CHAPTER **23**

CEREBRAL PALSY AND EQUINUS

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INTRODUCTION

Cerebral palsy (CP) is an "umbrella term covering a group of nonprogressive but often changing motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development."[1] With multiple motor and sensory deficits, it is no surprise that CP is a leading cause of chronic childhood disability. Despite advances in treatment, the incidence of 2 per 1,000 live births remains and various treatment options continue to be explored.[2]

Etiology

Lacking a single etiology or genetic origin of CP, various etiologies have been identified. The pathobiology of CP subtypes can be separated into different gestational age groups, most falling within the categories of preterm and term infants. Preterm births account for 12.7% of cases in the US, and because of the associated white matter disorder, are a major cause of spastic CP.[3] Nelson describes the incidence of white matter disorder and preterm birth to correlate with bouts of infection, which lead to infant sepsis from placental inflammation (chorioamnionitis). The cytokine cascade and the inflammatory mediators of infection have both been linked to damaging effects on the oligodendrocytes that comprise the white matter.[3]

CP seen in term infants is said to stem from perinatal ischemic stroke or congenital malformations. Often presenting with nonspecific symptoms, such as neonatal seizures and apneas, a resultant diagnosis of CP from stroke is frequently not recognized until after the perinatal period. Studies show that the events occurring within the placenta "hold the key to many mysteries of perinatal stroke," and the identification factors leading to them are still unknown.[3] Congenital malformations of the head, oropharangeal malformations, and gut atresias are more common in children with CP, suggesting a contribution of prenatal processes to some proportion of the disorder. Twins and other multiple births have also shown a higher incidence of CP, secondary in part to prematurity, but also to the death of a co-twin. Research suggests that monozygotic twinning with death of a twin is a major risk factor for CP in the surviving twin. With the death of one twin, a supposed vascular collapse occurs, leading to potential ischemic events and malformations in the survivor.[3]

Classification

Defined by their specific presentations and cerebral lesions, CP can be further classified to four clinical subtypes. Spastic, the most common presentation, is seen in 65% of all CP cases, and is characterized by hypertonia as a result of damage to the cerebral cortex.[4] The most common contracture seen, whether dynamic or static, is that of the gastro-soleal complex. As it is found to be tightened, weakened, and or sometimes shortened, a resultant equinus deformity is frequently seen. Usually coupled with spastic hamstrings and hip flexors, an unstable gait with a limited base of support often leads to long-term lower extremity complications.[5]

Making up approximately 25% of all CP cases, the athetoid or dyskinetic patient has a mixture of hypotonia and hypertonia. Seen with damage to the midbrain, this subtype is defined mainly by involuntary motions from muscles exhibiting normal mobility. These patients often have trouble holding themselves in an upright position, thus making the act of walking a difficult task. Athetoid patients also experience difficulty performing specific movements with their hands and feet.[4] Simple daily activities such as holding a toothbrush or pushing the gas pedal are often difficult maneuvers.

The ataxic group of CP (~5%) stems from damage to the cerebellum, and manifestations are directly related to a lack of coordination and motor skills. These patients will have obvious impairments when they attempt simple movements such as writing, typing, walking, and balance.[4] Hand to eye coordination is usually impaired, affecting many daily activities.

The fourth subtype of CP is more of a mixed group, often defined as rigid (~5%). Seen in more severe cases of CP, these patients are found to have widespread cerebral impairment. Clinical manifestations include a constellation of symptoms shared by the other three subtypes.[3] Often the patients exhibit a spastic equinus deformity that is recalcitrant to conservative therapy. More extreme types of intervention are often necessary to help these patients improve quality of life.

Equinus

Regardless of etiology or subtype, as podiatric physicians it is imperative to understand the most common deformity in children with CP. Defined as a "functional gait, or standing pattern characterized by disproportionate weightbearing on the metatarsal heads and increased plantarflexion at the ankle," equinus encompasses a number of problems that alter the gait cycle.[5] Over the years, research has focused not only on treatment options, but understanding the different forms and presentations of CP patients.

Because walking ability has been defined as a main concern of most parents of CP patients, the deleterious effects of long term equinus gait have been researched and identified. Described as spring loaded, a resultant equinus posture and gait develops secondary to the dominating calf spasticity.[5] Secondary complications include painful plantar metatarsal head calluses, decreased endurance and distance of ambulation, as well as lateral ankle instability. As the equinus foot attempts to clear the floor during swing phase, increased hip and knee flexion, circumduction, vaulting or increased external rotation of the lower extremity are necessary compensations. Due to these multi-level compensations, the energy requirement of walking for patients with CP is increased by two-fold.[5] As imagined, this factor alone can explain the early fatigue experienced with daily activities for CP patients. Some of the main reasons for concern with walking ability stems from the desire to achieve or maintain independent functioning.[6] Many treatment modalities are thus aimed at achieving better daily functioning through lower energy requirements and more efficient, biomechanical movements.

