

Clinical considerations on the use of IgG in neuropathies

Prof. Dr. Helmar C. Lehmann
Neurologische Klinik

Klinikum Leverkusen

Disclosures

In the last 3 years honoraria for talks and advisory boards from
Argenx, CSL Behring, Grifols, LFB Pharma, Pfizer, Takeda, UCB

Member of DGN, DGKN, Peripheral Nerve Society, World
Federation of Neurology

Immune-mediated diseases of the PNS

acute

Guillain-Barré syndrome

- demyelinating
- axonal (AMAN, AMSAN)

Miller Fisher syndrome

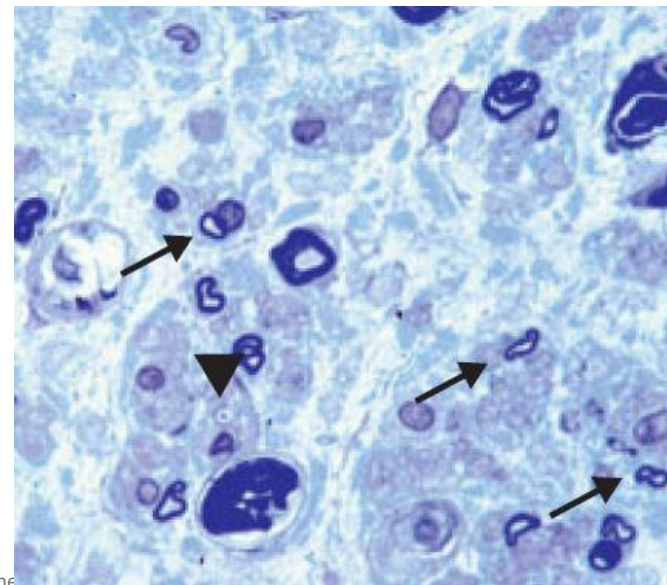
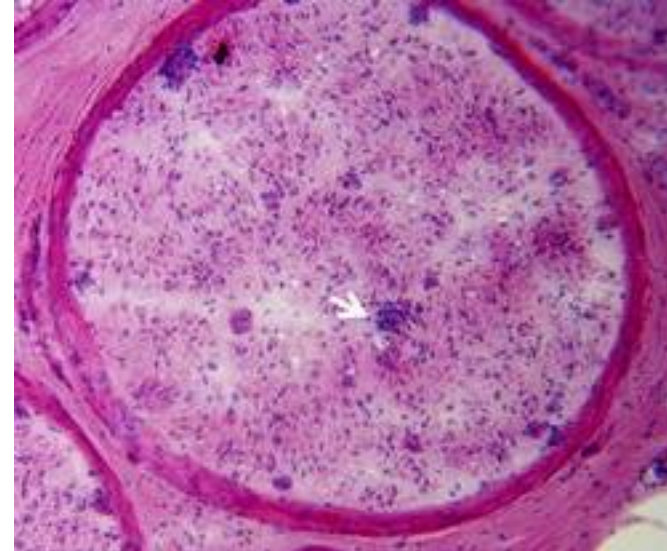
chronic

CIDP

MMN

Paraprot. Neuropathy

(Vaskulitische Neuropathie)



Treatment of immune-mediated neuropathies

	Steroids	PE	IVIg
• Guillain-Barré syndrome	✗	✓	✓
• CIDP	✓	✓	✓
• Multifocal motor neuropathy (MMN)	✗	✗	✓
• Paraproteinaemic polyneuropathy	✓	✓	✓

Guillain-Barré syndrome



Cranial nerve palsy:

36 %

sensory deficits:

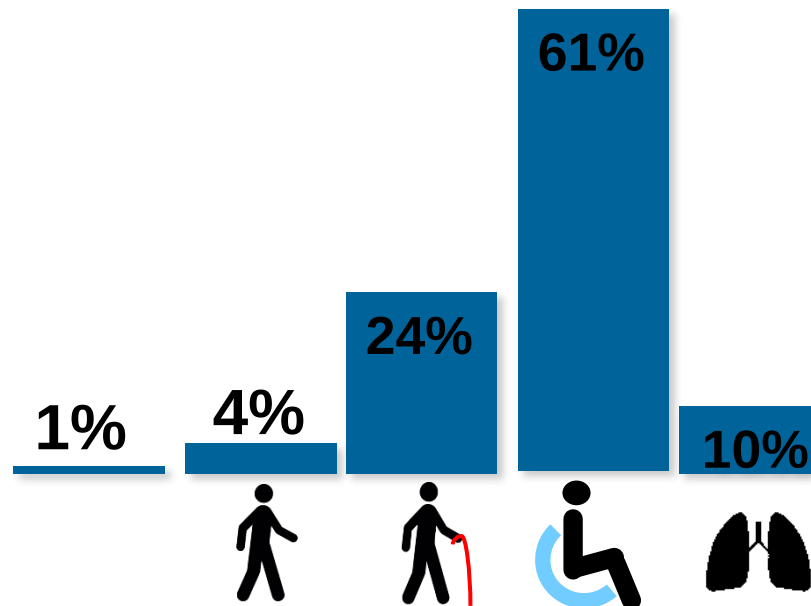
67 %

pain:

54 %

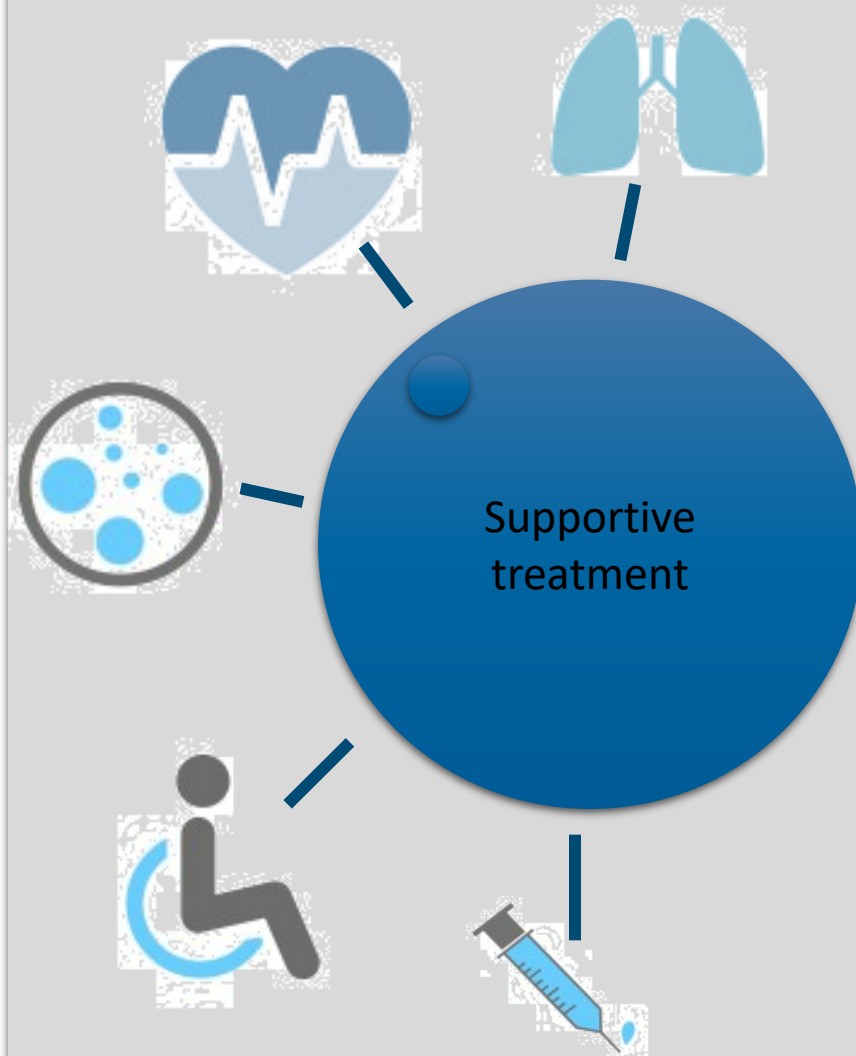
need for respirator:

