Advances in Clinical Medicine 临床医学进展, 2026, 16(4), 3960-3967
Published Online April 2026 in Hans. https://www.hanspub.org/journal/acm
https://doi.org/10.12677/acm.2026.1641664

慢性免疫介导周围神经病的免疫治疗进展

孙晓瑞[1]，邸　伟[2*]

[1]西安医学院研究生工作部，陕西　西安
[2]陕西省人民医院神经内科，陕西　西安

收稿日期：2026年3月21日；录用日期：2026年4月16日；发布日期：2026年4月21日

摘　要

慢性免疫介导周围神经病是一组由免疫系统调节功能紊乱导致的、以慢性进行性或复发性病程为特征的周围神经疾病，主要包括慢性炎性脱髓鞘性多发性神经根神经病、多灶性运动神经病、抗髓鞘相关糖蛋白抗体相关神经病以及自身免疫性郎飞结病。传统的一线治疗(糖皮质激素、静脉注射免疫球蛋白和血浆置换)虽能使多数患者获益，但仍有约20%~30%的患者疗效不佳、易复发或难以耐受长期治疗。近年来，随着对疾病免疫病理机制认识的深化，靶向B细胞、新生儿Fc受体、补体系统及细胞内信号通路的治疗策略取得了突破性进展，为难治性患者提供了新的选择。本文旨在系统综述慢性免疫介导周围神经病的新型免疫治疗进展，探讨其作用机制、临床证据及未来发展方向，以期为临床实践提供参考。

关键词

慢性炎性脱髓鞘性多发性神经根神经病，多灶性运动神经病，抗髓鞘相关糖蛋白抗体神经病，自身免疫性郎飞结病，免疫治疗，靶向治疗

The Progress of Immunotherapy for Chronic Immune-Mediated Peripheral Neuropathy

Xiaorui Sun[1], Wei Di[2*]

[1]Office of Graduate Student Affairs, Xi'an Medical University, Xi'an Shaanxi
[2]Department of Neurology, Shaanxi Provincial People's Hospital, Xi'an Shaanxi

Received: March 21, 2026; accepted: April 16, 2026; published: April 21, 2026

Abstract

Chronic immune-mediated peripheral neuropathies are a group of peripheral nerve disorders

*通讯作者。

文章引用: 孙晓瑞, 邸伟. 慢性免疫介导周围神经病的免疫治疗进展[J]. 临床医学进展, 2026, 16(4): 3960-3967.
DOI: 10.12677/acm.2026.1641664

characterized by chronic progressive or relapsing courses, resulting from dysregulation of the immune system. They mainly include chronic inflammatory demyelinating polyradiculoneuropathy, multifocal motor neuropathy, anti-myelin-associated glycoprotein antibody-related neuropathy, and autoimmune nodopathies. Although conventional first-line therapies (glucocorticoids, intravenous immunoglobulin, and plasma exchange) benefit most patients, approximately 20%~30% of patients still show poor response, experience relapses, or have difficulty tolerating long-term treatment. In recent years, with a deeper understanding of the immunopathological mechanisms underlying these diseases, breakthrough progress has been made in therapeutic strategies targeting B cells, the neonatal Fc receptor, the complement system, and intracellular signaling pathways, offering new options for refractory patients. This article aims to systematically review the advances in novel immunotherapies for chronic immune-mediated peripheral neuropathies, discuss their mechanisms of action, clinical evidence, and future directions, with the goal of providing a reference for clinical practice.

Keywords

Chronic Inflammatory Demyelinating Polyradiculoneuropathy, Multifocal Motor Neuropathy, Anti-MAG Antibody Neuropathy, Autoimmune Nodopathy, Immunotherapy, Targeted Therapy

1. 引言

免疫介导的周围神经病是一组由于免疫系统调节功能紊乱导致周围神经损伤的异质性疾病，可表现为急性、亚急性或慢性病程[1]。其中，慢性免疫介导周围神经病以病程迁延(超过 8 周)或复发缓解为特征，主要包括慢性炎性脱髓鞘性多发性神经根神经病(chronic inflammatory demyelinating polyradiculoneuropathy, CIDP)、多灶性运动神经病(multifocal motor neuropathy, MMN)、抗髓鞘相关糖蛋白(myelin-associated glycoprotein, MAG)抗体相关神经病，以及近年来从 CIDP 中独立出来的自身免疫性郎飞结病(autoimmune nodopathy, AN) [2] [3]。这类疾病的共同核心机制是免疫系统对周围神经成分的异常攻击，导致髓鞘脱失、轴索损伤或郎飞结/结旁区结构破坏。

