disease. Headache July/Aug 1999;39:486-489).

Migraine precipitating factors. Of 226 children with headaches, 148 (65%) had migraine without aura, 43 (19%) migraine with aura, 25 (11%) daily headache, and 16 (4%) had tension/migraine. Precipitating factors were elicited in 206 children (91%); 141 (62%) had one factor and 53 (29%) had two or more. In decreasing frequency, these included stress (23%), sleep deprivation (16%), hunger (11%), heat (11%), bright light (9%), exercise (8%), foods (7%), motion (6%), and medications (5%); MSG and caffeine, odors, and reading were precipitants in less than 5% each. Analgesic rebound occurred in 36% of the daily headache group compared to 1.5% of other headache patients. Environmental manipulations and avoidance of precipitating factors may obviate the need for daily medications in the treatment of childhood migraine. (Riback PS. Factors precipitating migraine headaches in children. Ann Neurol Sept 1999;46:540, abstract). In my own experience and that of colleagues in the UK, specific foods are more frequent precipitants of headache (see Progress in Pediatric Neurology I and II, PNB Publ, 1991;pp542-3, 1994;pp166-168).

Mast cell activation in migraine. Mean levels of urinary histamine, its metabolite, methylhistamine, and the mast cell enzyme, tryptase, were higher in children than in adults. In 8 of 10 children who successfully practiced relaxation imagery techniques for migraine, urine levels of tryptase were significantly lower than controls. Stress may activate mast cells that could be involved in the pathophysiology of migraine. (Olness K, Hall H, Rozniecki JJ, Schmidt W, Theoharides TC. Mast cell activation in children with migraine before and after training in self-regulation. Headache Feb 1999;39:101-107).

Intranasal sumatriptan for acute migraine. A randomized double-blind placebo-controlled crossover study showed that 12 of 14 patients (6-10 years of age) reported a decrease in pain intensity after sumatriptan versus 6 of 14 after placebo (p=0.031), and complete relief was obtained in 9 of 14 vs 2 of 14 (p=0.016). (Ueberall MA, Wenzel D. Intranasal sumatriptan for the acute treatment of migraine in children. Neurology April 1999;52:1507-1510).

HEREDO-DEGENERATIVE DISORDERS

SCREENING FOR SPHINGOLIPID-STORAGE DISEASES

A simple fluorescence assay was used to detect cells from patients with sphingolipidoses in a masked study at the Mayo Clinic, Rochester, MN. Replicate samples of 25 of 26 unique cell lines representing ten different lipid-storage diseases (Fabry's disease, gangliosidoses (Tay-Sachs and Sandhoff forms), metachromatic leukodystrophy, mucolipidosis type IV, Niemann-Pick disease (types A, B, and C), and sphingolipid-activator-protein-precursor (prosaposin) deficiency), and 18 of 20 unique cell lines representing controls were correctly identified. The sensitivity was 96.2% and the specificity 90%.

The artificial fluorescent lipid, a derivative of the natural sphingolipid, lactosylceramide, accumulated in the lysosomes of cultured fibroblasts from patients with sphingolipidoses. It was mainly confined to the Golgi complex in normal control cells and cells from patients with other types of lysosomal diseases. With increasing concentrations in the membranes of the lysosomes, the emission of the fluorescent lipid changed from green to red wavelengths. The method should be useful as an initial general screen for lipid-storage diseases,

and it may also allow screening of candidate drugs to treat the lipidoses. (Chen C-S, Patterson MC, Wheatley CL, O'Brien JF, Pagano RE. Broad screening test for sphingolipid-storage diseases. Lancet Sept 11, 1999;354:901-905). (Respond: Dr Richard E Pagano, Mayo Clinic and Foundation, Guggenheim 621-C, 200 First Street SW, Rochester, MN 55905).

COMMENT. This simple fluorescence assay should be helpful in the investigation of children with developmental disorders and progressive neurodegenerative diseases and the exclusion of sphingolipid-storage diseases in patients with atypical or mild variants.

In a commentary by Bryan Winchester, Institute of Child Health, London (Lancet Sept 11, 1999;354:879-880), the method is described as potentially an important advance. He suggests that adaptation of the method to allow measurement of the fluorescence using microtitre-plate technology in whole cells, preferably white blood cells, would simplify the procedure and avoid the delay and cost of culturing fibroblasts.

AUTOSOMAL DOMINANT JUVENILE AMYOTROPHIC LS

The clinical and electrodiagnostic findings in 49 affected family members and neuropathological findings from two autopsies of a Maryland kindred with autosomal dominant juvenile amyotrophic lateral sclerosis (ALS) are reported from Johns Hopkins University and Hospital, Baltimore, MD. The ALS was linked to the chromosome 9q34 region (ALS4). The mean age at onset was 17 years, and patients ranged in age from 12 to 85 years (mean 45 years). Clinically, the majority showed distal weakness and atrophy associated with pyramidal signs and normal sensation. Electrodiagnostic testing in 8 patients showed reduced evoked amplitudes and normal motor and sensory conduction. EMG showed distal chronic partial denervation and reinnervation. Spinal cord tissue was atrophic with loss of anterior horn cells, degeneration of corticospinal tracts, loss of neurons in the dorsal root ganglia, and degeneration of the posterior columns. Motor and sensory roots and peripheral nerves showed axonal loss and diffuse prominent swellings. This family extends the genetic heterogeneity of familial and juvenile ALS. (Rabin BA, Griffin JW, Crain BJ, Scavina M, Chance PF, Cornblath DR. Autosomal dominant juvenile amyotrophic lateral sclerosis. Brain Aug 1999;122:1539-1550). (Respond: Dr David R Cornblath, Pathology 627, Johns Hopkins Hospital, Baltimore, MD 21287).

COMMENT. Juvenile ALS is a chronic motor neuron disease with onset before 25 years and characterized by upper and lower motor neuron dysfunction in the absence of sensory abnormalities or ataxia. A family is described with a slowly progressive, non-fatal, autosomal dominant form of juvenile ALS linked to the chromosome 9q34 (ALS4).

SEIZURE DISORDERS

BENIGN PARTIAL SEIZURES OF ADOLESCENCE

Eight patients matching the description of benign partial seizures of adolescence (BPSA), as described by Loiseau et al in 1978, were found among 92 teenagers, including 37 with new-onset focal seizures, enrolled in a prospective first-seizure study at the University of Melbourne, Victoria, Australia. Four of the 8 patients with BPSA were boys and 4 girls, aged 11-17 years. All seizures were Jacksonian in pattern, with a sensory/motor march, 6 were secondarily