Supplemental data 1

Table e-1. Overview of clinical data excluded from the 52 publications identified in the systematic literature search

Study type, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM, IgM paraprotein, total IgM	Clinical outcome measures	Comment
Retrospective study, Codron *et al*. 2017 [16]	Plasma exchange (n=9)	No information available	**Responder (2/9)** Improvements in Hughes score **Non-responder (7/9)** No improvements in Hughes score	No anti-MAG IgM titres or paraprotein levels were measured. Short term reduction can be anticipated as patients underwent plasmapheresis cycles.
Retrospective and prospective study, Svahn *et al*. 2017 [17]	Various treatment interventions (n=202)	No information available	No information regarding change of the anti-MAG IgM levels and the clinical outcome measurements.	Detection of anti-MAG IgM was performed before treatment in 186 patients but only in 16 patients after treatment.
Case study, Noronha *et al*. 2006 [21]	Rituximab (n=1)	+30% paraprotein	**Acute deteriorating (1/1)** Flair in neuropathy	Waldenstrom's macroglobulinemia patient.
Case study, Rudnicki *et al*. 1998 [22]	Autologous bone marrow (n=1)	-99% in anti-MAG IgM titers	**Responder (1/1)** Fast electrophysiological response, slow symptomatic improvements	Waldenstrom's macroglobulinemia patient with atypical parkinsonism.
Placebo controlled, double blind and open label crossover study, Dyck *et al*. 1991 [18]	Plasma exchange (n=11) Sham exchange (n=10)	No information available	Clinical improvements observed in the patients. However, they did not reach significant in the PE group compare the sham exchange.	No anti-MAG IgM titres or paraprotein levels were measured. Short term reduction can be anticipated as patients underwent PE cycles.
Open label study, Oksenhendler *et al*. 1995 [19]	Chlorambucil (n=22) Chlorambucil and PE (n=22)	Limited information available	**Responder (8/22)** • Improvements in self-reported outcome **Non-Responder (14/22)** • Worsening in self-reported outcome (n=8) • Stabilization (n=6) **Responder (7/22)** • Improvements in self-reported outcome **Non-Responder (15/22)** • Worsening in self-reported outcome (n=7) • Stabilization (n=8)	PE seemed to confer no additional benefit in the treatment of polyneuropathy associated with monoclonal IgM.

Study type, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM, IgM paraprotein, total IgM	Clinical outcome measures	Comment
Randomized, crossover, placebo controlled trial, Comi *et al*. 2002 [20]	IVIg, placebo (n=11)	No information available	**IVIg phase** • Responder (10/22) • Non-responder (12/22), stable n=11, deteriorated n=1	Only modest benefit of IVIg in a minority of patients.
	Placebo, IVIg (n=11)		**Placebo phase** • Responder (4/22) • Non-responder (18/22), stable n=14, deteriorated n=4	
Open label study, Ellie *et al*. 1995 [23]	Various, PE, prednisone, IVIg, cytotoxic drugs (n=33)	Limited information available	**Responder (22/37)** • Only mild and transient improvements	Only modest benefit independent from the treatment. Four patients died during the follow-up phase.
			Non-responder (11/37) • No treatment response or worsening	

Table e-2. Overview of clinical data extracted from the 50 publications identified in the systematic literature search.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Retrospective study, Class VI, Pestronk *et al*. 2003 [24]*	Rituximab (n=7)	-57%	NR	NR	**Responder (7/7)** • Improvements in strength (+24%)	**Response** • 6 months (1st FU)	**Supportive** • Patients with other polyneuropathies were (e.g. anti-GM1 IgM) were included in the study as well.
	Placebo (n=5)	No change	NR	NR	**Non-Responder (5/5)** • No improvements (0%) in strength compare to pre-treatment after 24 months	**No response** • Stable for 24 months	
Double blind, placebo-controlled study, Class I Dalakas *et al*. 2009 [30]	Rituximab (n=13)	-50%	NR	-34%	**Responder (7/13)** • Improvements in INCAT (4/13) • Walking improved (7/13)	**Response** • 2 months (start to improve)	**Supportive** • Improvements would have been significant if one patient with a disability score of 0 at baseline was excluded.
	Placebo (n=13)	+37%	NR	+5%	**Non-Responder (13/13)** • No change in INCAT • No improvement in walking	**No response** • Stable for 8 months	
Open label study, Class IV, Gruson *et al*. 2011 [31]	Rituximab and fludarabine (n=2)	> -50%	-95%	NR	**Responder (2/2)** • Improvements in INCAT (-3.5) • Improvements in MCV (≥10%, range 10-50%) and decrease in DML (≥10%, range 10-25%)	**Response** • 6 months (end of treatment)	**Supportive** • One patient had baseline values of >70,000 BTU and the post treatment levels were 65,000 BTU. Therefore the actual reduction would be higher.
Case study, Class IV Weiss *et al*. 2014 [32]	Rituximab (n=1)	+404%	NR	+34%	**Acute deteriorating (1/1)** • Neurological deterioration (sensory ataxia and impaired ambulation) • Acute IgM flare	**Worsening** • 2 weeks	**Supportive** • Serological and neurological parameters returned to baseline after 6 weeks.
Case study, Class IV, Sala *et al*. 2014 [33]	Rituximab (n=3)	+440%	NR	NR	**Acute deteriorating (3/3)** • Deterioration in INCAT (+3.5) • Increased distal latencies and reduced MCV and cMAP	**Worsening** • 2 weeks	**Supportive** • Deterioration was reversible within some weeks to several months.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Open label study, Class IV, Baron *et al*. 2017 [34]	Plasma exchange (PE) (n=4)	-54%	-69%	NR	**Responder (4/4)** • Improvements in ONLS (-3)	**Response** • 1-2 months (1-6 PE)	**Supportive** • Plasma exchange was performed in anti-MAG patients after acute deterioration. • One patient showed immediate response to PE.
Open label study, Class IV, Levine *et al*. 1999 [25]*	Rituximab (n=1)	More than -50%	NR	NR	**Responder (1/1)** • Improvements in strength index (+20%)	**Response** • 3 months	**Supportive** • Only 1 anti-MAG neuropathy patient was included in the study.
Open label study, Class IV, Renaud *et al*. 2003 [35]	Rituximab (n=6)	More than -52%	NR	-58%	**Responder (5/6)** • Improvements in NDS (more than -3 points) • Increase in ulnar MCV	**Response** • 6-12 months (NDS)	**Supportive** • One patient was deteriorating, but was excluded due to severe occlusive arterial disease.
		-25%	NR	No change	**Non-responder (1/6)** • Stabilization in NDS • Decrease in ulnar MCV	**No response** • 12 months	
Follow up, open label study, Class IV, Renaud *et al*. 2006 [36] (responder of the previous study [35])	Rituximab (n=8)	-59% (median)	NR	-74% (median)	**Responder (6/8)** • Improvements in NDS • Improvements motor nerve conduction velocity by ≥10%	**Response** • 12 months	**Supportive** • One patient that did not respond to the low dose but did respond to the high rituximab dose (reduction of the titers). Unclear if improvements occurred before the FU at 12-month. • Two patients with Waldemstöm or Non-Hodgkin Lymphoma are included in this cohort.
					Non-responder (2/8) • Stabilization in NDS, n=1 • Deterioration in NDS (+2), n=1	**No response** • 12 months	