尽管糖皮质激素、静脉注射免疫球蛋白(intravenous immunoglobulin, IVIG)和血浆置换(plasma exchange, PE)作为一线治疗，可使约 70%~80%的 CIDP 患者获得良好反应[4]，但仍有相当比例的患者(约 20%~30%)对一线治疗反应不佳，或需要长期、高剂量维持治疗，面临感染、代谢紊乱等副作用风险[5]。而且，MMN 患者依赖反复 IVIG 输注，而抗 MAG 神经病和 AN 患者对 IVIG 的响应率普遍较低[6] [7]。因此，探索机制更精准、疗效更持久、安全性更优的新型免疫治疗策略，成为该领域的研究热点。下面将围绕 CIDP、MMN、抗 MAG 神经病及 AN 的免疫治疗最新进展分别阐述，探讨其作用机制、临床证据及未来发展方向。

2. 慢性炎性脱髓鞘性多发性神经根神经病

CIDP 是最常见的慢性免疫介导周围神经病，其发病机制涉及 T 细胞、B 细胞、巨噬细胞及补体系统的协同作用[8]。临床特征为对称性近端和远端肌无力，伴感觉障碍，病程超过 2 个月[7]。一线治疗包括糖皮质激素、IVIG 和 PE [7] [9]。近年来，针对 CIDP 的靶向治疗取得了重要突破。

2.1. B 细胞靶向治疗

利妥昔单抗(rituximab)是一种靶向 CD20 的嵌合单克隆抗体，通过抗体依赖或补体依赖的细胞毒性作用及诱导细胞凋亡的机制清除 B 细胞，减少自身抗体产生。早期开放标签研究显示其在难治性 CIDP 中具有良好疗效[10] [11]。一项纳入 17 例至少对两种一线治疗无反应 CIDP 患者的前瞻性开放标签研究显示，76.5%的患者在治疗 6 个月后病情改善，88.2%在 12 个月后仍维持改善[12]。中国的一项回顾性研究也证实，与常规治疗队列相比，联合利妥昔单抗治疗的患者病情改善更为显著[13]。然而，随后的一项多中心随机双盲安慰剂对照试验(RIM-CIDP)评估了利妥昔单抗在 CIDP 患者停用 IVIG 后的维持疗效，结果显示其预防临床恶化的效果并不优于安慰剂[14]。这一阴性结果可能与患者异质性、B 细胞清除不彻底或 CIDP 发病机制中存在不依赖 B 细胞的免疫通路有关。以上研究提示利妥昔单抗在 CIDP 中的应用可能需要更精准的患者筛选，例如针对特定亚群。

目前，针对抗 CD20 的其他药物如奥法妥木单抗(ofatumumab)和奥瑞珠单抗(ocrelizumab)仅有少数病例报道，其疗效有待验证[15] [16]。

2.2. 新生儿 Fc 受体抑制剂

新生儿 Fc 受体(neonatal Fc receptor, FcRn)是免疫球蛋白 G (IgG)的主要组织相容性复合物相关 Fc 受体，通过与 IgG 结合，负责保护 IgG 免于溶酶体降解，延长其半衰期[17]。FcRn 抑制剂通过竞争性结合 FcRn，阻断 FcRn 介导的 IgG 回收和再循环，加速包括致病性自身抗体在内的 IgG 清除，成为 CIDP 治疗的新靶点。

艾加莫德(efgartigimod)是一种人源 IgG1 Fc 片段，其皮下制剂在名为 ADHERE 的 II 期随机对照试验中取得了突破性结果：开放标签阶段，66%的患者显示出临床改善；在随机双盲安慰剂对照阶段，艾加莫德组较安慰剂组复发风险降低 61% [18]。基于此，美国食品药品监督管理局于 2024 年批准艾加莫德用于 CIDP 的治疗。然而，真实世界应用中，部分患者从 IVIG 转换至艾加莫德后出现快速且显著的恶化，提示该疗法存在个体差异，临床应用需谨慎评估[19]。其他 FcRn 拮抗剂如罗扎诺利珠单抗(rozanolixizumab)的 IIa 期研究未达到主要终点[20]，而巴托利单抗(batoclimab)和尼卡利单抗(nipocalimab)的临床试验仍在进行中。

