not causation.

In the majority of children aged 6 to 12 years, gross motor function has reached a plateau (Fig. 14-1). At the same time, gait parameters are noted to show deterioration as contractures and bony deformities increase (16–19). Changes in gross

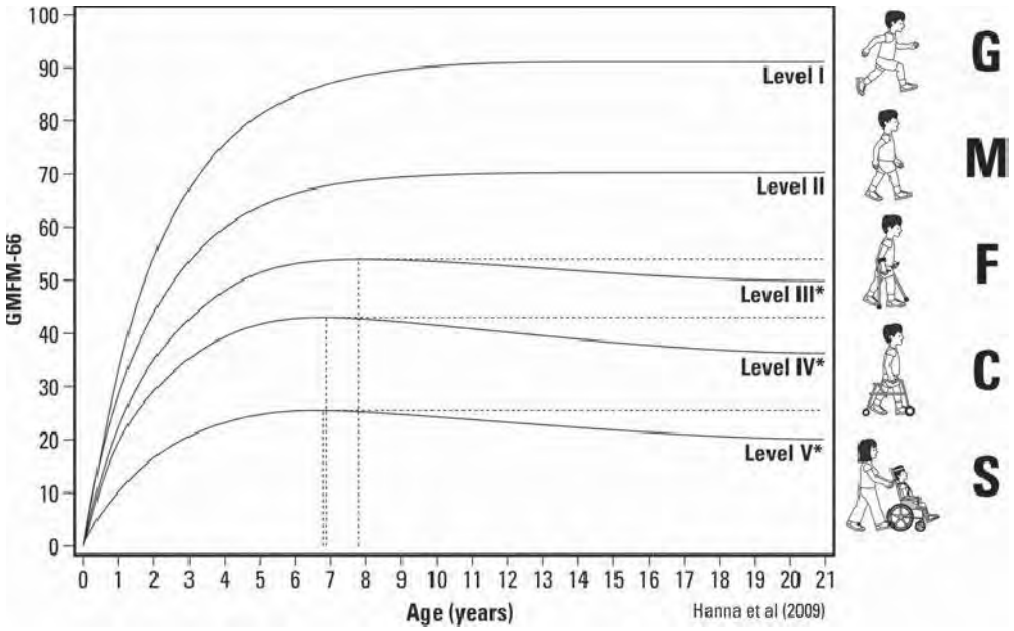



**FIGURE 14-1.** Gross motor curves in children with CP. The curves are based on longitudinal measurements of gross motor function, using the Gross Motor Function Measure (GMFM). Note the rapid acquisition of gross motor function between birth and age 2 years in all groups. Between the age of 2 and 6 years, the curves reach a plateau and level out into the five levels of the Gross Motor Function Classification System (GMFCS). (From Gallagher C, Sheedy M, Graham HK. Integrated management with botulinum neurotoxin A. In: Panteliadis CP, ed. *Cerebral palsy. A multidisciplinary approach.* Munchen, Germany: Dustri-Verlag; 2011:213–236, with permission.)

motor function and in gait during this plateau can be more realistically attributed to intervention, and longitudinal cohort studies are less liable to misinterpretation than in the birth to 6 years age group.

## PREVALENCE OF CEREBRAL PALSY AND CAUSAL PATHWAYS

The incidence of CP varies from 1 to 7 children per 1000 live births, according to maternal health, prenatal and perinatal maternal, and child health care services (6, 20). Prevalence rates are accurately reported in countries with well-developed health care services and are most reliable in countries with national CP registers including some European countries and Australia. In these countries, prevalence rates are comparable at around 2 per 1000 live births (20, 21). In most countries, prevalence rates are either static or increasing.

There is a paradoxical relationship between prevalence rates and the provision of neonatal intensive care. Sophisticated neonatal intensive care for premature and low birth weight infants may reduce the risk of brain injury in some and eliminate brain injury in other high-risk neonates. However, the lives of very premature and very low birth weight neonates with a severity of health problems, which would previously have resulted in premature mortality, are saved. These infants may survive with an increased risk of moderate and severe CP (6, 20).

Males are at higher risk of CP, perhaps due to gender-specific neuronal vulnerabilities (22). The risk of CP increases with decreasing gestational age. However, because births before 32 weeks contribute <2% of neonatal survivors, they contribute a minority (20% to 25%) of all CP in developed countries (6, 20, 23). The majority of CP cases are born at term.

The risk of CP increases 4-fold in twins and 18-fold in triplets (24–27). The widespread use of *in vitro* fertilization has greatly increased the rate of multiple births, which has resulted in an increase in CP rates (6, 28). The high CP rates in multiple births are in part explained by shorter gestation and low birth weight, but these are not the only factors. Any factor causing preterm birth may lie on a causal pathway to CP (6).

The introduction of statewide and national registers is extremely important in monitoring the prevalence of CP and detecting changes. In this way, causative factors and causal pathways may be identified that in turn may lead to primary and secondary preventive strategies (6, 21, 24).

## CENTRAL NERVOUS SYSTEM PATHOLOGY AND ETIOLOGY

CP is the most common cause of the upper motor neuron (UMN) syndrome in childhood, a syndrome characterized by positive features (spasticity, hyperreflexia, and co-contraction) and negative features (weakness, loss of selective motor control, sensory deficits, and poor balance) (Fig. 14-2). Clinicians have traditionally focused more on the positive features because it is possible to treat spasticity. However, it is the negative features, which determine the locomotor prognosis. Weakness and loss of selective motor control determine when or if a child will walk. Balance deficits may dictate long-term dependence on a walking aid (13, 15).

The brain lesion, which results in CP, is a "static encephalopathy." In other words, the brain lesion is not progressive and is unchanging. This is in contrast to musculoskeletal pathology in the limbs, which is progressive and constantly changing during growth and development (6, 29, 30).

At least 70% of cases have antecedents during pregnancy and only 10% to 20% have any relation to the child's delivery (6, 20, 31). The mechanism of causal pathways suggests that in any one case of established CP a number of factors may have contributed to the brain lesion resulting in the specific clinical phenotype. There are a number of genetic predispositions to CP that may require an addition of an environmental trigger such as a maternal infection to be expressed as a brain lesion and CP. The genotype may load the gun and the environment pulls the trigger. A large number of major brain malformations have a genetic basis, and subtle genetic polymorphisms may also play a role (31, 32).

About 10% of infants with CP weigh <1500 g at birth. In this low birth weight group, the risk of CP is 90 per 1000, compared to 3 per 1000 in infants born at term and weighing more than 2500 g. Maternal risk factors include viral infections, urinary tract infections in late pregnancy, dietary deficiency, some prescription drugs, drug or alcohol abuse, maternal epilepsy, mental retardation, hypothyroidism, preeclamptic toxemia, cervical incompetence, and third-trimester bleeding. Obstetric risk factors include multiple births, placental abruption, premature rupture of membranes, chorioamnionitis, and prolonged labor. Other obstetric factors include the administration of oxytocin, cord prolapse, and breech presentation, when accompanied by low Apgar scores (6, 28). Neither routine use of fetal monitoring during labor nor increasing rate of Cesarean section has resulted in a decrease in the prevalence of CP (6).

The older the child at the time of the acquired brain lesion, the more the clinical syndrome is likely to differ from classical CP. Age 2 to 3 years is an important watershed (5).

CP was formerly a clinical diagnosis with occasional confirmation by central nervous system (CNS) imaging. With the availability of MRI and safer anesthesia for children, the majority of children suspected of having a CP will have brain imaging (31). A recent practice parameter from the American Neurological Association recommended that the diagnosis of CP be confirmed by imaging (33). In a large recently published multicenter study, the brain lesions identified by MRI are given in Table 14.1.

On the basis of their MRI scans, only 20% of cases of CP were considered as *possibly* being secondary to some type

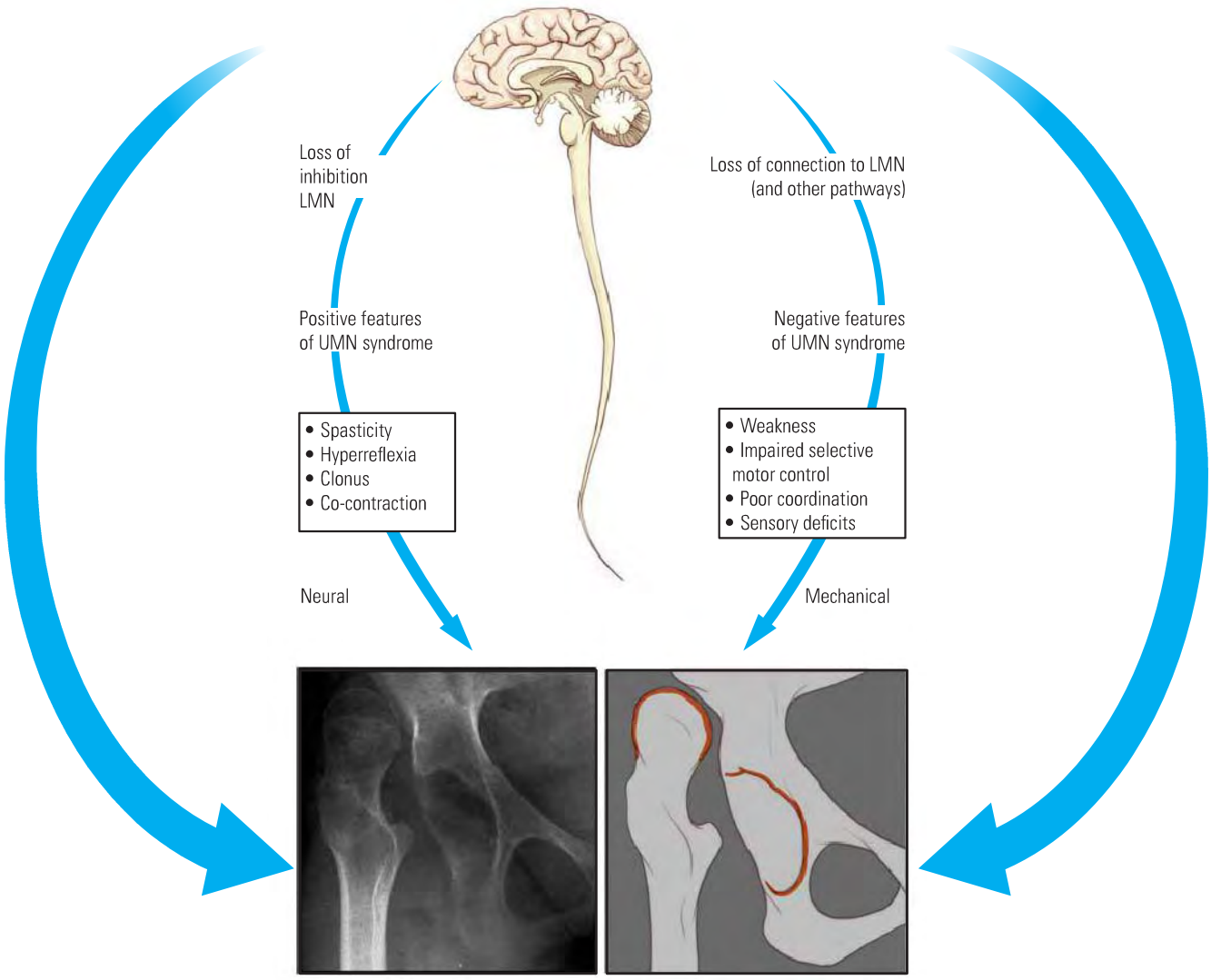



**FIGURE 14-2.** CP is a neuromusculoskeletal disorder. The CNS lesion has profound effects on the growing skeleton leading to deformities in both the upper and the lower limbs. Note the effects of both the positive and the negative features of the Upper Motor Neuron (UMN) syndrome.

of obstetric mishap. Twelve percent of cases, with a clinical diagnosis of CP, had a normal MRI scan (31).

## PROGRESSIVE MUSCULOSKELETAL PATHOLOGY: CP IS A NEUROMUSCULOSKELETAL DISORDER

By definition CP is a static encephalopathy, but the musculoskeletal pathology is progressive (6, 30). Chronic neurologic impairment affects the development of bones and muscles (Fig. 14-3). In spastic hemiplegia, the affected side demonstrates muscle atrophy and limb shortening, compared to the unaffected side. Thus, CP is a neuromusculoskeletal disorder (29, 30).

The key feature of the musculoskeletal pathology in CP is failure of longitudinal growth of skeletal muscle. An apt synonym for CP is "short muscle disease." The conditions for normal muscle growth are regular stretching of relaxed muscle, under physiologic loading conditions and normal levels of activity (29). In children with CP, skeletal muscle does not relax during activity because of spasticity and the

**TABLE 14.1 Frequency of Brain Pathology by MRI**

1. White matter damage of prematurity: 43%
2. Basal ganglia damage: 13%
3. Cortical/subcortical damage: 9%
4. Brain malformations: 9%
5. Focal infarcts: 7%
6. Miscellaneous lesions: 7%
7. Normal MRI: 12%

Bax M, Tydeman C, Flodmark O. Clinical and MRI correlates of cerebral palsy: the European Cerebral Palsy Study. *JAMA* 2006;296:1602–1608.

children have greatly reduced levels of activity because of weakness and poor balance (30). In animal models of CP, such as the hereditary spastic mouse, there is failure of longitudinal muscle growth, in relation to bone growth. Affected mice develop equinus deformities because of a failure of longitudinal gastrocsoleus muscle growth when compared to tibial growth (34). However, muscle growth can be enhanced by the injection of BoNT-A soon after birth (35). The newborn child with CP does not have contractures or lower limb deformities, and most do not show signs of spasticity (29, 30). With time, spasticity develops, activity levels remain low, the growth of muscle-tendon units (MTUs) lags behind bone growth and contractures develop. Although the MTU is short in CP, muscle fibers may not be short (36, 37). Because of disordered growth and abnormal biomechanics, torsional abnormalities persist or develop in the long bones and instability of joints including the hip and subtalar joint develop. Eventually, premature degenerative arthritis may develop (6, 30, 38–41).

An important therapeutic window exists for spasticity management before the development of fixed contractures (38, 39) (Fig. 14-3). A second therapeutic window exists for the correction of fixed musculoskeletal deformities, before the onset of decompensation (39). There are three important longitudinal studies of gait in children with spastic diplegia that confirm that the musculoskeletal pathology and the attendant gait disorder are progressive during childhood. These studies provide an important insight into natural history and a framework to interpret the results of surgical intervention (16–18).

## CLASSIFYING CEREBRAL PALSY

CP may be classified by the cause (when known) and the brain lesion as determined by MRI. Classification by movement disorder, topographical distribution, and gross motor function may be relevant to management, including orthopaedic surgery (13, 31, 42, 43) (Figs. 14-4 and 14-5A,B).

**Movement Disorder.** The most common approach to classification by movement disorder divides the disorders into pyramidal (spastic) and extrapyramidal (dystonic, athetoid) types (44). The majority of children with CP show features of both pyramidal and extrapyramidal involvement. When there is involvement of both pyramidal and extrapyramidal systems, spasticity and dystonia may coexist to varying degrees (43).

**Spasticity.** Spastic CP is by far the most common subtype and in most series comprises between 60% and 85% of all cases (21, 29, 43–46) (Fig. 14-4). A large population-based study of children with CP found that 85% of children had a primarily spastic movement disorder (21). Classically, spasticity is the result of a lesion affecting the pyramidal system and results in velocity-dependent increase in muscle tone with increased spastic tonic stretch reflexes. Spasticity is often associated with prematurity and the characteristic lesion of periventricular leucomalacia (PVL) on MRI (31).

**Dystonia.** Dystonia is the second most common form of movement disorder in CP and may not develop until late

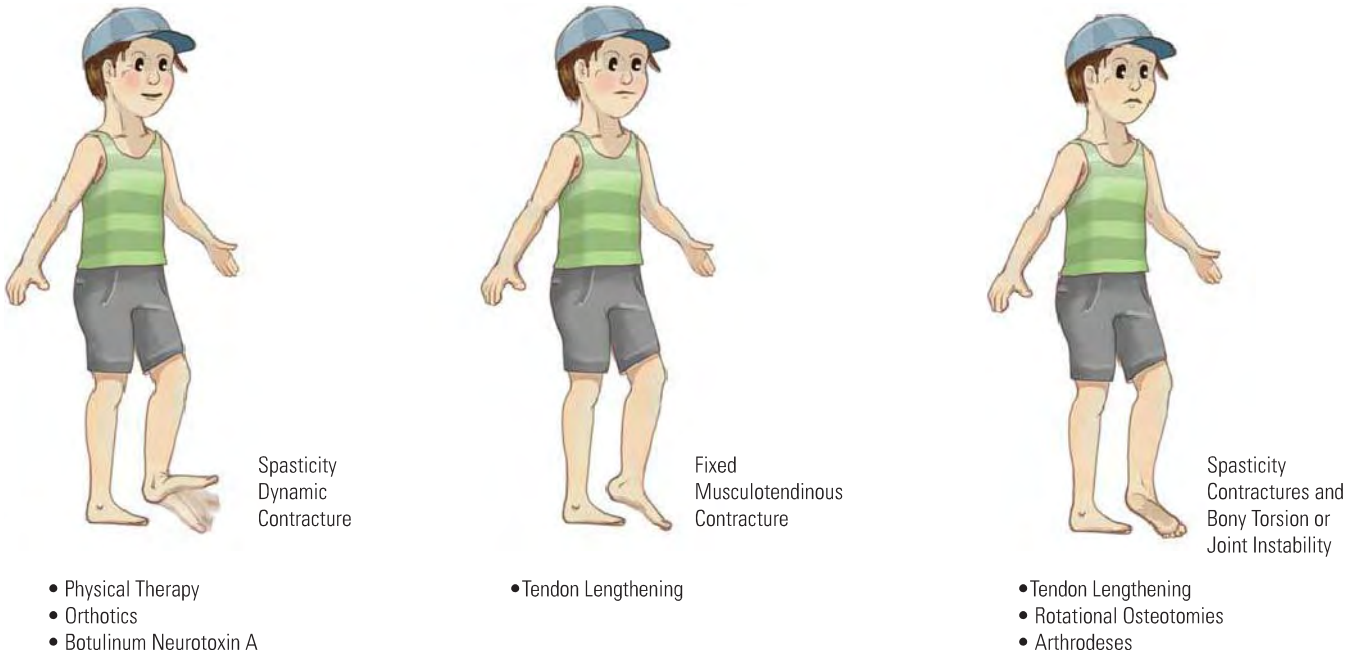



**FIGURE 14-3.** Musculoskeletal pathology in children with CP is progressive. At Stage 1, children have spasticity but no fixed contractures and can be managed nonoperatively. At Stage 2, there are fixed contractures and at Stage 3 contractures and bony deformities that may require corrective orthopaedic surgery. (Modified after Dr. Mercer Rang.)

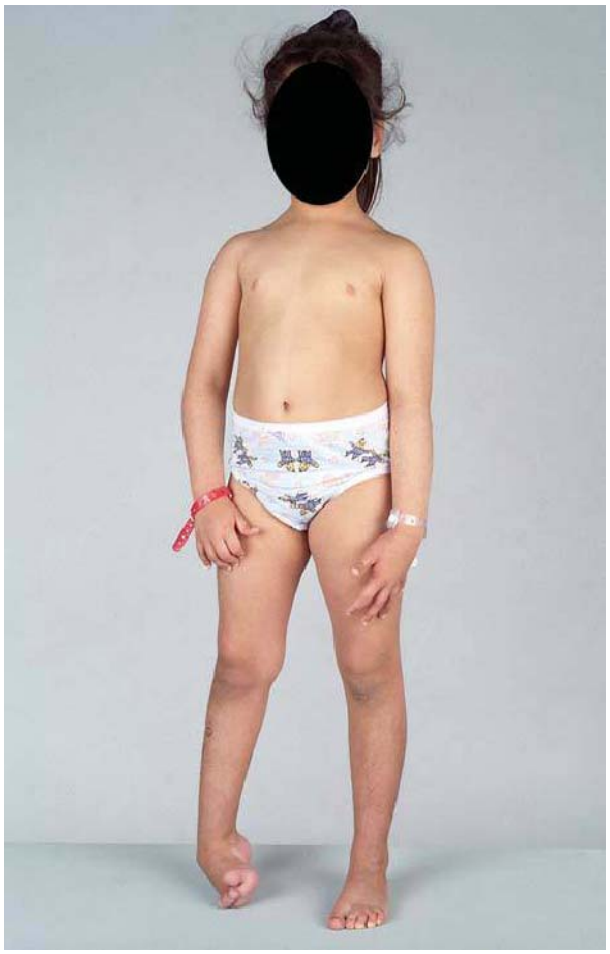


**FIGURE 14-4.** This girl has a right spastic hemiplegia. Her upper limb involvement is "dynamic." She has spasticity in her elbow flexors and this becomes apparent when she walks and even more pronounced when she runs. She has a severe varus deformity affecting her right foot because of spasticity in both tibialis anterior and tibialis posterior. The varus posture is consistent and does not change from day to day. Spastic CP tends to be predictable and amenable to corrective orthopaedic surgery.

childhood. Dystonia is often underreported in CP registers and population-based reviews (21, 45). Dystonia is diagnosed by observing abnormal twisting postures and writhing movements that vary in intensity. Dystonia may be triggered or worsened by attention, distraction, startling, overuse, fatigue, touch, or pain (43). Resting tone is variable and postures in both the upper and lower limbs vary with time. The brain lesion resulting in dystonia is usually in the basal ganglia and is more likely to be associated with a child who has had a term birth and widespread white matter lesions (31, 46). Uncontrolled movements seen in response to stimulation of the nervous system (e.g., volitional movements, loud noises, pain, etc.) are considered dystonic, while similar uncontrolled movements seen at rest are athetoid (43). Oral medications such as L-dopa and Artane may be beneficial in some children with dystonia. Severe dystonia can be managed by intrathecal baclofen (ITB) pump (38, 46) (Fig. 14-5A,B).

**Mixed Movement Disorder.** Many children with CP born at term have extensive brain lesions on MRI and have a mixture of pyramidal and extrapyramidal movement disorders. Defining the major movement disorder and the secondary and associated disorders can be challenging (43, 46). Dystonia and spasticity may occur in the same limb segments and distinction requires separation of the velocity-dependent from the action-induced and posture-responsive components of the hypertonia. Spasticity is evaluated by passively moving limb segments and joints at variable speeds. Dystonia is more easily confirmed by observation than palpation (39).

**Ataxia.** Ataxia may be part of a genetically determined syndrome and is uncommon as a pure movement disorder in CP (21, 45). Ataxia is disturbance of coordination and therefore most easily observed during walking. There may be associated signs of cerebellar dysfunction, including tremor. Because the resting tone is normal, and the majority of children have quite good walking ability, contractures are uncommon, and children with pure ataxia do not develop hip dysplasia or scoliosis (30).

**Hypotonia.** Many children who develop hypertonic CP are initially hypotonic, and this phase may last for several years. Hypertonia may not be expressed until myelination reaches a certain stage of completion (9) Many children with intellectual disabilities and certain other syndromes exhibit hypotonia, joint laxity, and developmental delay. Unless there is, in addition to the hypotonia, a defined static brain lesion, these children are not classified as having CP (6, 30).

## Topographical Distribution

### Unilateral Bilateral

1. *Unilateral*
   Monoplegia, hemiplegia
2. *Bilateral*
   Diplegia, quadraplegia

Topographical distribution is a classification of CP according to which limb segments are affected. As with classification by movement disorder, there is considerable variability in the terminology used, especially between different countries. The emphasis in Europe is the subdivision of CP into unilateral and bilateral types (44). The unilateral types can be subdivided into monoplegia (affecting only one limb) and hemiplegia (affecting one side of the body. The majority of children who seem to have a lower limb monoplegia with spastic equinus and toe walking, when asked to run, will show some abnormal upper limb posturing (21).

The common forms of bilateral CP are diplegia and quadriplegia (21, 45). Children with diplegia have bilateral lower limb involvement that may be symmetric or asymmetric. The involvement of the upper limbs is restricted to deficits in fine motor function, and overall upper limb function

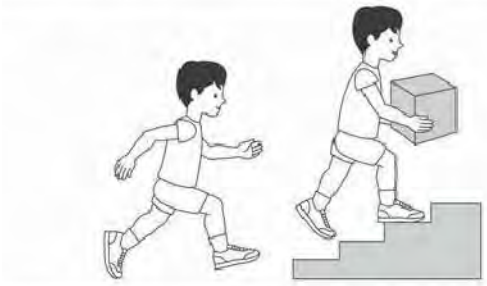

**FIGURE 14-5.** **A and B:** This teenage boy also has a right hemiplegia but his movement disorder is dystonic. He is using his uninvolved left hand to restrain his right hand. When the restraint is removed, his right arm undergoes variable dystonic posturing, with abduction at the shoulder, extension at the elbow, and abduction of the fingers. His dystonia is so severe that he requires a crutch and functions at GMFCS level III. Dystonic posturing can rarely be improved by orthopaedic surgery but may respond to medications including ITB.

is good. Spastic diplegia is most commonly associated with prematurity and PVL (31). The affected children usually have normal intelligence and a good prognosis for independent walking although many have visual deficits and learning difficulties. Quadriplegia refers to involvement of the upper and the lower limbs, and such children are usually born at term and have extensive brain involvement (46). They usually have a mixed movement disorder with spastic and dystonic features. The severity of involvement varies between the upper and the lower limbs and between the two sides. Because of the greater degree of brain involvement, quadriplegia is far more likely to be associated with comorbidities such as seizure disorder, learning challenges, and impairments of speech or cognition (5).

A small number of children appear to have a "triplegia" or three-limb involvement. This usually is a combination of hemiplegic involvement with both lower limbs involved to an asymmetric degree. One upper limb seems to be largely spared. This is an uncommon type of CP and is sometimes grouped with spastic quadriplegia (42, 45). Classification by topographical distribution is not a strictly functional classification, but there are functional implications. Almost all children with hemiplegia walk independently in the community, about 80% of children with diplegia walk either independently or with assistive devices, but only 20% of children with quadriplegia walk and then only with assistance (21). The separation of children into diplegia and quadriplegia is arbitrary and unsatisfactory, hence the need for a classification based on a valid and reliable measure of gross motor function (13, 42, 44).

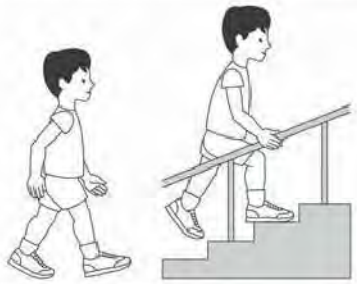
**Classification by Gross Motor Function.** The development of the GMFCS has for the first time given a common language to communicate about CP (13). The GMFCS is a five-level ordinal grading system in which a series of descriptors, supplemented by illustrations, can be used in five different age groups to classify gross motor function in CP (Figs. 14-6 and 14-7). The GMFCS has established validity

# GMFCS E & R between 6th and 12th birthday: Descriptors and illustrations



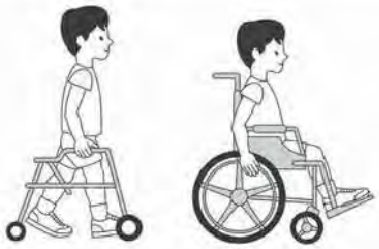
## GMFCS Level I

Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited



## GMFCS Level II

Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.



## GMFCS Level III

Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.

## GMFCS Level IV

Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.

## GMFCS Level V

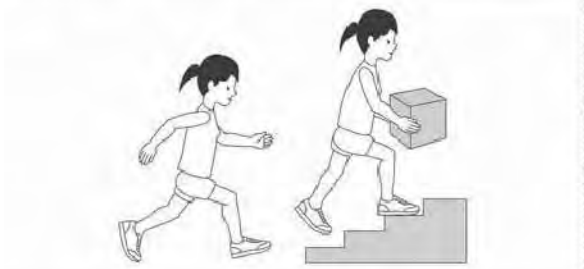
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca



**FIGURE 14-6.** GMFCS E & R (Expanded and Revised) between 6th and 12th birthday: Descriptors and illustrations.

# GMFCS E & R between 12th and 18th birthday: Descriptors and illustrations

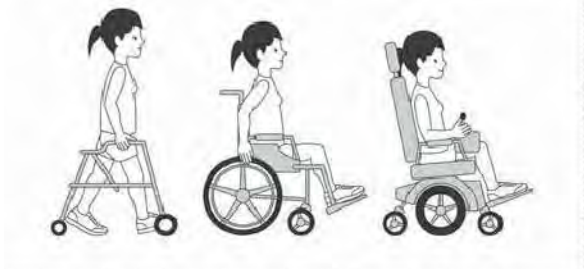


## GMFCS Level I

Youth walk at home, school, outdoors and in the community. Youth are able to climb curbs and stairs without physical assistance or a railing. They perform gross motor skills such as running and jumping but speed, balance and coordination are limited.

## GMFCS Level II

Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work they may require a hand held mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances.



## GMFCS Level III

Youth are capable of walking using a hand-held mobility device. Youth may climb stairs holding onto a railing with supervision or assistance. At school they may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community youth are transported in a wheelchair or use powered mobility.

## GMFCS Level IV

Youth use wheeled mobility in most settings. Physical assistance of 1-2 people is required for transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair.

## GMFCS Level V

Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca



**FIGURE 14-7.** GMFCS E & R (Expanded and Revised) between 12th and 18th birthday: Descriptors and illustrations.

(based on the GMFM), reliability, and stability. There is good agreement between clinicians and also between clinicians and parents (47). Given that the GMFCS is a grading system and not an outcome measure, it is the major prognostic information which must be considered in all children with CP. Knowing a child's long-term gross motor prognosis has management implications. For example, a child between the ages of 6 and 12 years at GMFCS level IV may perform some stepping with a heavily adapted walker, under the supervision of a therapist or a parent (13). However, following the pubertal growth spurt, useful walking is not sustained (15). It would be inappropriate to offer such children invasive treatments to improve or prolong walking because these will not be successful in the long term. Appropriate goal setting would be maintaining standing transfers (15).

The prevalence and severity of medical comorbidities shows good correlation with GMFCS (48). Severe respiratory disease, nutritional deficiencies, and premature mortality are largely seen at GMFCS levels IV and V. Children who are at GMFCS levels I and II do not have severe medical comorbidities (apart from epilepsy) nor do they show significant excess mortality.

Certain musculoskeletal features and deformities are also closely related to GMFCS level. The shape of the proximal femur shows a strong correlation with GMFCS level. Femoral neck anteversion (FNA) increases from GMFCS level I to level III and then plateaus at a mean of 40 degrees at GMFCS levels III, IV, and V. Mean neck shaft angle (NSA) increases stepwise from GMFCS levels I through to V (49). The incidence and severity of hip displacement is directly predicted by GMFCS level. In one study, children at GMFCS level I showed no hip displacement and those at GMFCS level V had a 90% incidence of having a migration percentage in excess of 30% (50). The relationship between GMFCS and hip displacement has direct implication for screening and management protocols.

## CEREBRAL PALSY: COMORBIDITIES

The spinal cord is not involved in CP and the most common neurologic impairment is epilepsy, affecting about 30% of children. Epilepsy is most commonly seen in hemiplegia, especially when it is acquired postnatally and in children with quadriplegia (48, 51). Intellectual disability is variable and more severe in children at GMFCS levels IV and V. However, many children at GMFCS levels I to III exhibit learning difficulties, autism spectrum disorders, and behavioral and emotional difficulties. Impairments of hearing, speech, and vision are also common and may adversely impact schooling and learning. The prevalence of visual problems is so high (about 50%) that routine screening is advised (52).

**Respiratory Disorders.** Children with CP have excess mortality at GMFCS levels IV and V, the levels previously described as spastic quadriplegia. The commonest cause of death is from respiratory disease especially aspiration pneumonia (48, 53). A major risk factor for respiratory disease is "pseudobulbar palsy" in which there are varying combinations of impaired swallowing, esophageal reflux, aspiration, and chest infection. Nocturnal coughing and asthma are also very common. The management of chronic respiratory disease involves careful assessment of swallowing and may necessitate investigation of the upper gastrointestinal tract. Fundoplication and the use of feeding tubes are often beneficial in the management of severe respiratory disease, when aspiration has been confirmed to be a major contributory factor (54).

**Gastrointestinal System.** The most severe problems are in GMFCS levels IV and V (48). Pseudobulbar palsy leads to impaired swallowing, vomiting, esophageal reflux, and aspiration. Oral feeding can be slow and inefficient leading to chronic malnutrition, impaired growth, and poor nutritional reserves (55). Malnourished children are at greatly increased risk of postoperative complications after hip reconstruction and scoliosis surgery. Assessment of nutritional status includes a careful dietary history and a radiologic assessment of swallowing in those with suspected gastroesophageal reflux. Nutritional status can be assessed by measurement of serum albumin, iron, and transferrin levels and examination for iron deficiency anemia. Correction of malnutrition may require the introduction of improved oral feeding. In many children, a percutaneous enterogastrostomy or a jejunostomy may transform the child's nutritional status and general health. Gastroesophageal reflux may respond to medical management but often requires a fundoplication (55, 56).

Of all problems affecting the gastrointestinal system, slow transit and chronic constipation is by far the most common (56). The excessively loaded colon is often noted as an incidental finding during hip surveillance radiographs. Chronic constipation assumes even more importance in the perioperative period when fecal impaction, vomiting, and the inability to achieve satisfactory intake of food and fluids postoperatively causes severe distress and results in prolonged hospitalization. Strategies to ensure that children at GMFCS levels IV and V have a satisfactory bowel management program prior to admission for orthopaedic surgery are advised.

## DIAGNOSIS AND ASSESSMENT: THE DIAGNOSTIC MATRIX

In the majority of children who are referred to an orthopaedic surgeon, the diagnosis of CP has already been established by a pediatrician or a pediatric neurologist. The exception is a small number of children with mild hemiplegia and monoplegia who present after walking age with toe walking or some other subtle disturbance of gait. The postural abnormalities associated with monoplegia and hemiplegia are best observed by asking the child to walk and then run in a sufficiently long, well-lit corridor (Fig. 14-8).

**History.** The "neonate at risk" and the majority of children with mild CP are diagnosed because they are observed to have

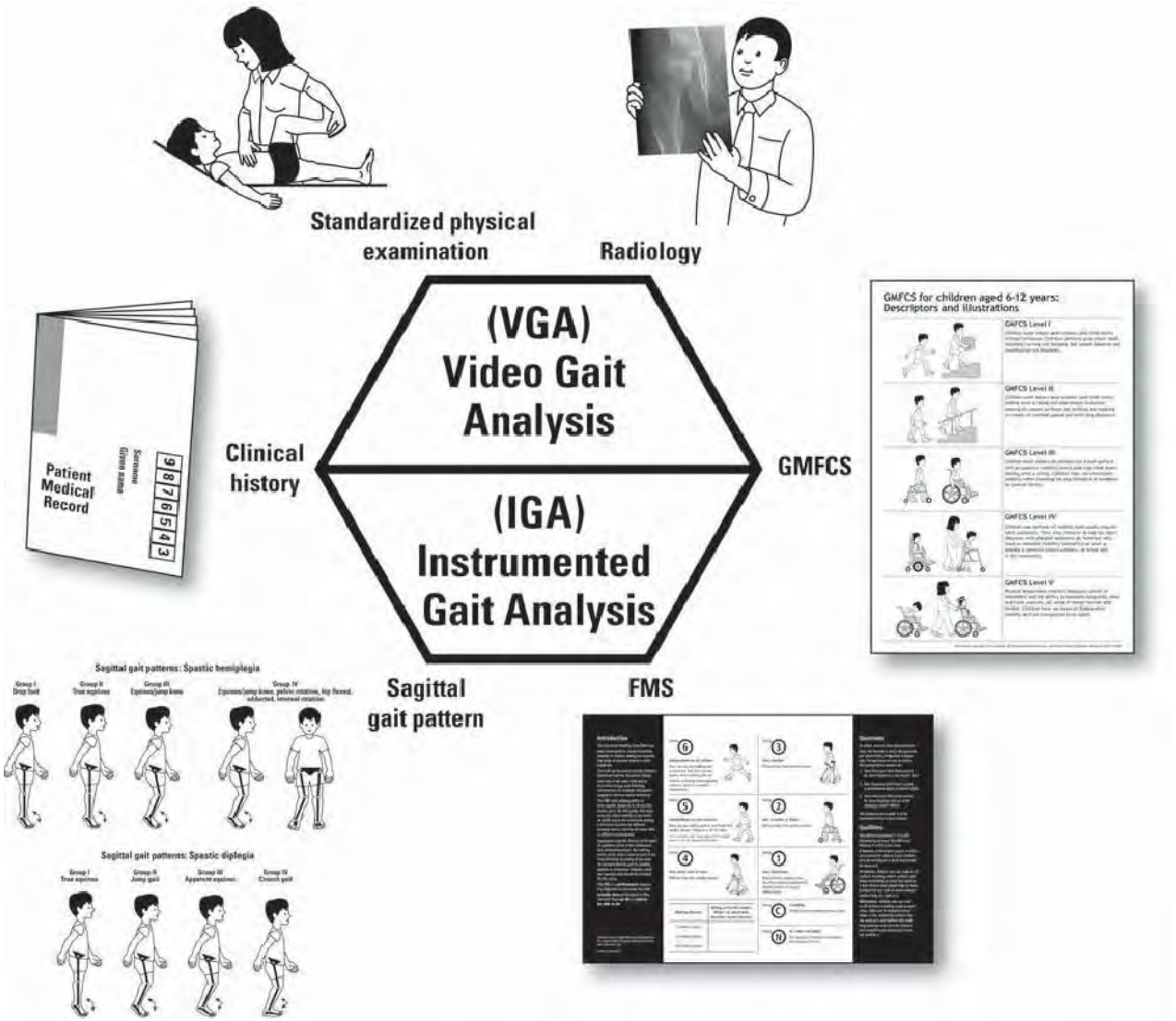



**FIGURE 14-8.** The diagnostic matrix consists of a standardized approach to clinical history, physical examination, radiology, GMFCS, functional scales (FMS and FAQ), and sagittal gait pattern identification. These components can be used in conjunction with either video gait analysis (VGA) or instrumented gait analysis (IGA). (Illustration copyright © Kerr Graham, Bill Reid and Adrienne Harvey.)

a delay in reaching gross motor milestones. A knowledge of the normal age (and range) of achieving these milestones is important. In typically developing infants, head control is acquired at age 3 months, sitting at 6 months, crawling and pulling to stand at 8 to 9 months, and independent walking at 12 months (10).

