MOVEMENT DISORDERS

TRANSIENT DYSTONIA IN LOW-BIRTHWEIGHT INFANTS

The long-term neuromotor, cognitive, and behavioral development of very low-birthweight (VLBW) infants with transient dystonia, and without cerebral palsy, was evaluated at the Department of Paediatrics, University of Bergen, Norway. Of 50 VLBW infants, 14 had transient dystonia (7 to 18 months) that was associated with low 1 min Apgar scores, bacterial infection, and lack of breast milk. At 5 years of age, Peabody Developmental Motor Scales, WPPSI, and Personality and Yale Children's Inventories showed scores that were comparable in infants with or without transient dystonia, although dystonic children had a greater tendency to eye-hand incoordination and gait problems. (Sommerfelt K, Pederesen S, Ellertsen B, Markestad T. Transient dystonia in non-handicapped low-birthweight infants and later neurodevelopment. Acta Paediatr Dec 1996;85:1445-1449). (Respond: Dr K Sommerfelt, Department of Paediatrics, Barneklinikken, 5021 Haukeland Sykehus, Norway).

COMMENT. Dystonia or increased muscle tone during the first year of life in VLBW children may be associated with later development of incoordination and motor problems but is not a risk factor for greater cognitive or behavioral impairments.

Determination of head circumference at 8 months of age is predictive of subsequent cognitive and behavioral problems in VLBW infants; subnormal head size is associated with lower IQ and higher incidence of hyperactivity at 8 years of age. (Progress in Pediatric Neurology II, PNB Publ, 1994:p194). Improvements in neonatal growth and development may depend on breast feeding and nutrition of chronically ill, low-birthweight neonates and environmental enrichment after discharge from hospital.

DEGENERATIVE DISORDERS

ATAXIA WITH VITAMIN E DEFICIENCY

Vitamin E adminstration (100 U/kg daily) resulted in improvements in ataxia, motor strength, and mobility in 2 siblings of a consanguineous Bedouin family with Friedreich ataxia and low serum vitamin E levels treated at Soroka Medical Center, Beer Sheva, Israel. The 2 siblings who responded to vitamin E had spinocerebellar symptoms but no peripheral neuropathy. Two siblings complicated by neuropathy failed to respond. Erythrocytes of vitamin E-deficent patients had reduced permeability to acidified glycerol that was partially corrected by vitamin E. (Shorer Z, Parvari R, Bril G, Sela B-A, Moses S. Ataxia with isolated vitamin E deficiency in four siblings. Pediatr Neurol Nov 1996;15:340-343). (Respond: Dr Shorer, Pediatric Neurological Unit, Soroka Medical Center, POB 151, Beer Sheva 84105, Israel).

COMMENT. Ataxia with isolated vitamin E deficiency is a rare type of Friedreich ataxia which may be benefited by early administration of large doses of vitamin E. Patients may be vitamin E deficient despite normal intake in the diet. (For further cases and commentary, see Progress in Pediatric Neurology II, PNB Publ, 1994:p497).