

PEDIATRIC NEUROLOGY BRIEFS

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J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

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TOXIC DISORDERS

RADIATION-INDUCED PARTIAL EPILEPSY

Intractable epilepsy in 3 patients treated with low-dose irradiation to "strawberry" scalp nevi in infancy is reported from the Montreal Neurological Institute and Hospital, Montreal, Quebec, Canada. Focal seizures developed at ages 8 to 20 years. All patients had localized alopecia and EEG abnormalities corresponding with the irradiated site. MRI showed thinning of the cranial vault with expansion of the brain into the skull defect in one patient, and mild perisylvian atrophy in one other. Neurologic deficits were not progressive. (Reutens DC, Andermann F et al. Intractable partial epilepsy following low-dose scalp irradiation in infancy. *Ann Neurol* December 1995;38:951-954). (Respond: Dr Andermann, Montreal Neurological Institute, 3801 University St, Montreal, Quebec, Canada H3A 2B4).

COMMENT. Strawberry nevi invariably involute spontaneously before 10 years of age and scalp irradiation is no longer employed. The risk of epilepsy following low-dose irradiation is low, and may reflect induction of intracranial tumor or delayed cerebral radiation necrosis. The syndrome of delayed cerebral radiation necrosis is characterized by progressive neurological deficits and sometimes raised intracranial pressure developing months to years after irradiation. MRI shows cerebral atrophy, white matter lesions, and enhancing foci. In the above patient reports, the postradiation syndrome was nonprogressive but intractable, and one showed mild cerebral focal atrophy. Irradiation to the skull should be avoided whenever possible.

NEUROLOGIC COMPLICATIONS OF FETAL COCAINE EXPOSURE

Cocaine-positive urine toxicology at birth in 51 newborns was associated with hypertonia during infancy in 21(41%) studied at the Harlem Hospital Center, New York. Cocaine-positive infants were four times more likely to show hypertonic tetraparesis than cocaine-negative infants. Hypertonia diminished over time and resolved by 24 months. Those with early hypertonia showed significantly lower developmental scores at 6 and 12 months than

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infants without hypertonia. (Chiriboga CA et al. Neurological correlates of fetal cocaine exposure: Transient hypertonia of infancy and early childhood. Pediatrics December 1995;96:1070-1077). (Reprints: Dr CC Chiriboga, Division of Pediatric Neurology, College of Physicians and Surgeons, Columbia University, 710 West 168th St, New York, NY 10032).

COMMENT. In our clinic for children with Attention Deficit Disorders at Children's Memorial Hospital, Chicago, I have observed an unusual incidence of a history of fetal cocaine exposure in those placed in foster homes soon after birth. Other complications of cocaine exposure in utero are small head circumference, cerebral infarction or hemorrhage, seizures, and SIDS. Disturbances in corticogenesis have been demonstrated in experiments on laboratory animals (see Progress in Pediatric Neurology I and II, PNB Publishers, 1991, pp452-3, and 1994, pp439-41).

GLUTEN SENSITIVITY AND NEUROLOGICAL ILLNESS

The frequency of IgG and IgA antigliadin antibodies, a measure of cryptic gluten sensitivity, and celiac disease was studied using ELISA in 147 adult patients admitted to the Royal Hallamshire Hospital, Sheffield, UK, for neurologic investigation. Of 53 patients with neurological dysfunction of unknown cause, including 25 with ataxia and 20 with peripheral neuropathy, 30 (57%) had positive antigliadin antibody titers, compared to only 5% of 94 patients with specific diagnoses, such as stroke, MS, and Parkinsonism, and 12% of 50 healthy blood donors. In antigliadin-positive patients with ataxia or neuropathy of unknown cause, duodenal biopsies revealed histological evidence of celiac disease in 35% and non-specific duodenitis in 38%. Only one had low vitamin B12 levels and the biopsy was normal. Gluten sensitivity was a common finding in this group of adult patients with ataxia and peripheral neuropathy of unknown cause. (Hadjivassiliou M et al. Does cryptic gluten sensitivity play a part in neurological illness? Lancet February 10, 1996;347:369-71). (Respond: Dr M Hadjivassiliou, Department of Clinical Neurology, Royal Hallamshire Hospital, Sheffield S10 2JF, UK).

COMMENT. This investigation underscores the importance of nutrition and diet in some neurological disorders of undetermined etiology. Antigliadin antibody estimation should be considered in the investigation of patients with neurological dysfunction of unknown cause, including those with refractory seizures, and especially if associated with occipital calcifications. Patients with histological evidence of celiac disease are treated with a gluten-free diet. However, those celiac patients with seizure complications and occipital calcifications are not always benefited by diet, and surgical resection of the involved occipital cortex may be required. (see Progress in Pediatric Neurology II, PNB Publishers, 1994, pp71-73).

CRANIAL NERVE DISORDERS

CONGENITAL FACIAL PALSY

The association between permanent congenital facial palsy in 61 children and recognized risk factors for traumatic birth was investigated in a retrospective case control study at the Departments of Plastic Surgery, Mount Vernon Hospital, Northwood, Middlesex, and the Hospital for Sick Children, Great Ormond Street, London, UK. The incidence of forceps assisted delivery