

concomitant thrombocytopenia. (Ballaban-Gil K, Callahan C, O'Dell C, Pappo M, Moshe S, Shinnar S. Complications of the ketogenic diet. Epilepsia July 1998;39:744-748). (Reprints: Dr K Ballaban-Gil, Epilepsy Management Center, Montefiore Medical Center, 111 E 210th St, Bronx, NY 10467).

COMMENT. Using the Johns Hopkins protocol for the ketogenic diet (KD) treatment of childhood refractory epilepsy, serious complications may occur in 10% of patients, often within one month of initiating the diet in a 4:1/ketogenic: antiketogenic ratio. Personally, I have preferred the Mayo Clinic method, which does not usually require the patient's hospitalization nor an initial period of starvation. The ratio of ketogenic to antiketogenic foods is modified over a 4 day period, beginning with a 1.1:1 ratio on the first day, 1.6:1 on the 2nd, 2.2:1 on the 3rd, and 2.8:1 on the 4th. A 4:1 ratio is rarely required to obtain ketosis and only in older children (Millichap JG. Nutrition, Diet, and Child's Behavior, Springfield, IL, Charles C Thomas, 1986). With this Mayo method of KD introduction, I have not encountered the adverse reactions reported with the Hopkins protocol. The concomitant use of valproate with the Hopkins ketogenic diet may also be a factor in the incidence of adverse events reported.

## BRAIN NEOPLASMS

### PROGNOSIS OF MEDULLOBLASTOMA

The prognostic role of clinical, pathological, and therapeutic factors in cases of medulloblastoma reported in the literature are reviewed at the University of Turin, Italy. Improvements in neurosurgical techniques and the addition of cranioc-spinal axis radiotherapy and chemotherapy can account for a drop in operative mortality from 32% before the 1960s to 10% after 1970, and an increase in 5-year survival rate from 2-11% to 50-70% in the same time periods. Local recurrence in the posterior fossa is the most frequent cause of failed treatment, and higher doses of irradiation (>50 Gy) may prolong survival but have long-term side effects. Low-dose cranioc-spinal radiotherapy plus adjuvant chemotherapy provides survival rates similar to those of high-dose irradiation, with less sequelae. Treatment is planned according to Chang's staging scheme, cases showing distant seeding receiving chemotherapy. The extent of surgical resection plays a controversial role in prognosis, but many surgeons favor a wide excision. The role of age at diagnosis is also uncertain, some authors concluding that 5-year survival is lower in children under 5 years, while others have found a better prognosis for infants, younger than 2 years. The prognostic significance of the pathology and cell differentiation of the tumor has been studied extensively with conflicting results. Tumors with a high proliferative rate are more susceptible to apoptosis and respond best to radiation. Almost 50% of medulloblastomas analyzed genetically exhibit deletion of the short arm of chromosome 17, a finding linked to shortened survival in some studies. (Giordana MT, Schiffer P, Schiffer D. Prognostic factors in medulloblastoma. Child's Nerv Syst June 1998;14:256-262). (Respond: Dr MT Giordana, Division of Neurology, Dept of Neuroscience, University of Turin, Via Cherasco, 15, 1-10126 Turin, Italy).

COMMENT. The prognosis of medulloblastoma is generally poor, 50% having recurrence, with death resulting within 5 to 7 years after diagnosis. Relapses after 8 years freedom from recurrence are rare. Factors having a significant influence on survival include radiotherapy and metastases at the time of diagnosis. The extent of surgical excision may also be important. Ongoing

research in cytogenetics of medulloblastoma may provide a better understanding of prognosis. See Progress in Pediatric Neurology II, PNB Publ,1994;p346, for reviews of long-term neurologic problems with medulloblastoma. Surveillance brain scans failed to detect recurrent disease and had no impact on outcome.

**Indications for ventricular drainage and V-P shunting in posterior fossa tumors** are reviewed from the Dept of Neurosurgery, Gdansk Medical University, Poland (Imielinski BL et al. Child's Nerv Syst 1998;14:227-229). Of 21 children with medulloblastoma, 20 had V-P shunts, 18 before and 2 after tumor resection. Symptomatic hydrocephalus occurred in 16 cases. Indications for V-P shunt included nonoperable tumor, acute hydrocephalus, and persistently elevated intracranial pressure after tumor removal.

**Multiple shunt failures in hydrocephalic children** are analysed for relevant factors at the Division of Neurosurgery, UCLA School of Medicine, Los Angeles, CA. (Lazareff JA et al. Child's Nerv Syst 1998;14:271-275). Of 244 with shunts, 136 had no failure (predominantly congenital hydrocephalus), 52 had one revision, 34 had 2 or 3 revisions, and 22 had 4 or more revisions. As the number of failures increased, the interval between revisions shortened. Repeated revisions were associated with an increase in CSF monocytes.

## VASCULAR DISORDERS

### **BASAL GANGLIA A-V MALFORMATION AND WRITER'S CRAMP**

A 12-year-old girl presenting with writer's cramp as the first manifestation of basal ganglia arteriovenous malformation (AVM) is reported from the Department of Neurosurgery, University of Tokyo, Japan. Difficulty in writing caused by too firm a pen grasp developed at 9 years of age and progressively worsened until at 11 years, she had weakness and involuntary movements of the right hand. At 12 years, she had severe headache with nausea and vomiting. MRI showed an unruptured left basal ganglia AVM localized to the globus pallidus and putamen, and extending to the left frontal lobe white matter. Cerebral angiography revealed a large high-flow AVM fed by lenticulostriate arteries. Treated conservatively, the dystonic cramp and weakness have not progressed during one year follow-up. (Kurita H, Sasaki T, Suzuki I, Kirino T. Basal ganglia arteriovenous malformation presenting as "writer's cramp." Child's Nerv Syst June 1998;14:285-287). Respond: Dr Hiroki Kurita, Dept of Neurosurgery, Faculty of Medicine, University of Tokyo, 7-3-1 Hongo, Bunkyo-ku, Tokyo, 113 Japan).

COMMENT. AV malformation presenting as writer's cramp is a novel case-report. The location and size of the lesion appear to have prompted conservative management. A review of the literature in 1994 revealed that AVM mortality was 23-57% with conservative management versus 8.5-11% postoperatively. The smaller the AVM, the higher the risk of hemorrhage and the greater the indication for surgery. (See Progress in Pediatric Neurology III, PNB Publ, 1997;p458).

### **CEREBRAL VASCULITIS IN JUVENILE RHEUMATOID ARTHRITIS**

A 16-year-old girl who developed a stroke after a 5-year history of polyarticular juvenile rheumatoid arthritis is reported from Tripler Army Medical Center, Honolulu, Hawaii. She was referred from Chuuk State, Micronesia,