The Care of Adults with Pediatric Onset Disabilities

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Cerebral Palsy: Transitions and Adult Care
Objectives

• Discuss Cerebral Palsy and develop a better understanding of the issues encountered by individuals with Cerebral Palsy.
• Discuss the care of these individuals when they become adults.
• Discuss the process of transitioning a child to adult care medicine.
Cerebral Palsy (CP)

- Cerebral Palsy is a non-progressive neurological disorder which occurs before, during or after birth in the first few years of development.
- Cerebral Palsy is the most common major disabling motor disorder of children.
- Cerebral Palsy affects movement and posture, causing activity limitations.
Epidemiology

- In the US there are 3.6 cases per 1,000 among 8-year-old children\(^1\)
- Data from the 2004 National Health Interview Survey indicate that \(\sim 1.4\) million children with some kind of activity limitation were in the 14-17 year old age group, a critical period for transition planning\(^2\)
- Over 90\% of individuals with CP live beyond their 18\(^{th}\) birthday\(^5\)
Comorbidities

- The degrees of motor function vary greatly among children and adolescents with CP
- Children with CP, in addition to physical impairments, may also have associated impairments:\n  - learning disabilities
  - communication difficulties
  - perception and sensation disturbances
  - epilepsy
Gross Motor Function Classification Scale (GMFCS)

**Level I** move freely in almost all situations.

**Level II** can walk, run and climb in most settings but may need held mobility equipment for safety and wheeled mobility for travelling long distances.

**Level III** can walk using assistance and can self-propel manual or powered wheelchairs.

**Level IV** can walk short distances with assistance but use wheeled mobility in most settings and need physical assistance for transfer.

**Level V** require extensive support in all situations.\(^7\)
Types of Cerebral Palsy

- **Spastic**: most common type of CP; reflexes are exaggerated, weakness and discoordination, as well as muscle movement is stiff.

- **Dyskinetic**:
  - Athetotic: Involuntary, slow, writhing movements
  - Dystonic: Trunk movements are more affected than limb muscles, resulting in a twisted position

- **Ataxic**: Voluntary muscle movements are not well coordinated

- **Hypotonic**: Muscle tone is decreased or floppy

- **Mixed**: Combinations of the symptoms listed above
## Distribution of Limbs Affected

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Quadriplegia</td>
<td>Both arms and legs are affected. The muscles of the trunk, face and mouth are often also affected</td>
</tr>
<tr>
<td>Diplegia</td>
<td>Both legs are affected.</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>One side of the body (one arm and one leg) are affected</td>
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![Diagram showing types of cerebral palsy]
Transitions From Pediatric Care to Adult Care

• Transitions involve a significant change or set of changes in a person’s life circumstances during a relatively limited time period rather than over the life span.²

• People with CP often achieve fewer developmental milestones, face more restrictions in social participation compared to healthy peers, and may lag behind in education and work, independent living, and relationships.³
# Transitioning

## Critical Steps to Successful Transition from Child-Oriented to Adult-Oriented Health Care for Youth with Special Health Care Needs.

1. Ensure that an identified clinician assumes responsibility for health care and planning for youth with special health care needs during the transition period so as to provide uninterrupted, comprehensive care.
2. Incorporate into physician training and certification the core knowledge and skills to provide developmentally appropriate transition services to young with special healthcare needs.
3. Maintain a portable, accessible, up-to-date medical summary to facilitate collaboration during the transition.
4. Write a transition plan by age 14 years with the patient and family that outlines services, providers, and payment. Review and update the plan annually and at the time of transfer.
5. Adhere to standard guidelines for primary and preventive care, recognizing that youth with special healthcare needs may require more resources to optimize their health.
6. Ensure the continuation of affordable health insurance that provides appropriate compensation for planning and coordination for youth with special healthcare needs.
What to address in a Transitioning Process?

The top four barriers identified:

- Lack of access to healthcare
- Lack of professional knowledge
- Lack of information provided
- Uncertainty regarding the transition process

Issues to address during health care transition:

- Changes in health insurance
- Identification of an adult-oriented primary care provider
- Identification of an adult-oriented subspecialist
- Need for disability-based services
- Capability for self-management
- Skills in health care advocacy
- Health promotion and disease/injury prevention
Adult Providers

- Few adult medicine practitioners have been trained in the care of the patient with CP, and may therefore be uncomfortable providing care.¹
  - General practitioners, gynecologists and medical students have all been shown to have limited knowledge about CP, which promotes misunderstanding, negative attitudes and stereotyping of patients.¹⁴
- Few medical facilities are prepared to treat adults with developmental disabilities.¹
- Treating adults with complicated musculoskeletal and neuromuscular impairments is challenging.
Adults with Cerebral Palsy

- Adults with CP require ongoing health services to continue to monitor for needs that were present in childhood.
  - Feeding difficulties, gastroesophageal reflux, constipation, seizures, progressive scoliosis, contractures and joint dislocations secondary to unremitting spasticity continue to be important medical issues to address.\(^5\)

- Adults with the CP often develop musculoskeletal and neurological symptoms, such as severe pain, chronic fatigue, and a premature decline in mobility and function, as they age.\(^1\)
Pain in Adults with Cerebral Palsy

