

extensive localizations and those with normal CTs often had areas of hypo or hyperperfusion. The pattern seen on SPECT was related to the clinical course and prognosis: extensive metabolic impairment on SPECT correlated with frequent seizure recurrence and mental retardation whereas all children with a normal SPECT had less than 2 seizures per year and normal neurological and intellectual development. (Denays R et al. Arch Dis Child Oct 1988;63:1184).

### INVOLUNTARY MOVEMENT DISORDERS

#### TRANSIENT TICS AND TOURETTE'S SYNDROME

The relation of transient tic disorder (TTD - motor or vocal tics lasting less than 1 year) to Tourette's syndrome (TS- multiple motor and vocal tics lasting longer than 1 year) has been evaluated in two Canadian Mennonite families at the Dept of Neurology, Univ of Rochester Sch of Med, NY. One girl aged 9 yrs experienced frequent repetitive throat clearing and eye blinking episodes that resolved after several months (TTD). Her father and brother had TS and 2 siblings had chronic tic disorder (CTC-motor or vocal tics, but not both, with duration more than 1 year). A boy aged 12 yrs had frequent head jerks resolving over several months (TTD). His father had TS and sibling had CTD. In adulthood, the patient had 3 children, one with TS, (Kurlan R et al. Transient tic disorder and the spectrum of Tourette's syndrome. Arch Neurol Nov 1988;45:1200-1201).

COMMENT. Transient tic disorder may be an expression of the TS gene that may be transmitted to offspring as an autosomal dominant. The frequency of transient tics in childhood is quoted at 4-16%. The observations in this study suggest that TS and related tic disorders are more prevalent than generally appreciated.

#### DYSTONIA AND HYPOPARATHYROIDISM

Recurrent attacks of flexion of the right hand and arm and bowing of the head initiated by sudden movement were associated with idiopathic hypoparathyroidism in a 12 yr old boy seen at the Children's Hospital of Philadelphia, PA. An EEG showed a right frontal spike wave focus, and attacks were initially controlled by phenytoin. The boy later developed massive myoclonic spasms of the legs. CT scan revealed calcification of the basal ganglia, frontal lobes, and the right cerebellar hemisphere. The serum calcium was 5.6 and phosphorus 11 mg/dl. The spasms responded to ergocalciferol, 25000 units twice daily, and calcium lactate. The authors propose this association of "paroxysmal dystonic choreoathetosis and hypoparathyroidism" as a syndrome distinct from "familial paroxysmal choreoathetosis without hypocalcemia" and "movement reflex epilepsy". They cite 2 similar cases in the literature. (Barabas G. Tucker SM. Idiopathic hypoparathyroidism and paroxysmal dystonic choreoathetosis. Ann Neurol Oct 1988;24:585).

COMMENT. In the differential diagnosis of dystonia, a CT scan showing calcification in the basal ganglia lead to tests for hypoparathyroidism whereas an MRI may be helpful in the diagnosis of Hallervorden-Spatz disease. Dystonia and striking MRI abnormalities in the globus pallidus ("eye-of-the-tiger" sign) are described in 2 patients with Hallervorden-spatz syndrome (Sethi KD et al. Ann Neurol Nov 1988;24:692). One patient developed arching of the body backward and a diagnosis of dystonia at 14 years. She slowly deteriorated and at age 20 had progressive difficulty with mouth closure and at 32 years, frequent falling. CT scan of the head was normal whereas the MRI T2-weighted images showed a low-signal area surrounding a relatively circumscribed region of high signal in the globus pallidus. The 'eye-of-the-tiger' sign is an appropriate name for this abnormality. The second patient, a 20-year-old woman, had a 1-year history of progressive difficulty with night vision and repeated forceful eye closure aggravated by sudden noise and attempted eye opening. The neurological findings were frequent blepharospasm, repetitive slow tongue protrusion, and tapetoretinal degeneration. Slow deterioration occurred over the next 2 years and reexamination showed facial grimacing, severe blepharospasm, tongue protrusion, resting tremor of hands, tongue, and jaw, and cogwheel rigidity of arms. CT scan was normal and the MRI showed the "eye-of-tiger" sign in the globus pallidus.

### CNS INFECTIONS

#### BACTERIAL MENINGITIS AND DEAFNESS

Dexamethasone (.15 mg/kg/bwt q 6 hr for 4 days) was considered beneficial in the treatment of infants and children with bacterial meningitis, particularly in preventing deafness, in two double blind, placebo-controlled trials involving 200 patients treated in the Dept of Pediatrics, University of Texas at Southwestern Medical Center, Dallas, TX. As compared to 98 patients receiving placebo, 102 treated with dexamethasone became afebrile earlier (1.6 vs 5 days;  $P < .001$ ) and were less likely to acquire bilateral sensorineural hearing loss (15.5 vs 3.3%;  $P < .01$ ). Twelve patients in the 2 placebo groups (14%) had severe bilateral loss as compared with 1(1%) in the 2 dexamethasone groups ( $P < .001$ ). (Lebel MH et al. Dexamethasone therapy for bacterial meningitis. Results of 2 double-blind, placebo-controlled trials. N Eng J Med Oct 13 1988;319:964-71).

COMMENT. An editorial in the same issue (Smith AL. Neurological sequelae of meningitis N Engl J Med 1988;319:1012) applauds the investigators for undertaking a difficult and complex study but notes that enthusiasm for the findings is dampened by the lack of follow-up of all patients enrolled in the study. The patients may have been restudied too early to detect improvement in auditory acuity