2.3. 补体系统抑制剂

补体激活在 CIDP 的髓鞘损伤中起重要作用，补体沉积可见于神经活检标本，血清和脑脊液中补体活化产物增加[21]。补体 C1s 抑制剂 Riliprubart (SAR445088)在 II 期研究中显示出良好的安全性和初步疗效，89%的难治性患者病情稳定或改善，且伴随神经丝轻链水平下降[22]。目前，该药的全球 III 期研究正在进行中。靶向 C5 的依库珠单抗(eculizumab)在少数 CD59 基因缺陷的儿童 CIDP 患者中显示出显著疗效[23]，但在更广泛的 CIDP 人群中的应用仍需探索。

2.4. 其他免疫调节策略

蛋白酶体抑制剂硼替佐米(bortezomib)通过靶向长寿命浆细胞减少自身抗体，在一项包含 10 例难治性 CIDP 的病例系列中，使 6 例患者病情稳定[24]。自体造血干细胞移植(autologous haematopoietic stem cell transplantation, AHSCT)通过免疫重建为难治性 CIDP 提供了潜在治愈可能。一项纳入 66 例患者的前瞻性研究显示，AHSCT 后 5 年，83%的患者达到长期无药缓解，辅助无行走率从术前的 32%提升至术后 1 年的 80%以上[25]。荟萃分析证实其在难治性 CIDP 中的高应答率(约 87%) [26]。嵌合抗原受体 T (chimeric antigen receptor T, CAR-T)细胞疗法在自身免疫病领域方兴未艾，其在 CIDP 中的探索刚刚起步，目前已

有开放标签临床试验正在评估其安全性和有效性[27]。

3. 多灶性运动神经病

MMN 是一种罕见的、以纯运动受累为特征的慢性免疫介导周围神经病，临床表现为不对称性、缓慢进展性肢体无力，以上肢远端为主，电生理检查可见运动神经传导阻滞[28]。其发病机制的核心在于抗 GM1 IgM 抗体与郎飞结上的 GM1 结合，并通过经典途径激活补体，导致膜攻击复合物形成，进而引起钠通道功能紊乱和传导阻滞，这一病理过程高度依赖补体系统的活化[28]。

IVIG 是目前唯一被 FDA 批准的一线维持治疗，可有效改善肌力并延缓残疾进展[29]。初始剂量通常为 2 g/kg，分 2~5 天输注，维持治疗需根据个体反应调整。皮下注射免疫球蛋白(subcutaneous immunoglobulin, SCIG)作为 IVIG 的替代，因其给药方便、全身副作用少，已被证实疗效不劣于 IVIG，且患者满意度更高[30] [31]。

针对难治性 MMN，探索性治疗包括利妥昔单抗和环磷酰胺，但缺乏随机对照试验证据支持[32]。利妥昔单抗在 MMN 中的研究结果不一，部分研究显示其可改善肌力并减少 IVIG 用量，而另一些研究则未显示明显获益[33]。鉴于补体激活在 MMN 发病机制中的核心地位，补体抑制剂成为极具前景的靶向治疗方向。一项在诱导多能干细胞来源的运动神经元模型中开展的研究显示，抗 C2 单抗(ARGX-117)能有效抑制 MMN 患者血清 IgM 诱导的补体激活[34]。基于此，ARGX-117 已进入临床试验阶段。依库珠单抗在 MMN 中的开放标签研究仅显示边缘性获益，未能改变对 IVIG 的依赖[35]。

4. 抗髓鞘相关糖蛋白抗体相关神经病

抗 MAG 神经病是一种与 IgM 单克隆丙种球蛋白病相关的慢性、远端、感觉性脱髓鞘神经病，典型特征为远端潜伏期显著延长、感觉性共济失调和震颤。其致病抗体为 IgM 型抗 MAG 抗体，通过激活补体导致髓鞘板层增宽[6]。

