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Abstracts

Articles appearing in the September 2017 issue

IgLON5 antibody: Neurologic accompaniments and outcomes in 20 patients

Objective To describe the phenotypes, treatment response, and outcome of IgLON5 autoimmunity.

Methods Archived serum and CSF specimens from 367 patients known to harbor unclassified antibodies that stained neural synapses diffusely (mimicking amphiphysin immunoglobulin G [IgG]) were reevaluated by indirect immunofluorescence assay (IFA) using a composite of mouse tissues and recombinant IgLON5-transfected cell-based assay (CBA; Euroimmun).

Results Available specimens (serum, 25; CSF, 9) from 26/367 patients (7%) had identical IFA appearance and robust IgLON5 CBA positivity. Clinical information was available for 20/26 patients; 13 were women. Median disease onset age was 62 years (range 46–75 years). Most patients had insidious onset and progression of neurologic symptoms affecting movement and sleep predominantly. Sleep disorders were sleep-disordered breathing (11) and parasomnias (3). Brainstem disorders were gait instability (14), dysphagia (10), abnormal eye movements (7), respiratory dysfunction (6), ataxia (5), craniocervical dystonia (3), and dysarthria (3). Findings compatible with hyperexcitability included myoclonus (3), cramps (3), fasciculations (2), and exaggerated startle (2). Neuropsychiatric disorders included cognitive dysfunction (6), psychiatric symptoms (5), and seizures (1). Dysautonomia, in 9, affected bladder function (7), gastrointestinal motility (3), thermoregulation (3), and orthostatic tolerance (1). Just 2 patients had coexisting autoimmune disease. Brain MRI findings were nonspecific and CSF was noninflammatory in all tested. Seven of 9 immunotherapy-treated patients improved: 6 of those 7 were stable at last follow-up. Three untreated patients died. Each IgLON5-IgG subclass (1–4) was readily detectable in ≥80% of specimens using CBA.

Conclusions IgLON5-IgG is diagnostic of a potentially treatable neurologic disorder, where autoimmune clues are otherwise lacking.

NPub.org/9013a

Reduced rich-club connectivity is related to disability in primary progressive MS

Objective To investigate whether the structural connectivity of the brain's rich-club organization is altered in patients with primary progressive multiple sclerosis (MS) and whether such changes to this fundamental network feature are associated with disability measures.

Methods We recruited 37 patients with primary progressi2ve MS and 21 healthy controls for an observational cohort study. Structural connectomes were reconstructed based on diffusion-weighted imaging data using probabilistic tractography and analyzed with graph theory.

Results We observed the same topologic organization of brain networks in patients and controls. Consistent with the originally defined rich-club regions, we identified superior frontal, precuneus, superior parietal, and insular cortex in both hemispheres as rich-club nodes. Connectivity within the rich-club was significantly reduced in patients with MS ($p = 0.039$). The extent of reduced rich-club connectivity correlated with clinical measurements of mobility (Kendall rank correlation coefficient $\tau = -0.20$, $p = 0.047$), hand function ($\tau = -0.26$, $p = 0.014$), and information processing speed ($\tau = -0.20$, $p = 0.049$).

Conclusions In patients with primary progressive MS, the fundamental organization of the structural connectome in rich-club and peripheral nodes was preserved and did not differ from healthy controls. The proportion of rich-club connections was altered and correlated with disability measures. Thus, the rich-club organization of the brain may be a promising network phenotype for understanding the patterns and mechanisms of neurodegeneration in MS.

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F. Mir, D. Lee, H. Ray, S.A. Sadiq. 2014;1:e21. doi.org/10.1212/NXI.0000000000000021

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Y. Hacohen, M. Absoud, C. Hemingway, et al. 2014;1:e2. doi.org/10.1212/NXI.0000000000000002