children's MEDICAL CENTER
The One for Children℠

FaxNotes
from Children's Medical Center Dallas

A MONTHLY UPDATE FOR THE PHYSICIAN COMMUNITY • JANUARY 7, 2008

GRAND ROUNDS

Jan. 9
Neonatal Herpes Simplex Virus Infection: presentation, treatment, and prevention
David W. Kimberlin, M.D., Professor of Pediatrics, University of Alabama at Birmingham.

Jan. 16
The Neuroscience, Taxonomy and Treatment of Maladaptive Aggression
Hans Steiner, M.D., Professor of Psychiatry and Behavioral Sciences, Child Psychiatry and Child Development, Department of Psychiatry and Behavioral Sciences, Stanford University School of Medicine.

Jan. 23
Abusive Head Trauma in Dallas: Fact, Fiction, and Future Directions
Matthew Cox, M.D., Assistant Professor of Pediatrics, Children's Medical Center.

Jan. 30
Update on Pediatric Diabetes Care
Bryan Dickson, MD, Associate Professor of Pediatrics, Medical Director, Endocrinology Clinic, Children's Medical Center.

Also available via videoconference at selected area hospitals, call 214-345-2330 for locations.

MEDICAL UPDATE: Neurometabolic Diseases and Hypoglycorrhachia

Whether acute or chronic, expected or incidentally encountered, hypoglycorrhachia always signifies central nervous system dysfunction and an increased risk for neural cell injury. Severe but brief (probably less than 30-minute) hypoglycemia is tolerable: cognitive and behavioral abnormalities may be followed by seizures, hemiparesis and coma, in no predetermined order, until all measurable cerebral electrical activity ceases and the electroencephalogram (EEG) f lattens, but normal neurological performance (including the EEG signal) returns when blood glucose is restored. Hypoglycorrhachia, however, heralds longer-standing problems.

The principal and all too well-known causes of hypoglycorrhachia are bacterial meningitis, hypoglycemia, aseptic meningitis, subarachnoid hemmorrhage and meningeal carcinomatosis. Few of these diseases evade clinical attention today. Others, however, are often missed when documented in the outpatient setting, despite being the most amenable to direct interpretation. Among these, disorders of cerebral energy metabolism stand out. The brain's energy budget includes provisions to supply and sustain a power consumption of 500 W, most of which derives from glucose. It is no surprise then that both the deficiency of the cerebral glucose transporter Glut1 and many mitochondrial diseases cause encephalopathy, but it is also not uncommon to encounter these diseases manifesting in subtle, nondescript ways.

In addition to epilepsy, a series of disturbances — including minimal mental retardation, ataxia or intermittent balance abnormalities, dystonia or abnormal limb postures, deceleration of head growth, and opsoclonus or rapid eye movements — are frequently caused by these diseases. In the case of Glut1 deficiency (a largely sporadic, treatable inborn error of metabolism also transmissible as an autosomal dominant trait), the defect in glucose transport accounts for the hypoglycorrhachia; in the case of mitochondrial diseases, increased anaerobic glycolysis probably results in reduction of glucose f lux to the CSF.

For more information on neurometabolic diseases and hypoglycorrhachia, contact Juan Pascual, M.D. at 214-456-2768 or juan.pascual@childrens.com.

UPCOMING CONFERENCE: Solutions in Childhood Obesity

"Solutions in Childhood Obesity: Identifying and treating patients in the primary care setting," will be presented Saturday, Feb. 23, at Children's Medical Center. The conference will educate primary care physicians regarding the identification and treatment of overweight and obese children, including identifying co-morbid conditions and risk factors. The $100 registration fee includes syllabus materials, breakfast and lunch. The University of Texas Southwestern Medical Center at Dallas, the accredited sponsor, is jointly sponsoring the event with Children's Medical Center. This activity has been approved for 7 AMA PRA Category 1 Credits™. To register or for more information, call 214-456-LEAN or send an email to leanfamilies@childrens.com.