LEARNING DISABILITIES IN NEUROFIBROMATOSIS TYPE 1

The frequency of specific leaning disabilities (SLD) in neurofibromatosis type 1 (NF1) was determined in a cohort of 81 patients (43 males, 38 females; mean age 11 years 6 months; age range 8-16) followed at Children's Hospital, Westmead, NSW, Australia. Academic underachievement occurred in 42 (52%) of NF1 children, 3.5-fold more frequently than in a control group. One or more SLDs was present in 16 (20%); 7 had reading disability, 7 spelling, and 8 a math disability. Of the 52% with academic difficulties, one third had a general learning disability (GLD) associated with IQ deficit. Only males with NF1 were at significant risk for SLD; Verbal IQ<Performance IQ discrepancy was predictive of SLD. NF1 patients with no SLD or GLD had normal IQ scores and average academic achievement but minor difficulties in sustained attention and visuospatial skills. ADHD was present in 31 (38%) NF1 children in the total group compared to 12% of 49 controls without NF1. The increase in frequency of ADHD was similar in those with comorbid SLD or GLD and lower but still significant in NF1 patients with no LD. (Hyman SL, Shores EA, North KN. Learning disabilities in children with neurofibromatosis type 1: subtypes, cognitive profile, and attention-deficit-hyperactivity disorder. **Dev Med Child Neurol** Dec 2006;48:973-977). (Respond: Kathryn N North MD, Clinical School, The Children's Hospital, Locked Bag 4001, Westmead, NSW 2145, Australia).

COMMENT. Children with NF1 have a 52% incidence of academic underachievement, and 20% have a SLD. One third of patients with LD have a GLD associated with a lowered IQ. Comorbid SLD and NF1 occur almost always in males, and in the above study, their verbal IQ was lower than performance IQ. The authors are examining VIQ<PIQ discrepancies in male preschool children as early predictors of risk of SLD. These findings are different from those of Eliason MJ (**Neurofibromatosis** 1988;1:17-25) who reported a preponderance of nonverbal learning problems (visual perceptual deficits) in children with NF1.

SEIZURE DISORDERS

DIAGNOSTIC ASSESSMENT OF STATUS EPILEPTICUS

Diagnostic methods of assessment of the child with status epilepticus (SE) are outlined following an evidence-based review of literature by the Quality Standards Subcommittee of the AAN and Practice Committee of the CNS. SE is a seizure of at least 30 min duration or 2 or more sequential seizures without full recovery of consciousness between seizures lasting 30 min. SE is classified by seizure type (focal, generalized or indeterminate) and etiology: 1) *acute symptomatic*, during an acute illness (26%); 2) *remote symptomatic*, a prior CNS insult with chronic encephalopathy or malformation and no acute illness (33%); 3) *progressive encephalopathy* (3%); 4) *febrile* illness, with no direct CNS infection (22%); and 5) *cryptogenic*, with no definable cause (15%). The etiologic incidence figures were obtained from 20 class III studies of 2,093 children. Laboratory studies were abnormal in 2.5%, CNS infection in 12.8%, AED levels were low in 32%. An inborn error of metabolism was

diagnosed in 4.2%. EEGs showed epileptiform abnormalities in 43%; 8% were generalized, 16% focal, and 19% both. MRIs were abnormal and indicative of the etiology of SE in 8%. Investigation of the cause of SE includes: 1) blood culture and lumbar puncture if there is clinical suspicion of a systemic or CNS infection; 2) AED blood levels: 3) toxicology and metabolic studies when clinically indicated or if etiology is unknown; 4) EEG to check for focal or generalized discharges, or if pseudostatus is suspected, or in diagnosis of nonconvulsive SE; and 5) MRI after SE is treated and stabilized, and if clinically indicated or if etiology is unknown. (Riviello JJ, Ashwal S, Hirtz D, et al. Practice parameter: Diagnostic assessment of the child with status epilepticus (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. **Neurology** November (1 of 2) 2006;67:1542-1550). (Reprints: American Academy of Neurology, 1080 Montreal Ave, St Paul, MN 55116).

COMMENT. The authors recommend further prospective studies to determine what factors may precipitate SE in children; the role for routine or selective laboratory investigations; indications and treatment significance of EEG; role of routine or selective MRI; and the frequency, etiology, and prognosis of nonconvulsive SE after control of convulsive SE.

Cognition and electrical SE during sleep (ESES). The duration of ESES and the localization of interictal foci play a major role in the degree and type of cognitive dysfunction following continuous spike-wave activity. ESES interferes with slow-wave activity at the site of the epileptic focus, impairing learning and other cognitive functions. (Tassinari CA, Rubboli G. **Epilepsia** Nov 2006;47(Suppl 2):40-43).

SHORT-DURATION ACTH THERAPY FOR WEST SYNDROME

Short-term developmental and seizure outcomes were assessed in 7 children, aged <12 months, with cryptogenic West syndrome treated with a short-duration (7-12 days) and low-dose synthetic ACTH at Nagoya City Hospital, Japan. All patients had received a trial of one or two AEDs before ACTH. Daily single dose of ACTH was 0.022-0.027 mg/kg/day (mean, 0.96 IU/kg), and total dose was 0.17-0.28 mg/kg (mean, 9.0 IU/kg). Interval between onset of spasms and initiation of ACTH was 12-105 days (median 14 days). Tonic spasms were controlled in all patients with no serious side effects. Spasms disappeared within 1 week, and hypsarrhythmia resolved within 2 weeks. EEG abnormalities persisted in 2. IQ or developmental quotients of 6 patients were 79-110 at age 2 to 6 years. One patient with a long treatment lag had a developmental quotient of 60. (Hattori A, Ando N, Hamaguchi K, et al. Short-duration ACTH therapy for cryptogenic West syndrome with better outcome. **Pediatr Neurol** Dec 2006;35:415-418). (Respond: Dr Hattori, Department of Pediatrics, Nagoya City Hospital, 1 Kawasumi, Mizuho-cho, Mizuho-ku, Nagoya 467-8601, Japan).

COMMENT. Vitamin B6 and AEDs, including clonazepam, valproic acid, and zonisamide, are used before initiating ACTH, because of possible adverse effects of hormone therapy. Delay in intiation of ACTH may result in poor outcome, and a favorable response to very low dose-short duration therapy should favor a recommendation for early trial of