Recognition of abnormal muscle tone is also important (43). This may include hypotonia in the infant and hypertonia in the child. The mothers of children with CP, who have previously had a typically developing child, often sense that "something is wrong" at a very early stage. It is clearly wise to take the concerns of an experienced parent seriously. Details of the history of the pregnancy, the birth, and early development are very important in establishing the diagnosis of CP. The family history is important to detect such conditions as hereditary spastic paraplegia (HSP) and congenital ataxias (57).

**Physical Examination.** Physical examination is important in both preliminary examinations to establish the diagnosis of CP and in subsequent assessments in which the child's tone, gross motor function, and secondary musculoskeletal pathology are evaluated:

1. Observation of posture, movement and gait
2. Assessment of gross motor function: GMFCS, FMS, and FAQ
3. Evaluation of muscle tone by a combination of observation, palpation, and testing of reflexes.
4. Assessment of soft-tissue contracture by evaluation of passive joint range of motion and muscle length measurement.
5. Assessment of torsional abnormalities in the long bones: FNA; tibial torsion; deformities of the spine, hands, and feet.
6. Sensory evaluation: especially the hemiplegic upper limb.

**Assessment of Gross Motor Function: GMFCS.** Knowing the child's GMFCS level is fundamental in establishing gross motor prognosis and monitoring changes. The GMFCS can be easily and reliably determined by age 4 years and all orthopaedic surgeons should be competent in assigning a grade after the age of 6 years. Changes in GMFCS levels should be carefully documented. The most common reason for a change in GMFCS is an error in the previous or current examination (58). Given that the GMFCS is a categorical grading system, true changes in GMFCS level sometimes occur and these may occur in both directions, that is improvement or deterioration. After major intervention such as selective dorsal rhizotomy (SDR) or multilevel surgery, a small number of children move up a level, but this is uncommon and should not be expected in more than about 5% to 10% of a study population. Deterioration in GMFCS level is more common. Lengthening of the Achilles tendons in children at GMFCS level II can result in progressive crouch gait and the need for assistive devices. Such children may deteriorate from GMFCS level II to III (59).

The GMFCS is a classification system and not an outcome measure. It is expected to be stable throughout a child's growth and development but is often misused as a proxy outcome measure in intervention studies. Simple scales of gross motor function that can be used as outcome measures are the Functional Mobility Scale (FMS) and the Functional Assessment Questionnaire (FAQ) (60, 61).

The World Health Organization's International Classification of Functioning (ICF) describes health conditions in several domains, including body structure and function, activities, and participation (62) (Fig. 14-9). These domains are modified by environmental factors and personal factors. Various tools exist to measure parameters relevant to CP in the ICF domains and new measurement tools are being developed. Fortunately, there are now valid and reliable tools to classify and measure gross motor function in both the upper and lower limbs.

**Functional Mobility Scale.** The Functional Mobility Scale describes the level of assistance a child requires to mobilize in three different environments, the home (distances of up to 5 m), school (distances of up to 50 m), and the community (distances of 500 m) (60). Hence, three numbers are assigned depending on the level of assistance required in each of these settings. For example, a child at GMFCS level III is frequently capable of independent walking in a sheltered familiar environment such as the home. The same child may require the use of Canadian crutches to move around in the school environment but may be too slow to keep up with the rest of the

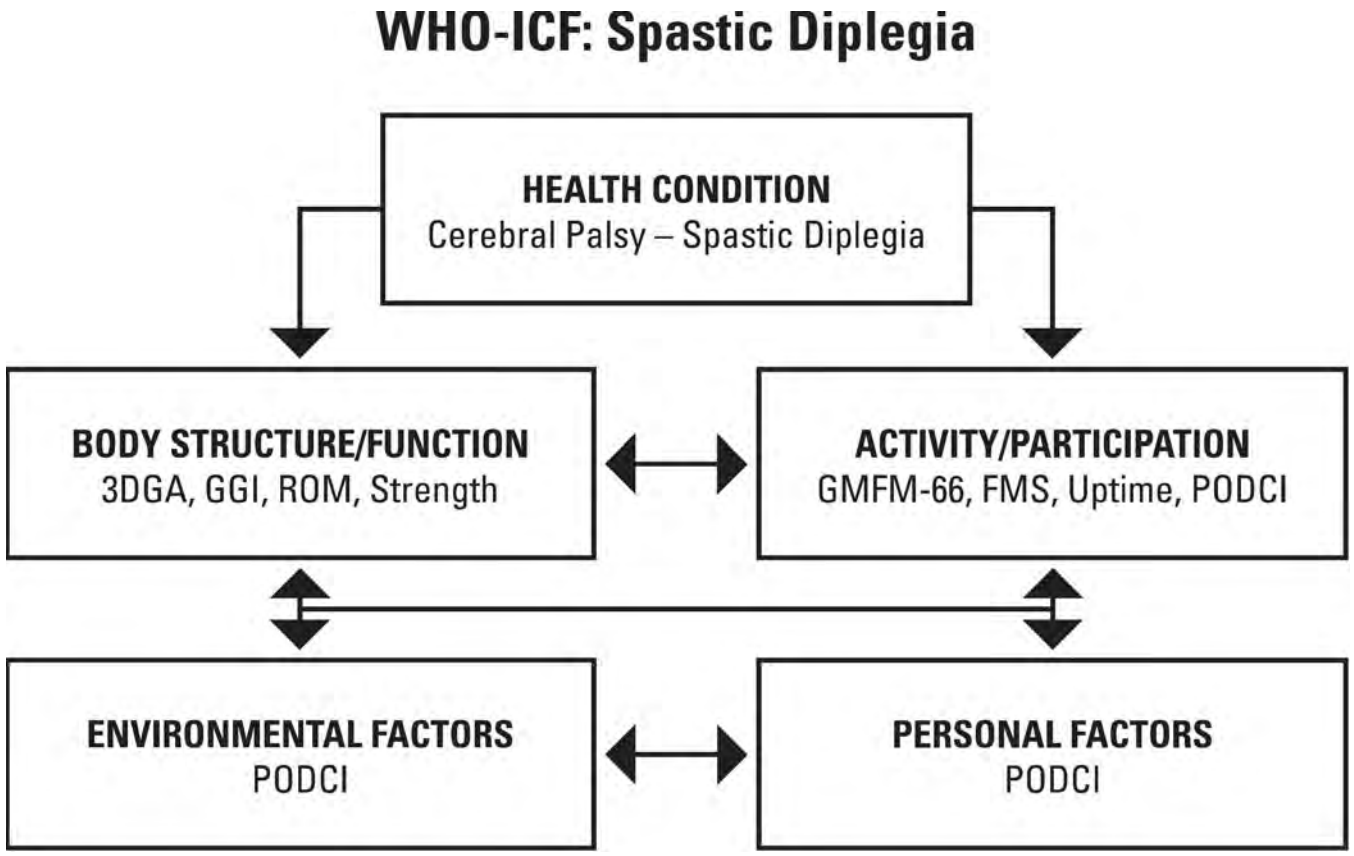



**FIGURE 14-9.** The World Health Organization International Classification of Functioning (WHO-ICF) as applied to spastic diplegia. Suggested assessment tools are indicated in each domain. (3DGA, three-dimensional gait analysis; GGI, Gillette Gait Index; PODCI, Pediatric Outcomes Data Collection Instrument.)

family during trips to the shopping mall, when a wheelchair may be preferred. The FMS grading for such a child is 5, 3, 1. Unlike the GMFCS, the FMS was designed as an outcome measure and is sensitive to change (59, 63). For example, children at GMFCS level III often require a posterior walker to ambulate prior to multilevel surgery. After optimum biomechanical realignment and correction of spastic contractures, these children can often progress to lesser levels of support. Some will be able to walk increasing distances independently; others will require crutches or sticks when previously they were dependent on a posterior walker. These important changes can be monitored and reported using the FMS (63).

**Functional Assessment Questionnaire.** The Gillette FAQ is a 10-level, parent report walking scale that describes a range of walking abilities across the spectrum of CP, from nonambulatory to independent ambulation at a high level. In addition to the 10-level walking scale, there is an additional list of 22 items describing a variety of higher-level functional activities requiring varying degrees of walking ability, balance, strength, and coordination. The scale has been shown to be reliable and validity has been established by comparison with other scales used in neuromuscular diseases. It has also been shown to have sensitivity to change with improvements seen in children with CP after such interventions as SDR and single-event multilevel surgery (SEMLS). It is a simple scale, which can be quickly completed by parents or caregivers and provides an excellent longitudinal view of the child's gross motor and walking abilities.

The FMS and FAQ are complementary scales and are both gaining acceptance in assessing children with CP as baseline measures and as outcome measures after intervention (59–61, 63).

**Additional Diagnostic Tests.** Between 10% and 20% of children with a clinical syndrome typical of CP will have a normal MRI of the brain and spinal cord (31). In these children additional investigation for causes such as HSP, genetic, dysmorphic, metabolic and muscular diseases and syndromes is important (57).

**Longitudinal Assessments with Radiology: Hip Surveillance.** The early stage of hip displacement is silent and formal screening by radiographs of the hips with careful positioning is advised (64, 65). The frequency of such radiographs should be directly related to the risk of hip displacement which is in turn related to the child's GMFCS level. At GMFCS level I, there is little risk of hip displacement and radiographs are only required if there are findings on clinical examination. More regular radiographs are required at GMFCS levels II and III. Radiographs every 6 to 12 months may be required at GMFCS levels IV and V to monitor progressive hip displacement (66).

Deformities of the feet are very common in all GMFCS levels. Standing weight-bearing radiographs of the feet are extremely useful in the longitudinal assessment of progressive foot deformity. Recently, a series of radiologic indices have been published in typically developing children that are an excellent baseline with which the results in CP can be compared (67).

**Instrumented Gait Analysis.** Evaluation of gait and functioning in children with CP can be considered in the format of a diagnostic matrix (68). The role of instrumented gait analysis (IGA) is crucial to the evaluation of gait dysfunction, especially in relation to planning and assessing the outcome of major interventions such as SDR and SEMLS (69). Historically, problems with reliability undermined the utility of IGA and the confidence in using such information for planning interventions (70–72). Recent work from several centers has reestablished confidence in the reliability of gait kinematics (73, 74). Only about half of the orthopaedic surgeons in North America who care for children with CP have access to IGA (75). There are children with symmetric gait deviations that through the eyes of experienced examiners are relatively easy to recognize. Even for those with experience, there are many children with complex movement disorders, asymmetric gait patterns, and complexities that require IGA for understanding and planning (40, 69). As gait analysis becomes more reliable, cheaper, and more accessible, the quality of assessments and outcomes should continue to improve.

**Video Gait Analysis.** Even in centers with access to IGA, video gait analysis (VGA) is a central part of the diagnostic matrix (38). A visual record of a child's gait and functioning on digital video is of much greater value than observational gait analysis and a written report (39). Digital video can be archived in a permanent fashion, is objective, and can be shared by multiple observers over time. It allows observation and recording of gait from multiple viewpoints, can be replayed in slow motion, and can be reviewed repeatedly, including late in the evening prior to an operating list, when real-time observation of a child's gait is not feasible (40). In an effort to quantify and objectify the outcome of observational gait analysis, a number of gait scores have been developed of varying degrees of complexity, sophistication, and reliability. These include the Physician Rating Scale, the Observational Gait Scale, and the Edinburgh Visual Gait Score (76–78).

VGA has wider application than IGA. IGA requires a child to be about a meter tall and to be able to follow simple commands over a 2-hour testing period. There is much useful information to be gained in children with hemiplegia and diplegia from when they first start to stand and walk which cannot be obtained from IGA. Longitudinal assessment of children following interventions such as injection of BoNT-A, the prescription or modification of orthoses, is also conveniently achieved using serial VGA (39, 79). Gait deviations for children at GMFCS IV may be so severe that the extra cost and effort of IGA may not be necessary (15). VGA and physical exam may be all that is needed.

## PHYSIOTHERAPY AND OCCUPATIONAL THERAPY IN CP

Physiotherapy is the most popular and widely used management strategy in children with CP (80–83). Some have access to therapy services integrated within the school program and others outside of the school program. The frequency of such programs may be related to the severity of the individual child's involvement but rarely reflect the family and the child's real needs. This type of background physical therapy support is valued by the parents of children with CP. These programs provide education and emotional support to the parents coping with the diagnosis of CP and starting the journey through the uncharted waters of raising a child with a lifelong physical disability. Roles that may be assumed by therapists may include education, counseling, coordination of access to other services including physical medicine and rehabilitation, orthotics, and orthopaedic surgery. The parents will often seek the view of the child's therapist on recommendations for spasticity management, the type of orthosis, and the timing and type of surgical intervention (80). The need for clear communication and teamwork within the multidisciplinary team is obvious. These "background" programs of physical therapy are rarely adequate to ensure an optimum rehabilitation from major interventions such as SDR or SEMLS. Therapy around such episodes needs to be carefully planned and is often best as a team approach involving the child's community therapist as well as providing the additional therapy in the tertiary hospital or rehabilitation center.

The physical therapy management of children with CP has been based on a variety of theoretical perspectives. The theoretical frameworks most commonly applied can be broadly categorized into biomechanical (splints, orthoses, stretching, and strength training), neurodevelopmental, cognitive (including conductive education [CE] and motor learning), and constraint-induced movement therapy (CIMT) (80–83). These approaches are not mutually exclusive. They can have shared components and most therapists use a combination of approaches. Given that the surgical management of CP has a biomechanical basis, surgeons find it easier to communicate with therapists who share this approach (80).

**Biomechanical Approach.** The biomechanical approaches (80) aim to maintain range of motion and muscle length. Techniques used include manual passive ranging of joints, stretching of muscles, and splinting and casting. Serial casting can be combined with Botox injections, for dynamic contractures (84). The introduction of a suitable ankle foot orthosis is often a critical step in providing an improved base of support and achieving progress in a child progressing to standing and walking (38). Muscle weakness is a significant functional and biomechanical problem in children with CP. In children with hemiplegia, muscle weakness is also often compounded by nonuse of the affected limb (39). Progressive resistance strength training may be used to increase muscle strength and endurance (85).

**Neurodevelopmental Therapy: The Bobath Approach.** Neurodevelopmental therapy (NDT) (81) is a widely used approach in children with CP. It aims to inhibit abnormal, primitive postural patterns and facilitate developmentally more mature movement patterns with improved function. NDT therapists use specific physical handling, including facilitation, to provide sensorimotor input and to guide motor output. Inhibitive casting is sometimes used with NDT to help achieve a reduction in tone and increased range of motion.

**Cognitive Approach.** Cognitive or learning approaches (82) focus on learning control of movement for function rather than emphasizing quality of movement. Motor learning or training programs use task analysis to breakdown functional tasks into basic motor components or patterns. These components are then practiced and learned as a motor skill for functional use. Conductive Education (CE) or the Peto method is a cognitive approach that utilizes rhythm, music, and counting to initiate and moderate movements. CE is used extensively by therapists to develop skills necessary for the performance of daily activities such as dressing and self-feeding.

**Constraint-Induced Movement Therapy.** CIMT (83) is used for learned nonuse of the affected upper limb, which is common in hemiplegia (39). It involves the forced use of the most affected upper limb by restricting use of the less involved hand in a cast, splint, or glove for periods of activity during the day.

The approach used by occupational therapists and physical therapists to manage children with CP is influenced by many factors. Intervention requires sensitivity to the child's age, cognitive, sensory, and perceptual factors as well as type and severity of CP. The child's environment, including family and culture, is important. Interventions will be less effective if they are not carried over into the child's management at home. The treatment setting, whether based in a hospital, at school or home, and the focus of these services are also factors. The setting and the frequency of treatments are often influenced by the financial resources available.

Despite widespread use, evidence on the efficacy of physiotherapy to improve function in children with CP is equivocal. In randomized, controlled trials, treatment effects have generally been small.

## MOVEMENT DISORDER MANAGEMENT AND THE SPASTICITY COMPASS

The "spasticity compass" can be used to classify and compare interventions for spasticity management, as focal or generalized in their effect and as temporary or permanent (84). Each intervention can then be located in the appropriate quadrant. For example, oral medications are general (all nerves or all muscles in all body areas) but temporary in effect (86). SDR is a neurosurgical procedure in which 30% to 50% of the dorsal rootlets between L1 and S1 are transected for the permanent relief of spasticity in a highly selected group of children with

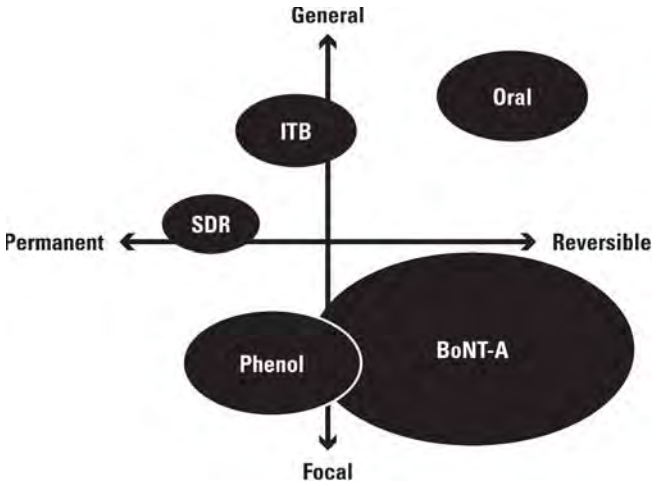



**FIGURE 14-10.** The spasticity compass as a guide to movement disorder management in CP. Interventions are classified on the north–south axis according to whether they are "general" or "focal" in their action. They are also classified on the east–west axis as to whether they are "permanent" or "reversible." (From Gallagher C, Sheedy M, Graham HK. Integrated management with botulinum neurotoxin A. In: Panteliadis CP, ed. *Cerebral palsy. A multidisciplinary approach.* Munchen, Germany: Dustri-Verlag; 2011:213–236, with permission.)

spastic diplegia. The principal effects are on the lower limbs although there may be minor effects on the upper limbs. The position on the grid is therefore permanent and half way between general and focal (87, 88) (Fig. 14-10).

Oral medications used for the management of spasticity in children with CP include diazepam, baclofen, dantrolene sodium, and tizanidine. Artane and L-dopa are used in dystonia. All are limited in usefulness by a combination of limited benefits and side effects. They have been extensively reviewed in several recent publications (84, 89).

The limited lipid solubility of baclofen when administered orally can be overcome by intrathecal administration using a programmable, battery-operated implantable pump connected to a catheter and delivery system to the intrathecal space (86). This is an invasive procedure with associated morbidity and mortality (38, 39, 88). However, it is the most effective current method available for the management of severe spasticity, dystonia, and mixed movement disorders in CP and a number of other conditions including spasticity of spinal origin and acquired brain injury. The role of ITB has been reviewed extensively in several recent publications (86, 88).

Chemodenervation is useful in the management of focal spasticity and dystonia. Phenol neurolysis was much more widely utilized before the introduction of Botulinum neurotoxin A (BoNT-A). The principal limitation on its use is pain at the site of injection and post injection dysesthesia. Phenol is not selective and has the same effect on sensory nerve fibers as motor fibers. The principal indications are neurolysis of the musculocutaneous nerve for elbow flexor spasticity and the obturator nerve for adductor spasticity. These nerves have limited sensory distribution (84, 90).

## BOTULINUM NEUROTOXIN A IN CEREBRAL PALSY

Injection of skeletal muscle with BoNT-A results in a dose-dependent, reversible chemodenervation, by blocking presynaptic release of acetylcholine at the neuromuscular junctions (84). Because of the toxin's rapid and high-affinity binding to receptors at the neuromuscular junctions of the target muscle, little systemic spread occurs. Neurotransmission is restored first by sprouting of new nerve endings, followed by the original nerve endings regaining their ability to release acetylcholine (91, 92). BoNT-A may be useful in children with CP to manage dynamic gait problems and to delay the need for orthopaedic surgery until the child is older (Fig. 14-11).

**Spastic Equinus.** The most common and most important indication for BoNT-A therapy in children with CP is the injection of the gastrocsoleus for spastic equinus (93, 94). Before widespread use of BoNT-A for spastic equinus, the majority of children with CP who walked on their toes had a lengthening of their Achilles' tendons by age 4 to 6 years (95). This resulted in crouch gait that was much more disabling than the original equinus gait.

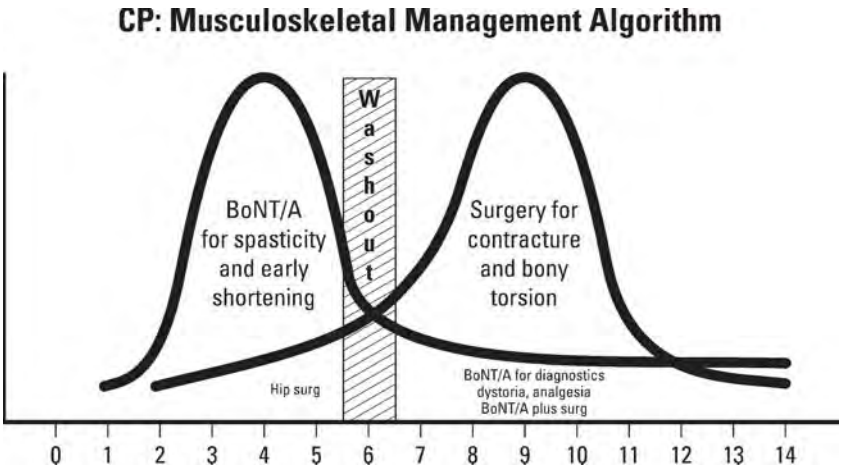



**FIGURE 14-11.** The CP musculoskeletal management algorithm. Neurolytic blocks, botulinum toxin A, and phenol are used in younger children with spasticity. Under the age of 6 years, the only surgery required is preventive hip surgery. Surgery for contracture and bony torsion is most often required between the ages of 6 and 12 years by which stage the role of Botulinum toxin is very limited. (From Gallagher C, Sheedy M, Graham HK. Integrated management with botulinum neurotoxin A. In: Panteliadis CP, ed. *Cerebral palsy. A multidisciplinary approach.* Munchen, Germany: Dustri-Verlag; 2011:213–236, with permission.)

Now children with spastic equinus usually commence BoNT-A therapy aged between 1 and 3 years, in conjunction with physical therapy and the use of appropriate AFOs. They receive injections every 6 to 12 months for several years until gross motor function plateaus, at 4 to 6 years of age. Residual contractures and bony torsion can then be dealt with as SEMLS (96). A program of care utilizing BoNT-A should be viewed as complementary to surgical reconstruction and not as an alternative (96, 97) (Fig. 14-11). Information about dosing, dilution, muscle targeting, and safety has been published elsewhere (98–102).

Injection of BoNT-A for spastic equinus increases the dynamic length of the gastrocsoleus with improvements in ankle dorsiflexion during gait, as determined by the Physician Rating Scale (93, 103). Improvements have been reported in studies using IGA, including kinematics, kinetics, and electromyography (94, 104). This may lead to small but important gains in gross motor function (105–107). The evidence base supporting the use of BoNT-A in CP is quite good as confirmed in several randomized controlled trials and systematic reviews (108, 109). However, the treatment effect is small and short lived. The drug is expensive limiting access and is not approved by the FDA in the management of children with CP in the United States. All current use in the United States is therefore "off label."

**Spastic Equinovarus and Equinovalgus.** Spastic equinovarus is the result of spasticity in the gastrocsoleus, tibialis posterior and/or tibialis anterior (110). In spastic equinovarus, the most effective strategy is to inject the gastrocsoleus and the tibialis posterior (94). Equinovalgus is not usually the result of muscle imbalance but altered biomechanics. It is best managed by injection of the gastrocsoleus and provision of an appropriate AFO (109).

**Injection of the Hamstrings and the Adductor Muscles in Cerebral Palsy.** Spasticity in the hamstring and adductor muscles is prevalent in the severely involved child and may result in scissoring postures and spastic hip displacement (38). Injection of the adductor and hamstring muscles with BoNT-A every 6 months combined with an abduction brace had no appreciable effect on the prevention of hip displacement, in a large RCT (111). The majority of the children required surgical stabilization of their hips either during the study or soon after the study concluded (111).

**Multilevel Injections of Botulinum Neurotoxin A in Cerebral Palsy.** Techniques have been developed for injecting the iliopsoas as part of a multilevel injection protocol for children with spastic diplegia. Multiple target muscles are injected under mask anesthesia and followed by supplemental casting, orthoses, and intensive rehabilitation. Temporary improvements in gait and function have been reported (112, 113). Phenol neurolysis for adductor spasticity can be combined with BoNT-A chemodenervation of the hamstring and calf muscles (90). The principal indication is the younger child with spastic hip displacement and walking difficulties (39, 84).

**Botulinum Neurotoxin A in the Upper Limb in Cerebral Palsy.** In typical hemiplegic posturing the most common target muscles are biceps, brachialis, pronator teres, flexor carpi ulnaris, flexor carpi radialis, and adductor pollicis. The long finger flexors should usually be avoided to prevent weakening grip strength, except when the aim is improved palmar hygiene (92, 114, 115). Precise targeting with electrical stimulation, electromyography, or ultrasound is mandatory (101, 102). Palpation is inaccurate. Upper limb dose guides have been published elsewhere (92, 98, 99, 115). Botox injections in the upper limb results in a reduction in muscle tone but robust evidence for improvements in function is limited, in studies that employed valid and reliable functional outcome measures (116–121).

## ADVERSE EVENTS AND BOTULINUM NEUROTOXIN THERAPY IN CEREBRAL PALSY

BoNT-A is generally safe in children with CP (93, 94, 98, 99). Most adverse events are localized, minor, and self-limiting. Systemic side effects including temporary incontinence and dysphagia have been reported (100). Dysphagia, aspiration, and chest infection are the most serious complications after injection of BoNT-A and if unrecognized or inadequately treated could lead to death from asphyxia (100, 122).

## BOTULINUM NEUROTOXIN A AS AN ANALGESIC AGENT IN CEREBRAL PALSY

BoNT-A can be used to treat muscle spasm following operative procedures, such as adductor-release surgery (123). Injection of BoNT-A can be useful for short-term relief of pain associated with hip displacement (124). Target muscles include the hip adductors, medial hamstrings, and hip flexors. Pain relief is associated with a decrease in spastic adduction and scissoring postures (123, 124). It is not clear if short-term pain relief can be sustained by repeat injections and the need for salvage surgery avoided. Some children with neglected hip displacement have limited life expectancy and may not survive salvage surgery. BoNT-A may provide useful palliation in such circumstances (124). Better still is to prevent painful hip displacement.

## SAGITTAL GAIT PATTERNS: SPASTIC HEMIPLEGIA—WINTERS, GAGE, AND HICKS

In 1987, a four-group classification of sagittal gait patterns in spastic hemiplegia was developed by Winters et al. (125). Their four-group classification has been extensively used by

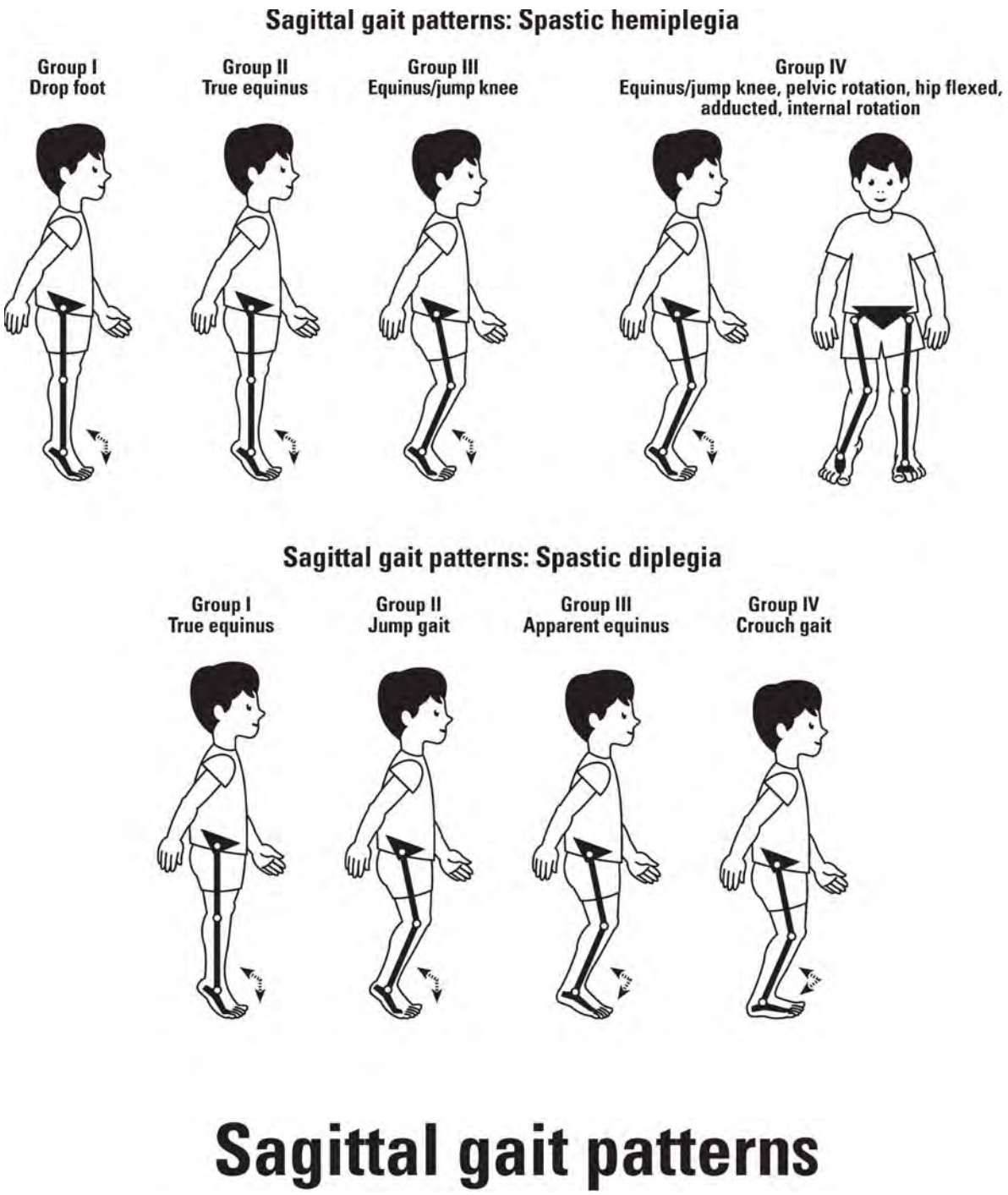



**FIGURE 14-12.** Sagittal gait patterns in spastic hemiplegia (based on the classification by Winters, Gage, and Hicks (127)). Sagittal gait patterns in spastic diplegia (based on Rodda and Graham (97)).

clinicians as a template for clinical management including prescription of orthoses, spasticity management by injection of BoNT-A, and musculoskeletal surgery (Fig. 14-12).

**Type I Hemiplegia.** In type I hemiplegia, there is a drop foot in the swing phase of gait due to loss of selective motor control in tibialis anterior. There is no contracture of the gastrocsoleus and second rocker is relatively normal. Neither spasticity management nor musculoskeletal surgery is necessary. Gait and function can be improved by the use of an AFO, usually a leaf spring AFO or a hinged AFO.

**Type II Hemiplegia.** In type II hemiplegia, there is spasticity in the gastrocsoleus that gradually becomes fixed resulting in a contracture and equinus gait. First and second rockers at the ankle are disrupted and there may be proximal deviations related to the excessive plantar flexion at the ankle, but these are not primary gait deviations. Management of type II hemiplegia

requires correction of fixed contracture in the gastrocsoleus (to correct second rocker) and provision of an AFO (to provide heel strike and first rocker in stance as well as swing phase clearance and appropriate prepositioning of the foot during preswing).

**Type III Hemiplegia.** In type III hemiplegia, there is a contracture of the gastrocsoleus at the ankle and knee involvement with co-contraction of the hamstrings and rectus femoris. Children in this transitional group may benefit from lengthening of the medial hamstrings and rectus femoris transfer (see Figs. 14-29 to 14-33), in addition to gastrocsoleus lengthening.

**Type IV Hemiplegia.** In type IV hemiplegia, pathology is present at all three joints of the lower extremity. In the sagittal plane, in addition to ankle equinus and knee stiffness, there is incomplete hip extension. In the coronal plane at the hip, there is excessive adduction and in the transverse plane, excessive internal rotation. Hip dysplasia is common and often presents late. In addition to the treatments outlined above for type III, correction of type IV hemiplegic gait includes lengthening of both the adductor longus and the psoas over the brim (POTB) of the pelvis as well as a proximal femoral derotation osteotomy.

Not all children with hemiplegia fit neatly into one of the four groups described (126). Nonetheless, this is an entirely logical and very useful way of classifying hemiplegic gait with direct relevance to clinical management (127).

## SAGITTAL GAIT PATTERNS—SPASTIC DIPLEGIA

Knee patterns in spastic diplegia have been classified as recurvatum knee, jump knee, stiff knee, and crouch (128). The knee classification has been extended to the sagittal plane as true equinus, jump gait, apparent equinus, and crouch gait (97) (Fig. 14-12).

**True Equinus.** True equinus is characterized by walking on tip toe with extended hips and knees, as is commonly seen in younger children with spastic diplegia when they first learn to walk. The plantarflexion-knee extension couple is overactive and the ground reaction force (GRF) is in front of the knee throughout stance phase. True equinus can be managed in the younger child by injections of BoNT-A to the gastrocsoleus and the provision of hinged AFOs. By the time children develop fixed contractures and require surgery, true equinus is rare. When it persists, there are usually occult contractures of the hamstrings and iliopsoas. Single-level surgery (gastrocsoleus lengthening) is almost never the correct strategy, no matter how tempting it may appear on observational gait analysis.

**Jump Gait.** Jump gait is characterized by equinus at the ankle associated with incomplete extension at the knee and hip. In the original description by Sutherland and Davids, the jump knee pattern is characterized by excessive flexion at initial contact with rapid extension in later stance to near-normal range (128). In the pattern described by Rodda and Graham, jump gait encompasses this pattern as well as patterns in which knee extension is more severely compromised and in which there is incomplete extension at the hip (97, 127). This is the most common pattern in the preadolescent with spastic diplegia. Many children benefit from SEMLS.

**Apparent Equinus.** Many children with spastic diplegia who walk on their toes, never achieving heel contact, have an ankle range of motion within the normal range. Such children are at risk of inappropriate management with injections of BoNT-A to the gastrocsoleus or even worse, lengthening of the gastrocsoleus. The important contractures are proximal at the level of the knee and hip. The recognition of "apparent equinus" in contradistinction to "true equinus" is very important to avoid inappropriate lengthening and weakening of the gastrocsoleus with further deterioration in gait and functioning. IGA is very helpful in differentiating "apparent equinus" from "true" equinus. Apparent equinus pattern is often transitional. With further growth and progression of lever arm deformities, the majority of children will eventually develop "crouch gait."

**Crouch Gait.** Crouch gait is characterized by excessive knee flexion in stance, incomplete extension at the hip, and excessive ankle dorsiflexion. Knee stiffness in swing is common. The soleus is excessively long and usually weak. This is a very common gait pattern in adolescence and is often the result of natural history, accelerated by lengthening of the gastrocsoleus, especially percutaneous lengthening of the Achilles tendons. In recent reviews of crouch gait, the majority of children had lengthening of the Achilles tendons in childhood (59). A key feature of crouch gait is that the majority of MTUs are excessively long. This is by definition true for all of the one joint muscles such as soleus, quadriceps, and gluteus maximus and often for the two joint hamstrings. The only consistent contractures are of the iliopsoas. In crouch gait, the hamstrings are short only in patients with a posterior pelvic tilt. When the pelvis is in the neural range, the hamstrings are of normal length and when the pelvis is anteriorly tilted, the hamstrings are excessively long. Without the use of IGA and the plotting of muscle lengths, it is very difficult to appreciate these findings. Consequently, the majority of children with crouch gait are managed by excessive hamstring lengthening to improve knee extension when in fact the hamstrings are of normal length or excessively long. Such surgery results in increased anterior pelvic tilt that in the long term may bring its own set of problems with low back pain and increased risks of spondylolisthesis and spondylolysis (59).

## BIOMECHANICAL STUDIES RELEVANT TO ORTHOPAEDIC SURGERY IN SPASTIC DIPLEGIA

In a study of muscle excursion and cross-sectional area, it was demonstrated that the gastrocsoleus and ankle dorsiflexors have such different physical characteristics that they cannot

be considered to be "in balance," in either normal subjects or in children with CP (129). The plantarflexors are six times as strong as the dorsiflexors. The plantarflexors of the ankle must be balanced against the GRF not the dorsiflexors. Lifting the foot and ankle during swing phase (dorsiflexor function) requires a very small muscle moment. Push-off in terminal stance (plantarflexor function) requires a large muscle moment. The concept of muscle balance should be redefined as a requirement for balance between the three anatomical levels, hip, knee and ankle, in the sagittal plane, not at a single level (129).

Lower limb muscles have different sensitivities to surgical lengthening, related to their gross anatomy and morphology. The soleus is exquisitely sensitive to lengthening, but the iliopsoas and semitendinosus are relatively resistant. A 1-cm lengthening of the soleus reduces its moment-generating ability by 30% and a 2-cm lengthening reduces its moment by 85% (130). A small error in terms of overlengthening the soleus may be disastrous. A 4-cm lengthening of the psoas is required to reduce its moment by 50% (130). The surgical implications are to lengthen the gastrocnemius only, when there is no contracture of the soleus. When the soleus requires lengthening, a precise and stable technique should be used, with careful control of the position postoperatively in a cast. By contrast, it is difficult to overlengthen the psoas. Intramuscular lengthening at the pelvic brim without immobilization postoperatively is safe and effective.

Many children who walk with flexed knee gait have hamstrings that are of normal length. It is the psoas that is shortened and requires lengthening, not the hamstrings. It is easy to do too much hamstring lengthening and not enough psoas lengthening (59, 131, 132).

