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Adults suffering from Orthopedic Conditions with Cerebral Palsy

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Introduction

The most common cause of physical disability in childhood is Cerebral palsy (CP). The overall society prevalence continues to increase due to increasing survival of preterm infants and increasing survivorship into adulthood. It is estimated that in the United States between 700,000 and 1 million adults have CP however the exact number is yet unknown [2]. Children suffering from CP have many transformation and they face many challenges. Children who have CP encounter with “premature aging” they are more liable to have a more speedy reduction in their mobility and walking as compared to other healthy population.

Patients with functional levels, Gross Motor Function Classification System (GMFCS) levels II to V, have been shown to lose some gross motor function, as measured by GMFM. Although CP is considered a nonprogressive condition of the brain, the musculoskeletal components tend to worsen and deteriorate over time.[3] Primary impairments of CP include abnormal muscle tone, loss of selective muscle control, impaired coordination and balance, weakness, and loss of sensation. These primary impairments coupled with the effects of growth, time, and developmental delays negatively affect the musculoskeletal system. Weakness and tone abnormalities may lead to progressive contractures. Persistent tone, increased body mass index, contractures, and lever arm dysfunction may lead to increased loads across joints with subsequent early arthritis and pain. [4]

Orthopedic care of adults with CP has not been well documented in the literature. There are few institutions in the country equipped to fully care for adults living with CP who have orthopedic issues. Orthopedic care for children with CP within the environment of children’s hospitals is often appropriate until ages 16 to 21 years. Many families with children living with CP experience the “falling off the cliff” that occurs during the transition from pediatric to adult health services. With more adults with CP living longer lives, new or increasing problems need to be addressed as we seek to improve quality of life in this patient population with focused treatments to decrease pain, maintain activity, and maximize function.[5]

Chronic Pain

Chronic pain due to early arthritis, especially of the neck, back, and lower extremities is relatively common in most of the adults with CP, with an estimated 25% experiencing degenerative joint disease in their 20s.Because the joints are not experiencing full range of motion with increased joint forces due to spasticity, they tend to wear out much quicker than in the general population. Solutions to pain are multimodal and include spasticity management (botox and oral medications such as baclofen and valium),

massage/chiropractic/healing touch, wheelchair modifications, and frequent changing of positions. Although pain medications are an option, substance abuse is an issue along with worsening of constipation. There may be a role of cannabinoids in the treatment of pain in people living with CP, although this must be further studied.

Spine Conditions

Neuromuscular scoliosis is a spine deformity that possibly has the most effect on longevity of the patient with CP. The cause of scoliosis in CP is not entirely clear, but it is thought to be due to a combination of muscle weakness, truncal imbalance, and asymmetric tone in paraspinous and intercostal muscles. It is more common in GMFCS IV and V patients. Unlike adolescent idiopathic scoliosis, a very high percentage of neuromuscular curves tend to severely progress with time, resulting in adverse effects on the heart and lungs, as well as leading to pelvic obliquity, which affects seating and leads to skin breakdown and decubitus ulcers.[6] Initial treatment of neuromuscular scoliosis is typically seating and wheelchair modifications. Bracing with spine orthoses may be offered but have not been shown to be effective. Once neuromuscular curves reach 50, they tend to rapidly progress. Posterior spinal instrumentation and fusion (PSIF) is the mainstay of treatment of severe neuromuscular curves. The goals of PSIF are to straighten the spine, balance the spine and trunk, lower the risk of progression, allow for easier sitting and positioning, relieve pain, decrease other complications including decubiti and lung disease, and improve quality of life. It is typically recommended in adolescence; however, many parents hesitate to pursue surgery until obvious curve progression. Although surgery can be performed in older adults, there is a higher complication rate, especially in patients with multiple medical problems. Yet, the benefits of surgery seem to outweigh the risks of complications as demonstrated through several high-quality studies published by the Harms study group. In most recent years, some CP programs have moved toward systematic processes of critically evaluating patients in a multidisciplinary setting to ensure the right surgery is performed on the right patient, thereby minimizing postoperative complications. Spinal stenosis or a narrowing of the spinal canal can lead to neurologic deficits from spinal cord compression ormyopathy. The most common type of spinal stenosis in adults with CP is cervical stenosis. Significant movement disorders such as dystonia or athetosis can contribute to development of cervical myopathy due to contorsional head and neck postures. In patients with dystonic CP, there is an 8-fold increase in degenerative joint disease and instability potentially leading to radicular pain,myopathy, and neurologic deficits.[7] Providers should be wary of spinal stenosis in patients complaining of loss of function. In patients at higher risk, serial neurologic examinations and MRIs every 2 years should be performed for surveillance. Prevention through medical management of dystonia is recommended along with seating and positional modifications. Surgical decompression (anterior, possibly both) and fusion may be necessary for rapidly progressing cases, those that fail to respond to conservative treatments, and if progressive functional loss is present. Providers caring for adult patients with CP need to be aware that increased spasticity, declining motorf unction, and changes in bowel and bladder function may represent a myelopathic picture as opposed to a change in neurologic function simply due to aging. Thoracic and lumbar stenoses are not as common as cervical stenosis with dystonia; however, they may be associated with progressive scoliosis, especially because what was initially an adolescent type of neuromuscular scoliosis erodes into a superimposed degenerative spine

