December 26 Highlights

Mortality in PD: L-Dopa or selegiline?
In a longitudinal study, Donnan et al. (p. 1785) compared mortality of patients with PD with that of controls, examining the effect of L-dopa and of selegiline. L-Dopa but not selegiline was associated with excess mortality. ◆ In the accompanying editorial, Langston and Tanner (p. 1770) consider this new information in light of the sometimes acrimonious debate as to whether selegiline is of preventive or only symptomatic benefit in PD. Regardless of that debate, this new study argues for the safety of selegiline—incriminated in mortality in earlier reports.

Spontaneous intracranial hypotension (SIH)
Alvarez–Linera et al. (p. 1895) report 11 patients with SIH who had prominent pituitary enlargement by MRI. The enlargement suggested pituitary tumor and was found to resolve in successfully treated patients. *Neurology* has had two other recent papers on SIH: *Neurology* 2000;55: 1321–1327 and *Neurology* 2000;55: 573–575. ◆ The accompanying editorial by Mokri and Posner (p. 1771) discusses the protean clinical and radiologic features of SIH, notes that its cause is almost always a spontaneous dual tear, and points out that treatment is usually successful in curing the syndrome.

Hereditary spastic paraplegia (HSP): The phenotype of SPAST mutations
Of the at least seven genes for autosomal dominant HSP, mutation in the SPAST gene on chromosome 2p (SPG4) is the most common cause. McMonagle et al. (p. 1794) studied 29 patients with SPAST mutations versus 22 patients in whom SPG4 was excluded by linkage analysis. Patients with SPG4 were clinically distinct from other HSP: more cognitive impairment, more rapidly progressive, but of later onset.

IV r-tPA: ER physicians versus neurologists
Akins et al. (p. 1801) note that in many centers neurologists are not readily available to treat patients with acute stroke, but that many ER physicians have had concerns about the generalizability of the NINDS treatment trials. They studied a small series of patients in a community setting treated by ER physicians. The outcomes were similar whether a neurologist or ER physician was the prescribing physician. However, protocol violations were only 5% for neurologists versus 30% for ER physicians.

More generalized seizures after successful epilepsy surgery?
Henry et al. (p. 1812) tailored a scale to assess the frequency of generalized seizures and showed that despite major reduction in number of seizures, those seizures that occurred were more likely to be generalized.

Limbic encephalitis without neoplasm: Temporal lobe epilepsy (TLE)
Bien et al. (p. 1823) describe four young adults who presented with intractable TLE, in whom there was MRI and pathology evidence for encephalitis. There was no evidence for a paraneoplastic syndrome.

Pupils: Ross syndrome—Mydriasis in carotid dissection
Shin et al. (p. 1841) describe five patients with the triad of segmental anhidrosis, hyporeflexia, and tonic pupils: Ross syndrome. Four had Horner syndrome. They provide pharmacologic evidence that Ross syndrome is a dysautonomic ganglion cell disorder and contrast the clinical features and pharmacologic testing of Ross syndrome with other pupillary disorders. ◆ Inzelberg et al. (p. 1934) report a 68-year-old woman who presented with transient right eye mydriasis as the sole neurologic finding. The finding resolved but the patient died of carotid and aortic dissection a few hours later.

AD: Cognitive tests that discriminate from normal
In a prospective community study, Chen et al. (p. 1847) retested 120 nondemented subjects who subsequently manifested AD and compared them with 483 controls who remained nondemented 10 years later. The better tests, most predictive of AD, were memory (delayed recall) and executive dysfunction.

Gelastic epilepsy: response to gonadotropin releasing hormone (GnRH)
Zaatreh et al. (p. 1908) describe two patients with gelastic epilepsy from hypothalamic hamartomas, in whom treatment of precocious puberty with GnRH was associated with cessation of seizures.

Leg fidgets from hypotension
Cheshire (p. 1923) describes six patients with autonomic failure in whom a distinctive syndrome of akathisia was evident while patients were sitting. Leg movements raised the blood pressure and presumably compensated for autonomic failure.

Dyferlinopathy: Identical mutation, different phenotypes
Why most myopathies are proximal and a few distal, as well as the peculiar distributions that gave rise to the names of individual muscular dystrophies, remains a mystery. Here, Illarioshkin et al. (p. 1931) describe 12 patients in a large Russian kindred with dysferlin mutations. Nine patients had a limb-girdle phenotype, one a distal (Miyoshi myopathy) phenotype, and two a distal myopathy involving both anterior and posterior compartments. Because sibships had the same phenotype an additional genetic factor seems likely.