10%

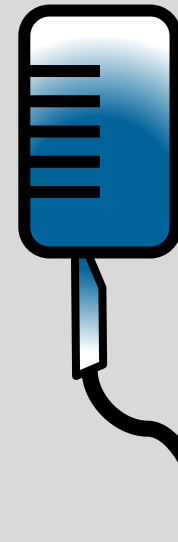


Guillain-Barré syndrome: treatment

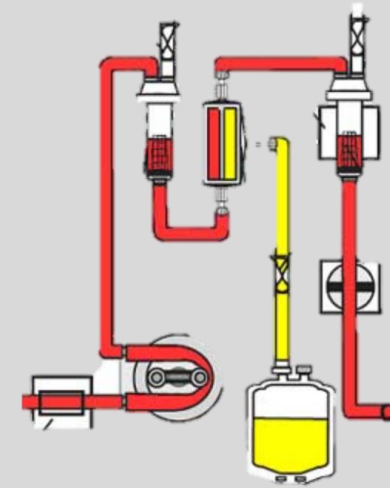
supportive:



causal:



IVIg
5days 0,4g/kg
bodyweigh



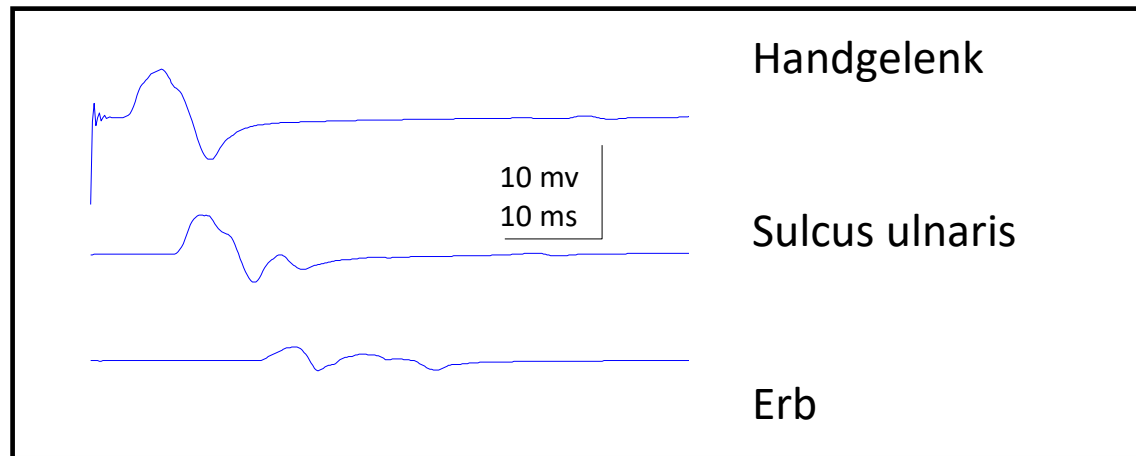
Plasmapheresis
4-6 cycles

Multifocal motor Neuropathy (MMN)

Rare, prevalence approx. 1:1,000,000

Slowly progressive paresis with only slight or no sensory deficits

Electrophysiologically characterized by the presence of persistent conduction blocks in motor nerves



