Difficulty Walking in an Adult with Cerebral Palsy

BETH STEPANCZUK, MD
Assistant Professor, Department of Physical Medicine and Rehabilitation
University of Pittsburgh School of Medicine

MARY SCHMIDT, DO
Resident, Department of Physical Medicine and Rehabilitation
University of Pittsburgh School of Medicine

Clinical Vignette

DJ is a 50-year-old man with a history of spastic diplegic cerebral palsy (CP) also resulting in scoliosis, dysarthria, strabismus, and seizure disorder. He presented to the office complaining of progressive difficulty with walking. Additionally, he reported right anterior and lateral knee pain without mechanical symptoms that was worse with weight bearing activities.

DJ has seen his primary care physician biannually for health maintenance and hypertension. In the remote past, he underwent bilateral tendo-Achilles lengthening and ocular surgeries. He lives alone in a wheelchair accessible home and ambulates with hinged bilateral ankle foot orthoses (AFOs). He uses a friend’s old power wheelchair for community mobility and to sometimes get around at home. He had been managing his own cooking, cleaning, and laundry, but he now requires assistance from an aide as these activities have begun to take longer and consume more of his energy. Previously, he worked as a bank clerk.

Examination of the patient was notable for thoracolumbar dextroscoliosis with pelvic obliquity and spastic paraparesis. Modified Ashworth scores were 1+ at the hips and knees. There was bilateral patella alta, extensor lag, and pain with palpation of the right patellofemoral joint line. There was no evidence of clonus, joint effusion, lower extremity edema, or sensory impairment. Using a wheeled walker, he ambulated slowly and with effort. Upon completing a 50-foot walk, he had mild tachypnea. Stance phase in both legs was characterized by excessive hip flexion, knee flexion, and dorsiflexion. The hips appeared adducted and internally rotated.
He would like to remain ambulatory and independent as long as possible. He also intends to seek employment. He asked for a comprehensive evaluation by a physiatrist in order to achieve these goals.

Definition and Epidemiology of Cerebral Palsy

Cerebral palsy (CP) is one of the most common conditions managed by pediatric physiatrists. As individuals with CP mature and transition to adulthood, there is a growing need for physiatrists who can address patients’ functional concerns. Physiatrists have unique training to help adults with CP maintain functional independence. Although the impairments of men and women with CP resemble those of individuals with acquired brain injury, there are many differences. Therefore, it is important for physiatrists to be familiar with common mobility concerns and associated medical conditions faced by adults with CP. Furthermore, as adults with CP age, they seek fewer health care services in part due to the multiple barriers they encounter when navigating the health care system. These barriers include lack of wheelchair accessible offices, adjustable exam tables, and providers trained in disability awareness. A physiatrist may be the only physician that an adult with CP will visit.

CP is defined as, “a group of permanent disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems.” CP can be classified by type of motor impairment as well as distribution of limb involvement, with spastic diplegia being most common overall (Table 1).

The occurrence of CP is 1.5 to 4 in every 1,000 live births, and the prevalence is estimated to be 1 in 323 children. In 2006, a survey of children with CP conducted by the Metropolitan Atlanta Developmental Disabilities Surveillance Program showed 40% had intellectual disability, 35% had epilepsy, and 15% experienced visual impairment. Risk factors for CP include low birth weight, prematurity birth, disruption of blood flow and oxygen supply to the fetal brain, infection in the mother, and infection during infancy.

CP has been traditionally viewed as a pediatric disorder, even though adults with CP can have an average lifespan. Over half of children with CP walk independently, and interventions by pediatric specialists have traditionally focused on maximizing motor function and ambulation. There is a growing school of thought that pediatric treatment should focus on anticipation for long-term mobility and function rather than short-term function in childhood. Surveys of the CP population have found that mobility and independence are the most important factors for psychological well-being and quality of life.