## MUSCULOSKELETAL MANAGEMENT IN CEREBRAL PALSY BY GMFCS LEVEL: INTRODUCTION

The GMFCS gives an accurate summary of a child's current gross motor function and long-term prognosis (13, 15). It is difficult to frame a logical discussion of musculoskeletal management outside of the context of the GMFCS. Most disagreements in the literature are apparent rather than real because those taking opposing views are often considering a child in a different GMFCS level. The terms mild, moderate, and severe diplegia along with mild, moderate, and severe quadriplegia are not meaningful or useful. Age-appropriate GMFCS descriptors are valid, reliable, stable, and clinically meaningful (47, 49, 50). Recommendations for both gait correction surgery and surgery for hip displacement are much more easily understood when the child's GMFCS level is known. In the following sections, musculoskeletal management will be discussed by GMFCS level, in conjunction with both topographical classification and sagittal gait patterns. Gait correction surgery is an option for many children with CP at GMFCS levels I to III but with differences at each level. SEMLS will be discussed primarily in the GMFCS level II section. Preventive hip surgery will be discussed in the GMFCS level III section, reconstructive hip surgery in the GMFCS level IV section, and salvage surgery in the GMFCS level V section. Obviously, reconstructive surgery may be appropriate at GMFCS levels III to V and salvage surgery is sometimes needed at both GMFCS IV and V but is best avoided.

## GMFCS I

1. **Between 6th and 12th birthday**
   Children walk at home, school, outdoors, and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance, and coordination are limited (13).
2. **Between 12th and 18th birthday**
   Youth walk at home, school, outdoors, and in the community. They are able to climb curbs and stairs without physical assistance or a railing. They perform gross motor skills such as running and jumping, but speed, balance, and coordination are limited (15).
3. **Risk of hip displacement:** Developmental dislocation of the hip occurs at the same rate as in the normally developing population. Spastic hip displacement is not seen (49, 50).
4. **Mean femoral neck anteversion (FNA):** 30 degrees
5. **Mean neck shaft angle (NSA):** 136 degrees
   Children at GMFCS level I participate in physical recreational activities, have a normal life expectancy, few medical comorbidities apart from epilepsy, and good levels of intellectual functioning.

**Movement Disorder.** Children at GMFCS level I usually have mild spasticity that is often distal with little proximal involvement. Some have mild dystonia. The movement disorder is mild and easily managed by injections of BoNT-A. Neither SDR nor ITB are appropriate choices (38, 39, 84).

**Topographical Distribution.** Children at GMFCS level I have either spastic hemiplegia or mild spastic diplegia. In terms of sagittal gait patterns, those with hemiplegia are usually type I or II. Those with diplegia usually have true equinus or mild jump gait.

**Musculoskeletal Impairments.** Hip disease and scoliosis are rare and occur with a similar prevalence as would be expected in typically developing children. If hip displacement is detected, it is usually a developmental dysplasia. If a spinal curvature develops, it will be an adolescent idiopathic-type curve.

### Musculoskeletal Management GMFCS Level I: Spastic Hemiplegia

**Lower Limb Surgery.** Children with type I hemiplegia have a drop foot in the swing phase of gait. They may benefit from a leaf spring or a hinged AFO. Orthopaedic surgery is not required (125).

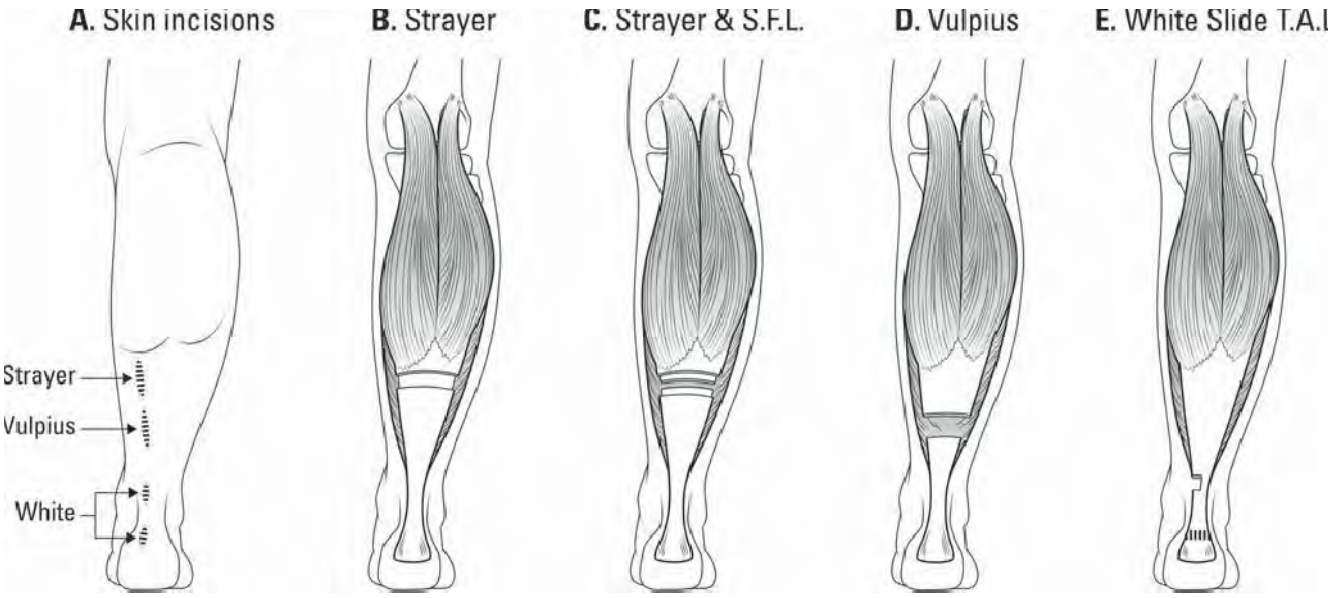



**FIGURE 14-13.** Surgery for equinus deformity in cerebral palsy. **A:** The location of the skin incisions is usually posteromedial and with accurate identification of the level can be kept small, typically 2 to 3 cm for the Strayer and Vulpius procedures. The White slide TAL may be performed percutaneously, but our preference is for two small posteromedial incisions each 1.5 cm long. We prefer to see the tendon and avoid accidental complete tenotomy. **B:** The Strayer procedure is a distal gastrocnemius recession and lengthens only the gastrocnemius portion of the gastrocsoleus. It is the most useful procedure in children with diplegia and "gastrocnemius equinus." **C:** The Strayer procedure can be combined with soleal fascial lengthening (SFL). This results in a lengthening of both gastrocnemius and soleus but by different amounts, a 2:1 ratio, that is, twice as much lengthening of the gastrocnemius as for the soleus. This is the most useful procedure in children with spastic diplegia who have a moderate contracture of the gastrocnemius and a less severe contracture of the soleus. **D:** The Vulpius procedure is a Zone 2 recession of the gastrocsoleus. The shape of the cut can be the familiar inverted V. However, a simple transverse cut requires a smaller skin incision and is just as effective. This is a useful procedure in children with hemiplegia who have a moderate degree of fixed contracture affecting both the gastrocnemius and the soleus. **E:** Slide lengthening of the Achilles tendon may be performed by double hemisection as described by White. This is the most useful procedure in children with hemiplegia who have a severe contracture affecting the gastrocnemius and the soleus. For further discussion please see reference 133.

Children with type II hemiplegia develop equinus contractures and may benefit from lengthening of the gastrocsoleus. Surgery can usually be deferred until age 4 to 6 years by the use of an AFO combined with injections of BoNT-A and physical therapy. The choice of lengthening procedure is based on a careful Silfverskiold test to determine the amount of contracture in the gastrocnemius and soleus, respectively (133). There are four main options as illustrated in Figure 14-13 (see Figs. 14-14 and 14-15).

**Upper Limb Surgery in Spastic Hemiplegia.** Many children with hemiplegia are not candidates for upper limb surgery. Occupational therapy and physiotherapy have small treatment effects alone but are essential adjuncts to surgical management. As with lower limb surgery, there is a move toward detailed preoperative analysis, the identification of component deformities and muscle imbalances, and the development of a detailed single-event, multilevel surgical plan which is followed by casting, splinting, and rehabilitation (134, 135) (Figs. 14-16 to 14-18).

The typical upper limb deformities in CP include adduction and internal rotation of the shoulder, pronation of the forearm, wrist flexion and ulnar deviation, finger flexion, and thumb in palm (134).

**Functional Impairment.** Functional deficits include problems with reaching, grasping, releasing, and manipulation and should be carefully evaluated in each child. The appearance of the limb is also a concern to children and caregivers. Gross motor function in the upper limbs in children with CP is classified using the Manual Ability Classification System (MACS), the upper limb equivalent of the GMFCS (136).

**Principles of Management.** Children with spastic hemiplegia function at a high level and may require interventions aimed at developing sophisticated fine motor control for bimanual hand activities. Improving cosmesis by reducing flexion posturing of the elbow during running and flexion of the wrist with grasping activities are important goals. Simpler hand activities such as grasping and releasing assistive walking devices are the main objectives of treatment in children with more severe involvement. In those still more severely involved, ease of dressing and hygiene are the primary reasons for correcting upper limb deformities (134).

**Assessment of the Upper Limb in CP.** Detailed history, standardized physical examination, and radiographs are the

*Text continued on page 508*

## The Strayer Distal Gastrocnemius Recession (Fig. 14-14)

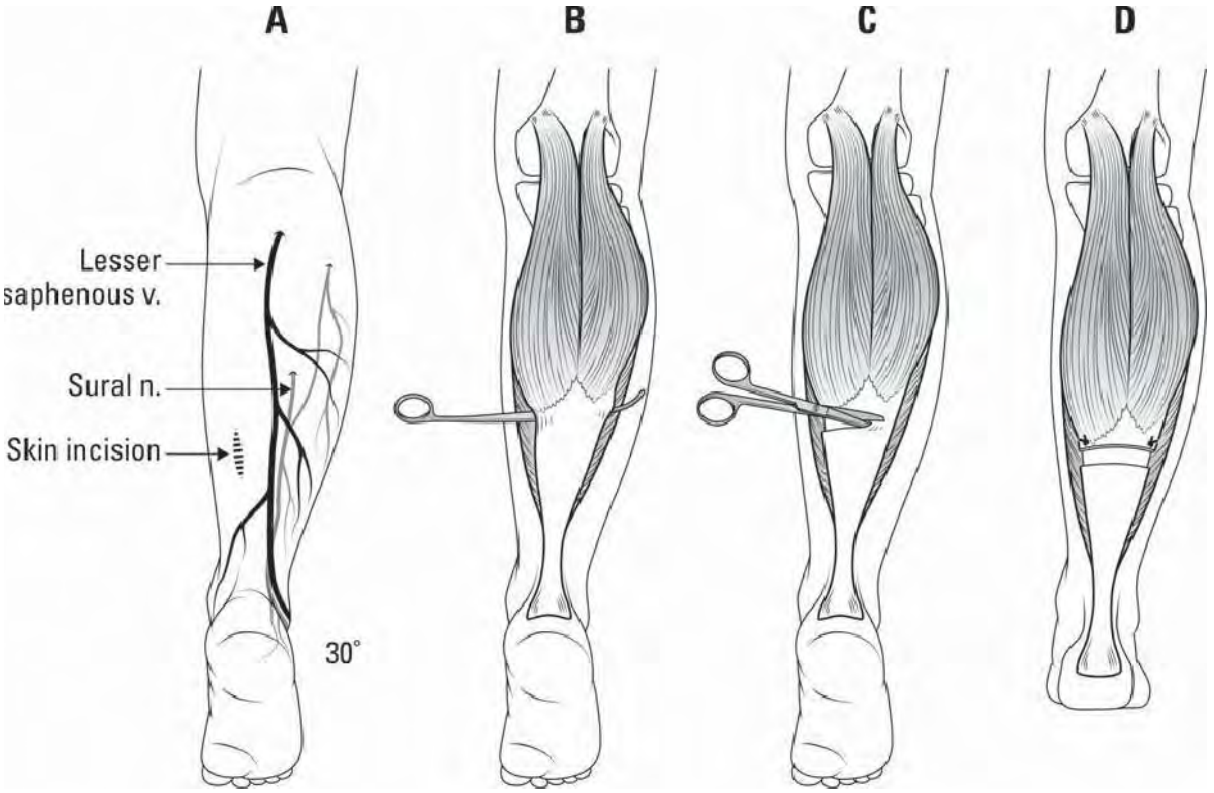



**FIGURE 14-14. The Strayer Distal Gastrocnemius Recession. A:** For all methods of gastrocsoleus lengthening, the superficial structures at risk of injury include the sural nerve and the lesser saphenous vein. **B:** The Strayer distal gastrocnemius recession is sometimes criticized because of the length of the skin incision required and subsequent scarring. However, with accurate identification of the interval between the gastrocnemius aponeurosis and the soleal fascia, the incision can be kept very small, typically 2 to 3 cm long. The termination of the distal medial belly of the gastrocnemius can be determined by palpation and the skin incision should be centered on this point. For those who are less experienced, this interval can be determined accurately by preoperative ultrasound and marking the skin with a surgical pen. The interval between the bellies of the gastrocnemius and the underlying soleal fascia can be developed by a combination of blunt dissection with the surgeon's finger and the use of a blunt dissector. A blunt dissector can be passed from medial to lateral through this interval and the lesser saphenous vein and sural nerve protected by retraction. **C:** Following mobilization of the gastrocnemius aponeurosis from the underlying soleal fascia, the aponeurosis is divided transversely from medial to lateral, using dissecting scissors. **D:** Following dorsiflexion of the foot, a gap opens up in the aponeurosis (without lengthening of the underlying soleus). With the knee in extension and the foot plantargrade, the gastrocnemius aponeurosis can be sutured to the underlying soleal fascia by two sutures to prevent excessive proximal retraction. We are unsure about the need for this step. We usually omit the suture in the interest of a shorter incision. Dorsiflexion of the foot to five degrees is sufficient for the majority of children with spastic diplegia because "a little equinus is better than calcaneus."

## Strayer Distal Gastrocnemius Recession Combined with Soleal Fascial Lengthening (Fig. 14-15)

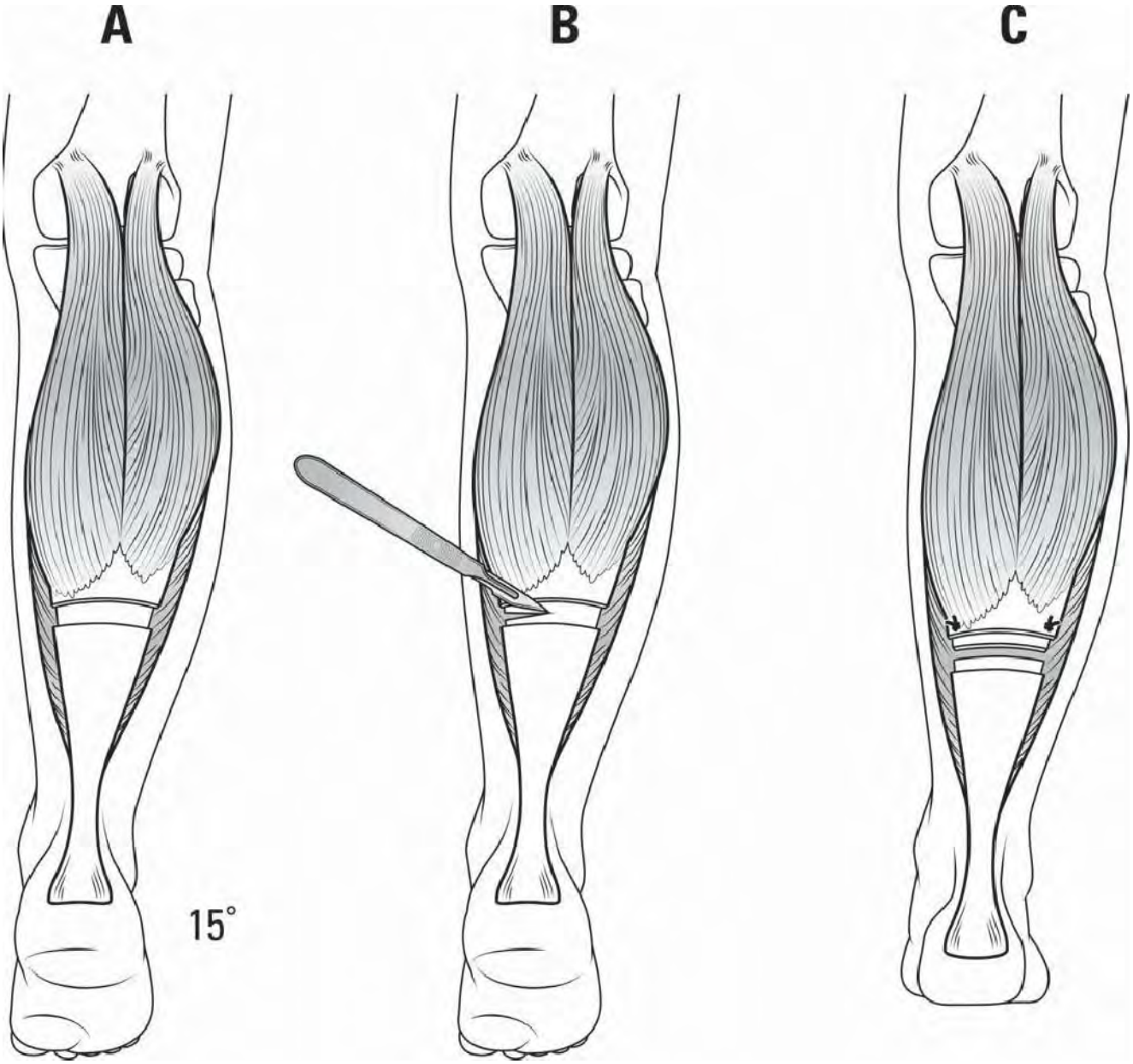



**FIGURE 14-15. Strayer Distal Gastrocnemius Recession Combined with Soleal Fascial Lengthening. A:** In some children, the Strayer procedure is insufficient to gain a plantargrade position of the foot. This can be confirmed by an intraoperative Silfverskiold test. When this is the case, lengthening of the soleal fascia should be considered. **B:** Following division of the gastrocnemius aponeurosis as previously described, the underlying soleal fascia can be divided transversely by sharp dissection using a scalpel. The midline raphé should also be identified and divided in the midline of the underlying soleal muscle. **C:** Following further dorsiflexion of the foot, the gastrocnemius can be sutured to the underlying soleal fascia as indicated.

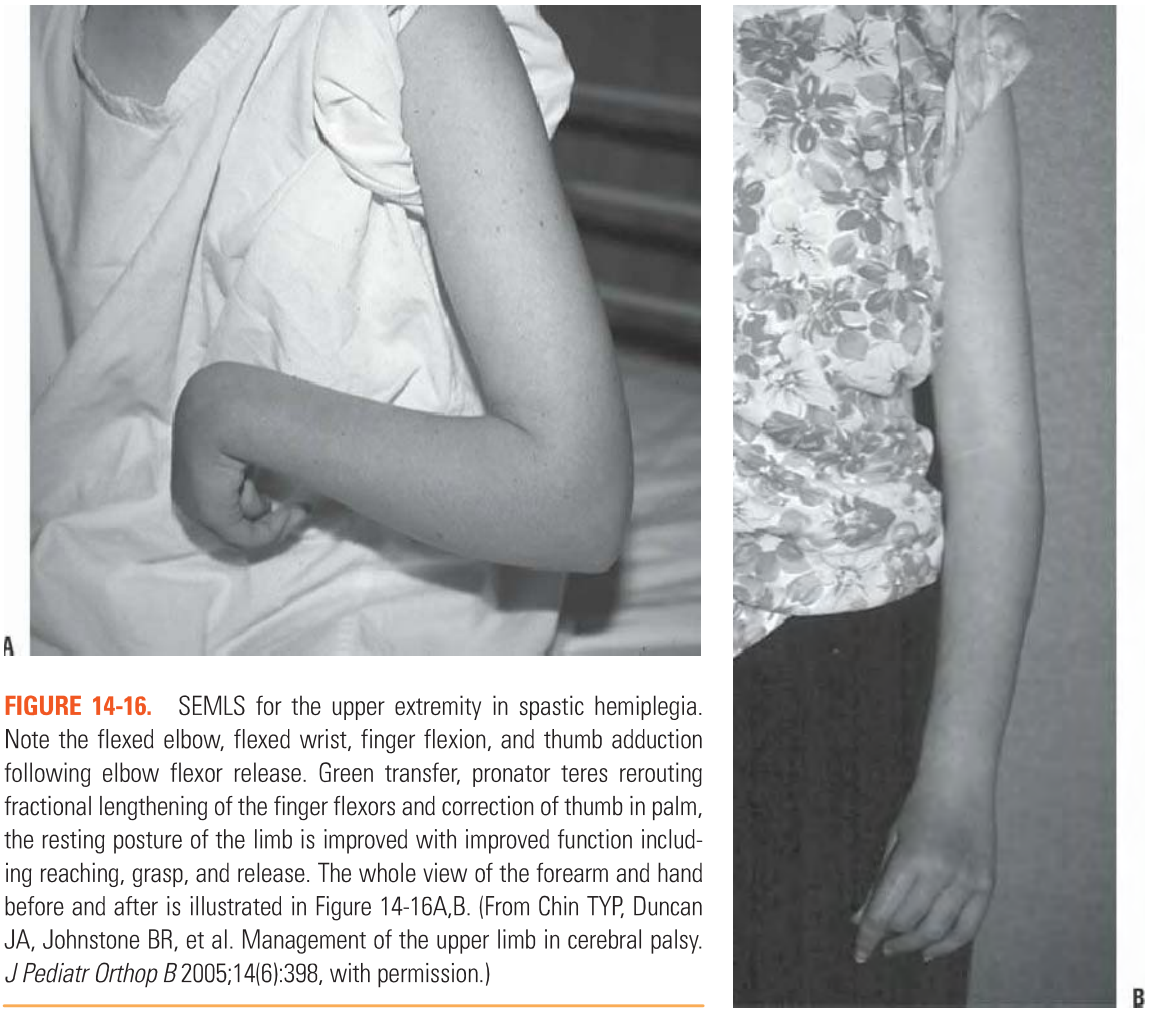


**FIGURE 14-16.** SEMLS for the upper extremity in spastic hemiplegia. Note the flexed elbow, flexed wrist, finger flexion, and thumb adduction following elbow flexor release. Green transfer, pronator teres rerouting fractional lengthening of the finger flexors and correction of thumb in palm, the resting posture of the limb is improved with improved function including reaching, grasp, and release. The whole view of the forearm and hand before and after is illustrated in Figure 14-16A,B. (From Chin TYP, Duncan JA, Johnstone BR, et al. Management of the upper limb in cerebral palsy. *J Pediatr Orthop B* 2005;14(6):398, with permission.)

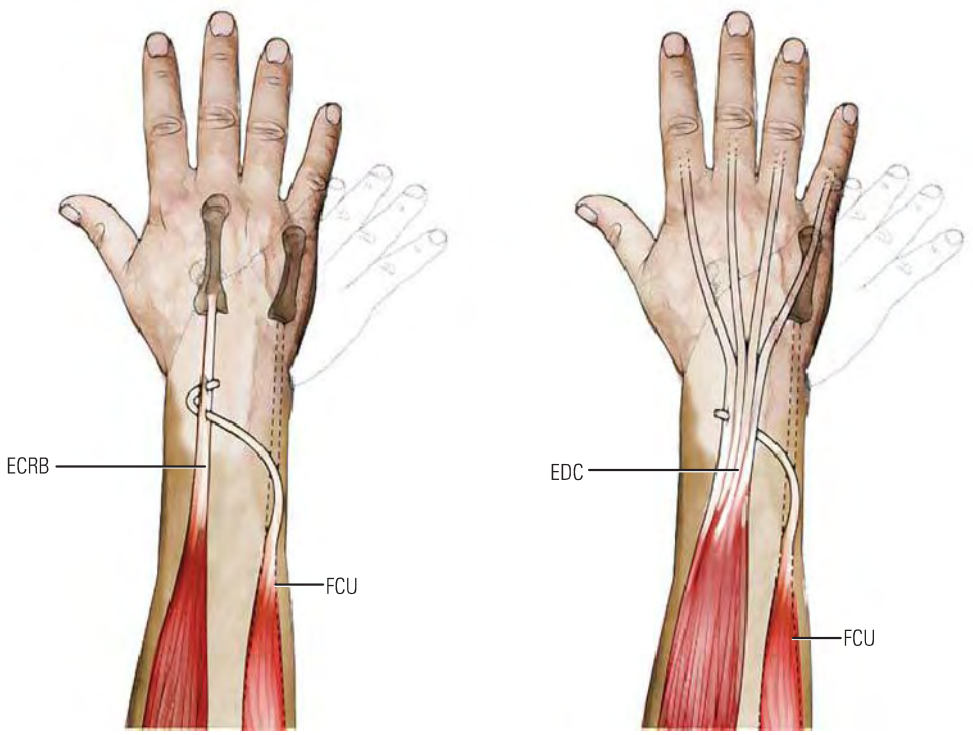



**FIGURE 14-17.** The Green transfer is the single most useful tendon transfer in the hemiplegic upper limb to improve function. The flexor carpi ulnarus (FCU) is the strongest wrist flexor and ulnar deviator of the wrist. When transferred around the subcutaneous border of the ulna to the extensor carpi radialis brevis (ECRB), wrist extension is strengthened and the tendency to ulnar deviation corrected. The line of the transfer also strengthens supination. An alternative site for the transfer is to the extensor digitorum communis (EDC).

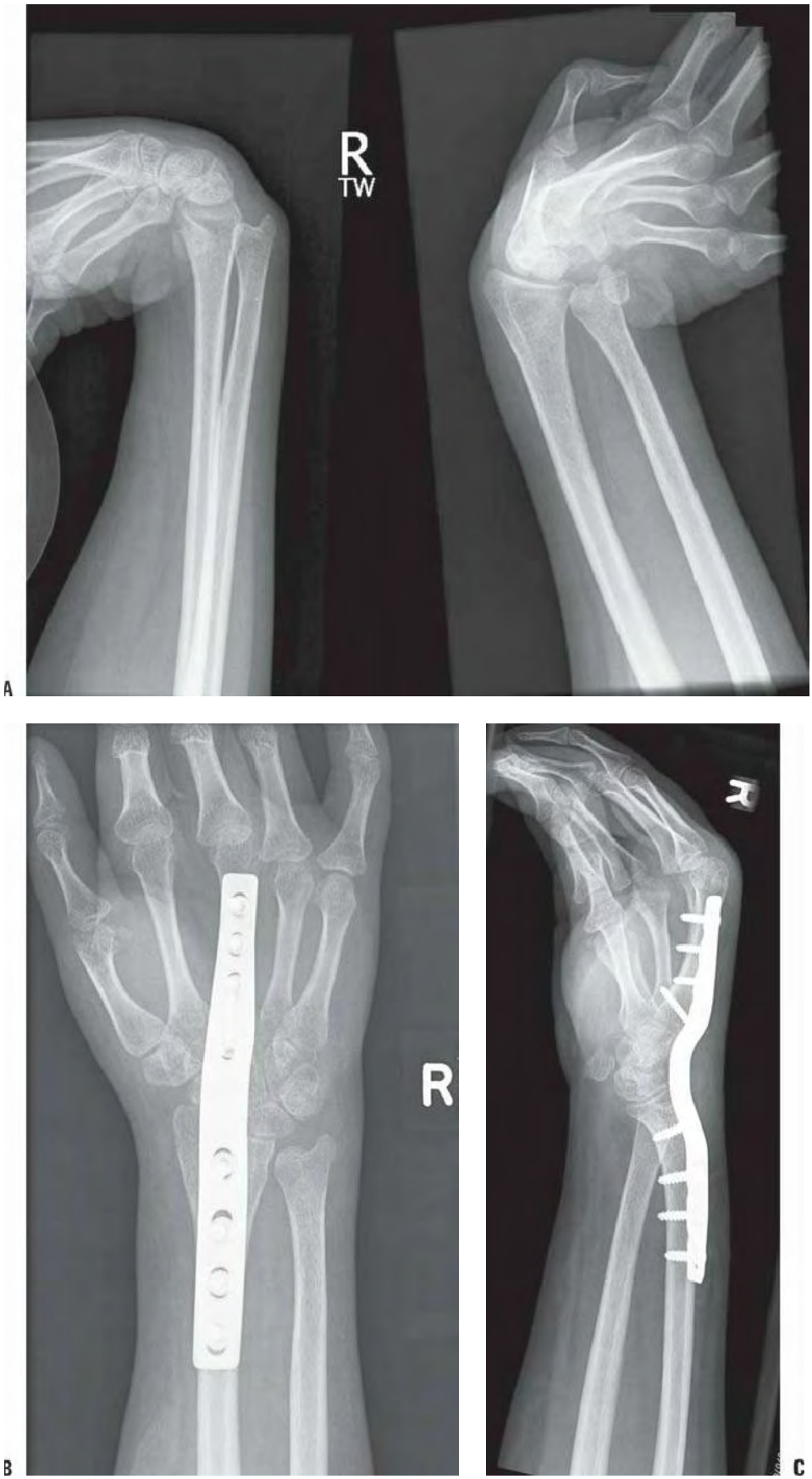



**FIGURE 14-18.** **A–C:** Severe wrist flexion and ulnar deviation in an adolescent with spastic hemiplegia before and after wrist fusion using a custom, contoured fixation plate. Fusion was combined with superficialis-to-profundus transfer of the finger flexors, and plication of the extensor tendons. Improvements in comfort and cosmesis were obtained with no change in function.

cornerstones of upper limb assessment. The active and passive range of motion, presence of spasticity, dystonia, contractures, selective motor control, muscle strength, and sensory deficits should be recorded (134). The child's functional use of the affected hand may be quantified according to the House Classification of Upper Extremity Functional Use (136). This nine-level classification is useful to establish baseline function, communicate functional levels and goals to parents and other clinicians, and monitor progress of treatment.

Objective evaluation of upper limb function using standardized, validated instruments such as the Melbourne Unilateral Upper Limb Assessment (Melbourne Assessment) or Quality of Upper Extremity Skills Test (QUEST) is strongly recommended to document baseline function and also assess changes following treatment (119, 120). Both scales have established reliability and validity. Video recordings of postural and functional assessments are very useful, especially when combined with an objective scoring system such as Shriner's Hospital for Children Upper Extremity Evaluation (SHUEE) (137). Kinematic analysis is developing rapidly but is not yet standardized or widely available.

**Principles of Surgical Management.** Almost any fixed contracture may benefit from lengthening but which procedures to use and when, requires experience and judgment (135, 138). Tendon transfers can be utilized to improve hand or wrist function. BoNT-A may also be used together with surgery as a spasticity-reducing measure or to aid with perioperative pain relief (139). Surgical results are most predictable in spastic movement disorders and are unpredictable in dystonia. Realistic expectations are vital because surgery cannot restore normal hand function or appearance (134, 135).

**Elbow.** Dynamic flexion contracture of the elbow is frequently seen in hemiplegic CP and is particularly marked as an associated movement during running. A transverse incision across the elbow crease can provide adequate access to perform Z lengthening of the biceps tendon, as well as a fractional lengthening of the brachialis (138) (Fig. 14-16)

**Forearm Pronation.** The pronator teres is the first MTU to develop a contracture in the hemiplegic upper limb. A fibrotic pronator teres can be simply released but if it has a reasonable excursion, it can be rerouted to act as a supinator (140). The forearm is immobilized in maximum passive supination with the elbow flexed to 90 degrees. Forearm pronation can also be improved by transferring flexor carpi ulnarus (FCU) to extensor carpi radialis brevis (ECRB) (Green transfer). By virtue of the dorsoulnar course of the transferred tendon, FCU becomes a secondary supinator in addition to its new role as a wrist extensor (138).

**Wrist.** The majority of children with hemiplegic CP have wrist flexion deformities. The two most useful procedures for wrist flexion deformities are the Green transfer and arthrodesis but for different indications. Children who have a functional hand with constant flexed wrist posturing, secondary to out-of-phase activity in FCU, may be candidates for the Green transfer (Fig. 14-17) (135, 138). Activation of FCU can be assessed by palpating the FCU tendon as patients open and close their fingers and confirmed using dynamic electromyography. Some children have poor finger extension and an FCU working in phase with finger extensors. These children may benefit from transfer of the FCU to the extensor digitorum communis (EDC). Contractures of FCR, Palmaris longus, and the long finger flexors must be addressed at the same time (134, 135, 138).

Adolescents with severe wrist flexion contractures and limited function may appreciate the cosmetic gains and improvements in palmar hygiene from arthrodesis of the wrist, combined with soft-tissue releases (Fig. 14-18). A dorsal wrist fusion plate provides stable fixation, permits early mobilization, and has good outcomes in terms of fusion rates and deformity correction (141). The soft tissues should be rebalanced by an extensive release of all contracted MTUs and plication of the redundant wrist and finger extensors. Improvements in cosmesis are substantial because the atrophic limb appears to be longer following correction of the severe wrist flexion deformity as well as partial correction of the digital contractures. There may be minor improvements in "helper hand" functions. In appropriately selected cases, satisfaction with the procedure is very high (134, 141).

**Fingers.** When wrist flexion is corrected, as described above, occult spastic contractures in the fingers and thumb may be unmasked. There are three main options. Mild spastic contractures in the long flexors may respond to Botox, combined with casting (139). Fractional lengthening at the musculotendinous junctions of FDS and FDP, especially when combined with injections of BoNT-A and splinting, is effective and preserves function (138). In severe contractures, when functional goals are more limited, FDS to FDP transfer may be performed (134). The FDS tendons are divided distally, close to the wrist. The FDP tendons are exposed and are divided more proximally toward the musculotendinous junction. The proximal FDS MTUs are then repaired en masse to the distal FDP tendons to provide a degree of tension. This works well in conjunction with wrist arthrodesis.

Release of the finger flexors may unmask swan neck deformities, particularly when the intrinsic muscles of the hand are spastic. An uncorrected wrist flexion posture has a tenodesis effect on the extensors, which is expressed by deformities at the PIP joint. In addition to rebalancing the flexor and extensor tension across the PIP joints, correction of unstable swan neck deformities is performed where there is incompetence of the volar plates (142, 143).

**Thumb in Palm.** The "thumb-in-palm" deformity is variable and may include adduction of the first metacarpal, flexion at the metacarpophalangeal (MCP) joint, and either flexion or extension at the IP joint (143). Many children have hyperextendable MCP joints and, with adduction of the metacarpal, this leads to a swan neck–type deformity of the thumb. This is managed with a release of adductor pollicis, and the flexor pollicis brevis from the flexor retinaculum. Release of the first dorsal interosseous and the overlying fascia is also frequently

required. A contracted first web space may be corrected by Z-plasty or "square flap" (143). Instability of the thumb MCP joint can be corrected by arthrodesis of the radial sesamoid of the thumb to the underlying metacarpal (143). Rerouting the EPL to the volar side of the wrist makes it an abductor, rather than an adductor of the thumb. EPL function can be augmented by transferring palmaris longus. An opponensplasty using the FDS to the ring or middle fingers can provide functional correction of a thumb-in-palm deformity.

**Surgical Results.** Several large retrospective studies have reported improvements in House scale, grasp and release, self-care, grip strength, and dexterity (132, 135, 144, 145). Satisfaction with both functional and cosmetic outcomes by both children and care givers is generally high (146).

**Spastic Diplegia—GMFCS Level I.** Children with spastic diplegia at GMFCS level I usually have a true equinus gait pattern with mild spasticity in the gastrocsoleus and little proximal involvement. The spasticity is typically too mild and too focal to require SDR. It responds well to injections of BoNT-A and the provision of an AFO. Some children develop a mild contracture, usually involving only the gastrocnemius and not the soleus. This can be managed by distal gastrocnemius recession as described by Strayer. Careful assessment by IGA is essential to identify proximal involvement that require simultaneous correction. Isolated, single-level surgery for equinus is rarely indicated and is associated with a 40% risk of severe crouch gait, in long-term follow-up (95). The correction of proximal gait deviations will be discussed in the section on children at GMFCS level II.

## GMFCS II

1. **Between 6th and 12th birthday**
   Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas, or confined spaces. Children may walk with physical assistance, a handheld mobility device or use wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping (13).
2. **Between 12th and 18th birthday**
   Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work, they may require a handheld mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances (15).
3. **Risk of hip displacement (MP > 30%):** 15%
4. **Mean femoral Neck Anteversion (FNA):** 36 degrees
5. **Mean Neck Shaft Angle (NSA):** 141 degrees (49, 50)
   The majority of children at GMFCS level II have either a type IV hemiplegia or a mild spastic diplegia.

**Type IV Hemiplegia.** In type IV hemiplegia, there is involvement of the entire lower limb. The usual pattern is equinus (equinovarus or equinovalgus) at the ankle; a stiff flexed knee; a hip that is internally rotated, adducted, and flexed; and a pelvis that is retracted (125, 147, 148). The lower limb is usually but not always spastic. The upper limb often has mixed spasticity and dystonia. In addition to increased FNA, there may be external tibial torsion (ETT) resulting in "malignant malalignment." The foot progression angle may be normal, but there is internal rotation of the femur and external rotation of the tibia. Complete correction will usually require an external rotation osteotomy of the femur and an internal rotation osteotomy at the supramalleolar level of the tibia and the fibula. Long-term reliance on the contralateral "sound" leg for push off may result in excessive ETT. It is difficult to evaluate "sound side" ETT without IGA, but it rarely requires correction.

Unilateral multilevel surgery is usually required between the ages of 6 and 10 years (147–150). It is important to note that type IV hemiplegia is associated with progressive hip displacement in a significant number of children. In the initial phases, this is clinically silent, so the hips should always have radiologic evaluation. Progressive subluxation of the hip is an indication to proceed with unilateral multilevel surgery in which stabilization of the hip and correction of the limb deformities is combined. IGA is essential in type IV hemiplegia because of the number of gait deviations and the need to differentiate between primary deviations, secondary compensations, and tertiary coping mechanisms. Shortening of both the leg and the shank is often significant (148). Clinical and CT measurement of limb segment lengths is strongly advised as well as periodic assessment of bone age. A number of children with hemiplegia benefit from contralateral epiphysiodesis to reduce limb-length discrepancy. In these children, bone age is often well ahead of chronologic age. Unilateral SEMLS in type IV hemiplegia can result in correction of hip displacement, improvements in lower limb alignment, correction of gait dysfunction, and significant improvements in both the efficiency and cosmesis of gait (147). Because of unilateral surgery and the intact lower limb, these children rehabilitate quickly and relatively easily.

