Sleep-Robbing Pain Common in Up to Half of Disabled Children

BY SHARON WORCESTER
Southeast Bureau

BOSTON — Pain frequently interferes with sleep in children and adolescents with physical disabilities, results of a study suggest.

Pain should be routinely assessed and managed to minimize the adverse effects it can have on patients and their caregivers, said Marilyn Wright of McMaster University and McMaster Children's Hospital, Hamilton, Ont.

The cross-sectional survey of 178 caregivers of children and adolescents with physical disabilities showed that up to half of those with certain disabilities experienced nighttime pain, and that this pain—and its effects on sleep—was associated with a number of sequelae in both patients and caregivers, she reported in a poster at the annual meeting of the American Academy for Cerebral Palsy and Developmental Medicine.

For example, 50% of the children and adolescents in the study with muscular dystrophy, 48% of those with cerebral palsy, 15% of those with developmental delay, and 15% of those with spina bifida experienced nighttime pain. The most common types of pain reported were muscle pain (18% of patients), joint pain (10% of patients), muscle spasms (9% of patients), and growing pains (7% of patients).

Among the factors significantly associated with pain in the children were waking during the night (9% reportedly wake up three or more times per night), the necessity of position change during the night, daytime irritability attributed to lack of sleep, and interference with school and social activities.

Caregivers also were affected. Nighttime pain experienced by a child was shown to be associated with caregiver daytime irritability and exhaustion attributed to lack of sleep; negative effects on caregivers' work outside the home; caregiver concern about the child's sleeping position; and concern about the negative effect of poor sleep on the general health of the child.

Furthermore, the children and adolescents who were reported as having nighttime pain that interfered with sleep were more compromised in their independent mobility, Ms. Wright reported.

Self-reports by 25 children and adolescents who were interviewed about nighttime pain, its effects on sleep, and sequelae of poor sleep showed trends similar to those seen with the data provided by caregivers, she noted.

The current study was undertaken following a prior finding that pain had a negative effect on sleep quality in children and adolescents with physical disabilities more often than in children without such disabilities (30% vs. 3%). With increasing attention to the effects of pain on sleep in this population, there has been greater awareness of the need for early intervention.

"Early and ongoing orthopedic surveillance is an integral part of preventing secondary impairments that can contribute to pain and interruption of sleep," Ms. Wright concluded.

Attention to positioning throughout the day also can help reduce pain. At McMaster Children's Hospital, a "24-hour positioning program" is offered to parents and caregivers through workshops and home visits, as well as through educational materials. It addresses issues like pain, muscle integrity, muscle contracture, and mobility, she said in an interview.

The program focuses on positioning throughout the day and promotes supportive, safe, well-aligned, and comfortable sleep positioning in an effort to reduce pain and promote optimal sleep throughout the night, she said. ■

Hyperbaric Oxygen Aids Cognition in Cerebral Palsy

BY DAMIAN McNAMARA
Miami Bureau

FORT LAUDERDALE, FLA. — Parents reported a nearly 50% overall improvement in their children with severe cerebral palsy following hyperbaric oxygen therapy, according to a study reported at a symposium on hyperbaric therapy.

To assess parental perception of hyperbaric oxygen therapy (HBOT), researchers at Nova Southeastern University in Fort Lauderdale, Fla., surveyed parents or caregivers of 73 children with cerebral palsy (CP). They rated levels of improvements for cognitive, motor, sensory, and overall function on a 0-10 scale via telephone. The ages of children ranged from 1 year to 26 years, and 51% were male.

"Preliminary analysis of the data thus far suggests hyperbaric oxygen can be an effective treatment for CP," Brandon Korman said at the symposium, sponsored by the Ocean Hyperbaric Neurologic Center in Fort Lauderdale.

"Parents reported quite a bit of improvement," said Mr. Korman, a graduate student at Nova Southeastern University. Parents found a 49.6% overall improvement in their children's conditions after HBOT.

Of the three domains on the survey, parents reported the greatest improvements in cognitive vs. motor or sensory areas. "This was a particularly interesting finding because mainstream medicine does not offer much for cognitive improvement in cerebral palsy," Mr. Korman said.

He conducted the telephone survey with graduate student Maritza Figueroa.

"When interpreting these data, keep in mind this is based on parent perception rather than a control group," Mr. Korman said. "There is no control group to rule out spontaneous improvement. Children at the program also received other therapies concurrently."

There was no indication of an age-related recovery effect. "This was somewhat surprising," Mr. Korman said.