IVIG 对该病仅有短暂且不确定的疗效，临床意义有限[36]。利妥昔单抗是目前应用最广的靶向治疗。尽管随机对照试验均未达到其预设的主要终点，但事后分析和荟萃分析显示，利妥昔单抗能显著改善患者的 INCAT 残疾评分[37]。早期治疗、保留轴索完整性的患者预后更佳[38]。有研究发现在两年多的随访期内，RTX 对临床活动性脱髓鞘抗 MAG 神经病变患者具有疗效[39]。目前，针对病程短、抗体滴度高的患者 III 期研究正在进行中。

布鲁顿酪氨酸激酶(Bruton tyrosine kinase, BTK)是 B 细胞受体信号通路的关键分子，在表达 MYD88 L265P 突变的淋巴浆细胞中异常活跃。鉴于超过 60%的抗 MAG 神经病患者携带此突变[40]，BTK 抑制剂为该病提供了新思路。病例系列报道显示，第一代 BTK 抑制剂伊布替尼(ibrutinib)及第二代药物泽布替尼(zanubrutinib)、阿卡替尼(acalabrutinib)可改善患者临床症状[41]。目前，多项评估 BTK 抑制剂单药或联合利妥昔单抗的 II 期研究正在进行中。

5. 自身免疫性郎飞结病

AN 是近年来被确立的独立疾病实体，由靶向郎飞结/结旁区蛋白(如神经束蛋白 155、接触蛋白 1、接触蛋白相关蛋白 1)的自身抗体(多为 IgG4 亚型)驱动。与 CIDP 不同，IgG4 亚型抗体不具备激活补体或介导炎症细胞浸润的能力，而是通过直接阻断蛋白–蛋白相互作用，破坏郎飞结/结旁区的正常结构，从而致病。这一独特的病理机制解释了 AN 对 IVIG 普遍不敏感的原因，也提示 B 细胞清除疗法在理论上具有优势[42]。

B 细胞清除治疗，特别是利妥昔单抗，已成为 AN 的一线选择。大量回顾性研究和病例系列显示，利妥昔单抗在 AN 患者中的有效率超过 80% [43]。一项最新的系统评价和荟萃分析纳入 29 项研究共 118

例患者，结果显示抗 CD20 单抗治疗的总体临床应答率高达 92.0%，且在抗 NF155 和抗 CNTN1 亚组中均保持较高水平[44]。对于对利妥昔单抗反应不佳的患者，可考虑使用第二代抗 CD20 单抗(如奥法妥木单抗)或靶向浆细胞的药物(如硼替佐米、达雷妥尤单抗) [44] [45]。糖皮质激素对部分患者也有效，但其地位不如 B 细胞清除疗法。

6. 总结

慢性免疫介导周围神经病的免疫治疗正经历着从广谱免疫抑制向精准靶向治疗的深刻变革。FcRn 抑制剂艾加莫德的获批，为 CIDP 患者提供了首个非激素、非免疫球蛋白的机制性治疗新选择，其快速降低 IgG 的特点有望改变部分患者的治疗模式。补体抑制剂、BTK 抑制剂以及 CAR-T 细胞疗法在特定亚型中的探索，代表了未来个体化治疗的希望。

然而，该领域仍面临诸多挑战。首先，疾病的异质性要求更精准的生物标志物来指导治疗选择。识别哪些患者最适合 FcRn 抑制剂，哪些患者能从利妥昔单抗中获益，是优化治疗的关键。其次，新型药物的长期安全性，尤其是感染风险，需要大样本、长期随访研究来确认。最后，高昂的治疗费用限制了其在临床的广泛应用。

展望未来，为推动该领域向更精准、更安全的方向发展，仍需在以下几个关键方向上开展深入研究：① 临床转化策略：针对 CIDP 患者从 IVIG 转换至 FcRn 抑制剂过程中出现的快速恶化现象，亟需建立基于临床和药效学指标的过渡方案，例如明确转换时机、联合用药策略及风险分层模型，以最大程度降低短期风险[19]。② 联合/序贯治疗探索：在抗 MAG 神经病中，BTK 抑制剂与利妥昔单抗可能通过抑制 B 细胞受体信号和清除 B 细胞产生协同效应，设计头对头比较研究(如 BTK 抑制剂单药、利妥昔单抗单药及两者联合)，将有助于明确最优治疗组合。③ 新型生物标志物开发：除神经丝轻链外，探索更多可客观反映疾病活动性和治疗反应的标志物，如神经影像学(如 MR 神经成像的神经增粗程度、脂肪抑制序列信号强度)、特定抗体亚型及其滴度动态变化、以及循环中浆细胞或 B 细胞亚群比例等，将有助于实现治疗效果的动态监测和个体化方案的动态调整。

