VISUAL FUNCTION IN WEST SYNDROME

The range and severity of visual abnormalities in 25 infants with West syndrome, and their relation to seizures, EEG patterns and brain lesions were studied at Catholic University, Rome; University of Pisa; and University of Pavia, Italy. All had normal funduscopic examinations. Tests of visual function (acuity, visual field, fixation, pursuit, eye movement, and visual attention) showed at least one abnormal result on one or more of the tests in 22 children. At spasm onset, visual impairment was related to EEG sleep disorganization rather than the hypsarrhythmic pattern. After 2 months, both EEG features were significantly correlated with visual function. With seizure control after 2 months, visual function improved in several cases. Cryptogenic and tuberous sclerosis patients had good visual function, whereas other patients with symptomatic spasms had more frequent visual dysfunction. The most severe abnormalities of visual function occurred with lesions involving the basal ganglia, posterior cortical areas, and optic radiations, but some infants with normal MRIs also had impaired visual function. Fluctuations in visual performance of fixation and pursuit in 17 patients were not related to the early EEG changes. Hypsarrhythmia was significantly related to low scores in visual function only after the first 2 months. Improvement in visual function after 2 months was correlated with good or partial control of seizures in 9 cases and only 4 without seizure control. (Rando T, Bancale A, Baranello, et al. Visual function in infants with West syndrome: correlation with EEG patterns. Epilepsia 2004;45:781-786). (Reprints: Dr F Guzzetta, Division of Child Neurology and Psychiatry, Catholic University, Rome, Italy).

COMMENT. Visual dysfunction in infants with West syndrome is caused by various factors and not only by brain structural abnormalities. The epileptic disorder per se is correlated with visual function. In drug or ACTH-responsive cases (idiopathic infantile spasms) a transitory hypsarrhythmia is predictive of normal visual function, whereas a persistent hypsarrhythmia and resistant spasms are risk factors for impaired visual function and attention.

Visual inattention, an early manifestation of West syndrome (WS). Jambaque et al found that visual inattention in WS was frequently associated with parieto-occipital abnormalities in SPECT studies (Epilepsia July/Aug 1993; and Ped Neur Briefs Sept 1993); and Inuma et al reported that two thirds of 17 infants with visual abnormalities and occipital EEG discharges developed WS with hypsarrhythmia at follow-up (Epilepsia 1994;35:806-809; Ped Neur Briefs Nov 1994).

PROLONGED UNCONSCIOUSNESS AND DELIRIUM WITH FEBRILE SEIZURES

The incidence and duration of unconsciousness and delirious behavior in children with febrile seizures were studied at Nagoya University Graduate School of Medicine and other centers in Japan. In 203 patients with 213 consecutive febrile seizures the duration of seizures was less than 5 minutes in 90.2%, duration of unconsciousness was less than 30 min in 93%, and delirious behavior observed in 2% of patients persisted for 10 to 60 minutes. Delirium appeared before the onset of the febrile seizure and resolved after the seizure.