COMMENT. The syndrome of mitochondrial neurogastrointestinal encephalomyopathy is an autosomal recessive disorder characterized clinically by severe gastrointestinal dysmotility, cachexia, ptosis, ophthalmoparesis, peripheral neuropathy, leukoencephalopathy, and mitochondrial abnormalities. The disease is caused by mutations in the thymidine phosphorylase gene.

## SEIZURE DISORDERS

### NEURONAL DYSFUNCTION AND TEMPORAL LOBE EPILEPSY

The relation of the neuronal dysfunction in the temporal lobes of children with temporal lobe epilepsy (TLE) to intractable seizures was investigated at the Montreal Neurological Institute and Hospital, and Montreal Children's Hospital, Canada. Reduction in N-acetylaspartate/creatine (NAA/Cr) ratios in the temporal lobes, as measured by proton magnetic resonance spectroscopic imaging, was used as an indicator of neuronal dysfunction. No significant differences were observed in the NAA/Cr ratios in 5 consecutive children with newly diagnosed TLE compared to 5 with long-standing intractable TLE. The patients had bilateral or unilateral reductions in NAA/Cr ratios of equal severity and extent in both groups. The results imply that the temporal lobe neuronal dysfunction in children with TLE is not specifically related to the intractable seizures and is present before seizures begin. (Miller SP, Li LM, Cendes F et al. Neuronal dysfunction in children with newly diagnosed temporal lobe epilepsy. Pediatr Neurol April 2000;22:281-286). (Respond: Dr Douglas L Arnold, Montreal Neurological Institute and Hospital, 3801 University Street, Montreal, Quebec H3A 2B4, Canada).

COMMENT. N-acetylaspartate/creatine ratio is a marker of the epileptogenic process and neuronal dysfunction in temporal lobe epilepsy, and reductions in the ratio are not specifically related to effects of refractory seizures.

### EPILEPTIC SYNDROMES, COGNITION AND CLASS PLACEMENT

A retrospective statistical analysis of IQ and school placement in 251 children with epilepsy, aged 3 to 17 years, is reported from the Hopital-Saint-Vincent-de-Paul, Paris, France. Age at onset and seizure frequency are associated with poor outcome. Children with idiopathic generalized or localization-related epilepsy have higher IQ scores and a greater likelihood of normal school placement than those with symptomatic or cryptogenic generalized epilepsies. IQ and schooling are related to the epileptic syndrome, age at onset and duration of epilepsy, and number of antiepileptic drugs. Cognitive deficits vary with the localization of an epileptic focus. (Bulteau C, Jambaque I, Viguier D et al. Epileptic syndromes, cognitive assessment and school placememnt: a study of 251 children. Dev Med Child Neurol May 2000;42:319-327). (Respond: Dr C Bulteau, Hopital Saint Vincent de Paul, Service de Neuropediatrie, 82 Avenue Denfert-Rochereau, 75674 Paris Cedex, France).

COMMENT. Epileptic syndromes vary in their association with impairments in IQ and school performance. IQ scores and class placement are least affected in children with idiopathic, generalized or localization-related epileptic syndromes.

**Autistic regression with Landau-Kleffner syndrome** is reviewed by Mantovani JF, Washington University School of Medicine, St Louis, MO (Dev Med Child Neurol May 2000;42:349-353). A cause and effect relationship between autism and EEG abnormalities is not established, and therapies for LKS, including

antiepileptic medications, steroids, and subpial resection, have no proven beneficial effects on the autistic regression. Since LKS may improve spontaneously, any claims for therapeutic efficacy must be subject to controls.

## ATTENTION DEFICIT DISORDERS

### SLEEP DISTURBANCES AND ATTENTION DEFICIT DISORDERS

The prevalence of parent-reported and self-reported sleep disturbances in 46 unmedicated school-aged children (mean age, 7 +/- 1 1/2 years, 74% male) with attention deficit/hyperactivity disorder (ADHD) was determined at Brown University School of Medicine, Providence, RI. The patients had been screened for marked symptoms of sleep-disordered breathing, and those diagnosed with obstructive sleep apnea were excluded. Results in the ADHD group of children were compared to those in 46 normal controls, matched for age and sex. A Children's Sleep Habits Questionnaire (CSHQ) completed by the parents includes subscales referring to bedtime resistance, sleep-onset delay, sleep duration, sleep anxiety, night wakings, parasomnias (talking in sleep, restlessness, sleepwalking, teeth grinding, nightmares, bed-wetting), sleep-disordered breathing, and daytime sleepiness. A corresponding Sleep Self-report (SSR) assessed similar sleep disturbances from the child's perspective.

Children with ADHD had significantly higher scores (and more sleep disturbances) on all, except for sleep-disordered breathing, subscales of the parent CSHQ compared to controls. The average number of hours of sleep reported by parents on the CSHQ was significantly lower in the children with ADHD than in controls. The child's SSR also showed a greater incidence of sleep disturbance, particularly relating to bedtime struggles and sleep resistance (P, .05-.001). Parent and child sleep reports showed a higher correlation in ADHD children than controls. Children with ADHD and comorbid sleep disorders should receive specific behavioral and pharmacological therapy. (Owens J A, Maxim R, Nobile C, McGuinn M, Msall M. Parental and self-report of sleep in children with attention-deficit/hyperactivity disorder. Arch Pediatr Adolesc Med June 2000;154:549-555). (Respond: Judith A Owens MD MPH, Division of Pediatric Ambulatory Medicine, Rhode Island Hospital, 593 Eddy Street, Providence, RI 02903).

COMMENT. The causes of sleep problems in children with ADHD have previously been addressed (Trommer BL et al. Ann Neurol 1988;24:322), and these include stimulant medications, comorbid anxiety disorder, and environmental behavioral sleep deprivation. A specific ADHD-linked dysregulation in sleep and arousal mechanisms has also been proposed (Corkum P et al. J Am Acad Child Adolesc Psychiatry 1998;37:6). Objective measurements of sleep habits, such as polysomnography (Greenhill L et al. Sleep 1983;6:91-101) and actigraphy, have shown conflicting results, while parental ratings of sleep behavior have revealed an increased prevalence of sleep problems in ADHD children. The authors of the present report have also included observations based on the child's perspective, and these correlate with the parental findings. Sleep disturbances, especially those occurring at bedtime, are frequently reported by parents and the children with ADHD. Their impact on the therapy of ADHD is problematic and requires further investigation.

Children presenting with ADHD should be screened for sleep disorders that may have a causative role or exacerbate the symptoms of inattention and hyperactivity. The use of clonidine should be considered as a substitute for stimulant medication in patients whose sleep problems require a change in