随着对疾病免疫病理学理解的不断深入和更多靶向药物的开发，慢性免疫介导周围神经病患者有望迎来疗效更好、安全性更高的个体化治疗时代。

基金项目

陕西省自然科学基金面上项目(2022JM-533)，陕西省人民医院科技人才资助项目(2021BJ-25, 2021YJY-26)。

参考文献

[1] Shastri, A., Al Aiyan, A., Kishore, U. and Farrugia, M.E. (2023) Immune-Mediated Neuropathies: Pathophysiology and Management. *International Journal of Molecular Sciences*, **24**, Article No. 7288. https://doi.org/10.3390/ijms24087288

[2] Rajabally, Y. (2024) Chronic Inflammatory Demyelinating Polyradiculoneuropathy: Current Therapeutic Approaches and Future Outlooks. *ImmunoTargets and Therapy*, **13**, 99-110. https://doi.org/10.2147/itt.s388151

[3] Briani, C. and Visentin, A. (2026) Immunotherapies in Chronic Immune-Mediated Neuropathies. In: *Handbook of Clinical Neurology*, Elsevier, 345-356. https://doi.org/10.1016/b978-0-323-90887-0.00019-5

[4] Oaklander, A.L., Lunn, M.P., Hughes, R.A., van Schaik, I.N., Frost, C. and Chalk, C.H. (2017) Treatments for Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP): An Overview of Systematic Reviews. *Cochrane Database of Systematic Reviews*, **2017**, CD010369. https://doi.org/10.1002/14651858.cd010369.pub2

[5] Kiers, L. and Cruse, B. (2025) Evaluation and Treatment of Refractory Chronic Inflammatory Demyelinating Polyradiculoneuropathy. *BMJ Neurology Open*, **7**, e001318. https://doi.org/10.1136/bmjno-2025-001318

[6] Min, Y.G., Visentin, A., Briani, C. and Rajabally, Y.A. (2025) Neuropathy with Anti-Myelin-Associated Glycoprotein

Antibodies: Update on Diagnosis, Pathophysiology and Management. *Journal of Neurology, Neurosurgery & Psychiatry*, **96**, 340-349. https://doi.org/10.1136/jnnp-2024-334678

[7] Van den Bergh, P.Y.K., van Doorn, P.A., Hadden, R.D.M., Avau, B., Vankrunkelsven, P., Allen, J.A., *et al*. (2021) European Academy of Neurology/Peripheral Nerve Society Guideline on Diagnosis and Treatment of Chronic Inflammatory Demyelinating Polyradiculoneuropathy: Report of a Joint Task Force—Second Revision. *European Journal of Neurology*, **28**, 3556-3583. https://doi.org/10.1111/ene.14959

[8] Kuwabara, S. (2024) Typical CIDP: Update of the Pathogenesis, Diagnosis, and Treatment. *Brain and Nerve*, **76**, 515-519. https://doi.org/10.11477/mf.1416202637

[9] 中华医学会神经病学分会, 中华医学会神经病学分会周围神经病协作组, 中华医学会神经病学分会肌电图与临床神经电生理学组, 中华医学会神经病学分会神经肌肉病学组. 慢性炎性脱髓鞘性多发性神经根神经病诊治中国专家共识 2022 [J]. 中华神经科杂志, 2023, 56(2): 125-132.

