Lefaucheur *et al.* recently investigated whether electrophysiological testing—which is objective and easy to perform—might have utility for showing functional decline over time in patients with HD.

Twenty patients with adult-onset HD underwent electrophysiological testing annually over a 2-year period; sympathetic skin responses (SSRs) and blink reflexes (BRs) to supraorbital nerve stimulation, long latency reflexes (LLRs) and somatosensory evoked potentials (SEPs) to median nerve stimulation, and cortical silent periods (CSPs) to transcranial magnetic stimulation were measured. Clinical evaluation was performed using the Total Functional Capacity (TFC) scale and the UHDRS motor score.

As expected, TFC score and UHDRS motor score deteriorated significantly over time ($P<0.05$ for both). BR latency, LLR presence, certain SEP parameters, and CSP duration also declined significantly over time ($P<0.05$ for all). No trend was observed in SSR amplitude and latency changes, but these parameters vary widely even among healthy individuals. Prolongation of BR latency and LLR disappearance correlated with UHDRS motor score deterioration, whereas attenuation of the peak of N20 (an SEP parameter) and CSP shortening were associated with TFC score decline.

The authors conclude that some electrophysiological parameters might be useful markers of HD progression.

Original article Lefaucheur J-P *et al.* (2006) Electrophysiological deterioration over time in patients with Huntington's disease. *Mov Disord* [doi: 10.1002/mds.20966]

Promising long-term results of zolpidem treatment for the permanent vegetative state

Zolpidem is an omega-1-specific indirect γ-aminobutyric acid (GABA) agonist used to treat insomnia; this drug has also previously been reported to improve the condition of patients with brain disorders ranging from the permanent vegetative state (PVS) to stroke-induced aphasia. A recent study of patients categorized to the PVS who were given long-term treatment with the drug has now also reported very encouraging results.

The researchers investigated the effects of a 10 mg daily dose of zolpidem in three patients. Two of the patients were aged 31 years and had been in PVS for 3 years following motor vehicle accidents; the third patient was 29 years old and had been in the condition for 5 years after almost drowning. Patients were scored on the Glasgow Coma Scale and the Rancho Los Amigos Cognitive Scale before and 1 h after drug treatment; in each patient, dramatic improvements on both scales were recorded after treatment.

At the time of the study report, the patients had been receiving zolpidem daily for between 3 and 6 years. All patients experienced daily arousal from their PVS following treatment, with dramatic improvements in their ability to interact meaningfully with others. The patients experienced maximum arousal 1 h after treatment, and the drug's effects subsided approximately 4 h later. No long-term adverse effects were observed.

The authors propose that zolpidem's mode of action could involve reactivation of dormant neurons in injured brain tissue. They suggest that the drug might have a wide application in patients with brain injury, and add that further testing is planned to assess the effects in larger groups of patients with brain damage.

Original article Clauss R and Nel W (2006) Drug induced arousal from the permanent vegetative state. *NeuroRehabilitation* **21:** 23–28

Initiation and progression phases of ALS involve *SOD1* mutations in different cell types

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that is characterized by loss of motor neurons. Most inherited ALS results from dominant mutations in the superoxide dismutase gene (*SOD1*), but the mechanisms behind neuron loss are unclear. Boillée *et al.* have used transgenic mice to investigate which cell types are damaged by mutant SOD1 and how this damage might influence the initiation and progression of the disease.

Transgenic mice carrying a mutant human *SOD1* gene, which could be deleted in specific cell types using the Cre–lox recombination system, were generated. All mice developed fatal progressive motor neuron disease, with symptoms including muscle atrophy and paralysis. Expression of mutant *SOD1* in motor neurons was shown to be a key determinant of the initiation