other measures of NPC severity was seen in patients aged ≥12 years receiving miglustat. All patients experienced at least one adverse event, but the adverse effects seen with miglustat were consistent with those seen in trials of the agent in type I Gaucher disease and were considered acceptable. The authors conclude that miglustat is well tolerated and improves or stabilizes symptoms of NPC.

Original article Patterson MC *et al.* (2007) Miglustat for treatment of Niemann-Pick C disease: a randomised controlled study. *Lancet Neurol* **6:** 765–772

Deleting redox modifier gene slows disease progression in ALS mouse model

Although the exact mechanism of neurodegeneration in amyotrophic lateral sclerosis (ALS) is not known, redox stress has been associated with disease progression in mouse models of the disease. In a recent study, Marden *et al*. examined the effect of NADPH oxidase gene deletions on disease progression in transgenic mice overexpressing a mutant form of human superoxide dismutase 1 ($SOD1^{G93A}$) that is found in patients with ALS.

The investigators demonstrated that deletion of the NADPH oxidase genes *Nox1* or *Nox2* significantly extended the life span of hemizygous $SOD1^{G93A}$ mice compared with control animals without *Nox1* or *Nox2* deletions, with the increase in 50% survival rate being markedly higher in *Nox2* knockout mice. Compared with controls, *Nox2* knockout also resulted in a significant decrease in spinal cord levels of NADPH-dependent superoxide production at the time of clinical death, and reduced the loss of motor neurons in the lumbar spinal cord. In female $SOD1^{G93A}$ mice heterozygous for the X-linked *Nox2* gene, levels of superoxide production in the spinal cord were intermediate between those of control and *Nox2* knockout mice, affording these animals approximately half the life span gain of *Nox2* knockout females.

A neuroprotective effect of reducing the activity of the Nox2 oxidase has been demonstrated in previous studies, but not to the degree seen here. Differences in genetic background of the mice studied might explain this discrepancy. Caution is needed when extrapolating the results to humans, but NADPH oxidase genes could prove to be a target for the treatment of ALS.

Original article Marden JJ *et al.* (2007) Redox modifier genes in amyotrophic lateral sclerosis in mice. *J Clin Invest* **117:** 2913–2919

A fast, noninvasive screening procedure for patients with functional brain disorders

In a 2006 paper, Georgopoulos *et al.* hypothesized that synchronous neural interactions observed at high temporal resolution using magnetoencephalography (MEG) could have utility as functional biomarkers of brain disorders. They have now tested this idea by comparing normal brain activity with that of patients diagnosed with a variety of functional brain disorders including Alzheimer's disease, schizophrenia, multiple sclerosis, chronic alcoholism and Sjögren's syndrome.

The authors used MEG to record magnetic field strength from 248 axial gradiometers while subjects fixated on a spot of light for 45–60 s. Autoregressive integrative moving average (ARIMA) modeling of raw data from 52 subjects generated stationary residuals from which synchronous cross-correlations and partial correlations were calculated. Linear discriminant analysis was used to extract relevant information and classify patterns of neural activity that were consistent among patient groups, yielding a number of disease-specific predictor subsets that gave 100% correct classification considerably more often than would be expected by chance ($z = 8.78$; $P < 10^{-50}$). Cross-validation of the predictor subsets was performed in a further 46 patients who were successfully classified into their respective patient groups more than 90% of the time. Excellent (100% correct) classification was obtained in a total sample of 142 subjects. The predictive value of this method will continue to improve as more subjects and different disease groups are included in the analysis.

As a clinical application, MEG could represent a quick, simple and noninvasive alternative to behavioral examinations in the screening or differential diagnosis of functional brain disorders.

Original article Georgopoulos AP *et al.* (2007) Synchronous neural interactions assessed by magnetoencephalography: a functional biomarker for brain disorders. *J Neural Eng* **4:** 349–355