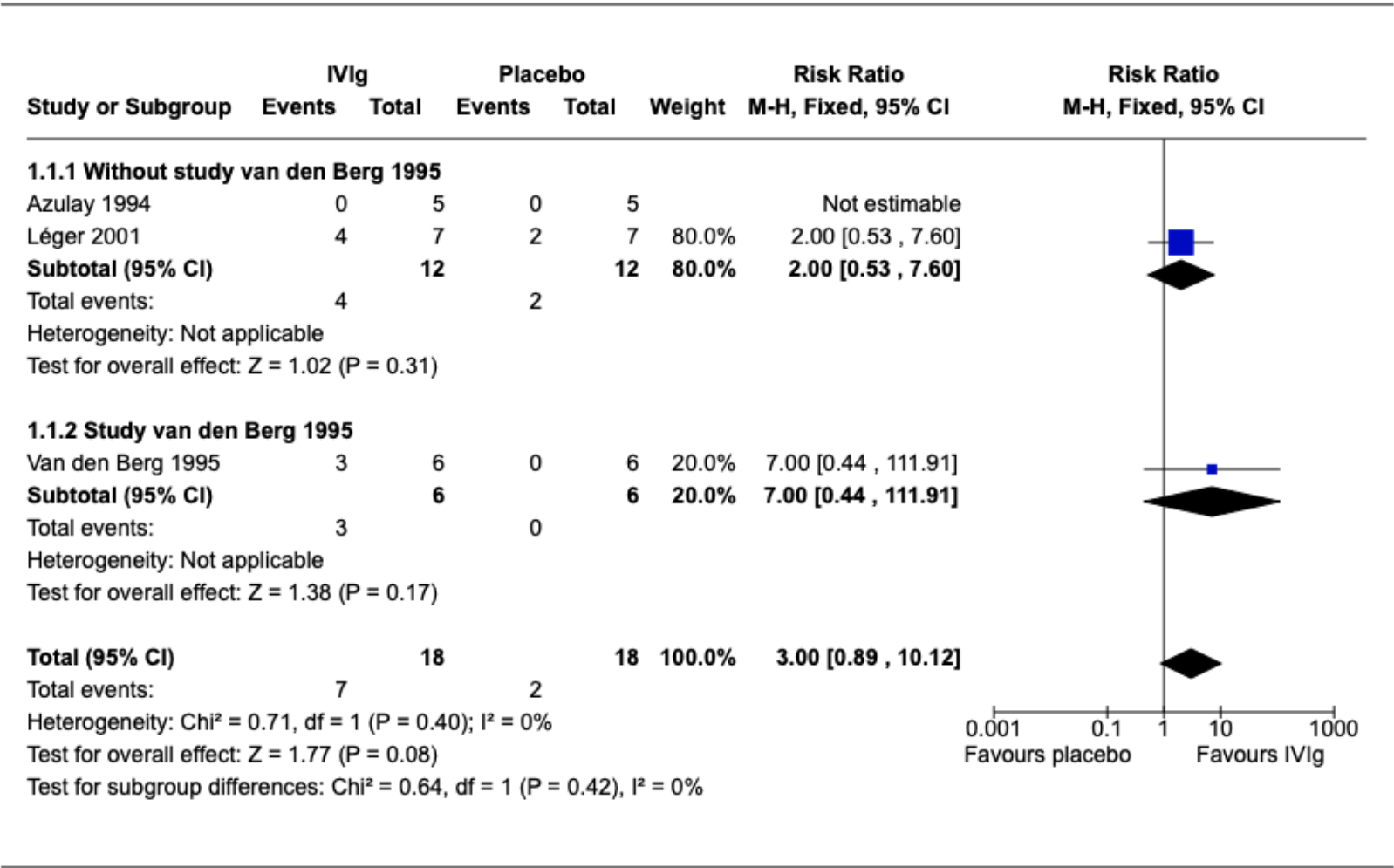
Often associated with IgM antibodies against gangliosides GM1, GD1a, and GD1b (Yuki et al., 2002; Nobile-Orazio, 2003)

IVIg in MMN

- IVIG mainstay of MMN treatment
- Best researched treatment

Figure 3

[Open in figure viewer](#)



IVIg versus placebo. Proportion of participants with an improvement in disability as determined and defined by the study authors. In all forest plots, the 'Total' columns show the number of observations.

