Behavioral problems in primary progressive aphasia

Rosen et al. found that semantic dementia is associated with significantly more behavioral dysfunction than other variants of primary progressive aphasia, and that the behavioral problems are similar to those seen in FTD.

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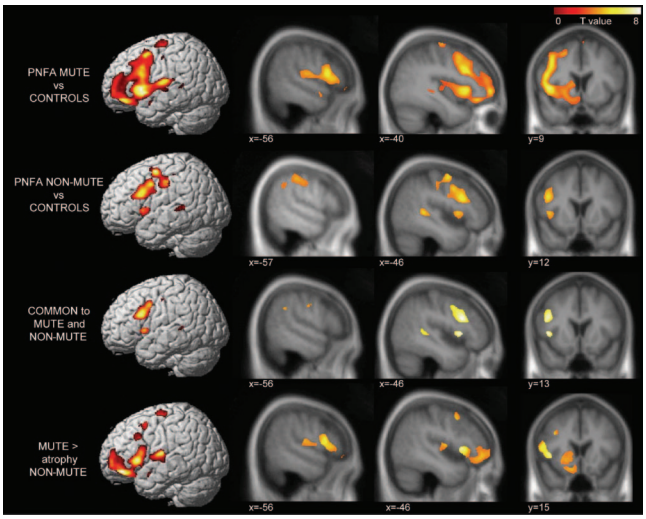
Atrophy of the speech network in mutism



Atrophy in mute patients vs controls (A); non-mute patients vs controls (B) (simple main effects). Area of common atrophy in mute and non-mute patients vs controls (C) and areas of greater atrophy in mute vs non-mute patients and controls (D).

Gorno-Tempini et al. used voxel-based morphometry on MRI scans of patients with progressive nonfluent aphasia (PNFA). The authors showed that patients with PNFA who presented with early complete loss of speech had greater gray matter atrophy in left pars opercularis of Broca's area and basal ganglia vs patients with PNFA who retained speech abilities, and healthy controls.

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The editorial by John Ringman and Argye Hillis about these two articles notes that disproportionate metabolic dysfunction or atrophy in the left posterior inferior frontal cortex identified through clinical imaging may predict relatively early mutism, which may be useful in planning for daily management and living arrangements.

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Pregnancy outcomes and levetiracetam

Hunt et al. report outcomes of human pregnancies exposed to levetiracetam. Major congenital malformations were found in none of 39 monotherapy exposures and 3 of 78 polytherapy exposures.

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Targeting Aβ with 3-amino-1 propanesulfonic acid in AD

Aisen et al. assessed the safety, tolerability, and pharmacokinetic/pharmacodynamic profile of 3-amino-1 propanesulfonic acid (3APS), a novel anti-amyloid compound in AD. 3APS demonstrated a satisfactory safety and tolerability profile and reduced CSF Aβ_{42} concentrations in patients. Results support the pharmacologic and potential therapeutic effects of 3APS on disease progression.

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Familial dystonia with cerebellar atrophy

Le Ber et al. report an unusual "dystonia plus" phenotype in eight families characterized by spasmodic dysphonia and upper limb dystonia. Mean age at onset was 27 years. The paucity of cerebellar signs contrasted with marked and global cerebellar atrophy.

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The editorial by H.A. Jinnah and E.J. Hess notes that because these patients did not have any known metabolic or degenerative causes, they collectively may justify the recognition of a new "dystonia plus" syndrome combining a slowly progressive ataxia with focal or segmental dystonia. Le Ber et al. suggest that dystonia, at least in their patients, arises from dysfunction of the cerebellum, challenging traditional views of the anatomy of dystonia, which focus predominantly on the basal ganglia. The link between dystonia and the cerebellum is that brain MRI revealed prominent atrophy of the cerebellum, without obvious abnormalities of the basal ganglia. In nearly all of the functional imaging studies showing abnormal basal ganglia function in dystonia, there is parallel evidence for abnormal cerebellar function. For dystonia, a role for the basal ganglia is undisputed, yet evidence similar to what was used to establish the link between the basal ganglia and dystonia is accumulating for the cerebellum.

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Driving ability of patients with PD

Uc et al. found that drivers with PD were more likely to experience worsening of driving safety errors during an auditory-verbal distraction task on a freeway in an instrumented vehicle vs controls. Measures of cognition, motor function, and sleepiness predicted effects of distraction on driving performance within the PD group.

