progressed rapidly during the growth period, reaching a 60° magnitude by 17 yrs. Those with scoliosis >60° were bedridden and had thoracolumbar curves. Risk factors for progression of scoliosis in spastic cerebral palsy are: a spinal curve of 40° before age 15 yrs; total body involvement; being bedridden; a thoracolumbar curve. Early surgical intervention to prevent progression is recommended in patients with these risk factors. (Saito N, Ebara S, Ohotsuka K, Kumeta H, Takaoka K. Natural history of scoliosisi in spastic cerebral palsy. Lancet June 6, 1998;351:1687-1692). (Respond: Dr Naoto Saito, Dept of Orthopaedic Surgery, Shinshu University School of Medicine, Asahi 3-1-1, Matsumoto 3900-8621, Japan).

COMMENT. Reports indicate that bracing is frequently ineffective for scoliosis with cerebral palsy and surgery is advocated in severe cases. Fusion is intended to facilitate sitting and standing, improve pulmonary and upper limb function, and benefit nursing procedures. The above study provides guidelines for early surgical intervention.

METABOLIC DISORDERS

FOCAL NEUROLOGIC DEFICITS WITH HYPOGLYCEMIA

Clinical manifestations and outcome of transient focal neurologic deficits (TFND) associated with hypoglycemia were evaluated in 44 children with insulin-dependent diabetes mellitus observed retrospectively during a 5 year period at the University of Trieste, Italy. Symptoms included transient hemiparesis in 54 episodes, usually during sleep, and alternating right- and left-sided in 3; aphasia in 16 episodes; and preceded by a brief convulsion in 8 episodes. Duration of episodes was <2 hrs in 30, 2-12 hrs in 13, and >12 hrs in 2. Hypoglycemia was documented in 26, and in 18 of these episodes, symptoms did not resolve promptly after sugar administration. The long-term outcome was benign, no patient having persistent neurologic abnormalities and none developing migraine at follow up. Invasive tests were not considered mandatory. (Pocecco M, Ronfani L, and Italian Collaborative Paediatric Diabetologic Group. Transient focal neurologic deficits asociated with hypoglycemia in children with insulin-dependent diabetes mellitus. Acta Paediatr May 1998;87:542-544). (Respond: Dr M Pocecco, Department of Paediatrics, Children's Hospital "Burlo Garofolo," University of Trieste, Italy).

COMMENT: Transient focal neurologic deficits in children with diabetes are often but not invariably asociated with hypoglycemia and their long-term outcome is good. Alternative causes for an episodic hemiparesis in diabetes include vascular spasm, hemiplegic migraine, and Todd's paresis.

COGNITIVE, MOTOR, AND BEHAVIORAL FUNCTION IN PKU

Measures of cognitive, frontal lobe (executive), behavioral and motor function were administered to 18 children (aged 12-101 months) with phenylketonuria followed at the University of Rochester School of Medicine, NY. Current phenylalanine levels were within recommended range (120-485 mcmol/l) in 65%, and lifetime levels ranged from 206-1331 (mean, 499). "Individual variation" (SD of lifetime level) ranged from 76-547 (av, 270). Lower current phenylalanine (PHA) levels were associated with higher cognitive functioning in children older than 3 yrs. Higher current and average levels correlated with more difficult temperament on behavior scales. Motor function was impaired in PKU children with current PHA levels above 360 mcmol/l, and lower motor scores tended to correlate with older age and current PHA levels.

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