Convective brain perfusion for Gaucher disease

Lonser et al. evaluated convection-enhanced delivery of glucocerebrosidase co-infused with a surrogate MRI tracer (gadolinium-DTPA) in the CNS. The authors used this delivery paradigm to perfuse targeted brain and brain-stem sites in a patient with neuronopathic Gaucher disease during MRI enzyme distribution in real-time.

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Can patients with epilepsy predict their seizures?

Many patients report that they can identify periods of increased risk for seizure. Haut et al. assessed seizure self-prediction in a prospective seizure diary study. Twenty percent of their subjects successfully self-predicted seizures; a positive prediction was associated with a 3.1-fold increase in risk of a seizure.

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“Pseudoseizure” dogs

Krauss et al. found that four of their six patients with seizure-assist dogs had psychogenic nonepileptic seizures rather than epilepsy. Seizure-assist dogs may provide companionship and help patients during seizures; however, this series shows the importance of accurately diagnosing epilepsy in patients prior to their obtaining specially trained service animals.

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Seizure alert dogs

Doherty and Haltiner present a patient with psychogenic nonepileptiform events who claimed her dog alerted her to her impending seizure. Seizure alert dog studies are critically reviewed in light of possible data contamination with nonepileptic patients.

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The editorial by Litt and Krieger asks: Why the focus on predicting seizures? The most problematic aspect of epilepsy, according to patients in one large survey, is uncertainty about if and when a clinical event may occur. Identifying periods of time when the probability of seizures is increased also offers patients an opportunity to avoid dangerous situations or take preventive medications. There are at least three sources of bias in the Haut et al. study: selection bias, nonresponse bias, and response bias. Because data in this study are self-reported, willingness of subjects to please investigators might have led to reports that improperly demonstrated prediction. Both studies of seizure-alert dogs suggest a potential therapeutic effect in owning such dogs, but that the benefit is more likely to be psychological than neurologic. In both studies it was only objective validation of patient-reported events that led the appropriate diagnosis.

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STN stimulation in early stage PD

Motor examination score (UPDRS III) in PD at inclusion (MO), and after 6, 12, and 18 months of medical treatment (left) or bilateral STN stimulation (right). Black bars: “off” levodopa; open bars: “on” levodopa.

Schüpbach et al. randomized in matched pairs 20 young patients with mild to moderate PD to best medical treatment or subthalamic nucleus stimulation. After 18 months, quality of life, motor signs and complications, and activities of daily living were significantly improved among patients with deep brain stimulation.

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The editorial by Riley and Lozano notes that there is a trend toward early DBS in PD management, and this trial offers the best support of that trend to date. Here an attempt was made to capture a group of young, active patients with PD before their livelihood was compromised. Despite the small sample size, the results significantly favored DBS over medical therapy alone in measures of quality of life, motor performance off medication, and medication dosage reduction. As a result, the severity of both fluctuations and dyskinesias also improved substantially. Currently only a minor fraction of patients with PD who would benefit from DBS experience this treatment. While neurologists and neurosurgeons are working to improve this ratio, this article actually enlarges the denominator. While many questions remain concerning patient selection, earlier application of DBS is likely to improve our approach to management of many patients with PD.

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Driving T-cell activation in inflammatory neuropathies

Hu et al. studied the CD28-related costimulatory molecule ICOS and its ligand (ICOSL), a potent driving pathway of T-cell activation, in inflammatory neuropathies. While absent in non-inflammatory controls, ICOS and its ligand were strongly upregulated in acute as well as chronic inflammatory neuropathies.

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Transdermal rotigotine in early PD

This randomized, controlled study by Watts et al. compared once-daily transdermal rotigotine vs placebo in 297 patients with early PD over 6 months. The rotigotine group had a significant reduction of UPDRS motor and activity of daily living subscores compared with placebo ($p < 0.0001$), and adverse events were mostly mild to moderate.

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Conversion from subtypes of mild cognitive impairment to AD

Fischer et al. compared the rates of conversion to AD between amnesic and nonamnesic MCI in a community-based birth cohort at age 75. Amnesic MCI predicted AD at a substantially higher rate than non-amnesic MCI.

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Soluble Nogo-A as a biomarker for MS

Jurewicz et al. found soluble 20 kDa Nogo-A product in 96% of CSF samples from MS patients but not from control subjects with other neurologic diseases including CNS autoimmune diseases. Nogo-A product were present both in RR-MS and SP-MS and thus has potential as a biomarker for assessing disease activity in patients with MS.

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Late recovery from the minimally conscious state

Concluding that it is ironic that despite all the attention generated by the Terri Schiavo case, little interest has been directed toward the needs of this underserved population, Fins et al. ask, "If America was confused by the Schiavo case, how will it respond to new knowledge about disorders of consciousness and the capability of the brain to recover in the face of overwhelming injury?" They consider the challenges identified by a recent Institute of Medicine meeting that considered the persistent vegetative state and the minimally conscious state. With a modest investment, future studies may well elaborate the basis for consciousness in the human brain and provide diagnostic and prognostic information that will prove helpful to clinical decision making at the bedside.

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Neurology®

January 23 Highlights

Neurology 2007;68;248-249

DOI 10.1212/01.wnl.0000255793.01489.64

This information is current as of January 22, 2007

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