Musculoskeletal Complications of Neuromuscular Disease in Children

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A wide variety of neuromuscular diseases affect children, including central nervous system disorders such as cerebral palsy and spinal cord injury; motor neuron disorders such as spinal muscular atrophy; peripheral nerve disorders such as Charcot-Marie-Tooth disease; neuromuscular junction disorders such as congenital myasthenia gravis; and muscle fiber disorders such as Duchenne’s muscular dystrophy. Although the origins and clinical syndromes vary significantly, outcomes related to musculoskeletal complications are often shared. The most frequently encountered musculoskeletal complications of neuromuscular disorders in children are scoliosis, bony rotational deformities, and hip dysplasia. Management is often challenging to those who work with children who have neuromuscular disorders.

Scoliosis

Scoliosis refers to deviation from normal spinal alignment. A commonly accepted definition of scoliosis is a curvature in the coronal plane of greater than 10°. The coronal curvature is almost always associated with a sagittal alignment abnormality, such as kyphosis, lordosis, or a rotational component. Scoliosis may be classified as idiopathic, congenital, or neuromuscular in origin. Overall, idiopathic scoliosis accounts for the significant majority of cases of scoliosis in children and adolescents, whereas scoliosis associated with neuromuscular disease, congenital deformity, and other causes occurs less frequently in the total population. Neuromuscular scoliosis can occur as...
a complication of a wide variety of disease processes in children, including upper and lower motor neuron conditions and myopathies.

Scoliosis may lead to functional deficits, such as decreased sitting balance. The upper extremities may be required to maintain upright posture, thereby reducing the availability of the arms for functional daily tasks. Neck, shoulder, and spine range of motion may be limited. In Duchenne’s muscular dystrophy, for example, the rigid neck, hyperextension deformity with associated marked increase of cervical lordosis forces patients to bend their trunk forward and assume an awkward posture to look straight ahead [1]. Scoliosis may result in skin breakdown or pain. As scoliosis becomes more severe, reduction in lung volumes and diaphragmatic heights may occur [2]. Beyond 100°, pulmonary hypertension and right ventricular hypertrophy may develop [3].

Epidemiology

Idiopathic scoliosis occurs in 2% to 3% of the adolescent population [4]. In contrast, the rates of spinal deformity in children who have neuromuscular disease are generally much higher and depend on the diagnosis (Table 1). For example, 20% of patients who have mild cerebral palsy may develop scoliosis, but nearly 100% of those who have thoracic spinal cord injury that occurs before puberty will develop this disease. Although idiopathic scoliosis is much more common in girls than boys [26], neuromuscular scoliosis does not discriminate between the genders. Children who have undergone selective dorsal rhizotomy for spasticity control seem to have a higher incidence of spinal deformity than those who have not undergone this procedure [27–30].

Origin

The origin of idiopathic scoliosis is unknown, although genetic, environmental, and undetected neuromuscular dysfunction are hypothesized causes [29,30]. In neuromuscular scoliosis, the situation is even more complex.

Table 1
Prevalence of scoliosis and hip dysplasia in children who have neuromuscular disease

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<th>Cerebral palsy</th>
<th>Myelomeningocele</th>
<th>Duchenne’s muscular dystrophy</th>
<th>Spinal cord injury</th>
<th>Charcot-Marie-Tooth</th>
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Upright posture may be impaired because of abnormalities in the intricate coordination among central nervous system, muscle, bone, cartilage, and soft tissue. Asymmetric weakness, spasticity, abnormal sensory feedback, or mechanical factors such as pelvic obliquity or unilateral hip dislocation may cause an initial, flexible spinal curve. However, which parameter contributes most or even determines the direction of the curve is still unknown. No significant correlation between muscle asymmetry or side of dislocated hip and side of scoliotic convexity has been discovered [7,15]. Whatever the origin or initial trigger, once a postural abnormality is present, a vicious cycle of progression may occur such that unequal compression on vertebrae causes unequal growth. Asymmetric growth may cause further unequal compression on the spinal structures, causing the cycle to perpetuate itself. If this cycle is sustained beyond a critical threshold of weight and time, fixed deformity with changes in vertebral and rib structure may follow, and spinal deformity develops [31]. Various triggers may cause the imbalanced spinal axis, but biomechanical forces may account for its progression [32]. Neuromuscular scoliosis is more likely to be rapidly progressive than idiopathic [11,33]. Some evidence indicates, however, that if the underlying origin is corrected, such as spinal cord untethering, the spinal curvature may improve [34,35].

**Evaluation**

Many neuromuscular diagnoses are confirmed at or around birth. In those circumstances, subsequent evaluations occur with full knowledge of expected outcomes related to spinal deformity. However, conditions such as the hereditary motor sensory neuropathies may not be recognized until later in childhood, and scoliosis may be the presenting symptom. The history of a child who has scoliosis should include information about pre- and perinatal events; developmental milestones; evidence of skill regression; age of onset of symptoms; other system disorders or anomalies (especially renal and cardiac); the presence of associated symptoms such as sensory loss, weakness, or pain; functional deficits; and family history.

