CHAPTER 24

Disorders of the Brain

Levels of Affection[1]

The neuromuscular system may be affected at various levels, each of which is characterized by changes in motor function peculiar to the site and extent of involvement.

At the *spinomuscular level,* motor activity is simple; the impulses arising in the anterior horn cells of the spinal cord are transmitted through the peripheral nerves to the myoneural junctions and then to the individual muscles. In disorders at the spinomuscular level, the loss of motor power is focal and segmental, with complete paralysis of the muscles or muscle groups that are supplied by a peripheral nerve or by the anterior horn cells in the spinal cord. Muscular paralysis is flaccid or hypotonic, with reaction of degeneration, atrophy, fibrillations, and fasciculations. The deep tendon and superficial reflexes are diminished or absent. Pyramidal tract signs, abnormal involuntary movements, and ataxia are absent. There may be trophic changes in the skin, nails, and bone.

Pathologic processes at the spinomuscular level may be further classified into various sublevels. When the disease originates in the anterior horn cells, as in poliomyelitis, the *spinal level* of the motor system is affected. Other examples of diseases at the spinal level are progressive spinal muscular atrophy of Werdnig-Hoffmann type, progressive bulbar palsy, syringomyelia, and intramedullary neoplasm. The loss of function of the anterior horn cells and the motor nuclei of the brain stem results in clinical findings of flaccid paralysis, atrophy, areflexia, reaction of degeneration, and fasciculations.

At the *neural level* of the motor system, the peripheral nerves and nerve roots are affected, common examples of which are obstetric brachial plexus palsy and progressive neural muscular atrophy (Charcot-Marie-Tooth disease). In affections of nerves, sensory fibers are usually involved, with resultant sensory changes such as anesthesia or hyperesthesia. Otherwise, the clinical findings are similar to those of spinal level affections; that is, there is flaccid paralysis, atrophy, reaction of degeneration, and areflexia as a result of loss of conduction of motor impulses. In the absence of sensory changes, it is difficult to distinguish between diseases of the peripheral nerves, anterior roots, and anterior horn cells.

When the pathologic process arises at the myoneural junction, as in myasthenia gravis and familial periodic paralysis, then it is a disease at the *myoneural level.* In diseases of primarily muscular origin, the motor system is involved at the muscular level. The muscular dystrophies are familiar examples of disturbance of the muscular level in diseases at the spinomuscular level. Paralysis is flaccid, but reflexes persist until the late stages, when marked atrophy has occurred. There is loss of contractibility without loss of excitability; that is, the muscle fibers have degenerated and have been replaced by fibroadipose tissue, but the peripheral nerves and anterior horn cells are normal.

In disorders of the motor system at the *extrapyramidal level,* there is generalized involvement of the muscles of the limbs and trunk. The muscle tone is hypertonic. Atrophy, fasciculations, and reaction of the degeneration are absent. Motion of the limbs is hyperkinetic, with loss of associated or automatic movements. The deep tendon and superficial reflexes are normal. There are no pyramidal tract responses and no sensory deficit. Athetoid cerebral palsy is a common example of a disease at the extrapyramidal level.

At the pyramidal or *corticospinal level* of involvement, motor deficit arises from affection of motor nuclei of the cerebral cortex. Paresis is usually generalized and associated with hypertonicity or spasticity of muscles. Pyramidal tract signs and pathologic reflexes are usually present. There is usually some atrophy that is not focal; it is caused by chronic paralysis and disuse. Fasciculations, trophic disturbances, reaction of degeneration, and abnormal movements are absent. The deep tendon reflexes are hyperactive and the superficial reflexes are diminished or absent. Spastic cerebral palsy illustrates the pyramidal level of motor involvement.

Cerebellar level lesions are characterized by loss of coordination and control, or ataxia. There is no real loss of motor power. Fasciculations, reaction of degeneration, atrophy, and trophic disturbances are absent. The deep tendon reflexes may be diminished, but the superficial reflexes are normal.

Differential feature of various levels of motor function are illustrated in Table 24–1.

REFERENCE

Levels of Affection

1. Tachdjian MO: Pediatric Orthopedics, 2nd ed. Philadelphia, WB Saunders Co, 1990.

TABLE 24–1 **Differentiation of Motor Disorders at Various Levels of Neuromuscular Function**

Type of Disturbance	Spinomuscular			Extrapyramidal	Pyramidal	Cerebellar
	Muscular	*Neural*	*Spinal*			
Loss of motor power	Focal-segmental Usually proximal and axial muscle groups Complete	Focal-segmental Usually distal limb musculature Complete	Focal-segmental Usually distal limb musculature Complete	Generalized Entire limb and movements Incomplete	Generalized Entire limb and movements Incomplete	None Ataxia may simulate loss of power
Tone	Flaccid	Flaccid	Flaccid	Rigid	Spastic	Hypotonic (ataxia)
Atrophy	Present	Present	Present	Absent	Minimal (due to disuse and chronic paresis)	Absent
Fasciculations	May be present	Absent	May be present	Absent	Absent	Absent
Reaction of degeneration	Present	Present	Present	Absent	Absent	Absent
Reflexes						
Deep	Diminished and preserved until late	Absent early	Absent early	Normal or variable	Hyperactive	Diminished or pendular
Superficial	Diminished	Absent	Absent	Normal or increased	Diminished or absent	Normal
Sensory deficit	Absent	Usually present	Absent	Absent	May be present	Absent
Trophic disturbance	Present	Present	Present	Absent	Usually absent	Absent
Ataxia	Absent	Absent	Absent	Absent	Absent	Present
Abnormal movements	Absent	Absent	Absent	Present	None	May be present (intention tremor and ataxia)

Adapted from DeJong RN: The Neurological Examination, 3rd ed, p 382. New York, Harper & Row, 1967; and Farmer TW: Pediatric Neurology, p 612. New York, Harper & Row, 1964.

Cerebral Palsy

Cerebral palsy was first described by William Little in 1862.[259] Little correlated the findings seen in young children with cerebral palsy and associated them with difficult births. Cerebral palsy was then known as Little's disease for decades. The term *cerebral palsy* originated with Freud.[157] *Static encephalopathy* has been used interchangeably with *cerebral palsy.*

DEFINITION

A succinct and accurate definition of cerebral palsy is difficult to construct.[308] There is wide variability in the manifestations of cerebral palsy. In all cases, however, the following must be true:

- Cerebral palsy is the result of a brain lesion. Therefore, the spinal cord and muscles are structurally and biochemically normal.
- The brain lesion must be fixed and nonprogressive. Thus, all of the progressive neurodegenerative disorders are excluded from the definition.
- The abnormality of the brain results in motor impairment.

The insult to the brain occurs either prenatally, perinatally, or during childhood. Although older children with brain damage were traditionally excluded from the definition, from an orthopaedic standpoint, this is not clinically relevant. Certainly, any orthopaedist caring for a child who has sustained an anoxic injury after nearly drowning or who is spastic following infectious meningitis would argue that these slightly older children do in fact have cerebral palsy.

The clinical manifestations of cerebral palsy depend on which part and how much of the brain is involved. The range of manifestations is huge, as both the young child who is intellectually bright but walks on his toe and the noncommunicative wheelchair-bound child with seizures both meet the above definition of cerebral palsy.

The orthopaedic surgeon is consulted by the pediatrician or family for the management of the musculoskeletal problems that follow from the underlying brain lesion. It is of utmost importance for the orthopaedist to evaluate the child thoroughly to ascertain why a child has cerebral palsy. If the child was born full-term, if there were no perinatal medical problems, and especially if the child began to develop normally and then regressed, prompt neurologic consultation should be sought. The neurologist will differentiate cerebral palsy from such dangerous entities as brain and spinal cord tumors and from progressive neurodegenerative diseases, some of which are treatable.

EPIDEMIOLOGY

The incidence of cerebral palsy is increasing slightly.[42] In recent reports the incidence is between 2.4 and 2.7 per 1,000 live births.[59,305,394] The prevalence of cerebral palsy appears to be increasing secondary to an increase in the number of very low birth weight babies being born and the increased survival for these tiny neonates, while the rate of cerebral palsy among infants of a given birth weight has remained stable.* This increase in incidence is of concern, as the

* See references 245, 308, 310, 314, 325, 352.

economic impact of cerebral palsy is significant, with the cost per case estimated at $503,000 in 1992.[146] The risk of cerebral palsy in a child born full-term is approximately 1 in 2,000.[394] The incidence of cerebral palsy has been correlated with both gestational age and birth weight.[197] Cerebral palsy was diagnosed in 12.3 percent of babies born between 24 and 33 weeks of gestational age.[438] Approximately 50 percent of children with cerebral palsy have low birth weights, and 28 percent of children with cerebral palsy weigh less than 1,500 g at birth.[103,354] Birth-weight-specific cerebral palsy prevalence ranges from 1.1 per 1,000 neonatal survivors in infants weighing 2,500 g or more to 78.1 per 1,000 in infants weighing less than 1,000 g.[353]

There is an increased incidence of cerebral palsy in multiple births. The incidence of cerebral palsy in recent studies of multiple births is 9 to 12 per 1,000 in twins, 31 to 45 per 1,000 in triplets, and 111 per 1,000 in quadruplets.[185,351,498] The predisposition toward cerebral palsy in twin pregnancies is present even when birth weight and gestational age are controlled for.[490] The risk of cerebral palsy is high for a surviving twin when the other twin dies in utero.[350,351]

ETIOLOGY

The etiology of cerebral palsy will be divided into three time periods: prenatal, perinatal, and postnatal.

Prenatal. The brain of the fetus is susceptible to damage from maternal infections and toxins. The TORCHES group of infections (toxoplasmosis, rubella, cytomegalovirus, herpes, and syphilis) are known to cause significant damage to the developing brain of the fetus, leading to very neurologically involved babies with mental retardation, microcephaly, and seizures. Orthopaedic deformities are noted in 82 percent of these children.[260]

Fetal exposure to drugs and alcohol through maternal use can also result in injury to the developing brain. Unfortunately, this is being seen more frequently in newborn nurseries. Cocaine, heroin, and marijuana all can cross the placental barrier and cause damage to the central nervous system (CNS) of the fetus.

Congenital malformations of the brain that occur during early pregnancy often result in severe cerebral palsy.

Rhesus blood group incompatibility resulting in kernicterus as a cause of cerebral palsy is decreasing in incidence with improvements in prenatal care. Rhogam treatment of Rh-negative mothers has led to a fall in kernicterus, which often led to the development of such movement disorders as athetoid cerebral palsy.

Maternal health problems, for example renal failure or infections, can lead to problems with brain development in the fetus.[359] Prenatal chorioamnionitis and maternal infection have been associated with an increased risk of premature onset of labor and cerebral palsy in the infant.[17,307,323,324] Placental abnormalities have been linked with a higher frequency of cerebral palsy.[104]

Fetal biophysical profile scores (BPS) are prenatal noninvasive tests used to monitor the health of the developing fetus. BPS scores are often obtained in high-risk pregnancies. Abnormally low BPS scores are thought to result from antenatal hypoxia and have been associated with an increased incidence of cerebral palsy.[271]

Perinatal. Anoxia as a result of perinatal complications may lead to the development of cerebral palsy. A tight nuchal cord[312] or placental abruption[439] can lead to anoxia resulting in cerebral palsy. Fetal hypoxia may be discovered by fetal heart rate monitoring, but changes consistent with hypoxia, such as late deceleration of the heart rate with uterine contractions, are quite common and not specific.[311] The frequency of cerebral palsy associated with birth asphyxia is estimated as 1 in 3,700 full-term live births.[499] Fetal distress during delivery has been documented in some children with cerebral palsy.[159] The mode of delivery—vaginal or cesarean—has not been found to influence the incidence of cerebral palsy.[413]

Premature delivery, either from premature onset of labor or from premature rupture of membranes, is commonly associated with cerebral palsy.[111]

Sepsis in the neonatal period can predispose to the development of cerebral palsy in the low birth weight baby.[491] Bronchopulmonary dysplasia and prolonged ventilation in preterm infants may result in hypoxia, which predisposes the infant to cerebral palsy.[12,180,306] Extracorporeal membrane oxygenation (ECMO) has been used to sustain babies with severe cardiorespiratory failure. Cerebral palsy has been diagnosed in up to 20 percent of surviving children who were treated with ECMO.[179]

Open heart surgery for the treatment of severe congenital heart disease has been linked with an increased incidence of cerebral palsy. Heart surgery before the age of 1 month resulted in cerebral palsy in 25 percent of babies.[292] Clearly, these children are quite ill, with an increased risk for hypoxia, sepsis, and prolonged ventilation.

Postnatal. Infections such as meningitis in early childhood can lead to cerebral palsy. Any episode of hypoxia, such as cardiopulmonary arrest, near drowning, and suffocation, can also produce brain damage leading to cerebral palsy.[6] Trauma, such as motor vehicle accidents producing head injury, severe falls, and child abuse, may result in cerebral palsy as well.

CLASSIFICATION

There are two different ways to classify patients with cerebral palsy. The first classification is physiologic and describes the type of movement disorder present. The most common movement abnormality is spasticity. Spasticity results from damage to the pyramidal system, particularly the motor cortex in the brain. There is disinhibition of pathologic reflex arcs, leading to increased tone in the extremities. The tone is velocity dependent, meaning that if a muscle is stretched rapidly, tone increases more than if the same muscle group was gradually and gently stretched.

Hypotonia is, as its name implies, abnormally decreased tone. Babies with cerebral palsy are often described as floppy or hypotonic. Hypotonia is usually a phase, leading most frequently to spasticity as the baby matures.

Dystonia is present when increased tone is present, but it is not velocity dependent. Whereas the tone in spasticity is described as "clasped knife," the tone in dystonic cerebral palsy is described as "lead pipe," meaning that the tone does not decrease with gentle prolonged stretching.

Athetosis is characterized by abnormal writhing movements which the patient cannot control. The movements become more exaggerated as the patient tries to complete a purposeful motion. Geographically, athetosis results from damage to the basal ganglia. Speech is often garbled and difficult to understand, yet the patients may be intelligent. Athetosis has frequently been the result of neonatal kernicterus.[156]

Cerebellar lesions lead to ataxic cerebral palsy. Balance is disturbed, leading to a wide-based and clumsy gait. Pure ataxic cerebral palsy is rare.

Patients with cerebral palsy frequently have a mixed form of movement disorder. It is important to correctly classify the movement disorder of a patient with cerebral palsy, as the results of surgical treatment are unpredictable for all but the purely spastic patient.

The second classification system is geographic, and describes what part of the body is affected by cerebral palsy. Hemiplegia is present when only one side of the body is involved, with the upper extremity usually more involved than the lower extremity (Figs. 24–1 and 24–2). Diplegia implies involvement of both sides of the body, with both lower extremities involved (although not always symmetrically) and lesser involvement of the upper extremities (Fig. 24–3). Involvement of both lower extremities and one upper extremity is termed triplegia. Quadriplegia, or total body involvement, is present when all four extremities are severely involved, with poor trunk control as well (Fig. 24–4). There is often disagreement among clinicians over the difference between severe diplegia and quadriplegia.[308]

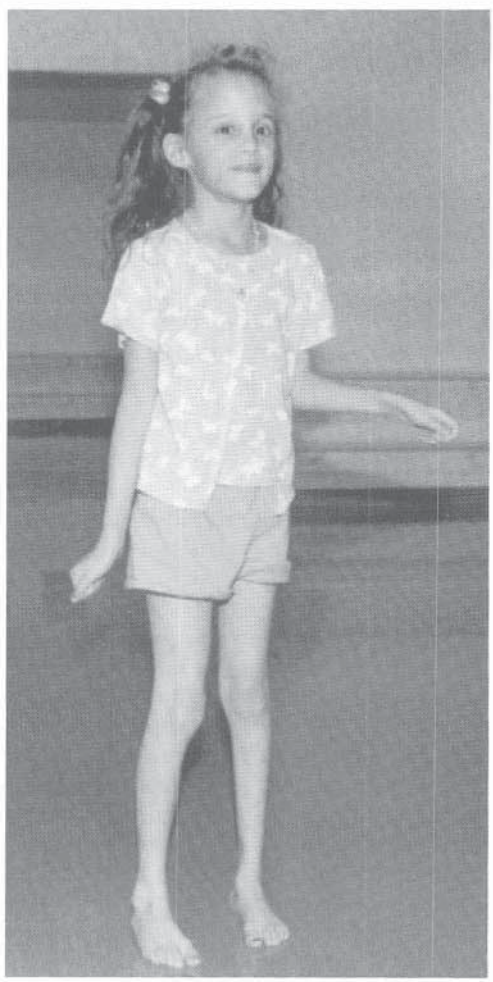


FIGURE 24–1 Girl age 7 years 6 months with left hemiparesis. Note the posturing of the left upper extremity in flexion and the relative atrophy of the calf.

A word of caution is needed. If the patient has abnormal tone in both lower extremities but the upper extremities are completely normal, the examiner should beware. Patients with diplegia and quadriplegia will have some abnormality in the upper extremities, such as decreased fine motor control, spasticity, or increased reflexes. If the upper extremities are normal, it is imperative to evaluate the spinal cord. Spinal cord pathology, including tumor, may masquerade as cerebral palsy.

There is one genetic neurologic disease resembling cerebral palsy in which the lower extremities are both spastic, yet the upper extremities are normal. This disease is known as familial spastic paraparesis. Various forms of the disease exist, and a history of other affected family members is helpful.

HISTOPATHOLOGIC FINDINGS

Two findings frequently described on histopathologic examination or imaging studies of the brain in children with cerebral palsy are periventricular leukomalacia and intra- and periventricular hemorrhage. Periventricular leukomalacia is defined as patchy areas of necrosis in the periventricular white matter adjacent to the lateral ventricles. It results from ischemic insult to the arterial watershed area close to the ventricular walls. Pyramidal tract fibers mapping to the lower extremities pass through this area and are therefore more susceptible to injury than fibers responsible for the upper extremities and face. The bigger the lesion is, the more fibers are injured and the greater the proportion of the body that is affected by the cerebral palsy.[44]

The areas of the brain immediately adjacent to the ventricles are also most susceptible to hemorrhage. Hemorrhage may be seen on ultrasound, magnetic resonance imaging (MRI), or computed tomography (CT). Mild hemorrhages involve the germinal matrix adjacent to the ventricles, while more severe hemorrhages extend into the ventricles themselves and into the parenchyma of the brain. Hypoxia is known to predispose to peri- and intraventricular hemorrhages.[44]

EVALUATION

The first step in the evaluation of a child presenting with cerebral palsy is obtaining a complete history. The birth history is particularly important and should include birth weight, gestational age at birth, and any complications that occurred following birth. Whether or not the child was hospitalized in the neonatal intensive care unit and whether the child required the assistance of a ventilator are important data. If the birth history is entirely normal, neurologic consultation should be considered.

The next step is a careful documentation of the motor milestones. Children with cerebral palsy have delay in reaching motor milestones, so the examining physician must be aware of the normal development of the infant. Head control

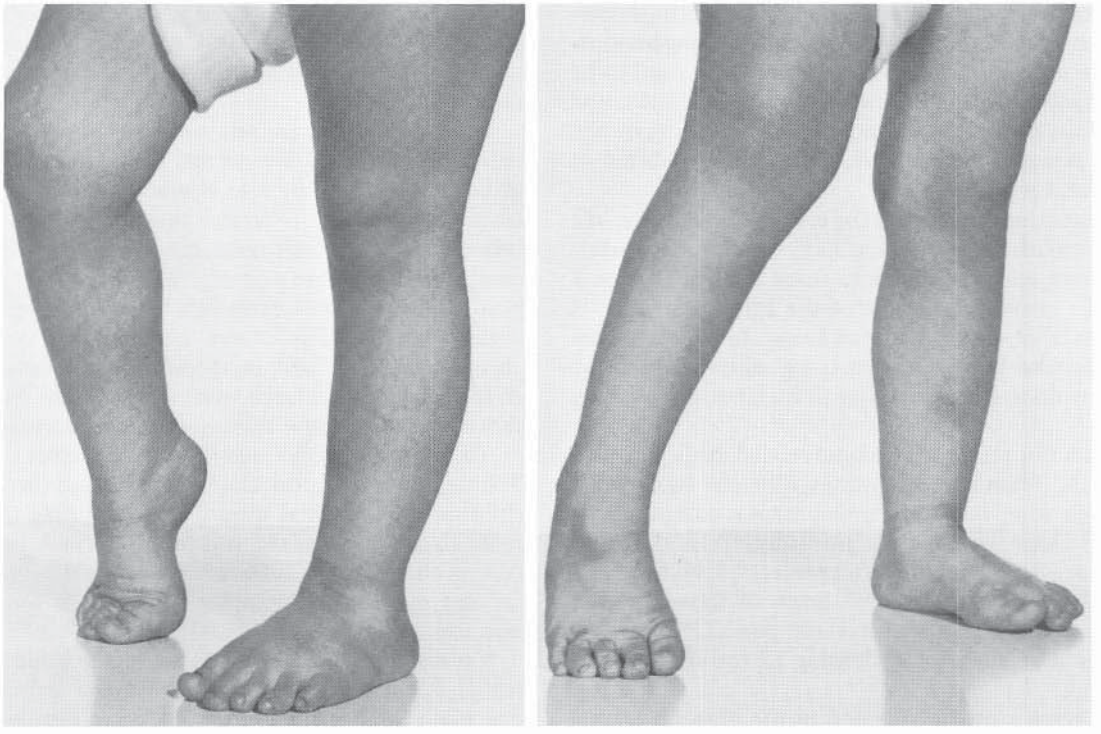


FIGURE 24–2 Right spastic hemiplegia with equinus deformity.

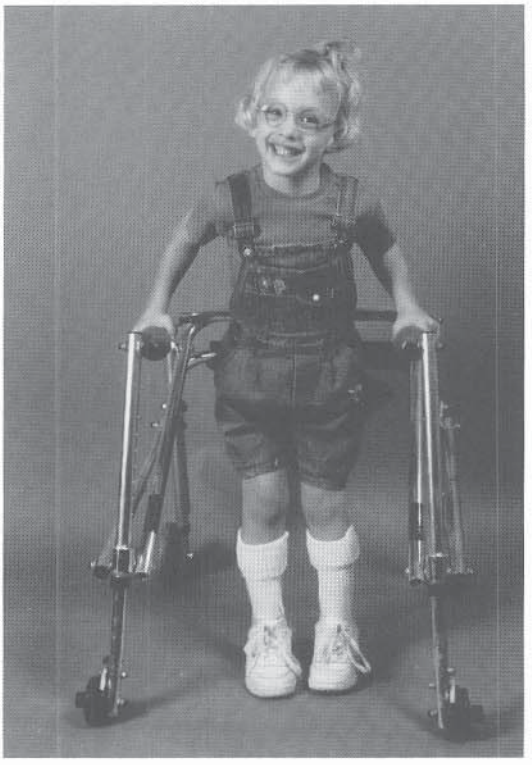


FIGURE 24–3 Girl age 5 years with spastic diplegia. She walks with the aid of a walker and bilateral ankle-foot orthoses.

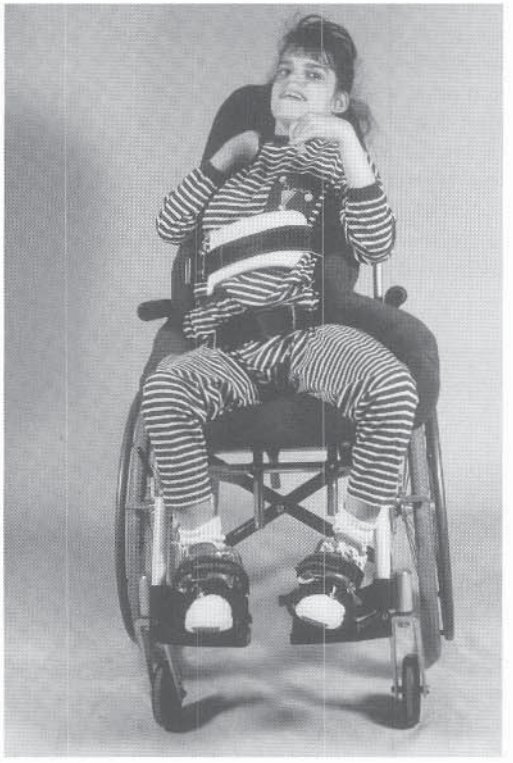


FIGURE 24–4 Girl age 15 with spastic quadriplegia.

should be present at 3 to 4 months (at the latest by 6 months), sitting is generally present by 6 months (at the latest by 9 months), crawling is present by 9 months, standing and cruising by 10 months to a year, and walking between 12 and 18 months. Adjustments for prematurity need to be made when assessing motor development. A baby who was born 3 months prematurely and who is not walking by age 15 months is not abnormal and should not be labeled as having cerebral palsy.

Next, the examiner questions the parents about the child's preferential use of one hand or leg. Early handedness, particularly apparent left-handedness in small infants, is often a clue that the neurologic status of the other extremity is abnormal and that spastic hemiparesis may be present. Likewise, infants who always drag one leg when crawling or scooting may do so because of spastic hemiparesis affecting that limb.

Finally, it is important to ask about related medical conditions. Does the infant have strabismus, difficulty swallowing, or frequent choking? Is speech development at an age-appropriate level? Does the child see normally? Does the child suffer from seizures? Some 15 to 60 percent of children with cerebral palsy have seizures, and many of these children began having seizures in the neonatal period.[8,194,246] These observations may all be clues leading to the diagnosis of cerebral palsy.

Specific items must be included in the physical examination of a child with cerebral palsy. First, the examiner should feel for increased muscle tone in the extremities. With the patient relaxed (even sitting in the lap of the parent), the examiner ranges the extremities. Spasticity feels like tightness in the muscles, which become tighter the quicker the limbs are passively moved. Greater range of motion can be gained by slowly and gently stretching the joints in question.

Deep tendon reflexes are increased in patients with cerebral palsy. Asymmetry may be present if the neurologic lesion is asymmetric (i.e., in hemiparesis versus diplegia). Repetitive tapping of the deep tendon reflexes may elicit clonus. Quick passive dorsiflexion of the ankle likewise may produce clonus, which establishes the presence of a neurologic abnormality.

Fine motor activities should be assessed. The examiner should pass the child a toy or a pen and watch as the child manipulates the item. A child with spastic hemiplegia will play with only one hand. Having the patient clap the hands or wiggle the fingers may bring to light difficulties with fine motor control.

Infantile reflexes are retained in children with cerebral palsy. These reflexes are normal in neonates but disappear in normal children by 3 to 6 months of age as the motor cortex matures and overrides them.[45] Bleck's textbook on cerebral palsy outlines these reflexes in clear detail.[51] The startle reflex, or Moro's reflex, is elicited by letting the baby's head drop back into extension with the child supine but slightly elevated. This causes the legs and arms to extend abruptly. Sudden loud noise can likewise cause the older child to extend and lurch from a wheelchair. Moro's reflex should disappear by 4 months of age in normal children.

The parachute reflex is easily tested in babies and toddlers. The examiner holds the child in the air and then lowers the child quickly headfirst toward the examining table. Babies more than 5 months old will reach out with both arms to protect themselves. Children with cerebral palsy cannot do so, and those with hemiplegia will reach out with only one arm.

Yet another simple reflex to test is the tonic neck reflex, which is elicited by turning the supine infant's head to one side. The ipsilateral arm and leg will extend while the contralateral arm and leg flex. This reflex should disappear in infancy. Its persistence should raise a suspicion about cerebral palsy.

Finally, the balance, sitting, and gait of the child are assessed. Can the child sit unsupported without using his or her hands? Can the child get into a sitting position without assistance? Is the child's balance easily disturbed in the sitting position or as the child walks?

The clinical assessment of gait requires that the child's joints be readily seen, so the child should be barefoot and in shorts or a short gown. The evaluation should be conducted with the examiner at the level of the child. Much more can be seen by an examiner sitting on a stool rather than standing and looking down on a small child. There should be enough room for the child to walk naturally. Can

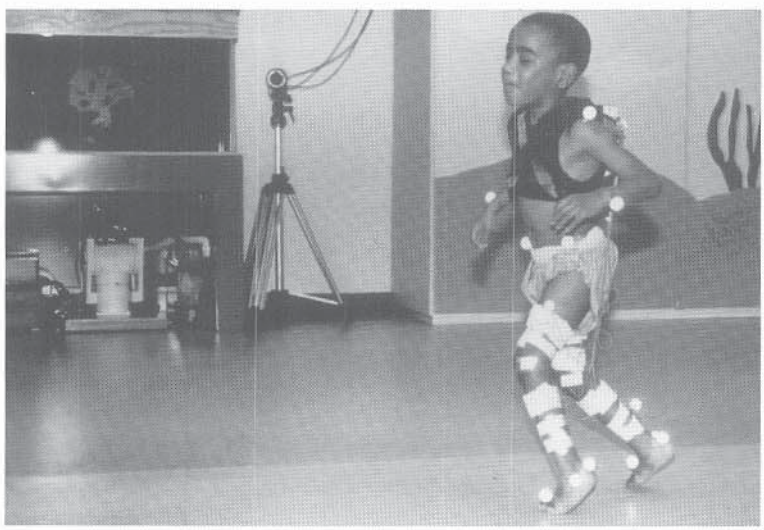


FIGURE 24–5 Young boy with spastic diplegia undergoing preoperative gait analysis.

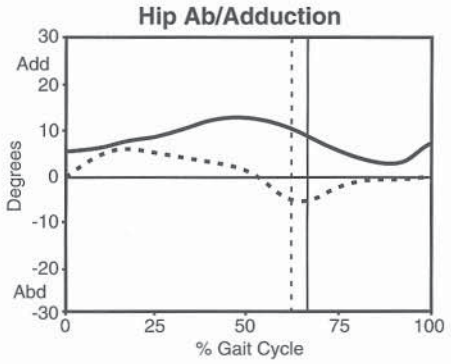


FIGURE 24–6 In normal gait, the hip adducts slightly in stance phase as the contralateral hemipelvis drops, and abducts slightly in swing phase, as represented by the *dotted line.* In patients with scissoring as a result of cerebral palsy, increased adduction of the hip is seen.

the child walk on the toes and heels, and hop on either foot? Finally, the examiner should have the child run if able. A patient with mild hemiplegia may walk nearly normally but exhibit abnormal movement patterns while running; the affected upper extremity will draw upward and not have a normal arm swing.

Gait should be observed from the front of the child and then from the side. A systematic routine should be developed to assess gait by looking at the hip, knee, and ankle from each perspective. In the child with cerebral palsy, crouch gait, with increased flexion at the hip and knee, should be specifically sought. The examiner observes for toe-walking and genu recurvatum. Is there a foot drop in swing phase? A disturbance in clearance of the swing phase limb may be due either to a foot drop or to inability to flex the knee.

OTHER ASSESSMENTS

Rarely are imaging studies ordered by orthopaedic surgeons when establishing the diagnosis of cerebral palsy. When questions persist regarding a correct diagnosis, referral to a pediatric neurologist is indicated. At the neurologist's discretion, such imaging studies as cranial ultrasonography, brain MRI, and CT may be pursued.[239] Similarly, laboratory studies may be necessary to look for evidence of metabolic diseases associated with delays in development and cerebral palsy-like symptoms, such as congenital hypothyroidism or dopa-responsive dystonia. A detailed discussion of these metabolic conditions is beyond the scope of this chapter.

Gait Analysis. Gait analysis has become very popular in the assessment of movement disorders in children with cerebral palsy (Fig. 24–5). The usefulness of such studies is controversial. Through the collection of three-dimensional data on joint movement (kinematics) and forces (kinetics), knowledge can be gained about each joint's movement and position dynamically during gait. Accurate documentation of dynamic range of motion may help in planning surgical treatment and is useful in assessing the results of orthopaedic operations, but does not replace a good clinical examination. When gait analysis graphs are scrutinized together with clinical examination information and slow-motion videotape, better understanding of a patient's gait can be gained. A detailed discussion of gait analysis can be found in Chapter 5, Gait Analysis.

Cadence parameters collected during routine gait analysis include walking speed, step length, cadence (number of steps per minute), and the proportion of time spent in stance and swing phase. There are usually disturbances in cadence parameters in patients with cerebral palsy. In good walkers with spastic diplegia, walking velocity is maintained despite decreased step lengths by increasing cadence—that is by taking many quick, short steps. Good walkers with cerebral palsy can increase their speed by increasing their cadence, but their spasticity interferes with increasing their step length.[4] In more severe diplegia and quadriplegia, walking speed becomes diminished, with decreasing cadence and step length. The proportion of time spent in stance phase, particularly double-limb stance, increases, as the child has greater difficulty with balance and advancing the limb. Children with hemiplegia show asymmetry in step lengths and in single- and double-support times.[316]

Certain kinematic patterns are commonly seen in patients with cerebral palsy.[118,163,217,457,494] Starting with the hip, scissoring is present when increased adduction leads to a narrow base of gait and difficulty advancing the swing phase limb past the stance phase one (Fig. 24–6). Scissoring is due to tightness in the adductor musculature, and probably in part to weakness of the hip abductors. In the sagittal plane, increased hip flexion and anterior pelvic tilt may be part of crouch gait, due to increased tone in the iliopsoas (Fig. 24–7). Increased femoral anteversion may be documented by gait analysis as increased internal rotation of the hips. Asymmetric pelvic rotation may be present, and gait analysis is particularly helpful when the examiner is trying to ascertain whether the abnormal rotation is coming from the pelvis, hips, or tibias (Fig. 24–8).[118]

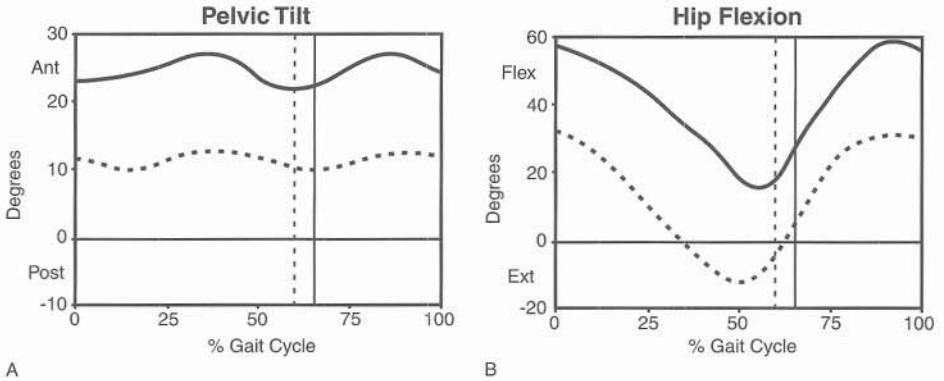


FIGURE 24–7 **A** and **B,** Patients with cerebral palsy may crouch at the hip joint, which is represented on gait analysis as increased anterior pelvic tilt and lack of hip extension at terminal stance phase. Normal kinematics for pelvic tilt and hip sagittal plane motion are represented by *dotted lines.* The area to the left of the vertical lines depicts stance phase, and to the right, swing phase.

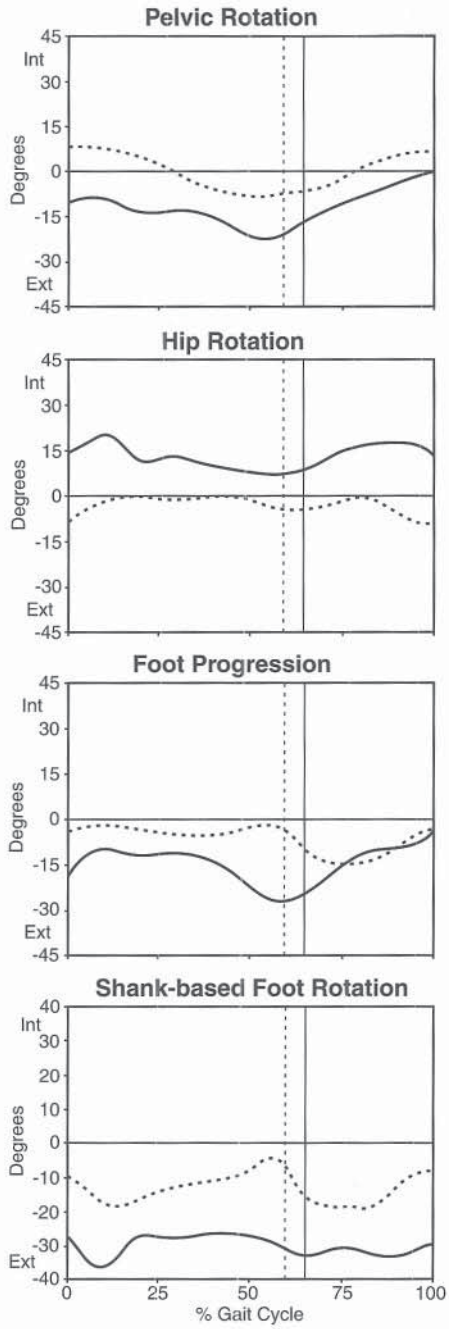


FIGURE 24–8 Transverse plane kinematics are useful in determining the etiology of intoeing or outtoeing in cerebral palsy. In this hemiplegic patient, the involved hemipelvis is characteristically externally rotated, there is excessive femoral anteversion, and the foot is externally rotated relative to the knee, due to midfoot break from equinus. This results in a net external foot progression angle of 14 degrees. Normal transverse plane kinematics are represented by the *dotted lines.*

At the knee, sagittal plane motion is usually abnormal. In patients with tight hamstrings, the knee remains flexed at initial contact and is unable to extend normally during stance phase. In swing phase, spasticity of the rectus femoris may inhibit the patient's ability to flex the knee and clear the floor (Fig. 24–9). Genu recurvatum during stance phase may be present in response to a tight tendo Achillis, with difficulty advancing the tibia forward over the foot.

Ankle kinematics often show disturbances in the three

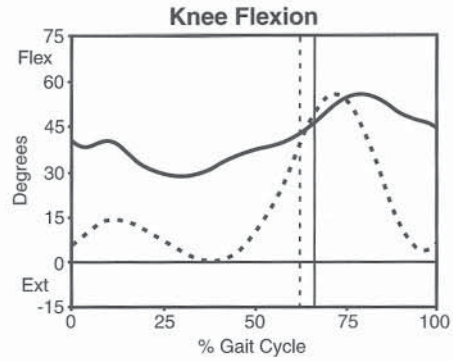


FIGURE 24–9 Crouch at the knee due to hamstring spasticity is documented as increased knee flexion throughout stance phase. As the leg enters swing phase (i.e., to the right of the vertical lines), the knee slowly flexes, reaching maximal flexion later than normal due to rectus femoris spasticity, which interferes with clearing the foot as it swings forward. The *dotted line* indicates normal knee kinematics.

rockers. First rocker, or plantar flexion on weight acceptance, is usually abnormal in patients with equinus contractures. Second rocker, which consists of progressive dorsiflexion during midstance, is diminished in the presence of a tight Achilles tendon. Third rocker, or push-off, is reduced if the ankle is already plantar flexed from the equinus. Finally, swing phase dorsiflexion may be absent owing to weakness or inactivity of the tibialis anterior, leading to foot drop (Fig. 24–10).

Gait analysis is particularly useful in assessing the cause of toe-walking in patients with cerebral palsy. It is tempting to attribute all toe-walking to tight Achilles tendons and to proceed with tendon lengthening. Some children, however, walk on their toes in response to crouch above the ankle and have neutral ankle dorsiflexion but increased flexion at the knee and hip. Lengthening of the Achilles tendon in these children results in calcaneus gait, with persistent crouch at the hip and knee but excessive dorsiflexion at the ankle, leading to inefficient push-off.

Electromyography (EMG) data is also collected in the gait analysis of the child with cerebral palsy. Spasticity leads to electrical overactivity on EMG during gait, and it seems

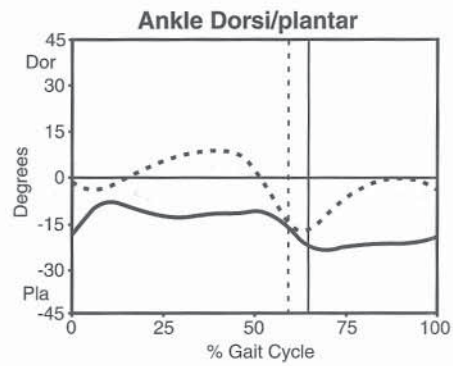


FIGURE 24–10 Sagittal plane kinematics of a patient with cerebral palsy who has equinus. The ankle remains in plantar flexion during stance phase, rather than progressively dorsiflexing in normal second rocker, as represented by the *dotted line.* This patient also has a foot drop, as the ankle does not dorsiflex in swing phase (represented to the right of the vertical lines).

that the more spastic the child, the greater the EMG signal seen and the less phasic the muscles are in their contraction (Fig. 24–11). Dynamic EMG data collected during gait analysis can be correlated with the kinematic and kinetic graphs to gain a fuller understanding of the biomechanics of gait.[342] For example, normally at initial contact, the ankle plantar flexes while the anterior tibialis fires on EMG. Kinetic plots show absorption of power as weight acceptance occurs. In stance phase in a child with cerebral palsy, there may be early heel rise, seen as plantar flexion on kinematic plots, which correlates with gastrocsoleus overactivity on EMG. When is EMG most useful? We find EMG data most useful when evaluating the child with a stiff knee in swing phase for rectus femoris transfer, and when assessing a child with an equinovarus foot for tendon transfer, either anterior tibialis or posterior tibialis split transfers.[344]

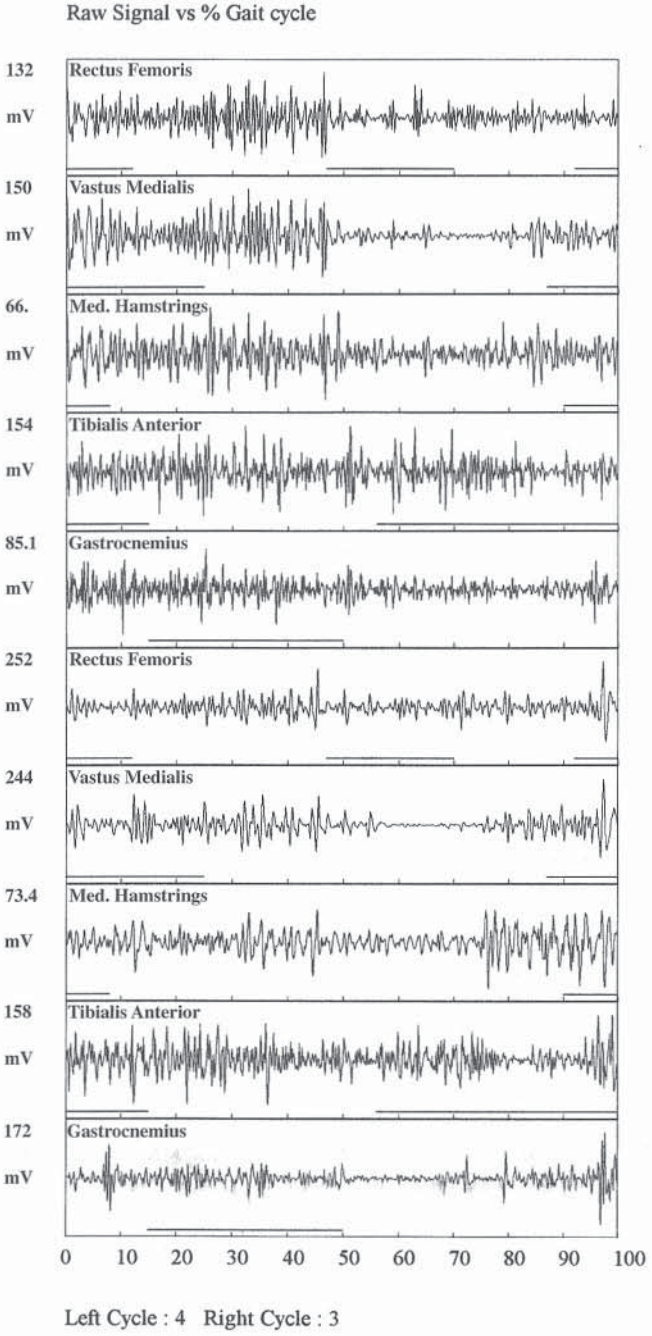


FIGURE 24–11 EMG graph. Typical EMG pattern during gait in a child with cerebral palsy. The horizontal bars represent when a muscle is normally "on." Stance phase is represented from 0 to 60 and swing phase from 60 to 100. There is inappropriate contraction of all muscles during gait.

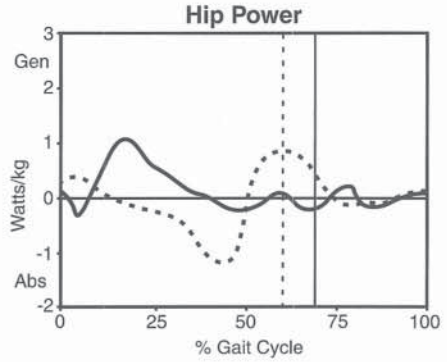


FIGURE 24–12 The normal hip generates power at terminal stance phase as the iliopsoas pulls the leg off the ground (*dotted line*). The vertical lines represent the division between stance and swing phase. In patients with cerebral palsy, hip kinetics can be disturbed. This patient generates no power at terminal stance phase and is therefore less efficient.

Kinetics are the forces exerted across joints during gait. Each joint has well-described kinetic patterns, and the reader is referred to work by Gage for descriptions of these.[162] Two particular forces are clinically interesting. Hip pull-off power, or the force exerted by the iliopsoas and other hip flexors to lift the stance phase limb off the ground and into swing phase, is often diminished in patients with cerebral palsy, leading to decreased energy efficiency (Fig. 24–12). Ankle push-off power, the force exerted by the gastrocsoleus at terminal stance to push the stance phase limb off the ground, is diminished in patients with equinus or calcaneus gait (Fig. 24–13). These powers have been used in research settings to assess the results of tendon lengthening operations.

Yet another means of collecting information in the gait analysis laboratory is the measurement of oxygen consumption. The goal of orthopaedic intervention is to improve the quality and efficiency of walking. By comparing oxygen cost and consumption to normal values through preoperative

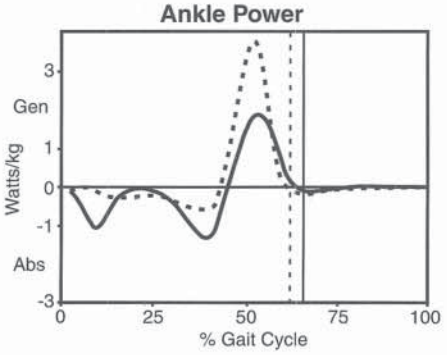


FIGURE 24–13 As the gastrocsoleus contracts at heel rise, the ankle generates a burst of power (*dotted line*). In patients with either equinus or calcaneus gait due to cerebral palsy, power generation is decreased (*solid line*).

and postoperative data collection, the examiner can objectively measure the efficiency of the child's gait.[57,140,482] Heart rate is an indirect measure of oxygen consumption and can be easily measured in the clinic.[313,391,392]

Critics of gait analysis in patients with cerebral palsy have pointed out several flaws with gait studies. Patients fitted with markers and electrodes and placed in front of video cameras do not walk as they do at home or school.[487] This cannot be refuted. Moreover, there is considerable variability in how patients walk from clinic visit to clinic visit. Basing treatment plans on the kinematic and kinetic data from a few strides across a gait lab may not always be in the patient's best interest. Gait analysis can be an adjunct to clinical examinations, but data should be scrutinized and the videotape reviewed to judge whether or not the data are representative of the gait seen in the clinic or described by the parents in the home. Very active gait analysis laboratories claim that their studies can change the surgical plan in patients with cerebral palsy in up to 52 percent of patients.[120,161] The decision-making process using gait analysis data is only as good as the astuteness and familiarity of the orthopaedic surgeon reading the graphs.

In summary, gait analysis in our center has the following purposes:

1. To clearly document the three-dimensional movement of the lower extremity during gait preoperatively.
2. To document changes in gait over time as the patient grows. The gait of many children with cerebral palsy deteriorates as the child grows into adolescence.[225]
3. To allow preoperative and postoperative comparisons following tendon or bony surgery to objectively assess results and to gather research data.
4. To analyze the rotational profile of the patient prior to surgery to help the surgeon select the correct site and amount of rotational change.
5. To confirm, when needed, a surgical plan.

Gait analysis does not tell the surgeon if he or she should operate, but it may help the surgeon fine-tune the operative plan when questions exist.

PROGNOSIS FOR AMBULATION

Many authors have proposed criteria for predicting the ultimate ability of the child with cerebral palsy to walk. Molnar and Gordon's primary criterion for walking is the ability to sit by age 2 years.[294] Bleck proposed that persistence of two or more infantile reflexes beyond 12 to 15 months implies a poor prognosis for ultimate ambulation.[47] Others find that the ability to control the head by 9 months and to sit by 24 months predicts the eventual ability to walk, while the lack of head control by 20 months is ominous.[105] Beals states that the severity of involvement of the lower extremities is the most important factor influencing the child's eventual ability to walk.[35] The presence or absence of mental retardation does not influence the ability to walk; all pediatric orthopaedic surgeons have observed in their own practices intelligent children who are wheelchair users and mentally retarded patients with cerebral palsy who walk without assistance.

The geographic type of cerebral palsy that a child has influences the probability that the child will walk. All children with spastic hemiplegia develop the ability to ambulate. Children with spastic diplegia become ambulatory in 86 to 91 percent of cases. Patients with spastic quadriplegia, however, learn to walk anywhere from 0 to 72 percent of the time. The differentiation between spastic diplegia and quadriplegia varies between studies, leading to the discrepant figures.[403]

The age at which children with cerebral palsy begin to walk varies with the severity of their neurologic disease. Patients with spastic hemiplegia generally walk between the ages of 18 and 21 months. Children with spastic diplegia who walk usually do so by age 4 years. Many agree that the ability to walk plateaus by age 7 years, implying that if a child is nonambulatory at age 7, the child will probably never become ambulatory.[35,47,102]

TREATMENT

Once the diagnosis of cerebral palsy has been made, the physician must choose a course of treatment. Treatment choices are now more numerous than ever, ranging from observation, physical therapy, botulinum toxin, and intrathecal baclofen administration to neurosurgery and orthopaedic surgery. Treatment, whether surgical or nonsurgical, must be goal-oriented. The goals of treatment of children with cerebral palsy that have been linked to productive lives as adults are communication, education, mobility, and ambulation.

Note that walking ranks below mobility. Although ambulation remains a desirable goal, it should not be pursued so fervently that attention to the overall development of the child is ignored.

Bleck quotes Rang as advising orthopaedists to remember that "the child with cerebral palsy becomes the adult with cerebral palsy."[47] Childhood is the optimal time to intervene to maximize the function of the patient with cerebral palsy. Bleck followed many patients with cerebral palsy into adulthood and found little scientific information on the problems faced by adults with cerebral palsy. Although social services are needed to provide medical equipment, transportation, and job training for adults with cerebral palsy, Bleck feels it is the orthopaedist's duty to ensure that the musculoskeletal treatment of the child prevents future problems with pain and deformity as an adult.[50] Patients with cerebral palsy usually do not have severely shortened life spans. In a study of all children with cerebral palsy born between 1966 and 1984, the 20-year survival rate was 89 percent. If the patients were ambulatory, had manual dexterity, and were not mentally retarded, the 20-year survival rate was greater than 99 percent.[218] The overall 30-year survival rate is estimated at 87 percent; it is lower in the presence of spastic quadriplegia, seizure disorders, and profound mental retardation.[101]

Nonsurgical Treatment

PHYSICAL THERAPY. Often the first treatment rendered to the child with cerebral palsy is physical therapy. Yet no controlled studies have confirmed that regular physical therapy improves the outcome of the child with cerebral palsy.[478] One of the first well-designed studies investigating the effect of physical therapy was by Wright and Nicholson in 1973. They found no difference in motor skills or reflexes between those children who had neurodevelopmental training physical therapy and those who had not, after 12 months.[496] Other studies followed and again showed no discernible im-

provement following different forms of therapy.[205,275,330,357] In defense of physical therapy, these studies are difficult to carry out, as they are done in different age groups, in children with varying severity of neurologic impairment, and usually over a brief period of therapy. Yet, as Bleck points out, "The burden of proof is on the proponents of the treatment. Critics need not prove ineffectiveness but can insist on positive data."[51] The efficacy of physical therapy can be proved or disproved only with a properly designed, collaborative, multicenter, randomized, controlled trial. Such a trial has yet to be undertaken.[477]

There are various schools of physical therapy, ranging from the medical model, which targets attaining specific goals such as ambulation, range of motion, or transfers; to neurodevelopmental training as promoted by Bobath; to sensory integration; and even to craniosacral schools. Electrical stimulation of the muscles has also been used in these patients.[76,200] Families like physical therapy, attributing the gains in the young child's abilities to the interaction with the therapist. However, some of these gains are due simply to the neurologic maturation of the child.

Our approach to physical therapy is to establish therapy to monitor the developmental milestones of the very young child, around the age of 2 or 3 years. Therapy is continued if gains are being made in attaining ambulation. School-based programs are used in elementary school, often including adaptive physical education. Postoperative intensive physical therapy is essential to reestablish range of motion and strength following surgical intervention. Strength training of weak muscles has been successful in improving the motor functions of these patients.[108,110] We also draw on the expertise of physical therapists in assessing orthotic needs and wheelchair seating when appropriate. There is no evidence to support the continued use of physical therapy for range of motion, particularly in the nonambulatory child. Physical therapists play an important consultative role in making treatment decisions for the patients they treat on a regular basis, and whom we examine on a relatively infrequent basis.

It is quite common for parents of children with cerebral palsy to resist discontinuing physical therapy. We feel that goals must be set for therapy, and if progress toward those goals is not being made, then either the goals need to be reassessed or therapy should be stopped, as it is not useful. Setting measurable functional goals has led to greater success following physical therapy.[58]

In the older child, transitioning from physical therapy to therapeutic recreation is desirable and is generally met with enthusiasm from the patient.[368] Adaptive sports or swimming allows the child to participate with peers and affords greater enjoyment than exercises in the therapy gym. Time in school should be spent on education at this age, and not on physical therapy.

CASTING. Inhibitive casting has waxed and waned in popularity as a mode of treatment for spasticity in cerebral palsy. It is based on the presence of areas on the feet that are target centers for increased tone at the ankle and, some feel, throughout the lower extremities in some patients. Usually short-leg casts are applied with extended toe plates, careful molding of the heel, and metatarsal head control. This has been performed by physical therapists, and by some physicians. The period of time spent in casts varies but is usually a minimum of 6 weeks, and is followed by the use of orthoses.

In our experience, there is a limited role for casting in patients with cerebral palsy. We have used casting in rare cases of very young children (less than 4 years old) with equinus contractures. Casting is done in an effort to buy time before surgical heel cord lengthening. Because equinus recurs following casting, permanent resolution of the equinus is not a goal.

ORTHOSES. Orthoses can be helpful in improving gait in ambulatory patients with cerebral palsy. Ankle-foot orthoses (AFOs) are most commonly prescribed to assist the child with positioning of the ankle and foot during gait. In young children, dynamic equinus can be improved, with a plantigrade foot position obtained and the tendency for genu recurvatum decreased.[395,432]

AFOs come in various designs (Fig. 24–14). Solid ankle AFOs are used when minimal dorsiflexion of the ankle is possible. They afford the easiest fit of the various types. Hinged AFOs permit a more natural gait by allowing some dorsiflexion during stance but blocking plantar flexion, and therefore eliminating equinus.[77,379] Hinged AFOs are bulkier and break more frequently, and they offer no benefit if the patient cannot passively dorsiflex the ankle or if crouch gait is present with the tendency for ankle calcaneus. Ground reaction AFOs have an anterior shell of plastic across the proximal tibia and are rear entry. The goal of the ground reaction AFO is to provide a push-back on the knee during stance phase and help the knee extend. Some patients with mild crouch at the knee benefit from this design. Posterior leaf spring AFOs are fabricated from a more pliable polypropylene and have more severe posterior trim lines at the ankle. They are touted as providing a little push-off at terminal stance. They tend to break and may not provide enough hindfoot control in most patients with cerebral palsy. Gait analysis has shown that they function similarly to articulated AFOs, that is, they allow some dorsiflexion in stance phase through deformation of the brace, and they control equinus in swing phase, but they do not return energy at push-off.[327]

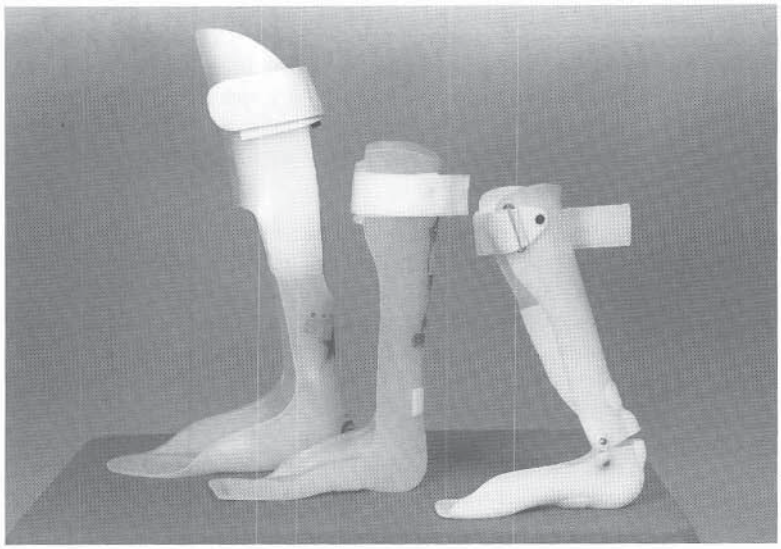
Tone-reducing AFOs incorporate an intensely molded footplate, termed an inhibitive footplate, and have higher trim lines that extend across the dorsum of the foot. The goal of these orthoses is to apply pressure similar to that of an inhibitive cast, and therefore reduce tone throughout the lower extremities. There is little scientific evidence to support this design, but it remains popular with many families.

Bracing above the knee with KAFOs or HKAFOs is generally not done in cerebral palsy. The weight and bulk of the braces usually interfere with rather than improve the child's ability to walk. Short-term use of KAFOs or knee immobilizers following soft tissue surgery for crouch gait can be helpful in retraining children to walk.

Our indications for bracing are as follows:

1. In patients with dynamic equinus, to obtain a plantigrade foot position and reduce genu recurvatum
2. When foot drop is present, to support the foot in dorsiflexion during swing phase
3. Postoperatively, while weakness is being addressed through physical therapy
4. When mild crouch is improved by use of AFOs

We do not advocate the use of AFOs in nonambulatory patients who are able to wear shoes, but we do on occasion prescribe them when shoe wear is difficult and is improved



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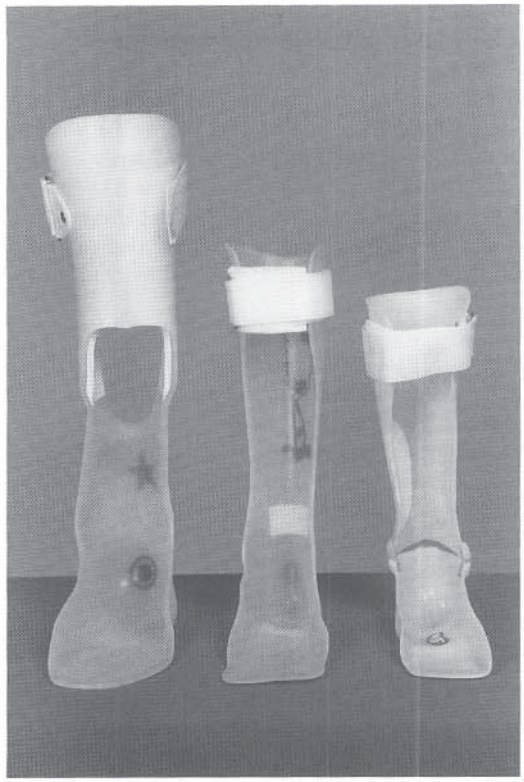

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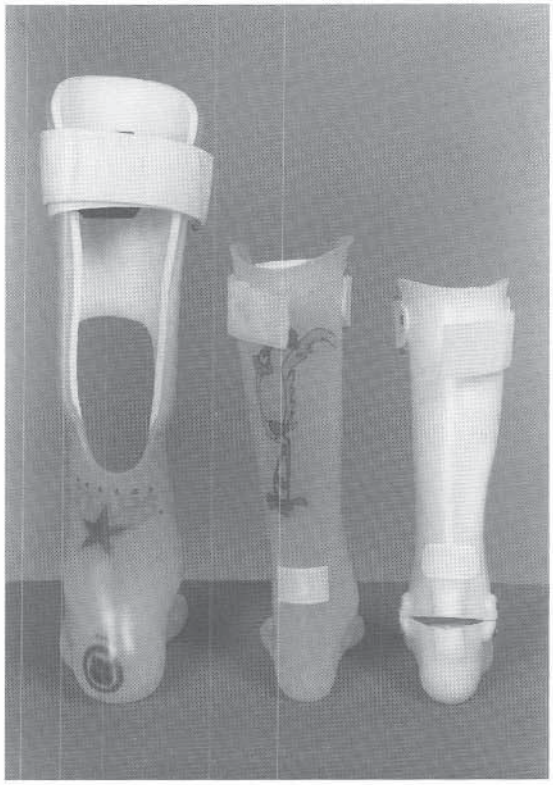


C

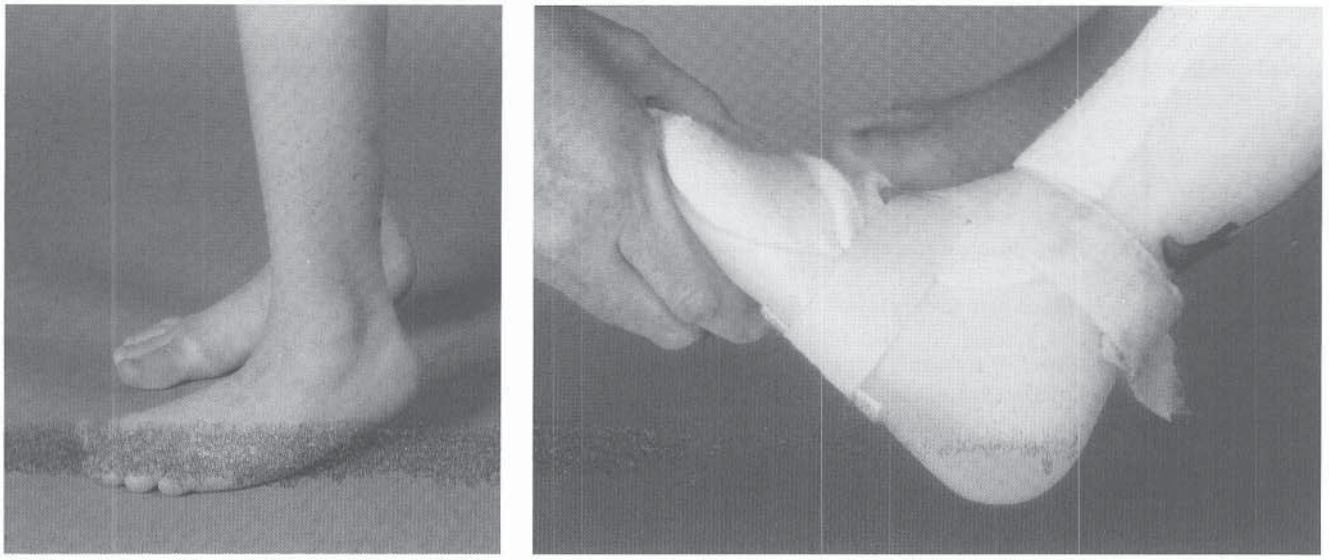
FIGURE 24–14 **A** to **C,** Different views of common ankle-foot orthoses used in patients with cerebral palsy. The orthosis on the left is a ground reaction AFO that extends across the anterior aspect of the tibia to prevent flexion of the knee in stance. The center orthosis is a conventional solid ankle AFO. The orthosis on the right is a hinged AFO that allows dorsiflexion of the ankle but prevents equinus.

with orthoses. We also do not use AFOs in the preambulatory young child, as our therapists feel that orthoses interfere with the child's ability to crawl and move about the floor.

