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

report, seizures occurred in 50% of cases (See Progress in Pediatric Neurology I, PNB Publ, 1991;p318).

ABSENT LABIAL FRENULUM IN HOLOPROSENCEPHALY

Absence of the superior labial frenulum is reported in 88% of 17 consecutive cases of holoprosencephaly examined at St Christopher's Hospital for Children, Philadelphia, PA. Brain imaging is recommended in children with this anomaly. Patients with midline facial abnormalities such as hypotelorism, smooth philtrum, or single central incisor should be examined for absent frenulum. (Martin RA, Jones KL. Absence of the superior labial frenulum in holoprosencephaly: a new diagnostic sign. J Pediatr July 1998;133:151-153). (Reprints: Rick Martin MD, Section of Clinical Genetics, St Christopher's Hospital for Children, Erie @ Front St, Philadelphia, PA 19134).

COMMENT. Absence of the superior labial frenulum may be added to the spectrum of midline anomalies characteristic of the holoprosencephaly sequence.

TRAUMATIC DISORDERS

INFLICTED AND NONINFLICTED TRAUMATIC BRAIN INJURY

Neuroimaging, physical, and neurobehavioral findings after inflicted and noninflicted traumatic brain injury (TBI) in 40 children, 0 to 6 years of age, were compared in a prospective longitudinal study at the University of Texas at Houston Health Science Center, and Baylor College of Medicine, Houston, TX. Signs of preexisting brain injury (cerebral atrophy, subdural hygroma, and ex vacuo ventriculomegaly) were present in 45% of 20 children with inflicted TBI and in none of 20 with noninflicted TBI. Neuroimaging findings included subdural hematoma in 16 (80%) inflicted TBI cases and 9 (45%) noninflicted cases, and epidural hematoma in none of the inflicted cases and 4 (20%) of noninflicted TBI. Seizures (13 cases) and retinal hemorrhage (13) were significantly more frequent in the inflicted TBI cases than in noninflicted cases: 65% and 65% cf 15% and 0 cases, respectively. Skull fractures, cephalhematomas, edema, and intraparenchymal hemorrhage were similar in the two groups. Glasgow Outcome Scale scores, cognitive development, and motor functioning assessed 1.3 months after TBI showed a significantly less favorable outcome after inflicted than noninflicted TBI. Despite comparable Glasgow Coma Scale scores and similar duration of impaired consciousness, the inflicted TBI group had significantly more cases of mental deficiency than the noninflicted TBI group. (Ewing-Cobbs L, Kramer L, Prasad M et al. Neuroimaging, physical, and developmental findings after inflicted and noninflicted traumatic brain injury in young children. Pediatrics August 1998;102:300-307). (Reprints: Dr Linda Ewing-Cobbs, Department of Pediatrics, University of Texas at Houston Health Science Center, 6431 Fannin, Houston, TX 77030).

COMMENT. Features of inflicted traumatic brain injury include acute CT/MRI evidence of old brain injury, subdural hematoma, seizures, retinal hemorrhages, and impaired cognitive function without prolonged impairment of consciousness. Chronic changes on acute CT/MRI in 45% of children with inflicted TBI, suggesting previous assault and cumulative brain injury, are found without previous reported history of brain injury. Neuroimaging is essential in diagnosis of acute brain injury and for identification of old injuries, especially in shaking-impact injuries which may have no external signs of physical abuse.