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Management of Patients With Multifocal Motor Neuropathy: A Global Quantitative Survey of Neurologists

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BACKGROUND

- MMN is a rare, immune-mediated, complement-driven, chronic neuropathy leading to demyelination and axonal degeneration and subsequent progressive, disabling, asymmetric limb weakness without sensory loss[1–3]
- Patients with MMN are frequently misdiagnosed with other MNDs, such as ALS[4]
- The EFNS/PNS 2010 Guidelines[5] on the definition, investigation, and treatment of MMN describe good practice points to define:[5]
  - Clinical and electrophysiological diagnostic criteria for MMN
  - Supportive criteria to categorize the diagnosis as either possible, probable, or definite MMN
- We report results from a 15-minute, 47-item online survey of neurologists treating MMN, aimed at understanding diagnostic and treatment patterns, perceived patient illness burden, and future treatment directions

STUDY DESIGN AND PARTICIPANTS

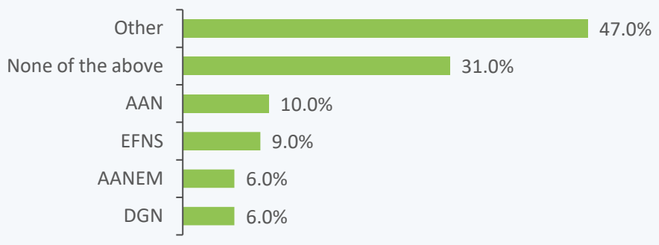
- Cross-sectional study collecting survey data from neurologists in the United States, Italy, Germany, Spain, Canada, the United Kingdom, Japan, the Netherlands, and Denmark
- Participating healthcare professionals remained anonymous to the investigators
- Criteria for participation included:
  - Neurology specialist
  - In practice for ≥2 years since residency
  - Treat/consult ≥2 MMN patients per year
  - Practice in one of the nine study countries (excluding Vermont in the US)
- 250 neurologists completed the survey

Country	Participants, n (%)
United States	100 (40.0)
Italy	41 (16.4)
Germany	30 (12.0)
Spain	30 (12.0)
Canada	18 (7.2)
United Kingdom	14 (5.6)
Japan	9 (3.6)
Netherlands	5 (2.0)
Denmark	3 (1.2)

RESULTS

Treatment Decisions: All Respondents

- **31%** of neurologists report making treatment decisions **without consulting a specific guideline**



- Neurologists who report not using a specific guideline use the following criteria to make MMN treatment decisions:
  - Clinical judgement
  - Response to IVIg
  - Patient response
  - Symptom severity
  - Grip strength/ complementary tests
  - Difficulty walking
  - Consensus among colleagues
- **79% of neurologists** surveyed report using **IVIg as 1L therapy** with 72% of their patients with MMN

	Total (n=250)		US (n=100)		OUS (n=150)	
	1L*	2L*	1L*	2L*	1L*	2L*
IVIg	79%	24%	75%	33%	82%	17%
CS	**33%**	**16%**	**36%**	**15%**	**31%**	**17%**
SCIg	18%	18%	16%	16%	20%	20%
PLEX	17%	27%	19%	27%	15%	27%
Rituximab	16%	31%	16%	25%	17%	35%

*As survey respondents did not specify if 1L therapies were used alone or in combination with other treatments, these data may exceed 100%.

- This survey produced responses that were **unexpected and inconsistent with EFNS/PNS MMN Treatment Guideline recommendations**:[5]
  - One-third of neurologists reported using **CS as 1L therapy** even though **they are not recommended to treat MMN**
  - Respondents report using an **average induction effective dose of 3.0 g/kg IVIg**, with 96% using between 0.1 and 10.99 g/kg [recommended range: between 1 and 2 g/kg[5]]

Survey Response Analysis

- Like all surveys, this survey is subject to limitations such as unintentional question and respondent bias
- Responses were filtered for those respondents **managing patients with MMN according to EFNS/PNS MMN Treatment Guidelines**

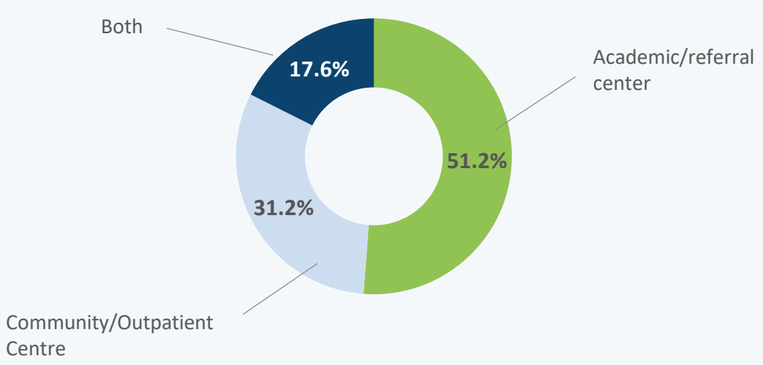
IVIg induction effective dose of 1.0–2.0 g/kg
AND
DO NOT USE CS as 1L
***n*=125**

