2000;42:595-603). A previous report by these authors (Roulet E, Deonna T, Despland PA. **Epilepsia** 1989;30:564-568) refers to the association of oromotor dyspraxia in a child with benign childhood epilepsy with centrotemporal spikes (BECTS). A fluctuant course of partial seizures involving the face and right arm correlated with the intensity of EEG paroxysms located in the lower rolandic fissure. No structural lesion was detected on MRI (**Ped Neur Briefs** Nov 1989). A family of 9 children in three generations affected by autosomal dominant BECTS and associated with oral and speech dyspraxia and cognitive impairment is reported from Melbourne, Australia (Scheffer IE et al. **Ann Neurol** 1995;38:633-642; **Ped Neur Briefs** Nov 1995). Partial seizures may be repetitive and frequent, more than 100 times daily, as in 2 children with BECTS who showed cognitive decline that improved after seizures resolved (Ong H-T, Wyllie E. **Neurology** 2000;54:1182-1185; **Ped Neur Briefs** April 2000;14:28). In one patient the role of antiepileptic medication in the cognitive decline was suspect. A decision to treat BECTS despite invariable spontaneous remission is controversial.

MOVEMENT DISORDERS

NEURODEVELOPMENTAL OUTCOME OF OPSOCLONUS-ATAXIA

Cognitive, adaptive behavior, academic, speech and language, and motor abilities were assessed in 19 children with opsoclonus-ataxia and neuroblastoma, followed at University of Southern California, Los Angeles, CA. Patients were examined twice with a minimum interval of 1 year. Age at onset of symptoms ranged from 11 to 31 months (median: 18 months). Age at the time of 1st and 2nd evaluations was a median of 3.1 and 6.4 years, respectively. Cognitive scores improved between testing sessions for the group as a whole, and a larger average gain in IQ/DQ was seen with younger children. Motor abilities improved in gross and fine motor function, but not at an age-appropriate rate. Behavioral and emotional functioning was overall stable and did not worsen significantly with age. Speech and language testing showed persistent deficits in the majority of patients, and all were receiving therapy. The necessity for treatment (steroids or immune-modulating medication) during the interval between evaluations did not influence change in test scores, but a history of relapse was strongly associated with deficits in cognition, academic achievement, and visual motor integration. Of 5 children without relapse, 4 had test scores in the normal range. Despite initial severity of symptoms, a minority of patients had an excellent outcome, with cognitive, academic, behavioral, and motor function within the normal range, and all immunotherapy had been discontinued. Fourteen with multiple relapses, and a chronic course associated with developmental sequelae, required prolonged therapy. (Mitchell WG, Brumm VL, Azen CG et al. Longitudinal neurodevelopmental evaluation of children with opsoclonus-ataxia. **Pediatrics** October 2005;116:901-907). (Respond: Wendy G Mitchell MD, Neurology Division, Children's Hospital Los Angeles, 4650 Sunset Blvd, Box 82, Los Angeles, CA 90027).

COMMENT. Early and periodic neuropsychological testing is recommended in children diagnosed with opsoclonus-ataxia. Normal cognitive and behavioral development can be expected in the minority of patients without relapse, but a chronic and relapsing course is associated with developmental deficits. This study confirms earlier reports of a

high incidence of delayed development, motor incoordination, behavioral, and speech deficits in children with opsoclonus-myoclonus syndrome (Hammer MS, Larsen MB, Stack CV. **Pediatr Neurol** 1995;13:21-24; Papero PH et al. **Dev Med Child Neurol** 1995;37:915-932; **Ped Neur Briefs** March & Aug 1995, Jan 1996).

DOPA-RESPONSIVE MOTOR DISORDER WITH SEPIAPTERIN REDUCTASE DEFICIENCY

The clinical findings in 7 children from Malta, at first suspected to have cerebral palsy and later diagnosed with sepiapterin reductase deficiency, are reported from Great Ormond Street Hospital, London, UK, and St Luke's Hospital, University of Malta. All had early motor delay with diurnal variation and cognitive impairment. Oculogyric crises occurred from an early age in 6, hypotonia followed by dystonia in 5, chorea in 4, bulbar involvement in 3, and Parkinsonian tremor in 2. Treatment with L-dopa was started at ages 1 to 10 years in doses of 1.5 to 4 mg/kg/day, and the response was dramatic, except for aggravation of chorea. Improvement was obtained predominantly in motor function and control of oculogyric crises, but cognitive function and learning remained moderately impaired. A worsening of symptoms in hot weather was relieved by an increase in dose of L-dopa. All had a novel mutation in the tetrahydrobiopterin pathway involving sepiapterin reductase, with autosomal recessive inheritance. None had an abnormality in the gene encoding guanosine triphosphate cyclohydrolase 1 (GTPCH1), consistent with the autosomal dominant Segawa disease. (Neville BGR, Parascandalo R, Farrugia R, Felice A. Sepiapterin reductase deficiency: a congenital dopa-responsive motor and cognitive disorder. **Brain** October 2005;128:2291-2296). (Professor BGR Neville, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK).

COMMENT. This autosomal recessive, dopa-responsive congenital motor disorder, characterized by dystonia with diurnal variation and dramatic response to L-dopa, resembles in some respects the autosomal dominant GTPCH1, dopa responsive deficiency known as Segawa's disease. Unusual features include a congenital onset, early hypotonia and frequent bulbar involvement, atypical manifestations of Segawa's disease (**Ann Neurol** 2003;54(Supp 6);S32-45). Both conditions are often incorrectly diagnosed with unexplained cerebral palsy. The authors recommend that infants with unexplained CP should be screened for sepiapterin reductase deficiency, although outside Malta, the disease is probably very rare. The report adds to a range of cases of dopa-responsive motor disorders and the necessity for a trial of L-dopa when the diagnosis is unclear.

INFECTIOUS DISORDERS

AUTOANTIBODIES IN POST-EPSTEIN-BARR ACUTE CEREBELLAR ATAXIA

Eight of 23 patients with acute cerebellar ataxia (ACA) following Epstein-Barr virus (EBV) infection (proven serologically) had increased IgM anti-triosephosphate isomerase (TPI) antibody titers, in a study at Kyorin University, Tokyo, and Kinki University, Osaka,