Hagberg B et al. Late onset globoid cell leukoencephalopathy (Krabbe's disease) - Swedish case with 15 years of follow-up. Acta Paediatr Feb 1995;84:218-21). The disease presented with visual dysfunction at 4 years of age. At 8 years he developed a limp and ataxia and within 6 months he was wheel-chair dependent. Epilepsy began at 14 years. Speech became dysarthric on entering school, but he was able to stay in the mainstream educational system. Leukocyte galactosylceramidase activity was reduced.

CONGENITAL LACTIC ACIDOSIS: PET AND MRS STUDIES

Positron emission tomography (PET) and proton magnetic resonance spectroscopy (MRS) identified an increase in rate of cerebral glycolysis (PET) and cerebral lactate (MRS) in 2 children with defective mitochondrial respiration and congenital lactic acidosis studied at the Universitat zu Koln, Germany, and the University Hospital, Nijmegen, The Netherlands. These changes were not apparent in a child with lactic acidosis and normal respiratory chain activity. Defects of oxidative phosphorylation may cause increases in glycolysis and accumulation of cerebral lactate. (Duncan DB et al. Positron emission tomography and magnetic resonance spectroscopy of cerebral glycolysis in children with congenital lactic acidosis. Ann Neurol March 1995;37:351-358). (Respond: Prof Dr Heiss, Max-Planck-Institut fur neurologische Forschung, Gleueler Str 50, 50931 Koln, Germany).

COMMENT. PET and MRS have been used to demonstrate the metabolic changes associated with defective mitochondrial respiration in the brain without resort to diagnostic muscle biopsy. For further discussion of MRS in mitochondrial disorders, see Progress in Pediatric Neurology II, 1994, p454.

VASCULAR DISORDERS

NEONATAL MIDDLE CEREBRAL ARTERY STROKE

The presentation, EEG, imaging studies, and outcome of six term neonates with middle cerebral artery infarcts are reported from Mannheim Hospital, University of Heidelberg, Germany. Birth was by cesarean section in 5 cases. Apgars were normal in 4. Seizures occurred in 4 within 1 to 3 days and in 2 at the 5th and 9th days. EEGs showed focal slowing followed by focal spike-wave activity, correlating with the structural lesion defined by neuroimaging. EEG abnormalities sometimes antedated a positive ultrasound. A late intra-uterine event was suggested. Only 2 had obvious hemiparesis at discharge, but all children showed developmental delay and spastic hemiparesis at 3 to 10 year follow up. One had infantile spasms and hemihypsarrhythmia, relieved only after drainage of a cyst and shunt procedure. Four developed epilepsy late. (Koelfen W, Freund M, Varnholt V. Neonatal stroke involving the middle cerebral artery in term infants: Clinical presentation, EEG and imaging studies, and outcome. Dev Med Child Neur March 1995;37:204-212). (Respond: Dr Wolfgang Koelfen, Kinderklinik Mannheim, Theodor Kutzer Ufer, 68127 Mannheim, Germany).

COMMENT. This study confirms previous reports of late onset of epilepsy and development of spastic hemiparesis and cognitive deficits following an apparent early favorable outcome in term neonates suffering middle

cerebral artery infarcts.

LATE PROGRESSIVE THALAMIC ATROPHY in four children with neonatal middle cerebral artery infarction is reported from the Centre Hospitalo-Universitaire, Dijon, France. (Giroud M, Dumas R et al. Child's Nerv Syst March 1995;11:133-136).

DEVELOPMENTAL DELAY AFTER HEART SURGERY

The developmental and neurologic status of 155 children were evaluated one year after heart surgery for D-transposition of the great arteries, comparing those randomly assigned to circulatory arrest or low-flow cardiopulmonary bypass, at Children's Hospital, Boston, MA. Circulatory arrest was associated with lower scores on the Bayley Scales of Infant Development, and the Psychomotor Development Index was inversely related to the duration of circulatory arrest. Risk of neurologic abnormalities also increased with the duration of circulatory arrest. A ventricular septal defect, present in 35 (23%), and seizure activity detected by continuous EEG monitoring in the early postoperative period were independent risk factors for a poor outcome. MRI abnormalities, and mental development and visual memory test scores were not correlated with the method of circulatory support. (Bellinger DC, Newburger JW et al. Developmental and neurologic status of children after heart surgery with hypothermic circulatory arrest or low-flow cardiopulmonary bypass. N Engl J Med March 2, 1995;332:549-55). (Reprints: Dr Newburger, Department of Cardiology, Children's Hospital, 300 Longwood Ave, Boston, MA 02115).

COMMENT. This report is a follow up of the perioperative neurologic effects of hypothermic circulatory arrest compared to low-flow cardiopulmonary bypass in 171 patients operated within the first three months of age at the Children's Hospital, Boston. (Newburger JW et al. N Engl J Med 1993;329:1057-64). Circulatory arrest was associated with a higher incidence of clinical and EEG seizures in the first 6 hours after surgery, but the incidence of neurologic abnormalities was similar in the two groups at time of discharge. (See Progress in Pediatric Neurology II, 1994, p386). It now appears that the technical advantages of total circulatory arrest may be outweighed by delayed motor development and neurologic abnormalities at one year and potential cognitive deficits at school age.

NEURODEVELOPMENTAL OUTCOME OF 11 INFANTS WITH SURGERY FOR HYPOPLASTIC LEFT HEART SYNDROME is reported from the State University of New York and Children's Hospital of Buffalo. (Rogers BT et al. J Pediatr March 1995;126:496-8). Testing at a mean age of 38 months showed microcephaly in 8 (73%), mental retardation in 7 (64%), gross motor delays in 5 (45%), and severe cerebral palsy in 2 (18%). The quality of life is obviously seriously impaired in these patients, a factor to be considered in treatment options.

BREATH-HOLDING SPELLS AND ANEMIA

Two children, ages 32 months and 19 months, with breath-holding spells that resolved after treatment for concomitant anemia, are reported from the