

VISUAL EVOKED POTENTIALS IN PHENYLKETONURIA

Visual evoked potentials (VEPs) were studied in 36 patients with phenylketonuria, and compared with MRI findings, dietary state, and IQ, at the Institute of Neurology, Queen Square, and the Institute of Child Health, Great Ormond Street Hospital, London, UK. Four patients presented with progressive neurological deficits. Twenty six were detected by routine neonatal screening, and 9 were diagnosed with developmental delay at age 5 years. All except one showing no symptoms had received a low phenylalanine diet until age 7 years or later. In 9 patients aged less than 14 years who were still on the diet, VEPs were normal in 8 and the MRI was abnormal in all but only mildly irregular in 6. The one patient with abnormal VEPs had the most pronounced MRI abnormalities and the lowest IQ score (74 compared to 90-119 in the remaining 8). Of 27 patients aged 14-31 years, >80% had abnormal VEPs, showing reduction of amplitude and prolonged latencies, whereas neuro-ophthalmological examination was normal. VEP amplitude was inversely correlated with MRI abnormalities and severity of white matter lesions in the parieto-occipital region, and with IQ. VEP abnormalities were marginally correlated with plasma phenylalanine concentrations but were not dependent on a sustained low phenylalanine diet. (Jones SJ et al. Visual evoked potentials in phenylketonuria: association with brain MRI, dietary state, and IQ. J Neurol Neurosurg Psychiatry September 1995;59:260-265). (Respond: Dr SJ Jones, Department of Clinical Neurophysiology, National Hospital for Neurology and Neurosurgery, Queen Square, London WC1N 2BG, UK).

COMMENT. This study confirms reports of the frequency of abnormal VEPs in patients with phenylketonuria, even in those diagnosed and treated early. VEP decreased amplitude and prolonged latency are correlated to some extent with subcortical myelin defects revealed by MRI. A striking increase in incidence of VEP and MRI abnormalities in older patients coincided with relaxed dietary management, plasma phenylalanine concentrations, and intellectual performance.

TOXIC DISORDERS

MRI IN KERNICTERUS

The magnetic resonance images (MRI) of three children with athetotic cerebral palsy and severe neonatal jaundice were examined in the Department of Pediatric Neurology, Ohzora-no-ye Hospital and Seirei-Mikatahara General Hospital, Shizuoka, Japan. High intensity areas in the posteromedial border of the globus pallidus on T2-weighted images were found bilaterally in all 3 children. No abnormalities were demonstrated on T1-weighted imaging. (Yokochi K. Magnetic resonance imaging in children with kernicterus. Acta Paediatr August 1995;84:937-9). (Respond: Dr K Yokochi, Ohzora-no-ye Hospital, 7448 Nakagawa, Hosoe, Inasa, Shizuoka 431-13, Japan).

COMMENT. Kernicteric encephalopathy is a rare neonatal disorder since the introduction of phototherapy. Autopsy findings have revealed bilirubin staining of the globus pallidus, subthalamic nucleus, hippocampus, and dentate and olivary nuclei. The posteromedial border of the globus pallidus is the most sensitive region to kernicterus in MR imaging. Perinatal hypoxic-ischemic encephalopathy is distinguished by involvement of the putamen and thalamus on pathological and MR

studies. The author lists other diseases with MR lesions in the globus pallidus including Leigh syndrome, Hallervorden-Spatz disease, hemolytic uremic syndrome (associated with E coli 0157:H7 and *Shigella dysenteriae* food poisoning), carbon monoxide intoxication, hepatic encephalopathy, and neurofibromatosis. See Progress in Pediatric Neurology II (PNB Publishers, 1994, pp242-3) for a previous article by the same author and commentary on MRI in 22 athetotic cerebral palsied children. The value of the MRI in the timing of basal ganglia pathology has been alluded to in other reports of dyskinetic and dystonic cerebral palsy (*ibidem*. pp243-4). Of 219 dyskinetic CP cases seen between 1955 and 1986 in the Cheyne CP Centre, Chelsea, London, 25% had been diagnosed with kernicterus.

VITAMIN A SUPPLEMENTS AND BULGING FONTANELLE

Safety of vitamin A supplements in early infancy was investigated by double-blind, randomized, placebo-controlled trial in 167 infants in the Urban Surveillance System area of the International Centre for Diarrhoeal Research, Bangladesh. Three doses of 25000 IU of vitamin A or placebo were given at 6, 12 and 17 weeks of age, and infants were examined by physicians on days 1, 2, 3 and 8 after supplementation. Bulging fontanelle occurred in 9 (10.5%) infants receiving vitamin A compared to 2 (2.5%) in the placebo group ($p < 0.05$). The side effect was not observed after the first dose, 3 infants were affected after the second supplement, and 9 after the third. A cumulative effect of vitamin A was likely. (Baqui AH et al. Bulging fontanelle after supplementation with 25000 IU of vitamin A in infancy using immunization contacts. Acta Paediatr August 1995;84:863-6). (Respond: Dr AH Baqui, Urban MCH-FP Extension Project, ICDDR,B, GPO Box 128, Dhaka 1000, Bangladesh).

COMMENT. The infants in this study received vitamin A supplements together with the routine DPT/OPV immunization. Bulging of the fontanelle has been reported in the US as a side effect of immunization with DTP vaccine and DT vaccine. (Gross TP et al. J Pediatr 1989;114:423-5 [cited in above study]). An additive or synergistic effect of the immunization cannot be excluded. The reliability of the clinical assessment of the fontanelle by observation and palpation is also debated, and a probable underestimation of vitamin A toxicity is suggested by the authors.

With present day enthusiasm for supplemental vitamins and a common attitude of nonchalance toward possible vitamin overdosage, the recognition of early symptoms and signs of vitamin toxicity is important. (Millichap JG. Environmental Poisons in Our Food, Chicago, PNB Publishers, 1993).

HEADACHE DISORDERS

BRAIN IMAGING INDICATIONS FOR HEADACHES

Charts of all children referred to the pediatric neurology clinic, Schneider Children's Hospital, New Hyde Park, NY, for evaluation of headaches over a 2-year period were reviewed retrospectively for headache characteristics, indications for performing CT and MRI studies, and imaging results. Of 133 patients ages 3 to 18 years, 52% had migrainous headaches, 21% chronic tension headaches, and 19% were unclassified. The indications for