

EPO-153 | Comparative analysis of paranodal antibody assays in autoimmune nodopathies: Accuracy and inter-laboratory agreement

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Background and aims: Autoimmune nodopathies (AN) are a subgroup of neuropathies that harbour antibodies targeting paranodal/nodal proteins (PNAbs): NF155, NF186, CNTN1 and CASPR1. However, laboratory strategies for their detection are not standardized. We aimed to evaluate the performances of PNAbs detection assays in two expert Neuroimmunology Centres.

Methods: A cohort of chronic inflammatory demyelinating polyneuropathy (CIDP) patients (n=45), pathological controls (n=34, 6 immune-mediated neuropathies, 12 non-immune-mediated neuropathies, 16 normal pressure hydrocephalus), and healthy controls (n=10) were tested using a commercial CBA (CCBA) or in-house assays: CBA on living cells (LCBA) and ELISA. PNAbs positivity required confirmation by both in-house tests at either Centre. CIDP patients with PNAbs were considered AN and “true positives.” We assessed analytic performances using sensitivity, specificity and accuracy, and inter-laboratory agreement using Fleiss’ Kappa test with 95% confidence intervals (CI).

Results: In-house assays detected PNAbs in 21 AN patients (NF155=11, PanNF=1, CNTN1=6, CASPR1=3), with an overall accuracy of 99.6% (sensitivity: 99.6%; specificity: 100%), with substantial agreement between Centres (Fleiss’ kappa: 0.668, 95% CI: 0.528 – 0.778). CCBA demonstrated comparable performance (accuracy: 98.4%; sensitivity: 92.8%; specificity: 98.8%). ELISA showed the lowest accuracy (95.5%; sensitivity: 97.6%; specificity: 95.4%). Substantial overall inter-laboratory agreement for PNAbs (Fleiss’ kappa: 0.735, 95% CI: 0.635 – 0.817) was observed, with only 3/89 (3.4%) discordant samples. CCBA showed the highest agreement (0.914), while ELISA the lowest (0.712).

Conclusion: PNAbs assays have overall good performances and reproducibility. CCBA represents a reliable alternative to in-house assays for their detection.

Disclosure: Nothing to disclose.

EPO-154 | GBS at the Norfolk and Norwich Hospital (NNUH): A retrospective study regarding our practice and adherence to guidelines

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Background and aims: Guillain-Barre Syndrome (GBS) is an autoimmune polyradiculoneuritis causing ascending paralysis, sensory impairment and potential bulbar and autonomic dysfunction. Diagnosis is clinical and supported by CSF analysis and NCS/EMG. In the UK, the incidence is estimated at around 2/100,000/year, with 3-7% mortality. National guidelines enforce early IVIg/PLEX treatment and close FVC monitoring. Our study aims to evaluate our department’s performance in diagnosing and treating the condition according to the guidelines and identify areas requiring improvement.

Methods: A retrospective analysis was conducted on GBS patients admitted to NNUH (January 2019 - July 2023). Data analysed included demographic details, Hughs score, CSF analysis, NCS/EMG and FVC.

Results: Thirty-nine GBS patients were identified (M:F=2:1, average age 59). 66% presented with typical ascending paralysis. Other presentations included ataxia, ophthalmoplegia, and bulbar palsy. Average Hughs score was 2.8 - 4. IVIg were administered to 80% of patients (100% of patient requiring it due to Hughs > 2). Albuminocytologic dissociation was detected in 48.7% cases. Supportive NCS/EMG were found in 49% patients. 23% patients required ITU admission (FVC<1.5). Incidence was 0.89/100,000/year with 10% mortality.

Conclusion: NNUH showed very good guideline compliance regarding investigations, monitoring and escalation of GBS patients. All eligible cases received prompt immunotherapy and ventilatory support in ITU. CSF analysis and NCS/EMG were performed to support the diagnosis, reflecting good clinical practice. Our study showed lower incidence and higher mortality in our catchment area compared to the national average, suggesting underdiagnosis of milder cases which did not reach hospital attention.

Disclosure: Nothing to disclose.

EPO-155 | Electrophysiological evaluation of peripheral nerve injuries after the february 6 earthquake in Turkey

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Background and aims: The February 6, 2023, earthquake in Turkey caused over 50,000 deaths and more than 100,000 injuries. This study aims to evaluate the peripheral nerve, plexus, and root injuries of earthquake-affected patients in terms of EMG findings.

Methods: The clinical and EMG findings of patients trapped under rubble and admitted to three different centers after the earthquake were retrospectively evaluated.

Results: Among the 47 patients examined in the EMG laboratory, 19 were male and 28 were female. The average age of the patients undergoing EMG was 29.5 years, with the youngest being 6 years old and the oldest 69 years old. Seventeen of the patients were under the age of 18. Upper extremity injuries were

found in 12 patients, lower extremity injuries in 34 patients, and both upper and lower extremity injuries in 1 patient. The average duration of being trapped under rubble was 29.5 hours. EMG examinations revealed peripheral nerve injury in 37 patients, plexus injury in 8 patients, root injury in 1 patient, and both plexus and peripheral nerve injury in 1 patient. The most common peripheral nerve injury was found in the peroneal nerve at a rate of 46%, followed by sciatic and tibial nerve injuries.

