Pelizaeus–Merzbacher disease (PMD): Intronic mutations
PMD is caused by mutations in the proteolipid protein gene (*PLP*). Hobson et al. (p. 1089) identified mutations in *non-coding* regions of the *PLP* gene. This discovery accounts for the previous inability to detect *PLP* mutations in 10 to 15% of patients with PMD. It may also explain some of the variations in the PMD phenotype. ◆ The accompanying editorial by Percy (p. 1072) summarizes previous work on the genetics of PMD and notes that the phenotypic heterogeneity of this disorder of CNS myelin protein is analogous to the various lesions in the single gene responsible for Charcot–Marie–Tooth disease 1A and hereditary neuropathy with liability to pressure palsies.

Benign childhood occipital epilepsy (Panayiotopoulos)
Caraballo et al. (p. 1096) studied a large Argentinian population of children with epilepsy and identified 66 children with the benign occipital epilepsy–Panayiotopoulos syndrome (PS) versus 145 with benign centrotemporal rolandic epilepsy. PS is defined clinically as a benign partial epilepsy with brief seizures consisting of ictal vomiting and eye deviation progressing to loss of consciousness and generalized seizures. The majority of seizures occur at night. ◆ The accompanying editorial by Berg and Panayiotopoulos (p. 1073) notes the consistency of studies in reporting PS to be a distinct syndrome and discusses possible reasons for divergent incidence figures reported in different populations.

Glutamate transporter splice variants: Not specific to ALS
A working hypothesis for the pathogenesis of ALS is glutamate toxicity to motor neurons. Decreased transport of glutamate and, recently, abnormal splice variants for the glutamate transporter EAAT2 have supported the hypothesis. The Expedited Publication by Honig et al. (p. 1082) reports that splice variants occur in AD and even controls, suggesting that motor neuron degeneration may not be related to abnormalities of the glutamate transporters.

Neuropathy in restless legs syndrome (RLS)?
Evidence for CNS abnormalities in RLS was the subject of a recent *Neurology* editorial (1999; 52:907) and also a number of papers. Polydefkis et al. (p. 1115), having noted that many of the patients they saw with small fiber neuropathy had symptoms of RLS, studied 22 patients with RLS. They report abnormalities of large motor and small sensory nerves (epidermal biopsy) in 36% of patients. This group showing evidence of neuropathy differed from the other patients with RLS by not having a positive family history and by their older age. The authors postulate that there may be two distinct forms of RLS.

Controlled trial of IVIg in MS
Noseworthy et al. (p. 1135) randomized 67 patients with muscle weakness due to MS to intensive IVIg or placebo for 3 months of treatment. There was no benefit to strength or in follow-up to relapses or functional impairment. The isometric muscle strength measurement used performed extremely well as an objective, quantitative test.

Exposure to organolead: A risk factor for progressive decline in cognition?
Schwartz et al. (p. 1144) compared 535 former organolead workers with controls. The workers (mean age 56 years) were an average of 16 years post-lead exposure. Tibial lead levels measured radiologically provided evidence of increased bone lead. Serial tests showed that lead workers had abnormal decline in visuo-constructive ability and verbal memory. Bone lead accumulation correlated with measures of decline.

Head injury and risk of subsequent AD
Plassman et al. (p. 1158) compared 548 male military veterans hospitalized with closed head injuries with 1228 who had no head injury. Both moderate and severe head injuries were associated with increased risk of AD. There tended to be a relationship between the number of *APOE* $\epsilon 4$ alleles and AD in head-injured men.

Vagus nerve stimulation (VNS) for suppression of pain
Kirchner et al. (p. 1167) studied experimental pain stimuli in 10 seizure patients with implanted VNS. VNS reduced pain from electrical stimuli and from tonic pressure without changing perception of tonic or of heat-induced pain. This reduction occurred both during and after acute VNS. VNS deserves study for pain treatment.

Boys with Rett syndrome
Villard et al. (p. 1188) describe two boys who died from an unexplained, severe neonatal encephalopathy and whose sister has classic Rett syndrome. A mutation in the *MECP2* gene was found in the sister and in DNA from one of the boys. The patients' mother also carried the mutation but did not have Rett syndrome because of skewed X-chromosome inactivation.

