

found in 44% of one sample of 108 patients (a DQ below 21 in 31%); 55% had a normal IQ. In studies of children with TS and near-normal IQ, 50% had ADD or hyperkinetic syndrome, and 25% had ODD. Infantile spasms and low IQ are significantly related. Cognition is also correlated with *tuber burden* (tuber number, size, and location). Genetic factors correlate with developmental outcome: *TSC1* cases have lower rates of mental retardation, seizures, and autism than *TSC2*. TS is linked to autistic spectrum disorder in 50 to 60% of cases (the rate is higher in those with infantile spasms), and these patients do not show the characteristic male preponderance seen in idiopathic autism cases. Temporal lobe tubers and EEG abnormalities increase the risk of autism.

Neurofibromatosis type 1 (NF-1) patients have an average mean IQ, despite earlier reports of an increased incidence of mental retardation, but learning disabilities, especially visual-perceptual deficits, occur in 30-60% of cases. ADHD is reported in 33%. Some studies show a resolution of childhood cognitive dysfunction in adults with NF-1. Cognitive deficits are correlated with the occurrence, number, and location of UBO's. Macrocephaly shows no correlation with cognitive functioning in NF-1. Adolescents with NF-1 and learning difficulties had social-skill problems, when compared to unaffected siblings. ADHD was the major risk factor for social problems. (Zaroff CM, Isaacs K. Neurocutaneous syndromes: behavioral features. *Epilepsy Behav* Sept 2005;7:133-142). (Respond: Charles M Zaroff MD, Comprehensive Epilepsy Center, New York University, 403 East 34th Street, New York, NY 10016).

COMMENT. Neurocutaneous syndromes are associated with an increased rate of mental retardation or learning disabilities. Problems in cognition and behavior are related to the underlying neurological disorders, especially seizures, and in tuberous sclerosis the genotype contributes to clinical heterogeneity. An increased prevalence of ADHD in all three syndromes is noteworthy, and tuberous sclerosis is strongly linked with autism.

SEIZURE DISORDERS

ETHOSUXIMIDE-INDUCED PSEUDOLYMPHOMA

A first case of pseudolymphoma induced by ethosuximide treatment in a 12-year-old boy is reported from the University of Sao Paulo, Brazil. He presented with a 2-month history of fever, weight loss, and non-painful lymphadenopathy in the neck, axillae, and inguinal regions. He had taken ethosuximide (30 mg/kg/day) for 3 months for absence epilepsy. The liver and spleen were not palpable. Blood count revealed a leucopenia of 3.6×10^3 /mcl (48% neutrophils, 3% eosinophils, 40% lymphocytes, and 9% monocytes), and decreased platelets of 119×10^3 /mcl. Cytomegalovirus, Epstein-Barr, herpes simplex virus, and toxoplasmosis were excluded. Biopsy of a cervical gland confirmed a diagnosis of lymphoma. After discontinuing ethosuximide, fever disappeared within 1 day, and lymph nodes decreased in size in 2 weeks and completely regressed in 2 months. Leukocyte and platelet counts normalized after 2 weeks of drug withdrawal. Fever and enlargement of lymph nodes recurred after 1 week following rechallenge with ethosuximide treatment. (Masruha MR, Marques CM, Vilanova LCP et al. Drug induced pseudolymphoma secondary to ethosuximide. *J Neurol Neurosurg Psychiatry* November 2005;76:1610). (Respond: Dr

MR Masruha, Division of Child Neurology, Department of Neurology and Neurosurgery, Botucatu Street 720, Federal University of Sao Paulo, Sao Paulo 04023-9000, Brazil).

COMMENT. This case is reported as the first example of pseudolymphoma associated with ethosuximide. Histologically, a lymphoid cell proliferation caused effacement of normal node architecture and a false appearance of malignancy. Pseudolymphoma may result from an hypersensitivity or idiosyncratic reaction, the drug acting as an antigen and triggering an immune reaction. Urticaria, Stevens-Johnson syndrome, systemic lupus erythematosus, eosinophilia, leukopenia, thrombocytopenia, aplastic anemia, and liver dysfunction have been reported with ethosuximide. Other anticonvulsants, phenytoin and lamotrigine, are reported to cause pseudolymphoma. Adverse effects of antiepileptic drugs, including idiosyncratic reactions are reviewed by Perucca E and Meador KJ (*Arch Neurol Scand* December 2005;112 (Suppl 181):30-35).

ETIOLOGY AND OUTCOME OF STATUS EPILEPTICUS

The etiology and outcome of status epilepticus (SE) in 135 children (76 boys, 59 girls), ages 1 month to 15 years, admitted to Nemazee Hospital, Shiraz, Iran, between 1999 and 2004, were studied retrospectively, and outcomes were rated according to the Glasgow Outcome Score (GOS). The mean \pm SD age was 4.14 \pm 3.8 years; 44 (33%) were younger than 12 months, 29 (21%) were 1-3 years old, and 62 (46%) were older than 3 years. Prolonged febrile seizure was the etiology in 69 (51%); 57% of FS patients were \leq 12 months old, 45% 1-3 years, and 50% $>$ 3 years. Idiopathic SE accounted for a total of 50 (37%) cases, and was associated with discontinuation of anticonvulsant drug in 38 (28%). Symptomatic SE was the cause in 16 (11.9%); 10 due to CNS infection, 4 metabolic disorders, and 2 trauma-related. Etiology did not differ significantly between age groups ($P=0.736$) or sex ($P=0.156$).

Fourteen (10.4%) died in hospital, and 81 (60%) were discharged well. Death was related to a prolonged febrile seizure in 4, idiopathic SE in 6, and symptomatic CNS infection in 3. Patients with symptomatic SE had the worst outcome, with 63% morbidity and mortality; the morbidity and mortality rate was 48% in idiopathic SE cases, and 29% for febrile seizure SE cases. Mean \pm SD duration of hospitalization was 7 \pm 9.7 days (range, 1-68 days); it did not differ with etiology but was shorter for patients with good outcome. (Asadi-Pooya AA, Poordast A. Etiologies and outcomes of status epilepticus in children. *Epilepsy Behav* November 2005;7:502-505). (Dr AA Asadi-Pooya, Shiraz University of Medical Sciences, Shiraz, Iran).

COMMENT. Fever is the most common cause of status epilepticus (SE) in children, even in those older than 3 years. The etiology of SE is significantly correlated with the outcome, symptomatic cases secondary to CNS infection having the worst, and prolonged febrile seizures the most favorable outcome. Factors in outcome of SE in adolescents and adults in the Netherlands were the underlying cause, noncompliance with AED treatment, systemic infection, and duration of the SE. (Scholtes FB et al. *Epilepsia* 1995;35:1104-1112). Noncompliance with AED therapy or inadequate instruction regarding the use of rectal diazepam administered in the home explained the need for admission of children with