Evaluation and Treatment of Hip Dysplasia in Cerebral Palsy

David A. Spiegel, MD*, John M. Flynn, MD

Division of Orthopaedic Surgery, Children’s Hospital of Philadelphia, 2nd Floor Wood Building, 34th Street and Civic Center Boulevard, Philadelphia, PA 19104, USA

Hip problems are commonly identified in patients with cerebral palsy, particularly those with a greater severity of neurologic involvement. Although ambulatory patients with hemiplegia and diplegia typically have torsional abnormalities of the femur, nonambulatory patients with spastic quadriplegia commonly develop neuromuscular hip dysplasia. Pain in the hip region can be a frequent complaint in patients with spastic quadriplegia, and may or may not be secondary to hip subluxation or dislocation.

The focus of this article is the diagnosis and management of neuromuscular hip dysplasia, or “spastic hip disease.” Clinical concerns include difficulties with seating or positioning, problems with hygiene and personal care, and occasionally hip pain. The goals of treatment are to maintain a level pelvis; a balanced spine; and mobile, pain-free hips. The natural history is variable, and a large subset of patients develops progressive subluxation of one or both hips, often leading to dislocation. The current approach to treatment focuses on establishing an early diagnosis to prevent these complications. When an early diagnosis is made, nonoperative treatment can be tried to prevent or delay the progression of deformity by maintaining range of motion and decreasing spasticity. In patients with established subluxation or dislocation, goals include improving positioning and ease of care, and treating any discomfort. Surgical options depend on the stage of disease at diagnosis, and may include soft tissue lengthening; reconstructive surgery by soft tissue release combined with a femoral osteotomy with or without a pelvic osteotomy; and salvage procedures, such as resection realignment, or replacement of the femoral head and neck.

Pathophysiology

The progressive changes associated with hip dysplasia in patients with cerebral palsy result from the effects of neuromuscular imbalance on the growth and development of the hip joint [1–4]. The primary problem is spasticity and muscular imbalance, and the musculoskeletal manifestations are secondary. Soft tissue abnormalities include a muscular imbalance between the stronger flexors and adductors, and the weaker extensors and abductors. With growth, this dynamic imbalance results in myostatic contractures in adduction with or without flexion. These contractures then tether growth, creating progressive changes in the femur and the acetabulum.

Bony abnormalities include femoral torsion with or without coxa valga, and progressive deformity of the femoral head or acetabulum. Most patients have internal femoral torsion (excessive femoral anteverision) caused by inhibition of derotation of the proximal femur during growth. Normal children have approximately 40 degrees of femoral anteverision at birth, which progressively decreases to approximately 15 degrees in the adult. Children with spastic hip disease have a persistence of fetal anteverision, and many have a progressive increase in the degree of anteverision over time. Using a three-dimensional
finite element model, Shefelbine and Carter [5] predicted a decrease in anteversion angle of 2 degrees per 6 months under normal loading conditions, and an increase of 1 degree per 6 months under the loading conditions typically seen in cerebral palsy. Although many children have a normal femoral neck-shaft angle, a subset has coxa valga. Excessive anteversion may appear to be coxa valga on the anteroposterior radiograph, and a true anteroposterior with anteversion corrected is required to evaluate the neck shaft angle.

A flexion-adduction contracture also shifts the center of rotation of the hip from the femoral head to the lesser trochanter, and the proximal femur is gradually displaced upward and outward [2]. As the femoral head displaces, pressure is concentrated along the posterolateral margin of the acetabulum, which progressively flattens. Asymmetric pressure may deform the femoral head, leading to degeneration of articular cartilage. Pathologic specimens have demonstrated wedging of the epiphysis with focal deformation of the femoral head [6].

In a small subset of patients (1%–2%), usually with extension posturing and hypotonia, the progressive subluxation (or dislocation) is anterior, as is the acetabular deficiency [7,8]. Selva and coworkers [8] found two types of deformity: those with an extension-adduction-external rotation contracture at the hip associated with an extension contracture at the knee, and those with an extension-abduction-external rotation contracture at the hip associated with a flexion deformity at the knee. Fifty percent of patients developed hip pain.

Physical examination

A comprehensive examination includes the spine, hips, and lower extremities. Patients should be evaluated both in their wheelchair and on the examining table. The range of motion at the hips (and lower extremities) is assessed. The spine should be inspected for scoliosis, and if present the location and flexibility are recorded. Sagittal spinal alignment is assessed, and any pelvic obliquity should be noted. On bench examination, a hip flexion deformity can be assessed with the Thomas test. Hip abduction is generally tested with both the hip and the knee in extension. Hip rotation is best assessed with the patient prone. Popliteal angles should also be measured, and the degree of spasticity should be quantified.