**Spastic Equinovarus at GMFCS II: Differences between Diplegia and Hemiplegia.** Spastic equinovarus is much more common in hemiplegia than in diplegia. Symptoms may include pain, tripping, brace intolerance, and callosities over the lateral border of the foot (110, 133). In diplegia, varus may be more apparent than real because of excessive FNA and "rollover varus." In diplegia varus is usually mild, flexible, and more prone to overcorrection into valgus than in hemiplegia. In hemiplegia, varus is often more severe, more stiff, and more likely to progress or relapse than in children with diplegia.

Evaluation should include IGA including dynamic EMG, pedobarography, and standardized radiographs in the weight-bearing position (67, 110).

There are many options for the management of spastic equinovarus in CP (133). There are few comparative studies and no clinical trials with high levels of evidence have been

| CLINICAL PRESENTATION | SUGGESTED MANAGEMENT (Fig. 14-19) |
|---|---|
| Mild, dynamic varus in the younger child | Inject GS and TP with BoNT-A + AFO (84) |
| Mild to moderate flexible varus: diplegia | IMT TP + GR + AFO + SEMLS (151) |
| Moderate, flexible varus: hemiplegia | IMT TP or SPOTT + GSR+ AFO (151–153) |
| Moderate to severe flexible varus hemiplegia | IMT TP + SPLATT + GSR + AFO (151,154,155) |
| Moderate fixed varus | Soft Tissue Balancing + cal. osteotomy/shorten lateral column (133) |
| Severe fixed varus | Soft tissue balancing + triple arthrodesis (133, 156) |

IMT, intramuscular tenotomy; GR, gastrocnemius recession; GSR, gastrocsoleus recession; SPOTT, split posterior tibialis tendon transfer; SPLATT, split anterior tibialis tendon transfer.

published. As with many management issues in CP, the stage of musculoskeletal pathology is important to determine and some appreciation of surgical "dose" is helpful.

In diplegia, intramuscular tenotomy of tibialis posterior, combined with correction of FNA, as part of SEMLS gives good results (157). In hemiplegia, equinovarus deformities are more variable in severity and more resistant to surgical correction. In younger children with documented overactivity in tibialis posterior, both intramuscular recession and SPOTT transfer are good options (110, 133, 150–152). Ideally this should be undertaken before deformities become fixed, avoiding the need for bony surgery. In children with documented overactivity in both tibialis anterior and tibialis posterior, a combination of SPLATT transfer and intramuscular tenotomy of tibialis posterior gives good long-term results (110, 133). It is easy to overestimate and overtreat the equinus component of the equinovarus deformity. An Achilles tendon lengthening combined with a tibialis posterior lengthening may result in excessive weakening of plantarflexion, overcorrection, and poor push off. A careful examination under anesthesia will confirm that a gastrocnemius or gastrocsoleus recession is all that is required for equinus correction, in most equinovarus feet.

In children with diplegia, overcorrection to valgus is common. In children with hemiplegia, relapse to recurrent equinovarus is common (133). Postoperative bracing with an AFO may be helpful. Bony surgery may be required for fixed deformities and for some recurrent deformities but must always be combined with soft-tissue balancing. A lateral closing wedge osteotomy of the calcaneum or heel shift is useful for fixed heel varus. Calcaneocuboid shortening/fusion is useful to correct adductus and supination. Triple arthrodesis should be avoided because it is unsatisfactory end-stage, salvage

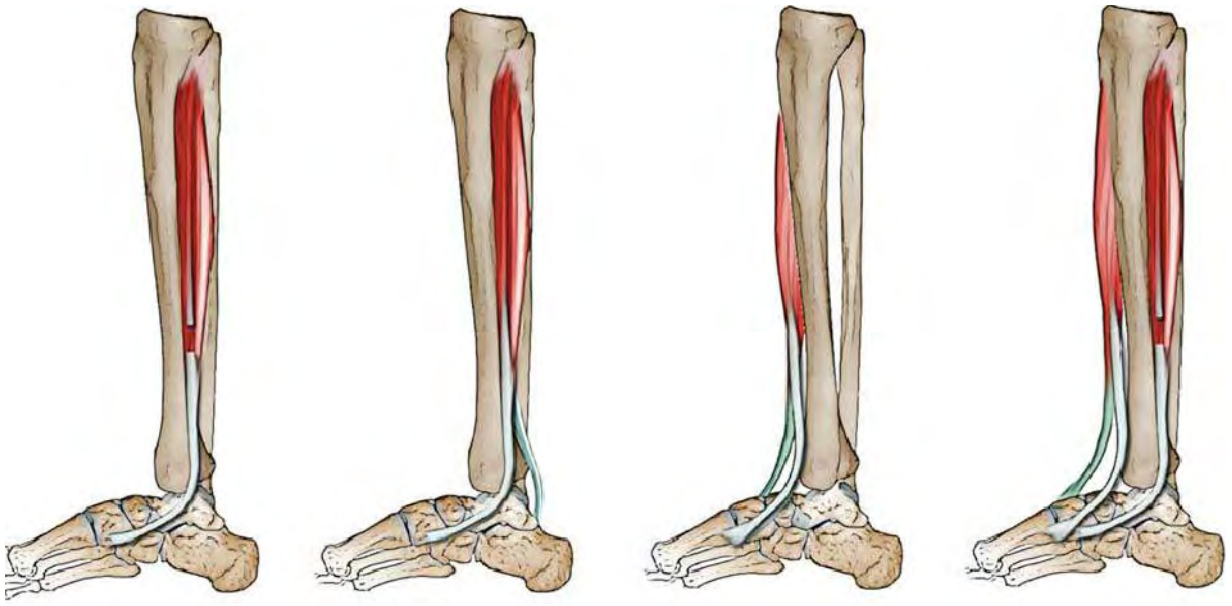



**FIGURE 14-19.** Surgical procedures for pes varus. (Modified from Graham HK. Cerebral palsy. In: McCarthy JJ, Drennan JC. *Drennan's the child's foot & ankle*, 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:188–218, Chapter 13.)

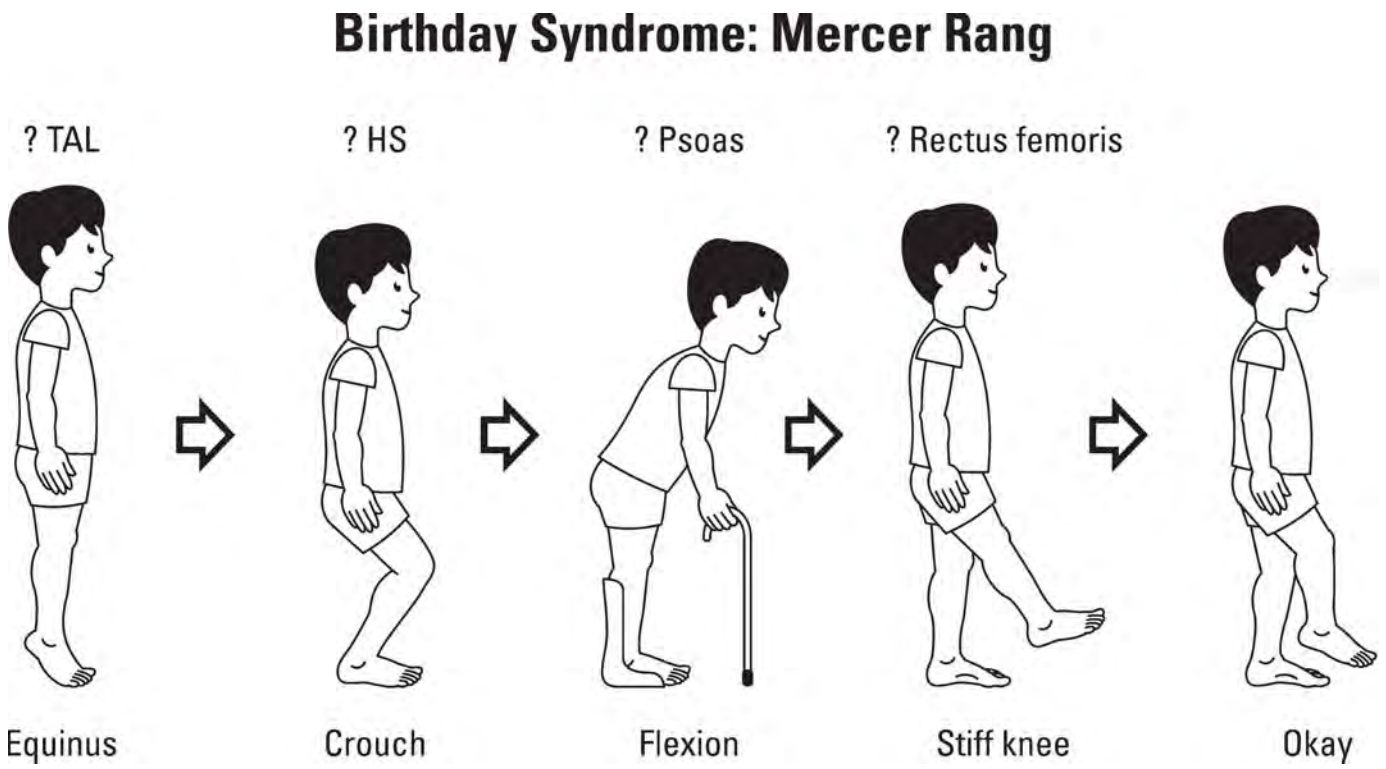



**FIGURE 14-20.** The Birthday Syndrome as described by Mercer Rang.

surgery (156). It will not be necessary if soft-tissue balancing is performed at the appropriate age and stage (133).

## GMFCS II—Moderate Spastic Diplegia

**Movement Disorder Management.** The movement disorder is usually spastic, especially in those born prematurely. If the spasticity is mild and mainly distal, it can be managed by multilevel injections of BoNT-A repeated at 6- to 12-month intervals. SDR may be a better option when the spasticity is severe, generalized, and adversely affecting gait and function (38, 88).

**Hip Displacement GMFCS II.** In spastic diplegia, GMFCS level II, the shape of the proximal femur is abnormal with a mean FNA of 36 degrees and a 15% risk of hip displacement (49, 50). The hip displacement is generally mild and progresses slowly. Preventive surgery consisting of lengthening of the hip adductors is usually effective (148). Lengthening of the psoas at the pelvic brim may be required (157, 158). Hip displacement and gait dysfunction can be successfully managed by intertrochanteric proximal femoral osteotomy with derotation and a very small amount of varus. The hip abductors are frequently weak and correction of the NSA should therefore be to normal values. Excessive varus weakens the hip abductors and causes a Trendelenburg gait.

**Musculoskeletal Pathology.** Musculoskeletal pathology in spastic diplegia GMFCS level II includes increased FNA and contractures of the two joint muscles, the psoas, hamstrings, and gastrocnemius (29, 30). There is usually pes valgus and in adolescents hallux valgus. There is sometimes excessive ETT resulting in lower limb malalignment. In asymmetric diplegia, pelvic retraction may make clinical estimation of rotational alignment during gait very difficult, without IGA (Fig. 14-20).

**The Birthday Syndrome and SEMLS.** The natural history of *deformities* in the lower limbs at GMFCS level II is for gradual progression during childhood with more rapid deterioration during the adolescent growth spurt (29). The natural history of *gait* is progressive deterioration including increasing stiffness throughout the lower limb joints and increasing tendency to flexed knee gait and ultimately crouch gait (16–18). The transition from equinus gait to crouch gait is often accelerated by procedures that weaken the gastrocsoleus, especially lengthening of the Achilles tendons (95) (Fig. 14-21).

Surgery for children with spastic diplegia used to start at the ankles with TALs for equinus gait. This achieved foot-flat but at the expense of rapidly increasing hip and knee flexion (95). The second stage of surgery was then to lengthen the hamstrings in order to improve knee extension. This resulted in increased hip flexion and anterior pelvic tilt, so eventually the hip flexors were lengthened. Finally, transfer of the rectus femoris was considered for knee stiffness. This approach was caricatured by Mercer Rang as the "Birthday Syndrome" (148). Children spent most of their birthdays in hospital, in casts, or in rehabilitation.

The current concept for the management of musculoskeletal deformities is SEMLS (40, 148). In this approach, the gait pattern is identified and evaluated by IGA as part of

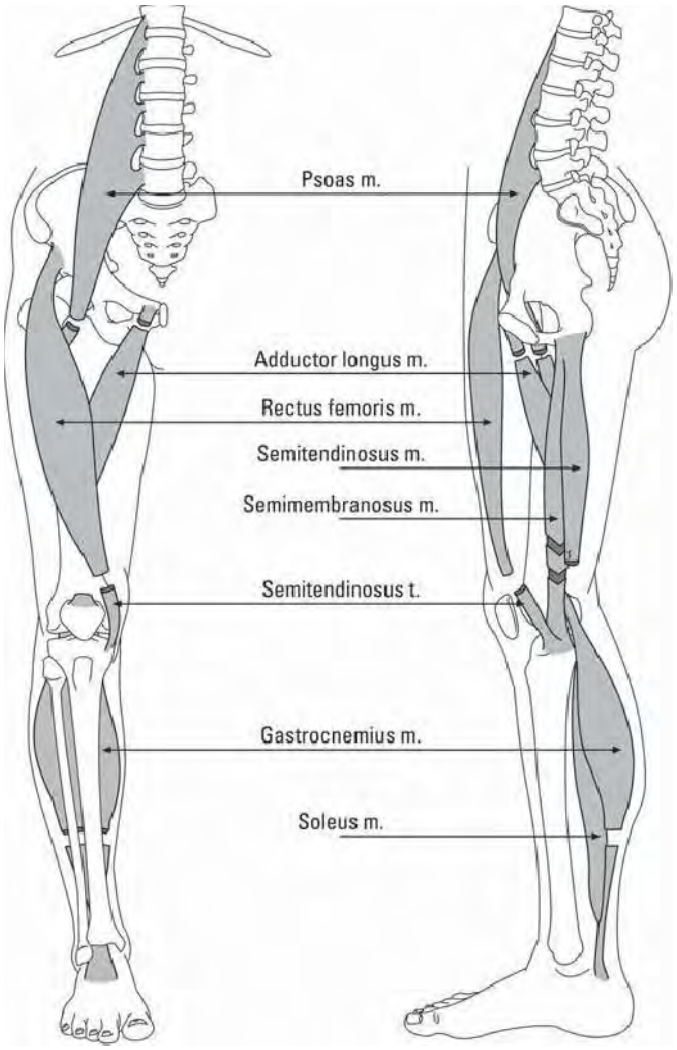



**FIGURE 14-21.** The most commonly used soft-tissue procedures in Single Event Multilevel Surgery. (From Bache CE, Selber P, Graham HK. Mini-Symposium: cerebral palsy: the management of spastic diplegia. *Curr Orthopaed* 2003;17:88–104, with permission.)

the diagnostic matrix (40, 68). A comprehensive plan is then developed for the correction of all muscle tendon contractures, torsional malalignments, and joint instabilities in one operative session (157, 159). Rehabilitation requires at least 1 year and improvements continue into the 2nd and 3rd years, postoperatively. The GMFCS descriptors for children at level II, aged 12 to 18, include the need for assistive devices for longer distances (15). Children who have optimal biomechanical alignment of their lower limbs by multilevel surgery continue to function independently throughout the second and third decades of life.

The principal components of a successful SEMLS program are:

1. Planning based on the diagnostic matrix, including gait analysis (68)
2. Preparation and education of the child and family (157)
3. Optimal perioperative care, including epidural analgesia (157)
4. Carefully planned and supervised rehabilitation (159)
5. Appropriate orthotic prescription (157)
6. Close monitoring of functional recovery (63)
7. Follow-up gait analysis at 12 to 24 months after the index surgery (159)
8. Removal of fixation plates and other implants (159)
9. Follow-up until skeletal maturity, for new or recurrent deformities (63).

The surgical team should consist of two experienced surgeons and two assistants. None of the surgical procedures are particularly complex, but a single surgeon is unable to perform 8 to 16 consecutive procedures without fatigue and diminished performance (159). Expert anesthesia and pain management is essential. Epidural analgesia is required to make SEMLS acceptable on social and humanitarian grounds (157). Postoperative nursing care must be vigilant. The use of epidural analgesia carries risks of masking the signs of compartment syndromes, nerve stretch palsies, and decubitus ulceration. The surgery is a series of steps that correct deformity. However, for 6 to 9 months after surgery, children are more dependent and less functional than they were prior to surgery. A child who walks into hospital with a typical diplegic gait pattern, leaves hospital in a wheelchair with straighter legs but may be unable to walk independently for weeks or even months. Only a carefully tailored and carefully monitored rehabilitation program can ensure that the child will reach a higher level of function (63, 157).

Weight bearing should commence within a few days if there has been no bony surgery, or a femoral osteotomy with stable internal fixation. The maximum acceptable delay to full weight bearing is 2 weeks, if there has been extensive reconstructive surgery at the foot-ankle level (157). Casts are only required after foot and ankle surgery. Removable extension splints may be used at the knee level after hamstring-rectus surgery. The goal is to achieve full extension of the knee, combined with regaining full flexion, so that the transferred rectus femoris does not become scarred and adherent in its new site. New AFOs must be prepared for immediate fitting after cast removal, usually 6 weeks after surgery. The initial postoperative brace is either a Ground Reaction or Saltiel AFO (GRAFO) or a solid AFO. The orthotic prescription must be carefully monitored throughout the first year after surgery (63, 157). A less supportive AFO, such as a hinged or a posterior leaf spring, may be introduced when the sagittal plane balance has been restored and the plantar-flexion, knee-extension couple is competent. Functional recovery and orthotic prescription can be monitored by a gait laboratory visit every 3 months for the first year after surgery and yearly thereafter.

## Soft-Tissue Surgery: Lengthening of Contracted Muscle-tendon Units (The Two Joint Muscles)

1. Lengthening of the psoas "over the brim" (POTB) (158, 160)
2. Percutaneous or open lengthening of adductor longus (157) (see Figs. 14-35 to 14-40)
3. Medial hamstring lengthening (MHS) (161)
4. Distal gastrocnemius recession (Strayer) (162) (Fig. 14-22) (see Figs. 14-14 and 14-15)

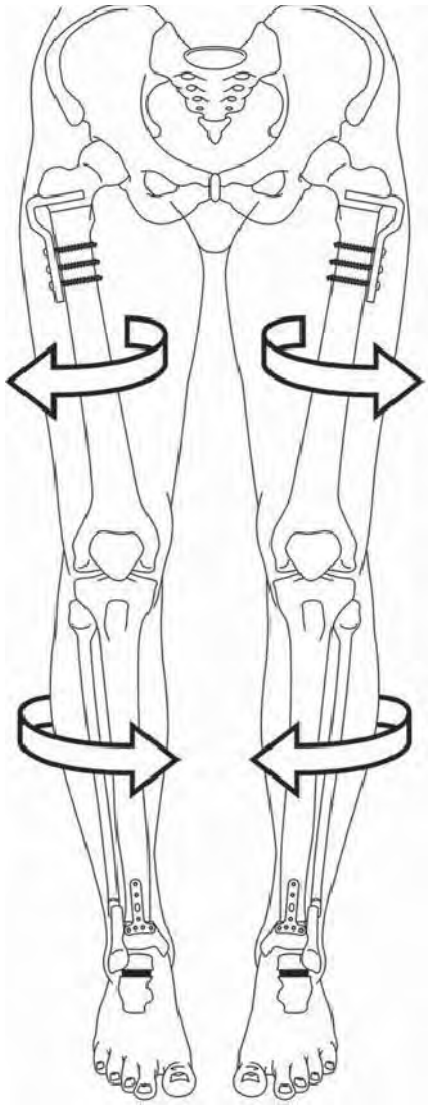


**FIGURE 14-22.** The most commonly used bony procedures in single event multilevel surgery; femoral derotation, supramalleolar osteotomy of the tibia, and stabilization of the midfoot. (From Bache CE, Selber P, Graham HK. Mini-Symposium: cerebral palsy: the management of spastic diplegia. *Curr Orthopaed* 2003;17:88–104, with permission.)

## Soft-Tissue Surgery: Tendon Transfers

1. Transfer of rectus femoris to the semitendinosus or gracilis (163, 164)
2. Transfer of the semitendinosus to the adductor tubercle.
3. SPLATT for the varus foot (154, 155)

## Bony Surgery: Rotational Osteotomies

1. External rotation osteotomy of the femur (165, 166)
2. Internal rotation osteotomy of the tibia (167–169) (Fig. 14-22)

## Bony Surgery: Joint Stabilization

1. Hip: varus derotation osteotomy (VDRO) (150, 169).
2. Os calcis lengthening (170).
3. Talo-navicular fusion.
4. Subtalar fusion (171).

## Occasional Procedures

1. Pelvic osteotomy (172)
2. Fusion first MTP joint for hallux valgus (173)
3. Epiphysiodesis for LLD (157)
4. "Guided Growth": Staples or "8" plates for knee flexion deformity (174, 175)

**Principles of Surgical Treatment: Dynamic Ankle Function.** Contractures of the gastrocnemius and soleus can be measured by comparing the range of ankle dorsiflexion with the knee flexed (soleus) and extended (gastrocnemius). The Silfverskiold test should be performed both before surgery and during surgery for equinus in order to select the correct surgical "dose" (133). Hindfoot valgus in weight bearing is often associated with breaching of the midfoot, lateral subluxation of the navicular on the talus, abduction of the forefoot, and an increasingly external foot progression angle. This reduces stance phase stability and the GRF is also maldirected out of the plane of progression, resulting in abnormal stresses on proximal joints (176).

Standardized weight-bearing radiographs of the foot and ankle mortise are required in all children (67). Excessive ETT is frequently found with the valgus/abducted foot, and careful clinical and radiologic assessment is required to determine how much of each deformity is present (133). Accurate measurement of tibial torsion by physical examination is difficult. Three techniques have been described: the thigh-foot-angle, the bimalleolar axis, and the "second toe test" (176, 177).

**Foot and Ankle: Soft-Tissue Surgery.** The gastrocnemius is always more contracted than the soleus in spastic diplegia, and selective lengthening of the gastrocnemius is best for the majority of children (162). Even when a contracture of the soleus is present, differential lengthening of the gastrocnemius and soleus by a combination of the Strayer procedure combined with soleal fascial lengthening (SFL) is biomechanically more appropriate and safer than other procedures. Only very severe and neglected equinus deformity requires lengthening of the Achilles tendons. The White slide technique, performed under direct vision, is a much more controlled and satisfactory procedure than the triple hemisection technique, performed percutaneously (133, 178).

The main complication is gradual failure of the plantar-flexion, knee extension couple, leading to calcaneus gait, which is more disabling and difficult to treat than the original equinus gait. In diplegia, "a little equinus is better than calcaneus" (148). Isolated lengthening of the gastrocsoleus will result in the crouch gait in up to 40% of children with spastic diplegia (95). The "overlengthening" is mediated by biomechanical changes and growth, not surgical imprecision. When the GRF falls behind the knee, the soleus responds to the continual stretch by adding more sarcomeres in series. In time, the soleus becomes functionally too long, biomechanically incompetent and calcaneus-crouch progresses rapidly (59). Deferring the surgery until age 6 to 8 years reduces the risks of


**FIGURE 14-23.** **A and B:** Sagittal and coronal views of a 10-year-old boy with spastic diplegia showing the characteristic musculoskeletal pathology. The sagittal view shows jump alignment with mild equinus at the ankle and significant flexion deformities at the hip and knee. In the coronal plane, there is internal rotation of both femora, external rotation deformities in both tibiae resulting in "malignant malalignment" that is asymmetric. The external foot progression angle is a combination of external tibial torsion and pes valgus.

both recurrence and overcorrection (95). The more proximal operations on the gastrocsoleus are the most stable and safest in terms of avoiding calcaneus (162, 179–181) (Fig. 14-23).

**Surgical Technique (Strayer).** With the patient in the prone position, a posteromedial incision, 2 to 3 cm long, is made, centered over the musculotendinous junction of the gastrocnemius. The deep fascia is divided longitudinally, and the sural nerve and lesser saphenous vein are identified and protected. The plane between the gastrocnemius and the soleus is identified from the medial side and developed by blunt dissection. Once the two layers have been separated, the aponeurosis of gastrocnemius is divided transversely, the muscle bellies are allowed to recess proximally and are then sutured in the appropriate position (ankle in neutral, knee in extension). If the range of dorsiflexion is still limited to less than plantigrade, with the knee in extension, the fascia overlying the muscle belly of soleus can be divided transversely. After wound closure, a below knee cast is applied, with the ankle at neutral. This remains in place for 6 weeks and is then replaced by an ankle foot orthosis. This surgery is inherently stable and immediate weight bearing is encouraged (133, 151, 162, 182, 183) (Fig. 14-24A,B).

**Foot and Ankle: Bony Surgery.** Equinus leads to excessive loading of the forefoot and with time may cause breaching of the midfoot. A series of complex segmental malalignments of the midfoot, hindfoot, and forefoot develops referred to as pes equinoplanovalgus, pes planoabductovalgus, or simply "pes valgus." The component parts are valgus of the heel, pronation of the midfoot with flattening of the medial longitudinal arch, pronation and abduction of the forefoot with hallux valgus (Figs. 14-25 and 14-26A,B). Symptoms may include pain and callosities over the collapsed medial arch, particularly the head of the talus. This leads to pain, inability to wear AFOs, and discomfort in shoes. Evaluation includes the usual

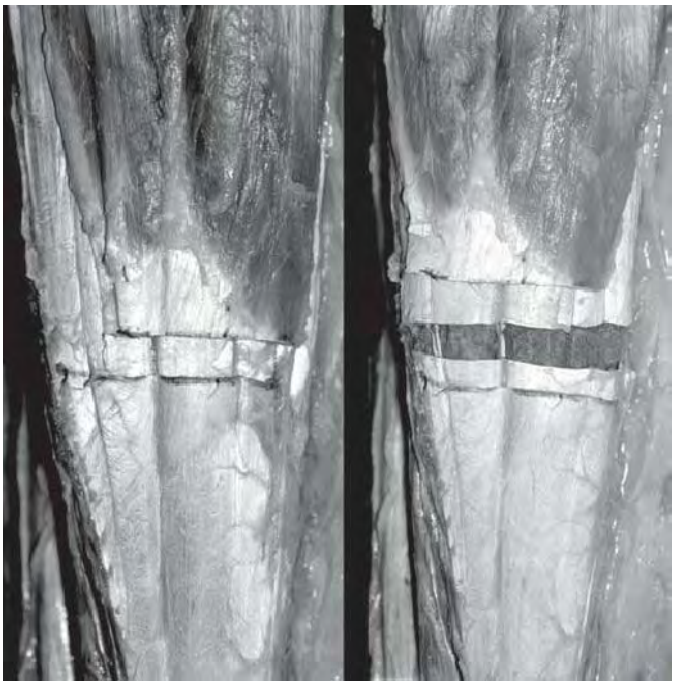


**FIGURE 14-24.** Cadaver dissection to demonstrate the distal gastrocnemius recession described by Strayer. The broad gastrocnemius aponeurosis has been divided transversely, at the distal extent of the medial gastrocnemius belly. This results in isolated lengthening of the gastrocnemius and is the safest procedure for equinus in diplegia because it avoids the risk of weakening of the soleus. In some children, lengthening of the soleus maybe required and this is illustrated on the right where the soleus fascia has been divided transversely exposing the soleus muscle fibers in the intervening gap. Note that when the Strayer procedure is combined with soleal fascial lengthening, the gastrocnemius is lengthened by more than the soleus that is biomechanically appropriate for the majority of children with spastic diplegia. (From Firth GB, McMullan M, Chin T, et al. Lengthening of the gastrocnemius-soleus complex. An anatomical and biomechanical study in human cadavers. *J Bone Joint Surg* 2013;95-A:1489–1496, with permission.)

components of the diagnostic matrix with special emphasis on weight-bearing radiographs of the feet and ankles, rather than motion analysis (68). A useful guide to the radiographic functional anatomy of the foot, with normal values for a series of radiographic parameters, has been published (67). Factors affecting the choice of operative procedure include the age of the patient and the clinical and radiographic severity of the deformity (133). The flexibility of the deformity is crucial because the commonly used surgical techniques depend on ligamentotaxis for the correction of all component parts of the deformity. The corrigibility of the deformity should be checked by placing the foot in an equinovarus position, while palpating the medial arch with special attention to the talonavicular joint. As the foot moves into equinovarus, the medial arch should be restored and the navicular should cover the head of the talus.

The midfoot can be stabilized and deformity corrected by lengthening of the lateral column of the foot (os calcis lengthening) or extra-articular fusion of the subtalar joint (170, 171, 184–186). Os calcis lengthening corrects subtalar joint eversion and midfoot breaching by elongating the lateral column of the foot, driving the heel out of valgus, into relative varus and raising the medial arch. This procedure has the advantage of preserving subtalar motion. The indication for os calcis lengthening is a flexible valgus deformity of the heel in association with an abductus deformity of the forefoot, in a patient who walks independently, GMFCS I or II (133, 186). Arthrodesis of the subtalar joint is a reliable means of correcting hindfoot valgus and with secondary correction of the midfoot. It is useful for more severe deformities in patients who require assistive devices and long-term orthotic support, GMFCS III and IV (133, 185). A modified Fulford technique is best, with a cannulated screw passed through the talar neck, across the sinus tarsi into the calcaneum, combined with iliac crest autograft or allograft (171). A third option for the correction of pes valgus in CP is calcaneo-cuboid-cuneiform (triple C) osteotomy (187). A fourth option which is gaining in popularity is isolated fusion of the talo-navicular joint.

Hallux valgus is commonly associated with deformities in the hindfoot, midfoot, and proximal gait deviations, such as stiff-knee gait, which causes toe scuffing. The most reliable procedure is fusion of the first MCP joint, either in conjunction with, or after correction of the proximal deformities. A cup-and-cone reamer technique with dorsal plate and screw fixation is effective and reliable (133, 173).

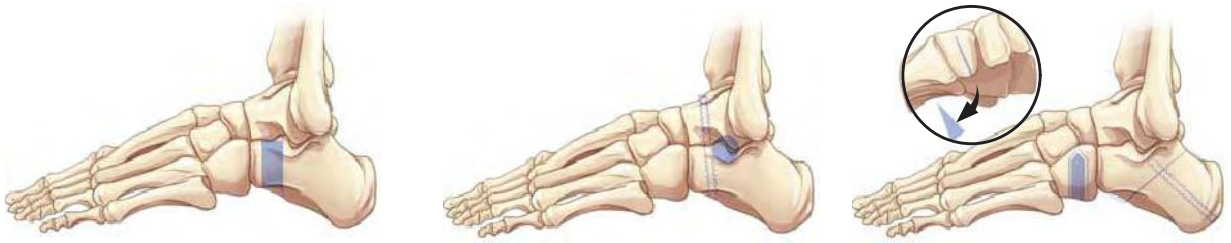



**FIGURE 14-25.** The three principal procedures for the correction of flexible pes valgus in CP. **A:** Lateral column lengthening. **B:** Extra-articular subtalar fusion (Dennyson and Fulford technique). **C:** The triple C osteotomy, closing lateral wedge of the cuboid, heel shift to the calcaneum, and opening medial wedge in the medial cuneiform. (Modified from Graham HK. Cerebral palsy. In: McCarthy JJ, Drennan JC. *Drennan's the child's foot & ankle,* 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:188–218, Chapter 13.)

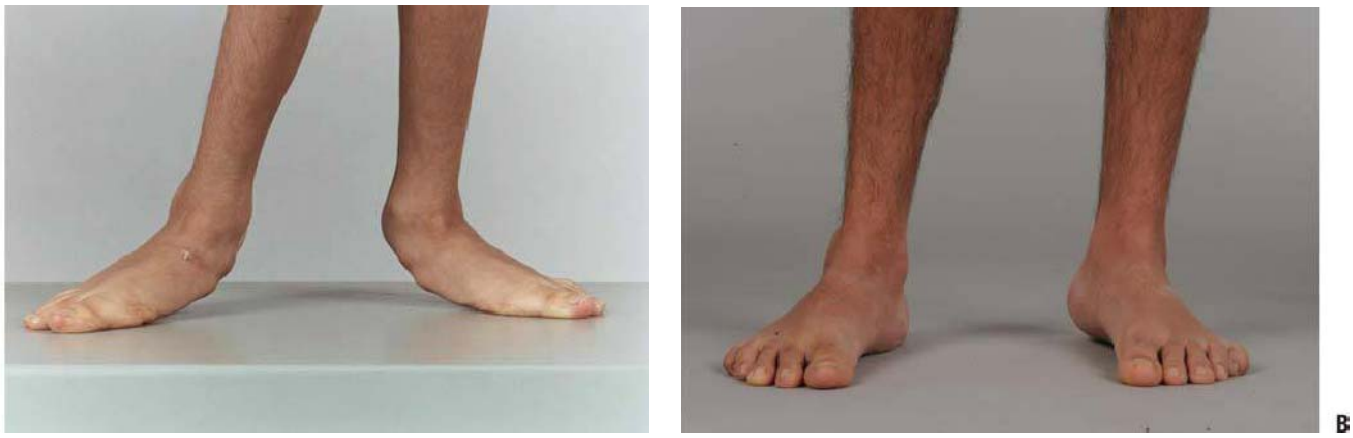


**FIGURE 14-26.** **A and B:** Severe pes valgus, and ETT in a 10-year-old boy with severe spastic diplegia before and after surgical correction that included Strayer gastrocnemius recessions, internal supramalleolar osteotomy of the tibia, and bilateral midfoot stabilization. Foot pain was relieved and he was able to resume wearing AFOs.

**Surgical Technique: Os Calcis Lengthening.** There are four principal steps, correct the equinus deformity, lengthen the lateral column, assess the medial column, and assess the ankle and tibia. Gastrocnemius recession to permit 5 degrees of dorsiflexion in subtalar neural is usually required (133) (Fig. 14-25).

An Ollier incision or a longitudinal incision can be used to approach the lateral aspect of the Os calcis. The aim of the surgery is to perform an osteotomy parallel to and approximately 1 cm proximal to the calcaneocuboid joint. The osteotomy should be between the middle and anterior facets of the subtalar joint. The osteotomy site is distracted with special distraction forceps, and a trapezoid of autologous or allograft corticocancellous bone is inserted. Some surgeons use a longitudinal K wire to transfix the osteotomy, the graft, and the calcaneocuboid joint. Calcaneocuboid subluxation is a risk that must be avoided. We prefer a stable, "press-fit" tricortical iliac crest allograft, without internal fixation, in the majority of children. After insertion of the graft, the alignment of the first ray and forefoot should be checked to determine if a plantarflexion osteotomy of the medial cuneiform is required to correct supination of the forefoot, unmasked by correction of the midfoot. The need for this procedure is not well defined and differences in practice in the literature may relate to the age of the patient and the stage of the deformity (133). Additional deformities that may need correction include ETT (SMO) and ankle valgus (SMO, guided growth).

Postoperatively, non–weight bearing in a below knee cast is advised for 2 weeks, followed by 4 weeks of weight bearing, protected in a cast, to a total of 6 weeks. At 6 weeks after surgery, graft incorporation is usually adequate to permit cast removal and fitting of an AFO. Short-term results are very good but failure may occur with time, particularly in more involved children, GMFCS III and IV (133, 170).

**Foot and Ankle: Supramalleolar Osteotomy of the Tibia.** ETT may occur in isolation or in conjunction with medial femoral torsion. In isolation, ETT results in an external foot progression angle and "lever arm disease" because the foot lever is effectively shortened and maldirected, in relation to the line of progression. Derotational osteotomy of the tibia is an effective means of addressing this problem (150). A very distal, supramalleolar osteotomy of the tibia is preferred 1-2 cm proximal to the distal tibial physis, which increases the cross-sectional area of contact between the two osteotomy surfaces. This increases stability, facilitates early weight bearing, minimizes the risk of secondary deformities, and is associated with reliable and rapid union. In order to gain the desired degree of rotation (which can be determined using transverse plane kinematic data), the fibula may sometimes need to be divided at the same level as the tibia. The osteotomy of the tibia may be stabilized using a straight DC plate, crossed Kirschner wires or the AO/ASIF small fragment, contoured T plate (150, 161, 163, 164). External foot progression angle is usually the result of combined pes valgus and ETT. The decision to perform os calcis lengthening, SMO, or a combination requires a very careful assessment using all components of the diagnostic matrix (68, 133). Finally, ETT may be disguised by increased FNA (malignant malalignment). The torsional deformities in both the femur and tibia should be corrected (Fig. 14-27).

**Knee.** The principal gait dysfunctions are stiffness and excessive flexion. Recurvatum is sometimes seen after excessive hamstring lengthening with an equinus contracture. Hamstring spasticity and contracture are often evaluated by measuring the popliteal angle. Unfortunately, the popliteal angle has little correlation with knee flexion during gait. Hamstring function is better assessed dynamically, based on muscle elongation rates and muscle lengths in the second half of swing (149). Improvements in knee extension during late swing occur when decisions regarding hamstring lengthening are made consistent with muscle length and velocity data. When hamstrings are lengthened in cases of normal hamstring function (normal muscle length and velocity in swing), excessive anterior pelvic tilt and pelvic range of motion may develop (Fig. 14-28).

