COMMENT. PET studies of human brain functional development have shown that the prefrontal and temporoparietal regions are relatively inactive before the sixth month of life and an adult pattern is seen at one year of age. (Chugani HT et al. Ann Neurol 1987; 22:487-497). The theory of developmental arrest in infants with Rett syndrome is supported by the finding of an immature pattern of cerebral blood flow. However, the results of the present study conflict with those of Naidu et al who found an increase in the metabolism in the frontal region and a lower metabolism in the occipital region of two patients with Rett syndrome studied with PET (J Child Neurol 1988; 3 (suppl): S78-S86).

SURAL NERVE AXONOPATHY AND RETT SYNDROME

The histopathologic findings of three sural nerve biopsies and one muscle biopsy from three patients with Rett syndrome are described from the Department of Pediatrics, National Sanatorium Yakumo Hospital, Yakumo, Hokkaido, Japan. The biopsies demonstrated mitochondrial changes in the cytoplasm of Schwann cells, occasional onion bulb formations, and mitochondrial alterations in myelinated axons with reduction in the number of large myelinated fibers. The muscle showed small dark angulated fibers with NADH-TR staining and dumbbell-shaped mitochondria. (Wakai S et al. Rett syndrome: Findings suggesting axonopathy and mitochondrial abnormalities. Pediatr Neurol Sept/Oct 1990; 6:339-343).

COMMENT. These findings suggest peripheral nerve involvement and mitochondrial abnormalities in Rett syndrome. An additional article describes cerebellar pathology at autopsy of five patients with Rett syndrome (Oldfors A et al. Rett syndrome: Cerebellar pathology. Pediatr Neurol Sept/Oct 1990; 6:310-314). Patients ranged in age from 7 to 30 years. All had reduced brain weights with small cerebella. There was loss of Purkinje cells, atrophy, astrocytic gliosis of molecular and granular cell layers, and gliosis and loss of myelin in the white matter. The cerebellar atrophy was greater in two patients treated with phenytoin. The pathology of Rett syndrome appears to involve the cerebellum, cerebral hemispheres, basal ganglia, especially substantia nigra, spinal cord, peripheral nerve, and muscle.

DEVELOPMENTAL DISORDERS

PRADER-WILLI SYNDROME IN NEONATES

A retrospective study of 16 patients identifying physical features of neonates with Prader-Willi syndrome is reported from the Department of Pediatrics, Division of Genetics, William Beaumont Hospital, Royal Oak, MI and Section of Genetics, Department of Pediatrics, University of Arizona, Tucson. Medical records of 16