Wichtige Paper der letzten 24 Monate in der Neuroimmunology

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Diagnosis of myelin oligodendrocyte glycoprotein antibody-associated disease: International MOGAD Panel proposed criteria

Brenda Banwell, Jeffrey L Bennett*, Romain Marignier*, Ho Jin Kim*, Fabienne Brilot, Eoin P Flanagan, Sudarshini Ramanathan, Patrick Waters, Silvia Tenembaum, Jennifer S Graves, Tanuja Chitnis, Alexander U Brandt, Cheryl Hemingway, Rinze Neuteboom, Lekha Pandit, Markus Reindl, Albert Saiz, Douglas Kazutoshi Sato, Kevin Rostasy*, Friedemann Paul*, Sean J Pittock*, Kazuo Fujihara*, Jacqueline Palace**

Serum antibodies directed against myelin oligodendrocyte glycoprotein (MOG) are found in patients with acquired CNS demyelinating syndromes that are distinct from multiple sclerosis and aquaporin-4-seropositive neuromyelitis optica spectrum disorder. Based on an extensive literature review and a structured consensus process, we propose diagnostic criteria for MOG antibody-associated disease (MOGAD) in which the presence of MOG-IgG is a core criterion. According to our proposed criteria, MOGAD is typically associated with acute disseminated encephalomyelitis, optic neuritis, or transverse myelitis, and is less commonly associated with cerebral cortical encephalitis, brainstem presentations, or cerebellar presentations. MOGAD can present as either a monophasic or relapsing disease course, and MOG-IgG cell-based assays are important for diagnostic accuracy. Diagnoses such as multiple sclerosis need to be excluded, but not all patients with multiple sclerosis should undergo screening for MOG-IgG. These proposed diagnostic criteria require validation but have the potential to improve identification of individuals with MOGAD, which is essential to define long-term clinical outcomes, refine inclusion criteria for clinical trials, and identify predictors of a relapsing versus a monophasic disease course.

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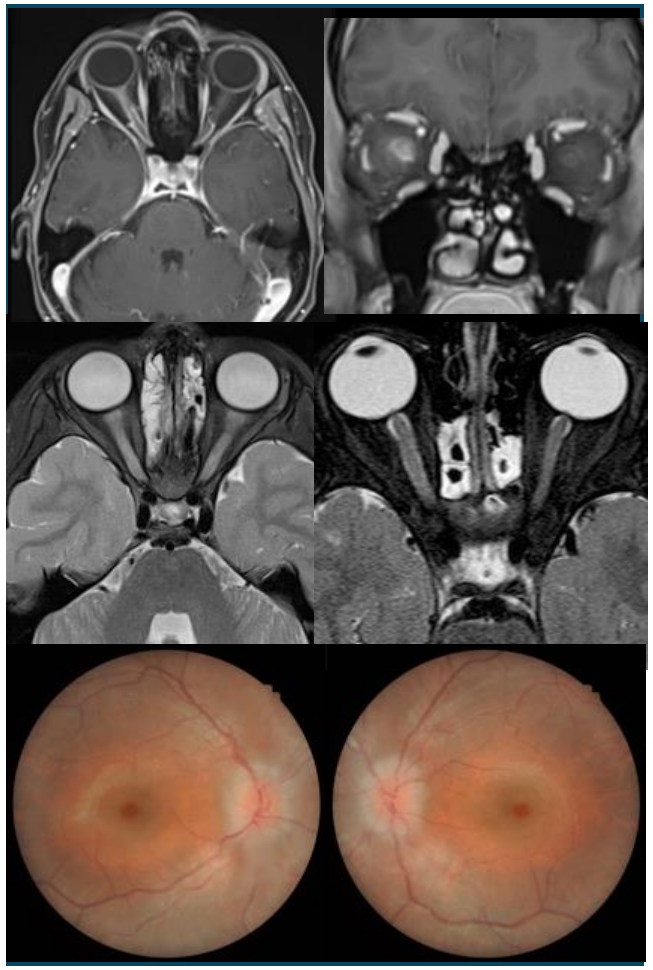
DIAGNOSIS OF MOGAD: ALL THREE CRITERIA MUST BE FULFILLED