With the pelvis viewed as the base of support, the relationship between the hips (infrapelvic alignment) and the spine (suprapelvic alignment) in both the coronal and sagittal plane should be evaluated. In the coronal plane, any pelvic obliquity should be correlated with both the spine and range of motion at the hips. Although the relationship between infrapelvic deformity and scoliosis is debated, several reports have suggested that a primary infrapelvic deformity (adduction contracture) results in pelvic obliquity, and may precede the development of a compensatory lumbar scoliosis with the apex toward the low side of the pelvis [9,10]. On occasion, a lumbar scoliosis with its convexity opposite to the low side of the pelvis may be seen. A primary suprapelvic obliquity (structural scoliosis) also causes pelvic obliquity. In the sagittal plane, although an increase in lumbar lordosis compensates for a hip flexion contracture (causes the pelvis to tilt anteriorly), a lumbar or thoracolumbar kyphosis compensates for an extension contracture of the hip (tight hamstrings, pelvis tilts posteriorly). In these cases, a flexible suprapelvic deformity compensates for the infrapelvic deformity. Although flexion contractures at the hips are less commonly associated with sitting imbalance, significant extension contractures may cause the patient to slide forward in the wheelchair.

In the coronal plane, there may be a windswept appearance of the hips with adduction of one hip and abduction of the other hip, termed the “wind-blown hip” [11]. Electromyographic data suggest that the adductors are active on both sides, whereas the abductors are overactive only on the abducted side [12]. The relationship between suprapelvic and infrapelvic obliquity remains controversial, and the available evidence suggests that infrapelvic obliquity (asymmetric soft tissue contracture) precedes the development of suprapelvic obliquity (scoliosis) [9–11].

Radiologic evaluation

Plain radiographs are commonly used to monitor patients with spastic hip dysplasia. A standardized technique should always be used, with the patient supine and the hip held in neutral adduction-abduction. In patients with a flexion deformity of the hip, the pelvis tilts anteriorly when the hips are extended to obtain the radiograph, which produces an inlet view of the pelvis and makes interpretation difficult. As such, the technician may need maximally to flex the contralateral hip and knee to flatten the lumbar prodosis. The next shaft angle usually appears increased, and “apparent” coxa valga may result from excessive femoral anteversion. Rotation of the hip to correct the excessive anteversion is required to assess accurately the next shaft angle. True coxa
valga may also be seen in patients with spastic hip disease.

The migration index is valuable to follow patients for progressive subluxation of the hip. This is calculated by dividing the width of the femoral head that lies outside the lateral margin of the acetabulum (Perkin's line) by the total width of the femoral head (Fig. 1) [13]. In patients without hip disease, the migration percentage is 0% for those less than 4 years of age, and 5% for those 12 to 16 years of age. In the population with cerebral palsy, normal is believed to be less than 30%, whereas subluxation is greater than 30%, and dislocation is greater than 90%. As for other radiographic measurement parameters, there is a normal range of variability. The same technique should be used for all films, and ideally the same observer measures the radiographs, with prior films available for comparison. Reimers [13] believed that a change of 10% represented a true change. More recently, Faraj and coworkers [14] found that for a single observer performing repeated measurements, a 95% confidence interval for a true change was 13%. Parrott and coworkers [15] also reported on the intraobserver (±8% for a true change) and interobserver (±11.6% for a true change) reliability for the migration percentage.

The acetabular index is commonly used to evaluate acetabular dysplasia. The normal acetabular index is 30 degrees at less than 1 year of age, 25 degrees at 1 to 5 years of age, and approximately 20 degrees in an adult. Kay and coworkers [16] found the 95% confidence interval for a true change to be 8 degrees, whereas Spatz and coworkers [17] found an interobserver variability of ±3 degrees and an intraobserver variability of ±3.8 degrees. These exclude variability in pelvic position between radiographs. In spastic hip subluxation, the acetabular index is generally in the range of 40 degrees when the migration index is 50%. Findings on plain radiographs suggest that the acetabular index is normal until approximately 30 months of age, and then becomes progressively higher [18]. It remains unclear whether the acetabulum is shallow [19] or normal [20] in depth. Cooke and coworkers [81] suggested that an acetabular angle of greater than 30 degrees was predictive of dislocation. Although arthograms are not commonly used in the evaluation of spastic hip disease, Heinrich and coworkers [21] found that the ossification of the lateral pelvic cartilaginous anlage is retarded by progressive subluxation, and was the greatest in hips with a migration index of 52% to 68%.