Therefore, idiopathic scoliosis is a diagnosis of exclusion. All children and adolescents who have scoliosis should undergo a careful neurologic and musculoskeletal examination. In one study, 23% of children referred to an orthopedic practice who had scoliosis and an atypical curve, congenital scoliosis, gait abnormality, limb pain, or weakness or foot deformity, had an MRI-identified spinal cord pathology [36]. In children who have no known neuromuscular disease, MRI should be obtained when a rapidly progressive curve (more than 1° per month), left-sided thoracic curve, neurologic deficit, limb deformity, or worrisome pain symptoms are identified.

The physical examination should include evaluation for pelvic obliquity, shoulder girdle asymmetry, waist crease asymmetry, rib prominence, or asymmetry with spinal flexion, leg length discrepancy, fixed foot deformity,
hip dislocation or subluxation, and limitation of spinal or extremity range of motion. A full neurologic examination should be performed, including an assessment of strength, muscle tone, reflexes (including abdominal reflexes), sensation, balance, cranial nerve function, speech and language, and cognition. A functional assessment is also an important component. Abnormalities in any of these areas may provide clues to origin, expected outcomes, and treatment strategies.

Radiographic evaluation includes a posteroanterior view of the entire spine. Standing films are most useful, although sitting films may be substituted when necessary. The Cobb method is the most commonly used technique to measure the degree of scoliosis (Fig. 1). A widely accepted grading classification denotes a mild curve if between 10° and 40°, a moderate curve if between 40° and 65°, and a severe curve if greater than 65°. Intra- and interobserver measurement variability is within the range of 3° to 10° for noncongenital scoliosis [37]. Curves are named for the location of the apex vertebrae, and are described as right or left based on their predominant convexity. They are designated C-shaped or double depending on their configuration. Idiopathic adolescent curves are more likely to be right-sided and thoracic in location. Experts have believed that neuromuscular curves have a higher incidence of left-sided convexity [11], although a recent retrospective study suggests that the curve patterns and apical levels in neuromuscular scoliosis are similar to those reported for idiopathic adolescent scoliosis.

Fig. 1. Cobb method of measuring scoliotic curve in which the vertebra with maximally tilted end plates above and below the apex are identified. The angle between lines drawn along the superior and inferior endplates or the angle of lines drawn perpendicular to them is the Cobb angle. (From Magee DJ. Orthopedic physical assessment, 4th edition. Philadelphia: Saunders; 2002. p. 461; with permission.)
Before surgery, curve flexibility may be assessed using supine lateral bending, fulcrum, or traction radiographs [39].

Nonoperative treatment

If the vicious cycle can be disrupted or the continuous state of asymmetric loading can be prevented early enough that significant spinal bony deformity has not occurred, some experts are hopeful that the progression of scoliosis may be mitigated [40]. A small body of literature suggests that exercise-based approaches in addition to bracing may be effective in some girls who have adolescent idiopathic scoliosis [41–43]. However, the daily use of a spinal orthosis is the mainstay of treatment for girls who have idiopathic scoliosis.

The effectiveness of nonoperative treatment in children who have neuromuscular scoliosis is controversial. Although intuitively attractive, the theory that controlling the mechanical forces acting on the spine will result in decreased curve progression has infrequently been translated into clinical practice [31]. Data are limited regarding efficacy of nonoperative treatment and bracing in preventing curve progression in neuromuscular scoliosis. Olafsson and colleagues [44] reported on brace use in 90 consecutive children who had various types of neuromuscular scoliosis. They observed a 28% success rate (defined as curve progression of less than 10° per year and good brace compliance) with a higher likelihood of improvement in ambulators with hypotonia and short lumbar curves of less than 40° and in nonambulators with spasticity and short lumbar curves. Those who had longer, hypotonic curves experienced less success. In another group of children who had myelomeningocele and a curve not exceeding 45°, a Boston brace was used successfully to arrest or slow the progression of scoliosis in most [45]. However, Miller and colleagues [46] reported no benefit after 67 months of bracing in 20 children who had spastic quadriplegia related to curve magnitude, shape, or rate of progression. Whether spinal orthoses and other conservative management techniques may be helpful in slowing the progression of scoliosis in certain subpopulations of children who have neuromuscular disease remains to be seen, but the prevailing attitude suggests that they are not.

Nonoperative interventions, including sitting supports and custom seating, spinal orthoses, and functional strengthening programs may be useful to improve sitting balance and functional independence [47–50]. In myelodysplasia, a soft thoracolumbosacral orthosis (TLSO) may be used to improve seating and positioning to free the upper extremities for functional tasks or as a temporizing measure to allow the child to develop increased trunk length before surgery [51].

Some are concerned that placing children who have neuromuscular disorders in a TLSO to improve postural function may cause further respiratory compromise, especially for children who have hypotonia. Bayar and colleagues [52] treated 15 children who had neuromuscular scoliosis who used
a polyethylene custom spinal orthosis for 8 to 10 hours and postural training, 
muscle strengthening, and stretching 5 days per week, with special emphasis 
on respiratory exercises for 4 weeks. Strength, range of motion, and balance 
improved although scoliosis did not. The forced vital capacity (FVC) while 
wearing the brace initially decreased by 18%. However, the negative effect 
on FVC lessened after the program, suggesting an improvement in coping 
with the restrictive effect of the brace. Further research showed that the use 
of a soft Boston brace did not impact negatively on the pulmonary mechanics 
and gas exchange in one group of children who had severe cerebral palsy and, 
in fact, decreased the work of breathing in some [53].

