5, 15 or 45 mg/kg. Idebenone was fairly well tolerated at all doses. In an overall comparison of the treatment groups, idebenone did not significantly affect the degree of change from baseline to 6 months in International Cooperative Ataxia Rating Scale (ICARS), Friedreich's Ataxia Rating Scale (FARS) or Activities of Daily Living (ADL) scores by ANCOVA (analysis of covariance), but it did produce a (non-significant) dose-related improvement in ICARS score on the Jonckheere trend test. In a subgroup analysis that excluded patients who needed wheelchair assistance, however, the ICARS score improved significantly with administration of idebenone, and a dose-related response was indicated in the ICARS, FARS and ADL scores. The authors suggest that their study might set the parameters for a future phase III trial to confirm the potential benefits of high-dose idebenone on neurological function in young patients with FA.

Original article Di Prospero NA *et al.* (2007) Neurological effects of high-dose idebenone in patients with Friedreich's ataxia: a randomised, placebo-controlled trial. *Lancet Neurol* **6:** 878–886

Oligoclonal bands add information to MRI in predicting development of multiple sclerosis

Oligoclonal bands (OBs) seen on protein electrophoresis analyses of cerebrospinal fluid are considered to be the best biological marker for predicting whether individuals with a clinically isolated syndrome will progress to clinically definite multiple sclerosis (CDMS). Tintoré *et al*. investigated whether OB testing adds information to that provided by MRI for predicting progression to CDMS and development of disability.

This prospective study enrolled patients who had experienced a single monosymptomatic MS-like attack that suggested demyelination of the optic nerve, brainstem or spinal cord. MRI and testing for OBs were performed within 3 months of the first attack. MRI scans were evaluated for abnormalities at baseline according to the number of positive Barkhof criteria (0–4) and the number of observed lesions. Patients were considered to have progressed to CDMS if they experienced a second MS-like attack with a new neurological abnormality at least 1 month after testing.

OBs were present in 61% of patients and were independently associated with development of CDMS ($P = 0.0001$). Of the patients with no Barkhof criteria or no lesions (low-risk MRI scans), 31% and 27%, respectively, were OB-positive, and 10% and 9%, respectively, developed CDMS. Adjusted analyses showed that presence of OBs was not associated with the likelihood of a patient reaching an Expanded Disability Status Scale score ≥3.0.

The authors conclude that the presence of OBs predicts progression to CDMS but not the development of disability, and is clinically useful in patients with a normal MRI scan who would otherwise be considered to be at low risk of progression.

Original article Tintoré M *et al.* (2007) Do oligoclonal bands add information to MRI in first attacks of multiple sclerosis? *Neurology* [doi:10.1212/01.wnl.0000280576.73609.c6]

Clinical benefit of miglustat in Niemann–Pick type C disease

Agents that prevent glycolipid biosynthesis have been shown to be beneficial in individuals with lysosomal storage diseases such as Niemann–Pick type C disease (NPC). In a recent study, Patterson *et al.* evaluated the efficacy and tolerability of the glycosphingolipid-synthesis inhibitor miglustat in patients with this incurable neurodegenerative condition.

Patients aged ≥12 years with NPC were randomly allocated to 12 months of either oral miglustat 200 mg three times a day ($n = 20$) or standard symptomatic care ($n = 9$). The study also included a cohort of 12 patients aged <12 years who all received a miglustat dose adjusted to body surface area. The authors assessed all patients at 1 week after initiation of therapy and monthly thereafter for changes in the following markers of NPC severity: horizontal saccadic eye movement (HSEM) velocity, swallowing ability, auditory acuity, ambulatory ability, and cognition.

Patients receiving miglustat showed an improvement in HSEM velocity compared with those receiving standard care; this improvement became statistically significant once patients taking benzodiazepines—which impair saccadic eye movement—were excluded. Similar improvement in HSEM velocity was seen in the patients aged <12 years. Improvement or stabilization of all