The morphology of the acetabulum can also be assessed by the shape of the sourcil [22]. The lateral corner is well defined and lies below the weight-bearing dome of the acetabulum in a type I sourcil. In a type II sourcil, the lateral corner is turned upward and lies above the weight-bearing dome. A CT scan of the hips with three-dimensional reconstruction may help to define the nature and location of acetabular deficiency, and on occasion deformity of femoral head also may be identified [19,23,24]. It remains to be determined whether MRI may play a role in evaluating femoral head morphology and cartilage thickness in those subluxated or dislocated hips being considered for reconstructive versus salvage surgery. Although not routine, the authors recommended a CT scan in selected patients in whom acetabular morphology cannot be adequately assessed on plain radiographs.

**Natural history**

Although the prevalence of hip dysplasia with cerebral palsy varies from 2% to 75%, progressive subluxation is most common in patients with more profound degrees of neurologic involvement [25–28]. Lonstein and Beck [28] identified hip subluxation or dislocation in only 7% of independent ambulators, but in 60% of nonambulators. In the subset of patients who develop progressive dysplasia, the evolution from muscle imbalance to fixed con-
tracture, subluxation, and finally dislocation usually occurs over several years. Bagg and coworkers [29] found that hips that were stable at 18 years of age remained stable, but that hips with severe subluxation all progressed to dislocation. Miller and Bagg [30] found that although hips with a migration percentage less than 30% were at low risk, those with greater than 60% all became dislocated. The intermediate group had a 25% risk of progression. The results of these studies suggest that close clinical and radiographic follow-up (every 6 months) is required to identify that subset of patients at risk for the development of progressive subluxation or dislocation.

The natural history of the severely subluxated or dislocated hip has also been studied, with variable conclusions. Although several reports have suggested that subluxated or dislocated hips are commonly associated with pain or other problems [27,31,32], which has prompted the current trend toward early screening and aggressive treatment, others have disputed these findings.

Cooperman and coworkers [27] studied 51 dislocated hips with a follow-up of 18 years, and found that half of the dislocated hips were painful. Moreau and coworkers identified hip pain in 11 of 21 institutionalized adults with a dislocated hip [4]. Boldingh and coworkers [31] reviewed 160 patients and found that hip pain was associated with migration and deformity of the femoral head. In contrast, Pritchett [33] studied 80 patients and found that a dislocated hip predisposed to lower-extremity fractures, but did not cause pain, problems with perineal care, or decubitus. The same author reviewed 100 patients with an established dislocation, 50 of whom had surgery for the dislocation, and concluded that there was no difference in outcome, and that pelvic obliquity and scoliosis were related to the degree of neurologic involvement rather than the status of the hips [34]. Knapp and Cortes [35] studied 38 dislocated hips, and found that 18% were definitely painful, whereas 71% were not painful. Noonan and coworkers [36] studied 77 subjects with a mean age of 40 years, of which 15% of the hips were dislocated and 12% were subluxated. Both subluxation and dislocation were associated with radiographic evidence of arthritis, but not necessarily with hip pain. Hip pain and problems with perineal care were identified in patients with less than 30 degrees of hip abduction, with greater than 30 degrees of hip flexion contracture, and with windswept hip deformities.

Chronic musculoskeletal pain is a complaint in up to 67% of adults with cerebral palsy, most commonly in the low back, hip, and leg [37]. Pain in the hip region is commonly encountered in nonambulatory children with spasticity, with or without subluxation or dislocation of one or both hips. The relationship between hip pain and subluxation or dislocation remains elusive in both children and adults. Hodgkinson and coworkers [38] found that the prevalence of pain was 47.2% (tolerable in 35.6%) of 234 non-ambulatory adolescents, and that pain could be provoked, linked to position, or spontaneous.

Identifying the source of pain in the region of the hip remains a challenge. Many patients are unable to articulate what they are experiencing, and the clinician must rely on the perception of the parents or caregivers to help identify the source. Pain may be observed at rest, with certain positions, or with such movements as passive abduction. Presumably, pain may originate in the skin or subcutaneous tissues, the musculature surrounding the hip, the osteoarticular structures, or may be referred from another location. Intra-abdominal problems, such as appendicitis, an ovarian cyst, or endometriosis, may present as hip pain. As such, a careful history and physical examination, supplemented by imaging studies or diagnostic injections, may be required to establish the cause and guide treatment. Pain that occurs with passive stretching may be muscular in origin (spasticity with or without contracture), or potentially from hinging of the femoral head on the lateral margin of the acetabulum (or iliac wing) in patients with subluxation or dislocation (with or without breakdown of articular cartilage). Positional pain may result from asymmetric pressure on the skin and subcutaneous tissues, especially in patients with pelvic obliquity caused by suprapelvic (scoliosis) or infrapelvic deformities. Discomfort experienced at rest may be muscular (spasticity), or originate from the articular surface in patients with subluxation or dislocation. On physical examination, the patient’s range of motion and response to passive stretching should be assessed.