Special mention of boys who have Duchenne’s muscular dystrophy is 
warranted. Significant progression of scoliosis is unusual while the child re-
 mains ambulant. Rapid progression of scoliosis seems to be related to the 
loss of walking ability and commonly corresponds with a growth spurt in 
adolescence [54]. The use of corticosteroids [55,56] and orthotics, such as 
knee-ankle-foot orthoses [57], have been shown to prolong ambulatory abili-
ity. This intervention seems to significantly delay onset and decrease severity 
of scoliosis so that a much smaller proportion of boys who have Duchenne’s 
require surgical stabilization [56,58]. Even without steroid treatment, not all 
boys who have Duchenne’s muscular dystrophy will need scoliosis surgery. 
It was recently recognized that up to 25% of nonambulant boys do not 
develop clinically significant scoliosis and therefore do not require surgical 
intervention [8]. As with other neuromuscular disorders, the primary indica-
tion for bracing is to improve postural control and seating rather than pre-
vent progression of curvature [54].

Surgery

The goals of surgical stabilization for spinal deformity in neuromuscular 
disease include correcting the curvature, preventing significant progression 
of the curvature, improving the balanced position of the spine, and, there-
fore, improving quality of life. Indications for surgical intervention include 
progressive deformity that compromises ability to sit or stand, cardiac or 
pulmonary function, skin integrity, and ability to perform nursing cares, 
and causes pain (Fig. 2). Reported outcomes of surgical intervention for 
neuromuscular scoliosis include improved Cobb angle, lung function, seat-
ing position and balance, and ability to perform activities of daily living, 
and decreased pain and time used for resting (Fig. 3) [59,60]. Self-esteem 
has also been shown to improve after surgery [60,61]. In Duchenne’s muscu-
lar dystrophy, most data do not show a significant effect of scoliosis surgery 
on respiratory function or survival [62,63].

Various surgical techniques have been described and their merits debated. 
Surgical considerations include anterior and posterior fusion versus poste-
rior-only fusion, one-stage versus two-stage procedures, various instrumenta-
tion techniques, and the extension of instrumentation across the
lumbosacral junction and sacroiliac joint [51,64–66]. From a surgical perspective, best results are achieved when the curve is progressive but not severe or rigid and when medical status is optimal [67].

Children who have neuromuscular scoliosis experience more complicated and costly hospitalizations from their scoliosis surgery than those who have idiopathic scoliosis. Before surgery, children who have neuromuscular disease are more likely to have gastrostomy tubes, failure to thrive, gastroesophageal reflux, and other medical diagnoses. Other challenges related to surgical procedures in children who have neuromuscular disease include curve severity that is characteristically worse and more rigid; osteoporosis; extension of deformity to include fixed pelvic obliquity; poor soft tissue coverage; deficiency of posterior spinal elements, such as in myelodysplasia; and tenuous neurologic status [33,51]. Postoperatively, they experience a higher frequency of pneumonia, respiratory failure, mechanical ventilation, urinary tract infection, surgical wound infection, central line placement, transient or permanent neurologic loss, and failure of the surgical procedure or hardware [51,68,69].

Among children who had cerebral palsy who underwent scoliosis surgery, the number of days in the intensive care unit and the presence of severe preoperative thoracic hyperkyphosis negatively affected survival rate [70]. Negative functional outcomes have been reported, such as loss of ability to roll, feed oneself, and walk [9,61].

Fig. 2. Preoperative radiograph of an 11-year-old girl who has idiopathic scoliosis.
Historically, children who have severe restrictive lung disease and an FVC of less than 30% of predicted have not been considered surgical candidates. However, several recent studies indicate that with aggressive team management by pulmonary, cardiac, anesthesia, and intensive care pediatric services, these children can safely undergo surgical spine stabilization without the need for tracheostomy or prolonged ventilation [71–73].

**Rotational deformities of bone**

Rotational malalignment of the lower extremities is a common outcome of neuromuscular disease. The spectrum of bony deformities has been referred to as *lever arm disease* [74,75]. Rotational deformities often occur at the femur and tibia and have a deleterious effect on function and cosmesis. Muscle efficiency may be reduced because the skeletal lever arms are not aligned with the line of progression during gait. For example, in cerebral palsy, intoeing occurs commonly. The increased internal foot progression angle may place muscle groups at a mechanical disadvantage and be associated with poor foot clearance, tripping, and falling and a cosmetically poor gait pattern. Torsional deformities may also be associated with premature degenerative processes at the hip and knee [76–79].
Epidemiology

In a recent retrospective gait analysis study of 412 children who had cerebral palsy, 37% of toeing gait had multiple causes. The most common contributors, either alone or in combination, were internal hip rotation in 55% and internal tibial torsion in 50%. Pes varus and metatarsus adductus also contributed [80]. Although experts have previously suggested that spasticity of hamstrings and adductors contribute substantially to an internally rotated gait, more recent evidence suggests that toeing in children who have cerebral palsy is almost universally associated with osseous deformity rather than hypertonia [80–82]. The overall prevalence of excessive internal hip rotation in cerebral palsy is 27%, with prevalence higher in those who have diplegia than in those who have hemiplegia [81].

