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