If a specific pain generator cannot be established despite a careful evaluation, empiric treatment is instituted. Simple measures include avoidance of certain positions, modification of the wheelchair, and nonnarcotic analgesics. When spasticity is believed to play a role, treatment options include oral agents (baclofen, tizanidine); intermuscular injections (botulinum toxin type A, phenol); or intrathecal baclofen. The authors have occasionally performed a diagnostic hip joint injection with a local anesthetic when an intra-articular source is suspected. Further study is required to establish the most appropriate guidelines for the evaluation and treatment of hip pain, and to define better the relationship between radiographic subluxation or dislocation and pain in the pediatric and adolescent population.
Spastic hip disease leading to progressive subluxation with or without dislocation is common in nonambulatory patients with spastic quadriplegia. Although it is clear that a subset of patients with neuromuscular dysplasia have hip pain, the relationship between radiographic findings and clinical findings remains elusive. Patients with progressive dysplasia should be followed closely, and the current trend is to treat these patients early to prevent further subluxation and dislocation. Because hip pain may or may not relate to any observed radiographic changes, a thorough clinical evaluation is suggested to determine the source and to plan treatment.

Nonoperative treatment

An early diagnosis is facilitated by both clinical and radiographic screening, and when there is evidence to suggest that the progression to subluxation and dislocation may be delayed or prevented by early diagnosis and treatment [39,40]. Patients with quadriplegia and marked spasticity should be monitored closely. The authors typically evaluate these patients every 6 months, and focus on the range of motion at the hip. Hips that cannot abduct beyond 30 to 45 degrees (in extension) are believed to be at risk. A baseline radiograph is obtained at 12 to 18 months of age, and the frequency of follow-up radiographs is based on both physical and radiographic findings. When an abduction contracture is present, or the migration percentage is greater than 25%, radiographs may be obtained every 6 months.

The primary problem is in the nervous system, but most treatment strategies have been directed at the end organ. Most of these children receive physical therapy, one goal of which is to maintain the range of motion. Although the effects of physical therapy alone on the natural history of a spastic hip disease remain unknown, efforts to maintain muscle length and prevent contractures should be encouraged. Abduction bracing may be used as an adjunct to stretching, but if used aggressively may cause wind-blown hips or hyperabduction deformity. Achieving compliance with a bracing program may be difficult. Hankinson and Morton [41] performed a prospective trial using a lying hip abduction (20 degrees) system in 14 children with less than 30 degrees abduction before treatment. Six of the 14 abandoned the treatment because of sleep disturbance, and one required surgical intervention. In the seven patients who completed the study, there was an improvement in the migration percentage. Further research is required to determine whether abduction splinting, with or without other therapies, can play a role in altering the natural history of spastic hip disease.

Although passive stretching and splinting are preventive measures directed at the end organ, several recent investigations have addressed the primary problem (spasticity). Intermuscular injection of the adductors with botulinum toxin A has been attempted, and although the psoas is also believed to play an important role in disease progression, the injection is technically more difficulty and has not yet achieved widespread acceptance. The injections need to be repeated at 3- to 6-month intervals, and are accompanied by passive stretching exercises with or without abduction splinting. Pidcock and coworkers [42] looked at the hip migration percentage in 16 patients (9–43 months of age) treated with botulinum toxin A injections to the adductors. Thirteen of these hips started with a migration percentage greater than 30%. At 14 to 49 months follow-up, 18 of 32 patients had a change of 10% or less in the migration percentage, and the greatest improvement was in patients less than 24 months of age with a migration percentage greater than 30%. Boyd and coworkers [43] compared patients treated with botulinum toxin A injections and hip bracing with a group who were observed. At 3 years follow-up, surgery was performed in 47% of patients who did not receive botulinum toxin A, and in 27% of those who did receive botulinum toxin A. Longer-term study is required to determine whether intermuscular injections, with or without other methods of therapy, may alter the natural history. Continuous intrathecal baclofen infusion has been successful in reducing muscle tone in patients with cerebral palsy. In a prospective study by Krach and coworkers [44], 33 patients had an assessment of the migration percentage before and after 1 year of treatment with intrathecal baclofen. A total of 73% of these patients were non-ambulatory, and some had had previous orthopedic surgery. The migration percentage remained stable in 91% of patients.

