

Supplementary data

Supplementary table 1 Clinical data at onset and at the most severe disease stage according to anti-MAG titre < 10 000 or ≥ 10 000 BTU

| Findings, number (%) | Patients (n=202) | Anti-MAG titre < 10 000 BTU (n=22) | Anti-MAG titre ≥ 10 000 BTU (n=180) |
|--|------------------------------|--|---|
| Onset | | | |
| Acute | 5 (2.5) | 0 | 5 |
| Subacute | 31 (15.3) | 2 | 29 |
| Progressive | 166 (82.2) | 20 | 146 |
| Symptoms at onset | | | |
| Paresthesias/dysesthesias | 180 (89.1) | 20 | 160 |
| Pure sensory deficit | 175 (86.6) | 20 | 155 |
| <i>Distal part of the LL</i> | 122 | | |
| <i>of the UL</i> | 12 | | |
| <i>of the LL and UL</i> | 37 | | |
| Hypoesthesia | 168 (83.2) | 19 | 149 |
| Sensory ataxia | 108 (53.4) | 12 | 96 |
| Neuropathic pain | 85 (42.1) | 12 | 73 |
| Sensorimotor deficit | 27 (13.4) | 2 | 25 |
| <i>Motor deficit at distal part of the LL</i> | 13 | | |
| <i>of the UL</i> | 3 | | |
| <i>of the LL and UL</i> | 4 | | |
| <i>Proximal motor deficit</i> | 7 | | |
| Postural and intention tremor | 22 (10.9) | 3 | 19 |
| Symptoms at the most severe disease stage | | | |
| Paresthesias/dysesthesias | 181 (89.6) | 19 | 162 |
| Sensory ataxia | 152 (75.2) | 16 | 136 |
| Neuropathic pain | 131 (64.9) | 11 | 120 |
| Postural and intention tremor | 64 (31.7) | 5 | 59 |
| Motor deficit | 87 (43.1) | 7 | 80 |
| Clinical examination at the most severe disease stage | | | |
| Asymmetric motor deficit in LL | 9 | 0 | 9 |
| Motor deficit purely distal in LL/UL | 50 / 10 | 7/1 | 43/9 |
| ONLS (arm scale/5) | 1.1 ^(0-5, NK= 1) | 0.8 | 1.2 |
| ONLS (leg scale/7) | 1.7 ^(0-7, NK= 1) | 1.7 | 1.7 |
| ONLS (global/12) | 2.9 ^(0-11, NK= 1) | 2.5 | 2.9 |
| Significant disability score (>2) [*] | 45 (22.4) ^(NK= 1) | 5 | 40 |

Statistical analysis did not show significant difference between the two groups.* According to the modified functional impairment scale described by Nobile-Orazio *et al.* (2000).

LL, lower limbs; n, number of patients; NK, not known; UL, upper limbs.

Supplementary table 2 EDX features at diagnosis

| Motor nerve conduction studies | DML (ms) | CMAP amplitude (mV) | CB (n) (%) | MCV (m.s⁻¹) | F-WAVE minimum latency (ms) |
|---------------------------------------|------------------------|---------------------------------|-------------------------------|---------------------------------------|------------------------------------|
| Peroneal n=366 | 10.7 (3.2-28.6) | 1.6 (0.1-9.2) UN=129 (38.4%) | 35 (9.6) | 27.3 (6-52) | 72.3 (47-118) NE=207 |
| Tibial n=337 | 10.9 (3.5-28) | 2.1 (0.1-18) UN=122 (36.2%) | 13 (3.9) | 28.7 (10-48.5) | 69.9 (36.2-104) NE=189 |
| Median* n=337 | 8.1 (3.2-23.2) | 5.4 (0.2-16) UN=7 | 28 (8.3) | 38.2 (8.7-68.1) | 39.1 (19.1-74) NE=45 |
| Ulnar** n=313 | 5 (2-17.2) | 6.4 (0.8-15.2) UN=6 | 39 (below elbow) (12.5) | 41.8 (7-69) (wrist-below elbow) | 38.2 (25.9-70.8) NE=32 |

Mean (limits).

Values in bold are considered as demyelinating according to European Federation of Neurological Societies and Peripheral Nerve Society (Joint Task Force of the EFNS and the PNS, 2010).