[10] Muley, S.A., Jacobsen, B., Parry, G., Usman, U., Ortega, E., Walk, D., *et al*. (2020) Rituximab in Refractory Chronic Inflammatory Demyelinating Polyneuropathy. *Muscle & Nerve*, **61**, 575-579. https://doi.org/10.1002/mus.26804

[11] Benedetti, L., Briani, C., Franciotta, D., Fazio, R., Paolasso, I., Comi, C., *et al*. (2011) Rituximab in Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy: A Report of 13 Cases and Review of the Literature. *Journal of Neurology, Neurosurgery & Psychiatry*, **82**, 306-308. https://doi.org/10.1136/jnnp.2009.188912

[12] Doneddu, P.E., Cocito, D., Fazio, R., Benedetti, L., Peci, E., Liberatore, G., *et al*. (2024) Prospective Open-Label Trial with Rituximab in Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy Not Responding to Conventional Immune Therapies. *Journal of Neurology, Neurosurgery & Psychiatry*, **95**, 838-844. https://doi.org/10.1136/jnnp-2023-332844

[13] Du, Y., Yan, Q., Li, C., Zhu, W., Zhao, C., Hao, Y., *et al*. (2024) Efficacy and Safety of Combined Low-Dose Rituximab Regimen for Chronic Inflammatory Demyelinating Polyradiculoneuropathy. *Annals of Clinical and Translational Neurology*, **12**, 180-191. https://doi.org/10.1002/acn3.52270

[14] Nobile-Orazio, E., Cocito, D., Manganelli, F., Fazio, R., Lauria Pinter, G., Benedetti, L., *et al*. (2025) Rituximab versus Placebo for Chronic Inflammatory Demyelinating Polyradiculoneuropathy: A Randomized Trial. *Brain*, **148**, 1112-1121. https://doi.org/10.1093/brain/awae400

[15] Wang, J. and Xiang, Q. (2024) Successful Treatment of Relapsed and Refractory CIDP with Ofatumumab: A First Case Report. *Frontiers in Immunology*, **15**, Article ID: 1437848. https://doi.org/10.3389/fimmu.2024.1437848

[16] Casertano, S., Signoriello, E., Rossi, F., Di Pietro, A., Tuccillo, F., Bonavita, S., *et al*. (2020) Ocrelizumab in a Case of Refractory Chronic Inflammatory Demyelinating Polyneuropathy with Anti-Rituximab Antibodies. *European Journal of Neurology*, **27**, 2673-2675. https://doi.org/10.1111/ene.14498

[17] Dalakas, M.C. and Spaeth, P.J. (2021) The Importance of FcRn in Neuro-Immunotherapies: From IgG Catabolism, *fcgrt* Gene Polymorphisms, IVIg Dosing and Efficiency to Specific FcRn Inhibitors. *Therapeutic Advances in Neurological Disorders*, **14**. https://doi.org/10.1177/17562864211997381

[18] Allen, J.A., Lin, J., Basta, I., Dysgaard, T., Eggers, C., Guptill, J.T., *et al*. (2024) Safety, Tolerability, and Efficacy of Subcutaneous Efgartigimod in Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy (ADHERE): A Multicentre, Randomised-Withdrawal, Double-Blind, Placebo-Controlled, Phase 2 Trial. *The Lancet Neurology*, **23**, 1013-1024. https://doi.org/10.1016/s1474-4422(24)00309-0

[19] Levine, T. and Muley, S. (2025) Early Deterioration of CIDP Following Transition from IVIG to FcRn Inhibitor Treatment. *Journal of the Neurological Sciences*, **468**, Article ID: 123313. https://doi.org/10.1016/j.jns.2024.123313

[20] Querol, L., De Sèze, J., Dysgaard, T., Levine, T., Rao, T.H., Rivner, M., *et al*. (2024) Efficacy, Safety and Tolerability of Rozanolixizumab in Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy: A Randomised, Subject-Blind, Investigator-Blind, Placebo-Controlled, Phase 2a Trial and Open-Label Extension Study. *Journal of Neurology, Neurosurgery & Psychiatry*, **95**, 845-854. https://doi.org/10.1136/jnnp-2023-333112

[21] Querol, L.A., Hartung, H., Lewis, R.A., van Doorn, P.A., Hammond, T.R., Atassi, N., *et al*. (2022) The Role of the Complement System in Chronic Inflammatory Demyelinating Polyneuropathy: Implications for Complement-Targeted Therapies. *Neurotherapeutics*, **19**, 864-873. https://doi.org/10.1007/s13311-022-01221-y

[22] Querol, L., Lewis, R.A., Hartung, H., Van Doorn, P.A., Wallstroem, E., Luo, X., *et al*. (2023) An Innovative Phase 2 Proof-of-Concept Trial Design to Evaluate SAR445088, a Monoclonal Antibody Targeting Complement C1s in Chronic Inflammatory Demyelinating Polyneuropathy. *Journal of the Peripheral Nervous System*, **28**, 276-285. https://doi.org/10.1111/jns.12551