Etiology

Abnormalities of muscle strength and tone from neuromuscular disease are believed to be ultimately responsible for the development of rotational deformity. Femoral anteversion in able-bodied infants is not significantly different from that in infants who have cerebral palsy. The average newborn shows 30° to 40° of femoral anteversion. This decreases to 10° to 15° by adolescence in a typically developing population [83]. However, children who have cerebral palsy are more likely to experience failure of the typical corrective lateral rotation that occurs with growth and development in their able-bodied counterparts [84]. Persistent hip flexor spasticity and tightness are believed to contribute because they prevent normal extension of the hip and concomitant external rotation, thus the usual remodeling of the infant torsion cannot occur [81].

Similarly, remodeling and lateral derotation of the usual infant internal tibial torsion may not occur in neuromuscular disease. At birth, the malleoli are level in the frontal plane. In typically developing children, most normal external rotation of the tibia occurs by 4 years of age, with an additional degree per year occurring up until skeletal maturity for a final average of 28° of external rotation [85]. Because of this lateral rotation of the tibia that occurs with normal growth, internal rotation abnormalities may improve with time. However, several factors, including muscle imbalance, soft-tissue contractures, associated congenital malformations, and mechanical abnormalities caused by habitually assumed posture over time, may impede this process causing internal tibial torsion to persist. In addition, other children, such as some who have myelomeningocele, may develop significant fixed external tibial torsion associated with valgus of the hindfoot, midfoot abduction, planus deformity, and genu valgum.

Evaluation of lower-extremity rotational deformity

Internal hip rotation, femoral anteversion, and medial femoral torsion all refer to an increased angle of the femoral neck relative to the transcondylar
axis of the knee. In other words, the axis of the hip is anterior or external to that of the knee [75]. Femoral anteversion may be assessed using physical examination, radiography, ultrasound, and CT scan and requires optimal positioning of the child for accurate measurement. The most commonly used physical examination maneuver (Craig’s test or the Ryder method) places the child prone with pelvis stable, hips extended, and knee flexed to 90°. The leg is then rotated outwardly with goniometric measurement of the angle between the shank and vertical. This angle is equal to the degree of femoral anteversion (Fig. 4).

Tibial torsion is defined as the angle formed between the articular axes of the knee and ankle joint. Tibial torsion is often measured using an assessment of the thigh–foot angle. The child is placed prone with the knee flexed to 90° and the ankle supported in a neutral position. The axis of the foot is then compared with the long axis of the thigh. Alternatively, the degree of tibial torsion can be measured in a seated position, using a goniometer to measure the angle between the visualized bimalleolar axis and the femoral epicondylar axis.

Nonsurgical intervention for torsional deformities

Experts widely believe that traditional exercise, night splints, shoe inserts, twister cables, and other conservative options cannot reverse fixed femoral

Fig. 4. Prone hip rotation measuring femoral anteversion. (Adapted from Magee DJ. Orthopedic physical assessment, 4th edition. Philadelphia: Saunders; 2002. p. 622; with permission.)
or tibial torsion [86]. However, aggressive treatment of spasticity may help prevent development or slow the progression of torsional deformities. Short-term improvements in functional outcomes (gait, Gross Motor Function Measure, and clinical examination) using botulinum toxin injections have been reported, but evidence is limited regarding the effect of botulinum toxin treatment on the development of bony deformity. In a nested case-control design, Desloovere and colleagues [87] reported an improved gait pattern characterized by fewer contractures at the level of the hip, knee, and ankle and decreased internal hip rotation at initial contact, toe-off, and mid-swing in children who had undergone multilevel botulinum A treatments. Botulinum injections were started at a young age and combined with common conservative treatment options. The authors concluded that children treated with multilevel botulinum A injections have a gait pattern less defined by bony deformity than their nontreated counterparts.

**Surgery**

Medial femoral torsion of greater than 40° to 45° that interferes with gait and function may be corrected surgically with a femoral derotational osteotomy (Figs. 5 and 6). Both proximal and distal surgical techniques have been described. A proximal osteotomy may be beneficial when a child has both femoral torsion and hip subluxation to allow varus angulation of the femoral neck and ensure stability of the hip through proximal femur internal rotation and distal femur external rotation. However, when the hips are stable, distal osteotomies are reportedly less invasive, provide quicker recovery time, and are as effective as proximal surgery in functional and cosmetic outcomes [88–90]. They also provide the added opportunity to correct a knee flexion contracture if needed. Long-term results indicate that partial

![Preoperative radiograph of a 4-year-old girl who has spastic diplegic cerebral palsy, bilateral coxa valga, and uncovering of the lateral one fourth of the femoral heads.](image-url)
recurrence of rotational deformity may occur in 0% and 33% of cases, with surgery before 10 years of age more likely to show deterioration [89,91]. Some centers avoid postoperative casting and encourage early mobilization [88]. Complications of femoral osteotomies include loss of fixation, delayed union, hardware failure, wound dehiscence or infection, and over- or under-correction [90,92].

Tibial torsion can also be surgically corrected using a tibial derotational osteotomy (Figs. 7–9). Various surgical techniques have been described, including proximal versus distal site of osteotomy, different shapes of osteotomy, various types of fixation, and possible simultaneous fibular osteotomy [86,92–94]. Complications include delayed union, cross-union, or nonunion; wound dehiscence; osteomyelitis; late fracture; distal physeal closure; and neurovascular compromise [93,94]. When combined with a split tibialis posterior tendon transfer for spastic equinovarus deformity, severe planovalgus or rigid equinovarus deformity has a higher rate of development presumably because of the increased difficulty in balancing the muscle forces across the spastic equinovarus foot [95].

