

# Health Care Transition for Youth with Cerebral Palsy

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**Abstract:** *In this review we draw from clinical experience and a review of the literature to describe important issues for the primary care provider to address during health care transition for youth with Cerebral Palsy (CP). Children and youth with CP are almost always living well into adulthood. Health care transition services for persons with CP must provide support to help them access adult specialty services and overcome insurance barriers while managing primary health issues associated with CP in the young adult such as spasticity. Moreover, providers caring for young adults with CP must carefully screen for and treat common co-morbidities of CP including joint and muscle pain syndromes, constipation, Gastro-Esophageal Reflux Disease (GERD) and behavior issues. Primary care and specialty services for adults with CP are not widely available due to issues related to provider training, insurance, and family and patient related barriers. Providers involved in health care transition for youth and young adults with CP must be active advocates for them in the adult health care system.*

## Introduction

Cerebral Palsy is a syndrome of motor impairment with a complex range of abnormalities in muscle tone from spasticity to hypotonia or weakness to choreoathetoid or other dyskinetic movement disorders.<sup>1</sup> The definition of CP is “a disorder of movement and posture due to a defect or lesion of the immature brain.”<sup>2</sup> CP is actually a group of disorders of movement and posture, causing activity limitations, which are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by seizures or disturbances of sensation, cognition, communication, and perception.<sup>3</sup> The incidence of CP is 2.12 to 2.45/1000 live births, which has increased over the past 40 years. In the 1970s the incidence was 1.7 per 1000 live births, and this has increased gradually until the 1990s when the incidence was 2.4 per 1000 live births.<sup>4</sup> A strong association with socioeconomic status is observed in studies in the UK, with an incidence of 3.33 per 1000 live births in the poorest quintile compared to 2.08 in the most affluent quintile.<sup>4</sup> Male infants form a small majority (58%) of those diagnosed with CP. Over 90% of cases of CP arise during gestation and can be attributed to intrauterine stroke due to one of a number of causes including prothrombotic polymorphisms, intrauterine infection and chorioamnionitis from Cytomegalovirus (CMV) or Rubella. Cerebral Palsy can also be caused by brain malformations associated with genetic syndromes. CP is associated with prematurity; with increasing rates seen in infants born weighing < 1,000 grams. Only 5-10% of CP is attributed to perinatal events such as birth asphyxia, neonatal jaundice, antepartum hemorrhage, neonatal infection, and instrument assisted birth. Only 5% of CP is attributed to postnatal causes such as meningitis or Shaken Baby Syndrome.<sup>4</sup>

The life expectancy of those with CP has risen dramatically over the past 40 years.<sup>5</sup> Life expectancy for persons with CP is related to the degree of severity of physical or cognitive disability.<sup>6</sup> Over 98% of people with mild CP reach the age of 30 years. Among those with severely disabling CP, two-thirds reach the age of 20 and 58% reach the age of 30. Similarly, among those with an IQ < 50, two-thirds reach the age of 20 and 60% reach the age of 30.<sup>6</sup> Because of the increased life expectancy, the adult population of persons with CP had grown dramatically. Currently, it is estimated that over half of the 800,000 people in the US with CP are adults.<sup>7</sup> Therefore, the vast majority of children with CP will require transition services to adulthood and ongoing services in the adult health care system.

## Important Transition Issues

The definition of appropriate transition, as stated in other articles in this issue, is “that Youth with Special Health Care Needs (YSHCN) receive the services necessary to transition to all aspects of adulthood, including from pediatric to adult health care, from school to employment and to independence.”<sup>8</sup> *Table 1* lists the most common barriers to successful transition for youth with CP. Barriers to appropriate transition start in adolescence before transition. Due to a lack of knowledge or experience on the part of pediatric providers of the clinical issues adults with CP face and their lack of knowledge of how the adult health care system works, few

### Table 1 Barriers to Transition

Termination of pediatric services with ineffective transition to an adult medical home

- Lack of education about services in the adult system
- Lack of education on the adult services
- Lack of an adult medical home model

Access to appropriate adult health care providers

- Lack of primary care providers with experience in CP
- Lack of adult rehabilitative services specialists with experience in CP

Inadequate preparation of YSHCN/families for transition

- ‘Letting go’ of adolescent by parents or caregivers
- Cognitive disability of the patient
- Developmental immaturity of patient