Operative treatment

With the goal of maintaining mobile, located hips to maintain seating balance and prevent pain, surgical treatment is performed in a number of patients with progressive subluxation or dislocation. Operative interventions include soft tissue lengthening [13,45–51]; hip reconstruction by a femoral osteotomy [25,26,52–54] with or without a pelvic osteo-
or salvage procedures, such as femoral head and neck resection, replacement, or re-

Soft tissue procedures have been recommended as a prophylactic measure against bony procedures when the hip is believed to be at risk (passive abduction less than 30–45 degrees, migration percentage greater than 25%), or when there is mild subluxation (migration percentage MP > 30%) without coexisting bony deformity. This option may also be considered in children with greater degrees of subluxation who are less than 4 years of age, given the higher risk of recurrence following bony surgery in this population, and as a means to improve motion in patients with greater degrees of deformity who are believed to be at too great a risk for bony surgery.

The goal of soft tissue surgery is to prevent progressive subluxation by rebalancing the muscles (decrease the deforming force in adduction with or without flexion) and restoring motion (relieve myostatic contracture). The specific surgical procedures have varied among published reports, but all authors agree that bilateral procedures should be used to decrease the risk of recurrence or imbalance. Standard components include an open myotomy of the adductor longus and the gracilis, and a partial myotomy of the adductor brevis with greater degrees of contracture. The goal is to achieve at least 30 degrees of passive abduction. Some authors perform routine release or lengthening of the iliopsoas [47,48], whereas others only perform this procedure in the presence of a myostatic contracture [13,45,51]. Obturator neurectomy (anterior branch) remains controversial given the risks of an extension and abduction contracture, and may be indicated in non-ambulators with severe spasticity [47,48,75]. An extension and abduction contracture may make it extremely difficult to sit, and may require additional soft tissue releases or shortening femoral osteotomies for adequate seating or positioning [3,8,76,77]. Most authors recommend splinting in abduction (approximately 30 degrees) postoperatively for a variable period of time [37]. Splinting in hyperabduction may predispose to an extension and abduction contracture, especially when an obturator neurectomy has been performed. Several authors have noted better results in patients less than 4 years of age at surgery [13,47], whereas others have suggested that the age at surgery has no bearing on the results [48,51]. Similarly, the migration percentage at the time of surgery may [45,47] or may not [48] predict the outcome.

Presedo and coworkers [48] reviewed the results of a standardized protocol in 129 hips (65 patients, 47 nonambulatory) with 10.8 years follow-up. The indication for an operation was a hip at risk (<45 degrees abduction, MP <25%), and the procedure included myotomy of the adductor longus and gracilis, a release or intramuscular lengthening of the iliopsoas, and an obturator neurectomy in non-ambulators. A proximal hamstring release was also performed on occasion. The mean age was 4.4 years, and the postoperative protocol included knee immobilizers and physical therapy. The results were graded as good in 49%, and the failure rate was 30%. Nineteen of 65 children required subsequent bony reconstructive procedures, and a second soft tissue release was required in 11 patients. Overall, the protocol was believed to be effective in 66% of cases. Ambulatory patients had a better outcome than nonambulators, and both the age at surgery and the migration percentage did not affect the outcome. The migration percentage at 1 year postoperatively was predictive of the long-term result. The authors suggest that with early screening and application of a standard protocol, osseous surgery should be required in only 5% of ambulatory and 45% of nonambulatory patients. Cornell and coworkers [45] reviewed 56 hips, and compared patients having a percutaneous release of the adductor longus and gracilis with those treated by an open release including an obturator neurectomy. Success was achieved in 83% when the migration percentage was less than 40%, and there seemed to be no difference between those treated by percutaneous or open techniques. This approach was successful in only 23% of those with a migration percentage of greater than 40%, and failed in all cases with a migration percentage greater than 60%. Reimers [13] found good results following soft tissue releases in 12 of 36 cases. Onimus and coworkers studied 40 hips who were treated at a mean of 4 years of age by myotomy of the adductor longus and gracilis, intramuscular lengthening of the iliopsoas, and obturator neurectomy [47]. At 3-year follow-up, the approach was successful in 67% overall, and in 90% of those less than 4 years of age with a migration percentage less than 33%. Turker and coworkers studied 90 hips treated by myotomy of the adductor longus (+brevis), gracilis, and release of the psoas (50%) with or without obturator neurectomy (or crushing the nerve for temporary denervation) at more than 8 years follow-up [74]. The approach was successful in 42% of patients, and the remaining patients either required additional surgery or developed a dislocation.