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Open label study, Class IV, Benedetti *et al*. 2007 [37]	Rituximab (n=7)	-87%	NR	-39%	**Responder (5/7)** • Improvements in ISS • Clinical improvement did not always correlate with electrophysiological improvement (MCV, DML, cMAP). • Electrophysiological improvement was usually more evident in the ulnar nerve than in the peroneal nerve.	**Response** • 12 months	**Supportive** • Improvements in ISS (1.9 point), as well improvements in MRC sum score and INCAT disability score, but not significant. Unclear if patients exhibited signs of improvements at earlier time points. • Deteriorating patient showed no chance in anti-MAG levels.
		-48%	NR	-2%	**Non-responder (2/7)** • Stabilization in ISS, MRC, INCAT, n=1 • Deterioration in ISS, MRC, INCAT, n=1	**No response** • 12 months	
Follow up open label study, Class IV, Benedetti *et al*. 2008 [38] (responder of the previous study [37])	No treatment (n=9)	-80%	NR	-40%	**Sustained responder (5/9)** • Improvements in INCAT (-1.2)	**Response** • Persistent for 24 months in 80% • Persistent for 36 months in 60%	**Supportive** • Deterioration coincided with or followed an anti-MAG IgM titers increase. • Not clear if all MGUS patients were included in the follow-up study.
		-20%	NR		**Transient responder (4/9)** • Deterioration in INCAT (+0.759)		
Open label study, Class IV, Kilidireas *et al*. 2006 [39]	Rituximab (n=2)	NR	-50%	NR	**Responder (1/2)** • Improvements in hand grip • Improvements in MRC • Improvements in10 m walk test • Increase in MNCV, SNCV at 6 weeks • Increase in cMAP, SNAP at 6 weeks	**Response** • 6 weeks	**Supportive** • Transient worsening of MRC in a patient 3 weeks after initiation of rituximab coincided with an IgM flair. Only SGPG and not MAG reactivity was assessed.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
		NR	No reduction	NR	**Non-responder (1/2)** • Stabilization in MRC • Decrease in MNCV, SNCV at 12 months • Increase in cMAP, SNAP at 12 months	**No response** • 12 months	
Open label study, Class IV, Souayah *et al*. 2013 [40]	Rituximab (n=3)	More than -90%	NR	NR	**Responder (2/2)** • Improvements in TNS (-10) • Only in one patient improvements in the nerve conduction studies were observed	**Response** • 2-6 moths	**Supportive** • Post-analysis was only done for 2 of 3 patients
Double blind, placebo controlled study, Class I, Leger *et al*. 2013 [41]	Rituximab (n=26)	-20% (median)	NR	NR	**Primary outcome: Non-responder (20/20)** • No significant difference in ISS compare to placebo) **Secondary outcome: responder (5/20)** • Improvements in INCAT disability score, n=4 (≥2) • Self-evaluated improvements (n=5)	**Response** • 12 months (1st FU)	**Partly supportive** • Withdrawal: n=6 rituximab, n=1 placebo. Typically, a reduction of anti-MAG IgM of at least around 50% is considered necessary for clinical improvements, which may explain the lack of clinical effect in this study[23].
	Placebo (n=28)	0% (median)	NR	NR	**Non-responder (27/27)** • No significant change in ISS • No change in INCAT disability score • No change in SF-36 questionnaire	**No Response** • 12 months	
Follow up study, Class I, Ferfoglia *et al*. 2016 [42] (Patients of the previous study [41])	Group 1 (2/7 rituximab and 5/7 no treatment) (n=7)	+6%	NR	NR	**Comparison of Group 1 (7/7) and Group 2 (8/8)** • No significant difference in ISS • No significant difference in INCAT disability score	Median FU 6 months	**Not applicable** • Cross-over design makes it challenging to assess the responder to the treatment. • Withdrawal: n=1 group 1, n=2 group 2. The authors

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
• Group 1: previously rituximab (n=8) • Group 2 previously placebo (n=10)	Group 2 (6/8 rituximab and 2/8 no treatment) (n=8)	-39%	NR	NR	• Worsening in the 10 meter walking test in Group 2		commented that considering the small number of patients and the heterogeneity of treatments during the FU period, they could not perform any comparison between the groups.
Retrospective study, Class IV, Hospital *et al*. 2013 [43]	Rituximab (n=26)	No change in anti-MAG IgM titres	NR	Reduction (in responder only)	**Responder (21/26)** • Improvements in mRS • **Non-responder (5/26)** • Stabilization in mRS, n=4 • Deterioration in mRS, n=1	**Response** • 9.5 months (median)	**Supportive** • IgM level felt only in responder • Anti-MAG IgM levels above the upper cut-off of the ELISA, therefore no difference was observed in the responder group. • Electrophysiological evaluation in 23 responders confirmed clinical improvements. • Significant improvements in mean median nerve distal latencies and cMAP of the peroneal nerve.
	Rituximab Combination (n=19)				**Responder (16/19**) • Improvements in mRS **Non-responder (3/19)** • Stabilization in mRS, n=2 • Deterioration in mRS, n=1	**Response** • 5 months (median)	
Open label study, Class IV, Gorson *et al*. 2001 [44]	Various treatment interventions (n=24) PE, IVIg, Prednisone, cyclophosphamide, PE and cyclophosphamide, INF-α2a chlorambucil, azathioprine	-11% (median)	-39% (median) -39% (mean)	-25% (median) -25% (mean)	**Sustained responder (4/24)** • Improvements in Rankin disability scale • Improvements in sensory score • Improvements in MRC (-1.4) • Only median motor nerve distal latency was more prolonged and the sural sensory nerve action potential was more often absent in responder and transient responders.	**Response** • 1-6 month • 4.8 years mean FU • 2.8 years median FU	**Supportive** • Due to frequent relapses or lack of a response, patients were treated with an average of three different modalities. The authors concluded that with a larger cohort (powered study) the difference would have been significant. Results in Table 1-3 are not consistent with the main text of the manuscript.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
		+29.3% (median)	+20% (median) +38% (mean)	+26% (median) +56% (mean)	**Transient responder (8/24)** • Transient Improvements in Rankin disability scale, sensory score, MRC • Improvements in MRC		
					Non-responders (12/24) • Deterioration in MRC (+0.5)	**No response** • 4.8 years mean FU • 2.8 years median FU	
Open label study, Class IV, Duncombe *et al*. 2017 [45]	Rituximab and cyclophosphamide (n=13)	-60%	-79%	NR	**Responder (13/13)** • Significant clinical improvements in ONLS and NCS	**Response** • 12 months (2nd FU)	**Supportive** • Unclear if a higher relative reduction in each single patient was associated with a better clinical outcome.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Open label study, Class IV, Nobile-Orazio *et al*. 1988 [46]	Chlorambucil (n=5)	-50%	NR	-54.5%	**Responder (2/5)** • Improvements in disability and ataxia score • Improvements in MCV and SNAP	**Response** • 2 months	**Supportive** • Non-responder did not show an alteration in the anti-MAG levels.
		No reduction	NR	-22.5%	**Non-Responder (3/5)** • No change in disability and ataxia score • Nerve conduction velocities were decreased in 2 non-responders	**No response** • 14 months	
Open label study, Class IV, Wilson *et al*. 1999 [26]*	Fludarabine (n=2)	NR	NR	-71.5%	**Responder (1/2)** • Improvements in mRS (-3) • Increase in median MCV and SAP	**Response** • 3 months	**Partly supportive** • No anti-MAG levels were measured
		NR	NR	-45%	**Non-responder (1/2)** • Stabilization in mRS • Increase in median MCV and SAP	**No response** • 6 months	
Retrospective study, Class VI, Campagnolo *et al*. 2017 [47]	Rituximab (n=25)	-60%	NR	NR	**Responder (15/25)** • Improvements in INCAT • Improvements in ISS	**No response** • 12 months (1st FU)	**Partly supportive** • Unclear if the patients with reduced anti-MAG levels where the same patients that showed clinical improvements.
					Non-responder (10/25) • No improvements in INCAT • No improvements in ISS	**No response** • 12 months (1st FU)	