Insurance Coverage

- Loss of SSI income due to rule changes at 18 years of age
- Ineligibility for the health insurance of caretakers
- Lack of adult health care providers who accept government insurance

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pediatric providers educate families of children with CP on what to expect when they leave pediatric services. Moreover, there are few adult health care providers with interest or experience in the management of people with CP, making it difficult for pediatricians to know how to transfer care to adult providers. Lastly, individuals with CP may have cognitive impairment or immaturity, requiring additional support to access appropriate health care services. For children these supports are provided by the family and by special support programs, e.g., Title V care coordination programs, available in the pediatric health care system. These special programs do not exist in the adult health care system. If the family is unable or unwilling to provide these supports for the adult with CP, they may experience significant barriers accessing appropriate services.

### **Availability of CP Trained Specialists**

Pediatric physical therapists, physiatrists and pediatric orthopedists commonly receive specialized training in the comprehensive management of the motor impairments of CP. Therefore, most children with CP residing in urban areas have access to a pediatric team of specialists that provides therapies, orthosis and orthopedic surgeries to reduce muscle spasticity and promote optimal growth. Once the child with CP ages out of the child health care system into the adult health care system, he/she experiences a dramatic decrease in the availability of specialists with training and experience in the management of CP.<sup>9</sup> Studies in Canada demonstrate that access to appropriate primary and specialty care is limited for adults with CP and hospitalization rates are much higher than expected.<sup>9</sup> One reason for the lack of services is that the training in the care of adults with cerebral palsy for adult-oriented therapists, neurologists, orthopedic surgeons and physiatrists is lacking and is more focused on acquired neurologic or orthopedic disease. One parent in the Jacksonville Health And Transition Services Program (JaxHATS) at the University of Florida–Jacksonville said, “When we left pediatric care, it was as if someone flipped the switch and turned the lights off.”<sup>10</sup> This parent searched for over three years before finding a primary care physician who had expertise in CP.

### **Common Conditions Requiring Management**

Adults with CP frequently experience serious co-morbidities that impact their daily functioning or can increase their risk for premature death. The most common conditions that occur with CP are listed in *Table 2 (p.46)* and include seizure disorders, GERD, chronic constipation, nutritional deficiencies, pressure ulcers, joint contractures and dislocations due to spasticity, scoliosis, osteopenia, and urologic disorders. While these conditions are much more common among persons with CP than in the normal adult population, they often manifest subtly. Diagnosis of these co-morbid conditions is complicated by the lack of communication in patients with cognitive impairment or the differential response to these co-morbid conditions by persons with severe physical disabilities. Moreover, the diagnosis and management of these

co-morbid conditions sometimes requires additional time and expertise by the primary care or specialty provider, especially among persons who are non-verbal or cognitively impaired. Primary care and specialty providers need to gain expertise in the special considerations for diagnosis and management of common co-morbid conditions in persons with CP.

In adults with CP, spasticity and other factors can lead to overuse syndromes and chronic pain and deterioration of motor performance. In fact without intervention, functional ability deteriorates in adulthood for a majority of persons with CP.<sup>7</sup> Also, as persons with CP age, they acquire other common health conditions such as cardiovascular disease, breast cancer, etc. Studies show that providers are less likely to follow preventive screening guidelines in adults with disabilities nor do these patients receive evidence based interventions for serious chronic health conditions.<sup>11-13</sup> As a result, persons with CP are more likely to be diagnosed later and die from cardiovascular disease and cancer than normal adults. Strauss studied mortality rates in adults with CP in California compared to adults in the general population and found that persons with CP had standardized mortality ratios 4.7 times higher for breast cancer and 3.6 times higher for ischemic heart disease.<sup>5,14</sup>

**Spasticity Management** – Persons with Cerebral Palsy have damage to the cerebral cortex from which the motor neurons originate, causing decreased inhibition of the spinal-motor reflex, increased muscle tone, spasticity, muscle contractures, muscle weakness and/or movement disorders.<sup>15</sup> The upper motor neuron syndrome and spasticity can result in multiple orthopedic health problems for the adult with CP including worsening scoliosis, joint contractures, decreased ambulation, overuse injuries, other muscle and joint-related pain syndromes, and decreased use of the upper extremities. Management goals to treat spasticity and Upper Motor Neuron (UMN) syndrome include the need for increased ease of care (e.g., perineal hygiene); prevention of pressure ulcers and skin breakdown; relief of muscle spasms, limiting progression of contractures, hip dislocations and scoliosis; improved seating and positioning; improved motor performance; and improved ability to perform activities of daily living or attend school or work.<sup>16</sup>

