Seizures were absence, generalized tonic-clonic, partial and complex partial in patterns. Seizures had disappeared at follow-up in 3 and epileptic activity in the EEG was absent in 4. The course of the aphasia was linked to the appearance and disappearance of electrical status epilepticus during slow sleep (ESES) which was found in 3 patients. Lasting EEG abnormalities and persisting clinical seizures, combined with the presence and duration of ESES, may impede language recovery. The disappearance of ESES may be associated with an improvement of language functions. The course of the aphasia was variable: slow and continuous improvement in 3, rapid recovery in 1, fluctuating in 1, and deterioration followed by slow improvement in 1. The follow-up period varied from 3 to 19 years. The response of the seizures and aphasia to anticonvulsants was variable; some benefit was obtained with phenytoin and phenobarbital in 1 patient, ethosuximide and valproate were helpful in 2. Prednisone in 1 patient had a variable and inconclusive effect (Paquier PF, Van Dongen HR, Loonen MCB. The Landau-Kleffner Syndrome or "acquired aphasia with convulsive disorder" Arch Neurol April 1992; 49:354-359). (Reprints: Dr. Van Dongen, Department of Neurology, Room EE 2287, University Hospital Dijkzigt-Rotterdam, P.O. Box 1738, 3000 DR Rotterdam, the Netherlands.)

COMMENT. The correlation between ESES and acquired epileptic aphasia has been emphasized previously (Jayakar PB, Seshia SS. J Clin Neurophysiol 1991; 7:299-311; Deonna TW. J Clin Neurophysiol 1991; 7:288-298). See also Ped Neur Briefs Sept 1991; 5:71; Jan 1991; 5:4-5). The beneficial effects of ACTH and cortico steroids have been reported from Tel Aviv University (Lerman P et al. See Ped Neur Briefs April 1991; 5:28-29). Cortico steroids administered early were more effective than delayed treatments.

MOVEMENT DISORDERS

PROGNOSIS OF TOURETTE'S SYNDROME

A retrospective study of 58 adults with a diagnosis of Gilles de la Tourette's syndrome (TS) during childhood is reported from the Department of Neurological Sciences, Rush-Presbyterian-St. Luke's Medical Center, Chicago, IL. The 41 men and 17 women had a mean current age of 27.1 years and a mean age of tic onset of 6.9 years. Forty six percent had a first degree relative with tics. All patients had tics as adults and most had both motor and phonic tics. Coprolalia occurred in 4% of adults. No patient experienced extreme adult disability and tics caused minimal to mild intrusion into private and professional life. All had received specialized medical care for TS in childhood, but only 26% were currently seeing a physician for tic disorders. Maximal disability had occurred during adolescence in most subjects, but 98% graduated high school, and 90% were full time students or fully employed. Childhood tic severity and coprolalia during development had no predictive value for risk of adult moderate/severe tics. Mild tic severity in adulthood correlated with mild tics during adolescence. (Goetz CG et al. Adult tics in Gilles de la Tourette's syndrome: description and risk factors. Neurology

April 1992; 42:784-788). (Reprints: Dr. C.G. Goetz, Department of Neurological Sciences, Rush-Presbyterian-St. Luke's Medical Center, 1725 West Harrison St., Chicago, IL 60612.)

COMMENT. In a study of TS in monozygotic twins, Hyde et al. at the National Institute of Mental Health, Washington, D.C. observed a significant effect of birth weight on the phenotypic expression of TS. Tic scores were greater in the lower birth weight twin as compared with the heavier group. Postnatal factors were not identified and did not play a major role in the subsequent expression of TS (Neurology March 1992; 42:652-658). Global neuropsychological performance was more impaired in the more severely affected twin particularly in tests of attention and visuospatial perception (Hyde TM et al. Neurology April 1992; 42(Suppl 3):396). Hypnosis was of benefit in reducing involuntary tics in 6 of 7 unmedicated children age 6 to 15 years with TS (Hollander H et al. Neurology April 1992; 42(Suppl 3):239). Deprenyl, an MAO-B inhibitor, was a safe and effective treatment of attention deficit disorder with hyperactivity in 80% of 20 children with TS. None of the patients noted exacerbation of their tics or other adverse side effects at doses up to 15 mg/day (Jankovic J. Neurology April 1992; 42(Suppl 3):238).

MUSCLE DISORDERS

MITOCHONDRIAL MYOPATHY AND HYPOTONIA

Three children with hypotonia, cardiac impairment, and defects of the mitochondrial respiratory chain complexes, but no ragged red fibers, are reported from the Hopital de la Timone, Chemin de l'Armee, d'Afrique, Marseille, France. Case 1 shared clinical and metabolic features with fatal infantile myopathy associated with cytochrome *c* oxidase deficiency as described by DiMauro: neonatal hypotonia and weakness, respiratory failure, and severe lactic acidosis. Post-mortem studies at age 7 weeks showed complex IV reduced in the liver but not in the heart and quantitative analysis of mtDNA showed depletion in muscle. Case 2 showed intractable cardiomyopathy, cyclic neutropenia, and 3-methylglutaconic aciduria. The boy died suddenly at age 27 months. Case 3 presented at age 16 months as an acute hypokinetic hypertrophic cardiomyopathy with transient hypotonia and mild lactic acidosis. After the acute episode the boy gradually improved and the neurological and cardiac examinations were normal at the age of 3 years. All cases showed lipid storage myopathy and decreased cytochrome *c* oxidase. Biochemical studies confirmed the cytochrome *c* oxidase deficiency in muscle in all cases (Figarella-Branger D et al. Defects of the mitochondrial respiratory chain complexes in three pediatric cases with hypotonia and cardiac involvement. (J Neurol Sci March 1992; 108:105-113.) (Correspondence: Dr. D. Figarella-Branger, Laboratoire d'Anatomie Pathologique, Hopital de la Timone, Chemin de l'Armee d'Afrique, F-13005 Marseille, France.)