*Median terminal latency index (TLI) $\leq 0.25 = 269/330$ (82%), **ulnar TLI $\leq 0.25 = 86/303$ (28%)

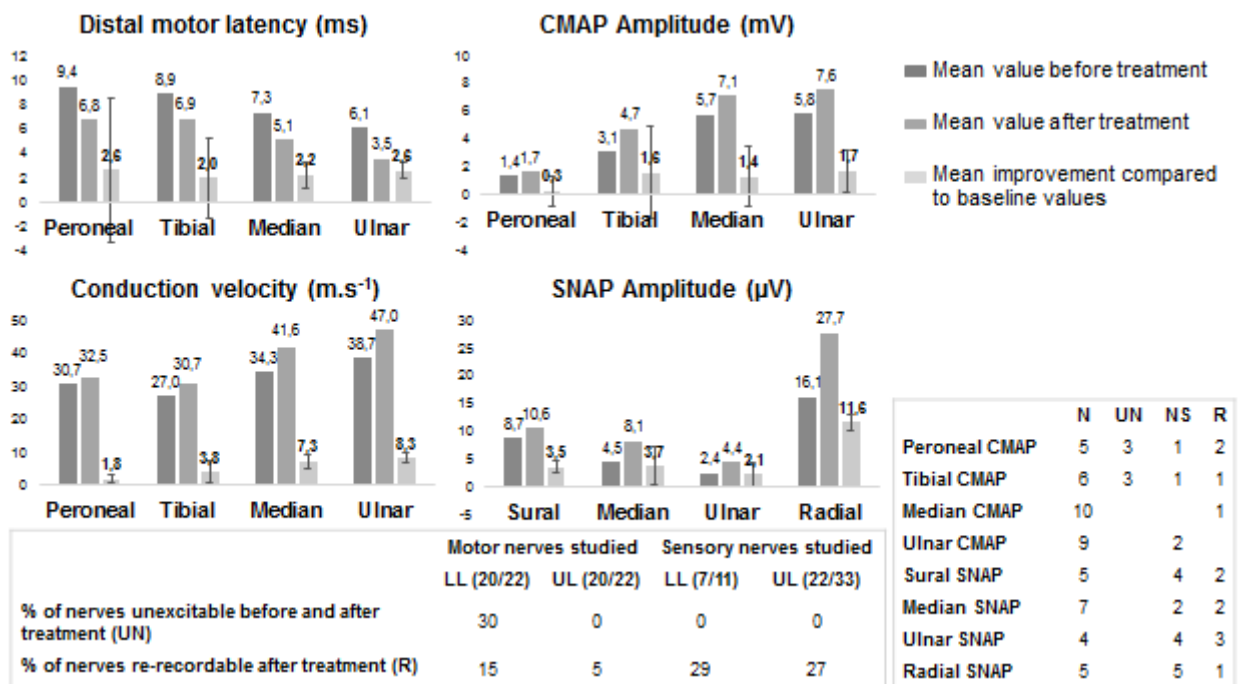
CB, conduction block; CMAP, compound muscle action potential; DML, distal motor latency; MCV, motor conduction velocity; n, number; NE, not elicitable; UN, unexcitable.

Supplementary table 3 Immunomodulatory therapies and clinical response

| Findings, number (%) | Patients (n=202) | Clinical response during | |
|---------------------------------|-----------------------------|---|--|
| | | the 6-month follow-up period | the 7-12-month follow-up period |
| IVIg courses | 68 (33.7) | 19 (27.9) | 3 (4.4) |
| Plasma exchanges | 24 (11.9) | 4 (16.7) | 1 (4.2) |
| Oral corticosteroids | 14 (6.9) | 0 | 3 (21.4) |
| IV corticosteroids | 7 (3.5) | 0 | 0 |

IVIg, intravenous immunoglobulins.

Supplementary figure 1



EDX data of responder or stabilised patients presenting with improvement of EDX parameters after rituximab associated or not with immunosuppressants (n=11).

Data are provided for the right side which was mainly studied. Only data concerning recordable nerves before and after treatment are provided in the graphics. Initially non recordable nerves for which a response has been recorded after treatment are designed as R=re-recordable.

CMAP, Compound Muscle Action Potential; N, number of nerves recordable; UN, number of nerves unexcitable before and after treatment; NS, number of nerves not studied; SNAP, sensory nerve action potential.