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Prospective uncontrolled trial, Class VI, Niermeijer *et al*. 2006 [49]	Fludarabine (n=6)	NR	NR	-60%	**Responder (2/6)** • Improvements in raking scale • Median values of EMG variables did not change significantly after treatment • Tendency for improvements of the MCV (>10%)	**Response** • 12 months (1st FU)	**Partly supportive** • Patients exhibited a switch monoclonal to polyclonal (n=4), and vice-versa (n=1).
		NR	NR	-42.75%	**Non-responder (4/6**) • Stabilization in raking scale • Median values of EMG variables did not change significantly after treatment	**No response** • 12 months (1st FU)	
Double-blind randomized, placebo controlled study, Class I, Niermeijer *et al.* 2007 [48]	Cyclophos-phamide and prednisone (n=16)	NR	-94%	NR (pre-treatment level)	**Responder (5/15)** • Improvements in Rivermead mobility index o(≥1), n=5 • More improvements in the Secondary outcome measures, including Rankin scale, MRC, and sensory sum score	**Response** • 6 months (1st FU)	**Supportive** • Supportive as more than 50% of the patients (placebo& treatment) exhibit the expected result. • One patient in the treatment group stopped because of angina pectoris. • Beneficial effect on most secondary outcome measures for impairment in addition to biologic effects on the M protein concentration and nerve conduction after 6 months and on the MRC sum score thereafter.
	Placebo (n=19)	NR	+106%	NR (pre-treatment level)	**Non-responder (15/19)** • Improvements in Rivermead mobility index o(≥1), n=4 • More improvements in the Secondary outcome measures compare to the treatment group	**Response** • 6 months (1st FU)	
Open label study, Class IV, Kelly *et al*. 1988 [50]	Various treatment interventions (n=5)	NR	-40%	NR	**Responder (3/3)** • Improvements in NDS	**Response** • 3 months	**Supportive** • Two patients were excluded due to the development of severe comorbidities.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Open label study, Class IV, Haas *et al*. 1988 [e1]	Plasmapheresis (n=1)	NR	NR	-20%	**Responder (1/1)** • Improvements in MRC • Conduction velocity and distal latency did not change appreciably	**Response** • 1 month	**Supportive** • Case study of repeated plasmapheresis.
Open label study, Class IV, Blume *et al*. 1995 [e2]	Plasma exchange and IV cyclophos-phamide (n=4)	-78%	NR	NR	**Responder (4/4)** • Improvements in strength (+34%)	**Response** • 3-9 months (depending on the FU time)	**Supportive** • All patients showed improvements.
Prospective, randomised, open clinical trial, Class I, Mariette *et al*. 1997 [e3]	IFN-α treatment (n=10)	NR	More than -50 % (in two responder)	NR	**Responder (8/10)** • Improvements in CNDS (-7.5) **Non-responder (2/10)** • No change in CNDS	**Response** • 6 months (1st FU)	**Not supportive** • No significant decrease in IgM paraprotein was noted. The authors suggest that IFN-α decreases the permeability of the blood neve barriers and therefore, explain why 6 patients showed clinical improvements without lowering the total IgM. • The mean value of ulnar motor-nerve conduction velocities and distal latencies were not different between the two groups. • Due to the large number of patients with no SNAP at baseline in the two groups, it was impossible to compare the evolution of sensory nerve conduction velocities.
	IVIg treatment (n=10)	NR	No reduction	NR	**Responder (1/10)** • Improvements in CNDS (only transient) **Non-responder (9/10)** • Worsening in CNDS (+2.3)	**No response** • 6 months	
Open label study, Class IV, Rakocevic *et al*. 2018 [14]	Obinutuzumab (n=2)	-98%	NR	-58%	**Non-Responders (2/2)** • No improvement or worsening in neuropathic symptoms	**No response** • 6 months	**Not supportive** • The authors suggest due to the patients' advanced disease and severe axonal degeneration no clinical response was detected.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Case study, open label, Class IV, Stino *et al*. 2017 [e4]	Lenalidomide (n=1)	No reduction	-71%	NR	**Responder (1/1)** • Improvements in I-RODS (22%) • No improvements in INCAT • Mild to modest improvements in NCS (median and ulnar DML), MCV unchanged	**Response** • 7 months (1st FU)	**Supportive** • Anti-MAG IgM levels are above the upper detection limit. Therefore, a reduction cannot be detected by ELISA.
Case Study, Class IV, Doneddu *et al*. 2017 [27]*	Rituximab (n=2)	NR	+8.5%	NR	**Acute deteriorating (2/2)** • Worsening in MRC • Worsening of tremor • Evidence of severe demyelinating neuropathy with significantly prolonged distal latencies	**Worsening** • 2-4 weeks	**Supportive** • The pre-treatment anti-MAG titers were already above the threshold of the ELISA (70'000 BTU) or rituximab potentially increase the permeability of the blood-brain barrier, allowing enhance migrating of the anti-MAG IgM in the CNS.
Case study, Class IV, Gomez *et al*. 2016 [e5]	Bendamustine/ Rituximab (n=1)	-88%	NR	NR	**Responder (1/1)** • Improvements in strength and Romberg test	**Response** • 1 month	**Supportive** • One year after starting Bendamustine/Rituximab treatment, new worsening symptoms with evidence of progressive increase anti-MAG IgM.
Case study, Class IV, Vo *et al*. 2015 [e6]	Rituximab (n=1)	NR	NR	-44%	**Acute deteriorating (1/1)** • Worsening in MRC • Worsening in INCAT • Worsening in grip strength • Worsening of previously noted demyelinating abnormalities (DML, cMAP, CMV)	**Worsening** • 2 weeks	**Not supportive** • Anti-MAG IgM levels were not assessed post treatment but patient improved after IVIg treatment.
Open label study, Class IV, Talamo *et al*. 2015 [1]	Rituximab and plasma exchange, rituximab, fludarabine (n=4)	-75%	NR	-76%	**Responder (4/4)** • Symptomatic improvements	**Response** • 6 months	**Supportive** • Only in two treated patient the total IgM was assessed pre- and post-treatment. One responder did not exhibit

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
	Untreated (n=3)	No change	NR	No change	**Non-responder(3/3)** • Stable symptoms	**No response** • 6 or 12 months	increased IgM levels (pre-treatment).
Prospective, open label study, Class IV, Zara *et al*. 2011 [28]*	Rituximab (n=5)	-20% (-49% to +53% range)	NR	NR	**Responder (3/5)** • Improvements in INCAT disability scale • Improvements in ISS	**Response** • 12 months	**Not supportive** • The authors indicated that two patients had anti-MAG IgM levels above the upper cut-off of the ELISA and therefore, a potential reduction could not be detected. Figure 1C is not consistent with the main text of the manuscript. • There was no evident correlation between anti-MAG serum antibodies and the electrodiagnostic data (except for absent SAP). Nor was there a correlation with the clinical scales, the slowing of motor conduction, TLI or cMAP amplitude reductions.
		-20%	NR	NR	**Non-responder (2/5)** • No improvements in INCAT • No improvement in ISS	**No response** • 12 months	
Open label study, Class IV, Delmont *et al*. 2011 [e7]	Rituximab (n=3)	-43%	-31%	NR	**Responder (3/3)** • Improvements in ISS, n=3 • Improvement in OLNS, n=2 • Improvement in MRC, n=3 • No change in individual or overall electrophysiological data	**Response** • 9 months (ONLS) • 3 months (ISS)	**Supportive** • Not specified which patient did show no improvements.
Case study, Class IV, Stork *et al*. 2013 [e8]	Rituximab (n=3)	-48%	+14%	-9%	**Acute deteriorating (3/3)** • Rapid worsening in MRC • NCS worsened in two patients	**Worsening** • during 1st/2nd treatment cycle	**Not supportive** • The authors suggested that the worsening might be related to significant side effects of rituximab, as seen in other studies [13, e6, e7].

