

An overview of the surgery of hydrocephalus: past, present and future, by Hirsch JF, Hopital Necker Enfants Malades, Paris, France is published in Acta Neurochir (Wien) 1992; 116:155-160. The need for research in the causes and prevention of hydrocephalus is stressed.

NEONATAL CEREBRAL VENOUS THROMBOSIS

The clinical presentation and management of 7 neonates with idiopathic cerebral venous thrombosis (CVT) are reported from the Floating Hospital for Infants and Children, New England Medical Center Hospitals, Tufts University School of Medicine, Boston, MA. Unexplained seizures in 3 infants and unexplained lethargy in 4 led to neurological evaluation and the diagnosis of CVT. Follow-up neurological evaluation through 6 months of age showed normal development. Conventional magnetic resonance images indicated more extensive thrombosis than was suggested by cranial CT. MR phase imaging confirmed thrombosis and established absence of blood flow in cerebral veins or sinuses. No infant received treatment with anticoagulants. (Rivkin MJ et al. Neonatal idiopathic cerebral venous thrombosis: an unrecognized cause of transient seizures or lethargy. Ann Neurol July 1992; 32:51-56.) (Correspondence: Dr. Rivkin, New England Medical Center Hospitals, Box 330, Division of Pediatric Neurology, 750 Washington St., Boston, MA 02111.)

COMMENT. Cerebral venous thrombosis may explain transient neonatal seizures or lethargy in newborn infants and may be diagnosed by MR and MR phase imaging.

METABOLIC DISORDERS

HYDROXYGLUTARIC ACIDEMIA

Increased urinary excretion of L-2-hydroxyglutaric acid, found in 8 mentally retarded patients from 5 unrelated families including 3 pairs of siblings seen in various centers in Europe, was reported from University Hospital Amsterdam, the Netherlands. The first symptoms occurred between 1 and 5 years of age and consisted of abnormal gait. Febrile seizures occurred in 7 of 8 patients. Cerebellar dysfunction was the principle neurological abnormality in later childhood and adulthood. MRI revealed subcortical leukoencephalopathy, cerebellar atrophy, and changes in the putamina and dentate nuclei. L-2-hydroxyglutaric acid was also increased in the CSF and plasma. Lysine was increased in CSF and plasma and glutaric acid was increased in the plasma of 2 patients. Loading and dietary studies failed to reveal the origin of the L-2-hydroxyglutaric acid. (Barth PG et al. L-2-Hydroxyglutaric acidemia: a novel inherited neurometabolic disease. Ann Neurol July 1992; 32:66-71.)

COMMENT. Glutaric acidemia, an autosomal recessively inherited disease caused by deficiency of glutaryl-Coenzyme A dehydrogenase, was manifested by acute dystonia in 3 infants reported from the Children's Hospital of Pittsburgh, PA (See Ped Neurol Briefs Jan