Patricia K Duffner, Department of Neurology, Children's Hospital of Buffalo, 219 Bryant Street, Buffalo, NY 14222).

COMMENT. The authors conclude that the longterm chemotherapy regimen for infantile brain tumors may delay and decrease the risk of irradiation-induced neurotoxicity at the expense of increasing the risk of secondary malignancies.

In an editorial, "Rethinking brain tumors in babies and more," Fisher PG of Stanford University comments that these discouraging results of prolonged chemotherapy should stimulate research in the clinical biology of brain tumors and prompt a more selective approach to aggressive oncology (Ann Neurol Sept 1998;44:300-302).

NEUROMUSCULAR DISORDERS

HEREDITARY INCLUSION BODY MYOPATHY

A new familial, autosomal dominant, myopathy and variant of hereditary inclusion body myopathy (HIBM) is described in 19 members of a large Swedish family followed in the Departments of Pediatrics, Genetics, and Pathology, Sahlgrenska University Hospital, Goteborg, Sweden. Onset was in the newborn period with congenital joint contractures in 14, hip dislocation in 4, limb-girdle weakness and muscular atrophy, external ophthalmoplegia, and decreased tendon reflexes. The course was nonprogressive in childhood, and joint contractures resolved. From 30 to 50 years of age, most patients showed deterioration, with progressive muscle weakness and atrophy, especially of quadriceps. EMG showed myopathic changes, and serum CK was elevated. Muscle biopsy showed focal disorganization of myofilaments in childhood cases, and dystrophic changes in adults, with rimmed vacuoles and cytoplasmic and intranuclear inclusions. (Darin N, Kyllerman M, Wahlstrom J, Martinsson T, Oldfors A. Autosomal dominant myopathy with congenital joint contractures, ophthalmoplegia, and rimmed vacuoles. Ann Neurol Aug 1998;44:242-248). (Respond: Dr N Darin, Department of Pediatrics, Sahlgrenska University Hospital-East, S-416 85 Goteborg, Sweden).

COMMENT. Inclusion body myopathies are sporadic and inflammatory or familial and hereditary. The above Swedish family appears to suffer from a unique form of autosomal dominant HIBM that presents at birth and shows a progressive deterioration in adult life.

PROGNOSIS OF BENIGN CONGENITAL HYPOTONIA

Twenty five children diagnosed with benign congenital hypotonia (BCH) between infancy and 2 years of age were examined at 6 to 8 years of age and compared to 26 controls, matched for sex, age, and weight, in a study at the School of Occupational Therapy, Hebrew University-Hadassah Medical School, and Child Development Institute, Jerusalem. Sensory, visual-perception, visual-motor integration, and behavioral measures were similar in the 2 groups, but the BCH group showed impairments in gross motor performance, bilateral coordination and strength on the Oseretsky Test of Motor Proficiency, despite recovery of near normal muscle tone. (Parush S, Yehezkehel I, Tenenbaum A et al. Developmental correlates of school-age children with a history of benign congenital hypotonia. Dev Med Child Neurol July 1998;40:448-452). (Respond: Dr Shula Parush, School of Occupational Therapy, Hebrew University-Hadassah Medical School, PO Box 24026, Mount Scopus, Jerusalem, Israel 91240).