

more days and were placed in foster care more often than control children. In order of decreasing frequency from 90% to 20%, the manifestations of CNS dysfunction were behavior problems, developmental delay, language delay, microcephaly, seizures, irritability, attention deficit disorder, hyperactivity, hearing loss, and learning disabilities. The most frequent facial dysmorphic features included a long, flat philtrum, low nasal bridge, short palpebral fissures, thin upper lip, ear malformations, flattened maxilla, short, upturned nose, and epicanthal folds. The frequencies of CNS dysfunction and dysmorphic features were lower in the incomplete FAS patients than in those with the full syndrome, except for ADHD, learning disabilities, mental retardation, flat philtrum, and low nasal bridge, which occurred with the same respective frequencies. (Kvigne VL, Leonardson GR, Neff-Smith M, et al. Characteristics of children who have full or incomplete fetal alcohol syndrome. **J Pediatr** November 2004;145:635-640). (Reprints: Thomas K Welty MD MPH, 5990 East Jeremy Lane, Flagstaff, AZ 86004).

COMMENT. Fetal alcohol syndrome diagnosis should meet all 5 of the following criteria: 1) prenatal alcohol exposure, 2) FAS suspected by physician, 3) one or more typical facial features, 4) growth deficiency, and 5) CNS impairment (CDC. **Mort Mor Wkly Rev** 1994;42:312-314). Children meeting only 1 to 4 of these criteria are defined as incomplete FAS. Children with FAS have numerous health, learning, and social needs. The diagnosis of FAS becomes more difficult as the child grows older since the pronounced growth retardation and dysmorphisms of early childhood tend to diminish with age. FAS in adolescence was studied in 44 patients followed for 10-14 years (Spohr HL et al. **Acta Paediatr** 1994;404:19-26). Although manifestations were less obvious, a characteristic "juvenile" pattern of FAS was recognized that included microcephaly, growth retardation, cognitive deficits, behavioral problems, and craniofacial dysmorphisms. (see **Progress in Pediatric Neurology III**, PNB Publ, 1997;524-525, for further articles on FAS).

VASCULAR DISORDERS

BRAIN BIOPSY IN DIAGNOSIS OF PRIMARY ANGIITIS OF THE CENTRAL NERVOUS SYSTEM

Two cases of histologically confirmed childhood primary angiitis of the central nervous system (PACNS) are reported from the University of California-San Diego, and Children's Hospital, San Diego, CA. Patient 1, a 12-year-old boy presented with morning headaches, vomiting, and perioral dysesthesias. Examination showed papilledema, ataxia, focal weakness, and brisk reflexes. CSF contained 16 white cells/mm³ (89% lymphocytes). Infections were excluded. Von Willebrand factor antigen was persistently elevated, and ESR was normal. MRI showed white matter abnormalities. Despite a normal angiogram, brain biopsy showed vasculitis involving larger leptomeningeal vessels. Treatment with steroids and cyclophosphamide resulted in resolution of weakness and ataxia and reduction of papilledema, with one period of relapse. Patient 2, a 3-year, 9-month-old girl presented with intermittent fever, vomiting, somnolence, and arthralgias. The child had chicken pox at 3 weeks of age. Blood count showed 13,000 leukocytes/mm³, and ESR was 81 mm/h. Infectious disease evaluation was negative. Symptoms resolved without treatment but recurred 3 months later. CSF revealed 150 WBC/mm³ (83% lymphocytes) but no pathogens.

MRI was normal at first but 3 months later, showed hyperintense, nonenhancing foci in the deep white matter. Eye exam showed anterior uveitis, responsive to topical corticosteroids. Within one month, a third episode with similar features, complicated by meningismus and photophobia, developed 3 days following a varicella zoster virus rash in the distribution of the left 4th thoracic root dermatome. CSF revealed 416 WBC/mm³ (31% neutrophils, 61% monocytes); PCR tests for VZV and HSV were negative. Angiogram was normal. Resolution of symptoms and MRI white matter changes followed steroid and methotrexate treatment. She was stable for 1 year and then relapsed with recurrence of fever, vomiting, meningismus, and uveitis, and also focal seizures. A right frontal brain and meningeal biopsy revealed perivascular lymphocytic infiltration of small to medium-sized vessels positive for CD4 lymphocytes. Remission followed treatment with steroids and methotrexate. (Yaari R, Anselm IA, Szer IS, et al. Childhood primary angiitis of the central nervous system: Two biopsy proven patients. *J Pediatr* November 2004;145:693-697). (Reprints: Joseph G Gleeson MD, UCSD School of Medicine, MTF 312, 9500 Gilman Drive, La Jolla, CA 92093).

COMMENT. These cases of primary CNS angiitis demonstrate the difficulties in diagnosis and the importance of early brain biopsy despite a normal angiogram. CSF examinations are typical of aseptic meningitis. The diagnosis of PACNS is based on clinical, radiographic, and biopsy correlation and exclusion of infection, toxin, drugs, neoplasm, or systemic disease. Ten previous references to cases with histological confirmation are cited. Treatment recommended is a combination of corticosteroid and cyclophosphamide. A VZV-induced vasculitis is a possible explanation in case 2; the negative CSF VZV PCR is against the diagnosis but CSF antiviral antibody titers were not known.

ATTENTION DEFICIT AND LEARNING DISORDERS

COGNITIVE DEFICITS AFTER FOCAL CEREBELLAR LESIONS

Patients with focal cerebellar lesions due to tumor or hematoma were evaluated by a neuropsychological test battery, neurological examination and MRI, and cognitive function was correlated with location of the lesions in a study of 21 adult patients at the Department of Neurosurgery, Christian Albrechts Universitat, Kiel, Germany. Compared to matched controls, patients showed deficits in general memory, delayed recall, and visual memory, but not in verbal memory; and deficits in executive function and in attentional processes such as working memory and divided attention. Patients with right-sided cerebellar hemisphere lesions were more impaired than those with left-sided lesions, and their deficits were verbal whereas those with left-sided lesions were more often non-verbal and spatial. The connection of the right cerebellum to the left cerebral hemisphere, which is dominant for language and right hand movements, explains the greater impairment of function with right-sided lesions. Motor impairments were not correlated with cognitive deficits. Cerebellar lesions lead to a “dysmetria of thought.” (Gottwald B, Wilde B, Mihajlovic Z, Mehndorn HM. Evidence for distinct cognitive deficits after focal cerebellar lesions. *J Neurol Neurosurg Psychiatry* November 2004;75:1524-1531). (Respond: Dr B Gottwald, Zentrum fur Integrative Psychiatrie ZIP, Niemannsweg 147, D-24105, Kiel, Germany).