Study type, Class of evidence, Reference	**Treatment and Nr. of anti-MAG neuropathy patients (n)**	**Change in anti MAG IgM**	**Change in para-protein**	**Change in total IgM**	**Clinical outcomes measures**	**Time to response[A]**	**Supporting change in anti-MAG IgM and clinical symptoms correlation and comments**
Case study, Class IV, Broglio *et al*. 2005 [e9]	Rituximab (n=1)	No reduction	NR	-50%	**Non-responder (1/1)** • Worsening in MRC • Wheelchair-bound because of ataxia	**Worsening** • 2 months	**Supportive** • Authors suggested that the pathogenic anti-MAG IgM is produced by $CD20^-$ cells.
Case Study, Class IV, Gironi *et al*. 2006 [e10]	Rituximab (n=1)	+21%	NR	+58%	**Acute deteriorating (1/1)** • Severe worsening of all neurological signs (specifically tremor)	**Worsening** • 3 months	**Supportive** • Patient with Waldestrom macroglobulinemia and neuropathy associated with anti-MAG IgM/k antibodies.
Open label, Class IV, Briani *et al*. 2019 [13]	Obinutuzumab and chlorambucil (n=2)	> -92% (n=1)	-45% (n=1)	-55% (n=1)	**Responder (2/2)** • Improvements in MRC and INCAT (-1) • Neurophysiology improved	**Response** • 3-6 months	**Supportive** • Patients had anti-MAG antibody neuropathy and concurrent chronic lymphocytic leukaemia. Both patients developed neutropenia and one a fatal pneumonia. • Patient had baseline values of >70,000 BTU, therefore the actual reduction would be higher • Only limited data are available from both patients
Case study, Class IV, Al-Bustani *et al*. 2016 [e11]	Rituximab (n=1)	-97%	-100%	-42%	**Responder (1/1)** • Improvements in NCS • Electrodiagnostic testing correlated with clinical improvement	**Response** • 1 month	**Supportive** • Clinical improvements were still persistent 7 years after first treatment.
Prospective pilot study, Class IV, Delarue *et al*. 2004 [e12]	Rituximab (n=4)	No reduction	No reduction	NR	**Non-Responder (4/4)** • No improvements of clinical neurological symptoms	**No response** • 24 months FU (median)	**Supportive** • One patient exhibited later improvements after high dose Melphalan followed by autologous stem cell transplantation.

Study type, Class of evidence, Reference	**Treatment and Nr. of anti-MAG neuropathy patients (n)**	**Change in anti MAG IgM**	**Change in para-protein**	**Change in total IgM**	**Clinical outcomes measures**	**Time to response[A]**	**Supporting change in anti-MAG IgM and clinical symptoms correlation and comments**
Prospective study, Class IV, Benedetti *et al*. 2019 [e13]	Rituximab (n=18)	-67% (after 31/46 rituximab cycles)	NR	NR	**Responder (16/18)** • Improvements in INCAT disability scale • Improvements in MRC sum score • Improvements in ISS	**Response** • Clinical improvements after the first rituximab cycles lasted two or more years	**Supportive** • No maintenance therapy was performed, unless patients exhibited relapses, then additional rituximab cycles wereused. One responder showed an increase in 10% anti-MAG IgM.
		+13% (+0% to +25% range)	NR	NR	**Non-responder (2/18)** • No change in INCAT disability scale • No change MRC sum score • No change in ISS	**No response** • Time of FU is unclear	
Uncontrolled open study, Class III, Hamidou *et al*. 2005 [e14]	Cyclo-phosphamide (n=9)	-7%	NR	-56%	**Responder (7/9)** • Improvements in Ranking scale • Improvements in muscle strength • No significant changes in the electrophysiological measures	**Response** • 6 months (1st FU)	**Supportive** • All patients showed improvements in muscle strength and a significant reduction in total IgM
		-3%	NR	-49%	**Non-responder (2/9)** • Stabilisation in Raking scale • Improvements in muscle strength (n=2) • No significant changes in the electrophysiological measures	**No response** • Stable over 18 months	
Case study, Class IV, Ghosh *et al*. 2002 [e15]	Cladribine (n=1)	-94%	disappearance of the IgM paraprotein	NR	**Responder (1/1)** • From effectively useless hands to grip objects, open hold a cup of coffee. • Able to climb stairs again and stand from a chair unaided. Walking improved.	**Response** • 10 months	**Supportive** • Disappearance of paraprotein and sustainable anti-MAG IgM reduction coincided with clinical improvements.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Open label study, Class III, Notermans *et al*. 1996 [e16]	Cyclo-phosphamide, prednisone (n=5)	NR	-56% (n=1, too low to quantify in n=4)	NR	**Responder (5/5)** • Reduction of bone marrow infiltration • Ulnar nerve conduction variables (DML, MCV, CMAP) were significantly better than before treatment	**Response** • 6 months (1st FU)	**Supportive** • Paraprotein was too low to quantify in 4 patients • Unclear if the clinical improvements occurred in the anti-MAG IgM MGUS cohort.
Open label study, Class IV, Notermans *et al*. 1997 [e17]	Dexamethasone (n=5)	NR	-40% (n=3, pre-treatment IgM too low to quantify in n=2)	NR	**Responder (5/5)** • Improvements in motor sum score • Improvements in disability scale	**Response** • 3-6 months	**Supportive** • Very high frequency of serious invalidating side effects occurred due to the treatment. • One patient showed the clinical improvements and paraprotein reduction (-60%) only after cyclophosphamide therapy
Case study, Class IV, Niemierko *et al*. 1999 [e18]	Immunu-adsorbption (Protein A column) (n=1)	NR	No reduction	NR	**Responder (1/1)** • Improvements in motor functional score (+2) • Improvements in gait, balance, and strength	**Response** • NR, potentially data were assessed at the quarterly treatment cycles.	**Not supportive** • 2nd IgM MGUS patient was included, however the reactivity of the paraprotein was nor reported. • As Prosorbat columns mainly remove IgG (95%) and only 30% of IgM, the authors suggest that reduced complement and/or enhanced clearance of soluble immune complexes may have occurred [e17, e18].

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
Case study, Class IV, Ernerudh *et al*. 1986 [e19]	Plasma exchange (n=3)	NR	NR	Approx. -90% (n=1)	**Responder (2/3)** • Improvements in muscle strength and vibration sense • Increase of motor conduction velocity • Painful paraesthesia disappeared • Only NCS improvements in the arm of one patient	**Response:** • 4-6 weeks	**Supportive** • Slight clinical deterioration occurred 3 and 10 months after treatment. • PE of the non-responder was stopped due to low IgG levels
		NR	NR	Approx. -60%	**Non-responder (1/3)** • No clinical or neurophysiological chance	**Non-response** • Non clinical improvements after 5 PE runs	
Open study, Class IV, Ernerudh *et al*. 1992 [e20]	Various treatment (n=5) Plasma exchange, chlorambucil, prednisolone, melphalan	Approx. -60% (reduction 2=n, and increase n=1)	NR	NR	**Responder (3/5)** • Improvements in motor function of hands • Improvements in muscle weakness score • Disability score	**Response** • 1-6 months	**Supportive** • In 3 patients there was clear correlation between clinical effect and IgM concentration. In 2 patients improvement corresponded to decrease and in 1 patient clinical status as well as antibody concentration was unchanged. • In 2 patients, there was no clear correlation (1 patient improved despite unchanged or increased antibodies and 1 patient did not improve despite lowered antibody concentrations.
		NR, (no reduction n=1, reduction n=1)	NR	NR	**Non-responder (2/5)** • No change in disability status and sensory status, as well as muscle weakness score	**Non-response** • NR	
Randomized, placebo controlled study, Class II, Dalakas *et al*. 1996 [e21]	IVIg (n=11)	Transiently decrease (approx. -50%)	NR	NR	**Responder (1/9)** • Increase in strength based on MRC • The electrophysiological findings remained unchanged	**Response** • 2 months	**Supportive** • Anti-MAG IgM did not appreciably change and only two patients modestly improved.

