al. Seizures induced by exercise. Neurology April 1988; 38: 633-634).

COMMENT. A cardiac pathogenesis is usually suspected when seizures occur during physical exercise, as noted by the authors. A full cardiac examination including EKG and echocardiogram is advisable in patients with seizures occurring during physical exertion, even in children. Exercise is an unusual precipitant of seizures and is generally accompanied by a reduction in EEG seizure discharges. A clinical seizure was not induced but a spike and wave discharge occurred in each of these three patients after 5, 7½ and 27 minutes of pedaling a stationary bicycle.

CORN OIL KETOGENIC DIET

The successful substitution of corn oil for MCT oil in 6 children treated with the ketogenic diet for intractable seizures is reported from the Depts of Pediatrics, Neurology and Psychiatry, University of Arkansas for Medical Sciences and Arkansas Children's Hospital, 804 Wolfe St., Little Rock, AR. Seizure types were mixed in all 6 patients, absence in 5, minor motor in 4, myoclonic in 3, and complex partial and generalized tonic-clonic in 1. All had been controlled with MCT oil diets but corn oil has the major advantages of being less expensive, more readily available without prescription, and better tolerated. Anticonvulsants were reduced in 5 patients and eliminated in 3, without deterioration in seizure control. (Woody RC et al. Corn oil ketogenic diet for children with intractable seizures. J Child Neurol Jan 1988; 3: 21-24.

COMMENT. The medium chain triglyceride (MCT) oil was advocated by Huttenlocher et al. (Neurology 1971; 21: 1097) as a substitute for dietary fats in the ketogenic diet. MCT's are more ketogenic and less restrictive of carbohydrates, they are more rapidly absorbed than dietary fat and may induce ketosis more quickly. A disadvantage of the MCT diet is the frequency of gastrointestinal side-effects, many patients suffering from bulky, loose stools, diarrhea, vomiting and abdominal pain. Perhaps the superiority and availability of corn oil will encourage a renewed interest in the ketogenic diet for the treatment of refractory seizures in children.

FOCAL MYOCLONUS AND CORTICAL DYSPLASIA

Four patients aged 9 to 19 years with focal myoclonus and intractable focal motor seizures beginning at age 4 to 6 years were treated surgically at the Montreal Neurological Hospital and Institute, McGill University, Montreal, Quebec, Canada. All 4 had a mild progressive hemiparesis, cognitive deficits, focal EEG seizure discharges in the contralateral rolandic areas, 3 showed cortical architectonic abnormalities on MRI, and evidence of focal cortical dysplasia with abnormally wide gyri at operation. All improved following surgery. The authors stress the value of the MRI in visualization and diagnosis of neuronal migration cortical dysplasias whereas other imaging techniques had been unrevealing. (Kuzniecky R, Berkovic S. Andermann F (correspondent) et al. Focal cortical myoclonus and rolandic cortical dysplasia; clarification by magnetic resonance imaging. Ann Neurol April 1988; 23: 317-325).

COMMENT. The authors consider the clinical, MRI and pathological

features of these 4 patients sufficiently similar to suggest a clinical entity not previously described. The focal seizure pattern, hemiparesis, focal epileptiform EEG discharges and focal cortical dysplasia all indicated a localized lesion, yet a generalized abnormality of EEG background activity and widespread cognitive defects pointed to a more diffuse cerebral dysfunction as well. The differential diagnosis included tumor, post-traumatic or ischemic lesions, metabolic disorder, and chronic localized encephalitis of Rasmussen T et al. (Neurology 1958; 8: 435). The report of macrogyria in 3 of the patients and giant astrocytes in both patients with histological studies is suggestive of a forme fruste tuberous sclerosis that might be entertained in the differential diagnosis.

DEGENERATIVE AND METABOLIC DISORDERS

RETT SYNDROME: DIAGNOSTIC CRITERIA

Diagnostic criteria for Rett Syndrome are proposed by the International Rett Syndrome Association and the Centers for Disease Control, Koger Center, F-37, Atlanta, GA. The criteria are separated into three categories: 1) necessary, 2) supportive, and 3) exclusion criteria. Female sex is not included as a necessary criterion because the possibility of undiagnosed male cases cannot be ruled out. Diagnosis is tentative unil 2-5 yrs of age, and the presence of one or more of the exclusion criteria is against the diagnosis, regardless of whether all of the necessary criteria have been met.

Necessary criteria include the following: 1) normal pregnancy, birth and psychomotor development through the first 6 or 18 months; 2) normal head circ at birth and deceleration of head growth between 5 mos and 4 yrs; 3) loss of purposeful hand skills between 6 and 30 months; 4) impaired language and psychomotor development; 5) stereotypic hand movements such as hand wringing; and 6) gait apraxia and ataxia between 1 and 4 years. Supportive criteria include breathing irregularities, EEG abnormalities, seizures, spasticity, scoliosis, growth retardation and small feet. Evidence of intrauterine growth retardation or perinatal acquired brain damage, microcephaly at birth, identifiable metabolic, degenerative or storage diseases are listed as exclusion criteria.

The clinical characteristics of Rett Syndrome and differential diagnoses are listed according to stages and age at onset: 1) Early onset deceleration stage, 6-18 mos; 2) rapid "destructive" stage, 1-3 yrs; 3) pseudostationary stage, 2-10 yrs; 4) late motor deterioration stage, 10+ years. (Trevathan E, Moser HW et al. The Rett Syndrome diagnostic criteria work group. Diagnostic criteria for Rett Syndrome. Ann Neurol April 1988; 23: 425-428).

COMMENT. Heller's dementia, an infantile dementia described in 1908, almost 60 years before the first description of Rett Syndrome, should be added to the differential diagnosis (Millichap JG. Lancet 1987; 1: 440; Rett A and Olsson B. Dev Med & Child Neurol 1987; 29: 835), especially as the female sex is no longer considered a necessary diagnostic criterion for Rett Syndrome. At this stage of our understanding, the diagnostic criteria of Rett Syndrome should not be too strict and too exclusive (Opitz J. Am J Med Genet 1986;