[23] Mevorach, D., Reiner, I., Grau, A., Ilan, U., Berkun, Y., Ta-Shma, A., *et al*. (2016) Therapy with Eculizumab for Patients with CD59 P.cys89tyr Mutation. *Annals of Neurology*, **80**, 708-717. https://doi.org/10.1002/ana.24770

[24] Pitarokoili, K., Yoon, M., Kröger, I., Reinacher-Schick, A., Gold, R. and Schneider-Gold, C. (2017) Severe Refractory CIDP: A Case Series of 10 Patients Treated with Bortezomib. *Journal of Neurology*, **264**, 2010-2020.

https://doi.org/10.1007/s00415-017-8599-4

[25] Burt, R.K., Balabanov, R., Tavee, J., Han, X., Sufit, R., Ajroud-Driss, S., *et al*. (2020) Hematopoietic Stem Cell Transplantation for Chronic Inflammatory Demyelinating Polyradiculoneuropathy. *Journal of Neurology*, **267**, 3378-3391. https://doi.org/10.1007/s00415-020-10010-6

[26] Zheng, Y., Hu, J., Sun, C., Zhao, C. and Lin, J. (2023) Efficacy of Hematopoietic Stem Cell Transplantation Treatment in Refractory Chronic Inflammatory Demyelinating Polyradiculoneuropathy: A Systematic Review and Meta-Analysis. *European Journal of Neurology*, **30**, 2570-2582. https://doi.org/10.1111/ene.15857

[27] Dong, M., Mei, Z., Zhou, L., Heming, M., Xu, L., Liu, Y., *et al*. (2025) Anti-bcma CAR-T Cell Therapy in Relapsed/Refractory Chronic Inflammatory Demyelinating Polyneuropathy. *Med*, **6**, Article ID: 100704. https://doi.org/10.1016/j.medj.2025.100704

[28] Claytor, B., Polston, D. and Li, Y. (2025) Multifocal Motor Neuropathy: A Narrative Review. *Muscle & Nerve*, **71**, 512-534. https://doi.org/10.1002/mus.28349

[29] Keddie, S., Eftimov, F., van den Berg, L.H., Brassington, R., de Haan, R.J. and van Schaik, I.N. (2022) Immunoglobulin for Multifocal Motor Neuropathy. *Cochrane Database of Systematic Reviews*, **2022**, CD004429. https://doi.org/10.1002/14651858.cd004429.pub3

[30] Sharawat, I.K., Panda, P., Dawman, L., Kasinathan, A. and Panda, P.K. (2025) Efficacy of Subcutaneous and Intravenous Immunoglobulin in Patients with Multifocal Motor Neuropathy: A Systematic Review and Meta-Analysis. *Annals of Indian Academy of Neurology*, **28**, 485-494. https://doi.org/10.4103/aian.aian_815_24

[31] Al-Zuhairy, A., Sindrup, S.H. and Jakobsen, J. (2021) Long-Term Follow-Up of Facilitated Subcutaneous Immunoglobulin Therapy in Multifocal Motor Neuropathy. *Journal of the Neurological Sciences*, **427**, Article ID: 117495. https://doi.org/10.1016/j.jns.2021.117495

[32] Pestronk, A. (2003) Treatment of IgM Antibody Associated Polyneuropathies Using Rituximab. *Journal of Neurology, Neurosurgery & Psychiatry*, **74**, 485-489. https://doi.org/10.1136/jnnp.74.4.485

[33] Chaganti, S., Hannaford, A. and Vucic, S. (2022) Rituximab in Chronic Immune Mediated Neuropathies: A Systematic Review. *Neuromuscular Disorders*, **32**, 621-627. https://doi.org/10.1016/j.nmd.2022.05.013

[34] Budding, K., Johansen, L.E., Van de Walle, I., Dijkxhoorn, K., de Zeeuw, E., Bloemenkamp, L.M., *et al*. (2022) Anti-c2 Antibody ARGX-117 Inhibits Complement in a Disease Model for Multifocal Motor Neuropathy. *Neurology Neuroimmunology & Neuroinflammation*, **9**, e1107. https://doi.org/10.1212/nxi.0000000000001107

