

and/or polyspike-wave synchronous with myoclonus; interictal EEG was normal. Seizures were controlled by valproate, and medication was discontinued after a seizure-free period of 3 years. The average duration of follow-up was 6 years 9 months (range, 4yrs 9 mos-9 yrs 2 mos). At the end of follow-up, 2 children with early onset of seizures (7 months) had mild or moderate mental retardation, deficits in fine motor coordination, attention deficit disorder, and language impairment. Three children had a borderline IQ and 1 a low average IQ, with learning disorders and attention deficits. Only one patient had a normal IQ. All children were oppositional, irritable, aggressive, and hyperactive. The mean Full Scale IQ of the group was 74, 5 had ADD, and 4 had learning disorders. Early onset of seizures is a major risk factor for poor cognitive and behavioral outcome. A delay in starting treatment is an additional risk factor. (Mangano S, Fontana A, Cusumano L. Benign myoclonic epilepsy in infancy: neuropsychological and behavioural outcome. **Brain Dev** April 2005;27:218-223). (Respond: E-mail: manganos@katamail.com).

COMMENT. BMEI, as described by Dravet and Bureau (1981), is characterized by myoclonic seizures in previously healthy infants, 4-36 months of age, and responsive to antiepileptic medication. The unfavorable outcome reported in the above study compared to previous series was linked to early onset of seizures and delay in starting treatment. Alternatively, a pre-existing mild encephalopathy may have been responsible. The authors conclude that BMEI interferes with the growth of developing neurons, resulting in long-term neuropsychological impairment.

BRAIN NEOPLASMS

POSTOPERATIVE CHEMOTHERAPY FOR MEDULLOBLASTOMA

The survival rate and cognitive function of 43 children, age <3 years, with medulloblastoma treated with intensive postoperative chemotherapy alone, without radiotherapy, were determined at the University of Wurzburg and other centers in Germany. Chemotherapy consisted of three two-month cycles of cyclophosphamide, methotrexate, vincristine, carboplatin, and etoposide. Intraventricular methotrexate was also administered in 36 doses through an implanted subcutaneous reservoir, beginning 2 to 4 weeks after surgery. The five-year progression-free and overall survival rates (+/-SE) were 82+/-9% and 93+/-6% for 17 patients with complete resection, 50+/-13% and 56+/-14% for 14 with residual tumor, 33+/-14% and 38+/-15% for 12 with macroscopic metastases, and 68+/-8% and 77+/-8% in 31 patients without macroscopic metastases. Tumor relapse occurred in 9 of 29 patients without residual disease after chemotherapy. Independent risk factors for tumor relapse were desmoplastic form of medulloblastoma (20 patients), metastatic disease, and age younger than 2 years. Asymptomatic leukoencephalopathy was detected by MRI in 19 of 23 patients. After treatment with chemotherapy, the mean IQ was significantly lower than that of healthy controls of the same age but higher than that of patients treated by radiotherapy in a previous trial. (Rutkowski S, Bode U, Deinlein F et al. Treatment of early childhood medulloblastoma by postoperative chemotherapy alone. **N Engl J Med** March 10, 2005;352:978-986). (Reprints: Dr Rutkowski, Department of Pediatric Oncology, Children's Hospital, University of Wurzburg, Josef-Schneider Str 2, D-97080 Wurzburg, Germany or at rutkowski@mail.uni-wuerzburg.de).

COMMENT. Intensive postoperative chemotherapy that includes intraventricular methotrexate provides long remissions in children with medulloblastoma, and results are especially promising for patients without initial metastases. Except in cases of relapse, radiotherapy with its adverse side effects may be avoided in the very young patient with medulloblastoma. (DeAngelis LM. Chemotherapy for brain tumors—a new beginning. *N Engl J Med* 2005;352:1036-1037).

VASCULAR DISORDERS

RISK FACTORS FOR CEREBRAL VENOUS SINUS THROMBOSIS

The various clinical and neuroradiological presentations, frequency of associated prothrombotic risk factors, and predictors of outcome were studied in 42 consecutive children (ages 3 weeks to 13 years) with cerebral venous sinus thrombosis (CVST) seen at 5 European pediatric neurology centers and from stroke registries. The majority (83%) presented acutely with seizures, focal signs and symptoms of raised intracranial pressure. Twelve (28%) were in coma. CVST was demonstrated on MR venography. Pre-existing chronic illness in 17 patients included 4 with surgical procedures (cardiac, ventriculoperitoneal shunt, brain tumor resection, and colectomy for colitis), 8 with recent infections, 4 with dehydration, and 2 with lupus. Recent infection and dehydration were triggers for CVST in 25 previously well children. Anemia (Hgb <2 SD below the mean for age) was present in 22 (52%); 5 were hemolytic, including 2 with sickle cell disease and one with thalassemia. Iron deficiency in 50% of patients was found in 17 with anemia and in 4 with microcytosis and normal Hgb. A risk factor for thrombophilia was diagnosed in 18 of 29 (62%) screened, most commonly high factor VIII. Iron deficiency, parietal infarction and lack of caudate involvement independently predicted CVST rather than arterial stroke. Follow-up ranged from 0.5 to 10 years. Five patients died, and 26 had sequelae, including pseudotumor in 12, cognitive and/or behavioral disorders in 14, and associated with epilepsy in 3 and hemiparesis in 2. Independent predictors of a good cognitive outcome were older age group, use of anticoagulation therapy (18 patients), and lateral and/or sigmoid sinus involvement. Of 19 patients with follow-up MR venography, 3 had persistent occlusion, associated with anemia. (Sebire G, Taabarki B, Saunders DE et al. Cerebral venous sinus thrombosis in children: risk factors, presentation, diagnosis and outcome. *Brain* March 2005;128:477-4898). (Respond: Dr FJ Kirkham, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK).

COMMENT. Cerebral venous thrombosis should be excluded by MR venography in children with acute neurological symptoms, especially in patients presenting with headache who are anemic, have iron deficiency, or are suffering from recent infection and dehydration. The authors recommend early anticoagulation therapy except in comatose patients. Pseudotumor cerebri is a common sequel of CVST and may be underdiagnosed in young children.

CVST was reported in 3 girls with headache and systemic lupus erythematosus (Uziel Y et al. *J Pediatr* 1995;126:722-727). After low-dose oral anticoagulation and treatment for lupus, none had further thrombotic events in 10-18 month follow-up.