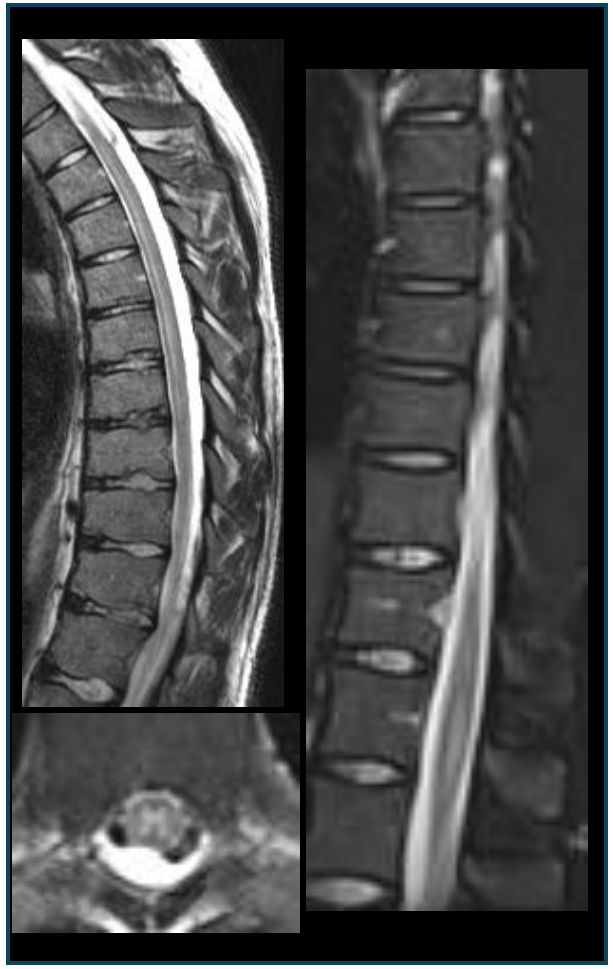
1- **First acute/subacute clinical event**	• Optic neuritis • Myelitis • ADEM • Cerebral syndrome with monofocal or polyfocal deficits • Brainstem or Cerebellar syndrome • Cerebral cortical encephalitis	
2- **Seropositive MOG-IgG test result**	**ADEM phenotype** seropositive by fixed- or live-CBA	No additional requirement
	Non-ADEM phenotype: high seropositive by live-CBA	Not additional requirement
	Non-ADEM phenotype: low seropositive by live-CBA or seropositive by fixed-CBA	At least one of the supporting clinical/MRI requirements needed [A]
3- **Exclusion of MS, AQP4-IgG associated disease, or any better explanation**		

A- **Supporting clinical / MRI requirements**	Optic Neuritis	Bilateral simultaneous clinical involvement; longitudinal optic nerve involvement (>50%); perineural optic sheath enhancement; optic disc edema
	Myelitis	Longitudinally extensive mielitis; central cord lesion; H sign; conus lesion
	Brain/ brainstem syndromes	Large ill-defined T2-hyperintense lesion/s in supratentorial or infratentorial (brainstem, middle cerebellar peduncle) WM; deep GM involvement; cortical lesion with/without lesional and overlying meningeal enhancement

