

FOCAL NEUROLOGIC DEFICITS WITH HYPOGLYCEMIA AND DIABETES

Focal neurologic deficits on 19 occasions in seven children with diabetes are reported from Children's Hospital, Winnipeg, Manitoba, Canada. None had a history of a seizure disorder or a febrile seizure prior to onset of diabetes. One had generalized seizures attributed to hypoglycemia. Two had a parent with migraine. The neurological examination was normal between events, all of which occurred during sleep. All seven patients had at least one episode of a focal motor deficit. Two children had separate episodes of right and left-sided hemiparesis, two had focal motor seizures followed by ipsilateral paresis, and two had headache beginning after the onset of the acute neurologic event. Hypoglycemia was demonstrated on the three occasions that blood glucose was measured at the outset of hemiparesis or focal seizures. (Wayne EA et al. Focal neurologic deficits associated with hypoglycemia in children with diabetes. J Pediat Oct 1990; 117: 575-577).

COMMENT. The authors consider that focal seizures induced by hypoglycemia are the most likely cause of these transient neurologic deficits and that the parietic episodes are examples of Todd paralysis. The administration of sugar did not always result in reversal of the neurologic deficit and the neurological symptoms were not a direct consequence of hypoglycemia. Hemiplegic migraine seemed to be an unlikely explanation. The anticonvulsant phenytoin has a hyperglycemic effect (Belton NR et al. Epilepsia 1965; 6:234) and may be beneficial in the treatment of seizures and transient hemiparesis associated with hypoglycemia in diabetic children.

CEREBRAL BLOOD FLOW IN RETT SYNDROME

Cerebral blood flow was studied with single photon emission computed tomography in seven girls with Rett syndrome at the John F. Kennedy Institute, Glostrup, Denmark and the Department of Clinical Physiology and Nuclear Medicine, Bispebjerg Hospital, Copenhagen, Denmark. Compared to results in an age matched control group of nine normal children, global cerebral blood flow was significantly lower in patients with Rett syndrome (54 vs 69 mL/100 g per minute). The blood flows in prefrontal and temporoparietal association regions of the telencephalon were markedly reduced, whereas the primary sensorimotor regions were relatively spared. The cerebral blood flow distribution in Rett syndrome was similar to the distribution of brain metabolic activity in infants of a few months of age. The most striking difference between the Rett syndrome and control groups was the pronounced frontal hypoperfusion in the Rett syndrome group; patients had a 30% lower anteroposterior flow ratio than the control group. These changes were not reflected in CT scans which showed cortical and central atrophy only in two of the seven patients. The age range of the patients was between 6.7 and 17.9 years with a median of 10.1 years. (Nielsen JB et al. Immature pattern of brain activity in Rett syndrome. Arch Neurol Sept 1990; 47:982-986).

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