deformity. Again, this condition should be suspected in any patients with myelopathic symptoms and loss of function. Conservative treatment includes nonsteroidal antiinflammatory medications, epidural injections, and physical therapy. Progressive pain and neurologic loss require surgical treatment with posterior spinal fusion and decompression. [8] Spondylolysis and spondylolisthesis are other common spinal conditions that affect the adult CP population. Spondylolysis is an acquired condition involving a stress fracture through the pars interarticularis, thought to be due to repetitive hyperextension. In the general population, 6% of people are affected. The prevalence is 20% among GMFCS I to III patients. This number is higher after selective dorsal rhizotomy. Typically, spondylolysis is responsive to conservative treatment including physical therapy and activity modifications; however, progression to spondylolisthesis may require L4-S1 posterior spinal fusion.

Hip Conditions

Neuromuscular hip dysplasia is a significant issue in children with CP. The incidence varies between 10% and 90% and increases with increasing GMFCS levels, most commonly affecting GMFCS IV and V. Untreated hip dysplasia in patients with CP can lead to frank dislocation over time due to abnormal forces acting on the femoral head. This eventually leads to degenerative joint disease and pain. Although young patients with hip dislocation may be asymptomatic, degenerative joint disease of the hip is the leading cause of pain in adults with CP, affecting up to 50% of adults living with CP. Pain from hip degenerative joint disease can lead to severe loss of quality of life. Treatment in children is focused on preventing dislocation and reducing dislocated hips before femoral head and acetabular dysplastic changes occur. Moderately high success rates are reported in the literature, which has led to a national push for formalized hip surveillance with serial radiographs until skeletal maturity. Once hip dysplasia, hip arthritis, and pain are present, treatment options are limited to either hip salvage or reconstruction; therefore, most orthopedic surgeons are united on taking a preventative approach. [9]

Proximal femoral resection was first reported by Castle and involves an osteotomy less than the level of the lesser trochanter with interposition of the abductors sewn over the acetabulum and the vastus lateralis over the femur. This procedure is meant to eliminate the source of pain. The McHale procedure moves the proximal femur laterally away from areas of impingement. The surgery involves removal of the femoral head (“Girdlestone” procedure) in combination with a valgus proximal femoral osteotomy. [10] These procedures are reserved for nonambulatory patients, including those who cannot craw. Although these procedures have been found to offer good pain relief, they are not without complications. Heterotopic ossification and proximal migration are the most commonly reported complications. [11] Both procedures have been found to provide good pain relief and improve seating and hygiene, although most studies recommend proximal femur resection due to decreased complication rates.

