Decreased executive functioning was strongly correlated with increased individual variation in lifetime average PHA levels. Maintained PHA levels of <360 mcmol/l may be necessary for optimal outcome in PKU children. (Arnold GL, Kramer BM, Kirby RS et al. Factors affecting cognitive, motor, behavioral and executive functioning in children with phenylketonuria. Acta Paediatr May 1998;87:565-570). (Respond: Dr G Arnold, Division of Pediatrics Genetics, University of Rochester School of Medicine, 601 Elmwood Ave, Box 777, Rochester, NY 14642).

COMMENT. These data support a strict dietary control of PKU, maintaining phenylalanine levels below 360 mcmol/l throughout childhood. Executive behavior and motor development, both dependent on frontal lobe function, are correlated with the degree of individual variation in phenylalanine levels during childhood. An increased incidence in attention deficit hyperactivity disorder might be expected in PKU children with frontal lobe dysfunction.

WERNICKE'S ENCEPHALOPATHY WITH SELF STARVATION

A 10-year-old boy was referred for inpatient psychiatric evaluation at the Children's National Medical Center, Washington, DC, because of a 2-month history of food refusal and a 9 kg weight loss after choking on a raisin. He ate only carbonated, sweetened beverages and occasional soft food. Complaining of diplopia, he was initially diagnosed with conversion disorder, and was treated for dehydration with 10% dextrose and water IV. He developed ptosis, vertical and horizontal nystagmus, ophthalmoplegia, ataxia and mild encephalopathy. Thiamine concentration was 0.12 mcmol/L (N. 0.16-0.23). Treatment with thiamine 100 mg IV was followed by neurologic improvement within 12 hours. Attention, short-term memory, and orientation to place and time were slower to resolve than eye muscle paresis and ataxia. He accepted a full diet after 1 week, and recovery was almost complete in 1 month (Gropman AL, Gaillard WD, Campbell P, Charya SV. Wernicke's encephalopathy due to self starvation in a child. Lancet June 6, 1998;351:1704-1705). (Respond: Dr AL Gropman, Departments of Neurology and Psychiatry, Children's National Medical Center, Washington, DC).

COMMENT. Wernicke's encephalopathy is rare in children in the US. Reduction of whole blood transketolase enzyme is a diagnostic test for thiamine deficiency (Menkes JH. Textbook of Child Neurology. 3rd ed. Philadelphia, Lea & Febiger, 1985). Infants fed thiamine-deficient formula, and children with emesis and weight loss during chemotherapy are at risk of Wernicke's disease. The diagnosis should be suspected in malnourished infants and children, especially those with persistent vomiting. Reports of 9 cases and pathological findings at autopsy are included in Progress in Pediatric Neurology I, PNB Publ, 1991;pp544-5). The diagnosis is easily missed during life, especially in acute cases. IV glucose may precipitate symptoms of WE by depleting thiamine levels.

INFECTIOUS DISORDERS

HIV ENCEPHALOPATHY IN PERINATALLY ACQUIRED DISEASE

The incidence and clinical progression of HIV encephalopathy among 128 HIV-perinatally infected children were studied at multiple US centers by the Women and Infants Transmission Study Group. During a median follow-up of 24 months, HIV encephalopathy was diagnosed in 27 (21%), with a median survival of 14 months and a mortality rate of 41%. In the encephalopathy cases, immunosuppression was present in 20 (74%), hepatosplenomegaly or

lymphadenopathy in 63%, and cardiomyopathy in 30%. Failure to gain weight predated the onset of encephalopathy in infected infants. HIV infected children without encephalopathy had a lower incidence of hepatosplenomegaly (29%) and cardiomyopathy (2%), diagnosed in the first 3 months of life. Risk of encephalopathy was related to a high viral load in infancy. (Cooper ER, Hanson C, Diaz C et al. Encephalopathy and progression of human immunodeficiency virus disease in a cohort of children with perinatally acquired human immunodeficiency virus infection. J Pediatr May 1998;132:808-812). (Reprints: Ellen R Cooper MD, Finland Laboratory, Boston Medical Center, 774 Albany St, Boston, MA 02118).

COMMENT. Encephalopathy is a frequent complication of perinatally acquired HIV infection, occurring in one in 5 infants. A high viral load during the neonatal period, failure to thrive, and early signs of organomegaly and lymphadenopathy are risk factors for HIV encephalopathy.

NEUROLOGIC COMPLICATIONS OF E. COLI H-UREMIC SYNDROME

The association between bacterial genotype of E coli O157:H7 and CNS manifestations of childhood gastroenteritis-associated hemolytic uremic syndrome (D+HUS) was studied in 51 patients with HUS treated at the British Columbia's Children's Hospital, Vancouver, Canada. Of 51 children with HUS, 11 suffered neurologic complications that included encephalopathy in 6 and seizures in 7, with 2 deaths. No association with bacterial genotype was demonstrated. (Cimolai N, Carter JE. Bacterial genotype and neurological complications of *Escherichia coli* O157:H7-associated haemolytic uraemic syndrome. Acta Paediatr May 1998;87:593-594). (Respond: Dr N Cimolae, Room 2G6, Department of Pathology and Laboratory Medicine, British Columbia's Children's Hospital, 4480 Oak Street, Vancouver, British Columbia, Canada).

COMMENT: Seizures, encephalopathy, visual disturbances, and transient hemiparesis are the neurologic complications of E coli hemolytic uremic syndrome. Seizures, the most common symptom, occur from 4 to 12 days after onset of diarrhea. A number of factors may cause the CNS symptoms, but a specific bacterial genotype of E coli is not implicated. Prevention or timely diagnosis and treatment may diminish the incidence of neurologic complications of E coli O157:H7 infection.

Risk of hemolytic uremic syndrome after sporadic E coli O157:H7 infection: Results of a Canadian collaborative study. (Rowe PC et al. J Pediatr May 1998;132:777-782). Of 205 children with HUS, 77% had E coli O157:H7 infection. A further 582 had E coli gastroenteritis. The risk of HUS in Alberta was 8.1%, compared to 31% in other Canadian referral centers.

SEIZURE DISORDERS

COGNITIVE AND BEHAVIORAL OUTCOMES OF FEBRILE SEIZURES

Of approximately 14,000 children enrolled in a British Child Health and Education Study, 398 identified with febrile convulsions (FC) were assessed at age 10 years at Addenbrooke's Hospital, Cambridge, and the University of Bristol, UK. Measures of academic progress, intelligence, and behavior in the FC patients were not significantly different from controls without FC. Patients with simple FC (287) and complex FC (94) showed similar results. The outcomes in those with recurrent FC and those with a single seizure were similar. Children with FCs in the first year of life required special schooling more often than those with late-onset FCs (7.5%