insipidus, and optic atrophy together with gait ataxia should suggest the diagnosis of Wolfram syndrome and indicate the need for an MRI, audiogram and renal sonogram.

RETT SYNDROME

The late infantile regression period (stages I and II) was analyzed retrospectively in 91 girls and women at the Department of Pediatrics, University of Goteborg and Pediatric Clinic, Ostersund Hospital, Sweden. The median age at onset of developmental stagnation (stage I) was 11 months, and loss of acquired skills (stage II) began at 19 months and lasted for 19 months. The onset of regression was distinct in 43%, dramatic in 16%, and insidious in 41%. The first observed signs of disease were delay in reaching expected gross motor milestones, dissociation of motor development, and disequilibrium. A triad of manifestations characterized the deterioration or loss of acquired skills: contact/communication, hand use/skill, babble/words. End of regression occurred at mean age 2.5 years. The girls gradually became more alert and showed an interest to act and interact. Handedness showed a preference for the left hand compared to the right, and was associated with spike activity in the left central leads in the EEG in 13 of 20 girls. (Engerstrom IW. Rett syndrome: the late infantile regression period - a retrospective analysis of 91 cases. Acta Paediatr Feb 1992; 81:167-172.) (Correspondence: Dr. Ingegerd Witt Engerstrom, Pediatric Habilitation Center, Tallasvagen 4, S-83142 Ostersund, Sweden.)

COMMENT. This further delineation of the stages of Rett syndrome may help in the diagnosis and pathophysiology of the disorder. In a study involving the analysis of beta-endorphin, as well as lactate, pyruvate and metabolites of norepinephrine, dopamine and serotonin in CSF from 12 girls with Rett's syndrome, the most consistent and significant difference from a control group was the elevation of beta-endorphin immunoreactivity in the CSF. Myer EC et al. from the Department of Child Neurology, Medical College of Virginia, Richmond VA found increased levels of beta-endorphin immunoreactivity in lumbar CSF in 90% of 158 Rett syndrome patients. (Neurology Feb 1992; 42:357-360.) The authors note that the symptoms of Rett syndrome are similar to the effects of centrally administered beta-endorphin or other opioids in experimental animals.

SEIZURE DISORDERS

RECURRENT STATUS EPILEPTICUS

The risk of recurrent status epilepticus in 95 children followed prospectively for a mean of 29 months was determined at the Montefiore Medical Center and affiliated hospitals of Albert Einstein College of Medicine, Bronx, New York. Ages ranged from 1 month to 18 years (mean 4 years). The cause was idiopathic in 24, remote symptomatic in 18, febrile in 29, acute asymptomatic in 18 or a progressive neurologic disorder in 6. Neurologically abnormal children (34% of the study population) accounted for

88% of the recurrent status group and for all 5 children with multiple episodes of status epilepticus. The risk was related to the severity of the neurological abnormality, the multiply handicapped children being at highest risk. Fifteen of 16 children with recurrent status epilepticus were being treated with anti-epileptic drugs at the time of recurrence. The morbidity and mortality of status epilepticus were low. (Shinnar S et al. Recurrent status epilepticus in children. Ann Neurol June 1992; 31:598-604.) (Correspondence: Dr. Shinnar, Division of Pediatric Neurology, VCP-207, Montefiore Medical Center, 111 E. 210th St., Bronx, NY 10467.)

COMMENT. The authors indicate that status epilepticus in neurologically normal children is by and large an isolated event that should not unduly influence decisions regarding further therapy. The high risk subgroup of neurologically abnormal children with recurrent status epilepticus underscored the need for better treatment strategies.

The more frequent use of neurosurgical treatment of refractory status epilepticus is suggested by Gorman DG et al. in a report from the UCLA Pediatric Epilepsy Research Program, Los Angeles, CA. (Epilepsia May/June 1992; 33:546-549.) The resection of a right frontal focus in a 9 year old boy with left focal motor seizures and status epilepticus was followed by immediate control of the seizures and no seizures for more than 1 year post-operatively while using carbamazepine monotherapy.

EEG MAPPING AND BECT

Sequential topographic EEG mapping performed to differentiate "epileptic" from "non-epileptic" rolandic spikes has shown a characteristic pattern significantly related to classical BECT (benign focal epilepsy of childhood with centro-temporal spikes) in a study at the Department of Clinical Neurophysiology and Department of Child Neurology, University Hospital Utrecht, The Netherlands. There were 2 patterns: 1) stationary potential fields, and 2) non-stationary potential fields. The non-stationary patterns represented by a double spike showed a specific sequence starting with a dipolar field, with the negative pole in the frontal region and the positive pole in the centro-temporal region, and changing to a unipolar or dipolar field, with a negative potential field in the centro-temporal region and a simultaneous positive potential field in the frontal region. This characteristic pattern appeared to be significantly related to classical BECT. (Van der Meij W. et al. Sequential EEG mapping may differentiate "epileptic" from "non-epileptic" rolandic spikes. EEG and Clin Neurophysiol June 1992; 82:408-414.) (Correspondence: Dr. W. Van der Meij, Department of Clinical Neurophysiology, University Hospital Utrecht, P.O. Box 85500, 3508 GA Utrecht (The Netherlands).)

COMMENT. The spike morphology in 40 uncontrolled childhood focal epilepsy patients was quantified by a computer-based technique and analyzed in relationship to epileptic syndromes at the Baylor College of Medicine, Houston, TX. The spikes of patients in the benign focal epilepsy category differed from those in other syndromes; they were