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,

because of right hemiparesis, facial weakness, joint pains, and cataract. MR angiography revealed vasculitis of the anterior and middle cerebral arteries. Both clinical and MR findings improved following treatment with methylprenisolone. Outpatient treatment with oral methotrexate, predisone, and aspirin resulted in complete remission. (Pedersen RC, Person DA. Cerebral vasculitis in an adolescent with juvenile rheumatoid arthritis. Pediatr Neurol July 1998;19:69-73). (Respond: Dr Pederson, Department of Pediatrics, 1 Jarrett White Road, Tripler Army Med Ctr, Honolulu, HI 96859).

COMMENT. Juvenile rheumatoid arthritis is an unusual cause of cerebral vasculitis and stroke in children. Cerebral vein thrombosis (CVT) and infarct associated with systemic lupus erythematosus (SLE) is reviewed in Progress in Pediatric Neurology III, PNB Publ, 1997;p173. Three girls with SLE and CVT, ages 11, 14, and 17, presented with a severe, persistent throbbing headache, unresponsive to analgesics.

DEVELOPMENTAL DISORDERS

CEREBELLAR MR SPECTROSCOPY IN WILLIAMS SYNDROME

Magnetic resonance spectroscopy was used to study brain biochemistry in 14 patients (age, 8-37 years) with Williams syndrome (WS) compared to 48 controls at the MRC Biochemical and Clinical Magnetic Resonance Unit, John Radcliffe Hospital, Oxford, UK. All patients had the facial dysmorphology typical of WS, and all showed the uneven cognitive-linguistic profile in psychological testing, with relative sparing of language and verbal skills and deficits in visuospatial, nonverbal tasks such as number, spatial cognition, planning, and problem solving. All WS subjects showed decreases in the phosphomonoester (PME) peak in 31P MRS ratios. Decreasing PME is associated with decreased cognitive performance. Ratios of choline- and creatine-containing compounds to N-acetylaspartate (Cho/Na and Cre/Na) were significantly elevated in the cerebellum of WS subjects, while the Cho/Cre ratio was not altered. The increased Na-containing ratios and suggested decrease in the cerebellar N-acetylaspartate (NA), a neuronal marker, correlated with impaired tests of cognitive ability, especially speed of processing. Cerebellar neuronal integrity may be important in cognition. (Rae C, Karmiloff-Smith A, Lee MA et al. Brain biochemistry in Williams syndrome. Evidence for a role of the cerebellum in cognition? Neurology July 1998;51:33-40). (Reprints: Dr Caroline Rae, Dept of Biochemistry, University of Sydney, 2006, Australia).

COMMENT. Williams syndrome (WS) is a rare genetic dysmorphic disorder with a hemizygous deletion on chromosome 7 and delayed motor and cognitive development. The chromosome abnormality affects a gene that programs elastin, accounting for the characteristic premature aging and wrinkles on the face, elfin facies, hernias, and supravalvular aortic stenosis. Adjacent genes included in the deletion may account for the mild mental retardation, visuospatial impairments, and disproportionate abilities in music and language (Rossen ML, Sarnat HB. Editorial. Why should neurologists be interested in Williams syndrome? Neurology July 1998;51:8-9). Biochemical abnormalities in the brain, demonstrated by magnetic resonance spectroscopy, show correlations with cognitive testing that may be specific for cerebellar dysfunction or may represent a more global cerebral anomaly. The dissociation between language and cognitive skills presents a specific neuropsychologic profile for WS. In one