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Deep brain stimulation and hypotension in PD

Stemper et al. analyzed orthostatic regulation in 14 patients with PD who had bilateral STN stimulators. The authors report that STN stimulation increased peripheral vasoconstriction and baroreflex sensitivity and stabilized blood pressure.

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LRRK2 mutations in early onset PD

Clark et al. found an association of G2019S mutation with both early and late-onset PD, confirming the higher frequency of G2019S in Jewish than in non-Jewish cases, especially those with a family history of PD. They estimate penetrance in predicted carrier relatives to be 24.0% (95% CI: 13.5% to 43.7%).

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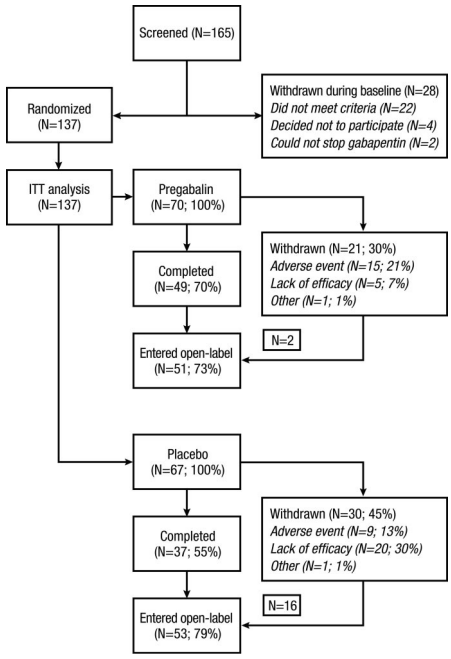
Pregabalin and neuropathic spinal cord injury pain



In a large, randomized controlled trial in people with central neuropathic pain following spinal cord injury, Siddall et al. found that pregabalin significantly improved pain, sleep, and anxiety and overall status vs placebo.

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Prediction of thrombolytic efficacy by CT

Kim et al. used thin-section noncontrast CT to estimate thrombus composition based on Hounsfield Units. Thrombi with lower Hounsfield Units were resistant to thrombolysis in acute ischemic stroke.

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Incidence of cervical artery dissection

Lee et al. report the average annual incidence of dissections in a population-based study. Internal carotid artery dissection was detected approximately twice as frequently as vertebral artery dissection in the overall study, but in the latter half of the study period, vertebral artery and internal carotid artery dissection incidence rates were equivalent. The majority of cervical artery dissection patients in the community have excellent outcome, and contrary to many tertiary referral series, re-dissection is rare.

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Laser identifies loss of optic nerve fibers in AD

Danesh-Meyer et al. found that compared to controls, patients with AD had an OR of 4.7 (95% CI: 2.3 to 9.8) for having a larger cup:disc ratio when the optic nerve head was measured with confocal scanning laser ophthalmoscopy (CSLO).

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The editorial by Neil R. Miller and David Drachman notes the observation that retinal ganglion cells and optic nerve fibers may be lost in AD, which raises three questions: Could CSLO or OCT serve as a marker for the diagnosis of AD or as a surrogate for severity of AD in clinical trials? Does retinal ganglion cell and optic nerve fiber loss indicate that AD is a widespread, generalized brain disorder that is not confined to the mesial temporal region and is not exclusively a dementia? What insight can retinal ganglion cell and optic nerve fiber pathology provide about the mechanisms of AD? Their review considers these, noting that while one might expect to see the hallmark pathology of AD—neurofibrillary tangles and neuritic plaques in the retina—these classic neuropathologic features of AD are notably absent in retinal tissue. This suggests that beta amyloid and tau protein may not be the prime cause of neuronal and nerve fiber degeneration in AD.

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Study of pirfenidone in adults with neurofibromatosis type 1

Babovic-Vuksanovic et al. studied the effect of pirfenidone in 24 patients with neurofibromatosis type 1, using three-dimensional MRI to assess outcome. At the end of treatment, four patients had a decrease in tumor volume by 15% or more, three had tumor progression, and 17/24 remained stable.

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Early vs delayed treatment of seizures

The Marson et al. randomized multicenter study of early epilepsy and single seizures provides evidence of an effect for carbamazepine as monotherapy in delaying time to first seizure recurrence, but mixed evidence of an effect for valproate. Immediate treatment was superior to delayed treatment, and remission rates at 1-, 3-, and 5-year time points were similar for the two drugs.

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