SCIG in MMN

Reference	Study Type and Duration	Participants	Outcome of SCIG Therapy
Eftimov et al.	Prospective open-label study, 6 months	10 patients, stable on IVIG	4 patients deteriorated, 4 patients stable
Harbo et al.	Randomized single-blind study, two crossover periods (IVIG and SCIG)	9 patients, stable on IVIG	(1) No difference in muscle strength or SF-36 score (2) 4 patients preferred SCIG, 2 IVIG, 3 had no preference
Misbah et al.	Prospective open-label study, 6 months	8 patients, stable on IVIG	No change in muscle strength or quality of life
Gentile et al.	Retrospective, follow-up of 6.4	8 patients, stable on IVIG	Stable or improved
Al-Zuhairy et al.	Randomized, observer-blinded crossover study of fSCIG and SCIG, 24 weeks for each therapy	20 patients, stable on conventional SCIG	No changes in strength between two SCIG formulations

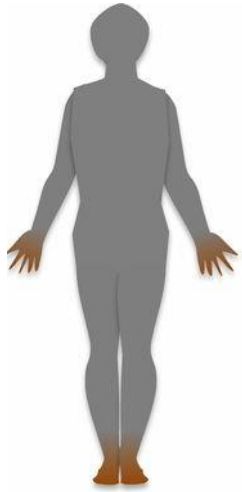
Paraproteinemic Neuropathy

Paraprotein in 3% of all 70-year-olds

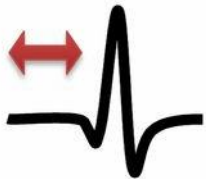
- - Of these, approximately 50% have neuropathy
- - Malignant underlying disease in about 1/3 (Multiple Myeloma, Osteosclerotic Myeloma, Waldenström's Disease, Primary Amyloidosis, Cryoglobulinemia)

Types and Clinical Manifestations:

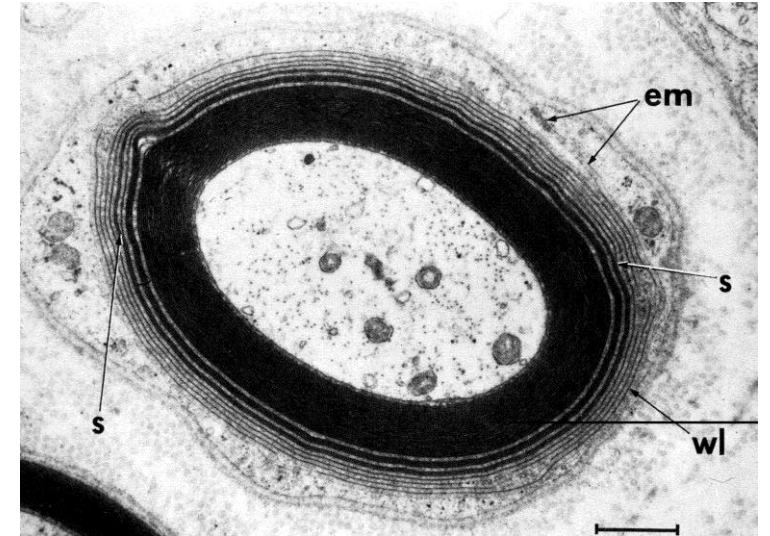
- - **40-50% IgG/IgA:** Clinically similar to CIDP
- - **50% IgM, of which 50% have MAG antibodies:**
 - - Clinically predominantly sensory, chronically progressive, poorly treatable
- Cryoglobulinemia: Axonal, predominantly sensory, symmetrical or multifocal



DADS



increased
dmL



SCIG and IVIG in Paraproteinemic Neuropathy

No RCT for SCIG

IVIG in this condition not well established (low evidence)

Rather monoclonal abs preferred

Thank you for your attention

contact:

- Tel. : +49-221-478-87091
- helmar.lehmann@klinikum-lev.de

