

this number, 14% occurred in the group treated early and 41% in the group with the longest period of delay in treatment. (Oller-Daurella L, Oller L F-V. Influence of the "lost time" on the outcome of epilep- sy. Eur Neurol May/June 1991; 31:175-177).

COMMENT. The sooner the correct diagnosis of epilepsy is made and treatment is begun, the fewer seizures a patient will suffer, and the greater the likelihood of successful antiepileptic control and subsequent withdrawal of antiepileptic drugs. Gowers, in 1881, pointed out that seizures beget seizures, and the greater the number of epileptic seizures the greater the likelihood of their continued reoccurrence. The results of this study should caution those who advocate delays in the initiation of anticonvulsant therapy and should encourage a more vigorous attempt to prevent seizure recurrences after the first epileptic seizure.

#### TONIC UPGAZE OF CHILDHOOD

A child with intermittent upward deviation of the eyes is reported from the Neuropediatric Unit CHUV, Lausanne, Switzerland. The boy was normal until nine months of age when brief intermittent upward eye deviation was noted and one month later these movements occurred for very long periods. At 14 months, vertical jerking of the eyes was associated with difficulty in downward gaze. He walked late at 16 months and fell often. When first examined at 21 months, the intermittent tonic upgaze lasted hours or days and was associated with a compensatory posture of the head, tilted with chin down. A downbeat nystagmus occurred when attempting to look down. His gait was wide-based and unsteady. The EEG, CT scan, NMR, and CSF exams were normal. The symptoms fluctuated and increased with fatigue and intercurrent illness. They were less marked in the morning on awakening from sleep. Treatment with acetazolamide was without effect. When last seen at 39 months of age the abnormal eye movements and head posture had almost resolved and the ataxia was mild. Since age 18 months he had had episodes of cyanosis, loss of contact, hypotonia, and falling, sometimes triggered by an emotional situation and resembling breathholding spells. (Deonna T et al. Benign paroxysmal tonic upgaze of childhood - a new syndrome. Neuropediatrics Nov 1990; 21:213-214).

COMMENT. This syndrome was first described by Ouvrier RA and Billson MD (J Child Neurol 1988; 3:177-180). These authors reported four cases. Ahn and Hoyt reported three infants with a similar syndrome (See Ped Neur Briefs Jan 1989). The eye movements are not seizures and improvement following levodopa therapy in one child suggests a closer analogy with dopa-sensitive dystonia.

#### CNS NEOPLASMS

##### COGNITIVE DEFICITS IN BRAIN TUMOR SURVIVORS

The results of studies of cognitive deficits in long-term survivors of childhood brain tumors are summarized from 31 published

reports between 1968-1989 in a review paper from the Division of Child Neurology, The Children's Hospital, Philadelphia and the Department of Neurology, Children's National Medical Center, George Washington University, Washington, DC. The increasing use of neuropsychological testing and the development of prospective studies during the first half of the 1980s has shown that from 40% to 100% of long-term survivors have some form of cognitive deficit in various intelligence quotients, visual/perceptual skills, learning abilities, and adaptive behavior. More cognitive deficits are detected the larger the time lapse from treatment to the testing evaluation. Factors predictive of subsequent cognitive deficits included a younger age at diagnosis, radiotherapy, methotrexate chemotherapy, and tumor location. Cognitive deficits occurred more frequently when medulloblastomas adhered to the brain stem, when tumors extended to the hypothalamus, and with hemispheric tumors. Surgery and preoperative hydrocephalus did not appear to modify cognition. (Glauser TA, Packer RJ. Cognitive deficits in long-term survivors of childhood brain tumors. Child's Nerv Syst Feb 1991; 7:2-12).

COMMENT. The quality of life for long-term survivors of childhood brain tumors becomes more important as improvements in treatment are developed. Long-term follow-up with neuropsychological tests is necessary since significant progressive deterioration in cognitive function may be demonstrated as the testing interval following treatment increases. (Duffner PK et al. Ann Neurol 1988; 23:575).

#### STROKE AS LATE SEQUELA OF CRANIAL IRRADIATION

The development of cerebrovascular disease manifested by strokes or transient ischemic attacks six months to four years after treatment of brain tumors in 11 children is reported from the Children's Hospital of Los Angeles, University of Southern California School of Medicine, Los Angeles, CA. All patients had received radiation therapy and seven chemotherapy. One patient, aged 4, with an incompletely resected craniopharyngioma, developed a right hemiparesis two years after treatment followed by left-sided focal motor seizures with quick progression to decerebrate posturing, central hyperventilation, coma, and death. Autopsy revealed bilateral cerebral infarctions as well as diffuse gliosis. Branches of the carotid artery showed severe thickening with subintimal proliferation of connective tissue. Smaller vessels showed cellular intimal proliferation with areas of leukocytic infiltration. (Mitchell WG et al. Stroke as a late sequela of cranial irradiation for childhood brain tumors. J Child Neurol April 1991; 6:128-133).

COMMENT. Damage to medium and large intracranial vessels in addition to small vessel damage may result in late onset of stroke or transient ischemic attacks caused by radiation and chemotherapy in children with brain tumors. The combination of methotrexate and cranial irradiation is more likely to produce a leukoencephalopathy than either treatment alone. The addition of chemotherapy may potentiate the damage to endothelial cells produced by radiation (mineralizing microangiopathy).