Some patients walk worse with AFOs than without them. In patients with significant crouch gait, flexion of the knee and hip with the foot supported in an AFO either forces the child to pull up out of the braces or to toe-walk despite the orthosis (Fig. 24–15). Because toe-walking has not ever been found to be harmful, often it is better to allow the child to walk on the toes brace-free or to proceed with surgery when appropriate. Valgus deformities of the hindfeet are often difficult to brace, producing painful calluses and blisters over the prominence of the talar heads. Comfortable shoe wear without orthoses is preferable for these feet. Finally, as young children with cerebral palsy become adolescents they often refuse to wear orthoses for cosmetic reasons. Allowing the patient to make a personal choice in whether or not to wear a brace is particularly important in this age group.

Medical Treatment of Spasticity

ORAL MEDICATION. Oral pharmacotherapy has been used in an attempt to reduce tone in patients with cerebral palsy.



A B

FIGURE 24–15 **A,** Bilateral equinus in child with spastic diplegia. **B,** The orthosis is unable to maintain a plantigrade foot position due to spasticity in the gastrocsoleus and a crouch at the hip and knee.

Medications used have included valium and baclofen.[106] Side effects include somnolence. Patients with movement disorders such as dystonia or choreiform movements have been treated by various medications, with varying success.[362]

INTRATHECAL BACLOFEN. The inability to adequately reduce tone with oral baclofen because of the side effects of the large dosages led to the trials of intrathecal baclofen. When baclofen is injected into the intrathecal space, larger doses reach the target neural tissue of the spinal cord, leading to a greater reduction in tone. Baclofen, a gamma-aminobutyric acid agonist, acts at the spinal cord level to impede the release of excitatory neurotransmitters that cause spasticity.[9] Following proof of efficacy with test injections of the drug, implantable pumps filled with baclofen are surgically inserted in patients and the dose of medication is titrated. Studies have shown decreased upper and lower extremity tone and improvements in range of motion with continuous intrathecal baclofen infusion.[10] Complications occur in approximately 20 percent of patients. The complications are usually related to catheter dislodgment or fracture, but serious infection occurs in 5 percent.[9] Overdose of intrathecal baclofen is very rare but serious, producing somnolence and hypotonia, which leads to respiratory depression requiring mechanical ventilation.[406] Currently there are no strict indications for intrathecal baclofen use, but most pumps are implanted in patients with severe spasticity that interferes with positioning and function in the upper and lower extremities. Many of these patients still need orthopaedic surgery for hip subluxation or contractures.[171]

BOTULINUM TOXIN. Botulinum toxin injections have become a popular form of treatment for patients with cerebral palsy. The toxin is produced by the anaerobic bacterium *Clostridium botulinum* and works by blocking the release of acetylcholine at the neuromuscular junction. It is injected into the sites of the neuromuscular junctions, often guided by EMG. The targeted muscle then becomes weak, due to lack of innervation, until the neuromuscular junction sprouts new endings.[79] Botulinum toxin is available in two forms, Botox and Dysport.

Muscles that are considered to be candidates for injection are those that produce dynamic deformities in the absence of fixed contracture. For example, the child who walks in ankle equinus due to spasticity in the gastrocsoleus but who has dorsiflexion on passive range of motion may be a candidate for injection into this muscle. The drug begins taking effect after 2 to 3 days, and its effect wears off after approximately 3 months. There is an upper limit to the amount of botulinum toxin that can be injected at any one setting, so it works best in children in whom only one or two muscles are to be injected. It offers little benefit to the child with multiple muscle involvement.

Most of the research that has been published in the orthopaedic literature on the use of botulinum toxin in cerebral palsy has been evaluations of the response of the ankle to gastrocsoleus injection. These gait studies have found improvements in sagittal plane stance phase dorsiflexion—in other words, less equinus—following botulinum toxin injection.[142,240,461] Cosgrove, one of the first proponents of this treatment, notes that patients who are younger, who have diplegia rather than hemiplegia, and who have greater dynamic shortening of the muscle with less contracture have the best response. He and others propose that botulinum toxin may allow tendon surgery to be delayed until a later age, when there is less risk of recurrence.[96,242] In a study comparing serial casting, which has also been used to delay Achilles tendon lengthening procedures, with botulinum toxin injection into the gastrocsoleus, the results were similar between the two groups.[94]

A few patients who have undergone Botox injections into other muscles have been described. Injection of the adductors is reported to lessen scissoring, and hamstring injections may decrease crouch at the knee.[74,96]

Botulinum toxin treatment is gaining in popularity with parents and neurologists alike. Often parents adopt the philosophy that with injection, no harm will be done, and

surgery can be performed later if necessary. Although this is true, our limited experience to date has not shown less overall tendon surgery in these patients, as the benefit of the temporary nature of the drug is also its downfall—a good result will wear off, and over time repeated injections become undesirable to the children.

Our current indications for botulinum toxin injections are as follows:

1. In a child with a dynamic equinus deformity and no fixed plantar flexion contracture
2. In a child with equinus gait without multilevel crouch
3. In a child less than 4 years old who cannot tolerate AFO orthoses because of dynamic equinus
4. If parents desire injections and refuse tendon lengthening surgery

Rarely, we recommend botulinum toxin injection prior to surgery to investigate possible response to a planned surgical intervention.

Surgical Treatment

GENERAL CONSIDERATIONS. When evaluating the child with cerebral palsy for surgical intervention, a few general principles must be kept in mind. First and foremost, it is very important to speak clearly and frankly with the family about the goals of surgery and the expected postoperative course. Often the family will have unrealistic expectations of surgery, believing that it will "cure" the child of cerebral palsy and help the child walk normally. Cerebral palsy is a brain disease, and no amount of lower extremity surgery can change that fact. Tendon and bony surgery will change the length of the muscles, hopefully improving joint motion during gait and the position of the legs, but inherent difficulties with balance, spasticity, and the brain's inappropriate signals to these muscles will not change following an orthopaedic procedure. Orthopaedic surgery in cerebral palsy will make the child walk differently, hopefully walk better, but it will not make them walk "normally." The parents should be told that the child will still limp, but that it is hoped that the limp will be improved.

Weakness is a frequent short-term sequela of lower extremity surgery in cerebral palsy. Plans should be made for frequent postoperative physical therapy shortly following surgery. If at all possible, the patient should be kept ambulatory or weightbearing following the operation. Prolonged time in a wheelchair adds to the overall weakness of the limbs. Patients with osteotomies are the exception to this rule. In these children, weakness may be even more of a problem once casts are discontinued.

There is a trend toward less immobilization following soft tissue surgery. In our practice, short-leg casts are still used following Achilles tendon lengthenings, but knee immobilizers rather than casts are becoming more frequently used following hamstring lengthenings, and removable abduction bars are preferable to Petrie casts after adductor release. The goal of soft tissue surgery is greater range of motion, so avoiding overimmobilization and secondary stiffness is logical.

TIMING OF SURGERY. Many authors have recommended combining multiple tendon surgeries and osteotomies into a single surgical event for children with cerebral palsy.[52,119] Rang advises avoiding "birthday surgery," or hospitalizing the child every year for yet another soft tissue surgery or osteotomy. Although some authors argue that early one-stage surgery prevents contractures,[63] until their studies include follow-up to skeletal maturity, this conclusion is premature. Aside from avoiding repeated hospitalizations as one joint is released at a time, contractures present at one joint affect the position and movement of the rest of the extremity, so addressing all concominant contractures simultaneously with one surgery is important in order to avoid recurrence or overcorrection of deformity.[372] Because gait changes and matures until about age 7 years, when feasible it is wise to avoid surgery until this time. At this age, multiple levels can be addressed simultaneously to optimize the ambulatory skills of the patient, followed by a single aggressive course of physical therapy to maximize the benefits from the surgery.

In some patients surgery cannot be delayed until age 7. Young children with hip subluxation are best served by surgery to improve coverage of their hips when the problem is first recognized. At younger ages, hip surgery is often less extensive, often consisting of only soft tissue release, whereas the older child requires additional osteotomies of the femur and pelvis.

Other children are nearly ambulatory but their progress has been halted by contractures in the lower extremities. Earlier surgery in these patients may allow them additional range of motion to make walking less cumbersome. Adductor release for scissoring and Achilles tendon lengthening with or without hamstring lengthening in these younger children may be indicated. Parents should be forewarned about recurrent contractures requiring repeat surgery in the future.

ANESTHETIC CONCERNS. Patients with cerebral palsy experience latex allergy with increased frequency. In suspected individuals, a latex-free surgical environment is prudent. Allergy testing is available for latex products. At-risk children are those who have undergone multiple previous operations or who have indwelling latex devices such as gastrostomy tubes or ventriculoperitoneal shunts.[122,132]

Seizure medications may produce alterations in clotting. Increases in bleeding times and decreased platelet counts are known side effects of some of the antiepileptic agents. In major operations such as hip reconstruction or spinal fusion, preoperative evaluation of clotting parameters is recommended. A routine clotting profile consisting of a prothrombin time and partial thromboplastin time will miss bleeding time abnormalities.

Postoperative pain control is difficult in patients undergoing hip reconstruction, spinal surgery, and multi-level tendon surgery. We have been using continuous epidural infusions at our hospital and find that if the pain is well controlled, problems with muscle spasm are lessened. Oral medication often needs to include both pain control medication and muscle relaxants such as diazepam.

Foot Surgery

EQUINUS. Equinus is defined as increased plantar flexion due to a plantar flexion contracture or dynamic plantar flexion due to overactivity of the gastrocsoleus during gait (Fig. 24–16). Patients who walk on their toes often have equinus, but some may be on their toes as a consequence of crouch

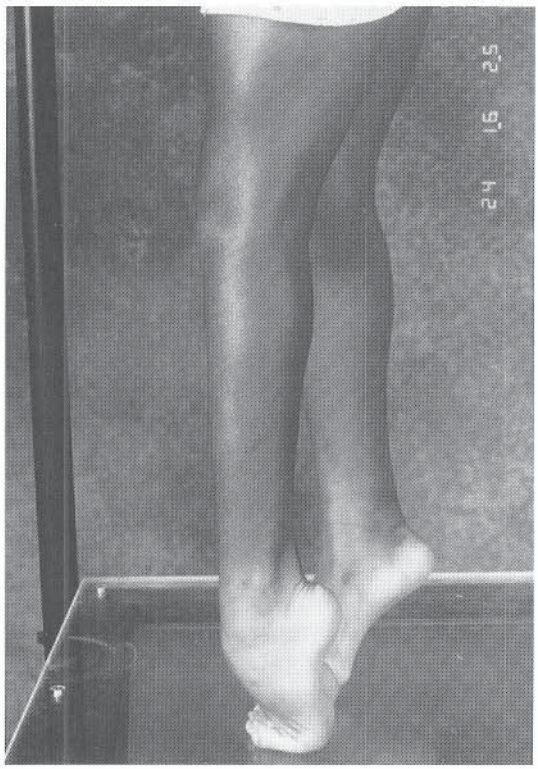


FIGURE 24–16 Bilateral severe equinus contracture in a child with cerebral palsy.

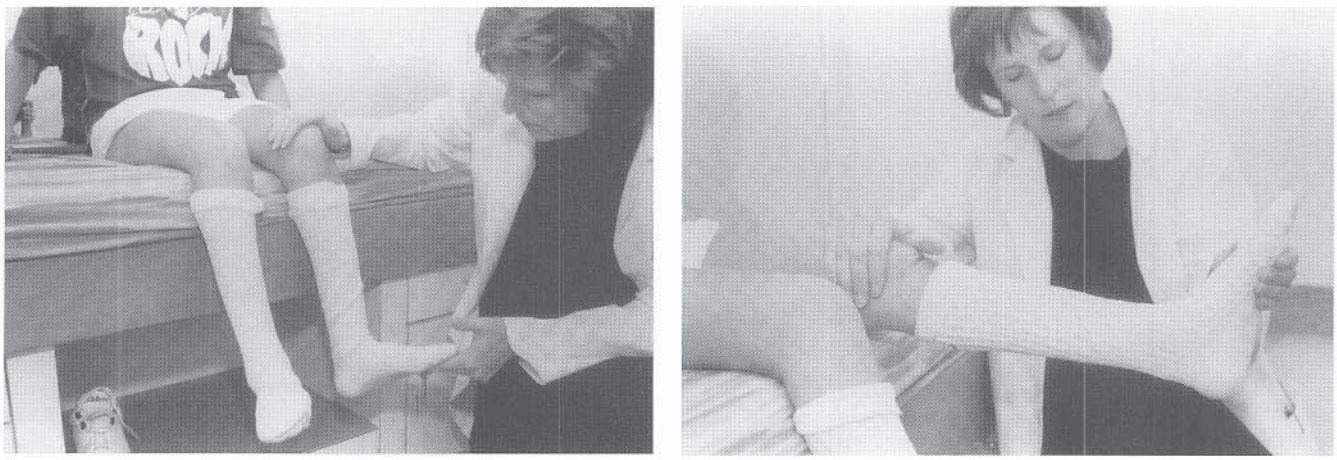
at the hip and knee and may in fact have a neutral ankle position. The physician must differentiate these two groups of children.

In a busy orthopaedic clinic it is tempting to attribute all toe-walking to cerebral palsy, but not all children who walk on their toes have cerebral palsy. Idiopathic toe-walking, also described as congenital short Achilles tendon, is a condition in which an otherwise neurologically normal child walks on the toes. Idiopathic toe-walking can be differentiated from the equinus of cerebral palsy first through the medical history: children with idiopathic toe-walking are not developmentally delayed and walk on time, but children with cerebral palsy walk at a later age than normal. Second, a child with idiopathic toe-walking has normal knee motion during gait, whereas the child with cerebral palsy usually does not fully extend the knee at the end of swing phase. Third, there will be no neurologic signs of spasticity in the child with idiopathic toe-walking; the reflexes are normal. Elegant gait studies looking at kinematics and EMG data have attempted to differentiate the two diagnoses on these grounds,[189,206,228,331] but a thorough history and physical examination by an experienced clinician are really all that is needed in the vast majority of children.

Yet another condition that produces toe-walking apart from cerebral palsy is muscular dystrophy. We have diagnosed Duchenne's muscular dystrophy in patients referred to the orthopaedic clinic for evaluation of equinus before obvious weakness affecting daily activities was present. The condition should be suspected in any young boy who walks on his toes and has a normal birth history. A delay in the age of walking is frequently seen in patients with Duchenne's muscular dystrophy, so the developmental history may not differentiate it from cerebral palsy. Testing for Gower's sign by having the child rise up rapidly from a sitting position on the floor will accentuate the presence of proximal muscle weakness. If the test for Gower's sign is suspicious, laboratory evaluation for serum muscle enzymes (creatine phosphokinase) is indicated.

Clinical examination of the child with equinus due to cerebral palsy reveals inability to fully dorsiflex the ankle. If the ankle can be passively dorsiflexed with the knee bent to 90 degrees but cannot be dorsiflexed with the knee extended, it is felt that the gastrocnemius is tight, but the soleus is not contracted (Fig. 24–17).[345] Some have used this test, named the Silverskiöld test, to determine which type of surgical lengthening to perform.

Equinus interferes with the tibia's forward progression over the foot during stance phase, and therefore shortens stride length. Because the ankle is already plantar flexed at terminal stance, little push-off power is generated, so the gait is less efficient. If the anterior tibialis is unable to lift the foot to neutral during swing phase, a foot drop results,



A B

FIGURE 24–17 **A** and **B,** Mild ankle equinus in a 17-year-old girl with spastic diplegia. The ankle cannot be dorsiflexed past neutral with the knee extended.

with possible problems in clearing the foot in swing phase and tripping. Knee disturbances also result from ankle equinus. Genu recurvatum is seen when the tibia is unable to move forward over the plantigrade foot due to tightness in the gastrocsoleus, so the knee thrusts backward during midstance (Fig. 24–18). The body and femur continue to move forward over the stationary tibia, and an extensor moment is produced at the knee. This aligns the ground reaction force anterior to the knee, reducing the demands on the quadriceps and improving the stability of the knee.[432,460] Likewise, compensatory knee flexion can be seen during stance phase in patients who walk on their toes.

Over time, ankle equinus leads to valgus positioning of the hindfoot and stretching out of the plantar arch, termed midfoot break. Although the foot may appear plantigrade, the talar head is very prominent in the longitudinal arch and the calcaneus is actually in equinus. Pain and callosities result over the head of the talus (Fig. 24–19). Hallux valgus can develop in response to the valgus positioning of the foot.

The surgical treatment for equinus is selective lengthening of the Achilles tendon or the gastrocnemius.[52] Advocates of gastrocnemius recession state that this operation preserves or even increases push-off power more than Achilles tendon lengthenings by not involving the soleus muscle or tendon.[393] Immobilization is minimized following gastrocnemius recession, whereas casting is necessary following Achilles tendon lengthening. There is a negligible risk of overcorrection into calcaneus. Perry and Hoffer believe that a gastrocnemius recession should be performed when a Silverskiöld test performed under anesthesia is positive and dynamic EMG shows more abnormality of the gastrocnemius than the soleus during gait.[344]

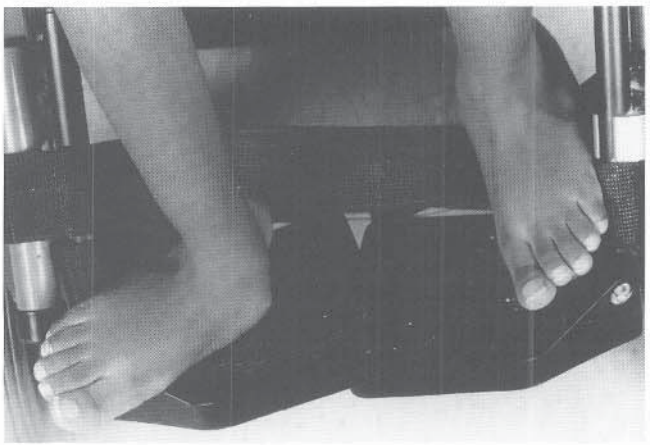


FIGURE 24–19 Equinovalgus of the right foot in an adolescent girl with spastic quadriplegia. The talar head is very prominent and a painful callus has developed.

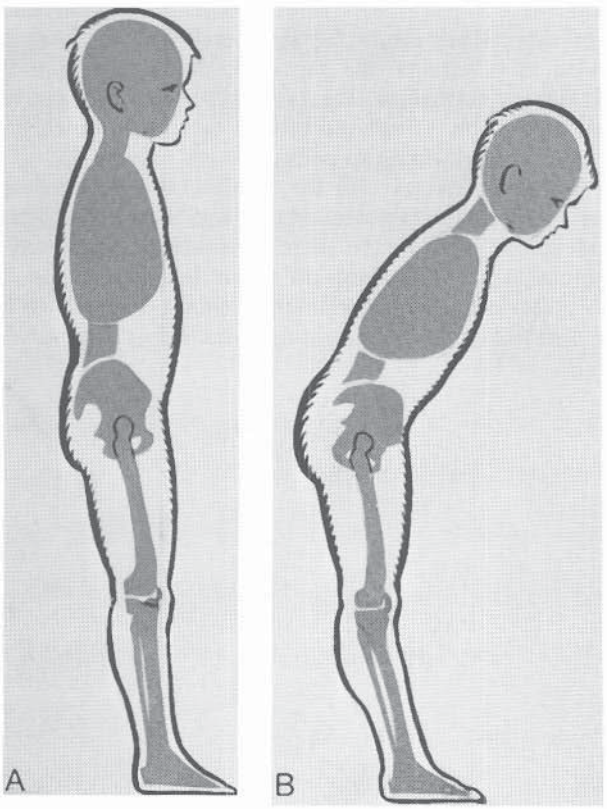


FIGURE 24–18 Hyperextension of the knees to compensate for fixed equinus deformity of the ankle. **A,** When the equinus deformity of the ankle is fixed and there is no compensatory accommodation at the hip and knee, the center of gravity is posterior to the base of support. **B,** One method of aligning the trunk and bringing the center of gravity over the feet is by knee hyperextension.

Those who prefer lengthening of the Achilles tendon recognize a greater recurrence rate following gastrocnemius recession, which is as high as 48 percent in some studies.[114,320] They also state that Achilles tendon lengthening may be done percutaneously, unlike gastrocnemius recession, and that it can be performed in ambulatory surgery without an overnight stay.[183,299] In a comparative gait analysis study, no significant difference was found between patients who had undergone gastrocnemius recession and those who had undergone lengthening of the Achilles tendon.[151]

Gastrocnemius recession may be done using the techniques of Strayer, Baker, or Vulpius. In a Vulpius procedure, the aponeurosis of the gastrocnemius is divided in a chevron fashion and the midline fibrous septum of the soleus is transected, but the soleus muscle fibers are not disturbed (Fig. 24–20). The fascia slides on the soleus and fills in with scar tissue.[220,396] The cut in the gastrocnemius is transverse in the Strayer procedure, which otherwise resembles a Vulpius recession (Fig. 24–21).[452] In the Baker technique, the gastrocnemius aponeurosis is cut tongue-in-groove and dissected free from the underlying soleus.[27] The fascia is allowed to slide on the underlying muscle, thereby increasing the overall length of the muscle, and the four corners of the aponeurosis are sutured in the lengthened position (Fig. 24–22).

Achilles tendon lengthening may be done using open or percutaneous techniques. In the open technique, a longitudinal incision is made just lateral to the Achilles tendon. The tendon is lengthened in a Z-fashion and repaired with stout nonabsorbable suture (Fig. 24–23). The tendon must be repaired with sufficient tension to avoid postoperative calcaneus.[28,29,165,168] The ankle is then immobilized in a short-leg cast for 6 weeks. An open sliding lengthening of the Achilles tendon may also be performed; this procedure does not require suturing of the tendon.

The percutaneous technique may require two or three cuts in the tendon. We prefer the two-cut technique de-

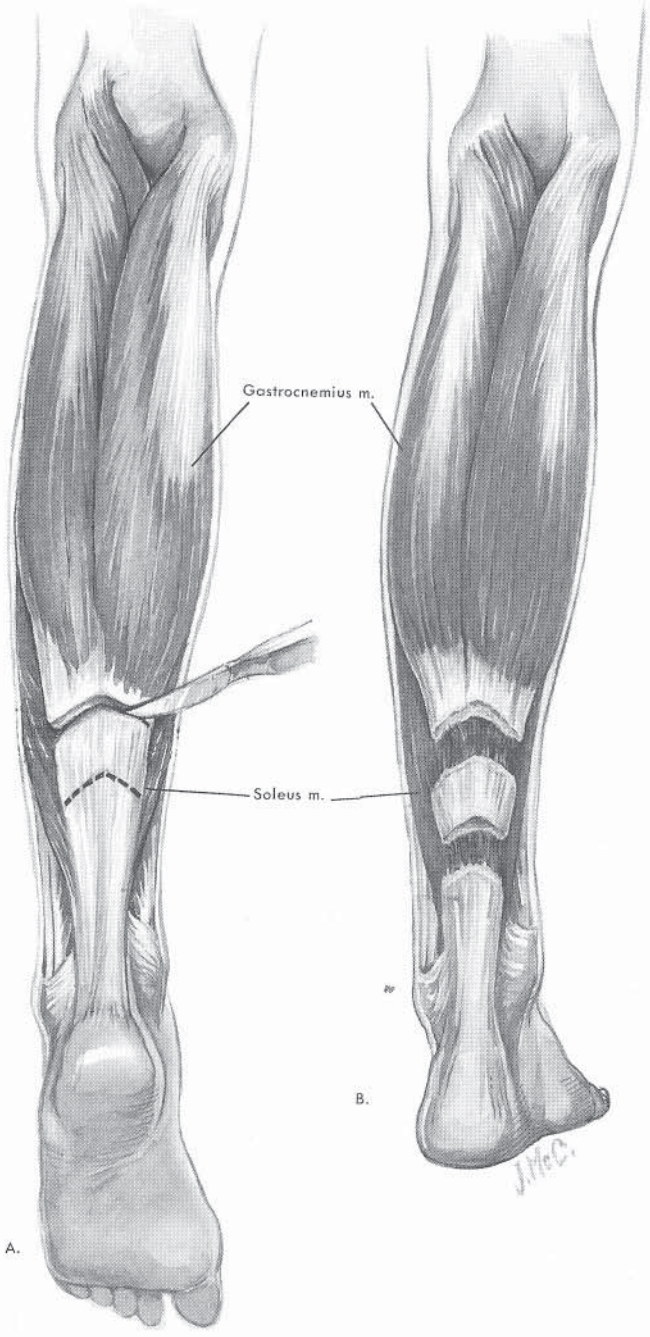


FIGURE 24–20 Lengthening of the gastrocnemius by the Vulpius technique.

scribed by White.[489] Just proximal to the calcaneal insertion, just over one-half of the tendon is divided medially by inserting a scalpel longitudinally into the center of the tendon, turning the blade out medially, and ballotting the tendon down onto the blade. The lateral one-half of the tendon is then divided using a similar maneuver more proximally. The heel is inverted into varus, and the ankle is dorsiflexed to the neutral position (Plate 24–1). The tendon can be heard lengthening as the ankle is gently manipulated. It is not unusual to hear a pop as the tendon slides and lengthens, although we prefer a more gradual and gentle lengthening to prevent overlengthening the tendon.

It is important to check the integrity of the tendon following percutaneous lengthening. The calf is squeezed while the surgeon observes the ankle. If the ankle plantar flexes when the calf is squeezed, the tendon can be presumed to be intact within the tendon sheath. If this is the case, no sutures are needed. Steri-strips are applied to the two stab wounds, followed by a cast. If there is no plantar flexion, the tendon may have been completely separated by the lengthening procedure and an open suture repair will be needed to reestablish continuity of the tendon. This is a very rare occurrence; usually the percutaneous technique proceeds without complication. Again, a short-leg cast is applied and used for 6 weeks, with weightbearing encouraged.

Hoke has described a three-cut technique for percutaneous heel cord lengthening.[212] One lateral and two medial cuts are made in the tendon and the ankle is then dorsiflexed as the tendon slides on itself and lengthens. Postoperative care is identical to that following the two-cut technique.

Neurectomy of either the gastrocnemius or soleus has been performed in conjunction with Achilles tendon lengthening in children with cerebral palsy. Although some groups have reported superior results with neurectomy,[75,133] most feel that the potential for overcorrection and calcaneus gait is greater. We do not advocate neurectomy in the treatment of ankle equinus, for that reason.

Yet another procedure that has been described to address equinus is anterior transposition of the Achilles tendon. The tendon is detached from its insertion on the calcaneal apophysis and is reattached anterior to the flexor hallucis longus tendon into a drill hole in the calcaneus.[356,453,476] Although some have reported satisfaction with this technique, we have no experience with it.

Following either heel cord lengthening or gastrocnemius recession, patients may tend to flex their knees following surgery. This is due to muscle spasm in the hamstrings and gastrocnemius (which crosses the knee joint). We find that use of a knee immobilizer for the first few days to weeks can help reduce the amount of muscle spasm and prevent the development of a postoperative knee flexion contracture.

An AFO is prescribed postoperatively in some patients. Children who have a foot drop in swing phase preoperatively will continue to have foot drop following heel cord lengthening, and patients and families should be forewarned of the continued need for orthoses following surgery to support the foot and improve toe clearance in swing phase. Many younger children benefit from the stability of an AFO postoperatively, while most older children and teens specifically state that to their minds, the goal of heel cord lengthening is to rid them of braces on their feet.

Complications are rare following heel cord lengthening or gastrocnemius lengthening. Recurrent equinus is the greatest risk, occurring in approximately 15 to 26 percent and correlating strongly with the age of the patient at surgery. Children who undergo heel cord lengthening at age 4 years or younger are particularly at risk for recurrence.[177,252,370] It is impossible to know whether it is the greater amount of growth the younger child has following heel cord surgery that leads to the high rate of recurrence or the preselection of those young children with the greatest tone to undergo surgery at an early age after failure of nonoperative treatment that leads to recurrent contracture. In a long-term study by Rattey

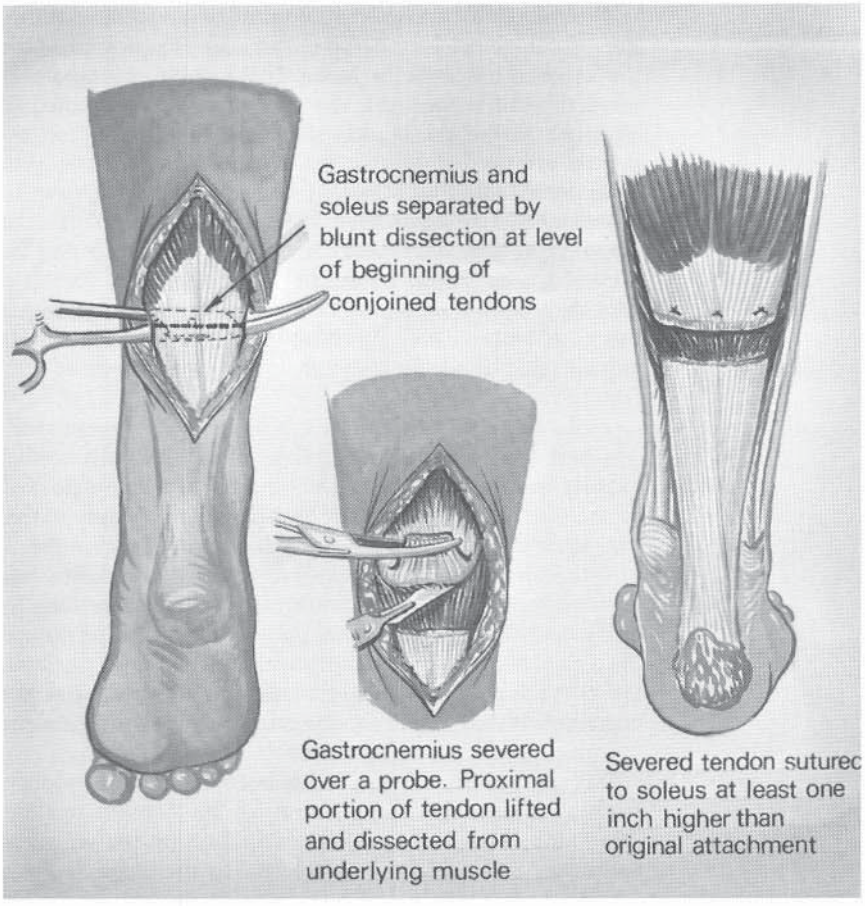


FIGURE 24–21 Distal recession of the gastrocnemius, the Strayer technique.

and colleagues, 26 percent of patients who had undergone Z-lengthening of the Achilles tendon required repeat lengthening for recurrent equinus at an average follow-up of 10 years. The risk of recurrent contracture was greater for patients with hemiplegia (41 percent) than for those with diplegia (18 percent).[370] Grant and associates found that inability to actively dorsiflex the foot preoperatively increased the risk for recurrence.[178] Repeat surgery is possible, and is quite commonly performed. With repeated lengthenings, the Achilles tendon becomes scarred and adherent, so that usually repeat lengthening must be done using the open technique.[177] Patients who have undergone gastrocnemius recession have a higher rate of recurrence than those who are treated by tendon lengthening.

Calcaneus deformity, defined as excessive dorsiflexion of the ankle during stance phase, may result for two reasons. First, the tendon may simply be overlengthened or may have lost continuity, and for all practical purposes it may have ruptured. When performing lengthening, the surgeon must be careful to bring the foot just to neutral. Careless overstretching of the tendon can lead to excessive length and calcaneus gait, with poor push-off and a tendency to develop progressive crouch of the knees.

The second reason calcaneus may occur following lengthening of the gastrocnemius or Achilles tendon is an unrecognized flexion contracture of the knee. If the knee remains crouched and the Achilles tendon becomes longer, the ankle sags into excessive dorsiflexion during stance phase and gait loses efficiency. A meticulous physical examination prior to surgery that assesses tone and range of motion of the knee is critical. If there is an increased popliteal angle, and if the child exhibits excessive knee flexion during stance phase or the inability to straighten out the leg as the heel makes contact with the ground, the patient will most likely develop postoperative calcaneus if the knee is not surgically addressed at the same sitting (Fig. 24–24).

The risk of calcaneus gait as a complication of heel cord lengthening averages approximately 5 percent. Segal and colleagues performed a gait analysis study in 20 children who had heel cord lengthenings an average of 5 years after surgery. They found that calcaneus could be documented on kinematic graphs in 30 percent of the patients.[419] Although this figure seems high compared with previous studies, it does highlight the need to address all levels of deformity during surgery and to avoid overlengthening at all costs.[370] Mathematical equations have been derived to calculate the exact amount of lengthening required for a particular patient, but most find the use of these equations somewhat cumbersome.[168]

In our clinical practice, we prefer the percutaneous approach to Achilles tendon lengthening in the surgical treatment of equinus. Postoperative immobilization is used for 6 weeks, and AFOs are prescribed on an individual basis. We agree with the philosophy that "a little equinus is better than any calcaneus," and avoid overlengthening. Most important, we thoroughly evaluate the child for other joint involvement, and we usually perform surgery on multiple levels of the lower extremity at the same time. We do see

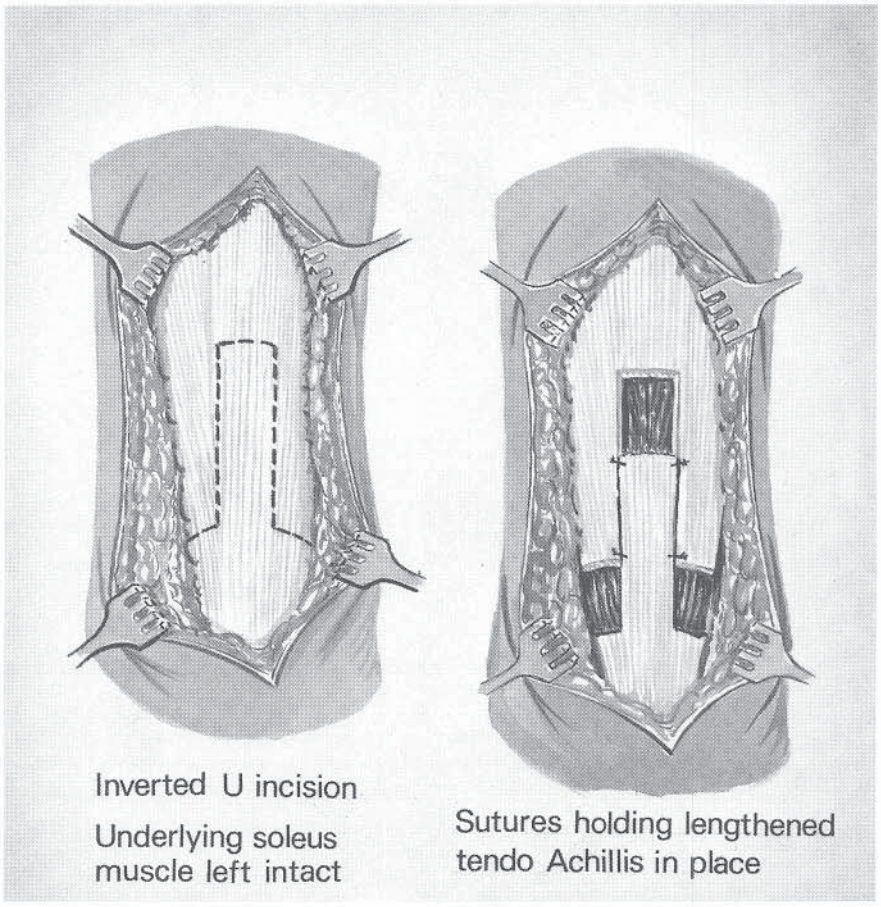


FIGURE 24–22 Tongue-in-groove lengthening of the gastrocnemius aponeurosis in its middle third, the Baker technique.

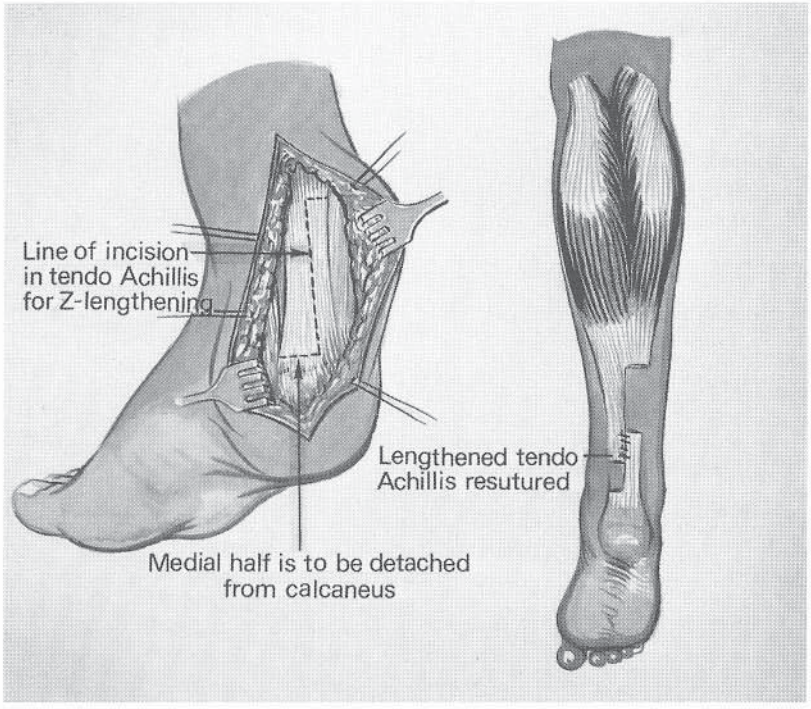


FIGURE 24–23 Z-lengthening of the Achilles tendon.

Technique for Percutaneous Achilles Tendon Lengthening

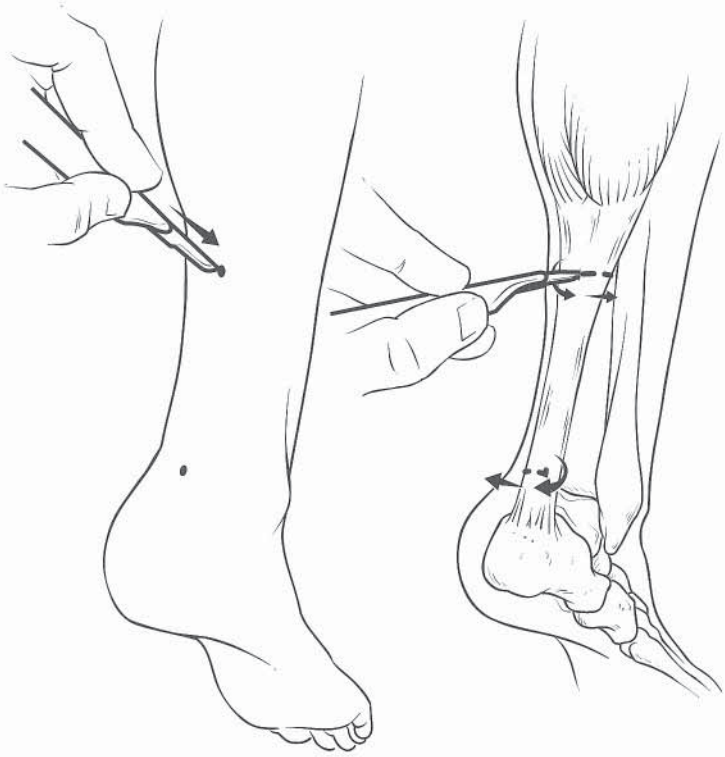
A, The two levels of lengthening are planned. The proximal level should be distal to the musculotendinous junction. The distal level should be close to the insertion into the calcaneus. The distance between the two cuts varies with the severity of contracture. The knife is inserted through a vertical stab incision into the tendon.

B, The scalpel is then rotated so that slightly more than half of the tendon is transected laterally at the proximal site and medially at the distal site.

C, The ankle is then dorsiflexed with gentle pressure until the desired degree of dorsiflexion is obtained. As the tendon lengthens, a "crackling" sound can be heard. If the tendon lengthens suddenly under force, a louder "pop" is heard. The surgeon should squeeze the calf and watch for plantar flexion of the ankle to ensure continuity of the tendon.

D, A short-leg cast and knee immobilizer are then applied.

PLATE 24–1. Technique for Percutaneous Achilles Tendon Lengthening



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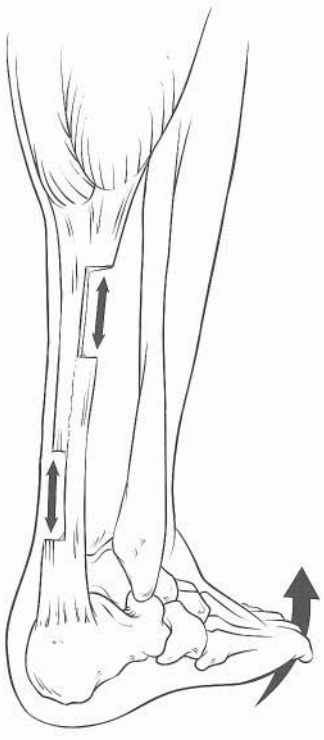

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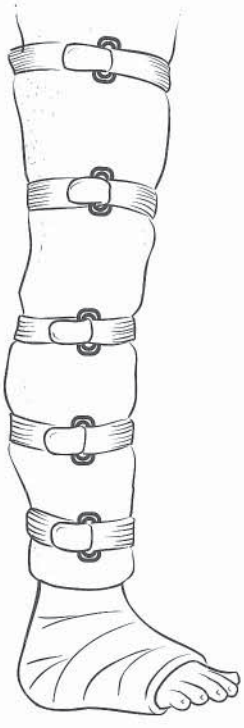

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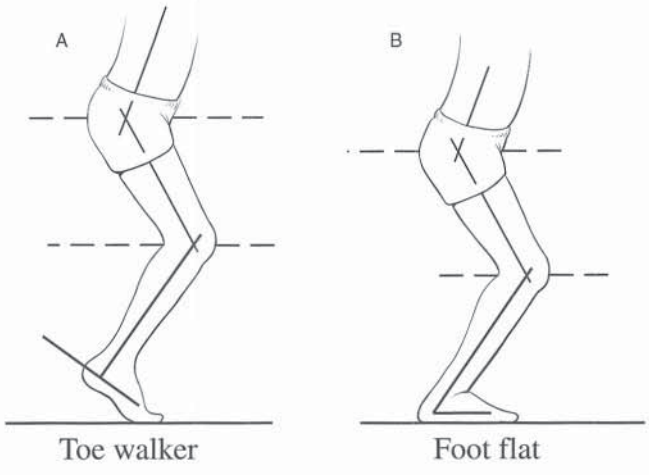


FIGURE 24–24 **A,** Child with a crouched knee and toe-toe gait. The ankle is actually in neutral position, but the child walks on the toes to compensate for the flexed knee. **B,** Following inappropriate Achilles tendon lengthening, flexion at the knee remains unchanged and the ankle is now excessively dorsiflexed, resulting in calcaneus gait.

some recurrent contractures with growth, and we reoperate when these contractures interfere with gait.

EQUINOVARUS DEFORMITY. Equinovarus deformity of the foot results from muscle imbalance in which the invertors of the foot, specifically the posterior tibialis and anterior tibialis muscles, overpower the evertors (the peroneals). The gastrocnemius contributes equinus to the deformity. Patients complain of walking on the lateral border of the inverted foot, and painful callosities can develop laterally over the fifth metatarsal. The deformity is most prevalent in patients with spastic hemiplegia (Fig. 24–25).

Gait analysis shows an internal foot progression angle due to the inversion of the foot. Ankle equinus is present, and foot drop may be evident during swing phase.

Nonoperative treatment is poorly tolerated. Bracing of a supple deformity can be done, but if the muscles are very spastic, the orthoses can exacerbate the blisters or calluses over the lateral border of the foot. Botulinum toxin injections have been tried, but there are no published studies showing long-term efficacy in equinovarus.

Surgery is indicated to improve foot contact and relieve pain and skin changes. If the foot can be passively corrected with manipulation in the clinic to a neutral position, tendon surgery can be performed. Lengthening procedures and split transfers have been described, and more detail will be provided about these procedures. If the deformity is stiff and the foot cannot be manipulated into a plantigrade position, bony surgery will be necessary to fully correct the deformity.

Surgical decision making in the soft tissue correction of spastic equinovarus focuses on distinguishing an anterior tibialis etiology from a posterior tibialis etiology of the deformity. The evaluation should start in the clinic with a careful physical examination. The confusion test is quite helpful in this setting. The patient is asked to flex the hip against resistance while seated, and the ankle is inspected. In most children with cerebral palsy the anterior tibialis fires while the hip is flexed; this is considered a positive confusion test (Fig. 24–26). The presence or absence of the reflex is not a significant finding in cerebral palsy, and the reflex has been seen in some normal children.[115] The examiner should look for the *position* of the foot as the anterior tibialis fires. If supination of the forefoot is seen, the anterior tibialis is most likely contributing to the equinovarus deformity. If the foot dorsiflexes without supinating, it is less likely to respond to surgery on the anterior tibialis.

Next, the examiner feels for spasticity in the posterior tibialis muscle. Passive manipulation of the hindfoot into valgus while feeling the posterior tibialis tendon can help the physician appreciate tightness in the posterior tibialis. The examiner should look at where the varus appears to be located. Hindfoot varus is most likely due to overpull of the posterior tibialis, whereas forefoot supination is more commonly due to the anterior tibialis. Persistent varus in both stance and swing phase is usually secondary to overactivity of the posterior tibialis. The examiner should observe for tension in the anterior tibialis and posterior tibialis during gait; the spastic tendon may be visibly taut as the patient walks.[236]

Gait analysis with dynamic EMG can help determine which muscle is acting inappropriately.[227,344,458,459] Surface electrodes suffice for measuring anterior tibialis activity, but the posterior tibialis is quite deep, and a fine wire needle electrode is necessary to measure its signal. The anterior tibialis should be quiet in midstance. Signal from the anterior tibialis throughout stance phase is indicative of overactivity (Fig. 24–27). The posterior tibialis serves to stabilize the foot during stance phase. Early-onset signal and prolongation of the activity during swing phase indicate an abnormality in the control of the posterior tibialis.[70]

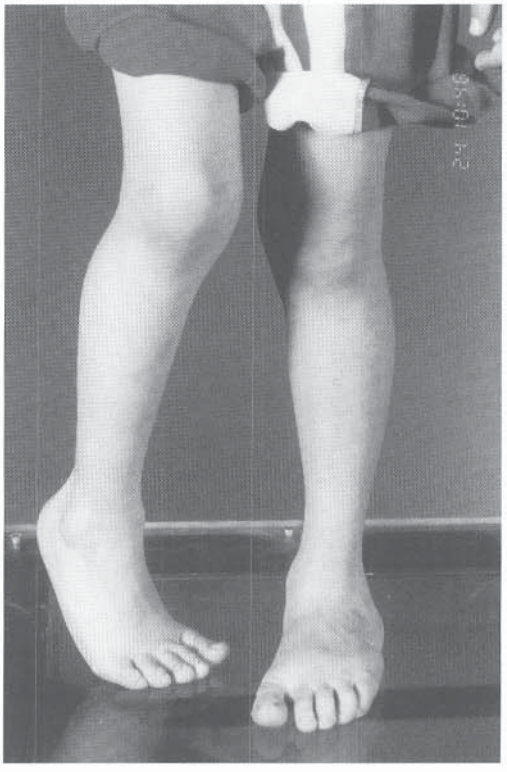


FIGURE 24–25 Equinovarus deformity in a 5-year-old boy with right spastic hemiplegia.

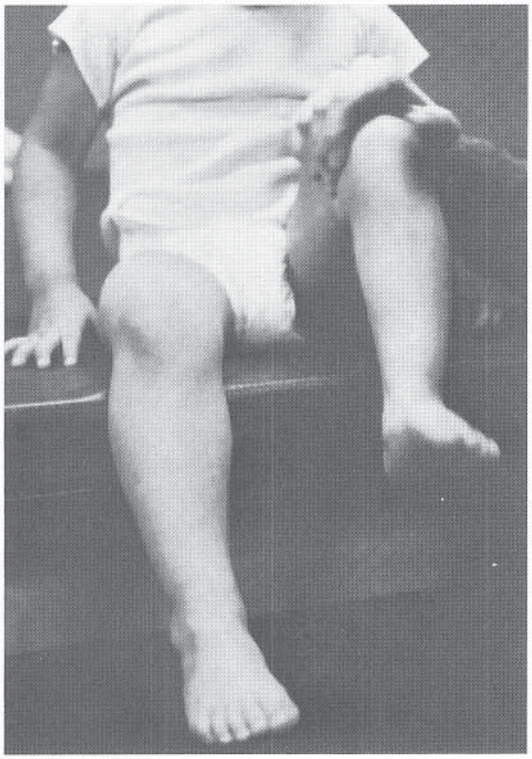


FIGURE 24–26 Confusion test: The anterior tibialis function can be tested by having the patient flex the hip against resistance. The foot will either dorsiflex or invert.

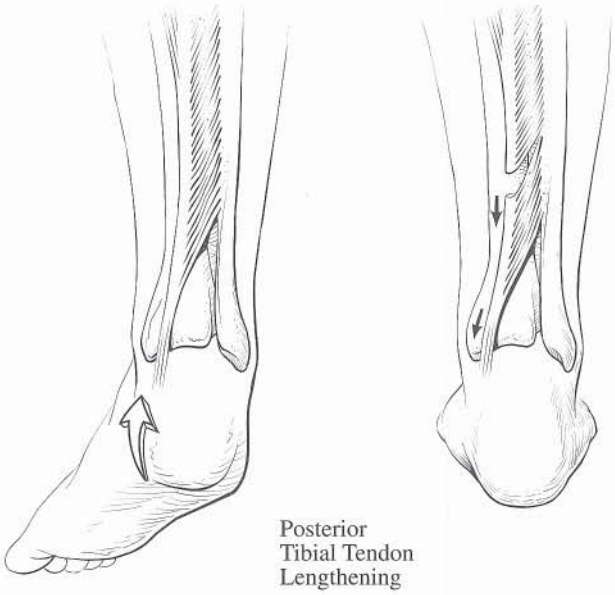


FIGURE 24–28 An intramuscular lengthening of the posterior tibial tendon can be performed in the distal one-third of the leg in patients with mild varus deformity.

Some patients with equinovarus feet have preexisting foot drops during swing phase. When this is seen, either in the clinic or during gait analysis, further weakening by anterior tibialis surgery will lead to a significant foot drop postoperatively, and an AFO will be needed.

Posterior Tibialis Tendon Lengthening. Surgical options for equinovarus deformity start with lengthening of the posterior tibialis tendon. This is usually done in conjunction with an Achilles tendon lengthening procedure in young patients with mild varus in conjunction with equinus. The tendon may be Z-lengthened distally, or an intramuscular lengthening can be performed in the distal one-third of the calf (Fig. 24–28).[398] The patient is placed in a short-leg cast postoperatively for approximately 6 weeks. Complications consist of recurrence of the deformity[387] and the development of postoperative valgus. The posterior tibialis is weakened by the lengthening, but the rebalancing of forces around the foot does not occur to the same extent as in a tendon transfer. Yet, in many milder cases, this is sufficient to obtain a plantigrade foot.

Transfer of the Posterior Tibialis Tendon to the Dorsum. Anterior transfer of the entire posterior tibialis tendon has been performed as a treatment for equinovarus.[191,389] Calcaneovalgus can be a disastrous result of this procedure, occurring in up to 68 percent of patients.[415,479] The only published indication for this procedure is if the posterior tibialis is completely silent during stance phase and active during swing.[293,344] Otherwise the posterior tibialis serves as a dorsiflexor during stance, and little resistance to the calcaneus position is offered by the lengthened Achilles tendon. We do not perform this operation in children with cerebral palsy at our hospital.

Split Posterior Tibialis Tendon Transfer. Overcorrection with complete tendon transfers led to the popularization of split tendon transfers in children with cerebral palsy. One of the most common operations for the treatment of the equinovarus foot is the split posterior tibialis tendon transfer, popularized by Kaufer and then Green.[182,232] In this procedure, the posterior one-half of the posterior tibialis tendon is detached from its insertion, split proximally to the level just proximal to the ankle, rerouted posterior to the tibia and fibula, and then woven into the tendon of the peroneus brevis (Fig. 24–29). The remaining posterior tibialis tendon attached to the navicular is then balanced by the lateral one-half of the transferred tendon, which acts as an evertor. In the original description of the operation four incisions were used, but Thompson has described using a Cincinnati incision for this transfer. Usually an Achilles tendon lengthening procedure is also required to address the equinus.

The prerequisite for a successful split posterior tibialis tendon transfer on gait analysis is swing phase activity seen

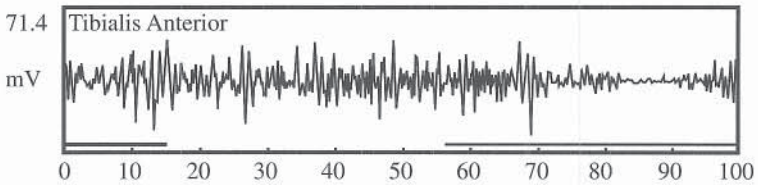


FIGURE 24–27 EMG tracing showing activity of the tibialis anterior muscle during gait in a child with an equinovarus foot due to cerebral palsy. The tibialis anterior normally fires at foot contact to gradually lower the foot to the ground, and again during swing phase to prevent foot drop. This EMG tracing shows activity throughout stance phase, occupying from 0 to 70 percent of this gait cycle, and lack of activity during swing phase.

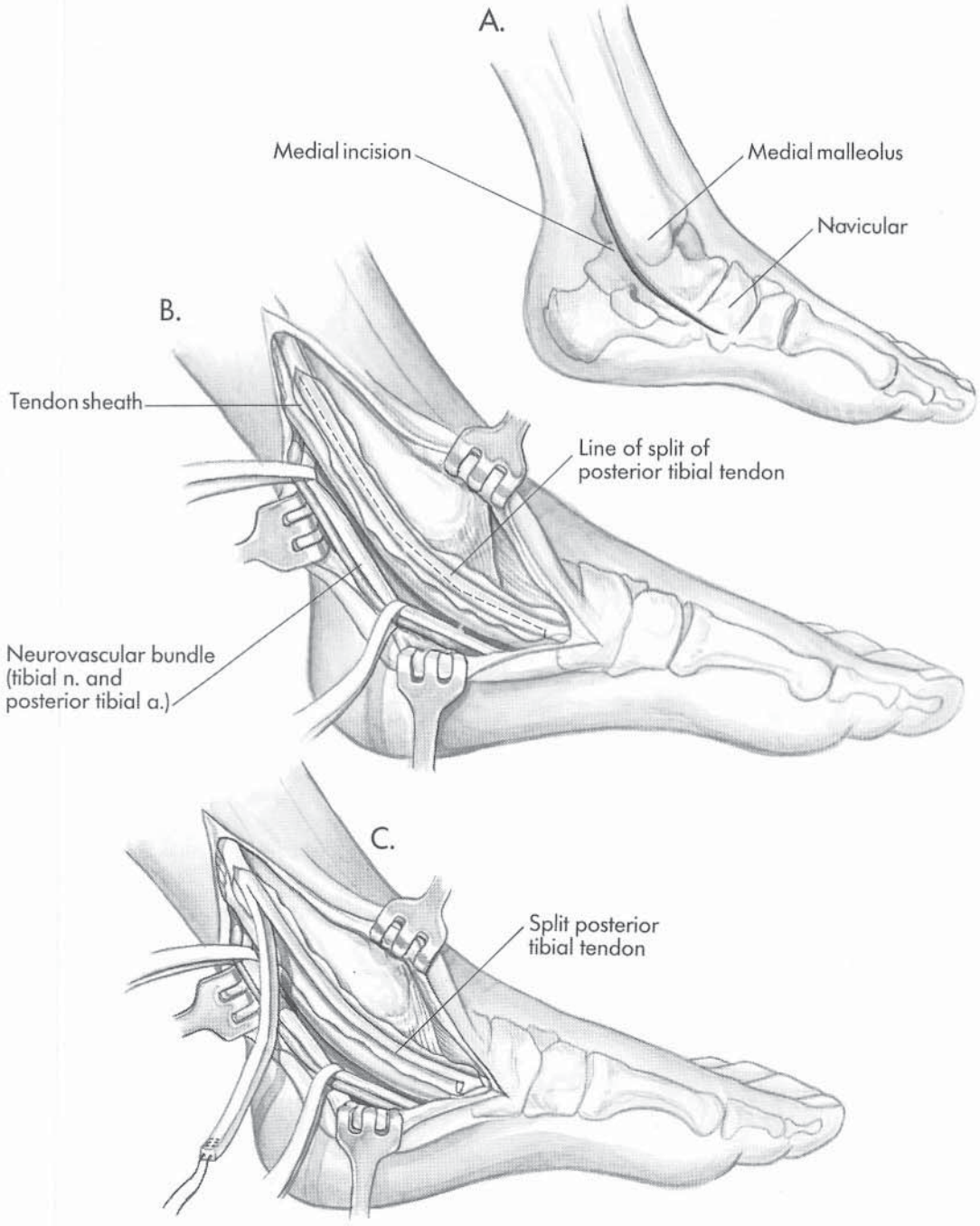


FIGURE 24–29 Surgical technique of the split posterior tendon transfer. **A,** Line of medial incision. Two smaller incisions may also be utilized. **B** and **C,** The posterior tibial tendon is split longitudinally and proximally to its musculotendinous junction; the dorsal portion of the tendon is left intact and attached to the navicular. The retinaculum of the ankle is not divided. Note that the neurovascular bundle and long toe flexors are gently retracted posteriorly.

on dynamic EMG. Yet several authors have published successful series of split posterior tibialis tendon transfers in which pre- or postoperative EMG was not performed.[236,467]

Usually patients can walk without orthoses following this tendon transfer.[467] As in any tendon transfer, the deformity must be passively correctable preoperatively for postoperative correction to be expected. Recurrent varus may be a complication following split posterior tibialis tendon transfer and usually results from inappropriate patient selection, where the deformity is too rigid. Green states that overcorrection into valgus has not occurred in his patients,[181] but we have occasionally seen overcorrection in older patients in our outpatient clinics several years following tendon transfer.