- Twenty-seven per cent of the adults with CP had chronic pain, compared with 15% in the general population.
  - However pain did not increase with age, which is different from the general population.
  - The most frequent site was back pain, both in adults with CP and in the general population.\(^\text{15}\)
- There is a high prevalence of neck and shoulder pain in persons with dyskinesia.\(^\text{15}\)
- Overuse syndromes, chronic pain and fatigue, osteoarthritis and osteoporosis, and an overall decline in mobility have all been reported among adults with CP.\(^\text{5}\)
Musculoskeletal Issues in Adults with Cerebral Palsy

- Studies of adults with CP have reported pain in 67 to 82%. Back, hip and lower extremities (knee) were the most common pain locations in many studies.¹

- Numerous musculoskeletal impairments, including patella alta, hip displacement, spondylolysis, cervical stenosis, scoliosis, foot deformities, and disuse osteoporosis.¹
  - Patella alta, most common in individuals with CP of the spastic diplegic type, can cause anterior knee pain and is complicated by stress fractures, tendonitis/bursitis, and subluxation/dislocations.
  - Hip subluxation/dislocation occurs in approximately 1% of individuals with spastic hemiplegia, 5% of those with diplegia, and up to 59% of those with quadriplegia.¹
  - Spondylolysis in weight-bearing adults with CP is reported to be up to 21 to 30%.
  - Cervical stenosis, most likely an overuse injury, has been found to be more common in individuals with CP than in the general population.¹
Osteoarthritis in Adults with Cerebral Palsy

Osteoarthritis was reported as a cause of pain in several studies; one study found clinical evidence of arthritis in 27% of young adults participates with CP compared with only 4% of the general population.¹

- Early osteoarthritis is a significant issue in many adults with CP.¹
- Joint destruction is believed to occur disproportionately in persons with CP, particularly of the hip¹

The epidemiology of osteoporosis and fractures in children and adults with CP is not well characterized.¹
Psychological Issues in Adults with Cerebral Palsy

It has also been shown that adults with CP tend to present with higher levels of depressive and stress symptoms when compared with the general population.\textsuperscript{15}

Physical pain was noted to occupy the thoughts of individuals with CP and took a lot on their energy level, which reduced their opportunities to participate in leisure activities, and this resulted in sadness and feeling of hopelessness.\textsuperscript{7}

Psychosocial issues associated with ageing with CP include the need for social support, self-acceptance and acceptance by others.\textsuperscript{12}
Fatigue in Adults with Cerebral Palsy

- Adults with CP reported significantly more physical but not mental fatigue than the general population. In a Norwegian study, 30% of adults with CP versus 22% of the reference population reported substantial fatigue.\(^{15}\)
- In a study of individuals with CP, an overwhelming amount of the complaints focused on fatigue and emphasized that fatigue occurred with walking and prolonged activity. Self-awareness of the individuals’ own bodies and adapting activity to continue to participate in various aspects of their lives emerged as the most important theme.\(^{8}\)
- Fatigue was most prevalent in persons at Gross Motor Function Classification System levels II and III.\(^{15}\)
Medical Care in Adults with Cerebral Palsy

- Research suggests that adults with CP use preventative care services less than do persons without disabilities.
  - Adults with CP use specialty health-care and rehabilitation services less, and emergency room cares more, than their non-CP peers.
  - Less than half of women with CP have Pap smears, and there have been no studies addressing the impact of menopause on bone health in adult women with CP.¹
- In Ontario’s healthcare, patterns have shown a greater rate of hospital admissions among people with CP compared to age-matched peers.⁵
Functional Outcomes in Adults with Cerebral Palsy

• CP has frequently caused a cycle of deconditioning, in which physical function deteriorates, followed by a further decrease in physical activity, and a cascade of functional decline.¹
  • About a third of adults with CP reported modest-to-significant decreases in walking or self-care tasks, especially in dressing and walking.¹

• The reasons for these early changes have similarly not been fully worked out; it is known, however, that the natural history of mobility in CP is one of early decline.¹

• Children with CP are less active than their non-disabled peers and have a lower maximum oxygen consumption, muscular endurance, and peak anaerobic power.¹
The Workplace

• It has been shown that individuals with physical impairments tend to be more loyal employees.

• Workplace accommodations may be needed to enhance continued employment for adults aging with CP.\textsuperscript{12}
Conclusions

• The diagnosis of CP is often thought of as a pediatric condition; it must be understood as a life-long disorder.

• Gaps in transition to the adult health system result in poor health outcomes and diminished opportunities to participate as productive members of the community.\textsuperscript{11}

• The goal of the Transitions process: Fostering autonomy and preparing adolescents for autonomous medical decision making. To maximize “lifelong functioning and potential through the provision of high-quality, developmentally appropriate (and technically sophisticated) health care services that continue uninterrupted as the individual moves from adolescence to adulthood.”\textsuperscript{2a}
More Conclusions

- Exercise and activity are effective means of pain management.¹
- Ensuring that persons with CP remain as active as possible is of critical importance.¹
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