In summary, definitive recommendations regarding the efficacy of soft tissue releases are difficult to establish from the existing literature given variations in the patient population (ambulatory and non-
ambulatory); the indications; the technical details of
the procedure; and the postoperative protocols. The
results seem to be best when the procedure is per-
formed as a prophylactic measure; however, the
lack of a control group in any of these studies makes
it difficult to be sure. Stott and Piedrahita [49] pub-
lished an evidence-based report in which 27 studies
involving soft tissue releases for spastic hip disease
were scrutinized. Radiographic subluxation was
improved in 168 of 530 hips, and the migration
percentage was improved in 241 out of 467 hips.
Because of the presence of confounding variables
(heterogeneity in patient populations, variability in
surgical procedures), any conclusions are preliminary
and further research is needed to make more spe-
cific recommendations.

Osseous reconstruction

Hip reconstruction is considered in patients who
have failed soft tissue surgery, or in those who have
developed progressive subluxation or dislocation
(migration percentage greater than 40%–60%) with
or without bony deformity (femoral valgus, acetabu-
lar dysplasia). Contraindications to reconstructive
surgery include deformity of the femoral head or
degenerative changes, and in borderline cases (usu-
ally severe subluxation or chronic dislocation) in-
spection of the femoral head at the time of surgery
may be required to decide between reconstruction
and salvage. The procedures are tailored to the path-
ologic findings, and may include a soft tissue release; a
proximal femoral varus derotational osteotomy (with
or without shortening); a peri-iliac acetabuloplasty;
and an open reduction and capsulorrhaphy when
necessary (Fig. 2).

For patients presenting without marked deformity
of the acetabulum (acetabular index below 25–
27 degrees), a proximal femoral varus derotational
osteotomy is performed. Components of this proce-
dure include correction of the neck-shaft angle (coxa
valga); correction of femoral torsion (persistent fetal
anteversion); and shortening of the femur to reduce
the muscle forces acting on the hip. A preliminary
soft tissue release is performed when passive abduc-
tion is less than 45 degrees, because the osteotomy
decreases passive abduction. This typically involves
the adductor longus, gracilis, and occasionally some
fibers of the adductor brevis. If passive abduction
remains limited despite these measures, release of the
medial capsule may be required. Lengthening or
release of the iliopsoas is also performed. In non-
ambulators, the psoas tendon may be released when
removing a medially based wedge of bone during the
osteotomy. In ambulators, the insertion on the lesser
trochanter should be preserved, and an intramuscular
lengthening may be performed proximally. The
appropriate goal for the postoperative neck-shaft
angle depends on both age and ambulatory status.
If possible, femoral osteotomies should be delayed
until at least 4 years of age [52,76]. Mazur and
coworkers [76] suggest that remodeling may result in

Fig. 2. (A) This 7-year-old boy with spastic quadriplegia had progressive right hip subluxation despite a soft tissue release several
years ago. His migration percentage was greater than 70%, and coexisting acetabular dysplasia was present. (B) He underwent
bilateral femoral varus derotational osteotomies (family chose bilateral osteotomies to preserve limb length and thigh symmetry),
with the goal of a neck-shaft angle of 100 degrees. A volume-reducing pelvic osteotomy was also performed on the right.
a 30-degree increase in neck-shaft angle in children less than 4 years of age, and approximately 20 degrees in patients between 4 and 12 years of age. There was no difference between ambulators and nonambulators. In ambulators, the authors suggest keeping the neck shaft angle between 110 and 120 degrees, depending on the age of the patient, to minimize the chance of abductor insufficiency. In nonambulators, the ideal neck shaft angle varies between 90 and 110 degrees depending on age. Femoral torsion is corrected by externally rotating the distal fragment. Many surgeons choose to fix the osteotomy at a point when there is an even balance between internal and external rotation. Alternatively, a guide pin placed in the femoral neck may be used to estimate both the degree of torsion and the version following derotation. Care should be taken to avoid excessive correction, because retroversion may increase the risk of posterior subluxation or dislocation of the hip. Perhaps the most important component of this procedure is limb shortening, which normalizes the muscle forces acting on the hip. Although the varus osteotomy helps to an extent, it is often beneficial to remove additional bone from the femoral shaft before fixation. The popliteal angle may be used to guide the appropriate amount of bone resected. With the hip flexed to 90 degrees, the amount of overlap at the osteotomy is measured with the popliteal angle at 0 degrees. This amount represents the length of bone that may be removed. The osteotomy is fixed with a small hip screw or a blade plate. Depending on both patient variables (degree of spasticity, bony stock) and the quality of fixation, a spica cast may be desirable for at least several weeks postoperatively. Some surgeons prefer to perform bilateral femoral osteotomies with the goal of equalizing limb lengths and muscle balance about the hips. If unilateral surgery is performed, there is an asymmetry in the appearance of the proximal thigh and in leg lengths. Whether the prevalence of recurrent deformity is greater following unilateral bony surgery remains to be determined.