A variation on the classic split posterior tibialis tendon transfer to the lateral side of the foot has been studied. In this procedure, the anterior one-half of the posterior tibialis tendon is transferred anteriorly through the interosseous membrane to the dorsum of the foot. It is felt that this

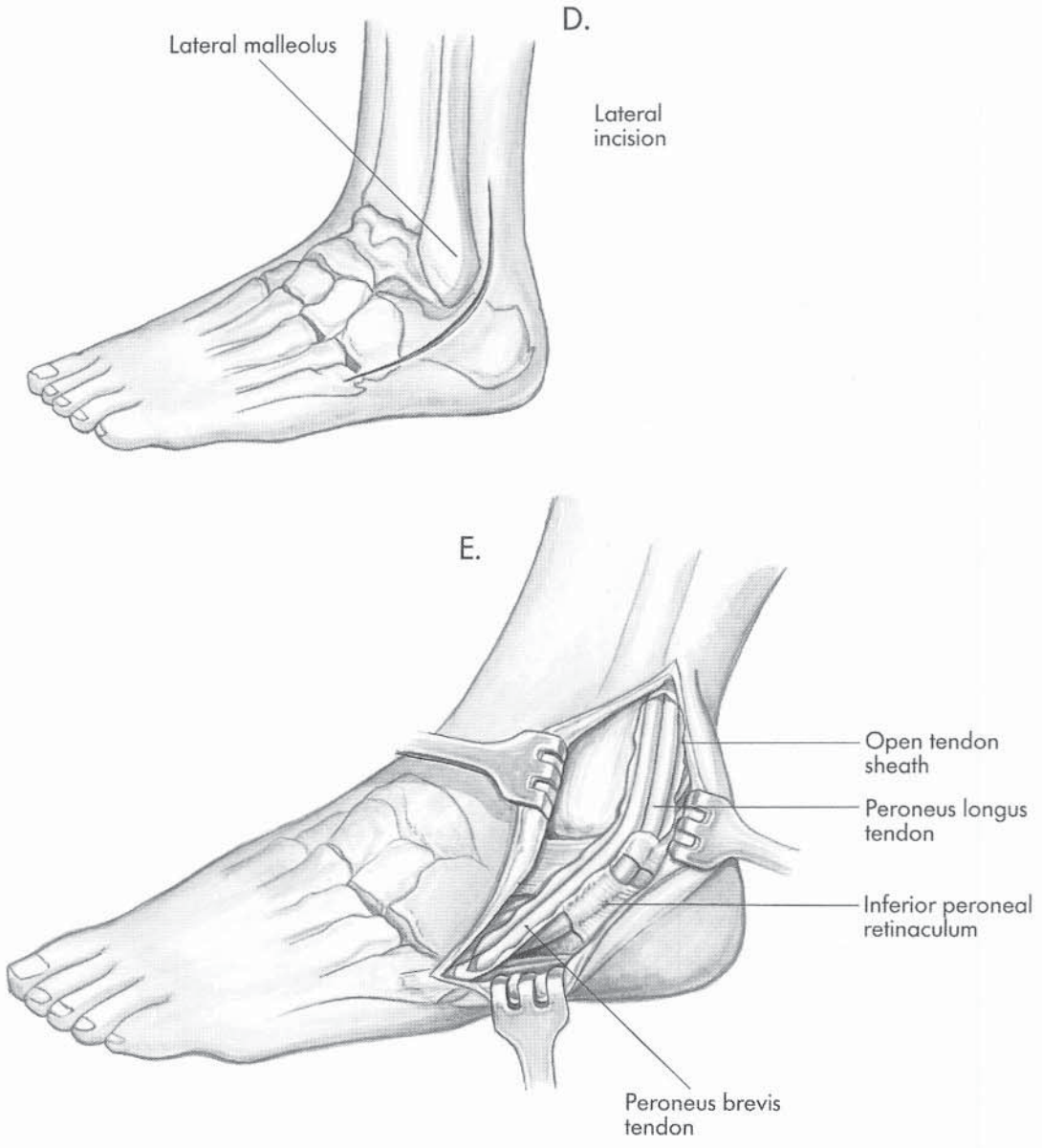


FIGURE 24–29 *Continued.* **D,** Lateral view of the foot and ankle showing the line of the lateral skin incision. Here also, two smaller incisions may be utilized. **E,** The peroneus brevis and longus tendons are identified and the tendon sheath is opened, exposing the peroneal tendons.

Illustration continued on following page

split transfer might assist dorsiflexion with less calcaneus deformity than in transfer of the entire tendon, because the posterior one-half of the tendon remains intact. Although early studies have shown promising results, this procedure has yet to be adopted universally.[304,400]

Split Anterior Tibialis Tendon Transfer. Another surgical option for the correction of spastic equinovarus is the split anterior tibialis tendon transfer (Plate 24–2). In this procedure, the lateral half of the anterior tibialis is detached from the base of the first metatarsal and split up proximal to the level of the ankle. The tendon is then passed beneath the extensor retinaculum, inserted through a bone tunnel into the cuboid bone, and tied over a button on the sole of the foot under tension with the foot positioned in 5 to 10 degrees of dorsiflexion. Again, the procedure is usually combined with an Achilles tendon lengthening procedure. When done in combination with a posterior tibialis lengthening, it is known as the Rancho procedure.

The clinical prerequisite for the procedure is overactivity of the anterior tibialis causing supination rather than dorsiflexion of the foot during the confusion test. Gait analysis will show inappropriate EMG activity of the anterior tibialis during stance phase, leading to a continuous signal.[208] Patients with profoundly weak anterior tibialis tendons and notable foot drops should not undergo split anterior tibialis tendon transfers.

The published results of split anterior tibialis tendon transfer and split posterior tibialis tendon transfer are similar and encouraging, with nearly all patients doing well, brace-free, and without overcorrection.[384] As in posterior tibialis tendon surgery, if the deformity is not flexible preopera-

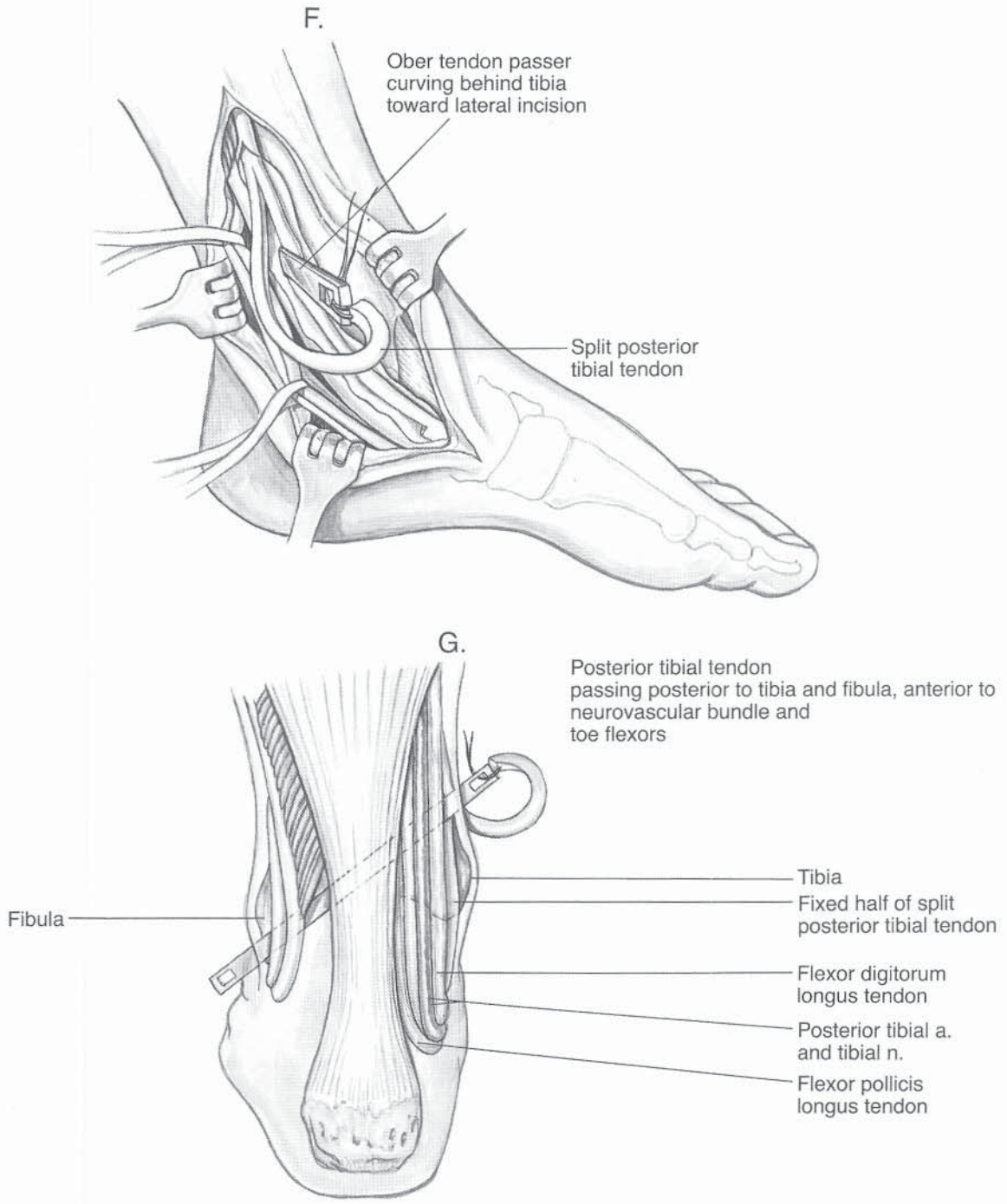


FIGURE 24–29 *Continued.* **F** and **G,** The split half of the posterior tibial tendon is transferred to the lateral side of the foot, passing anterior to the neurovascular bundle, long toe flexor tendons, and flexor digitorum longus. It is inferior to the lateral malleolus and deep to the peroneus brevis tendon.

tively, the split anterior tibialis tendon transfer will not be successful in correcting the deformity.[31]

Bony Surgery. If the varus deformity of the foot is fixed and it is felt that lengthening the posterior tibialis tendon would not provide correction, tendon transfer in and of itself will be unsuccessful and bony surgery should be performed. Two choices exist. Heel varus will respond to a calcaneal osteotomy. The calcaneus is approached laterally and a laterally based wedge of bone is resected.[430] Fixation, when used, can consist of a staple or screw to approximate the osteotomy on the lateral side of the calcaneus. A nonweightbearing cast is applied until healing begins.

If the deformity is more severe and there is a component of midfoot supination that is rigid, a calcaneal osteotomy will be insufficient and triple arthrodesis should be performed. A fused position in mild valgus is preferable, as it will provide a broad weightbearing surface. Pseudarthrosis may occur,

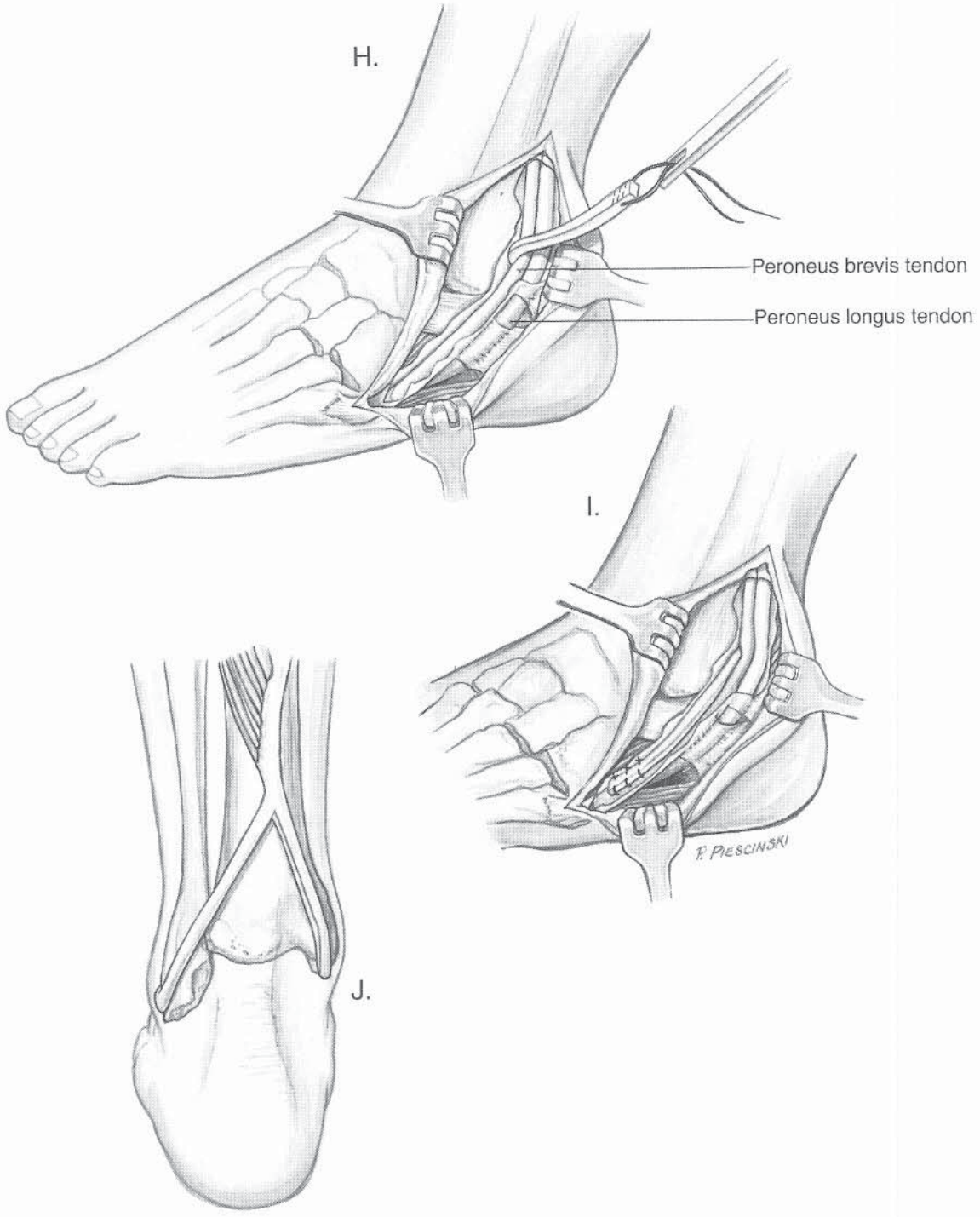


FIGURE 24–29 *Continued.* **H** and **I,** The split posterior tibial tendon is brought out into the peroneus brevis tension sheath, the tendon on it is adjusted, and it is sutured to the tendon as far distally as possible. It is best to weave it through the peroneus brevis tendon. **J,** Posterior view of the ankle and hindfoot, showing the direction of the tendon transfer. It is oblique from its musculotendinous junction above toward the tip of the lateral malleolus distally and laterally. The continuous contraction of the spastic posterior tibial tendon provides mechanical stability and control of the hindfoot in neutral position or 5 degrees of valgus inclination.

particularly in the talonavicular joint, which may or may not be symptomatic.[219] Long-term follow-up studies have reported degenerative changes in the ankles of many patients who had undergone triple arthrodesis, and some patients complained of pain limiting their ambulation. Pain at follow-up correlates with residual deformity, so it is imperative to fuse the foot in an optimal position.[471]

Even with bony procedures, muscle imbalance must be corrected, or the fusion or osteotomy will migrate over time into recurrent deformity. Often, Achilles tendon lengthening

Split Anterior Tibialis Tendon Transfer

A, A skin incision is made over the insertion of the anterior tibialis at the base of the first metatarsal.

B, A longitudinal split is made in the tendon.

C, The lateral one-half of the tendon is detached.

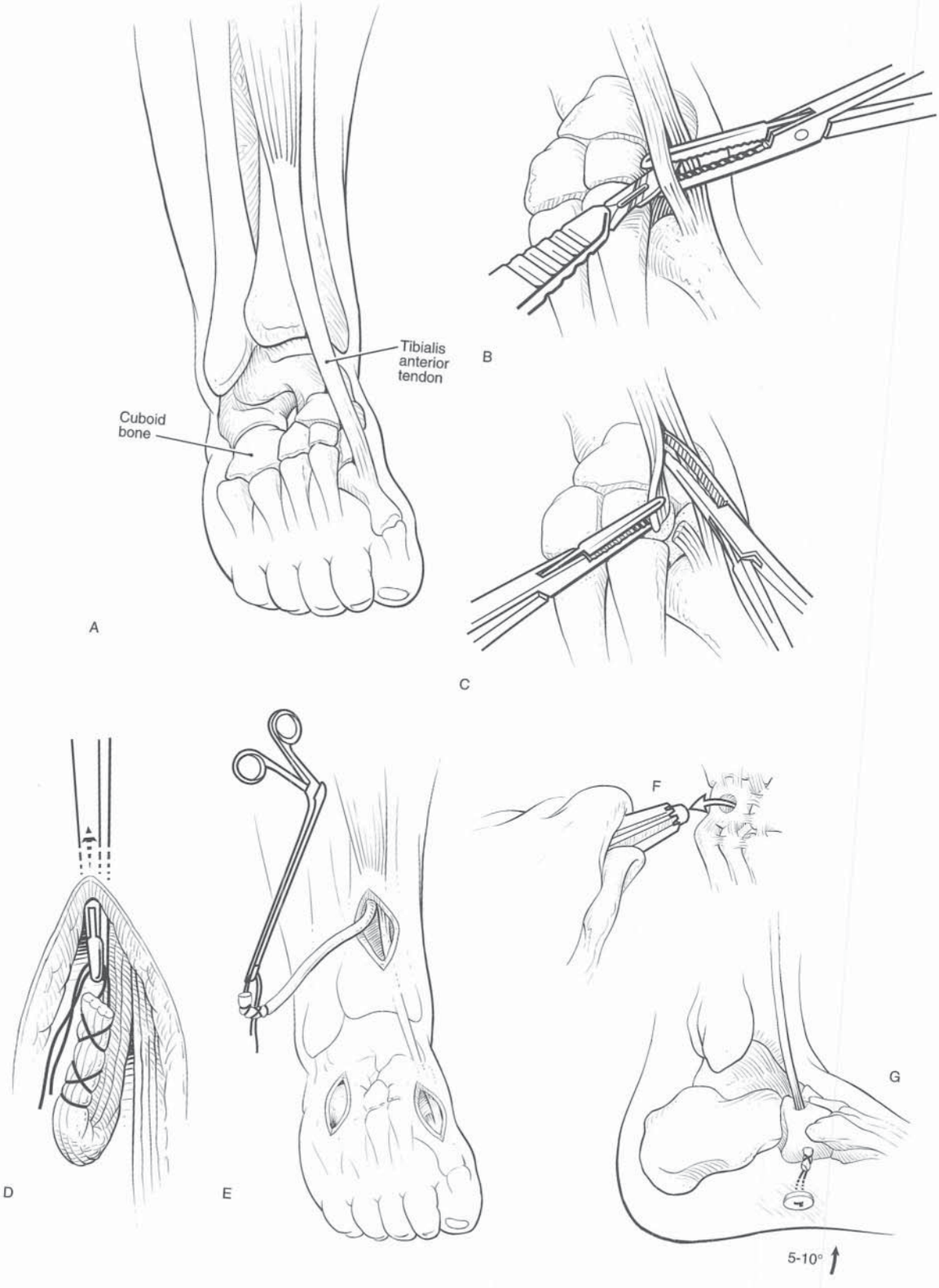
D, A stitch is woven into the detached tendon and the split is propagated proximally.

E, A second incision is made over the tibialis anterior just proximal to the extensor retinaculum. With a tendon passer, the split tendon is then delivered on its suture into the proximal wound.

F, A third incision is made over the dorsum of the cuboid. A trephine is used to create a bony tunnel in the cuboid.

G, With the foot held in neutral position, the tendon is transferred into the cuboid and the suture is passed on Keith needles out the sole of the foot. The suture is then tied over felt and a button with tension.

PLATE 24–2. Split Anterior Tibialis Tendon Transfer



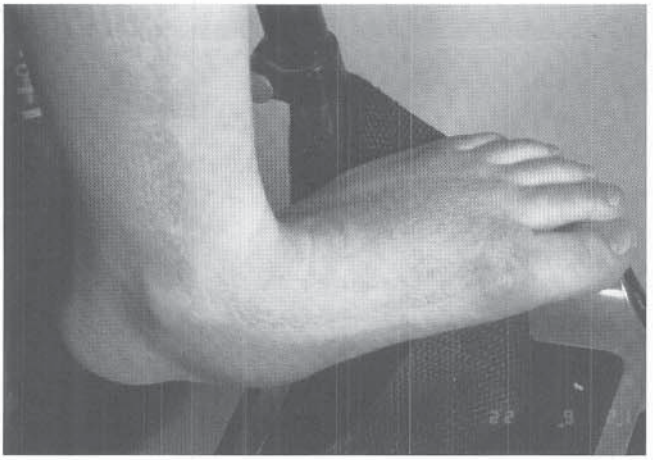


FIGURE 24–30 Patient with spastic diplegia and pes valgus. The prominent talar head was painful.

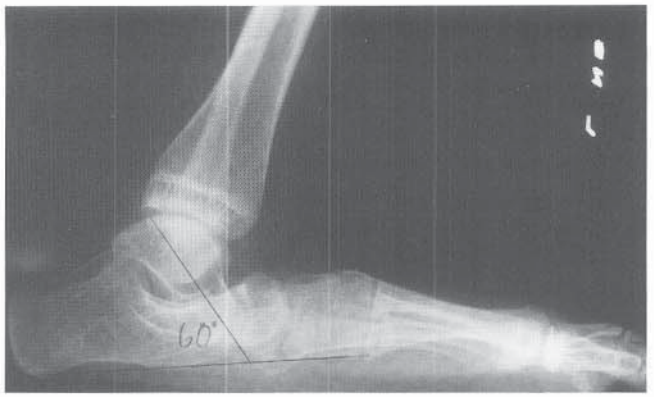


FIGURE 24–32 Lateral radiograph of patient with pes valgus. The talus is excessively plantar flexed.

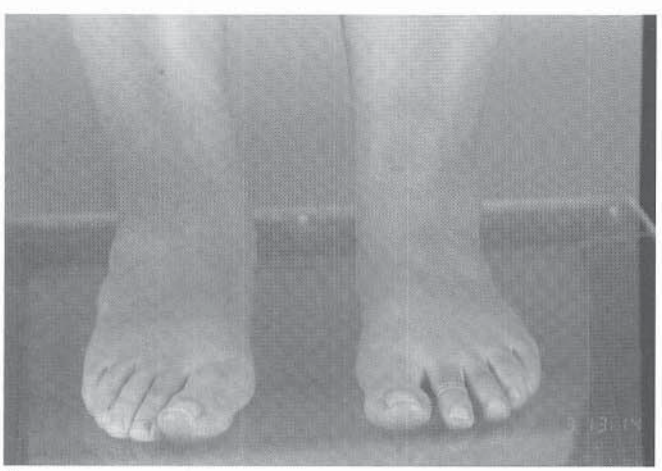
and posterior tibialis lengthening are required, but some have performed tendon transfers at the time of osteotomy or fusion.

PES VALGUS. Valgus deformity of the foot occurs in up to 25 percent of patients with cerebral palsy, being most common in older diplegic and quadriplegic patients.[41] Up to 42 percent of patients with spastic diplegia and up to 68 percent of patients with spastic quadriplegia develop equinovalgus deformities.[319] It is usually present bilaterally. Complaints are of abnormal shoe wear and of pain from calluses and blisters in the area of the talar head, which becomes very prominent in the arch of the foot (Fig. 24–30). Parents note that the ankles appear to roll in as the hindfoot valgus increases. With time, hallux valgus develops in response to the everted foot position, which may be painful (Fig. 24–31).[213]

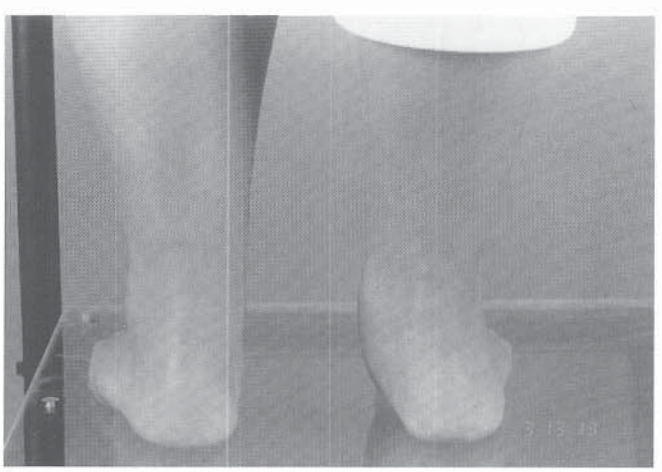
Pes valgus can be caused by spastic peroneal muscles, weakness of the posterior tibialis, and a tight gastrocsoleus, in any combination. EMG studies have shown that the peroneals may be continuous or may be phasic but inappropriate in pes valgus. The same studies found that there were some children in whom the posterior tibialis muscle was silent during stance phase, further enabling the peroneals to pull the foot out into abduction and valgus.[41,344,433]

Physical examination should assess for coexisting equinus or calcaneus deformities. Often the gastrocsoleus complex will not appear tight on initial examination. The examiner must be certain to maintain the hindfoot in varus and then passively dorsiflex the ankle. Valgus will mask equinus unless this is done. The foot may appear flat in the standing position, yet the hindfoot may be positioned in significant equinus, the talar head plantar flexed, and the midfoot overly mobile to maintain the plantigrade position. "Midfoot break" is the term used to describe plantar flexion of the talus and calcaneus, a collapsed longitudinal arch, and dorsiflexion and pronation of the forefoot.

Radiographs should be obtained in the standing position. Lateral radiographs are most helpful. The position of the hindfoot can be assessed for equinus or calcaneus deformity, and plantar flexion of the talus can be appreciated (Fig. 24–32). The navicular moves laterally, which is seen as uncovering of the talar head on an AP radiograph. Standing radiographs of the ankle should also be obtained. It is not uncommon to find ankle valgus on the AP radiograph of the ankle coexisting with hindfoot valgus. In such cases the fibular physis will migrate proximally, lying superior to its



A



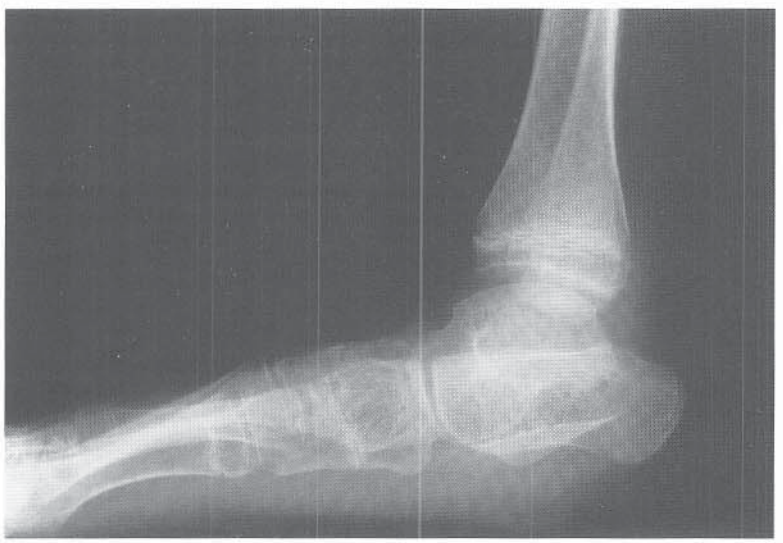
B

FIGURE 24–31 **A** and **B,** Patient with unilateral valgus deformity and symptomatic bunion.

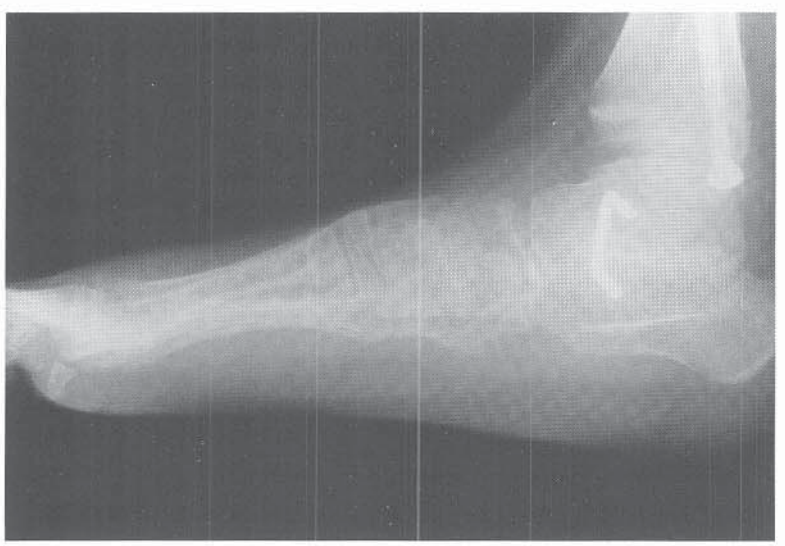
normal position opposite the joint line of the distal tibia (Fig. 24–33).

Treatment is controversial. Conservative treatment should be vigorously pursued, as shoe inserts and orthosis modifications may be adequate to relieve pain in some patients, and therefore surgery can be avoided. As long as the foot is painless, orthotic support of the valgus foot may not even be necessary, as some children do well in athletic shoes and develop symptoms only after rigid orthoses are placed on their feet.

After conservative measures have been exhausted, surgical treatment may be entertained. If the valgus is thought to be secondary to contracture of the Achilles tendon, heel cord lengthening may offer improvement in the young child. Clinically, these patients will have a normal longitudinal arch without medial prominence with the ankle held in



A



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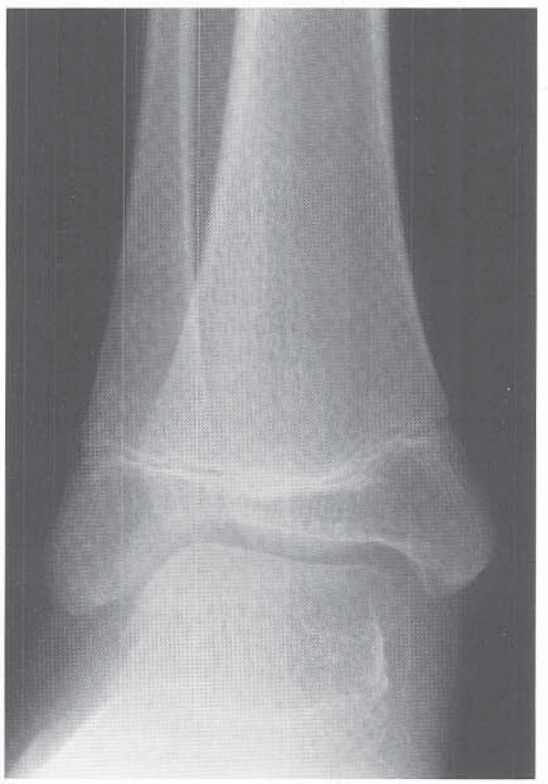

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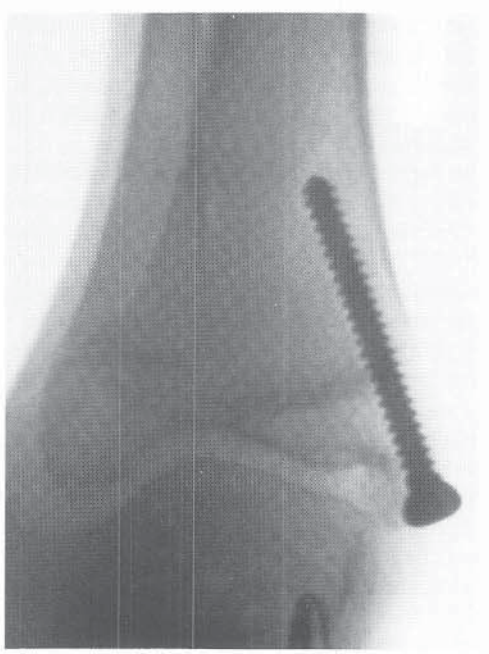


D

FIGURE 24–33 **A,** Radiographic appearance in an adolescent boy with hindfoot valgus deformity and pain in the medial arch over the prominent talar head. **B,** Standing AP radiograph of the ankle demonstrating coexistent ankle valgus that is due to the proximal location of the distal fibula. The fibular physis lies opposite the distal tibial physis rather than at the joint line. **C,** Lateral radiograph obtained 5 months following subtalar arthrodesis for correction of the hindfoot valgus. **D,** A medial malleolar epiphysiodesis using a screw was performed concomitantly to address the valgus of the ankle.

plantar flexion, and valgus becomes apparent as the foot is dorsiflexed to neutral. With further dorsiflexion, midfoot break occurs. Aggressive realignment of the valgus foot may not be required in this group of young patients. Postoperative support with orthotics is then needed to control the hindfoot once the equinus is corrected surgically. This treatment does not work in older children, as the valgus becomes fixed with age.

Tendon lengthening of the peroneals as a treatment for pes valgus has been studied[309] but has not been found to be successful in obtaining and maintaining a plantigrade foot, with some feet undercorrected and others drifting into varus over time.[51] Tenotomy of the peroneals can not only lead to a varus deformity, but a dorsal bunion of the first metatarsophalangeal joint may also occur owing to the absence of plantar flexion of the first metatarsal head by the peroneus longus. Transfer of the peroneus brevis to the posterior tibialis has also been performed,[344] with resultant overcorrection and dorsal bunion formation.[174] We do not advocate these procedures.

In valgus deformity, bony surgery is the only predictable alternative for full and lasting correction. Surgical options are (1) the Grice extra-articular arthrodesis, (2) lateral column lengthening of the calcaneal neck, (3) calcaneal osteotomy, and (4) triple arthrodesis.

Grice Extra-Articular Arthrodesis. The Grice extra-articular arthrodesis was first described by Grice in 1952 as a means of correcting pes valgus in young polio patients between the ages of 4 and 12 years (Plate 24-3).[186] The procedure was soon used widely for correction of pes valgus in children with cerebral palsy. A structural graft, such as from the fibula or tricortical iliac crest, is propped into a shallow trough in the sinus tarsi laterally to support the plantar flexed talus and correct the valgus of the subtalar joint. The advantage of this operation is that it does not interfere with the growth of the tarsal bones, because it is not a formal arthrodesis. Growth disturbance results from arthrodesis, as the articular surfaces of the tarsals are growth centers. The Grice arthrodesis is often combined with lengthening of the peroneal tendons or the Achilles tendon. Grice reported satisfactory results in 79 percent of patients in whom this procedure was used for pes valgus associated with a variety of diagnoses (Fig. 24–34).[187]

Although a few series have reported excellent outcomes following a Grice arthrodesis,[138,249] most groups have found that the results of the procedure are unpredictable at best.[276,300,397,418] The graft is not inherently stable in the Grice procedure, and loss of correction due to dislodgment of the graft is well documented. Bleck, in his book on cerebral palsy, points out that the orientation of the graft must be vertical for the forces through it to be compression rather than rotation. If the graft is aligned obliquely, dislodgment and fracture of the graft are likely.[51] McCall and associates attributed their failures with the Grice procedure to graft resorption, graft dislodgment, failure of the graft to unite, and undercorrection of valgus at the time of surgery.[276] One-third of the feet operated on for valgus due to cerebral palsy had unsatisfactory results, in part because of uncorrected contracture of the Achilles tendon. Similarly, Moreland and Westin found a 31 percent incidence of nonunion following Grice arthrodesis in children with cerebral palsy.[300] Ross and Lyne noted poor results in 12 of 17 patients who underwent a Grice subtalar arthrodesis, with most failures due to persistent valgus or graft slippage.[397] And Scott and associates reported failure of the Grice arthrodesis in 6 of 10 patients with cerebral palsy who were followed to maturity. Reasons for failure included overcorrection into varus, ankle valgus, and graft nonunion.[418]

Because of these problems, several modifications of the original procedure were proposed. First, the site from which the bone graft is taken was changed from the fibula to the iliac crest, as symptomatic fibular nonunions and progressive ankle valgus were recognized.[216,492] Some groups began using bone plugs or dowels inserted across the sinus tarsi to stabilize the subtalar joint.[61,166,193,358,422] Without internal fixation, there still is a risk of graft fracture and loss of correction, because the bone dowel is placed rather obliquely to the axis of weightbearing.[51,192]

Dennyson and Fulford, among others, have advocated using internal fixation to help maintain the position of the subtalar joint in combination with cancellous iliac crest graft.[125] Local bone graft from the calcaneus has also been used.[222] Less loss of correction occurs with the addition of internal fixation, and problems with screw breakage are very rare. Cancellous bone grafting accelerates bony fusion. Again, coexisting contractures must be addressed to improve results.[158] Improved results have been seen with the addition of internal fixation, with up to 70 to 95 percent of feet having satisfactory short-term outcomes.[33,196,222] Alman and associates used K-wires rather than a screw to maintain subtalar joint position while the graft healed, with good results in 48 of 53 feet.[16] Bioabsorbable screws were used in an extra-articular subtalar arthrodesis with comparable results in one small series, but this technique must be considered investigational at present.[333]

Arthroereisis of the Subtalar Joint. Stabilization of the subtalar joint in the correct alignment without fusion of the joint has been studied by several authors. The joint is propped open laterally, the talus is reduced on the calcaneus, and stabilization is achieved by inserting of either a staple or a polyethylene spacer. Crawford and associates first popularized this technique, finding lasting improvement in the pes valgus in up to 85 percent of children treated before age 6 years.[99] Yet almost one-half of the patients treated with staple arthroereisis in another series required revision or subsequent arthrodesis, leading the authors to discontinue use of the procedure in favor of osteotomy.[410] Bleck observed a lucency around the staple on radiographs of patients who had undergone arthroereisis without fusion, and he did not recommend this technique.[51]

Vendantam and colleagues have published the results of arthroereisis using a polyethylene peg spacer in children with cerebral palsy. They found satisfactory results in 96 percent of their patients at 4.6 years of follow-up.[486] We have no experience with this technique.

Lateral Column (Calcaneal) Lengthening. Lateral column lengthening was first described by Evans,[152] and has enjoyed recent popularity after a recent series by Mosca.[302] Mosca performed the procedure to correct pes valgus in 31 feet, including 26 procedures done for valgus due to cerebral palsy and myelomeningocoele.[302] Correction is achieved by lengthening the calcaneus, and therefore the lateral column of the

foot, effecting tightening of the plantar fascia and reduction of the lapsed talonavicular joint. The procedure is summarized in Plate 24–4 and below.

The calcaneus is approached through an oblique incision laterally that follows the skin lines. The peroneal sheath is incised and the tendons are retracted. The superficial peroneal nerve is identified and protected. The extensor digitorum brevis is reflected from the lateral surface of the calcaneus and the sinus tarsi. A subperiosteal exposure of the distal calcaneus is achieved, and the calcaneocuboid joint is identified but its capsule is left intact. An osteotomy is created 1.5 cm proximal to the calcaneocuboid joint, in the area between the anterior and middle facets. The osteotomy is opened laterally, and a tricortical iliac crest graft is inserted into the osteotomy. Care is taken to prevent dorsal subluxation of the distal calcaneus, and pinning of the osteotomy and the calcaneocuboid joint is performed when needed. Reefing of the posterior tibialis and medial talonavicular capsule is done when laxity persists. Nearly always, the Achilles and peroneal tendons must be lengthened to attain a plantigrade foot. The foot is then immobilized in a nonweightbearing short-leg cast.

Published series have reported very good results in patients with flatfoot deformity and valgus of different origins, including cerebral palsy, but also myelomeningocoele and idiopathic pes planovalgus (Fig. 24–35).[302,355] The preoperative rigidity of the foot did not correlate with the postoperative result. Several patients continued to use orthoses after surgery, but pain relief and resolution of the talar head callus or blisters were universal. Complications consisted of graft dislodgment and dorsal subluxation of the calcaneocuboid joint. The only contraindications to the procedure are advanced osteoarthrosis and the presence of other bony deformities of the foot.[302]

Calcaneal Osteotomy. Another surgical option for the child with pes valgus due to cerebral palsy is the calcaneal osteotomy described by Dwyer.[141] An osteotomy is performed obliquely from the sinus tarsi to the posterior margin of the calcaneus. Either a medial wedge can be resected or the lateral side can be propped open as an opening wedge and bone grafted.[428,429] Additionally, a sliding osteotomy can be performed in which the distal inferior fragment of the calcaneus is moved medially to reestablish the heel directly beneath the axis of weightbearing.[243,369] The advantage to these calcaneal osteotomies is that they preserve joint motion of the subtalar joint.[377] Results with these osteotomies have been very good, with Koman and associates publishing 94 percent excellent results following medial displacement osteotomy.[240] A contraindication to surgery is severe rigid valgus deformity, which is best treated with triple arthrodesis. A bony abnormality or malalignment in the midfoot or forefoot will not be amenable to treatment by a single osteotomy through the calcaneus, since realigning the calcaneus and hindfoot could exacerbate deformity more distally in the foot. In these feet, triple arthrodesis or more complex osteotomies such as the calcaneal-cuboid-cuneiform osteotomies described by Rathjen and Mubarak will correct deformity at more than one level in the foot.[369]

Triple Arthrodesis. Triple arthrodesis is the treatment of choice for rigid symptomatic pes valgus in the adolescent with cerebral palsy (Fig. 24–36). By resecting the subtalar, calcaneocuboid, and talonavicular joints, the growth of these bones is disturbed, leading to a small, shortened foot in younger patients. In the adolescent, however, a well-corrected triple arthrodesis can yield a stable plantigrade foot for future ambulation. The indications for triple arthrodesis are pain, skin ulcerations over the talar head, and deformity interfering with ambulation in a child with a deformity not amenable to osteotomy. Valgus alone without disabling symptoms does not merit a triple arthrodesis.

The technique used is identical to that used in the nonneuromuscular population.[212,248] Wedges of bone are resected with the articular surfaces, and internal fixation is used when the bones are sufficiently strong. Screws or staples can be used. A short-leg nonweightbearing cast is used, followed by a walking cast, and then an orthosis until the fusion is mature.

Satisfactory outcomes are found when the deformity is well corrected.[219] Postoperative malalignment usually results from undercorrection of the valgus deformity at the time of surgery.[420,471] It is technically more challenging to perform a perfect triple arthrodesis for a valgus foot than for a varus foot, and it is particularly difficult to achieve fusion at the talonavicular joint.[219] When visualization of the talonavicular joint is compromised, it is advised to make a second medial incision to complete the joint resection.

Patients are generally satisfied with the results of triple arthrodesis, and rarely is the talonavicular joint pseudarthrosis sufficiently symptomatic to interfere with function or to necessitate further surgery. Degenerative changes have been documented in the ankle joint at an average of 18 years following triple arthrodesis in 43 percent of the pediatric population, but functional limitations in this group of patients are unusual.[7,437,471]

Triple arthrodesis has also been used at our institution for the treatment of severe fixed deformity in nonambulatory patients who cannot wear shoes. The bony wedges resected in such cases are quite large, but patients and parents have been pleased with the improvement in the position of the feet and the ability to wear shoes in public.

ANKLE VALGUS. Patients with neuromuscular disease often develop valgus alignment of the ankle, which can contribute to the overall valgus deformity of the foot. Before surgical correction of pes valgus is undertaken, radiographs of the ankle with the patient standing should be obtained. Valgus of the ankle is present when the physis of the distal fibula is located proximal to the distal tibial articular surface.

Surgical correction of ankle valgus is either by epiphysiodesis or osteotomy. Hemiepiphysiodesis of the distal medial tibia provides gradual correction by tethering the medial malleolus and media tibia while allowing for growth of the fibula and lateral distal tibia. Three techniques have been used. When a permanent hemiepiphysiodesis is desired, such as in a child approaching the end of growth, an open epiphysiodesis of the medial malleolus is performed. When the surgeon anticipates full correction prior to the cessation of growth, a more temporary epiphysiodesis effect may be desired. In such cases, hemiepiphysiodesis using staples or a vertical medial malleolar screw will tether growth laterally, but allows for resumption of growth upon removal of the implants.[116,451] Staples in thin children with cerebral palsy

Text continued on page 1158

Extra-articular Arthrodesis of the Subtalar Joint (Grice Procedure)

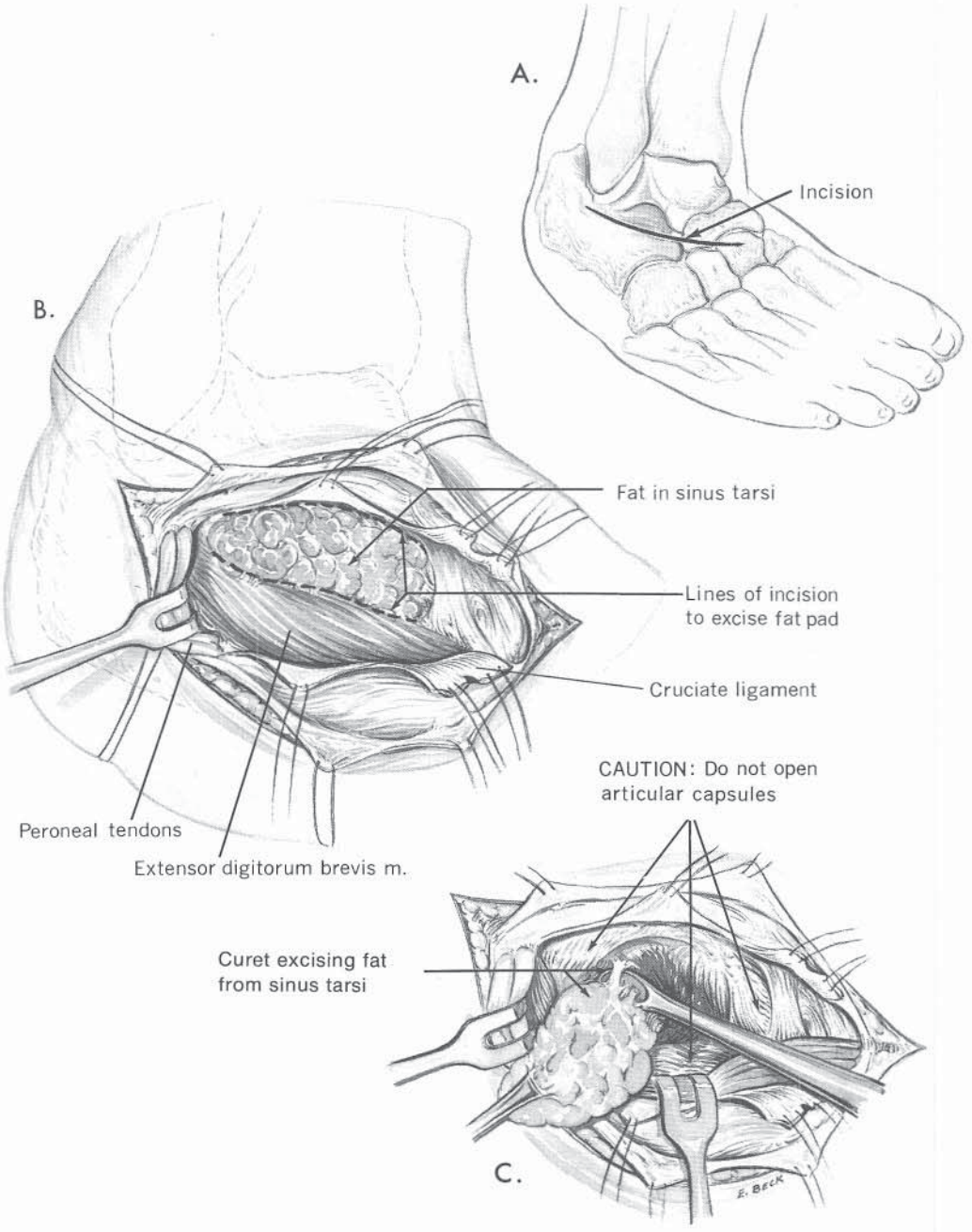
A, A 2½-inch long, slightly curved incision is made over the subtalar joint, centered over the sinus tarsi.

B, The incision is carried down to the sinus tarsi. The capsules of the posterior and anterior subtalar articulations are identified and left intact. The operation is extra-articular. If the capsule is inadvertently opened, it should be closed with interrupted sutures.

The periosteum on the talus corresponding to the lateral margin of the roof of the sinus tarsi is divided and reflected proximally. The fibrofatty tissue in the sinus tarsi and the tendinous origin of the short toe extensors from the calcaneus are elevated and reflected distally in one mass.

C, The remaining fatty and ligamentous tissue from the sinus tarsi is thoroughly removed with a sharp scalpel and curet.

PLATE 24–3. Extra-articular Arthrodesis of the Subtalar Joint (Grice Procedure)



Extra-articular Arthrodesis of the Subtalar Joint (Grice Procedure) *Continued*

D, Next, the foot is manipulated into equinus position and inversion, rotating the calcaneus into its normal position beneath the talus and correcting the valgus deformity. Broad straight osteotomes of various sizes ($\frac{3}{4}$ to $1\frac{1}{4}$ inches or more) are inserted into the sinus tarsi, blocking the subtalar joint and determining the length and optimum position of the bone graft and the stability that it will provide. The long axis of the graft should be parallel to the long axis of the leg when the ankle is dorsiflexed into neutral position.

E, The optimum site of the bone graft bed is marked with the broad osteotome. A thin layer of cortical bone ($\frac{1}{8}$ to $\frac{3}{16}$ inch) is removed with a dental osteotome from the inferior surface of the talus (the roof of the sinus tarsi) and the superior surface of the calcaneus (the floor of the sinus tarsi) at the site marked for the bone graft. It is best to preserve the most lateral cortical margin of the graft bed to support the bone block and to prevent it from sinking into soft cancellous bone.

F, A bone graft of appropriate size can be taken from the fibula or iliac crest. The corners of the base of the graft are removed with a rongeur so that the graft is trapezoidal in shape and can be countersunk into cancellous bone, preventing lateral displacement after operation.

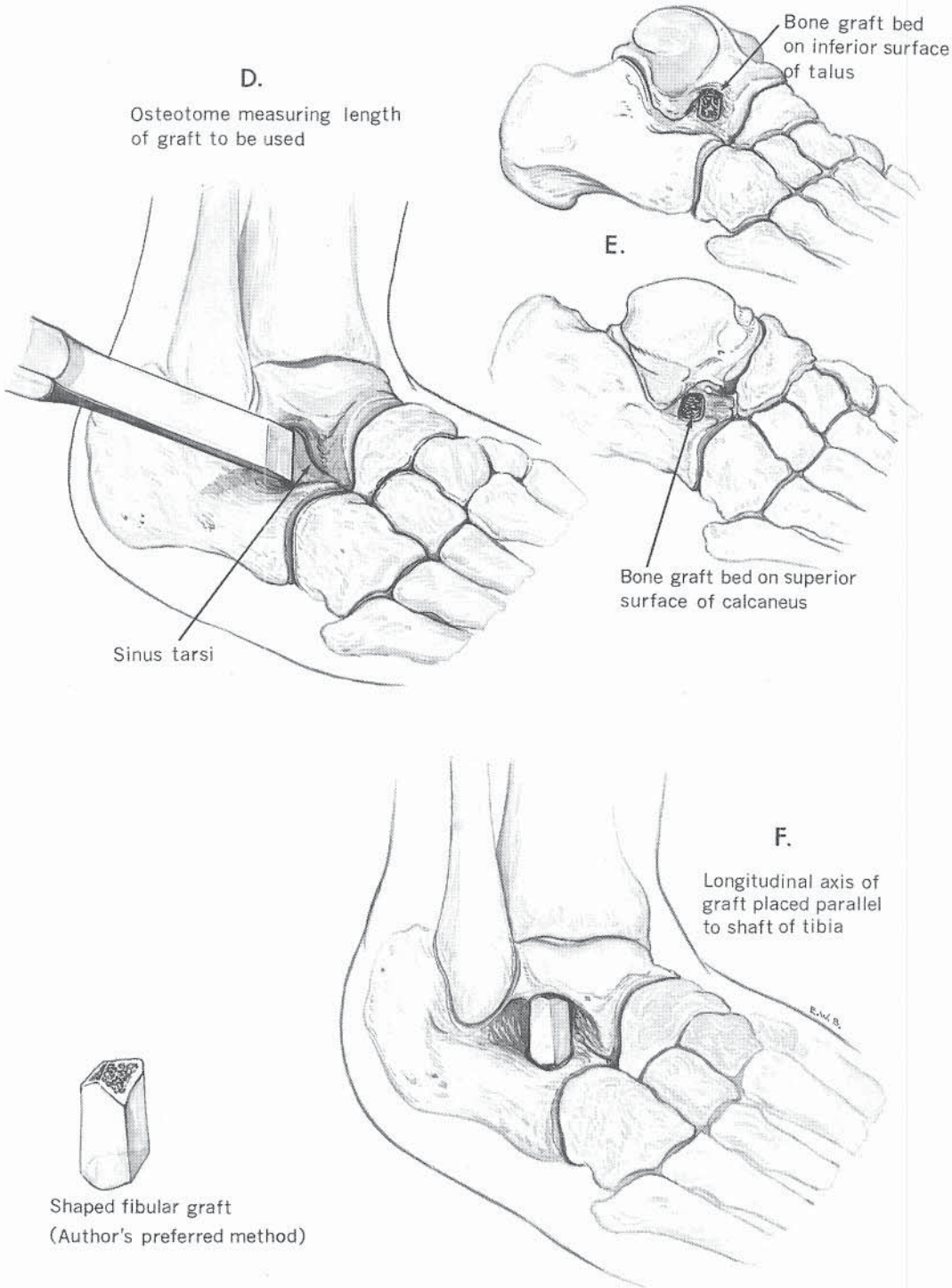
The bone graft is placed in the prepared graft bed in the sinus tarsi by holding the foot in varus position. An impacter may be used to fix the cortices of the graft in place. The longitudinal axis of the graft should be parallel to the shaft of the tibia with the ankle in neutral position.

With the foot held in the desired position, the distal soft tissue pedicle of fibrofatty tissue of the sinus tarsi, the calcaneal periosteum, and the tendinous origin of the short toe extensors are sutured to the reflected periosteum from the talus. The subcutaneous tissue and skin are closed with interrupted sutures, and a below-knee cast is applied.

POSTOPERATIVE CARE

The cast is removed 8 to 10 weeks after operation. If radiographs show solid healing of the graft, gradual weightbearing is allowed with the protection of crutches. Active and passive exercises are performed to strengthen the muscles and to increase the range of motion of the ankle and the knee.

PLATE 24–3. Extra-articular Arthrodesis of the Subtalar Joint (Grice Procedure)



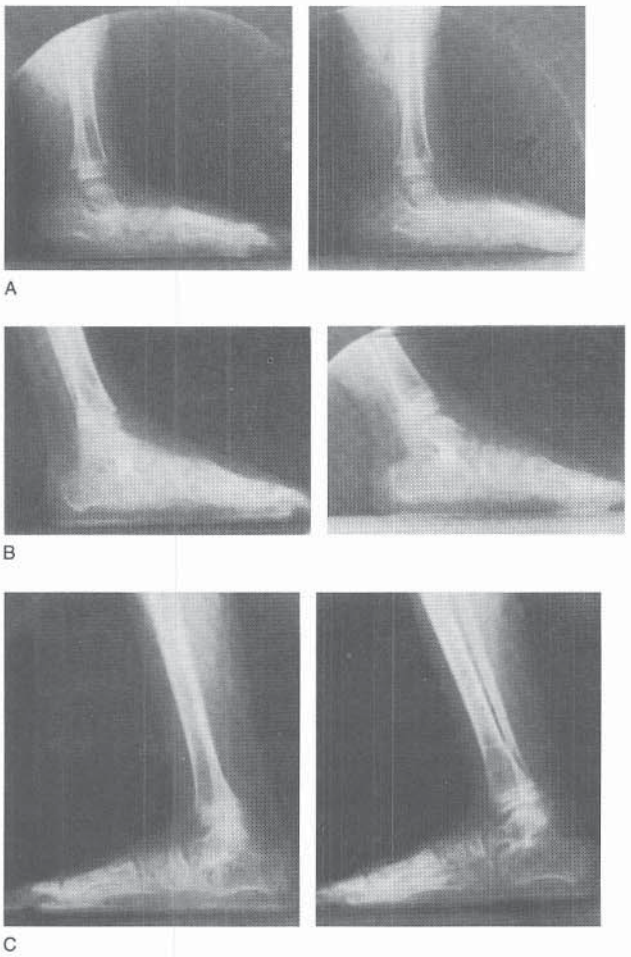


FIGURE 24–34 **A,** Weightbearing lateral radiograph of a patient with pes equinovalgus. **B,** Radiograph obtained 8 months after Achilles tendon lengthening and Grice extra-articular arthrodesis. **C,** Lateral radiograph obtained 3 years 6 months postoperatively.

may be quite prominent and cause rubbing on orthoses, resulting in skin problems or pain, so we have used the screw technique of late. The procedure can be done percutaneously, and immobilization is unnecessary. Although we have seen growth out of valgus with the medial malleolar screw, our experience is too new to comment definitively on growth following screw removal. Stevens and Belle[451] and Davids and colleagues[116] have published their results. Correction was obtained with minimal morbidity in a total of 79 feet, and further growth was obtained upon hardware removal, so much so that a few patients required replacement of the screw for recurrent valgus.[116]

When immediate correction of the valgus is desired, distal tibial osteotomy is useful. Internal fixation allows precise realignment. Usually a closing wedge osteotomy of the distal tibia, combined with distal fibular osteotomy, is performed.

HALLUX VALGUS. Hallux valgus in patients with cerebral palsy develops in response to an equinovalgus deformity of the hindfoot. The peroneus longus is spastic, leading to progressive eversion and abduction of the foot. This leads to lateralization of the origin of the adductor hallucis muscle, which results in increasing pull of the proximal phalanx of the great toe into adduction. When combined with external tibial torsion, the toe is pushed laterally as weight is borne by the everted forefoot. The big toe comes to lie beneath the second toe. The head of the first metatarsal becomes uncovered as the toe deviates laterally, and a painful bunion develops.[49] The patients complain of discomfort and swelling over the prominent head of the first metatarsal and difficulty in shoe wear.

Before embarking on the surgical correction of hallux valgus in a child or adolescent with cerebral palsy, the surgeon needs to carefully examine the child for concomitant malalignment in the tibia and foot. If the bunion is corrected but the external tibial rotation and crouch or pes valgus is not corrected, the hallux valgus deformity is likely to remain symptomatic. When the hallux valgus is mild, surgical treatment of the pes valgus will halt the progression of the toe deformity.

When the bunion is symptomatic, Goldner[174] and Bleck[49] describe soft tissue realignment, including release of the adductor hallucis tendon and lateral capsulotomy of the first metatarsophalangeal joint, combined with first metatarsal osteotomy and proximal phalangeal osteotomy for the treatment of hallux valgus in patients with cerebral palsy.[49,174] However, in studies reported from the Newington Children's Hospital and the A. I. Dupont Institute, first metatarsophalangeal fusion using the technique of McKeever led to better results with less recurrence than soft tissue realignment combined with either proximal or distal first metatarsal osteotomies.[221,378] The preferred position for fusion is 15 to 25 degrees of dorsiflexion and slight valgus.[158,283]

We prefer the first metatarsophalangeal arthrodesis for the surgical treatment of hallux valgus in patients with cerebral palsy (Fig. 24–37). We use internal fixation with screws whenever possible. In the rare cases in which pseudarthrosis occurs, revision surgery with additional internal fixation has been successful.

DORSAL BUNION. Dorsal bunion is a rare deformity in which the first metatarsal head is elevated but the great toe is plantar flexed, leading to a dorsal prominence of the metatarsal head (Fig. 24–38). The etiology is usually iatrogenic, following surgical procedures meant to balance the foot. It is argued whether the primary deforming force is an overpowering by the tibialis anterior of a weak peroneus longus or overpowering by the flexor hallucis of a weak extensor hallucis and gastrocsoleus. Symptoms are related to pain over the prominence with shoe wear. Surgery entails rebalancing the pull on the great toe, either by transfer of the flexor tendon to the extensor, or by flexor tendon tenotomy with or without anterior tibialis transfer, or by transfer of the flexor hallucis brevis to the metatarsal neck, or by these procedures in combination with a closing wedge plantar flexion osteotomy of the first metatarsal.[49,174,282]

Knee Surgery. Surgery at the knee in patients with cerebral palsy consists of hamstring lengthenings and rectus femoris transfer. The hamstrings are nearly always affected in patients with cerebral palsy. Tight hamstrings lead to crouch gait.[460] Findings during gait analysis include greater than normal knee flexion during midstance and inability to extend the knee fully at the end of swing phase and initial contact (Fig. 24–39). Step length then decreases as the knee loses excursion. Increasing demand is placed on the quadriceps to resist the progressive crouch, and energy expenditure

during gait rises.[343] The quadriceps and patellar tendon stretch, and patella alta may result.

It is important to note that the hamstrings cross two joints, the hip and the knee. At the hip the hamstrings serve as hip extensors, while at the knee they serve as knee flexors. The medial hamstrings also produce some dynamic internal rotation of the hip during gait.

Clinically, hamstring spasticity can be measured via the popliteal angle. The patient is positioned supine on an examining table and the hip is flexed to 90 degrees. The ipsilateral flexed knee is then extended, and the angle between the vertical and where the tibia may be extended to is the popliteal angle (Fig. 24–40). There is variability in normal popliteal angles, with a mean value of 26 degrees in normal children age 4 years and older.[231] Values greater than 50 degrees in this age range are considered abnormal. A decrease in the angle of straight-leg raising is also seen in the presence of tight hamstrings (Fig. 24–41).

In severe hamstring contracture, a fixed knee flexion contracture develops (Fig. 24–42). It is important to assess for a fixed contracture, as the presence of a contracture may lead to disappointing results following hamstring lengthening. When the contracture is most severe, the patient becomes unable to flex the hips and develops a poor sitting posture, with lumbar kyphosis and a slumped position (Fig. 24–43).

As discussed earlier in the section on equinus, the examining physician must carefully assess other joints for spasticity and contracture. A bent knee gait may be a compensation for equinus and toe-walking if the popliteal angle is normal. The hip must also be examined, as correction of hamstring contractures without addressing concomitant hip flexion contractures leads to increased hip flexion and forward lean during gait.

Mild tightness in the hamstrings may respond to orthotic management, usually with ground reaction AFOs. The posterior push on the knee from the brace in stance phase can improve mild crouch without fixed contracture. KAFOs are rarely prescribed in cerebral palsy.