The primary care provider should be familiar with the stepwise and multi-modal approach to spasticity/UMN management, which includes medical management of spasticity to interdisciplinary management with therapists, orthopedic and neurosurgeons and physiatrists to coordinate the care. The initial level of treatment usually includes preventive measures such as stretching and massage as well as preventing medical issues that can make spasticity worse (e.g., constipation, pain, urinary tract infection, decubiti, and others.). Most youth and adults with moderate to severe CP need ongoing daily stretching and exercises directed by physical and occupational therapy to optimize function over the course of their lives.

A second level of intervention includes positioning using serial casting or orthotics, which can be directed by physical

**Table 2 Co-Morbid Conditions in Persons with Cerebral Palsy**

<p><b>Cognitive Impairment</b></p> <p><b>Mental Health Disorders</b></p> <ul style="list-style-type: none"> <li>Attention Deficit Disorder</li> <li>Depression</li> <li>Behavioral syndromes</li> </ul> <p><b>Neuromuscular Challenges</b></p> <ul style="list-style-type: none"> <li>Spasticity</li> <li>Dysarthria</li> </ul> <p><b>Neurologic Disorders</b></p> <ul style="list-style-type: none"> <li>Seizure disorders</li> <li>Sleep disorders</li> <li>Aphasias</li> <li>Visual and hearing problems</li> </ul> <p><b>Orthopedic Issues/Bone Metabolism Disorders</b></p> <ul style="list-style-type: none"> <li>Scoliosis</li> <li>Spastic hip dysplasia</li> <li>Degenerative Joint Disease</li> <li>Osteopenia-fracture risk</li> </ul>	<p><b>Gastrointestinal/Nutritional Issues</b></p> <ul style="list-style-type: none"> <li>Gastro-Esophageal Reflux Disease</li> <li>Chronic constipation</li> <li>Nutritional deficiencies and inadequate calorie intake</li> </ul> <p><b>Urologic Impairment</b></p> <ul style="list-style-type: none"> <li>Bladder spasticity</li> <li>Incontinence</li> <li>Recurrent Urinary Track infection</li> </ul> <p><b>Oral Hygiene Concerns</b></p> <ul style="list-style-type: none"> <li>Preventive dental care</li> <li>Mouth breathers</li> <li>Palate abnormalities</li> </ul> <p><b>Integument Challenges</b></p> <ul style="list-style-type: none"> <li>Decubiti</li> <li>Spasticity/Contractures</li> </ul> <p><b>Pain Management</b></p> <ul style="list-style-type: none"> <li>Bone and Joint Disorders</li> <li>Muscle Spasms</li> </ul>
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and occupational therapists. Botulinum toxin is injected into specific muscles that are spastic to allow positioning and decreased contractures. Oral medications to reduce spasticity, such as benzodiazepines, baclofen, tizanidine and dantroline, are commonly used but can cause cognitive impairment or liver damage. Surgical tendon lengthening should be considered if joint contractures are present. Lastly, baclofen can be delivered via a subcutaneous pump through a catheter to the intrathecal space.<sup>15</sup> While this intervention requires surgery and has other risks, it can be a remarkably effective intervention for adults with CP that have functional limitations due to significant lower extremity spasticity. Patients that may benefit from this intervention should be referred to a neurologic or neurosurgical team experienced in the placement and management of baclofen pumps.