Hip dysplasia

Hip dysplasia, subluxation, and dislocation are orthopedic abnormalities encountered in children who have neuromuscular disorders. Hip dysplasia refers to a spectrum of conditions of the hip that may be present at or shortly after birth, including inadequate acetabular formation, femoral head subluxation, and femoral head dislocation [96]. Hip subluxation and
hip dislocation have typically been defined by the hip migration percentage or Riemers’ migration index, as measured on an anteroposterior radiograph. This measures the femoral head’s containment within the acetabulum in the coronal plane with respect to Perkin’s line [97–100] (Fig. 10). Shenton’s

Fig. 7. Preoperative radiograph of a 5-year-old girl who has lumbar myelomeningocele and bilateral internal tibial torsion with severe intoeing.

Fig. 8. Same girl as in Fig. 7 after bilateral distal tibial external rotation–producing osteotomies.
line, which is formed by the medial aspect of the obturator foramen and the medial aspect of the femoral neck, forms an unbroken arc in the normal hip. However, in a dislocated hip, this arc will be discontinuous (see Fig. 10). Hip subluxation is usually diagnosed with a hip migration percentage of greater than 33%, although others may classify subluxation as mild when it exceeds 20% [21,99,101,102]. Hip dislocation is diagnosed when the migration percentage is greater than 100% or the femoral head is completely uncovered [102].

Other bony abnormalities, such as a shallow acetabulum, coxa valga, and femoral anteversion, are commonly associated with or contribute to femoral subluxation or dislocation. Radiographic measurements are used to evaluate these hip abnormalities. The acetabular index measures the slope of the acetabular roof compared with Hilgenreiner’s line (see Fig. 10). An acetabular index of greater than 30° indicates dislocation, although accuracy of the measurement depends on patient positioning and age. Coxa valga is an increased neck–shaft angle of the femur. The neck–shaft angle of a newborn is typically 150° and typically 120° to 135° in an adult. Coxa valga in an adult is defined as an angle of greater than 135° (Fig. 11).

The most common functional impairments related to hip dysplasia include difficulty with seating, positioning, transfers, perineal hygiene, dressing, and pain [103,104]. Other potential issues include pressure sores and deformity. Seating issues are often complex because many of these children have concomitant pelvic obliquity and scoliosis.

In those who have milder disease or later presentation, the functional impairment may be less severe and occur late. For example, hip abnormalities
in children who have Charcot-Marie-Tooth disease are generally asymptomatic and may be found on radiographs obtained for other reasons. Often the hip abnormality goes undetected until adolescence or adulthood when the patient presents with a gait abnormality and pain. Pain tends to be seen in the later stages of the hip disorder when the joint may have marked subluxation or arthrodesis [105,106].

**Epidemiology**

In children who have no known neuromuscular disorder, the incidence of congenital hip dysplasia is 1 per 85 births with a 5:1 female-to-male ratio [107,108]. Risk factors for congenital hip dysplasia include a family history of congenital hip dysplasia, first born, female gender, and breech delivery; 25% are bilateral. When unilateral, it is four times more common in left hip [107–109].

In comparison, children who have neuromuscular disorders have an incidence of hip disorders of 8% to 82%, depending on the neuromuscular disorder, age of onset, and severity (see Table 1) [21–23,110,111]. The prevalence of hip dysplasia in cerebral palsy varies from 2% to 60%, with higher prevalence among children who are quadriplegic or nonambulatory, or have severe spasticity [110–112]. In cerebral palsy, the risk for subluxation or...
dislocation is directly related to gross motor function as measured with the Gross Motor Function Classification System (GMFCS) [113]. In children who have with spinal cord injury, the incidence of hip subluxation or dislocation is inversely related to age (ie, the older the child at injury, the lower the incidence of hip subluxation or dislocation) [21,22].

**Etiology**

In children who have congenital hip dysplasia without an underlying neuromuscular disorder, the most likely causes are related to intrauterine positioning, hormones, and joint laxity. In upper motor neuron disorders, such as cerebral palsy and spinal cord injury, the underlying cause of the hip disorder is a combination of muscular imbalance, spasticity, contractures, and limited ambulation. For example, muscular imbalance may be manifest through spastic hip flexors and hip adductors. This imbalance may cause asymmetric forces on the developing bony structures of the hip, resulting in deformities such as femoral anteversion, flattening of the femoral head and acetabulum, posterolateral migration of the femoral head, and flexion–adduction contractures [97,99,103,114,115]. In addition, children who have severe spasticity are often nonambulatory. The combination of a lack of ambulation and asymmetric muscle forces at the hip can exacerbate abnormalities of the femoral head and acetabulum. This results in increased forces at the lesser trochanter because of a shift in the mechanical axis of the hip with posterolateral migration of the femoral head. As the hip migration and increased lesser trochanter forces continue, subluxation, dislocation, and coxa valga may occur [111,114,116].