One procedure that has been historically used for the correction of spasticity in the hamstrings and crouch knee is that described by Eggers, in which the distal medial and lateral hamstring tendons are transferred to the posterior femoral condyles (Fig. 24–44).[147] Although crouch at the knee was improved, genu recurvatum was a frequent complication. Therefore, this procedure is rarely performed currently.[373]

Proximal hamstring release off the ischial origin has also been studied in patients with crouch gait.[423] Sharps and colleagues did not find deterioration in gait or an increase in the average lumbar lordosis in 32 patients studied retrospectively out of a series of 78 patients who had undergone proximal hamstring release. They did find hyperlordosis in 8 of 31 patients for whom radiographs were available, including one symptomatic patient with 75 degrees of lordosis and back pain.[424] Drummond and colleagues performed proximal hamstring releases in 30 children, and found that anterior pelvic tilt and hyperlordosis of the lumbar spine occurred frequently. Those patients at greatest risk had hip flexion contractures of 25 degrees or more. For this reason, they no longer recommend proximal release in patients who are able to walk.[135] Proximal hamstring release in the nonambulatory patient will be discussed further in the section on hip surgery.

Surgical lengthening of the distal hamstrings is now the preferred surgical treatment for crouch knee gait and is often performed in combination with other soft tissue procedures. The technique of hamstring lengthening varies among surgeons. We prefer to do an intramuscular aponeurotic lengthening of the semimembranosus, a Z-lengthening of the semitendinosus, and either a tenotomy or a Z-lengthening of the gracilis at a level just proximal to the knee. When lateral hamstrings are included in the procedure, an intramuscular aponeurotic lengthening of the biceps femoris is done (Plate 24–5). Usually two cuts are needed in the fascia of the semimembranosus and biceps femoris for adequate lengthening. The popliteal angle is rechecked, and adequate lengthening has been accomplished when the angle is reduced to around 20 degrees.

In the past, we always used a long-leg cast in the postoperative period. Currently, if the knee can be fully extended with ease following surgery, a knee immobilizer provides sufficient immobilization for 3 or 4 weeks. If there is residual contracture, a long-leg cast is applied. Early weightbearing and ambulation is encouraged in physical therapy, as the child who becomes nonambulatory in the immediate postoperative period loses strength and has more difficulty when immobilization is discontinued.

Postoperatively, improvements in knee extension in stance phase are expected. Greatest improvements in the knee flexion contracture are seen within 1 year following surgery. As a rule, the greater the contracture, the greater is the degree of correction.[112] A significant number of children improve at least one level in their ability to walk following hamstring lengthening, with up to 39 percent of preambulatory patients becoming able to walk at least around the house.[112,126] Reimers measured quadriceps strength in patients following distal lengthening of the hamstrings. He found that quadriceps strength was significantly reduced initially but returned to preoperative values by 6 months, and then improved by 1 year following surgery.[375] Damiano and colleagues found similar results, with hamstring strength also decreasing following surgery but improving by 9 months, and quadriceps strength increasing by 9 months as well.[109]

Often, anterior pelvic tilt increases following hamstring lengthening. This is due to weakening of the hamstrings. Since the hamstrings are also hip extensors, weakness leads to more relative hip flexion due to muscle imbalance, and forward tilt of the pelvis and trunk. De Luca and colleagues found that if just the medial hamstrings were lengthened, anterior pelvic tilt was not seen, but if the medial and lateral hamstrings were lengthened without psoas surgery, the pelvis did tip anteriorly.[121] Muscle length modeling has been done for the hamstrings and psoas in crouch gait and has shown that the hamstrings are usually not particularly short, since they also cross the hip, while the psoas is often shortened.[123] If there is a hip flexion contracture, it must also be surgically lengthened to minimize the postoperative tendency toward more hip flexion.[211]

It is common also to see a decrease in the flexion of the knee in swing phase following hamstring lengthening.[474] Normally the knee should flex at least 60 degrees in swing

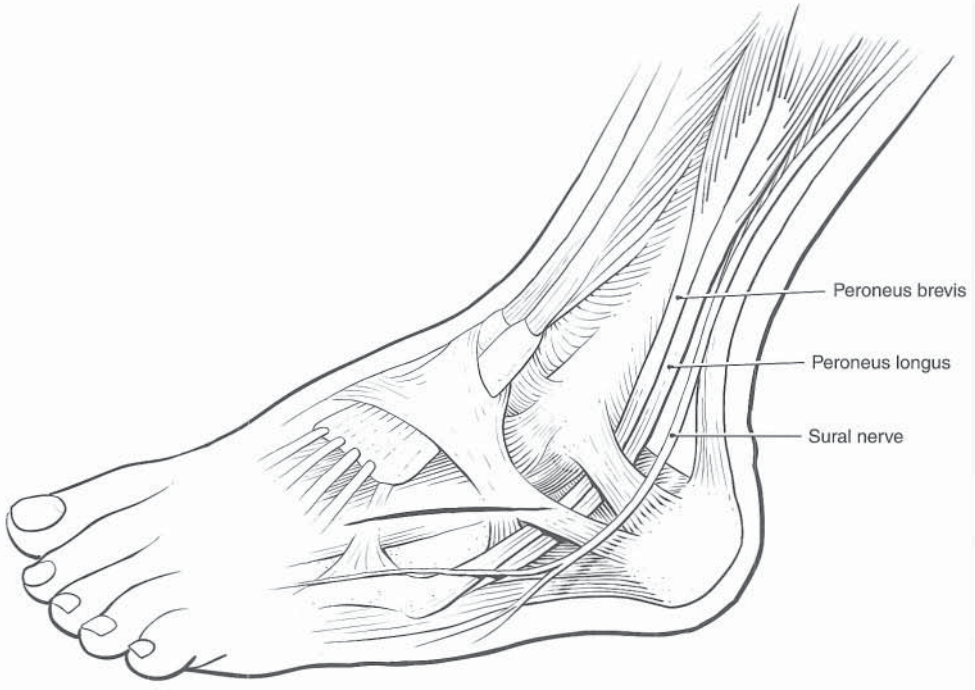
Text continued on page 1166

Lateral Column Lengthening

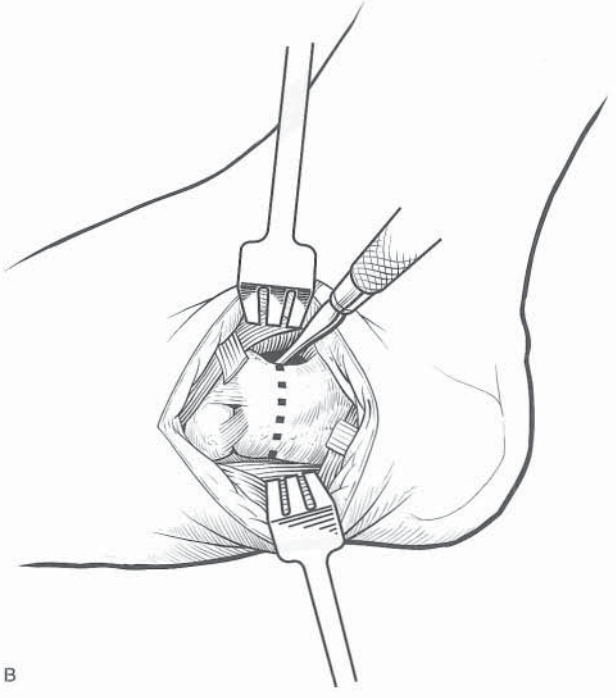
A, The calcaneus is approached laterally through a longitudinal incision.

B, The peroneal tendons are retracted plantarward and the neck of the calcaneus is exposed. The calcaneocuboid joint is identified but left undisturbed.

PLATE 24–4. Lateral Column Lengthening



A



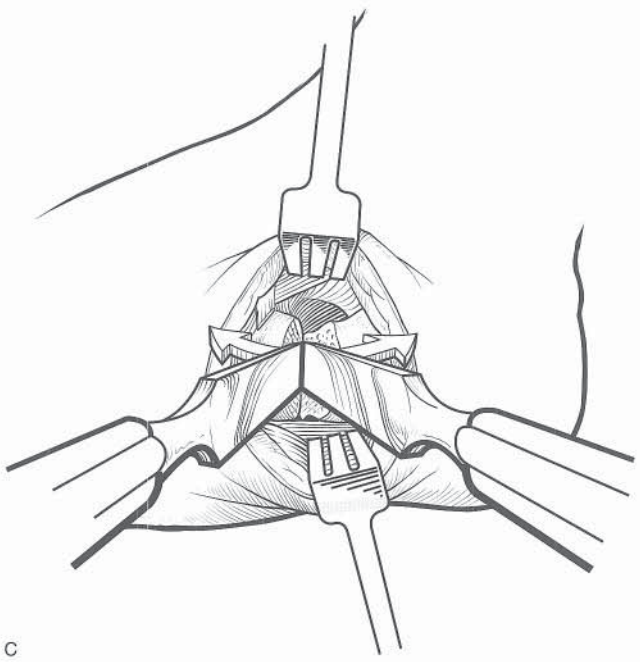
B

Lateral Column Lengthening *Continued*

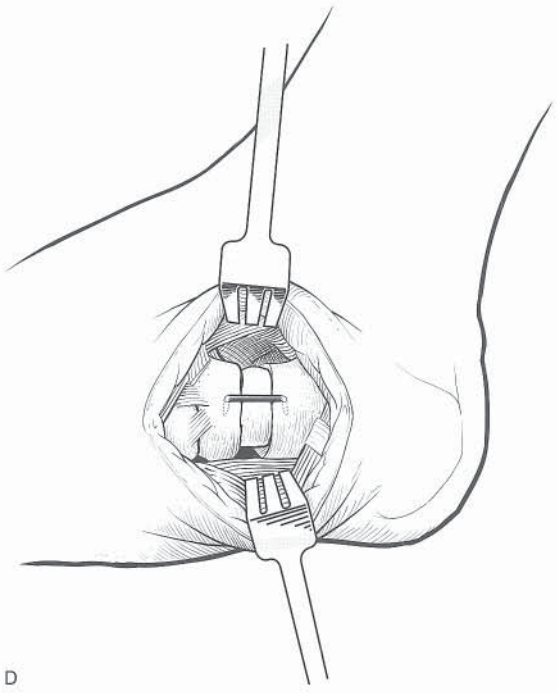
C, A vertical osteotomy is made in the neck of the calcaneus and hinged open laterally with osteotomes. A laminar spreader should not be used, as it will crush the bony fragments.

D, A tricortical wedge of iliac crest is placed in the osteotomy. The osteotomy may be stabilized with K-wires or a staple.

PLATE 24–4. Lateral Column Lengthening



C



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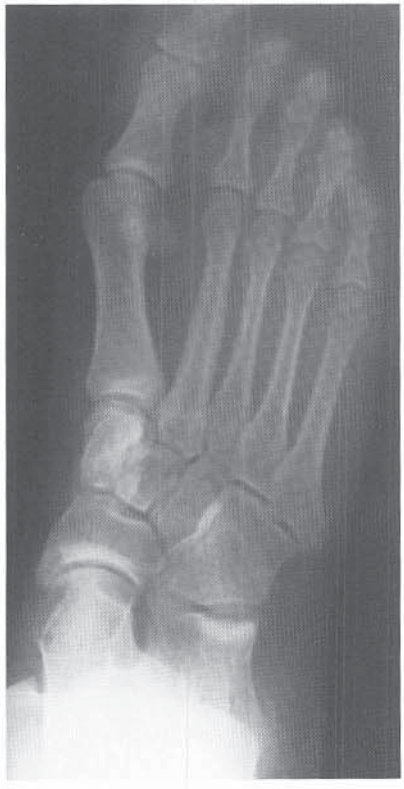
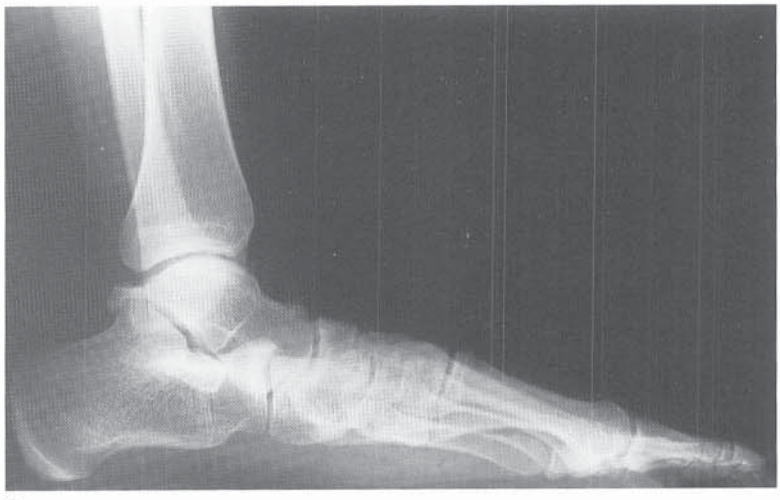
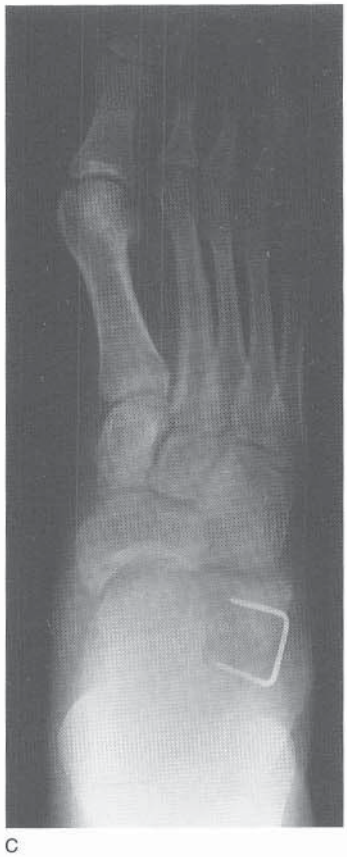


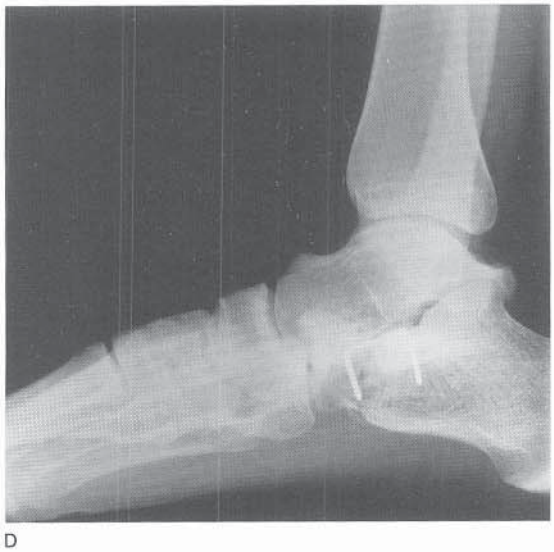
A



B

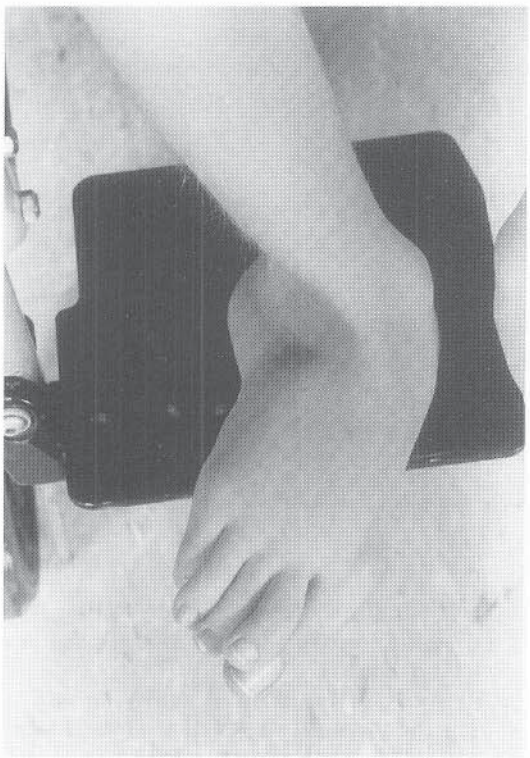


C

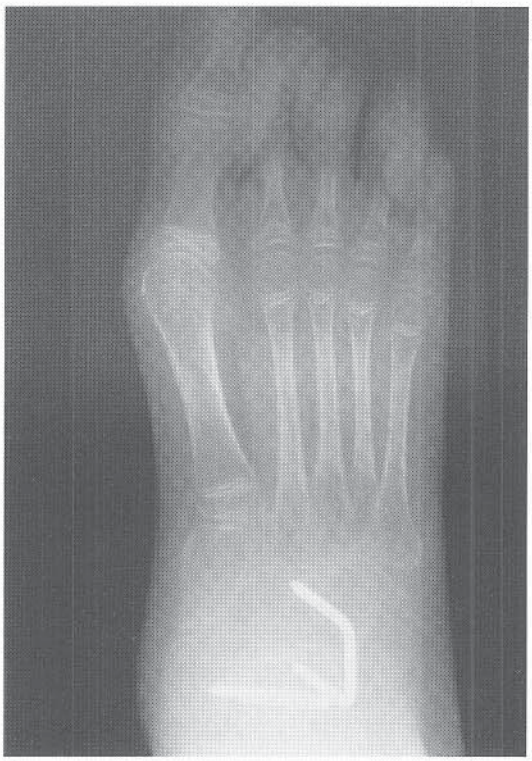


D

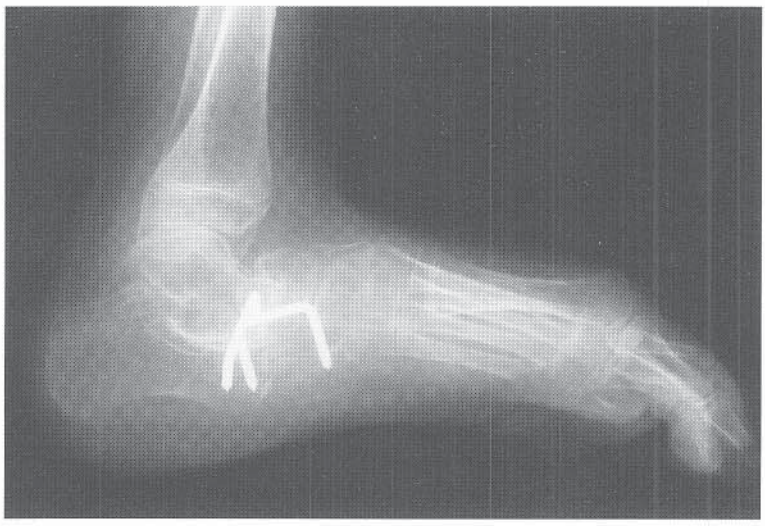
FIGURE 24–35 **A** and **B,** Preoperative AP and lateral radiograph of the foot of a 17-year-old girl with right hemiplegia and painful pes valgus. **C** and **D,** AP and lateral radiographs after lateral column lengthening.



A

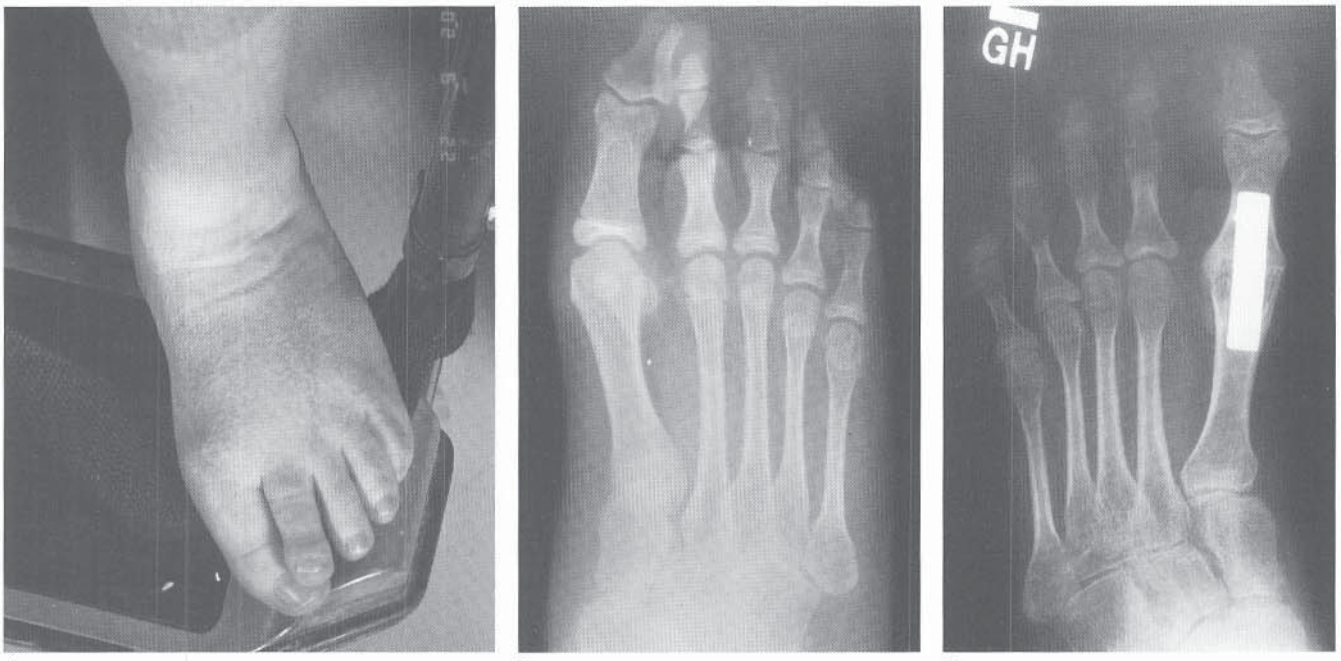


B



C

FIGURE 24–36 **A** to **C**, Triple arthrodesis for severe fixed painful valgus deformity in nonambulatory girl with cerebral palsy.



A B C

FIGURE 24–37 A to C, Hallux valgus in a 14-year-old girl with spastic diplegia treated with metatarsophalangeal fusion.

phase, and this flexion occurs quite early in swing phase.[160] Gage, Sutherland, and Perry all describe the inability of the knee to flex in swing phase resulting from spasticity of the rectus femoris muscle which becomes apparent following hamstring lengthening.[160,164,462] As a muscle that crosses two joints, the rectus acts to flex the hip at initial swing, and to extend the knee.[341] Preoperative EMG will often show inappropriate electrical activity in the rectus femoris during midswing. Gait analysis in affected patients shows a decrease in the amount of swing phase knee flexion and a delay in when the peak swing phase knee flexion occurs (Fig. 24–45).[474] When severe, this leads to problems with clearing the foot in swing phase, resulting in tripping and dragging the toe. Patients may complain of difficulty climbing stairs or surmounting a street curb, or in moving from a standing position to a seated position and vice versa, known as transitional movements.

Spasticity in the rectus femoris can also be tested for during the physical examination. The Duncan Ely test is performed by positioning the patient prone and then flexing the knee to 90 degrees. If there is spasticity in the rectus femoris, the ipsilateral buttock will rise from the table as a result of hip flexion caused by the rectus (Fig. 24–46). Unfortunately, this test is not specific to the rectus, as a patient with a hip flexion contracture due to tightness of the iliopsoas will also have a positive Ely test. Another clinical measure of rectus spasticity is the rectus grab. With the

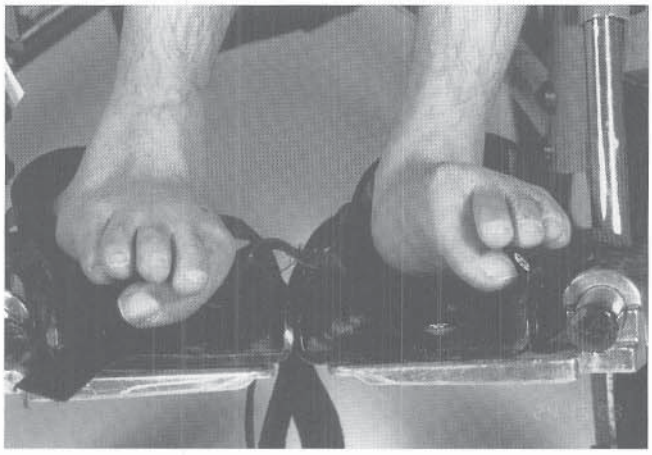


FIGURE 24–38 Painful bilateral dorsal bunion in a child with spastic quadriparesis.

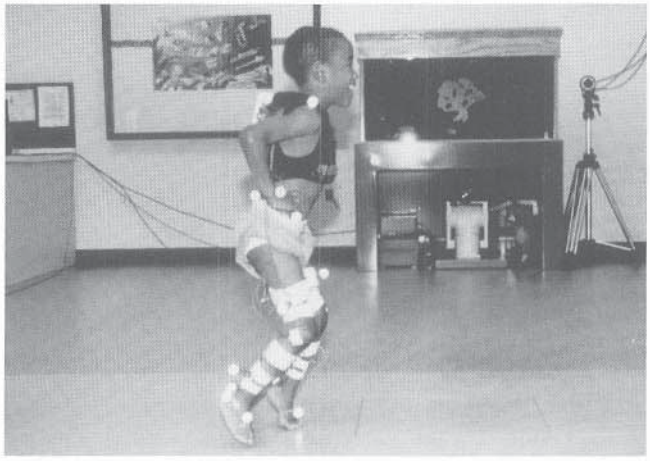


FIGURE 24–39 Young boy with crouch gait due to cerebral palsy. The knees are flexed due to hamstring spasticity, the hips are flexed, and the ankles are in equinus.

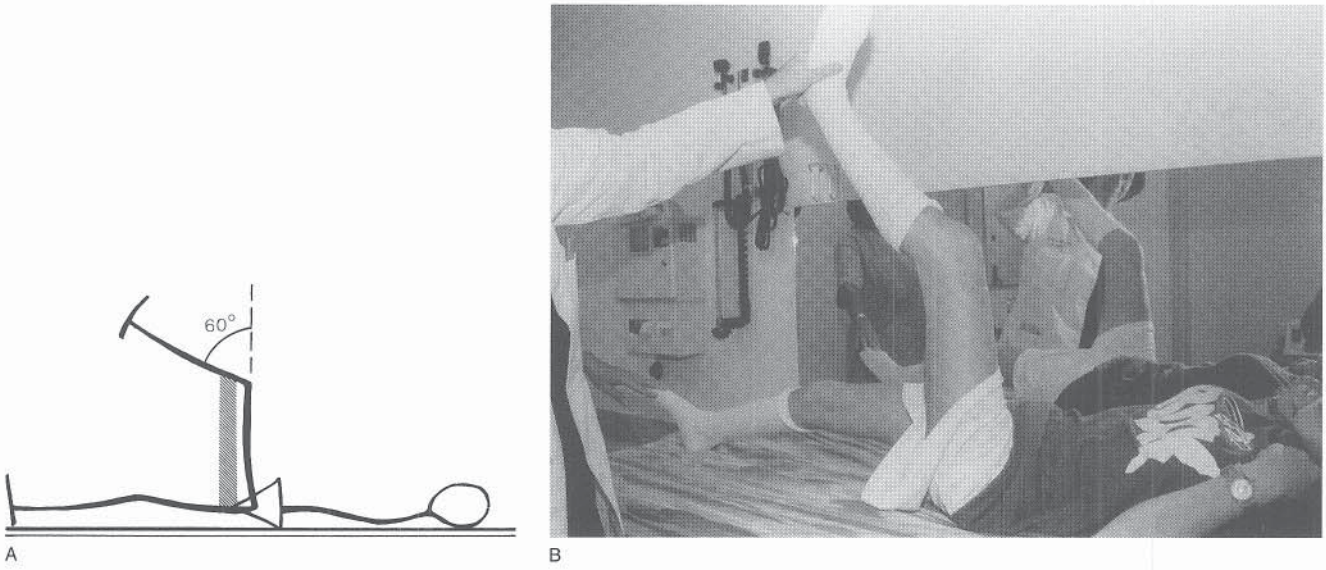


FIGURE 24–40 Holt's method of determining hamstring contracture. **A,** With the contralateral hip in extension, the tested limb is flexed to 90 degrees at the hip and the knee is extended passively. The angle between the anterior aspect of the leg and the axis of the thigh determines the degree of hamstring contracture. Bleck measures the angle on the popliteal surface between the leg and thigh. **B,** The popliteal angle of the left leg measures 40 degrees.

patient supine on the examining table, the knee is rapidly flexed. If resistance is felt, the rectus is spastic.

Not all patients with overactivity of the rectus femoris during swing phase will develop symptomatic stiff knees following isolated hamstring lengthenings. Although Damron and colleagues found that 71 percent of patients lost some knee flexion in swing phase following hamstring lengthening, only 13 percent of the ambulatory patients required rectus transfer for correction of stiff knee gait.[113] Dhawlikar and colleagues described recurvatum following distal hamstring lengthening, and the need for subsequent rectus femoris transfers in 17 percent of their patients.[126] In most studies up to 20 percent of patients develop recurvatum following hamstring lengthenings.[112,215]

The surgical treatment for stiff knee gait and inability to flex the knee in swing phase is a rectus femoris transfer.[164] This procedure is often performed simultaneously with hamstring lengthenings and other soft tissue procedures. The principle behind rectus transfer is to preserve the role of the rectus femoris as a hip flexor but to move the rectus distally, posterior to the axis of the knee, to eliminate its role as an inappropriate knee extensor during swing phase. Release of the proximal rectus femoris was studied but was found to increase swing phase knee flexion less than distal transfer of the rectus tendon.[463] Release of the rectus from the patella with mobilization of the muscle was also determined to be ineffective in treating loss of knee flexion in swing.[83,329] Neither of these procedures physically moves the distal rectus posterior to the knee joint, and this may be the

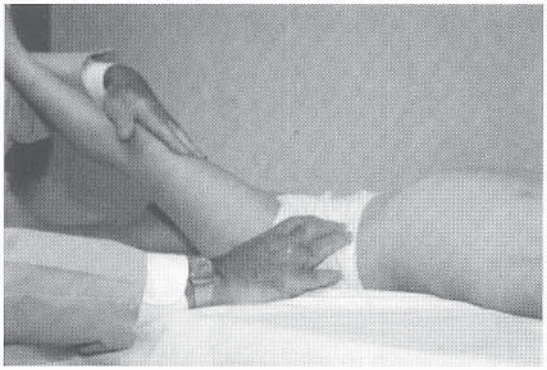


FIGURE 24–41 Method of determining hamstring tautness by straight-leg raising. The knee should be in complete extension and the pelvis should be stabilized. The angle between the lower limb and the examination table is measured.

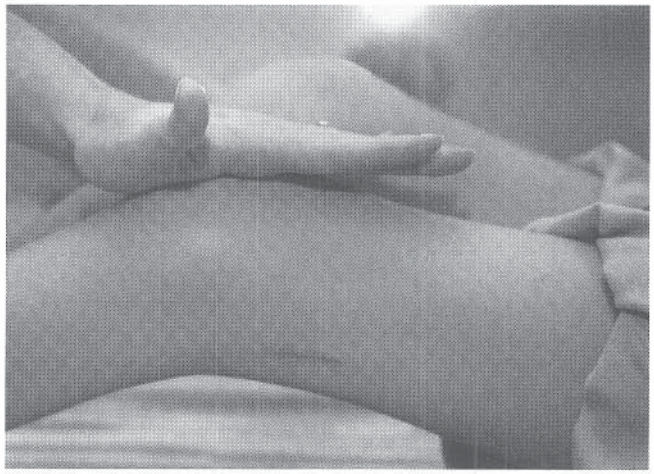


FIGURE 24–42 Fixed knee flexion contracture in a teenage boy with spastic quadriplegia. The contractures did not resolve following hamstring lengthening.

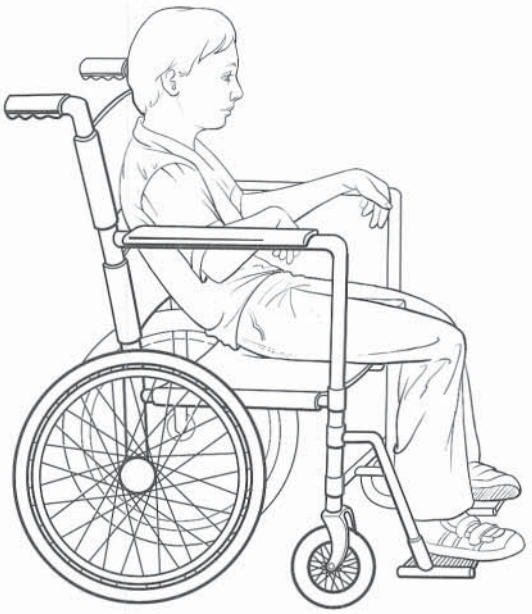


FIGURE 24–43 Hamstring tightness can lead to inability to flex the hips sufficiently to sit. The patient then thrusts forward in the chair and sits with lumbar kyphosis.

Postoperative care consists of either a long-leg cast or knee immobilizer, and early weightbearing and ambulation is again encouraged, as in hamstring lengthenings.

There has been abundant research published investigating rectus femoris transfer outcome. First, the preferred site for transfer was studied by Ounpuu, Gage, and others. While it was hypothesized that the rotation of the hip would become more external if the tendon was transferred medially and more internal if the tendon was transferred laterally, they found that rotation of the femur did not change, regardless of where the tendon was transferred.[328] They concluded that the site for tendon transfer could be determined based on the surgeon's preference and the existence of wounds from other concomitant surgeries such as simultaneous hamstring lengthenings. Miller transfers the rectus to the sartorius, while Gage prefers to transfer to the gracilis.[87,289]

Much has been written about the role of EMG in determining whether stiff knee gait will occur following hamstring surgery and in predicting the outcome of rectus femoris transfer. The results are conflicting. Chambers and colleagues found that preoperative EMG of the rectus femoris and vastus lateralis was not predictive of the amount of peak swing phase knee flexion following rectus release or transfer.[83] Miller and colleagues studied three groups of patients undergoing rectus transfer: those with phasic but inappropriate rectus EMG active in swing phase, those with

reason why they do not work as well as a rectus transfer. Riewald and Delp measured knee moments following rectus transfer and did not see that the rectus generated a knee flexor moment following surgery.[381] Regardless, rectus transfer has been found to increase swing phase knee flexion by an average of 16 degrees.[164] When rectus transfer is combined with a hamstring lengthening procedure, dynamic range of motion and crouch improve without a loss of swing phase knee flexion.[195,380]

The surgical technique for rectus transfer is as follows (Plate 24–6).[160] An incision is made superior to the proximal pole of the patella. Many incisions have been described, but I prefer to use a short transverse incision two to three fingerbreadths above the patella.[334] Through this cosmetic incision, the rectus femoris is dissected off the underlying vastus intermedius. Distally the two muscles and their tendons are quite adherent, so it is easier to start the dissection more proximally, where the tissue plane can be identified. The vastus lateralis and medialis also converge distally at the patellar insertion of the quadriceps tendon. Care must be taken to preserve these two muscles as well. Once the rectus femoris is dissected from the other parts of the quadriceps, the tendon is divided transversely just proximal to the superior pole of the patella, again taking great care to leave the tendon of the rest of the quadriceps undisturbed. A sturdy stitch is woven into the tendon of the rectus femoris, and a subcutaneous tunnel is made to the site of transfer. The tendon is then passed medially, usually through the posterior wound used for concomitant hamstring lengthening, and the rectus is sewn into either the stump of the gracilis tendon, the muscle of sartorius, or the lengthened semitendinosus. The remainder of the quadriceps tendon is then repaired by suturing the vastus lateralis to the medialis over the intermedius.

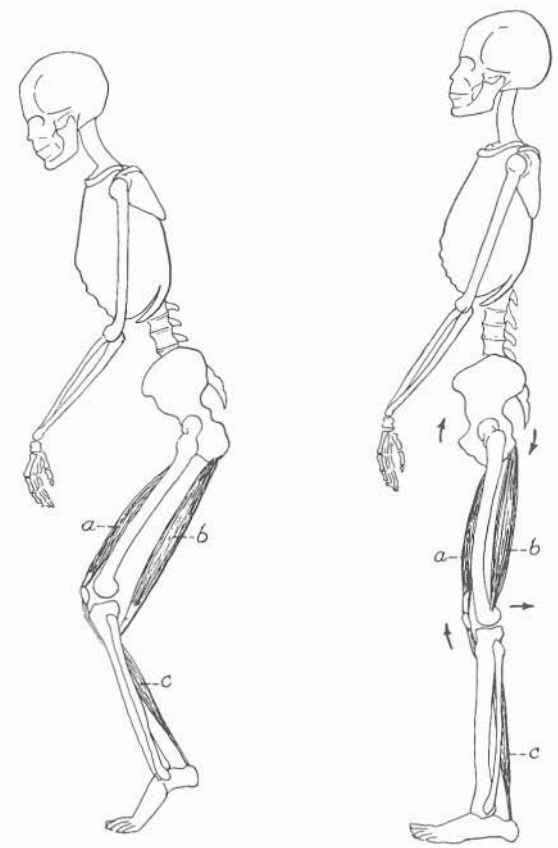


FIGURE 24–44 Eggers's transfer of hamstrings to femoral condyles. *a*, quadriceps femoris muscle; *b*, hamstring muscles; *c*, soleus muscle. (From Eggers GNM: Transplantation of hamstring tendons to femoral condyles in order to improve hip extension and to decrease knee flexion in cerebral spastic paralysis. J Bone Joint Surg 1952;34-A:827.)

continuous EMG, and those with normal EMG. They found that swing phase knee flexion improved in all groups but felt that the best results were seen in the patients who had phasic but inappropriate rectus EMG activity in swing phase.[289]

Other predictive variables used to study rectus femoris transfers include walking speed, dynamic range of motion, and joint kinetics. It has been found that patients whose walking speed is at least 80 percent that of age-matched normal subjects walk better following rectus transfers than their slower counterparts.[117] Patients who have 80 percent or more of normal dynamic range of motion of the knee on preoperative gait analysis do not appear to benefit from rectus transfer.[329] And those patients who have good power generation at the ankle and hip do best with rectus transfer.[160] It is logical that if a patient has difficulty initiating swing phase and cannot powerfully flex the hip to lift it from the ground, there is little momentum to produce swing phase knee flexion. If power is satisfactory, however, there is sufficient momentum to allow for knee flexion if the rectus femoris spasticity does not interfere. Hence the better results for transfers in the presence of good joint power. Inferior outcomes have been described in patients who underwent rectus transfers but who had rotational abnormalities exceeding 8 degrees.[164] If the knee and feet are not pointing straight ahead, swing phase knee flexion does not occur in the sagittal plane and rectus transfer is not optimal.

On review of the literature, our current indications for distal hamstring lengthening with simultaneous rectus femoris transfer are the following:

1. For significant crouch gait during stance phase with limited knee extension at midstance
2. For increased popliteal angle and positive rectus grab on clinical examination
3. If EMG shows activity in the rectus femoris during swing phase
4. In the case of sufficient hip pull-off power generation at late stance phase, or no preceding iliopsoas release
5. For velocity greater than 60 percent of normal
6. If there are no significant rotational abnormalities of the hips that interfere with gait

Although the popliteal angle improves following hamstring lengthening with or without rectus femoris transfer, there is usually a slow, gradual loss of correction over time with growth.[112] Recurrence of contracture requiring reoperation is not uncommon, occurring in 22 of 126 patients at 3 to 14 years of follow-up in one study[127] and in 12 percent in another study.[25] Loss of knee range of motion is commonly seen in many adolescents with cerebral palsy regardless of whether or not they have had prior hamstring surgery.[225] Repeat hamstring lengthening can be performed, but the procedure is more difficult because of scarring in the tendons from the first surgery. Recurrence has not correlated with the age of the patient at the time of initial lengthening.[126]

On occasion, hamstring lengthening in a severely contracted patient can lead to nerve palsy, particularly of the peroneal nerve. In our small experience, these patients are extremely difficult to manage, and assistance with medical management of paresthesias is often necessary. A mathematical equation is available to predict how much straightening can be performed safely, but clinical judgment in not aggressively stretching the posterior knee in patients with significant contractures is required.[22] When nerve palsy does occur, the pain from the paresthesias can be treated with such medications as Elavil or Neurontin.

ROTATIONAL OSTEOTOMIES. Spasticity in the lower extremities leads over time to the development of rotational abnormalities in the femur and tibia. Typically, persistent femoral anteversion is present in patients with spastic diplegia and in some patients with severe spastic hemiplegia. Femoral anteversion presents as intoeing in the schoolage child. Patients and their families complain of frequent falling and difficulty advancing one leg past the other during gait. When femoral anteversion is combined with scissoring and tight adductors, the inturned foot can become quite an obstruction in swing phase (Fig. 24–47).

Physical examination shows increased internal rotation and decreased external rotation of the hips. The patient's patellae appear internally deviated during gait, a finding that is made more apparent by circling the child's patella and watching the child walk toward the examiner. Care is needed when assessing a child's gait for femoral rotation, as pelvic rotation may also be present and confounds the clinical picture.

Over time, compensatory external rotation of the tibia develops and the foot progression angle turns more external. At this time rotational abnormalities may be missed without careful observation of gait. The child does not appear to be intoeing, yet the patellae are still pointing significantly inward. The foot progression angle may actually be external if the external tibial torsion is severe enough. Pes valgus is frequently present also.

Internal tibial torsion may also be present in children with cerebral palsy, specifically patients with spastic hemiplegia. Clinically, the torsion can be quantified by examining the bimalleolar angle. The lateral malleolus should be 25 to 30 degrees posterior to the medial malleolus when the patient is seated and the knee is pointing directly forward. Varus deformity of the foot due to spasticity in the posterior or anterior tibialis muscles can produce an internal foot-thigh angle, so the bimalleolar angle is more specific for internal tibial torsion.

In some patients, more precise information about the amount and levels of rotation can be obtained through gait analysis. The foot progression angle can be quantified accurately. Transverse plane rotation of the pelvis, femur, and tibia and foot can be documented and the appropriate level of osteotomy planned. Although computerized gait analysis is more accurate in complex cases than observation, it should be noted that patients with severe crouch gait may have measurement errors in the estimation of transverse plane rotation even on sophisticated gait analysis studies.[366]

The medial hamstrings, adductors, and gluteus medius and minimus can all produce dynamic internal rotation of the hips in children with cerebral palsy.[464] Lengthening of the medial hamstrings and adductors may in some cases lead to less dynamic internal rotation of the hip, but the amount of correction is usually slight and not very predictable. In 1980 Steel described a procedure in which the gluteus medius and minimus are transferred from the trochanter to the anterior surface of the proximal femur. He found improvement in femoral rotation postoperatively without abductor weakness in 90 percent of 42 hips. He noted prob-

Hamstring Lengthening

A, The hamstrings are approached through two longitudinal incisions. The medial incision is placed over the gracilis tendon and the lateral incision is placed lateral to the biceps femoris, to protect the peroneal nerve.

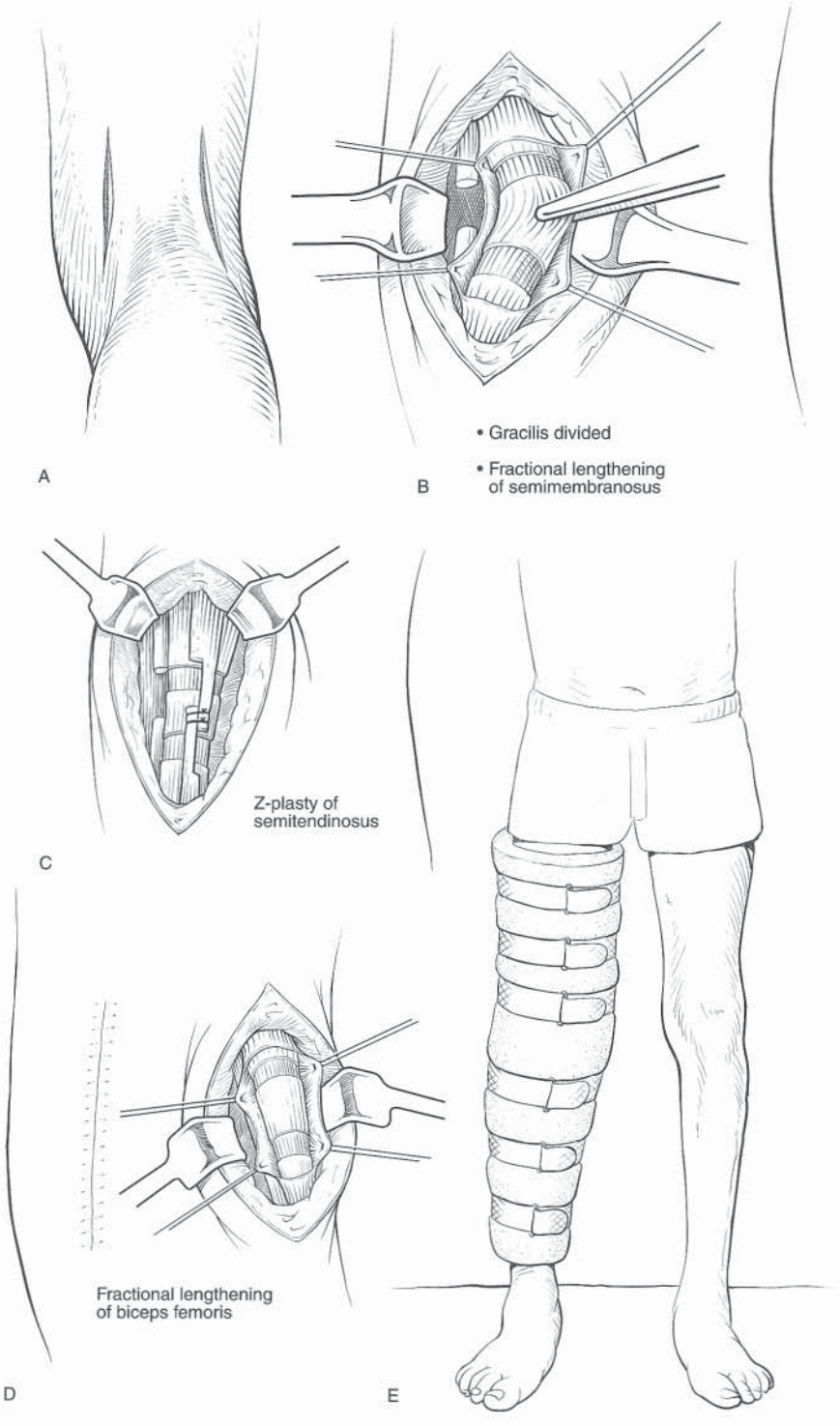
B, All three medial hamstrings are first identified. Following this step, the gracilis tendon may be divided or lengthened. The aponeurosis of the semimembranosus is cut, leaving the underlying muscle fibers in continuity. Usually two cuts in the aponeurosis are required, spaced approximately 1.5 to 2.0 cm apart. By extending the patient's knee and flexing the hip, the surgeon can perform a sliding lengthening.

C, The semitendinosus tendon is lengthened with a Z-plasty and repaired.

D, The biceps femoris is identified and the peroneal nerve protected, as it lies directly medial and deep to the tendon. An intramuscular lengthening procedure is performed by incising just the tendinous portion of the biceps, leaving the muscular fibers in continuity, as was done for the semimembranosus lengthening. Again, the hip is flexed and the knee extended to achieve a sliding lengthening.

E, The patient is then placed either in a knee immobilizer or in a long-leg cast with a straight knee. When other tendon or bony surgery is performed simultaneously, the mode of immobilization may vary. Early mobilization and weightbearing are encouraged.

PLATE 24–5. Hamstring Lengthening



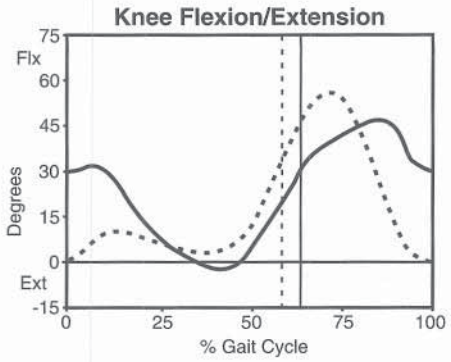


FIGURE 24–45 Hamstring spasticity leads to the inability to extend the knee to accept weight at initial contact (which occurs at 0 percent of the gait cycle). When the rectus femoris is spastic, the knee is unable to flex rapidly at initial swing phase, which is to the right of the vertical lines. The amount of swing phase knee flexion is decreased, and its timing is delayed. Normal kinematics are depicted by the *dotted line*, and those of a child with spastic diplegia by the *solid line*.

lems with the transfer pulling off the proximal femur in some patients, which could lead to weakness in abduction.[446] Continued problems with abductor weakness have led to a modification of the technique in which just the anterior fibers of the gluteus medius and minimus are transferred, with improved results in a small series.[226] Few centers continue to perform gluteal transfers for internal rotation gait in cerebral palsy.

Correction of rotational malalignment of the lower extremity is best achieved through derotational osteotomies.[234,481] Femoral anteversion is treated with femoral osteotomy, either proximally at the inter- or subtrochanteric level or distally at the supracondylar level (Fig. 24–48).

Those who advocate the proximal osteotomy feel that rotation of the extensor mechanism with the distal osteotomy is undesirable, although comparative studies have not been done. Computer simulation of intertrochanteric, subtrochanteric, and supracondylar osteotomies has shown minimal effect on the length of the hamstring and adductor muscles.[414] If the osteotomy is performed proximally, the patient is usually positioned prone on the operating table. Rotation to allow twice as much external rotation of the hip as internal rotation is the goal—in other words, 30 degrees of internal rotation and 60 degrees of external rotation.[377] Mathematical models have been devised to quantify the amount of rotation needed intraoperatively[365] but have not been widely used. Fixation with a blade plate or a plate and screws is performed,[444,480] and postoperative immobilization is used when the surgeon feels there is a risk of loss of fixation due to osteopenia.

Distal osteotomy is performed at the supracondylar level through a lateral approach with the patient supine and the legs draped free.[209] The femur is exposed by elevating the vastus lateralis anteriorly off the intramuscular septum. K-wires are used to quantify the amount of rotation intraoperatively.[237] Hoffer and colleagues used Steinmann pins to quantify rotation, and then used the pins for fixation by

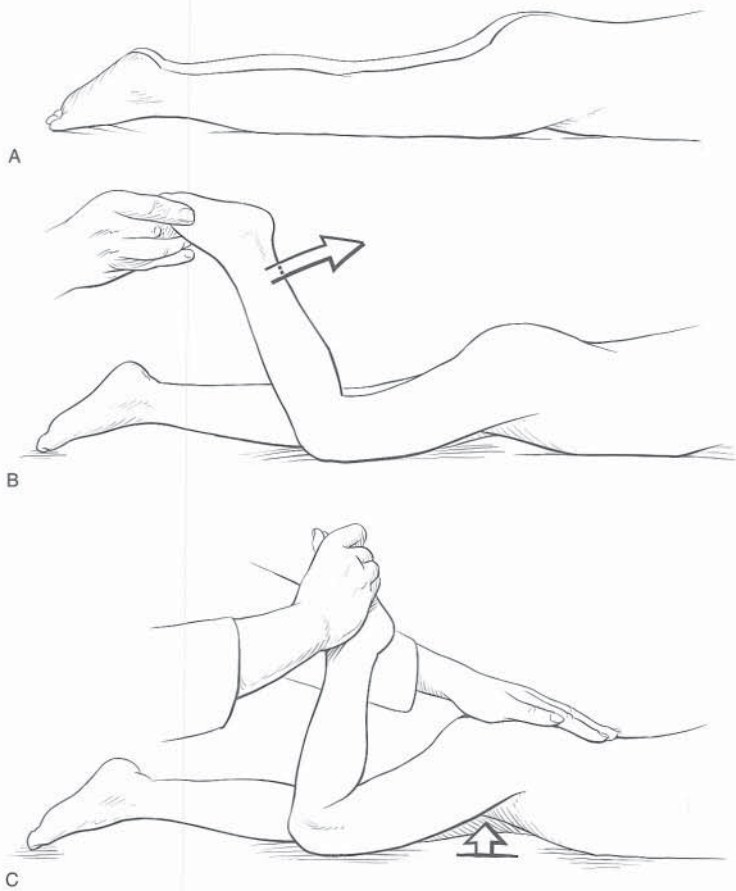


FIGURE 24–46 **A** to **C**, The Duncan Ely test. With the patient prone, the knee is passively flexed. A positive result occurs when the ipsilateral buttock rises, which may indicate rectus femoris spasticity.

incorporating them into the cast.[209] They did encounter pin tract infections, so those who perform distal osteotomy now generally use stable internal fixation with a plate and screws.[90] Hip rotation can be assessed in flexion following provisional fixation so that symmetry in internal and external rotation can be achieved. The benefits of performing the osteotomy distally are ease of the procedure and the ability to use a tourniquet. Immobilization with long-leg casts in knee extension allows standing and early weightbearing.

In patients with tibial rotational deformities, surgical correction should be performed at the distal level. Proximal osteotomies are associated with a higher risk of neurovascular injury.[412] Internal fixation with a plate and screws or crossed K-wire fixation can be performed with a low complication rate (Fig. 24–49).[131] Gait studies have shown that realignment of the tibia tends to normalize the forces working at the ankle and foot.[447]

Hip Surgery. Hip surgery in cerebral palsy can be divided into three areas: (1) correction of hip flexion contracture, (2) correction of increased hip adduction during gait, or scissoring, and (3) surgery to treat the subluxating or dislocating hip.

HIP FLEXION CONTRACTURE SURGERY. Hip flexion contractures are found most commonly in patients with spastic diplegia and spastic quadriplegia and are one component of the patient's overall crouch gait pattern. Hip flexion contractures are nearly always seen in combination with increased hip adduction and internal rotation, knee flexion due to hamstring spasticity, and either equinus, calcaneus, or valgus deformities of the feet. As such, surgery to improve hip flexion contractures is always done in conjunction with other soft tissue or bony procedures in patients with cerebral palsy.[385]

The flexion contracture is due to increased tone in the hip flexors, primarily the iliopsoas, and relative weakness of the hip extensors, such as the gluteal muscles. The contracture is identified during the physical examination by performing the Thomas and Staheli maneuvers.[32] In the Thomas test, described by Hugh Owen Thomas in 1876, the patient is positioned supine on the examining table. The opposite hip is fully flexed to flatten the lordosis of the lumbar spine and lock the pelvis against moving. The angle between the table and the hip in question is then measured, as the hip will rise up in flexion off the table in the presence of a contracture (Fig. 24–50). The Staheli test is performed by placing the upper body of the patient prone on the table, with the hips dangling off the edge of the table. The angle formed by the horizontal and the thigh, at the point at which further hip extension causes the pelvis to move, is the hip flexion contracture (Fig. 24–51).[441]

During gait, a hip flexion contracture is apparent as either increased flexion of the hip during the middle of stance phase (when the hip should be extended) or as increased anterior pelvic tilt. The anterior pelvic tilt produces either a forward lean of the upper body during gait or increased lumbar lordosis as the spine extends above the flexed pelvis.[198]

Radiographs can be used to objectively quantify the hip flexion contracture. Bleck in 1971 described use of the sacrofemoral angle. A standing lateral radiograph is taken to include the proximal femoral shaft and the lumbar spine. A line is drawn along the superior surface of the sacrum, and another along the femoral shaft. The intersection of these lines is the sacrofemoral angle, which normally should be between 45 and 65 degrees. In the presence of a hip flexion contracture, the sacrofemoral angle decreases (Fig. 24–52).[46]

Hip flexor surgery in the walking child is done to improve the hip flexion contracture, but more often than not the goal of the surgery is to prevent increasing hip flexion and anterior pelvic tilt when hamstring lengthening is performed. As noted earlier in the discussion on knee surgery, the hamstrings are not only knee flexors, they are also hip extensors. The hamstrings lose strength following lengthening, so any preexisting hip flexion contracture will be exacerbated following hamstring surgery.[121] Therefore, hip flexor lengthening is usually part of the overall surgical correction of crouch gait.

The recommended procedure to correct increased hip flexion is a psoas tenotomy done over the pelvic brim. The surgical approach is anterior, through an oblique incision just distal to the anterior-superior iliac spine. The psoas is located between the sartorius and the femoral sheath. The femoral nerve nearly overlies the psoas tendon. The tendon of the psoas is identified deep within the iliacus muscle, which is not lengthened. The tendon is then transected and slid within the iliacus, thereby increasing the overall length of the iliopsoas.[273,465] This is similar to the technique described by Salter as part of his innominate osteotomy.[404] Gait analysis studies have shown improvement in hip extension during stance, in hip moments and powers, and no loss of strength following lengthening either at the pelvic brim or over it.[86,465]

Release of the iliopsoas tendon off the lesser trochanter of the femur should not be done in ambulatory patients, as it results in loss of hip power and inability to forcibly flex the hip against gravity. Climbing stairs becomes extremely difficult, and gait deteriorates.[46] The gait of children who have undergone iliopsoas release is characterized by increased pelvic motion and decreased arc of hip flexion and extension, as the trunk tries to substitute for the weak hip flexors in pulling the leg forward off the ground. Others circumduct to advance the leg.

Bleck advised against simply releasing the iliopsoas as well, and suggested attaching the distal iliopsoas tendon anteriorly into the hip capsule. This would allow for additional length, yet hip flexion would be preserved.[46] Because psoas tenotomy over the pelvic brim is technically easier, transferring the tendon to the capsule is rarely done at present.

ADDUCTION CONTRACTURE SURGERY. Spasticity in the adductor muscles in cerebral palsy results in a narrow base of gait and scissoring. The patient has difficulty advancing one limb in front of the other as the swing limb contacts the ground in front of the other leg. Young children may be unable to progress in their ability to ambulate because of the scissoring (Fig. 24–53). Over time, the untreated adduction contractures, when combined with a hip flexion contracture, lead to progressive hip subluxation and possible dislocation. Surgery for the unstable hip in patients with cerebral palsy will be discussed in the next section.

Text continued on page 1178

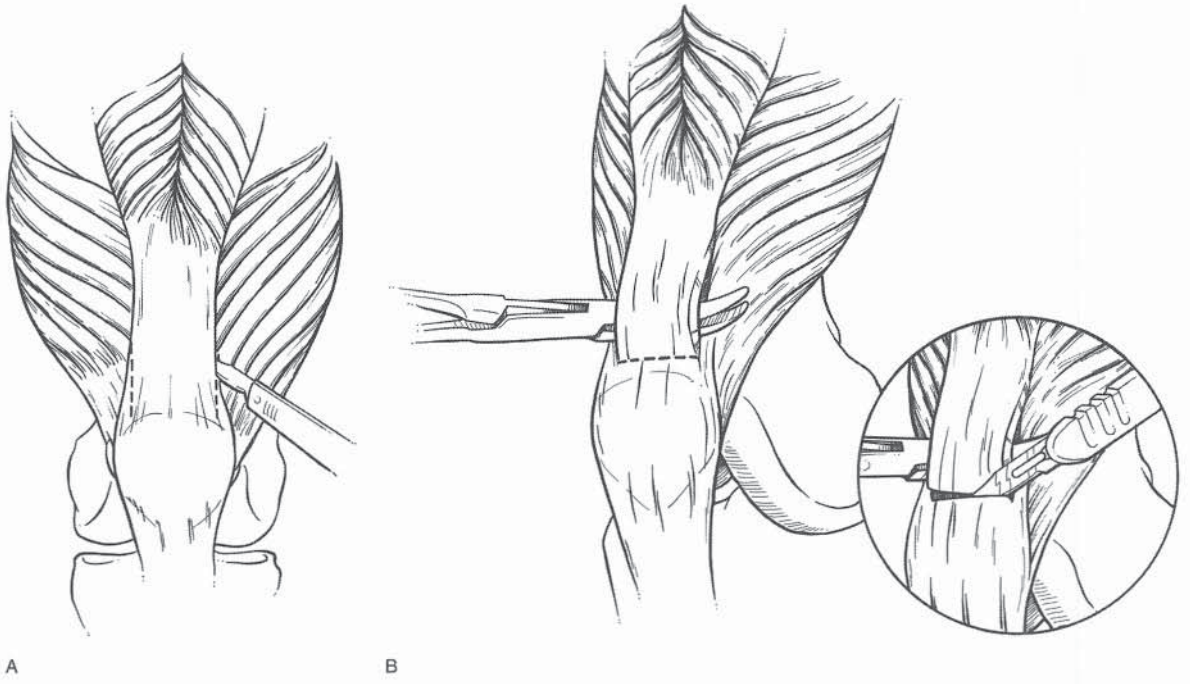
Rectus Femoris Transfer

A, The incision may be either a horizontal incision two fingerbreadths proximal to the proximal pole of the patella or a vertical incision. The conjoined quadriceps tendon is isolated and the rectus femoris component is identified. The rectus tendon is separated from the tendinous portions of the vastus medialis and lateralis.

B, The undersurface of the rectus must be carefully separated from the intermedius. This is most easily done proximally and extended by following the plane to the insertion on the patella. The rectus tendon can then be sharply released from the patella.

C, A strong suture is woven into the rectus tendon to be used in the transfer.

PLATE 24–6. Rectus Femoris Transfer



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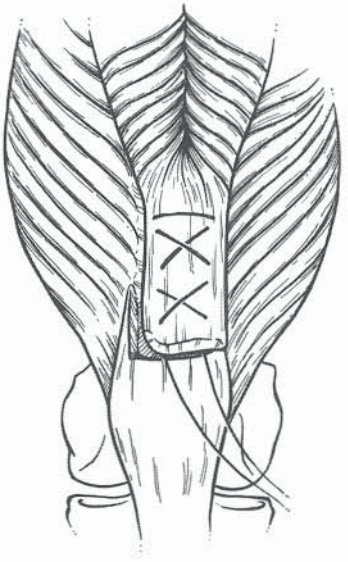


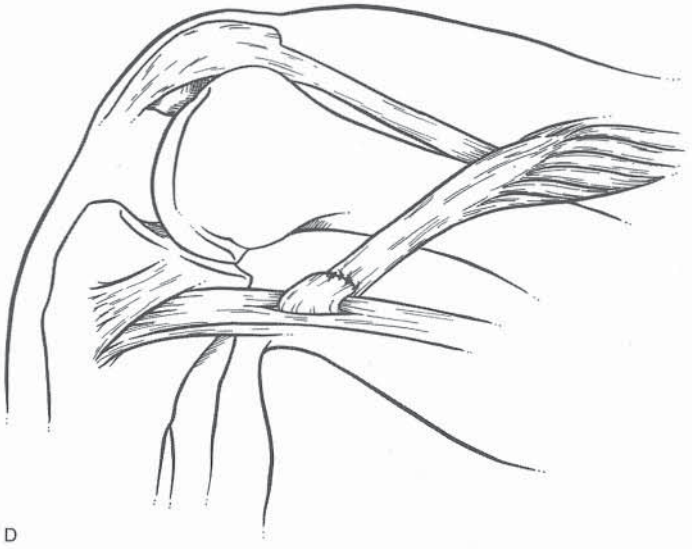
C

Rectus Femoris Transfer *Continued*

D, A medial posterior longitudinal incision is made and the gracilis tendon is isolated. The rectus is mobilized and transferred back into the posterior wound by grasping the suture. Then tendon is routed subcutaneously. The distal end of the rectus tendon is inserted through the tendon selected for the site of transfer and sutured back to itself. The gracilis is a popular site for the transfer, but use of the sartorius and biceps femoris has also been described.

