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).

COMMENT. Children diagnosed with benign congenital hypotonia in infancy should be reexamined at intervals through early childhood and should receive extended occupational and other therapy to build muscle strength, balance, and coordination. Hypotonic infants may appear to recover near normal tone by 6 years of age but demonstrate clumsiness and generalized muscle weakness on tests of motor proficiency. My own clinical experience would substantiate these findings. Whereas the office neurological examination appears normal, the mother complains that the child lacks normal stamina, tires easily on extended walks, and often wants to be carried. The symptoms described are sometimes suggestive of a possible myasthenia.

ACQUIRED NEONATAL BRACHIAL PLEXUS PALSY

Three infants presenting at 3, 15, and 21 days of age with brachial-plexus neuropathy were found to have a group-B streptococcal osteomyelitis, as reported from the British Columbia's Children's Hospital, Vancouver, Canada. Osteomyelitis was not recognized initially because the infants were afebrile and generally well. Patient 1 was normal at birth and for the first two days. The parents observed the left arm limp and painful when touched or moved on the 3rd day. An initial diagnosis of traumatic birth injury was changed at 15 days, on examination by neurologists, when the shoulder became warm and swollen, and reflexes were found intact. Blood cultures grew group-B streptococcus and radiographs showed a lytic lesion in the left humerus. EMG and nerve conduction studies on day 28 were consistent with brachial-plexus neuropathy. Intravenous penicillin for 6 weeks was followed by complete recovery by 3 months. Patients 2 and 3 had a similar history and recovered following penicillin therapy. (Sadleir LG, Connolly MB. Acquired brachial-plexus neuropathy in the neonate: a rare presentation of late-onset group-B streptococcal osteomyelitis. Dev Med Child Neurol July 1998;40:496-499). (Respond: Dr Mary B Connolly, Division of Neurology, Department of Paediatrics, British Columbia's Children's Hospital, 4480 Oak St, Vancouver, BC V6H 3V4, Canada).

COMMENT. The authors provide a list of the non-traumatic causes of brachial-plexus neuropathy. In addition to the most common obstetrical injury, a variety of rare, non-traumatic forms are reported, including vaccinations, cytomegalovirus, toxoplasmosis, congenital syphilis, and other infections. Brachial-plexus palsy associated with osteomyelitis may represent a pseudopalsy secondary to pain, or a true paralysis with involvement of the nerves by ischemia or swelling. Nerve conduction studies in the present case reports favor a true paralysis. Early diagnosis and intravenous penicillin therapy result in full recovery. Possible infection in bone should be considered in newborn infants who present soon after a non-traumatic birth with a painful limb paralysis resembling Erb's palsy, despite the absence of fever. For reports of the outcome and diagnosis of obstetric brachial plexus palsy, see Progress in Pediatric Neurology III, PNB Publishers, 1997;pp357-359.

INFLAMMATORY DEMYELINATING POLYRADICULOPATHY

A clinical and electrodiagnostic, retrospective study of 43 children with acute inflammatory demyelinating polyradiculopathy (AIDP) is reported from the Service de Neuropediatrie, Hopital de Bicetre, France. Age of onset ranged from one to 18 years, less than 3 years in 35%. An antecedent infection occurred within 2 months of onset in 74%. Presenting symptoms included pain (47%), weakness (23%), ataxia (15%), and sensory signs (5%). When the neurological impairment had ascended and reached a plateau, weakness was noted in 100%, pain in 63%,