Study type, Class of evidence, Reference	Treatment and Nr. of anti-MAG neuropathy patients (n)	Change in anti MAG IgM	Change in para-protein	Change in total IgM	Clinical outcomes measures	Time to response[A]	Supporting change in anti-MAG IgM and clinical symptoms correlation and comments
		No change	NR	NR	**Non-responder (8/9)** • No change in MRC • No clinically functional improvements • The electrophysiological findings remained unchanged	**Non-response** • Stable for 6 months	• Two patients were excluded as the anti-MAG reactivity couldn't be confirmed.
Open label study, Class IV, Sherman *et al*. 1984 [29]*	Plasma exchange (n=6)	-75%	-67%	NR	**Responder (2/6)** • Able to walk again with a walker • Able to extend wrist against gravity against gravity • No change in the electrophysiological studies despite improvement	**Response** • 1-2 weeks	**Supportive** • PE should be performed frequently enough to maintain the antibody titre at less than 50% of pre-treatment values
		-41%	-33.3%	NR	**Non-responder (4/6)** • No change, n=3 • Worsening of weakness, n=1 • No change in the electrophysiological studies		

*Hand selected publications; [A]After initiation of treatment; BTU: Bühlmann Titer Units; cMAP: compound motor action potential amplitude; CNDS: clinical neuropathy disability score; DML distal motor latency; F: Female; FU: Follow-up; INCAT: Inflammatory Neuropathy Cause and Treatment disability score; ISS: INCAT Sensory Score; I-RODS: Inflammatory Rasch-built Overall Disability Scale; M: Male; MCV: motor nerve conduction; MNCV: motor nerve conduction velocity; MRC: Medical Research Council sum score; mRS: modified Rankin Score; NDS: Neuropathy Disability Score; NR: not reported; OLNS: Overall Neuropathy Limitations Scale; SNAP: sensory nerve action potential; SNCV: sensory nerve conduction velocity; TLI: terminal latency index; TNS: Total Neuropathy Score

Table e-3. Overview of the participants from the 50 publications identified in the systematic literature search

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Pestronk *et al*. 2003 [24]	Responder (n=7) Treatment group	NR	NR	NR	NR	ELISA, immunofixation	**Anti-MAG IgM titers** In percentage of initial values, **Total IgM** In percentage of initial values,	• Instability of gait • Reduction of strength: 57% (4% SEM)
	Non-Responder (n=5) Control group	NR	NR	NR	NR		**Anti-MAG IgM titers** In percentage of initial values **Total IgM** In percentage of initial values	• Instability of gait • Reduction of strength: 63% (6% SEM)
Dalakas *et al*. 2009 [30], Treatment group	Responder (n=7)	66.8 (±7.9 SD)	12.9 (±7.2 SD) (mean disease duration)	2	11	Serum protein electrophoresis with immunofixation electrophoresis	**Anti-MAG IgM titers** 38.8 units/ml (±57.5 SD) **Total IgM** 599 mg/dl (±526 SD)	• INCAT: leg score: 1.46 (±1.0 SD) • 10m walk 8.3 sec (±3.2 SD) • MRC scale score: 134.6 (±11.9 SD) • Sensory score: 7.5 (±3.6 SD)
	Non-Responder (n=5)							
Dalakas *et al*. 2009 [30], Placebo group	Non-Responder (n=13)	67.6 (±8.4 SD)	12.9 (±6.5 SD) (mean disease duration)	7	6	Serum protein electrophoresis with immunofixation electrophoresis	**Anti-MAG IgM titers** 31.7 units/ml (±51.4 SD) **Total IgM** 698.5 mg/dl (±446 SD)	• INCAT: leg score: 1.45 (±0.7 SD) • 10m walk 9.5 sec (±4.2 SD) • MRC scale score: 131.6 (±11.2 SD) • Sensory score: 7.9 (±3.1 SD)
Gruson *et al*. 2011 [31]	Responder (n=2)	65 (64-66)	64 (63-65)	0	2	Electrophoresis, immunofixation, ELISA	**Anti-MAG IgM titers** 62’500 BTU (55’000 - >70’000)	• INCAT: 4 • Assessment of MCV, DML (ulnar, peroneal)

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Weiss *et al*. 2014 [32]	Acute deteriorating (n=1)	85	83	0	1	Serum protein electrophoresis, ELISA	**Anti-MAG IgM titers** 12’800 BTU **Paraprotein** Too small to detect **Total IgM** 190 mg/dl	• Advancing numbness in feet and imbalance • Stocking sensory loss • Mild sway with Romberg testing • Prolonged DML in upper and lower extremities • Reductions of MCV in the lower extremities • No motor conduction block
Sala *et al*. 2014 [33]	Acute deteriorating (n=3)	66 (63-69)	64.7 (62-67)	1	2	ELISA, total IgM NR	**Anti-MAG IgM titers** 50’461 BTU (1’366-86’567) **Total IgM** 4.64 g/dl (3.3-5.61)	• INCAT: 2 (1-3) • Leg paraesthesia, progressive ataxia, unsteadiness • MCV, DML, and cMAP in the peroneal, ulnar, and median nerve were assessed.
Baron *et al*. 2017 [34]	Responder (n=4)	68.5 (61-78)	63.5 (60-66)	1	3	ELISA, Paraprotein NR	**Anti-MAG IgM titers** 25’550 (18’600-38’943) **Paraprotein** 4.075 g/L (0-9.5)	• ONLS: 4.25 (2-6) • Ataxia, paraesthesia, tremor • Electromyogram was used to determine the characteristics of the neuropathy
Levine *et al*. 1999 [35]	Responder (n=1)	NR	NR	1	0	ELISA, serum immunofixation,	**Anti-MAG IgM titers** Only relative reduction reported **Total IgM** Only relative reduction reported	• Sensory loss, weakness • Reduced strength index (-20%)

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Renaud *et al*. 2003 [36]	Responder (n=5)	60 (48-77)	56 (42-75)	2	3	ELISA, immune electrophoresis	**Anti-MAG IgM titers** Only relative reduction of baseline shown **Total IgM** 1.5-15 g/L	• Only change in NSS shown • NDS: 30-70 • TLI >0.25 • Assessment of the Ulnar MCV
	Non-Responder (n=1)	73	66	0	1		**Anti-MAG IgM titers** Only relative reduction of baseline shown **Total IgM** Approx. 4 g/L	• Only change in NSS shown • NDS: approx. 36 • Assessment of the Ulnar MCV
Benedetti *et al*. 2007 [37]	Responder (n=5)	61.8 (53-69)	59.4 (51-68)	3	2	Western blot	**Anti-MAG IgM titers** 1:31'680 (1'600-51'200) **Total IgM** 495 mg/dl (300-887)	• ISS: 9.4 (9-11) • MRC: 56 (46-59) • INCAT: 3.6 (2-8)
	Non-Responder (n=2)	61.5 (62-62)	58.5 (57-60)	0	2	Western blot, ELISA	**Anti-MAG IgM titers** 1:435'000 (70'000-800'000) **Total IgM** 600 mg/dl	• ISS:10 (8-12) • MRC: 55 (54-56) • INCAT: 3 (2-4) • MCV, DML, cMAP was assessed in the peroneal and ulnar nerve