Fig. 11. Coxa valga denotes an increased neck–shaft angle compared with normal (125° in an adult). (Adapted from Magee DJ. Orthopedic physical assessment, 4th edition. Philadelphia: Saunders; 2002. p. 627; with permission.)
Similar factors occur in lower motor neuron disorders, such as spinal muscular atrophy and Charcot-Marie-Tooth disease. Global proximal weakness, limited weight-bearing, and ligamentous laxity may cause coxa valga. Trochanteric apophyseal growth may be diminished secondary to decreased weight-bearing and gluteal weakness, further promoting the coxa valga deformity [117,118]. In Charcot-Marie–Tooth disease, the proximal weakness is more subtle but may still result in a shallow acetabulum and a valgus anteverted femoral neck [106]. Children who have myopathies such as Duchenne’s muscular dystrophy are often independently mobile and ambulatory during crucial times of hip development. As a result, hip disorders are less likely but still possible given the muscular imbalance. Finally, the hip dysplasia associated with congenital torticollis has an uncertain origin. However, the conditions causing the development of the torticollis may participate in the development of hip dysplasia [119].

Natural history of hip dysplasia

In congenital hip dysplasia, congruent reduction achieved before 4 years of age typically results in normal hip development [120]. Evidence shows that proper hip development requires the femoral head to be centralized in the acetabulum by or around 5 years of age [121–123].

In a study of children who had quadriplegic cerebral palsy who did not walk, the mean hip migration index was 12% per year. In ambulatory children, the hip migration index was 2% per year. Ambulatory status and age were found to be the most influential factors on rate of progression of hip migration, and therefore young, nonambulatory children tend to have more rapid progression of hip migration [124].

Pelvic obliquity may coexist with hip subluxation, hip dislocation, scoliosis, and “wind-swept” hips (adduction of one hip and abduction of the other hip). The relationship between pelvic obliquity and hip dysplasia is controversial [96,121,125–128]. Suprapelvic obliquity refers to obliquity secondary to scoliosis, whereas infrapelvic obliquity refers to imbalance below the pelvis. One study of children who had spastic cerebral palsy found that the development of hip dysplasia was more related to infrapelvic obliquity than suprapelvic obliquity. Infrapelvic obliquity was noted to precede suprapelvic obliquity, and hip subluxation and dislocation almost always occurred on the high side [129].

Evaluation

Key elements of the history related to hip dysplasia include intrauterine abnormalities such as oligohydramnios; birth difficulties such as breech birth; and a family history of hip disorders in children or young adults. A review of symptoms, including the presence of pain, decreased sitting tolerance, autonomic dysreflexia, and skin ulcers, is critical [130,131]. A hip
physical examination for dysplasia includes an evaluation of the presence of
an asymmetry of fat folds of the thigh and buttocks; a Trendelenburg’s sign;
limitations of passive range of motion in all directions, including asymme-
try; “popping,” or pain. Hip flexion contractures are evaluated using the
Thomas test and rectus tightness using the Ely test. Hip adduction contrac-
tures may be assessed with the hip and knee in extension (gracilis stretch)
and flexion. Hip rotation is best assessed with the child prone. An apparent
leg length discrepancy may be evaluated using the Galeazzi sign (Fig. 12),
for which the child lies supine with knees and hips flexed. If the knees are
not at the same height, the low side may be posteriorly subluxed or dislo-
cated. One may also evaluate for a telescoping sign (Fig. 13) in which the
child is again placed supine with hips and knees flexed, and the femur is
pushed posteriorly toward the table and lifted up. A normal hip will show
little motion, but a dislocated hip will reveal an excessive telescoping or pis-
toning movement.

Special maneuvers, such as the Ortolani and Barlow tests, are used in in-
fants (Fig. 14). With the child calm and pelvis stable, the Ortolani test is per-
formed by first flexing the knee and hip. The thigh is then abducted while
applying slight traction to the distal thigh and slight anterior pressure
against the trochanters. If the hip is dislocated before starting the maneuver,
one may palpate a relocation “clunk.” The Barlow test continues from this
position. The hip is then adducted with a slight compressive force backward
and outward on the inner thigh while palpating for a “clunk.”

Fig. 12. The Galeazzi sign is useful in infants and toddlers for assessing unilateral hip disloca-
tion or dysplasia. (Adapted from Magee DJ. Orthopedic physical assessment, 4th edition. Phil-
adelpia: Saunders; 2002. p. 627; with permission.)
Anteroposterior radiographs of the pelvis with legs extended may show subluxation, dislocation, and lateral notching of the femoral head (Fig. 15). The lateral notching has been hypothesized to be caused by chronic pressure from ligamentum teres, the joint capsule, the reflected portion of the rectus femoris, and the hip abductor musculature, but was recently found to be most likely caused by a spastic gluteus minimus [132]. A systematic literature review evaluating the evidence on hip surveillance in children who have cerebral palsy concluded that all children who have bilateral cerebral palsy should have a radiograph of the hips at age 30 months or sooner if clinically suspicious. Children who have a migration index greater than 33% or acetabular index greater than 30° are most likely to require further treatment of their hips, particularly if noted by 30 months of age [110]. Others recommend that children who have more severe neuromuscular disorders, such as quadriplegic cerebral palsy, undergo a radiograph of the pelvis at 1 year of age and yearly thereafter until the natural history has been established. Children who have spastic diplegia should begin screening at 2 to 3 years of age, with subsequent radiographs every 2 to 3 years [124]. In infants who have Charcot-Marie-Tooth type 1, a screening ultrasound is recommended. In Charcot-Marie-Tooth type 2, screening with pelvis radiographs at least every 2 years is recommended [106].