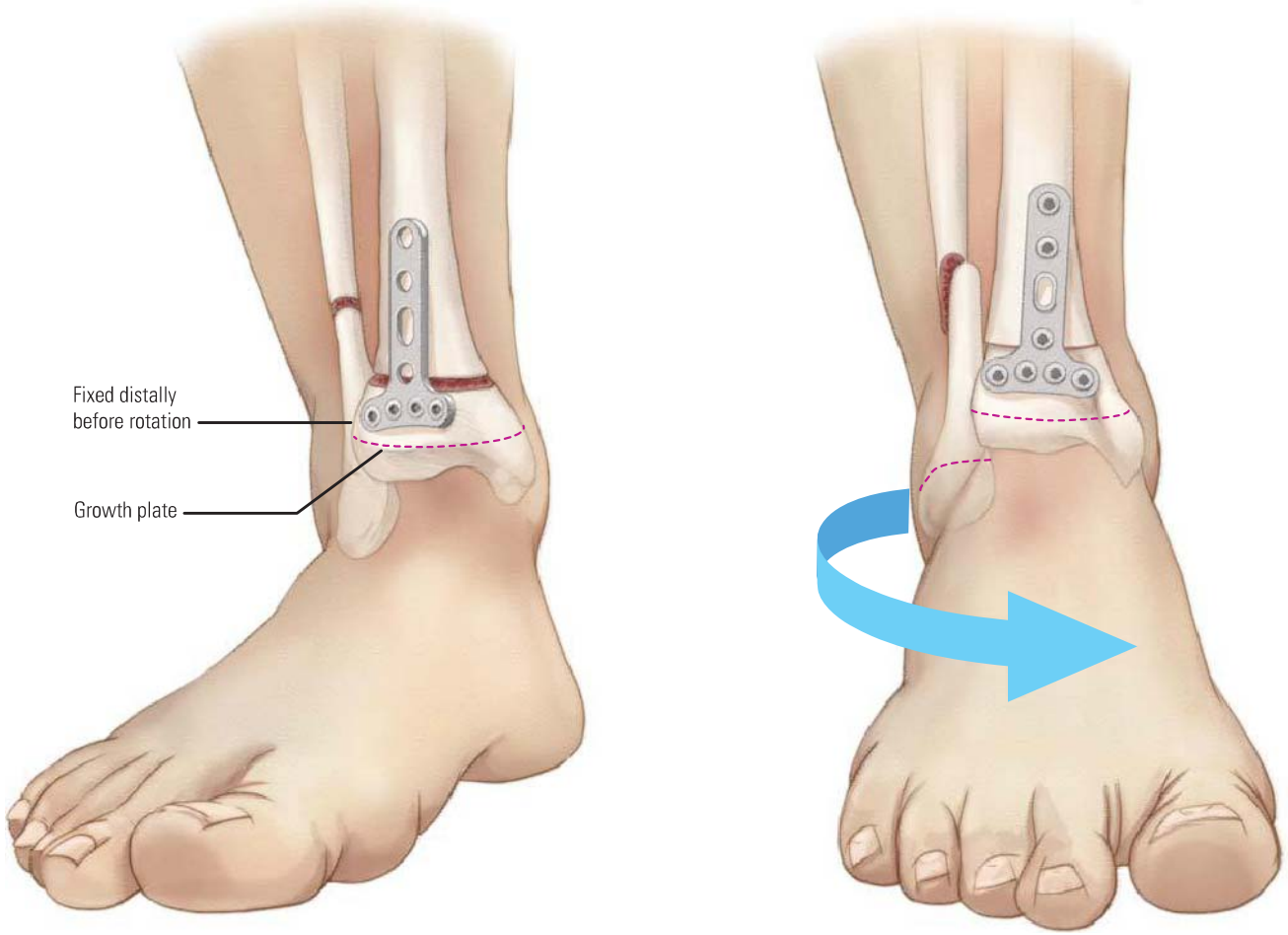



**FIGURE 14-27.** The technique of supramalleolar osteotomy of the distal tibia and fibula, with "T" plate fixation. The "T" plate is fixed to the distal tibia, 1 cm above the physis, prior to osteotomy of the tibia and fibula. The plate is then removed, the osteotomy is performed, the external rotation deformity is corrected, and the plate reapplied to fix the distal tibia, in the corrected position. (Modified from Selber P, Filho ER, Dallalana R, et al. Supramalleolar derotation osteotomy of the tibia, with T plate fixation: technique and results in patients with neuromuscular disease. *J Bone Joint Surg* 2004;86-B:1170–1175.)

**Knee: Soft-Tissue Surgery.** Fractional lengthening of the medial hamstrings can be accomplished through a midline posterior incision just above the knee. Gracilis and semitendinosus are lengthened in continuity by intramuscular tenotomy and the semimembranosus, by performing one or two stripes through its broad aponeurosis (157, 161). The semitendinosus or gracilis may be harvested at the time of medial hamstring lengthening, for subsequent transfer of the rectus femoris. The rectus can be detached distally from the patella and "tubed" around the harvested semitendinosus or gracilis tendon. Early mobilization is required to prevent adhesion formation (157) (Fig. 14-28).

There are two major problems with distal hamstring lengthening. Firstly, it frequently fails to achieve adequate knee extension during gait suggesting that other factors are causative. Secondly, it may cause or exacerbate anterior pelvic tilt (59). If an anterior pelvic tilt is present preoperatively, hamstring lengthening should either not be done or should be done cautiously and in conjunction with other efforts to treat the anterior pelvic tilt. Distal hamstring lengthening works best for mild dynamic deformities, in children at GMFCS levels I and II without any fixed flexion contracture at the knee. Distal hamstring lengthening is ineffective when knee flexion contracture exceeds about 5 to 10 degrees. When knee flexion deformity exceeds 5 degrees, in GMFCS III and IV, we prefer distal hamstring lengthening combined with transfer of the semitendinosus to the adductor tubercle (188) (see Figs. 14-29 to 14-33).

**Hip.** Hip flexion contractures are common and should be managed by lengthening of the psoas tendon at the brim of the pelvis (157, 158). Tenotomy at the lesser trochanter may result in excessive weakness of hip flexion and is reserved for the nonambulant patient. In the Sutherland technique, the femoral nerve is first identified and protected, before the psoas tendon is sectioned. It is inherently safer than the alternative technique, a modification of the approach to the psoas tendon described by Salter for innominate osteotomy of the pelvis. In this approach, the psoas is identified by palpation, lying between the iliacus muscle and the periosteum of the ilium, but the femoral nerve is not visualized (158). Both techniques are effective in the correction of hip flexion contracture and the associated gait

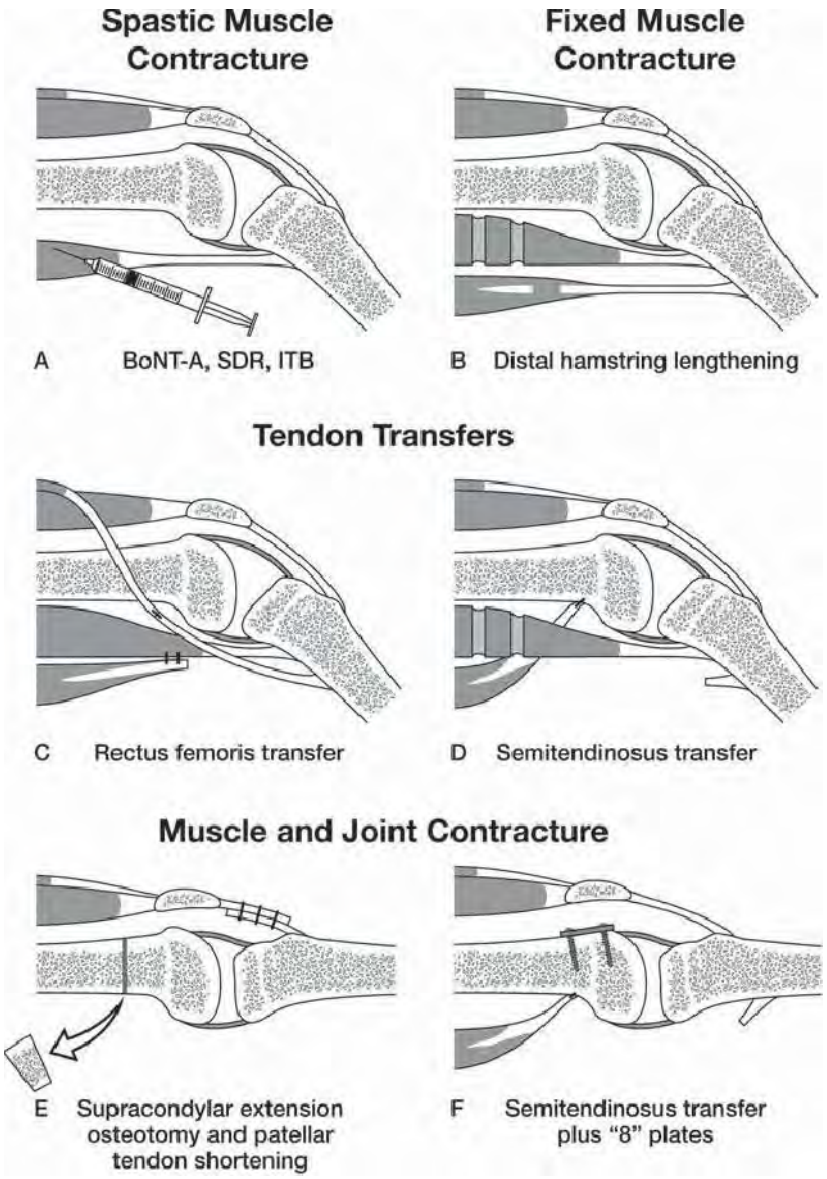



**FIGURE 14-28.** Management options for flexed knee gait in CP. (From Young JL, Rodda J, Selber P, et al. Management of the knee in spastic diplegia: what is the dose? *Orthoped Clin North Am* 2010;41:561–577, with permission.)

disturbance. The principal risk is injury to the femoral nerve because it is mistaken for the psoas tendon. Bilateral femoral nerve injuries have occurred but have not been reported by experienced surgeons. The Sutherland technique may be safer for less experienced surgeons (158). This is a stable lengthening and does not require immobilization. Prone positioning is required postoperatively to encourage hip extension.

**Hip: Bony Surgery.** Proximal femoral osteotomy is usually performed with the patient in the prone position, when the rotational arcs of both hips can be easily checked before and during surgery. A 90- or 100-degree AO/ASIF blade plate is used to achieve stable fixation. Anteversion is corrected to about 10 degrees, leaving only 10 to 20 degrees of internal rotation at the hip. The proximal osteotomy effectively lengthens the psoas and is the preferred technique (159) (see Figs. 14-35 to 14-40).

SEMLS is an exercise in correcting anatomical deformities based on clinical and radiologic examination and a biomechanical analysis of gait deviations. However, children with spastic diplegia have psychological and physiologic dimensions, which make successful surgical outcomes unpredictable. Weakness is a fundamental issue that is easily overlooked and may have a greater impact on energy cost of walking and function in the community than multiple musculoskeletal deformities (151). Careful preoperative assessment and goal setting helps to ensure that parent and surgeon goals are consistent. The majority of children have a satisfactory correction of gait but remain in their preoperative GMFCS level.

A recent systematic review of SEMLS found evidence for large improvements in gait dysfunction and moderate improvements in health-related quality of life. Changes in gross motor function were generally small and inconsistent (189).

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## Distal Medial Hamstring Lengthening Combined with Transfer of the Rectus Femoris to Semitendinosus (Figs. 14-29 to 14-33)

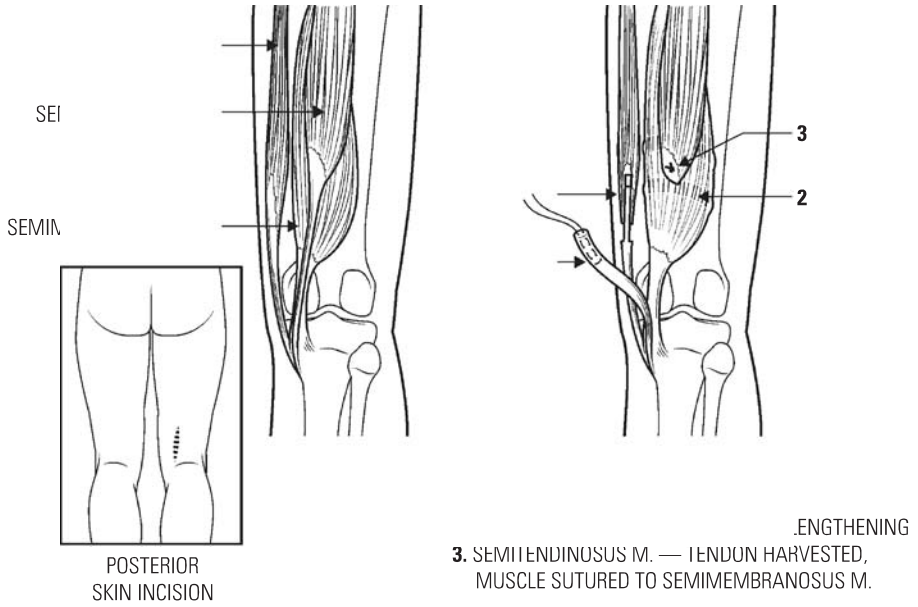



**FIGURE 14-29. Distal Medial Hamstring Lengthening Combined with Transfer of the Rectus Femoris to Semitendinosus.** In the majority of children, lengthening of the medial hamstrings is performed first, with the patient in the prone position. The position of the skin incision is indicated and with experience and good retraction, can be restricted to between 4 and 6 cm long on the posteromedial aspect of the distal thigh, terminating just above the popliteal crease. The gracilis is lengthened by an intramuscular technique. The semimembranosus muscle is lengthened by one or two transverse divisions of the fascial coat. The semitendinosus muscle is sutured to the underlying semimembranosus to prevent retraction and loss of important hip extensor function. The semitendinosus is then divided at the musculotendinous junction, secured with a whip stitch and mobilized distally to its insertion on the posteromedial aspect of the proximal tibia. The harvested semitendinosus tendon is placed in a subcutaneous position on the medial aspect of the distal thigh. A subcutaneous pocket is developed by blunt finger dissection. The incision is then closed in layers, a sterile dressing is applied. The patient is then turned into the supine position and the lower limbs prepared and redraped.

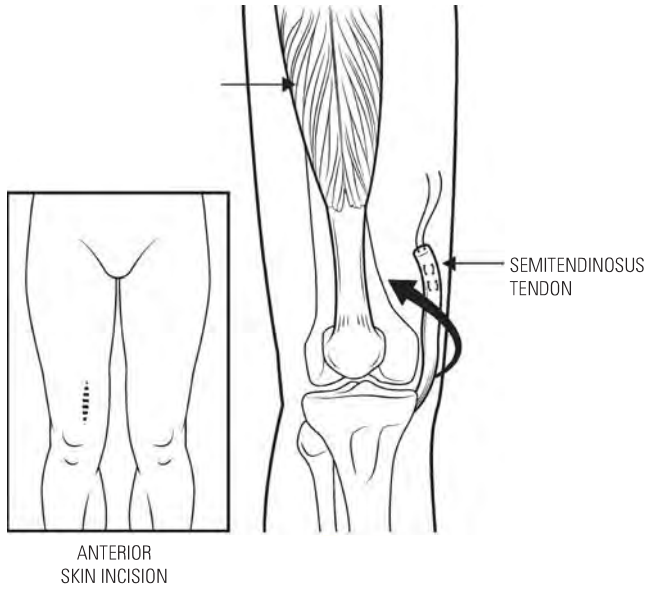



**FIGURE 14-30.** With the patient in the supine position a second incision is made on the anteromedial aspect of the thigh starting at the proximal medial pole of the patella and extending 4 to 6 cm proximally. The interval between the rectus femoris tendon and the underlying vastus intermedius is identified and mobilized by blunt dissection.

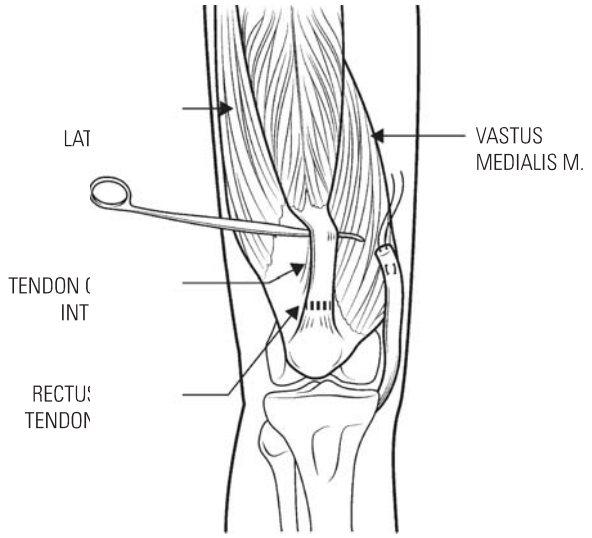



**FIGURE 14-31.** The rectus femoris tendon needs to be fully mobilized from the underlying tendon of the vastus intermedius distally. Proximally it needs to be separated from the overlapping bellies of the vastus medialis and lateralis. In the interests of cosmesis, it is important to use a small skin incision and to accomplish the proximal dissection by visualization using well placed retractors. Once the tendon of the rectus femoris has been fully mobilized from the underlying vastus intermedius, it is divided transversely just above its insertion on the patella.

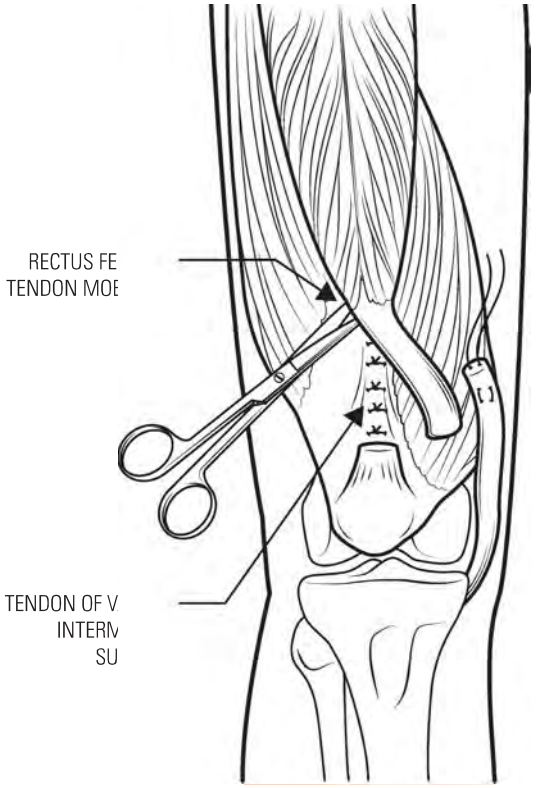



**FIGURE 14-32.** Any defects in the vastus intermedius and the capsule of the knee joint are carefully repaired. It is also helpful to approximate the vastus medialis to the lateralis underneath the rectus femoris to minimize dead space and prevent adhesions.

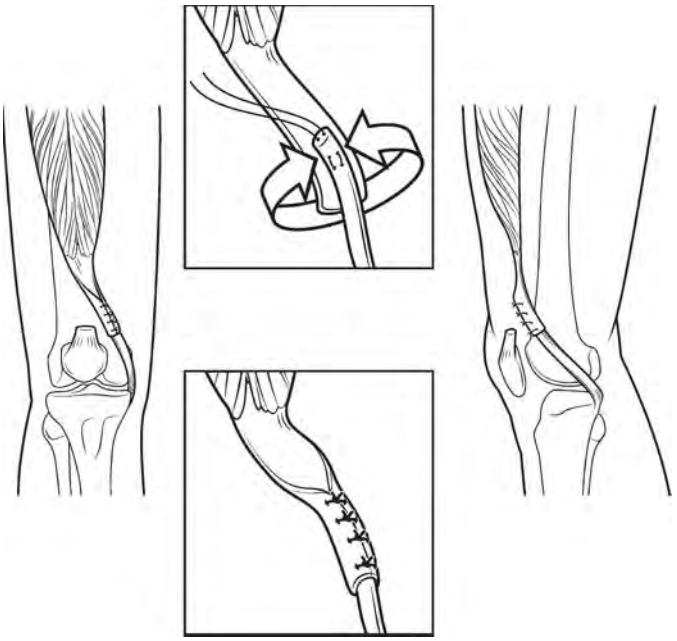


**FIGURE 14-33.** The relatively flat tendon of the rectus femoris is then transferred medially to meet the previously harvested tendon of the semitendinosus. The flat rectus femoris tendon is wrapped around the semitendinosus to form a tube and secured by a combination of the previous whip stitch in the semitendinosus combined with interrupted sutures to the rectus femoris. To permit a stable repair and early mobilization it is recommended that a nonabsorbable suture such as Ethibond is utilized. It is vitally important that the trajectory of the rectus femoris be smooth and unencumbered by adhesions or subcutaneous fascia as shown in both **A** and **B** from the anterior and medial aspects. Postoperative rehabilitation is crucial. Active and passive range of motion exercises are commenced on the first postoperative day. We recommend regaining 30 degrees of knee flexion by the end of the first postoperative week, 60 degrees of knee flexion by the end of the second postoperative week, and 90 degrees at the end of the third postoperative week. During this time, full knee extension is maintained by the use of a removable knee immobilizer. Cast immobilization cannot be used because adhesions will form and the transferred tendon will not regain gliding motion. If there are any concerns about the patient's ability to comply with early active and passive range of motion exercises then continuous passive motion (CPM) is a useful alternative.

The first RCT of SEMLS reported a 50% improvement in gait function (Gillette Gait Index, GGI) and a 4.9% improvement in gross motor function (GMFM-66) (159).

## GMFCS III

1. **Between 6th and 12th birthday**
   Children walk using a handheld mobility device in most outdoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances (13).

2. **Between 12th and 18th birthday**
   Youth are capable of walking using a handheld mobility device. They may climb stairs holding onto a railing with supervision or assistance. At school, they may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community youth are transported in a wheelchair or use powered mobility (15).
3. **Risk of hip displacement:** 41%
4. **Mean femoral neck anteversion (FNA):** 40 degrees
5. **Mean neck shaft angle (NSA):** 149 degrees (49, 50)

At GMFCS level III, children and adults ambulate in the community using an assistive device. The predominant movement disorder at GMFCS level III is spasticity, but some children have dystonia or a mixed movement disorder. Weakness of the major lower limb muscles, particularly those contributing to body support, is a major feature. Flexed knee gait patterns predominate and weakness is usually the primary determinant of long-term gait and community function rather than spasticity (79, 97). It is important to differentiate between those individuals who are "being pulled down" by spasticity and those who are "falling down" because of weakness (85). The strength and selective motor control of the muscle groups that contribute to the body support moment, the gastrocsoleus, quadriceps, and hip extensors is crucial (85).

The musculoskeletal pathology at GMFCS level III is similar to that at GMFCS level II, but the contractures of the MTUs are usually more severe and the deformities in the bony levers (femur and tibia) and joint instability (hip and foot) are more pronounced. Severe "lever arm deformities" are common at GMFCS level III with increased FNA, marked ETT, and pes valgus (30, 41).

**Hip Displacement at GMFCS III: Hip Surveillance and Preventive Hip Surgery.** Mercer Rang suggested many years ago that all children with CP should have regular hip examinations and radiographs (148). The goal was to prevent dislocation by early detection and early preventive (adductor release) surgery. He famously stated that no child with CP was actually helped by having a dislocated hip. Several centers in Europe and Australia have developed these concepts into formal "Hip Surveillance Programs." Children with a confirmed diagnosis of CP are offered regular clinical and radiographic examination of their hips and access to both preventive and reconstructive surgery. In both Southern Sweden and Victoria, Australia, the prevalence of late dislocation has decreased and the need for salvage surgery has been reduced (65, 190) (Fig. 14-34) (see Figs. 14-35 to 14-40).

Hip displacement in children with CP is different to DDH in typically developing children. The hip is normal at birth and then displaces because of limitations in activity, accompanied by contractures and bony deformities. Factors that may contribute to hip displacement include increased magnitude of muscle forces across the hip, which have been modeled to be increased sixfold (191). The shape of the proximal femur is also important and is predicted by GMFCS level, in terms of torsion (femoral neck anteversion, FNA) and NSA (49).

Hip displacement in children with CP can be reliably measured from AP hip radiographs, taken in the supine

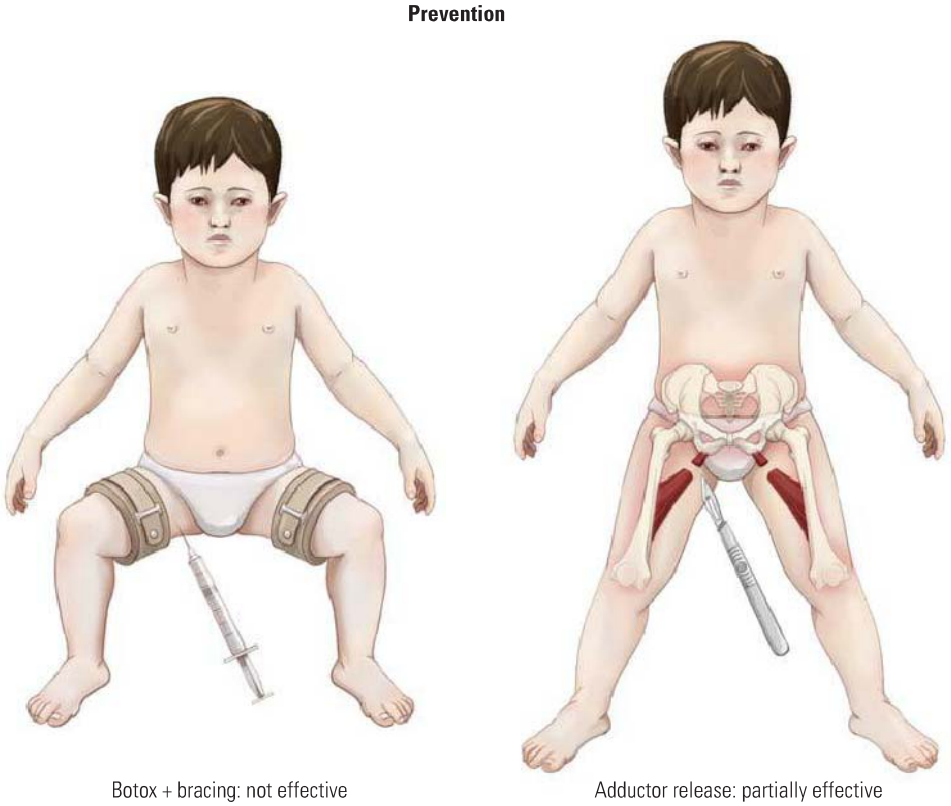
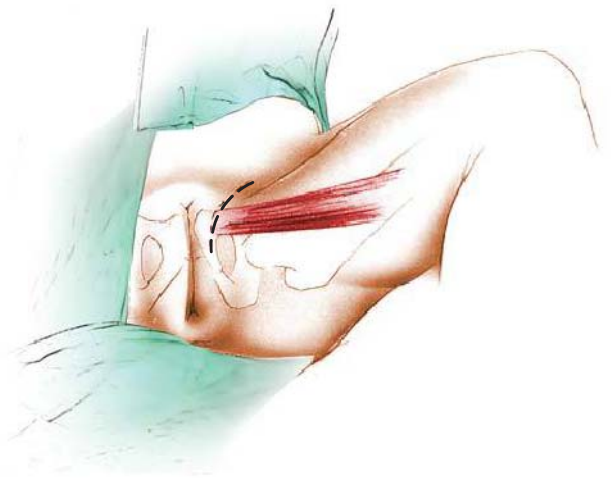



**FIGURE 14-34.** Prevention of hip displacement in younger children with CP is difficult. Repeated injection of Botulinum toxin to the hip adductors combined with an abduction brace is not effective. Adductor release surgery is partially effective. Success rate is high at GMFCS level II and lower at GMFCS level III. The failure rate in nonambulators, GMFCS IV and V, is very high.

*Text continued on page 525*

## Adductor and Iliopsoas Release (Figs. 14-35 to 14-40)

**FIGURE 14-35. Adductor and Iliopsoas Release.** The patient is placed supine, near the end of the operating table and the perineum is isolated with a waterproof dressing. Both legs are draped free so that the range of abduction in both flexion and extension can be checked. A transverse incision 2 to 3 cm long is made 1 cm distal to the groin crease, parallel to the groin crease, and centred over the adductor longus. The incision is opened down to the deep fascia with careful hemostasis.



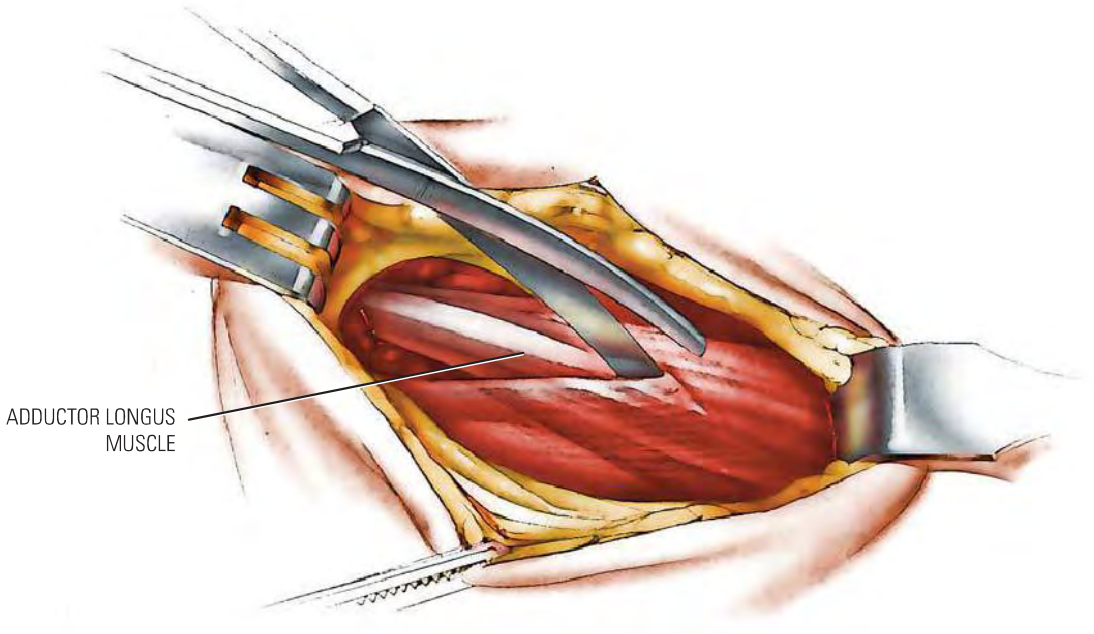



**FIGURE 14-36.** The adductor longus tendon and the interval between the adductor longus and brevis are carefully identified by blunt dissection. The fascia overlying the adductor longus is opened longitudinally, that is, at right angles to the skin incision and parallel with the axis of the adductor longus tendon.

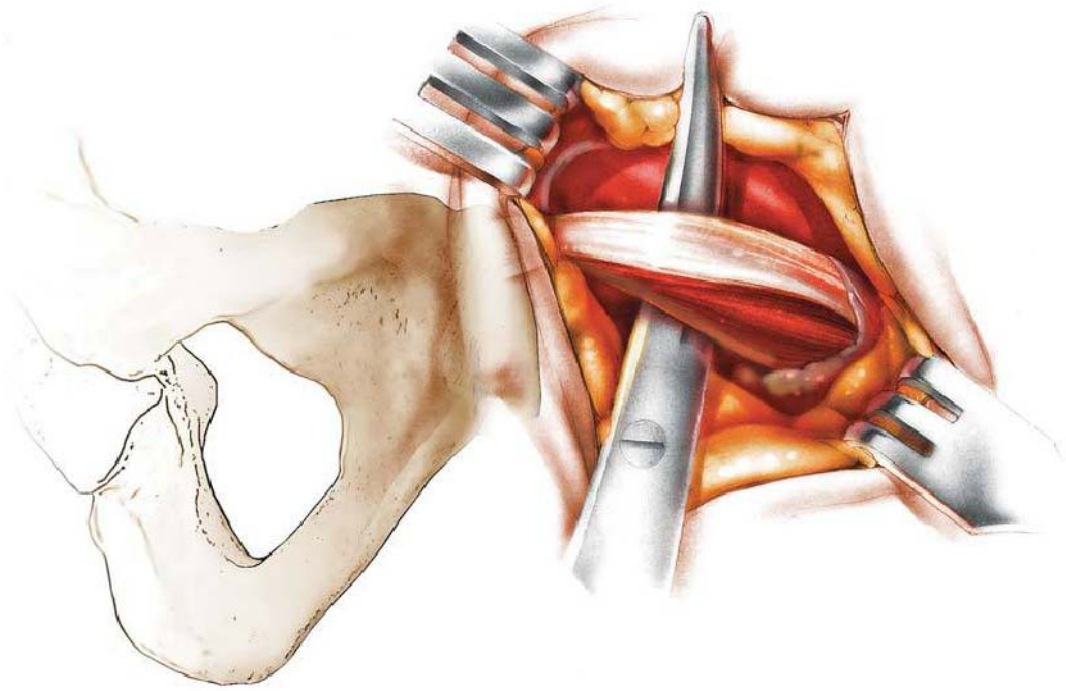


**FIGURE 14-37.** The tendon of the adductor longus is mobilized and divided close to its attachment to the pelvis, because this minimizes bleeding and dead space. The anterior branches of the obturator nerve can be identified in the interval between the adductor longus and brevis and should either be protected (in ambulators) or consideration given to phenol neurolysis in nonambulant patients with severe spasticity.

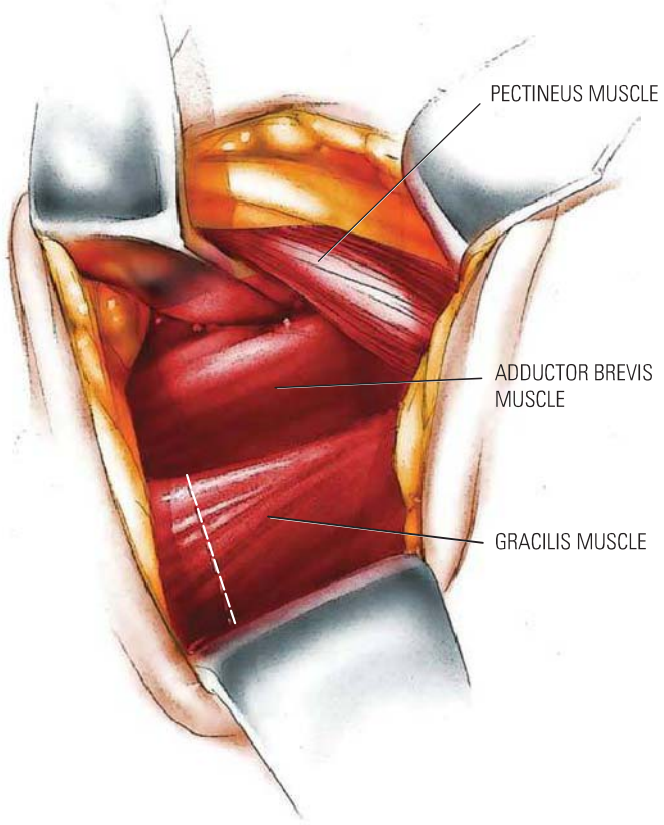
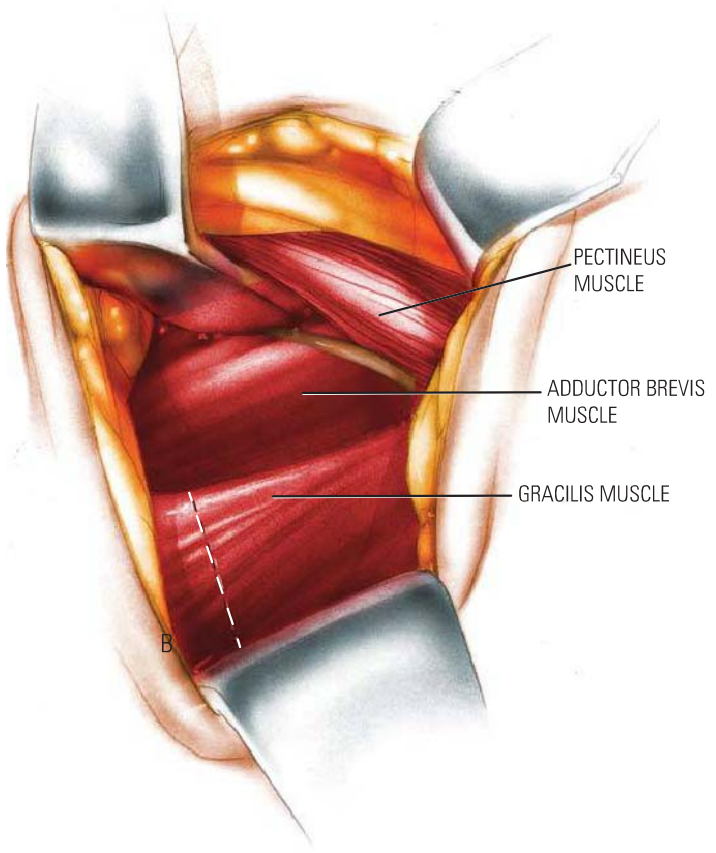



**FIGURE 14-38.** At this point the range of abduction in flexion should be checked carefully and the underlying adductor brevis can usually be manually stretched to provide sufficient abduction.

Next the origin of the gracilis is identified and mobilized. This can be facilitated by having the assistant extend the knee while abducting the hips. The gracilis is divided close to its origin from the pubis using electrocautery with attention to hemostasis.

**FIGURE 14-39.** The adductor brevis rarely requires division, it is better to manually stretch this muscle. In severe or neglected cases partial division may be required. Complete division should not be undertaken lightly because the posterior branch of the obturator nerve will be at risk. Excessive muscle division and/or injury to the posterior branch of the obturator nerve may lead to a fixed abduction deformity.