Respondent and Patient Profile: Respondents Following Treatment Guidelines

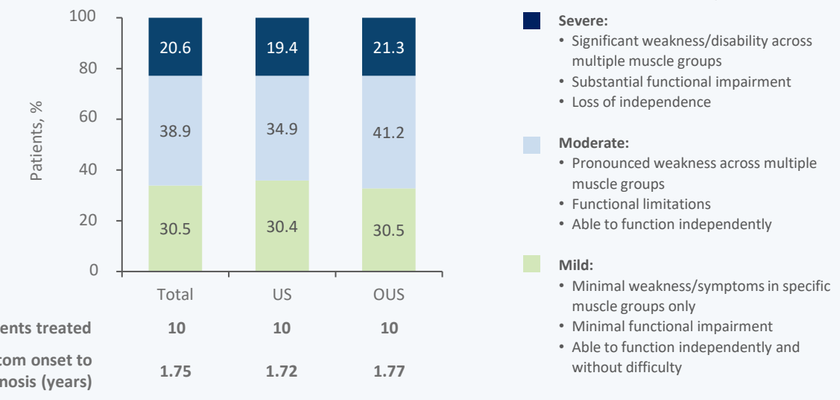
- Of the 250 neurologists surveyed, **125 follow EFNS/PNS IVIg dose and CS-use guidelines**

Country	Participants, n (%)
United States	46 (36.8)
Italy	23 (18.4)
Germany	18 (14.4)
Spain	12 (9.6)
Canada	11 (8.8)
United Kingdom	9 (7.2)
Netherlands	3 (2.4)
Denmark	2 (0.8)
Japan	1 (0.8)

- Over half (51.2%) of respondents report practicing at an academic or referral center and 31.2% at a community or outpatient center



- Neurologists report treating/consulting with a **median of 10 MMN patients per year**; the majority (39%) are considered to have moderate disease



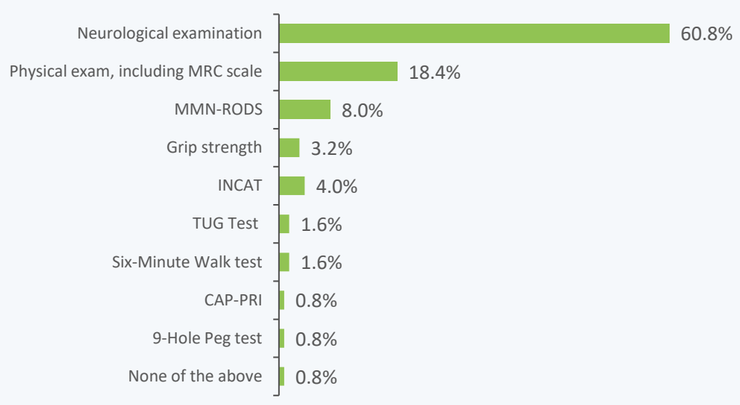
Diagnosis Decisions: Respondents Following Treatment Guidelines

- Of the neurologists surveyed, **70% of this subset report** their patients being **diagnosed with another peripheral neuropathy** before MMN

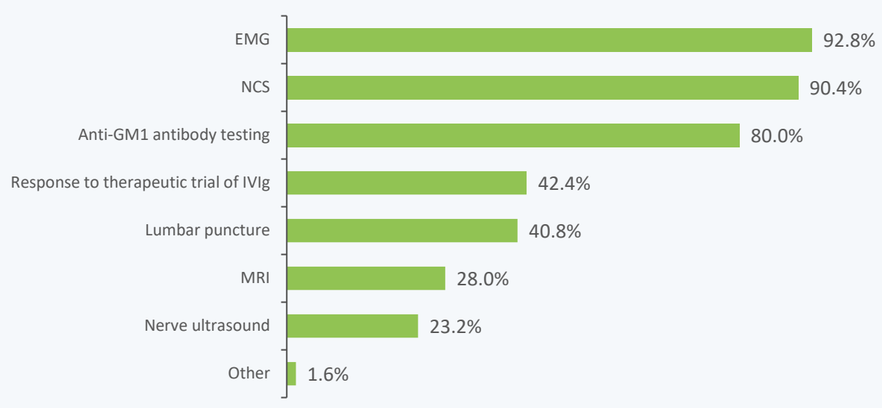
Initial diagnosis	Total, % (N=125)	US,% (n=46)	OUS, % (n=79)
Peripheral neuropathy	70.4	76.1	67.1
Compression neuropathies*	53.6	43.5	59.5
CIDP	51.2	47.8	53.2
ALS	50.4	47.8	51.9
GBS	32.8	32.6	32.9
Muscle disorder†	31.2	45.7	22.8
NMJ disorder‡	26.4	19.6	30.4
Hirayama disease	16.8	10.9	20.3