E, The vastus medialis and lateralis may be repaired to each other to recreate the quadriceps tendon. The patient can be immobilized either in a long-leg cast or in a knee immobilizer.

PLATE 24–6. Rectus Femoris Transfer



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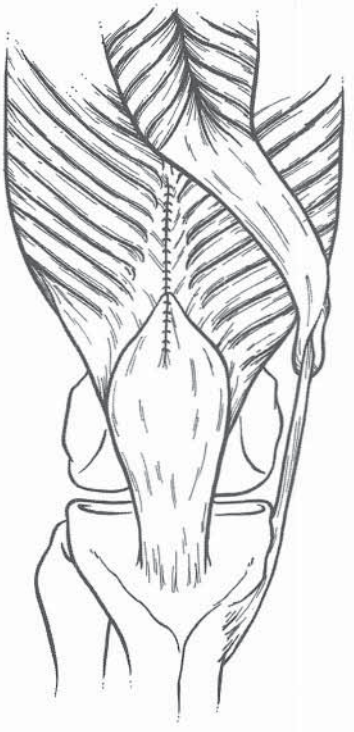

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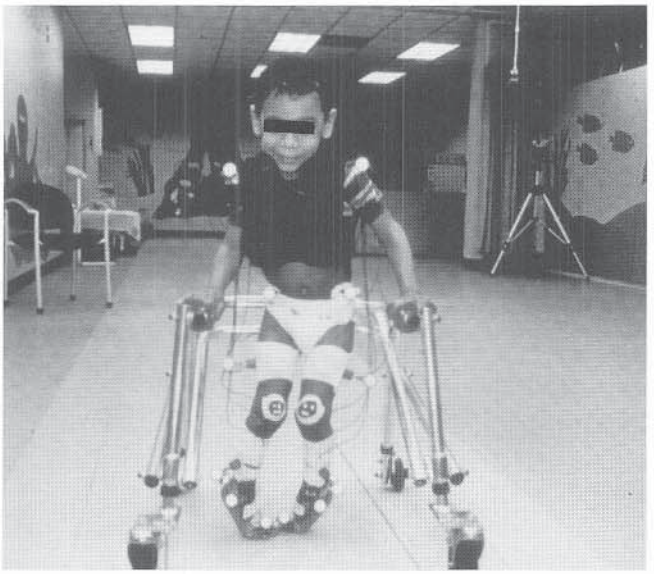

FIGURE 24–47 Excessive anteversion bilaterally in child with cerebral palsy. Tape outlines the patellas. He has difficulty clearing his foot forward in swing phase owing to intoeing from the anteversion and exacerbated by scissoring.

The muscles leading to the adduction contracture are the adductor longus, adductor brevis, adductor magnus, gracilis, and occasionally the pectineus.

Clinical examination reveals inability to abduct the hips in flexion and in extension. The tendon of the adductor longus is palpable and visibly tight in the groin. The child walks with knees knocking, and one foot scissors over the other in stance phase. The feet may appear locked together, as the child has difficulty initiating swing phase. EMG has shown abnormal swing phase electrical activity in the adductor muscles in patients with scissoring.[188]

A word of caution is needed here. Increased femoral anteversion when combined with crouch at the knee can produce the appearance of scissoring.[120] This clinical scenario has been termed pseudoadduction. Careful observation of the patellae during gait will alert the surgeon to the internal rotation. When the narrow base of gait is secondary to hamstring contractures and femoral anteversion, adductor surgery will be ineffective in improving the child's ability to walk.

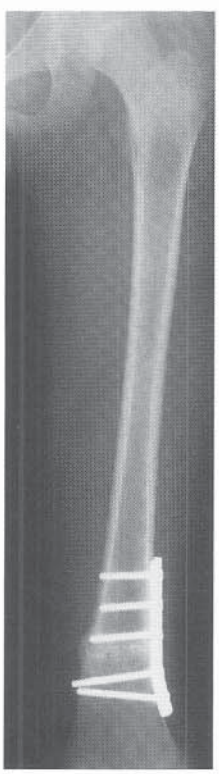

FIGURE 24–48 Distal femoral rotational osteotomy for treatment of excessive anteversion in a 10-year-old boy with spastic hemiplegia.

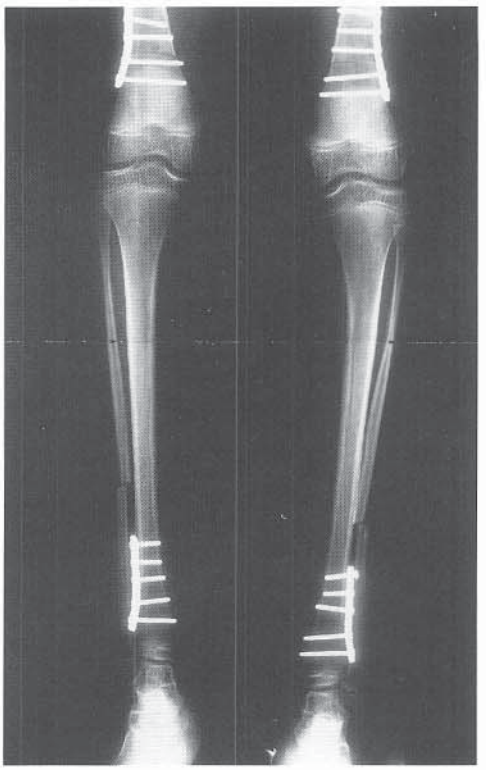

FIGURE 24–49 Bilateral rotational osteotomies of the femurs and tibias for the treatment of femoral anteversion and external tibial torsion.

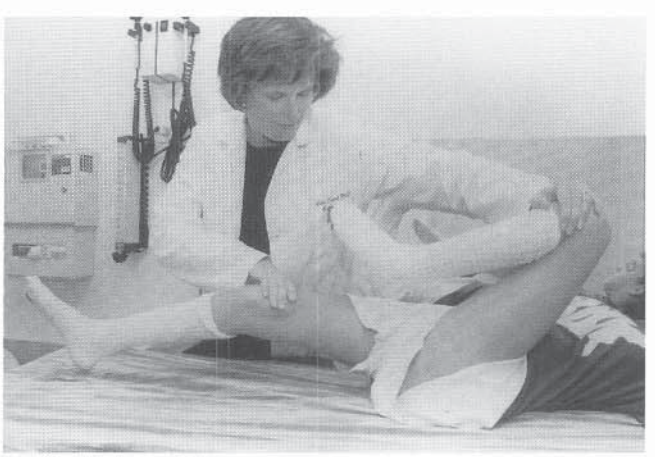

FIGURE 24–50 Thomas test reveals a 20-degree flexion contracture of the right hip. The opposite hip is fully flexed to flatten lumbar lordosis.

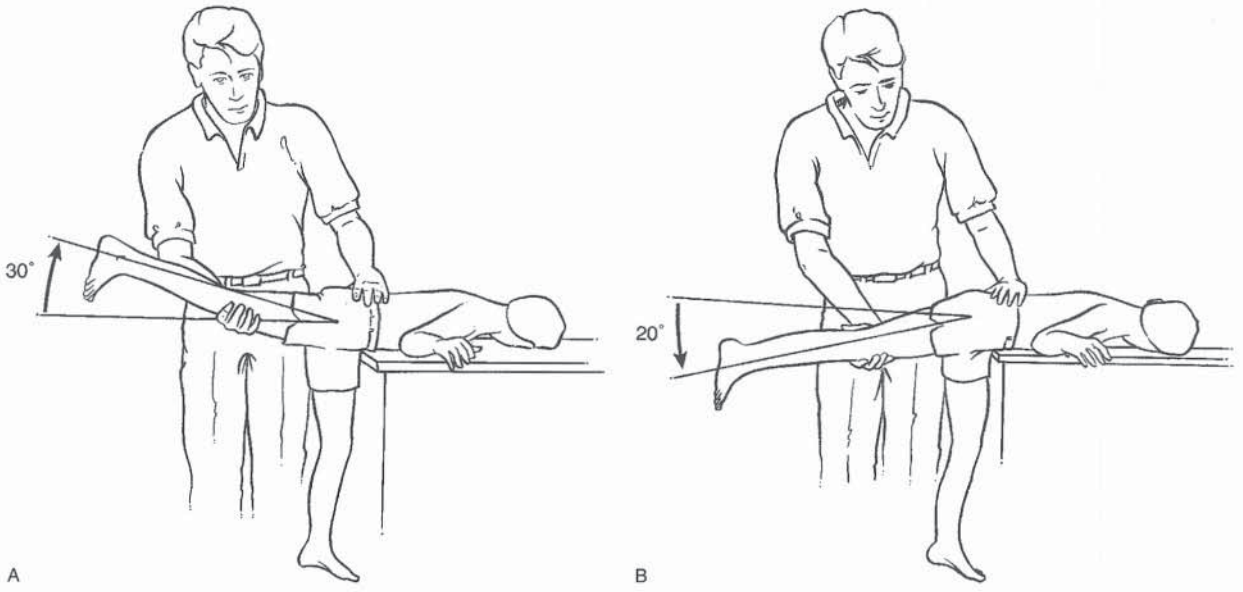

FIGURE 24–51 Staheli test, used to determine hip flexion deformity with the patient prone. **A,** The pelvis is stabilized, the patient's thigh is raised toward the ceiling, and the tested hip is extended. Normal extension is 30 degrees. **B,** The degree by which the hip fails to reach neutral position is the degree of deformity.

Bracing has not been shown to improve adduction contractures, and although botulinum toxin injections may relieve dynamic adduction, this treatment modality is still under investigation.[74] Surgery to improve adduction contractures is limited to adductor release, with or without obturator neurectomy, and posterior adductor transfer.

Adductor release was initially described by Banks and Green[30] and is commonly performed in the young child with cerebral palsy who is able to stand with support but has difficulty walking because of scissoring (Plate 24–7). It is also commonly a component of multiple soft tissue single-stage procedures in children with cerebral palsy who are ambulatory. A short transverse incision is made in the groin crease. The adductor longus tendon is detached from its origin on the superior pubic ramus, often along with at least

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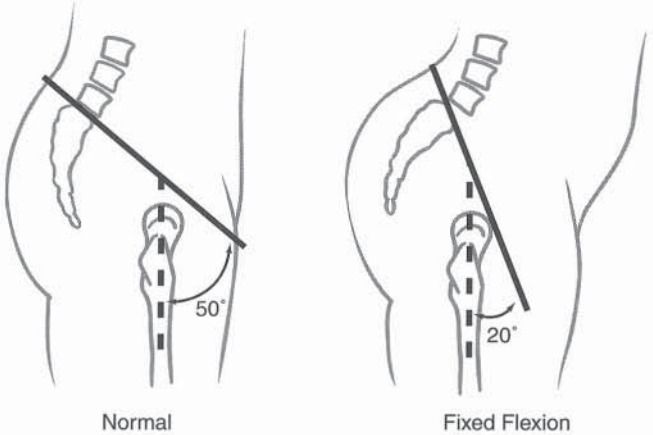


FIGURE 24–52 The *sacrofemoral angle.* With increasing hip flexion contracture the pelvis tips forward and the sacrum becomes more vertical. The angle formed between a line drawn along the superior aspect of the sacrum and the femoral shaft decreases with flexion of the hip.

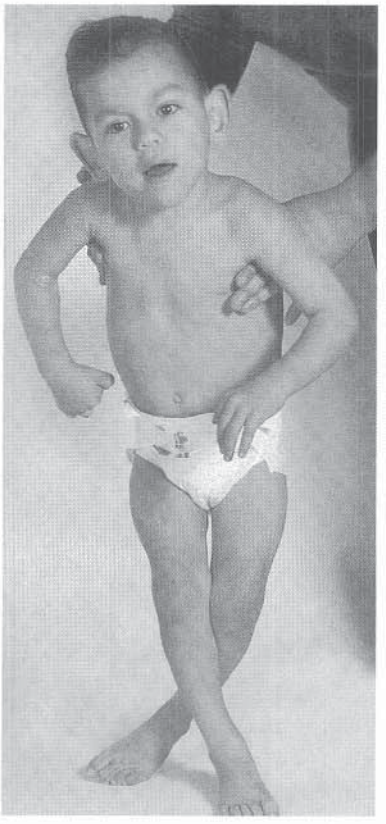


FIGURE 24–53 Three-year-old boy with cerebral palsy and scissoring of the hips.

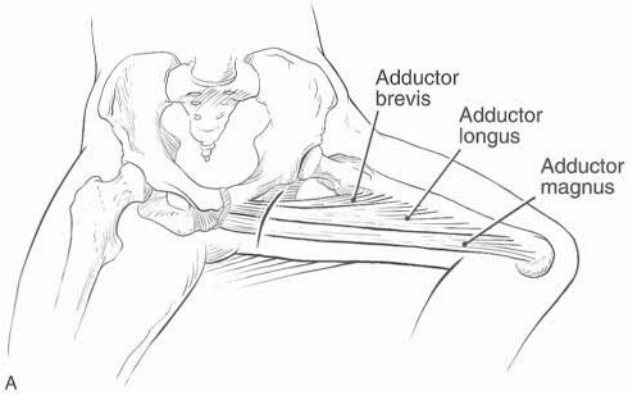
Technique for Adductor Contracture Release

A, A transverse incision is made in the groin crease and centered over the adductor longus tendon, which is easily palpated.

B, The adductor longus tendon is identified, isolated from the deeper adductor brevis, and divided using electrocautery.

C, The adductor brevis is then divided in part using electrocautery. Care is taken to identify the anterior branch of the obturator nerve, which should be preserved. The posterior branch of the obturator nerve, which lies deep to the adductor brevis, likewise should be preserved.

PLATE 24–7. Technique for Adductor Contracture Release



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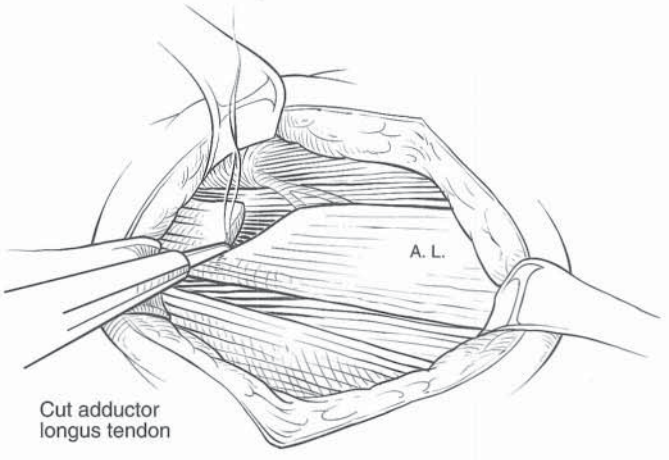

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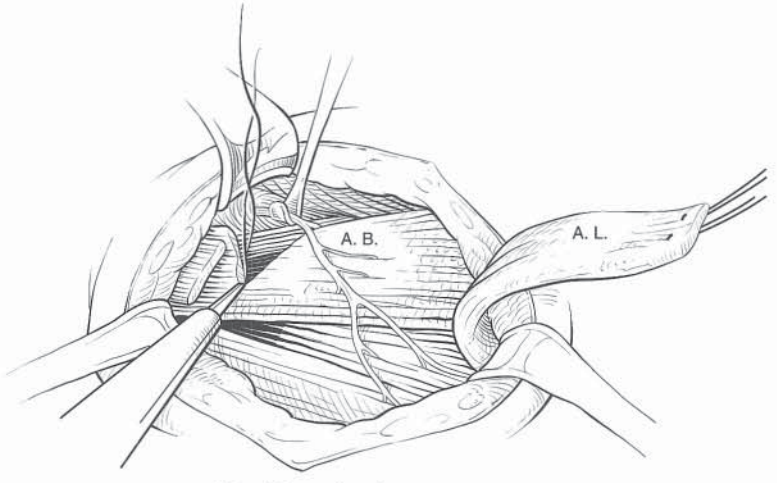


C

- Cut adductor brevis
- Keep obturator branches intact

Technique for Adductor Contracture Release *Continued*

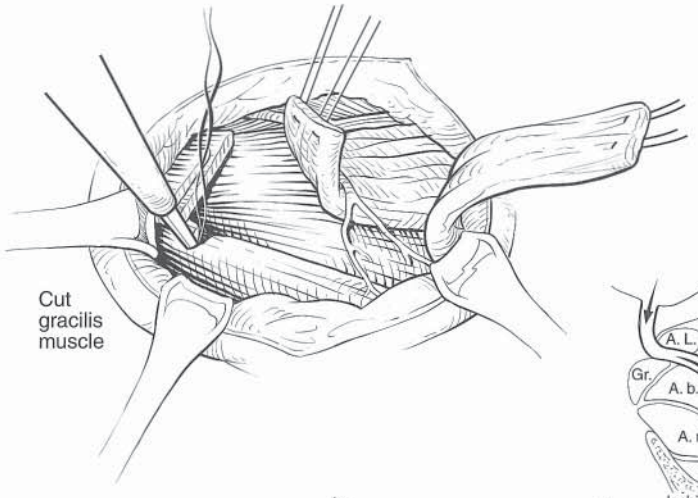
D, Just posterior to the adductor brevis and more superficial, the gracilis is identified. It is a flat, broad muscle. It is released from its origin using electrocautery.

E, If a concomitant release of the iliopsoas is performed in a nonambulatory child, its tendinous insertion on the lesser trochanter can be palpated deep to adductor brevis.

F, A fractional lengthening of the psoas tendon can be performed at this level, as illustrated, or preferably more proximally at the pelvic brim.

G, Immobilization consists of two long-leg casts with a removable abduction bar. The bar can be removed for range-of-motion exercises and transport but should be used most of the day for 3 to 4 weeks. The hip flexion contracture release is best treated by placing the child in the prone position at frequent intervals.

PLATE 24–7. Technique for Adductor Contracture Release



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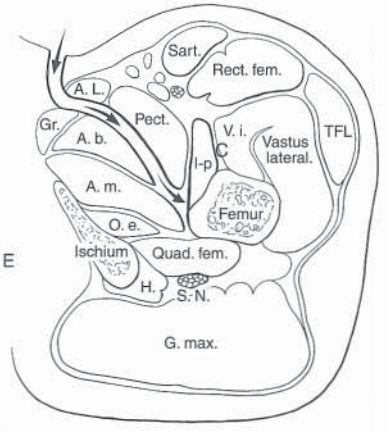

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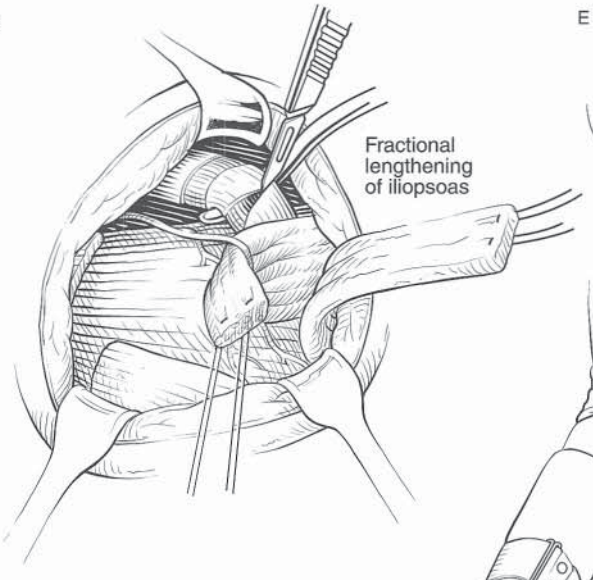

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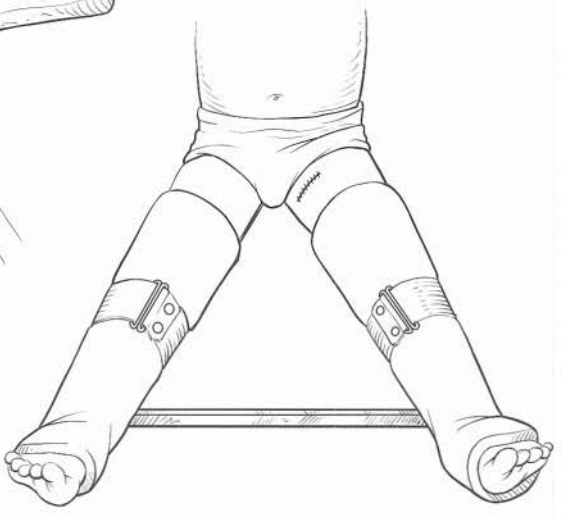


G

part of the adductor brevis and the gracilis origins. The adductor magnus is not released. The adductor brevis is sandwiched between the anterior and posterior branches of the obturator nerve, which innervate the adductor musculature. These branches should be identified to avoid injury to the nerves. The patient is then placed into either long-leg casts held in abduction (Petrie casts) or a removable abduction bar. A spica cast is unnecessary unless other procedures are performed concomitantly.

The advantage of adductor release is that it is a simple and quick procedure. It results in increased abduction, and therefore improves scissoring.[488] It has been linked with the development of postoperative abduction contractures and wide-based gait, particularly when combined with an anterior branch obturator neurectomy that denervates the adductor brevis.[274,408] For this reason, anterior branch obturator neurectomy should not be performed. The adduction contracture may recur with growth, and further surgery is necessary in 10 to 37 percent of all children who undergo adductor release.[390,408]

The rate of recurrent contracture led Perry in the early 1960s to devise a procedure in which the adductor longus, adductor brevis, and gracilis tendons are transferred from the pubic ramus to the ischium (Fig. 24–54). The new origin of the muscle would convert the adductors into hip extensors, thereby lowering the risk of recurrent contractures and further stabilizing the hips. The surgery was designed for patients with poliomyelitis, but it soon began to be used in the cerebral palsy population. Many studies of adductor transfers then followed.

Aronson and colleagues studied the results of adductor transfer in 42 patients with cerebral palsy.[19] In 88 percent of patients, the transfer was successful in improving or maintaining abduction (from an average of 20 to 44 degrees), extension, and stability of the hip. Ambulatory status improved in patients with spastic diplegia but not in patients with spastic quadriplegia. The failures were all in nonambulatory patients. Couch reported on 32 children who were either able to walk or nearly so, and found a 30-degree increase in abduction following adductor transfers.[97] Root and Spero in 1981 compared adductor release with adductor transfer.[390] They concluded that adductor transfer was preferable, because of better maintenance of abduction. Endurance, scissoring, and standing balance improved most in the ambulatory patients who had undergone adductor transfer. Others have found no significant difference and advocate adductor release because it is simpler and better tolerated by the child.[376]

Loder and colleagues performed an interesting study in 1992.[261] They tagged the adductor tendons with metallic clips following insertion into the ischium and found that only 19 of 33 transfers remained attached to the ischium. If the tendon detaches, it functions as a tenotomy, which is inherently a simpler procedure. What's more, if only one side pulls off while the other remains attached to the ischium, asymmetry of the pelvis and hips can result. In a study from Houston reported by Scott and colleagues, pelvic obliquity had developed in 85 percent of patients, and unilateral hip subluxation had developed in 36 percent of patients 10 years after posterior transfer of the adductor tendons, presumably because of unilateral loss of fixation of the tendons to the ischium.[417] Beals and associates published the results of a modified adductor transfer in 141 hips.[37] In their procedure the adductor longus and brevis are sutured into the lengthened gracilis, which remains attached to its origin. They believe this may increase the integrity of the transfers, and they noted an improvement in abduction that was maintained at an average of 4 years postoperatively.[37]

We currently do not use the adductor transfer at our institution for the treatment of adduction contractures in cerebral palsy. We continue to perform adductor tenotomy but we no longer recommend obturator neurectomy because of the problems with abduction contractures. We mobilize the patients earlier than previously, and we use removable abduction bars more frequently now to decrease postoperative stiffness.

HIP SUBLUXATION/DISLOCATION SURGERY. Prior to an extensive discussion about the surgical reconstruction of the subluxated or dislocated hip in the patient with cerebral palsy, it is important to understand the epidemiology and etiology of hip instability in this condition.

Hip dysplasia or instability is a common problem in patients with cerebral palsy, occurring in approximately 21 percent of 1,450 patients at the Hospital for Special Surgery.[377] Other series report a prevalence of subluxation or dislocation ranging from 3 percent to 47 percent.[91,214,265,298] The incidence of hip dysplasia varies with the severity of neurologic involvement.[26,214] Rang and colleagues found a 22 percent prevalence of hip dysplasia in patients living with their families, and twice that in institutionalized patients.[367] Patients with spastic hemiplegia rarely develop hip dysplasia and dislocation. Patients with spastic diplegia are at increased risk. Patients with spastic quadriplegia who have total body involvement have the highest rate of hip instability, with almost 50 percent developing hip subluxation or dislocation.[167] The incidence of hip subluxation and dislocation is also linked to the ability of the patient to walk. Nonambulatory patients are at much higher risk than those who can walk, accounting for 89 percent of patients with hip instability and cerebral palsy.[409] Lonstein and Beck found that subluxated or dislocated hips developed in only 7 percent of independent ambulators but in 60 percent of independent sitters.[265] The mean age at which patients with cerebral palsy present with subluxation or dislocation is 7 years.[409]

Hip subluxation develops in response to muscle imbalance. Spasticity and contracture of the adductors and flexors of the hip overpower the weaker and noncontracted hip extensors and abductors. Subluxation develops gradually, with increasing lateralization and proximal migration of the femoral head with respect to the acetabulum. This is completely different from developmental dislocation of the hip, in which soft tissue laxity leads to instability of the hip (Fig. 24–55). The hip in cerebral palsy is not grossly unstable on clinical examination; it is the extremely rare case (usually a hypotonic child) in which an Ortolani maneuver is positive with reduction of the hip. Rather than laxity, the hip is pried from the acetabulum over time by spastic muscles. It has been found through computer modeling that the forces exerted across a spastic hip in cerebral palsy are up to six times greater than normal.[291]

Bony deformity, then, occurs in response to the spasticity. The normal remodeling of the femoral anteversion seen in

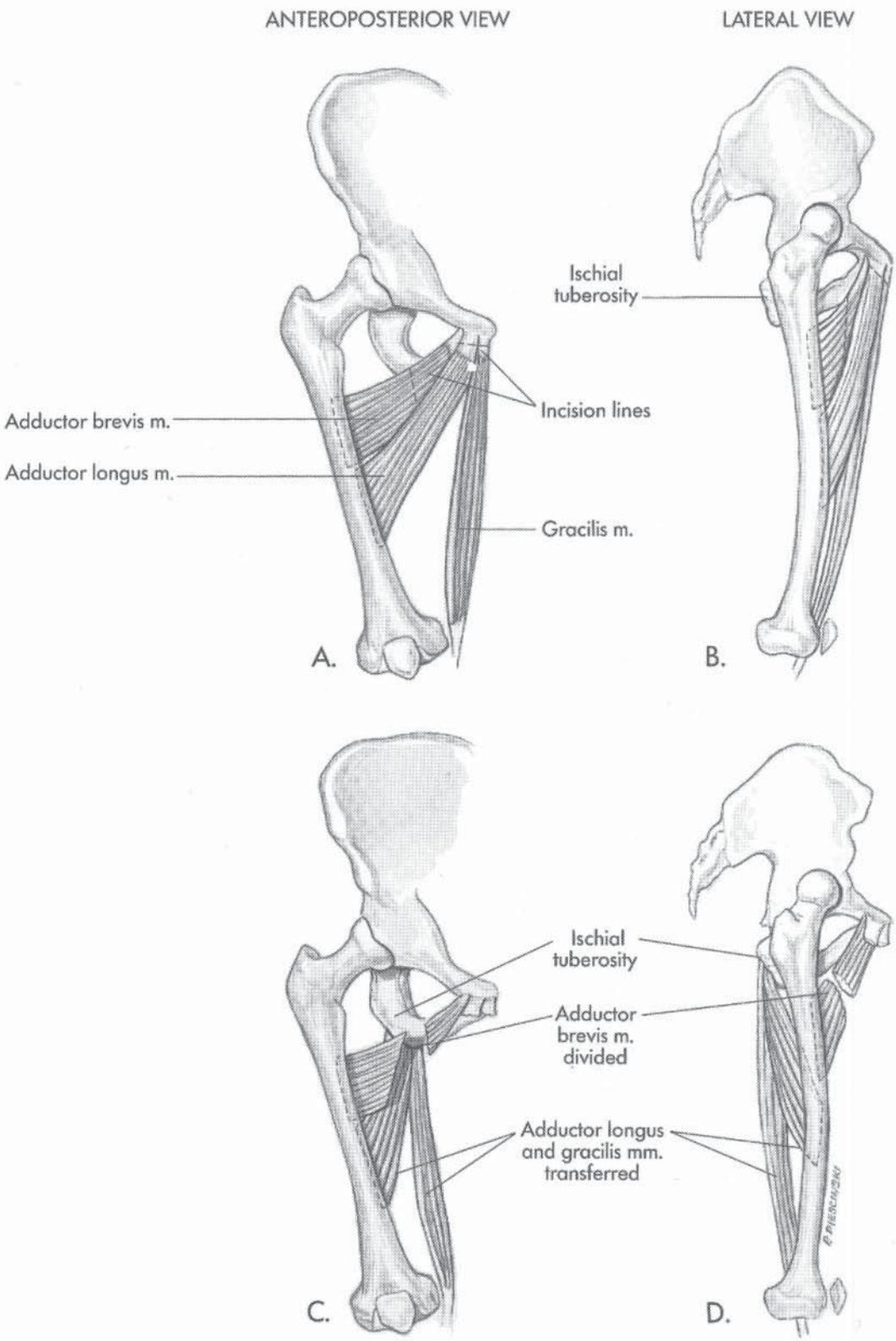


FIGURE 24–54 Posterior transfer of hip adductors to the ischium. **A** and **B,** AP and lateral views showing the line of division of the gracilis and adductor longus muscles at their origin and the line of myotomy of the adductor brevis. **C** and **D,** The adductor longus and gracilis muscles are transferred to the ischium, and the adductor brevis is divided.

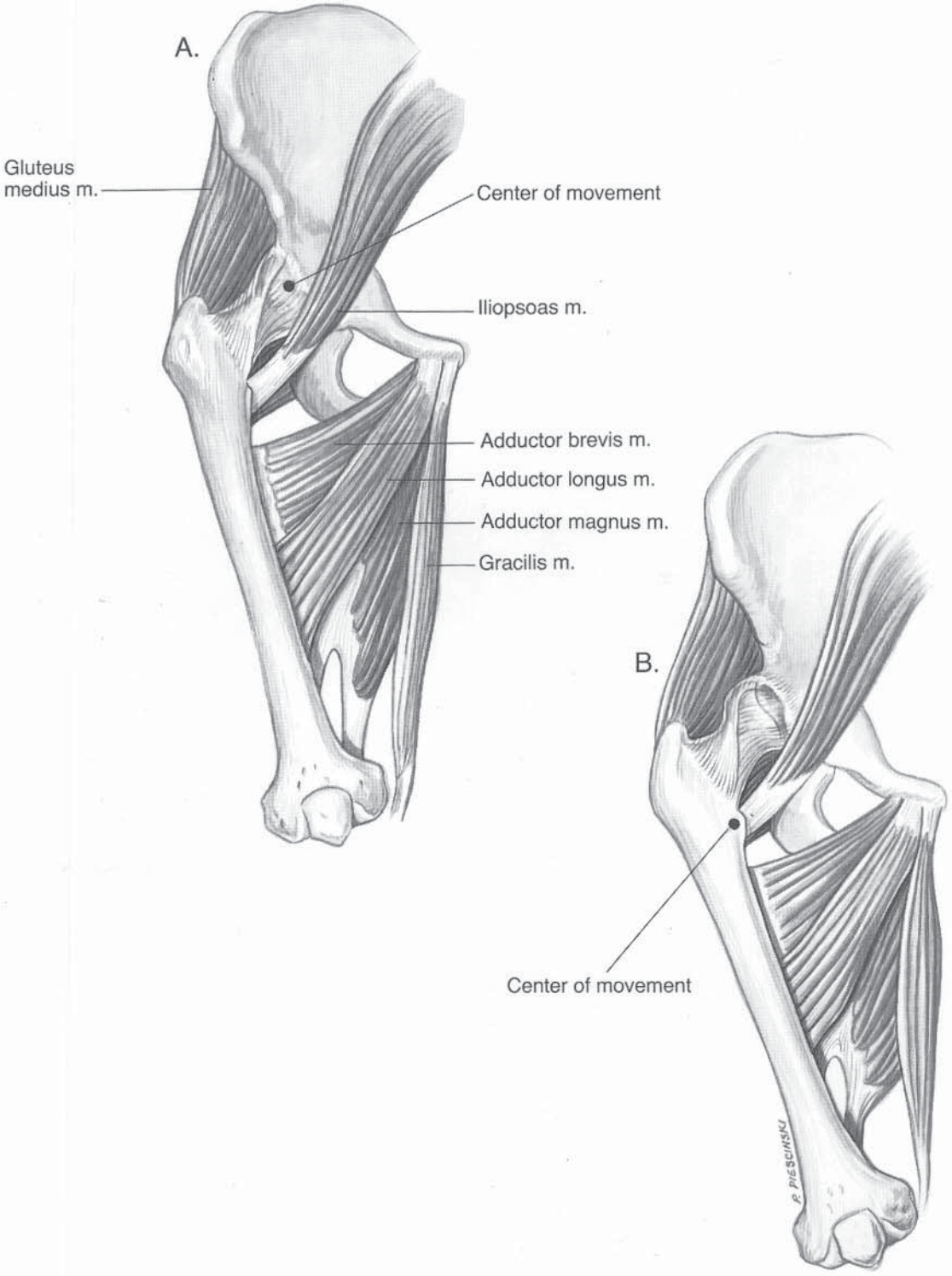


FIGURE 24–55 Mechanism of superior and posterior displacement of femoral head out of acetabulum. **A,** The normal hip. **B,** In cerebral palsy the hip adductor and iliopsoas muscles are spastic and shortened, and the gluteus maximus and medius muscles are weak. The center of movement of the hip is translated from the center of the femoral head distally to the level of the lesser trochanter. The hip joint capsule is elongated superoposteriorly, with gradual dislocation of the hip. (After Sharrard WJW: Paediatric Orthopaedics and Fractures, 2nd ed. Oxford, Blackwell Scientific Publications, 1979.)

the neurologically normal young child does not occur in patients with cerebral palsy, and anteversion persists into adulthood. The increased anteversion has been shown to correlate strongly with the development of hip dysplasia, particularly in nonwalking patients.[250] The neck-shaft angle becomes increased as coxa valga develops. The anteversion worsens the radiographic appearance of the valgus neck. The lesser trochanter becomes elongated due to pull from the iliopsoas. Acetabular changes occur as the hip subluxates, with an increased acetabular angle and erosion of the lateral lip of the acetabulum by the subluxating femoral head. Finally, changes in the shape of the femoral head take place, with superolateral and then superomedial notching from pressure from the capsule, the rim of the acetabulum, the abductors, and the ligamentum teres (Fig. 24–56).[44,266,409]

Because bony deformity develops in response to muscular spasticity, bony surgery in the absence of soft tissue release is ineffective in correcting subluxation or dislocation due to cerebral palsy. Likewise, by the time bony changes are seen, soft tissue surgery alone is likely to fail.

Hip subluxation or dislocation can be suspected from the results of the physical examination. Loss of range of motion is the first clue. Abduction will be limited, usually to less than 30 degrees, a hip flexion contracture is present, and there is increased internal and decreased external rotation of the hip. When dislocation is present and unilateral, a positive Galleazzi sign will be obvious, in which the thigh from the dislocated side appears shorter than the contralateral femur when both hips are flexed to 90 degrees and the knees are fully flexed bilaterally.

The diagnosis is then confirmed radiographically. The first signs of hip instability are a subtle break in Shenton's line and mild uncovering of the most lateral aspect of the femoral head by a shallow acetabulum. The amount of femoral head protruding past the lateral border of the acetabulum can be quantified by Reimers's migration index, which is the percent of the transverse diameter of the femoral head that lies lateral to Perkin's line, which is drawn at the edge of the acetabulum.[374] The acetabular index will be elevated due to acetabular dysplasia. The neck-shaft angle of the proximal femur is increased, indicating coxa valga and increased femoral anteversion (Fig. 24–57).[36]

With more significant subluxation, the lateral edge of the acetabulum becomes worn or eroded so that the acetabular index becomes very high and the capacity of the acetabulum appears reduced. The femoral head to teardrop distance increases as the hip begins to dislocate.

Treatment of hip dysplasia has the following goals: to provide a painless hip that allows stable sitting and positioning in the nonambulatory patient, and to reduce the hip fully so that ambulation can continue without groin or hip pain in patients who can walk. For the hip that is subluxated, surgery is done in large part to prevent dislocation. It is therefore important to know what a given hip's risk of dislocation is. Miller and Bagg studied 143 untreated hips in patients with cerebral palsy and determined the prevalence of progression of subluxation based on the age of the patients. They found that approximately 75 percent of hips that had Reimers's migration indices of less than 30 percent did not progressively subluxate, and those that did progress were in individuals less than 18 years old. All hips with migration indices of greater than 60 percent eventually dislocated. Subluxation persisted in the intermediate group with subluxation between 30 and 60 percent, with approximately 25 percent of hips worsening.[287]

Once it is determined that a given hip is likely to progress and therefore needs treatment, the risks and benefits of surgery should be weighed. The first question to be answered is, does treatment make a difference in hip subluxation and dislocation in the child with cerebral palsy? Great controversy exists, with several studies yielding differing results. Pritchett studied 100 severely involved nursing home patients with an average age of 26 years, 50 with hip dislocations that were untreated and 50 who had undergone surgical treatment for hip dislocations.[364] Only 50 percent of patients who underwent only soft tissue releases as treatment for hip subluxation had stable hips at follow-up, and 12 of 19 who had osteotomies of the femur with or without a pelvic osteotomy had stable hips. Pritchett included patients who had undergone salvage operations on the hip, such as resections and arthrodesis. There was no difference in pain, ease of perineal hygiene, or incidence of decubitus ulcers between the surgical and nonsurgical groups. Approximately 40 percent of patients in both groups had pain in the hip, which was usually classified as minor and did not interfere with daily activities. The incidence of scoliosis was the same irrespective of whether or not hip surgery had been done. He concluded that aggressive surgery to reduce dislocated hips offers no benefit to the well-being of the patient.[364]

Contrary to Pritchett's conclusion, Bleck advocates surgical reduction of hips in cerebral palsy, stating that one-third of patients with hip dislocations develop pain.[50]

Bagg and associates reviewed findings in their patients with hip subluxation and dislocation. Most of the patients had undergone some surgical treatment for their hips, generally either soft tissue releases or femoral osteotomy. Bagg and associates found that patients with hip dislocations had significantly more pain than those with either located or subluxated hips.[26] Interestingly, almost 25 percent of patients with reduced hips had mild pain, and a few even had severe pain. It remains difficult to know precisely what the influence of hip stability is on pain in this severely involved group of patients.

Cooperman and associates evaluated 38 patients who were not residing in institutions for the prevalence of pain with hip dislocation.[92] Twenty-four of 42 dislocated hips were not painful at 18-year follow-up and an average patient age of 26 years. In nine hips resection or fusion had been performed for pain, and nine additional hips were painful at follow-up. What makes this study different from the rest is the patient population: 50 percent of the patients were intelligent and communicative.

The next question to be asked is, does hip subluxation or dislocation lead to scoliosis? Letts and associates studied the association between the windblown hip, defined as one abducted reduced hip and a contralateral adducted subluxated hip, and pelvic obliquity. They found that the high side of the oblique pelvis correlated with the side of subluxation.[257] Next, Lonstein and Beck analyzed the prevalence of pelvic obliquity and scoliosis, and unilateral hip dislocation. They found that the proportion of dislocated hips was not related to the pelvic obliquity and that the direction of pelvic obliquity did not correlate with which hip was out. Scoliosis did correlate with pelvic obliquity, but the direction of the

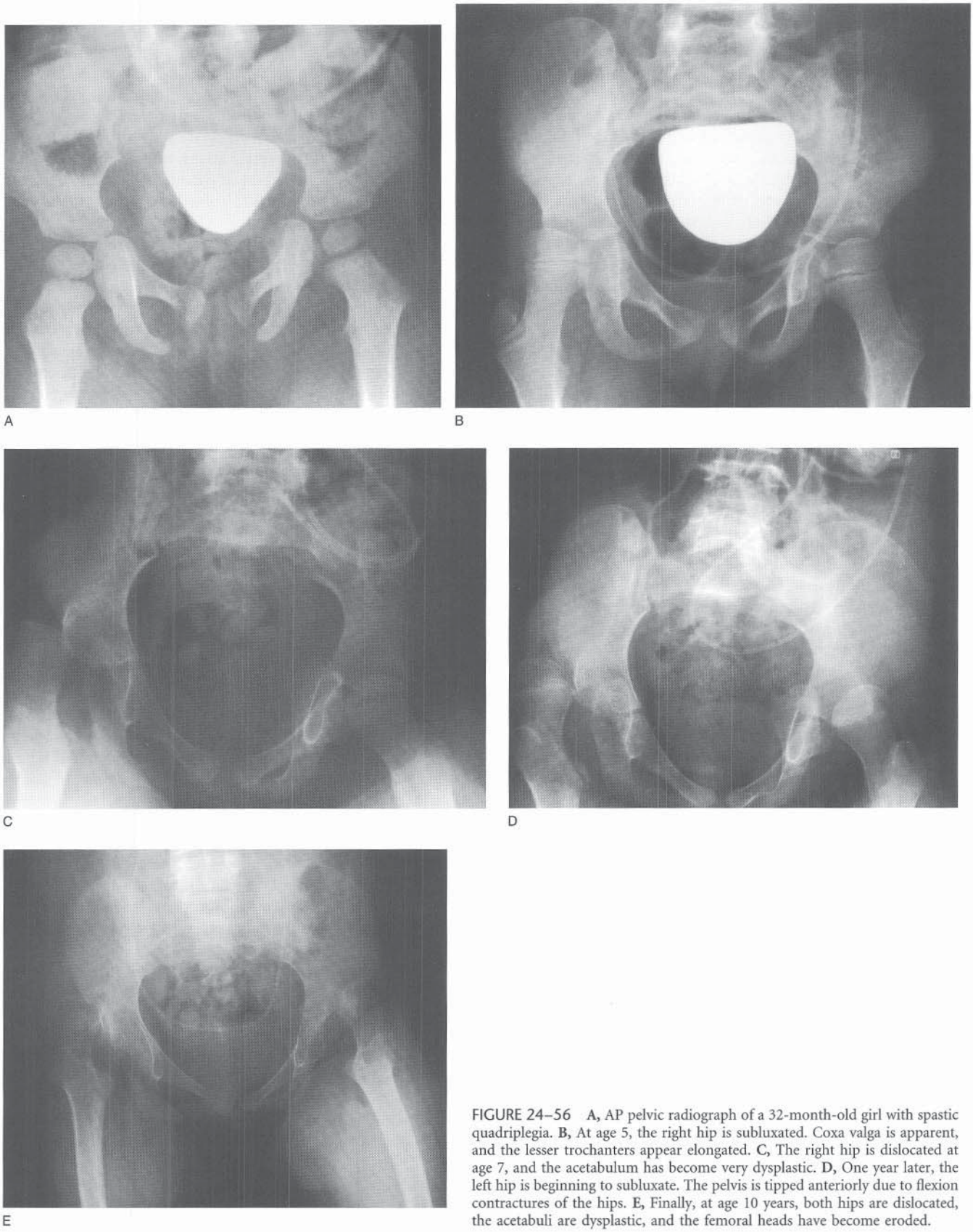


FIGURE 24–56 **A,** AP pelvic radiograph of a 32-month-old girl with spastic quadriplegia. **B,** At age 5, the right hip is subluxated. Coxa valga is apparent, and the lesser trochanters appear elongated. **C,** The right hip is dislocated at age 7, and the acetabulum has become very dysplastic. **D,** One year later, the left hip is beginning to subluxate. The pelvis is tipped anteriorly due to flexion contractures of the hips. **E,** Finally, at age 10 years, both hips are dislocated, the acetabuli are dysplastic, and the femoral heads have become eroded.

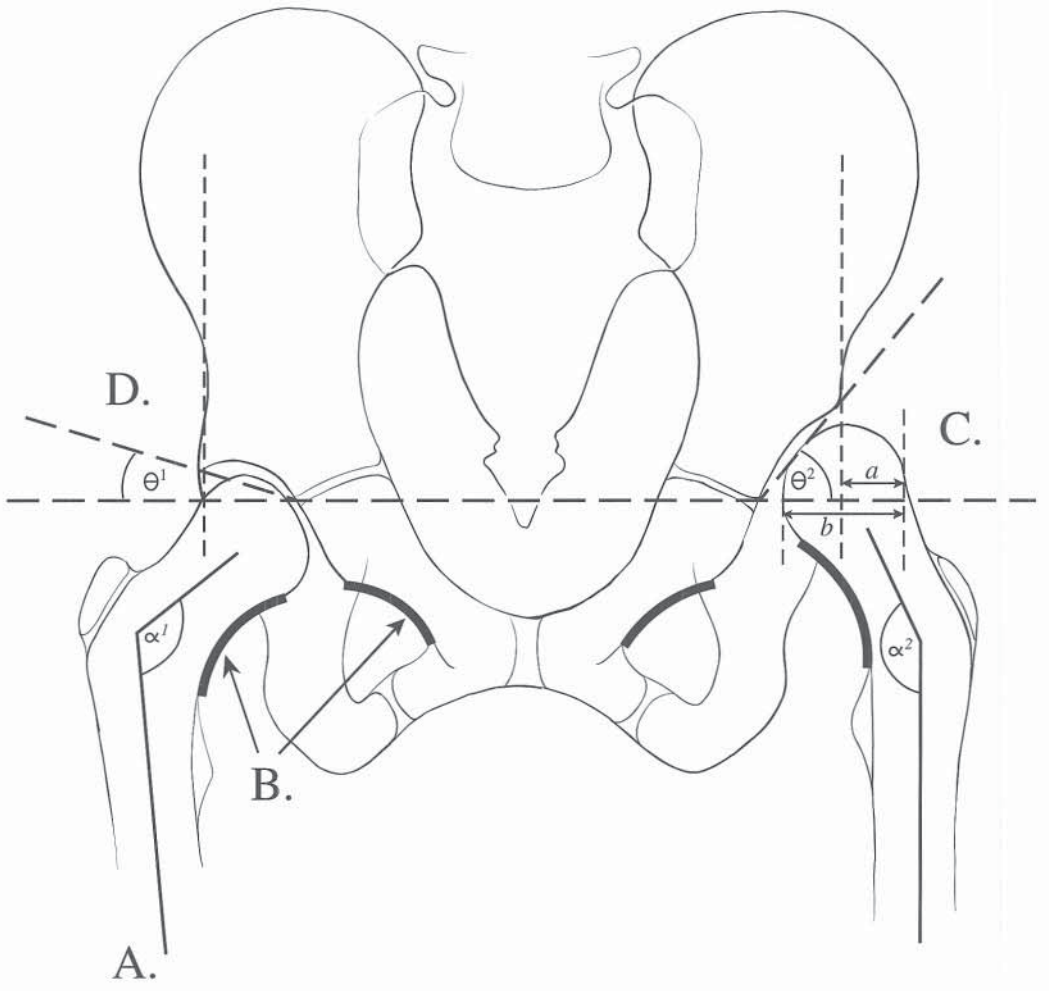


FIGURE 24–57 Radiographic measurements of hip subluxation in cerebral palsy. *A, The neck-shaft angle* (α^1 = normal; α^2 = subluxated hip) increases in patients with spasticity at the hip. *B, Shenton's line.* In the normal hip a line drawn along the inferior femoral neck matches a line drawn along the inferior aspect of the pubic ramus. In the spastic hip the femoral neck line is superior to the pubic line. *C, Reimers's migration index.* In the normal hip the entire femoral head is located medial to the lateral margin of the acetabulum. In the spastic hip, lateral migration is measured as *a/b*. *D, Acetabular index.* As the spastic hip progressively subluxates the acetabulum becomes more shallow and the index (θ^2) increases compared to normal (θ^1).

curve did not correlate with the side of the hip dislocation.[265] The pendulum then swung back again, with Black and Griffin finding that hip subluxation was consistent with the forces related to pelvic obliquity. In 21 patients with pelvic obliquity, unilateral subluxation occurred on the high side in 17 and on the low side in 4 patients.[43] We conclude that there is a strong tendency for unilateral hip subluxation to occur in conjunction with pelvic obliquity, with most dislocations on the high side of the pelvis (Fig. 24–58). Whether the incidence of scoliosis is increased in these patients because of the pelvic obliquity due to hip subluxation or whether the population at greatest risk for scoliosis is the same totally involved group of patients that is at risk for hip dysplasia remains unclear.

Surgical treatment is divided into three phases: (1) soft tissue surgery for the hip at risk, (2) reduction and reconstruction of the subluxated or dislocated hip, and (3) salvage surgery for long-standing painful dislocations.

Soft Tissue Release for Subluxation of the Hip at Risk. The hip at risk is defined as a hip that has significant adduction and flexion contractures but minimal subluxation, with a migration index of less than 30 percent. The patients are 5 years old or younger.[229] Surgical treatment is aimed at preventing dislocation of the hip. Soft tissue release of contractures is indicated when the abduction range is less than 30 degrees and the flexion contracture is more than 45 degrees. The procedure consists of adductor release and iliopsoas lengthening or release.[207] The surgical technique is described in Plate 24–7. Adductor transfer has also been used for the hip at risk.

Many studies have examined the surgical outcome following soft tissue surgery for the hip at risk. In 1975 Sharrard and others compared outcomes in a group of children with cerebral palsy who had undergone adductor tenotomy for abduction of less than 45 degrees with outcomes in a matched control group of patients who did not undergo

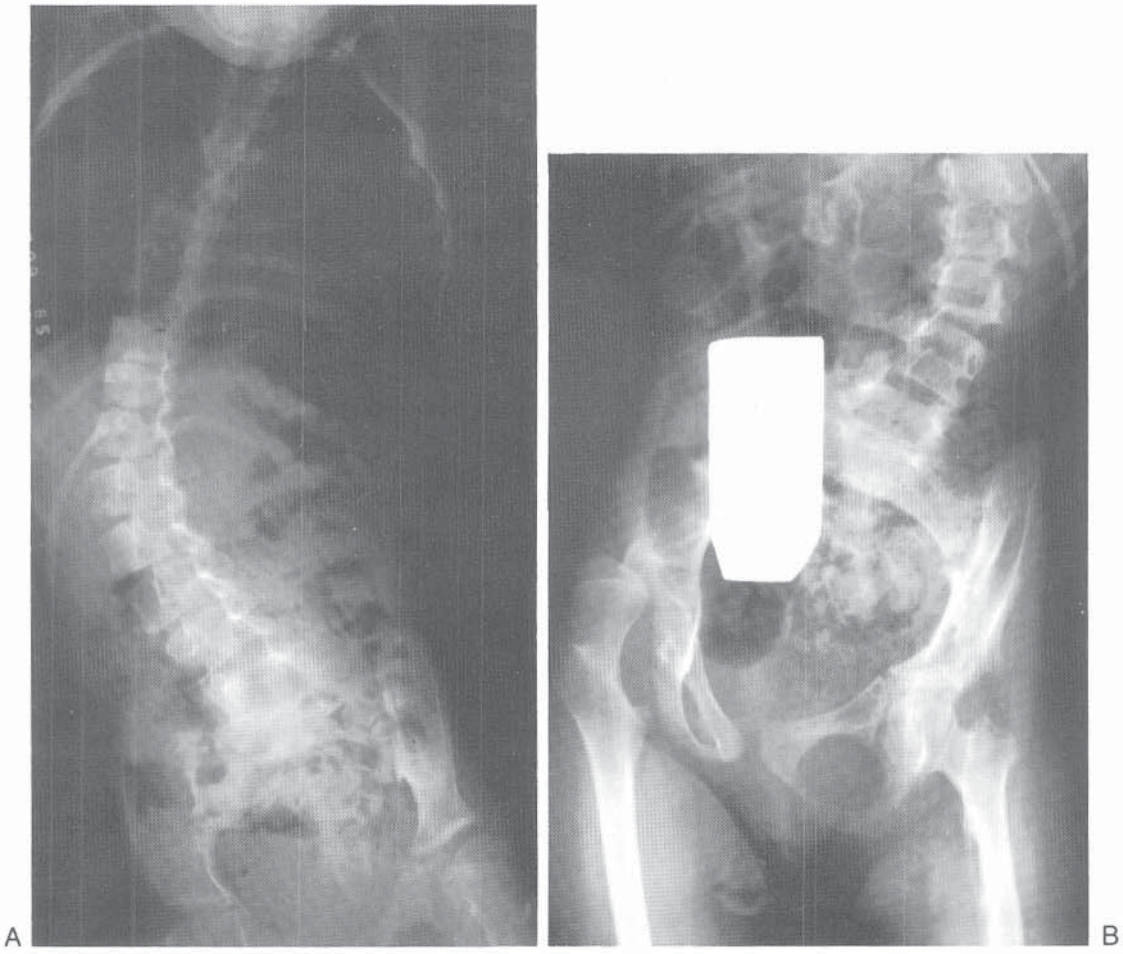


FIGURE 24–58 Paralytic dislocation of the left hip with severe scoliosis. **A,** AP view of the spine showing right dorsolumbar scoliosis (long C curve). Note the left hip subluxation. **B,** AP view of the left hip. Note the progressive subluxation.

surgery. At 4-year follow-up, 13% of hips treated by contracture release had subluxated and there were no dislocations. In the control group, 28 percent had subluxated and 11 percent were dislocated.[425] The average age of the surgical patients at follow-up was only 8.5 years, so it is impossible to know what effect subsequent growth may have on these findings. The early results of adductor surgery were promising. Wheeler and Weinstein published their results following adductor release in 41 hips, and noted that acetabular development and subluxation had improved postoperatively at 3.7 years of follow-up. The center-edge (CE) angle of Weiberg increased from an average of 4 degrees preoperatively to 32 degrees postoperatively. Fourteen of the hips remained subluxated or dislocated following the operation.[488]

Silver and colleagues published the results of adductor release in 86 hips, and found that dislocation was prevented in 80 percent.[431] Hips with a preoperative migration index of greater than 50 degrees had poorer results than hips with milder subluxation. Unilateral adductor release often led to contralateral contractures. They recommended that bilateral release be routine, and that hips that were significantly uncovered preoperatively be monitored closely for dislocation.[431] Cornell and colleagues reported that 83 percent of hips with preoperative migration indices of less than 40 percent that were treated by adductor release remained stable. Of the 77 percent of hips that were uncovered, more than 40 percent remained subluxated or eventually dislocated.[93] In another study, 10 of 16 soft tissue releases done on hips with an average preoperative migration index of 53 percent required further surgery for progression.[34] Clearly, the amount of preoperative subluxation present strongly influences the outcome of soft tissue hip releases.[288]

Kalen and Bleck reported outcomes in 99 patients who had undergone adductor release and compared the results in children who had undergone concomitant iliopsoas recession with those who had not.[229] They found a failure rate of 60 percent in the children who had undergone adductor release alone, compared to a success rate of 72 percent in those who had undergone concomitant iliopsoas recession. Patients who did best were age 5 years or younger, had mild subluxation, and had no pelvic obliquity.[229] Others have reported similar success rates with adductor release and iliopsoas tenotomy in patients no older than 5 years.[154,297,434] Very early release, at age 2 to 3 years, before the hip is dysplastic, has been advocated but is not generally accepted.[321]

A series of iliopsoas transfers as part of the surgical treatment of hip instability in cerebral palsy has been published. Hip stability was achieved in most patients, but there was

loss of the ability to flex the hip and a deterioration in the ability to sit. Iliopsoas transfer is not recommended in this patient population.[150]

Transfer of the adductor tendon origin posteriorly to the ischium has also been done in cases of mild subluxation of the hips. Aronson and colleagues found that nine hips in 20 patients with spastic quadriparesis failed to stabilize following adductor transfer and subsequently required osteotomy.[19] Root and Spero found that twice as many children who underwent adductor release required repeat surgery compared to those who underwent adductor transfer. In their series, however, an equal proportion of children in the two groups went on to have varus osteotomy of the femur for hip dysplasia.[390] Other series failed to show a significant difference in postoperative change in the migration index following adductor tenotomy versus transfer.[376,416] Most worrisome is a report by Scott and colleagues of a 36 percent incidence of postoperative unilateral hip subluxation following bilateral adductor transfer and an 85 percent incidence of pelvic obliquity, which was presumed to be secondary to unilateral detachment of the adductors from the ischium.[417] Based on these various reports, adductor transfer is rarely used currently for the prevention of hip dislocation, and we do not use it at all at our hospital.

Aside from failure to prevent progressive subluxation and dislocation, the complications resulting from adductor surgery and iliopsoas lengthening are few. Infection and hematomas may occur owing to the dead space created by the surgery and the location of the groin incisions.[390] These complications are more frequently seen in adductor transfers than in releases.

Some reports have noted extension-abduction contractures resulting from adduction-flexor releases. The children usually underwent prior tenotomy of the iliopsoas and obturator neurectomy at the time of adductor release. Spasticity in the gluteal muscles and hamstrings, which are not released, drives the development of this new contracture.[56] Extension is particularly disabling in patients who are nonambulatory, as they are unable to sit comfortably in their wheelchairs as a result of loss of hip flexion. Lumbar lordosis is lost, and thoracic kyphosis results. Surgical release of the gluteal muscles from their insertion on the greater trochanter combined with proximal hamstring release can be helpful. The proximal hamstrings are approached through an adductor incision transversely just distal to the groin crease (Plate 24–8). The tendons are identified at their origin on the ischium. The sciatic nerve is located just lateral to the hamstring origin, so it must be protected.[149] Some use a nerve stimulator to be sure that what they perceive as the hamstring tendons are not the sciatic nerve.[288] In severe cases, a sciatic nerve palsy has resulted from stretch following this release, and femoral shortening may be necessary.[468] This deformity can be prevented by performing a proximal hamstring release at the time of adductor and hip flexor releases in patients with the tendency to thrust their hips forward in their wheelchairs and when significant hamstring tightness is present.[149,434]

Another complication seen following bilateral adductor releases is a unilateral abduction contracture due to a preexisting windblown hip deformity.[4] On preoperative clinical examination, the surgeon should look carefully for abduction contracture of the contralateral hip when contemplating bilateral release for unilateral subluxation. The "pseudo-Galeazzi sign" has been described as asymmetry in the apparent lengths of the femora when the hips and knees are flexed in the supine patient in the face of reduced hips. This sign can occur in the setting of a mild windblown hip deformity, in which one hip is adducted and the other is abducted. Bilateral adductor release should not be performed in these cases, as the abduction may worsen postoperatively and interfere with positioning.[257,317]

Current indications for adductor and iliopsoas soft tissue release in patients with hips at risk for dislocation include adduction contractures in a young child, preferably no older than 5 years, with limited abduction of 30 degrees or less and a migration index of not more than 30 percent. Bilateral releases should be performed whenever bilateral contracture exists, but also in the absence of a contralateral abduction contracture. Proximal hamstring releases are added to the procedure in sitting patients who vault from their chairs because of their startle reflex. As up to 46 percent of young children will improve even in the presence of significant subluxation, patients less than 4 years old, especially those who are frail and have significant medical problems, may be treated with soft tissue release alone.[288] Some of these children will probably need further surgery later.

Femoral Osteotomy. In the setting of more significant hip subluxation, soft tissue release is inadequate to maintain a stable reduced hip. The most frequently performed procedure in this setting is a femoral varus derotation osteotomy (VDRO). An osteotomy is performed at the intertrochanteric level, usually accompanied by a closing wedge taken medially and/or femoral shortening, and the femur is fixed in an increased amount of varus with internal fixation (Fig. 24–59). Implants vary, but the two most frequently used are the hip screw and side plate, and the 90-degree blade plate.[38] The desired neck-shaft angle following varus osteotomy for hip instability is 90 to 100 degrees of varus.[148] Release of soft tissue contractures must also be done to balance the forces across the hip. Adductor tenotomy and iliopsoas lengthenings are performed. Usually the patient is then immobilized in a spica cast.

Hoffer and associates in 1985 published the results of femoral varus osteotomies after an average follow-up of 11 years.[210] Osteotomies were performed for subluxation in 18 hips and for dislocation in three. The results in subluxated hips were good, with an improvement in the CE angle to +17 degrees. Four hips remained subluxated, all of which had been more than two-thirds uncovered preoperatively. Three patients complained of pain at follow-up. Of note, of the three dislocated hips, one redislocated following surgery, and another developed avascular necrosis and became painful and stiff. Hoffer and associates concluded that VDRO is a good procedure for hip subluxation but is inadequate to treat the dislocated hip in cerebral palsy. Tylkowski and colleagues found improved coverage in 16 of 18 VDROs done for hip instability, including the successful reduction of two hips that were dislocated.[480] Follow-up in this study averaged only 3 years, however.