Conclusion: Severe peripheral nerve injuries were identified in earthquake survivors, causing significant disability. Long-term follow-up is necessary to assess nerve regeneration and the effectiveness of physiotherapy.

Disclosure: Nothing to disclose.

EPO-156 | Intraepineurial FF as a novel biomarker in TTR amyloidosis patients and asymptomatic carriers using MR neurography

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Background and aims: Transthyretin familial amyloid polyneuropathy (ATTRv-PN) is a rare and progressive neurodegenerative disorder characterized by axonal neuropathy. Early detection of disease onset and progression is crucial for timely therapeutic intervention. Intra-epineurial fat fraction (FF) could reflect lipid droplets in amyloid deposits or epineurial fatty-rich replacement due to nerve fibers loss. This study investigates the utility of quantitative magnetic resonance imaging (qMRI) biomarkers, particularly intra-epineurial FF, in differentiating ATTRv-PN from asymptomatic carriers (ATTRv-C) and healthy controls (HC).

Methods: 53 patients with TTR mutations, (31 symptomatic ATTRv-PN, 22 ATTRv-C), and 24 HC were included and imaged. Sciatic and tibial nerves were segmented on anatomical sequences. qMRI parameters including FF, MTR (Magnetization transfer ratio) and volume were computed from the sciatic and tibial masks to quantify nerve morphology, structural integrity, and intraepineurial fat fraction-like. Correlations between qMRI metrics, clinical and electrophysiological parameters were calculated.

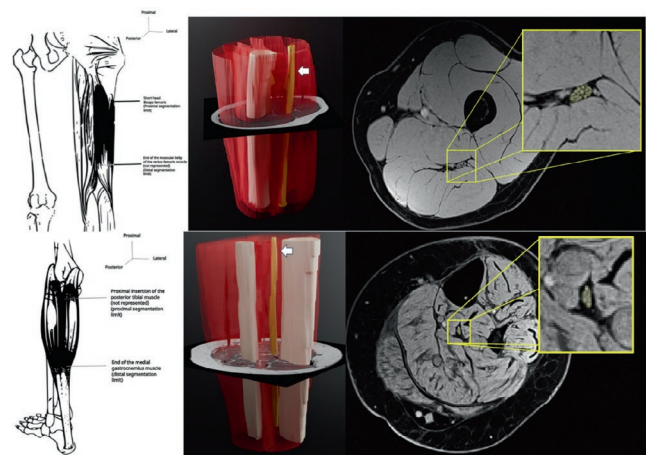


FIGURE 1 Anatomical Landmarks for segmentation limits are shown in the left column. Sciatic nerve (upper row) and tibial nerve (lower row) are marked by a white arrow (3D reconstruction, central column) and marked in yellow (right column).

Results: Symptomatic ATTRv-PN patients exhibited significantly higher intra-epineurial FF in both sciatic (median value 32.4% IQR [24.4-38.1]) and tibial nerves (median value 13.7%, IQR [9.97-20.7]) compared to controls (sciatic median value 22.3%, IQR [16.6-28.5]; tibial median value 9.74%, IQR [6.36-12.5] respectively) ($p < 0.05$). Intra-epineurial FF values were positively correlated in both uni- and multivariate analysis with clinical and electrophysiological scores. ATTRv-C also showed increased FF compared to controls ($p < 0.05$). Additionally, MTR and nerve volumes exhibited less pronounced differences across groups.

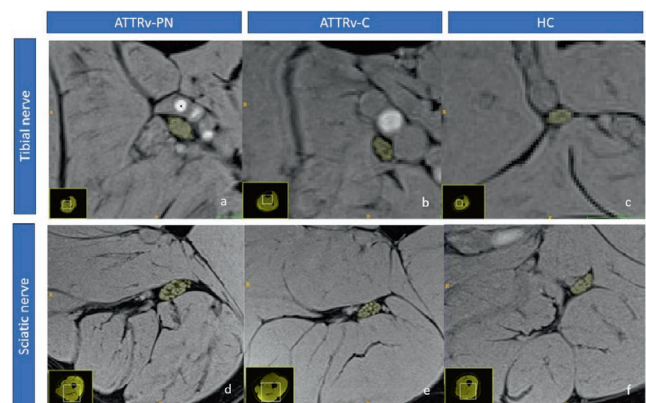


FIGURE 2 Enlarged views of nerve ROIs. Tibial nerve ROI in a symptomatic ATTRv-PN patient (a); ATTRv-C patient (b) and healthy control (c); Sciatic nerve ROI in an ATTRv-PN patient (d); ATTRv-C patient (e) and healthy control (f).