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SEIZURE DISORDERS

DEVELOPMENTAL OUTCOME OF WEST SYNDROME

Medical records of 32 patients with cryptogenic West syndrome were reviewed for factors correlating with developmental outcome in a study at Saitama Children's Medical Center, and the Jikei University School of Medicine, Tokyo, Japan. Clinical features were compared in a normal outcome group of 12 patients and a delayed outcome group of 20 patients. In the delayed outcome group, 5 patients had mild mental retardation (IQ, DQ 50-75), 8 were moderately retarded (IQ 25-50), and 7 were severely retarded (IQ <25). Age at onset of spasms and duration of follow-up were the same in the delayed and normal (IQ >75) outcome groups. The retarded group compared to the normal outcome group differed in the longer time from seizure onset to initiation of treatment (mean, 104.5 vs 40.8 days, $P<0.05$), and a longer time from onset to cessation of spasms (mean, 139.8 vs 77.0 days, NS). ACTH controlled spasms for > 28 days and hypsarrhythmia was resolved in all 32 patients initially (spasms disappeared after exanthem subitum in one patient without treatment). The duration from beginning ACTH to control of spasms was the same in both delayed and normal outcome groups (mean, 3.6 vs 2.8 days, NS). Relapse of spasms occurred in 7 of 20 delayed outcome patients (average time 2.8 +/- 1.6 months) and in none of the normal group ($P<0.05$). Other types of seizures (focal motor, complex partial, generalized) occurred in 11 of the delayed outcome group, evolving to focal epilepsy in 7. Paroxysmal EEG discharges did not reappear in 8 of the normal and 3 of the delayed outcome groups ($P<0.05$). Reappearance of EEG paroxysms occurred in 4 of the normal and 17 of delayed outcome groups; Average age of reappearance of EEG paroxysms was later in the delayed cf normal outcome groups (29.2 vs 10.8 months, $P<0.05$). Frontal and parieto-occipital regions were the most common locations for reappearance of paroxysmal discharges in the delayed outcome group. (Hamano S-I, Yoshinari S, Higurashi N, et al. Developmental outcomes of

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cryptogenic West syndrome. **J Pediatr** March 2007;150:295-299). (Reprints: Dr Shin-ichiro Hamano, Division of Neurology, Saitama Children's Medical Center, 2100 Magome, Iwatsuki-ku, Saitama-city, Saitama 339-8551, Japan).

COMMENT. The importance of early diagnosis and treatment of infantile spasms, demonstrated by this study, confirms previous reports of a correlation of improved response and prompt initiation of ACTH therapy (Millichap JG, Bickford RG. **JAMA** 1962;182:523-527). Of 21 patients referred to the Mayo Clinic, 80% of infants diagnosed and treated at < 1 year of age were benefited whereas only 22% older than 1 year at diagnosis showed reduction in spasms and EEG improvement. The shorter the treatment lag, the more favorable the outcome. (Koo B et al. **Neurology** 1993;43:2322-7; cited by authors).

In the above study, patients with a poor outcome have a greater risk of focal epilepsy and persistent paroxysmal EEG discharges in frontal regions. In a long-term study of 214 patients followed for 20-35 years at the University of Helsinki, Finland (Riikonen R. **Epilepsia** 1996;37:367-372), factors predictive of a good prognosis included cryptogenic etiology, normal development before onset of spasms, and a good response to ACTH. Focal abnormalities in the EEG were not necessarily indicative of a poor prognosis.

ICTAL SPECT FOCAL HYPERPERFUSION IN WEST SYNDROME

Ictal single photon emission computed tomography (SPECT) and EEG were used to determine the mechanism of clustered spasms in 3 patients with symptomatic West syndrome (WS), in a study at Tokushima University, Japan. Regional cerebral blood flow increased during ictus and decreased during the interictal period in the area coinciding with the CT/MRI delineated focal cerebral lesion. Ictal hyperperfusion of bilateral basal ganglia was detected in 2 of 3 patients. Ictal EEG showed a diffuse slow wave complex corresponding to a clinical spasm. Sharp waves that preceded the delta activity and spasm were located in the same area in which cerebral blood flow increased during ictus. No patient showed a partial seizure. Focal cortical discharge and secondary generalization generate cluster spasms and trigger the brain stem and basal ganglia to produce spasms. Administration of clonazepam or resection of the focal cortical lesion resulted in complete cessation of spasms and disappearance of hypsarrythmia. (Mori K, Toda Y, Hashimoto T et al. Patients with West syndrome whose ictal SPECT showed focal cortical hyperperfusion. **Brain Dev** May 2007;29:202-209). (Respond: Dr Kenji Mori, Department of Pediatrics, School of Medicine, Tokushima University, Kuramoto-cho, Tokushima, Japan).

COMMENT. A previous SPECT study from Tokushima University (Miyasaki M et al. **Epilepsia** 1994;35:988-992; **Ped Neur Briefs** Dec 1994) showed localized cerebral hypoperfusion in the temporal lobes in 7 of 10 patients with infantile spasms, EEG showed corresponding focal abnormalities in 5, and the MRI confirmed localized lesions in only 3. PET studies have shown hypermetabolism of the lenticular nuclei in 32 of 44 infants with both cryptogenic and symptomatic infantile spasms. (Chugani HT et al. **Ann Neurol** 1992;31:212-219; **Ped Neur Briefs** March 1992). Every infant with a focal lesion on CT/MRI had a focal abnormality on PET in the same location, but 17 of 28 infants with focal abnormalities on PET had no detectable focal abnormalities on CT/MRI. Chugani HT et al