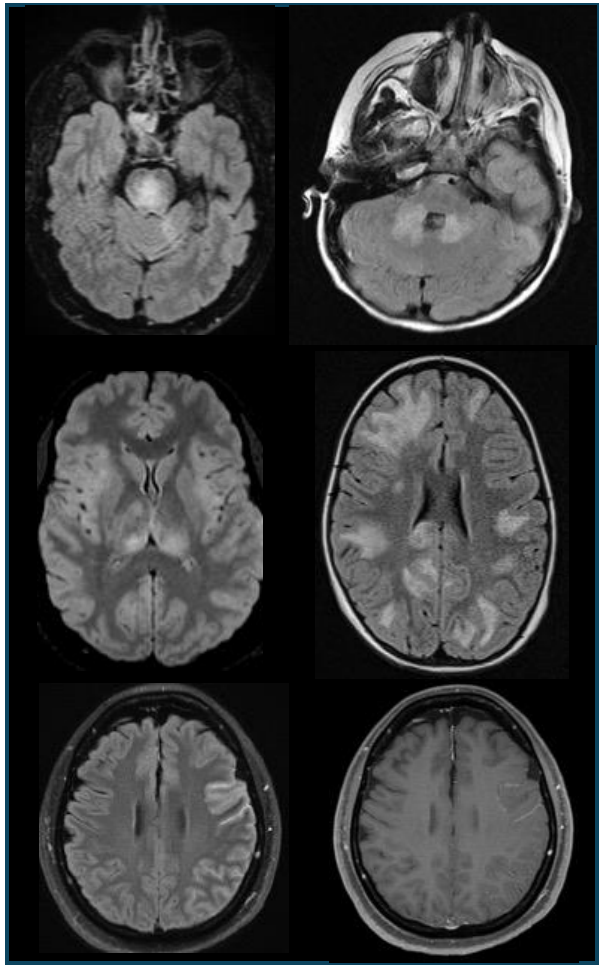
Optic nerve



Spinal cord



Brain



Original research

Oral corticosteroid dosage and tapeduration at onset in myelin oligodendrocyte glycoprotein antibody-associated disease influences time to first relapse

Benjamin P Trewin ,[1] Russell C Dale ,[2,3] Jessica Qiu,[1] Melissa Chu,[4,5] Niroshan Jeyakumar,[1] Fionna Dela Cruz,[4,5] Jane Andersen,[1,6] Pakeeran Siriratnam,[7] Kit Kwan M Ma,[4,5] Todd A Hardy ,[3,8] Anneke van der Walt ,[7,9] Jeanette Lechner-Scott,[10] Helmut Butzkueven,[7,9] Simon A Broadley ,[11,12] Michael H Barnett ,[3,13] Stephen W Reddel ,[3,8] Fabienne Brilot ,[6,14] Tomas Kalincik ,[4,5] Sudarshini Ramanathan ,[1,8] On behalf of the Australasian MOGAD Study Group

Neuro-inflammation

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AIM: To identify an optimal oral corticosteroid regimen at onset of MOGAD, which would delay time to first relapse while minimising cumulative corticosteroid exposure.

RESULTS: In this study of 109 children/adults with MOGAD, there was evidence that patients treated with at least 12.5 mg/day (0.16 mg/kg in children) of oral prednisone for at least 3 months had an 88% reduction in the risk of relapse compared with those who did not receive this regimen.

CONCLUSION: The optimal dose of 12.5 mg of prednisone daily for a minimum of 3 months at the onset of MOGAD delays time to first relapse!!!!

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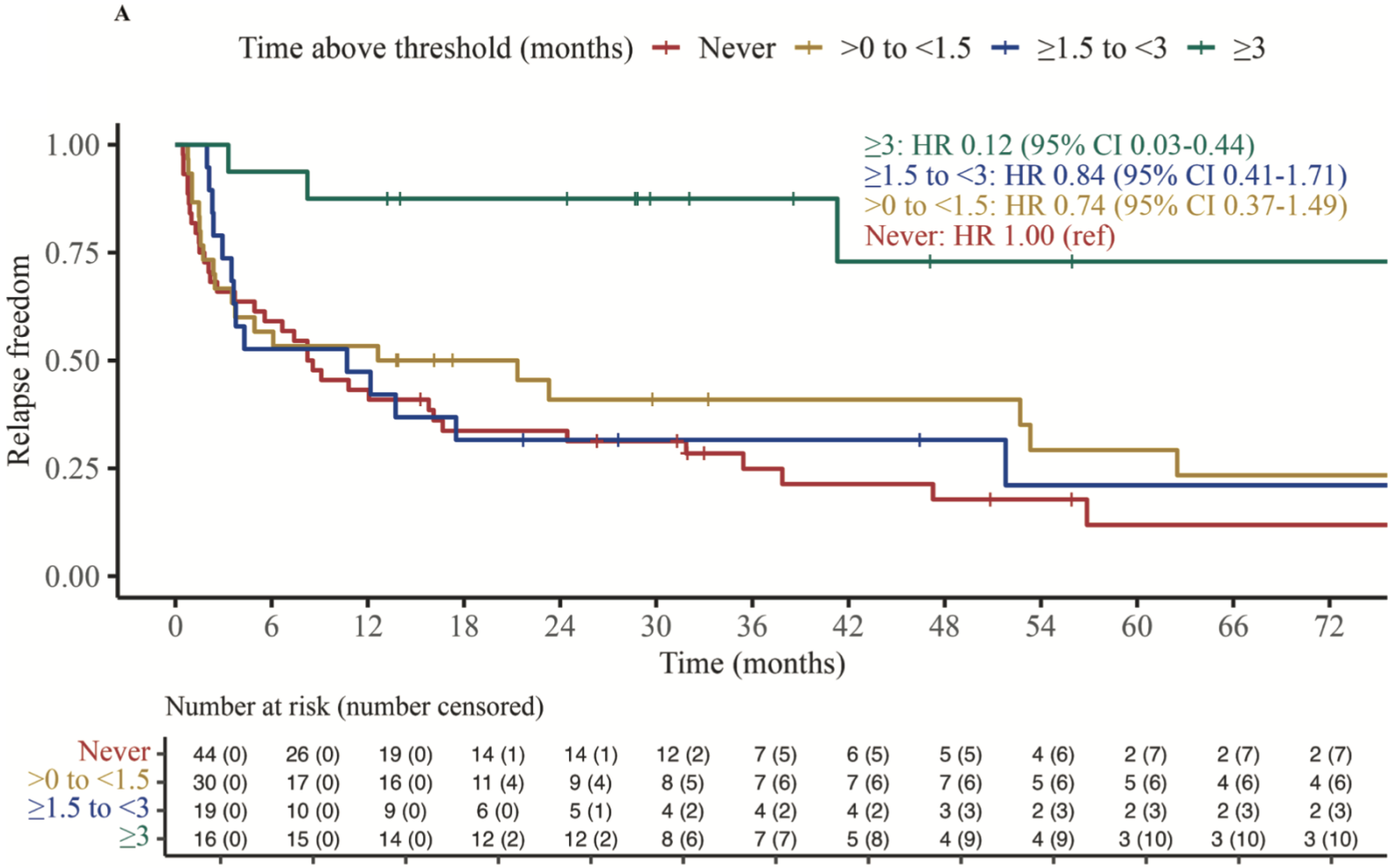