Brunner and Baumann also found deterioration in the coverage of the femoral head when a varus femoral osteotomy was performed for the treatment of hip dislocation.[66] After a mean of 15 years of follow-up, the CE angle in these hips had

Text continued on page 1196

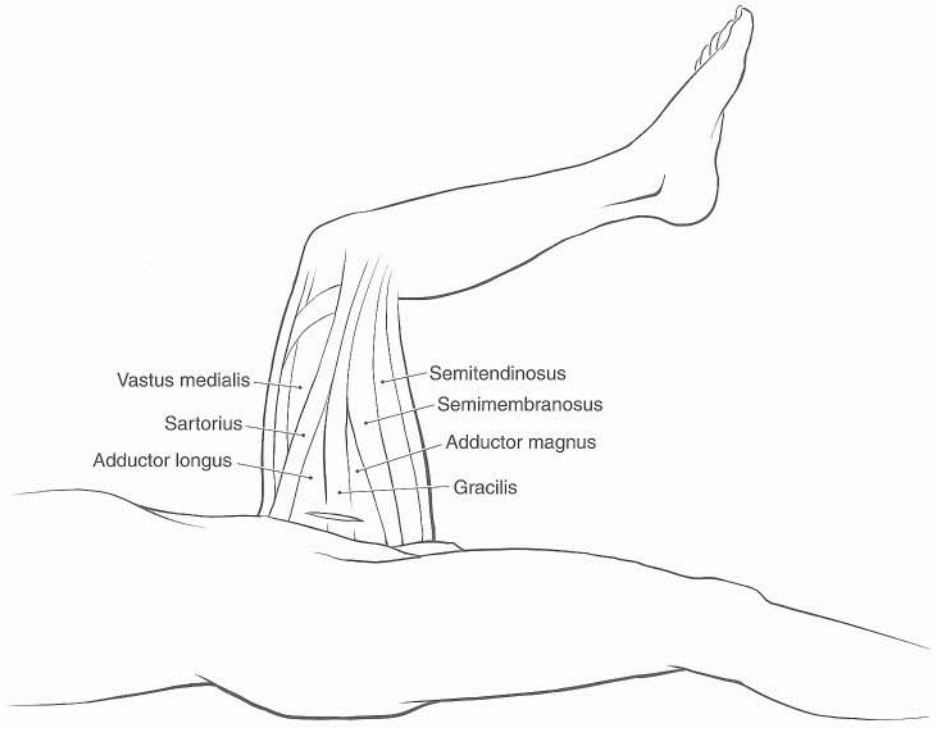
Technique for Proximal Hamstring Release

A, A transverse incision is made just distal to the groin crease and centered over the gracilis origin, just posterior to the adductor longus. When a proximal hamstring release is performed in conjunction with an adductor release, the surgeon simply extends the adductor incision more posteriorly.

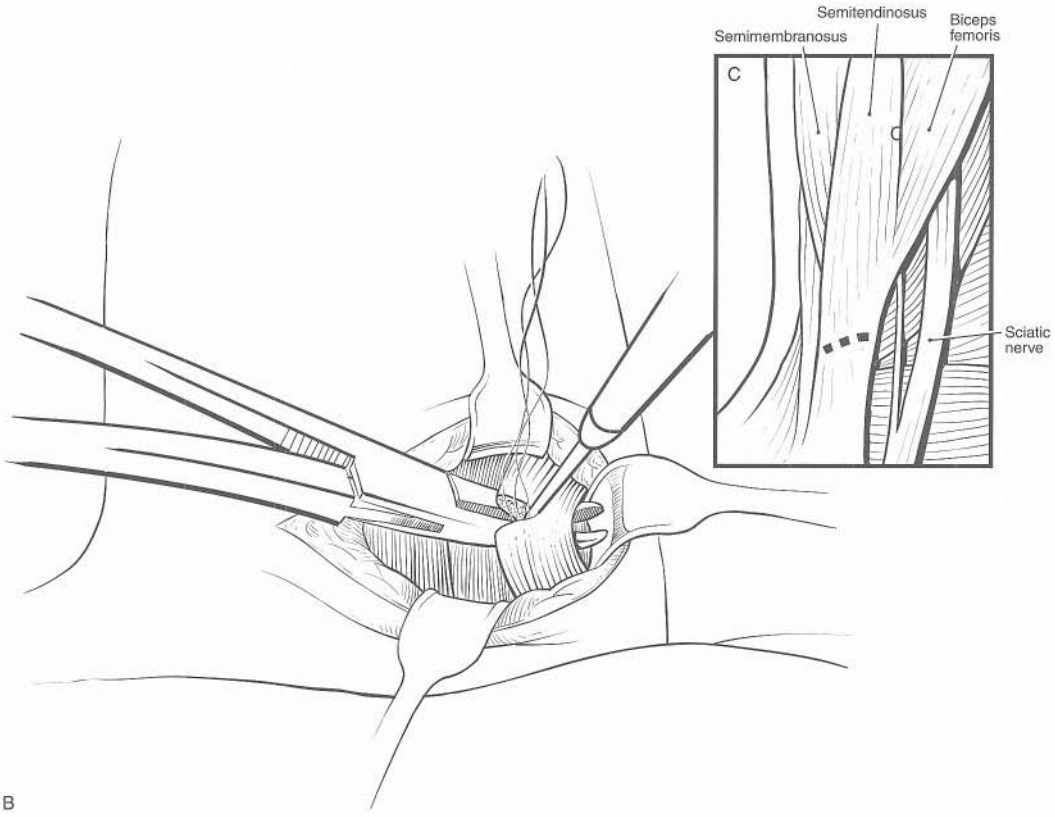
B, The gracilis is divided close to its origin using electrocautery.

C, The semimembranosus, semitendinosus, and biceps femoris are found posterior to the gracilis. Their common origin from the ischium should be verified by direct vision. The sciatic nerve lies just posterolaterally, so it is necessary to see the hamstring origins. A nerve stimulator can be used to make certain the hamstrings are not the sciatic nerve. The semimembranosus, semitendinosus, and biceps are then released from the ischium using electrocautery.

PLATE 24–8. Technique for Proximal Hamstring Release



A

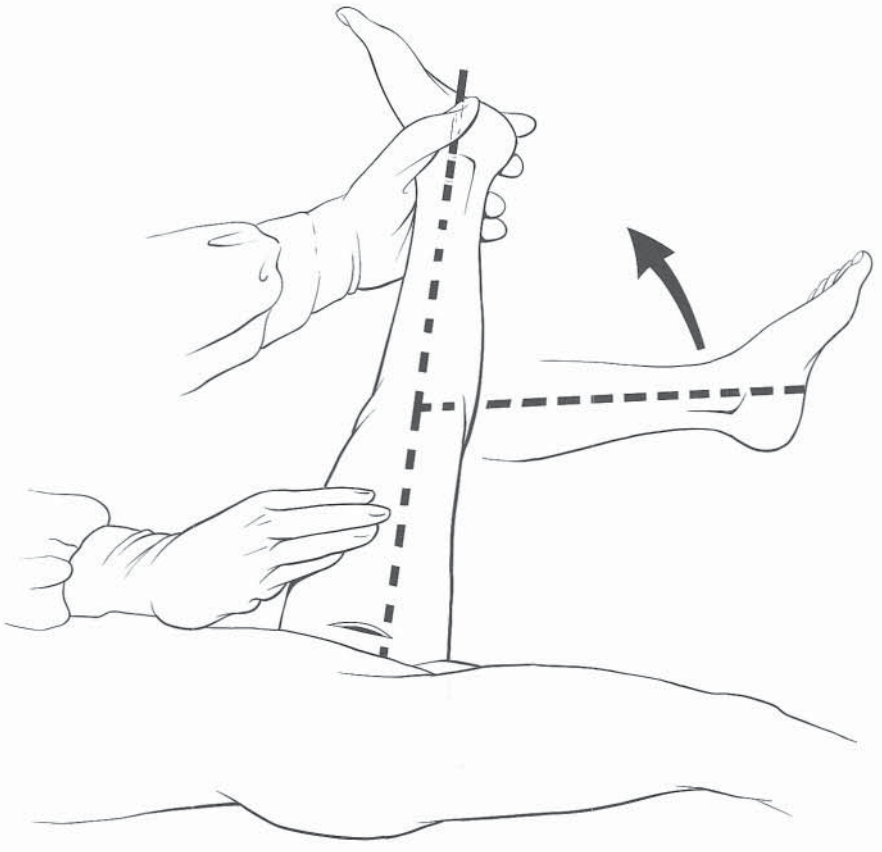


B

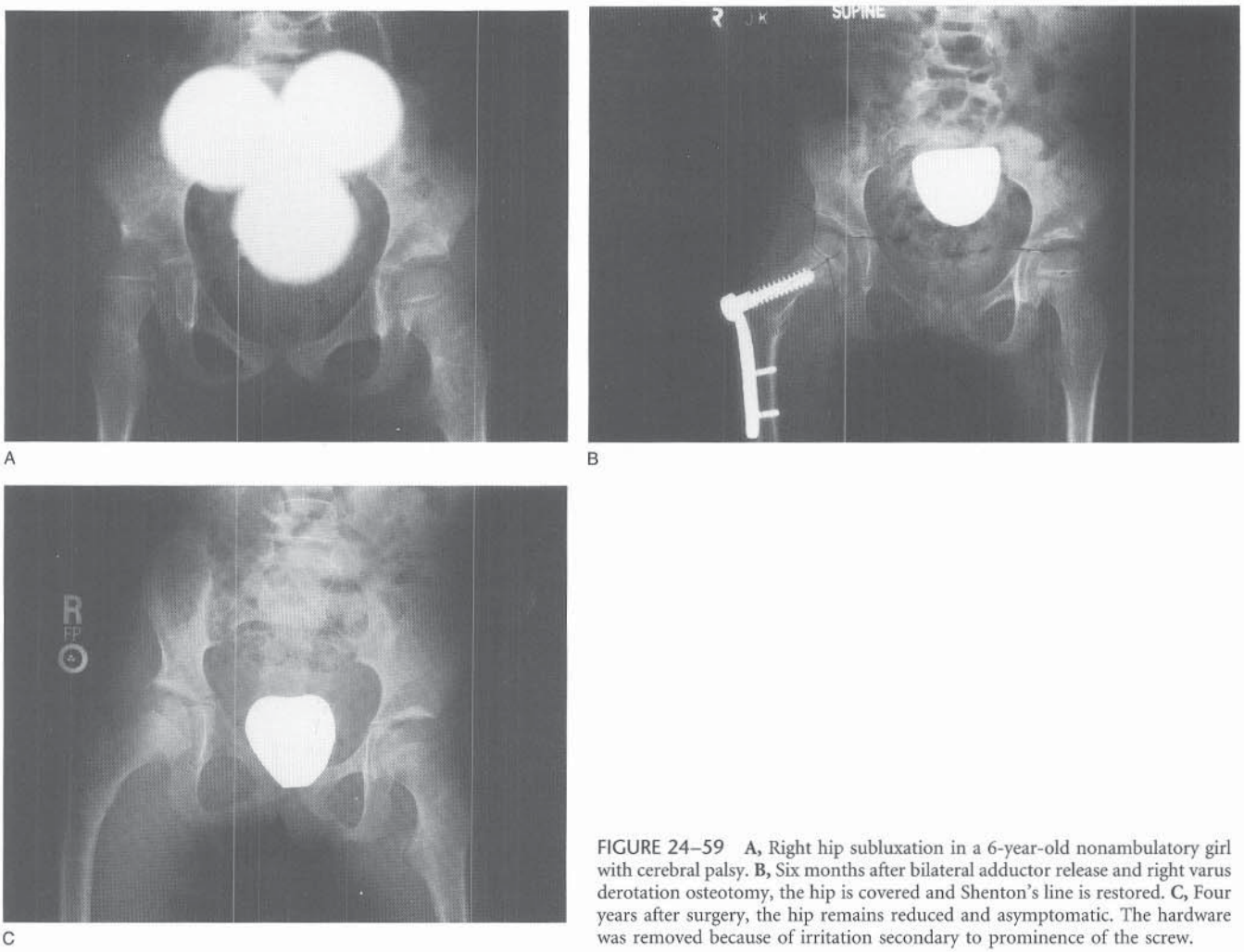
Technique for Proximal Hamstring Release *Continued*

D, The knee is then extended with the hips flexed. Postoperatively, the patient can be placed in straight-knee long-leg casts for 3 weeks. Sitting in the wheelchair in these casts can maintain correction of the hip extension contracture.

PLATE 24–8. Technique for Proximal Hamstring Release



D



A B C

FIGURE 24–59 **A,** Right hip subluxation in a 6-year-old nonambulatory girl with cerebral palsy. **B,** Six months after bilateral adductor release and right varus derotation osteotomy, the hip is covered and Shenton's line is restored. **C,** Four years after surgery, the hip remains reduced and asymptomatic. The hardware was removed because of irritation secondary to prominence of the screw.

returned nearly to preoperative values, and most patients needed further pelvic surgery to reestablish coverage. Yet the results in subluxated hips were uniformly good, with improvement in the CE angle from an average of 8 degrees preoperatively to 23 degrees at follow-up. Just as in soft tissue release, superior coverage is found when the VDRO is performed in hips with better coverage to begin with.[27,67]

Herndon and associates reported on 48 varus osteotomies performed for hip subluxation or dislocation in children with cerebral palsy. What is novel about this study is that a medial open reduction was also performed on 20 hips. The postoperative result closely correlated with the amount of preoperative coverage, the preoperative CE angle, and the acetabular index. Asymptomatic vascular changes within the femoral head subsequently developed in five of the 20 hips treated with open reduction. The authors concluded that the procedure is useful in subluxated hips, but if instability or poor coverage is present at the end of the case, pelvic osteotomy should be added.[204] Other studies comparing outcomes in hips treated by VDRO with hips treated by VDRO and pelvic osteotomy found superior results with addition of the pelvic osteotomy, and the authors have recommended against performing VDRO alone.[25] Song and Carroll advocate pelvic osteotomy if the preoperative migration index is greater than 70 percent.[436]

Remodeling does occur following femoral VDRO. Brunner's group documented remodeling of the proximal femur back into valgus with growth, which was most remarkable in children who had been operated on before age 4 years.[66,210] No significant remodeling can be expected in patients operated on at age 8 or older.[210]

Complications from the femoral VDRO other than recurrent dysplasia are loss of fixation and fracture. In a series of VDROs stabilized with the angled blade plate without casting, two patients lost fixation and five sustained proximal femoral fractures. Care to perform the osteotomy an adequate distance away from the insertion site for the blade can reduce the incidence of proximal femoral fractures.[38] We uniformly use a spica cast following VDRO in patients with hip instability due to cerebral palsy because we believe that weakness and stiffness will improve with time following physical therapy and are preferable to the potential loss of fixation in osteopenic bone. Delayed union and nonunion are rare but do occur.[436] Heterotopic ossification can result from any hip reconstructive procedure, including the varus osteotomy.

Combined Femoral Varus and Pelvic Osteotomies. In cases in which a femoral VDRO does not provide adequate coverage or stability to the hip, pelvic osteotomy should be performed.[35]

Among the various possible osteotomies are redirectional osteotomies, such as the Steel and Salter procedures; osteotomies that reshape the acetabulum, such as the Pemberton and Dega osteotomies; and salvage osteotomies that increase the area of the acetabulum with nonarticular cartilage, such as the Chiari osteotomy and the shelf augmentation procedure. Each of the osteotomies has its advocates, but it is important to understand the three-dimensional anatomy of the acetabulum and femoral head before choosing a particular osteotomy.

Three-dimensional CT reconstructions of dysplastic hips in patients with cerebral palsy have helped define the nature of the instability.[233] Abel and colleagues reviewed the three-dimensional CT scans of 31 hips in a mix of ambulatory and nonambulatory patients.[5] They found that subluxation and dislocation occur in a posterior direction in cerebral palsy. In patients who were able to walk, the acetabular volume was better, but there was significantly more anteversion in the proximal femur. In nonambulatory patients, global acetabular deficiency was seen, with lack of anterior and posterior wall development and decreased acetabular volume relative to the size of the femoral heads (Fig. 24–60). Based on their study results, the authors recommend against performing redirectional osteotomies that increase anterior coverage, such as the Salter osteotomy, because of the possibilities of decreasing the already deficient posterior coverage.[5] Posterior insufficiency was also found with CT in a study performed by Buckley and colleagues.[71] Another study of three-dimensional CT scans, reported by Brunner and colleagues, had differing conclusions. The authors did not think that the acetabulum was globally deficient but felt there was a rut driven by the femoral head through the acetabular roof as the hip subluxated, and that anterior and posterior wall coverage were adequate.[68] From these different studies, it appears that there may be varying patterns of acetabular deficiency in cerebral palsy, with many but not all patients having posterior insufficiency.

The two pelvic procedures with the longest historical use in patients with cerebral palsy are the Chiari osteotomy and the shelf acetabular augmentation procedure. The Chiari osteotomy is performed by making a horizontal osteotomy from the sciatic notch to a point just at the superolateral margin of the acetabulum. The osteotomy is displaced medially so that the hip capsule lies over the lateral bony surface of the ilium, which over time undergoes metaplasia to form fibrocartilage (Fig. 24–61).[84] A more detailed description of the surgical procedure can be found in Chapter 15, Developmental Dysplasia of the Hip.

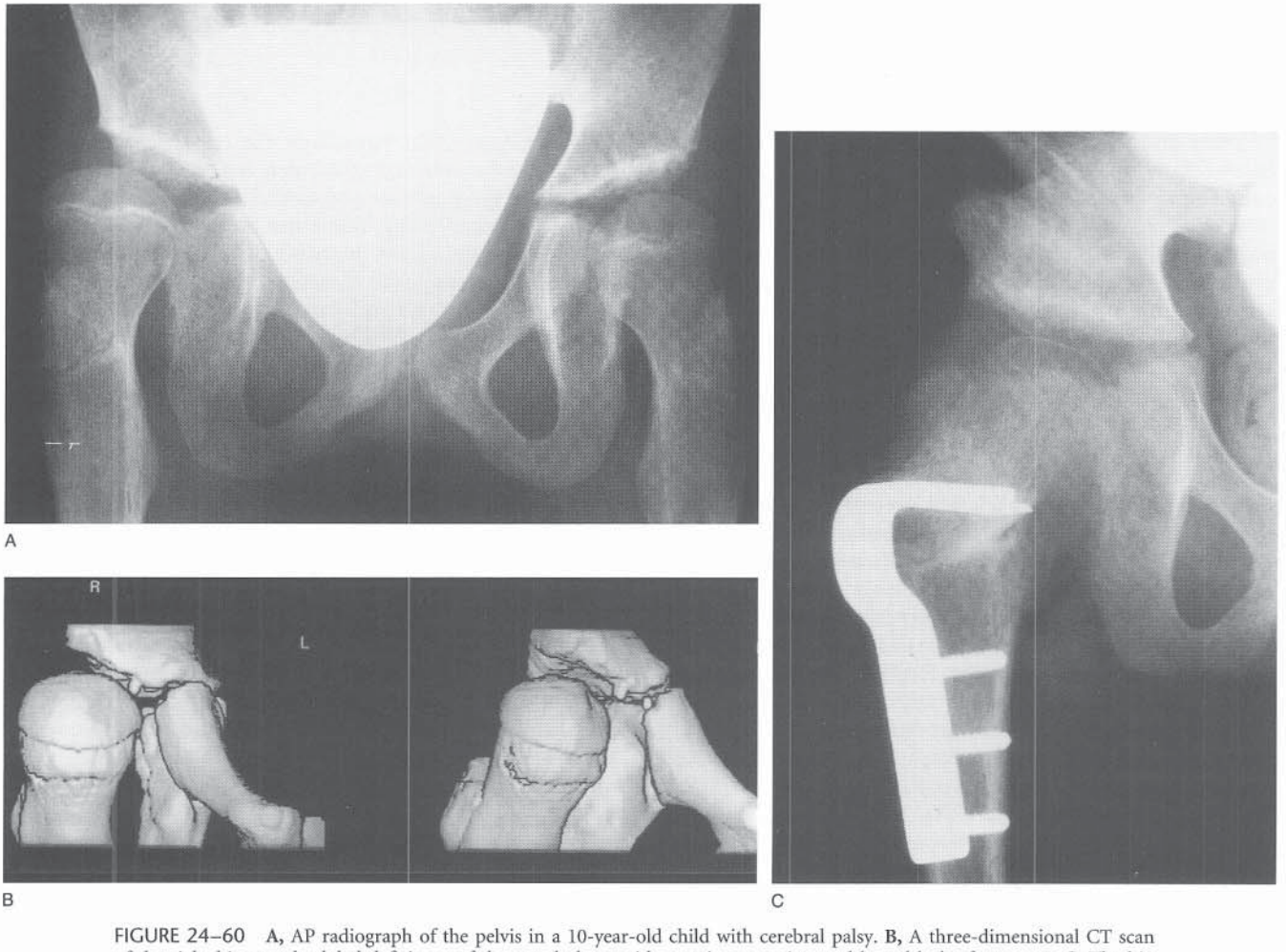


FIGURE 24–60 **A,** AP radiograph of the pelvis in a 10-year-old child with cerebral palsy. **B,** A three-dimensional CT scan of the right hip reveals global deficiency of the acetabulum, with anterior, superior, and lateral lack of coverage. **C,** The hip was reconstructed by muscle release, femoral varus derotation osteotomy with blade-plate fixation, and a Dega pelvic osteotomy.

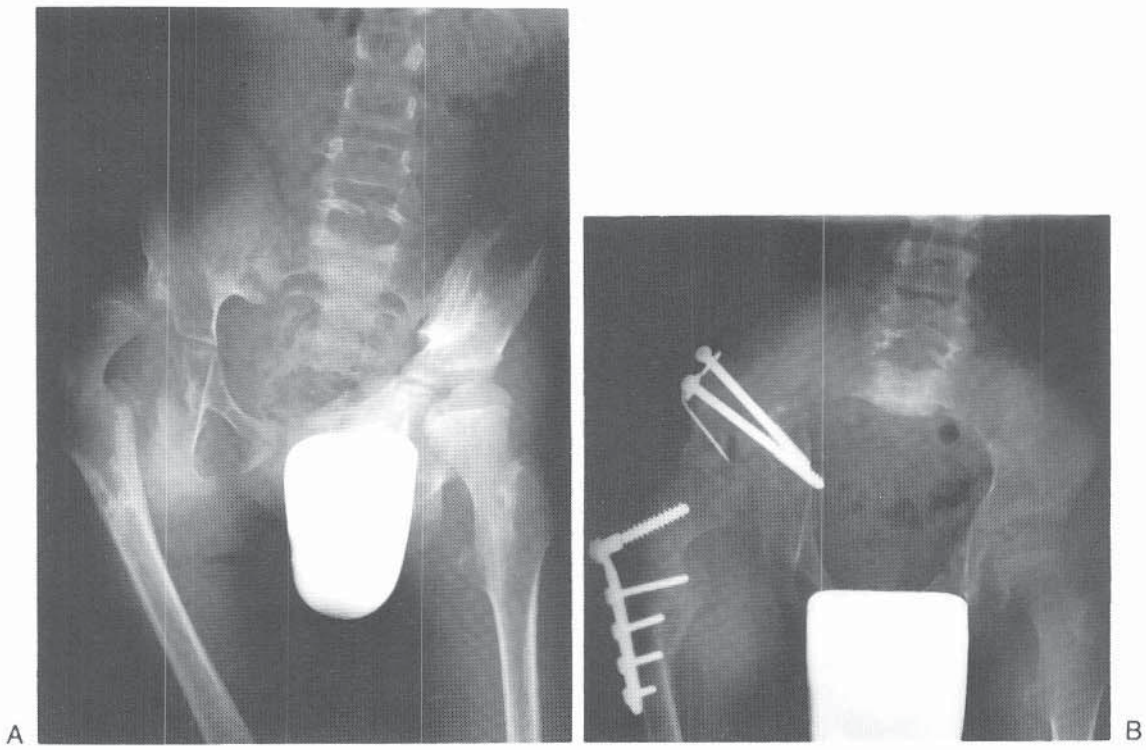


FIGURE 24–61 Paralytic dislocation of the right hip in severe spastic cerebral palsy. **A,** Preoperative radiograph. **B,** Postoperative radiograph following open reduction with femoral shortening, capsular plication, and Chiari pelvic osteotomy and shelf acetabuloplasty.

Dietz and Knutson studied 24 Chiari osteotomies at a mean follow-up of 7 years and found that a pain-free joint could be obtained in 79 percent of hips. Seven of the 24 hips still had migration indices greater than 30 degrees, indicating that subluxation persisted despite surgery. A simultaneous femoral osteotomy was not performed in this series, but the authors state that based on their results, it is indicated in patients with severe subluxation.[130] Osterkamp and colleagues published the results of Chiari osteotomy performed in conjunction with varus femoral osteotomy in nine patients and without femoral procedures in three patients.[325] Pain relief was obtained in five of six patients who complained of pain preoperatively. Two hips redislocated. Osterkamp and colleagues felt that the osteotomy was simple to perform and led to satisfactory results.[326] In a study reported by Pope and associates, five of seven hips remained reduced following VDRO and a Chiari osteotomy. Their indications for a Chiari osteotomy were the presence of severe subluxation or the presence of moderate incongruity on arthrography following the femoral osteotomy.[361] The series with the greatest success rate was that of Atar and colleagues, in which 14 of 15 hips remained stable following a Chiari osteotomy combined with a varus osteotomy and muscle release at 3-year follow-up (Plate 24–9).[23]

A second pelvic procedure that has been widely used is the shelf acetabular augmentation as described by Staheli in 1981. A notch is made in the outer wall of the ilium just at the margin of the acetabulum, and cancellous and corticocancellous strips of iliac crest are wedged into this notch above the hip capsule to increase the area of load bearing and therefore improve the stability of the hip (see Chapter 15, Developmental Dysplasia of the Hip).[442] The graft can be placed wherever acetabular deficiency is present, thereby improving the posterior and lateral deficiencies most common in cerebral palsy. Since the acetabulum is not redirected, coverage is not increased in one direction at the expense of the opposite direction. In fact, coverage that is increased beyond normal can be achieved, and according to Staheli should be.[443] The procedure is indicated in a hip in which spherical congruency cannot be achieved. In hips that remain congruent, a redirectional osteotomy is preferable to maintain coverage by articular cartilage. The shelf procedure is also helpful in hips in which there is global acetabular deficiency and a small articular surface.

Studies of the results of shelf procedures in cerebral palsy are encouraging, with hip stability obtained in 83 to 95 percent of patients.[267,436,500] Previous problems with resorption of the lateral graft are lessened with decortication of the lateral surface of the ilium and with radiographic localization of the inferior lip of the acetabulum, where the shelf needs to be placed. If it is placed too high, the acetabulum will appear to have a step-off and the graft will resorb. Since the shelf is built at the very margin of the acetabulum, it is possible to disturb further lateral growth of the acetabulum, so the shelf procedure should not be performed in very young patients.

A third pelvic osteotomy that is of particular benefit in hip dysplasia secondary to cerebral palsy is the Dega osteotomy (Fig. 24–62, Plate 24–10). This osteotomy, as described by Mubarak, extends through the outer table of the ilium from the anterior-inferior iliac spine to the sciatic notch. A bicortical osteotomy is performed only at the anterior-infe-

rior iliac spine and, using a kerasin rongeur, at the sciatic notch. The inner table of the ilium is not cut. The lateral osteotomy made through the outer table is extended with curved osteotomes to the triradiate cartilage under fluoroscopic guidance. The osteotomy is then pried down laterally and posteriorly with osteotomes, hinging on the triradiate cartilage and leaving the inner table of the ilium intact. Wedges of bone graft prop the osteotomy open, and the direction of desired coverage is addressed by where one places the bone graft. The sponginess of the triradiate cartilage closes the osteotomy around the bony wedges, so that fixation with pins is usually not necessary. A prerequisite for the Dega osteotomy is an open triradiate cartilage.[303]

Mubarak and colleagues reported their experience with the Dega osteotomy, which was performed in conjunction with adductor, iliopsoas, and proximal hamstring release and a shortening femoral VDRO. Seventeen of 18 hips remained stable at almost 7 years' follow-up, although AVN occurred in two hips. The authors commented that the Dega osteotomy allowed excellent correction of the superior and lateral deficiency seen preoperatively.[303]

Miller and colleagues used a similar osteotomy, again combined with a shortening femoral VDRO and aggressive muscle release. Only two hips redislocated and two hips remained subluxated out of 70 hips at follow-up. Their osteotomy differed from the Dega osteotomy in that it did not extend into the sciatic notch but was directed inferiorly to the triradiate at the posterior aspect.[290] Their indications for this combined procedure were (1) failure of a soft tissue release in a child less than 8 years old, (2) subluxation, defined as a migration index greater than 40 percent in children older than 8 years, (3) a recent hip dislocation (within 2 years), and (4) a painful subluxated or dislocated hip without significant femoral head deformity. They successfully did not use a spica cast postoperatively, but we do continue to use a cast in this group of patients, both for comfort and to reduce spasticity and maintain the position of the pelvic osteotomy during healing.

The benefit of the Chiari, shelf augmentation, and Dega osteotomies is that they provide coverage posteriorly and laterally and thereby improve the migration percentage and CE angle. Drummond warns against using the Salter osteotomy or Pemberton osteotomy in patients with cerebral palsy, as these osteotomies were designed to move coverage more anteriorly and laterally, without improving posterior coverage.[136,339] Salter himself indicated that his innominate osteotomy was not to be used in spastic dislocations.[405] Yet some surgeons have achieved good results with the Salter osteotomy[388] and the Pemberton osteotomy,[427] despite their theoretical disadvantages.[64,65,361] Root and colleagues advocated using the Salter osteotomy in children less than 12 years old and the Chiari osteotomy in older children.[388] We do not use the Salter or the Pemberton osteotomy in children with cerebral palsy and cannot explain these reports of satisfactory outcomes.

Patients with cerebral palsy who undergo surgical hip reconstruction with osteotomies are at significant risk for complications. The risk of complication following an osteotomy is significantly greater in the nonambulatory population, and in a recent series a worrisome 69 percent of patients with tracheostomy or gastrostomy sustained complications.[445] In every study some patients develop postoperative fractures, including pathologic fractures of the supracondylar femur during mobilization following cast removal and fractures in the area of the osteotomy, at times resulting in loss of fixation. It was this risk of fracture that stimulated Miller's group to try reconstruction without casting. We too have seen problems with fractures in this group of patients, and attribute the fractures not only to immobilization but also to preexisting osteopenia. Great care in the period of time following cast removal is needed, and attempts to increase passive range of motion should be pursued with extreme caution.

Patients with hip dislocations due to cerebral palsy are also commonly malnourished. Malnutrition increases the risk of postoperative pulmonary complications and the development of decubitus sores from the cast. The "felt suit" approach to spica casting is warranted: thick, soft felt is applied to all potential sites of breakdown and bony prominences and held in place with cast padding. These casts must be well padded.

Last, there is a debatable risk in a patient who undergoes surgical reconstruction of a unilateral dysplastic hip for subsequent instability in the contralateral hip (Fig. 24–63).[374,408,431] Studies have shown that the risk of contralateral subluxation is greatest if soft tissue release is performed prior to age 9 years in a nonambulatory patient.[78] Recent research has found minimal risk to the contralateral concentrically reduced hip following unilateral varus femoral osteotomy.[78,176]

Many patients who undergo femoral osteotomy develop symptoms due to prominence of the hardware. Bursae form over the lateral prominence that may cause pain while sitting. Hardware removal is frequently helpful but does carry a small risk of fracture in the postoperative period. Physical therapy should be discontinued for 6 weeks following removal of internal fixation to allow for bony healing.[39]

Recovery time is prolonged following combined soft tissue and bony reconstruction of the hip in children with cerebral palsy. The time interval following surgery until the patient regains the preoperative level of function and experiences pain relief is usually 6 months. Some have found that a rehabilitation period of 8 to 14 months occurs following hip reconstruction.[223] Parents should be forewarned that surgical improvement will not be seen for some time, and that prolonged aggressive postoperative care will be needed.

SURGICAL TREATMENT OF THE NONRECONSTRUCTABLE HIP. Surgical reduction of the dislocated hip in patients with cerebral palsy should be limited to those children who do not have significant bony changes in the femoral head. Pressure from the hip capsule, abductor muscles, and ligamentum can result in erosion and loss of sphericity of the femoral head, with a pointed superior aspect. Reduction of the hip will not lead to pain relief but may well exacerbate hip and groin pain. These patients are usually older, with most in adolescence. The indications for surgery in this group of patients include hip pain, inability to sit in a modified wheelchair, and difficulty with perineal hygiene due to contractures. There are four surgical options: proximal femoral resection, valgus osteotomy of the proximal femur, hip arthrodesis, and total hip arthroplasty.

The decision to abort reduction of the hip can be a difficult one to make. In the presence of arthritic changes

Technique for Shelf Acetabular Augmentation

A, The patient is positioned supine with a bump beneath the affected hip. Through an anterior approach, the outer table of the ilium is exposed down to the hip capsule. The rectus femoris is detached and tagged. After verifying the position with fluoroscopy, the surgeon uses a drill to outline the shelf just superior to the acetabular rim along the lateral aspect of the hip.

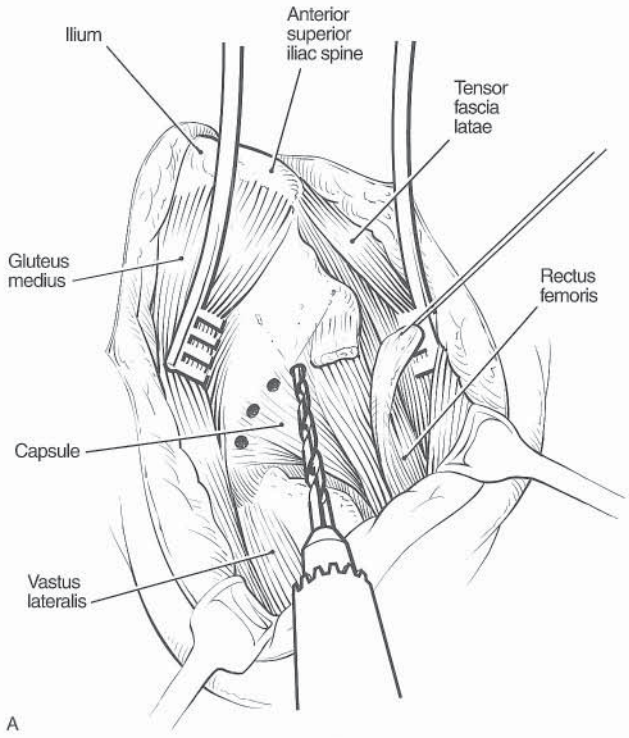
B, The drill is inserted approximately 1 cm *just above* the capsule.

C, A rongeur is used to connect the holes to create a trough for bone graft. Strips of corticocancellous and cancellous graft are obtained from the iliac wing. The graft is placed into the trough and over the hip capsule to form an awning covering the femoral head.

D, The strips are placed at 90-degree angles in layers, and morsellized bone graft is extended up the iliac wing.

E, The rectus femoris is repaired over the shelf and a spica cast applied.

PLATE 24–9. Technique for Shelf Acetabular Augmentation



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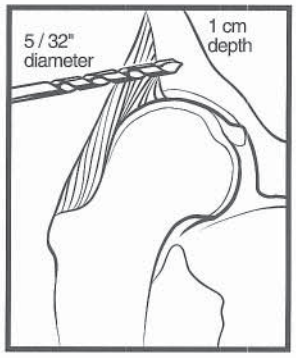

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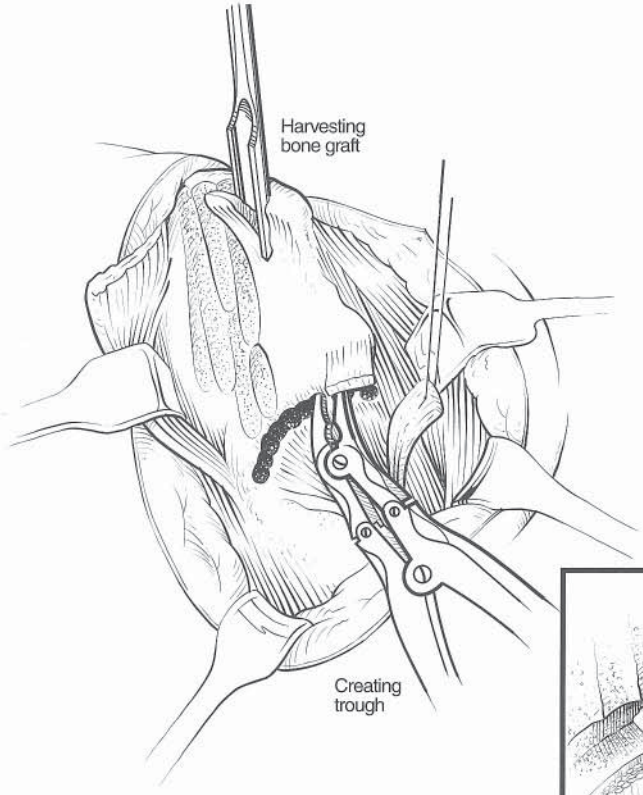

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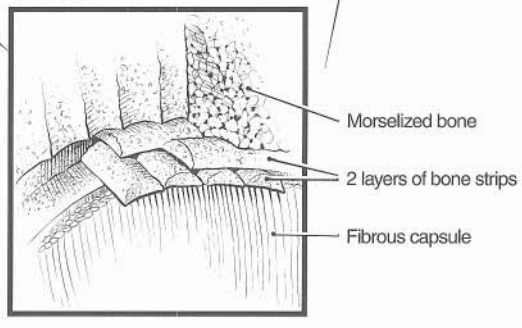

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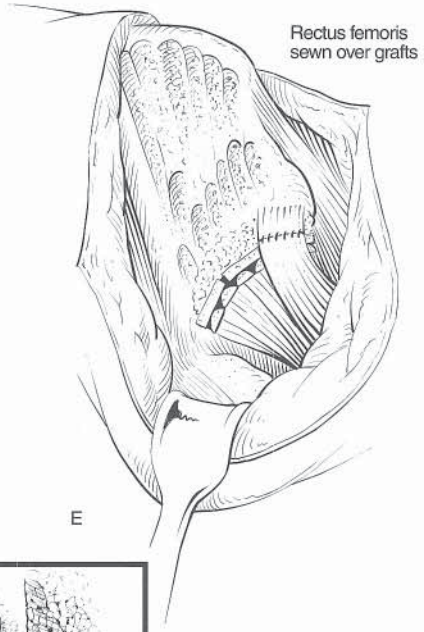

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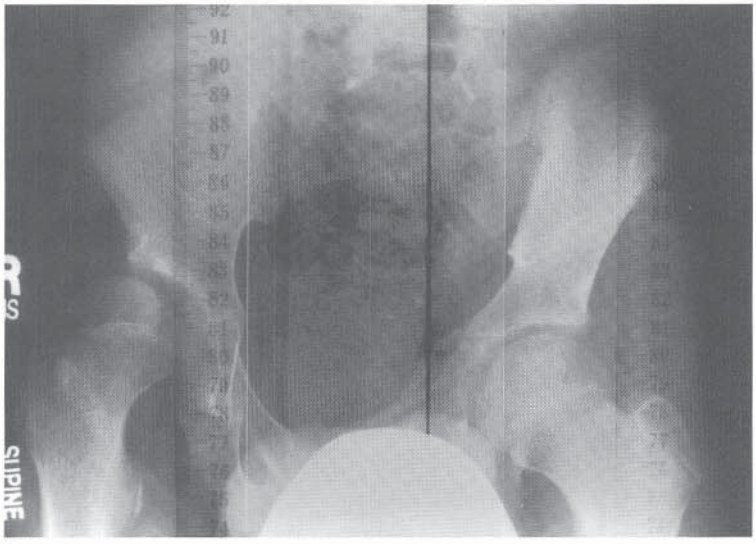

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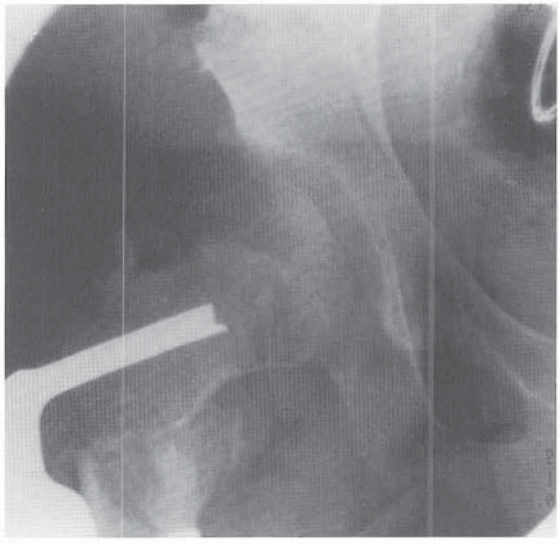

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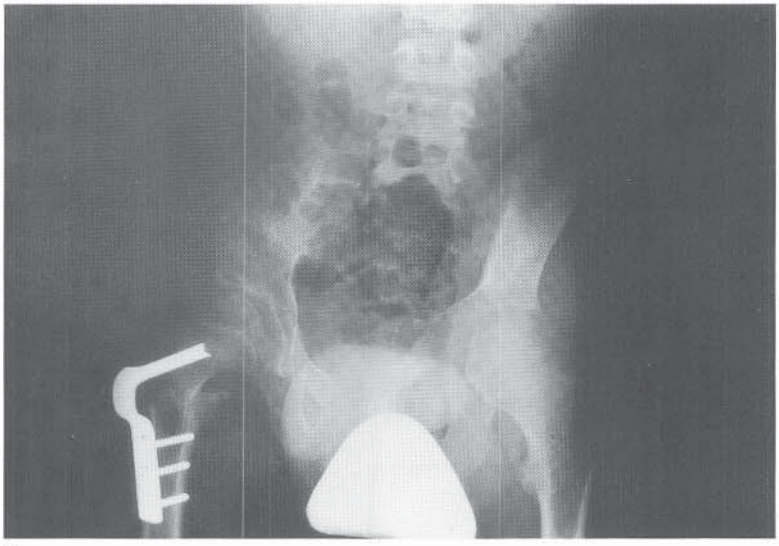


C

FIGURE 24–62 **A,** Right hip subluxation was noted on a scanogram taken for apparent leg length inequality in a 14-year-old ambulatory boy with spastic hemiplegia. **B,** Adductor release, femoral varus osteotomy, a Dega pelvis osteotomy, and an ipsilateral distal femoral epiphysiodesis were performed. **C,** At 3-month follow-up the hip was well covered and the osteotomies had healed. With aggressive physical therapy, the patient remained able to walk.

and severe erosion of the femoral head, it is clear that the hip cannot be reconstructed. Bleck has advocated opening the capsule prior to reduction in questionable cases, and proceeding with proximal femoral resection if the articular cartilage appears degenerated.[48]

Proximal Femoral Resection-Interposition Arthroplasty. Proximal femoral resection was popularized for the treatment of painful dislocations of the hip in nonambulatory patients by Castle and Schneider in 1978.[82] The surgical technique is as follows (Fig. 24–64). The proximal femur is approached laterally. An extraperiosteal dissection of the proximal femur is carried out. The abductors are sharply detached from the greater trochanter. An osteotomy is made across the proximal femur at a level 3 cm distal to the lesser trochanter. The iliopsoas tendon is transected. The hip capsule is detached from the proximal femur and is sewn shut following removal of the proximal femur, so as to cover the acetabulum. Then the quadriceps muscle is sewn over the end of the femoral shaft. The abductors are interposed between the acetabulum, which is covered with the capsule, and the femur, which is covered by the quadriceps. The patient is then placed in traction for a period of 3 to 6 weeks while the soft tissue interposition heals.

Technical caveats to be appreciated here include a more distal level of resection than that described in the Girdlestone procedure.[172] McCarthy and associates have advocated resection no more proximal than the level of the inferior aspect of the ipsilateral ischial ramus, or 3 cm distal to the lesser trochanter.[279] Resection at a more proximal level can lead to increased pain following surgery as the femoral shaft migrates proximally and abuts the acetabulum owing to spasticity.[340]

A second problem that can follow proximal femoral resection is heterotopic bone formation, with resultant stiffness of the resected hip.[238] In children with cerebral palsy this has been seen following soft tissue release of the hip and following spinal surgery,[244,255] but it is most commonly encountered following proximal femoral resection. The exposure of the proximal femur must be extraperiosteal, and the periosteum and all bony fragments must be thoroughly removed from the wound prior to closure to lessen the risk of this complication. When heterotopic bone blocks motion, it can be resected.

Recovery from proximal femoral resection is notoriously slow, with some patients not experiencing relief of pain and increasing mobility until 1 year following surgery.[34] Families should be warned that after resection, pain and spasticity may seem to worsen before improvement is noted. Some patients never become completely pain-free following proximal femoral resection and can pose difficult management problems (Fig. 24–65). Wheelchair modifications are always necessary following proximal femoral resection-interposition arthroplasty, as the operated leg will be markedly shortened.

Valgus Osteotomy of the Proximal Femur. Valgus osteotomy of the proximal femur is performed to allow the thigh to come out into abduction, thereby allowing perineal hygiene. The femoral head is repositioned farther lateral from the acetabulum, so that rubbing between the pelvis and the femoral head is lessened (Fig. 24–66). In this way, pain relief may occur.

The track record for proximal femoral valgus osteotomy is not particularly good, however. Sitting in a wheelchair can still be difficult because of the fixed abduction of the leg postoperatively. Samilson and colleagues found that pain relief was not predictably achieved after this procedure.[409] The femoral head produces a bump that can be felt laterally and can create pressure problems.

McHale and colleagues reported results in a very small series in which femoral head resection was combined with valgus osteotomy of the proximal femur.[280] The lesser trochanter was placed in the acetabulum and the capsule was repaired. At 3-year follow-up, sitting ability and pain had improved.[280] This procedure has not been widely used.

Hip Arthrodesis. Surgical fusion of the hip has been performed in patients with painful dislocations of the hip due to cerebral palsy. The desired position of fusion differs from that in the non-neuromuscularly population.[377] Because the primary position of the patient during daily activities is sitting in the wheelchair, a position of 50 degrees of flexion and 10 degrees of abduction is preferred in nonambulatory patients. Root and colleagues used a combined intra-articular and extra-articular technique, nearly always combined with a subtrochanteric osteotomy of the femur to allow positioning of the leg. They found that patients with cerebral palsy had a high complication rate, particularly of pseudarthrosis, following arthrodesis. Nevertheless, they advocate use of the procedure in young patients with normal spines and normal contralateral hips.[386] Bleck has rarely used arthrodesis and feels that it compromises both the sitting and supine positions and increases stress on the lumbar spine.[51]

Total Hip Arthroplasty. Root has been the greatest advocate of total hip arthroplasty in patients with cerebral palsy. The ideal candidate for joint replacement is an adult with cerebral palsy who is able to walk, stand, or transfer. Nearly all of the patients in whom he has performed total hip replacement had normal or nearly normal intelligence. Patients should not have coexisting pelvic obliquity or scoliosis.[51] Technical considerations are (1) the need to flex the acetabular component to provide posterior stability in sitting patients, and (2) the frequent loss of acetabular bone stock superolaterally, which requires bone grafting at the time of total hip arthroplasty. The use of constrained implants has led to poor results.[238] Based on their experience with instability in the early postoperative period following hip replacement with conventional nonconstrained components, Root and colleagues recommend using a spica cast for 4 weeks following surgery.[386] Despite concern about early loosening of the components, this has not been a problem in the vast majority of patients. Sixteen of 18 patients with cerebral palsy who underwent total hip arthroplasty with conventional implants at an average age of 30 years had relief of pain and improved function at an average of 10 years' follow-up.[73]

ANTERIOR DISLOCATION OF THE HIP. Anterior dislocation of the hip occurs much less frequently than posterolateral dislocation (Fig. 24–67). Children at greatest risk for anterior dislocation fall into two groups. In the first group are patients who have excessive adductor and hip flexor release, leading to extension and abduction contractures. In the second group, the dislocation appears to be due to the severe neurologic involvement of the child, with extension posturing.

Symptoms consist of inability to sit in a wheelchair due

Text continued on page 1211

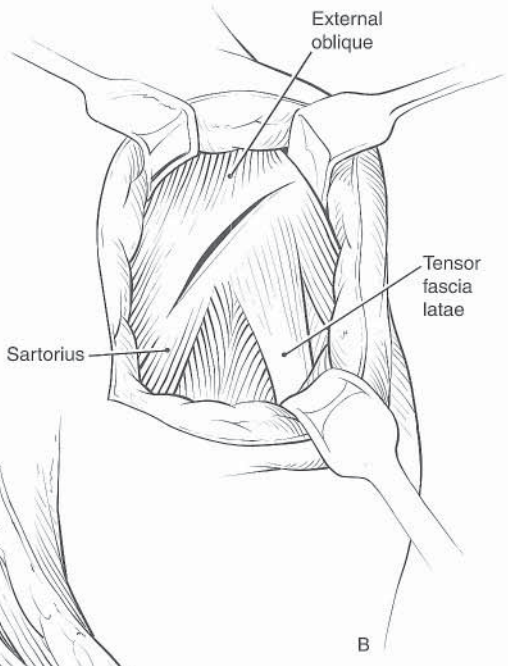
Technique for Dega Osteotomy

A, The patient is positioned supine, with the affected hip raised on a bump. An anterior incision is made over the iliac crest. The Dega osteotomy is usually performed during the same surgical setting as a VDRO, and will be illustrated as such.

B and **C,** The iliac apophysis is split and the inner and outer tables are exposed subperiosteally to the sciatic notch. The direct head of the rectus femoris is detached.

PLATE 24–10. Technique for Dega Osteotomy

A



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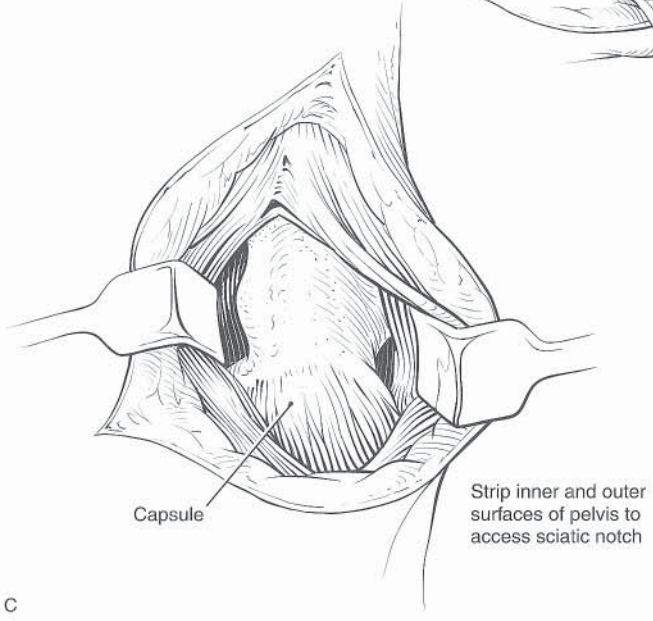


C

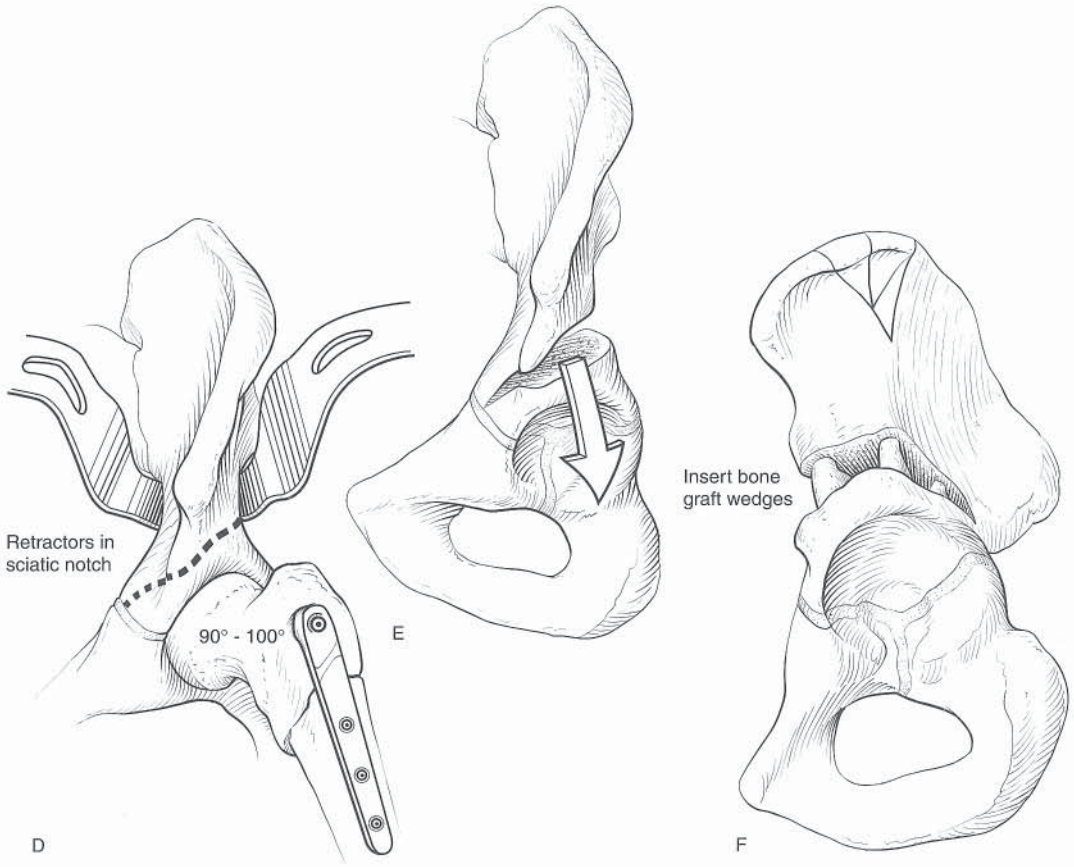
Technique for Dega Osteotomy *Continued*

D, Blunt Hohmann retractors are placed in the sciatic notch. The osteotomy is drawn on the pelvis at the level of the anterior-inferior iliac spine and extending back to the sciatic notch.

E, Osteotomes are then inserted from the outer table of the ilium down to the triradiate cartilage. The inner table is preserved. The anterior-inferior iliac spine is cut with the osteotome, and the sciatic notch is incised with a kerasin rongeur. Using osteotomes, the acetabulum is pried inferiorly and laterally. If a laminar spreader is used, great care should be taken that the bony surfaces are not crushed.

F, Tricortical wedges of iliac crest are harvested and stacked into the opening wedge of the osteotomy. The graft can be preferentially positioned to improve coverage more anteriorly, posteriorly, or just laterally. If the inner table remains intact, the osteotomy does not require fixation. Ranging the hip under visualization is recommended to verify that the osteotomy is stable. A spica cast is then used for 6 weeks to allow healing of the osteotomy.

PLATE 24–10. Technique for Dega Osteotomy



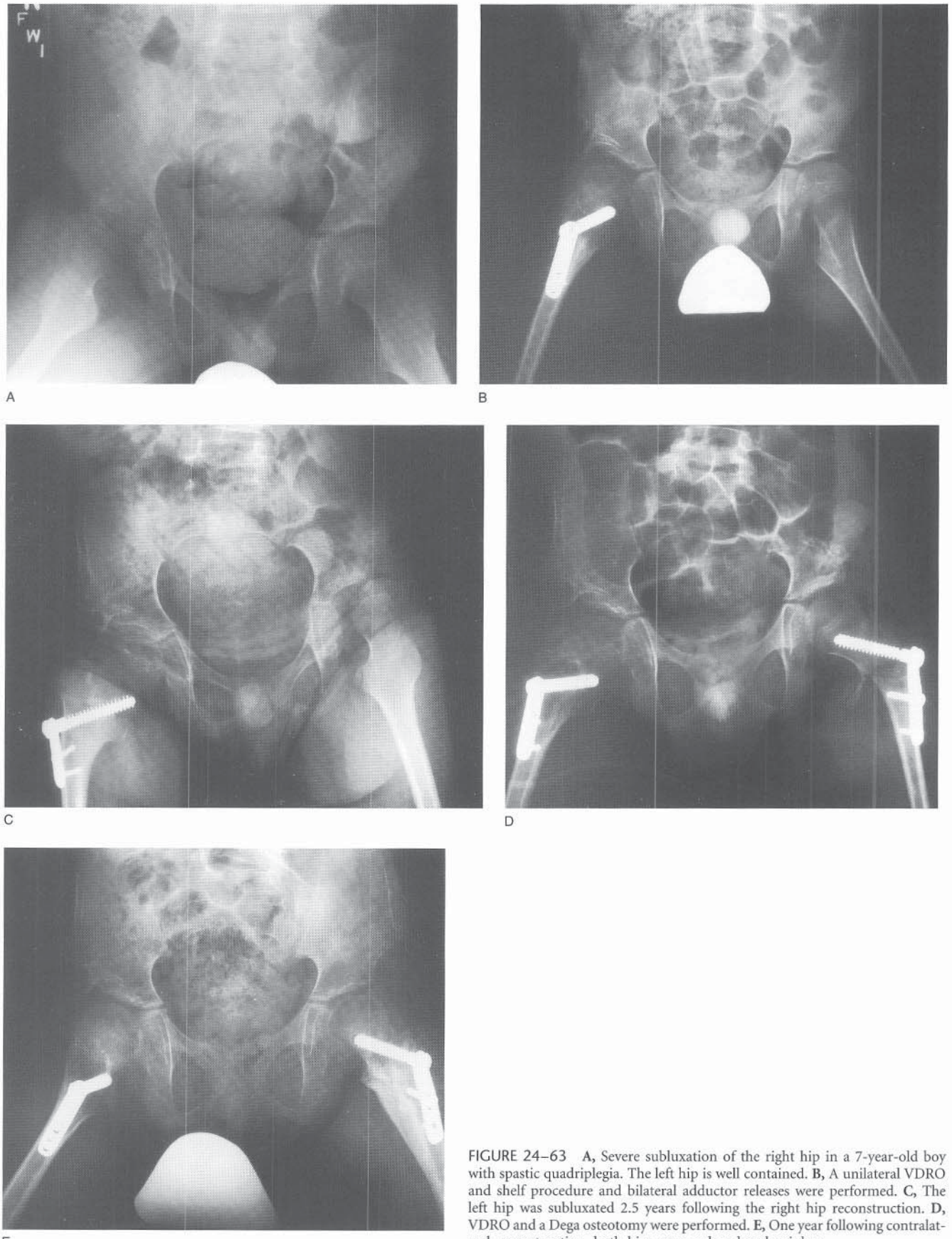


FIGURE 24–63 **A,** Severe subluxation of the right hip in a 7-year-old boy with spastic quadriplegia. The left hip is well contained. **B,** A unilateral VDRO and shelf procedure and bilateral adductor releases were performed. **C,** The left hip was subluxated 2.5 years following the right hip reconstruction. **D,** VDRO and a Dega osteotomy were performed. **E,** One year following contralateral reconstruction, both hips were reduced and painless.

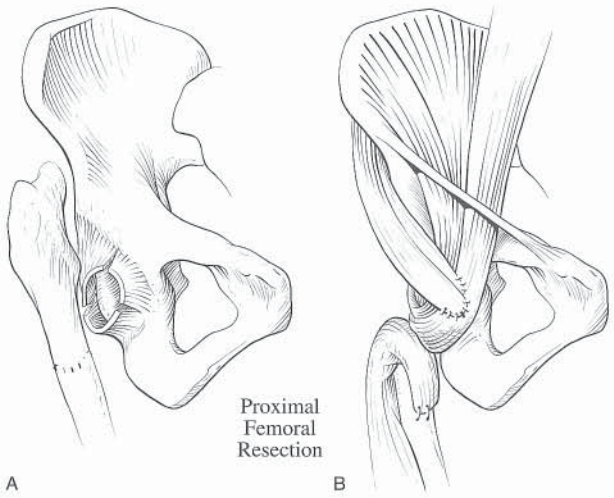
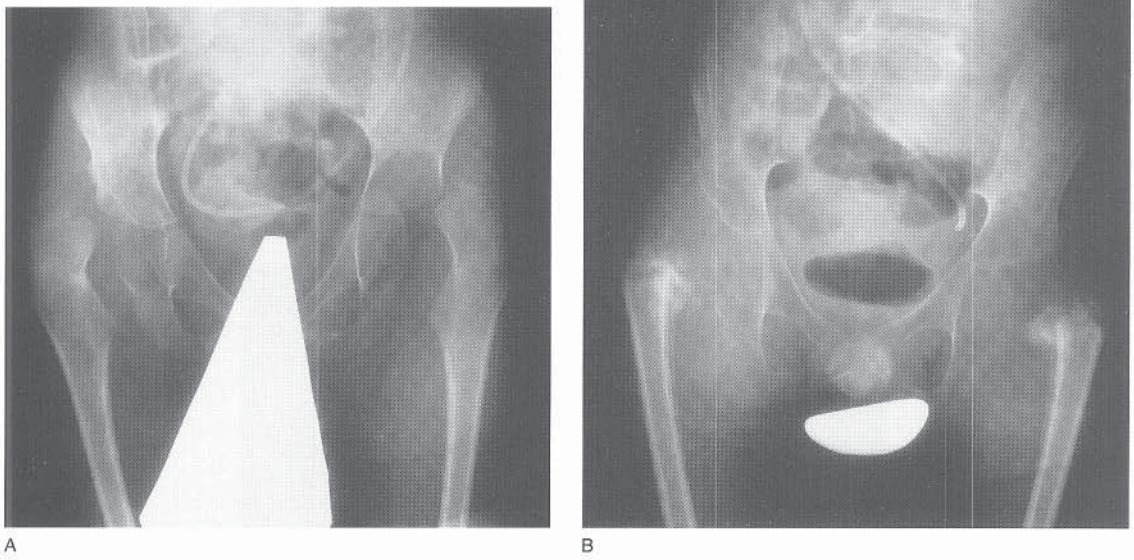


FIGURE 24–64 **A,** The planned level of resection for a proximal femoral resection is below the level of the lesser trochanter and below the level of the acetabulum. If less bone is resected, persistent pain and contracture are more likely. **B,** Interposition of soft tissue between the acetabulum and proximal femur includes closure of the capsule and suture of the iliopsoas to it. The quadriceps is sewn over the end of the proximal femur.