Evolution of Gait Dysfunction

There is general acceptance of a so-called “inevitable” decline in function for people with CP as they age. While CP is a static neurologic condition, over time musculoskeletal imbalance can lead to increased workload and abnormal stresses, which in turn predispose the body to secondary conditions that affect previously mastered abilities. Age is correlated with a decline in mobility, as is the case in the general population, however adults with CP experience a decline earlier in life than adults without CP. There appears to be a bimodal distribution of individuals with CP who stop ambulating: one group declining before their 30s and another in their 60s. While some young adults with CP continue to experience improvements in their ambulatory ability over time, a study by Day et al. showed that after age 25, individuals with CP are more likely to see no improvement in ambulatory ability. Adults with CP have an increased likelihood...

<table>
<thead>
<tr>
<th>TABLE 1: Classification of Cerebral Palsy</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Spastic</strong></td>
</tr>
<tr>
<td>• Monoplegia — One leg predominantly involved</td>
</tr>
<tr>
<td>• Diplegia — Both legs predominantly affected, with minor involvement of arms</td>
</tr>
<tr>
<td>• Hemiplegia — Ipsilateral arm greater than leg involvement</td>
</tr>
<tr>
<td>• Triplegia — Hemiplegia with contralateral monoplegia</td>
</tr>
<tr>
<td>• Quadriplegia — All four limbs involved</td>
</tr>
<tr>
<td><strong>Dyskinetic</strong></td>
</tr>
<tr>
<td>• Athetosis — Involuntary slow, writhing movements of extremities</td>
</tr>
<tr>
<td>• Chorea — Quick, random jerking movements</td>
</tr>
<tr>
<td>• Choreaathetosis — Combination of athetosis and choreiform movements</td>
</tr>
<tr>
<td>• Dystonia — Prolonged, alternating contraction and relaxation of antagonist and agonist muscles often accompanied by abnormal posture</td>
</tr>
<tr>
<td>• Ataxia — Uncoordinated intentional movements</td>
</tr>
<tr>
<td><strong>Mixed</strong></td>
</tr>
<tr>
<td>• Characterized by a combination of spastic and dyskinetic patterns</td>
</tr>
</tbody>
</table>
of gait deterioration if they began walking at an age greater than three years, need an assistive device for household mobility, have bilateral body involvement, or experience falls. A study by Strauss et al. examined walking ability in a large cross section of children and adults with CP who received services from the State of California. Of ambulatory 60-year-old individuals in this sample, the majority had stopped walking within 15 years. Among the subjects who were nonambulatory at age 60, only 60% survived until age 70, suggesting a correlation between a decline in physical function and mortality.

Pathophysiology and Treatment of Crouch Gait

Assessment of abnormal gait patterns and their root cause is challenging in the clinic because the deviations occur in multiple planes simultaneously. Most general physiatrists do not have access to quantitative gait lab equipment and must rely on observational gait analysis. Video recording in slow motion can allow for more careful visual analysis of each body segment as the subject may not have the stamina to repeat a walk several times. Crouch gait is a common gait pattern in diplegic CP that warrants further discussion. Without intervention, this gait pattern is progressive and leads to loss of ambulation. The key feature of crouch gait is excessive knee flexion during the stance phase of the gait cycle.

There are numerous contributors to crouch gait: plantar flexion, knee extension or hip extension weakness, flexion contractures or spasticity at the hip or knee joint, and lever arm dysfunction in the setting of malrotation at the femur, tibia, and feet. Soleus muscle weakness is thought to be the most significant factor. During the early through midstance phases, the soleus muscle contracts eccentrically to slow forward progression of the tibia, resulting in a ground reaction force anterior to the knee that promotes knee extension. Weakness in the soleus muscle results in a ground force reaction that is posterior to the knee and anterior to the hip, driving both joints into flexion. (Figure 1) Soleus weakness may be a result of the primary neurologic condition of CP, or from heel cord lengthening surgery as a child. Crouch gait progresses over time due to elongation of the patellar and quadriceps tendons, continued pull of the knee flexors, weight gain of the individual, and inability of antigravity musculature to resist these forces.