JAMA Neurology | Review

Differential Diagnosis of Suspected Multiple Sclerosis in Pediatric and Late-Onset Populations
A Review

Le H. Hua, MD; Andrew J. Solomon, MD; Silvia Tenembaum, MD; Antonio Scalfari, MD, PhD; Àlex Rovira, MD; Kevin Rostasy, MD; Scott D. Newsome, DO; Ruth Ann Marrie, MD, PhD; Melinda Magyari, MD, PhD; Orhun Kantarci, MD; Bernhard Hemmer, MD; Cheryl Hemingway, PhD; Mary Pat Harnegie, MLIS; Jennifer S. Graves, MD, PhD; Jeffrey A. Cohen, MD; Riley Bove, MD; Brenda Banwell, MD; John R. Corboy, MD; Emmanuelle Waubant, MD, PhD

IMPORTANCE While the typical onset of multiple sclerosis (MS) occurs in early adulthood, 2% to 10% of cases initially present prior to age 18 years, and approximately 5% after age 50 years. Guidance on approaches to differential diagnosis in suspected MS specific to these 2 age groups is needed.

OBSERVATIONS There are unique biological factors in children younger than 18 years and in adults older than age 50 years compared to typical adult-onset MS. These biological differences, particularly immunological and hormonal, may influence the clinical presentation of MS, resilience to neuronal injury, and differential diagnosis. While mimics of MS at the typical age at onset have been described, a comprehensive approach focused on the younger and older ends of the age spectrum has not been previously published.

CONCLUSIONS AND RELEVANCE An international committee of MS experts in pediatric and adult MS was formed to provide consensus guidance on diagnostic approaches and key clinical and paraclinical red flags for non-MS diagnosis in children and older adults.

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Supplemental content

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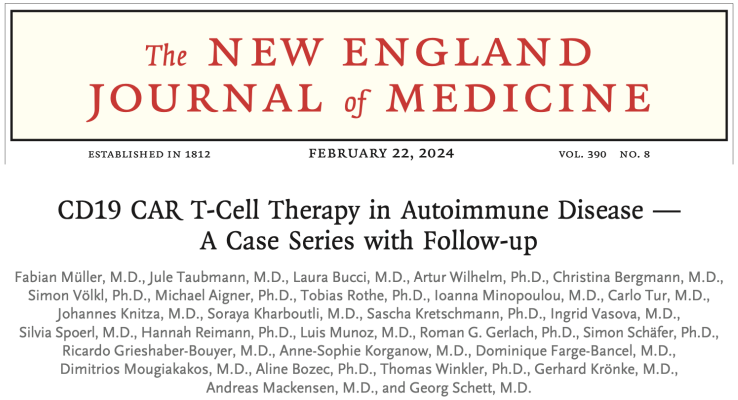