In newborns, ultrasound is the recommended imaging modality if a hip abnormality is suspected based on history or physical examination, because ultrasound can image cartilage. Because the femoral heads do not ossify until 3 to 6 months of age, radiographs may not completely show the femoral–acetabular relationship [107]. Repeat ultrasound imaging is recommended because false-positive findings are not uncommon in newborns.

Other imaging modalities, such as CT or MRI, may be considered in selected cases. Three-dimensional CT may provide additional detail about the femoral head and acetabular relationship, thus aiding in surgical
planning. CT, for example, may provide more comprehensive evaluation of
the location of acetabular dysplasia. The most common location of acetab-
ular dysplasia is posterior, but abnormalities have been noted in other loca-
tions, including anterior, midsuperior, anterosuperior, posterosuperior, and
global [133]. Although used infrequently, MRI may be useful for evaluating
the hip with an unossified femoral head that has been resistant to conserva-
tive treatment and may not be otherwise adequately imaged for presurgical
planning [134].

Nonoperative treatment for hip dysplasia

A physical therapy program performed by therapists and caregivers, with
daily focus on stretching of tight muscles, positioning, weight-bearing, and
orthotic devices is essential. Maintaining flexibility of two joint muscles,
such as the gastrocnemius, hamstrings, gracilis, and rectus femoris, is important. Standing or walking with or without orthoses has been shown to be crucial in delaying or preventing hip subluxation or dislocation in children who have upper and lower motor neuron disorders [25,123].

Nonoperative treatment approaches for developmental hip dysplasia include orthotics such as the Pavlik harness, Frejka pillow, Craig splint, or Van Rosen splint. The Pavlik harness is most commonly used [96]. These orthoses are intended to provide a prolonged stretch to hypertonic or tight hip adductors and promote correct acetabular development and spontaneous reduction of subluxed or dislocated hips. However, use of abduction bracing is contraindicated in patients who have lower motor neuron disorders, ligamentous laxity (Ehlers-Danlos syndrome), or fixed deformities (arthrogryposis) [106,135].

Other methods of postural management have been evaluated, although studies are small and use different postural devices. Postural devices include systems such as prone and supine lying supports, standing frames, and wheelchair seating systems, which all attempt to keep the hips in an abducted position. The amount of time the specific device is used depends on the severity of the hip migration, type of device used, and child’s tolerance. Some systems are recommended for up to 24-hour use. Studies have shown benefit when these devices are worn as intended [130,136,137].

Spasticity is believed to be a contributor to hip subluxation and dislocation in children who have cerebral palsy. Therefore, aggressive spasticity treatment has been speculated to reduce the progression of spastic hip disease. The effects of intrathecal baclofen on spasticity reduction are well known. One prospective, open-label, multicenter case series has been

Fig. 15. Radiographs of a 5-year-old boy who has linear sebaceous nevus syndrome and right hemiplegia. Bilateral coxa valga, right greater than left. Superolateral subluxation of the right femoral head, which is covered less than 10% by the shallow acetabulum. Less than one fourth uncovering of the left femoral head.
published on intrathecal baclofen and hip dysplasia in 33 children. The participants ranged from 4 to 31 years of age and included those who had paraplegic, tetraplegic, and diplegic cerebral palsy; most were nonambulatory. They were followed up for 1 year. The hip migration percentage stabilized or decreased in more than 90% of participants, with a trend toward greater improvement in younger participants. No controls were included, and more than two thirds of participants experienced at least one adverse event post-implant, including some serious drug-related events [138].

The effect of a single botulinum A injection to hip adductors was evaluated in one small retrospective study. Children who had an initial migration percentage greater than 30% who were younger than 24 months at injection were most likely to exhibit stabilization or improvement in the migration percentage during the 6-month follow-up [139]. In a randomized prospective study of children who had cerebral palsy, the group treated with botulinum A and a variable hip abductor brace required soft tissue surgery for hip adductor muscles less often than a control group who underwent standard physical therapy only. However, longer-term outcomes are not yet available [140]. Although more research is needed, a combination of botulinum A, hip abduction orthoses, and physical therapy starting in children younger than 24 months may prevent or delay hip disorders. In children who had cerebral palsy who underwent dorsal rhizotomies, the subsequent frequency of hip subluxation or dislocation was most often stable or reduced [141].

Operative treatment

The goal of operative intervention for hip dysplasia is to maintain mobile, located hips so that sitting balance, ambulatory ability, and comfort are enhanced. Operative interventions include soft tissue lengthening and hip reconstruction using femoral osteotomy with or without pelvic osteotomy. Salvage procedures are available for patients who have deformity of the femoral head, breakdown of articular cartilage, and established dislocation that cannot be repaired. In neuromuscular hip dysplasia, surgical intervention may be necessary when hip deformity or disability has progressed despite maximal conservative intervention. The timing of surgical intervention and type of intervention have been debated. However, hips with a migration percentage greater than 50% frequently require surgical intervention because of the risk for further progression and dislocation [124,142]. In addition, hips with greater than 70% of the femoral head uncovered preoperatively have a higher incidence of instability postoperatively [143].