Patients with coexisting acetabular dysplasia (acetabular index >25 degrees, type II sourcil) require a pelvic osteotomy. In most spastic subluxation cases, the acetabular deficiency is posterolateral or global. Because the acetabulum is typically shallow and saucer shaped, the most common procedures in patients with neuromuscular hip dysplasia are volume-reducing procedures rather than redirectional procedures. Although some authors have reported success with a Pemberton osteotomy [55,62], most have performed variants of the osteotomy described by Dega [3,22,56–58,60,61,63]. A pericapsular osteotomy is performed approximately 5 mm above the insertion of the joint capsule. The cuts are visualized using an image intensifier, and curved osteotomies are required. The osteotomy extends down to the triradiate cartilage, which serves as a hinge. The lateral margin of the acetabulum is then levered downward, and triangular bone grafts are inserted. Several options are available for graft material, including tricortical iliac crest allograft, triangular grafts from the iliac crest, or a triangular wedge of bone removed during the femoral osteotomy. The location of insertion may be used to maximize coverage in one area. Variations in technique typically surround whether or not the osteotomy is extended into the sciatic notch, and whether a bicortical cut is made anteriorly. Fixation is not required, and a spica cast is not mandatory. For patients with greater degrees of subluxation or dislocation, an open reduction also may be required. The indications for open reduction are variable, and the San Diego group has suggested that a migration percentage of greater than 70% is an indication for routine open reduction [57,58]. Miller and coworkers [22] have suggested doing a medial capsulotomy if there is less than 20 degrees of abduction and the femoral head does not reduce under the acetabulum after the pelvic osteotomy.

Postoperatively, both diazepam and narcotics are generally required. Complications include a variety of medical problems (pneumonia, atelectasis, urinary tract infection); pressure sores; heterotopic ossification (increased risk if spine fusion is performed within 6 weeks) [78]; avascular necrosis of the femoral head; and insufficiency fractures. Stasikelis and coworkers [79] found that complications developed in 68% of patients with spastic quadriplegia and a gastrostomy or tracheostomy tube (12% without) and in 29% of nonambulators (8% of ambulators).

McNerney and coworkers [57] reported the results of a combined reconstruction of 104 hips with a mean follow-up of 6.9 years. The indications for a pelvic osteotomy included an open triradiate cartilage, an acetabular index greater than 25 degrees, and a migration percentage greater than 40%. An open reduction was routinely performed when the migration percentage exceeded 70%. Ninety-five percent of the hips remained well reduced at follow-up and there were no redislocations. Eight percent developed avascular necrosis. Miller and coworkers [22] reviewed 49 subluxated and 21 dislocated hips treated by a single-stage reconstruction including soft tissue releases, a femoral osteotomy, and in most cases a volume-reducing pelvic osteotomy. At 34 months follow-up, all but two hips remained located, 82% of patients with hip pain had complete relief, there were
no cases of avascular necrosis and 80% of caretakers believed that the surgery had been of benefit.

For symptomatic, older patients with subluxation or dislocation in whom deformity of the femoral head or degenerative changes preclude hip reconstruction, several options are available for salvage. The decision between reconstruction and salvage is often made intraoperatively by direct inspection of the femoral head. There is no consensus regarding the optimal treatment for this difficult problem, and options include resection of the femoral head and neck; valgus osteotomy; femoral shortening osteotomy; resection, valgus, soft tissue interposition (McHale procedure); arthrodesis; and arthroplasty (total hip versus humeral prosthesis).