Conservative measures targeting the root causes of an individual’s crouched position may limit progression. Targeted spasticity interventions, stretching, and strengthening play a fundamental role. In crouch gait, the psoas and hamstrings may be short and hypertonic. Botulinum toxin injections can be considered in these muscles to provide focal reduction in spasticity. A randomized controlled trial that involved botulinum toxin A or placebo injections in 66 walking adults with CP indicated that chemodenervation reduced muscle stiffness in the short term for the treatment group versus placebo, but it did not affect joint kinematics. This data suggests that several concurrent treatment modes may be required to improve gait pattern. Similarly, evidence is lacking in treatment paradigms involving passive range of motion, aerobic activity, and balance training. Progressive resistance training is commonly prescribed without a clear understanding of its value. A systematic review by Jeglinsky et al. revealed moderate effectiveness of lower extremity strength training on gait parameters in adolescents and adults with CP; however, the populations and protocols

Figure 1. Crouch gait is characterized by excessive ankle dorsiflexion, knee flexion, and hip flexion. Left: In crouch gait the ground-reaction force (GRF, shown as the vertical arrow) is directed posterior to the knee joint and anterior to the hip joint. In severe crouch gait, the muscles resisting the GRF (hip extensors, knee extensors, and ankle plantar flexors) are weak and may become elongated. The principles for the correction of crouch gait include lengthening of shortened muscle-tendon units and support or strengthening of weak, elongated muscle-tendon units. Right: Use of a ground-reaction ankle-foot orthosis (GRAFO) limits forward motion of the tibia in midstance, placing the GRF (vertical arrow) anterior to the knee, thereby keeping the knee in an extended position.
studied varied widely such that specific recommendations could not be made. In our clinic, we emphasize strengthening of the glutei, vasti, and soleus muscles to promote hip extension, knee extension, and plantar flexion. Ground reaction AFOs with a rigid anterior shell and dorsiflexion stop are commonly used for crouch gait if joint contracture is not present.

Individuals with severely crouched posture often have multiplanar bony deformities and musculotendinous distortion in several areas of the lower extremities, and undergo what is referred to as single-event multilevel surgery (SEMLS). Common components of SEMLS are muscle lengthening (psoas, hamstrings, or adductor muscles), rectus femoris transfer, femoral or tibial derotation osteotomy, posterior knee capsulotomy, patellar tendon shortening, distal femoral extension osteotomy, and stabilizing foot surgery. These procedures are often performed during childhood. Outcomes for the adult CP population are unclear. A retrospective cohort analysis of 97 ambulatory individuals with spastic diplegic CP who were >17 years old at the time of surgery showed improvement in walking based on kinematic data from three-dimensional gait analysis. However, 21% experienced deterioration in ambulatory function. Of note, subgroup analysis of the 28 subjects who exhibited a crouch gait pattern preoperatively was not performed. The study also reported a longer time to regain walking in adults postoperatively (average 19.7 months) compared to the usual recovery time for children (12 to 15 months).

Selected Contributing Factors to Gait Dysfunction and Management

Chronic Medical Conditions

It is important to emphasize a holistic approach when considering the reasons why an individual with CP may stop walking. The degree of physical disability is associated with having chronic medical conditions. Adults with CP receive fewer health screening tests, such as colonoscopies, pap smears, and mammograms, and experience increased mortality from neoplasms, with breast cancer death rates, three times higher than in the general population. Undiagnosed medical conditions, such as malignancies or endocrine disorders, can be overlooked in the early stages since adults with CP commonly experience fatigue.