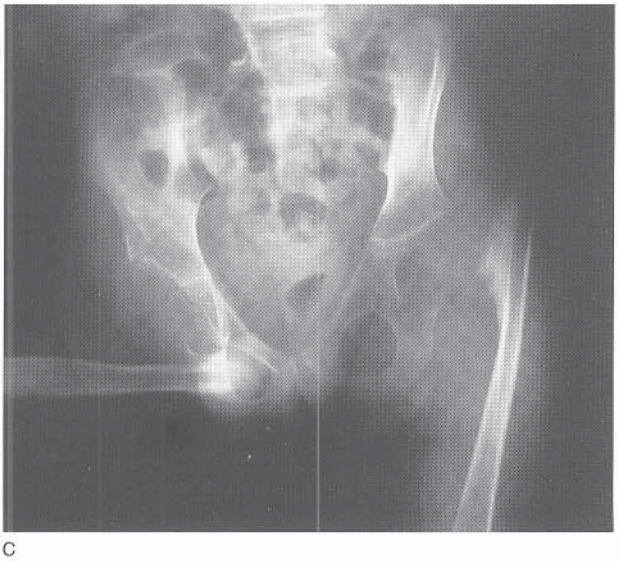
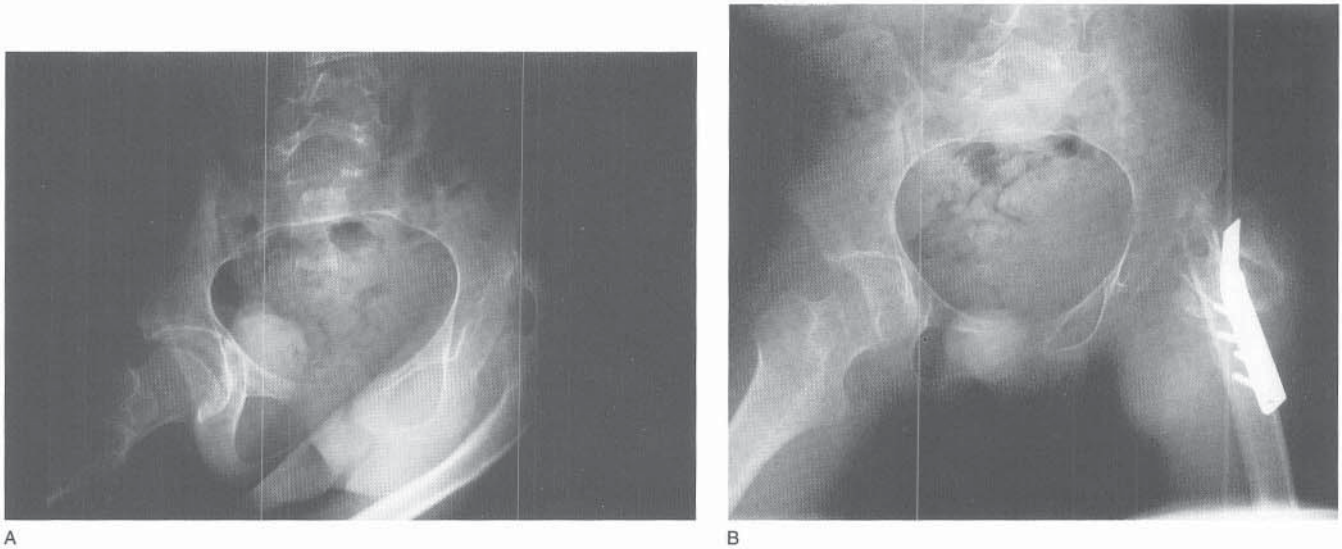


FIGURE 24–65 **A,** Bilateral painful dislocations of the hip in a 6-year-old boy with cerebral palsy. **B,** Heterotopic bone is present 2 months following proximal femoral resections. **C,** Four years later, sitting has been obstructed by recurrent contractures.



A B

FIGURE 24–66 **A,** Pelvic radiograph of a 16-year-old boy with a painful, stiff, dislocated left hip. **B,** A valgus osteotomy improved seating and hygiene.

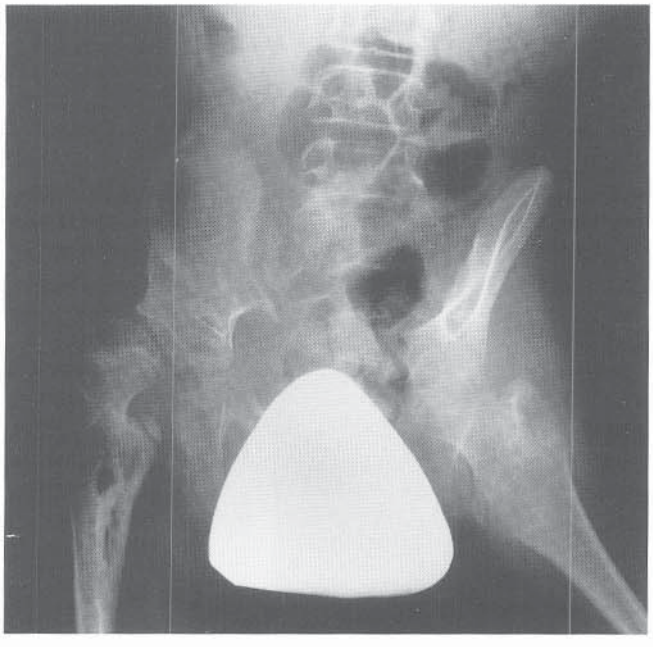


FIGURE 24–67 Anterior dislocation of the left femoral head in a 17-year-old boy with spastic quadriparesis. The right hip was posteriorly dislocated and remains subluxated despite surgery.

to extension contractures. Hip pain is present in half of these patients. The diagnosis is made on physical examination. The femoral head is palpable in the anterior groin of the patient. Flexion of the hip is limited. Radiographs may be confusing, as the hip may appear reduced as it lies anterior to the acetabulum.[69] If doubt exists, CT will confirm the diagnosis.

Treatment is difficult at best. Aggressive muscle release of the proximal hamstrings and, if the patient is abducted, the abductors and short external rotators must be included in the surgical reconstruction. A varus shortening osteotomy of the femur is recommended, in conjunction with acetabular osteotomy to gain anterior coverage. The Pemberton osteotomy is useful in this patient population, as it augments anterior coverage. Redislocation is a problem, particularly in hypotonic patients. If pain relief is not achieved, a proximal femoral resection is recommended.[421]

THE UPPER LIMB

Introduction. Operative treatment of the upper limb can improve the function of children with cerebral palsy, but only when the surgical procedure is chosen carefully and the goals are realistic. Two basic goals can be achieved when the child is functioning at a higher level: improvement in function and improvement in appearance. In children with more severe involvement who are unable to care for themselves, operative treatment may be a reasonable option if it facilitates nursing care (bathing and dressing). There are three major methods by which the surgeon can achieve some improvement: (1) lengthening of a tight musculotendinous unit, (2) augmenting a weaker muscle by tendon transfer, and (3) in the older child near the end of growth, arthrodesis.

With the judicious choice of one or more of these treatments, surgeons can alter the lives of patients in a most positive way. In the end, however, because the cause of the condition is in the brain and the operation is performed on normal muscles and joints in the extremity, the result is always to some degree unpredictable. This fact must be understood and accepted by the patient and family.

Sensory Impairment. Evaluation of sensory capacity in children with cerebral palsy is difficult, but sensory deficits are recognized to contribute more to the overall impairment in function than was previously recognized.[484] When somatosensory-evoked potentials (SSEPs) were included, impairment in at least one modality of sensory function was found in 88 percent of children with cerebral palsy.[92] Hemiplegic children who received intensive occupational therapy that concentrated on motor skills alone did not show improvement in their performance. A therapeutic focus on sensory rehabilitation is part of the nonoperative approach to the spastic upper limb.[497]

Effects on Growth. A limb that lacks normal neurologic input from any cause will often show abnormal growth. A progressive, nonproportional, and unpredictable limb length discrepancy is a common finding in spastic hemiplegia.[383] The overall rate of maturation of the limb as well as the length and girth are affected. The cause of this discrepancy is not related to the overall nutritional status of the child. In a study by Roberts and colleagues, delays in skeletal maturation on the affected side compared to the nonspastic side averaged 7.3 months.[383] Van Heest found a correlation between severity of sensory impairment and degree of growth impairment in the affected limb.[483]

Treatment. The orthopaedic management of limb problems in children with cerebral palsy has concentrated on the lower limb. It was felt that children affected by cerebral palsy would not benefit from surgical reconstruction of the upper extremity. In general, if the child has some volitional use of the hand or even awareness of its presence, surgical intervention can potentially lessen deformity and improve the function intrinsic to that limb.[484]

Success in the treatment of the spastic limb is determined by setting reasonable pretreatment goals and the by family's acceptance of these goals. The goals of treating a spastic limb include improvement in function, a decrease in deformity, improvement in appearance, and facilitation of custodial care. Volitional use of the hand is the best predictor of functional improvement following a change in the position of the hand. However, a change in the position of even a minimally functional hand may significantly alter its role as an assist hand and may improve the overall well-being of the child by normalizing the appearance and lessening the social stigma of spasticity in children who are not mentally or developmentally delayed.[251] An improvement in cosmesis can be dramatic but is often associated with disappointment in the lack of a concomitant improvement in function.

NONSURGICAL TREATMENT. Nonoperative treatment of spasticity is directed toward preventing of contractures, splinting for positional improvement, and hand therapy to improve dexterity, pattern use, and sensory reeducation. Inhibitory casting and aggressive splinting do not improve the results of standard therapy intervention and should be discouraged.[124] There is no evidence that one type of physical or occupational therapy is more beneficial than any other. Therapy protocols in which the timing, frequency, and type of intervention were varied did not show one combination to be of greater benefit than another. Recognition of the limb and a desire to use the limb are not features that can be taught by therapy or infused surgically.

Other forms of nonoperative treatment for spasticity include muscle relaxants such as baclofen, and botulinum toxin to effect a decrease in spasticity.

Botox. Botulinum toxin A (Botox) has application in some forms of cerebral palsy. Botulinum toxin prevents the release of acetylcholine at the neuromuscular junction, thereby interfering with muscle contraction. Recovery from a nonlethal dose is by nerve terminal sprouting, which occurs over the course of about 3 months. During the time of partial paralysis of the selected muscles, spasticity decreases and contractures can be stretched out more easily. The function of the antagonistic muscles may be unmasked, exposing the potential for better use of the hand. Surgical procedures can be planned based on the results of the selective Botox injections. Corry and co-authors compared botulinum toxin with sterile saline in a randomized, double-blind study and found improvement in elbow and thumb extension, elbow and wrist flexor tone, and grasp and release. They suggest that the ideal patient is one with marked flexor spasticity, no contracture, and some volitional control of the limb.[95]

Contraindications appear to be fixed contractures, lack of volitional control, and lack of spasticity. Repeated injections may be needed, owing to neural recovery. Potential problems include the development of antibodies to Botox and the cost of treatment.

SURGICAL TREATMENT. The surgical treatment of upper limb spasticity in cerebral palsy, a *neuromuscular* problem, is limited to what can be done at the *musculoskeletal* level. The goals of surgical treatment must be clearly defined and accepted preoperatively. These goals can be a position change to facilitate available function, to improve appearance, or to facilitate hygiene or custodial care. All of these goals are clear indications for surgical care and must be discussed thoroughly with the patient and the family, which must understand that surgery cannot increase volitional use or sensibility. Most surgical procedures attempt to restore balance by combinations of soft tissue releases and tendon transfers where possible, and by arthrodesis where soft tissue procedures are inadequate. Selective neurectomy of motor nerves within spastic muscles, or with chemical block, also has a place in the treatment of spasticity, although the results are rarely lasting and are difficult to control. Rhizotomy, a surgical procedure that ablates spinal motor nerve roots, is usually reserved for cases of uncontrolled spasticity in the lower limbs and has little application in the upper limb.

Preoperative Evaluation. General considerations: Other medical conditions must be under optimal control before elective surgery is performed, including any seizure disorder or dental condition. Coordination of other surgical services to address lower limb or eye deformity concomitantly may be appropriate.

Motor examination: The typical posture of the spastic upper limb is elbow, wrist, and finger flexion and forearm pronation. Muscles with fixed contractures and muscles that are consistently spastic are usually easy to identify. Athetosis in a muscle is manifested by the lack of a contracture and a variable tone in the muscle that allows intermittent full excursion, often without volitional control. An athetoid muscle should not be selected as a donor for a tendon transfer because the result is unpredictable (Fig. 24–68).

Dystonia is a movement disorder characterized by rigid co-contractions of opposing muscles that worsens with emotional overlay and is not amenable to surgical correction. Bony procedures that increase rigidity are contraindicated in dystonia. EMG with gait analysis can be used to provide this information; however, careful clinical examination may provide the same information to the careful examiner.

Sensory examination: Awareness of the limb and discrimination of pain and temperature are critical to incorporation of the limb into a use pattern. Detailed and discrete sensory evaluation is difficult in a child with cerebral palsy. Some patients who are able to articulate their perceptions will describe alterations in the sensibility of the involved hand that preclude manipulation and identification of small objects without visual input.

Surgical Planning. Surgical goals include release of contracture that interferes with function, cosmesis, or hygiene; tendon transfers to restore balance to forearm rotation, wrist, thumb, and finger position; and joint stabilization in the situation in which balance cannot be maintained by soft tissue.

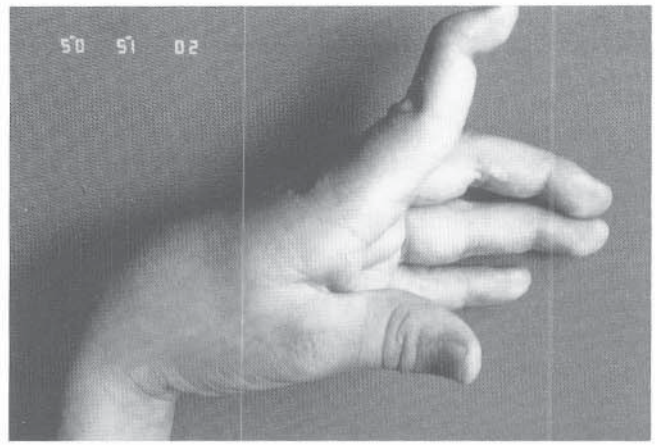


FIGURE 24–68 Hyperextensibility of the digits with variable tone may indicate an athetoid component and should signal caution in considering tendon transfers to these digits.

Elbow. Flexion contracture: The severe elbow flexion contracture occurs when there is very little hand function, and usually in the globally involved child. The indications for contracture release are to improve hygiene or facilitate custodial care. A shoulder adduction contracture may be present and can be ameliorated at the same time with an intramuscular lengthening of the pectoralis major muscle.

A severe elbow flexion contracture can be relieved by release of the lacertus fibrosis, Z-lengthening of the biceps tendon, and lengthening of the brachialis tendon with several chevron-type incisions through an anterior Z-plasty or long curvilinear approach. Loss of some active flexion is expected. The radial nerve must be protected, and the proximal origin of the brachioradialis muscle can be released if the elbow is still tight. The elbow is casted in comfortable extension for 3 weeks until wounds have healed, and then splinted intermittently to maintain a range of motion that allows ease of dressing and positioning for balance and limited function.

Forearm pronation: Pronation contracture of the forearm develops insidiously and, with growth, causes a rotational deformity of the radius and occasionally dislocation of the distal radioulnar joint (Fig. 24–69). When the forearm is passively correctable, the options for correction of the deformity are release or rerouting of the pronator tendon. If the pronator muscle is constantly spastic, with no in-phase activity, it should not be transferred. Rerouting of the pronator tendon uses a long Z-lengthening, prolonging the proximal slip of tendon with a strip of distal periosteum. Strong, nonabsorbable sutures placed in one-half of the tendon are used to pass the distal limb dorsally through the interosseous space and secure it to the proximal limb with the forearm positioned in full supination. Whether the pronator has been lengthened or transferred, the forearm is protected with a long-arm cast in full supination for 4 weeks.

Releasing or rerouting the pronator should be done cautiously when the flexor carpi ulnar is used for a transfer to radial wrist extensors because of the additional supinatory torque produced by that transfer.[401] A position of fixed supination is worse than one of pronation.

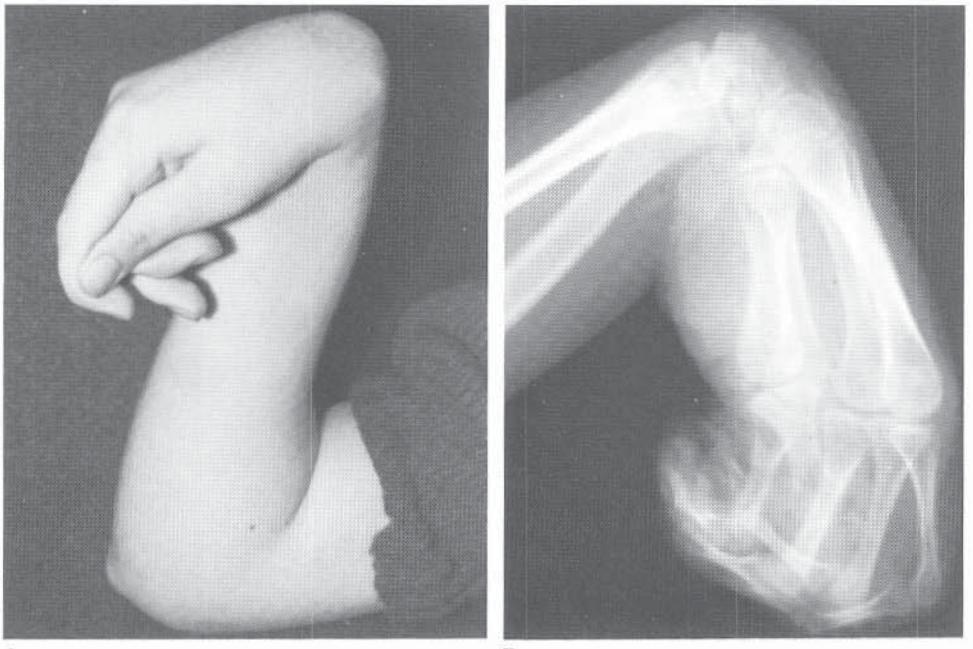


FIGURE 24–69 Acute flexed wrist and hand with pronation of the forearm in a child with cerebral palsy. **A,** The patient's upper limb. **B,** Lateral radiograph showing the volar subluxation of the carpus.

Wrist and Fingers. The goal of surgical procedures on the wrist and fingers is to allow the fingers to open with wrist flexion for release and to close with wrist extension for grasp. Fine motor and individual finger movement is not the intent of the surgical procedures for the wrist and fingers in cerebral palsy. Some wrist flexion must be preserved to allow finger extension, and wrist extension strong enough to resist the flexion force of the fingers is needed for grasp. Achieving this balance is often difficult.[360]

Judicious lengthening of the wrist flexors, the flexor carpi ulnaris at the intramuscular level, and the flexor carpi radialis with a Z-lengthening, and release of the palmaris longus and superficial fascia, will allow the wrist to be brought into an extended position. The extensor carpi ulnaris often is overactive as an ulnar deviator but remains in phase as an extensor and is the preferred tendon to transfer into the extensor carpi radialis brevis. This transfer has less of a supinatory torque than the Green procedure (transfer of the flexor carpi ulnar is to the extensor carpi radialis brevis). The tension is set with the wrist in maximum passive extension and the extensor carpi ulnaris in moderate tension as it is woven into the tendon of the extensor carpi radialis brevis distal to the first compartment muscles (Plate 24–11).

Extrinsic finger flexion contractures may limit both finger and wrist extension and can be addressed by lengthening in the forearm at the intramuscular, tendinous, or muscle-origin level. Fractional lengthening of the flexor profundus is feasible when the flexor tone is such that the surgeon cannot extend the patient's fingers passively with the wrist in neutral position (Plate 24–12). In general, greater length can be obtained by formal Z-lengthening than at the intramuscular level, but at the expense of muscle function and strength. If more than 45 degrees of wrist flexion is needed to fully extend the fingers, lengthening by a proximal flexor-pronator origin release is indicated if the desire is to preserve finger flexor function.

Lengthening of the extrinsic finger flexors may worsen the intrinsic contractures by altering the relationship between the superficialis and profundus tendon lengths. Greater relative lengthening of the superficialis weakens proximal interphalangeal joint flexion while increasing tension on the lumbrical (which takes its origin from the profundus tendon) and increases flexion of the metacarpophalangeal joint and hyperextension of the proximal interphalangeal joint, causing a severe swan's neck deformity in the face of intrinsic spasticity. Lengthening of the intrinsics or a superficialis flexor tenodesis may help address this deformity.

Severe deformity: For severe wrist flexion contractures, especially in the nonfunctional hand, wrist arthrodesis can offer a solution to hygiene and care problems (see Chapter 14, Disorders of the Upper Extremity, in the section on juvenile arthritis, under the subsection entitled Wrist Fusion). Resection of the proximal carpal row and fusion of the transected capitate and hamate to the distal radius will often allow sufficient soft tissue decompression and facilitate release or lengthening of the extrinsic and intrinsic finger contractures. The released wrist flexors then become available to augment finger extension in selected cases.

Thumb. The thumb-in-palm deformity is common with deforming forces, including the adductor pollicis and all thumb intrinsic muscles (Fig. 24–70). The approach to the thumb-in-palm deformity is to release contracted soft tissue and then augment weak extensors and abductors.[402] If the child demonstrates active thumb interphalangeal extension and a palpable extensor pollicis longus, simple release of the contracture will usually suffice to bring the thumb out of the palm. A comprehensive release of the origins of the thenar musculature, protecting the recurrent branch of the median nerve, and of the two heads of the adductor pollicis, protecting the deep branch of the ulnar nerve, will bring the thumb into a position of wide abduction (Fig. 24–71). A release of the first dorsal interosseous fascia with a release

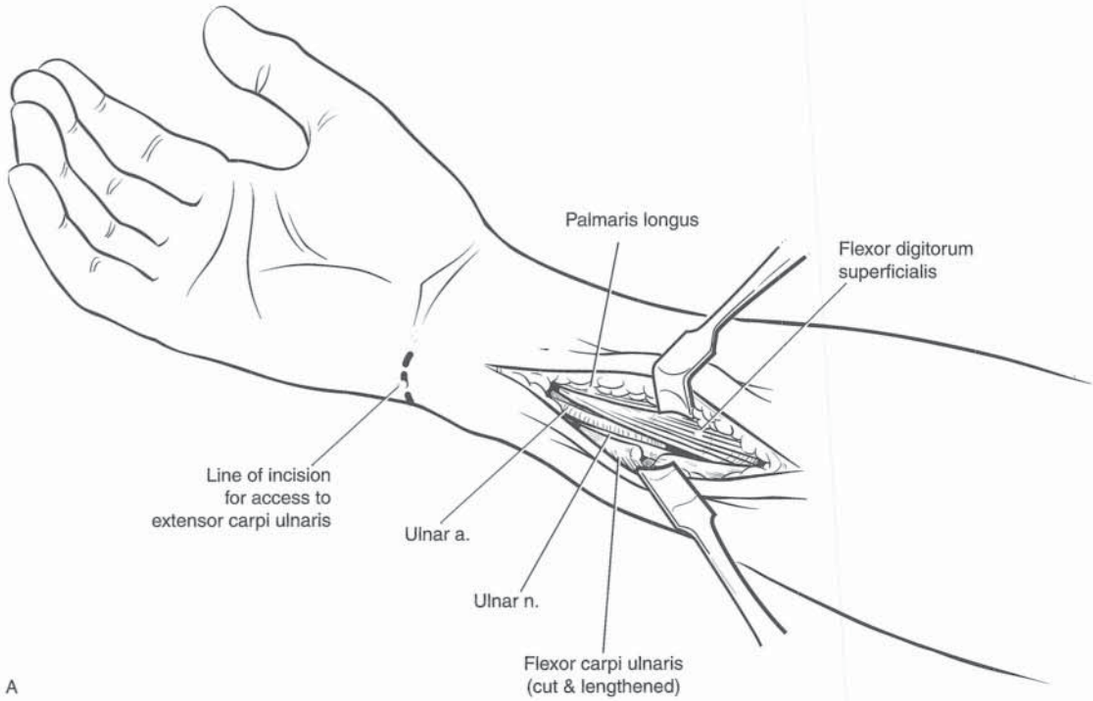
Text continued on page 1222

Extensor Carpi Ulnaris–Extensor Carpi Radialis Brevis Transfer

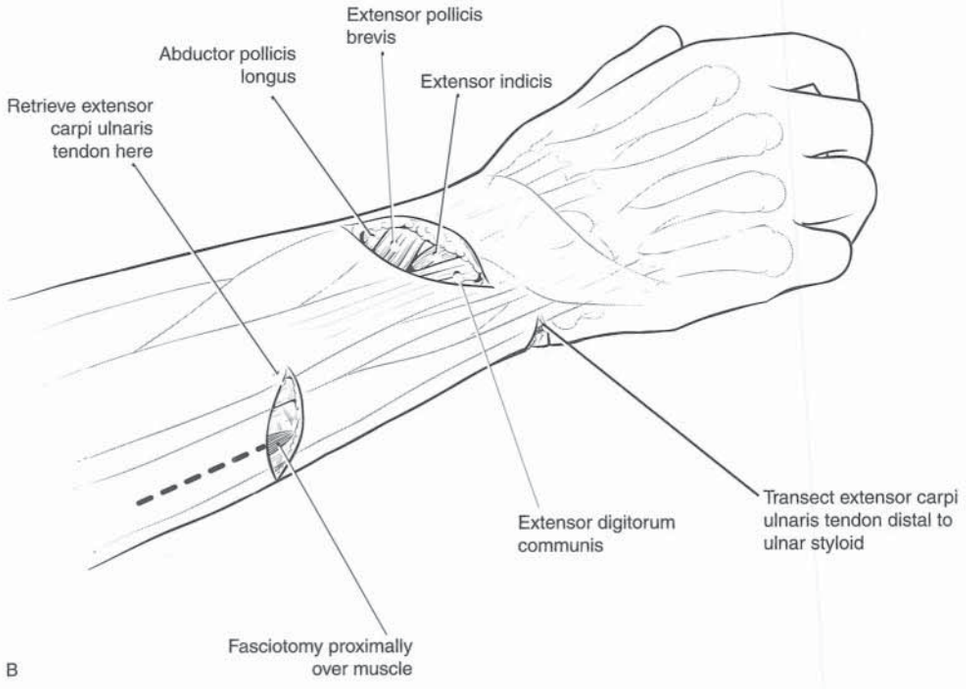
A, Lengthening of the flexor carpi ulnaris tendon is done at the musculotendinous level in the distal forearm. Lengthening of the other wrist flexors can be accomplished through the same incision if needed.

B, The incision to expose the extensor carpi ulnaris tendon is made just distal to the ulnar styloid. The extensor carpi ulnaris tendon is then retrieved into the proximal incision and transferred subcutaneously to the dorsoradial wrist incision.

PLATE 24–11. Extensor Carpi Ulnaris–Extensor Carpi Radialis Brevis Transfer



A

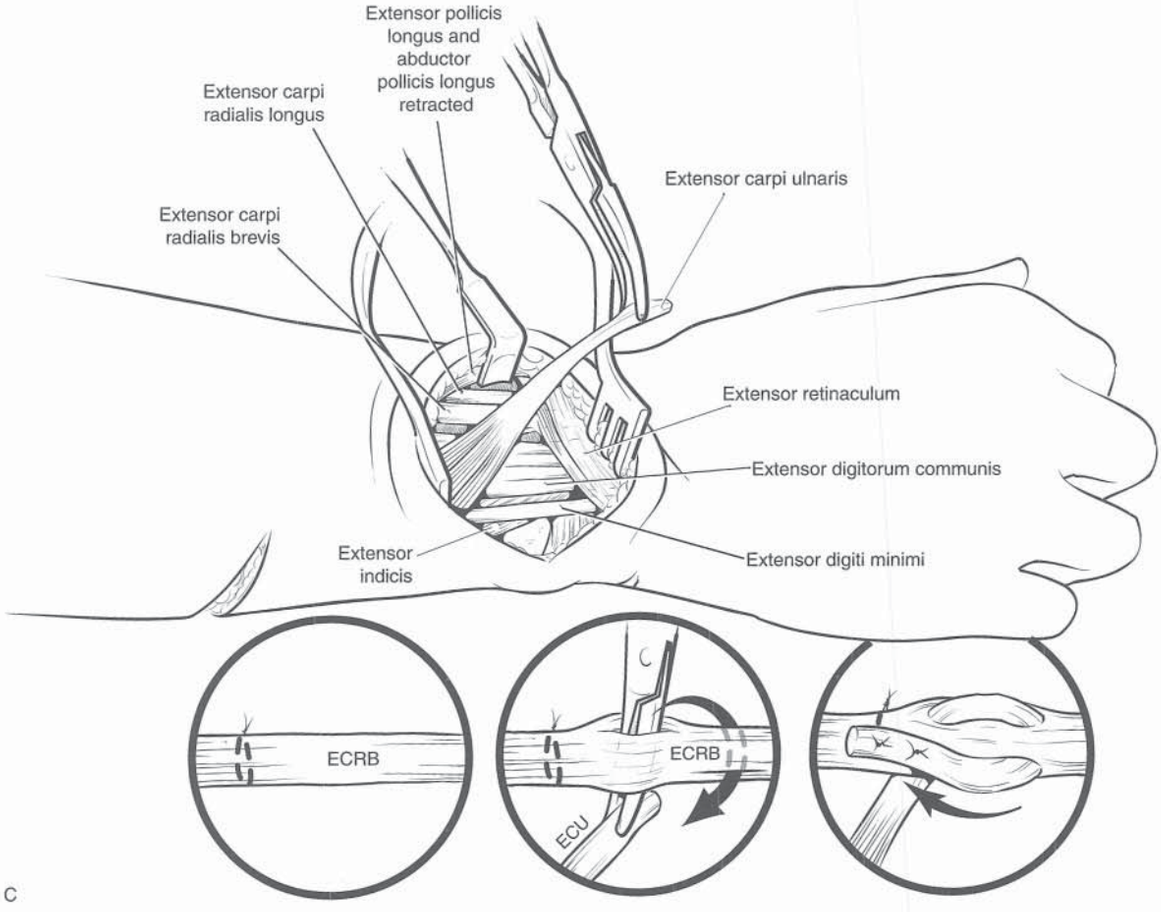


B

Extensor Carpi Ulnaris–Extensor Carpi Radialis Brevis Transfer
Continued

C, The tendon transfer is secured using a weave technique. The first suture is placed proximal to the site of the first pass of the donor tendon through the recipient tendon to prevent proximal migration of the tendon placement.

PLATE 24–11. Extensor Carpi Ulnaris–Extensor Carpi Radialis Brevis Transfer



C

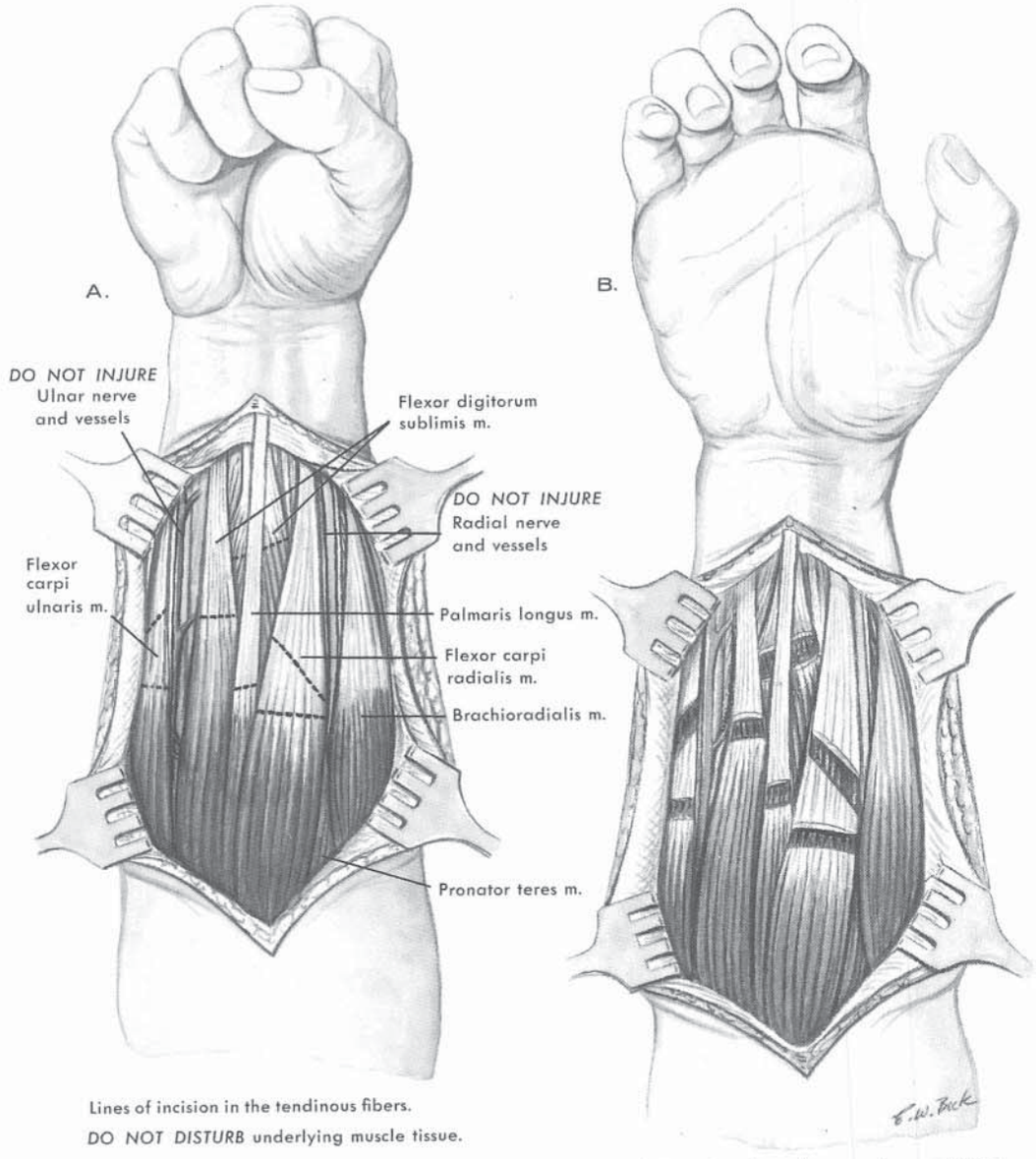
Fractional Lengthening of Finger and Wrist Flexors in Forearm

OPERATIVE TECHNIQUE

A, A midline longitudinal incision is made in the middle three-fourths of the volar surface of the forearm. The subcutaneous tissue and deep fascia are divided in line with the skin incision. The wound flaps are undermined, elevated, and retracted with four-prong rake retractors to expose the superficial groups of muscles. On the radial side of the flexor carpi ulnaris tendon, the ulnar vessels and nerves are identified and protected from injury; similarly, on the radial side of the flexor carpi radialis tendon, the radial vessels and nerve are isolated to protect them from inadvertent damage. Sliding lengthening of the flexor carpi radialis and flexor carpi ulnaris muscles is performed at the musculotendinous junction by making two incisions of their tendinous fibers, about 1.5 cm apart, without disturbing underlying muscle tissue. The proximal incision is transverse and the distal one is oblique. The palmaris longus and flexor digitorum muscles are lengthened by only one transverse incision in each.

B, The wrist and the fingers are passively hyperextended. The tendinous parts will separate while the intact underlying muscle fibers will maintain continuity of the muscles.

PLATE 24–12. Fractional Lengthening of Finger and Wrist Flexors in Forearm



Lines of incision in the tendinous fibers.
DO NOT DISTURB underlying muscle tissue.

Separation of tendinous parts on extension of wrist and digits

Fractional Lengthening of Finger and Wrist Flexors in Forearm
Continued

C and **D,** The deep volar muscles are exposed by retracting the brachioradialis muscle and radial vessels radially, and the flexor carpi radialis and flexor digitorum sublimis muscles ulnarward. The median nerve is identified and protected from injury by retracting it medially with the flexor carpi radialis muscle. The flexor pollicis longus and flexor digitorum profundus muscles are lengthened by making two incisions in their tendinous parts and sliding them in the same manner as described for the superficial volar forearm muscles. Continuity of muscles is maintained by gentle handling of tissues and by taking care that there is adequate muscle substance underlying the divided tendinous parts. Sliding lengthening is achieved by separating the tendinous fibers by slow but firm extension of the thumb and four ulnar digits.

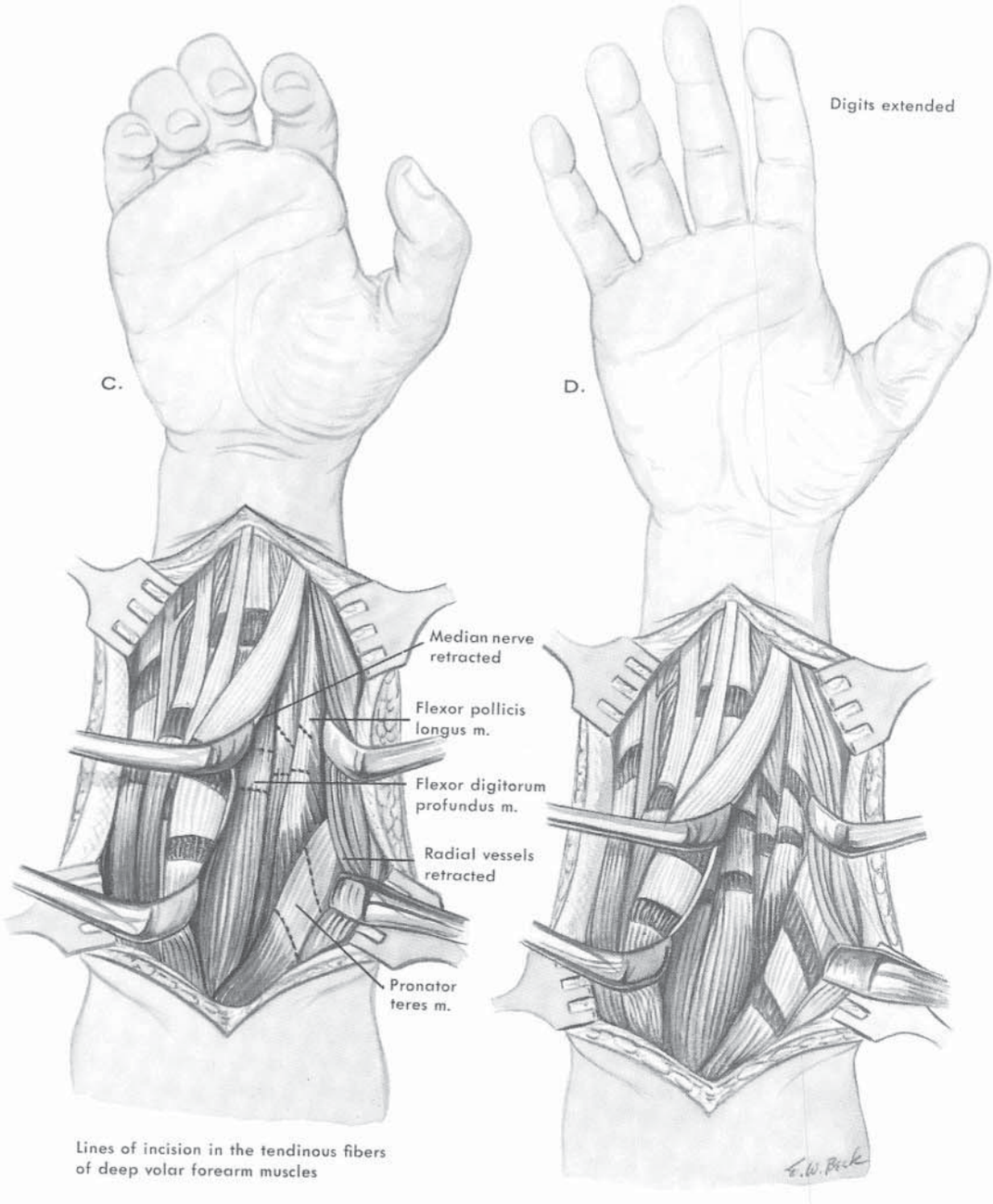
Next, the range of passive supination of the forearm is tested. If there is pronation contracture, the pronator teres muscle is lengthened by two oblique incisions, 1.5 cm apart, of its tendinous fibers. Again, underlying muscle tissue should not be disturbed. The forearm is forcibly supinated; the tendinous segments will slide and separate, elongating the muscle.

The tourniquet is released and complete hemostasis is obtained. The deep fascia is not closed. The subcutaneous tissue and skin are approximated by interrupted sutures. An above-elbow cast that includes all the fingers and the thumb is applied to immobilize the forearm in full supination, the elbow in 90 degrees of flexion, the wrist in 50 degrees of extension, and the fingers and thumb in neutral extension.

POSTOPERATIVE CARE

Four weeks following surgery, the cast is removed and active exercises are started to develop motor power in the elongated muscle. Squeezing soft balls of varying sizes and other functional exercises are carried out several times a day. An aggressive occupational therapy program is essential. The corrected position is maintained in a bivalved cast. As motor function develops in the elongated muscle and its antagonists, the periods out of the cast are gradually increased.

PLATE 24–12. Fractional Lengthening of Finger and Wrist Flexors in Forearm



Lines of incision in the tendinous fibers of deep volar forearm muscles

Note the sliding lengthening by separation of tendinous fibers

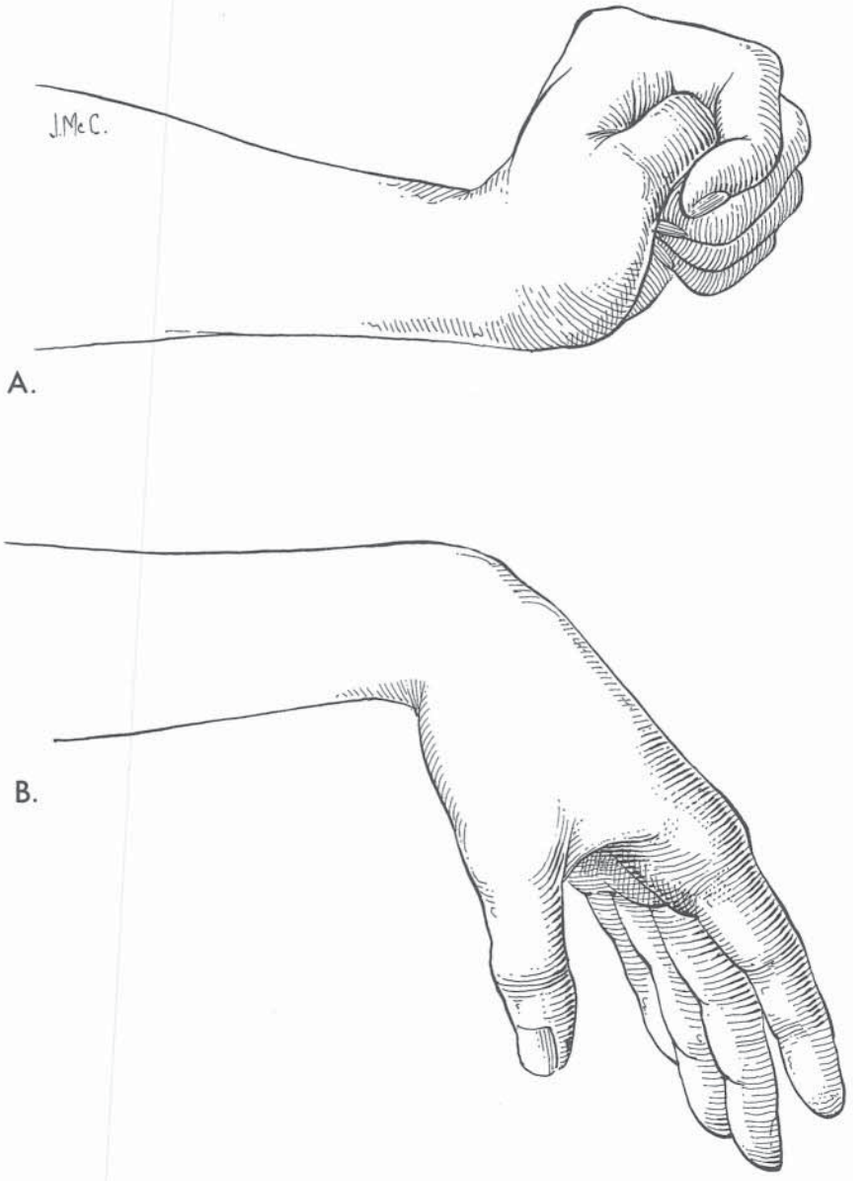


FIGURE 24–70 Thumb-in-palm deformity of the spastic hand. **A,** The fingers are clenched in the palm over the thumb. **B,** On hyperflexion of the wrist, the fingers and thumb extend out of the palm.

of the thumb-index web space may also be needed. The thumb metacarpophalangeal joint is often unstable and hyperextended. This must also be addressed so that extensor power to the thumb will lift the entire thumb ray and not increase hyperextension instability of the metacarpophalangeal joint (Fig. 24–72).[371] The joint may be stabilized by capsulodesis, sesamoid to metacarpal head fusion, metacarpophalangeal arthrodesis, tenodesis of the extensor pollicis brevis proximal to the metacarpophalangeal joint, or a combination of these procedures. Augmentation of the extrinsic extensors and abductors involves tendon transfers to the first dorsal compartment or extensor pollicis longus in its native or rerouted position.

Postoperative care for contracture releases includes immobilization for a minimum of 3 to 4 weeks, followed by splinting. If a tendon transfer has been done, immobilization should be prolonged to 6 weeks, followed by splinting full-time for an additional 6 weeks. Thereafter the splint is worn for protection during strenuous activity. Arthrodesis must be protected until radiographic evidence of healing.

SPINAL DEFORMITY

Scoliosis. Scoliosis is a significant problem in children with cerebral palsy, affecting between 25 and 68 percent of patients.[268,407] As is the case with hip instability, the incidence is highest in patients who are nonambulatory and who have total body involvement. Series that include patients who are ambulatory and have milder neurologic involvement yield smaller incidences, while those that study institutionalized patients show a higher frequency. Up to 64 percent of institutionalized adults with cerebral palsy have scoliosis.[268,475] Patients with spasticity are at highest risk, compared to patients with other movement disorders.[268]

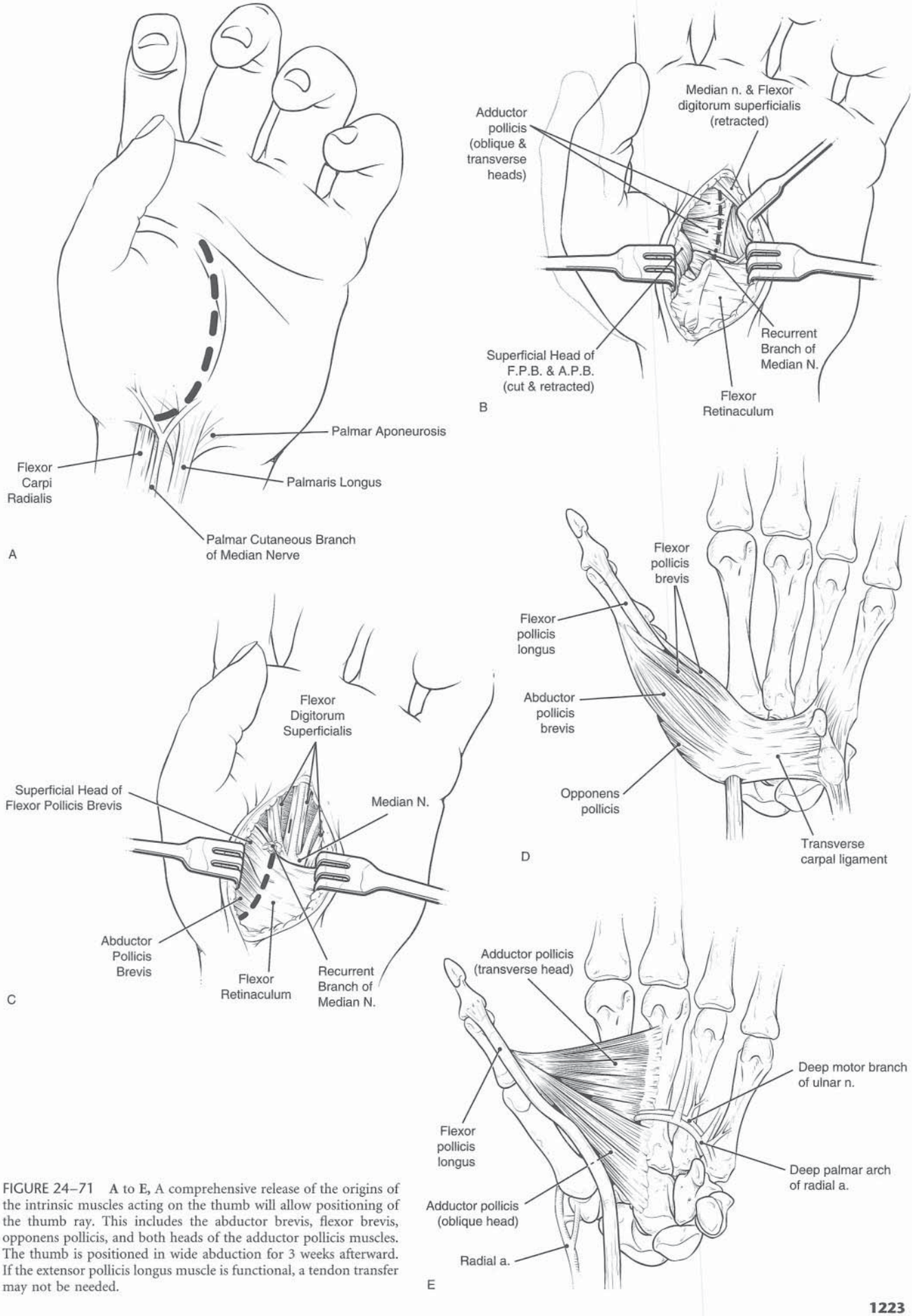


FIGURE 24–71 **A** to **E**, A comprehensive release of the origins of the intrinsic muscles acting on the thumb will allow positioning of the thumb ray. This includes the abductor brevis, flexor brevis, opponens pollicis, and both heads of the adductor pollicis muscles. The thumb is positioned in wide abduction for 3 weeks afterward. If the extensor pollicis longus muscle is functional, a tendon transfer may not be needed.

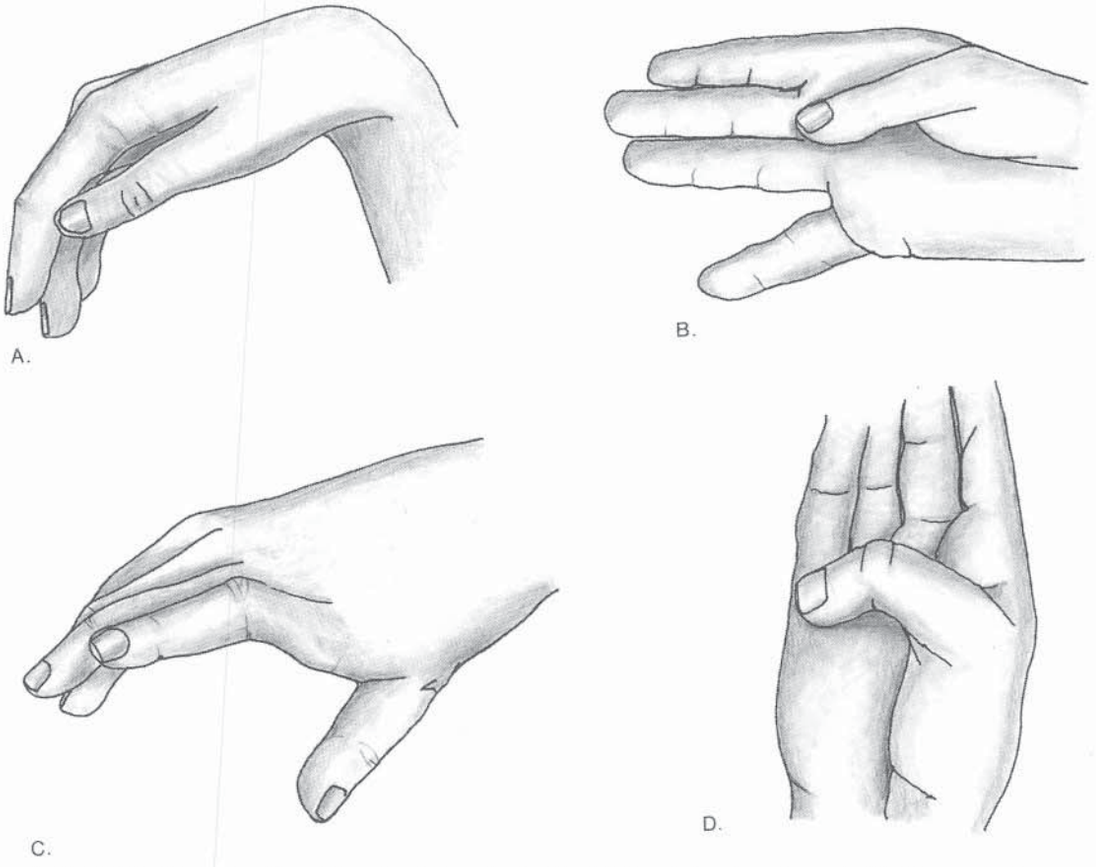


FIGURE 24–72 Deformities of the thumb in cerebral palsy. **A,** Type I, simple metacarpal adduction contracture. **B,** Type II, metacarpal adduction contracture and metacarpophalangeal flexion deformity. **C,** Type III, metacarpal adduction contracture combined with a metacarpophalangeal hyperextension deformity or instability. **D,** Type IV, metacarpal adduction contracture combined with flexion deformity of the metacarpophalangeal and interphalangeal joints.

There is a typical curve pattern in scoliosis secondary to cerebral palsy that differs from that seen in idiopathic scoliosis. The usual curve is a long sweeping curve that extends to the pelvis, with the apex of the curve at the thoracolumbar junction. Rotation accompanies the coronal plane curvature[20,466] (Fig. 24–73).

Scoliosis leads to difficulty in sitting, which is the functional position needed by the wheelchair-bound child. Curvature of the lumbar spine leads to pelvic obliquity, which can produce uneven pressure on the ischial tuberosities and eventual pressure sores.[435] Seating imbalance forces the child to lean on the upper extremities, becoming a hands-dependent sitter or even a propped sitter (Fig. 24–74).[367]

Nonoperative treatment of scoliosis with adaptive seating and orthoses has not met with success. Curve progression is not controlled by bracing, a fact that has been proven in many studies. Bracing had no impact on scoliosis curve, shape, or rate of progression in spastic quadriplegic patients who were followed by Miller and colleagues to fusion.[286] Bracing can lead to skin intolerance in these thin children, but Letts and colleagues found that seating was made somewhat easier when a soft orthosis was prescribed. The brace was used only to allow comfortable seating, not to treat the curve.[256] Wheelchair adaptations can help the child with scoliosis sit but do little to halt worsening of the curvature.

With discouraging results from nonoperative treatment, it would be logical that all patients with scoliosis due to cerebral palsy would undergo surgical correction. Yet this is not the case. Spinal fusion surgery carries sufficient risks that it is not felt to be in the best interest of every child with neuromuscular scoliosis. When deciding whether a specific patient should undergo spinal fusion, it is important to know what the natural history of scoliosis in this population of patients is.

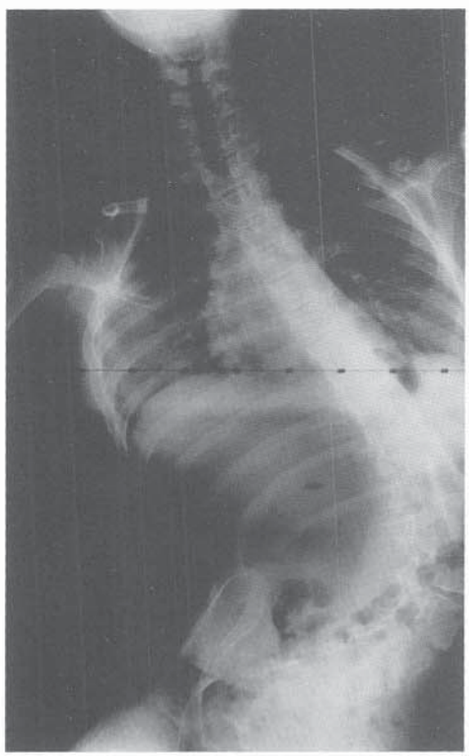


FIGURE 24–73 Long thoracolumbar scoliosis in a 14-year-old nonambulatory girl with cerebral palsy. The curve is associated with severe pelvic obliquity, which compromises seating.

Majd and colleagues followed all patients in a nursing home and documented whether or not they had scoliosis and whether their curves progressed.[269] They found that 18 percent of the patients had significant deterioration in their curves. The larger curves tended to progress in adulthood at a rate of 4.4 degrees per year. Three patients had decubiti, and their average curves were greater than 100 degrees, with more than 45 degrees of pelvic obliquity.[269] Thometz and Simon found similar results, but the rate of progression of curves greater than 50 degrees at skeletal maturity averaged only 1.4 degrees per year. They also found that thoracolumbar and lumbar curves tended to progress more than thoracic curves.[475] If a curve is more than 40 degrees by age 15 years, it is likely to progress.[399]

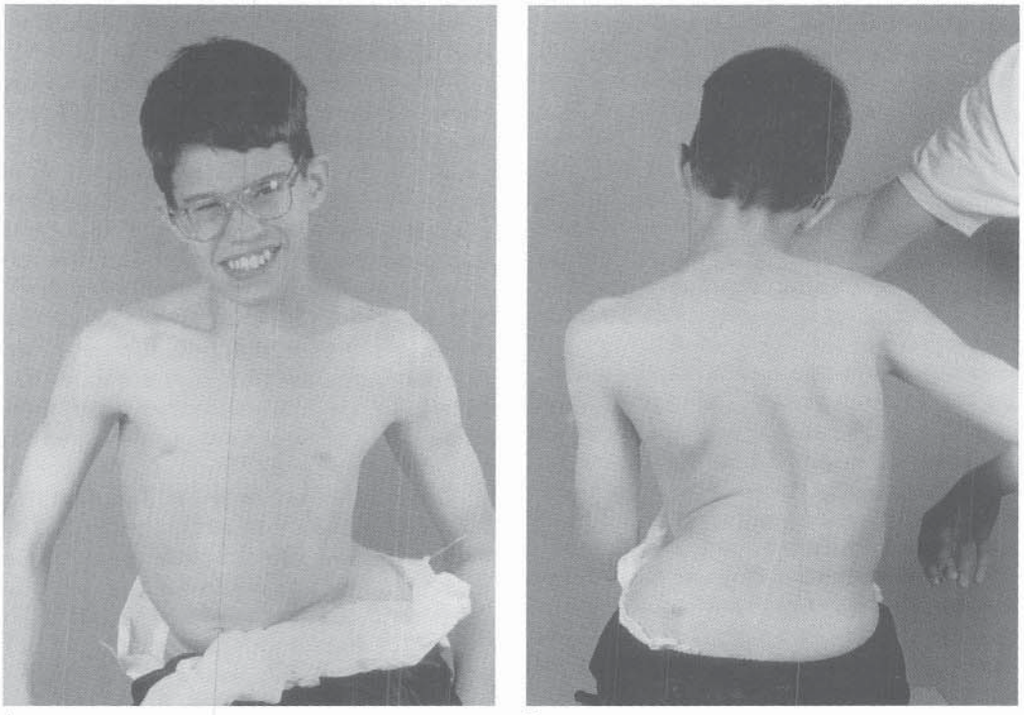
These studies looked at whether or not curves progressed following skeletal maturity, but they did not specifically address whether patients with larger curves were less healthy or more difficult to nurse. Kalen and associates compared 14 residents of a nursing home who had scoliosis of between 51 and 105 degrees with 42 residents with either no scoliosis or small curves. They found that the patients with larger curves had more orthopaedic deformities such as hip dislocations, and that they needed modified wheelchairs. There was no difference in the incidence of decubiti, functional level or loss of function, or oxygen saturation, however.[230] They concluded that problems with functional loss and decubiti were seen in equal proportions in both groups, so the surgical indications for spinal fusion in cerebral palsy were not clear.

Cassidy and colleagues took this question a step further and analyzed the health and nursing care of a group of institutionalized patients with cerebral palsy who had undergone spinal fusion surgery and a similar group of patients with scoliosis of more than 50 degrees who had not been operated on.[81] They found no significant difference in pain, pulmonary status, decubiti, function, or time required for daily care. The nurses caring for these patients, however, felt that those who had undergone spinal fusion were more comfortable. Based on this study and the study by Kalen and colleagues,[230] the indications for surgery in patients who are institutionalized and severely mentally retarded remain clouded.