**Seizure Disorder** – Overall, 30-40% of youth with CP have seizure disorders. Those with more severe motor or cognitive impairment have a higher prevalence of seizure disorders. Persons with hemiplegic or quadriplegic CP are unlikely to experience a remission of their seizures in adulthood, however if they have been seizure-free for several years, a trial of medication tapering can be attempted. Seizures common in CP include generalized seizures such as atonic, myoclonic and tonic-clonic seizures as well as focal seizures, with and without secondary generalization, and Lennox-Gastaut Syndrome.<sup>16</sup> The newer anti-epileptic medications are not approved for children. Therefore, many transition to adulthood on phenobarbital, valproate or carbamazepam. Adult epileptologists often change youth on these medications to newer anti-epileptic medications such as levetiracetam (Keppra®), topiramate (Topamax®), oxcarbazepine (Trileptal®), or lamotrigine (Lamictal®). All of these medications can cause cognitive impairment, dulling, irritability, and restlessness. These side effects may be difficult to identify in a person with severe physical or cognitive impairment. The provider should be careful to perform a good history and mental status

examination with a knowledgeable caretaker. Some cases that are refractory to more than one anti-epileptic medication may require either surgery or vagal nerve stimulation.<sup>17</sup> Fortunately, some seizure types get better and less frequent with age.

**Pain Management** – Pain is common in youth and adults with CP, however it often goes unrecognized by health care providers because individuals with cerebral palsy may not be able to describe the extent or location of their pain. Turk et. al (1997) found that 84% of women with CP reported pain, and more than 50% experienced pain daily.<sup>18</sup> One-third reported that the pain was constant.<sup>18</sup> Pain in persons with CP is most commonly attributable to musculoskeletal deformities, overuse syndromes, and arthritis. The most common sites for pain are: hip, due to overuse or dislocation; foot, after arthrodesis or due to muscle spasm; knee, due to chondromalacia patella, patella alta, or muscle spasm; and back or spine, due to degenerative changes or pathologic fractures from osteoporosis.<sup>19,20</sup> Most adults with cerebral palsy experience what is called *post-impairment syndrome*, a combination of pain, fatigue, and weakness due to muscle abnormalities, bone deformities, overuse syndromes (sometimes also called repetitive motion injuries), and arthritis. Individuals with cerebral palsy also have limited strength and restricted patterns of movement, which puts them at risk for overuse syndromes and nerve entrapments. There are almost no formal studies in pain management of persons with CP. Pain management should be multi-modal using physical therapy with stretching and positioning, whirlpool, ultrasound, medications such as non-steroidal anti-inflammatory drugs (NSAIDs) and correction of the underlying problem, such as vertebral fractures or joint dislocations.<sup>21</sup>

## Conclusion

Children and youth with Cerebral Palsy are almost always living longer into adulthood. Health care transition services for persons with CP must provide supports to help them access adult specialty services and overcome insurance barriers, while managing primary health issues associated with cerebral

palsy in the young adult such as spasticity. Moreover, providers caring for young adults with CP must carefully screen for and treat common co-morbidities of CP including joint and muscle pain syndromes, constipation, GERD and behavior issues. Primary care and specialty services for adults with CP are not widely available due to issues related to provider training, insurance, and family and patient related barriers. Providers involved in health care transition for youth and young adults with Cerebral Palsy must be active advocates for them in the adult health care system.

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## A Name Change After 55 Years!

To better convey the services provided to the community, Cerebral Palsy of Northeast Florida has changed its name to **"NEW HEIGHTS"**. Because programs are provided to infants, children and adults with disabilities, the new tagline – *"turning disabilities into capabilities"*, captures the services offered and provides a snapshot of what the agency is all about.

Located in a 12,000 sq ft building on the Southside, Cerebral Palsy of Northeast Florida Inc., was established over 55 years ago. Programs are provided for infants, children and adults with all types of disabilities, not only those affected by Cerebral Palsy.

The new name more accurately reflects the population New Heights serves. Many clients have disabilities caused by brain injury, down syndrome, mental retardation, spina bifida and other conditions. New Heights assists them as they progress along a journey of discovery and work toward their goal of becoming independent whether or not they live on their own, in a group home or with their family.

Therapy covering PT, OT and Speech are offered in addition to an in-house sensory room. Programs teach adults the skills needed for future independence; programs assist with the search for employment by providing job screening, resume preparation and job shadowing; and those seeking independence can enroll in the supported living program. New Heights also provides respite care and a fully inclusive day care.

New Heights programs are designed to help each individual strive to be as independent as possible within the limitations of their physical and/or intellectual challenges.

**For more information call 396-1462 and dial Ext 123 for M. Tucker regarding Adult Programs, or N. Naylor on Ext. 160 for Therapy Services.**

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