Obesity and Cardiovascular Disease

Obesity prevalence has yet to be studied in the adult CP population, although there is an increased prevalence in the pediatric population. In general, the CP population has a more sedentary lifestyle from birth, making them predisposed to weight gain and metabolic syndrome. Records from 1986 to 1995 reveal a two to three times greater risk of coronary heart disease in people with CP compared to people without CP, and a recent survey published in the *Journal of the American Medical Association* by Peterson et al. reveals a higher prevalence of hypertension and other heart conditions compared to the general population. Capriotti found women with disabilities had less knowledge about cardiovascular disease risks and received fewer screenings despite being at higher risk given a decreased baseline activity level. In addition to a low baseline activity level, weight gain may lead to changes in functional mechanics, which compound painful conditions and fatigue.

Fatigue

Fatigue is the feeling of loss of energy, tiredness, exhaustion, or weakness. Fatigue is a chronic disability itself, as well as a contributor to other aspects of function. Jahnsen et al. studied fatigue in adults with CP and found a higher occurrence in people with self-reported, moderately severe CP, which suggests this population has a higher energy expenditure for mobility. Fatigue was associated with older age, lower physical function, no physical activity, and lower scores on the Life Satisfaction Scale. In crouch gait, energy cost for ambulation is higher with greater knee flexion angles. Typically, individuals can reduce their speed of walking to reduce oxygen consumption. However, people with spastic diplegia require heightened activity by antigravity muscles to maintain an upright posture during stance phase, regardless of gait speed.

Long-term commitment to a well-rounded exercise program is felt to be important in boosting physiologic reserve so that individuals can complete activities of daily living and participate socially. A small controlled study by Kim et al. of 21 adults with CP suggested that a treadmill walking program could improve overground walking velocity and energy efficiency. This suggests that endurance training in this population is feasible and could impact symptoms of fatigue. If conservative measures do not allow the individual to overcome the considerable oxygen requirements of walking, a wheelchair may be helpful.

Pain

Pain is one of the most common complaints of adults with CP. Survey data that include various types of CP reveal that joint pain is very prevalent in this population, estimated to be 28%
versus 15% in the general population. Of those who received treatment for their pain, only 18% reported satisfaction with their treatment. In Norwegian adults with CP, the mean age of pain debut was 21 years. Occurrence was lower in subjects who were younger than 30 years old (18%) compared to those 60 years old (40%). Pain was most commonly located in the back, hips, and lower extremities. Neck pain, shoulder pain, and headache were most common in dyskinesia. Foot and ankle pain were most common in diplegia, and knee pain was most common in quadriplegia. Back pain was equally reported in all types. Also, there was a statistically significant association between reduced range of motion and pain. A majority of participants reported reduced range of motion in at least one joint. Pain can contribute to or arise from spasticity, contracture, weakness, and falls. Common etiologies of pain are poor ergonomics and biomechanics while performing coordinated tasks due to deformity or impaired motor control, underlying weakness causing overuse of less affected muscles, and osteoarthrosis. To address pain complaints, the primary objective is to identify the pain generator(s) so that specific interventions can be applied. Common disorders of muscle and bone (particularly the spine, hips, and knees) experienced by individuals with CP are discussed below.

Musculoskeletal Conditions

Spasticity

Spasticity is the most common form of abnormal muscle tone in people with CP and is an impairment that arises from the original neurologic injury. In CP, muscles that cross two joints are more severely affected by spasticity. Daily range of motion exercise and prolonged stretching of the affected areas, sometimes with splints, create the foundation of spasticity management. Progressive resistance training of weak antagonistic muscle groups and stretching of hypertonic muscles is thought to reduce the effect of spasticity. Slow contraction velocity of antagonistic muscle groups reduces the stretch reflex of the spastic muscles. Oral antispasticity medications (baclofen, tizanidine, diazepam, or dantrolene) can be helpful in tone reduction but are often limited by side effects, including cognitive impairment, fatigue, or hepatotoxicity. Botulinum toxin injections to the psoas, hamstrings, adductors, rectus femoris, and gastrocnemius muscles are indicated when spasticity is causing pain or gait deviation, although ultrasound guidance may be needed for accurate placement. Intrathecal baclofen pump can be considered in the case of widespread, refractory spasticity.

