

Cutaneous denervation in Progressive Supranuclear Palsy

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Progressive Supranuclear Palsy (PSP) is a rare neurodegenerative disease characterized by intraneuronal accumulation of hyperphosphorylated Tau. Because PSP is strongly linked biochemically and genetically to tau protein abnormalities there is a growing interest in new tau-directed therapies and in the search of new biomarkers able to monitor disease progression. We studied cutaneous innervation in PSP in order to identify new biomarkers of nerve degeneration through a minimally invasive technique such as skin biopsy. We performed 3 mm punch biopsies from thigh, leg and fingertip of 12 PSP patients (mean age = 68.4 ± 7.8). Samples were processed with indirect immunofluorescence using a large panel of antibodies to visualize and quantify sensory and autonomic cutaneous nerves. Results were compared with those obtained from a population of age and sex matched controls (mean age = 69 ± 3.3). In PSP patients, compared to controls, we found a severe length-dependent loss of intraepidermal nerve fibers associated to a severe loss of autonomic vasomotor, sudomotor and pilomotor nerves. Moreover, we observed a marked loss of Meissner corpuscles and intrapapillary myelinated endings in glabrous skin. The pattern of nerve degeneration appeared different compared to other neurodegenerative disorders. Skin may provide useful neuropathological biomarkers to study PSP progression.

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Multi-centric longitudinal study on electrophysiological diagnostic and prognostic markers in prolonged disorders of consciousness

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Diagnostic accuracy and reliable assessment of prognosis are challenging in patients in vegetative state (VS) and in minimally conscious state (MCS). This longitudinal multi-centric study aims to identify neurophysiological patterns with diagnostic and prognostic value. 59 patients in VS and 63 in MCS underwent clinical assessment by Coma Recovery Scale-Revised³ and neurophysiological evaluations of EEG background activity and reactivity, somatosensory evoked potential (SEP) and event related potential (ERP) at ≤ 3 months after brain-injury. Clinical outcome was evaluated at 6 months post-injury. EEG background activity and reactivity significantly differed between VS and MCS patients ($p < .001$), whereas the presence of cortical component N20 on SEP and P300 on ERP did not differ between the two groups. Good outcome was

significantly more frequent in patients with moderately abnormal to normal EEG background activity than in patients with poor EEG background organization ($p = .001$), in patients showing EEG reactivity ($p < .001$) and in patients showing P300 ($p = .016$). The presence of SEP did not differ significantly between the two prognostic groups. Multimodal clinical and neurophysiological assessment could provide useful diagnostic and prognostic information for disorder of consciousness. This multicentric project calls for international standardization of diagnostic and prognostic procedures.

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Steady-state visual evoked potentials in photosensitive patients with idiopathic generalized epilepsy

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To assess the visual system excitability of photosensitive patients with idiopathic generalized epilepsy (IGE) with the steady-state flash visual evoked potential (SS-FVEP) technique. We studied 13 adult patients with IGE showing a photoparoxysmal electroencephalographic response (PPR). Twenty normal subjects of similar age and sex acted as controls. We recorded FVEPs from occipital electrodes (1–1000 Hz; 10 kHz) with eyes closed. Stimuli were 3 s trains of flashes delivered at 8 and 16 Hz. Eighty sweeps were collected for any stimulus type and averaged off-line. SS-FVEPs were analyzed in the frequency domain with a discrete Fourier transformation and the amplitude of the first (1H) and second harmonic (2H) were measured. In patients, at 16 Hz, a significant ($p < 0.05$) increase in the first and second harmonic amplitude (i.e. 16 and 32 Hz) was found compared to controls. No significant changes at 8 Hz were detected. The SS-FVEP technique documents significant changes in the visual system of photosensitive patients with IGE, whose features likely underlie the PPR development.

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Neurophysiological prognosis in comatose patients after cardiac arrest: The Italian Multicentric Study (ProNeCA)

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SEPs are reliable predictor of poor outcome in comatose patients after cardiac arrest (CA). Recently also the role of EEG has been reassessed. To evaluate the prognostic value of EEG and SEPs association in post-anoxic coma at different recording time. We included comatose patients after CA. EEG and SEPs were recorded within 12 hs and at 72 hs after CA. EEG was classified into grade 1 ("continuous"/"neurally-continuous") and grade 2 (discontinuous/burst-suppression/suppression/isolectric). SEPs were dichotomized into "bilaterally

absent" (BA) and "present". Outcome was evaluated at 6 months by GOS. "Recovery of consciousness" was considered good outcome. Ten centers took part to the study and 282 patients were included. EEG and SEPs were recorded in 141 within 12 hs in 162 at 72 hs. Grade 1 EEG at 12 hs always predicted good outcome, grade 2 pattern at 72 hs predicted poor outcome with higher predictive power. BA SEPs always predicted poor outcome. Grade 1 EEG was always associated with present SEPs. EEG is a reliable time-dependent

predictor of good outcome (within 12 h) and poor outcome (after 72 h). SEP is time-independent predictor of poor outcome. Early grade 1 EEG is never associated with BA SEP. Early association recordings of EEG and SEPs increase of the reliability of prognosis prediction in post-anoxic coma.

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