

histological confirmation is not justified, (3) surgical removal, successful in 10 cases, is reasonably safe, (4) cranio-spinal axis radiation, either post-operatively or alone, is indicated for germinomas that have a tendency for CSF ependymal proliferation and/or seeding, (5) all children treated successfully by both surgery and radiation have not relapsed. (Schulte FJ et al. EUR J Pediatr 1987;146:233-245).

**COMMENT:** Pineal tumours are of 2 main types: (1) germinomas of the pineal or suprasellar region, and (2) pineal parenchymatous tumours (pineoblastomas in younger children and pineocytomas in older children and adults). Spinal seeding, more common with germinomas, occurs in 2-15% of pineal tumours.

Parinaud's syndrome (impairments of upward gaze, convergence and accommodation) is the classical presentation, usually with signs of increased intracranial pressure. Visual loss, diabetes insipidus, precocious puberty, and emaciation point to anterior hypothalamic involvement. Precocious puberty only occurs in males. Suprasellar teratomas present with visual loss, hypopituitarism, and diabetes insipidus.

Calcification occurs in 25-75% of pineal area tumours, rarely in children under 10 years of age, and especially in germinomas and embryonal carcinomas. CT scan demonstrates location but may not differentiate type of tumour. Oncofetal antigens (human chorionic gonadotrophin and alpha-fetoprotein) in serum and spinal fluid are helpful in diagnosis and response to therapy.

Operative mortality and morbidity in the past have been high. Shunt surgery and/or biopsy followed by radiotherapy has a 50-80% survival rate. The present authors make a point for surgical resection and post-operative radiation. (For further references see (1) Cohen ME, Duffner PK. Brain Tumours in Children, Raven Press, NY, 1984; (2) Sano K, Matsutani M, Child's Brain, 1981; 8:81-97; (3) Raimondi AJ, Tomita T. Child's Brain 1982;9:239-266).

#### DEGENERATIVE DISORDERS

##### ALPERS' PROGRESSIVE NEURONAL DEGENERATION

Reporting from the Hospital for Sick Children, Great Ormond Street, London WC1, the authors have selected 13 cases, 10 boys and 3 girls, with progressive neuronal degeneration of childhood (PNDC) that was complicated by liver disease and confirmed at post mortem in 11. During life, PNDC may be suspected by a characteristic clinical course, abnormal liver function tests, and abnormalities of EEG (grossly asymmetric, very slow activity of high amplitude mixed with polyspikes), VER, and CAT (cortical and central atrophy and areas of low density of the white matter). It is proposed that the term PNDC be reserved for a distinct syndrome characterized by normal initial development followed by developmental retardation and later onset of intractable seizures and liver degeneration, and by autosomal recessive inheritance.

Four patients received sodium valproate; 2 may have died from valproate toxicity although both had abnormal liver enzymes prior to treatment. Phenytoin was probably blameless; 8 patients never received it and the liver pathology of fatty degeneration, necrosis, and cirrhosis was not that expected in phenytoin toxicity.

Brain pathology revealed cortical atrophy with predilection for

the calcarine cortex, astrocytic proliferation, and spongy degeneration also involving the thalamus, basal ganglia, and brainstem. Hippocampal sclerosis and cerebellar infarcts resembled epileptic anoxic changes in some patients. (Egger J, Harding BN, Boyd SG, Wilson J, Erdohazi M. Clinical Pediatrics 1987; 26:167-173).

**COMMENT:** The syndrome of diffuse progressive degeneration of the cerebral gray matter was first described by Alpers in 1931. Ford (1951) differentiated infantile and juvenile types and reported familial cases. Huttenlocher et al (1976) emphasized a coincident hepatic cirrhosis. The cause is unknown. The cerebral pathology resembles anoxic encephalopathy secondary to status epilepticus in some reported cases and the liver disease might be the result of anticonvulsant toxicity, notably sodium valproate. In the author's cases, however, these causative factors were not generally accepted as primary, and a genetically determined metabolic explanation was preferred.

#### HEADACHE AND RELATED DISORDERS

##### DIET AND MIGRAINE

A team of investigators at the Department of Paediatrics, Rotherham District General Hospital, and Sheffield Children's Hospital, have carried out a controlled study in 39 children to assess the effects of exclusion of dietary vasoactive amines in migraine. The children were allocated at random to either a high fibre diet low in these substances or a regular high fibre diet for an 8 week period. Foods excluded were chocolate, cheese, yogurt, citrus fruits, bananas, pineapple, raspberries, plums, peas, beans, yeast, shellfish, smoked pickled fish, game, tea, coffee, and cola drinks containing caffeine.

Both test and control groups showed a significant decrease in the number of headaches and there was no significant difference between the two groups. A placebo effect was considered a probable explanation for the improvement in many. (Salfield SAW et al. Arch. Dis. Child. 1987; 62:458-460).

**COMMENT:** The relation of tyramine and other amine-rich foods to the occurrence of headaches in certain migraineurs is a theory frequently proposed (Hanington E. In Clinical Reaction to Food. New York, Wiley, 1983). The authors of the present study admit that their group was small and an idiosyncrasy to amines in occasional patients with migraine could not be ruled out.

An allergic mechanism for dietary migraine is an alternative theory investigated by use of a so-called "oligoantigenic diet", a diet that eliminates all but a few sensitizing food antigens. Cow's milk, egg, and wheat cereals were the most frequent offenders (Egger J et al. Lancet 1983; 2:865). To strictly avoid all foods listed as possible migraine precipitants is usually unnecessary and possibly hazardous to the child's health. If the possible benefits of an elimination diet are to be confirmed, however, the use of a control diet would be essential to exclude a placebo effect.

##### SOMNAMBULISM AND MIGRAINE

Neurologists in the EEG laboratory of the Hôpital d'Enfants,