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Special Report

Chronic Neurological Disorders Throughout the Lifespan

Evidence-based guidelines direct the long-term care treatment team

By Kathleen K. Frampton, RN, MPH

Each segment of the population diagnosed with chronic neurological conditions presents both unique challenges and opportunities to health professionals in the long-term care field. Starting with children, we know that the need for pediatric long-term care services appears to be on the rise. This is due primarily to an increase in the incidence of cerebral palsy over the last decade and the higher survival rate of these critically premature and frail infants with improved intensive care. When young and middle-aged adults with chronic adult-onset progressive neurological conditions require institutional care, they are most often admitted to traditional nursing homes. This necessitates their integration into a medical environment and social milieu that are tailored to the frail, aged resident. The older cohort poses some of the most complex challenges in the accurate recognition, diagnosis, and treatment of movement disorders coexisting with significant medical comorbidities. These trends result in patient care needs which require medical leadership, expert neurology consultation, and clinical team collaboration.

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—Audrius Plioplys, MD, FRCPC, FAAP, CMD

Cerebral Palsy in Pediatric Long-term Care

Children with cerebral palsy (CP) account for the largest percentage of pediatric residents with chronic neurological conditions in long-term care. CP is actually an umbrella-like term used to define a non-progressive disorder of posture or movement due to a lesion of the developing brain. Many etiologies—including perinatal asphyxia, low birth weight, and multiple births—result in the multitude of clinical symptoms that characterize CP.

There are five categories of CP: spastic, athetoid, dystonic, ataxic, and mixed. Spastic CP is the most common type, accounting for nearly 80% of cases, and is characterized by involuntary spontaneous movement and muscle contraction, hypertonia, and rigidity. Cerebral palsy is also described in terms of the location of the motor impairment and may be quadriplegic, diplegic, or hemiplegic. This disease is frequently associated with other deficits such as mental retardation in 60% of the cases, epilepsy in 50%, and hearing/visual defects in 10% to 15%. According to Audrius Plioplys, MD, FRCPC, FAAP, CMD, medical director at Little Angels in Elgin, Ill., and Marklund Children's Home in Bloomingdale, Ill., and former chair of AMDA's Pediatric Section, "the vast majority of pediatric residents in LTC are of the spastic

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The long-term care rehabilitation team will frequently use standing frames and other positioning devices when providing physical therapy to children with cerebral palsy.

type with quadriplegia and are neurologically devastated." Board-certified in both pediatric and adult neurology, he noted that "the biggest challenge for medical directors, attending physicians, and neurologists is in managing spasticity and epilepsy in this population."

Spasticity

Lesions of the motor areas in the brain in association with injury or deformity of the descending motor pathways (corticospinal tracts) result in spasticity in the child with CP. Spasticity is characterized by increased tone, hyperreflexia, clonus, and resistance to stretching that contribute to contractures, musculoskeletal deformities, and pain due to reduced longitudinal muscle growth. Scoliosis and hip dislocation are frequent problems for nonambulatory children with spastic CP and quadriplegia. Immobility and disuse also lead to osteoporosis and contractures, which can cause bone fractures. It is not uncommon for these problems to require fracture reduction or orthopedic surgeries such as tendon lengthening procedures or femural osteotomy.

The most recent "Treatment of Cerebral Palsy–Research Status Report" (2003) of the United Cerebral Palsy Research and Educational Foundation supports those treatments that diminish the degree of impairment and increase independent mobility. However, Dr. Plioplys stressed, "the overarching goal of care for the team is to avoid surgery because of the critical risks posed by pneumonia and infections...in this regard, the efforts of the nursing staff, along with the physical and occupational therapists, are so very important." Therapeutic rehabilitation interventions typically include range-of-motion exercises, participation in activities of daily living, strengthening and functional mobility training, and electrical stimulation. Splints, orthotic devices, and serial casting, as well as adaptive equipment, standing frames, and other positioning devices, are frequently employed and require intensive involvement of the entire rehabilitation team.

Drugs that are frequently used to control spasticity as a means to avoid surgery and provide relief for patients during range-of-motion exercises are benzodiazepines (e.g., chlorazepate dipotassium, diazepam) and skeletal muscle relaxants (e.g., baclofen, dantrolene sodium). Often muscle relaxants are used in combination with the benzodiazepines in order to obtain the additional anti-seizure properties necessary for some CP patients. Baclofen can be taken orally or delivered directly into the intrathecal space of the spine through a system consisting of a catheter and pump. While intrathecal baclofen pump therapy may be indicated to treat spasticity in other neurological disorders, it is not widely used in pediatric long-term care for residents with CP and quadriplegia.

Botulinum toxin type A (Botox) is used clinically in the treatment of spasticity by directly injecting it into muscle groups in order to produce denervation of the muscle, causing a localized reduction in muscle activity. It has been used very effectively to treat deformities and biomechanical abnormalities (e.g., ankle, hands) due to sustained muscle contraction around the joints. Botox is also used in the treatment of excessive drooling in cerebral palsy for non-cosmetic reasons in cases where aspiration of saliva is problematic. Typically, the botulinum toxin is injected into each parotid gland, the gland that produces most of the thin, watery part of the saliva.

Spasticity is difficult to treat, but it is equally difficult to evaluate treatment efficacy. Dr. Plioplys stressed, "This is an area of clinical research that is so very much needed in pediatric long-term care because there are no scientific evaluation tools or rating scales with objective criteria to measure progress. Since members of the treatment team often have divergent opinions and assessments of spasticity in each resident...it is critically important to stimulate interest in tackling this need."

Epilepsy

Children with medically resistant epilepsy have special needs since brain injuries resulting from seizures may be associated with cognitive impairment and emotional difficulties. Risk factors for developing persistent seizures include atonic, tonic, and atypical seizures, cryptogenic or symptomatic epilepsy, mental retardation, neurological deficits, and structural brain damage. Consequently, controlling seizures is another major challenge in pediatric long-term care.

The modern goal of epilepsy treatment has moved beyond seizure control to the elimination of seizures and side effects. Today there is a very large armamentarium of both old and new anti-epilepsy drugs (AEDs). In fact, since 1993, eight new AEDs have been introduced, including long-acting preparations. Vagus nerve stimulation (VNS) received FDA approval for treatment of refractory partial onset seizures in adults and adolescents in 1997. However, neither VNS nor epilepsy surgery (anterior temporal lobe resection) is generally considered for the severely compromised pediatric LTC residents.

The ketogenic diet has been used since the 1920s and has gained popularity in recent years. This is a very restrictive, high fat (80%), low carbohydrate, adequate protein (20%) diet that results in ketosis, mimicking starvation. While the exact mechanism of action is not known, there has been much success with the diet, particularly in the institutional setting with nonambulatory patients who are tube-fed and have no access to forbidden foods. A strong proponent of this diet, Dr. Plioplys finds it to be very effective and in his experience was able to decrease anticonvulsants after implementing the regimen.