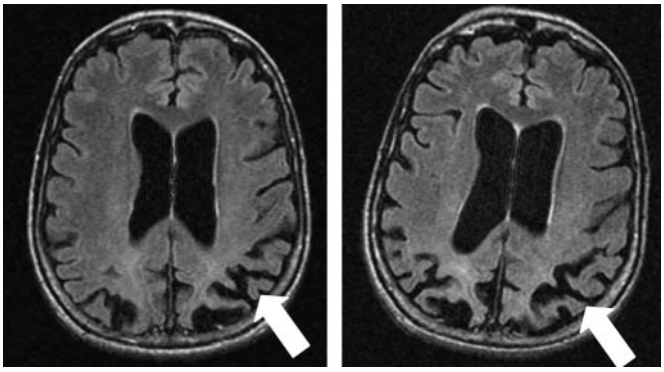
Posterior cortical dementia as a variant of Alzheimer disease



Serial MRI images of a patient with PCA showing occipito-parietal atrophy (large white arrows).

Tang-Wai et al. report the clinical, genetic, and neuropathologic characteristics of 40 patients with posterior cortical atrophy. Occipito-parietal dysfunction dominates the presentation. Their genetics and histopathology are like that of Alzheimer disease. However, the greatest concentration of neurofibrillary tangles is in extrastriate visual association cortex, not the hippocampus.

see page 1168

The differential diagnosis of posterior cortical dementia

Posterior cortical dysfunction presented with progressive visuospatial deficits in 27 patients described by Renner et al. These patients have a barrage of aphasias, apraxias, and elements of Balint's and Gerstmann's syndromes. Although Alzheimer neuropathology is most common, tauopathic and prion related pathologies occur. This syndrome is clinically distinguishable from other late-life dementias.

see page 1175

Lost but not forgetting: Posterior cortical Alzheimer disease

The accompanying editorial by Charles J. Duffy discusses how Tang-Wai et al. and Renner et al. echo Alois Alzheimer's description of focal cortical deficits. Phenotypic diversity in Alzheimer disease may betray genotypic heterogeneity or interactions between genetic predispositions and physiologic modulation. In either case, more specific diagnosis may be a further step toward clarifying pathophysiology and individualizing therapy.

see page 1148

Diabetes mellitus and risk of dementia in the elderly

In a follow-up study on a community cohort age 75 years and older, Xu et al. found that diabetes mellitus increased the risk of dementia, and vascular dementia in particular. The risk effect was increased by severe systolic hypertension and heart disease.

see page 1181

Hyperinsulinemia and the risk of AD: Effect of APOE-ϵ4

In a prospective study of elderly persons reported by Luchsinger et al., the risk of Alzheimer disease (AD) and memory decline increased with hyperinsulinemia. The authors also found that the increase in risk of AD related to hyperinsulinemia was higher for persons with the APOE-ϵ4 allele.

see page 1187

Morphometric determinants of verbal recall across weeks

In an MRI study of normal subjects who underwent testing of memory over a mean period of 11 weeks, Walhovd et al. found that hippocampal and cortical size predicted recall at different intervals. Hippocampal size mattered more across weeks than at short intervals, explaining variance not accounted for by age, total cortical, or white matter volume.

see page 1193

Education and the course of cognitive decline in AD

In a longitudinal study of nearly 500 people with Alzheimer disease (AD), Wilson et al. found that rate of cognitive decline gradually accelerated, particularly in people with relatively more education, supporting the idea that the course of cognitive decline in AD depends partly on premorbid life experiences.

see page 1198

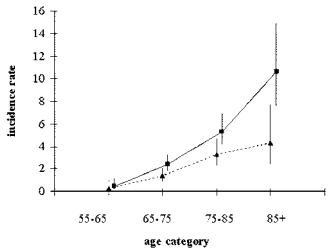
Stroke program improves discharge treatment utilization

Ovbiagele et al. assessed the impact of an aggressive, systematic hospital-based interdisciplinary stroke prevention treatment program and found that its implementation was associated with a substantial increase in treatment utilization of its key evidence-based therapy goals at the time of hospital discharge.

see page 1217

continued on page 1147

Incidence of parkinsonism and Parkinson disease (PD)



Incidence rates per 1,000 person years.

In a prospective population-based study in 6,839 participants with 39,879 person-years of follow-up, de Lau et al. found that the incidence of parkinsonism and PD was higher than most previously reported figures. More than one third of the incident PD cases, identified through in-person screening, had not been diagnosed with PD before and would not have been included in a study based on clinical data only.

see page 1240

Cannabis for dyskinesia in Parkinson disease

Carroll et al. report a double-blind randomized placebo-controlled study involving 25 patients, which showed that whole cannabis extract had no objective or subjective effect on dyskinesia in Parkinson disease.

see page 1245

POLG mutations cause ataxia and CNS abnormalities

Van Goethem et al. found recessive *POLG* mutations in eight patients presenting with adult onset sensory ataxia. Associated CNS features included myoclonus, epilepsy, and characteristic abnormalities on MRI and on autopsy. Ophthalmoplegia, myopathy, and mitochondrial abnormalities on muscle biopsy need not be present in patients with *POLG* mutations.

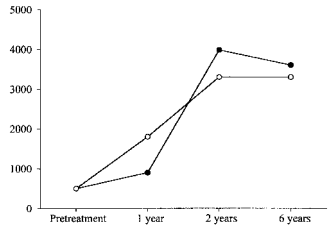
see page 1251

Hypocretin 2 receptor gene and cluster headache

Genetic factors play a role in cluster headache. In this expedited Brief Communication, Rainero et al. present evidence for an association between cluster headache and the hypocretin 2 receptor gene.

see page 1286

Long-term IVIg in multifocal motor neuropathy



Black circles: radial nerve; white circles: common peroneal nerve.

Periodic IV immunoglobulin (IVIg) therapy results in sustained clinical improvement in multifocal motor neuropathy, without secondary resistance to treatment. In this study, Vucic et al. shows that this improvement correlates with a decrease in the number of conduction blocks and development of reinnervation. The opposite conclusions of European studies may be related to their use of lower IVIg doses.

see page 1264

Volume reduction in Landau-Kleffner syndrome

Takeoka et al. detected volume reduction on MRI bilaterally in superior temporal areas (26–51%), in planum temporale (25–63%), in four children with Landau-Kleffner syndrome (LKS).

see page 1289

The accompanying editorial by Bourgeois and Landau notes that in the half century since Landau and Kleffner published their Neurology *article on six children with acquired aphasia and convulsive disorder, neither the pathogenesis nor proper treatment of LKS has been defined. The aphasia is predominantly receptive with secondary impairment of expressive language. Patients have few seizures, usually readily controlled and up to 1/3 have none. Epileptiform changes occur, usually in the temporal lobes but they, along with clinical seizures, resolve spontaneously before or during adolescence. More than 50% of patients followed into adulthood have language impairment. The Takeoka et al. article is the first evidence of anatomic change in LKS: all four LKS patients had a reduction (36 to 51%) in auditory association cortex volumes. They pose the fundamental question: Is the focal cortical atrophy the cause of LKS or the consequence of excitotoxicity? That the atrophy is the result of intractable epileptiform activity is suggested by the fact that: the subacute aphasia usually occurs after or simultaneously with seizures. Second, the physiologic changes associated with atrophy are likely to be irreversible, and the spikes are known to disappear with time. They call for a multi-institutional study with carefully defined clinical subgroups searching for etiology and conducting rigorous studies of treatment.*

see page 1152

October 12 Highlights

Neurology 2004;63;1146-1147
DOI 10.1212/WNL.63.7.1146