Our current indications for spinal fusion are the following:

1. Curves of more than 50 degrees in ambulatory patients
2. Progressive curves of more than 50 degrees in patients who are communicative and aware of their surroundings
3. Curves that interfere with seating and nursing in patients whose families desire surgical correction

Because the prevalence of scoliosis increases with the severity of neurologic involvement, the surgeon is often faced with the decision whether or not to operate on a child who is profoundly mentally retarded and unaware of his or her surroundings. In these difficult cases, we believe that it is the family that often makes the decision to undergo surgery or not. When surgery is not felt to be in the best interest of the child and when the family chooses not to "put the



A B

FIGURE 24–74 **A** and **B,** Clinical appearance of a 13-year-old boy with significant scoliosis who underwent a rhizotomy. He is unable to sit independently without propping with his arms.

child through" the surgery, modified seating can allow the child to be moved about.[367]

Once the decision to operate has been made, a thorough medical evaluation is necessary.[493] Malnutrition is frequently a problem in these patients and, when present, predisposes to infection and delayed wound healing. Laboratory studies, including measurement of serum protein and albumin and a total lymphocyte count, are useful in assessing the nutritional status of the child. A serum albumin level of 35 g/L and a total lymphocyte count of 1,500 cells/mm^3 have been established as levels below which complications occur more frequently.[224] Gastrostomy tube feedings may be necessary preoperatively in order to lessen the risk of complications. Aspiration has been documented in 69 percent of patients with total body involvement cerebral palsy.[137] If aspiration occurs in the postoperative period, pneumonia frequently results, prolonging the hospital stay of the child and even resulting in death in some children. Preoperative swallowing studies should be performed in patients suspected of aspirating, and referral to a pediatric surgeon should be made when these studies confirm aspiration. Certain seizure medications that are frequently used in patients with cerebral palsy can increase blood loss by interfering with coagulation. Those patients treated with Depakote or Depakene will have normal routine coagulation profiles—that is, prothrombin time and partial thromboplastin time—but prolonged bleeding times.[493] Preparations for large intraoperative blood loss must be made.

The surgical treatment of scoliosis in patients with cerebral palsy differs from the treatment for idiopathic scoliosis. The bone in nonambulatory patients is osteopenic, so that hook sites may be weak, leading to pull-out of hooks and loss of fixation. In patients with neuromuscular scoliosis, segmental fixation is preferred. The Luque technique, which uses sublaminar wires at each level, distributes the corrective forces equally throughout the spine. Loss of fixation rarely occurs, and bracing is not needed in the postoperative period. The children can be mobilized immediately following surgery, thus lessening the risk of pulmonary complications such as pneumonia and atelectasis. Correction is usually readily achieved and maintained.

The use of sublaminar wires does impose a greater neurologic risk, however, as each wire must be passed between the lamina and the dura. Generous removal of the ligamentum flavum with kerasin rongeurs can facilitate passage of the wires. Careful contouring of the wires must be done to minimize protrusion of the wire beneath the lamina. The wires must always be pulled up away from the dura, never pushed into the canal, and the wires are bent along the surface of the lamina following passage to prevent bouncing the wire down on the dura as the surgery continues.

Interspinous process segmental instrumentation has been previously described in patients with scoliosis due to cerebral palsy. Wires are placed at each level through the base of the spinous process and wired to either a Harrington or a Luque rod.[134,440] Although it is neurologically safer to keep the wires out of the spinal canal by placing them through the base of the spinous processes, most authors still prefer the Luque technique as it allows greater correction and better sagittal contouring, and there is less loss of fixation. Gersoff and Renshaw reported an average correction of 52 percent following Luque sublaminar segmental fixation, with only an average of 3 degrees loss of correction at follow-up.[170]

In patients with idiopathic scoliosis, it is the rule to

instrument and fuse as little of the spine as possible while correcting and stabilizing the curve. In scoliosis due to cerebral palsy, just the opposite is true. The rule is to instrument and fuse long, with the fusion extending from the second thoracic vertebra to the pelvis in nonambulatory patients, and to the lower lumbar spine in those who do walk. Fusing short often leads to additional levels adding on to the curve over time.[89] Some authors have had success fusing to L5 without inclusion of the pelvis, particularly in older patients with milder deformities.[456] Nonetheless, pelvic obliquity is best improved and the correction maintained by fusion to the pelvis. Isolated anterior fusions may be tempting in cases of lumbar and thoracolumbar curves, yet time has shown that short fusions may not be adequate in the long run, and the reoperation for a curve that has added on is rarely satisfying. Additionally, kyphosis may result from stopping the fusion in the midthoracic spine as the patient leans forward in the wheelchair.[60,382]

Use of the 1/4-inch stainless steel Luque rod is preferred over the 3/16-inch rod whenever possible. Rod breakage is significantly less with the larger rods.[60,170,382] Rigid cross-links should be used to prevent migration of the rods relative to one another with loss of correction of pelvic obliquity.

Fusion of the spine to the pelvis is accomplished using the Galveston technique, described by Allen and Ferguson (Fig. 24–75).[13] The posterior iliac wings are exposed by stripping the gluteal muscles from the outer table. The sciatic notch must be identified. A drill is used to create a passage for the rod from the posterior-superior iliac spine along the transverse bar between the inner and outer tables of the ilium. The rods should be inserted 6 to 9 cm within the ilium, and come to lie superior to the sciatic notch.[14]

Contouring of the Galveston bend requires practice, as the rod must make two bends which are three-dimensional.[15] The rod is divided into three sections: the spinal segment, the sacral segment, and the iliac segment.[13] The bend between the spinal segment and the sacral segment is made first. The less experienced surgeon must remember that the bend itself will take up approximately 1 to 2 cm of rod. A 60- to 80-degree bend is made and checked next to the patient's spine. A second bend is then made distally to drop the rod down into the ilium using a rod clamp and a tube bender. Finally, the spinal segment of the rod is bent to accommodate scoliosis and sagittal contouring of kyphosis and lumbar lordosis. The rod must fit easily within the prepared site in the ilium, since forcefully manipulating the rod may lead to it cutting out of the ilium.

Alternatively, the unit rod can be used. The unit rod is a single U-shaped rod that is precontoured with the Galveston bend for insertion into the iliac wings of the pelvis. With the rods seated within the iliac crests, the proximal aspect of the rod can be levered to correct pelvic obliquity, and then bent in situ and wired down onto the laminae for correction of scoliosis. Biomechanically, the technique offers stable fixation, but it is more technically challenging than using two Luque rods inserted into the pelvis with cross-links. It is particularly difficult to use when there is lumbar hyperlordosis.[129] Studies have shown superior correction of scoliosis, ranging from 55 to 78 percent.[40,128] Correction of pelvic obliquity with the unit rod can average up to 82 percent.[72,270]

Yet another form of fixation that is suitable for many patients with cerebral palsy is the Dunn-McCarthy technique, in which two S-shaped rods are placed over the sacral ala (Fig. 24–76). Upgoing laminar hooks or pedicle screws are placed more proximally, and distraction is applied to seat the rods firmly against the sacral ala.[278] Segmental fixation is then completed with wires or hooks. The advantage of this system is that the sacroiliac joint is not crossed, which should lead to less movement and loosening over time compared to the Galveston technique, following which loosening of the rods within the iliac wings commonly occurs. The exposure of the sacral ala sites is less time-consuming than preparation of the ilium for the Galveston rod. Correction of scoliosis averaged 70 percent in a group of 17 patients with cerebral palsy treated with S-rod fixation to the sacrum.[277] Postoperative complications are similar to those seen with Galveston fixation—wound infections, pressure sores, and occasional loss of sacral fixation.

There are no distinct guidelines for when anterior release, discectomy, and fusion is necessary in patients with cerebral palsy. Prior to the advent of segmental sublaminar fixation, combined anterior and posterior fusion was recommended for all patients with scoliosis and pelvic obliquity.[62] The addition of an anterior fusion reduced the pseudarthrosis rate from 22 to 5.4 percent in a series published in 1983 by Lonstein and Akbarnia.[264] Today, more secure segmental fixation has necessitated anterior fusion in only a subset of patients. Younger patients with open triradiate cartilages may develop the crankshaft phenomenon, with isolated posterior fusion in the face of postoperative anterior vertebral growth. These patients are best served by anterior fusion combined with posterior instrumentation and fusion. There is no role for posterior instrumentation without fusion in this patient population.[382] Severe, stiff curves may require anterior release and fusion to improve the surgical correction of the deformity.[128] Rinsky proposed that curves over 70 degrees be released anteriorly prior to posterior instrumentation and fusion.[382] Boachie-Adjei and colleagues recommended preliminary anterior release and fusion for curves over 90 degrees and for curves in which a stretch supine radiograph showed lack of correction of pelvic obliquity.[54] Anterior instrumentation offers no additional benefit over simple discectomy and fusion.

In the past, anterior release and fusion and posterior fusion were staged 1 to 2 weeks apart, with some surgeons placing the patient in traction between the two procedures.[55,382] More recently, anterior release and fusion have been done under the same anesthetic as posterior instrumentation and fusion. Although complications are frequent, regardless of the timing of the surgeries, Ferguson and associates found that nearly twice as many neuromuscular patients who underwent same-day anterior and posterior surgery remained without complications as patients who underwent staged procedures. Same-day surgery decreased the total anesthetic time, surgical blood loss, and hospital stay. Additionally, the nutrition of the patients was better if the anterior and posterior approaches were done on the same day, as they had only to recover from one very large surgery instead of two large surgeries.[155,318] Whenever possible, anterior and posterior surgery are carried out on the same day in our patient population.

The best way to avoid pseudarthrosis in this patient population is to perform a meticulous dissection and fusion. We

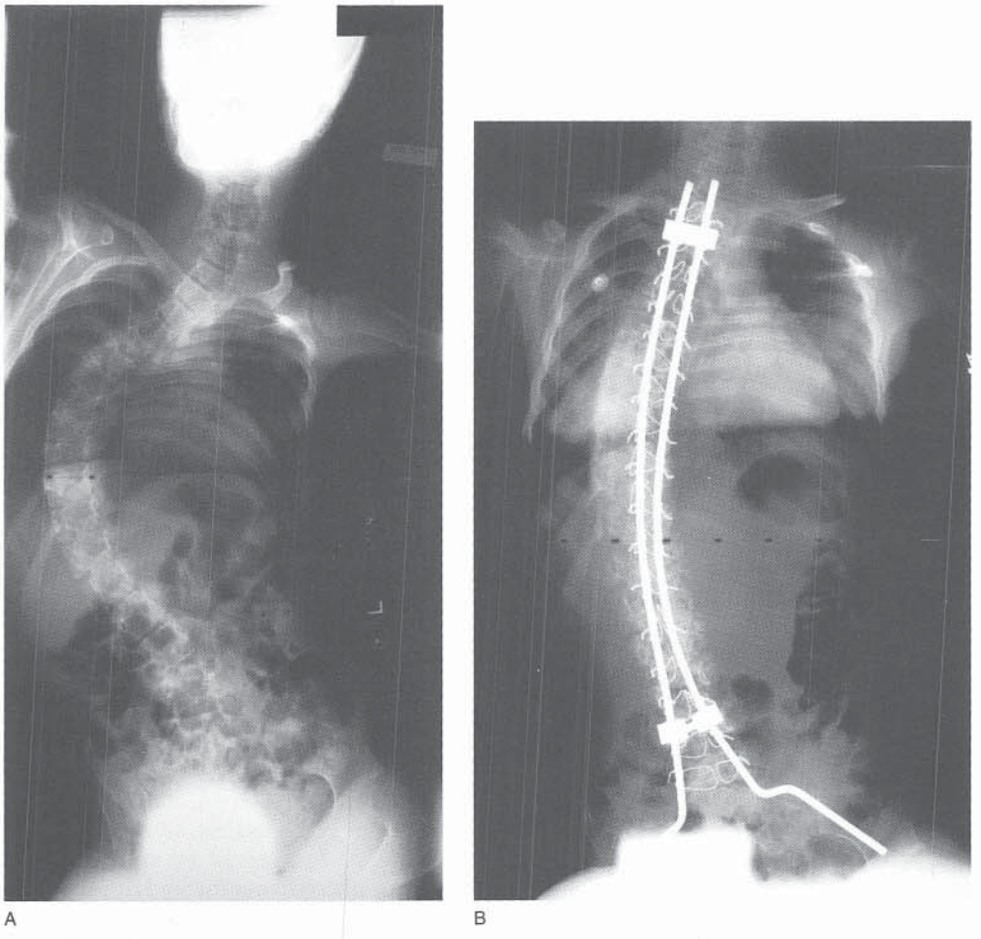


FIGURE 24–75 **A,** Eleven-year-old girl with spastic quadriparesis and scoliosis measuring 87 degrees. **B,** Posterior spinal fusion was performed with Luque rods, sublaminar wires, and Galveston technique, extending the fusion to the pelvis. **C,** The rods are contoured in the sagittal plane. **D,** Fixation was maintained and the fusion has consolidated 5 years postoperatively.

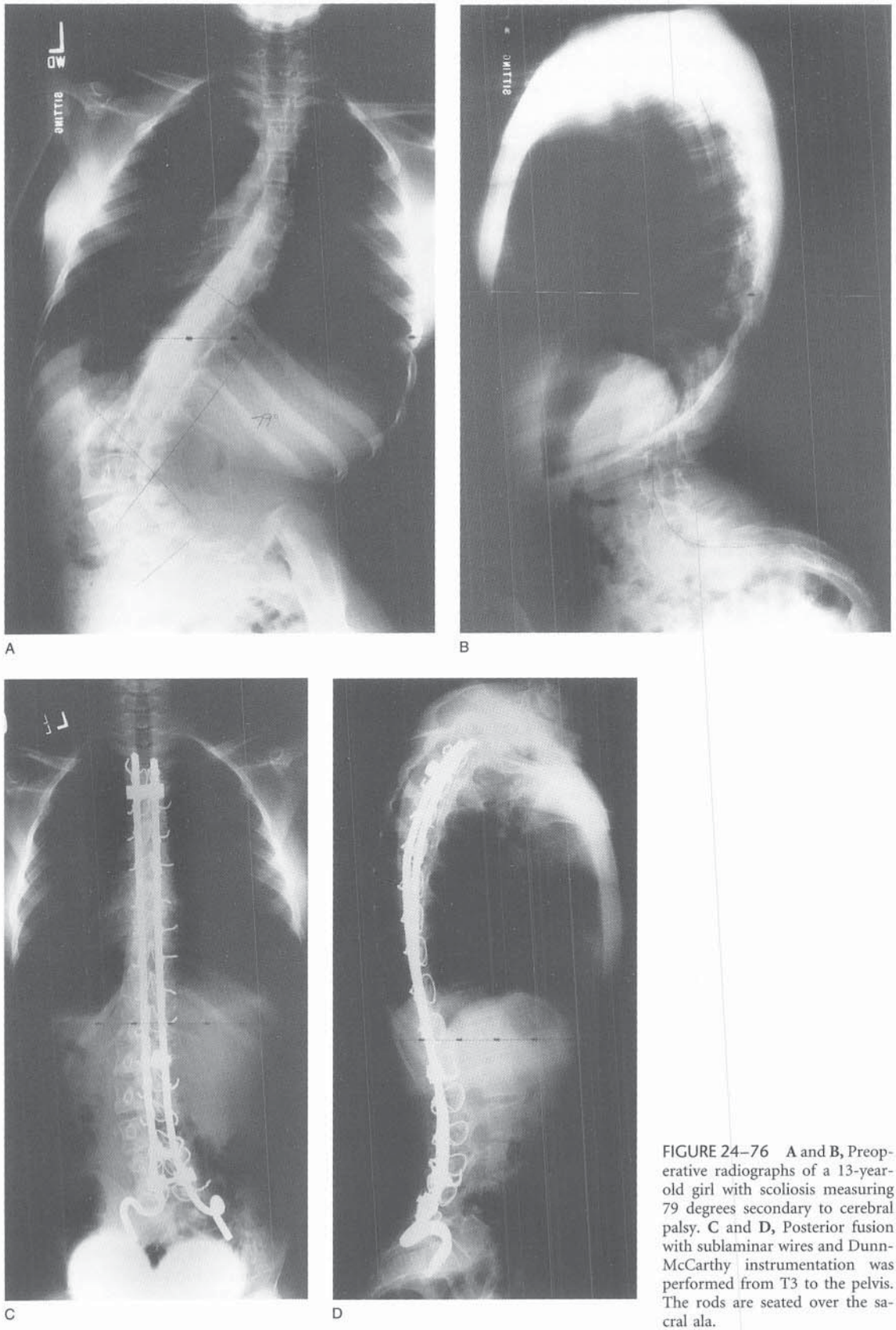


FIGURE 24–76 **A** and **B,** Preoperative radiographs of a 13-year-old girl with scoliosis measuring 79 degrees secondary to cerebral palsy. **C** and **D,** Posterior fusion with sublaminar wires and Dunn-McCarthy instrumentation was performed from T3 to the pelvis. The rods are seated over the sacral ala.

perform facetectomies and decorticate the entire spine and the exposed part of the sacrum, and apply copious amounts of bone graft to facilitate fusion. Because of the pelvic extensions of the rods, autograft from the iliac crest cannot be obtained in sufficient quantity to adequately graft the entire area to be fused. For this reason, allograft bone is used for the fusion.[295]

As always, it is important to contour the rods carefully in the sagittal plane for thoracic kyphosis and lumbar lordosis. Hyperlordosis is commonly present in lumbar and thoracolumbar curves and can lead to increased difficulty with sitting and discomfort. Spines with extreme hyperlordosis can be difficult to instrument posteriorly. If hip flexion contractures are contributing to the lordotic posture, hip flexor releases may improve the deformity prior to spinal surgery.

Intraoperative spinal cord monitoring should be performed during the surgical correction of scoliosis due to cerebral palsy. Although a series from Rancho Los Amigos showed only 53 percent successful tracings in cerebral palsy patients,[21] others have had greater success in being able to monitor the spinal cord of these children. Cortical responses can be more difficult to measure in patients with cerebral palsy. Ecker and colleagues were able to measure cervical/brainstem SSEPs in 31 of 34 cerebral palsy patients, with one false positive and no false negative tracings.[145] Loder and colleagues found that spinal cord monitoring in neuromuscular patients revealed many intraoperative changes, particularly during tightening of sublaminar wires, but that very few of these events led to neurologic change.[262] We continue to use spinal cord monitoring whenever possible in patients undergoing spinal surgery with cerebral palsy.

SURGICAL COMPLICATIONS. Patients who undergo surgical correction of scoliosis have a high likelihood of developing postoperative complications. In recent series, complication rates have been as high as 62 percent.[169] Pseudarthrosis may occur in up to 10 percent of children treated with modern instrumentation techniques. More likely is radiographic evidence of movement of the iliac extensions of the Luque-Galveston rods within the pelvis, seen as a radiolucency around the rods. This "windshield-wipering" was documented in 26 of 68 patients in a recent series but was not necessarily symptomatic in the majority of children (Fig. 24–77).[169]

Postoperative curve progression has been seen following posterior spinal fusion with Luque instrumentation. In a series reported by Comstock and associates, either the curve progressed or the pelvic obliquity progressed in 33 percent of patients. They attributed the high rate of curve progression to failure to fuse to the pelvis, not fusing proximally enough, and failure to perform anterior fusion in immature patients.[89] The crankshaft phenomenon has been observed in skeletally immature patients with cerebral palsy who underwent isolated posterior spinal fusions with either Luque or Luque-Galveston instrumentation.[411]

Wound infection is a common problem in patients with cerebral palsy who undergo scoliosis surgery.[264] Infection occurred in 8.7 percent of 172 patients who had surgical treatment for neuromuscular scoliosis recently.[469] Wound infection is most likely in malnourished patients.[138,235] Infection with gram-negative organisms is seen more frequently in this population because of contamination from the diaper

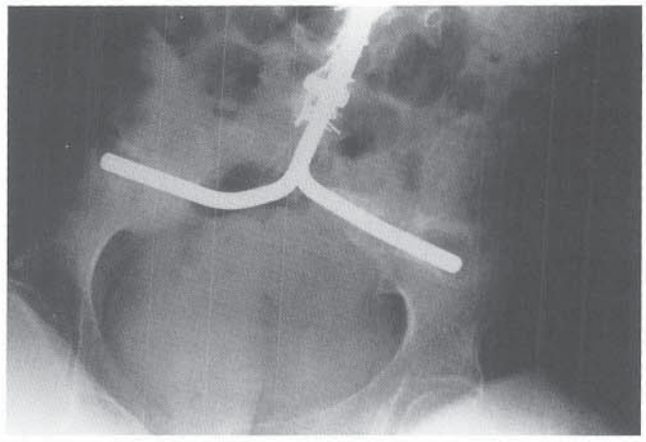


FIGURE 24–77 AP pelvic radiograph of a patient who underwent posterior spinal fusion for scoliosis secondary to cerebral palsy using the Luque-Galveston technique. There is radiolucency present around the ileal portion of each rod, indicating loosening. The patient was asymptomatic.

in patients with bowel and bladder incontinence. When infection occurs, it usually is located in the distal part of the incision.[469] Hematomas frequently accumulate in patients with cerebral palsy and may also become infected. When infection does occur, it usually responds to multiple irrigation and debridements, and hardware removal is rarely necessary.[224,346,469]

Decubitus ulcers can occur following spinal fusion. Prolonged recumbency in thin, malnourished patients predisposes to the development of bed sores. Persistent pelvic obliquity can also create uneven pressure over the ischium in sitting and lead to breakdown.[169]

Progression of deformity may occur following spinal instrumentation and fusion in patients with cerebral palsy. The reasons for curve progression are adding on to the curve through unfused and uninstrumented segments (i.e., below the distal end of a fusion that does not extend to the pelvis),[89] and pseudarthrosis with or without rod breakage. Additionally, cases have been described in which the iliac fixation was lost owing to rod perforation through the ilium. Revision surgery is possible but carries a high risk of subsequent complications.[129]

Spondylolysis and Spondylolisthesis. Bleck first proposed that hip flexion contractures and increased lumbar lordosis may lead to an increased incidence of spondylolysis and back pain in patients with cerebral palsy. Rosenberg and associates studied a nonambulatory group of patients with cerebral palsy and found that none of them had either spondylolysis or spondylolisthesis. It appears that weightbearing is a prerequisite for the development of the pars stress fracture, which occurs universally at L5, the bridge between the lumbar spine and the sacropelvis. Although Hennrikus and associates did not find an increased incidence of spondylolysis in their patients,[203] Harada and co-workers found spondylolysis in 21 percent of 84 patients with spastic diplegia. Those patients with spondylolysis had greater lumbar lordosis and smaller sacrofemoral angles than patients without pars fractures.[198]

Cervical Spine Spondylosis. Patients with athetoid-type cerebral palsy are prone to the development of cervical spon-

dylosis and resultant myelopathy based on the movement disorder. Athetoid patients have uncontrollable writhing movements, and the neck is rapidly flexed and extended in what has been described as a "whip movement."[144] This places bending and shear moments on the upper cervical spine, leading to spondylosis over time. Symptoms develop as early as late adolescence, but more commonly in early adulthood. Complaints consist of neck pain and arm pain, with weakness and decreased sensation in the arms and legs present on physical examination. Patients who are able to walk and communicate note deterioration in their gait. Deep tendon reflexes are increased, but this is commonly seen in most patients with cerebral palsy and may not be helpful in differentiating those with cervical cord embarassment.

Radiographs show flattening of the anterosuperior aspect of the vertebral bodies, with osteophytes at the anteroinferior margins. The disc spaces are narrow. Increased movement is seen on flexion-extension lateral radiographs. A study by Harada and co-workers also showed stenosis of the spinal canal in patients with athetosis, which may predispose to neurologic injury.[199] The most frequent levels involved are C3–4 and C4–5.[143]

Treatment is surgical. Laminectomy is contraindicated because it does not address the problem of instability. Nishihara and associates reported successful results with anterior interbody fusion and postoperative immobilization with a halo vest. They warn of the tendency for degeneration of the levels adjacent to the fusion.[315] Others advocate combined anterior and posterior spinal fusion.[199,285]

RHIZOTOMY

Selective dorsal (or posterior) rhizotomy is a neurosurgical procedure in which a percentage of the dorsal roots are severed at the level of the cauda equina in order to reduce spasticity.[322] Spasticity is improved by reducing the stimulatory inputs from the muscle spindles of the lower extremities that arrive via afferent fibers in the dorsal roots.[377] This surgery as currently performed was described by Fasano[153] and popularized in the United States by Peacock.[336]

The surgery entails laminectomy from L2 to L5 or S1. The facets are preserved. The dorsal roots are identified and subdivided into rootlets. Usually 25 to 50 percent of the posterior nerve rootlets from L2 to S2 are divided, using EMG guidance.[337] More recently, the laminae are replaced at the end of surgery.[88,448]

Postoperative care focuses on aggressive physical therapy to restore strength. Once spasticity is relieved, underlying weakness of the muscles becomes apparent, and physical therapy is prescribed three to five times a week to restore strength.

The rate of good results following rhizotomy depends most critically on proper patient selection. Ideal candidates are less than 8 years old, with some advocating rhizotomy at very early ages—between 2 and 4 years.[85] Candidates must have purely spastic cerebral palsy. Patients with ataxia, athetosis, dystonia, or rigidity are not candidates. Candidates should have no fixed contractures, and they must be able to ambulate without relying on spasticity for strength. There should be no weakness in the antigravity muscles or trunk musculature.[338,485] Finally, the patients must have supportive families and be able to cooperate with the postoperative physical therapy. Patients who fulfill all criteria are likely to have a good result from rhizotomy, as they are functional, intelligent, and mildly affected. It follows that these patients might also be the best candidates for orthopaedic surgery rather than rhizotomy, a debate that still remains unresolved.

Studies have shown decreased tone and increased joint range of motion following rhizotomy.[337,449] Peacock and Staudt found that 82 percent of patients continued to improve 3 to 7 years postoperatively.[337] Studies using gait analysis have shown improvements in sagittal plane hip, knee, and ankle motion following rhizotomy.[473,485] These improvements are seen at 1 year following rhizotomy, with little change occurring between 1 and 2 years postoperatively.[472] However, a recent study has shown that the improvements seen persist even at 10-year follow-up.[455] Because the hip flexors are more proximally innervated, anterior pelvic tilt is usually increased following rhizotomy.[55] Carroll and associates found that 29 of 112 patients who underwent rhizotomies used fewer walking aids postoperatively, and three who were unable to walk could do so following surgery. There were no objective improvements in ambulatory status in the remaining patients, although all had decreased spasticity.[80]

There is also an unexplained improvement in upper extremity use following lumbar rhizotomy.[11,263] Thirty-four percent experience an improvement in speech.[448] These suprasegmental improvements are theorized to occur as a result of the decrease in excitatory activity entering the spinal cord through the posterior roots from the lower limbs and spreading throughout the spinal cord along the propriospinal tracts.[448] Improvements in cognition and attention have also been described.[98]

Outcome studies using validated measurement tools are being performed to assess functional results following rhizotomy. In a small group of patients, self-care, mobility, and social functional skills were thought to improve when assessed by the PEDI tool.[53] A similar prospective study found that mobility and self-care improved following rhizotomy in patients with spastic diplegia, but not in those with spastic quadriplegia.[139] Other studies have used the gross motor functional measure (GMFM) to document functional improvement.[284,495]

Complications can occur from rhizotomy surgery.[450] As in any major surgery in this patient population, postoperative pulmonary complications occur frequently. Sensory disturbance has been described in patients who had rhizotomies as a result of sacrifice of some sensory rootlets.[2] Up to 40 percent of children experience painful postoperative dysesthesias of the legs. There is a risk of a neurogenic bladder developing, with inability to void. Patients at greatest risk are those who have spastic bladders preoperatively, a condition manifesting with frequent urinary tract infections, constipation, and daytime incontinence.[1] The changes in bladder function are usually transient.[448]

Orthopaedic complications are related to residual contractures, hip subluxation, and spinal deformity. Subsequent orthopaedic surgery is required in over half of patients who have rhizotomies, and Oppenheim suggests waiting 6 to 12 months after a rhizotomy to release contractures.[322] Carroll and colleagues found that 65 percent of patients who underwent a rhizotomy needed subsequent orthopaedic surgery.

Surgical correction of planovalgus deformity with the Grice procedure or lateral column lengthening was required in 37 percent of children, but only in those who became hypotonic following rhizotomy.[80] In the only study that has compared patients who underwent orthopaedic releases with patients who underwent rhizotomy, subsequent orthopaedic procedures were required in 62 percent of the rhizotomy group and 44 percent of the orthopaedic surgery group at an average follow-up of 4 years. The authors warned that follow-up to maturity is needed before the comparison can be valid. They also cautioned that the indications for surgery for the two groups were different and that the series was not randomized. It is difficult to compare the success rates of the two treatment courses in two different populations of children.[272]

In the series reported by Carroll and colleagues, hip reconstruction for subluxation was performed in 25 percent of children who had undergone a rhizotomy.[80] Greene and colleagues raised awareness of postoperative hip subluxation following rhizotomy in 1991, when they described six patients (treated at different centers) in whom progressive subluxation developed in the first postoperative year.[184] Park and colleagues followed up with a prospective series of 67 children with preoperative and postoperative radiographs of the hips. They found that the migration index increased in only 8 percent of hips after rhizotomy, and in 17 percent the coverage improved.[332] Unfortunately, in 20 of the 67 patients, follow-up radiographs were obtained between 6 and 10 months following rhizotomy. Longer follow-up is likely to reveal more cases of subluxation.

When femoral osteotomy is performed for progressive subluxation in patients who have undergone a rhizotomy, there is a peculiar predisposition toward developing heterotopic ossification. Ossification should be suspected in patients who lose range of motion postoperatively.[335]

A mechanism whereby subluxation might develop following rhizotomy has been proposed. The hip flexors are innervated by the L1 and L2 roots. Rhizotomy sections posterior rootlets, starting proximally at L2. The hip extensors are more distally innervated. There is greater denervation of the hip extensors than of the hip flexors following rhizotomy. Because spasticity in the iliopsoas contributes to hip instability, preservation of greater hip flexor tone can lead to hip subluxation in predisposed patients.[184]

Increased hip flexion in the presence of weak hip extensors can also lead to hyperlordosis of the lumbar spine after a rhizotomy (Fig. 24–78). Crawford and colleagues described two nonambulatory patients in whom extreme lumbar lordosis that interfered with sitting developed after rhizotomy.[100] Six additional patients with hyperlordosis or lordoscoliosis were described by Mooney and Millis. Five of the six patients were either nonambulatory or minimally ambulatory at the time of rhizotomy.[296] The surgical correction of this deformity is very difficult, and the risk-benefit ratio of rhizotomy in the nonambulatory population should be considered.

Spondylolysis, spondylolisthesis, and scoliosis have also been seen in patients who have undergone rhizotomies (Fig. 24–79). Peter and colleagues found an abnormally high rate of isthmic spondylolysis in patients who had rhizotomies.[349] Spondylolysis and spondylolisthesis occurred in 20 percent of their patients, but were asymptomatic.[347,348] Lumbar spinal stenosis has been described in two patients many years after a rhizotomy.[175]

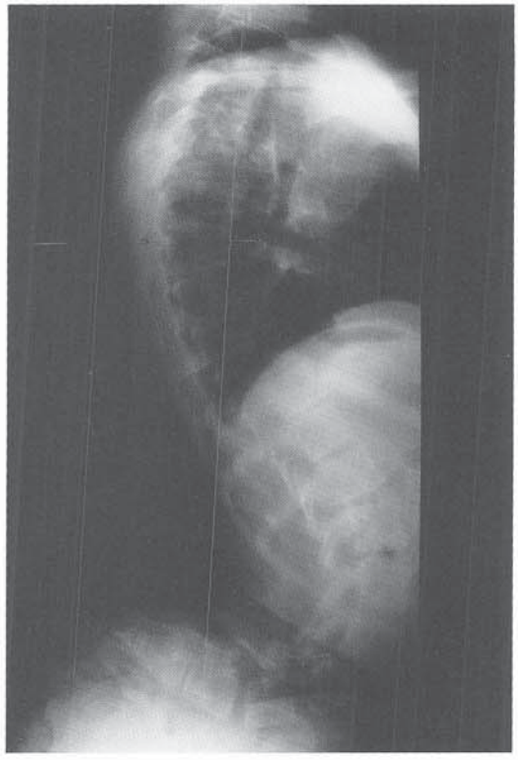


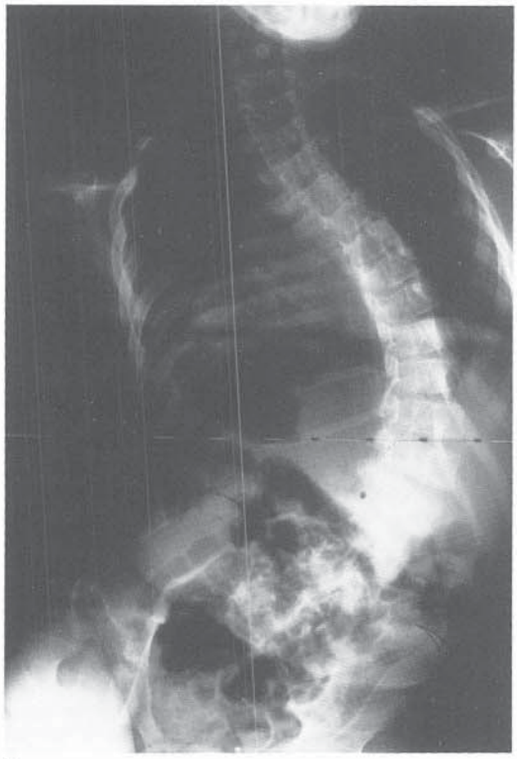
FIGURE 24–78 Hyperlordosis in a 12-year-old nonambulatory boy who underwent a posterior rhizotomy at age 7.

FRACTURES

Long bone fractures occur with frequency in patients with cerebral palsy.[67,253,258] Bone mineral density, measured with dual energy x-ray absorptiometry, is decreased in patients with cerebral palsy, with the most significant decreases seen in nonambulatory patients with poor nutrition.[202] Low levels of vitamin D have been documented in up to 42 percent of children with severe cerebral palsy,[253] but others did not find that the vitamin D level correlated with osteopenia or osteomalacia.[201] Hypovitaminosis D has been seen with increased frequency in patients who are on anticonvulsant medication.[18,253,254] Lack of exposure to sunlight has been implicated as yet another cause of rickets and fractures in institutionalized patients.[301]

Up to 74 percent of fractures occur in the femur, particularly at the supracondylar level.[67] Factors that are associated with an increased tendency for fracture are joint stiffness and recent surgery. Pritchett found a 20 percent incidence of femoral fracture in patients with untreated hip dislocations.[363] Yet patients who have undergone hip surgery are most likely to experience femoral fractures during the months following cast removal. A recent study found that 29 percent of nonambulatory children who were operated on for hip instability sustained a femur fracture within 3 months following cast removal.[454]

Treatment of osteopenia begins with vitamin D supplementation. Administration of bisphosphonates has been shown to increase bone mineral density in some patients



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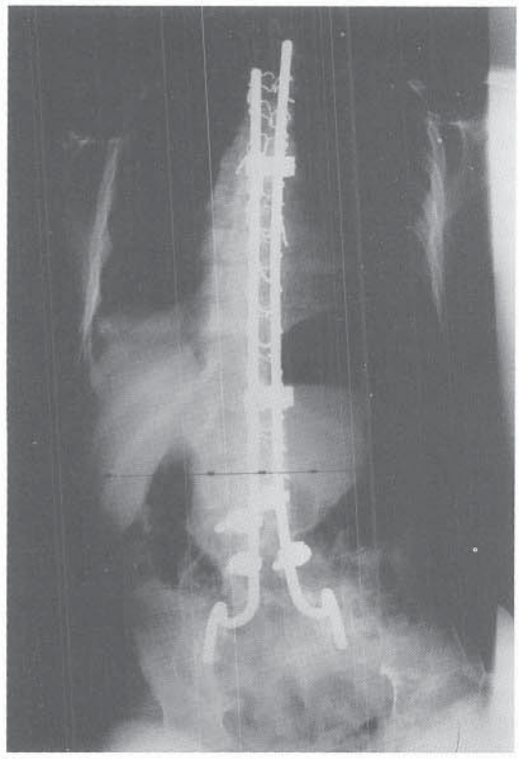

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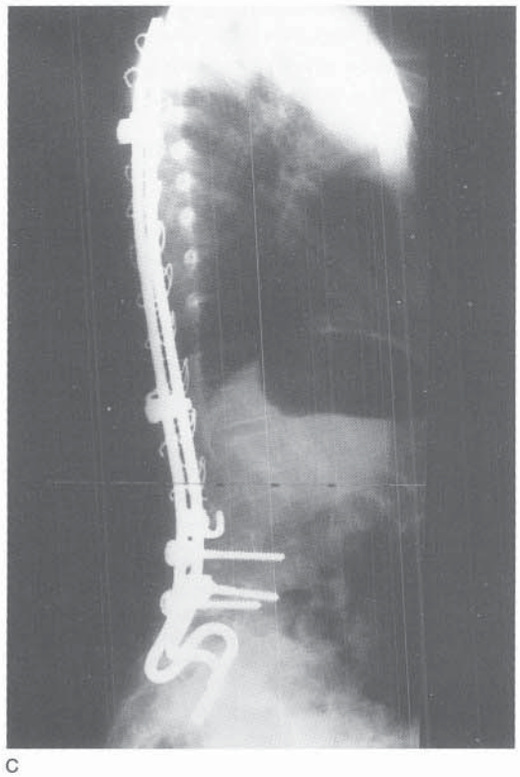

C

FIGURE 24–79 **A,** AP sitting radiograph of a 13-year-old child with scoliosis who has undergone a rhizotomy. Note the marked pelvic obliquity. **B** and **C,** Posterior spinal fusion using sublaminar wires proximally and pedicle screws in the previously operated area was performed. Pelvic obliquity was addressed using Dunn-McCarthy rods.

who sustain multiple fractures.[426] Treatment of fractures is usually by cast immobilization.[253,445] As immobilization leads to further demineralization, the time spent in cast should be kept to a minimum. Femoral shaft fractures are difficult to manage in traction[281] and may require internal fixation with plating or intramedullary devices.[247,454]

Orthopaedic surgeons who treat children with cerebral palsy should be aware that there is an increased incidence of child abuse in this patient population. Abuse may be the cause of the cerebral palsy, but there is an equally large proportion of patients who are abused following the diagnosis of cerebral palsy as a response to the diagnosis.[127] Suspicion should be increased whenever the history of the injury is peculiar.

OUTCOME ASSESSMENTS

The recent focus in medical research has been on the documentation of outcome following intervention, whether medical or surgical. Goldberg published a thought-provoking summary of the status of outcome analysis in cerebral palsy in l991. Outcome studies must address three areas: the technical outcome of the procedure, an assessment of functional health status, and patient satisfaction.[173] Current methods of assessing surgical outcome are gait analysis, the GMFM (gross motor function measure), and the WEE-FIM (a pediatric measure of functional independence).[107,190] The latter two are assessments generally administered by therapists of the abilities of the patient in crawling, running, or fine motor skills. In cerebral palsy in particular, many widely differing forms of treatment are available, all of which change the child without curing the disease. In the future, orthopaedic surgeons treating children with cerebral palsy can expect to see more objective documentation of how each intervention changes the abilities of the child.

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Rett Syndrome

Rett syndrome is a neurodegenerative condition seen in girls that consists of mental retardation, seizures, and characteristic wringing of the hands. The International Rett Syndrome Association has established the following criteria for the diagnosis of Rett syndrome: normal prenatal and perinatal

period; normal development through the first 6 months of life; normal head circumference at birth, with subsequent deceleration of head growth; loss of purposeful hand skills; severely impaired language; apparent severe mental retardation; and gait apraxia. Supportive criteria include respiratory dysfunction, seizures, spasticity, scoliosis, and growth retardation.[1] Loss of hand function and regression in development may be noted as early as 6 months of age to 18 months of age, although a definitive diagnosis usually cannot be made until 2 to 5 years of age.[2] After age 5 years, the disease becomes relatively static.[15] The patients' behavior resembles autism, and they scream and babble.

Rett syndrome is believed to be an X-linked dominant condition that is lethal in males.[1,14] Familial recurrence is rare but does occur on occasion in sisters.[19] Localization of the gene is underway.[16,18,19]

Hagberg divided the clinical course of the disease into four stages. The first stage, termed the early-onset deceleration stage, is characterized by hypotonia, deceleration of brain growth, and lack of developmental progress. This stage typically occurs between ages of 6 and 18 months. The stage of rapid destruction occurs between ages 1 and 3 years and is characterized by developmental regression, loss of purposeful hand function, and autism. The third stage, the pseudostationary stage, occurs between ages 2 and 10 years. During this period, seizures, ataxia, and dementia occur. Finally, the late motor deterioration stage occurs after age 10 years and is characterized by the development of scoliosis, muscle atrophy and contractures, and upper and lower motor neuron signs.[7]

Conditions from which Rett syndrome is to be distinguished include cerebral palsy, developmental delay, autism, and psychological disturbance. Patients are often misdiagnosed initially and are usually labeled as having cerebral palsy.[5,13]

MRI of the brain shows a global reduction in gray and white matter. There is a peculiar radiographic finding of short fourth metatarsals and short ulnas in 56 percent of girls.[4] Other radiographic findings include osteopenia (which may lead to fractures)[6,13] and scoliosis.

Orthopaedic manifestations of the disease are most commonly spasticity that initially affects gait, joint contractures, and scoliosis.[17] Spasticity may lead to joint contractures, and a small group of patients may benefit from soft tissue surgical procedures such as heel cord lengthening.[5] Coxa valga is seen in all patients.[13] Hip instability may also occur.[9]

Scoliosis occurs in 45 to 64 percent of patients with Rett syndrome.[8,13] The average age at onset is 8 years (Figs. 24–80A to C).[10,12] However, scoliosis may develop in very young children. Curves are long and sweeping, with the apex of the deformity usually in the thoracolumbar spine.[5] Progression of the curve is the rule[2,8,13] and occurs more rapidly than in idiopathic scoliosis (Fig. 24–80D).[12] Orthotic management may delay surgery but does not control progression.[10,11] Spinal fusion with segmental instrumentation is necessary for large progressive curves (Fig. 24–80E and F).[11]

Bracing has been largely unsuccessful in adolescent patients.[2]

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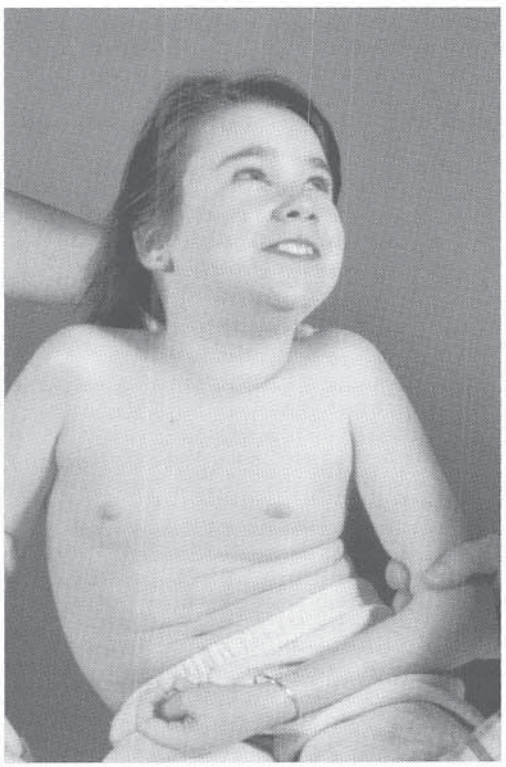
Hereditary Spastic Paraparesis

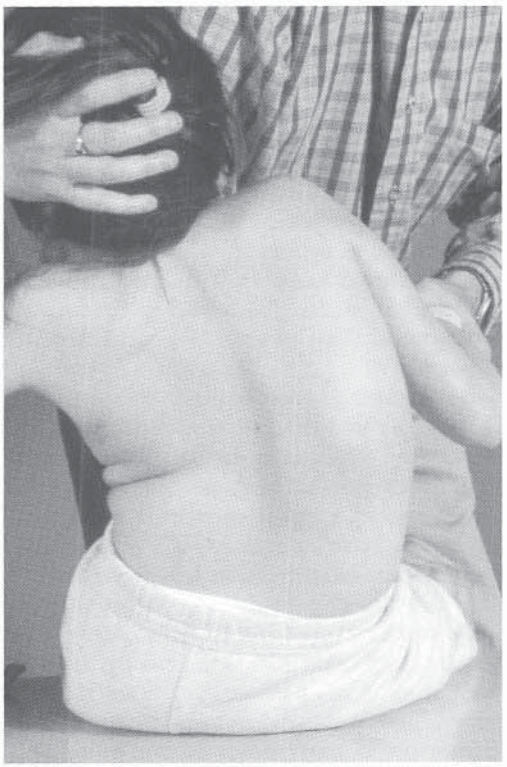
Hereditary spastic paraparesis is a neurodegenerative condition characterized by spasticity in the lower extremities with sparing of the upper extremities and cranial nerves. It is one of the more common progressive neurodegenerative diseases seen in children,[7] and has been seen regularly in our pediatric orthopaedic population. There are two forms, termed pure and complicated.[1,10] In pure hereditary spastic paraparesis, spasticity is the only neurologic finding. Dementia may occur in late adulthood.[15] In the complicated form, mental retardation, bulbar involvement, nystagmus, and dysarthria are also present.

Hereditary spastic paraparesis is usually transmitted as an autosomal dominant trait with complete penetrance, although X-linked inheritance has also been described.[5] Four genetic loci have been isolated, the most common of which is the 2p locus of chromosome 2.[4,11,14] There may be genetic anticipation, with offspring of affected parents having earlier onset of symptoms.[12]

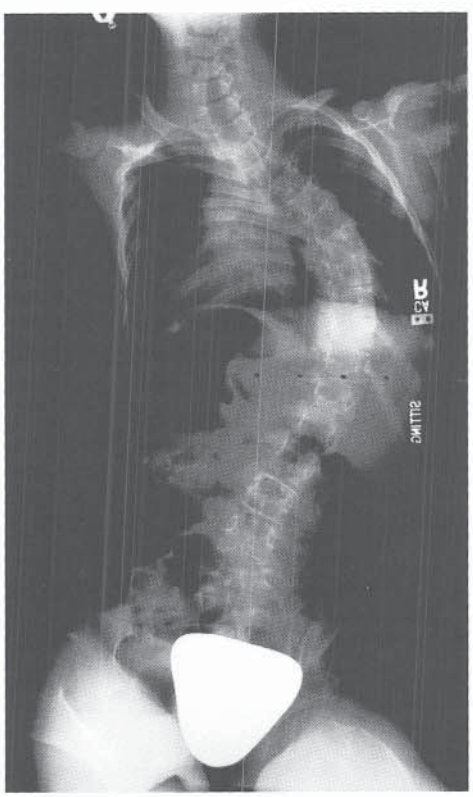
Patients with hereditary spastic paraparesis present with lower extremity spasticity and an abnormal gait. Toe-walking is a frequent complaint. Affected children may be delayed in walking, and clinical symptoms are usually present by age 3 years.[1] Other patients may be asymptomatic until adulthood.[9] Children with hereditary spastic paraparesis are often misdiagnosed initially as having cerebral palsy.[6,8] Patients with a family history of cerebral palsy should be sus-



A

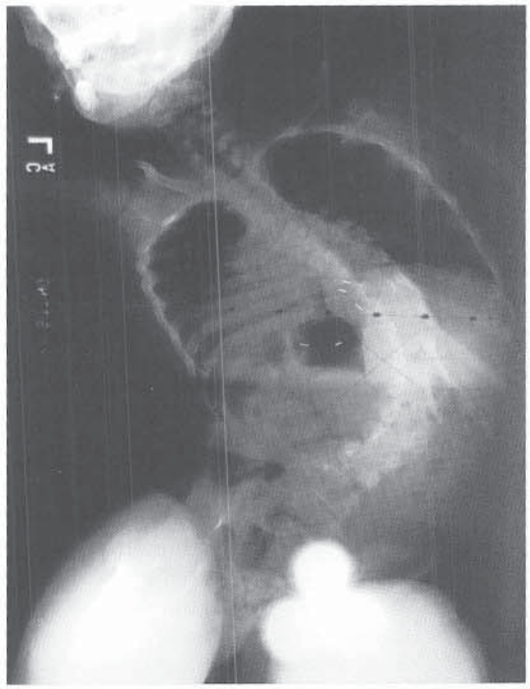


B

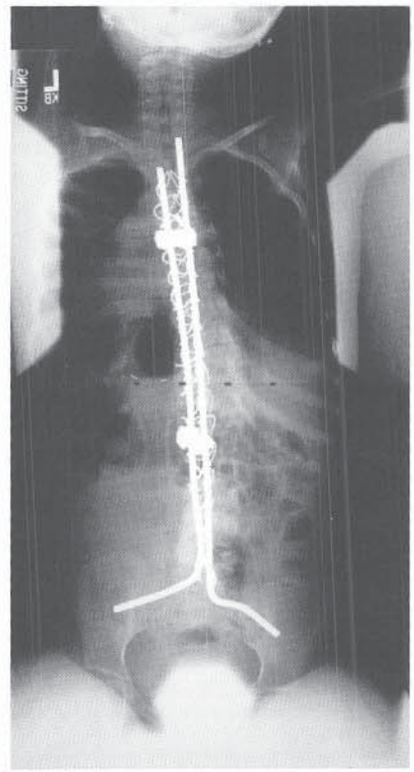


C

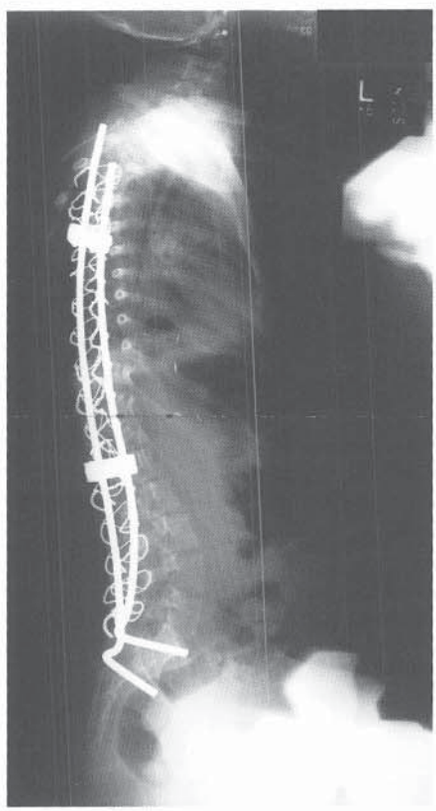
FIGURE 24–80 **A** and **B,** Clinical appearance of girl age 10 years 7 months with scoliosis due to Rett syndrome. **C,** Radiograph obtained at age 7 years 9 months.



D



E



F

FIGURE 24–80 *Continued* **D,** The curve had worsened by age 10 years 6 months. **E** and **F,** Posterior spinal fusion with Luque-Galveston instrumentation was performed.

pected of having hereditary spastic paraparesis, particularly if the upper extremities appear to be spared.

Physical examination reveals spasticity in the legs. Deep tendon reflexes are abnormally brisk, and the plantar reflexes are extensor.[8] Contractures, particularly of the Achilles tendon, may result from the increased tone.

Histopathologic examination reveals degeneration of the corticospinal tract and posterior columns, as well as degeneration of the spinocerebellar tracts in approximately 50 percent.[2] This can lead to subclinical sensory disturbances in patients with hereditary spastic paraparesis, which can be identified via somatosensory-evoked potentials.[3,13] The spinal cord degeneration is more advanced distally in the lumbar spine, with loss of myelin sheaths, leading to the predominance of lower extremity involvement.[8]

Orthopaedic treatment may be necessary if contractures are present, and resembles the management of cerebral palsy.

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Ataxia Syndromes

FRIEDREICH'S ATAXIA (HEREDITARY SPINOCEREBELLAR ATAXIA)

Friedreich's ataxia, first described in 1863,[10] is the most common of the hereditary ataxias, occurring in approximately 1 in 50,000 live births.[11] The disease is usually transmitted by an autosomal recessive gene, although autosomal dominant transmission has been reported. Genetic linkage analysis has localized the abnormal gene on chromosome 9.[4] The genetic defect is a triplet repeat expansion in the area of chromosome 9 responsible for encoding the mitochondrial protein frataxin.[21] The clinical severity of the disease is linked to the size of the triplet repeat.[6,15] In some families, affected persons may experience a milder form of Friedreich's ataxia. Prenatal diagnosis of the disease is available.[25] A variant of Friedreich's ataxia that is responsive to vitamin E has been described.[3,7,16]

In Friedreich's ataxia, both the cerebellar and the spinal cord pathways are involved. In the cerebellum, there is atrophy of the Purkinje's cells and the dentate nuclei. Changes may also occur in the brain stem. Degeneration of the corticospinal tract may occasionally occur above the level of the medulla and involve the cerebral cortex. In the spinal cord, there are degenerative changes in the dorsal and ventral spinocerebellar tracts, the corticospinal tracts, and the posterior column. The anterior horns are usually normal.

Clinical Features. A triad of clinical signs and symptoms is classically associated with the disease: ataxia, which is normally the presenting symptom; areflexia of the ankles and knees; and an extensor Babinski response.[24] The onset of symptoms usually occurs in childhood between 7 and 15 years of age,[12] with an average age at presentation of 12 years.[9] The onset often is insidious, making it difficult to precisely pinpoint when the condition first manifested.

An unsteady gait is almost always the first symptom. The child has a tendency to stagger and fall and has difficulty making sudden turns. The unsteady gait is more pronounced when the child attempts to walk in the dark. Over a period of years, the symptoms progress and ataxia of the upper limbs develops. The patient is usually not able to perform heel-to-toe walking. Stance is unsteady, and Romberg's sign is positive. The patient may demonstrate heel-to-shin ataxia and, later, finger-to-nose ataxia. Rapid alternating movements of the hands become more difficult for the patient to perform and gradually slow down.

Deep tendon reflexes usually are absent very early in the course of the disease, with areflexia one of the hallmarks of Friedreich's ataxia. Babinski's reflex becomes extensor. On sensory examination, position and vibration sense and two-point discrimination are lost. Later, as the disease progresses, the patient's speech becomes slurred, and dysphagia may become a serious problem.[20] Rotatory or horizontal nystagmus develops, and head tremor may be noted. Muscle weakness occurs symmetrically and is first apparent in the proximal muscles, such as the hip extensors.[18] Weakness first occurs in the lower extremities and then progresses to the upper extremities.[2]

Certain orthopaedic deformities are characteristic of Friedreich's ataxia. The most common is scoliosis, which occurs in 80 to 100 percent of patients.[13,17] The curves are most often located in the thoracic or thoracolumbar area and more closely resemble idiopathic curves than the long, sweeping neuromuscular curve patterns with associated pelvic obliquity.[1,5,17] Approximately two-thirds of patients who have scoliosis will have coexisting increased kyphosis.[5,17] Cavus deformity of the feet may be the presenting symptom

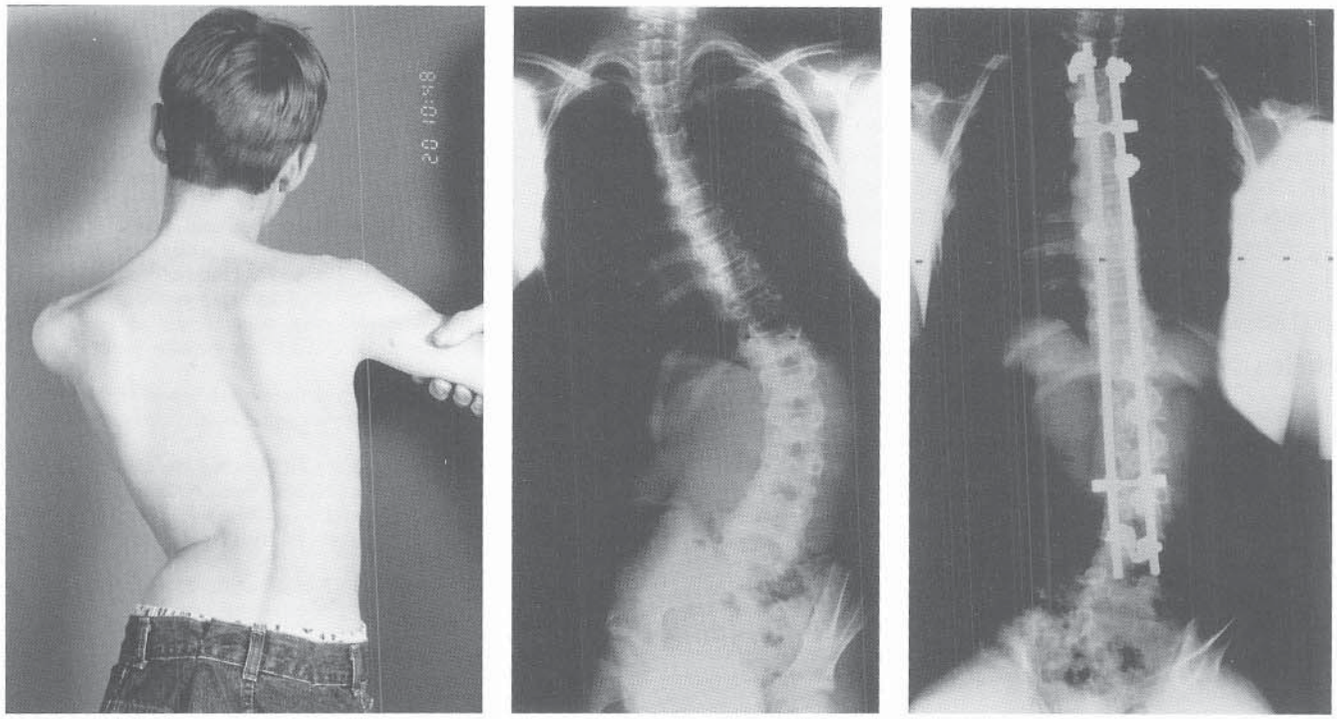

FIGURE 24–81 **A,** Clinical appearance of a 14-year-old boy with scoliosis secondary to Friedreich's ataxia. Because of poor balance he needed his father's assistance to stand for the photograph. **B,** Preoperative radiograph showing severe scoliosis and trunk decompensation. **C,** Radiograph obtained 6 months following posterior spinal instrumentation with TSRH instrumentation. The trunk balance has been improved.

for some patients. Early in the disease, the cavus is flexible, but as the disease progresses, the cavus becomes associated with varus and loses flexibility. Muscle imbalance, specifically peroneal muscle weakness, has been documented by Makin in most patients with cavus feet due to Friedreich's ataxia.[18] Diabetes mellitus has been seen in conjunction with the disease in approximately one-fourth of patients.[8,24] Cardiomyopathy develops in patients with Friedreich's ataxia, and eventual cardiac failure leads to a diminished quality and length of life.

Diagnosis. The presence of ataxia, scoliosis, and pes cavus, along with a positive family history, leads to the diagnosis of Friedreich's ataxia. Muscle biopsy shows denervation atrophy of both small and large muscle fiber groups. CPK levels are normal. Spinal tap reveals normal CSF. ECG findings include conduction defects, with bundle-branch block or complete heart block, and T-wave inversion. Ventricular thickening is notable on echocardiography.[22] EMG shows loss of motor units and an increase in polyphasic potentials. Nerve conduction studies show only a slight decrease in motor fiber conduction velocity but marked decrease in sensory action potential, unlike in the hereditary motor sensory neuropathies (e.g., Charcot-Marie-Tooth disease), in which the motor conduction velocity is significantly diminished. MRI of the spinal cord shows a decrease in the AP diameter of the cord and changes in the posterior and lateral columns.[19]

Orthopaedic Management. There is no definitive treatment for patients with Friedreich's ataxia. Orthopaedic treatment includes correction of cavus foot deformity (see Chapter 22, Disorders of the Foot). Surgery is rarely recommended for patients able to ambulate with AFOs. Aggressive surgical management of symptomatic pes cavus, though, can prolong ambulation. In a series of 34 patients, Makin reported improvement in ataxia after surgery.[18] Surgery usually consists of Achilles tendon lengthening, posterior tibialis tendon surgery (lengthening versus transfer), or triple arthrodesis.[24]

Posterior spinal fusion with instrumentation is the treatment of choice for progressive scoliosis[5,14] (Fig. 24–81). Curves should be fused if there is progression to 60 degrees or greater in magnitude. Spinal fusion should span the thoracic and lumbar spine,[23] as fusing short has been found to lead to further progression of the curve. A few curves stabilize at less than 40 degrees of magnitude and do not require surgery. Scoliosis progression despite brace wear has been reported.[5] In addition, bracing is poorly tolerated by patients and usually interferes with their ability to walk.

Prognosis. The clinical course of classic Friedreich's ataxia is a slow but relentless progression. The earlier the onset of disease, the poorer is the outcome. The age at onset and the rate of progression usually are similar in affected siblings. By 20 years of age, the ataxia is severe, and by the second or third decades of life, the patient usually requires a wheelchair. The average age at which patients were unable to walk was 26 years (±7.8 years) in a large Italian series,[9] and 20 years in a pediatric orthopaedic center.[5] Scoliosis usually is progressive, but if a curvature stabilizes with a Cobb angle of less than 40 degrees, progression rarely occurs. The younger the age of the patient at the onset of scoliosis, the

more likely it is that the curve will progress and require surgical intervention.

Progression of scoliotic curves has been linked to onset of ataxia at 10 years of age or less and age at onset of scoliosis less than 15 years.[17] The average duration of the disease from onset until death is 25 years. Death from hypertrophic cardiomyopathy or pneumonia usually occurs by 38 years (range, 21 to 69 years).[13] Milder forms of the disease are seen on rare occasions.

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