The technique of proximal femoral resection-interposition arthroplasty was initially described by Castle and Schneider [66], and involves an extra-periosteal resection of the proximal femur just below the lesser trochanter. The quadriceps muscle is sutured down over the femoral shaft, and both the joint capsule and the abductors are sutured down over the acetabulum. Postoperative Russell traction was used until soft tissue healing was complete. Several studies have reported adequate results following this procedure [64,66,70,74]. Postoperative concerns include the formation of heterotopic ossification and proximal migration of the femoral shaft. McCarthy and coworkers [70] modified this slightly by using a line across the distal ischia to define the level of resection, and used 90-90 skeletal traction for 6 weeks. They reviewed 56 hips in 34 patients after a minimum of 2 years and found that all patients had an improvement in motion, and all but one had an improvement in seating. Heterotopic bone was identified in most patients; however, additional surgery was only required in a small subset. Widmann and coworkers [74] reviewed 18 hips treated in this fashion, most of which had prior surgical procedures. At more than 7 years follow-up, all patients exhibited adequate pain relief, improvement in motion, and improvement in seating. Postoperative irradiation was successful in decreasing heterotopic ossification, and skin traction was as effective as skeletal traction in preventing proximal migration. Pain relief was maximal at an average of more than 5 months following the procedure. These results suggest that proximal femoral resection-interposition arthroplasty remains a good option for salvage. Care should be taken to minimize soft tissue trauma during the dissection, to maintain adequate hemostasis, and to use suction drainage postoperatively. Postoperative irradiation should be considered, and skin traction may be as effective as skeletal traction.

The McHale procedure involves an adductor release; a resection of the femoral head and neck at the level of the base of the femoral neck; a valgus proximal femoral osteotomy with plate fixation (to achieve 45-degree abduction); placement of the lesser trochanter into the acetabulum (suture the psoas tendon to the ligamentum teres); and a capsulorrhaphy [71]. Patients were placed in a spica cast for approximately 3 weeks. There was improvement in pain, range of motion, and sitting endurance in all five patients. One patient exhibited a small amount of heterotopic ossification, and there was no proximal migration. In a retrospective review of 36 hips, Leet and coworkers [69] compared the McHale procedure with proximal femoral head resection (postoperative traction or an external fixator to maintain length). The age at surgery was 19 years, and the average follow up was 3.4 years. Although both groups had an improvement in their pain and an increase in their sitting tolerance, those treated by the McHale procedure had a shorter length of stay, lower rate of complications, and decreased superior migration of the femoral head.

Other alternatives described in a limited number of studies include femoral shortening osteotomy [80], hip arthrodesis [67], and prosthetic reconstruction [65,68,72,73]. Terjesen and Hellum [80] performed a femoral shortening osteotomy (3–5 cm) at the subtrochanteric level in 15 patients with chronic dislocations, and found adequate symptomatic relief in all patients at 5 years follow-up. Five of the 16 hips were reduced at follow-up. Arthrodesis of the hip has been reported in a limited number of patients, and may be considered in nonambulatory patients with unilateral disease [68]. Prosthetic reconstruction has also been reported, most commonly in ambulatory patients with painful degenerative changes [65,72,73]. Buly and coworkers [65] reviewed 18 adults treated by a cemented total hip arthroplasty at 10 years follow-up. A spica cast was used in the immediate postoperative period. Pain relief was achieved in 94%, and at latest follow-up 95% had no evidence of loosening. Weber and Cabanela [73] reviewed 16 patients following total hip arthroplasty, and found adequate pain relief in 87%, with an improvement in function in 79%. Gabos and coworkers [68] implanted a noncemented humeral prosthesis in 14 hips (11 patients) with degenerative arthritis. In seven hips a glenoid component was inserted into the false acetabulum. At 5 years follow-up, 10 of 11 patients had complete pain relief, although four prostheses became dislocated. Caregiver satisfaction with seating and motion was high. Five patients developed heterotopic ossification, and one implant exhibited osteolysis.
Summary

Hip problems are very common in patients with cerebral palsy, particularly those who are nonambulatory with a large degree of spasticity. The emphasis has been on early detection of patients at risk for progressive dysplasia, and both clinical and radiographic screening may be useful for early detection. Although the natural history is variable, and not all hips progress to subluxation or dislocation, a subset of patients with chronically subluxated or dislocated hips has discomfort. Nonoperative strategies are currently under investigation, and it remains unclear whether physical therapy, abduction splinting, botulinum toxin A, or interthecal baclofen alter the natural history of spastic hip disease. For patients with early disease and the absence of bony deformity, a soft tissue release may play a role in stabilizing the hip. For those with established deformity, reconstructive or salvage options are available depending on the shape of the femoral head and the status of the articular cartilage. Reconstructive surgery usually involves a soft tissue release followed by a proximal femoral varus derotational osteotomy. Patients with coexisting acetabular dysplasia benefit from a volume-reducing pelvic osteotomy. A subset of patients may also benefit from an open reduction and capsulotomy, and this also facilitates inspection of the femoral head. For patients with established dislocation, or severe subluxation with deformity of the femoral head and breakdown of articular cartilage, a host of salvage options are available.

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