Soft tissue procedures are often recommended as a prophylactic measure against the development of bony deformity. In patients who do not have bony deformity, these procedures may play a role in stabilizing the hip. Procedures include iliopsoas, hamstring, and adductor release or lengthening. A review of the evidence for hip adductor release used to prevent progressive hip subluxation in children who had cerebral palsy was recently
published. Despite difficulties related to study design, a few observations were made. Radiographic improvement after adductor release was seen in approximately 50% of hips. However, the clinical significance and correlation to improvement of pain, function, or activities of daily living has not been systematically evaluated. Children who have a smaller preoperative hip migration index have a decreased incidence of postoperative hip subluxation or progression of migration index. Specifically, preoperative migration percentages of less than 30% to 40% were associated with successful outcomes in 75% to 90% of hips. Reported complications were few, although unilateral hip adductor release was often noted to have an adverse effect on the contralateral hip [144].

When bony abnormalities such as femoral torsion, coxa valga, and deformity of the acetabulum have occurred, bony procedures may be necessary and are often performed in conjunction with soft tissue releases. In patients who have no marked deformity of the acetabulum, surgical emphasis is placed on correcting femoral abnormalities. Possible interventions include derotational osteotomy of the femur, correction of the neck–shaft angle (coxa valga), and shortening of the femur to decrease muscle forces across the hip [111].

In patients who have coexisting acetabular deficiency, pelvic osteotomy may be required. The Pemberton osteotomy or acetabuloplasty (Figs. 16 and 17) is indicated if a deficiency of the anterior and superolateral walls of acetabulum is present. The Salter pelvic innominate procedure is used for anterolateral acetabular deficiency. The Dega osteotomy is typically indicated for posterior hip dislocations. The modified Dega adds femoral or intertrochanteric osteotomies or open hip reduction (Figs. 18, 19).

![Fig. 16. Same patient as in Fig. 15 who has undergone a right proximal varus and external rotation–producing osteotomy and Pemberton periacetabular osteotomy with bone graft from the iliac crest.](image)
San Diego procedure is used for anteroposterior acetabular deficiency and includes a femoral osteotomy and soft tissue releases. The Bernese (Ganz) periacetabular osteotomy may be performed in adolescents and adults who have dysplastic hips that require correction of congruency and containment to the femoral head. This procedure may be combined with a proximal femoral osteotomy to provide uninvolved acetabular and proximal femoral weight-bearing surfaces. The Chiari procedure is typically a salvage procedure that places the femoral head under a surface of cancellous bone rather than articular cartilage and is recommended in older children who have

Fig. 17. Four-year-old girl who has lumbar myelomeningocele. Lateral uncovering of 50% of the right femoral head by the acetabulum and one fourth uncovering of the left femoral head.

Fig. 18. Same girl as in Fig. 17 after undergoing right open hip reduction with capsulorrhaphy, bilateral Dega pelvic osteotomies, and bilateral proximal femoral varus and external rotation–producing osteotomies.
severe dysplasia and possibly subluxation when no other reconstructive options are available. A Shelf salvage procedure uses a bone graft for added support to the femoral head. The merits and outcomes of these various procedures are debated [102,103,128,142,143,145–149].

In nonambulatory children who have minimal symptoms or seating difficulties, operative treatment of hip subluxation or dislocation is controversial. Operative treatment options are similar for children who have upper and lower motor neuron disorders with a few exceptions. For individuals who have Charcot-Marie-Tooth disease and hip dysplasia, the acetabular deficiency has been recommended to be repaired first, because a primary femoral derotational osteotomy in the setting of weak hip abductors may exacerbate a Trendelenburg’s gait. If femoral derotational osteotomy is subsequently needed, the surgeon is suggested to proceed with internal fixation and early mobilization, because spica casts may exacerbate hip weakness from prolonged immobilization [106]. In children who have spinal muscular atrophy, a high frequency of resubluxation after surgical intervention has been reported [117,118,150]. Therefore, surgical intervention for subluxed or dislocated hips in children who have intermediate spinal muscular atrophy is not generally recommended. However, if surgical intervention is believed necessary, a single-stage combined procedure of appropriate soft tissue release and bony reconstruction is pursued [25,117,118,150]. A review of hip disorders in children who have spinal cord injury noted that operative treatment should include release of soft tissue contractures and appropriate bony interventions with muscle transfers in a select group of patients. Postoperatively, a hip abduction orthosis rather than casting is recommended to reduce risk for skin breakdown [130]. In congenital hip dysplasia, surgical correction usually involves closed reduction with casting. This procedure should be considered when a Pavlik harness trial of 6 to 12 weeks has failed
or the patient is older than 6 months [134]. If closed reduction is not possible or a child has a more advanced deformity, open reduction may be considered. Open reduction may combine soft tissue release and femoral shortening with varus derotational osteotomy, with or without acetabular osteotomy.

Summary

A wide variety of neuromuscular diseases affect children. Despite the vastly different primary pathophysiologic mechanisms of these disorders, certain secondary musculoskeletal complications are shared. Scoliosis, bony rotational deformities, and hip dysplasia are some of the most common sequelae in children. Care providers must recognize the musculoskeletal abnormalities and understand the natural history and nonoperative and operative treatment options for these children to prevent progression and functional loss.

References