*Including carpal tunnel syndrome or ulnar neuropathy. †Including muscular dystrophy. ‡Including MG

- Of the measures available to evaluate and monitor patients with MMN, over **60% of neurologists** report using **neurological examination**



- To confirm a diagnosis of MMN, over **90% of neurologists** report using **EMG and/or NCS**, and **80% report** using **anti-GM1 antibody testing**

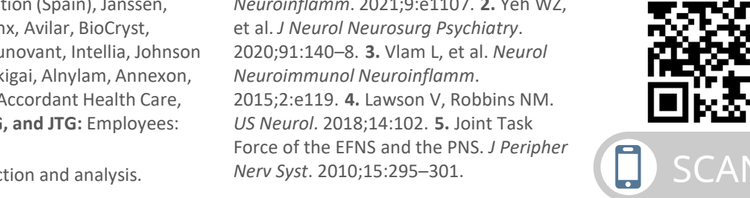


Treatment Decisions: Respondents Following Treatment Guidelines

- Respondents report using **IVIg as 1L therapy for 83%** and **SCIg therapy for 23%** of their patients with MMN
- IVIg was reported to be used at a mean induction effective dose of 1.8 g/kg and a mean maintenance dose of 1.2 g/kg
- SCIg was reported to be used at a mean induction effective dose of 2.8 g/kg and a mean maintenance dose of 1.6 g/kg
- Over the past 12 months, respondents reported switching an average **16.7% of their patients** with MMN to **2L therapy**
- Over **20% of respondents** report using **CS as 2L therapy** despite recommendations against their use

Top 5 2L Therapies	Total (n=125)
Rituximab	36.0%
PLEX	23.2%
CS	**21.6%**
SCIg	19.2%
Cyclophosphamide	18.4%

- Over three-quarters of respondents reported switching patients with MMN to 2L therapy due to lack of response to IVIg



*Reasons include a preferred treatment regimen (4.8%), patient development of antibodies (0.8%), new evidence/guidelines for treatment (1.6%), and change in patient insurance coverage (0.8%)

KEY TAKEAWAYS

Almost one-third of respondents report not consulting treatment guidelines and using CS as 1L treatment despite recommendations against their use

70% of neurologists using IVIg induction doses between 1 and 2 g/kg and not using CS as 1L report their patients have been previously diagnosed with another peripheral neuropathy

Over 75% of these neurologists report 1L treatments are ineffective in some patients

This survey highlights the unmet need for greater awareness of diagnostic and treatment guidelines in MMN

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ABBREVIATIONS

1L, first line; 2L, second line; AAN, American Academy of Neurology; AANEM, American Association of Neuromuscular and Electrodiagnostic Medicine; ALS, amyotrophic lateral sclerosis; CAP-PRI, Combined Assessment of Function and Performance Index; CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; CS, corticosteroids; DGN, Deutsche Gesellschaft für Neurologie (German Neurological Society); EFNS, European Federation of Neurological Societies; EMG, electromyography; GBS, Guillain-Barré Syndrome; INCAT, Inflammatory Neuropathy Cause and Treatment Group; IVIg, intravenous immunoglobulin; MG, myasthenia gravis; MMN, multifocal motor neuropathy; MND, motor neuron diseases; MRC, Medical Research Council; MRI, magnetic resonance imaging; NCS, nerve conduction studies; NMJ, neuromuscular junction; OUS, outside the US; PLEX, plasma exchange; PNS, Peripheral Nerve Society; RODS, Rasch-built Overall Disability Scale; SCIg, subcutaneous immunoglobulin; TUG, Timed Up and Go; US, United States.

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REFERENCES

1. Budding K, et al. *Neurol Neuroimmunol Neuroinflamm*. 2021;9:e1107. **2.** Yeh WZ, et al. *J Neurol Neurosurg Psychiatry*. 2020;91:140–8. **3.** Vlam L, et al. *Neurol Neuroimmunol Neuroinflamm*. 2015;2:e119. **4.** Lawson V, Robbins NM. *US Neurol*. 2018;14:102. **5.** Joint Task Force of the EFNS and the PNS. *J Peripher Nerv Syst*. 2010;15:295–301.

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