Spine Disorders

Spondylolysis is a defect in the pars interarticularis, most commonly at the L5 level. Acquired spondylolysis can be caused by repetitive stress and an imbalance of forces between the lumbar and sacral vertebral segments. Low back pain is the most frequent complaint at presentation, however, the disorder can also be asymptomatic. Prevalence of spondylolysis is as high as 30% in the CP population and even higher with a previous selective dorsal rhizotomy. Dystonia in the lumbar spine and anterior pelvic tilt are associated with increased risk of this disorder. Associated conditions that contribute to spondylolysis include lumbar lordosis, pelvic obliquity, hip contractures, and femoral head subluxation. Initial imaging includes posteroanterior, lateral, and oblique lumbar spine radiographs. Advanced imaging, such as computed tomography (CT) scan, may be necessary to confirm the diagnosis. Management of this condition includes physical therapy, assistive devices for gait, orthotics to minimize toe walking, and botulinum toxin injections to the paraspinal muscles. A typical physiotherapeutic program involves core strengthening, pelvic stabilization, and stretching of hip flexors and plantar flexors in an effort to decrease anterior pelvic tilt. Home exercises that are often recommended include prone lying, core strengthening, and stretching of the hip flexors, hamstrings, and gastrocnemius muscles. For the patient who has failed conservative treatment or with neurologic compromise, surgical segmental fusion may be necessary.

Scoliosis occurs in ambulatory individuals with CP, although curvatures tend to be more severe in nonambulatory individuals with spastic quadriplegia. In childhood, scoliosis rarely causes pain. However, after many years of abnormal loading, pain related to spondylolysis may arise in adulthood. Importantly, advanced scoliosis can adversely affect lung function and, secondarily, functional mobility. Progression typically worsens during periods of rapid growth and occurs most commonly in the thoracolumbar regions.

Cervical stenosis is found in higher rates in the CP population. In particular, individuals with neck dystonia or athetosis are more susceptible to development of cervical spondylosis and myelopathy. Murphy suggests screening magnetic resonance imaging (MRI) every two years in high risk patients, starting in young adulthood. Management of cervical dystonia is important in prevention of this disorder. Treatment options include botulinum toxin injections, postural supports, and oral antispasticity medications.
**Hip Disorders**

Hip dysplasia should be high on any differential diagnosis for someone with CP who reports difficulty with weight bearing. The prevalence is estimated to be 28% in people with spastic CP, although the highest rates are reported in nonambulatory patients with quadriplegia.\(^{21,22}\) Hip adductor and flexor hypertonicity results in femoral anteversion and coxa valga, which drive the femoral head superolaterally. Persistence of these forces causes progressive hip dysplasia via flattening of the femoral head, which in turn can lead to osteoarthritis and, ultimately, subluxation. Adults with CP who have increased tone in the hip adductors and flexors are at risk for hip dysplasia, and AP pelvis radiographs can confirm the diagnosis. Treatment and prevention involves stretching of spastic hip muscles and strengthening surrounding muscles of the hip. Oral or intrathecal spasticity medications, botulinum toxin injections to the adductor longus and hip flexors, or phenol obturator neurolysis may be useful.\(^{22}\) Finally, surgical treatment may be necessary with total hip arthroplasty, tenotomy, or varus derotation osteotomy.