Total hip arthroplasty is another option for painful CP hips; however, this procedure is considered to be high risk with many pitfalls. Severely dysplastic hips may make acetabular implantation technically difficult and impossible. Nonambulatory patients with CP may have too small femurs for implants. Loosening and fractures may result from osteopenic bone leading to poor bone stock. Repeat dislocation may occur due to spasticity. Historically, there has been bias against arthroplasty in CP with few surgeons comfortable with performing the procedure in this patient population. Although less commonly performed, data do show this procedure has a

high degree of patient satisfaction in addition to good pain relief. [12] A variation on total hip arthroplasty has been pioneered at DuPont, which involves a shoulder prosthesis interpositional arthroplasty. This procedure has been used in patients who have had a prior salvage procedure (Castle or McHale) but had recurrent hip pain. This procedure has been found to provide great relief of pain while also having high caretaker satisfaction. [13]

The knee is a common source of pain and gait dysfunctions in patients with CP. Most common orthopedic knee pathologies seen in adults with CP includes patella alta, knee flexion contractures, patella subluxations/dislocations, and inferior patellar pole fractures.

Foot and ankle deformities in adults with CP are similar in nature to those in the pediatric population; however, contractures and deformity tend to be much rigid and less responsive to soft tissue management.[14] These issues lead to difficulty with shoe wear, pain, and contribute to walking decline. Treatment goals differ among ambulatory and nonambulatory patients. In ambulatory patients, treatment is focused on providing stability for ambulation and function, preventing pain and further deformity. In the nonambulatory patient, having a braceable and pain-free foot is of utmost importance. The following are the common orthopedic deformities found in an adult patient with CP. Equinus is the most common type of foot and ankle deformity in the CP population. It is due to a shortened achilles and gastrocnemius/soleus complex tightness. Initial treatment of stretching and/or botox may be tried in adults but is not typically successful in a real contracture. Surgical treatment involves either lengthening of the fascia or tendoachilles lengthening. Types of fascial lengthening include the Strayer, Baker, or Vulpius techniques. Equinus alone is rarely seen in the adult CP population and typically found in combination with varus and valgus foot deformities as discussed later. Calcaneus deformity or pointing upward of the foot is usually iatrogenic in nature due to overlengthening of the achilles. Nonoperative treatment includes the use of floor reaction orthosis. Surgical treatment includes muscle transfers to the heel, heel cord tenodesis, and a calcaneal osteotomy with or without muscle transfer, but none of these procedures are highly effective. The best treatment of calcaneus deformity is prevention by not overlengthening Achilles tendons in children with CP. Equinovarus Varus deformity is the inward curvature of the heel. Equinovarus deformity is common in those with hemiplegia. Rigid deformities place stress on the lateral aspect of the foot, causing callosities and pain. [15] Surgical treatment options include the following: frost lengthening of the posterior tibialis muscle, split posterior tibialis tendon transfer, split anterior tibialis tendon transfer, Multiple osteotomies of the foot, including calcaneal, cuboid, cuneiform, and first metatarsal, and triple arthrodesis are more typically indicated in adults with CP due to the usual rigid deformities. Equinovalgus deformity of the foot is most commonly seen in diplegia and quadriplegia. Often called the “flatfoot,” it is commonly associated with midfoot break. Treatment options for the valgus foot include peroneal brevis tendon lengthening (with other procedures), calcaneal lengthening, subtalar arthrodesis, talonavicular arthrodesis, and triple arthrodesis.

Bunion deformities Dorsal bunion and hallux valgus are 2 types of bunion deformities common in adults with CP, affecting the first metatarsophalangeal joint and potentially leading to pain and difficulty with shoewear. Orthopedic surgery to treat bunions aims to improve stability by fusing the joint.

Summary

In the pediatric population, orthopedic surgery to treat neuromuscular foot and ankle deformities typically involves muscle lengthening and transfers or a combination of those with correction of bony deformities through osteotomies. In adults, tendon lengthening at the ankle may not have much of an effect. Osteotomies are often necessary but may not be powerful enough to correct the magnitude of the deformity. Arthrodesis can and should be used more frequently.

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