

## CONGENITAL DEVELOPMENTAL DISORDERS

### **LISSENCEPHALY: EEG AND EVOKED POTENTIALS**

The EEGs and evoked potentials were studied in 21 Dutch patients with lissencephaly type I at the Departments of Clinical Neurophysiology and Neurology, Juliana Children's Hospital, The Hague, The Netherlands. Compared to patients with atypical cortical dysplasia and to a control group the EEGs in the lissencephaly patients showed the following patterns significantly more often: (a) generalized fast activity (8-18/s) with an amplitude higher than 50  $\mu$ V, (b) sharp- and slow-wave complexes with high amplitude (range 500 - 3000 $\mu$ V and (c) an alternating pattern consisting of bursts of sharp waves alternating with periods of electrocerebral depression. Short latency somatosensory evoked potentials in 10 patients after stimulation of the median nerve were abnormal. (de Rijk-van Andel J.F. et al. EEG and evoked potentials in a series of 21 patients with lissencephaly type I. Neuropediatrics Feb 1992; 23:4-9.) (Correspondence: J.F. de Rijk-van Andel, MD, Ignatius Ziekenhuis, Molengracht 21, 4818 CK Breda, The Netherlands.)

**COMMENT.** Lissencephaly may be differentiated from an atypical cortical dysplasia by CT and/or MRI. In addition, the EEG and evoked potentials may be of diagnostic value. Generalized fast activity and sharp- and slow-wave complexes were noted in 20 of the 21 lissencephaly patients and in only 1 of the 21 patients with atypical cortical dysplasia. An abnormal PET scan suggested the microscopic pathology confirmed at autopsy in an infant with lissencephaly and normal gross cortical structure. The clinical features were intractable seizures and developmental delay but normal head growth (Korobkin R, Sarnat H, Chugani H. Neurology April 1992; 4(Suppl 3):354). Radiographic and clinical variability within the 3 subtypes of lissencephaly are reviewed in 19 patients ages 1 day to 6 years (Demos DS et al. Neurology April 1992; 42(Suppl 3):286).

### **TUBEROUS SCLEROSIS: DIAGNOSTIC CRITERIA**

The Diagnostic Criteria Committee of the National Tuberos Sclerosis Association reports primary, secondary, and tertiary features of the disease complex and considers the specificity and reliability of the various lesions. The histological pattern of a lesion when available is generally more reliable than visual or radiographic appearances. Definite, probable and suspect diagnoses are categorized. (Roach ES et al. Report of the Diagnostic Criteria Committee of the National Tuberos Sclerosis Association. J Child Neurol April 1992; 7:221-224.) (Correspondence: Dr. E.S. Roach, University of Texas Southwestern Medical School, Department of Neurology, 5323 Harry Hines Blvd., Dallas, TX 75235.)

**COMMENT.** In an editorial, Dr. Robert M. Shuman notes that assumptions about the specificity of lesions will need to be tested. The Committee intends these criteria to be adaptable and modified as data are collected.