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