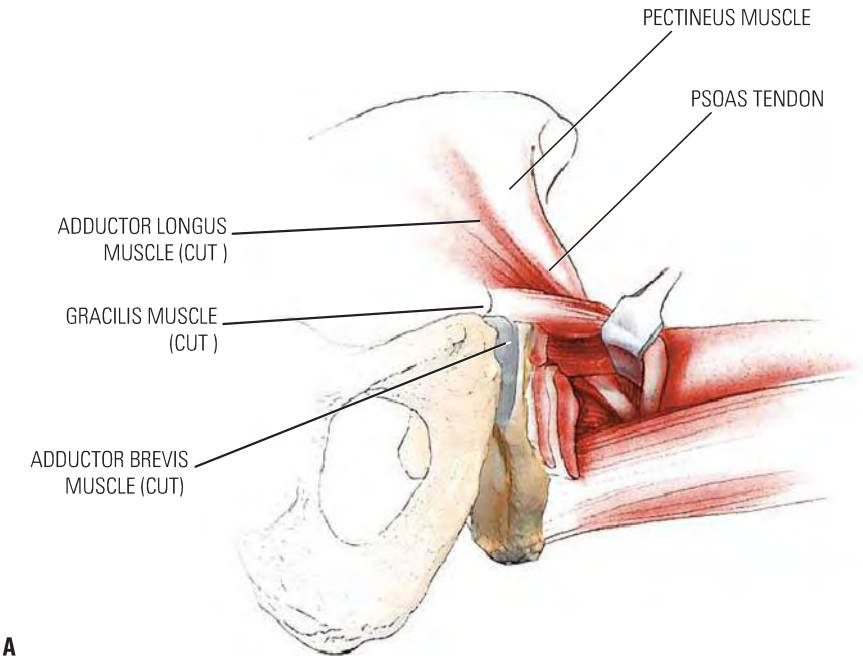




**FIGURE 14-40.** **A:** The decision whether to lengthen the iliopsoas at the lesser trochanter is based on the presence of a hip flexion deformity and the ambulatory status of the patient. If the hip flexion contracture is less than 5 degrees and the patient is an ambulator, lengthening at the lesser trochanter is not desirable. In nonambulators with a flexion contracture of more than 5 to 10 degrees, lengthening of the iliopsoas at the lesser trochanter is usually recommended. It is important to identify the correct intramuscular interval. This is best done by finger dissection along the posterior border of the divided adductor longus. The hip is held in abduction and flexion, with the surgeon's index finger palpating the proximal femur. The underlying femur can be identified by gently rotating the hip internally and externally followed by feeling the "bump" of the lesser trochanter. Then appropriate retractors can be inserted above and below the femur taking care to retract the neurovascular bundle anteriorly. It is important to place the retractors carefully and ensure a good view of the lesser trochanter with the psoas tendon running obliquely from the proximal thigh and inserting on the lesser trochanter. There is often a small amount of overlying fat which needs to be bluntly removed. A right angle clamp can be placed underneath the tendon of the iliopsoas which can then be divided completely under direct vision.

progressive hip displacement in children with CP (111). Hip surgery for children with CP can be classified as preventive, reconstructive, and salvage (191). Surgery to prevent hip displacement refers to soft-tissue releases of the hip adductors and flexors to prevent or reverse early hip displacement in younger children (194). Because the outcome of preventive surgery depends on the age of the child (younger children have better results) and the initial migration percentage (MP <40%), early and regular hip surveillance is recommended (64, 65, 195). The majority of children will require open lengthening of the adductor longus and gracilis with lengthening of the psoas over the brim of the pelvis (POTB). The results in children at GMFCS III, who are walking with external support, are very good. Several studies have reported that the outcome of preventive surgery is much better in ambulators than in nonambulators (194–196). The effectiveness of adductor surgery is predicted by GMFCS level (197).

**Surgical Correction of Crouch Gait in Spastic Diplegia.** Crouch gait may occur at GMFCS level I but is usually mild because children at GMFCS level I have good strength and good selective motor control. Severe crouch gait may occur at GMFCS level IV, but given that sustained ambulation is not feasible in adult life, correction by invasive surgery is not appropriate. Severe crouch gait is the major functional issue at GMFCS levels II and III. It can be part of the natural history of gait in spastic diplegia but in most recent series, the majority of affected individuals had prior lengthening of the Achilles tendons (59, 189).

There is usually a delay between lengthening of the gastrocsoleus and the development of crouch gait (95). The Achilles tendons are often lengthened in children with spastic diplegia between the ages of 3 and 6 years. It may take another 3 to 6 years before crouch gait becomes a significant functional problem, and it is often not until the adolescent growth spurt when the maximum deterioration in gait and functioning occurs (29, 95, 189). Instead of "growing up" the adolescent with progressive crouch gait "sinks down," with an inability to maintain an extension posture at the hip and knee during the stance phase of gait. Contributing factors seem to be a mismatch between the strength of the one-joint muscles contributing to the body support moment (gluteals, quadriceps, and soleus) and the increased demand because of rapid increases in height and weight at the pubertal growth spurt. This typically occurs in conjunction with progressive bony deformities known as lever arm disease. Around the time of the pubertal growth spurt, increasing patella alta (sometimes with fractures of the patella or avulsions of the inferior pole) increasing ETT and breakdown of the midfoot with severe pes valgus, all contribute to increasing crouch, fatigue and decreasing ability to walk (30, 41, 95) (Figs. 14-41 and 14-42). Understanding the biomechanics of severe crouch gait has led to improved surgical management in recent years with the development of more effective techniques to achieve lasting correction. This can be summarized by classifying surgical techniques as first-generation techniques, second-generation techniques, and hybrid techniques.

**FIGURE 14-41.** Sagittal alignment in crouch gait includes excessive dorsiflexion at the ankle, following previous TALs, with flexion deformities at the hips and knees.

## First-Generation Techniques

**Principles:** Lengthening of proximal contractures (psoas, hamstrings) and correction of lever arm deformities. External support using ground reaction AFOs (GRAFOs) is required until adaptive shortening of the quadriceps occurs. This mechanism is more effective in growing children (59) (Fig. 14-43).

**Advantages:** Familiar techniques with acceptable morbidity.

**Disadvantages:** Incomplete correction in many patients leads to early relapse and recurrence. This is often related to the inefficiency of distal hamstring lengthening to achieve full and lasting correction of knee flexion contractures of >5 to 10 degrees.

**Current Role.** First-generation techniques are most effective in younger children with crouch gait caused by Achilles tendon lengthening with good proximal strength and selective motor control, knee flexion contractures of <5 to 10 degrees, good cooperation with a rehabilitation program, and compliance with the use of ground reaction AFOs.

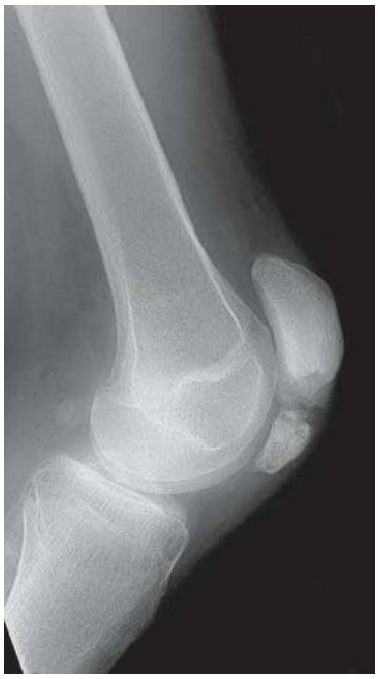


**FIGURE 14-42.** In crouch gait, there is frequently chronic overload of the extensor mechanism. In this 15-year-old boy, severe crouch gait was associated with bilateral fatigue fractures of the patellae.

Satisfactory correction of crouch gait using these techniques was reported with results maintained at 5 years (59). Increased knee extension was reported with healing of patellar fractures and resolution of knee pain. An important finding was that the soleus gradually shortened with a reduction in excessive dorsiflexion in the stance phase of gait, improved power generation at the ankle and the ability to ambulate without AFOs (59). Disadvantages include the slow recovery and reliance on ground reaction AFOs for at least 2 years after the surgery (59). Patella alta remains, even after the quadriceps has retensioned.

**Second-Generation Techniques.** First-generation techniques do not correct patella alta and distal hamstring lengthening is an inefficient method for the correction of knee flexion deformity of >10 degrees. More direct surgical approaches to the knee extensor mechanism insufficiency and the knee flexion deformity have been developed including distal femoral extension osteotomy (DFEO) with stable internal fixation, combined with shortening or advancement of the patellar tendon (198, 199) (Fig. 14-44).

**Principles:** Acute surgical shortening of the excessively long knee extensor mechanism combined with correction of knee flexion deformity by DFEO. Correction of all lever arm deformities is also required.

**Advantages:** Direct correction of the knee flexion contracture using extension osteotomy avoids weakening of MTUs (hamstrings) with more effective and predictable results. Direct correction of the patella alta and correction of quadriceps insufficiency by advancement or shortening of the extensor mechanism.

**Disadvantages:** These techniques are less familiar to many surgeons and are more invasive than first-generation techniques. They have a significant "learning curve," during which even experienced surgeons may report significant morbidity including neurovascular injury, loss of fixation, and incomplete correction.

**Current role:** DFEO combined with advancement or shortening of the extensor mechanism are powerful tools for the correction of severe crouch gait. Long-term studies are awaited to determine if the results will be durable (198, 199).

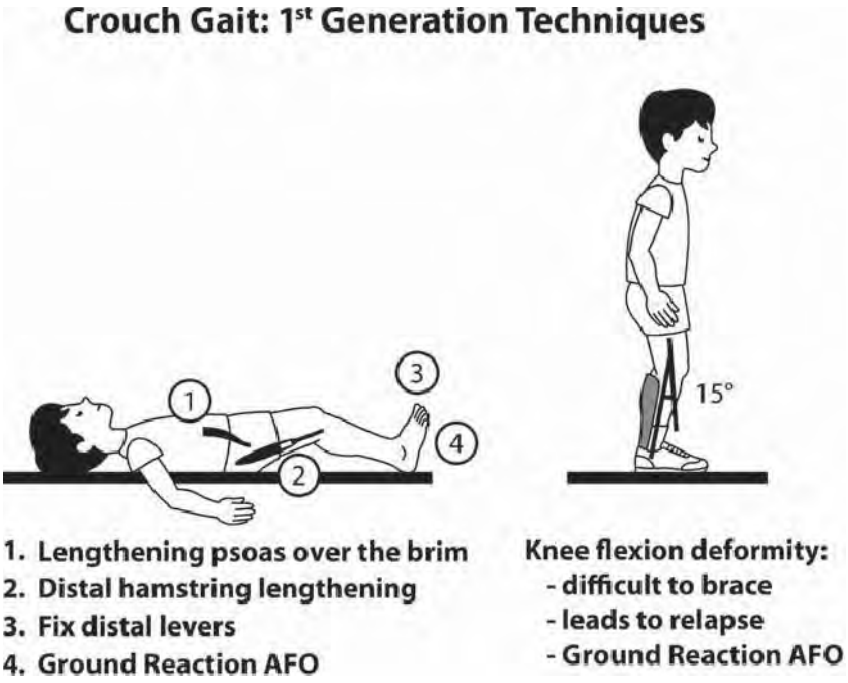



**FIGURE 14-43.** First-generation techniques for the management of crouch gait.

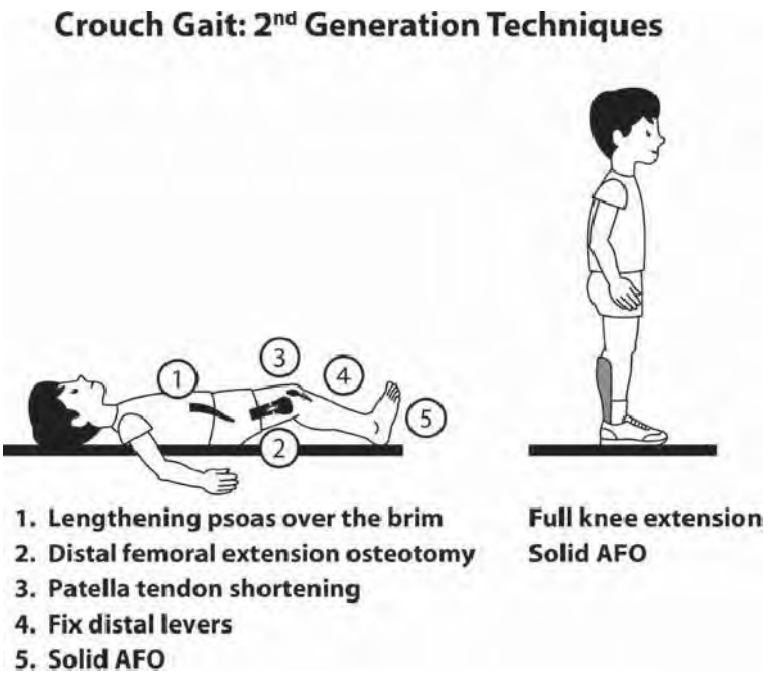



**FIGURE 14-44.** Second-generation techniques for the management of crouch gait.

**Hybrid Methods.** Some children develop severe crouch gait before skeletal maturity. In these children, a hybrid approach to surgical correction, consisting of a combination of hamstring surgery and distal femoral growth plate surgery, is showing promising results (168, 169, 176).

**Principles:** Distal hamstring lengthening/semitendinosus transfer deals with the spastic hamstring contracture (186). Guided growth deals with residual knee flexion deformity (174, 175).

**Advantages**: Semitendinousus transfer and guided growth are variations on familiar, existing techniques.

**Disadvantages**: Correction depends on growth and is gradual. Indications and outcomes not yet established in the literature.

**Current Role**: Preliminary reports have been published. Further reports and comparative studies are necessary.

Fractional lengthening of the semimembranosus, intramuscular tenotomy of the gracilis and transfer of the semitendinosus to the adductor tubercle can deliver improved knee extension, without weakening hip extension and without causing increased anterior pelvic tilt (188) (Figs. 14-45 and 14-46). Hamstring lengthening often reduces a knee flexion contracture but frequently fails to abolish it. In a typical case, a knee flexion deformity of 15 to 20 degrees will reduce to 5 to 10 degrees. The residual knee flexion deformity is enough to prevent correction of crouch gait and to impair the effectiveness of a ground reaction AFO (Fig. 14-45). Correction of

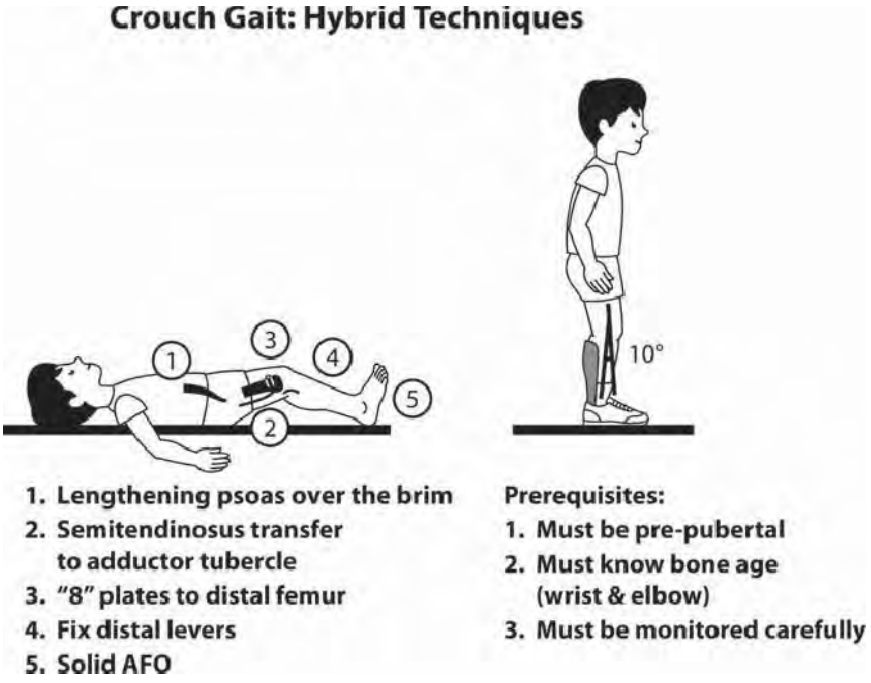



**FIGURE 14-45.** Hybrid techniques for the management of crouch gait.

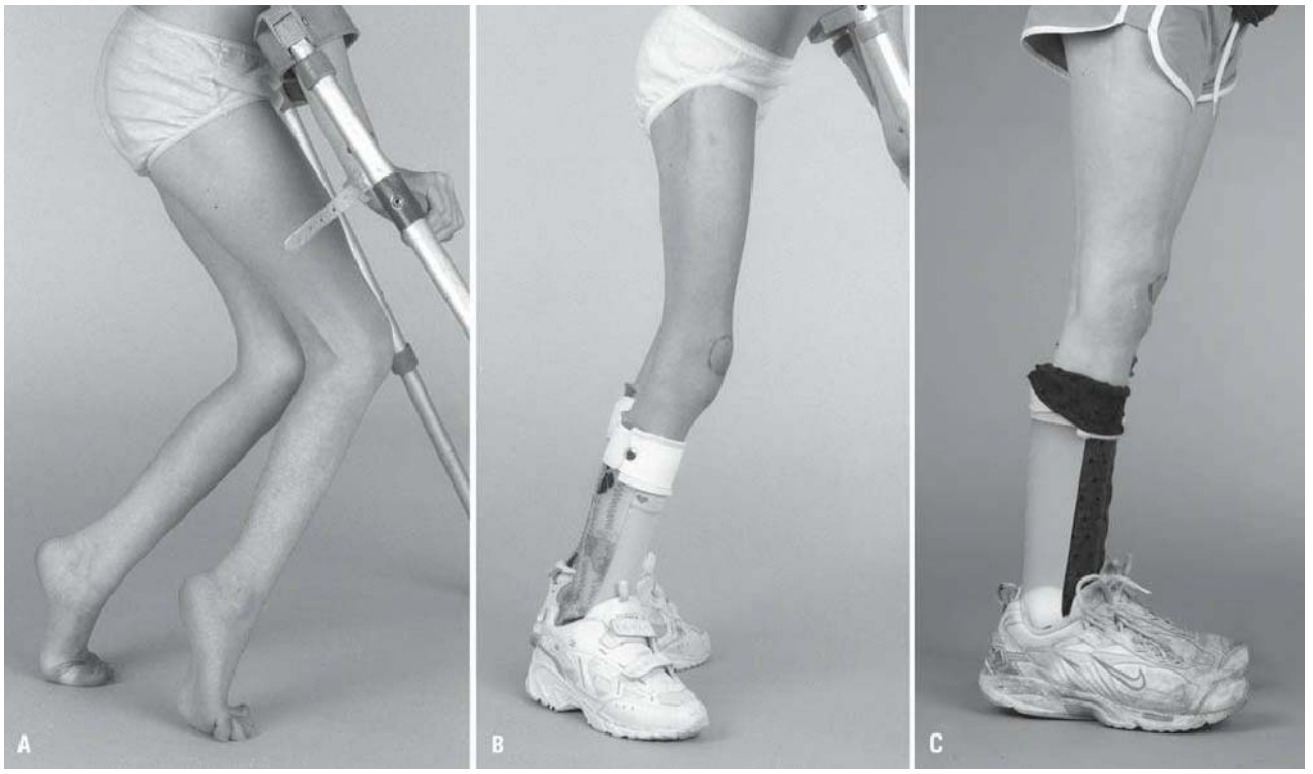



**FIGURE 14-46.** Severe jump gait alignment prior to SEMLS **(A)**. Improved alignment following SEMLS that included bilateral semitendinosus transfers and Strayer gastrocnemius recessions. Note the residual knee flexion deformities and external foot progression **(B)**. Full knee extension was achieved by a combination of guided growth, with staples to the anterior part of the distal femoral physis, and bilateral rotation supramalleolar osteotomies **(C)**. (From Young JL, Rodda J, Selber P, et al. Management of the knee in spastic diplegia: what is the dose? *Orthoped Clin North Am* 2010;41:561–577, with permission.)

the residual knee flexion deformity using growth plate surgery can be effective. This may be performed using staples or "8" plates placed in the anterior aspect of the distal femoral growth plate (175, 176) (Fig. 14-47). It is important to know both the chronologic age and the bone age of the patient. At least 2 years of remaining growth is necessary for clinically significant improvements in knee flexion contracture to occur.

Growth plate surgeries are consistent with early and full weight bearing and a rapid return of knee motion in most children (174, 175). However, some children can develop bursitis

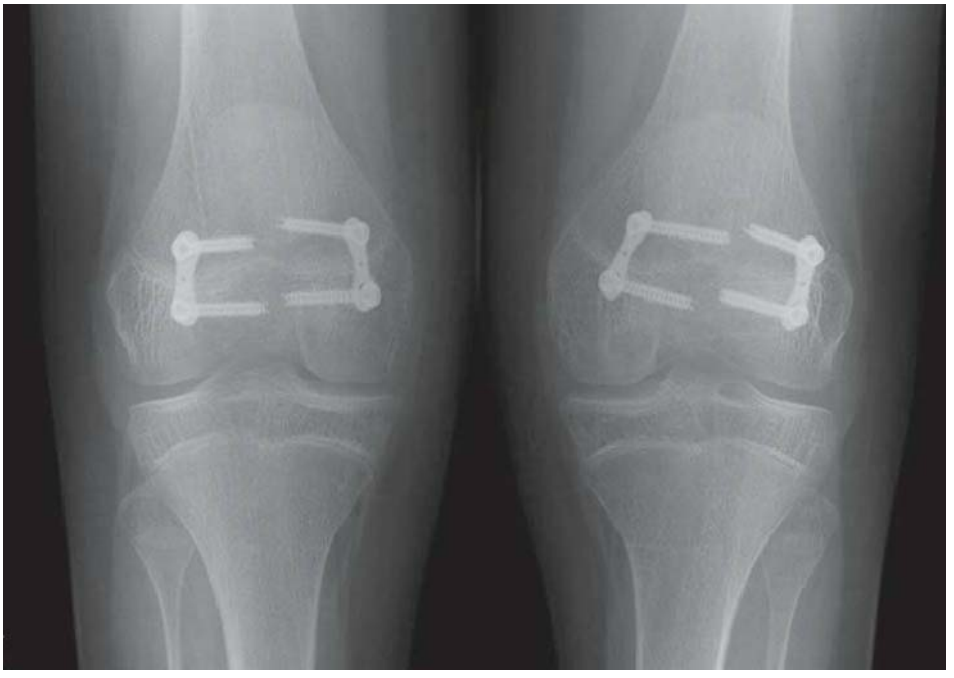


**FIGURE 14-47.** "8" plates for guided growth to correct residual flexion deformity at both knees. These are more prominent than staples and may cause bursitis and pain in some children.

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## Subtalar Fusion (Figs. 14-48 to 14-55)

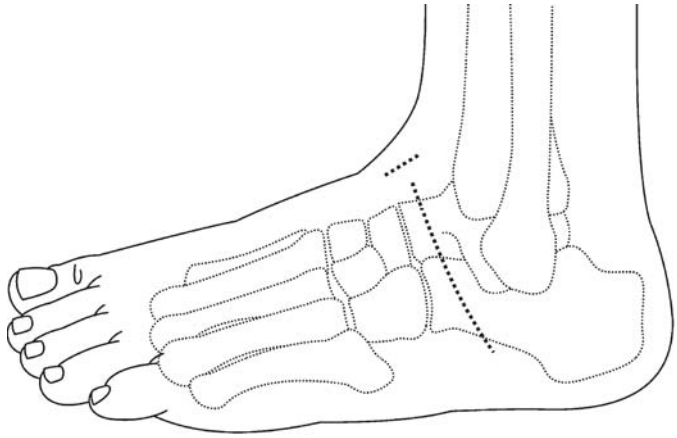


**FIGURE 14-48. Subtalar Fusion.** The skin incisions for subtalar fusion are as shown. They include a modified Ollier skin crease incision, which is ideal for exposing the subtalar joint, and a second small dorsal incision for the insertion of the cannulated screw. The operation is facilitated by having the patient in the supine position and using a pneumatic thigh tourniquet. The lower limb is prepared and draped free in the usual way. The flexibility and reducibility of the pes valgus is confirmed, and the margins of the sinus tarsi, the talonavicular joint, and calcaneocuboid joint are carefully palpated. The Ollier incision is made in a skin crease, extending across the midpoint of the sinus tarsi to give good access to the sinus as shown. At the inferior margin of the incision, the sural nerve and peroneal tendons need to be identified and protected. The dorsal incision for insertion of the screw is made by palpation of the anterior part of the talus and making a 1 cm incision in the midline of the talus. Blunt dissection is carried down to the anterior part of the talus avoiding injury to the extensor tendons and the neurovascular structures. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33: 431–438, with permission.)

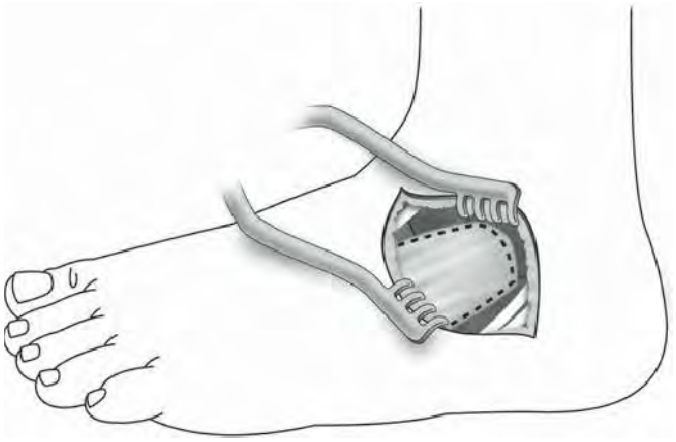


**FIGURE 14-49.** The skin incision extends down to the deep fascia overlying the extensor digitorum brevis (EDB) (which is often atrophic in neuromuscular patients) and a self retaining retractor is gently inserted. A distally based U-shape flap is outlined in the fascia and extensor brevis. This is carefully elevated with a combination of diathermy, cutting, and sharp dissection. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)

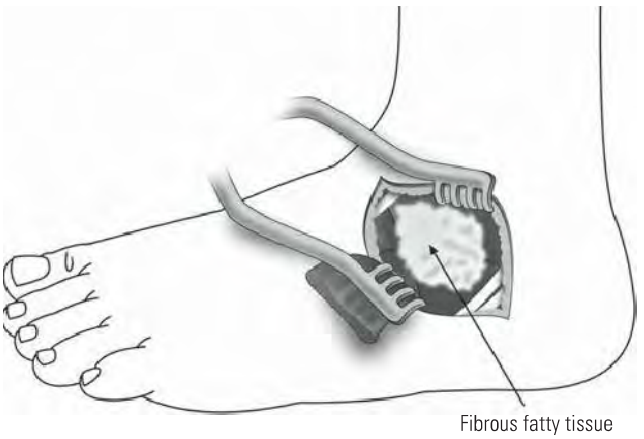



**FIGURE 14-50.** The elevated EDB is retracted from the sinus, and the fibrous fatty contents of the sinus are fully removed using a combination of sharp dissection, rongeurs, and best of all a hemispherical Coughlin reamer. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)

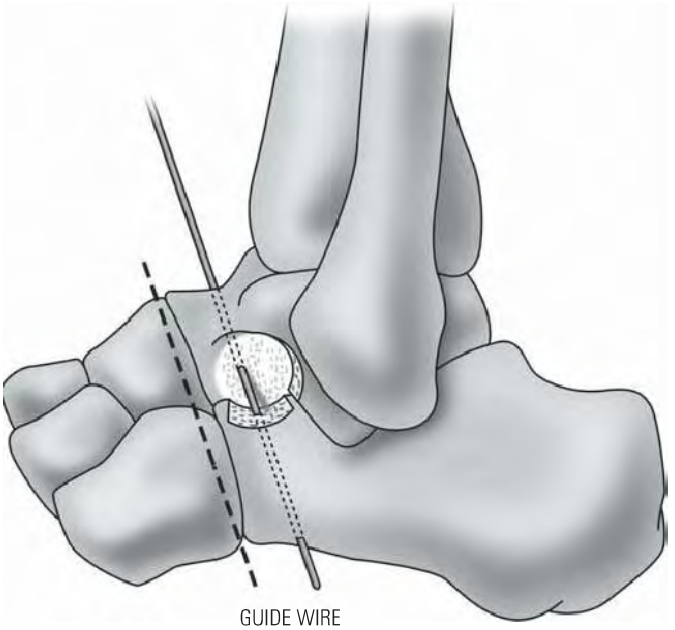



**FIGURE 14-51.** Once the sinus tarsi has been exposed and the contents cleared, the dorsal lateral peritalar subluxation (DLPTS) is manually reduced and a guide wire is inserted through the dorsal incision, across the depths of the sinus tarsi and into the anterior process of the os calcis, approximately parallel to both the talonavicular and calcaneocuboid joints. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)

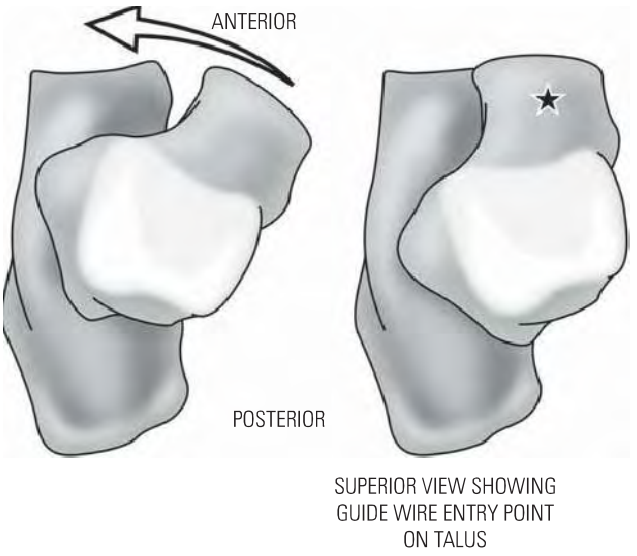



**FIGURE 14-52.** The reduction of the AP talocalcaneal subluxation is seen in Figure 14-53. The reduction needs to be checked both clinically and radiologically to ensure that full correction, but not over correction has been achieved. A useful guide is the navicular covering the head of the talus and the restoration of the medial arch of the foot. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)

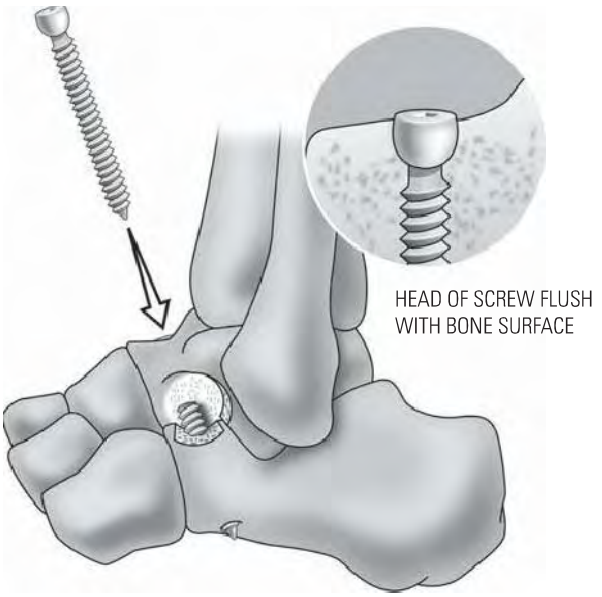



**FIGURE 14-53.** A fully threaded cannulated screw of appropriate length is now inserted across the guide wire to stabilize the reduction of the subtalar joint. It is very important to pay attention to the screw length. It should be fully counter sunk within the superior surface of the talus (to avoid impingement during ankle dorsiflexion). However, it must not protrude into the sole of the foot where it would cause pain and require early removal. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)

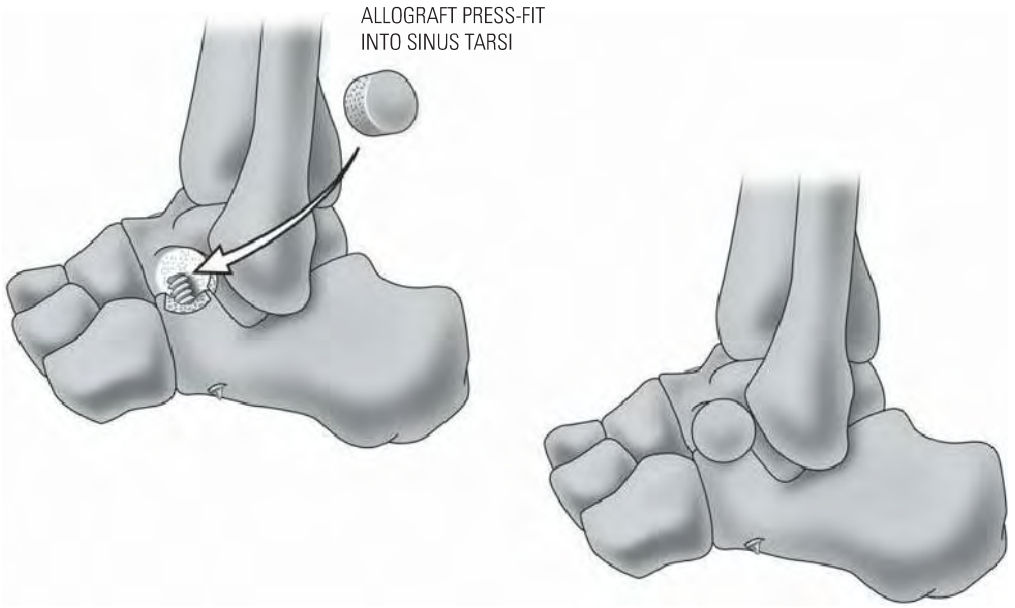
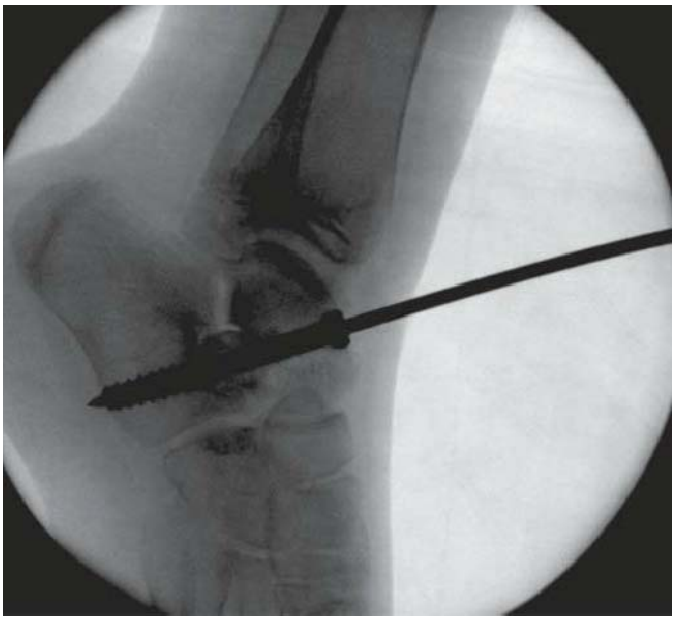



**FIGURE 14-54.** Following stabilization of the subtalar joint, the sinus tarsi is then grafted. The best options are a circular autograft (harvested from the patient's iliac crest) or a pre-cut circular allograft. Given the benefits of avoiding an additional incision to harvest an autograft, and the need for at least two grafts, our preference is a pre-cut circular cortico-cancellous allograft cut from an iliac crest. By using the Coughlin reamers to harvest this graft, it will be a secure press fit within the sinus tarsi. Given the circumferential contact between the cancellous bone of the allograft and the denuded margins of the sinus tarsi, rapid healing and incorporation is to be expected. In addition, the cortical surface of the iliac crest graft confers additional structural stability. Following irrigation, the incisions are closed in layers with interrupted nylon sutures to the skin. Following irrigation, the U-shaped flap of EDB is sutured back across the grafts covering the sinus tarsi. The incision is then closed in layers with fine interrupted nylon sutures to the skin. Using absorbable sutures risks wound separation and exposure of the allograft, which is in the relatively superficial position. We avoid this by using nylon sutures and leaving them in place for 3 weeks. The position of the graft and the fixation screw is checked on fluoroscopy prior to the application of a well padded below knee plaster cast. If a pneumatic tourniquet has been used or surgery has been prolonged it is wise to split the cast. We recommend non–weight-bearing for approximately 1 week until swelling has settled and then allow full weight bearing as tolerated. The combination of the press fit graft and the cannulated screw is stable for full weight bearing. After 2 to 3 weeks the first cast is removed, healing of the incisions is assessed, sutures are removed, and casting for a new solid AFO is done at this stage, if required. A second cast is applied for a further 4 to 6 weeks. It is often 6 to 9 months before complete healing and integration of the graft is noted on follow-up radiographs. However, bridging trabeculae can often be seen around the circular margins of the graft from as early as 3 months postoperatively. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)

**FIGURE 14-55.** The position of the screw and the graft is seen intraoperatively, and a clinical example of foot stabilization by this method is shown in Figure 14-26A, B. (From Shore BJ, Smith KR, Riazi A, et al. Subtalar fusion for pes valgus in cerebral palsy: Results of a modified technique in the setting of single event multilevel surgery. *J Pediatr Orthop* 2013;33:431–438, with permission.)



over the prominent "8" plates especially if they have a dystonic or a mixed movement disorder. Rehabilitation may be very slow in these children and in some, persistent pain and bursitis necessitate early removal of the hardware. The combination of correction of spastic hamstring contracture by semitendinosus transfer with fractional lengthening of the remaining medial hamstrings and dealing with the residual knee flexion contracture by growth plate surgery have not yet been reported in the literature. The place of these techniques in the correction of crouch gait is not yet established.

## GMFCS IV

1. **Between 6th and 12th birthday**
   Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors, and in the community, children are transported in a manual wheelchair or use powered mobility (13).
2. **Between 12th and 18th birthday**
   Youth use wheeled mobility in most settings. Physical assistance of one to two people is required for transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair (15).
3. **Risk of hip displacement:** 69%
4. **Mean femoral neck anteversion (FNA):** 40 degrees
5. **Mean neck shaft angle (NSA):** 155 degrees (49, 50)

**Movement Disorder.** The majority of children at GMFCS level IV have mixed tone, or "spastic-dystonia." In children and adolescents with severe generalized hypertonia, ITB by implanted pump offers the most reliable method of sustained tone reduction (86, 88). The position of the catheter in the intrathecal space can be adjusted to titrate the effects of the Baclofen in the upper limbs versus the lower limbs. Weakness of the trunk and paraspinal muscles may contribute to a postural kyphosis, and the incidence of scoliosis is high (30, 46).

Management goals, based on a realistic appraisal of long-term motor prognosis, include standing and assisted walking in early childhood. In later childhood, maintenance of comfortable sitting by detecting and treating early hip displacement and managing tone when necessary with ITB is important. Later, monitoring of spinal deformity with appropriate treatment and provision of custom seating are the most important issues.

**Preventive Hip Surgery GMFCS Level IV.** Lengthening of the hip adductors to achieve >50 degrees abduction in both hips, with the hips and knees extended, is an appropriate goal. This requires lengthening of the adductor longus and gracilis and sometimes a partial lengthening of the adductor brevis, depending on the age of the child and the degree of fixed contracture. Tenotomy of the iliopsoas at the lesser trochanter is helpful for hip flexion contracture. Lengthening at this level is more effective than lengthening at the pelvic brim. Weakness of hip flexion is of less consequence for a child at GMFCS IV. Phenol neurolysis of the anterior branch of the obturator nerve is safe and effective at the time of adductor release (90). Most reports of adductor releases with long-term follow-up report a high failure rate after adductor surgery in nonambulators (GMFCS IV and V) (196, 197).