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Kilidireas *et al*. 2006 [39]	Responder (n=1)	75	73	0	1	ELISA, paraprotein NR	**Anti-MAG IgM titers** Only SGPG reactivity was assessed, but classified as anti-MAG neuropathy **Paraprotein** 341 mg/L	• 9 peg hole test: 21.3 (R), 22.8 (L) • Hand grip: 56 (R), 56 (L) • MRC: 60 • 10m walk: 6.3 • Assessment of the MNCV, SNCV, cMAP, SNAP in the ulnar nerve
	Non-Responder (n=1)	60	58	0	1		**Anti-MAG IgM titers** Only SGPG reactivity was assessed, but classified as anti-MAG neuropathy **Paraprotein** 528 mg/L	• 9 peg hole test: 24.2 (R), 21.9 (L) • Hand grip: 86 (R), 82 (L) • MRC: 56 • 10m walk: 8.2 • Assessment of the MNCV, SNCV, cMAP, SNAP in the ulnar nerve
Souayah et al. 2013 [40]	Responder (n=2)	67.5 (62-73)	57 (53-61)	0	2	Anti-MAG IgM titers NR	**Anti-MAG IgM titers** 32’000 (12’800-51’200)	• Total neuropathic score: 14/36 • Assessment of DML, cMAP
Leger *et al*. 2013 [41], Treatment group	Responder (n=5)	64.6 (±8.6 SD	3.3 (1.4-4.8) median disease duration	8	18	ELISA, immunofixation and monoclonal protein according to standard procedures	**Anti-MAG IgM titers** ≥70’000 median (33’000-≥70’000) **Paraprotein** 6.9 g/L (4.2 SD), n=10 **Total IgM** 3.1 g/L (2.0-7.7), n=21	• INCAT disability score: 3 (2-4) • Median ISS: 6.5 (5-9) • 10m walk: 7.7 (6.0-10.7) • MRC: 56.5 (45-60)
	Non-responder (n=21)							

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Leger *et al*. 2013 [41], Placebo group	Non-responder (n=28)	67.2 (±8.6 SD)	3.8 (2.2-7.9) median disease duration	8	20	ELISA, immunofixation and monoclonal protein according to standard procedures	**Anti-MAG IgM titers** ≥70'000 median (14'000-≥70'000) **Paraprotein** 5.7 g/L (±2.9 SD), n=7 **Total IgM** 3.8 g/L (3.0-6.8), n=25	• INCAT disability score:3 (2-4) • Median ISS: 8 (6-10) • 10m walk: 9.0 (7.5-12.1) • MRC: 55 (51.5-60)
Hospital *et al*. 2013 [43] Rituximab treatment	Responder (n=21)	67 (47-86)	NR	12	14	ELISA, Paraprotein NR	**Anti-MAG IgM titers** 61'000 BTU (5'800->70'000) **Paraprotein** 0.35 g/L (0-1.52)	• mRS: 2.9 (2-5) • Sensory deficit, pain, ataxia, Motor deficit • Assessment of nerve distal latencies and cMAP
	Non-responder (n=5)							
Hospital *et al*. 2013 [43] Rituximab combination treatment	Responder (n=16)	68 (42-85)	NR	7	12	ELISA, Paraprotein NR	**Anti-MAG IgM titers** 60'000 BTU (1'000->70'000) **Paraprotein** 0.38 g/L (0-1.8)	• mRS: 2 (1-4) • Sensory deficit, pain, ataxia, motor deficit • Assessment of nerve distal latencies and cMAP
	Non-responder (n=3)							

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Gorson *et al*. 2001 [44]	Sustained responder (n=4)	64 (42-88)	2.5 (0.5-27) median disease duration	9	15	ELISA, serum immune-electrophoresis or immunofixation (e.g. high-resolution agarose gel technique or nephelometry)	**Anti-MAG IgM titers** 1:57'480 (6'400-1'600'000) **Paraprotein** 996 mg/dL (224-2'530)	• MRC: 36.3 (32-40) • Sensory score: 13.7 (8-22) • Ranking score: 2.7 (2-3) • Median, ulnar, peroneal, and tibial motor nerves and median, ulnar, and sural sensory nerves were sampled.
	Transient responder (n=8)						**Anti-MAG IgM titers** 1:309'605 (12'800-400'000) **Paraprotein** 624 mg/dL (69-2'083)	
	Non-responder (n=12)							• MRC: 37.2 (24-40) • Sensory score: 13.3 (6-24) • Ranking score: 2 (1-4) • Electrophysiological assessment see responder group
Duncombe *et al*. 2017 [45]	Responder (n=13)	NR	NR	NR	NR	NR	**Anti-MAG IgM titers** 38'925 (median) **Paraprotein** 4.7 g/L (median)	• ONLS: 3 (median) • MRC sum score: 76 (median, n=18)
Nobile-Orazio *et al*. 1988 [46]	Responder (n=2)	61 (60-62)	59	0	2	ELISA, total IgM NR	**Anti-MAG IgM titer** 7.85 (6.8-8.9, normalized value >3) **Total IgM** 0.95 g/L (0.8-1.1)	• Disability score: 2 (1-3) • Ataxia score: 1 (0-2) • Assessment of MCV (median, peroneal) and SNAP (median, sural)
	Non-Responder (n=3)	65 (54-72)	62 (53-69)	0	3	ELISA	**Anti-MAG IgM titers** 13.3 (9.8-19.5, normalized value >3) **Total IgM** 1.53 g/L (1-2)	• Disability score: 2.7 (2-3) • Ataxia score: 3.7 (3-4) • Assessment of MCV (median, peroneal) and SNAP (median, sural)

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Wilson *et al*. 1999 [26]	Responder (n=1)	45	41	1	0	Protein electrophoresis and quantified by densitometry	**Paraprotein** 7 g/l	• mRS: 4 • MRC sum score 56 • Sensory sum score: 4 • 10-meter walk 15s (with one stick) • Median MCV and SAP were assessed
	Non-Responder (n=1)	53	47	0	1		**Paraprotein** 5 g/l	• mRS: 2 • MRC sum score: 63 • Sensory sum score: 12 • 10-meter walk 7.1 • Median MCV and SAP were assessed
Campagnolo *et al*. 2017 [47]	Responder (n=15)	60.7 (44-72)	56.7 (40-68	7	8	Western blot, ELISA, total IgM NR	**Anti-MAG IgM titers** 52’480 BTU (10’000-100’000) **Total IgM** 3.2 g/L (1.6-7.9)	• INCAT: 2.7 (1-6) • ISS: 7.9 (1-18) • MRC: 56.3 (40-60)
	Non-Responder (n=10)	65.1 (49-87)	59.8 (47-71)	2	8		**Anti-MAG IgM titers** 141’525 BTU (7’500-800’000) **Total IgM** 3.3 g/L (1.08-6)	• INCAT: 2.5 (1-5) • ISS: 10.25 (2-18) • MRC: 57.1 (52-60)
Niermeijer *et al*. 2006 [49]	Responder (n=2)	57 (53-61)	44	2	0	NR	**Paraprotein** 4.5 g/L (<1-8) **Total IgM** 14.5 g/L (6.4-21.6)	• Raking scale: 3 • Assessment of MCV
	Non-Responder (n=4)	67.5 (60-74)	57 (55-60)	0	4	NR	**Paraprotein** 7.5 g/L (<1-16) **Total IgM** 14.2 g/L (6.4-21.1)	• Ranking scale: 2.25 (2-3) • Assessment of MCV