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- CAR-T-Zellen: Behandlung von malignen B-Zell-Erkrankungen.
- T-Zellen werden aus dem Blut des Betroffenen heraussortiert und mit Hilfe eines viralen Vektors gentechnisch mit einem chimären Antigenrezeptor (CAR), (z.B. B-Zell-Epitop CD 19) beladen.
- Nach Chemotherapie werden die CAR-T-Zellen zurückgegeben.
- CAR-T-Zellen können expandieren, das Zielantigen erkennen und zerstören.
- CD19-CAR-T-Zellen wurden erfolgreich bei SLE (Müller et al., 2024), MG und Stiff-Person Syndrom (Haghikia et al., 2023) eingesetzt.
- Erste Behandlungen von insgesamt 5 MS Betroffenen (Konitsioti et al., 2024).

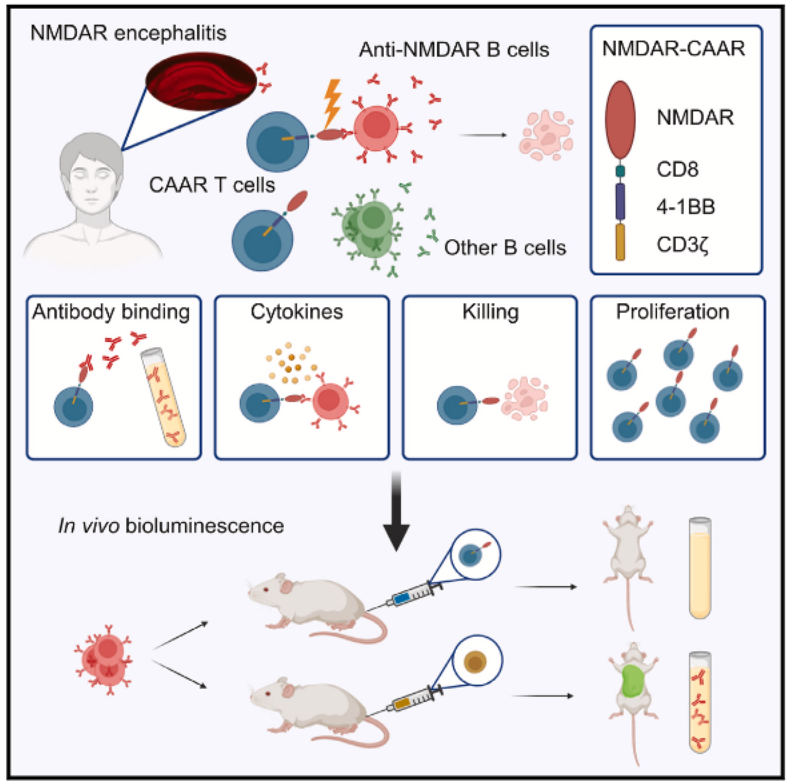
CAR-T-Zellen-Behandlungsansatz für die Behandlung einer schweren NMDAR-Enzephalitis:

Cell

Article

Chimeric autoantibody receptor T cells deplete NMDA receptor-specific B cells

Graphical abstract



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Dietmar Schmitz, Harald Prüss

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In brief

NMDAR-CAAR T cells precisely target an array of pathogenic B cells associated with NMDAR receptor encephalitis.

Highlights

- NMDAR-CAARs recognize a panel of patient-derived NMDAR autoantibodies
- Cytotoxicity against target cells expressing anti-NMDAR B cell receptors *in vitro*
- *In vivo* depletion of anti-NMDAR B cell line and reduction of autoantibody levels
- No histopathological signs of off-target toxicity by NMDAR-CAAR T cells

Neuro-inflammation

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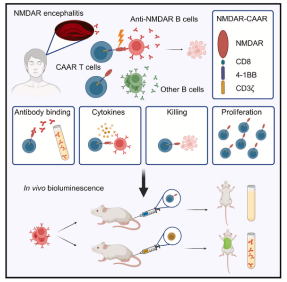

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Supplemental content

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