Before deciding upon appropriate treatment modalities, a proper diagnosis is imperative. First recognizing the difference between true equinus (plantarflexion of the foot and ankle with respect to the leg, seen in stance and swing phase) and apparent equinus (toe-walking secondary to increased hip and knee flexion) is essential to achieving successful outcomes. Stated by Goldstein, "in CP there is a race between bone and muscle growth that is only concluded at skeletal maturity. The pacemakers for this race are the bony physes (growth plates). In the case of the calf muscle, with growth of the proximal and distal tibial physes the spastic muscle will always come in second. Most equinus interventions are aimed at equalizing this race."[5]

TREATMENT

Physiotherapy

When deciding on physical therapy for intervention, it is recommended to ascertain the type of contracture the patient possesses, either dynamic or static. Those that have dynamic contractures have been shown to better respond to stretching, strength training, and orthoses. For these children, calf spasticity and/or dorsiflexor weakness are the underlying etiologies.[6] By directly addressing weakness, recent studies have shown structured strength training programs can provide improvement in strength and stability.[7] In an attempt to overcome both spasticity and weakness, serial casting of muscles in their stretched position has also become a popular form of treatment.[8]

For those patients with static contractures electing conservative therapy, it is important to recognize the significant shortening of their gastro-soleal complex. For these patients, a sustained muscle stretch (versus manual) of long duration has been supported for obtaining positive results.[8] It has also been stated that splinting a muscle in its elongated state will increase the number of sarcomeres by 19%.[9] On the other hand, some studies suggest only minimal improvements on length and range of motion, less than 10 degrees.[8] Through extensive review of the literature, the evidence for the effectiveness of passive stretching remains mixed.

Botulinum Toxin A

With spasticity to blame for much of the equinus deformity seen in CP patients, this form of intervention is targeted directly at the neuromuscular junction. Approved by the Food and Drug Administration for adjunctive treatment of dynamic focal leg spasticity, botulinum toxin A has to date, provided mixed results. Through intramuscular injections, a focal weakness is created by blocking acetylcholine release, hence directly reducing the power of spastic contractions. The injections are said to indirectly modify the reflex activity by altering muscle spindle sensitivity.[2]

More specific to the gait cycle, botulinum toxin A studies have shown reductions in equinus, improved developments of heel strike, and improved mid-stance and mid-swing dorsiflexion. By reducing muscle stiffness, the foot maintains a better position from initial stance and is able to conserve power for late stance and push off.[10] Achieving these results, however, has been debated in the

literature. Most physicians agree that the ideal candidate has a confirmed dynamic spasticity, and supplemental casting or orthoses is crucial.[10] Ultrasound guided injections have been shown to provide better results with avoidance of adverse reactions and errors. Significant improvements in muscle tone and gait patterns have been reported, with average durations of about 3-6 months. After six months, the motor end plates are said to begin releasing acetylcholine again, and contractures gradually return.[11]

Surgery

When faced with a deforming equinus contracture, modalities such as physiotherapy and botulinum toxin A often fall short of success. However before deciding upon surgical intervention, the nature of the equinus must be identified. As mentioned previously, the dynamic contractures should initially be addressed with more conservative measures.

Conversely, musculoskeletal modeling software has shown the gastro-soleal complex to be significantly shorter than average in static contractures, thus these tend to require more aggressive interventions.[6]

Defined as the inability to dorsiflex the ankle to neutral position with the knee extended, static deformities present with the need for a more definitive therapy such as the gastrocnemious recession. By surgically lengthening the limiting factor, children are returned to more anatomic positions for more efficient cycles of gait. Confirmed by length comparisons before and after gastrocnemious recession procedures, the calf muscles are no longer abnormally short postoperatively.[6]

Because some cases of equinus are the result of weakness and imbalance of invertors and evertors, an associated varus or valgus deformity is often present.[5] More specifically, hindfoot valgus is said to be just as common as equinus in children with CP. Described as a "complex three-dimensional malalignment of the subtalar joint," surgical management is often necessary. Correcting the plantar-flexion of the talus and calcaneus, along with valgus and external rotation of the calcaneus often requires procedures such as subtalar joint arthrodesis or Evans osteotomy. Various studies have evaluated the effectiveness of these procedures, and have found reductions in abnormal pressure distributions, callosity, and overall pain.[12]

CONCLUSION

Overall, it is important to remember that each patient has a unique deformity and goals for therapy that must be explicitly understood and catered to. Each patient is an individual and it is imperative they are viewed as such. No single course of intervention exists that can be the answer to every CP presentation. Specific patient goals must be identified and reviewed for realistic expectations. "CP is a disorder that manifests differently in each child, and for optimal results of interventions an individual analysis is needed together with an individual plan for treatment."[13]

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