**Reconstructive Hip Surgery GMFCS Level IV.** The most common indication for reconstructive surgery is a persistently high MP after adductor releases. If the MP remains >40%, at >12 months after adequate adductor releases and the child is >4 years, reconstructive surgery is indicated (191, 195). Reconstructive surgery is also advised as the index surgery in children >8 years presenting with MP >40% (191, 193) (Fig. 14-56).

Reconstructive hip surgery poses a major challenge to children with CP and general health should be optimized before embarking on bilateral hip surgery. Nutrition and respiratory status should be optimized before surgery (193). Potential sites for infection, including feeding tubes, chest, and bladder, should be screened. Many children have low-grade, iron deficiency anemia and may benefit from iron supplements. The single most common complication after reconstructive hip surgery is the exacerbation of chronic constipation that can be detected and corrected prior to hip surgery (56).

Reconstructive surgery consists of three main components: **Adductor Releases:** Lengthening of the soft tissues to ensure an adequate range of passive hip abduction (>50 degrees) is the first step in reconstructive surgery. For those children who have recurrent displacement after previous adductor releases, a revision adductor release at the time of bony reconstruction is always required (189, 190, 192). Revision adductor releases have a higher complication rate than first-time surgery. Meticulous hemostasis, suction drainage, and sealed waterproof dressings may reduce the rate of hematoma formation and deep infection. **Femoral Osteotomy**: Correction of the abnormal femur is achieved by varus derotation, shortening osteotomy of the proximal femur. Given that these children never ambulate independently and most are walkers for a relatively short period, correction of the NSA to about 100 degrees is an appropriate goal with reduction of anteversion to about 10 degrees (166, 169). The osteotomy should be performed at the intertrochanteric level. Most children at GMFCS IV have true coxa valga and will need excision of a 1 to 3 cm medially based wedge or trapezoid from the femur (49). The wedge includes the lesser trochanter and the psoas insertion. Shortening of the proximal femur is an effective way to reduce soft-tissue tension and restore range of motion and symmetry about the hips. In windswept deformities, symmetry can be achieved by a combination of soft-tissue releases and adjusting the amount of rotation in each osteotomy (191).

Stable internal fixation with the AO-ASIF 90- or 100-degree blade plate is the preferred fixation device (166). Older, two part fixation devices are not reliable but the newer proximal femoral locking plates are a good option, particularly in osteopenic bone. Hip spicas should be avoided, but poor bone quality may necessitate short-term use in a minority of children (191).

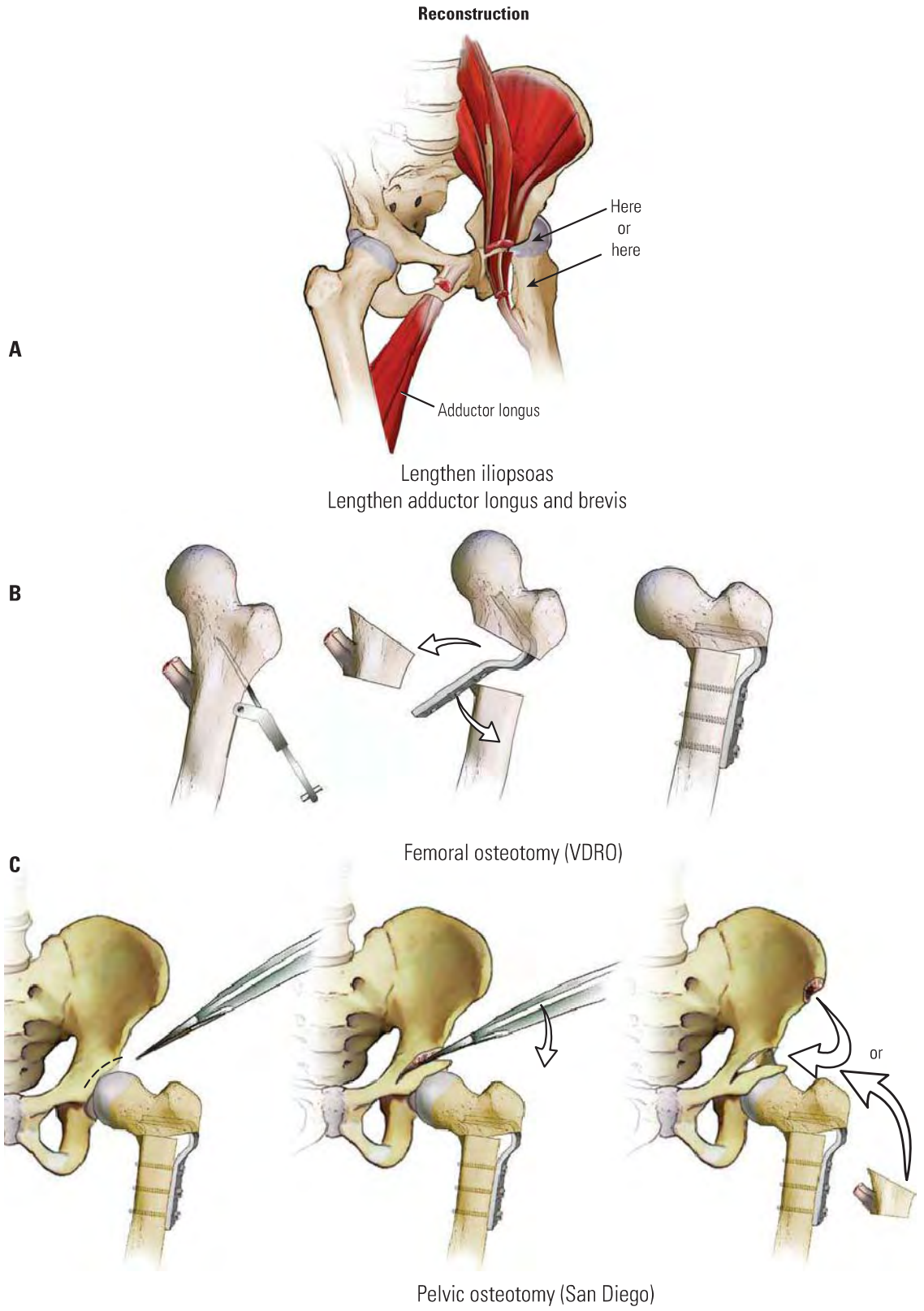



**FIGURE 14-56.** The essential three components of reconstructive hip surgery include the following: **A:** Primary or revision adductor and psoas lengthening. **B:** Femoral varus derotation osteotomy with appropriate shortening, internal fixation with a fixed angle blade plate or a proximal femoral locking plate and providing additional graft for **C:** Pelvic osteotomy of the San Diego type.

**Pelvic Osteotomy**: Significant acetabular dysplasia should be corrected by a pelvic osteotomy at the time of bony reconstruction. Innominate osteotomy is contraindicated in CP, and salvage procedures such as the Chiari or shelf procedures are not good options. A curved osteotomy, close to the acetabular margin as popularized by the San Diego and Du Pont groups, is by far the best option for the majority of younger children (172, 200, 201). The San Diego and Du Pont surgeons have refined the direction of the cut, the opening of the osteotomy site, and the stabilization with "press fit" bony wedges to make the procedure effective and reliable in CP. Older children and teenagers may benefit from a triple pelvic osteotomy or a periacetabular osteotomy (202, 203).

**Postoperative Care.** At the end of the reconstruction, the hip and the fixation should be stable and a hip spica should not be required (200). More preventable morbidity comes from hip spica immobilization than from the surgery. Expert pain management, nutritional and respiratory support should continue well into the postoperative period. General complications include respiratory infections, exacerbation of constipation, emesis, and weight loss. Surgical complications of reconstructive hip surgery include avascular necrosis, infection, nerve palsy, loss of fixation, periprosthetic fracture, and recurrent hip displacement. Windswept deformity, leg-length inequality, and heterotopic ossification are also seen.

**Outcomes:** One-stage correction of hip displacement, as described above, is a very effective and reliable method of stabilizing the severely subluxated or dislocated hip in CP as reported in several large series (172, 200, 201). The radiologic status of most hips remains satisfactory in the short and longer terms. Pain is prevented or relieved and sitting tolerance is usually improved (193). Prominent hardware (blade plates) are usually removed about 12 months after surgery. Radiologic monitoring of hip development should continue until skeletal maturity, at least. The biggest threat to hip status after successful hip reconstruction is progression of scoliosis and pelvic obliquity. It is very difficult to maintain hip stability on the high side of an oblique pelvis (191).

**Special Circumstances.** Anterior dislocations are rare and present clinically with extension posturing, restricted hip flexion, and inability to sit comfortably. Improved anterior cover, using a Pemberton osteotomy, is required as part of the reconstruction (191). Windblown hips require a very careful analysis of movement disorder, soft-tissue contractures, and a tailored asymmetric surgical prescription. In children with severe dystonic posturing, an ITB pump may be required before the hip reconstruction. The abducted hip may need an abductor release and a VDRO. The adducted hip may require an adductor release, a VDRO, and San Diego pelvic osteotomy (191).

**Lower Limb Surgery at GMFCS Level IV.** Flexion deformities at the hip and knee are very common as well as deformities at the foot and ankle, especially ETT and pes valgus. Hallux valgus and dorsal bunions are also quite common. In early childhood, if the child is enjoying assisted ambulation, it may be appropriate to employ some of the procedures described for children at GMFCS levels II and III, to help the child stand and achieve the goal of limited walking. However, parents, carers, and therapists should recognize that sustained ambulation into adult life is not achievable (15). Therefore, extraordinary measures to maintain ambulation are not indicated. Despite cospastic stiffness at the knee, transfer of the rectus femoris is contraindicated at this GMFCS level because it does not work and may weaken knee extension, which is needed for transfers. More invasive measures such as DFEO and patellar tendon shortening for crouch gait are also contraindicated because with or without these measures, ambulation will eventually be lost (15).

Managing deformity of the foot and ankle is important to allow bracing, the wearing of normal shoes, and for the feet to be able to rest on the foot rest of a wheelchair. Calcaneus is very common after gastrocsoleus lengthening and it is best to manage spastic equinus in these children by injections of BoNT-A or by a modest gastrocnemius recession (95). Correction of severe ETT by supramalleolar osteotomy of the tibia may be necessary for positioning on the wheelchair footrest. Stabilization of the foot for pes valgus is more reliably achieved by a subtalar fusion than by os calcis lengthening (133). Correction of hallux valgus and dorsal bunion by soft-tissue balancing and fusion of the first MTP joint is effective for deformity correction, pain relief, and comfortable shoe wear (133).

## Spinal Deformities at GMFCS Level IV

**Kyphosis.** Postural kyphosis is very common at GMFCS level IV due to paraspinal muscle weakness and impairments of posture and balance. Dorsal kyphosis remains flexible and corrects in prone lying in early childhood. It is best managed by appropriate seating and occasionally by the use of thoracolumbosacral orthosis (TLSO). With advancing age, the thoracic kyphosis and the secondary cervical lordosis may become more fixed. Cervical pain in adults with CP is common. Spinal reconstructive surgery may be indicated to improve alignment, minimize pain, and promote comfort in sitting.

**Scoliosis.** Progressive scoliosis is common at GMFCS level IV but differs from that seen at GMFCS level V (30). It tends to start a little later, is not so rapidly progressive, and the outcomes of surgery are better because medical comorbidities are fewer and less severe. Pelvic obliquity and hip disease are less severe. Management of scoliosis will be considered in more detail in the section on GMFCS level V. The question of orthotic management of scoliosis at GMFCS level IV is frequently raised because parents, caregivers, and therapists may wish to avoid the risks of spinal fusion surgery. Studies have suggested that bracing rarely prevents progression of spinal deformity at GMFCS level IV although it may slow down progression (204). Previous studies have not subdivided children according to GMFCS level. From the point of view of parents, it is important to provide information about the limited benefit of bracing. Even when surgery is likely to be required in the near future, it may be helpful for parents to feel that they have tried nonoperative measures. Curve progression should be monitored because it can be very rapid.

## GMFCS V

1. **Between 6th and 12th birthday**
   Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements (13).
2. **Between 12th and 18th birthday**
   Youth are transported in a manual wheelchair in all settings. They are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology (15).
3. **Risk of hip displacement:** 90%
4. **Mean femoral neck anteversion (FNA):** 40 degrees
5. **Mean neck shaft angle (NSA):** 163 degrees (49, 50)

**Seating Requirements GMFCS Levels IV and V.** Comfortable sitting requires a straight spine, over a level pelvis, with flexible hips that are in joint. Hip flexion should be >90 degrees and extension to within 30 degrees of full extension, with 20 to 40 degrees of abduction at each hip and no fixed abduction or "windswept" deformity. Flexible knees with little fixed flexion and no extension deformity and plantargrade feet that will rest comfortably on the foot plates of wheelchairs are also important. At GMFCS level IV, the wheelchair is a "total body orthosis" requiring prescription, fitting, and maintenance by an expert team (205). The ability to transfer independently in and out of specialized seating is crucial for the option of young adults to be able to live in a group home setting (Fig. 14-57).

**The Chair Back.** The back should support the patient's trunk from the shoulders to the pelvis and be wide enough to accommodate trunk and lateral supports that are often needed. The chair back should be firm enough to provide support and soft enough for comfort throughout the day. The chair back requires the ability to recline as this provides additional spinal support. A reclined position may inhibit extensor thrust as well as remove some of the demands on the paraspinal muscles to maintain the upright position (205) (Fig. 14-57A,B).

**The Seat.** The feet should reach the foot rests and the seat should be wide enough to accommodate a central pommel and lateral supports for the control of "windswept" hips. The seat should support the thigh segments that may be unequal in the windswept deformity. The seat should be firm enough to provide support and soft enough for comfort. Customized contouring for windswept deformity can be very helpful. The use

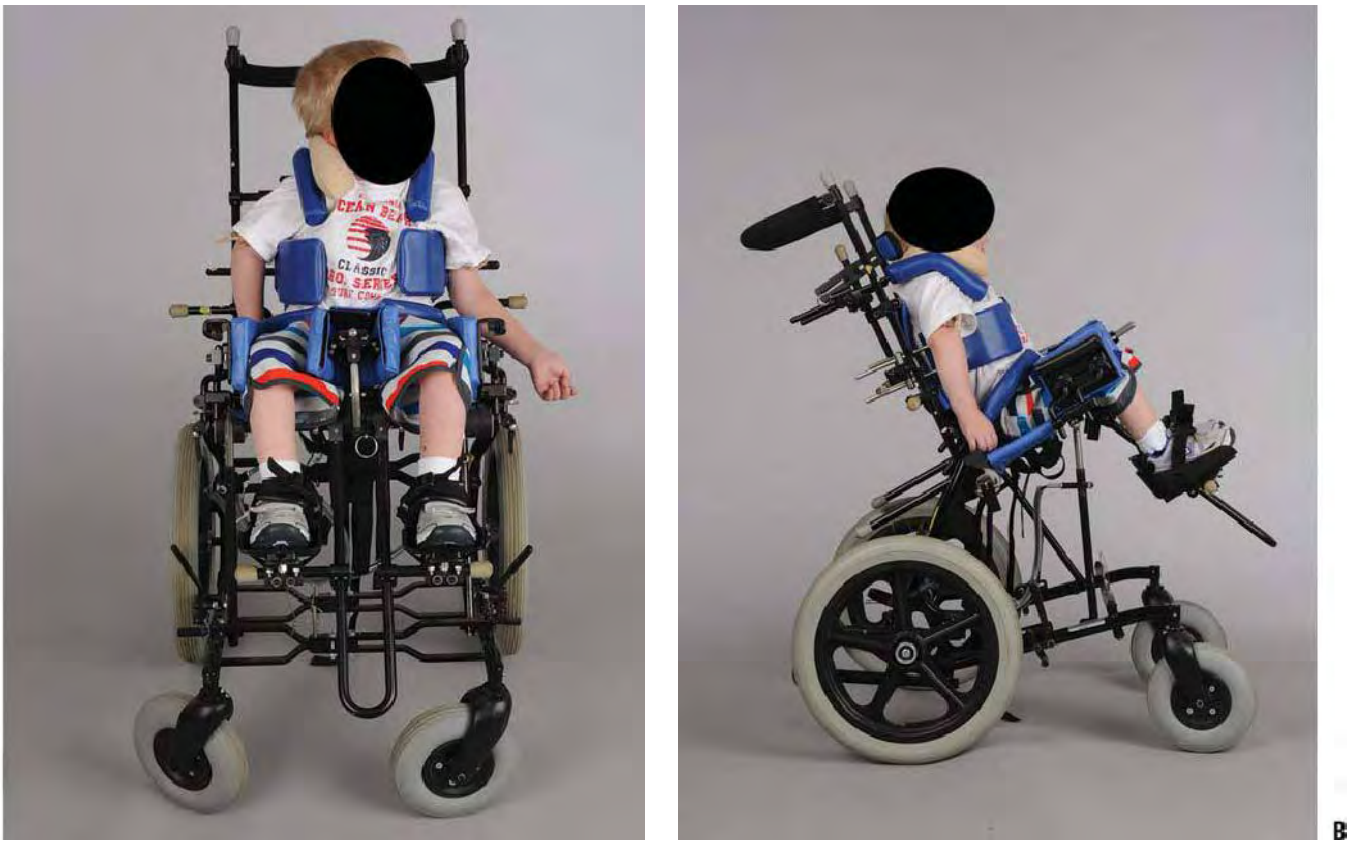



**FIGURE 14-57. A and B:** Modular, adjustable seating for a 5-year-old child with severe CP, GMFCS level V, who lacks head control and sitting balance. Note the ability to provide supports for the neck, shoulders, trunk, and legs. The chair is partially reclined to reduce extensor thrust and the feet are well supported by the foot plates. Maintenance of comfortable sitting at GMFCS level V frequently requires management of hypertonia, reconstructive hip surgery, and spinal fusion surgery.

of pressure mapping to define areas of high contact pressures can be helpful in problem children (205).

**The Foot Rest.** The foot rest should support the entire foot in a plantargrade position and be designed to swing out of the way during sitting and during transfers in and out of the chair. Given the high incidence of spastic dystonia, foot restraints, supplemented by straps at the knee level, may be helpful to avoid one or both lower limbs escaping from the chair and risking injury during transport. Additional supports or restraints may be helpful at the level of the head and neck, trunk, pelvis, knees, foot, and ankle (205).

**Functional Mobility.** The ability to move the chair easily in and out of an adapted vehicle is an important consideration for many families. Independent control by the patient using hand controls may add significantly to the patient's self-esteem, quality of life, and independence.

**Preventive and Reconstructive Hip Surgery: GMFCS V.** Preventive surgery has a high failure rate and should be considered to be a temporizing measure for most children at GMFCS V. Reconstructive surgery is technically easier and probably more successful in older children. Scoliosis and pelvic obliquity are so prevalent that hip and spine management should be considered together (193). Windswept deformities are more common and more severe at GMFCS V. If the hips are windswept, a more extensive release on the adducted side is required sometimes combined with phenolization of the anterior branch of the obturator nerve or a neurectomy. If there is fixed abduction contracture, this should be addressed by release of the hip abductors. Bilateral femoral VDRO should be performed with shortening and appropriate derotation, taking into consideration the patient's posture while awake and any torsional deformities identified clinically or confirmed by CT.

**Salvage Surgery GMFCS Level V.** The degree of femoral head deformity and acetabular deformity should be carefully evaluated in the context of the child's health, functioning, and life expectancy. The principal symptoms from neglected hip displacement are pain, which is reported to occur in between 10% and 90% of cases (191, 193). Fixed deformity, especially the windswept deformity, is also a major impediment to comfortable sitting and care. None of the salvage options that are available are reliable and predictable. The need for salvage surgery is best avoided by early hip surveillance and appropriately timed preventive and reconstructive surgery. Before considering salvage surgery, consultation with the multidisciplinary team to optimize the patient's general health is very important. Referral to an appropriate pain management service is important as a number of teenagers can be managed nonoperatively, in the short term. Reflex spasms of the hip adductors and flexors are almost always part of the pain problem in dislocated hips. Short-term symptomatic relief can often be achieved by injecting the hip joint with bupivicaine and corticosteroid and injecting the hip adductors and flexors with BoNT-A (123, 124). Open releases of the contracted hip adductors and phenolization of the obturator nerve may also help. These interventions have been reported to give short-term pain relief, but no long-term studies have been reported (123, 124).

It is also important to optimize tone management prior to salvage surgery. If an ITB pump is an appropriate choice for the child and accepted by the parents, this should be done before hip surgery (86). The marked reduction in tone around the hips may reduce pain and postural deformities to a degree that salvage surgery is not required. If salvage surgery is still necessary, it is much more easily performed in the context of global tone reduction afforded by the ITB pump (88). During salvage surgery, the pump can be reprogrammed to increase the amount of Baclofen available to the child in the immediate postoperative period resulting in reduced postoperative pain and a reduction in the need for narcotic analgesia. During surgery, the pump should be protected from hematogenous infection by perioperative antibiotics.

There is no single, reliable salvage surgery for the painful dislocated hip at GMFCS level V. The Castle procedure is an extraperiosteal resection of the entire proximal femur, below the lesser trochanter, with vastus lateralis and rectus femoris sewn over the end of the femur and as much hip capsule and gluteal muscle as possible interposed between the femoral stump and acetabulum (206). Postoperative care has included skin traction, skeletal traction, external fixators, hip distracters, hip spica casts, and bracing. Postoperative complications include pneumonia, decubitus ulceration, deep infection, wound breakdown, and death. Pain relief is usually delayed and unpredictable. After the Castle procedure, adolescents may take a year to show improvement in pain and there are high rates of heterotopic ossification, proximal migration, inadequate pain relief, and the need for revision surgery (207, 208). The family and caregivers must know that the hip will be unusually "floppy" and that weight bearing will no longer be possible.

The McHale combination of femoral head resection combined with valgus osteotomy is more stable, has a reduced risk of heterotopic ossification and less proximal migration than the Castle subtrochanteric resection (209, 210). However, impingement may occur between the lesser trochanter and the acetabulum or pelvic wall. Valgus osteotomy, without femoral head resection, has been reported in a recent study to have good pain relief in 24 patients followed for a mean of 44 months (211).

Interposition arthroplasty with a variety of devices has been reported in several small series with relatively short follow-up (212). Prophylaxis of heterotopic ossification with preoperative radiation or the administration of nonsteroidal anti-inflammatory drugs should be considered. Radiation may increase discomfort and affect healing. Nonsteroidal anti-inflammatory drugs are risky in this population but so is the development of severe heterotopic ossification.

Arthrodesis is very effective in terms of pain relief but is indicated only in unilateral hip disease in patients with dystonia in a hemiplegic distribution (213). Conventional total hip arthroplasty (THA) is very effective but is only indicated in a small subset of ambulant patients with well-controlled movement disorders (213). Metal on metal, resurfacing arthroplasty combined with proximal femoral derotation and shortening has a wider range of indications because the risk of dislocation is lower than with

conventional THA. Resurfacing arthroplasty can give good pain relief and improve function (214) (Figs. 14-58 to 14-60).

**Lower Limb Surgery GMFCS Level V.** Maintaining the ability to wear normal shoes and place the feet on a foot plate of a wheelchair is a basic but important goal. Deformities around the foot and ankle are often severe. Lengthening of the gastrocsoleus is contraindicated because it almost invariably leads to the subsequent development of a fixed calcaneus deformity. In the younger child, neurolytic blocks and AFOs are appropriate to maintain the feet in a plantargrade alignment. In the older child, soft-tissue surgery combined with bony stabilization for severe deformities may be appropriate. Triple arthrodesis may be necessary to manage problematic equinovarus or equinovalgus foot deformities. Occasionally, a severe calcaneovalgus deformity can be managed by a tibiotalocalcaneal fusion, eliminating all ankle motion and motion of the hindfoot, but resulting in a plantargrade foot that is easily accommodated within a normal shoe and on the wheelchair foot rest (215).

Dorsal bunion in severely involved children is common and often becomes symptomatic in the teenage years. Management requires soft-tissue rebalancing and a fusion of

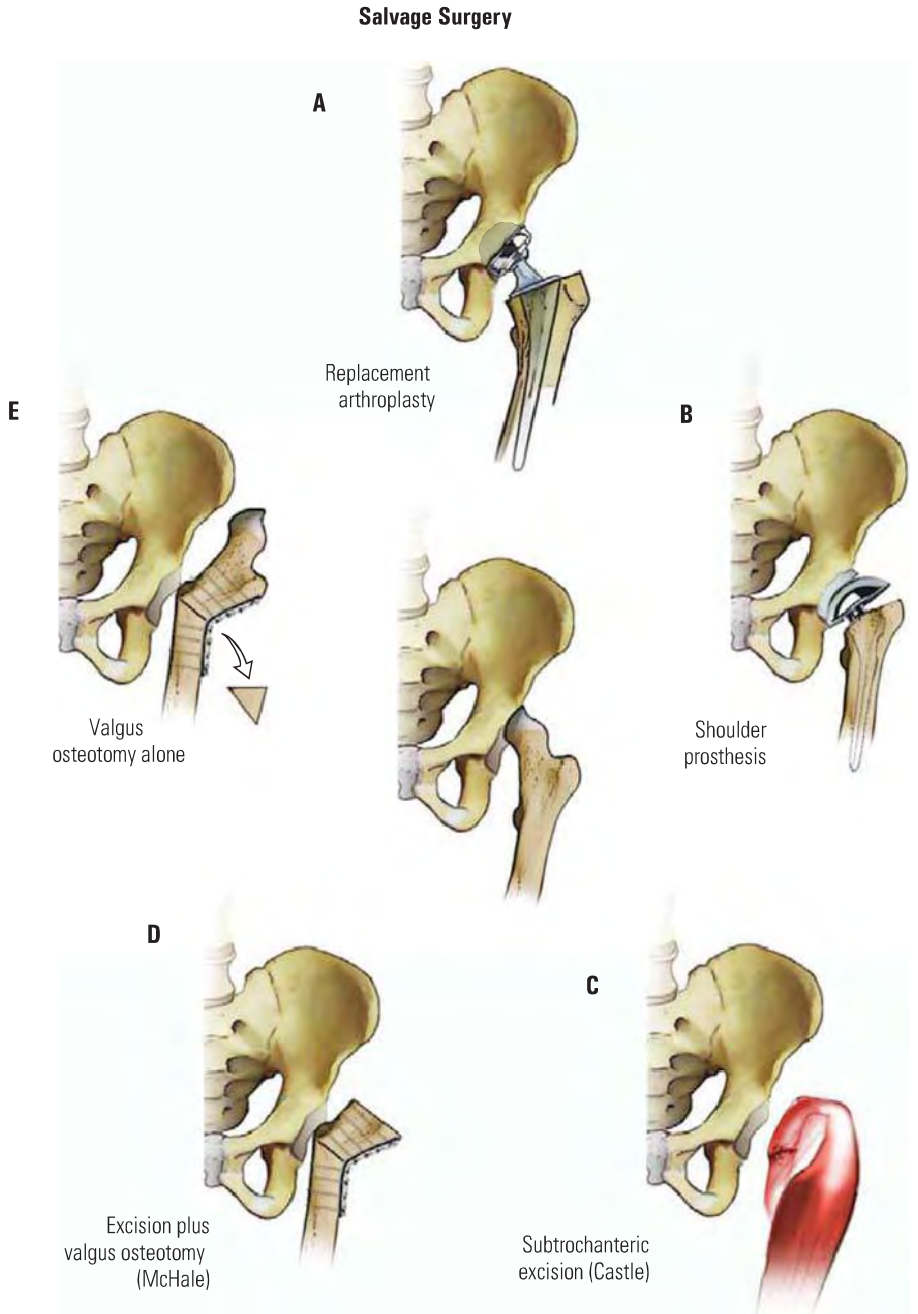



**FIGURE 14-58.** Options in salvage surgery for the hip in CP include the following: **A:** Replacement arthroplasty. **B:** Interposition arthroplasty with a shoulder prosthesis. **C:** Subtrochanteric excision of the proximal femur as described by Castle. **D:** Limited excision of the proximal femur along the intertrochanteric line combined with a valgus osteotomy as described by McHale. **E:** Valgus osteotomy without resection of the femoral head.

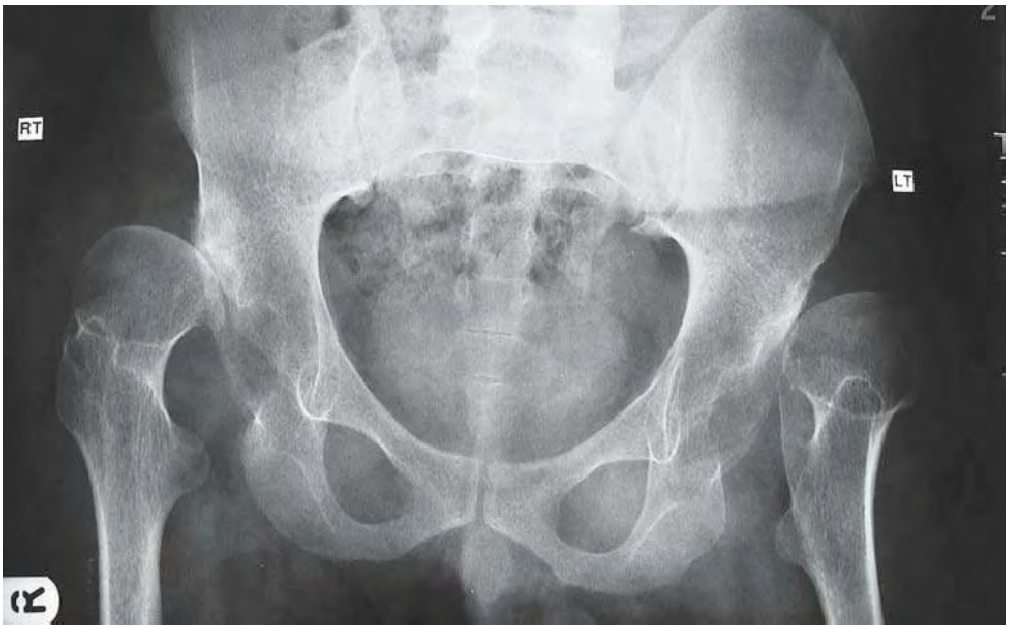



**FIGURE 14-59.** Bilateral painful hip dislocations in a young adult with CP, GMFCS level IV. Note the high riding dislocations, contact between the femoral head and pelvis, and severe acetabular dysplasia.

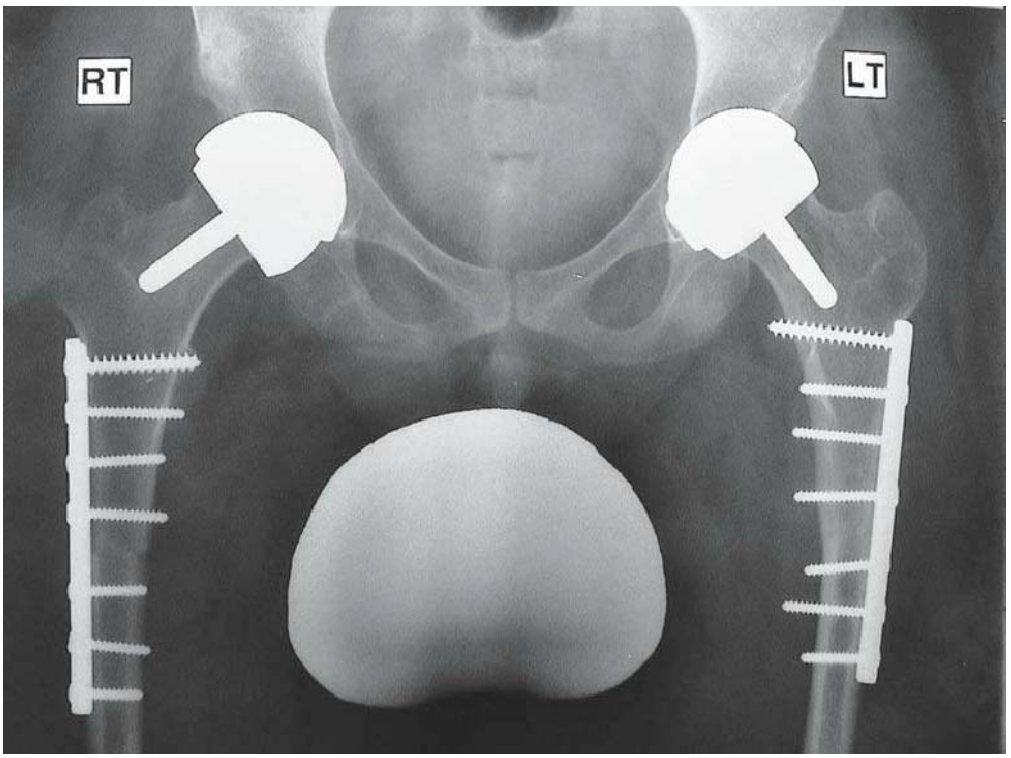



**FIGURE 14-60.** Post bilateral reconstruction including:

1. Bilateral femoral shortening with derotation and DCP fixation
2. Metal on metal resurfacing (Mr John O'Hara, Birmingham, England).

This type of reconstruction combines extensive soft-tissue lengthening, by virtue of the femoral shortening and the stability of large diameter metal on metal resurfacing. It effectively extends the range of joint arthroplasty to one of the most difficult patient populations (214).

the first MTP joint. One cm should be excised from the tibialis anterior, the remaining dorsiflexors should be lengthened, and the first MTP joint fused and fixed with a dorsal plate. The FDB and FHL will then act as a depressor of the first ray, as they act across a rigid first MTP joint (133).

## SPINAL DEFORMITY AND SCOLIOSIS SURGERY GMFCS LEVEL V

**Natural History.** Spinal deformity affects approximately two-thirds of children at GMFCS level V but is variable in its onset, severity, progression, and effects (216). Dorsal kyphosis is very common in younger children with weak paraspinal muscles and is best managed by having the wheelchair seat slightly reclined, the use of chest straps and occasionally a TLSO (204, 205). Kyphosis in the lumbar spine is less common and may be caused by tight hamstrings. Proximal hamstring recession at the time of adductor releases may be beneficial. Lumbar lordosis is much more common and is frequently related to hip flexion contractures. Lengthening of the hip flexors before the lordosis becomes fixed may help. An ITB pump may be helpful in the management of muscle imbalances about the hip that are contributing to lumbar lordosis or kyphosis (86, 88) (Figs. 14-61 and 14-62).

**Scoliosis.** Scoliosis in children with CP is particularly prevalent in nonambulant children, GMFCS levels IV and V. Curves in ambulant patients are uncommon, are more likely to be idiopathic in type, and are managed accordingly. The cause of scoliosis in CP remains speculative, but spasticity, dystonia, muscle imbalance, weakness, postural impairment, and immobility have been suggested as contributing factors. SDR may be associated with kyphosis, lumbar lordosis, spondylolysis, and spondylolisthesis (217). The high rate of scoliosis in CP and the lack of controls make the interpretation of this association difficult. As with hip displacement, there is growing evidence

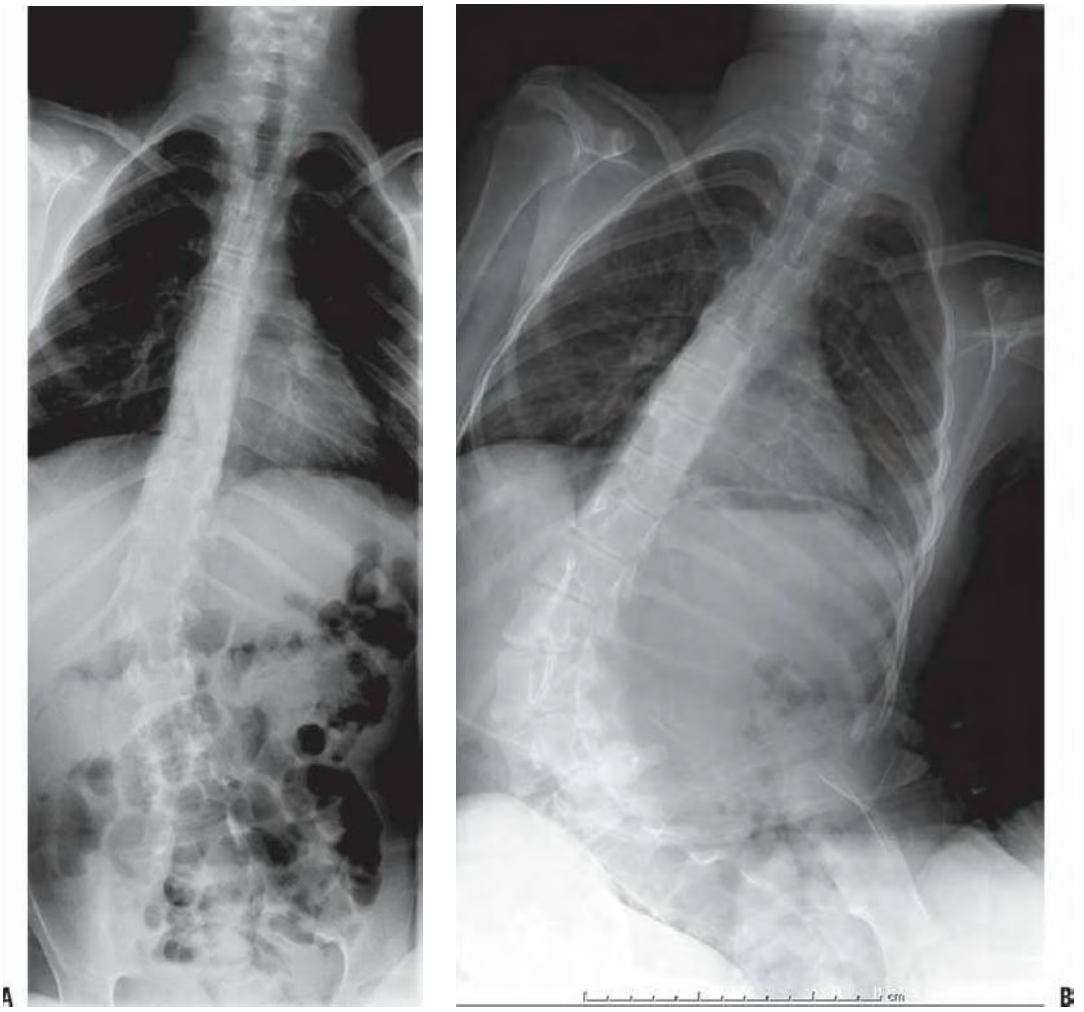



**FIGURE 14-61.** **A and B:** Scoliosis may progress very rapidly during the pubertal growth spurt in children with CP, especially at GMFCS level V. These two radiographs were taken only 14 months apart, at age 13 and just over age 14 years. Note the severity of the curve, and its extension into the sacrum and pelvis with marked pelvic obliquity.