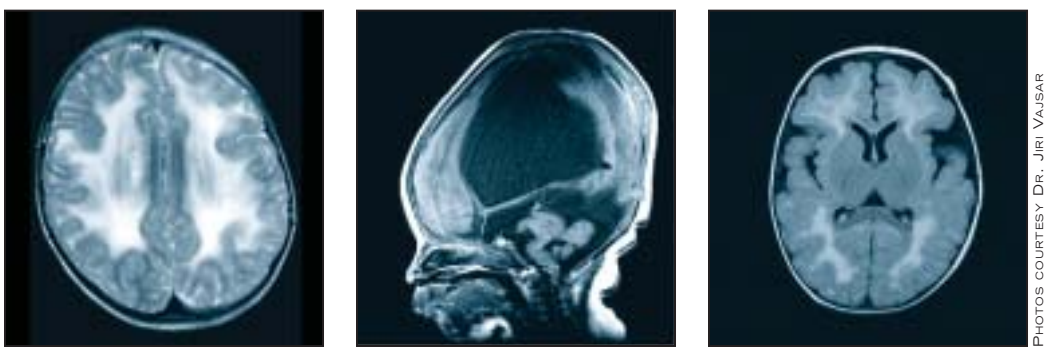
"It could be that HBOT is increasing the period of plasticity that otherwise drops off after infancy." Another finding that warrants further investigation, he said, was a lack of a significant dose-response relationship between the number of treatments and overall improvement.

A total of 73% of parents survey reported overall satisfaction with HBOT. ■

Brain MRI Aids Diagnosis of Congenital Muscular Dystrophies

BY AMY ROTHMAN SCHONFELD
Contributing Writer

MONTREAL — Characteristic changes on brain MRI can help diagnose and differentiate congenital muscular dystrophies with brain and eye abnormalities, reported Dr. Jiri Vajsar at the 10th International Child Neurology Congress.



PHOTOS COURTESY DR. JIRI VAJSAR

T2 image shows abnormal high signal in white matter (left) in CMD. Smooth cortex lacks sulcation; abnormalities of the corpus callosum and cerebellum and enlarged ventricles are consistent with WWS (middle). Thick cortex is indicative of MEB migrational abnormality (right).

Congenital muscular dystrophies (CMDs) with brain involvement share several common features, explained Dr. Vajsar, a neurologist affiliated with the Hospital for Sick Children in Toronto. All are autosomal recessive diseases that are characterized by early-onset hypotonia and weakness, delayed motor development, and elevated creatine kinase levels. Some CMD types also manifest mental retardation, delayed global development with subsequent regression, progressive contractures, seizures, variable ophthalmic findings, and cardiac and respiratory involvement.

Immunohistochemically, CMDs with brain involvement can be grouped into those with a deficiency in laminin-α2 chain (also known as merosin) or in α-dystroglycan, said Dr. Vajsar.

Both laminin-α2 chain and α-dystroglycan are part of a large complex of proteins and glycoproteins that protect the integrity of muscle cell structure during repeated cycles of contraction and relaxation. Poorly glycosylated α-dystroglycan corrupts laminin binding, weakening the link of muscle fibers to the extracellular matrix, and laminin-α2 chain deficiency results in disruption of the basal lamina of striated muscle.

MRIs of children with laminin-α2 chain deficiency show easily identifiable abnormalities in myelinated areas, although the corpus callosum and optic radiation remain normal. Despite the white matter abnormalities, these children maintain good cognitive function; however, about 30% are prone to seizures.

It is more difficult to generalize about the appearance on MRI of α-dystroglycan deficient CMDs, because several CMD phenotypes exist, said Dr. Vajsar. As a general rule, MRIs of these disorders show abnormalities in the posterior fossa, such as flattening of the pons, cerebellar hypoplasia or dysplasia, cerebellar cysts, and hypoplastic or absent vermis. The cortex takes on a cobblestone appearance, with disorganized cortical layers due to abnormal neuronal migration; multiple, abnormal, and coarse gyri with agyric regions; and variable thickness.

In Walker-Warburg syndrome (WWS), MRI findings include type II lissencephaly (cortical smoothing) and cerebellar malformation. Ventriculomegaly and abnormalities of corpus callosum and splenium are also common. Anterior (e.g., cataracts, microcornea, microphthalmia, lens defects) or posterior (e.g., retinal detachment, optic nerve atrophy, glaucoma) eye abnormalities are also frequent. Clinically, children with WWS are profoundly retarded, have seizures, and usually succumb to death within the first 3 years of life.

MRI findings in children with other CMD types show a spectrum of generally milder gray and white matter abnormalities, said Dr. Vajsar.

In muscle-eye-brain (MEB) disease, cortical, cerebellar, and callosum/splenium abnormalities are less prominent than in WWS. Polymicrogyria and thickened cortex may be noted in the frontal and parietal cortices, while agyria, cortical thinning, and lissencephaly may be evident in the occipital cortex.

MRI in patients with MDC type 1C may show normal brain with or without cerebellar cysts, said Dr. Vajsar. Occasionally, MRI shows other white and gray matter abnormalities, from cortical migrational anomalies and white matter changes to MEB-type or WWS patterns of abnormalities. ■