**Knee Disorders**

Patella alta is superior displacement of the patella on the femur and results in misalignment in the knee joint. It is often seen in crouch gait pattern where there are abnormal forces across the tibiofemoral and patellofemoral joints. Spasticity of the hip flexors, and excessive ankle dorsiflexion, leads to overstretching of the quadriceps muscles and infrapatellar tendon. Patella alta can be detected clinically by observing and palpating the knee. In static stance, the inferior border of the patella lies just above the tibiofemoral joint line. From a lateral viewpoint, a high-riding patella is suggested by the “camel sign,” in which one “hump” is the infrapatellar fat pad and the second “hump” is the patella. Additionally, extensor lag indicates quadriceps insufficiency and potentially patella alta. Extensor lag is the difference between active and passive knee extension in supine. Lateral plain radiographs at 30 degrees of knee flexion also can be used to evaluate the patellar height. The Insall-Salvati Index compares the patellar tendon length to the patellar bone length, where patella alta is defined as a ratio of >1.2.9. Prevalence is estimated at 58 to 72% of people with spastic CP.\(^{22}\) In childhood, it is often asymptomatic. Later in life, it can cause anterior knee pain. Complications include subluxation, dislocation, or stress fractures of the patella.\(^{11}\) Fragmentation of the lower pole of the patella may promote knee flexion contracture. Pain from patella alta can be addressed conservatively by taping techniques to improve the tracking of the patella, quadriceps strengthening, and stretching of the hip flexors and knee flexors. Slowing the progression of crouch gait may prevent worsening of knee pain. Surgical interventions for advanced cases typically involve patellar tendon advancement and distal femoral extension osteotomy to correct knee flexion contracture.

**Exercise for Health and Mobility**

Studies have shown that adults with CP who engage in regular physical activity have reduced risk of decline in mobility.\(^4\) When possible, weight bearing or closed kinetic chain exercises are best for maintaining bone health. Individuals with inadequate core strength should utilize physical assistance by another person, stabilization by their free arm, or seated, supine or prone positioning. For example, pushups can be performed from the knees or on an incline by standing and pushing from a waist-high countertop. Variations of the squat are a supine inclined leg press or sit-to-stand repetitions from a seat with a table-high surface in front of the exerciser to allow for progressive reduction in arm support to work on both leg strength and balance. Ankle weights applied to the wrist, or lifting gloves with a Velcro-secured grip to hold a barbell, can be used when strengthening paretic arms, although body weight alone is often sufficient in sedentary individuals to stimulate muscle growth. Recommended frequency of strength training is two to three times per week for each body area. Adequate rest between sets is required to avoid worsening of motor control from overfatigue.

Aerobic fitness training should initially consist of short and frequent bouts of light intensity work with incremental progression to achieve 20 to 40 minutes per aerobic session of moderate intensity, three days per week. Activities that involve simple motor patterns are walking, swimming, cycling, upper extremity cycling, stair climbing, rowing, and recumbent stepping with a NuStep. Individuals limited by imbalance may find seated, recumbent, and aquatic exercises to be the safest options.

**Vignette Outcome**

DJ completed six weeks of physical therapy, focusing on hip extensor, knee extensor, and abductor strengthening with concurrent stretching of the knee and hip flexors. He was transitioned to a comprehensive exercise program involving treadmill walking and strength training using weights. Bilateral custom-molded ground reaction force AFOs were prescribed. He was educated on ways to minimize the risk of cardiovascular disease. Six months after initial evaluation, DJ reported less pain and an improved ability to perform activities of daily living, allowing him to engage in volunteer activities.
Reference


About the Department of Physical Medicine and Rehabilitation

• UPMC is ranked by U.S. News & World Report as one of the nation’s best hospitals for rehabilitation.

• The Department of Physical Medicine and Rehabilitation is consistently a top recipient of NIH funding for rehabilitation-related research.

• The Spinal Cord Injury Program at UPMC is one of only 14 in the country selected by the National Institute on Disability and Rehabilitation Research as a model for other rehab providers.

• The Brain Injury Program at UPMC is one of only 16 in the country selected by the National Institute on Disability and Rehabilitation Research as a model for other rehab programs.

• Department clinicians lead UPMC’s rehabilitation network of more than 70 inpatient, outpatient, and long-term care facilities — one of the country’s largest.

Learn more about how UPMC is transforming rehabilitation.