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Niermeijer *et al*. 2007 [48] Treatment group	(n=16, anti-MAG IgM positive n=7)	64.3 (9.2 SD)	60.7 (9.3 SD)	3	13	Electrophoresis, immunofixation	**Paraprotein** 0.5 g/L (0.5–0.5) (interquartile range)	• Rivermead mobility index: 13.5 (12–14) • Rankin scale: 2 (2-3) • MRC sum score: 133 (123–138) • Sensory sum score: 39 (30–42)
Niermeijer *et al*. 2007 [48] Placebo group	(n=19, anti-MAG IgM positive n=10	64.2 (8.5 SD)	59 (9.8 SD)	11	8	Electrophoresis, immunofixation	**Paraprotein** 0.5 g/L (0.5–0.5) (interquartile range)	• Rivermead mobility index: 14 (12–14) • Rankin scale: 2 (2-3) • MRC sum score: 136 (131–140) • Sensory sum score: 40 (33-47)
Kelly *et al*. 1988 [50]	Responder (n=3)	59 (48-78)	28 (48-78) Disease duration in months	1	2	Western blot	**Paraprotein** 6.8 g/L (4.5-8.4)	• MRC distal legs and hands 4-4.5/5 • Weakness legs and hands • Only baseline electrophysiological assessments were performed
Haas *et al*. 1988 [e1]	Responder (n=1)	44	38	0	1	Serum immunofixation, immune-electrophoresis	**Paraprotein** 971 mg/dl	• Totally atrophic foot muscles (MRC 4- to 4+) • Assessment of the conduction velocity and distal latency of the median nerve

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Blume *et al*. 1995 [e2]	Responder (n=4)	54 (49-60)	52.8 (47-58)	1	3	ELISA, Western blot methods, serum immunofixation	**Anti-MAG IgM titers** 1:362'294 (5475-1'300'000)	• Strength in % of normal: 45% (10-85%) • Only baseline nerve conduction studies were performed (ulnar and sural never)l
Mariette *et al*. 1997 [e3] IFN-α treatment	Responder (n=8)	67 (60-67)	3.1 (0.3-6.1) Duration of the neuropathy	1	9	Immune-blotting on delipidated human myelin	**Paraprotein** Only relative reduction is reported	• Global score: 24.4 (±11.3 SD) • Motor score: 2.9 (±5.5 SD) • Sensory score: 16.0 (±5.7 SD) • Reflex score: 5.5 (±3.9 SD) • Assessment of cMAP, MNCV, distal latency, SNAP
	Non-responder (n=2)							
Mariette et al. 1997 [e3] IVIg treatment	Responder (n=1)	66 (52-85)	4.0 (0.4-17.8) Duration of the neuropathy	3	7	Immune-blotting on delipidated human myelin, total IgM NR	**Paraprotein** Only relative reduction is reported	• Global score: 28.7 (±11.5 SD) • Motor score: 3.5 (±3.3 SD) • Sensory score: 17.2 (±7.2 SD) • Reflex score: 8.0 (±4.0 SD) • Assessment of cMAP, MNCV, distal latency, SNAP
	Non-responder (n=9)							

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Blume *et al*. 1995 [e2]	Responder (n=4)	54 (49-60)	52.8 (47-58)	1	3	ELISA, Western blot methods, serum immunofixation	**Anti-MAG IgM titers** 1:362'294 (5475-1'300'000)	• Strength in % of normal: 45% (10-85%) • Only baseline nerve conduction studies were performed (ulnar and sural never)l
Rakocevic *et al*. 2018 [14]	Non-responder (n=2)	68 (65-71)	59.5 (52-67)	0	2	Anti-MAG titers by EIA, paraprotein NR	**Anti-MAG IgM titers** >1:102'400 **Paraprotein** 472 mg/dl (420-524 mg/dl)	• Sensory ataxia, muscle weakness • Feet paraesthesia, foot drop
Stino *et al*. 2017 [e4]	Responder (n=1)	76	73	1	0	Anti-MAG titers NR, paraprotein NR	**Anti-MAG IgM titers** 102'400 BTU **Paraprotein** 250 mg/dl	• Distal leg and intrinsic hand weakness MRC grade 4/5. • INCAT: 1 (lower limb • I-RODS: 32 • Assessment of the NCS (median and ulnar DML), MCV
Doneddu *et al*. 2017 [27]	Acute deteriorating (n=2)	74 (72-76)	60.5 (47-74)	0	2	ELISA, paraprotein NR	**Anti-MAG IgM titers** >70'000 BTU **Paraprotein** 4.05 g/L (2-6.1 g/L)	• MRC sum score: 53-61 • RODS: 17 (n=1) • NCS
Gomez *et a*l. 2016 [e5]	Responder (n=1)	74	49	0	1	ELISA	**Anti-MAG IgM titers** 1:51'200	• Progressive paresthesia in the bilateral anterior tibial • Only baseline electro-diagnostic studies were performed.

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Vo *et al*. 2015 [e6]	Acute deteriorating (n=1)	53	52	1	0	Anti-MAG IgM titers NR, total IgM NR	**Anti-MAG IgM titers** >1:102'400 **Total IgM** 443 mg/dl	• INCAT: 0 • MRC sum score: 60 • Grip strength: 76 • Assessment of DML, cMAP, CMV
Talamo *et a*l. 2015 [1]	Responder (n=4)	60.5 (51-73)	52 (29-66)	1	3	Western blot, ELISA, total IgM NR	**Anti-MAG IgM titers** >1:102'400 **Total IgM** 607 mg/dl	• Numbness in extremities, gait imbalance, tingling, weakness, pain • Electrodiagnostic studies were only performed for baseline assessment
	Non-Responder (n=3)	63.7 (62-66)	61.7 (62-66)	0	3		**Anti-MAG IgM titers** >1:72'533 (12'800->102'400) **Total IgM** 647 mg/dl	• Numbness in extremities, gait imbalance, tingling, weakness, pain • Electrodiagnostic studies were only performed for baseline assessment
Zara *et al*. 2011 [28]	Responder (n=3)	59 (43-72)	53.7 (42-60)	1	2	ELISA	**Anti-MAG IgM titers** 29'800 BTU	• INCAT Arm: 3-2 • INCAT Leg: 0-4 • MRC: 50-60 • ISS pinprick: 4 • TLI was assessed (median, ulnar, peroneal nerve)
	Non-Responder (n=2)	55 (48-62)	51 (46-56)	1	1		**Anti-MAG IgM titers** >70'000 BTU	• INCAT Arm: 0-4 • INCAT Leg: 1 • MRC: 48-60 • ISS pinprick: 2-6 • TLI was assessed (median, ulnar, peroneal nerve)

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Delmont *et al*. 2011 [e7]	Responder (n=3)	62.3 (57-62)	57 (54-62)	2	1	ELISA, paraprotein NR	**Anti-MAG IgM titer** Only relative reduction of 44-87% reported **Paraprotein** 9.7 g/L (NR)	• ONLS: 4.7 (3-6) • ISS: 8.3 (2-12) • MRC: 129.3 (123-136) • Assessment of electrophysiological status
Stork *et al*. 2013 [e8]	Acute deteriorating (n=3)	NR	NR	1	2	ELISA, paraprotein NR	**Anti-MAG IgM titers** 1:155'322 (7180-409'600) **Paraprotein** 3.4 g/L (0.3-9)	• MRC grade: 4 • Weakness of hands and foots • Extensive nerve conduction studies were performed including DML, MCV, SNAP, cMAP, TLI of the median, ulnar tibial and peroneal nerve
Broglio *et a*l. 2005 [e9]	Non-Responder (n=1)	75	71	1	0	Western blot, total IgM NR	**Anti-MAG IgM titers** 1:400'000 **Total IgM** 620 mg/dl	• MRC scale 4 • Modified RSS: 3 • Only baseline TLI was reported
Gironi *et al*. 2006 [e10]	Acute deteriorating (n=1)	64	56	1	0	ELISA, nephelometry	**Anti-MAG IgM titers** 144'000 BTU **Paraprotein** 4-5 g/L	• Sever tremor • Unsteadiness of gait
Briani *et al*. 2019 [13]	Responder (n=2)	83 (82-84)	84 (n=1)	1	1	ELISA, paraprotein NR, total IgM NR	**Anti-MAG IgM titers** >70'000 BTU **Paraprotein** 15.8 g/L (n=1) **Total IgM** 14.8 g/L (n=1)	• INCAT leg disability score: 2.5 (1-4) • Extensive nerve conduction studies were performed including DML, MCV, SNAP, cMAP, TLI