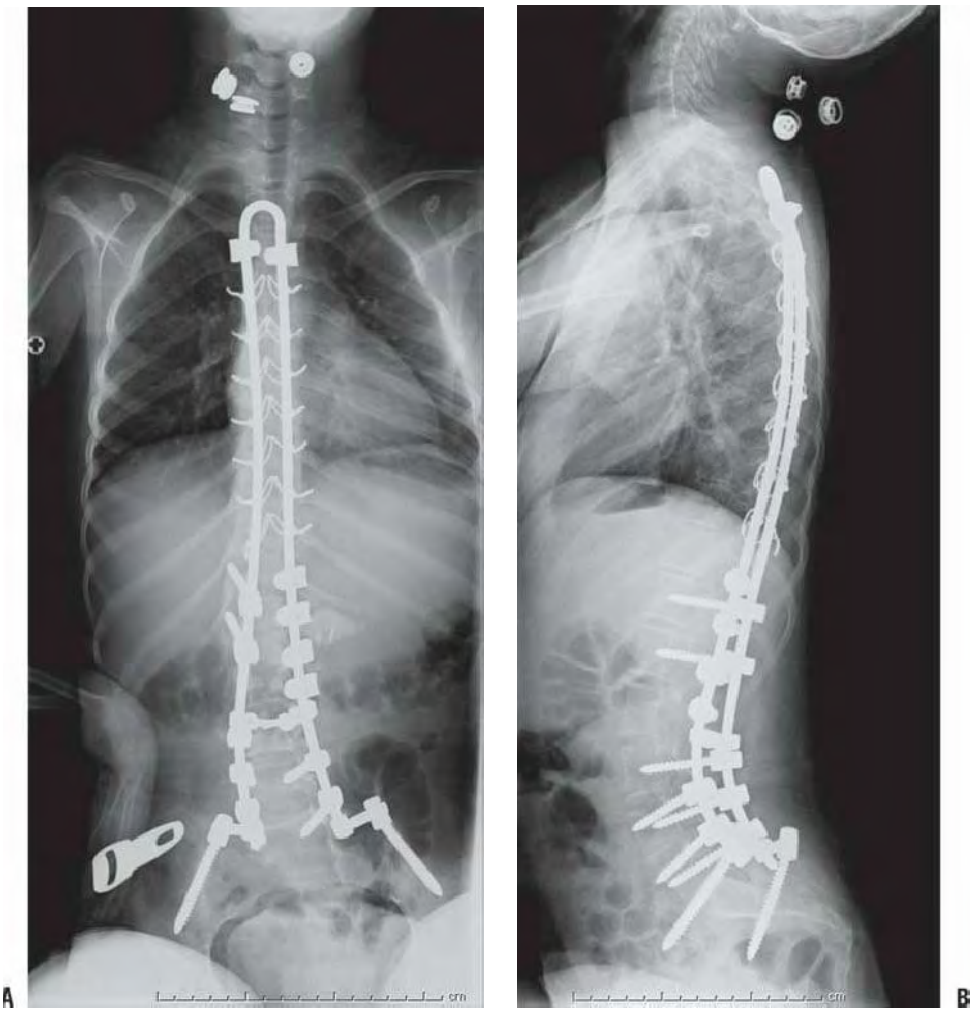


**FIGURE 14-62.** **A and B:** Long posterior instrumented fusion from T3 to the pelvis using a unit rod construct, and a combination of segmental fixation techniques, combined with iliac screw fixation to the pelvis in a manner similar to the Galveston technique.

that GMFCS level is the single strongest predictor of spinal deformity in children with CP (50).

**Natural History of Scoliosis in Cerebral Palsy.** The long "C"-shaped CP curves present earlier in childhood than idiopathic curves, are more likely to be progressive, progress more rapidly, and may continue to progress after skeletal maturity if the curve is more than 40 degrees (218–220). The curves may be convex to the left, which is rarely seen in idiopathic scoliosis. Scoliosis may present as early as age 6 to 8 years (and occasionally even younger). In the initial stages, curves are flexible. They progress faster than idiopathic curves. The rate of progression accelerates when the curve reaches 40 to 50 degrees and especially as the child enters the pubertal growth (Fig. 14-61A,B). The speed of progression may catch parents, pediatricians, and physical therapists unawares. Some adolescents present with acute loss of sitting ability, especially when there is pelvic obliquity and windswept hips. In the space of 1 to 2 years, it is possible for a curve to progress from a moderate flexible curve, easily correctible in single-stage posterior surgery with moderate risks, to a severe rigid curve requiring anterior and posterior surgery, with substantially increased risks of morbidity and mortality.

Neurologic deterioration, related to shunt malfunction, is also associated with rapid curve progression (216). Recent longitudinal studies of gross motor function in CP confirm that deterioration during the second decade is common, at GMFCS levels IV and V (19). Scoliosis may progress very rapidly, coincident with the deterioration in gross motor function. Curves progressed by 4.4 degrees per annum in a group of patients with a decline in function, as compared to 3.0 degrees per annum in a group with stable function (219). Larger curves progress more rapidly, including after skeletal maturity. In skeletally mature individuals, with curves <50 degrees, the progression was 0.8 degrees per annum and 1.4 degrees per annum for curves >50 degrees (219).

**Nonoperative Management.** Physical therapy, injections of BoNT-A, and electrical stimulation have been tried and are not effective (216). Customized seating with molded inserts improves sitting balance and comfort but does not slow curve progression. Bracing of scoliosis in CP is poorly

*Text continued on page 548*

## Galveston Pelvic Instrumentation (Figs. 14-63 to 14-73)

**FIGURE 14-63. Galveston Pelvic Instrumentation**. In the Galveston technique, the segment of the rod that is in the pelvis passes between the two tables of cortical bone in the thickest portion of the ilium, the transverse portion just cephalad to the sciatic notch.

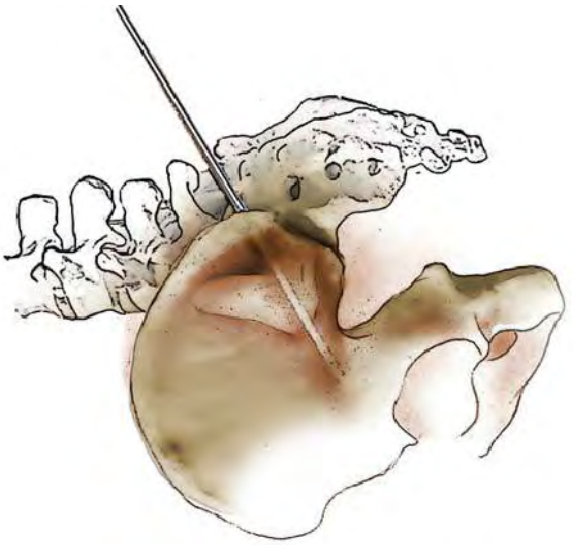
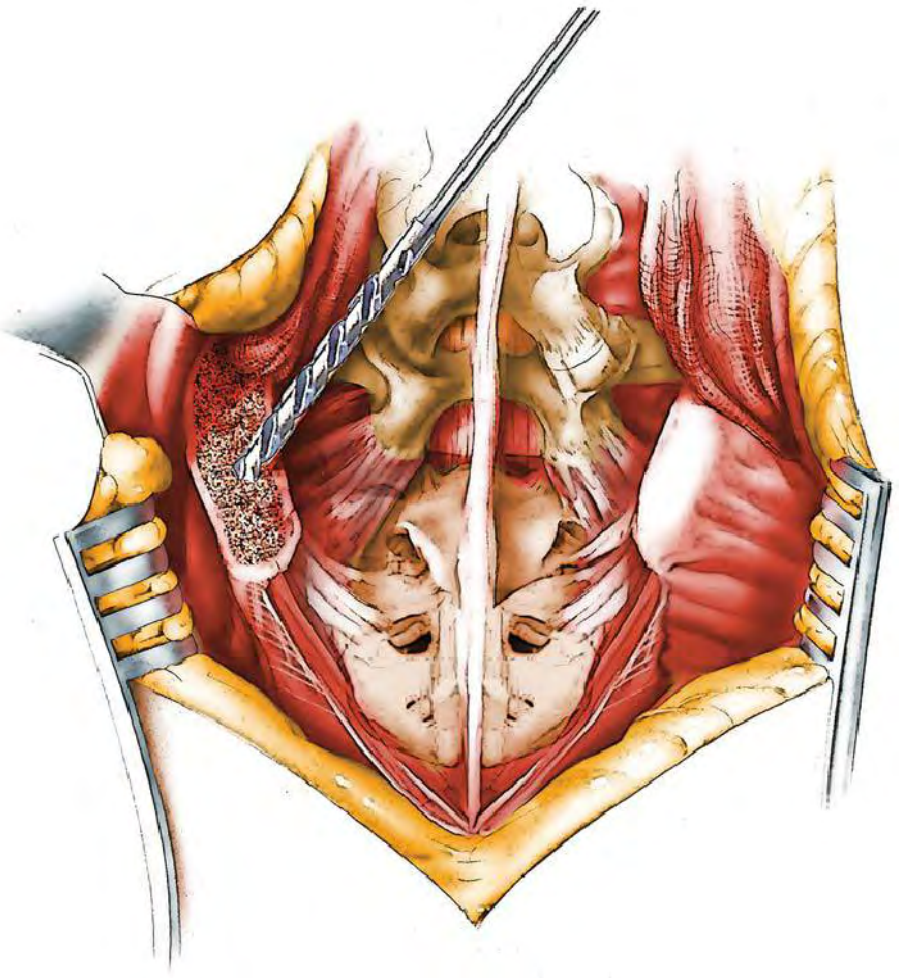


**FIGURE 14-64.** From the midline incision, both iliac crests are exposed. Unlike the exposure for obtaining a bone graft from a midline incision, this entire dissection is best carried out deep to the paravertebral muscles so that the rod can lie in contact with the bone and be covered with the muscle. Some surgeons prefer to split this muscle transversely for ease and speed of execution. Elevation of the muscle is aided by a transverse cut at the caudal extent of the muscle. The periosteum over the posterior crest is incised, and the posterior crest and the outer table of the ilium are exposed. The sciatic notch should be visible because it serves as a guide to the pelvic segment of the rod. The bone graft can be obtained from the more cephalad portion of the ilium, where it will not interfere with the purchase of the rod. In most cases in which this technique is used, however (e.g., paralytic scoliosis), the ilium is very thin, and what little bone is harvested does not make it worthwhile. After the area is exposed, a drill of correct size for the rod is used to drill the path for the rod.



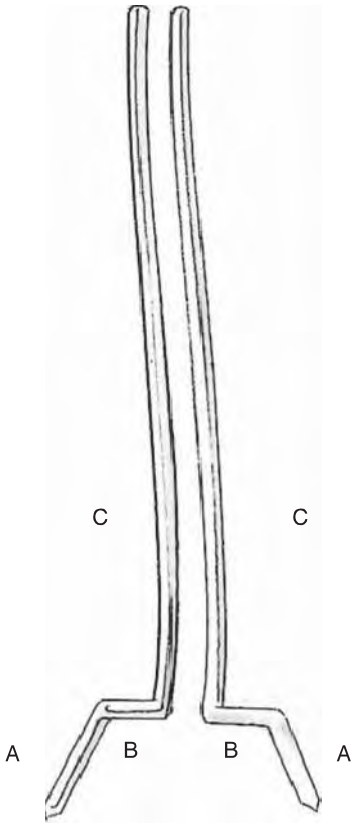



**FIGURE 14-65.** It will take two bends and one twist to produce the finished rod that consists of three segments. The first segment (*A*) is that which lies between the two cortical tables of the ilium and is called the *iliac segment*. The second part of the rod (*B*) runs from the ilium transversely to the area adjacent to the sacral spinous process and is called the *sacral segment*. The last segment (*C*) is that fixed to the spinal vertebrae and is called the *spinal segment*.

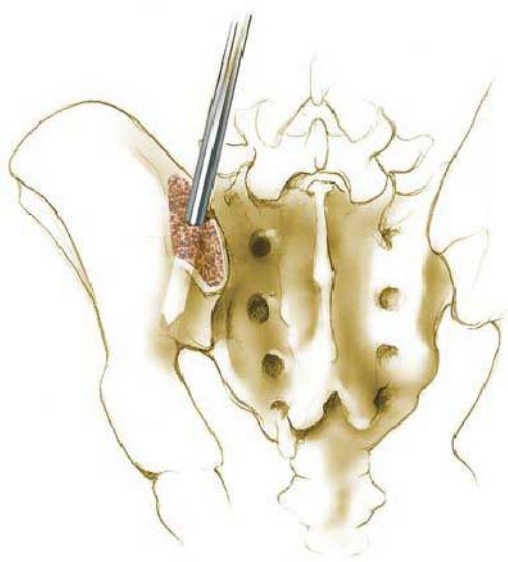


**FIGURE 14-66.** The hole for the iliac segment is made with a drill. The hole is started slightly cephalad to the posteroinferior iliac spine, and the drill is directed between the two tables of the ilium to pass just cephalad to the sciatic notch. The depth of the hole varies between 6 and 9 cm, depending on the size of the child. If desired, a guide pin can be inserted in this hole to be used with a special jig to aid in bending the correct contours into the rod (see Fig. 14-71). After a little experience, however, it is easier simply to bend the rods and make minor adjustments with the rod in place.

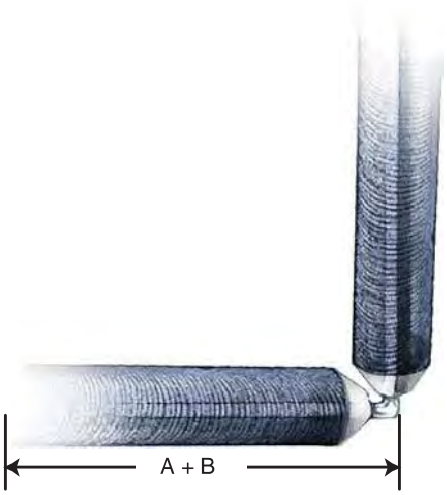
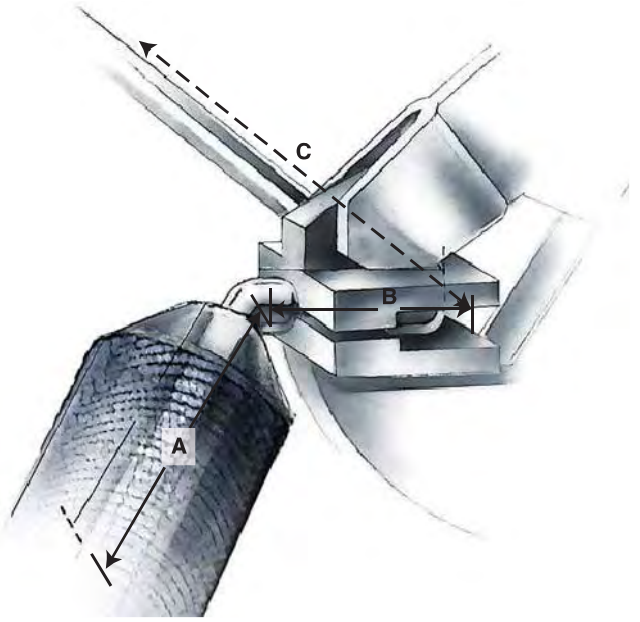



**FIGURE 14-67.** The depth of the hole should be noted; it is usually 7 to 8 cm. This is the length of the iliac segment of the rod (*A*). In addition, the distance from the hole to a point adjacent to the sacral spinous process should be noted. This is usually 2 to 2.5 cm (*B*) and represents the sacral segment of the rod. *C*: The spinal segment of the rod. The rod is now bent with two tube rod benders to place a 60-degree to 80-degree bend in the rod at a distance from the end of the rod that is equal to the length of both the iliac and the sacral segments of the rod. On the concave side of the curve, the rod fits better if the bend is less (i.e., approximately 60 degrees). On the convex side, 80 degrees is usually correct.

**FIGURE 14-68.** The next step is to place the bend that separates the iliac segment from the sacral segment. With a tube bender on the iliac section and a rod clamp on the sacral segment, a bend is placed that allows the rod to reach the sacral lamina when the iliac segment is inserted. In calculating the measurement with the bend, it should be remembered that the bend in the rod itself accounts for at least 0.5 cm. In addition, although the technique for bending the opposite rods is identical, the rods will be mirror images of each other.



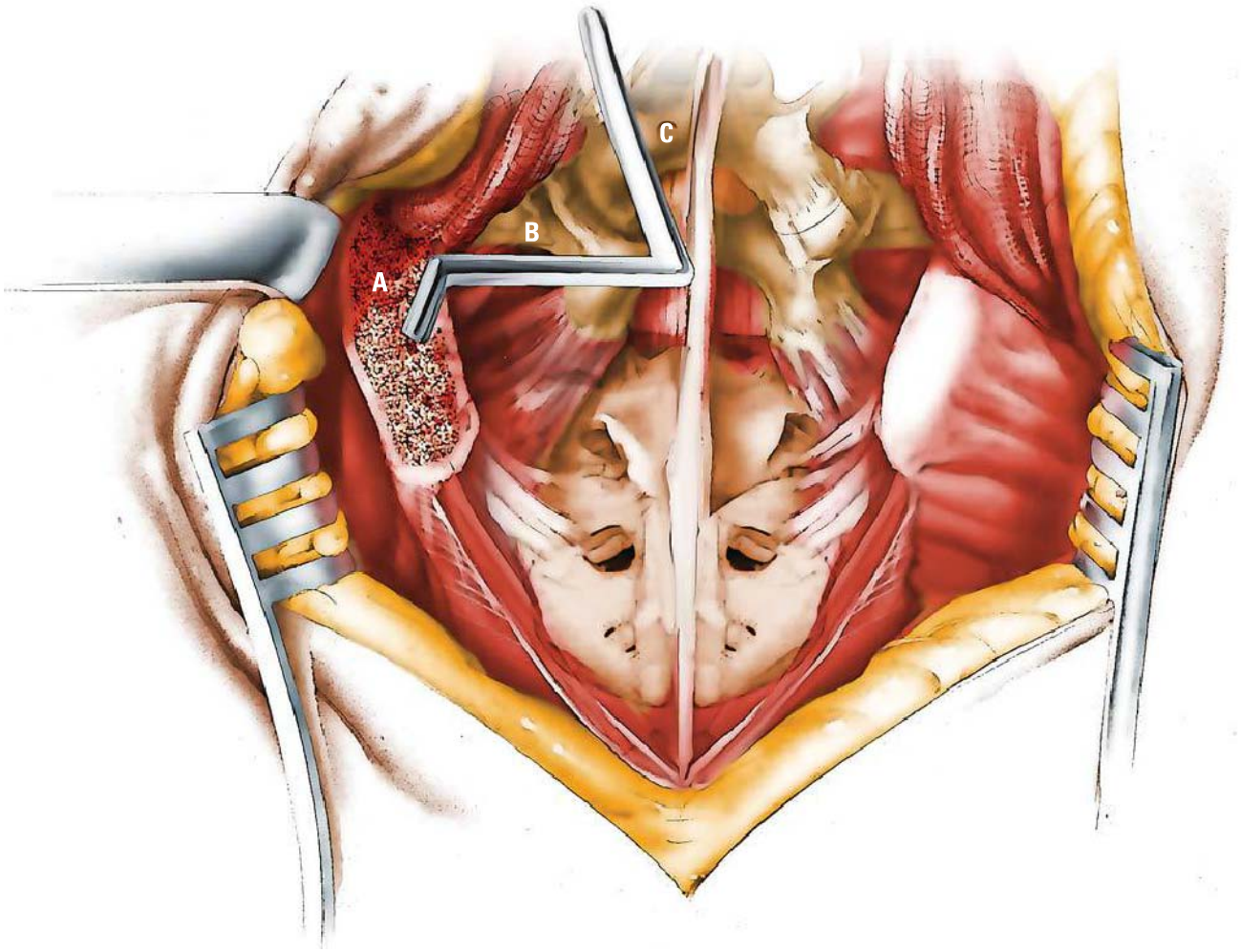




**FIGURE 14-69.** The three sections of the rod are now formed. At this point the rod cannot be placed.

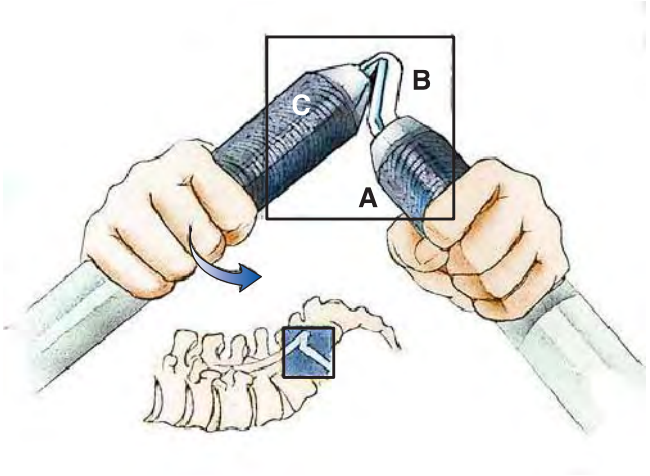



**FIGURE 14-70.** The last step (*B*) is to place a twist in the rod in the sacral segment. This allows the rod to conform to the sacral inclination. Although this can be done to some extent by bending lordosis into the rod, it is usually difficult to incorporate sufficient lordosis close enough to the junction of the sacral and spinal sections to have the rod lie on the sacral lamina. This twist is created by placing a tube rod bender on the spinal (*C*) and iliac (*A*) segments. The benders are brought toward each other. This produces a more ventrally directed spinal section, which conforms better to the sacrum. The amount of twist to be placed must be estimated because the rod cannot be placed at this point.

**FIGURE 14-71.** Finally, the desired spinal contours are bent into the rod. It is best to start with lordosis because it will not be possible to place the rod in the iliac hole and next to the spine until this is done. Although a rod guide can be used with a pelvic guide pin in the iliac hole and the double-rod guide, this technique usually results in a less than perfect fit and, after a short learning curve, is easily omitted.

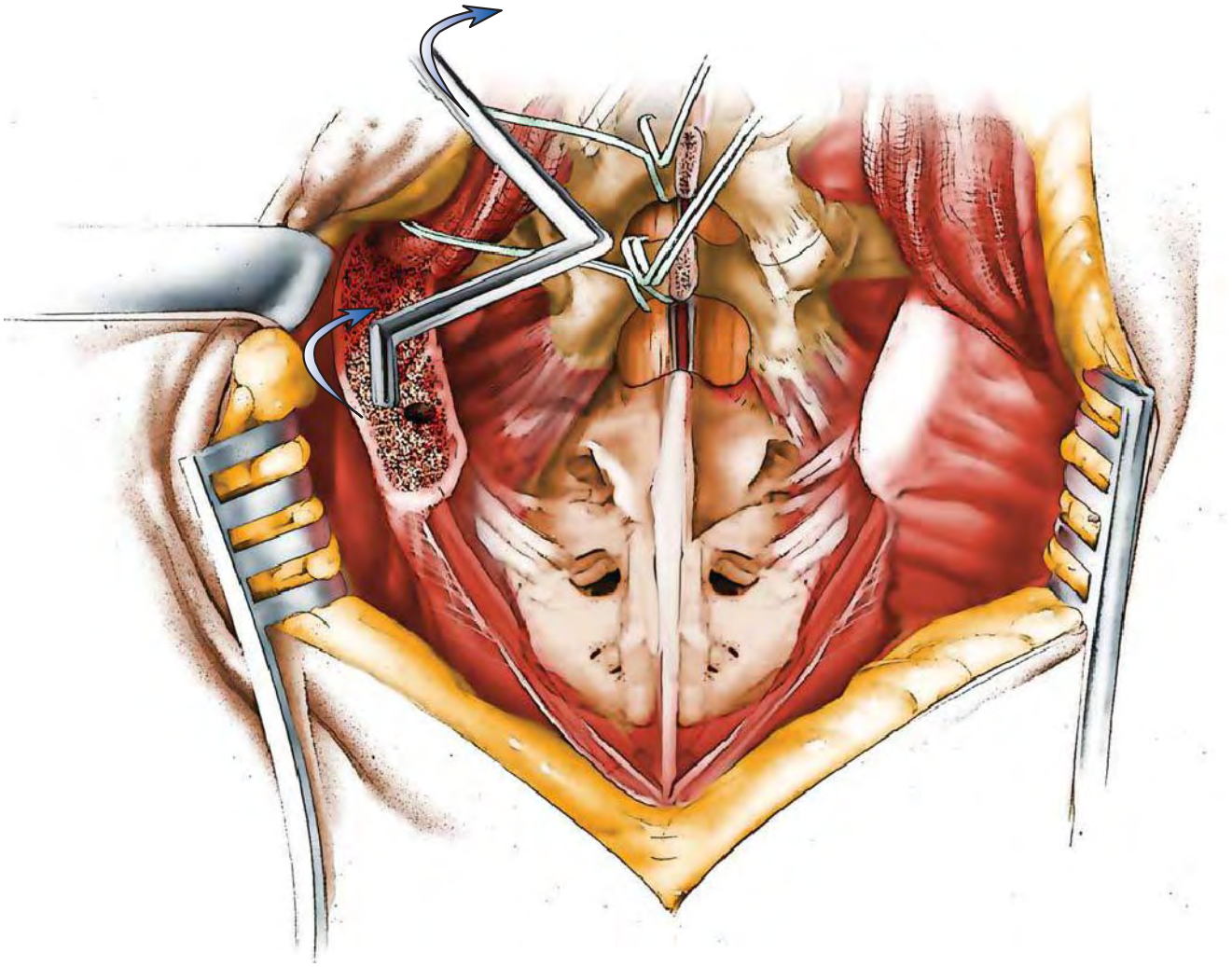


**FIGURE 14-72.** After the rod is contoured and the proper fit of both rods is ensured, the facet excision, any desired decortication, and passage of the sublaminar wires is completed. The rod can be inserted and wired into place.

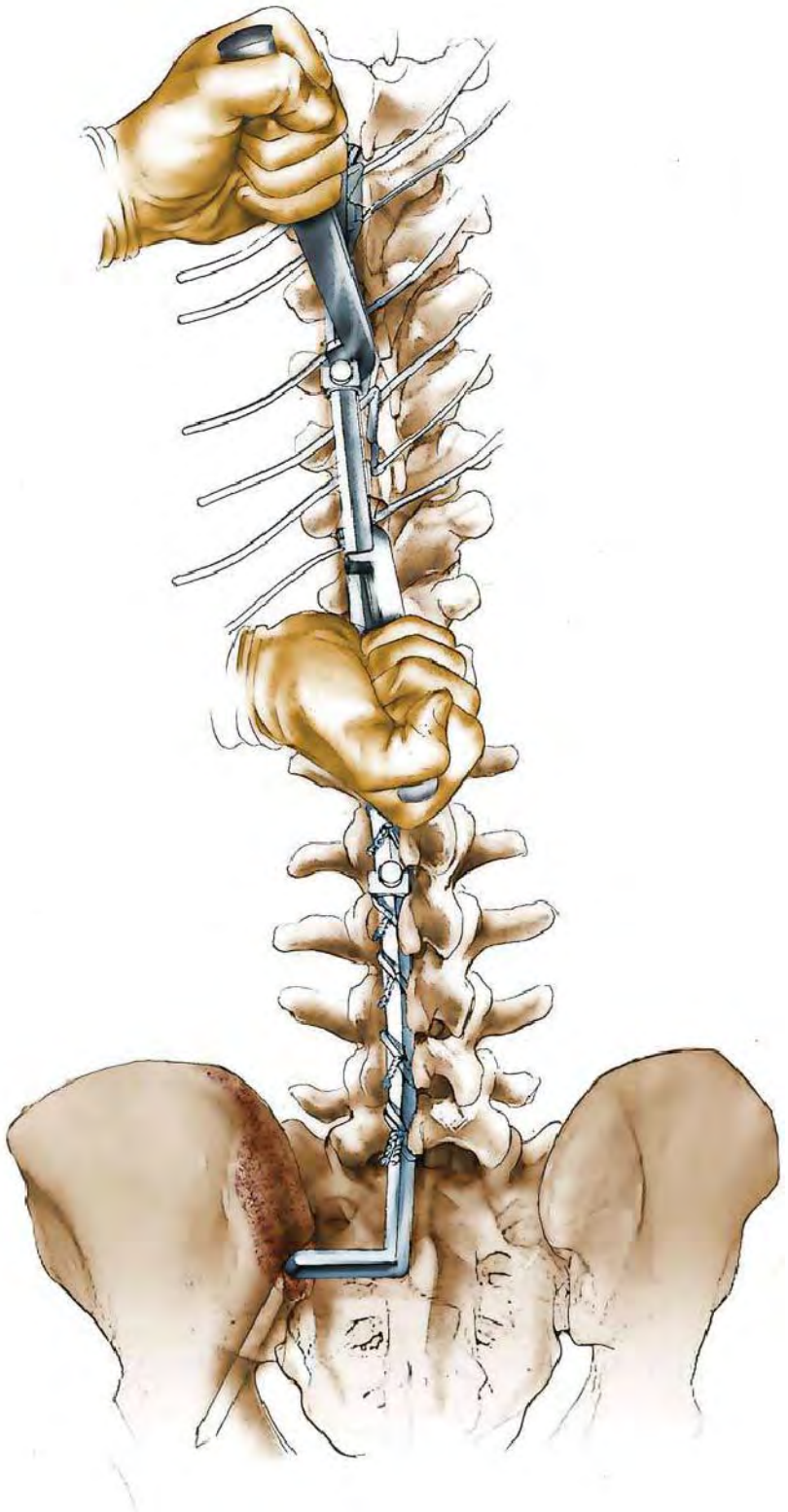


**FIGURE 14-73.** After the rods are in place, and even after some of the wires have been tightened, it is possible to make adjustments in the spinal segment with a pair of in situ rod benders. After the first rod is in place, consideration should be given to placing the second rod. It is likely that after tightening some of the wires on the spinal segment of the first rod, the contour of the spine would have changed. The contour of the spinal segment of the second rod may need to be adjusted. After all the adjustments are made, the cross-links are secured.

tolerated and is ineffective in avoiding progression in the long term (204). Nevertheless, in an effort to maximize spinal growth and to demonstrate that all reasonable steps have been taken prior to surgery, it is sometimes appropriate to offer bracing or seating modifications with close clinical and radiographic monitoring. This gives the parents and caregivers the opportunity to learn about the natural history of the curve in their child and come to terms with the need for major spinal surgery. In one study, bracing was helpful in curves <40 degrees in ambulant patients, but these are not the typical GMFCS V patients (204). Bracing in GMFCS V children may, at best, slow curve progression in immature patients with small curves and good compliance. Surgery may be postponed but will still be required for the vast majority of patients (216).

**Preoperative Assessment.** Poor nutritional status is associated with increased morbidity and mortality. Optimizing nutritional status requires adequate time for assessment and correction. Serum albumin <35 g/L and a lymphocyte count of <1.5 g/L were associated with increased rate of deep infection, prolonged intubation, and delayed discharge from hospital in one study (221). Nutritional supplementation is more effective and safer by the enteral route than by the parenteral route. Supplementation of gastrostomy feeds can be very effective in children with a feeding tube in place. Otherwise, supplementary feeding via a fine nasogastric tube may be offered for 3 to 6 weeks, prior to surgery. The period of preoperative preparation should be closely supervised and monitored by a gastroenterologist and dietician. Low serum iron and transferrin levels are common and may result in iron deficiency anemia. Gastroesophageal reflux is common and must be treated medically or by fundoplication, prior to major surgery. Perioperative conversion of the gastrostomy tube to a gastrojejunal feeding tube allows effective enteral nutrition early in the postoperative period when the risk of gastroparesis and gastroesophageal reflux is highest due to narcotic analgesics.

Good respiratory function is vital. Children with known respiratory disease require evaluation and preoperative chest management including physical therapy. Those with seasonal asthma should be offered surgery at the optimum time of year from the respiratory point of view.

Some seizure medications such as valproate are associated with both an anti–vitamin D effect (resulting in osteopenia and increased fracture risk) as well as increased bleeding (increased transfusion requirements) (216, 222). It may be prudent to wean valproate and convert to other antiepileptics as needed to control seizures if possible. Because the primary effect is on platelet function, routine screening parameters fail to detect the problem. The platelet function assay is the necessary test. Seizure management should be optimized and closely supervised throughout the perioperative period.

Comfortable sitting, for prolonged periods of time, is essential for participation in family life, schooling, and community activities. The functional goal of scoliosis surgery in CP is to improve sitting balance and ease the burden of care. The biomechanical goal is to achieve a stiff, well-balanced spine over a level pelvis with flexible, pain-free hips. The surgical goal is a long posterior spinal fusion with the fusion limits from high in the dorsal spine to the pelvis in the majority of children (223). If the fusion does not extend to high in the thoracic spine, there is a risk of a junctional kyphosis that may interfere with the patient's ability to see above the horizontal, make eye contact, and interact with their family and environment (see Figs. 14-63 to 14-73).

**Technical Issues (Figs. 14-14, 14-15, and 14-62 to 14-73).** Multisegmental fixation using sublaminar wires or combinations of hooks and pedicle screws to strong double rods are necessary to distribute the corrective forces throughout the length of the fusion. The unit rod has excellent results and costs a fraction of other systems (223, 224). Fusion should include the pelvis when pelvic obliquity exceeds 10 to 15 degrees, on an anteroposterior radiograph of the pelvis with the patient in the sitting position (216, 223). Without fusion to the pelvis, pelvic obliquity may continue to progress, resulting in impaired seating, recurrent hip displacement, and failure to reach the goals of the operation (225). Many surgeons recommend fusion to the pelvis in all GMFCS V patients regardless of preoperative pelvic obliquity. Fixation to the pelvis may include hooks, rods, and screws with the best documented and most reliable results reported using the Galveston technique (224, 225) (Fig. 14-62A,B) (see Figs. 14-14 and 14-15). To improve sitting, spinal balance in both the coronal and the sagittal planes is essential. Maintenance of this desired alignment is feasible with a solid fusion throughout the length of the spine. A strong fusion mass will necessitate large quantities of allograft, autograft, bone substitutes, and/or the addition of growth factors (223).

With regular monitoring and surgery at an appropriate age and stage of curve progression, anterior spinal surgery and anterior spinal fusion is not required. However, some children develop rigid curves and an anterior spinal release may be necessary as part of a one- or two-stage correction before instrumented posterior fusion. Specific indications for anterior spinal surgery may include the need to correct severe pelvic obliquity, to achieve balance in large rigid curves that do not correct to <50 degrees during lateral bending or traction radiographs. The release of severe rigid curves may be achieved by resection of the anterior longitudinal ligament, excision of the annulus, and removal of disc material and end-plate cartilage back to the posterior annulus and posterior longitudinal ligament. This may be done via an open or thoracoscopic approach (226). The role of anterior instrumentation is not established and may not be necessary when a rigid posterior instrumentation is performed. Anterior fusion may be indicated in immature children with growth potential to avoid the crankshaft phenomenon.

Spinal surgery in severe CP carries a significant risk of mortality and severe morbidity. Hence prior consultation with the multidisciplinary team, optimization of nutrition, respiratory function, and tone management are essential. If an ITB pump is present, this can be adjusted postoperatively to reduce tone and the need for analgesia. However, the skin over the pump needs to be protected during surgery from injury on the spine frame, hematogenous infection of the pump must be avoided by all means, and the catheter needs to be protected or resited

through the fusion mass. Postoperative fluid management is critical in these compromised patients. Intraoperative blood loss should be minimized by surgical technique and sometimes the use of additional agents such as Aprotinin (227). Both hypovolemia and coagulopathy must be avoided. The use of the cell saver intraoperatively may also be beneficial. The role of intraoperative monitoring of neurologic function remains controversial in nonambulant children with CP. There are problems with both somatosensory spinal evoked potentials and cooperation and communication during a wake-up test (216).

Postoperative bracing is not usually required. A brace may be helpful when osteopenia is identified intraoperatively and fixation is deemed to be tenuous. Parents and caregivers need clear instructions in regard to activity limitations and precautions in the immediate postoperative period.

### Outcomes of Spinal Surgery in Cerebral Palsy.

Spinal surgery in CP has been shown to effectively correct the deformity, with curve correction varying from 45% to 75% accompanied by good correction of pelvic obliquity. Curve progression after successful fusion is usually <10 degrees although this may be greater in immature individuals with growth potential (223–225, 228, 229). Surgical complications are common, often severe, and sometimes life threatening. Complications include fixation failure, respiratory failure, wound infection, decubitus ulceration, neurologic injury, intestinal obstruction, pancreatitis, and death. Major complication rates vary from 40% to 80% and mortality rates from 5% to 7% (230). Pseudarthrosis may be asymptomatic but may result in progression of deformity, implant failure, and the need for revision surgery. Deep wound infections are relatively common, compared to idiopathic scoliois but may be reduced by using allograft impregnated with antibiotics (231). Deep infection requires surgical debridement, antibiotics, nutritional and respiratory support. Vacuum-assisted closure may be very helpful in managing some wound complications.

Spinal fusion in children with CP improves sitting position and appears to improve upper limb function, eating, and respiratory function (216, 232). It may also prolong life (232). What is not yet clearly established is the effect of spinal surgery on the patient's quality of life and in reducing the burden of care (232, 233). It is very likely that quality of life and reduced burden of care can be achieved, but we have until recently lacked appropriate tools to measure these outcomes. Such tools are now being developed and will allow the effects of spinal surgery to be assessed in the domains of most importance to the patient and their families (234).

## ACKNOWLEDGMENTS

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