[35] Fitzpatrick, A.M., Mann, C.A., Barry, S., Brennan, K., Overell, J.R. and Willison, H.J. (2011) An Open Label Clinical Trial of Complement Inhibition in Multifocal Motor Neuropathy. *Journal of the Peripheral Nervous System*, **16**, 84-91. https://doi.org/10.1111/j.1529-8027.2011.00328.x

[36] Lunn, M.P. and Nobile-Orazio, E. (2016) Immunotherapy for IgM Anti-Myelin-Associated Glycoprotein Paraprotein-Associated Peripheral Neuropathies. *Cochrane Database of Systematic Reviews*, **2016**, CD002827. https://doi.org/10.1002/14651858.cd002827.pub4

[37] Léger, J., Viala, K., Nicolas, G., Créange, A., Vallat, J., Pouget, J., *et al*. (2013) Placebo-Controlled Trial of Rituximab in IgM Anti-Myelin-Associated Glycoprotein Neuropathy. *Neurology*, **80**, 2217-2225. https://doi.org/10.1212/wnl.0b013e318296e92b

[38] Tang, M.H., Mathis, S., Duffau, P., Cazenave, P., Solé, G., Duval, F., *et al*. (2020) Prognostic Factor of Poor Outcome in Anti-MAG Neuropathy: Clinical and Electrophysiological Analysis of a French Cohort. *Journal of Neurology*, **267**, 561-571. https://doi.org/10.1007/s00415-019-09618-0

[39] Parisi, M., Dogliotti, I., Clerico, M., Bertuzzo, D., Benevolo, G., Orsucci, L., *et al*. (2022) Efficacy of Rituximab in Anti-Myelin-Associated Glycoprotein Demyelinating Polyneuropathy: Clinical, Hematological and Neurophysiological Correlations during 2 Years of Follow-Up. *European Journal of Neurology*, **29**, 3611-3622. https://doi.org/10.1111/ene.15553

[40] Castellani, F., Visentin, A., Schirinzi, E., Salvalaggio, A., Cacciavillani, M., Candiotto, C., *et al*. (2023) Mutational Profile in 75 Patients with Anti-Myelin-Associated Glycoprotein Neuropathy. *Neurology Neuroimmunology & Neuroinflammation*, **10**, e200122. https://doi.org/10.1212/nxi.0000000000200122

[41] Castellani, F., Visentin, A., Campagnolo, M., Salvalaggio, A., Cacciavillani, M., Candiotto, C., *et al*. (2020) The Bruton Tyrosine Kinase Inhibitor Ibrutinib Improves Anti-Mag Antibody Polyneuropathy. *Neurology Neuroimmunology & Neuroinflammation*, **7**, e720. https://doi.org/10.1212/nxi.0000000000000720

[42] Querol, L. and Dalakas, M.C. (2025) The Discovery of Autoimmune Nodopathies and the Impact of IgG4 Antibodies in Autoimmune Neurology. *Neurology Neuroimmunology & Neuroinflammation*, **12**, e200365. https://doi.org/10.1212/nxi.0000000000200365

[43] Liu, B., Hu, J., Sun, C., Qiao, K., Xi, J., Zheng, Y., *et al*. (2023) Effectiveness and Safety of Rituximab in Autoimmune Nodopathy: A Single-Center Cohort Study. *Journal of Neurology*, **270**, 4288-4295.

https://doi.org/10.1007/s00415-023-11759-2

[44] Tao, Z., Jiang, Y., Gui, Q. and Ma, J. (2026) Efficacy and Safety of Anti-CD20 Monoclonal Antibody Therapy for Autoimmune Nodopathies: A Systematic Review and Meta-Analysis. *Frontiers in Neurology*, **17**, Article ID: 1759210. https://doi.org/10.3389/fneur.2026.1759210

[45] Scheibe, F., Ostendorf, L., Prüss, H., Radbruch, H., Aschman, T., Hoffmann, S., *et al*. (2022) Daratumumab for Treatment-Refractory Antibody-Mediated Diseases in Neurology. *European Journal of Neurology*, **29**, 1847-1854. https://doi.org/10.1111/ene.15266