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Al-Bustani *et al*. 2015 [e11]	Responder (n=1)	63	60	1	0	ELISA, serum protein electrophoresis	**Anti-MAG IgM titers** 1:25'600 **Paraprotein** 0.2 g/dl **Total IgM** 145 mg/dl	• Distal demyelinating sensory and motor polyneuropathy • No Romberg sign. • Extensive nerve conduction studies were performed including DML, MCV, SNAP, cMAP, TLI
Delarue *et al*. 2004 [e12]	Non-Responder (n=4)	64 (57-87)	60 (NR)	1	3	Anti-MAG IgM titers NR, paraprotein NR	**Anti-MAG IgM titers** No disappearance reported after treatment **Paraprotein** No reduction reported after treatment	• Peripheral sensory-motor polyneuropathy with clinical and electrophysiological symptoms
Benedetti *et al*. 2019 [e13]	Responder (n=16)	65 (48-77)	61 (46-73)	8	8	Western blot	**Anti-MAG IgM titers** 1:40'450 (1600-100'000)	• INCAT 2 (0-5) • MRC score: 57 (40-60) • ISS score: 6 (0-14)
Benedetti *et al*. 2019 [e13]	Non-Responder (n=2)	67.5 (61-74)	57.5 (45-70)	1	1	Western blot	**Anti-MAG IgM titers** 1:425'000 (51'200-800'000)	• INCAT: 2 • MRC score: 58.5 (57-60) • ISS score: 6 • Electrophysiology studies were performed only at the time of diagnosis
Hamidou *et al*. 2005 [e14]	Responder (n=7)	63 (±12 SD)	3.5 (±2.8 SD) mean disease duration	2	7	ELISA, total IgM NR	**Anti-MAG IgM titers** 101'547 BTU (60'220-224'000) **Total IgM** 5.3 g/L (2.8-8)	• Ranking scale: 4 (3-5) • Muscle strength 76 (70-80 • MCV, DML
	Non-Responder (n=2)						**Anti-MAG IgM titers** 27'420 BTU (22'240-23'600) **Total IgM** 5.5. g/L (5-6)	• Ranking scale: 3 • Muscle strength: 81 (78-84) • MCV, DML

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Ghosh *et al*. 2002 [e15]	Responder (n=1)	53	51	0	1	ELISA, protein electrophoresis	**Anti-MAG IgM titers** >70'000 BTU **Total IgM levels** 2.67g/L	• Ascending tingling, numbness • Tremor and neuropathic pain • Unable to use hands
Notermans *et al*. 1996 [e16]	Responder (n=5)	49.2 (46-60)	NR	NR	NR	Western blot, electro- and immune-electrophoresis	**Paraprotein** 9 g/L (n=1) >1 g/L (n=4)	• NR separately for the anti-MAG IgM MGUS cohort • MCV, DML, cMAP, TLI were assessed
Notermans *et al*. 1997 [e17]	Responder (n=5)	60.6 (47-70)	59 (±8 SD)	NR	NR	Paraprotein NR	**Paraprotein** 3.4 g/L (>1-5 g/L)	• Motor sum score: 110.6 (105-116) • Disability scale: 2.6 (2-3)
Niemierko *et al*. 1999 [e18]	Responder (n=1)	53	51	0	1	Anti-MAG IgM titers NR, paraprotein NR	**Anti-MAG IgM titers** 1:52'000 **Paraprotein** 800 mg/dl	• Motor functional score: -3 • Unable to work • Distal weakness, ataxic gait • Baseline EMG values were assessed
Ernerudh *et al*. 1986 [e19]	Responder (n=2)	52 (40-64)	Steady progression for at least 2-3 years	0	2	ELISA, agarose isoelectric focusing, immunofixation, autoradiography	**Anti-MAG IgM titers** Only myelin reactivity was demonstrated **Total IgM** 9.2 g/L (3.7-14.2)	• Painful paraesthesia • Motor velocity condition block in the legs • NCS were assessed in the arms and legs • Predominantly motor and sensory symptoms
	Non-responder (n=1)	59	Steady progression for at least 2-3 years.	1	0	ELISA, agarose isoelectric focusing, immunofixation, autoradiography	**Anti-MAG IgM titers** Only myelin reactivity was demonstrated **Total IgM** 8.0 g/L	• Predominantly sensory symptoms • No velocity condition block • NCS were assessed in the arms and legs

Study type Reference	**Treatment outcome** *(Number of patients)*	**Age** *Mean (Range)*	**Age at disease onset** *Mean (Range)*	**Sex**		**Laboratory testing**	**Pre-treatment anti-MAG IgM level (titers, paraprotein, total IgM** *Mean (Range)*	**Pre-treatment Scale/Score**
				F	**M**			
Ernerudh *et al*. 1992 [e20]	Responder (n=3)	57.7 (44-69)	52.7 (40-69)	1	2	ELISA, western blot, radial immune diffusion technique	**Anti-MAG IgM titers** Only relative change shown **Total IgM** 5.0 g/L (3.0-6.8)	• Disability status: 3.5 (3-4) • Ataxia score 3.7 (3-5) • Nerve conduction velocity: 10-43 m/s (motor), 0-51 m/s (sensory), only baseline reported
	Non-responder (n=2)	70 (65-75)	66.5 (62-71)	1	1		**Anti-MAG IgM titers** Only relative change shown **Total IgM** 9.3 g/L (8.6-10.0)	• Disability status: 3.3 (2.5-4) • Ataxia score: 2.5 (2-3) • Nerve conduction velocity: 30.45 m/s (motor), 0-45 m/s (sensory), only baseline reported
Dalakas *et al*. 1996 [e21]	Responder (n=1)	64	52	1	0	ELISA, thin-layer chromatographic	**Anti-MAG IgM titers** >1:10'000	• MRC: 120 • Neuromuscular symptom scores: 37 • Sensory score: 35
	Non-responder (n=8)	66.3 (56-77)	55.6 (37-70)	2	6	ELISA, thin-layer chromatographic	**Anti-MAG IgM titers** >1:10'000	• MRC: 146 (134-153) • Neuromuscular symptom scores: 50 (43-56) • Sensory score: 32.3 (19-46)
Sherman *et al*. 1984 [29]	Responder (n=2)	51.5 (45-58)	45.5 (35-56)	1	1	Immuno-electrophoresis	**Paraprotein** 470 mg/dL (390-550)	• Unable to walk or sit • Weakness against gravity • MCV in the median, peroneal, sural nerve
	Non-responder (n=4)	60 (53-67)	56.8 (48-66)	2	2		**Paraprotein** 1'025 mg/dL (600-1'200)	• Decreased sensation • Decreased vibration • MCV in the median, peroneal, sural nerve

*Hand selected publications; AAfter initiation of treatment; BTU: Bühlmann Titer Units; cMAP: compound motor action potential amplitude; CNDS: clinical neuropathy disability score; DML distal motor latency; F: Female; FU: Follow-up; INCAT: Inflammatory Neuropathy Cause and Treatment disability score; ISS: INCAT Sensory Score; I-RODS: Inflammatory Rasch-built Overall Disability Scale; M: Male; MCV: motor nerve conduction; MNCV: motor nerve conduction velocity; MRC: Medical Research Council sum score; mRS: modified Rankin Score; NDS: Neuropathy Disability Score; NR: not reported; OLNS: Overall Neuropathy Limitations Scale; SNAP: sensory nerve action potential; SNCV: sensory nerve conduction velocity; TLI: terminal latency index; TNS: Total Neuropathy Score

Supplemental data 2

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